

# Bovine Spongiform Encephalopathy

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 Great Britain.

BSE has had a substantial impact on the livestock industry in the United Kingdom. The disease also has been confirmed in native-born cattle in Austria, Belgium, Czech Republic, Denmark, Finland, France, Germany, Italy, Ireland, Japan, Liechtenstein, Luxembourg, the Netherlands, Northern Ireland, Portugal, Slovakia, Slovenia, Spain, and Switzerland. The U.S. Department of Agriculture's (USDA) Animal and Plant Health Inspection Service (APHIS) is enforcing import restrictions and is conducting surveillance for BSE to ensure that this serious disease does not become established in the United States.

## Clinical Signs

Cattle affected by BSE experience progressive degeneration of the nervous system. Affected animals may display changes in temperament, such as nervousness or aggression, abnormal posture, incoordination and difficulty in rising, decreased milk production, or loss of body weight despite continued appetite. Affected cattle die. The causative agent of the disease is not completely characterized, and there is neither any treatment nor a vaccine to prevent the disease.

The incubation period (the time from when an animal becomes infected until it first shows disease signs) 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 process usually takes from 2 weeks to 6 months. Most cases in Great Britain have occurred in dairy cows between 3 and 6 years of age.

Currently, there is no test to detect the disease 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. BSE is so named because of the spongy appearance of the brain tissue of infected cattle when sections are examined under a microscope.

## History

Since November 1986 (when BSE was first identified as a separate disease entity), over 178,000 head of cattle have been diagnosed with BSE in Great Britain. The epidemic peaked in January 1993 at approximately 1,000 new cases reported per week. Agricultural officials in Great Britain have taken a series of actions to eradicate BSE, including making BSE a notifiable disease, prohibiting the inclusion of mammalian meat-and-bone meal in feed for all food-producing animals, prohibiting the inclusion of animals more than 30 months of age in the animal and human food chains, and destroying all animals showing signs of BSE and other animals at high risk of developing the disease.

As a result of these actions, most notably the imposition of feed bans, the rate of newly reported cases of BSE has greatly decreased.

## Epidemiology

Epidemiologic data suggest BSE in Great Britain is an extended common source epidemic involving animal feed containing contaminated meat and bone meal as a protein meat source.

There are different scientific hypotheses concerning the origins of BSE. BSE in Great Britain may have been caused by feeding cattle rendered protein produced from the carcasses of scrapie-infected sheep or cattle with a previously unidentified TSE.

The practice of using products such as meat-and-bone meal as a source of protein in cattle rations has been common for several decades. Changes in rendering operations in the late 1970's and early 1980's may have played a part in the appearance of the disease.

There is no evidence that BSE spreads horizontally, i.e., by contact between unrelated adult cattle or contact between cattle and other species. Limited research suggests that maternal or vertical transmission may occur at a very low level. This low level most likely would not perpetuate the epidemic under British farming conditions. Research continues in this area.

BSE is classified as a transmissible spongiform encephalopathy (TSE). The agent responsible for BSE and other TSE's 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.

In cattle naturally infected with BSE, the BSE agent has been found only in brain tissue, in the spinal cord, and in the retina. The distal ileum, bone marrow, dorsal root ganglion, and trigeminal ganglion from experimentally infected cattle were also found to be infective. To date, there has been no evidence of infection detected in milk or muscle tissue.

The presence of the BSE agent in tissues is determined by inoculating animals, usually mice, with material believed to be infected with BSE. Mouse inoculation studies take a long time (up to 700 days) to detect the agent, and failure to identify it in tissues may indicate either true absence of the agent or simply the limited sensitivity of current diagnostic methods.

## Related Diseases

The TSE family of diseases includes scrapie, which affects sheep and goats; transmissible mink encephalopathy; feline spongiform encephalopathy; chronic wasting disease of deer and elk; and in humans, kuru, both classic and variant Creutzfeldt–Jakob disease, Gerstmann–Straussler–Scheinker syndrome, and fatal familial insomnia. TSE's have also been reported in captive exotic ruminants, and exotic and domestic cats. The agent isolated from several of these cases is indistinguishable from BSE in cattle suggesting the occurrence of TSE's in these species resulted from BSE-contaminated feed.

On March 20, 1996, the United Kingdom's Spongiform Encephalopathy Advisory Committee (SEAC) announced the identification of 10 cases of variant Creutzfeldt–Jakob disease. These 10 cases had a characteristic clinical and pathological phenotype which differed from other routinely diagnosed cases of classic (sporadic) CJD in that they are characterized by a younger age at onset of symptoms, have behavioral change symptoms, longer duration of illness, a non-diagnostic or normal EEG tracing, and, under microscopic examination, different lesions in brain tissues.

SEAC concluded that, although there was no direct scientific evidence of a link between BSE and vCJD, based on current data and in the absence of any credible alternative, the most likely explanation at that time was that the cases were linked to exposure to BSE before the introduction of a specified bovine offal (SBO) ban at slaughter in 1989. The SBO ban excluded from human consumption brain, spinal cord,

and other organs with potential BSE infectivity. As of Nov. 2, 2001, 114 cases of vCJD had been identified in the United Kingdom, one in Ireland, and five in France.

It is important to further clarify the difference between classic CJD and vCJD. Classic CJD occurs each year at a rate of 1 to 2 cases per 1 million people throughout the world, including in the United States and other countries where BSE has never occurred and among vegetarians and meat eaters alike. Classic CJD occurs sporadically (about 90 percent of cases), through iatrogenic transmissions (less than 1 percent) or genetically (about 10 percent).

Current evidence suggest that vCJD is a novel condition, clinically and pathologically. The epidemiological evidence is consistent with BSE and the causal agent and recent laboratory evidence provides strong support to the hypothesis of a causal link between BSE and vCJD.

## USDA Actions in Response to BSE

BSE has not been diagnosed in the United States, and USDA has worked proactively to keep it that way. In cooperation with USDA's Food Safety and Inspection Service (FSIS), APHIS has taken stringent measures in prevention, education, surveillance, and response.

To prevent BSE from entering the country, since 1989 APHIS has prohibited the importation of live ruminants from countries where BSE is known to exist in native cattle. Other products derived from ruminants, such as fetal bovine serum, bonemeal, meat-and-bone meal, bloodmeal, offal, fats, and glands, are also prohibited from entry, except under special conditions or under USDA permit for scientific or research purposes.

On December 12, 1997, APHIS extended these restrictions to include all of the countries in Europe due to concerns about widespread risk factors and inadequate surveillance for BSE.

As of December 7, 2000, USDA prohibited all imports of rendered animal protein products, regardless of species, from Europe. This decision followed the recent determination by the European Union that feed of nonruminant origin was potentially cross-contaminated with the BSE agent. The restriction applies to products originating, rendered, processed or otherwise associated with European products. USDA has taken emergency action to prevent potentially cross-contaminated products from entering the United States. The same type of rendered product from ruminant origin has been prohibited from BSE–infected countries since 1989.

APHIS educates veterinary practitioners, veterinary laboratory diagnosticians, industry, and producers on the clinical signs and pathology of BSE.

APHIS leads an ongoing, comprehensive, interagency surveillance program for BSE in the United States. BSE is a reportable disease by accredited veterinarians. APHIS veterinary pathologists and field investigators have received training, including training from their British counterparts in diagnosing BSE. The surveillance samples include field cases of cattle exhibiting signs of neurological disease, cattle condemned at slaughter for neurologic reasons, rabies-negative cattle submitted to public health laboratories, neurologic cases submitted to veterinary diagnostic laboratories and teaching hospitals, and sampling of cattle that are nonambulatory (downer cattle/fallen stock). APHIS has also begun to sample adult cattle that die on farms. APHIS' surveillance program is based on laboratories' histopathologically examining brains and using a technique called immunohistochemistry to test brain tissues for the presence of the abnormal prion protein. As of Feb. 4, 2002, more than 21,451 brains from the United States and Puerto Rico have been examined with no evidence of BSE or other TSE detected.

APHIS also monitors the remaining cattle imported from Great Britain, Belgium, and other European countries (before the bans on imports from those countries went into effect). As of Dec. 31, 2000, of the 496 cattle imported from Great Britain and Ireland between 1981 and 1989, three animals are still alive. The animals are quarantined and observed regularly. To date, no evidence of BSE or a TSE has been detected. There are five head of cattle from other European countries imported between 1986-97 that are still alive and under quarantine.

APHIS, in cooperation with the Food Safety and Inspection Service, has also drafted an emergency response plan to be used in the event that BSE is identified in United States. In addition, APHIS' TSE Working Group monitors and assesses all ongoing events and research findings regarding TSE's. APHIS continually revises and adjusts prevention and diagnostic measures as it receives new information and knowledge.

As an additional preventative measure, APHIS supports the Food and Drug Administration's (FDA) regulation (effective August 4, 1997) prohibiting the use of most mammalian protein (with certain exceptions) 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 tissues.

### Getting the Word Out

As part of its increased surveillance activities, 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.

For additional information, contact  
USDA, APHIS, Veterinary Services  
Emergency Programs  
4700 Riverdale Road, Unit 41  
Riverdale, MD 20737-1231  
Telephone: (609) 259-5825 or (301) 734-8073

For information about importing animals or animal products, contact

USDA, APHIS, Veterinary Services  
National Center for Import/Export  
Animals Program  
Telephone: (301) 734-8170  
Products Program  
Telephone: (301) 734-7885

For public health information, contact CDC at (404) 639-3091. For food safety information, contact FSIS at (202) 205-0293 or call the USDA's Meat and Poultry Hotline at (800) 535-4555. For more information about the ruminant feed ban, call FDA's Consumer Hotline at (800) 532-4440.

Current information on animal diseases and suspected outbreaks is also available on the Internet. Point your Web browser to <http://www.aphis.usda.gov> to reach the APHIS home page. For specific information on BSE, point your browser to <http://www.aphis.usda.gov/oa/bse>.

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