In 1985, cows in Britain began to die of a mysterious ailment that no one had ever seen before. The symptoms were strange. At first the cows staggered and drooled, their ears twitching. Then they began to show signs of fear, grinding their teeth, acting aggressively toward other animals. Soon they died. Farmers named the condition "mad cow disease" and the name stuck.

The disease is now known formally as "bovine spongiform encephalopathy" or BSE for short. BSE is one of a small family of diseases called transmissible spongiform encephalopathies, or TSEs. TSEs destroy neurons, the main nerve cells in the brain, creating holes that leave the brain resembling a sponge (thus "spongiform"). BSE, like the other TSEs, is an infectious disease, meaning that it can be transmitted from one cow to another.

During the 13 years since it first appeared, mad cow disease has killed more than 167,000 cows in Britain[1] and many more became infected but were slaughtered for food before symptoms appeared. Symptoms take an average of 5 years to show up after a cow is infected. Until the late 1990s, there was no test that could detect whether a cow was infected -- only the appearance of symptoms and microscopic examination of the brain could provide a definitive diagnosis. (Even today, tests require brain tissue, so they cannot be conducted on live animals.) British-type BSE has now been observed in cows in Switzerland, the Netherlands, Ireland, Portugal, France, Oman and the Falkland Islands. To date, the disease has not been observed in cows in the U.S.

When the disease first struck in Britain in 1985, health authorities insisted that British beef was safe to eat. For 10 years they defended that position aggressively, despite mounting evidence to the contrary. Then in 1996, the official story changed suddenly and the public was stunned. A panel of government scientists told Parliament in 1996 that the "most likely explanation" for 10 cases of a new TSE disease in humans was that BSE had moved from cows into people. That explanation is now widely accepted by most scientists, though airtight evidence remains elusive.

By the time of the stunning announcement in 1996, some British experts calculated that more than a million infected cows had already been consumed in Britain.[2]

In humans, the BSE-like disease is called "new variant Creutzfeld-Jacob disease," or nvCJD for short. Creutzfeld-Jacob disease (CJD) is a member of the TSE family, a brain-destroyer. CJD has been recognized for a long time as a rare disease of the elderly -- very similar to Alzheimer's disease -- but nvCJD is different. It has somewhat different symptoms, a different pattern of disintegration in the brain, and it strikes young people, even teenagers. Between 1995 and early 1998, at least 23 people died of nvCJD in Britain and at least one in France,[3] the oldest of them age 42 and the youngest 15.

In January 1997, British epidemiologists tried to estimate how large the outbreak of nvCJD might become. They concluded that the data were not sufficient to allow a precise estimate: somewhere between 75 people and 80,000 people would eventually die of the new disease, they estimated.[4] Only time will tell. More precise estimates of the size of the problem are not possible because no one knows for sure how long nvCJD "incubates," how much time elapses between infection and the appearance of symptoms.

An excellent recent book by Sheldon Rampton and John Stauber, MAD COW U.S.A.--CAN THE NIGHTMARE HAPPEN HERE? tells the story of the emergence of mad cow disease in Britain, and the serious and political battles that ensued.[5] Despite the evocative title, the book is thoroughly documented and carefully-written. Without oversimplifying the details, the book recounts a complicated story: the medical discoveries, strongarm tactics by the beef industry, and waffling and coverup by governments on both sides of the Atlantic -- yet it reads as easily as a detective story. As a piece of science writing -- a description of science as it plays out in the real world -- the book is an impressive accomplishment. By the end, the reader has absorbed several important lessons about public health policies distorted by big money, and about very serious threats to our first amendment right of free speech. Rampton and Stauber show that the U.S. beef industry will go to almost any lengths to try to prevent a public discussion of "mad cow" and the steps that the U.S. government still needs to take to prevent this disease from becoming an American problem.

In a nutshell: Mad cow disease developed because of a little-known but very-widespread modern agricultural practice -- farmers feeding dead cows to their cows, thus turning a vegetarian species into meat-eaters.

In Britain and the U.S., when a cow is slaughtered, about half of the animal cannot be sold for human uses -- the hide, bones, entrails, hooves, horns, fat, gristle and tough membranes are, by law, not permitted to be used in food. Obviously something else has to be done with these parts, to avoid creating a public health hazard.

Then there are "downer cows" that must be dealt with. Cows that cannot stand up, cows that collapse, and cows that die mysteriously are called "downers." Every year in the U.S. about 100,000 cows die of unknown causes. One day they are alive and the next day they are dead, and no one knows why. Something must be done with their carcasses.

In addition to downer cows there are thousands of pigs, horses, chickens, and sheep that die of unknown causes each year. To prevent public health problems, they must be disposed of. Then there is road kill -- deer, elk and other large mammals killed by motor vehicles.

In the U.S., all of these animals and animal parts end up at 280 "rendering" plants where they are ground up and boiled down. (The British call such plants "abattoirs." ) Up until the 1960s, the fat from rendering plants was generally used to make soap. But the invention of detergents, derived from petroleum, greatly reduced the demand for soap. As a result, the rendering industry had to develop new markets for its products. They hit on animal feed, and it became a great commercial success.

From the 1960s through the mid-1990s, rendering plants dried their rendered products, ground them into the consistency of brown sugar, and sold them for animal feed. Feed mills then mixed these animal by-products into various feed formulations -- about one-third for cattle, one-third for pigs and chickens, and one-third for pets.

Unfortunately, some of the animals sent to rendering plants (or abattoirs) are sheep killed by a disease called "scrapie." Scrapie is another TSE, a member of the same family as BSE, CJD, and nvCJD -- one of those diseases that eats holes in the brain and invariably kills its victims. Scrapie takes its name from the way sheep act once they get the disease -- they rub up against a fence or a barn until they scrape away their wool, leaving raw wounds. Then they die.

Scrapie has been a well-known, though mysterious, disease of sheep for at least 200 years, but only recently have scrapie-infected sheep been fed to cows. Scientists who study mad cow disease believe that the illness crossed the species barrier from sheep to cows through contaminated feed. Indeed, the British in 1988 banned the practice of feeding animal carcasses to other animals and within seven years new cases of mad cow disease diminished quite dramatically from 900 to 1000 per week to 280 to 300 per week.[6] The U.S., however, has been slow to act. Scrapie is well-established among sheep in this country. From October, 1988, to June, 1989, scrapie was diagnosed in 52 flecks of sheep in 20 states.[5,pg.104] Clearly, there was ample reason to ban the feeding of animal carcasses to animals in this country when the British took
action in 1988. However, the beef industry, and the rendering and feed industries, have generally opposed such precautionary measures.

TSEs have a very long incubation period. In cows, BSE takes three to eight years (average, five years) from the time of first infection to the appearance of symptoms shortly before death. CJD in humans has an incubation period of 10 to 40 years. Thus, by the time symptoms of BSE appear, many cows are likely to be carrying the infection silently. This was confirmed earlier this year in Switzerland when tests of brain tissues from 1761 cows revealed 8 infected animals without any symptoms, for a "silent" infection rate of 4.5 per 1000. This is 100 times as high as the rate of Swiss cows showing symptoms.[7] If this rate holds for Britain, it means that today there are about 460,000 British cows infected -- but symptom-free -- in a total herd of roughly 11 million.

TSE diseases are characterized by a long incubation period, and they are always fatal. Furthermore, the infectious agent is incredibly resistant to deactivation. Cooking infected meat, or even rendering it at high temperature, does not completely eliminate its infectivity. Animals get TSEs by eating infected animals or parts of infected animals, especially nerve tissues.

TSE diseases have now been identified in sheep, pigs, goats, cattle, deer, elk, mink, mice, hamsters, guinea pigs, domestic cats, puma, cheetah, eland, kudu, Arabian oryx, myland, marmosets, macaques, chimpanzees and humans. In addition, a TSE has been reported in ostriches in a German zoo.[8]

Thus one might think the U.S. Food and Drug Administration (FDA) would prohibit the feeding of any animals to any other animals, as the British did in 1988. But that is not what the FDA has done. Under pressure from the beef, rendering, and feed industries, in 1997 FDA only prohibited the feeding of ruminants and mink to ruminants.[1,8] Ruminants are animals that chew their cuds, including cattle, sheep, goats, deer and elk. Mink are included in the ban because they can get a TSE similar to mad cow disease.

FDA is still allowing the feeding of pigs to other animals, and the feeding of blood and gelatin from rendering plants to all animals. For example, many calves in the U.S. are being raised on a diet of dried blood taken straight from rendering plants. Pigs and chickens are still being fed rendered animal products. There are sound scientific arguments why this policy represents a form of Russian roulette being played with the health of the American public. Given that we are dealing with infectious diseases that invariably kill, the precautionary principle (see REHW #586) seems the only appropriate policy.

More next week.

--Peter Montague (National Writers Union, UAW Local 1981 AFL-CIO)

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Descriptor terms: mad cow disease; emerging diseases; creutzfeld-jakob disease; new variant creutzfeld-jakob disease; nvjcjd; cjd; great britain; consumers union; bse; tse; transmissible spongiform encephalopathies; sheldon rampton; john stauber; scrapie; britain; michael hansen;