Incidence of variant Creutzfeldt-Jakob disease diagnoses and deaths in the UK
January 1994 – December 2011

N J Andrews, Senior Statistician
Statistics Unit, Centre for Infections, Health Protection Agency

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Summary

In 2011 there were two new diagnosis of vCJD and five deaths. One of the new diagnoses related to a death in 2008. This brings the total number of cases reported in the UK to 176 of whom all 176 have died.

Results from modelling the underlying incidence of diagnoses and deaths indicate that the epidemic reached a peak in about the year 2000 when there were 27 diagnoses and 28 deaths and has since declined to a current incidence of about 1 to 2 diagnoses/death per year. There is evidence that the epidemic curve is skewed with an extended tail. Looking at onsets shows similar results with evidence of an extended tail.

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 in cases has been passed, it is possible that there will be future peaks, possibly in other genetic groups. To date, however, there is no evidence of a second wave.

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 2011.

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 Research & Surveillance Unit and classified as definite or probable by the end of December 2011 (Table 1).

Table 1. Cases of vCJD classified as definite or probable by end of December 2011.

	Died*	**Alive**	**Total**
Male	101	0	101
Female	75	0	75
Total	**176**	**0**	**176**

* Deaths including 122 definite and 54 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	6	7	6	5	34
2006	3	5	6	5*	30
2007	2	1	1	5*	24
2008	3	2	1	2	
2009	3	4	3	3	26
2010	1	2	4	3	
2011	0	2	2	5	
Total	176	176	176	176	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.

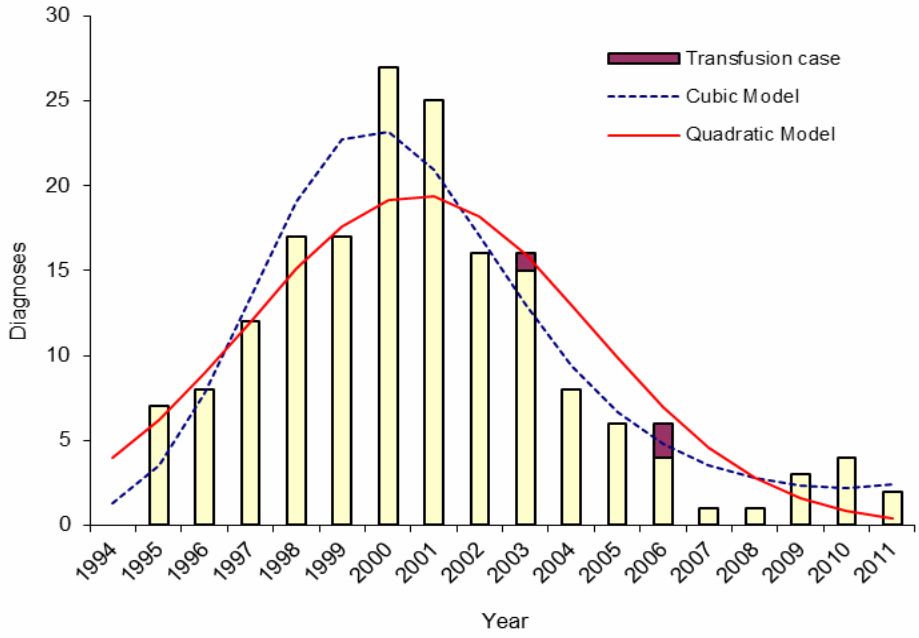
3. Methods

The incidence of deaths, diagnoses and onsets was modelled by Poisson regression using polynomials. Most deaths and diagnoses are reported quickly so an adjustment for reporting delay is not necessary. For onsets 2011 is not included. 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 (figure1), 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 positive skew in the epidemic curve (long tail). The peak is estimated to have occurred mid 2000.

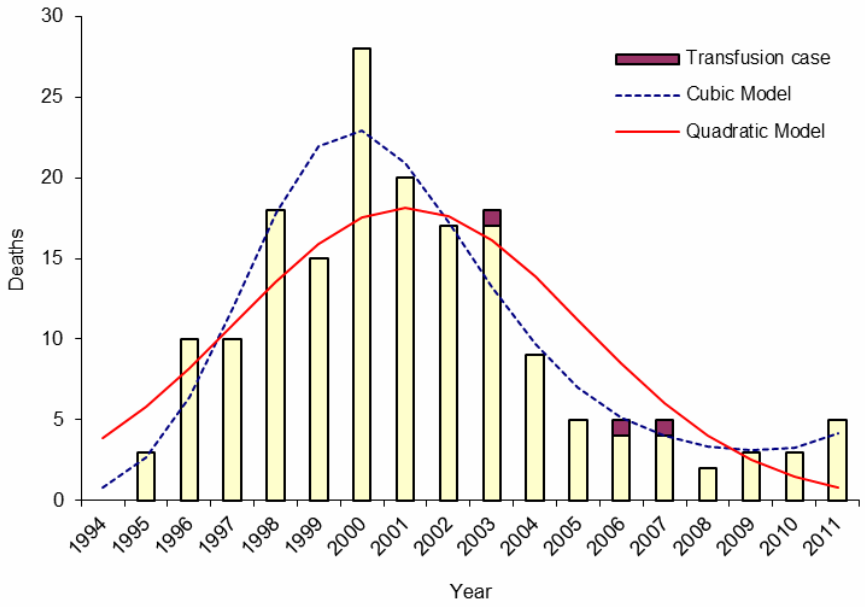
Figure 1: vCJD diagnoses by year with fitted quadratic and cubic trend lines



5. Results for Deaths

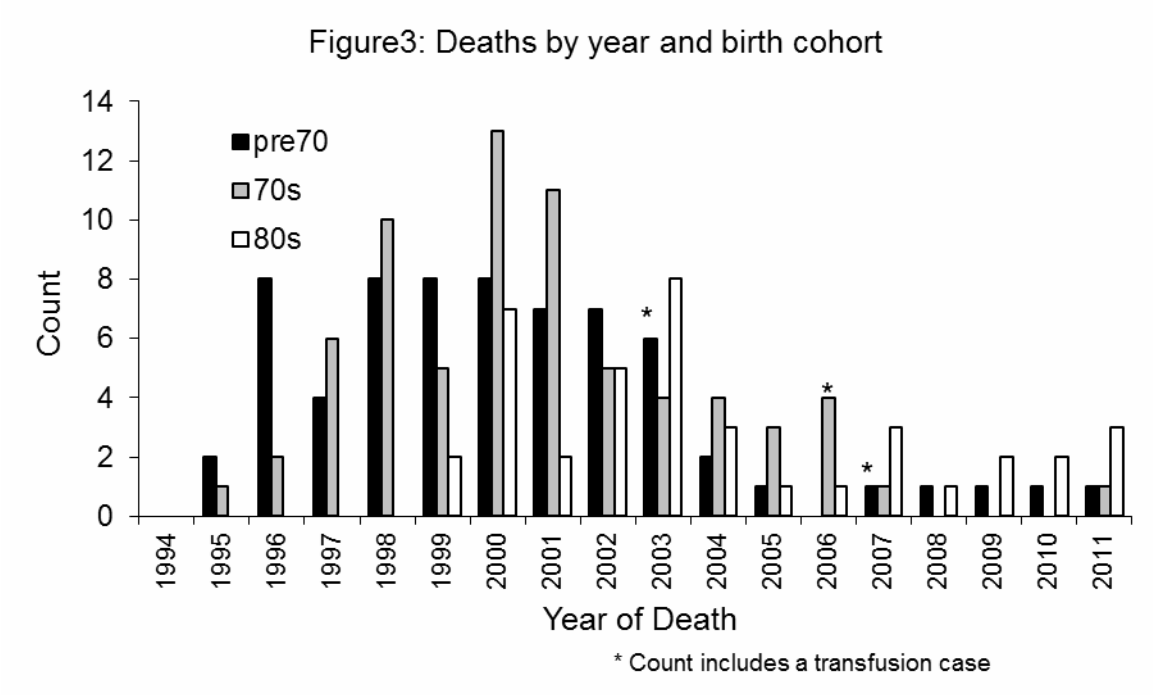
A quadratic trend model provides a good fit to the data (figure2) , however there was evidence ($p<0.01$) that a cubic model fitted better. This is due to the positive skew in the data. The peak is estimated to have occurred in mid 2000.

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.



6. Results for Onsets

Modelling onsets for 1993-2010 also gives a better fit the cubic model (p=0.001), although neither has a very good fit. The cubic model is better in the tails (at the start and end) and the quadratic model better around the peak. Both models predict a low current incidence of onsets (0 for quadratic and 2 for cubic).

Figure 4: vCJD onsets by year with fitted quadratic and cubic trend lines