PAEDIATRIC RESPIRATORY REVIEWS (2007) **8**, 8–16

ELSEVIER

Paediatric Respiratory Reviews

MINI-SYMPOSIUM: AIRWAY CLEARANCE IN CYSTIC FIBROSIS

Chest physical therapy, breathing techniques and exercise in children with CF

Maggie McIlwaine*

Clinical Instructor, School of Rehabilitation Sciences, University of British Columbia and Professional Practice Leader, Department of Physiotherapy, BC Children's Hospital, Vancouver, Canada

KEYWORDS
cystic fibrosis;
chest physiotherapy;
postural drainage and percussion;
active cycle of breathing;
forced expiration;
autogenic drainage;
exercise;
airway clearance techniques

Summary Chest physiotherapy in the form of airway clearance techniques and exercise has played an important role in the treatment of cystic fibrosis. Until the 1990s the primary airway clearance technique used was postural drainage combined with percussion and vibration (PD&P). It was introduced into the treatment of CF with little evidence to support its efficacy and once established, it has been difficult ethically to perform a study comparing PD&P to no treatment. A common question, yet unanswered is when should it be commenced, especially for the newly diagnosed asymptomatic CF patient? Recently, the technique of PD&P has been modified to include only non-dependant head-down positioning due to the detrimental effects of placing a person in a Trendelenburg position. In the 1990s other airway clearance techniques gained popularity, in that they could be performed independently, in a sitting position and avoided many of the detrimental effects of PD&P. These techniques include the Active cycle of breathing technique, formally called the Forced expiration technique and Autogenic drainage. Both these breathing techniques aim at using expiratory airflow to mobilize secretions up the airways and incorporate breathing strategies to assist in the homogeneity of ventilation. Studies suggest that both these techniques are as effective if not more effective than as PD&P and offer many advantages over PD&P. It has been suggested that exercise can be used as an airway clearance technique; however the literature does not support this. Rather, when exercise is used in addition to an airway clearance technique there is enhanced secretion removal and an overall benefit to the patient. Further research needs to be directed at assessing the effects of an airway clearance technique on the individual patient using appropriate outcome measures.

Chest physiotherapy (CPT) is an ambiguous term which refers to a variety of physiotherapy modalities used in the treatment of patients with underlying cardio-respiratory pathology. This includes airway clearance techniques (ACT), exercise, thoracic mobility exercises, positioning, breathing exercises, and inhalation therapy. Mistakenly, the term, 'CPT' has been used synonymously with the terms postural drainage and percussion with the latter being the traditional method of airway clearance used to treat cystic fibrosis patients until the early 1990s. In order to clarify terminology, the International Physiotherapy Group for Cystic Fibrosis (IPG/CF) has defined chest physiotherapy in the former term.

CPT should now only refer to, 'Cardio-pulmonary Physical Therapy' encompassing a more comprehensive approach to the cardiopulmonary system. Individual techniques are classified under subheadings such as airway

* Tel.: +1 604 875 2123; Fax: +1 604 875 2349.
E-mail address: mmcilwaine@cw.bc.ca.

1526-0542/$ – see front matter © 2007 Elsevier Ltd. All rights reserved.
doi:10.1016/j.prrv.2007.02.013

clearance techniques. The airway clearance techniques currently available to treat patients with cystic fibrosis (CF) include postural drainage and percussion (PD&P), Active Cycle of Breathing Technique (ACBT), Autogenic Drainage (AD), Positive Expiratory Pressure (PEP), High Pressure PEP, Oscillating PEP, High Frequency Chest Wall Oscillation (HFCWO) and Exercise.[1] This review will focus on the techniques of PD&P, ACBT, AD and the use of exercise in the treatment of cystic fibrosis.

The goals of airway clearance techniques in CF are to assist in the removal of airway secretions, thus improving ventilation, reducing airway resistance, correcting ventilation-perfusion mismatch and decreasing the proteolytic activity in the airway. As the mean life expectancy of CF patients increases due to better treatment, the goals of ACTs must also include long-term outcomes such as retarding lung disease and preserving physical function in order to improve quality of life.

COMMENCEMENT OF PHYSIOTHERAPY

Prior to newborn screening, the diagnosis of CF was made as a result of symptoms, either nutritional and/or respiratory. Physiotherapy was initiated immediately as it was considered the mainstay of therapy. However, over the past decade, the number of infants diagnosed with CF through newborn screening has increased worldwide. Often these infants are diagnosed before any recognizable respiratory symptoms appear. Now some CF centres are questioning when to begin physiotherapy in these infants, especially in ones who are apparently asymptomatic.[2]

The question arises as to whether these infants are truly asymptomatic or whether the present clinical tools used to assess the degree of respiratory involvement are inadequate to detect underlying lung pathology. Broncho-alveolar lavage has shown that airway inflammation and infection are present in the CF infant as early as 4 weeks of age.[3–5] Ranganathan[6] found that infants, who were judged asymptomatic on clinical examination, had diminished full and partial forced expiratory maneuvers. Tepper *et al.*[7] states, 'In the absence of respiratory symptoms, the clinical exam and radiological evidence may understate the degree of lung disease'. He concluded that it is not possible to assess a patient as being asymptomatic on the basis of clinical examination alone. One argument for commencing physiotherapy at diagnosis comes from Connett *et al.*[8] They compared the outcomes at 1, 3, 6, and 10 years of 73 CF patients with a delayed diagnosis of CF. The control was a group of CF patients diagnosed within 4 months of age who were commenced on treatment at diagnosis. Treatment consisted of physiotherapy, digestive enzymes, antibiotics and counseling. At 10 years the delayed diagnosis group required increased treatment to maintain their health and had significantly more radiological changes.

Although there is a question of when mucociliary transport becomes impaired in CF patients, we know that CF lung disease begins early in life. For this reason, many members of the multidisciplinary care team believe that physiotherapy should be commenced at diagnosis with the aim of preventing lung disease. This also lays a foundation for infants to accept physiotherapy as part of their daily life. Their parents learn to differentiate between the normal baseline for their infants and any early changes and when they need to communicate with the CF team.

In advocating for physiotherapy to commence at diagnosis, one must carefully balance the burden that is placed on the family to perform this treatment against the gain from the treatment as it must be acknowledged that there is no direct evidence showing the benefit in lung function of commencing ACT at diagnosis. In North America, the ACT most often initiated with newly diagnosed CF infants is a modified form of postural drainage and percussion.

POSTURAL DRAINAGE AND PERCUSSION (PD&P)

In the 1950s, postural drainage and percussion was introduced as a standard part of the CF care,[9] despite any large randomized controlled trials to assess its effectiveness. It includes placing a patient in a gravity dependent position and percussing the chest wall over the area being drained, for typically between 3–5 minutes. The patient is then asked to inhale deeply 3–4 times and on exhalation the chest wall is vibrated; this is followed by directed coughing.[10] There are 12 different postural drainage positions used, based on the nomenclature of the bronchopulmonary anatomy.[11] The physiological rationale for the use of postural drainage to assist in the clearance of secretions is based on the use of gravity to assist with the mucociliary action. In healthy individuals ciliary action moves secretions up the airways at a rate of 3–5 mm/min.[12,13] In people with CF where the mucociliary action is impeded, Wannemaker *et al.* found airway secretions moved slowly in the opposite direction towards the periphery of the lung, but when placed in a head-down gravity dependent position, secretions moved up the airways at the normal rate of 3–5 mm per minute.[13] This study suggests that gravity can assist in the removal of secretions from the periphery of the lungs. However, taking this study one step further, for PD to be effective in moving secretions for the basal segments of the lungs to the bifurcation of the right and left main stem bronchus, (approximately a distance of 30 cm in an adult) a patient would have to be placed in a head down position for between 60–100 minutes. MacKenzie *et al.*[14] in a study on 42 ventilated patients, demonstrated an increase in total lung compliance following a session of postural drainage, percussion and vibration

which lasted for a mean of 57 minutes. He suggested that chest physiotherapy needs to be of 1 hour duration to be effective.

To enhance the clearance of secretions, chest wall clapping or percussion was added to postural drainage. It is thought that percussion generates flow transients in airways beneath a percussed segment that improve the gas-liquid interactions.[15] Sutton *et al*.[16] demonstrated that the addition of percussion to postural drainage enhanced the removal of secretion in patients who have copious secretions. Mechanical percussors have been used to assist the patient and caregivers in performing the treatment with mixed reviews. Bauer *et al*., did not find them as effective as manual percussion.[17] Whereas, Flower[18] showed mechanical percussion increases intrathoracic pressure. In general, mechanical percussors have not been well studied; there is a variety of types on the market operating under different frequencies, the effect of which needs further evaluation.

Manual vibration is a combination of compression and oscillation of the chest wall during expiration. It is considered a component of the PD&P technique, but until recently was not well studied as a separate entity.[19] Recently McCarren and colleagues have been examining the effect of vibration on the respiratory system.[20,21] They found that vibrations increased peak expiratory flow rates (PEFR) by 50% over relaxed expiration. This was greater than the PEFR generated by chest wall compression or chest wall oscillation alone. They also measured changes in intrapleural pressures generated by vibration. With vibration, intrapleural pressure changed by 10 cmH_2O compared to only 7 cmH_2O on relaxed expiration. They concluded that the changes in intrapleural pressures occurring during vibration was the primary generating pressures for the increase PEFR and factors which contribute to this was a combination of lung recoil, compression and oscillation. Surprisingly, the frequency of oscillation of the vibration in the McCarren's study was only 5.5 Hz whereas other investigators have reported vibration creating an oscillation of 10–12 Hz. The work of McCarren *et al*. has greatly added to our understanding of the effects of vibration and in particular on expiratory flow rates. The physiological literature also suggests a rationale to support the use of vibrations with a frequency of <20 Hz. Other authors have demonstrated that vibrations, especially high frequency oscillations can cause a decrease in the viscoelastic properties of mucus.[22] This, together with increased expiratory flow rates seen with vibrations, provide some evidence to support the belief that vibrations improve mucociliary transport.[23]

Detrimental effects of postural drainage and percussion

Until recently, the main deterrents to performing this airway clearance technique were, the burden placed on the family, the requirement of a second person, and the time it took to perform the technique. However, recent studies have suggested that PD&P may have detrimental effects on patients. Both Giles and McDonnell reported hypoxic episodes occurring during PD&P in moderate and severely affected CF patients.[24,25] Button *et al*.,[26] demonstrated that PD&P performed in head-down positions may aggravate gastroesophageal reflux in infants with CF. As a result of this study, Button *et al*. modified PD positions to no tipping. They recently published the results of using these modified non-tip positions over a 5-year period in patients with CF, showing improved outcomes.[27] More recently Elkins[28] reported a decrease in maximum expiratory pressures and peak expiratory flow rates in head-down tilt positions as well as confirming a decrease in oxygenation and reflux scores in these positions. Rib fractures are another complication to be aware of in the very young CF patient with the incidence being reported as 1:1000 infants median age 3 months treated for bronchiolitis and pneumonia.[29] Other adverse reactions to PD&P performed in head-down positions include bronchospasm, changes in cardiac rhythm, and raised intracranial pressures.[30,31]

Many CF centres have now modified the use of PD&P to include only non head-down positioning.[32] This avoids many of the harmful side effects of PD&P. In Canada, head-down positions are no longer used to assist in secretion removal; modified PD positions are being used in infants. With other ACTs, patients are placed in positions to optimize ventilation to specific lung regions. It has been speculated that the redistribution of ventilation, as occurs with a change in body position, might alter the local airway patency and gas/liquid pump.[33,34] Consequently, it can be hypothesized that the physiological basis on which the concept of PD was originally developed may not be the only mechanism for the improvement seen with changes in position as used in PD positions. This hypothesis is partly supported by Lannefors and Wollmer[35] who noted that more secretions were cleared from the dependant lung rather than from the uppermost lung during postural drainage.

Evidence to support use of PD&P

As previously mentioned, PD&P was introduced into the treatment for CF in the 1950s without much scientific evidence to support its use. In 1971, Lorin and Denning were the first to study the effects of PD&P in 17 CF patients.[36] They showed a highly significant difference ($p < 0.0001$) in the volume of sputum produced with PD&P than with directed coughing alone. In 1975, Tecklin and Holsclaw studied 26 patients with CF and reported significant increases in FVC and peak expiratory flow (PEF) but not FEV_1 following PD&P.[37] This was followed by a study in 1983 by Desmond *et al*.[38] who attempted to compare PD&P to no airway clearance except directed

coughing in CF patients. In this study, pulmonary function declined significantly when PD&P was discontinued for a 3-week period, and when PD&P was recommenced pulmonary function returned to baseline. In 1988, the first long-term (3-year) study was published which compared PD&P to huffing and coughing alone.[39] There was a significant greater rate of decline in FEV_1 and FEF_{25-75} in the group who performed huffing and coughing alone ($p < 0.001$). The authors concluded that PD&P should remain a standard component of therapy in CF. They had wanted to compare the techniques of PD&P to no airway clearance technique, but were ethically not allowed to withdraw treatment from the CF participants and hence compared it to a form of the forced expiration technique. A meta-analysis of studies comparing PD&P with no physiotherapy found a significant benefit in favor of PD&P.[40] In 2004, van der Schans in a Cochrane review, attempted to compare any form of airway clearance technique to no treatment.[41] Only six short-term studies were included as any form of control treatments such as directed cough or huffing were excluded. Conclusions suggested that ACTs could have short-term effects in increasing mucus transport, demonstrated by improved mucus expectoration or radioactive clearance. Two other reviews have recently been published. Main and Prasad[42] in a Cochrane review compared PD&P with other ACTs and found no advantage of PD&P over other ACTs in terms of pulmonary function with patients tending to prefer self-administered techniques. Bradley *et al.*[43] in an evidence based review reinforced the findings of van der Schans's and Main's Cochrane reviews. Conclusions from all these studies suggest that there is evidence to support the short-term use of ACTs in the treatment of CF with PD&P being included as one of the ACTs. The problem is that there are no long-term studies comparing PD&P to no treatment and ethically such a study would be difficult to do. We can only draw inferences from the two long-term studies comparing PD&P to huffing or directed coughing which supports the use of PD&P in the treatment of CF.

Recommendations

Future research needs to be directed at trying to understand the physiological principles by which PD&P works and what are the essential components of the technique. Owing to the detrimental effects of PD&P performed in head-down positions and the recent evidence which shows a beneficial effect of using modified positioning, head-down positioning should no longer be used with PD&P.

BREATHING TECHNIQUES

Obstructed lung disease due to plugging of secretions in the airway is one of the main characteristics of cystic fibrosis. To assist in enhancing secretion removal, more recent efforts have been directed at using various breathing strategies which are directed at ventilating the area behind the obstruction and then utilizing increased expiratory flow rates to mobilize secretions up the airways. Various techniques have been developed based on these principles.

Breathing control

Interspersed with any ACT, there needs to be periods of relaxed tidal volume breathing called breathing control. This used to be termed, "diaphragmatic breathing", but due to altered lung mechanics which occurs with obstructed lung disease, the diaphragm is often flattened and at a mechanical disadvantage favoring the use of accessory muscles. Relaxed tidal breathing or breathing control, infers breathing a normal tidal volume in a relaxed position using the lowers chest and if needed accessory muscles with the aims of decreasing the work of breathing and airflow obstruction.[44]

Forced expiration technique (FET)

The forced expiration technique combines huffing with breathing control. Usually one to two huffs (forced exhalations) are performed followed by breathing control to avoid any increase in airway obstruction.[45] Huffing can be performed at various lung volumes. A huff from mid to low lung volume will help mobilize more peripheral secretions. While a huff from a mid to high lung volume will mobilize secretions from the larger, more proximal airways. The effectiveness of the forced expiration maneuver is based on the equal pressure point concept with compression of the airways downstream (towards the mouth) of the equal pressure point occurring on a forced exhalation.[44,46] Compression of the airways where the secretions are located creates a flow-limiting segment that increases gas velocity overcoming the shear forces required to mobilize the secretions stuck to the airway wall. This theory is supported by the work of McCarren[20] who demonstrated that a huff from TLC increased PEFR from 0.66 l/s to 7.76 l/s. This was just slightly less than that produced by a cough (8.14 l/s). Huffing produces less airway compression than coughing and is safer for patients with unstable airways.[46]

ACTIVE CYCLE OF BREATHING TECHNIQUE

The active cycle of breathing technique (ACBT) originated in Britain, and was developed by Webber and Pryor at the Brompton Hospital. It was originally called the forced expiration technique (FET) but was renamed in 1990 to ACBT to emphasize the fact that, in addition to forced expirations, breathing control and thoracic expansion exercises need to be included in the technique. Thoracic expansion exercises consists of 3–4 deep breaths which may or may not include a 3-second inspiratory hold followed by relaxed expiration. Increasing the lung volume

above tidal volume breathing reduces collateral ventilatory resistance, allowing air to flow behind secretions aiding their mobilization, while at the same time forces exerted between adjacent alveoli aid lung re-expansion. Thus ACBT is a cycle of breathing techniques consisting of breathing control, thoracic expansion exercises, and the forced expiration technique.

Several authors have found ACBT to be as effective as PD&P in mobilizing secretions.[40,42,46,47] It is less likely to cause oxygen desaturation[48] and allows more independence than PD&P. Originally it was used as an adjunct to PD&P performed in a postural drainage position, but as the technique has evolved and with improvements in medical management, various studies have been unable to demonstrate any advantage in performing ACBT in a head-down position.[49–51] At the time of this publication, positioning is only being used as a method of increasing ventilation to a specific lung region. ACBT can be introduced as early as four years of age and offers many advantages over PD&P in that it does not require any equipment, can be performed independently anywhere and is less tiring to perform than PD&P.

Recommendations

Breathing exercises can be incorporated into play activities as young as 18 months of age through the use of blowing games. Huffing is an excellent method of mobilizing secretions and can be introduced about 4 years of age. Thus ACBT can be taught to children as young as 4 years and offers another alternative to PD&P in this age group. Positioning should only be used as a method of increasing ventilation to a specific lung region.

AUTOGENIC DRAINAGE (AD)

This technique developed by Chevaillier in Belgium means 'self-drainage'. While observing a number of patients with obstructed lung disease, Chevaillier noted that their normal tidal volume breath was at an increased functional residual capacity (FRC) level. However, once they exhaled into their expiratory reserve volume (ERV), secretions were heard to be mobilized up the airways.

AD is based on a series of principles aimed at normalizing the breathing pattern. Ventilation behind obstructed lung units is achieved by inhaling a normal tidal volume breath followed by a 3-second breath hold. Exhalation is not forced as in FET, but is slightly accelerated to achieve sufficiently high airflow to overcome the shear forces with which the mucus is attached to the airway walls. The aim of the expiratory airflow is to achieve the maximum airflow velocity within the different generations of bronchi without causing compression of the airways. This is achieved by the patient learning to adjust the rate, depth and level of breathing according to their disease state and pulmonary mechanics. They must balance their maximum expiratory airflows against collapse of dynamically unstable airways. Fig. 1 illustrates this principle, showing the changes in a flow-volume loop during maximal forced exhalations and during AD breathing. It is believed that mucus moves centrally at a more rapid rate and for a longer duration when performing AD breathing.[52] The patient is trained to breathe at three lung volumes, beginning at low lung volume to "unstick" the peripheral mucus, moving to low to mid lung volume breathing to 'collect' the mucus in the middle airways and then progressing to mid to high lung volume to 'evacuate' the mucus from the central airways (Fig. 2). Once secretions are expectorated, the sequence is repeated until as much secretions as possible are cleared. AD avoids forceful expirations and high positive transthoracic pressures.

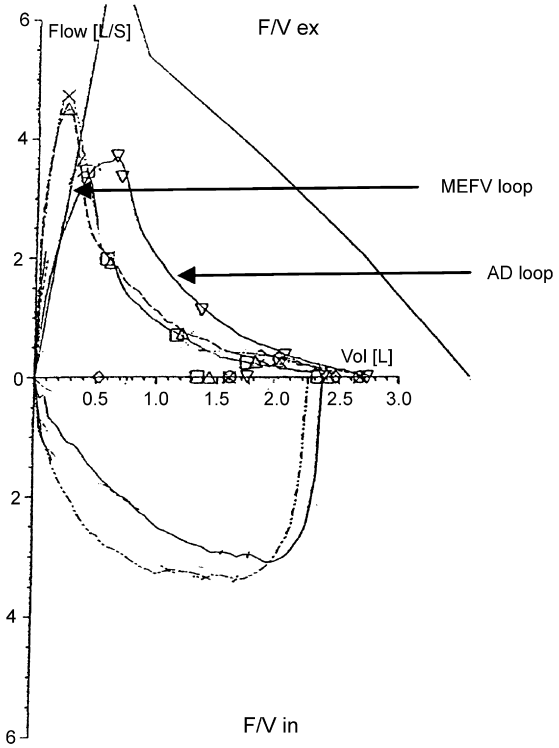

Figure 1 Flow volume loops performed during a maximal expiratory flow manoeuvre and during autogenic drainage.

There are few well powered studies examining the effects of AD in the treatment of CF. What we do know is that AD has been shown to be superior to PD&P and PEP with respect to sputum production[53] and it does not cause oxygen desaturation.[24,54] Thiebl *et al.*[55] demonstrated that AD was safer for patients with hyper-reactive airways in whom high-pressure PEP can cause bronchospasm. In the only long-term study with 36 CF patients comparing PD&P with AD,[56] patients were randomized to receive either AD or PD&P for one year and then cross-over for a second year. AD was found to be as effective as PD&P in maintaining pulmonary function over the first year period. The second year of the study had to be abandoned as patients who had been randomized to AD during the first year refused to change to performing PD&P for the second year as they preferred AD.

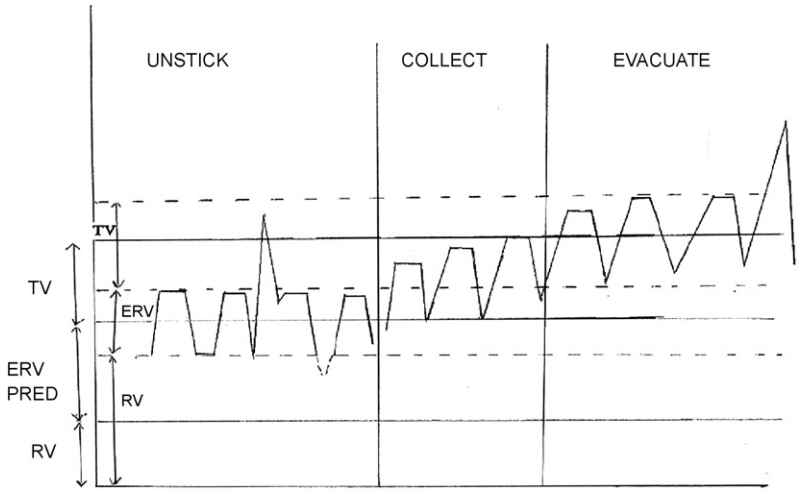

Figure 2 Breathing during Autogenic Drainage.

Recommendations

AD offers many advantages over PD&P, in that it can be performed anywhere independently without any equipment. It is a very effective ACT for patients with hyperreactive airways in whom PD&P or FET may induce more bronchospasm. AD can be difficult to learn as its performance is more an art than a science. It initially requires a high degree of concentration, but once learned, patients are generally able to perform this technique with much less effort. It is therefore not recommended for the younger child or for patients with seriously compromised lung function. To be taught effectively, AD requires a thoroughly trained physical or respiratory therapist who understands how to adapt the technique to the individual patient. As with many of the other ACTs, there still needs to be further long-term studies examining the efficacy of AD in the treatment of CF.

COMPARISON OF ACBT AND AD

Many people are confused between the techniques of ACBT and AD and think they are quite similar when in fact they are two different techniques based on different principles. ACBT uses high transthoracic pressures during forced exhalations and thus only 1–2 huffs can be performed at any time before being interspersed with breathing control. AD uses low transthoracic pressures avoiding airway compression and is a continuous cycle of inhaling a tidal volume breath followed by a controlled exhalation. ACBT ventilates obstructed airways using 3–4 deep breaths which need to be interspersed with breathing control to avoid hyperventilation, whereas AD uses a tidal volume breath with a 3-second breath hold on each consecutive breath. Each technique has their own merit, in that ACBT can be taught at a much younger age and is less tiring to perform in the severely compromised patient, while AD is a continuous normalized breathing pattern and once learned requires less thought. In the one and only comparative trial of ACBT versus AD, Miller and colleagues found both ACBT and AD equally effective in clearing secretions although AD cleared secretions at a faster rate as measured by xenon-133 ventilation studies.[57]

EXERCISE

Exercise is increasingly regarded as an essential part of the overall physiotherapy management of CF and would require a separate review to adequately describe its role and virtues. As the focus of this review is on airway clearance, this section will be limited to discussing the role of exercise as an airway clearance technique. Some proponents have suggested that exercise alone can be used as an ACT, but evidence to support this belief is limited with only one short-term in-patient study with limited number of patients demonstrating similar outcomes in those who received exercise and one daily session of PD&P per day compared to three daily sessions of PD&P.[58] A previous 3-week study performed at a CF camp was able to demonstrate an improvement in lung function when a vigorous exercise program of 6 hours a day was undertaken; however this much exercise is impractical in daily life.[59] Zach *et al*. demonstrated that exercise in the form of a swimming program can enhance secretion clearance.[60] In a meta-analysis of airway clearance techniques which included exercise, Thomas *et al*. reported exercise alone to be less effective than PD&P in mobilizing secretions, but as a complement exercise induces a significant increase in secretion clearance.[40] In two other studies by Salh *et al*.[61] and Bilton *et al*.,[62] physiotherapy plus exercise produced significantly more sputum than physiotherapy alone and exercise alone. There have been no long-term randomized controlled trails comparing exercise alone to any other ACT. The conclusion of all these publications is that

exercise plays a role in secretion clearance but should not be considered a replacement for an ACT.

On the other hand, there have been a number of studies examining the benefits of exercise as part of the overall physiotherapy management of the CF patient with very positive results. Two systematic reviews have summarized the effects of exercise for patients with CF.[40,63] They concluded that exercise in addition to PD&P, has beneficial effects on FVC and FEV_1. Exercise has the added advantage of improving general fitness, patient's self-esteem and measures of quality of life. Exercise is socially acceptable and helps to normalize the patient's life rather than adding a therapy that accentuates differences from peers.[64]

The prognostic importance of exercise testing is gaining importance after Nixon *et al.*[65] demonstrated that exercise capacity was an independent predictor of life expectancy in CF patients. Exercise testing is also an important outcome measure which is often underutilized in studies, but which may be more sensitive to long-term survival compared to pulmonary function parameters. One of the problems in performing exercise testing, is that many CF centres do not have the specialized laboratory testing equipment or manpower necessary to assess aerobic/anaerobic fitness. For these reasons, field exercise tests such as the shuttle test, or 6-minute walk test are being introduced.[66] Although they have been validated, there is still little data on normative values and what represents a significant change.[49]

CONCLUSIONS

Cystic fibrosis is a multifaceted disease with a high degree of variability in lung disease. This combined with patient's age, individual preferences, culture and motivation, mean that no one type of airway clearance technique will suit all patients. ACTs need to be individually adapted to suit the patient's and family's needs. There are short- to medium-term studies to support the use of ACTs in CF but only a few long-term studies. In a chronic illness, it is difficult to infer efficacy for one ACT over a lifetime, particularly since no one ACT has been shown to be superior to another ACT.[67] As many ACTs as possible should be taught to the patient, so that they will have a choice of ACTs based on the previously mentioned factors. If PD&P is the method of choice, only modified positioning should be used. Exercise should not be regarded as a form of airway clearance, but rather an adjunct which complements ACT.

In trying to evaluate ACTs, there are many methodological barriers. ACTs are not well standardized, they vary not only in regard to the specifics of the techniques but also the frequency, timing, duration and intensity. These methodological problems are exacerbated by variations in the criteria for patient selection, compliance monitoring and use of concurrent medications. All these factors make it difficult to design and conduct rigorous clinical trials to resolve contradictory results. In addition, it is not possible to conduct double-blind studies of ACT or ethically is it allowed to compare an ACT to no ACT in CF patients.

PRACTICE POINTS

- There is limited evidence to support the use of airway clearance techniques in the treatment of cystic fibrosis, but a lack of knowledge as to when it should be commenced.
- There are associated risks associated with performing postural drainage and percussion.
- Newer airway clearance techniques such as Active cycle of breathing and Autogenic drainage have been demonstrated to be as effective as postural drainage and percussion and offer many advantages.
- There is a lack of evidence to support the use of exercise as an airway clearance technique, although it contributes to the overall well-being of the CF patient.

RESEARCH DIRECTIONS

- Since the life expectancy in CF has increased over the past decade with a corresponding slower rate of decline in pulmonary function, studies need to be multi-centered of 2–3 years in length to show any statistical effect.[64]
- We need to decide which outcome measures are the most appropriate to use. In large clinical trials we may want to use pulmonary function measures or exercise capacity whereas if we are examining the physiological effect of a technique, mucociliary clearance index or expiratory flow may be more appropriate.
- The rational for commencing physiotherapy at diagnosis, needs further investigation.
- We still need to conduct small studies assessing the physiological effect of an ACT. McCarren[20] suggests that for airway clearance to be effective we need to have an expiratory bias to airflow. If this is true, then it may be that we still have not designed the best method of airway clearance for our CF patients.
- We know that the interaction between lung mechanics and the state of the individual's lung disease plays an important role in how a person with CF responds to an ACT. We need to determine which ACT is most suited to the individual patient based on their lung disease and lung mechanics. From this research it would be helpful to be able to provide some guidelines to healthcare professionals on the use of ACTs for the individual with CF.

REFERENCES

1. International Physiotherapy Group for Cystic Fibrosis. Physiotherapy in the treatment of Cystic Fibrosis. 2001 (3rd ED). Available from Cystic Fibrosis Worldwide. www.Cfww.org.
2. Wallis C, Prasad A. Who needs chest physiotherapy? Moving from anecdote to evidence. *Arch Dis Child* 1999; **80**: 393–397.
3. Khan TZ, Wagener JS, Bost T, Martinez J, Accurso FJ, Riches DWH. Early pulmonary inflammation in infants with cystic fibrosis. *Am Rev respir Crit Care Med* 1995; **151**: 1075–1082.
4. Armstrong DS, Grimwood K, Carlin JB *et al.* Lower airway inflammation in infants and young children with cystic fibrosis. *Am J Respir Crit Care Med* 1997; **156**: 1197–1204.
5. Nixon GM, Armstrong DS, Carzino R *et al.* Early airway infection, inflammation and lung function in cystic fibrosis. *Arch Dis Child* 2002; **87**: 306–311.
6. Ranganathan SC, Bush A, Dezateux C. Relative ability of full and partial forced expiratory maneuvers to identify diminished airway function in infants with cystic fibrosis. *Am j Respir Crit Care Med* 2002; **166**: 1350–1357.
7. Tepper Rs. Assessment of the respiratory status of infants and toddlers with cystic fibrosis. *J Pediatr* 1998; **132**: 380–381.
8. Connett GJ, Yeatman SL. Delayed diagnosis in cystic fibrosis patients homozygous for Delta F508 mutation results in increased treatment to maintain health in childhood. *Pediatr Pulmonol* 2002; Suppl 24): 318 (abst 407).
9. Matthews LW, Doershuk CF, Wise M, Eddy G, Nudelman H, Spector S. A therapeutic regimen for patients with cystic fibrosis. *J Pediatr* 1964; **65**: 558–575.
10. McIlwaine M. Postural drainage and percussion. In: International Physiotherapy Group for Cystic Fibrosis. Physiotherapy in the treatment of Cystic Fibrosis. 2001 (3rd ED). Available from Cystic Fibrosis Worldwide. www.Cfww.org.
11. Negus VE, Brock RC, Foster-Carter F. The nomenclature of bronchopulmonary anatomy. *Thorax* 1950; **5**: 222–228.
12. Wood RE, Wanner A, Hirsch J, Farrell PM. Tracheal mucociliary transport in patients with cystic fibrosis and its stimulation by terbutaline. *Am Rev Resp Dis* 1975; **111**: 733–738.
13. Wong JW, Keens TG, Wannemaker EM, Douglas PT, Crozier N, Levison H. Effects of gravity on tracheal mucus transport rates in normal subjects and in patients with cystic fibrosis. *Pediatrics* 1977; **60**: 146–152.
14. MacKenzie MB, Shin B, Hadi F, Imle PC. Changes in total lung/thorax compliance following chest physiotherapy. *Anaesthesia and Analgesia* 1980; **59**: 207–210.
15. Hardy KA. A review of airway clearance: New techniques, indications and recommendations. *Resp Care* 1994; **39**: 440–452.
16. Sutton PP, Lopez-Vidriero MT, Pavia D, Newman SP *et al.* Assessment of percussion, vibratory-shaking and breathing exercises in chest physiotherapy. *Eur J Resp Dis* 1985; **66**: 147–152.
17. Bauer ML, McDougal J, Schoumacher RA. Comparison of manual and mechanical chest percussion in hospitalized patients with cystic fibrosis. *J Pediatr* 1994; **124**: 250–254.
18. Flower KA, Eden RI, Lomax L, Mann NM, Burgess J. New mechanical aid to physiotherapy in cystic fibrosis. *BMJ* 1979; **2**: 630–631.
19. Thomas J, DeHueck A, Kleiner M, Newton J, Crowe J, Mahler S. To vibrate or not to vibrate: Usefulness of the mechanical vibrator for clearing bronchial secretions. *Physiotherapy Canada* 1995; **47**: 120–125.
20. McCarren B, Alison JA, Herbert RD. Vibration and its effect on the respiratory system. *Aust J Physiother* 2006; **52**: 39–43.
21. McCarren B, Alison JA, Herbert RD. Manual vibration increases expiratory flow rate via increased intrapleural pressure in healthy adults: an experimental study. *Aust J Physiother* 2006; **52**: 267–271.
22. King M, Phillips DM, Gross D. Enhanced tracheal mucus clearance with high frequency chest wall compression. *Am Rev Respir Dis* 1983; **128**: 511–515.
23. McCarren B, Alison JA. Physiological effects of vibration in subjects with cystic fibrosis. *Eur Respir J* 2006; **27**: 1204–1209.
24. Giles DR, Wagener JS, Accurso FJ, Butlersimon N. Short term effects of postural drainage with clapping versus autogenic drainage on oxygen saturation and sputum recovery in patients with cystic fibrosis. *Chest* 1995; **108**: 952–954.
25. McDonnell T, McNicholas WT, Fitzgerald MX. Hypoxia during chest physiotherapy in patients with cystic fibrosis. *Ir J Med Sci* 1986; **155**: 345–348.
26. Button BM, Heine RG, Catto-Smith AG, Phelan PD, Olinsky A. Postural drainage and gastro-oesophageal reflux in infants with cystic fibrosis. *Arch of Dis in Child* 1997; **76**: 148–150.
27. Button BM, Heine RG, Catto-Smith AG, Olinsky A, Phelan PD, Ditchfield MR. Chest physiotherapy in infants with cystic fibrosis. *Pediatr Pulmonol* 2003; **35**: 208–213.
28. Elkins MR, Alison JA, Bye PT. Effect of body position on maximal expiratory pressure and flow in adults with cystic fibrosis. *Pediatr Pulmonol* 2005; **40**: 385–391.
29. Chalumeau M, Foix-L'Helias L, Scheinmann P, Zuani P. Rib fractures after chest physiotherapy for bronchiolitis or pneumonia in infants. *Pediatr Radiol* 2002; **32**: 644–647.
30. Campbell AH, O'Connell JM, Wilson M. The effect of chest physiotherapy upon the FEV_1 in chronic bronchitis. *Med J Aust* 1975; **1**: 33–35.
31. Naylor JM, Chow CM, McLean AS. Cardiovascular responses to short-term head-down positioning in healthy young and older adults. *Physiother Res Int* 2005; **10**: 32–47.
32. Lannefors L, Button BM, McIlwaine M. Physiotherapy in infants and young children with cystic fibrosis: current practice and future developments. *J Royal Soc Med* 2004; **97**(Suppl 44): 8–25.
33. Menkes H, Britt J. Rationale for physical therapy. *Am Rev Respir Dis* 1980; **122**(Suppl 2): 127–128.
34. Oberwaldner B. Physiotherapy for airway clearance in paediatrics. *Eur Respir J* 2000; **15**: 196–204.
35. Lannefors L, Wollmer P. Mucus clearance with three chest physiotherapy regimes in cystic fibrosis. A comparison between postural drainage, positive expiratory pressure and physical exercise. *Eur Resp J* 1992; **5**: 748–753.
36. Lorin MI, Denning CR. Evaluation of postural drainage by measurement of sputum volume and consistency. *Am J Phy Med* 1971; **50**: 215–219.
37. Tecklin J, Holsclaw D. Evaluation of bronchial drainage in patients with cystic fibrosis. *Phys Ther* 1975; **55**: 1081–1084.
38. Desmond KJ, Schwenk WF, Thomas E, Beaudry PH, Coates A. Immediate and long-term effects of chest physiotherapy in patients with cystic fibrosis. *J Pediatr* 1983; **103**: 538–542.
39. Reisman JJ, Rivington-law B, Corey M *et al.* Role of conventional physiotherapy in cystic fibrosis. *J Pediatr* 1988; **113**: 632–636.
40. Thomas J, Cook DJ, Brooks D. Chest physiotherapy management of patients with cystic fibrosis. *Am J respire Crit Care Med* 1995; **151**: 846–850.
41. Van der Schans C, Prasad A, Main E. Chest physiotherapy compared to no physiotherapy for cystic fibrosis (Cochrane Review). The Cochrane Library 2004; 4: John Wiley & Sons, Ltd, Chichester.
42. Main E, Prasad A, van der Schans C. Conventional chest physiotherapy compared to other airway clearance techniques for cystic fibrosis. The Cochrane Library 2005; 1: John Wiley & Sons Ltd, Chichester.
43. Bradley JM, Moran FM, Elborn JS. Evidence for physical therapies (airway clearance and physical training) in cystic fibrosis: An overview of five Cochrane systematic reviews. *Respir Med* 2006; **100**: 191–201.
44. Partridge C, Pryor J, Webber B. Characteristics of the forced expiration technique. *Physiotherapy* 1989; **75**: 193–194.
45. van der Schans CP. Forced expiratory maneuvers to increase transport of bronchial mucus: a mechanistic approach. *Monaldi Arch Chest Dis* 1997; **52**: 367–370.
46. Pryor JC, Webber BA, Hodson ME, Batten JC. Evaluation of the forced expiration technique as an adjunct to postural drainage in treatment of cystic fibrosis. *Br Me J* 1979; **2**: 417–418.

47. van Hengstum M, Festen J, Beurskens C, Hankel M. Conventional physiotherapy and forced expiration maneuvers have similar effects on tracheobronchial clearance. *Eur Respir J* 1988; **1**: 758–761.
48. Webber BA, Hodson ME. Effect of chest physiotherapy on oxygen saturation in patients with cystic fibrosis. *Thorax* 1990; **45**: 77.
49. Pryor JA, Main E, Agent P, Bradley JM. Physiotherapy. In: Bush A, Alton EWFW, Davies JC, Griesenbach U, Jaffe A. eds: *Cystic Fibrosis in the 21st Century*. Prog Respir Res. vol 34, Basel, Karger, 2006; pp. 301–308.
50. Verboon JML, Bakker W, Sterk PJ. The value of the forced expiration technique with and without postural drainage in adults with cystic fibrosis. *Eur J Respir Dis* 1986; **69**: 169–174.
51. Cecins NM, Jenkins SC, Pengelley J, Ryan G. The active cycle of breathing techniques – to tip or not to tip? *Respir Med* 1999; **93**: 660–665.
52. Schoni MH. Autogenic drainage: a modern approach to physiotherapy in cystic fibrosis. *J Royal Soc Med* 1989; **2**(Suppl 16): 32–37.
53. McIlwaine PM, Davidson AGF, Wong LTK. Comparison of positive expiratory pressure and autogenic drainage with conventional percussion and drainage therapy in the treatment of cystic fibrosis. *Pediatr Pulmonol* 1988; (Suppl 2): 137.
54. McIlwaine PM, Davidson AGF, Wong LTK, Pirie G. The effect of chest physiotherapy by postural drainage and autogenic drainage on oxygen saturation in cystic fibrosis. *Pediatr Pulmonol* 1991; (Suppl 6): 291.
55. Theibl B, Pfleger A, Oberwaldner B, Zach M. Self-administered chest physiotherapy in cystic fibrosis: a comparative study on high pressure PEP and autogenic drainage. *Lung* 1992; **170**: 323.
56. Davidson AGF, Wong LTK, Pirie GE, McIlwaine PM. Long-term comparative trial of conventional percussion and drainage physiotherapy versus autogenic drainage in cystic fibrosis. *Pediatr Pulmonol* 1992; Suppl. 6): 298 (abst 235).
57. Miller S, Hall DO, Clayton CB, Nelson R. Chest physiotherapy in cystic fibrosis: a comparative study of autogenic drainage and the active cycle of breathing techniques with postural drainage. *Thorax* 1995; **50**: 165–169.
58. Cerny FJ. Relative effects of bronchial drainage and exercise for in-hospital care of patients with cystic fibrosis. *Phys Ther* 1989; **69**: 633–639.
59. Zach MS, Purrer B, Oberwaldner B, Hausler F. Cystic Fibrosis: physical exercise versus chest physiotherapy. *Arch Dis Child* 1982; **57**: 587–589.
60. Zach M, Purrer B, Oberwaldner B. Effect of swimming on forced expiration and sputum clearance in cystic fibrosis. *Lancet* 1981; **2**: 1201–1203.
61. Salh W, Bilton D, Dodd M, Webb AK. Effect of exercise and physiotherapy in aiding sputum expectoration in adults with cystic fibrosis. *Thorax* 1989; **44**: 1006–1008.
62. Bilton D, Dodd ME, Abbot JV, Webb AK. The benefit of exercise combined with physiotherapy in the treatment of adults with cystic fibrosis. *Respir Med* 1992; **86**: 507–511.
63. Bradley JM, Moran FM, Elborn JS. Evidence for physical therapies (airway clearance and physical training) in cystic fibrosis. An overview of five cochrane systematic reviews. *Respir Med* 2006; **100**: 191–201.
64. Orenstein DM, Higgins LW. Update on the role of exercise in cystic fibrosis. *Curr Opin in Pulm Med* 2005; **111**: 519–523.
65. Nixon PA, Orenstein DM, Kelsey SF, Doerschuk CF. The prognostic value of exercise testing in patients with cystic fibrosis. *New Eng J Med* 1992; **327**: 1785–1788.
66. Selvadurai HC, Cooper PJ, Meyers N, Blimkie CJ, Smith L, Mellis CM, Van Asperen PP. Validation of shuttle tests in children with cystic fibrosis. *Pediatr Pulmonol* 2003; **35**: 133–138.
67. Pryor JA, Tannenbaum E, Cramer D, Scott SF, Burgess J, Gyi K, Hodson ME. A comparison of five airway clearance techniques in the treatment of people with cystic fibrosis. *J Cystic Fibrosis* 2006; **5**: S77 (abst 349).