NeuroRehabilitation

A Multidisciplinary Approach

Dr.V.C.Jacob (PT)
Dr. Hema Biju (OT)
Dr. Alok Sharma

Published By:
NeuroGen, Brain and Spine Institute, Mumbai
Center for Stem Cell Therapy & Neurorehabilitation
www.neurogen.in

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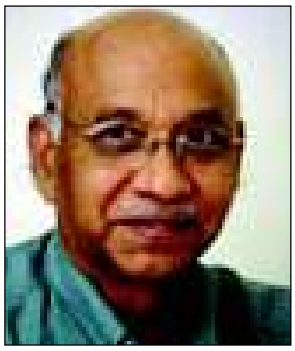

Dr. V.C. Jacob (PT) has been the Deputy Director of NeuroGen Brain and Spine Institute since its inception and is currently the Head of Neurorehabilitation at NeuroGen. He is a Trustee of Nina Foundation which is an NGO set up for the rehabilitation of spinal cord injury persons. He has been a physiotherapist at LTMG Hospital, Sion, Mumbai for 35 years and is now retired. For a period of 38 years, from 1968 to 2006 he was part of the Executive Committee of the Paraplegic Foundation, a charitable organization for the comprehensive care of the paraplegics. He has also organized several sports events for these paraplegics on a national level. In addition, he has also served as President of the Indian Association of Physiotherapists in the past. Apart from all these activities, Dr. Jacob has been involved in disaster management during calamities such as the catastrophic earthquake at Latur and also in Gujarat. He is wholeheartedly committed to the cause of Rehabilitation of patients suffering from neurological disorders.

Dr. Hema Biju (OT) is currently working as Head of Occupational Therapy Department at NeuroGen Brain and Spine Institute which is engaged in cutting edge research in the field of Neuroregenerative rehabilitation involving stem cell therapy.

With over 17 years of national and international work experience in the varied fields within Occupational Therapy, she has specialized in the field of neurological rehabilitation.

Having practiced in the U.S.A as a registered and licensed Occupational Therapist, she is currently associated as a consultant with numerous hospitals in Mumbai and was instrumental in setting up the rehabilitation departments at several of them .

Since 2002, along with her professional career, she has also been continuing her pursuit in the field of academics and research.

Dr. Alok Sharma is a Neurosurgeon who is presently the Professor & Head of Department of Neurosurgery at the LTMG Hospital & LTM Medical College in Sion, Mumbaias well as the Director of the NeuroGen Brain & Spine Institute in Chembur and Consultant Neurosurgeon at the Fortis Hospital in Mulund. He has been committed to both basic as well as clinical research in attempting to find an answer to the problems of paralysis and neurological deficits that occur following injury and diseases of the nervous system. He completed his graduate and post graduate studies from the Seth G.S. Medical College and KEM Hospital of Mumbai University. Subsequently he did fellowships in two departments that were the first to do Neural Transplantation.

In 1995, he worked at the Karolinska Hospital in Stockholm Sweden, where Neural transplantation was done for the first time in the world and in 1998 worked at the University of Colorado Health Sciences Center in Denver, USA where the world's first randomized trial for fetal cell transplantation was done for Parkinson's Disease. Its his life's mission and passion to bring about regeneration within the nervous system. He setup the stem cell and genetic research laboratory at the LTMG hospital which was the first of its type in Mumbai. He is a Neurosurgeon, Medical teacher and Scientist attempting to combine the best of science, medicine and humanity to alleviate the suffering of patients with neurological disorders. He is a staunch believer that stem cell therapy can relieve a lot of human suffering of neurological patients and makes every attempt to popularize this new approach amongst the medical community. This book is one such attempt for the same purpose. He can be reached at **alok276@gmail.com or Ph: +91 22 25283706, +91 9820046663**

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ISBN 81-86876-08-1

Cover design by

Sunakshi Savla & Sonam Gupta

Published by

NeuroGen Brain and Spine Institute

Printed by

SUREKHA PRESS
A-20, Shalimar Industrial Estate,
Matunga Labour camp,
Matunga, Mumbai 400 019.

Price : ₹ 2000

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This book is dedicated to
all rehabilitative therapists,
physicians, care-takers,
institute, rehabilitative centers,
NGOs, societies who have devoted
their lives looking after
the neurological and
musculoskeletal disease
and injuries.

Acknowledgement

Publishing a book of this nature could not have materialized had it not been from the valuable contribution of all the authors who have spent their valuable time and energy. Most of them are from the staff of NeuroGen Brain & Spine Institute and the visiting consultants from various disciplines. So rightly we can say that it is an effort of a multi disciplinary team. We acknowledge our sincere appreciation to all of them. We also thank all our juniors and family members who have also extended a helping hand in whatever way they could.

Dr. V. C. Jacob
Dr. Hema Biju
Dr. Alok Sharma

Preface

NeuroRehabilitation - A Renaissance

This book (which is the first of its kind published from India) has been written for the following purposes:-

1] To provide in one source a comprehensive overview of all aspects of NeuroRehabilitation

2] To provide both undergraduate and postgraduate students of physiotherapy and occupational therapy with all the information they will need from this field for their examinations.

3] To make available to our younger therapists the knowledge and wisdom of years of experience of the senior authors and contributors of this book.

4] To stimulate younger rehabilitation therapists to take up NeuroRehabilitation as a career choice.

5] To emphasize the importance of a multidisciplinary team approach in NeuroRehabilitation.

6] To introduce students, teachers, junior and senior practicing rehabilitation therapists to the fascinating possibilities of clinical improvements that can occur using Neuro-Regenerative-Rehabilitation-Therapy (which is a combination of Stem cell therapy with NeuroRehabilitation)

Of all the injuries and diseases that have afflicted mankind, those of the nervous system have proved to be the most difficult to treat. Whereas modern medical advances have significantly impacted the survival from infectious diseases and improved the longevity from life threatening cardiological and other critical illnesses, the management of neurological problems has not been as successful. There are still many neurological conditions where no drugs were available and where there are no further neurosurgical options. In such cases the only options for the patients to improve the quality of their lives was rehabilitation. But even in rehabilitation of these conditions, things were not so simple. Having to deal with patients who had no movements or limb strength when combined with other aspects such as spasticity, contractures, loss of sensations, cognitive impairment etc made the rehabilitation process very difficult. The work required to rehabilitate these patients was very physically intensive, requiring many hours of laborious work over long periods of time. The results after all this hard work were most often not upto the patients and their relatives satisfaction since what the patients would want is to start walking normally again and this was in most cases just not possible. The subsequent reluctance of patients to pay for the services appropriate to the efforts that were put in became another factor.

All these facts (the complicated nature of the rehabilitation, the poor response, the lack of willingness to pay) resulted in most younger therapists opting for the less physically demanding and more financially rewarding other options in rehabilitation that were also available more commonly. NeuroRehabilitation in its original sense was there becoming a dying science and skill. Another limiting aspect of treating this was the artificial barriers we had put up between the different branches of rehabilitation mainly physiotherapy and occupational therapy. We all tended to look at the patients from our specialty point of view and this often times did not let us see the whole picture. This was in a sense unfortunate since this was one aspect of rehabilitation where a real difference could be made.

However all this is now undergoing a major transformation. The availability of regenerative medicine using healthy cells (such as Stem cells) to replace damaged cells has opened up a entire new world of treatment options giving fresh hope to millions of patients who had given up on any hope for improvement. What Stem cells do is that they initiate a process of repair, replacement and regeneration of the damaged cells by a process of release of various growth factors, increased blood supply and cellular replacement. This sort of reenergizes the nervous and musculoskeletal system. However this is only part of the improvement process. The real functional changes in the patients lives come from rehabilitation. What the Stem cell therapy does is that it makes the rehabilitation process more effective and productive. Improvements not seen earlier just with rehabilitation are now visible. This combination of Regenerative therapy and NeuroRehabilitation has therefore opened up an entire new world of opportunities for

rehabilitation therapists. Not only is this work exciting and challenging and on the frontline of modern medicine, it is also very rewarding in all aspects i.e. clinically, academically as well as materially. A entire group of patients who had given up on rehabilitation since they were not getting encouraging improvements are now coming back to rehabilitation. The newer fancier corporate hospitals that are just about introducing stem cell therapy are now seeing rehabilitation in a new way. This is an incredible opportunity. A field that had been forgotten and given up as not being useful or remunerative enough is now being looked upon with a whole new interest. It's a rebirth or a renaissance of sorts. Much like how the older radio had almost died in the 80's and 90's with the advent of television but had a rebirth in the last decade with the introduction of FM radio.

In this book all the different aspects of rehabilitation (physiotherapy, occupational therapy, speech therapy, counseling, clinical and surgical aspects) have all been put together in individual chapters that are focused on an individual diseases or medical condition. This is intentional and meant to highlight the multidisciplinary approach to NeuroRehabilitation. Various theoretical aspects have been covered along with practical advice and suggestions on management. Extensive literature reviews have been done to incorporate the best of whatever is available from all across the world. Valuable individual information gathered from years of experience have also been incorporated. This is important. To highlight this let me state that at the NeuroGen Brain and spine institute we receive patients regularly from all over the world. These patients have already been to the best rehabilitation centers in the USA and elsewhere. And yet despite this virtually all the patients without any exceptions have one thing to say. " that they have never received the kind of rehabilitation in all the centers they had been to, which they received at NeuroGen". What is the difference between what is offered at NeuroGen and what is offered in the bigger rehabilitation centers all over the world. At NeuroGen there is the human touch. There are virtually no fancy electronic machines or equipment. Our rehabilitation makes a difference because its more hands on with the rehabilitation therapist working intensely and closely with the patients instead of depending on machines and watching from a distance.

This is work that requires dedication, commitment, hard work and perseverance. The joy of this treatment lies not as much in the money but in the small small functional improvements seen in the patients and their gratefulness and gratitude for the same. There were very few therapists who have dedicated their lives to this work. And these therapists belong to an older generation. It was important that the wisdom and knowledge of their years of experience was passed on to today's younger generation. This is therefore one of the important intentions of this book.

It is fortunate for the readers of this book that through this book they will get together in one place the lifetimes wisdom and experience of the senior author of this book Dr. V. C. Jacob. Dr. Jacob, who as you all well know, is one of the countries senior most and highly respected physiotherapist who had devoted his whole life to NeuroRehabilitation through his clinical and teaching work at the LTM Medical college and the charitable work at the Paraplegic Foundation. Inputs from the occupational therapy aspects of the work come from Dr. Hema Biju who has worked for several years in the USA before coming back to India. All the other contributors and authors are experts in their respective fields and we are grateful to their contributions which have resulted in this unique collection of knowledge and wisdom. On behalf of the Neurogen Brain and Spine Institute, I take this opportunity to thank all the authors and contributors. Grateful acknowledgement also goes out to all the members of the NeuroGen team who have despite great difficulties helped and supported all the work that had to be done to make this book a reality. The biggest thanks of course goes to Dr. Nancy Thomas, the scientific and editorial coordinator, who had worked tirelessly for several months to put all this wonderful information together. This book could not have been put together without her sincere efforts. All of us from Neurogen as well as all the contributors now place this book in your hands with the hope that it will inform, enrich an contribute to your lives and the lives of your patients in some important way. Should any of you need any more information please feel free to reach out to us. We are always more than happy to help any of you in any way that your desire. Wish you all students as well a practicing therapists happy reading and happy learning.

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Contents

Section 5: Stem Cell Therapy

Section 1

Spine

Ch.1 Spinal Cord Injury

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INTRODUCTION

"So many of our dreams at first seem impossible, then they seem improbable and when we summon the will they soon become inevitable."

– Christopher Reeve

Rehabilitation of the spinal cord injured individuals is by far one of the most challenging fields for the rehabilitation experts. Till today there is very little to claim as a panacea for the spinal cord injury (SCI) or regeneration of the damaged spinal cord. However the researchers all over the world find great hope in stem cell therapy (SCT) and it has been proved beyond doubt that the results with SCT coupled with good comprehensive rehabilitation is better than just giving SCT. Therefore rehabilitation is being given a better facelift by SCT.

As everyone knows that the role of all the members of the rehabilitation team is equally important and goes hand in hand with each other. No one can claim that other's role is less important. Perhaps in one case one of the team member's role may be more significant than other's, whereas it may be the other member's role in yet another case. But overall each one has to put in his best foot forward to achieve the desired goal. Therefore it always pays if we can achieve the co-operation and co-ordination of all the team members. It is not always possible to get everyone together all the time. But a good network of communication between the team would bring in good results.

In Latin, rehabilitare means make fit again.

Definition:

Rehabilitation is defined as

"Rehabilitation is a treatment or treatments designed to facilitate the process of recovery from injury, illness or disease to as normal a condition as possible."

– Medical Dictionary

In other words, it is the process of restoration of skills by a person who has had an illness or injury so as to regain maximum self sufficiency and function in a normal or as near as normal manner as possible. Having to give up independence and becoming totally dependent on others is a frustrating and debilitating affair for a person with SCI. with a unified team approach, a person with a SCI himself or herself would benefit in gaining not only physical independence but also economic and social acceptance. The relatives and the individual with SCI are as much a part of the team as professionals as eventually they will have to share the responsibility for the success in getting back the person into the society and not becoming a burden to the society.

Dr. Ketna Mehta. PhD., a SCI person herself says, it starts not just from movements of the limb done in an attempt to gain mobility and avoid contractures and other related complications but from mobilization of the mind by constant motivation so as to prevent from developing the worst ever contracture called "depression". Having to give up independence to a totally dependent life, all of a sudden can be largely frustrating and devastating. It definitely is a good growth medium for harmful parasitic thoughts of hopelessness, helplessness and worthlessness.

To help of patient come out of this dreadful situation, it requires effort from a team of members, which is rightly called as a multidisciplinary team.

MULTI DISCIPLINARY TEAM

An ideal multi disciplinary team should comprise of all of the following members:

1. Spinal surgeon
2. Urologist
3. Physiotherapist
4. Occupational therapist
5. Medical social worker
6. Psychological counselor
7. Rehabilitation nurse
8. Care givers

However the team would not be complete without the SCI person himself whose willingness to co-operate with the team members alone would make the entire process easier. Ideally the team members should take collective decisions in setting a goal or outcome measures, which is nothing but a guideline to achieve the expected level of recovery. For this each member has to set a realistic achievable goal. The patient's expectation may be too much, so also of the team members. The expectation may be realistic only if one takes into consideration the medical problems and the psychological status of the patient.

It is advisable and ethical to involve the patient in planning the treatment pathway from time to time. However very few patients have the technical knowledge regarding the management of their condition. Hence the therapists have to make efforts to explain the advantages and disadvantages of various approaches. Many a times it is beneficial to show other rehabilitated patients who have successfully undergone the rehab process and how they have solved their own problems, thereby living a productive life within their face of limitations. Before taking the final decision it is mandatory on the part of the therapist to consult other team members and each one of them has to make a thorough evaluation of the motor skills, functional skills, home assessment, ADL, etc. Rehab begins from the day of the injury. All the interventions planned for the patient from day one focuses on prevention of further damage to the spinal cord and to preserve the functional capabilities, most vital to save the patient. So a brief review of the SCI is discussed below. In the initial phase after the SCI, surgeon plays a vital role especially to decompress and stabilize the spine so that the actual physical rehabilitation can commence at the earliest. How big is the problem? To put light to the intensity of the SCI issue, the epidemiology is stated below.

Epidemiology:

- Before World War 2, the average life expectancy of a SCI person was just 2yrs. With the advent of antibiotics and improved therapeutic measures, the management of complications of SCI has improved remarkably. Acute care has also improved considerably. After the rehabilitation, they learn to live rather independently or with minimal help of the family members.
- The annual incidence of SCI has not changed but since life expectancy has improved considerably, the number of living SCI patients have increased. Though the prevalence is not much compared to the population, their presence is significant.

Causes and risk factors:

Spinal cord trauma can be caused by any number of injuries to the spine. They can result from motor vehicle accidents, falls, sports injuries (particularly diving into shallow water), industrial accidents, gunshot wounds, assault, and other causes.

A minor injury can cause spinal cord trauma if the spine is weakened (such as from **Rheumatoid Arthritis or Osteoporosis**) or if the spinal canal protecting the spinal cord has become too narrow (spinal stenosis) due to the normal aging process.

Direct injury, such as cuts, can occur to the spinal cord, particularly if the bones or the disks have been damaged. Fragments of bone (for example, from broken vertebrae) or fragments of metal (such as from a traffic accident or gunshot) can cut or damage the spinal cord.

Direct damage can also occur if the spinal cord is pulled, pressed sideways, or compressed. This may occur if the head, neck, or back are twisted abnormally during an accident or injury.

Bleeding, fluid accumulation, and swelling can occur inside the spinal cord or outside the spinal cord (but within the spinal canal). The accumulation of blood or fluid can compress the spinal cord and damage it.

Most spinal cord trauma happens to young, healthy individuals. Men ages 15 - 35 are most commonly affected. The death rate tends to be higher in young children with spinal injuries.

Risk factors include participating in risky physical activities, not wearing protective gear during work or play, or diving into shallow water.

Older people with weakened spines (from osteoporosis) may be more likely to have a spinal cord injury. Patients who have other medical problems that make them prone to falling from weakness or clumsiness (from stroke, for example) may also be more susceptible.

Symptoms:

Symptoms vary somewhat depending on the location of the injury. Spinal cord injury causes weakness and sensory loss at and below the point

of the injury. The severity of symptoms depends on whether the entire cord is severely injured (complete) or only partially injured (incomplete).

The spinal cord doesn't go below the 1st lumbar vertebra, so injuries at and below this level do not cause spinal cord injury. However, they may cause "cauda equina syndrome" -- injury to the nerve roots in this area.

1. CERVICAL LEVEL INJURIES

When spinal cord injuries occur in the neck area, symptoms can affect the arms, legs, and middle of the body. The symptoms may occur on one or both sides of the body. Symptoms can include:

- Breathing difficulties (from paralysis of the breathing muscles, if the injury is high up in the neck)
- Loss of normal bowel and bladder control (may include constipation, incontinence, bladder spasms)
- Numbness
- Sensory changes
- Spasticity (increased muscle tone)
- Pain
- Weakness, paralysis

2. THORACIC LEVEL INJURIES

When spinal injuries occur at thoracic level, symptoms can affect the legs:

- Loss of normal bowel and bladder control (may include constipation, incontinence, bladder spasms
- Numbness
- Sensory changes
- Spasticity (increased muscle tone)
- Pain
- Weakness, paralysis

Injuries to the high thoracic spinal cord may also result in blood pressure problems, abnormal sweating, and trouble maintaining normal body temperature.

3. LUMBO SACRAL INJURIES

When spinal injuries occur at the lumbo sacral level, varying degrees of symptoms can affect one or both legs, as well as the muscles that control bladder and bowel:

- Loss of normal bowel and bladder control (constipation, leakage, and bladder spasms)
- Numbness
- Pain
- Sensory changes
- Spasticity (increased muscle tone)
- Weakness and paralysis

COMPLETE SPINAL CORD INJURY

In complete lesion, there is no sensory or motor function below the level of lesion. It is caused by a complete transection or severe compression or extensive vascular impairment (due to lack of blood supply to the spinal cord).

INCOMPLETE SPINAL CORD INJURY

An incomplete spinal cord injury is the term used to describe damage to the spinal cord that is not absolute. The incomplete injury will vary enormously from person to person and will be entirely dependant on the way the spinal cord has been compromised.

TYPES OF INCOMPLETE SPINAL CORD INJURY

- **Central Cord Syndrome:**

This is the commonest of the incomplete syndrome (CCS). It is characterized by weakness in upper more than lower limbs with sacral sparing. In addition, sensory loss below the lesion with partial sensory deficit. Schneidan and others noted that the etiologic factor was hyperextension with simultaneous compression of the cord by either anterior osteophytes and posterior impingement caused by buckling of the ligmentum flavum. It is more common with elderly group of patients with cervical spondylosis, than with younger group of traumatic patients. Pathology of the CCS is probably due to the injury to the central part of the cord. Depending on the severity of the lesion both upper and lower limbs getting affected with upper more than lower because in cervical fibers are more centrally located compared to dorsal, lumbar and sacral spine.

Quencer etal by MRI study found that CCS is predominately a white matter peripheral injury and not intramedullary hemorrhage. CCS usually have good prognosis. The pattern of recovery also seen starting with lower limbs followed by bladder bowel function and then Upper limb starting from

proximal to distal and finally intrinsic muscle in Upper extremity. However younger patients recover faster than elderly patients. Similarly bladder recovery is also seen in younger more than elderly patients. Patients with ASIA scale 'D' are able to recover well and are ambulatory.

- **Brown –Sequard Syndrome (BSS) :**

It occurs in cases of stab injuries with ipsilateral loss of all sensory modalities at the level of injury:

- With ipsilateral flaccid motor paralysis.
- With ipsilateral loss of position sense and vibration sense below the lesion.
- With ipsilateral loss of motor function below the lesion.
- With contralateral loss of pain and temperature sensation.

This is because of the crossing of spinothalamic fibres which carry pain and temperature fibres whereas corticospinal tracts cross in the brainstem Brown sequard plus syndrome (BSPS) is much more common compared to pure BSS i.e. with ipsilateral motor weakness (hemiplegia) and contralateral hemiagnosia.

Numerous nontraumatic causes of Brown-Séquard syndrome have also been reported, including the following:

- Tumor (primary or metastatic)
- Multiple sclerosis
- Disk herniation
- Herniation of the spinal cord through a dural defect (idiopathic or posttraumatic)
- Epidural hematoma
- Vertebral artery dissection
- Transverse myelitis
- Radiation
- Type II decompression sickness
- Intravenous drug use

- **Anterior Cord Syndrome:**

Damage to the anterior 2/3 of the spinal cord preserving the posterior column due to retropulsed bone fragments either due to direct injury to the anterior cord or anterior spinal artery which supply blood to the anterior part of the spinal cord.(due to aortic or cardiac surgery, embolism, angioplasty etc.) Clinically they present loss of motor and pinprick sensation with relative preservation of proprioception, deep pressure sensation. Usually the prognosis is very poor with poor motor recovery and coordination.

- **Conus medullaries and Cauda equina lesion:**

It is the terminal part of the cord which lies at the inferior aspect of L1 vertebrae. The segment above conus medullaris is termed as epiconus consisting of cord segments L4-S1.Nerve roots then travel from conus medullaries caudal as the cauda equine.

Injuries to the conus will present LMN deficits of anal sphincter and bladder. If L3-S2 nerve roots are not affected, motor strength in the legs and feet may not be affected. If the roots are affected, it will give LMN damage with diminished reflexes.

Injuries below L1 usually give a LMN weakness supplying the lumbar and sacral segments causing muscle atrophy with bladder bowel involvement with loss of plantar reflexes, anal reflexes and bulbocaverous reflexes and impotence.

Cauda equine lesions have better prognosis than other SCI.

Fracture L1 may result in conus damage whereas fracture L2 downwards will affect cauda equina.

Isolated conus injury is very rare.

Incidence of conus and cauda equina injuries are common.

DIAGNOSTIC MEASURES:

Spinal cord injury is a medical emergency requiring immediate attention.

The health care provider will perform a physical exam, including a neurological exam. This will help identify the exact location of the injury, if it is not already known. Some of the person's reflexes may be abnormal or absent. Once swelling goes down, some reflexes may slowly recover.

The following tests may be ordered:

- A CT scan or MRI of the spine may show the location and extent of the damage and reveal problems such as blood clots (hematomas).
- Myelogram may be necessary in rare cases.
- Somatosensory evoked potential (SSEP) testing or magnetic stimulation may show if nerve signals can pass through the spinal cord.

- Spine x-rays may show fracture or damage to the bones of the spine.

As discussed earlier, the management of SCI begins at the location of the trauma. The initial focus should be to reach the patient immediately to hospital with utmost care not to aggravate the injury (well supported horizontal position of the spine).

MEDICAL MANAGEMENT:

A spinal cord trauma is a medical emergency requiring immediate treatment to reduce the long-term effects. The time between the injury and treatment is a critical factor affecting the eventual outcome. Initial treatment of patients with cord injury focuses on two aspects - preventing further damage and resuscitation.

Prehospital Management:

Spinal cord injury isn't always obvious. Mechanism of injury, pain in the vertebral column, or neurologic symptoms gives a clue to the emergency medical personnel regarding suspected spinal injury. Injuries involving the head, pelvis and those resulting from falling from heights should be suspected for spinal cord damage.

Emergency medical personnel stabilize and immobilize the spine at the scene of injury and transport the patient to the emergency department (ED) taking care in not moving the spine. Many a times it is the bad lifting and transport that causes damage to the spinal cord.

Emergency Department Management:

A. **ABC Resuscitation:** Airway, breathing and circulation may be compromised because of spinal cord injury or associated injuries. Resuscitation is aimed at airway maintenance, adequate oxygen saturation of peripheral blood, restoring blood pressure to acceptable limits, preventing bradycardia, done simultaneously to prevent any ischemic damage to the already compromised cord.

a. Airway management: Spinal cord injury makes the assessment and management of airway complex and difficult. Immobilization of cervical spine must be continued in neutral alignment during airway assessment and management.

   I. Restoring Airway Patency: Oral secretions are cleared to maintain airway patency and to prevent aspiration.

   II. Maintaining Airway Patency: To keep the airway patent, modified jaw thrust and insertion of an oral airway is required in some patients and, endotracheal intubation in others. When indicated intubation should be done carefully in these patients to avoid spine movement and further spinal cord injury.

b. **Circulation Management:** In patients with acute spinal cord injury, shock may be hemorrhagic and/or neurogenic. In these patients, a diligent search for occult sources of hemorrhage must be made before making the diagnosis of neurogenic shock as these patients have high incidence of associated injuries.

   I. **Excluding Haemorrhagic shock:** Computed tomography (CT) scan or X-Ray will reveal the most common sources of occult hemorrhage like chest, abdominal, retroperitoneal injuries and fractures of the pelvis or long-bones.

   II. **Neurogenic shock and its management:** History of spinal cord injury with exclusion of haemorrhage suggests spinal shock.

   i. **Fluid resuscitation:** Fluid resuscitation with isotonic crystalloid solution is the initial treatment of choice for neurogenic shock. These patients are at risk for the acute respiratory distress syndrome (ARDS) and hence volume overload should be avoided with judicious fluid resuscitation.

   ii. **Maintaining blood pressure:** Systolic blood pressure should be maintained above 90 mm Hg and hypotension should be avoided in these patients.

   iii. **Maintaining oxygenation:** Supplemental oxygenation and/or mechanical ventilation should be given to maintain adequate oxygenation and perfusion of the injured spinal cord.

   iv. **Monitoring heart rate:** Heart rate should be 60-100 beats per minute (bpm) in normal sinus rhythm. Atropine may be given for treatment of hemodynamically significant bradycardia.

   v. **Maintaining urine output:** Foley catheter

should be inserted to decompress the neurogenic bladder. Urine output should be monitoted and maintained more than 30 mL / h.

vi. **Inotropic support:** Dopamine or norepinephrine is should be reserved for patients who have decreased urinary output despite adequate fluid resuscitation. A low dose of dopamine in the 2- to 5-mcg / kg / min range is usually sufficient.

vii. **Prevention of hypothermia:** Hypothermia should be prevented in these patients.

B. **Assessment and management of associated injuries:** Twenty five percent of patients with spinal cord injury have associated head injury. A thorough evaluation for intracranial injury should be done in the presence of amnesia, external signs of head injury or basilar skull fracture, focal neurologic deficits, associated alcohol intoxication or drug abuse, and a history of loss of consciousness.

Corticosteroid Therapy:

Methyl prednisolone has been shown by the NASCIS-II (National Acute Spinal Cord Injury Studies) trial to be significantly beneficial in causing favorable neurological outcome if administered early enough (within 8 hours) of the primary insult.It appears to work by reducing damage to nerve cells and decreasing inflammation near the site of injury. To be effective corticosteroids should be given within 8 hours of injury. The recommended dose is 30mg / kg bolus to be administered over 15 minutes and after a 45-minute pause 5.4 mg / kg / hr over the next 23 hours.

Nesathurai and Shanker questioned the validity of the results of NASCIS trial. These authors cited concerns about the randomization, clinical endpoints and statistical analysis used in the study. In addition, the investigators noted that even if the benefits of steroid therapy were valid, the clinical gains were questionable. The steroid therapy is not free of side effects. An increased incidence of infection and avascular necrosis has been documented.

The Congress of Neurological Surgeons (CNS) has stated that steroid therapy "should only be undertaken with the knowledge that the evidence suggesting harmful side effects is more consistent than any suggestion of clinical benefit." Every hospital should have a policy regarding administration of steroids to spinal cord injury patients.

Monosialotetrahexosyl ganglioside (GM-1): GM-1 administered following acute spinal cord injury as an adjunct after the administration of methylprednisolone may be of benefit.

Spinal traction: Spinal traction is a more conservative and less invasive approach to allow the bones to heal naturally without surgery. Traction prevents movement of the spine. The skull may be held in place with tongs (metal braces placed in the skull and attached to traction weights or to a harness on the body). The spine braces may need to be worn for a long time.

Surgical Management:

Surgery may be required in the treatment of spinal cord injury patients. There are three major goals of surgery-

A. Spinal decompression: This involves removal of fluid tissue, bone fragments, disk fragments, or foreign objects that presses on the spinal cord.

B. Prevention of tethering: Neurological disorders related to pulling of the spinal cord at the base of the spinal canal causing stretching and damage. Surgery may prevent tethetering of the cord. [14]

C. Stabilization of the spine: The weakened vertebrae from fracture may not be capable of supporting the normal weight from the body and protect the spinal cord. Spinal instrumentation and fusion can be used to provide permanent stability to the spinal column. These procedures correct, join, and solidify the level where a spinal element has been damaged. A combination of metal screws, rods, plates or cage may be necessary to help hold the vertebrae together and stabilize them until the bones heal.

Timing of surgery: There are no established standards regarding when to operate the patients with spinal cord injury because of the lack of well designed studies. Emergency decompression of the spinal is suggested in the following indications-

1. Acute spinal cord injury with progressive neurologic deterioration, facet dislocation, or bilateral locked facets.

2. Spinal nerve impingement with progressive radiculopathy.
3. Extradural lesions such as epidural hematomas or abscesses.
4. Cauda equina syndrome.

Type of surgery:

The type of surgery performed (anterior vs. posterior), distraction forces during surgery, preoperative grade all influence the outcome. The surgery may be-

Spinal decompression:

The major types of surgery for spinal decompression are-

a. Microdiscectomy / Corpectomy
b. Laminectomy

Stabilization of the spine:

Can be achieved by spinal fusion with or without instrumentation.

1. In the Craniovertebral junction, stabilization may be achieved by
   a. C1-C2 fixation using pedicle screws / transarticular screw / sublaminar wires
   b. Occipitocervical fixation

Figure 1.1.1

2. In the cervical spine stabilization may be achieved by
   a. Anterior approach using vertebral body plates and screw
   b. Posterior approach using lateral mass screws

Figure 1.1.2 and 1.1.3

3. In the dorsal spine stabilization may be achieved by
   a. Anterior transthoracic approach for interbody cage placement
   b. Posterior approach using transpedicular screw and rod system
3. In the lumbar spine, types of stabilization procedures are-
   a. Anterior lumbar interbody fusion (ALIF) - the disc is accessed from an anterior abdominal incision.
   b. Posterior lumbar interbody fusion (PLIF) - the disc is accessed from a posterior incision. c. Transforaminal lumbar interbody fusion
   c. Transforaminal interbody fusion(TLIF) - the disc is accessed from a posterior incision on one side of the spine. Instrumentation with screws, rods, plates or cages may be required to provide permanent stability to the spinal cord.

Figure 1.1.4

Surgery for prevention of tethering: After complete release and reconstruction of the spinal cord, a Gore-Tex surgical membrane can be placed over the cord and fixed to the lateral dural surface with stay sutures to prevent tethering.

Post operative physiotherapy

Post operative physiotherapy starts immediately after the surgery and is continued during the convalescent period. Respiratory management plays a very important role to prevent post operative complications. Restorative physiotherapy starts immediately after the patient is stabilized.

REHABILITATION OF PATIENTS WITH LESIONS BELOW CERVICAL SPINE (PARAPLEGICS)

The acute stage of rehabilitation is followed by the convalescent stage, where the patient is thoroughly evaluated by the team for further management. The management is based on classification of SCI as per the ASIA Scale.

The American Spinal Injury Association (ASIA) first published an international classification of spinal cord injury in 1982, called the International Standards for Neurological and Functional Classification of Spinal Cord Injury. Now in its sixth edition, the International Standards for Neurological Classification of Spinal Cord Injury (ISNCSCI) is still widely used to document sensory and motor impairments following SCI. It is based on neurological responses, touch and pinprick sensations tested in each dermatome, and strength of ten key muscles on each side of the body, including hip flexion (L2), shoulder shrug (C4), elbow flexion (C5), wrist extension (C6), and elbow extension (C7).Traumatic spinal cord injury is classified into five categories on the ASIA Impairment Scale:

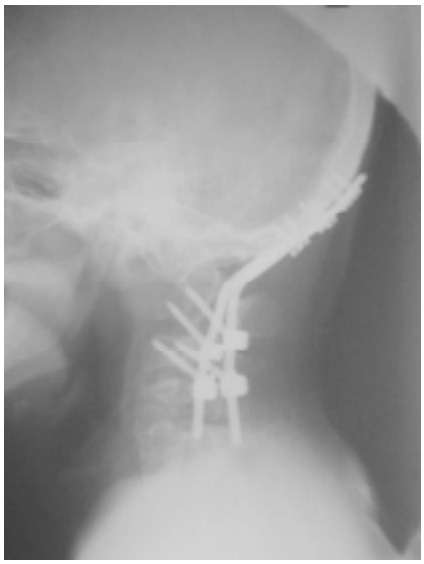


Fig 1: X-ray lateral view CV junction showing occipito cervical fixation.

Fig. 2: Intraoperative picture showing placement of vertebral body plate.

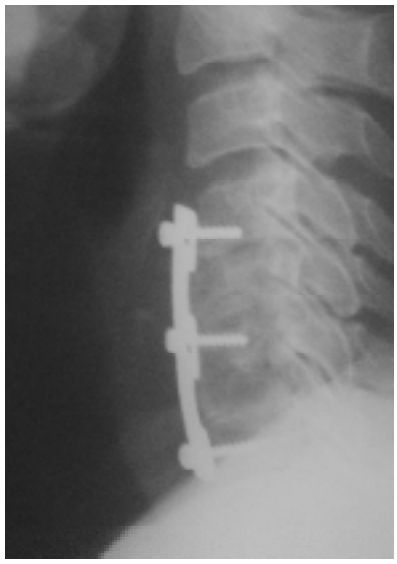


Fig. 3: X-ray lateral view cervical spine showing cervical fixation using vertebral plate and screws.

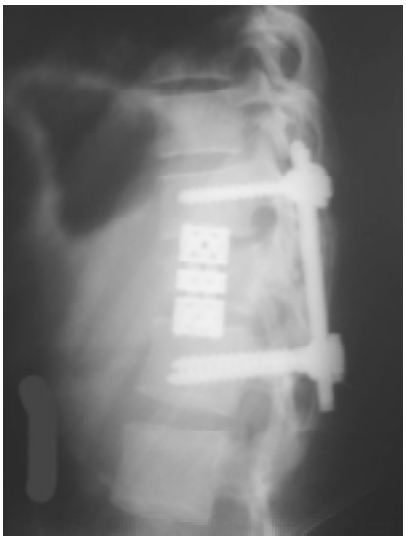


Fig. 4: X-ray lateral view lumbar spine showing instrumentation using transpedicular screws and rods and interbody cage placement.

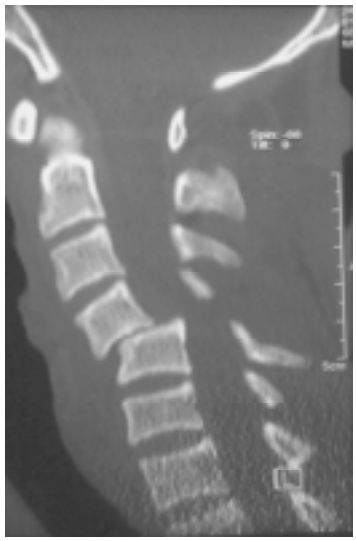


Fig. 1.1.4: CT Cervical spine sagittal view: showing post traumatic subluxation of C4 over C5 with canal compromise.

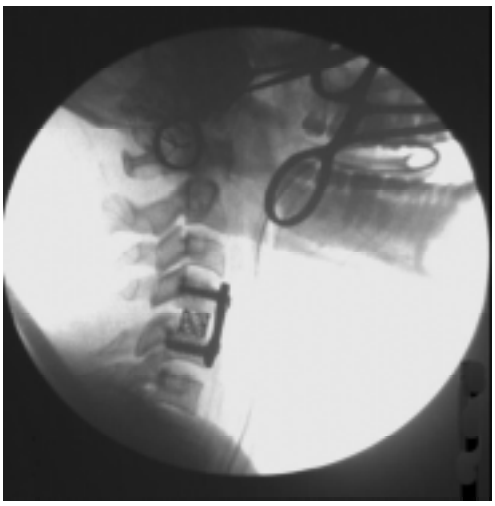


Fig. 1.1.5: Intraoperative image following reduction and stabilization of C4.C5 subluxation.

ASIA IMPAIRMENT SCALE

- A indicates a "complete" spinal cord injury where no motor or sensory function is preserved in the sacral segments S4-S5.
- B indicates an "incomplete" spinal cord injury where sensory but not motor function is preserved below the neurological level and includes the sacral segments S4-S5. This is typically a transient phase and if the person recovers any motor function below the neurological level, that person essentially becomes a motor incomplete, i.e. ASIA C or D.
- C indicates an "incomplete" spinal cord injury where motor function is preserved below the neurological level and more than half of key muscles below the neurological level have a muscle grade of less than 3, which indicates active movement with full range of motion against gravity.
- D indicates an "incomplete" spinal cord injury where motor function is preserved below the neurological level and at least half of the key muscles below the neurological level have a muscle grade of 3 or more.
- E indicates "normal" where motor and sensory scores are normal. Note that it is possible to have spinal cord injury and neurological deficits with completely normal motor and sensory scores.

MODIFIED ASHWORTH SCALE: FOR GRADING TONE

- **Grade 0**: no increased muscle tone
- **Grade 1**: Slight increase in muscle tone, manifested by a catch and release, or by minimal resistance at end of ROM when affected part is moved in flexion or extension.
- **Grade 1+ :** slight increase in muscle tone manifested by a catch, followed by minimal resistance throughout the remainder (<50%) of ROM.
- **Grade 2:** more marked increase in muscle tone through most of ROM and affected part is easily moved.
- **Grade 3:** considerable increase in muscle tone, passive movement is difficult.
- **Grade 4:** limb rigid in flexion or extension.

After evaluation of the client based on the classification, it is vital to set goals or achieving the realistic expected outcome.

Prediction of outcome:

Physical examination is the most important mehtod to predict the outcome.

Initial presence of muscle power would give a clear prediction. The earlier the muscle shows some power the faster muscle could come to functional level i.e above 3.

Within the first 6 months the recovery seen is maximum and the improvement in the first three months is usually more than the next three months.

If one gets back some power initially the chances of improvement is better than the muscles remaining “0”.

However those who have an incomplete injury initially have better prognosis for ambulation. MRI is the most superior radiological test for determining the prognosis. E.g. of a hemorrhage seen initially, the prognosis is poor. Contusion is worse than edema. If no hemorrhage is seen then damage to the cord is incomplete and the prognosis is better. The degree and the extent of edema is also determines the chances of recovery. In other words, along with clinical assessment, MRI can give the prediction better. The other investigations include nerve conduction studies, late responses (H-reflex and F-wave) and somatosensory evoked potentials.

Whatever the prognosis is, rehab has a definite role to play and whenever any scientific advances come, they could try at that stage. Till then the patient has to come to terms and try to become as independent as possible.

Outcome charts

These are merely meant to act as a guide. Perhaps the therapist’s expectations would be too much or too little. So the purpose is to describe the expected levels of safe functions in SCI patients. Considering associated medical problems and psychological status, therapists have to do systematic treatment planning.

Prescription of the equipments:

Things to be considered are:

1. Easy availability.
2. Accessibility
3. Durability

4. Safety
5. Comfort
6. Cost

It is advisable to involve the patients in planning. However very few patients can make their own decisions and therefore therapist has to explain advantages and disadvantages, often trying out different techniques. Perhaps, it will be more beneficial to show other patients who have experienced the same problems and have solved their problems. Many a times it would be essential to set a trial period, trying out trial calipers / shoes. Eventually the therapist can take a final decision in prescribing the necessary equipments. Before taking the final decision therapist has to make a thorough evaluation of motor skills, functional skills, home assessment, ADL and opinions of other team members and taking patients' consent.

Evaluation:

A) Data collection:

1. From patients and relatives.
2. From medical records including X-rays, MRI, and other reports.

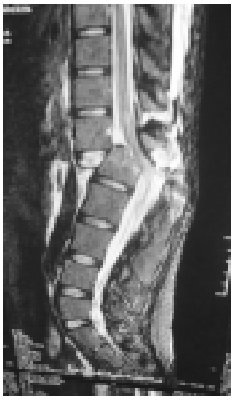


Fig1.MRI of Lumbar Spine

It is essential to explain to patients the need for evaluation and taking proper history so that he actively participates in the actual rehabilitation process.

B) Evaluation Procedures:

- Assessment of strength.
- ROM
- Sensations
- Tone
- Girth measurement

C) Physical Evaluation:

a) Skin Evaluation:

1. Bony prominences and vulnerable area of skin.
2. Scar tissue.
3. Pressure sores.
4. Secondary bad postures because of pressure sores.

b) Sensory Evaluation:

1. Superficial and deep sensations like pain, temperature, touch joint position etc.
2. Patient's awareness about sensory deficits.
3. Implications because of sensory deficits namely burns, bruises lack of coordination because of lack of kinesthetic sense.
4. Damage to skin due to faulty equipments like orthosis.

c) Respiratory Evaluation:

1. General observations:

a) Shape and symmetry of chest.
b) Muscle tone.
c) Wasting.
d) Presence of artificial airways like tracheostomy, intercostals tubes.

Respiratory history and associated medical history may indicate that he may require preventive measures for chest complications. Evaluation of upper and lower chest expansions may give a clue as to which lobes require to be attended. Lack of chest expansion and mobility may lead to decreased vital capacity. (Normal value of chest expansion at xiphoid process should be at least 2 1/2 to3 inches.) Lack of ROM at shoulder level may lead to poor chest expansion. It is essential to have normal flexibility of trunk and hips as it is easier for him to cough effectively. Normal posture of trunk is essential for rib mobility as excessive tightness of ribs would lead to poor Vital capacity.

2. Respiratory Muscle Strength Evaluation:

1. Diaphragm.
2. Abdominals.

3. Accessory muscles like intercostals, scalenes, serratus anterior, pectorals, erector spinae and sternocleidomastoid.

Implications : If abdominal muscles are weak, it may effect in poor coughing because of inability to build up intrathoracic pressure to expel air and therefore bronchial hygiene may be affected very badly.

1. An abdominal binder may be useful.
2. Increased respiratory rate indicate shallow breaths..
3. When abdominals are paralysed, they breathe best in lying.
4. Hypoventilation in patient may also cause drowsiness and irritability. This usually happens among quadriplegics or high level paraplegics because of weakness of accessory muscles.
5. Hypoventilation may cause fainting, tingling and numbness.
6. Poor co ordination of breathing with functional activities may cause lack of endurance.
7. Deep breathing exercises would increase inspiratory capacity which will improve coughing and ventilation of all the lobes thus decreasing respiratory complications.

Personal hygiene

1. Evaluation of cough and Vital Capacity
2. Evaluation of patients' knowledge of postural drainage.

IMPLICATION FOR PATIENTS

1. Vital capacity may be used as an indicator of weakness of respiratory muscles.
2. Decreased vital capacity leads to decreased cough effectiveness.
3. Postural drainage and coughing are very important for a SCI patient as it would prevent infection.
4. Movements would decrease the possibility of accumulation of secretions and infections in the lungs.

ROM of Joints:

- Rolling may be more difficult with overstretched back and trunk muscles.
- Selective tightness of the long flexors of the hand will help in grasp (Tendodesis action).
- Maintaining of adequate ROM is essential for helping the totally dependent patients to move for changing the positions.
- If a patient can be moved easily, there is less chances for pressure sores.

MuscleTone:

1. Mild increase: Resistance for passive stretching with full ROM, function not being affected
2. Moderate increase : More resistance and full ROM is possible but function being affected.
3. Severe increase: Full ROM is not possible and many functional skills impossible.

Description of quality of Tone:

1. Constant or Fluctuating?
2. Is it related to position change?
3. Symmetrical or Asymmetrical?
4. Fluctuation related to time.
5. Is increased tone beneficial in functional activities? e.g minimal increase in tone in extensors of hip & knee is beneficial in standing.

(C) Recovery process:

1. Spinal Shock: 3 to 6 weeks.

   In UMN lesions, tone usually increases for 1st 2 yrs and then levels off. Flexor tone is seen initially, extensor tone sets in after 6 months.

2. Effect of medications.

   Implications for the Patients:

   Muscle tone may benefit or hamper ADL.

   Chances for getting pressure sore are more with increased tone because of the difficulty in turning.

Strength:

Complete evaluation of all muscles.

Difficultes in evaluating the muscles:

1. Immobilisation device.

2. Poor stabilization because of weak muscles.
3. Trick movements.

Implications:

Some of the key muscles:

1. Scapular stabilizers useful in one man pivot transfer.
2. Pectoralis Major essential for rolling.
3. Triceps for bed mobility skills.
4. Wrist Extensors:
    1. Tenodesis.
    2. Moving wheelchair.
    3. use as a hook in other ADL.
5. Triceps: are essential for Wheelchair push ups. Lateral transfers.
6. Knee flexors and extensors for ambulation with AFO.
7. Hip flexors: very useful in ambulation.
8. Ankle Dorsiflexors: for clearing the ground and ambulation without AFO

Tolerance for Vertical Position:

1. Lying to sitting in bed.
2. Reclining wheelchair.
3. Tilt table.

Implications:

1. ADL independence, especially
    a) Transfers.
    b) Wheelchair movements.
    c) Dressing.

Balance :

- Evaluation of positive supporting reaction.
- Evaluation of equilibrium reaction.
- Evaluation of static balance / Dynamic balance.

Implications:

Unless patient had a head injury also, loss of balance is due to posterior column involvement and weakness of certain group of muscles.

Therefore he needs to learn to use some other group of muscles.

Providing a trunk support may also help.

Coordination:

a) Kinesthetic sense or awareness and accuracy of movements depend on timing. Find out as to how good was his performing skill prior to injury in comparison to the present skill.

b) It may require a lot of repetitions for improving the skill, also repeated instructions and explanations are beneficial.

c) ADL training is the ultimate goal.

Endurance :

1) Evaluation of cardiopulmonary status.

2) Evaluation of breathing with different activities.

3) Because of poor endurance, ADL may become slow.

4) Many a times Exercise sessions have to be made short for want of endurance.

Emotional Status:

1) Evaluation of patients understanding of the disability, whether he is realistic or not in his expectation of the outcome.

2) Evaluation of his accepting the disability.

3) Does he take active interest in solving his problems by himself? Is he interested in having equipments, which would improve the quality of life? e.g An orthosis may be useful for ambulation or perhaps he may have to use a W/C for the rest of his life

4) Is there a desire for him to be independent in his ADL or does he expect all the help from spouse or caretaker?

Relevant Information required in the Chart:

A) **Age:** Younger patients may be able to adapt or perform better. Would like to be independent in ADL

B) **Body build :**

1) Has he lost weight or put on with heavy weight? With heavy weights the movements are reduced, the chances of getting pressure sores are more. At the same time, the thinner the person more the bony prominences and they are also prone to have pressure sores.

2) Standard wheelchairs are easily available, where as over size wheelchairs and cushions are to be custom made. Over size wheelchair may not pass through standard doors. Wheelchair movements may also require larger area for mobility.

3) Heavier patients may require more number of people for ADL especially transferring. Many people prefer institutional life.

C) **Type of Injury:**

Type of injury and extent of injury would help in knowing the prognosis. e.g. Cases with complete transection of the cord, almost always will not have any chance of recovery and hence the patients have to live with disability. Similarly the level of injury would also give us an idea about the outcome as well as the chances of complications.

D) **General Health :**

Other injuries like head injury, limb fractures, cardiac, respiratory problems, ectopic bone formation, bladder and bowel problems are to be taken into consideration. They all affect the therapy and functional outcome. e.g. Ectopic bone formation affect ROM. Cardiac & Respiratory problems may affect the performance and endurance.

E) **Personality and Life style:**

1. Whether his work was sedentary nature or not.
2. Educational level / occupation
3. Willingness for change of lifestyle or job.
4. Acceptance of disability: Whether he is flexible or not?

F) **Family Support:**

1. Is family supportive?
2. Will he need another care giver?
3. Source of income for maintenance?
4. Will they continue the same therapy at home also?
5. Will they arrange for Vocational Rehabilitation so that he too can become an earning member and he too can live with dignity?

Evaluation is a continuous process. Initial evaluation is only a baseline evaluation to find out main problems of the patient. It is only to see whether we are able to go towards the expected goal and final evaluation before discharge is to see whether he has achieved the outcomes so that he can lead a meaningful life at home.

PHYSIOTHERAPY DURING THE REHABILITATIVE PHASE

BED MOBILITY

The most essential need of a SCI patient is some amount of mobility in the bed. The very first thing that he attempts is going on to the side by using upper trunk movements and scapular movements. The therapist can assist him initially by just doing passive rotation and asking him to assist as much as possible. Next, he is asked to do this by himself and the therapist helping only as much as he wants. When the patient can do by himself, more and more resistance has to be applied. He may use momentum, even weight cuffs may be applied to the hands to increase the momentum and facilitate movements. It also strengthens the muscles.

These activities give him awareness that with swift movements of the upper trunk he can initiate lower trunk movements.

Rolling can be made easy if he flexes his one hip and knee or both hips are flexed.

BED MOBILITY FOR PARAPLEGICS

Outcome: Patient should do all bed mobilities independently and without any specific equipment.

How to achieve:

- Rolling to the left and right.
- Rolling from prone to supine and back.
- Sitting with extended legs from supine as well as with flexed knees
- Moving to either side of bed.
- Moving to head and foot of the bed.
- Doing push-ups in bed to relieve pressure.
- Coming to quadruped(All Fours)position.
- Crawling
- Kneel

Considerations:

- Upper extremity strength and ROM.
- Spasticity.

Fig 2. Rolling

Quadruped

Crawling

Kneel Standing

- Pressure Sore.
- Age.
- Obesity.

Process:

- Evaluate and improve motor skills routinely.
- Appropriate bed mobility techniques.
- Make a routine exercise program to improve corresponding muscles.

PRESSURE RELIEF

Due to lack of sensation, they don't perceive the discomfort of pressure on certain vulnerable points because of squeezing of the local blood supply and resulting into ischemia over certain area and that will lead into pressure sore. They also do not have the ability to move the part to relieve the pressure. Therefore they have to develop new methods consciously. Patients who have very pointed bony prominences are prone to develop pressure sores than others (e.g. Some have anatomically pointed ischial tuberosities.) Therefore the therapists have to make a protocol for each patient and see as to how often they have to do the pressure relieving measures. After the patient becomes active in wheelchair activities, the chances of getting pressure sore are much less. Therefore careful clinical observations can help a patient to develop a treatment protocol which he can follow by himself.

Pressure Relieving Maneuvers:

1. Push- ups with hands on armrest of wheelchair lifting buttocks off the seat. (Strong triceps and shoulder extensors essential)

Push Ups

2. Forward leaning: Trunk muscles, pectoral muscles, triceps are essential. However

passively leaning forward can also relieve pressure to some extent.

3. Side lean by hooking on the rim of the wheelchair on the opposite side and leaning to one side and then the other. For doing this he needs strong biceps.
4. Electrically operated reclining wheelchair can also relieve the pressure.

Cushions

It is proven that no perfect cushion exists which is suitable for all the patients. We have to look into the lifestyle needs. Some of the cushions are too expensive. Some are easily punctured. Foam cushions need to be replaced every 6-18 months.

Arteriolar pressure in the skin capillaries is appropriately 32 mm hg but none of the cushions available in the market can maintain pressure below 32 mm. Perhaps, it is the posture control that matters most. Foam cushion has the benefit of being more porous with less moisture formation. However it is essential that the covering should be of light cotton material and not plastic. Another disadvantage of foam cushion is that if the atmospheric temperature is high, the skin temperature also tends to increase. Older patients tend to have lower skin capillary pressures (even as low as 20mm). Thin patients have less fat to cover bony prominences so also, flaccid patients. For these patients, cushions with lower contact pressure relief are recommended.

Those who have sensation intact, find contoured foam cushions uncomfortable. Those who have bladder bowel incontinence, less porous covers have to be given.

With contoured cushions, transferring is more difficult. All these factors should be taken into consideration. No cushion can substitute for frequent pressure releasing maneuvers, like wt shifting and pushups. Other factors like protein deficiency, anemia and infection also should be controlled in order to prevent pressure sores.

Mechanical Methods

Electrically operated reclining wheelchair is probably the best pressure relieving method especially for higher level paraplegic and tetraplegics.

Relieve pressure on ischium, sacrum, and greater trochanter. Factors to be considered are ROM (especially Hip and Elbow), Tone, obesity, cushions, adjustable arm rests, Ability to do pushups and other pressure relief skills like forward leaning, side leaning etc.

TRANSFERS

Moving from one surface to other is a major task for almost all SCI patients and needs special training and effort. They need the support of their strong hands to do that. They have to take extra care that they don't fall on the ground while transferring and also see that they do not hurt themselves.

As an initial step they are taught to maneuver to move themselves on the bed itself. Firmer beds are essential for the initial training. One has to learn step by step and may take several days in achieving each step. For instance,

i. Lifting and moving the legs to the left and right with their own hands;
ii. Managing to lift their body up by pushing on the beds.
iii. Shifting the body to the left and right on the bed; maintaining their balance.

    Some of them require improving their sitting balance especially the higher level paraplegics. Some may require strengthening their triceps. Some may require improving their ROM like rotation of trunk, elbow movements. How the various components are to be continued will depend on his skill. He should be encouraged to try different methods and should see that he never gives up trying. It would be ideal to show other patients who have achieved the task by trying different methods and have managed. Many a times the therapist has to try different methods before the patient finds a better method for him i.e. much experimentation has to be made.

iv. After achieving the ability to move from one end of the cot to the other he has to learn to transfer from the cot to the wheelchair. It would be easy for him if the WC is of the same height as the cot. If the WC is at a lower height he can easily go from the cot to WC but would find it difficult to transfer back. One needs very strong Upper extremities. A transfer board would facilitate the movements especially for higher level SCI cases who cannot maintain the balance while transferring. However with repeated attempts they learn to manage well even to a higher level. Keeping the Transfer board under the mid thigh is the most difficult thing.

v. The next transfer the paraplegics have to master is, from wheelchair to the commode and back. For this, bars need to be fitted on the side walls to make use of while transferring.

vi. It is ideal for paraplegics to learn to transfer to the floor and back, though this is not a very easy maneuver for obese and higher level paraplegics. Before attempting this it would be ideal for them to try it step by step. As a first step they can try corner- shifting in one attempt, then they can try holding on the two cots they can go up to the floor and back. Next they could go on to the floor with one hand on the floor and then going up with both hands on the cot and twisting their body. This is possible for patients with lower level of paraplegia and with satisfactory abdominal muscles.

Prior to learning, these transfer activities, it is necessary for the patient to develop good abdominals. He should attempt to go on all four position and do cat and camel exercises. Next he should attempt to do side-sitting on either side. Next he could attempt kneel standing with a hyper lordotic lumbar spine and balance. If he has a strong Quadratus lumborum and Lattissimus dorsi he can be taught kneel walking in parallel bars. All these exercises are preparatory exercises for walking with orthosis.

vii. Next he can learn a stand pivot transfer. This he can perform only if he has strong abdominals and trunk balance.

Dependent Transfers

1. One man pivot transfer
   - Anterior- arm around shoulder
   - Sideways – arm around the waist.
2. Two man lifts
3. Three man lifts.
4. Mechanical lift
   - Swivel bar transfer: Useful in cases of limited Hip ROM due to spasticity or ectopic bone formation around hip joints.
   - Bathroom Transfers: Before beginning these transfers, it is important to assess the patients own bathroom i.e. size of the bathroom; width of the entrance door, age, safety, amount of assistance required to transfer and time consumption. If the wheelchair would fit, lateral transfer is very practical. Raised toilet seats, grab bars are necessary. There is no one way to approach bath transfers. Selection of specific techniques will depend on each individual patient.

Transfer to Car and back to Wheelchair:

Difficulty is because of limited space for movement and bringing the wheelchair closer to the car seat. Therefore longer sliding boards are required. One man pivot transfer is also difficult because of the limited space. Finding the right method is a matter of experimentation usually while getting in legs should go in last and while getting out legs should come out first.

Transfer from Wheelchair to Floor:

This is a frightening maneuver for a patient who does not have strong abdominals and triceps. However, repeated attempts would give them confidence. First they have to scoot to the edge of the W / C and then come down straight on one hand and then the other with both knees following and coming on All fours position. The other alternative is going on the sides with straight legs sliding forward while landing on one hand, taking full weight with extended elbow and gradually bringing down the buttocks.

Floor to Wheelchair:

1. One of the easiest method is to have an intermediate step and then lifting the body to the wheelchair.
2. Another method in which patient has to get into kneeling position and then pull himself into the wheelchair. However getting into kneeling position is not that easy. Patients need a lot of strength and balance. Removing the foot pedals is important.
3. Yet another method is using the extensor tone which would assist in the pushing back to wheelchair. Here again the upper limb / lower limb strength and extensor tone are very important.

The factors that determine the process of transferring independently are the following:

1. Age. 2. Obesity. 3. Pressure sores. 4. Spasticity. 5. ROM.

The actual process includes:

1. Positioning of the wheelchair and management of its parts viz. brakes, detachable arm rests, foot rests.
2. Positioning of the transfer board (if necessary).
3. Ability to maintain sitting balance. Ability to lift body weight with elbows extended.
4. Ability to slide from one surface to the rest (lateral transfer) including the management of both lower extremities.

WHEELCHAIR MOBILITY

As a first step, the paraplegic has to accept, the very concept of using a wheelchair for mobility. This is not an easy thing for a person who was walking and running or driving a vehicle a few days or months back. For him to accept this slow moving mode of mobility is very difficult. Not only that, the stigma attached to the wheelchair is too much. However when he realizes that he is bedridden and that without moving from the bed, life becomes monotonous and a desire could come to him to get into a wheelchair. If he observes another wheelchair bound patient moving around comfortably, he too would consider getting into a wheelchair and attempt moving at least within house. When he observes people even get out of the house in a wheelchair and face other people in the society, slowly he gets a desire to accept a wheelchair for mobility rather than remain in a bedridden condition. This acceptance is the first step towards rehabilitation.

SCI above 1st lumbar spine should almost always consider wheelchair as a mode of mobility whereas cauda equina lesion with good abdominal muscles along with quadratus lumborum, should think of appliances in the form of orthosis like KAFO and a pair of crutches either elbow crutches or axillary crutches. Between wheelchair and orthosis the energy consumption is a much more with orthosis. However everyone has a desire in him for walking than being on a wheelchair. But when he realizes that wearing a caliper and crutches take time he would think in terms of getting into wheelchair and moving fast. However wheelchair needs more space to maneuver. Some of the bathrooms may not have enough space for movement.

Achieving good wheelchair mobility is essential for all high level paraplegics. For developing wheelchair skills, it is essential to have a good posture of the trunk and strong upper extremity. The muscles to be developed are shoulder flexors and extensors, elbow flexors and extensors, so also good hand grip. Maintaining a good posture is essential for various reasons mainly for conservation of energy, proper chest expansion, endurance, prevention of pressure sores (as awkward sitting can cause unequal pressure leading to pressure sore.) Bad alignment cause postural pain in the back. Wheelchair with higher back may help patient in maintaining a good posture. However, wheelchairs with higher back support are not good for fast movement. With lower back support, the movements of the arms are swifter.

A good pelvic support is essential. If there is any pelvic obliquity it needs to be corrected.

Sling seat causes adduction of lower extremities and causes the base of support becoming smaller and this would cause instability. Sometimes leg straps are necessary to keep the lower extremities in abduction.

Gel cushions also may cause instability. Some prefer higher foot rests in order to get pelvic stability, preventing forward movement of the pelvis.

Slight inclined back support with lumbar roll is a good option the lumbar roll will maintain in a good position; prevent kyphosis, scoliosis and pelvic obliquities. It also helps in maintain the pressure anterior to the ischial tuberosities and thighs.

Adjustment of the height of the arm rest is also important as they are essential for adjusting and maintaining the trunk in good posture.

Wheelchair movements are better with strong upper extremities. However more than the strength, good timing and coordination would help in skillful movement.

For forward movements, the muscle essential are: shoulder flexors, adductors and external rotators whereas for backward movements: shoulder extensors and adductors and internal rotators.

Propelling of Wheelchair:

1. Trunk flexors and extensors would facilitate forward push.
2. Avoid holding the tyre as it can get caught in the brake and cause injury. Grip should be always on the rim.

Wheelies:

Wheelie is one of the most advanced skills, a

paraplegic can learn i.e. to lift the front wheels up and maintain the balance on the back wheels only. This position would help a patient to negotiate curbs up. Without achieving a "Full Wheelie" a patient can negotiate curbs up to 2 inches. When he approaches the curb, he should stop and raise the front wheel then lean forward push the back wheel up.

For negotiating higher curbs i.e. more than 2 inches, one has to gain momentum by going fast for 5 to 10 feet and go into wheelie and drop front casters on the curb and then lean forward then push the rear wheels. One needs good rhythm and timing which everyone can't achieve.

For a patient to become independent in mobility both indoors and outdoors, he has to do step by step training like:

- Trunk mobility in sitting.
- Balance and equilibrium reach outs in sitting.
- Use of reach out.
- Use of upper extremity in sitting.
- Wheelies.
- Improve endurance.
- Forward and backward and turns.
- Management of brakes and removing and putting back arm rests, foot rests etc.
- Movements on surfaces like tiles, carpets etc.
- Movements through narrow passages, elevators, ramps.
- Movements curbs bumps, cracks in side walk etc.
- Crossing the street safely.

AMBULATION:
To Walk or not to Walk?

If you ask any patient this question, he would certainly say that he wants to walk. Initial stage he would say that he cannot accept a wheelchair as a mode of ambulation. However as time passes and he realizes that there is a lot of energy consumption he slowly accepts the fact that wheelchair may be better mode of locomotion. However those who are functionally able to manage with orthosis will never take the idea of a wheelchair. Therapists would like to sell the idea of wheelchair to those patients who have no hip flexors. This is because wheelchair is more practical and energy saving and time saving.

There is a debate on this issue even among the medical personnel. Those who are in favor of ambulation and standing believe there is a physiological benefit for the patient.

i. Most of the body's calcium is found in the bone.

ii. SCI patients have a high calcium washout and incidence of osteoporosis is evidently seen in the bones.

iii. This calcium washout is considered to be predisposing factor for formation of bladder stone and ectopic bone formation which are seen in SCI patients.

iv. Weight bearing can increase the bone density.

v. It is assumed that with ambulation and weight bearing, calcium excretion will be decreased and therefore less osteoporosis, ectopic bone formation and fewer bladder stones.

However there is a counterargument that standing and ambulation alone are not sufficient and more pressure is needed to promote bone growth. Muscle contraction can provide much more compressive force also shearing and torsion forces increase bone density significantly.

One study done by Jacqueline Claus-Walker and several physicians at TIPR in 70's found that calcium excretion increases during the first ten days, and continues to increase in the next six months and after 1 year it lowers to normal. They also found that position of the body will influence fluid distribution. Position is registered by the baroreceptors and transmitted via neural pathways to CNS. In the SCI patient, weight bearing is not often perceived as the neural pathways are not working. In their study they also found complete recumbency for 3 days did not increase hypercalciuera in quadriplegics whereas in normal healthy subjects, calcinera increased b 1.5 times.

The question still remains whether to encourage wheelchair mobility or weight bearing ambulation? There is no doubt that standing and walking would certainly boost up the morale of the patient as he feels that he is more like the normal people.

However in course of time, he would realize that walking is much more time consuming and energy consuming. Therefore, it is essential that he should be given a chance to be on his feet with appliances.

However considering the cost of the equipment before prescribing them it is advisable to try temporary trial orthosis like Push Knee Splints and high boots with posterior steel shank, for a few days and gait training could be given. If he is comfortable with these temporary orthotic devices, a prescription could be given for KAFO. Many SCI cases still prefer these temporary devices, as they are easy to wear and light in weight. The therapist should give the choice to the patients. Finally the patient forms his opinion as to which mode of ambulation he prefers.

GAIT TRAINING

A gait with orthosis like KAFO and crutches requires very high energy consumption because the patient has to maintain the balance in a PARAPLEGIC STANCE both in the dynamic movements and while resting, i.e. keeping the spine in a hyper extended position and taking the weight on the crutches. Therefore the patients get exhausted after walking a few steps. It is essential that the energy consumption should be bare minimum.

Depending on the patients muscle power, "**four point gait**" **or "swing to gait**" are the easiest to learn. If he can master these gaits, he can easily negotiate narrow spaces. Wheel chair may not pass through very narrow passages. Swing through gait needs more strength, coordination and are difficult to learn but once mastered the energy consumption is less.

Gait training should be started in the parallel bars first. Initially the patient is taught to be in the paraplegic stance i.e. hip in extension and spine in hyper lordotic posture. The therapist stands on the side and pushes the shoulders backwards and pelvis forward. Optimum position of the ankle is also important as with hyper dorsiflexion and plantarflexion, one tends to loose balance. High boots with posterior steel shank can maintain ankle mobility in neutral position with minimal ankle mobility required for ambulation. After this, patient is able to balance in standing. He is taught four point gait pattern which is the easiest to learn. It is easier for patients who have some power in hip flexors. If he has no hip flexors, he may use adductors by externally rotating at the hips and dragging his feet alternatively. The third option is by hiking the pelvis by quadratus lumborum. The other option is by "**swing to or swing through"** gait.

Once he is able to move comfortably in the parallel bars he is brought out first in the walker and later with crutches. Initially, he could be given axillary crutches and later on elbow crutches. Older patients prefer walker as it gives much more balance than any of the crutches.

Once he gains confidence in moving indoor, he can be brought outdoor. Initially he is taught to walk long distances so as to gain confidence and endurance.

The next step could be to climb stairs. Initially he is taught to hold onto one side on the railings and the other side on crutch and lift his body up and climb one step and bring the crutch to the standing level and then lift once again holding the crutch and railing climb another step.

Another method is facing the railing and holding it with both hands, swing one limb in one pendulum cycle and then hike the hip and place the foot on the higher step; then bring the other foot up. This is possible only if he has strong hip flexors or quadratus lumborum. There is yet another way of climbing up i.e. Jack kniving, Here the patient has to lift himself up higher than the height of a step with back facing the staircase and swinging both legs backwards so as to reach the higher step. The person has to use one crutch in one hand and holding the railing with the other hand or using crutches in both hands. This is possible only for patients who have very good control of the trunk as well as very strong Triceps and shoulder muscles.

OCCUPATIONAL THERAPY IN PARAPLEGIA

Paraplegia

Paraplegia affects the person's functional independence and has a major impact on quality of life, sense of self worth and consequential social participation. Whilst it is generally expected that the degree of functional independence achievable is dependent on a person's SCI level, a person's neurological level should not be viewed as strictly predictive but rather as indicative of potential function.

Expected Levels of Functional Independence at different thoracic and lumbar levels

Level	Abilities	Expected Levels of Functional Independence
T1	Has added strength and precision of fingers that result in limited or natural hand function.	**Daily tasks:** Can live independently without assistive devices in feeding, bathing, grooming, oral and facial hygiene, dressing, bladder management and bowel management. **Mobility:** Uses manual wheelchair. Can transfer independently.
T2-T6	Has normal motor function in head, neck, shoulders, arms, hands and fingers. Has increased use of rib and chest muscles, or trunk control.	**Daily tasks:** Should be totally independent with all activities. **Mobility:** A few individuals are capable of limited walking with extensive bracing. This requires extremely high energy and puts stress on the upper body, offering no functional advantage. Can lead to damage of upper joints.
T7-T12	Has added motor function from increased abdominal control.	**Daily tasks:** Able to perform unsupported seated activities.
		Mobility: Same as above.
		Health care: Has improved cough effectiveness.
L1-L5	Has additional return of motor movement in the hips and knees.	**Mobility:** Walking can be a viable function, with the help of specialized leg and ankle braces. Lower levels walk with greater ease with the help of assistive devices.
S1-S5	Depending on level of injury, there are various degrees of return of voluntary bladder, bowel and sexual functions.	**Mobility:** Increased ability to walk with fewer or no supportive devices.

OT in Paraplegia

The purpose of OT in paraplegia is to

1. Evaluate a person's ability and level of functioning in his/her home, at work, and while engaging in leisure activities and hobbies;
2. To provide individualized therapy to retrain people to perform daily living skills using adaptive techniques;
3. To facilitate coping skills that could help a person overcome the effects of SCI.

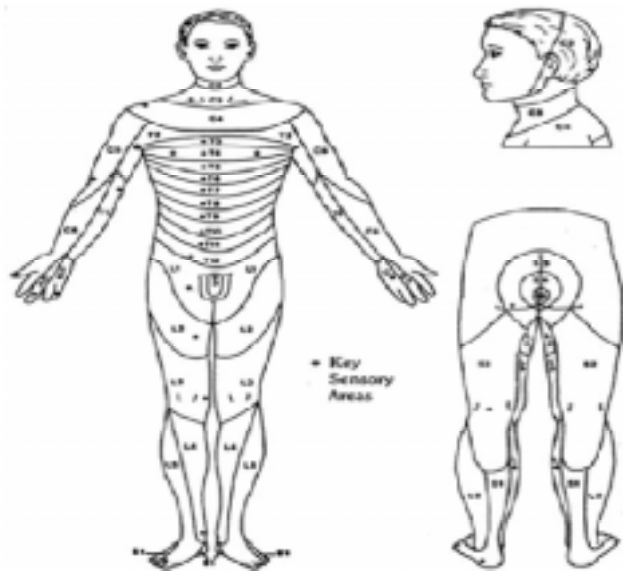
Role of the Occupational therapist

- The occupational therapist is particularly concerned in helping the paraplegic reach the highest level of independence both physically and psychologically that his injury, home, and work environment will allow. The paraplegic with the help of the occupational therapist overcomes difficulties successfully with the use of various aids and adaptive equipment and modifications in the home and place of work environment. A variety of purposeful activities are also used and modified to promote self-esteem by highlighting functional skills and emphasizing the tangible development of a client's physical, social, emotional, sensory and cognitive abilities. OT's unique contribution to occupational performance lies in its use of purposeful activities to promote psychological and physical health and maximum functional independence. As a part of complex rehabilitation, it is applied in solving the problems of occupation (self-care, work and leisure) of patients. It is emphasized that early OT, started immediately after stabilization of patient's functional state, is of great importance.

Occupational therapy Evaluation

Evaluation is an ongoing process that begins from the day of admission and continues long after discharge on an OPD basis. An accurate and comprehensive formal evaluation is needed to determine basic neurological, clinical and functional status of the patient. Medical files provide demographic data, the diagnosis and present and past history and other required information like associated injuries and complications, fractures, cardiac conditions etc. Occupational therapists evaluate

- **Passive and active range of motion (ROM)** - of all joints in the upper and lower extremity using Goniometry
- **Manual muscle testing-** of all muscles using accepted muscle testing techniques and
- **Sensory evaluation-** of all dermatomes of the body, including evaluation for light touch, pin prick, joint proprioception, stereognosis, and kinesthesia, to determine areas of loss or impaired sensation. It helps in identifying functional limitations and in establishing level of injury.



MRI of Lumbar Spine

- **The other areas assessed are:-**

ADLs, Reflexes present or lost, Muscle tone (once spinal shock subsides), Endurance and perceptual and cognitive skills in case of a combined SCI and TBI diagnosis. A work history, home accessibility, social, lifestyle, leisure and driving interests are also considered.

In order to determine the functional status of the patient the occupational therapists use the following assessment tools:-

1. **Functional Independence scale (FIM)**

   -It assesses six areas of function (Self-care, Sphincter control, Mobility, Locomotion, Communication and Social cognition), which fall under two dimensions (Motor and Cognitive).

   -It consists of 18 items assessing the 6 areas of function

2. Klein Bell ADL Scale

   -Measure ADL independence to determine current status, change in status, & sub activities to focus on in rehabilitation

   -Areas assessed are dressing, mobility, elimination, bathing & hygiene, eating & emergency communications

3. Spinal Cord Injury Functional Ambulation Inventory (SCI-FAI)

   -Assesses functional walking ability in ambulatory individuals with SCI (Field-Fote et al, 2001).

   - It is composed of three sub-scales, these include:

   - Gait
   - Assistive Device
   - Walking Mobility

4. The Spinal Cord Independence Measure (SCIM)

   -The SCIM, now in its third iteration, is a new disability scale developed to specifically address the ability of SCI patients to accomplish ADLs (Catz et al. 1997).

   - 19 items assesses 3 domains:

   Self-care (6 items, scores range from 0–20)

   Respiration and sphincter management (4 items, scores range from 0–40)

Mobility (9 items, scores range from 0–40)

Occupational Therapy Intervention

The Acute Phase:

When the patient arrives in the hospital he will mostly be confined to bed waiting for or recovering

from surgery to stabilize the spine which may be immobilized in traction or in a halo brace or body jacket and prohibited from flexing, extending, and rotating the spine. Occupational therapy begins within the first 48 hours of admission.

After the evaluation a daily range of motion program should be started with active and active-assisted ROM of all joints within strength, ability, and tolerance level. Positioning should be evaluated and instruction to the staff, patient, and family members should be given if necessary. Participation in self care activities (eating, combing, and writing) should be encouraged. Discussions regarding further therapy and rehabilitation are initiated to prepare the patient and the family members for discharge.

The Rehabilitation Phase:

The Rehabilitation Phase is also known as the Active or Mobilization phase. In this phase the patient can sit in a wheelchair therefore upright sitting tolerance should be developed. The occupational therapist also works on:-

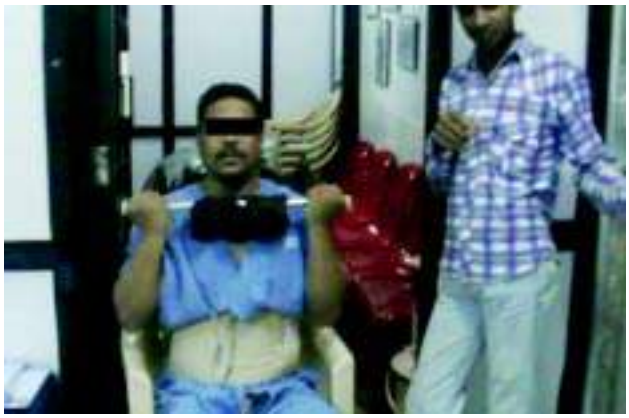
1. Bed Mobility

Bed mobility skills like rolling, coming to sit from supine, scooting and sitting at edge of bed techniques are taught to the patient. The patient is instructed and assistance is provided until patient is independent. Weight bearing while performing therapeutic activities in different positions like prone on arms, quadriped and kneeling are initiated to improve upper extremity and trunk stability, balance and develop skills for transfers.

Fig 2

2. Upper Extremity Strengthening

The Occupational therapist uses weights/ resistance for progressive resistive exercises and resistive activities to strengthen upper extremity muscles. As muscle strength increases, the amount of resistance should be increased to help the patient increase tolerance and endurance. Shoulder exercises should emphasize the shoulder depressors (latissimus dorsi), the flexors, abductors, and extensors (deltoids), and the scapular musculature. The triceps, pectoralis, and latissimus dorsi are required for weight shifts in the wheelchair and for transfers.



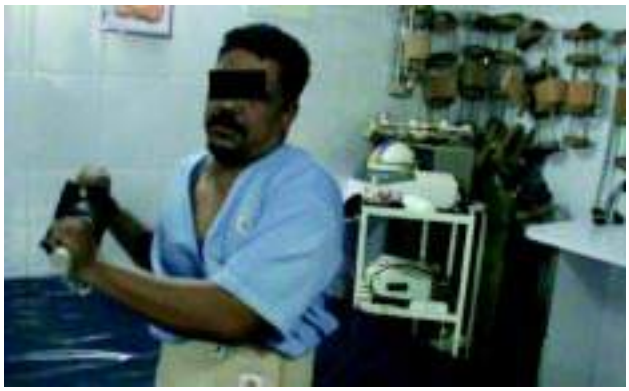


Fig 3 fig 4

3. Endurance Training

The intervention progamme should be graded to increase the amount of resistance that can be tolerated during the activity. As muscle power and endurance increases, increase the amount of time in wheelchair activities which helps patient participate in activities and occupation throughout the day.

4. Self care retraining-

Self-care retraining may commence whilst patient is still in bed, focusing on feeding and grooming. There are many assistive devices/ specialized items of equipment that O.T. prescribes to help you achieve greater independence in feeding, grooming,

showering, dressing, and bladder and bowel management.

- Grooming

  Grooming tasks include brushing teeth, washing face, combing hair, shaving and applying make-up. As with, upper body dressing a person with a paraplegia usually has full use of their arms and grooming is completed without difficulty from a wheelchair as long as items are in reach.

- **Feeding**

  Feeding, like upper body dressing and grooming, is usually not difficult for a person with a paraplegic level of injury.

- **Upper body dressing (UBD)**

  Upper body dressing (UBD) includes donning and doffing of clothing from waist upwards. As in paraplegics the upper extremities are spared so they can independently perform upper body dressing if the clothing's are in reach. In absence of independent sitting, supported sitting enables the patient to complete UBD.

- **Lower body dressing (LBD)**

  Lower body dressing (LBD) includes putting on and taking off any clothing item from the waist down. When dressing the lower body, persons with a paraplegic level of injury might find it helpful to use a combination of alternative techniques and adaptive equipment. The most common position for performing LBD is circle sitting or long sitting in bed. This allows the person to reach his/her feet from a large base of support, which increases balance. Technique to dress lower body is as follows:

Person sits up in the bed. One leg is bent at the knee and lifted over the other, making sure that the leg is not dragging on the bed. Socks can be put in this position while checking placement of seams to avoid pressure on toes. The process is repeated with the other leg. Trousers are put on after the socks to avoid toes being caught in the seams. The process is the same as the wearing of socks. Trousers are then pulled up as far as possible. Then the person lies on the bed and pulls up the trousers by rolling from one side to the other to pull it over the buttocks. By sitting up again, shoes can be put in the same way.

Fig. 5,6,7,8,9,10

Some of the most commonly used pieces of adaptive equipment (AE) used during dressing include:

- Dressing sticks
- Reachers
- Long-handled shoe horns
- Button hooks
- Velcro
- Elastic shoe laces
- Sock aids
- Legs straps

- **Toileting**

  Toileting includes the ability to pull down clothing in preparation for elimination, cleaning of the perineal area and pulling clothing up after completion. A person is often able to independently complete the process with the correct technique and needed equipment.

Examples include:

- Leaning on one elbow to raise a hip and pull down clothing from side to side

Leg straps to assist lifting legs

- Raised chair with a cut hole
- A standard-height toilet with a raised toilet seat and a cutout in the front and back to allow an individual or attendant access for digital stimulation and hygiene purposes
- Specially designed seats with attached arms, if needed.
- Sufficient area around the toilet to permit wheelchair transfer
- Grab bars at a height to maximize the person's ability

- **Bathing**

  The initial days or weeks following injury, will most likely be sponge bath from bed. Once patient is stable showering is allowed. The following aids assist with safety and completion of bathing:

  Shower chair

  Tub chair/tub bench with a back

  Transfer board

Fig. 5

Fig. 6

Fig. 7

Fig. 8

Fig. 9

Fig. 10

Hand held shower
Long handled sponge
Grab bars on three sides of the bathroom
Thermometer
Transfer board
Hand held shower
Thermometer Towel racks, soap dishes, and shelves within reach
Full length mirrors should be installed wherever possible
Non skid floors
Insulated hot water pipes to prevent burns

Bowel and bladder training

Therapists may assist in independent stimulation and applying a urinary collection device, with or without facilitatory equipment, by suggesting the best possible technique in the best possible position.

Transfers:

For independent transfers in paraplegics, muscle strength of upper limbs is very important. Upper limbs have to be strong enough to lift and support the body while transferring. Transfer board to bridge the gap between your wheelchair and the bed, commode, or bathtub.

The Two methods commonly used are taught to the patient for independent transfers:

Forward-on:

The wheelchair is placed at right angles to the bed and patient lifts the legs onto the surface (bed or

chair) one at a time. Footrests are removed or swayed away and wheelchair is moved closer. The person moves forward as necessary until buttocks are on the surface. Process is reversed to get back on the chair.

Legs down (side-transfer method):

The wheelchair is positioned beside the bed. Footplates are swung away or removed. The person moves forward on the seat until the feet are flat on the floor. The nearest armrest is removed, and the hand is placed on the farer armrest. And the person lifts the buttocks off the chair and on the bed or chair. Then legs are repositioned.

Some transfer rules and technique:-

- Transfers best done at same height levels
- Get the patient scoot as close as possible to the surface he has to be moved to
- Lock wheelchair if transfer is from or to a wheelchair.
- Get armrest out of the way on the side next to the surface patient is transferring to.
- Transfer using a closed fist with wrist in neutral for an increase in vertical excursion.
- Depress shoulders for additional height while transferring.
- When transferring, Lean trunk forward, head should move in the opposite direction of your hips. This is known as a head-hips relationship and can help with movement and clearing obstacles.
- To protect shoulders, position arms as close to the body as possible (about 30-45 degrees away from your body) while lifting weight.

Fig 11

- Lift-off

  Make sure that patient clears the surface during lifting to avoid shearing and pressure sores.

Mobility

The International Classification of Function defines mobility as "moving by changing body positions or locations by transferring from one place to another, by carrying, moving or manipulating objects, by walking, running or climbing, and by using various forms of transportation." Individuals with paraplegia due to the loss or impairment of motor function in lower extremities are able to move through use of technology such as wheelchairs, walkers, orthosis such as leg braces and crutches.

Wheelchairs

It is the most primary means of mobility. The occupational therapist helps the patient in selecting the most appropriate type of wheelchair (manual or powered), adjusting it well (wheelchair, armrests or casters height / width), training the patient in safe propulsion techniques and effective use in different settings. The Wheelchair Skill Test (www.wheelchairskillsprogram.ca.) can be used to determine which wheelchair skills are required to be addressed and to document any progress after intervention.

Parts of wheelchair and its uses, techniques to ascend and descend ramps, negotiating curbs and steps, taking a turn, transfers from wheelchair to different heights and surfaces and vice- versa, protecting oneself in case of falls are some of the wheelchair skills the patient needs training in. The family members are also educated regarding the care and use of the wheelchair.

Walkers

Walkers are metal frames designed to provide support and stability while walking. Walkers may be folding or fixed; height-adjustable or non-height-adjustable; equipped with wheels on the front, all four, or none of the legs; suited for stairs; and / or equipped with seats

Crutches

There are two basic styles of crutches: traditional under-arm axillary crutches or forearm crutches, whichever suits best can be prescribed.

Environmental Barriers, Home and Work (Job/

School/Play) Barriers assessment and recommendations

Environmental Barriers, Home and Work Barriers are physical impediments that keep patients from functioning optimally in their surroundings. Occupational therapists use the results of tests and measures to identify variety of possible impediments including:

– Safety Hazards (e.g. throw rug, slippery surfaces for patient with help of lower extremity orthosis & walker etc)

– Access problems (e.g. narrow doors, high thresholds & steps, absence of elevators)

– Home & Office design barriers (e.g. excessive distances to negotiate, multi storey environments, sinks, bathrooms, counters and placement of controls or switches)

Occupational therapists after identifying the impediments use the results of tests to suggest modification to the environment to improve functioning in the home, workplace and other settings:

- Construction of ramps or lifts to home.
- Railings and grabs around the house
- Electrical points fixed at a height accessible to patient from wheelchair
- Enlarged doorways and passages for easy access of wheelchairs
- Removal of cabinets from under sinks and platforms in kitchen
- Removal or rearranging furniture that hampers wheelchair access
- Alter thresholds to no more than 3/4" in height
- Position the heights of bed and chair in level with cushion of wheelchair to ensure easy transfers.

Tests and measures may include those that characterize or quantify:-

- Current & Potential barriers:- e.g. checklists, interviews, observations or questionnaires
- Physical space & environment:- observations, photographic assessments, questionnaires, structural specifications, technology assisted or videographic assessments

The environment directly impairs patients's ability to perform tasks and abilities that support physical, social and psychological well being. A variety of instruments have developed that address the impact of environmental factors on function for e.g.:

Craig Handicap assessment & reporting Techniques (CHART)- This instrument was developed to document an individual's functioning with in his or her societal context. It examines levels of involvement within six domains of functioning: physical independence, cognitive independence, mobility, occupation, social integration & economic self-sufficiency. The CHART-sf2 is a 20-item shortened version of the CHART.

Domestic retraining

As part of occupational therapy program, patients have the opportunity to practice homemaking activities in a simulated or in the environment the patient will be returning. E.g. cooking in a wheelchair accessible kitchen, where appropriate skills will be taught and opportunities to practice different pieces of equipment that can enhance patient level of independence in this area. Other domestic skills may also be addressed according to your individual need.

A number of products are commercially available to facilitate independence in performing home management tasks. Examples of these products include:-

Easy reachers, long-handled dustpans, brooms or vacuum cleaners

Trays or trolleys can be utilised to transport items or carry hot items to reduce the risk of burns on lower limbs. A front loading washing machine, and a lowered clothes line or front loading dryer, can facilitate independence in laundry tasks.

Assistance with return to driving

For a paraplegic who wants to resume driving, – a driving assessment with an Occupational Therapist who specializes in driving assessment and intervention is usually required. The driving assessment will determine readiness to resume driving and recommend modifications that are required to enable safe and functional driving, e.g. hand controls. O.T. aims to provide comprehensive education and retraining to help patient return to driving. Transportation options are also addressed (e. g. modified vehicle and public transport). Motor vehicle adaptations are selected based on physical capacity of an individual.

Common car adaptations include:-

- Hand controls to operate accelerator and brake
- Handles to assist in transfers from a wheelchair to a car or van,
- Ramps,
- Wheelchair and transfer lifts
- Raised ceilings,
- Lowered floors, and
- Wheelchair carriers attached to the outside of the car or van

Leisure skills

Leisure time means free time and excludes time spent on essential activities such as paid employment, chores, eating, sleeping, and going to school. Leisure activities not only put high emphasis on restful pursuits such as reading, watching TV, stamp collecting or watching performances, but also on more physical activities such as sports like basketball, swimming, table tennis and weight lifting. Many patients with spinal cord injuries are unemployed and therefore have more leisure time available. To maintain their self esteem suitable hobbies and sporting activities should be encouraged.

Together with the patient the O.T., investigates options for returning to previous leisure interests, and also developing new pursuits. Recreation and leisure activities assist people to not only pursue their individual talents, abilities and interests but also develop important and valuable social networks and relationships in the broader community.

Leisure activities are more important for the patients post injury than before injury as it helps to maintain the competitive instinct and to help integration back into the community.

Depending on the severity of their injury, they may have to learn new leisure skills. The therapists provide counseling about returning to pre-injury leisure interests, developing new leisure interests and possible adaptations needed to pursue those interests. People are also encouraged to identify benefits of their leisure time and to consider the important part leisure played in their lives before their injury.

Vocational rehabilitation

Vocational options are discussed with the patient by the occupational therapist. An occupation is of varying importance to patients, but most will see it as giving a sense of purpose to their life and will want to return to their former work if at all possible. Early contact with the patient's employer to discuss the feasibility of an eventual return to his previous job is important. If the degree of the patient's disability precludes this some employers are sympathetic and flexible, and will offer a job that will be possible from a wheelchair. If appropriate, a work site assessment may be arranged. An on-site work task analysis performed at the client's workplace as well as examination of the worker and the work environment will provide the therapist valuable insight about the present skills of the patient. However, many patients-find life outside hospital-difficult enough initially, even without the added responsibility of a job, and in these circumstances a period of adjustment at home is advisable before they return to work.

If a patient is planning to return to his previous employer, school, or college the occupational therapist should assess the suitability of the premises for wheelchair accessibility. Ideally, if a patient is considering returning to work the therapist assists him by assessing his work abilities in a simulated work environment. In addition the patient will build up his strength and stamina and both he and the staff will have a clearer idea of his employment capabilities. The patient should be taught proper body mechanisms and energy conservation to perform work in a safe manner. The occupational therapists provides referrals for a number of services to provide vocational counselling, rehabilitation and assistance with finding employment in the community.

REHABILITATION OF PATIENTS WITH LESIONS OF CERVICAL SPINE (TETRAPLEGICS)

Tetraplegia refers to partial or complete paralysis of all four extremities and trunk including the respiratory muscles; it results from lesion of the cervical cord.

In India approximately 15 lakh (1.5 million) people are living with SCI. Approximately, 10,000 new cases each year; about 8,000 are persons with paraplegia, and 2,000 persons with tetraplegia. 82.5% are males, age group 16-30.

Causes

1. Traumatic causes.

- Road traffic accidents,
- Fall from height (tree, train, buildings, govinda)
- Fall of objects on spine (buildings, earthquake, mining, load taken on head upper back, working under vehicles)
- Diving
- Violence
- Sports

2. Non Traumatic causes

- Transverse myelitis
- Syringomyelia
- Multiple sclerosis
- Amyotropic lateral sclerosis
- Pott's spine
- Neoplasms
- Disc prolapses.

C 5 is the most common injury level, followed by C4 then C 6.

Percentage of injuries by ASIA classification

- Incomplete Tetraplegia 34.5
- Complete Tetraplegia 18.4 %
- Incomplete Paraplegia 17.5 %
- Complete Paraplegia 18.4 %

PHYSIOTHERAPY MANAGEMENT OF TETRAPLEGIA

Physiotherapy (PT) which plays a very vital role in the rehabilitation and well-being of persons with spinal cord injuries can be broadly discussed as

1) PT management at the site of injury.
2) PT during the acute stage (intensive care)
3) PT in the sub-acute stage(outside intensive care, in the wards)
4) Active rehab / sports
5) PT at home and in the community

1) PT management at the site of injury

Move the patient with great care to avoid bending of neck / back. Have another person put the stretcher in place. If the neck is injured or broken, put sand bags or tightly folded clothing or boxes that are taped on sides of head to keep head / neck from moving. If there is more help the 10 steps should be followed

- Application of steady but gentle manual traction to the head and neck and airway opened by modified jaw thrust.
- Placing a rigid collar around the patient's neck.
- Board is placed parallel to the patient
- Kneel at patients side, opposite the board, leaving room to roll patient towards you, one at the shoulder, one at the waist, one at the knee, one maintains traction.
- Log roll to side, as one unit.
- Waist level person grips spine board and pulls into position against the patient.
- Roll the patient onto the board
- The patients head is secured to board with cravat or 3" tape. Place folded clothing besides head and neck for additional protection, these must be secured by a cravat over forehead and then tied to the sides of the board.
- Additional 3 straps, one across chest, one hips, and one across the knees are put, traction is maintained on the neck.
- Move patient and board on wheeled ambulance stretcher.

This safe lifting at site, on the ambulance, or at times onto a plane and then to a hospital which has expertise in treating people with spinal cord injuries will ensure that no further neurological damage occurs. With good handling, an incomplete injury will not progress to become a complete injury. When it is a high cervical spine injury, even the diaphragm, besides the intercostal muscles would be affected, and breathing is severely affected. Thus

an ambulance with well equipped artificial respiratory kits would be needed.

The physiotherapist can be part of this emergency evacuation team, or get involved in training personnel that safe lifting techniques of the person injured, and those lifting are adopted. This will prevent neurological damage to the person injured, and will prevent musculo-skeletal damage to those who are lifting.

2) Physiotherapy in the acute stage

From the time the patient is admitted in intensive care, physiotherapy is begun for

a) Chest care

b) Maintaining joint mobility

c) Posture

d) Pressure Relief

Other assessment includes

- Level of arousal
- Cranial nerves assessment
- Associated injuries like head injury, chest wall injury, fractures of bones, neuro-vascular injury.
- The type of respiration and chest mobility
- Ability to cough
- The nature of the injury
- Occupation of the person

The acute phase is very critical as complications can occur which we should be very much aware of

1. Pneumonia, atelectasis
2. Deep vein thrombosis (DVT
3. Pressure sores
4. Urinary tract infections(UTI)
5. Autonomic dysreflexia
6. Burns due to sensory loss
7. Heterotropic ossification
8. Osteoporosis
9. Orthostatic hypotension

FIGURE 1.1-2

Assessment tools for quadriplegia are the same as we use in evaluating paraplegia. i.e. ASIA Scale, Modified Ashworth Scale, etc. besides these, there are various other measures of assessment which are described below.

The neurological level is the most caudal neurological segment of the spinal cord that retains normal sensory and motor function on both sides of the body. Sensory and motor levels may vary from side to side and even on a given side.

4 levels, sensory and motor, both right side and left side are examined. 10 myotomes and 28 dermatomes are checked.

The 5 key muscles in upper limb and 5 key muscles in lower limb are

Upper limb:

- C5 elbow flexors
- C6 Wrist extensors
- C7 Elbow extensors
- C8 Finger flexors (distal phalanx of middle finger)
- T1 Finger abductor, little finger.

Lower limb:

- L2 Hip flexors
- L3 Knee extensors
- L4 Ankle dorsiflexors
- L5 Long toe extensors
- S1 Ankle plantar flexors.

Grading is done from 0 to 5 strength, total score 25 for each limb,50 for upper limbs, 50 for lower limbs, total 100 for key muscles of upper and lower limbs.

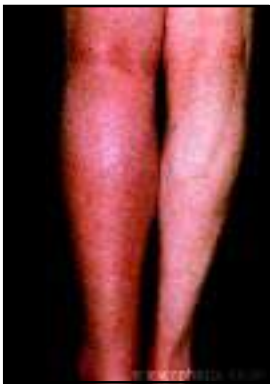
28 dermatomes from C2 to S4 / S5 are examined for pin prick and light touch

- 0 absent sensation
- 1 impaired sensation
- 2 normal sensation.

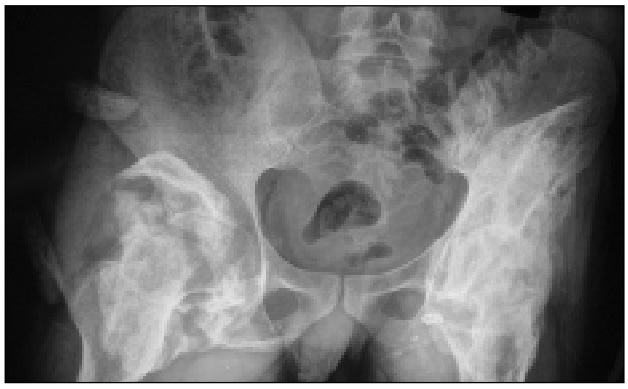
Thus a total score of 56 each for pin prick and 56 for light touch, total 112 for each limb.

Assessment of Respiratory system

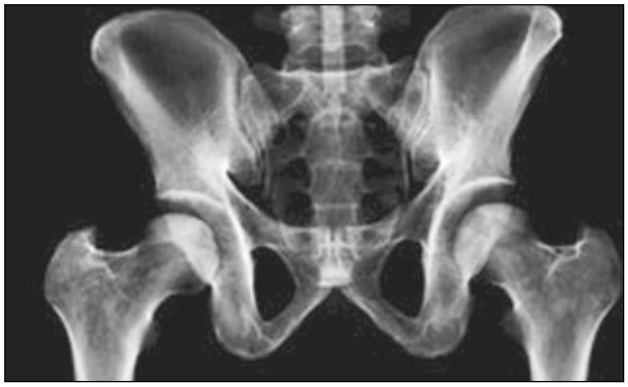
- Diaphragmatic movement
- Chest wall movement



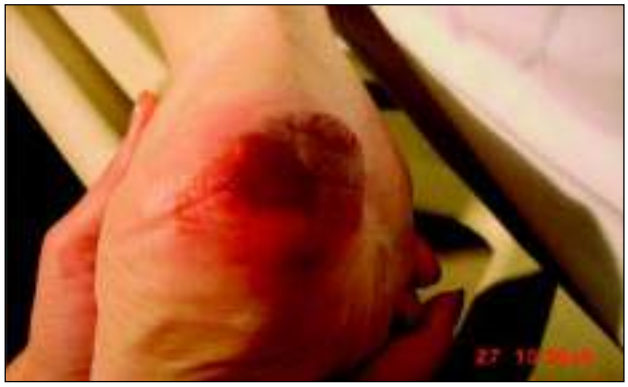
Deep Vein Thrombosis



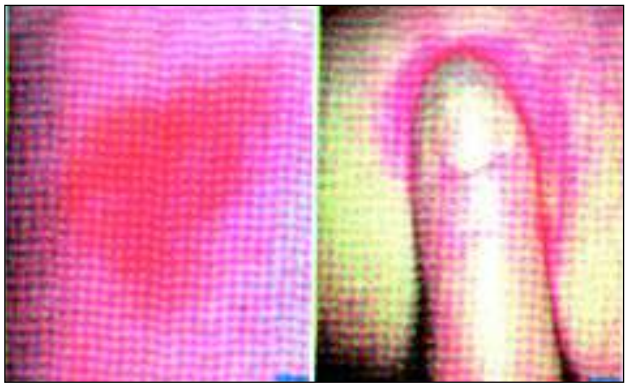
Heterotopic Ossification



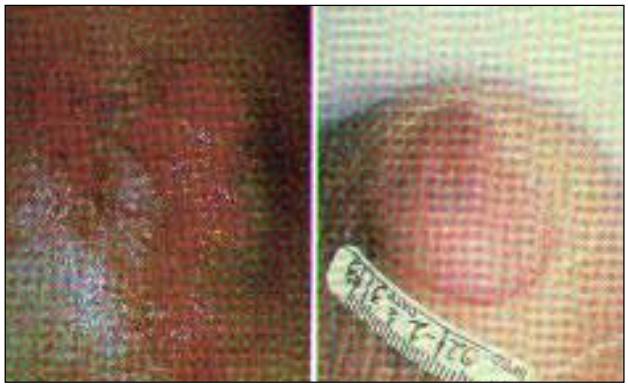
Osteoporosis



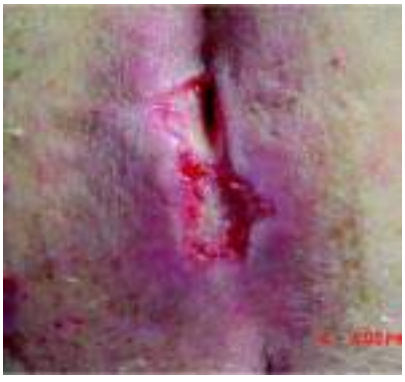
Pressure Sore Diagram



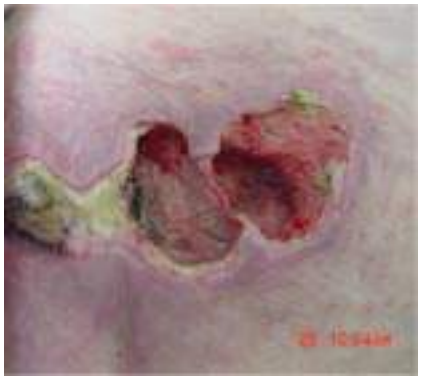
Pressure Sore – Stage 1



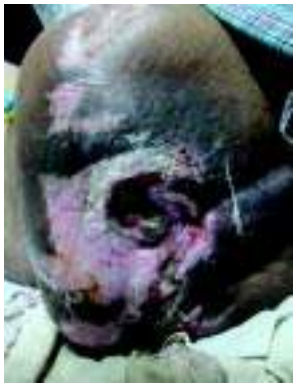
Pressure Sore – Stage 2



Pressure Sore – Stage 3



Pressure Sore – Stage 4



Pressure Sore – Stage 5

- Paradoxical movement
- Use of accessory muscles (sternocleido-mastoid, scalenes, trapezius)
- Rate and depth of respiration
- Chest expansion measurements, at axilla, at nipple level and at xiphisternum
- Breath sounds (air entry and added sounds)
- Assessment of ability to cough

Assessment of intugementary system

Occiput, scapulae, elbows, spinous process, sacrum, coccyx, ischial tuberosities, greater trochanters, lateral and medial femoral condyles, lateral and medial malleoli andheels are vulnerable to develop pressure sores.

These areas have a greater potential for breakdown, due to sustained loading in one posture, compounded by sensory loss makes these areas vulnerable.

Poor nutrition, smoking, swelling, incontinence and lack of pressure relief mechanisms are potential causes for skin breakdown.

In acute stage, chest physiotherapy is very vital. In first 3 months post SCI, death occurs mainly due to pulmonary complications.

Paralysis of trunk and extremity muscles causes immobility and due to abdominal muscle weakness, coughing is severely affected.

Loss of innervations of intercostal muscles and decreased diaphragm movement may result in decreased lung volumes.

Sympathetic cardiac innervation is lost making postural changes more precarious. There is decreased vital capacity, total lung capacity, expiratory reserve volume, and FEV1.

Use of accessory muscles for respiration is increased. wraps for the extremities, a G suit and Mas trousers may be used to minimize orthostatic hypotension until vasomotor control is established.

Due to reduced vital capacity, there is decreased deep breathing and inability to cough, inability to change position which are the causes for the retention of secretions, atelactasis and pneumonia. Assisted breathing exercises and coughing can help in the prevention of these complications.

Stryker frame is used for positioning and turning.

Percussions and vibrations are given to the anterior, lateral and posterior aspects of the chest wall.

Abdominal support given whilst coughing and huffing can also help.

Exercises for Sternocleidomastoid, trapezius,levator scapulae,platysma are given as they help to increase AP diameter and tidal volume.

Glossopharyngeal breathing

a. The mouth and throat are filled with air, the tongue, jaw and larynx are depressed.
b. The lips are closed and the soft palate is raised to trap the air.
c. The larynx is opened, the jaw and floor of the mouth and larynx are then raised.
d. With repeated motion of the tongue air is forced through the opened larynx in the trachea.
e. The larynx is closed and air is trapped in the trachea and lungs.
f. 14-48 strokes / min, can supplement air for effective coughing and secretion clearance.

Summed breathing

This may increase tidal volume. It is carried out by encouraging patient to take several quick shallow but cumulative breaths before expiration.

THE SUB-ACUTE STAGE (IN THE WARD)

BED MOBILITY

1) Rolling

Steps:

- Gaining momentum by swinging upper limbs side to side.
- Turning the head in direction of the movement, interlocking arms if weak triceps in one arm.
- Inhaling with extension, exhaling with flexion helps.
- Bending hip, knee and internal rotation helps, crossing legs over also helps.
- Weights cuffs (2-3lbs), bed rails and loops at the side of the bed would help.
- Air splints may hold the patients elbow straight as he / she swings arms.
- For log rolling, wedges and pillows can be used.
- Also draw sheet rolling starting from side-lying can also be done.

Rolling done in Group Therapy

2) Supine on elbows

Steps:

- Roll to one side and come up on bottom elbow. As rolling is done back to supine position, quickly extend the top arm and place the elbow under the shoulder. Shift weight onto the extremity and position the other elbow under the shoulder, good speed and timing are requisite for this.
- Place hands under their hips or hook thumbs into pockets. By using wrist extensors or their biceps to pull half way up. Then as they shift their weight from side to side they are able to reposition their elbows under their shoulders.
- Momentum of forcefully flexing arms and head to curl forward and then quickly throw elbow back behind the shoulders.

3) Supine/prone to long sitting

Steps:

- From supine on elbows, rocking from side to side attempting to build momentum, throw one arm behind and immediately shift weight onto this extended arm.
- Next throw the other arm behind and walk up on extended arms to a sitting position (can be maintained by pectoralis muscles with shoulder placed in extension and external rotation).
- If prone on elbows used, curled into C-curve, one arm is hooked under knee, pull with this arm, other arm flinged behind, locking the elbow into extension.
- The first arm is then thrown behind and patient walks with the extended arms the rest of the way up.

4) Prone on Elbows

Steps:

- Hands near shoulders, elbows close to the trunk, push of elbows on the mat and lifting upper trunk.
- Final position
  a. either patient shifts weight from side to side moving elbows up under the shoulders.
  b. pushes entire body backwards until elbows are under the shoulders.

SITTING STATIC

Long leg sitting on the bed is graded as follows:

1 **Poor minus,** requires maximum assistance to maintain static position

2 **Poor** requires moderate assistance to maintain static position

3 **Poor+** requires minimal assistance to maintain static position

4 **Fair minus,** requires contact guard to maintain static position

5 **Fair** maintains static position with closed supervision(< 2 minutes)

6 **Fair+** maintains static position with closed supervision,> 2 minutes.

7 **Good minus**, maintains static position against minimal resistance.

8 **Good** maintains static position against moderate resistance.

9 **Good+** maintains static position against maximal resistance.

DYNAMIC BALANCE

1) **Poor minus :** requires maximal assistance to move on either sides, unable to move voluntarily from the midline.

2) **Poor:** able to move through 25-50 % range, requires maximal assistance to return.

3) **Poor plus :** able to move through 50 % range, requires maximal assistance to return.

4) **Fair minus** : able to move through 50-75 % range, requires contact guard to return.

5) **Fair:** able to move through 75 % range with contact guard or 50-75% with closed supervision

6) **Fair plus :** able to move through full range with contact guard in all directions.

7) **Good minus:** independent in functional dynamic balance activities.

8) **Good:** independent in functional dynamic balance activities.

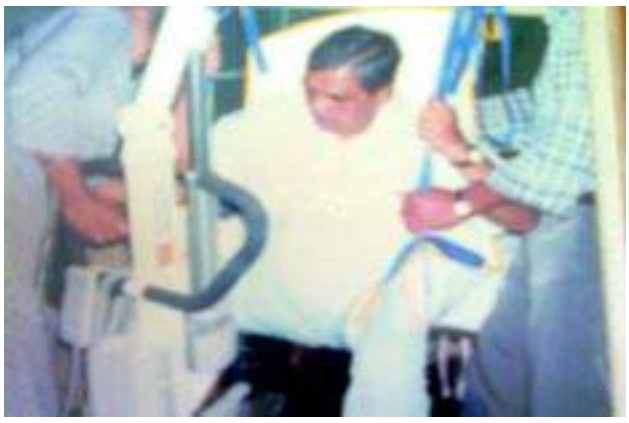
9) **Good plus:** independent in high level dynamic balance activities.

TRANSFERS

Transfers / transitions is the manner in which you move from one place to another, in bed, from bed to wheel-chair, from bed to furniture, wheel-chair to commode, wheelchair to car / vehicle.

For person with tetraplegia, sliding board transfers should be done first, antero-posterior transfers should be tried, so also lateral transfers, a wheel-chair with removable arm rest is useful for the sideways transfer.

Where the arm strength is very weak to do a push-up lift, assistance through sling, by one person or two persons assist is done. Mechanical and mechanized hoist also are safe lifting and transfer devices.



Lifters

Weight relieving maneuvers

1) Lifting leg through a thigh loop, holding this position for half to one minute.

2) Slump over feet, hooking handle or holding onto loops, sustain for half to one minute.

2) Leaning onto sides, holding arm rest of wheel-chair, or removing arm rest and leaning on cot, sustain position for half to one minute.

FIGURE4

4) Tilt back in a manual wheel-chair with assistance, or in a powered chair, hold for a minute.

5) If there is strength in triceps, do push- ups in wheel-chair, holding the tyres, or arm rests and sustain the push-up for one minute. To wear mittens to prevent injury to hand and callosities.

Along with pressure relieving maneuvers a good cushion, mattress will aid in pressure relief, and prevent skin breakdown.

Cushions come in different forms-

a) contoured foam

b) contoured foam with gel filled pad.

c) Viscoelastic foam with casing.

d) Polyurethane foam.

e) Air filled villous roho cushion

f) Water filled seat cushion

g) Low cost cardboard cushion.

FIGURE5

DEEP BREATHING EXERCISES

Goal is to strengthen diaphragm, intercostals if innervated, or accessory muscles for a deeper inhalation or to focus on intercostals if inervated for a more forceful exhalation needed for a productive cough.

Diaphragm position for maximum contraction-

Patient is supine with a pillow, and posterior pelvic tilt, with arms resting at the sides in shoulder adduction and internal rotation will slacken the accessory muscles and enhance diaphragmatic function. Patient is told to take deep breath from the belly, and to relax the shoulders, also therapist can provide firm manual pressure to the shoulders in the inferior direction. Stretch just before inspiration as in costophrenic cough technique. Weights can be used on the abdomen, but we should get correct motor performance, and discourage accessory muscle breathing. Later diaphragmatic activities in other functional positions used.

Strengthening the accessory muscles

The patient is instructed to take a deep breath, while raising his shoulders and contracting the innervated

Weight relieving Maneuver -1

Weight relieving Maneuver-2

Weight relieving Maneuver-3

Weight relieving Maneuver-4

Weight relieving Maneuver-5

Weight relieving Maneuver-6

Cushion-1

Cushion-2

muscles in the neck and pectoral regions. Position further facilitated by not using a pillow, shoulders in abduction and external rotation, and anterior pelvic tilt, mirror can be used to give feedback. Therapist can apply manual pressure at exhalation to inhibit diaphragmatic contraction. Prone on elbows is a diaphragmatic inhibiting position that allows the patient to better use accessory muscles.

Glossopharyngeal breathing

If only accessory muscles used, it is tiring. Patient should be taught glossopharyngeal breathing, also called frog breathing or air stacking. Muscles of the tongue, soft palate, pharynx and larynx work together to create a pumping action that forces gulps of air into the trachea and lungs. Patient traps air in a pocket of negative pressure within their mouths, which allows them to maximize that space, pulling in more air, then closing their lips, and forcing the air back and down the throat with stroking maneuvers of the tongue, pharynx and larynx. More than 60 % of patients with high SCI who are unable to breathe without the ventilator, can achieve autonomous breathing for hours or even all day with this technique.

Assisted Cough Techniques

- Heimlich assisted cough
- Costophrenic cough
- Anterior chest compression
- Tetraplegia long-sit assist
- Prone on elbows, self assist

Heimlich assisted cough

It is also called abdominal thrust assist.

i. Therapist places the heel of one hand just proximal to the patient's navel and the xiphoid process.

ii. Instruct the patient to take deep breath or air stack with hold for several seconds.

iii. Instruct the patient to cough, therapist simultaneously applies an anterior and superior force to increase the expiratory effort of the patient.

iv. Also can be done in sitting in a wheel-chair or when in a postural drainage position, including supine or side lying (with one hand on the posterior thorax for stability)

Costophrenic Assisted Cough (for those who cannot tolerate Heimlich assisted cough)

i. The therapist places the palmar surface of hands along the costophrenic angles of the rib cage with fingers laterally spread in the direction of the ribs.

ii. Quick stretch is given downwards and inwards on exhalation.

iii. This quick stretch can facilitate an improved contraction of the diaphragm and the intercostals muscles.

iv. After inspiration hold air and instruct to cough.

v. Quickly apply a lateral and inferior manual force to enhance the force of the patients exhaled air flow.

vi. Best position is side-lying.

Anterior chest compression

It is modification of the costophrenic assist, which facilitates the upper chest rather than the lower.

i. The therapist places one hand or entire forearm across the chest wall and the other across the lower chest wall.

ii. As the patient coughs, the lower hand or forearm still compresses to assist exhalation while the upper hand stabilizes and compresses the upper chest as well.

Counter-rotation Assisted Cough

It is also called Massery's Counter rotation assisted cough technique.

i. The counter-rotation assisted cough compresses the thorax in 3 planes for a maximal exhalation.

ii. It is performed in side-lying, for patients where there is no contraindication for spinal rotation.

iii. When patient is left side-lying with 45 degrees of hip flexion, therapist kneels behind the patient diagonally facing patient's shoulder.

iv. The therapist's left hand should be on patient's right scapula and right hand on patient's ASIS.

v. Patient is instructed to take a deep breath, therapist pushes the upper thorax superiorly and anteriorly with the left hand and pulls the pelvis inferiorly and posteriorly, with the right hand the therapist pulls the upper chest inferiorly and posteriorly with the left hand and simultaneously pushes the gluteal region superiorly and anteriorly with the right, patient is told to cough.

vi. This maneuver functions in the same manner as innervated intercostal muscles.

Tetraplegia long sitting self-assist

i. This is performed in long-sitting position or with legs externally rotated and flexed.

ii. The patient is instructed to take deep breath with cues to extend neck, scapular retraction, then instructed to cough while bringing head forwards to assist with upper chest compression.

iii. The short-sitting self assist is performed with the patient sitting in a wheel-chair or on side of bed or mat.

iv. Same process as above, hands interlocked, forearms in the Heimlich position press inward and upward on abdomen to assist with forced exhalation.

v. Patient can try it in prone position if it is not contraindicated.

vi. Also in quadruped position, but requires more balance and motor control.

vii. Instructed to rock forwards extending his head and trunk as he inhales.

viii. Then rocks back, bringing hips posterior and inferior and flexing trunk as he` coughs.

Role of Surgery (Tendon transfers)

1) Biceps to triceps transfer for elbow extension.

2) Brachioradialis to radial wrist extensor with FPL tenodesis. (Mobergs tenodesis)

3) Brachioradialis to flexor pollicis longus for active pinch.

4) Radial wrist extensor to flexor digitorum profundus.\

5) Transfer of posterior deltoid to triceps

Above transfers help in increasing pinch force, improve hand function, and activities of daily living, eliminates need of multiple adaptive equipments, restores reachable work space with the biceps transfer, eliminates reliance on shoulder external rotation and locking of the elbow joint, improves proximal control. In incomplete tetraplegia may help in holding walker / crutches.

DON'TS FOR PERSON WITH SCI

1) Do not exercise vigorously or jerkily (increase of spasticity, myositis ossificans, sprains, fractures.

2) Do not do hot water fomentation (burns)

3) Do not lie on hard bed, sit on hard chair without cushion (sores)

4) Do not stay immobile in one position for too long (sores, swelling, stiffness)

5) Do not sit for too long without support, (back pain)

6) Do not lie too curled up, in a flexed position (contractures and deformities)

7) Do not sit with legs dangling (swelling and contractures)

8) Do not put on weight (mobility severly hampered)

9) Do not change position abruptly, (postural hypotension)

10) If using axillary crutches, do not hang on them (neuropraxia)

11) Do not use improper aids (neck, back pain)

12) Do not wear ill fitting orthosis / worn out rubber tips, (abrasions, risk of falls)

13) Do not stay in sun too long (autonomic dysrefexia)

14) Do not smoke, consume alcohol (decreased cardio-respiratory endurance)

15) Do not travel without proper footwear, socks, wounds, burns.

16) Do not cut down on your water intake.

17) Do not seek advise from those who are not professionals.

DO'S FOR PERSONS WITH SCI

1) Exercise daily, put the joints, muscles under active control through full range of motion, 20 repetitions twice a day. (upper limbs can help the lower limb, stronger. Upper limb may assist the weaker limb, or assistive devices are used. Do warm up slowly and after exercise cool down, do gentle stretches.

2) If unwell do gentle ROM.

3) Do deep breathing exercises, twice daily, also huffing and coughing.

4) Do lie with a good posture, sit with an erect posture.

5) Do strengthen the whole body, head, neck, trunk, upper and lower limbs (whatever possible)

6) Inspect body parts twice daily, particularly bony prominences using a long handled mirror.

7) Do lie on soft, firm (not sagging) bed, do use soft cushion when you sit

8) Do sit up / stand slowly and lower yourself.

9) Do push-ups in prone, sitting, and if possible in standing.

10) Do roll on your sides every two hourly.

11) Do weight shifting and change of position frequently, every 15 minutes for 15 seconds to 1minute.

12) Do activities that improve trunk balance and co-ordination, especially of the trunk.

13) Do improve protective extension of arms in all positions.

14) Do your shifting in bed, chair and transfers smoothly, with adequate clearance, lift.

15) Do inspect aids, daily and see that they are in proper condition.

16) Do go out of the house daily / weekly by self or with help. If trunk balance is poor see that straps are fastened when on W / C.

17) Do be in the morning sun, in the house or outside (15mins to 30 mins), avoid afternoon sun.

18) Take part in sports, ball games. Exercise individually, also in group sessions.

19) Wear mittens when propelling a W / C.

20) Do seek advise from professionals only.

Lifting Principles and Musculo-Skeletal Care

The objective of any lift done with and for a patient is to change his or her position or place with ease and comfort to the person being lifted and the lifter, alone or with the help of other persons, with or without adaptations. The therapist should:

1) Let patients participate as much as they can, during the lift and shift.

2) Arch backwards few times before lifting.

3) Before lifting, bend hip, knee and ankle joints few times.

4) Work over bend hip, knee and ankles, vertebral column should be bending as little as possible.

5) Maintain lordosis while lifting.

6) Get close to the person and object they are lifting.

7) Keep a broad base while lifting.

8) Do dynamic bracing (lower abdomen contracted with a pull upwards and towards the sides of the waist).

9) Avoid jerky movements, leg movements should be fluid.

10) Lift in the direction of the movement.

When we lift a person (with help), we lift ½ of persons weight and 70% of our own body weight (HAT head, arms and trunk),multiplied by the distance from the body, multiplied by the degrees of bending(at 45 degrees multiplied by 5), also if rotation in the lift, load on the spine doubles.

If weight 20 cms from the body, moment at the lumbar spine	60 Nm.
If weight 40 cms from the body, Moment at Lumbar spine is	80 Nm.
Bending moment with spine straight, hips and knees bend	151Nm.
Bending with spine bend, knees straight	192.5 Nm.
Bending with spine bend, hips and knees bend, load away	212.5 Nm.

With trunk flexed, knees straight, force on LS junction goes up to 750 kg. Thus bending at hips and knees with a straight spine creates an air cushion in the abdomino-thoracic cavity unloading the spine.

Nachemsons chart:

- supine lying - 25,
- standing - 100,
- sitting with back rest - 150,
- sitting leaning forward - 175,
- knees straight - 200.

Bracing, lateral, antero-posterior rocking facilitate shifting and lifting, Gaining momentum aids the lift. Safe position of lifter / s and the person being lifted is very important. The choice of the lift, how many persons would be involved, who is holding where, the direction of movement, clear instructions and commands should be given., the start of the movement, the transit and the lowering should be smooth.

Aids to lifting/ shifting.

Grab rails, overhead bars, bed-ladder, draw sheet, push-up blocks, transfer boards, slings, belts, loops, manual or mechanized hoist.

TYPES OF LIFTS

Through arm lift.

Lifter

For patients who slump in sitting, a lifter alone and for moving up in bed, into and out of bath.

Orthodox lift.

Only should be used when a patient may not have pressure on the axilla or chest wall, as following mastectomy or when totally helpless.

Half-shoulder/ half orthodox lift.

When it is impossible because of patients condition to use shoulder lift on one side, then it may be necessary to use the orthodox lift on that side, while the shoulder lift is used on the opposite side (could be avoided, an oblique lift)

Grasps

Finger grasp, double wrist grasp, double forearm grasp, single wrist grasp, relaxed hand grasp, through arm grasp, axillary grasp,, elbow grasp, waistband grasp, buttock grasp, forearm grasp, palm-to-palm thumb grasp.

Stances for lifting and supporting

Walk standing

Lunge standing.

Stride standing.

Ten to two standing.

Ramps

Ramp

For wheel chair users, the incline / slope gradient is a very important consideration. For every inch of vertical rise there should be 12-14 inches of length.

This gradient is possible for average riders and strong persons with quadriplegia. This is the best slope for public buildings and rehab centres.

There should be width of 36 inches to 48 inches (91.4 -121.9 cms), continuous bilateral hand rails with height of 34 -38 inches situated 1.5 cms (3.8 cms) from the wall, with circular cross section of 1.25 -2 inches (3.2 -5.1 cms), with hand railings extending 12 inches (30.5 cms) beyond the top and bottom of the ramp.

There should be a non-skid surface and for a long and curved ramp there must be a level platform at 10 feet intervals.

If for each inch of vertical rise, there is length of 10 inches, it is a fairly steep slope and possible for riders with strong arms and person with paraplegia who are strong.

If only 6 inches length for each inch of vertical rise, it is a very steep slope, only possible for those using a powered chair or with help, rarely possible for rider alone, chair may tip backwards.

Orthosis

- Hard Cervical Collar (Thomas's Collar)
- Philadelphia Collar
- Sterno-Occipital Mandib
- Ular Immobilzer(SOMI),
- Poster Type Cervicothoracic Orthosis
- Minerva Cervicothoracic Orthosis

- Cuirass Type Cervico-Thoracic Orthosis
- Yale Orthosis
- Halo-Vest Orthosis
- Opponens Orthosis
- Opponens Orthosis With Lumbrical Bar
- Wrist Control Orthosis,To Assisting Tenodesis
- Universal Cuff
- Tailors Brace


Philadelphia Collar

Wheel-chairs

Wheelchair should be appropriate for the patients needs. Trial should be done indoors and outdoors in its safe use by patient and care-givers. Patient should learn skills of maneuvering the wheel-chair, manual for persons with paraplegia, with projections on outer rim, wheel or a powered chair for person with tetraplegia. Patient should learn some mode of transfer to and from the wheel-chair, and if possible learn assembly, disassembly, and maintenance. (Lubrication, and charging batteries of powered chairs)

Weight >23 kg for person weighing 114 kg, seat width 20”

Standard 18-23 kg, Lightweight 9-18 kg, Ultra light weight (sports wheel-chair) 7-9 kg.

Measurements of wheelchair

1) Back height distance from buttocks to level of scapulae.
2) Arm rest distance from buttocks to olecranon, elbows flexed 90 degrees.
3) Seat depth distance from back of buttocks to popliteal fold, add 1-2 inches.
4) Seat height distance from heel to popliteal fold, add 2 inches to provide clearance of foot rests when approaching slope, ramps.
5) Seat width widest distance across hips, add1-2 inches for clothing, splints

Types of Wheelchairs:

- Conventional wheelchair, non folding, fixed arm rests, small castors.
- Folding, removable arm rests, flipawayor elevating foot rests,
- Semireclining wheel-chair.
- Reclining wheel-chair
- Stand-up wheel-chair
- Manual wheel-chair with add on power unit.
- Electric wheel-chairs
- Tricycles
- Electric carts.
- Adapted vehicles.



Wheelchairs

Ambulation

The physiotherapist should be involved with training skills to gain independence through wheel-chair and later introduce ambulation if a person with incomplete tetraplegia wishes to do so.

Standing and weight-bearing have to be given to all whether injury is partial or complete, to prevent osteoporosis, prevent bladder and bowel complications, elevate their spirits and mood.

Let the person with tetraplegia(incomplete) decide whether ambulation is practical or not, tilt-table standing is done gradually.

Gait training should begin between parallel bars and with a mirror in front for postural reeducation.

Walking is begun in parallel bars with, KAFO's and abdominal binder / spinal brace, later one bar and one crutch, later bilateral crutches, if possible.

A modified walker, forearm gutter walker, a walker without wheels, or a wheeled-walker can be used.

Walking with HKAFO and spinal braces increases the energy cost of walking and is only possible for young athletic persons, for functional needs wheelchair becomes more practical.

If possible we should teach the skills of wearing and removing the appliances, standing from bed, lowering to bed from standing, walking forwards, sideways, backwards, going up and down small gradients, if possible stair climbing, and falling and rising.

Bladder management

1) During the acute phase over distension of the bladder is prevented by an indwelling catheter.
2) After 2-4 weeks, intermittent catheterization may be tried. Proper aseptic precautions should be followed, it needs to be done 4-6 times a day. For those who have finger control and power (C7)are able to do self catherisation, also C6 level quadriplegics may be able to do with tenodesis grip.
3) When the injury is above T12 level, there is spastic or reflex bladder, and below T12 / L1, there is flaccid / non-reflex bladder.
4) To avoid problems, we should not allow more than 400 cc of urine to fill in the bladder at any one point.
5) A voiding diary should be kept, a diary of water intake versus urine output.
6) Between catheterizations do not drink more than what your bladder will hold(<400cc).
7) A daily intake of 2 to 2.5 litres of fluid is sufficient.
8) Avoid drinking at bedtime, unless, patient or care-giver is going to catheterize.
9) Bladder stimulation by suprapubic jabbing may help, by pressing deep over the bladder to mechanically stretch its wall, rather than suprapubic tapping,or stroking or pinching the perineal skin.
10) Transurethral electrical bladder stimulation activates mechanoreceptor afferents which restore the sensation of bladder filling and in turn activate efferent nerves, resulting in detrusor contraction.
11) Valsalva manouvere- increase intravescical pressure by increasing intra abdominal pressure. It involves sitting and resting abdomen forward on the thighs for both men and women. During straining, hugging of the knees may prevent bulge of abdomen,.All of the increase in intra abdominal pressure is transferred to the bladder and pelvic floor. (in patients with vescicoureteral reflux, it is contra indicated. adverse effects may also include hemorrhoids, rectal prolapse, or hernia).
12) Crede's maneuvers increase intravescical pressure by manually pushing down on the bladder.
13) In the open hand credes maneuver, the thumb of each hand is placed over the left and right ASIS and the digits over the suprapubic area with slight overlapping of the tips. The slightly overlapped digits are then pressed into the abdomen. When well behind the symphysis, the pressure is directed downward to compress the fundus of the bladder. Both hands are then pressed as deeply as possible down ward into the pelvic cavity.
14) In the closed-hand Crede's method, compression of the bladder is done by using closed fist or a rolled-up towel.
15) Kegels exercises can be done in women.
    a. Females can use diapers,
    b. men can use condom catheters.
16) In those where there are complications of bladder stones, hematuria, bactermia, penile

and scrotal fistulas, uretheral stricture, uretheral diverticulum, bladder carcinoma, suprapubic catherization is preferrable.

17) Whenever there is urinary infection, appropriate antibiotics are given along with plenty of citrus fruits.

Bowel Care

1) Have a well-balanced fibre rich diet, with roughage. whole grain cereals, fruits with skin, green leafy vegetables.
2) Drink plenty of fluids.
3) Stay active.
4) Stick to a scheduled bowel program
5) Automatic bowel responds to a suppository or stimulation by a finger..
6) Preferably sit on a toilet seat / commode for 15 minutes after taking suppository, if you cannot, insert it while lying on your left side, use a gloved lubricated finger, push suppository 2 cm in anus, drink a cup of hot water, tea / coffee / lime juice or do after food, as gastro-colic reflex helps bowel movement / peristalsis.
7) Sitting time should not exceed 30 minutes, massing the abdomen from right to left and down several times helps.
8) If suppositories do not help then only do digital stimulation.

In summary, a physiotherapist's role is very vital in the total well-being of person with tetraplegia, right from the site of injury, in the intensive care unit, in the step down unit, in the home setting, and the community by optimizing functional skills. The therapist is cognizant and prevents negative effects of cardio-respiratory de-conditioning, prevents pneumonia, atelectasis, deep vein thrombosis, pressure sores, contractures and deformities, renal calculi and osteoporosis, and postural hypotension through graduated verticalisation and weight-bearing. Therapist later on strengthens all the innervated muscles, keeps up the mobility in the joints where active movement is not possible, enhances bed mobility skills, and transfers with / without assistive devices, improves dynamic sitting balance, wheel-chair skills for indoors and outdoors and also for getting in and out of vehicle,, weight relieving maneuvers, in persons with incomplete tetraplegia household ambulation with modified walkers / crutches, and lower limb orthosis, with abdominal binder, and if possible outdoor walking skills.

Besides physically helping, supporting the person with tetraplegia to gain functional independence in attaining motor skills, therapist cheers, boosts the morale, raises the spirit, infuses positivity in the person with tetraplegia, their families and the care-givers. The Physiotherapists should strive hard for giving them independence in ADLs like bed mobility skills, transfers, wheel chair mobility, bladder and bowel management rather than stressing on ambulation, which is more time and energy consuming.

OCCUPATIONAL THERAPY IN QUADRIPLEGIA

OT in Quadriplegia

Quadriplegia is defined as the partial or complete paralysis of all four limbs and trunk including respiratory muscles, as a result of damage to cervical spine.

Occupational therapy assessment

Regardless of where the patient begins in the rehabilitation process, an assessment is always completed on admission. The assessment will help in establishing a diagnosis as well as determining the most appropriate therapeutic intervention.

History Taking

The first step involves review of medical record to gather background information and identify medical precautions. The history should include demographic data, social history, occupational & leisure history, past and present medical & surgical history, associated conditions, social habits and allergies. Even though most of the information is already noted in the medical file, asking the patient about his hobbies, work or family often opens communication and helps establish a rapport between the therapist and the patient. Therapists conduct a Qual-OT assessment to identify which areas of potential quality of life are most important to the patient.

Physical Status

Before evaluating the physical status of the patient, the therapist should first obtain specific medical precautions from the physicians. The physical status includes:

- **Muscle tone & Deep tendon Reflexes**

  Muscle tone should be evaluated with reference to the quality, muscles groups involved and any factors that appear to increase or decrease tone. It is evaluated using the modified Ashworth Scale. Deep tendon reflexes evaluation is also done. The most common examined are the biceps, triceps, extensor carpi radialis longus, quadriceps and gastrocnemius.

- **Range of Motion**

  Range of motion evaluation includes measurements of all extremity joints and all digits. It should be measured before muscle testing to determine available pain–free range. It helps to identify if there are any contractures or potential to develop the same. In cases of spinal instability caution should be used while performing any movement.

- **Joint Integrity & mobility**

  The therapist should check for any soft tissue swelling, inflammation or restriction. Also joint hyper or hypo mobility should be checked for and noted.

- **Muscle strength**

  Manual muscle testing includes testing of the muscles of the scapula, shoulder, elbow, wrist, and digits, as well as grip and pinch strength measurements.

- **Sensation**

  Sensory evaluation of all dermatomes of the upper body includes evaluation for light touch, pin prick, joint proprioception, stereognosis, and kinesthesia. Sensation is indicated as intact, impaired or absent per dermatome. This helps in establishing the level of injury and to determine functional limitations. Using a dermatomal map aids in easy documentation.

- **Wrist and Hand function**

  A wrist and hand function evaluation determines the degree to which the patient can manipulate objects. This information helps the therapist to suggest the required splints (e.g. tenodesis splint) which will aid hand function. Sollerman Hand Test is a standardised hand function test based on seven of the eight most common hand grips. The test consists of 20 activities of daily living.

- **Cognitive and perceptual evaluation**

  If a head injury is also suspected, then the assessment also includes the patient's ability to initiate tasks, follow directions, carry over learning day to day, and do problem solving.

- Clinical observation is used to evaluate posture & trunk control, endurance, lower limb functional strength and total body function and more specific evaluation may be required depending on the individual.

Functional Status

A detailed functional evaluation is usually not carried out until the patient is in the active or rehabilitative phase where the patient will be medically stable. In the acute phase a functional evaluation includes performing light activities of daily living (ADLs) such as feeding, light hygiene and object manipulation, to determine present and potential levels of functional ability and should begin as soon as the patient is cleared of bedrest precautions, depending on the level of injury. During the rehabilitation phase performances in the following areas are assessed:-

ADLs

ADLs are the basic tasks performed on a daily basis in order to engage in daily routine. It includes bathing, toileting, dressing, getting in and out of bed or a chair, hygiene, and eating skills. Occupational therapy scales like FIM, Spinal Cord Independence Measure (SCIM) & Quadriplegic Index of Function (QIF) evaluate different areas of function. The QIF assesses 10 ADL's:

1. Transfers
2. Grooming
3. Bathing
4. Dressing
5. Feeding
6. Mobility
7. Bed activities
8. Bladder program
9. Bowel program
10. Personal care

The QIF was specifically designed for SCI patients and is focused on persons with tetraplegia

Environmental Barriers

Barriers present in the patients home and work environment can be identified using questionnaires or interviews. Also photographs or video recording of the particular environment aid in identifying barriers.

Other evaluations include assessment of performance in areas such as:

1. Vocation
2. Access to home and community
3. Leisure time activities
4. Driving
5. Appropriateness of treatment and equipment.

Goals

Once the patient is evaluated, goal planning is started. Goal planning is based on patient involvement as an active participant and not therapist-led practice, with the recognition and the utilization of the patient's strengths to meet identified needs significant to the patient in regard to his perception of his spinal cord injury. Goals are then set within the rehabilitation team with specific, measurable and realistic targets to be achieved in an agreed time. Regular review and monitoring success is necessary.

Some general goals of occupational therapy intervention are:

- Maintain / achieve full ROM in all joints
- Prevent deformities
- Increase strength of innervated or partially innervated musculature and increase physical endurance
- Achieve maximal level of functional independence
- To explore leisure interests and vocational potential
- To train the patient in use and care of necessary equipment.

Functional Expectations

The following are the anticipated functional abilities and limitations that a patient with quadriplegia may achieve at each level:-

C1 - 3

- Innervated facial and neck muscles permit chewing, swallowing, talking, blowing, and neck control, which permit:
  - Instructing others in care for pressure reliefs, skin precautions, upper extremity ROM exercises, equipment setup and maintenance, positioning, etc.
  - Independently propelling a power wheelchair with portable respirator and chin or breath control on hard, level surfaces
  - Operating communication devices, computers and environmental control systems electronic page turners, with head pointer, mouthstick or pneumatic control
  - Leisure activities, such as computer and electronic games, reading and painting
  - Vocational skills involving use of a computer
- Total paralysis of the trunk, UEs and LEs dictate:
  - Total ADL and respiratory dependence
  - Full-time attendant care

C4

- Innervated diaphragm(C3-5), trapezius, cervical and paraspinal muscles permit respiration, scapular elevation, and neck movements, which permit:
  - Limited self-feeding with a long straw and straw holder and externally powered flexor hinge splints and mobile arm supports
  - Propelling power wheelchair on uneven terrain using head, mouth, chin, breath or sip-and-puff controls
  - Note taking in school or business with an adapted tape recorder
  - Writing / art using pencil attached to mouthstick
  - Using a mobile with speaker phone and mouthstick
- Paralysis of trunk, UEs and LEs dictate:
  - Full-time wheelchair use
  - Part- or full-time respiratory assistance
  - Full-time attendant care

C5

- Innervated shoulder muscles, biceps, brachialis, brachioradialis, supinator,

infraspinatus, rhomboids and deltoid permit shoulder external rotation, abduction to 90° and limited flexion; elbow flexion and supination; and gravity assisted shoulder adduction, pronation, and internal rotation, which permit:

- Self-feeding with a mobile arm support or suspension sling, dorsal wrist splint with u-cuff, and other devices such as a plate guard or scoop dish, stabilized cup or cup holder, straw with holder and an angled spoon or fork
- Limited Upper Extremity dressing and fully dependent in lower extremity dressing
- Self-grooming, with a wash mitt and a quad grip long handle sponge/ hairbrush/toothbrush, to aid in face washing, upper extremity bathing, hair brushing, teeth brushing, with moderate assistance
- Maximum assistance with lower extremity bathing
- Upper extremity ROM with maximum assistance
- Propelling hand-controlled power wheelchair over obstacles with assistance, and manual wheelchair with projection knobs on level surfaces for short distances
- Pressure relief with power recline or tilt wheelchair with elbow or head switches
- Typing stick in dorsal wrist splint with u-cuff for operating computers and long writing orthosis for writing, turning book pages using a book holder stick in dorsal wrist splint with u-cuff.
- Driving a van with Hand controls

• Absence of elbow extension and pronation, all wrist and hand movements, total paralysis of trunk and LEs and Low endurance and low respiratory reserve result in:

- Inability to roll over or come to sitting without hospital bed with rails
- Assistance needed for transfers, skin inspection, bowel and bladder management
- Atleast part-time attendant care

C6

• Innervation of the pectoralis major(C5-8,T1), serratus anterior(C5-7), lattisimus dorsi(C6-8), pronator teres(C6,7), and Extensor carpi radialis longus and brevis(C6-7) permit strength for all shoulder movements, elbow flexion, forearm pronation and supination, and radial wrist extension (tenodesis grasp), which permit:

- Use of a tenodesis splint or universal cuff for increased ease and independence
- Self-feeding with a cup with large handles, a rocker or sharp paring knife for cutting, and does not need straws and plate guard
- Grooming, including teeth brushing, makeup application, and hair care, with tenodesis grasp and/or modified equipment
- Bathing, with a shower/tub bench, faucet within easy reach, and hand-held shower hose
- Typing, telephoning, turning appliances and equipment on and off
- Independent skin inspection and pressure reliefs when sitting using loops and forward weight shift
- Assist in bowel and bladder care,
- Dressing of upper extremity and pants in bed (using momentum and substitute movements to turn over, sit up and pull up clothing) or button hook and zipper pull, with maximum assistance required for shoes and socks
- Upper extremity ROM
- Propelling manual wheelchair, with friction material or rims on uneven surfaces for short or moderate length distances; moderate assistance needed for architectural obstacles
- Independent transfers with a transfer board and partial depression or swivel transfer
- Rolling and moving from supine to long-sitting with loops suspended over the bed or by grasping the bedrail
- Driving using hand controls with U-shaped Cuff attached to steering wheel

  - Light kitchen activities in a wheelchair-accessible kitchen
  - Use of specially Adapted electronic machines in offices
  - Participating in leisure activities like watching TV using the remote, listening to radio and playing adapted table top games or sports, such as quad rugby, swimming
- Absence of wrist flexion and total paralysis of the trunk and LEs result in the need for a part-time attendant for:
  - Some assistance with dressing
  - Moderate assistance for negotiating obstacles in a wheelchair and positioning
  - Maximum assistance with heavy work

C7

- Innervation of the triceps, extensor carpi ulnaris(C6-8), and flexor carpi radialis, flexor digitorum superficialis and profundus(C7-8, T1) permit elbow extension, wrist flexion and extension and PIP & DIP flexion which permit:
  - Self-care same as C6 except easier dressing, bathing/grooming, and bladder/bowel management
  - Propelling manual wheelchair over inclines and in/out of elevators, with some supervision for rough terrain and some assistance for doors
  - Independent pressure reliefs with push ups
  - Independent transfers to and from bed and wheelchair
  - Independent in Bed mobility, with minimal assistance with padding and positioning
  - Participating in preparation for sexual activity
  - Driving a modified car
  - All communication, recreational, and vocational activities for C6, but with greater ease
- Lack of trunk muscles result in lack of full shoulder stability, weak trunk control, reduced endurance due to low respiratory reserve, and paralysis of the lower extremities.

Acute Phase

Intervention Immediately after the initial evaluation:

1. A daily range of motion program should be started, with gentle, assisted, active movements given to all muscles. Progression is made to unassisted active exercises by encouraging the patient to move his arms independently and functionally within strength, ability, and tolerance levels. Basic reasons for range of motion exercises include a) prevention of contractures, b) prevention of joint pain caused by contractures, c) prevention of joint deformities, and d) mobility of the joints.
2. Total body positioning should be evaluated and instruction to the staff, patient, and family members should be given if necessary.

- **In supine,**

  The patient's shoulders should be externally rotated and abducted to 90°, elbows flexed, forearms supinated, and a pillow placed under the forearms. The upper extremities should be intermittently positioned in 80° of shoulder abduction, external rotation with scapular depression, and full elbow extension to alleviate pain in the shoulders and ROM limitations. In patients with injuries at the C5 level, the forearm should be positioned in pronation to prevent supination contractures

- **In side lying:**

  A pillow should be placed in a vertical position under the thoracic region of the trunk. The upper extremity under the patient should be placed in 100° of shoulder flexion. The elbow should be flexed or extended with the forearm in supination. The upper extremity on top of the patient should be placed in pillows in front of the patient

3. The occupational therapist may also provide the patient with hand splints to maintain proper hand positioning and prevent contractures and joint deformities. Dorsal, rather than ventral splints should be used to allow maximum sensation when the hand is resting on a surface.

- If there is inadequate musculature to support wrist and hands properly for function and/or cosmesis, the wrist should be supported in

extension, with the thumb in opposition. This will maintain the thumb web space and allow the fingers to flex naturally

- If there is at least F+(3+) strength of wrist extension, short opponens splints should be used to maintain the web space and support the thumb in opposition

4. Facilitation of a tenodesis grasp which involves fingers flexion while wrist is maintained in an extended position and extension of fingers while wrist is maintained in a flexed position should be begun during range of motion of the hand.
5. The patient should be provided with a basic environmental control, such as a television, telephone, and nurse call system, based on the patient's capabilities and needs, to promote the patient's sense of self-control and independence
6. Education of the patient is begun, including the importance of skin management, pressure relief, and daily ROM
7. Education of the family is begun, including discussion of anticipated medical equipment, home modifications, and caregiver training should be initiated
8. As the patient becomes medically stable, gentle resisted movements can be gradually introduced as indicated. Strong unilateral exercise for the whole arm involves head movement and is therefore completely avoided till spine is completely stabilized. All movements must be given with carefully graded resistance, avoiding any neck movements. Gentle static neck exercises are given 6 weeks post-injury if there are no contraindications. ADL training should be begun, particularly for patients who are on prolonged bed rest, followed by transfer to a wheelchair and training to tolerate an upright sitting position.

Active Phase

In the active phase emphasis is on maximizing functional independence. Passive and active Range of motion and strengthening the innervated musculature using resistive activities and building endurance should be continued. The use of weights, pulley systems, skateboards, suspension slings, and mobile arm supports are used depending on the patient's strength. Shoulder and scapular muscles are exercised for increasing proximal support. Wrist exercises should emphasize the extensors to maximize natural tenodesis function for functional grasp and release. Assistive devices prescribed to enable efficient performance should be cost effective and not bulky also. Use of assistive devices should be kept to a minimum with an emphasis on modified techniques.

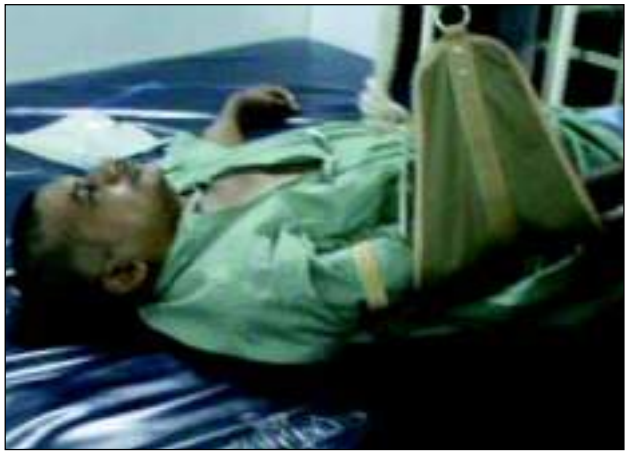


Fig 1 : Patient using suspension for active assisted exercises

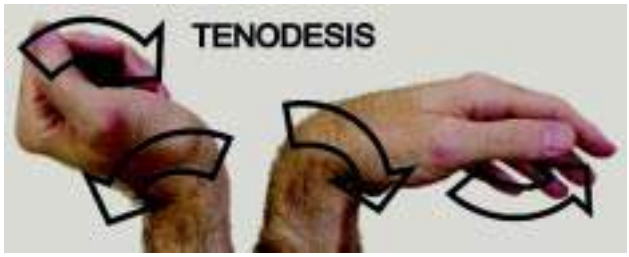


Fig 2 Facilitation of tenodesis action. Courtesy Electronic Textbook of surgery

ADLs

Activities of daily living (ADL) program can be expanded to include independent feeding, oral hygiene, and upper body bathing and dressing, with or without devices. Bowel and bladder care, such as independent stimulation and applying a urinary collection device, can be carried out with or without facilitory equipment, using the best possible technique in the best possible position. Transfers using a sliding board are taught to the patient. Communicating skills can be worked upon via writing & use of the telephone, tape recorder, stereo, and personal computer.

Fig 3 Patient using U-cuff for eating

Fig 4 Use of U-cuff for combing hair & brushing teeth

Fig 5

Fig 7

Fig 7

Fig 8

Fig 5, 6, 7 & 8 Patient performing dressing of upper extremity independently

Mobility

Wheelchair mobility provides high-level quadriplegic persons with one of their most achievable functional activities and also allows them to regain some control over the environment. When prescribing any wheelchair, it is important to consider the patient's living situation, educational and vocational potential, transportation, and maintenance. The occupational therapist has the primary role of informing the patient and the family of options, costs, maintenance record, and transportability of the wheelchair.

Orthosis

Splints are usually required to maintain position, correct a contracture or to encourage function, and can be made from several materials, e.g. plaster of Paris, synthetic plaster materials, thermo-plastics or neoprene. Some patients require two splints: one may need to be worn at night to maintain hand position and another during the day to aid independence. Some examples of the ways in which splints may be used:

- Long paddle splint (from forearm to finger tips with the fingers slightly flexed) for a patient

with a lesion at C4 or above to maintain the hand in a good position and give some control over the arm.

- Wrist extension splint for a patient with a lesion at C5 to inhibit lengthening of the extensor tendons and allow the hand to be used with simple gadgets.
- A patient with a lesion at C6 with wrist extensors less than grade 3, as for C5 above but may also need tapes over the fingers to hold them in finger flexion to encourage the tenodesis position.
- A knuckle duster splint to inhibit metacarpophalangeal hyperextension for a patient with a lesion at C7.
- A splint to encourage opposition of the thumb may be needed by a patient with an incomplete lesion or one at C7.

Leisure

Participation in Leisure activities is necessary as a means of self-expression, release, and socialization for the people with tetraplegia.

- Time should be taken out for leisure activities like viewing TV, listening to music, playing cards, table games, computer-assisted programs, reading materials, outings to the movies, theatre, sports events, sightseeing tours, museums and concerts, shopping, restaurants, and clubs etc.

Vocational rehabilitation

The likelihood of a person with tetraplegia being able to work is strongly dependent upon hand function. Tetraplegics without hand function have less than 2% likelihood of being employed. The recovery of hand function will increase the likelihood of a person working by more than fourfold. Prior education also enhances the likelihood of a person being able to work. (Spinal Cord Injury: Functional Outcomes in 2009 and Beyond Harry M. Koslowski, MD). Vocational therapy begins with a comprehensive vocational evaluation to determine a person's basic skills, including their dexterity and other physical capabilities, as well as their cognitive capabilities. The evaluation process also includes a component that determines changes in physical and cognitive capabilities and interests over time. Once the evaluation is complete, the occupational specialist helps the person to:

- Select and learn to use any assistive equipment they may need to enter or re-enter the work force or engage in another method of productivity.
- Develop personal supports, such as peer-to-peer mentor programs and appropriate interpersonal advocacy
- Identify potential work, educational, or other community environments where the person can be productive;
- Identify and implement the necessary assistive equipment, environmental modifications, task restructuring, task modification, use of coworkers, students, or other members of the self-directed, productive group as personal assistants, etc and
- Develop modified hand movements and assistive devices to compensate for lost hand function

Psychological support:

The therapist provides psychological support by allowing and encouraging the patient to express frustration, anger, fears, and concerns. The therapist also identifies and addresses each patient's psychological problems, such as denial, apathy, depression, etc. Then she identifies and emphasizes on the patient's strengths and skills. Group therapy with similar level of patients or forming of support groups is also encouraged.

Adaptive Equipment:

Following are some of the adaptive equipment suitable to help people with tetraplegia:-

Dressing and Grooming: Dressing sticks, zipper pulls and d-rings, Sock aid, Button hooker, Dressing ladder, Velcro fasteners, Loops in clothes, Long handle sponges and bath mitts, enlarged handles for toothbrushes, combs, razors and hairbrushes and Long handled mirrors.

Eating: Spoons, forks & knives with special handles, Plate guards, long straws, Universal cuffs to hold spoons, forks, or knives

Writing: Special holders for pens & pencils, Special pointing tools / mouthsticks to type on a keyboard

Bathing: rolling shower chair, grab bars, soap on a string, lever type taps

Cooking: Mirror over stove to view into vessel from the wheelchair. Special tools to open jars like reachers, universal cuff, Tenodesis splint

Environmental Control:-This technology allows patients to do things like turn on lights, open doors, answer the phone, and control the temperature. Many of these technologies are "hands-free," allowing the patient to control his or her environment by using puffs of air, voice, head movement, or even blinking. In addition, the home can generally be outfitted so that it operates off of one remote control unit

Driving: hand controls that allow braking and acceleration, easy-touch pads for ignition and shifting, and joysticks and spinner knobs.

Other Adaptive Equipment: Prism glasses, Mouthsticks, Telephone holders or a speaker phone. Special electronic devices for hands-free control of phones and to turn on radios, TVs, lamps, etc

Fig 9 - 17

Care giver Training

An important aspect in rehabilitation is educating the patient and the caregivers in proper handling, transferring and pressure relieving techniques. Energy conservation and joint protection methods are also taught to prevent pain related to overuse.

Complications Following Spinal Cord Injury:

Some of the common complications following SCI are :

- Autonomic dysreflexia
- Deep vein thrombosis
- Heterotopic ossification
- Neuropathic pain
- Osteoporosis
- Spasticity / contractures
- Pressure ulcers

1. AUTONOMIC DYSREFLEXIA

Autonomic dysreflexia (AD) is a syndrome of massive imbalanced reflex sympathetic discharge occurring in patients with spinal cord injury (SCI) above the splanchnic sympathetic outflow (T5-T6). Anthony Bowlby first recognized this syndrome in 1890 when he described profuse sweating and erythematous rash of the head and neck initiated by bladder catheterization in an 18-year-old patient with SCI. Guttmann and Whitteridge completed a full description of the syndrome in 1947.

This condition represents a medical emergency, so recognizing and treating the earliest signs and symptoms efficiently can avoid dangerous sequelae of elevated blood pressure. SCI patients, caregivers, and medical professionals must be knowledgeable about this syndrome and its management.

Epidemiology

Reported prevalence rates vary for Autonomic Dysreflexia (AD) in the United States, but the generally accepted rate is 48-90% of all individuals who are injured at T6 and above. Some incidence has been reported in SCI as low as T10.

The occurrence of AD increases as the patient evolves out of spinal shock. With the return of sacral reflexes, the possibility of AD increases. AD occurs during labor in approximately two thirds of pregnant women with SCI above the level of T6.

The male-to-female ratio for sustaining SCI is 4:1; therefore, autonomic dysreflexia is primarily a male phenomenon.

Etiology

Autonomic dysreflexia (AD) occurs after the phase of spinal shock in which reflexes return. Individuals with injury above the major splanchnic outflow may develop AD. Below the injury, intact peripheral sensory nerves transmit impulses that ascend in the spinothalamic and posterior columns to stimulate sympathetic neurons located in the intermediolateral gray matter of the spinal cord. The inhibitory outflow above the SCI from cerebral vasomotor centers is increased, but it is unable to pass below the block of the SCI.

This large sympathetic outflow causes release of various neurotransmitters (norepinephrine, dopamine-b-hydroxylase, dopamine), causing piloerection, skin pallor, and severe vasoconstriction in arterial vasculature. The result is sudden elevation in blood pressure and vasodilation above the level of injury. Patients commonly have a headache caused by vasodilation of pain-sensitive intracranial vessels.

Vasomotor brainstem reflexes attempt to lower blood pressure by increasing parasympathetic stimulation to the heart through the vagus nerve to cause compensatory bradycardia. The fact that this reflex action cannot compensate for severe vasoconstriction is explained by the Poiseuille formula, which demonstrates that pressure in a tube is affected to the fourth power by a change in radius (vasoconstriction); the pressure is affected only linearly by a change in flow rate (bradycardia). Parasympathetic nerves prevail above the level of

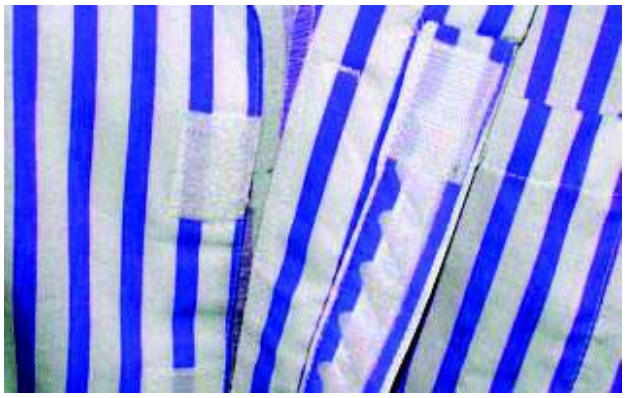


Fig 9: Use of Velcro instead of buttons for clothes

Fig 10: Enlarged handles of eating tools for ease in grasping

Fig 11 Use of mouthstick in operating a computer

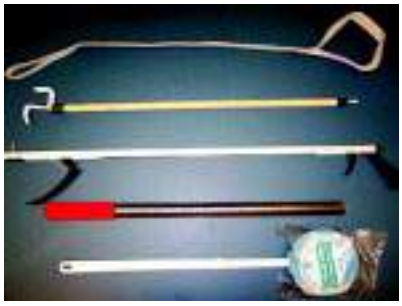


Fig 12 Different types of Reachers

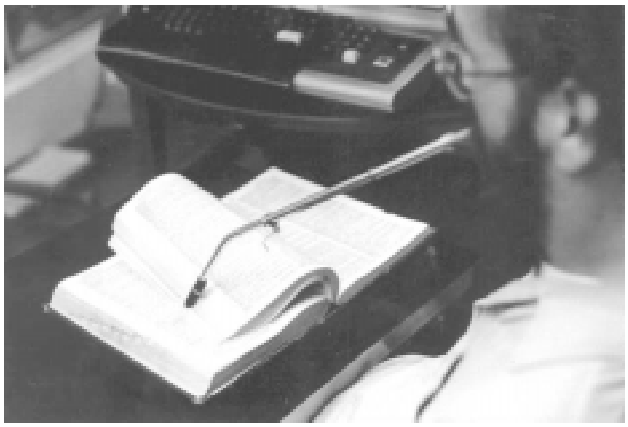


Fig 13 Use of a page turner to promote independence in reading

Fig 14 Example of Adaptive Seating in bathroom & toilet

Fig 15 A example of an adaptive bathroom

Fig 16 A example of a Wheelchair accessible kitchen

Fig 17 A example of a modified car with hand controls

injury, which may be characterized by profuse sweating and vasodilation with skin flushing.

There can be many stimuli that cause autonomic dysreflexia. Anything that would have been painful, uncomfortable or physically irritating before the injury may cause autonomic dysreflexia after the injury.

In general, noxious stimuli (irritants, things which would ordinarily cause pain) to areas of body below the level of spinal injury. Things to consider include:

- Bladder (most common) - from overstretch or irritation of bladder wall
  - Urinary tract infection
  - Urinary retention
  - Blocked catheter
  - Overfilled collection bag
  - Non-compliance with intermittent catheterization program
- Bowel - over distention or irritation
  - Constipation / impaction
  - Distention during bowel program (digital stimulation)
  - Hemorrhoids or anal fissures
  - Infection or irritation (eg. appendicitis)
- Skin-related Disorders
  - Any direct irritant below the level of injury (eg. - prolonged pressure by object in shoe or chair, cut, bruise, abrasion)
  - Pressure sores (decubitus ulcer)
  - Ingrown toenails
  - Burns (eg. - sunburn, burns from using hot water)
  - Tight or restrictive clothing or pressure to skin from sitting on wrinkled clothing
- Sexual Activity
  - Over stimulation during sexual activity [stimuli to the pelvic region which would ordinarily be painful if sensation were present]
  - Menstrual cramps
  - Labor and delivery
- Other
  - Heterotopic ossification ("Myositis ossificans", "Heterotopic bone")
  - Acute abdominal conditions (gastric ulcer, colitis, peritonitis)
  - Skeletal fractures

History and Physical Examination

History

The patient with autonomic dysreflexia (AD) generally gives a history of blurry vision, headaches, and a sense of anxiety. Feelings of apprehension or anxiety over an impending physical problem commonly are exhibited.

Physical Examination

A patient with AD may have 1 or more of the following findings on physical examination:

- Sudden, significant rise in systolic and diastolic blood pressure
- Profuse sweating above the level of lesion - Especially in the face, neck, and shoulders; rarely occurs below the level of the lesion because of sympathetic cholinergic activity
- Goose bumps above, or possibly below, the level of the lesion
- Flushing of the skin above the level of the lesion - Especially in the face, neck, and shoulders; this is a frequent symptom
- Blurred vision
- Spots in the patient's visual field
- Nasal congestion – A common symptom

With regard to the first item above, the sudden rise in blood pressure in AD is usually associated with bradycardia. Normal systolic blood pressure for SCI above T6 is 90-110 mm Hg; blood pressure 20-40 mm Hg above the reference range for such patients may be a sign of AD. However, patients with AD may display no symptoms, despite elevated blood pressure. Differentials for AD include essential hypertension and pheochromocytoma.

PATHOPHYSIOLOGY

Patient exposure to noxious stimuli (pain or pressure below the level of injury)

↓

Signal sent to autonomic nervous system via the sympathetic pathway

↓

Exaggerated Reflex (vasoconstriction and hypertension)

In normal cases	In spinal cord injury patients
↓	↓
Baroreceptors send signal to brainstem	Baroreceptor activity is interrupted
↓	↓
Vasodilatation	No compensatory mechanism
↓	↓
Slowing of heart rate	Elevated Blood pressure and heart rate

Prevention

The following are precautions you can take which may prevent episodes:

- Frequent pressure relief in bed / chair
- Avoidance of sun burn / scalds (avoid overexposure, use of #15 or greater sunscreen, watch water temperatures)
- Maintain a regular bowel program.
- Well balanced diet and adequate fluid intake
- Compliance with medications
- Persons at risk and those close to them should be educated in the causes, signs and symptoms, first aid, and prevention of autonomic dysreflexia.
- If you have an indwelling catheter:

  Keep the tubing free of kinks.
- Keep the drainage bags empty
- Check daily for grits (deposits) inside of the catheter.
- If you are on an intermittent catheterization program, catheterize yourself as often as necessary to prevent overfilling.
- If you have spontaneous voiding, make sure you have an adequate output.
- Carry an intermittent catheter kit when you are away from home.
- Perform routine skin assessments

Treatment of Autonomic Dysreflexia:

1. Recognise the symptom and remove the precipitating stimuli. Check the BP.
2. Check urinary blockage. Look for the kinks.
3. Flush the catheter.
4. If on intermittent catheter, do catheterization.
5. Consider medical management. If systolic BP remains above 150mm.
6. Check if there is a fecal impaction in rectum.

N.B.: Teaching the patient and the caretaker about this complication is essential.

2. DEEP VEIN THROMBOSIS

Deep Vein Thrombosis (DVT) is a common complication of acute spinal cord injury (SCI) but also major cause of morbidity and mortality. Morbidities from DVT include thromboembolism, prolonged edema, and pressure ulcers.

Definition

Deep vein thrombosis (DVT) is a condition in which a blood clot (thrombus) forms in one or more of the deep veins in the body, usually in the legs. Deep vein thrombosis can cause leg pain, but often without any symptoms.

There are changes in the normal neurologic control of the blood vessels that can result in occlusion of

the normal flow. DVT in the lower leg is almost universal during the early phases of recovery and rehabilitation. Thromboses in the thigh, however, are a great concern, as they are at risk for becoming dislodged and passing through the vessels to the lungs. A major obstruction of the arteries leading to the lung can potentially be fatal.

INCIDENCE

The incidence of DVT following acute spinal cord injury has been reported more than 20%. How ever the clinical symptoms are seen in only 15% of patients with acute spinal cord injury, and pulmonary edema develops in approximately 5% of these patients. The risk of DVT is highest within the first 2 weeks following injury, with peak occurrence between 7 and 10 days. DVT has been detected as early as 72 hours post injury; however, risk before this time appears to be low.

CAUSES

1. Inactivity after a spinal cord injury
2. Absence of pumping action in the leg musculature due to paralysis.
3. Trauma

SYMPTOMS

– Swelling in the affected calf muscle in the affected leg, including swelling in ankle and foot.

– Pain in the leg mainly in ankle and foot. The pain often starts in the calf and can feel like cramping. However majority of paraplegics do not have normal sensation and therefore one has to look for other symptoms to diagnose and treat the case immediately.

– Warmth over the affected area.

– Changes in the skin colour, such as turning pale, red or blue.

TREATMENT

The goal of deep vein thrombosis treatment

- Preventing blood clot from getting any bigger.
- Preventing the clot from dislodging and causing a pulmonary embolism
- Reducing the chances of recurrence of DVT.

Deep vein thrombosis treatment options include:

- Blood thinners. Medications used to treat deep vein thrombosis include the use of anticoagulants, whenever possible. While they don't break up existing blood clots, they can prevent clots from getting bigger or reduce the risk of developing additional clots.

  As soon as the DVT is suspected in SCI infusion of the blood thinner like heparin is given for a few days. After the course of heparin injections, oral medicines like warfarin are usually given. Oral blood thinners are required for at least for a minimum period of 3 months. However, these blood thinners have serious side effects like risk of bleeding especially when the dose is too high, how ever if the dose is too low there can be chances of getting additional blood clots. Therefore it is necessary to get blood clotting time bleeding time done periodically and the physician has to adjust the optimum dosage. In the event of a Paraplegic getting a life threatening complication like Pulmonary embolism, the physician may give other medications like tissue plasminogen(TPA)
- In cases where one cannot be given blood thinners, Filters are inserted in the venacava which prevents clots that break loose from entering the lung.
- Compression stockings. These help preventing swelling that is associated with DVT. These stockings are worn on the leg from foot to knee. They are to be worn for a period of at least one year.

3. HETEROTOPIC OSSIFICATION

INTRODUCTION:

Heterotopic ossification following spinal cord injury (SCI) was first described by Dejerine and Ceillier in 1918 as para osteoarthropathy. The ossification process involves the formation of mature lamellar bone, which is indistinguishable from normal bone, in soft tissues surrounding paralyzed joints. The bone is not connected to periosteum and becomes encapsulated as it matures.

The pathology is similar to that of fracture callus, except that bone forms in the connective tissue between the muscle planes (histologic findings in neurogenic heterotopic ossification are similar to those in healing fracture callus). Heterotopic ossification is also seen after other neurologic insults, such as traumatic brain injury (TBI) and

stroke, as well as after thermal injuries and orthopedic procedures (eg, total hip replacement).

In experimental models of heterotopic ossification formation, ischemia and tissue expression of bone morphogenic proteins have been shown to play important roles. Bone morphogenic proteins likely act on mesenchymal stem cells present in tissue, activating the cells to differentiate into osteoblasts.

INCIDENCE:

The incidence of heterotopic ossification in spinal cord injury is between 16% and 53%, depending on the incidence reports from various institutions. Once present, neurogenic heterotopic ossification is clinically significant in 18-27% of cases. Fortunately, only 3-5% of cases involve joint ankylosis.

There is no known race or sex predilection for neurogenic heterotopic ossification; however, the incidence of neurogenic heterotopic ossification after spinal cord injury is lower in pediatric patients than in adults, ranging from 3% to 10%. In addition, spontaneous resorption of the neurogenic heterotopic ossification is frequently seen in pediatric patients.

CLASSIFICATION OF HETEROTOPIC OSSIFICATION

HETEROTOPIC OSSIFICATION can be classified into the following 3 types:

1. **MYOSITIS OSSIFICANS PROGRESSIVA (FIBRODYSPLASIA OSSIFICANS PROGRESSIVA)** - this disorder is among the rarest genetic conditions, with an incidence of 1 case per 2 million persons. transmission is autosomal dominant with variable expression. the condition is characterized by (a) recurrent, painful soft-tissue swelling that leads to heterotopic ossification and (b) congenital malformation of the great toe. there is no treatment for this form of heterotopic ossification. limited benefits have been reported using corticosteroids and etidronate. most patients die early from restricted lung disease and pneumonia; however some patients live productive lives.
2. **TRAUMATIC MYOSITIS OSSIFICANS** - in this condition, a painful area develops in muscle or soft tissue following a single blow to the area, a muscle tear, or repeated minor trauma. The painful area gradually develops masses with a cartilaginous consistency; within 4-7 weeks, a solid mass of bone can be felt. Common sites include the pectoralis major, the biceps, and thigh muscles. a nontraumatic type of myositis ossificans also may exist.
3. **NEUROGENIC HETEROTOPIC OSSIFICATION** - this condition is the one that comes to mind when the generic phrase heterotopic ossification is used. This type of heterotopic ossification is the subject of this.. the various terms mentioned at the outset all refer to this type of heterotopic ossification.

PATHOPHYSIOLOGY OF HETEROTOPIC OSSIFICATION

The mechanism underlying heterotopic ossification following spinal cord injury is not fully understood but it appears to be initiated by mesenchymal cells into bone precursor cells. It has been noted that mesenchymal stem cells can differentiate into osteogenic cells given the right stimuli within the right environment, even soft tissues. These mesenchymal stem cells can generate cartilage, bone, muscles, tendons, ligaments or fat and are thought to play a pivotal role in the development of Heterotopic Ossification. Heterotopic Ossification then forms through a typical process beginning with the formation of a protein mixture created by bone cells (osteoid) that eventually calcifies within a matter of weeks. Over the next few months, the calcified osteoid remodels and matures into well-organized trabecular bone. Months following the initial trauma patients develop bone formation in muscle and soft tissues adjacent to a joint (paraarticular) with resultant restriction in range of motion, pain and ankylosis. The bony lesion has a high metabolic rate, adding new bone at more than three times the rate of normal bone. Osteoclastic (bone removal cell) density is more than twice that found in healthy bone. It is suspected there may be a neurogenic factor contributing to Heterotopic Ossification but the mechanism is poorly understood.

CAUSES:

No one is quite sure what causes heterotopic bone formation. Many reports propose it is related to a type of mesenchymal metaplasia, meaning that connective tissue cells change their characteristics into bone forming cells. It is not known why the cells change function but it is thought to be some type of inflammatory reaction.

In individuals with spinal cord injury, heterotopic ossification will usually begin forming in the intramuscular connective tissue within four months of the injury. When the reaction begins, there will be a deposition of calcium phosphate in the intramuscular tissue. However, ossification occurs only when the calcified material goes on to form hydroxyapatite crystals. Once bone formation has occurred, it rarely disappears spontaneously

HISTORY

The onset of heterotopic ossification usually is 1-4 months after injury in SCI patients, although it may occur in earlier weeks or as late as 1 year following injury. The condition may occur later with other precipitating circumstances (eg, fracture, surgery, severe systemic illness).

Commonly, incidental heterotopic ossification that was not noted clinically may be detected much later on radiographs.

HO always occurs below the level of injury in SCI patients.

HO tends to occur more frequently with complete injuries.

In SCI patients with HO, the hips are most commonly involved.

- At the hip, the flexors and abductors tend to be involved more frequently than are the extensors or adductors.
- At the knee, the medial aspect is most commonly affected by heterotopic ossification.
- Shoulders and elbows are the most commonly affected upper extremity joints.
- Involvement of the metacarpophalangeal joints of the hand is rarely seen.
- The lumbar paravertebral region also has been noted though very rarely.

PHYSICAL EXAMINATION:

A diagnosis of heterotopic ossification can be made clinically if localized inflammatory reaction, palpable mass, or limited Range of motion is observed.

Clinically, the onset of larger masses of heterotopic ossification is often characteristic of any inflammatory reaction.

Fairly, a warm and swollen extremity becomes obvious, and fever is present.

If sensation is intact, the area of swelling is painful. The swelling usually is localized more than it is in thrombophlebitis, and within several days, a more circumscribed, firmer mass is palpable within the edematous area.

If the mass is adjacent to a joint, gradual loss of passive range of motion may follow.

With the development of early heterotopic ossification at the hip or knee, effusion may be noted at the knee.

MANAGEMENT

MEDICAL MANAGEMENT

In the later stages of the development of mature bone, medical treatment is ineffective. Etidronate (Didronel) is the only available medication for the treatment of HO after SCI. Treatment with NSAIDs may be required initially, until the resolution of inflammation and the normalization of CRP levels.

PHYSICAL REHABILITATION

The use of physical therapy (PT) in HO has long been controversial. Rossier and co-investigators noted occasional transverse microfractures on sections of HO that they thought might be caused by spasticity or by overly aggressive Passive range of motion (PROM). Since then, the debate between resting the joint and aggressive PROM has continued. In the literature, however, the developing consensus appears to be that aggressive PROM and continued mobilization, once acute inflammatory signs have subsided, are indicated, because they help to maintain ROM and (in more extensive HO) they may lead to the formation of a pseudarthrosis. Resting the joint appears more likely to lead to decreased ROM or to ankylosis.

During the acute inflammatory stage, the patient should rest the involved joint in a functional position, and the physical therapist should initiate gentle PROM as soon as possible. The role of continuous PROM machines has not been studied in this situation. For patients with incomplete SCI or head injuries, maintaining ROM may be difficult because of pain from ROM exercises. The use of joint manipulation has been reported in patients with HO. but because of limited joint ROM, they have functional limitations. However, such manipulation is controversial owing to the risk of the formation of new hematoma and because of the chance that long-bone fracture will occur in patients with secondary osteoporosis.

NEUROGENIC PAIN

INTRODUCTION

Longstanding pain is experienced by many Paraplegics below the site of lesion. This is one of the most challenging medical problems after spinal cord injury (SCI).This neuropathic pain is very difficult to be treated and patients spend many sleepless nights. The incidence is very high. A review by Bonica an average of two-thirds of the Paraplegics experience this disturbing pain out of which half describe the pain as very severe, interfering with ADL.

CLASSIFICATION OF PAIN IN SPINAL CORD INJURY

Individuals with SCI can experience several types of pain. The most common ones can be classified into three groups.

- Neuropathic pain
- Musculo skeletal pain
- visceral

Neuropathic Pain

There are three varieties viz. SCI central pain, Segmental, nerve root entrapment pain, pain due to syrinx

SCI (Central pain) is described as sharp, shooting, burning pain, tingling numbness or throbbing. Though the patients do not experience sensations like touch, pin prick or any other sensation, he experiences a disturbing type of pain. This can begin within weeks or months after the injury and is felt at the level of injury or below the level where there is no touch sensation. It is thought that the pain signals are coming from elsewhere and not related to any action or position of the limbs.

Segmental pain often occurs around the border where there is normal sensation and where there is loss of sensation as a result of injury. It can be slightly above the level of injury or slightly below. It usually develops during the first few months after injury. Segmental pain is often associated with allodynia and hyperalgesia in the painful region. Allodynia is pain caused by something that does not normally cause pain. For example, something cold, warm or a very light touch to the skin can result in pain. Hyperalgesia means an extremely painful response to what is normally only mildly painful.

Nerve root entrapement pain often begins days to weeks after injury and may worsen over time. It occurs at or just below the level of injury and has a distinct pattern. You may feel brief waves of stabbing or sharp pain or a band of burning pain at the point where the normal feeling stops. Some times a light touch could trigger a severe pain.. The pain stems from compression of a nerve root by a bone or disk. Pain from damage to the cauda equina is a type of nerve root pain that is described as a burning feeling in the legs, feet, pelvis, genitals, and rectum.

Syringomyelia is a hollow, fluid filled cavity (syrinx) in the spinal cord. It is not common, but sometimes develops months or years after injury. The cavity can slowly increase in size and extend up or down the spinal cord. As the syrinx expands, it can result in pain along with an increased loss of sensory and motor function.

Musculoskeletal

This type of pain is also a concern for individuals with spinal cord injury. It occurs in parts of the body like the bones, joints, and muscles. Musculoskeletal pain is usually worsened by movement and eased with rest. It can generally be described as a dull or aching pain, but the pain can also be described in other terms.

Secondary overuse (pressure syndromes) is a very common cause of musculoskeletal pain. The pain can occur months or many years after injury. It is caused by the overuse of muscles in any part of the body. For example, many people develop tendonitis of the rotator cuff (shoulder) as a result of pushing a manual wheelchair for a long period of time.

Muscle spasm pain is experienced by some individuals after SCI. The spasms are involuntary movements of the body in areas that have lost some or all motor function. The pain is caused when muscles and joints are strained.

Mechanical instability of the spine is caused by damaged ligaments or fracture of bones. It occurs most often shortly after injury, but it can also develop later. The pain is usually around the area of instability.

Visceral

Visceral pain usually begins a short time following SCI. It occurs in the abdomen either above or below the level of injury. The pain is described as burning, cramping and constant.

PATHOPHYSIOLOGY

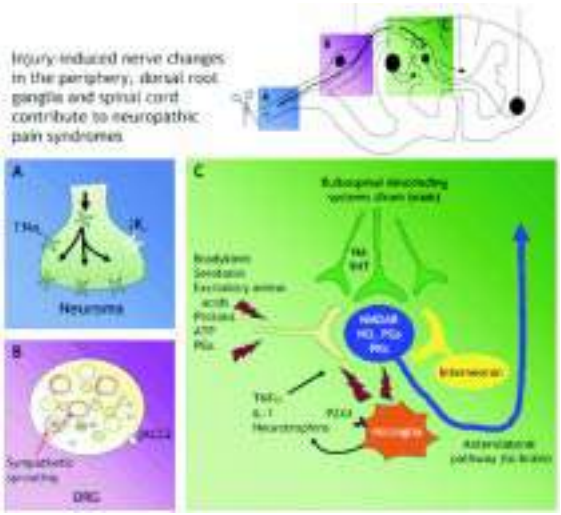


Fig: Neuropathic pain arises following nerve injury or dysfunction. A: After nerve damage, transcription and axonal trafficking of sodium channels to the site of injury is increased, with concomitant attenuation of potassium channels. The altered expression of ion channels results in neurons becoming hyperexcitable and generating ectopic activity, which is thought to lead to the genesis of spontaneous and paroxysmal pain. B: At the cell body of primary afferent neurons within the dorsal root ganglia (DRG), sympathetic neuronal sprouting occurs and may account for sympathetically maintained pain. C: Peripheral nerve injury causes a multitude of changes in gene transcription and activation of various kinases and proteins, including enhanced N-methyl-D-aspartate (NMDA) receptor activity. However, nerve injury also elicits hypertrophy and activation of glial cells, including microglia within the grey matter of the spinal cord. Microglia expresses P2X4 purinergic receptors, allowing them to be activated by adenosine triphosphate (ATP). Following activation, microglia release various pronociceptive cytokines, such as interleukin-1 (IL-1), tumour necrosis factor alpha (TNF-a) and neurotrophins, including brain-derived neurotrophic factor, which in turn exacerbates nociceptive transmission and contributes

to the sensitization and maintenance of neuropathic pain.

Note: Aß = A beta neuron, Ad = A delta neuron, C = C nociceptor, 5HT = serotonin, KCC2 = chloride transporter, NA = noradrenaline, Nav = sodium channel, NO = nitric oxide, Kv = potassium channel, PGs = prostaglandins, PKs = protein kinases, P2X4 = purinergic receptor.

CLINICAL PRESENTATION AND PATIENT EVALUATION

The blockade of nerve conduction in neuropathic conditions causes nerve dysfunction, which can result in numbness, weakness and loss of deep tendon reflexes in the affected nerve area. Neuropathic conditions also cause aberrant symptoms of spontaneous and stimulus-evoked pain. Spontaneous pain (continuous or intermittent) is commonly described as burning, shooting or shock-like. Stimulus-evoked pain includes allodynia (pain evoked by a nonpainful touch) and hyperalgesia (increased pain evoked by a painful stimulus). Allodynia can be caused by the lightest stimulation, such as skin contact with clothing or a light breeze. These sensory abnormalities may extend beyond nerve distributions, which may lead to the inappropriate diagnosis of a functional or psychosomatic disorder. The diagnosis of neuropathic pain is based primarily on history and findings on physical examination.

TREATMENT

Neuropathic pain can be very difficult to treat with only some 40-60% of patients achieving partial relief. Determining the best treatment for individual patients remains challenging. Establishing an accurate diagnosis is an important first step.

NONPHARMACOLOGICAL TREATMENT

Although many patients with neuropathic pain pursue complementary and alternative treatments, rigorous evidence supporting efficacy of nondrug therapy is limited. Some reports suggest benefits of conservative interventions such as exercise, transcutaneous electrical nerve stimulation, percutaneous electrical nerve stimulation, graded motor imagery and cognitive behavioural therapy or supportive psychotherapy. Relieving stress related activities is important because these intensify most pain disorders and may be helped by psychological counselling or psychiatric intervention.

PHARMACOLOGICAL TREATMENT

It is common practice in medicine to designate classes of medication according to their most common or familiar use e.g. as "antidepressants" and "anti-epileptic drugs" (AED's). These drugs have alternate uses to treat pain because the human nervous system employs common mechanisms for different functions, for example ion channels for impulse generation and neurotransmitters for cell-to-cell signaling.

No one pharmacological protocol can be used in the treatment of all patients. Usually the approach is to try one agent until pain relief is achieved, a maximum dose is obtained, or side effects prohibit increasing the dose, before trying another agent. Favored treatments are certain antidepressants e.g. tricyclics and selective serotonin-norepinephrine reuptake inhibitors (SNRI's), anticonvulsants, especially pregabalin (Lyrica) and gabapentin (Neurontin), and topical lidocaine. Opioid analgesics and tramadol are recognized as useful agents but are not recommended as first line treatments. Many of the pharmacologic treatments for chronic neuropathic pain decrease the sensitivity of nociceptive receptors, or desensitize C fibers such that they transmit fewer signals.

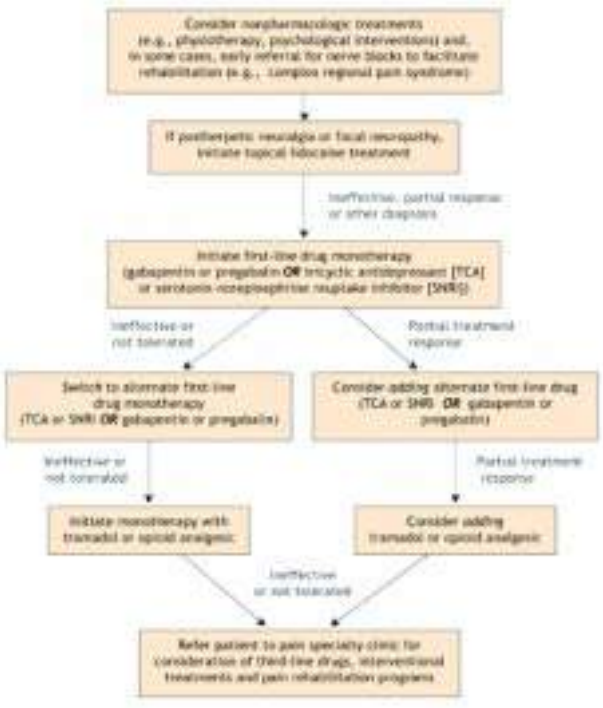
Some drugs may exert their influence through descending pain modulating pathways. These descending pain modulating pathways originate in the brainstem.

The use of intrathecal baclofen, morphine, and clonidine are newer approaches to decrease this type of pain.

SURGICAL TREATMENT

Surgical intervention is only considered if all else has failed and the neuropathic pain interferes with daily activities. The most common surgical techniques used include dorsal root rhizotomy, cordotomy, and lumbar sympathectomy.

ALGORITHM FOR THE MANAGEMENT OF NEUROPATHIC PAIN IN PRIMARY CARE.



Neuropathic pain is best managed with a multidisciplinary approach. Nevertheless, several different treatments can be initiated in the primary care setting. Treatments with the lowest risk of adverse effects should be tried first. Evidence supporting conservative nonpharmacologic treatments (e.g., physiotherapy, exercise, transcutaneous electrical nerve stimulation) is limited; however, given their presumed safety, nonpharmacologic treatments should be considered whenever appropriate.61 Simple analgesics (e.g., acetaminophen, NSAIDs) are usually ineffective in pure neuropathic pain but may help with a coexisting nociceptive condition (e.g., sciatica with musculoskeletal low-back pain). Early referrals to a pain clinic for nerve blocks may be warranted in some cases to facilitate physiotherapy and pain rehabilitation.

CONCLUSION

Neuropathic pain is a devastating chronic condition that generally can be diagnosed by history and findings on physical examination. For some neuropathic pain syndromes, available treatments are tolerable and afford meaningful relief to a considerable proportion of patients. Nevertheless, many patients report intractable and severe pain, and better treatment strategies are desperately needed. The field of neuropathic pain research and treatment is in the early stages of development, with many goals yet to be achieved. In particular, future laboratory, clinical and epidemiologic research into pathogenesis, treatment and prevention of neuropathic pain is expected as well as improved dissemination of new information to health professionals and the public. Over the years to come, many upcoming advances are expected in the basic and clinical science of neuropathic pain as well as in the implementation of improved therapies for patients who continue to experience these devastating conditions.

5. OSTEOPOROSIS

Introduction

One of the inevitable complications of spinal cord injury (SCI) is the associated osteoporosis that occurs mainly in the pelvis and the lower extremities. The acute treatment of patients with spinal cord injury has always been concentrated on the injury itself and on the subsequent complications. Osteoporosis or bone loss as a consequence of spinal cord injury has been of secondary concern.

Osteoporosis in persons with spinal cord injury was first studied in relation to calcium metabolism and the associated hypercalcemia and renal calculi that followed. The differences between osteoporosis induced by spinal cord injury and other causes of bone loss due to disuse because of prolonged bed rest, space travel, and lower motor neuron disorders, have since become clearer.

- Bone loss occurs below the level of the spinal cord injury, with no loss of any bone mass above the lesion.
- Trabecular bone is more affected than cortical bone, and in particular femur and tibia. Studies have shown that there is about 30% to 40% decease in bone density in the lower extremities after SCI.
- Osteoporosis can be detected on x-rays as early as six weeks after injury. Most researchers feel that bone loss is not that rapid after 6weeks and it levels out by around 2 years. But some studies show that there is still possibility of bone loss even after2 years..
- The lumbar spine does not show any osteoporosis, the reason being that SCI persons sit on the W / C for longer period and the loading is maximum on sitting than any other position. Loading of bone actually stimulates bone mineral density. The non-weight bearing lower extremities don't have this stimulation and therefore lose bone mineral density.
- Injury level

- Individuals with quadriplegia have more osteoporosis than paraplegics because of more area below the lesion. Paraplegics usually have bone mineral density preserved in their upper extremities.
- In the bone that is affected, the severity of bone loss is the same both in paraplegia and tetraplegia.
- Severity of injury: Individuals with complete injuries have more bone loss than those with incomplete injuries.
  - Spasticity may have a beneficial effect in maintaining bone mass after SCI, due to muscle pulling on the bone, similar to the effect of weight-bearing.
  - Duration of injury: The longer time the since injury, the greater the bone loss is.

Fractures and SCI

There is a risk of fractures as the bone mineral density decreases. The incidence of fractures of the lower limbs in SCI is quite high.. Most fractures while doing normal activities such as transferring and not from violent falls. Sometimes people cannot remember of any trauma at all, but when they notice swelling or redness get investigated and find they have afracture.

CAUSES OF OSTEOPOROSIS IN SCI

Various possibilities for osteoporosis are

- Disuse: or lack of mechanical loading on the bone would inhibit stimulation of bone-building cells.
- Disordered vasoregulation: sluggish blood flow to limbs may contribute to a decrease in bone mass.
- Poor nutrition: not taking healthy and balanced diet
- Hormonal alterations : proteins in the body are responsible for the maintenance bony formation and resorption.
- Metabolic disturbances (tissue acidosis, alkaline phosphatase, hypercalcemia / hyercalciuria, hydroxyproline excretion): disturbance in metabolites and acidity of the blood can influence the balance of bony formation and resorption.
- Autonomic disregulation: impaired control by the self-regulating nervous system can lead to increased imbalance between bone formation and resorption.

PATHOPHYSIOLOGY

The mechanism behind spinal cord injury (SCI)–induced osteoporosis is accepted as being multifactorial in the acute and chronic stages. These mechanisms differ from those observed in subjects without spinal cord injury after prolonged bed rest and in subjects with other neurologic deficits.

Disuse structural change and hypercalciuria

Spinal cord injury causes immediate and, in some regions, permanent gravitational unloading. The result is a disuse structural change with associated metabolic consequences. Hypercalciuria is seen by 10 days following the spinal cord injury and reaches a peak 1-6 months postinjury. This level of hypercalciuria is 2-4 times that of persons without spinal cord injury who undergo prolonged bed rest.

This marked increase in urine calcium is the direct result of an imbalance between bone formation and bone resorption.

Osteoblast and osteoclast activity

The activity of osteoblasts and osteoclasts is triggered by the spinal cord injury; however, markers of osteoblastic activity rise only slightly, whereas osteoclasts have a significant increase in their activity, peaking at 10 weeks following the injury with values 10 times the upper limits of normal (ULN). In addition, the increased bone resorption precedes the increase in osteoblastic activity. This model at the skeletal level following spinal cord injury resembles the high bone turnover rate seen in postmenopausal osteoporosis.

Bone muscle traction loss or neuronal factors

The loss of bone also may be enhanced by lack of muscle traction on bone or by other neural factors associated with spinal cord injury. These other factors further separate spinal cord injury–induced osteoporosis from other causes of disuse demineralization. Absorption of calcium from the gastrointestinal (GI) tract has been found to decrease in the acute period following injury. Even so, in the past, dietary calcium reduction was commonly recommended as a way to decrease calcium excretion and prevent the complications of hypercalciuria.

Parathyroid hormone

The body that has sustained spinal cord injury has been considered the model of premature aging, and the role of parathyroid hormone (PTH) in osteoporosis following spinal cord injury illustrates this point. Acutely, the parathyroid gland is relatively inactive, with low PTH levels observed up to the 1-year point following injury. Hypercalcemia seen immediately postinjury leads to this low level. A reversal in activity during years 1-9 is noted.

The parathyroid gland is stimulated to the point that PTH levels are above the reference range. The result is an increase in bone reabsorption or osteoporosis related to parathyroid dy nosfunction in the chronic stages of spinal cord injury. This chronic-stage mechanism of osteoporosis is balanced by an increase in bone mineral in regions of the body in which weight bearing is resumed (eg, in the upper extremities, spine) and adds to the demineralization observed in regions that are chronically non–weight-bearing (eg, the pelvis, lower extremities). In addition, the prevalence of vitamin D deficiency in SCI is increased, and this may exacerbate bone loss

SIGNS AND SYMPTOMS

There are no particular signs and symptoms for osteoporosis Only when an x ray is taken following a fracture it is revealed. There is no other physical examination to diagnose the condition. At times there may be some effusion in the knees or heterotropic ossification

Biomechanical Markers

The biomechanical markers that have been measured in studies of spinal cord injury (SCI)–induced osteoporosis include serum calcium, phosphorous, alkaline phosphatase, 1,25–dihydroxyvitamin D and calcitonin, and urinary calcium and hydroxyproline.

These markers may not be followed routinely in the ongoing care of the person with spinal cord injury. However, the sensitivity and early response of these markers indicate that they would be useful in the early identification of patients with spinal cord injury who are at risk of developing severe osteoporosis.

MANAGEMENT APPROACHES

Changes do occur very rapidly in the bones of a patient with spinal cord injury (SCI), and interventions must be undertaken quickly. Infact that there are no effective treatments to restore bone mineral once it has been lost and therefore early treatment is imperative. Thus, prevention is the main focus in treating spinal cord injury–induced osteoporosis.

In the evet of a fracture conservative treatment is to be tried first. Invariably healing would be seen in 3 to 4 weeks. Soft splints may be required. Hard splints and materials should be avoided. However if deformities are seen surgical intervention may be necessary.orthopedic surgeon's opinion has to be taken.

PREVENTION & TREATMENT

It is difficult to totally prevent bone demineralization after spinal cord injury. Individuals are at higher risk for osteoporosis following SCI. There is no "standard of care" to treat this problem. Each individual needs to be evaluated to determine if treatment is essential.

- **Exercise**

Physical activity is supposed to preserve or increase bone mass in able-bodied individuals. Studies has shown that very hard exercises are beneficial preventing bone loss in the upper extremities.. But this type of exercise did not stop demineralization of the lower body. Individuals with SCI need to be cautious when they begin an exercise program as overuse syndrome may set in. Therefore a physiotherapist has to prescribe the level o f exercises. Various activities are under study as forms of exercise to build bone strength in individuals with SCI. These include weight bearing using a standing frame or harness; treadmill training; Parastep; and functional electrical stimulation. Method.How ever there is controversy about the effectiveness of thisT of these different programs. Some of drawbacks to these activities are costs, the need to be done for long periods of time, and difficulty fitting it in to an individual's daily schedule. There is also an increased incidence of fractures if bones are already badly osteoporotic.

- **Extra calcium and vitamin D**

These need to be included in one's daily diet to help in preventing osteoporosis. Calcium helps build strong bones and vitamin D improves the absorption of calcium. But for individuals with SCI, high levels of calcium and Vitamin D may increase the risk of urinary calculii. There are no guidelines on the risks or benefits of calcium and Vitamin D supplements for individuals with SCI. A supplement of 1000 mg / day will be safe and beneficial if the excretion remains less than 250 mg / day and the individuals' parathyroid hormone levels are in mid-normal range. It may be necessary to monitor calcium levels in the urine if supplements are used.

- **Stop smoking.**

  Cigarette smoking reduces the body's ability to absorb calcium. This in turn can speed up bone loss in all populations.

- **Limit caffeine.**

  Caffeinated drinks act as a diuretic. They speed up the removal of calcium from the body in the urine by about 10 mg per day.

- **Avoid drinking too much alcohol.**

  This is linked to bone loss as well as poor nutritional habits.

- **Medications**

  A new class of drugs, bisphosphonates, can help prevent bone loss and increase bone density by 1 to 4%. These drugs are approved for preventing and managing osteoporosis in the general population.

- **Protect the Bones.**

Individuals with SCI need to protect their bones. When doing range of motion exercises, limit movement to stretches that are easily done. Do not force a joint or muscle to move past what is comfortable. Check with a physical therapist for exercises specific to an individual's needs and abilities.

Spasticity can put some force on the bones that helps to strengthen them. However strong spasms could cause a weak bone to fracture. Be cautious when transferring. Remove feet from heel loops or toe straps on the foot rests before transferring. If a person's balance or strength is weak, he / she needs to ask for help to avoid falling.

SPASTICITY

INTRODUCTION:

Spasticity is the involuntary movement (jerking) of muscles, which occurs because messages can travel from parts of your body to the spinal cord and cause reflex activity (muscle movement). This is possible because the spinal cord has certain normal automatic functions, which are under the influence of the brain. These functions include muscle tone and reflexes. Most spinal cord injured persons have a healthy, intact spinal cord below the immediate area of their injury, and thus these automatic activities can continue to exist. However, they are no longer under the regulating influence of the brain and are thus exaggerated. This is called spasticity.

Spasticity is common in varying degrees after spinal cord injury. Spasticity is a non-specific symptom, which may occur in many problems associated with spinal cord injury.

ETIOLOGY:

After spinal cord injury the nerve cells below the level of injury become disconnected from the brain. Following the period of spinal shock changes occur in the nerve cells that control muscle activity. Spasticity is an exaggeration of the normal reflexes that occur when the body is stimulated in certain ways. After spinal cord injury, when nerves below the injury become disconnected from those above, these responses become exaggerated.

Muscle spasms, or spasticity, can occur any time the body is stimulated below the injury. This is particularly noticeable when muscles are stretched or when there is something irritating the body below the injury. Pain, stretch, or other sensations from the body are transmitted to the spinal cord. Because of the disconnection, these sensations will cause the muscles to contract or spasm.

Some stimuli can cause a change in your spasticity. Anything that would ordinarily be uncomfortable or painful can cause an increase in your spasticity. If you experience a major increase in spasticity, possible causes are:

- Skin problems - a skin sore or ingrown toenail
- Bladder problems - high residuals, infection or bladder stones
- Bowel problems - constipation, impactions or hemorrhoids
- Medical problems - viral syndrome (infection, influenza, intestinal flu), heterotopic ossification or a spinal cyst.

TRIGGERING FACTORS:

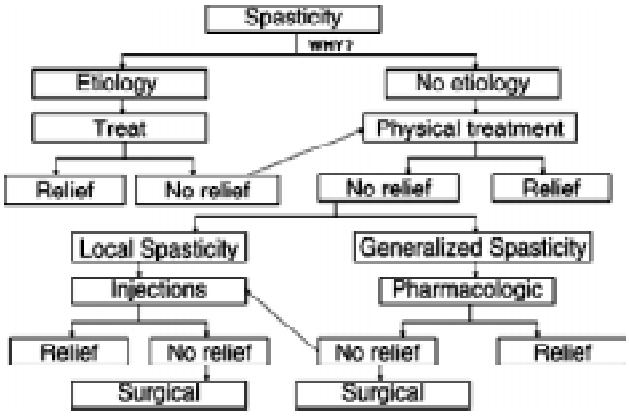
Almost anything can trigger spasticity. Some things, however, can make spasticity more of a problem. A bladder infection or kidney infection will often cause spasticity to increase a great deal. A skin breakdown will also increase spasms. In a person who does not perform regular range of motion exercises, muscles and joints become less flexible and almost any minor stimulation can cause severe spasticity.

BENEFITS OF SPASTICITY:

There are some benefits to spasticity. It can serve as a warning mechanism to identify pain or problems in areas where there is no sensation. Many people know when a urinary tract infection is coming on by the increase in muscle spasms. Spasticity also helps to maintain muscle size and bone strength. It does not replace walking, but it does help to some degree in preventing osteoporosis. Spasticity helps maintain circulation in the legs and can be used to improve certain functional activities such as performing transfers or walking with braces. For these reasons, treatment is usually started only when spasticity interferes with sleep or limits an individual's functional capacity

MANAGEMENT:

Some spasticity may always be present. The best way to manage or reduce excessive spasms is to perform a daily range of motion exercise program. Avoiding situations such as bladder infections, skin breakdowns, or injuries to the feet and legs will also reduce spasticity. There are three primary medications used to treat spasticity, baclofen, Valium, and Dantrium. All have some side effects and do not completely eliminate spasticity. Other interventions to help reduce or control spasms include injections of Botulinum Toxin A (Botox) or phenol directly into the muscles of concern. Injection therapy usually lasts 3-4 months. Surgical interventions include the insertion of an Intrathecal Baclofen Pump. A Baclofen pump trial is conducted first to make sure there is an adequate response to the intrathecal baclofen. Once it is determined that a person responds to the intrathecal baclofen therapy, surgery is scheduled. The pump must be refilled every three months but is dependent upon the amount of drug needed by each person.In the case of SCI, the distribution of spasticity tends to be more diffuse, making regional or systemic treatment preferable.34 The decision whether or not to treat spasticity and, if so, in what manner, is summarized nicely in a flow chart



CONSERVATIVE/PHYSICAL REHABILITATION MANAGEMENT

It is generally agreed that physical therapy / rehabilitation is an essential component in the management of spasticity as a first line of defence, as well as in a long-term regimen during and after the implementation of pharmacological or surgical strategies. The goal of physical therapy is to diminish spasticity in order to allow expression of voluntary mobility and movements and / or to improve the comfort and independence in tasks related to Quality of living, such as transfers, dressing, and using the washroom. The literature on the conservative / physical treatment of spasticity is sparse, and some have questioned the effectiveness of these management strategies. Table below summarizes the most common physical therapy approaches to spasticity management.

Table - Physical techniques in the management of spasticity.

Physical therapy technique description and comment on effectiveness	Purpose / suggested mechanisms
Positioning	
• In bed and during sitting • Reports of clinical effectiveness; impact remains to be proven scientifically	• Important to the maintenance of muscle length
Range of motion/stretching	
• Includes passive stretch and passive length-ening • Benefits may carry over for several hours • Effects remain to be quantified and the efficacy remains to be determined despite the clinical evidence for the benefits	• Prevents contractures • Causes temporary reduction in intensity of muscle contraction in reaction to muscle stretch • May cause plastic changes within the central nervous system and / or mechanical changes at the muscle, tendon, and soft-tissue level
Weight-bearing	
• Using a tilt table or standing frame •Benefits are greater than stretching alone and may persist into next day • Effectiveness has been questioned	• Prolonged stretch of ankle plantar flexor muscles • Mechanism remains uncertain; suggested to include a modulating influence from cutaneous and joint receptor input to the spinal motor neurons, resulting in decreased excitability
Muscle strengthening	
• Progressive addition of resistance to muscles with voluntary control	• Emphasis of balance of agonist and antagonist groups of muscles with voluntary control
Electrical stimulation	
• Various methods: stimulation to the antagonist muscle, application of tetanic contraction to the spastic muscle, functional electrical stimulation (FES), and transcutaneous electrical nerve stimulation(TENS) Reports of beneficial effects between only 10 min and 3 h	• Stimulation of the antagonist muscle: augmentation of reciprocal inhibition of the spastic muscle • Repetitive tetanic stimulation of spastic muscle: fatigue of the muscle due to repetitive tetanic stimulation • FES: change the mechanical properties of a spastic joint by strengthening the antagonists of the spastic muscle or might decrease the hyperactivity of spastic muscles through reciprocal inhibition • TENS: may involve the stimulation of large diameter afferent fibers that travel from mechanoreceptors to the spinal cord14
Epidural spinal cord stimulation	
•For mild spasticity and incomplete lesions: stimulation below the level of the lesion found effective (spasms) For severe spasticity: stimulation of dorsal roots of the upper lumbar cord segment found effective (hypertonus and spasms) Shown to lack long-term effectiveness	• May involve the activation of inhibitory networks within the spinal cord • More strongly affected patients require stronger stimuli and / or higher frequencies

Cold/heat application	
• Application of a cold pack or a vapocoolant spray, or superficial heat • Following cold application: tendon reflex excitability and clonus may be reduced for a short period of time (eg, <1 hr), allowing for intermittent improved motor function • Following heat application: subsequent passive stretch is facilitated	• Cold: may cause slowing of nerve conduction, decrease in sensitivity of cutaneous receptors, and alteration of CNS excitability • Heat: facilitation of uptake of released neurotransmitters and return of calcium to the
Splinting/orthoses	
• Helpful in the continuous application of muscle stretch • Use of splints is questioned	•Enables long-term stretch Joint can be maintained in a position that does not elicit a spasm

PRESSURE SORES

INTRODUCTION

Pressure ulcers are an extremely common complication after spinal cord injury. In the acute stage after spinal cord injury, more than one third of patients have pressure ulcers. Pressure ulcers may delay the onset of rehabilitation therapies, lengthen the total hospitalization, and make the ultimate adjustment to disability more difficult. In the long term, these ulcers impair an individual's quality of life because of activity restriction and increased need for medical and nursing care, often by hospitalization or surgery.

A pressure sore is known by many names, like pressure ulcer, decubitus ulcer, ischemic ulcer, bed sore or skin sore. No matter what it is called, it is a serious problem that can take days, weeks, months or even longer to heal.

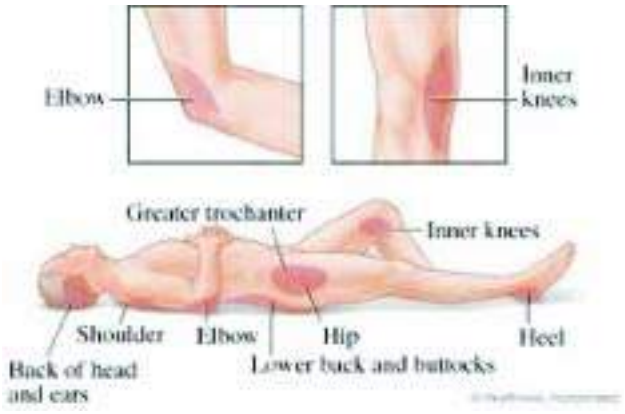
A Pressure Sore is an area of the skin or underlying tissue that is dead or dying as a result of the loss of blood flow to the area. It can begin in a number of ways. The most common is when you rest on a bony area for a prolonged period of time. The extended pressure leads to a pressure sore.

Damage from a pressure sore will range from slight discoloration of the skin (stage 1) to open sores that go all the way to the bone (severe). The affected area may feel warmer than the surrounding tissue. In light-skinned people, the discoloration may appear as dark purple or red. In darker-skinned people, the discoloration will appear darker than the surrounding tissue.

A pressure sore is serious matter and cannot be ignored as the sore may deepen in a very short time. With proper treatment, most pressure sores will heal.

A pressure sore is any redness or break in the skin caused by too much pressure on the skin for too long a period of time. The pressure prevents blood to the skin leading to necrosis of skin and even deeper tissues. Normally the nerves send messages of pain or feelings of discomfort to the brain and the person should change position to relieve the pressure, but due to damage to the spinal cord these messages cannot reach the brain.

Most common sites of pressure sores are as shown in the diagram below



The patient may need to learn new ways to change his / her position to prevent too much pressure. Pressure sores can occur, for example, when the patient sits or lies in one position too long. Shearing is also a kind of pressure injury. It happens when the skin moves one way and the bone underneath it moves another way. An example of this is if you slouch when you sit.

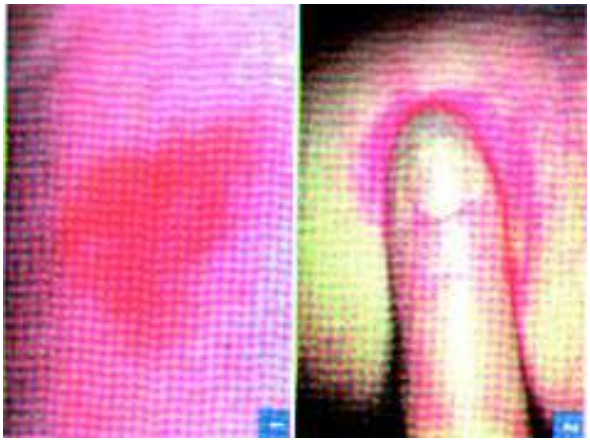
Another type of injury, an abrasion, can occur when pulling oneself across a surface instead of lifting.

This is an example of a friction injury. In addition, short exposure to high pressure, such as a bump or fall, may cause damage to the skin which may not show up right away.

STAGES OF PRESSURE SORES AND HOW TO CARE FOR THEM:

Category/Stage I: Non-blanchable erythema

Intact skin with non- blanchable redness of a localized area usually over a bony prominence. Darkly pigmented skin may not have visible blanching; its colour may differ from the surrounding area. The area may be painful, firm, soft, warmer or cooler as compared to adjacent tissue. Category I may be difficult to detect in individuals with dark skin tones. May indicate "at risk" persons.



How to recognize:

Skin is not broken but is red or discoloured. The redness or change in colour does not fade within 30 minutes after pressure is removed.

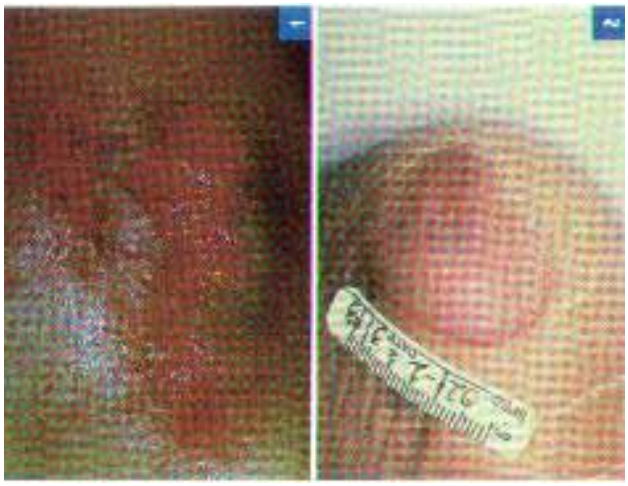
Category/Stage II: Partial thickness

Partial thickness loss of dermis presenting as a shallow open ulcer with a red pink wound bed, without slough. May also present as an intact or open/ruptured serum-filled or sero-sanginous filled blister, presents as a shiny or dry shallow ulcer without slough or bruising*. This category should not be used to describe skin tears, tape burns, incontinence associated dermatitis, maceration or excoriation.

***Bruising indicates deep tissue injury.**

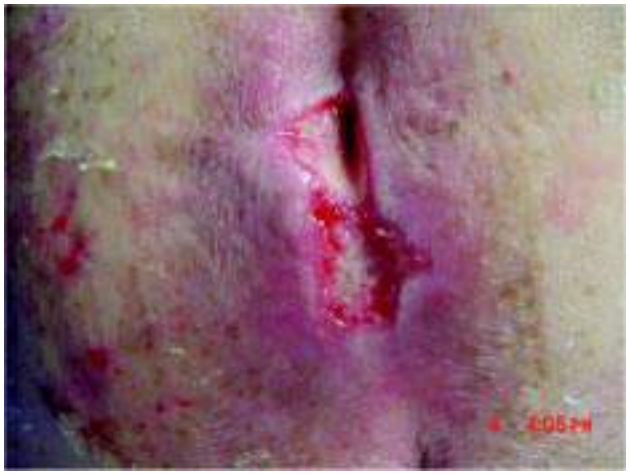
How to recognize:

The epidermis or topmost layer of the skin is broken, creating a shallow open sore. Drainage may or may not be present.



Category/Stage III: Full thickness skin loss

Full thickness tissue loss. Subcutaneous fat may be visible but bone, tendon or muscle are not exposed. Slough may be present but does not obscure the depth of tissue loss. May include undermining and tunnelling. The depth of a Category/Stage III pressure ulcer varies by anatomical location. The bridge of the nose, ear, occiput and malleolus do not have (adipose) subcutaneous tissue and Category/Stage III ulcers can be shallow. In contrast, areas of significant adiposity can develop extremely deep Category/Stage III pressure ulcers. Bone/tendon is not visible or directly palpable.



How to recognize:

The break in the skin extends through the dermis (second skin layer) into the subcutaneous and fat tissue. The wound is deeper than in Stage Two.

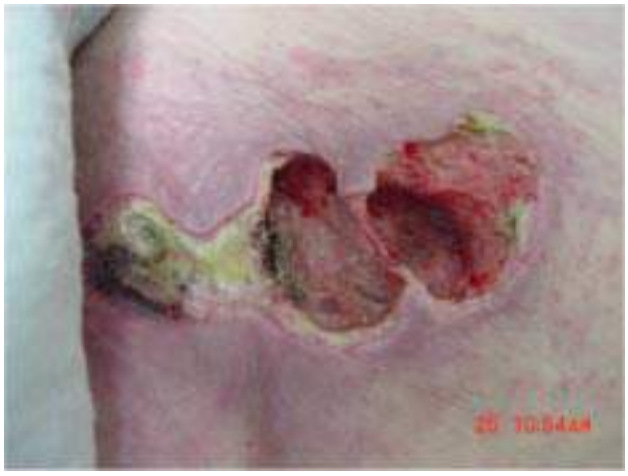
Category/Stage IV: Full thickness tissue loss

Full thickness tissue loss with exposed bone, tendon or muscle. Slough or eschar may be present. Often includes undermining and tunneling.The depth of a Category/Stage IV pressure ulcer varies by

anatomical location. The bridge of the nose, ear, occiput and malleolus do not have (adipose) subcutaneous tissue and these ulcers can be shallow. Category / Stage IV ulcers can extend into muscle and / or supporting structures (e.g., fascia, tendon or joint capsule) making osteomyelitis or osteitis likely to occur. Exposed bone / muscle is visible or directly palpable.

How to recognize:

The breakdown extends into the muscle and can extend as far down as the bone. Usually lots of dead tissue and drainage are present.



COMPLICATIONS CAUSED BY PRESSURE SORES:

- Can be life threatening.
- Infection can spread to the blood, heart, bone.
- Amputations.
- Prolonged bed rest.
- Autonomic dysreflexia.

PREVENTION:

1. **Turning:** Patients should be positioned properly in bed at all times. Patients should be turned frequently, every two hours at the beginning, and skin should be checked between turns. Turning time can be increased when hyperaemia over bony prominences resolves within 30 minutes. All positions, including the prone position and side-lying, should be used. In time, patients will be independent or able to give instructions to a caregiver in bed positioning, turning, and skin checks.
2. **Sitting:** Patients should sit no more than 30 to 60 minutes at the beginning. Their sitting time can be increased every few days, if hyperaemia resolves within 30 minutes. The patients' buttocks should be placed far back in the chair, and the footrests should be adjusted to permit the knees to be at or slightly below the level of the hips. Pressure reliefs should be performed frequently, every 15 to 30 minutes, for one minute. Recommended methods include leaning the patient forward, with chest toward the thighs, and tipping the wheelchair back 65 degree.
3. **Transfers:** Transfers should be performed carefully to avoid shearing.
4. **Equipment:**
   - During the acute care phase, the patient is frequently in a supine position and the sacrum, occiput, and heels are at high risk for pressure sores. To prevent pressure sores, orthotic devices can be used to completely elevate the head off the bed and minimal air loss beds are beneficial. Donut-type devices should not be used. Thoracic and lumbar fractures are managed by immobilization in special beds that allow movement.
   - Equipment used in the hospital should be as similar as possible to equipment that will be used after discharge in the home. Skin tolerance to standard mattresses and wheelchair cushions should be determined before discharge. Air-flotation and air-fluidized beds, for example, are suitable for hospital use in the beginning or to treat pressure ulcers, but they are not recommended for home use. An egg-crate foam mattress pad on top of a standard mattress is best suited to the home. Although wheelchair cushions reduce pressure over bony prominences, proper skin care and pressure reliefs are essential.
5. **Cushions:** Cushions should be used in wheelchairs, for many bed positions, etc. Most cushions are made of foam or gel or filled with air or water.
   - Foam cushions produce higher skin temperatures and less humidity. Custom contoured foam seat cushions are more effective than flat foam cushions.
   - Gel cushions maintain skin temperatures (although they may require cooling after

3 hours to continue to be able to do this) and produce more humidity than foam; some gel-type cushions also improve posture.

- Air-filled cushions need to be checked daily for proper air pressure.
- Water-filled cushions decrease skin temperature; however, humidity increases more on water-filled cushions.

Good body positions

Patient's position is important to relieve pressure on the sore and prevent new ones. He/she may need to switch positions whether in a bed or a chair.

While In Bed

1. Do not lie on the pressure sore. The use of pads or pillows to position the body will help relieve the pressure on specific areas.
2. Change positions at least every 2 hours.
3. Avoid lying directly on the hip bone. A 30-degree position is best.
4. When lying on the back, keep the lower legs up by placing a thin foam pad or pillow under the lower part of the legs (midcalf to ankle). Do not place the pad or pillow directly under the knee as this will reduce the flow of blood to the lower leg areas. Do not use donut shaped cushions as they will reduce the flow of blood.
5. Use pillows or small pads to keep the knees and ankles from touching.
6. Raise the head of the bed as little as possible (30 degrees or less).
7. Feed meals in an upright position to prevent choking.

Pressure Relief

Pressure sores form when there is constant pressure on certain parts of the body. Long periods of unrelieved pressure cause or worsen pressure sores and slow healing once a sore has formed. Taking pressure off the sore is the first step toward healing.

Pressure sores usually form on parts of the body over bony prominences (such as hips and heels) that bear weight when one sits or lies down for a long time. Pressure can be relieved or reduced by:

- Using special surfaces to support the body.
- Putting the body in certain positions.
- Changing positions often.

TREATMENT OF PRESSURE ULCERS ACCORDING TO DIFFERENT STAGES

STAGE I

1. Keep pressure off the sore!
2. Maintain good hygiene. Wash with mild soap and water, rinse well, pat dry carefully (but gently). Do not rub vigorously directly over the wound.
3. Evaluate patient's diet by asking - are you getting enough protein, calories, vitamins A and C, zinc and iron? All of these are necessary for healthy skin.
4. Review the mattress, wheelchair cushion, transfers, pressure releases, and turning techniques for possible cause of the problem.
5. If the sore seems to be caused by friction, sometimes a protective transparent dressing such as Op-Site or Tegaderm may help protect the area by allowing the skin to slide easily.
6. If the sore does not heal in a few days or recurs, consult the health care provider.

STAGE II

Follow steps 1-4 under Stage One. Consult the health care provider for further treatment, which may include the following:

- Cleanse the wound with saline solution only and dry carefully. Apply either a transparent dressing (such as Op-Site or Tegaderm), a hydrocolloid dressing (such as DuoDERM), or saline dampened gauze. The first two types of dressing can be left on until they wrinkle or loosen (up to 5 days). If using gauze, it should be changed twice a day and should remain damp between dressing changes.
- Check for signs of wound healing with each dressing change.
- If there are signs of infection consult the health care provider for alternative wound care ideas and review of possible causes (see step 4 under Stage One).

STAGE III

Follow steps 1-4 under Stage One and the additional steps under Stage Two. Always consult the health care provider. Wounds in this stage frequently need additional wound care with special cleaning or debriding agents. Different packing agents, and occasionally, antibiotics (creams or oral pills) may

be required. One may also qualify for a special bed or pressure-relieving mattress that can be ordered by the health care provider.

STAGE IV

Consult the health care provider right away. Surgery is frequently required for this type of wound.

How to know if the sore is healing:

- The sore will get smaller.
- Pinkish tissue usually starts forming along the edges of the sore and moves toward the centre; one may notice either smooth or bumpy surfaces of new tissue.
- Some bleeding may be present. This shows that there is good blood circulation to the area, which helps healing.

These are commonly seen complications in patients with SCI irrespective of levels of lesion, which become a challenge for the neuro rehabilitation team to tackle with.

UROLOGICAL REHABILITATION OF A SPINALLY INJURED PATIENT

Injury to the spinal cord leads to neurological damage below the level of injury & in turn produces temporary and or permanent changes in the body organs supplied by the respective nerves. In addition to commonly known changes such as muscle weakness or paralysis as well as impairment or loss of sensation, there are changes involving the urinary tract, sexual function as well as bowel. These changes add to significant physical & emotional stress and urological complications are often the major cause of morbidity & mortality in these patients. Last 3 decades have witnessed a sea change in the way we view the neurologically damaged urinary tract [neurogenic bladder]. With the concept of clean intermittent catheterization [CIC], the quality of life of patients with various forms of neurogenic urinary dysfunction has changed dramatically.

Normal urination

All of us have the ability to store & empty our bladder completely at will, at low pressure; without straining at bladder volumes within a certain limit. The reflex center for micturition [urination] is located in the sacral cord at S 2-3-4 segments. This in turn is modulated by parasympathetic autonomic system at S2-3-4 & sympathetic autonomic system located between cord level T10 to L2. The actual micturition center coordinating these is located at Pons & finally the higher social & permissive control happens at the level of frontal lobe. The sensory input is via the pelvic nerves [autonomic] & the motor output is via pelvic [autonomic] & pudendal [somatic] nerves. In general sympathetic system helps us store [S for storage] & parasympathetic system helps us empty [P for pee!] The first event during micturition is relaxation of external sphincter, followed by detrusor contraction followed by relaxation of bladder neck, leading to bladder emptying. The test to study the behavior of the lower urinary tract is called “Urodynamic evaluation”.

Types of neurogenic bladder

All patients with spinal injury go through a state of spinal shock or total unresponsiveness below the level of injury; for a period varying from a few days to several months. The urinary bladder during this phase is areflexic & asensate. Thus the patient has painless large volume retention with intermittent over flow dribble. Once the patient recovers from the spinal shock, depending on the completeness of injury, following patterns emerge. While describing the patterns, the detrusor & sphincter behavior has to be taken into consideration.

As the spinal reflex center for urinary bladder is situated in the sacral spinal cord, the injuries are divided into supra sacral, sacral or infra sacral. In supra sacral, the injury can be above the level of thoracic sympathetic outflow, that is above spinal level T6.

1 **Supra sacral injury above T6** – Here the bladder develops high pressure involuntary detrusor contractions at low volume, often leading to leakage of urine. The resting bladder pressure may be high leading to upper tract [kidney] damage. The outlet remains closed, sometimes with active sphincteric contractions during episodes of leak, leading to high pressure leak. Such patients may also develop rise in blood pressure with bladder distension, a condition called autonomic dysreflexia due to mass sympathetic discharge. This can produce dangerour rise in blood pressure & can rarely lead to intra cranial bleed. These patients clinically present with storage symptoms such as frequency with urinary leak, often associated with headaches & leg spasms. These patients usually have

constipation, with passage of hard stools every 3rd or 4th day.

2 **Supra sacral injury below T6** - These patients have a relatively safe bladder in that there is no autonomic dysreflexia. The remaining pattern is the same as that described above. The patients may also be able to predict when they will leak urine, thus enabling them to avoid incontinence. Bowel dysfunction remains the same as in group one.

3 **Sacral injury** – Those with injury at or below the sacral reflex center have poor active detrusor contractions with a fixed non contracting & non relaxing outlet. Thus they present with voiding symptoms, such as poor flow, straining to empty or frank retention associated with overflow leak & large residue. Bowels are usually constipated. The large residue predisposes them to infection & upper tract [kidney] damage.

Unfortunately majority of patients may have a partial and-or multiple level injury, thus the urinary bladder behavior also may show mixed picture.

Special mention must be made of spinal dysrhaphism or spina bifida, presenting in its various forms either at birth [meningo myelocoele] or later.[tethered cord]. Nowhere is urological rehabilitation as important as in these kids as it needs to be done right from birth with tremendous input for the parents. Typically low level lesions tend to spare the legs but involve only the bladder & bowel, manifesting as failure to empty. Higher lesions may present with various forms of paraplegia, often with upper motor neuron type bladder dysfunction +/- DESD. Such patients present with storage symptoms +/- vesico ureteric reflux. All these kids need rehabilitation in the form of starting ISC & anti muscarinic agents right from birth to avoid dangerous complication, especially renal failure in later life.

AIMS OF UROLOGICAL REHABILITATION

- To protect the upper tracts [kidneys] function
- Prevent urinary tract infections
- To prevent development of complications such as kidney stones
- To achieve continence.

Though to the patient the most important issue is to have continence; that is to be able to store & empty urine at will; medically that gets less importance as compared to the other issues such as protecting the kidneys & prevention of complications.

Management of neurogenic bladder

During the acute management of injury, all patients usually have an indwelling foley catheter. However once the patient is stable, clean intermittent catheterization [CIC] should be started either by teaching it to the patient [in paraplegics] or to the care giver [in quadriplegics.] The catheter needs to be passed every 4 to 5 hourly to mimic natural urination.

Once the state of spinal shock is over, all patients must have a baseline urodynamic evaluation, which shows the type of voiding dysfunction. The test can then be repeated every 2 years or earlier if a change takes place in the urination pattern. All patients must have an annual assessment of serum creatinine to check renal function & a sonography of the urinary tract to look for any upper tract dilatation of development of stones. As a rule adequate water intake [roughly about 2.5 to 3 liters] should be ensured to prevent super saturation of urine with crystals

Technique of CIC - The best catheter or tube for CIC is a 10 or 12 fr soft plastic catheter such as an infant feeding tube. This has the lowest chance of damaging an asensate urethra & is cheap & easily available. Though it is ideal to use a new catheter each time, the same tube can be reused if proper clean precautions are taken. The catheter & hands need to be washed thoroughly before & after CIC & the catheter should be dried from the outside & kept in a dry container without folding it. Though CIC is easy to learn for a male patient, in female patients a mirror needs to be used to help them identify the urethral opening initially. Patients should be given prophylactic antibiotics [such as Nitrofurantoin or Cotrimoxazol] in the initial learning phase to avoid infection. In female patients with severe leg spasms, special appliances can be made to keep the legs apart during catheterization. During travel or at work, such patients should keep extra catheters with them to avoid reusing the same catheter. However, in special situations, CIC is just not the most practical option, In that case any of the options mentioned below can be considered as per the clinical situation.

Timed urination - Some patients with detrusor over activity can actually time their urination. They visit the toilet at fixed times & sometimes produce reflex

voiding by supra pubic tapping with a finger or by manually pressing on the bladder. [Crede's maneuver]. Patients can also limit their liquid intake after 6 pm to avoid or reduce the night urination.

Condom catheter – In a male patient with a supra sacral injury, a condom catheter may be the most convenient management provided the bladder gets emptied with each void & there is no autonomic dysreflexia. It is also the most convenient form of management in those with leak due to a weak sphincter.

Diapers - In female patients with severe neurogenic incontinence and in some incontinent males; diapers are a practical option. Diapers are available in the market with a holding capacity of up to 1.5 litres. However in Indian weather, skin rashes & fungal infections are often the undesirable effect of having a wet diaper constantly in contact with the skin.

Indwelling catheter – In quadriplegics without round the clock care givers and in some paraplegics, an indwelling urethral or supra pubic catheter [16 Ch in size] may be the best & sometimes the only practical option. These can be changed monthly or 3 monthly depending on the type of catheter. In general incidence of infective complications is higher with an indwelling urethral catheter than with a supra pubic catheter. The urine drainage bag can be a hindrance especially during physiotherapy & mobilization. A belly bag or a leg bag is more convenient during these times. In special circumstances, a spigot can be used [to block the catheter] & released every 4 hours to empty the bladder. This, though not the most ideal option; can be considered if clean handling is ensured.

Medication for incontinence - As the commonest problem in neurogenic bladder is detrusor over activity, anti muscarinic [commonly known as anti cholinergic] drugs such as Oxybutinine or Tolterodine can be given to prevent episodes of frequency with leak. These drugs are also useful to control leak in between 2 catheterizations in those on CIC & to prevent leak by the side of the catheter in those with indwelling catheters. The commonest side effect of these drugs is constipation & dry mouth. In patients with failure to empty, sometimes alpha blocker group of drugs such as Tamsulosin or Alfuzosin are useful.

Botulinum toxin - This toxin when injected in a muscle, paralyses the muscle temporarily for a variable period from 6 months to over 1 year. Commercially available as Botox [Allergan] or Neuronox [Ranbaxy], it is injected in the detrusor muscle to control over activity or in the external sphincter to reduce its spasm. Patients with resistant form of detrusor over activity or those with DESD can benefit by this injection within the detrusor or sphincter respectively. Patients should be counseled about its temporary effect as well as about the need for ISC post such therapy. Patients with recurrence of symptoms after Botulinum toxin injection can be offered repeat injection with identiacl benefit as the first injection.

Surgical management of neurogenic voiding dysfunction - About 10% patients need some surgical procedures to maintain their urinary tract health. A detailed discussion on this is beyond the scope of this article. However, augmentation cystoplasty using a bowel patch [small or large bowel] to increase the bladder capacity may be required in severe cases of neurogenic detrusor over activity or a small capacity poorly compliant bladder. All these patients have to be conversant with the technique of self catheterization. This operation can be combined with creation of an abdominal catheterizable stoma for intermittent catheterization, especially in spastic overweight paraplegic female patients. [These patients otherwise may find it very difficult to self catheterize due to the location of their urethra.] An artificial sphincter may be required for cases with sphincteric leak. Also neurosurgical procedures such as placement of nerve root stimulator or a neuromodulator may be necessary in selected cases.

Complications in Neurogenic Bladder

With better understanding of urinary bladder dysfunction, most complications can be avoided by individualizing the management strategies.

Infection - The commonest complication is urinary tract infection. Although these patients may have only vague symptoms such as increase in leg spasms, frequent episodes of leak etc to indicate infections, presence of fever in a neurogenic bladder patient indicates urinary infection unless proved otherwise. Each attack has to be treated aggressively with appropriate antibiotics, often followed by prolonged course of preventive suppressive agents. Auxillary measures include ensuring adequate water intake with daily urine output of at least 3 liters, following correct clean technique of CIC, changing the indwelling catheters or diapers on time & ensuring proper perineal hygiene.

Calculus disease- Though the most dreaded complication, its incidence is decreasing at least in countries with appropriate bladder rehabilitation protocol. Prolonged immobilization, inadequate water intake, repeated infection, presence of foreign bodies such as catheters, inadequate bladder drainage, presence of dilated kidneys; encourage calculus formation in these patients. Smaller concretions in the bladder can be washed out but larger calculi need appropriate endoscopic management. [Cystoscopic or ureteroscopic or nephroscopic removal.] These patients may not need major anesthesia but their body deformities, spasms etc make them very challenging even for the most skilled urologist.

Renal insufficiency - Before the advent of CIC, most spinal injury patients eventually succumbed to renal failure! Thankfully that incidence has dropped substantially in recent times. In addition to the above measures, annual sonography & creatinine assessment should be carried out in all patients. Any rise in creatinine levels or development of kidney damage such as hydronephrosis must be referred to nephrologists or urologist on time. Whenever needed, dialysis should be started & in selected cases, even a kidney transplantation may be required. These transplants have to be performed in centres specialized in caring for such patients with compromised bladder function to ensure a good outcome.

In summary – From the above text, it is clear that patients with spinal injury need major urological rehabilitation. With proper management protocols, most of the management goals can be achieved & especially the complication can be prevented. Caring for these patient needs a team approach & an urologist becomes an important member of that team. The patient & his family members should be counseled about the need for urological rehabilitation right from the start. Needless to say, a sympathetic attitude of relatives, friends & colleagues at work goes a long way in boosting the morale of these patients who have to make a lot of adjustment with their body & life in general.

[No specific reference is sited in this chapter as all these treatment strategies are internationally accepted with plenty of references available in the literature. However as a comprehensive guide, the reader is requested to refer to the latest edition of Campbell's Urology textbook.]

SEXUAL REHABILITATION OF SPINAL CORD INJURED MALES

Can a Spinal Cord injured person lead a happy Family life ? Undoubtedly yes !

The physical problems faced by the patients with SCI are as:

1. Loss of Erection is called Erectile Dysfunction (ED).
2. Loss of Ejaculation is called Ejaculatory Dysfunction (EjD)

Erectile Dysfunction : Occurs when the sympathetic pathways T11,12, l1,2 and parasympathetic pathways S2,3,4 are involved.

Erectile Dysfunction can be treated with various modalities like

- Oral Erectogenic Drugs –
- Intra-Penile Self Injections
- Vacuum Erection Devices (VED)
- Penile Prosthesis

1. **Oral Erectogenic Drugs :** (mainly PhosphoDiEsterase type 5 Inhibitors) are:
   - Sildenafil
   - Tadalafil
   - Vardenafil

PDE5I have revolutionized treatment of Erectile Dysfunction. Sildenafil citrate, presently is the most commonly used oral erectogenic drug.

PDE5Inhibitors (PDE5I) like Viagra and similar drugs are available which is need to be taken only before sex but has mild to moderate side effects which is to be taken only with doctor's instructions and is not the permanent cure. These oral drugs show side effects like headache, back pain, dyspepsia, myalgia, nasal congestion, pain in limbs, visual disturbances, flushing of face with nitrates contraindicated in any form.

One should know that PDE5I is not an aphrodisiac i.e it does not increase libido nor make normal erection harder or last longer.

2. Intra-Penile Self Injections:

It is the clinician who decides what type of injection and the dose to be required. He gives the injections in prefilled syringes and trains the patient how to take it at home or giving to his wife before sex. But

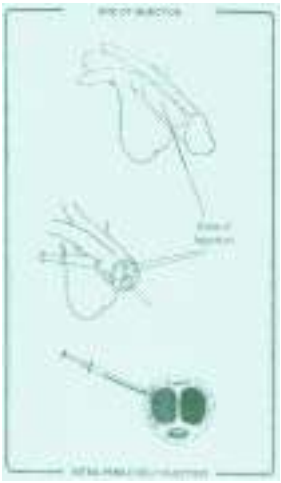


Fig1 : Intra penile Self Injection.

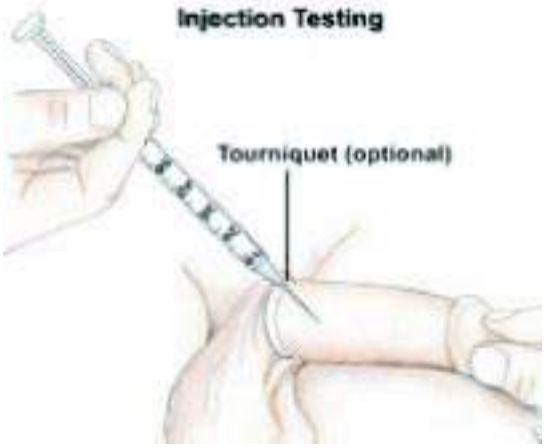


Fig2. Injection Testing.

Fig 3. Self Injection.

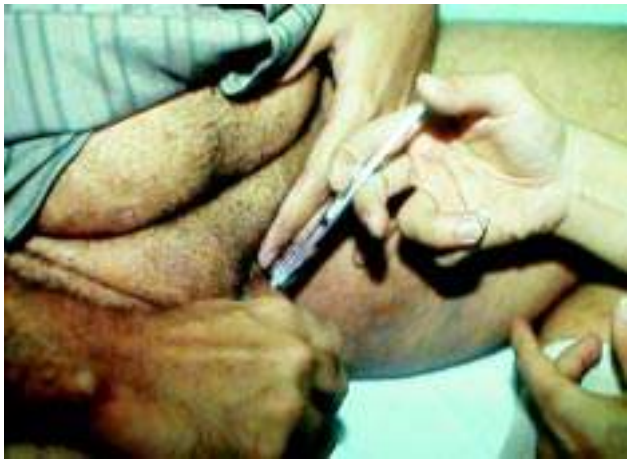


Fig 4: Wife assisted injection.

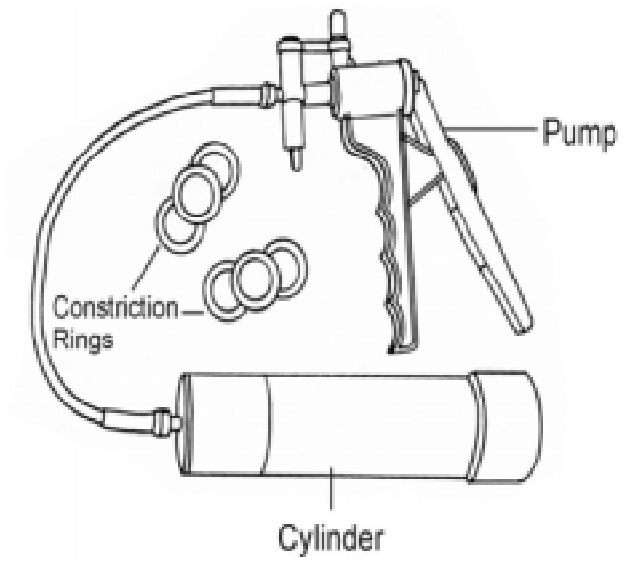


Fig 5: Basic design of Vaccum Erection devices.

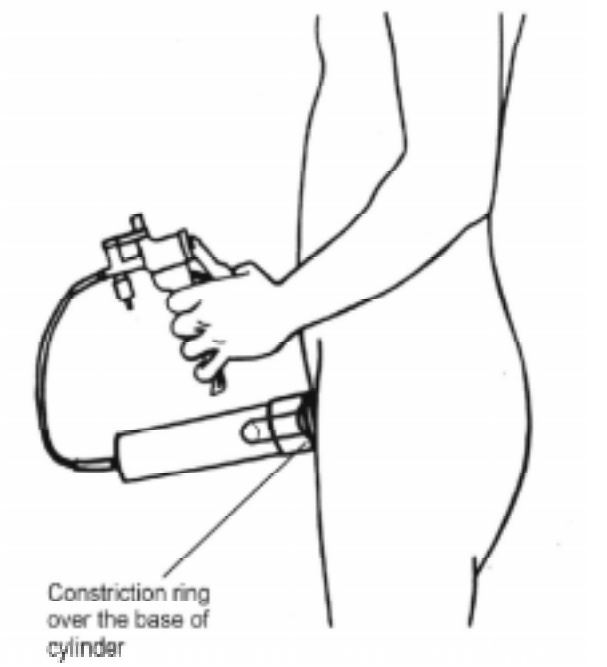


Fig 6: Constriction ring over the base of the cylinder.

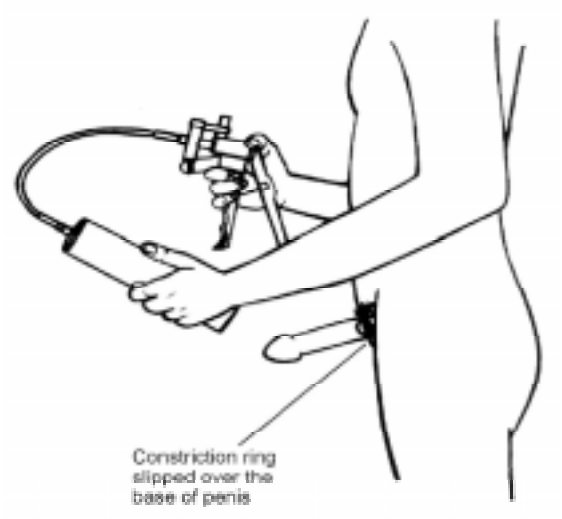


Fig 7: Constriction ring slipped over the base of the penis.

Fig 8 -10 After using the Vaccum Erection Devices.

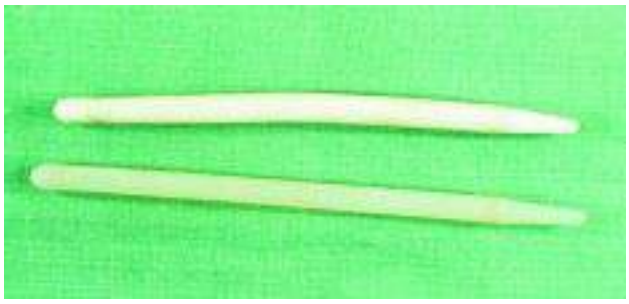


Fig 11: Malleable & Flexible Penile Prosthesis.

Fig 12a: Placement in body.

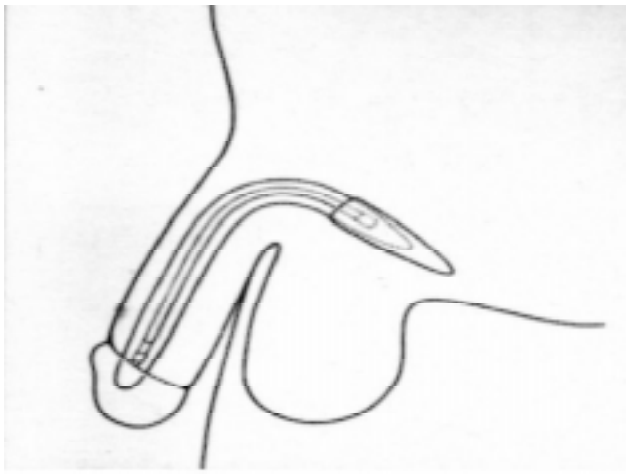


Fig 12 b: Placement in body.

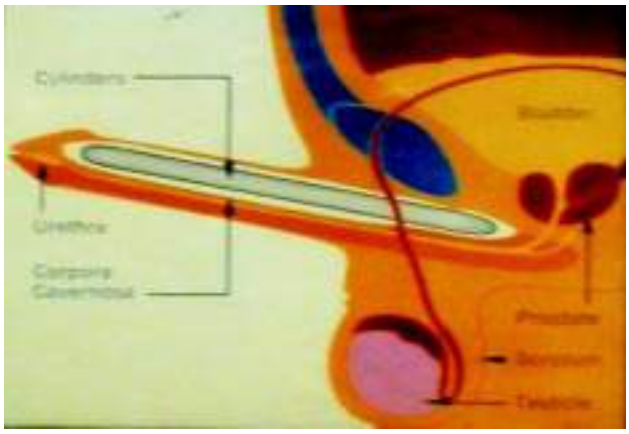


Fig 12 c: Erect State.

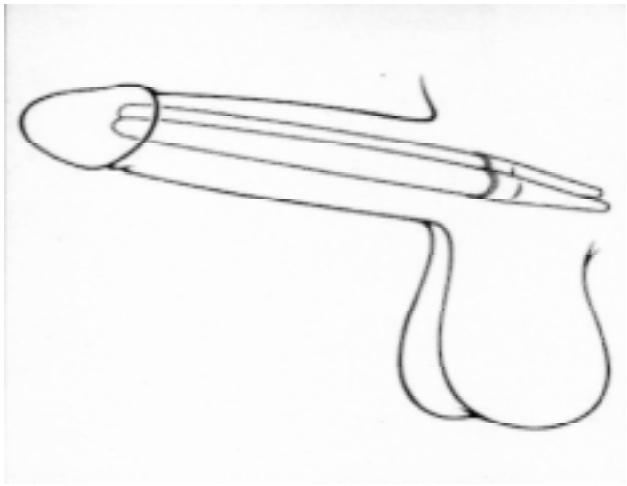


Fig 12 d: Erect State.

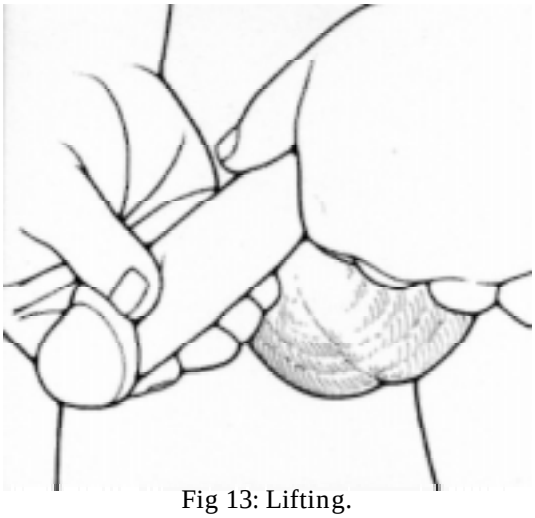


Fig 13: Lifting.

Fig 14: Inflatable Penile Prosthesis.

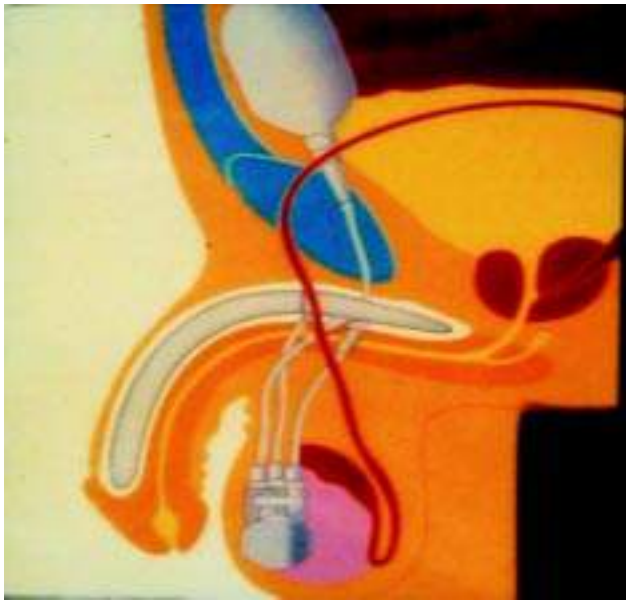


Fig 15 a: Placement in Body.

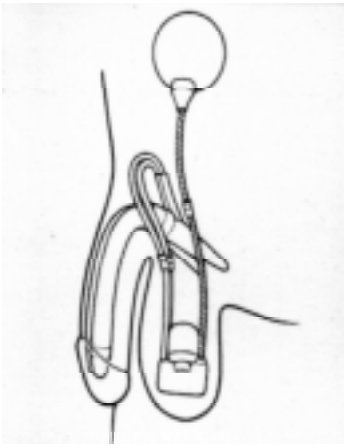


Fig15 b: Placement in Body.

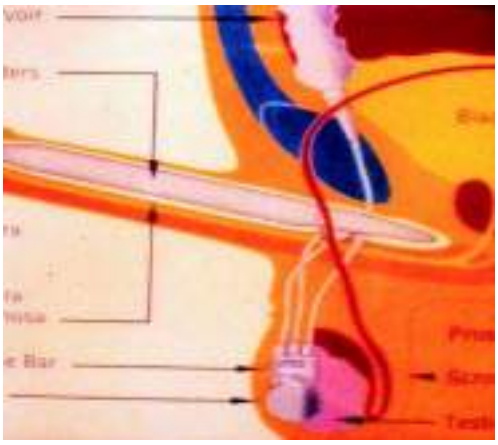


Fig 15 c: Erect State.

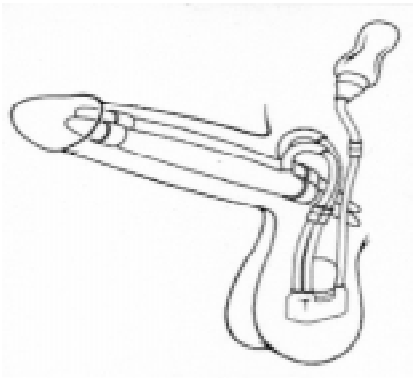


Fig 15 d: Erect State.

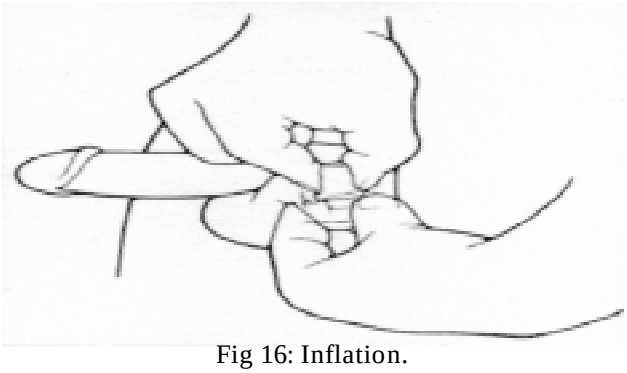


Fig 16: Inflation.

Fig 17 a : Vibration Therapy.

Fig 17 b: Vibration Therapy.

Fig 17 c: Vibration Therapy.

Fig 18: Seager's Electro Ejaculator.

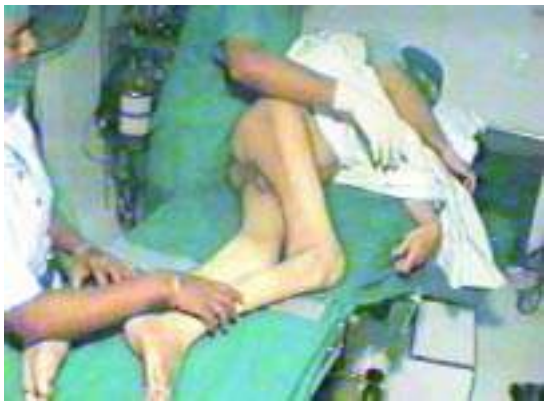


Fig 18a: Seager's Electro Ejaculator Procedure.

Fig 18 b: Seager's Electro Ejaculator Procedure.

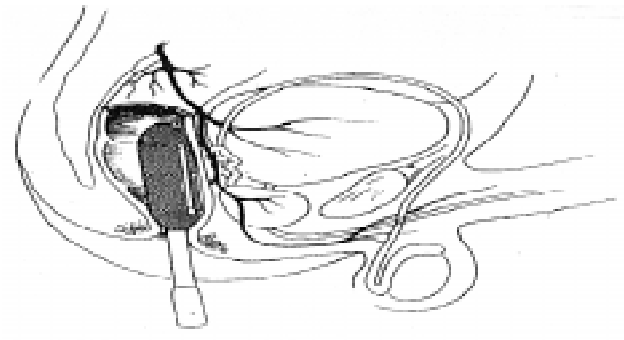


Fig 19: Ejaculation in antegrade direction.

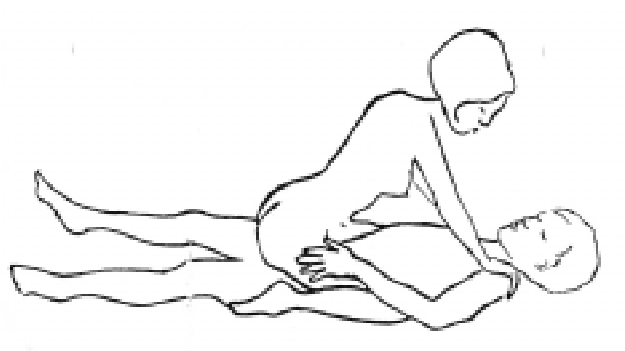


Fig 20: Position with women on top.

Fig 21: Testicular sperm retrieval techniques done percutaneous.

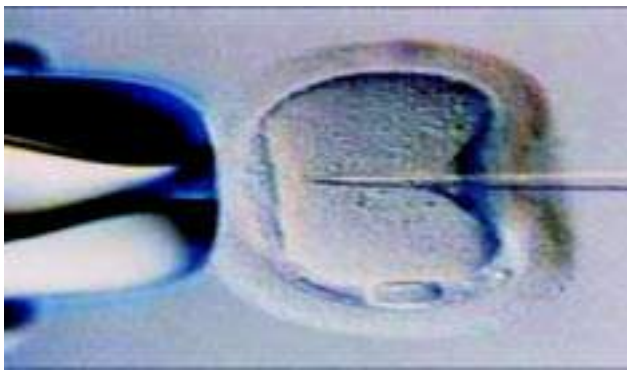


Fig 22: Intra Cytoplasmic Sperm Injection.

it is not a permanent cure. The commonly used drugs for Self Injections are Papaverine and Prostaglandin E1 in combination with other drugs like Phentolamine, Chlorpromazine, Atropine.

The Stepwise procedures done in Home Self Injection Program include the following:

1. Therapeutic dose determination by the clinician.
2. Training & supervised self injection by the patient
3. Home self-injection program.

Patient's Guide to Self Injection – A simplified pictorial guide to help the patient.

Fig1 : Intra penile Self Injection. Fig2. Injection Testing. Fig 3. Self Injection. Fig 4: Wife assisted injection.

Side effects:

The local side effects like noduleor small hematoma can happen but most important and dreaded side effect is PRIAPISM which is a rigid erection lasting for more than 4 hours. If not treated immediately, it can cause permanent inability to achieve erection.

3. Vacuum Erection Devices (VED):

1. Time-tested.
2. Non-invasive9.
3. For Select Population

BASIC DESIGN

1. Cylinder
2. Vacuum pump
3. Constriction rings

Fig 5: Basic design of Vaccum Erection devices. Fig 6: Constriction ring over the base of the cylinder. Fig 7: Constriction ring slipped over the base of the penis. Fig 8 -10 After using the Vaccum Erection Devices.

Patients who have limited frequency coitus can be advised to have a combination of intra penile injection and vaccum erection devices thereby avoiding surgery for prosthesis.

Surgery of Penile Prosthesis :

When none of the abovementioned modalities are effective enough to make penetrative intercourse possible, then surgery for prosthesis is resorted to.

There are essentially two types of prosthesis available.

- Non-Inflatable
- Inflatable

Non Inflatable Type- These are solid rods of medical grade silicon. There are two types again-

- Malleable
- Flexible

Fig 11: Malleable & Flexible Penile Prosthesis.

Fig 12a: Placement in body. Fig 12 b: Placement in body. Fig 12 c: Erect State. Fig 12 d: Erect State. Fig 13: Lifting.Fig 14: Inflatable Penile Prosthesis.

The components of inflatable penile prosthesis are as:

1. A pair of cylinders
2. A Reservoir
3. A Pump

Fig 15 a: Placement in Body. Fig15 b: Placement in Body. Fig 15 c: Erect State. Fig 15 d: Erect State Fig 16: Inflation

Thus with such wide ranging treatment modalities available, erection dysfunction in a Spinal cord injury male can be treated effectively.

Factors such as age, motivation, financial status and a cooperative partner plays an important role for the individual patient.

Ejaculatory Dysfunction :

Spinal cord injury does not disrupt sperm production but it affects the sperm transport to outside. The spinal centers of S2,3,4 (Onuf's nucleus), when they get involved in the spinal injury, the ejaculatory dysfunction occurs. In most cases, the sperm instead of coming antegrade (outside) tend to go Retrograde (backwards) into the urinary bladder. Hence inspite of being able to have sexual intercourse, the man cannot deposit his sperms into the vagina leading to infertility.

The main causes of infertility are as :

1. Inability to perform coitus.
2. Inability to ejaculate intra-vaginally.
3. Altered seminal parameters due to infection.

To overcome these problems, the couple requires doctor's assistance. Hence we have Assisted Reproduction Techniques (ART) which are as follows:

1. IUI (Intra Uterine Insemination) with sperm-preparations

2. IVF (In Vitro Fertilization)
3. ICSI (Intra Cytoplasmic Sperm Injection) / IVF

For every procedure mentioned above we need the patient's sperms.

Modalities to recover sperms from SCI patients include:

1. Vibration therapy
2. Electro-ejaculation
3. TESA (Testicular Sperm Aspiration)

Vibration therapy: is used to stimulate the undersurface of glans thereby enabling the patient to fantasy erotic pictures. The procedure takes upto one hour where patient sits comfortably. It requires privacy and needs upto four sessions.

Fig 17 a- 17 c.

If vibration therapy is not successful in producing semen from the SCI patient, then next modality used is:

Electro-Ejaculation under anesthesia: is a special instrument which uses direct current for stimulation. SCI patients do not need anesthesia but the equipment is quite expensive.

Fig 18: Seager's Electro Ejaculator. Fig 18 a-18b.

In this metal plate on the rectal probe is directed towards prostatic plexus and the current in increasing voltage (5-50 V) is delivered. At the optimum voltage for that particular patient, ejaculation mostly occurs in antegrade direction.

Fig 19: Ejaculation in antegrade direction. Fig 20 : Position with women on top.

The sperms thus obtained are injected into his wife on the day of her ovulation, resulting in pregnancy.

If this procedure also is unable to result in pregnancy, then further IVF techniques can be used where the sperms are directly collected from Testis for test tube baby. Thus, these sperms are injected in his wife's mature eggs where they show fertilization and when growth happens they are transferred back into her uterus resulting in pregnancy.

Obtaining Sperms directly from the testis for the purpose of IVF – ICSI (In Vitro Fertilization – Intra Cytoplasmic Sperm Injection) includes testicular sperm retrieval techniques which are done Percutaneously by the following methods:

1. TESA – Testicular Sperm Aspiration
2. NAB – Needle Aspiration Biopsy
3. Trucut Needle biopsy

Fig 20: Testicular sperm retrieval techniques done percutaneous.Fig 21: Intra Cytoplasmic Sperm Injection.

IVF-ICSI procedure is very expensive and exhausting where only 30-35% chance of success is seen.

ICSI has revolutionized the management of male infertility including azoospermia.

Thus Sexual Rehabilitation plays an integral part of total rehabilitation where the patient is guided correctly by giving the proper awareness of modalities of treatment available by the clinician. Socio-economic conditions also play a major role in availing of the treatment to the patient.

PSYCHOLOGICAL REHABILITATION OF SCI

Spinal Cord Injury (SCI) leaves a major impression on the person's body and mind. A new spinal cord injury patient usually has many queries regarding his future and at the same time has a sense that things are not going to be the same. A person who had been leading an independent satisfying life becomes immobilized, bowel and bladder incontinence, loss of sexual functioning and becomes dependent on others for every small necessity. The patient not only faces loss of body control but also experience changes in self worth, sense of independence, confidence, attractiveness, sexuality, and relationship with family and friends.

There are various stages that one goes through post spinal cord injury: 1) shock and denial 2) grieving followed by depression or vice versa 3) anxiety / frustration 4) anger / aggression 5) trying to adapt to the situation. The patients may go through all the above stages or they might not go through the stages given in the above order or they might skip some stages.

Shock and Denial:

When the patients come in terms with what has happened with them they are often not yet ready to acknowledge, the extent or permanence of their disabilities. At the other extreme, patients sometimes build denial systems based on unrealistically high hopes. Clear communication, emphasizing realistic expectations before introducing treatment, may prevent some of these

responses. They should be provided with information on obtaining recommended future.

Greif and Mourning:

Working through grief and loss is the way people adjust to losses they have sustained. This adjustment requires emotionally "letting go" of something that was valued but can't be replaced. It is only after letting go that the person is free to re-invest their emotions into new things. However, letting go can be difficult, complex, and require a great deal of time.

Tasks of Mourning:

Four tasks of mourning that a person needs to successfully work through while grieving.

1. **Accepting the Reality of the Loss:**

   Patients and families struggle long and hard with this reality. For some, the struggle and disbelief continues for years and some never do accept the reality of their loss and will forever be in mourning. Their emotional energy goes to fighting against the reality of the spinal cord injury / disease instead of concentrating on strengthening and using the remaining intact muscles and nerves. Those who do accept this reality, even a part of it, can then move forward to the next step.

2. **Experiencing the Emotional Pain Associated with the Loss:**

   Emotional pain is a reflection that something which has been valued or treasured has been lost. The emotions are often very strong and difficult to manage. Anger, sadness, hopelessness, fear and a sense of injustice are very common. Those individuals who do not successfully handle this task in the grief process may, over the long term, lose motivation and interest in most or all activities, withdraw from friends and family, become bitter and resentful, stay depressed, develop poor personal hygiene or eating habits, seriously abuse drugs and alcohol or deteriorate physically. Those persons successful at this step can move forward to the next.

2. **Adjustment to the Environment despite Spinal Cord Injury:**

   Adjustment means effectively learning to deal with the world despite the changes caused by the spinal cord injury / disease. A person may need to learn new skills because he cannot perform his old job.

3. **Withdrawing Emotional Investment:**

   The final stage of mourning involves the actual "letting go" of life as it used to be before the spinal cord injury / disease and investing energies elsewhere. Perhaps the person learns to participate in an activity that he did prior to hospitalization but now does it in a different manner; or the person who was very "body" oriented learns to use the "mind" more for stimulation, satisfaction, and productivity.

4. **Anxiety:**

   A panic like reaction of initial recognition of the enormity of the traumatic event takes place as the patient is relatively unaware. As, the patient comes into terms with the injury and the consequences the level of anxiety eventually subsides.

Depression:

Depression is a common illness and it can affect anyone. However, it is more common among SCI patients as about 1 in 5 people. Estimated rate of depression among people with SCI ranges from 11% to 37%. Krause, et al suggests that 48% of patients with SCI in 1997 had clinical symptoms of depression at a year or more after injury. Another study showed that 60% of Portuguese patients with spinal cord injury have depressive symptoms.

Suicide:

In Denmark, a suicide rate is 5 times higher than a general population and it is not related to the severity of the injury. Suicidal tendencies are higher during the initial days after spinal cord injury.

Independence:

Causes of depression after spinal cord injury and found that social support and recent stressful events can be used to identify patients at a high risk of depression but that they are less likely to become depressed if they are independent. Adjustment to spinal cord injury and quality of life can be adversely affected by inadequate home facilities that make a person more dependent (Seki, et al., 2002). Expectations of independence decline steadily with increasing age. In some patients, there are secondary gains in their dependent state, though they may not be consciously aware of this.

Body Image:

Many spinal cord injury patients value the fact that they look "normal" except for the wheelchair. The magnitude of disability may be "invisible." Patients sometimes report that people stare at them more. Their sense of "being different" and social discomfort increases. Spinal cord injury patients may not integrate disability into their self-concept for some time.

Adjustment:

Affective internalization, of the functional implications, of the disability along with behavioural adaptation to newly perceived life situation. True adjustment and adaptation begins after discharge from rehabilitation.

Family Issues:

Spinal Cord Injury is a life altering event not only for the person but also for family member. Family member also suffer from the various stages that a person goes through post spinal cord injury. Findings suggest that the spouse of a person with spinal cord injury usually suffers from emotional stress that is comparable to or greater than those of injured partner. The spouse has to now take on overall charge of the patient, himself or herself and other family members. They normally have to juggle out time for everything. This leaves the caregiver at a higher risk of physical and mental stress, burnout, anger, fatigue and resentment.

Spouses:

The burden of caregiving most frequently falls on the spouse. Caregiving spouses are often severely stressed, particularly due to health issues that arise after spinal cord injury. Caregivers have a higher incidence of physical stress, emotional stress, burnout, fatigue, anger, and resentment (Weitzenkamp, et al. 1997) than their partners or spouses who are not the caregivers. Chan, et al. (2000) report that the impact of spinal cord injury is more severe on marriages that began before than after the injury.

Parents:

When a person is young and not married, the burden of caregiving frequently is taken up by the parents. There is often nobody else to help ease out the burden of older parents. Parents often do not forget the incident for years and decades. It is not unusual to find tears in the eyes of a parent of a person with spinal cord injury when they talk about the accident and the events that follow. Feelings of helplessness and hopelessness, guilt, and depression often pervade their lives for a long time. There may also be differences between the responses of mother and fathers.

Children:

The disruptive and impoverishing effects of spinal cord injury on families, most people assume that spinal cord injury of a parent has deleterious effects on children. Killen (1990) assessed roles of children in families after spinal cord injury and found that spinal cord injury did not change the roles, i.e. mothers, fathers, husbands, and wives continued to play their traditional roles.

Marriage:

The marriage rate of people with spinal cord injury is lower than the general population but the factors that influence marriage rate may differ from what is commonly assumed. Disability appears to exert a greater effect on the marital status of females than males. Although medical complications have an adverse impact on quality of life of adults with pediatric-onset spinal cord injury, the presence of pressure ulcers, severe urinary tract infections, and spasticity have little effect on marriage rate of adults with pediatric onset spinal cord injury. In contrast, the presence of head injury has much more adverse effects on marital status.

Sexual satisfaction:

Spinal cord injury obviously impairs sexual function. However, sexual satisfaction does not appear to be related to physical factors such as erectile function, genital sensation, or orgasmic capacity as much as perceived partner satisfaction and relationship quality (Phelps, et al., 2001). Fisher, et al. (2002) assessed sexual function in 40 people (32 men, 8 women) with spinal cord injury. By 6 months after discharge from hospital, most of the participants in the survey had made significant changes in sexual behavior and activity. Most of the respondents had realistic concerns coupled with more requests for sexual health intervention. However, many were engaging in sexual activity. In Iceland, 55.5% of spinal-injured people are married and 71% had an active sexual life after injury (Knutsdottir, 1993). Thus, sex is important but is not necessary for sexual satisfaction or sexual activity, and is frequently not the most important factor in life satisfaction.

Psychological Assessments:

Psychological assessments are conducted on the patient to gauge the level of impact that the injury of the spinal cord has caused. As, this is a life changing event which causes the patient to make constant adjustments and impacts all the sphere of life, hence evaluation of all the aspects is required to see the coping mechanisms of the patient.

- Becks Depression Inventory (BBDI – II): This is a 21-question multiple-choice self-report inventory, one of the most widely used instruments for measuring the severity of depression. Each answer is scored on a scale value of 0 to 3. The cut-offs used differ from:

0–13: minimal depression; 14–19: mild depression; 20–28: moderate depression; 29–63: severe depression.

Higher total scores indicate more severe depressive symptoms.

- **Stress:** The Depression Anxiety Stress Scales (DASS): [1] is made up of 42 self report items to be completed over five to ten minutes, each reflecting a negative emotional symptom [2]. Each of these is rated on a four-point Likert scale of frequency. These scores ranged from 0, meaning that the client believed the item "did not apply to them at all", to 3 meaning that the client considered the item to "apply to them very much, or most of the time". It is also stressed in the instructions that there are no right or wrong answers.
- **Quality Of Life Questionnaire:** The SF-36: This is a multi-purpose, short-form health survey with only 36 questions. It yields an 8-scale profile of functional health and well-being scores as well as psychometrically-based physical and mental health summary measures and a preference-based health utility index.
- Suicide Risk Questionnaire to assess the suicidal ideation in a person with spinal cord injury.
- Hamilton's Psychiatric Rating Scale for Depression to assess the severity of depression.
- Hamilton Anxiety Rating Scale is conducted on the patients to gauge the level of anxiety in the patients.

PSYCHOLOGICAL INTERVENTION:

Psychological intervention is important as it acts like emotional catharsis, and gives the opportunity for the patient to tell his life-story, which enable him to gain a sense of order and perspective. It also helps the patient to vent out his emotions and feelings regarding the event when spinal cord injury took place. Also, the patient is able to discuss with the psychologist the emotional issues regarding family, work, and physical state.

Individualised Psychotherapy:

Hope is a potentially important coping strategy for both the person and family with spinal cord injury. Goal-directed hope based on realistic perceptions of life, focusing on progress, positive interpretation of events, and goal setting are important in helping people and families cope with spinal cord injury.

The psychologist can help the team of therapists to understand the patient's stage of adjustment, and provide consultation on behavioural management approaches [3]. Emotional responses dealt with by psychotherapy include a range of ego defences, most commonly repression and denial. Typically, as denial decreases over time, depression, anxiety, and anger increase. How these emotions are expressed depends largely on the patient's premorbid personality style. Psychotherapy can help via reinforcing adaptive coping skills and teaching new coping strategies. The psychologist may also work with the interdisciplinary team to develop behavioural modification programs, based on learning theory, to decrease these behaviours. Contingency management and behavioural "contracting" are, most frequently used in rehabilitation settings [4]. Approaches emphasizing positive reinforcement to "shape" desired behaviours are particularly effective. Cognitive therapy is used to help the client overcome the negative and distorted view of himself or herself or people around him or her.

Group Therapy:

Psychological treatment of spinal cord injury often includes group psychotherapy, which is an excellent method to both maximize patient learning and efficiently use therapist time. Patient groups can provide emotional support, peer role models; teach new coping skills, and decrease social discomfort. Likewise, multiple-family group psychotherapy is a powerful and effective tool for facilitating family adjustment to spinal cord injury. Family members experience similar emotional responses to the patient and similarly benefit from psychological intervention. If not included in the team effort, a well-meaning family member could inadvertently sabotage the independence-oriented rehabilitation approach, or be too psychologically

distressed to provide the emotional or physical care the patient needs.

Sexual Counseling:

Establishing a healthy sexual relationship may require professional help. Couples or individuals who get sexual counseling can learn effective ways to communicate feelings. Patients who are wheelchair bound usually are embarrassed about their body and their physical state and hence they need individual psychological session to help open up about their feeling and be comfortable or find solutions in their condition. Studies show that males with spinal cord injury want information about sexual issues. Those who receive the proper information have more positive sexual relationships.

Caring for the Caregivers:

Caregivers, play a major role in the life of patients with spinal cord injury. As, caregivers are mostly looking after the patient, all the time they should avoid having burnouts. Hence they should find time for themselves and their recreational activities, because if the caregivers are emotionally and physically well balanced they will be able to attend to the patients need in a better way. Also, they would be able to give their maximum support in improving the patients condition.

Other Therapies:

Other issues which need to be routinely addressed by the psychologist, in conjunction with the rehabilitation team, are vocational rehabilitation and pain management training. Prevention of medical complications particularly, those which have significant behavioural/emotional components, need to be emphasized. For example pressure sores, which often occur when depression and/or substance abuse lead to poor self care.

REHABILITATION – PERSPECTIVE OF A SPINAL CORD INJURED ACHIEVER

Dr. Ms. Ketna L. Mehta, PhD. is a founder Trustee of a 10 year old NGO, Nina Foundation – Rehabilitation of people with Spinal Injury. They have received prestigious awards such as NCPEDP Shell Helen Keller Award, NASEOH Award, featured in the Limca Book of World Records and empowered over 600 persons with Spinal Cord Injury. She is a Management Educationist, Editor & Associate Dean, Research, Welingkar Institute of Management Development & Research, Mumbai.

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I am Ketna Mehta, and I met with a paragliding accident on 12th February 1995 leading to a T12 burst fracture. That's when I came to know that God wills differently for me, because my life lay totally changed from a completely independent, working professional to being dependent on my near and dear ones even for my basic chores, initially after the accident.

Little did I know that I would be able to walk again with the help of a walker and come to lead a life that I can call independent. It only became possible due to the help and support of my family, medical team and consistent efforts of the Spinal Cord Injury (SCI) – REHABILITATION TEAM

It was like being reborn, re-learning how to perform every single activity of daily living in a new way, to adjust to my new self. But yes, I can surely say, as toddlers our parents rejoiced every new step we took towards growth when we had no clue as to why we were being hugged and kissed for standing up or walking those baby steps. Looking at the brighter side of my disability I got to rejoice and celebrate every new step I took towards independence.

Spinal Injury (SI) is an aspersion to the spinal cord resulting in a change, either temporary or permanent, in its normal motor, sensory, or autonomic function. People affiliated with Spinal Injury usually have permanent and often devastating neurologic deficits and disability.

Spinal Injury is a high cost disability leading to drastic changes in an individual's life. Due to the many changes in the life of a person with spinal injury emotional and psychological support becomes an essential factor. Also, the financial impact of Spinal Injury is extremely high as the disability leads to lengthy hospitalization, medical complications, extensive follow up care and recurrent hospitalizations.

Rehabilitation is a reiterative, active, educational, problem solving process focused on a person's behaviour (disability), with the following components:

- Assessment – Persons Problem
- Goal setting

- Intervention – Treatment and Support
- Evaluation – to check the effects of intervention.

The rehabilitation process aims to:

- Maximize the participation of the person in his or her social setting
- Minimize the pain and distress experienced by the person
- Minimize the distress of and stress on the person's families and corers.

A rehabilitation service comprises a multidisciplinary team of people who

- Work together towards common goals for each person
- Involve and educate the person and family
- Have relevant knowledge and skills
- Can resolve most of the common problems of their persons.

This definition emphasises the importance of the team skilled in achieving the professionally perceived clinical outcome for the individual with spinal injury.

On 12th Feb 1995 at 12 noon, on the outskirts of Mumbai, I was in a paragliding camp and this was my last flight. I trekked up the 40 feet hill and took off. Suddenly the wind turned and I swerved to my right without control crash-landing on the rocky ground with huge impact. T12 burst fracture with para paresis, was the verdict. I assimilate important pointers which were favorable in my journey of Rehabilitation (practical aspects of SI rehabilitation):

I was carried in the big tent cloth supine with four people holding the cloth on either side.

I was laid out straight on my back on rear seat of the ambassador car from the site of injury up to the town. (How we are handled at the time of accident is very important. I was fortunate!)

I was taken to Dr. Riten Pradhan an astute and knowledgeable orthopedic surgeon whose hospital was in Virar. He examined and diagnosed my condition as Spinal cord injury. He arranged to transfer me to Hinduja Hospital. He took me by train to avoid jerks and bumps of the Mumbai roads. He had administered methyl prednisolone which I was told would help in the recovery if taken within 6 hrs after the trauma. Since he accompanied me through the journey I had I felt reassured psychologically.

At Hinduja hospital, to my greatest luck Dr. S. Y. Bhojraj, an ace spine surgeon was available and he examined me and arranged for MRI, X-rays and other investigations and fixed me for surgery in the same night itself. To have the best doctors both at the site and hospital according to me was a critical aspect of my rehab journey.

After the surgery, Dr. S. Sagade, my Urologist gently informed me about my bladder situation and advised me to drink over 2 liters of water each day and keep a tab on the urine output. I had an indwelling catheter for 20 days in the hospital.

I was informed by Dr. Bhojraj that the surgery was for decompression and stabilization by a Steffi plate and 4 screws. I was also told that I was on a waterbed in the hospital and that I should turn every two hours on my sides to avoid bedsores. Later I was put on regular mattress and was asked to change positions frequently.

I was put on regular physiotherapy at the hospital by Sharon Vakharia my first Physiotherapist. She made me exercise regaling me with funny anecdotes and jokes. It was fun time everyday. She also told me about two other people with spinal injury in the ward and urged me to visit them once I was mobilized on a wheelchair. My urge to motivate and help others started right from the hospital. My sister Nina started me on homeopathic medicine immediately to aid my neurological recovery and keep me in a happy and jovial frame of mind. My movements, sensations, bladder and bowel were not functioning.

I was discharged from the hospital after 20 days stay.

My house was on the first floor and had to be carried up physically.

Dr. Milka Vivek was my 2nd Physiotherapist who attended me at home. She gave me a lot of courage. Later, Dr. V. C. Jacob from Sion Hospital came home to help me out. He was attending to me for 2 long years. His experience with SCI was indeed beneficial for me.

Dr. Jacob taught me several tricks – climbing stairs by personally getting railings installed, he ordered good thigh high calipers with shoes which locked at the knees, then graduating to ankle high shoes and also gave me a goal to help others with SI. To have an ace senior physiotherapist to guide, advice

and correctly assess my condition was a very important aspect of my rehab. He also took me to Paraplegic Foundation a charitable organization for comprehensive care for SCI persons and told me to dedicate one day of the week for my other friends with spinal injury. I shared, counseled and learnt about their issues and worked out solutions.

Education and Counseling by a Physiotherapist:

A person with spinal injury requires extensive physiotherapy to gain strength, lost muscle tone and to become largely functional again. During the treatment a physiotherapist plays a vital role in bringing about a positive approach towards the treatment of the person. A caring touch, constant reassurance and reaffirmation from the physiotherapist help the person to develop a positive approach as well as to look at the brighter side of life. The person needs to be explained the nature of the treatment, need for compliance and the importance of the regularity of the exercise.

She / he is informed about the condition and the likely outcomes. A team approach including physiotherapist, occupational therapist, social worker and person with Spinal Injury herself / himself will help the person gain not only physical independence but economic independence and social acceptance as well. The relatives and person with Spinal Injury are as much a part of the team as the professionals and must be considered at all times, because eventually it is they and the person herself / himself who will share the responsibility for the success or otherwise of attempts to restore her / him for an independent life in the community. In the case of the most severely disabled person the relatives may be able to undertake nursing care at home after suitable instructions and with the necessary equipments.

My mind was active professionally too. I had my management consultancy and continued with my market research projects – I had a PC & workstation and would work using the phone, hold meetings at home. It's an important aspect of rehab to be active and do what we enjoy.

With the interaction with the Rehab team I could learn a lot about the SCI, its complications like Osteoporosis, Hypercalciurea, Urinary calculii and their management. I also learnt the importance of dynamic weight bearing, risk of developing contracture and so on. I feel that it is the duty of Physical Therapists to explain to the SCI persons the benefits of Exercises and the complications that could happen in the absence of it.

It would be ideal to do a total period of 6 hours of exercises for 6 days a week for 6 months regularly and then continue for 2 years with other activities also which includes walking long distances (even in the crowd) and various sports activities.

My sister and brother -in- law being doctors taught me safe Clean Intermittent Catheterization (CIC) method. I slowly learnt to manage myself. Eventually, I could manage both the bladder and bowel program myself.

Dr. Jacob urged me to teach other female spinal injury persons the art of doing self catheterizations, which I did very proudly.

I took part in various sports activities. Sports are meant not only for recreation but also for improving strength and balance. No time should be devoted for idleness and self pity please! There should be social outing to parks, beaches, sea faces, movies, plays, wedding, temples etc. Each outing should be done with a sense of adventure. Never should feel embarrassed about the disability or the appliances.

Travelling was part of my profession. I even ventured travelling by public transport. People of Mumbai in general would always give a helping hand if you ask for it. I have no hesitation to hold on to two persons' shoulders and climb up or down. Therefore found that nothing is inaccessible. "God bless you" became my lexicon.

I took several trips abroad for holidays with my family. I even took a solo trip to Thailand for 7 days to attend a conference. This was a major adventure. It certainly boosted up my morale further, feeling a sense of achievement!

Sports and Recreation have helped me in my mental health. Playing Wheelchair games like Throw ball and W/C tennis also gave me a lot of pleasure.

Levels of Rehabilitation

(Source: "Market Potential Study for a World Class Spinal Cord Injury Rehabilitation Centre in Mumbai" PhD thesis by Dr. Ketna Mehta – India, 2008.)

Nina Foundation (www.ninafoundation.org) is an NGO founded by us in 2001 to spread awareness about prevention & rehabilitation of people with Spinal Injury. We also conduct educational seminars at various physiotherapy colleges, sensitizing the budding Physiotherapists.

Rehabilitation plays a major role in the life of a person with spinal cord injury.

Having a spinal injury is a life changing event for both the person and their loved ones. The aim of the entire team is to make the treatment sessions effective and enjoyable. All one needs is to explain to the person with spinal injury that they need to give it a try, work up ways and means, ask for help whenever required and not lose hope and calm; all they need to have in their mind is that they need to reach their goal. There is just a need to stimulate that urge for living and reignite the fire for setting goals and achieving them in a manner possible to them.

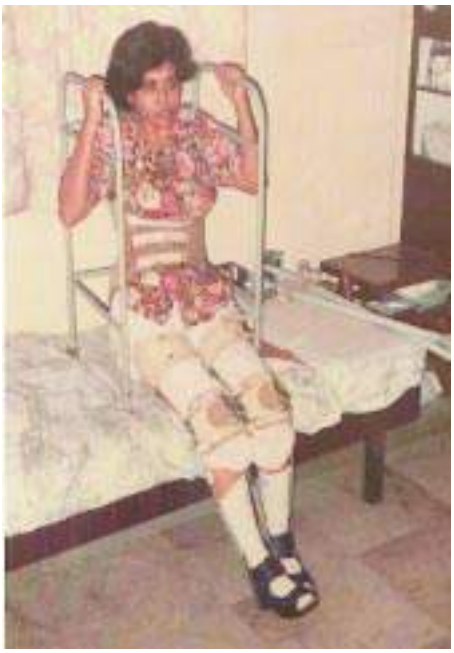
A physiotherapist becomes a friend, confidante, story teller, advisor and life coach. The trust reposed on the physiotherapist is immense and with tremendous patience can resurrect the life of the person with Spinal Injury bit by bit by bit – always being positive & keeping hopes alive.

My second life started after an accident at the time of an adventurous sport, but my adventure continues with a sense of achievement after going through a successful Rehabilitation.

Summary:

This chapter has dealt with almost all aspects of rehabilitation of SCI with special emphasis on team approach . Management of the SCI is a complex and challenging task, if all members take it up with commitment, great things can be achieved in different stages. Frequent communication among the team members , patient and family is the key factor and this alone can bring the person back to the society and lead a dignified and meaningful life.

PUSH UP

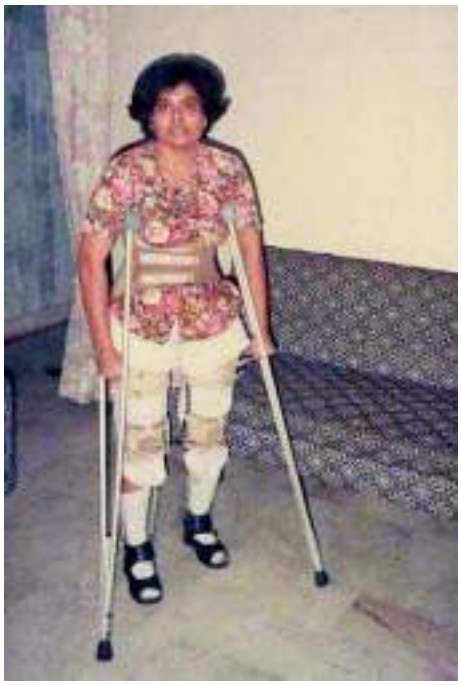


PULL UP

TRANSFERS FROM BED TO FLOOR

READY TO CRAWL

REACHOUTS IN ALL FOURS



WALKING WITH CRUTCHES

WALKING WITH A WALKER

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Section 2

Brain and Spine Related Disorders

Ch.2 Stroke

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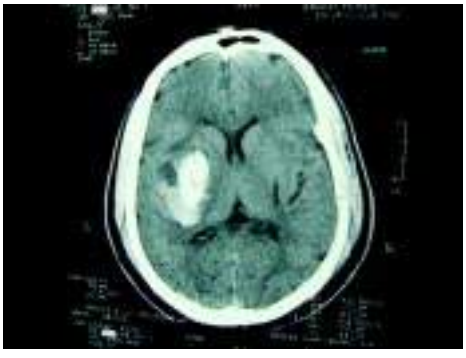
Definition:

Stroke is defined as sudden neurological deficit caused by focal vascular lesion in the brain. The term 'stroke' is synonymous with cerebrovascular accident (CVA) and is purely clinical definition which, according to the World Health Organisation, can be defined as a 'rapidly developed clinical sign of a focal disturbance of cerebral function of presumed vascular origin and of more than 24 hours duration' Included within this definition are most cases of cerebral infarction, cerebral haemorrhage and subarachnoid haemmorhage, but deliberately excluded are those cases in which recovery occurs within 24 hours. These are designated as 'transient ischaemic attacks' (TIA). The vascular lesion could be either hemmorhagic or thromboembolic phenomena (leading to ischaemia) involving the blood vessels supplying various parts of the brain. The extent of neurological involvement may range from mild motor deficit to gross involvement of various function, namely Sensorimotor, perceptual, emotional, behavioural, memory intelligence, speech & language function.

Etiology:

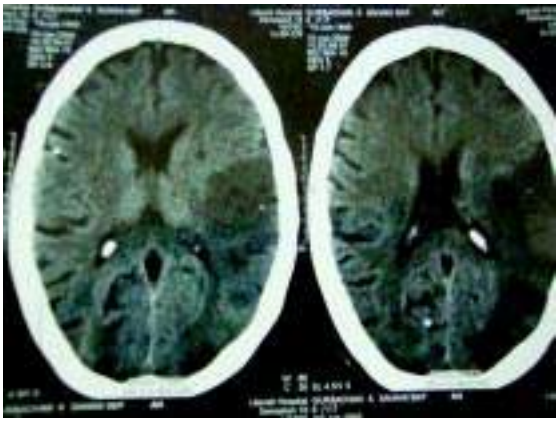
The brain is a unique organ in which functioning of the neurons depend on a continuous blood supply because metabolism is almost exclusively aerobic. Different mechanisms have been found to cause vascular insufficiency to the brain resulting in stroke. However, the most common causes are:

1. Thrombus: This is mainly due to the presence of atherosclerotic plaque in the cerebral arteries as a result of severe platelet adhesion, fibrinous coagulation & decreased fibrinolysis activity.



MRI Brain showing gliotic infarct

2. **Emboli:** These are free flowing bodies in the cerebral blood stream in the form of dislodged thrombus, fats, air, tissue particle, etc which gets trapped at any one point along their course, frequently at the bifurcation of the arteries, & cause occlusion to the cerebral Vessel.
3. **Hemorrhage:** It occurs due to the rupture of the blood vessels in the brain, commonest type being subarachnoid haemmorhage. Following hemorrhage tissue death occurs due to both ischemia & mechanical injury to the brain substance as a result of compression by the clot. Hemorrhage usually occurs either due to hypertension, arteriovenous malformation or due to trauma.



CT Scan Brain showing Hemorrhage

There are some risk factors that can predispose to stroke. The common ones are diabetes, high blood pressure & cardiac disorders. Modifiable risk factors are cigarette smoking, obesity, sedentary lifestyle & excessive alcohol intake.

Pathophysiology of Cerebral Ischemia & consequent infarction:

The two pathophysiological changes leading to cerebral infarction are loss in the supply of oxygen & glucose due to vascular occlusion, & various changes in cellular metabolism resulting in consequent collapse of energy producing processes with disintegration of cell membrane.

Complete occlusion to brain substance causes severe damage to it with a zone of infarction, which is however found to be smaller than the actual area supplied by the involved artery. The margin of this

infarcted zone consists of cells that are alive but metabolically less active. These surrounding areas are termed as ischemic penumbra. These areas are nourished by meningeal collaterals.

The necrotic tissue swells rapidly mainly due to excessive intercellular water content. Also, lack of oxygen is another factor that could contribute to swelling. This vascular lesion to the brain causes release of neurotransmitters like glutamate & aspartate by the ischemic cells, which excites neurons & produces an intracellular influx of Na & Ca leading to irreversible cell damage. Thus recent research attempts at blocking this action of gluatamate & aspartate on nearby cells, which will reduce the secondary involvement of surrounding viable cells.

Cerebral edema begins within few minutes & reaches a maximum by about 4 days, however it mostly disappears by 3 weeks. This edema can increase the intracranial pressure & can even cause contralateral & caudal shift of brain structure.

Neurovascular Syndromes:

Cerebral Blood flow (CBF) is controlled by a number of autoregulatory mechanisms (cerebral) that modulate a constant rate of blood flow through the brain. These mechanisms provide homeostatic balance, counteracting fluctuations in systolic blood pressure while maintaining a normal flow of 50 to 60 ml / 100 gm of brain tissue per minute. The brain has high energy requirements & very little metabolic reserves. Thus, it requires a continuous rich perfusion of blood to deliver oxygen & glucose to the tissues. Cerebral blood flow represents approximately 17 % of available cardiac output.

Depending on the areas of the brain & the arteries affected clinical symptoms vary.

A) Occlusion of proximal Middle Cerebral Artery (MCA)

It produces extensive neurological damage with significant cerebral edema & list of clinical manifestations called MCA Syndrome:

Structures involved	**Neurological deficit**
Motor area of face, arm, and fibres descending from leg area to enter the coronal radiata	Paralysis of contralateral face, arm and leg
Somatosensory area of face, arm and face leg	Sensory impairment over contralateral and leg
Motor (Broca's) area on dominant hemisphere	Motor speech disorder
Central language area and parieto-occipital cortex of the dominant hemisphere	Central aphasia, word deafness, anomia, jargon speech, alexia, agraphia, acalculia, and finger agnosia
Non-dominant parietal lobe	Perceptual disorder like unilateral neglect, anosognosial unawareness of hemiplegic side, apraxia, and spatial disorganization
Occipital lobe	Homonymous hemianopia and loss of conjugate gaze to the opposite side
Parietal lobe	Ataxia of contralateral limbs
Bilateral frontal lobe	Brun's ataxia or apraxia of gait
Supramarginal gyrus or inferior parietal lobe	Loss or impairment of optokinetic nystagmus
Posterior limb of internal capsule and adjacent corona radiata	Pure motor hemiplegia without sensory and visual involvement

B) Clinical Manifestations of Anterior Cerebral Artery Occlusion are as follows:

Structure involved	**Neurological deficit**
Motor leg area involvement of arm area of opposite arm although this is rare	Paralysis of opposite foot and leg involvement
Sensory area of foot and arm	Cortical sensory loss over foot and leg

Bilateral involvement of posteromedial part of superior frontal gyrus	Urinary incontinence
Medial surface of posterior frontal lobe	Contralateral grasp reflex, sucking reflex and gegenhalten (paratonic rigidity), frontal tremor
Severe frontal lobe infarction	Memory loss and behavioral impairments
Supplementary motor area of dominant hemisphere	Aphasia
Corpus callosum	Apraxia and agraphia
Bilateral motor area of leg	Cerebral hemiplegia

C) Clinical Manifestations of Posterior Cerebral Artery Occlusion are as follows:

Structure involved	**Neurological deficit**
Thalamus	Hemianesthesia (contralateral sensory loss) or thalamic sensory syndromes (unpleasant hemibody sensation with spontaneous pain)
Occipital cortex	Homonymous hemianopia, visual agnosia, prosopagnosia
Bilateral occipital cortex involvement	Cortical blindness
Temporal lobe ischaemia	Amnesic syndrome with memory defect
Midbrain	Skew deviation, athetoid posturing, postural tremor, hemiballismus)
Cerebral peduncle	Contralateral hemiplegia
Motor tract between red and vestibular nuclei	Decerebrate attacks

D) Lacunar Syndromes:

Lacunar syndromes are caused by small vessel disease deep in the cerebral white matter (penetrating artery disease). Lacunar syndromes are consistent with specific anatomic sites.

Structure Involved	**Neurological deficit**
Posterior limb of internal capsule, pons & pyramid	Pure motor lacunar stroke
Venterolateral thalamus or thalamocortical projections	Pure sensory lacunar stroke
Base of pons, genu of anterior limb or the internal capsule.	Dysarthria / Clumsy hand syndromes
Pons, genu of internal capsule, coronal radiate or cerebellum.	Ataxic Hemiparesis
Junction of internal capsule & thalamus	Sensory / Motor Stroke
Putamen, global pallidus, subthalamic nucleus.	Dystonia Movements

Synergy Patterns:

Stroke being a central Nervous system disorder, following the acute stage of flaccidity, there is severe spasticity in the affected parts of the body. This leads to abnormal, stereotyped, primitive, mass movement pattern, called Synergy patterns. Synergy can be either flexor or extensor. Synergy normally involves abnormal movement patterns that are not useful for functional activities.

Synergy type	Upper Limbs	Lower Limbs
Flexion Synergy	Shoulder girdle retractor and elevation, shoulder abduction, external rotation, supination flexion of elbow, wrist and finger flexion.	Hip flexion abduction and lateral rotation, knee flexion, dorsiflexion and inversion
Extension Synergy	Shoulder Girdle protaction and depression, Shoulder adduction, internal rotation, elbow extension, pronation, wrist and finger flexion.	Hip extension, adduction, internal rotation, knee extension, ankle plantarflexion, inversion and toe plantarflexion.

The abnormal hemiplegic attitude is due to combination of strongest component of the flexor and extensor synergy in both upper and lower limbs. The strongest components for the upper limb is the flexor synergy and for lower limb is extensor synergy. Hence, a hemiplegic patient has an attitude of shoulder retraction and depression, shoulder adduction and internal rotation, elbow flexion, pronation and wrist and finger flexion. The lower limb adopts an attitude of pelvic rotation, hip extension, adduction and external rotation, knee extension, and ankle plantarflexion and inversion.

Abnormal Synergy patterns of lower limbs leads to abnormal Gait:

Comparison of Normal and Hemiplegic Gait :

Walking is an independent, automatic, symmetrical and economical event. During a normal walking there is both vertical and horizontal displacement of CG in the form of a sinusoidal wave that causes horizontal displacement of 1.7 inch and vertical displacement of 1.8 inch. The gait determinants are as follows:

- Knee flexion at heel strike on loading which lowers the CG.
- Pelvic tilting
- Pelvic Rotation which increases with increase in the heel strike.
- Foot ankle mechanism
- Knee flexion on heel rise or heel off.
- Lateral displacement of pelvis

Hemiplegic stance.

Under normal circumstances walking is an automatic process which requires contribution and integration of various mechanisms, however after the brain damage like in cases of stroke all the automatic adaptation is lacking and the patient tries to compensate for it by putting more voluntary efforts. The phasic action of muscle group seen normally is replaced by abnormal muscular combination characteristic of primitive reflexes. The flexion and extension synergy interferes with the walking and greatly influence the hemiplegic gait.

The abnormality of gait in hemiplegic patient can be compared with normal gait taking into consideration the contribution of the joints for various phases of gait cycle.

Ankle Joint:

Early stance (heel strike to foot flat): In normals when the heel strikes the ground, the angle at the ankle is 90 degrees once the weight is transmitted to the fore foot, the sole is gradually lowered which

is controlled by eccentric contraction of the Dorsiflexors to prevent slapping of foot.

In hemiplegics, the anticipation of the weight bearing excites the extensor synergy which induces plantar flexion instead of dorsiflexion because dorsiflexion is not a part of the extensor synergy. The hemiplegic limb comes down on the entire sole and if the plantarflexion is marked then on the toes. Thus the heel strike is completely missing in hemiplegics. If there is pronounced inversion there is more weight borne by the lateral aspect of the foot and when it is moderate it gets corrected on weight bearing.

Mid stance:

In normal when the sole is firm on the ground the leg starts pivoting forwards above the ankle causing a relative dorsiflexion. This happens in response to the weight shift from hindfoot to the forefoot.The movement of relative dorsiflexion is controlled by eccentric work of the plantarflexors which gradually gets elongated.

In hemiplegics when the calf muscle is stretched by body weight, it stimulates the stretch reflex causing increase in spasticity of plantarflexors and it prevents any further stretch. Thus the forward shift is prevented in hemiplegics.

Late Stance (Unloading to push off) :

In normals towards the end of the stance phase the heel raises from ground and the knee flexes. This is achieved by forceful concentric contraction of plantarflexors for initiating a forward swing.

In hemiplegics, the knee does not flex due to lack of dorsiflexion at the ankle, in the earlier phase and also due to influence of quadriceps spasticity. Sometimes even if it does flex it flexs gradually. Moreover the combination of plantarflexion with knee flexion neither belongs to flexion or extensor synergy, hence is very difficult for a hemiplegic to propel himself. Due to lack of push off in hemiplegics the force required to initiate a swing is very less that causes slowness in walking.

Knee Joint:

Early and midstance:

In normals as heel strikes the ground the knee is extended but as the body weight shifts on to the loading foot, there is 15 degree flexion at the knee under the influence of body weight and it is controlled by the eccentric contraction of the quadriceps to prevent buckling. Immediately afterwards due to concentric work of quadriceps there is extension at the knee.

In hemiplegic patient in flaccid stage there can be buckling due to weight transmission. In stage of spasticity, the initial flexion does not take place and the knee continues to remain in hyperextension throughout. Some patients do have a tendency to buckle initially but immediately rectify by going into hyperextension.

Late stance :

In normals knee flexes with plantarflexion to give momentum to the swing phase.

In Hemiplegics the strong linkage between quadriceps and plantarflexors prevents the swing and the affected limb is moved forward by other compensatory mechanism.

Hip Joint :

Early to midstance:

In normals during weight bearing the abductors of the hip prevent the pelvis from sagging on the opposite side along with trunk flexors. The hip extensors work from heel strike to midtsnace. The short burst of activity reduces just before midstance.

In hemiplegics due to activation of extensor synergy, the adductors contract in place of abductors and hence there is tredelenberg's sign. Moreover when the adductor spasticity is very severe the affected leg is placed very close to normal limb or may even cross over interfering grossly with the forward propulsion.

Swing Phase:

In normals this begins in the late stance and is due to combined effort of muscles, gravitational force and momentum.EMG studies have shown that muscle activity level is minimal. The hip and knee flexors along with Dorsiflexors act to help in adequate ground clearance.

In hemiplegics the extensor synergy does not let go its grip and if it does then it is very gradually so that the body moves forward in slow manner. Hip and knee flexion is absent because of plantarflexion at the ankle and retracted pelvis. Thus, there is inadequate ground clearance. In the absence of forward rotation of the pelvis and hip flexion, the limb is brought forward by circumduction.

Assessment of Stroke Patient :

Detailed medical history, emphasizing on the onset, type of stroke, cause, territory of the brain affected. Following which examination of patient to confirm the extent of involvement and examine the voluntary control in limbs. Thorough assessment of Sensorimotor control is also mandatory. Few assessments scales are used as a standard tool :

STREAM Scoring.

The Brathel Index.

Functional Independence Measure (FIM)

NIH Stroke Scale Work sheet
(www.strokecentre.org)

Rehabilitation after Stroke:

Stoke rehabilitation has experienced tremendous growth in recent years. Much of the impact of stroke on functional independence is due to its effects on motor function. One of the main reasons of the impaired quality of life of these patients is not only related to the actual lesion but also to the result of immobilization and segregation from the community.

The type of rehabilitation and outcomes are determined by a variety of clinical and social factors, including length of time since injury, level of dependency, characteristics of the residual impairment, age of the patient, and resources available.

Is there any scope of remedial training in stroke?

Earlier the adult central nervous system was considered to be rigid and unalterable. Stroke rehabilitation was focused on compensatory approaches for motor impairments and most clinicians believed that therapeutic interventions had little impact on the process of neurological recovery. But research indicates that the adult central nervous system has great neuroplasticity and tremendous potential for reorganization, which has offered new hope to those treating patients with long-term disability and underlies the increasing interest in finding new and more effective ways to maximize this potential. In the recent years, it has become clear that a variety of interventions have the potential to favorably influence motor recovery.

The ICF model provides a conceptual basis and a universal common language for understanding and describing patients' health status, reaching beyond mortality, diseases, and rehabilitation. In the field of neuro-rehabilitation, it facilitates multidisciplinary team communication, to structure the rehabilitation process, for goal setting and assessment, for documentation, and for reporting.

ICF model defines various problems at four levels:

a) pathophysiology of the HEALTH CONDITION,
b) impairments at the BODY FUNCTIONS / STRUCTURE level,
c) disability at the ACTIVITY level, and
d) handicaps at the PARTICIPATION level. Based on the first three levels, Levin et al defined recovery and compensation to explain the differences and allow better understanding about motor recovery after stroke.

ICF Level	Recovery	Compensation
Health Condition: Neuronal Level	Restoring neuronal function in the penumbra (i.e., around the primary lesion) and in the disachisis, which was initially lost due to injury. -restructuring of brain circuits	Neural tissue acquires a function that it did not have prior to injury. -Abnormal activity in alternative brain regions
Body Functions / Structure: Performance Level	Restoring the ability to perform movement as normally as performed before the injury. -Reduction in truck displacement during reaching	Performing old movement in a new manner -Use of excessive shoulder elevation & retraction to lift arm
Activity: -Functional Level	Successful accomplishment of task normally as seen in nondisabled individuals -Opening package of chips using both hands as nondisabled individuals do	Successful completion of task but using alternate strategy. -Opening package of chips with one hand and mouth

The mechanism of recovery

Spontaneous recovery is the norm after stroke. This recovery is typically most rapid & early after a stroke and generally proceeds at a decelerating pace. In case of mild deficits due to small infarcts and transient ischemic attacks, recovery of near normal motor function may result. The mechanism of neurological recovery after stroke involves more than one process.

Recovery of the ischemic penumbra (brain tissue that has been affected by ischemia but not infarcted) and resolution of cerebral oedema early after stroke may account for much of the recovery that occurs within the first few days. The role of disachisis, a loss of brain function due to reduced neural input from remote infarcted tissue, may contribute to deficits that resolve during the early recovery period.

Stroke recovery may continue for months after a stroke. The latter portions of neurological improvement appear to be substantially due to cortical plasticity. The plasticity consists of various mechanisms like denervation supersensitivity, unmasking of silent synapses, regenerative and reactive synaptogenesis.

Basically it is the rewiring of synaptic connections within the surviving brain tissue to restore some degree of motor functioning. Cerebral plasticity has been clearly documented in animal model of cortical stroke and appears active in humans as well.

Animal studies of cerebral function have found changes in cortical function that result from exercise training for motor tasks in both the intact and the damaged brain and in the absence of exercise of the paretic limb, the cortical plasticity provides important evidence of the impact of exercise training on the brain. Thus cortical maps are dynamic However one should not forget to consider the various factors such as the patient's age, severity of the lesion, effects of drugs post injury that will have an impact on the outcome.

Mechanisms to induce recovery (plasticity i.e., at neuronal level) after brain damage:

Till date researchers have found following 10 principles, relevant to rehabilitation outcomes which might augment brain plasticity leading to neuronal recovery. Patients need to be explained, in details all of them to gain maximum benefit :

Principles

1. Use It or Lose It : Failure to drive specific brain functions with volitional movements can lead to functional degradation.
2. Use It and Improve It : Training that drives a specific brain function can lead to an enhancement of that function.
3. Specificity; The nature of the training experience dictates the nature of the plasticity.
4. Repetition Matters: Induction of plasticity requires sufficient repetition.
5. Intensity Matters: Induction of plasticity requires sufficient training intensity.
6. Time Matters: Different forms of plasticity occur at different times during training.Earlier the rehabilitation intervention better the prognosis.
7. Salience Matters: The training experience must be sufficiently salient to induce plasticity.
8. Age Matters: Training - induced plasticity occurs more readily in younger brains.
9. Transference: Plasticity in response to one training experience can enhance the acquisition of similar behaviors.
10. Interference: Plasticity in response to one experience can interfere with the acquisition of other behaviors.

It is becoming clear that the amount and type of physical and mental activity as well as attitude and motivation of the patient has an impact on reorganization of the brain. Hence, application of these new approaches with better understanding of recovery and compensation and mechanism for plasticity will help us in gaining better outcomes after stroke rehabilitation.

The role played by physiotherapists in stroke rehabilitation

Physical therapists specialize in treating disabilities related to motor and sensory impairments. Rehabilitation helps stroke survivors relearn skills that are lost when part of the brain is damaged. For example, these skills can include coordinating leg movements in order to walk or carrying out the steps involved in any complex activity.

Physical therapists help stroke survivors regain the use of stroke-impaired limbs, and establish exercise programs to help people retain their newly learned

skills. Disabled people tend to avoid using impaired limbs, a behavior called learned non-use. However, the repetitive use of impaired limbs encourages brain plasticity and helps reduce disabilities. Rehabilitation teaches survivors new ways of performing tasks to circumvent for any residual disabilities.

There is a strong consensus among rehabilitation experts that the most important element in any rehabilitation program is carefully directed, well-focused, repetitive practice - the same kind of practice used by all people when they learn a new skill, eg: playing the guitar or learning new game.

For some stroke survivors, rehabilitation will be an ongoing process to maintain and refine skills and could involve working with specialists for months or years after the stroke. Over time rehabilitation can be quite remarkable. It does take a team of professionals and the family to accomplish this.

Physiotherapists take a leading role in developing appropriate policies and strategies with other exercise professionals and services to address the transition from rehabilitation to an active lifestyle following stroke.

During this recovery phase, there are many caveats which physical therapists have to address to enhance the progression of recovery. Few of these issues are as follows:

- Paralysis or muscle weakness
- Impaired coordination
- Apraxia (patients loose their ability to plan the steps involved in a complex task and to carry the steps out in the proper sequence, having problems following a set of instructions.)
- Impaired muscle tone
- Loss of feeling
- Speech and language impairments
- Memory and reasoning problems
- Swallowing difficulties
- Psychological trauma (fear, anxiety, frustration, anger, sadness, and a sense of grief, clinical depression
- Problems with vision and visual perception

Goals of Treatment in Stroke patient:

- To normalise muscle tone
- To restore muscle function
- To control compensation strategies
- To maintain muscle length
- To re-educate balance
- To retrain walking and restore mobility
- To maximise functional ability while allowing on-going neuromuscular recovery

To deal with these issues, physical therapists adopt various approaches during the rehabilitation process.

Approaches used in Clinical Setting - Early Rehabilitation

Rehabilitation therapy begins in the acute-care hospital after the patient's medical condition has been stabilized, often within 24-48 hours after the stroke. Goals of treatment in acute stage are :

a) Prevent ignorance or unawareness of hemiplegic side.

b) Decrease the tendency to develop synergy in the chronic stage.

c) Prevention of any joint restriction or stiffness.

d) Prevention of complications due to immobilization like chest complications, deconditioning of bone muscles, etc.

e) Early weight bearing.

f) Pscychological councelling.

g) Education to the family.

The first steps involve promoting independent movement because many patients are paralyzed or seriously weakened. Patients are promoted to change positions frequently while lying in bed and to engage in passive or active range-of-motion exercises to strengthen their affected limbs. Exercises are generally begun with mat activities like rolling, sitting up from lying, in sitting position activities like side shifting are encouraged.Patients progress from sitting up and transferring between the bed and a chair to standing, bearing their own weight, and walking, with or without assistance.

Weight bearing exercises are necessary to promote development of tone in muscles and also to maintain the absorbtion of calcium in the bones.So activities like bridging, supine to elbows, sitting with weight bearing on affected arms and standing should be given as soon as possible with in the limitation of patients general limitation. Oromotor

exercises are also started so as to inhibit drooling and initiate adequate swallowing with out chocking.

Rehabilitation nurses and therapists help patients perform progressively more complex and demanding tasks, such as bathing, dressing, and using a toilet, and they encourage patients to begin using their stroke-impaired limbs while engaging in those tasks. Beginning to reacquire the ability to carry out these basic activities of daily living represents the first stage in a stroke survivor's return to functional independence.

Exercises on vestibular ball are also started to improve balance and also normalize tone of the trunk. Vestibular ball may be used to exercise the upper extremity mainly to achieve control at the proximal joint and facilitate extension of fingers through stimulation of the proprioreceptors at all the joints of upper limbs. Once the patient is comfortable in doing these exercises, then standing and gait training are initiated emphasizing on symmetrical weight bearing on both lower limbs

Fig : 2, 6, 7, 8, 9, 10, 12, 13, 14

Traditional therapeutic exercise program also consists of strengthening, mobilization, compensatory techniques, endurance training (e.g., aerobics). Traditional approaches for improving motor control and coordination: emphasize need of repetition of specific movements for learning, the importance of sensation to the control of movement, and the need to develop basic movements and postures.

Approaches used in Clinical Setting- Late Rehabilitation

Bobath Approach (Neurodevelopmental Therapy)

The bobath approach is widely utilized in rehabilitation following stroke and other neurological conditions. It is a problem solving concept that allows for a variety of strategies flexible enough to be adapted to the strengths and impairments of the individual client rather than a prescribed treatment of exercise.

Traditional Bobath approach was based on the belief that voluntary movements are built on reflexive movement and that treatment must follow the normal developmental sequence. Over the years, the Bobath approach has been modified with many changes and till today alterations continue.

Current Bobath concept has accepted that neural control is not a simple hierarchial function but it is more complex where multiple body systems participate in executing movement that is organized by the specific task and constrained by physical laws and the environment.

Limitation:

There is currently no evidence that Bobath interventions are more or less effective than other therapy approaches (kollen, et al., 2009, Luke, et al., 2004; Paci, 2003).

Hence, NDT approach requires evidence for its effectiveness over other treatment approaches.

Proprioceptive (or peripheral) Neuromuscular Facilitation (PNF)

PNF approach uses spiral and diagonal components of movements rather than the traditional movements in cardinal planes of motion with the goal of facilitating movement patterns that will have more functional relevance than the traditional technique of strengthening individual group muscles.

It uses resistance with the goal of facilitating "irradiation" of muscles to other parts of the body associated with the primary movement.

Constraint Induced Movement Therapy (CIMT)

CIMT is a behavioral approach consisting of different components derived from neuro rehabilitation from basic neuroscience. The treatment for humans after neurological injury has three components:

a) Repetitive, task oriented training of the impaired extremity or function following shaping principles for several hours a day for 10 or 15 consecutive weekdays (depending on the severity of the initial deficit);

b) Constraining patients to use the impaired extremity or function during walking hours over the course of treatment, sometimes by restraining the unimpaired extremity; and

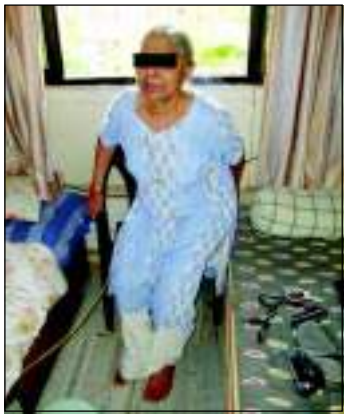
c) Applying a package of behavioral methods designed to transfer gains made in the clinical setting to the real-world.

Brunnstorm Approach (Movement Therapy)

Brunnstrom approach is based on the concept that damaged CNS regressed to phylogenetically older patterns of movements (limb synergies and

Fig : 2, 6, 7, 8, 9, 10, 12, 13, 14

Right Spastic hand with clawing of fingers and synergy pattern.



Stroke patient attempting to transfer from chair to bed.

Bedside shifting for Hemiplegic Patient, teaching independent transfers.

Bridging exs in Hemiplegic patient, to aid trunk strengthening and weight bearing of affected limbs.

Gait Training in Hemiplegic patient with Push knee splint and elbow splint for affected side of the body

Swiss ball activities to strengthen the trunk muscles.

All fours and hip extension of affected limbs to develop voluntary control & break synergy pattern.

primitive reflexes); thus, synergies, primitive reflexes, and other abnormal movements are considered normal processes of recovery before normal patterns of movements are attained. Hence, based on this principle, Brunnstorm approach uses primitive synergistic patterns in training to improve motor control through central facilitation. Treatment includes facilitation of specific synergies through cutaneous / Proprioceptive stimuli.

Motor Relearning Program/Carr and Shepard Approach (Carr et al., 1985)

This approach is based on cognitive motor relearning theory and influenced by Bobath's approach. Goal is for the patient to relearn how to move functionally and how to do problem solving while attempting new tasks.

In addition, this approach teaches general strategies for solving motor problems, instead of emphasizing repetitive performance of a specific movement for improving skills.

Sensorimotor Approach/Rood Approach (Noll, Bender, and nelson, 1996)

This approach is based on modification of muscle tone and voluntary motor activity using cutaneous sensory motor stimulation.

To facilitate muscle tone following cutaneous stimulation techniques are used: quick stretch, brisk icing, fast brushing, tendon tapping, vibration, and joint compression to promote contraction of proximal muscles. Similarly, to inhibit muscle tones, following cutaneous stimulation approaches are used: slow sustained stretch, prolonged icing, etc.

Newer approaches in late rehabilitation

Treadmill mediated gait training

Gait restoration is a major goal in post-stroke neurological rehabilitation.

For this reason, the recovery of independent walking is important in rehabilitation studies. Recently, gait training on a treadmill with body weight support (BWS) has received special attention. It consist of a suspension system to which a patient is connected so that weight shifting, balance, and stepping can be controlled; walking is facilitated by a treadmill. Increasing evidence has suggested that treadmill training in older subjects with hemiparesis improves locomotor capabilities during over-ground walking and motor relearning, because it provides task-oriented practice of walking and active repetitive movement training. It has been suggested that through training, functional movements of locomotors patterns, sensory inputs, and therefore central neuronal circuits, become activated. Hence, even with several studies having shown the feasibility of supported treadmill ambulation training in patients with stroke, whether it is superior to other therapies is still under dispute.

Functional electrical stimulation (FES)

People with hemiparesis often display abnormal gait patterns, such as equinovarus (excessive plantar flexion and inversion) or foot drop (excessive plantar flexion), in which selective control impairments are particularly prominent in the feet. During walking, a person's big toe and outer foot margin rub against the ground, thus putting the person at risk of sustaining sprains and other ankle injuries. To minimize these patterns, electrical stimulation to correct spastic foot drop in hemiplegia was first applied by Liberson and co-workers in 1961.

Functional electrical stimulation (FES) based on the concepts described by Liberson et al, uses electrical signals to activate peripheral nerves and control functional movements. This technique makes use of afferent feedback during contraction, a process that, with a patient's help, may minimize motor relearning during active repetitive movement training.

In routine clinical practice Functional electrical stimulation time (in minutes) can be adjusted according to verbal feedback from the patients during the 20 to 45 minute stimulation period. The patient should be interested to say when they felt fatigue related to dorsiflexion and eversion movements of the stimulated leg. In that situation, FES should be discontinued for 5 minutes and then activated again. As volitional control improves, the FES amplitude can be reduced.

Two main advantages of using FES combined with treadmill training.

- The first advantage was that the patients often report a preference for walking on the treadmill with BWS combined with FES. They report that gait training was more comfortable because it was easier to place their foot during early stance.
- The other advantage was that training with FES decreased the participation of the physical

therapists. Manual assistance can be provided to help the subjects optimize gait quality during training.

Task-oriented training:

No conclusive definition of a task-oriented approach exists in the literature. In the task-oriented approach, movement emerges as an interaction between many systems in the brain and is organized around a goal and constrained by the environment. (Shumway Cook & Woollacott 2001). Task-oriented training includes a wide range of interventions such as treadmill training, walking, training on the ground, bicycling programmes, endurance training and circuit training, sit-to-stand exercises, and reaching tasks for improving balance.

Task oriented training needs to be repetitive and meaningful for the individual. Many interventions in task-oriented training that have proved to be effective are usable in daily nursing practice, such as walking on the ground, moving from sitting to standing from different chairs, and sitting and reaching.

Hence, such training is task and patient focused and not therapist focused. Active use of task-oriented training in the daily nursing care of stroke survivors, will lead to improvements in functional outcomes and overall health-related quality of life.

Robotic-Assisted Therapy (Krebs et al., 1998)

Robotic-Assisted Therapy for stroke rehabilitation started only since 1990s. in general, robotics interfaces with computer software and hardware for improving upper extremity function. Most robots that have been developed for rehabilitation allow several modes of operation including acting as a low friction passive support to the upper extremity, providing active range of motion and/or soft guidance in a movement pattern when the individual is unable to initiate or complete a movement, and offering some resistance to the movement.

Five types of robots have been tested:

1) Assisted Rehabilitation and Measurement (ARM) Guide- made for straight line trajectory reaching (Reinkensmeyer,et al.2000)
2) Mirror Image Movement Enabler (MIME) - provides unilateral or bilateral shoulder/elbow movement (lure, et al., 2002; Kahn, et al., 2006)
3) Bi-Manu-Track- facilitates bilateral passive and active movement of the forearm (Prange et al., 2006)
4) NeReBot- enable movement of the shoulder, elbow, and wrist (Masiero, et al., 2007)
5) MIT-Manus/inMotion2- allows the person to person movement without interference from the robot (although the robot can help perform the motion when the person is unable to initiate or complete the motion)

Robotic-assisted therapy has the potential to facilitate improvements in motor control post stroke, but it is not clear whether this therapy improves outcomes to a greater extent than conventional therapy.

Bilateral Movement Training (Stewart et al., 2006)

A primary reason to perform bilateral arm training is that much of what we do everyday involves the use of both arms and therefore, bilateral re-training is necessary. For example, daily living activities such as bathing, dressing, feeding, toileting, as well as, many other mobility functions such as carrying objects, getting up from bed or chairs, and in driving requires use of both arms and hand. Instrumental activities such as keyboarding, shopping and cooking also rely heavily on bilateral arm use. Hence, bilateral training has potential to improve arm function post-stroke.

Mechanism for Bilateral Movement Training:

1) Bilateral coordinated movement, the upper extremities couple and act nearly as a single unit, reflecting inter-hemispheric coupling at the supplementary motor area (SMA), lateral premotor cortex, the premotor and sensorimotor areas that may facilitate movement in the affected extremity.
2) There is a reduction in intra-cortical inhibition with bilateral movement that may facilitate reorganization in the damaged hemisphere.

Limitation:

Studies of bilateral training have shown that people improve their motor skills with bilateral training after stroke, but the results are completely mixed on whether such training produces greater motor gains than unilateral training.

To design rehabilitation protocols for functional gains with bilateral movements is a challenge.

Mental Imagery Therapy (Page et al., 2006: 2007)

Mental practice, using first-person visual and kinesthetic imagery to practice performance of tasks without activating the muscles involved in physically performing those activities, activates many of the same neural areas as does physical practice. In stroke, mental practice has been paired with either practice of daily common tasks or nonfunctional motor tasks and has been performed from 3-5times per week. Studies have reported a positive effect of adding mental practice to therapy.

Thus, mental practice after stroke appears a promising adjuvant to motor rehabilitation. Although unexplored, the ability of individuals with stroke to produce quality detailed images may influence response to this therapy. How well motor imagery can be accomplished may depend on stroke location in addition to pre-stroke imagery ability.

Virtual reality therapy (Burdea, et al., 1994)

Virtual reality technology provides the capacity to create an environment in which the intensity of feedback and training can be systematically manipulated and enhanced in order to create the most appropriate, individualized motor learning paradigm. The potential benefits of training in virtual reality environment would be the ability to increase the duration, frequency, and intensity of therapy that could be provided to patients by using semi-automated programs. Virtual reality rehabilitation exercises can be made to be engaging, such that the patient feels immersed in the stimulated world, which is extremely important in terms of the patient motivation.

Limitation:

Subjects trained on a motor task in a virtual environment demonstrated the ability to improve performance on the task in that environment, but the learning did not always transfer to the real-world task.

Hence, these protocols should be explored more in order to ascertain the use of virtual reality training as an enhancement to traditional stroke therapy.

Speech Affection in Stroke :

Most of the individuals with stroke present with speech, language and swallowing problems depending on the area in the brain, subcortical areas and the brain stem affected. Usually, middle cerebral artery infarction causes problems in the speech and language areas like Broca's area (frontal lobe) which causes difficulty in expression, Wernicke's area (temporo - parietal lobe) which causes difficulty in comprehension and jargon speech, alexia and agraphia.

Stroke in the verbral and basilar artery affects more of the brain stem areas like the cerebellum, medulla and pons. This leads to dysarthria and dysphagia.

COMPLICATIONS IN THE HEMIPLEGIC PATIENTS :

1. ***Reflex sympathetic Dystrophy*** - many hemiplegic patients develop pain in the upper extremity. Pitting edema, cyanosis, and limitation of extension in the wrist and finger joints, associated with pain, are common and can be attributed to malpositioning and lack of therapy

Treatment :

Paraffin bath may be utilized prior to the passive assistive exercises, provided the paraffin glove is molded with the fingers in maximum flexion. The use of a platform or cockup splint, with the fingers in extension is advised..

2. ***Subluxation of Shoulder:*** is very common,and sling suspension along with weight bearing exercises are recommended.

Heterotopic calcification - extra articular calcification has been described in other neurologic disorders and it may occur in hemiplegic patients. It is seldom encountered in the patient who has been quite active and conscious during the early petiod following the stroke. Repeated insignificant trauma may play a role in the development of this, progressive refractory complication.it may be associated with parietal lobe dysfunction causing the patient to neglect the limb, predisposing it to repeated injury.

The extra-articular calcification occurs about the lesser trochanter of the femur producing a severe adduction flexion contracture of the hip similarly, it can produce a complete extra-articular bony ankylosis of the shoulder joint.The extensor surface of the elbow just above the olecranon process may also be the site of involvement. The elbow is held in an atypical attitude of relative extension rather than flexion as commonly seen in the hemiplegic patient.initially the elbow is quite painful, warm,and slighthly swollen, without any joint effusion. A small amorphous calcific deposit at the

insertion of the triceps tendon, seen in the lateral X-ray film,may be the initianl finding in other patients this complication may not recognized until the acute phase has subsided the calcification may progress to complete ossification

Treatment of this complication is very unsatisfactory, particularly when it is advisable to immobilize the elbow in flexion and avoid all motion until the swelling and pain have subsided has been recommended in traumatic conditions but has not been described in hemiplegic patients.

3. ***Frozen Shoulder / Adhesive Capsulitis:*** The painful shoulder is probably the most frequent and most disturbing complication encountered in the stroke patient. It can be quite disabling since it intereferes with active function, including the performance of simple dressing activities, and may even secondarily immobilize the hand.

The cause of the painful shoulder in the hemiplegic patient has been the subject of consider able discussion basically, several factors may be involved:

1) preexisting degenerative changes in the rotator cuff;
2) repeated small trauma to the periarticular structures, incurred as a result of traction on the shoulder when the patient lies unconscious on the hemiplegic arm or when he tries to move in bed with the arm flaccid at his side;
3) excessive traction on the shoulder capsule, with marked inferior displacement of the humeral herd in a flaccid limb;
4) vigorous stretching of the "tight" spastic shoulder during physical therapy, producing further trauma to the cuff and long head of the biceps tendon;
5) unrecognized trauma such as an impacted fracture of the surgical neck of the humerus, incurred as a result of falling during the onset of the stroke;and
6) contractures that occur in the untreated patient or develop as a result of peripheral nerve lesions and heterotopic calcification.

The treatment of the painful shoulder should be predicated on an accurate diagnosis rather than consist of routine procedures and exercises. Although the use of a sling may be of considerabie value in supporting the flaccid shoulder, it may only aggravate the adduction internal rotation contracture in a painful spastic shoulder. Mobilisation of the head of humerus and amnual therapy gives great results in breaking adhesions between the capsule.

Local infiltration of the tender long head of the biceps tendon with procaine and corticosterois may afford considerable relief when the diagnosis is peritendinitis.in some instances pendular exercises may produce sufficient relaxation to permit effective mobilization. Oral corticosteroids have been used extensively Physical therapy procedures are prescribed to relieve the pain and increase the range of motion.the application of hot packs or cold packs over the deltoid region has become more or less a routine procedure. Frequently it is more desirable to apply the packs over the spastic or contracted pectoral muscles while the shoulder is abducted and in some external rotation rather than adducted and internally rotated. The physical therapist can then intiate the shoulder exercises from a more advantageous position diathermy should be used with considerable caution, particularly in patients having a sensory deficit. Ultrasound therapy has been employed with varying results but may be indicated in treating a persistent localized tendinitis.

The application of a modality to the painful hemiplegic shoulder should always be followed by range-of-motion exercises. The patient must be in a supine position so that the shoulder can be adequately stabilized and the effects of gravity eliminared vigorous stretching should not be devoted to this phase of the treatment to permit the the pectoral and other adductor muscles to relax. Stretching will only elicit a stretch reflex and increase the spasticity in these muscles.

Pulley exercises should not be prescribed unless there is at least 60 to 70 degrees of passive abduction present; otherwise they will only increase the scapulothoracic substitution. The patient must be properly positioned so that the overhead pully is slightly behind him to effectively abduct and externally rotate the shoulder during this exercise.

4. ***Vascular complications.*** Thrombophlebitis in the hemiplegic lower extremity occurs not infrequently during the early convalescent period and may be overlooked. this could explain the persistent edema in the lower leg of some patients after they are started on ambulation activities.a simple elastic compression bandage or stocking applied before the patient gets out of bed is quite adequate. Ample provision must be made for the swelling about the

ankle when fitting the patient for shoes or a short leg brace. The T strap on the brace must be broader than usual and well padded to avoid any constriction about the ankle. The lower leg should be elevated when the patient is sitting in a wheelchair.

An incipient occlusion of the femoral artery may simulate a thrombophlebitis. Evaluation may be difficult, particularly when the patient is aphasic. Vascular changes may have been present prior to the onset of the hemiplegia. Pain in the lower limb is persistent, and there may be evidence of an ischemic neuropathy particularly involving the deep branch of the peroneal nerve, which is most sensitive to arterial insufficiency. The foot drops, if present initially, will become more pronounced, the hyperactive tendon reflexs may be lost, and a segmental sensry deficit involving especially the first web space may signal the onset of a major arterial occlusion.prompt surgical intervention is indicated at this stage rather than waiting for the appearance of the cyanosis of the foot and demarcation of the ischemic skin.

5. ***Seizure.:*** epileptic seizures may occur as a late complication of a cerebral vascular accident.the incidence may be as high as 10%to 15% among those patients who survive beyond the first year. Since the focal signs are localized in the hemiplegic extremities and in the speech mechanism, they often lead to the diagnosis of another stroke. However, these patients recover quite rapidly within the first 24 hours and resume their previous level of activity

6. ***Decubiti / Pressure Sore :*** The older hemiplegic patient with peripheral vascular disease must be carefully observed during the early phase of the illness to prevent pressure sores, particularly over the posterior aspect of both heels. Early mobilization helps in preventing it, by improving blood supply.

7. ***Trauma.;*** the hemiplegic patient is accident prone, especially when ataxia is the predominant peripheral defect. Fracture of the femoral neck is a frequent mishap, particularly in those ataxic patients who walk with a spastic adducted gait and invariably fall on the hemiplegic side. This injury may be associated with an unrecognized impacted fracture of the surgical neck of the humerus.

Careful consideration should be given to the management of the hip fracture. Replacement of the femoral head with prosthesis, followed by immediate ambulation with full weight bearing, has become the accepted procedure in fractures of the femoral neck in the elderly hemiplegic patient. More recently, consideration is being given to simply pinning the fracture after closed reduction followed by immediate progressive weight bearing.

Role of Occupational Therapist in Stroke:

Introduction

Following stroke, patients may be faced with occupational dysfunction. The physical, cognitive and psychosocial capacity to perform routine tasks such as self care, work and leisure activities that are meaningful to them, may be affected and in turn have an impact on the patients occupational identity, health and well being.

The role of the occupational therapist is to enable patients to regain competence, reengage in occupations and redevelop a positive occupational identity1. Occupational therapists are an integral part of the team working in the rehabilitation of patients suffering from stroke. Though stroke is a complex condition, there has been constant advances in the understanding of the condition, assessment and intervention techniques. It is hence vital that the OTs understand the condition itself, know the theoretical basis for intervention and keep themselves abreast of the latest advances in the care and rehabilitation techniques so as to provide the best possible care for the patients.

Theoretical Basis

There are several theoretical constructs, that help to describe and explain occupational function, guide assessments and interventions and predict outcomes. The prominent constructs are:

Conceptual models of practice

The Model of Human Occupation (MOHO)2 considers the complexity of human occupation that behaviour is dynamic and context dependent and that occupations shape a person's self-perception and identity.

This model and its associated tools help occupational therapists to understand the person and focus on an integrative view of human occupation. However, therapists are required to draw on other frameworks to understand and address patient's performance capacity.

The Canadian Model of Occupational Performance

and Engagement (CMOP-E)3 is a social model that considers the spiritual, physical, affective and cognitive components of the person whose self-care, productivity and leisure occupations occur in the context of the physical, institutional, cultural and social environment.

It is a client-centred outcome measure enabling clients to rate importance, performance and satisfaction with self-care, productivity and leisure activities that they 'need to', 'want to' or 'are expected to' do.

Similar but distinctive models emphasising person, environment and occupation are the Person-Environment-Occupation (PEO) Model4 and Person-Environment-Occupational Performance (PEOP) Model5.

The Australian Occupational Performance Model (OPM(A))6 describes eight interactive constructs, including occupational performance, occupational roles, occupational performance areas (self-maintenance, rest, leisure and productivity), components of occupational performance (biomechanical, sensorimotor, cognitive, intra- and interpersonal skills), core elements of performance (mind, body and spirit), the performance environment (sensory, social, physical and social contexts), time and space.

The Perceive Plan Recall and Perform (PRPP) System of Task Analysis7 was developed as one of the assessment tools within this model.

Activities Therapy8 combines psychodynamic, human developmental and behavioural frames of reference. This model suggests that adaptive (compensatory / functional) skills are re / learnt in a developmental sequence to achieve mature functioning and influenced by the environment and their biological composition. In relation to stroke, this model suggests that dysfunction arises when patients regress in their sensory integration, cognitive interaction, group interaction, self-identity and / or sexual identity skills. Rehabilitation is based on sequential relearning of adaptive (compensatory / functional) sub-skills through graded occupation and patient's innate need for mastery.

The Kawa (River) Model9 is an emerging model from an Asian perspective which may address cultural biases of existing models (which value individualism, autonomy and independence) to consider cultural values of collectivism, social hierarchy and interdependence. Appreciation of non-western perspectives enables therapists to be truly client-centred. Thus, in stroke rehabilitation, reduction of impairments may not be as significant as it is in western cultures. Maximising patients' personal attributes and resources, adapting environments and considering interdependence on family and social participation (social inclusion) may be more meaningful than addressing impairments and activity limitations.

Frames of reference

Client-Centred Frame of Reference is a humanistic approach which originated with psychotherapist Carl Rogers and was further developed by occupational therapists in Canada10. Key concepts of the approach include client autonomy and right to informed choice; partnership between client and therapist to work together to negotiate therapy goals and processes; responsibility of the client for his / her own health and ethical responsibility of the therapist to ensure no harm; empowering and enabling clients to achieve their occupational goals; understanding clients individual contexts through respect and listening; accessibility of services to meet clients needs; and respect for diversity.

Biomechanical Frame of Reference is a bottom-up frame of reference, useful for understanding occupational performance capacity in more detail. It considers the anatomy and physiology and mechanics of human movement (kinesiology) focusing on musculoskeletal, neuromuscular and cardiorespiratory systems. Occupational therapy approaches that fit within this frame of reference include graded activities to improve movement strength, endurance, range of motion and sensation, work hardening, energy conservation, ergonomics, assistive devices, splinting and joint protection. Thus approaches to prevent deterioration, restore function or compensate for limitations are significant here. Nevertheless, the primary assessment and outcome for occupational therapy should always be in the context of meaningful occupation.

Rehabilitative Frame of Reference draws on medical, physical and social sciences. It considers rehabilitation as the process of helping patients competently fulfill daily activities and social roles and focuses on therapists teaching, patients learning adaptive (compensatory / functional) methods, assistive equipment and environmental modifications to restore function when underlying impairments cannot be remediated and successful

rehabilitation is dependent on motivation and cognitive skills.

Motor Control Frame of Reference considers the relationship between the central nervous system in relation to motor function and reacquisition of coordinated skilled movement but recognises the influence of other systems (sensory input and cognitive processing),environmental context and learning principles (such as attention, feedback, active participation and goal-directed movement). In comparison to a biomechanical frame of reference, emphasis is on muscle tone, reflexes and movement patterns. Many restorative (remedial) intervention approaches fall under this heading, including Bobath's neurodevelopmental (normal movement) approach, Carr and Shepherd's movement science / motor relearning, Rood, Brunnstrom's Movement Therapy, Proprioceptive Neuromuscular Facilitation, Mental imagery and Constraint-Induced Movement Therapy.

Behavioural Frame of Reference considers learning principles arising from stimulus response models such as Pavlov's classical condition and Skinner's operant conditioning where behavioural responses to stimuli or triggers can be modified through exposure and manipulation of the consequences. This frame of reference is useful for behavior modification such as desensitisation or reduction of anxiety-related symptoms as well as for new learning principles such as repetition and positive feedback.

Cognitive Frame of Reference originated in psychiatry and psychoanalytical theory with the work of Aaron Beck. This frame of reference examines the links between the patients' automatic thinking, their behaviour and emotional response. Cognitive-behavioural therapy (CBT) links the cognitive and behavioural frames of reference together. It utilises a problem-focused approach to explore patients' underlying thoughts, beliefs and physiological responses associated with specific triggers and the consequences of dysfunctional behavioural responses that might maintain these.

Duncan (2006)11 cautions that a cognitive-behavioural frame of reference should be used in conjunction with an occupation-focussed conceptual model of practice to maintain professional role and identity and to enhance the therapeutic potential of the patient-therapist partnership.

Psychodynamic Frame of Reference originated with Sigmund Freud's controversial theories but has been developed to focus on understanding the relationship between past experience and present difficulties. It highlights links between unconscious motivations and emotions which are operationalised through interpersonal interaction, behaviour and occupation. For example, mechanisms such as repression, denial, projection, reaction formation, intellectualisation, rationalisation, regression, sublimation and compensation protect the psyche against anxiety arising from unconscious internal conflict. These internal conflicts and underlying emotions and motivations can be therapeutically explored and symbolically resolved through creative (projective) activities, meaningful occupations, reflection, group work processes and therapeutic relationships to achieve a sense of wellness12.

Cognitive Perceptual Frame of Reference draws on neuroscience and neuropsychology and focuses on the components and interaction of cognitive and perceptual skills that impact on occupational performance. Treatment approaches can be categorised into remedial / bottom-up / skills training or adaptive / top-down / strategy training approaches recognising the brain's capacity but limited potential to repair following brain injury13. A wide range of cognitive and perceptual tools and treatment strategies fall under this umbrella.

In addition to the above theoretical constructs which assist in guiding occupational therapy practice, the emerging theories of neuroplasticity are utilised in current neurological practice. A knowledge of neuroplasticity can assist the occupational therapist in selecting an intervention / approach for the individual patient and will assist in clinical reasoning and justification of the intervention administered.

Neuroplasticity

Recent advances in neuroscience and functional imaging have demonstrated evidence of neuroplasticity - the brain's considerable capacity for neural reorganization14.

Neuronal plasticity after injury occurs as a result of one of two main processes: either the rerouting and subsequent formation of new connections, or neurones substituting function of damaged neurones to enhance the effectiveness of existing connections15. This includes:

– The concepts of synaptic strengthening or potentiation - altering the effectiveness of

synapses (short-term potentiation / long-term potentiation).

– Unmasking of existing silent synapses whose function was previously blocked by inhibitory influences.
– Sprouting of new axon terminals.
– Changes in dendritic organisation.

Neuroplasticity redefined recovery as 'the ability to accomplish the goal in exactly the same way as before the injury'16. The loss of normal function deprives the CNS of the experiential feedback required to drive adaptive reorganisation and may subsequently permit maladaptive reorganisation17. This increased emphasis on remedial approaches for 'true neuroplastic recovery' early in stroke rehabilitation.

Intervention approaches

Despite evidence of neuroplasticity, predicting recovery potentials remains challenging. Some combinations of symptoms will be more amenable to true recovery while other combinations will have limited capacity, requiring an adaptive (compensatory / functional) approach to learn to adapt to activity limitations. Thus, occupational therapists will always need both restorative and adaptive treatment approaches as components of neurorehabilitation.

Restorative approach (remedial approach)

The restorative (remedial) approach relies upon theories of neuroplasticity and the ability of the brain to reorganise itself18. Neurophysiological approaches such as normal movement and motor relearning are included within the restorative (remedial) approach. Here, the therapist provides controlled visual, auditory, vestibular, tactile, proprioceptive and kinaesthetic stimulation to promote normal CNS processing of sensory information. Therefore, normal sensory processing should help the patient make normal perceptual motor responses required for performance of functional tasks. This approach therefore aims to reduce the impairment to subsequently improve activity and participation.

It is implicit within this approach that these tasks are appropriately graded to challenge the patient and encourage the brain to adaptively reorganise itself for successful behaviours.

Adaptive (compensatory/functional) approach

The adaptive (compensatory / functional) approach focuses on repetition of particular skills which are normally associated with activities of daily living (ADL). It is based on the belief that man is a functional animal and his ability to do so is essential for his well-being19.

Adaptive (compensatory / functional) approaches are traditionally used when restoration is unlikely and assumes that certain functions will not recover20. Compensation for loss of function is achieved by changing the activity, environment or patient behaviour by using external assistance, modifying the task or changing the goal or by practice until the task becomes easier in a variety of environments. The advantages of this approach are that it is patient-centred, easy to explain, uses problem solving, meets short-term needs and gives quick results.

Cognitive rehabilitation approach

Cognitive rehabilitation therapy is a systematic and functionally oriented approach to improve cognitive functioning either by restoring cognitive processing skills that are impaired and / or helping the patient learn new ways to compensate for the impairment(s)

Normal movement (Bobath-based approach)

The normal movement approach is the most commonly used restorative approach to physical

Neurorehabilitation. It is also known as Bobath or neurodevelopmental treatment (NDT)

and is based on neurodevelopmental reflex-hierarchical theory that hypothesised spasticity as a product of overactive reflexes. Originally, treatment utilized reflex inhibiting patterns and progressed patients through a neurodevelopmental sequence21. However, Bobath treatment techniques have changed since the last Bobath publication in the 1990s. The current 'Bobath Concept' of normal movement has evolved to incorporate present-day knowledge and a systems theory of motor control, motor learning, neural and muscle plasticity and biomechanics.

The Normal movement approach is a problem-solving or clinical reasoning process based on the assumption that 'too much effort by the patient and overuse of the unaffected side reinforce abnormal tone and movement of the affected side'. Thus, the approach aims to improve disturbances in function, movement and postural control. Key points are used to:

(a) Facilitate and control movements; and
(b) Alter postural tone.

Therapists use experience of movement, repetition, speed, voice, environmental manipulation and feedback.

Proprioceptive neuromuscular facilitation

Proprioceptive neuromuscular facilitation (PNF) as a neuophysiological treatment approach is based on the reflexive relationships between agonist and antagonist muscles which can be manipulated to control the contraction and relaxation of specific muscles groups and thus facilitate normal movement. It also emphasises that 'the brain registers total movement and not individual muscle action'. Assessment considers the relationship between proximal and distal functions, agonists and antagonists, in total patterns of movement observed during functional activities.

Rood approach

This intervention is based on reflexive and hierarchical models of the nervous system. Use of developmental postures and sensory stimulation applied to muscles and joints are used to stimulate a motor response that can either facilitate or inhibit muscle tone in preparation for normal movement. Rood's concept is therefore based upon the concept of correct sensory stimulation being applied to the sensory receptors and eliciting the correct motor reflex which can be utilised in normal movement patterns22.

Movement science

This remedial approach to physical neurorehabilitation is also known as motor relearning programming (MRP), functional and task-oriented approaches, founded by Carr and Shepherd in the 1980s 23. It emphasises the practice of the functional task or action itself as the remedial component promoted by principles of motor learning, including use of instruction, explanation, manual assistance, visual and verbal feedback on performance, reinforcement and contextual practice. Thus, it aims to facilitate motor relearning through use of meaningful activity, feedback and practice. This approach emphasises neuroplasticity and addresses concerns regarding negative effects of compensatory use of the affected side, learned non-use and use of adaptive aids on motor learning by altering task requirements.

Constraint-induced movement therapy approach

Constraint-induced movement therapy (CIMT) is a behavioural approach that involves restraint of the unaffected arm with intensive training of the paretic arm conducted by a clinician using shaping and repetition24. Shaping involves small steps of progressing difficulty and activities are designed to enable patients to carry out parts of a movement sequence; verbal feedback is always positive for any small gains made25.

Bilateral arm training/isokinematic training approach

Bilateral arm training is where the unaffected limb facilitates the affected limb in synergistic coordinated voluntary movements and is recommended for subacute and chronic phases of recovery26. It is based on theories that contralesional activation may activate the lesioned hemisphere or adaptively strengthen ipsilateral pathways to facilitate recovery of the affected limb.

Mental imagery approach

Mental imagery or mental practice has been described as the internal rehearsal of movements without any physical movements27. An essential part of mental imagery is the ability to create clear and powerful images of the task required on demand. The practice must have functional relevance and meaning to the individual to enable more successful visualisation.

With advances in neuroimaging techniques, these mechanisms could be better understood and assist in the selection of specific intervention strategies either in combination with mental practice or in isolation28. Using mental imagery may maintain neuronal activity that would deteriorate without stimulation and prime pathways in readiness to promote motor function.

Electromyographic (bio) feedback

Involves the use of external electrodes applied to muscles and instrumentation to convert electrical potentials from muscles into audio or visual information. This augmented feedback is based on behavioural and motor learning theory where extrinsic feedback is used to improve reacquisition of motor skills. There is some evidence to support its use to augment standard treatment29 but routine use outside of clinical trials is not recommended.

Functional electrical stimulation

Electrostimulation is thought to be beneficial to train and strengthen muscle contractions.

However, results remain inconclusive.

The occupational therapy process

Effective early management is vital in the care and rehabilitation process of a patient with a stroke. Key aspects of early management are accumulating relevant information about patient and completing effective assessments to identify areas of rehabilitation.

Putting all this information together helps to formulate a clear image of the patient, their impairments, skills, goals, motivations, so this information can be used to prepare a clear intervention plan.

Information gathering checklist:

Before assessing the patient, it is important to gather initial information from varied sources such as :

1. Medical history: It is important to document any co morbidities, associated medical problems, current medical or pharmacological management that the patient has, as these may affect their assessment and functional abilities.
2. Social history: The occupational therapist should document the patient's social network, occupation, main roles, financial information, driving status, leisure interests etc.
3. Current physical mobility: to ascertain how the patient can be moved and whether any equipment or assistance from others is required.
4. Premorbid level of functioning
5. Consistent level of functioning post stroke.
6. Information from CT Scan and MRI which indicate the area and extent of damage caused to the brain and the potential impairments that the patient may present with.

Assessment

Based upon their theoretical knowledge (conceptual model and frame of reference) and clinical experience, the OTs select specific assessment and evaluation measures that will identify and measure the factors affecting the functional abilities of the patient. Assessment provide information as a base line for intervention and are useful for planning, intervention and setting short term and long goals.

OT assessment at any stage of recovery will involve a detailed analysis of

I. PERFORMANCE SKILLS
   - Motor
   - Sensory
   - Cognitive
   - Perceptual
   - Psychological
   - Social

II. IMPACT OF ANY IMPAIRMENTS ON A PERSON'S ABILITY TO ENGAGE IN OCCUPATION WITHIN A DEFINITE ENVIRONMENT. (PHYSICAL, SOCIAL AND CULTURAL)

   a. Performance skills

A. Motor

i. Position, Movement, tone of affected extremity

ii. Ability to change or maintain position of trunk and extremities

iii. Presence of pain, shoulder subluxation, associated reactions etc.

iv. Ability to perform mobility in bed.

v. Ability to sit unsupported and maintain balance

vi. Ability to transfer from supine to sit, bed to wheelchair and chair, wheelchair or chair to toilet or commode

vii. Ability to stand on affected leg and reach out in standing while maintaining balance and safety.

viii. Ability to walk and tolerance in different environments. For eg: Indoors on even surfaces, open spaces, uneven surfaces etc.

ix. Ability to perform his or her ADLs for eg: in early stages of recovery, simple ADLs like feeding, grooming, washing and dressing. In later stages, shopping, travelling etc.

Standardised Assessments for Motor Impairments

There are two types of tests that are generally selected to assess motor impairment in stroke.

1. Generic activites of daily living for eg Barthel index30 is a generic ADL scale
2. Specific motor performance tests For eg: Motorocity index31 Rivermead motor assessment32 Nine hole peg test33:

B. Sensory

Sensory impairments like sensory loss are common following stroke. Also there may be problems in interpreting sensory information i.e. perceptual impairments of

i. Visual

ii. Somatosensory

iii. Auditory

iv. Vestibular

v. Olfactory and Gustatory

i. Visual

OT can observe for

a. visual acuity impairments: difficulty in reading, recognizing details or faces, squints while trying to focus.

b. Occulomotor control impairment: Blurred vision, double vision, difficult in focusing, difficulting in reading, watching TV etc.

c. Visual field impairment: Hemianopia and quandrantonopia

ii. Somatosensory:

OT can observe for

Impaired or absent tactile sensation or proprioception e.g. patients who appear to be clumsy and or burn or cut themselves. asstereognosis, e.g. patients who drop objects body image impairments e.g. patients who negect their limbs body scheme impairments like unilateral neglect, right / left discrimation impairment.

Standardised Assessments:

1. Rivermead assessment of somatosensory performance (RASP)34
2. Revised Nottingham sensory assessment (rNSA)35
3. Erasmus MC Modifications to the r NSA36
4. Stereognosis sub test of Chessington OT Neurologic assessment battery (COTNAB)37

iii. Auditory

OT can observe for

a. Hearing loss: e.g. Patients response to sound during conversation or patients name being called.

b. Phantom auditory perception e.g. tinnitus or hallucinations

c. Increased sensitivity to sound

iv. Vestibular

Vestibular Systems along with visual and somatosensory input greatly contribute towards balance38. Dysfunction leads to avoidance of activity and social isolation. OT can observe for:

a. balance reactions and reports of dizziness especially with eyes closed or reduced base of support.

Standardised Assessment: Sensory organization Balance test39

v. Olfactory and Gustatory processing:

Dysfunction in sense of smell and taste following stroke is rare. OT can observe for:

a. Patients response to pungent odours (like cloves, cinnamon, coffee, vanilla and lemon) and different tastes (like sweet, sour, salty and bitter).

Assessment of cognitive functions

Cognition refers to those mental functions which help us to acquire, organise, manipulate and use information and knowledge. It includes all of our 'thinking' processes.

In the acute phase of recovery from a stroke a person may be fatigued and may be dealing with a complex set of recovery issues which can impact on their emotional and psychological status. Testing specific cognitive skills at this stage may not give a true reflection of their abilities and it may be advisable to wait until the rehabilitation phase to do this.

Cognitive functions which may be impaired following a stroke include the following:

– Attention - the ability to focus on specific sensory stimuli and suppress distractions. Attention is required for many other cognitive functions to occur.

– Memory - the ability to retain and recall information.

– Perception - 'making sense of the senses' -

– Language - understanding and expression.

– Praxis - motor planning.

– Executive functions - skills which are needed to plan organise and execute a task.

OT can observe for cognitive dysfunction during assessment of occupational performance during selected daily living activites like grooming or preparing tea / coffee.

Cognitive Standardised assessment

Occupational therapists have a vast array of standardised assessments and screening tools available to them which can be used to contribute to the assessment of cognitive functions. Standardised, impairment-based assessments aim to provide valid and reliable assessments of performance skills, that is, specific cognitive components. These assessments should be used in combination with clinical assessments and reasoning to ascertain extent of impairment impact on occupational performance.

Standardised assessments: ***Standardised observational assessments of performance eg.*****:** MEAMS - Middlesex Elderly Assessment of Mental State40

– COTNAB - Chessington Occupational Therapy Neurological Assessment Battery41

– LOTCA - Lowenstein Occupational Therapy Cognitive Assessment42.

Perceptual assessment

Perceptual problems have been shown to be common following both right and left hemiplegic **Stroke.** These perceptual problems affect the patients' response to rehabilitation and their ability to perform activities of daily living.

Perceptual assessment will clarify whether patients have any perceptual impairments, including the type(s) of perceptual impairment(s) present and their severity. Perceptual ability can be assessed either functionally or by use of standardized assessments.

Functional assessment

Functional assessment of activities of daily living, such as personal or domestic activities, will demonstrate the effect of a mixture of impairments, both physical and cognitive impairments, upon the patient.

Standardised perceptual assessments

There are many standardised perceptual assessments available, some of which are for specific impairments only and some are more general. These can be split into three main categories and examples of each are shown below:

1. Neglect.
2. Spatial.
3. Multi assessments.
    1. Neglect assessments
        i. Baking Tray Test43.
        ii. Balloons Test44.
        iii. Behavioural Inattention Test45.
    2. Spatial assessments
        i. Location Learning Test46.
        ii. Rey Figure Copying Test47.
        iii. Visual Object and Space Perception Battery48.
    3. Multi assessments
        i. Cortical Visual Screening Test49.
        ii. Motor Free Visual Perceptual Battery50.
        iii. Occupational Therapy - Adult Perceptual Screening Test (OT-APST)51.
        iv. Rivermead Perceptual Assessment Battery52.
        v. Repeatable Battery for the Assessment of Neurological Status53.

Many assessments are available from Pearson Assessment (combining The Psychological Corporation, Thames Valley Test Company and Harcourt Assessment). In addition to the above assessments, assessments for provision of adaptive equipment may also be required.

Assessment for adaptive equipment and wheelchairs:

Wheelchair prescription for stroke patients is controversial as some OT's believe that self propelling the wheelchair increases patients muscle tone while using unaffected arm and leg. Indoor powered wheelchair could be considered with patients with severe physical disability and those who have chronic lung and heart conditions. Outdoor Powered wheelchair could be considered for patients with severe long term mobility problems.

While assessing for wheelchair, therapist should take into consideration

- patient's vision, cognition and perception
- patients home environment and local area
- Access to patient's home, type of accommodation, width of doorways, layout of furniture and fittings, door thresholds and floor coverings.
- Raised toilet seats and frames
- Non Slip mats for bathrooms

II. OTHER IMPAIRMENTS IMPACTING ON FUNCTIONAL ABILITY

1) Communication
2) Dysphagia
3) Depression and Anxiety
4) Fatigue
5) Emotional lability

1. Communication:

There are three main problems affecting communication:

– Aphasia / dysphasia-disorder of language which can result in difficulty in understanding and expressing things verbally, reading and writing.

– Dysartria-a speech disorder caused by damage to the nerve supplying the muscle used when speaking. It may involve problems with breath control for speech, voice production controlling whether air is directed orally or nasally during speech and articulation of speech sounds. It can range in severity from mildly slurred speech to inability to produce any intelligible speech.

– Verbal apraxia: a disorder affecting purposeful coordination of muscle movements for speech production. for eg: struggling to achieve correct sound for words or sequence the sounds in the right order.

Dysphagia:

Difficulty in moving a \bolus of food, liquid from the mouth to the stomach without aspirating withour aspirating and involves chewing and tongue movements, preparing food for swallow as well as actual swallow. The OT's can observe for:

– Loss of food or liquid from the mouth or drooling

– Coughing \choking while eating or drinking.

– Pocketing of food inside the mouth.

– Change in voice quality after eating or drinking

– Frequent pneumonias

Depression:

A depressed patient has decreased motivation to participate in assessments and intervention due to being preoccupied with their worries and thoughts54 and it was found that in patients, the severity of impairments in functional activity (ADL) and intellectual function was significantly correlated with the severity of post stroke depression early after stroke. Symptoms of depression include negative thoughts, irrational belief, distortion of reality, self blame low mood, poor appetite and weight loss or increased appetite and weight gain, disturbed sleep, poor concentration and indecisiveness.

Assessments for depression:

The assessments that may be used include the following:

(a) Hospital Anxiety and Depression Scale 55.

(b) Wakefield Depression Inventory56.

(c) Geriatric Depression Scale57.

(d) General Health Questionnaire58.

Anxiety

Patients may have anxieties relating to their stroke, for example, fear of having another stroke, fear of epilepsy, fear regarding their future in terms of home, social, sex, employment, etc.

Hospital Anxiety and Depression Scale59 may be used for assessment.

Fatigue

Fatigue can affect the individual physically, mentally, emotionally or as a mixture of all three60. Fatigue can lead to increased tone, poor performance, reduced motivation, etc. OTs should take care not to overtire the patient. Working with the therapist involves both physical and mental effort on patients part and it can be more effective to change the task than stop and rest when the

patient appears tired. Suitable relaxation techniques can be taught as an alternative to activity or rest. Therapist should try to understand the possible cause of tiredness. (for eg: not sleeping well at night, not able to wake up in the morning because of medication, have an infection or pathological condition, poorly nourished, or bored.) and discuss strategies of coping with it like keeping active, developing interest, using energy conservation techniques etc. The therapist can assist the patient in handling fatigue by providing a varied and challenging programme that is not impossible and not causing stress.

Goal Setting

is a collaborative process between the therapist, patient and their family (where appropriate) involving education and negotiation. Initially, therapists should ascertain patients' and their family's long-term goals or where they see themselves at the end of therapy. Long-term goals are aspirational, giving patients hope and motivation to engage in the therapeutic process. In contrast short-term goals need to be client-centred and collaborative, specific, measurable, achievable, realistic and timely (SMART). Short-term goals form the steps needed to work towards the long-term goal. These can be used to measure outcomes. In addition they allow patients, therapists and team members to maintain direction, motivation, monitor progress and gain insight into how achievable the longterm goal is or whether the long-term goal needs to be adjusted, thus allowing patients to transform their occupational identity to a more realistic sense of self.

Interventions

Rehabilitation enables the patients to meet their goals and ultimately aim to reduce the activity and participation limitation. Rehabilitation has been defined as a problem solving and educational process aimed at maximum recovery by using restorative (remedial) approaches to reduce impairments and adaptive (compensatory / functional) approach to prevent impairments from translating into functional disability. (Activity and participation limitation)

Following a thorough initial assessment, the therapist needs to incorporate the identified problems into an intervention plan. Specific interventions that should be offered (according to need) include:

- The opportunity to practise activities in the most natural (home-like) setting possible.
- Assessment for, provision of and training in the use of equipment and adaptations that increase safe independence.
- Training of family and carers in helping the patient.

The intervention plans should incorporate practicing tasks, particularly personal care tasks. Further, task-specific training should be used to improve activities of daily living and mobility: For eg. Standing up and sitting down.

Management of motor impairments:

Following a thorough assessment, th therapist needs to incorporate the identified problems into an intervention plan. The main aims of OT intervention regarding motor problems are:

1. To promote motor recovery in the most normal or efficient way to increase functional independence by practising graded activities of daily living using a restorative (remedial) approach.
2. To prevent secondary complications such as pain in the shoulder or swelling of the hand.
3. To maximise the patient's independence in activities of daily living by using an adaptive (compensatory / functional) approach, when the restorative (remedial) approach is felt not to be practical or achievable.
4. To train caretakers in safe techniques for handling and carry out risk assessment based on patient's functional level and equipment needs, either in preparation for discharge or as ongoing rehabilitation in the community.

In the early stages of recovery, when movements are restricted by the effects of their stroke, individuals are unlikely to be able to make the postural adjustments required, to maintain a symmetrical posture, without assistance. Proper positioning in bed help recovery by maintaining passive range of movement, allowing the individual to use the control they have and providing normal sensory and proprioceptive input.

Section II.Fig1.1 to Section II.fig 1.6

1. While side lying on the affected side keeping a pillow under the knee helps to reduce any developing hip adductor tone.

Positioning in bed lying on affected side
(Lt hemiplegia pt)

Positioning in bed sidelying on affected side (Rt hemiplegia pt)

Positioning in bed lying on unaffected side
(Lt hemiplegia pt)

Positioning in bed lying on unaffected side
(Rt hemiplegia pt)

Positioning in bed lying back
(Lt hemiplegia pt)

Positioning in bed lying back
(Rt hemiplegia pt)

2. Side lying on the unaffected side restricts use of sound upper extremity. However, this may be the position of choice, (atleast for some time for patients with over active sound side). This position provides proprioceptive feedback about midline, facilitates elongation of trunk on sound side and promotes weight bearing through over active side.
3. Positioning the patient on his /her back can restrict their visual fields and use of upper limbs. However this is a good position to allow pectoral muscles to be stretched. Pillows should be used to prevent affected shoulder and hip falling into retraction.

It is important to consider mattresses when positioning the patient. A firm, supportive surface will provide proprioceptive feedback, enable rolling and promote independence when sitting up. However, pressure areas also need to be monitored. Where hospital pressure care mattresses are used, the patient is likely to require more assistance to turn and sit up.

Encouraging the patient to sit up through side lying promotes head righting, weight transference and a sense of midline. More independence is offered to the early stroke patient in supported sitting and they gain a more normal visual perspective of their environment. There is scope for the unaffected arm to be used in a range of functional activities. The trunk muscles begin to be used actively and the lower limbs begin to form a stable base of support. It is important to note that sitting is not a passive task; the early patient may develop inappropriate muscle activity and 'holding' postures if they do not receive sufficient support from the chair or pillows. Those with sensory loss will require pressure areas to be monitored. Where head control is still lacking, support must be provided.

When the patient begins to gain some active sitting balance and transfers are progressing, positioning on a perching stool allows for more active sitting, improving dynamic control of balance, active extension of trunk and weight-bearing through lower limbs. The upper limbs are freed to perform a greater range of activities. The extra seat height and position of the pelvis in anterior tilt facilitates easier transfers into the standing position. However, perching stools should only be considered for relatively high functioning patients.

Self Care Activities:

All self-care activities should be graded, depending on the patient's level of functioning. The focus is on a restorative (remedial) approach, although it is a common practice to teach some adaptive (compensatory / functional) techniques early, that is, dressing techniques to maximize early independence.

When the patient's sitting balance has improved and the required assistance with transfers is minimal, the occupational therapist can consider sessions involving showering. Showering can be carried out on a shower chair or while seated on a bath board.

If the patient's overall mobility improves, standing should be incorporated into intervention, for example, standing in the shower or at the sink. The therapist should still provide facilitation and prompts if required, to achieve active incorporation of the affected arm and leg.

Dressing can be graded in a similar way to washing/ showering and should be part of the same intervention session.

- Patients early post stroke can be taught one-handed dressing techniques while seated in a wheelchair or armchair; the session would also focus on the patient's sitting balance, trunk control and position and incorporation of the upper limb as appropriate.
- Patients with improving trunk control could be taught dressing techniques while seated on a plinth. This would also involve practising standing with the necessary prompts or facilitation.
- The ultimate goal for dressing would be for the patient to be as independent as possible in the most normal environment, for example, sitting on the bed or standing in the bedroom or bathroom.

Improve flexibility and joint integrity:

Soft tissue or joint mobilization and range of motion exercises are intiated early to maintain joint integrity and mobility and prevent contractures. Positioining strategies are also important in maintaining soft tissue length. Effective positioning of the hemiparetic extremities encourages proper joint alignment while positioning the limb out of typically assumed abnormal postures.

Active and passive ROM with terminal stretch should be performed daily in all motions.

Full extension of elbow wrist and fingers is important as most stroke patients develop tightness

of flexor muscle group. This can be done functionally through sitting, weight bearing on paretic arm with wrist and fingers extended.

Splints: a resting hand splint can also be provided to prevent and manage length associated changes in muscle and connective tissues. Within the literature, there is conflicting evidence and opinions on whether to use splinting as a form of intervention with patients with stroke. The decision whether to splint must be made on sound clinical reasoning. Splints should not be considered when there is active movement that would be restricted if a splint is provided. It is important to teach the family and caregiver the correct manner to don-doff splint and the wearing schedule for effective long term management.

Graded Therapeutic / Remedial Activities:

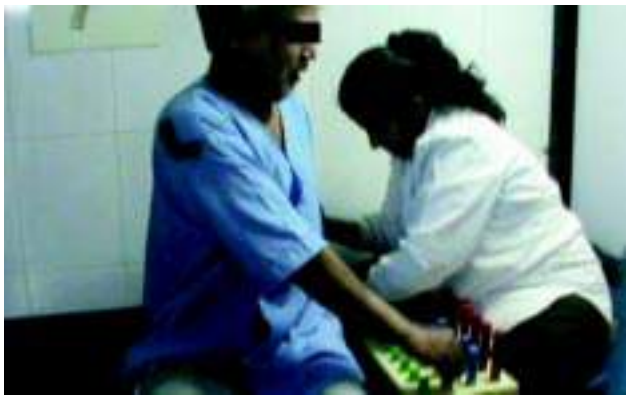
Activities that offer the patient movement experience in a controlled environment can be carried out. The OT sets up the environment and positions the task to gain specific movements and to maximize the effect of the intervention, for e.g.: adjusting the height of table the patient is seated or standing at, and to encourage the patient to reach forward and to pick up a card, carry out peg board activities, writing exercises, computer work etc.

Upper limb reeducation:

Activities to retrain upper extremity postural support, reaching and manipulation are essential elements of training. Two techniques that is bilateral arm training/ iso kinematic training and constrain induced movement training have been shown to be effective in some patients and can be incorporated into occupational therapy plans.

Joint approximation and extended arm weight bearing during early rehab phase promotes proximal stability and counter acts the effects of excessive flexor hypertonus and a dominant flexion synergy.

Weight bearing activites can be performed in sitting, quadruped, modified plantigrade and standing postions, reaching activities with unaffected extremities can be given in these postions using the affected upper extremity as a postural support. The occupational therapist should encourage the patient to use his affected extremity for postural assistance during functional training activities. E.g. Pushing from side lying into sitting. Graded reaching and manipulation activities can also be incorporated in the sessions61.



Facilitating weight bearing and trunk rotation during peg transfers

Avoiding secondary complications:

Pain: Post stroke, shoulder pain is very common and leads to difficulty in participation in functional activities and can cause low mood, altered sleep patterns and therefore have an impact on patients quality of life. Shoulder pain can be caused due to shoulder subluxation abnormal tone either hyper or hypotonicity, proprioceptive impairment and poor handling.

Intervention is aimed at normalizing tone and reducing pain using proper positioning and handling, gentle ROM and gentle stretching, biofeedback and relaxation techniques. The affected arm needs to be positioned properly in all positions lying sitting and standing. Support can be given by using pillows in supine side lying sitting, trays or lapboards for patients using wheelchair or a variety of slings in sitting standing or walking. Use of slings is controversial as it may lead to increased flexor tone and contractures and learned non use. An alternate to the traditional sling is a humeral cuff sling.

Slings should be used for certain upright activities like transfers and gait training and used only for short duration62. As patient recovers and voluntary movements emerge, use of slings should be discontinued.

Family caregivers and hospital staff are instructed and given practices in proper handling and mobilization of upper extremity, avoiding trauma and traction injuries during ROM exercises, transfers and wheelchair activities as well as proper use and care of slings and splints.

Oedema: Post stroke oedema results in restrictive mobility and functional use. Oedema is caused due to ineffective muscle pumping activity combined with poor positioning and effects of gravity. Oedema of the hemiplegic arm is commonly seen

as an isolated hand and wrist swelling but in some cases it can be a part of a more complex "shoulder hand syndrome"63.

Intervention includes a positioning program which includes supported elevation of the arm, passive ranging and light retrograde massage along with functional use of limb whenever possible.

Management of visual and sensory impairments

Intervention for visual processing dysfunction

Occupational therapists have a key role in identifying visual impairment and referring them to appropriate specialist. (Opthalmologist, optometrist or orthoptist) for an in depth assessment and to incorporate the identified impairments into any further interventions as vision has a key role to play in functions.

Following the International Classification of Functioning, Disability and Health model, intervention for visual impairments can be classified under impairment-based intervention, activity engagement and social participation.

Visual acuity and Occulomotor control

Impairment needs referral to the optician and orthoptist for recommended corrective lenses or glasses and occlusion for double vision, prisms etc.

Visual field impairments

This cannot be rectified by intervention and adaptive (compensatory / functional) measures are used to encourage the patient to scan and search within their affected visual field. Occupational therapists can provide simple tasks such as scanning sheets, telephone number copying, environmental searching, etc, to encourage scanning into the affected vision field. An orthoptist may consider use of prisms to compensate, so referral is worth considering.

Activity engagement

Often patients with visual processing impairments compensate very well and are able to adapt to their impairments; however, it is the role of therapy to not only increase their confidence in activity engagement but also their efficiency and reduce the effort required to complete familiar tasks. Intervention could include:

– Engaging the patient in an obstacle course, searching environment for hidden objects, Involving patient in games that require them to switch or track objects such as table tennis.

Therapy within the area of social participation is essential to ensure a patient continues to engage in their life roles and the wider environment while ensuring their safety. For e.g. Developing safe search strategies, for example, crossing the road - identifying the left curb, following it along and scanning from this curb till the patient sees the other curb. This ensures they have seen the whole road before crossing.

Management of somatosensory impairments:

Approximately 50% of stroke patients experience somatosensory impairment64. Functionally, somatosensory processing impairments have significant safety implications, particularly for the detection of protective thermal and pain sensations and for patients have difficulty regulating grasp for effective object manipulation; particularly for fasteners and writing. They are at an increased risk of developing learned non-use as spontaneous use of the affected hand is diminished contributing to further deterioration of motor function and their ability to relearn skilled movements is affected65. These difficulties may in turn impact on all personal domestic and community activities of daily living, sexual and leisure activities and thus participation in life roles66. Somatosensory impairments are significantly related to stroke severity and activity limitations, which negatively impact on motor recovery.

Intervention

Significant tactile, stereognostic and proprioceptive upper limb recovery can occur in the first six months post stroke; however, prognosis for recovery is poorer than in the lower limb67. Stroke severity is the strongest indicator of impairment and recovery and motor performance significantly influences recovery of stereognosis. However, there is no recognisable pattern to recovery68.

O Carey 69 summarises the principles of intervention from successful training programs which include the following:

– Attention to the sensory stimulus:

– Repetitive stimulation with and without vision:

– Use of targeted sensory tasks that are challenging and motivating, with

opportunities for success.

– Anticipation where patient draws on previous experiences of what the stimulus should feel like.

– Focus on the hand.

– Graded progression of tasks for the targeted modality from easy to more difficult discrimination. Variation in stimuli is required for generalisation to novel tasks.

– Intensity of training programme.

– Feedback on accuracy and execution in line with learning theory. Calibration of perceptions with the other hand and visual feedback may also be important.

Restorative (remedial) intervention at impairment level

Shabrun and Hillier70 reviewed the evidence for passive (excludes muscle contraction) and active sensory retraining following stroke and found that active stimulation involving exercises that practice discriminating and localising sensations, stereognosis and proprioception may be beneficial.

Restorative (remedial) intervention during functional tasks at activity and participation level

Encouraging some use of the affected limb during functional tasks to improve sensory feedback may help. The therapists should consider the dynamic nature of sensory input as the central nervous system responds to change and switches off when sensory stimulation is constant. Other techniques which may enhance normal sensory input include the following:

– Bobath principles of facilitation and use of handling to prevent abnormal movements and feed correct sensory input into the sensory system may have indirect effects on sensory improvement during functional tasks.

– Cognitive processing (inattention to tactile stimuli) to utilise attention / information processing strategies.

– Sensory re-education desensitisation - here the central nervous system is overwhelmed and is over-responsive to change, so needs to learn to cope with reduced amounts of input with slower changes.

– Constraint-induced movement therapy

Adaptive (compensatory/functional) intervention

– Use of vision to compensate.

– Enlarged handles to assist grips, universal splints when attention is divided.

– Safety - education, safety awareness, environmental adaptations.

– Pain - distraction, relaxation, pain clinic, Transcutaneous Electrical Nerve Stimulation (TENS), splinting (alignment), taping / Functional Electrical Stimulation (FES) / support for subluxation.

Intervention for auditory dysfunction

Speech and language therapist generally address impairment / restorative (remedial) intervention. Occupational therapists should liaise with these disciplines regarding recommended strategies and enable patients to incorporate these into everyday activities. Occupational therapy intervention should primarily focus on adaptive (compensatory / functional), activity / participation-based intervention.

Intervention for vestibular dysfunction

Occupational therapists should liaise with the physiotherapist regarding recommended strategies and consider application to everyday activities. The occupational therapy intervention should primarily focus on adaptive (compensatory / functional), activity / participation-based strategies including:

– Visual compensation.

– Stabilisation during functional tasks, for example, use of perching stool, chair, stabilizing pelvis against benches / sinks, propping through upper limbs.

– Grading activity demands related to weight transfer and dynamic sitting and standing balance and functional mobility.

– Graded activities requiring independent head / eye movements.

– Functional activities demanding balance that manipulate somatosensory input and visual feedback, such as

– Completing functional tasks with / without shoes on, on different surfaces inside and outside.

– Graded visual feedback starting from downgraded activities where visual feedback

is utilised and somatosensory feedback from the lower limb is stable to high-level retraining where somatosensory feedback is variable (e.g. outdoor surfaces) and vision is occluded.

– Activities which may include reading, tracking objects with / without head movements, coping in dynamic environments in the community, for example, crossing roads, shopping, travelling keeping gaze on fixed target and moving head horizontally and vertically versus moving eyes with head fixed, travelling on buses, in cars, mobility under more challenging conditions, for example, walking, running, moving, standing on moving surface.

– Falls prevention.

Intervention for Olfactory and Gustatory process dysfunction

Recovery of smell and taste have been reported in minor strokes71 and a restorative (remedial) approach could be considered on a theoretical basis. Adaptive (compensatory / functional) approaches should consider functional implications such as safety (e.g. alerting to spoiled food, gas, fire and smoke), nutrition and psychosocial implications.

Management of Cognitive Impairments

Cognitive rehabilitation

The main rehabilitative approaches used by occupational therapists, within cognitive rehabilitation are:

– Remediation (restoration) and.

– Adaptive (compensatory / functional).

Occupational therapists tend to favour a functional approach for the rehabilitation of people with cognitive impairment, including task-specific training and the use of activities which are meaningful and familiar.

Intervention for cognitive dysfunction:

Principles of intervention for the rehabilitation of people with cognitive impairment are:

– Goal orientated: Goals should be meaningful and relevant. Long and short-term goals are set and they should be, as far as possible 'SMART', that is, Specific; Measurable; Achievable (with some challenge); Realistic (within the environment and resources available) and Timescales should be set and there should be a regular review of goals with the patient, family / carer and team72.

– Individualised - a selection of strategies and intervention techniques may be required as people will have individual interests and responses to interventions.

– Educate and include relevant family / carers / friends and significant others - so that they understand the difficulties a person may be having and can assist with the application of strategies and provide support.

– Focus on functional improvement - including a way of measuring this improvement, such as goal attainment and performance measures.

– Include psychological and emotional support - people with cognitive problems can develop anxiety, depression and a sense of loss of control and self-esteem. These should be acknowledged and interventions provided to support management of these problems, such as anxiety management training, relaxation training and medication.

Intervention strategies

– Task-specific training - or functional retraining, stresses the value of the use of specific and relevant functional tasks. Emphasis is placed on task characteristics, in order to support behavioural change73.

– Practise - repetition over time and use of retained capacity assists learning.

– Errorless learning - people with brain injury, including stroke, may not learn from their mistakes so an approach which supports the achievement of a successful outcome by cueing the correct response is more likely to enhance learning. This has been evidenced in studies of people with memory problems74.

– Environmental adaptation - regulation of noise and distractions; clearing environmental clutter; and adaptations such as message boards.

– Compensation and strategy training - external aids and adapted methods - for example, use of memory aids such as pagers, diaries and calendars.

– Prompts and instruction - direct instruction and guided assistance may support relearning of skills.

– Restoration / skills training - Some studies of attention have reported improved skills when specific retraining of basic attention capacity is offered. Retraining tends to be more effective when embedded in a meaningful and functional context, targeting the specific level of attention impairment of the individual75.

Attention Intervention

Attention is required for most other cognitive functions to take place. It is dependent on an adequate degree of arousal and alertness and helps us to process a large amount of information on a daily basis. Attention is commonly affected after stroke, especially in the early stages of recovery. A functional approach using meaningful tasks can be used for intervention and it is suggested that intervention should be focused on training specific functional skills rather than the underlying processes.

As the patients attention improves, the challenge and complexity of the task can be increased to work on higher levels of attention. Repetitive tasks at the tabletop can be used, for example, letter cancellation and word searches, as long as the interventions are providing an appropriate challenge and can be graded as attention improves.

Adaptive (compensatory/functional) strategies

The adaptive strategies recommend that patients should be taught strategies to compensate for their reduced attention. This can be done by

1. providing structure to the patient's day such as using a diary system.
2. Minimise distraction in the patient's environment and
3. ensure the patient has a quiet place they can go to if they become overstimulated as this may manifest in agitated behaviour.
4. Use of prompting to maintain the patient's attention during tasks can be useful (prompts can be verbal or visual).

These techniques should also be taught to families and carers to alleviate the potential emotional stress attention problems can bring to both patient and their carers.

Memory Intervention

Intervention should be individualised, goal orientated and include psychological and emotional support. A restorative (remedial) approach, whilst of some use for those with mild problems, has limited effect for those with severe memory problems. The use of adaptive (compensatory / functional) approaches and assistive devices within the context of functional activities tend to be more successful77. A combination of approaches is recommended and these need to be selected according to where the memory information processing system breaks down.

Some of the strategies are:

1. make the information more meaningful, linking to previous learning or chunking information together.
2. Internal strategies and prompts - for example, use of mnemonics, visual imagery.
3. External strategies -If the patient is unable to store information, use of written and verbal prompts.
4. Compensatory aids - for example, electronic pagers, diaries, notebooks, calendars and computers.
5. work on consolidation through rehearsal and practice.
6. Errorless learning in practice of tasks to minimise performance errors and enhance learning.
7. If the client has 'tip of the tongue' syndrome and has difficulty recalling the information, give graded clues and prompts to elicit effortful but successful recall to facilitate memory.
8. Support from family, carers, colleagues and friends is required to implement strategies and provide prompts and support, no matter which approaches are taken.

Language

Aphasia may occur following a stroke and this may affect a person's understanding of the spoken word, verbal expression, reading and writing. It is usually assessed in more detail by the speech and language therapist; however, joint sessions between the occupational therapist and speech and language therapist may be of benefit to help to ascertain what elements of a person's performance are due to language difficulties or other cognitive problems.

Praxis

The principles of cognitive rehabilitation, 'goal orientated; individualised, educate and include

relevant family / carers / friends and significant others; focus on functional improvement; and include psychological and emotional support' should be used and an adaptive (compensatory / functional) approach is recommended.

Studies have shown that people with apraxia can improve their functional performance despite the lasting presence of apraxia, and that adaptive (compensatory / functional) strategies do not impede the recovery of the impairment78. A proposed 'strategy training' programme for people with apraxia can be carried out to improve performance by teaching internal and external compensatory strategies. As the assessment using ADL observations breaks the activity down into performance stages of initiation, execution and control, intervention targets the relevant stage using instruction, assistance and feedback. If initiation is the problem, instruction is given verbally and if the problems are mild and if the person is experiencing more problems initiating the activity, then the therapist may hand items to them one at a time. If execution is the problem, specific verbal or physical guidance is given and if control is a problem or performance errors are not corrected, then appropriate feedback is given.

Other smaller studies have shown some support for specific interventions:

– 'Activities in context80' - a small 3D movement analysis study suggested that motor performance and kinematic measures improved when the person with apraxia was supplied with the appropriate tools for a task and the correct contextual environment.
– 'Task-specific training81' - it was found that task-specific training could restore independence for trained activities. They also found that skills did 'not generalise' to other tasks and performance was retained only when tasks continued to be 'practised" in daily routines.

Intervention for Executive dysfunction

It is recommended that those with executive dysfunction and activity limitation should be taught adaptive (compensatory / functional) strategies, for example, electronic organisers, written checklists; and that family and other staff should be involved in discussions regarding the impairment and ways of supporting the person. It is recommended that82 'training of formal problem-solving strategies and their application to everyday situations and functional activities' and 'verbal self-instruction, self-questioning and self monitoring could be used to promote internalisation of self-regulation strategies'. Focusing on the functional activities which are limited by specific executive skills are encouraged, for example:

– Goal setting - specific and meaningful for the person. It is helpful to encourage people to set their own goals and then review the outcome to develop insight into their own abilities.
– Planning - activities can be graded to achieve success and gradually improve the difficulty.
– Organisation - a structured routine can be established with support, and responsibility for this gradually handed over.
– Self-initiation and self-direction - external aids such as pagers and alarms can assist with reminders. Taped or written prompts may assist with self-direction.
– Self-inhibition / monitoring and correction - develop strategies to monitor own behavior and make appropriate changes. Feedback and discussion can be used to develop a person's awareness of their own performance.
– Flexible problem solving - alternating scenarios can be presented and practised to develop a strategic approach to generating alternative solutions.

Perceptual Intervention

Intervention of perceptual impairments involves a mixture of restorative (remedial) and adaptive (compensatory / functional) approaches. The restorative (remedial) approach can be generalised, as practice on a particular perceptual task will affect the patient's performance on similar perceptual tasks.

The adaptive (compensatory / functional) approach can be interpreted as repetitive practice of particular tasks, usually activities of daily living, which will make the patient more independent in these particular tasks.

Occupational therapists use functional tasks as an intervention medium. Neglect is the most common perceptual impairment suffered by stroke patients and occurs over several sensory systems; vision, touch and auditory.

General intervention tips for perceptual dysfunction

- Consider the grade of the task; the complexity of the task increases the likelihood of errors.
- Consider the types of prompts, that is, visual, verbal, physical or questioning prompts and pausing before providing a prompt.
- Consider using written or visual instructions.
- Learning can be achieved through repetition and practice.
- Reinforce positive behaviours rather than negative ones.
- Stage components of the task, that is, break down the task and encourage the patient to complete one stage at a time.
- Use verbal rehearsal, that is, encourage the patient to talk through the task before completing it, errors can then be corrected before they are performed.
- Establish patterns and routines.
- Provide consistency in approach.

Specific intervention strategies for perceptual dysfunction

Body scheme

Restorative (remedial) strategies

- Ask the patient to verbally identify parts of the body83.
- Encourage the patient to verbalise positions of parts of the body to improve awareness.
- Provide tactile stimulation, for example, rub a rough cloth on the patient's arm while naming it before placing their arm through a sleeve84.
- Identify parts of the body before washing or dressing them.
- Incorporate bilateral activities that facilitate normal movement and improve body scheme.

Adaptive (compensatory/functional) strategies 85

- Provision of instructions that name parts of the body, such as 'wash your arm'.
- If the patient has functional awareness, provide cues such as 'move the part of the body that you use to hold things' instead of 'move your hand'.

Impaired midline awareness

Restorative (remedial) strategies

- For the patient to become aware of midline by using visual feedback, place a mirror in front of them and instruct the patient to self-correct themselves back to midline.
- In all postural sets, ask the patient to identify the position of their body and describe their relationship to supporting surface86.
- Get the patient to move between postural sets and for them to maintain their balance.

Adaptive (compensatory/functional) strategies

- Place pillows on the overactive side to provide extra supporting surfaces to enhance the patient's feeling of security.
- When seated in a wheelchair place the hospital bed in a high position on the overactive side to enhance feelings of security.
- Teach the patient to use vertical structures within the room such as door or window frames to adjust balance with reference to these markers87.

Unilateral neglect

Restorative strategies

- Use activities that cross midline, for example, personal care activities.
- During activities of daily living sessions place stimuli on the patient's affected side and prompt and encourage them to look over to their affected side. Place necessary items in midline and to their affected side using cues to locate all items and ask patients verbalise the location of items to practise spatial scanning.
- Practise shifting attention from left to right. Cue patients to target stimuli in neglected space to assist attentional shifts.

Move necessary items from midline to their affected side, such as the knife in midline and the butter further into the left side.

- Cancellation tasks such as maze or word searches to practise scanning left to right.
- 2D scanning tasks, that is, paper and pen tasks or more dynamic such as room searches.
- Computer games that require scanning from side to side.

– Tactile stimulation onto the neglected part of the body, using vibration, mildly hot or cold stimuli88.

Adaptive (compensatory/functional) strategies

– Place objects in midline and gradually move objects further into the patients' affected side.

– Approach patients from the midline.

– When reading, anchor the page and draw a red line down the affected side so that the patient becomes aware of how far across the page to start reading89.

– Adapt the environment; remove clutter on the affected side90.

– Encourage the patient to turn their plate round to ensure all the meal is eaten.

– Teach the patient to turn their heads to become more aware of the affected side.

Other intervention approaches

Constraint-induced movement therapy

CIMT attempts to reverse the learnt non-use of the affected arm; however, to be able to use this technique there needs to be enough return of movement that would allow the patient to functionally use their affected hand. CIMT reports to be a useful intervention method for unilateral neglect.

Eye patching

Studies have shown that using glasses that occlude the good (ipsilesional) side of vision in each eye, the patient is forced to direct their gaze to their contralesional side91. Compliance with this technique can be difficult as it is the patient's natural inclination to gaze towards the occluded side.

Prism glasses

There is evidence of the positive effects of prism adaptation92. A 10? rightward horizontal shift of the visual field can be achieved by wearing prism glasses. Using prisms is recommended if the unawareness is severe and persistent.

Visual discrimination

Restorative (remedial) strategies

– Teach the patient to retrieve items following verbal instructions with spatial concepts, for example, 'get the brush on top of the dresser behind the bed'.

– Teach the patent to place different items in different parts of the room.

– Use of tactile kinaesthetic strategies such as guiding the patient to the object.

– Encourage the patient to verbalise the position of parts of the body to improve awareness.

Adaptive (compensatory/functional) strategies

– Organise the objects so that they are in the same place.

– Mark drawers where key items are kept.

– Encourage the patient to feel and describe objects.

– Remove clutter in the environment93.

– Place objects on contrasting surfaces, for example, white soap on a dark coloured cloth.

Visual agnosia

Restorative (remedial) strategies

– Present objects in a straight position rather than other orientation.

– Encourage the patient to recognise differences and similarities between items.

– Start with items that are very different and gradually upgrade to items with subtle variations, for example, shape, size or colour.

– Encourage the patient to verbalise differences, that is, naming objects and differences between objects.

Adaptive (compensatory/functional) strategies

– Teach the patient to consider and think critically.

– Utilise verbal strategies where the patient describes the perceptual and functional characteristics of the object to aid retrieval of the object name.

– Use other senses to identify the object, that is, touch, smell or sound.

– Show the object in a natural context.

– Adding texture or edge orientation to objects may assist into providing cues to identification.

– Use premorbid orientation of objects, that is, did they keep a T-shirt kept in the drawer or on a hanger.

– If categorisation is intact ask the patient to identify which category the object would belong to.

– Provide labels for objects to maximise independence.

Tactile agnosia (stereognosis)

Restorative (remedial) strategies

– Exploratory hand movements for object identification. Explore the object by touching the surfaces and edges of the object, holding the object in the hand to obtain information on its size, shape and weight.

Adaptive (compensatory/functional) strategies

– Education of problems and how these affect function.

– Utilise other senses, that is, vision and touch from the unaffected hand.

– Teach the patient to focus on specific properties of the object.

– Use familiar objects within functional tasks.

– Use objects within context.

General assessment and intervention plan for perceptual impairments

A general plan for the assessment and intervention of perceptual impairments is shown below:

1. Assess perceptual abilities using functional tasks and standardised assessments.
2. Analyse the results and the effect of comprehension, concentration, reasoning (executive function), initiation, memory, anxiety, depression, apraxia, hemianopia / eyesight, inattention, etc.
3. Explain the perceptual problems and their likely effects in everyday life to the patient, their relatives and all staff involved with the patient.
4. Choose the intervention approach to be used, that is, restorative (remedial) or adaptive (compensatory / functional) or both.
5. Decide which intervention strategies to use.
6. Relate intervention to the patients' needs.
7. Remember that not everyone likes games and puzzles.
8. Remember we all learn in different ways.
9. Give mental stimulation.
10. Reassess perceptual and functional abilities.

Equipment

The assessment for and provision of equipment to stroke patients is generally viewed as an adaptive (compensatory / functional) method of reducing limitations. Most equipment is issued following a home assessment visit completed prior to weekend leave and / or discharge. However, some equipment can be used to facilitate normal movement and increase independence within the hospital setting. The pros and cons of timing and type of equipment should be carefully considered in conjunction with the patient, family and multidisciplinary team.

Wheelchairs

Wheelchair: wheelchairs can be prescribed to a stroke patients for two main reasons

– for correct positioning and to improve sitting balance and to increase stimulation during the early management.

– Indoor and outdoor mobility during the rehab stage.

A pressure care cushion should be provided with the wheelchair and monitored throughout the day by nursing staff and therapists.

A correctly fitted wheelchair provides a more active sitting posture, which encourages greater freedom of upper limb movements. When assessing any type of wheelchair on a long-term basis, the home environment and local area in which the patient will be living should always be taken into account. The access to the patient's home, the type of accommodation, the width of all internal / external doorways, the layout of the furniture and other fixtures / fittings, the door thresholds and the floor coverings should be considered for suitability of a wheelchair.

Pressure relief is an important consideration if the person is unable to change position without assistance, but this still needs to provide a stable base. Many specialist chairs are now available that provide additional postural support such as lateral supports, head supports, inclined seats and lap straps (to maintain the hips at 90?).

Toileting

Raised toilet seats and frames (which are safely fixed to the floor) will encourage a patient to move

from sit to stand in a more normal way than pulling on grab rails fixed to a wall.

Bathing/showering

Many stroke patients with independent sitting balance can manage transfers on and off a bath board, but a bath seat is generally too difficult due to the amount of effort involved. This in turn can increase muscle tone and be too strenuous for people with chronic heart and lung conditions and the frail elderly. Non-slip mats should always be provided or purchased to be used in conjunction with bath boards / seats. Chairs or seats that are fixed across the top of the bath for use with a shower or that lower into the bath require less effort for the stroke patient and carer and are much safer for those with poor sitting balance. Step-in shower cubicles have limited space for small stools or seats fixed to the wall and are therefore only accessible to the more mobile stroke patient who can wash themselves independently whilst standing or sitting on a stool.

Dressing

Clothing styles may change initially in the early stages of learning a dressing technique, the patient may wear more leisure wear which is easy to slip on until they become proficient in dressing techniques or their motor / cognitive problems improve, allowing the individual to dress in their desired style of clothing. Any change of clothing style must be carefully discussed with the patient in order to maintain the individual's autonomy and self-image.

Even in the early phase of recovery, occupational therapists can teach patients adaptive (compensatory / functional) strategies for functional tasks that will improve quality of life and that are considered not to have a detrimental effect on motor recovery. Adaptations such as elastic shoe laces or Velcro shoes / trainers are often useful, or teaching the patient a one-handed method of tying shoe lace is feasible.

Meal preparation

Some kitchen equipment such as large-handled utensils or cutlery issued by occupational therapists could be used by patients with some return of hand function to encourage further improvement or facilitate more normal movement. These could be used during meal preparation sessions in hospital or at home. Many other pieces of equipment are

Upper Body dressing activity

designed for one-handed use or to make heavy tasks lighter.

Fatigue is a major factor to consider when preparing a meal. The layout of the kitchen and its existing equipment or appliances can be looked at during a home assessment visit. Some portable items could be moved closer together in order to conserve the patient's energy. A perching stool could also help reduce fatigue.

Eating

During the acute stage, good positioning whilst eating will assist safer feeding and swallowing. Plates that retain heat will keep food warmer for a slow eater. Plateguards and large-handled mugs with lids reduce the risk of spillage. Dycem mats will keep plates in place. Large-handled cutlery could be used with the affected hand to encourage further return of movement. At a later stage, the one-handed patient may require a rocker knife or a fork with a serrated edge for cutting.

Evaluation of occupational therapy effectiveness 'is an ethical and professional imperative'. Standardised assessments are used in clinical practice to identify and quantify problem areas. These assessments can also be used as outcome measures. There are two types of tests that are usually selected, generic activities of daily living and specific motor performance tests. The results of such assessments should be useful for planning intervention and setting goals. The Barthel Index is generally recommended as a generic activity of daily living scale while the Motricity Index, the Rivermead Motor Assessment and the 9 Hole Peg Test as specific motor performance tests.

At the patient level, ongoing evaluation enables the appropriateness of intervention to be monitored, allowing opportunties for adjustment and to determine if therapy has been successful.

Follow-up

All patients with residual impairments after the initial period of rehabilitation should be offered a formal 6-monthly review and appropriate treatment recommendations. Particular areas to focus on during follow-up appointments include any changes in impairment (e.g. cognition), activity (e.g. activities of daily living), participation (e.g. fatigue), environmental and personal (e.g. mood, stress resistance, social support, quality of life) circumstances since discharge.

Young Stroke Patients

Younger stroke patients may have to make massive changes to their lives and this can take a long time to come to terms with: changes to jobs, loss of a car, changes to hobbies, breakdowns in personal relationships, depression, and reduction in wealth as well as any physical changes. Rehabilitation of younger stroke patients will include a wide multidisciplinary team and may need to be extended over many months.

As well as the rehabilitation programme, occupational therapists can help younger people with stroke access this website whilst in hospital, they can introduce them to other younger people and ensure they are provided with vocational rehabilitation.

Lifestyle and long-term management

The effects of having a stroke are often ongoing and therefore once patients are over the immediate rehabilitation phase of approximately six months, they start looking forward to resuming their lives as much as possible and making changes to accommodate their new limitations94. It is generally recommended that people with ongoing limitations after the initial rehabilitation phase should be offered a six monthly review and be provided with further rehabilitation if clear goals are identified. These review sessions may highlight areas that may have been previously ignored by the patient such as social participation, leisure, returning to work, transport and social support. the main emphasis at this point is for patients to start to plan, enquire, lead and be proactive about their lives, about the information they need and how they are going to undertake activities without the assistance of health professionals.

Patients may have changed their lives completely; they may have been given specific advice on diet such as a low-salt, low-fat, low-cholesterol to prevent a recurrent stroke; they may have given up work, driving and lost contact with friends and family. At about this time, contact with most rehabilitation services will have ceased. These life-changing actions happen over time and people often need support and reassurance to continue to explore new avenues.

Social participation

People who have had a stroke often feel dependent on others, lack knowledge and with one-third feeling socially isolated and one-quarter being depressed it is not surprising to find that patients have often lost the social networks they had before the stroke. Occupational therapists can initiate and enable people to start participating in society but over time people do not want to be always associated with professionals and they move to groups more normally associated with the

community such as work, family, visiting friends, voluntary groups such as social clubs, luncheon clubs, hobbies and religious activities. There are vast benefits to improving social participation on quality of life, personal choice, dignity and ultimately improving health by easier access to health centres, better physical health through exercise, reduction in falls through improved muscle strength and balance.

Leisure rehabilitation

It is known that participation in leisure decreases after stroke, even for patients with a good physical recovery95. The significance of this reduction lies in the fact that satisfactory leisure is related to life satisfaction. Consequently, such a decrease in leisure activity may reflect a decrease in quality of life.

Getting out of the house and transport

Many leisure and social activities require transport either to get to an event or to get supplies. However, even just getting out of the house 'for the sake of it' is an important rehabilitation target after stroke as it improves psychological and functional outcomes. Half of all people with stroke do not get out of the house as much as they would like even after rehabilitation.

An occupational therapy outdoor mobility rehabilitation intervention found that people who had received the intervention, which involved the patient practicing outside with a therapist, were

twice as likely to go out afterwards as those who had received the routine rehabilitation programme.

Vocational rehabilitation

it is important that people are able to undertake education, employment, re-training and voluntary work to improve their quality of life, fulfill a role in society, avoid low self-esteem and depression. There is evidence that this can be achieved by offering vocational rehabilitation (VR) to people after a stroke96.

Both paid and unpaid work fulfils a diversity of needs for the individual, and some of the many benefits of work are listed below:

- Increased self-esteem.
- Maintenance of routines and habits.
- Participation in a productive activity.
- Involvement in a socially accepted role that provides value to the community.
- Having 'a reason to get up in the morning'.
- Challenging someone to expand their horizons.

Occupational therapists can still assist the patient in establishing realistic goals and expectations towards work and produce a written joint plan of action.

Resuming sexual activity

As rehabilitation of stroke patients constantly improves, many new techniques develop but some activities are often neglected because of lack of research, lack or rehabilitation knowledge or because patients themselves do not wish to raise the subject. One such area is a return to their normal sexual activity. Generally, this is not routinely discussed with patients as part of their rehabilitation, although for many, it plays an important role in their life and discussion should be included in the assessment process.

Research has observed that after stroke there is a decline in sexuality in both genders, and partner dissatisfaction is high. There are many reasons for this decline: physical changes in the brain that may reduce the sexual urge; physical changes to the body that may make it difficult to move; psychological changes that make people not want to have sex. Stroke can cause physical limitations that influence body positioning and movement during sex which can lead to a reduction in sexual activity, misery and frustration. Other physical changes such as incontinence, drooling, emotional liability may be off-putting to partners and again lead to a reduction in sexual activity.

As well as physical difficulties, psychosocial impairments and depression may affect the will to engage in sexual relationships; however, these may not be recorded till late after stroke as patients may be unwilling to talk about sexual difficulties. Mood disorders, such as depression and anxiety are commonly observed after a stroke, frequently affecting sexual relationships and sexual function and conversely, sexual dysfunction may lead to depression.

After a stroke, generally both patients and partners want to know whether resuming sexual activity will cause another stroke or epileptic fit and do not know whom to approach for advice. Although there is only a low risk of stroke from sexual excitement, people are unwilling to have sex as they believe it may cause another stroke. Sexual activity is a subject which is important to both patients and partners, and should be included in stroke rehabilitation.

Occupational therapists are often approached to discuss sexual activity whilst they are dealing with other personal activities of daily living. This chance to contribute to part of patient's lives often ignored, should be embraced with knowledge and understanding.

Patients and partners need to know that returning to their normal sexual activity is considered routinely as an aspect of stroke rehabilitation. They also need to know there is the opportunity to discuss their sexual activity either alone or in couples, at a time which is appropriate to their needs.

Outcome measures

The consequences of stroke on an individual's functioning are often complex and varied in nature. Stroke not only effects neurological functioning, for example, movement / speech but may also lead to a dependence in activities of daily living and cognitive and perceptual difficulties. Outcome measures are tools or instruments used to quantify the change in a patient due to an intervention, and allow for the evaluation of the effects of interventions to be established.

Therefore, the measuring of outcomes is an essential component in determining therapeutic

effectiveness and therefore is central to the provision of EBP. Additionally outcome measures can be categorised according to the three categories; body function / structure, activity and participation.

Body functions and structures Activities Participation

- Beck Depression Inventory
- Action Research Arm Test
- Behavioural Inattention Test
- Barthel Index
- Performance Measure
- Canadian Neurological Scale
- Berg Balance Scale
- London Handicap Scale
- Clock Drawing Test
- Box and Block Test
- Medical Outcomes Study
- Fugl-Meyer

Assessment

- Chedoke McMaster Stroke Assessment Scale
- Nottingham Health Profile
- General Health
- Clinical Outcome

Variables Scale

- Reintegration to Normal Living Index
- Geriatric Depression Scale
- Functional Ambulation Categories
- Stroke Adapted Sickness Impact Profile
- Hospital Anxiety and Depression Scale
- Functional Independence

Measure

- Stroke Impact Scale
- Mini Mental State Examination
- Frenchay Activities Index
- Stroke Specific Quality of Life
- Modified Ashworth Scale
- Motor Assessment Scale
- Motor Free Visual Perception Test
- Nine-hole Peg Test
- National Institutes of Health Stroke Scale
- Rankin Handicap Scale
- Orpington Prognostic Scale
- Rivermead Mobility Scale
- Timed Up and Go

Other considerations

Standardised and non-standardised outcome measures - Standardised outcome measures have specified, standardised procedures for completion and scoring. These measures will usually have been tested for validity and reliability to ensure consistency in application of the measure. Additionally many measures will have been normatively standardised for scoring, over large populations. This means that therapists can compare patients' scores against a normal range and to other patients with similar conditions. Non-standardised outcome measures have not been subjected to the same rigorous testing procedure and are therefore often of poor quality, and generalisation of scores from non-standardised measures is problematic.

Use of outcome measures - Outcome measures can be used for a number of reasons.

Outcome measures can be used to evaluate:

– Improvement.
– Maintenance, for example, of function.
– Reduction, for example, of pain or discomfort.
– Prevention, for example, of disability or discomfort.
– Development / maturation.
– Recovery, for example, of function.
– Delay, for example, in rate of deterioration.

Outcome measures should be routinely used by occupational therapists not only to evaluate their own practices but also to inform and motivate patients and carers and provide feedback to the multidisciplinary team.

Psychological Aspect:

Psychology is the study of behaviour and the main goals of psychological intervention post stroke is re-gaining or improving the functioning in aspects

like: cognition, socialization, regaining sexual life, vocational guidance, and psycho-educating the caregivers and to improve the overall quality of life.

Psychological Assessments:

Neuropsychological assessment following stroke is undertaken for the following reasons:

- To provide prognostic information, in addition to monitor the rate and extent of natural recovery from stroke or improvement under therapy.
- To provide a baseline profile of cognitive functions against which to assess subsequent natural recovery and to judge the outcome of any intervention.
- To provide a basis on which to plan any cognitive remediation interventions.
- To provide a source of advice regarding suitable placement or long-term care following termination of active rehabilitation. This guidance can be used to help care staff and relatives gain better understanding of the stroke patients cognitive deficits and competencies and therefore to behave appropriately.

Assessments Post-Stroke:

- Mini Mental State Examination [112] to assess the cognitive functioning.
- Neurobehavioral Cognition Status Exam (NCSE) to assess the cognitive and behavioural functioning.
- The Clock Drawing Test to assess the cognitive functioning.
- The Montreal Cognitive Assessment [113] to assess the cognitive functioning.
- Hamilton's Anxiety Rating Scale to assess the level of anxiety.
- Beck's Depression Inventory to assess the presence of depression.

Cognitive Dysfunction Post Stroke:

Every stroke is unique and the cognitive effects and problems post stroke also differ depending on, where the stroke injured the brain, and your overall health. Stroke can cause vascular dementia, a greater decline in thinking abilities. Some experts believe that 10- 20% of patients over age 65 with dementia have vascular dementia. This makes it second only to Alzheimer's disease as a leading cause of dementia. The most common types of cognitive deficits arising from stroke are disturbances of attention, language syntax, delayed recall and executive dysfunction affecting the ability to analyze, interpret, plan, organize, and execute complex information [113, 114,115]

Memory Loss: Memory losses post stroke is common and there are many ways as to how their memory can be affected:

- Verbal Memory: Patients may have difficulty remembering names, stories and may have difficulty in forming sentences.
- Visual Memory: Patients have difficulty in remembering names, faces, shapes, directions and things that they see around.
- Recent Memory or Short term Memory loss: Patients may have have trouble remembering instructions, conversations, and recent events. They may also have trouble remembering upcoming events such as doctor's appointments, difficulty learning new things and new information or skills.
- Remote memory: Patients may have trouble recalling or retrieving past information or skills.

Reduced Attention Span:

Post stroke the patients may have difficulty in maintaining their attention span. When their attention span is reduced, these patients may be unable to attend to a specific task for more than a few minutes, at a time. Another effect of a reduced attention span is that survivors may have trouble dividing their attention between more than one task (such as reading the mail while listening to the news), or going back and forth between two or more different activities.

Reduced Problem Solving Skills

Difficulties with problem-solving skills can mean that a stroke survivor has trouble recognizing and solving common everyday problems, such as a car that will not start or a pot that is boiling over on the stove. Individuals with reduced problem-solving skills often require supervision at home in order to avoid accidents or injury. Stroke survivors with impaired problem-solving skills may also have difficulty with math and maintaining their personal finances.

Difficulty with Social Communication

Some survivors of right hemisphere strokes also have difficulty with social communication. This means that they may say inappropriate things to family, friends, or strangers. They may also have trouble understanding jokes and humour. Also, they may not be able to form statements and many times patient's speech is restricted to few words.

Post Stroke Depression (PSD):

Depression is the most under diagnosed and untreated squeal of stroke in spite of it being the most common complication of stroke. The reported prevalence of stroke widely ranges from 16 percent to 62 percent, with more reliable studies reporting 25 percent to 40 percent of stroke patients meeting the Diagnostic Stastical Manual, Fourth Edition, Text Revision criteria of minor depression. Most cases develop soon after the stroke persists from 6 to 12 months, with the frequency decreasing as time progresses, although a significant minority of patients with depression for 24 months or longer after the initial stroke.

The hindrances caused in diagnosing a stroke patient with depression is the overlap that exists between somatic manifestations of stroke such as alterations in sleep patter, decreased appetite, and feelings of sluggishness or appetite. Language and cognitive impairments, emotional liability and attention disturbances further complicate the diagnostic picture. Post stroke depression has been reported in patients who have a history of depression pre stroke, lack of social support and who have resulting severe disability including cognitive dysfunction, dysphasia or altered visual perception.

There are a few factors that raise the likelihood and severity of depression following a stroke:

- The location of the brain lesion
- Previous or family history of depression
- Pre - stroke social functioning

Stroke survivors, who suffer from major depressive disorder, may be less compliant with rehabilitation, more irritable and may also undergo personality changes.

Emotional Effects of Stroke:

Someone who suffers from stroke his/ her life suddenly changes from an able bodied person to someone who is unable to move one side of the body or who has difficulty communicating or who suffers from frequent emotional outbursts. Medical treatments have although reduced both the severity and frequency of stroke and on the other hand the rehabilitative services have made it easier to adapt to the changes stroke effects such as physical disabilities and problems with thinking processes. However, there are significant changes that take place in the person's relationship, in their feelings and in their independence. In particular the relationship between the strokes, suffers and the caregivers may unalterably change especially id it becomes clear that recovery may be limited.

Common emotional effects of post stroke sufferers:

- Anxiety
- Loss of independence
- Emotional Liability
- Depression
- Confusion
- Apathy
- Memory loss
- Irritability

Stroke suffers have different symptoms depending on the area of brain injury, that is they may experience both physiologically induced emotions and psychological emotions that are a result of brain injury and changes that has brought about it. People who suffer from stroke on the right side of the brain tend to experience memory problems, time disorientation and emotional highs and lows. They may also be less able to use abstract thinking, may talk excessively, may have a shortened attention span, and may be more impulsive than before the stroke. On the other hand, people who have had a stroke on the left side of their brain tend to find their language and reading abilities more seriously affected. They will also be more cautious and easily frustrated. As well, left brain injuries are most often associated with depression. Whether this is a result of the location of the brain injury, or whether it is a consequence of the difficulties in communication that these people experience, is not known.

Personality Changes Post Stroke:

Changes in personality following a Stroke may be very disturbing to the family. Personality is the unique combination of an individual's thoughts, feelings and reactions toward themselves, others and their environment. Post stroke the individual

may not be the same as he/ she was pre stroke. The way in which they think, feel and react may be altered. Family and relatives need to understand the new and puzzling changes. Problems and activities once tackled easily may be difficult or impossible, while other tasks are unaffected. People may become confused, self-centred, uncooperative and irritable, and may have rapid changes in mood. They may not be able to adjust easily to anything new and may become anxious, annoyed or tearful over seemingly small matters.

Loss of motivation:

Decreased or absent motivation and impaired ability to initiate activity may be experienced by stroke. Generally, referred to as adynamia and is a direct result of brain trauma. Stroke suffers may do little beyond self-care tasks and may seem lazy. The patient may appear to have a lack of initiative in activities. With extra guidance and prompting after time they will have a better view in engaging in familiar activities followed by trying in new activities.

Sexual Desires Post Stroke:

Recovery from stroke would take a long time and would lead through many stages. If being sexually active was important to a person before stroke then there would be the same need post stroke. A few hindrances to having an active sexual life post stroke would be:

- **Fear:** The common fear following a stroke is that having sex will bring on another stroke. There is no reason why after a couple of weeks you cannot begin to have sex if you feel ready to do so. Medical evidence supports this. If you still feel unsure about having sex then arrange to speak to your own doctor.
- **Emotional changes:** Both men and women experience similar emotional problems after a stroke. How you feel about yourself and how you perceive others feel towards you, can lead to you losing confidence in yourself. It can take time to adjust to and come to terms with the changes in your life after a stroke and many people experience anxiety and depression as a result. This can have a knock on affect on your desire for sex.
- **Change in relationships:** Post stroke there could be a sexual awkwardness that a patient would experience. This could be especially true if the patient needs assistance in maintaining personal hygiene. This could affect the relationship as the partner would be deprived of his/ her sexual needs.

Caregivers:

Common Emotional side-effects of Stroke Caregivers:

- Anger
- Guilt
- Doubt
- Impatience
- Helplessness
- Resentment
- Depression and anxiety

There are many good emotions that will result from caregiving, but negative emotions about your situation are also normal. Unfortunately, negative emotions toward your care recipient are normal as well. Rather than beating yourself up when the frustration overwhelms you, acknowledge that these feelings are part of caregiving - and they don't make you any less of a caregiver. Also recognize that it is in everyone's best interest for you to take regular breaks. To be the best caregiver, you need to rejuvenate yourself from time to time. And your care recipient needs breaks from you too!

Even if you are the primary caregiver, your family and friends need to understand that their support is needed - and part of that means supporting you. Your ability to care for your loved one is dependent upon your well-being. You need many of the same things that your care recipient needs:

- Affection and love
- Emotional support
- 'Away' time
- Exercise and a healthy diet
- Proper rest
- Relaxation time

How do you manage all this? Creating a care plan, can help.

Psychological Intervention:

Pharmacological Treatment for Depression:

Pharmacological treatment for post stroke depression is still questionable as the potential side effects of antidepressant medications is quiet

evident. If a person is only mildly or moderately depressed, psychological treatment alone may be effective. However, if depression is severe or persists, medication is often necessary as well. Medical research shows that depression is often associated with an imbalance of certain chemicals in the brain. Antidepressants can help rebalance these chemicals. Antidepressant medication can take seven to 21 days to work effectively and should not be started or stopped without medical advice.

Psychological Counseling:

Patients with chronic physical illness and co-morbid depression may have a very negative view of their situation. It could also lead to feeling helpless and hopeless about the situation. Counseling would help the patients identify and change these negative ways of thinking and find ways to find hope and meaning to their lives with the existing level of functioning. Counseling session would involve usage of various psychological therapies depending on the individual patients needs.

Cognitive Rehabilitation:

Aim of Cognitive Rehabilitation:

The chief aim of cognitive rehabilitation is to help the stroke survivor to achieve the most independent level of functioning as possible. The objectives of cognitive rehab also vary from person to person. Some stroke victims would want to have the ability to express needs verbally in simple terms while others would be happy with just pointing to pictures. But majority of stroke survivors want to improve their capability of defining words and to identify the cause and effect of relationships.

When a stroke patient undergoes cognitive rehabilitation program the first 2 steps to rehabilitation are:

- To restore functions which, can be restored
- To teach new compensatory patterns of cognitive activities

After which different cognitive methods are used to rehabilitate the patient like:

- Solving arithmetic problems
- Solving logical puzzles
- Concentrating on writing and reading skills
- Playing memory games

Recovery of the brain follows a systematic pattern of brain development. Just like in infants, a stroke damaged brain must first learn or re-learn gross or large-scale systems before fine and complex systems. The gross cognitive systems include attention, focus and perceptual skills. And when these are redeveloped, complex intellectual activity will follow through.

When to start cognitive therapy:

Cognitive therapy should start when the patient is still in the hospital as time is very crucial after a stroke attacks. Early cognitive therapy will focus on increasing alertness and attention. It will also focus on improving the stroke victim's orientation to person, place, time and situation. It will also help the stroke victim to comprehend speech, another problem associated after a stroke attack. Post Discharge, the stroke patients could be on home cognitive rehabilitation or on outpatient basis depending on the needs of individual patient.

Strategies of Cognitive Rehabilitation:

- Form and follow a routine during the day, which would help to improve the sequential memory.
- The patient should be oriented to date, day, time, place and person.
- Break down tasks and activities into steps which would help to re-learn the lost activities and functions. It would also help to learn new information and recalling it faster.
- Make note of things, activities and important events. For example: medication time or doctor's appointment, etc.
- Arranges the things that the patient uses in fixed places and cupboards, this will help the patient remembering where he has to look out for particular things and to find it easily.
- The patient should be given responsibilities within his capacity which would help him be cognitively active and would also help increase his confidence if he is successful. For e.g. Making phone calls, making online payments, etc.

Sexual Counseling Post Stroke:

Talking about ones feeling about one's body and appearances to a psychologist or the partner would be very helpful post stroke. You can express your feelings in many different ways, through talking but also with body language and physical contact such as kissing and cuddling. Taking the first step

may be the biggest hurdle to overcoming your anxiety and shyness about resuming sexual contact. Retaining closeness and intimacy within your relationship will help to overcome difficulties. It is important to keep communicating with each other. There can be a subtle change within a relationship when a partner becomes a carer, especially when assistance is needed with personal care. This can sometimes cause embarrassment or affect the way you feel about each other.

Changing Roles and Assigning Duties:

Caring for Caregivers:

Part of what makes the adjustment to a disability so difficult is the disarray it throws your life into. All of a sudden the old rules don't work anymore, and new ones need to be drawn out. This can be an extremely frustrating time, particularly for the care recipient and their caregivers. You can minimize these frustrations by drawing up a care plan [115].

Every family tends to have 'tried and true' ways of making decisions. Family members may also have 'roles' that give them certain duties and responsibilities. If a family member becomes disabled, one person may immediately assume the role of primary caregiver, whether the arrangement is implicit or explicit. Because the responsibility of a primary caregiver is so great, and so important, it is best to sit down as a group and openly discuss the care of your loved one. Rather than anticipating help from people, who may or may not be aware of what is expected of them, having everyone come together and talk about what needs to be done can help prevent family conflicts. Family members can also use this time to talk about their own feelings.

When holding the family meeting or conference, the different responsibilities for your loved one's care can be assigned to everyone who is able and willing. No one should be forced to accept a role unwillingly - particularly the role of primary caregiver. Perhaps one family member can be responsible for finances, another for transportation, another for housing. A back-up plan in the case of respite care, changing needs or an emergency should also be discussed. And be flexible. If the initial plan doesn't work out, hold another meeting and try to sort out the problems. Some decisions won't be easy, but working together as a family is the best solution.

If the loved one has to move, in with family members, the changes that this will cause in the household need to be discussed frankly [116]. The family may have less time for leisure and privacy. Children may also be confused by the new addition to their home. Still, if you explain the situation to children and offer ways for them to help, they may volunteer to help with certain duties, like cleaning, preparing dinner or spending time with your care recipient.

Even if one person accepts the role of primary caregiver, a role that may include housing and daily care activities, they should be able to ask for significant help from other family members. The primary caregiver will also need regular breaks from time to time, including longer 'vacations'. Other family or friends can take over care temporarily, or perhaps the care recipient can stay in a nursing home for a week or two. As well, most states have government-run programs that offer respite services. [117]

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Ch.3 Cerebral Palsy

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Definition:

Cerebral palsy (CP) is a collection of motor disorders resulting from damage to the brain that occurs before, during or after birth. The damage to the child's brain affects the motor system, and as a result the child has poor coordination, poor balance, or abnormal movement patterns-or a combination of these characteristics. The universally recognized definition of cerebral palsy is ***"a non progressive but not unchanging disorder of movement and / or posture due to an insult to or anomaly of the developing nervous system".*** CP is a static disorder of the brain, not a progressive disorder. This means that the disorder or the disease process will not get worse as the time goes on. However the child may appear to worsen if not given proper intervention, not because of an increase in the lesion but just because the damaged brain is not able to cope up with the physical demand of the growing body and the increasing demand of the environment surrounding the child.

History :

In the late 1800's Sir William Osler, a British Physician, coined the term CP. Around the same time, Sigmund Freud (1856-1939), a Neuropathologist in Austria, conducted extensive research and published some of the earliest papers on the subject. But William John Little (1810-1894),an English surgeon was responsible for the first clinical description, and the recognition of the link between low birth weight and perinatal events in 1843.Dr. Little theorized that lack of oxygen damaged the sensitive brain tissues, which controls movement. Sigmund Freud disputed Dr. Little's conclusions. He pointed out that most CP children suffer from additional problems like seizures, mental retardation and visual disturbances. This led him to believe that CP might be caused during the brain's development in the womb many months before birth. However doctors only seriously began investigating CP in 1861 when Dr. William John Little,published a paper describing children with spastic diplegia. Spastic diplegia is still sometimes called Little's Disease.

For many years CP was treated from a surgical perspective that was based upon surgeon's experiences in treating poliomyelitis. The enthusiasm for surgical intervention to correct deformities, to provide stability and to improve motor control decreased as assessment showed that deformities recurred or new ones developed.

In 1932,Winthrop M.Phelps began developing a new approach. Phelps became aware of the necessity for including exercise, muscle training, and bracing in the treatment of CP. Since Phelp's era various non surgical approaches have evolved, emphasing Neuromuscular training.They include Neuro Muscular Reflex therapy (Fay & Doman), Neurodevelopmental treatment (Bobath), Neurophysiological Approach (Rood), Proprioreceptive Neuro Muscular Facilitation (Kabat, knott & Voss) and Sensory Integration.

Prevalence & Incidence:

Prevalence rates for CP vary somewhat in different countries and regions, largely reflecting economic and nutritional factors and access to good obstetric care. Because of the absence of any "test " for CP, the vagueness and elasticity of even the best definition, inaccurate ascertainment (many cases are never reported to physicians or public health authorities) and lack of agreement between different experts in the inclusion of the individual cases, precise figures are difficult to obtain. Further confusion results because many cases die in infancy and others may be so mild as to be unrecognized. Prevalence rate for CP therefore should be presented as the prevalence at a given age per 1000 live births of the defined population under study.

Of every 2000 infants born, 5 are born with CP. This incidence had remained constant for over 30 years. Although improvement in medical care have decreased the incidence of CP among some children who otherwise would have developed the disorder. Medical advances have also resulted in the survival of children who previously would have died at a young age, and many of these young children survive with an impairment or a disability such as CP. Throughout the industrialized world a prevalence of about 2 per 1000 is found. In contrast estimates for India are around 45 per 1000, rather more in rural & less in urban populations.5 to 8 percent of premature infants under 1500 grams birth weight, who survive have CP. The marked difference in prevalence provides a challenge for Indian Preventive medicine to make the best

obstetric and perinatal care available to all communities. Essential keys to progress include improved education and nutrition to mothers to be.

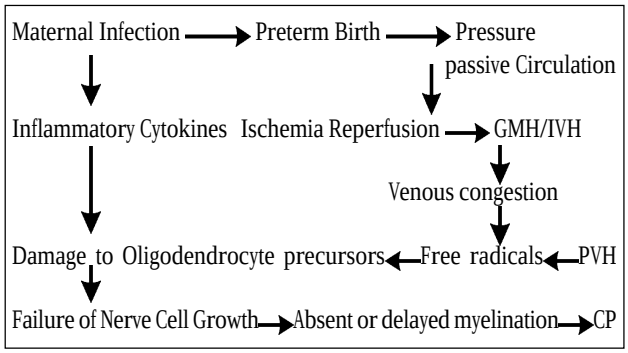
Etiology :

Many causative factors produce CP, infact it is the multiplicity of causative factors that led Denhoff and Robinault to refer to CP as a group of diseases. The factor common to the group is time of occurrence of the brain dysfunction in the prenatal or natal period. The brain damage that leads to CP can be caused by :

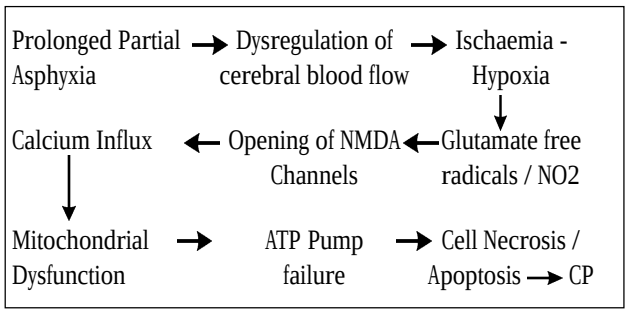
1. Idiopathic (no known cause of damage to brain during pregnancy) -still the most common cause.
2. A viral infection during pregnancy such as cytomegalovirus (CMV) or rubella.
3. Iso-immunization reactions eg : Jaundice due to immunization mechanism related to mixing of fetal Rh positive blood with maternal Rh negative blood
4. Hydrocephalous either before or after birth.
5. Developmental Defects.
6. Defects in Biochemical maturation
7. Defects in Hereditary material (genetic)
8. A blood clot in the fetus brain causing a stroke while in vitro.
9. Haemorrhage (Bleeding into the brain) : while inutero this could be due to a bleeding disorder, after birth, this can be seen as a complication of extreme prematurity.
10. Prolonged period of Asphyxia (lack of oxygen) for example abruptio placenta, i.e. when the placenta tears away from the uterine wall during labor, by cutting off the baby's blood supply.
11. Infections, eg : Bacterial meningitis after birth.
12. Head trauma from shaken baby syndrome (child abuse) during the first year of life.
13. Toxins & Poisons, eg : Lead poisoning during the first two years of life.

Etiopathology:

The underlying basis of most neurodevelopmental sequelae in CP infant is white matter damage, collectively called periventricular Leukoence-phalopathy. This term encompasses germinal matrix haemorrhage (GMH), Periventricular Haemorrhage (PVH), Intraventricular Heammorhage (IVH), Periventricular Heammorhagic Infarction, and Periventricular Leukomalacia (PVL) Preterm infants are more prone to GMH, PVH, and IVH, because their cerebral circulation is sensitive to changes of bold pressure (pressure passive) and they lack supporting glia in their germinal matrix.



Events leading to CP in preterm Infant.



Events leading to Cerebral Palsy in Term Infant

Classification of CP :

The diverse motor and non motor associated defects in CP, would suggest the difficulties in categorizing this disorder. Therfore children with CP are categorized on the basis of the types of neuromotor deficit and topographic distribution of the deficit. According to, the following schema, which uses only major groupings of the classification of the American Academy of Cerebral palsy.

Neuromotor Classification by Minear (1956)

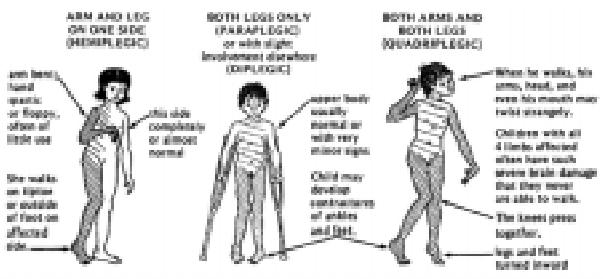
1. **Spastic :** This type is characterized by exaggeration of the stretch reflex and increased deep tendon reflexes in the affected parts.
2. **Athetoid :** The chief characteristics of this type are the slow worm like, involuntary uncontrollable, unpredictable and purposeless motions at rest.
3. **Rigidity :** When the part is moved, there is a continuous resistance in agonist and

antagonist muscles, simulating the sensation of bending a lead pipe. The degree of rigidity may, from time to time be referred to as "lead pipe ", "cog wheel " or "intermittent " rigidity. The principal clinical finding is the hypertonicity of the muscles which in some patients is so great that no motion is present.

4. **Ataxia:** The principal sign noted in the ataxic patient is disturbance of balance and equilibrium. The ambulation pattern has been described as reeling, drunken type gait.
5. **Tremor :** The chief characteristics of this type are the involuntary, uncontrollable motions that are reciprocal and regular in rhythm.
6. **Mixed :** Not all children with CP can be diagnosed as true spastics, athetoics, or ataxics.About 1% of the total may be mixed cases in which there is more than one type of the above described characteristics.

Topograhic Distribution Of Neuromotor Involvement

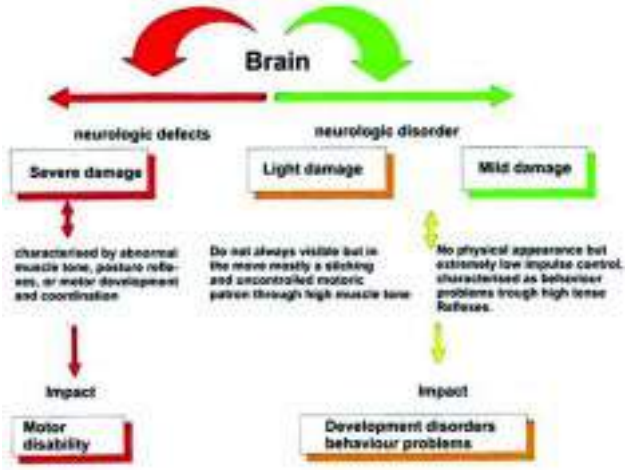
1. **Paraplegia:** involvement of the lower extremities. Patients with paraplegia are practically always of the spastic type.
2. **Hemiplegia:** involvement of an upper and lower extremity on the same side of the body. Patients with Hemiplegia are almost always spastic, but occasionally an athetoid hemiplegic patient may be seen.
3. **Triplegia:** involvement of three, extremities, usually both lower extremities and one arm. The disability is usually of spastic type.
4. **Quadriplegia or tetraplegia :** involvement of all four extremities. The term diplegia is some times used to indicate that the lower extremities are more involved than the upper. Almost all patients of athetosis have all four extremities involved.



Classification based on Severity :

This type of classification is used emphasing on the quantification of neuromotor involvement that limits patient's ability to perform Activities Of daily living.

1. Mild: The patient needs no treatment, since he has no secondary problems, is able to care for his daily needs and ambulates without the aid or any appliance.
2. Moderate: The patient need treatment, since he is inadequate in self care, ambulation, and had associated problems. Braces and self help appliances are needed.
3. Severe: The patient needs treatment, but the degree of involvement is so severe that the prognosis of self care, ambulation is poor. And also the associated secondary complications are major concern hampering patient's quality of life.



The following list presents the medical problems most often associated with CP.

Neurological Problems:

a) Mental Retardation.
b) Learning Disabilities.
c) Attention Deficit Hyperactivity Disorder / Behavioral issues.
d) Seizure Disorder (epilepsy).
e) Visual Impairment.
f) Swallowing Difficulties.
g) Speech Impairment.
h) Hearing Loss.
i) Hydrocephalus.

Orthopaedic Problems:

a) Scoliosis.

b) Hip Dislocation.

c) Contractures of Joints.

d) Discrepancy in leg length.

Secondary Effects:

a) Communication Disorder.

b) Drooling.

c) Poor Nutrition / growth

d) Aspiration Pneumonia and gastro esophageal reflux.

e) Depression.

f) Fragile bones and frequent fractures.

g) Cavities-Tooth decay and gum disease.

h) Constipation or Bladder bowel incontinence.

Quadriplegic CP child with spasticity and poor voluntary control of limbs.

Summary:

DYSTONIC Predominantly abnormal tone	Spastic	65%	Quadriplegia (25%) Diplegia (20%) Hemiplegia (20%)
	Dystonic	Usually regarded as Spastic	
	Hypotonic	May eveolve to spastic or athetoid types	
DYSKINETIC Predominantly abnormal movement	Athetoid, Choreo-athetoid tension	15%	
	Ataxic	Rare	
MIXED		20 % or more	

A study of 1000 children with cerebral palsy in northern India showed spastic Quadriplegia to be the most common group (61%), followed by spastic diplegia(22%), with dyskinetic cases amounting to about 8 %.Mental retardation (75%)appeared to be very much common in this population than in industrialized nations.

Diagnosis: Early signs of CP may be present from birth. Most children with CP are diagnosed during the first 2 years of life. But if a child's symptoms are mild, it can be difficult for a doctor to make a reliable diagnosis before the age of 4 or 5.The growth and development of infants born with CP, may be erratic, showing normal patterns of development in some areas but not in others. It is at these points of perceived delay that the parents need to become concerned, seek professional advice, investigate the problem and if necessary, obtain remedial help for the child. Thus, diagnosis can be based on following broad criteria's :

A) Concept of Normal Neurological Maturation:

Overviews of Developmental milestones

Age	Gross Motor	Visual / Fine Motor	Languages	Social
1 months	Prone, Lifts head	Head usually fisted; stares or objects	Soothes to voice	Regards face
3 months	Supports chest in prone position	Gross placed rattle; follows slow moving objects with eyes	Coos / laughs	Smiles easily, spontaneously
6 months	Rools and sites well, without support	Reaches and grasps, transfers hand to hand	babbles; plays peek-a-boo	Fear of Strangers; smiles at self in mirror

12 months	Walks alone	Pincer grasp of raisin	Say "mama," "dada", + 2 other words	Shy, but play game, gives affection
18 months	Walks up steps	Stacks 3 blocks; manages spoon	Points to named body parts; follows simple commands	Helps with simple tasks; imitates play
24 months	Alternates feet on stairs; kicks balls	Stacks 6 blocks; turn books pages	At least 50-word vocabulary; understands 2-steps commands	Washes / dries hands; helps get dressed
30 months	jumps with both feet	Holds pencil in hand, not fist	Use pronouns "I," "ME," "YOU" correctly; states full name	Plays tags; asserts personality
36 months	Balances on 1 foot, 5 sec,; rides tricycle	Imitates block bridge; buttons	Recognizes 3 colour	Plays with children, takes turns

Note:- It is not uncommon for a child to lag behind in one area and be advanced in another. However, there are generally accepted limits for what is considered "normal development".

Source:- Adapted from The Harriet Lane Handbook, 17th ed. (Philadelphia Elsevier Mosby, 2005).

B) Overview of Abnormal Infant Neurological Development:

Every child development involves gaining mastery of four major types of skills: gross motor, fine motor, communication and social. Development in these areas occurs simultaneously to prepare the child to meet physical, social, linguistic and emotional demands. Significant delays in early child development are "red flags" that should prompt parents to discuss their concerns with the child's doctors.

Developmental Red Flags

Milestone	Normal	Concern if Not Acquired by
GROSS MOTOR		
Head up / chest off in prone position	2 months	3 months
Rolls front to back, back to front	4-5 months	6-8 months
Sits well un supported	6 months	8-10 months
Creeps, crawls, cruises	9 months	12 months
walks alone	12 months	15-18 months
Runs; thros toy, from standing without fall	18 months	21-24 months
Walks up and down steps	24 months	2-3 years
Alternates feet on stairs; pedals tricycle	3 years	3½-4 years
Hops, skips; alternates feet going down stairs	4 years	5 years
FINE MOTOR		
Unfists hands, touches objects in front of them	3 months	4 months
Moves arms in unison to grasp	4-5 months	6 months
Reaches either hand, transfers	6 months	6-8 months
Pokes forefinger; pincer grasp; finger feeds; holds bottle	9 months	1 years
Throws objects, voluntary release; mature pincer grasp.	12 months	15 months
Scribbles in imitation; holds utensil	15 months	18 months
Feeds self with spoon; stacks 3 cubes	18 months	21-24 months

Turns pages in boos; is stready cup drinker; removes shoes and socks	24 months	30 months
Urbuttons; has adult pencil grasp	30 months	3 years
Draws a circle	36 months	4 years
Buttons clothes; catches a ball	4 years	5 years
LANGUAGE		
Smile socially after being talked to	6 weeks	3 months
Goos	3 months	5-6 months
Orients to voice	4 months	6 months
Babbles	6 months	8 months
Waves bye-bye; says"dada," "mama" indiscriminately	8-9 months	12 months
1-2 words other than dada / mama; follows 1-steps command with gesture	12 months	15 months
7-20 words; knows 1 body part; uses mature jargoning	18 months	21-24 months
2-word combination; 20 words; points to 3 body parts	21 months	24 months
50 words; 2-words sentences; pronouns (inappropriate); understands 2-step commands	24 months	30 months
3-word sentences; plurals; minimum 250 words	36 months	3½-4 years
knows colour; asks questions; multiple-word sentences (tells story)	4 years	5 years
SOCIAL		
Regards face	1 month	1-2 months
Recognizes parents	2 months	2-3 months
Enjoys viewing surroundings	4 months	5-6 months
Recognizes strangers	6 months	7-8 months
Reciprocal games: so big, pat a cake	9 months	12 months

Primitive Reflex Patterns:

These are patterns seen in the early stages of development with disappear later on or with advanced age. These reflexes are essential for normal progressive motor development.However not overcoming these primitive reflex patterns at the right time should be definitely considered as abnormal. Initially lower centers such a s spinal cord control these movements but later on higher centres like midbrain and cortex take control over them and dominate the lower ones thus integrating them for various voluntary functional task. Accordingly we have four levels at which reflexes are regulated:

1. Spinal cord: Flexor with drawal, Extensor thrust, Cross extension, Palmar grip, Plantar grip, Sucking reflex, Rooting reflex, Primitive walking.
2. Brainstem: Symmetrical Tonic neck reflex, Assymmetrical Tonic neck reflex, Tonic Labrinthine reflex, Positive and negative supporting reaction.
3. Midbrain: Optical Righting, Labrinthine reflex, Body on neck, Body on body, Neck righting reflex.
4. Cortical reflex: Equilibrium reactions, which can be in prone, supine, kneeling, sitting and standing. Equilibrium reaction in supine and in prone position in present from 6 months onwards, in quadripud position is present from 8 months, and in sitting starts normally from 10-12 months. Kneeling equilibrium occurs from 15 months onwards and finally in standing occurs from 18 months onwards.

Automatic reflexes: Moro's reflex, Gallants trunk incurvatum, Landau's reflexes and parachute reflex.

Abnormal Manifestations and Prognosis

1) A child with spinal cord dominance will not be able to walk and carry out ADL and therefore he / she will be bedridden throughout. This condition leads to a lot of contractures and thus aim of treatment is to

prevent secondary complications due to lack of ambulation like preventing tightness or contractures, respiratory complications, etc.

2) Child with brain stem dominance will also be bedridden and unable to walk.Aim of treatment is same as above and all secondary complications due to lack of ambulation need to be prevented.

3) Midbrain dominance children will have a relatively better prognosis, such a child can ambulate due to and or absent equilibrium reactions, some walking aids amy be required.

4) Cortical Dominance children will have near mormal development.

C) Prediction of Developmental Abnormality:

1) Biochemical:

Apgar Scoring system based on observation of color, respiratory effort, heart rate, tone, and reflex activity has been a guide to assess child's condition at birth. Predictability is improved with the addition of a low 5 - min Apgar score and the need for intubation at birth. Various newborn spinal fluid components have been shown to be effective markers of Hypoxic ischaemic encephalopathy. Predictive effects of newborn urine products have also been considered since asphyxia results in injury to the kidney. Paucity of urine flow (oliguria) and elevated ß2 microglobulin at 36 hours have been found to correlate with later neurological deficits.It should be kept in mind that even if these or other biochemical markers of hypoxic ischaemic stand the test of time an experience, they deal only with asphyxia and ischaemic injury to the brain, which represent but one etiological basis among many for static encephalopathy.

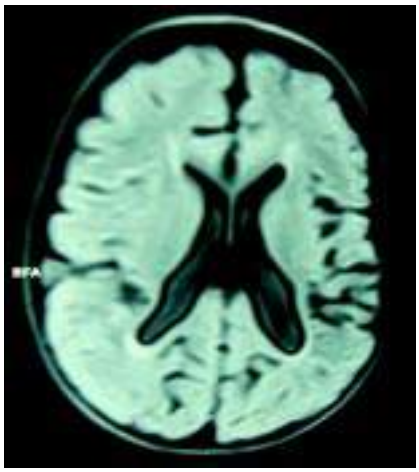
2) Prenatal:

Prenatal screening includes amniocentesis, chorionic villus sampling, and use of ultrasound are used to monitor the pregnancy and pick up early cases of CP or developmental disabilities such as Down's Syndrome.

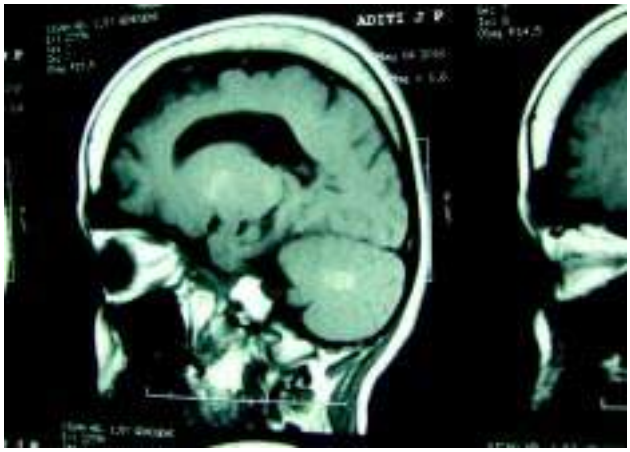
3) NeuroImagining:

Doctors diagnose CP by evaluating a child's motor skills and taking a careful and thorough look at medical history. In addition to checking for the most characteristic symptoms - slow development, abnormal muscle tone, unusual posture - doctors have to rule out other disorders that could cause similar symptoms. Neuroimaging techniques like MRI can also show a doctor the location and type of brain damage and can hence, help in predicting the severity and type of disorder and prognosis accordingly.

MRI Brain showing perinatal hypoxic brain injury



MRI brain showing periventricular leukomalacia.



MRI Brain showing perinatal hypoxic brain injury

Symptoms seen in CP correlating to area of brain damage seen on MRI

ANATOMICAL AFFECTION	MRI FINDINGS	CLINCAL FINDINGS
Bilateral white matter necrosis,descending motor fibres,optic,acoustic radiations	Periventricular leucomalacia	Spastic diplegic, quadriplegic, visual & cognitive defects.
Focal, multifocal ischaemic brain necrosis	Specific vascular infarction, typically left MCA	Hemiplegic, seizures.
Basal ganglia neuronal injury	Status marmoratus	Choreoathetosis or mixed picture.
Selective neuronal necrosis	Lateral geniculate, thalamus, basal ganglia	Mental retardation, seizures
Parasaggital cerebral injury	Bilateral medial & posterior portions of cortex	Upper extremity more severely affected than lower.

Physical Assessment and Developmental Screening Scales:

A large variety of rating / screening instruments have burgeoned within recent years, with varying degrees of accuracy for prediction of

1. Developmental delay
2. Definite developmental abnormality
3. Specific Developmental Diagnosis.

Among those most commonly used at present at the present tiem are the following :

I) Developmental Scales:

1. Amiel - Tison
2. Bayley
3. Brazelton
4. Dubowitz
5. Haataja et al
6. Peabody
7. Prechtl

II) Developmental Screening Testes:

1. Denver Developmental Screening Test (Denver II) (most commonly used).
2. Batelle Screening Test
3. Clinical Adaptive Test (CAT)/ Clinical Linguistic and Auditory Milestone Scale (CLAMS)
4. Knobloch Revised Screening Inventory
5. General Movement Assessment (GMs).

III) Motor Assessment Instruments:

1. Alberta Infant Motor Scale (AIMS)
2. Early Motor Pattern Profile (EMPP)
3. Gross Motor Function Measure (GMFM)
4. Movement Assessment Inventory (MAI)
5. Test of Infant Motor Performance (TIMP)

Developmental Evaluation Procedure

A) Developmental History

1) Chief Complaint
2) Family and genetic history
   a) Pregnancy
   b) Labor / Delivery
   c) Perinatal / Neonatal
3) Developmental Milestones
4) Other developmental Features.
5) Reviews of systems
6) Past medical history

B) Developmental Physical Examination.

C) Developmental Neurological Examination.

1) General Observation.
2) Quality of general movements.
3) Tone
4) Patterns of motor behavior.
   a) Primitive
   b) Postural
5) Sensation
6) Cranial nerves.
7) Cerebellar Function.
8) Dystonia.

9) Motor signs
   a) Upper motor
   b) Lower motor
10) Neurological Soft signs.

D) Developmental Screening Instruments.

E) Laboratory Evaluation.

Speech Affection in Cerebral Palsy:

Children with cerebral palsy have higher risks of having a concomitant problem like autism, hearing impairment, mental retardation, learning disability etc. The symptoms that the children present with depend on the severity of problem and presence of the associated problems.

Children with cerebral palsy can have problems in comprehension, delayed expressive speech i.e children may speak few sentences to complete absence of spoken language, difficulty in grasping new concepts and learn normally due to mental retardation. Developmental dysarthria or impaired speech clarity is also very common in children with cerebral palsy. Due to the muscle affected, the children with cerebral palsy may present severe dysphagia and are many of the times fed on semi solids and liquids. In the developmental stage of life, they fall victim to malnourishment and aspiration pneumonia due to which they further deteriorate and experience frequent hospitalization.

Treatment:

The latest and the most effective way of treating Cerebral Palsy children comprise of an integrated approach. The team effort is required to make the child independent in most ADL for his age and thereby improve his quality of life.

The team includes:

- Parents
- Physicians
- Teachers
- Nurses
- Physical Therapists
- Occupational Therapists
- Speech Language Pathologists
- Audiologists
- Rehabiliattion Workers
- Orthotists
- Clinical Pscychologists
- Vocational Counsellors
- Social Workers.

The therapy aims at developing rapport with parents and patients and is extremely important as any goal will be difficult to achieve without the cooperation of the patient and relatives. The patient has to be motivated regularly to gain confidence. Only when the motivation level of the patient is very high will the child cooperate in the treatment session. The goal set for the patient should be challenging but at the same time achievable. The goal of any therapy is to maximize each child's functioning to all that it can be.

Physiotherapy Management of CP:

Physical and Occupational therapist's approach overlap because their focus is to help the child develop motor skills. Areas in common include seating assessments, early intervention therapy, and developmental testing. Physical therapists, however focus mainly on gross motor or large muscle activities involving the legs, such as walking, bracing using crutches, and rehabilitation after surgery.

Treatment in CP has evolved from focusing on impairments (such as spasticity and contractures) to activities (such as walking), and then to considerations of participation (such as child's family role as a sibling) Thus a child can be impaired and not necessarily be disabled, and a person can be disabled without being handicapped. To make it clear:

Impaired means a deviation from normal

Disabled denotes restricted ability to perform normal activities of daily living; and

Handicapped means being unable to achieve an age appropriate role in the society.

Treatment Interventions:

Various interventions have been designed and developed by various scientists and therapists to treat CP children. At times a combination of few help to overcome problems in them. The theories and the treatment interventions are explained in brief :

I) Neuro Developmental Technique (NDT) :

The most renowned of all interventions addressing the movement and posture of the child with CP is NDT.The concept was developed by Karel and Berta Bobath in the 1940's and it views development dynamic, sequential, cephalocaudal, proximal to distal, automatic before conscious, responsive and lastly adaptive.

Bobath's promoted the use of handling techniques

to inhibit abnormal tone and primitive reflexes and to facilitate normal movement. They believed that children with CP needed the experience of normal movement. For children unable to move, a therapist's hands provided the experience. Quality of movement was considered very important. Bobath's also used reflex- inhibiting positions to reduce the effects of the tonic reflexes. Treatment progression centered on the normal developmental sequence, assuming carryover to functional tasks.

Today, NDT emphasizes functional goals. Principles of treatment include weight shifting, weight bearing, and normalizing muscle tone. Quality of movement is still important and may reduce abnormal stresses on joints, possibly preventing secondary impairments and deformity. Modalities include the use of balls, bolsters, horseback riding, and swimming. Therapists combine NDT principles with a variety of other approaches such as strengthening and the use of adaptive equipment.

The art of NDT:

The intervention process begins with the assessment of the individual's functional performance. Analytical problem solving is used to develop a treatment plan. Treatment focuses on increasing function by building on the individual's strengths while addressing the impairments.

Therapeutic handling is one strategy, which is utilized to help the individual achieve his or her functional goals. Therapeutic handling is integral in NDT approach.

Therapeutic handling allows the therapist to:

- Feel the child's response to changes in posture and movement
- Facilitate postural control and movement synergies that broaden the client's options for selecting successful actions.
- Provide boundaries for movements that distract from goal and
- Inhibit or constrain those motor patterns that, if practiced, lead to secondary deformities further disability, or decreased participation in society
- The therapist's hands are purposefully and specifically placed on the child's body during facilitation techniques. Mrs. Bobath called this therapeutic handling through "key points of control".

Based on the child's motor control, presence or absence of abnormal muscle tone and the quality of volitional movement present, strategy adopted to treat child may contain either the facilitation approach (absence of tone) or the inhibitory approach (presence of abnormal tone)

Facilitation:

The use of primitive or tonic reflexes, quick stretching, tapping, vibration, approximation, and weight bearing may be required to prepare the child for the performance of functional activities. In addition, facilitation can also be in terms of education, mechanical, postural, manual, motivation, etc based on the clinical presentation of the child and goal of therapy.

Principles of facilitation:

The most important Kinesiological parameters need to be considered are

- Range of motion
- Alignment
- Base of support
- Movement in all three planes (sagittal, frontal and transverse plane)

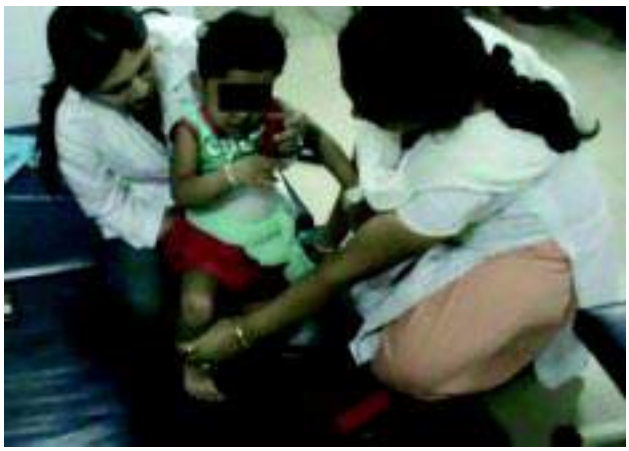
Inhibition:

Therapist also uses inhibition to restrict the child's atypical postures and movements that may prevent the development of more selective motor patterns and efficient performance.

- Prevention or redirection of components of movements those are unnecessary and interfering with intentional coordinated movement.
- Constrain the degree of freedom to decrease the amount of force the child uses to stabilize posture.
- Balance antagonistic muscles
- Reduce spasticity or excessive muscle stiffness that interferes with moving specific segments of the body.

Child performing quadrapud activity

Child prone on elbows to promote weight bearing on the upper limbs.



Child performing half kneeling activity.

Sensory integrative therapy:

Sensory integrative (SI) therapy, was developed by Jean Ayres in the 1970's, is based on promoting the organization and processing of sensory information. Proponents believe that information coming into the body from the proprioceptive, tactile, and vestibular systems is disorganized and not processed well. This lack of cognitive processing is considered a perceptual problem. These processing problems cause difficulties with motor planning and motor control. Therapy focuses on movement and environmental awareness. The use of swings, scooters, and other moving objects helps the child process sensory input and use sensory information to plan movement and gain postural control. Traditionally, occupational therapists are more involved in the use of SI treatment.

Children with CP often have sensory processing dysfunction (SPD) and show sensory based motor dysfunction, Sensory processing evaluates how the brain takes it what it receives through the senses.In SI therapy state of arousal of child is very important for learning movement.Depending on arousal therapist will upregulate or downregulate sensory stimulation. Use of varying textures like cotton, rough gauge, deep pressure are used to stimulate kinesthetic and Proprioreceptive input.

Normalizing tone of the muscles:

In the infant with CP,spasticity is seldom significant in the first 6 months, but sometimes between 6 months to 24 months, it starts becoming apparent.Spasticity is defined as hyperexcitability of the stretch reflex, resulting in a velocity dependent increase in muscle tone. Secondary to the brain lesion, there are also mechanical changes in the muscles of children with CP.the muscles produce more force for a given change in length and therefore feel stiff and resist passive lengthening.

In children with hypertonicity slow passive movements, sustained stretch, cryotherapy over the muscles for 15 to 20 minutes, stimulation of antagonist movement and vibrations are used. On the contrary, for the cases with hypotonicity weight bearing, joint compression, rhythmic stabilization, vibrations, cryotherapy in brisk manner and tapping is used.

Aaprt from therapy medications like Lioresal, Tizanidine or Dantrium are also prescribed orally. In severe cases Baclofen Pumps are also recommended. A dorsal Rhizotomy, is another surgical treatment, in which nerves as they exit from the spinal cord are identified and the ones to be found most involved with spasticity are cut.

Serial Casting and intramuscular botox injections are also recommended in few cases where spasticity is interfering with functions.But effect of botox lasts for 6 -8 months and needs a good back up with exercises.

Swiss ball activities to normalize tone and strengthen trunk muscles.

Developing postural reaction:

Postural adjustments are essential if the child is to move freely and to adjust to various environmental demands rapidly. Motor milestones can be best achieved by good postural reactions. Postural reaction consists of righting reactions, protective extension and equilibrium reactions. Initially children first develop righting reactions. Righting reaction allow the child to orient his head in space so that the eyes and mouth are horizontal regardless of the position of the body. It also helps in maintaining the proper alignment of the head with respect to the body and vice versa. Once righting reaction develop in a particular position protective extension comes up. Lastly equilibrium reaction emerge which helps the patient to counteract the opposing force and enable them to maintain balance. These reactions are best developed by various exercises on vestibular ball and tilt board.

Rood approach:

Rood utilized both the sensory and motor systems to facilitate movement with her treatment techniques. She felt that preventing the development of abnormal movement patterns was important. She used sensory input to achieve or relax muscles to promote normal movement. For example, during therapy gentle stroking is used to have relaxatory effect on the muscles thereby used to inhibit spastic muscles.On the other hand brisk stroking, tapping, quick ice, etc, has been found to develop tone in the muscles, hence can be used for cases like flaccidity.

Pediatric Constraint Induced Therapy:

Constraint induced therapy is used to improve the use of affected upper extremity in a child with Hemiplegic CP.The normally functioning or stronger upper extremity is immobilized for a variable duration in order to force the use of the affected or weaker upper extremity over time.

The efficacy of this approach has not been established, and the adverse effects of prolonged immobilization often normally developing upper extremity are a significant concern.

Vojta's Technique:

This approach was introduced by Dr.Vaclac Vojta and used afferent sensory stimulation through touch, stretch and pressure for facilitation of movement. Reflex locomotor patterns and proprioreceptive input are the basis for treatment. Trigger points were used to reflexly to stimulate creeping, rolling, or crawling activities. Proponents of Vojta suggest that very early intervention does not allow the development of abnormal crawling patterns and subsequently promotes normal walking patterns.

Strengthening Programs:

Children who cannot walk are by definition weak. There is no evidence that strengthening increases spasticity. Infact, there is evidence that weakness is a problem in this population of children and strengthening is effective in reducing weakness and improving function. Therapist need to lengthen spastic agonist and strengthen the antagonist. For very young children, weight training may not be appropriate but therapists can incorporate strengthening activities in to child's therapy program through games and repetition of functional movements. eg: In Partial weight treadmill training child is suspended in a specialized harness and helps strengthening the weak antigravity muscles and facilitate normal gait patterns.

Stretching and Mobility:

The muscles should be maintained at the appropriate physiological length for normal movement control and normal postural adjustments. In CP because of delay or absence of normal movement muscles are usually in a shortened position hence stretching of the muscles is essential to increase the neuromuscular control. Length of the muscles should be maintained not only through stretching but also through various functional activities. Thus home exercises and activities are very important in additional to the therapy at the pt clinic.

Taping :

Taping provides joint stability, supports weak or over lengthened muscles. It provides sensory proprioreceptive inputs. It is known to increase or decrease activation.

Electrical Stimulation:

Neuromuscular electrical stimulation is supposed to increase range of motion, strengthen muscle and bring about muscle reeducation. Therapeutic Electrical Stimulation (TES) helps in reducing tone and strengthening of muscles, but tolerance is sometimes an issues with children. Therefore

treatment is done during sleep, and recommended age for beginning TES is 2 years.

Aqua therapy or hydrotherapy:

Hydrotherapy is therapy performed in water. The effects of water give children a feeling of weightlessness, which helps to reduce tone and allow those children better motor control. It is also a good modality for gait training, especially in an overweight child who may be able to walk in water with relative weightlessness. In addition, swimming as a recreational activity is excellent in children with CP. For many children for whom walking consumes a great deal of energy, learning to swim, and using this a physical conditioning is an excellent option.

Theratogs:

They provide joint stability and increase body awareness. Improves posture, balance, gait and movement skills. More information available on www.theratogs.com. Stabilising Pressure Input Orthosis (SPIOS) are flexible, provide dynamic stability and balance thereby increasing body awareness in space.

Hyperbaric Oxygen Therapy (HBOT)

Hyperbaric oxygen therapy (HBOT) is the inhalation of 100% oxygen inside a chamber that is pressurized to greater than 1 atmosphere (therefore described as hyperbaric because the pressure is above atmospheric pressure). HBOT is typically administered at I-3 atmospheres of pressure.

It is not clear from a scientific standpoint how HBOT could help overcome damages to brain tissue that occurred years before in a child with CP. When HBOT was studied in a scientific manner in two group of children with CP, with a Control group placed in pressurized room air and a treatment group in pressurized oxygen, both groups improved, without any difference between the two groups. Similar results were found in a second such controlled study of children with CP.

Ear pain / discomfort and bleeding from the ear are by far the most commonly reported adverse events during HBOT. In addition, there may be an increased risk of seizures in those treated with HBOT.

Equine therapy:

It is also known as Horseback riding therapy or hippo therapy. The underlying theory is that the positioning and large movements provided by horseback riding are very helpful in establishing balance and relaxation of spastic muscles.The vertical motions of horseback riding are thought to provide sensory stimulus which decreases muscle tone. Sitting on horse helps with stretching hip adductors and improves pelvic tilt and trunk positioning. This allows better movement and range of motion for the therapist to work with, after the child finishes with session.

Bracing and Orthosis:

The role of orthoses in CP is :

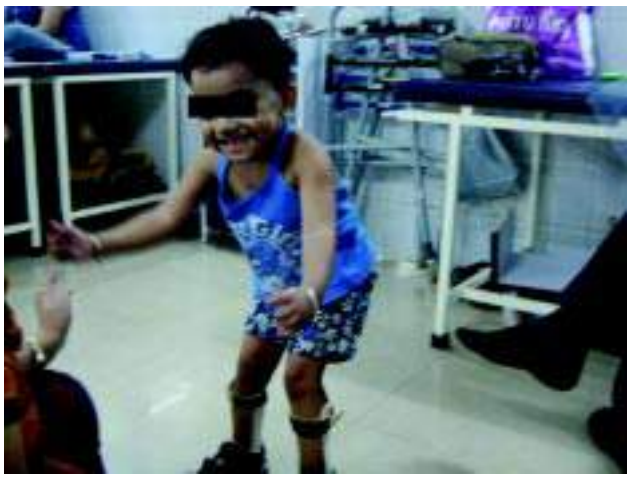
1. Improve function and efficiency.
2. Improve joint biomechanics and alignment.
3. Prevent Deformity.
4. Protection after surgery.
5. Encourage a normal motor patterning.

   The most commonly used Orthoses are :

   a. Ground reaction AFO (GRAFO)

   b. AFO's Solid and hinged.

   c. Supra malleolar Orthoses.



Child made to walk with AFO's

of rehabilitation, controlling ankle joint motion produces an effect on the knee during gait and by using an AFO to manipulate the ankle rockers, therapist can increase or decrease the plantar flexion knee extension couple. The correct selection and prescription of lower limb orthosis is essential to maximize a child's rehabilitation. Orthosis for correct period of time gives the best results.

Occupational Therapy Intervention

Cerebral palsy (CP)

CP is a non progressive disorder of posture and movement. It is often associated with epilepsy and abnormalities of speech, vision and intellect (resulting from a defect/lesion in the developing brain). It is a condition that occurs early in life and is present throughout a person's lifetime. It can affect all aspects of a person's development throughout their life

Occupational therapy Intervention

Occupational therapy is the art and science of enabling engagement in everyday living, through occupation; of enabling people to perform the occupations that foster health and well-being; and of enabling a just and inclusive society so that all people may participate to their potential in the daily occupations of life (Townsend& Polatajko, 2007, p. 372).

The practice of occupational therapy in Cerebral palsy means the therapeutic use of occupations, including everyday life activities with individuals, groups, populations, or organizations to support participation, performance, and function in roles and situations in home, school, workplace, community, and other settings.

Frames of Reference

Conceptual frameworks and Models are essential for determining "best practice". Best Practice involves imaginative problem solving, creative application of knowledge and research and evaluation of effectiveness of Intervention to bring about improved client outcomes and effect organizational change. Depending upon the impairments in a child a suitable frame of reference or combination of approaches is used

Examples of specific occupational therapy Frame of Reference models which are used in evaluation and treatment in CP include the:

- Biomechanical model
- Canadian Model of Occupational Performance (CMOP)
- Cognitive Disabilities model
- Lifestyle Performance Model
- Model of Human Occupation (MOHO)
- Neurodevelopmental therapy (NDT)
- Occupational Performance Model (OPM)
- Person Environment Occupation Performance Model (PEOP)
- Sensory Integration model

Occupational therapy assessment scales

Occupational therapy services may include comprehensive evaluations of the home and other environments (e.g., school), recommendations for adaptive equipment and training in its use, and guidance and education for family members and caregivers.

OT interventions can be classified into five specific intervention categories,

1. Training of sensorimotor functions including play activities to facilitate motor performance.
2. Training of daily activities skills including feeding, dressing, bathing, personal hygiene, writing etc.
3. Parental counseling in which parents are educated how to stimulate Independence in their child.
4. Advice and instructions regarding use of assistive devices including provision of mobility aids like wheel chair and bathroom devices.
5. Provision of splints such as hand orthosis to facilitate hand functions.

The above Classification is based on the International Classification of Functioning Disability and Health (ICF) and enables the categorization of all intervention possible in OT

The following are the occupational therapy tests that can be used specifically in the assessment of Cerebral palsy:

- BRUININKS-OSERETSKY TEST OF MOTOR PROFICIENCY (BOTMP)

  Purpose: Developmental motor skills

  Age Range: 4.5 -14.5 years

Areas Tested: Balance, strength, coordination, running speed and agility, upper limb coordination (ball skills), dexterity, fine motor control, visual-motor ability.

- CANADIAN OCCUPATIONAL PERFORMANCE MEASURE

  Purpose: To detect changes in parent or child's self-perception of performance over time.

  Age Range: Any age

  Areas Tested: Satisfaction and disability rating of daily activities and routines, which are, identified by the child and family as important part of daily life

- FUNCTIONAL INDEPENDENCE MEASURE FOR CHILDREN (WeeFIM)

  Purpose: To determine the severity of a child's disability, the measurement of caregiver assistance needed in the performance of functional activities, and outcomes of rehabilitation

  Age Range: Children without disabilities: 6 months to 8 years; Children with developmental disabilities: 6 months to 12 years; Children with developmental disabilities and mental ages less than 7 years

  Areas Tested: Eighteen items grouped into two major categories of function, motor, and cognition that are divided into six domains divided into sub domains:

  Motor, Self-care: eating, grooming, bathing, dressing, toileting, Sphincter control: bladder and bowel management, Transfers: chair, wheelchair, toilet, tub, and shower, Locomotion: wheelchair / crawl, stairs, Cognitive -Communication: comprehension, expression, Social cognition: social interaction, problem solving, and memory.

- GROSS MOTOR FUNCTION MEASURE (GMFM)

  Purpose: To evaluate change in gross motor function in children with cerebral palsy, describe a child's current level of motor function, and determine treatment goals.

  Age Range: No specific age range is recommended by the authors; however, the test has been validated on children between 5 months and 16 years. Seems best suited for children two to five years

  Areas Tested: Eighty-eight items of gross motor function divided into five dimensions: -Lying and rolling-Sitting Crawling and kneeling-Standing-Walking, running, and jumping. Items were selected to represent those typically performed by children by age five

- GROSS MOTOR FUNCTION CLASSIFICATION SYSTEM (GMFCS)

  Purpose: To classify a child's present gross motor function.

  Age Range: 12 months to 12 years

  Areas Tested: based on self-initiated movement, with emphasis on sitting, transfers, and mobility

- HOME OBSERVATION FOR MEASUREMENT OF THE ENVIRONMENT (HOME)

  Purpose: A screening tool to identify the quality and quantity of social, emotional and cognitive supports available to the child in the home environment

  Age Range: Infant and toddlers version birth to three years of age.

  Areas Tested: Infant and toddlers version: forty-five items clustered into six subscales: Parental responsivity, acceptance of child-Organization of the environment-Play materials-Parental involvement with the child.

- ORAL MOTOR / FEEDING RATING SCALE

  Purpose: To document oral motor / feeding patterns and feeding function

  Age Range: One year through adulthood

  Areas Tested: Two major areas of oral motor / feeding behavior: Oral motor / feeding patterns lip / cheek movement, tongue movement, jaw movement. Related areas of feeding function: self-feeding, adaptive feeding equipment, diet adaptation, position, sensitivity, food retention, swallowing, oral-facial structures

- PEDIATRIC EVALUATION OF DISABILITY INVENTORY (PEDI)

  Purpose: To determine functional capabilities and performance, monitor progress in functional skill performance, and evaluate therapeutic or rehabilitative program outcome in children with disabilities

Age Range: Six months to seven years, six months

Areas Tested: Two hundred seventy-one items divided into three subtests in the Functional Skill Scale:-Self care: eating, grooming, dressing, bathing, toileting-Mobility: transfers, indoors and outdoors mobility-Social function: communication, social interaction, household and community tasks. Also environmental modification and amount of caregiver assistance is systematically recorded in Modification Scale and Caregiver Assistance Scale

- SENSORY INTEGRATION AND PRAXIS TEST

  Purpose: Measures sensory systems contributions to balance and motor coordination

  Age Range: 4-8 yrs 11 months

  Areas Tested: Numerous tests of postural control, motor coordination & planning, fine and gross motor function, & sensory integration

- THE ALBERTA INFANT MOTOR SCALE (AIMS)

  Purpose: To identify motor delay and to evaluate maturation over time. Fifty-eight items related to posture, movement, and weight bearing in prone, supine, sitting, and standing AIMS has been designed to assess gross motor maturation, to trace motor retardation and to identify infants that might benefit from early intervention. In addition, the AIMS may also be useful in designing and monitoring a treatment program

  Age Range: Used during first year of life

  Type of Test: 58 item, performance-based, norm-referenced, observational too

- TEST OF INFANT MOTOR PERFORMANCE (TIMP)

  The TIMP is a test of functional motor behavior in infants

  Purpose: a criterion-referenced measure designed to evaluate motor control and organization of posture and movement for functional activities in infants

  Age Range: 32 weeks gestational age to age 4 months

  It predicts 12-month motor performance with sensitivity 92% and specificity 76% and preschool motor performance with sensitivity 72% and specificity 91% at 3 months of age

  Areas Tested: 27 observed behaviors and 26 elicited behaviors assessing the ability to orient and stabilize the head in space and in response to auditory and visual stimulation in supine, prone, side lying, upright, and during transitions from one position to another, body alignment when the head is manipulated, distal selective control of the fingers, wrists, hands, and ankles, antigravity control of arm and leg movement

- SCHOOL FUNCTION ASSESSMENT (SFA)

  Designed to facilitate collaborative program planning for students with a variety of disabling conditions.

  Purpose: Used to measure a student's performance of functional tasks that support his or her participation in the academic and social aspects of an elementary school program.

  Type of Test: criterion-referenced assessment

  Areas tested: Three parts: Participation in school activity settings; Task supports; Activity Performance. Includes physical and cognitive / behavioural tasks.

Occupational therapy strategies

The focus of OT Treatment should be on the facilitation of Independence. The management of a child with CP is done with the objective of optimizing functional abilities. OT focuses on development of skills necessary for performance of Activities of Daily Living. These activities include play, self care activities such as dressing, grooming and feeding and fine motor tasks such as writing and drawing. OT also addresses cognitive and perceptual disabilities especially in the visual motor area. Another aspect of OT is the adaptation of equipment and seating to allow better upper extremity use and to promote functional independence. Parental counseling is another important aspect of the occupational therapist with regards to optimizing parental support for improving functional abilities of child with CP. Different approaches to treatment are taken and considered. Since no child is the same, intervention for each child is specific and unique and related to

the skills the strengths and limitations of the child. To treat sensorimotor problems like spasticity, hypotonicity, non integrated primitive reflexes, immature postural or equilibrium reactions, sensory dysfunction and other issues like lack of age appropriate play behavior, difficulties in performing ADLs, cognitive and social impairments, Occupational therapists work on the following :-

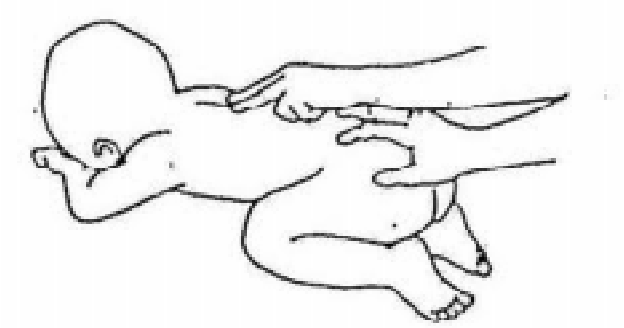
1. Development of Postural Control

Postural control is the ability to maintain balance and alignment while upright in space. The child with CP has poor postural control due to delayed or incomplete motor development, decreased voluntary control, lack of stability, difficulty in antigravity movements, lack of dissociative movements, exhibition of stereotypical movements with compensatory mechanisms. The development of postural control is also affected in presence of deficits in the organization of sensory inputs.

Therefore Interventions are aimed at:-

- Facilitation and development of antigravity movements, e.g. lowering a child from a sitting to supine position to facilitate neck flexion against gravity. Neck and trunk extension can be promoted by placing the child in prone positions over a wedge or large therapy ball
- Facilitation of automatic reactions like Righting, equilibrium and protective reactions, e.g. placing the child on unstable surfaces and moving the surface using variations in speed, ranges and rhythms. Reach out activities while sitting on ball, bolster or balance board.
- Facilitation of sensory organization. E.g. walking on different texture surfaces with various visual conditions like closed eyes or dim lights. Reaching out while swinging on different types of swings
- Facilitation of anticipatory control. e.g. catching and throwing a ball. Kicking a ball.

Facilitation of neck flexion while going from sit to supine



Stimulating back extensors to facilitate extension in prone position

Ocupational therapist's use Neurodevelopmental Treatment Approach which is aimed at facilitating and normalizing hyper- or hypotonic muscle reflexes using facilitation-inhibition mechanisms; reaction and movement patterns; and managing the specific reactions to the treatment of equilibrium.

2. Development of Hand Skills

Hand function involves the following components like reach, grasp, carry voluntary release, in hand manipulations, bilateral hand use. Often the CP child has impaired hand function skills due to problems in isolation of movements and tending to use total patterns, insufficient force application, improper timing of movements, limitations in trunk control and inability to bring hands in midline. Intervention aims at

- Proper positioning of the child. E.g. first consider the optimal position for eliciting the skills desired. Certain body positions can be used to elicit specific hand skills like Supine position for arm movements and visual regard of hands, prone position with forearm weight bearing for shoulder stability, side lying position for unilateral movements. While sitting at table for fine motor tasks the child should be seated at an appropriate table height on a stable chair with foot supports, with arms on table surface without elevating shoulders.
- Improvement of postural tone and control E.g. (refer to development of postural control above) and inhibition of tone using weight bearing or slow movement activities
- Development of hand skills by:-
  Promoting isolated arm and hand movements e.g. stabilizing proximal muscles (trunk and shoulder) to promote opportunity for the isolated hand movements.

Enhancement of reach, grasp, carry voluntary release, in hand manipulations, bilateral hand use

e.g. using pegs, beads or marbles of various sizes and shapes, clay activities, stringing and stacking activities

Proper positioning in sitting to promote hand functions

3. Sensory Integration (SI)

SI is the ability of the CNS to process information to make adaptive response to the environment. The CP child may have sensory integration problems which are:

- Sensory modulation problems
- Sensory defensiveness
- Sensory dormancy / Sensory registration problems
- Adaptive movement response problems
- Vestibular processing problems
- Sensory discrimination and perceptual problems
- Tactile discrimination
- Proprioceptive perception
- Visual perceptual
- Other sensory problems
- Praxis problems

Occupational therapists work with children who have sensory processing disorders, by engaging them in activities that promote sensory integration.

SI therapy in CP enables the child to make purposeful adaptive responses to sensory input in the environment. It refers to a therapeutic intervention which uses strong kinesthetic and proprioceptive stimulation to attempt to organize the CNS.

Guiding principles in SI:

- Sensory input can be used systematically to elicit an adaptive response
- Registration of meaningful sensory input is necessary before an adaptive response can be made
- An adaptive response contributes to development of sensory integration
- Better organization of adaptive responses enhances the child's general behavioral organization
- More mature and complex patterns of behavior are composed of consolidations of more primitive behaviors
- The more inner- directed a child's activities are the greater the potential of the activities for improving neural organization.

Intervention techniques should address the underlying sensory deficit and not the behavior. SI uses all senses but focuses primarily on the Vestibular, Proprioceptive and Tactile Senses together known as the power senses. A child's brain organizes sensory stimulation from touch and movement in order to learn and respond successfully to the environment. Vestibular stimulation includes linear, circular and rotatory movements like lying on the top of a large exercise ball while receiving rocking or bouncing movements and sitting inside an inner tube swing. Proprioceptive stimulation includes compression and traction like weight bearing activities like wheel barrow walking and jumping on trampoline and hanging from a trapeze. Tactile stimulation includes different textures stimulation like sand playing, brushing painting activities. Different types of stimuli influence a child's muscle tone and normalizing muscle tone enables a child to move and function well. Children with increased muscle tone typically benefit from calming sensory stimuli such as slow movement, rhythmic music, dim lights and a quiet environment. Children with low muscle tone typically benefit from erratic music, bright lights and irregular movement such as jumping.

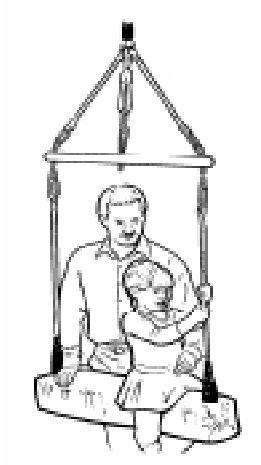


Fig 4 Vestibular stimulation provided in the form of swinging on a bolster swing

4. Development of Visual Perceptual Skills

Visual Perception is defined as the total process responsible for the reception (sensory functions) and cognition (specific mental functions).

Visual receptive Functions

- Acuity
- Accommodation
- Binocular fusion
- Steropsis- Binocular depth perception or three dimensional vision
- Convergence and divergence

Visual Cognitive Functions

- Visual Attention - alertness, selective attention, visual vigilance and divided or shared attention
- Visual memory- short term and long term
- Visual discrimination-
    - Object (form) perception- form constancy, visual closure and figure ground
    - Spatial perception-position in space, depth perception and topographical orientation
- Visual Imagery

CP children with Visual Perceptual problems demonstrate many problems like in ADLs e.g. difficulty in combing, tying laces, matching clothes or in Play e.g. difficulty in sports, cutting, constructing, doing puzzles.

Intervention focuses on visual perceptual training involving Developmental, Neurophysiologic, Sensory Integration and Compensatory approaches which helps in improving skills that limit function and also compensate for the limitation.

5. POSITIONING AND SEATING

Proper positioning can be achieved by use of handling techniques, pillows/ wedges, bracing, inserts etc to minimize the effects of abnormal tone and reduce secondary complications.

Evidence supports that children with CP should be fitted for wheelchairs that place them in a functionally safe position, which includes; a hip-belt, an AO, footrests, and a cutout tray, with the addition of a sloped forward seat to improve upper-extremity function.

Adaptive Devices

- Nonslip surface on chair to prevent slipping (e.g. Dycem)
- Bolster, rolled towel, or blocks for feet
- Adapted or alternate chair, stander
- Custom fitted wheel chair or insert

6. ACTIVITIES OF DAILY LIVING (ADLs)

Activities of Daily Living-ADL involve taking care of one's body such as toileting, bowel and bladder management, bathing personal hygiene and grooming, eating and feeding, dressing, functional mobility sleep and rest. Approaches to improve Performance of ADL involve Establishing, restoration, maintenance and Modification or Adaptation and Prevention and education.

Occupational therapists use many Behavioral techniques like chaining (backward and forward), shaping and reinforcements to teach ADL skills.

The following are the issues to be considered while applying the functional approach (Activities of Daily Living-ADL) in a combined intervention of child-centered activity and structural-developmental intervention respectively:

1. Developing a feeling of interest in performing daily activities and motivation
2. Experiencing learned functions by using daily life devices
3. Practicing self-care skills

4. Developing visual-spatial skills within the environmental setting
5. Developing creative self-expression through play, artistic activity, movement and other activities
6. Developing perceptual and visual-motor functions which are necessary for learning
7. Developing more complex play levels

- **Feeding**

  Evaluation should encompass the patient's visual, perceptual and cognitive skills, physical control of head, trunk and extremities, oral structures and ability to suck, masticate, and swallow. Also face and mouth sensitiveness, ability to sense temperature, facial muscles, interference of primitive reflexes like rooting, bite, sucking. Impaired oral reflexes like gag coughing reflexes, Outer oral motor assessment (facial expression, lip control and jaw control) and inner oral motor assessments (palatal functioning, tongue musculature and motor control) are conducted.

  The family's desires and expectations in regard to the child's feeding capacities should also be considered.

  Intervention in feeding involves positioning, handling and compensatory strategies.

  Proper positioning should be emphasized while feeding to promote oral motor function.

Appropriate positioning for feeding:

- Neutral pelvic alignment of trunk. Pelvic alignment is facilitated when the child s well supported against a flat back, on a flat seat and square on the buttocks with hip and knee in 90° of flexion.
- Good head, neck and shoulder alignment in slight neck flexion or in neutral.
- Chin tuck with the back of the neck elongated
- Providing the child with external postural stability enhances stability, good alignment and easy feeding.

After positioning the child, the therapist can use handling techniques to aid oral movements:

- Tapping or quick stretch, vibration to increase tone
- Deep and firm pressure to reduce tone
- The therapist can also use her hands and fingers to provide external support e.g. using one finger to promote chin tuck and the other under jaw for support from the side or from front

Oral motor stimulation to improve oral motor control

Adaptive devices

Non slip mats, wet towels, suction cups are used to stabilize eating utensils

Adapted cutlery may be more suitable, e.g. rocker knife, extended handle cutlery (e.g. foam handle on utensil), Adaptive drinking devices (e.g. cup with cut out rim)

Adapted chairs, tray attached to table

Practical tips

- Normalize child's tone as much as possible before beginning the feeding process
- Observe the child while he / she is eating.
- Make sure the child can see the plate, the food which is on it and the spoon bringing the food from the plate to her mouth.
- Talk to her / him about the process and let her / him see, feel and smell the food, feel the plate and the spoon
- The feeding person should be seated directly in front in order to maintain proper position
- Table should be positioned at axilla height and close to chest so distance from plate to mouth is reduced
- Chewing may need to be encouraged slowly and patiently by very gradually increasing the density of texture and later on the 'lumpiness' of the food offered.

- Encourage her / him to pick up food (such as cakes, biscuits, bread or fruit) in her hands and bring it to her mouth.
- Use different food temperatures and textures to increase child's awareness of what is in his mouth.
- Firm pressure is more acceptable to the child than light touch
- Placing a mirror in front of the child during mealtimes helps his feeding skills.
- Give the child the drink he likes best allowing him to make a choice when you teach him to drink
- Therapist can guide the extremity in the correct pattern if required

- **Dressing**

  Learning and participation in dressing is a major step in step in achieving independence. Even with limited motor and sensory skills dressing can be made easy by motivating the child to actively participate and reducing demands placed on the child.

Adaptations

Adaptive clothing

- Loose clothing free from unnecessary fastenings which should be limited to few layers
- Use of Velcro and elastic band clothing instead of buttons or zippers
- Front openings, Pullovers, large buttons, Stretchy clothing

Practical tips

- Work on undressing first, as it is an easier skill than dressing.
- Always put the clothes on the most affected part of the body first.
- Positioning child on floor is safer than on a chair.
- Techniques that inhibit spastic postures will facilitate the movements in and out of clothing, If her legs are bent before putting on socks and shoes it may help ease any stiffness in her ankles and feet and her toes are less likely to curl under.
- Encourage to stand up and hold on to furniture.

- **Bathing**

Achieving cleanliness is essential for maintaining good hygiene. Bathing can be made fun and a special bonding time with the caregiver. Occupational therapists can use bathing therapeutically to enhance motor and sensory skills.

Adaptive devices

Hand held Faucet

- Soap on a string or soap tied in a lofah or liquid soap
- Railing, grab bars
- Adaptive chairs
- Semi-inflated inner tube can be used as a positioning device
- Nonskid mats
- Hydraulic grab bars

Practical tips

- Positioning the child in a such way that normalizes his muscle tone as much as possible providing a sense of security, rather than challenge his sense of balance.
- Bath sponge wedge can be used to bathe an infant in both sitting and lying positions.
- Bathing can be time which can be used to teach various concepts for example, arm in, out, through: tub full, empty, wet, warm, cold and working on body parts identification. Thus bathing provides the child with many opportunities to learn cause-and-effect relationships, spatial relationships and tolerance of different sensations.
- The time following a bath can also be very productive. For example, a fast run with a terry towel can help the child learn to process sensations and can lead to better body awareness.

- **Toileting**

  Independent toileting is very important in achieving self maintenance. Toilet training can be quite challenge in a CP child as the child must be temperamentally and physically ready to accept toilet training as well as be able to understand the process in order to have any success.

Adaptive devices

Hand held jet sprays

- Use of Toilet paper
- Soap on a string or soap tied in a lofah or liquid soap

- Railing, grab bars
- Adaptive commodes or chair with hole placed over commode
- Footstools to support feet
- Use of straps to maintain safe and proper posture
- Nonskid mats

Practical tips

- A fairly casual, nonconfrontational introduction to the process will help reduce any stress related to toilet training.
- Visual break down of task on wall
- Habit training to go at same times to toilet
- Reinforce them for their success or for sitting patiently on the toilet and trying.
- Instruct caregivers to follow same routines at school

MOBILITY

Being mobile enhances a person's ability to learn, interact with others and participate in the community. For children with mobility impairments, a variety of mobility aids and devices are available to provide support, motion and access, as well as to enable them to lead active and fulfilling lives.

Mobility aids include:

- Canes
- Crutches
- Walkers
- Manual wheelchair
- Powered wheelchair with joystick, head switch, or sip / puff controls
- Grab rails and Railings

7. Play Skills

Play can be an important part of the learning experience and development of motor skills for a child with cerebral palsy. Selecting an activity should incorporate the child's interests and the skill he possess to participate in safe playing.

- Appropriate adapted equipment, such as wedges, bolsters, bean bags, CP chairs, may be used.
- Make certain your child changes positions frequently. Children should be encouraged to play on different surfaces and at safe heights
- Position child with both arms forward when playing with toys.
- Make certain that the child can see what is happening.
- Talk to the child at the child's eye level.
- Give the child ample time to respond
- Maintain a good balance between noisy, active play and quieter, less strenuous activities.
- Present toys that encourage your child to reach and grasp with the hand that is more difficult to use, but allow the child to use whichever hand he chooses.
- When teaching dressing to the child, put the more affected arm or leg into the clothing first.
- When interacting with your child, take the more affected hand.
- Encourage bilateral activities such as rolling clay or throwing a large ball,
- Provide multi sensory input toys that have interesting things to see, hear and feel.
- Avoid too noisy small play items
- Grade level of activities, gradually increasing its complexity
- A mirror can be a great aid in playing so that the child can get visual feedback
- Toys that have enlarged handles or knobs to grasp

Age Appropriate toys used to enhance play behaviors

0 -2 Years

- Play mat and frame with dangling toys
- Rocking and bouncing games
- Making lots of babbling and cooing noises
- Tickling games
- Building blocks
- Story books - with voice
- Peek a Boo games
- Banging on musical drums
- Imitation and turn-taking
- Playing with mirrors, press toys

- Unwrapping toys
- Lentils, rice and pasta in tubs to sit in, put your hands in or just throw about

1-3 Years

- Finger puppets
- Tunnel games
- Surprise bags full of toys and interesting objects for your child to find
- Pulling and pushing
- Object identification
- Pretend games with dollies and teddies
- Painting, using fingers or brushes
- Cars and trains
- Paper tearing and crumpling

2-4 Years

- Story books
- Clay modeling
- Ball games
- Obstacle courses
- Messy play (i.e. sand castle, finger painting)
- Sticking
- Stamping (leaves, crumpled paper prints etc.)
- Spot the difference
- Shape and color matching
- Make believe
- 'Simon says'-imitation games
- Imaginative Play e.g. Turning boxes into toys (such as castles, cars or space ships)
- Action rhymes
- Listening games
- Making music with home-made instruments

Games involving "spotting the difference", or pointing out which part of a picture or drawing does not belong, help the child's neurological development.

8. Hand writing Skills

Writing is a fine motor task required to compose stories, complete written examinations, copy numbers etc. It is a complex task which requires synthesis and integration of memory retrieval, organization, problem solving, and language and reading ability, ideation and graphomotor function. The building blocks of early handwriting are:

- Spatial and body awareness
- Postural control
- Visual perception
- Fine motor control
- Directionality
- Handwriting

Intervention focuses on

Neurodevelopmental approach: postural and limb preparation activities like jumping on a trampoline, pushups, bear walks etc to modulate tone

Biomechanical approach: sitting posture should be with feet firmly planted on the floor, height of table such that it should be 2inches above flexed elbow, paper position should be slanted so that it is parallel to the forearm of the writing hand, pencil grip should ideally be a dynamic tripod grasp, pencil with a wide diameter, various writing paper (lined, unlined, double lined, textured, margins)

Sensorimotor approach: involves controlling sensory input through selected activities to enhance sensory systems. An inclined, vertical or horizontal writing surface, writing tools like crayons, sketch pens, vibratory pens, and using different textures like sand, shaving cream, talcum powder

Adaptive writing equipment

Pencil Grippers and weighted pencils and handi-writers (soft elastic looped around the writing utensil and wrist that keeps the utensil in the appropriate space of the hand)

Adaptive writing paper that is color-coded, sticky or rough on one side or has larger lined area

9. Use of Assistive technology

The cerebral palsy assistive technology includes hardware and software which helps maximize their abilities to access information and services. It has the potential to increase abilities of person with disabilities and can lead to independent living.

- Typing Master Software
- Text to Speech Software
- Voice recognition computer applications
- On - Screen Keyboard setting, Magnifier

setting, Narrator setting, Mouse Keys Utilization and setting.

The objective is to see how the technology can be used by a person with disabilities to enable him or her lead a more purposive life with some skills to do work and at the same time too have time for relaxing and enjoying life.

Some Assistive Technology used in

WRITING are

- Pencil or pen with adaptive grip
- Adapted paper (e.g. raised lines, highlighted lines, and so on)
- Slantboard
- Type writer
- Portable word processor
- Computer
- Text to Speech Software
- Voice recognition computer applications
- On - Screen Keyboard setting, Magnifier setting, Narrator setting, Mouse Keys Utilization and setting.

Alternate Computer Access

- Keyboard with easy access or access DOS
- Keyguard
- Arm support (e.g. ergorest)
- Track ball, track pad, joystick with onscreen keyboard
- Alternate keyboard (e.g. Intellikeys, Discover Board, TASH)
- Mouth stick or head pointer with standard or alternate keyboard
- Head mouse or head master/tracer with onscreen keyboard
- Switch with scanning
- Voice recognition software
- Word prediction (e.g. Co:Writer) to reduce keystrokes

Composing Written Material

- Word cards, word book, or word wall
- Pocket dictionary or thesaurus
- Electronic or talking electronic dictionary, thesaurus, or spell checker (e.g. Franklin Bookman)
- Word processor with spelling and grammar checker
- Word processor with word prediction (e.g. Co:Writer) to facilitate spelling and sentence construction
- Talking word processor for multisensory typing
- Voice recognition software
- Multimedia software for expression of ideas (assignments)

Communication

- Augmentative and Alternative Communication (AAC)
- Communication board or book with pictures, objects, letters, or words
- Eye gaze board (Eye gaze communication)
- Simple voice output device (e.g. Big Mack, Cheap Talk, Voice-in-a-Box, Micro Voice, Talking Picture Frame, or Hawk)
- Device with speech synthesis for typing (e.g. Cannon Communicator, Link, Write:Out Loud with laptop computer)

READING, STUDYING AND MATH

Reading

- Changes in text size, spacing, color, or background color
- Use of pictures with text (e.g. Picture It, Writing with Symbols)
- Book adapted for page turning (e.g. page fluffers, 3-ring binder, cardboard in page protector)
- Talking electronic device to pronounce challenging words (e.g. Franklin Bookman)
- Scanner with talking word processor
- Electronic books

Learning and Studying

- Print or picture schedule
- Low tech aids to find materials (i.e., index tabs, color coded folders)

- Highlight text (e.g. markers, highlight tape, ruler)
- Software for manipulation of objects or concept development (e.g. Blocks in Motion, Toy Store). Consider alternate input device (e.g. switch or touch window)
- Software for organization of ideas and studying (e.g. Inspiration, Claris Works Outline, PowerPoint)
- Recorded material (books on tape, taped lectures with number coded index)
- Key guard to go over keyboard to help select the right keys and forearm supports to help stabilize upper extremity.

Math

- Abacus or math line
- Calculator, with or without print out
- Talking calculator
- Calculator with large keys or large LCD print out
- On screen calculator
- Software with templates for math computation (consider adapted input methods)
- Tactile or voice output measuring devices (e.g. clock, ruler)

RECREATION AND LEISURE

- Adapted toys and games (e.g. toy with adaptive handle)
- Use of battery interrupter and switch to operate a toy
- Adaptive sporting equipment (e.g. lighted or bell ball, Velcro mitt)
- Universal cuff to hold crayons, markers, or paint brush
- Modified utensils (e.g. rollers, stampers, scissors)
- Ergo Rest to support arm for drawing or painting
- Drawing or graphic program on computer (e.g. Kid Pix, Blocks in Motion)
- Playing games on the computer
- Music software on computer

ENVIRONMENTAL CONTROL

- Light switch extension
- Use of Powerlink and switch to turn on electrical appliances (e.g. radio, fan, blender, and so on)
- Radio or ultrasound remote controlled appliances

VISION

- Eye glasses
- Magnifier
- Large print books
- Screen magnifier (mounted over screen)
- Screen color contrast (e.g. CloseView)
- Screen magnification software (e.g. Closeview, Zoom Text)
- Screen reader (e.g. OutSpoken, Jaws)
- Braille Keyboard and Note taker (e.g. Braille N Speak)
- Braille Translation Software

HEARING

- Hearing aid
- Classroom amplification
- Captioning
- Signaling device (e.g. vibrating pager)
- Screen flash for alert signals on computer

10. Environment modifications

To ensure safe mobility and increase independence of the CP child, the physical environment in which he functions may need modifications in the form of:

Accessible Entrance / Exit - includes adding ramps, widening doorways, making entrance locks and door handles accessible and providing an emergency exit, considering lightweight doors or automatic door openers

Accessible Interior - includes widening hallways or interior doors, moving electrical switches and outlets related to door widening, reinforced wall for grab bars, railings

Accessible Bedroom - including widening doorways, making the closet accessible (lowered

shelves and hanging rods), relocating electrical switches and outlets

Accessible Bathroom - includes modifying design of commode, sink and cabinets, tub or shower, widening entrance, moving switches and outlets, faucet hardware

Other modifications- includes reinforced ceiling if need a lift, roll under sink in kitchen and bath, assigning workspace in close proximity to school supplies and equipment, modifying workspace or desk design and height

Splinting

The Occupational Therapist evaluates and recommends use of Upper extremity splinting to improve, maintain and prevent contractures and deformities or to improve functional movements. Hand splints to improve thumb abduction, wrist extension and functional positioning of digits are generally prescribed. E.g. resting pan splint is one which keeps the wrist in 20° to 30° extension, the metacarpophalangeal joints in 60° flexion and the interphalangeal joints in extension. This type of splint is used at night and during periods of inactivity with the hope of preventing deformity.

An example of a functional splint is an opponens splint to bring the thumb out of the palm of the hand, allowing for better grasp. This type of splint is used in everyday activities. However, it is still unknown whether a thumb abduction orthosis improves use and manual function of the affected hand in children with hemiplegia

Precautions during splint use

The caregivers should be educated about the wearing of the splints which includes donning and doffing of the splint, wearing schedule and care of skin and splint.

Non verbal children having poor sensations may not be able to report sensory problems occurring during wearing of the splint, so a thorough skin inspection should be taught to the caregivers

Starting with wearing of the splint for few minutes the wearing time should be gradually be increased to about 8 hours a day for static splints. Use of dynamic splints can be increased for additional more hours according to the child's tolerance.

Hand orthoses may inhibit the active use of the extremity. Hence it is important for the child to spend a certain amount of time in between the wearing schedule without the splints

OT intervention in school for CP children

OT in school focuses on child's ability to participate in functional school activities. A problem solving approach is used in identify the difficulties the child faces and to identify intervention strategies. Mental retardation and learning disabilities are some of the problems which the CP child may have to face.

Parents may require counseling regarding the type of school which best suits the child. The Occupational therapist is one of the members of a child's IEP (Individualized Education Program) team.

Strategies used in the school:-

- Reframe teacher's perspective. e.g. by explaining the issues and the underlying deficit the child seems to be facing
- Improve child's skills. e.g. use of practice worksheets
- Adapt the task e.g. use of keyboard to take down notes
- Adapt the environment. e.g. keeping visual distractions to a minimum
- Adapt the routine. e.g. extra time to complete worksheets.

Adaptive aids

Adaptive chair (CP chair)

Adaptive writing devices, aids and paper

Railings along staircase, classrooms and toilets

Adaptive commodes

Schedule boards, checklists timers, calendars

Vocational and prevocational Rehabilitation:

Vocational and prevocational activities are ones that help to prepare students for a future job they may pursue as they get older.

Pre-Vocational skills training: Activities are developed that will prepare an individual for employment. It includes support and training in behaviors related to following directions, attending to task, task completion, problem solving, and safety and assisting the person to adjust to the productive and social relationship demands of a work place.

In the classroom it is important to give children a

chance to experience what sorts of tasks they may encounter in the workplace. Some examples of these tasks are: sorting, putting objects together, alphabetizing, filing, data entry, and packaging items.

Vocational skills training

Prior to vocational skills training a Transition Programme can be conducted, in which the skills learnt already in the pre vocational training are transferred to vocational training. Occupational therapists specialized in Vocational rehabilitation, conduct workshops to give training in a variety of occupations like tailoring, greeting card making and pot painting exercise, soap and phenyl, pickle, card making, craft works etc.

Management of associated problems in CP

Seizures Management

Almost half of children with cerebral palsy experience seizures.

Practical Things to remember in event of any child having a seizure are:

- Do not attempt to hold the child still or to prevent physical movement; instead, make the environment safe so that she cannot be physically hurt.
- If the child is in a sitting position, or standing or walking when you observe the seizure, help her to lie down, so that she will not fall and get hurt. If at all possible, place the child on a blanket or protected surface. Position the child on her side, supporting the head. Move all sharp objects out of the way, as well as any furniture. Pad any sharp objects that cannot be moved, to prevent the child from getting hurt. Loosen clothing, especially in the chest and abdominal area.
- Do not put anything in the child's mouth. Do not interfere with the seizure or try to stop it.
- Allow the seizure to continue without interruption. Check the child for breathing. If the child has stopped breathing, clear the airway and perform mouth-to-mouth breathing.
- Turn the child on her side so the saliva can flow out of her mouth.
- After the seizure has run its course, let the child rest and be supportive.
- Although, medications are very effective in preventing or reducing seizures when given regularly, they may also produce a variety of side effects. For example, they may cause hyperactivity behavior, irritability, sleep problems, lethargy, depression, or sedation which may affect therapy.

Sexuality issues

Adolescents with cerebral palsy have delayed and prolonged puberty. They may develop precocious puberty as well. The therapist can guide the parents in preparing the child about puberty and the bodily and behavioral changes he may experience through books or pictures. Try and recognize the timing of sexual maturation and educating parents to provide age-appropriate sexual education. Pose questions about sexuality privately, using normalizing statements and open-ended questions helps the adolescent open up to discussions.

Conclusion:

Although there is no evidence that any specific approach in occupational therapy treats CP therapy to enhance functional skills is important. Increased Performance in all ADLs is very important which is specifically addressed by OT. Also, parental education and counseling along with home programs given by therapists help to address the needs of the family as a whole.

Psychological Intervention:

Introduction:

Cerebral palsy causes a lack of muscle control and motor coordination. Children with cerebral palsy are at an increased risk of developing emotional and behaviour problems. They may develop a feeling of learned helplessness and may feel socially isolated. Preschoolers with cerebral palsy are unable to explore the world around them and spend more time passively unengaged as compared to other preschoolers. By school age, social contact is reduced with the majority of free-playing time to non-play or other activities. As these children move on to adolescence, they emphasise on activities planned by adults than on spontaneous activities with peers. As, adults involvement is strictly limited. A lack of participation can lead to the development of physical, medical, cognitive,

emotional, or psychosocial secondary conditions with adverse outcomes in health, wellness, and quality of life.

Emotional Problems in Cerebral Palsy: About two thirds people with cerebral palsy suffer from severe emotional stress.

Depression: It is often seen that adults suffering from cerebral palsy have a sense of lack of emotional support, lack of coping skills and a very negative view of the future. Patients may suffer from pain which may lead to depression. However, it is seen that it may not be so much due to the severity of the disability but it may depend but would depend on how well they cope with the disability. Children and adults suffering from cerebral palsy may develop depression as being unable to control their body, embarrassment about their body in social situations and lack of information about their situation.

Anxiety: As adults with cerebral palsy may age they may develop age related issues like arthritis, bone fractures, chronic pain and fatigue. Due to these additional problems they may develop anxiety about their worsening condition and how their condition can limit their functioning. Patients may also develop sleep problems which in turn can contribute to anxiety and other emotional problems.

Low Self- esteem: Due to perceived physical limitations, loss of body control and medical condition, patients with cerebral palsy may suffer from low self-esteem. As, parents are over protective about their children, this in turn may lead to dependence and low self -worth in patients. Parents who engage their child in conversation about other topics, such as the child's likes, dislikes, achievements and ambitions, are likely to overcome this feeling of a lack of individuality.

Cognitive Deficits in Cerebral Palsy:

Learning Difficulties: Children with cerebral palsy may experience specific learning difficulties which may include short attention span, motor planning difficulties, perceptual difficulties and language difficulties. It is also seen that children suffering from cerebral palsy who display disruptive or avoidance behaviours and low self concept may have underlying learning issues. Students suffering from cerebral palsy may get tired quiet easily as they need to put more effort into concentrating on their movements and sequence of actions than others. Learning may also be affected by problems in fine motor and gross motor coordination and communication.

Mental Retardation: It has been estimated that around 65 percent of the individuals living with cerebral palsy also have some form of mental retardation. About 50% are full mentally retarded i.e. an IQ below 70. Because cerebral palsy and mental retardation can be co-morbid, they can contribute to emotional stresses as well. Learning disabilities may be present, depending on the area of the brain that was damaged. About a third of individuals with cerebral palsy have mild intellectual impairments, a third have moderate-to-severe intellectual impairments, and another third have normal intellectual functioning.

Behavioural Problems in Cerebral Palsy:

Behavioural problems and cerebral palsy usually correlate, depending on the degree of mental retardation. The child may have behavioural problems or emotional issues that in turn, may affect psychological development and their ability to have social interaction.

1. Frustration: Patients suffering from cerebral palsy may face difficulty in completing a task, which may lead to getting angry and discouraged about their condition. This problem can be overcome by helping them with the task and finishing it which would foster a sense of achievement.
2. Communication difficulties: Lack of ability to communicate efficiently can cause disturbance associated with behavioural problems. During such situations the children call for a lot of physical and mental stress to the parents. Excessive attention should be discouraged whereas the child should be kept involved by talking or just maintain eye contact.
3. Attention Deficit Disorder: Many of those who are immobile let their attention wander. In such cases, there should be minimal distraction while teaching them such as teaching them in the corner of a room. The television sets and other modes of distraction should be kept away, in order to increase their attention span.

Employment Issues: Several studies suggest that about 30% to 50% of adults suffering from cerebral palsy were competitively employed. Findings of a study suggest that speech deficits can lead to verbal

difficulties and could lead to decrease in competitive employment. It is often seen that individuals with hemiplegic cerebral palsy had regular jobs as compared to those with other types of cerebral palsy. The severity of cognitive and motor impairment, seizure disorder and types of cerebral palsy were predictive factors in competitive employment. Cognitive ability is a very important factor in employment.

Psychosocial Factors: The development of both intelligence and personality relies heavily on developmental experiences and the opportunity for self-expression. The child may find it easier to withdraw towards social isolation. They should be encouraged to take independent decisions and physical tasks. Early choices can be made by the child regarding the clothes to wear or which task to do first.

Parents need to resolve their own way the emotional impact of the child's disability. Most parents feel inadequate, ignorant and relatively helpless at being unable to remedy the situation for the child. They need help in feeling good about themselves before they can effectively guide the child towards self -acceptance as an adequate human being. Parents need guidance to provide themselves with opportunities to rest and renew their energies.

Psychological Testing:

Children with cerebral palsy might find it difficult to respond to the tests that are timed or that require manipulation of objects, such as some of the subtests of Wechsler. Alternative tests such as Pictorial Teat of Intelligence, the Columbia Mental Maturity Scale or the Peabody Picture Vocabulary Test are recommended. There are various measures of gross motor functioning amongst these are Bruininks - Oseretsky Test of Motor Proficiency, the McClenaghan and Gallahue Checklist and the Vulpe Assessment Battery. Raven's Coloured Progressive Matrices is a fast, easy-to-administer test able to obtain a measure related with linguistic, visuo-perceptual, and memory cognitive functioning in persons with CP despite their motor and speech disorders.

Psychological Treatment:

Education and vocational preparation come into the foreground by school age. Concern with the physical disability should not distract attention from the emotional and social needs of childhood and adolescence. Disabled youngsters need the same variety of life experiences as all other children to develop emotional resilience, personal determination, and social skills. As the child with cerebral palsy grows older, the need for and types of therapy and other support services will continue to change.

Neuro-cognitive therapy: A new approach to treating cerebral palsy from Snowdrop. It is based upon two proven principles. (1) Neural Plasticity. The brain is capable of altering its own structure and functioning to meet the demands of any particular environment. Consequently if the child is provided with an appropriate neurological environment, he will have the best chance of making progress. (2) Learning can lead to development. Lev Vygotsky proposed that children's learning is a social activity, which is achieved by interaction with more skilled members of society.

Counselling and behaviour therapy, for emotional and psychological challenges may be needed at any age, but is often most crucial during adolescence. Behaviour therapy is often used to enhance child's ability and discourage destructive behaviours. Behaviour therapy might include planning activities that are rewarding which could provide a sense of accomplishment; use of reinforcements can encourage a behaviour change, enhance learning and solidify gains. For example behaviour therapy might include hiding a toy inside a box to reward a child for learning to reach into the box with his weaker hand. In other cases the therapist may deal with unhelpful or destructive behaviours like biting, or hair pulling by selectively presenting a child with rewards such as praises or rewarding with extra play time.

Teaching relaxation strategies such as systematic desensitization, abdominal breathing to the client or the caregiver can help reduce stress and anxiety and effect behavioural change. Token economy is an effective way to reinforce a positive change. Aversion therapy i.e. to reward rather than punish on negative consequences can help enhance self - esteem. Expressive therapies are usually used with people who have difficulty verbalizing their feelings such as art, music, poetry, etc which could help freeing and empowering oneself. Sometimes children with cerebral palsy can become violent or aggressive, resorting to things such as biting or hair-pulling to they can be helped to release their

aggression and frustrations, by either being vocal or, if the child is able to control his or her hand enough, perhaps drawing or writing.

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Ch.4 Head Injury

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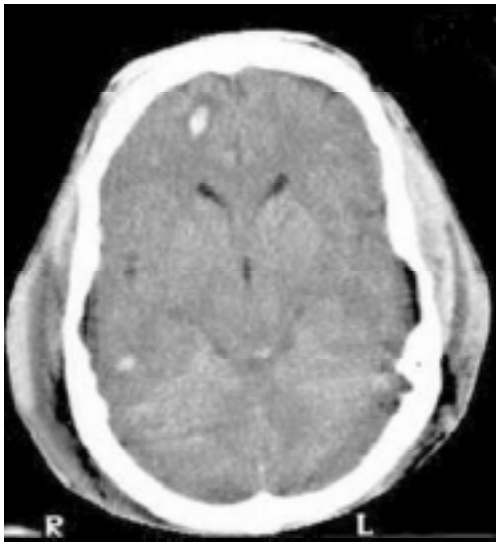
Head injury includes injuries of the scalp, skull, or brain. The most common cause of head injury is trauma which may be road traffic accident, fall or physical assault.

Head injuries can be classified in to closed or open depending on whether the duramater is intact or breached. A closed head injury is one in which duramater remains intact. There may be injury to the scalp or cranial bones. An open head injury occurs when a penetrating object breaches the duramater.



A head injury may result in only scalp wounds. In more severe cases it may result in skull fracture and direct or indirect injury to the brain. Injury to the brain may be in the form of concussion, contusion, haemorrhage or haematoma.

Patients with head injury may be aymptomatic at the time of injury and may develop symptoms over a few days. Symoptomatic patients may present with nausea, vomiting, headache, confusion, drowsiness or seizures. There may a lucid interval during which the patients remains conscious after head injury and deteriorates later. Unconscious after a head injury even for a short period is not normal. The patient may have other symptoms like difficulty concentrating, increased mood swings, lethargy or aggression, and altered sleep habits. These symptoms can occur in concussion type of head inury and can easily be missed.

The physical examination and the history of the exact details of the injury are the first steps in caring for a patient with head injury. Patient should be assessed for symptoms suggestive of severe head injury- severe headaches, fluid draining from nose / mouth / ears, loss / alteration of consciousness, confusion, drowsiness, slurred speech, blurred vision, stiff neck or vomiting, paralysis.

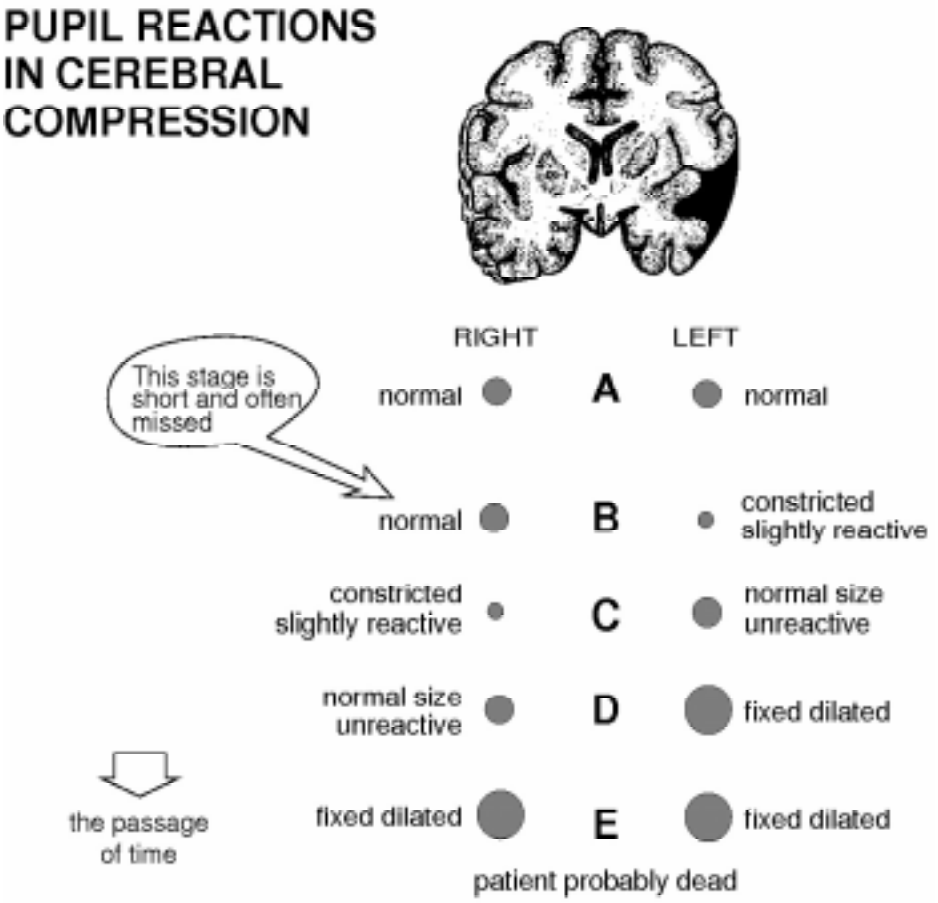
The Glasgow Coma Scale (GCS) should be used to assess the level of consciousness, severity of head injury and to determine the prognosis.

- Minor head injury: GCS 13-15; mortality 0.1%
- Moderate head injury: GCS 9-12; mortality 10%
- Severe head injury: GCS <9; mortality 40%.

The GCS should be assessed periodically to determine the neurological status of the patient.

Examination of pupil forms a vital part of neurological assessment. The response of pupils to light and their size should be examined. Anisocoria, unequal pupil size, is a sign of serious head injury. Pupil not reacting or reacting sluggishly to light is suggestive of raised intracranial pressure.

Neuroimaging helps in determining the diagnosis, prognosis and in deciding the treatment. Non contrast Computerized tomography (CT) scan of the head can show bleeding and swelling in the brain. It can also evaluate bony injuries to the skull



and bleeding in the sinuses of the face associated with basilar skull fractures. CT does not assess brain function, and patients suffering axonal shear injury may be comatose with a normal CT scan of the head.

Management of head injury

Medical Management:

Acute management:

Immediate resuscitation including assessment and stabilization of the airway, breathing and circulation is a priority in the management of acute head injury. Cervical spine should be stabilized by head and neck immobilization. After resuscitation and stabilization, secondary brain injury can be prevented by keeping- mean arterial pressures above 90 mm Hg and arterial oxygen saturation greater than 90%. Urgent non contrast CT scan is done to assess the brain damage and hemorrhage (hematoma).

Intracranial pressure reduction and monitoring is critical in the management of head injury. Increase in ICP reduces cerebral blood flow, causes cerebral compression and is an independent predictor of poor outcome. The following ICP reduction strategies should be used if the ICP is above 20-25 mm Hg- hyperventilation, intravenous Mannitol, CSF drainage, hypertonic saline, high-dose barbiturate therapy can be used if the intracranial pressure does not respond to the above conventional treatments. Caution is required as barbiturate may reduce blood pressure.

In addition to reducing ICP, maintaining cerebral perfusion pressure is critical in the management of head injury. For adequate perfusion, cerebral perfusion pressure should be maintained at 50-70 mm Hg. [1] With ICP reduction strategies, the cerebral perfusion pressure may fall and should be maintained with- volume expansion and vasopressors. Normal saline is preferred over albumin. [2]

A systematic review has concluded that hypothermic therapy is of no benefit in patients with head injury. [3] A recent study has shown that very early hypothermia induction may improve outcomes in patients with surgically-evacuated hematomas and worsen outcomes in patients with diffuse head injuries. [4]

Neuroprotection is the administration of some agent, which should reverse some of the damage or prevent further damage. [5] The majority of neuroprotective agents are antioxidants. Role of neuroprotective agents in improving the outcomes of patients with brain injury is not well established. Agents with neuroprotective activity in brain innjuries include-

Nimodipine has been shown to reduce mortality and severe disability when given acutely in patients with head injuries and traumatic subarachnoid hemorrhages. [6] Magnesium chloride did not show benefit in clinical studies with moderate and severe head injury. [7] Intravenous progesterone has been shown to reduce 30-day mortality in patients with moderate and severe head injury. [8] Cyclosporin is an inhibitor of mitochondrial membrane permeability. Dexanabinol, a weak N -methyl-D-aspartic acid (NMDA) antagonist, showed no efficacy in outcome improvement in a recent trial, when given within 6 hours to patients with severe closed head injuries. [9] Rosuvastatin given in the acute phase of moderate head injury has been shown to reduce amnesia. [10] Creatine has shown to reduce cortical damage primarily through stabilizing mitochondrial functioning in pre-clinical studies. [11] Melatonin is a free radical scavenger, and has shown neuroprotective effect by significantly reduced levels of lipid breakdown products in pre-clinical studies. [12]

Phenytoin is efficacious in controlling early posttraumatic seizures. Some patients may develop an allergic reaction with this drug. [13] Posttraumatic seizure prophylaxis should be discontinued after 1-2 weeks unless further seizures supervene. [14]

Patients with head injury are in hypermetabolic state and therefore in need of adequate nutrition early. Early nutritional therapy has shown to reduce mortality in severe head injury. [15]

The risk of deep venous thrombosis is increased in head injury as coagulation parameters are altered. [16] Early DVT prophylaxis (with enoxaprin / heparin) is found to be safe in patients with head injury. [17] The risk of IC Bleed extension should be balanced with the benefits of thromboembolic prevention.

Steroids have found to be of no benefit in the treatment of acute head injury and may be associated with higher mortality. [18]

Long-term management: Long-term management of head injury includes management of spasticity, cognition enhancing medications and management of psychiatric complications.

Dantrolene, baclofen, diazepam, and tizanidine can be used orally for hypertonicity. Baclofen can also be used intrathecally. Botulinum toxin has been shown to decrease hypertonia in patients with head injuries. [19]

The cognition enhancing medications include methylphenidate, donepezil, levodopa. Methylphenidate has shown promise by improving motor outcomes and attention in patients with head injuries. [20] Donepezil has shown improved visual and verbal memory as well as attention in patients with head injury. [21, 22] Levodopa has been shown to produce cognitive improvements in patients with head injury. [23]

The management of psychiatric complications like emotional lability and depression are an integral part of long term management of head injury. Selective serotonin reuptake inhibitors have shown to be effective in pathologic laughing and crying. [24] Sertraline has shown efficacy in depression in mild head injury. [25]

Surgical Management:

The factors that help in decision making regarding the need for surgery include age, neurological status, mechanism of injury and pupillary reactions. The major questions addressed by the surgeon are:

1) Is the conscious level depressed?
2) Are there other signs of raised ICP (eg oculomotor palsy)?
3) Does the scan show a haematoma or contusion with mass effect (midline shift / ventricle effacement / dilatation of contralateral ventricle / loss of basal cisterns / sulcal effacement)?
4) Is expansion of the mass anticipated (eg contusions)?
5) Where is the mass (temporal lobe lesions are considered dangerous due to the association with uncal herniation compressing the midbrain)?

These questions must be appropriately addressed rapidly and when required the surgery should be performed with an appropriate sense of urgency.

1. **Extradural Haematoma:** The indication of surgery for EDH include-

a. EDH greater than 30cm3 should be surgically evacuated regardless of patient's GCS.

b. An EDH less than 30cm3 and with less a 15-mm thickness and with less than a 5-mm midline shift in patients with a GCS score greater than 8 without focal deficit can be managed nonoperatively with serial CT scanning and close neurological observation in a neurosurgical centre.

It is strongly recommended that patients with an acute EDH in coma (GCS score < 9) with anisocoria undergo surgical evacuation as soon as possible. There are insufficient data to support one surgical treatment method. However, craniotomy provides a more complete evacuation of haematoma.

2. **Subdural Haematoma:** The indication of surgery for SDH include-

a. An acute SDH with a thickness > 10 mm or a midline shift > than 5 mm on CT scan should be surgically evacuated, regardless of GCS score.

b. All patients with acute SDH in coma (GCS score < 9) should undergo ICP monitoring.

c. A comatose pt with SDH <10-mm thick and a midline shift < than 5 mm should undergo surgical evacuation of the lesion if the GCS score decreased between the time of injury and hospital admission by 2 or more points on the GCS and/or the patient presents with asymmetric or fixed and dilated pupils and/or the ICP exceeds 20 mm Hg.

In patients with acute SDH and indication for surgery, surgical evacuation should be performed as soon as possible. If surgical evacuation of an acute SDH in a comatose pt (GCS < 9) is indicated, it should be performed using a craniotomy with or without bone flap removal and duroplasty. In extreme cases a "burst" lobe may be evident. Such severe haemorrhagic contusions may necessitate resection of brain in the form of a lobectomy. Brain resection requires rapid but careful surgery preserving the middle and anterior cerebral arteries.

3. **Traumatic Parenchymal Lesions:** The indication of surgery for traumatic parenchymal lesions include-

a. Patients with parenchymal mass lesions and signs of progressive neurological deterioration referable to the lesion, medically refractory intracranial hypertension, or signs of mass effect on CT scan should be treated operatively.

b. Patients with GCS scores of 6 to 8 with frontal or temporal contusions > 20 cm3 in volume with MLS of at least 5 mm and/or cisternal compression on CT scan, and patients with any lesion greater than 50 cm3 in volume should be treated operatively.

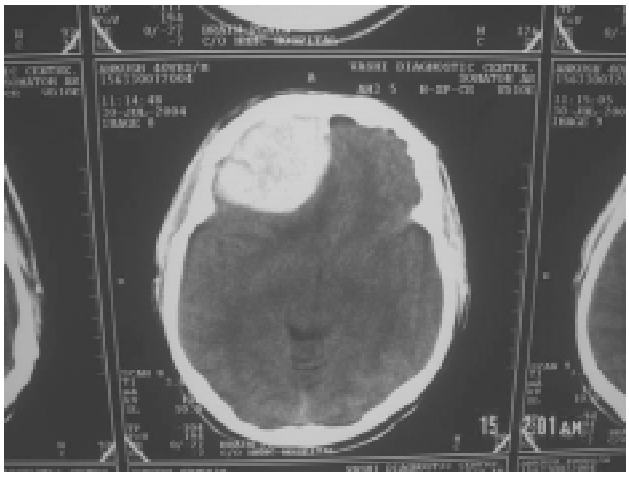
Craniotomy with evacuation of mass lesion is recommended for those patients with focal lesions and the surgical indications listed above. Bifrontal decompressive craniectomy within 48 hours of injury is a treatment option for patients with diffuse, medically refractory post traumatic cerebral edema and resultant intracranial hypertension. Decompressive procedures, including subtemporal decompression, temporal lobectomy, and hemispheric decompressive craniectomy, are treatment options for patients with refractory intracranial hypertension and diffuse parenchymal injury with clinical and radiographic evidence for impending transtentorial herniation.

4. **Posterior Fossa Mass Lesions:** The indication of surgery for traumatic parenchymal lesions include-

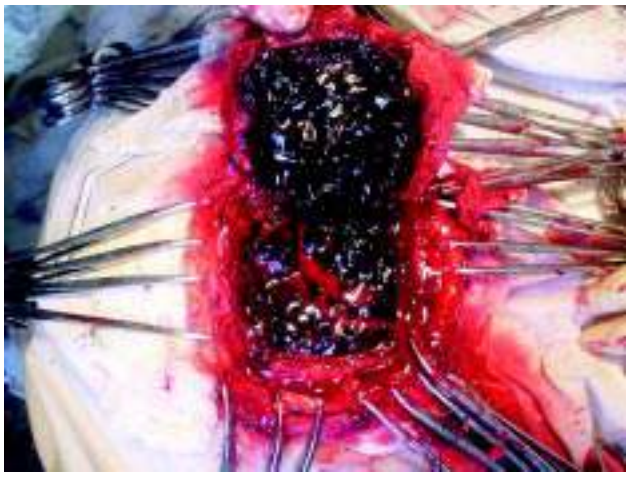
a. Patients with mass effect on CT scan or with neurological dysfunction or deterioration referable to the lesion should undergo operative inter- vention. Mass effect on CT scan is defined as distortion, dislocation, or obliteration of the fourth ventricle; compression or loss of visualization of the basal cisterns, or the presence of obstructive hydrocephalus.

b. Patients with lesions and no significant mass effect on CT scan and without signs of neurological dysfunction may be managed by close observation and serial imaging.

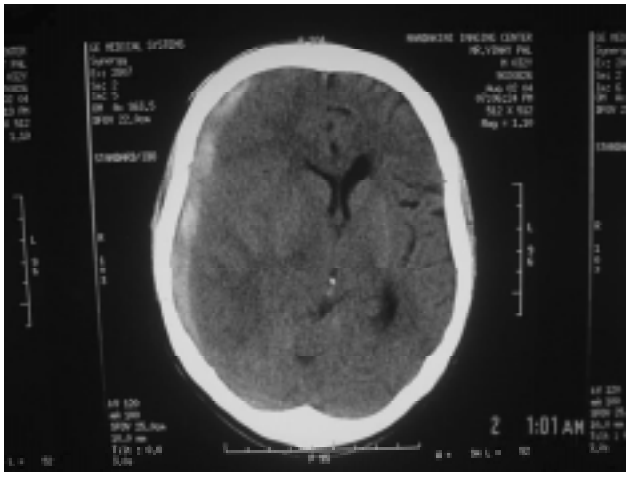
In patients with indications for surgical intervention, evacuation should be performed as soon as possible because these patients can deteriorate rapidly, thus, worsening their



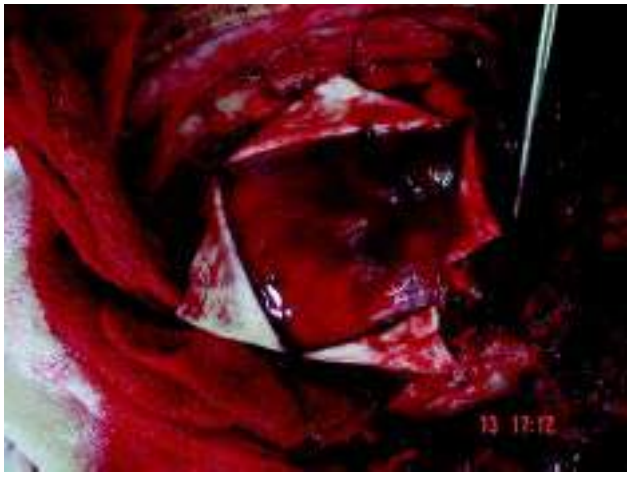
Compted Tomography brain (axial view) showing a large biconvex hyperdense lesion in the right frontal region suggestive of extradural haematoma.



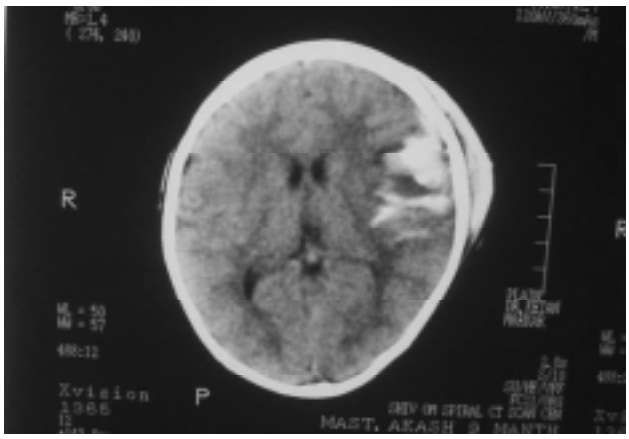
Intraoperative picture showing large extradural haematoma below the craniotomy flap.



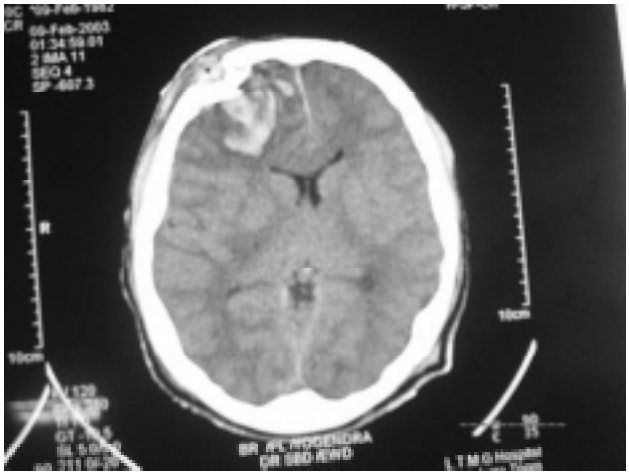
Compted Tomography brain (axial view) showing a large concavo-convex hyperdense lesion over the right frontoparietal region with mass effect suggestive of subdural haematoma.



Intraoperative picture showing large subdural haematoma on opeing the dura.



Compted Tomography brain (axial view) showing a large hyperdense lesion in the left frontal region suggestive of haemorrhagic contusion.



Compted Tomography brain (axial view) showing a right frontal depressed fracture with underlying contusion.

prognosis. Suboccipital craniectomy is the predominant method reported for evacuation of posterior fossa mass lesions, and is therefore recommended.

5. **Cranial Fractures:** The indication of surgery for cranial fractures lesions include-
   a. Patients with open (compound) cranial fractures depressed > the thickness of the cranium should undergo operative intervention to prevent infection.
   b. Patients with open (compound) depressed cranial fractures may be treated nonoperatively if there is no clinical or radiographic evidence of dural penetration, significant intracranial hematoma, depression greater than 1 cm, frontal sinus involvement, gross cosmetic deformity, wound infection, pneumocephalus, or gross wound contamination.
   c. Nonoperative management of closed (simple) depressed cranial fractures is a treatment option.

   Early operation is recommended to reduce the incidence of infection. Elevation and debridement is recommended as the surgical method of choice. Primary bone fragment replacement is a surgical option in the absence of wound infection at the time of surgery. All management strategies for open (compound) depressed fractures should include antibiotic.

Speech Affection in Head injury:

Patients with head injury present with a myriad of problem which include language problems, speech difficulty, cognitive issues and personality disorder depending upon the area of the brain affected. The left hemisphere damage affects language to a greater extent as compared to right hemisphere damage which affects the pragmatics of language. Individuals with left sided brain damage present with problems of understanding spoken or written language, difficulty in expressive language, alexia, agraphia and naming problems.

An individual with a mild traumatic brain injury may present with poor speech clarity (dysarthria) or even milder being the cognitive issues like affected attention and concentration, difficulty in problem solving issues, and changes in personality which is just a tip of the ice berg.

Management explained in Speech Rehabilitation Chapter.

Physiotherapy In Head Injuries :

A Traumatic Brain Injury (TBI) is a devastating and life changing event for the individual and his or her family. A TBI leads to a much more wider range of conditions than from sustaining any disruption to the vascular supply of the brain making it one of the most challenging conditions for physiotherapists. There is a multitude of issues with these patients as affection for the different parts of brain will be responsible for their own aspects of impairments and secondary impairments. Patients with a moderate to severe TBI require extensive rehabilitation during the different stages of the recovery & physiotherapist will be involved in the most of them depending upon the sensory motor deficit & their recovery. Physical recovery does not always go hand in hand with the cognitive & behavioral recovery. One of the deficits may show very good recovery without any in other during different stages and the durations.

Many traumatic brain injury (TBI) survivors are left with significant disability. The brain is, however, very adaptable and, with the correct physiotherapy input, recovery can take place over a period of years. People often witness a rapid recovery in the first few weeks following a TBI, followed by a slower recovery over the following years.

To gain the maximum recovery, physiotherapy should be continued until patient's full potential has been reached. With the correct physiotherapy input and advice patient has lot of potential to long term improvements regaining as much movement and function over the years.

Physiotherapy in TBI can help to:

- prevent or improve the respiratory distress to minimise / prevent secondary brain damage
- prevent the skin, soft tissues, joints dysfunction while unconscious or bed ridden
- reduce muscle spasms, pain and stiffness
- increase strength
- retrain normal patterns of movement
- increase affected arm and leg function
- initiate the ability & do independent ADLs from roll / move in bed / sit to stand and walk
- improve balance and walking
- increase energy levels

- increase independence and quality of life
- decrease risk of falls

The physiotherapist must follow the physiotherapy examination before the interventional planning. Interventions required will be as per the motor control, cognitive, behavioral or any other deficits presented in a particular patient. The interdisciplinary rehabilitation team offers a unique opportunity for physiotherapist to understand the patient better and so to do the best to reach the maximum functional recovery. Individual tem member contribute their expertise in their specialized area results in the enhancement of the team's overall effectiveness. Communication & open mindedness are the key to any team. All the members must share their skills & findings with the whole team & be willing to learn from each other to promote optimal return of function. The Physiotherapist must be willing to share a unique knowledge of motor control, but be open to learn from other team members which will lead to a consistent and complete approach to care.

Any one or some of the rehabilitation team members may play more prominent role in a patient depending upon the area of deficit he or she may be having due to TBI and stages of recovery also decide the dominance of roles.

SEQUELAE OF TBI

Though physiotherapists are primarily concerned with the treatment of neuromuscular impairments, the associated cognitive and behavioral changes become the hindrance in getting back the functions and abilities.

Various aspects of impairments in TBI seen are as follows:

Neuromuscular :	Abnormal tone of spasticity
	Primitive postures - Decorticate
	Decerebrate rigidity
	Alterations in sensation - light touch, pain, deep pressure and temperature.
	Impairments in proprioception and kinesthesia
Cognitive :	Coma / Vegetative state / Minimally Conscious State (MCS) / Stupor / Obtunded
	Disorientation
	Memory deficits
	Post traumatic amnesia (PTA)
	Declarative memory / & Procedural memory
Behavioral :	Sexual disinhibition
	Emotional disinhibition
	Apathy
	Aggressive disinhibition
	Low frustration level tolerance
	Depression
Communication:	Expressive or Receptive aphasia
	Deficits in reading comprehension, written comprehension, language skill
	Dysarthria
Visual-Perceptual:	Visual impairments-hemianopia, very rarely cortical blindness.
	Perceptual deficits - spatial neglect, apraxia, spatial relations syndrome, somatagnosia and right-left indiscrimination
Swallowing :	Dysphagia caused by damage to cranial nerves
	Motor control impairments, apraxia, and poor postural control.
Indirect impairments :	Skin breakdown or bedsores
	Soft tissue contractures
	Deep Vein thrombosis
	Heterotopic ossification
	Decreased bone density
	Muscle atrophy
	Decreased cardio pulmonary endurance
	Infections

ASSESSMENT

The first step in beginning an examination is to conduct a complete review of the case paper of the patient before actually seeing the patient as he / she may not be medically stable & to know about the complications as well as the precautions to be taken during the examination & subsequent treatment. Patient's medical status may be dynamic at these stages, it is important to check with the intensivist / anesthetist / house surgeon / nurse before beginning any treatment session. Owing to the possibility of various infections, physiotherapist should always observe precautions & will require wearing gowns, gloves, and masking when treating the patient.

Initially, should focus on observing the patient & response to stimuli to know:

- What posture is the patient in? Is there evidence of primary postures or reflexes?
- Are the patient's eyes open or closed?
- Is the patient able to respond to auditory or visual stimulation?
- Is the patient able to vocalize?
- Does the patient exhibit any active movement? Is the movement purposeful or nonpurposeful?
- Does the patient react to tactile/ painful stimulation?
- Do the patient's vital signs change when external stimulation is presented?

Respiratory State

Respiratory distress and, therefore, inadequate ventilation may be caused by:

1. **Obstruction of the airway** due to inhalation of blood, vomitus, false teeth, foreign body; the tongue flopping, jaw fractures
2. **Depression of the respiratory centre due** to damage to the respiratory centre in the brainstem
3. **Associated injuries** like primary damage to the chest wall & lung tissue

Neurological examination with Consciousness level

Consciousness assessment is very important in TBI.

Glasgow Coma Scale(GCS) is the most accepted objective examination. Any alteration in response can be seen easily if the observations are plotted on a graph.

-movement of limbs (lateralizing & focal signs)

-pupillary light reflex

Cardiovascular Respiratory State

Pulse & Blood Pressure: Hypotension will lead to reduced cerebral perfusion & neurological deterioration respiratory state.

If it is not medically contraindicated, the examination should include an attempt to make the patient sit on the edge of the bed with assistance. The therapist should monitor the vital signs & document any changes in vital signs as well as in muscle tone, head or trunk control.

Associate injuries

Many patients with a HI have additional injuries. These injuries may be relatively minor & can wait attention, or they may demand urgent attention, i.e. chest injuries, abdominal bleeding from damaged viscera, fracture dislocation of the cervical spine, fractures of the face, ribs,

Often it may require many sessions to complete the entire examination. Signs of progress or regression should be carefully monitored.

From the examination, an assessment of severity of the injury can be made.

PROGNOSIS & GOAL SETTING

The data gathered during the initial examination is used to evaluate the patient's status & develop a prognosis & plan of care. Based on the findings from the initial examination, impairments (primary & secondary) contribute to the patient's functional limitations.

Brain injury leads to a wide range of cognitive, motor & neurobehavioral impairments, so, it is difficult to establish & predict long term outcomes & set goals for these patients. When setting goals, the patient, family, caregivers, & entire interdisciplinary team should be consulted. The patient's potential for cognitive & behavioral improvement will greatly affect future motor ability & social participation.

Though it is difficult to use general guidelines to predict with precision for any individual patient due to widespread neurological damage & range of resulting impairments, initial severity of injury as measured on GCS, duration of coma and length of Post Traumatic Amnesia (PTA) may be used as predictors of outcomes.

The majority of subjects who experience a coma of less than 1 week duration, get either moderate disability or a good recovery, whereas, when coma is greater than 2 weeks, possibility is for moderate or severe disability.

The majority of subjects with PTA lasting 4 weeks or less have a good recovery or moderate disability. If it lasts more than 12 weeks, then most of the subjects show moderate to severe disability.

PATIENT CARE : PHYSIOTHERAPY MANAGEMENT

ACUTE STAGE

The main priority at this stage is to ensure the patient is medically stable. Proactive management at this early stage, of potential secondary complications, will enhance patient outcomes.

At this stage, the goals & anticipated outcomes will be:

- To handle carefully & immediately known or suspected raised ICP
- To treat an unconscious patient as a whole person & not a series of joints & muscles
- To facilitate arousal, increase the level of alertness & physical function
- To reduce the risk of secondary impairments
- To prevent building up respiratory secretions and enhance oxygenation of the brain
- To work on the response by stimulating the sensory system
- To work for the normalization of the muscle tone
- To improve joint mobility & control
- To provide early family education & support- by means of involvement in aspects of the patient's care, providing time out to discuss the patient's progress & potential outcomes of interventions, advice on support groups and sources of information.

The unconscious patient is best nursed in an intensive care unit as the high levels of care is required. The patient may be on a ventilator, monitoring of ICP may be ongoing; there may be weight bearing precautions & movt. restrictions due to musculoskeletal injuries, open wounds or presence of bone fixators. Frequent neurological observations are required to know any deterioration, if any.

Chest care: Hypoxia causing further brain damage is an important cause of neurological deterioration. Cerebral hypoxia may be produced by: airway obstruction from inhalation of blood or vomit or head position; depression of the respiratory drives; associated chest injury, infection or pulmonary edema; & epileptic fits.

Patients who are managing to maintain their own airways & adequate oxygenation may not require intubation & mechanical ventilation. They are nursed in a position which allows the head to be raised at 30º & in side lying to prevent inhalation of secretions. Regular turning will prevent consolidation & collapse of lungs & the pressure sores. Positioning & turning may be difficult, if there are associated injuries which require splinting, such as plaster casts or traction.

If the patient is unable to maintain his own airway, a cuffed endotracheal (ET) tube is required. This may be sufficient to obtain sufficient oxygenation & remove any retained secretions to stop any further deterioration. Any evidence of respiratory inadequacy, need of artificial ventilation (IPPV) is required.

It is important to keep the chest clear of secretions as patchy collapse with sub clinical hypoxia & hypercarbia is probably more damaging to the brain than any other factor. Due to the traumatic brain a small rise in IC volume may cause a dramatic rise in ICP. Various physiotherapeutic procedures for the chest care such as postural drainage, shaking, bag squeezing, prolonged suction & coughing may precipitate the raise in ICP. Therefore, the patient must be protected during chest physiotherapy; effective treatment can be carried out without provoking large increases in ICP. This can be achieved by drug therapy for sedation, analgesia & muscle paralysis.

This is best carried out to coincide with turns & before bolus feeds, a continuous feed should be switched off. The chest is vibrated on the expiratory phase of breathing. Tipping the patient with his head down is not advisable due to the risk of elevating ICP. When suction is required to remove secretions, nasal suction is contraindicated if CSF Rhinorrhoea is present: this indicates a CSF leakage through skull fracture with a risk of ascending infection.

The frequency of treatment depends on the individual patient, but even if there are no obvious lung problems, treatment is given to prevent the unnecessary complications of retained secretions.

Techniques of shaking & vibration are best used while the patient is being ventilated by bag squeezing, i.e. off the ventilator. Careful suction via the ET tube should clear secretions.

Patients having controlled IPPV usually need continuous sedation using anesthetic agents & analgesics. A bolus dose of sedation is given prior to physiotherapy. Total treatment time is a crucial factor & should be kept at minimum, shorter

sessions, more frequently should be given as per the requirement. Suction, in particular, is a relatively dangerous procedure because of its effect on ICP.

General care: Handling these patients is often made more difficult from spasticity with the release of primitive neuromuscular activity such as reflex patterns. Primitive postures may include those associated with decorticate (abnormal flexor response) or decerebrate (abnormal extension response) rigidity.

Patients with TBI are at a high risk of developing contractures owing to prolonged periods of immobilization & abnormal reflexing posturing. Techniques to preserve mobility & inhibit primitive responses are employed as far as possible. These, combined with careful positioning after treatment sessions & every turn, should help reduce the problems to which hypertonicity leads at a later stage. Never assume that these patients are unaware of what is happening around them. Talking to them, telling them what you are doing & why, asking them to try & help, provide important stimuli from other sensory pathways along with the stimuli from movement & handling. As consciousness returns, the clinical features resulting from the head injury are evidently seen, but the predominant problem of spasticity will be evident early on.

Unless there are reasons to delay the start of an active physiotherapy, associated injuries such as limb fractures, chest injuries, patients should be introduced to changes of position out of bed & movement early on. Aim for normal sitting posture, avoiding tipping chairs.

The unconscious patients will also require to be treated for their oromotor impairments for oral hygiene, sensory motor training, chewing, swallowing, as well as speech therapy.

By Passive Range of Motion(PROM) exercises, joint mobility & integrity improvement can be achieved. During PROM exercises of upper extremities, care should be taken to mobilize the scapula, otherwise, impingement of glenoid fossa becomes the cause of the pain in shoulder. PROM exercises when performed, forceful or aggressive movts. should be avoided to avoid heterotopic ossification. It is more prone in the proximal joints & becomes the cause of pain & hypo mobility. Proper positioning will help in the reduction of secondary impairments such as contractures, bedsores, pneumonia, deep vein thrombosis, which not only hamper the progress but may lead to life threatening consequences. Good positioning will help in preventing skin breakdown, contractures, improve pulmonary hygiene & circulation. In bed, head should be positioned in neutral to prevent the neck contractures & to lessen the effects of tonic neck reflexes. The hips & knees should be slightly flexed. Turning frequently will help in preventing the skin breakdown & pneumonia. Caregivers should be told about the turning the patient every 2 hours when in bed. Specialized air mattresses with electrical / pneumatic pressure relieving system are useful for the alternate weight relieving from the load bearing body parts in the bed & avoiding the pressure sores. Splints help in positioning the head, the legs & the feet in these bedridden patients.

Stimulation of the sensory organs: Sensory stimulation is an intervention used in an attempt to increase the level of arousal & elicit movement in individuals in a coma or persistent vegetative state. It is proposed that, by providing stimulation in a controlled multisensory manner, with a combination of stimulation and rest, the reticular activating system may be stimulated causing a general increase in arousal. The following sensory systems are systematically stimulated: auditory, olfactory, gustatory, visual, tactile, kinesthetic & vestibular. During these types of interventions, the patient must be closely monitored for subtle responses such as eye movts., facial grimacing, and changes in posture, head turning, or vocalization. Overstimulation may lead to the accomodation making all efforts useless. Patient should be subjected for a single type of stimulus as multistimulii may lead to the confusion for the patient. During this Coma stimulation, physiotherapist must look for the warning signs like flushing, perspiring, increase in respiratory rate, agitation, eye closure, decreased level of arousal or increase in muscle tone. If any of these symptoms are seen, coma stimuation therapy should be immediately stopped. For the motor response, therapist should communicate with the patient by all the possible means.

The patients who are slow to recover & minimally conscious & who remain in this stage of recovery for a longer time duration are often placed in long term rehabilitation centre / nursing home.

Family members and caregivers of patients who have experienced a TBI play a critical role in the recovery process. Family members commonly experience a high level of stress related to worries

about the future, less free time, and increased conflict. Family education is an important component of the management. The goal of patient & family education is to teach the family about the stages of recovery and what can be expected in the future. By becoming informed, the family may not be so helpless. The family can also become involved in performing ROM exercises, positioning, & sensory stimulation. Although it is difficult to predict long term outcome at these early levels of recovery, families should be informed of possible outcomes. The therapist should be realistic but provide hope for the family. It is often beneficial to have a medical social worker consult with the family to provide support and guidance. When the patient begins to exhibit improved mobility skills, may lack the insight to recognize that he or she may not yet be safe to ambulate or transfer alone. The patient should be educated in how to best compensate for the residual impairments or disabilities. In severe cases, counseling will be required for the patient and the family members by a medical social worker or a neuropsychologist. Family members should learn how to assist the patient with functional mobility including bed mobility, transfers, ambulation and wheelchair mobility skills. They should be trained for the proper body mechanics when assisting the patients to avoid the injury to themselves or the patient. They should be taught the home exercises for the strengthening and PROM exercises.

SUB ACUTE STAGE

At this stage, the goals & anticipated outcomes would be:

- To preserve the integrity of the neuro musculoskeletal system, thereby preventing or minimizing adaptive muscle shortening and contractures
- To mange the effects of abnormal Tone & spasticity
- To provide an appropriate level of sensory stimulation
- To improve the motor control, postural control & functional level

Once the acute phase gets over & the patient with TBI is medically stable, continuation of the care can be in a variety of set ups. Patients with minimally conscious state or in coma may get the therapy in hospital or a long term care nursing home. Patients who start recovering from coma with moderate to severe cognitive, behavioral, and physical impairments often continue rehabilitation in either in-patient / out-patient rehabilitation centre or a multispecialty hospital. The cognitive abilities and behavioral presentation of individuals with TBI will have an impact upon their level of function. It is therefore vital that these elements be assessed and taken into account when agreeing goals for interventions. Access to a neuropsychologist, who can assess and advise on the most effective way to deal with these factors when planning treatment, is invaluable.

As the patient begins to emerge from coma, he or she often experiences a period of acute post traumatic agitation. The confusion, amnesia and disorientation often result in agitation, aggression, noncompliance and combative behavior. The patient may be markedly agitated and prone to emotional outbursts ranging from verbally acting out to physically attempting to hurt themselves or others or attempt sexually inappropriate behaviors. The patient is always confused & with the short & long term poor memory. Attention span is decreased, and so, get easily distracted.

Examination at this stage would be difficult as the patient is often uncooperative. The therapist must utilize observational skills by seeing the functional mobility, balance both in sitting and standing (if possible), ROM, strength, motor control, tone, sensation and reflexes. Patient's cognitive abilities also have to be examined like orientation, attention span, memory, insight, safety awareness and alertness. Neuropsychologist's help will be required to manage the patient's agitated behavior & help in setting up behavioral modification technique such as positive reinforcement using a reward system, redirection and compliance training which will help in managing inappropriate behaviors and improving in therapy. Severe cases may require medications too.

Due to the confused state of the patient, it is very important to maintain the consistency in the talking as well as in dealing with the inappropriate behaviors by all the members of the team as well as the family members. It is important to maintain the familiarity to the patient by keeping the same timing, place of the treatment & the same therapist. Patient should be frequently provided orientation information.

At this stage, patient may require one to one staff supervision and assistance throughout the day.

Family support required at this stage ranges from the supervision due to cognitive deficits to the physical assistance in all the ADLs due to motor deficits. For discharge from the hospital, this factor of strong family support & ability to help is as important as the health status of the patient.

The therapist will be primarily responsible to improve the motor & postural control which will be a prerequisite to improve the functional level of the patient. Maintaining the normal physiological length of the soft tissues is very important to prevent or minimizing adaptive shortening and contractures. This can be achieved by passive movements and correct positioning of the patient. Utmost care has to be taken to avoid the over-stimulation of the tissues and the joints. Taking care of the painful joints, especially the shoulder is very important at this stage as this pain if persists, become the cause of the inability to get the range even passively. If it prolongs for a longer time, to get the motor control over the movements of the upper extremities will be delayed & some times terminal ranges are permanently affected. Prophylactic splinting to the hypertonic muscular body parts helps to prevent the shortening or contractures. Worst cases of hypertonicity may require the medications.

Physical & motor problems of abnormal tone in the form of spasticity or rigidity is a common problem throughout the recovery phase or thereafter for many patients with TBI. More than 80% patients with TBI develop contractures due to the spastic hypertonicity. Increased tone may be responsible for the difficulty in personal hygiene, transfers or may be a source of pain or pressure sores. Increased tone in LEs may help the patient in weight bearing and bed transfers or make it easier for the caregiver to transfers.

Physiotherapists use two basic treatment strategies as compensatory (to improve functional skills by compensating for the lost ability) and restorative (to restore the normal use of the affected part) approach.

Many therapeutic skills are available with Physiotherapist to deal with the abnormal tone and in turn to avoid its adverse effects.

i) Basics of spasticity management are therapeutic stretching-sustained & gentle, strengthening exercises with adjunctive modalities and functional retraining. PROM and selective strengthening of the antagonist muscles can help to decrease spasticity. Positioning is an important adjunct in the management of tone abnormalities. Maintaining the head and neck in a neutral position is important for minimizing the effects that primitive postures may have in increasing tone. Keeping the whole body in proper alignment is also important.

ii) Cryotherapy or air splints can also be used to reduce the hypertonia, though their effect is temporary.

iii) Serial casting helps in decreasing hypertonia. Serial casting is often used for plantar flexors or biceps contractures resulting due to either increased tone or prolonged shortening of the muscle. Progressive maximal stretched position is achieved for the cast to maintain for a week before trying to get the new more stretched position is tried for the casting. PROM exercises are continued to have the new increased ROM. There should be constant check on the skin coming in contact with the cast as it can breakdown if the patient has sensory, communication or behavioral impairments.

iv) Various medications either systemic (Baclofen) or local (phenol or Botulinum toxin) can also be used to reduce the hypertonia but their use should be weighed in comparison with the side effects they can produce in a particular patient

v) Increased tone in lower limbs that requires more than manual techniques to inhibit and maintain range of movement may benefit from the application of below knee weight bearing plasters. This is a skilled task and care must be taken to obtain correct alignment of foot and ankle.

The tilt table can be used for periods of standing in conjunction with standing with necessary assistance & standing transfers. Lack of head control can impede progress to activities in sitting & standing.

Many patients with TBI get affected with ataxia, who will need the coordination & balance therapy. Some of these may require a walking aids either temporary or for a longer time. Sensory disturbance will involve perceptual problems including vision & hearing, apraxia & agnosia. Emotional & intellectual disturbance will involve memory & behavioral disorders. For these patients co treatments will be beneficial if done along with

Speech therapy, Occupational therapy & Neuro psycho therapy.

Upright sitting is very important to work towards the treatment goals for the early levels of recovery. Upright position is vital for the proper functioning of many organs like, stimulation of bowel movements and bladder emptying, improved ventilation due to moving down of abdominal contents leading to redistribution of air flow to basal lobes and changing perfusion / ventilation ratio. As soon as medically stable, the patient should be transferred to a sitting position and out of bed to a wheelchair / chair. All precautions should be observed. The head should be properly supported, as the patient may have very inadequate neck and head control to maintain an upright posture without support. Often, it is beneficial to perform co treatments with an Occupational therapist when first sitting & transferring the patient. Use of a tilt table is very useful as it allows early weight bearing through lower extremities. The upright position in sitting by the side of the bed, in a wheel chair or standing on a tilt table improves overall level of alertness. Sitting training promotes movement and learning by allowing the patient's body in functional position to do the tasks. Assisted movements provide tactile, proprioceptive and kinesthetic stimulation while training to perform a ADL task.

Constant efforts are required by the physiotherapist to improve the motor control & motor learning. Neuro physiotherapeutic techniques can be used to initiate, progress, improve the sensory motor control and finally helping the patient to get back to the as normal life as possible depending upon the severity of the lesion, personal as well as external contextual factors. Due to the newer understandings and the theories of the motor learning and the practices, many new skills are developing but still most of the older skills also hold true today and specifically in the initial and subacute phase, they are most useful. The therapist should work as per the patient's physical level of function and attempt to improve endurance. Progressing to more challenging skills which would require new learning, should be as per the patient's capacity for new learning.

Depending upon the skills and experience of the therapist, clinical conditions and the co-morbidity of the patient any or many of the followings are used.

i) **Developmental sequence:** Developmental postures are utilized to facilitate and help restore movement and functional mobility. Once, the patient gets the fair control over the different parts of the body as per the involvement due to their cerebral or the cerebellar trauma, postural training should be started on the basis of developmental sequence & the biomechanical stability basis. This may start with the supine-side turning as a log first & then by girdle initiation, cephalocaudally or caudocephalic-prone lying- coming on the forearm & then on hands- all four with elbow / forearm loading to hands loading- quadruped position with ability to relieve the load of one or two extremities (crossed)-independent sitting -kneel sitting-half kneel sitting-kneel standing- sitting to standing- kneel standing to standing-squatting -walking & then the training for the dynamic postures. All the static postures first to be trained in assisted & guarded situations. Once active control is gained, then they are made more & more difficult so that in adverse situations they will be mastered. This can be against the resistance or against the smaller base of support or against the unstable base. The gymnastic ball has specific qualities which can be used to gain mobility, stability & postural control. Other injuries, if any, than TBI also should be taken care of during training all these postures. As and when, the patient masters the posture, ability should be used for the related possible activities performance first in the therapeutic area & then in life situations.

ii) **Proprioceptive Neuromuscular Facilitation (PNF):** This is a good technique for the initiation of mass movements of the proximal girdles to the distal components as they are closer to the activities of the daily life. These can be first initiated passively by the therapist with the commands till there is response and then assisted to learn actively by the patient. Full range movements along with the strength and endurance are achieved. All these activities should be trained for the near normal equilibrium responses. Sufficient practice will give this maximum perfection to which the patient can reach a level & with the maximal endurance. Once this is met in the treatment area, this has to work equally good in the actual life situational areas, where patient will have to be multiresponsive due to the environmental demands depending upon the crowd, architectural barriers & the vehicular traffic.

iii) **Neuro Developmental Therapy(NDT):** This is based on Ms. Bobath's core ideas of rigorous observation of posture and movement, careful alignment of body segments and joint position before asking patients to move, constant analysis of how the patient moved throughout the entire treatment session, the contribution of the sensory systems to movement, family involvement in treatment, and home programming.

Task oriented approach is currently advocated for this locomotor training, utilizing

iv) **Body Weight Support (BWS) and a treadmill:** Suspending the patient in a parachute like overhead harness which allows partial relief / support of the body weight. Therapist needs to support the patient by pelvis / trunk for the weight shifting & the forward advancement of the LEs. This can be done in parallel bars, with walker or with the treadmill. Training as well as corrections can be done in this system in each phase of the gait cycle. Difficulty can be progressively increased by decreasing the harness & physiotherapist's support and increasing the speed of walking or treadmill as the patient's ability improves.

v) **Constraint Induced Movement Therapy(CI/ CIMT):** It involves promoting the use of the most affected UE for up to 90% of waking hours and reducing the use of the lesser affected UE. Intensive, task oriented training is provided for the affected UE for up to 6 hrs per day over 2-3 weeks period.

vi) **Modified Constraint Induced Movement Therapy(MCIMT):** A mesh is worn to the unaffected hand for gentle restraint of the actvities for several hours for 10 weeks period.Training will be same as CIMT.

vii) **Hand Arm Bimanual Intensive Training (HABIT):** This is also a form of intensive task training, but the more emhasis is on the bilateral hand manipulations in ADLs. Unaffected hand may play a major role at the initial phase, but, emhasis should be given on the more and more equal participation of the affected hand during the tasks.

Patient should be started training the simple daily functional activities like bed activities, feeding & the ambulation before expecting him / her to learn new skills. Use of some audiovisual aids like charts / pictures / photos will help the person to learn new skills & the progress each day. Patient will be required to give a recall each day of the previous days teaching as carryover will be difficult in many patients.

It is important for the therapist to perceive the patient's emotions & behavior, so that he / she should be able to make feel the patient safe & secured. Even the family members' & caregiver's attitude towards the patient will have an effect on the behavior of the patient. Patient will not be in a position to understand the other people's views & most likely egocentricity may be present. Due to the limited attention span of the patient and the easy distractibility, concentration on the activities training is going to be limited for early days of the treatment.

PHYSICAL REHABILITATION

At this stage the main goals of physiotherapy interventions are to:

- Encourage the return of active movt. that carries over into function
- Prevent secondary deformities
- Prevent unnecessary and potentially damaging compensatory movt. strategies
- Maximize respiratory function
- Encourage social and vocational reintegration
- Provide advice to the family, caregivers and other members of the team on aspects of the patient's management
- To educate family members on patient's condition, management goals & outcomes

At this stage, when patient shows the signs of improvement in the consciousness & the awareness or may be only in the sensory motor components with respiratory independence, patient will be most of the times either in half way rehabilitation centre or at home & comes for the therapy on out patient basis.

The examination should be done at this level to access the status on following points:

- attention & cognition
- cranial nerve integrity
- balance
- gait
- joint mobility

- motor control
- muscle strength
- ROM
- Reflexes
- ADL skills & sensory integrity
- Functional abilities of the patients (better in closed environments than an area of multiple distractions)
- Gait is generally examined by Observational Gait Analysis to know the deviations & asymmetries so the impairments can be found to treat the causative factors. Gait speed & the endurance can be measured by 10 meter walk or 6 minute walk test
- Their motor control & functional status including the ability to perform ADL, transfers, bed mobility, stairs, & locomotion can be measured by using Functional Independence Measure (FIM) or Fugl-Meyer Assessment of Motor Recovery (FAM) or Chedoke -MacMaster Stroke Assessment.

For the task performance training, it is to be seen about the balance during the performance, initiation & completion, consistency, energy required, shift of the body weight & maintenance of body alignment.

Those having cognitive & behavioral deficits will have difficulty in reacquisition of the motor skills due to their mental as well as physical fatigue during treatment sessions. That can be known by the signs like increased irritability, decreased attention and concentration, deterioration in the performance of physical skills and delayed initiation. Treatment sessions with sufficient rest periods will avoid the mental & physical fatigue and will maximize the motor relearning.

TBI patients will have many different physical, cognitive and emotional impairments making the therapist to have an individual approach as per the patients capabilities and the difficulties due to the uneven recovery of these different aspects of the brain functioning. For every patient, the emphasis is given for the initiating, ability to get it actively and perfecting the motor tasks by making them progressively increasingly difficult and complex to blend them in the daily activities successfully.

TBI patients have a wide variety of impairments and functional limitations, they may be helped to compensate for the lost function by giving them some form of adaptive device. It depends upon the predicted probable residual disability as per the opinion of the rehabilitation team, to decide how much help should be given. Most severe patient who may not be able to make to walk again will require a wheel chair. Depending upon his or her regained cognitive skills, wheel chair can be advanced with the computer based controls powered one to the simple manual one which may be operated by the patient or the caregiver.

Therapist should have more options of the activities for the training, so after few days, different combinations & the options to the patient can be given for breaking the monotonicity as well as to give choice to the patient.

Prior to discharge from a comprehensive interdisciplinary day treatment settings, cognitive, behavioral, emotional and psychological issues have to be tackled before community re-entry and return to school / work. Skills in judgment, problem solving, planning, self awareness, health and wellness and social interaction should be taught by the respective professionals with lesser and lesser supervision needs. Once the patient gets the insight into his / her own strength and weaknesses, they should be involved in decision making for their working towards reintegration back to home and community. Group treatment sessions and Support groups are crucial for the patient to learn how to function in society with their present abilities and limitations. Adaptations are required most of the times by the family, work place colleagues or schoolmates to accommodate the patient.

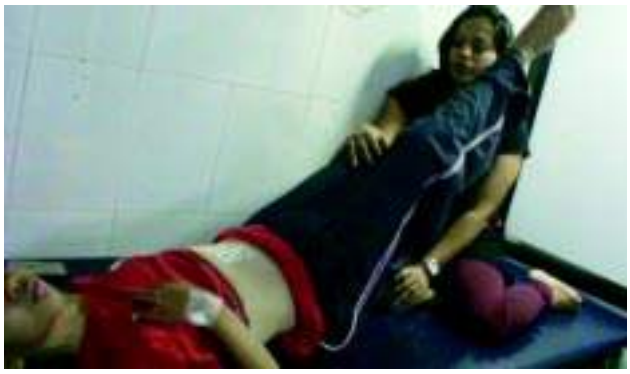
Occupational Therapy and Traumatic Brain Injury

Brain injury is a life-altering event which affects every area of a person's life including his or her relationship with family members and others close to him or her. The effects of brain injury often change roles and responsibilities within the family.

TBI can result in physical, cognitive, behavioral, or emotional difficulties. People with TBI may experience short- term memory loss, have difficulty concentrating or paying attention, have impaired judgment, experience headaches or migraines, have slurred speech, experience seizures, become fatigued, depressed, or easily agitated, or experience increased anxiety and impulsive behaviors.

Half Rolling

Bridging



One leg Bridging

Side sitting in Quadrapud

Crawling

Hip extension in Quadrapud

Unilateral arm raise in Quadrapud

Kneel walking(1)

Ball Catching in Kneeling Position

Forwar bending at the edge of bed in sitting

Long leg shifting in forward direction

Long leg shifting in backward direction

Getting up from sidelying

Getting up from supine

Getting up from low stool

One leg Standing

Reachouts with weight bearing on affected side

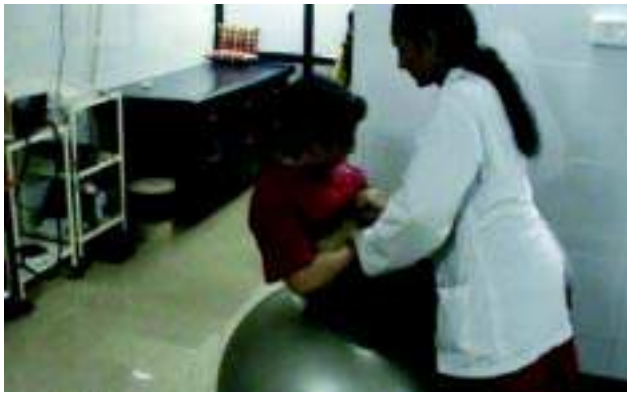
Coordination exercises for Upper limb

Balance board exercises

Cognitive Therapy

Hip adduction on Swiss Ball

Hip flexion on swiss ball



Upper abdomen on Swiss Ball

Back extension on Swiss Ball

Post trauma manifestations may also include occasional loss of consciousness, dizziness, headache or vertigo provoked by sudden changes in position; confusion and disorientation about time and place; and emotional reactions such as combativeness. Behavioral disturbances may be blatant, particularly during intital recovery period. Personality disorders, ammensia and delirium may also occur. These manifestations may be accompanied by intellectuals deficits, blindness, diplopia, hemianopsia, olfactory dysfunction and auditory deficits. Physical symptoms include quadriparesis, unilateral or bilateral hemiparesis, initial decerebrate rigidty and speech deficits.

These deficits prevent the head injured person from effectively interacting with the environment and may hinder rehabilitation. The patient may seem to have lost contact with the surroundings and to be able to focus only on the narrow yet overwhelming feelings, concerns and behaviour imposed by the sudden disability. The clinical picture represents a pattern of fluctuating symptoms in these areas, subject to frequent change evident throughout the recovery as the patient tries to adjust to the deficits imposed by the injury, to learn to decrease and eliminate handicapping effects and to renew self confidence through awareness and acceptance of the immediate and future levels of function.

Occupational therapists can help people who have sustained a TBI. Injuries can range from minor to extremely severe and call for various levels of intervention and treatment. The type and duration of the intervention will also depend on the severity of the injury.

As part of the TBI rehabilitation process, occupational therapy is designed to assess functions and potential complications related to the movement of upper extremities, daily living skills, cognition, vision and perception. Together with the patient and family, the occupational therapist will help determine the best ways to perform daily living skills including showering, dressing and personal hygiene. The therapist will identify equipment that can help the patient when eating, dressing and bathing.

The occupational therapist will also address specific skills to prepare the patient for their return home. These skills may include activities for daily living, such as Cooking, Grocery Shopping, Banking, Budgeting, Readiness for returning to work by assessing prevocational and vocational skills, etc.

Interventions and Practices

Development of Occupational Profile:

Prior to evaluation the occupational therapist needs to gather as much information as possible (from varied sources such as diagnostic reports, (CT scan and MRI reports), medical notes, and interviews of client and or care takers and other team members) regarding patients condition such as presence of any co morbidities (associated fractures and or pheripheral nerve injuries) and pre morbid status. The CT scan and MRI reports are invaluable in indicating the area and extent of damage caused to the brain and the possible potential impairments that the client may have. The therapist then evaluates the client to develop a clear understanding of the areas of impairment and its effect on the abilities and capabilities of the patient and its impact on the occupational performance of the patient. This information is then used to assist in directing any assessments and identifying underlying causes for problems in functional abilities of the client.

Analysis of Occupational Performance:

Information from occupational profile informs the Occupational Therapist about the extent of impairment, activity and participation limitations while considering the environmental and social contexts. In the early phases of recovery the client may be unable to participate in purposeful and goal directed activity due to the altered state of consciousness. Hence OTs analyse the occupational performance on return of mental alertness and purposeful behavior. The occupational therapist should observe and anlyse the clients occupational performance in the natural or least - restrictive environment. The therapist must endeavour to note the effectiveness of the following performance skills: (i.e. the observable elements of action of an occupation.)

1. Motor and praxis skills
2. Sensory -perceptual skills
3. Cognitive skills
4. Emotional regulation
5. Communication
6. Social skills

It is also important that the occupational therapist gathers information about

- The clients performance patterns e.g. habits, routines, rituals and roles

- The client factors like underlying abilities, values, beliefs, and spirituality, body functions and body structures which influence individuals occupational performance.
- Environment and contextual factors such as influence of cultural, personal, physical and social context on occupations and activities.
- Activity Demands The demands of an activity are aspects of the activity that include the tools needed to carry out the activity, the space and social demands required by the activity, and the required actions and performance skills needed to take part in the given activity.

The occupational therapist theoretical knowledge and clinical expertise help in selecting specific assessment and evaluation methods to identify and measure the factors affecting clients performance at a particular time.

Occupational Therapist may elect to use client - centred evaluation approach that may focus on possible impairments affecting performance of function. (Bottom - up approach) or an evaluation approach that analyses the role of individual with TBI and areas of occupation that encompass a typical day (top - down approach)

Occupational therapist may select dynamic assessments that emphasise the process involved in learning and change to gather information to guide treatment planning and intervention or may select performing evaluations that include body structures and functions, activity and participation enabling the occupational therapist to compile a comprehensive view on the persons functioning.

Prefered assessments are assessments with proven

- Validity with traumatic brain injury population
- Efficacy in detecting and quantifying the typical pattern of impairment seen in clients with TBI.

Based on the assessment data, the occupational therapist then endeavors to

- Identify factors that support or hinder clients performance such as memory impairments affecting personal hygiene, home management skills, work tasks and social participation.
- Establish goals that are meaningful to the client and family and address the client's desired outcomes.
- Identify potential approaches guided by best practices and evidence and as per discussed with client and family.

Interventions:

As part of the OT process, the occupational therapist develops an intervention plan that considers the clients goals, values beliefs, health and well being, the client's performance skills, performance patterns, collective influence of context, environment activity demands and client factors on client's performance.

The intervention plan outlines and guides the therapists' action and is based on best available evidence to meet the targeted outcomes. The therapist also determines the intervention approach that is best suited to address the identified goals. The Intervention approaches used by OT include

Prevent: an intervention approach designed to address clients with or without disability who are at risk for occupational performance problems; for example, intervention to prevent development of secondary impairments such as joint contractures during the coma phase of recovery.

Establish and restore, an intervention approach designed to change client variables to establish a skill or ability that has not yet developed or to restore a skill or ability that has been impaired; for example, restoring hand coordination to engage in functional activities such as cooking.

Modify activity demands and the contexts in which activities are performed to support safe, independent performance of valued activities within the constraints of motor, cognitive, or perceptual limitations.

Create or promote a healthy and satisfying lifestyle that includes adherence to medication routine, appropriate diet, appropriate levels of physical activity, and satisfying levels of engagement in social relationships and activities by providing enriched contextual and activity experiences that will enhance performance for all persons in the natural contexts of life.

Maintain performance and health that the individual with TBI has previously regained or that neuropathology has spared.

Types of intervention:

The OT consider the type of intervention when deciding the most effective treatment plan for a client. The types of interventions include:

- Therapeutic use of self (therapist's use of his / her personality, perception and judgement).
- Therapeutic use of occupations and activities which include
  - preparatory method : (e.g. Serial casting, sensory stimulation.)
  - purposeful activity : (e.g. role playing of social situations, practicing grocery shopping in stimulated environment)
  - occupation based activity: (e.g. Interviewing for a job or preparing meal for one's family.)
- Consultation and
- Education

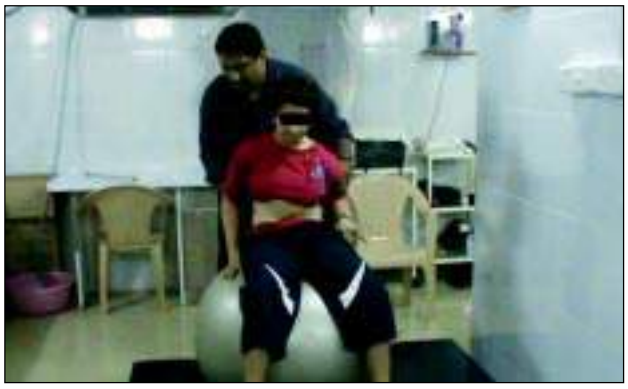
I. Intervention during early period of recovery:

During the early period of recovery from a TBI, the occupational therapist working with the client in coma typically focuses the intervention on establishing or restoring the client factors or impairments that resulted from the injury such as impairment in voluntary control abnormal tone. Intervention also focuses on preventing the development of secondary impairments that occur in the period of unconsciousness like prevention of pressure sores and heterotrophic ossification. Global mental functions, such as level of consciousness and alertness, are often addressed through a program of coma arousal or sensory stimulation.

The focus of intervention may shift among establishing, restoring, or maintaining occupational performance; modifying the environment and / or contexts and activity demands or patterns; promoting health; or preventing further disability and occupational performance problems.

Occupational Therapy Intervention Approaches and Examples of Their Use with Clients at Various Levels of Recovery from Traumatic Brain Injury

Intervention Approaches Commonly Used During the Coma Phase of Recovery (Rancho Los Amigos Levels I-III) Interventions typically focus on state of consciousness and WHO ICF areas of impairments.	
Intervention Approach	Occupational Therapy Treatment Activities
Prevent	Prevent loss of muscle length and joint mobility by • performing range of motion (ROM), • serial casting, • tone-inhibiting techniques, and • positioning of patient in the bed and wheelchair. Prevent skin breakdown and postural deformities by providing • Providing the client with proper body alignment in tilt-in-space wheelchair with head • rest, lap tray, gel seat cushion, and trunk inserts and • providing the nursing staff with a splint-wearing schedule.
Establish / Restore	• Restore the client's connection to the external environment by positively reinforcing appropriate behavioral responses to sensory stimulation.
	• Restore the client's ability to follow one-step demands for a motor response in relation to sensory stimulation within 15 seconds of request or stimulus.
Modify	• Modify environment to vary levels of stimulation
	• prevent accommodation and attenuation to environmental stimuli (e.g., vary lighting, noise level, visual stimulation, temperature).
Maintain	• Maintain muscle length and joint mobility by instructing caregivers in routine stretching exercises.



Ball bouncing

Tilt in space wheelchair

Gel seat cushion

Attached lap tray to wheelchair

Basic self care grooming activity- Wiping face

A) Object manipulation using marbles

B) Object manipulation using Shape sorting activity

A) Restore cognitive skills using Stacking activity

B) Restore cognitive skills by naming of objects using flashcards

Neuromusculo skeletal Recovery Programs

Occupational therapy intervention for neuromusculo-skeletal and movement-related functions after brain injury is focused on either impairments that are a primary consequence of that injury (e.g., impairments in voluntary movement, abnormal tone, balance) or secondary impairments resulting from the immobility or excessive muscle tone, such as contractures of the muscles or joints and diffuse weakness and deconditioning. At the coma phase of recovery, intervention for neuromuscular motor impairments is generally passive in nature, using more preparatory methods such as passive range of motion (PROM), splinting and serial casting, and positioning in the bed and wheelchair to either establish and restore motor control or prevent the development of secondary joint and muscle contractures. Joint contractures can result in significant limitations in self-care, particularly dressing and hygiene.

PROM:

Prolonged periods of disuse and immobility can lead to change in muscle and joint postural alignment and other impairments (such as muscle tightness, atrophy, fibrosis, contracture, postural deformity). Early intervention is critical in maintaining joint motions, tissue extensibility, physical ability and function. Additional benefits include improved circulation and tissue nutrition to the limbs and pain inhibition along with decreased hypertonicity and increased sensory input.

In the coma phase of recovery, when the patient is unresponsive, the therapist or the caregiver performs ROM exercises passively through the patients full available range. The limbs are well supported with stable positioning of the patient to prevent joint trauma. Movements should be slow and rhythmic taking care to avoid excess force and pain. This is especially important in clients with TBI as they are at risk for heterotropic ossifications.

When moving the Upper Extremity during PROM, care should be taken to mobilize the scapula and to maintain proper joint mechanics at the glenohumeral joint. Maintaining ROM at the hip and ankle are especially important as a decrease range of motion at these joints can greatly impact functional activities like transfers, sitting, wheelchair positioning ambulation and stair climbing.

Splinting and Serial Casting: Splints are indicated when

- Spasticity interferes with movement
- Joint range of motion limitations are present
- Soft tissue contractures are possible.

Splints are thought to provide elongation and inhibition by positioning the joint in a static position with the muscle and soft tissues on stretch.. Splinting of elbows, wrists and hands is often implemented to maintain functional position at rest and to reduce tone. For eg. Resting hand splint or functional position splint. Once splints have been fitted, then:

- A wearing schedule is established for the care givers to follow.
- Caregivers are trained in proper application and removal of each splint.
- The client must be monitored frequently for skin breakdown or tonal changes that may change the initial fit of the splint.

Serial Casting:

A serial casting program is a more aggressive intervention indicated in the presence of moderate to severe spasticity that cannot be managed by splints. The goal of serial casting is to increase range of motion and decrease tone by using progressive succession of separately fabricated casts each worn continuously for a period of weeks. Casts are often left on for five to seven days which puts the muscle and tendons on a prolonged stretch and reduces tone. Successive casts are designed to increase range of motion further until a functional joint range is achieved and maintained. The common difficulty that prevents the success of serial casting is skin breakdown.

Positioning in Bed

Proper bed positioning is critical in early stages of TBI to prevent pressure sores and to facilitate normal muscle tone. If a client exhibits abnormal tone or posturing a side line or semi-prone position is preferred as it assits in normalizing tone and providing sensory input. A supine position might elicit tonic labyrinthine reflex and extensor tone. Supine position with head in lateral position may elicit asymmetrical tonic neck reflexes. Clients with TBI generally have bilateral involvement requiring a program for side-lying on both sides.

Pillows, foam wedges and splints may be incorporated in a bed positioning program to facilitate normal position and prevent abnormal postures such as extreme elbow flexion, head and neck extension and foot drop deformity.

Wheelchair Positioning:

Positioning the client in coma in a wheelchair with adequate supports for the head and trunk also can decrease muscle tone, foster increased upright motor control in a functional posture, and improve awareness of stimuli within the environment.

A proper wheelchair positioning helps prevent skin breakdown and joint contractures. Facilitate normal muscle tone, inhibit primitive reflexes, increase sitting tolerance, enhance respiration and swallowing and promote functioning.

While seated on a wheelchair, the client's buttocks should bear weight evenly with both ischail tuberositities firmly resting on the wheelchair seat. The knees should be positioned at 90 degreees with heels slightly behind the knees. It is desirable to maintain both feet securely on the footplate to provide propio-ceptive inputs and facilitate weight bearing in both heels to normalize tone. The upper extremity should be positioned with the scapulae in neutral position (neither elevated nor depressed, shoulders slightly externally roated and abducted, the elbows in a neutral position of slight flexion with forearm pronation and the wrist and digits in a functional position. Slightly reclining the wheelchair between 10 to 15 degrees prevents clients head from falling forward and facilitates visual interaction with the environment. A good wheelchair positioning can be achieved by using several positioning devices as and when necessary such as:

- A firm solid seat (padded with foam and covered with vinyl) facilitates neutral to anterior pelvic tilt and prevents internal rotation and adduction of the hips.
- A wedged seat insert with downward slope pointed towards the back of the chair facilitates hip flexion and inhibits extensor tone in hips and lower extremities.
- Lateral trunk supports can be used to reduce scoliosis and lateral trunk flexion caused by imbalanced tone of the intrinsic muscles of the back.
- An abduction wedge between the lower extremities can help in reducing hip adduction and internal rotation.

Management of Heterotopic Ossifications (HO)

Monitoring clients for the development of HO (e.g., observation for an inflammatory reaction, palpable mass, or limited ROM in joints of the limbs) in clients with abnormal tone is important. During the acute inflammatory stage, the occupational therapist should position the patient's involved limb in a functional position and initiate gentle PROM, monitoring the patient for signs of pain (e.g., facial grimace, change in vital signs). Once acute inflammatory signs have subsided, continued mobilization is indicated to maintain range. Resting the joint appears more likely to lead to decreased joint range, whereas continued mobilization may lead to formation of a pseudarthrosis.

Sensory Stimulation Programs

The goal of sensory stimulation program is to increase the client's level of awareness by trying to increase arousal with controlled sensory input. Introduction of sensory stimulation in the form of isolated visual, auditory, tactile, olfactory and gustatory stimulants to the individual with brain injury may heighten arousal. For e.g.

1. Flash light may be used to elicit eye opening and visual tracking.
2. Playing familiar music may cause changes in respiratory rate or blood pressure.
3. Olfactory stimulants like variety of scents may elicit eye opening or head turning.
4. Gustatory stimulation like salty, sweet, bitter or sour taste to clients lip and tongue, etc.
5. Kinesthetic input in the form of guided movements while performing simple movements like rolling side to side or wiping the mouth.

The theoretical aim of functional sensory stimulation is to reactivate highly processed neural pathways that have been established before the injury.

II. Intervention During the Acute Rehabilitation Recovery Phase (RLA Levels IV to VI)

Intervention Approaches Commonly Used During the Acute Rehab Phase of Recovery (Rancho Los Amigos Levels IV-VI) Interventions typically focus on motor and cognitive skills and WHO ICF areas of impairments and activity limitations.	
Intervention Approach	• Occupational Therapy Treatment Activities
Prevent	• Prevent aspiration during feeding by modifying the food texture and head positioning if the client displays signs of dysphagia.
Establish / Restore	• Establish the client's ability to release energy constructively during agitated periods by providing structured and familiar activities with minimal challenges to areas of impairments. • Restore ability to perform self-care by engaging the client in a daily self-care program of showering, dressing, and grooming, providing verbal and physical cues as needed. • Restore normal patterns of movement by engaging the client in various functional motor tasks (e.g., grooming, self-feeding, object manipulation) with gradual increases in the unpredictability and complexity of the contextual and activity demands, providing tactile input to guide and normalize movement patterns. • Establish skills to safely and efficiently transfer from wheelchair to various surfaces (e.g., toilet, bed, chair, car). • Establish and restore cognitive skills by teaching cognitive strategies to improve performance; engage in a variety of activities related to roles, responsibilities, and interests (e.g., financial management, cooking, parenting, leisure pursuits). • Establish strategies and new routines to accurately use external memory aids to recall scheduled appointments and events and to take medications. • Establish habits to ensure accuracy of work (e.g., self-monitoring of work for errors, timely completion, match with instructions).
Modify	• Modify the client's hospital room to provide environmental cues to minimize confusion and to provide orientation to person, time, and place. • Modify tasks and environments to enable independence (e.g., provide adaptive equipment to increase independence in ADLs and IADLs, such as checklists for activity sequences and external memory aids).
Maintain	• Maintain the client's postural alignment while sitting by providing structural wheelchair supports. • Maintain the client's maximum ROM obtained with serial casting by providing resting cast / splint for night wear.

The acute rehabilitation phase of recovery begins as the client is medically stable and emerging from the coma. Occupational therapists, as part of the rehabilitation team working with clients who have TBI, must address the client's physical, cognitive, communicative, emotional, and spiritual needs. This phase of recovery from TBI encompasses a spectrum of behaviors by the client that fluctuate with changes in situational factors such as environmental stimulation, task demands, and time of day. During this phase the client is alert but often displays confused, agitated and inappropriate response which might hinder his ability to participate in the rehabilitation program.

Behaviour modification techniques such as positive reinforcements using a point or reward system, redirection and compliance training are useful in managing inappropriate behavior and improving participation in therapy.

As the agitation lessens, the cognitive and motor challenges presented to the client can be gradually increased to address underlying impairments.

Addressing Motor Recovery

As the client with TBI emerges from coma and performs more voluntary movement, the occupational therapist begins to address impairments seen within the sensory and neuromusculoskeletal and movement-related systems. Clients with persistent spasticity resulting in joint contractures interfering with performance of functional activities, who have not been successfully managed with more conservative rehabilitation techniques, may be candidates for botulinum toxin A injections, motor point or neural blocks, surgical release of the contracted tissue, or intrathecal baclofen pump placement. Occupational therapy intervention following these procedures can be helpful to increase functional integration of the limb into daily activities.

Occupational therapists apply the principles of practice and feedback, task-specificity, and training intensity when providing intervention focused on motor skill recovery. Two commonly used approaches to address movement related impairments after TBI are motor learning and constraint-induced movement therapy (CIMT).

Motor learning is a process of learning to produce skilled movement that involves practice and experience. Occupational therapists using a motor learning approach set up the therapeutic learning environment, typically the occupational therapy clinic, to promote skill acquisition by varying the tasks and environment to meet the patient's current learning abilities.

CIMT, a motor skill intervention approach based on the learning principles of shaping and preventing learned nonuse, was initially designed to increase the use of the impaired arm in chronic stroke patients. CIMT involves three main components: (1) intensive training of the affected arm, (2) practice to promote transfer of therapeutic gains from the clinical environment to real-world situations, and (3) constraint of the less-affected arm during the entire period of intervention.

Some treatment approaches utilize the developmental sequence and muscle facilitation techniques. Developmental postures like prone on elbows, quadruped, sitting, kneeling half kneeling and standing can be used to facilitate and help restore movement and functional mobility.

Other Considerations:

A brain injury can cause fatigue and conditions such as seizures, spasticity and swallowing difficulties visual problems and bladder and bowel changes.

Fatigue

Fatigue may result from the injury (and other injuries in cases of trauma) or from additional physical and mental effort required to do tasks that once were performed with little or no effort. Physical functioning, attention and concentration, memory and communication can be adversely affected by fatigue. In time, the person's stamina and energy level likely will improve, and the ability to engage in activities may be increased. The following strategies may be useful in helping the person with brain injury learn to manage fatigue:

- Encourage use of a calendar or planner to help manage mental fatigue.
- Set a schedule that includes regular rest breaks or naps. (For example, one nap in the morning and one in the afternoon after some physical or mental activity.) Rest breaks or naps should not exceed 30 minutes and evening naps should be avoided.
- Gradually decrease the length and number of breaks as the person's ability to tolerate activities with less fatigue improves.
- Resume activities gradually, over weeks or even months.
- Start with familiar tasks that the person can complete without fatigue.
- Gradually increase the complexity of the task, encouraging breaks as needed, and slowly increase the length of time.
- Become familiar with indicators of fatigue for the person such as increased inattention or distractibility, repetition of tasks or comments, irritability or increased errors.
- Encourage breaks, every five minutes, during tasks, before or as soon as signs of fatigue appear.

- If the health care team recommends, use assistive aids (for example, a cane for walking) to conserve energy or a wheelchair for long distances.
- Plan ahead for fatiguing activities, such as visitors, trips, going out.
- Schedule a nap before visitors come or before going out.
- Consider limiting the person's time with visitors or a rest break during visits.

Seizures

Seizures can be caused by a sudden, excessive, disorderly electrical discharge of brain cell activity. This risk of ongoing seizures is related to the severity and characteristics of the brain injury, such as the type and location of the brain injury. Risk seems to be greatest in the months after injury, and generally declines with time gradually.

Several types of seizures may occur after brain injury. The most frequent types are generalized (grand mal, tonic/clonic) and partial (partial complex and simple partial) seizures.

Most seizures are self-limited and last only a few minutes. The person may cry out, stiffen and fall, have jerking movements, turn flushed or blue and have some difficulty in breathing. The OT can educate the immediate caretakers on the steps that need to be taken in case of a seizure, such as:

- Make sure the person is in a safe area and lay the person's head on something soft if a fall has occurred.
- Loosen tight clothing such as a necktie or belt and remove eyeglasses.
- Clear away hazardous objects that may be nearby.
- Position the person lying on his or her side to keep the chin away from the chest. This will allow saliva to drain from the mouth.
- Do not force your fingers or any object into the person's mouth.
- Do not restrain the person since a seizure cannot be stopped.
- After the seizure, the person usually will be temporarily confused and drowsy and hence do not offer food, drink or medication until the person is fully awake. Someone should stay with the person until full recovered has occurred.

Driving privileges should be restricted until a predetermined seizure-free interval has been achieved (often six months to one year). During this time, extreme caution should be taken if the person will be working around heavy or dangerous equipment.

Swallowing

Problems that affect swallowing after brain injury can vary widely and may include one or more of the following:

- Poor head or upper body control
- Decreased lip and tongue strength, range of motion and coordination
- Impaired memory or concentration
- Any or all of the above may cause aspiration (inhaling food or liquid into the lungs)

Exercises, treatment techniques and positioning may help improve a person's ability to chew and swallow. An occupational therapist along with a speech therapist will teach the person with brain injury and caregivers how to perform these exercises and techniques. Most people regain the ability to swallow after brain injury, though it may take longer for some than others.

Visual Problems

Occupational therapy practitioners are recognized experts in addressing the vision processing deficits that occur from traumatic brain injury (TBI). Practitioners provide interventions to improve visual attention, search and speed, and efficiency in visual processing. They work closely with optometrists and ophthalmologists to ensure that clients with TBI are able to compensate for deficits in acuity, visual field, and eye movements to complete important daily activities safely and effectively.

Clients with TBI either have cortical visual impairment (CVI; affecting areas of the brain that process visual information) or ocular visual impairment (affecting the eyes). Clients with either CVI or ocular impairment benefit significantly from coordinated and comprehensive services to enable them to learn to use their remaining vision more efficiently and learn non-visual methods to complete activities.

Bowel and bladder changes

Brain injury may affect bowel and/or bladder

function. The injured person may need help re-establishing and maintaining a pattern of regular bowel and / or bladder emptying.

Bowel management

The goals of bowel management include establishing a regular emptying pattern, maintaining dry, healthy skin, and avoiding incontinence, diarrhea, and constipation. Each person is assessed by a physician and recommendations are made as needed.

Bowel problems can occur if the person with brain injury:

- cannot control bowel emptying voluntarily
- cannot recognize bowel fullness and the need to have a bowel movement
- cannot ask for help to the bathroom
- cannot walk to the bathroom
- cannot eat enough food with fiber and drink enough fluids
- cannot plan ahead and allow enough time to get to the bathroom

To maintain optimal bowel function, a person with brain injury should eat at regular times, focus on eating foods with fiber, drink the amount of fluids recommended by the dietitian or physician, and be as active as possible. Meeting with a dietitian to discuss a diet plan may be helpful. The person also may be asked to follow a bowel care schedule, which includes attempting to schedule a bowel movement at the same time daily and establishing regular times for meals.

At certain stages of recovery, other methods for bowel emptying (fiber supplements, stool softeners, suppositories, and / or laxatives) may need to be used. However these methods are not used regularly because they decrease the colon's natural abilities, and these methods may be habit forming. Most individuals with brain injury regain the ability to regularly and effectively empty their bowels.

Bladder management

People with brain injury also may have a problem with urination (bladder emptying) during the post-injury period. TBI may result in the inability to:

- Ask for help
- Control urination
- Recall when last urination occurred
- Plan ahead to get to the bathroom
- Walk to the bathroom in time
- Recognize the sensation of bladder fullness or the need to urinate

Premorbid conditions (such as an enlarged prostate in men or a pattern of infrequent urination in men or women) may add to bladder problems after brain injury.

The goals of bladder management include preserving kidney function, preventing incontinence (accidental urination), preventing bladder overfilling and bladder infections, establishing a regular patter of urination with complete bladder emptying, and maintaining dry, healthy skin in the genital area.

Problems with bladder management may include:

- Urinary retention (an inability to void or pass urine)
- Urinary incontinence
- Increased urgency to urinate
- Increased frequency of urination
- Incomplete emptying of the bladder
- Bladder infections
- Skin problems because of incontinence

If the person cannot sense the need to urinate, other approaches to bladder management are considered, including:

- Keeping the indwelling catheter in the bladder
- Intermittent catheterization (inserting and removing a catheter several times a day to regularly empty the bladder)
- Scheduling attempts at urination
- Using an external condom catheter for men
- Using an adult diaper
- Other methods of bladder retraining to control urination may be recommended

To maintain optimal bladder function, a person with brain injury should drink fluids as recommended by the dietitian or physician. The majority of individuals with brain injury regain the ability to regularly and effectively empty their bladder.

Intervention Addressing Cognitive Impairments

Interdisciplinary and comprehensive cognitive rehabilitation is an integral component of intervention with individuals who have sustained TBI. Even in those clients with good physical and medical recoveries, cognitive impairments in the area of attention, memory, and executive functions can limit the person's ability to engage in functional, social, and vocational activities. As the client emerges from the agitated phase of recovery and can more actively engage in the rehabilitative process, the occupational therapist begins to address cognitive impairments and the resulting limitations to functional activity performance and social participation more formally.

Cognitive Rehabilitation Approaches

Occupational therapists select cognitive rehabilitation intervention approaches that focus on the individual, the task, or the environment, on the basis of an understanding of the client's ability to learn and generalize information. When there is potential for change in the client's underlying cognitive impairments, the occupational therapist uses a remedial approach focused on improving and restoring the client's attention, memory, or executive function skills.

Functional approaches, which capitalize on the client's strengths and abilities, shift the focus of intervention from restoring underlying impairments to minimizing their limitations on engagement in activities and participation within the community. Therapists may modify the client's environment (e.g., placing cue cards or signs in key locations, labeling closets or drawers to identify their contents) or modify the activity (e.g., pre-selecting and pre-arranging items needed for completion of the task or presenting items for only one step of a task at a time). For clients with limited ability to learn and generalize information, a functional skill training program incorporating vanishing cues and errorless learning in the natural environment may be used.

Capitalizing on procedural memory, occupational therapists may teach the client to perform specific activities (e.g., completing morning hygiene, preparing simple snacks, following a medication schedule) that would decrease caregiver burden and provide some level of independence. Clients are trained on the same activity that they are expected to perform in the same environment in which it would be performed.)

Intervention Addressing Areas of Occupation

As the client emerges from the period of agitation, occupational therapy intervention can begin to focus on the client's performance of more client-centered areas of occupation, including ADLs (e.g., feeding, dressing, grooming, toileting, bathing, transfers) and instrumental activities of daily living (IADLs) (e.g., meal preparation, shopping, financial and home management, child rearing, caring for pets). The occupational therapist may delay focusing on some areas of occupation (e.g., work, education, leisure) until the client is transitioning back to his or her community.

Intervention approaches using adaptations to the task or environment require repetitive practice of the task and caregiver support and education on how to structure tasks and the environment for optimal performance. Although some adaptations to the environment may be fixed (e.g., a door alarm to prevent wandering or color-coded labels on the inside or outside of drawers and closets to reduce memory demands), other modifications depend upon the consistency and reliability of another person (e.g., filling and programming an alarmed pill box to aid the client in following a medication schedule). Throughout intervention focused on modifying and adapting tasks and the environment, the occupational therapist is acutely aware that acceptance of these interventions requires the client to accept a new vision of himself or herself and a willingness to accept change to the pattern or conditions of how he or she performs activities.

If the individual with TBI demonstrates potential for improvement in underlying cognitive and motor impairments, shows awareness of current limitations, and shows the ability to alter performance when provided cues and feedback, the occupational therapist may choose to focus intervention on restoring underlying cognitive and motor impairments that contribute to difficulties in the performance of functional tasks. Addressing executive-planning skills such as organization by developing cognitive strategies (e.g., checklists, self-monitoring strategies), occupational therapists may focus intervention on addressing underlying impairments to restore ability to enable more consistent, independent occupational performance. Application of cognitive strategies will be practiced in performance of a variety of ADLs and IADLs and individual and group intervention sessions to encourage generalization of the strategy to multiple areas of occupation.

A comprehensive occupational therapy intervention program addressing ADLs and IADLs considers multiple parameters that contribute to successful performance, including familiarity of the environment and the items used; the client's typical performance patterns (i.e., habits and routines); safety risks resulting from motor and cognitive-behavioral impairments; possible adaptive devices and compensatory techniques to improve performance; team and family support for implementation of the selected approaches; and the client's ability to monitor and correct performance.

Intervention During the Community Recovery Phase

Intervention Approaches Commonly Used During the Community Phase of Recovery (Rancho Los Amigos Levels VII-X) Interventions typically focus on higher cognitive skills and WHO ICF areas of activity limitations and participation restrictions.	
Intervention Approach	Occupational Therapy Treatment Activities
Prevent	• Prevent development of substance abuse and depression by educating the client about the risks and developing healthy alternative coping strategies. • Prevent client injury by modifying the home environment to decrease safety risks (e.g., installing grab bars and raised toilet seats, removing throw rugs and potential obstacles, installing automatic turn-off switches for stove burners and safety locks on cabinets).
Establish / Restore	• Restore cognitive and social communication skills by having the client plan and complete a community outing with family and friends; practice social pragmatics in group activities and role-playing. • Establish daily routines to enable the client to complete desired morning rituals in a timely manner and prevent late arrival at work or school. • Restore joint mobility and motor function after surgical excision of heterotopic ossification or botulism toxin injections for muscle spasticity. • Work with local brain injury support group to establish leisure skill program to increase social networks for community-dwelling individuals with TBI.
Modify	• Modify home and community environments to support independent performance of activities.
	• Modify daily routines to plan physically and cognitively challenging activities when well rested (e.g., pay bills in the morning when well rested; perform activities requiring fine motor demands when muscles are not fatigued). • Modify community mobility to accommodate for nondriving status.
Create / Promote	• Promote a healthy lifestyle that includes engagement in occupations that support physical and mental health (e.g., develop exercise program, create list of healthy food and meal selections, identify healthy social activities to foster social relationships.
Maintain	• Maintain gains made in ROM achieved by serial casting by wearing resting elbow splint for several hours per day. • Maintain social support systems in the community by engaging in leisure activities with friends.

Note: Rancho Los Amigos levels taken from Hagen, C. (1998). The Rancho Los Amigos Levels of Cognitive Functioning: The revised levels (3rd ed.). Downey, CA: Los Amigos Research and Educational Institute.

Adjustment to the variety of physical, cognitive, and neurobehavioral impairments resulting from TBI require functional coping skills from both the client and family. The ability to maintain existing friendships and develop new friendships is challenged when a person experiences a TBI. Impairments in cognitive and social skills, as well as limitations in the ability to engage in shared occupations, can result in the distancing of friends from the individual who sustained the injury.

Occupational therapy practitioners begin addressing social skills during inpatient rehabilitation, but often these impairments in social skills become more evident when the individual with TBI is discharged home to the community and reassumes social roles. In the community setting, the occupational therapy practitioners may conduct social skills training groups to address cognitive components of social interaction, the pragmatics of social conversation, and tasks involved in developing and maintaining friendships and relationships. Techniques such as goal setting, individualized written contracts, role playing and rehearsal, peer mentoring and role modeling, and videotaping social interactions with self-reflection and supportive feedback may be used in individual and group sessions.

Family-focused intervention may help the family unit manage the cognitive and neurobehavioral symptoms of their member with TBI upon return home to the community.

Intervention Addressing Social and Coping Skills

Adjustment to the variety of physical, cognitive, and neurobehavioral impairments resulting from TBI require functional coping skills from both the client and families. Family members and others close to a person with brain injury may struggle to cope with behavioral changes caused by the brain injury. The injured person also may struggle to adjust. Family members and others close to the person may feel stressed, burdened, even depressed by the major changes in activities, responsibilities, daily schedules, leisure and support that are required to adjust to the consequences of acquired brain injury. Often, major adjustments are best made in small steps. Simple changes may help the person with brain injury, family and friends to find more enjoyment in their relationships and activities.B y taking each stressful situation one step at a time, the person with brain injury and family may feel that life is becoming a little more "normal" again.

Self-esteem

Self-esteem is a person's assessment of self-worth that is often adversely affected by brain injury. The greater the self awareness the person with brain injury has, the more likely are negative changes in self-esteem. To counter the negative self worth felt by the client, the therapist needs to:

- constantly make the client express their feelings and focus on the positives.
- redirect conversation to positive or neutral thoughts.
- Express their concern and desire to understand the person's feelings.
- Point out the person's successes, even partial successes.
- Encourage as much independence as possible.
- Give caring, realistic feedback.
- Help the person plan ahead to maximize opportunities for success.
- Choose activities and tasks that the person can successfully complete.

Brain injury support group are vital links to education, life planning, and emotional support for clients and families. These community groups offer information on life planning and real-world problem solving. They also may offer leisure and social networking opportunities in groups where the client's neurobehavioral problems are more easily understood and accepted. Occupational therapists encourage clients and their families to connect to local brain injury support groups and may work with the groups to provide educational information sessions or develop programming to address leisure and social needs of the members.

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Individuals with TBI and impaired coping skills can show signs of depression and poorer outcomes. When impaired coping skills are coupled with neurobehavioral symptoms such as impulsivity, the person with TBI may be at greater risk for alcohol and drug abuse.

Depression

As confusion decreases and self awareness improves and as the person struggles to adjust to a temporary or lasting disability caused by TBI, he or she may suffer from depression. This might also occur if the injury has affected areas of the brain that control emotions. Depression is an illness (and not a sign of weakness, nor is it anyone's fault) caused by biochemical and structural changes in the brain. Some of the symptoms of depression are:

- Persistent sadness
- Irritability, moodiness
- Anxiety
- Loss of interest or pleasure in life
- Neglect of personal responsibilities or personal care
- Changes in eating habits or sleeping patterns
- Fatigue, loss of energy, lack of motivation
- Extreme mood changes
- Feeling helpless, worthless or hopeless
- Physical symptoms such as headaches or chronic pain that do not improve
- Withdrawal from others
- Thoughts of death or suicide

Fortunately, medication and other therapies can help most people who have depression. Effective treatments are available, including individual and group therapy, medication or a combination. Early treatment can help prevent needless suffering. Mental health professionals, including rehabilitation psychologists and social workers, can be consulted to provide help to both the client and to the immediate caretakers.

Family-focused intervention may help the family unit manage the cognitive and neurobehavioral symptoms of their member with TBI upon return home to the community.

Care for the Caretaker

Providing companionship and emotional support for the person with a brain injury may be necessary, in addition to physical care. Caregivers also may have many other responsibilities, including employment outside the home and caring for the home and children. Being a caregiver can be overwhelming, and adapting to these changes is challenging. The OT needs to prepare the primary caretakers regarding the varied emotional and physical challenges arising due to the differences and changes in circumstances in both the short term and over the long term and the ways to cope with the same: The occupational therapist can provide the following suggestions:

- Ask for help when needed: Caregivers frequently try to handle everything alone. Expecting too much of oneself may add to the stress. The therapist can provide suitable options for assistance such as home health care or respite care.
- Set limits. There are only so many hours in the day and only so many things a person can do. Its important to priortise and understand that some things can wait.
- the caretaker should plan something to look forward to each day for both the client and self.
- Take time away from the person being taken care for. Taking an hour, a day, a weekend or a week away can do wonders to restore emotional well being.
- Maintain contact with friends and family to discuss concerns or have fun.
- Take care of self. Caregivers are vulnerable to stress-related illnesses. Caretakers should also try to exercise since it increases stamina, lessens anxiety and depression, improves or maintains muscle tone and strength, and increases self confidence. These benefits make exercise a worthwhile use of limited time.
- Learn relaxation techniques such as breathing

exercises, meditation or progressive muscle relaxation.

- Join a support group. Support groups are an outlet for sharing problems and concerns. People with similar issues can not only empathise but also provide valuable suggestions on varied issues and coping techniques.

Sexuality

Love, affection and sexual feelings are healthy human desires. If these desires are not understood or expressed, confusion, distress and feelings of inadequacy may result. Sexuality involves the expression of male or female identities through sexual actions, attitudes and behavior in relationships. The ability to appropriately express these learned behaviors may be lost after a brain injury. Further, the adult with brain injury may not understand when it is appropriate to kiss, hug and touch. These changes in the client, who may act differently after the injury can cause feelings of confusion, anger, fear, frustration and helplessness in the family members.

Addressing relationships between client and family members should be done early in the rehabilitation process. The occupational therapist should empathise that it might be difficult for the client and or care takers to discuss sexual matters. The goals of rehabilitation for the person with brain injury include independence, self-reliance and healthy personal relationships.

The ability to develop and maintain social relationships may be the most important measure of successful rehabilitation. Therefore, recognizing and discussing concerns about love and sex are important. The closest family member of a person with brain injury is usually the most effective person to help the injured person relearn how to express sexual feelings appropriately. This family member may benefit from professional support and guidance in addressing sexual issues. It might have to be clarified that a person with brain injury might not recognize sexual cues, and hence not respond to a partner or would not initiate sexual activity. Social workers, rehabilitation nurses and rehabilitation psychologists are resources for support in addressing sexual issues and concerns.

Recreation and Leisure

Through leisure, basic human needs are met, including:

- Feeling good about ourselves
- Being part of a group
- Competing with ourselves and with others
- Experiencing success
- Laughing and having fun (reducing stress)
- Developing useful skills
- Developing friendships
- Strengthening social relationships

Participation in meaningful leisure experiences is essential to the recovery of the person with a brain injury. Most people naturally select certain activities that they enjoy and help them meet some basic needs. People with brain injuries may face barriers to experiencing this kind of fulfillment, including:

- Attitude (some people may not realize the importance of leisure activities)
- Physical disability (they may no longer be able to enjoy the activities they once did)
- Lack of cognitive skills (skills needed to participate in some activities - attention, concentration, initiation, planning, problem solving - may be impaired)
- Interruption of social and / or language skills
- Lack of knowledge (some may not know how to engage in certain leisure activities or how to adapt them so they can participate)

Occupational Therapist can help by:

- Helping the person identify leisure interests
- Assisting with structuring time and daily schedules so that leisure balances with necessary tasks and activities
- Planning ahead for recreation to keep life interesting
- Investigating community resources (city parks / recreation departments,libraries, churches, and other avenues for leisure options)

Intervention Addressing Occupational Performance

Although acute inpatient rehabilitation typically focuses on the performance of ADLs, individuals more than 10 years after their brain injury can show clinically significant improvements in functional skills when engaged in rehabilitation programs

focused on retraining specific skills or training of new skills previously not part of the client's roles. Using elements of procedural learning in a natural environment with no expectations for generalization or improvement in cognitive functioning, an occupational therapist may develop a program that incorporates errorless learning, practice of a specific task with fading cues, positive prompts, and praise and encouragement.

Occupational therapists also may need to assist the client and family in adapting strategies taught during acute inpatient hospitalization to sustain the same level of independence within the home environment. The natural cues offered by the familiar home and community environments may support greater independence, but these environments also challenge cognitive and physical skills due to their unpredictable nature.

Intervention Addressing Education and Work Activities

School reintegration and vocational rehabilitation are important aspects of community recovery for clients with TBI.

For children and teenagers, returning to school is important for both social and educational growth. At school, young people find friends and peer support, develop social skills, and increase their knowledge. School also provides a place to monitor young children's intellectual and social growth. Sometimes the effects of a brain injury are not apparent in young children but become more apparent in later years when the thinking and social demands at school increase.

Clients who wish to return to academic studies need to practice strategies that will support success in the student role. Occupational therapists may create simulated classroom instructional sessions for the client to practice taking notes and processing complex information, and review study habits and test-taking strategies. Cognitive orthotics such as personal digital assistants (PDAs), portable tape recorders, alarm watches, and laptop computers with scheduling software may be explored for their ability to compensate for residual cognitive impairments.

For those returning to high school and college, developing specific accommodations can help the person with brain injury to be successful in school. In most colleges, an office for students with disabilities assists in assuring that individual teachers provide such accommodations. Some common accommodations that may assist a person with brain injury to learn in school are:

- Extra time for tests to compensate for thinking or information processing that may be slower
- Taking tests privately in a distraction-free environment to accommodate for difficulties with attention, concentration and increased distractibility
- Placement in classrooms with less noise and distractions
- Tape recording lectures to compensate for attention, concentration and memory problems
- Access to teachers' or peers' class notes to compensate for difficulty in dividing attention between listening to a lecture and taking notes
- Tutoring with a peer or professional tutor

Cell phones, smart phones, and other forms of personal digital assistants can be used by clients with cognitive issues to remember essential information, navigate daily tasks, be reminded of appointments or times to take medication, and stay focused on treatment goals. Counselors can send the client text messages or voice reminders (these can be automated), and they can help the client program the devices to provide timer beeps, visual cues, maps, or other cognitive aids.

Work is defined as productive activity. It plays a major role in the lives of most people. Work may provide many benefits such as a sense of achievement, recognition, responsibility, financial independence, social interaction and structure. A brain injury can cause many changes in behavior, emotions and thinking skills. This can make it difficult to keep a job, even if it was the same job held before the injury. Regardless of whether or not the person with the brain injury returns to work, discovering how to use his or her talents to the best of his or her ability will make his or her life more rewarding. Those who return to work after brain injury are generally healthier and have a higher self-esteem than those who do not work.

Returning to work after a brain injury depends on a number of factors:

- Availability of jobs
- Health
- Desire to work

- Physical abilities
- Ability to adjust to changes
- Social and behavioral abilities (for example, ability to control behaviors and get along with co-workers)
- Thinking and problem-solving abilities
- Self-awareness of deficits and limitations
- Vocational interests and capabilities
- Willingness to receive further training
- The willingness of an employer to adapt the job or workplace

Returning to work after a brain injury can be challenging and rewarding. Case-coordinated early intervention focused on vocational skills can reduce unemployment among clients with TBI. Occupational therapists' unique ability to analyze task demands and environmental conditions and match these to the client's capabilities makes them well qualified to address vocational issues in individuals with TBI. Occupational therapists can provide job coaching, instruction, and education in safe work practices. They also may recommend modifications to job tasks, work hours, or work positions or may recommend specialized equipment or cognitive orthoses that enable efficient and accurate job performance. Those clients with TBI who are able to return to work activities may need additional coordinated interventions and support to sustain their work status.

Intervention Addressing Community Mobility

When the client is discharged to his or her home, issues of community mobility, and driving specifically, should be addressed with the client and family. Some occupational therapists specialize in driver rehabilitation and can assist the client and family in determining if and when a return to driving for community mobility is possible. Driving assessments should investigate the client's driving skills considering performance in clinic-based assessments as well as on-road evaluations to determine fitness to drive.

Holistic, intensive, and multidisciplinary neurorehabilitation can help individuals with TBI return to safe driving. Occupational therapists may use driving simulators both to evaluate the client's judgment, problem solving, and reaction times and to practice responding to simulated driving events in a safe although virtual environment. Clients who perform well in clinic-based and behind-the-wheel or on-road driving assessments typically participate in a trial of driver's training to practice and reinforce safe driving behaviors in gradually more challenging situations. Periodic follow-up on driving skills may be warranted.

For clients unable to resume independent driving, occupational therapists can provide intervention in use of alternatives for community mobility, including community-based transportation services for people with disabilities, taxi services, and public transportation systems.

Intervention Review

Intervention review is a continuous process of reevaluating and reviewing the intervention plan, the effectiveness of its delivery, progress toward targeted outcomes, and the need for future occupational therapy and referrals to other r professionals. Reevaluation may involve re-administering assessments used at the time of initial evaluation, a satisfaction questionnaire completed by the client, or questions that evaluate each goal. Reevaluation normally substantiates progress toward goal attainment, indicates any change in functional status, and directs modification of the intervention plan, if necessary. Because recovery from TBI involves multiple stages of client functioning and lengthy intervention, it is important for occupational therapists to periodically review the intervention plan to determine whether it reflects the client or family's current priorities, incorporates intervention approaches that meet those needs, and integrates current available evidence.

Discharge Planning, and Follow-Up

When clients with TBI are discharged from structured inpatient rehabilitation programs to their homes and communities, the true extent of their limitations may be revealed, often at a time when their financial and supportive resources are depleted. Occupational therapists' strength in analyzing and adapting functional tasks can be of great assistance in helping clients with TBI living in the community in resuming meaningful roles and occupations.

Conclusion

Since individuals with TBI present with many different physical, cognitive, and emotional impairments, therapists should be well versed in a

variety of treatment options and not just one approach. No matter which intervention technique is chosen, an emphasis should be placed on shaping the task to the patients abilities cognitive as well as physical to optimize success while progressively increasing the complexity and demands of the tasks and relating the intervention to a meaningful functional goal.

Rancho Los Amigos Scale

The Ranchos Los Amigos (Revised) Cognitive Scale is used by many health care teams to can begin treatment that will develop skills and promote appropriate behavior. Health care professionals often suggest the following simple measures to family and friends while the patient is still in coma:

- Always talk as if the patient hears when you are nearby.
- Speak directly to the patient about simple things and frequently reassure them.
- Explain events and noises in the surrounding area. Tell the patient what has happened and where they are.
- Touch and stroke the patient gently. Tell the patient who you are each time you approach the bedside. Hold their hand.
- Play the patient's favorite music for them.
- For parents of young children, tape yourself singing or reading your child's favorite stories.

Levels of Cognitive Functioning

Level I - No Response: Total Assistance

- Complete absence of observable change in behavior when presented visual, auditory, tactile, proprioceptive, vestibular or painful stimuli.

Level II - Generalized Response: Total Assistance

- Demonstrates generalized reflex response to painful stimuli.
- Responds to repeated auditory stimuli with increased or decreased activity.
- Responds to external stimuli with physiological changes generalized, gross body movement and/or not purposeful vocalization.
- Responses noted above may be same regardless of type and location of stimulation.
- Responses may be significantly delayed.

Level III - Localized Response: Total Assistance

- Demonstrates withdrawal or vocalization to painful stimuli.
- Turns toward or away from auditory stimuli.
- Blinks when strong light crosses visual field.
- Follows moving object passed within visual field.
- Responds to discomfort by pulling tubes or restraints.
- Responds inconsistently to simple commands.
- Responses directly related to type of stimulus.
- May respond to some persons (especially family and friends) but not to others.

Level IV - Confused/Agitated: Maximal Assistance

- Alert and in heightened state of activity.
- Purposeful attempts to remove restraints or tubes or crawl out of bed.
- May perform motor activities such as sitting, reaching and walking but without any apparent purpose or upon another's request.
- Very brief and usually non-purposeful moments of sustained alternatives and divided attention.
- Absent short-term memory.
- May cry out or scream out of proportion to stimulus even after its removal.
- May exhibit aggressive or flight behavior.
- Mood may swing from euphoric to hostile with no apparent relationship to environmental events.
- Unable to cooperate with treatment efforts.
- Verbalizations are frequently incoherent and/or inappropriate to activity or environment.

Level V - Confused, Inappropriate Non-Agitated: Maximal Assistance

Glasgow Coma Scale

The Glasgow Coma Scale is based on responses to stimuli in three areas: motor, verbal performance and eye opening. The scale assesses the depth and duration of coma and impaired consciousness. The Glasgow Coma Scale helps to gauge the impact of brain damage caused by traumatic and/or vascular injuries or infections, metabolic disorders, such as hepatic or renal failure, hypoglycemia, or diabetic ketosis.

Glasgow Coma Scale Eye Opening Response

- Spontaneous--open with blinking at baseline; 4 points
- To verbal stimuli, command, speech; 3 points
- To pain only (not applied to face); 2 points
- No response; 1 point

Glasgow Coma ScaleVerbal Response

- Oriented; 5 points
- Confused conversation, but able to answer questions; 4 points
- Inappropriate words; 3 points
- Incomprehensible speech; 2 points
- No response; 1 point

Glasgow Coma Scale Motor Response

- Obeys commands for movement; 6 points
- Purposeful movement to painful stimulus; 5 points
- Withdraws in response to pain; 4 points
- Flexion in response to pain (decorticate posturing); 3 points
- Extension response in response to pain (decerebrate posturing); 2 points
- No response; 1 point

Categorization

Coma: No eye opening, no ability to follow commands, no word verbalizations (3-8)

Head Injury Classification

Severe Head Injury -- Glasgow Coma Scale score of 8 or less

Moderate Head Injury -- Glasgow Coma Scale score of 9 to 12

Mild Head Injury -- Glasgow Coma Scale score of 13 to 15

PSYCHOLOGICAL ASPECT:

Introduction:

A person who sustains traumatic brain injury (TBI) can have dramatic and wide-reaching effects for both the person who sustained the injury and his or her caregivers. The effects of impairment include physical, cognitive, emotional behavioural and psychosocial disturbances depending on the areas of the brain affected and the severity of it [1]. These disturbances may affect the patients availability of social contact, return to work or school and changes in leisure activities. Patients with traumatic brain injury may also undergo personality changes and may lack awareness of, or would have difficulty in adjusting to post-injury outcomes.

Acute Consequences Post Traumatic Brain Injury:

A brain injury can be assumed as a head trauma, if it is accompanied by alteration in consciousness, neurological impairments or cognitive be deficits and can result from any object striking the head or the brain after coming in contact with the skull [2]. The injuries caused to the brain may be focal, multi focal or diffuse and can often involve structures away from the initial site of impact. The severity of the initial impact may be predictive of outcomes depending on the factors such as age, pre-existing conditions, psychological sequel and other factors also impact the long-term outcomes. The outcomes involve physical, cognitive and behavioural impairments, which may require prolonged hospitalization and post acute rehabilitation programmes.

Consequences of Traumatic Brain Injury:

The deficits that arise following a brain injury where physical deficits are visible, socially acceptable and may very often recover quickly. Cognitive impairments, emotional changes and behavioural problems may be less visible and are more likely to limit the range of a person's activity and impacts on the ability to reintegrate into the society. The deficits are listed below:

Cognition:

The commonly affected cognitive functions post traumatic brain injuries are: memory, impairment of attention, visual - spatial abilities and executive functions. These difficulties are compounded by lack of flexibility in attending, thinking and acting slowly and inefficiency in processing of information, difficulty with learning, poorly organised behaviour and verbal expressions.

Cognitive functions affected post TBI are as follows:

Memory:

Post brain injury it is often seen that there are mild, moderate and severe brain injuries due to impairment of all processes involved in memory

like encoding, maintenance and retrieval. Memory disturbances has a major impact on psychosocial outcomes and has been identified as an important predictor of work status, with severe verbal learning deficits often existing ten to twenty years post the injury.

Attention:

Attention problems most often include impairment of arousal, focused attention and divided attention. The persons alertness is affected i.e. the person's ability to focus on a particular stimulus and ignore other interfering materials if often affected. The person may experience difficulty to maintain a conversation in a noisy setting, or has impairment in reading complex instruction or has difficulty in perceiving the intentions and actions of others.

Language:

Impairments of language functions include deficits in fluency, understanding and naming objects. Language impairments may lead to talkativeness, verbal expansiveness, frequent use of words and phrase and eventually these may lead to emotional and behavioural changes. It is often seen that language difficulties in people post traumatic brain injury causes frustration, anxiety and the level of embarrassment increases.

Visual and Spatial Abilities:

The ability of a person to represent and manipulate spatial information is crucial for a wide range of perceptual, cognitive and motor functions [3].

Executive Functions:

Lezak (1995) [4] describes executive function as the ability which enables a person in independent and purposive behaviour. These include higher order skills such as problem solving, abstraction, concept formation, cognitive flexibility and self -regulation. Judgement of the patient may be impaired due to difficulties in scanning and assessing relevant components of a current situation and in controlling impulsivity. The reduced cognitive ability to perform complex actions may lead to disturbances in initiation and abstract reasoning.

Emotional and Behavioural Changes:

TBI can cause an impact on the persons emotional, behavioural and social functioning which leads to affecting the way a person behaves in social situations. Common behavioural deficits include reduced ability to initiate responses, verbal and physical aggression, learning difficulties, shallow self awareness, altered sexual functioning, impulsivity and social disinhibition. Mood disorders, personality changes, egocentricity, emotional liability, depression, anxiety and isolation are also prevalent after TBI [5]. Due to the affected cognitive difficulties, there could be problems that a person faces in social interaction. Hoofien and his colleagues (2001) reported that brain injury survivors and family members perceived their social functioning as being the most problematic, as compared to other areas.

Emotions and Motivation:

Post TBI the patients may experience impairments in emotions and may have adaptive function of providing sensations of comfort or discomfort that indicate whether a situation is safe or threatening, while motivation provides their impetus work towards a desirable goal. Emotions and motivations may be influenced by environmental factors, pre-morbid characteristics and neurotransmitter disturbances. Emotional reactions post injury would be mostly related to recognition of impaired abilities and a changed self-concept. Apathy, which is closely linked to motivation and emotion, has been described as diminished motivation that is not a result of diminished level of consciousness, cognitive impairment, or emotional distress, and in traumatic brain injury can result from disruption to a core circuit involving the anterior cingulum in the prefrontal cortex, and the nucleus accumbens, ventral pallidum and ventral tegmental areas (Marin & Chakravorty, 2005).

Mental Disorders Post Traumatic Brain Injury:

People with neurological injuries such as traumatic brain injury, have elevated risk of developing mental health disorders. The symptoms are most likely to worsen during the first six months post-trauma which include variability in mood, depression, emotional withdrawal, agitation/ hostility and apathy.

1. Depression:

Affective disorders are the most common psychological outcomes post traumatic brain injury. It is found that patients with traumatic brain injury are at "great risk" for developing depressive symptoms. The prevalence of major depressive disorder is between 15.6 percent and 6 percent (Kin at al., 2007). Certain factors were significantly related to the depression - time after injury, injury

severity, and post injury marital status. Therefore, the degree of initial injury, be it mild, moderate, or severe, can all lead to crippling depression. A person with traumatic brain injury may appear depressed, with symptoms of memory dysfunction, slowed movements, apathy, lack of initiation and blunted emotional expression in the absence of major depressive disorder. Usually, patients may feel worthless, helpless, frustrated and demonstrate loss of interest in work and family activities. Reactions to psychosocial changes may have an impact on prolonged or delayed onset depression.

2. Anxiety:

Individuals with traumatic brain injury may have reduced ability to adapt to their environment and as a result, are less likely to be able to manage anxiety or use it as a signal to indicate potential areas of threat. Another area of concern is social phobia identified as the most frequently occurring phobic disorder, which may be influenced by withdrawal from work and social situations. Obsessive compulsive disorder is another symptom which may appear as the person tries to achieve maximal control over his or her environment [6] with the development of some obsessive characteristics as an adaptation to impairment in the cognitive abilities such as memory and attention. Individuals with TBI may also experience Post Traumatic Stress Disorder, which is characterised by sleep disturbances, social withdrawal, ruminations, and hyperirritability.

3. Aggression:

Aggressive behaviours may result as a reaction to irritability or rise of organic factors. These may include damage to the limbic system, poor insight, reduced impulse control, or per injury personality traits. Aggression particularly involves physical assaults, which is especially directed towards the caregivers.

4. Personality Alterations:

Individuals post injury may have difficulties with frustration, problem solving, self- monitoring, concrete thinking, new learning and memory, impairment of judgement and poor insight. There could also be presence of inappropriate expression of affection, irritability, impulsivity and lack of motivation. Premorbid characteristics of the patient may frequently be exaggerated post injury, whereas other patients may show drastic alterations.

5. Suicide:

The association of depression post traumatic brain injury and suicide has been investigated with an emphasis on identification of risk factors. Suicide risk can continue for a number of years post injury.

Denial of Deficits:

Altered self awareness can be due to neurological and non neurological origin, such as social, emotional and motivational factors which can interact with organic problems to determine a person's level of self awareness. There can be two factors which contribute to the person's level of self awareness:

1. Organic Denial:

Organic denial can result from damage to certain areas of the brain which could disrupt the individual's self - concept and cognition. Consciousness and self awareness are the highest attributes of the frontal lobes (Stuss, 1991). Lack of awareness due to frontal lobe affection can have an impact on the rehabilitation activities, as the person will not perceive the need for rehabilitation if he or she does not recognize changes from pre-injury abilities.

2. Psychological Denial:

An emotional and protective response in the face of increasing emotional distress due to recognition of the disability may lead to denial. This is not necessarily harmful as persons post traumatic brain injury should be allowed to maintain their denial especially if it is not interfering with their rehabilitation. As, denial may help the person to pace his or her recovery following trauma by reducing excessive amounts of distress, with the need for denial, which would reduce the person's sense of self-competence and self-esteem.

Social Difficulties:

Individuals with traumatic brain injury experience a wide range of difficulties in terms of returning to their prior work situations, particularly those who were previously very successful and were holding higher positions at work. Emotional changes and disturbed behaviour were very often serious problems for relatives, while loss of social contact was the injured person's most disabling handicap in everyday life, with families and individual's often becoming isolated. A person's difficulties with interpretations and reaction to social situations, and

inappropriate content of speech may make social interactions generally unrewarding for others. Post the injury there may be decreased opportunities for establishing new social contacts and friends, and engaging in leisure activities, often due to a high incidence of people living at home with their families. [7]

Attempting to resume former social and work activities prematurely may lead to failure and rejections, which in turn may lead to increasing reliance on family members for support and a decline in satisfying relationships with peers. As people with traumatic brain injury become more isolated, they become more vulnerable to psychiatric disability.

Family Difficulties:

Traumatic Brain Injury affects the patient, caregiver and the family member. Family members often provide support and act as a caregiver for a prolonged period of time, and it is necessary to consider their psychological needs as well as those of the injured persons (Kay & Cavallo, 1994). The family members may also be in denial post trauma and they may have an unshakable belief in the injured person's potential for future recovery, underestimating changes and thinking of the person's potential for future recover, underestimating changes and thinking for the person they remember for before the injury, when they are often faced with different people. Denial should be taken care of when it prevents realistic planning for the future and it may serve as an adaptive function when it maintains family stability and role functions. Family members also undergo depression post the injury and there could be marital problems also, as the spouse may feel he or she is with a completely different person altogether who has aggressive or childish inappropriate behaviour. Divorce may often follow marital difficulties associated with traumatic brain injury. Families may have a prolonged mourning period which may eventually lead to negative feelings towards the injured person.

NEUROPSYCHOLOGICAL EVALUATION PROCESS:

Neuropsychological Assessment evaluates the cognitive processes and behaviours, using psychological testing to assess the central nervous system function and to diagnose specific behavioural or cognitive deficits or disorders. Neuropsychological assessments include sensitive tests that are used to detect subtle cognitive changes, severity of injury and improvement over time. Neuropsychological assessment may make a contribution to the differential diagnosis of neurobehavioral disorders, and the cumulative effect of multiple brain injuries.

i. In the acute setting neuropsychological consultation and assessment in moderate / severe brain injury is indicated for:

A) Determining emergence from post-traumatic amnesia

B) Documenting the early course of improvements in attention functioning, memory, visual-perceptual abilities, and language and executive functions. This information may be utilized in:

- Treatment planning and team consultation
- Family education / support
- Education and / or psychotherapy

ii. During the sub acute phase cognitive / physical stamina is reduced, availability for testing may be limited due to medical priorities, and other rehabilitation commitments.

Selective neuropsychological testing may be indicated to:

- Identify cognitive strengths and weaknesses
- Provide intervention such as psychotherapy
- Educate individual and family about TBI
- Assess or recommend behavioural management interventions

Based on an analysis of the profile of standardized scores and trained clinical observations of the individual's mental processes, evaluation is made regarding:

(1) Structural brain condition,

(2) Deficiencies caused by brain trauma versus other conditions such as pain, emotions, personality, and pre-injury conditions that contribute to functional status,

(3) Strengths in cognitive and psychosocial skills,

(4) Comprehensive diagnostic understanding of the physiological, psychological, and cognitive impact of the injury,

(5) Extent of injury and prognosis for recovery,

(6) Specific treatment needs along with identification of barriers to and assets for recovery,

(7) Objectively-based prognosis for return to work, school, and other activities,

(8) Foundation for life-care planning.

Psychological Assessments Tools for Traumatic Brain Injury:

The below listed tests are used to assess the cognitive deficits, behavioural changes and emotional disturbances in individuals with traumatic brain injury. These tests are conducted, scored and interpreted by psychologists or neuropsychologists.

1. Mini Mental Status Exam (MMSE):

The MMSE was developed to standardize and quantify the examination of an individual's cognitive state. It is used to screen for cognitive impairment and to follow individual's progress over time.

2. STROOP Neuropsychological Screening Test:

The STROOP NST is a test developed to predict the probability of an individual having organic impairment. It measures what is known as "higher executive function". The STROOP is a neurological screening test developed to predict the probability of an individual having brain damage.

3. Weschler Adult Intelligence Scale - III:

The WAIS-III is a standardized intelligence test which measures 15 different aspects of cognition and gives three different IQs: Verbal, Performance and Full Scale. The WAIS - III also offers four additional measures of cognition by analyzing verbal comprehension, performance organization, working memory and processing speed. The WAIS-III's use as a screening device for neuro-psychological impairment is well documented.

4. Weschler Memory Scales (WMS-III Abbreviated):

The Wechsler Memory Scale-Third Edition Abbreviated is a reliable survey of auditory and visual memory abilities. The WMS-III contains four subtests measuring auditory and visual immediate and delayed memory. This provides differences in patterns of memory performance have been found useful in discriminating among clinical groups with cerebral dysfunction or functional disorders resulting from various neuropathological or psychological processes.

5. MMPI-2:

The MMPI-2 consists of 567 items and is a self-administered standardized questionnaire. The MMPI-2 elicits a wide range of self-descriptions scored to give a quantitative measurement of an individual's level of emotional adjustment and attitude toward test taking. There are 10 major scales, four validity measures and several supplementary measures. The contents for the majority of the MMPI-2 questions are relatively obvious and deal largely with psychiatric, psychological, neurological and physical symptoms. The MMPI-2 has direct relevance to forensic applications and includes the test's ability to measure various symptoms of psychopathology.

6. Trail Making Test:

The TMT9 is a measure of attention, speed, and mental flexibility. It is brief, widely used by neuropsychologists, sensitive to TBI-associated cognitive impairment, and reliable.

7. Satisfaction With Life Scale:

The SWLS is a global measure of life satisfaction. The SWLS consists of 5 items that are completed by the subject.

8. Becks Depression Inventory (BBDI - II):

This is a 21-question multiple-choice self-report inventory, one of the most widely used instruments for measuring the severity of depression. Each answer is scored on a scale value of 0 to 3. The cut-offs used differ from:

0-13: minimal depression;
14-19: mild depression;
20-28: moderate depression;
29-63: severe depression.

Higher total scores indicate more severe depressive symptoms.

9. Stress: The Depression Anxiety Stress Scales (DASS):

This [8] is made up of 42 self report items to be completed over five to ten minutes, each reflecting a negative emotional symptom. [9] Each of these is rated on a four-point Likert scale of frequency.

These scores ranged from 0, meaning that the client believed the item "did not apply to them at all", to 3 meaning that the client considered the item to "apply to them very much, or most of the time". It is also stressed in the instructions that there are no right or wrong answers.

10. Quality Of Life Questionnaire: The SF-36:

This is a multi-purpose, short-form health survey with only 36 questions. It yields an 8-scale profile of functional health and well-being scores as well as psychometrically-based physical and mental health summary measures and a preference-based health utility index.

PSYCHOLOGICAL REHABILITATION POST TRAUMATIC BRAIN INJURY:

Goals:

The goals for individuals post traumatic brain injury are to improve the person's ability to function by enhancing his or her capacity to organise the daily activities, to attend to and process information and to interact with others in a socially appropriate manner. It would basically focus on improving the overall quality of life of the patient and the caregivers. This could be done with the help of clinical psychologists and neuropsychologists who would perform assessments and depending on the results of the tests would make recommendations for future rehabilitation programmes. Neuropsychological assessment is necessary prior to implementation of cognitive remediation, in order to provide a person's areas of relative strength and weakness. The psychologists on the other hand would provide psychotherapy services to people with traumatic brain injury and their families [10].

Cognitive Rehabilitation:

The main aim of cognitive rehabilitation is to improve the individual's functional and integrative performance, where this is impaired by the individual's cognitive deficits. Restorative training focuses on improving a specific cognitive function, whereas compensatory training focuses on adapting to a deficit [11]. First the neuropsychologists assess the cognitive strengths and weakness and then the patient is provided with learning compensation techniques or new ways of doing things.

Complementary techniques may focus on the use of external methods such as cue cards, watch alarms, diaries, address books and computers to record notes, thoughts and other data, can be used to address a number of cognitive deficits.

Internal strategies may involve self-instructional routines that regulate behaviours using the inner speech. Self- instructional routines can help in memory training and attentional deficits by helping to maintain focus on specific tasks. Self instructional techniques can help with initiating actions, planning and problem solving to patients who have executive function deficits due to which they demonstrate inappropriate social and work behaviour, particularly when they are in environment or engaged in activities that are not routine. Patients should learn to apply compensatory strategies in functional situations when they are suffering from cognitive impairments as this is an important part of cognitive rehabilitation.

Behavioural Rehabilitation:

As the focus for individuals with traumatic brain injury is physical recovery, with less emphasis of their ability to adapt to social behaviour and changing situations. However, even after significant changes in the physical functions it is very difficult to reintegrate the individuals into the community due to the behavioural and emotional changes. Behaviour Modification techniques could be very helpful for patients with traumatic brain injury as the focus of this technique would be to address the inappropriate social behaviour, attention and motivation, lack of awareness, memory, language and motor disturbances (McGlynn, 1990). Professionals should come up with an individualised plan for the patient by first assessing the behavioural issues and then apply strategies to increase or decrease particular behaviours.

Behavioural and Cognitive Behavioural Interventions:

Behavioural approaches can include behaviour modification techniques and skills' training that addresses personality and behavioural effects from traumatic brain injury. These techniques may also be incorporated within a Cognitive Behavioural Therapy (CBT) approach. The cognitive approaches to psychopathology regards the processing of external events as biased which distorts the individual's evaluation of experience which leads to a variety of cognitive errors, for example, over-generalisation, selective abstraction and personalisation(Beck, 2005).

The main aim of cognitive behaviour technique is to identify the underlying distorted beliefs that have been incorporated into enduring schemas or core belief systems. The psychologist records the patient's negative automatic thought patterns and conditional core beliefs. Behavioural techniques can be used in the early stages of therapy as some people with brain injury often operate at a very concrete level (Cicerone, 1989; Khan -Bourne & Brown, 2003), while cognitive techniques help to identify the person's beliefs about the current situations and abilities. CBT aims to break the negative cycles that maintain depression following the brain injury, by identifying the maladaptive strategies, and promoting more adaptive behaviours. Post TBI, there is a reduced control over mood shifts and impulsive behaviour and this may have a negative impact on a person's sexuality. CBT provides in individual therapy or group therapy to improve the sexual and interpersonal functioning.

Managing Attention Difficulties:

An option to manage attention difficulties would be to use compensatory strategies and environmental support.

The following strategies are very helpful:

1. Orienting procedure:

This involves monitoring an activity by checking what is involved at each step. For example things needed to make a sandwich, listing down the items needed to make a sandwich, the procedure used to make a sandwich and eventually checking it.

2. Pacing:

As there is mental fatigue while maintaining attention for a prolonged period of time, the pacing strategy suggests that one needs to develop a realistic expectation about what can be achieved over a period of time.

3. Keeping Notes:

Patients can learn to jot down the key questions or ideas that come to mind which can be useful to solve a problem; it's very difficult for patient with TBI to switch between tasks.

4. Changing the surroundings:

Making a surrounding at home or workplace which would be easy accessible: For example:

- Getting rid of clutter
- Arranging things needed in one cupboard.
- Labelling, the cupboards and jars.
- Setting up filling systems
- Getting rid of distractions.

Managing Memory Difficulties:

Memory functions on the coordination of a number of processes i.e. attention, encoding, storage and retrieval.

Strategies for Memory Difficulties: (Further explained in psychological chapter).

- Learning more effectively
- Making use of mnemonics
- Making use of external aids like alarms, tags, memory diaries, organisers, etc.

Managing Aggression and Irritability:

A common complaint following TBI of patients would be the inability of the patients would be their reduced ability to manage frustration, anger and aggression. The psychologist working with the patient should record his level or irritability, frustration, anger and aggression. Along with this a note should be made as to what are the situations in which the person gets angry and the severity of responses in such situations.

Anger management strategy:

A - Anticipate the trigger situations

N - Notice the signs of the rising anger.

G - Go through your rising temper by using relaxation techniques.

E - Extract yourself from the situation, if everything fails.

R - Record how you coped.

ASPECTS OF COGNITION AND PHASES OF IMPROVEMENT POST TRAUMATIC BRAIN INJURY: [12]

Aspects of Cognition:

1. Attention:

This is in terms of holding objects, events, words or thoughts in consciousness. Cognitive components related to attention are attention span, filtering, selectivity, filtering, maintaining, shifting and dividing.

Early Phase: During the early phase of Traumatic Brain Injury:

- Patient shows severely decreased arousal or alertness.
- Patient has minimal selective attention and has difficulty in shifting his focus.
- Attention would be present primarily due to internal stimuli.

Middle Phase: During the middle phase of Traumatic Brain Injury:

- Attention is generally focused on external events
- The patients attention span is short
- Patient finds great difficulty in fixing his attention and is usually highly distracted.

Late Phase: During the late phase of Traumatic Brain Injury:

- Patient's attention span is much reduced
- Concentration is relatively weak, attention is selective and there are fluid attention shifts.
- The patient finds difficulty organizing things or thoughts and there is absence of goal.

2. Perception:

Perception is recognizing features and being able to find relationship between events, thing, etc. Where the aspects which are taken into consideration while perceiving a situation or object is intensity, duration, significance, context and familiarity of the stimuli.

Early Phase: During the early phase of Traumatic Brain Injury:

- Patient begins to recognize and use familiar objects.
- When exposed to a stimuli the patient may be able to recognize only one aspect of the stimuli
- Patients may start adapting to continuous stimulation

Middle Phase: During the Middle phase of Traumatic Brain Injury:

- Patient can clearly recognize familiar objects and events.
- Patients undergo sharp deterioration as there as an increase in the complexity of stimuli.
- Patients have difficulty in distinguishing whole form the part.

Late Phase: During the Late phase of Traumatic Brain Injury:

- Patients could undergo subtle versions of perceptual problems related to complexity, rate and amount.
- Patients could have inefficient shifting of perceptual set.
- Patients usually have weak perception of relevant feature.

3. Memory and Learning:

This consists of recognition, interpretation and formulation of information including internal code. Coding is affected by knowledge, personal interests and goals.

Early Phase: During the Early phase of Traumatic Brain Injury:

- There is progression in comprehension from minimal responses to vocal modulation and stress to recognition of simple, context bound instructions.
- There is however no evidence of encoding or storage of new information.

Middle Phase: During the Middle phase of Traumatic Brain Injury:

- Patients undergo weak encoding due to poor access to knowledge base, poor integration of new with old information, or inefficient attention or perception.
- Inefficiently encoded information is often lost after short delay.
- Recognition is stronger than recall i.e. receptive vocabulary is superior to expressive vocabulary.
- Patients often have difficulty in organized search of storage system.

Last Phase: During the Last phase of Traumatic Brain Injury:

- Patients have increase in cognitive stress depending on the level of cognitive functioning prior to TBI.
- Patients experience memory problems related to recalling information related to personal experience or abstracted knowledge.

4. Organizing or analyzing ability:

We classify, integrate, and sequentially arrange

relevant features of objects and events; comparing for similarities or differences by integrating into organized description. These processes are presupposed by higher level reasoning and efficient learning.

Early Phase: During the Early phase of Traumatic Brain Injury:

- There is no evidence of significant improvement in this phase.

Middle Phase: During the Middle phase of Traumatic Brain Injury:

- Patients have weak or bizarre association and analysis of objects into features.
- Patients have disorganised sequencing of events.
- Patients are poor at identifying similarities and differences in comparisons and classifications.
- Patents can integrate concepts into propositions but they have difficulty in integrating propositions into main idea.

Last Phase: During the Last phase of Traumatic Brain Injury:

- Patients have difficulty in maintaining goal directed thinking.
- They easily get lost into details.
- They can organize unstructured stimuli with prompts.

5. Reasoning:

Reasoning means drawing inferences and conclusions involving flexible exploration of possibilities and making use of past experiences. There are 3 types of reasoning: deductive, inductive and analogical reasoning.

First Phase: During the First phase of Traumatic Brain Injury:

- There is no major evidence of improvement in these processes.

Middle Phase: During the Middle phase of Traumatic Brain Injury:

- There is minimal inferential thinking in the patients which may help deal with concrete cause and effect relationship.
- There is general inefficiency that the patient experiences in abstract ideas and relationships.

Last Phase: During the Last phase of Traumatic Brain Injury:

- The patients have fair to good concrete reasoning in controlled setting and disorganized thinking in stressful or uncontrolled setting.
- Patient's abstract thinking is deficient.

6. Problem Solving and Judgement:

This takes place when a goal cannot be achieved directly. Ideally goals involve identification, consideration of relevant information, exploration of possible solutions and selection of the best solution. Judgement is based on the way a person decides over other options.

Early Phase: During the Early phase of Traumatic Brain Injury:

- No evidence of substantial improvement in the early phase has been seen.

Middle Phase: During the Middle phase of Traumatic Brain Injury:

- The patient finds it difficult to see relationships among problems, goals and relevant information.
- There is inflexibility in generating or evaluating possible solutions and impulsive trial and error approach.
- Patients usually have severely impaired safety and social judgement.

Last Phase: During the Last phase of Traumatic Brain Injury:

- Patients are usually impulsive, disorganised and have difficulty in problem solving in this phase.
- They have inflexible thinking and shallow reasoning.

7. Long term Memory:

This contains of knowledge of concepts, words, rules, strategies, procedures and organizational principles.

Early Phase: During the Early phase of Traumatic Brain Injury:

- There are emerging evidences of remote memory, recognition of familiar objects and persons improvement in patients with TBI in this phase.

Middle Phase: During the Middle phase of Traumatic Brain Injury:

- Patients have recognition of strong associations of basic semantic relations and common two or three event sequences.

Last Phase: During the Last phase of Traumatic Brain Injury:

- Patients undergo variable growth of long term memory, depending on type and severity of residual cognitive deficits.

8. Response Systems:

This controls all output including speech, facial expression and fine and gross motor activity including motor planning.

Early Phase: During the Early phase of Traumatic Brain Injury:

- The patients are grossly limited and often have preservative responses.

Middle Phase: During the Middle phase of Traumatic Brain Injury:

- The patients during this stage usually speak or begin with augmentative systems.

Last Phase: During the Last phase of Traumatic Brain Injury:

Patients in this phase usually have functional communication system.

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Ch.5 Autism

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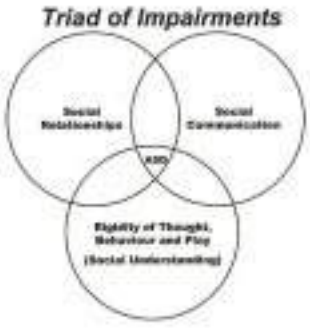
History

In 1943, Leo Kanner, in his classic paper "Autistic Disturbances of Affective Contact" coined the term infantile autism and provided a clear, widespread account of early childhood syndrome. Kanner suspected that the syndrome was more frequent than it seemed and suggested that some children with the disorder had been misclassified as mentally retarded or schizophrenic. Before 1980, children with pervasive developmental disorder were generally diagnosed with childhood schizophrenia. Over time, it became evident that autistic disorder and schizophrenia are two distinct psychiatric entities. [1]

Definition:

- The Diagnostic and Statistical Manual of Mental Disorders, DSM-IV (American Psychiatric Association, 1994) defines autism spectrum disorders as pervasive developmental disorders characterized by:
  - qualitative impairment in social interaction
  - qualitative impairment in communication
  - restricted, repetitive and stereotypic patterns of behaviour, interests and activities.



Triad of Impairment in Autism

- Autism is diagnosed when a child or adult has abnormalities in a 'triad' of behavioural domains: social development, communication, and repetitive behaviour / obsessive interests (APA, 1994; ICD-10, 1994).
- Autism Society of America (ASA) defined autism as: "Autism is a complex developmental disability that typically appears during the first three years of life and is the result of a neurological disorder that affects the normal functioning of the brain impacting the development in the areas of social interaction and communication skills. Both children and adults with autism typically show difficulties in verbal and non-verbal communication, social interactions and leisure or play activities. Autism is one of the five disorders that fall under the Pervasive Developmental Disorder (PDD), a category of neurological disorders characterized by "severe and pervasive impairment in several areas of development".

Epidemiology:

Prevalence:

Autistic disorder is believed to occur at a rate of about 8 cases per 10,000 children (0.08 percent) and it continues to grow at an alarming rate however, because of inadequate data, these numbers may underestimate autisms true prevalence. By definition, the onset of autistic disorder is before the age of 3 years, although in many cases it noticed at a much older age. [1]

Sex Distribution:

It is seen that autistic disorder is four to five times more common in boys than in girls. However, girls with autistic disorder are more probable to have more severe mental retardation as compared to boys.

Socio economic Status:

Over the past 25 years, no epidemiological studies have demonstrated an association between autistic disorder and any socioeconomic status.

Etiology and Pathogenesis:

1. Genetic Factors:

Current evidence supports a genetic basis for the development of autistic disorder in most cases. Family studies have demonstrated a 50 to 200 times

increase in the rate of autism in siblings of an index child with autistic disorder. Studies suggest that even if the siblings are not affected with autism they are an increased risk for a variety of developmental disorders which are often related to communication and social skills. Linkage analyses have demonstrated that regions of chromosomes 7, 2, 4, 14 and 19 are likely to contribute to the genetic basis of autism.

The concordance rate of autistic disorder in the two largest twin studies was 36 percent in monozygotic pairs versus 0 percent in dizygotic pairs in one study and about 96 percent in monozygotic pairs versus about 27 percent in dizygotic pairs in the second study. Approximately 1 percent of children with autistic disorder also have fragile X syndrome, who tend to show gross motor and fine motor difficulties as well as relatively poorer expressive language compared with children with autism without fragile X syndrome.

Recent research was conducted where, the DNA of more than 150 pairs of siblings with autism was analyzed and they found extremely strong evidence that two regions on chromosome 2 and 7 contain genes involved with autism. The likely locations for autism - related genes were found on chromosome 16 and 17, although the strength of correlation was somewhat weaker. [1]

2. Biological Factors:

Approximately 70 percent of children diagnosed with autistic disorder have mental retardation. About one third of children with autistic disorder have mild to moderate and close to half of these children are severely or profoundly mentally retarded. Children with co-morbid autistic disorder and mental retardation display marked deficits in social understanding, abstract reasoning and verbal tasks than in performance tasks, such as digit recall and block design.

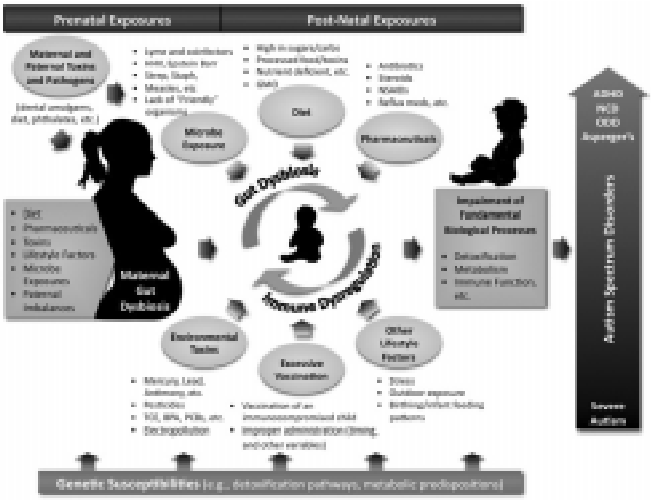
Of persons with autism, 4 to 32 percent have grand mal seizures at sometime and about 20 to 25 percent show ventricular enlargement on computed tomography (CT) scans. Various electroencephalogram (EEG) abnormalities are found in 10 to 83 percent autistic children and although no EEG finding is specific in autistic disorder, there is some indication of failed cerebral lateralization. [1]

3. Immunological Factors:

The lymphocytes of some autistic children react with maternal antibodies, which raise the possibility that embryonic neural or extra embryonic tissues may be damaged during gestation. [1]

4. Perinatal Factors:

A higher than expected incidence of perinatal complications seems to occur in infants who are later diagnosed with autistic disorder. Maternal bleeding after the first trimester and meconium in the amniotic fluid has been reported in the histories of autistic children than the normal population. It is seen that in the neonatal period, autistic children have a high incidence of respiratory distress syndrome and neonatal anemia. Males with autism, as a group have been found to be the products of longer gestational age and were heavier babies than babies in the general population. On the other hand females with autism are more likely to be the product of post term pregnancies than babies in the general population. [1]



Causal Factors of Autism i.e. pre natal and posy natal effects

5. Neuroanatomical Factors:

The neuroanatomical basis of autism remains unknown; however, recent evidence suggests that enlargement of the gray and white matter cerebral volumes, but not at 2 years of age. It is seen that the head circumference appears normal at birth; however the head circumference growth appears to emerge at about 12 months of age. MRI studies, where the brain volume of the autistic subjects and normal controls was compared the results revealed that the brain volume was larger in those with autism, however it seen that autistic children with severe mental retardation generally have smaller heads. The greatest average percentage increase in size occurred in the occipital lobe, parietal lobe and temporal lobe, however no difference was found in the frontal lobe. The increased volume can occur from three different possible mechanisms: increased

neurogenesis, decreased neuronal death and increased production of non-neuronal brain tissue, such as glial cells or blood vessels. Brain has been suggested as possible biological marker for autistic disorder.

The temporal lobe damage is reported to be one of the critical areas of the brain abnormality in autistic disorders as also animal studies reveal that when the temporal region of the animals is damaged, normal social behaviour is lost, there is presence of repetitive motor behaviour and restlessness. Some brains of autistic individuals exhibit a decrease in cerebellar Purkinje's cells, which is believed to account potentially for abnormalities of attention, arousal and sensory processes.

Areas of the Brain Affected in Autism

An interesting study, which reveals the differences between male and female brains suggests the capacity to predict and respond to feelings and behaviour of others by inferring their emotional states, is a stronger trait in females than in males are a population level i.e. empathizing. On the other hand male population is stronger at systemizing, that is, inferring rules that govern "cause and effect" relationships of behaviour. People with pervasive developmental disorders are characterized by deficits in empathizing and those with high intellectual capacity have been reported to have relative strengths in rule bound thinking.

A neural basis of empathy or social intelligence was first proposed by Brothers (Brothers, 1990) [2]. She suggested from animal lesion studies (Kling & Brothers, 1992), single cell recording studies (Brothers, Ring, & Kling, 1990) [3], and neurological studies that social intelligence was a function of three regions: the amygdala, the orbito-frontal cortex (OFC), and the superior temporal sulcus and gyrus (STG). Together, she called these the "social brain".

The amygdale:

There are four lines of evidence for an amygdala deficit in autism (Baron-Cohen et al; 2000) [4]

(a) Post-mortem evidence:

A neuroanatomical study of adults with autism at post-mortem found microscopic pathology (in the form of increased cell density) in the amygdala, in the presence of normal amygdala volume (Bauman & Kemper, 1994; Rapin & Katzman, 1998) [5]

(b) Similarities between autism and patients following amygdalotom:

Patients with amygdala lesions show impairments in social judgement [6] that have been likened to "acquired autism" (Stone, 2000). People diagnosed with autism tend to show a similar pattern of deficits to those seen in patients with amygdala lesions (Adolphs, Sears, & Piven, 2001).

(c) Structural neuroimaging:

A recent structural magnetic resonance imaging study of autism reported reduced amygdala volume (Abell et al; 1999) [7].This is not the only structural abnormality in the brain (see below), but the amygdala abnormality has some potential relevance to the social symptoms observed. It is not yet known why this difference occurs.

(d) Functional neuroimaging:

Using single photon emission computed tomography (SPECT), patients with autism spectrum conditions show significant reductions in temporal lobe blood flow. This is not simply an effect of temporal lobe epilepsy (Gillberg, Bjure, Uvebrant, Vestergren, & Gillberg, 1993). In a recent functional magnetic resonance imaging (fMRI) study, adults with High Functioning Autism (HFA) or Asperger Syndrome (AS) showed significantly less amygdala activation during a mentalizing task (Reading the Mind in the Eyes task), compared to normal (Baron-Cohen, Ring et al; 1999).

Other brain areas that might be abnormal in autism:

Reduced neuron size and increased cell-packing density has also been found in the limbic system, specifically the hippocampus, subiculum,

entorhinal cortex, amygdala, mammillary bodies, anterior cingulate, and septum in autism [8].

6. Biochemical Factors:

A number of studies report that about one third of patients with autistic disorder have high plasma serotonin concentrations, however, this is not specific to autistic disorder as children with mental retardation without autistic disorder also display this trait. In some autistic children, a high concentration of homovanillic acid in the brospinal fluid (CSF) is associated with increased withdrawal and stereotypes. Some evidence indicated that symptom severity decreases as the ratio of 5-hydroxyindoleacetic acid (5 - HIAA, metabolite of serotonin) to homovanillin acid in CSF increases. The 5 - HIAA concentration in CSF may be inversely proportional to blood serotonin concentration, which is increased in one third of autistic disorder patients, a non - specific finding that also occurs in mentally retarded persons.

7. Psychosocial and Family Factors:

Studies comparing parents of autistic children with parents of normal children have shown no significant differences in child- rearing skills. It is seen that children with autistic disorder respond like children with other disorders respond to exacerbated symptoms like family discord, the birth of a new sibling or a family move, etc. Some autistic children may be extremely sensitive to even small changes in the families and immediate environment. While making the diagnosis the evaluators should consider prenatal history, perinatal history, birth complications, developmental milestones and outlining family history via interviews with the parent or relevant caregivers with the help of formal and informal assessment. Autistic disorder can be diagnosed by a physician, a psychologist, or a psychiatrist. Specific tools can be used to check for ASD.

Diagnosis and Clinical Features:

Autistic disorder has been diagnosed using the Diagnostic Stastical Manual - IV th Edition - Text Revision, this is a manual with diagnostic categories approved by the American Psychological Association and International Statistical Classification of Diseases and Related Health Problems, tenth revision (ICD-10) published by The World Health Organization (WHO). In 1994, when the fourth edition of DSM was published, five categories appeared under the Pervasive Developmental Disorder (PDD) heading: Autism, PDD-NOS, Asperger's syndrome, Rett's syndrome, and childhood disintegrative disorder. PDD is an umbrella term for disorders that involve impairments in reciprocal social interaction skills and communication skills, and the presence of stereotypical behaviours, interests and activities. The term Autistic Spectrum Disorder is used to represent the fact that while these individuals share common characteristics, how these characteristics are manifested will differ with each individual. As a result, no two individuals are the same.

It is sometimes complicated diagnosing autistic disorder because there are no medical tests, blood tests or any radiological images which will definitively indicate or diagnose a person with autistic disorder. Also it could be difficult as the person has other issues related to medical, emotional, sensory or learning difficulties. An accurate diagnosis is made on the basis of specific intellectual, social and behavioural characteristics that are listed in the DSM - IV - TR Manual. The diagnosis can be made by a physician, a psychologist, or a psychiatrist.

The DSM - IV - TR diagnostic criteria for autistic disorder are as follows:

Diagnostic code according to DSM - IV - TR: 299.00

A. A total of six (or more) items from (1), (2) and (3), with at least two from (1) and one each from (2) and (3):

(1) Qualitative impairment in social interaction as manifested by at least two of the following:

- a) Marked impairments in the use of multiple nonverbal behaviors such as eye-to-eye gaze, facial expression, body posture, & gestures to regulate social interaction
- b) Failure to develop peer relationships appropriate to developmental level
- c) A lack of spontaneous seeking to share enjoyment, interests, or achievements with other people, (e.g; by a lack of showing, bringing, or pointing out objects of interest)
- d) Lack of social or emotional reciprocity

(2) Qualitative impairments in communication as manifested by at least one of the following:

- a) Delay in or total lack of, the development of spoken language (not accompanied by

an attempt to compensate through alternative modes of communication such as gesture or mime)

b) In individuals with adequate speech, marked impairment in the ability to initiate or sustain a conversation with others

c) Stereotyped & repetitive use of language or idiosyncratic language

d) Lack of varied, spontaneous make-believe play or social imitative play appropriate to developmental level

(3) Restricted repetitive & stereotyped patterns of behavior, interests & activities, as manifested by at least two of the following:

a) Encompassing preoccupation with one or more stereotyped & restricted patterns of interest that is abnormal either in intensity or focus

b) Apparently inflexible adherence to specific, nonfunctional routines or rituals

c) Stereotyped & repetitive motor mannerisms (e.g. hand or finger flapping or twisting, or complex whole-body movements)

d) Persistent preoccupation with parts of objects

B. Delays or abnormal functioning in at least one of the following areas, with onset prior to age 3 years:

1) social interaction

2) language as used in social communication

3) symbolic or imaginative play

C. The disturbance is not better accounted for by Rett's Disorder or Childhood Disintegrative Disorder

(From American Psychiatric Association. Diagnostic and Statistical Manual of Mental Disorders. 4th ed. Text rev. Washington, DC: American Psychiatric Association; copyright 2000) [9].

The International Statistical Classification of Diseases and Related Health Problems, tenth revision (ICD-10) diagnostic criteria for autistic disorder are as follows:

Diagnostic code according to ICD - 10: F84.0

A type of pervasive developmental disorder that is defined by: (a) the presence of abnormal or impaired development that is manifest before the age of three years, and (b) the characteristic type of abnormal functioning in all the three areas of psychopathology: reciprocal social interaction, communication, and restricted, stereotyped, repetitive behaviour. In addition to these specific diagnostic features, a range of other nonspecific problems are common, such as phobias, sleeping and eating disturbances, temper tantrums, and (self-directed) aggression.

Autistic disorder

Infantile:

- autism
- psychosis

Kanner's syndrome

Excl.: autistic psychopathy (F84.5)

Atypical autism: F84.1

A type of pervasive developmental disorder that differs from childhood autism either in age of onset or in failing to fulfill all three sets of diagnostic criteria. This subcategory should be used when there is abnormal and impaired development that is present only after age three years, and a lack of sufficient demonstrable abnormalities in one or two of the three areas of psychopathology required for the diagnosis of autism (namely, reciprocal social interactions, communication, and restricted, stereotyped, repetitive behaviour) in spite of characteristic abnormalities in the other area(s). Atypical autism arises most often in profoundly retarded individuals and in individuals with a severe specific developmental disorder of receptive language.

Atypical childhood psychosis

Mental retardation with autistic features

Use additional code (F70-F79), if desired, to identify mental retardation.

Other Pervasive Development Disorders:

The diagnostic category known as Pervasive Developmental Disorders includes: Autistic Disorder, Asperger's Disorder, Rett's Disorder, Childhood Disintegrative Disorder and Pervasive Developmental Disorder Not Otherwise Specified. All of these disorders share common features.

However, there are differences in some areas, such as the number of symptoms, age of onset, developmental pattern and level of cognitive functioning.

Physical Characteristics:

Children with autistic disorder do not show any physical signs indicating the disorder on the first glance. A greater than expected number of children do not show lateralization and remain ambidextrous at an age when the cerebral dominance is established in most children.

Seizures:

It is estimated that 25% of autistic individuals also develop seizures, some in early childhood and others as they go through puberty (changes in hormone levels may trigger seizures). These seizures can range from mild (e.g; gazing into space for a few seconds) to severe, grand mal seizures.

Many autistic individuals have subclinical seizures which are not easily noticeable but can significantly affect mental function. A short one- or two-hour EEG may not be able to detect any abnormal activity, so a 24-hour EEG may be necessary. Although drugs can be used to reduce seizure activity, the child's health must be checked regularly because these drugs can be harmful.

Sleep Problems:

Many autistic individuals have sleep problems. Night waking may be due to reflux of stomach acid into the oesophagus.

Pica:

30% of children with autism have moderate to severe pica. Pica refers to eating non-food items such as paint, sand, dirt, paper, etc. Pica can expose the child to heavy metal poisoning, especially if there is lead in the paint or in the soil.

Low Muscle Tone:

A study conducted by the first author found that 30% of autistic children have moderate to severe loss of muscle tone, and this can limit their gross and fine motor skills. That study found that these children tend to have low potassium levels.

Chronic Constipation and/or Diarrhoea:

Analyses of the ARI's autism database of thousands of cases show over 50% of autistic children have chronic constipation and / or diarrhoea. Diarrhoea may actually be due to constipation-i.e; only liquid is able to leak past a constipated stool mass in the intestine. Manual probing often fails to find an impaction. An endoscopy may be the only way to check for this problem. Consultation with a paediatric gastroenterologist is required.

Behavioural Characteristics:

Difficulties in Social Skills:

Have you ever made a mistake in social situations? or Told a joke that did not fit the event? Or misread the intentions of others? Then it is easier to understand why individuals with autism have difficulty in social situations. Social situation are very demanding as they need us to make quick judgements, respond to unpredictable events, read facial expressions and body gestures of others. For individuals with autism, these skills are missing or are functioning at a deficit.

Social difficulties are the primary that individuals with autistic disorder face challenge being employed or in employment. As when young they usually have minimal interest in playing or interacting with peers, hence when social engagement occurs it is typically on their own terms or it gets very awkward. These responses reflect a lack of skill in knowing how to interact rather than a lack of desire in socialization. Also as they stick to their ritualistic routine, they have difficulty in changing or alternating routine for which they need additional support and transitioning time to new and unfamiliar experiences. [1]

Poor Eye contact

Autistic children do not display the expected level of subtle reciprocal social skills that demonstrate relatedness to parents and peers. Children with autistic disorder may lack social smile and may have anticipatory posture as an adult approaches. They may also have less frequent or poor eye

contact. Autistic children display impaired social development i.e. they may display poor attachment behaviour towards parents or peers. They may not be able to differentiate between the most important people in their lives like their parents, siblings, and teachers.

Anxiety experience by autistic child

Autistic children strictly adhere to their routine and when disrupted they may display extreme anxiety and irritability. When autistic children have reached school age, their withdrawal may have diminished and may be less obvious, particularly in higher-functioning children. A prominent deficit is seen in ability to play with peers and to make friends; their social behavior is awkward and may be inappropriate. Cognitively, children with autistic disorder are more skilled in visual- spatial tasks requiring skill in verbal reasoning. The cognitive style of children with autism is that they cannot infer the feelings or mental state of others around them. That is, they cannot make attributions about the motivation or intentions of others and thus, cannot develop empathy. This lack of a "theory of mind" leaves them unable to interpret the social behaviour of other s and leads to lack of social reciprocation.

Autistic person in late adolescence, often desire friendship, but their difficulties in responding to another's interests, emotions and feelings are major obstacles in developing them. They are often shunned by peers because of their awkward behaviour that alienate them form others. Autistic adolescents and adults experience sexual feelings, but their lack of social competence and skills prevent many of them from developing sexual relationships. [1]

Disturbance of Communication and Language:

Autistic children are not simply reluctant to speak and their speech abnormalities do not result from lack of motivation. Language deviance, as language delay, is characteristic of autistic disorder. In contrast to normal and mentally retarded children, autistic children have significant difficulty putting meaningful sentences together even when they have large vocabularies. In the first year of life, an autistic child's pattern of babbling may be minimal or abnormal. Some children emit noises like clicks, screeches and nonsense syllables - in a stereotyped fashion, without a seeming intent of communication. [1]

Autistic children with delays in learning to talk and use language may not compensate by using extensive gestures and pointing. Instead young children with this disorder may put an adult's hand on what they want, pull the adult over to the object they want to access, or engage in problematic behaviors to express their message. Individuals with underdeveloped speech may grunt, point a finger, pull an adult to an area, or use a picture cue and some may never develop a meaningful speech and may need to use singing and / or augmentative communication devices. Others begin their journey towards developing oral communication skills by using echolalia. Some have a very restricted repertoire of use of their communication skills and need direct instruction and support to expand their skills. Even those who develop oral skills often have other language / communication problems in social and academic situations.

Individuals with autistic disorder are very concrete in their understanding of the world and can have significant comprehension problems and significant gaps in their store of background knowledge; hence they may see jokes and sarcasm as lies and then mistrust the speakers. [1]

Stereotypes Behaviour:

Restrictive, repetitive and stereotypical behaviour may vary with individuals, circumstance, and age and by the level of awareness about others. In an autistic child's first few years of life, the expected spontaneous exploratory play is absent. The toys and objects are often manipulated in a ritualistic manner, with few symbolic features. Autistic children generally do not show imitative play or use abstract pantomime. The play and other activities of these children are often rigid, repetitive and monotonous. The ritualistic and compulsive phenomenon's are common in infants and adolescents years. Autistic children often use inanimate objects in a very vague manner like

spinning bottles, banging toys and may exhibit attachment to particular inanimate objects like wires, rods, etc. Autistic children generally show resistance to transition and change like moving to a new house, moving furniture in a room, or a change, such as having breakfast before a bath when the reverse was routine was the routine may evoke panic, fear or temper tantrums. [1]

Rocking one's body for example is a repetitive behaviour which occurs when the child is anxious, agitated at home, school and social situations. Repetitive behaviours might include lining things up, ensuring that all cupboard doors are closed, aligning chairs in a certain fashion, and making certain noises. Stereotyped movements might include hand flapping, rocking, spinning, jumping, and other patterns. Restrictive behaviour involves having a narrow set of interests. Some become walking encyclopaedias of facts on certain topics.

Instability of Mood and Affect:

It is very common for autistic children to exhibit mood changes, with bursts of laughing or crying spells for no apparent reason. It is however, a difficult to task to learn about these episodes if the child cannot express the thoughts to the effects. [1]

Response to Sensory Stimuli:

Our senses are conditioned to organize and interact with the world around us. It is important to realize, understand, and accept that some individuals on the autism spectrum may actually feel, hear, see, smell, and taste at an extreme level. They may be hypersensitive (overreactive) or hyposensitive (underreactive) for example to sound and pain.

Touch Sensation:

Touch is an important sense that is needed for social interactions with loved ones or to show care for a person. However, some with autistic disorder don't like being touch and respond aggressively on doing so, whereas on the other hand some have a high tolerance for pain and may not realize a shoulder is broken until it is swollen for several days.

Auditory Senses:

Imagine how it would feel to hear a fire alarm, vacuum cleaner, or a room full of children at a birthday party at a magnified level at the same time within the environment. This is what autistic children go though and they have difficulty in blocking and auditory senses. An individual may have difficulty discriminating between sounds, remembering directions, paying attention to a voice, and / or reading aloud.

Vestibular sensations:

Vestibular sensations occur in our inner ear. The inner ear receptors register every movement we make and every change in head position. This encompasses messages from our neck, eyes, and body. Rotary movements involve moving in circles (e.g; spinning). Linear movements (i.e; back and forth, side to side, up and down), especially when rapid (i.e; rocking in a chair, swaying, swinging on a tire), may cause most to become dizzy, nauseated, or to get a headache. Individuals with autistic disorder may actually crave these sorts of movements and this means that they need a lot of vigorous activity in order to get started. Some become distressed and show anxiety due to a fear of falling, being picked up, standing up, or sledding down a hill.

Visual Senses:

Individual with autistic disorder may be distracted with objects hanging from the ceiling; they may feel blinded by the sun or may be unable to focus in the presence of florescent lighting. Whereas others may have their eyes glued on to spinning or bouncing objects or reflection of objects.

Smell and Taste Sensation:

Some individuals with autistic disorder may be highly agitated by perfumes, the odour of foods or animal or hand lotions and may avoid people, places or foods so that they are not around that odour. Whereas, individuals hyposensitive to smell may crave odours or tastes and may lick or taste inedible objects (e.g; clay, chalk) or prefer spicy, hot, or sour foods.

Proprioceptive feedback helps us position our bodies and move through the environment. Proprioceptors exist in our muscles, joints, ligaments, tendons, and connective tissue. Proprioceptors\ work closely with tactile and vestibular systems through body awareness, motor control / planning, and postural stability. Autistic children may need to keep their eyes open in order to know how their own body is moving. Motor control / planning involves coordinating one's gross and fine motor skills within the environment. Those who have difficulty in this area may bump into people or obstacles or frequently fall or trip. Some cannot regulate how much pressure to exert when

grasping an object. As a result they hold pencils too tightly, and their written work is messy. Individuals on the autism spectrum may have poor posture, lean their head on their hands, and be unable to balance on one foot. In all of these areas, individuals may feel emotionally insecure due to feeling uncomfortable or inadequate, rigid, or intimidated by the environment. It is imperative to accommodate individuals with sensory needs throughout the day to help them calm their nervous systems so they can function in home, community, or work. These breaks should not be considered a reward but as a necessary requirement for a person's day. Many autistic children reportedly enjoy music, where they frequently hum a tune or sing a song or commercial jingle before saying words or using speech.

Associated Behavioral Symptoms:

Self Injurious Behaviour

Hyperkinesis is a common behaviour problem in young autistic children whereas hypokinesis is less frequent; but when present it often alternates with hyperactivity. Aggression and temper tantrums are observed, often prompted by change or demands. Self injurious behaviors include head banging, biting themselves or others, picking their scabs, scratching, and hair pulling. Others may react to their environment by being physically hurting their siblings, peers or family members, and such behaviors may escalate during the adolescent years due to the hormonal changes. Some children may flap their arms, brush their hands against their face repeatedly, or may hum as a self -stimulatory behaviour. Short attention, poor ability to focus on task, insomnia, feeding and eating problems and enuresis are also common among children with autism.

Head Banging behaviour

Difficulties with Executive Function:

Individuals with autism have difficulty in multi tasking, completing assignments and organizing. They are better at carrying out one task at a time. Executive functions also include deficits in the area of making transitions and organizing oneself.

Intellectual Functioning:

Approximately 70 to 75 percent of children with autistic disorder fall in the mentally retarded range of intellectual function. About 30 percent of children function in the mild to moderate range, and about 45 to 50 percent are severely to profoundly mentally retarded. Epidemiological and clinical studies show that the risk of autistic disorder increases as the IQ decreases. About 1 / 5 of all autistic children is have a normal, non-verbal intelligence. The Intelligence Quotient scores of autistic children tend to reflect most served problems with verbal sequencing and extraction still, with relative strength in visuo-spatial or rote memory skill. This finding suggests the importance of the effects in language related functions.

Unusual order precocious cognitive or visuo-motor abilities occur in some autistic children. The abilities, which may exist even in the overall retarded functioning, are referred to as splinter functions or islets of precocity. Perhaps, the most striking example is autistic savants, who have exceptional rote memories or calculating abilities, usually beyond the capabilities of their normal peers. Other gifted abilities in young autistic children include hyperlexia, an early ability to read well (although they cannot understand what they read), memorizing and reciting and musical abilities (singing or playing tunes or recognizing musical pieces).

When an individual is gifted in specific areas, he or she is often referred to as being savant. These individuals may have exceptional talents in calculating numbers, playing a musical instrument, or drawing.

Early Diagnosis:

Although autism is usually seen to occur during the early gestational abnormalities of brain development, but for the parent it may seem to begin after the first year of life. The babies are often "too good", very quiet and are passive or are too irritable and intense; the eye contact may be absent from the start and the infants may seem to be unresponsive and different as compared to the other children. The symptoms of autism in a two year old are very different as compared to a four year old; hence it is extremely important to make the diagnosis depending on the age.

Speech:

Delayed speech and language milestones must be taken very seriously and form the most common presenting symptoms in the autistic spectrum. Hearing impairment should definitely be ruled out with the help of tests. It is often seen that autistic children usually lack "communicative intent" and do not seem to "listen" to adult conversations. Absolute indication of preliminary assessment include absent babbling at 12 months of age, no single word at 12 months, no phrase at 24 months and at eventually may either lead to developing a restricted speech or absence of speech.

Social Interaction:

Poor eye contact and delay in social smiling within the first few months of life may require urgent consideration. The one year old who ignores gestures, does not shake or nod the head appropriately during conversations should raise suspicion. Pointing is a fundamental communicative act which is usually seen when a child is around 15 months of age but if there is a failure to point to objects at 18 months of age; this would be a strong suspicion of autism. However, absence of pointing may be seen in children who are mentally retarded or with visual impairment. They may also lack interest in imaginative play or social interaction and if play is present it could be present in the form of repetitive behaviour, rejection of new toys, or bizarre use of toys.

The critical step in early diagnosis is to recognize the grounds for suspicion and to arrange for an assessment process to begin. Some children may be seen to be mildly affected with this disorder but eventually may lead to full blown syndrome. Early diagnosis may help to formulate an intervention plan which would significantly help improving the quality of life and functioning of the child.

Differential Diagnosis:

Autism must be differentiated from one of the other pervasive developmental disorder such as Asperger's disorder and pervasive developmental disorder not otherwise specified. Further it must be differentiated from other developmental disorders, including mental retardation syndromes and developmental language disorders. It is sometimes difficult to make the diagnosis of autism because of over- lapping symptoms specified below:

Schizophrenia with Childhood Onset:

Although a wealth of literature on autistic disorder is available, few data exists on children under age 12 who meet the diagnostic criteria for schizophrenia.

Mental Retardation with Behavioural Symptoms:

Approximately 40 percent of autistic children are moderately, severely or profoundly retarded and retarded children may have behavioural symptoms that include autistic features. When both disorders are comorbid, both should be diagnosed. The prominent distinguishing feature between autistic children and mentally retarded children are that mentally retarded children usually relate to adults and other children in accordance with their mental age, use the language they do have to communicate with others, and exhibit a relatively even profile of impairments without splinter functions.

Mixed Receptive - Expressive Language Disorder:

Some children with mixed receptive - expressive language disorder have mild autistic like features and may present a diagnostic problem.

Acquired Aphasia with Convulsion:

Acquired aphasia with convulsion is a rare condition that is sometimes difficult to differentiate from autistic disorder and childhood disintegrative disorder. Children with this condition are normal for several years before losing their receptive and

their expressive language over a period of weeks or months. A profound language comprehension disorder then follows, characterized by deviant speech pattern and impairment. Some children recover, but with considerable residual language impairment.

Congenital Deafness or Severe Hearing Impairment:

Autistic children are often mute or show selective disinterest in spoken language, they are often thought to be deaf. The prominent distinguishing factors are as follows:

- Autistic infants may babble once in a while, whereas deaf infants have a history of relatively normal babbling that then gradually tapers off and may stop at 6 months to 1 year of age.
- Deaf children respond only to loud sounds, whereas autistic children may ignore the loud or normal sounds and respond to soft or low sounds.
- Most importantly audiogram or auditory evoked potentials indicate significant hearing loss in deaf children.
- Unlike autistic children, deaf children usually relate to their parents, seek affection and enjoy being held as infants.

Psychosocial Deprivation:

Severe disturbance in the physical and emotional environment (e.g. maternal deprivation, psychosocial dwarfism, hospitalism, and failure to thrive) can cause children to appear apathetic, withdrawn and alienated. Language and motor skills can be delayed. Children with these signs almost always improve rapidly when placed in a favorable and enriched psychosocial environment, but such improvement is not the case with autistic children.

Course and Prognosis:

Autism is generally a lifelong disorder with the guarded prognosis. Autistic children who have intelligence quotient about 70 and those who use communicative language by ages five to seven years tend to have the best to prognosis. It is seen that symptoms such as repetitive and ritualistic behaviour did not seem to improve overtime. Studies on the outcome of children with autistic disorder reported that about two third of autistic adult's remains severely handicapped and live in complete dependence or semi-dependency with their caregivers or relatives. Only about 2 percent are able to lead a normal, independent life with employment and about 5 to 20 percent of them are able to achieve a borderline status.

The prognosis of children is believed to improve if the environment or home is supportive and capable of meeting the extensive need of such a child. The symptoms may decrease in some cases as the age increase but with others severe self-mutilation and aggressiveness and aggression have been seen to increase. About 4 to 32 per cent at a grand mal seizures in late childhood or adolescence, and seizures adversely affect the prognosis. [1]

Treatment for Autism:

Intervention for autism is a very intensive, comprehensive and one which involves the child's entire family and a qualified team of professionals. Based on the issues that the child with autism has and a comprehensive evaluation, the therapies and therapists are narrowed down for the child. Treatment programs may combine therapies for both core symptoms and associated symptoms.

Medical Management:

Medical management makes an important contribution to the overall wellbeing and outcome in individuals with autism and to their family members, even though the primary intervention is psychological and educational in nature. The physician is usually the only resource for the families in search of up- to -date and accurate information on the biology and neuroscience of autism.

Bowel functions (diarrhoea and constipation):

This occurs with the same frequency in children with autism and other neurological disorders such as cerebral palsy. Constipation usually reflects low muscle tone and immobility and deserves very energetic treatment along conventional lines in order to prevent fissures and pain. Family history of severe constipation should lead to the exclusion of Hirschsprungs disease. Good hydration is essential in the management of both constipation and diarrhoea.

Seizures:

There is substantial evidence that certain nutritional supplements, especially vitamin B6 and

dimethylglycine (DMG), can provide a safer and more effective alternative to drugs, for many individuals.

Sleep:

Sleep is a frequent concern for parents of autistic children and can be very difficult to treat. A strict going to sleep routine is even more important for these children than for others, and here their strong need for routine and ritual may be used to help them. Stimulating foods and drinks should be avoided. With the exception of melatonin no drugs are recommended for insomnia in children, unless a simple antihistamine such as Benadryl is given for a few nights to give the parents a break from incessant crying or hyperactivity. Some truly hyperactive children diagnosed with autism and ADHD may be helped to sleep by methylphenidate (Ritalin) but most will be kept awake even longer. Placing bricks under the head of the bed may help keep stomach acid from rising and provide better sleep. Melatonin has been very useful in helping many autistic individuals fall asleep. Other popular interventions include using 5-HTP and implementing a behaviour modification program designed to induce sleep. Vigorous exercise will help a child sleep, and other sleep aids are a weighted blanket or tight fitting mummy-type sleeping bag.

Dental Treatment:

As some autistic children are non- verbal they may communicate the pain of dental cavities or issues by being self-abusive, hyperactive or otherwise by a very difficult behaviour.

Nutrition and Diet:

It is often observed that autistic children are very fussy feeders who incorporate their obsessions and need for the sameness into their feeding behaviour. Regular measurement of weight, height and head circumference will allow calorie deficiencies to be detected early along with clinical evaluation for signs of malnutrition. As sub- optimal nutrition with micro-deficiencies in many vitamins and mineral supplements should be recommended for all but those whose meal -time behaviour approaches normal. Zinc deficiency may be associated with sensory blunting, poor appetite, and disturbed bowel functions and perhaps with aggressive behaviour so the supplementation should often be considered. Autistic children may have low plasma fatty acids and supplementation with evening primrose oil or fish oil can do no harm. Non - specific or placebo like benefits seem to occur with almost any dietary improvement or modification.

Gluten Free, Casein Free Diet (GFCF): A very popular dietary treatment for symptoms of autism is removal of gluten (a protein found in barley, rye, oats, and wheat) and casein (a protein found in dairy products), in what is known as a Gluten Free, Casein Free diet, or GFCF. This is based on the hypothesis that these proteins are absorbed differently in children with autism spectrum disorders and act like false opiate-like chemicals in the brain. There are ongoing studies to prove the effectiveness of this dietary intervention. However, many families report that dietary elimination of gluten and casein has helped to control bowel habits, sleep activity, habitual behaviour and enhance the overall progress in their children.

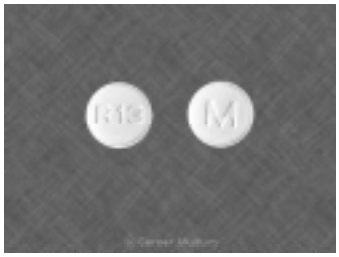
Pharmacotherapy in Autism:

Current psychopharmacologic trials are under way to investigate efficacy of a variety of classes of agents on promoting social interactions and reducing disruptive behaviours in children and adolescents with autism and other pervasive development disorders. Currently, no specific medications with proved efficacy in the treatment of the core symptoms of autistic disorders are available; however medications have been shown to be promising in reducing hyperactivity, obsessions and compulsive behaviours, irritability, aggression and self - injurious behaviours.

The administration of antipsychotic medication has been shown to be efficacious in the reduction of aggressive and self- injurious behaviour. One early study indicated that haloperidol reduced the behavioural symptoms such as hyperactivity, stereotypes, withdrawal, fidgetiness, irritability and labile affect and accelerated learning. Given its potentially serious adverse effects, haloperidol is no longer the antipsychotic agent of choice in the treatment of self injurious behaviours in children with autistic disorders.

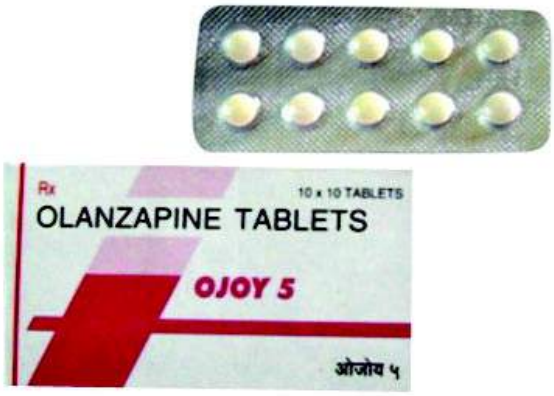
The atypical antipsychotic agents are known to have a lower risk of causing extra pyramidal adverse effects, although some sensitive individuals cannot tolerate the extra pyramidal or anticholinergic adverse effects of the atypical antipsychotic agents. The atypical antipsychotic agents include risperidone, olanzapine, quetiapine, clozapine and ziprasidone.

Risperidone, a high-potency antipsychotic with combined dopamine D2 and serotonin 5 - HT2 receptor antagonist properties, has been used to subdue aggressive or self - injurious behaviours. Several reports have suggested that risperidone is effective in diminishing aggressiveness, hyperactivity and self-injurious behaviour in children with autistic disorder. In some cases, it reportedly encouraged socially acceptable behaviours. The U.S. Food and Drug Administration has approved Risperdal as autism medication to treat irritability in autistic children and adolescents. This is the first time the FDA approved a drug to treat behavior-related problems associated with autism in children. The drug can be used to treat aggression, deliberate self-injury and temper tantrums. Risperdal is considered an atypical antipsychotic drug manufactured by Janssen Pharmaceutica N.V. in Beerse, Belgium. For autism, lower dosages ranging from 0.5 to 4 mg per day are used in clinical practice. Extra pyramidal effects and akathisia have been reported adverse effects, as well as sedation, dizziness and weight gain. Drooling was reported more in the risperidone group compared with the placebo group. In this sample, extrapyramidal symptoms were not reported more commonly in the risperidone group. The side effects that caused the most concern were somnolence and weight gain. [10]



Risperidone 3 mg MYL,
white, round, film coated

Olanzapine specifically blocks 5- HT 2A and D2 receptors and also blocks muscarinic receptors. No studies provide specific guidelines regarding the use of olanzapine in children with autism. Dosages that have been used clinically to target aggression and self-injurious behaviours range from 2.5 to about 10 mg per day. Among olanzapine's most common adverse effects are sedation, orthostatic hypotension, and weight gain.



Olanzapine

Clozapine has a hetrocyclic chemical structure that is related to certain conventional antipsychotics, such as loxapine, although clozapine carried lower risk of extrapyramidal symptoms. It is generally used in treatment of aggression and self- injurious behaviour unless those behaviours coexist with psychotic symptoms. It's most serious adverse effects are agranulocytosis, which necessitates monitoring white blood cells count weekly during clozapine's use. Its use is generally limited to treatment - resistant psychotic patients. Lithium can be administered in the treatment of aggressive or self - injurious behaviours when anti psychotic medications fail.

Clozapine C11 M, 100,g, green colour,
9.00 mm, round.

Psychotropic Medication: Psychotropic medication is commonly used to treat disruptive behaviors, agitation, inattention, and hyper-activity in children with ASD (Myers, et al; 2007).

Class	Medication (Brand name)	Level of Evidence	Target Symptoms	Significant Potential Side Effects	Studies
Antipsych -otics	Risperidone (Risperidal)	**Established Evidence**	Irritability, hyper-activity, and stereotypy	Weight gain, drooling, dizziness, fatigue, involuntary muscle movement	(Jesner, Aref-Adib, & Coren, 2007; McDougle, et al; 2005; Miral, et al; 2008; RUPP, 2002)
	Haloperidol (Haldol)	**Established Evidence**	Aggression	Tardive dyskinesia, sedation, irritability	(Anderson, et al; 1989; Anderson, et al; 1984)
Stimulants	Methylpheni-date (Ri-talin)	**Established Evidence**	Hyperactivity	Social withdrawal, irritability, agitation, stereotypy	(Handen, Johnson, & Lubetsky, 2000; Quitana, et al; 1995)
Norepineph-rine Reuptake Inhibitor	Atomoxetine HCI (Strattera)	**Preliminary Evidence**	Attention deficit, hyperactivity	None	(Arnold, et al., 2006)
Alpha 2 Agonist	Clonidine (Catapres)	**Preliminary Evidence**	Hyperactivity, irritability, inappropriate speech, stereotypy, oppositionality	Drowsiness, low blood pressure, irritability	(Jaselskis, Cook, Fletcher, & Leventhal, 1992)
	Guanfacine (Tenex)	**Insufficient Evidence**	Hyperactivity, inattention, impulsivity, aggression	Transient sedation	(Posey, Puntney, Sasher, Kem, & McDougle, 2004)
Selective Serotonin Reuptake Inhibitors (SSRIs)	Fluoxetine (Prozac) & Citalopram (Celexa)	**Insufficient Evidence** (conflicting results)	Repetitive behavior	Celexa: Hyperactivity, insomnia, inattention, impulsivity, diarrhea, dry skin	(Hollander, et al., 2005; King, et al., 2009)
Others	Clomipramine (Anafranil)	**Preliminary Evidence**	Stereotypy, ritualistic behavior, social behavior	Insomnia, constipation, twitching, tremors	(Gordon, State, Nelson, Hamburger, & Rapoport, 1993)
	Valproic Acid (Depakote)	**Insufficient evidence**	N / A	Rash, weight gain, hair loss, fatigue	(Heillings, et al., 2005; Hollander, et al; 2006)
	Naltrexone (Revia)	Insufficient evidence	N / A	Increased stereotypy	(Willemsen-Swinkels, Buitelaar, Weijnen & van Engeland, 1995)

Role of Psychology in Autism:

Diagnostic and Assessment Instruments For Children with Autistic Spectrum Disorder:

The below listed scales and tool are used by psychologists, educators and researchers to assess children suspected of or previously diagnosed with a pervasive developmental disorder. The instruments listed below are used to measure specific dimensions of a child's development in different domains of functioning, which would help develop a proper intervention plan for the child.

Diagnostic Assessment:

Autism Diagnostic Interview - Revised: This is a semi-structured, investigator-based interview which is useful for diagnosing autism, having an intervention plan and distinguishing autism from other developmental disorders. The ADI-R has proven particularly effective in assessing syndrome boundaries, identifying new subgroups, quantifying Autistic symptomatology, and determining the clinical needs of groups in which a high rate of Autism Spectrum Disorders might be expected, such as people with severe language impairments. It is conducted on children and adults with a mental age of above 2 years. It takes approximately 1 1/2 to 2 1/2 hours, including scoring.

Prelinguistic Autism Diagnostic Observation Schedule: The Prelinguistic Autism Diagnostic Observation Schedule (PL-ADOS) (DiLavore, Lord, & Rutter, 1995) is a semi-structured observation scale for diagnosing children whose speech is not yet developed and who are suspected of having autism. This instrument provides an opportunity to observe specific aspects of the child's social behavior, such as joint attention, imitation, and sharing of affect with the examiner and parent. PL-ADOS scores are reported to discriminate between children with autism and children without autistic developmental disabilities. [11]

Childhood Autism Rating Scale Second Edition (CARS- 2): This scale helps to distinguish children from developmentally handicapped children who are not autistic. It helps distinguish between mid to moderate to severe autism. Standard Version Rating Booklet (CARS2-ST) is equivalent to the original CARS; for use with individuals younger than 6 years of age and those with communication difficulties or below-average estimated IQs. High-Functioning Version Rating Booklet (CARS2-HF) is an alternative for assessing verbally fluent individuals, 6 years of age and older, with IQ scores above 80. Questionnaire for Parents or Caregivers (CARS2-QPC) is an unscored scale that gathers information for use in making CARS2-ST and CARS2-HF ratings. The CARS2-ST and CARS2-HF each include 15 items addressing the following functional areas like: relating to people, social and emotional understanding, imitation, body use, visual responses, verbal communication, taste, smell, and touch response and use, etc. It is conducted on children 2 years and older and the approximate administration time is 5 to 10 minutes. [12]

Checklist for Autism in Toddlers (CHAT): This is a screening instrument designed to detect core autistic features to enable treatment as early as 18 months. CHAT offers psychologists a means of diagnosing autism in infancy so that educational programs can be started months or even years before most symptoms become obvious. It is a test which aims to look at behaviors which, if absent at 18 months, put a child at risk for a social-communication disorder. These behaviors are (a) joint attention, including pointing to show and gaze-monitoring (e.g. looking to where a parent is pointing), and (b) pretend play (e.g. pretending to pour tea from a toy teapot).

Gilliam Autism Rating Scale - Second Edition (GARS - 2): GARS-2, a revision of the popular Gilliam Autism Rating Scale, assists teachers, parents, and clinicians in identifying and diagnosing autism in individuals ages 3 through 22. It also helps estimate the severity of the child's disorder. Items on the GARS-2 are based on the definitions of autism adopted by the Autism Society of America and the Diagnostic and Statistical Manual of Mental Disorders: Fourth Edition-Text Revision (DSM-IV-TR). The instrument consists of 42 clearly stated items describing the characteristic behaviors of persons with autism. The items are grouped into three subscales: Stereotyped Behaviors, Communication and Social Interaction. Using objective, frequency-based ratings, the entire scale can be completed and scored in 5 to 10 minutes. A structured interview form is included for gathering diagnostically important information from the child's parents. [13]

Developmental Assessment:

Psycho- educational Profile-Revised (PEP-R): This offers a developmental approach to the assessment of children with autism or related developmental

disorders. It is an inventory of behaviors and skills designed to identify uneven and idiosyncratic learning patterns. The test is most appropriately used with children functioning at or below the preschool range and within the chronological age range of six months to seven years. The PEP-R provides information on developmental functioning in imitation, perception, fine motor, gross motor, eye-hand integration, cognitive performance, and cognitive verbal areas. The PEP-R also identifies degrees of behavioral abnormality in relating and affect (cooperation and human interest), play and interest in materials, sensory responses, and language.

Adaptive Assessment:

Vineland Adaptive Behavior Scales - Second Edition (Vineland-II): This scale comes in three forms varying in degree of detail and proposed setting. The VABS is administered by interviewing the child's parents, teachers, or care providers. The scales range in age from birth to nineteen years. It takes approximately 20 to 60 minutes for the conduction and scoring of the test. [14]

Standardized Tests of Intelligence:

Wechsler Intelligence Scale for Children (3rd ed.): This is used to assess intelligence as a global but multifaceted entity that can be inferred from a child's performance on a series of tasks. It is valuable for psycho-educational assessment, diagnosis, placement, and planning. WISC-III can be used to diagnose exceptionality among school-aged children and has a strong place in clinical and neuropsychological assessment and in research. Like the WPPSI-R, the WISC-III is widely used and generally regarded as the best standardized measure of intelligence. [15]

Differential Ability Scales: The Differential Ability Scales (DAS) (Elliott, 1990) [16] measures overall cognitive ability and specific abilities in children and adolescents. It is better suited for intellectually higher-functioning children with autism. The DAS assesses multidimensional abilities in children ages two years and six months to seventeen years and eleven months. It is administered individually and takes 45 to 65 minutes for the full cognitive battery. The achievement test takes 15 to 25 minutes to administer. The seventeen cognitive and three achievement subtests yield an overall cognitive ability score and achievement scores. The three achievement subtests are Basic Number Skills, Spelling, and Word Reading.

Stanford-Binet Intelligence Scale (4th ed.): The Stanford-Binet Intelligence Scale (4th ed.) (SBIS-IV) (Thorndike, Hagen, & Sattler, 1986) is for individual's ages two years to adult. It provides scores in four areas: Verbal Reasoning, Abstract and Visual Reasoning, Quantitative Reasoning, and Short-Term Memory; and a Composite Score that is equivalent to the Wechsler Scales Full Scale IQ. [17]

Academic Screening:

Wide Range Achievement Test 3 (WRAT3): The Wide Range Achievement Test 3 (WRAT3) measures reading, spelling, and arithmetic in persons from five to seventy-four years old. Two equivalent forms make pre- and post testing possible. The test takes 10 to 15 minutes to administer. The WRAT3 provides a good method for measuring basic academic skills in children who perform below their peers.

Behavior Assessment:

Achenbach Child Behavior Checklist: The Achenbach Child Behavior Checklist (ACBC) is for children four to eighteen years old and is completed by an adult informant. It has two major scales - externalizing and internalizing behaviors - each of which has four subscales. It has been used as a follow-up measure. The child's primary caregiver (in most cases, the client's mother) serves as the informant.

Analysis of Sensory Behavior Inventory (Rev. ed.): The Analysis of Sensory Behavior Inventory (Rev. ed.) (ASBI-R) (Morton & Wolford, 1994) is designed to collect information about an individual's behaviors as they are related to sensory stimuli. Six sensory modalities are assessed: vestibular, tactile, proprioceptive, auditory, visual, and gustatory-olfactory. Ratings can be made about both sensory-avoidance and sensory-seeking behaviors within each modality. Information obtained from this tool may be helpful in completing a functional analysis of behavior and in designing effective intervention strategies, including accommodations and reinforces for the individual.

Psychological Intervention for Autism:

The goals of psychological intervention for children with autism is to eliminate or reduce the undesirable behaviours and target the behaviours that will help the child integrate into schools, develop and meaningful social communication with peers and family members and increase the

likelihood of maintaining an independent living as an adult. To do this, treatment intervention aim to increase socially acceptable and pro-socially behaviour, to decrease or behavioural symptoms, and to improve verbal and non-verbal communication. It basically aims at increasing the level of independence and increasing the quality of life. Both language and academic remediation often required. In addition, treatment goal generally include a reduction of disruptive behaviour that may be exacerbated especially during translation and in school. Children with mental retardation need intellectual appropriate behaviour intervention to be reinforcing socially acceptable behaviour and encourage self-care skills. In addition, parents, often distraught, need support and counseling. Insight oriented individual psychotherapy has proved ineffective. Educational and behavioural interventions are currently considered the treatment of choice. Structured classroom training, in combination with behavioural method, is the most effective treatment for many autistic children.

Well controlled studies indicate that the gain in the area of language and cognition and diseases in the maladaptive behaviour are achieved by consistent behavioural programs. Careful training of parents in the concept and skills of behaviour modification and resolution of the parents' concerns me yield considerable gain in children's language, and cognitive and social area of behaviour. These training programs, however, are rigorous and require much parental time. And autistic child requires as much structure as possible, and the daily program for as many hours as feasible is desirable.

Behaviour Therapy:

Behaviour Therapy Plan:

Identification: The behaviours which are undesirable and which need to be either replaced with a desirable behavior or reduced needs to be identified and noted down including the level of severity in it.

Gathering Information: Information need to be gathered from all the family members or caregivers, therapists and teachers who are taking care of the child, teaching or treating the child to understand the behaviours, the pattern of the behavior and the situations where and when the undesirable behaviours occur.

Observation: The child's behaviour needs to be observed in different situation as this helps to understand the response of the child in various situations and the level of acceptability of the behavioural plan.

Making a Behavioural Plan and Implementing: A behavioral plan needs to be formulated, where also the strategies that are to be used are to be listed down and it should be implemented. Along with it a close track needs to be kept on whether, the plan benefiting the client or is it further accelerating the client's undesirable behaviours. If it is accelerating the client's undesirable behaviors then, the techniques should be replaced with one which will help reducing the undesirable behaviors.

Reward System: A reward system needs to be set up, depending on what the child considers as a reward and something which in the long run would not lead to another issue. Examples of rewards which are safe and encouraging are praise, star on the hand, food items, etc.

BEHAVIOUR MANAGEMENT PLAN FOR AUTISM

Name of the patient: Date:

Age: Session Number:

Define the behaviour problem, severity and obstacles caused by it:

Hypothesis regarding the behaviour problem:

Identified skills to be taught to reduce the behavioral problems:

Antecedent strategies used to put in place to prevent the behaviours:

Hierarchy of consequential strategies to use when behaviours occur:

Reward system for good behavior:

Traditional Approaches to Deal with Poor Behaviour:

Aversives: Aversives are defined as the use of negative reinforcers to interrupt or stop problem behaviours as and when they occur. Aversives are response to inappropriate behaviours and not a preventive measure for inappropriate behaviours. For example: When a child is being very hyperactive and is disturbing the class the teacher would direct the child to stand outside the class for 10 minutes.

Overcorrection: This a type of aversive technique which requires the child to complete the

appropriate behaviour several times because he did not do it when asked, or because he continued with the negative behaviour in spite of being asked to stop. For example:

Restraints: Restraints are defined as techniques that are used to "hold back" a child from doing some act which is harmful or dangerous to him. For example: If an autistic child has a self- injurious behaviour of banging his head hard on the table or the ground, then this behaviour can be prevented by having a custom made helmet for the child. This would either stop the child from banging his head or at least prevent from inflicting injury to himself.

Behavior Modification:

Behaviour Modification is the use of a collection of techniques that are compiled together to increase the number of desired behaviours and to extinguish or reduce the number of undesirable behaviours. Behaviour modification takes account of only those behaviours which are observable and measureable. The way to change an undesirable behaviour is to "target the behaviour" and then break them down into tiny pieces and steps. After which the tiny pieces of behaviors need to be practiced and rewarded till it is not mastered. Punishments like withdrawal of privileges or timeouts are used to reduce the number of undesirable behaviours. It is again important to remember that behaviour modification is a commonly used technique for handling behavioural problems and that it does not take into consideration the reasons for such behaviours. For example if an autistic child is exhibiting self-stimulatory behaviour because he is stressed, then the application of overcorrection may lead to replacing one self-stimulatory behaviour to another.

Behaviour Therapy

Behaviour Strategies: Social Language Difficulty:

Autistic children have social language inadequacy which due to which they may not be able to initiate a conversation or sustain it and it may lead to not having peers or peer group. Social skills deficits must be addressed in individualized educational plans, as they are the core symptoms of the autistic spectrum disorder. [18]

The following are few listed strategies to improve social language:

1. Individualized training of facial expression and behaving in a socially acceptable manner would help grasp and understand better. Gradually then exposure to practicing this in a group would be really helpful.
2. Usage of drama and role playing to teach the voice inflection, modulation and facial expression is seen to be very helpful. It helps to reduce the fear of speaking in public and helps boosting the child's confidence.
3. Use videotapes and flashcards to imitate and practice facial expression. This would also help to improve eye contact and help increase attention and concentration. For example: Expose the child to various expressions on a flash card and ask him to imitate the expression.
4. Peers and siblings can be trained to promote help the client maintain a naturalistic kid friendly conversation.
5. Teach the child how to initiate a conversation, respond and how to sustain it.
6. Also, a very important aspect that is poor in autistic children is maintaining eye contact. Help the child understand the importance and practice this by reminding the child to maintain eye contact while role-playing.

Behaviour Strategies: Social Behaviour Difficulty:

Autistic children have difficulty in displaying socially appropriate behaviours due to which they are usually bullied or may face rejection for people around or they themselves prefer to stay in isolation. [19]

Group Social Interaction

The following are few listed strategies to improve social behaviour:

1. Arrange the situation for the child in such a way where, he needs to find clues to ascertain how someone is feeling or predict how they will respond. He or she needs to be taught what various facial expression, emotions and how one's sounds in different emotions.
2. Use videotapes or television to reinforce learning about facial expressions and sounds of emotions.
3. Use role play, group therapy and practice scripts to respond to moments of teasing or being bullied.
4. Teach the child, the social rules and regulations which would help them behave appropriately to a situation.
5. Use stories with a moral and explain to the client the story step by step which would help him / her understand the behaviours that are acceptable socially.

For example: An autistic patient would have trouble invading someone's personal space, by positioning themselves very close to the individual they are listening to, which would place the speaker to be very uncomfortable with the close proximity. To avoid this vocal reminders need to be given and in spite of this if there are no changes then, a strategy to solve this would be to tie a rope around the child's waist, with leaving a 3 foot tail behind. Practice this with peers holding the tail and demonstrating to the client the meaning of 3 feet difference.

Behaviour Strategies: Difficulties with Transitions and Changes:

Autistic children yearn for sameness and routine. If there is a change in their routine, then they would try their best to stick to their routine or would throw temper tantrums. It's seen that children with autism work better and learn better with people who are more structured in their daily routine.

The following are few listed strategies to help autistic children to accept and adjust to transitions:

1. Teach children to adjust and accept transitions by making small changes in their routine as and when they get very comfortable with it.
2. Dim the lights of the room, when a new activity is being presented to them.
3. Give prior explanation of the transition as getting them prepared before hand and understanding why a particular change is taking place would help the be prepared rather than having a flaring temper after the change has occurred.
4. Make the transition as enjoyable and as stress free as possible because otherwise the child's autistic feature will shoot up.

Behaviour Strategies: Problems with Self-Esteem and/ or Depression:

Often children with autism have a poor self-concept and lack confidence. This is normally seen to be present by the child having a poor eye contact, difficulty socializing or stammering. This is a very common feature in autism as these children undergo a lot of rejection from peers, they hear people around speaking about their behaviour and problems and being bullied.

The following are few listed strategies to help boost self -esteem in autistic children:

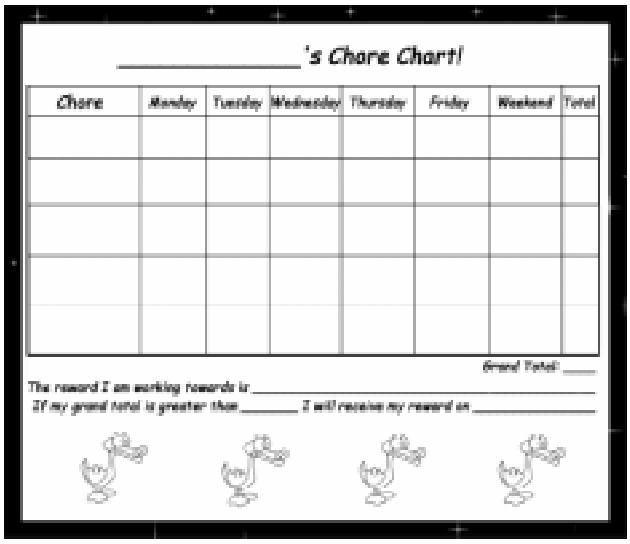
1. Keep a track of the child's behavioural achievements and encourage it by rewarding the child in front of the family members.
2. If the child is demonstrating poor or undesirable behaviours then do not take away his previously earned rewards.
3. Reward him / her for minor changes in the behaviour.
4. Be specific about the behaviours that are liked and encourage them with a reward. Behaviours that are undesirable need to be changed by desirable ones.

5. Phrases and words like "good boy" or "bad boy" should not be used rather "You followed the direction well" should be used to help increase the confidence.
6. The child's strengths should be motivated and ways to enhance it should be used. For example: A child who enjoys music should be encouraged to learn an instrument and excelling at playing it.
7. Encourage the client to maintain a photo book of his achievement or good behaviours. For example: If he / she won some competition or made some clay model, take a picture and keep it in front of the child.
8. When an autistic child is using negative words, phrases or sentences referring to himself encourage the child to make changes in his / her dialogues to a more positive view about himself / herself.
9. The child's grades, health concerns and short comings should be kept confidential and not publicized about.

Behaviour Strategies: Managing Stress and Anxiety:

Helping children with autism managing stress and anxiety is extremely important in preventing undesirable behaviours.

1. Maintain a time- table for all the daily activities and keep a reward system for it.
2. Teach the child breathing exercises, help him understand when to use it and practice it, so that the child is aware that he has to use it when he is stressed and anxious.
3. Exercise and play session is extremely important for the child, as it would help the client relieve his / her stress.
4. Self- talk method should be taught to the client and he should be also taught that this should be only practiced when he is alone in the room.
5. The client should have some hobby which he or she enjoys and feels relaxed when taking it up.



Cores Chart for managing stress

Behaviour Strategies: Attention Problems:

Attention problems can lead to poor behavior. It is often assumed that a child is non-compliant when he or she does not pay attention to directions or is non-focused when a task is being performed.

1. The desk on which the child sits to study should be dark in colour and should have very few items on the desk. It is seen that dark colour desks are said to be able to maintain attention for a longer time.
2. A separate room should be allotted to the child, to sit and study. The room should have less furniture and the room should not be of a very bright colour. While the child is studying the windows should be closed as most of the autistic children love to stare outside the window.
3. Demonstrate to the client how one's boy should look when one is paying attention i.e. modeling technique should be used.
4. The child's interests should be known in term of activities with high interest and low interests. The low interest activities should be separated with breaks.
5. Arrange for tests and difficult assignments to be completed in quiet rooms away from distractions.

Cognitive Rehabilitation to increase attention span and level of comprehension.

Strategies: Improve Academic Skills

Autistic children have academic problems especially if they fall into the category of mental retardation or if they have learning difficulties.[20]

1. Modify the homework and class work assignments for the child.
2. Use different modalities to teach in a way which will help the child grasp and understand better like audio and visual modality i.e playing nursery rhymes with video.
3. Create motivators and rewards that are clearly established and known to the child.
4. Have individualized and personalized session with the client to make the child understand concepts in a better way.
5. If the child is comfortable with a certain special educator or a teacher see to it that he/ she is there with the client throughout the academic year. Changing teachers or educators would confuse and upset the child as autistic children if they get attached to a particular educator they can be very high chances of improvement in them.

Rewards and Motivators:

A very crucial aspect of any behavior modification / management plan is to have reinforcements listed down which would encourage a child to have a desirable behaviour and eliminate or reduce undesirable behavior. Punishment teaches children with autism that they have done something wrong and it does not teach them the right behaviour but instead has a bad impact on their self-esteem and confidence.

Reward

Rewards can include:

Edible reinforcers: fruits, biscuits, chocolates, etc.

Material reinforcers: toys, stickers, star on the hand, etc.

Social reinforcers: smiling, patting, praising, etc.

Activity reinforcers: play time, or T.V. time, etc.

Reward Systems:

Token Economy: When a child completes a task or activity assigned to him successful he is awarded with a token for example a poker chip, which he can exchange for various things like toys or T.V time.

Points earned versus points lost: This is where the child earns points on daily basis i.e. if a child's behaviour is good then he receives a point but if his behaviour is unacceptable then a point is subtracted. The list of acceptable and unacceptable behaviours should be specified to the child.

Sticker Card: The child here is presented with a sticker card consisting of 20 grid rectangle. If the child demonstrates good behaviour then he receives a sticker and after the completion of card he can trade it for a reward.

Positive notes: A positive word communicated to an autistic child is very encouraging but autistic children are visual learners and if the same words are put on papers then it would help boost the child's confidence.

Above mentioned are a few reward systems for autistic children, it is very important to understand that a meaningful reward would encourage a child to have a good behavior and maintain it. Initially the child may be very excited about the reward but eventually may lose out interest in the reward, at this time the rewards should be revised. It is important that the child understands the reward system.

Consequences:

Like rewards are an important factor in contributing to the child's good behaviour, in the same way consequence is equally important for the child to understand that if not behaved in an undesirable manner then he will be punished.

Types of Consequences:

- Vocal chastise (no hitting).
- Loss of pleasure activities (loss of T.V time).
- Verbal threats or warnings (If you do that again, you will be made to sit in the timeout zone).
- Time-out (the child should be made to sit in the corner of the room, without making any noise for a decided time limit and should think about his behaviour and why he was punished? If he argues then his time limit would increase).

APPLIED BEHAVIOUR ANALYSIS:

Applied Behaviour Analysis (ABA) is based on the theory that influencing a response associated with a particular behaviour may cause that behaviour to be shaped and controlled. The behavioural methods in ABA are used to measure behaviour, teach functional skills, and evaluate progress. There are various studies which suggest the success of ABA technique in treating deficits in the behaviours of children with Autism Spectrum Disorder (ASD) at any level.

Approaches of Applied Behaviour Analysis:

1. Discrete trial training (DTT)
2. Pivotal Response Training (PRT)
3. Picture Exchange Communication System (PECS)
4. Self-Management
5. Social skills training techniques

The goal of the session is to find ways to motivate the child with the help of a number of strategies and to ensure that the session is enjoyable and productive. The purpose of the ABA program is to improve the language skills, play, and socialization and to eliminate or reduce self-injurious or ritualistic behaviours that interfere with the learning process. It also focuses on to improve the eye contact and encourage on the desire to learn. Even if the child does not achieve a "best outcome" result of normal functioning levels in all areas, nearly all autistic children benefit from intensive ABA programs.

Specific targets of the interventions are chosen based on the child's individual issues and disorder. In an intensive behavioural intervention program, goals are set and progress is continuously monitored and evaluated. If there are any positive changes in the child's behaviour then the goals are eventually changed, and the focus is shifted on to improving other disruptive behaviours.

Basic Principles of Behavioural and Educational Intervention Approaches:

Behavioural therapies incorporate specific approaches to help the individual acquire or modify behaviours. The basic theory that ABA is based on is operant conditioning which involves presenting a stimulus (request) to a child, and then providing a consequence (a "reinforcer" or a "punisher") based on the child's response.

Reinforcement: A reinforcer is anything, which when presented as a consequence of a response, increases the probability or frequency of that response being repeated. Examples of possible reinforcers for young children may include verbal praise, or offering the child a desired toy.

Punishment: A punisher is a consequence that decreases the probability or frequency of that response. Possible punishers for young children may include verbal disapproval or withholding a desired object or activity. The term "punisher" is a technical term used in behavioural therapy and does not imply the use of punishment (physical abuse such as hitting, slapping, spanking, or pinching).

It is necessary to keep in mind that, reinforcers and punishers are different for each child. Many different specific behavioural and educational techniques have been used as part of interventions for individual children with autism. These techniques are effective in a wide body of research

based on a common set of behavioural and learning principles. Behavioural interventions involve the therapist controlling the activity and/or consequences to shape the child's responses.

Discrete Trial Teaching (DTT):

This is an ABA based therapy which was developed in the 1970's by psychologists Ivar Lovaas, and Robert Koegel, at the University of California at Los Angeles (UCLA). This principle makes use of the idea that when children with autism are rewarded for desired behaviors, they are likely to repeat that behaviour. For example: the therapist gives the child a motivating cue, such as a request to stand still or maintain eye contact, along with a correct response. The therapist then rewards the child by a praise, toys or food to reward the child for completing the task.

Discrete Trial Teaching consists of a series of distinct repeated lessons or trials taught one- to - one. Each trial consists of a prior, a "directive" or request for the individual to perform an action; a behaviour, or "response" from the person; and a consequence, a "reaction" from the therapist based upon the response of the person. Positive reinforcers are selected by evaluating the individual's preferences.

Many children initially respond to, recognizable or concrete reinforcers such as food items. These concrete rewards are faded as fast as possible and replaced with rewards such as praise and hugs. Early intensive behavioural intervention such as the Lovaas program is usually implemented when the person is young, before the age of six. Parent training is a necessary part of an effective ABA-based program. A maintenance schedule allows for periodic checking so the person does not regress in mastered skills. Discrete trial training is a technique that can be an important element of a comprehensive educational program for the individual with autism spectrum disorder. In some cases, a much less intensive, informal approach of discrete trial training may be provided by a knowledgeable professional to teach specific skills such as sitting and attending.

Pivotal response therapy (PRT):

This therapy is also referred to as pivotal response treatment or pivotal response training, is a behavioural intervention therapy for autism. The main principle of this therapy suggests that behaviour links primarily on two 'pivotal' behavioural skills, i.e. motivation and the ability to respond to multiple cues, and that development of these skills will result in overall behavioural improvements.

The primary pivotal areas of pivotal response therapy involve:

1. Motivation
2. Initiation of activities
3. Self - Management
4. Feeling to respond to multiple cues
5. Ability to respond to multiple cues

The PRT training is child - directed: where the child makes the choice in which direction to direct the therapy session. Emphasis is also placed upon the role of parents as primary intervention agents. This training focuses on increasing a child's motivation to participate in learning new skills.

Pivotal response training involves specific strategies such as

- Clear instructions and questions presented by the therapist
- Child choice of stimuli (based on choices offered by the therapist)
- Intervals of maintenance tasks (previously mastered tasks)
- Direct reinforcement (the chosen stimuli is the reinforce)
- Reinforcement of reason for purposeful attempts at correct respond
- Turn taking to allow modeling and appropriate pace of interaction

Pivotal response training has a naturalistic training method that is structured enough to help children learn simple through complex play skills, while still flexible enough to allow children to remain creative in their play. The child can be reinforced for single or multiple step play. The therapist has the opportunity to model more complex play and provide new play ideas on his/her turn.

Reciprocal Imitation Training:

Reciprocal imitation training (RIT) is a variation on the pivotal response training procedure for teaching play skills. This training was developed to teach spontaneous imitation skills to young children with autism in a play environment; however, this intervention technique has also been shown to increase pretend play actions. This procedure

includes unexpected simulation in which the therapist imitates actions and vocalizations of the child.

Self-Management Training:

This is an additional approach for teaching children with autism to maximize in independence and generalization without increased reliance on a teacher or parent. Self-management typically involves some or all of the following components: self-evaluation of performance, self-monitoring, and self-delivery of reinforcement. Ideally, it includes teaching the child to monitor his / her own behaviour in the absence of an adult. This therapy uses a self-management treatment package to train school-age children with autism to engage in increased levels of appropriate play. Self-monitoring procedures have also been used to increase social initiations while reducing disruptive behaviour and to increase independent interactions with typical peers.

In a study children displayed very little independent appropriate play before training, and typically engaged in inappropriate or self-stimulatory behaviour when left on their own. With the introduction of the self-management training package, the children increased their appropriate play in both supervised and unsupervised settings, and across generalization settings and toys. Decreases in self-stimulatory and disruptive behaviours were maintained in the unsupervised environments.

Video Modeling:

This is like vivo modeling which uses predictable and repeated presentations of target behaviours; however, these behaviours are presented in video format, thus reducing variations in model performance. Studies suggest that video modeling improves various skills in individuals with autism, including conversational speech like verbal responding, helping behaviours and purchasing skills. This medium has also been claimed to increase vocabulary, emotional understanding, attribute acquisition, and daily living skills.

Video modeling interventions can be used in 2 forms:

1. Self-as-model: In this individuals act as their own models and the video is edited so that only desired behaviours are shown.
2. Other-as-model methods: This employs taping other individuals, typically adults or siblings, performing target behaviours

Applications of video modeling as an intervention technique are now being extended to teaching and increasing play in children with autism.

Therapeutic Use of Applied Behavioural Analysis:

It is extremely important to have a trained ABA therapist and to give adequate training to the parents or caregivers. ABA is the best method to manage the undesirable and anomalous autistic behaviours such as: self-injurious, repetitive, ritualistic, aggressive and disruptive behaviours. This approach has been seen and studies to extinguish or reduce these behaviours and promote alternative pro-social behaviours simultaneously. The process of ABA is successful as it breaks complex tasks into smaller parts making them less daunting for the child. ABA can also be used to train a child to learn a new adaptive behaviour, such as dressing and toileting and to promote functional communication.

ABA whether they are observed or taught they function on 3 aspects:

- Antecedent (A) - what triggered a behaviour or what happens before the behaviour,
- Behaviour (B) - the behaviour itself, and
- Consequence (C) - what happens after the behaviour.

The consequence is whatever the behaviour accomplishes, for example it can be getting attention (negative or positive) or relief of stress. The consequence is not always obvious, especially in the case of unusual behaviour (odd behaviours the child does such as arm flapping or repetitive actions), which is why keeping data is helpful to identify what the function of the behaviours are, as well as what triggers them.

Applied Behavior Analysis assessment focuses on:

- Exactly what behaviours are performed by the child
- When these behaviours are performed
- At what rate the behaviours are occurring
- What happens before and after the behaviours
- What purpose does the behaviour serves

Skills that are to be promoted are broken down into small sequential steps. The ABC principles of

behaviour intervention are used to teach the child each step:

- **A (antecedent):** Each instruction is given clearly, in as few words as possible. Assistance is provided; for example prompting through demonstration or physically guiding.
- **B (behaviour):** An appropriate behaviour is observed.
- **C (consequence):** A consequence is an outcome that will reward the child and increase the likelihood that the behavior will be repeated again in the future, also called a positive reinforcer.

ABC Model

This "ABC" process is repeated f for each behaviour both in structured teaching situations and in the course of everyday activities using PRT techniques.

i. Instructions are given to emphasize metacognition (learning how to learn). In this case learning how to listen, to watch, to imitate, to ask and do.

ii. As the child's learned behaviours improve, the structured guidance is systematically reduced and the prompts are used less frequently and eventually faded out. This so that the child learns to perform the trained behaviour independently.

iii. It is also important to change the context of teaching (different people giving the antecedent, more people around, different situations, etc.) in order to generalize the learned behaviours.

iv. When each behaviour consisting of single sequential steps are acquired, the person is taught to combine them to produce more complex behaviours.

v. Problematic behaviours are not reinforced; instead the child is consistently redirected to engage in appropriate behavior.

vi. The child's responses during each step are meticulously recorded. The information is later used to determine if the child is progressing at an acceptable rate. If progress is not satisfactory, the learning steps are analyzed for possible flaws and the program modified.

Applied Behaviour Analysis: Example of the teaching procedure:

Objective: The child will be able to make a chutney cheese sandwich.

Materials Required: Bread slices, butter, chutney, cheese, knife and plate.

Number of trials in set: Up to 5

Teaching Procedure:

Step	Presentation	Response	Consequence
1.	Mother making a sandwich		
2.	Tell the child to make the sandwich one at a time: 1. Get the plate and the knife and assist him. 2. Get the chutney, butter and cheese from the fridge and then assist him. 3. Get the bread out of the bag and assist him. 4. Open the butter packet and take cheese slices and assist him. 5. Spread the butter and assist him 6. Spread the chutney and cheese slice and a ssist him.	Child performs part of each step with assistance. 3 consecutive trial	Correct = Praise Incorrect = Error interruption, re-administer with increased prompts No response= System of most prompts Re- administer with increased prompts
3.	Tell the child to make the sandwich Model the steps " 2 - 5"	The child is able to perform step 1 without any prompt.	Same
4.	Tell the child to make the sandwich Model the steps " 3 - 5"	The child is able to perform step 1 and 2 without any prompt.	Same

5.	Tell the child to make the sandwich Model the steps " 4 - 5"	The child is able to perform step 1, 2 and 3 without any prompt.	Same
6.	Tell the child to make the sandwich Model step " 5"	The child is able to perform step 1, 2, 3 and 4 without any prompt.	Same
	Tell the child to make the sandwich	Child performs step 1 - 5 without any prompts.	Same

Social Stories: Social stories are used to teach social skills through the use of accurate information about the situations that the child with autism may find difficult or confusing. Social stories can be used for different purposes like to prepare the child for an upcoming routine, or learn to communicate appropriately in social situations. The concept behind this form of therapy is that the child rehearses the story ahead of time with an adult, which would help the child to act in an appropriate socially acceptable way when the situation arrives.

Social Stories use different types of sentences for example:

1. Descriptive Sentence: This explains the, who, what, where and why about the situation, which helps the child recognize the situation as and when it occurs. For example: There will be many shoes to choose from.
2. Directive Sentences: This suggests the appropriate social response in the situation. For example: When I decide about the shoes, I will tell the grown-up.
3. Perspective Sentences: This describes the possible feelings or the response. For example: I might not know which shoes I like.
4. Affirmative Sentences: This describes the laws or rules that are commonly shared. For Example: That is okay with everyone.
5. Cooperative Sentences: Describe how other people will help out in the given situation. For example: The grown-up will go get the shoes for me.
6. Control Sentences: This helps the child remember strategies that work for him or her. For Example: I can hold onto my string while I decide.

Theory behind Social Story:

Autistic individuals have difficulty with reciprocal social interaction and this impairment might result from unusual activity or functioning of certain areas of the brain that are used for social skills. [20] Some researchers believe that individuals with autism have trouble understanding what others believe, know, or don't know. This difficulty is sometimes called the theory of mind deficit in autism. [21] Recent research studies show that social stories can help reduce problem behaviours, increase social awareness, and / or teach new skills. In some cases, the new behaviours were maintained and generalized to other situations, even after the story was faded out. Social stories are most useful for children who have basic language skills. [22]

Floortime (DIR):

This therapeutic technique which is based on the Developmental Individual Difference Relationship Model (DIR) was formulated by Dr. Stanley Greenspan in 1980s. The premise of floortime is that an adult can help a child increase his circles of communication by meeting him at his developmental level and building on his strengths. Therapy is often incorporated into play activities on the floor.

There are six developmental milestones that contribute to emotional and intellectual growth:

1. Emotional Ideas
2. Emotional Thinking
3. Complex communication
4. Two-way communication
5. Self -regulation and interest
6. Intimacy or a special love for the world of human relations

The therapist or parent engages the child at a level where the child enjoys the activities and the therapist then follows the child's lead. The parent is instructed how to move the child toward more increasingly complex interactions, a process known as opening and closing circles of communication. Floortime does not separate and focus on speech, motor, or cognitive skills but rather addresses these

areas through a synthesized emphasis on emotional development. The intervention is called Floortime because the parent gets down on the floor with the child to engage him at his level. Floortime is considered an alternative to and is sometimes delivered in combination with behavioural therapies. Floortime is usually delivered in a low stimulus environment, ranging from two to five hours a day by a psychologist or a special educator.

Role of occupational Therapy in Autism

Occupational Therapy

Occupational therapy services focus on enhancing participation in the performance of activities of daily living (e.g., feeding, dressing), instrumental activities of daily living (e.g., community mobility, safety procedures), education, work, leisure, play, and social participation. For an individual with an ASD, occupational therapy services are defined according to the person's needs and desired goals and priorities for participation. [23]

Occupational therapy services for individuals with an ASD include evaluation, intervention, and measurement of outcomes. Throughout the process, collaboration with the child or adult with autism, family, caregivers, teachers, and other supporters is essential to understanding the daily life experiences of the individual and those with whom he or she interacts. Occupational therapy services can focus on personal development, quality of life, and the needs of the family and individual.

The occupational therapy evaluation process is designed to gain an understanding of the individual's skills-his or her strengths and challenges while engaging in daily activities (occupations). The occupational therapy intervention process is based on the results of the evaluation and is individualized to include a variety of strategies and techniques that help clients maximize their ability to participate in daily activities at home, school (if relevant), work, and in the community environment. The evaluation process looks at the child's development in a number of domains including motor, perceptual, communication and interaction skills; habits, and routines. An understanding of the child's abilities, needs, and goals is gained through interviews with the child, parents, siblings, teachers, and / or caregivers; standardized tests; and observation of the child during activities at school and home such as classroom tasks, mealtimes and play.

Progress or a successful outcome is noted through improved performance (or adaptation), enhanced participation in necessary or meaningful daily activities, personal satisfaction, improved health and wellness, and successful transitions to new situations and roles. These measures can help the individual, family, and team appreciate success and refocus and change priorities of the intervention plan as needed.

Occupational therapy practitioners help people with autism adjust tasks and conditions to match their needs and abilities. Such help may include adapting the environment to minimize external distractions, finding specially designed computer software that facilitates communication, or identifying skills they need to accomplish tasks

Occupational Therapist focuses on:

Evaluating an individual to determine whether he or she has accomplished developmentally appropriate skills needed in such areas as grooming and play and leisure skills.

- Provide interventions to help an individual respond to information coming through the senses. Intervention may include developmental activities, sensory integration or sensory processing, and play activities.
- Facilitate play activities that instruct as well as aid a child in interacting and communicating with others.
- Devise strategies to help the individual transition from one setting to another, from one person to another, and from one life phase to another.
- Collaborate with the individual and family to identify safe methods of community mobility.
- Identify, develop, or adapt work and other daily activities that are meaningful to enhance the individual's quality of life. [24]

Assessment:

A combination of standardized evaluations and observations of behavior provides a holistic picture of child's ability and needs.

Assessment strategies -

Assessment of sensory integration for a child with ASD includes:

- Evaluation of sensory responsiveness (Over,under,or labile responsiveness)3

- Sensory preferences (likes and dislikes) [25]
- Ability to attach meaning to sensation and use of sensation for adaptive behavior
- Assessment of praxis
- Assessment of participation in daily living activities

Useful Assessment Tools:

In autism, it is important to consider both structured and unstructured evaluation with children in Autism Spectrum.

Children with high functioning ranges often are able to be tested with structured evaluation of sensory integration such as SIPT (Sensory Integration and Praxis Test)

Useful assessment strategies for evaluating Sensory processing deficits:

- Skilled observation in school, home, and other environments.[26]
- Sensory histories / questionnaires such as sensory profile
- SIPT (Ayres,1989) for those children with intelligence in normal range.
- **(Sensory Integration and Praxis Test)**

Sensory Intervention mainly focuses on:

- Helping to organize behavior
- Improving feedback about body
- Helping the child attach meaning of sensation
- Providing foundation for praxis
- Encouraging rapport and social interaction
- Incorporating the child's sensory needs into his / her daily life

Occupational Therapy Intervention for Children with Autism

The goal of occupational therapy interventions is to enable individuals to participate in everyday occupations. This may be achieved through a range of therapy approaches such as modification of tasks and / or the environment to match individuals' abilities, developing skills such as posture and coordination, or development of daily routines to facilitate adaptive behaviours. [27] Occupational therapists work with children in their natural settings; in the home with their families, in the school and in the community.

- Research suggests that the behaviours of children / youth with ASD have a significant impact on family roles and activities (Werner DeGrace, 2004). The effect of having a child with ASD varies among family members and depends on available community supports (Galvin-Cook, 1996). Families are devoted to the needs of the child and consequently may have difficulty engaging in positive family experiences (Werner DeGrace, 2004).

Treatment Effectiveness:

Research in the area of autism has developed a great deal over the past decade. There are currently several studies going on within Canada which have potential to provide valuable information for the planning and implementation of health services for these children and their families.

Intervention for Challenging Behaviour

In the past, the use of intrusive treatments, such as seclusion or restraints, were once used to treat problem behaviour (Perry et al., 2003). It is now the practice to use non-intrusive methods emphasizing positive-based strategies. Intrusive methods are used only as a last resort (when non-intrusive methods are ineffective) (Perry et al., 2003).

This review concluded that the most efficacious strategies for problematic behavior in children with autism spectrum disorder were: behavior enhancement strategies, behavior reduction strategies, educational strategies, and ecological strategies (NIH 1991- Canada).

The Children's Mental Health Ontario completed an analysis of interventions for challenging behaviours in children with autism (Perry et al., 2003). Positive behavioural supports, which are non-intrusive methods for treating such behaviour, were recommended as the first course of treatment. Several studies conducted in Ontario support its effectiveness in treating symptoms of self-injury, aggression, and disruptive behaviour (Perry et al., 2003). Horner et al (2002), in a research synthesis on problem behaviour intervention for young children with autism, found that the most common problem behaviours were aggression / destruction, disruption / tantrums, self-injury, and stereotypy. Stimulus-based or instruction based interventions are the most common form of treatment for autistic children with problem behaviour. Horner et al. (2002) also reported that early use of behavioural

interventions may reduce problem behaviour by 80-90%.

Comprehensive Programs:

Intensive Behavioral Intervention (IBI) has been researched extensively and has been shown to be an effective intervention with children with autism. This intervention teaches children to respond to specific words and environmental stimuli using repetition. According to Couper and Sampson (2006), IBI is thought to provide a superior outcome since it specifically targets the deficit areas in children with autism. Treatment needs to become more naturalistic, and within the child's own environment.

Occupational therapy treatment has focused on two main areas:

1. Sensory Motor Integration

Sensory processing is the normal neurological function that all people experience when their brain processes sensory information from the environment around them. Most of us unconsciously learn to combine our senses (sight, sound, smell, touch, taste, balance, body in space) in order to make sense of our environment. Children with autism face difficulty while doing this.

Sensory integration therapy is a type of occupational therapy (OT) that places a child in a room specifically designed to stimulate and challenge all of the senses. During the session, the therapist works closely with the child to encourage movement within the room.

Sensory integration therapy is driven by four key principles

1. The child must be able to successfully meet the challenges that are presented through playful activities (Just Right Challenge);
2. The child adapts her behavior with new and useful strategies in response to the challenges presented *(Adaptive Response);*
3. The child will want to participate because the activities are fun (Active Engagement); and
4. The child's preferences are used to initiate therapeutic experiences within the session (Child Directed).

Sensory integration therapy **is based on the assumption that the child is either overstimulated or understimulated by the *environment* Therefore, the *aim of sensory integration therapy* is to improve the ability of the brain to process sensory information so that the child will function better in his *daily activities.***

In the past, one of the more frequent sensorimotor interventions in occupational therapy was sensory motor integration. There is evidence that children with autism do process sensory information differently from other developing children (National Institute of Child Health and Development, 2005- Canada). Therefore, the focus in occupational therapy has shifted to understanding how and when a child is reacting poorly to a sensory experience and structuring the environment to accommodate or minimize such reactions.

Occupational therapists can use a mediator or consultation approach to work with parents and teachers to provide strategies to prevent reactions to sensory experiences from limiting daily activities.

By adapting the tasks and environments as well as working with the families on how to teach new skills and build calming or alerting activities into their everyday routines, occupational therapists can make a difference in the family's day to day life. In particular, occupational therapy focuses on self care issues, feeding, bathing, hygiene and sleep which are significant issues for children and enormous stressors for the family.

In the school setting, a student's occupational performance may be impaired by sensory, developmental, attentional and/or learning challenges (Sahagian Whalen, 2003). Occupational therapists may adapt classroom tasks and the school environment to promote a child's participation.

Occupational therapists can assist teaching assistants and teachers with understanding the impact of sensory processing difficulties on daily functioning and how they can modify what they do to maximize the child's participation and reduce behavioural difficulties.

As some children with autism find changes to routines, or unstructured time difficult to comprehend and adjust to, environmental supports and structures can improve the quality of life for children with autism.

Common sensory integrative -related behaviors of children with ASD [29]

- Difficulty intiating and maintaining social interaction and relationships

- Communication impairments, including language delay or echolalia
- Repetitive, stereotypical play
- Often visually focused
- Sometimes good motor skills
- Difficulty calming and regulating states of arousal
- May have cognitive deficits
- Confusion about the effect and consequences of behaviors

Common occupational performance difficulties [30]

- Poor social participation due social isolation or poor social skills
- Difficulty initiating and maintaining social interaction and relationships
- Repetitive, stereotypical play
- Some children having difficulty with daily life skills, such as eating a healthy selection of foods or wearing a variety of weather appropriate clothing due to atypical sensory responsiveness
- Difficulty with adaptive behaviors, such as toileting, eating and other daily life skills
- Academic problem due to cognitive deficits
- Poor sleep-wake, cycles that interfere with daily routines
- Engagement in restrictive, repetitive, and potentially self- injurious behaviours and interests that limit the child's ability to learn and fit in with peers
- Delayed play skills often results in delays in learning and social skills
- Communication deficits, including language delays or echolalia, limit opportunities for social participation and play
- Difficulty carrying out daily living tasks, such as playing with other children, participating in mealtime with family, and participating in family routines and outings.

Key considerations in using a sensory integration approach with children with ASD

Children with ASD may demonstrate a wide variety and range of sensory processing difficulties ranging from extreme sensitivities to considerable lack of responsiveness. They may also demonstrate:

Inability to cope with unexpected or intense sensations

- Difficulty registering and attending to salient sensory input
- Many children with autism demonstrate difficulty maintain selecting and allocating attention due to poor registration of sensory input (courchesne et al,1994)
- Heightened sensitivities
- Variability in reactions to sensation
- Gravitational insecurities
- Sensory- seeking and sensory- avoidance behaviors in relation to movement, audition, touch, smell and taste
- Self stimulatory behaviours
- Problems in processing tactile information
- Strengths in visual memory and visual manipulation of objects
- Auditory processing problems
- Motor planning deficits resulting in:
- Poor ability to initiate new ideas for play
- Disorganized behavior
- Inability to carry out daily routines independently
- Poor initiation, especially mouth and facial gestures (Poor oral praxix)

Poor sensory processing may affect the child's ability to participate in his or her daily life activities, including self care activities, such as brushing his or her teeth, eating and social activities (eg. Playing with his or her peers)

Many children with ASD demonstrate difficulties with praxis in general, but especially ideational praxis. Ideational praxis is essential for self-initiated play, and thus poor ideational praxis may play role in the tendency for children with ASD to prefer sameness and routine.

Poor sensory processing associated with impaired motor planning cause difficulty in imitation and poor social reciprocity.

Intervention to Organize Behaviour

Useful sensory activities help child become calmer and more organized

Occupational therapist provides proprioceptive and deep pressure activities, linear vestibular input to become calmer and more organized.

Intervention to improve Sensory Feedback from the Body

Many children with autism have poor body awareness because they are not obtaining adequate somatosensory feedback from their body (ie, tactile proprioceptive, vestibular and interocetive sensations). Occupational therapist give activities, which are rich in somatosensory sensations and provide foundation fpr body awareness and motor planning needed for praxis.

Activities to Improve Tactile Sensation

Tactile activities help children gain awareness of their bodies and are useful for helping children learn to move and manipulate their body and hands in a coordinated, planned manner.

Activity to improve tactile sensation
(clay dough activity)

Activities rich in tactile sensations:

- Go to petting zoo
- Messy play with hands in pudding, ice creams, mashed potatoes, cookie dough, etc.
- Find objects hidden in a container of a sand, beans, macroni, rice, corn packing peanuts, feathers and pompom balls, foam wedge, buttons and beads, dirt, flour and sugar mixture or cinnamon and oatmeal etc.
- Water play with squirt guns, water balloons and spray bottles
- Write and colour with vibrating pen
- Textured balls, cloths, mitts
- Feel magic bags filled with surprises
- Finger paint- add texture with sand or rice for variety
- Apply lotion after bath with firm pressure
- Warm towels / cloth with blow dryer after bath
- Walk barefoot outdoors on variety of surfaces
- Draw letters and shapes with finger on a carpet square and erase them with hands or feet
- Have child identify shapes, letters, and numbers drawn on his or her arm, leg or back

Activities to improve Proprioceptive Sensation

Proprioception is the sensation that has been observed to help children regulate their responsiveness to sensation (Balance & Schaaf,2001). Proprioception occurs when we move our muscles. Movement of our muscles against gravity or against a weight increases the amount of proprioceptive stimuli.

Home Activities to improve proprioception

Pushing the wall to increase
proprioception

- Play on pull-up bars
- Jump on trampoline
- Use in-line skates or roller skates
- Ride a scooter or bicycle
- Jump or climb in and out of inner tubes
- Participate in climbing activities
- Play running and jumping games
- Swing from trapeze bar
- Play in sand box with damp, heavy sand
- Use cardboard boxes with blankets and pillows to make forts
- Put heavy objects in a cardboard box and have the child push it through cone- driving a car
- Play catch with heavy ball, or bounce and roll heavy ball

- Things to climb on
  - Balls
  - Crash cushions
  - Mats
  - Rock wall
  - Large bolster lying on angle
  - Tables and chairs
  - Rope ladder
  - Large inflated mats / coushions
  - Large boxes
- Push against a wall
- Fill up big toy trucks with heavy blocks, pushing with both hands to knock things down

Swinging - Vestibular Stimulation

Transition Activities to improve Proprioception:

Wheel Barrow Walking

- Roll, walk, or run up a hill
- Wheelbarrow walking
- Open and close door for people
- Walk up a ramp or incline
- Animal walk (Crab walk, bear walk)

Activities Rich in Vestibular sensation:

Vestibular activities include any movements that involve the head moving through space. To stimulate the vestibular system use activities that are stop and go and that occur in a variety of planes. For the child who is overly sensitive to vestibular movement, or who needs calming, use vestibular sensations that are in the linear plane and that are slow and rhythmical like swinging or rocking.

Sensory Integration Room

Swinging and Sliding Activities:

- Have the child sit on a swing and drop beanbags placed between knees or feet onto a target
- Have the child stand on a swing (with supervision) and jump off into pillows or inner tube
- To teach pumping, have the child extend legs as swing moves forward so therapist can push on bottom of his or her feet
- Hang the swing from single point so that movement occurs in more planes, including rotational plane
- Use sitting disc (inflated, cushioned disk) for child during homework or mealtime
- Ride tricycle / bicycle
- Jump on a trampoline

- Do cartwheels and / or somersaults
- Use rocking horse or rocking chair
- Sit on parent's lap and rock back and forth or bounce up and down
- Roll or bounce on big balls
- Swing in hammock- two adults and blanket can form a hammock swing for a very small child
- Ride a scooter
- Do "stretching" exercises with head, such as rolling head around or side to side

Intervention to develop praxis-

It is very important for children with ASD to develop praxis skills because they are needed to manage everyday situations that are constantly changing. Even routine activity, such as riding the bus to school, requires that the child adapt to new stimuli and situations. (different children, different driving conditions, different seat mates, different seat belts).

Occupational Therapist help child to develop praxis by providing him or her with varied sensory experiences grounded the organizing adaptive responses. Once a desired level of organization is achieved, introduce the activities that challenge the child , and scaffold sensory experiences to facilitate skill development. For example expand activity to include increasingly complex motor planning, interaction, and language skills.

Structure the environment for simple adaptive responses and move to more complex ones, include activities that facilitate skills such as imitation, sequencing, bilateral coordination, timing and imitation.

Activities to improve Praxis:

Ayes(1989) showed that there is relationship between sensory processing and praxis, specifically somatosensory processing (Blanche, 2001). Sensory information from the body and the environment is necessary for creating ideas for movement, putting together the plan and providing feedback during and following the movement activity. Many children with difficulties processing sensory information also have poor motor planning or dyspraxia. Ayes described some activities which provide significant somatosensory input, thus underscoring the relationship between body awareness and praxis.

- Ride tricycles and bicycles
- Create and complete an obstacle course
- Participate in imaginative play (housekeeping, fireman,doctor, post office)
- Ask child to perform animal walks
- Play "simon says" games
- Play balloon volleyball
- Play "mirror image" child makes a mirror image of another child's posture
- Play "red light / green light"- child run on green light and stops on red light
- Play "follow-the-leader"- leading and following

Encourage Rapport and Social Interactions

The child's enjoyment and motivation during sensory-rich activities can improve a child's tolerance for, and satisfaction in , social arenas. Motivating sensory experiences can be used to elicit eye contact, social cuing, or social language for the child.

2. General Skill Building

Occupational therapists work with children with autism and their families to provide intervention that improves the child's ability to participate in activities of daily living, play and school. In occupational therapy, the focus is on task analysis, breaking down a task into manageable steps for the child, teaching those steps to the key people in the child's environment and structuring task and / or environment to support successful completion. For example, if a child with autism is having difficulty dressing himself, the occupational therapist could use a backward chaining behavioural approach which teaches skills by breaking the tasks down into manageable steps which are always performed in the exact same order (CanChild, 2006). Occupational therapists work as collaborative consultants, providing strategies for intervention to families, teachers, in order to develop a child's skills within their natural environments.

Some Proprioceptive rich activities during the course of daily activities and routines:

- Unpack heavy groceries
- Carry heavy items (baskets with cardboard blocks, groceries,etc.)
- Climb into weighted blankets for bed.

- Push or pull boxes with toys or few books in it (more resistance is provided if boxes are pushed / pulled across a carpeted floor)
- Take the cushions of the sofas, vacuum under them, and then put them back ; canalso climb on them or jump and "crash" into them
- Go "shopping" with a child's shopping cart filled with items, or have the child push the shopping cart when you go shopping
- Rearrange bedroom furniture
- Put large toys and equipments away
- Do chair push-ups

The focus of therapy intervention is consultation rather than direct intervention.

Speech Therapy:

Children with autism present with delayed speech and language development, severe social and pragmatic issues and behavioural problems. Speech and language problems may range from severe comprehension issues to inability to express their needs and wants. They may rely on pointing and just crying and vocalizing to express himself. Autistic children have severe problems in social skills and use of language. Poor eye contact, poor social smile, solitary play behaviour and irrelevant speech patterns are very commonly seen in these children.

Complementary therapies:

Complementary therapies are typically used in addition to behavioral and educational approaches.

Music Therapy:

Music therapy is seen to be an effective intervention for emotional recognition deficits in autism. However, researchers have yet to propose for the neurological and cognitive components that are responsible for such improvements. Individuals with autism show poor perception of affective cues within the social domain and experimental evidence suggest that such individuals fail to interpret and recognize vocal and facial expression of emotions. The two essential goals when music therapy is conducted for individuals with autism is improving communication / language and improving socio-behavioral skills (Kaplan & Steele, 2005). Buday (1995) observed that the number of signed and spoken words correctly imitated by children with autism was higher using sung rather than spoken text [32].

Music as a mode of intervention has yielded beneficial effects for the modification of behavioural problems. Orr, Myles, and Carlson (1998) investigated the effect of rhythmic entrainment on erratic classroom behaviours in their case study of a girl with autism [33]. Rhythmic entrainment involves the use of music to aid relaxation by the introduction of externally produced rhythms, designed specifically to re-entrain the body to its natural rhythmic patterns. They found that rhythmic entrainment helped in reducing problematic classroom behaviour.

Music therapy in autism

Even though music therapy has been used with success, there is still a shortage of theoretical papers that deal with understanding the neural substrates and cognitive mechanisms underlying the improvement in such adaptive skills [9].

Improvisational music therapy, a form of music therapy where there is interactive use of live music for engaging clients to meet their therapeutic needs. This is widely used in the treatment of children with autism and is gaining growing recognition as an effective intervention addressing fundamental levels of spontaneous self-expression, emotional communication and social engagement for individuals with a wide range of developmental disorders. Within this therapeutic intervention, the music therapist identifies musical elements (temporal beat, rhythmic patterns, dynamics of expression, pitch range and melodic contour) in the child's musical and non-musical behaviour, and then provides a predictable, empathic and supportive musical structure to attract and engage the child

Art Therapy:

What Is Art Therapy?

According to the American Art Therapy Association, "art therapy is a mental health profession that uses the creative process of art making to improve and enhance the physical, mental and emotional well-being of individuals of all ages. It is based on the belief that the creative process involved in artistic self-expression helps people to resolve conflicts and problems, develop interpersonal skills, manage behavior, reduce stress, increase self-esteem and self-awareness, and achieve insight."

For children and adults with autism, it can be a very useful way to emote and express themselves.

Art Therapy in Autism

Why Use Art Therapy to Treat Autism?

One of the hallmarks of autism spectrum disorders is difficulty with verbal and social communication. In some cases, people with autism are literally non-verbal: unable to use speech to communicate at all. In other cases, people with autism have a hard time processing language and turning it into smooth, easy conversation. People with autism may also have a tough time reading faces and body language. As a result, they may have difficulty with telling a joke from a statement, or sarcasm from sincerity. [34]

Meanwhile, many people with autism have an extraordinary ability to think visually - "in pictures." Many can turn that ability to good use in processing memories, recording images and visual information, and expressing ideas through drawing or other artistic media. Art is a form of expression that requires little or no verbal interaction which can open doors to communication.

All too often, it's assumed that a non-verbal person or a person with limited verbal capabilities is incompetent in other areas. As a result, people on the autism spectrum may not be exposed to opportunities to use artistic media -- or the opportunities may be too challenging in other ways (in large class settings, for example). Art therapy offers an opportunity for therapists to work one-on-one with individuals on the autism spectrum to build a wide range of skills in a manner which may be more comfortable (and thus more effective) than spoken language. Group Art based therapy will improve social interaction and will help the child vent out his emotions.

Group Art Based Therapy

Play Therapy:

If play is a child's language, then toys can be thought of as the words. Through play therapy the child can work through their challenges and issues using the toys that they choose, revealing their inner dialogue. Through play the child is able to test out various situations and behaviors in a supportive environment. Unconditional positive regard and acceptance encourages the child to feel safe enough to be able to explore their inner selves without censorship. In this environment children are able to try out different roles, work through conflicting emotions and thoughts, and try to figure out what the world is like. The child is able to form a relationship with the provider, and through this relationship they are able to develop trust, improved self-esteem, and self efficacy.

In non-directive play therapy, the child is in control, within some gently but firmly set limits. Children often feel that they do not have control over situations in their lives. Through play therapy they are able to work through these experiences in an environment that they are able to control. They can make the story be how they want it to be, they are in charge of the outcome. This feeling of control is vital to their emotional development as well as

positive mental health. Children are able to use play as a means for developing problem-solving skills, ways to relate to others, expressing their feelings, and working on their behaviors, all at a safe psychological distance from reality.

Play Therapy

Play Therapy in Autism:

As play therapy has been shown to be effective with children who have social and emotional difficulties, it fits with some of the treatment goals of autism. Using unconditional positive regard, the child is accepted at their current level of functioning along with the assumption that they have an intrinsic drive towards improved functioning. This allows children who have autism to be able to work at a pace and focus on change that is fitting to them, increasing self-efficacy and autonomy. In autism research it has been strongly advocated that the therapeutic interventions need to be attuned to the individual child's developmental level. Play therapy automatically operates at the child's current level and is highly individualized, as the children determine the pace and focus of change (Josefi & Ryan, 2004).

Animal therapy, such as horseback riding and swimming with dolphins, improves the child's motor skills while increasing self-confidence.

Animal Therapy

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Ch.6 Dementia

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INTRODUCTION:

Dementia is defined as a progressive impairment of cognitive functions occurring in clear consciousness (i.e. in absence of delirium). This is a disease which has a significant global impairment of intellect, manifested as difficulty with memory, attention, thinking and comprehension. Other psychological functions can be included including mood, personality, judgment, and social behaviour. There are specific diagnostic criteria that are found for various dementias, such as Alzheimer's disease or vascular dementia, however all dementias have certain common elements that result in significant impairment in social or occupational functioning and cause a significant decline from a previous level of functioning.[1]

The disorder can be progressive or static, permanent or reversible. An underlying cause is always assumed, although in rare cases, it is impossible to determine specific cause. Approximately 15 percent of patients with dementia have reversible illness if treatment is initiated before irreversible damage takes place.

French Physician, Philippe Pinel who coined with the term "demence" i.e dementia.

HISTORY:

Western literature for centuries has described a disease process what we call as "dementia" today. "Dementia is a word which is given by a French physician named Philippe Pinel. In 1801 he was working on a 34 year old woman who lost her memory, speech, ability to walk or use common objects like a fork or a hairbrush and Pinel called this process "demence". He used the words"demence" to mean an "incoherence" of mental faculties to describe her disease. Today the word dementia indicates a person having cognitive impairment significant enough to interfere with daily functioning and describe one of more than 48 types of these diseases in the brain.

When Pinel's patient died, he autopsied her brain. Using a primitive microscope, he studied the brain tissue. With his microscope, he was only able to describe two distinct features of her disease. He wrote that the woman's brain was full of fluid and it had dramatically shrunken in size.

In 1907, a German physician published a paper on a patient of his who exhibited behaviours similar to Pinel's patient. This woman was in her fifties and she appeared to have the same disease Pinel described. The woman suffered a "failure of memory, paranoia, loss of reasoning powers, incomprehension and stupor."

When the German physician looked at her brain however, he had a more advanced microscope, an optical microscope. And in writing a research paper about his patient, Alois Alzheimer described the disease process for which he is known today. Alzheimer described a brain that was (1) shrunken and (2) full of fluid, but also (3) suffered structural damage in the form of neurofibrilary tangles and (4) had bone structures growing in the brain tissues. These are the four hallmark features of a brain with Alzheimer's disease.

Alzheimer sent his paper documenting his findings to his mentor, Dr. Emil Kraepelin. When Dr. Kraepelin published his eighth medical textbook, Alzheimer's paper and research were included and the disease became known as Alzheimer's disease. One of the challenges for us today is the pronunciation of Alzheimer's. Some people have a tendency to pronounce Alzheimer's like the words

"Old-Timers," which continues to enforce the thought that as we age we lose our mental abilities. Alzheimer's is actually a disease and is not considered a part of the normal aging process. Most people do not develop any type of dementia. And most people remain cognizant throughout their lifetimes. Indeed, people learn at the age of eighty at the same rate they learned at the age of twenty. Dementia is truly not a part of normal aging.

DEFINITION OF DEMENTIA:

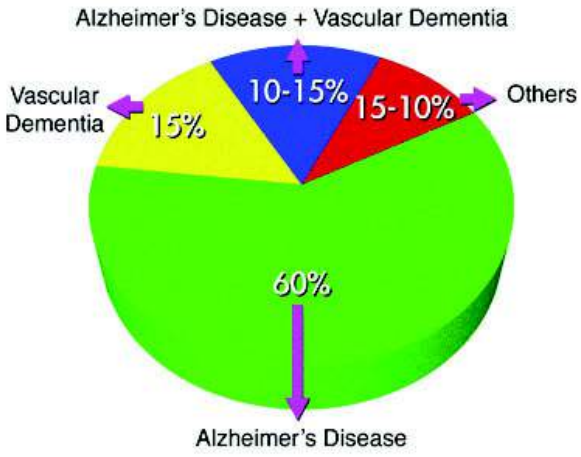
The World Health Organization (2007): "a syndrome due to disease of the brain, usually of a chronic or progressive nature, in which there is disturbance of multiple higher cortical functions, including memory, thinking, orientation, comprehension, calculation, learning capacity, language, and judgment. However, consciousness is not clouded. The impairments of cognitive function are commonly accompanied, and occasionally preceded, by deterioration in emotional control, social behavior, or motivation which occurs usually in Alzheimer's disease, in cerebrovascular disease, and in other conditions primarily or secondarily affecting the brain."

EPIDEMIOLOGY:

The prevalence of dementia is rising, with the growing aging population. The prevalence of moderate to severe dementia:

- 5 percent - general population more than 65 years of age
- 20 to 40 percent - general population more than 85 years of age
- 15 to 20 percent in outpatient general medical practices
- 50 percent in chronic care facilities

Of all the other types of dementia, 50 to 60 percent of patients suffer from the most common type of dementia i.e. the Alzheimer's type. Dementia of the Alzheimer's type increases in prevalence with increasing age. For persons aged 65 years men have a prevalence rate of 0.6 percent and women of 0.8. At age 90 years, rates are 21 percent. The second most common type of dementia is vascular dementia, which accounts for 15 to 30 percent of all cases. Vascular dementia is most common in persons between the ages 60 and 70 and is more common in men than in women.



Prevalence rates of different types of dementia

Other common causes of dementia, each representing 1 to 5 percent of all cases, include head trauma, alcohol related dementias and various movement disorder - related dementias, such as Huntington's disease and Parkinson's disease.

ETIOLOGY:

The most frequent causes of dementia in individuals older than 65 years of age are: (1) Alzheimer's disease (2) Vascular dementia and (3) Mixed vascular and Alzheimer's dementia. Other illness that account for approximately 10 percent include Lewy body dementia, Pick's disease, frontotemporal dementias normal pressure hydrocephalus, alcoholic dementia, infectious dementia, such as human immunodeficiency virus (HIV) or syphilis and Parkinson's disease. Many types of dementias evaluated in clinical settings can be attributable to reversible cases, such as metabolic abnormalities, nutritional deficiencies or dementia syndrome caused by depression.

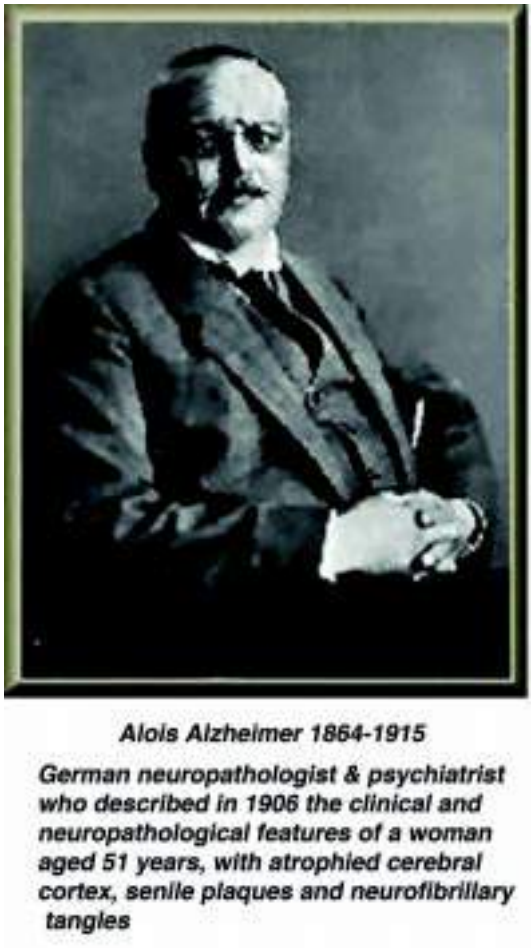
DIAGNOSIS:

Dementia is a type of chronic encephalopathy that can have many causes, including irreversible degenerative and potentially reversible nondegenerative causes. The most important diagnostic step in evaluating dementias is to determine whether a chronic encephalopathy results form a degenerative or other potentially reversible causes. Historical clues suggesting a reversible process include fluctuating severity, altered level of consciousness or hypersomnolence, and visual hallucinations. Clues on mental status testing include findings the patient to be inattentive, disoriented and somnolent but not particularly

amnestic. Clues on physical examination include a variety of findings that may be common in elderly patients are not part of the typical picture of Alzheimer's disease, such as ataxia, hyper-reflexia and tremulousness. Laboratory evaluation should be directed towards.

Dementia of The Alzheimer's Type:

In 1907, Alois Alzheimer first described the condition that later assumed his name. He described a 51 year old woman with a 4 ½ year course of progressive dementia. The final diagnosis of Alzheimer's disease requires a neuropathological examination of the brain; nevertheless, dementia of the Alzheimer's type commonly diagnosed in the clinical setting after other causes of dementia have been excluded from diagnostic consideration. The clinically defined syndrome, Alzheimer's dementia has been considered as the paradigm of a cortical dementia syndrome. The hallmark of, cortical dementia include not only memory loss, which is common to most dementia syndromes but also elements of aphasis, apraxia and agnosia.



Alois Alzheimer after whom the condition assumed his name Alzheimer 's disease.

Most clinical studies are in agreement that the earliest clinical signs of AD are memory loss, which precedes the actual dementia. Abnormal cerebral metabolism, demonstrated by positron emission tomography (PET), may precede even mild memory loss [2]. A few studies have correlated other relative cognitive inefficiencies occurring much earlier in life with the subsequent development of dementia, although interpretation of these findings have been a source of controversy.

Recent memory is easier to access reliably then remote memory and is thought to be disproportionately severely involved. Nonetheless, remote memory is also abnormal, and there is a gradient effect regarding recall over a retrograde time interval: The oldest memories appear to be the best preserved, with proportionately greater forgetting as the retrograde interval shortens [3]. In contrast procedural memory appears to be relatively spared. Alzheimer's patients are able to learn simple skills as easily as normal controls and better than patients with subcortical patterns of dementia or patients with various types of sensorimotor deficits [4]. One very important skill that needs to be addressed clinically is driving. Despite the relative preservation of procedural memory, patients with mild Alzheimer's dementia have a higher rate of collisions and moving violations than age-matched controls, although estimates of risk vary, especially during the first 2 years of the disease. Whether this actually results from impaired procedural memory, attentional factors, other cognitive aspects or a combination is unclear, although visual tracking, memory, and Mini-Mental State score all correlate with a laboratory-based driving score.

Aphasia, apraxia and agnosia are the other categories of cognitive impairment that typically occur in cortical dementia syndromes and particularly in AD. In Alzheimer's dementia, these aspects do not usually dominate the clinical picture, but they can in variant syndromes. In mild to moderate stages of dementia, anomia is prominent and readily detectable with neuropsychological testing using a variety of naming tests. However, patients are fluent and may have relatively good comprehension, so clinical detection is not always easy.

Apraxia can be confused with impaired comprehension in mild to moderate stages of Alzheimer's dementia. Patients have difficulty in learning new procedures such as a new car or

performing tasks that they were previously adept at. In moderately advanced stages, patients have difficulty in dressing and performing other activities of daily living.

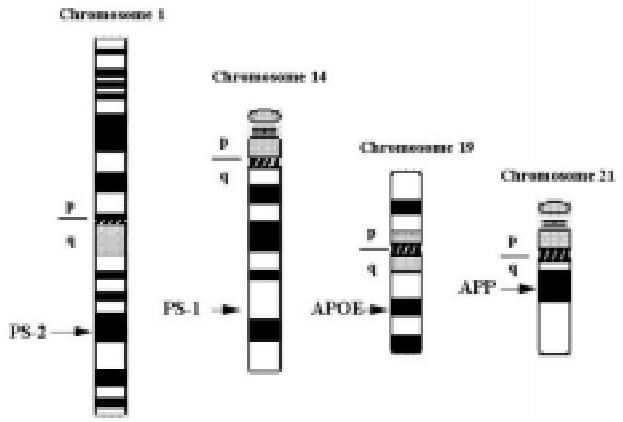
Anosognosia is the failure to recognize illness, this is a cardinal feature of Alzheimer's dementia and this is typically present, even in mild stages of the disease. In AD, a patient may deny significant memory problems and will actively try to explain away the observations of concerned family members and friends, even to a point of becoming hostile and accusative.

Psychiatric symptoms include both affective and psychotic disturbances. Depression interferes with an individual's functional status and an accurate cognitive assessment, erroneously leading to the diagnosis of dementia; such is the nature of pseudodementia, which can be estimated to account for 4 percent or 5 percent of dementia cases [5]. Psychotic symptoms, most commonly involves paranoid delusions and less commonly hallucination.

Sleep wake cycle disturbances are common and may be present even during relatively mild stages of their illness. They may become more common and more severe during moderately severe stages. There are two aspects to the sleep cycle disturbance. The first is the so called sun-drowning effect, which means that the patient becomes more confused, agitated, and difficult to manage during the evening. The second regards not sleeping at night, waking up during very early hours or going to sleep very early in the evening. As, the disease progresses to more advanced stage, patients may become generally less active and eventually in terminal stages, are bedbound with little apparent conscious activity. Weight loss is common during the latter stages as well. Incontinence is uncommon in mild stages but becomes increasingly frequent as the disease progresses and is universal in late stages.

Genetic Factors:

Although the cause of dementia of the Alzheimer's type remains unknown, progress has been made in understanding the molecular basis of the amyloid deposits that are a hallmark of the disorders neuropathology. Some studies have indicated that as many as 40 percent of patients have a family history of dementia of the Alzheimer's type thus genetic factors are presumed to play a part in the development of the disorder, at least in some cases. In several well-documented cases, the disorder has been transmitted in families through an autosomal dominant gene, although such transmission is rare. Alzheimer's type dementia has shown linkage to chromosomes 1, 14 and 21.



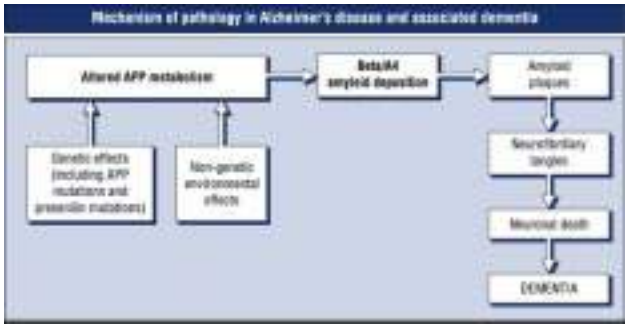
Genetic affect in Alzheimer's dementia

Amyloid Precursor Protein:

The gene for amyloid precursor protein is on the long arm of chromosome 21. Whether the processing of abnormal amyloid precursor protein is of primary causative significance in Alzheimer's disease is unknown, but many research groups are studying both the normal metabolic processing of amyloid precursor protein and its processing in patients with dementia of Alzheimer's type in an attempt to answer this question.

Multiple E4 Genes:

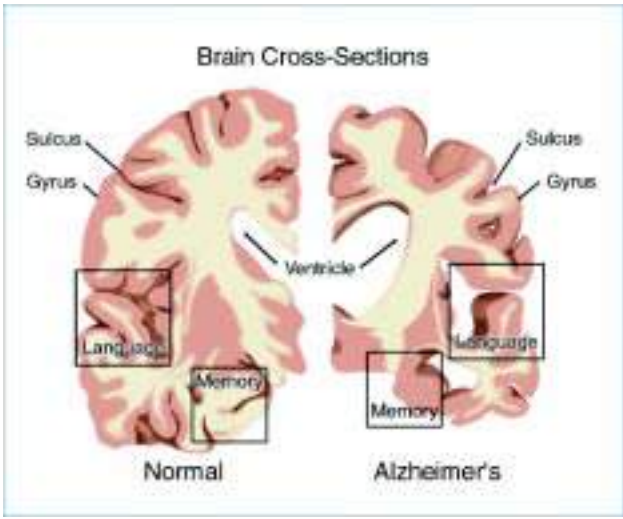
One study implicated gene E4 in the origin of Alzheimer's disease. People with one copy of the gene have Alzheimer's disease three times more frequently than do those with no E4gene, and people with two E4 genes have the disease eight times more frequently than do those with no E4 gene. Diagnostic testing for this gene is not currently recommended because it is found in persons without dementia and not found in all cases of dementia.



Mechanism of pathology in Alzheimer's disease and associated dementia

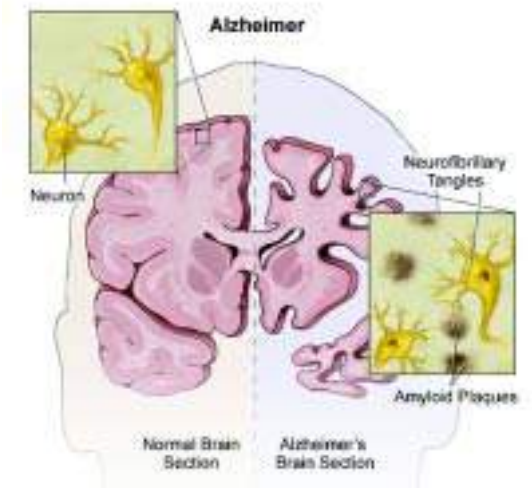
Neuropathology:

The classic gross neuroanatomical observation of a brain from a patient with Alzheimer's disease is diffuse atrophy with flattened cortical sulci and enlarged cerebral ventricles. The classic and pathognomonic microscopic findings are senile plaques, neurofibrillary tangles, neuronal loss (particularly in the cortex and hippocampus), synaptic loss (perhaps as much as 50 percent in the cortex) and granulovascular degeneration of the neurons.



Neuroanatomical Observation diffuse atrophy with flattened cortical sulci and enlarged cerebral ventricles.

Senile plaques also referred to as amyloid plaques more strongly indicate Alzheimer's disease, although they are also seen in Down syndrome and to, some extent, in normal aging. The number and the density of senile plaques present in postmortem brains have been correlated with severity of the disease that affected the person.



Neurofibrillary tangles and Amyloid Plaques in Alzheimer's patients

Neurotransmitter:

The neurotransmitters that are most often implicated in the pathophysiological condition of Alzheimer's disease are acetylcholine and norepinephrine both which are hypnotized to be hypoactive in Alzheimer's disease. Decreased norepinephrine activity in Alzheimer's diease is suggested by the decrease in norepinephrine containing neurons in the locus ceruleus found in some pathological examination of brains from persons with Alzheimer's disease. To other neurotransmitter implicate in the pathophysicological condition of Alzheimer's disease are the neuroactive peptides somatostatin and corticotrophin; diseases concentrations of both have been reported in person with Alzheimer's disease.

Other Causes:

Another theory to explain the development of Alzheimer's disease is that an abnormality in the regulation of membrane phospholipid metabolism results in membranes that are less fluid - that is, more rigid - than normal. Aluminium toxicity has also been hypothized to be causative factor, because high levels of aluminium have been found in the brains of some patients with Alzheimer's disease; but this is no longer considered a significant etiological factors. Excessive stimulation by the transmitter glutamate that may damage neurons is another theory of causation.

Familial Multiple System Taupathy with Presenile Dementia: A recently discovered type of dementia, familial multiple system taupathy, shares some brain abnormalities found in people with Alzheimer's disease. The gene that causes the disorder is thought to be carried on chromosome 17. The symptoms of the disorder include short term memory problems and difficulty in maintaining balance and walking. The onset of the disease occurs in the 40s and 50s and persons with the disease live an average of 11 years after the onset of symptoms.

As, in patients with Alzheimer's disease, tau protein builds up in neurons and glial cells, of persons with familial multiple system taupathy. Eventually, the protein build up kills brain cells. The disorder is not associated with the senile plaque seen with Alzheimer's disease.

Diagnosis:

Diagnostic criteria for Dementia of the Alzheimer's Type (cautionary statement).

A. The development of multiple cognitive deficits manifested by both (1) memory impairment (impaired ability to learn new information or to recall previously learned information) (2) one (or more) of the following cognitive disturbances:

   (a) aphasia (language disturbance)

   (b) apraxia (impaired ability to carry out motor activities despite intact motor function)

   (c) agnosia (failure to recognize or identify objects despite intact sensory function)

   (d) disturbance in executive functioning (i.e., planning, organizing, sequencing, abstracting)

B. The cognitive deficits in Criteria A1 and A2 each cause significant impairment in social or occupational functioning and represent a significant decline from a previous level of functioning.

C. The course is characterized by gradual onset and continuing cognitive decline.

D. The cognitive deficits in Criteria A1 and A2 are not due to any of the following:

   (1) other central nervous system conditions that cause progressive deficits in memory and cognition (e.g., cerebrovascular disease, Parkinson's disease, Huntington's disease, subdural hematoma, normal-pressure hydrocephalus, brain tumor)

   (2) systemic conditions that are known to cause dementia (e.g., hypothyroidism, vitamin B or folic acid deficiency, niacin deficiency, hypercalcemia, neurosyphilis, HIV infection)

   (3) substance-induced conditions

E. The deficits do not occur exclusively during the course of a delirium.

F. The disturbance is not better accounted for by another Axis I disorder (e.g., Major Depressive Episode, Schizophrenia).

Code based on presence or absence of a clinically significant behavioral disturbance:

294.10 Without Behavioral Disturbance: if the cognitive disturbance is not accompanied by any clinically significant behavioral disturbance.

294.11 With Behavioral Disturbance: if the cognitive disturbance is accompanied by a clinically significant behavioral disturbance. (e.g., wandering, agitation)

Specify subtype:

With Early Onset: if onset is at age 65 years or below

With Late Onset: if onset is after age 65 years

Coding note: Also code 331.0 Alzheimer's disease on Axis III. Indicate other prominent clinical features related to the Alzheimer's disease on Axis I (e.g., 293.83 Mood Disorder Due to Alzheimer's Disease, With Depressive Features, and 310.1 Personality Change Due to Alzheimer's Disease, Aggressive Type).

Prognosis:

Alzheimer's disease is a progressive disorder and as the patients are elderly, and susceptible to a variety of other disease processes, AD however is not the common cause of death. However, if no other disease process supervenes, death usually ensues approximately 6 t 10 years after the onset.

VASCULAR DEMENTIA:

The primary cause of vascular dementia is from impaired blood flow to the brain. Formerly referred to as multi- infarct dementia is presumed to be multiple areas of cerebral vascular disease, resulting in a symptom pattern of dementia. However, the name was changed to vascular dementia to reflect the array of conditions that can impair the blood's ability to circulate to the brain. Vascular dementia often occurs alongside Alzheimer's disease, resulting in mixed dementia between 1% to 4% of people over the age of 65 years have vascular dementia, and the risk of developing it increases dramatically with age. Vascular Dementia most commonly is seen in men especially those with preexisting hypertension or other cardiovascular risk factors.

Impaired blood flow in the brain in Vascular Dementia

Causes of Vascular Dementia:

Vascular dementia can occur either due to narrowing or a complete blockage of blood vessels in the brain, which deprives brain cells from nutrients and oxygen they need to function properly. Vascular dementia often results from several small strokes that occur over time. It can also occur after a single major stroke, which is sometimes referred to as post-stroke dementia. Not all strokes lead to dementia, but up to one-third of those who have a stroke will develop dementia within six months. Conditions like high blood pressure and diabetes that don't block blood vessels, but simply narrow them, can also lead to vascular dementia.

Epidemiology and Risk Factors for Vascular Dementia:

Incidence and prevalence rates of vascular dementia are affected by the definition used. The prevalence of post-stroke vascular dementia ranged from 11.3 percent using the NINDS- AIREN criteria to 20. 1 percent using the ICD- 10 NA criteria, and the incidence ranged from 2.6% with the ADDTC to 5.2 percent with the ICD - 10 - NA[5]. The incidence rate of AD is higher among individuals with prevelant cardiovascular disease than among those without it.

Individuals who develop vascular dementia often have a history of one or more of the following: heart attack, stroke, high blood pressure, diabetes, or high cholesterol. In particular, if an individual has a history of multiple strokes, the risk of developing vascular dementia increases with the number of strokes experienced over time.

Clinical Feature and Associated Disorders:

People with vascular dementia often display multiple cognitive problems, including memory impairment and possibly aphasia, apraxia, agnosia, or problems with executive functioning. In maximum cases this makes it difficult for the patients to have a successful career or hold a job, take care of the household activities or socialize. People with vascular dementia also experience neurological symptoms such as exaggerated reflexes, problems with walking and balance, and / or weakness in the limbs, hands, and feet. Depending on the individual and on the cause of the dementia, delusions, confusion, agitation, urinary problems, and / or depression can also accompany vascular dementia.

Patients have a more subcortical pattern of dementia with psychomotor slowing, poor learning curve, and relative preservation of naming and other language skills. The degree of impairment, however, correlates better with the degree of neurofibillary pathology than with cerebrovascular pathology at autopsy.

Interestingly, memory loss usually occurs later in the disease compared to when it appears in Alzheimer's. In vascular dementia, the first symptoms are often the neurological ones, such as problems with reflexes, walking, and muscle weakness. On the other hand, memory problems and behavioral symptoms are commonly the first issues noticed in Alzheimer's. Additionally, vascular dementia often progresses in a step-wise fashion. For example, the person will seem stable for a period of time, then suddenly get much worse, then continue to alternate between stable periods and sudden drops in functioning. On the other hand, Alzheimer's disease progresses in a more gradual, downward fashion.

Diagnosis of Vascular Dementia:

As with Alzheimer's disease, a complete diagnostic workup should be performed in order to rule out other possible causes of the person's symptoms. Vascular dementia is usually identified through imaging procedures, which can reveal strokes and narrowed or blocked arteries. Neuropsychological tests might also be conducted to determine the nature and extent of cognitive impairment.

DSM-IV Criteria for the Diagnosis of Vascular Dementia

A. The development of multiple cognitive deficits manifested by both:

    1. Memory impairment (impaired ability to

learn new information or to recall previously learned information).

2. One or more of the following cognitive disturbances:

a. Aphasia (language disturbance)

b. Apraxia (impaired ability to carry out motor activities despite intact motor function)

c. Agnosia (failure to recognize or identify objects despite intact sensory function)

d. Disturbance in executive functioning (i.e., planning, organizing sequencing, abstracting)

B. The cognitive deficits in criteria A1 and A2 each cause significant impairment in social or occupational functioning and represent asignificant decline from a previous level of functioning.

C. Focal neurological signs and symptoms (e.g., exaggeration of deep tendon reflexes, extensor plantar response, pseudo bulbar palsy, gait abnormalities, weakness of an extremity) or neuroimaging evidence indicative of cerebrovascular disease (e.g., multiple infarctions involving cortex and underlying white matter) that are judged to be etiologically related to the disturbance..

D. The deficits do not occur exclusively during the course of delirium.

Differential Diagnosis:

There is a broad range of cerebrovascular disorders that may result in cognitive impairment. Multiple large artery territory infarction and hemorrhages may result in more extensive physical disability and cortical cognitive signs such as aphasia. Non-artherosclerotic forms of cerebrovascular disease may have their own disease associations that, in turn, may influence cognition. Cerebral vasculitis may present as a rapidly progressive fulminant encephalopathy, a sub acute course of recurrent cerebral infarction or a more chronic dementia - like course.

Evaluation:

Considerations are the same as that for AD except that causes of cerebral infarction also need to be included.

Prognosis for Vascular Dementia

Currently, there is no cure for vascular dementia. If the dementia was caused by multiple strokes, the person may get worse in a step-wise progression, where stable periods are interrupted by sudden downward episodes. Life expectancy for someone with vascular dementia is highly individual and depends on the nature of the cardiovascular problems that are causing the dementia, along with the person's age and other medical conditions.

BINSWANGER'S DISEASE:

Binswanger's disease, also known as subcortical arteriosclerosis encephalopathy, is characterized by the presence of many small infarctions of the white matter that spare the cortical regions. Atherosclerosis (commonly known as "hardening of the arteries") is a systemic process that affects blood vessels throughout the body. Although Binswanger's disease was previously considered a rare condition, the advent of sophisticated and powerful imaging techniques, such as magnetic resonance imaging (MRI), has revealed that the condition is more common than previously thought.

Symptoms of the Binswanger's Disease:

The symptoms associated with Binswanger's disease are related to the disruption of subcortical neural circuits that control what neuroscientists call executive cognitive functioning:

- Short-term memory,
- Organization,
- Mood,
- The regulation of attention,
- The ability to act or make decisions, and
- Appropriate behavior.

The most characteristic feature of Binswanger's disease is psychomotor slowness - an increase in the length of time it takes, for example, for the fingers to turn the thought of a letter into the shape of a letter on a piece of paper.

Other symptoms include:

- Forgetfulness (but not as severe as the forgetfulness of Alzheimer's disease),
- Changes in speech,
- Unsteady gait,
- Clumsiness or frequent falls,

- Changes in personality or mood (most likely in the form of apathy, irritability, and depression), and
- Urinary symptoms that aren't caused by urological disease.

Diagnosis:

Brain imaging, is very essential as it reveals the characteristic brain lesions of Binswanger's disease, and is essential for a positive diagnosis.

Prognosis:

Binswanger's disease is a progressive disease; there is no cure. Changes may be sudden or gradual and then progress in a stepwise manner. Binswanger's disease can often coexist with Alzheimer's disease. Behaviors that slow the progression of high blood pressure, diabetes, and atherosclerosis -such as eating a healthy diet and keeping healthy wake/ sleep schedules, exercising, and not smoking or drinking too much alcohol -- can also slow the progression of Binswanger's disease

PICK'S DISEASE:

Arnold Pick, was the first one who described the disease in 1892, he reported that Pick's disease causes an irreversible decline in a person's functioning over a period of years. It is most commonly confused with the much more prevalent Alzheimer's disease; Pick's disease is a rare disorder that causes the frontal and temporal lobes of the brain, which control speech and personality, to slowly atrophy. It is therefore classified as a frontotemporal dementia, or FTD.

Pick's disease is a relatively rare form of dementia that causes a slow shrinking of brain cells due to excess protein build-up. Patients with Pick's initially exhibit marked personality and behavioral changes, and then a decline in the ability to speak coherently.

Pathogenesis and Pathophysiology:

In Pick's disease the brain tissue changes and loss occurs in focal areas rather than the generalized damage of Alzheimer's. Pick's disease affects the frontal and temporal lobes of the brain. Marked shrinkage, called atrophy, of the frontal lobes of the brain occurs that can be seen on brain scans.

Pick's disease is marked by the presence of abnormalities in brain cells called Pick's bodies. These are found in the affected areas as well as elsewhere in the brain. Pick's bodies are fibers that look very different from the neurofibrillary tangles found in Alzheimer's disease. Pick's bodies are straight rather than paired and helical.

Changes showing in the hippocampus gyrus and balloned achromatic neurons and pick bodies

Symptoms of Pick's Disease:

Many of the early symptoms of Pick's disease are frontal lobe symptoms. It is these symptoms that tend to mark out the differences between Pick's dementia and the other types, such as Alzheimer's. In Alzheimer's disease the initial symptoms tend to be memory impairment. In Pick's disease because the frontal lobes of the brain are affected, the first symptoms occur in emotional and social functioning. It is the mood changes, often biased towards euphoria, disinhibition and deterioration in social skills that are so noticeable.

Behavioural Symptoms Include:

- Repetitive or obsessive behavior
- Overeating or drinking to excess (when this was not previously a problem)
- Impulsivity and poor judgment
- Extreme restlessness (early stages)
- Lack of attention to personal hygiene
- Sexual exhibitionism or promiscuity
- Withdrawal or decreased interest in activities of daily living
- Decline in function at work and home

Emotional Symptoms Include:

- Unaware of the changes in behavior
- Apathy
- Abrupt mood changes
- Easily distracted; poor attention span
- Rudeness, impatience, or aggression
- Lack of warmth, concern, or empathy

Language Problems Include:

- Complete loss of speech
- Difficulty speaking or understanding speech
- Weak, uncoordinated speech sounds
- Repeating words others say

- Decrease in ability to read or write
- Trouble finding the right word
- Loss of vocabulary

Physical Symptoms Include:

- Memory loss
- Difficulty moving about
- Lack of coordination
- Increased muscle rigidity or stiffness
- Urinary incontinence
- General weakness

Pick's disease generally occurs between the ages of forty and sixty years of age.

Pick's disease affects slightly more women than men.

Diagnosis of Pick's Disease:

The diagnosis from a doctor can help to rule out other diseases or conditions which can be treatable or cured. A psychologist can help establish the type of dementia.

Distinguishing Feature to Diagnose:

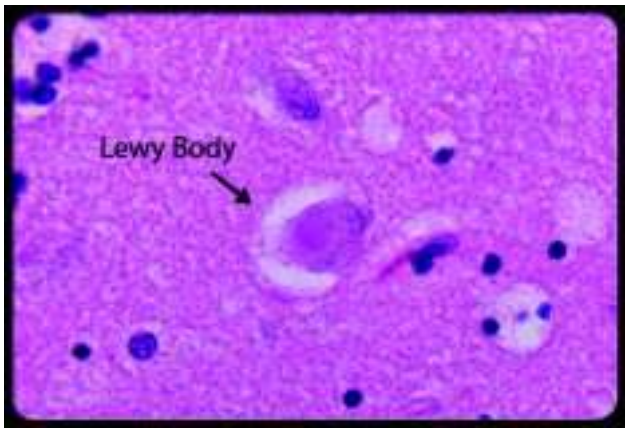
There are a few distinguishing feature that Pick's disease has which distinguishes it from other disorders: (i) Onset before 65 years of age, (ii) Lack of inhibition, (iii) Initial personality changes (iv) Loss of normal controls for example: hyper sexuality (v) Roaming behaviour.

Prognosis in Pick's Disease:

The length of progression varies in individuals from less than 2 years to 10 years in some.

LEWY BODY DISEASE:

Lewy Body Disease is a dementia that is clinically similar to Alzheimer's disease and often characterized by hallucinations, parkinsonian features and extra pyramidal signs. These degenerative brain diseases are linked to abnormal clumps of a protein called alpha-synuclein. These clumps are called as Lewy bodies, which are found in nerve cells throughout the outer layer of the brain (the cerebral cortex) and deep inside the midbrain and brainstem. These patients usually experience progressive cognitive decline, although there is considerable variability between the symptoms. The common symptoms comprise of problems with movement, visual hallucinations, and fluctuations in thinking skills or attention.



Lewy Bodies found in the midbrain and brain stem

Causes:

The causes of Lewy body dementia, is unknown in majority of cases. Monogentic forms of Lewy body disorders, where a patient inherited the disease from one parent, are rare and comprise about 10% of cases. These familial variants are more common in patients with an earlier age of disease onset. The majority of cases are thought to result from complex interactions among different "susceptibility" genes and environmental risk factors. The primary environmental factors that have been implicated in increasing risk of some cases of Lewy body disorders include pesticide or other chemical exposure and head injury. There are undergoing researches to study the role that genetics and environment play in Lewy body disorders.

The underlying brain changes of Lewy body dementias involve the clumping of a protein in the brain called alpha-synuclein. These clumps are called "Lewy bodies." The regional distribution of these clumps in the brain influences the types of symptoms (e.g., cognitive, motor, psychiatric). Lewy body dementias are also associated with reductions in important brain chemicals called neurotransmitters, including dopamine and acetylcholine.

Diagnosis:

An extensive neurological and neuropsychological evaluation is essential for Lewy body dementia diagnosis. Structural brain imaging for example MRI Brain and CT scan and laboratory tests are used to rule out other diagnoses. Lewy body dementias can be difficult to diagnose because they can resemble and overlap with other causes of dementia like Alzheimer's disease, other parkinsonian syndromes and vascular dementia.

As in many neurodegenerative diseases, a definitive diagnosis is only available after an autopsy.

Differential Diagnosis:

Parkinson's disease versus Lewy Bodies: A few patients with Parkinson's disease (PD) experience a subtle cognitive decline and their primary limitation is their motor disorder. Although patients with, Parkinson's disease will develop dementia as a consequence of the disease. When dementia develops after an established motor disorder, we call the disease PDD. In contrast, when the dementia develops prior to or at the same time as the motor disorder, we call the disease DLB. Although the initial sequence of symptoms differs in PDD and DLB, as the disorders progress, the symptoms and the underlying brain changes are much more similar than they are different. As such, many researchers and clinicians think of PDD and DLB as being on a continuum of a similar disease process rather than as two distinct entities.

Signs and Symptoms:

The parkinsonian motor syndrome is similar in PDD and DLB and can include slowed movement (bradykinesia), rigidity (muscles feel still and resist movement), tremor and gait difficulties. In terms of the cognitive difficulties, both disorders can involve progressive problems involving visual spatial processing, attention, executive dysfunction (e.g., planning and multi-tasking) and memory. Psychiatric symptoms are common and can include anxiety, depression, hallucinations (usually visual) and delusions (false beliefs). Sleep problems including excessive daytime drowsiness and difficulty staying asleep are common. Many patients exhibit REM sleep behavior disorder (RBD) that manifests by the patient acting out his or her dreams.

Many patients experience constipation, repeated falls, syncope and loss of smell. Individuals diagnosed with Lewy body dementias often have adverse reactions including confusion when taking medications that affect the brain, such as anti-anxiety drugs (examples: Valium®, Ativan®), anticholinergic drugs (examples: Benadryl, Detrol), and antiparkinson's drugs (examples: Sinemet®, Mirapex). To avoid adverse reactions to medications, physicians should carefully monitor medications, introduce medications one at a time and prescribe minimal doses when possible.

Disease Progression:

The onset of the disorder usually occurs in geriatric adults, and the disease risk increases with age. There is substantial unpredictability in the course of the illness. In general, symptoms progress over several years as cognitive decline becomes more prominent and psychiatric symptoms emerge or worsen. The progression of symptoms represents increasing pathology in the brain including more widespread Lewy bodies, loss of neurotransmitters (examples: dopamine and acetylcholine) and nerve cell death.

Prognosis:

The prognosis for Lewy Body Dementia (LBD) can be highly variable. There are no known therapies to stop or slow the progression of. LDB. The average life expectancy following the diagnosis is between five and seven years.

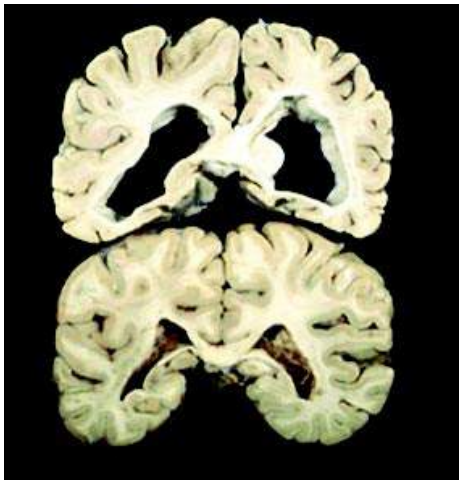
HUNTINGTON'S DISEASE:

Huntington's disease is classically associated with the development of dementia. The dementia seen in this disease is the subcortical type of dementia, characterized by more motor abnormalities and fewer language abnormalities than in the cortical type of dementia. Grossly, the cortical gyri appears normal to slightly atrophic. Coronal sections reveal striking caudate greater than putamen and pallidum atrophy. Milder degenerative changes occur in the neocortex and thalamus. Evidences exist for excitotoxic damage and mitochondrial DNA mutations and oxidatives stress in the pathogenesis of cortical pathology in hungtington's disease.

Clinical Feature and Associated Disorder:

Lieberman and associates [7] reported that around 90 percent of patients were demented by a mean age of 48.3 years and that dementias preceded the onset of chorea in 24 percent. Whether or not it is the critical determinant of cognitive decline, and cognitive decline correlates to pathologically and radiologically with degree of caudate atrophy and hypometabolism [8]. The area of particular interest in dementia of HD are frontal lobe relate or frontostriatal - related neuropsychiatric deficits, declarative memory impairment, and motor learning difficulties. Regarding cognitive disturbances, prospective study of cognitive decline in early cases has shown that psychomotor slowing is a characteristic early feature and memory

impairment although present early on does not deteriorate rapidly, or correlate as well with caudate atrophy, as does psychomotor speed. Huntington's disease causes memory loss but it appears to be less severe as compared with that of Alzheimer's disease at a matched stage of severity, although this factor has been more consistent at milder stages of dementias. Most studies have found that patients with HD have greater difficulty learning a task, but benefit more by cued recall or recognition as compared with free or unassisted recall. This has been thought to reflect greater impairment of retrieval than storage mechanisms and it affects remote memory retrieval as well as more recently learned material. There are many similarities in memory disturbances between HD and PD, suggesting similarities between subcortial dementia illnesses but overlap is imperfect. Evidence shows that the basal ganglias in patients with Huntington 's disease show a decrease in activity of the mitochondrial pathway, complex II-III. Such deficiencies are often associated with basal ganglia degeneration [9]. This degeneration of striatal neurons projecting to GPe leads to disinhibition of the indirect pathway, increased inhibition of STN, and therefore, reduced output of the basal ganglia [10].



The impact of Hungtion Disease on the brain structure in the basal ganglia

Differential Diagnosis:

In absence of chorea, the family history is critical in reaching a diagnosis. In the absence of both chorea, the family history is critical in reaching a diagnosis. In the absence of chorea and family history, the differential diagnosis is that of other subcortical dementias and chronic progressive encephalopathies.

Evaluation:

The clinical examination may provide compelling evidence for HD, which can be confirmed genetically. Ancillary tests include MRI and CT to ascertain caudate atrophy and SPECT or PET to ascertain caudate hypoperfusion or metabolism. Neuropsychological assessment is useful to document the nature and severity of cognitive decline, particularly in mild to moderate stages of illness.

Prognosis and Future Perspectives:

Death occurs within 10 to 20 years after onset, although suicide is more prevalent in at risk and early onset HD patients.

HIV - RELATED DEMENTIA:

Encephalopathy in HIV infection is associated with dementia and is termed acquired immune deficiency syndrome (AIDS) dementia complex or HIV dementia. The HIV virus will attack the nerve cells in the brain, which will lead to AIDS Dementia. So a person who carries the HIV virus may contract AIDS Dementia. As said earlier even comparatively healthy people with HIV positive status can fall prey to it. At times, the immune system has to grow very weak for the symptoms to surface properly. Patients infected with HIV experience dementia at an annual rate of approximately 14 percent. An estimated 75 percent of patients with AIDS have involvement of the central nervous system (CNS) at the time of autopsy. The development of dementia in people infected with HIV is often paralleled by the appearance of parenchymal abnormalities in MRI scans. Other infectious dementias are caused by Cryptococcus or Treponema palladium.

Symptoms of HIV Dementia: lack of concentration, withdrawal, forgetfulness, long term and short term loss of memory, mania, altered personality, irritability, poor coordination, impaired judgment, blurred or hazy vision, reduction in attention span, apathy and loss of bowel and bladder control.

Diagnosis:

The diagnosis is done on the basis of neurological brain scans, coordination tests and mental activities.

Head Trauma - Related Dementias:

Dementia can be a sequela of head trauma. The so -called punch- drunk syndrome occurs in boxers after repeated head trauma over many years. IT is

characterized by emotional liability, dysartharia and impulsivity.

Dementia Due to Other General Medical Conditions:

The DSM IV TR lists six specific causes of dementia that can be coded directly: HIV disease, head trauma, Parkinson's disease, Huntington's disease, picks disease and Creutzfeldt - Jakob disease. A seventh category allows clinicians to specify other nonpsychiatric medical conditions associated with dementia.

Substance induced persisting dementia: alcohol-related dementia has been estimated to account for, approximately 4% of dementias.

DIAGNOSIS AND CLINICAL FEATURES:

The dementia diagnosis in DSM-IV-TR dementia of the Alzheimer's type, vascular dementia, dementia due to other general medical condition, substance induced persisting dementia, dementia due to multiple etiologies, and dementia due to not otherwise specified. The most important step in making the diagnosis is to determine whether a chronic encephalopathy results from a degenerative or due to other potentially reversible causes. A reversible process includes fluctuating severity, altered level of consciousness or hypersomnolence and visual hallucinations.

The diagnosis of dementia is based on the clinical examination, including a mental status examination, and information from the patient's family, friends, and employers. Complaints of a personality change in patients older than age 40 suggest that the diagnosis of dementia should be carefully considered.

Clinicians should note patients complain is about intellectual impairment and forgetfulness as well as evidence of patients evasion, denial, or rationalization aimed at concealing cognitive deficits. Excessive orderliness, social withdrawal, or a tendency to relate event in minute detail can be characteristic, and sudden outburst of anger sarcasm can occur. Patients in appearance and behaviour should be observed. Liability of emotions, sloppy gloomy, uninhabited remarks, silly jokes, or a dull, apathetic, or vacuous facial expression and manners suggest the presence of dementia, especially when coupled with memory impairment.

Memory impairment is typically an early and prominent feature in dementia, especially in dementia involving the contacts, such as dementia of the Alzheimer's type. Early in the course of dementia, memory impairment is mild and usually most marked for recently went; people Forget telephone numbers, conversation, and events of the day. As the course of dementia progress is, memory impairment becomes severe, and only the earliest learned information example of a person's place of birth is retained. In as much as memory is important for orientation two per cent, place, and time, orientation can be progressively effected during the course of a dementing illness. For example patients with dementia may forget how to get back to their room after going to the bathroom. No matter how severe the disorientation seems, however, patients show no impairment in their level of consciousness.

Dementing processes that affect the cortex, primarily dementia off the Alzheimer style and vascular dementia, can affect patients language abilities. DSM-IV-TR includes aphasia as one of the diagnostic criteria. The language difficulties may be characterized by awake, stereo type, imprecise, circumstantial locution and patient may also have difficulty naming objects.

PSYCHIATRIC AND NEUROLOGICAL CHANGES:

Personality:

Changes in the personality of a person with dementia are especially disturbing for their families. Preexisting personality traits may be accentuated during the development of a dementia. Patients with dementia may also become introverted and seem to be less concerned than they previously were about the effects of their behaviour on others. Persons with dementia who have paranoid delusions are generally hostile to family members and caretakers. Patients with frontal and temporal involvement are likely to have marked personality changes and may be irritable and explosive.

Hallucinations and Delusions:

An estimated 20 to 30% of patients with dementia have had hallucination and 30 to 40% have delusions, primarily of paranoid or persecutory and unsystematized nature, although complex sustained and well systematised illusions are also reported by these patients. Physically aggression and other forms of violence are common in

demented patients who also have psychotic symptoms.

Mood:

In addition, to psychosis and personality changes, depression and anxiety major symptoms in an estimated 40 to 50% of patients with dementia, although the full syndrome of depressive disorder may be present in only 10 to 20 per cent. Patients with dementia also may exhibit pathological laughter or crying that it is, extremes of emotions with no apparent provocation.

Cognitive changes:

In addition to appraise in patients with dementia, apraxias and agnosias are common, and they are included as potential diagnostic criteria in DSM IV TR. Other neurological signs that can be associated with dementia seizures, seen in approximately 10% of patients with dementia of the Alzheimer style and in 20% of patients with vascular dementia and a typical neurological presentation, such as nondominant parietal lobe syndrome. Primitive reflexes, such as the grasp, snout, suck, tonic foot and palmomental reflexes may be present on neurological examination and myoclonic jerks are present in five to 10 percent of the patients.

Patients with vascular dementia may have additional neurological symptoms, such as headaches, dizziness, faintness, weakness, focal neurological signs, and sleep disturbances, possibly attributable to the location of the terrible or vascular disease.

Behavioural symptoms of dementia:

Patients with dementia may experience behavioural and psychological symptoms and during the course of their illness. These may include:

Behavioural Symptoms: screaming, restlessness, physical aggression, agitation, shaking, screaming, wandering, culturally inappropriate behaviours, sexual disinhibition, hoarding, cursing, and shadowing.

Psychological Symptoms: depression, delusions, hallucinations, delirium and apathy.

Causes of Behavioural Symptoms:

Biological:

- Genetic abnormalities related to the structure of particular parts of the brain.
- Changes of chemicals in the brain.
- Changes in the structure of different parts of the brain due to dementia.

Medical:

- Patients with dementia who also suffer from infections, pain, dental problems and arthritis may be unable to articulate the pain they are experiencing and instead may express their distress by vocalising or becoming aggressive.
- Co-morbid conditions such as delirium, depression, anxiety or psychosis. Individuals with dementia are more susceptible to delirium and the illnesses / environmental stressors that lead to delirium.
    - o Side effects caused due to medication can be numerous and may have a significant effect on a person's behaviour
    - o Hearing or vision impairment that is not well managed (for example, malfunctioning hearing aids may lead to frustration and changed behaviours).
    - o Sleep disturbances are common in people with dementia and can cause agitation and restlessness during the day and night.

Environmental and social:

- o Changes in social routine (for example, alteration in meal times or introduction of a new care routine can cause confusion and a feeling of loss of control for the person with dementia, contributing to behaviours of concern).
- o Change in environment (for example, relocation to a new room or home can increase agitation and disorientation).

Psychosocial Determinants:

The severity and course of dementia can be affected by psychosocial factors. The greater a person's premorbid intelligence and education, the better the ability to compensate for intellectual deficits. People who have a rapid onset of dementia use fewer defenses than do those who experience an insidious onset. Anxiety and depression can intensify and aggravate the symptoms. Pseudodementia occurs in depressed people who complain of impaired memory, but in fact are suffering from a depressive disorder. When the depression is treated, the cognitive defects disappear.

Catastrophic reaction:

Patient with dementia also exhibit a reduced ability to apply what Kurt Goldstein called the abstract attitude. Patients have difficulty generalizing from a single instance, forming concepts and grasping similarities and differences are among concepts. Furthermore, the ability to solve problems, to reason logically and to make sound judgement is compromised. Goldestine also described a catastrophic reaction marked by agitation secondary to the subjective awareness of the intellectual depth received under stressful circumstances. Persons usually attempt to compensate for defects by using strategies to avoid demonstrating failures in intellectual performance; they may change the subject, make jokes or otherwise divert the interviewer.

Sundrowner Syndrome:

Sundrowners Syndrome is characterised by drowsiness, conclusion, ataxia, and accidental faults. It occurs in older patients who are overly sedated and inpatients with dementia that reacts adversely to even a small dose of psychoactive drug. The syndrome also occurs in demented patients when external stimuli, such as light and interpersonal orientation is cues are diminished.

GENERAL PATHOLOGY, PHYSICAL FINDINGS AND LABORATORY EXAMINATION:

A comprehensive laboratory workup must be performed when evaluating a patient with dementia. The purposes of the workup are to detect reversible causes of dementia and to provide the patient and family with a definitive diagnosis. The range of possible causes of dementia mandate selected use of laboratory test. The evaluation should follow clinical suspicion, based on the history and physical and mental status examination results. The continued improvements in brain imaging techniques, particularly MRI, have made differentiation between the dementia of the Alzheimer type, and vascular dementia, in some cases somewhat more straightforward then in the past. And active area of research is the use of single photon emission computed tomography (SPECT) to detect patterns of brain metabolism in various types of dementia; the use or off (SPECT) images may soon help in the clinical differential diagnosis of dementing illnesses.

A reasonably recent complete physical evaluation should be obtained in patients who are suspected of suffering from dementia. It may reveal evidence of systematic disease causing brain dysfunction, such as an enlarged liver, and hepatic encephalopathy, or it may demonstrate systematic disease related to particular CNS processes. Patient should also undergo structural neuroimaging with computed tomography (CT) or magnetic resonance imaging (MRI) of the brain. When a nondegenerative based chronic progressive encephalopathy is suspected, electroencephalography (EEG) may be helpful in demonstrating severe dysrhythmic slowing. Cerebrospinal fluid (CSF) examination may show evidence of a chronic meningoencephalitic process such as an elevated IgG index and synthesis rate and occasionally oligoclonal bands, suggesting an intrathecal inflammatory reaction. Ultimately is suspicion remains high based on noninvasive tests for nondegenerative causes, cerebral angiography and meningeal and brain biopsy can be considered, or in the absence of such invasive testing, an empirical therapeutic trial of prednisone could be considered for a possible steroid-responsive inflammatory meningoencephalitis. Focal neurological findings, such asymmetrical hyperreflexia or weaknesses are seen more often in vascular then in degenerative diseases.

DIFFERENTIAL DIAGNOSIS:

Dementia of the Alzheimer type versus Vascular Dementia:

Classically, vascular dementia has been distinguished from dementia of the Alzheimer type by the decremental deterioration that can accompany cerebrovascular disease overtime. Although the discrete, stepwise deterioration may not be apparent in all cases, focal neurological symptoms are more common in vascular dementia of the Alzheimer's type, as are the standard risk factors for cerebrovascular disease.

Vascular Dementia versus Transient Ischemic Attacks:

Transient Ischemic Attacks (TIA's) are brief episodes of focal neurological dysfunction lasting less than 24 hours. Although a variety of mechanisms may be responsible, the episodes are frequently the result of microembolization from a proximal intracranial arterial lesion that produces transient brain ischemia, and the episodes usually

resolve without significant pathological alteration of the parenchymal tissues. Approximately one third of the person's with untreated TIA's experience a brain infarction later; therefore, recognition of TIA's is an important clinical strategy to prevent brain infarction.

Clinicians should distinguishing episodes involving the vertebrobasilar system from those involving the carotid arterial system. In general, symptoms of vertebrobasilar disease reflect a transient functional disturbance in either the brainstem or the occipital lobe; carotid distribution symptoms reflect unilateral retinal or hemispheric abnormality. Anticoagulant therapy, antiplatelet agglutinating drugs such as aspirin, and extracranial and intra cranial reconstructive vascular surgery are effective in reducing the risk of infarction in patients with TIA's.

Delirium:

In general, delirium is distinguished by rapid onset, brief duration, cognitive impairment fluctuation during the course of the day, nocturnal exacerbation of symptoms, marked disturbance of the sleep - wake cycle, and prominent disturbance in attention and perception.

Depression:

Some patients with depression have symptoms of the cognitive impairment difficult to distinguish from symptoms of dementia. The clinical picture is sometimes referred to as pseudodementia, although the term depression- related cognitive dysfunction is preferable and more descriptive. Patients with depression - related cognitive dysfunction generally have prominent depressive symptoms, more insight into their symptoms than do demented patients, and often a history of depressive episodes.

Factious Disorder:

Person who attempts to stimulate memory loss, as in factitious disorder, do so in an erratic and inconsistent manner. In true dementia, memory for time and place is lost before memory for persons, and recent memory is lost before remote memory.

Schizophrenia:

Although schizophrenia can be associated with some acquired intellectual impairment, its symptoms are much less severe than are the related symptoms of psychosis and thought disorder seen in dementia.

Normal Aging:

Aging is not necessarily associated with any significant cognitive decline, but minor memory problems can occur as a normal part of aging. These normal occurrences are sometimes, referred to as benign senescent forgetfulness or age - associated memory impairment. They are distinguished from dementia by their minor severity and because they do not interfere significantly with a person's social or occupational behaviour.

Other Disorders:

Mental retardation, which does not include memory impairment, occurs in childhood. Amnestic disorder is characterized by circumscribes loss of memory and no deterioration. Major depression in which memory is impaired responds to antidepressant medication. Malingering and pituitary disorder must be ruled out, but they are unlikely.

COURSE AND PROGNOSIS:

The classic course of dementia is an onset in the patients 50s or 60s with gradual deterioration over 5 to 10 years, leading eventually to death. The age of onset and the rapidly of deterioration vary among different types of dementia and within individual diagnostic categories. The average survival expectation for patients with dementia of the Alzheimer's type is approximately 8 years, with a range of 1 to 20 years. Data suggest that in persons with an early onset of dementia or with a family history of dementia the disease is likely to have a Rapid course.

The most common course of dementia begins with a number of subtle signs that may, at first, be ignored by both the patient and the people closest to the patient. A gradual onset of symptoms is most commonly associated with dementia of the Alzheimer's type, vascular dementia, endrinopathies, brain tumors and metabolic disorders. Conversely, the onset of dementia resulting from head trauma, cardiac arrest with cerebral hypoxia, or encephalitis can be sudden. Although the symptoms of the early phase of dementia are subtle, they become conspicuous the as the dementia progresses, and family members may then bring a patient to a physician's attention. People with dementia may be sensitive to use of benzodiazepines or alcohol, which can be precipitate agitated, aggressive or psychotic behaviour. In the terminal stage of dementia,

patients become empty shells of their former selves-profoundly disoriented, incoherent, amnestic and incontinent of urine and feces.

With psychosocial and pharmacological treatment possibly because of the self- healing properties of the brain, the symptoms of dementia may progress slowly for a time or may even recede somewhat. Symptoms regression is certainly a possibility in reversible dementias once treatment is initiated. The course of the dementia varies from steady progression to an incrementally worsening dementia to a stable dementia.

Stages of Dementia:

1-Early stage (developed in 1-2 years):

The early stage of dementia is often overlooked because the onset of dementia is gradual; it is often difficult to be sure exactly when it begins. The person may for example:

- Individual may have problems talking properly (language problems).
- Individual may have significant memory loss - particularly for things that have just happened.
- The individual may not be oriented.

2-Middle stage (developed in second to fifth year):

As the disease deteriorates gradually, limitations become clearer and more restricting. The person with dementia has difficulty with day-to-day living and:

- May become very forgetful - especially of recent events and people's names.
- Can no longer manage to live alone without problems.
- Is unable to cook, clean or shop.

3-Late stage (developed in fifth year or after):

This stage is one of near total dependence and inactivity. Memory disturbances are serious and the physical side of the disease becomes more obvious. The person may:

- Have difficulty eating.
- Be incapable of communicating.
- Not recognize relatives, friends and familiar objects.
- Display inappropriate behaviour in public.
- Be confined to a wheel chair or bed.

Morbidity and Mortality associated with Dementia Global Morbidity and Mortality:

- In 2000, age-standardized dementia mortality rate was 6.7 and 7.7 for 100,000 male and female respectively. [10]
- 24.3 million have dementia [11]
- 4.6 million new cases per year [11]
- Worldwide dementia contributes 4.1% of all disability-adjusted life years (DALYs) and 11.3% of years lived with disability and 0.9% of years of life lost.

India Morbidity and Mortality:

- Age standardized death rate of 12.1 per 100,000 reference number?
- 1.8 million have dementia in India and South Asia in people >60 years of age. [11]
- 400,000 new cases per year for India + South Asia.[11]
- 1,034 per 100,000 DALYs.

TREATMENT FOR DEMENTIA:

PHARMACOTHERAPY:

Clinicians may prescribe benzodiazepines for insomnia and anxiety, antidepressant for depression and antipsychotic drugs for delusions and hallucinations, but they should be aware of possible distinctive drug effects in older people. In general, drugs with high anticholinergic activity should be avoided.

Donepezil, rivastigmine, galantamine and tacrine and cholinesterase inhibitors used to treat mild to moderate cognitive impairment in Alzheimer's disease. They reduce the inactivation of the neurotransmitter acetylcholine and thus potentiate the cholinergic neurotransmitter, which in turn produces a modest improvement in memory and goal-directed thought. These drugs are most useful for persons with mild to moderate memory loss who have sufficient preservation of their basal forebrain cholinergic neurons to benefit from augmentation of cholinergic neurotransmission.

Donepezil: AriceptTM is the trade name for donepezil. It is a cholinesterase inhibitor, making it a drug from the main class of compounds now

used to treat people with Alzheimer's disease. AriceptTM has been used in a large number of clinical trials, including some from Canada that have used a variety of individualized symptom-based tests of its effectiveness. In 2006, the US Food and Drug Administration approved AriceptTM as a treatment for severe dementia associated with Alzheimer's disease.

Dosage: This is taken once a day, with food. The starting dose is 5 mg / day. After a month on the drug, if it is well tolerated, most patients will be increased to the 10 mg / day dose. Usually, at least two months more are needed to know whether the drug is meeting the goals of treatment.

Rivastigmine: ExelonTM and Exelon TMPatch are the trade names for rivastigmine. It is a cholinesterase inhibitor for the treatment of Alzheimer's disease. It is also used to treat people with Lewy body dementia, and with the dementia of Parkinson's disease.

Dosage: The starting dose is 1.5 mg, twice a day. Typically, after one month, this is increased to 3.0 mg, twice a day, which is often the lowest effective dose. Patients who tolerate that dose can have it increased to a dose of 4.5 mg twice a day and later to a dose of 6.0 mg twice a day. These are the recommended doses.

Exelon TMPatch is now available in some countries, including the United States and Canada. It is applied to the back, chest or upper arm and provides continuous delivery of medication through the skin over 24 hours. Treatment is initiated with Exelon TMPatch 5 (9mg / 5cm2) with a release rate of 4.6mg over a 24 hour period to a maximum dosage of Exelon TMPatch 10 (18mg / 10cm2) with a release rate of 9.5mg over a 24 hour period.

Galantamine: Reminyl ER is the trade name for galantamine. It is a cholinesterase inhibitor, making it a drug from the main class of compounds now used to treat people with Alzheimer's disease.. In addition to acting like the other cholinesterase inhibitors, it also directly stimulates a class of receptors to which acetylcholine binds.

Dosage: Briefly, galantamine (extended release) is taken once a day, with food. The starting dose is 8 mg / day. After a month on the drug, if it is well tolerated, most patients will be increased to the 16 mg / day dose, and there is also a 24 mg / day dose. Usually, at least two months after starting at the 16 mg / day dose are needed to know whether the drug is meeting the goals of treatment.

Memantine: Memantine goes by several trade names, or brand names, including EbixaTM, NamendaTM (in the United States) and AxuraTM. It is an NMDA, or N-Methyl-D-aspartic acid. (N-methyl-D-aspartate) receptor antagonist, making it distinct from cholinesterase inhibitors, the other class of compounds commonly used to treat people with Alzheimer's disease. The reason to block the NMDA receptor is to block transmission of a brain chemical called glutamate. Glutamate is a neurotransmitter, which is used widely throughout the brain, but too much glutamate is felt to cause calcium overload in brain cells. The theory is called Glutamatergic Excitotoxicity.

Dosage: Briefly, memantine is taken twice a day, with food. The starting dose is 5 mg / day. After a week, this is increased to 5 mg, twice a day. If it is well tolerated, most patients will be increased to the 10 mg / once a day and 5 mg once a day, after which the dose increases to 10 mg twice a day. Usually, after starting at the 10 mg twice a day dose, at least two months are needed to know whether the drug is meeting the goals of treatment.

Donepezil is well tolerated and widely used. Tacrine is rarely used, because of its potential for hepatotoxicity. Fewer clinical data are available for rivastigmine and galantamine, which appears more likely to cause gastrointestinal and neuropsychiatric adverse effects than donepezil. None of these medications prevents the progressive neuronal degeneration of the disorder.

Other Treatment Approaches: Other drugs being tested for cognitive-enhancing activity include general cerebral metabolic enhancers, calcium channel inhibitors, and serotonergic agents. Some studies have shown that selegiline, a selective type B monoamine oxidase inhibitor, may slow the advance of this disease. Ondansetron, a 5 HT 3 receptor antagonist, is under investigation.

Estrogen replacement therapy: Estrogen replacement therapy may reduce the risk of cognitive decline in postmenopausal women; however, more studies are needed to confirm this effect. Complementary and alternative medicine studies of ginkgo biloba and other phytomedicinals to see if they have a positive effect on cognition. Reports have appeared of patients using nonsteroidal anti-inflammatory agents having a lower risk of developing Alzheimer's disease. Vitamin E has not been shown to be of value in preventing the disease.

PSYCHOLOGICAL INTERVENTION:

Psychological Intervention is very often required in geriatrics but for individuals who are suffering from serious disorders it is very important that they undergo psychological treatment. Psychological treatment very often involves working in an orchestrated way with several professionals.

Intervention Plan:

- Early testing and diagnosis
- Optimization of physical health, cognition, activity and overall well being.
- Detection and treatment of Behavioural and Psychological Symptoms of Dementia.
- Educating carer and providing long term support to carer.

PSYCHOLOGICAL ASSESSMENT FOR DEMENTIA:[12]

1. ADAS- Cog: This test is used to measure cognition which consists of 11 tasks measuring the disturbances of memory, language, praxis, attention and other cognitive abilities which are often referred to as the core symptoms of Alzheimer's disease. This is recommended for the second stage on Alzheimer's disease. The estimated evaluation time of this test is 30 to 45 minutes. This test is usually administered by a neuropsychological or a clinical psychologist. [13]
2. Quality of Life - Alzheimer's disease Scale (QOL - AD): This test is used to measure the quality of life. This scale consists of 13 domains to measure quality of life. [14]
3. DEMQOL: This test assesses the quality of life in persons with mild to moderate dementia. It takes approximately 10 to 20 minutes for assessment.
4. Modified Mini - Mental Status Examination (3MS): The Modified Mini - Mental Examination is a brief widely used test to measure the cognitive functions with good reliability and validity. The administration time taken is approximately 15 minutes. Qualified health-care professional (at least trained in the Mini Mental) interviews the patient using a standard set of questions. Scoring takes 5 min. [15]
5. Holden Communication Scale: This is a scale which is used to assess the communication changes in the patient. This scale is completed by staff or family caregivers and covers a range of social behaviour and communication variables. [16]
6. Cornell Scale for Depression in Dementia: This scale is used to assess depression. This scale consists of five broad categories using the information from interviewers, with staff and participants [17].
7. Rating Anxiety in Dementia Scale (RAID): This scale assesses anxiety in the individual suffering from dementia. This scale rates anxiety in four main categories and uses interviews with staff and participants. [18]
8. Neuropsychiatric Inventory (NPI): This test is used to assess behavioural disturbances in patients with dementia. To assess psychopathology in the person with dementia and to help distinguish between the different causes of dementia. Acute, Primary, Community and Residential Care. Includes symptoms known to be rare in Alzheimer's disease, but are characteristic of fronto-temporal dementias. It also assesses the level of caregiver distress engendered by each of the neuropsychiatric disorders. This test assesses 10 behavioural disturbances occurring in dementia patients. It takes around 10 to 20 minutes to administer. [19]
9. Short Form-12 Health Survey (SF-12): This scale is used to measure generic health concepts relevant across age, disease, and treatment groups. The SF-12 includes 8 concepts commonly represented in health surveys. It is a self administrative measure that provides a comprehensive, psychometrically sound, and efficient way to measure health from the patient's point of view by scoring standardized responses to standard questions. This measure is only used when a family caregiver is available from the community sample participants. [20]
10. Frontal Executive Function: The FAB is a brief tool that can be used at the bedside or in a clinic setting to assist in discriminating between dementias with a frontal dysexecutive phenotype and Dementia of Alzheimer's Type (DAT). The FAB has validity in distinguishing Fronto-temporal type dementia from DAT in mildly demented patients (MMSE > 24). Total

score is from a maximum of 18, higher scores indicating better performance. It takes approximately 1o minutes to administer and 5 minutes to score.

11. Behavioural Pathology in Alzheimer's Disease (BEHAVE-AD): The purpose of this test is to measure behavioural and psychological symptoms of dementia in persons with Alzheimer's disease. It takes approximately 20 minutes.

12. Clinical Dementia Rating (CDR): The purpose of this test is to assess the severity of cognitive and functional impairment in dementia. The severity of dementia is rated on the following severity scale:

    0 - No Impairment

    0.5 - Questionable Impairment

    1 - Mild Impairment

    2 - Moderate Impairment

    3 - Severe Impairment

    This is a very comprehensive test and takes approximately 40 to 75 minutes.

13. Clock Drawing Test:

Instructions for the Step 1: Give patient a sheet of paper with a large (relative to the size of handwritten numbers) pre drawn circle on it. Indicate the top of the page.

Step 2: Instruct patient to draw numbers in the circle to make the circle look like the face of a clock and then draw the hands of the clock to read "10 after 11."

Clock drawing Test

Scoring:

Score the clock based on the following six-point scoring system:

1- Perfect: No errors in the task

2- Minor visuospatial error

3- Inaccurate representation of 10 after 11 when visuospatial organization is perfect or shows only minor deviations.

4- Moderate visuospatial disorganization of times such that accurate denotation of 10 after 11 is impossible.

5- Severe level of disorganization as described in scoring of 4

6- No reasonable representation of a clock

The first step in intervention of dementia is verification of the diagnosis. Accurate diagnosis is imperative, for the progression may be halted or even reversed if appropriate therapy is provided. Preventive measures are important, particularly in vascular dementia. Such measure might include changes in diet, exercise and control of diabetes and hypertension. Pharmacological agents might include antihypertensive, anticogagulant or antiplatelet agents. Blood pressure control aim for the higher end of the normal range, because that has been demonstrated to improve cognitive function in patients with vascular dementia. Blood pressure below the normal range has been demonstrated to further impair cognitive functions in the patient with dementia. Surgical removal of carotid plaques may prevent subsequent vascular events in carefully selected patients. The general treatment approach to patients with dementia is to provide supportive medical care, emotional support for the patients and their families, and pharmacological treatment for specific symptoms, including disruptive behaviour.

Treatment for Behavioural Issues in Dementia:

Treatment strategies for behavioural problems in dementia include:

- Nonpharmacological Approach (e.g., behavior therapy, cognitive stimulation therapy, psychoeducation)
- Pharmacological Approach (e.g., antipsychotics, antidepressants, mood stabilizers, cognitive enhancers)

PHARMACOTHERAPY FOR BEHAVIOURAL AND PSYCHOLOGICAL PROBLEMS:

Pharmacotherapy should be initiated only if the patient's symptoms have not responded adequately to nonpharmacological interventions, if there is no underlying medical condition causing these symptoms, and/or if these symptoms are not related to a medication effect. Although these nonpharmacological and pharmacological treatments are effective in decreasing the burden of behavioural symptoms, they usually require sustained input from a multidisciplinary team and ongoing staff training to maintain superior quality of care for patients with behavioural symptoms.

Medications used to treat Behaviour and Psychological Symptoms of Dementia: These medications are prescribed by a psychiatrist after unsuccessful nonpharmacological interventions.

Class of Medication	Name of Medication	Dosage Range (mg/day)	Common Side Effects of Medication Class
Antipsychotics	Aripiprazole Haloperidol Risperidone Quetiapine Olanzapine	2.5-15 0.5-5 0.25-2 25-200 2.5-15	Sedation, extrapyramidal symptoms, neuroleptic malignant syndrome, metabolic syndrome, QTc prolongation
Antidepressants	Fluoxetine Citalopram Paroxetine Sertraline Trazodone	10-80 10-60 10-50 25-200 25-200	Sedation, extrapyramidal symptoms, neuroleptic malignant syndrome, metabolic syndrome, QTc prolongation
Mood stabilizers	Fluoxetine Citalopram Paroxetine Sertraline Trazodone	100-400 250-1000 300-600	Sedation, gait and balance issues, falls, liver dysfunction, hyperammonemia, thrombocytopenia Cognitive enhancers Donepezil Galantamine Rivastigmine Memantine 5-10 8-24 3-12 5-20 Sedation, gastrointestinal disturbance, confusion,
Cognitive enhancers	Donepezil Galantamine Rivastigmine Memantine	5-10 8-24 3-12 5-20	Sedation, gastrointestinal disturbance, confusion, Agitation

Current evidence indicates the efficacy of short-term pharmacological treatment for behavioural symptoms and psychological symptoms. Although limited, available evidence remains in favor of using atypical antipsychotic drugs, especially risperidone, aripiprazole, and, to a lesser extent, olanzapine in the first-line treatment of BPSD that is resistant to nonpharmacological interventions [21, 22] Evidence indicates that these antipsychotic medications generally should not be continued for longer than 12 weeks11-13, [22] however, longer-term therapy may be needed in patients who have persistent symptoms, although the data for longer-term treatment with antipsychotics are limited[23].The

possible benefits of prescribing these medications should always be weighed carefully against the risks. Careful monitoring and treatment of risk factors for cerebrovascular adverse events such as hypertension, atrial fibrillation, hyperlipidemia, and diabetes mellitus should help reduce not only the risk of cerebrovascular adverse events, but also the risk of death [23]. Antipsychotics should also be prescribed within the recommended dosage ranges to reduce the risk of serious adverse events such as cerebrovascular adverse events and death. The best pharmacological alternatives to antipsychotics in the treatment of BPSD are anticonvulsants, antidepressants, cholinesterase inhibitors, and memantine, although these drugs may have better efficacy in treating nonaggressive behaviours such as apathy than in treating agitation, aggression, and psychotic symptoms such as delusions and hallucinations. Despite the growing evidence for pharmacotherapy for BPSD, none of the aforementioned medication classes have been FDA-approved for these behaviors and symptoms. Information on common scales used to assess the severity of dementia is listed in table an algorithm for the treatment of BPSD.

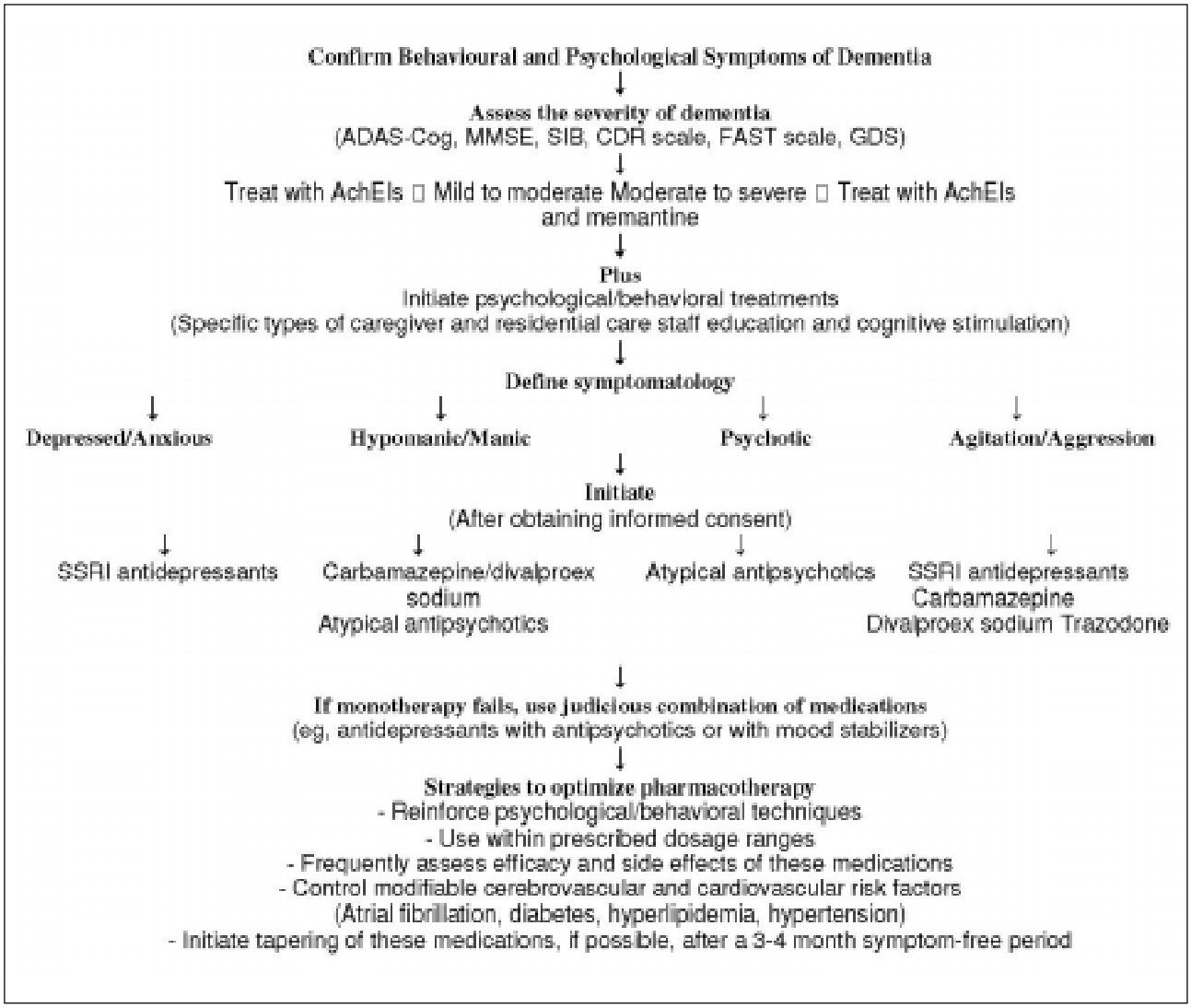


Figure. Algorithm for the treatment of BPSD.

Abbreviations: AchEIs, acetylcholinesterase inhibitors; ADAS-Cog, Alzheimer's Disease Assessment Scale-Cognitive; BPSD, behavioral and psychological

symptoms of dementia; CDR, Clinical Dementia Rating; FAST, Functional Assessment Staging; GDS, Global Deterioration Scale; MMSE, Mini-Mental State

Examination; SIB, Severe Impairment Battery; SSRI, selective serotonin reuptake inhibitor.

PSYCHOTHERAPY:

Psychotherapy for older people has flourished despite the now famous statement by Sigmund Freud that older people are not amenable to psychoanalysis. According to him anyone over the age of 50 years lacked "the elasticity of the mental processes on which treatment depended....old people are not educatable" [24].Eventually this view was not encouraged and now psychotherapy is offered to the older population. Psychotherapy is used to work on various psychiatric problems, emotional issues, and adjustment problems.

The deterioration of mental faculties has significant psychological meaning for patients with dementia. The experience of a sense of continuity over time depends on memory. Recent memory is lost before remote memory is most cases of dementia, and many patients are highly distresses by clearly recalling how they used to function while observing their obvious deterioration. At the most fundamental level, the self is a product of brain functioning. Patients identities begin to fade as the illness progresses, and they can recall less and less of their past. Emotional reactions ranging from depression to severe anxiety to a catastrophic terror can stem from the realization that the sense of self is disappearing.

Patients often benefit from a supportive and educational psychotherapy in which the nature and course of their illness are clearly explained. They may also benefit from assistance in grieving and accepting the extent of their disability and from attention to self- esteem issues. Any areas of intact functioning should be maximized by helping patients identify activities in which successful functioning is possible. A psychodynamic assessment of defective ego functions and cognitive limitations can also be useful. Clinicians can help patients find ways to deal with the defective ego functions, such as keeping calendars for orientation problems, making schedules to help structure activities and taking notes for memory problems.

Psychodynamic intervention with family members of patients with dementia may be of great assistance. Those who take care of patient struggle with feelings of guilt, anger and exhaustion as they watch a family member gradually deteriorate. A common problem that develops among caregivers involves their self-sacrifice in caring for patients. The gradual developing resentment from this self - sacrifice is often suppressed because of the guilt feelings it produces. Clinicians can help caregivers understand the complex mixture of feelings associated with seeing a loved one decline and can provide understanding as well as permission to express these feelings. Clinicians must also be aware of the caregiver's tendencies to blame themselves or others for patient's illness and must be to appreciate the role that patients with dementia play in the lives of family members.

Dynamics:

Psychotherapy is mostly based upon a dynamic view of psychiatric (and some physical) illnesses. However, the word dynamic suggests activity where psychiatric symptoms arise, conflict often leads to a deadlock. Such a conflict might be between the drive to assert oneself and to make no trouble. Dynamic factors are important in old age psychiatry. The main aim of psychotherapy is to resolve the conflict and, in doing so, reduce symptoms and help the person function more effectively. This also focuses on improving the patient's quality of life. This is achieved through words exchanged between the patient and the therapist, and the relationship which forms between them. Pitt (1982) [24] identified dynamic factors in three-quarters of consecutive referrals to an old age psychiatric service.

Transference:

The dynamic theory is of the outlook that disturbed relationships, past and present, contribute to illness. Transference takes place when the patient reacts to the therapist as a key figure from their past, such as a parent, sibling, child or spouse. The therapist can use transference by helping the patient become aware of subconscious feelings to these important figures. Although the patients are usually older than their therapists, this does not prevent them from being regarded as parent figures, as well as child (or grandchild) figures.

Dependency:

Dependency is commonly seen in later stages of life. This takes place in people who are emotionally deprived during the earlier days of life and who had good coping mechanisms during adulthood but in older age feared of being neglected and not loved as the see their useful role slipping away. Goldfarb (1965) [25] described the frantic search for help from a strong parent figure, which is frustrated because the demands are too clamorous. In partnership, i.e. between husband and wife or child

and parent, one of them is usually comes out to be stronger than the other but in fact they are dependent on each other. The 'strong' partner preserves an illusion of mastery through the wreaker's reliance on them. In these cases the therapist tries to establish a supportive relationship to meet the dependency needs of the patient, without being taken over, or turning the patient into a child. For some very old patients, supportive therapy may mean an involvement for life.

Confrontation:

Confrontation may play a very important part in therapy, in spite of the pleasant personal remarks and warmth that a client shares with the therapist. Negative reactions to the therapist such as plaintiveness; exaggeration of symptoms; seeking help elsewhere; or lateness for appointments, should be explored. Skilfully handled these confrontations allow the patient to acknowledge resentment and aggression, and find relief that therapy can continue.

Group therapy:

Group therapy is a good medium of meeting other patients with the same problem and working through the problems including dealing with the dependency issue. This form of therapy is very intense and than an individualized session and this is less likely to lead to regression. Group therapy often helps the older isolated patient to get back to socialization. Group therapy works as a stress buster and works as a ventilation for patients as patients are quite comfortable sharing their feelings and emotions to people with the same problem than someone normal who finds it difficult to understand their problem. Dobson & Culhane (1991) [26] describe a therapeutic group run for older women. They emphasise the importance of having a clear purpose for a group and considering selection criteria carefully. In the early stages, rules such as not talking while others do, and valuing others' contributions, helped to harness good intentions.

Family therapy:

As caregivers are present with the patient for a longer period of time especially of the patient's functioning has reduced then family therapy is extremely important to maintain the family dynamics or make it more fulfilling for the patient. Most of the times the caregivers are totally involved with the patients physical and emotional needs and due to which very often the caregivers might be at a fix between their needs and those of the patient. There might be other problems with communication, for example deafness or poor vision, which can compound the effects of ageing in reducing information processing capacity. The overall effect is to make demanding tasks such as therapy very difficult. Physical or mental illness can be used to scapegoat the older person. Conversely, symptoms of physical illness can be accentuated or become an important vehicle for the older person's status and power (West & Spinks, 1988). Benbow & Marriott (1997) [27] listed the following ideas as being useful in family therapy with older adults:

(a) The family life cycle: looking at how families evolve. Key issues in later family life include retirement and becoming a grandparent.

(b) Cross-generational interplay - life cycle changes in different generations may not 'fit'. One generation may be more family orientated (e.g. during childbirth) while others are more outward looking (e.g. early retirement). Expectations may vary across the generations.

(c) Genograms - drawing a family tree is a useful way of collecting, organising and considering family information.

(d) Circular questions - these are in terms of relationships. Examples include, "If your mother says this, what does your brother do?" or "who in the family would this affect?"

(e) Reflecting teams - members of the multi-disciplinary team talk about the family while they listen, offering different perspectives. The systemic approach derived from family therapy can also be applied to the care of older people in institutions [28] Beckenham: Croom Helm.. Sometimes the problems attributed to one or more residents are better addressed by looking at the social network and relationships in the home.

Marital therapy:

Marital therapy is extremely important as very often the one who cares the maximum for the patient and looks after him is his/her partner. Throughout the life the patient and the caregivers are struggling with various things like bringing up children, running the household, etc and after retirement when its now time for them to rest and relax, either of the partner suffers from dementia and this puts the other partner in a position to take complete responsibility of the partner. Illness in one

partner may result in guilt in the other. It would also lead to frustration and irritations but the caregiver would not be able to express it and would in return feel guilty about his / her thoughts.

Cognitive and behavioural therapies:

The most frequently reported developments have been with younger people (Williams, 1984), but there have been examples of applications to older people which have now been reviewed. There are three reasons why cognitive and behavioural therapies are particularly suited to the elderly:

(a) Depression in later life is prone to relapse and there is evidence that cognitive and behavioural approaches have longer-term benefits. Patients may develop preventative skills.

(b) Older people are more susceptible to adverse side-effects from drug therapy, especially those who are physically ill or taking medications for other illnesses.

(c) Spontaneous remission is less likely in older depressed people (Lambert, 1976; Thompson et al, 1987). Lambert, M. J. (1976) [28] Evidence suggests that depression often goes unrecognized or untreated when it occurs with physical illness (Koenig et al, 1992). Only 10% of their sample of depressed older men in hospital received some form of psychological therapy, and 44% received no treatment at all. They suggest that behavioural and cognitive techniques are a viable therapeutic option for the 50% of patients or more who have medical contraindications to antidepressants.

Anxiety management:

The prevalence of phobic disorders is higher than might be expected, with a one month prevalence of 10% (Lindesay, 1991) [29]. Disability from phobias can be significant but specific treatment is rare. The majority of these patients suffer from a late-onset agoraphobia following a traumatic experience, typically an episode of serious physical illness. They have a higher than average contact with primary care services but tend not to be referred for specialist treatment. While it may be necessary to adapt anxiety management for older people, it is just as effective as in younger patients. Older people may benefit from anxiety techniques such as relaxation therapy with tapes. By using headphones it is possible to deliver soothing instructions loudly, so overcoming all but the most severe hearing loss.

Grief Counseling:

Although bereavement is not exclusively a problem of later life it is more common, and can mean the loss of an intimate relationship which has lasted many years. One ageist stereotype is that older people are better at handling loss because it is expected, or because they have had more practice. In fact older people are not only more likely to experience loss, but the effects are cumulative in terms of risk for depression (Murphy, 1986) [30]. Being recently widowed is a major risk factor for mortality (Rees & Lutkins, 1967).

Losses can also reduce social support, either because friends and family have died, or because physical illness enforces isolation. Appropriate interventions include bereavement therapy and practical help to improve social networks. Important steps are recognizing that something is wrong and encouraging the older person to accept help.

Therapies for Dementia:

Reality Orientation:

Reality orientation is perhaps the best known specific psychological treatment for older people (Holden & Woods, 1988) [31]. Reality orientation aims to help patients with dementia by directly focusing on some of the deficits of the disorder, including disorientation and impaired short-term memory. It also helps to preserve skills. The approach can be divided into brief sessions (classroom reality orientation) and a pervasive approach influencing staff-patient interactions throughout the day (24-hour reality orientation). Underlying both of these types of approach is the principle that staff enhances orientation by using identifying names and other information. This is supported by cues such as the commonly used reality orientation board, showing the date, weather, etc. (providing it is kept up to date!) and cues to everyday behaviour such as making tea or visiting a pub. Activities within formal reality orientation sessions include prompting basic information such as the names of group members; looking at current events; and using tactile, olfactory and other stimuli to encourage active cognition.

The enthusiasm with which reality orientation has been embraced by staff has sometimes exceeded the evidence for its effectiveness (Powell- Procter & Miller, 1982). There is evidence that reality

orientation is beneficial, but the benefits tend to be modest, and only sustained with continued effort. There is little evidence to suggest that generalisation occurs (i.e. encouraging orientation for time does not lead to gains in orientation for place). There are also doubts about the suitability of reality orientation for use by informal carers at home.

Perhaps the biggest impact of reality orientation has been on staff attitudes, where it has resulted in staff improving the environments in which they work. When you orientate someone to their environment you become more aware of it yourself. There is little point in orientating someone to the day if all days are the same.

Reminiscence:

Thinking about the past is not exclusive to older people. Younger people reminisce, although their memories may not extend back so far. Similarly, older people are not always reminiscing. Some avoid doing so because it is painful, others because they prefer the present. Nevertheless for many older people looking back is an important part of making sense of themselves and their lives. It allows the kind of integration that Erikson (1959) suggests is important as a developmental task of later life. The ability to reminisce is preserved in early dementia. People with dementia are better at remembering what happened many years ago than what happened this week or earlier this morning. The act of reminiscence therefore offers a good way of engaging people with mild to moderate cognitive impairment without reminding them of their cognitive shortcomings. It is quite possible to use reality orientation and reminiscence in combination. Starting with reality orientation before progressing to reminiscence appears to offer better results (Baines et al, 1987) [32]. Whether the activity of reminiscence, per se, can be considered a 'therapy' is open to question. Certainly reminiscence can have powerful emotional consequences and may be used with specific therapeutic targets in mind. However, it is neither a panacea nor is it always appropriate for older people. The overlap between reminiscence and post-traumatic stress disorder requires more careful study. Certainly there are older people whose problem is not how to remember but rather how to forget memories from wartime or other past trauma. The effects of reminiscence, like reality orientation, are dependent on the environment. There is a more obvious effect in a less interactive environment (Head et al, 1990).

Validation therapy:

Concerns about the specific effects of reality orientation in confronting dementia sufferers with their failings have crystallised around the development of an alternative therapy which is directed at emotional needs. There can, not surprisingly, be problems in reorientating people when the reality is upsetting. A common example given is of a disoriented person who wants to go home to his or her spouse, not remembering that they have died. Is reorientation the best solution in this situation? Validation therapy focuses on the phenomenology of dementia at an emotional, rather than a factual level. It views the disoriented person as struggling to cope with a complex and confusing world. It is hypothesised that the content of 'confused' talk reflects the emotional meaning of past events. For example, worrying about getting home in time to meet the children may reflect that parenting was a time of reward and security.

The response to the disorientation is directed at exploring what things were like for that person, and how this relates to how they are feeling now. It is suggested that even the most confused behaviour has some meaning for that person. Validation therapy has begun to be reported but more investigation is needed (Jones, 1997) [33]. It remains to be demonstrated how validation therapy and reality orientation differ. Of course bad reality orientation can certainly be frustrating and even unkind. However, closer scrutiny of the practice of experienced therapists suggests that they are sensitive to the emotional content of 'confusion', and avoid inappropriate confrontation. We need to find the best responses to people with dementia.

Memory therapy:

The emergence of specific approaches aimed at helping those with memory problems would seem to be of particular interest to those treating the elderly. On the whole, however, the work has been directed at younger brain damaged patients. This is because the conditions for memory therapy are not met so well in dementia. In particular the use of mnemonics or memory aids require insight into memory loss, as well as preserved language and psychomotor skills. Older people with mild memory problems can make use of simple aids such as lists, alarms and placing notices and instructions in key areas. A useful list of such aids is given by Burnside (1988) [34]. However, these are of major significance in only a few cases. These tend to involve people with mild or relatively static

Colour Recognition

Problem solving with colours and numbers

Problem Solving with colours

Recognition of various denominations

memory problems, such as following a stroke. For those with progressive dementia prompts supplement, rather than replace, the presence of an alert carer.

Resolution therapy:

Resolution therapy has been introduced as a companion to reality orientation (Stokes & Goudie, 1990) [35]. It shares with validation therapy the assumption that there is meaning in the behaviour and confused talk of patients with dementia. But, unlike validation therapy, it looks for that meaning in the 'here and now'. In other words it sees such behaviour or speech, as an attempt to make sense of what is happening now, or to communicate a current need. In order to try and understand these hidden meanings, the therapist must use reflective listening, exploration, warmth and acceptance.

Psychological Home Care Strategies:

- Prepare a meaningful time table for the patient i.e. brings activities that are of interest to the person keeping in mind their level of ability. Also this will help the patient practice and remember his daily routine sequentially.
- Historic snapshots: Gather as much as information about the patient and his past experiences and memory. For example: routines, eating patterns, personality traits, relationships with family friends, hobbies, etc. This would help the client remember things from the past and would help the family members compare the past and present state of the patient.
- Interview the patient Alone: This will help to know the patients personality, the way he responds to situations, his social and family structure and way of functioning.
- Attune the environment - Wherever possible interview the person with dementia in his / her own home or room. Familiarity is imperative to a person with dementia feeling safe and comfortable. Be sure that the environment is free from noise, interruptions and distractions; i.e., is quiet, pleasant and calm.

Communication Strategies for Psychologists: Listening to a patient with Dementia:

- Communicate interest and respect to patients with dementia by maintaining eye contact and posing a relaxed body language.
- Facial expressions and gestures communicate about the person's feelings and emotions in the situation, hence learning to read these signs are very important.

- Be calm, patient and don't interrupt while the patient is speaking as this would interrupt the client's thought process or would stop the emotional flow.
- Enter the world with them as they remember whatever they are expressing is actually where they are in time. (Their past is their present, the present is their future, and the future doesn't exist because they can't store memory.)
- Avoid criticism, correcting and arguing as this can be traumatic for the person. Focus on feelings, not facts, and encourage non-verbal communication.
- Offer best guess if you don't understand what is said by the patient as this will make the person feel secured and he will feel understood.
- Engage the person's "body memory" - called the "chaining" technique - to help them initiate or sustain an activity. For example place a glass of water in the hand.
- Confabulating serves to fill gaps in memory. Persons with dementia may make assertions that are not true to cover for memory loss. Trying to argue someone out of such beliefs is usually futile because the person is not lying.
- Refusal to cooperate may be due to sadness, anger, frustration, embarrassment, anxiety. Step back calmly to previous activity and assure the person that he / she is safe.
- Recollection of old memories are key ingredients to success such as humming a favorite song, talking about a pet, offering a familiar photo or object for a story. Smells, taste and touch are also strong memory triggers.
- Offer comfort and reassurance especially when the person is having difficulty expressing self; offer praise for success in accomplishment (e.g., completing a thought, reciprocating in an activity).
- Use "bridging" technique, a sensory connection that increases focus / attention and decreases anxiety. This could be a touch, a light guide on the elbow to steer, humming, stroking the skin with an object that has a unique surface such as satin. Be sure to ask permission before touching; tell the person what you are doing as you do it.

Communication Strategies for Psychologist: Speaking to Patients with Dementia

- Use short, simple, familiar and precise words, sentences and commands.
- Take on a similar posture to theirs to develop rapport non-verbally be on their level.
- Go to their eye level and be sure that they can see you clearly when you talk and listen.
- Other forms of communicating include music, touch, food and joint activity.
- Treat person as an adult and don't be condescending.

It is important to remember that the person who is suffering from dementia and has associated problems had at some point of time a life rich with history, experience, relationships, skills, hopes and dreams. In your role, you may have a wealth of information about this person as he / she declines, and it is important to share this information to keep continuity of care and interests alive for the person. Be sure to inquire into the activities that have had meaning to see if they are still being initiated.

Behavioural Strategies for Dementia Caregivers:

- Families and carers may find the following strategies helpful when responding to the behavioural and psychological symptoms of dementia:
- Every individual has a specific behaviour pattern, so do people suffering from dementia have. One needs to look for behaviour pattern and try to identify any triggers, as certain behaviours are displayed on at certain times or during particular activities. For example: at a very noisy place, or very bright light which may add to the confusion and restlessness.
- Establishing a routine is very helpful as it is helps patients with dementia be oriented, have sequential memory feeling secure. The person with dementia may become upset if they find themselves in a strange situation or among unfamiliar people, and may become confused, anxious, or agitated
- Clear and simple communication is very important i.e. giving specific commands. The person with dementia may become agitated if they do not understand what is expected of them. They may also feel frustrated with their

inability to make them self understood. It is important to face the person, speak slowly in a calm and reassuring voice and use simple sentences. Be patient and allow extra time. Ensure the person with dementia participates as much as possible in daily activities. Make sure the task isn't too complicated or the person with dementia may become overwhelmed.

- Regular exercise is very important. Simple exercise, like a walk outside or a game, can reduce anxiety and depression and can minimise other behaviours such as wandering and restlessness
- Distraction often helps. If possible, direct the person with dementia away from a particular task or environment that seems to be triggering the behaviour. Suggest a different activity that the person with dementia may like, such as listening to a favourite song, or going outside.
- Avoid punishment. The person with dementia may not remember the event, and therefore may not be able to learn from it. Try to stay positive.
- Be consistent. If there are strategies for modifying behaviours that work, try to ensure other people who also care for the person with dementia use the same strategies.
- Communication is important. Always explain to the person with dementia what is going to happen. Speak slowly in a calm and reassuring voice, and use simple sentences. Be patient.
- Involve the person with dementia as much as possible. Even if the person with dementia cannot participate verbally, they can still enjoy certain activities, such as reminiscing about their past by looking at old photos, or listening to a favourite song.
- Be sensitive to the reactions of the person with dementia. Therapies that involve remembering about the past, such as reminiscence or music therapy, may prompt happy memories, but may also prompt painful or sad memories.
- Avoid creating discomfort for the person with dementia. Sometimes the person with dementia may not feel comfortable being touched, so therapies such as massage may not be appropriate for these people.

OCCUPATIONAL THERAPY IN DEMENTIA:

The World Health Organization defines dementia as: "a syndrome due to disease of the brain, usually of a chronic or progressive nature, in which there is disturbance of multiple higher cortical functions, including memory, thinking, orientation, comprehension, calculation, learning capacity, language, and judgment. However, consciousness is not clouded. The impairments of cognitive function are commonly accompanied, and occasionally preceded, by deterioration in emotional control, social behavior, or motivation which occurs usually in Alzheimer's disease, in cerebrovascular disease, and in other conditions primarily or secondarily affecting the brain." (World Health Organization,2007). Dementia is not a specific disease, but rather symptoms caused by other illness or conditions that affect the brain. Contrary to what many people believe, it is not a normal part of the aging process. It is a severe loss of cognitive ability, such as thinking, memory, and reasoning, that interferes with a person's daily functioning. Symptoms include personality changes, behavioral problems, and memory problems.

Goal of Occupational Therapy:

The primary goal of rehabilitation is to enable people to achieve their optimal level of function. The challenge of occupational therapy is to create a comfortable balance between patient safety and maximum independence. The focus is on the strengths and the preserved capabilities of the individual to help them achieve quality of life regardless of level of cognitive status. Occupational therapists assist individuals to maintain or develop skills, which are needed to achieve functional independence and maintain their occupational role in everyday life using meaningful activities. OT assists patients and family members with changes needed in the living situation, different ways / techniques of performing daily activities, and / or tests patient's safety awareness. Occupational therapy is shown to improve quality of life for dementia patients and their carers.

OT Assessment:

The primary focus for the dementia patient is on habilitative, compensatory, non pharmacologic approaches that modify behaviors and / or the physical and social environment to help individuals

cope with the disease and continue to participate in everyday activities of living for as long as possible. An occupational therapy assessment includes

- History taking
- Cognitive and mental state examination (include examination of attention and concentration, orientation, short and long-term memory, praxis, language and executive function)
- Physical examination

Occupational Therapist evaluate present abilities and functional performance and determine the type of assistance, compensatory strategy, and environmental modification needed to successfully and safely complete activities. They also provide caregiver training in problem-solving, task simplification, communication and stress-reduction techniques to ease caregiver's burden. A holistic assessment of the resident's abilities and background is necessary to provide care and assistance that is tailored to the individual's needs.

People with dementia, which is a cognitive disability often report functional difficulties. Therefore Functional assessments should be done to identify impairments not demonstrated on formal cognitive testing. A functional assessment by an OT provides data that enables the other team members to gather a more complete picture of the person and their abilities. An OT assessment considers:

- The person,
- The task / activity &
- The environment.

The purpose of an occupational therapy evaluation is to design an intervention plan to increase participation, maintain occupational performance or modify activity demands, or prevent deterioration in performance capability. It is important to discern between what the patient can do and what they actually do. Ongoing evaluation should be adopted to ensure that any strategy / intervention used remains appropriate.

Assessments Utilized in Dementia are:

Disability Assessment For Dementia (DAD): Designed to quantify functional abilities in activities of daily living in patients with dementia and other cognitive impairments. Contains 40 items relating to basic self-care and instrumental aspects of activities of daily living. Scored from 0 (most impairment) to 100 (least impairment).

Cornell Scale for Depression in Dementia: The Cornell Scale (Alexopoulos et al, 1988) is specifically for the assessment of depression in dementia. The 19-item scale is rated on a three-point score of 'absent', 'mild or intermittent' and 'severe' symptoms, with a note when the score is unevaluable. A score of 8 or more suggests significant depressive symptoms. It is the best scale available to assess mood in the presence of cognitive impairment.

Mini-Mental State Examination (MMSE)

- It is a rating of cognitive function and takes 10 minutes to administer (Folstein et al, 1975). It is the most widely used measure of cognitive function.
- 26-30 (Cognitive impairment may still be present - especially in Fronto-temporal dementia and PD) - may require further assessment.
- 20-25 mild cognitive impairment
- 10-19 moderate cognitive impairment
- 0-9 severe cognitive impairment (Molloy, 2000)
- a score of 23 or less in an individual with more than 8 years education indicates cognitive impairment (Folstein, Folstein, McHugh, and Fanjiang, 2001)
- reliable and valid

Clinical Dementia Rating (CDR): This scale is used as a global measure of dementia (Hughes et al, 1982; Berg, 1984). It has become one of the main methods by which the degree of dementia is quantified into stages. Six domains are assessed: memory; orientation; judgment and problem-solving; community affairs; home and hobbies; and personal care. Ratings are 0 for healthy people, 0.5 for questionable dementia and 1, 2 and 3 for mild, moderate and severe dementia as defined in the CDR scale.

Alzheimer's Disease Functional Assessment and Change Scale (ADFACS) : It is used for the assessment of activities of daily living in patients with Alzheimer's disease with particular reference to outcomes in clinical trials (Galasko et al, 1997). It is informant-based and takes 20 minutes. Consists of ten items for instrumental activities of daily

living: ability to use the telephone; performing household tasks; using household appliances; handling money; shopping; preparing food; ability to get around both inside and outside the home; pursuing hobbies and leisure

Cognistat

- It identifies basic strengths and weaknesses
- A standardized evaluation to provide a basic cognitive profile including tests for Orientation, Language, Visual Construction, Memory, Calculation, & Reasoning.
- For adolescents, and adults in three age groups: 60-64, 65-74 and 75-84
- High level of reliability and validity

Clock drawing test

- It is a simple screening tool
- It is used with people who have executive cognitive dysfunction and a normal MMSE
- Measures a range of cognitive functions including visuospatial construction which is a skill known to be impaired in the early stages of dementia (Schramm et al., 2002)
- A normal clock suggests that a number of functions are intact and contributes to the weight of evidence that the patient may, for example, be able to continue independently. Alternatively, a grossly abnormal clock, is an important indicator of potential problems warranting further investigation or resource (Braunberger, 2001)
- good reliability
- more sensitive in identifying cognitive impairment than the MMSE (Flood and Buckwalter, 2009)

Hierarchic Dementia Scale (HDS)

- To determine a baseline and monitor a person's cognitive function.
- To assist with the formulation of care plans.
- Consists of 20 subscales each worth a maximum of 10, therefore the maximum obtainable score is 200.
- 160-190 mild dementia
- 159-42 moderate dementia
- 40-0 severe dementia
- Sensitive to change over time
- High inter rater reliability, testretest reliability, concurrent validity and internal consistency (Cole and Dastoor, 1996)

Assessment of Motor and Process Skills (AMPS)

- An observational assessment that is used to measure the quality of a person's occupational performance objectively.
- Useful for assisting with determining return to independent living and guardianship hearings.
- AMPS is designed to allow the person evaluated to choose what ADL task he or she will perform for the evaluation based on (a) the familiarity and relevance of the task to the clients daily life needs, (b) the degree of challenge that the tasks offer the client.
- 16 ADL motor and 20 ADL process skill items the person being assessed chooses two familiar and life-relevant ADL tasks to complete.
- Fully standardized valid reliable and sensitive assessment tool.

Modified Barthel Index (MBI)

- Measure dependence in ADLs
- Measures a person's performance in 10 ADLs with a maximum score is 100.
- Lower score the higher the level of dependence
- Good reliability and validity
- High inter-rater reliability

Functional Independence Measure (FIM)

- Assesses physical and cognitive disability assessment focusing on the burden of care
- 18 items scored 1-7 for level of independence. Scores can range from 18 to 126 with higher scores indicating more independence
- Good reliability and validity

When deficits and strengths in performance components and performance areas have been identified, occupational therapists work with patients and their caregivers to reduce the barriers in daily functional activities and facilitate maximum engagement in their environments, the human and physical context for daily living.

MOHO: MOHO is intended for use with any person experiencing problems in their occupational life and is designed to be applicable across the life span. Focus is on

- Systemic, holistic approach for persons of varying needs and populations across the lifespan
- Stresses the importance of the mind/body connection in its depiction of how motivation (internal) and performance of occupations (external) are interconnected
- Human occupation is described as the "doing" of work, play, or activities of daily living within a temporal, physical, and socio cultural context.
- Interactive nature between the person and his environment and how this relationship contributes to one's source of motivation, patterns of behavior, and performance.

OT intervention:

Through assessments there is clarification of the presence or absence of skills, the person strengths and weaknesses. The present skills are then fully utilized and developed. Absent skills are compensated through the development of various strategies. And this helps to minimize the impact of the condition on daily life and promote functional independence. Involving the person with dementia and the family members in the decision making process helps in easy acceptance of therapy. The plan, then developed should reflect client and caregiver needs.

ADLs

Dressing:

Dressing is a very complex task with numerous steps involved which can be overwhelming for people with cognitive impairment.

Some of the problems encountered in area of dressing are:-

- forgetting how to dress, to change clothes or dressing in the incorrect order (e.g., underwear over trousers)
- wearing extra layers as judgment and sensation of temperatures may be impaired
- may recognize the item of clothing but forget which body part goes into that clothing item
- difficulty with clothes selection
- difficulty manipulating clothes fasteners e.g., buttons, snaps, hook and eye, zippers
- Undressing frequently.

Possible strategies:

- Prompt or remind them of how to dress through verbal and physical prompts/cues. Using pictures or drawings of steps for dressing
- Be patient and allow as much time as is necessary for the person to complete steps
- Encourage the person to change regularly. If the patient wants to wear the same clothes every day, make duplicates available so that favorite clothes can be laundered.
- Use simple one step instructions
- Arrange closets and drawers systematically separating individual clothing
- Break task down into manageable steps
- Remove unnecessary clutter from the cupboard
- A mirror maybe useful for providing visual feedback while dressing
- Ensure environment provides adequate light and temperature
- Limit number of choices provided
- Utilize color contrast techniques for people with visual problems e.g., light color clothing on dark bedcover
- Select non crease clothing so ironing is not required
- Clothes with Velcro tape or elastic waist bands are preferred

Examples of labeling on drawers

Toileting and Continence:

A number of difficulties experienced by the person when using the toilet including perceptual difficulties, falls, hygiene difficulties, anxiety or behavioral problems are:

- Difficulty locating and transferring to the toilet
- Distance to the bathroom
- Difficulty undressing / maintaining hygiene
- Urinating in places other than toilet
- Falls while going to toilet
- Difficulty sitting still to use toilet

Possible solutions:

- Simplify clothing. Use Velcro / elastic instead of buttons and zippers.
- Select clothing that is easy to wear like elastic waistbands for trousers and wrap around skirts and also easy to wash to wash
- Use visual cues to assist with locating the toilet. Place a sign or label on the toilet door such as a picture or a photograph of the toilet in a prominent position. Leave the toilet door open and close all other doors leading to the toilet to discourage urinating in other rooms.
- Paint the bathroom / toilet door / door knobs / toilet seat / floor tiles & walls a contrasting color e.g., dark blue to make it stand out in a lighter colored hallway.
- Using large arrows to direct to the toilet from the living room or bedroom
- Utilizing sensor lights or night lights to avoid having to enter a dark
- Glow in dark strips placed around light switches or in hallway to assist in finding the light switch or direct the person to the bathroom
- Keep from passageways or stairways free from clutter
- Stick contrasting tape on rails to help locate them
- Install grab rails or equipment to assist with transfers
- Consider keeping a voiding diary (frequency and amount) which can assist with establishing a voiding routine e.g., every two hours.
- Once a routine is established try to avoid unnecessary changes
- Use simple one step instructions
- Use positive reinforcement to promote independence

Incontinence:

Incontinence is the loss of control of the bladder or bowel function. Being in control of these functions depends on having an awareness of bodily sensations and the memory of how, when and where to respond. Incontinence is always a symptom of an underlying problem. The cause of the incontinence should always be investigated as it may be due to numerous medical reasons such as infection, constipation, hormonal changes and prostate enlargement.

Dementia can impact on a person's continence by interfering with their ability to:

- Recognize the need to go to the toilet
- Short term memory loss resulting in frequent urination
- Be able to wait until it is appropriate to go to the toilet
- Knowing when the bladder / bowel is empty
- Locate the toilet
- Recognize the toilet
- Use the toilet correctly
- Manage clothing
- Attend to hygiene

Suggestions for managing incontinence:

- Be sure the person is drinking adequate fluids, preferably water, 5-8 glasses. Try to establish a regular routine for drinking fluids
- Reduce the person's caffeine intake by using decaffeinated beverages
- Observe the person's toileting pattern and suggest they use the toilet
- At regular times that follow their established pattern
- Utilize protective garments or disposable pads
- Utilize suitable aids or appliances.
- Ensure person consumes a high fiber diet
- Regular exercise

Eating:-

Eating habits and behaviors change during the course of dementia, and may be caused by physiologic or psychological factors.

- Straws with one way valves
- Modified eating utensils, plates with lips or rims
- Minimal reflections / glare from polished surfaces
- Reminders or prompts for meals and medications e.g., alarm clock or phone call allow and encourage finger feeding when person is no longer able to manipulate cutlery. Present finger foods on a flat plate at a comfortable reaching distance
- Reduce clutter: avoid lots of cutlery, crockery etc.
- Keep background noise and activity to a minimum, turn off TV
- Serve only one plate of food at a time
- Allow time for the persons memory to respond
- Physical prompts initially to use cutlery
- Modeling eating so patient can follow
- Offer meals at regular times
- Encourage physical exercise to promote a healthy appetite
- If assistance is required ensure the carer utilizes appropriate feeding techniques such as allowing sufficient time to chew and swallow, not over loading the person's mouth, using hand over hand techniques
- For chewing difficulties try light pressure on the lips or under the chin, tell the person when to chew, demonstrate chewing, offer small bites
- For swallowing difficulties verbally propmt the person to swallow, stroke throat gently, check mouth to see food has been swallowed, avoid foods that are hard to swallow, moisten foods
- Cut food into small pieces if over stuffing is an issue
- Monitor food temperatures

Use of contrasting colours in eating utensils

Bathing and Personal Hygiene

Personal care is often a sensitive issue for people with dementia. Tasks associated with hygiene were once completed independently and privately.

As dementia progresses the person may require physical assistance with the tasks resulting in a lack of privacy and autonomy. By focusing on the person rather than the task the person with dementia may be more willing to accept assistance. Identifying potential triggers and interpreting what the person is trying to communicate can assist in making personal care activities positive experiences. Being prepared and familiar with the person's usual routine and normal level of involvement will also assist in positive hygiene experiences.

Possible problems or concerns:

- Difficulty with manipulating taps
- Forget to turn off taps
- Change in sense of perception of hot and cold
- Unable to regulate water temperature risk of scalds
- Fear of water
- Fear of drowning particularly if water is being poured over their head
- Fear of falls
- Access to potentially dangerous items e.g., razors, mouth wash, electrical items
- Overwhelmed by complexity of the task (undressing, showering, drying, dressing etc.)
- Difficulty shaving, combing hair, applying makeup
- Poor oral hygiene, forgetting to brush teeth, forgetting how to use toothbrush or nail cutter

Possible solutions:

- Automatic taps that turn off if user forgets
- Put a few drops of blue food colouring in the water to strengthen its visual impact
- Non skid floor tiles which contrast with wall tiles
- Hand held shower level floor surfaces
- Non-slip floor tiles
- Grab rails, powder coated provides more grip (Calkins, M 2001)
- Keep access ways free from clutter
- Wide entry doorway with outward swinging or sliding door or hinges to allow removal of door / easy emergency access to bathroom use laminate signs or posters of bathing / grooming steps and hang them where the person can see them during the different stages of each task. If reading is difficult, use pictures or drawings.
- Soft calming music may be helpful in the background
- Choose the best time of day for the person for bathing i.e. when they are most relaxed or the time they used to bathe earlier
- Establish a routine
- Break down the task to manageable simple steps. Gently explain each step
- Lay out items that are required for task for example soap, shampoo, towel in the order they will be required
- Use simple clear one step directions
- Place items within easy reach to accommodate reduced mobility
- Use brightly colored soap, towels etc.,
- Model the action or give visual or verbal prompts when necessary
- The visual prompt could be a picture of the person performing the task, drawing or picture of another person performing the task, a drawing of the objects or a label specifying the task or objects
- Put grooming items out in the sequence they will be used
- Schedule regular visit to the dentists, saloons to maintain personal hygiene
- Use positive reinforcement and provide compliments

Examples of contrasting colours's use in the bathrooms.

Anti- skid mats

Sleep:

Sleep problems are often regarded as one of the most difficult symptoms of dementia. The person with dementia, their families and carers all require adequate sleep.

Possible problems or concerns:

- Disturbance to the person's biological clock i.e. being awake and restless during the night and sleeping during the day
- No longer able to distinguish between day and night
- Going to bed too early or sleeping too much during the day
- Overtiredness impacting on a person's ability to fall asleep
- Reduced activity resulting in reduced need for sleep
- Glare or reflections from glass or mirrors can be disturbing
- Temperature of bedroom: maybe too hot or cold
- Nightwear that is too restrictive, hot or cold
- Bed wetting due to incontinence
- Difficulty in locating bed

Suggestions:-

- Establishing a sleep routine that is familiar and predictable
- Avoid having day clothes on display at night as this may trigger a response to get up
- Try to incorporate exercise into their daily routine
- Listening to relaxing music, radio or television themes associated with preparing for bed
- Water proof covers
- Keep the environment as consistent as possible and free from distractions
- Utilizing a contrasting top and bottom sheet may assist the person to locate their bed
- Gently remind the person it is night time and time to sleep
- Reduce time duration of sleep during day time

Medication Management:

Medication should be reviewed by a doctor regularly.

Possible problems or concerns:

- forgetting to take medication or taking twice or more times
- incorrect use or storage of medication
- using expired medication or taking medication that is not required
- difficulty opening bottles or containers.

Possible solutions:

- Medical / nurse review re - need for medication, dose and frequency
- Leave medications in a visible location (if safe)
- Reminders in the form of Alarm system, notepads
- Link medication times to routine activities e.g., taking medications at meal times
- Count and keep medications according to date and dosage in separate packs

Attention and concentration:

Strategies to improve attention and concentration:

- Remove distractions whenever possible (turn off the TV and radio during conversations)
- Talk to the person one at a time
- Concentrate on relevant material
- Simplify information and written instructions
- Reduce the amount of information that is presented at one time
- Get the person to repeat back information to ensure he or she has understood what was told
- Get the person to focus on one task / step at a time
- Keep the person focused by breaking down tasks into manageable parts
- Allow enough time to process information

Memory:

Due to the cognitive deficits the person with AD struggles with memory losses, increasing forgetfulness disorientation and confusion. The following ideas serve as memory aids to help the person with dementia:-

- Labeling cupboards and drawers, maybe using pictures rather than words - for example, a photo of a cup and jar of coffee

- Keep a large calendar showing the day, month and year in every room
- Visual schedule of activities and Alarms
- Signs, line drawings or arrows to help identify rooms or objects
- Keep a notepad / board by the phone for messages
- Use an answering machine for phone messages
  - Decide on a consistent place to keep handbags, keys and wallets
  - Attach important items so they cannot be mislaid (using a neck cord for glasses, tying keys to a belt)
  - When a person is trying to recall an event, assist them by providing a meaningful context (who was there, what the event was for, how long ago it occurred); providing cues and prompts to aid recall
- Write down all appointments in a clear and simple diary or wall-calendar.
- Only use one diary / whiteboard or wall-calendar; use it consistently and encourage the person to refer to it every day
- Set an alarm to remind the individual of things that he or she needs to complete - if the person is used to using a mobile phone or electronic daily planner these are ideal tools.
- Make sure the person has his or her name, address and contact in his or her wallet / purse and consider an identity bracelet if disorientation is becoming a problem.
- Use of remembering techniques e.g. Mnemonics

Games to improve memory

Examples of labeling different rooms of the house

Speech and communication:

Can be improved by:-

- Allowing the person more time to communicate
- If the person is struggling to find a word in conversation, offer assistance after allowing a reasonable amount of time
- Stand in front of the person and minimize distractions
- Place yourself at eye level
- Speak gently and clearly using statements rather than questions
- Wait for a response from the person with dementia before continuing

If the ability to communicate through spoken word or written language is lost, then other forms of communication can be used. Encourage and use lots of Nonverbal communication like Facial expressions, Gestures and Eye contact.

Assistive Technology:

Assistive technology is any item or object that enables a person to complete a task that they would otherwise be unable to do. Independence in task completion impacts positively on a person's sense of self worth, self esteem and self reliance. Assistive technology should not replace human contact.

When considering assistive technology interventions a simple approach should be utilized and only changing what needs to be changed. Caution should be utilized as every person's reaction to technology will be individual and in some cases it may have a negative impact. Involving the person with dementia in the decision making process and trialing of items will lead to greater acceptance and use of the technology solutions.

Examples of some of the technology currently available:

- Electronic assistive technology: automated door openers, alarms, computers, smart stove tops etc.
- Mobile phones: features vary on each phone but can include speed dialing, call blocking, larger buttons and screen, calendar with reminders/ to do lists, medication/schedule reminding software, GPS tracking etc
- Computers: larger keys or keyboard; touch screen; screen enlarger or magnifier, speech and voice recognition programs; screen readers that read all screen contents; software programs that use speech synthesizers etc.
- Use of a Universal remote.

Mobility and Falls:

Falls in dementia can occur for a wide variety of reasons. Even though people with cognitive decline are three times more likely to fall, mobility should always be encouraged.

Daily mobility helps to:

- maintain strength and endurance
- improve balance
- maintain joint mobility
- promote cardiovascular health

Tips to assist with mobility and falls prevention:

- Make movement/ daily physical activity enjoyable
- Ensure furniture that the person transfers from is a suitable height, has a firm base, where possible use chairs with arm rests
- Ensure suitable footwear is worn
- Minimize clutter by arranging furniture simply and leave it in the same place
- Keep the floor free from hazards such as rugs or items that can be walked into or tripped upon
- Strategically place sturdy furniture for use when mobilizing
- Eliminate glare from all areas where the person is transferring or mobilizing
- Give time for the care recipient's eyes to adjust to changes in light when moving from one area to another.
- Avoid use of furniture with sharp edges, carpets with turned edges
- Use single word commands when directing transfers or ambulation
- Training may be required to teach safe transfer and handling techniques. The carer may also benefit from back protection or energy conservation techniques.
- Provide positive reinforcement when the person displays safe behaviors
- Use visual, verbal or physical prompts
- Use antiskid mats and floorings

Leisure and social Activities:

Engaging in leisure activities promotes physical, social and emotional health. People with dementia gradually withdraw from leisure and social activities and become socially isolated. Although Simple games can provide enjoyment for people with dementia, the person should not be treated as a child, when planning recreational activities. Gardening or other hobbies, arts and crafts, or pets can all be enjoyable sources of stimulation. Activities should be tailored to the individual's personality.

Some solutions to improve leisure skills and social interaction are

- Place one interesting activity, game or item in a prominent position where it will be noticed. Once interest declines replace with another item of similar skill level and interest
- Choose activities that are familiar, repetitive, require one step
- Incorporate former interests or hobbies
- Choose movies and music that are era specific for the person
- Plan leisure as a part of the daily routine
- Encourage exercise and activities that require no strenuous gross motor actions. Walking with another person is an excellent activity.

- Avoid competitive activities. Choose activities that meet the person's capabilities
- Simplify the rules of games to encourage success
- Utilize talking books if reading has become difficult
- Make a family history book / family photo boards. Use captions to assist with memory / orientation
- Talking photo albums: available from specialty photographic stores
- Compile photos / videos to make DVD's which could be viewed during passtimes
- Utilize easy cookbooks i.e. large font, spiral bound with step by step instructions
- Encourage the person to watch the activities of the neighborhood from a window or veranda
- Traveling is encouraged if the person can tolerate the changes. Try to preserve the daily routine, plan frequent rest breaks, take a third person if able to assist with the caring role. Take a recent photo of the person in case they become lost. A card with the care givers details and person's details can be placed in a pocket or purse.
- Encourage them join a support group for people with dementia.

Conclusion:

Thus an occupational therapist uses a combination of education, setting up of feasible goals, adaptations in physical environment, training of compensatory skills, training supervision skills, and also changing dysfunctional cognitions on patient behavior and caregiver role.

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Ch.7 Cerebellar Ataxia

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Introduction

Normal motor movements are achieved in human beings with constant neuro-developmental maturation following birth and gets completed when normal postural mechanisms become localized, followed by the ability to maintain balance in different positions and lastly by the formation of muscular coordination.

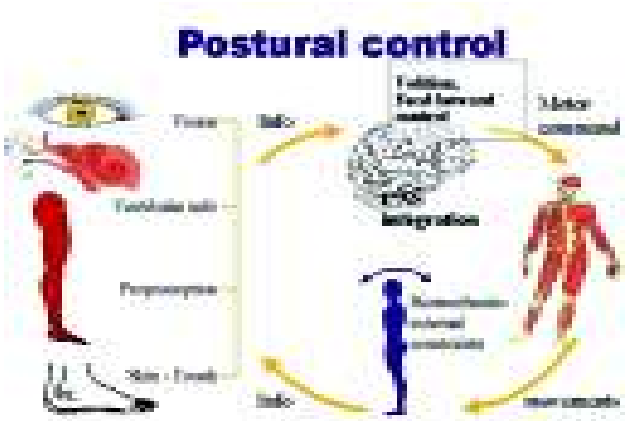
Normal postural tone helps in keeping posture erect in standing against gravity, adaptation to changes on support surface and proximal stabilization. Balance can be defined as postural adaptation to changes in gravitational center with the contribution of normal postural tone. Muscular coordination on the other hand, is the functioning of all muscles active during the voluntary motor movement in appropriate rhythm, velocity and amplitude. A person may perform daily life activities through normal motor movement formed by the above mentioned three components.

Nervous system diseases and/or injuries usually affect postural control mechanisms where in some of these diseases, balance problems are more dominant and in others coordination problems related to extremities are prominent.

Ataxia meaning 'disorderly' (Bastian 1997) is defined as "the in-coordination of movements" (Bastian 1997, Mariotti et al. 2005) and where both **insufficient postural control and incoordination of multi-joint movements are observed. (DeSouza 1990) This results in postural** instability leading to imbalance and walking disturbances. It has three subcategories, which are sensory, cerebellar and vestibular ataxia. Some researchers has given frontal ataxia as the 4th category whereas 5th category termed as mixed ataxia which involves symptoms of at least two basic types of ataxia together have been put forward.. In some cases incoordination is usually accompanied by balance dysfunction and gait problems whereas in other cases, balance dysfunction is observed without the existence of in-coordination of movements which includes muscle weakness. For example, if normal walking is considered to be the perfectly timed sequence of muscle activity that occurs in response to both internal and external forces, then it is easy to consider walking as a task requiring coordination as well as balance (Crutchfield et al. 1989). Since ataxia is resistant to medical treatments, physical treatment plays an important role which involves proprioceptive training, balance exercises, stabilization techniques for the extremity ataxia and vestibular exercises for accomplishing functional improvement and restoration of the ataxic patients. Compensatory techniques can be used by adaptive devices thereby improving quality of life(1)

Anatomical Structures Responsible for Balance Development and Coordination

Normal motor control is the output of normal postural tone, muscle coordination, and balance working together. Postural control within the normal functioning of the nervous system is maintained by the different sensory-motor subsystems working together within a circular network. The vestibular, visual, somatosensory systems and cerebellum work in a coordinated manner to produce postural orientation depending upon the goal of the movement and environmental conditions. Each subsystem may be dominant at different times. For example, whenever the type or quality of the surface on which the patient is standing changes, the changes will be registered by the somatosensory system whereas when the information in the visual area changes, the visual system will register changes. The subsystems collecting the information at that particular moment will dominate thereby determining the appropriate motor response (Crutchfield et al. 1989).



S**omatosensory System (proprioceptive and superficial senses):** Sensation from lower extremities, position of cervical (Treleaven 2008) and lumbar area, length of muscles and positions of joints are transmitted through proprioceptors located in joints, ligaments, muscles and tendons. The contribution of proprioceptive senses (position and kinesthesia) is particularly important in the formation of normal motor function (Sherrington 1907, Bear et al. 2001).

Position and kinesthetic senses are transmitted via fibres with thick myelin which convey messages rapidly and are transmitted to central nervous system through two paths. The first one is the dorsal column- medial lemniscal system through which conscious sensations are conveyed to sensory areas 3,1,2 of cerebral cortex (Bear et al. 2001). Proprioception and the visual system, provides information about speed, form and size of the movement that motor cortex has to generate. (Scneider et al. 1977).

Ventral and dorsal spinocerebellar tracts are the second path through which proprioceptive information is transmitted from spinal cord to brain. The sensory input which is carried by these tracts comes to an end in the area called spinocerebellum within the cerebellum, and does not reach consciousness. In other words, the sensory input carried by these tracts is the subconscious proprioceptive sense. This sensory input enables the cerebellum to correct the faulty motor commands that the motor cortex may send by informing the cerebrum and cerebellum simultaneously about the size, speed, form and timing of the movement before the movement is performed (Guyton 1976, Ramnani et al. 2001).

Dysfunction of the dorsal columns of the spinal cord results in loss of proprioception which generally carries the proprioceptive information to the brain leading to condition called **Sensory ataxia.** At times it may be due to dysfunction of various brain parts which receives those information, including the thalamus, and parietal lobes.

Sensory ataxia shows the following features:

1. High steppage gait and postural instability due to lack of proprioception which cannot be compensated by visual input.
2. Romberg's Test is positive when patient's stands with his feet together and eyes closed resulting into instability with wide oscillations and possibility for fall. This finding differentiates sensory ataxia from the other ataxia.(Bannister 1992).
3. Loss of vibration sense in the extremities and loss of deep tendon reflexes.
4. Worsening of the finger-pointing test with the eyes closed.

Sensory ataxia are generally observed in:

1. Hereditary types of ataxia such as Friedreich's ataxia and spinocerebellar ataxia.
2. Other diseases like diabetic or alcoholic neuropathy, vitaminB12 inadequacy neuropathy, tabes dorsalis,
3. Tumoral conditions found in the posterior cord of the medulla spinalis,
4. Multiple sclerosis (Edwards 1996).

Vestibular System:

Vestibular system has two pathways:

1. Central Pathway.
2. Peripheral Pathway.

The central vestibular pathways coordinate and integrate information about head and body movement and use it to control the output of motor neurons that adjust head, eye, and body positions.

Primary vestibular axons from cranial nerve VIII make direct connections to the vestibular nucleus of the same side of the brain stem and to the cerebellum. The vestibular nuclei also receive inputs from the cerebellum, visual and somatic sensory systems, thereby combining incoming vestibular information with data about the motor system and other sensory modalities.

The vestibular system, is composed of otolithic organs (utricle and saccule) and semicircular canals.

1. **Vestibulo-spinal reflex:** Axons from otolith organs project to the lateral vestibular nucleus, which then projects via the vestibulospinal tract to excite spinal motor neurons controlling leg muscles which helps to maintain posture even on unstable surface.
2. Axons from semicircular canals project to the medial vestibular nucleus, which sends axons via medial longitudinal fasciculus to excite motor neurons of trunk and neck muscles that orient the head. These pathways help the head to stay straight even if the body moves around.

One important function of the vestibular system is to keep eyes fixed in a particular direction, which is actualized by vestibulo-ocular reflex which works by sensing rotations of the head and immediately commands a compensatory movement of the eyes in the opposite direction. The movement helps to keep gaze fixed on a visual target. (Guyton 1976, Bear et al. 2001).

Vestibular ataxia: develops as a result of peripheral or central diseases which directly affects the vestibular nuclei or the afferent and efferent connections of the vestibular nuclei.

A patient with vestibular ataxia shows :

1. Symptoms of balance disturbances in standing and sitting. Patient represents stagger, has a broad base support and may tend to lean backwards or towards the side of the lesion while walking.
2. Head and trunk motion along with arm swing is often reduced (Borello-France et al. 1994). The patient is limited particularly when crossing the street and shopping at the market
3. Balance is disrupted when performing a head or eye movement.
4. It may be accompanied by vertigo, nausea, vomiting, blurred vision and nystagmus due to the vestibular system's role in sensing and perceiving self-motion and stabilizing gaze via the vestibulo-ocular reflex (Horak&Shupert 1994). Extremity ataxia is by no means observed in vestibular ataxia.
5. Deep tendon reflexes are normal.

Vestibular ataxia can seen in :

1. Central factors such as medullar stroke and multiple sclerosis.
2. Peripheral vestibular diseases such as Menier's, benign paroxysmal vertigo, or vestibular neuronitis.

Cerebellum:

Anatomy and Physiology of Cerebellum:

The transverse lobular arrangement of cerebellum shows neuro anatomical presentation and 7 longitudinal mediolateral parallel zones on each side of midline and are termed as functional units of the cerebellar cortex.These zones are apparently formed via developmental mechanisms and with evolution the cerebellum has expanded mediolaterally.

The medial zone is involved in adaptive control of somatic and autonomic reflexes and compound movements such as locomotion and saccadic eye movements commonly seen in vertebrate species.

The intermediate zone is developed in relation to voluntary movement in mammals.

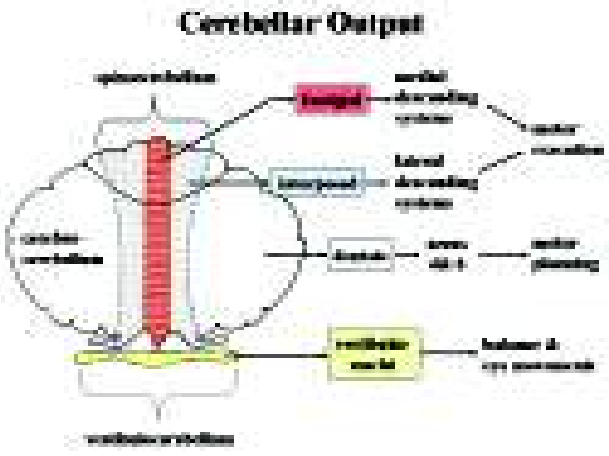
The lateral zones are related to higher functions of the cerebral association area. In humans, it is likely to be associated with cognition.

Each zone receives afferents from discrete areas entering the cerebellum via mossy or climbing fibers. These two fiber systems transmit various types of information and influence the efferents cerebellar Purkinje cells to relay the information to cerebellar or vestibular nucleus. The mossy fiber system carries afferent information from the spinal cord, brain stem, and cerebral cortex via pons and is responsible for moment-to-moment, rapid firing of Purkinje cells and then modulates ongoing movements. The climbing fibers relay information to the cerebellum from the inferior olivary nucleus which results in slow firing of Purkinje cells that seems to be important for motor learning.

The input into the cerebellum is from all 3 peduncles with the ascending input through the inferior and the cortical input through the middle cerebellar peduncle. The superior cerebellar peduncle is responsible predominantly for the output from the cerebellum. The afferents received by the cerebellum have specific functions in terms of locomotion, postural control, voluntary movements, and finally cognition within the cerebellum.

Functions of cerebellum:

The cerebellum has a crucial role in balance and locomotion where different zones are responsible for different functions. In humans. cerebellar



damage results in postural sway with difference seen in their amplitude, frequency, direction and movements in relation to lesion affecting the zone.

- The medial zone of cerebellum : This zone integrates spinal and vestibular inputs and subsequently projects out to vestibulospinal and reticulospinal tracts through the fastigial nucleus.

  Functions :

  1. They exert modulatory control of the rhythmic flexor and extensor locomotor pattern generated by vestibular and reticular nuclei.
  2. They also control extensor tone to maintain upright balance and stance.

A lesion in this zone leads to a significant balance problem and impairment of postural tone with low frequency, high amplitude postural sway, without a preferred direction and without intersegmental movements.

Intermediate zone (paravermal region): receives input from the spine (via spinocerebellar tracts) and projects out to the red nucleus and cerebral cortex. through the globose and emboliform nuclei It integrates spinal and cortical inputs and influences locomotion through projections to motor cortical areas.

Function of the intermediate zone in relation to limb placement includes :

1. Timing of limb placement.
2. Elevation of limb placement
3. Trajectory of limb elevation.
4. Descent of limb placement.

Lesion to this region leads to gait ataxia and swing phase overshoot of legs with no change in balance or postural tone.

It includes increased postural sway of high velocity and low amplitude in anteroposterior direction, increased postural tremor, increased intersegmental movements of the head, trunk, and legs.

- **Lateral zone:** This area receives input primarily from cerebral cortical area via pontine nucleus (corticopontocerebellar fibers) and projects out to the thalamus and cerebral cortical areas via the dentate nucleus through the red nucleus.

  Functions:
  1. It helps in voluntary modification of motor activities and the locomotion.
  2. Lateral cerebellum is especially active in novel walking conditions where precise limb placement is necessary.
  3. It modulates visually guided motor activities because of the robust projection it receives from the visual cortex.

- A lesion in this region leads to limb ataxia and locomotion problems in normal walking and challenging situations with slight postural instability or sway.

In normal walking, balance deficits is seen more in cerebellar gait ataxia (medial and intermediate zone) whereas visually guided leg control deficits are seen in the lateral zone.(2)

Voluntary movements controlled by cerebellum:

Cerebral cortical association areas plan voluntary movements and the plan is executed by the motor cortex. But there is a robust cerebellocerebral loop that modulates these motor functions. These loops connect the intermediate part of the cerebellum to the association cortex and the motor cortex. It helps in planning and monitoring the movements..

Cognitive function of cerebellum

A closed cerebellocerebral loop is found in the prefrontal cortex and thus the cerebellum provides a forward model for mental functions in the cerebral cortex which is analogous to cerebellocerebral loop concerned with motor functions. A primary cerebellar injury in premature infants has shown to be associated with contralateral decrease in cerebral volume.[7] This show the evidence of the importance of the cerebellocerebral connections responsible for important cognitive functions.

A mental model of image, idea, or concept is formed in the temporoparietal association cortex which are manipulated by the prefrontal cortex. After repeated exercise, the cerebellum copies a mental model to form an internal model through cerebello-cerebral loop. Because of this internal model formed by the cerebellum, we are able to conduct movements and thoughts unconsciously

Clinical phenotypes show considerable overlap; however, the genetic, molecular, and biochemical

causes for these disorders are often distinct. In dominant ataxias phenotypes show considerable genetic heterogeneity. These phenotypes may manifest with pure ataxia or involve multiple levels of the nervous system (including dementia, seizures, disturbance in proprioceptive function, movement disorders, and polymyoclonus).

Thus, from the proprioceptors in the periphery, cerebellum learns the position of the body in space. It receives information about balance from the vestibular system as well, and through cortico ponto cerebellar tract about the features of the intended motor movement. Based on this information, the cerebellum facilitates coordinated and balanced movement by making the appropriate adjustments. (Guyton 1976, Young&Young 1997, Herdman 1998).

Cerebellar ataxia

Cerebellar ataxia develops as a result of lesions to the cerebellum or the afferent and efferent connections of the cerebellum.

1. Vestibulo-cerebellar dysfunction corresponds to lesion in the flocculonodular lobe (flocculus and nodulus) and involves problems regulating balance and controlling eye movements thereby resulting with postural instability, which worsens when standing with the feet together irrespective of whether the eyes are open or closed: this is a negative Romberg's test. (Liao et al. 2008, Morton & Bastian 2004).
2. Spino-cerebellar dysfunction corresponds to the vermis and paravermis.

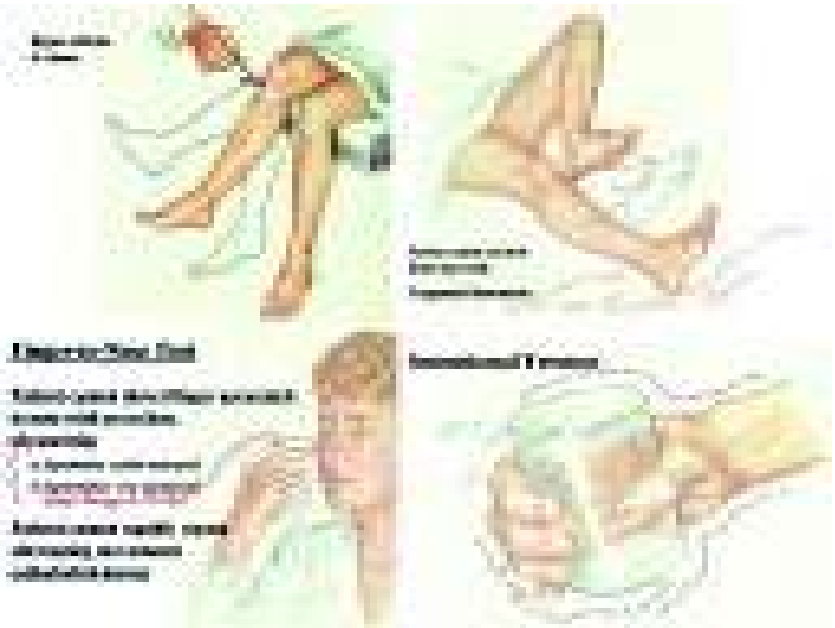
It results :

1. With a wide-based gait, characterized by uncertain start and stop,
2. Lateral deviations.
3. Unequal steps.
4. Abnormal inter-joint coordination patterns.

When this part of the cerebellum is damaged, gait ataxia or walking in-coordination occurs.

1. Cerebro-cerebellar dysfunction indicates a lesion of the deep pontine nuclei connections with the cerebellum. The cerebro-cerebellum contributes to planning and monitoring of movements and any lesion results in disturbances in performing voluntary, planned movements.

Symptoms associated with cerebellar ataxia include:



1. **Dysmetria:** This refers to inaccuracy in achieving a final end position (hypermetria equals overshoot; hypometria equals undershoot). This clearly is demonstrated by the patient attempting the finger-nose test.
2. **Tremor:** Kinetic tremor, which is oscillation that occurs during the course of the movement.
   - Intention tremor: Increase in tremor which is seen towards the end of the movement.
   - Postural tremor: occurs when holding a limb in a given position.
   - Titubation: Tremor affecting the head and upper trunk showing lesion in the vermis.
   - Postural truncal tremor: affecting the legs and lower trunk, which is seen in anterior cerebellar lobe lesions.
3. **Dyssynergia:** is particularly seen while doing multi-joint movements especially when:
   1. Agonist-antagonist and synergistic muscles may not be able to contract in correct order during voluntary movement;
   2. Antagonist muscle may be failing to control eccentric contraction during the concentric contraction of agonist muscle.

   With the combination of these two factors, the extremity undergoes a sudden velocity resulting in inappropriate and uncontrolled motor movement.
4. **Dysdiadockokinesia:** This is the inability to perform rapidly alternating movements such as alternately tapping with palm up and palm down. The rhythm is poor and force of each tap is variable.
5. **Hypotonia:** This occurs in acute cerebellar lesions, but it is rarely seen in chronic lesions. Hypotonia is seen in proximal and antigravity muscles.
6. **Weakness and fatigue:** A generalized non-specific weakness as a feature of cerebellar dysfunction often observed with extensive and deep lesions and is most apparent in the proximal musculature. Fatigue has also been noted as a common feature of cerebellar dysfunction.
7. **Dysarthria:** This occurs due to in-coordination between tongue and lip muscles. The patient speaks like he is drunk.
8. **Nystagmus:** Abnormal eye movements that develop in horizontal and vertical directions mostly as nystagmus at the end point (Edwards 1996).
9. Deep tendon reflexes are maintained in cerebellar lesions, and gain a pendular characteristic.(5)

Cerebellar ataxia can be observed:

1. Spino-cerebellar ataxia among hereditary ataxias.
2. Friedreich's ataxia.
3. Chronic alcoholism
4. Paraneoplastic cerebellar degeneration.
5. Pontocerebellar angle tumors.
6. Multiple sclerosis.

As the underlying mechanisms of disease begin to be understood, the inherent challenges are apparent; for instance, several ataxias are caused by defects in DNA repair, while others may result from protein folding and chaperoning defects. Advances in genomics, proteomics, transcriptomics, and metabolomics are paving the way towards understanding of gene function, protein synthesis and transcription, and gene-gene and protein-protein interactions.(6)

Frontal ataxia

Frontal ataxia (also known as gait apraxia) is observed when tumors, abscesses, cerebro vascular accidents and normal pressure hydrocephalus effect the frontal area. It shows the following features like:

- Patient has difficulties standing erect
- Even with use of support, patient tends to lean towards hyperextension
- Patient's legs are in scissors-cross position during walking and there is incoordination between the legs and trunk
- Ataxia is accompanied by frontal dementia, urinary incontinence, frontal release signs and perseveration (jeffmann.net / Neuro Guide-maps / gait).

Mixed ataxia

Mixed ataxia refers to the type of ataxia when symptoms of two or more types of ataxia are

observed together, such as occurrence of sensory and cerebellar ataxia symptoms.

In some diseases, mixed ataxia may be observed frequently. For instance, in Multiple Sclerosis, cerebellar, vestibular and sensory ataxia may be observed together; whereas in cases of spino-cerebellar ataxias, cerebellar and sensory ataxia may be seen.

Features of basic types of ataxia are briefed in Table 1 below:

Table 1: Clinical Differences Between Basic Types of Ataxia

Type	Cerebellar Ataxia	Sensory Ataxia	Frontal Ataxia	Vestibular Ataxia
Head posture	Upright and sometimes fixed	Leans forward	Leans forward	Upright and definitely fixed
Trunk posture	Stooped-leans forward	Stooped-upright	Upright	Upright
Stance	Wide-based	Wide-based	Wide-based	Wide-based
Initiation of gait	Normal	Normal-wariness	Start hesitation	Normal
Postural reflexes	+/-	Intact	May be absent	+/-
Steps	Stagger-lurching	High-stepping	Small-shuffling	Normal
Stride length	Irregular	Regular	Short	Normal
Leg movement	Variable, ataxic	Variable - hesitant and slow	Stiff, rigid	Normal
Speed of movement	Normal-slow	Normal-slow	Very slow	Normal-slow
Arm swing	Normal, exaggerated	Normal	Exaggerated	Normal
Turning corners	Veers away	Minimal effect	Freezing - shuffling	Dysequilibrium
Heel-toe test	Unable	+/-	Unable	Unable
Romberg's test	+/-	Increased unsteadiness	+/-	–
Heel-shin test	Usually abnormal	+/-	Normal	Normal
Falls	Uncommon	Yes	Very common	Common

Genetic-biochemical basis for classification

Initially on the basis of anatomical localization of pathological changes, inherited ataxias were classified.

In 1993, Harding introduced another classification in which the ataxias were placed into 3 categories:

1. Congenital.
2. Inherited metabolic syndromes with known biochemical defects.
3. Degenerative ataxias of unknown cause.[8]

The last category was subdivided further into early onset (< 25 y) and late-onset types

In ataxia:

1. Presentation can be variable (eg, static vs progressive, intermittent vs chronic, early vs delayed).
2. Mode of inheritance also varies such as Autosomal dominant, recessive, and nonmendelian inheritance patterns where nonmendelian inheritance patterns refers to disorders of inheritance for which the rules of Mendelian genetics do not apply.

Disorders of triplet repeat expansion and certain mitochondrial defects are examples of nonmendelian inheritance.

Causes of Ataxia:

1. Idiopathic or Sporadic Ataxias.
2. Vitamin B12 deficiency: with other neurological abnormalities can cause overlapping cerebellar and sensory ataxia.
3. Inherited Ataxias: caused by a genetic fault inherited either from the mother or father or

both. A faulty gene that is passed down through generations can result in cerebellar ataxia. At times, the severity worsens from one generation to the another with age of onset getting more younger group. This type of worsening inherited ataxia is known as anticipation.

For getting inherited ataxia, the chances depends on the type of ataxia that parent has.

To develop Friedreich's ataxia, the faulty gene would need to be carried by both the mother / father and the father (recessive inheritance).

Spinocerebellar ataxia: Faulty gene requires one parent to carry the defective gene(dominant inheritance) and each of his / her offspring would have a 50% risk of developing the condition.

3. **Non-inherited Ataxia:** Without any family history.

The following are the:

1. **Brain Surgery**.
2. **Head injury**.
3. **Alchol abuse.**
4. **Drug abuse:** can cause ataxia as they have a depressant effect on central nervous system function. E.g Ethanol is capable of causing reversible cerebellar and vestibular ataxia. Antiepileptic drugs produces cerebellar ataxia as a adverse effect.
5. **Infections** such as Chicken pox.
6. **Brain tumour.**
7. **Exposure to toxic Chemicals:** Inability to excrete copper from the body is seen in Wilson's Disease which is autosomal-recessive gene disorder. Copper accumulates in the nervous system and liver causing ataxia as well as other neurological and organ impairments.
8. **Multiple sclerosis**, **Cerebral palsy** and some other neurological conditions.
9. **Malformation** of the cerebellum while the baby is still in womb.
10. **Radiation Poisoning:** Ataxia can be induced as a result of severe acute radiation poisoning with an absorbed dose of more than 30 grays.

Diagnosis: With patient's medical history for causative factors such as brain injury as well as family history for inheritance can help in reaching upto diagnosis.

The following tests are performed:

1. Magnetic resonance imaging (MRI) or Computerized Tomography (CT) scan to determine whether there is any brain damage.
2. Genetic tests: to determine whether the patient has inherited ataxia.
3. Urine tests: Urinalysis may suggest specific abnormalities that are associated to some types of ataxias.
4. Blood test: to rule out any specific type of ataxia.

Assessment :

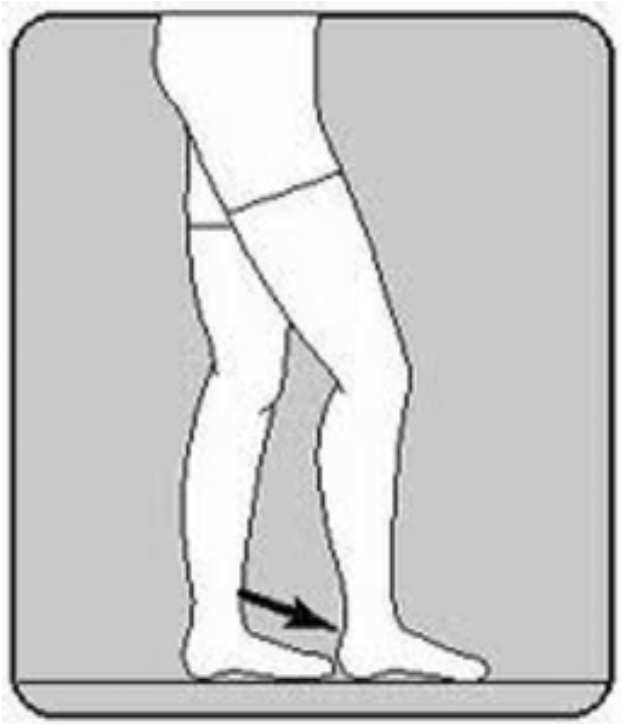
1. Age of onset.
2. Sex
3. Mode of onset (ie, acute, subacute, chronic)
4. Clinical History
5. Natural history (ie, nonprogressive / static, episodic, progressive)
6. Associated symptoms / signs that provide localizing information such as:
    1 Presence of dystonia or chorea suggesting involvement of the striatum
    2. Proprioceptive dysfunction suggesting involvement of spinocerebellar pathways
    3. Visual deficits (retinitis pigmentosa), auditory involvement (Refsum disease)
    4. Cognitive dysfunction possible early and / or late

Examination1,3 of cerebellar Signs:

1. Evaluate muscle power, tone and reflexes:

    In cerebellar disorders, Tone remains mild hypotonia and reflexes shows hyporeflexia.

- Evaluate upper limbs for limb ataxia :
    1. Rebound of outstretched arms.
    2. Finger-nose test
    3. Dysdiadochokinesis.
- Evaluate co-ordination in leg with heel-shin test.
- Ask patient to sit up with arms crossed to check for truncal ataxia.
- Ask the patient to walk heel-to-toe (Tandem Walking) to check any gait ataxia.



- Ask the patient to stand with feet together where in cerebellar disorders, patients will be unsteady with eyes open or closed - not a true Romberg's positive.
- Ask the patient to take their tongue out and move it from side to side (movement slowed).
- Ask patient to repeat "baby hippopotamus" thereby looking for dysarthria and abnormal speech rhythm and syllable emphasis.
- Evaluate eye movement for ophthalmoplegia or nystagmus.
- Evaluate fundi for papilloedema.
- If unilateral signs - check V, VII, and VIII (cerebellopontine angle pathology)(8)
- Other systemic features like:
    1. Dysmorphic features and associated congenital malformation suggesting of a specific association or clinical syndrome.
    2. Cardiac (Friedreich ataxia), renal (NPCA), and cutaneous (xeroderma pigmentosa) features are examples.
- Family history and pedigree analysis provides diagnostic clues and information on possible patterns of inheritance, which are useful for planning investigations and genetic counselling.
- MRI is useful to identify the location of the lesion (e.g cortex, white matter, cerebellum, striatum, brainstem, etc), and patterns of involvement.
- Other investigations like MR spectroscopy, cytogenetic studies and metabolic screening tests are also useful.

MEDICAL TREATMENT

Several drugs used to control ataxia including muscle relaxants like diazepam which also reduces muscle spasm.

Speech Affection in Cerebellar Ataxia:

Cerebellum along with the other parts of central nervous system forms a control circuit to refine and fine tune speech. Individuals having cerebellar affection have typical ataxic dysarthria. Their speech may sound "drunkard" type with lot of slurring. The rhythm and stress pattern of speech is affected. Language is usually normal in these patients as there is no higher cerebral involvement.

The therapist can guide them about how to make the voice sound clearer by :

1. Changing the posture thereby improving the quality of voice.
2. To strengthen the muscles with exercises which are used for talking.
3. Teaching to speak slowly to make it more emphatic.
4. Breathing techniques used to improve speech.
5. If dysarthia worsens, speaking aids, such as a laptop computer that is connected to a voice synthesizer can be used as modification.
6. In case of dysphagia, therapist can teach the exercises which will stimulate the nerves which will trigger the swallowing reflex and strengthen the muscles used during swallowing.
7. Physical techniques like tucking the chin forward during swallowing will prevent any food from entering the airways.

Nutritionist: will guide about what should be incorporated to have a healthy and balanced diet like more of fibre type of food and liquids which will be easier to swallow and digest.

For e.g mashed potatoes are a good source of carbohydrates, and scrambled eggs and cheese are high in protein and calcium.

Physical therapy and rehabilitation of ataxia

The goal of the physiotherapist is to improve the functional level of the patient through restorative techniques. When this is not possible, the therapist makes use of compensatory strategies to make the patient perform as independent as possible within the present functional level. The goals can be briefly described as:

1. Improving balance and postural reactions against external stimuli and gravitational changes.
2. Improving and increasing postural stabilization following the development of joint stabilization.
3. Developing accuracy of limb movements incorporating placement of both upper and lower extremity when performing all functional activities.
4. Developing independent and functional gait including clearance of obstacles,
5. Increasing the patient's independence while performing activities of daily living.

Main principles of training

1. Whatever be the pathology, treatment requires repetitions of a task and task sequence which will help in execution of both slow and rapid movements.
2. Exercises should be practiced consciously at first, and in later stages should be followed by automatic exercise activities.
3. Exercises should progress from simple to complex activities.
4. Activities should be practiced first with the eyes open and later with the eyes closed.
5. After achieving proximal tonus and stabilization, the coordinated movement of the distal segments should be taken into consideration.
6. Compensation methods and supportive aids and equipment should be employed whenever necessary.
7. Treatment should be supported by an appropriate home exercise program and sports activities.

Measurement and assessment

Evaluation of a patient with cerebellar lesion should include the following:

1. Bed mobility and posture.
2. Ability to sit up from a reclining posture.
3. Maintenance of sitting posture on surfaces normally used by the client.
4. Ability to stand up from a sitting position and transfer to and from a commode as if the person was within the home environment.
5. Maintenance of standing posture.
6. Ambulation and the environment within which the person will ambulate.
7. Ability to dress, groom and eat as normal daily living activities.

Performance can also be measured by:

1. Assistance required to perform the activities.
2. Level of effort involved.
3. Time required to complete the activity.
4. Potential Hazards to the client.
5. Unusual accompanying movements.

Although the observational methods and scales mostly designed to assess balance are easy to use,their ability to provide standardized measurements is limited, and the results can vary depending on the person who has done the observation.(9) Though computerized systems are highly reliable, they are costly systems which require working within the laboratory environment. Balance assessment tools frequently used by physiotherapists are shown in Table 2.

Table 2: Methods of Balance Assessment

Assessment Tool	Purpose of Tool
External Perturbation Test - Push and Release test (Jacobs et al.2006, Valkovic et al. 2008)	Static balance External Perturbation
Test - Pull test (Hunt&Sethi 2006, Munhoz et al.2004, Horak et al. 2005)	Static balance in different sensory conditions

Clinical Sensory Integration Test (Smania et al. 2008, Chaudry et al.2004) conditions	Dynamic balance in different sensory
Sensory Integration Test of Computarised Dynamic Posturography (Mirka&Black 1990, Jackson et al. 1995, Cham et al.2006)	Static and dynamic balance
Static and Dynamic Posturography (Mohan et al. 2008, Federica et al. 2008, Buatois et al. 2006)	Static balance
Single Leg Stance Test (Soyuer et al. 2006, Mann et al. 1996)	Static balance
Functional Reach Test (Martin et al. 2006, Jacobs et al. 2006)	Functional static and dynamic balance
Berg Balance Scale (Yelnik&Bonan 2008, Ryerson et al. 2008, Enberget al. 2008)	Functional static and dynamic balance
Five Times Sit to Stand Test (Buatois et al. 2008)	Functional dynamic balance and gait
Time Up and Go Test (Zampieri& Di Fabio 2008, Vereeck et al. 2008)	Gait and functional dynamic balance
Dynamic Gait Index (Herman et al. 2008, Chang et al. 2008)	Dynamic balance and gait
Tandem Walking (Ravdin et al. 2008)	Dynamic balance
Four Square Step Test (Blennerhassett&Javalath 2008)	Dynamic balance

Measurements such as gait duration, step length, step width can be used apart from these tests. Moreover, self-perception scales filled in by the patient such as Dizziness Handicap Inventory, Activity Specific Balance Confident Scale and scales for daily living activities such as FIM™ and Barthel Index can be employed to assist in assessment methods (Wrisley & Pavlou 2005).

There are a limited number of scales which have been developed to assess truncal ataxia and extremity ataxia together, and tested for validity and reliability. (Table 3)

Table 3: Scales of Ataxia

Assessment Tool	**Purpose of Tool**
International Cooperative Ataxia Rating Scale (D'Abreu et al. 2007)	Evaluating truncal and extremity ataxia, gait ataxia, nystagmus and talking
Scale for Assessment and Rating of Ataxia (Yabe et al. 2008)	Evaluating truncal and extremity ataxia, gait ataxia and talking
Ataxia Functional Composite Scale (Assadi et al. 2008)	Evaluating gait speed, upper extremity ataxia and visual acuity
Nine Hole Peg Test (Lynch et al.2005)	Evaluating upper extremity ataxia
Computer Graphics Tablet (Erasmus et al. 2001)	Evaluating upper extremity ataxia
Brief Ataxia Rating Scale (Schmahmann 2009)	Evaluating truncal and extremity ataxia, gait ataxia, nystagmus and talking
Friedreich's ataxia impact scale (Cano 2009)	Speech, upper limb functioning, lower limb functioning, body movement, complex tasks, isolation, mood, self perceptions
Composite cerebellar functional severity score (du Montcel 2008)	Upper limb functions

Physical therapy approaches

A physical therapy program is tailor made from the interpretation of the measurement and assessment results. The contents of the treatment program can vary depending on the type and characteristics of ataxia. For instance, while approaches which improve proprioception and incorporate visual aids are used more commonly in patients with sensory ataxia, stabilization training is more important to reduce truncal and extremity ataxia in patients with cerebellar ataxia. The patient with vestibular ataxia should be given habitation exercises in order to reduce vertigo, and also vestibulo-ocular, vestibulo-spinal reflexes should be stimulated to improve balance. In some cases, a problematic condition which requires the use of a number of approaches, such as mixed ataxia, may arise. In such cases, Physiotherapy helps in improving the gait, balance and trunk control of people with ataxia and reduce activity limitations and increase participation.(10)

Cerebellar signs are seen in many pathologies like multiple sclerosis, head injury, cerebellar stroke, brain tumour, cerebellar degeneration, central vestibular dysfunction and Friedreich's ataxia and interventions are to be tailor made for individuals varying in terms of type, intensity, duration and frequency.

Interventions like PNF, Frenkel's exercises, dynamic training of postural stability with task and activity focus, gait and balance training; along with strengthening and flexibility can be included. Therapeutic equipment was often provided to support function.

Dynamic task practice for Strength and flexibility would improve stability and gait. (11)

The use of orthotics and devices along with movement retraining reduces the degree of freedom but it enables the patient to do the ADLs better.

The classification of treatment applications can be briefly described as follows:

Head and trunk control:

A patient with postural instability needs to be assisted in maintaining posture in all activities in which the patient engages. Learning to sit on noncompliant surfaces versus compliant surfaces requires the cerebellum to process more proprioceptive input in the former and more vestibular input in the latter.

Biofeedback can be used to promote upright head position in severely affected patients for example, wearing helmet provides visual and auditory clue which can be used when the vertical position of the head is not maintained.

Stretch reflex activation may be used to increase muscle contraction in the initial phases of treatment as it is required to maintain muscle contraction in the core trunk muscles.

- Firstly, the proximal muscles and stabilization of the trunk should be improved (Edwards 1996). It is appropriate to use the mat activities of the PNF techniques.
- Bridging
- One leg bridging.
- Abdominal and back strengthening exercises.
- Long leg shifting in circles and in forward backward directions.
- Quadrapud
- Wt shifting in Quadrapud.
- Hip extension in Quadrapud.
- Side sitting in Quadrapud.
- Crawling
- Kneel standing.
- Trunk rotation in kneeling.
- Kneel walking.
- Perturbation (pushing and pulling in different directions).
- Weight transferring and functional extension
- Sitting balance: When the patients can hold their heads up and can hold some trunk control in a static position then they can be challenged to various sitting postures which will challenge their balance which includes treatment on the edge of the mat table or in a chair without back or arm support like able to get up from sidelying position and side shifting in bed,
- Joint approximation has been used to promote trunk stability.
- Rhythmic stabilization for trunk rotation can be used to sustain contraction of the trunk muscles where it is to give more of sensation of stability
- Rotation of the trunk also may be valuable in gaining stability.

Bridging

One leg bridging

Long leg shifting in forward direction

Long leg shifting exercises

Quadrapud

Weight shifts in Quadrrapud

Side sitting in Quadrapud (a)

Side sitting in Quadrapud (b)

Crawling

Trunk rotation in kneeling (1)

Trunk Rotation in kneeling (2)

Getting up from sidelying (2)

Getting up from sidelying position (1)

Side shifting at edge of bed in sitting unsupported

Side shifting at edge of bed in sitting unsupported (1)

Side shifting at edge of bed in sitting unsupported (2)

Forward reachouts in sitting

Ball Bouncing

If a patients cannot sustain a contraction which requires co-activation of the trunk muscles, a pattern of slow-reversal -hold over a steadily decreasing range of trunk rotation can be used.

- Push-pull activities with hands joined or with the cane or pole are helpful as well in promoting coactivation needed for stability.
- Therapeutic Ball is also a good activity as gentle bouncing on the ball may promote activity in the small trunk extensors thereby regaining awareness of being vertical.
- Weight shifts in all directions while sitting with both the hands support progressing to no support from the hands further with arms overhead and the trunk rotated as these positions are used in our daily activities.

Techniques to improve Limb Ataxia

Exercises are designed in a manner to provide fixation by establishing balance between the eccentric and concentric contractions within the multi-joint movements of lower extremities and the upper extremities in particular. During the performance of these exercises, it is important to establish slow, controlled and reciprocal multi-joint movement and stabilization, thus Frenkel's coordination exercises were introduced.(Edwards 1996, Danek 2004). Actively repeated contractions similar to PNF can be utilized on their own or by combining them with Frenkel's coordination exercises (Armutlu et al. 2001). While these two types of exercise are effective in cases with mild extremity ataxia, they can be insufficient in severe cases. In such cases, rhythmic stabilization and combination of isotonic techniques are more effective than PNF (Adler et al.2000).

Use of proprioceptive neuromuscular facilitation (PNF) patterns of rhythmic stabilization or slow-reversal-hold for the lower extremities will allow clients to ambulate with better control.

Functional tasks involving the arms can be preceded by PNF patterns for the arms.

Frenkel's exercises are effective for some patients and can be performed in the supine, sitting or standing positions. Each activity is to be placed slowly, with the patient watching the extremity carefully. When the client has gained reasonable control of one activity, she or he should proceed to the next. The patient starts with moving a limb with support, to moving without support at one joint of a limb, to moving the limb as a whole. Fig 6.18

Frenkel's exercises in Lying

Frenkel's exercises in lying (1)

Frenkel's Exercises in Standing

Frenkel's exercises in Standing (1)

Frenkel's exercises:

1. Supine:

a) Flex and extend one leg, heel sliding down a straight line on table.

b) Abduct and adduct hip smoothly with knee bent, heel on table.

c) Abduct and adduct leg with knee and hip extended, leg sliding on table.

d) Flex and extend hip and knee with heel off table.

e) Place one heel on knee of opposite leg and slide heel smoothly down shin toward ankle and back to knee.

f) Flex and extend both legs together, heels sliding on table.

g) Flex one leg while extending other leg.

h) Flex and extend one leg while abducting and adducting other leg.

2. Sitting:

1. Place foot in therapist's hand, which will change position on each trial.
2. Raise leg and put foot on traced footprint on floor.
3. Sit steady for a few minutes.
4. Rise and sit with knees together.

Standing:

1. Place foot forward and backward on a straight line.
2. Walk along a winding strip.
3. Walk between two parallel lines.
4. Walk, placing each foot in a tracing on floor.

The sequence used will depend upon the way the patient sleeps as well as the client's weight, side of involvement, underlying muscle strength, age and comorbid conditions. The patient should be provided with a variety of ideas for rising from the bed. Methods for rising from the floor should be taught and fall can also be practiced with lying to sitting.

Independent Transfers :

If the clients have adequate sitting balance but are not considered candidates for safe ambulation, they should be taught as many independent transfers as possible. Transfers can be taught with the caregiver to increase follow-through and safety at home. A sliding transfer from a wheelchair to

another chair or bed will be safest. Swivel sliding boards may assist the caregivers for transfers in and out of a car. A trapeze over a bed or bars in the bath may increase the level of independence if the accuracy of limb movements allow such activity.

Progressing towards for Ambulation:

If the goal is to progress the patients toward ambulation, a series of preliminary activities would be beneficial before they attempt to stand. These activities may include exercises such as bridging. The patient may practice transferring from sit to stand many times through the day. A simple way to increase strength and practice in this activity is to have the patient stand after sitting one to five times, she or he sits down or stands(expect for when the patient is transferring to the toilet for hygiene.

Moving from sit to stand and stand to sit can be further progressed by using their hands on the table(before and after meals) or with the use of a walker or wheelchair.

If the patient has been in a wheelchair or has been in bed for a long time, the therapist may need to prepare the person's cardiovascular system for being upright by placing him or her on a tilt table. Standing activities may be started in the parallel bars.

When standing up from sitting, the patient need to slide forward in the chair and flex their trunks considerably, placing the center of gravity over the feet. The trunk and legs should be extended only after gaining balance on the feet. This may be the most difficult step for an ataxic individual, who will either lean too far forward or extend the trunk too early and drop back into the chair.

Once upright, the person should practice balancing which can be reinforced by approximation through the hips and shoulders. Weights have been used to increase stability in standing and walking. A study by Widener indicated that small weights carefully placed for the individual was effective in improving standing balance.

In the standing position, weight transferring onto the front, back and sides, narrowing the support surface and balance training in tandem position, balance training on one leg should be performed thereby challenging their stability. Fig 6.19-6.26

The best indicator of dynamic stabilization / balance is gait. Therefore, gait training should be given including the following applications : walking on two narrow lines, tandem gait, backward gait, slowed down gait (soldier's gait), stopping and turning in response to sudden directions, flexion, extension and left-right rotations of the head and balance board exercises.

It is hoped that the patient can learn to come to and maintain standing without pulling on the bars, however, for some people this will be impossible. Those individuals who rely on the bars will not become independent in ambulation but may with assistance of another person or an assistive device, be able to get up and walk.

Once standing and stable, the patient needs to practice walking on a level surface as well as walking over obstacles and uneven surfaces to be considered as functionally independent in ambulation.(21)

Ambulation:

When the patient begins to walk within the parallel bars, he or she will need precise verbal feedback as to step length, body rotation, accessory movements and trunk position. Ambulation over the rungs of a ladder or lines on the floor can be used to increase visual clues and feedback regarding foot placement.

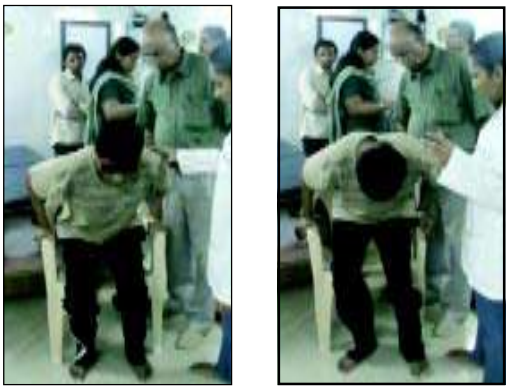
A visual clue will be needed to prevent the patient from walking too far into the walker and falling backward. A four-wheeled walker may be easier to manipulate and does not need to be lifted just pushed forward.

Crutches or canes may be used but require reciprocal movement of the arms and legs with appropriate timing and placement. The patient may actually do better using a walking stick for support. One can measure the patient progress in ambulation by the number of times the patient loses his or her balance in a treatment sessions, frequency of a specific error, The distance ambulated or the level of assistance needed.

The more closely the treatment activities mirror the patient's life, the more likely the treatment will be successful.

Relaxation and Biofeedback

Guercio et al (1997, 2001) describe two case studies of individuals with severe ataxic kinetic tremor following a TBI that used behavioural relaxation training with EMG auditory biofeedback to reduce tremor severity and improve functional performance. Findings reported benefits in functional performance and reduction of tremor.



Sit to stand from the chair

Getting up from low stool

Getting up from sidelying

Balancing exs in standing

Tandem Walking

Crossing the Obstactles

Balance board exercises.

Baram and Miller (2007) investigated the effect of auditory biofeedback for 14 people with MS and gait dysfunction due to cerebellar ataxia. Positive results were reported in terms of speed and steplength but not maintained for doing ADLS

Interventions used to improve tremor in upper limb:

Lesions affecting the cerebellar hemispheres give rise to ipsilateral limb symptoms including tremor in addition to dysynergia, disdiadochokinesia and rebound phenomenon.

An action tremor occurs during movement i.e. is produced by voluntary contraction of muscle. Postural tremor occurs when voluntarily maintaining a position against gravity e.g. holding an arm out straight.

Kinetic tremor occurs during any type of voluntary movement and further can be subdivided into:

1. Simple kinetic tremor which occurs during voluntary movements that are not target-directed (e.g.flexion / extension or pronation / supination).
2. Intention tremor, which occurs during target directed, visually guided movements (e.g. finger-nose test), and worsens at the terminal phase of the movement as the target is approached (Deuschl et al, 1998).

In addition to affecting activities of daily living (Feys, et al, 2004) the psychosocial consequences of upper limb tremor can be significant (McGruder et al, 2003).

The use of weights and the carryover when the weights are removed will require more careful investigation may indeed be patient or disease specific..The clinician must analyze whether the weights are increasing joint approximation in a closed chain or increasing joint distraction in an open chain. Closed chain enhancement may help the cerebellum, while open-chiar distraction may increase the imbalance between agoinst and antagonist during a movement pattern.(12)

Wrist weight cuffs

Wrist weighting is an intervention used to reduce upper limb tremor, however mixed effects are reported. Use of supportive devices enables the

Coordination exercises for upper limb

Picking the Beads

Picking the Beads1

Picking the beads 2

Rolling the theraband

Cutting the page

Hand functions-Picking up

Putting the beads

patient to function more easily within his present functional level. In cases of severe ataxia, suspending weights from the extremities and the use of weighted walkers can be preferred (Gibson-Horn 2008).

Cold Therapy

It has been reported that cold therapy has significant beneficial effect in the cases of tremors in MS in upper limbs.

Interventions used to improve proprioception:

The aim is to increase proprioceptive input by mechanically stimulating the joint surfaces, muscles and tendons, and decreasing postural instability by improving body awareness. There are many approaches that can be used for this purpose. These are: Proprioceptive Neuromuscular Fascilitation (PNF), rhythmic stabilization, slow reversal techniques (Adler et al. 2000, Gardiner 1976), resistive exercises (DeSouza 1990, Arai et al. 2001), use of Johnstone pressure splints (Armutlu et al. 2001), gait exercises on different surfaces (hard, soft, inclined surfaces) with eyes open and closed and balance board exercises.

Vibration has been a frequently used application where it is directly applied to the muscle and tendon, and also it can be applied by exposing the whole body to vibration (Schunfried et al. 2007, Hatzitaki et al. 2004, Semenova 1997).

In addition yoga, and body awareness exercises can be included in the program.

Theraband exercises can be used to maintain resistance and improve joint approximation.

Interventions used to improve vestibular dysfunction:

Since dizziness accompanies balance dysfunction in vestibular problems, repetitive head movements and Cawthorne and Cooksey exercises (Dix 1979) are of great importance. A vestibular exercise program consists of repetitive, progressively more difficult, eye, head and body movements designed to encourage movement and facilitate sensory substitution.

Exercises

Exercises should be encouraged as part of health promotion.Exercise should be tailored by exploring several different options as well as building motivation and sustainability into the exercise prescription (Dean, 2009;Rhodes and Fiala, 2009)

1. Endurance/aerobic Training

Fillyaw and Ades (1989) investigated the physiological adaptation to aerobic training in Friedreich's Ataxia where training took place over a period of nine weeks and consisted of 27 ECG monitored, bicycle ergometer sessions of 20-25 minutes of continuous cycling at training level intensities.Prior to commencement of training the participant, who had no history of cardiac symptoms, underwent a cardiovascular examination and echocardiogram that confirmed normal cardiac size and function. Increases in cardiorespiratory and work measures demonstrated clinically important physiological adaptations to aerobic conditioning suggesting that people without cardiomyopathy, aerobic training offers a means of promoting activity and reducing deconditioning which may provoke functional as well as other health benefits.

2. Hydrotherapy and Swimming

Cook (2007) advocates the use of hydrotherapy and swimming for people with ataxia because water activities offer risk and challenges providing freedom of movement and may be beneficial for speech. Hydrotherapy is also considered to offer beneficial effects on health related quality of life. No studies directly evaluate the efficacy of hydrotherapy for people with ataxia and can be taken as a form of exercise.

3. General fitness training, Yoga and Pilates

Benefits of general fitness training, yoga and Pilates for people with ataxia can help in maintaining strength, flexibility and balance thereby resulting in overall improvements in psychosocial aspects.

ROLE OF OCCUPATIONAL THERAPY IN CEREBELLAR ATAXIA

Occupational therapist form an important part of the multi-disciplinary care team in ataxia. OT is an important intervention for patients with progressive neurological conditions in maintaining independence and quality of life and enable people to perform simple tasks that are meaningful to them. (13)

Occupational therapist should use assessment and outcome tools that measure the persons satisfaction with the performance of an activity, since use of tools that measure only impairment would not demonstrate the effectiveness of OT

intervention(14). Evidence suggests that people with ataxia may have a lower quality of life in the early and end stage of the condition. It is therefore important to recognize that even at the early stage of the condition, difficulties with roles and activity engagement may benefit from support. (15)At most ataxias are progressive an important consideration is proactive planning for future needs.

Patients with ataxia present with insufficient postural control and incoordination of multijoint movements resulting in postural instability leading to balance and mobility dysfunction. Some patient may also present with hypotonia and hyperkinesia i.e tremors, titubation which interferes in all their functional activities. Hence it is essential that occupational therapist addresses these issues

Management for Hypotonia:

Intervention techniques to increase tone for patients with hypotonia (flaccidity) can include quick stretch, tapping, resistance, approximation, and positioning. Patients typically also demonstrate weakness and at times it is difficult to differentiate between two. Strengthening exercises that do not overload the weak, hypotonic muscles are indicated. Postural instability is common problem. Intervention should be designed to improve postural stability in functional positions

Management to improve postural control

In patients with ataxia instability is associated with excessive postural sway, wide BOS, a high guard hand position or handhold, and loss of balance. The therapist can provide training through postures that demand increasing amount of upright(antigravity) postural control eg. prone- on-elbows to quadruped and finally sitting. As trunk becomes more stable, the patient is expected to assume active control in stabilizing the posture. For patients with ataxia, the PNF technique of stabilizing reversals (slow reversals) is appropriate

The therapist can have the patient stabilize while seating on a therapy ball. Gentle bouncing provides joint approximation through the verbal joints, facilitating extensors and an upright posture. For patients requiring more assistance, sitting control can first be practiced on a complaint surface placed on a platform mat or sitting on a ball with a ball holder. Task difficulty can be increased by reducing the BOS.

Aquatic therapy can also be used to enhance proprioceptive loading. The water provides degree of unweighting and resistance to movement. This can be quite effective in reducing hyperkinetic movements and enhancing postural stability. For example, a patient who demonstrates significant ataxia may be able to sit or stand in the pool with minimal assistance while these same activities outside the pool are not possible.

Although active movement is the goal, assistance may be required during initial movement attempts for both the dynamic movements' as well as the stabilizing body segments. Specific task- oriented training (eg. Reaching, stepping) are more motivating, especially if the task is important to the patient.

Therapy ball activities are effective in developing dynamic stability control. For example, the patient sits on a ball and gently moves the ball side-to-side, forward-backward, or in a combination (pelvic clock motions or the patients sits on ball while performing voluntary movements of the arms or legs alternate legs or arm raises)

Reachouts in standing

E.g. reaching for pegs / kicking a football.

Progression is from unilateral to bilateral and finally to reciprocal limb movements. Voluntary trunk motions can be practiced while sitting on the ball (e.g. head and trunk rotations are forward / backward leans). Resistance can be introduced by using elastic resistance bands or weight cuffs on the ankle or wrists. Difficulty can also be increased by adding a second task (dual task training) such as catching and throwing a weighted ball, balloon volleyball, or kicking a ball. Group activities can be introduced when patients can safely perform each of the activities individually.

Management for Ataxic Movements (16):-

- Control of ataxic movements can be achieved through proprioceptive loading and light

resistance. Resistance can be provided by application of elastic resistance bands / light weights to stabilize movements. Weight cuffs, weighted boots or weighted jacket / belt can reduce tremors of limbs / trunk.

- The extra weight will also increase energy expenditure and must therefore be carefully balanced against increased fatigue it might cause.
- Weighted canes / walkers can be used to decrease ataxic lower extremities movements that interfere with use of assistive device during ambulation.
- Weighted spoons and fork can be used to enhance eating for patients with significant tremor, these devices may mean the difference between dependent and independent function.
- A soft neck collar can be used to stabiles head and neck tremors.
- All these strategies however are temporary and compensatory.

Strategies to improve Safety (16):

Prevention of falls for the patient with balance deficiency is an important goal of therapy. Lifestyle counseling is important to help recognize potentially dangerous situations and reduce the likelihood of falls. For example, high risk activities likely to result in falls include turning, sit to stand transfers, reaching and bending over. Patients should also be discouraged from early hazardous activities such as climbing on step stools, ladders, and chairs, or walking on slippery surfaces. The education plan should stress the harmful effects of a sedentary lifestyle. Patients should be encouraged to maintain an active lifestyle, including a program of regular exercise and walking. Medications should be reviewed and those medications linked to increase risk of falls (e.g., medications that result in postural hypotension) should be addressed.

Assistive devices should be used assist to balance when necessary.

Falls management

Falls may occur in any area that a person mobilizes. At home, the person should be taught fall recovery techniques and where appropriate, consider the use of community care alarms such as pendant alarms, and techniques to avoid further injury such as pressure sores while waiting for help to arrive. OTs should consider joint assessment with or referral to a physiotherapist, and referral to a falls programme / group locally. If there is a family member or carer involved, the occupational therapist and physiotherapist should consider the safety of the carer.

Fall Prevention Strategies: Modifying the Home Environment

- **Adequate lighting** is essential. Both low light and glare can be hazardous, particularly for the elderly. Glare can be reduced with translucent shades or curtains.
- **Light switches** should be positioned at the entrance to a room and fully accessible.
- **Carpets with loose edges** should be tacked down. Scatter or throw rugs should be removed.
- **Furniture** that obstructs walkways should be removed or repositioned.
- **Chairs** should be adequate height and firmness to assist in sit-to stand transfers. Chairs with armrests and elevated seat heights may be required.
- **Stairs** are the site of many falls. Ensure adequate lighting. Contrast tape using bright warm colors (red, orange, or yellow) can be used to highlighting steps. Handrails are important for safety on stairs and, if not present, may need to be installed.
- **Grab bars or rails** reduce the incidence of falls in the bathroom. Nonskid mats or strips in the bathtub along with a tub or shower seat can also improve safety. Toilet seat can be elevated to facilitate independent use.

When ADL activities are difficult to manage or perform OTs support people by changing and adapting their relationship with their physical and social environment to develop new valued activities and role. Focus of occupational therapist on engagement in activity, rather than the disorder is important in progressive conditions. OT intervention should focus on goals that support the person and carers and improve their quality of life.

The main focus of occupational therapist is participation in activities rather than focus on the progressive condition of disorder thus enhancing the present lifestyle and improving quality of life.

Evidence suggests that people with ataxia may have a lower quality of life in the early and end stages of the condition (15). As most ataxias are progressive an important consideration is proactive planning for future needs.

Common Occupational Therapy management for Cerebellar Ataxia

Self care and toileting

Treatment planning includes reducing the impact of excessive movement and helping the patient to increase their independence. Mainly people with ataxia face difficulty in balance, so support bar can be used to provide stability during transfers. Dressing and undressing to toilet and personal hygiene are other problems encountered during toileting

Practical Suggestions

- Encourage bath in seating position with support for the back and arms.
- Use of hand held shower instead of using taps.
- Use of rail bars in toilet to minimize risk of fall
- Instead of Zip or button use of Velcro.

Eating and drinking

Due to multiple difficulties faced by ataxic patients Feeding needs to be considered for effective eating and drinking. Due to multiple impairments impacting on safe and effective eating and drinking feeding needs to be considered. Proper positioning helps to maximize posture and support core stability, which helps to reduce the impact of excessive limb movements. Occupational therapist works in tandem with the speech and language therapist to find appropriate feeding solutions. (20).

Feeding Assessment (Use of oral -motor / feeding rating scale)

Practical suggestions

- All the utensils and necessary things while eating should be organized to increase independence.
- Use of Plate guards, and non-slip matting to prevent unnecessary movement of plate.
- Use lidded / insulated cups or cups with straws for drinking, especially hot liquids - such as tea and coffee
- Use of weighted spoon or fork while eating

Food preparation

Preparing food is one of the common concerns in the early stages of ataxia due to poor imbalance and Inco-ordination. OTs should carry out an activity analysis of food preparation tasks and suggest a variety of methods and aids that may compensate for difficult or unsafe aspects of tasks. Kitchen platform should be on wheelchair level so cutting vegetables, cooking and doing other work will be easier and independent.

Household management

In ataxia, people are cognitively able to manage the home but may have difficulty in physically doing heavy housework such as cleaning floor and washing clothes. Fatigue is also main concern, so people should use energy conservation techniques organization and adaptive devices.

Practical Suggestions

- Use Kettle tipper devices can help making drinks safer
- Using a travel mug with a lid can sometimes assist with carrying a drink
- Waist height ovens; use of full-length oven gloves; sliding food to a level surface (or level trolley) rather than lifting
- A microwave oven can provide a safer alternative to standard ovens
- Chopping boards with an attached cutting blade can be safer than a separate knife
- A food processor can help with slicing or chopping vegetables.

Bed, chair and toilet transfers

Ensure that the height of the chair and bed should be on same level for safe transfer and the hip and knee angle is at 90 degrees and that the feet are flat on the ground. While transferring it should be in noticed that the chair is stable and armrest is of suitable height and the patient should be in a position to do a push up. Educate the patient and care giver on sit to stand techniques and adequate training of transfers should be taught. In particular, provide the most support possible for safety reasons.

Indoor Mobility

Indoor mobility should be ideally assessed in the environments that the person uses. According to

patient needs OTs should modify the home as well as work place environment. Use of railings, grab bars, walking frames should be considered to increase independent mobility in daily living. OTs should consider the use of walking aids in the home and other environments. eg a walking frame to the toilet and then the use of hand rails inside the toilet. If wheelchair mobility is considered the environment should be modified accordingly to ensure access to the areas they need to. For eg. Modify door widths, arrangement of furniture, ramps/ lifts, work table height.

Practical Suggestions

- Consider the height of the bed and location within the room
- Mattress variators, or profiling beds may be of benefit
- Ensure surfaces are the optimal height to ensure the most efficient and safest transfers
- Firmer mattresses will aid bed mobility
- If patient is wheelchair bound or bedridden and mobility is severely restricted, pressure care needs should be considered.
- Wear tight fitting clothing instead of loose clothing and avoid carrying items while walking to avoid imbalance while walking.
- Use of trolleys to carry items, especially food, drinks and heavy items at work or in the home
- Use of nonskid mats at floor and remove loose electrical cables to avoid risks to mobility in the home environment
- Good lighting will help optimize performance of tasks and decrease the chance of further complications.

Outdoor and community mobility

In ataxia, incoordination and imbalance while walking is main problem so mobilising outdoors is often difficult for the person with ataxia. Educate the carer and the person with ataxia about energy conservation techniques like rest regularly while walking outdoors and use of walking aids, use of wheelchair for outdoor can help to reduce fatigue or maximize safety. If patient is using wheelchair, proper assessment for optimal seating position should be done, like use of cushions and back support should consider. Ongoing assessment for seating position can be useful in all the stages. A compromise between optimizing function and providing adequate support is important.

Practical Suggestions

- Public transport and rail providers offer subsidized fares and can provide a meet and greet service
- Outdoors motorized scooters or wheelchairs can maximize independence

Handwriting

Handwriting is another area of concern in progressive ataxia

Practical suggestions

Use of weighted wrist cuffs and weighted pens may be helpful in reducing the tremors.

- Proper posture is advisable to maximize independence while writing
- Use of Dictaphones or voice-activated computer software to compensate for
- handwriting and speed difficulties

Computer use

Patients with Cerebellar Ataxia gradually find difficulty in using computers. If the patient has dysarthria, consideration of voice-activated software should be carefully thought about.

Practical suggestions

- Keyboard and mouse modifications can be made to adjust the sensitivity and speed of response
- Alternative mouse such as a Tracker ball can be helpful
- Smaller keyboards or key guards may help-check it
- Location and setup of computer equipment should be easily accessible.

Work

Patient should consider continue to work as long as possible. When faced with difficulties. Occupational Therapist should conduct onsite evaluation and suggest or modify environment accordingly. Work hours, Transportation, seating arrangement, type of equipment etc should be considered.

Practical Suggestions-

- Adequate rest periods should be provided between working hours.
- Ergonomically appropriate seating
- Use of lift instead of stairs
- Educate patients about safe use of equipments

Leisure

If the patient has loss leisure role such as participation in sport, gardening, painting then occupational Therapist should consider alternate leisure activities like Watching on television, painting with adaptive devices such as stencils, block stamping, care of potted plants. Active participation in hobbies should be encouraged. Outings, get-togethers with loved ones can prove to be therapeutic.

Practical suggestions

- Educate the person and carer on allowing the car door to be opened fully and to consider the height of the transfer being undertaken
- Ensure the person sits their bottom down first before moving their legs into the car
- Use of transfer board with transfer mat and if the car seat is low, use firm foam

Cushions for safe transfer

- Choose a model of car that optimizes transfers, door access and storage space

Activity Analysis & Synthesis

A key skill of an OT is to analyze the component parts of an activity in order to use it purposefully, meaningfully and therapeutically (Finlay, 2004).

Activity Summary of Expected Performance

This activity analysis format is taken from Lamport, Coffey, Hersch (2002).

1. Name of activity:
2. Brief Description of the activity:
3. Tools / Equipment (non-expendable), Cost and Source:
4. Materials / Supplies (expendable), Cost and Source:
5. Space / Environmental Requirements:
6. Sequence of Major Steps
7. Precautions:
8. Special considerations:
9. Acceptable criteria for completion:
10. Performance Components

Components	Required ?	Required for	Synthesis
A. Sensorimotor			
Sensory			
Sensory Awareness			
Sensory Processing			
Tactile			
Proprioceptive			
Vestibular			
Visual			
Auditory			
Gustatory			
Olfactory			
Perceptual Processing			
Stereognosis			
Kinesthesia			
Pain Response			
Body Scheme			

Components			Required ?	Required for	Synthesis
		R-L discrimination			
		Form constancy			
		Position in space			
		Visual-Closure			
		Figure Ground			
		Depth Perception Spatial Relations			
		Topographical Orientation			
Neuromusculoskeletal					
		Reflex			
		Range of motion			
		Muscle Tone			
		Strength			
		Endurance			

Components	Required ?	Required for	Synthesis
Postural control			
Postural Alignment			
Soft tissue Integrity			
Motor			
Gross coordination			
Crossing the midline			
Laterality			
Bilateral Integration			
Motor control			
Praxis			
Fine coordination / Dexterity			
Visual-Motor Integration			
Oral-Motor Control			

B.Cognitive Integration and cognitive components				
Components	**Required ?**		**Required for**	**Synthesis**
	Level of arousal			
	Orientation			
	Recognition			
	Attention Span			
	Initiation of activity			
	Termination of activity			
	Memory			
	Sequencing			
	Categorisation			
	Concept formation			
	Spatial Operations			
	Problem Solving			
	Learning			
	Generalisation			

C. Psychosocial Skills and Psychological Components					
Components		**Required ?**		**Required for**	**Synthesis**
Psychological					
	Values				
	Interests				
	Self-concept				
	Social				
	Role performance				
	Social Conduct				
	Interpersonal Skills				
	Self-expression				
	Self-management				
	Coping skills				
	Time management				
	Self-control				

Psychological Aspects in CEREBELLAR ATAXIA

Problems due to Cerebellar damage:

When the Cerebellum is affected it leads to the following impairments: in executive function, visual-spatial analysis and selected deficits in language skills as well as changes in personality and behavior. There may be major problems in multi tasking, planning and organizing. The cognitive flexibility, which the patient used to perform earlier but after the onset of the disease they may require conscious effort and new strategies. The patients may suffer from mood changes like depression, apathy, irritability and limited frustration tolerance.

Over 65% of individuals with cerebral ataxia develop mood disorders at time after the onset of their neurological illness, usually a form of depression. While low mood is usually a part of clinical depression, some or all of a long list of other symptoms are also present and may be more prominent than low mood: loss of interest in usual activities, excessive anxiety or agitation, irritability or changes in sleeping patterns, changes in appetite r sexual drive, etc. Episodes of depression typically last weeks to months, but may be longer or shorter. This type of depression is caused by abnormalities in brain functions and is not a psychological reaction to adverse events in a person's life.

Treatment: All treatment efforts must first start with education. It is essential that individuals with the disease and their family members must be aware of the psychiatric and the cognitive problems that may arise during the course of their illness. Just like they experience trouble with balance or coordination, psychiatric and cognitive problems are a part of the disease and are not the fault of the affected individual, family members or health care providers.

The recognition of depression is essential, since among all the other complications of the disease, this may be the most treatable. The approach to the treatment is multipronged and this includes antidepressants, support and environmental changes. Antidepressants are very effective in treating depression i.e. Serotonin specific reuptake inhibitors (SSRI's), of which Prozac (fluoxetine) is the most famous example. It is prudent to start with a relatively low dose and advance the dose relatively slowly, since individuals with neurodegenerative disorders are more prone than other people to become confused if drug doses are too high or are advanced too fast. After 1 or 2 months of no response to one antidepressant at a typical dose it is reasonable to switch to another agent. Helpful environmental interventions often involve establishing a structure for the patient, including setting regular hours for sleeping, eating, exercising and getting out of the home. Meanwhile, the patient can be freed from burdensome responsibilities, which then can be gradually reinstated as the patient improves. As always, treatment must be individualized. Unlike depression, the goal for management of personality change or impaired cognition is not elimination of the condition, but rather adaptation to it. Medicines, typically antipsychotics drugs (such as haloperidol, resperidone, or olanzapine) or mood stabilizers (such as valproate or gabapentin) may prove useful in minimizing moodiness or irritability. Apathy may improve with stimulants (such as amphetamines) or other agents affect the chemistry of the dopamine system (such as amantadine). Establishing routines that emphasize the strengths of the affected individual often prove very helpful in managing personality change and cognitive impairment. For instance, a set schedule of regular daily activities, minimizing change from day-to-day, is often helpful for patients who react with anger, suspicion, or confusion to change. New routines with increased stimulation may be of some value in overcoming apathy.

Learning the proper response to irritability (ignoring it, switching to another topic of conversation, or distraction by introduction of a different activity) may help prevent escalation of temper into verbal or physical violence. Patients may partially compensate for mild cognitive impairment by keeping lists and allowing themselves more time for tasks. The stress on patients and families of more severe cognitive impairment or personality change may be decreased by removing the responsibility of the patient for performing tasks which are beyond their capacity (maintaining finances, supervising small children, driving, employment), while maintaining the patient's responsibility for tasks within their capacity. Such interventions require considerable delicacy. Developing, implementing, revising, and maintaining treatment for cognitive and psychiatric complications of cerebellar disease requires an accurate assessment of the patient's strengths and weaknesses, individualization of treatment, and

flexibility to change the plan as needed. Neurologists and primary care physicians with expertise in depression and personality change may assume overallmanagement of the treatment. In more complicated cases, or with neurologists or primary care physicians who are less comfortable with these issues, consultation with a psychiatrist with experience in the complications of neurodegenerative diseases may be necessary. Neuropsychological testing can provide precise information on cognitive strengths and weakness, and occupational therapists can assess an individual's capacity to perform both routine and more complicated tasks of daily living. Involvement of the family is essential in every step of the process of diagnosis and treatment.

To summarize, diseases that result in cerebellar degeneration are commonly complicated by cognitive impairment and psychiatric disorders, especially depression. These complications are probably more frequent and more severe when other brain regions in addition to the cerebellum are also involved. Based on our current knowledge of neurodegenerative diseases affecting other brain regions, both pharmacologic and non-pharmacologic interventions may prove helpful to patients with cerebellar degeneration and their families. Successful treatment is dependent on thorough evaluation and careful individualization to the strengths, weakness, and environment of each affected person.

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Informative Website for Cerebellar Ataxia

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2. www.ataxia.org.uk
3. National ataxia foundation- Support for research in ataxia

Ch.8 Multiple Sclerosis

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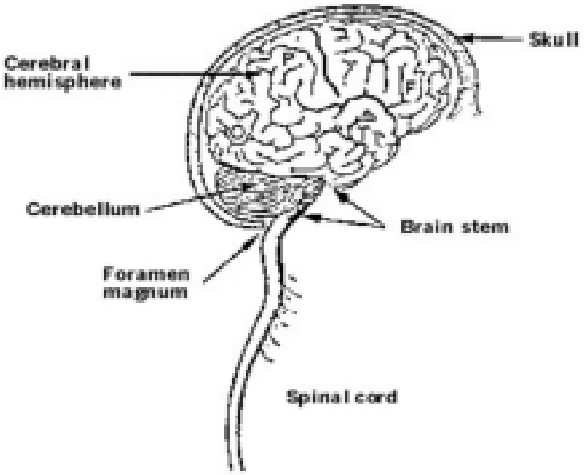
Multiple Sclerosis is an inflammatory relapsing or progressive disorder of CNS white matter and is a major cause of disability in young adults and affects between the ages of 20 and 40 years.(1)



It was identified and established by Dr. Jean Martin Charcot in 1868 who described it as Sclerose en Plaques.(2)

Epidemiology

MS is not a rare disease and it affects millions worldwide. Symptoms usually begin during adulthood, with peak onset around the age of 24. Approximately 0.3% of MS cases are diagnosed before age 15. Multiple sclerosis (MS) affects women more than men.

MS is most predominant in whites of Northern European ancestry and is rare in some races like African blacks and Eskimos. By studying people who move from one area to another, researchers have learned that individual risk changes based on location. They have concluded that some exposure in the environment increases the risk of for MS.

The incidence of MS is seen more in places which are farther from the equator, MS occur more prominently over there. Studies have shown that distribution of MS depend on areas of high, medium and low frequency.

The temperate zones of the northern United States, northern Europe, southern Canada, New Zealand, and Southern Australia are high frequency areas; with incidence reported of 30 to 80 per 100,000 populations. Southern United States and Europe and rest of Australia which are closer to the equator are medium frequency areas, with incidence reported of 10 to 15 per 100,000 population. Asia, Africa and South America are low frequency areas, with incidence reported of less than 5 per 100,000.

Perhaps the most incriminating evidence for the role of environmental factors in the development of MS is the changing risk with migration and the occurrence of MS clusters and epidemics. Immigrant populations tend to acquire the MS risk inherent to their new place of residence. Migration from high to low prevalence before the age of 15 lowers the risk, whereas migration after this age does not affect risk. Migration from high to low prevalence areas increases the risk of MS, but the effect of age is less clear.

Other environmental factors associated with the development of MS include cigarette smoking (odds ratio of 1.81), animal fat intake, and deficiency of vitamin D.

It has been observed that, place where person spends the first 15 years of life may determine to a greater or lesser likelihood of developing MS as opposed to where he or she lives at the time of diagnosis.(3) Minor respiratory infections relapses 27% of relapses in patients with established MS.(4)

A multitude of other environmental factors have been suspected to alter the risk for MS (cold climate, precipitation, amount of peat in the soil, exposure to dogs, and consumption of meat, and dairy products), but none has been verified to be an independent risk factor.

Etiology

The exact cause of MS is not known though it is believed to be caused by damage to the myelin sheath, the protective covering that surrounds nerve cells. When this nerve covering is damaged, nerve signals slow down or stop. The nerve damage is caused by inflammation which occurs when the body's own immune cells attack the nervous system. This can occur along any area of the brain, optic nerve, and spinal cord.

The presence of increased immunoglobulin (IgG) and Oligoclonal bands in CSF of 65 to 95% of MS patients suggestive of a precipitating infection eliciting an autoimmune response with resulting pathological changes.(5)

It is believed that viruses such as Epstein-Barr (mononucleosis), varicella zoster, and the hepatitis vaccine may be the cause of MS to till date. However, this belief has not been proven.(6)

The definitive genetic association in MS is with the serologically defined human leukocyte antigen (HLA) DR15, DQ6. This is one of the DR2 haplotypes, also known as Dw2 in cellular terminology and DRB1*1501, DQA1*0102, DQB1*0602 in molecular nomenclature.7, 8, 9

Other susceptibility genes likely contribute, possibility the T-cell receptor variable ? region and the IgG heavy-chain variable region (especially the VH2-5 gene)

Fine mapping data suggests that the DRB1 gene itself is responsible for a significant portion of this risk.(10)

Another theory suggests that, the immune system mistakes a portion of myelin protein for a virus that is structurally similar and targets it for destruction, known as Molecular Mimicry.(11)

Genetically identical monozygotic twins are more often concordant for MS than dizygotic twins (26% and 2.4% respectively), indicating a genetic component; however, even after following monozygotic twins past age 50 or using MRI data, less than 50% are concordant, suggesting a role for environmental factors.

Smoking is being implicated as risk factor in MS as it has been associated with increased blood-brain barrier disruption, and greater atrophy in multiple sclerosis.(12)

Pathophysiology:

In patients with MS, the immune response triggers the production of T-Lymphocytes, macrophages, & immunoglobulin's (antibodies). In turn, a command or trigger protein, the antigen is activated, producing autoimmune cytotoxic effects within the CNS.

The blood brain barrier fails and myelin sensitized T - Lymphocyte cells enter and attack the myelin sheath that surrounds the nerve. Myelin serves as an insulator, speeding up the conduction along nerve fibres from one node of ranvier to another termed salutatory conduction. It also serves to conserve energy for the nerve because depolarization occurs only at the nodes. Disruption of myelin sheath produces active demyelination, slowing neural transmission & causing nerves to fatigue rapidly. With severe disruption, conduction block occurs with resulting disruption of function. 13

Clinical Course of Multiple Sclerosis

Clinical course of MS is unpredictable. Patients, usually have one of the following clinical courses:-

Benign MS: Some patients get only rare relapses of their disease, in which patients' remains functional till 15 years after disease onset. It affects 20% of MS patients

Benign multiple sclerosis" is often temporary as apparently benign disease often becomes disabling.14

Malignant MS: In this type there is rapid progression of the disease which sometimes cause significant disability or death within short period after the onset

Relapsing/Remitting type: It is the most common type and is seen among 70% of MS patients and occur frequently, one to several times per year.

Primary Progressive MS: In the primary progressive form continuous worsening occurs with steady progression but not interrupted by distinct relapses. It affects 10% of MS patients

Secondary-Progressive MS: In this type relapsing remitting disease followed by progression with or without occasional relapse, minor remissions and plateaus.

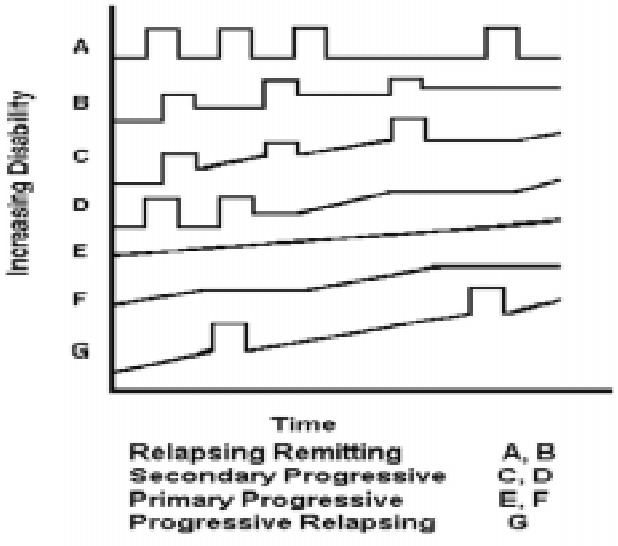
Progressive-relapsing

Progressive disease from onset but with clear, acute relapses that may or may not have some recovery or remission; Periods between relapses are characterized by continued progression.

That mainly seen in people, whose disease onset after 40 years.(15)

Signs and Symptoms

Signs and symptoms of MS vary depending on the location of specific lesions. Symptoms can develop quickly, within hours, or slowly over several days or weeks.



From the study of 697 patients with MS
Most Frequent Symptoms of MS

1. Fatigue 85%
2. Walking problems- 87%
3. Bladder problems- 65%
4. Pain and other sensations- 60%
5. Visual disturbances- 58%
6. Cognitive problems- 44%
7. Tremors- 41%

It has been concluded that-

- 50 to 70% of people with MS shows evidence of some cognitive impairment
- Majority have mild impairment
- 10% to 20 % have significant dysfunction

Common Symptoms in Multiple Sclerosis

Sensory Symptoms:	Hypoesthesia,numbness, Paraesthesia(tingling) Dysesthesias(burning)
Motor Symptoms :	Weakness or Paralysis Fatigue Spasticity Incoordination Intentional Tremor Impaired Balance Gait Disturbances.
Pain:	Lhermitte's Sign: Tingling in spine with limbs on neck flexion. Chronic Pain.
Visual Symptoms:	Blurred or double vision Diminished acuity or loss of vision Scotoma Nystagmus. Optic or trigeminal Neuritis
Bladder Symptoms: Bowel Symptoms:	Urinary Urgency, Frequency Nocturia Incontinence Urinary Hesitancy, Dribbling Constipation Diarrhoea Incontinence
Speech & Swallowing:	Dysarthria Diminshed verbal Fluency Dysphonia Dysphagia

Cognitive Symptoms:	Memory or recall problems Decreased attention, concentration Diminished abstract reasoning Diminished problem solving, judgment. Diminished speed of information processing. Diminished visual -Spatial abilities.
Emotional Symptoms:	Depression Pseudo bulbar Affect Anxiety
Cardiovascular Dysautonomia	Very rare.

Sensory Symptoms: This is the most common presenting manifestation in MS (21% to 55%) and ultimately develope in nearly all the patients. The symptoms like Hypoesthesia, numbness, Paraesthesia (tingling) Dysesthesias(burning) may occur in one or more limbs, trunk, face or in combinations. The more distinctive sensory relapses of MS consist of the sensory cord syndrome and sensory useless hand syndrome. The sensory symptoms may ascend to the trunk, producing a sensory level, or may involve the upper extremities. In most of the MS patients it has seen that they develop persistent sensory loss, usually consisting of diminished vibratory and position sensations in distal extremities.

Motor Symptoms: Motor symptoms like Weakness, Spasticity or Paralysis, Fatigue, Incoordination, Intentional Tremor, Impaired Balance, Gait Disturbances occur due to Pyramidal tract dysfunction. It can occur acutely or in a chronic progression with weakness of one or more limbs and facial weakness, leg stiffness that impairs gait and balance, or extensor or flexor spasms. Exercise or heat frequently worsens subtle deficits.

Muscle atrophy is usually due to disuse, but lesions of lower motor neuron fibers or of the anterior horn itself can cause pseudoradiculopathy with segmental weakness, atrophy, and diminished reflexes. Motor symptom manifestations of MS in 32% to 41 % of all cases and their prevalence is higher than 60% in long-standing MS.

Visual Symptoms: The initial symptoms of MS includ optic neuritis (ON) in 14 % to 23% of patients, and more than 50 % experience a clinical episode of optic neuritis during their lifetime. The most common manifestation is visual loss in one eye that evolves over a few days. Periocular pain, especially with eye movement, usually accompanies and may precede the visual symptoms. Bilateral simultaneous optic neuritis is uncommon in adults, but formal visual field testing reveals unexpected defects in the clinically normal eye in a substantial number of patients.

Examination shows an afferent pupillary defect, diminished visual acuity, subdued colour perception, and often a central scotoma.

There may be persistent visual blurring, altered colour perception, or Uhtoff's sign. MS patients without a clinical history of optic neuritis often have evidence of optic nerve involvement on funduscopic examination or visual evoked potentials. Recurrent optic neuritis can occasionally be seen without evidence for dissemination to other areas of the CNS.

Patients with frequent and severe optic neuritis events in the first 2 years were more likely to convert to NMO; they also had a higher likelihood of significant persistent vision loss.

Cerebellar Symptoms: Cerebellar pathways are frequently involved during the course of MS, but a predominately cerebellar syndrome is uncommon at onset. The manifestations include dysmetria, dysdiadochokinesia, action tremor with terminal accentuation, dysrhythmia, breakdown of complex motor movements, and loss of balance. Patients with long-standing MS may develop a "jiggling" gait and an ataxic dysarthria with imprecise articulation, scanning speech, or varying inflection, giving it an explosive character.

Bladder Symptoms: Urinary urgency, frequency, and urge incontinence (due to detrusor hyper-reflexia or detrusor-sphincter dyssynergia) result from spinal cord lesions and are frequently encountered in MS patients. The combined incidence of bowel and bladder dysfunction in MS is more than 70%. Symptoms of bladder dysfunction may be transient and occur with an

exacerbation but are commonly persistent. Impaired vesicular sensation causes a high capacity bladder and may lead to bladder atonia with thinning and disruption of detrusor muscle. Due to incontinence, there is constant dribbling of urine in this irreversible condition. Interruption of brain stem mictuirition center input sometimes lead to cocontracture of the urinary sphincter and detrusor muscles (detrusor=sphincter dyssnergia). If it is untreated, high pressure may lead to hydronephrosis and chronic failure.

Bowel Symptoms: Constipation is a common problem, occurring in 39% to 53% of MS Patients, especially with limited activity and spinal cord involvement. Fecal incontinence is a socially devastating symptom that is often associated with perineal sensory loss in MS patients.

Sexual Dysfunction: It is a frequent problem in MS. Nearly two thirds of patients report diminished libido. One third of men have some degree of erectile dysfunction, and a similar percentage of women have deficient vaginal lubrication. Besides direct neurological impairment, sensory loss, physical limitations, depression, and fatigue additionally contribute to sexual difficulties in MS patients and sometimes the partner's attitude and psychological factors dealing with self-image, self-esteem, and fear of rejection may also lead to impotence or loss of libido.

Intense vertigo assosciated with nausea and emesis is an occasional manifestation of MS relapse. **Speech and Swallowing: Dysphagia** is often due to impairment of cranial nerves IX, X and XII and generally appears late in the course of some patients.

Cognitive Symptoms: Cognitive symptoms are present in 40% to 70% of MS patients. The pattern of cognitive decline is typified by decrease of episodic memory, processing speed, verbal fluency and difficulty with abstract concepts and complex reasoning. To a lesser extent, executive functioning and visual perception, semantic memory and attention span may also decrease. General intelligence is not typically affected.

Emotional Symptoms: Anxiety and depression are more frequent in MS patients than in the general population. In long-term studies, the incidence of depression in MS patients is close to 75%. Patients sometimes experience uncontrollable weeping or less commonly laughter incongruent with their mood. Interruption of inhibitory corticobulbar fibers is responsible for these symptoms (pseudobulbar effect).

Fatigue: It is a pervasive symptom among MS patients that is not related to disability or depression. Over 75% of MS patients experience fatigue during their disease course. A diurnal pattern is characteristic and follows the normal circadian pattern of body temperature fluctuations, with the worse symptoms occurring in afternoon hours (peak core body temperature) often giving way to improvement in the late evening.

Pain: Intense pain and ipsilateral or crossed sensory symptoms may accompany them. Paroxysmal weakness occurs, but it is uncommon. Paroxysmal sensory symptoms like tingling, prickling, burning, or itching may occur in MS patients. Sharp neuralgic pain is also common. Trigeminal neuralgia may appear in patients with MS. The occurrence of trigeminal neuralgia in a person younger than age 40 is suggestive of MS.

Lhermitte's sign (transient sensory symptoms usually precipitated by neck flexion)- It is an electrical or tingling sensation that travels down the spine or into the extremities. Although quite common in MS, Lhermitte's sign can also occur with a wide variety of other disorders, such as vitamin B12 deficiency, spondylosis.

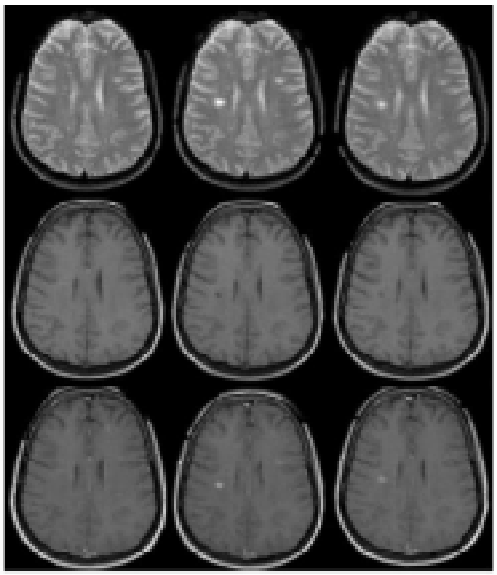
Seizures: It has seen that seizures occur in larger proportion of MS cases compared to normal control subjects. Cortical and juxtacortical lesions may be responsible for the increased incidence of seizures in MS patients. Focal motor seizures, possibly with secondary generalization, are the most frequent. The occurrence of seizures usually follows one of two patterns. Some times focal onset seizures begin early in the course of MS and become a chronic problem that may be difficult to control.(16)

Diagnosis

MS is a disease disseminated in time and disseminated in space (Harrison's)

Diagnosis of MS is largely clinical. The diagnosis of MS is generally done by a neurologist on the basis of history, clinical findings, and supportive clinical tests, including magnetic resonance imaging(MRI), cerebral spinal fluid(CSF), and evoked potentials(EP)

Magnetic resonance imaging (MRI): The brain MRI is the most sensitive test for detecting structural abnormalities due to MS-related disease



Each column contains three identical MRI slices obtained the same day with different image sequences. From top to bottom: T2-weighted, T1-weighted and T1-weighted with gadolinium injection. The three sets of images were made one month apart.

activity. MRI scans show focal brain abnormalities in more than 90% of patients with clinically definite MS. MRI with gadolinium scan distinguish between new or old lesion. Because the imaging abnormalities seen in MS patients can also be seen in other medical conditions, a diagnosis of definite MS cannot be based solely upon the MRI.

Evoked Potentials: In 90% of MS patients shows 90% of abnormal evoked potentials Evoked potentials reflect changes in the electrical activity that occurs within the CNS due to sensory input (a stimulus). Visual evoked Potentials (VEP), Brain Stem auditory evoked potentials(BAEP), and Somatosensory evoked potentials provide evidence of altered nerve conduction. These tests are abnormal in 70-90% of patients with clinically definite MS and often detect abnormalities that are not apparent on neurological examination. Because these tests measure function within the brain or spinal cord, they complement the information about brain structure provided by the MRI.

Lumbar Puncture (Spinal Tap): Cerebrospinal fluid abnormalities are detected in 80-90% of patients with clinically definite MS. These abnormalities include an increase in the number of cells and immunoglobulin proteins suggesting an inflammation or a heightened immune response. This test may be used to establish a diagnosis in patients who have experienced a slowly progressive decline in function without exacerbations (i.e; patients with so-called primary progressive MS) and who have no abnormalities seen on the brain MRI scan. In such instances, a diagnosis of definite MS cannot be made without an abnormality in the spinal fluid. The spinal fluid analysis may also be useful in excluding an infection that may be difficult to distinguish from MS.(17)

Prognosis: The prognosis for visual recovery after each episode of optic neuritis is good, and most patients regain normal visual acuity. Profound visual loss, recurrent optic neuritis, and age older than 35 are associated with a higher risk for poor recovery.

Factors that are associated with an increased risk of developing MS as a disseminated illness are the presence of venous sheathing, recurrent optic neuritis, family history of MS, white race, previous vague or non-specific neurological symptoms, and the presence of oligoclonal bands (OCBs), elevated IgG index, or IgG synthesis rate in CSF. The severity of acute transverse myelitis is inversely related to the risk of acquiring further symptomatic demyelinating lesions. Complete transverse myelitits with profound loss of motor, sensory, and sphincter function imparts a relatively low risk of 3 to 14 for the later diagnosis of MS.

Partial transverse myelitis with preservation of significant motor function at peak is associated with a much higher incidence of MS.

Prognosis depends on the subtype of the disease; the individual's sex, age, and initial symptoms; and the degree of disability the person experiences. Female sex, relapsing-remitting subtype, optic neuritis or sensory symptoms at onset, few attacks in the initial years and especially early age at onset, are associated with a better course

The life expectancy of people with MS is 5 to 10 years less than that of unaffected people. Almost 40% of patients reach the seventh decade of life. Nevertheless, two-thirds of the deaths in people with MS are directly related to the consequences of the disease. There is higher prevalence of suicide in MS than in the healthy population, while infections and complications, depressions are especially hazardous for the more disabled ones.

Patients with sensory or visual symptoms as a dominant neurological sign have a more positive prognosis.(18)

Although most patients lose the ability to walk prior to death, 90% are still capable of independent walking at 10 years from onset, and 75% at 15 years.(19)

Medical Management

Disease -Modifying Agents

There are three disease- modifying agents, which are recognised by the U.S Food and Drug Administration (FDA) for relapsing-remitting MS. It has seen that these drugs are effective on Primary and secondary progressive disease.

Drugs are:-

- Interferon beta1a (Avonex)
- Interferon beta1b (Betaseron)
- Glatriramer acetate (Copaxone)

These drugs have shown approximately one third reductions in relapse rate.

Management for Relapses

Natural improvement occurs in 4 to 12 weeks and degree of improvement varies from person to person

For Acute Relapse- High dose intravenous corticosteroids

Eg- Methylprednisolone (Depomedrol)

Given over 3to5days, gradually followed by tapering does of Oral corticosteroids (eg. Prednisone)

Sometimes Oral corticosteroids may be used to treat a mild or moderate relapse, and intravenous methylprednisolone used to treat Optic Neuritis and retrobulbar neuritis. There are some side effects of having long term corticosteroids, such as Osteoporosis, Hypertension, Cataracts, Muscle wasting etc.

Management for Symptoms

1. **Spasticity-** Most commonly used medications are
   Baclofen (Lioresol), Tinzanidine (Zanaflex)
   Other drugs are-
   Sodium dantrolene (Dantrium) - induce weakness
   Diazepam (Valium)- Spasms (in Night)
   Cyclobenzaprine (Flexoril)- Back spasms
   Carbamepazine (Tegretol)- Tonic Spasms
   There are possibilities of some side effects with overdose of these drugs including sedation, weakness and fatigue but reduction in spasticity.
   Surgical Procedures- Phenol blocks and tendon release for severe spasticity
2. **Fatigue -**
   First line treatment for fatigue- *Amantadine Hydrochloride (effective in 40% cases of MS)*
   Sometimes CNS stimulants like-*Pemoline (cylert) and methylphenidate (Ritaline)* are given but there are some side effects (Anorexia, Irritability, and Insomnia)
3. **Bladder and Bowel Symptoms-**
   Bladder symptoms-
   - Neurogenic bladder:- Urinary tract infection, should be taken care off.
   - Smooth Muscle relaxants or nerve blockers - Propantheline (Pro-Banthine) and Oxybutynin (Ditropan) reduce urinary frequency and urgency.
   - Bethanechol (Urecholine) or Phenoxybenzamine - For urinary retention
   - Crede techniques and intermittent catheterization to empty the bladder.
   - If problem cannot be solved with medication or intermittent catheterization then continuous catheterisation (Foley Catheterisation)
   - Sometimes antibiotics given to treat the bladder infections.
   - Dietary recommendations include drinking 8 glasses of fluid per day

   Bowel Constipation/ Incontinence
   - For Stool Softeners- Docusate (Colace)
   - Laxatives - Milk of magnesia
   - Suppositories- Glycerine or Bisacodyl (Dulcolax)
   - Adequate intake of fluids and fiber in diet.

4. Pain- It is not common in MS

But many people with MS suffer from-

- Paroxysmal Pain

  Medications - Carbamazepine (Tegretol), Amitriptyline (Elavil), Phenytoin (Dilantin), Diazepam (Valium)

- Dysesthesias - Low doses of Amitriptyline (Elavil), Imipramine (Tofranil) or Desipramine (Norpramin)
- Pain and Numbness - Corticosteroids

5. Tremors

- Medications - Hydroxyzine (Atarax, Vistaril), clonazepam (klonopin), propranol (Inderal), buspirone (Buspar), ondansetron (Zofran), and primodone (Myosoline) reduce the tremors
- Dizziness and Vertigo- medications like - Meclizine

6. Sexual Dysfunction

- Erectile difficulties, orgasmic dysfunction and decreased libido

  Medications - sildenafil (Viagra), alprostadil (muse)

  Injections - prostaglandin for erectile dysfunctions

  Sometimes, sexual aids and prosthetic devices also advised.

7. Cognitive Impairments and Emotional Problems (Depression)

For Cognitive Impairments- Retraining and teaching of compensatory techniques

Depression - In studies it has been seen that in MS Depression is 7.5 times higher than general populations.(20)

Rehabilitation Management

Examination :

1. Detailed history including current chief complaints and functional status, family history, medical and surgical history.
2. Strength
3. Tone
4. Range of Motion
5. Balance (Static and Dynamic Sitting and Standing Balance)
6. Coordination
7. Ambulation
8. Bed Mobility
9. Transfers
10. Fatigue
11. Cardiovascular and Respiratory status
12. Visual Impairments
13. Bowel / Bladder / Sexual impairment
14. Speech
15. Swallowing
16. Sensory Status (Deep and Superficial)
17. Cognition
18. Activities of daily living (BADL, IADL)
19. Vocational and Avocational Status
20. Psychosocial Status

Specific Measures for MS

Scales and Assessment tools:

1. Expanded Disability Status Scale (EDSS)- Measures clinical impairments
2. Functional Independence Measures(FIM) and Barthel Index- For functional Status
3. Multiple Sclerosis Quality of Life Inventory (MSQLI)
4. Modified Fatigue impact scales (MFIS)
5. MS functional Composite (MSFC)- measures leg function and ambulation, arm and Hand function and cognition. It can be used for periodic baseline function.(21)
6. Assessment of motor and process skills- A standered assessment for ADLs

Speech affection in Multiple Sclerosis:

Multiple sclerosis affects various parts of the central nervous system. Along with ambulatory problems, individuals with multiple sclerosis experience a lot of problem in speaking and swallowing if the affected areas are the speech and swallowing areas. This affects the normal tone and strength of the muscles of jaw, tongue, lips, soft palate and the pharyngeal constrictors.

The commonest speech problem seen in individuals with multiple sclerosis are slurring of speech due to imprecise consonants and scanning. Scanning problems lead to problems

in rhythm and suprasegmentals of speech. Slurring of speech leads to affected clarity of speech. Swallowing problems in multiple sclerosis are not uncommon. The problems can range from weak oral musculature to disordered swallowing due to reduced pharyngeal peristalsis.

Management is explained in detail in chapter speech rehabilitation.

Physiotherapy Management in Multiple Sclerosis

Physical therapist assesses an MS patient with emphasis on measures of voluntary movement, sensory appreciation, ROM, strength, balance, fatiguability, mobility, gait and functional status.

Framework for Physical therapy management in MS:

According to the National MS Society's Medical Advisory Board, rehabilitation referral should be initiated whenever there is an abrupt or gradual worsening of the function or an increase in impairment that has significant impact on the individual's mobility, safety independence and / or quality of life.

Specific Measures for MS

Scales and Assessment tools: Items in these are included to provide information about the disease process and outcomes and ideally document clinically meaningful change over time.

Expanded Disability Status Scale (EDSS) and Functional Independence Measures(FIM).

Goals of PT in MS :

1. To Improve muscle performance in terms of strength, power and endurance:

   Prescription is based on four interrelated elements (the FITT Equation)

   a) Frequency of exercise: Daily exercises sessions should be scheduled, preferably in the morning, when body core temperature tends to be lowest and before fatigue sets in.

   b) Intensity of exercise :Submaximal Exercise intensities (50 to 70 % of MVC-Maximal Contraction)

   c) Time or Duration of exercise : Exercising to the point of fatigue is contraindicated so frequent rest intervals are advised as time to fatigue varies greatly among individuals with MS. So respect patients desire to rest and allow him to rejuvenate himself between sessions.

   d) Type of Exercise: Circuit training in which improved work capacity is developed through the use of various different stations that alternate work between upper and lower extremities, distributes the load among muscles and may prove best for reducing the likelihood of fatigue.

Symptomwise Management :

1. **Spasticity :** Apart from oral medications,

   a) Topical cold or hydrotherapy can temporarily reduce spasticity by decreasing tendon reflex excitability and clonus and by slowing conduction of impulses in nerves and muscles.

   b) Intermittent static stretching held for minimum of 30 to 60 seconds be applied ideally for 5 to 10 repetitions.

2. **Coordination and Balance training :** Frenkel's Exercises including upper and lower extremity coordination exercises. Also exercises on balance board are recommended in patients who are able to stand, but have poor standing balance and coordination.

3. **Tightness:** Flexibility exercises and ROM exercises, to ensure adequate joint ROM. Mainly stretching advised for hip flexors, adductors,hamstrings and heel cords in lower limbs.

   In upper limbs pectoralis major / minor and lattismus dorsi as these are likely to develop shortness due to slumped posture.

4. **Pain:** Use of TENS (Transcutanious Electrical Stimulation), multidisciplinary stress pain clinic, stress management are adviced.

5. **Urinary incontinence:** Apart from medications, patients are also taught Kegel's exercises to strengthen pelvic girdle muscles, so as to assist in improving stress incontinence due to poor muscular control.

Occupational Therapy Management:

Role of Occupational Therapy

Primary objectives and aims

The primary objective of occupational therapy is to enable individuals to participate in self-care, work and leisure activities that they want or need to perform, thereby optimising personal fulfilment, well-being and quality of life. The occupational therapist evaluates whether people with MS are limited in the life domains that are important to them Figure and determine strategies for overcoming these difficulties. Possible strategies include restoration, compensation, adaptation and prevention.

Interventions

Evaluation is the first step in the occupational therapy programme. The therapist assesses the over all performance areas like ADL, IADL, work leisure, social participations, participation skills like Motor, process and communication skills.

A home and work environment assessment is generally completed to evaluate potential need for modifications and a customised occupational therapy rehabilitation program. As Multiple sclerosis has an unpredictable cause, periodic re-evaluation may be necessary. Patients endurance / fatigue, sleep, patterns, visual abilities, perceptual processing and cognitive emotional and balance should also be reassessed periodically.

Multiple sclerosis is a progressive condition, hence OT focuses on the need of the patients to adapt to their deteriorating / disabling condition. It is essential to educate the patients and their families about the condition and to obtain their support for making long term realistic plans and for consideration, modification or adaption as and when required

Programme improves the individual's ability to perform daily activities within his or her own unique situation, thus, often requiring a combination of different techniques for making long term realistic plans.(22)

In patients with multiple sclerosis, occupational therapy can be used to develop strength, stamina and coordination. Occupational therapist emphasizes on active and purposeful occupations, which have an advantage of engaging the clients attention and interests as well as help in psychological upliftment.

Occupational therapist may initiate coordination training with neuromuscular education and progress to repetitions activities requiring desired coordinated movement patterns. Placing blocks, marbles, cones, paper cups or pegs, are enabling activities that demands repetition pattern of non-resistive movements.

Activities should be structured to enable the client to use precise movement pattern and to work at speeds consistent with the maintenance of precision. Therapist should avoid constantrepetition of an incoordinated pattern as this reinforces incoordination. Practice that increase incoordination such as fear, poor balance, faulty posture, pain, too much resistance, too much fatigue, strong emotions, prolong inactivity and excessively prolong activity should be avoided or dealt with prior training.

Purposeful activities such as leather lacing, mosaic tile work, needle craft and house hold task such as wiping, sweeping and dusting also provide repetitions in movement along with psychological benefits such as alleviating feelings of guilt and reinforcing feelings of self worth.

Fatigue is one of the most common symptoms in MS(reported by at least 75% of MS patient) which impaired quality of life among MS patients, independent of depression or disability. Fatigue also imposes significant socioeconomic consequences, including loss of work hours and in some instances, loss of employment.

Role of occupational therapist is to teach energy conservation techniques, so individuals can participate in self-care, work and leisure activities.(23)

Occupational therapy management to improve Coordination, strength and endurance

Activities that can be given to improve balance and co-ordination, strength and endurance

- Reach outs in all fours position like pegs reach outs to improve gross motor and eye hand coordination.
- Reach outs in kneeling position in different directions to improve balance and overhead peg transfers to improve bilateral hand coordination
- Ball throwing in kneeling and standing position to improve balance

- Wand exercises to improve gross motor coordination
- Beads stringing and picking small beads by cloth pins improve fine motor coordination

Management for Spasticity -

Active and passive ROM exercises

- Weight bearing exercises
- Hydrotherapy
- Serial casting in inhibitive postures has been seen to be effective in tone reduction.(25)
- Stretching exercises

The aims of stretching in spasticity are to improve muscle extensibility, reduce muscle stiffness, and improve function. Clinically, a range of stretching techniques are used including static, dynamic, Proprioceptive Neuromuscular Facilitation (PNF), as well as prolonged and ballistic stretching.(26)

When sitting for long periods in a wheelchair, there is a greater risk of tone increase and muscle shortening (for example of hip adductors and hip flexors). Rehabilitation should then include instruction on sitting correctly in a tone inhibiting posture (for example, with knees separated). Tone-inhibiting postures enable long-term stretching of the spastic muscles and are effective in preventing muscle shortening.

Use a tilting table to stretch hip flexors and calf muscles using body weight. For the arms, one can consider individually adapted splints, for example to maintain an open hand position, which is important for hand hygiene.(27)

Management for Anxiety and Depression- Deep breathing and relaxation techniques.

Regular relaxation can lead to decreased tension in muscles, lower blood pressure and slower heart rate. Relaxation can help with fatigue as it promotes good sleep patterns; increases benefit from rest periods during the day and can be used to manage stressful situations.

Deep breathing is the basis for many relaxation techniques. However, focusing on how you breathe and creating a slow, deep and even pattern helps to feel calmer and more relaxed and can create a distraction from the causes of stress.

Managing Heat during exercise in Multiple Sclerosis Rehabilitation.

Some patents with MS are affected by heat, but some are particularly sensitive. Hot weather, an over-heated room and exercise can all make MS symptoms worse. This is a temporary effect but when the body cools down again, symptoms return to the level they were before. If patients are sensitive to heat, keeping cool during or shortly before exercise may help to do exercise for longer, or more strenuously, without bringing on heat-related symptoms. Patient can take ice drinks, cooling garments, or give regular breaks to prevent overheating. Research showing benefits for these cooling techniques is not conclusive, and they may not help everyone, but they are unlikely to be harmful. Sometimes lowering the body's temperature, with cold baths or cooling garments, cold packs reduce muscle stiffness temporarily. In contrast, some people with MS find that cool temperatures make their spasms or stiffness worse. For these people, exercising in a warm swimming pool may help with stretching and relaxing muscles. Precaution is needed while taking hot and cold therapies. When applying cold directly to the skin, or when using cooling garments or cold water to cool the body, care should be taken not to damage the skin. MS can cause changes in the way the patient experience temperature, distorting the feeling that would normally tell them when something is too hot or too cold.

Strategies to manage fatigue:

- Frequent, short rests can be beneficial.
- Balance between periods of rest and activity. Regular rest periods should be built into the daily schedule. Rest should be taken before fatigue sets in. The balance can be difficult to find -- too much exercise can induce fatigue, but decreased physical activity can also lead to tiredness and lack of energy.
- Organize the tasks Priorities: most important and valued activities should be done first. A valued leisure activity can boost motivation and well-being, better equipping the person to take on the next task.
- Because of fatigue, full time work may not be possible hence part time work should be considered.
- Ergonomics are important. Good, supportive seating posture should be encouraged.

Ball throwing in sitting to improve balance

Ball throwing in standing to improve balance and coordination

Weighted resistive therapeutic exercises to decrease tremors in upper limbs

Rearrange the environment to avoid unnecessary lifting, carrying or stooping. Use proper lifting techniques -- bend with the knees instead of the back. Use a trolley, if necessary, to avoid unnecessary carrying.

- Simplify tasks where possible. Hard tasks can be done with help of another person, to reduce fatigue.
- Smoking can increase fatigue as can infection.
- A healthy, balanced diet can help -- involve a dietician. Unnecessarily hefty meals can invoke fatigue as can an inadequate diet.
- Extremes of temperature can exacerbate fatigue -- for example. Hot bath
- Stress can contribute to fatigue, so explore methods of minimizing stress.
- Always set realistic goals, Unrealistic goals can cause stress. Relaxation techniques may be helpful.

Occupational therapy in the home: - Meeting the individual and the family in their environment can provide the occupational therapist with valuable information that may not be readily available when assessing the person within a healthcare setting. The assessment includes an evaluation of the individual's current functional status in relation to the performance of activities of daily living.

An assessment of the home situation includes the evaluation of the need of any modifications For example:- Kitchen modification or need for assistive equipment and training.

Finally, the occupational therapists visit to the home can serve to determine whether additional paid assistance, e.g. Part time paid caretaker may be useful.

Kitchen Modifications - Use pull-out shelves; install these just below waist height or at horizontal reach. Use lightweight dishes and pans, and for larger and heavier items use a trolly.

Use of assistive devices- Assistive devices can help the patient to be independent. Sometimes individuals have poor balance and difficulty in overhead functions, so tools like long-handled shoe horns, reachers, and dressing sticks can be helpful

- Use of dressing and grooming aids (demonstrating button hooks, suggesting replacing zippers and small buttons with Velcro, making closets easier to use)
- Use eating and drinking devices (helping clients use assistive devices, such as double-handled cups, lightweight or weighted cups)
- Helping patients learn how to use assistive or mobility devices (wheelchairs, canes, scooters, walkers) efficiently in their home (transferring to / from the devices and beds, toilets, chairs; maneuvering them in various spaces; installing ramps to make using devices easier)
- Computers open a world of recreation and job-related activities, but MS symptoms may require special adaptations. For example, people with vision problems can use computers with enlarged keyboards, alternative mouse such as a Tracker ball can be helpful or text-to-speech programs that read aloud the material onscreen. Location and setup of computer should be easily assessable.

Electronic remote controls can be used for virtually all the appliances in an environment-lights, radios, telephones, televisions, air conditioners, An OT can help the patient to determine which devices are most appropriate. Patients with MS with memory problems may find a day times very useful to schedule their daily activities. It will help them to remember things to do and also conserve their energy.

Patient should try to do the hardest or necessary tasks for times when they have the most energy and save the easier tasks for less energetic times.

Use mobility aids whenever required. Occupational therapists can help the patient to find the right ones. Canes, walkers, wheelchairs and scooters may be warranted.

Occupational Therapist can help the patient learn to use all these equipments efficiently in their home (transferring to / from the devices and beds, toilets, chairs; maneuvering them in various spaces; installing ramps to make using devices easier). (28,29,30)

Vocational Rehabilitation:

People with MS constitute a valuable labour resource for the societies in which they live. They are often well-educated and skilled workers with extensive employment histories, which is not surprising given that MS typically manifests itself in early-to-middle adulthood (after people have begun and, in many cases, established their careers). Too often, however, these highly trained, productive, and experienced workers leave the work force shortly after MS diagnosis, usually of their own choosing and usually before the illness has rendered them unable to work.

Experts do not know exactly why so many people with MS leave the work force prematurely, but some of the most commonly reported barriers to continued employment among people with MS include

- The unavailability of transportation to and from work,
- Difficulties in obtaining on-the-job accommodations
- Lack of awareness of assistive technology
- Limited awareness of employment issues on the part of treating physicians, disincentives to work in government assistance programmes
- The unpredictable and sometimes progressive nature of MS

Studies of people with MS show that men, people with

- Higher levels of education
- People who do not experience cognitive impairments
- Workers whose jobs require little physical exertion or exposure to heat
- Management or professional-level workers,
- Employers who have formal disability related personnel policies are the most likely to retain employment over time.

Vocational rehabilitation provides a mechanism for people with MS to make adjustments in their careers and to continue working as long as they wish to. The purpose of vocational rehabilitation is to provide services, supports, and training that enable people with disabilities to obtain, maintain, and advance in jobs that are compatible with their

interests, abilities, and experience. Early intervention is emphasised. Occupational therapist can assess and direct patients to get job placement. Career counselling services offered to people with MS who have left the work force but wish to return to it. Occupational therapist conduct vocational evaluation of vocational interests, limitations, strengths, work behaviours, transportation, identification and implementation of workplace accommodations, training in the use of assistive technology, self advocacy training, and consultation with employers on a wide range of disability-related matters.

For people with MS, the workplace accommodations that have proven effective in helping them stay in the work force include

Schedule modifications (the most common form of workplace accommodation implemented on behalf of people with MS),

Memory aids to combat cognitive impairments,

Motorised scooters to combat fatigue and mobility problems, climate control in the work station,

- Low vision aids (e.g. magnification machines, voice output software),
- Accessible parking, building renovations to allow for wheelchair access,
- Cooling vests,
- Ergonomic keyboards
- Voice-activated computer programs
- Telecommuting or home-based employment.

Occupational therapy focuses on learning strategies for managing daily life, based on the person's physical, social and psychological needs.

PSYCHOLOGICAL ASPECT IN MULTIPLE SCLEROSIS

Multiple sclerosis is an extremely unpredictable and complex disease as no two patients suffering from the disease have the same psychological effect. Numbness, fatigue, pain, visual disturbance, parenthesis, muscle weakness, muscle spasm - these troubling symptoms are invisible to other people as compared to a cane or a wheelchair. This makes multiple sclerosis like an iceberg with a huge hidden part that often causes intense mental anguish.

The significant stress of being diagnosed with chronic illness can trigger off psychological issues such as depression and anxiety. When behavioural changes are addressed, the changes are described simply as emotional reaction to the life situation or poor adjustment to chronic illness. Patients with multiple sclerosis don't go through a definite orderly set of stages culminating in adjustment. Rather, adjustment is an ongoing, life process that flows through the unpredictable changes brought about by the disease.

Psychological Process and Problems:

Emotional Issues:

Major Depressive Disorder: Depression is the most common psychological symptoms in multiple sclerosis. The incidence rate in multiple sclerosis is three times that of the general population. The 12-month prevalence of major depressive disorder among persons with multiple sclerosis is 15.7%, nearly double the prevalence of major depressive disorder in persons without multiple sclerosis (7.4%). Reports of the lifetime risk for major depressive disorder in multiple sclerosis populations have ranged from 27-54%. It could be very difficult to diagnose multiple sclerosis as there are many symptoms that are seen in both like fatigue, weight loss, etc. Also, suicidal ideations are commonly seen in patients suffering from multiple sclerosis, occurring7.5 times higher than in general in the population. The point prevalence of anxiety in multiple sclerosis ranges from 19% to 34%. In order to slower the progression of multiple clerosis patients are put on various medications such as Baclofen, Dantrolene, Betaseron and Avonex etc. these could cause changes in mood in either direction. Also, as some of them are on steroids, it could also lead to periods of hyperactivity and euphoria, which could be followed by a low feeling period. Also certain steroids can cause or worsen depression. Causes of depression in multiple sclerosis patients is due to psychosocial challenges, maladaptive coping, brain lesions resulting from multiple sclerosis disease in process and immune deregulation associated with multiple sclerosis exacerbation.

Studies suggest that 13% patients with multiple sclerosis also have bipolar disorder. Bipolar may be an initial symptom of multiple sclerosis, preceding other neurological symptoms. While manic episodes in multiple sclerosis may be precipitated by steroid therapy, there is increasing evidence that affective disturbance may be due to

organic changes in the brain. Both euphoria and pathologic weeping or laughing in multiple sclerosis patients may be due to lack of emotional control in multiple sclerosis.

Treating depression in MS is often rewarding and drugs in the Specific Serotonin Reuptake Inhibitor (SSRI) group are probably safest. Common side effects, usually transient, include nausea, sexual dysfunction and gastrointestinal disturbance. Withdrawal from these drugs should be gradual, especially with paroxetine (Seroxat).

Euphoria: This is a mental condition where there is a feeling of happiness, confidence, or well-being sometimes exaggerated in pathological states as mania. It can often lead to having unrealistic expectations and thought process. It is seen that patients in this state lack insight. Cognitive problems like inattention, disturbed thinking, etc usually trigger euphoria.

Emotional Liability: Emotional liability means that the patient may easily burst into tears, both at happy and sad events or might become suddenly very angry or aggressive which may be inappropriate or exaggerated for the situation. This is different from pathological crying or laughter. In patients with multiple sclerosis, the symptom of emotional liability may vary from mild to severe.

Lack of Insight: Patients with multiple sclerosis may lack insight about their condition and this could cause a lot of problems. Lack of insight may lead to problems in day-care or rehabilitation. For example, when a patient suffering from multiple sclerosis may try to regain walking, when everybody around would be aware that he or she may not be able to walk again. This is when the caregivers and the therapists may face a problem as they would be in a soup whether to encourage or discourage the patient.

There could be a number of emotional aspects that a patient may go through like anger, isolation, loss of control and dependence, self doubt and dislike which is discussed in the chapter of psychological rehabilitation.

Cognitive Deficits:

As multiple sclerosis is a disease that causes damage to nerve fibers in the brain and spinal cord because of which demyelination occurs in the brains of most patients with multiple sclerosis. A neuropsychological point of view suggests multiple sclerosis often affects higher order cognitive functioning; typically causing deficits in speed of information processing, attention, executive functions, verbal fluency and memory were clearly associated with less brain volume, and with higher lesion loads, in particular at frontal and occipital areas. There has been research that between 45% to 65% of all people, with multiple sclerosis experience problems with memory, attention, problem -solving and other cognitive functions, which may vary from one patient to another.

Problems in neurocognitive functioning in multiple sclerosis are mainly modulated by speed and stability of speed processing; in particular when attention-demanding controlled information processing is required. Cognitive difficulties can be particularly stressful for people because they are changes that nobody sees. Understanding the type and the source of cognitive problems is the first step for patients and their family members to begin coping with these symptoms effectively. The cognitive issues may vary from patient to patient. The range of cognitive dysfunctions which can be associated with multiple sclerosis:

Cognitive Fatigue: Research shows that patients with multiple sclerosis even if they have no other cognitive problems, they can get fatigued from doing tasks that are mentally challenging work, even if they aren't exerting themselves physically. Patients with multiple sclerosis once they are cognitively fatigued they are more likely to work slowly and make errors than people who do not suffer from multiple sclerosis.

Attention and Concentration: Problem arises when patients suffering from multiple sclerosis are asked to focus on more than one thing. Often patients as their disease progresses they feel that they can work on one thing at a time, in a quiet and distraction free place whereas, before they could multi-task quiet easily.

Processing Speed: Cognitive slowing in individuals with multiple sclerosis (MS) has been documented by numerous studies employing explicitly timed measures in which speed of responding is an obvious focus of task performance. Often the patients experience an overall slowing the way they process information as compared to before.

Memory: Memory is a complex neuropsychological function. There are two separate systems for memory:

1. **Procedural memory** is the memory for how to do things. Procedural memory guides the

processes we perform and most frequently resides below the level of conscious awareness. When needed, procedural memories are automatically retrieved and utilized for the execution of the integrated procedures involved in both cognitive and motor skills. In multiple sclerosis, this almost always remains intact and unaffected, so although you may forget to do something, you will not forget how to do it.

2. **Semantic memory** needs several steps to occur before a memory can be stored or recalled, and interruption at any stage, such as may be caused by the inability to concentrate, can interfere with the ability to store or retrieve information. This memory can be affected in multiple sclerosis. It's often seen that patients with multiple sclerosis have trouble recalling recent everyday events since the diagnosis. Some patients could also experience difficulty recalling things that they see but have no trouble recalling things that they have heard, while others can remember things that they see with clarity but are unable to remember what they have heard. In very advanced cases, memory problems could be as severe as to completely disrupting the learning and recall process.

Language: In patients with multiple sclerosis they could be issues related to language fluency. Especially, there could be difficulty in finding words i.e. accidentally saying the wrong word and problems with naming i.e. problems with recalling uncommon words or the names of acquaintances. Patients with multiple sclerosis can also have problems with dysphagia or dysarthria, which refer to problems coordinating the movements of the lips, mouth, tongue, and throat. They also suffer from slurred speech or physical difficulty saying certain words or syllables, without any cognitive problems at all.

Problem Solving: At times, people with multiple sclerosis have difficulty solving problems in new situations or they may have difficulty making decisions because it is hard to identify which of a number of choices is the best.

Visual Spatial Skills: Patients may find themselves getting lost more frequently, losing items, or having trouble understanding visual-spatial information like maps, diagrams, and charts.

Evaluating the Cognitive Changes: The first step to dealing with cognitive changes is to recognize that they have been changes. Usually patients with multiple sclerosis do not take these changes quiet seriously. Adequate assessments of cognitive deficits need to be carried out by a psychologist or a neuropsychologist with the help of standardized neuropsychological tests. It is also important to evaluate depression, stress, anxiety which may be a contributing factor to cognitive problems with the help of scales like Beck's Depression Inventory, Hamilton Anxiety Rating Scale, etc. An overall assessment would help the psychologist formulate a good rehabilitation plan like vocational rehabilitation, improved family life, cognitive rehabilitation, etc.

Treatment for Cognitive Deficits:

There is as such no "pill" which could improve the cognitive functions in multiple sclerosis. However, there is a class of drugs called potassium channel blockers (4-aminopyridine (4-AP and 3, 4 diaminopyridine) have been studied for several years. The potassium channel blockers increase the speed of nerve conduction in demyelinated nerve fibers, suggesting that they might improve neurologic function and cognition. However, these drugs lead to high risk for seizures as they speed up the nerve conduction. A recent study of 69 patients suffering from multiple sclerosis with memory deficits showed that Aricept had modest benefits for verbal memory i.e. the ability to remember a list of words. Five drugs are now approved by FDA of which Avonex has demonstrated to be the most positive benefit on cognition. However, none of the medications have shown dramatic benefits, on slowing the progression of cognitive dysfunctions or reversing the effects.

Cognitive Rehabilitation: Cognitive rehabilitation is a systematic program designed to improve cognitive functions. There are a few stratergies mentioned which are used during a cognitive rehabilitation session:

- If the patients complain of weak memory then substitute it for an organizer for example maintain a dairy where one needs to write down everything down, or maintain planners and set up sections for appointments, addresses, phone numbers, driving directions, etc.
- Maintain particular place for everything as this would help to avoid misplacing things.

- Try to focus more attention on things that are important and keep repeating it to yourself so that it registers in the long term memory. Meditate regularly to improve attention and concentration.
- Work slowly and the focus should be on accuracy rather than speed as a patient with multiple sclerosis suffers from poor processing.

Sexual Issues:

Sexuality is an integral part for each one of us and it has an impact on our identity, personality, interpersonal relationship and our life span. Multiple sclerosis is a disease which can target any part of the nervous system like if it affects the nerves servicing the reproductive organs, which in turn could lead to changes in sexual functioning. Multiple sclerosis can cause problems for the patients on two aspects: neurologically and psychologically. Neurologically, the sexual excitement and response arises in the central nervous system which is normally affected. While the brain sends messages to the sex organs through spinal cord but due to multiple sclerosis the affected area is the nerve pathways which is caused by demyelination. This can cause impairment in sexual functioning like:

- Loss of libido or sexual interest
- Slower arousal time
- Difficulty masturbating
- Changes in genital sensation
- Decreased vaginal lubrication and muscle tone
- Difficulty in erectile functioning and ejaculation
- Difficulty attaining climax

Psychologically, the patient may go through problems like bowel and bladder incontinence, weakness, fatigue, decreased sexual desire and loss of interest. Also, as physical contact and sexual positioning may become uncomfortable due to sensory changes this could lead to further impact psychologically like:

- Depression, stress or anxiety
- Performance anxiety
- Altered body image or sexual self-image
- Fear and worries about the future of the relationship
- Low self esteem
- Fertility and pregnancy issues
- Genetic factors

How do couples regain their sex life?

1. **Communication:** The first step is to accept that there is a problem and that to verbalize it with your partner or the person you are able to confide in. By being open about the situation would reduce the level of embarrassment and doubt about the situation.
2. **Consulting a Doctor:** Discuss the side effects of medication that one takes for multiple sclerosis and medications taken for muscle relaxants and pain as this could lead to being intimate more readily.
3. **Alternatives:** Doctors could suggest various alternatives.
4. **Sexual Counseling** is necessary as it helps the couple to come in terms with the challenges of multiple sclerosis and also come up with different alternatives. Sexual Counseling is further discussed in the psychological rehabilitation chapter.

Family Issues:

Families have their own style of functioning, coping and communicating. If anyone the family suffers from multiple sclerosis eventually the whole family would be involved physically, psychologically and financially. It can lead to a lot of stress and challenge to the coping mechanisms of everyone in the family.

A person who was once there for the family as financial, social and psychological support to the family members, after diagnosis may have to reduce working hours which may lead to reduces family income. Also, the person who may have been very active socially may now avoid social functions and interaction and may also need emotional support as a result of his deteriorating condition. Family members may be expected to take on additional responsibilities of the family member who is suffering from multiple sclerosis. This leads to change in functioning and family role, also people might feel that this change would be temporary which is not true, it would be a lifelong change and responsibility.

Often the family members may stop spending time on leisure activities that they would enjoy doing previously. Family members should spend some

time on their hobbies or what they enjoy doing, otherwise this would lead to caregivers burnout.

Multiple sclerosis can be very challenging to a married life as when the patient is diagnosed as multiple sclerosis the couple is usually planning and working towards future goals. The couple needs to have a strong bond which would help them to come out with effective coping mechanisms. As the course, of the disease could be very unpredictable with high chances of relapse. The best possible way to deal with the situation is to forget the old ways of doing things and adapting to the progression of the disease and finding out alternatives to old ways of doing things. Work out a specific plan for determining how practical needs are handled and who will be responsible for what, when. Patience, compromise and enlisting help from other family members could prove to ease out the situation.

Hiding the diagnosis from the children would not be an idle way to deal with the situation as the children would sense something wrong. Also, they would not be able to understand the behaviour of the parent suffering from multiple sclerosis like being inactive and unavailable most of the time. It could lead to misunderstanding that the parent is not interested in the child. It's better to inform and educate the children about the disease, and how the children need to cope with the situation. Children may go through a number of emotions like guilt, stress and anxiety. Let the children ask questions and if you don't know the answer, help them find those answers. Children often take pride in being able to contribute in a small ways, allow them to do so and appreciate their concern. What children need the most? Is time, love and attention, so find alternatives as to how this could be done.

Family Therapy: Family therapy is based on the belief that the family is a unique social system with its own structure and patterns of communication. These patterns are determined by many factors, including the parents' beliefs and values, the personalities of all family members, and the influence of the extended family. Before getting on with family therapy it is very important to evaluate the strength and weakness of how the family functions.

Psychological Tests and Battery: The below listed are discussed in detail in psychological rehabilitation chapter.

Beck's Depression Inventory - To assess depression

Hamilton's Anxiety Rating Scale - To assess anxiety

Wechsler's Memory Scale - To assess cognition

Psychological Intervention for Multiple Sclerosis: The below listed are discussed in detail in psychological rehabilitation chapter.

- **Individualized Psychotherapy:** This allows the patient with multiple sclerosis to deal with emotional difficulties, lack of motivation, negative impact of the disability and inability to handle change in functional skills typical of this condition.
- **Group Therapy:** A new study at The University of Nottingham has revealed that offering emotional support to Multiple Sclerosis sufferers through group therapy sessions could improve their quality of life.
- **Sexual Counseling**
- **Cognitive Rehabilitation**
- **Family Therapy**

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Section 3

Muscle and Nerve Related Disorders

Ch.9 Muscular Dystrophy

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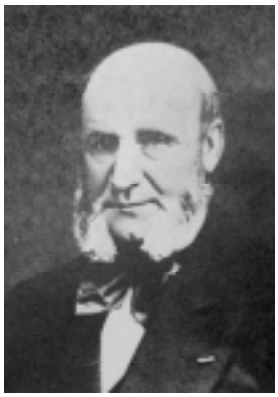
Sir Charles Bell, in 1830 was the first one to note the progressive weakness seen in boys which was studied further by another scientist who reported generalized weakness, muscle damage & replacement of damaged muscle tissue with fat& connective tissue on two brothers and was thought to be tuberculosis. By 1850s, these boys grew progressively weaker, unable to walk & later on died at an early age and made a marked impression on medical field. Later in 19th century French neurologist Guillaume Benjamin Amand Duchenne did pioneering work & gave a comprehensive account of 13 boys with the most common and severe form of the disease(which now carries his name-Duchenne muscular dystrophy) & revealed of the disease having more than one form, and that these diseases are affected in people of either sex and of all ages. Duchenne was the first to do a biopsy to obtain tissue from a living patient for microscopic examination & used his biopsy needle on boys with DMD and concluded to have this disease of muscle origin.



Guillaume Benjamin Amand Duchenne

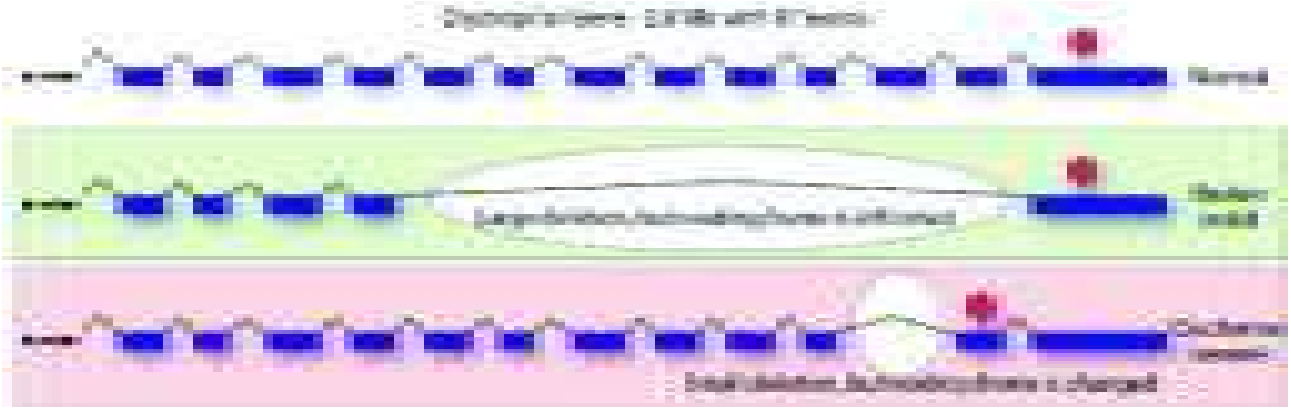
Muscles are composed of many muscle fibers which are separated by each other by connective tissues called endomysium and arranged in bundles called fasciculues where individual fibers are arranged parallel to each other. Each fasisculus has a outer connective tissues membrane called perimysium and muscle has a whole consists of all these fasisculues together with outer layer called epimysium

When an impulse travels from brain down to neuromuscular junction through the peripheral nerves, a chemical named acetylcholine is released which causes series of events resulting in muscle contraction.

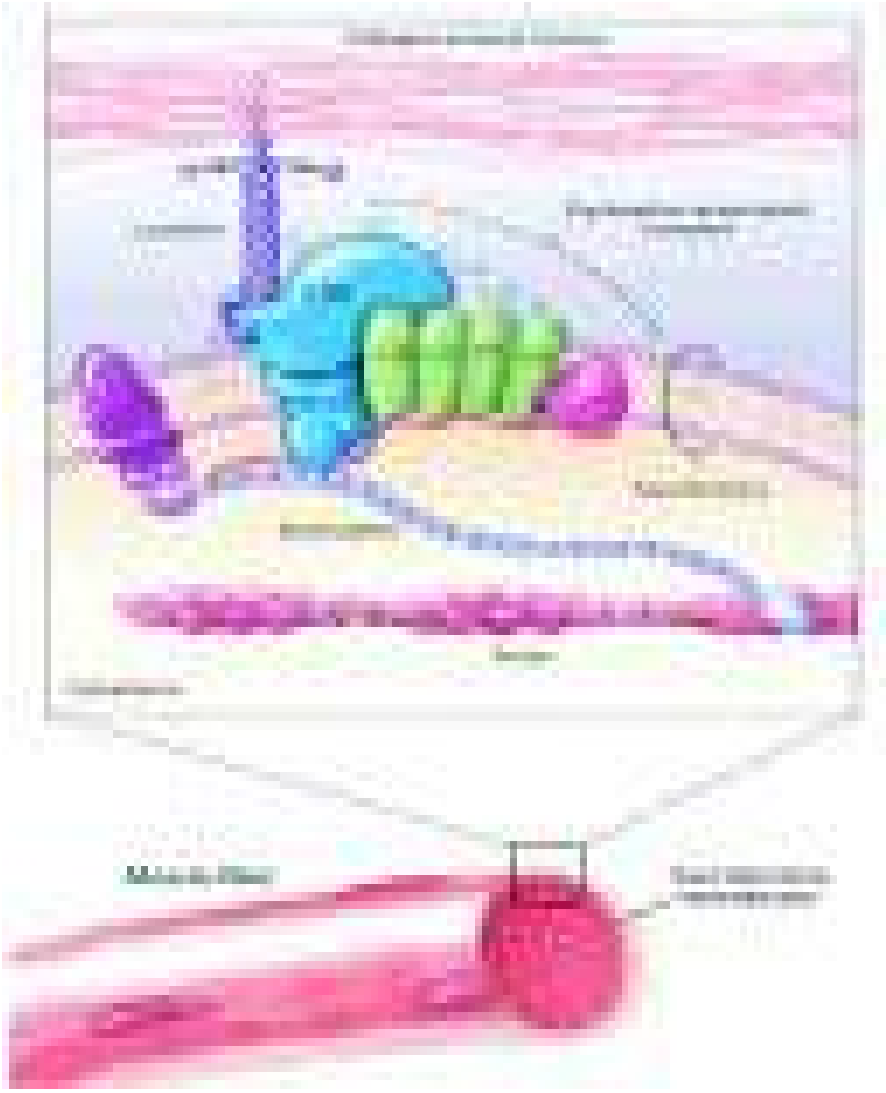
During contraction and relaxation of muscle, muscle membranes are protected by group of protein named dystrophin- glycoprotein complex which are inbuild in membranes which contains protein creatine kinase needed for the chemical reactions which produces energy for muscle contractions. When this protective membrane gets damaged, there is a leakage of protein creatine kinase leading to excess calcium influx in muscle fibers, which causes further harm and eventually death thereby resulting to progressive muscle degeneration.

All the muscular dystrophies are inherited and result from different mutations in the gigantic gene that encodes dystrophin and which is required to maintain muscle integrity. The body cells don't work properly when a protein is altered or produced in insufficient quantity (or sometimes missing completely. Many cases of MD occur spontaneous mutations that are not found in the genes of either parent, and this defect can be passed to next generation. There are 79 exons in DMD gene spanning at least 2,300 kb. Deletions cause deficiencies in 1 or more of these as a result of which mental retardation and cardiomyopathy sometimes accompany DMD, which is seen in clinical presentation depending on what the deletions was removed from the dystrophin gene.

Dystrophin protein is found in muscle fibre membrane, acting like a spring. It joins the membrane actin filiments. The protein is rod shaped around 150nm in length, 3684 amino acids long, 427kDa molecule weight. It is hydrophobic (does not like water). Conformation is alpha-helical, allowing protein to act as a shock absorber, preventing overstress. Dystrophin links actin (cytoskeleton) and dystroglycans of the muscle cell plasma membrane, known as the sarcolemma (extracellular). Dystrophin functions in two ways; mechanical stabilisation and regulated calcium levels. Cycles of degenerating and regenerating occur in presence of dystrophin deficiency. It's a mystery as how these cycles are related to altered strength & stability of cell membrane, increased cell permeability of cell membrane, altered



Dystrophin Gene



Dystrophin Protein

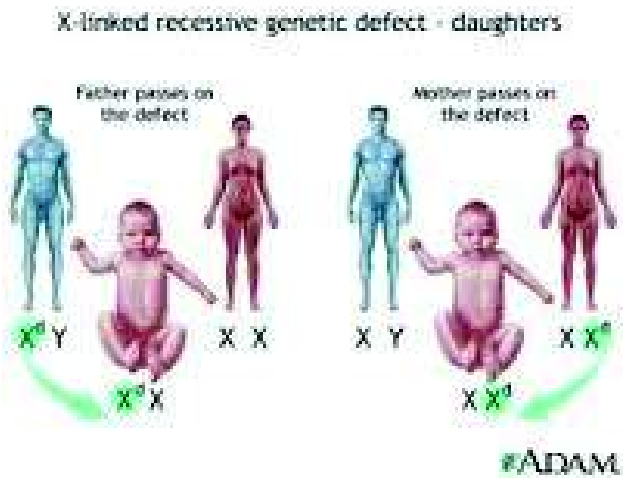
mechanism of Ca+2 regulation, calpain activity, ischaemia, mast cell infiltration, satellite cell function & proliferative potential, basic fibroblast growth factor activity, activity of platelet derived growth factor receptors, endomysical & perimysical fibrosis& finally replacement of muscles by fibrofatty connective tissue.

In Muscular dystrophy, integrity of the muscles fibers are predominately affected leading to muscle degeneration, progressive weakness, fiber death, fiber branching and splitting, phagocytosis and at times leading to chronic or permanent shortening of tendons and muscles which results in overall decreased or absent in muscle strength and tendon reflexes due to replacement of muscle by connective tissue and fat.

(Genes are like blueprints containing coded messages that determine a person's characteristics or traits; arranged along 23 rod-like pairs of chromosomes,*- with one half of each pair being inherited from each parent. Each half of a chromomsome pair is similar to the other, except for one pair, which determines the sex of the individual.)

Muscular dystrophies can be inherited in three ways:

1. **Autosomal dominant** inheritance occurs when a child receives a normal gene from one parent and a defective gene from the other parent. Autosomal means the genetic mutation can occur on any of the 22 non-sex chromosomes in each of the body's cells. Dominant means only one parent needs to pass along the abnormal gene in order to produce the disorder. In families where one parent carries a defective gene, each child has a 50 percent chance of inheriting the gene and therefore the disorder. Males and females are equally at risk and the severity of the disorder can differ from person to person.
2. **Autosomal recessive** inheritance means that both parents must carry and pass on the faulty gene. The parents each have one defective gene but are not affected by the disorder. Children in these families have a 25 percent chance of inheriting both copies of the defective gene and a 50 percent chance of inheriting one gene and therefore becoming a carrier, able to pass along the defect to their children. Children of either sex can be affected by this pattern of inheritance.
3. **X-linked (or sex-linked) recessive** inheritance occurs when a mother carries the affected gene on one of her two X chromosomes and passes it to her son (males always inherit an X chromosome from their mother and a Y chromosome from their father, while daughters inherit an X chromosome from each parent). Sons of carrier mothers have a 50 percent chance of inheriting the disorder. Daughters also have a 50 percent chance of inheriting the defective gene but usually are not affected, since the healthy X chromosome they receive from their father can offset the faulty one received from their mother. Affected fathers cannot pass an X-linked disorder to their sons but their daughters will be carriers of that disorder. Carrier females occasionally can exhibit milder symptoms of MD.

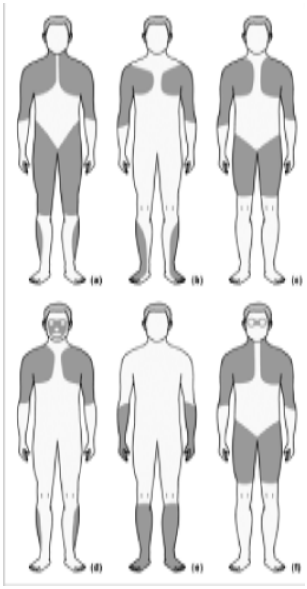


Types of Dystrophies:

There are nine types of the muscular dystrophies which are classified on the basis of the :

1. Extent and distribution of muscle weakness.
2. Age of onset
3. Rate of progression.
4. Severity of symptoms
5. Family history (including any pattern of inheritance).

Some forms of MD appears in infancy or childhood while others appear in middle age or later. Even though incidence rates and severity varies, each of these dystrophies causes progressive skeletal muscle deterioration, where some types affect cardiac muscle also.



Infancy or childhood onset Muscular dystrophy are classified into four types:

1. Duchenne Muscular dystrophy.
2. Becker Muscular dystrophy.
3. Congential Muscle dystrophy.
4. Emery- Dreifuss Muscular dystrophy.

1. **Duchenne Muscular Dystrophy (DMD)** is the most common and severe childhood form of muscular dystrophy, affecting one in 3,500 male births. DMD results from an abnormal gene which is seen on X-chromosome at band Xp21, which encodes for dystrophin, a427-kD cytoskeletal protein in membrane. Since it has X-linked recessive inheritance, it primarily affects boys, whereas girls and women who carry the defective gene may show some symptoms. About one-third of the cases reflect new mutations and the rest have family history where sisters of boys with DMD have a 50 percent chance of carrying the defective gene.

In most of cases, DNA analysis is normal and diagnosis gets confirmed by immunoblotting or immunostaining techniques where complete absence of dystrophin from muscle tissue is seen. Pathological tests show creatine Kinase (CK) elevated more than 100 times in early stages of disease and these levels decrease over a period of time with loss of muscle mass. Elevated CK is evident at birth long before symptoms are evident

DMD usually becomes apparent when a child begins to walk with onset of muscle weakness typically seen in between ages 2 and 5. Progressive weakness and muscle wasting is seen in proximal group of muscles in lowerlimbs (a decrease in muscle strength and size) caused by degenerating muscle fibers before spreading into the upper arms. Other symptoms include loss of some reflexes, a waddling gait, frequent falls and clumsiness in movements especially while running or jumping, difficulty when rising from squatting position, low chair, sitting or lying position or while climbing stairs, early fatigue, postural imbalance, impaired breathing, lung weakness, and cardiomyopathy (heart muscle weakness that interferes with pumping ability).The muscles are wasted and are replaced by an accumulation of fat and connective tissue, causing them to look larger and healthier than they actually are hence called pseudohypertrophy). As the disease progresses, the respiratory muscles (diaphragm) which assist in breathing and coughing weakens resulting into breathing difficulties, respiratory infections, and swallowing problems. Bone thinning and scoliosis are common complications with slight impairment in cognition. Between ages 3 and 6, children may show brief periods of physical improvement followed by progressive muscle degeneration and becomes wheelchair-bound by age 12 and usually die in their late teens or early twenties from progressive weakness of the heart muscle, respiratory complications, or infection.

2. **Becker Muscular Dystrophy** is closely related to DMD with less severity. It is slowly progressive X-linked recessive disorder with partial but insufficient function of the protein dystrophin. The onset of weakness is manifested between the age 10 and 15 years, and patients generally live upto the middle age or later. The rate of progression, symmetrical pattern, muscle atrophy and weakness varies greatly among affected individuals. Most of the patients are able to walk until they are in their mid-thirties or later, while others are unable to walk past their teens where as some doesnt need any wheelchair. As in Duchenne MD, muscle weakness in Becker MD is typically noticed first in proximal muscles like the upper arms and shoulders, thighs and pelvis.

Early symptoms of Becker MD include tip toe walking, frequent falls, and difficulty rising from the floor. Calf muscles may appear large and healthy as damaged muscle fibers are replaced by fat, and muscle activity at times cause cramps in some individual. Cardiac and mental impairments are not as severe as in DMD.

3. **Congenital Muscular Dystrophy** refers to a group of autosomal recessive muscular dystrophies

that are either present at birth or become evident before age 2. They affect both boys and girls with hypotonic type of weakness.The degree and progression of muscle weakness and degeneration vary with the type of disorder. Weakness may be first noted when children show delayed milestones in motor function and muscle control. Muscle degeneration may be mild or severe and is restricted primarily to skeletal muscle. Most of the patients has no sitting or standing balance and some may never achieved walking ability.

There are three groups of congenital MD:

- merosin-negative disorders, where the protein merosin (found in the connective tissue that surrounds muscle fibers) is missing;
- merosin-positive disorders, in which merosin is present but other needed proteins are missing; and
- neuronal migration disorders, in which very early in the development of the fetal nervous system the migration of nerve cells (neurons) to their proper location is disrupted.

Defects in the protein merosin is mostly the main cause seen nearly in half of all cases of congenital MD.

Contractures (chronic shortening of muscles or tendons around joints, which prevents the joints from moving freely), scoliosis, respiratory and swallowing difficulties, and foot deformities are the complications seen in congenital muscular dystrophy. Some patients have normal intellectual development while others become severely impaired depending upon central nervous system affection, resulting in vision and speech problems, seizures, and structural changes in the brain.Respiratory failure is also seen due to weakness in diaphragm. Some children die in infancy while others may live into adulthood with only minimal disability.

4. **Emery-Dreifuss Muscular dystrophy** is X-linked recessive primarily affects boys with classic triads of:
    1. Early contractures especially of elbows, Achilles tendon, and posterior cervical muscles.
    2. Cardiac conduction defects
    3. Slowly progressive weakness and atrophy in a humeroperoneal distribution.

Onset of Emery-Dreifuss MD is usually apparent by age 10, but symptoms can appear as late as the mid-twenties. This disease causes slow but progressive wasting of the upper arm and lower leg muscles and symmetric weakness. Early onset of contractures before the onset of any significant weakness is classic presentation of this disorder. Contractures may cause elbows to become locked in a flexed position. The entire spine may become rigid as the disease progresses. Other symptoms include shoulder deterioration, toe-walking, and mild facial weakness. Serum creatine kinase levels may be moderately elevated. Nearly all Emery-Dreifuss MD patients have some form of heart problem by age 30, often requiring a pacemaker or other assistive device. Female carriers of the disorder often have cardiac complications without muscle weakness. Death results in mid-adulthood due to progressive pulmonary or cardiac failure.

Youth / adolescent-onset muscular dystrophies are classified in two types:

5. **Facioscapulohumeral MD (FSHD)** is an autosomal dominant with progressive weakness initially seen in muscles of the face (facio), shoulders (scapulo), and upper arms (humera). It is also known as Landouzy-Dejerine disease. Life expectancy is normal, but some individuals become severely disabled. Disease progression is typically very slow, with intermittent spurts of rapid muscle deterioration. Onset is usually in 15 years of age but may occur as late as age 40. Muscles around the eyes and mouth are often affected first, followed by weakness around the lower shoulders and chest. A particular pattern of muscle wasting causes the shoulders to appear to be slanted and the shoulder blades to appear winged. Although reflexes are impaired only at the biceps and triceps, muscles in the lower extremities may also become weakened. Changes in facial appearance may include the development of a crooked smile, a pouting look, flattened facial features, or a mask-like appearance. Some patients cannot pucker their lips or whistle and may have difficulty swallowing, chewing, or speaking. Other symptoms may include hearing loss (particularly at high frequencies) and lordosis, an abnormal swayback curve in the spine. Contractures are rare. Some FSHD patients feel severe pain in the affected limb. Cardiac muscles are not affected, and the pelvic girdle is rarely significantly involved. An infant-onset form of FSHD can also cause retinal disease and some hearing loss.

6. **Limb-girdle MD** refers to inherited conditions marked by progressive loss of muscle bulk and symmetrical weakening of proximal muscles, primarily those in the shoulders and around the hips. At least three forms of autosomal dominant limb-girdle MD (known as type 1) and eight forms of autosomal recessive limb-girdle MD (known as type 2) have been identified. Some autosomal recessive forms of the disorder are due to a deficiency of any of four dystrophin-glycoprotein complex proteins called the sarcoglycans.

The recessive limb-girdle muscular dystrophies occur more frequently than the dominant forms, usually begin in childhood or the teenage years, and show dramatically increased levels of serum creatine kinase. The dominant limb-girdle muscular dystrophies usually begin in adulthood. Rate of progression disease depends upon earlier appearance of clinical signs affecting both the genders. Some forms of the disease progress rapidly, resulting in serious muscle damage and loss of the ability to walk, while others have very slow progression over the years causing minimal disability, leading to normal life expectancy. At times, the disorder appears to halt temporarily in some cases, but later symptoms resume back.

Weakness is typically noticed first seen in proximal group of lower limbs affecting around the hips before spreading to the shoulders, legs, and neck. They have waddling type of gait and has difficulty in getting up from squatting position or low chair, climbing stairs, or carrying heavy objects. They have a history of frequent falls andare unable to run. Contractures are rare but can develop in the back muscles, which gives them the appearance of a rigid spine. Proximal reflexes are also affected. Cardiomyopathy and respiratory complications have been seen in limb girdle muscular dystrophy with intelligence remaining normal. They become severely disabled within 20 years of disease onset.

Adulthood onset of Muscular dystrophy are classified into three types:

7. **Distal MD,** also called distal myopathy, consists of a group of at least six specific muscle diseases that primarily affect distal muscles like in the forearms, hands, lower legs, and feet. Onset of distal MD is between the ages of 40 and 60 years affecting both genders. Distal dystrophies are less severe, with slow progression, and involve only fewer muscles. Distal MD can affect the heart and respiratory muscles, and patients may eventually require the use of a ventilator. Patients finds difficulty in performing fine motor activity with difficulty in straightening the fingers. As leg muscles become affected, walking and climbing stairs become difficult and some patients may be unable to hop or stand on their heels., The cause in one of distal MD is a muscle membrane protein complex called dysferlin which is suppose to be lacking.

Although distal MD is primarily an autosomal dominant disorder, autosomal recessive forms have also been reported in young adults. Symptoms are similar to those of DMD but with a different pattern of muscle damage. An infantile-onset form of autosomal recessive distal MD has also been reported with slow progressive weakness noticed around age 1, when the child begins to walk, and continues to progress with weakness throughout his adult life.

8. **Myotonic MD,** also known as Steinert's disease and dystrophia myotonica, is the most common adult form of Muscular Dystrophy, affecting both men and women with autosomal dominant inheritance. Myotonia, or an inability to relax muscles following a sudden contraction, is only seen in myotonic muscular dystrophy. People with myotonic MD has long life expectancy, with variable but slowly progressive disability. Onset of disease is seen between ages 20 and 30, but can develop earlier. Myotonic MD affects the central nervous system and other body systems, including the heart, adrenal glands and thyroid, eyes, and gastrointestinal tract. Muscles in the face and the front of the neck are the first to show weakness and may produce a haggard, "hatchet" face and a thin, swan-like neck. Wasting and weakness can be appreciated in forearm muscles with other symptoms including cardiac complications, difficulty swallowing, ptosis, cataracts, poor vision, early frontal baldness, weight loss, impotence, testicular atrophy, mild mental impairment, and increased sweating. Patients may also have tendency to feel drowsy and have an excess need to sleep.

Females may have irregular menstrual periods and may be infertile. The disease occurs earlier and is more severe in successive generations. A childhood form of myotonic MD appears at age between 5 and 10 years. with symptoms of generalized muscle weakness particularly in the face and distal muscles, lack of muscle tone, and mental impairment.

An expectant mother with myotonic MD can give birth to an infant with a rare congenital form of the disorder. Symptoms at birth may include difficulty swallowing or sucking, impaired breathing, absence of reflexes, skeletal deformities (such as club feet), and noticeable muscle weakness, especially in the face. Children may also experience mental impairment and delayed motor development. This severe infantile form of myotonic MD occurs almost exclusively in children who have inherited the defective gene from their mother, who may not know she is a carrier of the disease.

The inherited gene defect that causes myotonic MD is an abnormally long repetition of a three-letter "word" in the genetic code. In unaffected people, the word is repeated a number of times, but in people with myotonic MD, it is repeated many more times. This triplet repeat gets longer with each successive generation. The triplet repeat mechanism has now been implicated in at least 15 other disorders, including Huntington's disease and the spinocerebellar ataxias.

9. Oculopharyngeal MD (OPMD) generally begins in a fourth or fifth decade of life and affects both men and women. The disease is most common in families of French-Canadian descent and among Hispanic residents of northern New Mexico. Patients complaints of symptoms of drooping eyelids, followed by weakness in the facial and pharyngeal muscles in the throat, causing difficulty in swallowing. The tongue may atrophy and changes to the voice may occur. Eyelids may droop so dramatically that they compensate by tilting back their heads. At times they have double vision and problems with upper gaze, while others may have retinitis pigmentosa (progressive degeneration of the retina that affects night vision and peripheral vision) and cardiac irregularities. Muscle weakness and wasting in the neck and shoulder region is predominately seen with limb muscles also being affected. Persons with OPMD may find it difficult to walk, climb stairs, kneel, or bend and with those persons most severely affected will eventually lose the ability to walk.

Stages of Muscular dystrophy:

As some muscular dystrophies are progressive in their disease course while others are very slow; these disease can be divided into stages according signs and the pattern of involvement.

Stage 1: Early Pre-symptomatic :

During Presymptomatic stage, diagnosis remained unnoticed unless there is some positive or strong family history or unless for some reasons any blood investigations are done. Symptoms of delay in early developmental milestones like crawling, walking and in speech remained unnoticed with child been flaccid and less physically active than normal children. Given a new diagnosis like Muscular dystrophy shatters the families and there comes sensitive counselling and support which can influences the family's ability to manage the child in the future.

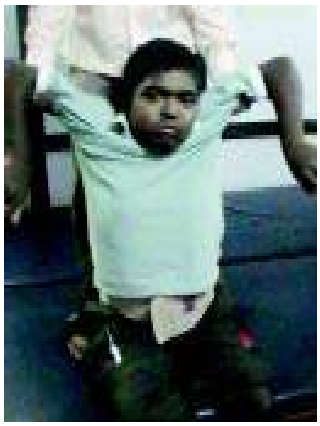
Stage 2 : Early Ambulatory (Walking)

Boys shows classic traids like **Gower's Manoeuvre** in which the child gets up from the floor by using his arms to crawl up his own legs,Has waddling type of gait and has toe walking with difficulty in activities such as running, hopping and climbing stairs.

Stage 3: Late Ambulatory (going off feet)

Mobility becomes labourous with difficulty in climbing stairs and walking distances. They present with frequent falls and have difficulty or unable to getting up on their feet without assistance. Fractures are the main cause because of frequent falls resulting into loss of mobility. Also early involvement of scapular stabilizers results in decrease in arms and hands movements during reachout activities. **Classic test of scapular stability called Meryon sign** where child slips from examiner's grip as the child is being lifted from under the arms.

Wrong pattern of lifting.



Meryon's sign.

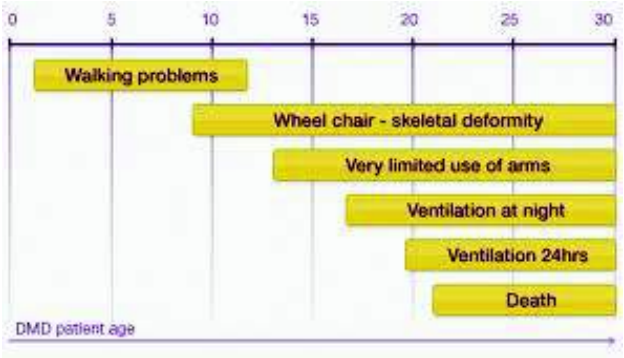
Stage 4 : Early Non-Ambulatory

Boys are able to sit up of their own independently but are unable to walk. In this stage, wheelchair becomes a main aid for propulsion where they can self propel for some period of time. Good postural management is required as most of time they will be maintaining sitting posture. Since there is poor trunk control because of muscle imbalance, there is increased risk of developing scoliosis and management of same is mandatory.

Stage 5: Late Non-Ambulatory

Postural imbalance and malalignment is the main concern seen in this stage with decrease in upper limb function which results in decline on performing ADLS like dressing, feeding and difficulties with oro-motor function can lead to nutritional problems. Because of no mobility happening, there is impairment seen in respiratory and cardiac functions and require interventions for same.

Stage 6: Palliative Care/ End of Life.



Life span of DMD.

Functional Transitions in patient with Muscular Dystrophy.

1. Walks with mild waddling gait with lordosis. Running becomes strenuous but can ascend, descend steps, curbs.
2. Walking with moderate waddling gait with lordosis. Running becomes impossible. Climbing stairs and curbs becomes difficult. Uses Gower's maneuver while getting up from floor but can rises from chair independently.
3. Walks with moderately severe waddling gait and lordosis. Can raise himself from chair independently but ascending descending curbs or stairs or rise from floor becomes totally dependent.
4. Walks with assistance or with bilateral KAFOs. Can have surgical release for contractures. Can propel manual chair slowly and wheelchair propulsion for community mobility. Independent in bed and self -care even though some help is needed in dressing and bathing because of time constraints.
5. Unable to walk independently but can bear and shift weight to walk with orthoses and can transfers from wheelchair independently. Can propel self in manual chair but with limited endurance whereas motorized wheelchair becomes more functional. Independent in self care with transfer assist for bath or shower.
6. Independence in motorized wheelchair but requires trunk support or orthosis. Assistance is needed in bed and with major dressing, is independent for performing self -grooming but dependent for toileting and bathing.
7. Independence in motorized wheelchair but need to recline intermittently while in chair. Needs assistance for hygiene and most self care requiring proximal upper limb control.

8. Use both hands for single hand activities, can perform simple table- level hand activities, and can perform self-feeding with arm support.
9. Can sit in wheelchair only with trunk support and intermittent reclining or transfer to supine position. May require nighttime ventilatory support or intermittent daytime PPV. Some hand control can be achieved if arms are supported and needs help for turning at night.
10. Totally dependent and cannot tolerate upright position and needs home ventilatory support if needed for prolonged ventilation, then tracheostomy is needed.

Diagnosis:

To diagnose any muscle disease thoroughly medical and a complete family history should be reviewed to determine whether it is secondary to a disease affecting other tissues or is an inherited condition or it is result of prior surgery, exposure to toxins, or current medications that may affect the patient's functional status.

The first suspicions are usually raised by one of the following three signs (even when there is no history of DMD in the family):

1. Cluminess in Movements :

Boys who have DMD walk slower and are less active than other boys of their age group. They have pseudohypertrophy of calf muscles and have difficulty in running, jumping or climbing stairs. They have frequent falls and have a tendency of toe walking. They may also have a speech delay. The classic signs likethe "**Gowers'" manoeuvre or sign,** where the boy uses his hands and arms to "walk" up his body in order to push himself to an upright position. This is due to weakness in the hips and thigh muscles and Meryon sign in which the child slips from examiner's grip as the chld is being lfted from under the arms.

2. High levels of the muscle protein creatine kinase (CK) in a blood test :

Serum creatine kinase is elevated more than 100 times normal in early stages of disease.Elevated CK is evident at birth long before symptoms are evident and CK levels decrease over time with loss of muscle mass.

- Some have delay in their speech development also.

Investigations:

Laboratory tests such as:

1. **Blood and urine tests** can help in identifying defective genes which are responsible for identify specific neuromuscular disorders. For example:. Serum aldolase level, an enzyme involved in the breakdown of glucose, is measured to confirm a diagnosis of skeletal muscle disease.

   Creatine kinase is an enzyme that leaks out of damaged muscle and its elevated levels may indicate muscle damage, before physical symptoms become apparent in the early stages of Duchenne and Becker MD. Testing can help in identifying whether a young woman is a carrier of the disorder or not.

- Myoglobin is an oxygen-binding protein found in cardiac and skeletal muscle cells and measured when injury or disease in skeletal muscle is suspected. High blood levels of myoglobin are found in patients with MD.
- Polymerase chain reaction (PCR) / Molecular diagnosis or Genetic testing: can detect mutations in the dystrophin gene method for generating and analyzing multiple copies of a fragment of DNA.
- Serum electrophoresis is a test to determine quantities of various proteins in a person's DNA.
- **Electron microscopy:** can identify changes in subcellular components of muscle fibers that characterize cell death, mutations in muscle cell mitochondria, and an increase in connective tissue seen in muscle diseases such as MD.

Changes in muscle fibers that are identified in a rare form of distal MD using an electron microscope.

Exercise tests can detect elevated rates of certain chemicals and are used to determine the nature of the MD or other muscle disorder following exercise. It also assess muscle strength by measuring muscle function against gravity and detecting slight muscle weakness. If weakness in respiratory muscles is suspected, respiratory capacity may be measured by having the patient take a deep breath and count slowly while exhaling if respiratory muscles are involved.

Genetic testing identifies genes which are

responsible for the cause or associated with inherited muscle disease. DNA analysis and enzyme assays can confirm the diagnosis of certain neuromuscular diseases, including MD. Genetic linkage studies can identify whether a specific genetic marker on a chromosome and a disease are inherited together. They are particularly useful in studying families with members in different generations who are affected.

Genetic counselling can help parents who have a family history of MD determine if they are carrying one of the mutated genes that cause the disorder. Two tests can be used to help expectant parents find out if their child is affected.

- **Amniocentesis**: is usually done at 14-16 weeks of pregnancy, where a sample of the amniotic fluid in the womb for genetic defects is taken as the fluid and the fetus have the same DNA.
- **Chorionic villus sampling or CVS**: involves the removal and testing a small sample of the placenta during early pregnancy.

Magnetic resonance imaging (MRI) is a noninvasive, painless procedure,used to examine muscle quality, any atrophy or abnormalities in size, and fatty replacement of muscle tissue, as well as to monitor disease progression.

Other forms of diagnostic imaging for MD include phosphorus magnetic resonance spectroscopy, measuring cellular response to exercise and the amount of energy available to muscle fiber, and ultrasound imaging (also known as sonography), uses high-frequency sound waves to obtain images inside the body and can be used to measure muscle bulk.

Muscle biopsies are used to monitor the course of disease and treatment effectiveness. The muscle specimen is examined to determine whether the patient has muscle disease, nerve disease (neuropathy), inflammation, or another myopathy. Muscle biopsies also assist in carrier testing. With the advent of accurate molecular techniques, muscle biopsy is no longer essential for diagnosis. In muscular dystrophy, degeneration with gradual loss of fiber, variation in fiber size & proliferation of connective and adipose tissue is seen.

Immunofluorescence testing can detect specific proteins such as dystrophin within muscle fibers.

Histochemical studies indicate loss of subdivision into fiber types, with a tendency toward type 1 fiber predominance.

Neurophysiology studies can identify physical and / or chemical changes in the nervous system.

- **Electromyography (EMG)** can record muscle fiber and motor unit activity. Increase in insertional activity is seen in early stage of disease which can decrease later as fibrotic tissue replaces muscle and is characteristic of MD. Patterns of low-amplitude, short-duration polyphasic motor unit action potentials is seen in EMG with patients of Muscular dystrophy affection.
- **Nerve conduction velocity studies** measure the speed with which an electrical signal travels along a nerve. Sensory NCSs are normal and Motor NCSs have normal latencies,conduction velocities and F-wave latencies, but amplitude of compound muscle action potential (CMAP) decreases as disease progresses.
- **Repetitive stimulation studies** involve electrically stimulating a nerve 5 to 10 times, at a frequency of 2-3 shocks per second, to study muscle function.

Medical Management:

There is no specific treatment that can stop or reverse the progression of any form of MD. All forms of MD are genetic and cannot be prevented. Treatment is aimed at keeping the patient independent for as long as possible and preventing complications that result from weakness, reduced mobility, and cardiac and respiratory difficulties. Treatment may involve a combination of approaches, including physical therapy, drug therapy, and surgery.

Assisted ventilation is required to treat respiratory muscle weakness especially seen in the later stages. Oxygen is fed through a flexible mask to keep the lungs inflate fully. some patients may need overnight ventilation when respiration becomes more strenuous. A mask worn over the face is connected by tube to a machine that puts out intermittent bursts of forced oxygen. Obese patients with Duchenne MD may develop obstructive sleep apnea and require nighttime ventilation. Patients on a ventilator may also require the use of a gastric feeding tube.

Drug therapy may be prescribed to delay muscle degeneration.

Corticosteroids such as prednisone can slow the rate of muscle deterioration in Duchenne MD and help children retain strength and prolong independent walking by as much as several years. However, these medicines have side effects such

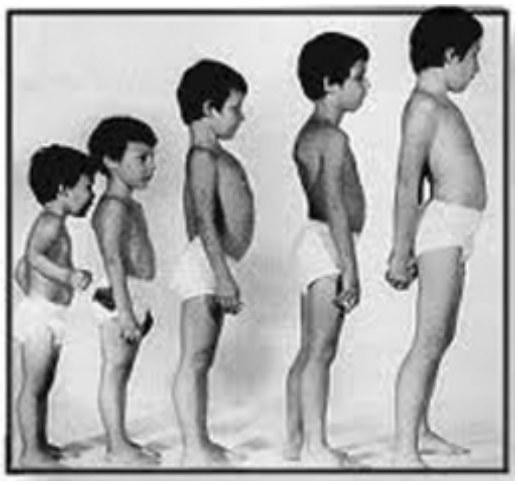
as weight gain and bone fragility that can be especially troubling in children. **Immuno-suppressive** drugs such as cyclosporin and azathioprine can delay some damage to dying muscle cells.

Drugs that may provide short-term relief from myotonia (muscle spasms and weakness) include mexiletine; phenytoin; baclofen, which blocks signals sent from the spinal cord to contract the muscles; dantrolene, which interferes with the process of muscle contraction; and quinine.

Anticonvulsants, also known as antiepileptics, are used to control seizures and some muscle activity. Commonly prescribed oral anticonvulsants include carbamazepine, phenytoin, clonazepam, gabapentin, topiramate and felbamate.

Respiratory infections may be treated with antibiotics.

PATTERNS OF WEAKNESS AND COMPENSATION ACCORDING TO STAGES WISE:



Progression of Weakness.

Early stage:

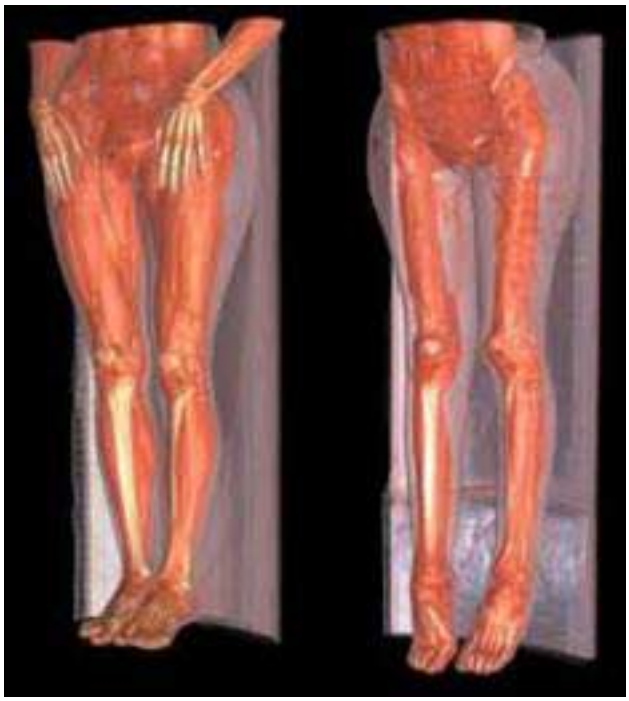
Weakness seen in hip extensors, ankledorsiflexors, hip abductors, hip adductors, abdominals, neck flexors, shoulder depressors, extensors and abductors and elbow extensors.

Compensation :

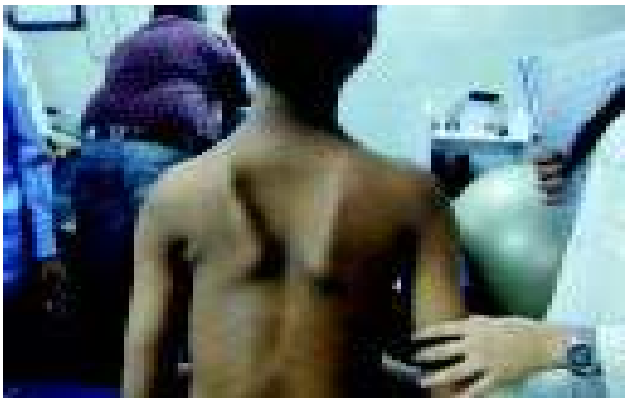
1. Increased lumbar lordosis to keep force line behind hip joint.
2. Lack of heel strike.
3. Foot may be pronated and everted.
4. Hip waddling gait
5. Decreased in Cadence.
6. Gower's Maneuver.

Standing posture.

Waddling gait.



Normal standing vs Muscular dystrophy.



Winging of scapulae.

Most comfortable position.

Middle stage:

Weakness seen in quadriceps and ankle evertors in addition to muscles mentioned above in early stage.

Compensation:

1. Line of gravity infront of knee joint and behind in hip joint.
2. Base of support widens to maintain balance and due to tightness in iliotibial band
3. Increase ankle plantarflexion and equinus posture to avoid knee flexion.
4. Frequent falls.
5. Strong action of tibilias posterior results in inversion attitude at ankle.

Tightness develops in iliotibial bands & tensor fascia lata, hip flexors, hamstrings, gastrosoleus and posterior tibialis.

Functional Losses: All activities against gravity like ability to rise from floor, stair climbling and rising from chair.

Compensated Gait: Standing and toe walking with wide base of support and extreme lumbar lordosis, extreme lateral trunk lurch and upper limb abduction.

Prescription of long leg braces like KAFO with or without surgery should be done to prolong ambulation.

Late or Non-Ambulatory Stage :

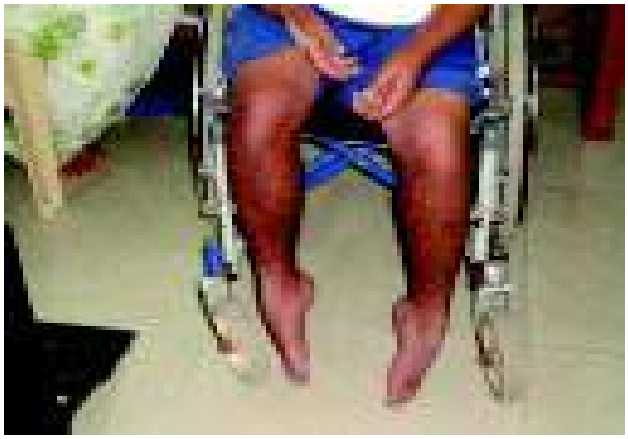
Weakness in upper limb interferes functional, elbow extensor, forearm supinator, wrist and finger extensor with above mentioned muscles in two stages. Distal hand functions are preserved.

Compensation: Here in this stage compensation are used to maintain upright posture and facilitate ambulation, achieve support and stability in sitting and upper limb function.

Tightness is seen elbow flexors, pronators, wrist and finger flexors and neck extensors results in development of upper limb contracture.

Scoliosis results as there is more of sitting posture with loss of ambulation.

Loss of functional activity is seen in upper limb activities, sitting ability and in doing ADLS.



Talipes equino varus position.

COMPLICATIONS:

1. **Scoliosis** can be explained into two phases: one in ambulatory phase and non-ambulatory phase.

During ambulatory stage, scoliosis is flexible, functional and is minimized by protective spinal hyperextension and lateral trunk lurching.

Factors that influence whether or not scoliosis appears prior to final loss of ambulation:

1. Age at which walking ceases.
2. Intervention used or not used to prolong ambulation.
3. Final gait pattern.

In non-ambulatory phase, scoliosis becomes more prominent as there is constant use of wheelchair and no ambulation. It can lead to decline in pulmonary function, upper extremity function and sitting ability.

Sign of scoliosis

Scoliosis.

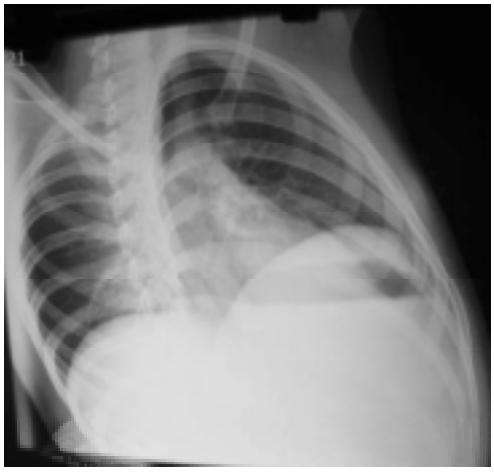
Factors that results in the development of scoliosis are as:

1. Factors that make spine vulnerable

   Severe symmetrical weakness in trunk musculature. Rapid vertebral growth during adolescent coincides with loss of ambulation. Loss of protective spinal hyperextension.

2. **Factors that initate asymmetry.**

   Compensatory movement pattern used for stability, upper limb function, pelvic position and lower extremity position. The deforming force of gravity on vertebral column increase in presence of asymmetrical spine and pelvic alignment that compromise the simple mechanical ability of vertebral column to withstand the force of gravity. Inaddition, resultant unequal distribution of weight on epiphyseal growth plates increased the potential for an initial flexible scoliosis to become structural.



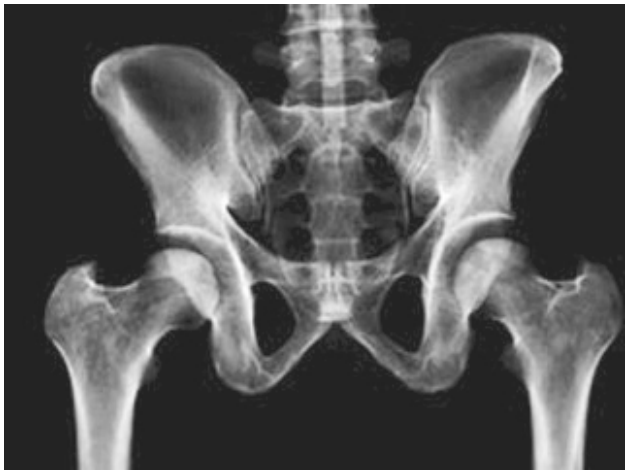
X-Ray showing scoliosis

2. **Obesity:**

   Since the muscles are replaced by the fat and fibrous tissues in later stage of disease with the loss of ambulation, there is less calorie expenditure resulting into obesity.

Obesity.

3. Osteoporosis:



Osteoporosis.

It is more common in vertebral column than in long bones, worse in lower extremities than in upper extremities and is aggravated by steroids.

4. Respiratory Affection.

It is compromised by no. of factors:

1. Decrease in thoracic and spinal mobility due to progressive muscle weakness leading to replacement into fibrous tissue and restricted pattern of breathing.
2. Asymmetrical breathing pattern due to muscle weakness.
3. Total lung capacity, vital capacity and forced inspiratory and expiratory abilities decreases and residual volume increases.
4. Breathing becomes strenuous from initial shallow breathing to more rapid breathing to less chest or lung volume / expansion leading to decline in breathing volume.

   Decrease in lung expansion leads to areas of lung collapse and weakness in abdominals and in muscles of forced expiration results in decreased coughing.
5. **Cardiac Involvement :**
   Cardiac muscles are too affected by dystrophin. Cardiac involvement can also compromise by scoliosis and respiratory status. It is so progressive that it shows ECG abnormalities, hypertrophic cardiomyopathy and dilated cardiomyopathy. Cardiac abnormalities may also include AV block, atrial paralysis, atrial fibrillation or flutter, ventricular arrhythmia, conduction defects and reduced ejection fraction.

Physical Therapy Assessment:

Pathokinesiology :

Imbalanced muscle weakness, compensatory movement patterns, postural habits and influence of gravity add to weakness progression in a pattern of proximal to distal. As weakness increases, postural alternations and movement which produced mechanically locked joints thereby substituting for adequate muscle strength. The substitutor are responsible for optimizing function eventually leading to muscle tightness, contracture and deformity thereby increasing disability.

In Muscular dystrophies, assessment is ongoing process where specific type of weakness and tightness and compensation are identified and interventions should be designed inorder to maximize strength, prevents deformity and provide effective adaptive functioning.

1. Postural alignment.
2. Range of motion
3. Manual muscle testing
4. Girth Measurement
5. Respiratory status
6. Activities of Daily living
7. Gait
8. Functional Status.
9. Transfers
10. Orthosis / Casting / Bracing
11. Mobility status:
12. Wheelchair mobility: Manual
    Motorized
13. Physical environmental and Accessibilty.

Outcome Measures:

1. Vignos Functional Rating Scale.
2. Brooke's clinical protocol.
3. WeeFIM.
4. Pediatric evaluation of disability inventory.
5. Muscle, Pulmonary and ROM testing.
6. GSGC Assessment which includes noting timed tasks in 10 m timed walkng, Stair climbling, sit to stand from chair and rising from floor.

Physiotherapy Interventions for Muscular Dystrophy:

Physiotherapy in Early stage:

1. Educating the family about the condition and coping strategies
2. Prevention of contractures and deformity which can further lead to disability and pain.
3. Maintenance of maximal strength to prevent disuse atrophy.
4. Maintenance of maximal functional capabilities by using appropriate adaptive equipment
5. Emphasizing home programs.
6. Interventions required to maintain ambulation.

Physiotherapy in Middle stage Following loss of ambulation:

1. Continuation of early stage program.
2. Spinal care and management to prevent further deformity because of loss of ambulation.
3. Transfers requiring less energy expenditure should be taught.
4. Educating about body mechanics to further prevent the contractures and deformity.
5. Adaptive devices to modified ADLS.

Physiotherapy in Later stage :

1. Continuation of above program.
2. Evaluating the endurance and fatigue level required for maintaining any posture or activity.
3. Maximizing Upper limb function.

Short term Goals:

1. To increase / maintain range of motion.
2. To increase / maintain strength & endurance
3. To promote optimal body alignment & symmetry.
4. To minimize compensatory movement, patterns & position used for function.
5. To prevent the development of scoliosis.
6. To maintain functional ambulation.
7. To maintain sitting ability.
8. To maintain functional, independent mobility throughout all phases of disease progression.
9. To provide active respiratory program and to establish & maintain most effective breathing pattern.
10. To maintain chestwall mobility.
11. To strengthen respiratory muscles & develop endurance.
12. To To teach principles of pulmonary hygiene & assisted coughing.
13. To help preserve maximal hand function.
14. To teach caregivers proper handling & transfer techniques

Long term Goals:

1. To prevent deformity & contracture.
2. To maximize & maintain strength & endurance within the limits of fatigue.
3. To maximize & maintain respiratory status.
4. To maintain ambulation.
5. To maintain functional mobility.
6. To maintain highest possible level of functional independence using adaptive equipment & orthotic devices.
7. Provide patient, family & caregivers with timely information helping in overall management of disability.

As DMD represents with severe muscular weakness resulting in contractures of the hip flexors and plantar flexors which later interfere with ambulation and as walking mobility is lost, contractures of the hips, knees, ankles, shoulders, wrists, and fingers set in quickly. Loss of ambulation leads to another major complication of the development of scoliosis which will lead to severe thoracic distortion comprising with respiration. Again, a loss of ambulation results in reduced caloric expenditure resulting to obese. Obesity along with loss of strength may lead to pressure sores, as there are unable to do wheelchair push-ups or move the body for extended periods. As there is complete loss of mobility, respiratory complications sets in resulting to death.

Exercise will be proven beneficial as increase in strength will improve performance of daily activities such as stair climbing, chair rising, and walking. To stretch the contractures, flexibility

program should be included to reduce the progression of contractures, thus allowing persons with DMD to ambulate longer. Finally, strengthening of postural muscles may decrease the formation of scoliosis. Increased energy expenditure due to exercise may reduce the prevalence of obesity as there is no ambulation.

Endurance

In one of the animal studies, results showed that mdx mice ran significantly less distance per week and ran at a significantly slower daily speed compared to normal mice and when exercise was administrated it showed improvement in resistance to fatigue, increased proportion of oxidative fibers, and improvements in force production of the soleus and extensor digitorum longus of exercised mdx when compared to sedentary mdx mice.

Muscular Strength

A certain amount of muscle activity has been thought to be beneficial in preventing disuse atrophy, maintaining strength, functional status and flexibility. Studies proving which type of muscle activity can be beneficial or detrimental is limited.

Eccentric exercise increases muscle damage thus it should be eliminated, i.e., dynamic constant external resistance exercise, stepping down off a step as it results in fiber splitting,vacuoles & necrosis with proliferation of connective tissue as well as fiber hypertrophy. Recent report also shows that isokinetic eccentric exercise did not elevate CK in healthy individuals. With dynamic constant external resistance, (ex. machines or free weights) there may be an increase in muscle damage with the change in the direction of the velocity at the point where the concentric contraction is followed by an eccentric contraction whereas submaximal isokinetic exercise if given in early stage of disease is beneficial when the strength was at its maximum.

General Exercise Guidelines

	Type	**Frequency**	**Intensity**	**Duration**
Flexibility	Passive / Active	Daily	Low	3x (10-30 sec)
Endurance Exercise	Walking, Cycling, Swimming	Variable 1-7 x per week	Low	Variable 1-20 minutes
Muscular Strength	Isokinetic, Concentric, or Eccentric only	Variable 1-5 x per week	Low Low	Variable 1-3 sets 5-15 repetitions

Specific Exercise Guidelines

	Muscle Groups
Flexibility	Hip flexors, Shoulder extensors, Elbow flexors, Plantar flexors, Intrinsic hand flexors, Knee flexors
Endurance Exercise	Muscles used for ambulation
Muscular Strength	Hip extensors, Elbow extensors, Shoulder flexors, Dorsiflexors, Hand and wrist extensors, Back extensors Abdominals.

Precautions and Contraindications to Exercise

Muscle damaging eccentric exercise (i.e., downhilll running or walking)

1. Ballistic stretching
2. Sitting or lying for extended periods of time.

Stretching and Positioning to prevent deformity:

Effects of chronic positioning, unopposed influence of gravity and imbalanced muscle activity around the joints contribute to development of hypo extensibility and which can be prevented by adequate positioning by daily ROM / stretching and use of splinting, casting and standing on standing board.

Mostly severe contractures are seen in 2 joint muscles and those which are maintaining posture.

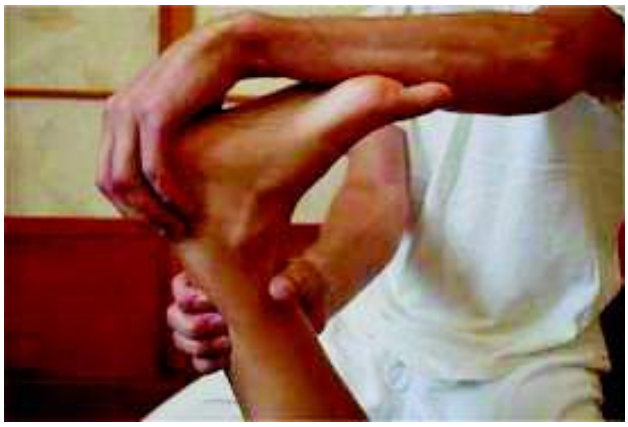
In upper limb:
1. Elbow flexors.
2. Forearm pronators.
3. Wrist / Finger flexors.

In Lower limb:
1. Illiotibial band.
2. Tensor fascia lata.
3. Hip fexors.
4. Hamstring
5. Gastrosoleus
6. Posterior tibialis

In spine: Neck extensors.

Interventions:

1. **Passive stretching:** should be done daily and is best achieved by standing.



Soleus stretching

PNF hamstring

Tendoachilles Stretching

Knee flexor stretching

Psoas_Stretch

Hip flexor stretching in Prone

2. **PNF-** Contract Relax to improve flexibility
3. **Joint Mobilization -**Traction to all joints.
4. **Myofascial release techniques.**
5. **Modalities:** Hot packs to increase plasticity and comfort but should be avoid excessive generalized heat which can induce fatigue and reduce strength.

Strengthening:

Submaximal levels of isokinetic and aerobic exercise have been practiced inorder to prevent disuse atrophy and to maintain residual strength while avoiding overwork weakness. Exercise in early stage of disease proves to be beneficial as long periods of rest and immobility results in functional loss.

Loss of muscle strength is seen in:

1. Neck flexors.
2. Abdominals
3. Shoulder girdle musculature especially deltoids, latissimus dorsi, stabilizers.
4. Pelvic girdle musculature especially gluteals.
5. Knee extensors.
6. Ankle dorsiflexors.

Strengthening can be achieved by

1. Maintaining proper alignment thereby maximizing the muscle work and biomechanical advantage.
2. Positioning and supporting the muscle in their optimal length so that they are functional.
3. Movement facilitation.
4. Submaximal exercises.
5. PNF active assist.
6. Functional movements which are used in ADLS.

Exercises can be categorized according to the stagewise depending upon:

1. Remaining strength in the available muscles.
2. Ability to perform the functional activity.
3. Endurance.

In Early stage:

muscle power remains functional i.e more than Grade 3 but has difficulty in climbling stairs and getting up from floor showing weakness especially in gluteal muscles and abdominals muscles.

Exercises to strengthen these muscles in early stage compromises:

1. **Bed mobility which includes:**
    1. Rolling.
    2. Leg rotation.
    3. Abdominal and back strengthening exercises.
    4. Bridging.
    5. Quadrapud.
    6. Cat and camel exercises in Quadrapud.
    7. Hip extension in Quadrapud.
    8. Side sitting in Quadrapud.
    9. Kneeling
    10. Kneel walking
    11. From quadrapud coming to kneeling position.
    12. Trunk rotation in kneeling.
    13. Hip extension in prone.
    14. Hip flexion in supine.
    15. Hip abduction in sidelying.
    16. Strengthening of scapular and neck muscles.

Rolling - 1

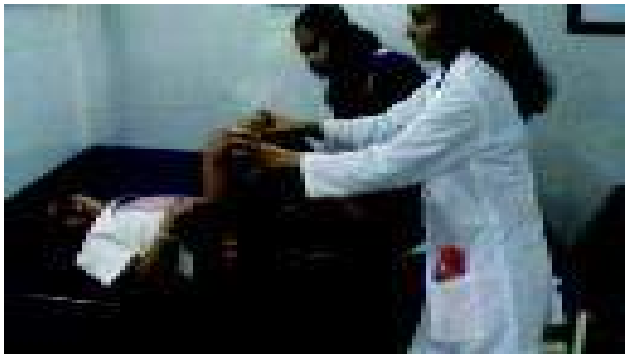
Rolling - 2

Rolling - 3

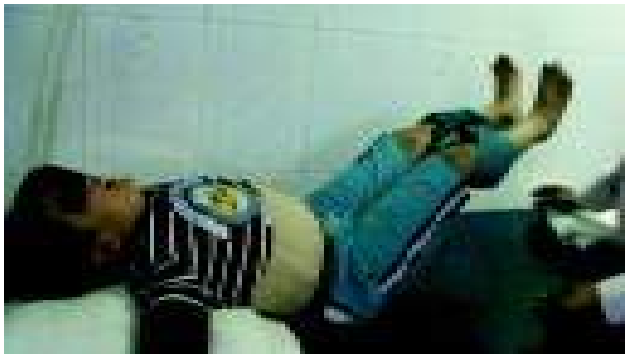
Bridging

Leg Rotation-1

Leg Rotation-2



Grade-II Lower Abdomen-1



Grade-IV Lower Abdomen-2

Upper Abdomen

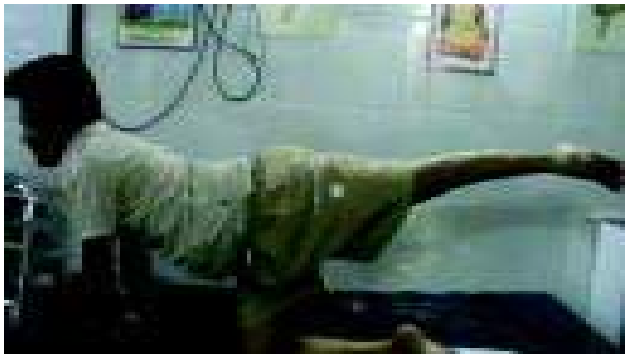
Back Extension

Reachouts in Quadrapud

Crawling

Cat camel exercises in Quadrapud

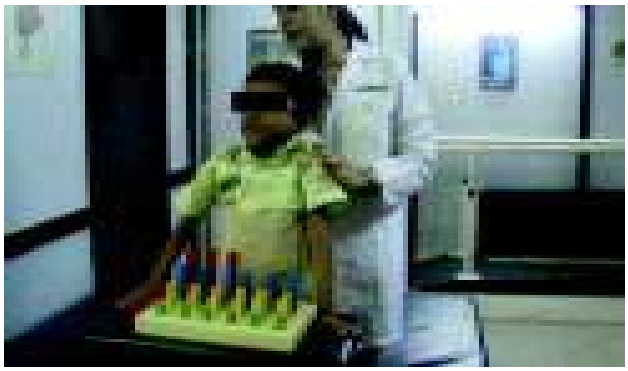
Cat camel exercises in Quadrapud 1



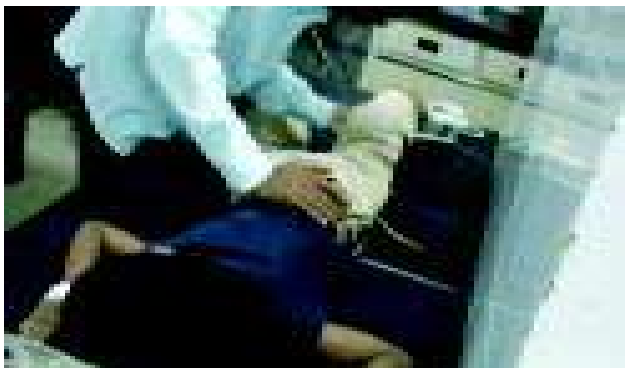
Hip extension in Quadrapud

Hip extension in Quadrapud

Kneeling - 1



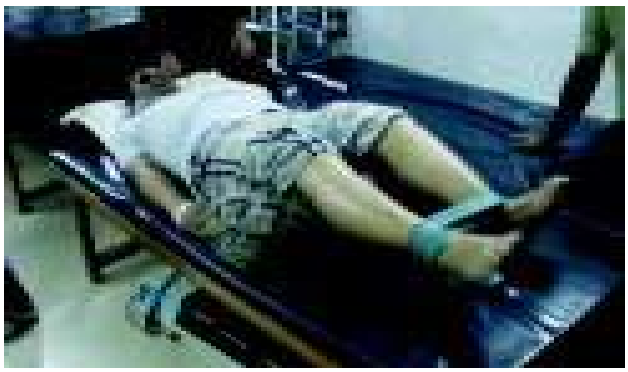
Kneeling - 2



Hip Extension in Prone

Hip abduction in sidelying

Active assisted Hip flexion



Knee extension Strengthening

Hip extension on Swiss Ball

Back Extension on Swiss Ball

Resisted knee flexion

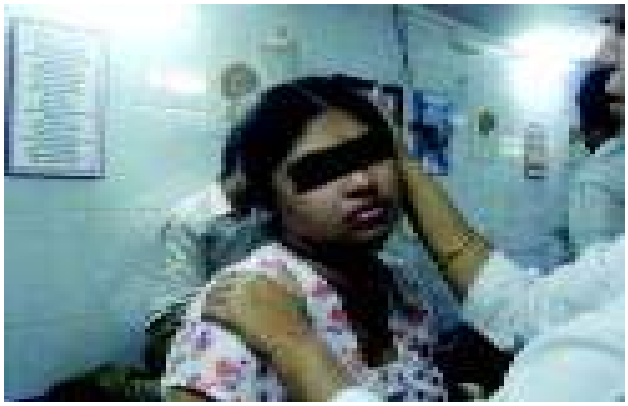
One leg bridging

Balancing exercises in sitting

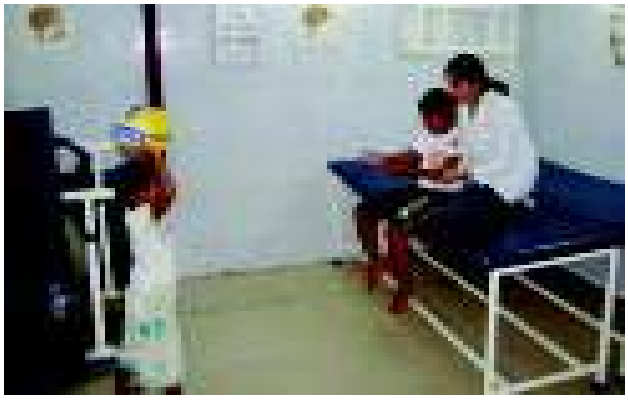
Balancing exercises in sitting(1)

Going to lying position

Going to lying position 1



Neck strengthening exercises



Overhead activities

Rhomboids Strengthening with wand and theraband

2. Getting up from low stool or chair.
3. Quadriceps drill.
4. Stepping exercises with the low stepper.
5. Swiss ball exercises to strengthen the trunk and hip musculature.
6. Hydrotherapy.

In Middle Stage:

Patients shows the muscle power around Grade 2++ to Grade 2- with frequent falls while walking as patient develops toe walking because of tendoachilles contracture and lumbar lordosis compensating for the weakness of hip extensors and abdominals and difficulty in getting up from floor and rising from chair showing quadriceps weakness. Initial weakness starts developing in upper limb with difficulty in doing overhead activities.

Exercises in this stage includes strengthening of muscles and stretching of tight muscles.

Exercises such as:

1. **Suspension therapy exercises** for strengthening the hip, knee and trunk muscles from Grade 2- to Grade 3-.
2. **Active assisted exercises** for abdominal and back musculature.
3. **Quadrapud position** achieved with the help of elbow splint for developing control on the shoulder, trunk and hip muscles.
4. **Cat and camel** exercises and weight shifting in forward, backward and sideways direction exercises are included.
5. **Kneeling position** is achieved with the help of elbow splint and foot stool infront.
6. **Getting up** from sidelying position.
7. **Standing** with the help of push knee splint and high boots with posterior steel shank and walker or wall support for an half an hour to one hour to stretch the knee flexor contracture and Tendoachilles contracture.
8. **Walking** is prolonged either with push knee splint and high boots with posterior steel shank and walker or with KAFO.
9. **Stretching** the hip flexor contracture in pronelying position for about half an hour to one hour and constant splinting the lower limbs at night to prevent the legs going into hip abduction external rotation, knee flexion and foot into talipes equino varus position.

Late or Non-Ambulatory Stages:

In this stage the muscle is almost replaced by fat and fibrous tissue thereby muscle power showing Grade 1+ with only flicker contraction at times to Grade 0. The child becomes wheelchair bound with spinal deformities developing later. Functionally the child becomes totally dependent for all his ADLS.

In this stage, exercises such as:

1. Breathing exercises are incorporated to improve the strength of the respiratory muscles and to build up the endurance.
2. Suspension therapy exercises and passive - assisted exercises are included inorder to maintain ROM and strength in available muscles.
3. Stretching to prevent further worsening of the contractures.
4. Standing on a standing board compulsory for an hour twice in a day for physiological benefit.
5. Transfering the child in this stage is difficult as holding the axilla and lifting is almost impossible. The only way is to lift him by holding the child with both the arms around, lift him and rotate.

Orthosis and casting : Fig 21.

1. At night or during daytime while taking rest to maintain proper positioning.
2. Ankle foot orthosis(AFO) for stretching plantarflexors.

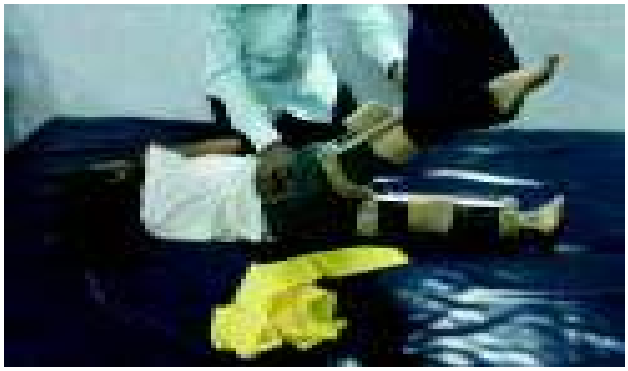
Assisted Bridging with bedsheet

Wt shifting in Quadrapud

Suspension exercises-Hip abduction adduction

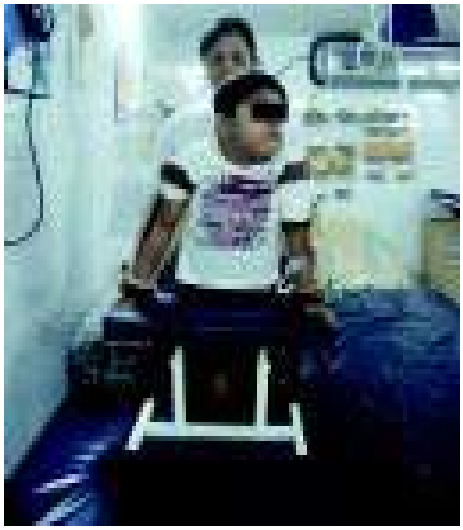
Suspension exercises-Resisted Hip adduction

Suspension exercises-Resisted Hip flexion



Suspension-Hip extension

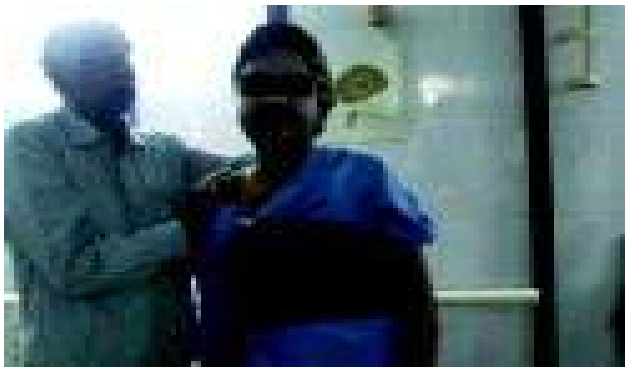
Suspension exercises-Knee extension



Kneeling with support of foot stool and elbow splint

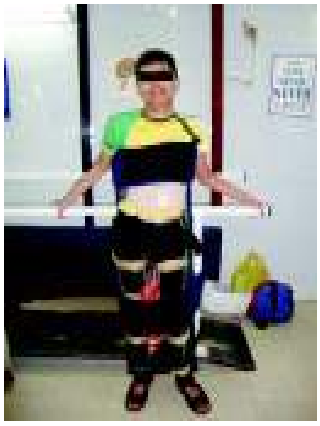
Elbow flexion with wand

Upperlimb strengthening with theraband



Neck side flexion exercises

Neck rotation exs



Standing on standing board (2)

Walking with push knee splint

Lifting 1 Lifting 2 Lifting 3 Lifting 4

3. Knee extension splints for stretching knee flexor contractures.
4. Wrist and finger splints for wrist and finger flexor stretching.
5. Serial Casting.

Positioning:

1. Prone lying.
2. Supine -Tieing both the thighs to avoid the leg to go to abduction.

Standing :

1. With long leg braces (KAFO) on tilt table or standing board to minimize osteoporosis and to prevent lower extremity contractures.ss

Emphasis on Standing and Ambulation with KAFO :

Whether to release the contractures and then give braces for walking or straightway give braces and make him walk till he could prolong with walker.

Importance of standing with braces:

Standing promotes functional status in walking & ADL.

Lower extremity contractures are stretched resulting in release of stiffness, and flexibility.

Standing results in minimization of severe osteoporosis.

Standing provides weight control.

It improves cardiovascular and cardiopulmonary functions.

Standing delays the development of scoliosis.

Wheelchair: When independent or dependent walking i.e with or without orthosis becomes difficult, wheelchair as a mobility orthosis is used as it acts like a functional mobility aid.

Usually self propelled wheelchair is given to a child to used it in indoors as this increases the chest expansion and muscle power of upper limb. When the patient is unable to use the self-propelled wheelchair in later stages of disease, then powered or attendant propelled wheelchair can be used. Wheelchair should be light weight, strong and should provide good sitting posture to the patient leaving minimum space on either side of the sitting area. Feet should be resting at 900 to prevent equines deformity and the armrest of the chair should be at proper height so that when the elbows rest over arm rest and the shoulders are in relaxed position.

Spinal Management

1. It is required to prolong ambulation and standing.
2. To promote spinal extension in sitting.
3. To maintain symmetry while sitting in wheelchair.
4. To optimize upper extremity function in symmetrical pattern.

Interventions:

A) Sitting posture:

1. Proper sitting posture in wheelchair should be attained to avoid any compensation happening at spine level. One should be constantly leveling his pelvis without rotation. If scoliosis develops, a spinal jacket is prescribed inorder to prevent further increased in curvature. Avoiding kyphotic posture, maintaining lower extremity position with no hip abduction and having proper foot placement.
2. Evaluating all functional activities which can produce asymmetrical movement patterns.

B) Standing posture:

1. To help in controlling Lower extremity contracture.
2. To promote spinal extension in standing posture on standing board or tilt tables.
3. To optimize more physiological benefits.

Parents and caregivers should be asked to monitor symmetry and asymmetry posture attained during sitting or standing and correcting those by visual feedback periodically or by changing position or support while maintaining those postures.

Surgical Management:

Segmental instrumentation in spine allows stabilization with immediate postoperative mobilization with no external support required whereas in lower limbs subcutaneous release of Achilles tendons and hamstring muscles and fasciotomy of iliotibial bands. At times, rerouting of tibialis posterior to the dorsal surface of the scond or third cuneiform to balance the foot thereby preventing severe varus position of foot.

AFO

Push Knee Splints

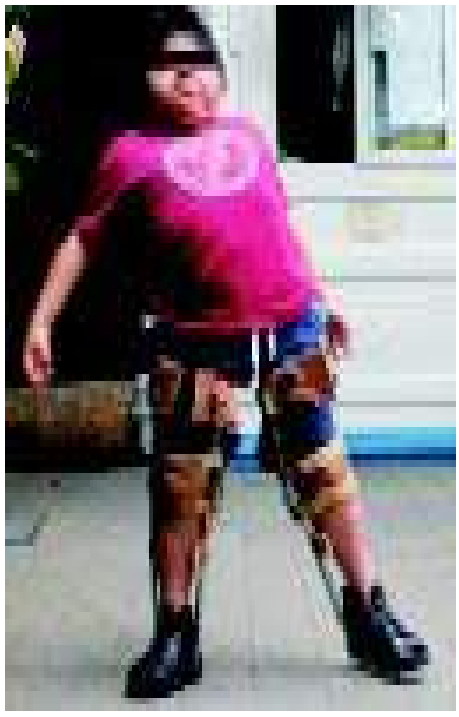
Knee-ankle-foot Orthosis

KAFO

KAFO with T-Strap for preventing inversion

Body Jacket

Standing with KAFO independently inside the Parallel Bar



Walking with KAFO independently outdoors without any aid

Scoliosis before correction with spinal brace

Scoliosis corrected with brace

It is seen that early surgery for contractures followed by intensive physical therapy can prolong brace free ambulation. Gait training can begin within 48 hours after surgery thereby regaining the sense of standing.

Respiratory Management:

As there is involvement of respiratory muscles, proper respiratory management has.to be administrated.

It is required :

1. To maintain thoracic wall mobility.
2. To maintain strength and endurance in respiratory muscles.
3. To establish proper breathing pattern.
4. To make use of non-invasive inspiratory and expiratory aids.

Interventions:

1. **Inspiratory exercises /Segmental** breathing to improve lung expansion and increase chest wall mobility and to increase the strength of diaphragm muscles.
2. **Swimming** is advised to improve endurance and breathing patterns.
3. To teach **efficient coughing.**
4. **Postural drainage** to remove secretions lobewise should be taught.
5. **Inspiratory muscle aids** e.g Nocturnal or daytime IPPV with volume ventilator BiPAP if required.
6. **Expiratory muscle aids** e.g mechanical insufflation - exsufflation.

Spirometer exercises

Nutritional management :

Always, steroids becomes main protocol in treating in Muscular dystrophy. But it produces weight gain by means of fluid retention, increasing appetite and body fat redistribution and increase in body weight becomes a burden on the already weak muscles. Treatment induced excess weight gain can be prevented through a healthy diet regimen having four meals per day and avoid multiple snacks or long fasting periods.

Foods like all vegetables especially leafy and green varieties, raw vegetables salads, Fruits, whole grams and pulses, sprouted grams, dals, lean meat, Chicken without skin, Fish & egg should be included.

Foods like Fast foods, Junk foods, Fry foods (Poori, Vadai),Salty foods, Chocolates & milk sweets, Cookies / cakes, Candies, Cheese, Butter, Ghee, Vanaspathi, Palm oil, Coconut oil, Nuts & oily seeds, Bakery products, Organ meats (Liver, Brain) Shellfish (Prawns, Shrimp) should be avoided.

OCCUPATIONAL THERAPY FOR MUSCULAR DYSTROPHY

Occupational therapists have a unique ongoing role in supporting and working with patients with muscular dystrophy as the patient's needs and the needs of their caretaker are constantly changing. They need to assess and evaluate an individual's physical, psychological and social needs and focus on maximizing skills, promoting and enabling independence, as well as improving the quality of life of the affected individual and his family.

Assessment

The OT will be responsible for occupational performance areas and components. All Dystrophy areas assessed will be considered in relation to functional performance and skills in order to identify realistic and client centered treatment goals. The initial step in management of the child with MD involves taking:

1. Medical History with family concerns
2. Aerobic capacity and endurance assessment

Assessments Of Performance Areas :

I. Activities of Daily Living: Basic ADL (BADL) and Instrumental ADL (IADL) skills

BADL:

- Personal care
- Eating and drinking skills
- Dressing
- Bathing
- Toileting
- Bed Mobility
- Transfer: moving and handling, mobility aids used
- Ambulation and stair climbing

IADL:

- Domestic chores
- Transportation
- Banking
- Shopping

II. Play and Leisure Skills

- Recreation
- Hobbies
- Pets
- Sports
- Peer Group

III. Work and Productivity Skills

- School and Nursery assessments
- Pre Vocational testing
- Vocational / Workplace assessment
- Architectural barriers

Assessment of Performance Components:

The following key performance components need to be assessed.

A. Sensory Processing

Proprioception

B. Neuromusculoskeletal

i) Reflex
ii) Range of motion
iii) Muscle Tone
iv) Muscle strength
v) Endurance
vi) Postural control
vii) Postural alignment
viii) Integumentary (when using orthoses, adaptive equipment, or wheelchair)
ix) Contracture / deformities
x) Atrophy / Wasting

C. Motor

i) Motor control
ii) Gross coordination
ii) Bilateral integration
iv) Fine coordination or dextertity

D. Cognitive Integration

i) Attention span
ii) Spatial operations
iii) Problem solving
iv) Learning
v) Generalization

E. Psychosocial Skills

i) Values
ii) Interests

iii) Self concept

iv) Role performance

v) Social conduct

vi) Interpersonal skills

vii) Self expression

viii) Coping skills

ix) Time management

x) Self control

F. Respiratory Status

G. Sleep

H. Need for Assistive and Adaptive Devices and Technologies

I. Home Assessment

J. Wheel Chair Assessment

Frequency of Assessment

An occupational therapy assessment or review of the young person should be carried out systematically and at least annually. More frequent reviews may be necessary at times of change, such as following periods of ill health or after surgery and following loss of ambulation.

Standardised Assessment Tools

The main goal of assessment in occupational therapy is to get a clear understanding of the individual, their social circumstances and their environment, in order to develop a treatment plan which will improve the quality of life of the person and their family.

The quality of the assessment carried out will have a direct correlation with the quality of the treatment interventions (Turner et al., 2002). Ideally, in the atmosphere of evidence-based practice, standardised assessments should be used to measure the effectiveness of occupational therapy interventions. Occupational therapists and other professionals have created many standardized tests that could be used to assess certain functions that are problematic for people with muscular dystrophy.

1. ADL Scales : FIM, Barthel Index, WeeFIM,
2. Muscle test (gonimeter, pinchometer, dynamometer, hand functions)
3. Range of Motion (R.O.M.)
4. Vignos Functional Rating Scale.
5. Brooke's clinical protocol.
6. Fall Risk Assessment

PLANNING

Following the assessment process, short and long-term occupational therapy goals have to be set with the individual and the family. These goals must be based on the person's preferences.

INTERVENTIONS

STAGE I & II: Early/pre-symptomatic and Early ambulatory (Walking) Stage

At this stage, education regarding the condition and counseling to the patient and family is of utmost importance. One of the primary considerations in the early management program is to retard the development of contractures. Contractures have not been shown to be preventable, but the progression can be slowed with positioning and an ROM program.

A home ROM program should be emphasized and the family instructed in the stretching exercises. Cycling and swimming are excellent activities for overall conditioning and are often preferred over formal exercise programs Standing or walking for a minimum of 2 to 3 hours daily is highly recommended.

Breathing exercises have been shown to slow the loss of vital capacity and forced expiratory flow rate. Game activities such as inflating balloons or using blow-bottles to maintain pulmonary function can easily be included in a home program and will decrease the severity of symptoms during episodes of colds or other pulmonary infections. Night splints are helpful to slow the progression of ankle contractures.

ADLS-Doffing of Upper Body Dressing

Play

Play is essential in the psychological development of children. Occupational

therapists use play activities in treatment to enhance the developmental and functional skills of a child and to increase the child's enjoyment of play and playfulness. Play can also be a valuable communication tool used by children to communicate their feelings and anxieties. All play activities should be based on the child's interests, not their medical condition. Activities requiring repetitive muscle building types of exercise should be avoided, as they are likely to damage muscle tissue further

Sports

Active exercises and participation in sports activities should be encouraged to help delay the development of contractures. Swimming can be good fun at any age and is an enjoyable form of exercise for people with muscular dystrophy. A child in the early stages of muscular dystrophy will enjoy riding and it is a good exercise for helping him to maintain his balance reactions.

Hobbies

Collecting specialised items is a hobby that fosters social interaction. Shopping, as well as having a functional purpose like buying food or clothes, can also be a social experience at the large shopping malls, the use of computers and video games in occupational therapy treatment programmes is beneficial to people with muscular dystrophy. There are a number of interests that can be carried out with limited upperlimb function; these include reading and creative writing, painting, photography, graphic art and some crafts, such as model-making. People can enjoy leisure pursuits

on their own, but Passmore and French. found that social leisure activities were important, as they fostered feelings of self-worth and gave participants a sense of belonging.(12)

Stage III and IV: Late Ambulatory and Early non-ambulatory

Loss of function in personal-care activities is a constant and stark reminder of the progression of muscular dystrophy confronting both the individual and their family in various ways on a daily basis. It is time consuming and physically demanding for all involved personal care is an area that needs to be addressed with the utmost sensitivity.

Forward planning is also vital to ensure that the young person's and their family's changing needs are provided for in a timely manner.

In the first stages of loss of function, small independence aids may be useful in maintaining independent self-care skills. As the condition progresses, these aids become more difficult to use and personal-care tasks a more passive experience for the young person. When considering self-care tasks, it is essential to discuss upper-limb function, as this is crucial for independence in this area.

Eating

For the individual with muscular dystrophy, this basic survival task becomes very demanding as muscle weakness progresses, grip strength becomes poor and it becomes increasingly difficult to lift the hands / arms against gravity.

Possible options include:

- lightweight cutlery and cups or mugs with built-up handles
- rocker knife;
- cuffs with inserts for cutlery;
- Plate with a rim to contain the food when scooping;
- non-slip mats;
- mechanical eating aids
- long straw for drinks

Other alternatives such as elevating the plate height, and angled cutlery will

minimize the amount of active arm, wrist and hand movement required. Mobile arm supports provides support to the forearm to facilitate eating and drinking.

Grooming

Consideration should be paid to the design features of items of equipment for shaving, combing hair such as long-handled brush / comb, and cleaning teeth, including weight and the type of grip. For grooming tasks normally carried out at the basin, access for a wheelchair to fit underneath (i.e. without a vanity unit, or wall-mounted) together with support for the elbows at each side of the sink is necessary.

Bathing

Bath board and shower aids such as hand held

shower, non skid mats, shower chair may be sufficient to provide independence in this area.

Dressing

For postural stability, balance and energy conservation, a seated position with feet firmly on the floor can be helpful for dressing / undressing. As the amount of assistance required increases, small items of equipment such as a dressing stick and reachers may prove useful. Clothing should be comfortable and easy to get on and off: loose, with big head opening and minimal fastenings zips can be made longer to allow easier access for toileting and various sorts of fastenings can be considered, such as Velcro and hooks.

Toileting

Many boys suffer from constipation due to immobility, self-limited diet, reduced fluid food intake to avoid the need to go to the toilet and slowing of peristaltic movement.

A regular toileting routine can help avoid disruption, discomfort and stress, particularly in relation to the school and work environment. Whilst the young person maintains the ability to carry out weight-bearing transfers, rails and a raised toilet seat or a toilet frame may be sufficient. However, as postural control deteriorates, increased support may be necessary in order to allow for a well-supported and relaxed position on the toilet. This type of support generally falls into two categories:

- support which wheels over the toilet;
- frames which fix onto the toilet itself

Transfers

Information and training on how to move and handle an individual can be offered by the occupational therapists, along with advice on equipment that can help when transferring the individual from one position to another. Some of the common moving and handling equipment supplied by therapists are listed below:

- transfer boards;
- hoists and slings;
- sliding sheets;
- handling belts.

Raising the height of beds and chairs from the floor can be useful to the young person in the early stage of the condition as a higher surface requires less muscle power to stand up from, and since the legs can be lowered to the floor in a straight-leg position, rather than trying to rise against gravity from a flexed-knee position. Bed-height adjustment is also helpful if the young person is able to manage to side transfers on / off the bed using a transfer board.

Postural Management

Individuals with muscular dystrophy can develop spinal problems fairly quickly once they stop walking, so they need good postural management interventions to slow down the rate of spinal curvature.Postural management is an approach to the handling, treatment and positioning of children and adults with muscular dystrophy that will reduce the risk of contractures and the development of postural deformities. Passive and active movements of limbs will also slow down the development of contractures.

Good positioning will allow the person to carry out everyday activities with more ease and without adopting abnormal postures. If postural problems are not addressed, it can lead to pain, spinal problems and breathing difficulties. The main pieces of equipment that can help with postural management are:

- sleep systems;
- postural seating;
- wheelchairs with postural seating systems;
- splints / orthotics.

Sleeping

The young person's postural needs must be managed throughout their daily lives which includes overnight positioning. Once pelvic instability is apparent, a postural management plan should be developed to address the sitting, standing and lying positions that the young person will need. This is essential to minimize the risks of deformity, such as the limitations of movement and pain caused by joint contractures or spinal curvatures that impact upon lung capacity and respiratory function.

The young person's postural needs will require regular review and the postural management programme will require to be adjusted accordingly.

Postural Seating

The aims of good seating are: to achieve a good postural position; to maintain functional ability; and

to ensure comfort. Seating which promotes a good sitting posture will also promote effective upper-limb function which is essential for a variety of activities, including feeding, writing and play. It is crucial that seating needs are considered from an early age to prevent or delay deformities and promote optimal function. This should be monitored and reviewed on a regular basis to accommodate any changes as the person's condition progresses.

Wheelchairs

Wheelchairs are essential forms of transport for people with muscular dystrophy; they need them to participate in everyday life when they have difficulty walking. They will need different types of wheelchairs at different stages in their illness. Occupational therapists are involved in the assessment and provision of wheelchairs. They may also have to train the individual in how to use their wheelchair. The therapist will have to give recommendations regarding the postural support and pressure relief required for the chair, as well as the type of controls needed to operate the wheelchair.

Transport Issues

Transport is vital to children and adults with muscular dystrophy. They need

transport to access education, hospitals, and employment and leisure pursuits. The type of transport needed will change over the course of their illness and the methods of transport used will vary to meet their travel needs. Occupational therapists will often be involved in assessments relating to the transport requirements of people with muscular dystrophy. They need to teach the individuals and their caretakers on how to assist the patient onto different forms of transport. They can also suggest car modifications.

Access To Play Equipment

It is important that young patients with muscular dystrophy have the opportunity to play to develop their skills. Occupational therapists can suggest toys and activities that will help with their development.

IT equipment: hardware and software: If an individual cannot use a standard

mouse and keyboard, details of alternative types of keyboards, word-recognition software and joysticks can be supplied for accessing the internet and playing console games and for socializing and other leisure activities.147.

Support groups

Many occupational therapists can provide information about and links to support groups for the individuals with muscular dystrophy, their parents or their siblings. Friends and family are the most important factor to maintaining an active social life. Peer-group friends can provide opportunities for discussion about all topics, including sensitive issues that cannot be easily discussed within the family. Pets with a loving and protective temperament can also give hours of enjoyment and company to people with muscular dystrophy.

Housing, School And Workplace Adaptations

There are many housing adaptations that the therapist can recommend, that will make life easier for the person with muscular dystrophy and their caretakers. A feware listed below:

- ramps;
- bathroom alterations;
- extensions;
- handrails;
- door alterations;
- hoists;
- lifts.

Equipment

Occupational therapists can advise and provide many pieces of equipment that can help the person to maintain their independence in daily living tasks, school tasks or work tasks. Equipment can also help the caretaker with their care tasks. The following are a minute selection of the equipment that could assist a person with muscular dystrophy:

- hoists and slings;
- shower chairs;
- bath lifts;
- eating aids;
- toilet equipment;
- writing aids.

Written Work / Graphic Skills

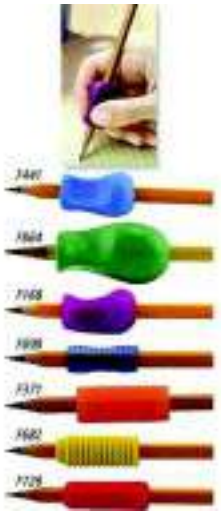
Handwriting is a major occupation of education. Once handwriting is established, it is important to assess the following aspects of it:

- Speed of written work? Does speed reduce with sustained effort?
- Effects of gradual postural changes and deterioration
- Legibility of written work? Does legibility deteriorate with sustained effort?
- Effects of writing - does the child experience fatigue and / or cramps in the hands?
- Child's preference - how does the child feel about using technology? Would they prefer to use a scribe / writer?

Children with Duchenne muscular dystrophy may encounter problems with pencil skills on account of any of the following factors: reduced muscle strength; reduced range of movement; reduced grip strength; reduced stamina; and postural and coordination difficulties. Learning difficulties may also be present and these can further impact on graphic skills. Possible solutions include:

- pencils grips (various types), angled writing boards, resistance provided by

both the writing implement and the paper, paper stands / 'page-ups' may improve performance in early stages;



Various Pencils Grips

- reduction in the amount of writing required, such as by using worksheets on which the child fills in missing words / phrases;
- word-processing technology, including voice-activation programs;
- use of a scribe or writer;
- more oral responses;
- timetabling to allow alternation of passive and active tasks throughout the day to limit fatigue, such as listening activity preceding written work.

Information and computer technology

Word processing: Use of computers should be introduced at an early stage as complimentary to handwriting.

Keyboard alternatives: As power and active movement are lost from the shoulders and upper limbs, it becomes very difficult for the child to extend their arms to the top and edges of the keyboard. Trunk flexion is used to compensate, which is tiring and encourages poor postural positioning. Possible solutions include:

- on-screen keyboard with mouse;
- mouse alternatives, such as touch-pad mouse, joystick, trackball or finger operated integral joystick;
- compact keyboard and or laptops
- Voice-recognition software

Teaching New Methods

Everyone is used to carrying out activities in their own way. An occupational therapist can look at how the individual carries out a task and suggest alternative ways to do it. This may allow the person to complete the task independently. Examples are:

- teaching a person to get dressed on the bed if they have balance problems;
- using a computer to do homework as opposed to having to write it all by hand;
- substitute a battery-operated toothbrush for an ordinary toothbrush.

Pre Vocational Testing

Children need to select subjects that they find motivating and stimulating that could lead to careers that they can pursue. But they also need to be realistic in the courses that they select. Developing a vocational identity is an important part of adolescent development, regardless of their health status. Occupational therapists can have a role by encouraging them to talk about what they want to do when they are older. They can also raise the subject of the boy's expectations regarding employment, as well as establishing what their parents' and their teachers' views are of the boy's

work prospects. This will ensure that everyone has a realistic view.

Workplace Assessments

Occupational therapists can also offer practical help in suggesting adaptations to the workplace and work methods to enable the individual to carry out their job. An employment assessment helps to find out what skills a person with muscular dystrophy can bring to an employer. It may also identify skills that have to be developed to improve the person's employment opportunities. The assessment will also discuss the types of work that the person is interested in obtaining and how their medical condition may influence their choice of work.

Fatigue Management

Energy-conservation methods can be used, to reduce fatigue and pain by planning and pacing activities. Some methods of saving energy are listed below:

- If it is not important to the individual to do the task, can someone else do the work?
- Does the task need to be done every day?
- Spread the tasks over the whole day rather than trying to do everything in one time period.
- Can any tools, equipment or adaptations make the tasks easier?
- Stress reduction and relaxation techniques can also help with fatigue and pain management.

Ongoing Assessments

Once the actions and programmes have been put in place, the occupational therapist needs to make sure that these interventions are fulfilling the original goals set by the individual and their caretakers following the assessment process. If their goals have not been met, the therapist will have to re-evaluate their treatment plan and seek alternative ways for the person with muscular dystrophy to achieve their goals.

Stage V & VI: Late non-ambulatory and Palliative Care Stage

As the condition progresses, the individual find that the aids become more difficult to use and personal-care tasks a more passive experience.

Call Systems

A call system should be put in place which can be easily operated by the young person and alerts the caretaker to their needs.

Diet

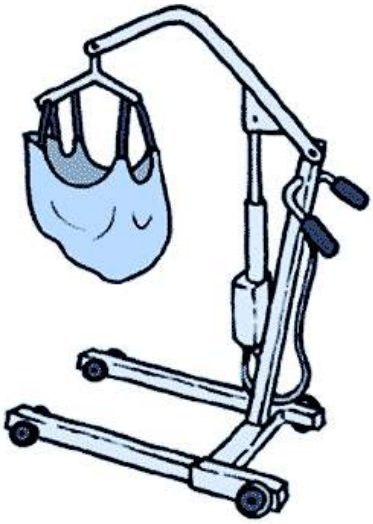
An immobile individual may gain weight very quickly. This is obviously

detrimental to health and increases the physical strains on caretakers. Often, the patient tends to select dry / finger foods in order to avoid the physical difficulties involved in cutting or tearing and reaching the mouth. It is important that the occupational therapist helps the patient make healthy choices.

Transfers: Moving and Handling

Moving and handling needs and the needs of the individual's and caretakers will change over time; therefore, regular reviews need to be carried out. Before any handling task is carried out, it should be explained and consent taken for the move. Postural issues such as trunk and head control have to be assessed to ensure that any equipment or movement approaches used have the right level of support, such as chairs with lateral supports or slings with head supports. The condition of the young person's skin will also influence moving and handling methods. If his skin is vulnerable, make sure that any equipment used will not cause soreness or rubbing. A profiling bed may be useful as part of a postural management positioning programme. Profiling beds also allow the height of the bed from the floor to be adjusted. Caretakers will also find the ability to raise the bed to an optimum-working height invaluable for transfers, helping with dressing, carrying out stretches or helping the young person to move. The risk of back strain is then minimised. Occupational therapists can provide advice regarding the number of transfers required and can also advise on how to eliminate unnecessary moves. Several equipment can be utilized to facilitate transfers under different conditions and requirements. For eg.

- Mobile Shower Chairs, Shower Trolleys And Lifting Bath Seats can be used for bathroom and toilet transfers.
- Hoists and Slings are often used for safe transfer of individuals within their home and also in different locations outside. Depending on the hoist design, slings are made with loops, rings or clips to attach to the hoist. Mesh slings are used for bathing, as they dry quickly. Padded slings should be used where the person's skin is vulnerable.



Lifters

- Stair-climbers and Lifts Stair-climbers and lifts are obviously used to move 151 people and so they can be deemed manual handling equipment. Stair climbers are often operated by carers, who therefore need training in how to use them.

Seating

- There are several aspects involved in the assessment for specialised seating, including seat height, width and depth, arm rests, footplates and head rest.
- As the individual becomes more immobile, pressure relief, possibly in the form of a pressure cushion, becomes increasingly important.
- Tilt-in-space facilities in a chair as well as independently adjustable back rests and footrests facilitate a change in position for an individual who may be unable to achieve this himself.

Sleep Management

As the condition progresses, it may be necessary to provide an increased level of support to manage the young person's lying posture effectively. At this stage, a sleep system is worth considering. The aim of a sleep system is to combine symmetrical positioning with a comfortable and supportive position for sleep. Other sleep systems consist of a mattress overlay that can be moulded, by the positioning of padded supports, to provide contoured all-round body support. For any sleep system, an assessment is required to create an individually customized combination of supports. The following factors would need to be considered: o the quality of sleep that the person gets and how many times a night the person's and caretaker's sleep is disturbed.

- Establish the cause of sleep disturbances. Is it respiratory, dietary, pain-related or psychological?
- Check whether the bed used is a standard or specialist bed.
- Does it meet the needs of the individual and their caretakers?
- Check whether the mattress has pressure-relieving qualities or whether they are using a sleep system to provide positioning support.

Pain Management

There are a number of interventions that occupational therapists can suggest that can help with pain management. This may be the provision of pressure relief equipment, such as the following:

- mattress;
- seating and wheelchair seating;
- pressure cushions for commodes, shower chairs and baths;
- padded and sheepskin slings.

Skin Protection and Management

It is vital to ensure that any equipment issued will not damage the individual's skin.

If the skin is vulnerable, pressure-relieving

materials should be used where the skin comes into contact with the equipment and measures should be taken to limit moving and handling tasks. It is advisable to review how the person is moved and how many times a day he has to be moved, as it may be possible to change the methods of handling to reduce skin contact or to reduce the number of times the person is handled throughout the day. If the individual wears splints, ensure that these are not causing marking or chaffing of the skin. Advice on changing the individual's position when seated in one chair or a bed for long periods of time will also help to prevent skin problems. This can be made easier for the caretaker and the individual by providing adjustable beds and tilt-in-space chairs so that the area that pressure is on can be changed easily with the push of a button.

Sexual Health and Well-Being

Sexuality is fundamental to an individual's health and well-being, irrespective of whether a disability is involved. These needs to be addressed in adults with muscular dystrophy, It is not just about the sexual act. It may be about how medication or incontinence issues affect this aspect of their life. It is also about how they view themselves as a sexual person.

Bereavement and Anticipatory Grief

Individuals with muscular dystrophy experience the loss of muscle strength and associated functions and skills. The loss experienced is ongoing as the condition progresses. This loss is observed but not always understood by the health professional. In addition, in Duchenne's muscular dystrophy as the young man reaches his late teens and early twenties, he becomes acutely aware of his own prognosis. This is compounded by the deterioration and death of his peers. The impact of these deaths and the proximity to the young man himself cannot be underestimated, although it is not always fully recognised. When a realisation or anxiety of impending loss is experienced in advance of the loss, this is anticipatory loss Anticipatory loss can be experienced by people close to the person, too. Bereavement can be understood to be an emotional and psychological event, which

may occur several times in one's life. It affects one's sense of well-being and provokes questions of a spiritual and religious nature, challenging one's existence, sense of meaning and purpose. Bereavement and the associated mourning can also accompany traumatic loss of aspects of oneself, as in paralysis, injury and relationship breakdown.

Thought needs to be given to the fragility of one's confidence, self-esteem and identity when a young person is still growing and developing with a deteriorating condition. It is not always recognised that children grieve, as bereavement is often understood tobelong to adulthood.

Hospices and Palliative Care

A hospice is defined as a programme, or a facility, to provide palliative care and attend to emotional, physical, spiritual and social needs of terminally ill patients and their families, at the hospice or within the home.153. The emphasis is on the relief of pain and promoting quality of life. In this way, it can be seen that hospice care has developed into a concept of care, as it is not limited to the hospice building itself. The term 'palliative rehabilitation' has been developed in recognition that there is an ongoing adaptation and a re-adjustment to living with a deteriorating condition

Caring for the Caregivers

Occupational therapists also have a duty of care to ensure the needs of the parents and caretakers are addressed separately from those of the person with muscular dystrophy. It is necessary to be both aware and sensitive to the possibility of the different experiences and depths of loss when working with children and young people with muscular dystrophy.

The focus at all times for the occupational therapist is on living and enabling

independence but the attitude and approach of the therapist are fundamental to a positive working relationship with the young man and his family. Tact, sensitivity and diplomacy are required by the occupational therapist together with an insight into the difficulties which a family may be experiencing.

PSYCHOLOGICAL REHABILITATION IN MUSCULAR DYSTROPHY

Introduction:

A patient diagnosed with Muscular Dystrophy and his caregivers go through a lot of psychological changes. This starts with having very little knowledge about the disease which leads to confusion, eventually getting to know about not

many ways to cure it, or lack treatment plan, deteriorating condition, family issues, social withdrawal, and embarrassment about the condition and eventually coming in terms about the issue of death. During this process the patient and the caregivers undergo many emotional and psychological changes like confusion, stress, anxiety, restlessness, irritability, etc. Hence, as the patients undergo psychological distress they need psychological help which would enhance their overall well being and would help the patient and their caregivers to cope with the situation in a healthier manner.

Comorbid Disorders:

Autism:388

Research shows an increased risk of Autism in children with Duchenne's muscular dystrophy. Problems with, nonverbal communication skills, language delay, repetitive language, excessive fixation to an object, poor socialization skills and deficits in attention appears to be a common feature of autism in children with Duchenne's. Children suffering from Duchenne's muscular dystrophy who are suspected of suffering from autism or showing symptoms of autism should be assessed by a mental health or behavior professional such as a psychologist, psychiatrist, neurologist, or developmental pediatrician.

It has been seen that as age increases the autistic symptoms in these children reduce in the following areas: In addition, some autistic behaviors in children with Duchenne may improve with age, including: Reciprocal conversation, sharing interest or enjoyment, make a believe play activity, verbal and nonverbal communication.

Obsessive-Compulsive Disorder:

There may be an increased prevalence of obsessive and compulsive like behaviors in patients with Duchenne's muscular dystrophy. In some cases obsessive-compulsive (OC) behaviors may be due to sensory sensitivities or due to deficits in mental flexibility or adaptability. While many Duchenne children may have these tendencies, most would not be severe enough to receive an OCD diagnosis.

Attention-Deficit/Hyperactivity Disorder (ADHD):

Patients with muscular dystrophy are at a higher risk of suffering from attention deficit hyperactivity disorder. Symptoms would signify inattention, hyperactivity or impulsivity, which would differ in their type like predominantly hyperactive-impulsive type, predominantly inattentive type and combined type. These patients suffer from muscle weakness and physical limitations, the symptom of hyperactivity may be less likely in patients with muscular dystrophy.

Cognitive Deficits:

Role of dystrophin: While scientists are still figuring out the exact role of dystrophin in the brain, as some research suggests that not having dystrophin seems to cause an increased risk for specific weakness and learning difficulties, this does not suggest that all patients with muscular dystrophy will have deficits in these areas.

Developmental Delay and Intellectual Ability:

The most common delays are in gross motor skills like sitting, walking, etc. However, patients with Duchenne's muscular dystrophy are at an increased risk for delays in speech development, fine motor skills, etc. Patients suffering from Duchenne's muscular dystrophy are at an increased risk for having an intelligence quotient that ranges from below average to mental retardation (i.e. IQ below 70).

Attention Span and Memory:

The amount of information that a patient with muscular dystrophy is able to grasp in one time (short term memory) may be less as compared to other children. It is usually seen that Duchenne's muscular dystrophy patients with memory cognitive deficits or who have good IQ score would have difficulty in verbal information and visual information. These patients have slow processing speed and also have difficulty multi-tasking.

Executive Functioning:

They have difficulty in planning, organizing, goal oriented behaviours and self-analysis. Mental flexibility in particular appears to be more problematic in Duchenne's than in the general population because of which these patients have difficulty in adapting to expected changes or transitioning from one activity to another.

Learning Disabilities:

Research suggests that children with Duchenne are at increased risk for all three types of specific learning disabilities: dyslexia (reading disorder), dyscalculia (mathematics disorder), and dysgraphia (disorder of written communication) (13)

Emotional Impact in Muscular Dystrophy:

The patients go through a lot of emotional changes and for which they need emotional support, understanding, love and patience to ensure their emotional well being. There may be times when it's difficult to keep up with the progression of the disease, anger, frustration, embarrassment, sadness or anxiety. These emotional issues may occur especially during the developmental period or as the disease progresses (14).

Social Isolation:

As the level of mobility decreases, there is a sense of loss of independence and this could lead to social isolation and depression. Patients with late onset in muscular dystrophy may prefer living in isolation if they don't have enough family support to keep them socially isolated. These patients have a low self-esteem and concept which hinders them from socializing.

Depression and Anxiety:

According to studies it has been found that as compared to normal people, people suffering from Muscular Dystrophy stand higher chances from suffering from depression and anxiety.

Behavioural Issues:

Behaviour problems are quiet common in children who are diagnosed as Duchenne's muscular dystrophy. Young boys with Duchenne's have more difficulty with impulsivity and emotional control than other children of the same age. Most of them are likely to be inflexible in their thinking which may result into non-compliance or oppositional.

Aggression: Patients with muscular dystrophy are at a higher risk of having significant problems with following directions, temper tantrums, problems with arguing and refusing to do what they are asked to do. Behaviour Therapist with the help of Behaviour modification therapy would be helpful in developing alternative strategies to modify the likely triggers to negative behaviours.

Family Issues:

When the patient's starts showing signs of muscular dystrophy initially the parents overlook the disease but as the disease progresses and after the diagnosis is confirmed, the parents go through an emotional turmoil. As, the patients and family members undergo, emotional stress, frustration, anxiety, depression, etc. they should be given the opportunity to discuss the impending death in an accepting environment with the psychologist who is experienced in handling patients with muscular dystrophy (15)

PSYCHOLOGICAL ASSESSMENT:

The most crucial time to consider assessment includes the time around diagnosis as this is the window period of adjustment after diagnosis for the patients and the family members.

Behavioural, Emotional Adjustment or Coping: Patients with muscular dystrophy should undergo a brief screening of emotional status either in every 6 months or annually, as many of the patients slip into depression and this may worsen their physical condition. Standardised tests like Beck's depression inventory, Hamilton's anxiety rating scale, etc. can be used to evaluate their emotional state.

Neurocognitive:

Comprehensive developmental or neuropsychological assessments is recommended at near the time of diagnosis as this would help parents either to go in for a normal school or special school to cater to the special needs of the child. Standardised performance and verbal based tests or rating scales are used to evaluate the IQ or the cognition of the patient like Wechsler's Intelligence Scale for Children, Wechsler's Adult Memory Scale, etc. (16)

PSYCHOPHARMACOLOGICAL INTERVENTIONS:

This should be considered for the treatment of moderate to severe psychiatric symptoms as a part of multimodal treatment plan that includes appropriate psychotherapies and educational interventions.

Intervention for Co- morbid Disorders:

Autism: A patient who is suffering from both Muscular dystrophy and Autism needs to be treated for both but keeping in mind which disorder or disease is causing maximum dysfunction needs to be tackled at first. There are several interventions specifically to reduce autistic features like pharmacotherapy for under the guidance of a psychiatrist, applied behaviour analysis which is conducted by a psychologist, cognitive therapy, floortime, occupational therapy, animal therapy, group therapy and TEACCH. Other interventions are usually problem-focused, and designed to address specific problem areas.

Obsessive Compulsive Disorder:

After listing the obsessions and compulsions of a patient the patient usually undergoes various psychotherapies, of which cognitive-behaviour therapy has been very effective in treating OCD patients, along with it other forms of therapy like systematic desensitization and pharmacotherapy is administered if nonpharmacological treatment is not successful. Few strategies for the caregivers of OCD with comorbid muscular dystrophy patients are:

1. A calm, supportive atmosphere is important to overcoming OC behaviours.
2. If your child is struggling with significant OC behaviours at school, provide the school with a written summary of your child's OCD challenges and needs.
3. School accommodations need to be individualized based on a student's unique pattern of OCD behaviours and coping skills.

Attention Deficit Hyperactivity Disorder:

Treatment and intervention often has three components:

- The use of medication is the most effective component for many boys. Approximately 80 to 90% of children with ADHD obtain some benefit from medication, although this is not always complete improvement. Stimulants are the most commonly prescribed medications. Most boys with Duchenne respond well to treatment with stimulant medication. However, stimulant medications should be used with extreme care in any child with heart problems, which are common in older boys with Duchenne. Thus, the doctor should closely monitor the cardiac status of boys with Duchenne who are prescribed stimulant medications.
- Another component of ADHD intervention consists of psychosocial strategies. Behavioural therapy is very effective in changing the core features of hyperactivity, impulsivity, and inattention. Behavioural therapies are usually the most effective before the child is 10 or 11 years old, so early intervention is the key.
- The third component of ADHD intervention usually involves modifying or adapting the child's environment to reduce the impact that ADHD has on their daily activities. This includes things like increasing more structure and oversight during activities, implementing compensatory strategies at school or home, developing supports for memory and organizational weaknesses, and implementing routines that can be followed consistently.

Intervention for Emotional and Behavioural Issues:

- **Psychotherapy:**

  The primary focus of psychotherapy should be the patient, where the patients emotions, maladaptive behaviour, poor socialization should be taken care of with the help of various psychotherapies like cognitive therapy, cognitive behaviour therapy, rational emotive behaviour therapy, reality therapy, play therapy, art therapy, etc should be administered during the psychological session depending on the patients problems.

- **Individual Therapy:**

  This is suggested for internalizing behaviours like depression, anxiety, low self- esteem, adjustment and coping difficulties.

- **Group Therapy:**

  This is recommended as the patients are mostly socially withdrawn and cuddle into a nutshell, as this is quiet common in patients with muscular dystrophy as with their deteriorating condition they become increasing shy, embarrassed so in order to get them out its necessary that they undergo group therapy which would help them resolve their issues about themselves and help them build confidence. Group therapy for children could be of various forms i.e. Art therapy, Play therapy, etc.

- **Family Therapy:**

  This is quiet important as the patient becomes dependent on the family members for all his needs, which leads to added burden to the family members and a change in their lives. The family members should be well equipped to cope with the situation avoid caregiver's burnout and at the same time help support the patient. Early in the child's life, the family should be guided to encourage the child's independence and to discourage

overprotection. It is important to help the child and the family identify realistic goals for independence (17).

- **Psychoeducation for Caregivers:**

  Parental management and training, where the parents and caregivers are guided on the ways to help them and the patient cope with the situation and to avoid parent-child conflict which could aggravate and add on to the existing problems.

- **Applied Behaviour Analysis:**

  This is especially needed if a patient has comorbid, psychological disorders like autism or attention deficit hyperactivity disorder.

- **Pharmacological Intervention:**

  Selective Serotonin re-uptake inhibitors are prescribed for patients with muscular dystrophy who also suffer from depression, anxiety and obsessive compulsive disorders. Mood stabilisers are prescribed for aggression, anger and emotional dysregulation. Stimulants are prescribed for attention-deficit hyperactivity disorder.

Strategies for Caregivers:

Ways to Mange Aggressive and Difficult Behaviour:

- Develop a routine for the patient for the week and stick to it. Explain to the patient the time-table and do incorporate rewards as children are usually noncompliant after exercising everyday.
- Recreational activities need to be also incorporated in the time table as, it is very important to have something to look forward to during the day that motivates and does not let the patient be depressed about his or her condition For example: Painting or playing board games with siblings or watching television.
- Explain to the patient the situation if there are any changes made or if he is taken for some therapy that he denies to undergo, for example if the patient does not want to exercise on a particular day explain to him the situation, how doing so would have repercussions in the long run and set up a reward system if necessary.
- Try to keep calm when a child is misbehaving. Angry parents and teachers tend to escalate the situation. This would worsen the situation as the child would feel neglected and that nobody understands him / her.
- Focus on the positive. Strategies that only focus on punishment do not promote positive behaviours, increase motivation, or change attitudes. Rewarding / praising / encouraging good behaviour is more effective in the long run. Look for opportunities to say "yes" instead of "no." ("Yes, you can have a cookie, after you").

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Ch.10 Motor Neuron Disease

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MND is also known as Charcot's disease, and Lou Gehrig's disease or Amyotrophic lateral Sclerosis. It is characterized by progressive degeneration of anterior horn cells of the spinal cord causing lower motor neuron type of weakness ; the corticospinal tracts causing upper motor neuron symptoms ; and certain motor nuclei of the brain stem leading to bulbar symptoms. It is one of the most devastating types of neurological disorder, with no known cause.

Prevalence :

The prevalence of this disorder is 5 per 1,00,000 and there is a male to female ratio of 2:1[17]

In about 10% of cases, ALS is caused by a genetic defect. In the remaining cases, the cause is unknown.

There are no known risk factors, except for having a family member who has a hereditary form of the disease.

MND manifests in different forms like:

1. Amyotrophic lateral sclerosis (ALS)
2. Progressive muscular atrophy (PMA)
3. Progressive bulbar palsy (PBP)
4. Primary lateral sclerosis (PLS)

Amyotrophic lateral sclerosis (ALS) can be defined as a neurodegenerative disorder characterized by progressive muscular paralysis reflecting degeneration of motor neurons in the primary motor cortex, brainstem and spinal cord. It usually begins at the age of 40 years of age. "Amyotrophy" refers to the atrophy of muscle fibers, which are dennervated as their corresponding anterior horn cells degenerate, leading to weakness of affected muscles and visible fasciculations. "Lateral sclerosis" refers to hardening of the anterior and lateral corticospinal tracts as motor neurons in these areas degenerate and are replaced by gliosis. Thus ALS exhibits mixed presentation of UMN and LMN symptoms which is the diagnostic criteria.

The syndrome of progressive muscular atrophy (PMA) accounts for 5-10% of patients with MND, and indicates a pure lower motor neuron syndrome without accompanying upper motor neuron signs. There is gross wasting and weakness, along with fasciculations in small muscles of all four limbs, leading to foot drop and reduced grip strength. Unlike ALS, this disease progresses very gradually upto 10-15 years. [19]

Primary lateral sclerosis (PLS) is a clinically progressive pure upper motor symptoms that cannot be attributed to any other disease process. Patient may have involvement of both corticospinal and corticobulbar paths. Pathologically there is loss of pyramidal cells in the precentral gyrus and cerebral cortex, with degeneration of corticobulbar and corticospinal tracts.

Progressive Bulbar Palsy is a condition with involvement of motor nuclei in the lower brain stem. There is primary involvement of the bulbar muscles like the jaw muscles, facial muscles, tongue, larynx and pharynx. Hence patient suffers from dysphagia and dysarthria. As muscles of mastication and deglutition are affected the food particles get stuck to one corner of the mouth Weakness of the pharyngeal muscles causes improper pushing of food particles into oesophagus giving rise to choking. Fibrillation of tongue is a common symptom. It is a fast progressive disorder leading to death with in a span of 2 years of beginning of symptoms and mainly due to respiratory failure.

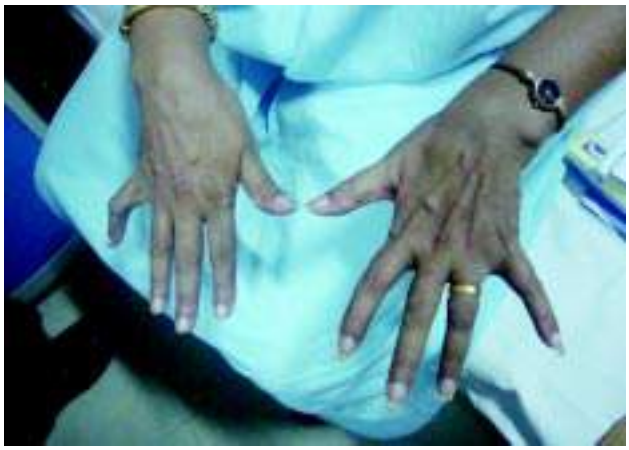
Subtype	Nervous System Pathology
Amyotrophic Lateral Sclerosis	Degeneration of the corticospinal tracts, neurons in the motor cortex & brain stem & anterior horn cells in the spinal cord.
Primary Lateral Sclerosis	Degeneration of upper motor neurons
Progressive Bulbar Palsy	Degeneration of motor neurons of cranial nerves IX to XII
Progressive muscular atrophy	Loss or chromatolysis of motor neurons of the spinal cord & brainstem.

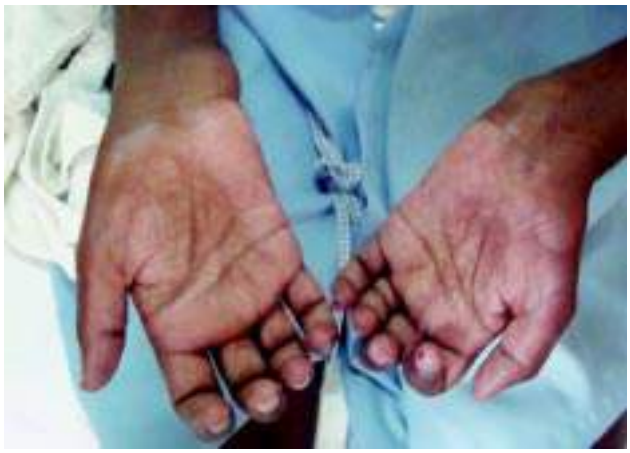
(Reference: Rowland, LP:Diverse forms of Motor neuron disease.Adv Neurol 36:1, 1982.)

Clinical Features:

The features of ALS were first clearly described as a clinico-pathological entity by Jean Martin Charcot in 1869

Approximately two thirds of patients with typical ALS have a spinal form of the disease (classical 'Charcot ALS'). They present with symptoms related to focal muscle weakness where the symptoms may start either distally or proximally in the upper limbs and lower limbs. Patients may have noticed fasciculations (noticed as involuntary muscle twitching) or cramps preceding the onset of weakness or wasting for some months (or years). The weakness is usually of insidious onset, and patients may notice that symptoms are exacerbated by cold weather. Although it is usually asymmetrical at onset, the other limbs develop weakness and wasting sooner or later, and most patients go on to develop bulbar symptoms and eventually respiratory symptoms.



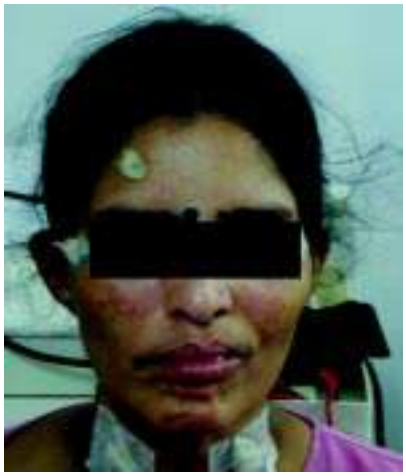


Lower motor neuron symptoms showing weakness and wasting of thenar and hypothenar muscles.

Patients with bulbar onset ALS usually present with dysarthria. Rarely, patients may present with dysphagia for solid or liquids before noticing speech disturbances. Limbs symptoms can develop almost simultaneously with bulbar symptoms and in the vast majority of cases will occur within 1-2 years. Almost all patients with bulbar symptoms develop sialorrhoea (excessive drooling) due to difficulty in swallowing saliva and mild UMN type bilateral facial weakness which affects the lower part of the face. 'Pseudobulbar' symptoms such as emotional lability and excessive yawning are seen in a significant number of cases.

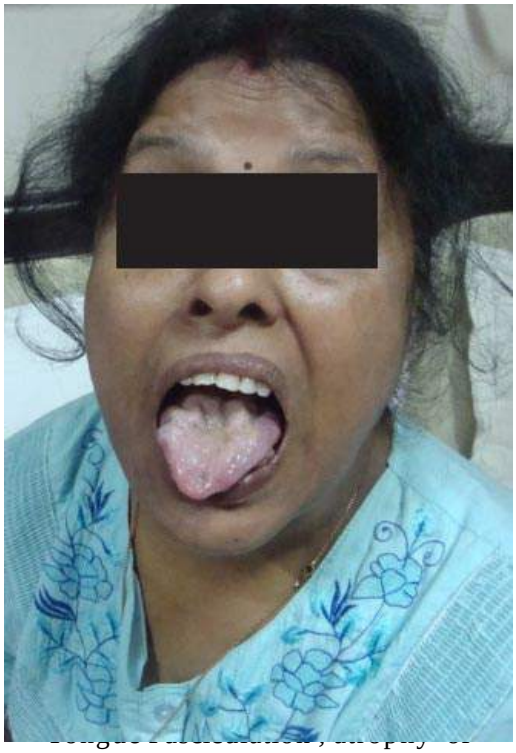
Common Impairments Associated with MND

Type of Impairment / Location	Clinical Manifestation of Pathology
Impairments related to LMN pathology	Muscle weakness, hyporeflexia, hypotonicity, atrophy, muscle cramps, fasciculations.
Impairments related to UMN pathology	Spasticity, Pathological reflexes, hyperreflexia, muscle weakness.
Impairments related to bulbar pathology	Dysphagia,dysarthria,sialorrhea,peudobulbar affect.
Respiratory Impairments	Exertional dyspnea, nocturnal respiratory difficulty, orthopnea,hypoventi lation.
Other Impairments	Dementia, Cognitive Impairments
Rare Impairments	Ocular Palsy, bowel & bladder dysfunction.
Indirect & Composite Impairments	Fatigue, weight loss,cachexia,decreased ROM,pain,gait disturbance,depression,anxiety,deconditioning,balance & postural control impairments



Facial Muscles Weakness

On examining the cranial nerves, the jaw jerk may be brisk, especially in bulbar-onset disease. An upper motor neurone type facial weakness affects the lower half of the face causing difficulty with lip seal and blowing cheeks, but often varying degrees of UMN and LMN facial weakness coexist. The gag reflex is preserved and is often brisk while the soft palate may be weak. Patients develop fasciculations and wasting of the tongue, and tongue movements are slowed due to spasticity. Sensory examination is almost always normal. Respiratory failure and other pulmonary complications are the usual cause of death in ALS. [19]



tongue muscles.

Summary of Revised El Escorial Research Diagnostic Criteria for ALS (Brooks et al., 2000)

The diagnosis of ALS requires:

1. Evidence of LMN degeneration by clinical, electrophysiological or neuropathological examination;
2. Evidence of UMN degeneration by clinical examination, and
3. Progressive spread of symptoms or signs within a region or to other regions, as determined by history or examination,

Together with the absence of:

1. Electrophysiological and pathological evidence of other disease that might explain the signs of LMN and / or UMN degeneration, and
2. Neuroimaging evidence of other disease processes that might explain the observed clinical and electrophysiological signs

UMN signs: Clonus, Babinski sign, absent abdominal skin reflexes, hypertonia, loss of dexterity.

LMN signs: atrophy, weakness. If only fasciculation: search with EMG for active denervation. Regions reflect neuronal pools: bulbar, cervical, thoracic and lumbosacral.

Pathogenesis of motor neuron degeneration in MND

Most ALS cases are sporadic but 5-10% of cases are familial, and of these 20% have a mutation of the SOD1 gene and about 2-5% have mutations of the TARDBP (TDP-43) gene. Two percent of apparently sporadic patients have SOD1 mutations, and TARDBP mutations also occur in sporadic cases. The diagnosis is based on clinical history, examination, electromyography, and exclusion of 'ALS-mimics' (e.g. cervical spondylotic myelopathies, multifocal motor neuropathy, Kennedy's disease) by appropriate investigations. The pathological hallmarks comprise loss of motor neurons with intraneuronal ubiquitin-immunoreactive inclusions in upper motor neurons and TDP-43 immunoreactive inclusions in degenerating lower motor neurons. Signs of upper motor neurone and lower motor neurone damage not explained by any other disease process are suggestive of ALS.

The clinical diagnosis is confirmed by Electromyography (EMG) and muscle biopsy. On EMG, the motor neuron conduction velocity is normal until late in the disease, but the amplitude is reduced. On needling, the mechanical stimulation of the needle causes fibrillation potentials, and spontaneous fibrillation and fasciculation potential are seen when the needle is stationary in relaxed muscle, with greater duration and amplitude of the action potentials. When the muscle contracts there is marked decrease in the number of spikes leading to incomplete interference pattern.

Examination :

1. Joint Integrity, Range of Motion and Muscle length: is examined using standard tools.
2. Pain : seen in ALS is assessed by Visual Analog Scale (VAS)
3. Muscle Performance: is examined by Manual Muscle Testing (MMT), isokinetic muscle strength testing or hand -held dynamometer.
4. Motor Function: Due to Spasticity, and weakness of muscles there could be many manifestations like Impairments in dexterity, incoordination of both gross and fine movements as well as loss of motor control. Therefore Functional assessment of both upper and lower extremities should be done.
5. Tone and Reflexes: Tone can be assessed by Modified Ashworth Scale and reflexes by deep tendon reflexes.
6. Cranial Nerve involvement should be assessed. Pseudo Bulbar and Progressive Bulbar varieties of MND only show involvement of cranial nerves.
7. Postural malalignment and imbalance are seen which can be assessed by Tests like Tinetti Performance Oriented Mobility Assessment (POMA), Berg Balance Scale, Timed Up andGo Test and Functional Reach Test
8. **Gait:** Deviations due to muscle imbalance should be assessed, so also endurance.
9. Respiratory Function: There could be involvement of respiratory muscles resulting into breathlessness, Low vital capacity and lack of cough effectiveness. Therefore Respiratory Function evaluation should be done in detail by using a hand-held spirometer. Aerobic capacity and cardiovascular pulmonary endurance should also be tested to evaluate aerobic conditioning.
10. Because of being in bed for long time without mobility there are chances of getting trophic ulcers: periodic skin inspection should be done.
11. Functional Status: Functional Independence Measure (FIM) can be used to document functional status.
12. Environment Barriers: should be considered for easy accessibility and safety.
13. Fatigue: Fatigue Severity Scale to be used.

Speech Affection in MND :

Motor neuron disorder affects the motor neurons of the brain and the spinal cords. Depending on the type of motor neurons affected, the signs and symptoms vary. The patients where the upper motor neurons are affected present with brisk reflexes / hypertonicity in the jaw, tongue, lips and palatal and pharyngeal areas. The patients where the lower motor neurons are affected present with reduced tonicity, weakness, muscle wasting and fasciculations. Usually, individuals with motor neuron disorder have both upper as well as lower motor neuron affected hence present with mixed symptoms.

Specific Measures for MND :

ALS Functional Rating Scale (ALSFRS): The functional status of ALS patients can be rated by ALS Functional Rating Scale (ALSFRS) and revised version ALSFRS-R It correlates with muscle strength of both upper and lower limbs. ALSFRS-R includes respiratory muscles measures also.

Conventional therapies:

The management of ALS / MND has considerably changed over the past two decades, with an emphasis on coordinated multidisciplinary care between specialist, like physician, speech therapist occupational therapist, physical therapist, dietician and nurses. Advanced directives are on end of life care, respiratory and nutritional management during late stages of life are focused on.

Riluzole is the only approved drug that has been shown to have a modest effect on prolonging life in ALS patients. The mechanism of action of riluzole is not entirely certain but is thought to include interference with N-methyl-D-aspartate (NMDA) receptor mediated responses, stabilisation of the

inactivated state of voltage-dependent sodium channels, inhibition of glutamate release from pre-synaptic terminals, and increasing of extracellular glutamate uptake, thereby preventing degeneration of motor neurons.

Treatments to control symptoms are also helpful:

- Baclofen or diazepam may be used to control spasticity that interferes with daily activities.
- Trihexyphenidyl or amitriptyline may be prescribed for patients with problems in swallowing their own saliva.
- Choking is a common symptom. Patients may decide to have a tube placed into their stomach for feeding. This is called a gastrostomy or Percutaneous Endoscopic Gastrostomy (PEG). A nutritionist's help is very important for them as they tend to lose weight. The illness itself increases the need for food and calories. At the same time, problems with swallowing make it difficult to eat enough.

Role of Physiotherapist in Motor Neuron Disease:

The efficacy of therapeutic interventions is related to:

1. Timing of interventions,
2. Motivation and persistence of patient in carrying out the program.
3. Support from family members.

Rehabilitation intervention plan depends on the following:

1. The rate of progress of the disease
2. Presence of spasticity, bulbar involvement, respiratory involvement causing hypoxia and fatigue.
3. Phase of Disease. Exercises are to prescribed according to level of impairment, functional limitation and level of disability

Phase I : Patient is independent in all ADL, is ambulatory, except for mild weakness and clumsiness in movements.

Stage 1: In case of mild weakness advice is to continue normal activities.

In case of clumsiness, stretching exercises like Yoga

In case of ambulatory patients, gentle resisted exercises with caution not to fatigue or over work.

Stage 1 Patient, exercises to strengthen the knee flexor muscles with resistive theraband exercises.

Stage 1 Patient, exercises like all fours given for weight bearing and strengthening of trunk and limb muscles.

Stage 1 Patient, exercises to strengthen upper limb muscles with weights within fatigue tolerance.

Stage 2: Patient presents with moderate selective weakness in ankles, wrist and hand

Advice : Stretching exercises to avoid contractures.

In case of decreased independence in ADLs like climbing, overhead activities and difficulty in buttoning etc, strengthening exercises to be prescribed avoiding fatigue.

In case of difficulty in Ambulation, Orthotic devices like AFO, hand splints to be considered.

Stage 2 Patient, exercises to strengthen hip extensor muscles (antigravity muscles of lower limb), in all fours position.

Stage 3: Patient presents with fatigability in long distance ambulation, needs supervision in ADL.

Advice : Continue stage 2 and emphasize on deep breathing exercises to be added.

In case of non-ambulatory patients, consider wheelchair, (standard or motorized).

Phase 2: (Partially Independent)

Stage 4: Patient may present with additional pain and edema in hand and feet,

Advice : Consider modalities like massage, elevation and active exercises.

In case of severe weakness in extremities, caution is to be taken to support the joints while doing rotations.

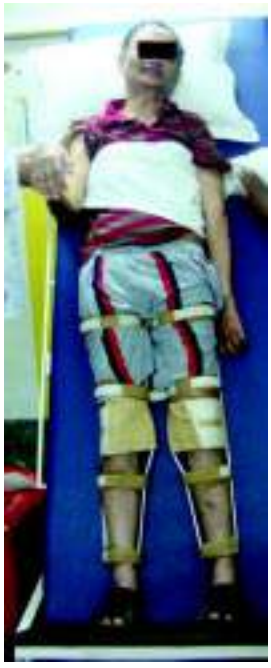
In case of Fatigability in ADLs, encourage isometric upto level of tolerance and to consider slings or arm support, motorized chairs etc.

Stage 5: Patient presents with severe lower extremity weakness,

Advice : Emphasize on teaching family members proper techniques of transfer and positioning of patients limbs.

In case of severe upper extremity weakness, consider modifications at home like high dinning table to facilitate eating etc.

Stage 5 patient with weakness of antigravity lower limb muscles and suspension exercises given within fatigue threshold. Mainly knee musculature suspension strengthening exs.



Stage 5 Patient, treated with Passive movements of limbs and made to stand on standing board for physiological benefits of standing.

Phase 3 : (Dependent)

Stage 6: In case of totally bedridden patients with dysphagia, consider suction, soft diet, tube feeding, PEG feeding etc.

In case of severe breathing difficulty, frequent clearing of airways, tracheotomy and respiratory support if needed.[19]

Neck muscle strengthening exercises.

The research evidence suggests:

1. Overuse weakness does not occur in muscles with MMT grade 3(fair) or greater out of 5(normal).
2. Moderate resistance exercises can increase strength in muscles with a MMT grade3 or greater out of 5.
3. Strength gains are proportional to initial muscle strength.
4. Heavy eccentric exercise should be avoided.
5. Exercises may produce functional benefits.
6. Psychological benefits have yet to be determined.

Patients with severe respiratory and bulbar complications may not benefit from active exercise programs. The goal in end stage is to optimize health and increase quality of life.

- Secondary complications in the form of pressure sore and contractures should be avoided and when the patient goes into respiratory failure, chest physiotherapy becomes necessary. The therapist should make all efforts to maintain the chest expansion of the patient by teaching and supervising breathing exercises in the form of incentive spirometer.
- Proper positioning of patient is necessary because once the limbs become weaker and wasting progresses, it becomes increasingly difficult to maintain upright position for the patient. This difficulty is noted while standing, sitting in wheel chair or on bed, hence proper positioning is necessary. At times patients also develop neck drop. In order to minimize the effect of gravity on the body, it is advised to incline the patient back from vertical position, as in this position the line of gravity passes in front of head and neck through thorax. It also causes relief of pressure of thorax on abdomen, allowing diaphragm to work efficiently and aid in breathing with ease, as their breathing is restricted due to intercostal muscle weakness.

Role of occupational Therapy in Motor Neuron Disease

An occupational therapist (OT) is an integral part of multidisciplinary team that cares for patients with motor neuron disease. They help to maintain mobility, function and independence in long term. As the patient's condition deteriorates, OTs can advise on different ways of performing tasks and the selection, acquisition and adaptation using disability aids.

Calman describes quality of life (QoL) in terms of a match between an individual's hopes and expectations and current reality. For illnesses such as MND, where there is no curative treatment, the role of occupational therapist to emphasize on the maintenance and improvement of Quality of life.[21].

Primary objectives and aims

The primary objective of occupational therapy is to enable individuals to participate in self-care, work and leisure activities that they want or need to perform, thereby optimising personal fulfilment, well-being and quality of life

The tendency toward rapid progression of the disease necessitates regular review and reassessment, and makes planning for the future essential. Liaison with family and care givers is important, to ensure they are aware of the implications of the condition.

Occupational Therapy Intervention

Model of Human occupation- it facilitates ongoing assessment of the impact of the disease throughout human system and the impact it has on occupational performance, patterns and order of behaviour and choices and motivation.

It provides a good position from which to assess the individual's social, physical and temporal context, emphasising the experience of individual and the uniqueness of their life. MOHO describes three subsystems that contribute to our innate drive to act on the environment

Volition: the motivations, values, interests and choices that drive our will to act on and engage with the environment

Habituation: the automatic patterns, habits and roles that shape our occupational behaviour

Mind-brain-body performance: the musculoskeletal, neurological, perceptual and cognitive structures and system required to produce behavioural output.

These three subsystems in this model resonate throughout each other. MOHO provides a reasoned approach to framing the presenting problems in context of the individual's unique experience of them.

This approach considers occupational therapy assessment and intervention designed to enhance and compensate for deteriorating function.

Management for Upper extremity strength of the disease

In Motor neuron disease, after partial or complete denervation of muscle tissue and during inactivity or disuse, muscle strength decreases. When strength is inadequate substitution pattern or trick movements are likely to develop active, active-assisted exercises are used to increase strength.

A muscle must contract at or near its maximal capacity and for enough repetitions and time to increase strength.

Many purposeful activities can be given to maintain/improve strength. For instance, leather lacing can offer slight resistance to the anterior deltoid if lace is pushed in upward direction.. Suspension exercises are advised for all the shoulder muscles.

If the patient is able to hold pegs or blocks, reach outs in sitting position to strengthen abductors and adductors of shoulder. Kneading dough and forming clay objects give resistance and thus strengthening arms and hands.

Neck mobility and Strengthening exercises like - Flexion, extension and rotation of neck can be given for neck support patient can use soft collars and under chin supports.

Management of Spasticity

Active and passive range of motion exercises in the full range are given.

1) **Stretching:** it reduces the spasticity directly in the muscle being stretched by depressing muscle spindle (Kaplan 1962) and it also reduces possibility of contractures.
2) **Bed mobility exercises,** like rolling, shifting, all fours, forward backward shifting in all fours etc (weight bearing exercises) reduce spasticity.
3) **Casting-** casting in inhibitive postures has been shown to be effective in tone reduction.

Joint problems: Muscle weakness can put strain on a joint, causing pain and stiffness. Inactive muscles in the shoulder area may also result in a "stiff shoulder", which causes painful movement. So gentle stretching and hot water fomentation can help to relieve pain.

Postural issues: The loss of normal postural control may cause pain in the muscles of the neck, shoulders, hips or knees. Electrically operated beds and chairs are useful and special cushions and mattresses providing extra padding and support can be helpful. The basic philosophy of seating is same for all patients that "the body should be maintained in balanced, symmetrical and stable posture that is both comfortable and maximizes function (Barnes 1993).

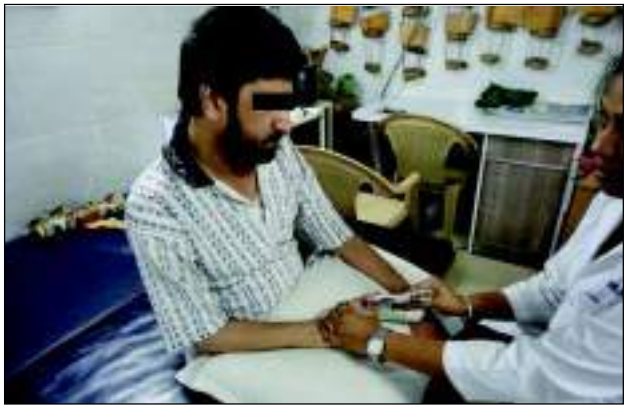
Adaptive devices for Hand functions

Operating locks, door handles and light switches may become difficult and may require special modification. Hands free telephones can alleviate problems with the weight of a telephone receiver. Environmental control systems should be considered earlier rather than later, as they can involve a lengthy wait -- additional elements (TV, lights, door openers, etc.) can be added in at a later date. Book rests, page turners and talking books can prove valuable.[22]

Unilateral weight bearing during peg transfers

Gripper exercise to improve hand muscles

Finger flexor exercise to improve fine motor activity



Therapeutic activity to improve introssei function

Suspension exercises to maintain upper extremity muscles

Modification for self care management

With reduced use of arms and hands, toileting can be a problem, so according to patient conditions Occupational therapist should advise for toilet modification like raised toilet seats. Clothing can also be difficult to manage because of poor hand functions like difficulty in buttoning, zipping or doing other fine motor activities. Velcro can overcome problems with zips and buttons and it will facilitate dressing and independence for toilet purposes. Devices such as dressing sticks and sock aids may be difficult due to their weight. Bathing is another likely problem. The solution needs to take into account the likely progression. If an adaptation to the bathroom is being considered, grab rails, a level access shower will allow for longer term needs whilst an over bath shower is likely to serve only in the short term. Commodes and shower chairs can be useful.

Equipment for Dressing, Grooming and Feeding

Dressing

velcro for assistance with clothing and shoes

long handled aids for washing and hair

Grooming

toothbrush holders and toothpaste

squeezers

wash mitts

Feeding

modified cups, plates and cutlery

non-slip mats

cup holders

collars

splints

Management for Transfer and Mobility

With increased difficulty in transferring, moving and handling equipment may be useful. An adjustable height bed can help reduce risk to carers if it is necessary to provide personal care in bed. Increasing problems with mobility bring increased risk of pressure sores, and regular formal assessment of risk is essential. Special cushions and mattresses may be necessary. When providing hoist slings, seating, shower chairs, mattresses and pressure relief cushions, consideration should be given to pain experienced by the person, often caused by unstable joints, weight loss exposing bony prominences, muscle cramps and spasticity.

Many patients will begin to have difficulty walking. Loss of balance due to foot-drop, muscle atrophy, and spasticity can make walking extremely difficult and dangerous. A typical progression in mobility aids is: ankle-foot orthosis (AFO), then cane, then walker, then manual wheelchair, then power wheelchair. While most patients will understandably resist the use of a mobility aid for as long as possible, it is important that they accept that they are going to get weaker and that mobility equipment can help them maintain independence, conserve energy and most importantly, avoid the perils of a serious fall and related injury. This last point cannot be over emphasized. Catfish Hunter, the famous baseball player who was stricken with ALS, died relatively soon after diagnosis as a result of a head injury sustained as a result of a serious fall. Living with ALS is challenging enough without the added burden and pain of injury.

Ankle Foot Orthosis (AFO)

One common mobility symptom resulting from MND is the inability to hold the toe of one or both feet up while walking. This is commonly referred to as foot-drop and results in the patient having to lift the foot more than normal while walking to avoid tripping. Correcting foot-drop with a lightweight ankle-foot orthosis can be helpful to minimize falls and maintain endurance. Alternatively, low-heeled cowboy boots may be helpful if the patient does not have hip weakness and if the extra weight of the boots is not burdensome.

AFO's are made of lightweight plastic and are available in a variety of types, styles and cost ranges. The simplest and least expensive is the off the shelf, catalog variety. These are a one-piece unit, which come in a variety of sizes. The best and most expensive are custom made devices, which incorporate a spring-loaded hinge at the ankle. Unfortunately, as with most things, you get what you pay for. The catalog variety tends to be less functional and much less comfortable due its one piece, one size fits all design.[23]

Example of walking aids

Canes

There are basically three styles of cane available,

the standard cane, tripod cane and the quad cane. The standard cane has a single tip, the tripod has a triangular (three tip base) while the quad cane has a rectangular four tip base for improved stability. The quad cane is typically considerably heavier and can actually be more awkward making it less stable depending on patient balance and strength. Newer model quad canes are designed using lightweight plastic making them lighter and allowing slight flex which provides some self-leveling. Which style works best for an individual patient will vary depending on condition and can only be adequately determined by having the patient try each style.

Walkers

When a cane does not provide enough support and the risk of falling becomes more frequent, it is time to upgrade to a walker. There many styles and designs of walkers. The standard walker has wheels in front, grippers for hard surfaces or glides for carpets in the back and typically folds up for travel. More elaborate walkers are available with features such as larger wheels, three or four wheel designs, hand brakes, baskets for carrying items and fold down seats. Most tend to be larger and heavier than the standard walker but typically fold up for transport.

Manual Wheelchairs

If walking becomes more difficult and exhausting, it is usually a good idea to obtain a manual wheelchair to use for longer outings. This wheelchair can also be utilized long term as a backup to a power wheelchair. Manual wheelchairs, like power wheelchairs, are optimally custom fitted for the individual patient. Important features include lightweight folding construction, removable wheels, removable flip up footrests, removable or flip up armrests and a comfortable seat.

Power Wheelchairs

As the disease progresses many MND patients will eventually need a power wheelchair. There are a large variety of power wheelchair designs and options available with costs varied range. While a low cost wheelchair will probably suffice early on it will quickly become inadequate as the disease progresses.

Equipments for Mobility

- walking sticks
- walking frame
- rails / ramps
- splints
- wheeled trolley
- wheelchairs - carer or patient operated,
- manual / electric

Management for fatigue

Fatigue is common in MND. By recognising the factors that worsen symptoms and by learning how to conserve energy, people with MND can improve their quality of life. Some strategies are: plan activities in advance, take regular rest periods, rest between activities and before going out, do not exercise to the point of excessive fatigue, cramps, or muscular weakness. (refer to MS chapter for some energy conservation techniques)

Home Modification

1-Use of grab bars to prevent falls and further injury.

2- If the patient is wheelchair bound, there should a ramp will increase independence in indoor mobility. Home should be modified according to patient's needs like adjustment of furniture, increasing door width, removal of architectural barriers etc.

Kitchen tasks are likely to be problematic early in the course of the disease, with difficulties in lifting, carrying, opening jars and reaching higher shelves. Equipment such as vegetable draining baskets, jar openers, tin openers and trolleys may help, as may a rearrangement of the kitchen surfaces to facilitate sliding objects from one area to another instead of lifting. Upper limb difficulties may also necessitate use of modified cutlery, cups and plates, non-slip mats, cup holders and arm supports.[24]

Use of Adaptive Devices in MND

Adaptive devices promotes greater independence by enabling people to perform tasks that they were formerly unable to accomplish, or have great difficulty accomplishing, by providing enhancements to, or changing methods of interacting with, the technology needed to accomplish such tasks.

Accepting any adaptive equipment can be difficult for the person, as the rapidity of the progression of the disease allows little time for adjustment. The

psychological impact of having to rely on a hoist can be devastating, so the issue needs to be approached carefully, and with a positive emphasis on maintaining independence.

Adaptive devices for Communication

Communication problems can include writing, using phones and speaking. Some people need to use speech synthesisers or other communication equipment.. Dysphagia is another difficulty to be aware of, due to damage to some of the cranial nerves (hypoglossal, accessory, vagus, trigeminal, facial, glossopharyngeal).

Augmentative and alternative communication (AAC)

Impairments or restrictions on the production or comprehension of spoken or written language.[10] AAC systems are extremely diverse and depend on the capabilities of the user. They may be as basic as pictures on a board that they are used to request food, drink, or other care; or they can be advanced speech generating devices, based on speech synthesis, that are capable of storing hundreds of phrases and words.[11]

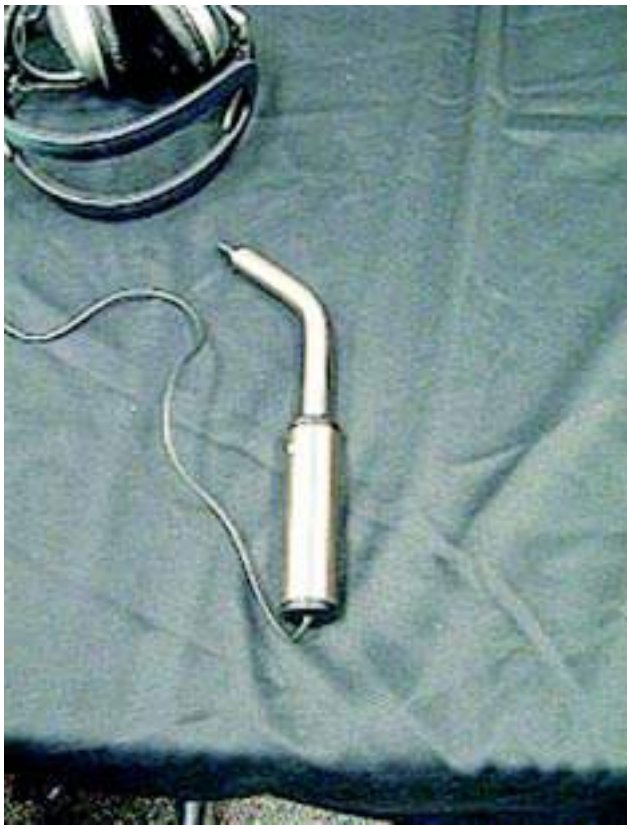
AAC interventions are highly individualized, taking into account specific abilities of language comprehension, social-relational characteristics, learning strengths and weaknesses, and developmental patterns for specific types of intellectual disabilities.[14] AAC can be used to aid both spoken and written language, and can supplement or replace speech and writing as necessary. AAC can be a permanent addition to a person's communication or a temporary aid.[10] The systems used in AAC include gestures, hand signals, photographs, pictures, line drawings, words and letters,[25] which can be used alone or in combination to communicate.[26]

Adult AAC users generally have satisfying relationships with family and friends and engage in pleasurable and interesting life activities.

Accessible input devices for computers

This is a sip-and-puff device which allows a person with substantial disability to make selections and navigate computerized interfaces by controlling inhalations and exhalations.

QWERTY Key board is the most common modern-day keyboard layout.



Ergonomic accessories like, height-adjustable furniture, footrests, wrist rests, and arm supports to ensure correct posture. Key guards fit over the keyboard to help prevent unintentional key presses. Alternatively, Assistive Technology may attempt to improve the ergonomics of the devices themselves:

- Ergonomic keyboards reduce the discomfort and strain of typing.
- Chorded keyboards have a handful of keys (one per digit per hand) to type by 'chords' which produce different letters and keys.
- Expanded keyboards with larger, more widely spaced keys.
- Compact and miniature keyboards.
- Dvorak and other alternative layouts may offer more ergonomic layouts of the keys. There are also variants of Dvorak in which the most common keys are located at either the left or right side of the keyboard.

Input devices may be modified to make them easier to see and understand:

- Keyboards with lowercase keys
- Keyboards with big keys.
- Keyboards with less and big keys, or multifunctional keys, such us the special keyboard Pi- Tech, with only five big rounded keys, which is used with a special software for writing[5]
- Large print keyboard with high contrast colors (such as white on black, black on white, and black on ivory).
- Large print adhesive keyboard stickers in high contrast colors (such as white on black, black on white, and black on yellow).
- Embossed locator dots help find the 'home' keys, F and J, on the keyboard.
- Scroll wheels on mice remove the need to locate the scrolling interface on the computer screen.
- Footmouse - Foot-operated mouse.

More ambitiously, and quite crucially when keyboard or mouse prove unusable, AT can also replace the keyboard and mouse with alternative devices such as the LOMAK keyboard, trackballs, joysticks, graphics tablets, touchpads, touch screens, foot mice, a microphone withspeech recognition software, sip-and-puff input, switch access, and vision-based input devices, such as eye trackers which allow the user to control the mouse with their eyes.

Accessibility software

In human-computer interaction, computer accessibility (also known as Accessible computing) refers to the accessibility of a computer system to all people, regardless of disability or severity of impairment

Human Computer Interactions for Amyotrophic Lateral Sclerosis Patients

Human Computer Interactions is a communication and device control channels, which are helpful for Amyotrophic lateral sclerosis (ALS) patients.

Human computer interactions (HCIs) will be discussed in three respects; **electrical brain activities, eye movements and hemoglobin level in the blood.** With technological advances, fighting or minimization side effects of the diseases is the main purpose of biomedical research. Gradually when disease progress patients feel difficulty in controlling muscles and consequently have problems in moving the entire body. Some of these patients can only move their eyes. In severe conditions of the progressive motor neuron diseases, patients cannot move their eyes nor can they speak. Establishing an efficient communication channel without overt speaking and hand motions makes the patient's life a bit easier and increases their quality of life. HCIs are a research field which includes interactions such as communication and device / machine control between a user and a computer. The aim of the HCI is to improve performance of the interaction, meaning a minimization of the barrier between the human and the computer. [27]

Man-machine interface (MMI), brain-machine interface (BMI) and BCI can be thought of as applications of HCIs. If communication or control is established directly from the brain, it is called BCI and it is the only method of interaction for the individuals with complete Amyotrophic Lateral Sclerosis.

Research in this field is typically focused on several areas of improvement for HCIs in order to increase its usefulness and effectiveness. These areas are:

i. High performance
   a. Accuracy
   b. Reliability
   c. Fast
   d. Robustness
ii. User friendliness (including user training)
iii. Ease of application
iv. Cost effectiveness.

Equipments for Communication

- hands-free telephone
- call bells
- personal alarms
- computerised communication aids including
- light touch keyboards and voice synthesisers
- eye-gaze boards
- computer programs
- voice ampli?ers

Durable medical equipment (DME)

- Seating products that assist people to sit comfortably and safely (seating systems, cushions, therapeutic seats).
- Standing products to support people with disabilities in the standing position while maintaining / improving their health (standing frame,standing wheelchair, active stander).
- Walking products to aid people with disabilities who are able to walk or stand with assistance (canes, crutches, walkers, gait trainers).
- Advanced technology walking products to aid people with disabilities, who would not at all able to walk or stand (exoskeletons).
- Wheeled mobility products that enable people with reduced mobility to move freely indoors and outdoors (wheelchairs / scooters)
- Vehicles modified with Height adjustable suspension, to allow wheelchair entry to the vehicle

Use of Splints and Orthoses

Certain of the specific disabilities resulting from selective muscle atrophy may be overcome by the use of splints or other appliances. Atrophy of the thenar muscles, which affect fine finger movements and pinch grip, are assisted by a suitable splint which holds the thumb in the abducted position and allows opposition to the index finger. Power of finger flexion is also seriously impaired in the presence of wrist drop, and is aided by application of a static wrist extension splint. Foot drop due to weakness of the long toe extensor muscles may be aided by a suitable splint. In mildly affected cases a lightweight slipper type of appliance, extending up the calf of the leg and fitting inside the shoe, may be adequate. Lightweight boots are preferable to shoes because of the added ankle stability. Later, a more sturdy external appliance with toe-spring attached t o the shoe or boot proves necessary, often with an external below-knee caliper. Weakness of the quadriceps muscles causing sudden instability of the knees in walking may be aided by a long-leg caliper with knee-locking device. A cervical collar, Sometimes with suitable chest plate support, is frequently necessary to compensate for paravertebral muscle paresis and flopping of the head onto the chest. Lower limb appliances may be used in conjunction with a walking stick or crutches, depending on the degree of disability. Eventually a wheelchair proves necessary, especially for outdoor excursions. A suitable folding chair which is easily stowed in a car is preferable for outdoor use. When severe weakness affects both upper limbs, independence within the home may be maintained by the provision of a battery-powered wheelchair with suitable microswitch controls. The latter may also advise about the provision of Velcro fastenings rather than buttons or buckles. Similarly, alterations to clothing, with application of Velcro fastenings in place of buttons or hooks, will facilitate dressing and independence for toilet purposes.

Relaxation: Relaxation techniques help to relieve anxiety and have been of great value to many people with MND, especially those with breathing or swallowing problems.

Positive coping

Despite the physical and emotional suffering of MND, there is a large number of patients who cope well and find positive meaning in life.

Role of Psychology in Motor Neuron Disease:

Psychological Aspect:

Motor Neuron Disease is an incurable illness involving the progressive degeneration of upper and lower motor neurons [1]. The physical aspects of motor neuron disease, frequently receives maximum attention whereas, the psychological aspects are most often overlooked or are secondary [2]. As MND is a rapidly deteriorating disease, it requires the individual to continually adjust to new losses and continue making changes. Shorter survival times and higher rates of mortality have been reported for patients with high levels of helplessness and hopelessness. Depression is relatively common i.e. a prevalence rate of about 50% is noticed, as there are other forms of psychological distress in the MND population, which is not associated with illness severity and functional status [3]. Depression strongly correlates with the quality of life.

Mood Disturbances:

It is only natural that adapting to changes caused by MND may lead to changes in mood. Also, as the condition in MND is constantly, there is a need to make constant adjustment or adaptation to new ways of functioning. Many people with MND will experience feelings of frustration, anxiety, stress with changing situations, and guilt. For a small number of people these feelings can be overwhelming and as a result, they become depressed. If someone finds it difficult to engage with a task or concentrate on things, it may be due to feeling low, rather than cognitive change. Some people may choose to take certain medication when they feel like this, e.g. anti-depressants or they may seek counselling.

Depression, Anxiety and Psychological Distress:

Brown and Mueller first studied the psychological state of individuals with MND, describing them as 'stoic' and cheerful, with a capacity to rule out dysphoric affect from consciousness [4]. Studies which have used the psychological tool to assess depression i.e. Beck's Depression Inventory with a sample size of over 100 have reported a prevalence of depression between 11% and 15% [5]. Depression has been correlated to suffering, hopelessness, fast deteriorating and social withdrawal which is in agreement to the quality of life. Although MND is most often associated with persistent physical deterioration, and providing relief for this is very essential, the view has been expressed that a solely physical focus is limited. "Concentration of rehabilitation efforts in this respect leads to a serious underestimation of the significance of assessing and managing the major psychological distress apparent in most patients with ALS."

Emotional Liability:

Some people with MND experience 'emotional liability'. This can result in uncontrollable laughter or crying in response to something that is only moderately funny or sad, for example a television programme. This can seem inappropriate at times and cause confusion.

Social Support and Social Disability:

There is a reciprocal relationship between social support and depression. The stress-buffering model explains that stress, such as that linked with a chronic or terminal illness can be appreciably moderated by good social support. However, the social interaction of many patients with MND is quiet restricted. Also, as the family and spouse are completely involved in taking care of the needs of the patient, right from visiting doctors, to therapy sessions to day - to - day work, patients might lose a lover or companion. Patients might experience increasing isolation from friends, family and socializing, which is what is known as "social disability" [6]. Some patients either withdraw from others as they see their own mortality and are very scared, where as others prefer to live in "social vacuum". Social support is very helpful and acts as a protection from depression which is associated with low social support, withdrawal and feeling of being bounded at home. The loss of or difficulty in communication and mobility are examples of the limitations to socialisation that MND imposes.

Hopelessness and Suicidal Ideation:

Hopelessness is often associated with loneliness and irrational beliefs, a perception of loss of control over life, and loss of 'Purpose in Life' [7]. As, patients with MND deal with deteriorating condition and increasing dependence, this leads to losing hope with every loss of function. Physical function is not a significant predictor of hopelessness; however, depression is a major predictor. As the patient gets to know his/ her diagnosis the dreams and aspirations that he or she has for himself or herself

no longer are significant. The focus is then shifted to visiting doctors, medication, therapies i.e. fighting against deterioration. During these times hopes are an important aspect, as a diagnosis of MND threatens hopes, dreams and expectations on has from life. Patients usually feel helpless about their condition and that they can't do anything to stop the deterioration.

Goggin et al. [8] emphasise the relevance of hopelessness to MND, reporting that hopelessness scores of MND patients exceeded those of HIV/ AIDS patients. A study recently reported that out of 136 MND patients 22% were moderately hopeless and 10% severely hopeless. Another study reported that higher levels of hopelessness were characteristic of MND patients who considered hastened death and that those who were more hopeless also experienced more suffering [9].

Quality of Life (QoL):

Calman [10] describes the quality of life in terms of the match between individual's hopes, expectation and the present reality. As, MND is a disease with no definite cure, the focus of the disease should be on preservation and improvement of the quality of life. Clarke et al. [11] reported that the patients with greater disability are more likely to be affected due to psychosocial aspects of life rather than the physical aspect. Simmons et al. [12] also found no correlation between QoL and physical functioning or strength, but that psychological and existential domains of life were important contributors to QoL. Various studies report that psychosocial aspects of care are important, and that there is more to maintaining QoL than simply attending to a person's physical state.

Cognitive Changes in Motor Neuron Disease:

With regard to cognition, patients suffering from Motor Neuron Disease fall into one of the four groups:

1. No cognitive changes experienced
2. Subtle cognitive and behavioural changes
3. Severe cognitive and/ or behaviour changes where the patient eventually develops frontotemporal dementia (FTD)
4. Patients with motor neuron disease who go on to develop motor impairments i.e. motor neuron disease is diagnosed after dementia.

Frontotemporal Dementia is a type of dementia which involves severe cognitive changes, executive functions, language, and behaviour changes. Approximately 15% of people with motor neuron disease suffer from frontotemporal dementia [13].Approximately 5 percent of patients with motor neuron disease develop frontotemporal dementia. Further, it is reported that about 35% have mild cognitive impairment such as mild aphasia and/or may have behavioural change.

Consequences of Cognitive Changes:

The few effects of cognitive changes seen in patients with motor neuron disease are mentioned below [14]:

- Changes in personality (rigidity or aggressiveness)
- Slowing down of psychological processes like decision-making, answering questions, etc.
- Emotionality (uncontrollable crying, laughter or anger)
- Difficulties with problem solving and generating new ideas and strategies when old ones prove unsuccessful
- Difficulties in divided attention (being unable to do two things at once, like walking and talking)
- Patients have difficulty in concentration i.e. difficulty during reading or dealing with household bills.
- They find it extremely difficult to learn new activities.
- To finish a task through conclusion.
- To start a conversation and to sustain it if there is distraction in the background.
- They find it quiet stressful and difficult to have a sequential activity or plan ahead.
- Patients also have difficulty in responding to social situations.
- Patients have difficulty in comprehend complex sentences.
- Patients have difficulty in finishing a task.

Diagnosis of Cognitive Impairment:

Cognitive and/or behavioural change may occur at the onset of the disease. A full neuropsychological assessment and advice by a clinical neuropsychologist is warranted, where

cognitive impairment has been seen to be very helpful in overcoming the cognitive deteriorations. Paucity of local neuropsychology services may be an issue but this must note prevent assessment and should not add to delays in this vulnerable group of patients. It has been proven that ventilatory failure if develops during the course of the illness can exaggerate cognitive symptoms. Both respiratory muscle weakness and dehydration can be a cause of cognitive change, which should therefore be excluded.

Behavioural Changes:

Behavioural changes occur in the patients as a result of their deteriorating condition, changes in the family structure, work place and constant changes in their functioning, which would escalate their emotional and behavioural problems like:

- Feeling restless and impatient
- Lacking drive or initiative
- Acting impulsively without thinking about the consequences
- Having unhealthy eating habits i.e. eating excessive quantity of sweets.
- Becoming fixated on one activity or routine
- Lacking empathy and appearing indifferent, e.g. when you or someone else close to the person with MND is distressed.

Psychological Intervention:

Positive Coping Strategies:

Motor Neuron Disease leads to physical and emotional suffering, in spite of which many patients cope well and find positive meaning in life. It is important to find purpose of life by using and developing things around us rather than focusing on things which cannot be controlled. This can be done by recreational activities, replacing the negative self-concept to more positive ones and living each day as it comes. There are many studies which report that patients who have a positive effort to confront problems are at a lower risk to experience anxiety and depression. On the other hand, persistent wishful thinking and denial are generally unhelpful coping strategies and associated with higher anxiety and depression even though denial may be adaptive for short periods to allow an individual to deal with a particular aspect of the illness

Hope is the "ability to see a path to the future" and is important to maintaining psychological and physical health. Hope has also been described as the 'driving force' of perseverance and most significantly, the key to the maintenance of quality of life. In the study of Dal Bello-Hass et al. [15] the majority of patients reported a religious practice and there was a positive correlation between religious well-being and QoL.

Cognitive Rehabilitation:

A neuropsychologist, depending on formal assessments i.e. neuropsychological tests, and informal assessment i.e. interviewing the client and observation and a few other medical assessments would suggest strategies to help the patient regain the lost cognitive functions or stop from further losing those functions. For example: work on the orientation of the patient.

Talk Therapy:

This would help the patient take professional help to vent out their anxieties, frustrations, doubts, irrational thoughts and issues related to survival and death. The various therapies used would be cognitive behaviour therapy, cognitive therapy, rational emotive behaviour therapy, gestalt therapy, group therapy, etc.

Caregiver's Support:

Psychoeducating the caregiver and family member, as to how to deal or respond to the patient's emotions, behaviours, mood changes and cognitive changes is extremely important. The caregivers should distribute the work of the patient amongst the family members, which would not burden a particular individual in the family. The caregivers should not neglect their health and overlook their needs of leisure time, work and other needs by being there for the patient at all times. As, this would increase the level of frustration and anxiety within them and which would eventually lead to, not being able to give complete support to the patient.

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Ch.11 Polyneuropathy

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Polyneuropathy describes damage to the peripheral nervous system that transmits information from the brain to the spinal cord and to every other part of the body. Peripheral nerves also send sensory information from the body, back to the brain and spinal cord

The Nervous system can be divided into two parts

- **Central nervous system (CNS) :** The CNS consist of the brain and the nerves that make up the spinal cord, which runs through the vertebrae.
- **Peripheral nervous system (PNS):** The PNS consists of the sensory-somatic nervous system which carries information about sensations and transmits instructions to carry out actions (such as movements). The autonomic nervous system which controls and regulates bodily function (for example, breathing).

Polyneuropathy/peripheral neuritis is the dysfunction of the peripheral nervous system by nerve damage. It can be the result of the underlying disease and also the diseases affecting the nerves. **The main symptoms** are **numbness of feet with tingling sensation, weakness, incoordination, pain, burning sensation and invisible 'glove-like' sensation, abnormal heart rate, reduced sweating and sexual problems.**

Infection, injury, nutritional deficiency, excessive alcohol consumption and disorders such as diabetes can cause peripheral neuropathy. The cause varies according to the specific neuropathy.

Polyneuropathy can be classified into more than 100 forms and produces different set of symptoms and have different prognosis. Motor, sensory and autonomic nerves damage can occur. If the motor nerves are affected then imbalance occurs in the coordination of walking, holding things and other voluntary muscle movements. Damage in sensory nerves results in the loss of sensations like touching or pain whereas autonomic nerves affect the involuntary nerves controlling vital organs.

CLASSIFICATION OF PERIPHERAL NEUROPATHY

Mononeuropathy

When only one nerve is affected then it is called as mononeuropathy. It can be caused by compression to the nerve, carpel tunnel syndrome or some infection and nerve inflammation causing tingling feet.

Mononeuritis multiplex

Mononeuritis multiplex occurs when multiple nerves are damaged in the body due to diabetes mellitus (diabetic neuropathy), Churg-Strauss syndrome, HIV, amyloidosis and rheumatoid arthritis. It is present with dull pain in legs and back mostly at night. Diabetics may have severe pain in thigh of either side with weakness and knee reflex absence.

Polyneuropathy

Polyneuropathy affects nerve cells anywhere in the body irrespective of the nerve path. It can cause changes in axon, neurons cell bodies and myelin sheath surrounding axons. Distal axonopathy is the condition affecting only the axons with intact neurons. In sensory neuronopathy and motor neuron disease sensory and motor neurons are affected respectively. Polyneuropathy produces symptoms such as numb feet, burning, erectile dysfunction and imbalance in bladder function. This neuropathy treatment involves three steps. It starts with removing the cause, then strengthening muscles and their function and in the last pain relief by using neuropathy creams containing capsaicin.

Autonomic Neuropathy

The fourth pattern in the peripheral type of neuropathy is the autonomic neuropathy causing alterations in the autonomic nervous system. It affects the non-involuntary nerves reaching urinary bladder, digestive system, sexual organs and the heart. Chronic diabetic patients are prone to this neuropathy. Autonomic neuropathy can also be present in combination with other neuropathies. It produces symptoms such as incontinence of urine, pain in abdomen with vomiting, diarrhea or constipation, tachycardia, hypotension and impotency.

Etiology

Polyneuropathy may be either acquired or inherited.

Causes of acquired peripheral neuropathy include

physical injury (trauma) to a nerve, tumors, toxins, autoimmune responses, nutritional deficiencies, alcoholism, and vascular and metabolic disorders.

I. Acquired peripheral neuropathies are grouped into three broad categories:

- Those caused by systemic disease,
- Those caused by trauma from external agents, and
- Those caused by infections or autoimmune disorders affecting nerve tissue.

One example of an acquired peripheral neuropathy is trigeminal neuralgia (also known as tic douloureux), in which damage to the trigeminal nerve causes episodic attacks of excruciating, lightning-like pain on one side of the face. In some cases, the cause is an earlier viral infection, pressure on the nerve from a tumor or swollen blood vessel, or, infrequently, multiple sclerosis. In many cases, however, a specific cause cannot be identified. Doctors usually refer to such neuropathies with no known cause as idiopathic neuropathies.

Physical injury (trauma) is the most common cause of injury to a nerve. Injury or sudden trauma, such as from automobile accidents, falls, and sports-related activities, can cause nerves to be partially or completely severed, crushed, compressed, or stretched, sometimes so forcefully that they are partially or completely detached from the spinal cord. Less dramatic traumas also can cause serious nerve damage. Broken or dislocated bones can exert damaging pressure on neighboring nerves, and slipped discs between vertebrae can compress nerve fibers where they emerge from the spinal cord.

Systemic diseases are disorders that affect the entire body and often cause peripheral neuropathy. These disorders may include metabolic and endocrine disorders. Nerve tissues are highly vulnerable to damage from diseases that impair the body's ability to

- Transform nutrients into energy,
- Process waste products, or
- Manufacture the substances that make up living tissue.

Eg. Diabetes mellitus, characterized by chronically high blood glucose levels, is a leading cause of peripheral neuropathy.

Kidney disorders can lead to abnormally high amounts of toxic substances in the blood that can severely damage nerve tissue. A majority of patients who require dialysis because of kidney failure develop polyneuropathy. Some liver diseases also lead to neuropathies as a result of chemical imbalances.

Hormonal imbalances can disturb normal metabolic processes and cause neuropathies. For example, an underproduction of thyroid hormones slows metabolism, leading to fluid retention and swollen tissues that can exert pressure on peripheral nerves. Overproduction of growth hormone can lead to acromegaly, a condition characterized by the abnormal enlargement of many parts of the skeleton, including the joints. Nerves running through these affected joints often become entrapped leading to neuropathy.

Vitamin deficiencies and alcoholism can cause widespread damage to nerve tissue. Vitamins E, B1, B6, B12, and niacin are essential to healthy nerve function. Thiamine deficiency, in particular, is common among people with alcoholism because they often also have poor dietary habits. Thiamine deficiency can cause a painful neuropathy of the extremities. Some researchers believe that excessive alcohol consumption may, in itself, contribute directly to nerve damage, a condition referred to as alcoholic neuropathy.

Vascular damage and blood diseases can decrease oxygen supply to the peripheral nerves and quickly lead to serious damage to or death of nerve tissues, much as a sudden lack of oxygen to the brain can cause a stroke. Diabetes frequently leads to blood vessel constriction. Various forms of vasculitis (blood vessel inflammation) frequently cause vessel walls to harden, thicken, and develop scar tissue, decreasing their diameter and impeding blood flow. This category of nerve damage, in which isolated nerves in different areas are damaged, is called mononeuropathy multiplex or multifocal mononeuropathy.

Connective tissue disorders and chronic inflammation can cause direct and indirect nerve damage. When the multiple layers of protective tissue surrounding nerves become inflamed, the inflammation can spread directly into nerve fibers. Chronic inflammation also leads to the progressive destruction of connective tissue, making nerve fibers more vulnerable to compression injuries and infections. Joints can become inflamed and swollen and entrap nerves, causing pain.

Cancers and benign tumors can infiltrate or exert damaging pressure on nerve fibers. Tumors also can arise directly from nerve tissue cells. Widespread polyneuropathy is often associated with the neurofibromatoses, genetic diseases in which multiple benign tumors grow on nerve tissue. Neuromas, benign masses of overgrown nerve tissue that can develop after any penetrating injury that severs nerve fibers, generate very intense pain signals and sometimes engulf neighboring nerves, leading to further damage and even greater pain. Neuroma formation can be one element of a more widespread neuropathic pain condition called complex regional pain syndrome or reflex sympathetic dystrophy syndrome, which can be caused by traumatic injuries or surgical trauma. Paraneoplastic syndromes, a group of rare degenerative disorders that are triggered by a person's immune system response to a cancerous tumor, also can indirectly cause widespread nerve damage.

Repetitive stress frequently leads to entrapment neuropathies, a special category of compression injury. Cumulative damage can result from repetitive, forceful, awkward activities that require flexing of any group of joints for prolonged periods. The resulting irritation may cause ligaments, tendons, and muscles to become inflamed and swollen, constricting the narrow passageways through which some nerves pass. These injuries become more frequent during pregnancy, probably because weight gain and fluid retention also constrict nerve passageways.

Toxins can also cause peripheral nerve damage. People who are exposed to heavy metals (arsenic, lead, mercury, thallium), industrial drugs, or environmental toxins frequently develop neuropathy. Certain anticancer drugs, anticonvulsants, antiviral agents, and antibiotics have side effects that can include peripheral nerve damage, thus limiting their long-term use.

Infections and autoimmune disorders can cause peripheral neuropathy. Viruses and bacteria that can attack nerve tissues include herpes varicella-zoster (shingles), Epstein-Barr virus, cytomegalovirus, and herpes simplex-members of the large family of human herpes viruses. These viruses severely damage sensory nerves, causing attacks of sharp, lightning-like pain. Postherpetic neuralgia often occurs after an attack of shingles and can be particularly painful.

The human immunodeficiency virus (HIV), which causes AIDS, also causes extensive damage to the central and peripheral nervous systems. The virus can cause several different forms of neuropathy, each strongly associated with a specific stage of active immunodeficiency disease. A rapidly progressive, painful polyneuropathy affecting the feet and hands is often the first clinically apparent sign of HIV infection.

Lyme disease, diphtheria, and leprosy are bacterial diseases characterized by extensive peripheral nerve damage.

Viral and bacterial infections can also cause indirect nerve damage by provoking conditions referred to as autoimmune disorders, in which specialized cells and antibodies of the immune system attack the body's own tissues. These attacks typically cause destruction of the nerve's myelin sheath or axon (the long fiber that extends out from the main nerve cell body).

Some neuropathies are caused by inflammation resulting from immune system activities rather than from direct damage by infectious organisms. These inflammatory neuropathies can exhibit a pattern of alternating remission and relapse. Acute inflammatory demyelinating neuropathy, better known as Guillain-Barré syndrome, can damage motor, sensory, and autonomic nerve fibers. Most people recover from this syndrome although severe cases can be life threatening.

1-GBS (Guillain-Barre syndrome)

Guillain-Barre syndrome (GBS) in developed countries in the post-poliomyelitis era, is the most common cause of acute progressive flaccid paralysis. Guillain-Barre syndrome typically, manifests as rapidly progressing limb weakness, accompanied by paresthesias and often cranial nerve dysfunction. The age of onset is 40 years.

Pathogenesis and Pathophysiology

All subdivisions of GBS appear to be autoimmune diseases resulting from aberrant immune response against various components of peripheral nerve fibers. With AIDP (acute inflammatory demyelinating polyradiculoneuropathy), both humoral and cell-mediated factors appear to be operative, with both antiganglioside antibodies and activated T cells functioning in an attack on myelin sheaths and possibly Schwann cell membranes by sensitized macrophages. Pathophysiologically,

these focal areas of demyelination result in conduction slowing or conduction block, depending on the severity of the process

Epideiology and risk factors:

AIDP is a relatively rare disorder, having an average yearly incidence rate of 1-2 cases per 100,000 population. It occurs worldwide, affects all races, and attacks more males than females. Persons of any age are at risk, although the incidence is higher in the elderly. AIDP is responsible more than 90% of the GBS cases that occur in North America, Europe and Australia. It occurs principally in epidemics and during the summer months, has a very strong association with preceding Campylobacter jejuni infections, and typically affects the children and young adults. AMAN(Acute motor-axonal neuropathy) also occurs in North America, Japan, India and Central America.

Clinical features and associated disorders

The cardinal features of GBS are weakness, paresthesias, and diminished or absent DTRs.

The initial neurological symptoms vary from patient to patient. **Distal, usually symmetrical, paresthesias** involving the toes, finger, or both herald the onset of the disorder in at least 50% of patients. As the disease progress, these paresthesias typically spread proximally but seldom extended beyond ankles and wrists. Facial paresthesias, which are usually perioral, are less common. Gradually weakness in lower extremities especially in proximal muscles, so that patients may feel difficulty in climbing stairs and rising from chairs. Weakness soon spreads to the upper extremities. However, some times the illness progresses, leading to complete paralysis of arms and legs. About one quarter of the time, the paralysis continues up the chest causing respiratorydysfunction

Cranial nerve (VII, Facial diaplegia), occasionally, fasciculations are noted. Sometimes deep pain also occur, which is more prominent in shoulder girdle, back and posterior thighs and is notoriously more severe in night.

Substantial sensory abnormalities are found infrequently and contrast with the high incidence of sensory symptoms. Typically, vibration and proprioception, the sensory modalities mediated over large myelinated fibers, are the most severely affected.

Management:

The treatment of GBS has two components: supportive care and specific therapy.

Supportive care remains the corner stone of therapy, because most patients recover function if they advance past the acute phase of the illness. The major reductions in the deaths from GBS over the past few decades have been due to advances in supportive care, particularly in mechanical ventilation. Respiratory compromise is the most common serious complication and underlines the reason that approximately 30% of GBS patients who are hospitalized require treatment in an ICU. Weakness of the respiratory muscles, particularly the diaphragm, is most often responsible for respiratory failure. Other factors include weakness of various bulbar-innervated muscles and pulmonary complications, including atelectasis, pneumonia, and pulmonary embolism. Vital capacity measurements provide the most useful information regarding the degree of respiratory compromise. Respiratory function may be monitored closely. Intratracheal intubation is complicated in patients with GBS because of dysautonomia, which can result in marked hypotension, arrhythmias, and sudden death due to various drugs and to airway manipulation and because of hyperkalemia, induced by succinylcholine, which can cause arrhythmias and cardiac arrest. Dysautonomia occurs to some extent in majority of GBS patients but more frequently and more severely in those with respiratory failure and severe motor deficits. Pulmonary embolism may occur in about 5% of patients immobilized with GBS, typically after the second week of immobilization. Preventive measures include subcutaneous heparin-switched to warfarin if patients are long term-and intermittent calf pneumatic compression devices. Plain support stockings can reduce the incidence of thrombosis significantly.

Two non life threatening complications of GBS that require attention are pain and psychological trauma. Simple analgesics and nonsteroidal anti-inflammatory medications frequently prove inadequate. Gabapentin or carbamazepine often is beneficial during the acute stage. Narcotic analgesics are required in many patients. However, they must be used with caution because they tend to exacerbate respiratory failure and to produce ileus. The specific treatment for GBS since 1978 has been plasmapheresis. With plasmapheresis, less

mechanical ventilation is required, and both ICU and hospitalization time is decreased. The drawbacks of plasmapheresis include its expense and rare complications such as hypotension and sepsis.

Prognosis:

Overall, approximately 5% of patients die; mortality rates of 15% to 20% have ocurred in ICUs, most often from adult respiratory distress syndrome. Approximately 75% of patients recover without serious neurological residuals, and recovery typically ensues over the 6 to 12 months after onset and is usually maximal by 18 months after onset. Some patients may be left with persistent minor weakness, paresthesias, or sensory loss. Approximately 7% to 15% of patients have permanent, substantial neurological sequelae (e.g., bilateral footdrop, intrinsic hand muscle weakness and wasting, sensory ataxia, burning dysesthesias). Only a very few, however, are permanently bedridden by the disorder

Differential Diagnosis: Transverse myelitits and Vasculitic neuropathies may superficially resemble GBS,. Acute intermittent porphyria can produce a PNS disorder that can be mistaken for GBS.

2-Chronic inflammatory demyelinating polyneuropathy (CIDP), generally less dangerous, usually damages sensory and motor nerves, leaving autonomic nerves intact.

This slowly progressive steroid-dependent polyneuropathy can occur at any age (mean age of onset in the 5th decade).

Pathogenesis and Pathophysiology : CIDP is an apparent immune-mediated disorder of PNS. The term chronic inflammatory demyelinating polyradiculopathy was coined to emphasize that the disorder is a chronic process that results in demyelination as well as inflammatory cell response in peripheral nerves and spinal nerve roots; typically, there is a mononuclear cell infiltration involving the endoneurium and epineurium of peripheral nerve fibers. The predominant physiological feature is Segmental demylination, although usually there is some degree of axon loss as well. These pathological changes have been found to involve roots, plexuses, proximal nerve trunks, as well as some autonomic nerves.

The evidence that CIDP is immune-mediated is compelling, although still somewhat inconclusive. In many respects, it is similar to the evidence advanced for AIDP being an immune- mediated disorder. The onset or relapses of CIDP on occasion seem to be triggered by a preceding event, such as an infection or vaccination that could initiate an immune-mediated response. In addition, most patients with CIDP response to corticosteroids and other immunosuppressants, plasma exchange, and IVIG, therapies designed to treat immune-mediated disorders.

Epideiology and risk factors: The age of onset of CIDP has been reported to range from 1 year through the eight decade or later, with mean in the fourth decade. With the mean age of onset being in the fourth decade, women of childbearing age may develop CIDP. Furthermore, there is evidence that the disorder may relapse or worsen during pregnancy and in the postpartum period.

Clinical Features and Associate Disorders

CIDP is a chronic disorder that typically evolves slowly, almost always requiring a period of 8 weeks or longer for symptoms to reach their peak, and they typically do so over many months. CIDP can follow either a slowly progressive monophasic course, which occurs in approximately two thirds of patients, or a cyclical relapsing course seen in the remainder.

The symptoms of CIDP can be variable, but usually weakness tends to predominate over sensory symptoms. The distribution of weakness often conforms to a typical peripheral neuropathy pattern of symmetrical involvement, with the lower extremities being affected more than the upper extremities and distal muscles being affected more than the proximal muscles. However, in many patients, the weakness is asymmetrical, and in others, it is more prominent proximally. The degree of weakness varies markedly from patient to patient. In majority of patients, it is mild to moderate in degree, but rarely, the weakness can be severe and generalized, sometimes associated with respiratory failure necessitating ventilatory support. Muscle cramping and fasciculations are infrequent symptoms.

Sensory symptoms are usually confined to mild paresthesia or modest sensory loss. However, in those patients with chronic progressive courses, sensory symptoms may become more prominent. In addition to numbness and parathesia, pain sometimes is reported, although this occurs in less than 20% of patients. Very infrequently, CIDP may

present with predominately sensory symptoms, including numbness, parathesia, and sensory ataxia. However, in most instances in which the disorder begins as sensory neuropathy, it later evolves to a more typical pattern of sensory and motor involvement. On occasion, the cranial nerves may be affected, causing symptoms of diplopia, facial weakness or numbness, dysarthria and dysphagia. Autonomic symptoms are uncommon, although incontinence and erectile impotence have been reported.

The physical signs noted in CIDP reflect the clinical symptoms. Distal symmetrical weakness, with the lower extremities more severely affected than the upper extremities. However, asymmetrical involvement and more prominent proximal involvement can occur, and they serve as important clinical clues to diagnosis of CIDP. Generalized hyporeflexia or areflexia is characteristic. Sensory loss often conforms to a distal to proximal gradient, in the so-called stocking-glove pattern, with all modalities affected to a nearly equal extent. Occasionally, sensory ataxia may be observed. Cranial nerve findings may include signs of facial, bulbar and neck weakness.

Some patients with CIDP have abnormalities on MRI scans of the head consistent with multiple sclerosis.

Differential Diagnosis, CIDP may be confused with a wide variety of chronic sensorimotor polyneuropathies, HIV infection,Familial hypertrophic polyneuropathies

On occasion, CIDP may be difficult to differentiate with AIDP. Patients with subacute courses of CIDP have been described and sometimes CIDP may begin with a relatively acute onset. Overtime, CIDP usually declares itself by a typical chronic relapsing course or a more slowly progressive monophasic course. The distinction between CIDP and AIDP is significant because the ultimate course, prognosis, and treatment are different.

Prognosis

The prognosis is favorable for most patients with CIDP who are properly diagnosed and treated. In one large group of patients whose disease duration averaged 5.7 years and who were followed for a minimum of 24 months, nearly 40% were asymptomatic and nearly 50% had either minor symptoms that did not have any impact on their level of function or had only very modest restriction in their lifestyle.

3-Chronic idiopathic sensory neuropathy (CISN): This is basically a sensory form of CIDP with limb ataxia, numbness and pain, loss of proprioception, normal muscle strength but generalized areflexia.

4-Alcoholic Polyneuropathy: Alchoholic Neuropathy is a disorder involving decreased nerve functioning caused by damage that results from excessive drinking of alcohol. The onset is often insidious and presents with distal symmetric dominant sensory-dominant polyneuropathy confined to the legs. Painful sensations with or without burning represents the initial and major symptom. Other manifestations are dysesthesias, paresthesias or sensory ataxia. Autonomic features may be present (commonly abnormal sweating or diarrhea, less frequently postural hypotension, vomiting, micturition difficulties, impotence and retrograde ejaculation). Electrophysiological studies show axonal neuropathy predominantly affecting the sensory nerves. Progression is gradual continuing over months or years. Abstinence reveals gradual improvement in the polyneuropathy. Rapidly progressive polyneuropathy resembling GBS but without raised protein or slowed NCVs can occur in alcoholics.

5-Idiopathic peripheral facial palsy (Bell palsy): The incidence of Bell palsy is 2-3‰. Retroauricular pain usually precedes the paralysis by 1 or 2 days. Almost 50% show maximal paralysis in 2 days. Recovery takes weeks - 2 month. Ramsay-Hunt syndrome (zoster around the ear, acute peripheral facial palsy, and symptoms involving the VIII cranial nerve) is caused by a reactivation of VZV(Varicella Zoster Virus). The same virus causes acute peripheral facial palsy without skin lesions (zoster sine herpete). About 15% of patients will have permanent sequelae. The facial palsy associated with Ramsay-Hunt syndrome is more severe and has a lower recovery rate than that of Bell palsy. The diagnosis of Ramsay-Hunt syndrome is relatively easy in the presence of typical skin rash.. A combination of acyclovir (4000 mg daily for 5 days) and prednisone (60 mg / day for 4 days) provide a 100% cure rate if started within 3 days after the onset.

6-Diabetic Neuropathies

i. **Diabetic polyneuropathy** Diabetic neuropathies are a family of nerve disorders caused by diabetes. People with diabetes can, over time, have damage to nerves throughout the body. Neuropathies lead to numbness and sometimes pain and weakness in the hands,

arms, feet, and legs. Problems may also occur in every organ system, including the digestive tract, heart, and sex organs. People with diabetes can develop nerve problems at any time, but the longer a person has diabetes, the greater the risk.

An estimated 50% of those with diabetes have some form of neuropathy, but not all with neuropathy have symptoms. The highest rates of neuropathy are among people who have had the disease for atleast 25 years

ii. **Diabetic small-fiber sensory polyneuropathy:** This is most prevalent in IDDM (Insulin- Dependent Diabetes Mellitus) of more than 20 years duration, and in those with AHT(antihypertensive treatment) or poor glycemic control. The majority of patients present initially with relative symmetric sensory symptoms including paresthesia, burning and lancinating pain in the legs with stocking distribution. Pain and temperature sensation is affected; vibratory sensation and joint position sense are lost in the toes. Positive Romberg sign with sensory gait ataxia may be found. Ankle jerks are lost, while others are preserved. Autonomic features may include warm dry foot with hard vulnerable skin with cracking. Neuropathic joints may develop. Distal muscle weakness (with atrophy) is unusually except in longstanding cases.

iii. **Distal polyneuropathy:** Distal, symmetrical, primarily sensory neuropathy is the commonest form of chronic neuropathy in diabetes. The lifetime incidence is estimated at 37-45% in type 2 diabetes and 54-59% in type 1diabetes. The clinical presentation consists of distressing numbness and paresthesias in the feet and lower legs, worse at night. The pain is often reported as superficial presenting as allodynia, sharp, stabbing, or burning pain in the feet. The ankle and knee jerks are absent. Trophic changes may occur. There is generally a family history of diabetic polyneuropathy and obesity is commonly found. Tendon reflexes and distal sensation are reduced.

iv. **Proximal diabetic radiculopathies:** There are two distinctive forms of radiculopathy in diabetic patients; the painful asymmetric radiculopathy often referred to as "diabetic amyotrophy" and the painless proximal symmetric amyotrophy. The diabetic amyotrophy is most commonly observed in NIDDM (non-insulin dependent diabetes mellitus) patients in their 6th or 7th decade. Pelvic girdle and thigh muscle weakness and atrophy are evident, but mainly affecting the quadriceps muscle and developing over the next few days or weeks. The knee jerks are lost. Despite the unilateral onset, bilateral weakness eventually occurs in 50% of patients. The plantar responses may be extensor! Upper extremities are rarely affected. Deep and superficial sensation may be intact or mildly affected. The neuropathy is associated with profound weight loss. Femoral NCV is delayed. CSF is slightly elevated. Improvement of the condition is usually seen after some months (or years). Only 20% of patients have complete recovery of muscle strength. 1/5th of patients experience recurrence and often affecting the opposite limb.

v. **Diabetic mononeuritis multiplex:** This painful unilateral or asymmetrical multiple neuropathy tends to occur in elderly with mild diabetes or unrecognized diabetes. It tend to occur during periods of transition, when severe hyperglycemia or hypoglycemia arises, following the initiation of insulin therapy or in association with rapid weight loss.

vi. **Diabetic Mononeuropathy:** Painful oculomotor nerve palsies, often sparing the pupil are common in older diabetics and resolve spontaneously.

vii. **Diabetic Truncal neuropathy:** Attacks of truncal pain and sensory loss affecting one or more thoracic roots can occur. It typically occurs in the 5th - 7thdecade in NIDDM and is associated with weight loss. Focal paralysis of the abdominal wall muscles may be seen. EMG shows denervation of paraspinal muscles. Recovery is spontaneous but may take months.

7-Uremic polyneuropathy: 50% of patients with end-stage chronic renal failure have clinical or electrophysiologic evidence of polyneuropathy. Uremic polyneuropathy develops gradually and occurs with glomerular filtration rates below 10 ml / min. In contrast to the underlying causes of renal failure, which are often resulting in focal or demyelinating polyneuropathy, uremic polyneuropathy is axonal in nature. Restless legs (burning paresthesias, itching sensation) are often the initial manifestation, followed by muscle

cramps and fatigability and, finally, muscle distal weakness and atrophy. The earliest objective signs are loss of vibration at the toes and absent ankle jerks. NCVs indicate axonal degeneration of motor and sensory fibers. Renal transplant greatly improves the condition. Occasionally an acute uremic polyneuropathy may develop that resembles GBS. NCVs may show demyelinating features and raised CSF protein.

8-Thyroid/Pituitary Neuropathies

Mucinous deposits in soft tissue resulting in nerve compression and carpal tunnel-like symptoms have been implicated in neuropathy associated with hyperthyroidism. Neuropathy associated with excess growth hormone or acromegaly has been associated with subperineurial-tissue proliferation and diminished myelinated and unmyelinated fibers.

9-AIDS-associated Neuropathy

Polyneuropathy affects as many as one-third of individuals with acquired immunodeficiency syndrome (AIDS), most commonly manifested as distal, symmetrical polyneuropathy.

10-Toxic polyneuropathy

i. **Acrylamide polyneuropathy:** High-dose intoxication which occurs after drinking of contaminated water causes subacute encephalopathy followed some days later by mild polyneuropathy. Chronic low dose intoxication (construction workers) results in polyneuropathy within 4 weeks after exposure. Diffuse areflexia is an early sign. Ataxia may be prominent. The neuropathy involves both sensory and motor. Contact dermatitis, blistering and hyperhydrosis of the palms and the soles may occur. SNAPs are small or absent. Sural nerve biopsy shows degeneration and regeneration of axons. Complete recovery is possible.

ii. **Acute arsenic neuropathy:** This type of poisoning occurs either acute or chronic (smelting workers). In acute poisoning neurologic symptoms appear 2-3 weeks after the initial gastrointestinal manifestations (abdominal burning pain, nausea, vomiting and diarrhea within hours-days of ingestion) or shock. Subacute polyneuropathy (7-14 days) presenting as numbness and often painful paresthesias with loss of vibration and position sense are the initial manifestations, subsequently followed by leg muscle weakness. Ankle jerks are invariably lost. SNAPs are absent and motor conduction is initially mildly slowed. Sural nerve biopsy shows axonal degeneration. Improvement over years occurs but is often incomplete. Other features in acute poisoning are encephalopathy, pancytopenia, eosinophilia, liver and / or kidney failure depending on the amount. Chronic arsenic exposure in industrial workers may produce an asymptomatic sensorimotor neuropathy detectable only by NCVs.

iii. **Thallium polyneuropathy:** High doses of thallium (rodenticide) cause shock due to gastroenteritis and dehydration. When this initial phase is survived sensorimotor neuropathy becomes apparent in a few days. Sensory symptoms occur first and consist of painful paresthesias and allodynia affecting the feet. The reflexes remain preserved. The neuropathy progresses rapidly to involve the respiratory and bulbar muscles resembling GBS. Associated autonomic neuropathy results in tachycardia and hypotension. CNS manifestations are optic neuropathy, ataxia, confusional psychosis and involuntary movements. Systemic features include dark pigmentation at the hair followed by rapid complete alopecia, dry skin, Mees lines on the nails helps to differentiate this form of neuropathy from other forms of acute polyneuropathy. Sural nerve biopsy shows axonal degeneration.

iv. **Lead polyneuropathy:** Workers in metal smelting and battery (fumes) manufacturing are often the subject of chronic inorganic poisoning. The presentation is very similar to porphyria. Abdominal cramps are often the first manifestation. Classically a pure motor neuropathy develops affecting most often the radial or peroneal nerves. Sensory loss is uncommon. Anemia, basophilic stippling of red cell precursors and free erythrocyte protoporphyrin level are the best guide to chronic lead exposure. Motor NCVs may be slowed. Sural nerve biopsy shows loss of the large myelinated axons.

v. **Mercury polyneuropathy:** Chronic exposure to inorganic or elemental mercury produces mild sensorimotor peripheral polyneuropathy. Elemental mercury poisoning resembles

MND. Organic mercury poisoning (e.g. Minamata disease) typically causes the combination of paresthesias, sensory ataxia, and visual field constriction. Paresthesias around the mouth and fingers and toes with sometimes normal NCVs may point towards the diagnosis. Measurement of mercury levels in blood, urine and hair confirm the diagnosis.

II. Inherited forms of peripheral neuropathy

Inherited forms of peripheral neuropathy are caused by inborn mistakes in the genetic code or by new genetic mutations. Some genetic errors lead to mild neuropathies with symptoms that begin in early adulthood and result in little, if any, significant impairment. More severe hereditary neuropathies often appear in infancy or childhood.

The most common inherited neuropathies are a group of disorders collectively referred to as Charcot-Marie-Tooth disease. These neuropathies result from flaws in genes responsible for manufacturing neurons or the myelin sheath. Main symptoms of Charcot-Marie-Tooth disease include extreme weakening and wasting of muscles in the lower legs and feet, gait abnormalities, loss of tendon reflexes, and numbness in the lower limbs.

1-Hereditary Sensorimotor neuropathy (HSMN)/ (Charcot-Marie-Tooth disease)

This is the most common cause of distal leg muscle wasting and weakness ("peroneal muscular atrophy" syndrome), usually accompanied by pes cavus. The age of onset is variable and asymptomatic, yet affected elderly relatives may be identified. In HSMN males are commonly affected, whereas females are more often asymptomatic. Positive sensory symptoms (paresthesias) are unusual and should rather suspect acquired neuropathy. Associated features (spastic paraparesis, optic atrophy, retinitis pigmentosa, deafness and mental retardation) can occur. With the availability of genetic testing, noninvasive accurate diagnosis is now possible and omits the need for nerve biopsies.

i. **HSMN - I (Charcot-Marie-Tooth disease type 1 or adult-onset demyelinating)**: The age of onset is 1st to 2nd decade of life and this is mostly autosomal dominant (autosomal recessive, and X-linked forms exist). Clinically, distal leg muscle atrophy ("inverted champagne bottle"), lost ankle jerks and weakness, usually accompanied by pes cavus characterize the disorder. Some hand weakness, tremor and limb ataxia may ultimately develop. All modalities of sensation may be impaired distally in the limbs and acrodystrophic changes secondary to sensory loss may develop. Scoliosis, pupil abnormalities, or extensor plantar responses occasionally occur. Diaphragmatic weakness may cause dyspnoea or respiratory failure. Palpable nerve thickening (great auricular nerve) is found in 25% of patients.

   Progression of disease is slow.

ii. HSMN - II (Charcot-Marie-Tooth disease type 2 or adult onset axonal form): The age of onset is 2nd to 3rd decade of the life and this is mostly autosomal dominant (X-linked and autosomal recessive forms exist) Males are more commonly affected than females. Common symptoms are distal leg muscle wasting and weakness, with absent ankle jerks. Hand weakness, tremor and ataxia of the arms, sensory loss, generalized areflexia or pes cavus are less common than in HSMN - I. There is no nerve hypertrophy. Nerve biopsy shows axonal loss. Late-onset HSMN should be differentiated from chronic idiopathic axonal polyneuropathy in which sensory features and progression are characteristic.

2-Hereditary neuropathy with liability to pressure palsies (HNPP): This condition also known as tomaculous (sausage-like) neuropathy is autosomal dominant with the onset of symptoms is in the 2nd or 3rd decade of life and translates in a tendency to develop painless focal and recurrent demyelinating sensory and motor peripheral mononeuropathies due to unusual vulnerability to pressure or traction. Exposed nerves such as ulnar nerve, radial nerve and superficial peroneal nerve are especially vulnerable. Painless brachial plexus lesions may result from traction or prolonged abnormal postures. Recovery occurs over days, weeks or months, but permanent disability may develop after recurrent episodes. Typically, patients experience tingling of the fingertips when using scissors.

NCVs show prolonged distal motor latencies or reduced SNAPs with conduction blocks and minor slowing of motor conduction velocities both in affected and asymptomatic gene mutation carriers. Almost 80% of patients have a deletion of PMP22 gene at chromosome 17p11.2.

Nerve biopsy shows sausage-like pattern of the nerve (also found in familial brachial plexus

neuropathy, Ehler-Danlos and paraproteinemic neuropathy).

3-Hereditary sensory and autonomic neuropathy:

Five clinical different entities have been described under hereditary sensory and autonomic neuropathies - all characterized by progressive loss of function that predominantly affects the peripheral sensory nerves. Their incidence has been estimated to be about 1 in 25,000.

i. **(Hereditary Sensory Radicular Neuropathy)**: It is the most common of the hereditary sensory and autonomic neuropathies (HSAN). It is transmitted as autosomal dominant trait and is characterized by a sensory deficit in the distal portion of the lower extremities, chronic perforating ulcerations of the feet and progressive destruction of underlying bones. Symptoms appear in late childhood on early adolescence with trophic ulcers as pain sensation is affected. Many patients have accompanying nerve deafness and atrophy of the peroneal muscles. Histopathologic examination reveals a marked reduction in the number of unmyelinated fibers. Motor nerve conduction velocities are normal, but the sensory nerve action potentials are absent.

ii. **(Congenital Sensory Neuropathy):** It is characterized by onset of symptoms in early infancy or childhood. Upper & lower extremities are affected with chronic ulcerations and multiple injuries to fingers and feet. Pain sensation is affected predominantly and deep tendon reflexes are reduced. Autoamputation of the distal phalanges is common and so is neuropathic joint degeneration. The NCV shows reduced or absent sensory nerve action potentials and nerve biopsy shows total loss of myelinated fibers and reduced numbers of unmyelinated fibers. It is inherited as an autosomal recessive condition.

iii. **(Familial dysautonomia, Riley-Day syndrome):** It is an autosomal recessive disorder seen predominantly in Jews of eastern European descent. Patients present with sensory and autonomic disturbances. Newborns have absent or weak suck reflex, hypotonia and hypothermia. Retarded physical development, poor temperature and motor in coordination are seen in early childhood. Other features include reduced or absent tears, depressed deep tendon reflexes, absent corneal reflex, postural hypotension and relative indifference to pain. Scoliosis is frequent. Intelligence remains normal. Many patients die in infancy and childhood. Histopathology of peripheral nerve shows reduced number of myelinated and non-myelinated axons. The catecholamine endings are absent.

iv. **(Congenital Insensitivity to Pain and Anhidrosis):** It is an autosomal recessive condition and affected infants present with episodes of hyperthermia unrelated to environmental temperature, anhidrosis and insensitivity to pain. Palmar skin is thickened and charcot joints are commonly present. NCV shows motor and sensory nerve action potentials to be normal. The histopathology of peripheral nerve biopsy reveals absent small unmyelinated fibers and mitochondria are abnormally enlarged.

v. **(Hereditary Sensory and Autonomic Neuropathy):** It also manifests with congenital insensitivity to pain & anhidrosis. There is a selective absence of small myelinated fibers differentiating it from type 4

Diagnosis

Diagnosing peripheral neuropathy is often difficult because the symptoms are highly variable. A thorough neurological examination is usually required and involves;

- Taking an extensive patient history (including the patient's symptoms, work environment, social habits, exposure to any toxins, history of alcoholism, risk of HIV or other infectious disease, and family history of neurological disease),
- Performing tests that may identify the cause of the neuropathic disorder, and
- Conducting tests to determine the extent and type of nerve damage.

A general physical examination and related tests may reveal the presence of a systemic disease causing nerve damage. Blood tests can detect diabetes, vitamin deficiencies, liver or kidney dysfunction, other metabolic disorders, and signs of abnormal immune system activity. An examination of cerebrospinal fluid that surrounds the brain and spinal cord can reveal abnormal antibodies associated with neuropathy. More

specialized tests may reveal other blood or cardiovascular diseases, connective tissue disorders, or malignancies. Tests of muscle strength, as well as evidence of cramps or fasciculations indicate motor fiber involvement. Evaluation of a patient's ability to register vibration, light touch, body position, temperature, and pain reveals sensory nerve damage and may indicate whether small or large sensory nerve fibers are affected.

Based on the results of the neurological exam, physical exam, patient history, and any previous screening or testing, additional testing may be ordered to help determine the nature and extent of the neuropathy.

Computed tomography, or CT scan, is a noninvasive, painless process used to produce rapid, clear two-dimensional images of organs, bones, and tissues. Neurological CT scans can detect bone and vascular irregularities, certain brain tumors and cysts, herniated discs, encephalitis, spinal stenosis (narrowing of the spinal canal), and other disorders.

Magnetic resonance imaging (MRI) can examine muscle quality and size, detect any fatty replacement of muscle tissue, and determine whether a nerve fiber has sustained compression damage. The MRI equipment creates a strong magnetic field around the body. Radio waves are then passed through the body to trigger a resonance signal that can be detected at different angles within the body. A computer processes this resonance into either a three-dimensional picture or a two-dimensional "slice" of the scanned area.

Electromyography (EMG) involves inserting a fine needle into a muscle to compare the amount of electrical activity present when muscles are at rest and when they contract. EMG tests can help differentiate between muscle and nerve disorders.

Nerve conduction velocity (NCV) tests can precisely measure the degree of damage in larger nerve fibers, revealing whether symptoms are being caused by degeneration of the myelin sheath or the axon. During this test, a probe electrically stimulates a nerve fiber, which responds by generating its own electrical impulse. An electrode placed further along the nerve's pathway measures the speed of impulse transmission along the axon. Slow transmission rates and impulse blockage tend to indicate damage to the myelin sheath, while a reduction in the strength of impulses is a sign of axonal degeneration.

Nerve biopsy involves removing and examining a sample of nerve tissue, most often from the lower leg. Although this test can provide valuable information about the degree of nerve damage, it is an invasive procedure that is difficult to perform and may itself cause neuropathic side effects. Many experts do not believe that a biopsy is always needed for diagnosis.

Skin biopsy is a test in which doctors remove a thin skin sample and examine nerve fiber endings. This test offers some unique advantages over NCV tests and nerve biopsy. Unlike NCV, it can reveal damage present in smaller fibers; in contrast to conventional nerve biopsy, skin biopsy is less invasive, has fewer side effects, and is easier to perform

Management

Physical Therapy assessment includes:

1. **History:** including family and medical history or any exposure to any other drugs should be assessed.

   Family history: Majority have a family history of the condition, or having neuropathy in family history. Presentation of pes cavus or abnormal gait represents of having some inherited disorder like CMT. Family tree tracking both sides of the family for 3 generations to rule out who was affected and what mode of inheritance is present whether anybody had trouble with walking, balance, tripping, falling, or with their hands or sensations should be find out.

   An autosomal dominant type will have people affected in all generation and male-to-male transmission can be seen thus males are severely affected. Whereas autosomal recessive pedigrees may have only one person or sibling affected with no family history

- At times with no family history does not preclude the diagnosis, as new mutations are relatively caused.

  Medical History: includes difficulty in balancing and problems in finding well-fitting shoes owing to high foot arches and as history of surgery been done of tendon transfer like tight heel cords, hammer toe straightening and arthrodesis of ankle.

2. **Observation:**

   Analysis of movement, observation of the condition and shape of muscle contours should be noted. Functional difficulty like clumsiness in gripping a cup or difficulty in manipulating small items such as bottle tops because of wasting of the intrinsic hand muscles. Even muscle imbalances will lead to shortening of unopposed muscle groups resulting in contractures and deformities.

3. **Cranial Nerve Examination:** Generally cranial nerves are normal at times abnormalities in papillary constriction and in optic nerve.

4. **Sensory Examination:** like pinprick, light touch, proprioception, vibration, graphesthesia and temperature should be assessed in terms of extent and pattern of involvement. Both small and large sensory nerve fibres show decreased or absent sensation. Reduction in pinprick and vibration sensation is seen and more pronounced at the toes than in more proximal muscles with Sensory ataxia (i.e., imbalance and in-coordination) due to loss of proprioception may also be seen.

5. **Muscle Strength:** should be assessed with MMT where decreased strength is noted in the distal muscles of the arms and legs like intrinsic hand and foot muscles and tibialis anterior as distal muscles degenerate (axonal loss) first while maintaining strength in proximal muscles and in the gastrocnemius muscle. They are also associated with atrophy and weakened foot eversion. Hand weakness is also seen in terms of poor finger control, poor handwriting, and difficulty using zippers and buttons, and clumsiness in manipulating small objects.[4, 5]

   A hand held myometer has been shown sensitive and reliable in assessing neuropathies. Weakness in hand muscles can be assessed by using a grip dynamometer which is used in distal neuropathies.

6. **Reflex Testing:** should be tested as deep tendon reflexes are diffusely absent (areflexia) or reduced (hyporeflexia) and can occur in any condition involving nerve damage.

7. **Girth measurement:** is done in order to detect the degree of wasting or atrophy of the muscles.

8. **Deformities:** like pes caves are common and are associated with areflexia; give the likelihood of having CMT. It results from muscle imbalance as anterior tibialis and the intrinsic foot muscles are affected with sparing of the gastrocnemius muscle thus the stronger pull of gastrocnemies overcomes the weaker pull of anterior tibialis, leading to structural foot deformities. In upper limb, claw hand deformities can be seen.

9. **Gait:** Assessment of the gait reveals difficulty in walking, twisting of ankles, slapping of the feet, or loss of a heel-to-toe pattern, and the patient may walk with a high-steppage gait (lifting legs up excessively to clear the toes) which is due to tight heel cord, and weakness in tibialis anterior leading to inadequate strength to pull the foot up during ambulation. Fractures are common. At times may require bilateral aids, such as ankle-foot orthoses, to ambulate. If quadriceps muscle weakens, hyper extension occurs at the knees to produce a rigid structure when weight bearing whereas when proximal muscles in lower limbs like hip abductors weakness is seen it results in positive Trendelenburgs sign positive.

10. **Autonomic Dysfunction:** Sweating due to poor vasomotor control leading to cold feet with blotching or pallor of the skin of the feet is noticed.

11. **Neuropathic pain (burning, tingling, shooting)**: may also occur as a result of the neuropathy, while bone and joint pain may result from pressure on the feet. Muscle cramps and restless legs is also been noticed. Neuropathic pain can be treated with gabapentin, pregabalin, duloxetine, and amitriptyline. Topical lidocaine patches may help localized pain. Narcotic analgesics need to be carefully considered. Joint, bone, and muscle pains require different approaches and should be treated appropriately.

12. **Respiratory System:** is examined in terms of rate of respiration, chest expansion and vital capacity as in acute phase of polyneuropathy especially in GBS, respiration is compromised and at times the patient may need ventilator for respiratory assistance.

13. **Fatigue testing:** Fatigue can seen in any activities such as getting tired while writing letters or walking distances. Fatigue severity

scale has been demonstrated to be reliable measure in neuropathies. The patient is asked to maintain a diary documentation activities and length of time performing each one and the symptoms of fatigue. Later it is to be analyzed to try and develop a structure to the patients activities that prevent further fatigue and also to check whether this is not leading to disuse by cutting out all activity that cause any degree of fatigue.

14. **Functional Assessment:** is also measured to any change in performance level. For e.g. ability to rise from a chair and standing on heels in case of diabetic neuropathy.

15. **Electrophysiological tests:** is done to differentiate between a demyelinating and an axonal process. It serves as a prognostic factor in determining whether there are any axonal changes as seen in GBS where once axonopathy is permanent, there are no changes left for repairing the axon.

16. **Gene testing and counselling:** Genes for inherited neuropathies can be identified by deoxyribonucleic acid (DNA) testing. Once a patient is shown to have the gene, counseling can be offered and the implications for other family members are discussed. Prenatal testing can be identify a fetus at risk of developing an inherited neuropathy and give the parents the option of termination.

Management:

Physiotherapy Interventions:

Acute Neuropathies:

Physical management in acute neuropathies includes prevention of contractures, control of pain and respiratory care.

1. Stretching to prevent contractures.

Neuropathy may result in severe disability making the patient totally bed-ridden. When the muscles are not stretched adequately but are left in a shortened position, structural changes, involving loss of sarcomeres, occur and compromises potential recovery. The therapist should ensure that all structures including the nervous system are moved through their full range. Adverse neural tension signs can occur if neural tissues are not stretched.

Continuous passive motion machine has been suggested as a mean to maintain range of movement. Even passive ROM is given 2-3 times in a day. Tightening of posterior crural muscle group develops rapidly. Able patients should be taught self -stretches in a weight-bearing position. For bedridden patients, foot drop splints should be provided and gentle stretches performed. Thus muscles which crosses two or more joints must be stretched to its full length.

2. Positioning:

Frequent position changes are recommended in bed ridden patients to prevent selective muscle shortening and pressure sores. Truncal weakness can cause the patient to lie with scapulae retracted, unless a wedge is positioned under the thoracic region as well as the head and shoulders. The pelvis should be supported in a position of posterior tilt with the hip flexors stretched, to prevent shortening. Every two hourly turning helps in preventing pressure sores. If the sore is developed, ultra-violet radiation is given to enhance the healing process. The patient's position is maintained with the help of sand bags, pillows etc. If splint is given, then frequent checking is maintained to avoid the skin breakdown around the application area.

3. Maintenance of Circulation:

Passive exercises help in maintaining the circulation.

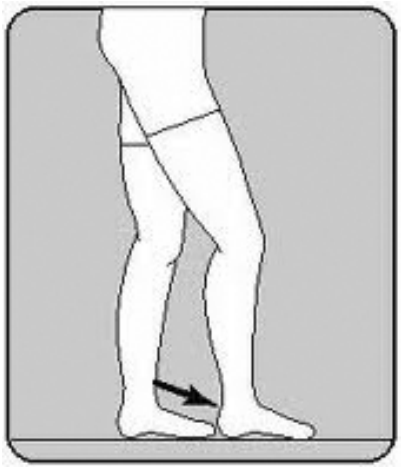
4. Pain :

Pain is an early symptom in acute neuropathy such as GBS due to spontaneous discharge in demyelinated sensory neuropathies. Freeman suggested that the reason why patient with acute GBS enjoy large amplitude mid range movements is because of their pain relieving properties.

5. Respiratory Care:

In acute cases, the patient may be ventilated and the role of physiotherapist is to help prevent atelectasis when breathing is compromised. Since the respiratory system is involved, the aim would be to maintain clear airway by removing the lung secretions through the following technique;

1. Intermittent positive pressure breathing.
2. Postural drainage.
3. Suction catheter.
4. Vibration and rib shaking.



Tandem walking

Side Sitting in Quadrapud (a)

Side sitting in Quadrapud (b)

Weight Shifts in Quadrrapud

One leg standing

Balance Board Exercises.

Once the patient is weaned from the ventilator, he is taught effective coughing and good breathing exercises.

When there is facial muscle weakness, care must be taken to ensure there is lip seal around the mouth piece when measuring vital capacity. Where the autonomic system is affected, disturbed blood pressure is seen when attempting suction or making early sitting.

Chronic Neuropathies:

Physiotherapy plays a major role in chronic neuropathies which help in maintaining ambulation and prevention of contractures. It helps in facilitating functional recovery in bedridden patients of neuropathies thereby preventing complications and improving the quality of life of patients with chronic neuropathies.

Physiotherapy interventions in chronic neuropathies include:

1. Strength Training :

The aim is to maintain the strength of weak muscles which can be achieved by the following techniques:

1. Free active exercises.
2. Proprioceptive Neuromuscular Facilitation (PNF).
3. Progressive resisted exercises.
4. Suspension therapy exercises.
5. Equilibrium and righting reactions.

Low-intensity to moderate intensity strength training programs in patients with neuromuscular disease, including Charcoat Marie Tooth disease (CMT), show varying results.

Kilmer et al.1994 found that higher resistance exercise was no better than low resistance exercises for strengthening muscle and there was some evidence of damage when tested eccentrically. Endurance training can improve cardiopulmonary fitness seen in established neuropathies. Six week home programmed of strengthening, stretching and aerobic conditioning increases strength and quality of life.

Concentrating on the hip and knee helps in compensating and producing a more stable gait pattern. The additional AFO would also increase stability by allowing the development of plantar flexion movement despite very weak muscles, as well as preventing tripping. Abnormally prolonged muscle ache following exercise or sudden loss of functional ability indicates damage due to overuse and modification of exercise to be tailored made in order to allow recovery.

In one randomized study where CMT patients underwent home-based strength training were evaluated using outcome measures such as isokinetic knee extension and flexion; maximal voluntary contraction; endurance at 80% MVC; ability to descend and climb stairs and stand up from a chair or from lying supine; and time to walk 6 m at a comfortable pace or 50 m quickly showed a significant reduction in the time taken for a 6-meter walk at 24 weeks after starting the exercise with no other significant improvements in time-scored functional activities

Both studies observed limited improvement in upper-body and lower body strength, but Kilmer and coauthors [88] observed a high number of injuries in their patients and concluded that increases in training frequency, volume, and especially intensity may put patients with neuromuscular disease at increased risk of training-induced injury.

Recently 24-week cycling program using a cycloergometer in limited number of patients (4 with CMT1A and 4 with CMT2) proved to be effective in improving exercise tolerance and functional ability in the absence of significant changes in fatigue resistance. Another study tested the effects of a 12-week, home-based resistance exercise program on strength, body composition, and activities of daily living (ADLs) in 9 men and 11 women with CMT1A (n=18) and CMT2 (n=2). Strength and ADLs improved equally in men and women, and there were no differences between CMT1A and CMT2. Therefore, similar rehabilitative approaches may be useful in CMT, but with the safety and efficacy. Also, overloading of the weakened muscles can cause fatigue and weakness thus overloads principle need to be considered.

2. Stretching:

Gentle stretches for muscle groups particularly in HMSN, are done to the muscles that are liable to shorten thereby preventing muscle tightness and loss of strength. Tightness and contracture affects the extensibility of muscle and surrounding connective tissues. Regular stretching of 15 to 30 seconds trice a day can prevent or reduce joint deformities that may

result from uneven pulling of muscle on bones. Serial night casting for 4 weeks induced a small increase in ankle dorsiflexion range in children and young adults with CMT.

A recent case report suggested that when triceps surae muscles are weakened, stance and gait gets affected however range of motion is affected.

3. **Re-education of sensory awareness:** The sensory system can be stimulated by the cutaneous stimulation which can be given by the different material, textures, shapes and weights. Equilibrium and righting reactions help in reeducating the proprioception of sensory system. Vision can also be used as an alternative system in reeducating the sensory awareness.

4. **Postural kinesiotherapy:** May be helpful in reducing the need to control joints from three joints (hip, knee, and ankle) to one joint (hip), and proprioceptive kinesitherapy may help to improve coordination.

5. **Balance Training :**

   Balance training plays major role as weakness and deformities at foot reduces balance thereby increasing walking in stability. Thus it should be emphasize by including the following exercises:

   1. Standing with feet apart in comfortable posture.
   2. Standing with feet apart and keeping the arms in different positions for support.
   3. Standing with close feet.
   4. One leg standing.
   5. Repeat all the exercises with eyes closed.
   6. Balance boards exercises.
   7. Quadruped side sitting.
   8. Hip extension in quadruped.
   9. Bilateral arm and leg raise in quadruped.
   10. Weight shifts in forward, backward and sideways direction in quadruped.
   11. Tandem walking. Walking sideways and backward.
   12. Walking and stopping alternately.
   13. Ask the patient to walk in circles.
   14. Walk on toes.
   15. Walk on heels.
   16. Intrinsic strengthening.

6. Functional and mobility aids used for normalizing the gait:

   Aids used for function and mobility include a range of orthotics and wheelchairs.

Gait and balance problems are important risk factors for falls in CMT as well as in other peripheral neuropathies. Where ever there is an abnormal gait pattern, mainly due to the weakness of the dorsiflexors and / or the intrinsic muscles of the feet, the use of orthosis should be advised. It has been seen that foot deformities have become irreversible and insoles are necessary to allow the foot to sit comfortably in a standard shoe and to redistribute the weight across the total surface of the sole. When foot drop exists, light polypropylene splint should be advised.

It is essential to discuss skin care with the patient as most of it has sensory deficit also.

Ankle-foot orthoses (AFOs) may be useful in treating patients with CMT. AFOs compensate for weakness and correct foot drop, can offer a control of the foot, can help control unwanted inward rotation of the foot, and facilitate a more normal gait pattern.

Prescription of AFO for a patient with CMT can enhance physiological performance and perceived exertion at sub maximal activity levels.

Falls can also be prevented by paying attention to the floor, avoiding uneven ground, being wary of rugs and carpets, avoiding dark places, using a handrail when going up and down the stairs, avoiding haste, and staying slim. When falls are frequent, a walking stick, one or two crutches, or a walking frame is required; it can also be advisable to use a wheelchair outside of the house.

Wheelchairs: In some of cases, the neuropathy progresses to render the patient dependent on a wheelchair. For e.g. in the early stages of rehabilitation, a patient with GBS, a reclining wheelchair is valuable to coping up possible fluctuations in blood pressure and to allow gradual accommodation in the upright position.

7. Manual dexterity training :

Hand function is also compromised through weakness or paralysis of intrinsic muscles of the thumb and fingers causing difficulty in pincher gripping and finger movement. Normal daily activities such as dressing, bathing, holding cutlery, or writing becomes difficult. Tripod pinch strength and thumb

opposition are major determinants of manual dexterity in CMT and should therefore be the focus of intervention strategies that aim to preserve or enhance manual dexterity in CMT. Simple thumb opposition splint may allow a patient to produce legible writing for a longer time, or to grip a cup or knife. At times when there is thenar eminence weakness and wasting is being seen, a night splint cast in a functional position will help prevent severe contracture. Use of putty and rubber bands of different strengths can be used to improve strength of hand and forearm muscles.

8. Adaptive devices:

Special tools modification is given at times in order to avoid overwork and weakness thereby improving performance. Hand or forearm splinting may be advised later once wasting of the hand muscles sets in. Adaptive equipment may be prescribed to compensate for the hand deformities, sensory loss, and weakness which include a button-hook, a long-handled shoehorn, and elastic shoelaces.

9. Neuropathic Pain Management:

Malalignment of joints due to muscle imbalance often leads to pain. In chronic neuropathies, neuropathic pain is seen which is caused by damage or dysfunction in the nervous system, which includes the spinal cord. It can generally be described as a sharp, shooting, or burning pain,

Management of neuropathic pain becomes very important as it hinders to the day-to-day activities of patients. Pain management can be divided into physiotherapy, behavioural and cognitive interventions.

A) Physiotherapy interventions include:

1. **Thermotherapy** - can reduce muscle spasm thereby relaxing the muscle.
2. **Cryotherapy** - can be used in case if there is any swelling at some distal joints.
3. **Massage** - can reduce muscle tension by gentle touch thereby increasing local blood circulation and relaxing muscles and soft tissues.
4. **Transcutaneous Electrical Nerve Stimulation (TENS) :** is proved to be useful in neuropathic pain. Low rate TENS or Acupuncture Like TENS acts on small diameter C fibers and acts on mechanisms of endogenous opiates thereby blocking pain gate mechanisms.
5. **Ultrasound:** has also been used in peripheral neuropathy as it reduce muscle spasm and pain thereby acting on both large diameter A fibers and small diameter Cfibers.

B) **Cognitive Strategies:** are used in treating together the body, mind and soul.

C) **Behavioural interventions:** Distraction - listening to music or using imagery techniques can be helpful during brief episodes of pain or painful procedures. Music therapy has been used successfully to reduce disruptive behaviour or aggression attitudes.

10. **Aerobic Exercises:**

Patients with chronic neuropathies show reduced peak oxygen consumption and decreased functional aerobic capacity, and studies shows aerobic exercise improve functional ability and aerobic capacity. Aerobic walking has been used in neuromuscular disorders and was effective in ameliorating peak power output and peak oxygen intake, walking ability, and metabolic changes.

11. **Hydrotherapy:** is advised as it increases the muscle power, improve coordination and balance of the patient. Care is taken in case of flaccid joint. Pain which is also a feature of polyneuropathy can be relieved by hydrotherapy as water has pain relieving property.

Occupational Therapy Management

Occupational therapy professionals should be involved early in the rehabilitation program to promote upper body strengthening, ROM, and activities that aid functional self-care. Both restorative and compensatory strategies can be used to promote functional improvements. Energy conservation techniques and work simplification also may be helpful, especially if the patient demonstrates poor strength and endurance. Participation in recreational therapy assists in the patient's adjustment to disability and improves integration into the community. Recreational activities, either new or adapted, can be used to promote the growth, development, and independence of a long-term hospital patient.

Occupational Therapy assessment includes:

- History
- Muscle Strength
- Range of Motion
- Physical Endurance
- Gross Motor coordination
- Fine Motor coordination, manipulation and dexterity
- Sensory Registration or awareness
- Sensory processing: discrimination of light touch, stereognosis, proprioception, and two-point discrimination
- Cranial Nerve Examination
- Pain
- Respiration
- Swallowing
- ADLs
- IADLs
- Productivity history, skills, interest and values
- Leisure

Occupational Therapy Intervention:

Occupational therapy is instrumental in helping the patient cope with the functional, vocational, and social impact of peripheral neuropathy by:

- Improving sensory-motor skills
- Teaching self-care activities
- Teaching the patients management of fatigue.
- Teaching the patient safety issues, (e.g., paying more attention to the terrain when walking since falling or tripping may pose a risk for patients with PN)

Management to improve Sensory Motor Skills:

People with lack of protective sensation are at risk of serious injury since they can not feel pinprick or hot or cold exposures

Some protective sensory reeducation techniques include:

- Protect from being exposed to sharp items or to cold or heat
- Try to soften amount of force when gripping an objects
- Use built-up handles on objects whenever possible to distribute gripping pressure over a greater surface area.
- Do not persist an activity for prolonged periods of time. Instead, change the tool used and the work task often.
- Visually examine the skin for edema, redness, warmth, blisters, cuts or other wounds (tissue heals more slowly when there is nerve injury).
- Before bathing or showering, use a thermometer to test the temperature of the water.
- Always use pot holders when cooking or handling items on a stove or in an oven, and use thick gloves when washing dishes or working with sharp utensils. Wear low- or flat-heeled shoes that are of an appropriate size, and wear thick socks to protect against blisters.

Motor intervention

After partial or complete denervation of muscle tissue and during inactivity or disuse, muscle strength decrease. Active- assisted, active exercises are used to increase strength when strength is inadequate substitution pattern or trick movements are likely to develop.

Suspension exercises are advised for all the shoulder muscles. Many purposeful activities can be given to maintain / improve strength.

Clay pressing

In the presence of contracture, Passive ROM with gentle movement of joints should be given and it should not exceed the point of pain.

Manual dexterity is of utmost importance in performing activities of daily living and is described as the ability to move the hands easily and skillfully, to work with the hands in turning and placing motions.

Some Standardized Test for Hand Functions: In HSMN, distal hand muscles weakness leads to

difficulty in gripping object and performing fine motor activities.

There are some standardized tests to evaluate dexterity of hands.

Sollerman hand function test (SHT): This test is used for the evaluation of dexterity in HSMN. This test was developed to provide an overall measure of hand and grip function when engaging in ADLs (Sollermen & Ejeskar 1985). It was designed to measure grips that are needed for certain ADLs such as eating, driving, personal hygiene, and writing. The test includes subtests that represent common handgrips (volar, transverse volar, spherical volar and pinch positions - pulp, lateral, tripod, and the five finger) and activities (using a key; picking up coins from a flat surface; writing with a pen; using a phone; and pouring water from a jug).

Functional Dexterity test (FDT):

The FDT measures the ability to perform a tripod pinch through the timed manipulation of pegs (administration time about 5 min). A tripod pinch pattern is frequently used during daily activities such as eating, writing and tying. This grip pattern in particular, may become problematic when the intrinsic muscles of the hands are affected.

Activity to Improve Hand Strength and Dexterity

Play dough / cookie dough-Rolling it out, pulling it apart, forming a ball between the palms or a small ball with the fingers, pinching it with thumb and index finger.

Clay rolling

Posting activities-Pushing pennies or dried pulses into a slot or hole in the top of a Pringles tin box or a plastic container.

Tearing activities-Tearing paper, junk mail etc into strips then crushing it into a ball and filling up a box to hide things inside. Encourage tearing with the thumb, index and middle fingers to perform fine motor coordination.

Clay pinhing

Vegetable stamping-Cut vegetables such as onions and potato in half and dip in paint. Print on paper taped to wall to strengthen shoulder, wrist and fingers muscles.

Tongs-Use tongs of different sizes and stiffness to develop hand strength. Pick up objects such as cotton balls, marbles, small dried peas etc; this can be graded to increase resistance and skill.

Elastic band pulls-Use elastic bands of different thickness resistance and, stretch them over pegs in a board or nails hammered into wood etc to strengthen intrinsic muscles of hand.

Sealing bags-Use Jiffy bags or equivalents that can be sealed by pressing the edges together. Get your child to use just the thumb and index / middle fingers if possible

Tug of war-This increase gross grasp strength and contraction in the bigger joints up the arms. Use a towel, sheet or soft rope in sitting or standing position.

Screwing and unscrewing-Use a variety of bottles and containers with screw tops to build finger strength. Screwing and unscrewing of nuts and bolts increase strengthen the small muscles in the fingers.

Buttering bread-Good for grasp strength and also provides an opportunity to develop bilateral skills and motor planning and sequencing skills.

Peeling fruit-peeling fruit develops finger strength

Vertical drawing/vertical activities-Drawing on a vertical surface is an excellent way to improve grasp strength. Break the chalk or crayon into small pieces of around half to one inch long as this helps to

develop the tripod grasp and encourages a better wrist position. Working on an elevated vertical surface also improves shoulder stability.

Because many neuropathy patients lose sensations in their hands and feet, it may be necessary to take several steps to ensure that everyday tasks are safe.

Management for maximizing independence in ADLs: (Self Care Activities)

Adaptive and assistive devices may be introduced to decrease effort during daily activities

BATHROOM:

A. BATHING:

- Sit on a tub bench or bath stool
- Use a bath mitt or long handled brush / sponge
- Install grab bars around the tub
- Install lever-type faucets or build up the faucet handles to decrease stress on hand / finger joints
- Use a non-skid rubber mat or strips in the tub or shower
- Keep towels in easy reach
- Soap on a rope or liquid soap to avoid dropping soap
- Do not use glass containers that could be dropped and broken

B. TOILETING:

- Put grab bars around the toilet
- Use a raised toilet seat
- If a raised toilet is too high, then push a low footstool under your feet once you are sitting

C. GROOMING:

- Build up or extend the handles on brushes, combs, toothbrushes etc., using rulers, foam rubber, or pipe insulation
- Use an electric toothbrush
- Use pump dispenser type toothpaste. If you have to squeeze out toothpaste, squeeze the tube between both palms or place the tube over a damp washcloth and lean onto it
- Place a foam curler over eyeliner or lipstick to build up a handle
- If holding one or both arms up to wash or style hair is fatiguing, try sitting at a desk or dressing table. Prop an elbow on a book (or 2) covered with a soft towel.

D. DRESSING:

- Sit down to dress
- Reorganize closet so that the shelves are lower and clothes are hung at a lower height
- Use long handled shoehorns and sock aids
- Wear supportive and proper fitting shoes
- Wear "pretend" or clip-on neckties
- To make zippers easier to grasp, use a zipper pull, add a loop chain or large paper clip
- Replace buttons with velcro if possible, or use a buttonhook

Management for fatigue

An important aspect of treatment is to educate the client and family members to use energy conservation and work simplification strategies when engaging in ADLs, IADLs and leisure occupations.

II. KITCHEN:

A. ORGANIZING SPACE/WORK AREA

- Keep pots and frying pans near the stove
- Keep frequently used appliances (e.g. Toaster) on the counter
- Set up a specific area to make coffee (i.e. A coffee station where you keep everything you need nearby.
- Keep all baking equipment in one cupboard
- Keep items to the front of the shelves and the rows shallow
- Use plastic lid holders on inside of cupboard doors to keep lids organized
- Eliminate clutter by organizing drawers and dividers
- Keep unused duplicate items in other areas if needed or give away or throw out (i.e. scissors)
- Keep frequently used items to the front of the shelf and items used less often to the back

- Use space savers (e.g. lazy susans) and pullout shelves
- Keep heavy jars and boxes at waist level
- Store only light objects on the higher shelves only if absolutely necessary and use a long handled reacher or tongs to reach items
- Keep heavy items on top shelf of refrigerator, near the front

B. COOKING:

- Plan meals ahead to decrease / prevent last minute tasks
- Use precut vegetables, chopped nuts (convenience food)
- Sit town while preparing vegetables, meat, etc., for cooking
- Use a mirror over the stove to monitor food while sitting
- Use electric appliances (e.g. Microwave, electric mixer / can opener / knives / fry pan)
- Can use egg slicer for any soft vegetables
- Place a damp cloth underneath a bowl or plate to keep it from moving
- Use lightweight dishes / pans and serve from them
- Use a wheeled cart to move heavy items around
- Re-arrange kitchen to make it accessible from a seated position
- Prepare large amounts of food that freezes well, then freeze for later meals
- Use built up handled utensils for more comfortable grip
- Use wall mounted manual or electric jar and can openers.

C. CLEAN-UP:

- Use disposable aluminum baking pans or plastic baking bags
- Use a nonstick product on pans or line with foil before baking or frying
- Sit on a stool while washing dishes
- Use a wheeled cart to collect dishes from table
- Use an easy to grip sponge to clean up rather than a thin dish cloth

III. LAUNDRY:

- Do only what is necessary: buy permanent press clothing
- Use separate baskets to sort clothes before bringing them to the laundry room or keep hampers / baskets in the laundry room to collect dirty laundry
- Use a rolling cart to move wet clothes to the clothesline if a dryer is not available
- Sit when sorting, folding and ironing clothes (some ironing boards are adjustable)
- Label a basket for each family member and have each one put their own laundry away
- Sort clean clothing and linen into different baskets and have other family members put them away.
- Use a rolling cart if you put the laundry away

HOUSECLEANING:

- Store cleaning supplies everywhere they are used
- Wear an apron with large pockets to carry necessary cleaning supplies
- Use a sponge mop with an easy lever-type squeezer, a "janitor's pail". move the bucket / pail of cleaning solution on a rolling dolly / caster
- Use a long handled feather duster or dust with a mitt
- Use automatic toilet bowl cleaner & spray on mildew remover to eliminate scrubbing
- Put casters on furniture so they can be moved easily
- When making beds, finish one side at a time
- Alternate heavy and light tasks, and remember to take a break between / during tasks
- Do only one major cleaning task a day (e.g. Laundry, cleaning the bathroom)

WORKPLACE:

- Keep frequently used, necessary items in easy reach
- Sit in a swivel chair with proper adjustable back support
- Set the work surface at a comfortable height
- If you are required to make many errands, use the elevator and organize the errands to make as few trips as possible

LEISURE:

- Use a card rack for holding playing cards
- Use a bookstand. Lay the newspaper on an open table rather than holding it up with your arms
- Use felt tip pens which require less pressure to write. Also, use larger sized pens for easier grip
- Use a push-button phone or pen tip to dial
- Attach a gooseneck to the table then to the telephone receiver to eliminate holding the receiver.
- Get a headset for the phone to eliminate the need to hold the receiver.

GARDENING OR YARDWORK:

- Use lightweight tools with extended or built up handles.

o Use a gardening stool

o Use raised flower boxes or window boxes

FOR TRAVEL:

o ask for a wheelchair escort to the gate

o Ask a porter to assist with luggage or get a luggage cart, or get luggage with wheels

o use elevators, escalators moving walkways

CAR:

o get power options steering, windows, brake, locks, seat controls

o use a wide-angled mirrors if you have decreased movement in your neck

o build up the key tops or use key holders to make turning easier

General Precautions:

- Watch for redness over bony areas of the body and change the person's position every two hours
- Watch the person for signs of fatigue. Do not continue activity if signs of fatigue are present
- Have the person maintain good posture and positioning at all times to protect joints while muscles are weak.
- Avoid irritation of inflamed nerves. Go slowly on exercise program.
- Teaching the patient to pay attention to issues which involve automatic functions (e.g., learning how to change positions smoothly to avoid a sudden drop in blood pressure and the risk of falling)

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Section 4
Miscellaneous

Ch.12 Spasticity

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Lance in 1980 gave the definition of spasticity as 'Spasticity is a motor disorder characterized by a velocity-dependent increase in tonic stretch reflexes (muscle tone) with exaggerated tendon jerks, resulting from hyperexcitability of the stretch reflex, as one component of the upper motoneuron syndrome 1

Upper motor neurons originate in the brain and brain stem and project to lower motor neurons within the brain stem and spinal cord.2 The lower motor neurons are of two types, both of which originate in the ventral horn of the spinal cord: (1) alpha motor neurons project to extrafusal skeletal fibers and (2) gamma motor neurons project to intrafusal muscle fibers within the muscle spindle.2 With a lesion of the CNS comes interruption of the signals sent via the upper motor neurons to the lower motor neurons or related interneurons.

Spasticity usually is accompanied by paresis and other signs, such as increased stretch reflexes, collectively called the upper motor neuron syndrome. Paresis generally affects distal muscles, with loss of the ability to perform fractionated movements of the digits. The upper motor neuron syndrome results from damage to descending motor pathways at cortical, brainstem, or spinal cord levels and spasticity evolves in the days and weeks after injury. When the injury that leads to spasticity is acute, muscle tone is flaccid with hyporeflexia before the appearance of spasticity. The interval between injury and the appearance of spasticity varies from days to months according to the level of the lesion. In addition to weakness and increased muscle tone, the signs in spasticity include clonus, the clasp-knife phenomenon, hyperreflexia, the Babinski sign, flexor reflexes, and flexor spasms.

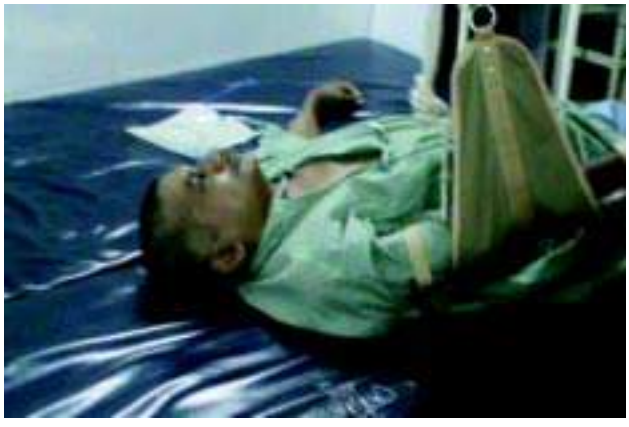


Fig. 1: The stretch reflex arc.

The stretch reflex arc is the most basic neural circuit contributing to spasticity. When a muscle is stretched, an impulse is generated in the muscle spindle and is transmitted via the sensory neuron to the grey matter of the spinal cord. Here the sensory neuron synapses with the motor neuron, and the transmitted impulse results in muscle contraction. While agonist muscles contract in response to stretching, antagonist muscles must relax. Their relaxation is brought about via an inhibitory neuron within the spinal cord.

The alpha motor neuron and the muscle comprise the final common pathway in the expression of motor functions, including spasticity. There are numerous excitatory and inhibitory modulatory synaptic influences on this pathway. An imbalance in these influences results in hyperexcitability of the stretch reflex arc, which is thought to be the basis for spasticity. Some of the factors that play a role in suppressing hyperactivity of the final common pathway (Fig. 2) include cerebral inhibitory pathways (from the brain) and spinal mechanisms such as nonreciprocal Ib inhibition (from golgi tendon organ receptors in tendons), presynaptic inhibition of the Ia terminal (at the axoaxonic synapse between 2 axons), reciprocal Ia inhibition (inhibition of antagonistic muscles) and recurrent Renshaw inhibition (inhibitory feedback of the alpha motor neuron cell body by the inhibitory interneuron).3

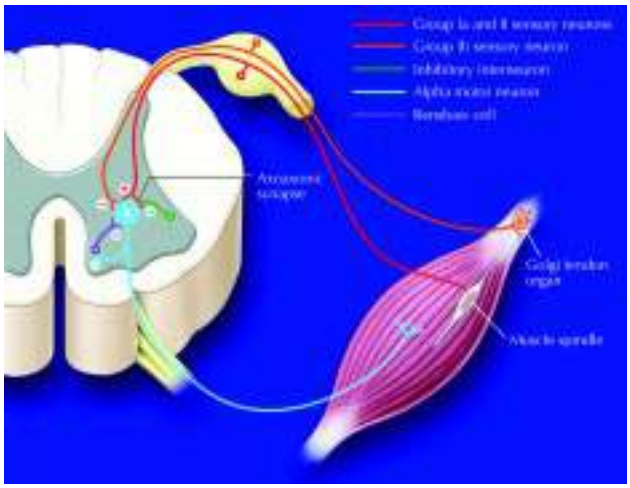


Fig. 2: Potential spinal mechanisms of suppression of hyperactivity in the final common pathway (alpha motor neuron and muscle). There are numerous excitatory and inhibitory modulatory synaptic influences on this pathway. An imbalance in these influences results in hyperexcitability of the stretch reflex arc, which is thought to be the basis for spasticity. Factors that play a role in suppressing hyperactivity of the final common pathway at the spinal cord level include nonreciprocal Ib inhibition (from golgi tendon organ receptors in tendons), presynaptic inhibition of the Ia terminal (at the axoaxonic synapse between 2 axons), reciprocal Ia inhibition by the inhibitory interneuron (inhibition of antagonistic muscles [see Fig. 1]) and recurrent Renshaw inhibition (inhibitory feedback of the alpha motor neuron cell body by the inhibitory interneuron).

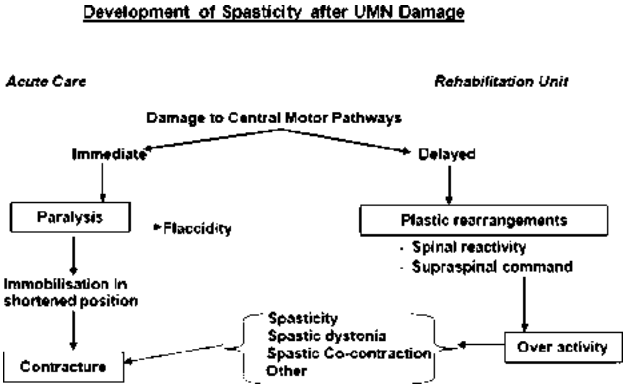
Pathophysiology

Spasticity arises from prolonged disinhibition of spinal reflexes as a result of UMN lesion. These spinal reflexes include stretch, flexor and extensor reflexes and are under supraspinal control by inhibitory and excitatory descending pathways. Stretch reflexes are proprioceptive reflexes, and are either phasic or tonic. The tonic stretch reflex arises from a sustained muscle stretch and is the cause of spasticity (Sheean 2002). Stretch reflex is dependent on tendon lengthening and excitatory post synaptic potentials (EPSPs) carried by I-a afferents but Inhibitory post synaptic potentials (IPSPs) arising from antagonistic muscle spindles, oligosynaptic and polysynaptic pathways also have an important role in the maintenance of tone (Lance 1980, Young 1994, Nathan 1973).

Damage to pyramidal tracts alone does not result in spasticity. It occurs only when the lesion involves premotor and supplementary motor areas. Selective damage to area 4 in the cerebral cortex of primates produces paresis that improves with time, but increases in muscle tone are not a prominent feature. Lesions involving area 6 cause impairment of postural control in the contralateral limbs. Combined lesions of areas 4 and 6 cause both paresis and spasticity to develop.[3] Physiologic evidence suggests that interruption of reticulospinal projections is important in the genesis of spasticity.[4] In spinal cord lesions, bilateral damage to the pyramidal and reticulospinal pathways can produce severe spasticity and flexor spasms, reflecting increased tone in flexor muscle groups and weakness of extensor muscles.4

Spastic dystonia is primarily due to abnormal supsraspinal descending drive, which causes a failure of muscle relaxation and is sensitive to the degree of tonic stretch imposed on that muscle (Denny-Brown 1966). There is inappropriate recruitment of antagonist muscles in spastic co-contraction upon triggering of the agonist under volitional command. This occurs in the absence of phasic stretch and is sensitive to the degree of tonic stretch of the co-contracting antagonist (Gracies et al. 1997). For instance, triceps will be recruited during volitional action of biceps and will lead to elbow stiffness Associated reactions are found in muscles that are not particularly stretch sensitive. They include, when there is extra-segmental co-contraction due to cutaneous or nociceptive stimuli, or inappropriate muscle recruitment during autonomic or reflex activities, such as yawning.

The resultant pattern is determined by the age, size and location of the lesion. Supra-bulbar lesions present predominantly with flexor patterns of spasticity, whereas spinal cord lesions produce extensor patterns predominately. Patients with partial lesions, where sensation is intact or partially intact, are typically bombarded by nociceptive inputs and display greatly increased ?-motor neuron activity. Different patterns emerge early on after the neurological insult and later, when patients may find themselves in a rehabilitation unit. The following figure shows the effects of the different scenarios.



An explanation of Figure 1 is available

Immediately after injury, a period of neuronal shock occurs and spinal reflexes are lost, which include stretch reflexes. A flaccid weakness in seen, but even during this, the positive features of hypertonia can start to be seen. Limbs are not sufficiently stretched and may be immobilized in shortened positions. Rheological changes occur within

muscles in the form of loss of proteins and sarcomeres and accumulation of connective tissue and fibroblasts (Ward 1999). Unless treated, tendon and soft tissue contracture and limb deformity are established. Altered sensory inputs such as pain, recurrent infection and poor posture, maintain a further stimulus to lead to yet further shortening, and this cycle is difficult to break.

Spasticity is set up later on, as plastic rearrangement occurs within the brain, spinal cord and muscles.5,6,7,8,9,10 This attempt at restoration of function through new neuronal circuitry creates movement patterns based on existing damaged pathways. Neuronal sprouting occurs at many levels with interneuronal endings moving into unconnected circuits from decreased supraspinal command through the vestibular, rubrospinal and reticulospinal tracts (Krenz and Weaver 1998). The end-effect is muscle over activity and exaggerated reflex responses to peripheral stimulation (Farmer et al. 1991). This process occurs at anytime, but is usually seen between one and six weeks after the insult. Muscle over activity declines over time and the following are suggested as possible causes:

- Structural and functional changes due to plastic rearrangement
- Axonal sprouting
- Increased receptor density

Intrinsic tonic spasticity

Decq2 has differentiated intrinsic tonic spasticity (increased muscle tone) as that component of spasticity resulting from an exaggeration of the tonic component of the stretch reflex. Briefly, the stretch reflex is a monosynaptic reflex pathway that originates in the muscle spindles embedded parallel to the muscle fibers and travels via a Ia afferent to the spinal cord, where it synapses either first with interneurons or directly with an alpha motor neuron innervating the muscle from which the stimulus originated.11 The tonic component of the stretch reflex associated with increased muscle tone results from a maintained stretch of the central region of the muscle fibers and the reflex is polysynaptic.11 Upon a sustained stretch, both type Ia and type II afferents (from secondary spindle endings) synapse with interneurons within the ventral horn of the spinal cord. Synapses of the interneurons with alpha motor neurons facilitate contraction in the muscle being stretched.

It is the hyperexcitability of this tonic stretch reflex that is commonly thought to result in increased muscle tone in response to passive stretch following SCI. This hypertonia is velocity-dependent, with faster stretching velocities being associated with greater amounts of reflex activity. The development of tonic stretch reflex hyperexcitability could be due to a lower threshold, an increased gain of the stretch reflex, or a combination of the two. The resultant increase in muscle tone is thought to be due to a combination of increased denervation hypersensitivity and changed muscle properties.11, Denervation leads to an initial downregulation of neuronal membrane receptors, followed by an upregulation, with enhanced sensitivity to neurotransmitters.2 Gradual changes in muscle properties also occur following SCI, such as fibrosis, atrophy of muscle fibers, decrease in the elastic properties, decrease in the number of sarcomeres, accumulation of connective tissue, and alteration of contractile properties toward tonic muscle characteristics, which likely contribute to the increased passive tension.11,

Intrinsic phasic spasticity

Intrinsic phasic spasticity encapsulates symptoms such as tendon hyper-reflexia and clonus, and is due to exaggeration of the phasic component of the stretch reflex.2 Tendon hyper-reflexia is identified as an exaggerated muscle response to an externally applied tap of deep tendons.7 Reduced presynaptic Ia inhibition is thought to play an important role in this hyper-reflexia, as the occurrence of reduced presynaptic inhibition of group Ia fibers appears to correlate with the excitability of tendon reflexes.36

Clonus has been defined as 'involuntary rhythmic muscle contraction that can result in distal joint oscillation'37 and most often occurs at the ankle.2, 7, 9 Clonus is elicited by a sudden rapid stretch of a muscle.38 The prevailing theory explaining the underlying mechanism responsible for clonus is that of recurrent activation of stretch reflexes.11, 37, 38 According to this theory, dorsiflexing the ankle causes activation of the Ia muscle spindle afferents and induces a reflex of the triceps surae, resulting in plantar flexion of the ankle.11, 37, 38 This reflex contraction is brief, essentially phasic, and ceases rapidly.2 The muscle then relaxes, causing the ankle to be dorsiflexed once again, due either gravity or the stretch being sustained by an examiner.2 The result is a new stretch reflex, etc.2, 37 Ultimately, it is the disinhibition of the stretch

reflex due to interruption of descending influences with SCI, that is thought to cause exaggeration of the phasic stretch reflex pathway and, hence, clonus.3

The second theory is that clonus is the result of activity of a central oscillator or generator within the spinal cord, which rhythmically activates alpha motor neurons in response to peripheral events.37, 38 Beres-Jones et al37 outline observations that they feel support such a hypothesis: (1) reports of similar frequencies of clonus among ankle, knee, and wrist muscles, (2) observations that the clonus frequency is not entrained by the input frequency, suggesting that clonus cannot be solely stretch-mediated, (3) the finding that stimuli other than stretch evoke clonus, and (4) the observation of a refractory period following the clonic EMG burst where tendon tap, H-reflex stimulation, and vibration fail to elicit an efferent response. Therefore, whereas reduced presynaptic inhibition of group Ia fibers appears to be among the contributing factors to tendon hyper-reflexia, the underlying mechanism of clonus has not been clearly elucidated.

Extrinsic spasticity

In addition to the various intrinsic factors that contribute to symptoms of spasticity, involuntary muscle spasms can also occur in response to a perceived noxious stimulus originating extrinsic to the muscle: extrinsic spasticity.2, 3, 7 Flexion spasms are the most common form of extrinsic spasticity, triggered by afferent input from skin, muscle, subcutaneous tissues, and joints (collectively referred to as 'flexor reflex afferents'). These flexor reflex afferents mediate the polysynaptic reflexes involved in the flexion withdrawal reflex.3, 35, 39 SCI can interrupt the inhibition of these reflexes by supraspinal pathways, making them hyperexcitable.2, 3, 40 In other words, whereas flexor withdrawal reflexes occur normally in individuals without SCI, upon disruption of normal descending influences, the threshold for the flexor withdrawal reflex may become lowered, the gain of the system may become raised, or both may occur together.3 A recent study has provided evidence to implicate plateau potentials in the spinal interneuronal and motoneuronal circuitry in the hyperexcitability of flexion withdrawal reflexes in individuals with chronic SCI.41 Intrasegmental polysynaptic connections cause the flexor reflex initiated by a localized stimulus to generate a widespread flexor spasm, which can appear as a coordinated flexion of all joints of the leg.35, 39

spasticity can have a negative impact on quality of life through restricting activities of daily living (ADL), inhibiting effective walking and self-care, causing pain and fatigue, disturbing sleep, compromising safety, contributing to the development of contractures, pressure ulcers, infections, negative self-image, complicating the role of the caretaker, and impeding rehabilitation efforts.12-20 Spinal Cord (2005) 43, 577-586. doi:10.1038/sj.sc.3101757; published online 19 April 2005

But it has been suggested it also has beneficial effect as symptoms of spasticity may increase stability in sitting and standing, facilitate the performance of some ADL and transfers, increase muscle bulk and strength of spastic muscles (thereby helping prevent osteopenia), and increase venous return (possibly diminishing the incidence of deep vein thrombosis). This potential for a beneficial effect of spasticity on QOL has a large impact upon decisions regarding its management.11

Aetiology

Spasticity typically occurs in patients following:

1. stroke,
2. brain injury (trauma and other causes, e.g. anoxia, post-neurosurgery),
3. spinal cord injury,
4. multiple sclerosis and
5. cerebral palsy.
6. Other disabling neurological diseases.

Measurement of Spasticity:

Spasticity depends on several factors like presence of noxious stimuli, the patient's physical and mental status and the position of the body. Therefore it is difficult to measure spasticity because of its multifactorial nature. Different methods are available for measurement as measurement is essential to assess the response to treatment but none of them is precise and reliable enough to quantify the severity of spasticity clinically.

Ashworth Scale

This scale is based on the assessment of resistance to stretch when a limb is passively moved. It was

originally validated for patients with multiple sclerosis and was validated by Ashworth (1964). Its reliability is questioned by the subjectivity required by the observer to carry out the test and by the fact that it measures multiple aspects of limb stretch. However, it is in general use and has good inter-and intra-rater reliability (Ashworth 1964). The original Ashworth scale is only validated for measuring spasticity in the lower limb (Lee et al. 1989). In addition, it does not distinguish between increased neurogenic muscle tone and mechanical limb stiffness. The major modification (Modified Ashworth Scale) was proposed to differentiate between mild and moderate spasticity, as discrepancies appeared in clinical judgement at the lower end of the original scale. Bohannon validated the scale in elbow flexion in post-stroke patients and attempts have been made to widen the validity (Bohannon and Smith 1987). A grade 1+ was added and the top of the scale was reduced from 5 to 4.

Score	Ashworth (Ashworth 1964)	Modified Ashworth (Bohannon and Smith 1987)
0	No increase in tone	No increase in tone
1	Slight increase in tone giving a catch when the limb is moved in flexion / extension	Slight increase in tone giving a catch, release and minimal resistance at the end of range of motion (ROM) when the limb is moved in flexion / extension
1+		Slight increase in tone giving a catch, release and minimal resistance throughout the remainder (less than half) of ROM
2	More marked increase in tone, but the limb is easily moved through its full ROM	More marked increased in tone through most of the ROM, but limb is easily moved
3	Considerable increase in tone - passive movement difficult and ROM decreased	Considerable increase in tone - passive movement difficult
4	Limb rigid in flexion and extension	Limb rigid in flexion and extension

Tardieu Scale

The angle at the point of resistance is noted by stretching a limb passively. This is performed during as slow a movement as possible (V1), under gravitational pull (V2) and at a fast rate (V3). The examiner will feel a catch in a muscle under the influence of an overactive stretch reflex. Five levels have been described at the point of this catch to capture the quality of the muscular reaction. In essence the scale assesses dynamic and static muscle length as well as joint range of motion. The inter and intra-rater reliability is generally good (Gracies 2001).

Stretch Velocity

1. V1: Slow as possible
2. V2:Speed of limb falling under gravity
3. V3 Fast as possible

Y Angle (Dynamic Range of Motion)

1. R1 Fast Velocity: Movement through full range of motion.
2. R2 Slow Velocity: Passive joint range of motion or muscle length.

Quality of Muscle Reaction

Course of Passive Movement

0 No resistance

1. Slight resistance
2. Clear catch at precise angle, interrupting the passive movement,followed by release
3. Fatiguable clonus at precise angle,interrupting the passive movement,followed by release
4. Unfatiguable clonus at precise angle, (less than 10 sec when maintaining the pressure) occurring at a precise angle, followed by release.
5. Rigid limb & joint

Wartenberg Pendulum Test

In this, the leg moves under gravity and the observer measures the pendular activity of a spastic

limb as it relaxes. It is best carried out on the lower limb, for it is not so reliable for other limb segments.

Other methods for evaluating or assessing spasticity include:

1. Muscle grading,
2. Deep tendon reflexes
3. Range of Motion measuring,
4. Bilateral adductor tone score,
5. Visual analogue scale,
6. Spasm frequency score.
7. Torque devices
8. Electrophysiological studies (including dynamic multichannel EMG, tonic vibratory reflexes and electrical tests related to the H reflex and F wave).

Most of these methods are:

1. Time consuming,
2. Expensive,
3. Require specialised equipment and
4. Mainly used in research.

Treating Spasticity:

Spasticity can be disabling in itself and, if left untreated, may lead to consequences, such as:

- muscle shortening,
- contractures (leading to abnormal body segment loading and sensory change),
- limb deformity and altered body mechanics,
- altered body image,
- the need for special wheelchairs and seating and pressure-relieving equipment,
- loading on pressure points,
- pressure sores,
- difficulty in the management of pressure sores,
- pain from muscle spasms,
- degenerative joint disease,
- loss of function, and
- mood problems and inability to participate in rehabilitation.

Complications will prevent patients from achieving their optimal functioning and deconditioning from ill-health and pain will have a negative effect and patients and their carers may find reduced quality of life. Complications that may result due to spasticity are interference with function, nursing care and hygiene, pain, deformity and disfigurement, contractures, joint subluxation and dislocation, peripheral neuropathy and pressure ulcers. Although associated with complications, spasticity is beneficial to some patients. It may help to transfer, stand and ambulate, maintain muscle bulk, prevent deep vein thrombosis and osteoporosis.

Indications of Antispastic Treatment

Non-ambulatory patients who have moderate to severe weakness, hyperflexia, clonus and painful flexor spasms which interferes with their ADLS usually require treatment of spasticity. Patients may fulfill more than one indication, e.g. pain relief and care management.

1. To improve the functional improvement in terms of mobility by enchancing the speed and endurance of person in gait or wheelchair propulsion thereby improving transfers. It also improves hand functions in dexterity and in reachouts. It eases the person while performing sexual acts.
2. It helps in relieving pain and muscle spasms thereby keeping the legs in anticontracture postures enabling the person to wear the splints or orthosis.
3. It enhances the body image thereby improving posture.
4. It decreases the burden of caretakers interms of dressing, positioning the patients for feeding and in personal hygiene.
5. It helps in carryout the rehabilitative therapies thereby delaying or preventing surgery.

Principles of Management

The main goal of therapy is to increase functional capacity, relieve symptoms and decrease carer burden. This should be clear to the physician, the patient and the care giver. The consequence of reduction of spasticity should be assessed. If spasticity offers stability to a joint, its reduction may decrease the patients function. But, if there is minimal weakness with significant spasticity, treatment will result in considerable improvement in the patient's function.

Spasticity requires treatment when it is causing harm and this is the sole indication. Some patients early on after their stroke or brain injury are helped by their spasticity. For example, patients may start to support their weight by using their spastic lower limb when the degree of weakness in the leg would not allow it. Physical management (good nursing care, physiotherapy, occupational therapy) through postural management, exercise, stretching and strengthening of limbs, splinting and pain relief is the basis of spasticity management (British Society of Rehabilitation Medicine 1992). The aim of treatment is to reduce abnormal sensory inputs, in order to decrease excessive a-motor neuron activity (Ward 1999). All pharmacological interventions are adjunctive to a programme of physical intervention. Stretching plays an important part in physical management, but needs to be applied for several hours per day (Tardieu et al. 1998). Limb casting has been developed in this field to provide a prolonged stretch. Some studies have suggested that task-specific training might be more effective (Socialstyrelsen 2006).

Patient Assessment

Spasticity is a movement disorder and patients cannot be adequately assessed unless they are observed during movement and function. Physiotherapists and occupational therapists contribute to the observation and examination process, but some patients with complex movement patterns need assessing in a gait laboratory. The assessment process highlights the differences in patterns of limb posture and movement following an upper motor neuron lesion. Where there is no movement, the assessment process is fairly straightforward, but where there is loss of motor control rather than a spastic dystonia, one has to attempt to identify the different aspects of motor impairment. Patients with longstanding problems also develop compensatory movements, which may or may not require treatment and the clinician has to be clear about the underlying pathophysiological processes.

One can then identify how function is impaired and whether the problem is generalised, focal, or more regional. This will then point to the options for treatment. The indication for pharmacological treatment therefore is when spasticity is causing the patient harm. Some patients early on in their rehabilitation following a stroke or brain injury use their spasticity to walk on, when their weakness would otherwise not allow it. Clearly, treating the spasticity here would not be helpful and physical measures to utilise the developing movement patterns would be the treatment of choice, but where the spasticity gives rise to problems for either the patient or the carer, then treatment is required.

It is sometimes quite difficult to distinguish between severe spasticity and contracture formation, but it is important to do so. The clinicians and the patient/carer can then know what anti-spastic treatment can or cannot achieve and realistic expectations can then be identified. Severe, inadequately treated spasticity will go on to develop a limb contracture through shortening the muscle and tendons. A contracture may be fixed and will require serial splinting or surgery to correct it, but before it becomes fixed, the spasticity contributes to a dynamic contracture and treating the underlying spasticity may allow easier treatment of the contracture. One way to do that is examination under sedation. It is advisable to use a general anaesthetic for children. This relaxes spastic muscles and allows the range of passive joint movement to be assessed. One particular use is in assessing patients, who externally rotate their leg during walking. The adductor muscles can compensate for weak hip flexors and the patient rotates the leg accordingly. Blocking the obturator nerve reduces the function of the adductors and it is then possible to see the degree of hip flexor weakness, so that a programme of muscle strengthening can be started rather than of BTX injections to weaken the adductors.

Medical Management of spasticity:

Spasticity is a symptom and not a disease. Management of spasticity begins with the assessment of the underlying disorder.

Medical Management:

Treatment options:

A.Oral Drug Therapy: A number of drugs are available for the treatment of spasticity. These drugs reduce muscle tone and painful spasms, however their use is often limited by their side effects. The efficacy of oral antispastic drugs is small and evaluation of the effect on patient's quality of life is lacking from the available studies. 23

1. **Baclofen:** Baclofen is a GABA (?-aminobutyric acid) agonist and acts by inhibiting both monosynaptic and polysynaptic spinal cord reflexes.24The oral dose of baclofen used to treat spasticity ranges from 30-100 mg/d in

two to three divided doses. Oral baclofen may cause considerable side effects such as sedation, respiration problems and muscular weakness in higher doses. 25Baclofen must be tapered slowly to prevent withdrawal effects like increase in spasticity, fever, altered mental status, seizures, malignant hyperthermia.

2. **Benzodiazepines:** (Diazepam and clonazepam) Benzodiazepines act by increasing the affinity of GABA receptors for endogenous GABA.26Diazepam can be started at 5 mg at bedtime, and if daytime therapy is indicated, the dosage can be increased slowly to 60 mg / d in divided doses. Clonazepam can be started at 0.5 mg at night and slowly increased to a maximum of 20 mg / d in 3 divided doses. The side effect includes sedation, confusion, habituation and tachyphylaxis. 22

3. **Dantrolene:** Dantrolene is more useful for spasticity of cerebral origin. Dantrolene interferes directly with the excitation-coupling reaction. It acts at the level of the muscle fiber, affecting the release of calcium from the sarcoplasmic reticulum of skeletal muscle and thus reducing muscle contraction.]27 Dantrolene is given in a dose of 0.5-3.0 mg / kg / d. Dantrolene may cause side effects like muscle weakness and hepatotoxicity. 22

4. **Tizanidine:** Tizanidine decreases the excitability of α and γ motor neurons in the spinal cord by reducing the release of excitatory neurotransmitters in the spinal cord and decreasing the action of these excitatory neurotransmitters at their receptors. 28 Tizanidine also acts by inhibiting the release of substance P from small sensory afferent nerve fibers as well as slowing the firing of the locus ceruleus. 29 Tizanidine should be started at a low dose, 2-4 mg, preferably at bedtime. The average maintenance dosage of tizanidine is 18-24 mg / d. The maximum recommended dosage is 36 mg / d. The side effects of tizanidine are sedation, dizziness, dry mouth, and hypotension. Tizanidine does not cause any muscle weakness. 30 Therefore, it might be preferable over other antispasticity medications that do cause weakness, such as baclofen and dantrolene, especially in patients whose strength is already compromised by neurological disease.

2. Intrathecal Baclofen pump: The poor penetrataion of blood-brain barrier and significant side effects of oral baclofan can be minimized by intrathecal administration (directly into CSF) via a programmable pump. Considerably lower doses are required in intrathecal injection and it is without the development of tolerance. [11,12,13] It should be considered in patients unresponsive to oral pharmacotherapy and a severity of 3 on the Ashworth scale for at least 12 months. A test dose should first be given intrathecally before the pump is implanted. The dose range is 12-2000 mcg / d and should be fine tuned according to the severity of symptoms and response to therapy. The complications of intrathecal baclofen pump implantation are relatively few and usually are limited to mechanical failures of the pump or the catheter. Adverse drug effects are usually temporary and can be managed by reducing the rate of infusion.

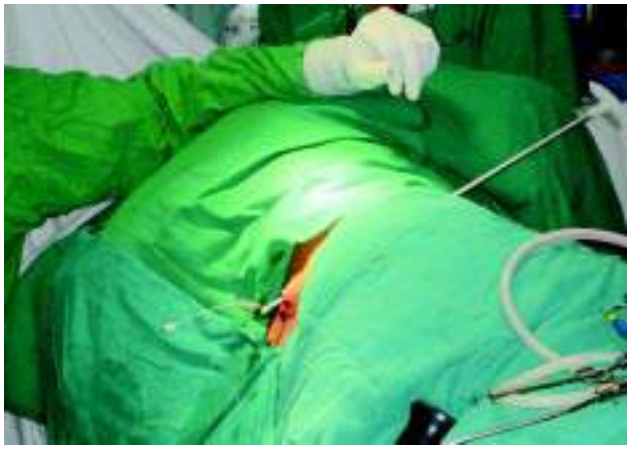


Figure 4.1.1 and 4.1.2

3. **Neurolysis with Neurotoxins: Botulinum toxin:** Botulinum toxin is a neurotoxin produced by the clostridium botulinum

bacterium. Botulinum toxin acts by causing reversible block of neuromuscular transmission by inhibiting acetylcholine release. 34There are seven different serotypes of botulinum toxins (A,B,C,D,E,F,G). Botulinum toxin is injected intramuscularly into the spastic muscles using a very fine needle. Injections should be targeted to spastic muscles responsible to functional loss using EMG guidance. 35 The effect of botulinum toxin starts within two weeks after injection. The clinical effect appears 4-7 days after injection, reaches a maximum after about 2 months thereafter the effect tapers off. The effect of botulinum toxin is not permanent and lasts for 3-4 months. After 3-4 months the effect gradually fades away and repeat treatment may be required depending on the symptoms / dysfunction caused by spasticity. [16] Botulinum toxin dosing has to be individualized and is dependent upon muscles involved, prior response, and functional goals. Botulinum toxin is used in patients with localized or multifocal spasticity. 37,38 American Academy of Neurology recommends botulinum toxin to reduce muscle tone and improve passive / active function. [19] Botulinum toxin therapy is approved for the treatment of cervical dystonia, strabismus, and blepharospasm in patients older than 12 years.

Antibodies may form against botulinum toxins and are a common cause of absence of any beneficial effect. Botulinum toxin injections are usually needed at 3- to 6-month intervals to maintain therapeutic benefit. Repeated, high-dose injections are likely to result in antibody formation. To decrease the possibility of antibody formation, repeat injections should not be given in less than 3 months. The smallest amount of botulinum toxin necessary to achieve therapeutic benefit should be used, and the interval between treatments should be extended as long as possible.

4. **Chemical neurolysis (Phenol or alcohol Injections):** A peripheral nerve innervating the spastic muscles is injected with phenol or alcohol solutions, which destroys myelin. It is used in patients with focal spasticity. The effectiveness of the injection diminishes over time and repeated injections are needed.

5. **Electrical Stimulation:** Electrical Stimulation of muscles, peripheral nerves and spinal cord has been used in the management of spasticity. Surface electrical stimulation of spastic muscles cause reduction of spasticity by stimulating cutaneous afferents and suppressing motoneuronal excitability. 40 Electrical stimulation of peripheral nerves reduces spasticity by inducing complete and reversible conduction block.41 Electrical stimulation of the dorsal columns of the spinal cord through epidurally placed electrodes may reduce spasticity. 42 Direct stimulation of spinal cord suppressess excitability of spinal motoneurons and cause reduction of spasticty.

6. **Transcranial magnetic stimulation:** Repetitive high-frequency (5 Hz) and low-frequency (1 Hz) transcranial magnetic stimulation may improve spasticity.

Surgical Management:

Surgery can play a very important role in the treatment of chronic spasticity or to allow more normal bone and muscle growth. Surgical treatment of spasticity involves neurosurgery and orthopaedic surgery. Neurosurgical treatment for spasticity is reserved for severe cases in which medical management has been ineffective or has lost its effectiveness. Neurosurgical procedures for spasticity include Selective Posterior (Dorsal) Rhizotomy (SDR), microsurgical DREZotomy, Peripheral Neurotomy, Longitudinal Myelotomy and Neurectomy.

Selective Posterior (Dorsal) Rhizotomy (SDR) involves cutting of the dorsal nerve roots that lie just outside the vertebral column and transmit nerve impulses to and from the spinal cord. SDR is primarily indicated in conditions exhibiting severe spasticity that interferes with mobility or positioning. 43 Microsurgical DREZotomy (Dorsal Root Entry Zone-otomy) is a type of selective rhizotomy. It involves cutting the nerve fibers at the entry zone and suppresses afferent discharges to the spinal cord. It is more effective in the treatment of severe spasticity limited to the upper or lower limbs. [24] Peripheral neurotomy involves cutting of peripheral nerves at the point at which they enter the muscle. It is indicated in the treatment of spastic neck, elbow, hand, hip, and foot. 45Longitudinal Myelotomy involves longitudinal division of the spinal cord to sever crossing sensory fibers and produce localized analgesia. It used to be performed earlier for severe and painful

spasticity. 46 Neurectomy involves the cutting of the nerve branches as they enter the targeted muscle. It is indicated in patients with focal spasticity refractory to botulinum toxin. 44

Orthopaedic surgerical procedures for the management of spasticity include muscle / tendon surgery and bone surgery. Muscle / tendon surgery involves lengthening or release of muscles and tendons. In contracture release surgery, contractured muscle is cut and the joint is then positioned at a more normal angle allowing regrowth of the tendon to this new length. Tendon transfer involves moving the attachment point of a spastic muscle to allow improved active function or retention of only passive function. Bone surgery can be osteotomy or arthrodesis. Osteotomy involves removal of a small wedge of bone to allow it to be repositioned or reshaped. Arthrodesis is fusion of bones that normally move independently and thereby limiting the spastic muscle to pull the joint into an abnormal position.

Physical Therapy Management:

Spasticity sets in gradually after the initial insult to the central nervous system. It becomes noticeable in the first few months, but the timing varies depending on the extent of underlying neurologic insult. Once recovery from the neurologic deficit stabilizes, the spasticity also tends to stabilize. Spasticity is not always detrimental as a weak flaccid limb can interfere with such daily activities as transferring, dressing, grooming and perineal care, spasticity provides posture and tone to a limb that can assist with weight bearing even if the patient cannot walk. However, excessive tone may interfere with these activities. Thus, it is only when spasticity interferes with function or puts the individual at risk of hurting himself or herself, it needs to be treated.

According to Lockley 2006, physical interventions helps in minimizing the viscoelastic properties of the connective tissue; altering the neural patterns of spasticity or spasms and maintaining levels of function for the individual.

The goal of physiotherapy in the management of spasticity should include :

1. Reduction of excessive tone.
2. Facilitate normal movement patterns.
3. To give a sense of normal position and normal movement.

1) Body Positioning:

It is advisable to inhibit the unwanted activity of the released reflex mechanisms by positioning the head and neck of the patient as they have tendency to elicit strong postural reflex mechanisms. Proper head and neck positions can inhibit these reflexes thereby

preventing the rest of the body from going into the reflex pattern. Sidelying with pillow support is most preferred position as it avoids stimulation of the tonic labyrinthine reflex and the asymmetrical tonic neck reflexes. Rhythmical trunk rotations can be easily given in both passive and active assisted exercises which helps in reduction of spasticity.

- Equipment that Improves Positioning

Controlling the position of a child's pelvis, knees, and ankles can help decrease arching, increased head control and shoulder stability, and improve functional hand use. Children may need different types of equipment at different times during their growth and development. Again, the goal is to increase control of movement and manipulation of objects in the environment. Some examples of equipment that can help improve positioning are:

1. Adapted strollers
2. Wheelchairs
3. Bath chairs
4. Adapted car seats
5. Adapted seating for general use
6. Standers
7. Gait Trainers

2) Rotatory Movements:

Trunk rotation produces lower limb to extend, abduct and externally rotate which is the position opposite to reflex pattern. Limb rotations are also very effective in normalizing the muscle tone of the patient.

3) Pressure over undersurface of Foot:

When the pressure is applied to the ball of the foot it stimulates an extensor reflex which is a pathological pattern of extension, adduction, and medial rotation of hip with plantar flexion of the foot, which is undesirable event in case of spasticity. When the pressure is applied under the heel of the foot, purposeful contraction of muscle occurs giving a normal supporting pattern. One should facilitate

normal Movements Patterns thereby avoiding triggering factors. Due to reflex release, some motoneurone pools are already in an excitatory state and any volitional effort will act as a trigger to those motoneurone pools resulting in muscle contraction in the spastic pattern. In such patients strong volitional effort should be discouraged as this facilitates the movement in the spastic pattern.

Other factors such as quick movements, abruptly performed, noisy surroundings, anxiety, excitement, over exertion should also be avoided as it may increase spasticity.

4) Slow Sustained Stretching:

Stretching forms the basis of spasticity treatment. Stretching helps to maintain the full range of motion of a joint, and helps prevent contracture, or permanent muscle shortening as it activates muscle spindles (Ia & II endings), golgi tendon organs (Ib endings) which are sensitive to length changes, inhibits or dampens muscle contraction and tone due largely to peripheral reflex effects. It is effective in extensor muscles than flexors due to the added effects of II inhibition. Stretching when done forcefully against severe spasticity, it increases hyperexcitable stretch reflex more strongly damaging periosteum of bone when excessive tension is applied to the tendons of the stretched muscles.

Techniques used to maintain slow sustanied stretching are as:

1. Manual contacts
2. Inhibitory casting or splinting
3. Reflex-inhibiting patterns
4. Mechanical low-load weights

5) Prolonged Cold Application:

Application of cold packs to spastic muscles (usually for 10 minutes or longer) may improve muscle tone. While the effect is not long lasting, it helps to improve function for a short period of time, or to ease pain by activating thermoreceptors thereby decreasing neural, muscle spindle firing leading to inhibition of muscle tone.

Techniques used

1. Immersion in cold water; ice chips
2. Ice towel wraps
3. Ice packs
4. Ice massage
5. Ice application with exercises

6) Neutral Warmth:

Retention of body heat stimulates thermoreceptors, autonomic nervous system mainly parasympathetics, which produces generalized inhibition of tone, calming effect, relaxation and decreases pain. It should be applied for about 10 to 20 minutes. Overheating should be avoided as it might increase arousal or tone.

Techniques used are as:

1. Wrapping body or body parts: ace wraps, towel wraps
2. Application of snug fitting clothing (gloves, socks, tights) or air splints

7) Relaxed Passive Movements:

Rhythmical, passive movements performed slowly throughout the range of motion in normal pattern may help in reducing spasticity.

8) Deep Rhythmical Massage (Tendon Rolling):

Deep rhythmical massage with pressure over the muscle insertions can be given to reduce spasticity.

9) Inhibitory Pressure (Weight-Bearing):

Prolonged pressure to long tendons inhibits the hypertonicity of a muscle as it activates muscle receptors (muscle spindles, golgi tendon organ) and tactile receptors. It can be applied manually or by body weight. Weight bearing postures provides inhibitory pressure, such as quadruped or kneeling postures can be used to promote inhibition of quadriceps and long finger flexors and sitting, with hands open, elbow extended, and upper extremity supporting body weight can be used to promote inhibition of long finger flexors.

10) Biofeedback:

Biofeedback is the use of an electrical monitor that creates a signal usually a sound as a spastic muscle relaxes. Thus, the person with spasticity may be able to train himself to reduce muscle tone consciously.

11) Functional Electrical Stimulation:

Electrical stimulation may be used to stimulate a weak muscle to oppose the activity of a stronger, spastic one thereby improving motor activity in agonistic muscles and reducing the tone in antagonistic muscles. Once stimulation has been stopped, the effect last for less than 1 hour probably because of neurotransmitter modulation within reflex arc. It improves standing, walking, and exercise training as well as decreases upper extremity contractures.

12) Tone Reducing Orthosis:

Orthotics are designed to help provide support to weak muscles and minimize the risk of joint deformity. There are a variety of orthotics made from a number of different materials. The goal is to use an orthotic which can give support depending on a child's pattern of movement, avoid skin breakdown, and be comfortable. If areas of the skin become red, this indicates that the orthotic may not be fitting appropriately, especially if the redness lasts more than half an hour after removing the orthotic.

1. Ankle foot orthoses (AFO's): These are plastic AFO's in which foot plate and broad upright are designed to modify reflex hypertonicity by applying constant pressure to the plantarflexors and invertors. Foot plate may be modified which maintains the toes in an extended or hyperextended position, thus assisting individual to walk with better foot and knee control thereby preventing the foot to go into equinovarus position.
2. Leg braces or casts
3. Hand splints
4. Soft body jackets

13) Serial Casts to Gain Range of Motion

Serial casts can either be used for the arms or the legs. The goal is to maintain or increase range of motion of a muscle, tendon, or joint. The casts provide a sustained stretch across the joint. Prolonged stretch can help muscles relax. The cast may also help "soften" tendons. Serial casts are normally changed one time per week. The number of total weeks of casting varies depending on the need of each child. The goal is to slowly gain more joint range without causing significant discomfort to the child.

14) Inhibitive Casts to Improve Function

Inhibitive casts are used to increase function more than to improve range of motion. Sometimes, inhibitive casts will be used to "give more information" (proprioception) to the ankle joint or the foot in order to prevent a child from using "reflex patterns" of movement. They are often utilized when a child with muscle imbalance is learning to walk. They can give better proprioceptive input to the foot when compared to the AFO since they are heavier and can have special features built in to them. For example, inhibitive foot plates can be built into the base of each cast. The inhibitive foot plates apply pressure to different areas of the foot to give better proprioceptive input to the joint which inhibits reflex patterns of movement. Often times, we will use inhibitive casts prior to prescribing orthotics, depending on a child's function.

15) Slow Maintained Vestibular Stimulation:

Low-intensity vestibular stimulation such as slow rocking produces generalized inhibition of tone as it facilitates primarily otolith organs (tonic receptors); less effects on semicircular canals (phasic receptors). Slow rolling movements, assisted rocking in a weight-bearing position or rocking with equipments like rocking chair, Swiss ball, equilibrium board, Hammock can help in reducing the tone.

16) Proprioceptive Neuromuscular Facilitation Techniques :

Proprioceptive neuromuscular facilitation is a neuromuscular treatment that uses repetitive stretches where one muscle is contracted while another muscle is relaxed at the same time. There are several techniques performed in proprioceptive neuromuscular facilitation which increases range of motion, improves range of stretch, increase strength and develop healthy muscle tissue.

Techniques used in reducing spasticity are as:

1. Rhythmic Initiation - Voluntary relaxation followed by passive movements through increments in range, followed by active movements progressing to resisted movements using tracking resistance to isotonic contractions.

2. Rhythmic Rotation - Voluntary relaxation combined with slow, passive, rhythmic rotation of the body or body part around a longitudinal axis, followed by passive movement into the antagonist range.
3. Contract Relax Active Contraction - Isotonic movement in rotation is performed followed by isometric hold of the range limiting muscles in the antagonist pattern against slowly increasing resistance followed by voluntary relaxation and active movement into the new range of the agonist pattern.

17) Manipulating Key Points:

Spasticity can be reduced by manipulating the key points in the upper limb and lower limb like the movements with thumb in abduction helps in reducing the spasticity in upper limb and in lower limb in sitting position, placing one hand over the lower back and other near the xiphoid process and moving the patient in the figure of 8 pattern forwards and backwards thus manipulating the pelvis helps in reducing spasticity.

18) Examples of different therapy models include:

1. Neurodevelopmental therapy (NDT),
2. Conductive education.
3. Manual therapy (myofascial release and craniosacral therapy).
4. Hippotherapy, or horseback riding, can help a child increase balance and improve trunk control.
5. Aquatic therapy can help promote more typical patterns of movement that are often affected by gravity.

19) Alternative Treatment

Alternative and complementary therapies include approaches that are considered to be outside the mainstream of traditional health care. Techniques that reduce stress, such as yoga, Tai Chi, meditation, deep breathing exercises, guided imagery, and relaxation training, may be helpful to induce relaxation and manage spasticity. Acupuncture and biofeedback training also may help induce relaxation.

OCCUPATIONAL THERAPY INTERVENTIONS IN SPASTICITY

Intervention for hypertonicity and spasticity is necessary when it interferes with ADL's, gait, sleep or wheelchair positioning or when it causes severe pain and limits hygiene (for eg.the client is unable to wash hand or axilla) or when spasticity leads to contractures or pressure sores50.

Spasticity, if not managed can lead to secondary problems such as 51:

- Deformity of the limbs, specially the distal upper limb (elbow to digits)
- Maceration of the palm tissue. Possible masking of underlying motor control.
- pain syndromes resulting from loss of normal kinematics
- impaired ability to manage BADL tasks (especially upper extremity dressing and bathing of the affected hand and axillawhen flexor positioning is present in a patient with stroke).
- loss of reciprocal arm swing during gait activities.

However before treating hypertonicity, the therapist should closely evaluate the function of the tone. Hypertonicity can have beneficial effects such as aiding him in standing and transfers, maintaining muscle bulk and prevent deep vein thrombosis, osteoporosis and oedema. 50

A uniformly acceptable, reliable and practical measure of spasticity continues to elude the therapists 52. Objective assessment of muscle tone in clients with hypertonia and spasticity is difficult. The postural reflex mechanism, the position of body and head in space, the position of head in relation to body and stereotypical reflexes and associated reactions all influence the degree and distribution of abnormal muscle tone. 53

Manual rating scale for hypertonicity and spasticity:

The Ashworth Scale 54 and the Modified Ashworth Scale 55 are 2 most widely used scales to manually rate spasticity 56, 57. There is a controversy in literature about the validity and reliability of these scales.

Brashear et al concluded that in upper limb spasticity, the Ashworth Scale had good intra and

inter - rater reliability when used by trained medical professionals 58.

Tardieu Scale: Modified Tardieu Scale 59 and the Tardieu Scale58 both measure spasticity. Modified Tardieu Scale has an interrater reliability coefficient of 7 and was shown to be more reliable than Modified Ashworth Scale 61.

Mild-Moderate-Severe Spasticity Scale and Preston's Hypertonicity Scale: Some therapists find it easier to use these scales.

MILD-MODERATE-SEVERE SPASTICITY SCALE:

Mild: The stretch reflex (palpable catch) occurs at the muscles end range (i.e. the muscle is in a lengthened position).

Moderate: The stretch reflex (palpable catch) occurs in mid range.

Severe: The stretch reflex (palpable catch) occurs when the muscle is in a shortened range.

PRESTON'S HYPERTONICITY SCALE:

0: No abnormal tone detected during slow passive movement.

1. **Mild:** First tone or resistance is felt when the muscle is in a lengthened position during slow passive movement.
2. **Moderate:** First tone or resistance is felt in the mid range of the muscle during slow passive movement.
3. **Severe:** First tone or resistance occurs when the muscle is in the shortened range during slow and passive movement.

MECHANICAL AND COMPUTER RATING SYSTEMS FOR HYPERTONIA AND SPASTICITY

Mechanically determined parameters are more reliable than the manual methods. McCrea et al, concluded that using a linear spring damper model to assess the hypertonic elbow was reliable and valid 62. However it is time consuming and certain muscle groups are difficult to assess with it.

Leonard et al investigated the construct validity of myotonometer, a newly developed computerized, electronic device. The authors concluded that the myotonometer could provide objective data about the tone reducing efficacy of various tone reducing procedures 63.

In the past, sensorimotor approaches were used to treat patient's with abnormal skeletal muscle activity. These approaches were developed by Rood, Bobath, Knorr and Voss (PNF) and Brunnstorm and are based on an understanding of CNS dysfunction. Although these interventions are commonly used, their effectiveness is being challenged as occupational therapist move towards models of evidence-based practice64.

For years, occupational therapists have been using weight bearing activities to reduce hyper-tonicity and to remediate paresis in patients with upper motor neuron lesions. At present there is a limited amount of research to support these neurofacilitation approaches. Brouwer and Ambury concluded that cortico-spinal facilitation of motor units occurred during weight bearing. They believed that afferent input from weight bearing increased motor cortical excitability65.

Chakerain and Larson studied the effects of upper extremity weight bearing on hand opening and prehension in children with spastic cerebral palsy. Computer calculation of the patients hand surface area were measured and it was found that there was an increase in surface area after weight bearing and an increase in the maturity of movement components needed for prehension.66

McIllroy and Maki demonstrated that if the affected arm is used when weight bearing, postural responses occur throughout the weight bearing extremity and occur during other perturbations of Posture.67Although, not many studies have documented how and why weight bearing works physiologically, it certainly is a requirement for improving performance 50

An important occupational therapy objective is to have client manage muscle tone to engage in and complete basic and instrumental activities of daily living. Positioning and movement in patterns opposite to hypertonic or synergistic patterns are important to expand the motor repertoire and develop movement that is as close to normal as possible.

The client should also be taught how to incorporate the affected extremity as much as possible into all ADLs. ADLs, crafts, games, and work activities can be used to teach incorporation of the extremities for a total approach to treatment 68

Casting in inhibitive postures has been shown to be effective in tone reduction 69,70. The beneficial effect of casting on hypertonia is well documented in literature71,72,73. Serial casting is effective in presence of a contracture and its use should cease when desired position is achieved and tone is manageable using a splint.

Multiple commercially available spasticity reducing splints are used to keep hand and wrists in inhibitive postures. The client and family need to be educated in continuing to incorporate the extremities in occupations and to bear weight on extremities as much as possible; to retain the ROM gain achieved during casting 74

Physical agent modality such as cold, superficial heat, ultrasound and Neuromuscular Electric Stimulation can be used as a preparation for or in conjunction with purposeful activity and muscle reeducation, provided the therapist has the appropriate training62.

The Functional Tone management (FTM) Arm Training Program is based on distal activation model focusing on the key points of early initiation of upper extremeity movements that incorporate grasp and release. In order to incorporate the hand into FTM arm training, a dynamic orthosis for the hand called the SaeboFlex is used. This orthosis assists an individual who exhibits hypertonia in the hand to place the hand in an open functional position and also to produce a graded muscle contraction of the finger flexors in order to grasp an object. The FTM program combines high repetition grasp and release with task specific arm training drills to progress the client towards a functional goal. A significant body of research support the FTM arm training program 26, 27, 28. Clinically observed improvements with FTM arm training Program include increased AROM at the shoulder, elbow and wrist, improved upper extremity Fugal- Meyers Scores and decreased modified Ashworth Scores78.

In conclusion, the impact of spasticity can be far-reaching on an individual's lifestyle and that of family and carers. Health care professionals can provide vital care support to individuals and their families and refer them appropriately towards other professionals and organizations for required intervention and support.

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Ch.13 Orthosis

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LOWER LIMB ORTHOSIS

A lower limb orthosis is an external device applied or attached to a lower extremity to improve function by controlling motion, providing support, reducing pain through transferring of the load to another area, correcting flexible deformities, preventing the deformities getting worsened.

Orthosis is the medical terminology for brace or splint.

- **AFO:** Ankle Foot Orthosis.
- **KAFO:** Knee Ankle Foot Orthosis.
- **HKAFO:** Hip Knee Ankle Foot Orthosis.
- **THKAFO:** Trunk Hip Knee Foot Orthosis.

Orthosis are of two types :

1. **Static:** Support the weakened joint.
2. **Dynamic:** Allows joints movements during locomotion.

Materials used are of:

1. Metal
2. Plastic
3. Leather
4. Synthetic fabrics.

Plastic materials such as thermosetting materials / thermoplastic are commonly used.

They can be easily moulded after heating and hardened after cooling.

Low temperature thermoplastics can be fabricated easily.

Principles:

In all orthosis :

1. **3 point pressure** is essential.
2. Orthotic joints should be at the **anatomical joint level**
3. Should be **light weight, strong, durable** and **cosmetically acceptable.**
4. **Static or dynamic stabilization.**
5. **Flexible material.** Tissue tolerance to compressive and shear force.
6. **Thermoplastics (polypropylene)** requires high temperature (150 degrees Celsius) for molding and it is used mainly for high stress activities.
7. **Rubber is used for padding**
8. **Metals** are used for joint components and uprights.

External Shoe Modifications:

1. **Cushion heel:** to minimize impact on heel, encourage plantarflexion and minimize knee flexion.
2. **Heel wedge:** a) **Medial wedge** to promote inversion (for flat feet).
   b) **Lateral wedge** to promote eversion (for varus foot)
3. **Crooked and elongated heels:** Medially (for flat feet) Reverse crooked and elongated heels (C / E Heels) i.e. elongated lateral part of heel (for varus foot)
4. **Heel elevation:** to compensate fixed equinus deformity or length discrepancy.
5. **Compensation platform:** for length discrepancy

Sole Modifications

1. **Metatarsal bar** at posterior metatarsal head to shift the weight bearing on metatarsal heads, commonly used in Diabetes Mellitus and anesthetic foot.
2. **Sole wedge : Medial wedge** to promote supination of the foot

   Lateral wedge to promote pronation.

Internal Shoe Modifications:

1. **Cushion Heel:** Soft pad with scooped out central part under the painful points.
2. **Heel wedge: When given medially**, can promote inversion of hind foot.

   When given laterally, can promote eversion of hind foot.
3. **Metatarsal pad:** shifts stress from metatarsal head to meta tarsal shaft.

4. **Innersole excavation:** at one or more Metatarsal heads.
5. **Valgus pad or medial arch support.** (for flat feet)

Foot Orthosis:

1. **Plastic insert:** This is a rigid plastic insole for manual correction. It has rigid medial, lateral or posterior walls.
2. **Heel Cup:** Plastic cup under the heel with rigid wall posterior and medial and lateral aspect of the heel (for pronated heel)

Custom made orthosis are always better than ready made devices.

Ankle Foot Orthosis (AFO):

- **Prescribed for weakness** of 1) ankle dorsiflexors 2) plantarflexors 3) inverters 4) everters.
- They are **used** for maintaining the feet in the neutral position while walking.
- They are also used to **prevent** deformities.
- In a case with an UMN weakness of the foot with spasticity in the inverters and plantarflexiors, which would give an equino-varus tendency of the foot, it is prescribed to maintain the foot in corrected position and also prevent foot drop. This would help in the gait training
- The position of foot affects the stability of knee indirectly.
    1) Equinus foot with spasticity could lead to hyper extension of knee.
    2) Calcaneal foot may lead to knee flexion.
    3) varus foot may cause Genu varum
    4) valgus foot could lead to Genu valgum
- An AFO can reduce energy consumption in Diplegics, Hemiplegics and in LMN foot drop.

TYPES OF AFO

1. **Fixed Ankle:** a) Plastic insert
   b) High boots with steel shank.
2. **Hinged AFO:** Adjustable ankle hinges with desired ankle plantarflexion or dorsiflexion.
3. Conventional below knee braces with ankle or without ankle joints, along with medial T-strape (for valgus foot) or lateral T-strape (for varus foot)

High boots with steel shank

Plastic AFO

Plastic AFO

KAFO (Knee Ankle Foot Orthosis)

KAFO is prescribed whenever there is an inability to lock the knees due to weak muscles around the knees (Quadriceps and Hamstrings). There is a tendency for buckling of the knee.

Types of KAFO

1. With Knee Joint
2. Without Knee Joint
3. Ischial Weight Bearing
4. With Free Knee Joint
    - Knee Joint is provided for the patient to wear a brace and fold the knee.
    - Ischial Weight Bearing KAFO is prescribed when the weight bearing has to be on the ischium (in cases of fractures of the weight bearing bones like femur or tibia)
    - KAFO with free knee joint is prescribed whenever there is strong quadriceps but

the knee joint has tendency for hyperextension.

Many a times, trial splints like **Push Knee Splints** are prescribed to see whether the patient can manage locking the knee and walking. (Provided the foot has got proper control)

Knee Ankle Foot Orthosis

Push Knee Splints

Hip Knee Ankle Foot Orthosis (HKAFO)

HKAFO is prescribed whenever there is a lack of control of hip and knee. A pelvic bandis provided in order to give Hip stability. Hip is controlled mainly by hip extensors and abductors,in the absence of which, the hip becomes wobbly and with bilateral involvement walking is very unstable. Thus patient may require use of a pair of crutches or a walker also.

Hip Knee Ankle Foot Orthosis

SPINAL ORTHOSIS

Spinal Orthosis are prescribed for

1. Unstable Spine due to weak musculature
2. Unhealed Fractures of the spine
3. Correction of deformities like Scoliosis, Kyphosis, etc.
4. Spinal Dysraphysm e.g. Meningomyocoele, Spina Bifida
5. Post surgical conditions like Laminectomy

Types of spinal orthosis

1. Lumbosacral belt or corset
2. Thoracolumbar Corset
3. Close Contact Spinal Orthosis
4. Milwaukee Brace
5. Taylor's Brace
6. Minerva Jacket
7. Anterior Hyperextension Brace

By far, a close contact spinal jackets can give the best support for scoliosis and other deformed spine.

Close Contact Spinal Orthosis

Lumbosacral belt or corset

Anterior Hyperextension Brace

Taylor's Brace

CONCLUSION:

Various types of orthotic devices are useful for preventing deformities getting worse, maintaining corrected position of the lower extremities and also improving the gait pattern as much as possible. They compensate for the weak muscles for stability. Prescribing the correct orthotic devices is one of the most essential part of Rehabilitation, especially in Neurological conditions

UPPER LIMB ORTHOSIS

Definition

An orthosis is a device that is applied to the body in order to protect and stabilize body parts, to prevent or correct scarring and deformities, or to aid in performance of certain functions. Upper limb orthoses are applied to the shoulder, elbow, arm, wrist, or hand. These devices may be called orthoses, orthotic devices, or splints.

Upper extremity orthoses are used frequently on patients who have

- musculoskeletal problems like those resulting from trauma, sports, and work-related injuries.
- neurologic problems, such as stroke, traumatic brain injury(TBI), multiple sclerosis (MS), cerebral palsy (CP), spinal cord injury (SCI), and peripheral nerve injury.
- arthritic conditions

Purpose of Splinting

Upper limb orthoses can be used for a wide variety of purposes. Some of the more common uses include:

- stabilizing fractures or unstable joints
- immobilizing joints to promote healing
- preventing or correcting joint contractures
- Protect weak muscles from overstretch
- correcting subluxation of joints or improper alignment of tendons
- Helping to provide enhanced function
- Serving as an attachment for assistive devices
- Blocking unwanted movement of a joint
- maintaining correct joint alignment
- assisting movement of joints
- reducing muscle tone in spastic muscles
- To reduce weight bearing forces for a particular purpose
- To otherwise correct the shape and/or function of the body, to provide easier movement capability or reduce pain

Team Approach

Creating and employing upper limb orthoses often involves a team approach, especially in rehabilitation settings. Custom-made upper extremity orthoses may be fabricated by physical, occupational, and hand therapists. The therapist may help the patient learn to use the orthosis but can also recommend ready-made devices or refer the patient to an orthotist.

Health care professionals who create and fit upper limb orthoses must have a good understanding of the anatomy and physiology of the upper limbs. They must also understand the mechanics and forces involved in making various body movements, and they must be familiar with the materials and tools involved in constructing orthoses. Certified orthotists are specialists who focus exclusively on fitting and building orthoses.

The professional relationship between the physical/ occupational/ hand therapist and orthotist is very important in identifying patient functional goals and a variety of predictable outcome.

Basic Principles in Splinting:

I. **Mechanical Principles:** Use of mechanical principles helps make splints comfortable, durable, effective and diminishes of additional injury secondary to splint application.

Mechanically, splints may be grouped into two categories:

1. **Those that apply three point pressure:** Three point pressure splints function through a series of reciprocal forces with the middle force directed opposite to the two end forces.
2. **Those that pull adjacent articulated bony segments together** through circumferential pressure. In the circumferential pressure splint, the middle reciprocal force is absent.

Pressure and shear can be reduced by increasing the surface area of the splint. Short narrow splints or components are often problematic since they apply pressure to a small area creating pressure necrosis of underlying soft tissues. Longer splint

designs increase mechanical advantage and make splints less susceptible to causing pressure problems. (Willard)

Other Mechanical principles that need to be taken into consideration are

1. Use Optimal Rotational Force

- **Rotational element-** To produce joint rotation
- **Translational element-** To produce joint distraction or compression

2. Consider the Torque Effect

- Increase torque by increasing the distance between the joint axis and the point of attachment of the dynamic assist

3. Consider the **Relative Degree** of **Passive Mobility of Successive Joints**

4. Consider the **Effects of the Reciprocal Parallel Forces**

- Use first -class lever system
- If Mechanical Advantage is increased, the middle opposing force is decreased

5. Provide Contours

- Eliminate Frictions by proper padding, forces, stockinet, etc

II. General Principles of Design

1. Consider individual patient factors such as age, intelligence, motivation, body size, activity level, socio economic status etc.
2. Consider the length of time the splint in to be used
   - Shorter the anticipated need of splint, simpler its design, material type and construction should be.
3. Strive for simplicity and pleasing appearance
4. Allow for optimum function of the extremity
   - Avoid needless immobility of normal joints
5. Allow for optimum sensation
   - Palmar tactile surface areas should be free
6. Allow for quick, low cost and efficient construction and fit.
7. Provide for ease of application and removal
   - Avoid dependence on others for assistance
8. Design for several functions into one splint to avoid confusion and simplify the wearing and exercise routines.

III. Specific Principles of Design

1. Identify the key joints to be splinted by careful evaluation.
2. Review purpose to immobilize, to increase passive motion or to substitute for active motion by re-evaluating for functional objective of a given splint.
3. Decide whether to employ Static or Dynamic forces.
4. The surface of the extremity on which the splint will be based is determined and secondary joints that need to be controlled or positioned are noted.
5. Identify area of diminished sensibility to avoid pressure necrosis.
6. Consider kinetic effects: Since application of the splint often alters internal and external forces to proximal or distal joints, the kinetic effects of the splint must be considered including what will be the forces on un-splinted joints and what will be the ramifications to extrinsic and intrinsic musculo-tendinous structures.
7. Choose the most appropriate materials and adapt to general properties of the selected splint materials.

IV. Principles of Construction

1. Use fabricating equipment appropriate to material.
2. Use type of heat and temperature appropriate to the material
   - Over heating or under heating can disrupt the material.
3. Strive for good cosmetic effect.
4. Round corners and smoothen edges
   - For increased strength, durability, cosmesis and comfort
5. Stabilize Joined Surfaces
   - By using one or two rivets as per the requirement

6. Rivets should be flush with the material surface or apply tape / moleskin over internal metal rivet to prevent rusting.
7. Provide small ventilation holes to improve air circulation
8. Perforation should be made from inside out to give a smoother inner surface
9. Secure Padding
   - Allow ease of splint application and removal
   - Apply extra padding to the edges, to allow curl around splint edges
10. Secure straps to provide splint stability by gluing or riveting and the strap ends should be rounded and smoothed to prevent fraying.

V. Principles of Fit

a. Mechanical Considerations

1. Use Principles of mechanics
   - The mobilizing force should always be directed perpendicular to both t2he segment being moved and to the rotational axis of the joint
   - The leverage system applied (to the placement of straps, finger cuffs and fingernail hooks) should help in dissemination of the applied force and reduce pressure and elimination of friction.

b. Anatomic Considerations

Contiguous fit of the splint to the extremity reduces pressure on bony prominences as well as soft tissue.

1. **Bone**
   - Accommodate bony prominences
   - To avoid tissue necrosis
   - Incorporate Dual Obliquity concepts
   - The progressive decrease of length of the metacarpals from the radial to ulnar aspects of the hand
   - The immobility of the second and third metacarpals as compared to the mobile first, fourth, and fifth metacarpals
2. **Ligaments**
   - Consider ligament stress
   - Preserve the ligament structure in correct position and tension to prevent inflammation, attenuation and disruption of ligament tissue
3. **Arches**
   - Maintain the skeletal arches (proximal transverse, distal transverse and longitudinal arches)
4. **Joints**
   - Align splint axis with anatomic axis
   - Use optimum rotational force in 90 degree angle of pull
5. **Skin**
   - Use skin creases as boundaries

Kinesiologic Considerations

1. Allow for Kinematic changes when fingers and thumb are in flexion or in extension
2. Employ kinetic concepts

Technical Considerations

1. Develop patient rapport and trust
   - Give instructions and explanations regarding the importance and function of the splint
2. Work efficiently
3. Change method according to properties of material used

Prefabricated or Custom fabricated Orthoses

Prefabricated orthoses are easily available in a cost effective and timely manner. As a general rule, prefabricated orthoses should be considered for patients having a normal anatomy, and or who will require the orthosis for a short period of time. Though these prefabricated orthoses are easy to apply, the therapists need to understand the indications, contra indications and limitations of these devices.

Custom made orthoses are generally used when orthotic device is required for extremities or when the spine have deformities or in case of unusual sizes and / or when they have to be used indefinitely

The **advantages** of custom fabricated orthotics are:

- Designed to patient specification
- easily adjusted if uncomfortable

- Any position requested can be achieved with the low-temperature plastic
- Able to clean as needed
- Can get wet
- Adjustable as swelling decreases or as able to get into a better position

Materials and construction

Although ready-made orthoses are available for some applications, many are custom made to fit the specific needs of each patient. The material used in orthotic devices typically includes low-temperature thermoplastics that are readily shaped for fit and function. Other materials used include casting, metal, straps, and hook-and-loop. Most orthoses have employed lightweight thermoplastic materials, which are plastics that become pliable when they are heated and retain their shape once they cool. The thermoplastic sheets can be molded to fit body parts exactly, and some can be reshaped repeatedly as the treated body part changes shape. The resulting orthotic device is lightweight and relatively easy to use and maintain.

Thermoplastic materials are usually classified into high- and low-temperature types, based on the temperature at which they become pliable. High-temperature thermoplastic materials must be molded at a temperature that is too high to come in contact with human skin. These materials must be molded over a plaster model of the body part, but have the advantage of being stronger and more durable than low-temperature thermoplastics. They are used in situations where the orthosis will undergo a lot of stress or will be used for a long time. High-temperature thermoplastics require special tools for cutting and shaping, and orthoses made from these materials are usually constructed by an orthotist, a technician who specializes in making these devices.

Many upper limb orthoses are constructed of low-temperature thermoplastics. These material becomes pliable below 180°F (80°C), and can be molded directly against the body. It is relatively easy to cut and shape, and many therapists construct orthoses using these materials. Precut shells made from low-temperature thermoplastics are also available. The therapist can use a precut thermoplastic shell as the base for a device and then modify it to fit by trimming and adding pads and straps. Orthoses made from low-temperature thermoplastics are commonly used in situations in which the orthosis will receive relatively little stress or is intended for temporary use. These orthoses are especially important when a device is needed quickly, such as in postsurgical or trauma treatment.

Both high- and low-temperature orthoses must be attached to the body. Most modern orthoses use straps made of hook-and-loop tape for this purpose. This material is lightweight, durable, and readily adjustable, and comes in a variety of widths and colors. Orthoses can also include padding to cushion sensitive areas, as well as specialized linings. Patients often use a separate interface that absorbs perspiration and protects the skin, and which can be washed or replaced as needed.(7)

Types of orthoses

The upper limbs comprise a complex system of muscles, joints, ligaments, and tendons, which are capable of a number of distinct movements. For this reason, a wide variety of upper limb orthoses have come into existence. These devices often go by multiple names, reflecting the name of the manufacturer, the name of the person who developed the device, or the anatomy and function it serves.

In the 1970s, a group of American professionals involved in the field of orthotics-put forth an effort to classify orthoses by their function and acronyms describing the joints that are encumbered by the orthoses. From this effort sprung the current nomenclature : AFO- ankle foot orthosis, TLSO- thoracolumbosacral orthosis, WHO- wrist hand orthosis, etc. However, the nomenclature based on the function of the orthosis; such as assist dorsiflexion at ankle, limit wrist flexion to 10 degrees, resist thoracolumbar rotation has not found wide spread acceptance.

Orthoses are usually classified under the following three categories:

- static,
- serial progressive
- dynamic or functional

Static orthoses hold a body part in a fixed position and do not allow joint movement. Some static orthoses do not contain joints, as with fracture orthoses that stabilize the long bones of the arm after a fracture. Most others simply maintain the joint at a particular angle, providing support and proper positioning. For example, a static wrist

orthosis can be used to hold the wrist in a neutral position to promote healing and prevent injury during activities. Sometimes static orthoses include attachments that help patients perform functional activities. For example, a hand-wrist orthosis may include an attachment for pens or eating utensils. Static orthoses sometimes serve the function of promoting eventual joint movement.

Complications associated with use of static orthoses

1. Skin breakdown
2. Contractures
3. Infection

Serial or progressive orthoses loosen joints that have become frozen due to contractures or arthritis. Serial orthoses involve several similar devices used in a series, with each successive device gradually increasing the range of motion of the affected joint by providing a gentle stretching action.

Progressive orthoses accomplish similar goals, but do so by allowing adjustments in the device so that it gradually increases the amount of stretch created in the joint.

Serial and progressive orthoses must be designed and used carefully to provide the correct amount of stretching in the joint. Excessive stretching can damage the tissues, and inadequate stretching will be ineffective.

Dynamic/functional orthoses allow or create joint movement. These devices hold the joint in the proper position while assisting movement using springs, rubber bands, or other mechanical features. Dynamic orthoses are useful for patients who have weakened muscles or limited neuromuscular control, because they allow the patient to perform actions that would be difficult or impossible without assistance. These devices promote independence in patients who have handicapping conditions, and they are common in rehabilitation settings. Since no single device can perform all the movements that the human hand can perform, the patient may need to use several different dynamic devices in order to carry out activities of daily living. Some dynamic splints have a dual or bilateral tension-providing mechanism that can safely accommodate moments of spasm and thus potentially limit or prevent soft-tissue injuries.

One study indicated that patients who have sustained a stroke can be aided by a gravity-compensating arm orthosis that helps to loosen the grip of the affected hand and help stroke patients gain greater functional independence.

However, a systematic review that examined 4 trials including 126 patients with stroke or other nonprogressive brain lesions found that upper limb orthoses showed no effect on upper limb function; ROM at the wrist, fingers, or thumb; or pain.

Types of upper-limb orthoses

- Upper-arm orthoses
- Clavicular and shoulder orthoses
- Arm orthoses
- Functional arm orthoses
- Elbow orthoses
- Forearm-wrist orthoses
- Forearm-wrist-thumb orthoses
- Forearm-wrist-hand orthoses
- Hand orthoses
- Upper-extremity orthoses (with special functions)

Clavicular and shoulder orthoses include the following:

- **Figure-8 harness/clavicular brace** - This is used to restrict motion in patients with clavicular fractures so as to allow tissue healing and bone remodeling
- **Shoulder sling** - This is used to restrict motion in subluxated shoulders by providing humeral cuff and chest straps to keep the humeral head in the glenoid cavity
- **Overhead sling suspension** - This is used for patients with proximal arm weakness or paralysis to allow hand or arm use when the muscles are at least antigravity in strength
- **Hemiplegic arm sling** - This is used for immobilization of the hemiplegic shoulder, which helps to decrease pain and subluxation (see the image below)

Functional arm orthosis are used primarily in patients with proximal arm weakness involving the shoulder and arm,[7] such as that resulting from spinal cord injury (SCI) or peripheral nerve lesions. They include a shoulder saddle from which a proximal forearm cuff is suspended by means of straps or a Bowden cable.

Elbow orthoses include the following:

- **Posterior elbow splint -** This is used particularly for elbow immobilization in patients
- **Serial cast -** This is used for preventing or correcting contractures through promotion of soft-tissue stretch and passive range of motion (ROM)
- **Air splint -** This is a form of circumferential inflatable sleeve, used to maintain or increase elbow extension and also used for contractures and elbow immobilization
- **Dynamic elbow splints**, static progressive elbow splints, turnbuckle elbow splints-gently elongate the soft tissues over a long period to attempt to reverse joint malalignment (contractures, burns, and late phase of fracture). They are not used in spastic muscles as they may further increase tone.
- **Dorsal elbow-extension mobilization orthosis -** extend the elbow as well as provide mediolateral elbow stability and rotational forearm stability
- **Dorsal elbow - flexion mobilization orthosis -** flex the elbow and provide mediolateral stability and rotational forearm stability

Forearm orthoses

Balanced forearm orthosis (BFO) - This supports the weight of the forearm and arm against gravity and is used primarily in patients with high-level tetraplegia or severe proximal arm weakness or paralysis[7] ; it may be attached to a wheelchair or table; patients may be able to perform tabletop activities; prerequisites for its use include a power source, such as neck or trunk muscles (to shift the trunk center of gravity) or adequate scapular movement

Requirements

- Some residual muscle strength (MMT at least poor or grade 2) and coordination of elbow flexion (can be used for C5 quad)
- Adequate trunk stability and balance
- Adequate endurance in a sitting position
- Preserved ROM of the shoulder and elbow joints

Other uses: they also may be used in spastic patients to allow them to self-feed by dampening muscle tone through a friction device

Forearm mobilization (corrective) orthosis- dynamic supination / pronation splints used to increase supination or pronation in forearm rotational contracture, or to increase passive or active-assisted ROM in spinal cord injury

Shoulder-elbow-wrist-hand orthosis (SEWHO) consists of a forearm trough (attached by a hinge joint to a ball-bearing swivel mechanism) and a mount (which can be mounted on the WC, on a table or working surface, or onto the body jacket). It helps support the forearm and arm against gravity and allows patients with weak shoulder and elbow muscles to move the arm horizontally and flex the elbow to bring the hand to the mouth (e.g., patients with spinal cord injury, Guillain-Barré Syndrome, polio, muscular dystrophy, and brachial plexus injury).

Forearm-Wrist Orthoses: Forearm-wrist orthoses may be either volar or dorsal and either gutter-based or circumferential. Ideally, the wrist should be positioned in 15-30° of dorsiflexion (wrist extension), except in carpal tunnel syndrome. The volar type should allow for metacarpophalangeal (MP) flexion by ending before the distal palmar crease.

Examples of forearm-wrist orthoses include the following:

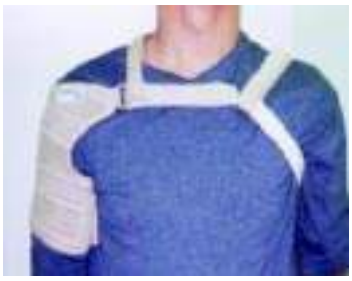
- Wrist cock-up splint
- Wrist extension splint
- Ulnar gutter splint

Volar wrist-hand stabilizers/wrist cock-up splint

- **Wrist-hand orthosis (WHO)** extending from distal two-thirds inch of forearm to about one-quarter proximal to the distal palmar crease to allow full metacarpophalangeal flexion while maintaining the functional position of the wrist and hand
- Uses include resting wrist and hand in acute arthritis (RA), wrist sprain / contusion, flexor / extensor tendinitis, carpal tunnel syndrome, postsurgical wrist extensor tendon repair, wrist fusion, and skin grafting, contractures prevention, reduce pain, reduce spasticity, prevent ulnar / radial deviation wrist / hand (e.g., RA)

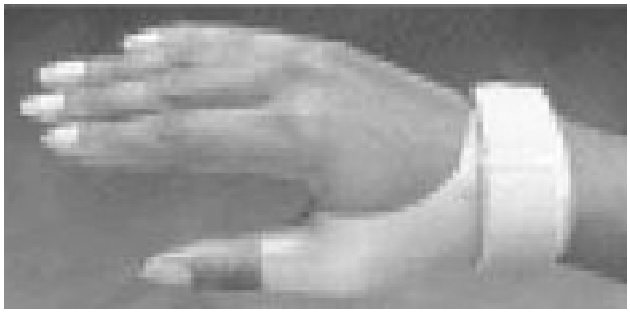
Dorsal wrist-hand stabilizers

- WHO are used to provide the same functions

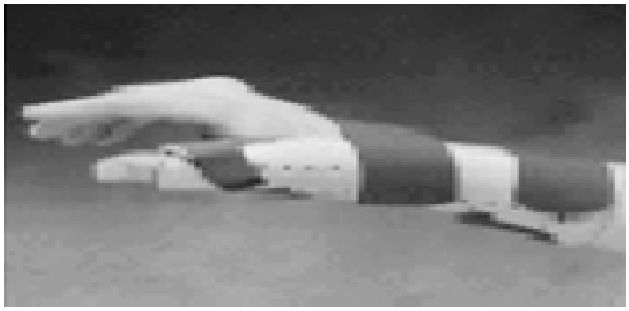


Hemiplegic arm cuff / sling

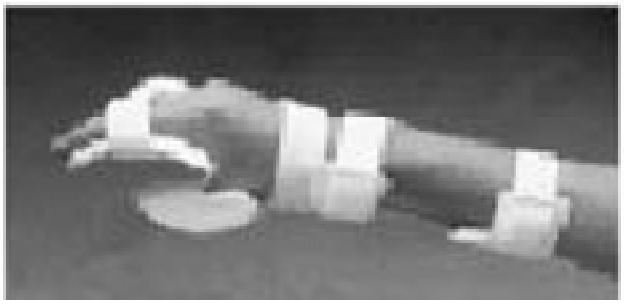
Resting hand splint



Basic Opponens Splint



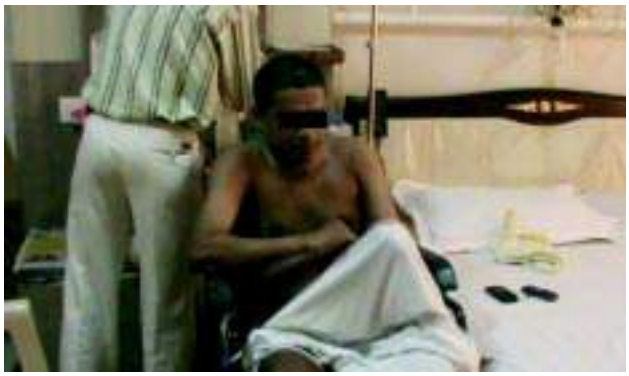
Long Opponens Splint



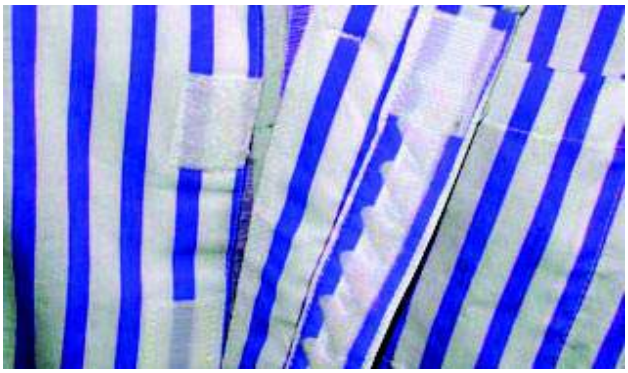
'C' bar splint

Ulnar deviation correction splint

(A) Antispasticity "Ball" splint (B) Hand Cone splint



Universal Cuff



Patient Using U-cuff for brushing teeth

of the volar WHO as well as greater stabilization because of rigid dorsal hand section

- More difficult to fabricate and fit than the volar WHO

Forearm-Wrist-Hand Orthoses: The basic types of forearm-wrist-hand orthoses are the resting hand splint (see the image below), the functional resting splint, and the static hand splint. Other types include the burn splint and the weight-bearing splint.

Fig 2

Placement may be dorsal, volar, or circumferential and extends from the tips of the fingers to cover two thirds of the distal forearm. The dorsal type of splint is particularly useful in patients who demonstrate palmar hypersensitivity and grasp reflex. The position of the wrist is neutral or in slight dorsiflexion. Immobilization in this position preferred because metacarpophalangeal joint and Interphalangeal collateral ligaments are kept stretched, minimizing future joint capsule contractures. In addition, it provides functional thumb position for opposition and three-jaw chuck pinch.

Functions of forearm-wrist-hand orthoses include the following:

- Immobilization in patients who have hand flexor and extensor tendinitis or who are undergoing tendon, nerve, or fracture repair.
- Maintenance of passive range of motion (ROM) in patients with upper motor neuron lesions and contractures

Wrist Orthoses

1. **Wrist control orthosis**

   Promotes slight extension of the wrist or prevent wrist flexion, thus assisting weak grasp (via tenodesis effect)

   a. Volar wrist-flexion control orthosis (cock-up splints)

- **Wrist-hand orthoses (WHOs)** in which the palmar section is extended (usually 20°. They are used to tighten finger flexors (via tenodesis effect) and prevent wrist flexion contracture in patients with radial neuropathy.

   b. Wire wrist-extension assist orthosis (Oppenheimer splint)

- Prefabricated from spring steel wire and padded steel bands to assist wrist extension by tensing the steel wire, thus aiding finger flexion through tenodesis effect

Wrist-driven prehension orthosis (tenodesis orthosis, flexor hinge splint)

Used in C6 complete tetraplegia (in which no muscles to flex or extend fingers remain innervated but wrist extension, through the extensor carpi radialis muscle, is intact) to provide prehension trough tenodesis action and maintain flexibility of the hand, wrist, and elbow.

- Wrist extensors should be 3+ or better to use body-powered tenodesis
- May interfere with manual WC propulsion.
- Rarely accepted by C7 and C8 tetraplegics who prefer to use their residual motor power or utensil holders.

RIC tenodesis splint

- Orthosis made of low-temperature thermoplastics in three separate pieces (wristlet, short opponens, and dorsal plate over index and middle finger)
- Easily and quickly fabricated; made as a training and evaluation splint for patients; light weight.
- Uses a cord/string running from the wrist piece, across the palm and up between the index and ring fingers. The string is lax when the wrist is flexed and tightens with wrist extension, bringing the fingers close to the immobilized thumb, accomplishing three-jaw chuck prehension.

Wrist driven prehension orthosis.

I. Wrist extension: three jaw chuck

II. Wrist flexion: release

Hand Orthoses

Wrist, Hand, and Finger Static Orthoses

Positional orthoses

Opponens orthoses : primarily used to immobilize the thumb to promote tissue healing and/or protection or for positioning of the weak thumb in opposition to other fingers to facilitate three-jaw-chuck pinch. Examples: short opponens splints, C-bar splints, cone splints, static thumb splints.

Opponens orthoses with wrist control attachments

- Ex: long opponens splints and thumb spica splints
- In addition to the benefits already mentioned for opponens orthoses (stabilizes first MCP), forearm bar maintains wrist in extension and prevent radial and ulnar deviation deformities

Opponens orthoses with lumbrical bar

- Finger orthosis that prevents metacarpal phalangeal joint hyperextension but allows full MCP flexion
- Prevents claw hand deformity (in addition to the benefits already mentioned for opponens orthoses)

Opponens orthoses with finger extension assist assembly

- Similar to basic opponens orthoses plus finger orthosis that assist proximal interphalangeal and distal interphalangeal extension
- Used for interphalangeal flex contracture, boutonnière deformity, or postsurgical release of Dupuytren's contracture.

Utensil holders/universal cuff (splints)/ADL splints

- Consist of a handcuff with palmar pocket onto which a utensil can be inserted.

Protective orthoses: used to protect wrist, hand, and / or fingers from potential deformity or damage by restricting active function / limiting motion

Digital stabilizers:

1. **Finger stabilizers/static finger orthoses (FOs):**

   Interphalangeal stabilizers (DIP, PIP, and DIP+PIP gutter splints, static finger splints, stax orstack splints, egg-shell finger casts, etc.)

   - FOs used to restrict motions at the PIP and DIP
   - Generally, IPs maintained in full extension to keep the collateral ligaments stretched and to prevent IP flex contracture (unless condition dictates otherwise)
   - Used to promote healing (e.g., phalanx fx, PIP / DIP dislocation, etc.) and to provide prolonged finger stretch (e.g., burns and contractures)

2. **Finger mobilization orthosis**

a. **Interphalangeal extension-mobilization orthoses-passively extend the PIP joints**

- **Uses:** Finger IP flexion contracture, Boutonnière deformity, and postsurgical release of Dupuytren's contracture
- **Examples:** Dynamic IP extension splints, reverse finger knuckle benders, Capener splints, safety-pin splints, spring coil assist, eggshell finger extension casts, buddy splints

b. **Interphalangeal flexion mobilization orthoses-passively flex PIP joints**

- **Use:** Finger IP extension contracture
- **Examples:** Dynamic IP flexion splints, finger-knuckle benders, fingernail book orthoses, buddy splints

Ring Stabilizers:

- **Swan neck ring-FO** that prevents hyperextension of the PIP joint (via three point pressure system) but allows full IP flexion
- **Boutonnière ring-FO** that immobilizes the PIP in extension (prevents flexion) through a three point pressure system.

Metacarpophalangeal ulnar-deviation restriction orthosis:

- FO used to limit ulnar deviation of the MCP with unrestricted (if possible) MCP flex / extension in arthritic patients with ulnar deviation at the MCPs.

Metacarpophalangeal (MCP) mobilization orthoses

a. **MCP- extension mobilization orthoses-**

- **Uses:** extend MCP joints in MCP-flexion contractures, burns, and post ORIF of metacarpal fracture, patients with weak finger extension (e.g., radial nerve lesion and brachial plexus lesion).
- **Examples:** Reverse MCP knuckle benders, dynamic MCP extension splints with dorsal outrigger, MCP extension assists; radial nerve splints

b. MCP-flexion mobilization orthoses

- **Uses:** used to flex MCP joints in MCP collateral ligament contractures, extensor tendon

shortening, median / ulnar lesion, claw hand, postcapsulotomy, post ORIF of metacarpal fracture.

- **Examples:** MCP knuckle benders, dynamic MCP flexion splints with volar outrigger and fingernail hooks, MCP flexion assists

Thumb stabilizers

Thumb carpometacarpal stabilizers/thumb posts

- Thumb orthosis that stabilizes the first CMC and MCP joints in neutral position to protect the thumb form inadvertent motion. Thumb-web space stabilizers / thenar web spacers / c-bar splint.
- FO that consists of a rigid C-shaped splint held firmly in the thumb and index finger web space.
- **Function:** increases or maintains the thenar space and prevents web-space contractures
- **Uses:** Burns, postsurgical revision of scar, web-space contractures

Thumb mobilization orthoses

a. **Thumb extension-mobilization orthosis-** dynamic thumb IP extension splints. Use: thumb IP flexion contracture.

b. **Thumb flexion-mobilization orthosis-** dynamic IP flexion splints

Use: Thumb IP flexion contractures.

c. **Thumb abduction-mobilization orthosis-** dynamic thumb abduction splint.

Use: Thumb-adduction contracture.

Upper Extremity Orthoses With Special Functions

Tone-Reducing Orthosis

Theoretical basis for tone-reducing orthosis

- Inhibition of reflexes
- Pressure over muscle insertions
- Active and static prolonged stretch
- Orthokinetics

Inhibition of reflexes

- A reflex consists of a motor act that is elicited by some specific sensory input
- Primitive reflexes appear at birth and become integrated once more complicated movements emerge
- When the CNS is damaged, primitive reflexes reemerge and again dominate motor activity

Pressure over muscle insertion

- Farber reported in 1974 that continuous firm pressure at point of insertion reduces tone

Active and static prolonged stretch

- Decrease reflex tone by providing mechanical stabilization of the joint and altering properties of the muscle spindle

Orthokinetics

- Originally developed in 1927 by Julius Fuchs, an orthopedic surgeon
- Focuses on physical effects to materials placed over muscle bellies
- Passive field materials (those that are cool, rigid, and smooth) produce inhibitory effect
- Active field materials (those, warm, expansive, and textured) produce facilitatory effect

Examples of Upper Extremity Tone-Reduction Orthoses are

A. Anti-spasticity "Ball" Splint.

B. Hand Cone Splint.

- They can be either hand-based wrist-hand orthoses (e.g., hand-cone splints) or forearm-based wrist-hand-finger orthoses (e.g., antispasticity ball splints and Snook splints).
- They can be volar based, dorsal based, or circumferential
- Typically worn two hours on and two hours off throughout the day
- Forearm based splints usually are more effective because of the extension positioning of the extrinsic finger flexors

Rationales of efficacy of tone-reduction orthoses include:

- **Reflex-inhibiting positioning-NDT** technique approach (Bobath)
- **Firm pressure into volar surface (PALM)-** Rood (sensorimotor) approach
- **Dorsal-based splints (eg, Snook):** facilitation of muscle contraction by direct contact-it is

theorized that stimulation of extensor surface might produce extensor muscle contraction and balance muscle tone and/or avoid increase flexor tone.

- **Functions:** flexor tone reduction, prevent skin breakdown/maceration of palm by fingernails, increase passive range of motion via low-load, prolonged stretch (serial static splinting)
- **Indications:** spasticity-upper motor neuron lesions (cerebral vascular accident, HI, multiple sclerosis, cerebral palsy)

Evaluation

The therapist must understand the complex and intricate interrelations of normal anatomic structures, their kinesio- logic functions and their biomenchanical and physiologic ramifications before attempting to interpret, define and treat the abnormalities that accompany upper extremity problems.

Prior to evaluation, gathering patient history is important and can be used to establish a rapport with the patient and the family members. Information that are required from the patient, caretakers and health care professionals include details/ cause of initial injury, previous medical care, reason for seeking additional care and desired outcomes of new treatments based on the wide range of specific functions a patient performs daily.

The evaluation of upper extremities requires inputs regarding strength, ROM, condition of soft tissues and sensation. In addition, ambulatory status, bilateral or unilateral condition, status of vision, and condition of the spine and head need to be considered before deciding on the appropriate orthotic device for the patient.

Since minor changes in strength dramatically change the orthotic need, many critical muscle tests must be made in patients with the upper extremity dysfunction. A patient with unilateral involvement can be provided with a typical prefabricated positional wrist hand orthosis (WHO) to prevent contracture and injury and a supporting shoulder orthosis for shoulder subluxation. On the other hand for a patient with bilateral involvement, conservation for grooming, feeding mobility, etc must be considered and the patient may be needed to be fitted with a more complex and customized orthoses.

There are similarities in the orthotic management of orthopedic and neurologically impaired patients. However, the neurologically impaired patients additional factors that challenge prescription criteria for the rehabilitation team. Lack of propriception, sensation, (hypher or hyposensitivity) and spasticity represent some of these special considerations and possible probems with communication add to these patient management complications.

Evaluation of the neurologically impaired patient must be comprehensive. The therapist must assess the client or family motivation, client ability to tolerate and or function with the orthosis and risks associated with orthotic intervention. Further a total evaluation of the patient environment is important in developing the patient plan.

The medical diagnosis should alert the evaluator to the patterns associated with identified impairments aand should be used to confirm potential findings. In addition to ROM evaluations, muscle power testing, assessment of sensation, skin sensitivity or lack, integrity of the affected limb or spine the therapist must assess what limitations initiating the orthotic care may have on other important functions.

Goals of Orthotic Intervention

The clinical experience of the therapist and patient evaluation must be used to create a plan of treatment. Only a well thought out plan that is thoroughly communicated to all the participants can ensure success of the intervention. Several other key factors that have an impact on the success of intervention are:

- Need to address the major complaint of the patient.
- Need to increase function without complication or patient risk.
- Need to establish a baseline of function so that results of intervention are measureable.
- Ability of the patient to independently donn and doff the orthotic device.

In some cases, concentrated instructions, orthotic modifications and time are required before improved function can be observed. It is essential that the orthotic intervention be as simple as possible and to ensure that the least amount will help achieve the goals of intervention. The treatment goals need to be realistic, manageable and well discussed with the patient and care takers to be successful.

Operation

Although there is a wide variety of upper limb orthoses, most of these devices operate on similar principles. The general goal of most orthoses is to provide stability and support while allowing as much motion as possible. Immobilizing joints for long periods has proven deleterious for most patients since muscles atrophy, joints stiffen, skin tightens, and the healing process is ultimately slowed. By allowing movement while restricting motion that would create stress on joints, muscles, or tendons, orthotic devices allow healing and preserve range of motion and function.

Exact fit is a key element for many upper limb orthoses. In order to work properly, the orthosis must hold the body part in an exact position since if the orthosis does not fit exactly, it may not work and may actually cause harm. This can become a problem in situations where the patient has experienced swelling and may require a new fitting for the orthosis once the swelling has resolved. Poor fit can also lead to discomfort and the development of pressure sores.

Dynamic orthoses usually operate with the aid of attached outriggers. These provide a place to attach rubber bands, springs, or other materials that assist motion. They also provide leverage and help to ensure that the joint stays in proper position during movement. These devices require exact fit, as well as adjustment to ensure that the device works properly.

Many upper limb orthoses require a period of training for the patient to learn how to use the device properly. This is especially true with devices that assist motion, because the patient must initiate the motion properly in order for the orthosis to work. Patients with a long history of paralysis or immobilization may require considerable time in order to learn how to use the device. Orthotic treatment must maximize what little muscle strength and ROM the patients have. Further, orthoses used during the day for functional purposes might have to be replaced with positional orthoses to preserve gain and to prevent decline.

It is very important to consider the patient's motivation and attitude toward the orthosis as part of the treatment plan. Since most upper limb orthoses are removable, patients can choose whether or not to use these devices. Patients may object to orthoses because of discomfort, unattractive appearance, or restrictiveness of the device. Health care professionals must work closely with the patient to ensure that the patient will accept the orthosis and use it properly.

Exercise: Splints and exercises must be carefully integrated to allow the patients to achieve their full rehabilitation potential. Splints are used to improve passive motion, substitute for weakened or lack of active motion and are infrequently fabricated to provide resistive exercise. Evaluation measures provide the guidelines from which splinting and exercise programs are coordinated. Because patients with identical diagnosis may respond differently to therapeutic interventions, it is insufficient and often detrimental to adhere to rigid pre determined protocols and the exercise and splinting programs will require constant alterations to keep pace with the changing requirements. Patients are generally instructed to repeat a splint wearing and exercise routine every two hourly through out the day. This involves wearing the splint for one hour and forty five minutes and then removing the splint and exercising for 15 minutes. Conversely if stiffness is a problem, exercise may be prescribed more often.for e.g. every 45 minutes. Written instructions for splint wearing and exercise routine should be used to augment verbal instructions.

The patients and the family should also be instructed regarding

- Donning and doffing
- Wearability
- Precautions

Patients must be taught to monitor the status of their splinted extremities. The presence of pain, reddened areas, blisters, swelling, rashes, or other problems associated with wearing a splint should be noted immediately and the use of splint discontinued until the appropriate corrective measures have been taken to rectify the problem.

Patients should be cautioned about exposing their splints to warm or hot temperatures such as hot water heaters or stoves or leaving the splints in hot car interiors since thermoplastic splints may lose or alter their configurations in these conditions.

Maintenance

Many upper limb orthoses require little or no maintenance. This is especially true for static orthoses and for those intended for temporary use. The plastic shell can be wiped clean, and materials

worn underneath the orthosis can be washed or replaced. The patient may need to be checked periodically to ensure that the orthosis fits. Dynamic orthoses may require adjustments and replacement of worn springs, rubber bands, and the like

Splinting in stroke patients.

Within the literature there is conflicting evidence and opinions on whether to use splinting as a form of intervention for stroke patients. There is a lengthy debate on the theoretical basis for splinting within neurology with two conflicting theories of biomechanical and neurophysiological approaches (Copley and Kuipers, 1999). Biomechanical rationale argues that splinting is used to prevent and manage length-associated changes in muscles and connective tissues. The neurophysiological rationale recommends that splinting is used to inhibit reflexive contracture of the muscle. The decision on whether to splint must be made on sound clinical reasoning. There are a wide variety of materials and types of splints and all have different qualities. Splints should not be considered when there is active movement that would be restricted if a splint should be provided.

Lannin and Herbert (2003) found a lack of evidence for splinting following stroke. In 2007, Lannin et al. conducted a randomised control trial and found no evidence to support the provision of a splint to prevent contracture in the acute phase for spasticity as a method of prevention of contracture. Regular passive range of movement and stretching is recommended and it is important to provide teaching to the patient and carer to perform this programme. The type of splint chosen will vary depending on the ideal position, but may include cones or resting splints in volar, dorsal or mid prone.

Reduction of pain

Following a stroke, pain can occur in various joints although wrist pain is a common complaint among patients. This can be due to the wrist being in a prolonged flexed position due to spasticity or flaccidity, leading to overstretching and/or shortening of muscles. Provision of a volar resting splint may be beneficial to prevent further harm and provide support. This must be alongside regular range of movement.

Maintaining joint alignment

Within the early stages of low tone, the hand may lose the curvatures due to prolonged resting in a flat position. A resting splint may be considered to maintain the hand's natural curves and prevent secondary complications developing.

Functional aims of splinting in stroke patients

- To improve grasp in functional activities.
- To increase range of movement to open hand easily to enable daily hygiene.
- To use the upper limb pain free within activities.

Orthoses for SCI patients: Though the evaluation and rationale for determining the appropriate splint per injury level are agreed upon, the time of providing the splint and whether permanent orthoses enhance long term functions is debated. However early splinting for positioning to prevent deformity has proven to be effective at all injury levels.

Clients with C1 to C4 tetraplegia require resting hand splints to assist with proper positioning and maintain the support of the wrist and web space

Clients with Tetraplegia at C5 level can be independent with communication, feeding and hygiene only with the assistance of an orthosis. They must have joint stability and support at the wrist the hand to perform these skills. Static splints such as wrist cockup, long opponence and resting hand splints can be given. A universal cuff can be provided which include a slot for a spoon, fork, comb, tooth brush or hair brush so that a patient can effectively feed or groom himself.

Clients with injury at the C6 level can use their wrist for a tenodesis grasp. Clients who are not strong enough to use their wrist for tenodesis may require splinting to support their wrist until they can use their wrist against gravity. Long opponens splint can be used to position the thumb for function but support the weak wrist. Once the wrist strengthens, the long opponens splint can be cut down to a hand based short opponens splint to maintain proper web space and thumb positioning and maximize tenodesis.

Clients with C8 to T1 injuries or clients who have incomplete injuries may experience "clawing". A metacarpo-phalangeal block splint to block the MCP joints and promote weak muscle function can be given. Finally educating the client on the splint wearing schedules, skin checks and splint care are very important to prevent skin breakdown.

Orthoses for patients with cerebral palsy: In children with cerebral palsy, orthotic intervention is used in combination with an active task oriented therapy program. The new and wide variety of thermoplastic materials available to fabricate customized orthotics provides many options for upper and lower extremities as well as for trunk. The least restrictive devices, soft splints can be constructed from webbing, neoprene, hook and loop fastener material and other substances. Soft splints do not limit dynamic mobility and sensory feedback as much as do thermoplastics. For children with moderate degree of spasticity stronger moulded thermoplastic materials are necessary to provide stability. They are lighter in weight than the original metal and leather braces and are easily cleaned. Some of the low temperature plastics can be altered as growth or change occurs. OT have found that upper extremity casting which provides prolonged and gentle stretch to spastic or contracted muscles is an effective adjunct to therapeutic techniques. Results of upper extremity casting have been significant with increased strength, control, and spontaneous use of impaired arm as well as bilateral hand use during play and transitional movements. The casting program is always integrated into the therapy program with the same functional goals.

Summary:

Therapist's clinical expertise alongwith a sound understanding of orthotic principles, knowledge of multiple orthotic options available are vital to reach the desired outcome of the orthotic intervention. Since every patients' need for an orthotic intervention is unique, it is essential to develop an individualized splint wearing and exercise schedule with periodic reassessments. Coordination between individual team members such as Orthotist, therapist (occupational physical or hand therapist) patient and family (especially in case of children) is essential for successful orthotic intervention.

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Ch.14 Psychological Rehabilitation

Ms. Akshata Shetty, M.A.(Clinical Psychologist)

DEFINITION OF PSYCHOLOGY:

The word psychology is derived from the Greek words Psyche (which means soul) and logos (which means study). Hence, psychology could be defined as a "study of the soul". However, today it is defined as the scientific study of the behaviour of individuals and their mental processes (American Psychological Association).

IMPORTANCE OF PSYCHOLOGY:

Psychology is important in our daily living as it is concerned with the study of behaviour and mental processes and at the same time, it is also applied to many different things in human life. It is primarily a study of who and what we are, why we are like that, why we act and think like that and what we could be as a person. Psychology helps us in understanding about human perceptions, cognition, emotion, personality, behaviour and interpersonal relationships.

Psychology also attempts to understand the role these functions play in social behaviour and in social dynamics, while incorporating the underlying physiological and neurological processes into its conceptions of mental functioning. It helps the patients who have emotional, cognitive and behavioural deficits or issues to understand the problem, the severity of it, to accept the situation and ways to either reduce the discomfort caused or eliminate it with the help of various psychotherapies.

Process of a Psychological Intervention:

- Build rapport with the patient.
- To take down the case history and document it
- To assess the patient with the help of formal and informal assessments and make reports
- To diagnose the patient
- To educate the patient and family members, about the diagnosis and explain the intervention plan
- To make referrals to other professionals id needed
- To conduct various psychotherapies depending on the issues of the patient
- Help the patient to maintain the condition and
- Educate the patient for further maintenance

EMOTIONAL, BEHAVIOURAL AND COGNITIVE PROBLEMS THAT PATIENT WITH DETERIORATING INCURABLE DISEASE OR DISORDERS AND LIFE CHANGING TRAUMA:

Cognitive Changes or Deficits:

Cognitive changes or deficits in patients could be depending on the whether it is due to traumatic brain injury, or due to a deteriorating disease like dementia, or cognitive deficit since birth or taking place due to general medical condition. Patients could have a wide variety of cognitive changes or deficits like problems in perception, orientation, episodic memory, shot-term memory, sequential memory, long-term memory, working memory, poor judgment, poor problem solving, difficulty in concept formation, difficulty in decision making, etc.

Personality Changes:

Changes in the personality of patients are especially disturbing for their families. Pre-existing personality traits may be accentuated during the course of the illness. Patients may become introverted and seem to be less concerned than they previously were about the effects of their behaviour on others. Patients could also have paranoid delusions are generally hostile to family members and caretakers. The personality changes could take place depending on the disease for example patients with frontal and temporal involvements are likely to have marked personality changes and may be irritable and explosive. Also, patients with traumatic brain injury could have overt sexual behaviour, which could be completely different as compared to their premorbid personality.

Mood:

In addition, to psychosis and personality changes, depression and anxiety major symptoms in patients, although the full syndrome of depressive disorder

may be present in only 10 to 20 per cent. Patients may also may exhibit pathological laughter or crying that it is, extremes of emotions with no apparent provocation. Some could even quality for a diagnosis of mood disorders.

Depression, Anxiety and Psychological Distress:

Depression has been correlated to suffering, hopelessness, fast deteriorating or inability to cope with the disorder and social withdrawal which is in agreement to the quality of life. It is often seen that adults suffering from an incurable disease have a sense of lack of emotional support, lack of coping skills and a very negative view of the future. Patients may suffer from pain which may lead to depression. However, it is seen that it may not be so much due to the severity of the disability but it may depend but would depend on how well they cope with the disability. Children and adults may develop depression as being unable to control their body, embarrassment about their body in social situations and lack of information about their situation. Also guilt could be a reason of depression as the patients may see their caregivers suffering due to them.

Loss of Control:

Patients often undergo a feeling that they have lost control of their body, thoughts or feelings. This could eventually lead to impetuous decisions like resigning from job, or thinking of living alone as they feel their caregivers have to go through a lot because of them, etc. The patients should often learn to identify the control issues, to manage effectively and to cope up with situations which are difficult to change.

Poor or Low Self-Image and Self Doubt:

Am I really sick? Am I imagining this? Am I weak? Did I cause this illness? If I had eaten better or exercised more, would I be well today? Patients find it very difficult to accept themselves. They often compare the things that they were able to do before and how their condition has deteriorated now. This often leads to a lot of frustration and eventually being unable to accept their self -image. It would be difficult to accept ones self-image as conditions which are deteriorating or incurable need constant adjustment to new changes. It is very important to examine why the lost abilities were important and to find an alternative method of meeting those needs.

Anger and Aggression:

Anger is a common and natural reaction to the frustrations one encounters while dealing with a disease. While having some anger about living with the frustrations, symptoms, and anxieties associated with having a chronic illness is appropriate and healthy, sometimes anger is expressed indirectly as sarcasm, cynicism, or irritability. Other people may feel consumed by anger. Either way, anger about the directed toward others or bitterness can have damaging effects on a person's relationships with others. An important task for many people suffering from a deteriorating disease or a disorder is recognizing the anger at the disease, learning to direct it properly at the disease rather than at loved ones, and managing anger and aggression effectively.

Isolation:

Many people with a disease or a disorder, as well as families and caregivers, can become socially isolated. Early in the disease, people may avoid talking to friends or family about their disease. Sometimes they avoid doing so because they fear others will treat them differently, or sometimes they avoid these discussions because they don't want to burden others or make them worry. However, this avoidance can lead to increased withdrawal, resulting in isolation, loneliness, and depression, both for the person living with the disease and his or her loved ones. People who are more severely affected physically by a disease sometimes find it physically harder to have contact with others. Sometimes, people with disease (particularly those who don't know anyone else with a similar disorder or disease) feel that no one could understand what it is like living with this disease, so there is no point in trying to talk with others about it. However, there is a strong relationship between isolation, depression, and increased health problems.

While maintaining relationships is not always easy, it is very important to ones mental and physical health. Maintaining social contact often requires new methods of approaching relationships. Often it requires becoming more open with people about how the disease affects you and how it might impact your relationship. Support groups, volunteering, involvement in religious or spiritual practices and organizations, and taking classes at junior colleges are but a few ways people find meaningful contact with others. Many people have begun using Internet chat rooms. While the Internet

can be helpful, we caution against spending so much time that it interferes with actual contact with other people. New research suggests that too much time on the Internet, at the expense of time face to face with other people, may increase depression and social isolation.

Loss of Independence:

Loss of independence can be a major issue for the patients and the family members. These issues can be emotional, physical, medical and financial. Some people rush headlong into a dependent role, only to find themselves, then feeling empty, hopeless, or useless. Others may fight their fears of dependency to the point where it is physically dangerous. For example, someone who is uncomfortable with dependence on walking aids may avoid using a cane or crutches, even though he or she is falling and getting injured. Often, people have uncomfortable feelings about starting new medications, and may reject treatments which can be beneficial. Recognizing ones own issues around dependency is the first step towards preventing them from putting you at risk.

Abandonment:

Fear of abandonment can be one of the most frightening issues a person faces. Many people, particularly in the earlier stages of the illness, fear their spouse or partner will want to leave them. These fears are often so anxiety-provoking that couples avoid talking about this issue. For people with very severe disease late in the illness, people may fear being moved to nursing facilities to be abandoned. For those who are not married or in a relationship, there are often questions about whether they will ever find a life partner, or if they will lead a life abandoned by others. These thoughts can be so frightening that people avoid discussing them with anyone. Such fears are sometimes self-fulfilling, as people fearing abandonment may push others away through angry, withdrawing, or other behaviors. Avoiding this topic has never been known to save a relationship. Discussing these fears openly often results in feelings of relief for all concerned.

Uncertainty:

Because the course of certain diseases like Multiple Sclerosis, Motor Neuron Disease, Muscular Dystrophy, etc is so unpredictable, many people experience considerable uncertainty and anxiety. This uncertainty often makes life planning difficult. Major milestones, such as changing jobs, taking on a mortgage, or having a child, all weighty decisions for anybody, can seem impossible to resolve when the uncertainty of MS is involved. Sometimes, people will spend so much of their time and resources preparing for things they fear in the future they miss out on pleasures, goals, and desires in the present.

Of course, there are also numerous other issues which can be experienced by the patient, including feelings of hopelessness, anxiety, issues around work and disability, and many others. These issues can be overwhelming at times. However, they are not insurmountable. Very few people with a particular disorder or disease have difficulties in all areas all the time. Most people experience periods of regular day-to-day life interspersed with periods when one or more of these issues become prominent. Most people will also find that they are able to cope with many of these issues much of the time; yet from time to time, one or more of these issues might becomes overwhelming.

Emotional Instability:

The emotional instability is manifested by irritability, anxiety and depression, as well as euphoria, even to the extent of a manic or depressive psychosis. Denial is manifested by projection of frustration and hostility onto other people, or by unrealistic optimism. Threat, to personality functioning, produces the compensatory psychological defence mechanisms of denial and projection.

Self-dislike:

Why should I try to look attractive when I'm so dependent? Who wants to employ someone who might be ill all the time? Why can't I overcome this illness?

Family Problems:

Patients who are suffering from a deteriorating or incurable disorder can cause serious problems within the family dynamics. Patients usually undergo feelings of fear, that if they demand anything from their partner, his demands may be rejected. Often the relatives become overprotective with underlying feelings of hostility. This may be a serious when a parent becomes overprotective, especially if he sufferer is married. Other patients would deny the disease so much that they refuse to acknowledge any limitations. Life then becomes

difficult for all members of the family in a different way.

Quality of Family Support:

The most important question to be answered is "What kind of relationship the patient has with his close relatives (children, spouse, and extended family)?" It is important to know because the psychological suffering of a patient may result from tensions within his family (rejection, stigma, exclusion, indifference). Often a psychological maltreatment develops between the patient and his relatives; they feel unable to bear the daily progress of the disease.

Support for the Caregivers:

Many patients require long-term physical, financial, and psychological support from family and friends. Both the physical and mental health of the caregiver is critical. The burden may be considerable and threaten both. In fact, the more hours dedicated to the patient, the more depressed the caregiver is likely to be. This leads to a cycle of poor caregiving followed by reduced functioning in the patient.

According to Boot et al. (2008) psychological input can have benefits in the following areas:

- Managing their mood better
- Coping better
- Improved levels of daily activity
- Better understanding of their difficulties
- Improved relationships
- Less prone to feelings of suicide
- More confident about managing their future with MS.

Stigma:

While self-image refers to how you think about yourself, stigma refers to how others view you. Unfortunately, in some environments it is common for people to stare, be patronizing or overly solicitous, or to be avoidant of a patient whose symptoms that are visible to others. If your symptoms are not immediately visible to others, such as cognitive problems or fatigue, some people may say you're just ignoring me or you're just lazy, rather than attribute the symptoms to the disease. It is difficult for many people to adjust to the changes in how they are treated by others. However, it is also important to distinguish between true stigma, and difficulties one has in ones own self-image which are projected onto others. For example, a person who grew up feeling uncomfortable around people in wheelchairs, and who now finds him / herself in a wheelchair, may imagine that others are uncomfortable even when they are not. In this way, people sometimes imagine there is stigma even when there is none. How people choose to deal with discrimination and stigma varies from situation to situation, and from person to person. Some people find that confronting discrimination directly and advocating socially and / or politically for the rights of people with chronic illnesses and disabilities helps them feel empowered. Other people find taking an educational approach to discriminatory situations is best for them, and still other people find that viewing discrimination as their problem / weakness / loss, not mine, and instead focusing on their own health, needs, and well-being is best.

PSYCHOLOGICAL TESTING:

Intelligence Testing:

Intelligence is defined as the ability to assimilate factual knowledge to recall either recent memory or remote events, to reason logically to manipulate concepts (either numbers or words), to translate the abstract to the literal to the abstract, to analyze and synthesize forms and to deal meaningfully and accurately with problems and priorities deemed important in a particular setting. Intelligence varies for individual to individual.

Wechsler Adult Intelligence Scale® - Third Edition (WAIS®-III):

Description: The WAIS- III is the best standardized and most widely used intelligence test in the clinical practice today. It was constructed by David Wechsler at New York University Medical Centre and Bellevue Psychiatric Hospital. WAIS - III has 11 subtests which are made up of six verbal sub-tests and five performance sub-tests, which yield a verbal IQ, a performance IQ and a combined or full scale IQ. The sub-tests are as follows:

- **The Verbal subtests are:** Vocabulary, Similarities, Arithmetic, Digit Span, Information and Comprehension.
- **The Performance subtests are:** Picture Completion, Digit Symbol - Coding, Block Design, Matrix Reasoning and Picture Arrangement

Age group: Between 16 to 89 years of age

Target Group: Adults with cognitive deficits due to age related or trauma to the brain, or due to other diseases, etc.

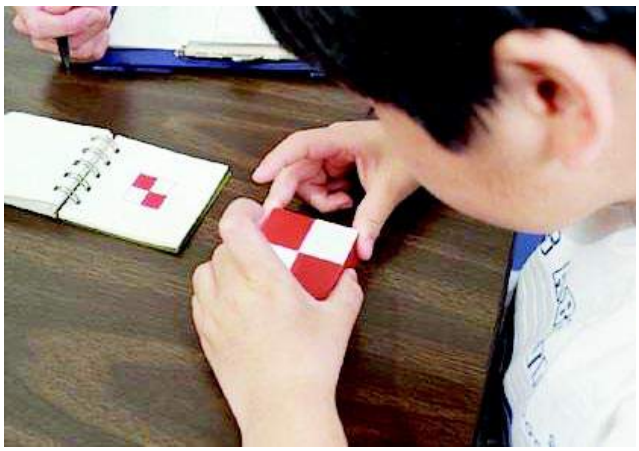
Wechsler Intelligence Scale for Children (WISC):

Description: This test was developed by Dr. David Wechsler, is an individually administered intelligence test for children that can be completed without reading or writing. It is also used to diagnose attention-deficit hyperactivity disorder (ADHD) and learning disabilities. The WISC can be used as part of an assessment battery to identify intellectual giftedness, learning difficulties, and cognitive strengths and weaknesses. When combined with other measures such as the Adaptive Behaviour Assessment System-II and the Children's Memory Scale its clinical utility can be enhanced. Combinations such as these provide information on cognitive and adaptive functioning, both of which are required for the proper diagnosis of learning difficulties and learning and memory functioning resulting in a richer picture of a child's cognitive functioning.

Age Group: Between the ages of 6 and 16

Time taken for Administration: 65-80 minutes

Target Group: Attention Deficit Hyperactivity Disorder, Learning Disability, Mentally Retarded Children, Cerebral Palsy, Autism, Children with Duchenne's Muscular Dystrophy with comorbid Mental Retardation, etc.



Wechsler's Intelligence Scale for Children

Memory Test:

Memory is defined as "the ability of an organism to store, retain, and recall information and experiences".

Wechsler Memory Scale Fourth Edition (WMS - IV):

Description: This is a neuropsychological test designed to measure different memory functions in a person. The current version is the fourth edition (WMS-IV) which was published in 2009 and which was designed to be used with the WAIS-IV. WMS-IV is made up of seven subtests: Spatial Addition, Symbol Span, Design Memory, General Cognitive Screener, Logical Memory, Verbal Paired Associates, and Visual Reproduction. A person's performance is reported as five Index Scores: Auditory Memory, Visual Memory, Visual Working Memory, Immediate Memory, and Delayed Memory.

Age Group: From 16 to 90 years.

Time taken for Administration: 75 minutes

Target Group: Individuals with Memory deficits.

Neuropsychological Tests:

Neuropsychological Tests are specifically designed tasks used to measure a psychological function known to be linked to a particular brain structure or pathway. Tests are used for research into brain function and in a clinical setting for the diagnosis of deficits. They usually involve the systematic administration of clearly defined procedures in a formal environment. Neuropsychological tests are typically administered to a single person working with an examiner in a quiet office environment, free from distractions.

Mini Mental Status Examination:

Description: The mini-mental state examination (MMSE) or Folstein test is a brief 30-point questionnaire test that is used to screen for cognitive impairment. It is commonly used in medicine to screen for dementia. It is also used to estimate the severity of cognitive impairment at a specific time and to follow the course of cognitive changes in an individual over time, thus making it an effective way to document an individual's response to treatment. The MMSE test includes simple questions and problems in a number of areas: the time and place of the test, repeating lists of words, arithmetic such as the serial sevens, language use and comprehension, and basic motor skills.

Time Taken for Administration: 10 minutes

Target Group: Patients with dementia

Scoring:

25 - 30 : Normal

21 - 24 : Mild Cognitive Impairment

19 - 20 : Moderate Cognitive Impairment

10 - 20 : Mild Cognitive Impairment

00 - 10 : Severe Cognitive Impairment

A low to very low scores correlate closely with the presence of dementia, although other mental disorders can also lead to abnormal findings on MMSE testing. The presence of purely physical problems can also interfere with interpretation if not properly noted; for example, a patient may be physically unable to hear or read instructions properly, or may have a motor deficit that affects writing and drawing skills.

Luria-Nebraska neuropsychological battery:

Description: This is a standardized test based on the theories of Alexander Luria regarding neuropsychological functioning. There are 14 scales: motor functions, rhythm, tactile functions, visual functions, receptive speech, expressive speech, writing, reading, arithmetic, memory, intellectual processes, pathognomonic, left hemisphere and right hemisphere. It is used with people who are 15 years or older; however, it may be used with adolescents down to 12 years old. Part of A.R. Luria's legacy was the premium that he placed on the observation of a patient completing a task; intra individual differences. The modern practice of standardized testing tends to neglect this aspect of psychology. The Luria-Nebraska Neuropsychological Battery (now in its third iteration) attempts to create an alloy of standardized testing and idiosyncratic observation by allowing comparison to the normative sample, and at the same time giving the test administrator flexibility in the administration.

Age Group: 15 years and up

Time taken for Administration: 1 ½ hours and 2 ½ hours.

Rey-Osterrieth Complex Figure Test (ROCF):

Description: This is a neuropsychological assessment in which examinees are asked to reproduce a complicated line drawing, first by copying and then from memory. Many different cognitive abilities are needed for a correct performance, and the test therefore permits the evaluation of different functions, such as visuospatial abilities, memory, attention, planning, and working memory (executive functions). First proposed by Swiss psychologist André Rey in 1941 and further standardized by Paul-Alexandre Osterrieth in 1944, it is frequently used to further elucidate any secondary effect of brain injury in neurological patients, to test for the presence of dementia, or to study the degree of cognitive development in children.

Halstead-Reitan Neuropsychological Battery:

Description: This is a combination of neuropsychological tests used to assessment the possible physical aspects and localization of neurological damage. The Halstead-Reitan is typically used to evaluate individuals with suspected brain damage. The battery also provides useful information regarding the cause of damage (for example, closed head injury, alcohol abuse, Alzheimer's disorder, stroke), which part of the brain was damaged, whether the damage occurred during childhood development, and whether the damage is getting worse, staying the same, or getting better.

The Battery includes:

- **Trails A and B** (which see how quickly a patient can connect a sequence of numbers (trail A) or numbers and letters (trail B).
- **Controlled Oral Word Association Test (COWAT, or Verbal Fluency)** - a measure of a person's ability to make verbal associations to specified letters.
- **Halstead Category Test** (including seven subtests which form three factors: a Counting factor (subtests I and II), a Spatial Positional Reasoning factor (subtests III, IV, and VII), and a Proportional Reasoning factor (subtests V, VI, and VII).)
- **Tactual Performance Test**
- **Rhythm Test**
- **Speech Sounds Perception Test**
- F**inger Oscillation Test**

Age Group: 15 years and older

Personality Tests:

Personality is defined as the particular combination of emotional, attitudinal, and behavioral response patterns of an individual. A personality test is a questionnaire or other standardized instrument designed to reveal aspects of an individual's

character or psychological makeup. The first personality tests were developed in the early 20th century and were intended to ease the process of personnel selection, particularly in the armed forces.

Minnesota Multiphasic Personality Inventory-2 - Restructured Form:

Description: The original authors of the MMPI were Starke R. Hathaway, PhD, and J. C. McKinley, MD. This is one of the most frequently used personality tests in mental health. The test is used by trained professionals to assist in identifying personality structure and psychopathology. The MMPI-2-RF aids clinicians in the assessment of mental disorders, identification of specific problem areas, and treatment planning in a variety of settings. **The test can be used to help:**

- Assess major symptoms of psychopathology, personality characteristics and behavioural proclivities.
- Evaluate participants in substance abuse programmes and select appropriate treatment approaches.
- Assess medical patients and design effective treatment strategies, including chronic pain management.
- Support classification, treatment, and management decisions in criminal justice and correctional settings.
- Identify high-risk candidates in public safety screening and selection settings.
- Give strong empirical foundation for expert testimony in forensic evaluations.
- Support college and career counselling recommendations.
- Provide valuable insight for marriage and family counselling.

Age Group: 18 years to 80 years

Time taken for Administration: Individual - 35 to 50 minutes.

MMPI Test Answer Sheet

Sixteen Personality Factor Questionnaire Revised - 5th Edition (or 16PF- R 5th Ed.): This is a multiple-choice personality questionnaire which was developed over several decades of research by Raymond B. Cattell and his colleagues.

Each factor can be measured on a scale, determined by completing a questionnaire, and the word pairs below indicate the extremes of each scale. The letter codes were ascribed to each scale as a shorthand notation.

Factors		**Descriptors**	
A	Warmth	Reserved	Outgoing
B	Reasoning	Less Intelligent	More Intelligent
C	Emotional Stability	Affected by feelings	Emotionally stable
E	Dominance	Humble	Assertive
F	Liveliness	Sober	Happy- go - lucky
G	Rule Consciousness	Expedient	Conscientious
H	Social Boldness	Shy	Venturesome
I	Sensitivity	Tough-minded	Tender-minded
L	Vigilance	Trusting	Suspicious
M	Abstractedness	Practical	Imaginative

N	Privateness	Straightforward	Shrewd
O	Apprehension	Self-assured	Apprehensive
Q1	Openness to Change	Conservative	Experimenting
Q2	Self- Reliance	Group - dependent	Self -sufficient
Q3	Perfectionism	Self - conflict	Self - control
Q4	Tension	Relaxed	Tense

Age Group: Individuals 16 years and older

Projective Tests:

In psychology, a projective test is a personality test designed to let a person respond to ambiguous stimuli, presumably revealing hidden emotions and internal conflicts. This is different from an "objective test" in which responses are analyzed according to a universal standard (for example, a multiple choice exam). The responses to projective tests are content analyzed for meaning rather than being based on presuppositions about meaning, as is the case with objective tests.

Rorschach inkblot test:

Description: The test is named after its creator, Swiss psychologist Hermann Rorschach. This is a psychological test in which subjects' perceptions of inkblots are recorded and then analyzed using psychological interpretation, complex algorithms, or both. Some psychologists use this test to examine a person's personality characteristics and emotional functioning. It has been employed to detect underlying thought disorder, especially in cases where patients are reluctant to describe their thinking processes openly.

The core of scoring revolves around coding the response according to all of the blot features that have contributed to the formation of the response. The following characteristics are coded:

- Form
- Movement - when any movement occurred in the response
- Chromatic Color - when color is used in the response
- Achromatic Color - when black, white or grays are used in the response
- Shading-texture - when texture is used in the response
- Shading-dimension - when dimension is used in the response with reference to shading
- Shading-diffuse - when shading is used in the response
- Form dimension - when dimension is used in the response without reference to shading
- Pairs and reflections - when a pair or reflection is used in the response.

Rorschach Inkblot Card No. 2

Rorschach Inkblot Card No. 10

Thematic Apperception Test (TAT):

Description: The TAT is often administered to individuals as part of a battery, or group, of tests intended to evaluate personality. It is considered to be effective in eliciting information about a person's view of the world and his or her attitudes

toward the self and others. As people taking the TAT proceed through the various story cards and tell stories about the pictures, they reveal their expectations of relationships with peers, parents or other authority figures, subordinates, and possible romantic partners. In addition to assessing the content of the stories that the subject is telling, the examiner evaluates the subject's manner, vocal tone, posture, hesitations, and other signs of an emotional response to a particular story picture.

The TAT is usually administered to individuals in a quiet room free from interruptions or distractions. The subject sits at the edge of a table or desk next to the examiner. The examiner shows the subject a series of story cards taken from the full set of 31 TAT cards.

Children's Apperception Test (CAT):

Description: The Children's Apperception Test was developed in 1949 by Leopold Bellak and Sonya Sorel Bellak. It was an offshoot of the widely used Thematic Apperception Test (TAT), which was based on Henry Murray's need-based theory of personality. Bellak and Bellak developed the CAT because they saw a need for an apperception test specifically designed for children. The CAT is intended to measure the personality traits, attitudes, and psychodynamic processes evident in prepubertal children. By presenting a series of pictures and asking a child to describe the situations and make up stories about the people or animals in the pictures, an examiner can elicit this information about the child.

Age Group: 3 to 10 years

Time taken for administration: 20 to 45 minutes

Childhood Apperception Test

Draw - A - Person Test:

Description: Draw-A-Person Test (DAP): This test was developed by Florence Goodenough in 1926. This is a psychological projective personality or cognitive test used to evaluate children and adolescents for a variety of purposes. The tester is asked to draw a man or a woman whatever they prefer, after the first figure is drawn i.e. if a boy is drawn then the client is asked to draw the other sex i.e. a girl or a woman. After which the client is inquired as to what he drew and a few related questions are asked for interpretation and scoring purpose.

Aspects such as the size of the head, placement of the arms, and even things such as if teeth were drawn or not are thought to reveal a range of personality traits (Murstein, 1965). The personality traits can be anything from aggressiveness, to homosexual tendencies, to relationships with their parents, to introversion and extroversion (Machover, 1949).

Time taken for administration: 20 to 30 minutes

Advantages:

1. Easy to administer
2. Helps people who have anxieties taking tests (no strict format)
3. Can assess people with communication problems
4. Relatively culture free

Disadvantages:

1. Restricted amount of hypotheses can be developed
2. Relatively non-verbal, but may have some problems during inquiry
3. Little research backing

Draw- A - Person Test (Male figure)

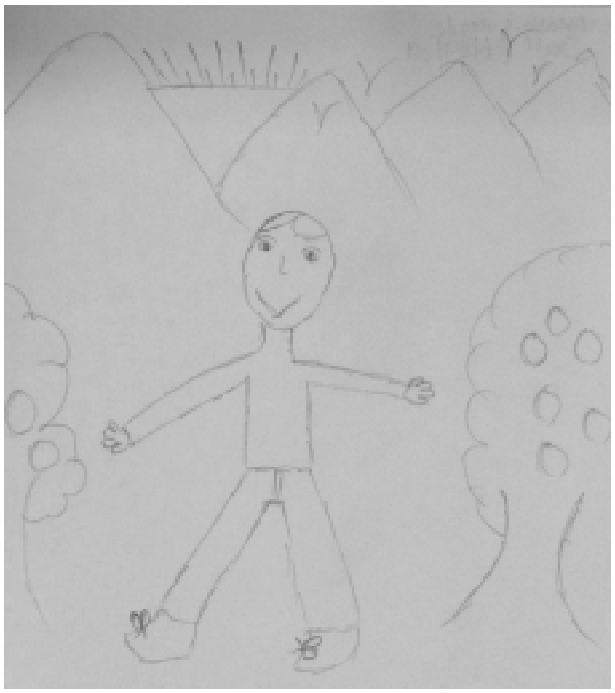
Draw - A - Person Test (Female figure)

House-Tree-Person Test (HTP):

Description: This test was created by Buck in 1948, provides a measure of a self-perception and attitudes by requiring the test taker to draw a house, a tree, and a person. The picture of the house is supposed to conjure the child's feelings toward his or her family. The picture of the tree is supposed to elicit feelings of strength or weakness. The picture of the person, as with other figure drawing tests, elicits information regarding the child's self-concept. The HTP, though mostly given to children and adolescents, is appropriate for anyone over the age of three.

House- Tree - Person Test (House Drawing)

House- Tree - Person Test (Tree Drawing)

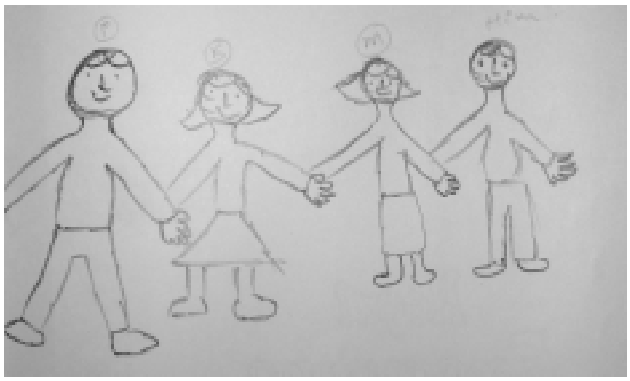


House- Tree - Person Test (Person Drawing)

House- Tree - Person Test Complete House - Tree - Person Test

Kinetic Family Drawing Test:

Description: The Kinetic Family Drawing, developed in 1970 by Burns and Kaufman, requires the test taker to draw a picture of his or her entire family. Figure drawings are projective diagnostic techniques in which an individual is instructed to draw a person, an object, or a situation so that cognitive, interpersonal, or psychological functioning can be assessed. Children are asked to draw a picture of their family, including themselves, "doing something." This picture is meant to elicit the child's attitudes toward his or her family and the overall family dynamics. The KFD is sometimes interpreted as part of an evaluation of child abuse.



Kinetic Family Drawing Test

Behaviour Tests:

Beck's Depression Inventory - Second Edition (BDI - II):

Description: The Beck Depression Inventory (BDI, BDI-II), is created by Dr. Aaron T. Beck, is a 21-question multiple-choice self-report inventory, one of the most widely used instruments for measuring the severity of depression.

Time taken for administration: 10 minutes

Age Group: From 13 to 80 years

Scoring:

0 - 13: Minimal Depression

14 - 19: Mild Depression

20 - 28: Moderate Depression

29 - 63: Severe Depression

Higher total score indicates severe depression

The tests consists of 21 questions, which consists of sadness, pessimism, past failures, loss of pleasure, guilt feelings, punishment feelings, self- dislike, self-criticalness, suicidal thoughts or wishes, crying, agitation, loss of interests, indecisiveness, worthlessness, loss of energy, changes in sleeping patterns, irritability, changes in appetite, concentration difficulty, tiredness or fatigue and loss of interest in sex.

Beck Hopelessness Scale® (BHS®):

Description: This is a 20 item test developed by Aron Beck. This is a powerful predictor of eventual suicide to help you measure three major aspects of hopelessness: feelings about the future, loss of motivation, and expectations. Responding to the 20 true or false items on the Beck Hopelessness Scale® (BHS®), patients can either endorse a pessimistic statement or deny an optimistic statement.

Age Group: 17 through 80 years

Time taken for administration: 5 to 10 minutes self-administered.

Beck Scale for Suicide Ideation® (BSS®):

Description: This was developed by Aron. T. Beck, where it assess the severity of suicidal ideation in an individual.

Age Group: 17 years and older

Time taken for administration: 5 to 10 minutes self-administered.

Bender Visual-Motor Gestalt Test, Second Edition (Bender-Gestalt II):

Description: Originally published in 1938 by Lauretta Bender, M.D., the Bender Visual-Motor Gestalt Test is one of the most widely used psychological tests. The Bender Gestalt provides helpful information in preschool screening as well as geriatric assessment. And it can offer insight into many conditions, including ADHD, mental retardation, giftedness, learning disabilities, autism, and Alzheimer's disease.

Time taken for Administration: No time Limit

Age Group: 4 to 85 years old

Condition Specific tests:

Autism: The psychological tests that are conducted on autistic children and adults are as follows: Autism Diagnostic Interview - Revised, Prelinguistic Autism Diagnostic Observation Schedule, Childhood Autism Rating Scale Second Edition (CARS- 2), Checklist for Autism in Toddlers (CHAT), Gilliam Autism Rating Scale - Second Edition (GARS - 2), etc. Detailed description including other tests are mentioned in the autism chapter i.e. Chapter 4.

Dementia: The psychological tests that are conducted on patients suffering from dementia are as follows: ADAS - Cog, Quality of life - Alzheimer's disease scale (QOL - AD), DEMQOL, Holden Communication Scale, Neuropsychiatric Inventory, Rating Anxiety in Dementia Scale, etc. Detailed description including other tests are mentioned in the dementia chapter i.e. Chapter 5.

PSYCHOTHERAPIES:

Psychotherapy is a general term referred to a therapeutic interaction or intervention plan between a trained psychologist and a client, patient,

caregiver or a group. The psychological problems are addressed depending on the kind of problem or deficit or the severity of the problem. The main goal of psychotherapy is to help the client face the situation, deal with it and come up with solution to the problem during a therapeutic session.

Most forms of psychotherapy use spoken conversation. Some also use various other forms of communication such as the written word, artwork, drama, narrative story or music. Psychotherapy with children and their parents often involves play, dramatization (i.e. role-play), and drawing, with a co-constructed narrative from these non-verbal and displaced modes of interacting. Psychotherapy occurs within a structured encounter between a trained therapist and client(s). Purposeful, theoretically based psychotherapy began in the 19th century with psychoanalysis; since then, scores of other approaches have been developed and continue to be created.

Therapy is generally used in response to a variety of specific or non-specific manifestations of clinically diagnosable and/or existential crises. Treatment of everyday problems is more often referred to as counseling (a distinction originally adopted by Carl Rogers). However, the term counseling is sometimes used interchangeably with "psychotherapy".

While some psychotherapeutic interventions are designed to treat the patient using the medical model, many psychotherapeutic approaches do not adhere to the symptom-based model of "illness/cure". Some practitioners, such as humanistic therapists, see themselves more in a facilitative/helper role. As sensitive and deeply personal topics are often discussed during psychotherapy, therapists are expected, and usually legally bound, to respect client or patient confidentiality. The critical importance of confidentiality is enshrined in the regulatory psychotherapeutic organizations' codes of ethical practice.

The various forms of psychotherapies are listed below:

Cognitive Therapy:

History:

Aaron. T. Beck developed cognitive behaviour therapy and he himself had many difficulties during childhood himself. He used to be ill very often, had many anxieties and phobias, including a blood injury phobia, a fear of public speaking and fear of suffocation. Beck during such trying times used reasoning to alleviate these anxieties and was very successful in overcoming his fears and anxieties. [1]

Principles of Cognitive Therapy:

The practice of cognitive therapy is based on the following principles:

- Changes in thinking, lead to changes in feelings and acting.
- The treatment focuses on the present although attention is paid to the past when required.
- Treatment needs to be short-term, problem focused and goal oriented.
- This therapy has a structured and active approach to treatment.
- Treatment requires a sound and collaborative therapeutic alliance.
- Careful assessment, diagnosis and treatment planning are integral.
- This therapy believes in teaching the client to identify, evaluate and modify their own cognitions, this promotes emotional health and prevents relapse.
- To help the client accurately assess his/ her assess her cognitions, inductive reasoning and Socratic questioning is extremely important.

The Process of Cognitive Treatment:

Cognitive treatment is usually time limited and it usually takes 4 to 14 sessions for solving the issue that the client has. The session usually has 10 steps:

1. To establish the agenda that is meaningful to the client.
2. Determine the intensity of the person's mood.
3. To identify and review the presenting problems.
4. To elicit the person's expectations from the treatment.
5. To educate the client about the therapy and his contribution in the therapeutic alliance.
6. Explain the client about his problems and diagnosis.
7. Establish clear goals.
8. Recommend homework.
9. Summarize the session.
10. Obtain the feedback of the session.

Goal:

The goal of cognitive therapy is to help the client identify the errors that take place in his information-processing system and correct them. To help the client to work through this process the therapist helps the client identify both the client's immediate thoughts (automatic) and global thoughts (core) and beliefs as well as the associated emotions and behaviours.

Cognitive Therapy

Distorted Thinking Patterns: A few of the distorted thought processes are listed below:

- Overgeneralization: The individual here draws conclusions that do not have any justifications or evidences of truth in them. For example: 'I am not a good parent as my son is suffering from muscular dystrophy and might have inherited the disease from me".
- Mental Filters: The individual focuses more on the negative details and does not look at the broad picture.
- Mind Reading: Attributing negative thoughts and reactions to others without checking whether they are present.
- Magnifying or Minimization: The individual draws negative or positive information in excess.

Strategies to Alter the Dysfunctional thoughts:

- Mental and emotional Imagery: This is to help the client adapt to new ways of thinking and feeling. Also to help them envision new ways of coping successful.
- Activity Scheduling: Here the therapist helps the client to try out new behaviours and ways of thinking and encourages them to remain active despite feelings of sadness or anxiety experienced by the person.
- Thought Stopping: This is done by saying "STOP" to oneself either loudly or subvocally. This is to help the client stop the persisting thoughts that are harmful to the patient.
- Self - Talk: Give positive messages and use encouraging phrases to oneself.
- Maintaing Diaries: The patient should be explained the idea of maintain a diary, where he/ she should write down distorted cognitions, emotions and the efforts to make positive changes should also be jotted down.

Cognitive Thought Record Sheet used by psychologists to help the client a clear view of his thoughts:

Where were you?	Emotions or Feelings	Negative automatic thought	Evidence that supports the thoughts	Evidence that does not support the thoughts	Alternative Thoughts	Emotion or feelings
Where were you? What were you doing? Who were you with?	Emotions can be described with one word. For example: Angry, sad, or scared. Rate the emotions: o - 100%	What thoughts were going through your mind? What memories or images were in your mind?	What facts support the truthfulness of this thoughts or image?	What experiences indicate that this thought is not completely true all the time?	Write down a new thought which takes into account the evidence for and against the original thoughts.	How do you feel about the situation now? Rate 0 to 100%

Importance of cognitive Therapy for Patients with Disability:

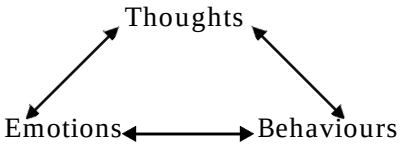
Patients with disability have many emotional and behavioural problems which arise due to negative and distorted thinking of the world and people around them. With incurable disease or deteriorating condition they may have negative view of themselves, they may have feelings of helplessness, hopelessness, may bring on death by not working on improving their condition or giving up easily. For all these reasons cognitive therapy is important for patients with disability. Again it is important to keep in mind that some disabled people function quiet well.

Rational Emotive Behaviour Therapy (REBT):

History: Albert Ellis was the man who developed Rational Emotive Behaviour Therapy; he was born in1913 in Pittusburgh. He like Beck used to be very ill i.e. nephiritis for which he used to be frequently hospitalized. To overcome these difficulties he decided not to be miserable about his condition and maintained strong positive thoughts about his competence and worth. Eventually he was very successful, published over 700 papers and 55 books.

Principal of Rational Emotive Behaviour Therapy:

Rational Emotive Behaviour Therapies core belief is that although thoughts are emphasized emotions, thoughts and behaviours are interrelated and inseparable. [2]



The widely used model of Rational Emotive Behaviour Therapy is the ABCDEF Model:

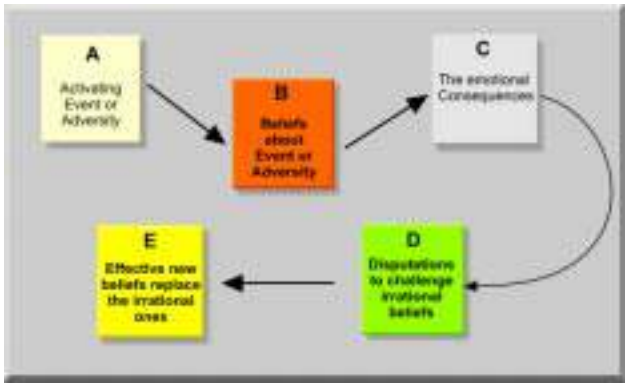
A: Activating Event

B: Belief which may be rational or irrational

C: Consequence of the belief

D: Dispute i.e. questioning the belief whether it is rational or irrational

E: Effective i.e. effectively describing the outcomes



ABCDE Model of Rational Emotive Behaviour Therapy

Goals:

Rational Emotive Behaviour Therapy is a goal oriented system which focuses on outcomes such as reduction in symptoms and changes in beliefs. It helps people to learn to accept themselves and experience greater happiness in life

REBT Strategies to Dispute Irrational Belief: A few strategies are mentioned below: [3]

- **Empirical Disputes:** This focuses on the accumulation of evidence.
- **Rational Alternative belief:** Taking up an alternative rational belief.
- **Humorous disputed:** Disputing the irrational beliefs in a lighthearted way.
- **Didactic style:** This is an educational, explanatory and efficient way in giving appropriate information for developing rational beliefs.

Importance of Rational Emotive Behaviour Therapy for people with Disability:

This therapy believed that an individual's thoughts, emotions and behaviours are inter-related and if they have an irrational belief then it would affect the person in his overall development. It is important for the patient to view his condition positively and should be motivated to work upon and improve his condition; this would be only possible when the negative irrational beliefs are changed to positive rational belief.

Behaviour Therapy:

History: Behaviour Therapy was first developed during the 1950s and 1960s. Many psychologists and physiologists have contributed to the development of this therapy: B.F. Skinner (Operant

Conditioning), Ivan Pavlov (Classical Conditioning), John Watson, Joseph Wolpe, Albert Bandura (Modelling), John Dollard and Neal Miller. [4]

Five Models of Behaviour Therapy:

1. **Applied behaviour Analysis: This was derived by B.F. Skinner. ABA whether they are observed or taught they function on 3 aspects:**
   - **Antecedent (A) -** what triggered a behaviour or what happens before the behaviour,
   - **Behaviour (B) -** the behaviour itself, and
   - **Consequence (C) -** what happens after the behaviour

ABC Model of Behaviour Therapy

There is 47 % of success rate of applied behaviour analysis working in Autistic Patients.

2. **Neo - behaviorism:** This takes up Ivan Pavlov's classical conditioning theory and focuses on the process of conditioning and learning responses.
3. **Social Learning Theory:** This theory is based on Bandura's research and this approach seeks to understand to interaction of cognitive, behavioural and environmental factors in shaping behaviour.
4. **Cognitive Behaviour Therapy:** This approach looks at how cognitions shape behaviours and emotions. This treatment makes use of both cognitive and behavioural strategies to effect change.
5. **Multimodal Therapy:** This theory was developed by Arnold Lazarus, which is based on principles of behaviour therapy but systematically integrates strategies from a wide range of approaches

Principles of Behaviour Therapy:

- Behaviour is learned and acquired largely through modelling, conditioning and reinforcement.
- Behaviour has a purpose.
- Behaviour therapy seeks to understand and change behaviour.
- Education, promoting a new learning and transfer of learning to the natural environment.
- Focus of the therapy should be the present.

Strategies of Behaviour Therapy are incorporated in Cognitive Behaviour Modification below:

Cognitive Behaviour Modification:

Miechenbaum in 1993 developed cognitive behaviour modification in an effort to integrate psychodynamic and cognitive treatment system with the technology of behaviour therapists.

Three assumptions of Cognitive Behaviour Therapy are:

1. **Constructive Narrative:** This suggests that people construct their own reality and reality is a product of personal meaning.
2. **Information Processing:** People experience negative emotions because they distort the reality as a result of cognitive errors and misperception.
3. **Conditioning:** Cognitions are viewed as covert behaviours that have been conditioned

Strategies of Cognitive Behaviour Modification: A few strategies are mentioned below:

1. **Behaviour Rehearsal:** This strategy gives people an opportunity to practice a challenging task. The rehearsal might involve role-play, tape-recording or rehearsing in front of the mirror. This strategy is extremely useful for people who have fear speaking in public or with children who lack social interaction like that seen in autistic children, etc.
2. **Reinforcements:** The use of a reward or reinforcement can encourage behavior change, enhance learning and solidify gains. There are different forms of rewards, like praises, a toy, candy, T.V time, play time, etc. For example: A child when shows desired behaviour is praised or a star is given on his hand, or a food item that he enjoys.
3. **Activity Scheduling:** Planning activities that are rewarding and provide a sense of accomplishment can help people in many ways. Having a schedule or a time -table can help give patients focus and direction. For

example: A child suffering from muscular dystrophy and who is not cooperative during physical therapy is rewarded if he sticks to the time-table.

4. **Visual Imagery:** This seen to be very effective in patients, where they are asked to visualize the activity that is their chief goal and they want to achieve. For example: A spinal cord injury patient who is unable to walk is asked to visualize and see himself walking, this motivates the patient and help him set goals to achieve what he had visualized.
5. **Aversion therapy:** This therapy is based on the principle that undesirable behaviours with a negative experience can be powerful motivator for a change in the behaviour. For example: Time-outs given to a child who throws temper tantrums.
6. **Diaphragmatic Breathing:** This is based on taking slow, deep breaths and focusing on the breathing process, this can be very calming and also sleep induced. This can be helpful for people with sleep disturbances especially seen in patients with stroke and dementia.

Psychoanalytic Therapy: Freud formulated this theory.

According to Freud, the mind can be divided into two main parts:

Psychoanalytic Theory of Personality

1. The conscious mind includes everything that we are aware of. This is the aspect of our mental processing that we can think and talk about rationally. A part of this includes our memory, which is not always part of consciousness but can be retrieved easily at any time and brought into our awareness. Freud called this ordinary memory the preconscious.
2. The unconscious mind is a reservoir of feelings, thoughts, urges, and memories that outside of our conscious awareness. Most of the contents of the unconscious are unacceptable or unpleasant, such as feelings of pain, anxiety, or conflict. According to Freud, the unconscious continues to influence our behavior and experience, even though we are unaware of these underlying influences.[5]

Basic system of belief:

- Id, Ego and Superego: The three parts of the personality.
- Stages of Development: Oral, anal, phallic, latency period and genital.
- Defence mechanisms: Repression, projection, reaction formation, fixation, regression.
- Dreams: Dreams are viewed as the gateway to the unconscious.
- Catharsis: The release of pent-up emotion.
- Corrective Emotional Experience: The therapist helps clients alter their self-perceptions and behaviour.

Treatment Procedures:

- Therapy is long-term and focuses on exploring unconscious issues through interpretation, dream analysis, free association, transference and other methods. [6]
- **Detachment:** The therapist is detached, objective and neutral so that the client can project onto the therapist things from the client's unconscious.
- **Transference:** Traditionally, transference is a projection of unconscious desires onto the therapist.
- **Counter transference:** These are projections the therapist makes onto the client.

Existential Therapy:

There are many contributors to this therapy: Victor Frankl, Rollo May, Irvin Yalom, et al,. The theory underlying existential therapy focuses not on the treatment processes but on the universal issues that people go through. Through their understanding of these issues, clinicians can connect with people at a very deep and personal level and help them change their lives so that they offer more meaningful life. [7]

Existentialists believe that human condition is

difficult one. Life has no inherent meaning and is repellent with sadness and loss. They believe that the following aspects of the human conditions are typically at the root of emotional difficulties: inevitability of death, existential alienation, meaninglessness of life, anxiety and guilt. These are some aspects that the existentialists have spoken about and they believe in. [8]

Goal:

The fundamental goal of existential therapy is helping people find value, meaning and purpose to their lives. The purpose of psychotherapy is not to cure the client but to help increase the level of awareness of what they are doing and to get them out of the victim role. Existential therapists help people confront their deepest fears and anxieties about the inevitable dimensions of life.

Importance of Existential Therapy for people with Disability:

As people who are suffering from incurable diseases or disorders that do not have any treatment option for it and after visiting many doctors and not receiving proper amount of information or treatment, patients usually stop making goals for themselves and do not find meaning to their life. Hence existential therapy is quiet important as it helps the patient find purpose and meaning to lives.

Person-Centered Therapy (PCT):

Person Centered Therapy is also known as person-centered psychotherapy, person-centered counseling, client-centered therapy and Rogerian psychotherapy. PCT is a form of talk-psychotherapy developed by psychologist Carl Rogers in the 1940s and 1950s. The goal of PCT is to provide patients with an opportunity to develop a sense of self wherein they can realize how their attitudes, feelings and behaviour are being negatively affected and make an effort to find their true positive potential. [9] In this technique, therapists create a comfortable, non-judgmental environment by demonstrating congruence (genuineness), empathy, and unconditional positive regard toward their patients while using a non-directive approach. This aids patients in finding their own solutions to their problems.

The core concepts of Person Centered Therapy:

Rogers (1957; 1959) stated that there are six necessary and sufficient conditions required for therapeutic change:

1. **Therapist-Client Psychological Contact:** a relationship between client and therapist must exist, and it must be a relationship in which each person's perception of the other is important.
2. **Client incongruence or Vulnerability:** that incongruence exists between the client's experience and awareness. Furthermore, the client is vulnerable to anxiety which motivates them to stay in the relationship.
3. **Therapist Congruence or Genuineness :** the therapist is congruent within the therapeutic relationship. The therapist is deeply involved him or herself - they are not "acting" - and they can draw on their own experiences (self-disclosure) to facilitate the relationship.
4. Therapist Unconditional Positive Regard (UPR): the therapist accepts the client unconditionally, without judgment, disapproval or approval. This facilitates increased self-regard in the client, as they can begin to become aware of experiences in which their view of self-worth was distorted by others.
5. Therapist Empathic understanding: the therapist experiences an empathic understanding of the client's internal frame of reference. Accurate empathy on the part of the therapist helps the client believe the therapist's unconditional love for them.
6. Client Perception: that the client perceives, to at least a minimal degree, the therapist's UPR and empathic understanding.

Transpersonal Psychology:

The word "transpersonal" comes from the Latin "trans," meaning beyond and through, and "persona," meaning mask or personality. This is a school of psychology that studies the transpersonal, self-transcendent or spiritual aspects of the human experience. A short definition from the Journal of Transpersonal Psychology suggests that transpersonal psychology "is concerned with the study of humanity's highest potential, and with the recognition, understanding, and realization of unitive, spiritual, and transcendent states of consciousness" [10] Issues considered in transpersonal psychology include spiritual self-development, self beyond the ego, peak experiences, mystical experiences, systemic trance and other sublime and/or unusually expanded

experiences of living. This therapy is commonly used to help patients with depression.

Family Therapy:

Family therapy, also referred to as couple and family therapy, family systems therapy, and family counseling, is a branch of psychotherapy that works with families and couples in intimate relationships to nurture change and development. It tends to view change in terms of the systems of interaction between family members. It emphasizes family relationships as an important factor in psychological health.

Family therapy uses a range of counseling and other techniques including:

- Communication theory
- Media and communications psychology
- Psychoeducation
- Psychotherapy
- Relationship education
- Systemic coaching
- Systems theory
- Reality therapy
- Attachment-focused family therapy

Family Therapy session

The number of sessions depends on the situation, but the average is 5-20 sessions. A family therapist usually meets several members of the family at the same time. This has the advantage of making differences between the ways family members perceive mutual relations as well as interaction patterns in the session apparent both for the therapist and the family. These patterns frequently mirror habitual interaction patterns at home, even though the therapist is now incorporated into the family system.

Group Therapy:

The term group therapy, however, was first used around 1920 by Jacob L. Moreno, whose main contribution was the development of psychodrama, in which groups were used as both cast and audience for the exploration of individual problems by re-enactment under the direction of the leader. Group psychotherapy, like individual psychotherapy, is intended to help people who would like to improve their ability to cope with difficulties and problems in their lives. But, while in individual therapy the patient meets with only one person (the therapist), in group therapy the meeting is with a whole group and one or two therapists. Group therapy focuses on interpersonal interactions, so relationship problems are addressed well in groups.

Goal of Group Psychotherapy:

The aim of group psychotherapy is to help with solving the emotional difficulties and to encourage the personal development of the participants in the group. The therapist (called conductor, leader or facilitator) chooses as candidates for the group people who can benefit from this kind of therapy and those who may have a useful influence on other members in the group.

Group Therapy Session

Different types of Groups:

There are many kinds of groups in the group-psychotherapy field. The techniques used in group therapy can be verbal, expressive, psychodramatic etc. The approaches can vary from psychoanalytic to behavioral, Gestalt or encounter groups. Groups vary from classic psychotherapy groups, where process is emphasized, to psychoeducational, which are closer to a class. Psychoeducational

groups usually focus on the most common areas of concern, notably relationships, anger, stress-management etc. They are frequently more time-limited (10 to 15 sessions) and thus very appealing in a managed care environment. Each approach has its advantages and drawbacks, and the participant should consult the expert which technique matches her / his unique personality.

Benefits:

Group psychotherapy is suitable for a large variety of problems and difficulties, beginning with people who would like to develop their interpersonal skills and ending with people with emotional problems like anxiety, depression, etc. There are support groups for people in the same situation or crisis (e.g. groups for bereaved parents, groups for patients with various disability, etc). Groups are ideally suited to people who are struggling with relationship issues, disability, caregiving, etc. The groups interactions help the participants to identify, get feedback, and change the patterns that are sabotaging the relations. The great advantage of group psychotherapy is working on these patterns in the "here and now" - in a group situation more similar to reality and close to the interpersonal events.

Clinical Music Therapy:

In music therapy improvisation is defined as a process where the client and therapist relate to each other. The client makes up music, musical improvisation, while singing or playing, extemporaneously creating a melody, rhythm, song, or instrumental piece. In clinical improvisation, client and therapist (or client and other clients) relate to one another through the music. Improvisation may occur individually, in a duet, or in a group. The client may use any musical or nonmusical medium within his or her capabilities. Musical media includes voice, body sound, percussion, and string, wind, and keyboard instruments. Nonmusical media can consist of images, titles, and stories.

Goals of Music Therapy :

According to Bruscia (1998), clinical goals that can be achieved through improvisation are as follows:

1. Establish a nonverbal channel of communication, and a bridge to verbal communication
2. Provide a fulfilling means of self-expression and identity formation
3. Explore various aspects of self in relation to others
4. Develop the capacity for interpersonal intimacy
5. Develop group skills
6. Develop creativity, expressive freedom, and playfulness with various degrees of structure
7. Stimulate and develop the senses
8. Play, on the spot, with a decisiveness that invites clarity of intention
9. Develop perceptual and cognitive skills

Music Therapy

Therapeutic Techniques:

- Imitating is a basic technique of empathy in which the music therapist copies or repeats a client's response, after the response has been displayed. The music therapist focuses on any sound, rhythm, interval or even facial expression.
- Reflecting is a technique in which the music therapist expresses the same moods or feelings which have been presented by the client.
- Rhythmic grounding is implemented by establishing a steady beat or rhythm, supporting the client's improvisation. The use of a rhythmic ostinato is an example of rhythmic grounding.
- Dialoguing is a process in which the music therapist and the client communicate through their improvisations. Lastly, accompanying is a technique in which the music therapist supports the client's improvisation by giving an accompaniment that consists of rhythm, melody, and chord progressions.

Clinical Music Therapy is extremely useful for people with various diseases and disorders like autism, learning difficulties, dementia, emotional and psychological problems, etc.

COUNSELING CHILDREN:

The goals of the session are very important to be listed when working with children.

There are 4 levels of goals:

Level 1: Fundamental goals: These goals are applicable to all children in therapy.

- To enable the child to feel good about himself or herself.
- To enable the child to deal with painful emotional issues.
- To enable the child to achieve some level of congruence in regards to thoughts, emotions and behaviours.
- To enable the child to change behaviours, that have negative consequences.
- To enable the child to accept his or her limitations and strengths and to feel OK with the.

Level 2: Parents goals: These goals are set by the parents who bring their children for therapy session. They are usually related to the parent's agendas and based on the child's current behaviour. For example if the child throws temper tantrums and is very aggressive then the goal of the parent would be either to reduce or extinguish this negative behaviour.

Level 3: Goals formulated by the psychologist: These goals are formulated by the counselor as a consequence of hypothesis which the counselor may have about why the child is behaving in a particular way. The counselor may have goals of addressing and resolving the child's emotional issues.

Level 4: The child's goals: These goals emerge during the therapy session and are effectively formulated as the child's own goals although the child wouldn't be able to verbalize them. If a counsellor goes into a session with a specific agenda, there are many times when sticking to the agenda will be effective and appropriate. But there is generally a danger of holding rigidly to a pre-determined agenda because the child's own needs might be overlooked rather than addressed.

Attributes of Child and Counsellor Relationship:

- Exclusive: The counsellor should build a good rapport with the client and the child should experience a unique relationship with the counsellor which is not comprised by unwanted intrusion of others such as parents or siblings.
- Safe: The counsellor should create a permissive environment in which the child feels free to act out and to gain mastery over the feelings in safety. The child should feel safe to make disclosures with confidence that doing so will not have repercussions or consequences which may be emotionally harmful or damaging.
- Connecting Link: The child and counsellor relationship should be a connecting link between the child's world and the counsellor. The focus of the relationship is primarily about connecting with the child and staying with the child's perceptions. The child may see the environment in which he lives quiet differently from the way in which his parents see his environment.
- Authentic: The child and counsellor relationship should be authentic i.e. it should be genuine and honest. The authentic relationship allows the child to give up the pretence of being someone she is not and to allow raw inner self to be exposed.
- Confidential: When working with the children the counsellor tries to create an environment where the child feels safe enough to share very private thoughts and emotional feelings. In order for the child to feel safe there should be some amount of confidentiality that is required. There would be times when the child would share information with the counsellor with the counsellor believes needs to be shared with others, for example in case of a child abuse or sexual abuse.
- Non - Intrusive: When working with children the counsellor needs to join with the child in a way which is comfortable for the child. Some counsellors believe in questioning the child and inquiring about the child's family and background, during the joining processes which is a useful way of getting to know the child's world. However asking to many questions could scare the child as he or she may feel intrusion into his or her personal space.

- Purposeful: Children enter into a therapeutic process more willingly and confidently if they know exactly why they are coming to see the counsellor. They need time to prepare themselves for counseling and will usually do so if given prior suitable notice and if told of the reason as to why they are being brought for the counseling session.

OTHER FORMS OF THERAPIES FOR CHILDREN:

Play Therapy:

If play is a child's language, then toys can be thought of as the words. Through play therapy the child can work through their challenges and issues using the toys that they choose, revealing their inner dialogue. Through play the child is able to test out various situations and behaviours in a supportive environment. Unconditional positive regard and acceptance encourages the child to feel safe enough to be able to explore their inner selves without censorship. In this environment children are able to try out different roles, work through conflicting emotions and thoughts, and try to figure out what the world is like. The child is able to form a relationship with the provider, and through this relationship they are able to develop trust, improved self-esteem, and self efficacy.

Play Therapy Session

In non-directive play therapy, the child is in control, within some gently but firmly set limits. Children often feel that they do not have control over situations in their lives. Through play therapy they are able to work through these experiences in an environment that they are able to control. They can make the story be how they want it to be, they are in charge of the outcome. This feeling of control is vital to their emotional development as well as positive mental health. Children are able to use play as a means for developing problem-solving skills, ways to relate to others, expressing their feelings, and working on their behaviours, all at a safe psychological distance from reality.

Furniture and Associated Items:

Play Therapy Room

Toys: Dolls, doll house, doll clothes, doll family, telephones, mirrors, play money, kitchen set, child chairs and bean bags, etc.

Equipments and Materials: Sand tray, clay, play dough, papers, pencils, paints, crayons, glue, scissors, wool, coloured papers, wooden blocks, farm animals, zoo animals, etc.

Dress up Materials: Variety of clothes, jewellery, wigs, swords, doctor's set, masks, etc.

Games: Card games, board games, etc.

Books: Colouring books, drawing books, story books and worksheets, etc.

Art Therapy:

Art therapy for children can provide kids with an easier way to express themselves since children are more naturally artistic and creative. A young child is likely to be more comfortable initially expressing him / herself with some crayons and markers, for example, than he / she is going to be at expressing emotions and feelings through words. A question and answer type of format can be daunting and intimidating for a child, especially when they have to try and explain themselves with their already limited vocabulary. Because of this, art therapy for children can be a much more viable solution for communication than simply having a conversation and talking about things. This can be especially true when it comes to children and traumatic events.

If a child experiences something tragic, that event usually gets buried in their subconscious where it affects them in the future. These types of things are not easy for kids to talk about, especially when there

are deep-rooted emotional issues in play. Through art therapy children can help bring these suppressed emotions to the surface so the art therapist can then focus on healing the child's issue(s). Art therapy can also aid a child in achieving better self-awareness, relief from stress or anxiety, learning disorders, autism, and other traumatic experiences.

Art Therapy Children Supplies:

1. Crayons
2. Pastels
3. Colored pencils
4. Set of brightly colored markers
5. Tempera paints and / or watercolor set
6. Spiral bound pads of art paper / sketch pads
7. Scissors and glue
8. Old magazines

And for Younger Children:

Finger paint set

Play-Dough set

Art Therapy

Strategies of Art Therapy:

- I am the most sad when I remember_____. This is my picture of feeling sad.
- I sometimes get angry when I think about the death. This is my picture of anger.
- These are the things I liked the most about (deceased).
- I get scared when I think of _____. This picture shows my fear.

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Ch.15 Speech Rehabilitation

Dr. Manasi Jani (Speech Therapist).

One of the features that put a human being apart from other animals is the ability to communicate. Human beings communicate through various modes or channels called modalities. The most commonly used modality to communicate is verbal modality or spoken modality. Speech is a result of co-ordination between different systems in the body namely respiration, phonation, resonance and articulation. The organs which actively participate in speech production are the lungs, the larynx (voice box), the vocal tract, the lips, the tongue, the soft palate and all the sinuses. Also, hearing is an important sensory organ which has direct connection to speech production and perception. An audiologist and speech language pathologist is a professional who deals with diagnosis and treatment of disorders related to speech and hearing.

Hearing, speech and language defects can arise due to various neurological problems. Neurological disorders that lead to speech and language disorders can be classified into congenital and acquired causes. Acquired causes can be classified in to static and progressive disorder. The main causes of congenital neurological disorders are cerebral palsy, muscular dystrophy, autism, viral encephalitis and mental retardation. Acquired static causes can be due to stroke, post encephalitis, head injury while the progressive causes are ALS, multiple sclerosis, muscular dystrophy, parkinson's disease to name a few.

The commonest problems seen in children and adults with a neurological defect are:

- **Impaired comprehension -** this includes problems like inablility to understand concepts which normal children or adults of that age can. It also includes problems like inability to follow simple commands like 'close your eyes' or can you show me your nose?'.
- **Impaired expression -** this includes inability to verbally express. The patient may speak a few words to ask for needs or may just use pointing to express themselves. Sometimes, it is observed that the patients use words which do not exist in the language and are meaningless. These are called jargon speech.
- **Impaired pragmatics -** this includes affected social interaction, irrelevant answers to questions, lack of humour and inability to understand metaphorical elements.
- **Impaired swallowing functions -** this category of symptoms have a variety of presentations depending on the stage of swallowing affected. The stages of swallowing are the oral, pharyngeal and esophageal phase. Problems like affected lip seal, poor tongue control, inability to chew properly affect the oral phase of swallowing. Problems like delayed or absent swallow reflex, multiple swallows needed to swallow a single bolus of food are included in the pharyngeal phase. Regurgitation of food after swallowing is included in the esophageal phase of swallowing.
- **Affected speech clarity due to imprecise sound production -** this includes substitution or inability to produce a certain phoneme or sound appropriately.
- **Hypernasality.**
- **Affected loudness, pitch and quality of voice** - every person has a certain speaking pitch. Neurological disorders have a drastic effect on the vocal system too. They can result in reduction in the loudness of voice, change in the pitch, harshness or hoarseness of voice.
- **Monotonous speech.**
- **Affected oromotor structures and functions-** affected lip seal, drooling, affected tongue control, and affected palatal movements.

Role of A Speech Therapist as A Swallowing Therapist:

Swallowing being in a vegetative function is extremely crucial and requires immediate attention. The long term effects of swallowing problems can pose detrimental consequences like aspiration pneumonia and malnourishment. What may look like a mild coughing and choking could have serious implications. Swallowing problems can occur in any age group. Hence, it is of vital importance that these problems be tackled under strict supervision of a qualified speech therapist.

Role of A Speech Therapist as A Voice Therapist:

Larynx or the voice box is an important system in speech production. Hence, speech therapists play an important role in optimizing the vocal parameters. Neurological disorders affect the nerves supplying the larynx thereby affecting the function of the vocal cords. These can be corrected by certain breathing exercises, vocal function techniques or surgery in case speech therapy doesn't work. Also, voice therapy should be considered before considering any patient for a phonosurgery or surgery to improve voice because the efficacy of voice therapy in neurological voice problems like vocal cord palsy is excellent.

Speech Therapy:

1) **Oromotor and PNF exercises** are exercises involving stimulation of jaws, lips, cheeks, tongue and palate. The speech therapist aims to improve the oral musculature and attain optimal function. The adequate function of oral functions is important for swallowing and speech functions.

2) **Hearing defects** are seen in many children with cerebral palsy especially athetoid cerebral palsy children and in adults with peripheral neuropathy. These disorders have to be diagnosed for the degree and type of hearing loss and differentiate a temporary hearing loss from a permanent one. The diagnosis is made using a routine test like pure tone audiometry (PTA) or brain stem evoked responses (BERA) in children who cannot respond to the heard sounds. A sensorineural hearing loss or a hearing loss affecting the inner ear is treated by fitting proper hearing aid and check for auditory perception and discrimination.

3) **Comprehension deficits** are seen in children with mental retardation, autism, cerebral palsy and adults after a stroke or head injury. In these patients the main aim is to achieve near normal comprehensive skills. By pointing tasks like pointing to flash cards, or answering riddles, the patient can be taught to understand the skills which the patient has not achieved or has lost due to the disorder causing it.

   While working on comprehension, it is necessary to ensure that the patient is giving complete attention to the task and hearing the therapist correctly. It is always advised to initiate with simple words and few distracters and then include complex and indirect comprehension tasks with multiple distracters.

4) **Complete loss of speech or restricted speech** is seen in children with delayed speech and language, autism, cerebral palsy and individuals with stroke, or head injury. While dealing with children with speech and language problems, it is always advisable to work on language in the most naturalistic way possible as that facilitates better generalization of abstract concepts. Using flash cards and real objects to elicit responses is the widely used method to teach language.

   The child can be shown a card or an object and asked "what is this"?. If the child responds appropriately, the clinician target to achieve elaborate expression for eg response from a ball to this is a red round ball. If the child is unable to respond at all, the clinician acts as a model and the child repeats after the clinician. This technique is called modelling. Also, expansion and extension of responses can be used to achieve more specific and detailed responses. More opportunities should be provided to the children to use the learnt target.

   In adults with expressive deficits, various techniques like word retrieval tasks, melodic intonation technique etc can be used depending on the baseline evaluation. Many a times when a patient presents with severe expressive deficits, the functional communication should be considered.

5) Many of the children with cerebral palsy, autism, head injury and adults with stroke, parkinson's disease, ALS, multiple sclerosis, muscular dystrophy present with affected speech intelligibility called **dysarthria.**

   Typical **signs** seen are imprecise or weak consonants, slurring, reduced loudness of voice and inability to speak many words in one breath to name a few.

   The speech therapist **aims** to achieve near normal speech intelligibility within patient's impairment limits. Also, working on the monotony in speech and stress patterns, naturalness in speech can be achieved.

Augmentive and Alternative Communication in Neurological Patients:

In our daily clinical practise, we come across very severely affected patients or patients with poor prognosis in speech production. Also, when we encounter patients with progressive neurological conditions like motor neuron disorders, multiple sclerosis, dementia etc, a long term solution should be kept in mind. Therefore, a regular follow up with the speech therapist and evaluation of the deterioration is necessary.

In patients with a chronic brain damage or severe level of deterioration, verbal communication cannot be solely depended upon. In such cases, an alternated mode of communication or another mode which will augment the current verbal production should be considered. According to the strengths of the patient i.e patient's level of understanding, patient's level of performance and motivation, the appropriate mode of communication can be selected. Using drawing board, using pantomime or using computer assisted technologies are the widely used systems today. Also, for severely affected individuals like patients in vegetative state, head pointers or eye blinks to indicate needs are used. In patients with severe dysarthria, a specialized system which converts typed words in to spoken signals is advised. Hence, along with technological advancement, the function of AAC has gained momentum.

Swallowing Therapy:

This is one of the most critical areas a speech therapist works into. Diagnosis in terms of site of lesion and the stage affected is crucial as the treatment plan changes according to the stage affected.

A patient having an oral stage of swallowing affected has to focus more on strengthening his oromotor areas and achieve optimal functioning in order to manipulate the bolus in the mouth and achieve proper chewing. Certain manoeuvres like supraglottic swallow; effortful swallow, Shaker's exercise and Mendelson's manoeuvre are helpful.

Few bodily postural changes like chin back, chin tuck and head tilts as recommended by the speech therapist is very helpful. Also, many a times patients find it easier to swallow a certain consistency like liquids or solids better than the other. Hence, bolus modification which suits the patients and which also fulfils the nutritional needs of the patient can be made by the speech therapist.

At every point of time in the treatment, airway safety has to be monitored closely and chances of silent aspiration and aspiration pneumonia should be minimized. In patients having a chronic or long standing dysphagia, an enteral feeding like nasogastric tube or peritoneal esophageal gastrostomy can be considered along with oral trial feeds.

Any neurological condition having bulbar symptoms reveals speech, language and cognitive deficits as one of the first indications. Therefore, the role of a speech language pathologist diversifies in to a that of a voice therapists, hearing specialist and swallowing therapist apart from a traditional speech therapist.

Section 5

An Overview on Stem Cells and Stem Cell Therapy

Ch.16 An Overview on Stem Cells and Stem Cell Therapy

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An Overview on Stem Cells and Stem Cell Therapy

Regenerative medicine is a newly evolving branch of modern medicine that deals with cell based therapies which use healthy cells cultured in the laboratory to replace damaged cells in adult organisms to treat disease. This could therefore potentially hold the key for addressing ailments which currently have no proven treatments or cures, such as, neurological disorders (spinal cord injury, cerebral palsy, brain stroke, muscular dystrophy, Alzheimer's disease, multiple sclerosis, etc.), diabetes, cardiovascular disorders, bone disorders, hematopoietic disorders, cancers, hepatic, renal and dermatological disorders.

One of the building blocks of this therapy is stem cells. Regenerative medicine aims to repair or re-grow parts or tissues which are lost as a consequence of disease or injury. Stem cells have the capability to multiply manifolds and convert or differentiate into any specialized cell types of the body. Hence, the potential of these invaluable assets could even be projected as far as, sometime in the near future, to replace organ transplantation.

Depending on the source, the potency or plasticity of stem cells varies. Stem cells procured from the 5-6 day embryo (usually from wasted or excess fertilized embryos from IVF clinic), referred to as embryonic stem cells, have theoretically the capacity to give rise to the whole embryo and cells of all the germ layers (pluripotent).However, they are surrounded by hordes of ethical issues regarding the source of these cells. Also, formation of "teratomas" is a serious possibility in the long-term with these cells.

In order to bypass the ethical and medical issues associated with embryonic and fetal stem cells, researchers and clinicians have researched and developed other sources of stem cells, such as haematopoietic and mesenchymal stem cells from the bone marrow and umbilical cord, stem cells from the adipose tissue, olfactory ensheathing, endometrium, neural stem cells, etc., which have varying potencies for differentiating into different cell types. A body of work has been ongoing on the use of these cells, in various specialties and disorders.

The nervous system is like the central processing unit of the animal body. In humans, it is more evolved and specialized. Since, disorders and injuries affecting the nervous system lead to irreparable damage and disability, this area has become a major focus point in the arena of regenerative medicine. The hope is that by using the plasticity of the nervous system and combining it with the regenerative potential of the stem cells it would be possible to evolve definitive treatments for degenerative and traumatic disorders of the nervous system.

Basics of Stem Cells

Every cell in the human body can be traced back to a fertilized egg that came into existence from the union of the egg and the sperm. The body is made up of over 200 different types of cells. All of these come from a pool of stem cells in the early embryo. During early development as well as later in life, the stem cells give rise to the specialized or differentiated cells that make up our body. Over the past 2 decades scientists have been gradually deciphering the processes by which unspecialized stem cells become the different types of specialized stem cells. Stem cells can regenerate themselves or produce specialized cell types. This is the property that makes them appealing as a method for creating medical treatment that can replace lost or damaged cells. In this chapter we will look at some of the fundamental basic properties of Stem cells.

What Are Stem Cells?

A stem cell is defined by two properties. First, it is a cell that can divide indefinitely, producing a population of identical offspring. Second, stem cells can, on cue, undergo an asymmetric division to produce two dissimilar daughter cells. One is

identical to the parent and continues to contribute to the original stem cell line. The other varies in some way. This cell contains a different set of genetic instructions (resulting in an alternative pattern of gene expression) and is characterized by a reduced proliferative capacity and more restricted developmental potential than its parent. Eventually a stem cell becomes known as a "progenitor" or "precursor" cell, committed to producing one or a few terminally differentiated cells such as neurons or muscle cells. (1)

Developmental Hierarchy in Stem Cell (SC) Compartment:

There exists a hierarchy in the stem cell compartment, depending on their 'potency' or fate restriction.1) Totipotent stem cells give rise to embryonic as well as the extra embryonic tissue. This means, it has the capacity to form the whole of the embryo, including the placenta. The physiological totipotent stem cell is a fertilized oocyte (zygote) or first blastomere which comprises of the 8 cell stage. The artificial counterpart is a clonote obtained by somatic cell nuclear transfer (SCNT) to an enucleated oocyte.2) Pluripotent stem cells in turn have the capacity to give rise to cells of all the three germ layers of the embryo, i.e., endoderm, mesoderm and the ectoderm. Pluripotent stem cells are cells from the inner cell mass of the blastocyst (ICM), epiblast (EPSC) and SC obtained as immortalized cell lines - blastocyst derived embryonic stem cells (ES) and Primordial Germ Cell-derived embryonic germ cells (EG). 3) Multipotent stem cells give rise to cells of one of the germ cell layers only, either ecto-, meso- or endoderm. Sources range from 8 day old embryo to adult bone marrow. 4) Monopotent stem cells are tissue-committed stem cells that give rise to cells of one lineage, e.g., hematopoietic stem cells, epidermal stem cells, intestinal epithelium stem cells, neural stem cells, liver stem cells or skeletal muscle stem cells. (2)

Though the above classification has evolved over decades, understanding of the potency of these cells are everchanging. Many of these cells, which were earlier considered to be multipotent, have shown limited pluripotent properties. Also, transdifferentation of monopotent / unipotent cells by external stimulation or manipulation have shown that these classifications, based on fate restriction or potency, are fast becoming redundant.

Classification of Stem Cells

Stem cells are classified as embryonic stem cells, umbilical cord stem cells and adult stem cells on the basis of their origin.

Embryonic Stem cells:

Embryonic stem cells are pluripotent in nature which are derived from the inner cell mass (ICM) of 5 to 7 day blastocyst, obtained from IVF clinics. (3)

Developmental studies in mouse revealed that the fertilized oocyte, the zygote, has the capacity to form the whole embryo. It further divides progressively to give rise to an 8 cell staged, 16 celled, 32 celled blastomere and then finally the blastocyst.

The blastocyst is demarcated into the outer transparent trophoblast (which forms the extra embryonic tissue / the placenta) and the Inner cell mass (ICM) which is a 30-34 celled clump.

The ICM ultimately gives rise to the three germ layers and subsequently the whole embryo. Hence, the inner cell mass is the source for the derivation of the embryonic stem cells, which has lost the "totipotency" of the zygote, but is now "pluripotent".

The potential of the embryonic stem cell to form the "germ layers" & its capacity to self renew indefinitely as well as its ability to form any cell type of the body, has led to opening up of this field widely, not only with respects to its use in regeneration, but has thrown up debates regarding ethics and legalities.

However, even before the first embryonic stem cell line was derived in 1981, embryonal carcinoma cells derived from germline tumors called "teratocarcinomas" were widely studied. After transplantation to extra-uterine sites of appropriate mouse strains, these "funny little tumors" produced benign teratomas or malignant teratocarcinomas. (4)

Uses of Embryonic Stem Cells:

1. Embryonic stem cells as cellular models

Experiments designed to understand gene function in the context of an organism require genetic strategies. Enhancer and promoter traps, gene traps, random activation of gene expression (RAGE) and genome-wide cell-based knockout (GECKO) represent genome-wide strategies to identify,

isolate, or determine gene function. Because of gene-targeting techniques, transgenic mice have also proven critical to the creation and evaluation of some models of human disease. Embryonic stem cell lines have proven to be useful mediums for genetic manipulation, for understanding developmental processes and correction of genetic defects. (5)

2. Embryonic stem cells in pharmacology and embryotoxicology

Stem cells also represent a dynamic system suitable to the identification of new molecular targets and the development of novel drugs, which can be tested in vitro for safety or to predict or anticipate potential toxicity in humans. (6)

Human ES cell lines may, therefore, prove clinically relevant to the development of safer and more effective drugs for human diseases. Three aspects are relevant to this issue. 1) At present, insufficient methods exist in some areas of in vitro toxicology to predict target organ toxicity. 2) In embryotoxicology, interspecies variation complicates data analysis, and human cell systems may enhance the identification of hazardous chemicals. 3) Human ES-derived cells cultured in vitro may reduce the need for animal testing in pharmacotoxicology.

The application of hES cells in pharmacology and embryotoxicology could have a direct impact on medical research, but to date, such an approach has primarily been used with mouse ES cells.

3. In stem cell based therapies:

The in vitro developmental potential and the success of ES cells in animal models demonstrate the principle of using hES-derived cells as a regenerative source for transplantation therapies of human diseases. Before transfer of ES-derived cells to humans can proceed, a number of experimental obstacles must be overcome. These include efficient derivation of human ES cells in the absence of mouse feeder cells, and an understanding of genetic and epigenetic changes that occur with in vitro cultivation. It will be necessary to purify defined cell lineages, perhaps following genetic manipulation, that are suitable for cell-based therapies. If manipulated, then it will be important to guard against karyotypic changes during passaging and preparation of genetically modified ES-derived cells. Once introduced into the tissue, the cells must function in a normal physiological way. Finally, assurances against the formation of ES cell-derived tumors and donor/ recipient immunocompatibility are additional requirements of stem cell-based therapies. As pointed out, significant progress has been made in the isolation of defined cell lineages in mouse, and important advances have already been seen with hES cells. Before therapeutically applicable, any ES-based treatment must, however, show limited potentials for toxicity, immunological rejection, or tumor formation, and at present, human ES cell research has not reached this threshold.

The availability of human ES cells, however, represents an extraordinary opportunity for cell transplantation that may be applicable to a wide range of human ailments. Three properties make ES cells relative to adult stem cells very attractive for replacement therapies.1) Human ES cells can be grown indefinitely in culture.2) ES cells can be genetically manipulated, and loss of function genes (e.g., CTFR) can theoretically be repaired by the introduction of transgenes into ES cells either by random transgenesis or through gene targeting. 3) Numerous differentiation protocols have already been established that permit the generation of almost any cell type, either through the use of established culture conditions or when coupled with genetic manipulations. In theory, hES cells could be applied to a wide range of human ailments, but the proof of principle has largely come from the use of mouse ES cells.(7-8)

Adult Stem Cells

Adult stem cells are pluripotent, clonogenic, self renewing, having ability to differentiate into the mature cell of it resident environment and also, may have transdifferentiating abilities.

Adult stem cell niches have been found in most organs of the human body,

eg. liver, brain, bone marrow, adipose tissue, heart, etc. The primary role of these adult stem cells is initiation of repair process in the organ following an injury. These cells have been, in practicality, difficult to obtain due to the following reasons:

1) Inaccessibility and small numbers (e.g. neural stem cells)
2) Lack to markers for characterization and isolation of the "stem cell population" from various organs.(9)

The field of Regenerative medicine, which got

opened up widely following the discovery of the embryonic stem cells, is now in search of the "almighty" pluripotent stem cell, following ethical, legal and medical questions raised against the ES cell research and therapeutic use.

The search has now been directed towards adult stem cell niches, which pose a non controversial and safe option for use in human subjects. However, the debate over its pluripotency is ongoing and the fields as well as the concept of adult stem cell plasticity have been extremely dynamic.

Bone Marrow Derived Cells

Bone marrow is the most accessible and most studied source of adult stem cells. Different types of stem cells have been found to be present in the bone marrow, which differ in their potential to differentiate and form cells from one or more germ layers.

Initially, the bone marrow was thought to contain only haematopoietic stem cells. The excitement regarding HSCs diminished after it was found to have limited potency. However, increasingly, evidence is pouring in regarding the heterogenous population of cells having varying plasticity.

Potential Pluripotent Stem Cells candidates identified in adult tissues (especially, bone marrow)

1) Mesenchymal Stem Cells (Multipotent Mesenchymal Stromal Cells):

Human mesenchymal stem cells (MSCs) are thought to be multipotent cells that have the potential to differentiate into multiple lineages including bone, cartilage, muscle, tendon, ligament fat and a variety of other connective tissues. Indeed, marrow-derived cells seem to retain a remarkable plasticity, since they have much wider differentiation potential than previously thought. Marrow cells have been reported to contribute to angiogenesis, somatic muscle development, liver regeneration, and the formation of central nervous system cell types. It is likely that MSC may be contaminated by other populations of primitive non-hematopoietic stem cells. This possibility should be considered whenever a "transdedifferentiation" of MSC into cells from other germ layers is demonstrated. Because various inconsistencies have come to light in the field of MSC research, the International Society for Cellular Therapy recently recommended avoiding the name of MSC stem cells and changing it to multipotent mesenchymal stromal cells instead. (10)

2) Multipotent Adult Progenitor Cells (MAPC):

MAPC are isolated from BM as well from various adult organs as a population of CD45 GPA-A- adherent cells and they display a similar fibroblastic morphology to MSC. Interestingly MAPC are the only population of BM derived stem cells that have been reported to contribute to all three germ layers after injection into a developing blastocyst, indicating their pluripotency. (11) The contribution of MAPC to blastocyst development, however, requires confirmation by other, independent laboratories

3) Marrow-isolated adult multilineage inducible (MIAMI) cells:

This population of cells was isolated from human adult BM by culturing BM MNC in low oxygen tension conditions on fibronectin . MIAMI cells were isolated from the BM of people ranging from 3- to 72-years old. Colonies derived from MIAMI cells expressed several markers for cells from all three germ layers, suggesting that, at least as determined by in vitro assays, they are endowed with pluripotency. However, these cells have not been tested so far for their ability to complete blastocyst development. The potential relationship of these cells to MSC and MAPC is not clear, although it is possible that these are overlapping populations of cells identified by slightly different isolation / expansion strategies

4) Multipotent Adult Stem Cells (MACS):

These cells express pluripotent-state-specific transcription factors (Oct-4, Nanog and Rex1) and were cloned from human liver, heart and BM-isolated mononuclear cells. MACS display a high telomerase activity and exhibit a wide range of differentiation potential. Again the potential relationship of these cells to MSC,MAPC and MIAMI described above is not clear, although it is possible that these are overlapping populations of cells identified by slightly different isolation / expansion strategies.

5) Very Small Embryonic Like (VSEL) Stem Cells:

Recently, a homogenous population of rare (~0.01% of BM MNC) Sca-1+ lin- CD45- cells was identified in murine BM. They express (as determined by RQ-PCR and immunhistochemistry) markers of pluripotent stem cells such as SSEA-1, Oct-4, Nanog

and Rex-1 and Rif-1 telomerase protein (12) Direct electron microscopical analysis revealed that VSEL (2-4 µm in diameter) display several features typical for embryonic stem cells such as i) a large nucleus surrounded by a narrow rim of cytoplasm, and ii) open-type chromatin (euchromatin). Interestingly, these cells despite their small size possess diploid DNA and contain numerous mitochondria. VSEL, however, do not express MHC-1 and HLA-DR antigens and are CD90- CD105- CD29.

Umbilical Cord Stem Cells

Umbilical cord blood stem cells can be obtained from the umbilical cord immediately after birth. Like bone marrow, umbilical cord blood is another rich source of hematopoietic stem cells, since 1988. The blood remaining in the umbilical vein following birth contains a rich source of hematopoietic stem and progenitor cells, has been used successfully as an alternative allogeneic donor source to treat a variety of pediatric genetic, hematologic, immunologic, and oncologic disorders. Fresh cord blood is also a promising source of non-hematopoietic stem cells. Among others, it contains endothelial cells, MSCs and unrestricted somatic stem cells (USSC). These hematopoietic stem cells are less mature than those stem cells found in the bone marrow of adults or children.

Umbilical cord blood contains circulating stem cells and the cellular contents of umbilical cord blood appear to be quite distinct from those of bone marrow and adult peripheral blood. The characteristics of hematopoietic stem cells in umbilical cord blood have recently been clarified. The frequency of umbilical cord blood hematopoietic stem cells equals or exceeds that of bone marrow and they are known to produce large colonies in vitro, have different growth factor requirements, have long telomeres and can be expanded in long term culture. Cord blood shows decreased graft versus host reaction compared with bone marrow, possibly due to high interleukin-10 levels produced by the cells and/or decreased expression of the beta-2-microglobulin. Cord blood stem cells have been shown to be multipotent by being able to differentiate into neurons and liver cells.

The advantages of using cord blood as a source of stem cells are:

1) It is a non-invasive source and can be obtained from the umbilical cord immediately after birth.
2) Available in vast abundance; thousands of babies are born each day and the umbilical cord and placenta are discarded as waste.
3) Despite its high content of immune cells, it does not produce strong graft-versus-host disease
4) Therefore, cord blood grafts do not need to be as rigorously matched to a recipient as bone marrow grafts. A 4 out of 6 match is sufficient for clinical use.

Hence, cord blood has recently emerged as an alternative source of hematopoietic stem cells for treatment of leukemia and other blood disorders.

All over the world, innumerable cord blood banks have cropped up for storage of umbilical cord stem cells. These are generally either pure public banks or private banks. There are certain banks which offer both types of banking (mixed type). Umbilical cord stem cells banks also differ in the type of biological material that they store. Some banks only store the cord blood (from the umbilical vein) which predominantly carries the haematopoietic stem cells. Increasingly, banks have started storing pieces of the placenta and cord, which are a rich source of mesenchymal stem cells.

MECHANISM OF ACTION

Stem cells are instrumental in the formation of new tissues and thereby promoting repair and regeneration. Their role, in the normal wear and tear of the body, appears to be assistance of repair and maintenance of normal tissue structure and function. Recreation of this ability in vitro as well in animal models of various diseases is the basis of devising therapeutic modalities for degenerative disorders through remodeling of the injured tissues. Cell-based therapy could therefore potentially be used to treat a wide array of clinical conditions where cellular damage is the underlying pathology.

More importantly, the use of adult stem cells as opposed to human embryonic stem cells for therapy avoids ethical problems and has two additional advantages:1) Adult stem cells can be isolated from patients, and this overcomes the problem of immunological rejection and 2) The risk of tumor formation is greatly reduced as compared to the use of embryonic stem cells.(13)

Plasticity, Pluripotency and Production

While pluripotency and plasticity are considered properties of early ESC, adult stem cells are

traditionally thought to be restricted in their differentiation potential to the progeny of the tissue in which they reside. However, a remarkable plasticity in differentiation potential of stem cells derived from adult tissues has been seen. (14)

The events underlying stem cell plasticity could relate to a variety of mechanisms such as dedifferentiation, trans-differentiation, epigenetic changes, and / or cell fusion. Rerouting of cell fate may result from the multistep process known as dedifferentiation where cells revert to an earlier, more primitive phenotype characterized by alterations in gene expression pattern which confer an extended differentiation potential.

Another mechanism put forward to explain stem cell switch to a novel phenotype is a process known as trans-differentiation. Cells may differentiate from one cell type into another within the same tissue or develop into a completely different tissue without acquiring an intermediate recognizable, undifferentiated progenitor state. (15) or may undergo cell fusion resulting in nuclear reprogramming and changes in cell fate. (16,17) It is now recognized that adult stem cells from bone marrow may fuse with cells of the target organ. So far, bone-marrow-derived cells were shown to form fusion heterokaryons with liver, skeletal muscle, cardiac muscle, and neurons. There is evidence that such fused cells become mono-nucleated again, either by nuclear fusion or by elimination of supernumerary nuclei.(18)

The Paracrine Effect

Exploration of the various cellular processes occurring (both during normal physiology as well as after tissue injury) in the process of stem cell renewal and differentiation, suggests that stem cell treatment or transplantation of stem cells remodels and regenerates injured tissue, improves function, and protects tissue from further insult. Stem cells transplanted into injured tissue express paracrine signaling factors including cytokines and other growth factors, which are involved in orchestrating the stem cell-driven repair process through increasing angiogenesis, decreasing inflammation, preventing apoptosis, releasing chemotactic factors, assisting in extracellular matrix tissue remodeling and activation of resident / satellite cells which is discussed further in details.

Increased Angiogenesis

Stem cells produce local signaling molecules that may improve perfusion and enhance angiogenesis to chronically ischemic tissue. Although the particular growth factors contributing to this neovascular effect remain to be defined, the list includes vascular endothelial growth factor (VEGF), hepatocyte growth factor (HGF), and basic fibroblast growth factor (FGF2). (19,20)

Decreased Inflammation

Stem cells appear to attenuate infarct size and injury by modulating local inflammation. When transplanted into injured tissue, the stem cell faces a hostile, nutrient-deficient, inflammatory environment and may release substances which limit local inflammation in order to enhance its survival. Modulation of local tissue levels of pro-inflammatory cytokines by anti-inflammatory paracrine factors released by stem cells (such as IL-10 and TGF-?) is important in conferring improved outcome after stem cell therapy. (21)

Anti-Apoptotic and Chemotactic Signaling

Stem cells in a third pathway promote salvage of tenuous or malfunctioning cell types at the infarct border zone. Injection of MSC into a cryo-induced infarct reduces myocardial scar width 10 weeks later. MSCs appear to activate an anti-apoptosis signaling system at the infarct border zone which effectively protects ischemia-threatened cell types from apoptosis.

Beneficial Remodeling of the Extracellular Matrix

Stem cell transplantation alters the extracellular matrix, resulting in more favorable post-infarct remodeling, strengthening of the infarct scar, and prevention of deterioration in organ function. MSCs appear to achieve this improved function by increasing acutely the cellularity and decreasing production of extracellular matrix proteins such as collagen type I, collagen type III, and TIMP-1 which result in positive remodeling and function.

Activation of Neighboring Resident Stem Cells

Finally, exogenous stem cell transplantation may activate neighboring resident tissue stem cells. Recent work demonstrates the existence of endogenous, stem cell-like populations in adult hearts, liver, brain, and kidney. These resident stem cells may possess growth factor receptors that can be activated to induce their migration and proliferation and promote both the restoration of dead tissue and the improved function in damaged

tissue. Mesenchymal stem cells have also released HGF and IGF-1 in response to injury which when transplanted into ischemic myocardial tissue may activate subsequently the resident cardiac stem cells. (22)

To sum up, although the definitive mechanisms for protection via stem cells remains unclear, stem cells mediate enhanced angiogenesis, suppression of inflammation, and improved function via paracrine actions on injured cells, neighboring resident stem cells, the extracellular matrix, and the infarct zone. Improved understanding of these paracrine mechanisms may allow earlier and more effective clinical therapies

Remyelination

Remyelination involves reinvesting demyelinated axons with new myelin sheaths. Previous attempts aimed at regenerating myelin-forming cells have been successful but limited by the multifocal nature of the lesions and the inability to produce large numbers of myelin- producing cells in culture. Stem cell-based therapy can overcome these limitations to some extent and may prove useful in the future treatment of demyelinating diseases.

Contrary to the general expectations that stem cells would primarily contribute to formation of tissue cells for repair, other mechanisms such as paracrine effects and remyelinations appear to be important ways via which stem cells seem to exert their effect. More Basic research to understand these mechanisms is underway throughout the world.

SURGICAL ASPECTS OF STEM CELLS THERAPY

The stem cell therapy process using autologous bone marrow derived stem cells consists broadly of 3 stages. (1) Procurement of the stem cells from the bone marrow via a bone marrow aspiration in the operating theatre (2) separation, harvesting, enriching &/or expansion and differentiation in the laboratory and finally (3) transplantation or delivery of the cells to the desired location. The laboratory aspects have already been dealt with in the previous chapter therefore in this chapter the procurement and transplantation aspects will be discussed.

Procurement of Stem cells - Bone marrow aspiration

The choice of site may be dependent on various factors such as age, weight, marrow distribution, physical status of the patient, physicians experience etc. However the most common site is the pelvis. The aspiration is easily done from either of the iliac crests (posterior or anterior). The posterior superior iliac spine is easily accessible and identifiable, however to access this, the patient has to be turned in the lateral or prone position which can be troublesome and cumbersome. The anterior superior iliac spine can be accessed with the patient lying comfortably in the supine position. In obese patient, the landmarks may be obliterated due to fat distribution. Sampling is not normally discordant between the anterior or posterior iliac spines.

The site of the aspiration is palpated. For the posterior superior iliac spine, in thin individuals, it is usually palpated as the bony prominence superior and three finger breadth laterals to the intergluteal cleft. The anterior superior iliac spine can be palpated as an anterior prominence on the iliac crest. The overlying skin is prepared in a manner similar to preparation of any site for surgery. The area is anaesthetized by intradermally administering a local anesthetic such as lignocaine using a 25G or 26G needle. A 1 cm area is anesthetized.

A standard bone marrow aspiration needle is inserted through the skin till the bone is felt. Before using the needle it is flushed with heparin. Some surgeons make a small incision with a surgical blade and expose the bone before putting in the needle, however in our experience this is rarely required. The needle which is firmly fixed to the obturator is firmly inserted inside, clockwise and anticlockwise, in a screwing motion with exertion of downward pressure, until the periosteum is reached. With similar motion, the needle is inserted till it penetrates the cortex. At this point initially a sudden giving way of the resistance is felt as the needle enters the soft trabecular bone and then the needle feels firmly fixed in the bone. The angle of insertion of the needle is important as it has to be in alignment with the curve of the bone. If this is not done properly the needle will make a through and through penetration across both the cortical surfaces with the tip now being outside the marrow. A study of the anatomy of the pelvis with a model and personal experience over time make this a very simple procedure.

The stylet is now removed and a 10 ml or 20 ml syringe, with some heparin in it, is attached and

the aspiration is done. A total of 100-120 ml is aspirated in adults and 80-100 ml in children. This is collected in heparinized tubes which need to be appropriately labeled. The bone marrow collected is transported to the laboratory in a special transporter under sterile conditions.(23)

Transplantation of Stem Cells

The other surgical aspect in the process of stem cell therapy is the delivery of the cells which may either be done systemically (through intravenous or intraarterial routes) or locally (intrathecal or direct implantation into the spinal cord or brain). Different centers are following different routes to transplant the cells and as of now there are no comparative studies that could tell us which is the preferred method. However keeping in mind the existence of the blood brain barrier, local delivery would seem to be a more logical option.

Intrathecal delivery

The patient is positioned in the lateral decubitus position, in the curled up "foetal ball" position. Occasionally, the patient is made to sit, leaning over a table- top. Both these maneuvers help open up the spinous processes. The back is painted and draped and local anaesthetic is injected into the L4-5 or L3-4 space. An 18G Touhy needle is inserted into the sub-arachnoid space. After ascertaining free flow of CSF, an epidural catheter is inserted into the space, far enough to keep 8-10 of the catheter in the space. The stem cells are then injected slowly through the catheter, keeping a close watch on the hemodynamics of the patient. The cells are flushed in with CSF. The catheter is removed and a benzoin seal followed by a tight compressive dressing is given. This procedure is usually done under local anesthesia. General anesthesia is given to children.

A spinal needle instead of a catheter is preferred in patients with cardiac problems, where excessive intravenous infusion is to be avoided, in patients on anti-coagulant or anti-platelet drugs so as to avoid bleeding into the sub-arachnoid space, in case where the spine is scoliotic which happens often in patients with muscular dystrophy and in some previously operated cases of lumbar spine surgery.

Sometimes in patients with severe spinal deformities such as scoliosis it is very difficult to get the needle intrathecally and at times assistance has to be taken of the C arm to exactly locate the point and direction of needle placement.

Callera et al (2007) demonstrated for the first time that autologous bone marrow CD 34+ cells labelled with magnetic nanoparticles delivered into the spinal cord via lumbar puncture (LP) technique migrates into the injured site in patients with spinal cord injury. They conducted the trial on 16 patients with chronic SCI. 10 of them were injected intrathecally with labelled autologous CD 34+ cells and the others received an injection containing magnetic beads without stem cells. Magnetic resonance images were obtained before and 20 and 35 days after the transplantation. Magnetically labelled CD 34+ cells were visible at the lesion site as hypointense signals in five patients, which were not visible in the control group.(24)

Intraspinal transplantation

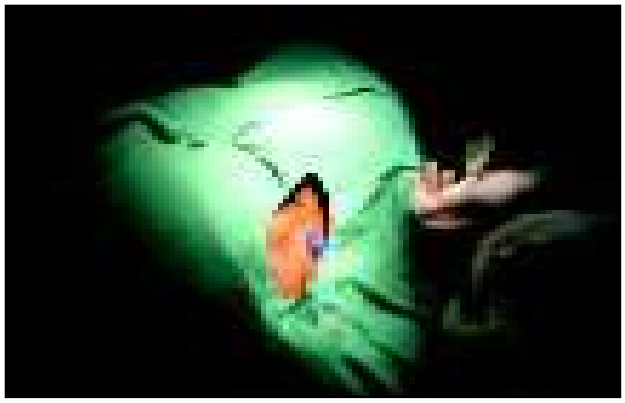
Direct implantation into the spinal cord may be done in one of many ways:-

a) Through a complete laminectomy from one level above to one level below the injury site so that there is sufficient access to the transplantation site. The dura is incised, sparing the arachnoid, which is subsequently opened separately with microscissors. The dorsal surface of the contusion site is located under high-power microscopic magnification. After exposure of sufficient surface in the contusion site, 300µL aliquots of cell paste (total volume, 1.8 mL) are injected into six separate points surrounding the margin of the contusion site. To avoid direct cord injury, 2 × 108 cells are delivered at a rate of 30 µL / min, using a 27-gauge needle attached to a 1-mL syringe. The depth of the injection site is 5 mm from the dorsal surface. To prevent cell leakage through the injection track, the injection needle is left in position for 5 min after completing the injection, after which the dura and arachnoid are closed. The muscle and skin are closed in layers. (25)

b) Though a minilaminectomy and exposure of the spinal cord. The dura is opened and a 27 gauge scalp vein is used by cutting one of the wings. The other wing is held by a hemostat and inserted at a 45 degree angle into the dorsal root entry zone. It is inserted 3mm deep into the spinal cord. Two injections are made on either side above the injury site and two injections are made below the injury site. In China, surgeons are injecting 35 µL of stem cells. In his planned trials, Wise Young is

Bone marrow aspiration

Bone marrow samples



Lumbar Puncture

Intrathecal injection of stem cells

Intramuscular injection of bone marrow derived stem cells

intending to inject an escalating dose of 4 µL, 8 µL and 16 µL.

c) In their ongoing trials, Geron and Neuralstem are using stereotactic systems specifically designed for intraspinal injections. They have the advantage of precision as well as being less invasive. Geron is using a stereotactic frame with a straight needle and injecting 25 µL.

Intra-arterial injection

Following revascularization surgery such as Carotid endartrectomy or Superficial Temporal artery to Middle Cerebral artery bypass, stem cells could be injected directly intra-arterially immediately after the completion of the revascularization procedure. The advantage of this approach is that the stem cells would go directly to the ischemic brain and also that since the artery is already exposed no separate procedure needs to be done for the stem cell injection. The other method of direct intra-arterial injection would be via the endovascular interventional route. This is done by making a puncture in the femoral artery and negotiating a catheter to the arteries supplying the brain. The advantage of this is that it is a relatively non invasive procedure and the limitations of intravenous injection are avoided.

Stereotactic implantation into the brain

Cell transplantation for neurological conditions started with stereotactic implantation of fetal cells for Parkinson's disease. (26) However, after a randomized trial done by Freed et al showed that the clinical outcomes were not significantly different from non transplanted patients this has now been given up. (27) There are many stereo tactic systems available all over the world however the two most popular ones are the Leksell Stereotactic system and the CRW Stereotactic system. The Leksell system involves fixing the frame on the patients head and then getting a MRI done with the frame on. The area where the tissue is to be transplanted is identified on the MRI scan and then using the MRI software the X, Y and Z coordinates are obtained. The patient is now shifted to the operating room where a small burr hole is drilled into the skull and then through this the cells to be transplanted and inserted at the desired location using the X, Y and Z coordinates. The entire procedure is done under local anesthesia.

Intramuscular injection

In certain disorders, especially Muscular dystrophy, cells are also transplanted into the muscle. The points at which these have to be injected are termed as the "motor points. At these motor points, the area is cleaned with povidone iodine. The cells diluted in CSF are injected with the 26G needle going into the muscle at an angle (approx. 45 degrees).The piston / plunger of the syringe is slightly withdrawn to verify the needle is not inside a blood vessel. The cells are then injected, the needle removed and the site immediately sealed with a benzoin seal.

Stem cell transplantation, in its various forms, has been practically attempted for various degenerative disorders, including diabetes, cardiac disorders and neurodegenerative disorders. World wide reports reveal the use of bone marrow derived mononuclear cells and mesenchymal cells, umbilical cord blood stem cells, mesangioblasts, myoblasts, neural stem cells for various incurable and intractable neurological disorders.

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Ch.17 Role of Rehabilitation Therapist in Stem Cell Therapy

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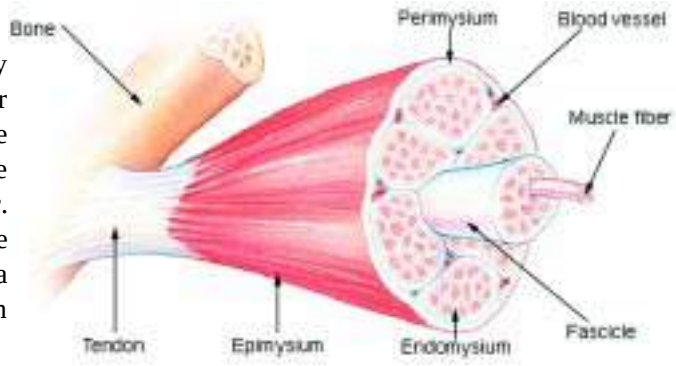
STRUCTURE OF A MUSCLE

In human body, muscles are composed of many muscle fibres, which are separated from each other by connective tissues called endomysium and are arranged in bundles called fascicules, where individual fibres are arranged parallel to each other. Each fasciculus has an outer connective tissue membrane called perimysium and muscle as a whole consists of all these fascicules together with outer layer called epimysium.



Types of Muscles in human body

There are three major types of muscles skeletal, smooth and cardiac. The characteristics of each type are summarized below.

Type of Muscles in Human Body	Characteristics	Location in the human body
Skeletal Muscle	striped, striated, somatic, or voluntary muscles, most abundant	attached to skeleton
Smooth Muscles	plain, unstriped, non-striated, visceral, or involuntary muscles	often encircle or surround the viscera
Cardiac Muscle	intermediate in structure, being striated and at the same time involuntary.	form myocardium of the heart
Myoepithelial Cells	Function: assist in expulsion of secretion from the acini.	present at the bases of secretary acini of sweat gland

Out of the four kinds of muscles, the skeletal muscles are most abundant in the body and have three major types.

TYPES OF SKELETAL MUSCLE FIBERS

The human body has three major types of skeletal muscle fibers: fast fibers, slow fibers, and intermediate fibers.

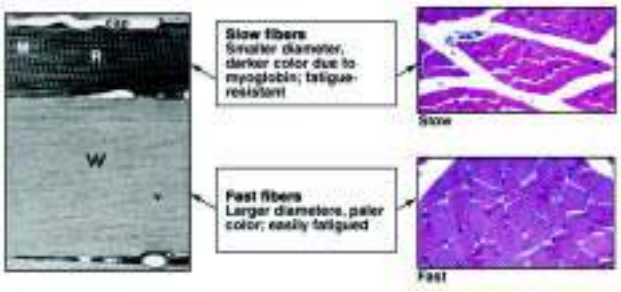
i. Fast Fibers:

Most of the skeletal muscle fibers in the body are called fast fibers, because they can contract in 0.01 sec or less after stimulation. Fast fibers are large in diameter; they contain densely packed mofibrils, large glycogen reserves, and relatively few mitochondria. The tension produced by a muscle fiber is directly proportional to the number of sarcomeres, so muscles dominated by fast fibers produce powerful contractions. However, fast fibers fatigue rapidly because their contractions use ATP in massive amounts, so prolonged activity is supported primarily by anaerobic metabolism. Several other names are used to

refer to these muscle fibers, including white muscle fibers, fast-twitch glycolytic fibers, and Type II-A fibers.

ii. Slow Fibers:

Slow Fibres are only about half the diameter of fast fibers and take three times as long to contract after stimulation. Slow fibers are specialized to enable them to continue contracting for extended periods, long after a fast muscle would have become fatigued. The most important specializations in them is improved mitochondrial performance. Slow muscle tissue contains more extensive network of capillaries than in a typical fast muscle tissue and so has a dramatically higher oxygen supply. In addition, slow fibers contain the red pigment myoglobin . This globular protein is structurally related to hemoglobin, the oxygen-carrying pigment in blood. Both myoglobin and hemoglobin are red pigments that reversibly bind oxygen molecules. Although other muscle fiber types contain small amounts of myoglobin, it is most abundant in slow fibers. As a result, resting slow fibers contain substantial oxygen reserves that can be mobilized during a contraction. Because slow fibers have both an extensive capillary supply and a high concentration of myoglobin, skeletal muscles dominated by slow fibers are dark red. They are also known as red muscle fibers, slow-twitch oxidative fibers, and Type I fibers.



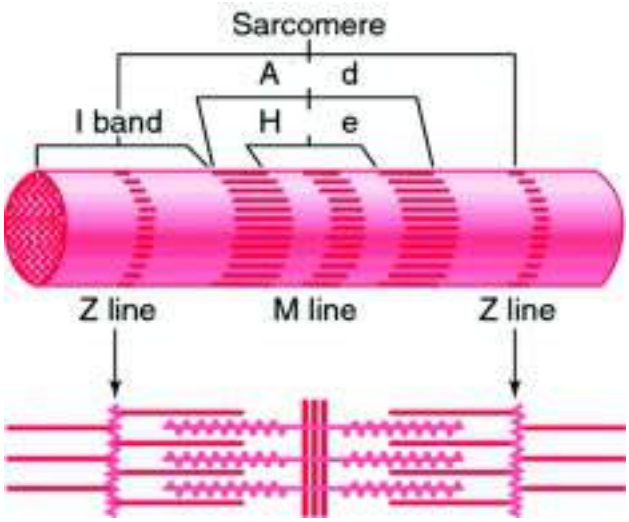
iii. Intermediate Fibers:

The properties of intermediate fibers are intermediate between those of fast fibers and slow fibers. In appearance, intermediate fibers most closely resemble fast fibers, for they contain little myoglobin and are relatively pale. They have a more extensive capillary network around them, however, and are more resistant to fatigue than are fast fibers. Intermediate fibers are also known as fast-twitch oxidative fibers and Type II-B fibers.

MUSCLE PHYSIOLOGY

Sarcomere is the contractile unit of a myofibril, which are repeating units and delimited by the Z bands along the length of the myofibril.

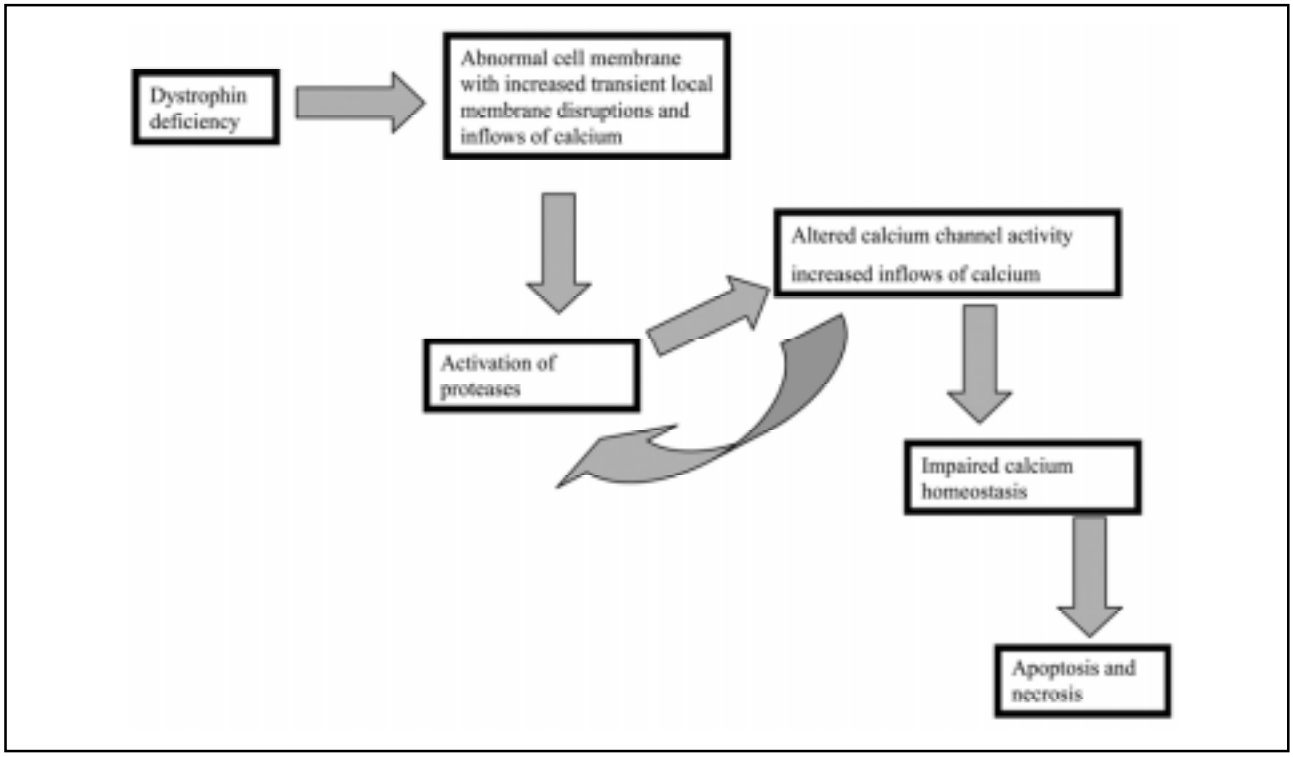
Muscle units are separated from other muscle groups by plasma membranes called the sarcolemma and the cytoplasm within is called the sarcoplasm. Within the sarcoplasm are multiple long protein bundles called myofibrils, and many ATP producing mitochondria, as well as glycogen (a form of stored glucose for energy) and myoglobin (oxygen stored in blood for the breakdown of glycogen). Bundles of parallel myofilaments make up the myofibrils which is where most of the action takes place. In the myofilaments are contractile proteins called myosin (thick filaments), and actin (thin filaments).When signaled, the actin and myosin interlock and slide over each other to stretch or slide into one another to contraction. They are signaled from the nervous system followed by a series of chemical reactions involving ATP, calcium, sodium and potassium ions.



There are many other proteins involved in the process. Aside from the contractile proteins, there are regulatory proteins called tropomyosin and troponin which act like a switch to determine when to contract and when to relax. On the muscle fiber the 'I band' is the space between the myosin (thick) filaments, where lies only the thin filaments. In the middle of each 'I band' is a dark disc called the 'Z disc' made of titan, (elastic filament), which is connected to the sarcolemma by the cytoskeleton. The space between each Z disc, where these filaments interact, is called the sarcomere. As the muscle contracts the 'I band" shrinks and the sarcomere shortens and as the Z disc's come closer

together pulling on the sarcolemma shortening the cell. This is how the muscle contracts. One of the most clinically important accessory proteins here is dystrophin which is located just under the sarcolemma in the cytoplasm in the area of the 'I band'. It is produced by specific genes and links the actin filaments to the protein extracellular matrix in the membrane known as the dystrophin-associated protein complex. Elements of the dystrophin gene and the protein structure have been identified, yet the exact functional role is still a bit unclear. However, as research continues it is thought that its primary function is to provide mechanical reinforcement to the structure of the sarcolemma and thereby protecting the membrane from the stress or tearing during contraction.

In Muscular Dystrophy patients, as dystrophin is defective or absent, the membrane breaks down and molecules like proteins and enzymes leak out of the fiber into circulation. These enzymes and chemicals that leak out are responsible for certain chemical reactions and disruption of the process of muscle contraction which thus causes irreparable damage.



Pathophysiology of Protein deficient Muscle:

To summarize, important abnormalities of dystrophin-deficient muscle cells have been demonstrated in three areas:

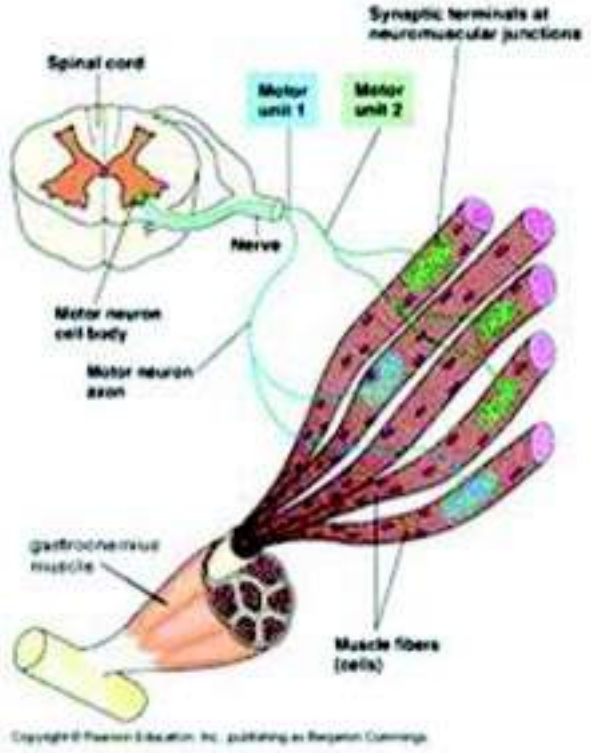
1. Calcium homeostasis,
2. An increased susceptibility to oxidative toxins, and
3. Increased (and stress enhanced) membrane permeability.

MOTOR POINT

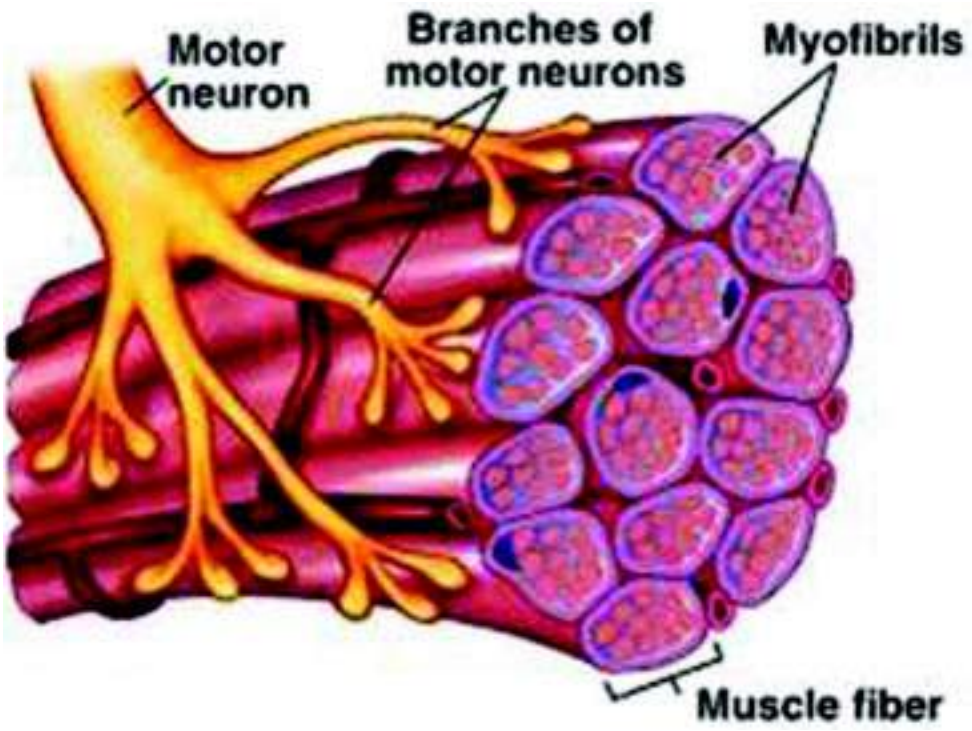
Motor point is the point at which the main nerve enters the muscle or, in case of deeply placed muscle, the point where the muscle emerges from under covers of the more superficial ones.

Facts about Motor points:

Motor points are frequently at the junction of the upper & middle one thirds of the fleshy belly of the muscles, although there are exceptions e.g.: the motor point of vastus medialis, whose nerve enters the lower part of the muscle, is situated a short distance above the knee joint. Deeply placed muscles may be stimulated most satisfactorily where they emerge from beneath the more superficial ones, e.g.: extensor hallucis longus in the lower one third of the lower leg. This is the point on the skin region where an innervated muscle is most accessible to percutaneous electrical excitation at the lowest intensity. This point on the skin generally lies over the neuro vascular hilus of the muscle & the muscles band or zone of innervations. Muscle fibres do not always extend the whole length of a muscle & myoneural junctions are not uniformly spread out all over the muscle but are concentrated in a confined area-the zone or band of innervations where the greatest concentration of



A Neuromuscular Junction



The Motor Unit

motor endplates & the other large diameter nerve fibres may be reached with less concurrent painful stimulation of the smaller diameter cutaneous fibres.

The exact location of motor point varies slightly from patient to patient but the relative position follows a fairly fixed pattern. Some motor points are superficial & are easily found, while others belonging to deep muscles are more difficult to locate.

CONCEPT OF MOTOR POINT STIMULATION

When a nerve is stimulated at a nerve cell or an end organ, there is only one direction in which it can travel along the axon, but if it is initiated at some point on the nerve fibre it is transmitted simultaneously in both directions from the point of stimulation.

When a sensory nerve is stimulated the downward travelling impulse has no effect, but the upward travelling impulse is appreciated when it reaches conscious levels of the brain. If impulses of different durations are applied, using the same current for each, it is found that the sensory stimulation experienced varies with the duration of the impulse. Impulses of long duration produce an uncomfortable stabbing sensation, but this becomes less as the duration of the impulse is reduced until with impulses of 1 ms & less only a mild prickling sensation is experienced.

When a motor nerve is stimulated, the upward - travelling impulse is unable to pass the first synapse, as it is travelling in the wrong direction, but the downward travelling impulse passes to the muscles supplied by the nerve, causing them to contract.

When a stimulus is applied to a motor nerve trunk, impulses pass to all the muscles that the nerve supplies below the point at which it is stimulated, causing them to contract.

When a current is applied directly over an innervated muscle, the nerve fibres in the muscle are stimulated in the same way. The maximum response is thus obtained from stimulation at the motor point.

Preparation of the patient

Clothing is removed from the area to be plotted & the patient is supported comfortably in good light. The skin has high electrical resistance as the superficial layers being dry, contain few ions. The resistance is reduced by washing with soap & water to remove the natural oils & moistening with saline immediately before the electrodes are applied. Breaks in the skin cause a marked reduction in resistance which naturally results in concentration of the current & consequent discomfort to the patient. To avoid this broken skin is protected by a petroleum jelly covered with a small piece of non absorbent cotton wool to protect the pad. The indifferent electrode should be large to reduce the current density under it to a minimum. This prevents excessive skin stimulation & also reduces the likelihood of unwanted muscle contractions, as it may not be possible to avoid covering the motor points of some muscles.

Preparation of apparatus

Faradic type of current

A low frequency electronic stimulator with automatic surger is commonly used. A faradic current is a short -duration interrupted direct current with a pulse duration of 0.1 - 1 ms & a frequency of 50 - 100 Hz. Strength of contraction depends on the number of motor units activated which in turn depends on the intensity of the current applied & the rate of change of current. To delay fatigue of muscle due to repeated contractions, current is commonly surged to allow for muscle relaxation.

Stimulation of Motor points

This method has the advantage that each muscle performs its own individual action & that the optimum contraction of each can be obtained, by

Electrical stimulator used for stimulation and plotting of motor points.

stimulating the motor point. The indifferent electrode is applied & secured in a suitable area. The indifferent electrode is placed over the motor point of the muscle to be stimulated. Firm contact ensures a minimum of discomfort & where possible the whole of operators hand should be in contact with the patient's tissues so that she /he can feel the contractions produced.

Selection of the Individual muscles for intramuscular injection of stem cells in Muscular Dystrophy Patients:

Patients with Muscular Dystrophy have primarily weak antigravity muscles like hip knee extensors , back extensors and ankle Dorsiflexors in lower limbs. In upper limb the proximal shoulder girdle muscles like deltoids, biceps triceps and scapular stabilizers like rhomboids and serratus are the most commonly affected. All of these muscles are needed for mobility and activities of daily living , but as they progressively get weaker , patients begin to get dependent for functional activities.

Figure 4 : Preparation of the patient for motor point plotting

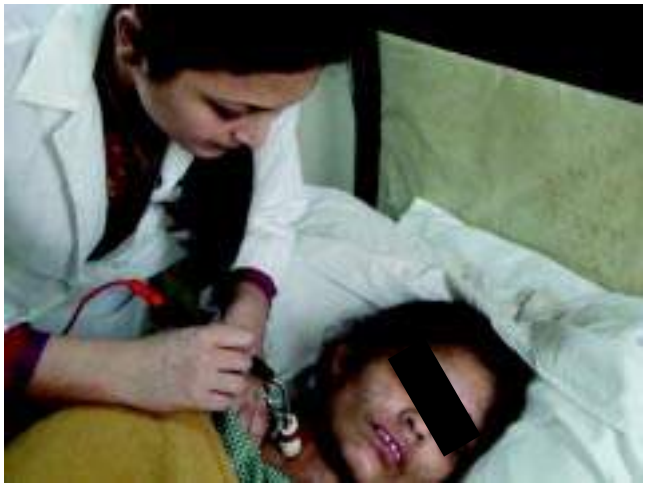


Figure 5 : Plotting of motor point (strenomastoid muscle)

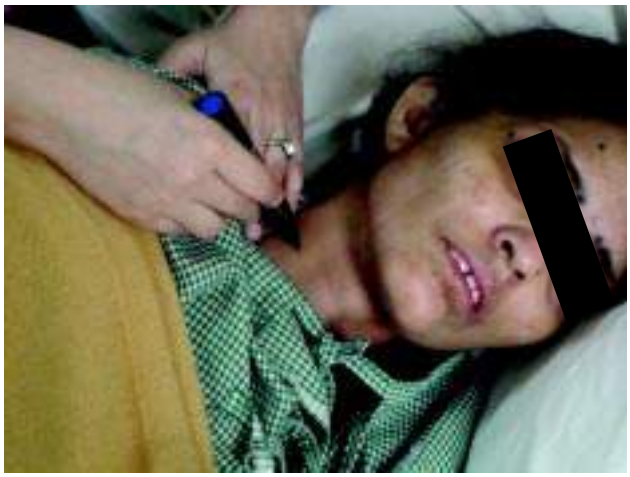
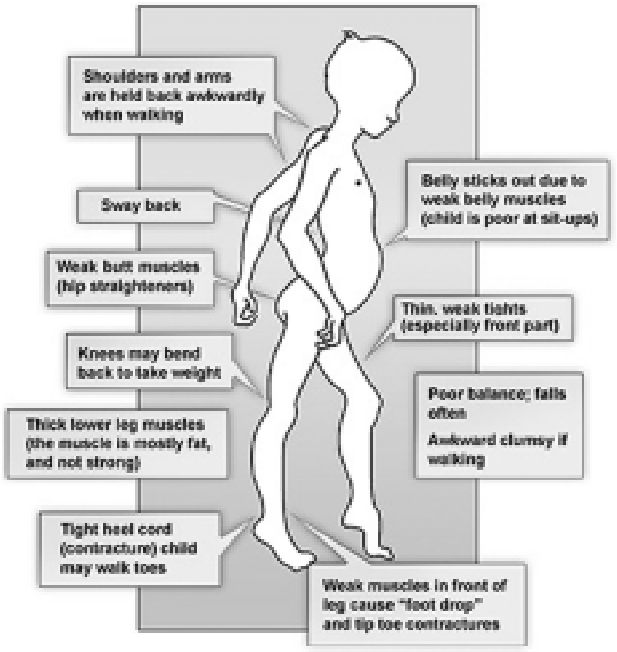


Figure 6 : Marking of sternomastoid muscle motor point.

Although MD can affect several body tissues and organs, it most prominently affects the integrity of muscle fibers.It causes muscle degeneration, progressive weakness, fiber death, fiber branching and splitting, phagocytosis (in which muscle fiber material is broken down and destroyed by scavenger cells), and in some cases, chronic or permanent shortening of tendons and muscles. Also, overall muscle strength and tendon reflexes are usually lessened or lost due to replacement of muscle by connective tissue and fat.

So selection of muscles (motor points) for intramuscular injection depends on manual muscle testing & patient's complain of weakness & difficulty in activities of daily living. So rehabilitation team (Physiotherapists and Occupational therapists) decides motor points of



which muscles need to be injected with stem cells. Also the Electromyography and Musculoskeletal MRI , aid in locating muscles with severe affection in the form of fatty infiltration or reduced interference pattern on voluntary contraction.

In few selective types of Muscular Dystrophies like Oculopharyngeal MD , facial muscles are weak and are therefore considered for intramuscular injection.

Commonly considered muscles for injection are as follows

A) Major muscles of upper limbs that are generally considered:

a) Deltoid: Anterior, middle & posterior fibres.

b) Biceps brachialis.

c) Triceps: long, lateral & medial heads.

d) Thenar muscles: Opponens pollicis & abductor pollicis brevis & flexor pollicis brevis.

e) Hypothenar muscles: abductor, flexor & opponens digiti minimi.

B) Major muscles of lower limbs that are generally considered:

a) Quadriceps: vastus medialis, vastus lateralis, rectus femoris.

b) Hamstrings: Biceps femoris, Semimembranosus & semitendinosus.

c) Glutei.

d) Dorsilflexors: Tibialis anterior, Peronei longus & brevis, EHL.

C) In trunk:

Abdomen & back extensors are considered, & in neck muscles sternocleidomastoid.

D) Facial Muscles:

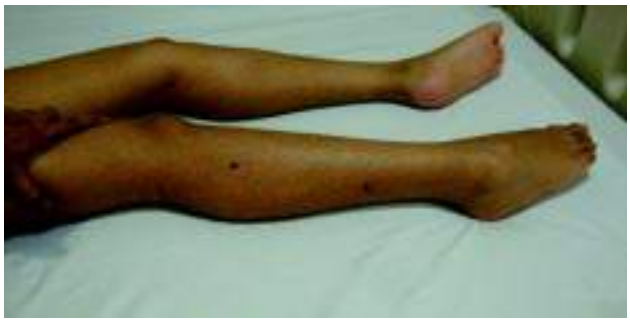
In case of facial muscle weakness : orbicularis oris, orbicularis oculi, Buccinator, rhizorius, frontalis, mentalis, etc.

Intramuscular stem cells injection in motor points within the muscle, ie the area with high concentration of motor end plates is very specific transplantation. Also multiple motor points in choosen muscle group allows for a graded response, thus allowing increment in muscle strength clinically depending on, further specific training & strengthening of individual injected muscles. An injection of stem cell in the motor end plate potential, can be identified in the neuromuscular system within few hours, although the onset of clinical effects is noticed as early as 72 hours post transplant, which varies from patient to patient.

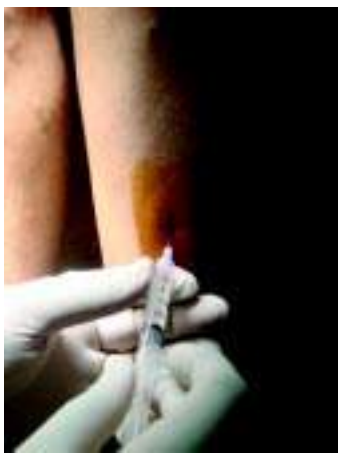
MECHANISM OF ACTION OF INTRAMUSCULAR STEM CELL INJECTION AT MOTOR POINTS

As motor point is the point at which the main nerve enters the muscle. Delivery of stem cells at this point facilitates further specific implantation of the stem cells in isolated individual muscles and aids in enhancing the healing of the degenerated muscle. Also the stem cells promote regeneration by enhancing angiogenesis, suppression of inflammation and improved function via paracrine actions on injured cells ,neighboring resident stem cells , extracellular matrix , and the infarcted zone. (*Refer chapter 12)*

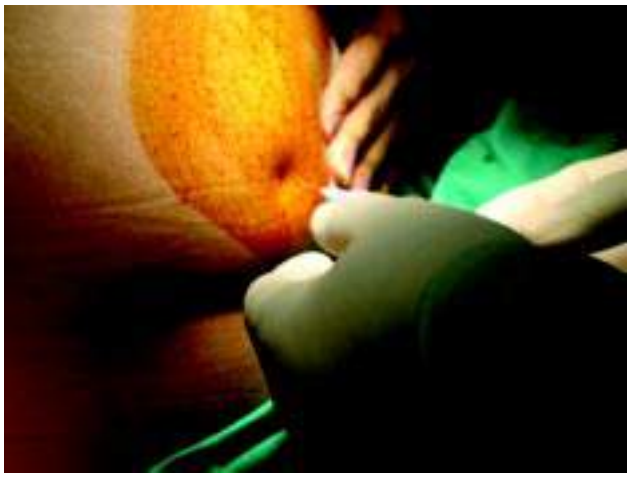
Post stem cell injection these muscles need specific training & individual muscle strengthening program so that results are seen by following mechanisms:



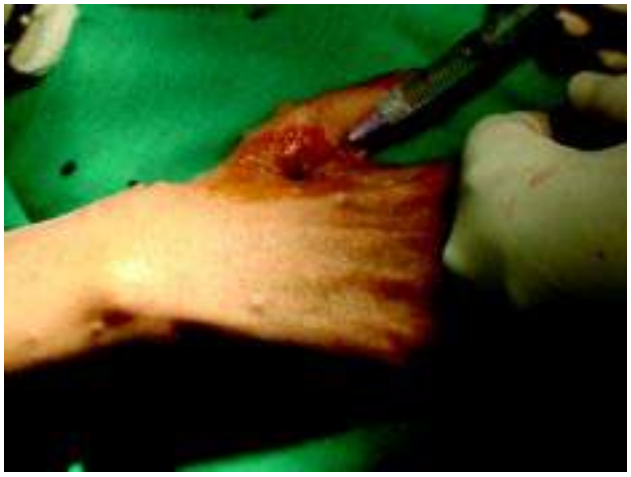
Plotted motor points of tibialis anterior and peronei muscle



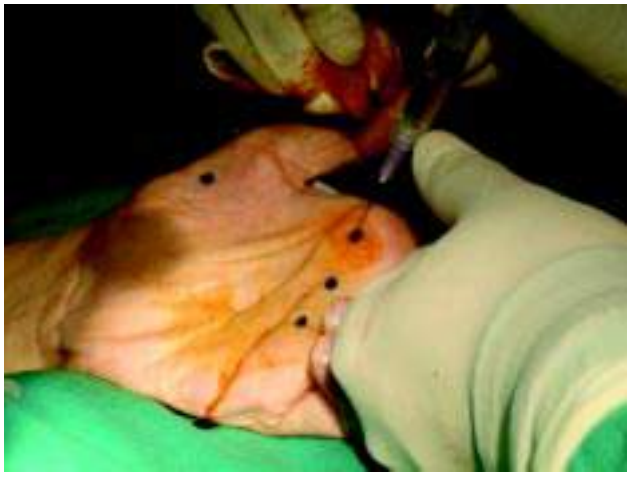
Injection of stem cells in tibialis anterior muscle motor point.



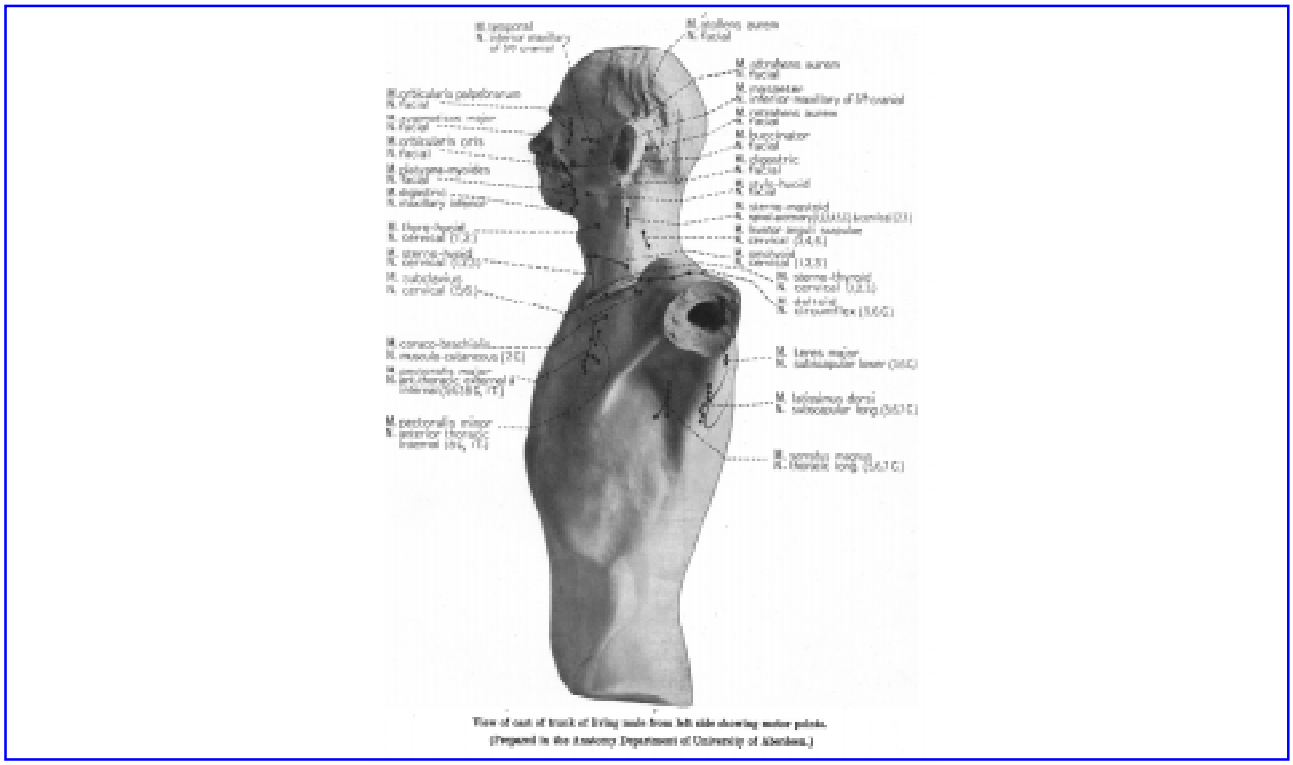
Injection of stem cells in the glutei muscle motor point.

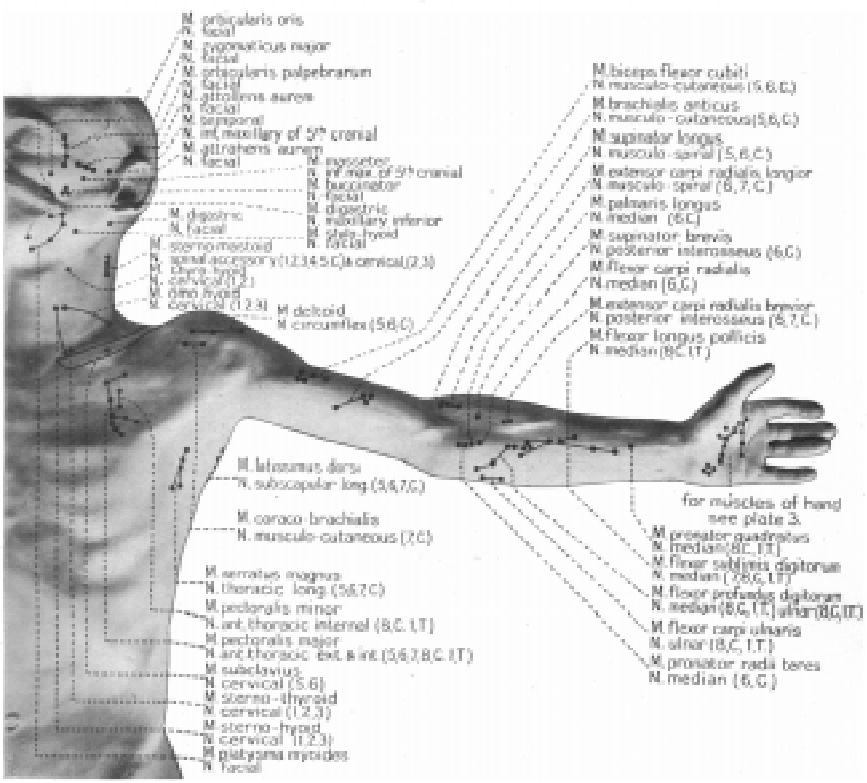


Injection of stem cell injection in the adductor pollicis muscle motor point.

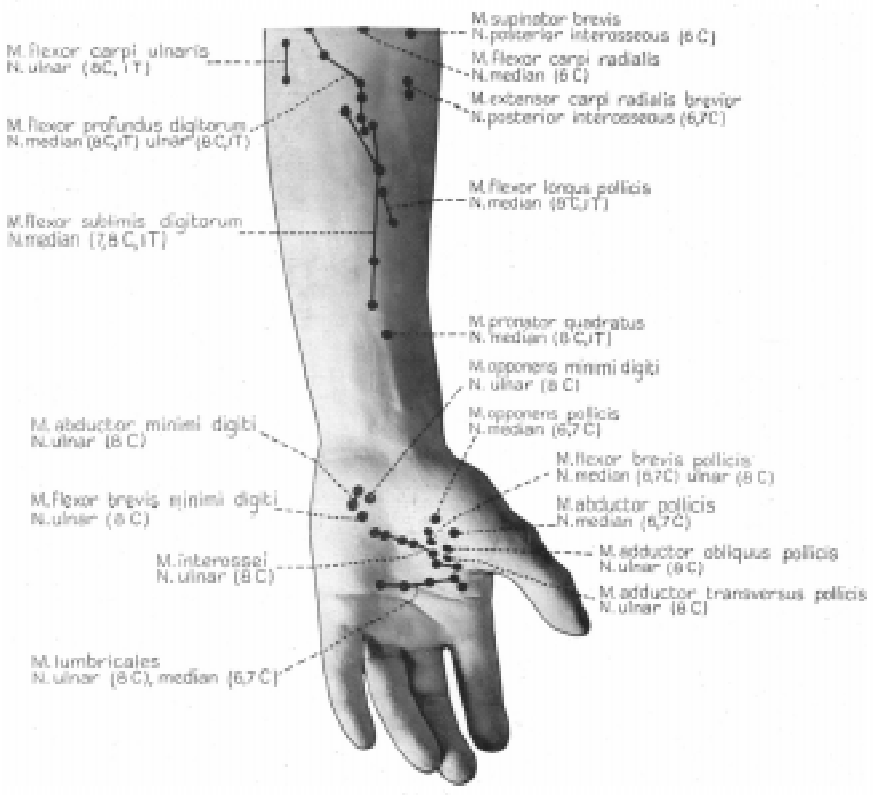


Injection of stem cells in the lumbrical muscle motor points

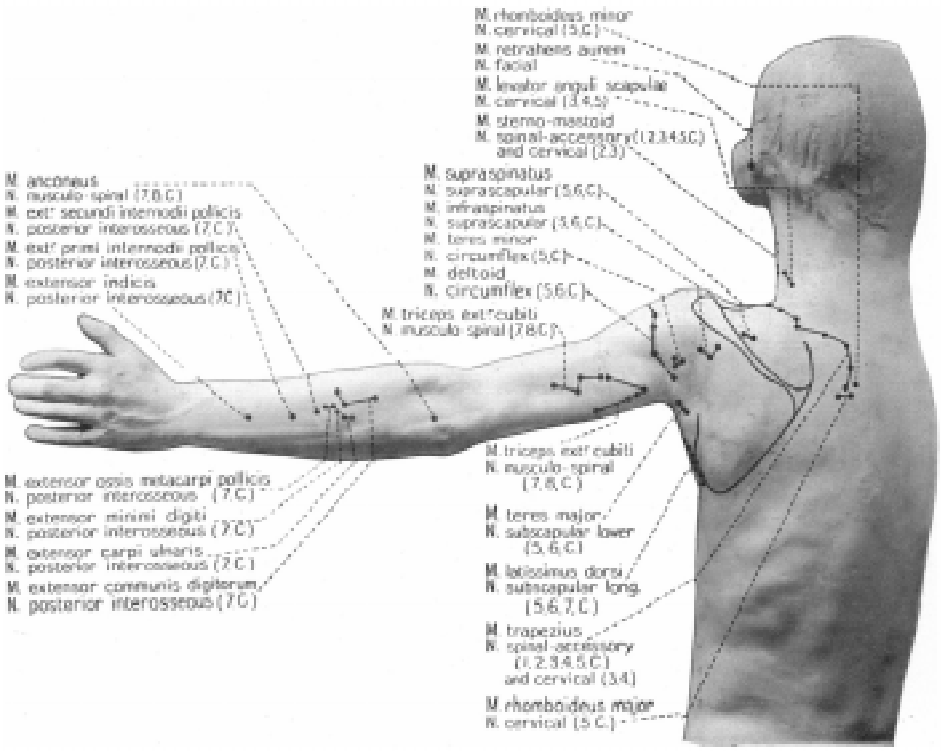




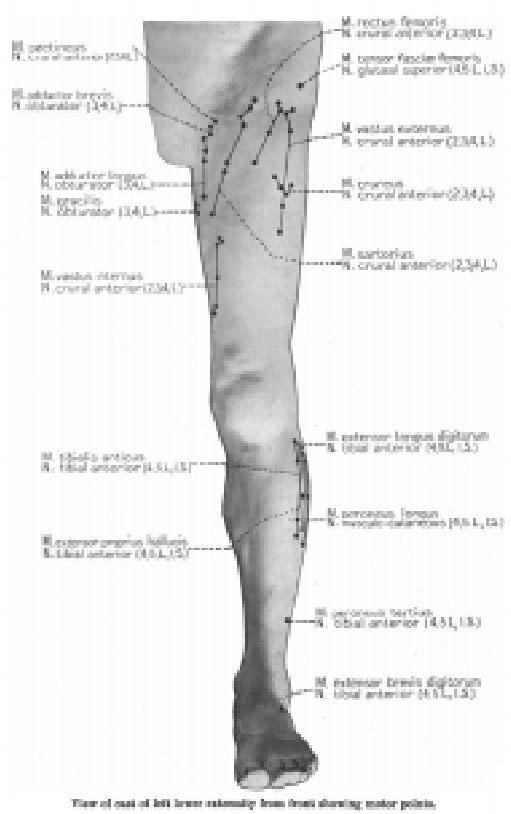
View of cast of left side of trunk and upper extremity from front showing motor points.
(Prepared in the Anatomy Department of University of Aberdeen.)



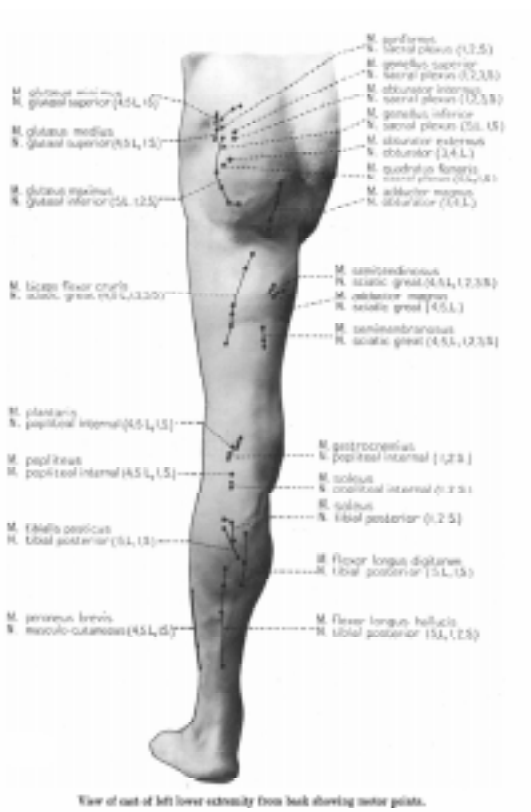
View of cast of left fore-arm and hand from front showing motor points.
(Prepared in the Anatomy Department of University of Aberdeen.)



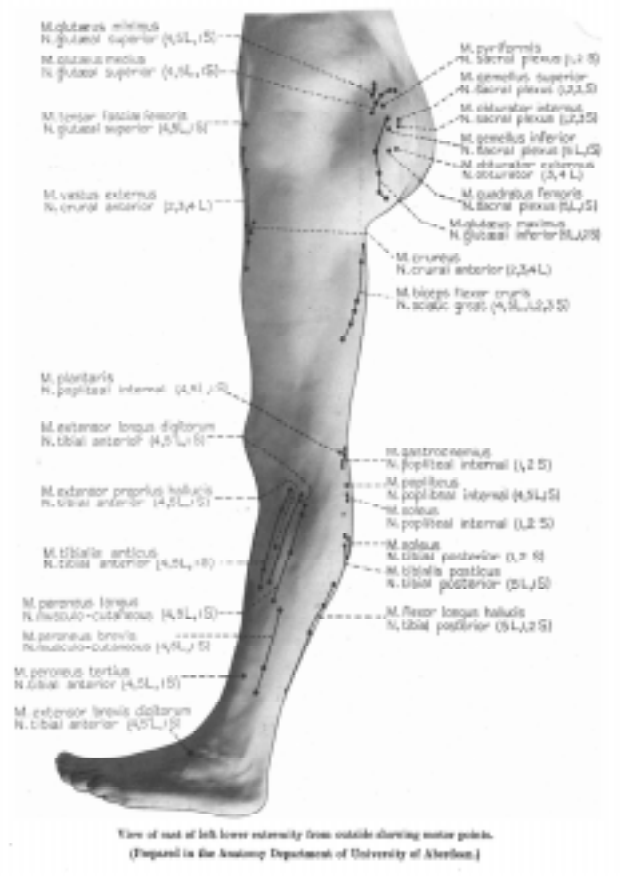
View of cast of left side of trunk and upper extremity from behind showing motor points.
(Prepared in the Anatomy Department of University of Aberdeen.)



View of cast of left lower extremity from front showing motor points.
(Prepared in the Anatomy Department of the University of Aberdeen.)



View of cast of left lower extremity from back showing motor points.
(Prepared in the Anatomy Department of University of Aberdeen.)



1. In muscles that contain a mixture of fast and intermediate fibers, the proportion can change with physical conditioning. For example, if a muscle is used repeatedly for endurance events, some of the fast fibers will develop the appearance and functional capabilities of intermediate fibers. The muscle as a whole will thus become more resistant to fatigue.
2. Exercise leads to stimulation of Satellite cells (special stem cells which lie adjacent to skeletal muscle fibre and play a role in muscle regeneration and repair)
3. As dystrophy patients muscles lack enzyme , which produces nitric oxide , which in turn leads to vasodilatation , in order to stimulate satellite cells .Natural stimulation of satellite cells in them is very slow , thus leading to rapid degeneration and braek down to muscles.But direct stem cell intramuscular transplantation and exercise leads to angiogenesis and vasodilatation , leading to stimulation of satellite cells and thus repair and regeneration of muscles.

Gradually as the muscle strength increases patient gains efficiency & independency in activities of daily living (ADL).

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Ch.18 Clinical Improvements in Neurological disorders after Stem Cell Therapy

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At NeuroGen Brain and Spine Institute, stem cell therapy was carried out on disorders like muscular dystrophy, cerebral palsy, multiple sclerosis, stroke, spinal cord injury, motor neuron disease and other rare incurable neurological , neuromuscular diseases including genetic disorders based on the Helsinki Declaration. (1-8)

Till date over 700 patients of various disorders (as mentioned above) have been administered stem cell therapy. A detailed analysis of their outcome, with an average followup of 18 months for 300 patients has been presented in this chapter.

Out of these 300 patients,73% of the patients were observed to have improvements, ranging from mild to significant category (graph 1).The other 25% were mainly status quo, with no changes, while 2% of the patients deteriorated in their condition , despite the therapy , all were suffering from progressive neurological disorder (which were revealed to be patients of motor neuron disease). (Graph 1)

Neurological Disorder	Number of patients
Muscular Dystrophy	72
Cerebral palsy	16
Spinal Cord Injury	74
Autism	22
Stroke	11
Multiple sclerosis	21
Motor Neuron Disease	47

The majority of this above cohort was spinal cord injury and muscular dystrophy.

Muscular Dystrophy

Seventy two muscular dystrophy patients who underwent intrathecal autologous bone marrow derived mononuclear cell transplantation, could broadly be categorized as Duchene Muscular Dystrophy type,(41) Limb Girdle Muscular Dystrophy (17), Congenital Muscular Dystrophy (11), Becker's Muscular Dystrophy(2) and Fascio Scapulohumeral Dystrophy(1). (Graph 2)

Their mean follow up of 18 months showed that out of 72 patients , 67 (93%) showed positive results while non responders were only 5 (7%).The major area showing improvements were increase in trunk strength (32), lower extremity strength (30) , upper extremity strength(20) and improved gait pattern (11).Many of these patients showed improved muscle strength on manual muscle testing. Muscle tightness was reduced in 40 patients. (Graph 3, 4 , 5)

Not only that, these changes also lead to functional improvements, as reflected by the shift in the Functional Independence Measure scores (FIM scores) in 38 patients. Biochemical response in terms of reduction in serum creatine phosphokinase was seen in 47 patients. Objective evidence of response to treatment was obtained by:

a) increase in amplitudes of compound motor units action potential as well as increased interference pattern while muscle contraction on electromyography (EMG changes seen in 9 patients)

b) Imaging (MRI) of the musculoskeletal system(both upper and lower limbs) showed reduction in fatty infiltration, some regeneration of muscle fibres and molecular changes on MR Spectroscopy of the muscles- indicating molecular flux in the intramyo-cellular and extramyocellualr lipids. (8)

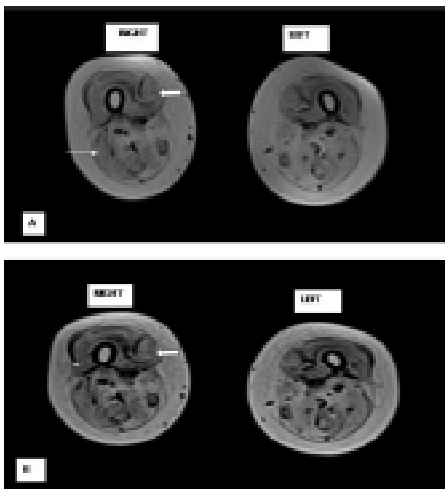

Fig.1: Axial T1W images at the level of upper thigh

(A)Pre-stem cell therapy show marked fatty infiltration of the right vastus medials (thick arrow) and lateralis muscle (thin arrow), seen as high signal intensity. (B) Post-stem cell therapy shows reduced high signal in both the vastus medialis (thick arrow) and lateralis (thin arrow) suggestive of less fatty infiltration and regeneration of muscle fibres

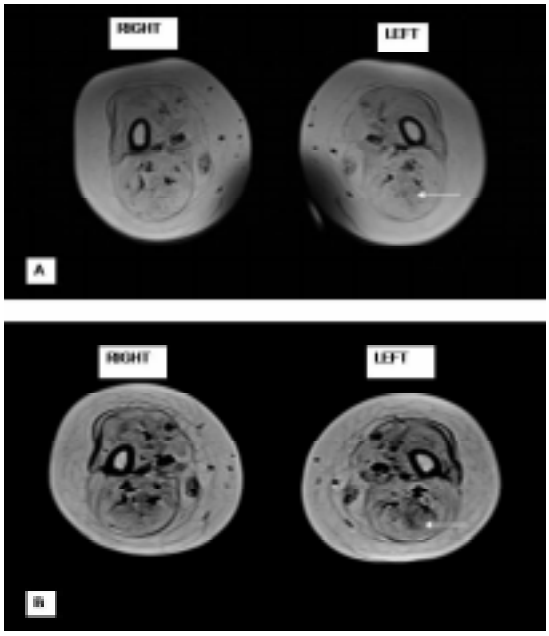

Fig. 2: Axial T1W images at the level of upper thigh

(A)Pre-stem cell therapy show marked fatty infiltration of the left semitendinosus (thin arrow) seen as high signal intensity. (B) Post-stem cell therapy shows reduced high signal in the left semitendinosus (thin arrow) suggestive of less fatty infiltration and regeneration of muscle fibres.

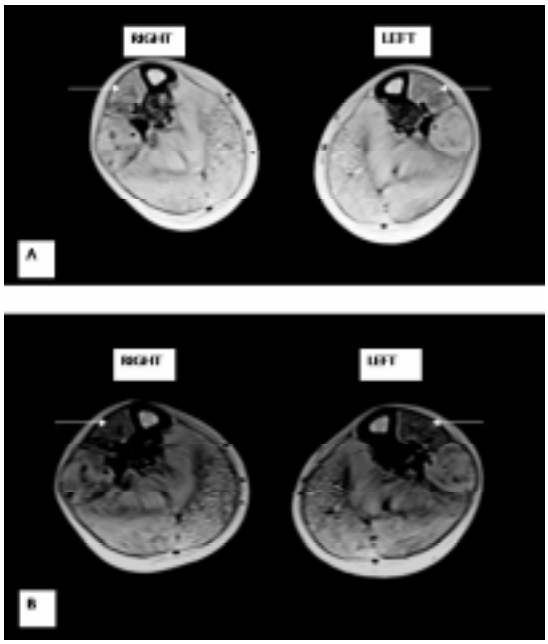

Figure 3: Axial T1W images at the level of the calf

(A) Pre-stem cell therapy show marked fatty infiltration of the bilateral tibialis anterior muscle (thin arrows) seen as high signal intensity. (B) Post-stem cell therapy shows reduced high signal in bilateral tibialis anterior muscle (thin arrows) suggestive of less fatty infiltration and regeneration of muscle fibres.

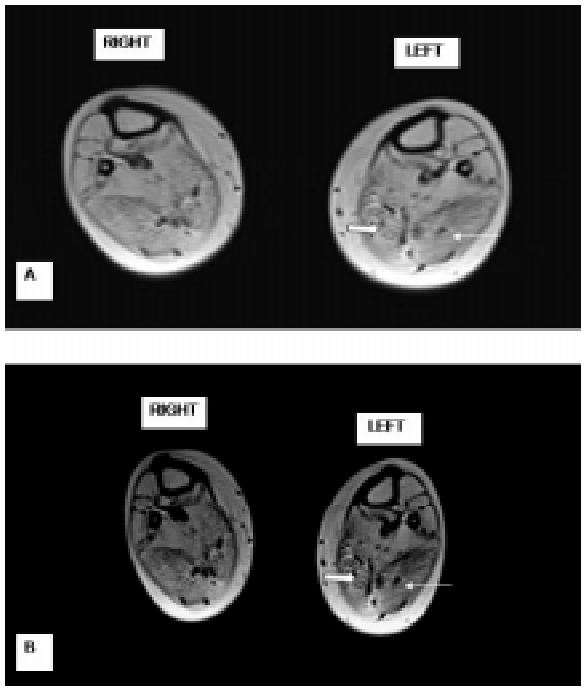

Figure 4: Axial T1W images at the level of the calf

(A)Pre-stem cell therapy show marked fatty infiltration of the left medial (thick arrow) and lateral gastrocnemius muscles (thin arrow)) seen as high signal intensity. (B) Post-stem cell therapy shows reduced high signal in left medial (thick arrow) and lateral gastrocnemius muscles (thin arrow) suggestive of less fatty infiltration and regeneration of muscle fibres.

Cerebral Palsy

Cerebral palsy patients who underwent stem cell therapy (16), showed symptomatic improvement in oromotor functions (37.5%) like speech & swallowing , improved neck holding (37.5%), sitting balance (62.5%),range of limb movements (50%), cognition (50%) and normalization of overall muscle tone (62%).Overall improvement was seen in 87.5% of the treated patients. Apart from functional changes, objectives changes in neuroimaging corroborating with actual improvements is now emerging. This is in the form of improvement in neurometabolism as seen in PET CT Scan of the brain over 6 months. (graph 7-9)

Spinal Cord Injury

Out of the 74 patients treated, 50 were paraplegics, while 24 were quadriplegics, with road traffic accident (83.7%) being the most important cause .Majority of these were males (52), while 22 were females. (Graph 9- 11)

Following stem cell therapy, 81% (60) patients improved, while 14 (19%) patients maintained status quo (neurologically the same). On assessing them symptom-wise 48 patients showed improvement in sitting balance (static and dynamic),13 showed improvement in muscle strength, 4 showed complete recovery in bladder and bowel functions, 9 showed sensory recovery and 8 of them showed complete recovery from postural hypotension. In addition 50% (42) patients had a reduction in spasticity and 15 were able to ambulate with assistance of walker. These changes reflected on a shift on ASIA scale by 2 grades in 12 of these who showed improvements in muscle power and gait with assisted devices. Changes on FIM score were appreciated in 38 out of 74 patients, score shifts ranging from 2 to 45.(graph 12-16)

Autism

Out of 22 cases who underwent intrathecal stem cell therapy,18 (82%) of the children showed improvements to varied extents. These changes seen were mainly in social interaction(72%), increased attention span (54%), improved memory (45%), eye contact(81%) and behavior(77%) which includes reduction in hyperactivity, temper tantrums and emotional lability. A positive shift in these areas has been beneficial in overall management and training of the children, thereby helping towards the goal of integrating them into the mainstream. In addition, PET CT Scan of the brain done before the therapy helps to pinpoint areas of brain which are either hypo or hypermetabolic. A followup analysis over a period of 6-8 months with a repeat PET CT Scan has shown improvement in neurometabolism which corroborates with the clinical findings. (graph 17,18)

Stroke

Out of 11 patients, 55% were suffering from non haemorrhagic while 45% from haemorrhagic stroke, with a female versus male ratio of 7:4. (Graph 19-21)

Reduction in spasticity and improvement in cognition (60%),were noticed, followed by improvement in hand function and gait(47% and 67% respectively), speech improvement in 75% patients while cognition improved in 60% of those affected. (graph 22-24)

Multiple Sclerosis

Twenty two multiple sclerosis patients underwent intrathecal autologous bone marrow derived mononuclear cell transplantation. The mean follow up of 6 months showed that 11 patients shifted on EDSS Scale showing objective Neurological Improvement. (Graph 25, 29 ,30)

On assessing them symptom-wise 17 patients showed reduction in spasticity,9 showed improved upper extremity and trunk coordination,, 6 improved in speech clarity and 8 showed increased in muscle strength.(graph 26 , 27 , 28,31)

Motor Neuron Disorder

Forty seven motor neuron disease patients underwent intrathecal autologous bone marrow derived mononuclear cell transplantation. In contrast to other neurological disorders, results seen in this group of rapidly deteriorating condition are not very encouraging, with either no improvement or deterioration in seen in over 70% of the patients. Patients with bulbar symptoms were seen to have continued progression. 14 showed some minor improvements whereas 33 patients kept on deteriorating with symptoms of early fatigue, weakness, muscle wasting and bulbar symptoms, which progressed with the natural course of the disease. Out of the 14 patients who improved, symptoms which showed results were improved neck holding, speech, swallowing, and reduction in fasciculations and a halt in the progression of muscle weakness. (graph 32-35)

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Index

G

H

I

K

L

M

N

O

P

R

S

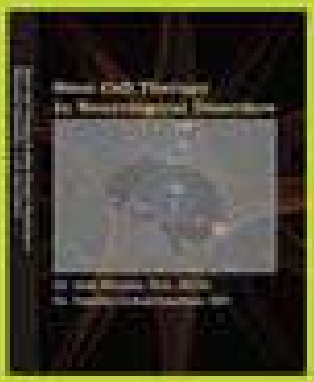
T

U

V

W

Other Publications:



Stem Cell Therapy in Neurological Disorders:
This is the first book published in India on Stem Cell Therapy giving insight into exciting field of neuroregeneration. It is a comprehensive book which covers history of stem cells, the basic concepts of cell repair, mechanism of action of stem cells, procedural details, clinical role in various disorders alongwith case reports and future of this novel invention. It explains in details about the different types of stem cells, the laboratory separation, options for administration and their advantages as well as disadvantages. This book also emphasizes the key aspect of rehabilitation in neuroregeneration. This unique book will be helpful in understanding the fascinating stem cells.

Stem Cell Therapy & Other Recent Advances in Muscular Dystrophy
The book covers three broad sections wherein section one deals with the basic information on muscular dystrophy, its genetic inheritance, treatment options available including gene therapy. The next section introduced the novel concept of Neuro Regenerative Rehabilitation therapy (NRRT) which explains role of autologous (self derived) stem cell therapy along with neurorehabilitation. It also reviews other types of cell transplantations which have been reported by other researchers. The last section details out 120 case reports of muscular dystrophy patients who have undergone autologous bone marrow derived stem cell therapy, highlighting their significant improvements.
This book was written with a purpose of giving a comprehensive overview on Muscular Dystrophy, which incorporates the various aspects of the clinical features, conventional treatment and newer/recent advances .Though majorly it is aimed for the medical fraternity, the book also contains, interesting information for the layman as well as the patients about stem cell therapy and its potential.

NeuroGen Brain and Spine Institute

The Neurogen Brain and Spine Institute has been set up to help patients, with incurable neurological disorders, get relief from their symptoms and physical disabilities using the safest and most effective available treatments and technologies from the feild of Neurosciences and Regenerative medicine in a professional and scientific as well as holistic and caring manner.

We have introduced a novel concept of NeuroRegerative Rehabilitation Therapy (NRRT), wherein, our strategy is to promote the recovery of neural function with a close integration of stem cells and physical, occupational and speech therapies. We recognize that even small functional gains may have significant effect on the quality of life of our patients. In addition to the medical treatment there is a significant emphasis on both clinical as well basic research, so that the best therapeutic strategies can be evolved and practiced at the same time.

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