Mad cow disease appeared for the first time in Britain in 1985. Since that time it has killed roughly 170,000 cows in Britain, and it has spread to humans.[1] In humans the disease is called "new variant Creutzfeld-Jakob disease," or nvCJD for short. At this point nvCJD has killed 24 people in Britain and one in France. More human deaths are expected in Britain[2] because several million people ate diseased beef before the British government (or the beef industry) acknowledged that mad cow disease could infect people.

From a U.S. perspective, the obvious question is, How can an outbreak of mad cow disease be prevented here?

Mad cow disease is a member of a family of rare diseases called transmissible spongiform encephalopathies, or TSEs. TSEs have different names in different animals (for example, scrapie in sheep, chronic wasting syndrome in deer and elk, and bovine spongiform encephalopathy or BSE in cows). However all TSEs share a few common features: they attack the central nervous system, causing disintegration of the brain; they have a long incubation period between the time when infection first occurs and the appearance of symptoms; TSEs are always fatal; and they are transmitted by the eating of animals or animal parts, especially brains and spinal cords.

TSEs are now thought to be caused by a unique disease agent, called a prion (pronounced PREE-on). A prion is simply a particular kind of protein. All mammals have prions, and some non-mammalian species have them as well. Prions are normal.

According to modern biology, a prion should not be able to reproduce itself, and therefore should not be able to cause disease, because prions contain no DNA. Without DNA, reproduction should not be possible. However, it is now becoming widely accepted that prions do reproduce themselves and do cause disease. Somehow a normal prion goes bad --it gets folded into an abnormal shape and in its abnormal shape it can destroy nerve cells in the central nervous system. The abnormal prions also cause other nearby prions to become folded into the same shape, thus creating more abnormal prions by a domino effect. After a long period of time (months or years, or even decades) the symptoms of disease appear, followed a few weeks or months later by death. Thus animals and humans can be carrying an infectious prion disease for months or years without showing any symptoms.

The prion theory of disease is still not accepted by 100% of the scientific community, but the inventor of the theory, Stanley B. Prusiner of the University of California at San Francisco, received the Nobel Prize for his work in 1997.[3]

In this country, the government agency with primary responsibility for preventing an outbreak of mad cow disease or its human variant, nvCJD, is the U.S. Food and Drug Administration (FDA).

The FDA in 1997 issued a rule declaring it illegal for farmers to feed animal protein from ruminants or mink to other ruminants --a preventive step that had been taken by the British government in 1988. Ruminants are animals that chew their cuds, including cattle, sheep, goats, deer and elk. Mink are included in the FDA's ban because they can get a TSE similar to mad cow disease.

When cows, pigs, and chickens are slaughtered, much of the animal cannot be used for food and is sent to a rendering plant to be ground up, boiled down, dried to the consistency of brown sugar and sold as feed for cows, pigs, chickens, and pets. It is this rendered "animal protein" derived from ruminants (and mink) that FDA has banned from feeding to ruminants.

The FDA's ruminant-to-ruminant ban still allows animal protein of all kinds to be fed to pigs and chickens, and it allows animal protein derived from pigs and chickens to be fed to ruminants. The FDA ban also allows blood and gelatin derived from ruminants to be fed to other ruminants. In the U.S., many newborn calves are fed a high-protein diet consisting mainly of dried blood. Blood cells carry prions just as nerve cells do.[4]

A small group of scientists, led by Dr. Michael Hansen of Consumers Union, has challenged the adequacy of FDA's ruminant-to-ruminant rule. [5] They argue that the FDA ban does not go far enough, "does not adequately protect human health, and is not scientifically defensible."[6] Consumers Union is the publisher of CONSUMER REPORTS magazine.

Scientists on both sides of the controversy agree that mad cow disease probably developed in Britain in one of two ways. Possibly cows ate parts of sheep that had been infected with the TSE called scrapie, and the scrapie, once in cows, evolved into mad cow disease. Or, alternatively, a prion spontaneously went bad (via genetic mutation of the gene that produces normal prions) in a cow, and that cow was fed to other cows, which were fed to other cows until the disease was amplified into an epidemic. In either case, it was cows (which are vegetarians by nature) being forced to eat animals that created the problem.

FDA officials say they are confident that their ruminant-to-ruminant ban has prevented, and will continue to prevent, an epidemic of mad cow disease in this country because (a) mad cow has never been observed in cows in the U.S., and (b) Creutzfeld-Jakob (CJD) disease is not increasing in the U.S.[7] If mad cow were occurring in U.S. cows, some form of CJD should be increasing, and it isn't, the FDA argues.

Michael Hansen of Consumers Union offers evidence that the government may be wrong on both counts. Here are his arguments:

Mad cow may have already appeared in U.S. cows. Hansen offers evidence from seven studies that some "downer" cows may have a form of mad cow disease, though with symptoms somewhat different from those in British cows. Downer cows are cows that cannot stand up, cows that collapse, and cows that die mysteriously.

In 1985 an outbreak of transmissible mink encephalopathy (TME) --a brain-destroying TSE of mink --occurred on six mink farms in Wisconsin. Because all the farms used the same ready-mix feed which came from the same feed plant, investigators assumed that the feed was the source of the infectious agent.[8] Two years later, in 1963, an outbreak of TME occurred on two more Wisconsin mink farms. Based on the 1961 outbreak, scientists suspected feed and they examined the two farms' feed records carefully. They learned that "downer" cows from farm A had been fed to mink on Farm A and Farm B. The researchers wrote, "Since mink on both farms developed the disease almost simultaneously, we believe this feed component has to be incriminated."[9]

In 1985 an outbreak of TME occurred on a mink ranch in Stetsonville, Wisconsin. Dr. Richard Marsh of the University of Wisconsin investigated and found that the mink had been fed 95% downer cows and 5% horse meat.[10] When brains from infected mink were injected into two calves, within 19 months both calves had a bovine TSE but they did NOT exhibit the symptoms of Britain's mad cows. The Stetsonville cows simply became lethargic and then fell over. In other words, they exhibited typical "downer cow" symptoms. When brains from these cows were injected into mink, the mink got TME, confirming the kind of disease that had killed the cows. Marsh and his colleagues concluded, "These results suggest the presence of a previously unrecognized scrapie-like infection in cattle in the United States."[10]

Marsh's cattle inoculation experiments have been repeated and, again, mink TME was transmitted to cows and back to mink and the cows exhibited "downer" symptoms, nothing like British mad cow disease.[8] Furthermore, in 1979 U.S. Department of Agriculture researchers in Mission, Texas, inoculated 10 cows with sheep...
scrapie. Three of the 10 cows developed neurological symptoms, but they were more like "downer cow" syndrome than British mad cow disease: "progressive difficulty in rising, a stiff-legged gait, incoordination, abnormal tail position, disorientation, and terminal recumbency [lying down]," according to Dr. Clarence Gibbs, Acting Chief of the Laboratory of Central Nervous System Studies at the National Institutes of Health.[11] Ten years later, when a test for mad cow disease became available, Dr. Gibbs confirmed a bovine TSE disease in the three cows, whose brains had been preserved.[11] Dr. Gibbs concluded, "Susceptibility of cattle to scrapie further suggests the possibility that sporadic cases of BSE [mad cow disease] may have occurred in the United States under the clinical picture of the downer cow syndrome..."[11]

After Gibbs confirmed that the Mission, Texas cows had indeed died of a TSE, the U.S. Department of Agriculture repeated the experiments at Ames, Iowa under the direction of Randall Cutlip.[12] Dr. Cutlip described the results: "All calves kept longer than one year became severely lethargic and demonstrated clinical signs of motor neuron dysfunction that were manifest as progressive stiffness, posterior paresis [partial paralysis], general weakness, and permanent recumbency [lying down]." In other words, cows infected with a sheep TSE had all the signs and symptoms of downer cows. Thus Hansen argues, there is considerable evidence that a TSE has been present in some U.S. cattle for several decades.

But if mad cow disease is already present in some number of cows in the U.S., where are the human victims? People should be getting some form of CJD [Creutzfeld-Jakob disease], and this disease is thought to be very rare and not increasing in the U.S. population. So where are the victims?

Hansen argues that CJD may be more prevalent in the U.S. population than is presently thought. The official figures say that CJD is exceptionally rare -- one case in every million people. In the U.S., this would mean there are 250 CJD cases at any given time. Hansen points to two studies in which people diagnosed with Alzheimer's were examined after death. In one study, among 54 presumed Alzheimer's victims, 3 (or 5.5%) were found to actually have CJD.[13] A Yale University study of 46 victims of Alzheimer's found that 6 (or 13%) actually died of CJD.[14] There are 2 million people with Alzheimer's in the U.S. (or 5.5% of them actually have CJD). There are 110,000 cases of CJD in the U.S., not 250 cases. If 2% of the 2 million have CJD, there are 260,000 cases of CJD in the U.S., not 250. If even 1% of the 2 million had CJD, it would mean there was an epidemic of 20,000 cases of CJD masquerading as Alzheimer's. Thus the FDA's argument that CJD is very rare, and not increasing, needs to be re-examined. [To be continued.]

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Descriptor terms: mad cow disease; emerging diseases; creutzfeld-jakob disease; new variant creutzfeld-jakob disease; nvcej; cjd; great britain; consumers union; bse; tse; transmissible spongiform encephalopathies; scrapie; britain; michael hansen; prions; prion theory of disease; fda; stanley prusiner; bans; ruminants; pigs; chickens; cows; consumers union; richard marsh; clarence gibbs; randall cutlip; alzheimer's disease;