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## **Bovine Spongiform Encephalopathy (BSE)**

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*Present situation in the United States:* The U.S. Department of Agriculture (USDA) made a preliminary diagnosis of BSE in a single nonambulatory, disabled (“downer”) Holstein cow in Washington State on December 23, 2003. The diagnosis was confirmed on December 25, 2003 by an international reference laboratory in Weybridge, England. The BSE-positive cow was 6.5 years old and was slaughtered on December 9, 2003. Although beef from the slaughter cow had been processed for human consumption, all central nervous system tissue was diverted out of the human food supply and into rendering. On December 23, 2003, the Food Safety and Inspection Service (FSIS) issued a recall on all beef slaughtered at the involved slaughter plant on December 9, 2003. USDA conducted a trace back investigation, indicating that the affected cow was likely imported from Canada in 2001. Prior to this incidence, the United States sampled over 57,000 brains since September 30, 2003 through an active BSE surveillance program and no evidence of BSE was detected. This incidence resulted in the modification of several regulations designed to eliminate the possibility of human exposure to materials that may contain the BSE agent, as well as the implementation of new policies to further minimize this risk.

*What is BSE?* Bovine Spongiform Encephalopathy (BSE) is a chronic degenerative disease that is classified in the family of diseases called Transmissible Spongiform Encephalopathies (TSE). BSE affects the central nervous system of cattle. The disease was first diagnosed in Great Britain in 1986. BSE has a long incubation period; from the time when an animal first becomes infected until it shows disease symptoms is presumably two to eight years. After the onset of clinical symptoms, death will occur between two weeks and six months.

*Is BSE a virus or a bacterial infection?* BSE does not appear to be a typical viral or bacterial infection. The evidence indicates a protein material or “prion” as the cause of the disease. The infective agent of BSE is resistant to heat, ultraviolet light, ionizing radiation, and common disinfectants. In addition, the infective agent causes no detectable immune or inflammatory response.

*What is a prion?* Prions have been defined as “small proteinaceous infectious particles which resist inactivation by procedures that modify nucleic acids.” Prions are formed from abnormal protease-resistant protein or PrP<sup>res</sup>. These prions have only been found in the brain, spinal cord, and retina of infected cattle. Accumulation of PrP<sup>res</sup> in brain cells will alter the function of the cells and eventually kill the cells. Prion diseases are often called spongiform encephalopathies due to the post mortem histopathologic appearance of the brain with large vacuoles (a spongy look) in the cortex and cerebellum. Specific examples include:

- Scrapie: sheep
- CWD: (chronic wasting disease): mule deer, elk
- TME: (transmissible mink encephalopathy): mink
- BSE: (bovine spongiform encephalopathy): cattle

Humans are also susceptible to several prion diseases:

- CJD: Creutzfeldt-Jakob Disease
- Kuru
- GSS: Gerstmann-Straussler-Scheinker Syndrome
- Alpers Syndrome
- FFI: Fatal Familial Insomnia

*When and where was BSE first detected?* BSE was first diagnosed and reported in Great Britain in 1986. From November 1986, when BSE was first identified as a separate disease, until February 2001, an estimated 182,000 head of cattle in almost 35,000 herds were diagnosed with BSE in Great Britain.

*Where has BSE been found?* An estimated 95% of BSE cases have been found in the United Kingdom, which includes England, Scotland, Ireland, and Wales. BSE has also been confirmed in native cattle from Austria, Belgium, Canada, Czech Republic, Finland, France, Germany, Greece, Ireland, Israel, Italy, Japan, Luxembourg, Liechtenstein, the Netherlands, Northern Ireland, Poland, Portugal, Slovakia, Slovenia, Spain, and Switzerland. Oman, the Falkland Islands, and the Azores have detected BSE, but these cases may be associated with the import of infected live animals or infected animal products for cattle feed from the United Kingdom or countries known to have BSE.

On May 20, 2003, Canada confirmed its most recent positive case of BSE in an 8-year-old black beef cow on a remote commercial farm in Alberta. Previously, only a single case of BSE in Canada had been reported in 1993; the infected cow had been imported from Great Britain.

*Have we imported cattle from BSE at-risk areas?* Between 1981 and 1989, the United States imported 334 cattle from the United Kingdom and 162 cattle from the Republic of Ireland. None of these cattle are still alive today. Additionally, 5 head of cattle imported from other European countries in 1996-97 remain under quarantine today.

*What are the symptoms of BSE in cattle?* Cattle affected by BSE first appear alert but agitated, anxious, and apprehensive. As the disease progresses, the animal begins to display an abnormal posture, uncoordinated frenzied movements with an abnormal and exaggerated gait causing tumbling, and skin wounds. The animal also loses body weight while maintaining a normal appetite. These conditions led to the terminology "Mad Cow Disease" to describe BSE.

*How do cattle get BSE?* Some scientists believe that BSE originated in England by feeding cattle rendered protein produced from the carcasses of scrapie-infected sheep. These infected cattle then were rendered and fed to cattle all over the United Kingdom causing BSE in those cattle. Scientists also hypothesize that BSE may be spread through maternal transmission either genetically or true transmission, but at such a low level this means of transmission would not sustain the BSE epidemic.

*Can animals get BSE from physical contact with BSE affected animals?* Scientific evidence indicates that BSE does not spread directly from cattle to cattle or from cattle to other species by physical contact.

*Can humans get BSE from eating beef?* Scientific evidence indicates that the infective agent for BSE has not been found in beef or dairy products, or that BSE can be transmitted through direct physical contact, close association or consumption of those products.

*What is Creutzfeldt-Jakob disease?* German scientists Hans Gerhard Creutzfeldt and Alphonse Maria Jakob first identified Creutzfeldt-Jakob Disease (CJD) in the 1920s. CJD is a rare progressive fatal encephalopathy caused by a prion and marked first by lethargy, visual disturbance, and a loss of balance. Advanced symptoms are dementia, agitation, and muscle twitching. The average length of survival after the onset of symptoms is four months. No cure is available for CJD.

*What causes Creutzfeldt-Jakob disease?* Scientific evidence points to four different modes of infection. CJD may be contracted through an inherited genetic form. Secondly, a sporadic form of CJD accounts for 85 percent of the CJD cases and has an unknown origin. Thirdly, infection could result from contact with CJD contaminated surgical equipment used for brain surgery or spinal taps. Finally, CJD could be contracted through the use of human growth hormones.

*What is the incidence of Creutzfeldt-Jakob disease?* CJD occurs at a consistent rate of approximately one case per million people per year around the world, including nations where BSE has never occurred. There have been 4,751 deaths resulting from CJD in the United States from 1979 to 1998. CJD also occurs at a consistent rate among vegetarians and meat eaters alike, which implies that its cause is not associated with meat consumption.

*What is Variant CJD?* Variant CJD (vCJD) is a new strain of CJD first detected in 10 Britons in 1996. Variant CJD had been identified as being very similar to BSE symptoms, incubation time, and brain lesions.

*How are CJD and vCJD different?* Variant CJD affects a much younger individual at an average age of 28 years old, whereas CJD affects people over the age of 63. The electrical activity of the brain with vCJD is different than CJD. BSE is more closely related to vCJD than CJD. Researchers believe that vCJD may develop in younger adults through the

consumption of BSE infected nervous system tissue. In addition, the average duration of vCJD symptoms are 10 months longer than CJD.

*What is the incidence of vCJD?* As of January 5, 2004, there have been 155 definite and probable cases of vCJD (145 in the United Kingdom, six in France, one in Ireland, one in Italy, one in Canada, and one in the United States). However, scientists concluded the patients in Canada and the United States contracted the disease while living in the United Kingdom. The patient in the United States has never donated or received blood, plasma, or organs, and the patient has no history of any major surgeries.

*Is there a test to detect BSE in live animals?* Currently there is no test to accurately detect BSE in live animals. Several European companies are applying for patents and testing procedures for a live animal BSE test. Any test made available for use must not yield false positives; the publication of false positives would have a devastating effect on beef demand. Currently, two laboratory methods are used to test for BSE: 1) microscopic examination of the brain tissue to identify characteristic changes; 2) techniques to detect the partially-proteinase resistant form of the prion protein. These techniques are immunohistochemistry, immunoblotting, and ELISA.

*What is the FDA Feed Ban?* The FDA Feed ban prohibits feeding mammalian-derived proteins to ruminants. Products that are not covered under the ban are tallow, fats, oils, grease, amino acids, dicalcium phosphate as a by-product of the gelatin manufacturing process and paunch material. The FDA Feed ban also requires persons feeding ruminant animals to maintain copies of purchase invoices and labeling for all feeds and feed ingredients containing animal protein products to show proof that no mammalian protein has been fed. The FDA rule also requires that persons feeding ruminant animals make records available to FDA for inspections. Following the confirmation of one positive BSE case in the United States, FDA issued an interim rule to implement four changes to the ban. The interim rule eliminates the exemption in the ban that allows mammalian blood and blood products to be fed to ruminants. Secondly, the rule prohibits the use of "poultry litter" as a feed ingredient for ruminant animals. Poultry litter consists of bedding, spilled feed, and fecal matter that is collected from poultry houses and fed to cattle as a protein source. Furthermore, the rule bans the use of "plate waste" as a feed ingredient for ruminant animals. Plate waste is uneaten meat and scraps collected from large restaurant operations and are then rendered into meat and bone meal for animal feed. Finally, the rule minimizes the possibility of cross-contamination of ruminant and non-ruminant feed by requiring separate equipment, facilities, or production lines for the manufacturing of non-ruminant feed that contains proteins prohibited in ruminant feed.

*What are the USDA, FDA, FSIS, NIH, CDC, and APHIS doing to prevent BSE from entering the United States?*

1. In 1986, APHIS established a BSE surveillance program in the U.S. Over 60 veterinary diagnostic laboratories participate in the BSE surveillance program along with the National Veterinary Services Laboratories in Ames, Iowa.
2. Beginning July 21, 1989, importation of live ruminants was banned from countries where BSE was known to exist.
3. December 6, 1991, USDA/APHIS banned the importation of most ruminant products from countries with known cases of BSE.
4. Since 1991, there has been a voluntary ban on the use of rendered products from adult sheep in animal feeds.
5. On August 4, 1997, an FDA regulation went into effect banning the use of mammal-derived animal by-products in cattle feed.
6. December 13, 1997, USDA/APHIS banned the import of all live ruminants and at-risk ruminant products from Europe.
7. December 7, 2000, APHIS prohibits the importation of all rendered animal protein products from Europe.
8. January 17, 2001, Purina Mills inadvertently mixed ruminant meat and bone meal in a supplement for cattle feed delivered to a feedyard in South Texas. Purina immediately notified FDA and the feedyard, which had already fed the supplement to an estimated 1,222 head of cattle. This was a regulatory issue, because since 1997, the FDA has banned the feeding of meat and bone meal supplements to cattle.
9. Inspectors from FSIS perform postmortem and antemortem slaughter inspection in all federally inspected slaughter establishments. All central nervous system disorder suspect cattle are condemned and tested for BSE.
10. USDA has trained approximately 300 state and federal veterinarians on the diagnosis of BSE.

*What other preventative measures have been implemented since the diagnosis of one BSE case in the United States?* FSIS issued three regulations and a notice in the Federal Register on January 12, 2004. The regulations and notice will minimize human exposure to materials that have been demonstrated scientifically to contain the BSE agent in infected cattle.

1. FSIS issued a notice announcing that inspectors will no longer pass and apply the mark of inspection to the carcasses and parts from cattle that are selected for testing by USDA's Animal and Plant Health Inspection Service (APHIS) for BSE until the sample is determined to be negative.
2. Brain, skull, eyes, trigeminal ganglia, spinal cord, vertebral column (excluding the vertebrae of the tail, the transverse processes of the thoracic and lumbar vertebrae, and the wings of the sacrum), and dorsal root ganglia (DRG) of cattle 30 months of age or older, as well as the tonsils and the distal ileum of the small intestine from all cattle are "specified risk materials" (SRMs). FSIS declared that SRMs are inedible, and therefore, their use in human food is prohibited.
3. Non-ambulatory disabled cattle are considered unfit for use as human food and shall be condemned. These cattle may be on the premise housing the slaughter establishment, but cannot enter the facility. FSIS has defined non-ambulatory disabled livestock as livestock that cannot rise from a recumbent position or that cannot walk, including, but not limited to, those with broken appendages, severed tendons or ligaments, nerve paralysis, fractured vertebral column or metabolic conditions.
4. In addition to a regulation prohibiting spinal cord tissue in any advanced meat recovery product labeled as "meat," FSIS expanded the prohibition to include dorsal root ganglia. Furthermore, since the vertebral column and skull of cattle 30 months or older is considered inedible, it cannot be used in advanced meat recovery.

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