Bovine spongiform encephalopathy (BSE) is an incurable degenerative neurological disorder of cattle that is invariably fatal. It was first identified in 1986 in the United Kingdom and by 1996 it had been associated with a form of the Creutzfeldt-Jakob disease in humans. Though largely concentrated in the United Kingdom, BSE has infected cattle in most of Europe, North America, and Japan, disrupting the international trade of live cattle, beef for human consumption, and beef by-products. BSE has attained global significance by revealing the potential for the intercontinental spread of an infectious animal disease and its implications for risk management, animal health, and human health.

BSE in cattle is associated with a broader class of transmissible spongiform encephalopathies (TSEs) that affect domestic herbivores such as sheep and goats (there it is known as scrapie), domestic carnivores such as house cats and farmed mink, wild and farmed ungulates such as deer and elk (there it is known as chronic wasting disease), and humans in the form of a variant of the Creutzfeldt-Jakob disease (vCJD). While the common form of CJD is a fatal dementing neuropathy of the elderly that is endemic worldwide, vCJD is associated with consumption of tissue contaminated by the active agent that causes BSE. Unlike common CJD, vCJD is distinguished by distinctive clinical and neuropathological features, the biochemical properties of the prion protein, and an average age at mortality which is less than 30. By the end of 2009, a total of 167 deaths in the United Kingdom, 25 in France and 22 in other countries had been attributed to vCJD as the probable or definite cause since 1995.

The agent that causes TSEs is known as a “prion,” a self-reproducing proteinaceous infectious particle. The prion was discovered and named by Dr. Stanley Prusiner in the early 1980s; for this he was awarded the Nobel Prize in Medicine in 1997. Prions were initially associated with scrapie in sheep, and it was not until later that the prion disease family was recognized as TSEs, of which BSE has had the highest profile. Of all the TSEs, BSE has had the greatest economic impact on animal agriculture and the greatest zoonotic effect by infecting humans with vCJD.

BSE in the United Kingdom and Europe

As an animal disease, BSE was first identified in Britain in November 1986, after a dairy cow’s abnormal behavioral symptoms were recorded in December 1984. While there was no evidence that BSE was not transmissible to humans, the UK government assured the public repeatedly and authoritatively that British beef was safe and that BSE was not a danger to human health. The British public was taken by surprise in March 1996, when the government announced that ten cases of vCJD in people under the age of 42 had been confirmed, and the most likely explanation was that those cases were linked to...
exposure to BSE. Seven days later, the European Union prohibited the export of all live cattle and beef products from the UK. In an effort to eradicate the disease, Britain belatedly announced that no cattle over the age of 30 months – those most likely to be infected by BSE – would enter the food or animal feed chains.

In the United Kingdom the incidence of BSE increased rapidly, because meat-and-bone meal derived from infected sheep and cattle was recycled and fed back to cattle, which amplified the outbreak. The epizootic peaked at 37,000 cases in 1992, diminishing gradually to only seven reported cases by 2009. The international spread of the disease resulted initially from British exports of infected live cattle or of animal feed containing infected bovine by-products. For the disease to spread, the practice of feeding protein supplements to cattle, derived from rendered cattle tissues, had to be well established in the importing country. BSE spread first to the Republic of Ireland in 1989 and then to Europe, notably to France, Germany, Belgium, Italy, Spain, and Portugal, the number of reported cases peaking between 1998 and 2002. In 2009 the total number of reported cases worldwide was only 31: 7 in Britain, 22 in Europe, and 1 each in Canada and Japan. Thus the global spread of BSE appears to be under control, and likely to disappear altogether.

**BSE in North America and Globalization**

An embargo on live cattle imports from the UK and a ruminant feed ban in 1997 were thought to be sufficient to keep BSE out of North America. In Canada, the discovery of a single cow infected with BSE on May 20, 2003, from a farm in the Peace River country of northwestern Alberta, heralded an economic catastrophe for Canadian cattle producers. Over 33 countries embargoed Canadian beef products. The potential impact was forecast to translate into a $2-billion loss in gross domestic product (GDP), a $5.7-billion decline in total output, and a loss of 75,000 jobs. BSE made the front pages because of the enormous disruption caused to Canada's cattle markets, livestock producers, and regional agricultural economies. By March of 2010, a total of 17 cases of BSE had been identified in Canada since 2003 and 2 cases had been reported in the United States, one of which was imported from Alberta.

The BSE experience is significant in the annals of globalization for three reasons. It offers the most recent and the most costly evidence of the capacity for intercontinental transmission of animal disease in the absence of regulatory controls over the trade of live animals and animal by-products. The BSE experience in the United Kingdom illustrates the conflict of interest in state agencies that are responsible for promoting domestic agricultural food safety and for protecting public health. And, finally, the BSE experience has become a lightning rod in growing concerns about an agri-food industry that has become global in extent, an example of zoonotic risk at a global scale, a cautionary tale to warn against over-reliance on global food sources, and further justification for a return to a more localized diet.

SEE ALSO: Diseases, Borderless; Livestock and Meat Industries.

**FURTHER READING**
