

## Could Mad Cow Disease Already be Killing Thousands of Americans Every Year?

January 7, 2004 by Michael Greger, M.D. for the  
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October 2001, 34-year-old Washington State native Peter Putnam started losing his mind. One month he was delivering a keynote business address, the next he couldn't form a complete sentence. Once athletic, soon he couldn't walk. Then he couldn't eat. After a brain biopsy showed it was Creutzfeldt-Jakob disease, his doctor could no longer offer any hope. "Just take him home and love him," the doctor counseled his family.[1,2,3] Peter's tragic death, October 2002, may have been caused by Mad Cow disease.

Seven years earlier and 5000 miles away, Stephen Churchill was the first in England to die. His first symptoms of depression and dizziness gave way to a living nightmare of terrifying hallucinations; he was dead in 12 months at age 19.[4] Next was Peter Hall, 20, who showed the first signs of depression around Christmas, 1994. By the next Christmas, he couldn't walk, talk, or do anything for himself.[5] Then it was Anna's turn, then Michelle's. Michelle Bowen, age 29, died in a coma three weeks after giving birth to her son via emergency cesarean section. Then it was Alison's turn. These were the first five named victims of Britain's Mad Cow epidemic. They died from what the British Secretary of Health called the worst form of death imaginable, Creutzfeldt-Jakob disease, a relentlessly progressive and invariably fatal human dementia.[6] The announcement of their deaths, released on March 20, 1996 (ironically, Meatout Day[7]), reversed the British government's decade-old stance that British beef was safe to eat.[8]

It is now considered an "incontestable fact" that these human deaths in Britain were caused by Bovine Spongiform Encephalopathy (BSE), or Mad Cow disease.[9] Bovine means "cow or cattle," spongiform means "sponge-like," and encephalopathy means "brain disease." Mad Cow disease is caused by unconventional pathogens called prions--literally infectious proteins--which, because of their unique structure, are practically invulnerable, surviving even incineration[10] at temperatures hot enough to melt lead.[11] The leading theory as to how cows got Mad Cow disease in the first place is by eating diseased sheep infected with a sheep spongiform encephalopathy called scrapie.[12]

In humans, prions can cause Creutzfeldt-Jakob disease (CJD), a human spongiform encephalopathy whose clinical picture can involve weekly deterioration into blindness and epilepsy as one's brain becomes riddled with tiny holes.

We've known about Creutzfeldt-Jakob disease for decades, since well before the first mad cow was discovered in 1985. Some cases of CJD seemed to run in families; other cases seemed to just arise spontaneously in about one in a million people every year, and were hence dubbed "sporadic." The new form of CJD caused by eating beef from cows infected with Mad Cow disease, though, seemed to differ from the classic sporadic CJD.

The CJD caused by infected meat has tended to strike younger people, has produced more psychotic symptoms, and has often dragged on for a year or more. The most defining characteristic, though, was found when their brains were sampled. The brain pathology was vividly reminiscent of Kuru, a disease once found in a New Guinea tribe of cannibals who ate the brains of their dead.[13] Scientists called this new form of the disease "variant" CJD.

Other than Charlene, a 24 year old woman now so tragically dying in Florida, who was probably infected in Britain, there have been no reported cases of variant CJD in the U.S.[14] Hundreds of confirmed cases of the sporadic form of Creutzfeldt-Jakob disease, however, arise in the United States every year,[15] but the beef industry is quick to point out these are cases of sporadic CJD, not the new variant known to be caused by Mad Cow disease.[16] Of course, no one knows what causes sporadic CJD. New research, discussed below, suggests that not hundreds but thousands of Americans die of sporadic CJD every year, and that some of these CJD

deaths may be caused by eating infected meat after all.

Although the fact that Mad Cow disease causes variant CJD had already been strongly established, researchers at the University College of London nevertheless created transgenic mice complete with "humanized" brains genetically engineered with human genes to try to prove the link once and for all. When the researchers injected one strain of the "humanized" mice with infected cow brains, they came down with the same brain damage seen in human variant CJD, as expected. But when they tried this in a different strain of transgenic "humanized" mice, those mice got sick too, but most got sick from what looked exactly like sporadic CJD! The Mad Cow prions caused a disease that had a molecular signature indistinguishable from sporadic CJD. To the extent that animal experiments can simulate human results, their shocking conclusion was that eating infected meat might be responsible for some cases of sporadic CJD in addition to the expected variant CJD. The researchers concluded that "it is therefore possible that some patients with [what looks like]... sporadic CJD may have a disease arising from BSE exposure." [17] Laura Manuelidis, section chief of surgery in the neuropathology department at Yale University comments, "Now people are beginning to realize that because something looks like sporadic CJD they can't necessarily conclude that it's not linked to [Mad Cow disease]..." [18]

This is not the first time meat was linked to sporadic CJD. In 2001, a team of French researchers found, to their complete surprise, a strain of scrapie--"mad sheep" disease--that caused the same brain damage in mice as sporadic CJD. [19] "This means we cannot rule out that at least some sporadic CJD may be caused by some strains of scrapie," says team member Jean-Philippe Deslys of the French Atomic Energy Commission's medical research laboratory. [20]

Population studies had failed to show a link between CJD and lamb chops, but this French research provided an explanation why. There seem to be six types of sporadic CJD and there are more than 20 strains of scrapie. If only some sheep strains affect only some people, studies of entire populations may not clearly show the relationship. Monkeys fed infected sheep brains certainly come down with the disease. [21] Hundreds of "mad sheep" were found in the U.S. in 2003. [22] Scrapie remains such a problem in the United States that the USDA has issued a scrapie "declaration of emergency." [23] Maybe some cases of sporadic CJD in the U.S. are caused by sheep meat as well. [24]

Pork is also a potential source of infection. Cattle remains are still boiled down and legally fed to pigs (as well as chickens) in this country. The FDA allows this exemption because no "naturally occurring" porcine (pig) spongiform encephalopathy has ever been found. But American farmers typically kill pigs at just five months of age, long before the disease is expected to show symptoms. And, because pigs are packed so tightly together, it would be difficult to spot neurological conditions like spongiform encephalopathies, whose most obvious symptoms are movement and gait disturbances. We do know, however, that pigs are susceptible to the disease--laboratory experiments show that pigs can indeed be infected by Mad Cow brains [25]--and hundreds of thousands of downer pigs, too sick or crippled by injury to even walk, arrive at U.S. slaughterhouses every year. [26]

A number of epidemiological studies have suggested a link between pork consumption and sporadic CJD. Analyzing peoples' diet histories, the development of CJD was associated with eating roast pork, ham, hot dogs, pork chops, smoked pork, and scrapple (a kind of pork pudding made from various hog carcass scraps). The researchers concluded, "The present study indicated that consumption of pork as well as its processed products (e.g., ham, scrapple) may be considered as risk factors in the development of Creutzfeldt-Jakob disease." Compared to people that didn't eat ham, for example, those who included ham in their diet seemed ten times more likely to develop CJD. [27] In fact, the USDA may have actually recorded an outbreak of "mad pig" disease in New York 25 years ago, but still refuses to reopen the investigation despite petitions from the Consumer's Union (the publishers of Consumer Reports magazine). [28]

Sporadic CJD has also been associated with weekly beef consumption, [29] as well as the consumption of roast lamb, [30] veal, venison, brains in general, [31] and, in North America, seafood. [32,33] The development of CJD

has also, surprisingly, been significantly linked to exposure to animal products in fertilizer,[34] sport fishing and deer hunting in the U.S.,[35] and frequent exposure to leather products.[36]

We do not know at this time whether chicken meat poses a risk. There was a preliminary report of ostriches allegedly fed risky feed in German zoos who seemed to come down with a spongiform encephalopathy.[37] Even if chickens and turkeys themselves are not susceptible, though, they may become so-called "silent carriers" of Mad Cow prions and pass them on to human consumers.[38] Dateline NBC quoted D. Carleton Gajdusek, the first to be awarded a Nobel Prize in Medicine for his work on prion diseases,[39] as saying, "it's got to be in the pigs as well as the cattle. It's got to be passing through the chickens." [40] Dr. Paul Brown, medical director for the US Public Health Service, believes that pigs and poultry could indeed be harboring Mad Cow disease and passing it on to humans, adding that pigs are especially sensitive to the disease. "It's speculation," he says, "but I am perfectly serious." [41]

The recent exclusion of most cow brains, eyes, spinal cords, and intestines from the human food supply may make beef safer, but where are those tissues going? These potentially infectious tissues continue to go into animal feed for chickens, other poultry, pigs, and pets (as well as being rendered into products like tallow for use in cosmetics, the safety of which is currently under review[42]). Until the federal government stops the feeding of slaughterhouse waste, manure, and blood to all farm animals, the safety of meat in America cannot be guaranteed.

The hundreds of American families stricken by sporadic CJD every year have been told that it just occurs by random chance. Professor Collinge, the head of the University College of London lab, noted "When you counsel those who have the classical sporadic disease, you tell them that it arises spontaneously out of the blue. I guess we can no longer say that."

"We are not saying that all or even most cases of sporadic CJD are as a result of BSE exposure," Professor Collinge continued, "but some more recent cases may be--the incidence of sporadic CJD has shown an upward trend in the UK over the last decade... serious consideration should be given to a proportion of this rise being BSE-related. Switzerland, which has had a substantial BSE epidemic, has noted a sharp recent increase in sporadic CJD." [43] In the Nineties, Switzerland had the highest rate of Mad Cow disease in continental Europe, and their rate of sporadic CJD doubled. [44]

We don't know exactly what's happening to the rate of CJD in this country, in part because CJD is not an officially notifiable illness. [45] Currently only a few states have such a requirement. Because the Centers for Disease Control (CDC) does not actively monitor the disease on a national level, [46] a rise similar to the one in Europe could be missed. [47] In spite of this, a number of U.S. CJD clusters have already been found. In the largest known U.S. outbreak of sporadic cases to date, [48] five times the expected rate was found to be associated with cheese consumption in Pennsylvania's Lehigh Valley. [49] A striking increase in CJD over expected levels was also reported in Florida [50] and New York (Nassau County) [51] with anecdotal reports of clusters of deaths in Oregon [52] and New Jersey. [53]

Perhaps particularly worrisome is the seeming increase in CJD deaths among young people in this country. In the 18 years between 1979 and 1996, only a single case of sporadic CJD was found in someone under 30. Whereas between 1997 and 2001, five people under 30 died of sporadic CJD. So five young Americans dying in five years, as opposed to one young case in the previous 18 years. The true prevalence of CJD among any age group in this country remains a mystery, though, in part because it is so commonly misdiagnosed. [54]

The most frequent misdiagnosis of CJD among the elderly is Alzheimer's disease. [55] Neither CJD nor Alzheimer's can be conclusively diagnosed without a brain biopsy, [56] and the symptoms and pathology of both diseases overlap. There can be spongy changes in Alzheimer's, for example, and senile Alzheimer's plaques in CJD. [57] Stanley Prusiner, the scientist who won the Nobel Prize for his discovery of prions, speculates that Alzheimer's may even turn out to be a prion disease as well. [58] In younger victims, CJD is more often

misdiagnosed as multiple sclerosis or as a severe viral infection.[59]

Over the last 20 years the rates of Alzheimer's disease in the United States have skyrocketed.[60] According to the CDC, Alzheimer's Disease is now the eighth leading cause of death in the United States,[61] afflicting an estimated 4 million Americans.[62] Twenty percent or more of people clinically diagnosed with Alzheimer's disease, though, are found at autopsy not to have had Alzheimer's at all.[63] A number of autopsy studies have shown that a few percent of Alzheimer's deaths may in fact be CJD. Given the new research showing that infected beef may be responsible for some sporadic CJD, thousands of Americans may already be dying because of Mad Cow disease every year.[64]

Nobel Laureate Gajdusek, for example, estimates that 1% of people showing up in Alzheimer clinics actually have CJD.[65] At Yale, out of a series of 46 patients clinically diagnosed with Alzheimer's, six were proven to have CJD at autopsy.[66] In another study of brain biopsies, out of a dozen patients diagnosed with Alzheimer's according to established criteria, three of them were actually dying from CJD.[67] An informal survey of neuropathologists registered a suspicion that CJD accounts for 2-12% of all dementias in general.[68] Two autopsy studies showed a CJD rate among dementia deaths of about 3%.[69,70] A third study, at the University of Pennsylvania, showed that 5% of patients diagnosed with dementia had CJD.[71] Although only a few hundred cases of sporadic CJD are officially reported in the U.S. annually,[72] hundreds of thousands of Americans die with dementia every year.[73] Thousands of these deaths may actually be from CJD caused by eating infected meat.

The incubation period for human spongiform encephalopathies such as CJD can be decades.[74] This means it can be years between eating infected meat and getting diagnosed with the death sentence of CJD. Although only about 150 people have so far been diagnosed with variant CJD worldwide, it will be many years before the final death toll is known. In the United States, an unknown number of animals are infected with Mad Cow disease, causing an unknown number of human deaths from CJD. The U.S. should immediately begin testing all cows destined for human consumption, as is done in Japan, should stop feeding slaughterhouse waste to all farm animals (see <http://organicconsumers.org/madcow/GregerBSE.cfm>), and should immediately enact an active national surveillance program for CJD.[75]

Five years ago this week, the Center for Food Safety, the Humane Farming Association, the Center for Media & Democracy, and ten families of CJD victims petitioned the FDA and the CDC to immediately enact a national CJD monitoring system, including the mandatory reporting of CJD in all 50 states.[76] The petition was denied.[77] The CDC argued that their passive surveillance system tracking death certificate diagnoses was adequate. Their analysis of death certificates in three states and two cities, for example, showed an overall stable and typical one in a million CJD incidence rate from 1979 to 1993.[78] But CJD is so often misdiagnosed, and autopsies are so infrequently done, that this system may not provide an accurate assessment.[79]

In 1997, the CDC set up the National Prion Disease Pathology Surveillance Center at Case Western Reserve University to analyze brain tissue from CJD victims in the U.S. in hopes of tracking any new developments. In Europe, surveillance centers have been seeing most, if not all, cases of CJD. The U.S. center sees less than half. "I'm very unhappy with the numbers," laments Pierluigi Gambetti, the director of the Center. "The British and Germans politely smile when they see we examine 30% or 40% of the cases," he says. "They know unless you examine 80% or more, you are not in touch." [80] "The chance of losing an important case is high." [81]

One problem is that many doctors don't even know the Center exists. And neither the CDC nor the Center are evidently authorized to reach out to them directly to bolster surveillance efforts, because it's currently up to each state individually to determine how--or even whether--they will track the disease. In Europe, in contrast, the national centers work directly with each affected family and their physicians.[82] In the U.S., most CJD cases--even the confirmed ones--seem to just fall through the cracks. In fact, based on the autopsy studies at Yale and elsewhere, it seems most CJD cases in the U.S. aren't even picked up in the first place.

Autopsy rates have dropped in the U.S. from 50% in the Sixties to less than 10% at present.[83] Although one reason autopsies are rarely performed on atypical dementia cases is that medical professionals are afraid of catching the disease,[84] the primary reason for the decline in autopsy rates in general appears to be financial. There is currently no direct reimbursement to doctors or hospitals for doing autopsies, which often forces the family to absorb the cost of transporting the body to an autopsy center and having the brain samples taken, a tab that can run upwards of \$1500.[85]

Another problem is that the National Prion Disease Pathology Surveillance Center itself remains underfunded. Paul Brown, medical director for the National Institutes of Health, has described the Center's budget as "pitiful," complaining that "there isn't any budget for CJD surveillance." [86] To adequately survey America's 290 million residents, "you need a lot of money." UK CJD expert Robert Will explains, "There was a CJD meeting of families in America in which... [the CDC] got attacked fairly vigorously because there wasn't proper surveillance. You could only do proper surveillance if you have adequate resources." [87] "I compare this to the early days of AIDS," says protein chemist Shu Chen, who directs the Center's lab, "when no one wanted to deal with the crisis." [88]

Andrew Kimbrell, the director of the Center for Food Safety, a D.C.-based public interest group, writes, "Given what we know now, it is unconscionable that the CDC is not strictly monitoring these diseases." [89] Given the presence of Mad Cow disease in the U.S., we need to immediately enact uniform active CJD surveillance on a national level, provide adequate funding not only for autopsies but also for the shipment of bodies, and require mandatory reporting of the disease in all 50 states. In Britain, even feline spongiform encephalopathy, the cat version of Mad Cow disease, is an officially notifiable illness. "No one has looked for CJD systematically in the U.S.," notes NIH medical director Paul Brown. "Ever." [90]

The animal agriculture industries continue to risk public safety, and the government seems to protect the industries' narrow business interests more than it protects its own citizens. Internal USDA documents retrieved through the Freedom of Information Act show that our government did indeed consider a number of precautionary measures as far back as 1991 to protect the American public from Mad Cow disease. According to one such document, however, the USDA explained that the "disadvantage" of these measures was that "the cost to the livestock and rendering industries would be substantial." [91]

Plant sources of protein for farm animals can cost up to 30% more than cattle remains. [92] The Cattlemen's Association admitted a decade ago that animal agribusiness could indeed find economically feasible alternatives to feeding slaughterhouse waste to other animals, but that they did not want to set a precedent of being ruled by "activists." [93]

Is it a coincidence that USDA Secretary Veneman chose Dale Moore, former chief lobbyist for the National Cattlemen's Beef Association, as her chief of staff? [94] Or Alison Harrison, former director of public relations for the Cattlemen's Association, as her official spokeswoman? [95] Or that one of the new Mad Cow committee appointees is William Hueston, who was paid by the beef industry to testify against Oprah Winfrey in hopes of convicting her of beef "disparagement"? [96] After a similar conflict of interest unfolded in Britain, their entire Ministry of Agriculture was dissolved and an independent Food Safety Agency was created, whose sole responsibility is to protect the public's health. Until we learn from Britain's lesson, and until the USDA stops treating this as a PR problem to be managed instead of a serious global threat, [97] millions of Americans will remain at risk.

For updates on this evolving crisis, visit the OCA Mad Cow page or send a blank email to [DrGregerMadCowUpdates-subscribe@lists.riseup.net](mailto:DrGregerMadCowUpdates-subscribe@lists.riseup.net)

For background on this important issue, read the excellent book *Mad Cow U.S.A.*, the full text of which is available free online at <http://www.prwatch.org>, or my report *U.S. Violates WHO Guidelines for Mad Cow Disease*.



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