SECOND CASE OF CJD IN DAIRY-FARMER

DH were informed last week by Dr Will of the CJD Surveillance Unit in Edinburgh of a case of CJD (confirmed by brain biopsy) in a 64 year old dairy farmer, who is presently hospitalised near his home in the West Country. The farmer is thought to have had at least two cases of BSE in his herd, which were diagnosed in 1992. The farmer is reported to have assisted in calving and to have drunk the milk from his herd. The history does not suggest that this is anything other than a sporadic case of CJD.

There are similarities between this case, and the confirmed case of CJD in a dairy farmer reported earlier this year (see attached case report from The Lancet). This previous case attracted a good deal of media attention, and this current case is likely to do the same, especially as his family are known to be concerned. MAFF are aware of the situation and are briefing their ministers.

We are taking expert advice with respect to this further case, and we will keep you informed. At the moment, the existence of this case is confidential. We therefore suggest the following line to take in case of enquiries:

"DH is aware of a second case of CJD in a dairy farmer who has had BSE in his herd. We cannot comment on the details of the case, but we know of nothing to suggest this is anything other than a sporadic case of CJD. The Department continues to monitor the incidence of CJD in humans."
If pressed:

The numbers concerned are very small, and it is not possible to draw any conclusions from such small numbers. This issue is being considered by the Government's expert advisers.

More detailed Q & A briefing will be provided as soon as we have more information.

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The notion that CJD is always acquired (as opposed to inherited) and that the existence of any hypothetical risk factor must therefore be the cause of the disease led to the much cited claim that the high incidence of CJD among Libyan Jews was due to their consumption of sheep's eyeballs, despite a lack of evidence that their dietary habits differed from their ethnic neighbours in whom no increased incidence of this disease was recorded. The high frequency of CJD in the Libyan Jews is now known to be due to a codon 200 mutation in the PrP gene in affected families in that ethnic group.

CJD is a peculiar disease that does not fit into any single pattern of distribution. The great majority of cases cannot be attributed to environmental exposure. Very particular precautions are required to prevent transmission in the great majority of cases of human and animal spongiform encephalopathy since, when this does occur, a major outbreak of disease can arise. Under these circumstances it is especially important that the occurrence of CJD is viewed from an epidemiological rather than an anecdotal perspective.

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R. M. RIDLEY
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Creutzfeldt-Jakob disease in an individual occupationally exposed to BSE

Sir,—The occurrence of bovine spongiform encephalopathy (BSE) has led to public and professional concern about the possibility of a risk to human health and the reinstatement of surveillance for Creutzfeldt-Jakob disease (CJD) in the UK. We have identified an individual with pathologically confirmed CJD who had previously had occupational contact with BSE.

A 61-year-old right-handed man was admitted for investigation of progressive dysphasia and impairment of short-term memory for 4 months. He had had progressive dysphasia, dysgraphia, constructional dyspraxia, and mild impairment of short-term memory. Cranial nerves were normal. The only non-specific abnormality was reduction in dexterity in the right hand; there were no involuntary movements.

Computed tomography of the brain revealed a slightly enlarged ventricular system and mild cortical atrophy. Cerebrospinal fluid was normal. The initial electroencephalogram showed continuous high-amplitude slow activity in the right hemisphere but serial recordings revealed more frequent periodic complexes at about 1 per second, consistent with the diagnosis of CJD.

Dysphasia and apraxia worsened, with akinetic mutism at 3 weeks, associated with frequent myoclonic jerks and several generalized seizures. The patient developed brachymnesia and died 3 months after admission. Postmortem histological examination showed spongiform change typical of CJD throughout the cerebral cortex, with particularly severe changes in the occipital lobe. Immunocytochemistry for prion protein (Ab4 antibody, from Dr J. Hope, Neuropathogenesis Unit, Edinburgh) showed intense staining in the neocortical area adjacent to areas of spongiform change, especially in the occipital lobe.

The patient had been treated for hypertension for the preceding 16 months and had undergone an operation for intestinal volvulus as an infant. There was no history of previous neurosurgery and no family history of dementia. The open-ended frame of the prion protein was sequenced at the Centre for Genome Research, Edinburgh, and was normal, excluding any of the known pathogenic mutations associated with familial CJD.

The patient had been a dairy farmer throughout his working life and in 1998 had had a case of BSE in his herd (confirmed histologically, J. Wilesmith, Central Veterinary Laboratory). The animal had been potentially exposed to contaminated feed before July 1986, when the feeding of ruminant proteins to cattle was banned. The farmer had had no contact with the cow's internal organs or tissues (e.g., in assisting veterinary surgery or at the animal's destruction). He had drunk pooled milk from the herd which included milk from the affected animal.

This is the first report of CJD in an individual with direct occupational contact with a case of BSE and raises the possibility of a causal link. About 120,000 individuals work in dairy farming in England and Wales and over one-third of farmers have had at least one case of BSE. The national incidence of CJD is about 0.5 cases per million per year and a crude calculation suggests that in the 2 years since the start of our survey, we would have expected about 0.05 cases in dairy farmers. With a BSE-affected herd, this calculation takes no account of other groups with increased exposure to affected animals and we found no case of CJD in other potentially "at-risk" groups, such as abattoir workers or veterinarians. We have identified individuals with occupation (e.g., vicar, art teacher) that are statistically less likely to have occurred by chance than potentially "at-risk" occupations.

The course of symptoms and signs in our case, the investigations (including electronmicroscopy) and the necropy findings are consistent with previous experience in CJD.2 Risk factors for CJD, including intravenous transmission and genetic predisposition, have been largely excluded by the history and gene analysis. The Southwood Committee recommended surveillance of specific occupational groups because of the risk of direct inoculation of bovine tissue.3 The history suggests no occurrence in our case and the only possible direct exposure was by drinking milk. Milk does not contain detectable titres of infectivity, even from animals clinically affected with natural disease4 and epidemiological evidence (e.g., the absence of vertical transmission in human cases) largely precludes milk as a route of transmission in spongiform encephalopathies.

CJD in our case is most likely to have been a chance finding and a causal link with BSE is at least conjunctural.

We thank the various neurologists, neuropathologists, neurophysiologists, other colleagues, and in particular the relatives of affected patients for their co-operation.

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R. G. WILL


CORRECTION

More long-term bacterial meningitis.—In this commentary by Dr O'Neill (Feb 27, p 350) we regret that we introduced an error into the second sentence of the fifth paragraph, which should have begun "Chloramphenicol is inferior to clindamycin in children's meningitis".