

## An issue comes to a head

***North Americans haven't tested rigorously enough for mad-cow disease. Have we underestimated its prevalence in our midst? demands science writer ANDREW NIKIFORUK***

One mad cow is messy; two are messier. And in the next few months, if and when North American regulators actually begin to gather some real science by testing thousands of cows, the picture will likely get even dirtier.

Many experts on bovine spongiform encephalopathy now suspect that BSE/mad cow has been in North America for at least a decade, that the beef industry and regulators have fought proper regulation from day one, that the current surveillance system is a don't-look-don't-find model and that the public-health risk from contaminated meat could be greater than most are prepared to admit.

"We have to take some serious actions," notes Yale University pathologist and mad-cow expert Laura Manuelidis. "It's here now, and we have to do something about it."

Let's begin with what's known. Mad cow is just one of many transmissible spongiform encephalopathies (TSE) that put holes in the brains of mammals, including elk and deer. The human disease is called Creutzfeldt-Jakob disease (CJD) and it bears so many similarities to Alzheimer's disease that doctors commonly misdiagnose it (more on that alarming connection later).

The infectious agent responsible for the current mayhem may be an abnormal protein or prion (the dominant view) or an unconventional virus. Environmental pollutants capable of altering the amount of key trace metals in animals may also play an unexplained role in the progress of this enigmatic disease. Scientists generally agree that TSEs are spread by bone, blood and nerve tissue in contaminated feed. Or by inoculation, which explains why the Canadian government has banned U.S. animal vaccines derived from rendered cows. More than one strain exists, and not every strain looks like classic British BSE.

The first North American case of mad cow probably appeared in 1985, on a Wisconsin mink farm. That's when Richard Marsh, a veterinary pathologist at the University of Wisconsin, discovered that mink fed "downer cattle" (technically any cow that has difficulty walking) from local dairy farms, went crazy and died. Prof. Marsh took samples of these mink brains and inoculated and fed them to bull calves. Each bull developed holes in the brain. He then fed infected cattle-bits back to mink, which developed more spongy brains.

His conclusion: "There must be an unrecognized scrapie-like disease (BSE-like agent) in cattle in the United States."

This peculiar strain of BSE didn't have the same clinical symptoms of classic British BSE. Instead of acting aggressively, these infected cows behaved sleepily, like downer cattle. Inspectors looking for drooling or rabid cows as a sign of mad-cow infection would miss this disease.

Even the brain pathologies were different. Canada's two detected cases of mad cow last year showed no symptoms of madness at all.

Before Prof. Marsh died in 1997, he pressed for a ban on feeding cattle-bits to cattle, and he warned that waiting for the first case of mad cow was like closing the barn door after the proverbial horse had run off. "With a disease having a three-to-eight-year incubation period, thousands of animals would be exposed before we recognized the problem and, if that happens, we will be in a decade of turmoil," he wrote.

Prof. Marsh was vilified and denigrated by the U.S. cattle industry for his work. His grant proposals to test more cattle were routinely turned down by government. When a consumer's group sued the U.S. government last year for not banning downer cows from the food chain, the U.S. government, like the industry, retorted: "BSE has never been found in the country's livestock," and said that the threat was "not real or immediate." It was as if Richard Marsh's work never existed. Speaking to U.S. journalist John Stauber, co-author of *Mad Cow USA*, Prof. Marsh once confessed: "By issuing warnings to industry, I thought industry would do the right thing. How could I have been so wrong?"

Many BSE experts now regard Prof. Marsh's work as prescient. "It was good work. It was ignored unfairly, and it was years ahead of its time," notes David Westaway, a molecular biologist and prion specialist at the Centre for Research in Neurodegenerative Diseases at the University of Toronto. Dr. Westaway, a cautious conservative scientist like Prof. Marsh, says the current system is anything but science-based. He notes that U.S. Cattlemen's Association has been "virulently" against testing, and that they have influenced Canadian policy.

The tests aren't perfect, and are mostly designed to pick up the tail end of an infection. "But tests are better than no testing," adds Prof. Westaway. "We have to get the prevalence. It's unlikely we have an enormous epidemic -- but we don't know what's out there."

Prof. Westaway's plea for more testing has gone unanswered for three years. No one from the Canadian Food and Inspection Agency has ever called him about the issue. Nor has anyone investigated why Alberta's animal-disease surveillance system, one of the best in North America, was drastically downsized after an imported cow from England with BSE was discovered in 1993.

There may be a bigger public-health concern out there. In 1989, Laura Manuelidis and colleagues at Yale University performed autopsies on the brains of Alzheimer's patients and found that 13 per cent of the patients actually suffered from Creutzfeldt-Jakob disease -- the human form of mad cow.

A University of Pittsburg study made similar findings. Until then, most scientists assumed that CJD only occurred in one in a million people. What the results could well mean is that "at least some people diagnosed with Alzheimer's have CJD," says Ms. Manuelidis. Instead of the official caseload of approximately 30 CJD cases a year, Canada, which has 364,000 cases of Alzheimer's and related dementias, just might already have much higher numbers of CJD.

So things are about to get messy. That is not to say that a higher incidence of CJD is necessarily connected to BSE; there could be more sporadic CJD than previously thought, or an infectious prion could be responsible. What we do know is that we have mad cows; we have enigmatic brain-wasting diseases in people; and we don't have much science in between.

*Andrew Nikiforuk, a Calgary journalist, has been writing about the beef industry for a decade. Last year his book *Saboteurs* won the Governor-General's award for non-fiction.*