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 Great Britain.

BSE has substantially damaged the livestock industry in the United Kingdom. The disease has also been confirmed in native-born cattle in Belgium, the Czech Republic, Denmark, France, Germany, Greece, Italy, Ireland, Liechtenstein, Luxembourg, the Netherlands, Northern Ireland, Portugal, Spain, and Switzerland.

BSE has not been diagnosed in the United States, and the U.S. Department of Agriculture’s (USDA) Animal and Plant Health Inspection Service (APHIS) is enforcing import restrictions and conducting surveillance for BSE to minimize the risk of this disease becoming established here. In addition, the Food and Drug Administration (FDA) has prohibited the use of most mammalian protein in ruminant feed.

Clinical Signs

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. Affected cattle will die. 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 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.

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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.

APHIS photographer Dr. Art Davis, a veterinarian at the National Veterinary Services Laboratories (NVSL) in Ames, IA.

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.

History

Between November 1986 and July 2001, more than 178,000 head of cattle in over 35,000 herds were 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 eliminate BSE, including making it a reportable 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 potentially exposed 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 decreased and continues a downward trend.

USDA Actions in Response to BSE

Implementation of Stringent Measures

BSE has not been diagnosed in the United States. The USDA policy has been to be proactive and preventive. In cooperation with USDA’s Food Safety and Inspection Service (FSIS), APHIS has implemented stringent measures in prevention, education, surveillance, and response.

Entry Ban—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 owing to concerns about widespread risk factors 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 rendered products of nonruminant origin were potentially cross-contaminated with the BSE agent. The restriction applies to products originating, rendered, or processed in Europe or otherwise associated with European-rendered animal protein products. USDA has taken this 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.

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. Microscope slides showing typical BSE lesions have been distributed to Federal and State diagnostic laboratories, and Federal foreign animal disease (FAD) diagnosticians have been trained in Great Britain in BSE recognition. More than 250 Federal and State veterinarians throughout the United States have been trained to recognize FADs, including BSE.

APHIS veterinary pathologists and field investigators have received training that has included instruction from their British counterparts in diagnosing BSE. 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.

Surveillance and Monitoring—APHIS leads an ongoing, comprehensive, interagency surveillance program for BSE in the United States. Samples of BSE are obtained from five sources: field cases of cattle exhibiting signs of neurological disease, cattle condemned at slaughter for neurological reasons, rabies-negative cattle submitted to public health laboratories, neurological cases submitted to veterinary diagnostic laboratories and teaching hospitals, and aged cattle that are nonambulatory (downer cattle and fallen stock).

APHIS’ surveillance program is based on laboratories’ histopathologically examining brains and using a technique to test brain tissues for the presence of the abnormal prion protein. As of mid-August 2001, more than 14,600 bovine brains from the United States and Puerto Rico have been examined with no evidence of BSE or any other transmissible spongiform encephalopathy (TSE) detected.

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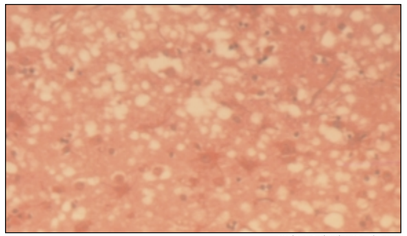
APHIS leads an ongoing, comprehensive interagency surveillance program for BSE in the United States to ensure the health of America’s cattle herd.

APHIS monitors the remaining cattle imported from Great Britain and other European countries before the bans on imports from those countries went into effect. As of July 1, 2001, of the 496 cattle imported from the United Kingdom and Ireland between 1981 and 1989, 3 animals are still alive. The animals are quarantined and observed regularly. To date, no evidence of BSE or a TSE has been detected. APHIS continues to attempt to purchase those three live animals for diagnostic research purposes. The six European cattle imported in 1996 and 1997 that are still alive are currently under quarantine, and APHIS is attempting to buy these animals as well.

All cattle presented for slaughter in the United States are inspected before slaughter by FSIS for signs of central nervous system disorders. Any animals exhibiting neurologic signs during this inspection are condemned, and the meat is not permitted for use as human food. The brains from these animals are submitted to APHIS’ National Veterinary Services Laboratories for analysis.



NVSL veterinarian Dr. Al Jenny

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.

Cooperation With Other Agencies

In cooperation with FSIS, APHIS has also drafted an emergency response plan to be used if BSE is identified in the United States. The plan specifies a step-by-step series of actions to be taken if BSE is detected here. In addition, APHIS' TSE Working Group monitors and assesses all ongoing events and research findings regarding TSEs. APHIS continually revises and adjusts prevention and diagnostic measures as it receives new information and knowledge.

APHIS supports the FDA's 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 tissue, thus preventing the possibility of the transmission of BSE to cattle.

Origins of the Disease

On the basis of epidemiological data, researchers believe that the source of BSE in Great Britain was feed containing meat-and-bone meal. 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 1970s and early 1980s may have played a part in the appearance of the disease.

There is no evidence that BSE spreads horizontally, that is, by contact between unrelated adult cattle or from cattle to 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 on this issue continues.

BSE is classified as a TSE. The agent responsible for BSE and other TSEs 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 abnormal form of a normal protein known as cellular prion protein, and (3) the agent is a virino—an "incomplete" virus composed of nucleic acid protected by host proteins. 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. In experimentally infected cattle, the distal ileum, bone marrow, dorsal root ganglion, and trigeminal ganglion also were found to be infective.

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

TSEs

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 (CJD), Gerstmann–Straussler–Scheinker syndrome, and fatal familial insomnia. TSEs have also been reported in captive exotic ruminants. The strain of agent isolated from the exotic ruminants and cats is indistinguishable from BSE in cattle, suggesting that the occurrence of TSEs 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 CJD. The disease process in 10 patients had a characteristic clinical and pathological phenotype differing from other routinely diagnosed cases of classic (sporadic) CJD. The 10 individuals experienced the onset of symptoms at a younger age, exhibited behavioral changes, were sick for longer than patients with classic CJD, displayed a nondiagnostic or normal electroencephalogram, and experienced brain lesions that were, under microscopic examination, different from lesions seen in brain tissue from patients with classic CJD.

SEAC concluded that, although there has been no direct scientific evidence of a link between BSE and variant CJD, on the basis of current data and in the absence of any credible alternative, the most likely explanation is that the cases resulted from exposure to BSE before the introduction of a specified bovine offal (SBO) ban at slaughter in 1989. The SBO ban excluded brain, spinal cord, and other organs with potential BSE infectivity from human consumption. As of August 6, 2001, 106 cases of probable or confirmed variant CJD had been identified in the United Kingdom, 1 in Ireland, and 3 in France.

Difference Between Variant CJD and Classic CJD

It is important to clarify the difference between classic CJD and variant CJD further. 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), iatrogenically (less than 1 percent), or genetically (about 10 percent). According to the U.S. Centers for Disease Control and Prevention (CDC), no cases of variant CJD have been identified in the United States.

Current evidence suggests that variant CJD is a clinically and pathologically new condition. The epidemiologic evidence is consistent with BSE, and the causal agent and recent laboratory evidence provide strong support for the hypothesis of a causal link between BSE and variant CJD.

Contacts for More Information About BSE

For general information about BSE, contact USDA, APHIS, Veterinary Services, Emergency Programs at (301) 734–8073.

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 FSIS at (202) 720–9113.

For questions related to human health or Creutzfeldt–Jakob disease, contact CDC at (404) 639–7292.

For questions related to science or BSE research, contact the National Institutes of Health at (301) 496–5751.

For questions related to food, 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/oa/bse.

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An Overview