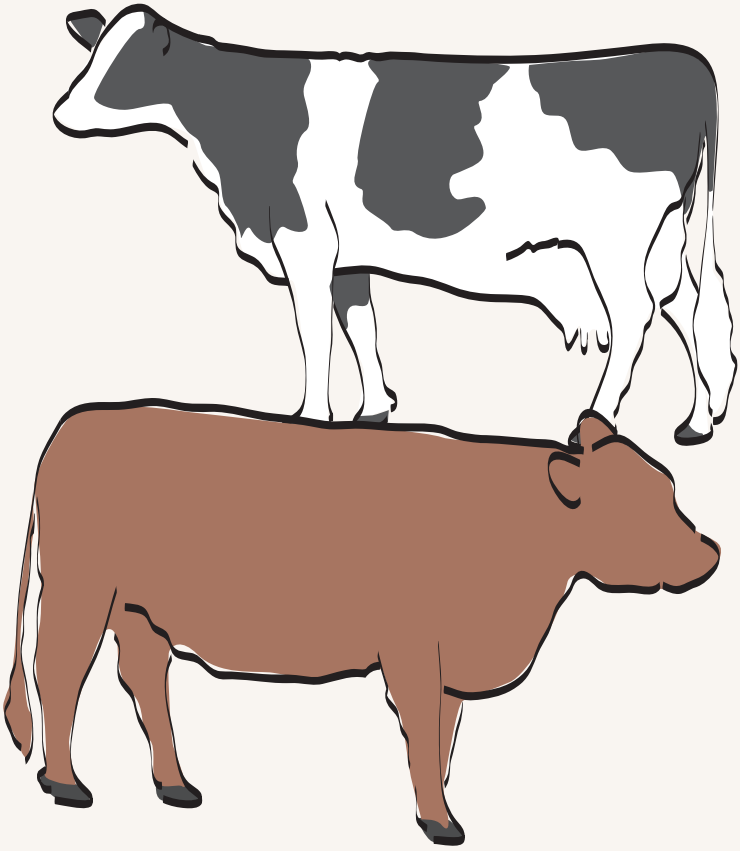
United States Department of Agriculture
Animal and Plant Health Inspection Service

Program Aid No. 1705

Bovine Spongiform Encephalopathy:

An Overview



Introduction

Bovine spongiform encephalopathy (BSE), widely referred to as "mad cow disease," is a chronic degenerative disease affecting the central nervous system of cattle. The disease was first diagnosed in 1986 in the United Kingdom. Since that time, it has been found in many European countries, and also in countries outside of Europe, including Japan, Canada, and the United States. To date, however, more than 95 percent of the total cases worldwide have occurred in the United Kingdom.

What Causes BSE and How It Progresses

BSE is a progressive and fatal neurological disease of cattle caused by an unconventional transmissible agent. BSE belongs to the family of diseases known as transmissible spongiform encephalopathies (TSEs). In addition to BSE, TSEs include scrapie, which affects sheep and goats; transmissible mink encephalopathy; chronic wasting disease of deer and elk; and in humans, kuru, both classic and variant Creutzfeldt–Jakob disease (CJD), Gerstmann–Straussle–Scheinker syndrome, and fatal familial insomnia.

The agents that cause BSE and other TSEs have yet to be fully characterized. The theory most widely accepted in the scientific community is that the agent is a prion–an abnormal protein. The BSE agent is extremely resistant to heat and to normal sterilization processes. It also does not evoke any detectable immune response or inflammatory reaction in host animals.

Cattle affected by BSE experience progressive degeneration of the nervous system. Affected animals may display nervousness or aggression, abnormal posture, difficulty in coordination and rising, decreased milk production, or loss of body weight despite continued appetite. All infected cattle die. There is neither any treatment nor a vaccine to prevent the disease.

Figure 1—APHIS supports the FDA regulation (effective August 4, 1997) prohibiting the use of most mammalian protein in the manufacture of animal feeds given to ruminants. In addition, the final regulation also requires process and control systems to ensure that ruminant feed does not contain the prohibited mammalian tissue.

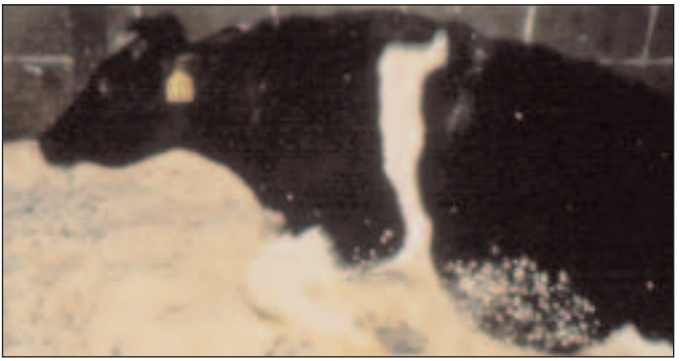


Figure 2 and 3—Cattle affected by BSE experience progressive degeneration of the nervous system. Changes in temperament (e.g., nervousness or aggression), abnormal posture, incoordination and difficulty in rising, decreased milk production, and/or loss of weight despite continued appetite are followed by death.

The incubation period (the time from when an animal becomes infected until it first shows disease signs) averages 4 to 6 years, although the period can be longer or shorter. Following the onset of clinical signs, the animal's condition deteriorates until it either dies or is destroyed. The process of deterioration usually takes from 2 weeks to 6 months.

Currently, there is no test to detect the disease in live cattle; veterinary pathologists confirm BSE by postmortem microscopic examination of brain tissue or by the detection of abnormal prions in brain tissue. BSE is so named because of the spongy appearance of the brain tissue of infected cattle when examined under a microscope.

Transmission

BSE is not a contagious disease and therefore is not spread through casual contact between cattle. The primary source of BSE infection in cattle is commercial feed contaminated with the infectious agent. Scientific evidence shows that feed contamination results from incorporating ingredients (for example, meat-and-bone meal) that contain protein derived from rendered infected cattle. Standard rendering processes do not completely inactivate or kill the BSE agent. Therefore, rendered protein such as meat-and-bone meal derived from infected animals may contain the infectious agent. Regulations prohibiting the inclusion of mammalian or ruminant protein in ruminant feed, including cattle feed, are used to prevent BSE transmission.

Consumption of feed contaminated with the BSE agent is the only documented route of field transmission of BSE. However, limited research cannot rule out the possibility of maternal, or vertical, transmission. Although such transmission may be possible, scientific evidence suggests that it is unlikely to occur at any appreciable level, if at all.

Studies have indicated that most infected cattle were likely exposed to the BSE agent as calves and became infected during the first year of life. These findings suggest that susceptibility in cattle declines with age; therefore,

young animals are most susceptible. Studies also indicate that the incubation period is inversely related to dose. For example, an animal exposed to a low dose of the BSE agent would have a longer incubation period. Similarly, a very young animal (at the most susceptible age) exposed to a large dose would have a much shorter incubation period.

Tissue Distribution and Infectivity

Research conducted to date indicates that only certain tissues in animals affected with BSE are actually infective. Most of this information has been derived from experimental studies conducted in the United Kingdom. In these studies, different tissues derived from infected cattle were inoculated into mice to determine if disease transmission occurred. The same process has been repeated with inoculation into cattle. In cattle naturally infected with BSE, infectivity has been found only in brain tissue, the spinal cord, and the retina of the eye. In experimentally infected cattle, the distal ileum of the small intestine, tonsil, dorsal root ganglion, and trigeminal ganglion also were found to be infective.

Creutzfeld–Jakob Disease

In 1996, scientists in the United Kingdom announced the finding of 10 cases of a new variant of Creutzfeld–Jakob disease (vCJD), a chronic and fatal neurodegenerative disease of humans. These new cases presented a different clinical and pathological picture from routinely diagnosed cases of classic (sporadic) CJD. Experimental and epidemiologic studies have linked the occurrence of vCJD to exposure to the BSE agent, most likely through the consumption of food containing ingredients derived from infected cattle.

Figure 4—APHIS leads an ongoing, comprehensive interagency surveillance program for BSE in the United States to ensure the health of America's cattle herd.

USDA Actions in Response to BSE

The U.S. Department of Agriculture's (USDA) Animal and Plant Health Inspection Service (APHIS), in cooperation with the Food and Drug Administration (FDA) and USDA's Food Safety and Inspection Service (FSIS), has taken aggressive measures to prevent the introduction and potential spread of BSE in the United States.

APHIS has maintained stringent restrictions since 1989 to prevent importation of the highest risk animals and products. The primary animal-health protective measure is maintained by the FDA. In 1997, the FDA implemented regulations that prohibit the feeding of most mammalian proteins to ruminants, including cattle. This feed ban is the most important measure to prevent the transmission of disease to cattle.

Public or human health protective measures are maintained by both the Food Safety and Inspection Service and the FDA. The most important public-health protective measure is the removal from the human food supply of specified risk materials–those

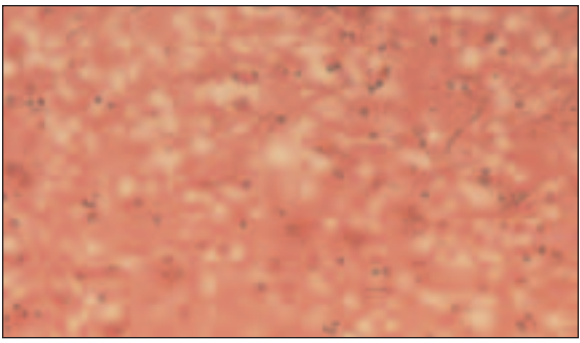


Figure 5—Vacuoles—microscopic holes in the grey matter—give the brain of BSE-affected cows a spongelike appearance when tissue sections are examined in the lab.

tissues where the BSE infective agent would be found if present. Other controls include banning nonambulatory disabled cattle from the human food chain; prohibiting air-injection stunning of slaughter cattle; requiring additional process controls in advanced meat-recovery systems; and forbidding the use of mechanically separated meat in human food. Additionally, protection from BSE and other disease is achieved through antemortem inspection of slaughter cattle and the exclusion of animals with any clinical signs of neurological disease or other abnormalities.

Surveillance and Monitoring

APHIS has conducted BSE surveillance since 1990, including an enhanced surveillance effort that was initiated after an imported cow tested positive for the disease in December 2003. The goal of the enhanced effort, which began in June 2004, was to test as many animals in a targeted population as possible over a 24-month period. This intensive effort was intended to provide sufficient data to allow USDA to more accurately estimate the prevalence or level of BSE within the U.S. cattle population.

Analysis of the data collected over 7 years of BSE surveillance shows that the prevalence of BSE

in the United States is less than 1 infected animal per million adult cattle. Two mathematical models were used in this analysis, and they estimated that the most likely number of infected animals present in the adult cattle population could be 4 or 7 infected animals out of 42 million adult cattle.

APHIS continues to conduct an ongoing BSE surveillance program that samples approximately 40,000 animals annually. This level of surveillance will continue to monitor the BSE status of U.S. cattle and will provide a mechanism for detection of BSE prevalence if it were to increase above 1 infected animal per million adults. In addition, this level of surveillance exceeds the guidelines set forth by the World Animal Health Organization.

Education, Training, Outreach

APHIS educates veterinary practitioners, veterinary laboratory diagnosticians, industry, and producers about the clinical signs and pathology of BSE. Videotapes of cattle showing clinical signs of BSE and BSE factsheets, risk assessments, and reviews have been widely distributed to State and Federal veterinarians, private practitioners, other industries, and producers. APHIS is continuing an education effort to inform U.S. cattle producers and veterinarians about this disease. Numerous briefings have been held for industry groups. In addition to press releases and factsheets, a videotape on BSE and an information packet were distributed to all APHIS field offices, State veterinarians, extension veterinarians, colleges of veterinary medicine, and industry groups.

Contacts for More Information About BSE

For general information about BSE, contact USDA, APHIS, Veterinary Services at (301) 734–6954.

For information about importing animals or animal products, contact USDA, APHIS, Veterinary Services, National Center for Import/Export Animals Program at (301) 734–8170 or the National Import/Export Products Program at (301) 734–7885.

For questions related to food safety, meat and meat products, or meat inspection, contact the USDA's Food Safety and Inspection Service at (202) 720–9113.

For questions related to human health or Creutzfeldtñ-Jakob disease, contact the Centers for Disease Control and Prevention at (404) 639–7292.

For questions related to BSE research, contact the USDA's Agricultural Research Service at (301) 504–1638.

For questions related to food, animal feed, drugs, cosmetics, or biological products, contact the FDA at (301) 443–1130.

Current information on animal diseases and suspected outbreaks is also available on the Internet at <http://www.aphis.usda.gov>. Specific information on BSE can be found at <http://www.aphis.usda.gov/newsroom/hot_issues>.

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