



## **MAD COW DISEASE FACT SHEET**

### **Q. What is mad cow disease?**

**A.** The popular name for bovine spongiform encephalopathy (BSE), it belongs to a group of related diseases known as “transmissible spongiform encephalopathies” (TSEs), or brain-wasting diseases. This group includes scrapie in sheep and Creutzfeldt Jakob Disease in humans. TSEs are caused by a protein that has folded itself into an abnormal shape, called a prion. Prions somehow induce other healthy proteins to fold incorrectly, leaving patches of useless debris and holes that turn brains to sponge, resulting in death.<sup>1</sup> The disease has an incubation period in cattle of up to eight years.<sup>2</sup>

### **Q. How do cattle get the disease?**

**A.** TSEs are thought to occur spontaneously along with being spread by consumption of infected central nervous system tissue. Cattle can get the disease by eating *less than one gram* of diseased meat and bone meal fed to them as a protein source.<sup>3</sup> The disease is not known to be contagious from one live animal to another.

### **Q. Can people get mad cow disease?**

**A.** Humans can contract a TSE from eating certain kinds of meat from cattle infected with BSE. This human form of the disease is known as variant Creutzfeldt-Jakob disease (vCJD). 146 people in Britain have been infected with vCJD from contaminated beef.<sup>4</sup> Tissue from the infected cow’s central nervous system (including brain or spinal cord) is the most infectious part of a cow; such tissue may be found in some hot dogs, taco fillings, bologna and other products made from ground or chopped meat, due to contamination during the extraction of the last bits of meat from cow carcasses.<sup>5</sup>

Transmission of vCJD has also occurred in over two dozen cases as a result of transplants or injections of body tissue from infected people.

### **Q. Have people in the US contracted vCJD from eating infected beef?**

**A.** So far, none of the CJD cases diagnosed in the US have been linked to US-produced beef, but this fact may have little bearing on the reality of the situation: the disease has a long incubation period and few dementia-related deaths in the US are investigated.<sup>6</sup> Only one person in the United States has been confirmed to have variant vCJD – she grew up in Britain and was probably infected there.

### **Q. What is Creutzfeldt-Jakob disease?**

**A.** “Classic” CJD inflicts about 250 people a year in the US. 85% of cases occur spontaneously, while 5-10% are hereditary and less than 1% of cases are acquired by exposure to infected nervous-system tissue through medical procedures. A person with naturally occurring CJD usually starts having symptoms in their 60s, with a rapid decline. Symptoms are similar to, and perhaps often confused with, Alzheimer’s disease. Victims rapidly develop staggering, memory loss, impaired vision and dementia, fall into comas, and die.<sup>7</sup>

The variant, vCJD, slowly deteriorates the brain and years, or even decades, later ends in death. Symptoms begin with confusion, depression, and behavioral changes, then impaired vision and coordination, and later dementia. vCJD usually kills its victims 6-9 months after the first appearance of symptoms. There is no cure, treatment, or vaccine for any TSE.

**Q. What is so unusual and risky about this disease?**

**A.** When scientists were trying to determine the cause of scrapie in sheep, they could not identify the organism that carries the disease. This is because it is not a virus nor a bacteria. And unlike other infections, it seemed to provoke absolutely no response from the body's immune system. Without antibodies (a response to infection), the only way to confirm an animal had scrapie was to examine its brain after death. Similarly, there is currently no test on live cattle to determine if they have BSE. Due to the long incubation period, symptoms of a sick cow won't appear for many years after infection. In addition, normal disinfection procedures do not stop this disease – so even well-cooked contaminated meat can infect humans. The rendering process (cooking of dead, often disease-ridden, animals), used to make supplements for animal feed, also cannot kill the infection – and only serves to spread it.

**Q. What countries have had cases of BSE in cattle?**

**A.** Mad Cow was first detected in England in the mid-1980s, where it has killed over 180,000 cattle. From Britain it spread to Europe, infecting over 4,200 cattle in 19 countries by mid-2003. The disease still kills 2-3 cattle each day in Europe.<sup>8</sup> Japan has had 11 confirmed cases since first found there in 2001. In May of 2003 a bull tested positive in Canada and the US had its first case in December of that year. The Czech Republic confirmed one case in January 2004. BSE has also appeared in Israel, Oman, and the Falkland Islands.<sup>9</sup> vCJD cases have been reported in the UK, France, Canada, Ireland, Italy and the US.<sup>10</sup>

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<sup>1</sup> Blakeslee, Sandra, and Marian Burros. "Danger to Public is Low, Experts on Disease Say." *New York Times*, December 24, 2003.

<sup>2</sup> Burros, Marian, and Donald G. McNeil Jr. "U.S. Inspections for Disease Lag Behind Those Abroad." *New York Times*, December 24, 2003.

<sup>3</sup> The BSE Inquiry, Report to the U.K. House of Commons, October 2000. <http://www.bseinquiry.gov.uk/index.htm>

<sup>4</sup> U.K. Department of Health. Monthly Creutzfeldt Jakob Disease Statistics, March 1, 2004.

<sup>5</sup> Blakeslee and Burros.

<sup>6</sup> Seltzer, Jed, and Elinor Mills Abreu. "U.S. Mad Cow Link Questioned in Creutzfeldt-Jakob Cases." Reuters, December 26, 2003.

<sup>7</sup> King, Warren. "What you need to know about this disease." *The Seattle Times*, December 24, 2003.

<sup>8</sup> Office of International des Epizooties. [http://www.oie.int/eng/info/en\\_esbmonde.htm](http://www.oie.int/eng/info/en_esbmonde.htm).

<sup>9</sup> Rampton, Sheldon, and John Stauber. *Mad Cow U.S.A. – Could the Nightmare Happen Here?* Maine: Common Courage Press, p. 157.

<sup>10</sup> King.