



Prions In 1972, American scientist Stanley Prusiner became interested in scrapie, an infectious disease in sheep for which the exact cause was unknown. Although he first suspected a virus, experiments suggested that the disease might actually be caused by tiny particles found in the brain. Unlike viruses, these particles contained no DNA or RNA, only protein. Prusiner called these particles **prions**, short for “protein infectious particles.”  **Prions are misfolded versions of a normal cellular protein, called PrP (meaning "Prion Protein").**

What makes prions infectious, according to Prusiner's hypothesis, is that each misfolded prion can cause other PrP proteins to misfold in the same way. Eventually, so many prions accumulate, especially in the nervous system, that cells become damaged or destroyed. Prions are resistant to heat and digestive enzymes, so they are not destroyed by cooking infected meat.

 **CHECKPOINT** What is a prion? How do prions cause disease?

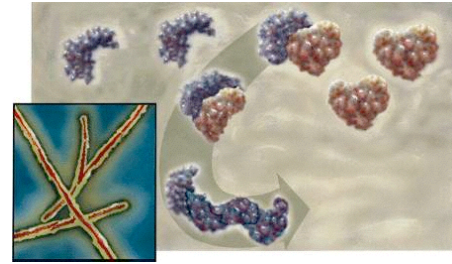
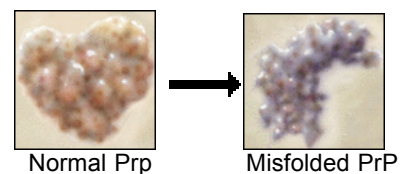
Mad Cow Disease In 1985, veterinarians in Great Britain found cows suffering from a disease that, like scrapie in sheep, attacked and destroyed parts of the brain. The disorder was called **BSE** (for "bovine spongiform encephalopathy"), but the erratic behavior of infected cattle led to the common name of "mad cow disease." When evidence emerged that mad cow disease and Creutzfeldt-Jakob disease (CJD), a similar disease in humans, might be caused by prions, people began to worry. Was it possible that more than 100 people in Britain had died from CJD caused by prion-infected beef?

In 1996, British authorities concluded that the practice of using tissue from sheep and cows to prepare cattle feed had made it possible for BSE to spread rapidly to cattle and then to humans who ate contaminated beef. They banned the use of cattle tissue in feed, and a similar ban was put in force in the United States the very next year. The epidemic of BSE, which had infected more than 100,000 cattle in Britain, gradually began to end.


In 2002, however, BSE was discovered in cows in Canada, and near the end of 2003, tissue from a Washington state cow with BSE was discovered after its meat had been processed. Public health authorities are now working aggressively to determine the sources of these BSE cases, and to ensure that infected tissue does not enter the human food supply.



Figure 19-17 This cow is suffering from BSE, a disease that gradually destroys its nervous system. **Applying Concepts** What is a prion? Can you classify prions as living or non-living? Explain.



prions spread by causing other proteins to misfold

 **Figure 19-18** Prions are misfolded versions of a normal cellular protein called PrP. Prions damage cells by causing other PrP proteins to misfold. Infected cells fill up with fiber-like clusters of prions (lower left)

Is BSE a Danger to Public Health? The British experience with mad cow and CJD in the 1990s shows two things:

- The first is that a widespread epidemic of BSE (more than 100,000 cows) can result in an increase in human cases of CJD. In Great Britain, more than 100 people developed a form of CJD, which is generally fatal, that can be traced to BSE. The exact number of deaths is difficult to determine, since some human cases of CJD are caused by mutations or spontaneous misfoldings of the PrP protein.
- The second is that a careful program regulating the preparation of cattle feed and requiring inspection of ranches and slaughterhouses can dramatically reduce the incidence of BSE.

Prior to 2003, public health authorities in the US and Canada had not gone quite as far as their European counterparts in acting against BSE. The fact that not a single case of BSE had been found in the US seemed to suggest that more extreme health measures might not be necessary. However, now that BSE has been found in both the United States and Canada, authorities are considering stricter measures. Since US exports of beef play an important part in the economies of many states (**Figures 19-19** and **19-20**), keeping the food supply safe is important both at home and abroad.



Figure 19-19 Cattle raising is an important part of the US economy, especially in western regions .



Figure 19-20 This worker at a restaurant in Korea is posting a sign to reassure its customers that it does not use US beef. **Inferring** How might the emergence of mad cow disease in the United States affect the exports of US beef?



Be sure to visit the Mad Cow page of the **millerandlevine** web site for the latest information on BSE and CJD:



<http://www.millerandlevine.com/news/bse/>

Biology News from millerandlevine.com

"Mad Cow" Disease appears in the United States

"Mad Cow" Disease is the common name for **Bovine Spongiform Encephalitis (BSE)**, a disease of cattle that was noticed in the United Kingdom in 1986. Cows with BSE behaved strangely - they staggered and drooled, sometimes acted aggressively, and at other times were unable to stand. When the cattle were slaughtered their brains were shot full of holes - filled with empty spaces where cells seemed to have died from a mysterious disorder. At first, the government in Britain assumed the holes were from a mysterious disease passed to their farm humans. But in 1988, medical authorities admitted that at least 10 people who had died from a similar human disorder (**Creutzfeldt-Jakob disease**, or **CJD**) had probably contracted CJD by eating meat products from BSE-infected cows.

Teacher Resources

Looking for teaching ideas on CJD (mad cow)? Here are several lesson plans recommended by the American Association for the Advancement of Science (AAAS) through their [Science, Med, and Life Program](#).

Diseases without Borders - studying the spread of disease across geographical boundaries. (Students will use the European Union's struggle with Mad Cow Disease as a starting point to study the spread of infectious disease across geographical boundaries.)

Invisible Invaders - Studying the effects of epidemics on different aspects of society. (In this lesson, students research various epidemics that have devastated the world population at different points in history, focusing on the historical events being discussed during the time of the epidemic.)

Since 1986, Britain and many other countries have adopted strict inspection rules to keep infected meat products from the human food supply. Many authorities hoped that similar rules in the United States would keep BSE from appearing here. But in December 2003, a confirmed case of BSE was found in tissues of a cow that had recently been slaughtered.

NY Times article on the first BSE case in the US
[Click to read on Mad Cow](#)
[Report from the Houston Chronicle](#)

Many aspects of BSE and CJD are still very poorly understood. But scientific opinion on the disease generally accepts the hypothesis put forward by [Maurice L. Stanley](#), Ph.D., an American biologist who has argued that these diseases are caused by protein molecules called "Prions." [Click here](#) to visit the web page of Dr. Prasher's laboratory.

The Prion Hypothesis suggests that diseases like mad cow and human CJD are caused by the misfolding of a protein known as PrP that most cells contain. Once a few copies of the protein become misfolded (the form shown in the right-hand image), they cause other PrPs to misfold, leading to an accumulation of unstable proteins in the cell. By mechanisms that are not understood, these unstable proteins cause cell death and damage the nervous system.

More Information about Mad Cow:

[National Cattlemen's Beef Association information on BSE](#)

BSE Resources and Articles from New Scientist:
<http://www.newscientist.com/biotech/bse/>