USDA United States Department of Agriculture

Animal and Plant Health Inspection Service

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Factsheet

Bovine Spongiform Encephalopathy

Bovine spongiform encephalopathy (BSE) is a progressive neurological disease of cattle caused by prions, which are infectious agents made up of protein material. The prion proteins affect the brain structure of infected animals, causing the animal to lose motor skills and eventually causing death. Clinical signs include behavioral changes, coordination problems, weight loss and decreased milk production. Once clinical signs appear, the animal's condition deteriorates until it either dies or is euthanized. There is no treatment or vaccine to prevent BSE.

There are two general types of BSE - classical and atypical. Neither form is contagious.

Classical BSE

Classical BSE spreads through the ingestion of certain materials (brain, spine, etc.) from infected animals. The main way classical BSE spread was through contaminated animal feed containing meat or bone meal from infected cattle. Because of BSE, the Food and Drug Administration banned the use of ruminant protein in feed for ruminants in 1997 and additionally prohibited the use of certain high-risk tissues of cattle in feed for all animals in 2009. The feed ban is proven to keep animals from becoming infected with classical BSE.

The incubation period for classical BSE from the time of infection until the onset of clinical signs averages three to six years.

The first diagnosis of classical BSE occurred in 1986 in the United Kingdom, where most of the worldwide cases occurred. Since then, many other countries have confirmed cases. The first case of BSE in the United States was classical BSE. The United States Department of Agriculture (USDA) confirmed the case in 2003, was an animal imported from Canada born before the establishment of feed bans.

Atypical BSE

Atypical BSE occurs rarely and spontaneously, typically in cattle that are eight years old or older. There is no association of atypical BSE with contaminated feed or ingestion of infected materials. Instead, a spontaneous change in the prion proteins in older adult cattle cause atypical BSE. Because changes in prion proteins are sporadic and very rare, scientists do not fully understand what causes prions to change from normal to abnormal forms.

Scientists are currently studying whether genetics or environmental factors may contribute to atypical BSE. As part of this research, scientists typically examine the offspring of animals diagnosed with atypical BSE and animals born in the same location at the same time as affected animals.

Four of the five cases of BSE identified in the United States were diagnosed as atypical BSE. In all four cases, the infected animals were at least 10 years old.

BSE and Trade

The World Organization for Animal Health (OIE) officially recognizes a country's risk status for BSE after an evaluation of the country's history with the disease, the implementation and enforcement of their feed ban, and their BSE surveillance. The OIE guidelines outline criteria for safe trade in cattle and their products according to their risk status. The United States' import requirements follow these guidelines, and USDA encourages other countries to do the same regarding imports from the U.S.

After the 2003 BSE finding, many countries stopped importing U.S. beef. This caused substantial losses for the cattle industry. In 2013, the OIE upgraded the U.S. BSE-status to negligible risk, the best status available. In 2015, the OIE determined that atypical BSE occurred spontaneously at a low rate in all cattle populations and would be excluded when determining BSE risk. Today, most countries that prohibited U.S. beef due to BSE have removed those restrictions.

BSE Surveillance

USDA began conducting BSE surveillance testing in 1990 as a way to detect any cases that might be present in the United States. If no cases are found, surveillance results also prove the absence of the disease in the country.

The current U.S. surveillance program, in place since 2006, far exceeds the current surveillance guidelines provided by the OIE.

The testing program focuses on cattle populations that are at higher risk for BSE, including animals 12 months of age and older that display central nervous system signs; and animals over 30 months of age that are excluded from slaughter due to poor health status (non-ambulatory, unhealthy, or dead).

Producers can support USDA's efforts to identify BSE, especially cases of atypical BSE that fall outside of normal surveillance efforts:

- Immediately report any unusual or suspicious signs of disease to your veterinarian, State or Federal animal disease control officials, or to your county agricultural agent;
- Watch cows that show nervous or aggressive behavior, difficulty with coordination, trouble standing up, decreased milk production, and weight loss; and
- Report the location of any potential offspring for affected cows.

Related Diseases
BSE is part of a group of diseases known as transmissible spongiform encephalopathies (TSEs). Some TSE diseases include scrapie in sheep and goats, chronic wasting disease in deer, elk and moose, and classic and variant Creutzfeldt-Jakob disease (CJD) in people.

BSE is considered a public health concern because cases of variant CJD in people are linked to the consumption of food containing ingredients derived from classical BSE-infected cattle.

Additional Information
For more information about BSE, contact:

USDA, APHIS, Veterinary Services
Surveillance Preparedness and Response Staff
4700 River Road, Unit 41
Riverdale, MD 20737–1231
Phone: (301) 734–8073
Fax: (301) 734–7817

Current information on animal diseases and disease outbreaks is also available on the Internet at
https://www.aphis.usda.gov/aphis/ourfocus/animalhealth/animal-disease-information.