Situation:

Bovine spongiform encephalopathy (BSE), more commonly known as “mad cow disease,” is a chronic degenerative disease affecting the central nervous system of cattle. BSE first came to the attention of the scientific community in 1986 when cattle in the United Kingdom (UK) were diagnosed with a newly recognized neurological disease. From 1986 until September 1999, over 175,000 head of cattle in more than 34,000 herds were diagnosed with BSE in the UK. In early 1993, as many as 1000 cases/week were being reported. Agricultural officials in the United Kingdom have taken a series of actions to reduce the incidence of BSE in the UK and to minimize the spread of the disease to other parts of the world. Economic repercussions of BSE have included the slaughter of infected and at-risk herds and a 1990 European Union ban on the importation of cattle from Britain.

To date, BSE has not been reported or detected in the United States. However, there is some concern that BSE could find its way into the U.S food supply and become a significant foodborne contaminant. The introduction of BSE would create a significant challenge to the food processing industry. Many forms of conventional food processing techniques are incapable of destroying or inactivating the infectious agent. While the United States Department of Agriculture, Animal Plant and Health Inspection Service (APHIS), suggests that it is extremely unlikely that BSE could become a foodborne hazard in this country, proactive and preventative measures have been taken to ensure the safety of the American public. APHIS is enforcing import restrictions and is conducting surveillance for BSE to ensure that this serious disease does not become established in the U.S.

Little is known about the emerging BSE infectious agent. Current and future research efforts will help scientists better understand where the infectious agent comes from and how it can be controlled.

What we know:

BSE has been attributed to a transmissible infectious agent that is not fully understood. Most scientists classify the BSE agent as a modified form of a normal cell protein, referred to as a “prion.” These prion proteins are smaller than all other infectious agents such as bacteria and viruses.

The disease is called ‘mad cow’ because afflicted animals exhibit unusual behavior. BSE can be fatal within weeks to months after onset of the illness.
The incubation period (the period of time from when the animal first becomes infected until the first signs of disease) ranges from 2-8 years. This makes it very difficult to identify the source of the infection and the possible extent of the spread. Currently, there is no test to detect the disease in the live animal. Veterinary pathologists confirm BSE by postmortem microscopic examination of brain tissue.

BSE has had a substantial impact on the livestock industry in the UK. The disease has also been confirmed in domestic cattle in Belgium, France, Ireland, Liechtenstein, Luxembourg, Portugal, Switzerland, and the Netherlands. However, the infectious agent has NOT been found in the U.S. thus far. The USDA-APHIS has established a surveillance program; and to date, the brain tissue of over 6000 cattle has been examined for the disease. No sign of the disease or the disease agent has been found.

BSE is one of a number of related neurological diseases of animals and humans. These include ‘scrapie’ of sheep and goats and diseases in mink, mule deer, elk, and cats. Related diseases have also been reported in humans. “Kuru” is a severe and fatal human disease found only in New Guinea. The cause was traced to cannibalistic practices involving the consumption of the brains of the recently deceased. Creutzfeldt-Jakob disease (CJD) is another related human disease that occurs worldwide but is very rare. Of significance to the BSE outbreak is a new variant form of CJD in humans. There is strong epidemiologic and laboratory evidence for a causal association between new variant CJD and BSE (source: The Centers for Disease Control and Prevention).

Of most importance to food safety specialists and the food processing community is the ability of the BSE agent to survive unusually harsh environments. Because a prion is extremely small in size, it is more difficult to destroy when compared with larger infectious agents such as bacteria. It is a highly stable agent that can survive high heating temperatures and is resistant to freezing and drying. The agent will survive normal cooking and may also survive thermal processing conditions used for pasteurization and commercial sterility of foods (source: USDA). If the BSE agent does become an agent of foodborne contamination in the U.S., processing conditions must be changed to ensure destruction.

What we don’t know:

Because mad cow disease is a relatively new concern in animal production, there are still many unknowns regarding the possible impact to production agriculture and to the food industry. Of greatest importance is to confirm that mad cow disease is not likely to be transmitted to humans by consuming food. Although there is evidence for a causal association between BSE the human new variant CJD, it is not known how the disease is transmitted. If mad cow disease is identified as an agent transmissible in foods, considerable research efforts will need to be done to identify how we can control the agent during food processing and preparation. Because the onset of illness symptoms occurs years after exposure, it is very difficult to determine the source of the infectious agent and how many animals could be carriers. At present, detection is limited to postmortem microscopic observation and analysis. Research needs to be focused on identifying these types of infectious agents. An ideal detection package would be capable of detecting low levels of the infectious agent in the live animal that is accurate, rapid, and affordable for industry use. Better detection would help to identify infected animals, which would help to determine the source and prevention.

Keep an eye on mad cow disease. Scientists are learning more and more everyday.

For additional food safety information about BSE, call the toll-free USDA Meat and Poultry Hotline at (800) 535-4555 or USDA Food Safety Inspection Service at (202) 205-0293. Much of the information contained in this publication and additional information on the subject can be found at websites for USDA (<http://www.usda.gov/>) and the Centers for Disease Control (<http://www.cdc.gov/>). Specific documents that address mad cow disease can be found at <http://www.cdc.gov/ncidod/diseases/cjd/cjd.htm> and <http://www.aphis.usda.gov/oa/bse/>.