FactSheet

Para Su Información

Bovine Spongiform Encephalopathy (BSE)

What is Bovine Spongiform Encephalopathy (BSE)?

BSE (bovine spongiform encephalopathy) is a progressive neurological disorder of cattle, and has been called "mad cow disease." Its symptoms are similar to "scrapie," a brain disease that occurs in sheep. Cattle affected by BSE experience progressive degeneration of the nervous system. Affected animals may display changes in temperament, such as nervousness or aggression, abnormal posture, incoordination and difficulty in rising, decreased milk production, or loss of body weight despite continued appetite. Affected cattle die and currently preventive treatment, including a vaccine, does not exist.

The incubation period (the time from when an animal becomes infected until it first shows disease signs) is from 2 to 8 years. Following the onset of clinical signs, the animal's condition deteriorates until it either dies or is destroyed. This process usually takes from 2 weeks to 6 months. Most cases in Great Britain have occurred in dairy cows between 3 and 6 years of age.

As of November 2003, more than 183,000 cases of BSE were confirmed in the United Kingdom in more than 35,000 herds. The BSE epidemic in the United Kingdom peaked in January 1993 at almost 1,000 cases per week.

What causes BSE?

BSE and scrapie both result from infection with a very unusual infectious agent. However, the exact causative agent of the disease has not yet been completely characterized. Three main theories on the nature of the agent have been proposed: an unconventional virus; a prion or abnormal partially-proteinase K-resistant protein, devoid of nucleic acid, capable of causing a cell to produce more abnormal protein; or a virino or "incomplete" virus composed of naked nucleic acid protected by host proteins.

Currently, the most accepted theory is that the agent is a modified form of a normal cell protein known as a prion. A prion is not a bacterium, parasite, or virus, and thus treatments usually used for treating or prevention bacterial infections (e.g. antibiotics) or viral infections are not effective against prions.

Currently, it is accepted that the BSE agent (1) is smaller than most viral particles and is highly resistant to heat, ultraviolet light, ionizing radiation, and common disinfectants that normally inactivate viruses or bacteria; (2) causes no detectable immune or inflammatory response in the host; and (3) has not been observed microscopically.

The outbreak of BSE in the United Kingdom may have resulted from the feeding of scrapie-infected sheep meat-and-bone meal prepared for cattle to young calves.

Does BSE or a similar disease occur in humans?

BSE belongs to a group of progressive degenerative neurological diseases known as transmissible spongiform encephalopathies (TSEs), which are always fatal. The TSE diseases include scrapie, which affects sheep and goats; transmissible mink encephalopathy; feline (cat) spongiform encephalopathy; and chromic wasting disease of deer and elk. There are six TSE diseases that affect people: kuru, classical Creutzfeldt-Jakob disease

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(CJD) and variant Creutzfeldt-Jakob disease (vCJD), Gerstmmann-Straussler-Scheinker syndrome, fatal familial insomnia, and sporadic fatal insomnia. The human diseases are very rare; for example, classical CJD has been well studied and occurs sporadically worldwide at a rate of about one case per one million people.

Where is the BSE agent found?

In cattle naturally infected with BSE, the BSE agent has been found in brain tissue, in the spinal cord, and in the retina of the eye. Additional experimental studies suggest that the BSE agent may also be present in the small intestine, bone marrow, and dorsal root ganglia.

How was BSE spread or transmitted?

It is thought that BSE was spread via meat-and-bone meal fed to cattle. The practice of using this material as a source of protein in cattle feed has been common for several decades. In the late 1970s there was a change in the production (rendering) process used to make this meat-and-bone meal. One hypothesis is that this change permitted the infectious agent of scrapie (a transmissible spongiform encephalopathy, or TSE, of sheep)to survive the rendering process, and get transmitted to other animals, such as cows, that are fed meat-and-bone meal nutritional supplements.

Does BSE occur in the United States?

On Dec. 23, 2003, the U.S. Department of Agriculture (USDA) announced a presumptive diagnosis of BSE in an adult Holstein cow from Washington state. USDA has launched an epidemiologic investigation to determine the source of the disease. For up-to-date information access the USDA website at www.usda.gov.

Is BSE a foodborne hazard in the United States?

Strong evidence indicates that BSE has been transmitted to humans primarily in the United Kingdom, causing a variant form of Creutzfeldt-Jakob disease (vCJD). In the United Kingdom, where over 1 million cattle may have been infected with BSE, a substantial species barrier appears to protect humans from widespread illness. As of Dec. 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.

What countries have reported BSE?

The vast majority of cases of BSE (more than 99% as of 1999) have been reported from the United Kingdom during an epidemic. However, endemic cases have also been reported in other European countries including: the Republic of Ireland, Switzerland, France, Liechtenstein, Luxembourg, Netherlands, Portugal and Denmark. The numbers of reported cases by country are available on the web site of the Office International des Epizooties (http://www.oie.int/). These numbers should be interpreted with caution, however, because the intensity and methods of surveillance probably vary over time and by country.

How is the BSE agent detected?

The presence of the BSE agent in tissues is generally determined by injecting animals, usually mice, with material believed to be infected with BSE, then observing the mice to see if they die and have characteristic brain tissue changes. Mouse inoculation studies take a long time (up to 700 days) to detect the agent, and a negative result (that is, lack of brain tissue changes in the injected mice) may only mean that there was too little of the infectious agent to cause symptoms, not that the material was really free of the infectious agent altogether. It is also possible to detect the presence of the abnormal prion protein in tissue (such as brain) using special staining procedures, although these methods do not allow an accurate assessment of infectivity of the infected material.

What has the British government done in response to the BSE epidemic?

In response to the BSE epidemic, the British Government instituted a series of measures to minimize the risk of disease transmission among both animals and humans. These included a ban on feeding ruminant protein (ruminants are animals, such as cows, sheep and goats) to ruminants (1988), removal of some “high risk” materials (such as brain, spinal cord and intestines) from cattle at slaughter

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(1989 and 1995), and a ban on cattle over 30 months of age from being used for food (1996). Following institution of these measures, Great Britain has seen a decrease in the number of cattle with BSE from a peak incidence of 36,680 confirmed cases in 1992 to 2,254 confirmed in 1999 (information on the BSE epidemic in Great Britain is available at http://www.defra.gov.uk/).

Where can I get more information?

Access the Centers for Disease Control and Prevention website at www.cdc.gov, or the USDA website at www.usda.gov. Contact your doctor or the Southern Nevada Health District, Office of Epidemiology at (702) 759-1300.

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Updated 8-09