Debate over 'mad cow' disease resurfaces in UK

Fears that bovine spongiform encephalopathy (BSE), also known as 'mad cow' disease, could cause a related form of dementia in people who eat contaminated beef have been resurrected in the UK by recent reports that two British teenagers with no known risk factors have developed Creutzfeldt–Jakob disease (CJD). One of the teenagers has since died of the disease, which is exceptionally rare in people under 30.

Scientists say they hope to be able to establish whether the agent that causes BSE is the cause of the two cases. The Spongiform Encephalopathy Advisory Committee, a panel of experts set up in 1990 to advise the Ministry of Agriculture and the Department of Health, said it would be essential to study the patients in great detail. Professor John Pattison, the committee's chairman, said the committee was due to meet on 23 November and would be advising the two Government departments on what experimental work should be undertaken.

In early October, shortly before the publication of the reports about the teenagers, the committee said there was no evidence for an emerging CJD epidemic. The committee, which was responding to the fourth annual report of the CJD Surveillance Unit in Edinburgh, said it was satisfied that adequate controls were in place to protect public health with regard to CJD.

There were 55 cases of CJD in the UK last year, up from 42 cases the year before. The unit is conducting a long-term study to identify any changes in the pattern of CJD since the emergence of BSE. In October, health department officials said that "to date, no such changes have been detected."

Commenting on the unit's report, the government's Chief Medical Officer, Kenneth Calman, said: "I continue to be satisfied that there is currently no scientific evidence of a link between meat eating and CJD and that beef and other meats are safe to eat."

BSE was first recognized in cattle in the UK in 1985 and is thought to have resulted when cattle were given feed made from the byproducts of sheep infected with the scrapie agent. BSE, scrapie and CJD all cause the brains of affected individuals to become vacuolated or have a spongy appearance hence the name 'spongiform encephalopathies'.

In about 15 percent of cases of CJD, there is a family history of the disease. No one knows what causes the remaining 85 percent of so-called sporadic cases, such as those reported recently in two letters to the *Lancet* (**346**, 1155–1156; 1995).

The first, a girl who was 16 when she first developed neurological problems in March 1994 and who is still alive, was born in the UK of Turkish-Cypriot parents. She had no family history of dementia or similar illnesses. She normally ate lamb but had occasionally eaten corned beef and beefburgers and had eaten cow's brain in Cyprus — where there have been no reports of BSE in about 1989. A brain biopsy in August showed changes typical of CJD. The second case involved an 18-year-old man who began to suffer memory loss before developing dementia. He died after an illness lasting between 9 and 12 months. He had no relevant family history. He used to visit his aunt's farm each year, where he would have been exposed to cows, but no cases of BSE had been reported in this herd. Neither had received treatment with growth hormone derived from human pituitary glands, or undergone tissue grafting or neurosurgical procedures.

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Teenage cases of Creutzfeldt–Jakob disease have sparked renewed fear of possible link with BSE.

John Collinge, head of the Prion Disease Research Group at St Mary's Hospital Medical School in London, and one of the authors of both letters, said it was surprising to see two cases of CJD in teenagers in the UK in the same year. Although it could be simply a coincidence, the recent cases emphasize the need for continued surveillance, he says. If further cases appear, "this may be telling us that there is a new risk factor for Creutzfeldt–Jakob disease."

The BSE agent produces a very characteristic pattern of pathology in the brains of affected animals: For example, the brain stem is severely damaged. This pattern persists even when the agent is passed from, say, cattle to cats to mice.

Collinge wants to find out what range of pathological patterns results when brain tissue from sporadic cases of CJD is inoculated into mice. Because normal mice are not readily susceptible to infection with the agents that cause the spongiform encephalopathies, he will be using transgenic mice which carry a gene that codes for human prion protein — the protein that builds up in the brains of people with CJD. Collinge will compare the patterns with the pattern produced when the BSE agent is inoculated into the mice, and with that produced using brain tissue from the two teenagers with CJD.

"If the sporadic cases of CJD are all caused by one strain and that produces a pattern that is very different from BSE, then it might be possible to determine if individual cases have arisen from BSE," says Collinge. "But if there are multiple strains and some of them cause damage to the brain stem then it may be difficult to tell whether these cases were caused by BSE."

Robert Will, head of the CJD Surveillance Unit, emphasizes that there have been four other reported cases of CJD in teenagers in other countries (in which there were no reports of BSE at the time), as well as some cases in people in their twenties. Nevertheless, he adds, "these cases [in the UK] are very unusual . . . I understand that they are a matter for concern."

Will says the incidence of CJD in other countries in Europe that either do not have BSE, or that have a very low incidence of it, is very similar to that in the UK. One unexpected finding has been a statistical excess of cases of CJD in dairy farmers in the UK. A similar pattern has prevailed in Europe. Yet there have been no cases in the UK among abattoir workers, who are thought to be at relatively high risk. Official figures state that there have been just over 10,000 cases of BSE so far this year in the UK, a drop from almost 24,000 last year.

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