What is BSE, or “Mad Cow” Disease?
Mad Cow Disease is the nickname for an animal disease called bovine spongiform encephalopathy or BSE. The disease has struck European cattle, a handful of cattle in other countries including Japan, Canada and the United States. Ninety-five percent of BSE cases have occurred in the United Kingdom. The disease causes cattle to appear anxious, lose physical coordination and ultimately die.

When was BSE first discovered?
BSE was first diagnosed in cattle in the United Kingdom in 1986. When the BSE epidemic reached its peak, cases were occurring at a rapid rate, but as more information became available and scientists implemented aggressive prevention and control measures to prevent the disease, the rate of disease onset has slowed dramatically. Less than 200 cases worldwide were diagnosed in 2007 compared to more than 36,000 diagnosed in Great Britain in 1992 at the height of the epidemic. Only 7 cases were diagnosed in 2013.

What causes BSE?
The original source of BSE infectivity in cattle has not been definitively determined. BSE is spread through feed that contain the infective BSE agent.

What steps has the U.S. taken?
The United States recognized in the mid-1980s the seriousness of BSE when it witnessed the disease’s devastating impact on the U.K. cattle herd. As a result, the U.S. government, as well as the U.S. cattle, animal feed and beef industries have taken aggressive actions starting in 1989, to ensure that if BSE was introduced into the U.S. that it would not result in an outbreak in cattle similar to what Europe experienced.

The U.S. approach has been one of prevention using a “multiple firewalls” strategy. Together, these firewalls prevent BSE from being introduced into the U.S. and would quickly control its amplification and spread to animals and humans.

The first critical firewall in protecting U.S. cattle involves protecting U.S. borders. As early as 1989, the U.S. Department of Agriculture (USDA) banned the import of cattle and beef products from countries with BSE. As more scientific information became available, many governments including the United States began to focus on preventive controls.

The second critical firewall involves aggressive surveillance. USDA veterinarians are stationed at every U.S. meat packing plant and check cattle for signs of any disease - including BSE. No animal can be processed for meat without a veterinary inspection. All non-ambulatory cattle are excluded from processing. If an animal shows any symptoms that could possibly indicate BSE, the animal is condemned and removed for BSE testing.

USDA veterinarians, working with state and local counterparts and the rendering industry, target surveillance testing to high risk animals such as dead and non-ambulatory cattle. Since 2004, USDA has tested more than 800,000 cattle that are most at risk.

USDA also intends to develop a national ID system, which will assist with surveillance.

The third critical firewall involves controlling what cattle are fed. Evidence indicates that BSE is spread through infected feed. The feeding of protein derived from ruminant animals (cow, sheep, goat or deer) to ruminants has been banned since 1997. The latest FDA survey shows what the agency termed an “extraordinary” compliance rate of 99 percent. More recently, FDA published a proposed rule to remove certain materials from animal feed.

The fourth critical firewall is the removal of specified risk materials (SRMs). The BSE agent can be found in specified risk materials like the
brain of an infected animal. Therefore, the government requires the precautionary removal of SRMs to protect human health. SRMs are skull, brain, trigeminal ganglia, eyes, spinal cord and the dorsal root ganglia from cattle 30 months of age and older. And small intestine and tonsils from cattle of all ages. Additional firewalls include:

- A ban on non-ambulatory cattle from food supply.
- The prevention of carcasses that are tested for BSE from entering the food supply until a negative result is achieved.

These efforts provide a high degree of assurance that U.S. beef supply is and will remain safe.

In May 2013, the OIE recognized the United States’ proactive BSE prevention and control efforts classified the United States as a negligible risk country for BSE.

**Does beef from BSE-infected animals make people sick?**

No. The BSE agent accumulates primarily in brain and spinal cord tissue in infected cows. Evidence suggests that the same agent causes BSE and variant Creutzfeldt-jakob Disease, the neurological disorder that affects humans. However, vCJD is not the same as so-called “sporadic” CJD, which strikes older people worldwide at a rate of about one in every million people for unknown reasons.

About 200 cases of vCJD have been identified worldwide since BSE was first recognized in 1986. Most have occurred within the United Kingdom. It is widely believed consumption of BSE infected tissue is the apparent cause of BSE.

This underscores how rare the disease is and lends support to the theory that development of vCJD may require a combination of exposure to the BSE agent and a certain genetic predisposition.

In the United States, brain, spinal cord and other specified risk materials are banned for human consumption.

**People get classic CJD in the U.S. What causes it?**

Sporadic CJD has been a rare (one per million),yet recognized illness since the 1920s and occurs worldwide in countries at a relatively uniform rate regardless of dietary patterns. No evidence exists to suggest that CJD is caused by any food, including beef.

**What do the experts say about the ‘real’ risk of BSE?**

Following the release of the BSE Risk Assessment, done by Harvard University’s Center for Risk Analysis, the U.S. Department of Agriculture said, “measures taken by the government and industry make the United States robust against the spread of BSE to animals or humans.” To quote prominent health writer Abigail Trafford of The Washington Post, “The perception of risk is wildly out of proportion to the actual risk.”

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### HELPFUL LINKS

**North American Meat Institute**

- [www.meatinstitute.org](http://www.meatinstitute.org)
- [www.meatsafety.org](http://www.meatsafety.org)

**World Organization for Animal Health**

- [www.oie.int](http://www.oie.int)

**Animal and Plant Health Inspection Service**


**United Kingdom Department of Environment, Food and Rural Affairs**

- [www.defra.gov.uk](http://www.defra.gov.uk)

**USDA FMD Hotline**

- 800-601-9327

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**A Comparison of Bovine Spongiform Encephalopathy (BSE) in the United States and Europe**

<table>
<thead>
<tr>
<th></th>
<th>United States</th>
<th>Europe</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of BSE Cases</td>
<td>3</td>
<td>190,000</td>
</tr>
<tr>
<td>Cases of variant Creutzfeldt-Jakob Disease (vCJD)</td>
<td>4</td>
<td>229</td>
</tr>
<tr>
<td>Year Full Feed Controls Were Implemented</td>
<td>1997 (Seven years before first case)</td>
<td>1988 (two years after first case was detected)</td>
</tr>
</tbody>
</table>

* Non-native U.S. cases