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Just the Facts...

Health Care Provider Fact Sheet

Bovine Spongiform Encephalopathy (BSE) or "Mad Cow Disease" and variant Creutzfeldt-Jakob Disease (vCJD)

The following information will help you to become familiar with the history, epidemiology and clinical course of Bovine Spongiform Encephalopathy and Variant Creutzfeldt-Jakob Disease, and ways to reduce the risk of exposure. A complete healthcare provider education packet is under development.

WHAT IS BSE?

BSE is a slowly progressing degenerative brain disease of cattle. The disease is fatal for cattle within weeks to months from onset of clinical signs. BSE is one of a family of diseases called Transmissible Spongiform Encephalopathies (TSE) characterized by spongy degeneration of the brain. This group of diseases infects several different species of animals including humans (see Table 1). BSE was first diagnosed in cattle in the United Kingdom (UK) in 1986. Since that time, the disease has been found throughout Europe, in Asia, and it is considered a risk in the following countries: Albania, Andorra, Austria, Belgium, Bosnia-Herzegovina, Bulgaria, Croatia, the Czech Republic, Denmark, the Federal Republic of Yugoslavia, Finland, France, Germany, Greece, Hungary, the Republic of Ireland, Israel, Italy, Japan, Liechtenstein, Luxembourg, the Former Yugoslav Republic of Macedonia, Monaco, the Netherlands, Norway, Oman, Poland, Portugal, Romania, San Marino, The Slovak Republic, Slovenia, Spain, Sweden, Switzerland, and the United Kingdom. There have been no cases of BSE found in the United States and there have been very active surveillance programs since May 1990.

Table 1. Transmissible Spongiform Encephalopathies in Different Species

Species	Disease
Cattle	Bovine Spongiform Encephalopathy (BSE)
Sheep	Scrapie
Elk, Deer	Chronic Wasting Disease
Mink	Transmissible Spongiform Encephalopathy in mink
Cats	Feline Spongiform Encephalopathy
Humans	Creutzfeldt-Jakob Disease (CJD) (classic and variant), Kuru

WHAT CAUSES BSE?

BSE is associated with a transmissible agent. This agent affects the brain and spinal cord of cattle and lesions are characterized by sponge-like changes visible with an ordinary microscope.

The exact nature of the BSE agent is unknown but two theories are being researched. The first is the prion theory, in which the agent is composed largely, if not entirely, of a protein, referred to as a prion. Abnormal forms of a specific protein are found in the brains of infected animals. These abnormally shaped proteins "replicate" by causing the normal animal protein to change shape upon contact. The amyloid plaques found in the brains of infected animals are deposits of these abnormal proteins. The second theory argues that the agent is virus-like and possesses nucleic acids, which carry genetic information. Although strong evidence collected over the past decade supports the prion theory, the ability of the BSE agent to form multiple strains is more easily explained by a virus-like agent.

The agent is highly stable, resistant to freezing temperatures, resistant to drying, and cooking at normal temperatures such as those used in pasteurization and sterilization.

HOW IS BSE TRANSMITTED?

In the cattle industry it has been common practice to use the by-products of rendering, ground and processed into a product called meat and bone meal (MBM). This MBM was then combined with other ingredients and processed into feed for cattle and sheep. Changes in this feed processing in the UK in the 1970s were thought to have contributed to the emergence of BSE.

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Ingestion seems to be the most common form of transmission. There have been cases of transmission inutero from cow to calf. About 1% of calves born to cows with BSE will die of BSE. Injection of tissues from infected animals into the brain of experimental animals 28 February 2001has produced the disease. The World Health Organization has categorized some bovine tissues of infected cattle as having a higher level of infectivity. For classic, sporadic Creutzfeldt-Jakob Disease (CJD) the infectious agent has been transmitted from human to human through grafts of human meningeal tissues, corneal transplants, and natural human growth hormone injections. Transmission has also been shown from intra-cranial surgical instruments. There has been no known transmission from blood transfusion. The theoretical risk of infection with the BSE agent via blood transfusion is under active research at this time.

WHY IS BSE IMPORTANT?

In 1996, a new form of fatal human disease called variant Creutzfeldt-Jakob Disease (vCJD) was diagnosed in a group of people in the UK, and subsequently in France and Ireland. The agent identified as the cause of this new disease appears very similar to the BSE agent. The consumption of meat products from cattle infected with BSE has been implicated as a potential risk factor in the development of vCJD. The incubation period is uncertain, but thought to be from 10 to 20 or more years. There is no curative treatment, and there is no reliable screening test available, although researchers are working to develop one. Diagnosis can only be confirmed during autopsy. At present, there are no cases of vCJD in current/previous service members or family members. Although the risk of adverse health effects is small, the exact mechanism for infection and the effect of repeated exposures to BSE contaminated beef is unknown.

WHAT ARE THE CLINICAL SIGNS AND SYMPTOMS OF vCJD?

Both forms of CJD are marked by the onset of rapidly progressive presenile dementia, myoclonus and progressive motor dysfunction. Classic CJD has a more rapid course, with death usually occurring in the first half-year after symptom onset. Variant CJD differs from classic CJD by having more prominent psychiatric symptoms and signs at onset, a longer course (death usually in the second year after symptom onset) and a lack of characteristic EEG findings often found in classic CJD. Variant CJD also affects a younger age group than the classic form of CJD. Finally, there are some differences in pathologic findings in brain tissue of patients with variant CJD.

WHAT IS THE RISK OF EXPOSURE TO THE AGENT THAT CAUSES BSE/vCJD?

Public health control measures have been recommended by the World Health Organization (WHO) to prevent potentially BSE-infected tissues from entering the human food chain. The most stringent of these control measures have been applied in the UK and appear to be highly effective. In addition, strict bans on the use of cattle protein for cattle feed have been recommended throughout Europe. BSE has not been found in the United States, thus meat procured from the US is considered to be free of BSE. US military dining facilities, commissaries, BX/PX/NEX facilities and MWR activities are directed to sell only beef and beef products procured from US approved sources. According to the Centers for Disease Control and Prevention, the current risk of acquiring vCJD from eating beef (muscle meat) and beef products produced from cattle in Europe appears to be extremely small (perhaps fewer than 1 case per 10 billion servings), if it exists at all. However, to reduce this possible risk, travelers to Europe and other areas where European beef could be used may wish to consider either avoiding such beef and beef products altogether, or selecting solid pieces of beef muscle meat (instead of beef products such as burgers and sausages). Milk, dairy products, poultry, and pork are not believed to pose any risk for transmitting the BSE agent.

For more information, please refer to the Food Safety Fact Sheet or go to:

1. U.S. Army Center for Health Promotion and Preventive Medicine Website.
chppm-www.apgea.army.mil/madcowdisease/

2. Bovine Spongiform Encephalopathy. USDA, Animal Plant Health Inspection Service, APHIS Website.
www.aphis.usda.gov

3. Bovine Spongiform Encephalopathy. USDA Food Safety Inspection Service Website.
www.fsis.usda.gov/OA/topics/bse.htm

4. Emerging and Other Communicable Diseases (EMC) World Health Organization Consultation Report, March 1997.

5. WHO Fact Sheet #180 and #11. WHO Website.
www.who.int

6. Centers for Disease Control and Prevention. National Center for Infectious Diseases, Travelers Health Website.
www.cdc.gov/travel.madcow