

## What is Nor98-Like (Nonclassical) Scrapie?

Nor98-like scrapie is a prion disease. The 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 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” in the literature.

Since 1998, almost every country in Europe, as well as the Falkland Islands, New Zealand, Canada, and the United States have found similar cases. The first cases in the United States were identified in 2007.

Nor98-like (nonclassical) scrapie and classical scrapie are separate diseases with distinct features.

- Transmission
  - Classical scrapie is an infectious disease that is transmitted to other sheep and goats under natural conditions.
  - Nor98-like scrapie is either not transmitted or is poorly transmitted, under natural conditions. Many scientists believe that Nor98-like scrapie is not an infectious disease under natural conditions and that it is instead caused by a random conversion of the normal prion protein into the abnormal form (often referred to as “sporadic”).
- Distribution
  - Nor98-like scrapie has been found in all countries where extensive surveillance has been conducted using sensitive test methods; whereas, classical scrapie has not been reported in some of these countries.
  - Nor98-like scrapie cases are widely distributed and proportionate with sheep/goat populations; whereas, classical scrapie cases often occur in clusters.
- Number of affected sheep or goats in a flock or herd
  - Classical scrapie usually infects more than one animal in an infected flock. In the US, approximately 18 percent of the mature, genetically susceptible sheep in an infected flock are infected.
  - 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.
- Average age of onset of clinical signs in animals
  - With classical scrapie, clinical signs typically first appear and result in death in animals that are between 3-5 years of age.

- In Nor98-like scrapie, clinical signs are rarely documented and the animals are typically diagnosed when they are sampled at slaughter, usually at greater than 5 years of age.
- Variations in protection against disease conferred by genotype
  - Sheep with genotypes that are resistant to classical scrapie are susceptible to Nor98-like scrapie.
- Clinical signs associated with disease
  - Unlike classical scrapie, clinical signs are rarely reported in Nor98-like scrapie cases.
  - 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, collapse while running, tremors, loss of condition, leg biting, nibble response and/or behavioral changes.
  - One potential clinical difference is that intense rubbing is a frequently occurring clinical sign in classical scrapie cases; whereas, it has not been reported in Nor98-like scrapie cases.
- Laboratory findings readily 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.

#### **How has APHIS' policy changed regarding Nor98-like scrapie?**

APHIS will no longer require the depopulation or movement restriction of Nor98-like scrapie exposed sheep and goats. APHIS will 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 scrapie types (or certain/specific types of scrapie)-- such as Nor98-like scrapie -- that are determined to pose minimal risk of lateral transmission under natural conditions.

In the meantime, APHIS is conducting a national scrapie control pilot project for Nor98-like scrapie. The project is for flocks and herds in which animals positive to Nor98-like scrapie were born, lambed, or kidded.

#### **What will the pilot project do for producers?**

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.

#### **What will happen if Nor98-like scrapie case is found on or traced back to an owner's flock or herd?**

The owner will be contacted by a Federal or State veterinarian who will schedule a visit to the farm with the owner. The activities listed below will be performed.

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

- 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 authority 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.