NVAP Reference Guide: Bovine Spongiform Encephalopathy

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Bovine Spongiform Encephalopathy (BSE), widely referred to as “mad cow disease,” is a chronic degenerative TSE disease affecting the central nervous system of cattle. It is caused by an abnormal prion protein. It has been identified in two forms: classical BSE (C-Type) and atypical BSE forms (L-type or H-type).

C-type BSE has been linked to variant Creutzfeldt-Jakob disease (vCJD) in humans, hence the zoonosis importance of the disease. C-type BSE in cattle results from the ingestion of cattle feed (i.e. meat-and-bone meal) containing the abnormal prion protein. Feeding mammalian protein to cattle have been prohibited in the U.S. since 1997. As a result of this control measure C-type BSE cases have not been diagnosed in the U.S., except for a C-type BSE case in 2003 where the cow originated from Canada.

The Atypical BSE forms, L- and H-type, occur spontaneously at very low levels in all cattle populations, particularly older cattle 8 years of age or older and does not appear to be associated with contaminated feed. Bioassay data support the hypotheses that these strains are biologically distinct from C-type BSE and might not pose a risk to humans. In 2015, the World Animal health Organization (OIE) excluded

atypical BSE forms from the classical BSE general risk provisions.

In 2013, the OIE upgraded U.S. Status for BSE to negligible risk, the highest status available.

History

BSE was first diagnosed in 1986 in Great Britain. Since then, more than 185,000 cases have been confirmed worldwide. More than 95 percent of these have occurred in the United Kingdom, but the disease has also been confirmed in native-born cattle in the following countries: Austria, Belgium, Canada, the Czech Republic, Denmark, Finland, France, Germany, Greece, Ireland, Israel, Italy, Japan, Luxembourg, Liechtenstein, the Netherlands, Poland, Portugal, Slovakia, Slovenia, Spain, Switzerland, and the U.S.

Five cases of BSE have been identified in the U.S. The first case in 2003 was confirmed as C-type BSE and the subsequent four cases were confirmed as atypical BSE forms. The first case was detected in 2003 in Washington State in a 6 year-old dairy cow imported from Canada. The second, in 2005, was a 12 year-old beef cow in Texas. The third, in 2006, was a 10 year-old beef cow in Alabama. The fourth, in 2012, was a 10 year-old dairy cow in California. The fifth case, in 2017, was an 11 year-old beef cow in Alabama. As a result, U.S. beef exports dropped 80% after 2003 and have not yet fully recovered. It was not until May 2017 that China allowed U.S. beef exports to flow directly to the country since 2003 when the C-type BSE case was confirmed in U.S.

Clinical Course

Cattle affected by BSE experience progressive degeneration of the nervous system. Affected animals may display changes in temperament (nervousness or aggression), abnormal posture, incoordination and difficulty in rising, decreased milk production, or loss of condition without noticeable loss of appetite. There is no treatment or vaccine to prevent BSE. The incubation period 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.

Etiologic Agent and Tissue Distribution

The agent responsible for BSE has not been completely characterized. There are a variety of theories regarding the nature of the agent. The most widely accepted is that disease is caused by an abnormal prion protein that accumulates in the central nervous system and causes the normal cellular version of the protein to change shape such that it can no longer be degraded by the cell, causing the protein to accumulate and damage the cell.

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, evidence of disease 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 from experimentally infected cattle were also found to be infective.

Transmission

BSE is not a contagious disease and therefore is not spread through casual contact between cattle or with other species. The primary source of C-type 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 the BSE agent. Therefore, rendered protein such as meat-and- bone meal derived from infected animals may contain the infectious agent. The preferred method for disposal of BSE-infected carcasses is alkaline digestion or complete, high- temperature incineration. Under no circumstances should BSE suspects be used for human or animal consumption.

Testing

Currently, there is no validated test to detect the disease in a live animal. BSE testing is done by examining the obex portion of the brain stem for the accumulation of abnormally folded prion protein. BSE screening is generally done by enzyme-linked immunosorbent assay (ELISA) on fresh tissue. BSE can be confirmed by

immunohistochemistry or Western Blot. Histopathology may also be utilized to look for spongiform changes in brain stem tissue that are characteristic of the TSEs.

The accumulation of abnormal prion protein material and other changes in brain stem tissue that are diagnostic for BSE are not apparent in the early stages of the disease. Thus, the failure to detect BSE is not equivalent to a negative test or the absence of infectivity. Current testing methods are surveillance tools only. They are not intended to protect human health or animal health nor can they guarantee food safety.

Surveillance

BSE surveillance has been conducted in the U.S. since 1990. After the initial case of BSE was detected in the U.S. in late 2003, APHIS conducted a BSE Enhanced Surveillance Program from 2004 to 2006. This was a one-time intensive effort to detect BSE if present at a very low level and to provide information about prevalence. More than 830,000 animals were tested. Subsequent data analysis indicated that the prevalence of BSE in the U.S. was very low—less than one infected animal per million based on a population of 42 million adult cattle.

Beginning in 2006, the BSE Ongoing Surveillance Program was implemented. This program follows a stringent U.S. standard of detecting one case of BSE per one million adult cattle with 95% confidence. This standard far exceeds the current surveillance standards provided by the OIE for the U.S. under BSE negligible risk or *Type B* surveillance which is to detect one case of BSE per 50,000 adult cattle with 95% confidence. Both standards are based on a point system that reflects the likelihood of finding BSE. The BSE Ongoing Surveillance Program focuses on populations of cattle at higher risk for BSE, including those animals that are 12 months of age and older that display CNS signs and those over 30 months of age that are condemned on ante-mortem inspection at slaughter and thus are excluded from slaughter due to poor health status (non-ambulatory, unhealthy, or dead). From 2006 to 2015, the BSE Ongoing Surveillance Program tested approximately 40,000 samples per year. On 2015, the BSE sample collections were reduced to 25,000 per year. In the last 11 years the U.S. BSE sample collection resulted in point totals that far exceed both the OIE and U.S. requirements.

Eradication and Control Efforts

Agricultural officials in the United Kingdom and other countries affected with BSE have taken actions to eradicate or control the disease. These entail prohibiting the inclusion of mammalian meat-and-bone meal in animal feed; prohibiting the use of specified risk materials or SRMs (those tissues, e.g., brain and spinal cord, known to have the highest infectivity) in food, feed, or other products; and destroying 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 in the United Kingdom has greatly decreased.

In 1997, the U.S. Food and Drug Administration (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. In 2008, the ban was strengthened by prohibiting the inclusion of SRMs (brains and spinal cords from animals 30 months of age or older) in any animal feed. The 2008 rule also prohibits the use of entire carcass of cattle not inspected and passed for human consumption, unless the cattle are less than 30 months of age, or the brains and spinal cords have been removed.

Public Health Significance

In March 1996, the United Kingdom’s Spongiform Encephalopathy Advisory Committee (SEAC) announced the identification of 10 cases of variant Creutzfeldt-Jakob disease (vCJD) in people. These cases had a characteristic clinical and pathological phenotype that differed from other routinely diagnosed cases of classic (sporadic) CJD. SEAC concluded, and scientific evidence later confirmed, that these cases were linked to exposure to C-type BSE before the feed bans were implemented. It is not known whether atypical forms of BSE (L- or H-type) are casually linked to forms of human prion diseases.

In the U.S., public or human health protective measures are maintained by both the Food Safety and Inspection Service (FSIS) and the FDA. The most important public health protective measure is the removal of SRMs from the human food supply. 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 from slaughter of animals with any clinical signs of neurological disease or other abnormalities.

Identifying Affected Animals

The U.S. also maintains import regulations, consistent with the OIE guidelines for BSE, to prevent BSE from entering the US through imports. Beginning in 1989, APHIS prohibited the importation of ruminants and most ruminant products from countries that had identified BSE in native cattle or that were at risk for BSE.

An ongoing, comprehensive interagency surveillance program for BSE has been in place in the U.S. since 1990. APHIS also supports the FDA’s regulation prohibiting the use of most mammalian proteins in ruminant feed. Currently, USDA allows the importation of some animals and commodities under permit or by regulation from minimal BSE-risk countries.

Because of the clinical history that can be obtained, samples collected on the farm from cattle exhibiting symptoms of central nervous system disease are particularly valuable to the BSE Ongoing Surveillance Program efforts in the U.S. Accredited veterinarians with proper training can play a key role in the sampling of these animals and should contact their VS field office for more information. BSE is a reportable disease. Any suspicious cases should be reported to the APHIS – VS Area Offices or the State animal health official as a suspected foreign animal disease (FAD).

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