

Bovine Spongiform Encephalopathy in the United Kingdom, 1986 – 2003

An epidemic of bovine spongiform encephalopathy (BSE) was first recognized in the United Kingdom in 1986 and has since affected over 179,900 cattle on more than 35,740 U.K. farms (as of 28 February 2003). The epidemic peaked in January 1993 at almost 1,000 new cases per week. This outbreak may have resulted from the feeding of scrapie-containing sheep meat-and-bone meal to cattle or it may have arisen as a rare spontaneous formation of a spongiform encephalopathy in a cow that then spread to other cattle through contaminated meat and bone meal. There is strong evidence and general agreement that the outbreak was amplified by feeding rendered bovine meat-and-bone meal to young calves. An outbreak of new-variant Creutzfeldt-Jakob disease (vCJD) that has killed 127 people (as of April 2003) is most likely a result of the consumption of beef products contaminated by central nervous system tissue from cattle with BSE. As of April 2003, a total of 134 cases of vCJD had been reported, mainly in the United Kingdom. An average of 10–15 cases have been seen each year since vCJD emerged in 1994; the magnitude and geographic distribution of this epidemic in countries that have imported British cattle or cattle products cannot yet be predicted. The possibility that large numbers of apparently healthy people might be incubating the disease raises concerns about the possibility of transmission through medical instruments and blood and organ donations.

BSE developed into an epidemic as a consequence of an intensive farming practice – the recycling of animal protein in ruminant feed. The question of how to handle the BSE agent, a known hazard to cattle and potential hazard to humans, is key to the BSE story. The government took measures to address both hazards, but they were not always timely or adequately implemented and enforced because the basic biology of BSE was unknown and it was believed that BSE was not a potential threat to human life.

What are BSE and vCJD?

BSE is a progressive neurological disorder of cattle that results from infection by an unconventional transmissible agent. The causative agent of BSE and other transmissible spongiform encephalopathies (TSEs) is yet to be fully characterized. The BSE agent 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. It causes no detectable immune or inflammatory response in the host and has not been observed microscopically. The incubation period for BSE ranges from two to eight years and clinical disease usually occurs in older animals. Most cases in Great Britain have been seen in dairy cows between three and six years of age. 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. Following the onset of clinical signs, the animal's condition deteriorates until it dies or is destroyed. This usually takes from two weeks to six months. There is no treatment.

Creutzfeldt–Jakob disease (CJD) is a rare and fatal human neurodegenerative disease of unknown cause. Patients with the conventional form are usually between 50 and 75 years of age. The new variant form (vCJD) in the United Kingdom mainly affects younger people; the median age at death is 28 years.

The first cases of BSE

Individual cattle were probably first infected by BSE in the 1970s. If they lived long enough to develop signs of disease, these were not reported to or investigated by the Central Veterinary Laboratory (CVL) of the State Veterinary Service (SVS). The first clinical cases were reported in 1984, although it was two years before the nature of the disease was actually recognized.

On December 22, 1984, Peter Stent of Pitsham Farm in Sussex called Dr. David Bee, a private veterinarian, to examine Cow 133. The cow had an arched back and had lost weight. Dr. Bee visited the farm several times over the following months, and continued to see animals showing unusual symptoms. Cow 133 developed a head tremor and incoordination before dying on February 11, 1985. By the end of April, five more cows on the farm had died. Dr. Bee requested assistance from Dr. J. M. Watkin–Jones, a veterinarian at the Winchester Veterinary Investigation Center (VIC) of the Veterinary Investigation Service. A number of samples of body tissue were submitted to the CVL for pathological analysis. Various possible ailments were identified, but despite a wide range of tests there was no definite diagnosis. The CVL suggested that Mr. Stent submit a live affected cow for slaughter and post–mortem. Cow 142 was sent live to the CVL in September for euthanasia and a post–mortem examination. The pathologist on duty examined the tissues and concluded that the problem was associated with fungal contamination of feed and mycotoxin production. An April 1985 laboratory report stated that a fungal toxin called citrinin had been found in the feed at the farm. New cases ceased to develop on the farm and the veterinarians assumed that the problem had run its course (from www.bseinquiry.gov.uk/report/volume3/chapterd.htm).

The mysterious disease soon reappeared, however, on other farms. At the end of 1986, the Pathology Department of the CVL considered four more cases of unusual neurologic disease in cattle from farms in Kent and Bristol. They identified these cases as a probable transmissible spongiform encephalopathy in cattle and named the new disease bovine spongiform encephalopathy. By the end of 1987, the CVL Epidemiology Department concluded that the cause of the reported cases of BSE was the consumption of meat–and–bone meal (MBM), which was made from animal carcasses and incorporated into cattle feed. At first it was thought that the source of infection in the MBM was tissues derived from sheep infected with conventional scrapie and that the MBM had become infectious because rendering methods that had previously inactivated the conventional scrapie agent had been changed. However, the cases of BSE identified between 1986 and 1988 were not index cases, and they were not the result of the transmission of scrapie. They were the consequences of recycling BSE–infected cattle into

MBM. BSE probably originated from a novel source early in the 1970s, possibly a cow or other animal that developed disease spontaneously. The origin of the disease will probably never be known. The theory that BSE resulted from changes in rendering methods is probably not correct because rendering methods have never been capable of completely inactivating TSEs.

Precautions taken

In June 1988, the Southwood Working Party, set up to provide advice on the implications of BSE, recommended that cattle showing signs of BSE be destroyed and that compensation be paid to farmers. In February 1989, the Southwood Working Party submitted a report to the government that concluded that the risk of transmission of BSE to humans appeared remote and that 'it was most unlikely that BSE would have any implications for human health.' This assessment of risk was made assuming that BSE was probably derived from scrapie and could be expected to behave like scrapie. The Southwood Report never underwent a scientific review by experts in the field. Precautionary measures were put in place that went beyond those recommended by the Working Party and an expert committee was set up to advise BSE research.

Once MBM was identified as the probable vector of BSE in 1988, the government implemented a ban on incorporating ruminant protein in ruminant feed. This ban reduced the escalating rate of infection. After BSE was experimentally transmitted to a pig in 1990, new measures to protect pigs and poultry from BSE were introduced. However, the measures were unenforceable and widely disregarded. It was later discovered that a cow can become infected with the BSE agent by eating an amount of infectious tissue as small as a peppercorn. Cross-contamination in feedmills caused thousands of cattle to become infected, but because of the long incubation period this was not apparent until later. In 1994, because of the continuing infection, regulations were revised and a rigorous enforcement campaign was initiated. After March 1996, the incorporation of all animal protein in animal feed was banned. The BSE epidemic in the United Kingdom peaked in 1993 and, as a result of these control measures, now appears to be subsiding.

Recognition of the potential risks to humans

In June 1989, specified bovine offal (SBO) was banned from use in human food. Specified bovine offal includes the brain, spinal cord, spleen, thymus, tonsils, and intestines of cattle. At the time of the ban, some questioned whether all of the spinal cord could be removed during the abattoir process. Questions were also raised about the process of mechanical recovery of scraps left attached to the vertebral column for use in human food (mechanically recovered meat). However, this was not pursued and, in 1995, instances of failure to remove all of the spinal cord from the carcass were discovered. In December 1995, the extraction of mechanically recovered meat from the spinal column of cattle was banned. Mechanically recovered meat can include dorsal root ganglia that have been demonstrated to be infectious in the late stages of incubation.

In May 1990, a domestic cat was diagnosed as suffering from a ‘scrapie-like’ spongiform encephalopathy. This generated widespread public and media concern that BSE had been transmitted to the cat and might also be transmissible to humans. As time passed, the increasing knowledge about BSE made the theory that it would behave like scrapie less and less viable. The public was not informed of any change in the perceived likelihood that BSE might be transmissible to humans and in fact was repeatedly reassured that it was safe to eat beef. There was, nevertheless, some recognition that the pathways by which bovine products or by-products might come into contact with humans or other animals needed to be examined. Known or suspected pathways included meat, vaccines, cosmetics, surgical instruments, bovine or human tissues, agricultural fertilizer, and agricultural waste. However, no coordinated consideration was implemented until March 1996.

The first human cases

Scientists suspected that if BSE were to spread to humans it would resemble Creutzfeldt–Jakob disease. In 1991, surveillance for atypical cases or changing patterns of CJD was put in place. Three dairy farmers who had had BSE in their herds were diagnosed with CJD in August 1992, July 1993, and December 1994. The fourth annual report of the CJD Surveillance Unit (CJDSU), issued in August 1995, noted the apparently high incidence of CJD in farmers. The Spongiform Encephalopathy Advisory Committee (SEAC) released a press release about suspected CJD in a cattle farmer in October 1995.

In May 1995, Stephen Churchill, age 18, died. He was later confirmed as the first known victim of vCJD. His was one of three vCJD deaths in 1995. The CJDSU identified its second suspect case of CJD in a remarkably young patient in August 1995. A third individual died in November 1995. Both cases were later confirmed as vCJD. The CJDSU announced the emergence of vCJD and on March 16, 1996, the SEAC announced that the most likely explanation for the cases of a new variant of CJD in young people was exposure to BSE. This has since been compellingly supported by scientific evidence. A policy of banning consumption of cattle over 30 months of age was introduced. However, the incubation period for transmissible spongiform encephalopathies is long and, not surprisingly, cases of vCJD continue to be diagnosed. Three vCJD deaths were reported in 1995 and 10 deaths in 1996. Ten deaths attributed to vCJD occurred in 1997, 18 in 1998, 15 in 1999, 27 in 2000, 20 in 2001, 17 in 2002, and six during the first three months of 2003. The vast majority of vCJD cases have occurred in the U.K., but six cases were also reported in France and one each in Ireland, Italy, and the United States. Nearly all of these people had lived in the U.K. during multiple years between 1980 and 1996 and had been exposed to BSE there.

BSE outside the United Kingdom

When the BSE epidemic became evident, the European Union Commission prohibited the export from the U.K. of live bovine animals, their semen and embryos, mammalian–derived MBM, or the meat of bovine animals slaughtered in the U.K. that is liable to enter the animal

feed or human food chain. The export of materials destined for use in medicinal products, cosmetics, or pharmaceuticals was also banned. Despite these measures, BSE has spread to countries outside the U.K. Eighteen European countries have reported at least one case of BSE in indigenous cattle. Portugal has the highest incidence rate; in 2001, Portugal reported more than 100 indigenous cases per million cattle aged over 24 months. Significant numbers of cases have also been reported from the Republic of Ireland, with 62 BSE cases per million (cpm), Switzerland (49 cpm), Belgium (28 cpm), Spain (24 cpm), Germany (20 cpm), France (20 cpm), Slovakia (18 cpm), Italy (14 cpm), and the Netherlands (10 cpm). Denmark, Slovenia, Greece, the Czech Republic, Finland, Japan, and Austria have reported between one and seven BSE cases per million. Other countries likely to have BSE include Albania, Bulgaria, Croatia, Cyprus Republic, Estonia, Hungary, Latvia, Lithuania, Luxembourg, Poland, Romania, San Marino Republic, Slovic Republic, and Turkey. Japan reported its first case of BSE in indigenous cattle in 2001 and Israel in 2002. Infections have also been seen in imported cattle in Oman, Liechtenstein, the Falkland Islands, and Canada; however, eradication efforts centered on such imported animals may prevent BSE from becoming established in a country.

BSE and the United States

In 1989, to prevent BSE from entering the United States, restrictions were placed on the importation of live ruminants and certain ruminant products from countries where BSE was known to exist. These restrictions were later extended to include the importation of ruminants and certain ruminant products from all European countries. On August 4, 1997, the Food and Drug Administration (FDA) established regulations that prohibit the feeding of most mammalian proteins to ruminants. Active surveillance efforts by the USDA Animal and Plant Health Inspection Service were instituted in May 1990. As of September 30, 2003, over 57,362 bovine brain specimens from cattle with undiagnosed neurologic disease had been examined by an ongoing BSE surveillance system and no evidence of BSE was found. Regularly updated numbers of bovine brain samples tested as part of the nationwide BSE surveillance program are available at a Web site maintained by the USDA (see below). BSE was detected in one beef cow in Canada in May, 2003. The United States diagnosed BSE in a dairy cow in Washington state in December 2003. This cow had been imported from Canada in September 2001, along with 81 others. Samples were taken from 255 animals located on farms in the U.S. where cows from the index herd in Canada were being raised. All tested negative to BSE.

Sources of Information

USDA APHIS web site

<http://www.aphis.usda.gov/lpa/issues/bse/bse.html>

http://www.aphis.usda.gov/lpa/issues/bse/updates/bse_aphisupdates.html

Statistics from the nationwide BSE surveillance program

<http://www.aphis.usda.gov/oa/bse/bsesurvey.html#charts>

Bovine Spongiform Encephalopathy (BSE) Surveillance, March 2003

<http://www.aphis.usda.gov/lpa/issues/bse/bse-surveillance.html>

CDC web site

<http://www.cdc.gov/ncidod/diseases/cjd/cjd.htm>

Update 2002: Bovine Spongiform Encephalopathy and Variant Creutzfeldt–Jakob Disease

http://www.cdc.gov/ncidod/diseases/cjd/bse_cjd.htm

New Variant CJD: Fact Sheet Centers for Disease Control

http://www.cdc.gov/ncidod/diseases/cjd/cjd_fact_sheet.htm

U.K. DEFRA web site

<http://www.defra.gov.uk/animalh/bse/>

Monthly Creutzfeldt–Jakob Disease Statistics. April 7, 2003.

U.K. Department of Health.

<http://www.doh.gov.U.K./cjd/stats/apr03.htm>

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<http://www.bseinquiry.gov.uk/report/>

(Information from the sections titled “The first cases of BSE,” “Precautions Taken,” and “The First Human Cases” is from this report.)