Government of Western Australia
Department of Health

Policy Frameworks
MP 0120/19
Effective from: 3 September 2019

Creutzfeldt-Jakob Disease Policy

1. Purpose

The *Creutzfeldt-Jakob Disease Policy* (CJD) sets out the infection prevention management procedures and the reporting requirements for CJD that are to be implemented to minimise the risk of transmission of CJD in Western Australian (WA) healthcare facilities.

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 the prion protein (a normal cell surface glycoprotein).

The infective agent of CJD, the prion, is resistant to routine reprocessing procedures used for reusable medical devices (RMDs) for example surgical instruments. Although transmission of CJD in the healthcare setting is very rare, healthcare workers need to be aware of the potential for transmission via RMDs contaminated with the prion, and the need to implement additional reprocessing procedures where an identified risk is determined based on the infectivity of the tissue to which the RMD is exposed and the patient risk factors for CJD.

Any patient with suspected CJD shall have access to appropriate evidence-based healthcare without discrimination or disadvantage.

Variant CJD (vCJD) is linked to bovine spongiform encephalopathy and is excluded from the scope of this document as vCJD has not been reported in Australia.

This Policy is a mandatory requirement under the Public Health *Policy Framework* pursuant to sections 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.*

This Policy supersedes *Operational Directive 0456/13 Creutzfeldt - Jakob disease (CJD) Risk Assessment and Management.*

2. Applicability

This Policy is applicable to all Health Service Providers, excluding Health Support Services.

3. Policy requirements

CJD Infection Prevention Management

Health Service Providers are required to comply with the risk assessment approach and infection prevention guidance articulated in the Australian Department of Health *Infection Control Guidelines for Creutzfeldt-Jakob disease* (the Guidelines).

3.1. Health Service Providers must ensure all patients undergoing surgical or diagnostic procedures involving higher-infectivity tissue shall have their patient risk category determined as per Section 2.3 and Appendix 3 of the Guidelines.

3.2. On the basis of the patient's risk as being high, low or background, the risk assessment matrix at section 2.4 of the Guidelines is to be used to identify whether routine reprocessing or additional reprocessing procedures will be required.

3.3. In the event a patient is identified to be in a high or low-risk category for CJD and undergoing a procedure involving higher-infectivity tissue, the Health Service Provider must have a documented action plan initiated to ensure admission and treatment is not delayed. It is essential to ensure patient care is not compromised and the patient is not discriminated against. Any reasons for variations or delays in treatment must be clearly communicated to the patient in order to encourage all patients with risk factors for CJD to disclose their risk status.

3.4. Any patient in a high or low-risk category and undergoing a procedure involving higher-infectivity tissue will require implementation of additional procedures. All staff involved in the care of the patient, equipment reprocessing or environmental cleaning are to be fully informed of the implementation of these additional procedures and have knowledge to carry out the additional procedures.

3.5. Where additional procedures are determined to be required, Health Service Providers are required to follow the requirements set out at Section 3, Table 3 of the Guidelines. Single use instruments and equipment should be used, wherever possible, and when their use will not compromise patient care.

CJD Reporting and Management

3.6. The Director of Medical / Clinical Services of each Health Service Provider are required to notify the Director, Communicable Disease Control Directorate (CDCD) Department of Health, should an adverse event arise, that is, where after routine reprocessing, RMDs used on a patient with asymptomatic CJD have subsequently been used unknowingly on other patients.

Note that in the event of an adverse event, the Director, CDCD will assume responsibility for the investigation, equipment management, patient risk assessment and the scope of a look-back investigation where required.

3.7. On receipt of a CJD notification, the Department of Health Healthcare Associated Infection Unit (HAIU) will undertake a public health risk assessment in consultation with the Australian National CJD Registry (ANCJDR), the notifying Medical Practitioner and Medical/Nursing staff where the case may have received care within two years of diagnosis.

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

3.9. In addition, medical practitioners are required to notify cases to the ANCJDR which has responsibility for assisting the Australian Department of Health with the ongoing surveillance of CJD cases in Australia.

4. Compliance monitoring

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

The HAIU undertakes the following compliance monitoring activities in relation to the policy requirements:

- monitors CJD notifications from Health Service Providers
- monitors CJD notifications received directly from the ANCJDR
- liaises with the ANCJDR to identify those cases not notified to CDCD by Medical Practitioners and ensures corrective action is undertaken.

5. Related documents

The following documents are mandatory pursuant to this Policy:

- Infection Control Guidelines – Creutzfeldt-Jakob Disease Australian Department of Health

6. Supporting information

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

- Appendix 1: Creutzfeldt-Jakob Disease – Additional Information

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.
Medical Practitioner	A doctor of medicine registered and accredited to practice within Australia.
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.
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 Guidelines.
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.

8. Policy contact

Enquiries relating to this Policy may be directed to:

Title: Healthcare Associated Infection Unit

Directorate: Communicable Disease Control Directorate

Email: hiswa@health.wa.gov.au

9. Document control

Version	Published date	Effective from	Review date	Effective to	Amendment (s)
MP0120/19	3 September 2019	3 September 2019	February 2022	Current	Original version

10. Approval

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

APPENDIX 1: Creutzfeldt-Jakob Disease – Additional Information

1. Variant Creutzfeldt-Jakob Disease

Although vCJD is excluded from the scope of this document, if a patient is suspected to have vCJD, the Director, Communicable Disease Control Directorate (CDCD), Department of Health, must be contacted immediately. For the purpose of this document, the term CJD is used to describe all forms of human TSE (sporadic, inherited and acquired) except vCJD.

2. Occupational Exposure

Although cases of CJD have been reported in HCWs, there have been no confirmed cases linked to occupational exposure. There is no epidemiological evidence to indicate that HCWs are at an increased occupational risk for acquiring CJD. Any occupational exposure should be reported in accordance with local HCF procedures and managed in accordance with local HSP protocols.

3. Post Mortem Procedures

In WA, all deceased persons for autopsy, who are suspected CJD or high or low-risk for CJD are required to be transported to the Royal Perth Hospital mortuary, where there must be staff who are appropriately trained in CJD infection prevention and control procedures.

The CDCD provides the funding for the transport of these patients to and from the RPH mortuary and funeral service companies should be instructed to invoice the CDCD, WA Department of Health.

4. Additional Resources

The Australian National CJD Registry (ANCJDR)

The Florey Institute of Neurosciences and Mental Health

30 Royal Parade, PARKVILLE, Victoria 3052

Phone: (03) 8344 1949 Fax: (03) 9349 5105

Email: ancjd-reg@unimelb.edu.au Web: http://ancjdr.path.unimelb.edu.au/

Office hours

Registry office: Weekdays 9am – 5 pm.

Specimen reception: Weekdays 8am – 4 pm

CSF samples collected at times when the ANCJDR is closed can be stored frozen until the ANCJDR reopens.

Closed on Victorian public and university holidays

The ANCJDR conducts ongoing surveillance of CJD in Australia. The registry acts as a resource to assist with clarification of cases into high-low-background risk for CJD. They offer diagnostic services to enhance ante-mortem diagnostics; the 14-3-3 protein CSF test and genetic testing. The ANCJDR is a World Health Organisation reference centre for human TSE.

CJD SUPPORT GROUP NETWORK (CJDSGN)

Phone: 1800 052 466 (Toll Free). This is a 24 hour hotline.
Email: contactus@cjdsupport.org.au
Web: http://www.cjdsupport.org.au

The CJDSGN is funded by the Australian Department of Health. It is contracted to assist and support people and families affected by CJD. The CJDSGN is also a source of information for health professionals working with Australians affected by CJD and other prion diseases.

AUSTRALIAN HUMAN PITUITARY HORMONE PROGRAM

For information and queries about hormone recipients, contact the Department of Health, Canberra. Phone: 1800 802 306 (0900–1700 EST)

WORLD HEALTH ORGANIZATION

The WHO infection control guidelines for transmissible spongiform encepahalopathies can be found here.

This document can be made available in alternative formats on request for a person with a disability.