



7. Creutzfeldt-Jakob disease

Surveillance objectives

The objectives of Creutzfeldt-Jakob disease (CJD) surveillance are to:

- Identify and investigate all suspect cases of CJD in Victoria;
- Identify and investigate any potential clusters or transmission events that may have exposed others;
- Monitor the epidemiology of CJD in terms of time, person, place and risk factors.

Summary of notifications

During 2005, the Australian National Creutzfeldt-Jakob disease Registry (ANCJDR) notified 19 cases of suspected CJD to the department. Of these, 11 cases were confirmed as definite CJD cases by neuropathological examination, six were classified as non-CJD following detailed investigation (two with neuropathological examination) and two were still under investigation at the time of writing (one of which was still alive). The 11 confirmed cases corresponded to an age standardised CJD mortality rate in 2005 of 2.2 deaths per million population. Of the 11 definite cases, five were female and six were male. The median age of death was 69 years (range: 18–75 years) and the median duration of disease was 7.3 months (range: 2–42.5 months).

Risk factors

No cases of iatrogenic CJD, acquired through pituitary hormone treatment, dura mater grafts or surgical instrument contamination, were identified. The ANCJDR did not identify any other risk factors for the cases notified in 2005 other than age.

Outbreak and other investigations

No outbreaks were identified.

Comment

In general, Victorian cases were slightly older and had a longer duration of disease compared to the national average (64 years and four months respectively). The Victorian standardised CJD mortality rate was higher than the global annual mortality rate of approximately one case per million. However, a higher number of autopsies were performed in Victoria on suspect CJD cases in 2005, suggesting greater ascertainment of definite CJD cases, which may partly explain the higher notification rate. Elevated annual mortality rates of this magnitude have also been observed in other Australian states.

Variant CJD (vCJD), the human disease form associated with the ingestion of dietary products contaminated with prion disease in cattle, has not been detected in Australia to date. The ANCJDR continues to monitor the occurrence of – and provide diagnostic services for – all suspect cases of vCJD in Australia.