Government of Western Australia
Department of Health

Communicable Disease Control Directorate Guideline

Creutzfeldt-Jakob Disease -

Infection prevention and reporting requirements in Western Australian Healthcare Facilities

Guideline 0028 / June 2025

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1. Definitions/Acronyms

Term	Definition
Australian National Creutzfeldt-Jakob Disease Registry (ANCJDR)	Monitors and reports on cases of CJD and other TSEs on behalf of the Australian Government. It also provides diagnostic testing and expert advice on diagnoses, epidemiology, and infection control matters in relation to TSEs in Australia.
Creutzfeldt-Jakob disease	CJD is a fatal neurogenerative disease belonging to a group of prion diseases known as transmissible spongiform encephalopathies (TSE). It includes sporadic, inherited and acquired CJD (iatrogenic and Kuru). These conditions are caused by an accumulation in the brain of an aberrant form of a normal cell surface glycoprotein, prion protein. It excludes variant CJD.
14-3-3 protein	An increased concentration of 14-3-3 protein in cerebrospinal fluid supports the diagnosis of CJD in a patient who has a compatible clinical illness and characteristic features on EEG and MRI and in whom other possible causes of rapidly progressive dementia have been excluded. In diagnosing CJD, the sensitivity of 14-3-3 ELISA is 81.3%, and its specificity is 84.4%.
Prion	The term prion means proteinaceous infectious particles. Prions are the infectious agents responsible for several neurodegenerative diseases in mammals, like, CJD. This happens due to the abnormal folding of the proteins in the brain.
Real-time quaking-induced conversion (RT-QuIC)	RT-QuIC a highly sensitive assay for prion detection. In diagnosing CJD, the sensitivity of RT-QuIC is 92%, and its specificity is <99%
WA health system	The WA health system is comprised of: (i) the Department; (ii) Health Service Providers (North Metropolitan Health Service, South Metropolitan Health Service, Child and Adolescent Health Service, WA Country Health Service, East Metropolitan Health Service, PathWest Laboratory Medicine WA, Quadriplegic Centre and Health Support Services); and contracted health entities, to the extent they provide health services to the State.

2. Purpose

This document describes the infection prevention management and the reporting requirements for Creutzfeldt-Jakob Disease (CJD) in Western Australian (WA) healthcare facilities. This document is to be used in conjunction with *Australian Department of Health Infection Control Guidelines for Creutzfeldt-Jakob disease (the CJD Guidelines)* which provides recommendations for infection prevention and control procedures to minimise the risk of transmission of CJD in health care settings and ensure patients with suspected CJD

have access to appropriate evidence-informed healthcare without discrimination or disadvantage.

For this document, the term CJD is used to describe all forms of human TSE (sporadic, inherited and acquired) except variant CJD (vCJD), which is linked to bovine spongiform encephalopathy, and is excluded from the scope of this document as it has not yet been reported in Australia.

3. Introduction /Background

CJD is a rare and rapidly progressive fatal neurodegenerative disease for which there is no known cure. CJD belongs to a group of prion diseases that affect humans known as TSEs. These conditions are caused by an accumulation in the brain of an aberrant form of a normal cell surface glycoprotein, prion protein.

Although transmission of CJD in a healthcare setting is very rare, there is the potential for transmission to occur via contaminated reusable medical devices. The infective agent of CJD, the prion, is resistant to routine reprocessing procedures. Therefore, additional reprocessing procedures must be implemented when an identified risk is determined based on the infectivity of the tissue to which the reusable medical device is exposed, and the patient risk factors for CJD.

Even though cases of CJD have been reported in healthcare workers, there are no confirmed cases linked to occupational exposure. There is no evidence to indicate that staff are at an increased occupational risk for acquiring CJD.

CJD is a nationally notifiable disease and under the Western Australian (WA) Public Health Act 2016 must be notified by the treating medical practitioner to the Communicable Disease Control Directorate (CDCD) pursuant to Part 9, Division 2 of the *Public Health Act 2016*. This includes all probable cases and those confirmed via neuropathology.

The Florey Institute manages the Australian National Creutzfeldt-Jakob Disease Registry (ANCJDR) and is responsible for diagnostic services and national surveillance of clinically suspected and diagnosed human prion disease on behalf of the Australian Government. The ANCJDR follows-up each case referred to the Registry, gathering detailed information and investigating potential causes. Early involvement of the ANCJDR allows liaison with the clinical team to encourage appropriate investigations.

There is a Confidentiality Agreement between WA Health and The Florey Institute which allows exchange of data concerning CJD to assist with the performance of their respective functions.

4. Requirements (of the Guideline)

CJD Infection Prevention Management

Health Service Providers are to ensure they:

- Comply with the risk assessment approach and infection prevention requirements outlined in *The CJD Guideline*.

- Develop local procedures to effectively screen, identify, risk assess and manage patients and reusable equipment to minimise the risk of CJD transmission.
- Have a documented plan to ensure admission and treatment is not delayed in the event a patient is identified in a high or low-risk category for CJD and is undergoing a procedure involving higher-infectivity tissue.
- Clearly communicate any reasons for variations or delays in treatment to the patient.
- Assess all patients undergoing surgical or diagnostic procedures involving higher-infectivity tissue have their risk category determined in accordance with *The CJD Guideline*.
- Use the risk assessment matrix as described in *The CJD Guideline* to identify whether routine reprocessing or additional reprocessing procedures will be required.
- Follow the additional procedures as described in *The CJD Guideline.* Single use instruments must be used, wherever possible, and when their use will not compromise patient care.
- Inform and provide training to relevant staff involved in the care of the patient, equipment reprocessing, or environmental cleaning of the need for the additional procedures.
- Arrange for deceased persons undergoing autopsy, with probable CJD to be transported to the Royal Perth Hospital (RPH) mortuary. The WA Department of Health CDCD will cover all costs associated with transportation to and from RPH.

CJD Reporting and Management

- The Director of Medical/Clinical Services of each Health Service Provider is required to notify the Director, CDCD, should an adverse event arise i.e. where after routine reprocessing, reusable medical devices used on a patient with symptomatic CJD have subsequently been used unknowingly on other patients.
  Note: If an adverse event occurs, the Director, CDCD will take responsibility for the investigation, equipment management, patient risk assessment and the scope of a look-back investigation where required.
- All medical practitioners who identify a probable or confirmed case of CJD, including sporadic, familial or acquired cases are required to complete a WA Infectious Disease Notifications to the CDCD pursuant to Part 9, Division 2 of the *Public Health Act 2016*.
- The Department of Health, Infection Prevention Policy and Surveillance Unit (IPPSU) is responsible for undertaking a public health risk assessment for any probable or confirmed case of CJD. This will be done in co-operation with the notifying medical practitioner and medical/nursing staff where the case may have received care in a WA healthcare facility within two years of diagnosis and in liaison with the ANCJDR.
- The IPPSU is responsible for:

    - ensuring the treating medical practitioner has completed the WA Notifiable Disease Notification form
    - distribution and collation of information on the CJD REDCap surveillance form
    - liaising with the ANCJDR and sending relevant documentation to the Registry i.e. WA Disease Notification form, REDCap form, copies of diagnostic test results and any neuropathology reports.

- Medical practitioners are to liaise with the ANCJDR to assist with clarification of cases and use their services to enhance ante-mortem diagnostics including the 14-3-3 protein or RT-QuIC cerebrospinal fluid test and genetic testing. In addition, medical practitioners are required to notify cases to the ANCJDR, the office responsible for assisting the Australian Department of Health with the ongoing surveillance of CJD cases in Australia.

- Any occupational exposure in healthcare staff must be reported and managed as per local reporting procedures.

- Although vCJD is excluded from the scope of this document, if a patient is suspected to have vCJD, Health Service Providers must notify the Director, CDCD, Department of Health, immediately.

5. Relevant Legislation

CJD is a notifiable infectious disease and reporting of confirmed, probable and possible cases is a mandatory requirement pursuant to Part 9, Division 2 *Public Health Act 2016*.

6. Additional Resources

- Australian Government. Department of Health and Aged Care Creutzfeldt–Jakob disease – Infection control guidelines

- Australian Government. Department of Health and Aged Care. Communicable Disease Network Australia. Creutzfeldt–Jakob disease (CJD) – Surveillance case definition.

- CJD Support Group Network - offers support, information and assistance for family members and friends of patients suffering with suspected CJD and other prion disease and for those at increased risk of developing CJD.

7. Guideline Contact

Enquiries relating to this Guideline may be directed to: Infection Prevention Policy Surveillance Unit (IPPSU).

Directorate: Communicable Disease Control Directorate

Email: IPPSU@health.wa.gov.au

8. Document Control

Guideline number	Version	Published	Review Date	Amendments
0028	V.1.0	03/06/2025	03/06/2028	Original version. Contents of the rescinded Mandatory Policy transfer to Guideline.

9. Approval

Approved by	Dr Paul Armstrong, Director, Communicable Disease Control Directorate, Department of Health
Approval date	14/04/2025

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