Bovine Spongiform Encephalopathy

Bovine spongiform encephalopathy (BSE) is a progressive neurological disease of cattle. It is 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 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 (FDA) banned the use of ruminant protein in feed for ruminants in 1997. FDA additionally prohibited the use of certain high-risk cattle tissues 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 3 to 6 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 classical BSE in the United States was confirmed by the U.S. Department of Agriculture (USDA) in 2003. The animal was imported from Canada and born before feed bans were established.

Atypical BSE

Atypical BSE occurs rarely and spontaneously, typically in cattle that are 8 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.

Six of the seven cases of BSE identified in the United States have been diagnosed as atypical BSE. In most cases, the animals were 10 years of age or older. Two of the six detections of atypical BSE involved animals aged approximately 5 years or older.

BSE and Trade

The World Organisation for Animal Health (WOAH) officially recognizes a country’s BSE risk status after evaluating the country’s history with the disease, the implementation and enforcement of their feed ban, and their BSE surveillance. WOAH 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 U.S. imports.

After the 2003 BSE finding, many countries stopped importing U.S. beef. This caused substantial losses for the cattle industry. In 2013, WOAH upgraded the U.S. BSE-status to negligible risk, the best status available. In 2015, WOAH 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 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 WOAH. 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
- 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, by:

- Immediately reporting any unusual or suspicious signs of disease to your veterinarian, State or Federal animal disease control officials, or to your county agricultural agent;
- Watching cows that show nervous or aggressive behavior, difficulty with coordination, trouble standing up, decreased milk production, and weight loss; and
- Reporting 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). Other 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.

More Information

For more information about BSE, email USDA’s Ruminant Health Center at VS.SP.Cattle.Health.Center@usda.gov. Current information on BSE detections in the United States is available online at www.aphis.usda.gov/animalhealth.