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EXPLORATIONS



Tempest in a T-Bone?

Researchers go around in circles on mad cow disease

MEATY FEARS.

Recent evidence indicates that mad cow disease can jump the "species barrier" to sheep.

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EXPLORATIONS**

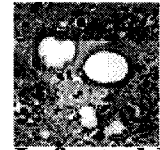
As bovine spongiform encephalopathy, or BSE, it was a mysterious malady that caused brain degeneration among cows. As "mad cow disease" prevalent in British herds, it prompted concerns about unhealthy cattle feeding practices. But this past March, when an advisory committee to the British Parliament declared that BSE might be able to jump the species barrier and cause Creutzfeldt-Jakob disease (CJD), a rare but horrifying brain disorder among humans, it became the stuff of hysterical talk shows. Oprah Winfrey described BSE as "a medical mystery spreading panic across the Atlantic." The true extent of the risk remains murky, however, as researchers know frustratingly little about how BSE works.

So it came as relatively little surprise to most scientists working in the field when the European Union announced on July 23 that BSE can be transmitted from cows to sheep. Indeed, there was already good reason to think that the disorder could travel in the other direction. One of the best guesses for the origin of BSE, which has affected close to 200,000 animals in England and Europe, is that it arose when cows ate feed supplemented with the brains and spinal cords of sheep infected with scrapie. (Strange though it may sound, such use of animal by-products in cattle feed was common practice in the U.K.) Scrapie, a degenerative brain disease, has been known to sheep farmers for at least 250 years; its symptoms are much like those of BSE. Moreover, BSE -- or something remarkably similar to it -- has also appeared in pigs, cats and several zoo animals whose diet was augmented with cow-derived protein.

Then again, researchers probably would not have been surprised if BSE had proven harmless to sheep. Scrapie manifests itself in a number of different strains. The specific version of the disease that is able to infect cows might well be expected to be innocuous to its original host -- after all, the genetic differences between species are usually sufficient to halt the spread of infectious agents. And cows injected with scrapie agent do not develop the typical symptoms of BSE. Such conflicting findings give a sense of the confusion that still surrounds a field in which simple tests of contagion may take three to five years to carry out.

Molecular biologists continue to search for an understanding of BSE's mechanism of action. Even the precise identity of the infectious agent is unknown. Many scientists pin the blame on prions, a peculiar class of aberrant proteins that, unlike familiar pathogens, contain no genetic material at all. But information about how prions cause disease -- or even whether they truly exist -- remains elusive.

Meanwhile, human carnivores wait with apprehension for confirmation that there exists a type of BSE that attacks them. The March declaration by the British Parliament was prompted by reports of a dozen cases of "atypical"



Infected Brain Creutzfeldt-Jakob disease, a deadly neurological ailment whose symptoms resemble those of BSE and scrapie. CJD, which was first recognized about a century ago, primarily kills people over age 45.

Atypical CJD causes a different pattern of brain damage than usual, and the 10 instances thus far reported in the U.K. have occurred among people in their teens and twenties. Responding to the possibility that this variant is a human version of BSE, the British government banned any use of potentially infectious cow, sheep and goat parts at the end of May.

Both BSE and atypical CJD appear to be spreading. Although the annual number of new "mad cow" cases in the U.K. is down by more than half from its peak of 37,000 in 1992, farmers in continental Europe, who had once thought that their herds remained untouched, have now reported several dozen cases of BSE. B.E.C. Schreuder of the DLO-Institute for Animal Science and Health in Lelystad, the Netherlands, estimates the true number to be more like a couple thousand cases. Young adults with brain lesions almost identical to those of their British counterparts have reportedly died in both France and Germany. Even more troubling are anecdotal reports of an increasing prevalence of CJD in the U.S., where BSE has yet to be confirmed in any animals. One report from Oregon counted 50 cases of CJD during a five-year period among a population in which only 12 or 13 cases would have been expected.

There is no direct evidence, however, that the new human disease -- if there even is one -- is actually linked to the British epidemic of BSE. The agent responsible for BSE has yet to be identified with certainty, so detecting it in humans is problematic. Furthermore, the amounts of beef that the CJD victims consumed varied wildly, and cooking the meat may or may not affect the spread of BSE. And the number of atypical CJD cases is still a small fraction of the number of typical CJD deaths: researchers examined more than 100 diseased brains in the process of confirming the dozen victims. Meanwhile, no one has any idea whether the kinds of cooking that various cuts of beef undergo might affect their putative infectivity.

Most references quote the annual incidence of "typical" CJD at one case per million people each year, but no one knows the real

numbers. In the U.S, for example, there is no active surveillance for the disease. Statisticians simply count up the number of times it appears on death certificates -- about 250 in an average year. Because CJD can only be diagnosed with certainty by sectioning the brain after death (its symptoms are broadly similar to those of Alzheimer's disease), there is a significant chance for undercounting or overcounting, depending on how officiously doctors fill out their former patients' paperwork. Since the late 1980s, neurologists everywhere have had mad cow disease on the brain, and it takes only a tiny increase in the number of people diagnosed with CJD to yield a big jump in the statistics.

The British government established a special reporting system for CJD in 1990, after the potential dangers of BSE became apparent, but the resulting disease statistics will be difficult to compare with those from other countries or with earlier figures from the U.K. Until researchers unravel the mechanism of the agent behind BSE and related diseases in other animal -- or, more grimly, until far more people start dying -- it will be impossible to determine whether meat-eating humans are at the same risk as meat-eating cows. In the meantime, hold off on the breaded calves' brains.

--Paul Wallich, staff writer

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[BSE fact sheet from the World Health Organization](#)

[BSE information from the U.S. Department of Agriculture](#)

[Historical overview of BSE](#)

[BSE news and resources](#)

[CJD news and resources](#)

[Overview of transmissible spongiform encephalopathies](#)

["The Prion Diseases,"](#) by Stanley B. Prusiner; Scientific American, January 1995
