



# Scrapie

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## Introduction

Scrapie is a transmissible spongiform encephalopathy (TSE) of sheep and goats. It is similar to other TSEs, such as bovine spongiform encephalopathy (BSE) or "mad cow disease" of cattle, chronic wasting disease (CWD) of deer and elk, and a number of diseases that affect humans. These diseases occur because of a buildup of an abnormal cellular protein in the brain, known as a prion. Susceptibility to these diseases is genetically controlled in some species (including sheep and goats) but in some other species a genetic component is less apparent. However, in all species affected, the central nervous system including the brain starts to degenerate; there is no known cure for TSEs, and they are always fatal.

## Causative Agent

Prions, short for 'proteinaceous infectious particles,' are believed to be responsible for causing scrapie. Prions are an abnormal form of a normal cellular protein. These proteins are in very high concentrations in lymph tissues (specialized cells involved with the immune system) and nervous tissues throughout the body. Once the abnormal form of the protein occurs, it causes the other normal proteins around it to change into the abnormal form; this is how it 'spreads' after an animal is infected. Prions are very resistant to destruction by cooking, rendering, normal heat sterilizing methods and many detergents. Therefore, once they are in the environment, it is very difficult to get rid of them. Although the prion itself is infectious, a sheep or goat must have genetic susceptibility to scrapie in addition to becoming infected with the prion in order to develop the disease.

## Disease Transmission

Susceptible sheep are typically infected as young lambs through contact or ingestion of infected placenta or birth fluids from infected ewes (which may not necessarily be mother-to-offspring transmission), but susceptible adult sheep also can become infected through this route. The disease also can be transmitted by lambs ingesting colostrum or milk from infected ewes. Scrapie is not passed from the ewe to the lamb in the uterus, but it can be transmitted via blood transfusion of scrapie-infected blood to another susceptible sheep. It is possible that transmission could occur through infected urine, feces, saliva or nasal secretions. Although rams are susceptible to the disease, they are not known to transmit the infection to other animals through the act of breeding. However, ram genetics will contribute to the scrapie susceptibility of their offspring. The incubation period (time from infection to developing signs of the disease) is generally two years to

five years, and scrapie always results in death of the animal. Although the disease is much more prevalent in black-faced sheep (Suffolks, Hampshires and crosses of these), all breeds are susceptible.

## Clinical Signs

Clinical signs (symptoms) include progressive weight loss with a normal appetite, behavioral changes (including suddenly becoming aggressive), incoordination, head tremors, intense pruritus (itchiness), biting at or pulling wool out of their sides or legs, cachexia (muscle wasting), weakness, recumbency (going down and being unable to get up), blindness and death. Affected animals may have a high-stepping gait in the front limbs or a 'bunny-hopping' gait in the hindlimbs. The name "scrapie" was coined because infected animals will often scrape their wool/hair off from all the itching and rubbing. Once these signs develop, the animal will usually have one month to three months to live.

## Disease Diagnosis

Diagnosis of scrapie in live animals is from lymph tissue biopsy of the third eyelid, certain lymph nodes or rectal mucosa. Postmortem diagnosis (diagnosis after death) is from brain tissue. These tests determine actual infection and are generally only used on scrapie suspect and scrapie exposed animals. However, only testing of the brain is guaranteed to correctly identify an animal infected with scrapie. Testing a live animal for scrapie can result in false negatives (the animal may have scrapie but still have a negative test result). Animals do not develop an immune response to scrapie, so a blood test for antibodies to scrapie cannot be used.

## Treatment & Prevention

Unfortunately, there is no treatment for scrapie. Scrapie is no different than any other livestock disease. If proper biosecurity measures are followed, producers can limit the risk their flock will be infected. There are several precautions producers can take to protect the flock from scrapie, many of which will help limit the exposure to other diseases as well.

1. **Limit outside exposure.** Limit contact of nonfarm personnel with livestock and livestock areas, wash hands and boots when visiting other livestock facilities or wear plastic boots, and clean and disinfect livestock trailers after hauling livestock from other farms. Fairs, sales and exhibitions require special attention due to the number of animals commingling from across the country. Ewes

and does that are in close proximity need to have special confinement to prevent contact with vaginal fluids.

2. **Lambing/kidding management.** Remove placentas and bedding soiled by birth fluids from birthing areas right away and thoroughly clean the birthing area between lambings/kiddings.
3. **Flock/herd additions. Close the flock to female additions. The best way to maintain a healthy flock is to maintain a closed flock.** Once the genetics of the flock has been established, replacement females should be selected from within the flock. Since scrapie is transmitted primarily by females during lambing, keeping the disease out of the females is essential. If new additions are to be made, purchase ewes that have been genotyped and have the QR or RR genotype, or buy sheep from flocks certified by the National Scrapie Flock Certification Program.
4. **Use genotyping to select genetically resistant breeding stock.** Disease vulnerability of offspring can be well addressed by using RR rams alone without having to focus on the breeding females.

There is no definite way to prevent scrapie on a farm except to maintain a totally closed flock/herd and completely prevent any contact with other sheep or goats.

## Scrapie Genetics & Genotyping

Genotyping is a DNA test that identifies the genes for susceptibility or resistance. This tool measures vulnerability, not infection. Blood is the sample most generally used by approved laboratories. However, the U.S. Department of Agriculture (USDA) will only recognize tests when the blood sample was collected by an accredited veterinarian. There are multiple genes that influence susceptibility, Codon 171 is the gene most generally focused on in the U.S. At this gene, QQ animals are highly susceptible to scrapie infection IF EXPOSED; QR animals are rarely susceptible; and RR animals are resistant. In essence, a 171 QQ fetus exposed to a scrapie infected placenta will most likely result in scrapie infection of the newborn. Using this information, sheep producers can select breeding stock that will produce resistant offspring. For example, if an RR ram is bred to a QQ ewe, a QR lamb will be produced every breeding. This is the reason RR rams are desirable, because they can be bred to susceptible ewes and still produce offspring that is "rarely susceptible." So, essentially, disease vulnerability can be well addressed by ram selection alone without having to focus on the breeding females. There are many traits that must be evaluated when determining breeding potential. Genotype is another to be added to that list. Genotyping can be performed by any accredited veterinarian.

## Atypical Scrapie

Another form of scrapie, termed 'atypical scrapie' or the 'Nor98 variant' of scrapie is very uncommon, but has been reported in the U.S., as well as other countries. It may occur

sporadically and is believed by some researchers to be a random conversion of the normal form of the prion protein to the abnormal form. Genetic susceptibility to this form of scrapie is almost the exact opposite of the genetic susceptibility to 'classical' scrapie (described above). Animals that would be resistant to the classic form would be susceptible to atypical scrapie.

## National Scrapie Eradication Program

In 2001, the USDA announced an accelerated program to eradicate scrapie in the nation's sheep and goat herds. This program is succeeding, but to meet the goal of substantially eradicating the disease, the cooperation of sheep and goat producers everywhere is needed. The National Scrapie Eradication Program (NSEP) coordinated by the USDA is a joint effort that includes participation by state governments, industry and particularly producers. Veterinarians and producers can contribute significantly to this program by:

- Officially identifying sheep and goats per federal and state regulations. In Oklahoma, sheep and goats of any age must be officially identified before being moved for sale, exhibition, slaughter or change of ownership. If unsure whether a particular sheep or goat needs to be identified, tag it. Official ear tags and applicator pliers are available free of charge from USDA Veterinary Services.
- Reporting suspect animals to a state (405-522-8396), federal (405-751-1701) or accredited veterinarian.
- Use genotyping to select scrapie resistant animals. Join the USDA Scrapie Flock Certification Program.

Other excellent sources of information regarding scrapie and the NSEP include:

- <http://www.eradicatescrapie.org/>
- [http://www.sheepusa.org/ResearchEducation\\_Online\\_Education\\_Scrapie](http://www.sheepusa.org/ResearchEducation_Online_Education_Scrapie)

More information can be found at the USDA web site.



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