

Nor98-Like (Nonclassical) Scrapie

Nor98-like scrapie is a prion disease. Other prion diseases include classical scrapie in sheep and goats, bovine spongiform encephalopathy (BSE) primarily of cattle, chronic wasting disease (CWD) of deer and elk, and in humans Creutzfeldt-Jakob disease (CJD) and variant Creutzfeldt-Jakob disease (vCJD). What each of these diseases has in common is that they cause a progressive and ultimately fatal degeneration of the nervous system. While the underlying cause(s) are still debated, the theory most widely accepted in the scientific community is that the infectious agent is a prion—an abnormal form of a normally occurring cellular protein.

Unlike BSE, classical scrapie and Nor98-like scrapie have not been shown to be a threat to human health. Classical scrapie has been known to exist for

over 250 years, and cases have occurred in the United States since 1947. It is likely that Nor98-like scrapie has also existed for a long time; however, it was first identified in Norway in 1998, hence the name Nor98. This type of scrapie is referred to as “atypical scrapie,” “Nor98 scrapie,” “Nor98-like scrapie,” or “nonclassical scrapie.” Since 1998, similar cases have been found in almost every country in Europe, as well as Australia, Canada, the Falkland Islands, New Zealand, and the United States. The first cases in the United States were identified in 2007.

Nor98-like (nonclassical) scrapie and classical scrapie are separate diseases with distinct features. Laboratory findings quickly distinguish Nor98-like scrapie from classical scrapie. In 2009, the World Animal Health Organization (OIE) recognized Nor98-like scrapie as a separate disease from classical scrapie because of differences in laboratory findings, transmissibility, and distribution. This determination means that Nor98-like scrapie is not a reportable disease to OIE, and should be of no trade concern.

	Nor98-like (nonclassical) scrapie	Classical scrapie
Transmission	Nor98-like scrapie is either not transmitted or is poorly transmitted, under natural conditions, meaning thorough exposure to infected animals on farms. Many scientists believe that Nor98-like scrapie is instead caused by a random conversion (often referred to as “sporadic”) of the normal prion protein into the abnormal form.	Classical scrapie is an infectious disease that is transmitted to other sheep and goats under natural conditions.
Distribution	Nor98-like scrapie has been found in all countries where extensive surveillance has been conducted using sensitive test methods. Nor98-like scrapie cases tend to be isolated in many different herds and appear to be randomly distributed.	Classical scrapie has not been reported in some of the countries where Nor98-like scrapie is found. Classical scrapie cases often occur in clusters.
Number of affected sheep or goats in a flock or herd	Nor98-like scrapie is rarely found in more than one animal in a flock or herd. When an additional case is found, it is usually in flocks with more than 500 sheep.	Classical scrapie usually infects two or more sheep or goats in an infected flock.

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Average age when clinical signs start to appear in animals	In Nor98-like scrapie, clinical signs are rarely documented and the animals are typically diagnosed when sampled at slaughter, usually at greater than 5 years of age.*	With classical scrapie, clinical signs typically first appear and result in death in animals that are between 3-5 years of age.
Clinical signs associated with disease	Clinical signs are rarely reported in Nor98-like scrapie cases. Furthermore, intense rubbing against fixed objects has not been reported in Nor98-like scrapie cases.	Intense rubbing against fixed objects is a frequently occurring clinical sign in classical scrapie cases.
Variations in protection against the disease conferred by genotype	In Nor98-like scrapie, no protection is conferred by genotype.	Some sheep have genotypes that are resistant to classical scrapie but are still susceptible to Nor98-like scrapie.

* In the few Nor98-like scrapie cases where clinical signs were reported, the signs observed were indistinguishable from those described for classical scrapie. These include incoordination, gait abnormalities, collapsing while running, tremors, loss of condition, leg biting, behavioral changes and/or nibble response. The nibble response is when a scrapie-affected animal is scratched somewhere on its body (usually along its back) and it causes involuntary lip and/or tongue movements.

APHIS Policy Regarding Nor98-Like Scrapie

APHIS will no longer require the depopulation or movement restriction of Nor98-like scrapie-exposed sheep and goats. APHIS is planning to propose changes to the Code of Federal Regulations (CFR) in 2010 to allow the APHIS Administrator to eliminate or reduce post-exposure requirements for certain or specific scrapie types—such as Nor98-like scrapie—that pose minimal risk of transmission between animals under natural conditions.

In the meantime, APHIS is conducting a national scrapie-control pilot project for Nor98-like scrapie. The project involves flocks and herds in which animals positive for Nor98-like scrapie were born, lambled, or kidded. The pilot project will allow producers that have sheep and goats exposed to Nor98-like (nonclassical) scrapie to retain, sell, exhibit, or move them for any purpose.

Response Measures

If a Nor98-like scrapie case is found in or traced back to an owner's flock or herd, certain measures will be taken. The owner will be contacted by a Federal or State veterinarian who will schedule a visit to the farm accompanied by the owner to:

- Confirm the identification of the positive animal;
- Provide the owner with information about scrapie and its control; and

- Determine if the positive animal was born in or gave birth in the flock/herd. If so, the veterinarian will:
 - Work with State animal health authorities to ensure animals are not moved from the premises until they have been officially identified;
 - Develop a Nor98-like scrapie flock plan and a 5-year monitoring plan with the producer;
 - If not already officially identified, apply official eartags to sheep and goats exposed to Nor98-like scrapie; and
 - Inventory all sheep and goats, and any sheep or goat embryos.

The officially identified sheep and goats will then be classified and handled as low-risk exposed animals, allowing the owner to move the animals from the premises for any purpose including sale.

Additional Information

To learn more about classical scrapie, Nor98-like scrapie, and any updates to the scrapie control pilot project, visit http://www.aphis.usda.gov/animal_health/animal_diseases/scrapie/.

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