

# Bovine Spongiform Encephalopathy

Bovine Spongiform Encephalopathy (BSE), also known colloquially as Mad Cow disease, is a transmissible spongiform encephalopathy (TSE) disease, affecting cattle. TSE disorders are thought to be caused by an infectious, misfolded prion protein found in the brain.<sup>1</sup> BSE is believed to be transmitted by consumption of nervous tissue containing infectious prion protein. Some BSE symptoms include ataxia (uncoordinated gait, falling) and altered behaviors (i.e., general change in temperament, aggression). Initial signs are subtle and behavioral and most animals that contract the disease reach a terminal state by three months after clinical onset.<sup>2</sup> Research into BSE transmission methods indicates that eliminating exposure to the specified risk materials from BSE infected cattle stops the spread of the disease. Furthermore, the United States Food and Drug Administration imposed the Ruminant to Ruminant Feed Ban in 1996 which prohibits the use of specific animal products in ruminant (beef cattle) feed.

Though the method of transmission is not well understood, it is known that “BSE is a progressive neurological disorder of cattle that results from infection by an unusual transmissible agent called a prion.” As a result of the “strong epidemiologic and laboratory evidence for a causal association between a new human prion disease called variant Creutzfeldt-Jakob Disease (vCJD)” which made its first appearance in the United Kingdom in 1996 during the height of that country’s worst outbreak of BSE,<sup>3</sup> identification of transmission vectors which enable BSE to pass to humans are being researched worldwide. According to the USDA Food Safety and Inspection Service, the specified risk material (SRM) which has the potential for carrying the infective agent for BSE are as follows: the brain, skull, eyes, trigeminal ganglia, spinal cord, vertebral column (excluding the tail vertebrae, thoracic and lumbar transverse processes, and sacral wings) and dorsal root ganglia (DRG) of cattle 30 months of age and older, and the tonsils and distal ileum of the small intestine of all cattle, regardless of age.<sup>4</sup> Further specific descriptions of portions of the cattle carcass which are considered “prohibited from being used in edible rendering” may be found at 9 CFR 318.6 b (4).

Atypical, H-type, BSE was studied in Sweden in 2008 after that country’s first case of BSE was detected in a 12 year old cow in 2006. The Swedish experiments were inconclusive as to the “origin and infectivity of atypical BSE cases for humans and animals.”<sup>5</sup> However, research coming out of Germany indicates successful transmission of C-Type (typical) and H-Type and L-Type (atypical) BSE to lab mice.<sup>6</sup> With the proper application of current regulations and laws

---

<sup>1</sup> G.A.H. Wells and J.W. Wilesmith, “Bovine Spongiform Encephalopathy and Related Diseases,” in *Prion Biology and Diseases*, 2nd Ed., (Cold Spring Harbor, NY: Cold Spring Harbor Laboratory Press, 2004), 595-628.

<sup>2</sup> *Merck Veterinary Manual*, 9<sup>th</sup> Edition, ed. C. Kahn (Whitehouse Station, NJ: Merck & Co., 2008), accessed April, 2010, <http://www.merckvetmanual.com/mvm/index.jsp>.

<sup>3</sup> “BSE (Bovine Spongiform Encephalopathy, or Mad Cow Disease),” Centers for Disease Control and Prevention, last modified April 22, 2010; accessed May 13, 2010; <http://www.cdc.gov/ncidod/dvrd/bse>.

<sup>4</sup> “Bovine Spongiform Encephalopathy (BSE): Key Points for the Public Health Veterinarian,” United States Department of Agriculture, Food Safety Inspection Service, March 29, 2009; accessed September 1, 2010, <http://origin-www.fsis.usda.gov/PDF/PHVt-BSE.pdf>.

<sup>5</sup> Gavier-Widen, et al., “Bovine spongiform encephalopathy in Sweden: an H-type variant,” *Journal of Veterinary Diagnostic Investigation* 20 (2008):2-10, accessed August 31, 2010, <http://jvdi.org/cgi/content/abstract/20/1/2>.

<sup>6</sup> Buschmann, et al., “Atypical BSE in Germany--proof of transmissibility and biochemical characterization,” *Veterinarian Microbiology* 117 (2006):103-16, accessed August 31, 2010 at <http://www.ncbi.nlm.nih.gov/pubmed/16916588>.

governing removal of SRM's, USDA believes meat products taken from undetected classical (C-Type) or atypical (H-Type, L-Type, M-Type) BSE infected cattle do not provide a threat to public safety. Consequently, the risk associated with traceback covered by the uslp Warranty would be somewhat mitigated by negligent liability of processing plants.

There have been three cases of BSE identified in cattle in the United States. The first reported case occurred in December of 2003 when a six year old Holstein cow in the state of Washington was diagnosed with the disease. The resulting investigation determined this cow had been imported to the United States from the province of Alberta, Canada in 2001 as part of a shipment of 81 animals. The cow had produced two calves during the period 2001 through 2003 and these animals were identified and also destroyed. Of the original 81 imported cattle, 25 were classified as high-risk of which the USDA was able to account for 14.<sup>7</sup>

The second case of BSE in the United States occurred in June of 2005 in a 12 year old cow in Texas. The Texas outbreak was the first endemic case in the United States.<sup>8,9</sup> Though multiple sources, including USDA Foreign Agricultural Service and Food and Drug Administrations, confirmed this occurrence of BSE in Texas, it was not reported in the data received from OIE.

The third and most recent outbreak occurred in March of 2006. A ten year old cow in Alabama was diagnosed with the disease. The affected animal lacked identification or distinctive marks. Intensive investigation failed to determine the animal's origin because of these limitations. In August 2008, several investigators reported that a rare, genetic abnormality that may persist within the cattle population at large is considered to have caused atypical (H-type) BSE in the Alabama animal. In an atypical H-type BSE occurrence, it is hypothesized that BSE occurs spontaneously rather than through feed transmission.<sup>10</sup> The following chart illustrates the number of cattle destroyed or slaughtered due to BSE in the United States. These numbers reflect the trace-out and trace-in cattle destroyed or slaughtered as a result of their association with the affected cattle.

---

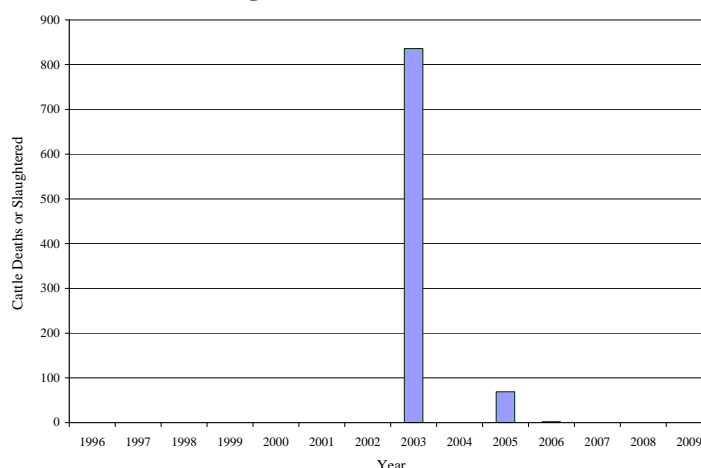
<sup>7</sup> "Bovine Spongiform Encephalopathy," California Department of Food and Agriculture, accessed August 31, 2010, [www.cdffa.ca.gov/ahfss/Animal\\_Health/BSE\\_info.html](http://www.cdffa.ca.gov/ahfss/Animal_Health/BSE_info.html).

<sup>8</sup> "BSE (Bovine Spongiform Encephalopathy, or Mad Cow Disease)," Centers for Disease Control and Prevention, last modified April 22, 2010; accessed May 13, 2010; <http://www.cdc.gov/ncidod/dvrd/bse>.

<sup>9</sup> "Bovine Spongiform Encephalopathy," California Department of Food and Agriculture, accessed August 31, 2010, [www.cdffa.ca.gov/ahfss/Animal\\_Health/BSE\\_info.html](http://www.cdffa.ca.gov/ahfss/Animal_Health/BSE_info.html).

<sup>10</sup> "BSE (Bovine Spongiform Encephalopathy, or Mad Cow Disease)," Centers for Disease Control and Prevention, last modified April 22, 2010; accessed May 13, 2010; <http://www.cdc.gov/ncidod/dvrd/bse>.

### Cattle Deaths or Slaughtered Due to BSE (U.S. 1996 to 2009)



Source: Final epidemiology reports for the Texas and Alabama outbreaks and the Summary Report for the Washington outbreak as published by APHIS.

Though there have been only three cattle in the United States positively confirmed to have been infected with BSE, USDA APHIS has depopulated or euthanized 907 cattle.

According to the 2008 U.S. Animal Health Report published by APHIS, a 2006 analysis of surveillance data concluded that BSE might occur in this country, but levels would be extremely low – less than one case per million in the U.S. adult cattle population.<sup>11</sup> This study was done using the BSurvE model (Wilesmith et al., 2004) and also the Bayesian Birth Cohort (BBC) model. The results of the two methods were similar, with the expected number of infected adult cattle at 4 and 7 respectively for BBC and BSurvE. The 90 percent confidence interval of BSE infected cattle for the two models was between 1 and 8 and between 3 and 24 respectively in a population of approximately 42,000,000 cattle. Both models strongly support a conclusion that the prevalence of BSE in the United States is below one case per million adult cattle.<sup>12</sup>

All beef products offered for sale in the retail/wholesale markets must be inspected through the federal (interstate/international export markets) or state (intrastate markets) meat and poultry inspection program.<sup>13</sup> Furthermore, each federally or state inspected packing plant must, by law, have a Hazard Analysis and Critical Control Point (HACCP) plan which includes the removal and disposal of all SRMs prior to carcass fabrication.<sup>14</sup> This redundant system of meat and meat byproduct inspection results in the opportunity for fabricated meat and meat byproducts becoming contaminated with prions being prevented. Furthermore, currently, treatment of BSE is ineffective and methods to control the contraction and spread of BSE in both Europe and the U.S. center on banning the use of mammalian-derived protein in the rations for all farm animal

<sup>11</sup> “2008 U.S. Animal Health Report,” United States Department of Agriculture, Animal and Plant Health Inspection Service (APHIS), pp. 21, accessed April, 2010, [http://www.aphis.usda.gov/animal\\_health/animal\\_health\\_report/downloads/AHR\\_08/2008\\_US\\_Animal\\_Health\\_Report.pdf](http://www.aphis.usda.gov/animal_health/animal_health_report/downloads/AHR_08/2008_US_Animal_Health_Report.pdf).

<sup>12</sup> “An Estimate of the Prevalence of BSE in the United States,” United States Department of Agriculture, Animal and Plant Health Inspection Service (APHIS), 2006, accessed April, 2010, [http://www.aphis.usda.gov/newsroom/hot\\_issues/bse/downloads/BSEprev-estFINAL\\_7-20-06.pdf](http://www.aphis.usda.gov/newsroom/hot_issues/bse/downloads/BSEprev-estFINAL_7-20-06.pdf).

<sup>13</sup> “Rules of Practice Governing Proceedings under the Animal Welfare Act.” Code of Federal Regulations Title 9, Pt. 4, 2010.

<sup>14</sup> “Validation, Verification, Reassessment.” Code of Federal Regulations Title 9, Pt 417.4(a)(3), 2010 and “Specified Risk Materials from Cattle and their Handling and Disposition.” Code of Federal Regulations Title 9, Pt 310.22(e)(1), 2010.

species.<sup>15</sup> U.S. regulations “provide for the importation of certain ruminants, and ruminant products and byproducts from regions that pose a minimal risk of introducing bovine spongiform encephalopathy (BSE) into the United States (U.S.),...”<sup>16</sup> USDA APHIS has developed an overview document describing BSE and the USDA actions in response to a BSE outbreak. This document may be found at:  
[http://www.aphis.usda.gov/publications/animal\\_health/content/printable\\_version/BSEbrochure12-2006.pdf](http://www.aphis.usda.gov/publications/animal_health/content/printable_version/BSEbrochure12-2006.pdf).

The APHIS Factsheet on BSE, namely “Bovine Spongiform Encephalopathy Epidemiologic Process and Testing Protocols,” may be found at  
[http://www.aphis.usda.gov/publications/animal\\_health/content/printable\\_version/fs\\_bsetestingprotocols.pdf](http://www.aphis.usda.gov/publications/animal_health/content/printable_version/fs_bsetestingprotocols.pdf).

The last animal documented to be infected with BSE in the United States died in 2006. Canada had a confirmed outbreak in February 2010. Mexico reports no occurrences of BSE in their history. BSE occurs sporadically and sometimes spontaneously worldwide. There have been no documented cases of BSE being horizontally transmitted between live cattle.

In addition to the response protocols in force in the United States and the specified removal of SRMs from slaughtered animals, the animal health industry has also implemented training and testing classes for all levels of animal health practitioners and inspectors. Practitioners are given intense training on medical identifiers for the disease and current medical research relating to identifying the disease in a suspect animal visually, extraction of tissue samples from the suspected animal, proper protocols for tissue handling and chain of custody procedures, laboratory testing procedures, confirmation of infection protocols, notification procedures, among others. Inspectors and surveillance personnel within the industry are trained and tested on the visual indicators of the disease on cattle carcasses at the slaughter facility, visual inspection techniques for carcasses, visual inspection techniques for ensuring proper and complete SRM removal, notification procedures for suspect cases of infection, and proper handling and security of the suspect carcass and tissue. Coordination efforts between and among federal and state agencies, both civilian and law enforcement, are practiced, assessed and modified during focused worst case scenario (FMD and High Path Avian Influenza) training situations and response protocols are practiced. Lessons learned from these simulated exercises are used by public and animal health professionals to keep abreast of current protocols and procedures during emergency response situations. Producers and stakeholders are also invited to attend and participate in those training modules relevant to their particular area of interest.

---

<sup>15</sup> “An Estimate of the Prevalence of BSE in the United States,” United States Department of Agriculture, Animal and Plant Health Inspection Service (APHIS), 2006, accessed April, 2010,  
[http://www.aphis.usda.gov/newsroom/hot\\_issues/bse/downloads/BSEprev-estFINAL\\_7-20-06.pdf](http://www.aphis.usda.gov/newsroom/hot_issues/bse/downloads/BSEprev-estFINAL_7-20-06.pdf).

<sup>16</sup> “Bovine Spongiform Encephalopathy: Minimal-Risk Regions and Importation of Commodities,” United States Department of Agriculture, Animal and Plant Health Inspection Service (APHIS), (Docket No. 03-080-8, 70 FR 460 – 553), accessed May, 2010, [http://www.aphis.usda.gov/newsroom/hot\\_issues/bse/downloads/Docket2006-0026-3.doc](http://www.aphis.usda.gov/newsroom/hot_issues/bse/downloads/Docket2006-0026-3.doc).