

SHEEP HEALTH

Fact Sheet . . . No. 9



SCRAPIE OF SHEEP

Scrapie is a transmissible degenerative disease of the nervous system of sheep characterized by a long incubation period and a protracted, debilitating degeneration of the central nervous system. Changes produced in the brain are microscopic. There is no inflammatory response and no immunity produced by the disease.

Related Diseases

Several diseases of other species have features similar to scrapie in sheep:

1. Kuru is a disease of humans restricted to the native Fore tribe of Papua New Guinea. Incidence of the disease has declined dramatically with the demise of ritualistic cannibalism in this tribe.
2. Creutzfeldt-Jakob disease also affects humans but is widespread throughout the world. It is characterized by pre-senile dementia. There is no known exposure risk, such as scrapie of sheep, as the disease exists in parts of the world where scrapie does not occur.
3. Transmissible mink encephalopathy is a rare disease of ranch-reared mink. Evidence suggests that it may be caused by feeding infected livestock carcasses to mink.
4. Chronic wasting disease of captive mule deer and elk may be a natural disease caused by exposure, either prior to or after capture.
5. Bovine spongiform encephalopathy (BSE) is a disease of dairy cattle in Great Britain and other countries that is believed to have been caused by feeding contaminated protein concentrate prepared from scrapie-infected sheep carcasses to cattle. It has been transmitted experimentally to laboratory mice and appears to be similar to scrapie.

Incidence

Scrapie is widespread throughout the world. The disease was eradicated from Australia and New Zealand after outbreaks in 1952 and 1954. In the United States the disease was first diagnosed in Michigan in 1947. In the United States, scrapie has been found in Cheviots, Corriedales, Dorsets, Finns, Hampshires, Merinos, Montadales, Southdowns, Suffolks, and several crossbreeds.

Pathogenesis

The scrapie agent most likely enters the body via the oral route. During the first year or two of life, the infectious agent remains in the lymph tissue at low levels. After two to three years, it affects the central nervous system, where it multiplies more rapidly. The means of shedding the organism from sheep is not known, although it may be excreted in feces, nasal secretions, and fetal membranes.

Transmission

The organism that causes scrapie is one of the most unusual of all disease producing organisms known. It is closely associated with cellular fractions of the host (sheep) tissue. It does not possess the same characteristics as viral agents. It can be thought of as a piece of protein material that transmits the disease. It is resistant to environmental conditions, many disinfectants, and is substantially resistant to cooking or rendering. In addition to the infectious organisms, a "prion" gene has been detected that is an important factor in controlling both host susceptibility to scrapie and the length of the incubation period. This gene apparently reacts closely with the transmissible agent, resulting in clinical disease. Based on current knowl-

edge, scrapie can be regarded as an infectious disease that is affected by genetic susceptibility.

The scrapie organism has not been detected in a wide variety of tissues and body fluids. It has not been found in embryos, fetuses, kidneys or urine, salivary glands or saliva, testes or semen, mammary tissues or milk from infected animals. It has not been found in urine or feces from infected animals or in pasture grass, drinking water, or bedding used by infected animals. This information should, however, be evaluated with caution at the present time.

Scrapie can be spread from animal to animal in infected flocks, including those managed under pasture conditions. The most common method of transmission of scrapie is from dams to their own and other offspring. The age of greatest susceptibility to natural infection appears to be during the first few months following birth. Transmission from sire to offspring is minimal, probably because sires and offspring rarely mingle under most management conditions. Goats are highly susceptible.

The extent to which scrapie is transmitted by contaminated premises such as corrals, barns, feed, water, animal excretions, and bedding is not known. Infection of sheep by exposure to such premises has been reported, although exposure may have been due to contact with infected sheep. There is no evidence that scrapie is transmitted by birds, small mammals, or external parasites. There is also no evidence that scrapie is transmitted to the lamb prior to birth.

Susceptibility

The period of greatest susceptibility is the first few months of life. There also is greater likelihood of intense exposure at this time, due to the intimate contact of lambs with infected dams.

Susceptibility to scrapie is strongly influenced by the genetic make-up of animals. A gene that controls the length of the incubation period has been identified. Alleles for both short and long incubation periods have been identified. Characterization of the genetic

alleles is expected to provide a method to predict the susceptibility of sheep to scrapie.

Sheep with the "short" incubation allele appear to develop scrapie about 2-4 years after exposure. Sheep with the "long" allele usually fail to develop the disease, because they die from natural causes before the incubation period is completed. It is not known to what extent or under what conditions sheep with the long incubation allele might be a source of infection for other sheep.

Clinical signs

Due to damage to nerve cells, scrapie infected sheep will usually show behavioral changes, tremors, and incoordination that progresses to recumbency and death. A high-stepping gait of the forelimbs and "bunny hop" movement of the rear legs is characteristic. Animals react abnormally to stimulation, noise, movement, and handling. Infected sheep lose weight while maintaining a normal appetite. Itching may occur, but is often not dramatic. Some animals will bite at their legs and pull wool from their sides. Animals will live from one to six months after the onset of symptoms.

Bovine spongiform encephalopathy

A neurologic disorder called bovine spongiform encephalopathy (BSE) has been diagnosed in cattle in Great Britain. British veterinary scientists believe the disease resulted from contamination of protein supplement with scrapie-infected sheep offal. Primarily dairy cattle have been involved in the episode. A large number of herds but few individuals within most of these herds have been involved. BSE has not been identified in cattle in the United States, although an active surveillance program is in operation. The incidence of BSE in the U.K. is now decreasing.

Diagnosis

Control of scrapie is greatly complicated by the lack of a simple diagnostic test. No immunity to the disease is developed, so traditional serological tests do not work. Currently, tested livestock tissues may

provide the method for flock surveillance. This is an intense area of research.

Treatment

There is no treatment for scrapie, or for any of the other spongiform encephalopathies of animals and humans.

Prevention

The major method of prevention of scrapie is to maintain a closed flock. Replacement of ewes or rams should originate from "scrapie-free" flocks. In reality, this is difficult to do since no pre-purchase test is currently available to ensure freedom from the disease. Buyers must rely on the integrity of the seller; but sellers may not always be aware that their flock is infected because of the long incubation period of the disease. Buyers should ask sellers to sign a statement indicating to the best of the seller's knowledge that no affected or exposed animals have been in the flock. Because contagion is more likely from ewes than rams, raising replacement ewe lambs and purchasing only rams reduces the risk of acquiring scrapie. No procedures other than maintaining a totally closed flock and avoiding all contact with other sheep can guarantee freedom from the disease, however.

Public Health Concerns

There is no evidence that scrapie or BSE is transmissible to humans.

Proposed federal program

Instituting an effective scrapie control program will be difficult in the absence of a diagnostic test on live animals. One alternative would be the slaughter of all infected and exposed flocks. This would be very costly. Substantial federal funding for control and research has been sought, but is uncertain at this time. A scrapie-free certification plan has been proposed.

State programs

Iowa quarantines infected and bloodline flocks, but does not have a state indemnification program.

Summary and conclusions

Scrapie is a serious disease that has caused economic loss to several producers in the state. Many producers fear contracting the disease. Disease control efforts are hampered by incomplete knowledge of the infectious agent, epidemiology of the disease, and especially by the lack of a diagnostic test. Federal control efforts are hampered by the absence of decisive, clearly defined objectives, and by lack of adequate funding.

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