

Bovine Spongiform Encephalopathy (BSE): Risks and Implications for the United States

Introduction

Bovine spongiform encephalopathy (BSE), or mad cow disease, was first recognized in the U.K. in the mid 1980's. The disease is one of several transmissible spongiform encephalopathies (TSE), variants of which affect sheep, elk, deer, mink, and other species. In 1996, U.K. government scientists reported that new variant Creutzfeldt-Jacob Disease (vCJD), a fatal disease in humans, was linked to consumption of beef from animals infected with BSE. As of June 2002, vCJD has been confirmed or suspected in the deaths of 113 individuals in the U.K., six in France, and one each in Italy and Ireland. Given uncertainty about the incubation period of the disease, estimates about the likely number of total victims vary from fewer than a 1,000 to over 50,000 (***)

The 1996 announcement of the link to vCJD resulted in an immediate ban on U.K. exports of live cattle and all beef products, a 30 to 40 percent reduction in domestic beef consumption, and significant losses to beef producers, agribusinesses, and the treasury. A report by the Food Standards Agency estimated the total annual direct costs of BSE controls to U.K. taxpayers at 425 million British pounds (approximately \$650 million). In late 2000, a similar set of events played out in the EU following the announcement of the first native-born cases of the disease in Germany, Spain, and Denmark, and in Japan when it reported its first case in September 2001. The direct costs of BSE control measures to the EU and Japanese treasuries were estimated at 1 billion Euro and 155.4 billion yen (approximately \$1 billion and \$1.3 billion) in 2001.

In this paper we present an overview of the science of BSE and of the events associated with the disease in the U.K., Europe, and Japan. We then review the situation in the U.S. where the disease has not yet been discovered. We describe counter measures already in place, and argue, based on the experience with the disease in other countries, that counter measures should be strengthened. Abbreviations used in the manuscript are listed in Table 1.

Facts about the Disease

Several questions remain unanswered about the cause, original source, incubation periods, and routes of infection of BSE. As to the cause, most but not all scientists believe the infective agent to be a modified prion¹ protein. From a biological perspective prions are simple structures without DNA, which are more difficult to destroy than viruses or bacteria. In fact, no feasible food processing intervention including irradiation destroys prions.

Some experts speculate that gene mutation created the original modified prion responsible for BSE (Phillips *et al.*), while others suspect cross transmission of sheep scrapie (Horn *et al.*). Scrapie has long been known as a disease in sheep and is found worldwide with the exception of Australia and New Zealand. The first recorded case in the U.K. dates from 1732 and there are an estimated five to ten thousand cases there every year. Scrapie could have spread from sheep to cattle through changes in the rendering² system in the 1970's and the introduction of meat and bone meal (MBM) as a protein supplement in rations for young dairy calves (Horn *et al.*).

Regardless of the original source, once it became established in cattle, BSE spread within cattle and then to humans and other species, as tissue from BSE-infected animals entered the animal and human feed chains. It is possible but not yet proven that maternal transmission can occur in cattle, and scientists have not ruled out the possibility of lateral transfer, via contaminated feces, between cohort animals. However, the small number of cases detected in animals born after the UK enhanced its feed ban in 1996 suggests that these routes of transmission do not operate at significant levels.

Most cases occur in animals over four years old, although there are rare instances in which BSE has been found in animals as young as 20 months. The pattern of disease in the U.K. suggests that most animals become infected as calves. During the incubation period infective prions accumulate in nervous tissue—spinal cord, eyes and brain—before symptoms, which involve loss of motor and neurological control, appear. Once symptoms appear, progression of the disease is relatively quick with death occurring within a year. To date, there is no known treatment in either humans or cattle, and the disease can only be definitively diagnosed post-mortem.³

Infective doses for humans are not known with certainty but experiments with cattle and sheep have shown that as little as a half of a gram of infective material is sufficient to transmit BSE when administered orally. Genetic factors are known to play a role in determining both human susceptibility to vCJD and sheep susceptibility to scrapie. Research is ongoing to find corresponding genetic markers in cattle. Thus far transmission to humans has only been associated with consumption of prion-infected nervous tissue from bovines. It is believed unlikely that consumption of muscle tissue or

milk from infected animals can cause the disease, although Bosque *et al.* recently demonstrated the presence of infectious prions in muscular tissue in mice.

BSE Chronology

United Kingdom

The earliest recorded case of what was later recognized to be “mad cow disease” was reported in 1984 on a farm in Sussex in the south of England. Within a year, six more cows from the same farm had succumbed to the illness, but it was not until late in 1986 that the new disease was confirmed as a bovine TSE (see Table 2). As of June 14, 2002 a total of 179,280 cases had been confirmed on over 35,000 farms (Figure 1). For each confirmed case it is likely that several sub-clinical cases were slaughtered. About 80% of cases have been in dairy cows, with over 60% of U.K. dairy farms having at least one case.

Transmission of the new disease was soon linked to MBM, and in July 1988, the U.K. banned the feeding of “animal protein” to ruminants, where animal protein was defined as any protein material derived from the carcass of a ruminant. Because most MBM was used for pigs and poultry the ban was not expected to have significant economic consequences.

Having lost a portion of their domestic market, U.K. renderers turned to exports. Most MBM exported from the U.K. went to EU countries, where it was incorporated into animal rations that were often exported outside of EU. The U.K. was not proactive in warning other countries about the risk from MBM. Publication in the U.K. *Veterinary Record* and a report at the annual meeting of the OIE (Office International des

Epizooties) in May 1989, resulted in several EU countries banning imports of U.K. MBM and the Netherlands imposing a feed ban similar to that of the U.K. It was not until February 1990 that direct correspondence about the issue was made with non-EU countries which had imported MBM from the U.K.⁴

In 1988 an expert group under the chairmanship of Sir Richard Southwood was set up to examine potential human health implications of the new disease. The group recommended incineration of carcasses of infected animals (which was enacted in July 1988), extension of the ruminant feed ban, and destruction of milk from animals suspected of having BSE. They also considered the use of bovine brain and other potentially infective tissues in foods for human consumption, vaccines, and other medicinal products. However, they failed to recommend measures against their use except to avoid the use of bovine offal in baby food.

In June 1989, the U.K. government went beyond the recommendations of the Southwood report by banning the use of specified bovine offal (SBO) for human consumption, thereby removing from the food chain potentially infective tissues from animals not showing symptoms of the disease.⁵ Following discoveries that BSE could be transmitted to mice, domestic cats, and pigs, and on advice from the newly commissioned Spongiform Encephalopathy Advisory Committee, the ban on SBO was extended to all animal feed in September 1990. With both the ruminant feed ban and SBO ban in operation, government officials felt confident that the disease would be contained. The official line was that BSE was not a risk to humans and that it was safe to eat beef. As quoted in the Phillips report (V.6, p4.465), a scientist affiliated with the meat industry

claimed “*a human would have to eat an impossible amount of pure cow brain at the height of infection to reach an equivalent dose to that needed to infect a cow.*”

In March 1991 came the first case of BSE in an animal born after the 1988 feed ban (referred to as a ‘BAB’—born after ban). BABs were initially attributed to delays in the clearing of MBM stocks and while there was some evidence of deliberate breaches of the regulation, officials remained confident that the ban would be effective in eradicating the disease. However, in 1994 there were almost 10,000 BABs, suggesting serious problems with cross-contamination in mills producing feed for both ruminants and non-ruminants, and, by extension, a very high level of infectivity in tissues derived from sub-clinical animals, (since clinical cases were being destroyed). In 1994, officials began finding cases in animals born after the 1990 SBO ban. To date there have been over 12,000 such cases suggesting that in addition to cross-contamination, SBO was not being properly removed.

Figure 2 describes BSE infection pathways and the objectives of measures taken to halt the disease. All three of the numbered pathways must operate for the disease to persist. Destroying clinically infected animals was not sufficient to halt the disease (pathway 1), because sub-clinically infected animals are also infective. The ruminant feed ban sought to block pathway 3, but failed because of cross-contamination. Cross-contamination was (and remains) difficult to detect because of the lack of reliable tests for banned proteins in compound feeds. The SBO ban, which would block pathway 2, failed because of non-compliance. Part of the problem was that the animal SBO ban was

poorly designed and essentially unenforceable with no means of identifying whether or not MBM contained SBO (Phillips *et al.*).

Despite continued reassurance from the government, public concern about the risk to humans had been growing since BSE was first reported. When it was learned in 1990 that the disease had spread to cats, the media began to report that some scientists were concerned about the risk to humans. One scientist, Professor Richard Lacey, was quoted as saying that due to BSE, *“in the years to come our hospitals will be filled with thousands of people going slowly and painfully mad before dying.”* Public concern was heightened in 1993 when it was reported that two dairy farmers had died from CJD. The official position on human health risk remained unchanged with the health minister being quoted in December 1995 as saying *“there is no conceivable risk of BSE being transmitted from cows to people.”*

Early in 1996, officials at the ministry of agriculture were informed that ten people had died of a new strain of CJD. Unlike sporadic CJD which strikes elderly people, all the victims were in their teens or twenties. On March 20, 1996, the government announced that there was a probable link between BSE and this new type of CJD. New control measures included a ban on the use of all MBM of mammalian origin in farm animal feed and the removal of beef from animals over thirty months from the human food chain. This over-thirty-months-scheme remains in operation and has accounted for over 5 million animals. As of June 6, 2002, there have been eighteen as yet unexplained cases of BSE in animals born after the enhanced feed ban was enforced.

Spreading the disease

BSE spread from the U.K. through exports of live animals and contaminated feed. Infected animals exported from the U.K. were detected in Canada, Oman, and the Falkland Islands, in addition to six European countries. The EU banned imports of live animals from the U.K. in 1989, but the ban applied only to animals born prior to the July 1988 feed ban—on the premise that the feed ban would prevent new infections. The import ban was extended the following year to only allow imports of animals aged less than six months. These restrictions were not significant for the U.K. since the majority of its live exports were young calves destined for veal production.

Other countries reacted more vigorously than the EU. Israel banned cattle imports from the U.K. in 1988, followed by Australia, New Zealand, Sweden, and the U.S. in 1989. Of course, not all of the exported cases were detected; many ended up in the animal and human food chain, and likely contributed to the spread of both BSE and potentially vCJD.⁶ However it is likely that exports of contaminated MBM were responsible for the greater part of the spread of BSE beyond the U.K.

Exports of MBM from the U.K. were relatively small during the 1980's, reaching approximately 15,000 tons (4 percent of production) in 1988, most of which went to EU countries, but also to non-EU countries including Indonesia, Thailand and Sri Lanka, and indirectly via animal rations produced in the EU to the Middle East and North Africa. After the U.K.'s ruminant feed ban in 1988, exports to the EU and other countries increased markedly—essentially doubling between 1988 and 1989 (Figure 3). When EU

countries banned U.K. MBM or initiated their own feed bans starting in 1989, MBM exports were redirected to non-EU countries.

As mentioned earlier, the U.K. did not communicate the risk directly to non-EU countries until February 1990, and until September 1991 there was no prohibition on exporting MBM containing SBO to those countries. Given the continuing revision of what constituted SBO, until the mid 1990's MBM was manufactured in the U.K. containing potentially infective material and exported to countries that did not have a ban on the feeding of ruminant protein to ruminants.

BSE in Europe

Following the initial EU ban on live imports from the U.K., restrictions were gradually tightened. In 1990, live exports were limited to animals less than six months old, exports of SBO were prohibited, and U.K. exports of bone-in beef were permitted only from herds free of BSE. Individual European countries banned imports of MBM from the U.K. or adopted ruminant feed bans in 1989 and 1990.

But these measures were taken too late. The first country outside the U.K. to detect BSE was Ireland with ten non-imported cases in 1989. Subsequently, the disease was discovered in Switzerland (two cases in 1990), France (five cases in 1991), and Portugal (twelve cases in 1994). In 1994, the EU banned the use of mammalian protein in ruminant feed and in March 1996, following the announcement of the link to CJD, banned exports of all cattle and beef from the U.K. On January 1, 1998, new legislation required the destruction of Specified Risk Material (SRM) from cattle, sheep, and goats—introducing to other EU countries the restrictions on SBO in place in the U.K. As

had occurred with SBO in the U.K., the definition of what constituted SRM continued to expand. In December 2000, for example, the definition was amended to include intestines of bovines of all ages.

From the mid to late 1990's additional BSE cases were discovered in France, Portugal and Switzerland at relatively low rates,⁷ with cases also appearing in Belgium and the Netherlands in 1997. Then in the fall of 2000, the first "homegrown" cases were discovered in Germany, Spain, and Denmark, followed by Italy's first case in early 2001. The market reaction was instant and dramatic. Beef consumption fell by about 30 percent, and exports by the EU to non-EU countries—notably Russia and Egypt—were halted. German consumers, having been continually reassured by their government that their beef supply was free of BSE, reacted particularly strongly, with beef demand falling by about 50 percent as manufacturers of processed meat products scrambled to reformulate with pork instead of beef. The immediate price effects were so great (see Table 3) that at one point in the subsequent months the trade pattern for beef within the EU was almost completely reversed—with Germany exporting beef to Ireland.

In the wake of the crisis, the EU took a series of measures designed to safeguard public health and restore confidence in beef (see Byrne). These included: (1) BSE tests for all animals over 30 months intended for human consumption.⁸ Animals not tested were eligible for a Purchase-for-Destruction scheme which ran from January 1 to June 30, 2001. (2) All casualty animals over 24 months were tested for BSE. (3) The use of MBM in all animal feed was suspended and all MBM stocks were to be destroyed. (4)

The list of SRM was extended to include vertebral column and the entire intestine, and (5) MRM was banned.

Beyond Europe

The announcement of the first BSE case in Japan came on September 10, 2001. As of June 1, 2002, there had been four confirmed cases, all in dairy cows born in 1996. Initially, it was thought that the outbreak was caused by MBM imported from the U.K. during the early 1990's.⁹ If that were true, the age of the affected animals would suggest that they were second-generation cases—infected from domestic MBM produced from animals infected by the original imports. This recycling scenario would imply that the disease is widespread in Japan, but the slow pace of discovery of new cases suggests otherwise. An alternative explanation involved milk substitute manufactured with beef tallow imported from the Netherlands. However, infectivity has never been detected in tallow. What is now considered the most likely explanation is that the disease was introduced through inadequately sterilized MBM imported from Italy between 1995 and 1998.

Following the announcement, beef consumption fell dramatically. September retail sales were reported to be down 40 to 50 percent, while at regional markets wholesale prices were 30 to 60 percent below normal. According to a national survey by Asahi newspaper in mid-October, one out of four consumers had stopped eating beef altogether, and only 26 percent indicated that they had not altered their beef consumption pattern. Restaurants specializing in beef reported more than 50 percent loss in sales

revenue during the first half of October, while school lunch programs around the nation altered their menus to exclude beef.

In its response to BSE, the Japanese government removed animals over 30 months from the human food chain and instituted BSE testing of all slaughtered cattle for human consumption.¹⁰ It also banned imports, processing, and distribution of MBM for all uses. All livestock waste was to be processed into MBM and incinerated. The total budget of over \$1.3 billion announced by the Ministry of Agriculture, Forestry, and Fishery (MAFF) included an income stabilization scheme for cattle farms, incineration costs of MBM, and the cost of a new electronic tagging system for all livestock. The government also decided to purchase and incinerate the entire beef inventory prior to the initiation of BSE testing at a cost of over 10 billion yen (approximately \$100 million).

The government's handling of BSE has been heavily criticized due to several administrative blunders. For example, MAFF first announced that the infected cow had been incinerated, but three days later acknowledged that it had in fact been processed into MBM. This resulted in an effort to track and incinerate 150 metric tons of MBM, a portion of which had already been transported elsewhere. The requirement to incinerate all MBM also caused problems; several slaughter facilities were forced to temporarily shut down because they had no means to dispose of their waste.

The Situation in the United States

Overview

To date there have been no cases of BSE in the U.S. Other animal TSE's are present including scrapie in sheep, transmissible mink encephalopathy (TME), and

chronic wasting disease (CWD) in deer and elk. Scrapie was first found in the U.S. in 1947 and since then has been reported in over 1000 flocks. The Federal Government has implemented a program designed to eradicate scrapie by 2017. TME was first found in the 1960's in ranched mink and was attributed to the feeding of scrapie-infected sheep tissue (McKenzie *et al.*).

CWD was first identified in the 1960's in captive deer at a research station in Colorado. During the past decade, it has spread to wild deer in several mid-western states and Canada, and to farmed elk. In Wisconsin, testing in 2001 indicated that up to 3 percent of the deer population might have the disease. Much remains unknown about CWD—in particular its modes of transmission and original source. At this time there is no evidence to suggest that CWD can be transmitted to humans. Nevertheless, the World Health Organization (WHO) advises that tissue from deer or elk with CWD should not be used in animal or human food.

To prevent BSE, the U.S. banned imports of live animals from countries with BSE in 1989, imports of MBM from those countries in 1991, and extended those restrictions to countries at risk for BSE (i.e., all of Europe) in 1997. A ban by the Food and Drug Administration (FDA) on the feeding of most ruminant protein to ruminants was implemented in 1997. A recent report by the Harvard Center for Risk Analysis and the Tuskegee University Center for Computational Epidemiology (Cohen *et al.*) has been widely quoted to support the idea that the risk of BSE in the U.S. is extremely low. However, as is stated in the report, its purpose was to evaluate the robustness of measures

to prevent the spread of BSE **if** it were to occur, and it goes on to state that “BSE is extremely unlikely to *become established* in the U.S.” (emphasis added).

The EU Scientific Steering Committee classifies the U.S. as a Geographical BSE-Risk (GBR) level II country—one in which the presence of BSE is ‘unlikely, but not excluded.’¹¹ While U.S. meat exports now carry a label stating, “The United States meets or exceeds all of the international guidelines to be considered free of BSE,” the EU requires that GBR level II countries remove SRM from any beef exported to the EU.

There are several ways BSE might occur in the U.S., one of which is through importation. Currently the U.S. bans imports of live animals and animal feed from all countries at risk for BSE, but the U.S. imported about 1,000 cattle during the past 20 years from countries in which BSE was later found (GAO). These included 334 animals from the U.K. between 1980 and 1989, 443 animals from other EU countries between 1983 and 1997, and 242 animals from Japan between 1993 and 1999. It is likely that material from several hundred of these animals has been used for animal feed and, given the incubation period of the disease, the possibility that some of those animals were infected with BSE cannot be ruled out.

Another possibility is transmission of sheep scrapie or gene mutation within cattle, one of which is the likely origin of BSE in Europe. Moreover, given the prevalence of CWD in the country, cross transmission of CWD from deer to cattle is also a concern, since little is understood about the transmission mechanism for these TSE’s.

Deliberate introduction is also a possibility. Because the BSE agent is stable, infectious in small amounts, apparently harmless unless ingested, relatively accessible

(compared to bio-weapon grade anthrax), and capable of having a significant economic impact, one cannot discount its use as a biological weapon.

Adequacy of U.S. Measures

In February 2002, the General Accounting Office (GAO) responded to a congressional request to evaluate the effectiveness of measures taken to prevent BSE. The GAO concluded that measures adopted in the U.S. did not sufficiently ensure that BSE would be prevented, citing weaknesses in import inspections, inadequate testing of animals that die on farms, and non-compliance with the FDA feed ban. The report also pointed out that the U.S. still allowed cattle brains and other central nervous system tissue to be sold as human food and that consumers were not adequately informed about foods that contained such tissue.

Consider the BSE infection route depicted in Figure 2. In the U.S., the FDA feed ban, adopted in 1997, corresponds to measures to block pathway 3. To compare the FDA feed ban with those adopted elsewhere, we classify bans as follows:

- Level 1 Prohibits feeding *ruminant* protein to *ruminants*
- Level 2 Prohibits feeding *mammalian* protein to *ruminants*
- Level 3 Prohibits feeding *mammalian* protein to *all farm animals (and fish)*
- Level 4 Prohibits feeding *mammalian* protein to *all animals (including pets)*

In this classification, the U.S. feed ban is in fact less restrictive than that described as Level 1 because it permits certain ruminant proteins (milk, blood) to be included in ruminant feed, in addition to allowing protein from non-ruminants such as pigs and horses. The ban adopted in the U.K. in 1988 was at Level 1, and was not fully effective

due to cross contamination between ruminant and non-ruminant feed. In fact, over 43,000 cases of BSE (almost 25 percent of the total to date) have been confirmed in animals born after that ban was implemented. The feed ban adopted by the EU in 1994 was at Level 2. In 1996, the U.K. enhanced its ban to Level 3 by banning the use of MBM for all farm animals and fish. In 2001, the EU adopted a complete ban on the use of MBM; Japan has recently taken a similar measure.

The GAO found weaknesses in FDA enforcement of the feed-ban, particularly in its efforts to re-inspect firms that had not complied with requirements to label feed containing prohibited protein. The GAO also found major weaknesses in the FDA database of inspection records, which led to underreporting of the number of firms not in compliance. Given the extent of cross-contamination problems experienced in the U.K. and the inadequacies in enforcement reported by the GAO, there can be little assurance at present that ruminant feed does not contain prohibited material.

Regarding pathway 2 (Figure 2), the Harvard/Tuskegee report identified additional measures that would reduce the risk from BSE. These included prohibiting the rendering of animals that die on farm, and the implementation of a European type ban on SRM from human and animal feed. A ban on SRM was estimated to reduce the predicted number of BSE cases in an outbreak by 80 percent, and reduce potential human exposure by 95 percent.

For countries at risk for BSE, the WHO recommends removal from the food chain of tissues likely to contain the BSE agent, and as noted above, the EU requires removal of SRM from product exported from the U.S. At present, the U.S. does not have a ban on

human consumption of bovine brain or other central nervous system (CNS) tissue.

According to the GAO, products such as beef stock, beef extract, and beef flavoring can contain CNS tissue, and since 1997, U.S. Department of Agriculture has found CNS tissue in 12 of 63 samples of meat derived from advanced meat recovery (AMR) systems. In its response to the GAO recommendation that consumers be informed about products that might contain CNS, USDA stated that labeling and warning statements should be reserved for known hazards (USDA). Because the U.S. does not have BSE, there is no hazard.

Lastly, effective surveillance is necessary to block pathway 1 (Figure 2). Testing for BSE in the U.S. began in 1990. The surveillance program targets cattle diagnosed with central nervous system disorders at slaughter and “downer” animals, which are those unable to walk at the time of slaughter. As of mid 2001, over 13,000 tests had been conducted. The GAO report noted that very few animals that died on farm, a relatively high-risk population, were included in the testing program. Animals that die on farm may be buried or collected for rendering—unlike the situation in Europe where deaths of all animals over 24 months must be notified, the animal tested for BSE, and the carcass collected for incineration. The Harvard-Tuskegee analysis showed that excluding such animals from the rendering process would significantly reduce the potential for BSE to spread.

In February 2002, USDA announced that it would increase, from 5,000 in 2001 to 12,500 in 2002, the number of cattle brains tested for BSE. However, with between 5 and 6 million cows slaughtered each year, those numbers represent a small fraction of a

higher-risk category of animals. Compared to Europe, where almost 8.5 million animals out of a total adult cattle population of 40.8 million were tested in 2001,¹² the level of testing in the U.S. is miniscule. Of the animals tested in Europe, 770,000 were designated “at risk”—a category including animals that died on-farm or those found sick or injured at slaughter. In the “at risk” category, 760 animals tested positive for BSE, an incidence of about 1 per thousand. If the U.S. had a similar level of incidence among “at risk” animals, 12,500 tests per year in that category provide a 99.999 percent probability of detecting at least one positive case. But with no clinical cases to date, the incidence of sub-clinical BSE in the U.S. herd, if any, must be at a much lower level. If, for example, it were present in U.S. “at risk” animals at the same level as in “healthy” animals in Europe (279 positives in 7.6 million tests, or approximately 1 per 25,000), the current level of testing provides only a 39 percent probability of detection, and it would require almost 120,000 tests to increase the likelihood of detection to 99 percent. In short, the fact that BSE has not been found provides little assurance that it is not present.

It could reasonably be argued that if the disease is present at an extremely low level, the U.S. would be better off not detecting it—especially if there is little chance of it spreading due to controls already in place. However, the infectivity of sub-clinically infected animals (as demonstrated by the failure of the U.K.’s feed ban and SBO controls) is a characteristic of this disease that suggests the potential peril of ignoring its presence—even at an extremely low level.

Implications for the U.S.

It is clear from the experience in Europe and Japan that BSE would have disastrous consequences for the beef industry. With a single confirmed case, exports, which account for about 12 percent of production, would cease and domestic demand would inevitably fall. If the response mirrors that of EU and Japanese consumers, beef consumption would decline by 30 percent, implying that the industry could face an overall reduction in revenue of 40 percent in the short- to intermediate-run.¹³ If demand recovers over a period of four to five years, as happened in the U.K., the cumulative loss to the beef sector would be on the order of \$30 billion.

The response to BSE can be viewed in terms of finding an appropriate balance between costs (to industry) and benefits (to consumers) of actions that mitigate the risk. The problem is that the level of risk is difficult to quantify. If one believes that BSE is inevitable or perhaps already present in U.S. cattle, then a case can be made for taking additional measures to protect human health and eradicate the disease. But if one takes the position that BSE is not here and is extremely unlikely to occur, current measures are more than adequate (see Murphy for an example of this viewpoint). However, the absence of BSE is not so reassuring given the low level of detection efforts. In light of the EU's assessment of the risk level for the U.S., and the discovery of the disease in Germany, Japan, and other countries where it was thought not to exist,¹⁴ a case can be made for both modifying the "official line" on BSE and enhancing preventive measures.

Discovery of BSE in Germany and Japan illustrated two points: i) the fact that BSE has not been detected does not imply its absence, and ii) promoting consumer

confidence in a country's "BSE free" status when there is any risk may be unwise. Official assurances that those countries were free of BSE were unwarranted, contributed to loss of trust in government when BSE was discovered, and likely exacerbated the extent to which consumers moved away from beef. The "official" USDA position at present appears very similar to that taken in Japan and Germany prior to their BSE discoveries. The emphasis is on the fact that the disease has not been detected and the Harvard/Tuskegee report is cited to support the idea that the risk is very small. Because significant economic consequences would result from the discovery of just a single case, it would seem prudent for the U.S. to explicitly acknowledge the possibility that BSE may be discovered, while at the same time enhancing some counter measures.

Particular consideration should be given to measures that reduce potential for human exposure. In the event of a discovery, having such measures in place would provide more credibility to reassurances about the safety of consuming beef. As already noted, for countries that cannot exclude the possibility of BSE, the WHO recommends an SRM ban.¹⁵ If BSE were discovered, the fact that bovine brain and other CNS tissues are not banned for human consumption could be very damaging given the tendency for the media to sensationalize risk issues. Limiting human exposure to bovine CNS tissue might also involve restrictions on AMR.¹⁶ Different AMR systems likely result in different rates of CNS contamination, but until it can be established that one or more systems result in an acceptably low level of contamination, a ban on the use of vertebral column for AMR may be the only way to guarantee absence of CNS in meat.

Other options include extending the feed ban to pigs and poultry. The U.K. experience with cross-contamination suggests that the current U.S. feed ban is inadequate, particularly if enforcement is weak. Eliminating animals that die on-farm from rendering would remove a high-risk source of infectivity, while increasing the level of surveillance testing would provide greater assurance about the continued absence of the disease. Another potential measure is the introduction of animal tracing systems. In the event of BSE being detected, U.S. export markets would be compromised indefinitely because there is no way to trace an animal to its farm of origin and then forward-trace its cohorts or offspring. However, given the substantial costs, it would be difficult to obtain support for a broad range of measures to counter a theoretical risk. Any enhancement in BSE measures at this stage should focus on human health protection in an effort to minimize the market disruption that would result from a discovery.

Acknowledging the possibility of BSE and enhancing measures to safeguard human health should help engineer, in political parlance, a “soft (or at least a softer) landing” if the disease is discovered. It may allow whatever additional measures are taken in response to a discovery to be more proportionate to the risk that is presented. The goal should be to avoid the extreme inefficiency of the Japanese situation in which massive costs are being incurred in response to what now appears to be an extremely low threat. Achieving that goal entails additional up-front costs, but the rate of return on “two stitches in time” is still favorable.

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Table 1. List of Abbreviations

AMR	advanced meat recovery
BAB	born after ban
BSE	bovine spongiform encephalopathy
CJD	Creutzfeldt-Jacob Disease
CNS	central nervous system
CWD	chronic wasting disease
FAO	Food and Agriculture Organization of the United Nations
FDA	Food and Drug Administration
GAO	General Accounting Office
GBR	geographical BSE-risk
MAFF	Ministry of Agriculture, Fishery, and Forestry, Japan
MBM	meat and bone meal
MRM	mechanically recovered meat
OIE	Office International des Epizooties
SBO	specified bovine offal
SRM	specified risk material
TME	transmissible mink encephalopathy
TSE	transmissible spongiform encephalopathy
USDA	U.S. Department of Agriculture
vCJD	variant Creutzfeldt-Jacob Disease
WHO	World Health Organization

Table 2. BSE Chronology

Date	Event*
Dec. 1984	First recorded case of the new disease
June 1987	BSE confirmed
Dec. 1987	Epidemiologists link BSE to MBM
July 1988	U.K. bans <u>ruminant derived</u> MBM in ruminant feed
Nov. 1989	SBO (brain, spleen, spinal cord, thymus, tonsils, intestines) banned <u>for human consumption</u>
Sep. 1990	Ban on use of SBO in <u>all animal feed</u> (including pet food)
Mar. 1991	First BSE case in animal born after the July 1988 feed ban
<i>June 1994*</i>	<i>EU bans feeding of <u>mammalian</u> protein to ruminants</i>
Aug. 1995	SBO definition expanded (entire skull incl. brains, eyes)
Dec. 1995	Ban on use of spinal column in MRM
Mar. 1996	Government announces probable link between BSE and CJD
Aug. 1996	Enhanced MBM feed ban (<u>all farm animals</u>) enforced
<i>Jun. 1997</i>	<i>FDA feed ban introduced</i>
Jan. 1998	Introduction of EU-wide SRM regulations
June 2000	First confirmed case born after the enhanced feed ban
<i>Nov. 2000</i>	<i>Germany, Spain report first cases (Italy in Jan. 2001)</i>
<i>Jan. 2001</i>	<i>EU wide ban on MBM for <u>all farm animals</u></i>
<i>Sep. 2001</i>	<i>First case born outside Europe (Japan)</i>

* *Italics denote events outside the U.K.*

Table 3. Beef Prices in Selected Countries: 2000-01^a

	<u>Aug. 2000</u>	<u>Aug. 2001</u>	<u>Change</u>
Ireland	202	178	-12%
U.K.	224	213	- 5%
France	230	178	-22%
Italy	252	195	-22%
Germany	214	161	-25%
Australia	118	137	+16%
U.S.	204	238	+16%

^a Irish pounds per kilogram of dead weight. Compiled by the authors from various national sources.

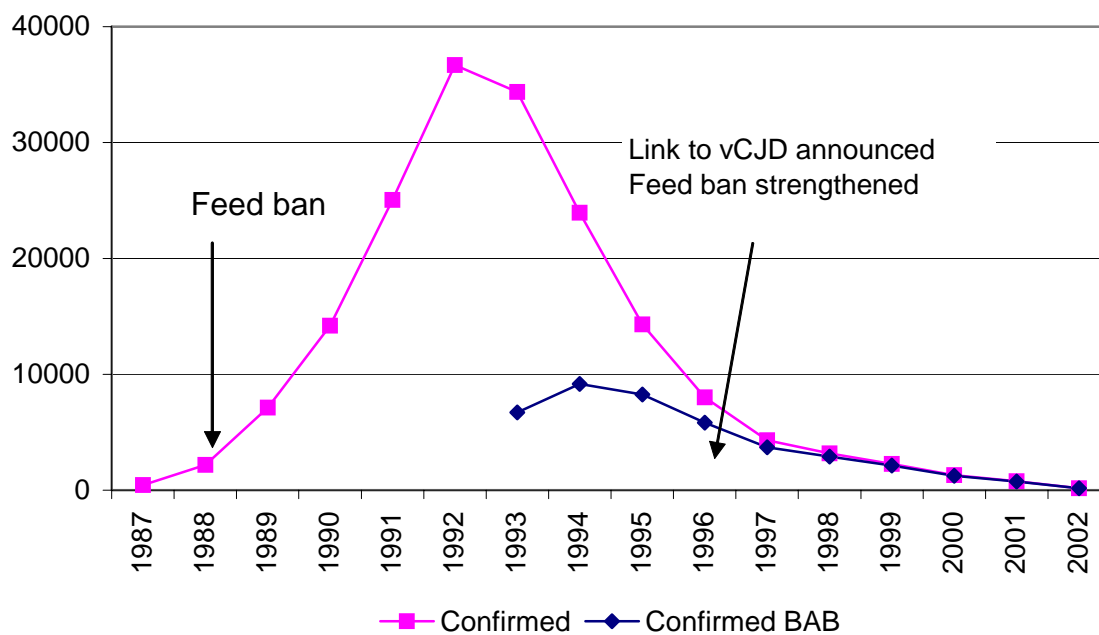


Figure 1. BSE in the U.K. Cases detected by passive surveillance as of May 10, 2002.
Data from U.K. Department for Environment, Food, and Rural Affairs.

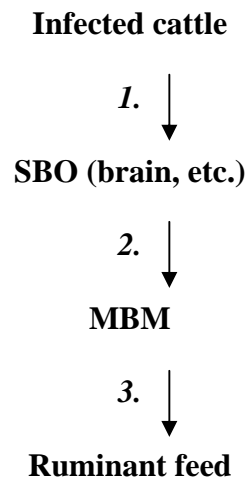


Figure 2. BSE infection route.

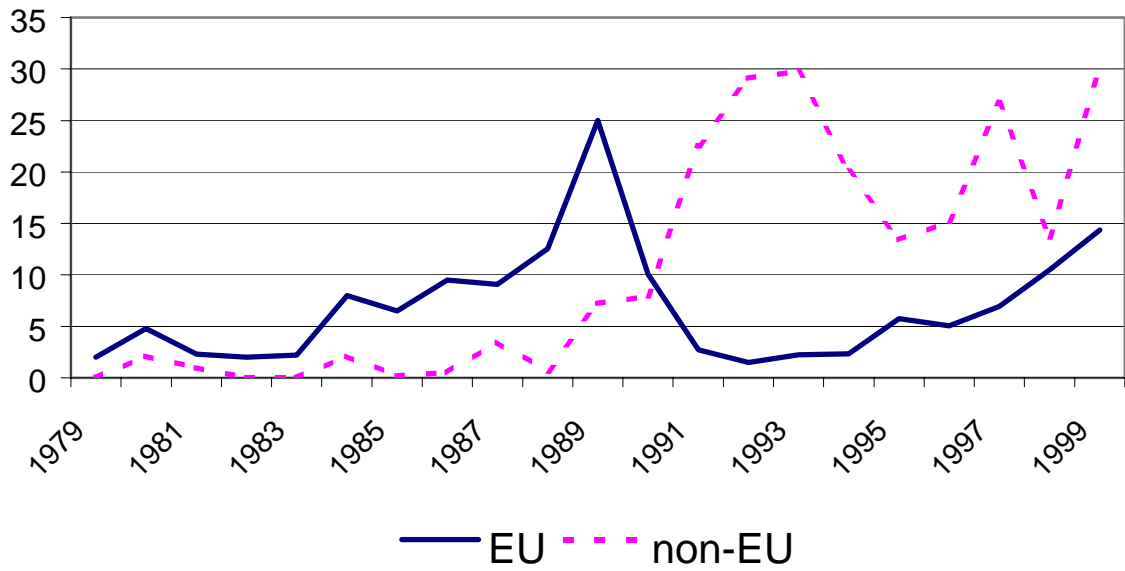


Figure 3. U.K. exports of flours, meats and pellets of meat or meat offal, unfit for human consumption (greaves).

Footnotes

¹ While several strains of scrapie have been identified, thus far only one strain of the BSE prion has been identified in cattle with the disease. The same strain has been identified in other species suffering from BSE type diseases and in humans suffering from vCJD.

² Rendering is the processing of offal (organs, intestines) and other parts of the carcass to produce meat and bone meal, and tallow. It involves drying and cooking, and separating the solids (meal) from the melted liquid fraction (tallow).

³ Recently tests have been developed to detect chronic wasting disease in live deer. Progress has also been reported on tests for TSE's using urine and blood samples.

⁴ The countries were: Norway, Sweden, Switzerland, Czechoslovakia, Hungary, Nigeria, Thailand, South Africa, Malaysia, Taiwan, Hong Kong, South Korea, Japan, Canada, U.S., Turkey, Kenya, Malta, Liberia, Lebanon, Saudi Arabia, Sri Lanka, Puerto Rico, Curaçao and Finland.

⁵ Initially SBO was defined to include brain, spinal cord, tonsils, spleen, thymus and intestines from cattle more than six months old—tissues in which the BSE agent was most likely to be present. The definition was expanded several times in subsequent years. In 1995 the government banned mechanically recovered meat from the spinal column.

⁶ “During 1985-1990 the U.K. exported 57,900 cattle. These animals would have resulted in 1,668 cases of BSE had they remained in Great Britain, but only a small fraction of these cases have been reported by the recipient countries. It has been suggested that only one in six probable cases of BSE within the EC has been reported.” (from Phillips *et al.*). Because importing countries had no bans on human consumption of SBO, these exports constituted a threat to human health.

⁷ In 1999, there were 31 cases in France, 91 in Ireland, 159 in Portugal, and 50 in Switzerland—compared to 2,254 in the U.K.

⁸ In 2001, approximately 8 million animals over the age of 30 months were tested with

approximately 1,000 positive cases discovered.

⁹ Data from COMEXT (an EU trade database) indicate 132, 62, 43, 31, and 64 metric tons of flour, meals and pellets, of meat or meat offal unfit for human consumption were exported from the U.K. to Japan annually between 1990 and 1994.

¹⁰ Japan is the only country that implements comprehensive BSE tests, regardless of age.

¹¹ The U.S. is classified at level II due to imports of live animals from the U.K. and Ireland prior to 1990. In December 2001, three GBR-II countries—Austria, Finland, and Slovenia—detected their first domestic cases of BSE.

¹² Data available at: http://europa.eu.int/comm/food/fs/bse/testing/bse_results_en.html

¹³ Pennings *et al.* conjecture that the response to BSE by U.S. consumers would likely be similar to that of German consumers, since for both groups the consumption decision is driven more by risk attitude than by their perception of the level of risk. For Dutch consumers, who have not responded quite as dramatically to BSE as have Germans, the consumption decision was more closely related to risk perception than to risk attitude.

¹⁴ Israel, which had been one of the first countries to ban imports of MBM and live animals from countries with BSE, discovered its first case in June 2002.

¹⁵ Adoption of the human SBO ban in the U.K. in 1989 is probably the only example in the BSE story of a government going beyond expert opinion in taking a precautionary measure. It turned out to be the correct decision, and likely saved thousands of people from exposure to the disease.

¹⁶ AMR provides around 10 pounds of meat per animal, most of which is used in processed meat products such as hotdogs. The EU banned the process in 2001.