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.

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.
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.

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