AABP FACT SHEET

Facts about Bovine Spongiform Encephalopathy

Report from the
American Association of Bovine Practitioners
Food Quality, Safety and Security Committee
for distribution to clients and the general public

While "mad cow disease" has widespread name recognition, considerable misinformation continues to circulate. The following questions and answers are intended to give you a basic understanding of the disease, its control and how to protect public health.

What is Bovine Spongiform Encephalopathy (BSE)?

The disease commonly referred to as "Mad Cow Disease" is known in veterinary circles as Bovine Spongiform Encephalopathy (BSE). BSE is a progressive, fatal disease of the nervous system of cattle. The disease was first reported in the United Kingdom in 1986 and has been reported in Europe, Asia, the Middle East, and North America. There is no treatment or vaccination for this disease.

What causes BSE?

Scientists have isolated a very small abnormally shaped protein from BSE-affected cows called a prion. The BSE prion can infect other cattle if it is injected or fed to them. The BSE prion accumulates in brain and spinal tissue of affected cattle, interfering with normal brain function. Prions are different from viruses and bacteria. They are much more resistant to destruction than most pathogens and not all rendering and sterilization processes will adequately destroy this protein.

What are the signs of BSE?

Cattle with BSE show clinical signs including nervousness, increased startle response to stimuli (like noises or contact), depression, severe muscle tremors, unusual gait or stance, reduced rumination, altered heart rhythm and weight loss. Unfortunately, the signs of BSE are similar to those seen in many other diseases that affect the nervous system of cattle, for example: Grass Tetany, Nervous Ketosis, Listeriosis (circling disease), Polio and Rabies. Cattle infected with BSE appear healthy for several years (2-8) before showing any signs, as the brain destruction occurs very slowly at first. Once signs begin to occur, the disease progresses rapidly and the cow will generally die within six months.

Is there a test for BSE?

Yes, but all of the currently available tests use samples that can only be collected from cattle after death. The brain can be examined under a microscope to find characteristic lesions, essentially holes in the brain tissue which give it a 'sponge-like' appearance, or brain tissue can be tested for the abnormal protein. No test is currently available to check live cattle for BSE.

Is BSE unique?

Yes and No.

Yes, BSE is a new disease of cattle discovered a little over 20 years ago. Unfortunately, examination of the brain tissue from infected animals shows that BSE is indistinguishable from Feline Spongiform Encephalopathy, and variant Creutzfeldt-Jakob Disease (vCJD), suggesting that the source of Feline Spongiform Encephalopathy, and vCJD apparently was BSE contaminated feed/food.

No, BSE is not the only brain-destroying disease like this. BSE belongs to a group of diseases known as Transmissible Spongiform Encephalopathies (TSE). In animals, these diseases also include Scrapie in sheep and goats, Chronic Wasting Disease (CWD) in elk and deer, Transmissible Mink Encephalopathy in mink, and Feline Spongiform Encephalopathy in both wild and domestic cats (including large cats like lions). Both exotic (what we would see in a zoo) and domestic ruminants have been diagnosed with BSE, all associated with consumption of contaminated feed. There are a number of TSE diseases in humans as well: Kuru, Gerstmann-Straussler-Scheinker Syndrome, and Creutzfeldt - Jakob disease (CJD).

How is BSE transmitted?

The primary source of BSE infection in cattle is feed contaminated with the infectious agent. Scientific evidence shows that feed contamination results from incorporating ingredients (for example, meat-and-bone meal) that contain rendered protein derived from infected cattle. Standard rendering processes do not completely inactivate or kill the BSE agent. Therefore, rendered protein such as meat-and-bone meal derived from infected animals may contain the infectious agent and transmit the disease. BSE is not a contagious disease and therefore is not spread through casual contact between cattle.

The widespread use of ruminant-derived protein, including brain and spinal tissue, in cattle diets recycled the infection. Since less than one gram of infected material is necessary to cause infection, the disease was rapidly amplified in the cattle population by this feeding practice. Offspring of BSE infected cattle are more likely to develop BSE; however, it is not known why this occurs.

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Is BSE in North America?

As of November 2009 there have been three cases of BSE in the United States, one from a cow that originated in Canada and two from cattle of US origin. Within the same time frame Canada has discovered one cow with BSE that was imported from the United Kingdom and 16 cases of BSE in cattle of Canadian origin. All the US cases were in cattle that were born prior to the US ban of ruminant derived protein feeding to other ruminants. Eleven of the Canadian cases were in cattle that were born after the ruminant derived protein feeding ban implemented by Canada.

What are we doing to prevent BSE in our cattle population?

The key to preventing BSE is to stop imports of potentially infected cattle and feed, and the implementation of bans on the use of rendered animal protein in ruminant feeds. The USDA and the FDA are continually reviewing management practices that may place the US cattle population at risk. As research shows that something is a potential risk, regulatory measures are enacted to prevent the risk. Similar restrictions have been implemented by the Canadian Government and the Canadian Food Inspection Service.

BSE concentrates in certain tissues in infected cattle known as specified risk material (SRM). To prevent BSE spread among cattle, the United States and Canada banned the feeding of ruminant-derived protein to ruminants, including SRM, in 1997. To provide further animal health protection, as of July 12, 2007, Canada has banned the use of SRM from all other animal feeds, pet foods and fertilizers. In 2009, the US enacted a similar rule banning most SRM in animal feed.

Although the United States and Canada previously did allow the feeding of ruminant-derived protein to ruminants, the risk of BSE exposure has always been lower than in the United Kingdom because cattle feeding practices have been quite different between the two regions. In United Kingdom, ruminant-derived protein served as an inexpensive protein source for calves and cattle. Here in North America, we have many more options for protein supplementation because of a large supply of relatively inexpensive, high-protein plant sources, such as cottonseed meal or soybean meal.

What is the current situation in Europe?

The outbreak of BSE in the United Kingdom peaked in 1992-93 at approximately 1000 new cases per week. The rate of new cases has been declining rapidly since that time with only 67 cases in 2007, 37 in 2008, and only 7 in 2009 (through September 30). The numbers of cases in other European countries have similarly declined in recent years. While BSE has been found in a number of other countries, approximately 95% of the all cases diagnosed to date have been found in the United Kingdom.

The European Union has implemented control measures to halt the spread of BSE. The most stringent control measures include a UK program that excluded all animals more than 30 months of age from the human food and animal feed supplies. Due to the continued decline in cases (and the increasing age of those cases that were found), this restriction was altered in 2006 and ultimately ended in 2008. In June 2000, the European Union Commission on Food Safety and Animal Welfare strengthened the European Union's BSE control measures by requiring all member states to remove specified risk materials from animal feed and human food chains as of October 1, 2000; such bans had already been instituted in most member states. Other control measures include banning the use of mechanically recovered meat from the vertebral column of cattle, sheep, and goats for human food and BSE testing of all cattle more than 30 months of age destined for human consumption.

What about "the human form of BSE"?

There is now strong scientific evidence that the agent responsible for the outbreak of prion disease in cows, BSE, is the same agent responsible for the outbreak of Variant Creutzfeldt-Jakob Disease (vCJD) in humans. Both disorders are invariably fatal brain diseases with unusually long incubation periods measured in years, and are caused by an unconventional transmissible agent. While the signs of vCJD are similar to classical CJD, a rare disease found around the world, the diseases differ in many distinct ways. In vCJD, the age of onset is much younger, the duration of the disease (once signs are apparent) is longer, and the microscopic appearance of the brain is different. Tests show that proteins present in the brain tissue of affected individuals are different between cases of CJD and vCJD.

Since variant CJD was first reported in 1996, a total of 217 patients with this disease from 11 countries have been identified. As of November 2009, variant CJD cases have been reported from the following countries: 170 from the United Kingdom, 25 from France, 5 from Spain, 4 from Ireland, 3 from the United States, 3 in the Netherlands, 2 each in Italy and Portugal, and one each from Canada, Japan, and Saudi Arabia. Two of the three U.S. cases and the single Canadian case all had ties to the United Kingdom, while the third US case had ties to Saudi Arabia.

No one knows yet how many people will be affected with vCJD in the future; however, the annual number of cases in the United Kingdom has decreased over the past few years. It is important to note that vCJD has not been associated with the consumption of lamb or mutton products. If Scrapie were transmissible to humans, then the consumption of sheep brain (considered a delicacy by some) would likely have caused the emergence of vCJD long ago.

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Just how big is the risk of getting vCJD if I eat beef products?

CDC has identified a total of 4 people in North America that have been diagnosed with vCJD. All of those people had ties to either the United Kingdom or Saudi Arabi. CDC has concluded that their exposure to BSE most likely occurred outside of North America.

The prevalence of BSE in both the US and Canada is very low compared to other countries that have had outbreaks. Early detection of BSE and aggressive preventive measures have been taken to prevent human exposure to BSE. According to the CDC, the risk to human health from BSE in the United States is extremely low.

What about Chronic Wasting Disease (CWD) in deer and elk?

Chronic Wasting Disease is another transmissible spongiform encephalopathy. CWD was first identified as a fatal wasting syndrome in captive mule deer in Colorado in the late 1960s and in the wild in 1981. CWD does not appear to be connected to BSE in any way. CWD is considered endemic in free-ranging deer and elk located in a contiguous area in northeastern Colorado and southeastern Wyoming. A few privately owned herds also have CWD. In recent years, CWD has been found in areas outside of this disease-endemic zone, including areas east of the Mississippi River. The geographic range of diseased animals currently includes 11 U.S. states and two Canadian provinces.

Public health surveillance has not found any evidence that CWD can cause illness in humans consuming brain or spinal cord tissue from infected animals.

Sources of reliable information:

United States Department of Agriculture http://www.aphis.usda.gov/newsroom/hot_issues/bse/index.shtml

United States Department of Agriculture; Food Safety Inspection Service (FSIS) http://www.fsis.usda.gov/Fact_Sheets/Bovine_Spongiform_Encephalopathy_BSE/index.asp

United States Food and Drug Administration http://www.fda.gov/AnimalVeterinary/GuidanceComplianceEnforcement/ComplianceEnforcement/BovineSpongiformEncephalopathy/default.htm

Canadian Food Inspection Agency http://www.inspection.gc.ca/english/anima/heasan/disemala/bseesb/bseesbindexe.shtml

Department for Environment, Food and Rural Affairs (United Kingdom) www.defra.gov.uk

World Animal Health Organization http://www.oie.int/eng/info_ev/en_BSEHome.htm

The US Centers for Disease Control http://www.cdc.gov/ncidod/dvrd/bse/

For information on variant Creutzfeldt Jakob Disease at http://www.cdc.gov/ncidod/dvrd/vcjd/index.htm

American Association of Bovine Practitioners (AABP) www.aabp.org

Center for Food Safety and Public Health at Iowa State University www.cfsph.iastate.edu

National Cattlemen's Beef Association www.beef.org

The American Veterinary Medical Association (AVMA) http://www.avma.org/public_health/default.asp#bse

Your State Veterinarian/Department of Agriculture is also a source of information on BSE and other reportable diseases.

Information used in this article was taken from the sources listed above. Population estimates used in calculations were from the US Census Bureau, the CDC, the Office International Des Epizooties and the United Kingdom Dept of Health websites.