BOVINE SPONGIFORM ENCEPHALOPATHY
- THE AUSTRALIAN RESPONSE

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BACKGROUND

Bovine spongiform encephalopathy (BSE) is a progressive degenerative disease of the central nervous system of cattle first identified in 1985-86 in the United Kingdom (UK). Affected animals may display behavioural changes such as aggression or nervousness, abnormal posture, poor coordination, reduced milk output, fine muscle twitching and weight loss. Death is inevitable, generally within six months of diagnosis. The incubation period is from two to eight years. In the UK the incidence of BSE increased rapidly and by December 1995 there were 158,271 confirmed cases in an epidemic which appears to have peaked in 1993. BSE has occurred in a number of countries including Switzerland and France, but the UK is the only country with a high incidence.

BSE is one of a group of related diseases described as transmissible spongiform encephalopathies (TSE). These occur in a number of species including sheep and goats (scrapie), mink (transmissible mink encephalopathy), cats (feline spongiform encephalopathy) and also humans (Creutzfeldt-Jakob disease, kuru, Gerstmann-Straussler syndrome and fatal familial insomnia). Scrapie has been endemic in sheep in the UK for at least 200 years and no link between eating sheep meat or offal, including brains, and Creutzfeldt-Jakob disease (CJD) has been established. CJD occurs at the same rate in Australia which is free of scrapie, as in countries where scrapie is endemic, such as the UK, France and the Middle East.

Although the transmissible agent for TSE has not been isolated, the predominant theory holds that the infective agent, classified as a prion, consists principally of a modified form of a host encoded glycoprotein, the prion protein. The agent causes the accumulation of an abnormal proteinase-resistant form of host protein in the brain. The agent is extremely resistant to heat, normal sterilisation processes and chemicals such as formaldehyde and glutaraldehyde. Brain and spinal cord are the primary infected tissue, but pituitary gland, nerves, spleen, adrenal gland, lymph node, thymus, lung, liver and kidney may also be infected.

A 1987 epidemiological study concluded that the only viable hypothesis for the cause of the BSE epidemic was animal food supplements containing ruminant derived meat and bone meal from rendering plants in the UK.
Bovine spongiform encephalopathy

The practice of using processed animal products as a source of protein for cattle had been common for several decades. However, in the late 1970s and early 1980s changes in rendering processes, including lower sterilisation temperatures and the omission of a solvent extraction, rendering processes, including lower sterilisation temperatures, may have allowed the disease - which may have been present at a low level in cattle in the UK before 1986 - to infect a large number of cattle. Alternatively, the inclusion of inadequately inactivated protein derived from scrapie-infected sheep meat, offal and bonemeal, into animal food supplements resulted in cattle being exposed to high doses of the scrapie agent and the agent was then able to break the species barrier.

Cattle products contaminated with BSE continued to be processed through the rendering plants until at least 1988. Control measures implemented in 1988 included a ban on the feeding of ruminant-derived proteins to ruminants including cattle. The decline in incidence of BSE in the UK since 1983 is believed to be related to these measures. Figure 1 depicts the incidence of BSE in the UK.

### FIGURE 1

**Notifications of BSE in the United Kingdom**

![Graph showing notifications of BSE in the UK]

BSE has been most prevalent in Holstein-Friesian dairy cattle, the predominant breed of dairy cattle in the UK. Dairy cattle have a longer life - of seven to ten years - than beef cattle, which are generally slaughtered before three years of age. Given the long incubation period of the disease, it is not surprising that dairy cattle are more likely to exhibit the disease. The meat from these cattle, slaughtered at the end of their milk production, tends to be used in pies, sausages, minces, manufactured meat products and for the production of food additives rather than sold as steak. These are the very products which are more likely to contain offal (such as brain tissue) as an ingredient.

In 1990 a CJD surveillance unit was set up in the UK. The CJD Surveillance Unit recently identified 10 cases of CJD with onsets between February 1994 and October 1995 which displayed a new and specific neuropathological profile. The neuropathological samples from each of the 10 infected people were virtually indistinguishable. They were also significantly younger than is typical, with an average age of 27.8 years. Sporadic CJD in the UK has a mean onset of 65 years. The 10 infected people also displayed a prolonged duration of illness, averaging 13.1 months with a range of 7.5 to 23 months, whereas normally CJD is fatal within six months. Electroencephalogram features typical for CJD were absent in these cases. A causal link to BSE has been suggested for this cluster of a previously unrecognised variant of CJD. However, a link with BSE cannot be confirmed on present evidence.

### THE RESPONSE IN AUSTRALIA TO BSE

Australia has not allowed the importation of live cattle, cattle semen or embryos from the United Kingdom or other BSE-affected countries since 1988. There are 31 cattle of UK origin remaining in Australia, eight of them in NSW. These animals are 12 years of age or more, are free of BSE symptoms and have been under quarantine surveillance. The cattle will not enter the food chain. NSW Agriculture has undertaken surveillance for BSE in NSW cattle since 1991. About 1,500 brains from cattle showing neurological signs have been examined and all have been negative for BSE.

Scrapie is not present in Australian sheep and BSE is not present in Australian cattle herds. Very little ruminant-derived protein is used in Australian cattle feed, and readily available vegetable proteins have been the main source of protein supplements. However, NSW Agriculture proposes implementation of the April 1996 recommendation of the WHO Consultation on public health issues relating to bovine spongiform encephalopathy that all countries ban the use of ruminant tissues in ruminant feed.

There have been no commercial imports of fresh or frozen beef from the UK in recent years. The Federal Government has stopped the importation of a small range of food products such as beef soup and meat-based flavourings, which may contain beef from the UK. The National Food Authority and NSW Health have overseen a recall of these products from retail outlets.

No medical devices of bovine origin have been imported into Australia from the UK. Medical devices supplied in Australia, such as haemostats, coated vascular grafts, coated heart valves, collagen implants and bone are manufactured from materials obtained from herds in such countries as Australia, New Zealand, the US, the Netherlands and Brazil. Therefore people who have received implants of therapeutic devices made from bovine material have almost certainly not been exposed to BSE-infected material.

A CJD case register was established in 1994 at Melbourne University to monitor the incidence of the disease in Australia.

### DISCUSSION

Australian consumers of imported foods containing beef from the UK, and travellers to the UK, may have been exposed to some risk, but the risk would appear to be very low.

The predicted and apparent decline in incidence of BSE in the UK suggests that control measures implemented in 1988 have been effective and that the greatest risk probably occurred between 1985 and 1991.

The association between BSE and CJD has attracted widespread media interest and international controversy. The implications for the British beef industry have been devastating, with serious consequences for the British economy.
The British Government has failed to convince the public and trade partners that BSE in beef from the UK presents an acceptable public health risk. Control measures have sometimes appeared not to be timely, appeared half-hearted and presented contradictory messages. This is illustrated by the 50 per cent subsidy initially offered to farmers to destroy symptomatic stock. At the same time the public was told beef was safe to eat. This appeared to offer little incentive to farmers to destroy stock, which the Government agreed was safe to eat, when the market place would probably offer full value if symptoms were not too obvious. The Government appeared to have no contingency plans to deal with an issue that had been threatening to explode for many years.