

Scrapie

Scrapie is a fatal, degenerative disease affecting the central nervous system of sheep and goats. It is among a number of diseases classified as transmissible spongiform encephalopathies (TSE). Infected flocks that contain a high percentage of susceptible animals can experience significant production losses. Over a period of several years the number of infected animals increases, and the age at onset of clinical signs decreases making these flocks economically unviable. Female animals sold from infected flocks spread scrapie to other flocks. The presence of scrapie in the United States also prevents the export of breeding stock, semen, and embryos to many other countries. TSEs are the subject of increased attention and concern because of the discovery of bovine spongiform encephalopathy (BSE) in cattle, the link between BSE and variant Creutzfeldt-Jakob disease (vCJD) in people, and feline spongiform encephalopathy (FSE) in cats in Europe. This increased concern has led to the following:

- Packers and producers have had difficulty finding options for disposal of sheep offal and dead sheep causing packers and producers to incur significant increases in disposal costs,
- Other countries have expressed concerns and have indicated that they may prohibit or restrict certain ruminant products because the United States has scrapie, and
- Domestic and international markets for U.S. sheep-derived meat and bone meal have been adversely affected.

The combination of all of these factors has led to the decision to develop a strong scrapie eradication program in the United States.

Epidemiology and Transmission

The agent responsible for scrapie and other TSEs is smaller than the smallest known virus and has not been completely characterized. There are three main theories on the nature of the scrapie agent: (1) the agent is a prion, which is an abnormal form of a normal cellular protein, 2) the agent is a virus with unusual characteristics, and (3) the agent is a virino, a very small piece of DNA that acts like a virus. 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 sheep and goats.

The scrapie agent is thought to be spread most commonly from the ewe to her offspring and to other lambs through contact with the placenta and placental fluids. Signs or effects of the disease usually appear 2 to 5 years after the animal is infected but may not appear until much later. Sheep may live 1 to 6 months or longer after the onset of clinical signs, but death is inevitable. The genetics of the sheep affects their susceptibility to scrapie.

In the laboratory, the scrapie agent has been transmitted to hamsters, mice, rats, voles, gerbils, mink, cattle, and some species of monkeys by inoculation. There is no scientific evidence to indicate that scrapie poses a risk to human health. There is no epidemiologic evidence that scrapie of sheep and goats is transmitted to humans, such as through contact on the farm, at slaughter plants, or butcher shops.

Clinical Signs

Signs of scrapie vary widely among individual animals and develop very slowly. Due to damage to nerve cells, affected animals usually show behavioral changes, tremor (especially of head and neck), rubbing, and locomotor incoordination that 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, weakness, 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.

On the farm, veterinarians diagnose scrapie based on the appearance of its signs combined with knowledge of the animal's history. Scrapie can be diagnosed in the live animal by biopsy of the lymphoid tissues on the inside of the third eyelid. This test is used by the U.S. Department of Agriculture's (USDA) Animal and Plant Health Inspection Service

(APHIS) to determine whether exposed flocks are infected. Scrapie is most often diagnosed by microscopic examinations of brain tissue at necropsy or by procedures that detect the presence of the abnormal prion protein in brain tissue.

Research

Scrapie research efforts are currently focused on developing more practical live-animal tests to diagnose infected sheep before they show signs, investigating transmissibility of the agent, identifying the scrapie agent and its different strains, identifying genes that influence scrapie infection and evaluating genetic selection as a tool for scrapie eradication. Substantial evidence has accrued to show that the risk of scrapie transmission by embryo's is negligible provided that the embryos are properly handled between collection and transfer, but additional experimental data are needed to support existing evidence.

Related Diseases

The TSE family of diseases includes BSE: transmissible mink encephalopathy; FSE; chronic wasting disease of deer and elk; kuru; both classical and variant Creutzfeldt-Jakob disease; Gerstmann-Straussler-Scheinker syndrome; and fatal familial insomnia. TSEs have also been reported in Europe in captive wild ruminants in the bovid family, cats, and monkeys. The occurrence of TSEs in captive wild animals is believed to have resulted from BSE-contaminated feed.

Eradication Program

USDA has initiated an accelerated scrapie eradication program. The program is based on the following key concepts:

- Identification of preclinical infected sheep through live animal testing and active slaughter surveillance,
- Effective tracing of infected animals to their flock/herd of origin made possible as a result of the identification requirements, and
- Providing effective genetic based flock cleanup strategies that will allow producers to stay in business, preserve breeding stock, and remain economically viable. APHIS provide the following to exposed and infected flocks/herds that participate in cleanup or monitoring plans:
 1. Indemnity for high-risk, suspect, and scrapie positive sheep and goats, which owners agree to destroy,
 2. Scrapie live-animal testing,
 3. Genetic testing, and
 4. Testing of exposed animals that have been sold out of infected and source flocks/herds.

Operating an effective program to deal with this insidious disease requires cooperation among producer organizations, allied industries, and governmental agencies.

History

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. Only two countries are recognized by the United States as being free of scrapie: Australia and New Zealand.

The first case of scrapie in the United States was diagnosed in 1947 in a Michigan flock. The flock owner had imported sheep of British origin through Canada for several years. APHIS conducted a slaughter surveillance study from April 1, 2002, to March 31, 2003, which determined the prevalence of scrapie in mature U.S. cull sheep to be 0.2 percent or one positive out of 500 cull sheep.

In the United States, scrapie has primarily been reported in the Suffolk breed. It also has been diagnosed in a Border Leicester, Cheviots, Corriedales, a Cotswold, Dorsets, Finn sheep, Hampshires, Merinos, Montadales, Rambouillets, Shropshires, Southdowns, and a number of crossbreeds. Through October 2003, approximately 2,350 cases in sheep and 12 cases in goats have been reported.

Additional Information

For more information about scrapie, contact your local APHIS, Veterinary Services, area office or contact:

USDA, APHIS, Veterinary Services
National Animal Health Programs
4700 River Road, Unit 43
Riverdale, MD 20737-1231
Telephone (301) 734-6954
Fax (301) 734-7964

Current information on animal diseases and suspected outbreaks is also available on the Internet. Point your Web browser to <http://www.aphis.usda.gov/vs/nahps/scrapie/> to reach the APHIS scrapie home page.

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