INFECTIOUS DISEASES

Is the U.S. Doing Enough to Prevent Mad Cow Disease?

U.S. officials say they're taking every reasonable step to keep mad cow disease out. But critics still see chinks in the country's armor

On a cold spring morning, when the hills in East Warren, Vermont, were covered with a fresh pack of snow, the Faillace family lost its livelihood. It happened in a government action that—if you hear Larry Faillace recount it—was every bit as dramatic as the one that wrenched Elián González from his Miami relatives last year. At 5:30 a.m. on 23 March, says Faillace, armed federal agents in flak jackets entered the family farm and ordered his three children to stop feeding the sheep. Shortly after, an enormous truck pulled up, and U.S. Department of Agriculture (USDA) agents began loading all of the Faillaces' 126 sheep. A few hours later, the truck was gone, leaving the family, the town, and several dozen protesters behind in anger and shock.

The early morning raid is perhaps the most dramatic example of the U.S. government's efforts to keep "mad cow disease," or bovine spongiform encephalopathy (BSE), out of the country. USDA suspected that the sheep, which the Faillaces had imported from Belgium and the Netherlands in 1996, were infected with a sheep version of BSE. So they took no chances: The entire herd was destroyed days after the animals were seized.

To prevent a BSE outbreak, USDA, the Food and Drug Administration (FDA), the U.S. Customs Service, and other government agencies have put in place a long list of safeguards—from barricading the borders to analyzing brains of people suspected of having died from the human form of mad cow disease, called variant Creutzfeldt-Jakob disease (vCJD). Yet public interest groups and others have long argued that the government's response has been too little, too late. Because of this lax response, the critics say, the disease may well be among us. And if it is, the government is not vigilant enough to detect it, they warn, nor tough enough on the meat industry to keep it out of the human food chain.

Government agencies say they've taken "aggressive" measures to prevent the disease, and many scientists agree. They admit that the precautions are not failsafe and that the disease could emerge in the country—but say the risk is vanishingly small. Even so, the concerns are reverberating on Capitol Hill, where House and Senate committees have summoned officials to discuss the risks. Senator Richard Durbin (D–IL) announced recently that he will introduce a bill that would beef up border inspections and other controls to keep BSE out of the food chain.

But underlying the argument is a broader question: How much prevention is enough? Scientists point out that the U.S. defense against BSE consists of multiple tiers, each of which would have to break down for an outbreak to occur. Although the risk could be reduced further, the necessary control measures would become increasingly costly and draconian. "You don't go spending half the budget to reduce the risk to zero," says Paul Brown, a senior scientist at the National Institute of Neurological Disorders and Stroke (NINDS) in Bethesda, Maryland, "especially in view of much more serious public health problems that afflict us."

Sad day. Larry and Linda Faillace of Vermont watch as their sheep are being seized.

CREDITS: TOBY TALBOT/AP

Multitiered containment

BSE is one of the so-called transmissible spongiform encephalopathies (TSEs), a mysterious class of fatal brain diseases. Scientists are still debating their etiology, but the leading theory is that they're caused by abnormal forms of proteins, called prions. Several TSEs have the scary ability to jump the species barrier; in Britain, for instance, 99 people are known to have died or are presumed to be dying of vCJD, most likely contracted after eating meat products from infected cattle. Epidemiologists expect more cases in the United Kingdom, but they're not sure how many; there could be tens of thousands.

In the United States, the first line of defense is to block entry of the BSE agent, and most people agree that the government has been thorough. As early as 1989, USDA banned the importation of all ruminants (cattle, goats, and sheep) and many animal products from the United Kingdom and other countries with BSE. In 1997, when BSE cases started showing up in several other countries, that ban was extended to all of Europe. The 500 or so animals that were imported from those countries before 1997—such as the Vermont sheep—have almost all been quarantined or purchased and killed.

But closed borders offer no guarantees. Researchers still don't know how BSE arose in Britain, but whatever the process, it could happen here, too. One prominent theory is that the agent that causes scrapie, a TSE in sheep, crossed the species barrier and ignited the cattle epidemic in Britain—specifically, when cattle were fed meal that contained infected sheep tissue. That practice is now banned in the United States, making such a scenario unlikely.

But BSE could also arise out of nowhere. Each year about one in every million humans worldwide gets CJD spontaneously, and it's possible that the same happens in cattle—or indeed all mammals. Last year a U.K. panel chaired by Lord Andrew Phillips supported the theory that such a "sporadic" case may have started the British outbreak.

Work by Richard Marsh, a veterinary virologist at the University of Wisconsin, Madison, who died in 1997, suggests that sporadic cases of a cattle TSE may have already arisen in the United States. Five times between 1947 and 1985, a disease called transmissible mink

encephalopathy decimated populations on U.S. mink farms. After investigating the last outbreak, Marsh concluded that cow carcasses fed to the mink were the most likely source of the disease agent. He speculated that at least one of the cows must have had a TSE.

Another potential source of BSE is a homegrown prion disease that afflicts deer and elk. Conceivably, this ailment, called chronic wasting disease, could jump to cattle or sicken people who eat infected venison (see sidebar).

But would anybody have noticed if the United States had a couple of cases of BSE? Probably not, say some critics. USDA now tests some 50 suspect cows a week. The test program pales in comparison to the massive effort started last year in the European Union, where every cow over 30 months old is tested after slaughtering. The United States should do something similar, says Thomas Pringle, a molecular biologist with the Sperling Biomedical Foundation in Eugene, Oregon, who maintains a Web site about BSE. "You can try all these containment measures, but at the end of the day the question is: How much BSE do you have?" he says. "The way to find out is to run hundreds of thousands of tests."

Testing at that level would be silly, replies Linda Detwiler, a senior staff veterinarian at the USDA, because BSE has never been found in the country. Even so, she says, this year the agency will double the number of tests it performs.

Cows eating cows

Even if a cow got BSE and it went undetected, that wouldn't spell doom for the rest of the nation's livestock. The only plausible way for an outbreak to occur would be if that cow were fed to other cows, thereby passing on the infectious agent. For decades, cows did eat other cows, when they were fed meat-and-bone meal, a protein concoction produced by milling and boiling (or "rendering") the carcasses of, say, sick farm animals, road kill, and dead pets. The epidemic in Britain is believed to have been fueled after infected cattle were recycled on a large scale.

But this route is now cut off in the United States, at least in theory: FDA banned feeding most mammalian protein to all ruminants in 1997. Those same proteins can still be fed to pigs and poultry, however, so FDA has ordered rendering plants and feed mills to prevent commingling of the two types of feed. Enforcing this separation has proven difficult, however. A March 2001 FDA inspection report showed that about one in seven feed mills and rendering plants didn't have adequate procedures to prevent commingling; many haven't been inspected yet.

Indeed, Purina Mills in Texas discovered in January that a new employee had mistakenly let cattle protein slip into a batch of cow feed. After 1222 animals that had been given the suspect feed were quarantined, Purina paid for the entire herd to be destroyed. "Who knows how many other cases have been swept under the rug?" asks Peter Lurie, a researcher at Public Citizen, a consumer watchdog group in Washington, D.C., and a member of FDA's advisory committee on TSEs. Lurie would like to see the FDA get much tougher on the feed industry.

Although that may not be a bad idea, others say, the current situation is hardly a recipe for disaster. Suppose a BSE-infected animal did end up in cattle feed, says NINDS's Brown, and a few cows became infected and went to the slaughterhouse undiagnosed. For the outbreak to continue, they would have to be rendered themselves and mistakenly turned into cattle feed again. "A regulatory breakdown of this magnitude is virtually impossible," Brown wrote recently in *Emerging Infectious Diseases*. Similarly, Will Hueston, a veterinary epidemiologist at the University of Maryland, College Park, says the risk of even a single case of BSE is "pretty darn small."

The chance that humans might get vCJD from eating infected cattle is even smaller. But here, too, critics see loopholes that they would like closed. European countries now require brains and spinal cords to be removed from a carcass directly after slaughter; no such safeguard exists in the United States.

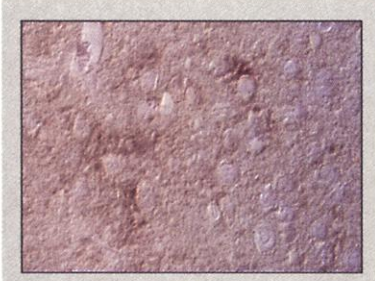
Another route of infection could come from the local health food store. In 1994, Congress deregulated dietary supplements. Many of these contain animal parts—including brain tissue. Although the FDA has asked manufacturers not to use such materials from countries known to have BSE, it can't ensure that no cow brains make it in, says Lurie. Supplements are a problem as long as FDA lacks jurisdiction over them, agrees Brown, who chaired FDA's advisory panel on TSEs until last January.

How much is enough?

In the end, nobody disputes that more can be done to prevent BSE; the question is how much the country is willing to invest. For instance, banning the use of all animal proteins in livestock feed would all but eliminate any risk, says Brown. But it would be the end of the $2.5 billion rendering industry, and it might make meat more expensive, he says.

In his recent commentary, Brown summed up seven holes in the safety net that critics are sure to pounce on if a BSE case were ever to occur. Even so, Brown thinks the current safeguards earn "high marks." Rather than closing each and every hole, he suggests that the money could be better spent on other public health issues, such as diabetes, hypertension, or car accidents.

Mule deer



CJD brain

PRION DISEASES FOUND IN THE U.S.

Scrapie. First case diagnosed in 1947; now 40 to 60 infected sheep farms are reported per year.

Chronic wasting disease (CWD). Afflicts wild deer and elk in Colorado, Wyoming, and Nebraska; also found on elk farms in other states and in Canada.

Transmissible mink encephalopathy (TME). Five outbreaks reported at mink farms in Wisconsin, Minnesota, and Idaho between 1947 and 1985.

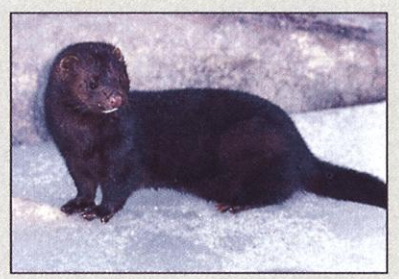
Creutzfeldt-Jakob disease (CJD). An estimated 250 to 300 cases per year; about 85% "sporadic," 15% genetic.

Gerstmann-Sträussler-Scheinker disease and fatal familial insomnia. Two extremely rare genetic human diseases.

NOT FOUND IN THE U.S.

Bovine spongiform encephalopathy (BSE), or "mad cow disease"

Variant Creutzfeldt-Jakob disease (vCJD), the human form of BSE



Mink

CREDITS: (CLOCKWISE FROM TOP) J. H. ROBINSON/PHOTO RESEARCHERS; PIERLUIGI GAMBETTI; STEPHEN J. KRASEMANN/PHOTO RESEARCHERS

America's Own Prion Disease?

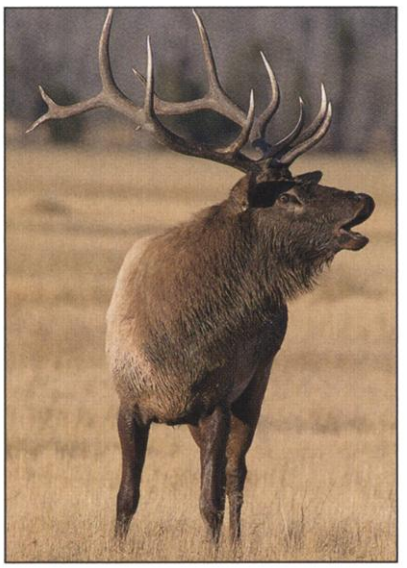
Hunting just isn't what it used to be—at least not in certain parts of Colorado and Wyoming. Not only do hunters have to wear orange jackets to avoid being shot; some scientists now worry that their trophy animals may kill them.

Since the 1960s, deer and elk in these states have been known to suffer from a chronic wasting disease (CWD), a fatal condition whose first symptoms are emaciation and abnormal behavior. In the 1970s, CWD was discovered to belong to the so-called transmissible spongiform encephalopathies, like scrapie in sheep and Creutzfeldt-Jakob disease (CJD) in humans. But hunters and others gave it little thought until 1996, when Britain concluded that a similar disease affecting cattle, bovine spongiform encephalopathy (BSE), could cause a new and fatal form of CJD in humans, vCJD. If eating beef can give you vCJD, then what about venison? So far, there's no good answer.

CWD, which affects white-tailed deer, mule deer, and elk, was first diagnosed in captive deer in research centers, although scientists don't know if it originated there. Today, it occurs in the wild throughout northern Colorado and southeastern Wyoming; two cases have been found just across the border in Nebraska and one in the Canadian province of Saskatchewan. It has also hit elk farms in several U.S. states and in Saskatchewan. Unlike BSE, CWD doesn't seem to be food-borne; instead, it's probably transmitted through saliva or feces, says Elizabeth Williams, who studies the disease at the University of Wyoming in Laramie. Williams estimates that 4% to 6% of mule deer and less than 1% of free-ranging elk in the endemic areas are infected. So far, CWD hasn't been found elsewhere in the world—but then, few countries have been looking.

One reason Williams and other researchers worry about CWD is that it might trigger a new animal disease. For instance, cattle in the endemic areas could pick it up and develop a new strain of BSE. Three years ago, Williams and colleagues at the Colorado Division of Wildlife and the National Animal Disease Center in Ames, Iowa, started experiments to determine the likelihood of that scenario by exposing cattle to CWD. The first results, although not clear-cut, are worrisome. The team injected brain tissue from infected deer into the brains of 13 cows. Last January they reported that three of the 13 had become sick with a disease resembling BSE and had to be killed. Admittedly, injecting material into the brain is an unnatural way to induce an infection, so perhaps infection doesn't occur outside the lab, says Williams. So far, cattle that were fed infected brains or were housed with infected deer are still healthy, she notes. The researchers plan to monitor the animals for at least another 7 years.

The direct risk to humans from eating meat from infected deer or elk is equally puzzling. Recently, researchers at the Centers for Disease Control and Prevention (CDC) in Atlanta have taken a close look at three people who died from CJD and had a history of eating venison. One, a woman whose father used to hunt in Maine, ate the meat when she was a young child; the other two were hunters from Utah and Oklahoma—where CWD has never been found—who frequently dined on venison. A worrying common denominator is that all three were under 30, says CDC epidemiologist Ermias Belay. Such cases are "extremely rare," he says; most CJD victims are over 55. In Britain, however, vCJD strikes people at an average age of 29.



Health hazard? Some elk carry a BSE-like disease.

Although suggestive, this small sample is a long way from proving a link between CWD and CJD, says Belay. For one, vCJD patients all share a typical brain pathology that sets them apart from classic CJD patients. The brains of the three venison eaters, however, suggested classic forms of CJD, says Pierluigi Gambetti of the National Prion Disease Pathology Center in Cleveland, Ohio, who studied their brains. Furthermore, the three each had different subtypes of the disease. But most important, when Belay interviewed their relatives and physicians, he couldn't find hard evidence that they ever ate meat from any of the endemic areas.

Still, Belay says he can't exclude the link either—and that's why CDC plans to keep tabs on future cases of CJD. In the meantime, health officials in Colorado and Wyoming are advising hunters not to shoot or handle sick-looking deer and to avoid eating brain, spinal cord, and other possibly infected tissues.

Even the next best thing to eating your own prey—taking the head home as a trophy—might be risky, says molecular biologist Thomas Pringle of the Sperling Biomedical Foundation in Eugene, Oregon. To sever a head, hunters have to cut right through the spinal cord. "It's potential biosafety level 3 material," says Pringle. "I'm not sure Joe Sixpack should be throwing that in the back of his pickup truck."

–M.E.

George Gray, a risk analyst at Harvard School of Public Health in Boston, agrees. "Every bit of attention and effort we put into [BSE] takes away from something else," says Gray. "And I think there are considerably bigger risks out there in the food supply." An estimated 5000 people a year die from microbial contamination in food alone—many more than would be harmed by BSE in any plausible scenario, he asserts. At USDA's request, Gray is studying the risks of BSE and related diseases in the United States. The study, which will guide future policy, will be presented to the agency within the next 2 months.

Lurie dismisses such comparisons. "By that argument, we should not worry about microbial contamination because many more people die from cancer," he says. Although the risk may be low, he says, the worst-case scenario would have such disastrous public health and economic consequences that extreme caution is warranted.

That's the argument that led USDA to kill the Faillaces' sheep and another nearby flock, says Detwiler. Tests carried out last year on four slaughtered animals showed signs of a BSE-like disease, although it wasn't clear whether it was scrapie or a sheep version of BSE. Sheep have been infected with BSE in the lab, but no natural cases have been found in the world. If the Vermont sheep did have a form of BSE, they would be the first. Better to err on the side of caution, says Detwiler, than for the United States to have that dubious honor.

The Faillaces, who fought the seizure in a long legal battle, claim the sheep were healthy and the tests were sloppy. Additional tests of the Faillaces' sheep are now being performed at the National Animal Disease center in Ames, Iowa. The results, says Detwiler, will be available in a few months—about the time that Gray's risk assessment is due.

–MARTIN ENSERINK

CREDIT: C. K. LORENZ/PHOTO RESEARCHERS