

## **Bovine Spongiform Encephalopathy (Mad Cow Disease)**

### **Background Information and Recommendations**

Bovine spongiform encephalopathy (BSE, or “mad cow disease”) is a progressive neurological disorder of cattle that results from infection by an unconventional transmissible agent. In 1993, the BSE epidemic in the United Kingdom (U.K.) resulted in about 1,000 new cases per week. The outbreak may have resulted from the feeding of scrapie-containing sheep meat-and-bone meal to cattle. There is strong evidence and general agreement that the outbreak was amplified by feeding rendered bovine meat-and-bone meal to young calves.

On December 23, 2003, the U.S. Department of Agriculture (USDA) announced a presumptive diagnosis of BSE in an adult Holstein cow from Washington State. The National Veterinary Services Laboratory, in Ames, Iowa, made the BSE diagnosis. The diagnosis was confirmed by a laboratory in Weybridge, England. Preliminary trace-back based on an ear-tag identification number suggests that the BSE-infected cow was imported into the United States from Canada in August 2001.

Strong evidence indicates that BSE has been transmitted to humans causing a variant form of Creutzfeldt-Jakob disease (vCJD), a rapidly progressive, invariably fatal neurodegenerative disorder. The Centers for Disease Control and Prevention (CDC) monitors the trends and current incidence of CJD in the United States by analyzing death certificate information. The average annual CJD death rate in the United States has remained relatively stable, about one case per million population per year. In addition, CJD deaths in persons aged <30 years in the United States remain extremely rare (<1 case per 100 million per year). In contrast, in the United Kingdom, over half of the patients who died with variant CJD were in this young age group.

The nature of the transmissible agent that causes BSE or vCJD is unknown. Currently, the most accepted theory is that the agent is a modified form of a normal cell surface component known as prion. In the U.K., where over 1 million cattle may have been infected with BSE, a substantial species barrier appears to protect humans from widespread illness. As of December 1, 2003, a total of 153 vCJD cases had been reported worldwide; of these, 143 cases had occurred in the United Kingdom. The risk to human health from BSE in the United States is extremely low.

### **Worker Protection**

Workers involved in the slaughtering, processing, packaging, and preparation of food items that may be from infected livestock should follow all pertinent regulations and guidelines, such as those in FDA’s Food Code <http://www.cfsan.fda.gov/~dms/fc01-toc.html>. Precautions to avoid cuts or punctures that could bring contaminated equipment or product in contact with the blood stream are advisable. In any situations where there is handling of product containing central nervous tissues (brain, spinal cord), where the prions are more concentrated, precautions similar to those taken against blood-borne pathogens may be appropriate.

### **Workplace Cleanup**

The prion protein is not a living organism and is resistant to heat, radiation and disinfectants used in food handling industries. Thorough scrubbing of equipment and work surfaces, particularly of porous surfaces, to avoid cross contamination, should be performed. Again, good guidelines are in FDA's Food Code.

### **Is BSE a foodborne hazard?**

Although there is very strong evidence that the agent responsible for the human disease is the same agent responsible for the BSE outbreaks in cattle, the specific foods that might be associated with the transmission of this agent from cattle to humans are unknown. The current risk for infection with the BSE agent among travelers to Europe is extremely small, if it exists at all. In the U.K. the current risk of acquiring vCJD from eating beef and beef products appears to be about one case per 10 billion servings. According to the CDC, the risk to human health from BSE in the United States is extremely low. To reduce the possible current risk of acquiring vCJD from food, travelers to Europe or other areas with indigenous cases of BSE may wish to consider either: 1) avoiding beef and beef products altogether or, 2) selecting beef or beef products, such as solid pieces of muscle meat (versus calf brains or beef products such as burgers and sausages), which might have a reduced opportunity for contamination with tissues that may harbor the BSE agent. Milk and milk products from cows are not believed to pose any risk for transmitting the BSE agent.

### **Purchase of meat and meat products**

The best protection is to avoid introduction of contaminated food products in the human food chain. Meat in food products to be used at facilities on DOE sites should be purchased only from slaughtering or processing facilities that: 1) meet regulatory sanitation performance standards; 2) have written sanitation standard operating procedures (SSOP); and 3) have Hazard Analysis and Critical Control Point (HACCP) plans.

### **Meat handling and preparation:**

Facilities at DOE sites that handle and prepare food, including meat products, for human consumption should follow the guidelines given in the Food Code <http://www.cfsan.fda.gov/~dms/fc01-toc.html>, which was developed by the Food Safety and Inspection Service of the U.S. Department of Agriculture. It provides practical, science-based guidance and manageable, enforceable provisions for mitigating risk factors known to cause foodborne illness. The Code is a reference document for regulatory agencies that oversee food safety in food service establishments, retail food stores, other food establishments. Inspection and enforcement is handled locally, and DOE should ensure that proper authorities routinely inspect food establishments at DOE sites.

The above information has been abstracted from the CDC homepage at [www.cdc.gov](http://www.cdc.gov)