



BOVINE SPONGIFORM ENCEPHALOPATHY ¹ *EI -- 03-04*

What we know about BSE. ²

Bovine spongiform encephalopathy (BSE), widely known as "mad cow disease," is a chronic, degenerative disease affecting the central nervous system of cattle. Worldwide there have been more than 180,000 cases since 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 has also been confirmed in native-born cattle in Austria, Belgium, Canada, Czech Republic, Denmark, Finland, France, Germany, Greece, Ireland, Israel, Italy, Japan, Luxembourg, Liechtenstein, the Netherlands, Northern Ireland, Poland, Portugal, Slovakia, Slovenia, Spain and Switzerland. However, over 95% of all BSE cases have occurred in the United Kingdom. BSE was not known to exist in the United States until a Holstein dairy cow was diagnosed on December 23, 2003 in Washington State.

BSE belongs to the family of diseases known as the transmissible spongiform encephalopathies (TSE's). These diseases are caused by a transmissible agent which is yet to be fully characterized. They share the following common characteristics

- a. a prolonged incubation period of months or years;
- b. a progressive debilitating neurological illness which is always fatal;
- c. when examined by electron microscopy, detergent treated extracts of brain tissue from animals or humans affected by these diseases reveal the presence of scrapie associated fibrils (SAF);
- d. pathological changes appear to be confined to the CNS and include vacuolation, and astrocytosis;
- e. the transmissible agent elicits no detectable specific immune response in the host which has inhibited the development of a preclinical live animal diagnostic test to date.

Similar Diseases of Humans and Other Animals

TSE's are caused by similar uncharacterized agents which usually produce spongiform changes in the brain. TSE's include scrapie (which affects sheep and goats), transmissible mink encephalopathy, feline spongiform encephalopathy, chronic wasting disease of deer and elk, and in humans, kuru, Classical Creutzfeldt-Jakob Disease (CJD), Gerstmann- Straussler syndrome, fatal familial insomnia, and vCJD. More about scrapie, transmissible mink encephalopathy, and chronic wasting disease are available on the APHIS web site on BSE.

Clinical Signs of BSE in Cattle

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 condition despite continued appetite. There is no treatment, and affected cattle die.

The incubation period ranges from 2 to 8 years. Following the onset of clinical signs, the animal's condition deteriorates until it dies or is destroyed. This 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.

The Causative Agent of BSE

¹ This information has been excerpted from the APHIS web site list elsewhere in this paper. Information is updated at the APHIS site regularly.

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The causative agent of BSE as well as other TSE's is yet to be fully characterized. Three main theories on the nature of the agent have been proposed:

- An unconventional virus.
- A prion or abnormal partially-proteinase K-resistant protein, devoid of nucleic acid, capable of causing normal prion protein in the host to change and form more abnormal protein.
- A virino or "incomplete" virus composed of naked nucleic acid protected by a host protein.

The BSE agent (1) is smaller than most viral particles and is highly resistant to heat, ultraviolet light, ionizing radiation, and common disinfectants that normally inactivate viruses or bacteria; (2) causes no detectable immune or inflammatory response in the host; and (3) has not been observed microscopically.

How BSE Is Currently Diagnosed

There is no test to detect the disease in a live animal. Currently there are two laboratory methods to confirm a diagnosis of BSE: 1. microscopic examination of the brain tissue to identify characteristic changes; 2. techniques to detect the partially-proteinase resistant form of the prion (PrP^{res}) protein. These techniques are immunohistochemistry, immunoblotting and ELISA.

What risk is there of a BSE outbreak occurring in the United States?

There are still a number of unknowns regarding the origin and transmission of BSE. Given these scientific uncertainties, we cannot assure zero risk from BSE. However, we can and will continue to monitor new scientific findings and world events and adjust our regulations and policies to keep the risk of BSE infecting the national herd as low as possible.

What About Other Animal TSE's in the US?

These other TSE's HAVE been found in the United States: Scrapie in sheep and goats, transmissible mink encephalopathy, and chronic wasting disease of deer and elk.

The Cause of BSE in Great Britain

Epidemiological data suggest that BSE in Great Britain is a common-source epidemic involving animal feed containing contaminated meat and bone meal as a protein source. The causative agent is suspected to be from either scrapie- affected sheep or cattle with a previously unidentified TSE. Changes in rendering practices in the late 70's—early 1980's may have potentiated the agent's survival in meat and bone meal.

For more information about BSE in the United Kingdom, please visit the Department for Environment, Food and Rural Affairs (formerly the Ministry of Agriculture, Fisheries and Food, UK) web site at <http://www.defra.gov.uk/animalh/bse/index.html>.

Countries Other Than the United Kingdom With Confirmed Cases of BSE

In native cattle: Austria, Belgium, Czech Republic, Denmark, Finland, France, Germany, Greece, Ireland, Israel, Italy, Japan, Luxembourg, Liechtenstein, the Netherlands, Northern Ireland, Poland, Portugal, Slovakia, Slovenia, Spain and Switzerland. While there is a decline in the number of cases of BSE in the United Kingdom, confirmed cases of BSE have risen in other European countries.

Oman, the Falkland Islands, Canada, and the Azores have detected BSE in cattle imports from other countries known to have BSE.

On May 20, 2003, Canada had a cow test positive for BSE. Previously, there had been one case of BSE in a single cow in Canada in 1993. It had been imported from Great Britain and was dealt with by destroying the affected cow and all its herdmates, as well as other cattle determined to be a risk by animal health officials in Canada.

On December 23, 2003 a Holstein dairy cow tested positive for BSE in the state of Washington. The animal, along with about 80 herdmates, was imported into the U.S. from southern Alberta, Canada. All animals and their offspring are being traced and will be tested. It is presently suspected that a common contaminated feed source may be

responsible in both cases. Both animals were born before the feed ban was put into effect in August, 1997 in both Canada and the U.S.

For more information, please see Office International des Epizooties at http://www.oie.int/eng/info/en_esb.htm.

Transmission of BSE

There is no evidence that BSE spreads horizontally, i.e., by contact between unrelated adult cattle or from cattle to other species. Some evidence suggests that maternal transmission may occur at an extremely low level. Results of British research show that there is approximately a 9-percent increase in the occurrence of BSE in offspring of BSE-affected dams as compared to calves born to dams where BSE was not detected. The study did not ascertain if this was the result of genetic factors or true transmission. The research did however point out that at this level if maternal transmission does occur it alone will not sustain the epidemic (Wilesmith et al. 1997).

A recently published study found no evidence of disease transmission via embryos collected from cows with BSE. The embryos were collected and handled in accordance with international health standards (Wrethall et. al., 2001).

About Classical Creutzfeldt-Jakob Disease (CJD)

CJD is a slow degenerative disease which affects the central nervous system of humans. CJD occurs sporadically worldwide at a rate of approximately 1 case per 1 million people per year. More rare are the other TSE conditions affecting humans: Gerstmann- Straussler syndrome, kuru, vCJD, and fatal familial insomnia. More is available at <http://www.cjd.ed.ac.uk/>.

Classical CJD in the USA and in Britain

The incidence of classical CJD in the United States (about 1 case per 1 million population per year) is similar to the incidence found in the rest of the world, which includes Australia and New Zealand--countries that have NOT reported either scrapie or BSE. CJD, which was first diagnosed in the 1920's, occurs with roughly the same frequency in Britain as in other countries at the present time.

For more information on CJD in the United States, please visit the Centers for Disease Control and Prevention's National Center for Infectious Diseases website at <http://www.cdc.gov/ncidod/index.htm>.

BSE and vCJD—Human Health Concerns

On March 20, 1996, the UK's Spongiform Encephalopathy Advisory Committee (SEAC) announced the identification of 10 cases of a new variant form of CJD (vCJD). All of the patients developed onset of illness in 1994 or 1995. The following features describe how these 10 cases differed from the sporadic form of CJD:

- The affected individuals were much younger than the classical CJD patient. Typically, CJD patients are over 63 years old. The average patient age for the onset of variant CJD was 28 (range of 12 to 74) years.
- The course of the disease in the vCJD averaged 14 months. Classical CJD cases average a 4–6 month duration.
- In the variant cases, electroencephalographic (EEG) electrical activity in the brain was not typical of classical CJD.
- Although brain pathology was recognizable as CJD, the pattern was different from sporadic CJD, with large aggregates of prion protein plaques.

Epidemiological and case studies have not revealed a common risk factor among the cases of vCJD. According to the SEAC, all victims were reported to have eaten beef or beef products in the last 10 years, but none had knowingly eaten brain material. One of the affected individuals had been a vegetarian since 1991.

The 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 control measures, in particular, the specified bovine offal (SBO) ban in 1989.

Research reported in later 1996 and 1997 has found evidence to further support a causal association between vCJD and BSE. Two significant studies published in the October 2, 1997 edition of *Nature* lead the SEAC to conclude that BSE agent is highly likely to be the cause of vCJD. Dr. Moira Bruce and colleagues at the Institute for Animal Health in Edinburgh, Scotland inoculated 3 panels of inbred mice and one panel of crossbred mice with BSE, vCJD and sporadic CJD. Results indicate that mice inoculated with BSE showed the same pattern of incubation time, clinical signs and brain lesions as mice inoculated with tissues from patients with vCJD. This provides evidence that BSE and vCJD have the same signature or are the same "strain". In addition, sporadic CJD and known scrapie strains were not similar to vCJD or BSE.

Results from a study published by Dr. John Collinge and colleagues of Imperial College School of Medicine, London, UK strongly support Bruce's results. Collinge's paper reports findings of BSE transmission to transgenic mice expressing only human PrP.

Another paper by Collinge et. al. in the October 24, 1996 edition of *Nature* also provides data to support the association between vCJD and BSE.

More recently, studies using transgenic animals expressing the bovine PrP have supported the view that BSE infected cattle are responsible for vCJD. These mice not only propagated the BSE infectious agent in the absence of a species barrier, but also were highly susceptible to vCJD. Furthermore, the transgenic mice inoculated with either vCJD or BSE had indistinguishable disease characteristics.

FOR FURTHER READING TRY THESE LINKS

Consumers with food safety questions can phone the toll-free USDA Meat and Poultry Hotline at 1-888-MPHotline. The hotline is available in English and Spanish and can be reached from 10 a.m. to 4 p.m. (Eastern Time), Monday through Friday. Recorded food safety messages are available 24 hours a day.

USDA's Animal and Plant Health Inspection Service web site
<http://www.aphis.usda.gov/lpa/issues/bse/bse.html>

ERS (Economics Research Service), USDA updated information on the beef sector, BSE, etc:
<http://www.ers.usda.gov/News/BSECoverage.htm>

A Chronology of Events surrounding the BSE Case in the US is available at:
<http://www.usda.gov/news/releases/2003/12/bsechronology.htm>

Beef Industry information page
www.BSEInfo.org

The Livestock Marketing Information Center
www.lmic.info

Kansas State University's Ag Manager Livestock Page
<http://www.agmanager.info/livestock/>

Iowa State University Beef Center
<http://www.iowabeefcenter.org/>

Centers for Disease Control and Prevention's National Center for Infectious Diseases
<http://www.cdc.gov/ncidod/index.htm>

Canadian Food Inspection Agency

<http://www.inspection.gc.ca/english/anima/heasan/disemala/bseesb/bseesbindexe.shtml>

U. S. Food and Drug Administration BSE hot topics page

<http://www.fda.gov/oc/opacom/hottopics/bse.html>

Food Safety Inspection Service

<http://www.fsis.usda.gov/oa/topics/bse.htm>

Biotechnology Industry Organization

<http://science.bio.org/bse.news.html>

Government Accounting Office 2002 study on the animal feed ban and BSE in the U. S.

<http://www.gao.gov/new.items/d02183.pdf>

University of Illinois at Urbana-Champaign BSE information page

<http://w3.aces.uiuc.edu/AnSci/BSE>