When a new form of an old human disease appeared in England in 1995, some medical specialists immediately suspected that it might be a human version of "mad cow disease," but they had no proof. Mad cow disease had appeared in British dairy cattle for the first time in 1985 and during the subsequent decade 175,000 British cows had died from it. British health authorities spent that decade reassuring the public that there was no danger from eating the meat of infected cows. They said a "species barrier" prevented mad cows from infecting humans. A "species barrier" does prevent many diseases from crossing from one species to another -- for example, measles and canine distemper are closely related diseases, but dogs don't get measles and humans don't get distemper.

While the British government was placing its faith in the species barrier, British citizens began to die of a new disease, called "new variant Creutzfeld-Jakob disease" or nvCJD. A similar disease, CJD (Creutzfeld-Jakob disease) had been recognized for a long time but it almost never occurs in people younger than 30; nvCJD, on the other hand, strikes people as young as 13. There are several other differences between CJD and nvCJD, so nvCJD represents something new. To date, nvCJD has killed 48 people in England and one or two others elsewhere in Europe. The main feature of both mad cow disease and nvCJD is the progressive destruction of brain cells, inevitably leading to total disability and death.

New research published late in 1999 showed that nvCJD is, in fact, a human form of mad cow disease,[2] dashing all hope that a species barrier can protect humans from this deadly bovine affliction.

Mad cow disease is formally known as "bovine spongiform encephalopathy" or BSE. BSE is the cow version of a larger class of diseases called "transmissible spongiform encephalopathies," or TSEs. TSEs can afflict sheep, deer, elk, cows, mink, cats, squirrels, monkeys, humans and other species. In all species the symptoms of TSEs are the same -- progressive destruction of brain cells leading to dementia and death.

Traditional Creutzfeld-Jakob disease (CJD) is a rare human affliction. The visible symptoms are similar to Alzheimer's disease; in fact, CJD is sometimes diagnosed as Alzheimer's and therefore may go unrecognized. CJD strikes one in a million people almost all of whom are older than 55. In people younger than age 30, CJD is extremely rare, striking an average of 5 people per billion each year, worldwide (not counting the recent outbreak in England).

In cows, the latency (or incubation) period for mad cow disease is about 5 years, meaning that cows have the disease for five years before symptoms begin to appear. No one knows the latency period for nvCJD in humans, but it is thought to be around 10 years. Because of this uncertainty, no one is sure how many people in England already have the disease but are not yet showing symptoms. The British government's chief medical officer, Professor Liam Donaldson, said December 21, 1999, "We're not going to know for several years whether the size of the epidemic will be a small one, in other words the hundreds, or a very large one, in the hundreds of thousands." The epidemic of mad cow disease was caused by an agricultural innovation -- feeding dead cows to live cows. Cows are, by nature, vegetarians. But modern agricultural techniques changed that. Cows that died mysteriously were sent to rendering plants where they were boiled down and ground up into the consistency of brown sugar, and eventually added to cattle feed. It was later determined that mad cow disease was being transmitted through such feed, and especially through certain specific tissues -- brain, spinal cord, eyes, spleen and perhaps other nerve tissues.

Ten new cases of nvCJD were reported in England in 1999, bringing the total to 48. It has been more than 10 years since government authorities banned the use of the particular parts of cows thought to transmit mad cow disease. The appearance of new cases of nvCJD in 1999 implies either that the latency period for the disease is longer than 10 years, or that infected meat was not effectively eliminated from the food chain when government authorities said it was, or both.

The SUNDAY TIMES of London reported in late December that some meat banned for human consumption is still being marketed in England. After the mad cow scandal erupted, the British government attempted to eradicate the disease by requiring that all cows older than 30 months be slaughtered. As a result, by last September more than 2.5 million British cows had been killed. But the TIMES reported that British investigators have documented at least 50 cases of farmers and cattle dealers using bogus identity documents to falsify the ages of cows in order to sell them for human consumption. Furthermore, the Agriculture Ministry acknowledged that as many as 90,000 cattle could not be accounted for. About 1600 new cases of mad cow disease are still being reported each year in England.

In December, French health authorities announced finding a second case of nvCJD, a 36-year-old woman in Paris. France has continued to refuse to import British beef, even though the European Union on August 1, 1999, formally declared British beef as safe as any in the European Union. The European Union said in December it will take France to the European Court of Justice to force it to import British beef. Germany is also refusing to import British beef.

The U.S. government says mad cow disease has never been observed in any U.S. cows. However, a closely-related TSE disease, called chronic wasting disease (CWD), has been increasing for almost 20 years among wild deer and elk in northern Colorado and southern Wyoming. Since 1981, CWD has been spreading slowly among wild deer and elk herds in the Rocky Mountains and now afflicts between 4% and 8% of 62,000 deer in the region between Fort Collins, Colorado and Cheyenne, Wyoming.

During 1999, CWD erupted among a herd of elk on the David Kesler Game Farm near Philipburg, Montana, which raised elk commercially. A few of Mr. Kesler's elk had been shipped to Oklahoma and Idaho, and perhaps elsewhere, and CWD was discovered in some of those animals, too. In early December, Montana health authorities slaughtered 81 elk on Mr. Kesler's farm. They initially announced plans to incinerate the carcasses, but later decided that incineration would be too expensive. The animals were finally buried at the High Plains Sanitary Landfill north of Great Falls. Equipment used to feed, water and care for the animals was also buried in the landfill. Montana authorities announced that the fence line at the elk farm would be decontaminated, but they did not say what procedure they would use. Nor did they announce what would become of Mr. Kesler's contaminated land. The disease agent that causes CWD -- a prion protein -- is very hardy and resists destruction by traditional sterilization techniques like alcohol and heat.

The diseased elk carcasses in the High Plains landfill have been buried under a mound of garbage but will still be accessible to rainwater and perhaps to scavenging animals.

In northeastern Colorado and southeastern Wyoming, state officials are urging hunters to protect themselves when dressing wild deer and elk they have shot. Hunters should wear rubber gloves, minimize contact with brain and spinal cord tissues, discard the brain, spinal cord, eyes, spleen and lymph nodes and definitely not eat them. There is no evidence that CWD can cross over from deer and elk to humans, but there was no firm evidence that mad cow disease could afflict humans until 1999, so wildlife officials in the Rocky Mountain states say caution is warranted.

Writing in the BOSTON GLOBE, Terry J. Allen reported in late 1999 that, since 1996, Creutzfeld-Jakob disease has been identified in 3 Americans younger than age 30.[3] All three are known to have hunted extensively or eaten venison. There is no evidence that CWD disease has jumped from deer or elk to humans, but the appearance...
of this extremely-rare disease in young people was the first evidence of a problem in England, so health authorities in the U.S. say they are aggressively investigating all the possibilities.

A statistician at the federal Centers for Disease Control (CDC) in Atlanta told Terry Allen that, if one more case of CJD had surfaced in a person younger than 30 in the U.S., it "might tip the balance," meaning it might convince authorities that something truly unusual was occurring. Dr. Michael Hansen of Consumer's Union says, "Given how rare the disease is in young people and how difficult it is to make a diagnosis, the possibility that some cases go undetected cannot be ruled out."[3]

Indeed, of the 3 cases detected in the U.S. since 1996, one nearly went undetected. Last year in Utah, Doug McEwan, 28, began to show an array of mysterious symptoms: loss of memory, loss of motor control, mood swings, and disorientation. His wife, Tracey, says his doctors conducted hundreds of tests but could not diagnose his disease. She happened to see a TV program on mad cow disease and she insisted that Doug's doctors must test for CJD. A brain biopsy confirmed the diagnosis.

One of the three young CJD victims had eaten deer shot near Rangely, Maine, so last November federal officials took samples of brains from 299 deer shot in western Maine. Authorities said at the time they were quite sure Maine deer are not harboring CWD. So far, test results have not been released.

Federal authorities have quarantined two herds of sheep in Vermont because they say the sheep may have been given feed that contained parts of animals afflicted by mad cow disease. The sheep had been imported into Vermont from Belgium and the Netherlands, where they may have been fed improperly. A similar herd of sheep in New York state was recently purchased by the federal government and slaughtered.[4]

Meanwhile, a 68-year-old Indiana man with a fondness for beef-brain sandwiches died of CJD last summer. Beef-brain sandwiches are a local delicacy in Indiana, introduced years ago by German immigrants. The EVANSVILLE (INDIANA) COURIER reported that John Hiedingsfelder, a forensic pathologist in Evansville, said he had seen three cases of CJD in the past year. No connection to mad cow disease has been established in the Indiana cases, Roberta Heiman, a staff writer for the EVANSVILLE (INDIANA) COURIER reportedly received a warning from a cattleman's association not to publish any further articles about this subject.

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

[1] Unless a specific source is cited, information in this issue of Rachel's was taken from www.mad-cow.org, a web site maintained by Thomas Pringle of Eugene, Oregon. Sources of information are cited at www.mad-cow.org.


Descriptor terms: mad cow disease; england; france; montana; wyoming; vermont; maine; deer; elk; bse; tse; central nervous system disorders;