7. Creutzfeldt-Jakob disease

Creutzfeldt-Jakob disease (CJD) is a rare and fatal disease with an occurrence rate in Australia of one person per million head of population, comparable with worldwide data. CJD was scheduled as a notifiable disease in Victoria from 31 January 2004. The Australian National CJD Registry (ANCJDR), situated in the Department of Pathology at The University of Melbourne, is contracted to provide surveillance and investigation of CJD for the Department of Human Services. Confirmation of a suspected case of CJD is reliant on post-mortem investigations that may take up to eight weeks to complete.

The ANCJDR has established the total Victorian figures for 2004 as five confirmed cases, of which four were female and one was male. The median age at death was 64 years (range: 38–76 years) and the age-standardised mortality rate in Victoria for 2004 was 0.96 CJD deaths per million population. No cases of acquired CJD including iatragenic or variant CJD were identified in Victoria during 2004.

Risk factors

The ANCJDR assesses risk factors during investigation of all suspect cases of CJD and notifies the department of its findings. In 2004 a possible case with a history of neurosurgery was considered a risk to public health, the investigation of which is discussed below.

Outbreak and other investigations

The department, together with a metropolitan health service, was involved in the investigation of possible iatragenic transmission of CJD in 2004 when it was recognised that a newly confirmed case had undergone two neurosurgical procedures. Although the risk was considered minimal, the national CJD Incident Panel was notified and expert advice requested. As a consequence, several actions were undertaken: 1,200 patients from the hospital concerned were notified of the incident; a hot line was established for patients and general medical practitioners to contact the hospital for additional information and; CJD fact sheets were provided to patients and placed on the Department of Human Services and Australian Government Department of Health and Ageing websites. Patients who had undergone dura mater piercing procedures within three months of the index case's neurosurgical procedures were also mailed advice to provide for health providers about infection control measures to be undertaken in the event of the person requiring surgical procedures where the potential for further possible intragenic transmission may exist.

In addition, the destruction of all neurosurgical and other instruments potentially used in either neurosurgical procedure was considered appropriate as the health service was unable to track individual instruments to the patient. Furthermore, patients involved in this incident will be followed over time, although the risk of transmission is considered to be very low.

The investigation highlighted the importance of surgical instrument tracking systems to reliably identify those patients at risk of iatrogenic transmission and the absence of a CJD investigation protocol for retrospective diagnoses in the national *Infection control guidelines for the prevention of transmission of infectious diseases in health care settings*. The latter issue is being addressed through a revision of the CJD chapter of the national infection control guidelines, and will draw on the outcomes of a Victorian Advisory Committee on Infection Control consensus conference that discussed interpretation of the guidelines and the provision of consistent CJD advice for various clinical situations.

Comment

In the health care setting, risk factors for CJD include past use of cadaver acquired pituitary hormones and Lyodura (dura mater) from cadavers with undiagnosed/unrecognised CJD. These practices ceased in 1985/6. Another risk factor includes the surgical transmission from a CJD infected patient by instruments that have not been processed adequately to remove prions. This type of transmission (neurosurgical) occurred overseas and the last reported incidents were over 30 years ago and there have been major improvements worldwide in the cleaning, disinfection and sterilisation of reusable medical and surgical instruments and equipment since then. Information from the Australian Government Department of Health and Ageing about cadaver acquired pituitary hormone recipients is available from http://www.health.gov.au/internet/wcms/publishing.nsf/Content/health-pubhlth-strateg-phi-index.htm

Variant CJD – resulting from the ingestion of infected meat products – has not been reported in Australia to date. As a precaution against the possible transmission of variant CJD through blood products, the Australian Red Cross Blood Service (in alignment with most countries in the world) will not accept blood donations from people who have lived in the United Kingdom for a period of six months or more during the years 1980 to 1996. Information from the Australian

Government Department of Health and Ageing about Australia's response to the possibility of Variant CJD in Australia is available from http://www.health.gov.au/internet/wcms/publishing.nsf/Content/bovine+spongiform+encephalopathy+%28BSE%29-1

Information about CJD is also available from the department's website at http://www.health.vic.gov.au/ideas/diseases/cjd_facts.htm