NVAP Reference Guide - Scrapie (Control and Eradication)

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There are two types of scrapie; classical and nonclassical. Nonclassical scrapie is also referred to as atypical, Nor98, or Nor98-like scrapie. Nonclassical scrapie appears to occur sporadically and has occurred in sheep of all the common genotypes and goats. It is either not transmissible or poorly transmissible under natural conditions. Given this the scrapie eradication program focuses on classical scrapie. Here after where “scrapie” is used it is intended to mean “classical scrapie”.

Scrapie is a fatal, degenerative TSE disease affecting the central nervous system of sheep and goats. First recognized as a disease of sheep in Great Britain and other countries of Western Europe more than 250 years ago, scrapie has been reported throughout the world. In the United States, scrapie has primarily been reported in the black-face meat breeds and their crosses. It also has been diagnosed in numerous other breeds and crossbreeds including wool and hair sheep, and in goats. At the end of FY 2016, the percent of cull sheep found positive at slaughter and adjusted for face color was 0.001 percent with an upper confidence limit of 0.009 percent. This measure of prevalence has decreased by 99 percent since slaughter surveillance started in FY 2003. Based on all goats sampled at slaughter through FY 2016, the prevalence of scrapie in U.S. cull goats is 0.002 percent with an upper 95 percent confidence limit of 0.004 percent.

The agent responsible for scrapie and other TSEs is smaller than the smallest known virus and has not been completely characterized. There are a variety of theories regarding the nature of the agent. The most widely accepted is that disease is caused by an infectious protein, or prion, that causes the normal cellular version of the protein to change shape such that it can no longer be degraded by the cell, causing the protein to accumulate and damage the cell.

The scrapie agent is extremely resistant to heat and to normal sterilization processes. It does not evoke any detectable immune response or inflammatory reaction in host animals. The scrapie agent is thought to be spread most commonly from the ewe to her offspring and to other lambs in contemporary lambing groups through contact with the placenta and placental fluids and through milk and colostrum. Signs or effects of the disease usually appear 2 to 5 years after the animal is infected but may take longer to appear. Sheep usually live 1 to 6 months after the onset of clinical signs and in some cases longer, but death is inevitable.

On the farm, veterinarians identify scrapie suspects based on the appearance of its signs combined with knowledge of the animal’s history and signalment. Signs of scrapie vary widely among individual animals and develop very slowly. As the result of nerve cell damage, affected animals usually show behavioral changes, tremor (especially of the head and neck), pruritus, and locomotor incoordination, which progresses to recumbency and death. Early signs include subtle changes in behavior or temperament. These changes may be followed by scratching and rubbing against fixed objects, apparently to relieve itching. Other signs are loss of coordination, weight loss despite retention of appetite, biting of feet and limbs, lip smacking, and gait abnormalities, including high-stepping of the forelegs, hopping like a rabbit, and swaying of the back end.

An infected animal may appear normal if left undisturbed at rest. However, when stimulated by a sudden noise, excessive movement, or the stress of handling, the animal may tremble or fall down in a convulsive like state. Several other problems can cause clinical signs similar to scrapie in sheep, including the diseases ovine progressive pneumonia, listeriosis, and rabies; the presence of external parasites (lice and mites); pregnancy toxemia; and toxins.

Testing

The official test currently used for scrapie diagnosis in the United States is immunohistochemistry.
Histopathology, Western Blot, and ELISA may be used as supplemental tests or when tissues are not suitable for immunohistochemistry. Histopathology is used to identify pathological changes in the brain following necropsy. Pathological changes of scrapie are confined to the central nervous system. The lesions are characteristically found in the grey matter of the brainstem. They include neuronal vacuolation, other forms of neuronal degeneration, astrocytosis, and a vacuolar or spongy alteration called status spongiosis. Immunohistochemistry, Western Blot, and ELISA can be used to detect the abnormal prion protein in brain or lymphoid tissues.

Third-eyelid and rectal lymphoid tissue biopsy are APHIS-approved tests for scrapie detection in live animals. The tests use a biopsy of lymphoid tissue from the third eyelid or rectum and IHC. Rectal biopsies are preferred over third-eyelid biopsies because they typically have more follicles so fewer samples are read out as invalid test due to insufficient follicles. Most collectors also find it easier to collect rectal biopsies compared to third eyelid biopsies. A single rectal biopsy or two third eyelid biopsies done at the same time have a sensitivity of approximately 87 percent when compared to the result of IHC testing on lymph node and brain. When animals over 14 months are tested, held and necropsied upon death the sensitivity of the third eyelid is reduced to about 70 percent as would be expected due to the long incubation period of the disease. It is likely that the rectal biopsy, if evaluated long term, would have similar results.

Identifying Affected Animals

Animals that are incubating the disease and may be shedding the agent are rarely identified until the onset of clinical signs. The only absolute way to prevent an introduction of scrapie into a flock is to prohibit all movements of sheep and goats into a flock. Until a more sensitive, cost-effective live-animal test is available, the risk can be substantially reduced by maintaining a closed ewe flock; by acquiring female animals only from certified free flocks, zones, or countries; and/or by acquiring ewes that are genetically resistant or less susceptible.

If scrapie develops in a flock, the risk of further spread, reintroduction of the disease, or both can be minimized through

- Removal of genetically susceptible exposed sheep and exposed goats;

- If all susceptible exposed animals are not removed live-animal testing and removal of test-positive animals;
- Breeding for genetic resistance;
- Careful cleaning and disinfection of lambing facilities;
- Improved management of animals at lambing time with particular attention to segregating them into small groups or keeping them alone when possible, maintaining the risk classification of animals in each group at the same level, and removing and burying, landfilling or incinerating placenta and soiled bedding immediately following lambing; and
- When warranted by extremely valuable seedstock, employing embryo transfer, cesarean section, or both.

The National Scrapie Eradication Program

The National Scrapie Eradication Program has two major components: a regulatory eradication program called the Accelerated Scrapie Eradication Program (ASEP) and a voluntary certification program called the Scrapie Flock Certification Program (SFCP).

Accelerated Scrapie Eradication Program

- In September 2001, the scrapie regulations were revised to require the official identification of sheep and goats not in slaughter channels (except low-risk commercial goats) and any sheep over 18 months of age in interstate commerce with some exceptions. In addition, the revision required States to implement and enforce official identification of most sheep and goats on change of ownership intrastate in order to move sheep and goats interstate with minimal restrictions.
- APHIS provides free official sheep and goat eartags to producers and accredited veterinarians. Call 866-USDA-TAG to request tags;
- Infected sheep are identified through active slaughter surveillance, reporting of suspect animals by producers and accredited veterinarians, testing of mature sheep or goats that die on farm or at other locations, and live-animal testing of higher risk animals;
- Effective tracing of infected animals to their flock or herd of origin and tracing and testing of exposed animals made possible as a result of the new identification requirements; and

- Providing effective cleanup strategies that will allow producers to stay in business, preserve breeding stock, and remain economically viable.

APHIS provides the following assistance to owners of exposed and infected flocks or herds that participate in cleanup plans including owners of exposed animals that have been sold out of infected and source flocks or herds:

- Indemnity for high-risk, suspect, and scrapie-positive sheep and exposed goats that owners agree to destroy;
- Genetic testing of sheep for scrapie susceptibility; and
- Scrapie testing on live or dead animals

Scrapie Flock Certification Program (SFCP)

The SFCP is a voluntary program that is open to all sheep and goat producers in the United States. The overall objective of the SFCP is to minimize the scrapie risk of participating flocks and herds, thereby improving the marketability of animals from participating flocks and herds and contributing to the national scrapie eradication program.

The SFCP has two categories: Export and Select. The Export category has two statuses (Export Monitored and Export Certified), and the Select category has one status (Select Monitored).

Export Category. The objective of this category is to certify participating flocks and herds as scrapie free establishments through limiting the acquisition of does and ewes from flocks of the same or higher status, annual inspections including reconciliation of the animal inventory, official individual animal identification (ID) requirements, recordkeeping requirements, and animal sampling requirements. Animals from Export Certified flocks are considered highly unlikely to be infected with scrapie, and have greater marketing opportunities both domestically and internationally.

Select Category. The objective of this category is enhance scrapie surveillance by increasing the reporting of animals with clinical signs of scrapie and submission of samples for scrapie testing. Select flocks are required to submit samples from at least one mature animal for scrapie testing every 1-3 years, depending on the size of the flock or herd. Participating flocks do not advance to a certified status.

All requirements of the SFCP are outlined in the SFCP program standards, available in electronic form at: Scrapie Free Flock Certification Program

Role of Accredited Veterinarians in Scrapie Eradication Accredited veterinarians play an integral role in the eradication of scrapie. The accreditation standards require reporting of live or dead scrapie suspects to State and Federal authorities. In addition to this critical role, accredited veterinarians are the producers' primary source of education about all aspects of the program, including identification, recordkeeping, and movement requirements. When requested by the producer, accredited veterinarians can apply official eartags, collect and submit samples for official genotype testing, scrapie testing on obex, lymph node, third-eyelid lymphoid tissue, or rectal lymphoid tissue; and issue Certificates of Veterinary Inspection for interstate movement. Additionally, accredited veterinarians play a very important role in educating producers about the disease and in assisting producers with the prevention and elimination of scrapie.

Information on the issuance of Certificates of Veterinary Inspection for sheep and goats and on identification of sheep and goats can be found in the "Sheep and Goat" section under the "Animal Movement" heading. Additional information on the APHIS National Scrapie Eradication Program may be found at: National Scrapie Eradication Program

Guidance on the scrapie regulations can be found in the Scrapie Eradication Uniform Methods and Rules at : Scrapie Eradication Uniform Methods and Rules (271.72 KB)

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