

Bovine Spongiform Encephalopathy (B.S.E.)— A New Disease?

P. J. N. Pinsent, *F.R.C.V.S.*
Saxon Place, Langford
Bristol, England

Introduction

This disease is probably a genuinely new disease. It apparently only exists in Great Britain, and it seems unlikely that it was present in this country before 1984. It is a slowly progressive encephalopathy of adult cattle producing, microscopically, a spongiform appearance of sections of brain tissue. It runs a relatively lengthy course which invariably ends in death. It is probably due to an "unconventional viral" agent similar to that of scrapie.

Incidence

Between 450 and 500 cases have been confirmed to date, mostly in Friesian or Holstein-Friesian dairy cows. Most cases are between two and five years of age, four years being a common age. There is no obvious relationship to pregnancy or lactation, and most incidents are sporadic. Where more than one case has occurred on a farm, the numbers affected have been very small in relation to the total cow population. So far, only one bull has been affected, but this is not surprising for the majority of dairy herds use artificial insemination. Cases have occurred in most areas of Great Britain, but have been most common in the South and South West, and least common in Wales, Scotland and the North East of England.

Clinical Signs

There is insidious, progressive, locomotor and behavioural change, which develops after a very long incubation period, possibly of several years. Milk yield slowly falls and there is loss of bodily condition. Temperature, pulse and appetite remain normal and there are no specific physical signs. At first there is a tendency towards an abnormal degree of apprehension. If the animal is cornered or hustled, the attitude may change from apprehension to aggression, and marked excitability may develop. The cow is particularly apprehensive on approaching doorways and may well refuse to enter the milking parlour. Handling or milking stimulates kicking. Handling of the head is particularly resented. Persistent grinding of the teeth occurs; tremors develop along the lower part of the neck, over the shoulders, and occasionally along the flank. Occasionally, cows show a generalised muscular tremor. There are semaphore like movements of the ears, and a

hind leg ataxia with swaying of the hindquarters. There may be a marked high stepping hind leg gait.

Pawing at the ground occurs, and there may be continual licking of the muzzle and nose. If the cow turns sharply there may be knuckling of the hind fetlocks, stumbling and even falling. From recumbency she may be able to rise on to her front legs, but not on to her hind legs, which appear very weak.

Response to sound is grossly exaggerated and a loud handclap may cause the patient to fall over.

If the cow is made to trot she may "pace" with front and hind legs on each side moving in unison. The tail head is raised at the trot and the hindquarter ataxia much worse.

The syndrome deteriorates gradually over a varying number of months until the cow eventually becomes unmanageable, or goes into convulsions, or even dies. Occasionally there is temporary improvement, but the condition always worsens again.

Differential Diagnosis

Differential diagnosis includes listeriosis, hypomagnesaemia, and nervous ketosis. Hypomagnesaemia and nervous acetoanaemia develop much more rapidly and respond equally rapidly to appropriate treatment. Listeriosis is also more acute in onset, and often shows a raised temperature. It usually produces signs of 7th cranial nerve involvement, with drooping of the eyelid and ear on one or both sides with facial paralysis; or 5th, 9th or 12th nerve involvement producing difficulty in chewing, manipulating the food in the mouth, or swallowing. Rabies does not occur in Britain.

Pathology

Wells and others (1987) describe the pathological findings in detail. Histopathological examination of the brain showed bilaterally symmetrical degenerative changes in certain areas of brain stem grey matter. There were a certain number of discrete ovoid and spherical vacuoles in the neuropil.

Neuronal perikarya and neurites of certain brain stem nuclei contained large intracytoplasmic vacuoles. These could be single or multiple. The contents of vacuoles, both in the neuropil and in neurons, remained unstained and clear after histological staining for glycogen in paraffin sections, and for lipids in fixed cryostat sections.

The changes described were clearly pathological, and were distinguishable from those seen in the brains of healthy cattle.

Treatment and Control

No treatment appears to be effective in this disease. By virtue of the Animal Health Act (1981), and the Bovine Spongiform Encephalopathy Order (1988) the disease is notifiable, and suspected cases must be reported to the relevant Divisional Veterinary Officer of the Animal Health Department of the Ministry and Agriculture, Fisheries and Food.

The animal is slaughtered and the head removed so that the brain can be submitted to the relevant Veterinary Investigation Centre for confirmation by histological examination. Compensation amounting to 50% of the market value is paid for confirmed cases.

The carcasses of confirmed cases may not be used for human consumption. Substances of ruminant origin may not be incorporated into bovine feed stuffs.

Discussion

There is no evidence to suggest that environmental toxins or drug reactions play any part in causing this disease. There is no firm evidence that it is associated with the movement of cattle, or the close presence of sheep. There is no direct evidence of contagion, although it is obviously wise to isolate suspect cows if they calve while awaiting slaughter. Membranes and discharges should be destroyed and the area disinfected. Progeny of suspected or confirmed cases should be monitored continually.

Wells and his co-authors (1987) regard the histological evidence of this disease as compelling provisional evidence of similarity with diseases such as scrapie caused by unconventional infectious agents, although there is as yet no conclusive evidence.

Studies on the background and heredity of affected animals are important. Nevertheless it seems possible that the cattle population was widely exposed to a common infective source two or three years before the first cases occurred. It is possible that this infective source was the "unconventional viral" agent of scrapie present in meat and bone meal products derived from sheep and included in dairy rations. Such scrapie like organisms are not easily inactivated by heat or other processes (Morgan K (1988) *Veterinary Record* Vol 122 p445). There are, of course, very many more sheep in the country than formerly, and as a result of diversification by dairy farmers affected by milk quotas, many of these sheep were reared and maintained on dairy farms.

References

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This paper was prepared at the request of the Editor and is a review of the available literature on this new disease. Dr. Pinsent is Emeritus Professor of Veterinary Medicine, Bristol University.