

CONFIDENTIAL

CONFIRMED CASE OF CJD IN A DAIRY FARMER

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Mr J Howard
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Mr Lister - DoH
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1. The Minister has asked for more detailed briefing about the latest case of Creutzfeldt-Jakob Disease (CJD) in a farmer in whose herds cases of BSE have been found. This is attached and, so far as it deals with human rather than animal health issues, the content has been agreed with Department of Health officials.
2. I should emphasise that there is no reason to think that this case will become known to the public in the near future, although the possibility cannot be ruled out. The purpose of the previous notes to the Ministers was simply to alert them to the possibility. The previous case did not attract public attention until details were published in a letter in the Lancet in March 1993, the farmer having died during the autumn of 1992.
3. Neither Dr Will nor the CJD Surveillance unit intend to disclose the existence of this case or make any comment at present unless it attracts media attention. If details are published in a medical journal we will ask to be forewarned of the date of publication. Information will eventually be included in the Annual Report on CJD Surveillance in the UK: the second annual report is in its final draft, so it will be at least a year before information is made public by this route.
4. Arrangements have been made for a special meeting of the Spongiform Encephalopathy Advisory Committee (the Tyrrell Committee), and a small group (ie, not all members of the Committee will attend) will meet at 11.00am on Tuesday 20 July. Written advice can be expected very soon after the Committee has met.

5. Close liaison between the Department of Health and the Ministry over this case will be essential. Demarcation of territory will be difficult as the media are certain to claim a link between CJD and BSE. Nevertheless it is important that, as far as possible, comment on human health issues is left to the Department of Health.

K. C. Taylor

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CONFIRMED CASE OF CREUTZFELDT-JAKOB DISEASE (CJD) IN A DAIRY FARMER

BRIEFING FOR MINISTER

1. HUMAN CASE DETAILS (PERSONAL AND GEOGRAPHICAL INFORMATION
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64 year old dairy farmer named Mr
clinical symptoms first seen May 1993
still alive in hospital
CJD confirmed by brain biopsy July 1993

2. FARM DETAILS (CONFIDENTIAL)

- i. 120 dairy cow unit.
- ii. 131 dairy cow unit.

3. CATTLE CASE DETAILS

- i.
 - a) Dairy cow born 14 December 1986
Clinical onset 18 May 1992
Suspect disease notified 18 June 1992
Slaughtered 18 June 1992
BSE confirmed 22 July 1992
 - b) Dairy cow born 7 September 1988
Clinical onset 18 March 1993
Suspect disease notified 12 May 1993
Slaughtered 12 May 1993
BSE being confirmed this week

ii.

Dairy cow born 7 September 1986
 Clinical onset 26 February 1993
 Suspect disease notified 26 March 1993
 Slaughtered 26 March 1993
 BSE confirmed 25 May 1993

4. WHAT IS CJD?

CJD is one of a family of transmissible spongiform encephalopathies, so called because of the spongy appearance of the brain under the microscope. The family includes scrapie in sheep and sporadically worldwide, with about 1 case per million people per year. Some cases are described as "familial" and are due to a gene. The remaining 80% are sporadic. A small number, described as variant CJD, result of surgical and biological products. In the UK, all such cases are monitored at the Western General Hospital, Edinburgh, and the Department of Health.

of transmissible spongiform encephalopathies because of the vacuolated (ie spongy) appearance of the brain under the microscope. The family of diseases includes BSE in cattle and scrapie in sheep. BSE is a rare disease which occurs at a rate of approximately 0.5-1 new cases per year. Some 10-15% of cases are described as "sporadic". The remaining cases are described as iatrogenic, have been the result of contamination by human growth hormone. Cases in the UK are monitored in a study conducted by Dr R Will, Edinburgh, which is funded by the Scottish Office.

5. UK INCIDENCE OF CJD

The incidence of CJD in other parts of the world is 0.86 cases/million/year and probable CJD cases in the UK. CJD is a notifiable disease, and case ascertainment may be incomplete - particularly in the period before 1990.

The incidence of CJD in the UK is similar to the incidence in other parts of the world. In 1992 the UK incidence was 0.86 cases/million/year. A histogram showing definite CJD cases in the UK and Wales only, from 1970 is attached: until 1984 CJD was not a notifiable disease, and case ascertainment may be incomplete - particularly in the period before 1990. Attention was focused on BSE in the UK in 1988.

6. CJD IN FARMERS

The second annual report on CJD surveillance in the UK, which is about to be published, gives occupational history details of 29 definite and probable CJD cases recorded in people who had a history of employment at any time in particular occupational groups of potential significance for the occurrence of the disease. The 29 cases were amongst 95 diagnosed over a 3 year period: the other 66 cases did not fall into such occupational groups.

These relevant details are:-

- medical/paramedical/dentistry 7
- animal laboratory 1
- pharmaceutical laboratory 0
- research laboratory 0
- farmers/veterinary surgeons 7
- butchers/abattoir workers/occupation involving direct contact with animals or carcasses 5
- occupation involving animal products 9

Although the small numbers make interpretation difficult, case control studies, which provide comparative data on occupational frequencies in cases and an age and sex-matched control population, do not suggest that any significant risk of developing CJD is associated with any of the specified occupational groups. In other words, current information does not suggest that occupation is linked to an increased risk of developing CJD, and this includes occupations which might involve an increased exposure to the BSE agent.

7. WHAT IS BSE?

BSE is a progressive neurological disorder of adult cattle, resulting from infection with an unconventional agent, the precise nature of which still remains uncertain. The first case was identified in November 1986,

Bov 173

although unrecognised cases are thought to have occurred from April 1985. 100,000 cases have been confirmed in Great Britain, mostly in dairy cows. Measures to control and eradicate the disease have been taken. There is no evidence that BSE is transmissible to humans, but measures to prevent any risk to human health are in place.

8. LINE TO TAKE

- make no comment unless case becomes public knowledge.
- there is no scientific evidence to link BSE with the human disease, CJD.
- this case of CJD is most likely to be a chance phenomenon, particularly as the interval between the first case of BSE on the farm and the onset of clinical CJD was only 12 months. Iatrogenic CJD has an incubation period measured in years.
- the steps needed to protect human health from any remote risk posed by BSE have already been taken. The independent Spongiform Encephalopathy Advisory Committee (the Tyrrell Committee) in their report published in July 1992 commented that they were "satisfied that all the necessary safeguards are in place to prevent any risk of transmission to humans".
- the Tyrrell committee is nevertheless considering this case, and will give advice

TABLE 3

DEATHS FROM CJD
(DEFINITE & PROBABLE CASES)
1970 - 1993

