Known prion Diseases

Human Diseases:
- Creutzfeldt-Jakob Disease (CJD)
- Variant Creutzfeldt-Jakob Disease (vCJD)
- Gerstmann-Straussler-Scheinker Syndrome
- Fatal Familial Insomnia
- Kuru

Animal Diseases:
- Bovine Spongiform Encephalopathy (BSE, aka “Mad Cow Disease”) - cows
- Chronic Wasting Disease (CWD) - deer, elk, and moose
- Scrapie - goats and sheep
- Transmissible mink encephalopathy - mink
- Feline spongiform encephalopathy - felines
- Ungulate spongiform encephalopathy - exotic ungulates

What is Scrapie?
- Scrapie is a TSE (Transmissible Spongiform Encephalopathy)
- Caused by a prion (pathogenic protein)
- Leads to death of infected victim
- Scrapie was the first recognized prion disease
- Prevalence in sheep has went down in US by 99% since 2003
- Last known case of scrapie in goats was detected in 2015
When diseased prion protein (PrPSc protein) comes in contact with a healthy prion protein (PrPC protein) then the healthy protein changes shape and becomes diseased and also infectious to other healthy prion proteins.
Certain prion diseases, such as CJD, kuru, and BSE are only somewhat transmissible, if transmissible at all. They are generally transmitted by contaminated food, or for some cases, genetically.

Scrapie, on the other hand, appears to spread readily and rapidly from individual to individual.

- Routes of infection include: contaminated skin, feces, urine, mucosal secretions, or placental remnants
- There is also a level of persistence within the environment not seen with BSE
- Scrapie spreads easily probably due to lymph tissue involved with infection
Known Species of Goats Affected with Scrapie (5):

- Alpine
- Angora
- Boer
- Nigerian Dwarf
- Nubian
- Saannen
- Toggenburg Cross
- Other crosses

Symptoms of Scrapie in Domestic Goats:

- Tremors (notably of head and neck)
- Gait abnormalities including loss of coordination, high stepping, and swaying of back end
- Pruritus (urge to scratch)
- Weight loss with retained appetite
- Biting of extremities (6)
- Head pressing
- Star-gazing
- Sensitivity to sudden noise or movement
- Difficulty rising (7)
Diseases with Similar Symptoms

Because there are many different diseases that can cause scrapie-like symptoms, it is important to have goats tested for scrapie if no other diagnosis is confirmed.

- listeriosis
- exposure to toxins
- pregnancy toxemia
- pneumonia
- rabies
- lice
- mites
- Goat polio
- heavy worm loads
- Johnes disease.
Diseases with Similar Symptoms

Linignathus stenopsis goat sucking louse
If you have a goat that exhibits symptoms of scrapie that is over 12 months of age, the Animal and Plant Health Inspection Service will cover the cost of shipping labels, shipping of tissues, and testing of the tissues. They will pay for the testing of up to 30 animals from a flock per year.

This means there is no cost to the producer to send in samples of suspected scrapie cases (up to 30 animals) (8).
Why does Scrapie matter to you?

- Costs $10-20 million (or more) in lost revenue due to exportation loss and production cost increases
- Many countries including Canada ban the import of sheep and goats unless the animal comes from a farm enrolled in the voluntary scrapie eradication program
- Although scrapie has never been proven to be transmissible to humans, the WHO has recommended all countries attempt to remove prion diseases from animals raised for meat (9).
Why does Scrapie matter to you?

- Europe and Eastern Bloc countries have banned all live sheep and goats, semen, and embryos from being imported from the United States. This is a huge potential market for breeding stock in both the goat and sheep industry.
- China and Japan ban the import of sheep and goat bone meal and tallow.
- Israel, a previously active importer of US goats, banned their import due to confirmed scrapie cases.
- Mexico, a huge importer of US products, requires all rendered meat to be extensively heat treated before import (10).
- Possibility of BSE infecting goats/sheep.
Surveillance activities

- Enhancing awareness of the disease
- Ensuring samples from targeted and clinical animals reach approved laboratory testing facilities
- Expanding existing slaughter surveillance to include new collection sites
- Increase efforts to reach underrepresented flocks and geographical areas
- Increase compliance with identification requirements (7)

Goal of Scrapie Surveillance: Eradicate Classical Scrapie in the United States
US Scrapie Eradication Program

Identification of Goats
Goats that change ownership or cross state lines must be identified with the Scrapie Eradication Program.

Exceptions
Goats raised for meat, fiber, or not for breeding purposes. Castrated goats also do not need to be identified.

Means of Identification
Ear tags, tattoos, and microchips. Ear tags and microchips must be sufficiently tamper proof and provide a unique identification to each animal.
US Scrapie Eradication Program

**Recordkeeping**
Sex, identification number (and date applied), breed, date animal was acquired, address of previous owner, date sold, and address of new owner must be kept in farm records for 5 years (11).

**Tracing of Infected Animals**
Infected animals are traced back to their flock or origin through recordkeeping and scrapie tags (6).

**Clean-Up Strategies**
With the goal to keep producers in business and breeding stock alive, live animal testing, genetic testing, and individual destruction of infected flock members is the preferred method of scrapie infection cleanup (6).
Scrapie Prevention

Ultimately, prevention of scrapie within a producer's flock/herd should be used as a first line of defense in the eradication of scrapie as a whole.

Tips to Prevent Scrapie Within your Herd

• Keep a closed flock/heard
• If you must buy breeding stock from another heard, do so from one that is not known to have scrapie
• Keep does/ewes that are high risk for scrapie away from other does/ewes at kidding time to reduce exposure to bodily fluids
• Selectively breed stock with PrP resistant genotypes
• Cull animals with PrP susceptible genotypes (8).
Scrapie Prevention

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Testing for Scrapie Resistance:
- S146 gene
- K222 gene
- These genes cause significant resistance to scrapie occurring naturally as well as introduced in laboratory settings
- Detection of these genes allow for selective breeding for scrapie-resistant goats
- Testing currently available at the UC of Davis and soon to be available at NeoGen Genomics
- Herd cleanup projects based upon genetic testing for these alleles could be considered in the future
- Discounted testing prices are available for members of the American Boer Goat Association as well as the American Dairy Goat Association (12)

Ultimately, prevention of scrapie within a producer's flock/herd should be used as a first line of defense in the eradication of scrapie as a whole.
Success So Far

• No classical Scrapie or source flocks have been identified since 2016
• Only one positive goat has been found though RSSS since 2003, and only 41 total field cases since 2002. This does not include quarantined flocks in Colorado (13).

With the continued support of goat and sheep producers, the United States is well on its way to being declared scrapie free.


