

Bovine spongiform encephalopathy

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Bovine Spongiform Encephalopathy (BSE), commonly known as **Mad Cow Disease** is a fatal, neurodegenerative disease of [cattle](#), which infects by a mechanism that surprised biologists on its discovery in the late [20th century](#). While never having killed cattle on a scale comparable to other dreaded livestock diseases, such as [foot and mouth](#) and [rinderpest](#), BSE has attracted wide attention because it seems possible to transmit the disease to humans; it is thought to be the cause of [variant Creutzfeldt-Jakob disease](#) (vCJD), sometimes called new variant Creutzfeldt-Jakob disease (nvCJD), a human brain-wasting disease, although this has never been proven.

Infectious agent

Unlike the other kinds of infectious disease which are spread by [microbes](#), the infectious agent in BSE is a specific type of [protein](#). Misshapen ("misfolded") [prion](#) proteins carry the disease between individuals and cause deterioration of the [brain](#). BSE is a type of [transmissible spongiform encephalopathy](#) (TSE). TSEs can arise in animals that carry a rare [mutant](#) prion [allele](#), which expresses prions that contort by themselves into the disease-causing [shape](#). Most TSEs, however, occur sporadically in animals that do not have a prion protein mutation. Transmission can occur when healthy animals consume tainted tissues from others with the disease. In the brain these proteins cause native cellular prion protein to deform into the infectious state which then goes on to deform further prion protein in an exponential cascade. These aggregate to form dense plaque fibers, which lead to the microscopic appearance of "holes" in the brain, degeneration of physical and mental abilities and ultimately death. Some TSE's are resistant to extreme temperatures and are not affected by household disinfectants.

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The BSE epizootic in British cattle

The British BSE [epizootic](#) in cattle was recognized in [1986](#). It was first believed to have originated in [sheep](#), in which the related prion disease [scrapie](#) is common. However, sheep and cattle [TSEs](#) are quite different and it is now thought more likely that BSE could have originated with a case of sporadic BSE in a single bovine. Prior to the BSE epizootic, cattle were fed with [meat and bone meal](#), a high-protein substance obtained from the remnants of butchered animals, including cows and sheep. This practice allowed the accumulation of prions over many generations. As more animals became ill, more infectious tissue got into the feed, and so the number of cases reached epizootic proportions. The tissues that contain most of the pathogenic molecules are those of the

[brain](#) and the [nervous system](#), although infectious amounts have been shown experimentally to be present elsewhere, such as in blood.

The use of meat and bone meal as a protein supplement in cattle feed was widespread in [Europe](#) prior to about [1987](#). Worldwide, [Soybean](#) meal is the primary plant-based protein supplement fed to cattle. However, [soybeans](#) do not grow well in Europe, so cattle raisers throughout Europe turned to the less expensive animal byproduct feeds as an alternative. A change to the rendering process in the early [1980s](#) may have resulted in a large increase of the infectious agents in the cattle feed. A contributing factor seems to have been a change in British laws that allowed a lower temperature sterilization of the protein meal. While other European countries like Germany required said animal byproducts to undergo a high temperature steam boiling process, this requirement had been eased in Britain as a measure to keep prices competitive.

Following an epizootic of BSE in [Britain](#), 157 people (as of [2004](#)) acquired and died of a disease with similar neurological symptoms subsequently called [vCJD](#), or (new) variant Creutzfeldt-Jakob disease. This is a separate disease from 'classical' [Creutzfeldt-Jakob disease](#), which is not related to BSE and has been known about since the early [1900s](#). Of the 157 cases of vCJD in humans so far, 148 occurred in the [United Kingdom](#), 6 in [France](#), and one in Italy. Three cases of vCJD occurred in people who had lived in or visited Britain--one each in Ireland, Canada and the United States. There is also some concern about those who work with (and therefore inhale) cattle [meat and bone meal](#), such as [horticulturists](#), who use it as fertilizer. Up to date statistics on all types of CJD are published by the [UK CJD Surveillance Centre](#) in Edinburgh.

For many of the vCJD patients, direct evidence exists that they had consumed tainted [beef](#), and this is assumed to be the mechanism by which all affected individuals contracted it. Disease incidence also appears to correlate with slaughtering practices that led to the mixture of [nervous system](#) tissue with [hamburger](#) and other beef. It is estimated that 400,000 cattle infected with BSE entered the human food chain in the 1980s. Although the BSE epizootic was eventually brought under control by culling all suspect cattle populations, people are still being diagnosed with vCJD each year (though the number of new cases currently seems to be dropping). This is attributed to the long incubation period for prion diseases, which are typically measured in years or decades. As a result the full extent of the human vCJD outbreak is still not fully known.

The scientific consensus is that infectious BSE prion material is not destroyed through normal cooking procedures, meaning that contaminated beef foodstuffs prepared "well done" may remain infectious.[\[1\]\[2\]](#)

In [2004](#) researchers reported evidence of a second contorted shape of prions in a rare minority of diseased cattle. In other words, this implies a second strain of BSE prion. Very little is known about the shape of disease-causing prions, because their insolubility and tendency to clump thwarts application of the detailed measurement techniques of [structural biology](#). But cruder measures yield a "biochemical signature" by which the newly discovered cattle strain appears different from the familiar one, but similar to the

clumped prions in humans with traditional CJD ([Creutzfeldt-Jakob Disease](#)). The finding of a second strain of BSE prion raises the possibility that transmission of BSE to humans has been underestimated, because some of the individuals diagnosed with spontaneous or "sporadic" CJD may have actually contracted the disease from tainted beef. So far nothing is known about the relative transmissibility of the two disease strains of BSE prion.

In 2005 a controversial paper in [The Lancet](#) suggested that BSE might have originated in British cattle when they ate imported animal feed that included infected human remains from Hindu funeral ceremonies in India. [3]

UK epizootic and UK licensed medicines

During the course of the investigation into the BSE epizootic, an enquiry was also made into the activities of the [Department of Health](#) and its Medicines Control Agency. On [May 7, 1999](#), in his written statement [number 476](#) to the BSE Inquiry, [David Osborne Hagger](#) reported on behalf of the Medicines Control Agency that in a previous enquiry the Agency had been asked to:

"... identify relevant manufacturers and obtain information about the bovine material contained in children's vaccines, the stocks of these vaccines and how long it would take to switch to other products." It was further reported that the: "... use of bovine insulin in a small group of mainly elderly patients was noted and it was recognised that alternative products for this group were not considered satisfactory." A medicines licensing committee report that same year recommended that: "... no licensing action is required at present in regard to products produced from bovine material or using prepared bovine brain in nutrient media and sourced from outside the United Kingdom, the Channel Isles and the Republic of Ireland provided that the country of origin is known to be free of BSE, has competent veterinary advisers and is known to practise good animal husbandry." In [1990](#) the British Diabetic Association became concerned regarding the safety of bovine insulin and the government licensing agency assured them that: "... there was no insulin sourced from cattle in the UK or Ireland and that the situation in other countries was being monitored." In [1991](#) a European Community Commission: "... expressed concerns about the possible transmission of the BSE/scrapie agent to man through use of certain cosmetic treatments." Sources in France reported to the British Medicines Control Agency: "... that there were some licensed surgical sutures derived from French bovine material." Concerns were also raised: "... regarding a possible risk of transmission of the BSE agent in gelatin products."

Husbandry practices in the United States relating to BSE

Soybean meal is cheap and plentiful in the [United States](#). As a result, the use of animal byproduct feeds was never common, as it was in Europe. However, U.S. regulations only partially prohibit the use of animal byproducts in feed. In 1997, regulations prohibited the

feeding of mammalian byproducts to [ruminants](#) such as cows and goats. However, the byproducts of ruminants can still be legally fed to pets or other livestock and poultry such as pigs and chickens. In addition, it is legal for ruminants to be fed byproducts from some of these animals. [4] A proposal to end the use of cow blood, restaurant scraps, and [poultry litter](#) (fecal matter, feathers)^[1] in January 2004 has yet to be implemented [5], despite the efforts of some advocates of such a policy, who cite the fact that cows are herbivores, and that blood and fecal matter could potentially carry BSE.

In [February 2001](#), the [USGAO](#) reported that the [FDA](#), which is responsible for regulating feed, had not adequately policed the various bans. [6] Compliance with the regulations was shown to be extremely poor before the discovery of the Washington cow, but industry representatives report that compliance is now 100%. Even so, critics call the partial prohibitions insufficient. Indeed, US meat producer [Creekstone Farms](#) alleges that the [USDA](#) is preventing BSE testing from being conducted [7].

Japan was the top importer of U.S. beef, buying 240,000 tonnes valued at \$1.4 billion in 2003. After the discovery of the first case of BSE in the U.S. on December 23, 2003, Japan stopped U.S. beef imports in December 2003. In December 2005, Japan allows imports of U.S. beef once again, but reinstated its ban in mid-January 2006 after a technical violation of the U.S.-Japan beef import agreement: a vertebral column, which should have been removed prior to shipment, was included in a shipment of veal.

Tokyo yielded to U.S. pressure to resume imports, ignoring consumer worries about the safety of U.S. beef, said Japanese consumer groups. Michiko Kamiyama from Food Safety Citizen Watch said about this: "The government has put priority on the political schedule between the two countries, not on food safety or human health."

Possibly due to pressure from large agribusiness, the United States has drastically cut back on the number of cows inspected for the BSE (USA Today, August 4, 2006, archived at [8].) Also, the U.S. is prohibiting the sale of test kits that detect BSE.

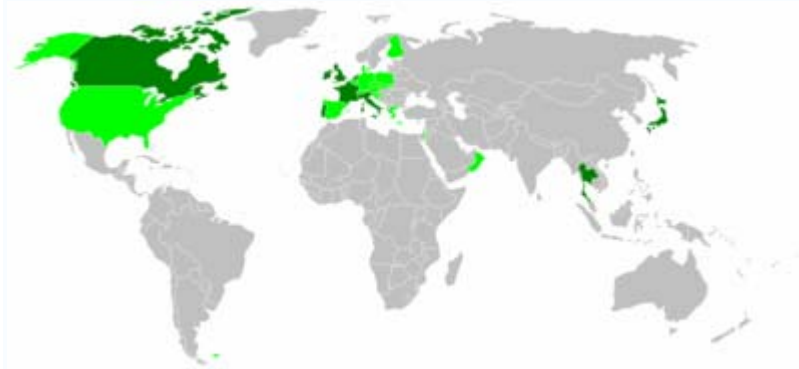
Sixty-five nations have full or partial restrictions on importing U.S. beef products because of concerns that U.S. testing lacks sufficient rigor. As a result, exports of U.S. beef declined from \$3.8 billion in 2003, before the first mad cow was detected in the US, to \$1.4 billion in 2005. (USA Today, August 4, 2006, archived at [9].)

1. [^] The term "chicken litter" also includes spilled chicken feed as well as fecal matter and feathers. It is still legal in the United States to use ruminant protein to feed chickens. Thus, ruminant protein can get into the food chain of cattle in this round about way.

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BSE statistics by country

The following table summarizes reported cases of BSE and of vCJD by country. BSE is the disease in *cattle*, whilst vCJD is the disease in *people*.



Dark green areas are countries that have confirmed human cases of variant Creutzfeldt-Jakob disease and light green are countries that have bovine spongiform encephalopathy cases.

Country	BSE cases	vCJD cases
Austria	2	0
Belgium	125	0
Canada	5 (plus one pending)	1
Croatia	0	0
Czech Rep	9	0
Denmark	13	0
Falkland Is	1	0

Finland	1	0
France	891	11
Germany	312	0
Greece	1	0
Hong Kong	2	0
Ireland	1353	2
Israel	1	0
Italy	117	1
Japan	26	1
Lichtenstein	2	0
Luxembourg	2	1
Netherlands	75	1
Oman	2	0
Poland	21	0

Portugal	875	1
Slovakia	15	0
Slovenia	4	0
Spain	412	0
Sweden	1	0
Switzerland	453	0
Thailand	n/a	2
UK	183803	160
US	3	0
Total	188515	170 (+ 6 results pending)

The figures given above for BSE are certainly too low, and most likely by a considerable amount. The tests used for detecting BSE vary considerably as do the regulations in various jurisdictions for when, and which cattle, must be tested. For instance, in the EU the cattle tested are older (30 months+), while many cattle are slaughtered earlier than that. At the opposite end of the scale, Japan tests all cattle at the time of slaughter. Tests are also difficult as the altered prion protein has very small levels in blood or urine, and no other signal has been found. Newer tests are faster, more sensitive, and cheaper, so it is possible that future figures may be more comprehensive. Even so, currently the only reliable test is examination of tissues during an autopsy.

It is noticeable that there are no cases reported in Australia and New Zealand where cattle are mainly fed outside on grass pasture and, mainly in Australia, non-grass feeding is done only as a final finishing process before the animals are processed for meat.

As for vCJD in humans, autopsy tests are not always done and so those figures too are likely to be too low, but probably by a lesser fraction. In the UK anyone with possible vCJD symptoms must be reported to the UK Creutzfeldt-Jakob Disease Surveillance Unit and so it is unlikely that any cases would be missed. In the U.S., the CDC has refused to impose a national requirement that physicians and hospitals report cases of the disease. Instead, the agency relies on other methods, including death certificates and urging physicians to send suspicious cases to the National Prion Disease Pathology Surveillance Center (NPDPS) at Case Western Reserve University in Cleveland, which is funded by the [CDC](#).