

**Incidence of variant Creutzfeldt-Jakob disease diagnoses and deaths in
the UK**

January 1994 – December 2010

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Summary

In 2010 there were four new diagnosis of vCJD and three deaths. This brings the total number of cases reported in the UK to 174 of whom 170 have died.

Results from modelling the underlying incidence of diagnoses and deaths indicate that the epidemic reached a peak in the year 2000 when there were 27 diagnoses and 28 deaths and has since declined to a current incidence of about one diagnosis/death per year.

An analysis that looked at deaths by birth cohort (pre 1970, 1970s, 1980s) showed that the shape of the epidemic differs between cohorts, mainly due to the fact that deaths of individuals born in the 1980s were only seen from 1999 onwards.

It is important to note that although a peak has been passed, it is possible that there will be future peaks, possibly in other genetic groups. There is also the possibility of ongoing person to person spread as seen with four cases of transfusion association vCJD infection to date, who received blood in 1999 or earlier from donors who were later diagnosed with clinical vCJD. Three of these individuals developed vCJD (one diagnosed in 2003 and two in 2006), whilst the fourth died from causes unrelated to vCJD, but was found on post mortem examination to have abnormal prion protein present in the spleen and a lymph node.

1. Introduction

Each year data on diagnosed cases of variant Creutzfeldt-Jakob disease (vCJD) in the UK are reviewed in order to investigate trends in the underlying rate at which deaths and diagnoses are occurring. The present report reviews the data for all individuals who had been classified as definite or probable cases by the end of December 2010.

2. Background information

Definite cases are those confirmed neuropathologically. To date all probable cases for which neuropathological data have become available have subsequently been confirmed as definite. The date of diagnosis is taken as the date when diagnosed as probable or, when this is not available, the date of confirmation of a definite case.

For these analyses we have included all cases notified to the National CJD Surveillance Unit and classified as definite or probable by the end of December 2010 (Table 1).

Table 1. Cases of vCJD classified as definite or probable by end of December 2010.

	Died*	Alive	Total
Male	97	2	99
Female	73	2	75
Total	170	4	174

* Deaths including 119 definite and 51 probable (without neuropathological confirmation).

Numbers of cases by onset, notification, diagnosis and death are given below by year along with the median age at death by year of death (Table 2).

Table 2. Annual cases by onset, notification, diagnosis and death (including median age at death by year of death).

Year	Onset	Notification	Diagnosis	Death	Median age at death
1994	8	0	0	0	-
1995	10	8	7	3	-
1996	11	9	8	10	30
1997	14	13	12	10	26
1998	17	20	17	18	25.5
1999	29	16	17	15	29
2000	24	29	27	28	25.5
2001	17 [^]	21	25	20	28
2002	14	15	16	17	29
2003	5 [^]	16	16	18*	28
2004	9	6	8	9	26
2005	5	7	6	5	34
2006	3	5	6	5*	30
2007	2	1	1	5*	24
2008	3 [^]	2	1	1	-
2009	3 [^]	4	3	3	-
2010	0	2	4	3	-
Total	174	174	174	170	28

* Three cases have arisen to date who had a blood transfusion from earlier cases. These cases, who were all male, died (were diagnosed) in 2003 (2003), 2006 (2006) and 2007 (2006). These cases are included in the analyses although are likely to part of secondary spread.

[^] This indicates the year of onset for the four living cases.

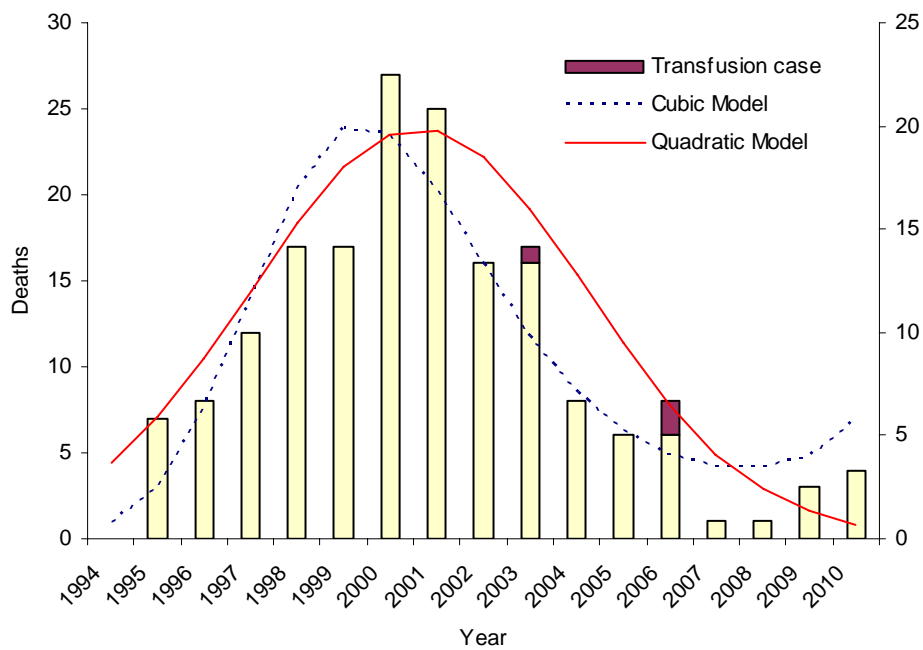
3. Methods

The incidence of deaths and diagnoses was modelled by Poisson regression using polynomials. Most deaths and diagnoses are reported quickly so an adjustment for reporting delay is not necessary. The age at death has not increased as may have been expected, assuming that most exposure to BSE ceased in the early 1990's. In order to examine this further the cases were stratified by year of death and birth cohort (pre1970, 1970s and 1980s). Trends in deaths over time were compared between these cohorts.

4. Results for Diagnoses

A quadratic trend model provides a good fit to the data (Figure 1), however there was evidence ($p < 0.01$) that a cubic model, which would give a longer tail to the epidemic, fitted better. This is due to the somewhat greater than expected number of diagnoses in 2009 and 2010. The peak is estimated to have occurred in mid 2000.

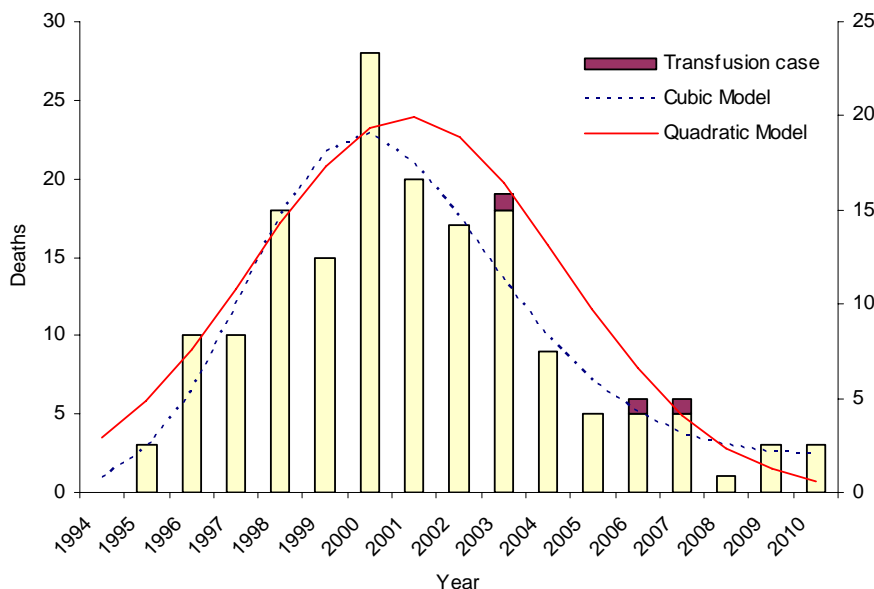
Figure 1: vCJD diagnoses by year with fitted quadratic and cubic trend lines



5. Results for Deaths

A quadratic trend model provides a good fit to the data (Figure2), however there was evidence ($p < 0.01$) that a cubic model fitted better. This is due to the somewhat greater than expected number of deaths in 2009 and 2010. The peak is estimated to have occurred in mid 2000.

Figure 2: vCJD deaths by year with fitted quadratic and cubic trend lines



Deaths by cohort

The epidemic curves vary significantly ($p < 0.001$) by birth cohort. The main difference is due to the fact that in the 1980s cohort no deaths were seen prior to 1999 (Figure 3). This finding is consistent with those born in the 1980s being infected towards the end of the BSE epidemic when they were older rather than at the beginning. This requires a lower exposure/susceptibility in the very young, which is reasonable because no cases have been seen to date in individuals born in the 1990s.

Figure3: Deaths by year and birth cohort

