

# Scrapie

Extended version

**Classic case:** Adult sheep; weight loss, incoordination, behavior change, **pruritis** & wool loss

## Presentation:

- Adult sheep, black-faced breeds in US – 96% of cases
- Suffolk, Hampshires and crosses
- Many breeds in other countries
- Moufflon (*Ovis musimon*), a wild sheep breed
- Rare, but is seen in goats
- Usually > 2 years of age, incubation is 2-5 years
- Death within weeks to months once C/S present



Scrapie in Rambouillet-Cheviot ewe;  
Image by Dr. ML Crocheck,  
APHIS-USDA-VS-NVSL

Clinical signs may vary:

- Weight loss w/ normal appetite
- **Progressive neurologic signs:**
  - Fine head tremors
  - Ataxia, incoordination-1<sup>st</sup> seen in hinds; 'bunny-hopping'. In fores; 'high-stepping', prancing.
  - **Intense pruritis** (70% of cases), hypersensitivity- nibble at legs or at air
  - Chewing motions, lip-smacking seen with stimulation
  -
- Behavior changes, separation from flock
- Trembling, hyperexcitable, even convulsions if handled
- **Wool loss**, due to rubbing
- Eventually, recumbency and death

***Pruritis is the classic sign of Scrapie; seen in ~70%;  
Sheep rub skin constantly – scraping off their wool***

## DDx:

Dentition problems  
External parasitism  
Listeriosis  
Meningitis

Toxic encephalopathy  
Johne's disease  
Pseudorabies  
Caseous lymphadenitis

Ovine progressive pneumonia (visna)  
Abomasal emptying disease  
Tick borne encephalitis

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**Test of choice:** Must detect prion proteins in tissue

Post mortem tests – most common

- Histopathology - characteristic changes - vacuoles, plaques - now replaced by:
- **Immunohistochemistry (IHC)** - worldwide **gold standard**
  - Brain tissue, most often the OBEX – post-mortem
  - Cerebellum, cortex for 'atypical' Scrapie
  - Lymphoid tissues – PrP<sup>Sc</sup> seen here first in some cases
- **ELISA for screening** - Brain or lymphoid tissues
- Western Blot when tissues are autolyzed

**Antemortem tests: becoming more common**, but false negs seen

- **Biopsy of lymphoid tissue inside 3<sup>rd</sup> eyelid** – IHC
- Biopsy of tonsils – used in Europe – IHC or ELISA
- Detection of prions in placenta – used as herd screening test

**Treatment:**

None; Euthanasia

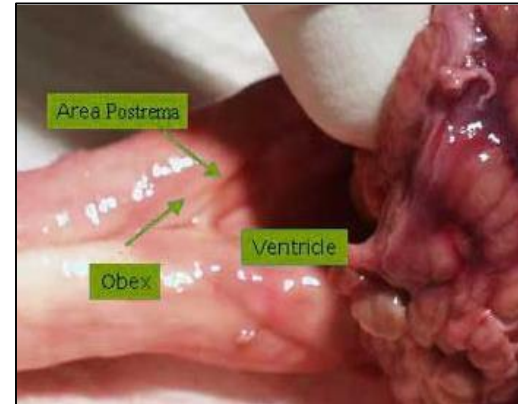
*No serology for Scrapie or other TSE's –  
No immune response = No antibodies!*

**Prognosis:**

Grave, always fatal disease

**Prevention:** See Scrapie Eradication program below

- Purchase and breed only genetically resistant sheep
- Do not feed ruminant proteins to other ruminants
  - Banned in most countries since 90's
- Maintain closed herds; Euthanize positive sheep
- Test all sheep that die with neurologic disease
- Caesarian section in positive dams – not practical
- Carcasses – Incineration or alkaline digestion
- Sodium hydroxide or other approved disinfectant
- *Take GREAT care - handling or transporting tissues*



OBEX – in brainstem where 4<sup>th</sup> ventricle narrows caudally; Image courtesy of CDC



Scrapie in a Montedale ewe;  
Note weight loss w/ obvious dorsal spine;  
depressed, head down posture;  
Image courtesy USDA



Hair loss over pelvis from rubbing;  
Image courtesy USDA

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**Pearls:** Scrapie is a transmissible spongiform encephalopathy (TSE)

TSEs are caused by 'misfolded' *prion* proteins - **Prions (PrP)**

PRIONS	SCRAPIE - PrP <sup>Sc</sup>
<ul style="list-style-type: none"> <li>• <b>Altered form of a <u>normal</u> protein</b></li> <li>• Same primary a.a. sequence</li> <li>• Secondary structure – 'misfolded'</li> <li>• PrPs – <i>very hard to kill</i> <ul style="list-style-type: none"> <li>▪ Protease &amp; disinfectant resistant</li> <li>▪ <b>Not 'foreign' - no immune response</b></li> </ul> </li> <li>• Prions 'reproduce' - convert normal proteins to PrP's</li> </ul>	<ul style="list-style-type: none"> <li>• Transmission - contact w/placenta/allantoic fluid</li> <li>• Usually soon post parturition in lambs Not in utero – amnion is barrier</li> <li>• Direct contact between sheep</li> <li>• Fomites, environment - less common</li> <li>• <i>Genetic susceptibility required</i> in ovine <ul style="list-style-type: none"> <li>▪ Polymorphism at codons 136,154,171</li> <li>▪ ARR/ARR most resistant genotype</li> <li>▪ VRQ/VRQ most susceptible</li> </ul> </li> </ul>

USA Mandatory Scrapie Eradication Program – Goal - *Scrapie-Free by 2017*

- Breeding sheep leaving original home – must be identified – individual and premise origin
- Slaughter surveillance since 2003 – sheep with signs of Scrapie are tested
- Positives - Quarantine farm of origin, test all sheep; all sheep in contact traced & tested

Many US producers also following voluntary guidelines to Eradicate Scrapie

Oldest TSE known – ID over 250 yrs ago

- Worldwide distribution; first reported in US 1947
- Sheep and goats only natural hosts

'Atypical' Scrapie – Nor98 - first ID in Norway 1998

- May be a 'spontaneous' version of Scrapie
- Not spread via ingestion or contact

**Refs:** Merck Vet Manual, 19<sup>th</sup> Ed., Online: Overview of Scrapie; Pugh & Baird Diseases of Sheep & Goats, 2<sup>nd</sup> ed., pp. 385-7.

**My Notes:**