# SPONGIFORM ENCEPHALOPATHY IN RUMINANTS: EPIDEMIOLOGY, DIAGNOSIS AND SOCIO-ECONOMIC IMPLICATIONS

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## INTRODUCTION

Until 1985 six transmissible spongiform encephalopathies (SE), or prion diseases were known, three in man, kuru, Creutzfeldt-Jakob disease (CJD) and Gerstmann-Sträussler-Scheinker disease, all of which are rare or very rare and three in animals, scrapie of sheep and goats, which is relatively common, transmissible mink encephalopathy (TME) of farmed mink and chronic wasting disease of some species of deer and Rocky Mountain elk which are also rare diseases. CJD has a worldwide distribution but scrapie does not exist in several countries including Australia, New Zealand and some South American countries. The other diseases are geographically restricted and none of the animal diseases are implicated in the causation of the human diseases. All the diseases are fatal. There is no certain way of confirming the diagnosis in a live animal, there is no practical test for infection and no effective treatment is available.

## EPIDEMIOLOGY

Since 1996, when bovine spongiform encephalopathy (BSE) was first discovered in the UK, fourteen additional diseases have been reported. The SE in moufflon is probably scrapie. The SE in the other thirteen species are probably caused by BSE. There have been nineteen cases of SE in eight species of captive wild ruminants, ten cases in captive wild FELIDAE and seventy eight cases in domestic cats (seventy six in the U.K, one in Norway and one in Liechtenstein).

The cattle and captive wild ruminants were infected orally by concentrate feed containing meat-and-bone-meal (MBM) which was the vehicle for the scrapie-like agent responsible for the disease. The domestic cats were probably also infected *via* feed but the specific

ingredient is unknown. The captive wild FELIDAE were infected *via* feeding uncooked central nervous tissue from affected cattle.

The origin of the epidemic was either from a scrapie-like agent from sheep (the only known reservoir) or a cattle-adapted scrapie-like agent. Commercially motivated changes in the UK rendering industry were responsible. This industry processes waste tissues from abattoirs and butchers and converts it into MBM (which is the carrier of infection) and tallow (fat), which does not carry infection even if it was present in the raw ingredients. Once it was discovered that feed was the source of BSE a ban was introduced (in July 1988 in the UK) which prohibited the feeding of ruminant protein to ruminant animals. This, if perfectly enforced, and if there was no other source would result in the eradication of BSE once the incubation period was complete. The modal incubation period is five years. There have been over 30,000 cases of BSE born after the feed ban was introduced, it is now clear that the greater majority of these were due to cross contamination of ruminant diets with those prepared for pigs and poultry using the same equipment. Failure to clean delivery wagons between deliveries of raw materials or finished feed for different species could also have contributed. Progressive tightening of the controls, and importantly the introduction of the specified bovine offals ban helped to effect control of BSE. This was primarily introduced to protect public health in 1989 from any risks associated with infected tissues from clinically healthy, infected cattle but was extended in 1990 to protect all species of animal and bird. However, this ban too, was 'leaky' and enforcement had also to be tightened to reduce, and finally prevent, exposure *via* feed. Because the security of the ruminant feed ban could not be 100% guaranteed, since April 1996 mammalian MBM has not been permitted to be supplied or fed to any food animal species in the UK, including horses and fish. These measures have resulted in a progressive and sustained decline in the UK epidemic. In the absence of any significant cattle to cattle spread, for which currently there is little evidence, the UK is *en route* to eradicate the disease by the early years of the next century. A few cases of BSE in Europe and one in Canada (<300 in any other country) have resulted from the importation of infected animals, or feeding of infected imported MBM, or MBM from indigenous sources, to cattle. Only Switzerland of the five other countries with BSE in native-born cattle has brought the disease under control by adopting similar methods to the UK at an early stage.

## DIAGNOSIS OF BSE

BSE is suspected clinically but the signs are not pathognomic and even in the best circumstances only about 85% of suspect cases will be confirmed *post mortem*. Confirmation is normally made by microscopic examination of the brain looking for the specific lesions and locations of spongiform change and neuronal vacuolation. Additional methods seek the disease-specific protein $PrP^{Sc}$ which can be detected in tissue sections (by immunohistochemistry) or by immunoblotting of detergent extracts of unfixed brain or spinal cord treated with proteinase K or by examination of similar extracts for scrapie associated fibrils (SAF, which are morphologically distinct aggregates of the truncated form of $PrP^{Sc}$ - $PrP_{27\text{-}30}$) again using unfixed tissue. Recently there have been reports of successful detection of cases by examining samples of cerebrospinal fluid for disease-associated proteins or urine for disease-associated changes in analyte composition. These show some promise of detection of disease in the live animal but probably only in clinically sick animals.

## SOCIO-ECONOMIC IMPLICATIONS

Several researchers have addressed this problem scientifically over the period of the epidemic both in the UK and elsewhere. The ingredients of the BSE story stimulated extensive media interest which was fuelled by some irresponsible scientists and reporters with gossip, and false claims on subjects of which they had little knowledge. Although some sections of the media persisted for years in misrepresenting the truth and consistently presenting unbalanced views at the extreme end of the spectrum of rationality, others have become much more responsible and sought the truth, based upon their own intense research into the subject. For the most part in the UK now, reporting tends to be factually accurate with only the headlines being sensationalist. However, the same poor historical record of British journalists does seem to be repeating itself now in some European and North American countries.

How did all this come about and what were the features of BSE that distinguish it from other diseases and accidents? BSE scares followed on from the food scares of the 1980s relating to salmonella contamination in eggs and contamination of sheep meat with radioactivity resultant upon the Chernobyl nuclear reactor explosion and subsequent fall-out over parts of Wales, Northern England and Scotland. The Government was quick to act in all three circumstances and took measures to protect the public from any risks there may theoretically have been.

The consumer already had a high profile and demanded more recognition as the end user of most animal products. Consumer's views were put forcibly by powerful and power-seeking Consumer Groups but these were sometimes ill-informed and this had to be corrected and was over a period of time. The public's perception of food safety was already at a high level when BSE struck. Whereas earlier the 'man in the street' was little concerned about how meat was prepared in abattoirs or about what happened to edible offals and waste, or what MBM was and how it was fed to food animals, there was growing interest in these issues which created a great concern because of the newly perceived implications for food safety. Most people found it abhorrent for cattle or other ruminants, regarded as herbivores, to be fed the cooked, defatted and dried remains of their relatives. This was vividly explained to them, mainly inaccurately, by the media. That it had been happening for decades without apparent concern was no reason for it to continue and it did not. The practice was banned in 1988, but for scientific reasons connected with BSE.

The main ingredients of the BSE story that ignited the public's attention was the mystery surrounding the disease - it was entirely new - and the agent which caused it was not known. So we had a new risk, an unknown cause and publicly expressed controversy fuelled by some media-seeking individuals, the classic ingredients for a good and long running story. Although from the outset there was a perceived risk to humans from BSE and control measures were put in place to protect public health from 1988, every step of the way brought forth new confidence-damaging information. BSE was as expected found to be transmissible, first to mice. Subsequently it was unexpectedly transmitted to pigs. Cats succumbed naturally with an entirely new disease (feline SE) in 1990 which, like a few SE cases in captive wild animals, seemed closely related to BSE. Then despite the 1988 feed ban cases of BSE started to occur in cattle born after this time (>30,000 to date) and these were linked to weaknesses in the enforcement of the various bans and to the very small amount (1g) of brain that could transmit BSE to cattle *via* the oral route which was established by research. The final and most important feature of all was the occurrence, during 1995/6, of 10 cases of a new variant (NV) form of CJD. Though there is still no direct link with BSE, there was no other obvious explanation for the temporal and geographical occurrence of this new disease. The most likely cause (so it was announced by the Secretary of State for Health on 20 March

1996) was exposure to BSE prior to the SBO ban in 1989 (possibly *via* consumption of cattle offals such as brain or spinal cord).

This announcement caused the European Commission, stimulated by the Member States of the EU, to demand a ban on the export of cattle and most bovine products from the UK immediately with the object of restoring confidence in beef. Milk, hides and semen were excluded after discussion. Currently only meat (with certain rare and approved exceptions) from cattle under 30 months of age can be consumed in the UK. Milk from all clinically healthy cattle is internationally agreed to be safe. All products other than hides from cattle older than 30 months of age when they are killed are rendered safely for later incineration. Over one million cattle over thirty months have been killed up to 1 January 1997 and over 100,000 more are being culled compulsorily to meet the terms of the EU for lifting the ban. The longer term environmental effects from the stored material prior to final safe disposal are now being considered.

To say the UK and European beef industry has been seriously affected would be an understatement. Export sales of live cattle, beef and cattle products from the UK and some other countries have been reduced or stopped. Beef consumption has fallen dramatically in most countries of the EU but has partially recovered in the UK. The consumption of prime beef joints and minced meat certified to only consist of beef, is now close to its previous level in the UK but, offal and products like pies, sausages and burgers containing cattle products are purchased and consumed less than before and the recovery is slower. Major effects have occurred in the rendering trade now converted substantially to a waste disposal industry. Other significant effects have occurred in the gelatin manufacturing industry though gelatin too is regarded as safe by world authorities providing the raw materials are safely sourced and approved manufacturing protocols are used.

What has been the cost? This is very difficult to calculate but in the UK about £200M will have been spent on slaughter and compensation alone, and the cost of BSE legislation to the abattoir industry to 1995 was calculated to be about £18M pa. All this is likely to be exceeded by the additional costs accrued from the export ban of 1996 which remains in place at present and does not yet seem to have achieved the objective of improving consumer

confidence. The costs of the over 30 months scheme, the compulsory culls and storage and final disposal of the end products will make a formidable contribution to the overall costs.

Perhaps it is better to look on the bright side. Farmers will be developing more economic and environmentally friendly beef production systems. Much improved quality assurance procedures are being developed with traceability from farm to table. The attraction of beef from pure beef breeds rather than from cross-bred dairy animals will gain a further foothold in the export market, especially as such cattle have been largely untarnished by the BSE stigma. They were rarely fed MBM. In the end the UK farming industry, especially the beef industry, will be the stronger for its experiences with BSE. The U.K will be out of the tunnel before anyone else and though all beef (*i.e.* meat) from all countries can be safely consumed if the current meat hygiene laws are enforced, it is in the UK that the best guarantees can be given. Control of the specified risk materials in countries outside the UK is imperative. In the UK no mammalian MBM is permitted to be fed to any food animal species, no beef from cattle >30 months old can be consumed and the last born animal to develop BSE was born in September 1993. Any current risk is therefore imperceptibly small from UK cattle. However, the final arbiter of public confidence will be the number of NV-CJD cases that occur in the new few years and whether or not any definite link can be made between such cases and BSE. At present there is no evidence for such a link. Let us hope it remains that way.

## FURTHER READING

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