

**ATTACHMENT 4**  
**MAY, 2000**  
**OIE BSE CASES**  
**UK nvCJD STATISTICS**

# Bovine Spongiform Encephalopathy

## NUMBER OF CASES OF BOVINE SPONGIFORM ENCEPHALOPATHY (BSE) REPORTED IN THE UNITED KINGDOM<sup>(1)</sup>

	1987 and before (2)	1988 (2)	1989	1990	1991	1992	1993	1994	1995	1996	1997	1998	1999 (3)
Great Britain	442	2 469	7 137	14 181	25 032	36 682	34 370	23 945	14 302	8 016	4 312	3 179	1 771
Northern Ireland	0	4	29	113	170	374	459	345	173	74	23	18	5
Isle of Man (4)	0	6	6	22	67	109	111	55	33	11	9	5	3
Jersey	0	1	4	8	15	23	35	22	10	12	5	8	3
Guernsey <sup>(5)</sup>	4	34	52	83	75	92	115	69	44	36	44	25	7
<b>Total United Kingdom</b>	<b>446</b>	<b>2 514</b>	<b>7 228</b>	<b>14 407</b>	<b>25 359</b>	<b>37 280</b>	<b>35 090</b>	<b>24 436</b>	<b>14 562</b>	<b>8 149</b>	<b>4 393</b>	<b>3 235</b>	<b>1 789</b>

(1) Cases are shown by year of restriction.

(2) Cases prior to BSE being made notifiable are shown by year of report, apart from cases in Great Britain which are shown by year of clinical onset of disease.

(3) As at 31 October 1999.

(4) In the isle of Man BSE is confirmed on the basis of a laboratory examination of tissues for the first case on a farm and thereafter by clinical signs only. However, all cases in animals born after the introduction of the feed ban have been subjected to histopath/SAF analysis. To date, a total of 277 animals have been confirmed on clinical grounds only.

(5) In Guernsey BSE is generally confirmed on the basis of clinical symptoms only. To date, a total of 575 animals have been confirmed without laboratory examination.

## NUMBER OF REPORTED CASES OF BSE WORLDWIDE\* (EXCLUDING THE UNITED KINGDOM)

COUNTRY	1989	1990	1991	1992	1993	1994	1995	1996	1997	1998	1999	2000
Belgium	0	0	0	0	0	0	0	0	1	6	3	2(a)
Denmark	0	0	0	1(c)	0	0	0	0	0	0	0	1(a)
France	0	0	5	0	1	4	3	12	6	18	31(b)	14(a)

<b>Ireland (Rep.)<sup>(d)</sup></b>	15 <sup>(b)</sup>	14 <sup>(b)</sup>	17 <sup>(b)</sup>	18 <sup>(b)</sup>	16	19 <sup>(b)</sup>	16 <sup>(b)</sup>	73	80	83	91	24 <sup>(a)</sup>
<b>Liechtenstein</b>	0	0	0	0	0	0	0	0	0	2 <sup>(a)</sup>		
<b>Luxembourg</b>	0	0	0	0	0	0	0	0	1	0	0	0 <sup>(a)</sup>
<b>Netherlands</b>	0	0	0	0	0	0	0	0	2	2	2 <sup>(a)</sup>	
<b>Portugal</b>	0	1 <sup>(c)</sup>	1 <sup>(c)</sup>	1 <sup>(c)</sup>	3 <sup>(c)</sup>	12	14	29	30	106	170	4 <sup>(a)</sup>
<b>Switzerland</b>	0	2	8	15	29	64	68	45	38	14	50	13 <sup>(a)</sup>

\* Cases are shown by year of confirmation.

*(a) Belgium - date of the last detection of a case: 19 February 2000 -*

*date of the last confirmation of a case: 2 March 2000;*

*Denmark - date of detection of the case: 20 January 2000 - date of confirmation of the case: 25 February 2000;*

*France - date of the last detection of a case: 23 March 2000 -*

*date of the last confirmation of a case: 3 April 2000;*

*Ireland - data as of 29 February 2000;*

*Liechtenstein - date of the last confirmation of a case: 30 September 1998;*

*Luxembourg - data as of 31 January 2000;*

*Netherlands - date of the last detection of a case: 10 March 1999;*

*Portugal - date of the last detection of a*

*case: 9 December 1999 - date of the last confirmation of a case: 11 January 2000;*

*Switzerland - data as of 3 March 2000 - New surveillance system since 1 March 1999.*

*(b) France: includes 1 imported case (confirmed on 13 August 1999).*

*Ireland: includes imported cases: 5 in 1989, 1 in 1990, 2 in 1991 and 1992, 1 in 1994 and 1995.*

*(c) Imported case(s).*

*(d) No cases were confirmed in Ireland before 1989. All the cases reported by Ireland to the OIE*

*have been in female animals, apart from one imported 5-year old bull which was confirmed*

*positive in 1989. There have been no cases reported to date in young male animals, i.e. steers or bulls.*

## THE FOLLOWING COUNTRIES/TERRITORIES HAVE REPORTED CASES ONLY IN IMPORTED ANIMALS

(in parentheses: date of initial detection)

**Canada:** 1 case (11/93); **Falkland Islands:** 1 case (1989); **Germany:** 6 cases (1 in 1992, 3 in 1994 and 2 in 1997); **Italy:** 2 cases (10/94); **Oman:** 2 cases confirmed in 1989.

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## MONTHLY CREUTZFELDT-JAKOB DISEASE STATISTICS

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2000/0251 - Tuesday 2nd May 2000

### MONTHLY CREUTZFELDT-JAKOB DISEASE STATISTICS

The Department of Health is today issuing the latest information about the numbers of known cases of Creutzfeldt Jakob disease. The position on variant Creutzfeldt Jakob disease (vCJD) - the form of the disease thought to be linked to BSE - is as follows:

Deaths of definite and probable cases in the UK

Year	Referrals	Sporadic	Iatrogenic	familial	GSS	vCJD probable still alive	vCJD probable deaths awaiting p.m. results	nvCJD confirmed*	Total
1985	-	26	1	1	0	-			28
1986	-	26	0	0	0	-			26
1987	-	23	0	0	1	-			24
1988	-	22	1	1	0	-			24
1989	-	28	2	2	0	-			32
1990	53	28	5	0	0	-			33
1991	75	32	1	3	0	-			36
1992	96	43	2	5	1	-			51
1993	78	38	4	2	2	-			46
1994	116	51	1	4	3	-			59
1995	87	35	4	2	3	-		3	47
1996	134	40	4	2	4	-		10	60
1997	161	59	6	4	1	-		10	80
1998	152	61	3	3	1	-		17	85
1999	161	56	6	2	0	-	1	12	77
2000*	70	6	0	0	0	10	1	4	21

\* To 28 April 2000. Total number of definite and probable cases of vCJD = 68.

\* including 3 historic probable deaths from vCJD where neuropathological confirmation will never be possible.

The next table will be published on Monday 5 June 2000.

**New information:** this table now includes the number of living patients known to be suffering from 'probable' vCJD. This is because recently agreed criteria for diagnosing vCJD in living sufferers has been confirmed by SEAC, the scientific advisory body which advises the Government on CJD issues, as being reliable enough to enable such information to be published. Further details were set out in

the Department of Health Press Release of 16 March (ref R227 - 34).

**Referrals:** a simple count of all the cases which have been referred to the National CJD Surveillance Unit for further investigation in the year in question. CJD may be no more than suspected; about half the cases referred in the past have turned out not to be CJD. Cases are notified to the Unit from a variety of sources including neurologists, neuropathologists, neurophysiologists, general physicians, psychiatrists, electroencephalogram (EEG) departments etc. As a safety net, death certificates coded under the specific rubrics 046.1 and 331.9 in the 9th ICD Revisions are obtained from the Office for National Statistics in England and Wales, the General Register Office for Scotland and the General Register Office for Northern Ireland.

**Deaths:** Apart from the 'still alive' column, all columns show the number of deaths which have occurred in definite and probable cases of all types of CJD and GSS in the year shown. The figures include both cases referred to the Unit for investigation while the patient was still alive and those where CJD was only discovered post mortem (including a few cases picked up by the Unit from death certificates). There is therefore no read across from these columns to the referrals column. The figures will be subject to retrospective adjustment as diagnoses are confirmed.

**Definite cases:** This refers to the diagnostic status of cases. In definite cases the diagnosis will have been pathologically confirmed, in most cases by post mortem examination of brain tissue (rarely it may be possible to establish a definite diagnosis by brain biopsy while the patient is still alive).

**Probable vCJD cases:** are those who fulfil the 'probable' criteria set out in the Annex and are either still alive, or have died and await post mortem pathological confirmation. Those still alive will always be shown within the current year's figures, until they ultimately transfer across into the 'awaiting p.m.' or the 'vCJD confirmed' column. It follows therefore that the figures in these columns will be subject to retrospective adjustment, for example as and when post mortem confirms diagnosis. After death, some cases\* are never confirmed pathologically because a post mortem examination does not take place (for instance where the relatives of the patient refuse consent) and these cases remain permanently in the probable category.

**Sporadic:** Classic CJD cases with typical EEG and brain pathology. Sporadic cases appear to occur spontaneously with no identifiable cause and account for 85% of all cases.

**Probable sporadic:** Cases with a history of rapidly progressive dementia, typical EEG and at least two of the following clinical features; myoclonus, visual or cerebellar signs, pyramidal/extrapyramidal signs or akinetic mutism.

**Iatrogenic:** Where infection with classic CJD has occurred accidentally as the result of a medical procedure. All UK cases have resulted from treatment with human derived pituitary growth hormones or from grafts using dura mater (a membrane lining the skull).

**Familial:** Cases occurring in families associated with mutations in the PrP gene (10 - 15% of cases).

**GSS:** Gertsmann-Straussler-Scheinker syndrome - an exceedingly rare inherited autosomal dominant disease, typified by chronic progressive ataxia and terminal dementia. The clinical duration is from 2 to 10 years, much longer than for CJD.

**vCJD:** Variant CJD, the hitherto unrecognised variant of CJD discovered by the National CJD Surveillance Unit and reported in The Lancet on 6 April 1996. This is characterised clinically by a progressive neuropsychiatric disorder leading to ataxia, dementia and myoclonus (or chorea) without the typical EEG appearance of CJD. Neuropathology shows marked spongiform change and extensive florid plaques throughout the brain.

**Definite vCJD cases still alive:** These will be cases where the diagnosis has been pathologically confirmed (by brain biopsy).

## ANNEX

### DIAGNOSTIC CRITERIA FOR VARIANT CJD

- I A) PROGRESSIVE NEUROPSYCHIATRIC DISORDER  
 B) DURATION OF ILLNESS > 6 MONTHS  
 C) ROUTINE INVESTIGATIONS DO NOT SUGGEST AN ALTERNATIVE DIAGNOSIS  
 D) NO HISTORY OF POTENTIAL LATROGENIC EXPOSURE
- II A) EARLY PSYCHIATRIC SYMPTOMS \*  
 B) PERSISTENT PAINFUL SENSORY SYMPTOMS \*\*  
 C) ATAXIA  
 D) MYOCLONUS OR CHOREA OR DYSTONIA  
 E) DEMENTIA
- III A) EEG DOES NOT SHOW THE TYPICAL APPEARANCE OF SPORADIC CJD \*\*\*  
 (OR NO EEG PERFORMED)  
 B) BILATERAL PULVINAR HIGH SIGNAL ON MRI SCAN
- IV A) POSITIVE TONSIL BIOPSY

**DEFINITE:** IA (PROGRESSIVE NEUROPSYCHIATRIC DISORDER) and  
 NEUROPATHOLOGICAL CONFIRMATION OF vCJD \*\*\*\*

**PROBABLE:** I and 4/5 OF II and III A and III B  
 or I and IV A

\* depression, anxiety, apathy, withdrawal, delusions.

\*\* this includes both frank pain and/ or unpleasant dysaesthesia

\*\*\* generalised triphasic periodic complexes at approximately one per second

\*\*\*\* spongiform change and extensive PrP deposition with florid plaques, throughout the cerebrum and cerebellum.



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This page last updated 4 April 2000