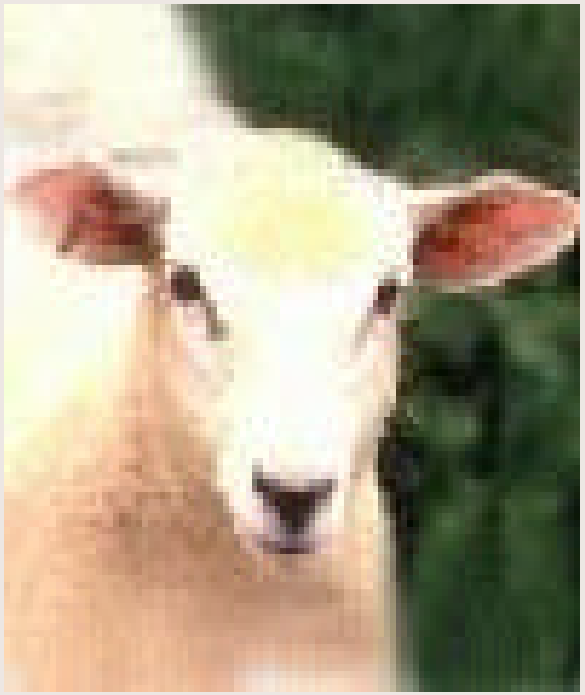
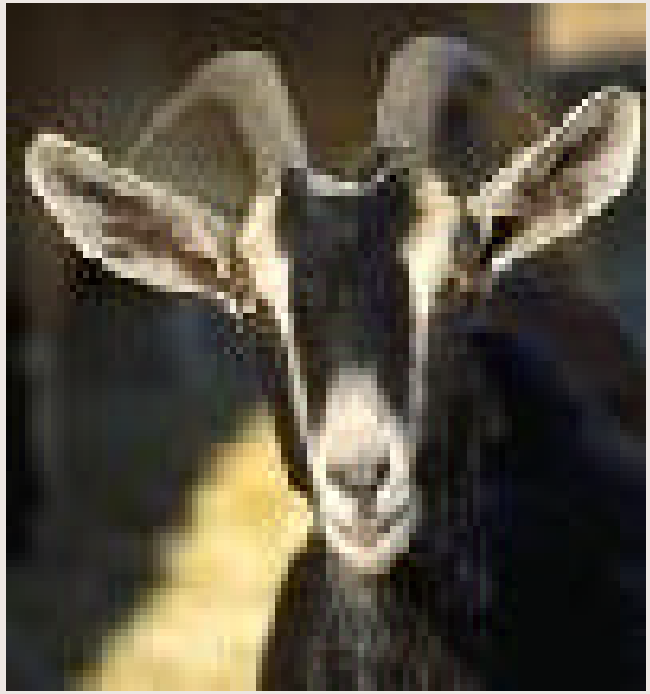
Scrapie





By

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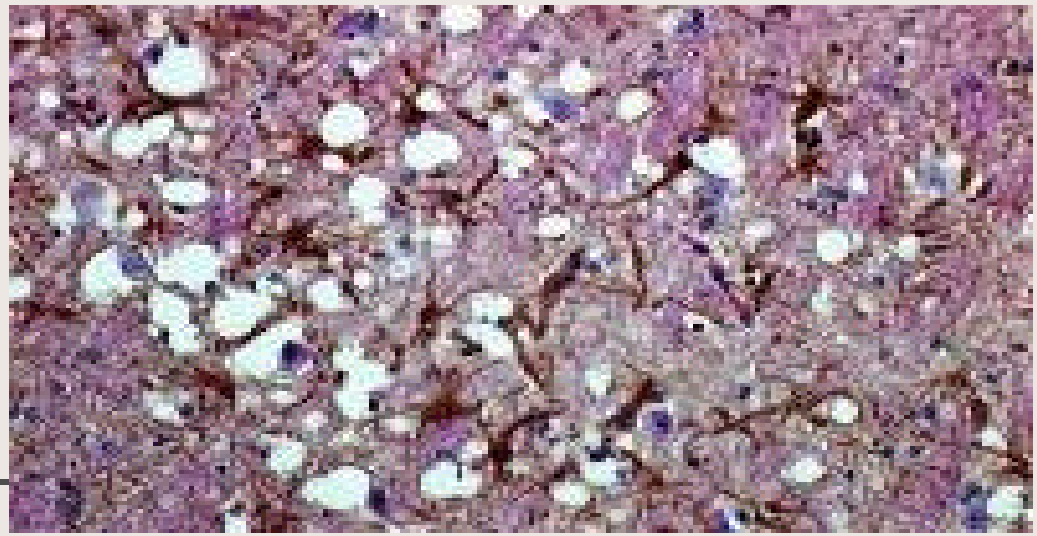
What is it?

- ✓ A fatal neurodegenerative disease of sheep and goat
- ✓ Very Infectious
- ✓ Scrapie attacks the brain and causes it to start looking like a sponge
- ✓ The name reflects the intense itching it causes; many sheep scratch themselves against fencepost or other handy objects

The typical sponge-like appearance of brain parenchyma



Scrapie (sheep)

How did it start?

✓ First recognized as a disease of sheep in UK and Western Europe over 200 years ago

✓First case of scrapie in the US, 1947

✓1281 sheep in 850 flocks has been reported (Sept 30, 1996).

✓Effect on global market place is far greater

✓Australia and New Zealand, free of scrapie

Causative Agent

✓ Prions (PrP^{Sc})

✓ Proteinaceous infectious particles that lack nucleic acid (neither DNA or RNA)

✓ Composed largely, if not entirely of an abnormal isoform of a normal cellular protein (PrP)

Prions

✓Small enough to pass through a filter that stops most viruses

✓Can survive treatments that killed most viruses

✓Not effected by nuclease, proteases, formalin or boiling

✓Inactivated by 1 N NaOH, 4.0 M guanidinium hydrochloride, sodium hypochlorite, or steam autoclaving at 132C for 4.5 hours (highly heat resistant)

Structure of PrP^{Sc}

✓In the lower middle range in size (35,000 Daltons)

✓ Octapeptide coding repeat region

✓ Two asparagine-linked sugar moieties enclosed within a disulphide bridge and a glycolipid membrane anchor

✓Primary structure is identical in normal and diseased states, except conformational change from alpah-helix to beta-sheet rich

Related Disease

The mammalian prions cause scrapie and other related neurodegenerative diseases of humans and animals.

- ✓Scrapie – sheep & goat
- ✓Transmissible mink encephalopathy (TME) – mink
- ✓Chronic wasting disease (CWD) – mule deer and elk
- ✓Bovine spongiform encephalopathy (BSE) – cattle
- ✓Feline spongiform encephalopathy (FSE) – cats
- ✓Kuru – humans
- ✓Creutzfeldt-Jakob disease (CJD) - humans

Symptoms

✓Rubbing themselves against fences and other objects to relieve the itching, and scrape off wool

✓Restless, chew their own skin and excitable

✓Walk unsteadily

✓Suffer from thirst

✓Weak, become paralyzed and then die

Picture of a sheep affected with Scrapie (July 29th, 1998)

What breeds get the disease?

- 

✓Over 90% of confirmed scrapie cases in the US have been in black-faced sheep.

✓In other countries, it occur mostly in white-faced breeds.

Where is it found?

✓Highest concentration of prions in the Central Nervous System and its covering

✓Also in spleen, thymus, lymph nodes and lungs

✓Pockets of lymphoid tissue in sheep's nictitating membrane or third eyelid (used in live-animal test)

Mode of Transmission

Suspected mode of transmission

✓Consumption of prion-infected food

✓From animal to animal (direct contact)

✓Species to species (scientist has proven that scrapie can pass between both animals and species by injecting pureed brain of sick animals into healthy animals)

Transmission to human??

Not proven one way or the other.

Diagnosis

✓Very long incubation period, similar clinical signs from external parasites, listeriosis, plant poisoning, rabies, etc..

✓Therefore, clinical signs must be confirmed by microscopic examination of brain tissue or by immunohistochemistry

✓Does not stimulate the immune system, there is no current blood test for preclinical diagnosis

✓Live animal test are still developing (monoclonal antibody)

Treatment & Prevention

✓Because the means of natural transmission are still unknown, no adequate control measures are available

✓Federal scrapie control program, Voluntary Scrapie Flock Certification Program (VSFCP)

✓Prevention is based on maintaining a closed flock

✓Strict regulation for the handling of animal carcasses to protect farmers

Research

✓The mechanism by with a normal protein is converted to PrP^{sc}

✓Live animal test for early diagnosis (using prion protein collected in sheep's thrid eyelid)

✓Breeding sheep with scrapie-resistant genes

Summary

- ✓ An infectious disease of sheep and goats
- ✓ Causative agent, PrP^{sc}
- ✓ Means of natural transmission are still unknown
- ✓ No cure yet