Government of Western Australia
Department of Health

Policy Frameworks
MP 0120/19
Effective from: 13 May 2022

Management and Reporting of Creutzfeldt-Jakob Disease Policy

1. Purpose

The *Management and Reporting* of *Creutzfeldt-Jakob Disease (CJD) Policy* describes the mandatory infection prevention management and the reporting requirements for CJD to minimise the risk of transmission of CJD in Western Australian healthcare facilities.

This Policy will ensure that patients with suspected CJD shall have access to appropriate evidence-informed healthcare without discrimination or disadvantage.

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 transmissible spongiform encephalopathies (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.

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.

Although cases of CJD have been reported in healthcare staff, 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.

This Policy is a mandatory requirement under the *Public Health Policy Framework* pursuant to section 26(2)(c) of the *Health Services Act 2016*.

In addition, 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*.

2. Applicability

This Policy is applicable to the following Health Service Providers:

- Child and Adolescent Health Service
- East Metropolitan Health Service
- North Metropolitan Health Service
- South Metropolitan Health Service
- Western Australia Country Health Service.

The requirements contained within this Policy are applicable to the services purchased from contracted health entities where it is explicitly stated in the contract between the contracted health entity and the State of Western Australia or Health Service Provider. The State of Western Australia or Health Service Provider is responsible for ensuring that any obligation to comply with this Policy by the contracted health entity is accurately reflected in the relevant contract and managed accordingly.

3. Policy Requirements

3.1. CJD Infection Prevention Management

Health Service Providers must:

3.1.1. Comply with the risk assessment approach and infection prevention requirements outlined in the Australian Department of Health Infection Control Guidelines for Creutzfeldt-Jakob disease (the Guidelines).

3.1.2. Develop local procedures to effectively screen, identify, risk assess and manage patients and reusable equipment to minimise the risk of CJD transmission.

3.1.3. 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.

3.1.4. Through the medical practitioner, clearly communicate any reasons for variations or delays in treatment to the patient.

3.1.5. Ensure all patients undergoing surgical or diagnostic procedures involving higher-infectivity tissue have their risk category determined in accordance with section 2.3 and appendix 3 of the Guidelines.

3.1.6. Use the risk assessment matrix as described in section 2.4 of the Guidelines, to identify whether routine reprocessing or additional reprocessing procedures will be required.

3.1.7. Follow the additional procedures as described in section 3, table 3 of the Guidelines. Single use instruments must be used, wherever possible, and when their use will not compromise patient care.

3.1.8. 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.

3.1.9. Arrange for deceased persons undergoing autopsy, with suspected CJD to be transported to the Royal Perth Hospital (RPH) mortuary. The Department of

Health Communicable Disease Control Directorate (CDCD), will cover all costs associated with transportation to and from RPH.

3.2. CJD Reporting and Management

3.2.1. 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.

3.2.2. The notifying medical practitioner and medical/nursing staff, where the case may have received care within two years of diagnosis, must cooperate with all investigations relating to a CJD notification that is conducted by the Department of Health, Infection Prevention Policy and Surveillance Unit (IPPSU) to assess any public health risk and in consultation with the Australian National CJD Registry (ANCJDR).

3.2.3. All medical practitioners who identify a possible, probable or confirmed case of CJD, including sporadic, familial or acquired cases are required to complete a statutory notification to the CDCD pursuant to Part 9, Division 2 of the *Public Health Act 2016*.

3.2.4. 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 4-3-3 protein 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.

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

3.2.6. 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.

4. Compliance Monitoring

Health Service Providers are responsible for ensuring compliance with this Policy.

The IPPSU will undertake compliance monitoring to ensure all suspected or confirmed cases of CJD are notified in accordance with the *Public Health Act 2016*.

The IPPSU will undertake compliance monitoring to ensure Health Service Providers are compliant with all aspects of this Policy when undertaking the public health risk assessment for each case notified to the CDCD.

5. Related Documents

The following documents are mandatory pursuant to this Policy:

- Infection Control Guidelines – Creutzfeldt-Jakob Disease Australian Department of Health
- WA Health Metropolitan Notification Form and Rural Notification Form

6. Supporting Information

The following information is not mandatory but informs and/or supports the implementation of this Policy:

- Notification of Infectious Diseases and Related Conditions
- AS/NZS 4187 Cleaning, disinfecting and sterilizing reusable medical and surgical instruments and equipment, and maintenance of associated environments in health care facilities

7. Definitions

The following definition(s) are relevant to this Policy.

Term	Definition
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.
Contracted health entity	A non-government entity that provides health services under a contract or other agreement entered into with the Department CEO on behalf of the State, a health service provider or the Minister.
Medical practitioner	means a person registered under the *Health Practitioner Regulation National Law (Western Australia)* in the medical profession.
Patient risk categories	Patients are categorised into high-risk, low-risk and background risk for CJD transmission risk. Refer to section 2.3 of the Guidelines.
Reprocessing	The cleaning, disinfection and when applicable the sterilisation of reusable medical devices in accordance with AS/NZS 4187.
Reusable medical devices	A reusable medical device is any device designated or intended by its manufacturer as suitable for reprocessing and reuse. It is not a device designated or intended by the manufacturer for single use.
Tissue infectivity	Human body tissues and fluids have known or predicted infectivity for CJD. They are categorised as high or medium infectivity and low or no detectable infectivity. Refer to Table 1 of the Guidelines.

8. Policy Contact

Enquiries relating to this Policy may be directed to:
Title: Infection Prevention Policy and Surveillance Unit
Directorate: Communicable Disease Control Directorate
Email: hiswa@health.wa.gov.au

9. Document Control

Version	Published date	Effective from	Review date	Amendment(s)
MP 0120/19	3 September 2019	3 September 2019	February 2022	Original version
MP 0120/19 v.2.0	13 May 2022	13 May 2022	May 2025	Policy review with major amendments as stated below:
• Transferred to new mandatory policy template • Policy title amended to clearly reflect the subject of the policy • Updated purpose section to ensure clarity and to include some information previously contained in Appendix 1 • Applicability updated to list the applicable health service providers • New policy requirements relating to the development of local procedures, staff training, transporting deceased persons with suspected CJD for autopsy, liaising with the ANCJDR for assistance and reporting of occupation exposure and vCJD • Compliance Monitoring updated to link to the policy requirements • Related documents updated to include statutory notification forms • Supporting information updated to include new information and remove the previous Appendix 1 • Definitions amended to include additional terms.				

10. Approval

Approval by	Dr David Russell-Weisz, Director General, Department of Health
Approval date	28 August 2019

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