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ERADICATE SCRAPIE!

Scrapie Fact Sheet

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Scrapie is a fatal, degenerative disease affecting the central nervous system of sheep and goats. Infected flocks that contain a high percentage of susceptible animals can experience significant production losses. In these flocks 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. Animals sold from infected flocks spread scrapie to other flocks. The presence of scrapie in the U.S. also prevents the export of breeding stock, semen, and embryos to many other countries.

More recently, increased attention and concern has been paid to all transmissible spongiform encephalopathy (TSE) diseases, including scrapie, as a result of the discovery of bovine spongiform encephalopathy (BSE) in cattle, and the link between BSE and new variant Creutzfeldt-Jakob disease (*nv*CJD) in people and feline spongiform encephalopathy (FSE) in cats in Europe. This increased concern has led to the following effects:

- Packers and producers have had difficulty in disposing of sheep offal and dead sheep causing them 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 U.S. has scrapie, and
- Our domestic and international markets for 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 full-fledged scrapie eradication program in the U.S.

History

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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. From this first case through July 2001, scrapie has been diagnosed in more than 1000 flocks in this country.

In the United States, scrapie has primarily been reported in the Suffolk breed. It also has been diagnosed in Border Leicester, Cheviot, Corriedale, Cotswold, Dorset, Finnsheep, Hampshire, Merino, Montadale, Rambouillet, Shropshire, Southdown, and a number of crossbreeds. Through August 2001, approximately 1,600 cases in sheep and seven (7) cases in goats have been reported.

Epidemiology and Transmission

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Scrapie is classified as a transmissible spongiform encephalopathy (TSE). 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 virus with unusual characteristics, (2) the agent is a prion, an exclusively host-coded protein that is modified to a protease-resistant form after infection, and (3) the agent is a virino, a small, noncoding regulatory nucleic acid coated with a host-derived protective protein. 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. Signs or effects of the disease usually appear two (2) to five (5) years after the animal is infected but may be longer. Sheep may live one (1) to six (6) months or longer after the onset of clinical signs, but death is inevitable.

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, or through the consumption of sheep or goat meat/milk products.

Clinical Signs

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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), pruritus, 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, 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. There is a test currently under-going evaluation by USDA's Animal and Plant Health Inspection Service (APHIS) for scrapie detection in live animals. Scrapie can currently only be confirmed by microscopic examinations of brain tissue at necropsy or by procedures that detect the presence of the abnormal prion protein.

Genetics

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Susceptibility to scrapie in sheep is affected by the amino acid sequence of the sheep's prion protein. It appears that codons 136, 154 and 171 of the prion protein play the largest role in regards to natural scrapie in sheep. The clinical and pathological differences observed following exposure appears to be controlled by both the scrapie strain and the host PrP genotype. With very few exceptions, naturally infected sheep of a number of breeds in the US, UK, Europe and Japan carry either 136 Valine (136 Valine/Valine or 136 Valine/Alanine) or 171 Glutamine/Glutamine (QQ) (Belt at al 1995, Clouscard et al 1995, Hunter et al 1993, Hunter et al 1994, Ikeda et al 1995, Laplanche et al 1993a & 1993b, Westaway et al 1994, O'Rourke et al 1996). There has been only one report of a scrapie-affected Suffolk carrying 171 Arginine/Arginine (RR) (Ikeda et al 1995) and four published reports of scrapie-affected Suffolks with 171 Glutamine/Arginine (QR) (Junghans et al., 1998; Hunter et al., 1997; Ikeda et al., 1995).

This suggests that genetic selection may be helpful in controlling and possibly eliminating clinical disease within flocks.

Research

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Scrapie research efforts are currently focused on developing a practical live animal test 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 control, and examining the role of artificial insemination and embryo transfer in the transmissibility of the scrapie agent. Research studies using experimentally infected sheep suggest that embryos may play a role in the spread of scrapie.

Related Diseases

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The TSE family of diseases includes bovine spongiform encephalopathy (BSE), which affects cattle; transmissible mink encephalopathy; feline spongiform encephalopathy; chronic wasting disease of deer and elk; and kuru, both classical and variant Creutzfeldt-Jakob disease, Gerstmann-Straussler-Scheinker syndrome, and fatal familial insomnia, five rare diseases in humans. 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.

Control Program

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The United States Department of Agriculture (USDA) has initiated an accelerated scrapie eradication program. The program is based on the following key concepts:

- Identification of pre-clinical 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 new identification requirements.
- Providing effective cleanup strategies that will allow producers to stay in business, preserve breeding stock, and remain economically viable. USDA/APHIS will do this by providing the following to exposed and infected flocks/herds that participate in cleanup 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.

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