



RESEARCH NOTE

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Mad Cow Disease: Horror or Hysteria?

Bovine Spongiform Encephalopathy (BSE), or 'mad cow disease', was first identified in the UK in November 1986. By July 1993, 100,000 cases of the disease had been confirmed and in 1995 alone, almost 20,000 British cows succumbed to the illness. The afflicted animals become anxious, develop an abnormal gait, suffer muscular spasms and cease eating. Upon autopsy, the animal's brain exhibits characteristic spongy changes in the structure of its nervous tissue with extensive nerve cell degeneration.

BSE is only one of a number of spongiform encephalopathies (SEs) found in many species, including man. In humans, the closest analogue is Creutzfeldt-Jacob disease (CJD), but other human SEs include Gerstmann-Straussler-Scheinker disease, Alpers disease and kuru, this latter disease having afflicted certain tribes in Papua-New Guinea. Sheep can exhibit SE in the form of 'scrapie' while similar illnesses are also found in mink, deer, cats and other animals. All of these conditions are fatal.

Most important is the fact that these illnesses involve some infectious agent and are transmissible. CJD, for example, has been transmitted amongst humans through human growth hormone, corneal transplantation and infected surgical instruments. BSE has been transmitted, in the laboratory, to cattle, mice, sheep

and goats both orally or through the injection of infected tissue.

The Infectious Agent

There is still some doubt as to the infectious agent involved in transmissible SEs. Researchers have not found any evidence of a virus or other organism which multiplies by replicating its DNA. Such organisms could be destroyed in the laboratory by using radiation, heat or chemicals, yet the tissue found in transmissible SEs remains infectious when subjected to these treatments. The agent can even survive temperatures of up to 360 degrees C.

The most likely explanation is that the agent is a 'prion' (proteinaceous infectious particle). This is a slightly modified protein which can convert a normal protein in the brain into a dangerous one simply by altering its physical structure. The immune system does not recognise these changes and does not combat the disease.

Prion diseases appear to arise in three ways. Some of them (e.g. scrapie, Gerstmann-Straussler-Scheinker disease) have a hereditary origin. Even in CJD, up to 15 per cent of cases have a familial connection. Secondly, as is hypothesised in the remaining cases of sporadic CJD, there may be a random mutation of a prion protein which sets in train a chain reaction amongst normal proteins. Thirdly, the agent

may be transmitted via food or tissue.

Prions from one species appear capable of infecting certain other species. It is likely that the closer the molecular structure of the prion protein of different species, the easier it is for cross infection to occur. The BSE prion differs to some extent from the normal protein in humans, so it is difficult to assess whether this species barrier can be overcome.

Cause of BSE

Epidemiological studies in the UK have concluded that the most likely cause of BSE was the feeding of cows with meat and bone meal made from the remains of sheep infected with scrapie. An alternative hypothesis is that the meal contained offal from cows with latent BSE. In the late 1970s, the method of processing carcasses into meal was changed and may have permitted infectious agents to survive in the processed fodder. Since BSE, like all other SEs, can have a long incubation period, this could explain why the first cases of the disease in cows were not detected until 1986.

The UK Government banned the use of such feed for ruminants from July 1988 and one might expect BSE to die out with that generation of cows. Disturbingly, the disease has appeared in many cows born after the ban. A few researchers consider that the disease

may be passed on from mother to calf, but others attribute the phenomenon to the continued use of animal meal, still allowed for pigs and poultry until September 1990, after the ban. If no vertical transmission is involved and if stock feed is the mode of infection, BSE should eventually be eliminated. Reported cases of BSE are beginning to decline.

The Danger to Humans

The UK Government has taken a number of steps to address the BSE problem. Infected cows have been culled and compensation paid to farmers. In 1989, meat processors were banned from using 'specified bovine offal' (brain, spinal cord, spleen, thymus, tonsils and intestines from all cattle over 6 months of age).

In March 1996, a UK Government advisory committee announced that 10 cases of a CJD-like disease had been detected in humans and that it could not rule out the possibility that the disease was linked to BSE. The new disease is an SE but manifests differently to CJD. In cases of CJD, the victims are usually older, tend to die within one year of diagnosis and display mainly neurological symptoms such as muscle spasms, dementia and sensory and visual decay. The ten new cases were much younger, had a prolonged duration of illness and the initial symptoms were mainly psychiatric (depression, anxiety, aggression).

If there is a link between BSE and this new form of CJD and if these victims were infected in the mid 1980s when BSE was in its infancy, a major epidemic of CJD might well occur in the UK. If the incubation period is shorter and they were infected at the height of the BSE infection, the number of victims could be much lower.

Overseas Reaction

The existence of a possible danger to human life stimulated the European Union to impose a worldwide ban on British beef and cattle exports in March 1996. The UK Government has announced a £550 million scheme to prevent beef from cattle most at risk from entering the food chain. From the end of April 1996, all cattle over 30 months old coming onto the market will be purchased and destroyed. The EU will bear 70 per cent of the cost of the package.

Australian Action

There seems little chance of BSE being transmitted to Australia. In July 1988, Australia was one of the first countries to ban the import of cattle, bovine embryos and semen from the UK. A certification scheme is also in place to ensure that no cattle can be imported from the UK via a third country. The importation of stockfeed of animal origin was banned from all countries except NZ prior to 1988. Pharmaceutical products of ruminant origin from BSE or scrapie infected countries can-

not be used for animal use, and in humans only on the advice of the Therapeutic Goods Administrations. In March 1996, the importation of a small number of UK processed food items was also banned. Australia does not import beef from the UK.

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