



4. 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 2006, the Australian National Creutzfeldt-Jakob Disease Registry (ANCJDR) notified 11 cases of suspected CJD to the department. Of these, six were confirmed as definite CJD cases by neuropathological examination and one was confirmed as probable CJD after detailed investigation. Two cases were classified as non-CJD following neuropathological examination and two were still under investigation at the time of writing. The seven confirmed cases corresponded to an annual age standardised CJD mortality rate of 1.3 deaths per million population. Of the seven definite or probable cases, five were female and two were male. The median age of death was 67 years (range: 40–82 years) and the median duration of disease was three months (range: 1–13 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 2006 other than age.

Outbreak investigations

No outbreaks or common exposures between cases were identified.

Comment

In general, Victorian cases of CJD in 2006 were slightly older but had a similar duration of disease compared to the national average (65 years and four months respectively). The Victorian standardised CJD mortality rate was marginally higher than the global annual mortality rate of approximately one case per million. Annual mortality rates of this size have also been observed previously in Victoria and other Australian jurisdictions.

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 and classical CJD in Australia.