USDA

Key Facts

November 2001

Bovine Spongiform Encephalopathy (BSE)

What is BSE?

Bovine spongiform encephalopathy (BSE), commonly known as “mad cow disease,” is a chronic, degenerative neurological disorder of cattle. Worldwide, there have been more than 180,000 cases of BSE since the disease was first diagnosed in 1986 in the United Kingdom (UK). The disease has also been found in native-born cattle in Belgium, the Czech Republic, Denmark, France, Germany, Greece, Ireland, Italy, Japan, Liechtenstein, Luxembourg, the Netherlands, Portugal, Slovakia, Slovenia, Spain, and Switzerland. However, more than 95% of all BSE cases have occurred in the United Kingdom. No confirmed cases of BSE have been found in the United States despite more than 10 years of active surveillance.

BSE belongs to a family of diseases known as the transmissible spongiform encephalopathies (TSEs). TSEs share some common characteristics including a prolonged incubation period ranging from a few months to years and progressively debilitatingneurological illnesses, which are always fatal. Examples of other TSEs include scrapie (sheep and goats), chronic wasting disease (deer and elk), feline spongiform encephalopathy (cats), kuru (humans), and Creutzfeldt-Jakob Disease (CJD) (humans).

TSEs are caused by a transmissible agent that is not yet fully understood. The agent responsible for BSE is smaller than the smallest known virus and has not been completely characterized. There are three main theories on the nature of the agent: (1) the agent is a virus with unusual characteristics; (2) the agent is a prion – an exclusively host-coded protein that is modified to a partially protease-resistant form after infection; and (3) the agent is a virino – a small, noncoding regulatory nucleic acid coated with a host-derived protective 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.

Epidemiological data suggest that the outbreak of BSE was caused by a common source material, involving animal feed containing contaminated meat-and-bone meal as a protein source. The disease in Great Britain may have been caused by feeding cattle rendered protein produced either from the carcasses of scrapie-infected sheep or from cattle with a previously unidentified TSE. Changes in rendering operations in the early 1980s may have permitted an infectious TSE agent to survive the rendering process. The resulting contaminated meat-and-bone meal could then have been fed to cattle, eventually resulting in the epidemic.

Currently, there is no test to detect TSEs in a live animal. Veterinary pathologists confirm BSE by postmortem microscopic examination of brain tissue or by the detection of the abnormal form of the prion protein using supplemental tests described below. BSE is so named because of the spongy appearance of the brain tissue of affected cattle when sections are examined under a microscope.

Supplemental tests are available to enhance the diagnostic capabilities for TSEs. Research shows a partially protease-resistant form of the prion protein is found in the brain of TSE-infected animals. Two tests that have been used routinely to detect the abnormal prion protein in animals showing clinical signs of a TSE are immunohistochemistry and a Western-blot technique. These tests permit diagnosis of a TSE based on finding prion proteins even if the brain has been frozen or if tissue breakdown has occurred.

The presence of the TSE agent in tissues is generally determined by injecting animals, usually mice, with material believed to be infected with the TSE, then observing the mice to see if they die and have

characteristic lesions in the brain. Mouse inoculation studies take a long time (up to 700 days) to detect the agent. Since this method may take more than 2 years, it is not considered practical for routine testing.

To date there has been no evidence of infectivity detected in milk or muscle tissue.

BSE: Implications for Cattle

Cattle affected with BSE experience progressive degeneration of the nervous system. Affected animals may display changes in temperament, such as nervousness or aggression, abnormal posture, lack of coordination and difficulty in rising, decreased milk production, and loss of body weight despite a normal appetite. The disease is always fatal. Currently, there is neither a treatment for the disease nor a vaccine to prevent it.

The incubation period typically (the time from when an animal becomes exposed until it first shows signs of disease) is from 2 to 8 years. Following the onset of clinical signs, the animal's condition deteriorates until it either dies or is destroyed. This usually takes from 2 weeks to 6 months. Most cases in the United Kingdom have occurred in dairy cows between 3 and 6 years of age.

There is no evidence that BSE spreads by contact between unrelated adult cattle or from cattle to other animal species. However, some evidence suggests that maternal transmission may occur at an extremely low level. This low level would not be sufficient to sustain the epidemic in the UK if it occurs at all.

BSE: Implications for Humans

BSE is a disease that affects cattle. However, other TSEs affect humans. The most commonly identified TSE in humans is classic (sporadic) Creutzfeldt-Jakob Disease. Classic CJD occurs sporadically at a rate of approximately one case per one million people per year, worldwide.

Scientific evidence (epidemiological and laboratory) supports a causal relationship between BSE outbreaks in Europe and another TSE disease in humans, called new variant Creutzfeldt-Jakob disease (vCJD).

The disease vCJD is most likely caused by the ingestion of products contaminated with the BSE agent. As of November 2, 2001, 111 cases of vCJD have been suspected or confirmed in the UK. There have also been four cases in France and one case in Ireland.

Patients with vCJD have primarily been younger and exhibit clinical signs of the disease longer than patients with classic CJD. The median age of death of persons afflicted with vCJD is 28 years – compared to 68 years for CJD. The average time between the onset of clinical symptoms and death is 13 months for vCJD and less than 6 months for CJD.

For More Information

For questions concerning BSE, BSE surveillance, and agricultural import bans, contact APHIS
Media Inquiries: (301) 734-7799
Technical Inquiries: (609) 259-5825
APHIS Web site: www.aphis.usda.gov

For questions concerning food safety, AMR, or MSM, contact FSIS
Media Inquiries: (202) 720-9113
Technical Inquiries: (202) 690-6566
Consumer Inquiries: Call USDA's Meat and Poultry Hotline at 1-800-535-4555. In the Washington, DC, area, call (202) 720-3333. The TTY number is 1-800-256-7072.
FSIS Web site: www.fsis.usda.gov

For questions concerning CJD, vCJD, and other human TSEs, contact CDC
Media Inquiries: (404) 639-3286
CDC Web site: www.cdc.gov