Mad Cow Disease: Uncertainty Rules

Following an anti-beef shopping panic in Britain, Europe and the United States are taking steps to guard against a brain disease of sheep that has been afflicting British cattle

A BRITISH SUNDAY LUNCH without roast beef? Unthinkable, but increasingly true. Sales of beef are about 20% down since the beginning of the year, and in the supermarkets even drastic specials are failing to shift the meat into shoppers' carts. The reason: fear of bovine spongiform encephalopathy, or BSE, a disease that has already claimed the lives of more than 18,000 cattle in Britain since it was first diagnosed in November 1986.

The disease came to cattle from sheep that were ground up to make cattle feed. And the panic in Britain is being driven by the possibility that it could now jump to people who eat beef. Most scientists believe the chance of that happening is small, but disagree over the exact amount of risk—which leaves governments in the difficult position of having to deal with this scientific uncertainty in drawing up public policy. The result in Britain has been a public relations disaster and, some critics charge, inadequate public health measures.

So far, the disease has turned up outside the United Kingdom only in cows exported from Britain. But other countries are now drafting their own policies on BSE, and they are hoping to avoid Britain's experience. The rest of Europe, for example, is taking no chances of an outbreak and is moving toward a ban on sheep-derived protein in cattle feed. The United States is prepared to wait and see, although it has banned the import of live British ruminants and feed made from their offal.

BSE is the cow's version of scrapie, a long-standing disease of sheep endemic in Europe and the United States. It is caused by a so-called unconventional virus-like agent. The main symptoms are abnormal gait and behavior, but there is no diagnostic test, and the disease can be confirmed only in a postmortem by the presence of the holes it creates in the brain. It also has a very long incubation time, so that it can be months or years between infection and symptoms.

Britain may have been at greater risk than other countries when the scrapie agent unexpectedly moved from sheep to cattle, apparently sometime in the 1980s. Feed practices in Britain differ from those in other countries, and British farmers raise a large number of sheep—and hence scrapie-infected carcasses—relative to cattle.

But it was not the first time spongiform encephalopathies had crossed a species barrier. In the United States, mink have long been known to have contracted the disease from sheep, and elk have also shown the characteristic spongy brain. And in Britain, a BSE-like disease turned up in May in a pet cat. Officials from Britain's Ministry of Agriculture Fisheries and Food (MAFF) played down the episode, saying it was unlikely to have come from pet food. Cats, they argued, were entitled to their own spongiform encephalopathy; perhaps no one had seen it before because no one had looked. But Hugh Fraser, an authority on scrapie at the Medical Research Council's neuropathology unit in Edinburgh, disagrees. "Overwhelmingly the most likely explanation," says Fraser, is that the disease came from cat food. "It's wishful thinking to suggest otherwise." (Pet food includes sheep and cattle not fit for human consumption, although manufacturers say they no longer include those organs most likely to be contaminated by scrapie.)

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Burning issue. *Health officials in Britain have ordered the destruction of thousands of infected cattle—like this one in Devon—since an unusual brain disease appeared in 1986. Beef sales have plummeted.*

The limits of cross-species infectivity are being probed in several medical centers, but results are a long time coming. Just this week, however, MAFF scientists announced they had infected a pig by injecting ground-up brains from a cow directly into the pig's brain.

From a public health standpoint, of course, the key question is: Could BSE jump yet another species barrier and become HSE—human spongiform encephalopathy? There already is one scrapie-like human spongiform encephalopathy, known as Creutzfeldt-Jacob disease. It crops up very rarely—roughly one case per million people per year—and is no more common among people who eat plenty of sheep, including sheep's brains. Optimists take comfort from this. "I don't think there's any risk to humans," says Richard Marsh, professor of veterinary science at the University of Wisconsin at Madison and an expert on scrapie. Nevertheless, pessimists, such as Heino Diringer, who studies unconventional viruses at the Robert Koch Institute in Berlin, say it is irrelevant. "We do not know if [the disease] will change in cattle. We cannot assume that because [eating] sheep is relatively safe, ergo beef is safe."

Faced with these conflicting scientific assessments, Britain has taken steps to minimize the risk to the public. The British government in 1988 banned the use of sheep in cattle cake and late last year it prohibited the sale of certain organ meats, notably brains and sweetbreads (thymus), for human food. To begin with, it paid farmers half the market value of sick cows that proved to have BSE after slaughter. Later, it agreed to full compensation because of public fears that farmers were being tempted to sell suspect cows before they became obviously ill, thus spreading BSE into the food chain. And this week, following the laboratory infection of a pig, MAFF banned the use of cattle offal that is already prohibited from human consumption from being used in pig feed and pet food.

But these moves have drawn fire because

there has been no attempt to keep tabs on the offspring of sick cows, and meal made from infected sheep is still being fed to pigs and poultry and exported to Third World countries. The government defends its decision to permit sheep protein in pig and poultry feed because "pigs are not at risk because they are natural scavengers (and have evolved defenses against pathogens) and poultry are not at risk because there is an enormous zoological divide between cattle and poultry." Experts disagree. "That's so much baloney," says Fraser. Although Fraser himself thinks that the risk to humans is small, he notes that "pigs get anthrax and botulism and half a dozen other diseases as a result of exposure in their diet."

Since farmers in many countries continue to be allowed to feed animal protein—including that from scrapie-infected sheep—to cattle, the fact that no BSE epidemic has appeared outside Britain is seen as a good sign that the disease will not spread widely. In France, for example, despite reports of a "mystery illness" misdiagnosed as rabies (and still unidentified), there is no BSE. "I don't think we have the evidence," says Michel Brahic, a virologist at the Pasteur Institute in Paris. But, he adds, there has been no epidemiological survey.

The French Ministry of Agriculture has, however, started a national monitoring program centered at the national Bovine Disease Laboratory in Lyon. Mark Savey, the virologist and veterinarian who heads the lab, says he would be "very happy to publish the first case of BSE in France," but so far has not found one. "Perhaps tomorrow," says Savey. He doubts that France will suffer a massive outbreak, mostly because French dairy farmers prefer soya-based feed for their cows. "One day we will have one or two cases," he told *Science*, but they will be in imported cattle or those fed rations from Britain. Exports of feed from Britain to France jumped between July 1988, when sheep protein was banned in Britain, and December 1989, when the French acted to ban its import. French farmers are still currently allowed to feed French sheep protein to cattle, although there are plans in the ministry to restrict ruminant protein to pigs and poultry. Savey accepts that this is a risk: "No one can say what will happen."

Dairy farmers in the United States continue to feed sheep protein to cattle, and at present there is no legislation limiting sources of protein in feed, nor plans to introduce any. However, the feed industry did stop using fallen and sick sheep as a protein source in December 1989, prompted largely by fears of legal liability should BSE break out.

EM Unit, Weybridge/Science Photo Library

Rogue agent. *Damage to cows' brains resembles "slow virus" diseases like scrapie in sheep and Creutzfeldt-Jacob disease in humans.*

Yet there are some who see signs of trouble. "I think we have a BSE-like infection here already," says Wisconsin's Marsh, referring to his investigations of outbreaks of spongiform encephalopathies on mink farms. Marsh is quick to point out, however, that cattle in the United States are less likely to suffer from BSE because feeding practices differ from those in the United Kingdom. In Britain, dairy cows receive animal protein from their first year and are kept alive longer. Their American cousins do not get animal feed until their second year and are often slaughtered by the age of five. Because BSE has a long incubation time, this difference could limit its spread in the United States. Nevertheless, says Marsh, "it would be nice if the [U.S. Department of Agriculture (USDA)] would discontinue feeding ruminant protein to calves. . . . I guess they're going to wait until they have the first case—which will be too late."

"To change the whole structure of the feed industry here we want more evidence," says Lonnie King, veterinarian and deputy administrator for veterinary services at the USDA. King concedes that the epidemiological evidence that BSE came from scrapie is "pretty compelling." But those who worry about it infecting U.S. cattle, he says, make two unproven assumptions: that what happened in Britain to enable scrapie to jump from sheep to cattle will happen in the United States and that the scrapie agent is the same in Britain and America. He sees no reason for alarm and expects no change in U.S. policy until certain "data gaps" are plugged. "We'll have to wait and see."

While other countries ponder the public health implications of BSE, no country other than Britain has yet had to deal with a secondary epidemic related to the disease: public panic. In banning from human food any organs known to harbor BSE, the British government followed the advice of its scientists in trying to make an unlikely event—transmission of the scrapie agent to people—even less likely. But, instead of honestly reflecting scientific uncertainties, MAFF has always insisted that British beef is "completely safe." Many Britons—including scientists advising MAFF—simply don't believe that.

Daphne Barrett, a director of the London public relations company Infoplan, describes MAFF's handling of BSE as "inept." Barrett masterminded Perrier's response to the benzene found in its bottled water, which "was never a risk to health. . . . But it was a risk to the brand." Just so with BSE. "It may be a slight risk to human health," says Barrett, but "it is a far greater risk to MAFF's credibility." She says MAFF should have responded more quickly and with more responsibility, informing people rather than nannying them.

At issue is public faith in government. Scientists working with spongiform encephalopathies are disappointed, though not surprised, by the desire for certainty. "It's difficult to explain to people that we don't know some things," says Savey. "If you are an optimist, you say the risks are so small they do not matter. If you are a pessimist, you say we must act as if the risk were big. But it is not a scientific position." Scrapie has existed in sheep for at least 200 years without spreading to people or worrying consumers unduly. Its leap to cattle provoked panic. That response bothers the scientists.

"Nobody worries about scrapie, so I don't see why we should worry about BSE," says Michel Brahic at the Pasteur Institute. He pauses. "Or maybe we should. If we worry about BSE then we should worry about scrapie. There is a lack of logic."

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