**Dyddiad:** 13 Hydref 1999

**Amser:** 9:15am - 13:30pm

Lleoliad: Ystafell Bwyllgora 1, Adeilad y Cynulliad Cenedlaethol

### YSTADEGAU DIWEDDARAF AR ACHOSION vCJD A BSE

Mae'r tablau ynghlwm yn rhoi'r ffigurau diweddaraf i'w cyhoeddi ar:

achosion vCJD, gan yr Adran Iechyd, ac

achosion BSE, gan y Weinyddiaeth Amaethyddiaeth, Pysgodfeydd a Bwyd.

## Ysgrifenyddiaeth

Hydref 1999

Cyhoeddir yr atodiad hwn yn yr iaith y'i derbyniwyd gan Gynulliad Cenedlaethol Cymru

DEPARTMENT OF HEALTH 04 October 1999

### MONTHLY CREUTZFELDT-JAKOB DISEASE FIGURES

The Department of Health is today issuing the latest monthly table, giving the numbers of deaths of definite and probable cases of Creutzfeldt-Jakob disease in the UK.

Year Referrals

Deaths of definite and probable cases in the UK

		Sporadic	Iatrogenic	familial	GSS	nvCJD	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	150	57	3	3	0	16	79
1999*	116	24	2	0	0	б	32

\* To 31 August 1999. Total number of definite and probable cases of vCJD = 46 (which includes one case who died after August)

1. The next table will be published on Monday 1 November 1999.

2. At its meeting on 18 March 1999 the Spongiform Encephalopathy Advisory Committee (SEAC) agreed that variant CJD (vCJD) should now be used in preference to nvCJD in line with current practice in many scientific journals.

Referrals: This is 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: These 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 figure includes 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 and Probable: 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 cases have not been confirmed pathologically; 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 andmyoclonus (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).

Probable vCJD: Cases in which post-mortem (or brain biopsy) has not been carried out and which fulfil preliminary criteria for the clinical diagnosis of vCJD. These criteria cannot yet be fully validated because of the limited experience of vCJD.

Cyhoeddir yr atodiad hwn yn yr iaith y'i derbyniwyd gan Gynulliad Cenedlaethol Cymru

MAFF BSE information: Incidence of BSE - Monthly Statistics

# **GENERAL STATISTICS - AS AT 27/08/99**

		PER CENT
TOTAL FARMS	34878	n/a
TOTAL CASES	175404	n/a
		% OF TOTAL
DAIRY FARMS	22041	63.19
SUCKLER FARMS	9437	27.06
MIXED FARMS	2086	5.98
NOT RECORDED	1314	3.77
DAIRY CASES	142160	81.05
SUCKLER CASES	20652	11.77
MIXED CASES	10373	5.91
NOT RECORDED	2219	1.27
PURCHASED CASES	56725	32.34
HOMEBRED CASES	117340	66.90
NOT RECORDED	1339	0.76

a. - *In the table above*, 'NOT RECORDED' = *data not yet entered in appropriate part of BSE database*.

CONFIRMED DAIRY HERD INCIDENCE	61.1%
CONFIRMED SUCKLER HERD INCIDENCE	16.4%
CONFIRMED TOTAL HERD INCIDENCE	37.3%
YOUNGEST CONFIRMED CASE	20 months
OLDEST CONFIRMED CASE	18 years 10 months

## YOUNGEST AND OLDEST CASES BY YEAR OF ONSET AS AT 01/09/99

YR OF ONSET	AGE YOUNGEST CASE (mths)	AGE 2nd YOUNGEST CASE (mths)	AGE 2nd OLDEST (yrs.mths)	OLDEST CASE (yrs.mths)
1986	30	33	5.03	5.07
1987	30	31	9.09	10.00
1988	24	27	10.06	11.01(2)
1989	21	24(4)	12.00(2)	15.04

1990	24(2)	26	13.03	14.00
1991	24	26(3)	14.02	17.05
1992	20	26	15.02	16.02
1993	29	30(3)	14.10	18.10
1994	30(2)	31(2)	14.05	16.07
1995	25	32	14.09	15.05
1996	29	30	15.07	17.02
1997	37(7)	38(3)	14.01	14.09
1998	34	36	14.07	15.05
1999	39	41	13.05	13.07

## **CONFIRMATIONS IN BULLS - AS AT 01/09/99**

The following table lists the number of bulls in which BSE has been confirmed, by breed, and with crosses included under main breed type.

Aberdeen Angus	5	Jersey	5
Ayrshire	5	Limousin	71
Belgian Blue	17	Lincoln Red	1
Blonde D'Aquitaine	13	Marchigiana	1
Brown Swiss	1	Murray Grey	2
Charolais	72	Red Poll	3
Devon	3	Saler	2
Friesian	114	Simmental	80
Gelbvieh	4	South Devon	7
Hereford	72	Sussex	4
Highland	4	Not recorded	9
Holstein	8		
Total	503		

# NUMBER OF CATTLE BORN AFTER FEED BAN (BAB) AS A PERCENTAGE OF BSE CASES BEING REPORTED - AS AT 01/09/99

(Note that these are suspects placed under restriction, NOT confirmed cases)

Month o	of Report	<b>Total Reported</b>	Number Non-BAB	Number BAB	Percent BAB
1	1993	4165	3657	508	12.20

2	1993	3933	3407	526	13.37
3	1993	4384	3746	638	14.55
4	1993	3639	3030	609	16.74
5	1993	3215	2619	596	18.54
6	1993	3104	2523	581	18.72
7	1993	3375	2685	690	20.44
8	1993	3299	2550	749	22.70
9	1993	3617	2730	887	24.52
10	1993	3360	2585	775	23.07
11	1993	3599	2666	933	25.92
12	1993	3241	2340	901	27.80
Mont	h of Report	<b>Total Reported</b>	Number Non-BAB	Number BAB	Percent BAB
1	1994	3511	2397	1114	31.73
2	1994	3096	2136	960	31.01
3	1994	3442	2249	1193	34.66
4	1994	2729	1789	940	34.44
5	1994	2484	1572	912	36.72
6	1994	2313	1411	902	39.00
7	1994	2044	1205	839	41.05
8	1994	2249	1247	1002	44.55
9	1994	2203	1256	947	42.99
10	1994	2082	1205	877	42.12
11	1994	2155	1160	995	46.17
12	1994	1951	1063	888	45.52
Mont	h of Report	<b>Total Reported</b>	Number Non-BAB	Number BAB	Percent BAB
1	1995	2017	985	1032	51.17
2	1995	1572	750	822	52.29
3	1995	1839	870	969	52.69
4	1995	1482	686	796	53.71
5	1995	1517	633	884	58.27
6	1995	1334	534	800	59.97
7	1995	1259	511	748	59.41
8	1995	1468	578	890	60.63
9	1995	1314	482	832	63.32
10	1995	1220	478	742	60.82
11	1995	1603	605	998	62.26
12	1995	1320	464	856	64.85

Month	n of Report	<b>Total Reported</b>	Number Non-BAB	Number BAB	Percent BAB
1	1996	1405	437	968	68.90
2	1996	1251	377	874	69.86
3	1996	1343	436	907	67.54
4	1996	945	277	668	70.69
5	1996	968	291	677	69.94
6	1996	690	180	510	73.91
7	1996	775	194	581	74.97
8	1996	755	162	593	78.54
9	1996	723	187	536	74.14
10	1996	762	177	585	76.77
11	1996	585	109	476	81.37
12	1996	495	76	419	84.65
Month	n of Report	<b>Total Reported</b>	Number Non-BAB	Number BAB	Percent BAB
1	1997	536	90	446	83.21
2	1997	501	94	407	81.24
3	1997	521	83	438	84.07
4	1997	523	77	446	85.28
5	1997	447	58	389	87.02
6	1997	432	50	382	88.43
7	1997	450	65	385	85.56
8	1997	454	52	402	88.55
9	1997	412	50	362	87.86
10	1997	460	59	401	87.17
11	1997	427	47	380	88.99
12	1997	441	55	386	87.53
Month	n of Report	<b>Total Reported</b>	Number Non-BAB	Number BAB	Percent BAB
1	1998	459	39	420	91.50
2	1998	403	38	365	90.57
3	1998	436	43	393	90.14
4	1998	384	48	336	87.50
5	1998	325	29	296	91.08
6	1998	334	23	311	93.11
7	1998	343	29	314	91.55
8	1998	307	24	283	92.18
9	1998	324	32	292	90.12
10	1998	371	39	332	89.49

11	1998	315	29	286	90.79
12	1998	290	15	275	94.83
Month	of Report	<b>Total Reported</b>	Number Non-BAB	Number BAB	Percent BAB
1	1999	316	25	291	92.09
2	1999	307	23	284	92.51
3	1999	353	17	336	95.18
4	1999	256	17	239	93.36
5	1999	279	11	268	96.06
6	1999	259	15	244	94.21
7	1999	224	9	215	95.98
8	1999	205	7	198	96.59