

Bovine Spongiform Encephalopathy

“Mad Cow Disease”

Questions and Answers

Are BSE and “mad cow disease” the same thing?

Yes. BSE stands for bovine spongiform encephalopathy, and it is widely referred to as “mad cow disease.” It is a chronic degenerative disease that affects the central nervous system of cattle. BSE is named because of the spongy appearance of the brain tissue of infected cattle examined under a microscope.

Is BSE related to any other diseases?

BSE belongs to a family of diseases known as the transmissible spongiform encephalopathies (TSEs). TSE animal diseases found in the United States include scrapie in sheep and goats, chronic wasting disease in deer and elk, transmissible spongiform encephalopathy in mink, feline spongiform encephalopathy in cats, and in humans: kuru, both classic and variant Creutzfeldt-Jakob disease, Gerstmann-Straussler-Scheinker syndrome, and fatal familial insomnia.

(Note: The one case of variant Creutzfeldt-Jakob disease in the United States is in a young woman who likely contracted the disease while living in the United Kingdom. Symptoms appeared after she moved to the United States. The Centers for Disease Control and Prevention has not found additional cases in the United States through its surveillance program.)

What causes BSEs and other TSEs?

The agent that is responsible for BSE and other TSEs has not been fully characterized. Although other types of agents have been implicated, the theory that is most accepted in the scientific community is that the agent is a prion, which is an abnormal form of a normal protein known as a cellular

prion protein. The TSE agents are extremely resistant to heat, ultraviolet light, ionizing radiation, normal sterilization processes, and common disinfectants that normally inactivate viruses and bacteria.

What about the possibility of BSE coming from other TSEs already in the United States, such as deer and elk with chronic wasting disease, or other sources such as the practice of feeding cattle parts to pigs?

As mentioned before, there are several TSEs in the United States. However, there is no evidence to date that BSE has emanated from TSEs in other animals.

Regarding feeding practices, it is known that cattle can become infected with BSE by eating feed contaminated with the infectious BSE agent. This is why in 1997 the U.S. Food and Drug Administration (FDA) prohibited the use of most mammalian protein in the manufacture of animal feed intended for cattle and other ruminants. For additional information on the feed ban, visit FDA's Center for Veterinary Medicine Web site at <http://www.fda.gov/cvm/>.

Where is the BSE agent found in cattle?

Current scientific research confirms that BSE infectivity occurs in the brain, trigeminal ganglia, tonsils, spinal cord, dorsal root ganglion, and distal ileum of the small intestine of cattle experimentally infected with the BSE agent. Research also confirms that BSE infectivity is in the brain, spinal cord, and retina of the eyes of cattle infected with the agent under field conditions. Although bone marrow has demonstrated infectivity in experimentally infected cattle, these findings are not conclusive.

Can BSE be transmitted from one cow to another cow?

No. BSE is not a contagious disease. There is no evidence that the disease is transmitted through direct contact or animal-to-animal spread. The

primary means by which animals become infected is through consumption of feed contaminated with the infectious BSE agent.

What is FSIS doing to protect the public from BSE?

While FSIS believes that the food supply is safe, the Agency has taken a number of steps to ensure that the public does not receive product that could have the BSE infectious agent – however remote that risk is to begin with. On December 23, 2003, after the discovery of a presumptive positive of BSE found in a Holstein dairy cow slaughtered at an establishment in Moses Lake, Washington (see recall release [FSIS-RC-067-2003](#)), FSIS immediately issued a press release that announced the firm's voluntary recall of 10,410 pounds of raw beef. This product might have been exposed to tissues containing the infectious agent that causes BSE. The recall was made out of an abundance of caution, since muscle meat does not contain the high risk neural tissues such as brain and spinal cord, and is considered safe.

In addition, on December 30, 2003, Agriculture Secretary Ann Veneman announced new policies that would further strengthen an existing solid food safety system against BSE. On that date, an immediate ban was enacted to prevent all non-ambulatory disabled cattle from being used in the human food supply. This group contains the highest risk population of cattle that could possibly have BSE. However, even before this ban, FSIS inspectors at slaughterhouses were condemning all cattle they suspected of showing central nervous system disorders.

The four policies that Secretary Veneman announced on December 30, 2003 were made effective by FSIS on January 12, 2004. These included:

- **Product Holding** – FSIS inspectors no longer mark cattle tested for BSE as “inspected and passed” until confirmation is received by FSIS and the plant that the cattle have, in fact, tested negative for BSE.
- **Specified Risk Material** – FSIS declared that skull, brain, trigeminal ganglia, eyes, vertebral column, spinal cord and dorsal root ganglia of cattle 30 months of age or older and the small intestine of all cattle are

specified risk materials that are prohibited in the human food supply. Tonsils from all cattle are also not allowed in the human food supply.

- **Advanced Meat Recovery** – FSIS expanded a prior prohibition on spinal cord from being allowed in product produced from a technology called advanced meat recovery (AMR). This new regulation prohibits dorsal root ganglia, clusters of nerve cells connected to the spinal cord along the vertebral column, in addition to spinal cord tissue from being in AMR product.
- **Air-Injection Stunning** – FSIS banned the practice of air-injection stunning to ensure that portions of the brain are not dislocated into the tissues of the carcass as a consequence of humanely stunning cattle during the slaughter process.

FSIS has implemented these measures as further safeguards in an existing strong food safety infrastructure to protect public health. For more information about these regulations, visit FSIS' Web page at: <http://www.fsis.usda.gov/oa/news/2004/bseregs.htm>.

Is there a BSE test for meat?

No. The only USDA approved testing for the agent is post-mortem analyses of brain tissue. This is a laboratory screening test for BSE.

How does one test for BSE?

Currently, there is no test to detect the disease in a live animal or in muscle meat. Veterinary pathologists confirm BSE by postmortem microscopic examination of brain tissue using sophisticated laboratory techniques, such as a histopathological examination to detect sponge-like changes in the brain tissue and immunohistochemistry to examine the BSE fibrils. These are “gold-standard” tests, and they take more than a week to run.

More rapid tests that provide results within 36 to 48 hours have been developed to detect the abnormal prion in brain or spinal cord tissue of dead animals. Rapid tests can be used to determine if BSE exists in a population and to obtain an indication of its prevalence or detect animals with the disease which are not yet showing clinical signs.

What are the clinical signs that cattle have BSE?

Cattle affected by BSE experience progressive degeneration of the nervous system. Affected animals might 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.

Is there any cure for BSE?

No. There is no treatment for BSE. The course of the disease varies from two weeks to 14 months, usually resulting in death or humane destruction within four months in countries where the disease is present.

How long can BSE be in an animal before it shows signs of the disease?

The incubation period (the time from when an animal becomes infected until it first shows disease signs) is from 30 months to eight years with only a few rare exceptions in younger animals. Following the onset of clinical signs, the animal's condition deteriorates rapidly. This process usually takes from two weeks to six months. Most cases in Great Britain occurred in dairy cows between three and six years of age.

Are humans susceptible to BSE? *

Although not scientifically proven, there is strong epidemiologic and laboratory data linking a rare, degenerative, fatal brain disorder in humans

called variant Creutzfeldt-Jakob Disease (vCJD) to the consumption of BSE-contaminated product. This type of disease begins primarily with psychiatric symptoms and affects younger patients (median age, 28 years).

How many cases of vCJD have there been and have there been any in the United States? *

As of December 1, 2003, a total of 153 cases of vCJD had been reported in the world: 143 from the United Kingdom, six from France, and one each from Canada, Ireland, Italy, and the United States.

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How is variant Creutzfeldt-Jakob Disease different from classic Creutzfeldt-Jakob Disease? *

The classic form of Creutzfeldt-Jakob Disease is endemic throughout the world, including the United States. The median age at death of patients with classic CJD in the United States is 68 years, and very few cases occur in persons under 30 years of age. In contrast, the median age at death of patients with vCJD is 28 years.

The vCJD can be confirmed only through examination of brain tissue obtained by biopsy or at autopsy, but a “probable case” of vCJD can be diagnosed on the basis of certain clinical criteria developed in the United Kingdom. The incubation period for vCJD is unknown because it is a relatively new disease. However, it is likely that ultimately this incubation period will be measured in terms of many years or decades. In other words, if a person develops vCJD from consuming a BSE-contaminated product (not yet scientifically proven), he or she likely would have consumed that product a decade or more earlier.

In contrast to classic CJD, vCJD predominantly affects younger people, has atypical clinical features, with prominent psychiatric or sensory symptoms at the time of clinical presentation. There are delayed onset of neurological abnormalities, including ataxia within weeks or months, dementia and myoclonus late in the illness. Typically, the duration of illness is at least six months.

Can BSE be transmitted to milk and other dairy products?

There is no scientific evidence to suggest that milk and dairy products carry the agent that causes BSE.

What do I do if I ate recalled meat associated with BSE?

The recalled meat (class II from December 23, 2003) is considered safe by USDA, as the tissues that would carry the BSE agent were completely removed at slaughter and not used in meat cuts or products that might have been consumed by humans. The recall from December 23, 2003 was made out of an abundance of caution. If you have concerns that you might have contracted a foodborne illness, then you should contact your health care provider.

Will cooking (including microwave cooking) kill the BSE agent?

Current scientific research indicates that cooking will not kill the BSE agent.

Will irradiation kill the BSE agent?

Current scientific research indicates that irradiation will not kill the BSE agent.

Are baby foods safe?

Beef products processed by mechanical separation may not be used in the formulation or production of baby, junior, or toddler foods.

Advanced meat recovery (AMR) products, which are processed by removing muscle tissue without breaking bones and do not include spinal cord tissue, is allowable for these products (However, there are further prohibitions of material allowed in AMR. See FSIS' [rule](#) which became effective January 12, 2004).

Are meats used in the National School Lunch Program safe?

Yes. USDA's Agricultural Marketing Service (AMS), by specification, does not allow beef that is mechanically separated from bone with automatic deboning systems, advanced lean (meat) recovery (AMR) systems, or powered knives for any commodity programs. USDA procurement specifications for beef specifically prohibit the use of meat from downer animals – animals too sick or injured to walk.

For questions concerning animal feed for livestock or pets, contact the U.S. Health and Human Service's Food and Drug Administration (FDA).

Center for Veterinary Medicine
7519 Standish Place
Rockville Maryland 20855-0001
(301) 827-3800 or 1-888-INFO-FDA
<http://www.fda.gov/cvm/>

For questions concerning animal health, surveillance, and BSE, contact the U.S. Department of Agriculture's Animal and Plant Health Inspection Service (APHIS).

USDA-APHIS
4700 River Road
Riverdale, MD 20737
(301) 734-7799
<http://www.aphis.usda.gov/>

*** For questions concerning vCJD or CJD, or any of the specific human diseases and technical terms mentioned on this Web page, contact the U.S. Health and Human Service's Centers for Disease Control and Prevention (CDC).**

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1600 Clifton Road
Atlanta, GA 30333
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