Subject: Creutzfeldt-Jakob Disease (CJD) Risk Assessment and Management

Compliance with this Operational Directive is mandatory for all Western Australian public healthcare facilities and those licensed private healthcare facilities contracted to provide services to public patients.

This Operational Directive describes the infection prevention and control procedures that are required to be implemented to minimise the risk of transmission of Creutzfeldt-Jakob disease (CJD) in healthcare settings.

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A/DIRECTOR GENERAL
DEPARTMENT OF HEALTH WA

This information is available in alternative formats on request for a person with a disability.
1. INTRODUCTION

The infective agent of Creutzfeldt-Jakob disease (CJD), the prion, is resistant to routine reprocessing procedures used for surgical instruments or diagnostic equipment.

This Operational Directive describes the infection prevention and control procedures that are required to minimise the risk of transmission of CJD in healthcare facilities. It is based on the Creutzfeldt-Jakob Disease Infection Control Guidelines, revised by the Communicable Diseases Network Australia (CDNA), and released by the Commonwealth Department of Health and Ageing in January 2013.¹ This directive should be read in conjunction with the guidelines. All patients with suspected CJD should have access to appropriate evidence-based healthcare without discrimination and 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. If a patient is suspected to have vCJD, the Communicable Disease Control Directorate (CDCD) must be contacted immediately.¹

2. 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 Human 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). For the purpose of this document, the term CJD is used to describe all forms of TSE except vCJD, as outlined in Table 1.

Table 1: Categories of CJD disease

<table>
<thead>
<tr>
<th>Sporadic CJD</th>
<th>Inherited CJD</th>
<th>Acquired CJD</th>
</tr>
</thead>
<tbody>
<tr>
<td>There is no recognised cause</td>
<td>• Familial CJD</td>
<td>• Healthcare associated (iatrogenic) CJD</td>
</tr>
<tr>
<td></td>
<td>• Gerstmann-Sträussler-Scheinker disease (GSS)</td>
<td>• Kuru</td>
</tr>
<tr>
<td></td>
<td>• Fatal Familial Insomnia (FFI)</td>
<td></td>
</tr>
</tbody>
</table>

Although transmission of CJD in the healthcare setting is very rare, healthcare workers (HCWs) need to be aware of the potential for transmission by instruments or equipment contaminated by CJD. As the prion is resistant to routine reprocessing procedures, it is essential that HCFs implement the risk assessment process and the additional procedures outlined in this document to minimise the risk for CJD transmission. The decision to implement additional procedures is based on the infectivity of the tissue to which the instrument is exposed and the patient risk factors for CJD.

For patients in long term residential, palliative or community care facilities and for routine hospital procedures that do not involve exposure to higher-infectivity tissues for CJD, standard precautions and routine reprocessing procedures are all that is required for the management of suspected CJD patients or patients who are at high or low-risk for CJD.

3. DIAGNOSIS

There is currently no minimally invasive test available to detect CJD infection before the onset of symptoms. A pre-symptomatic period exists during which disease transmission is presumed to be possible. Definitive diagnosis of CJD is by neuropathological examination of brain tissue following biopsy or autopsy.
However, pre-mortem brain biopsy is not recommended as a routine procedure to confirm clinical suspicion of CJD. Methods which may assist in the diagnosis of CJD, and the exclusion of other causes of subacute dementia in symptomatic patients, include:

- electroencephalograph (EEG)
- presence of protein 14-3-3 in cerebrospinal fluid (CSF)
- magnetic resonance imaging (MRI)
- direct amplification of misfolded prion protein (PrP\text{Sc}) in the CSF using Real Time-Quaking Induced Conversion (RT-QUiC).

4. MODES OF TRANSMISSION

The majority of CJD cases are sporadic. Both sporadic and familial forms of CJD are caused by prion mutations and are not acquired by transmission of the disease directly from other people. However, there is evidence of healthcare associated transmission through the use of neurosurgical instruments contaminated with central nervous system tissue and through contaminated tissue implants or products (e.g. dura mater grafts, corneal grafts and pituitary products). There is no epidemiological evidence that CJD can be transmitted through respiratory normal social or sexual contact, mother-to-child transmission or via blood or blood products.

5. INFECTIVITY OF HUMAN TISSUE

The known or predicted infectivity of body tissues and fluids of symptomatic and asymptomatic patients with CJD are listed in Appendix 1.

6. PATIENT RISK CATEGORIES

The risk categories used to identify individuals who may pose a risk of transmitting CJD are described in Table 2. The specific classification criteria for high and low-risk individuals are outlined in Appendix 2 and 3 and should be referenced when assigning risk category.

Table 2: Patient risk categories for CJD

<table>
<thead>
<tr>
<th>High-risk</th>
<th>Low-risk</th>
<th>Background risk</th>
</tr>
</thead>
<tbody>
<tr>
<td>People who represent a <strong>definite</strong> risk of CJD transmission. These patients typically report neurological symptoms and display neurological signs of disease.</td>
<td>People who represent a <strong>potential</strong> risk of CJD transmission. These patients may report neurological symptoms or be showing neurological signs or may have an identified risk factor for CJD.</td>
<td>The general population who represent <strong>no</strong> identified increased risk of CJD transmission. The rate of CJD deaths in the general Australian population is 1.15 per million people per year.</td>
</tr>
</tbody>
</table>

(Refer to Appendix 2 ) (Refer to Appendix 3)
7. **RISK ASSESSMENT AND MANAGEMENT**

7.1 **Assessment**

All patients undergoing surgical or diagnostic procedures involving higher-infectivity tissue described in Appendix 1 shall have their patient risk category (Table 2) determined prior to the procedure. A questionnaire template to assist with risk assessment for CJD is available (Appendix 4).

The patient risk assessment questionnaire shall be administered to patients by the medical practitioner performing the procedure, prior to consent being obtained. The completed questionnaire shall be included in the patient medical record.

7.2 **Management**

Healthcare facilities (HCFs) must ensure processes are in place to ensure the risk assessment (Appendix 5) is undertaken, documented and actioned appropriately. 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 HCF is to have an action plan in place 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 to disclose their risk status.

The planned procedure may require modification or initiation of processes for the implementation of additional procedures. All HCWs involved in the care of the patient, equipment reprocessing or environmental cleaning are to be fully informed of the proposed implementation of these additional procedures.

7.3 **Risk assessment matrix**

The risk assessment matrix (Table 3) is used to identify whether routine reprocessing procedures or additional procedures are required. Additional procedures are only to be implemented when:

- the diagnostic or therapeutic procedure performed involves the exposure of higher-infectivity tissue (Appendix 1)

**AND**

- the patient is identified as high or low-risk for CJD (Table 2, Appendix 2 and 3).

<table>
<thead>
<tr>
<th>Patient CJD Risk Categories</th>
<th>Procedures Involving Exposure to Higher-Infectivity Tissue</th>
<th>Procedures Involving Exposure to Lower-Infectivity Tissue</th>
</tr>
</thead>
<tbody>
<tr>
<td>HIGH RISK</td>
<td>Use additional procedures</td>
<td>Use routine reprocessing procedures*</td>
</tr>
<tr>
<td>LOW RISK</td>
<td>Use additional procedures</td>
<td>Use routine reprocessing procedures</td>
</tr>
<tr>
<td>BACKGROUND RISK</td>
<td>Use routine reprocessing procedures</td>
<td>Use routine reprocessing procedures</td>
</tr>
</tbody>
</table>

* See note 3 in Appendix 1: recommendations relating to surgery involving the cornea or anterior segment of the eye.
8. ADDITIONAL PROCEDURES

The relevant additional procedures that apply to the handling and reprocessing of surgical instruments and diagnostic equipment are described in Table 4.

Single use instruments and equipment should be used wherever possible and when their use will not compromise patient care.\(^2\)

If reusable instruments are used they shall either be destroyed or only reprocessed in the following circumstances:

- they need to be quarantined pending clarification of patient risk status
- they are used exclusively on a patient who is high or low-risk for CJD
- they are used exclusively for autopsies on cadavers.

8.1 Additional reprocessing procedures

The prion is resistant to routine reprocessing procedures using heat and chemical sterilisation. Chemicals that are known to have some activity against prions include anionic detergents, hypochlorite and harsh acids and alkalis, however, their practical effectiveness and use in reprocessing is influenced by prior cleaning and prion strain. Multiple reprocessing steps will reduce infectivity, but the number of reprocessings which render instruments completely safe, has never been established.

All reprocessing procedures must be in accordance with the Australian and New Zealand Standard AS/NZS 4187 and any subsequent revisions.\(^3\) The additional reprocessing procedures to be implemented are described in Table 4, in conjunction with the following requirements for cleaning:

- instruments shall be thoroughly washed and cleaned in anionic detergent prior to further additional reprocessing procedures
- new enzymatic cleaning products for surgical equipment are available that have been registered on the Australian Register of Therapeutic Goods (ARTG). However, there is currently insufficient evidence of the efficacy of these products in the removal or inactivation of prions, therefore, their use is not recommended for use as part of additional reprocessing procedures
- HCFs must ensure that new cleaning products or systems that come on the market in the future are registered on the Australian Register of Therapeutic Goods (ARTG) together with a specific statement confirming that the product or procedure is indicated for the removal or inactivation of prions
- instruments are not to be exposed to high-level disinfectants, such as glutaraldehyde, prior to cleaning as they enhance the adherence of prions to surfaces. Harsh acids and alkalis are also not recommended due to occupational health issues and potential for damage to instruments and equipment
- ultrasonic cleaners may be used as part of the additional reprocessing procedures.

9. INSTRUMENT TRACKING

HCFs and companies that provide loan, demonstration or trial equipment for use in procedures exposing higher-infectivity tissues shall have systems in place to track individual instruments and equipment to the level of the individual patient. Tracking of instruments and trays will minimise the number of patients implicated in a look-back, where a patient thought to be at background risk is subsequently diagnosed with CJD.
### Table 4: Additional procedures

<table>
<thead>
<tr>
<th>Activity</th>
<th>Additional Procedures</th>
</tr>
</thead>
</table>
| **Operating Theatre Preparation and Set-up** | • Schedule patients to allow for appropriate preparation and cleaning following the procedure.  
• Remove unnecessary equipment and supplies from the operating suite.  
• Where appropriate, and it will not present a fire hazard, cover equipment not exposed to higher-infectivity tissue with plastic to protect from splash. Incinerate after use. |
| **Personal Protective Equipment** | • Wear fluid repellent single use PPE including gloves, gowns, masks and protective eyewear or full face-shields. Incinerate after use.                                                                                          |
| **Anaesthetic Equipment**       | • Routine management and reprocessing.                                                                                                                                                                                  |
| **Surgical Drapes**            | • All drapes should be single use and incinerate after use.                                                                                                                                                            |
| **Tracking of Instruments**     | • HCFs performing procedures exposing higher-infectivity tissue and companies providing loan equipment shall have systems in place to track individual reusable items to the level of the individual patient, to minimise the number of patients implicated in a look-back. |
| **Instrument Use**             | • Use single-use instruments wherever possible and incinerate OR  
• Reusable instruments should be kept for exclusive use on an individual patient or quarantined. Reprocess separately and quarantine instruments pending determination of risk status. If determined high or low-risk patient: incinerate OR keep for the exclusive use of the patient and incinerate on completion of the therapy OR place back in circulation if risk found to be background only. |
| **Intra-operative Handling of Instruments** | • Separate instruments used on higher-infectivity tissue from general instruments to avoid cross-contamination.  
• Where possible, to prevent tissue residues drying on instruments during surgery, regularly wipe instruments with a pre-moistened radio-opaque pack or keep in a tray/kidney dish covered with a moistened radio-opaque pack. |
| **Reprocessing Instruments** (Only for quarantine, exclusive patient use, or autopsies) | • To prevent drying prior to reprocessing, immerse instruments in sterile water in a dedicated container after surgery.  
• Reprocess separately.  
• Do not mix with any other instruments or equipment at any stage.  
• Instruments are not to be exposed to chemical disinfectants prior to initial cleaning.  
• For cleaning information refer to Section 8.1.  
• Steam sterilise at 134°C for 3 minutes  
• Any item identified as difficult to clean shall be destroyed or advice sought from the National CJD Incident Panel. |
| **Quarantine Process**         | • Ensure instruments are separated, reprocessed, contained, labelled and stored in a secure environment pending incineration or return to circulation once risk status determined.  
• Any quarantine system must minimise the risk of accidental reintroduction of potentially infected equipment. |
| **Collection of Specimens**     | • Standard specimen collection, handling and transportation. Tissue or fluid samples should be collected into sealed containers with the CJD risk status of the patient clearly labelled to alert laboratory workers. Higher- infectivity tissues should be handled under Physical Containment Level 2 (PC2). |
| **Environmental Cleaning and Spills** | • Routine containment and cleaning procedures should be used for the whole operating room, including surfaces, unless a spill of higher-infectivity tissue has occurred.  
• Spill-Kits containing either 20,000ppm (free chlorine) sodium hypochlorite or 1M sodium hydroxide (NAOH) are to be available in areas of increased risk such as neurosurgery operating rooms, mortuaries and laboratories. Occupational Health and Safety recommendations and material data sheets (MDS) must be available.  
• For spillage of higher-infectivity tissues from low or high-risk CJD patients decontaminate the contaminated area by exposing it to freshly prepared solution for 1 hour at room temperature and then rinse with water. Surfaces that cannot tolerate NaOH or sodium hypochlorite should be cleaned using anionic detergent. |
| **Waste Disposal**             | • All items including spent specimens, operative tissue and body fluids involved in the case, are to be disposed of in clinical waste for incineration or other appropriate alternate approved method of medical waste destruction. Routine management of sharps disposal is to occur. |
| **Endoscopes**                 | • Any endoscope used in a procedure in a high or low-risk patient where higher-infectivity tissue has been exposed (e.g. ventriculoscope) shall be destroyed by incineration or kept exclusively for use on that patient.  
• In all other situations, endoscopes may be reprocessed using routine processes  
• Note: nasal endoscope procedures do not reach the olfactory epithelium. |
10. NOTIFICATION AND ADVERSE EVENT MANAGEMENT

CJD is scheduled as a notifiable disease in all Australian States and Territories and notification is required to the CDCD for all cases in which a strong clinical suspicion for CJD exists or on receipt of confirmatory pathology.

Since there is no test to reliably detect CJD prior to onset of symptoms, it is possible that surgical instruments used on a patient with asymptomatic CJD might subsequently be used unknowingly on other patients after routine reprocessing, with a potential risk of transmission. In this event, the equipment having direct contact with higher-infectivity tissue should be identified via instrument tracking systems and quarantined (refer Table 4 and Appendix 5).

The Executive of the HCF and the Director of CDCD are to be notified of all possible adverse events and will assume responsibility for the investigation, equipment management, patient risk assessment and the scope of a look-back investigation if required. The National CJD Incident Panel, established by the Australian Government, is available to provide expert advice in the event of an adverse event involving CJD.

11. 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. There are no extra requirements following exposure to tissues of a patient with CJD or a patient in the high or low-risk category.\textsuperscript{1, 4}

12. ORGANS AND TISSUES FOR TRANSPLANTATION

For information on organ and tissue transplantation, including people who should be excluded from the routine donation of organs, tissues, blood and plasma, refer to the \textit{Creutzfeldt-Jakob Disease Infection Control Guidelines, 2013}.\textsuperscript{1}

13. DENTAL PROCEDURES

Instruments used on all patients, undergoing routine dental procedures in contact with lower-infectivity tissue (Appendix 1), can undergo routine reprocessing procedures.

Dental procedures on high or low risk patients involving higher-infectivity tissues should be performed at a facility that has HCWs who are familiar with additional procedures for CJD (Table 4).

Dental procedures (and an example of the higher-infectivity tissue exposed) requiring additional procedures would include:

- major oral surgical procedures such as maxillectomy with orbit enucleation (optic nerve)
- injection of the trigeminal ganglion (cranial nerve exposure)
- oral surgical cancer procedures that also combine a neurosurgical approach (potential brain tissue, central nerve exposure).

Dentists should contact the Australian Dental Association for further advice on the management of patients identified as high or low-risk for CJD.
14. POST MORTEM PROCEDURES

In WA, all patients 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. Facilities should contact the CDCD to discuss funding arrangements for the transport of these patients to and from the mortuary.

Additional procedures are required for autopsies involving exposure to higher-infectivity tissue in patients who are high or low-risk or suspected CJD (Table 4). Single use instruments or a set of instruments dedicated to suspected CJD patients should be used for autopsies and must be kept separate to any instruments used to harvest organs and tissues for donation. Following autopsy, the body should be sealed in plastic to contain fluid leakage. For further autopsy requirements, refer to the Creutzfeldt-Jakob Disease Infection Control Guidelines, 2013.¹

Cadavers from high or low-risk patients should not be used for teaching purposes.

15. FUNERAL INDUSTRY PROCEDURES

If the bodies of CJD patients have not undergone a brain autopsy, the transportation, preparation, disinfection, and final disposition can be safely performed using standard precautions.

Transport following an autopsy can be performed as above, however it is recommended that the body is placed in a leak proof bag which is lined with absorbent material to absorb leakage of body fluid. In instances where there is excess fluid, a double bag can be utilized

For further information for the funeral industry refer to the Creutzfeldt-Jakob Disease Infection Control Guidelines, 2013.¹

16. ADDITIONAL RESOURCES

The Australian National CJD Registry (ANCJDR), Melbourne
Phone: (03) 8344 1949   Fax: (03) 9349 5105
Email: ancjd-reg@unimelb.edu.au   Web: http://ancjdr.path.unimelb.edu.au/

The ANCJDR conducts ongoing surveillance of TSE in Australia. The registry will act 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)
Email: contactus@cjdsupport.org.au   Web: http://www.cjdsupport.org.au

The CJDSGN is funded by the Department of Health and Ageing. 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 Department of Health and Ageing, Canberra
Phone:  1800 802 306   (0900–1700 EST) for information and queries about hormone recipients.
17. REFERENCES


3. Australian/New Zealand Standard AS/NZS 4187 (2003), Cleaning, disinfecting and sterilizing reusable medical and surgical instruments and equipment, and maintenance of associated environments in health care facilities.

**APPENDIX 1**

**KNOWN OR PREDICTED INFECTIVITY OF HUMAN BODY TISSUES AND FLUIDS FOR CJD**

<table>
<thead>
<tr>
<th>Infectivity Category</th>
<th>Tissues</th>
<th>Secretions</th>
</tr>
</thead>
</table>
| **HIGH or MEDIUM Infectivity**(1) | • Brain  
• Dura mater  
• Spinal cord  
• Pituitary gland  
• Posterior segment of the eye*  
• Cranial and dorsal root ganglia  
• Olfactory epithelium (not normally encountered in routine nasal or sinus surgery) | |
| **LOW or NO DETECTABLE Infectivity**(2) | • Cornea (3)  
• Anterior segment of eye(3)  
• Kidney  
• Liver  
• Lung  
• Lymph nodes / spleen / tonsils  
• Placenta  
• Uterus  
• Adipose tissue  
• Adrenal gland  
• Blood and blood products  
• Bone marrow  
• Oral tissue (teeth, gingival tissue, dental pulp)  
• Heart muscle  
• Intestine  
• Peripheral nerve  
• Prostate  
• Skeletal muscle  
• Ovaries  
• Testes  
• Thyroid gland  
• CSF  
• Amniotic fluid  
• Faeces  
• Breast milk  
• Nasal mucus  
• Saliva  
• Semen  
• Serous exudate  
• Sweat  
• Tears  
• Urine | |
### INDIVIDUALS IN THE HIGH-RISK CATEGORY FOR CJD*

<table>
<thead>
<tr>
<th>Classification of CJD</th>
<th>Clinical signs and risk factors</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>1. Sporadic TSE</strong></td>
<td><strong>Clinical signs</strong></td>
</tr>
<tr>
<td>1.1 Definite</td>
<td>I. Rapidly progressive dementia</td>
</tr>
<tr>
<td></td>
<td>II.</td>
</tr>
<tr>
<td></td>
<td>a) Myoclonus</td>
</tr>
<tr>
<td></td>
<td>b) Visual or cerebellar problems</td>
</tr>
<tr>
<td></td>
<td>c) Pyramidal or extrapyramidal features</td>
</tr>
<tr>
<td></td>
<td>d) Akinetic mutism</td>
</tr>
<tr>
<td>1.2 Probable (refer to clinical signs in adjacent column)</td>
<td>III. Typical EEG</td>
</tr>
<tr>
<td></td>
<td>rapidly progressive dementia (I), and at least 2/4 clinical signs of group II and typical EEG (III)</td>
</tr>
<tr>
<td></td>
<td>possible rapidly progressive dementia plus a positive 14-3-3 CSF assay.</td>
</tr>
<tr>
<td></td>
<td>possible rapidly progressive dementia (I), and 2/4 clinical signs of group II and duration of &lt;2 years.</td>
</tr>
</tbody>
</table>

| 2. Accidentally transmitted (iatrogenic) TSE | Recognised health care acquired risk factors: |
| 2.1 Definite | Treatment with human cadaver-derived pituitary growth hormone, human cadaver-derived pituitary gonadotrophin or human dura mater graft. |
| 2.2 Probable | Corneal graft recipient in which the corneal donor has been classified as definitely or probably having a human prion disease. |
|              | Exposure to surgical instruments that have come into contact with higher-infectivity tissues previously used in a case of definite or probable human prion disease. |
|              | The relevance of any exposure to disease causation must take into account the timing of exposure in relation to disease onset. |
|              | This list is provisional, as previously unrecognised mechanisms of human prion disease may occur. |

<table>
<thead>
<tr>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>3.1 Definite</td>
<td>A large number of PRNP mutations in the prion protein gene have been described. Refer to the CJD Infection Control Guidelines¹.</td>
</tr>
<tr>
<td>3.2 Probable</td>
<td>People who have had the PRNP gene sequenced and are shown not to carry the disease-linked mutation can be classified as ‘background’ risk, unless they have other demonstrated risk factors.</td>
</tr>
</tbody>
</table>

* Table Adapted from Creutzfeldt-Jakob Disease Infection Control Guidelines 2013.
**APPENDIX 3**

**INDIVIDUALS IN THE LOW-RISK CATEGORY FOR CJD***

<table>
<thead>
<tr>
<th>Low-risk patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>- People with a progressive neurological illness of less than one year’s duration, with or without dementia, for whom a determination to assign a high-risk status or background risk status cannot be made following competent professional review.</td>
</tr>
<tr>
<td>- People with a progressive neurological illness of less than one year’s duration, with or without dementia awaiting the outcome of a professional review to assign a high-risk status or background risk.</td>
</tr>
<tr>
<td>- Patients undergoing a diagnostic brain biopsy for progressive brain disease or patients undergoing neurosurgical investigations (including brain biopsy) or therapeutic procedures for a progressive disorder that includes dementia if &lt;1 year duration and where professional review is unable to assign a high-risk status or a background risk status.</td>
</tr>
<tr>
<td>- All genetically related members of any family in which there is a strong family history (two or more first degree relatives*) of dementia or neurological illness, and in which affected individuals have not been competently and completely assessed neurologically, specifically for CJD.</td>
</tr>
<tr>
<td>- Recipients of cadaver-derived human pituitary hormones (growth hormone and gonadotrophins) before 1986.</td>
</tr>
<tr>
<td>- Recipients of dura mater homografts or transdural neurosurgery before 1990 or neurosurgical patients for whom the use of dura mater homografts cannot be excluded by reference to patient records.</td>
</tr>
<tr>
<td>- Individuals who have been contacted by the Health Department as part of a look-back procedure from exposure to surgical instruments that had previously been used on high or medium infectivity tissues from patients later found to have contracted CJD are likely to have a very low, but unquantifiable risk for CJD that is thought to be above background risk. Until further information on the likely risk of these individuals is available, they are conservatively placed in a low risk category.</td>
</tr>
</tbody>
</table>

* Table Adapted from Creutzfeldt-Jakob Disease Infection Control Guidelines 2013.

* First degree relative: parent, sibling or child
  Note: second degree relative: grandparent, grandchild, aunt, uncle, niece, nephew, half-sibling.
IDENTIFICATION OF POTENTIAL CREUTZFELD-JAKOB DISEASE (CJD) RISK

Admitting Medical Practitioner to Complete

The following questions should be asked of a patient prior to undergoing surgery, investigations or a procedure involving any of the following higher-infectivity tissues:
- brain, pituitary gland or dura mater
- cranial and dorsal root ganglia
- spinal cord
- posterior segment of eye (includes retina and optic nerve)
- olfactory epithelium (not normally encountered in routine nasal or sinus surgery)

Note: for procedures of the anterior segment of the eye (includes cornea), which are lower-infectivity tissues, consider the use of single-use instruments in known high-risk patients.

If this is a repeat procedure and the following questions have already been answered, they do not need to be completed again providing the patient’s neurological condition remains unchanged.

To be completed by patient’s medical officer to determine risk status:

<table>
<thead>
<tr>
<th>QUESTIONS</th>
<th>YES</th>
<th>NO</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 Do you think the patient may have CJD?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2 Has the patient had two or more first or second-degree relatives* with CJD? (It is important to know about any relatives with CJD, but having a single affected relative with sporadic CJD does not place the patient in a low or high-risk category.)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>3 Does the patient have an unexplained progressive neurological illness of less than 12 months?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>4 Does the patient have a history of receiving human pituitary hormone for infertility or human growth hormone for short stature prior to 1986?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>5 Has the patient previously had surgery on the brain or spinal cord that included a dura mater graft prior to 1990?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>6 Has the patient been involved in a ‘look-back’ for CJD or shown you a ‘medical in confidence’ letter regarding their risk for CJD?</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

* First degree relative: parent, sibling or child
  Second degree relative: grandparent, grandchild, aunt, uncle, niece, nephew, half-sibling.

If the patient answers ‘YES’ to any of the above questions, please contact <insert name of responsible person> for further advice. Additional procedures may need to be implemented for reprocessing of equipment. DO NOT proceed to surgery prior to obtaining this advice.

Advice sought from: ................................................................................................................ <Print Name>
Admitting doctor: .................................................................................................................... <Print Name>
Signature: ................................................................................................................................. Date:  /  /  

Please use Patient Identification Label or Block Print

SURNAME MRN
GIVEN NAME
DOB SEX
DOCTORS NAME

Creutzfeldt-Jakob Disease (CJD) Risk Assessment and Management
Is the patient undergoing a procedure where higher-infectivity tissue will be exposed? (Appendix 1)
- brain, pituitary gland, dura mater
- cranial and dorsal root ganglia
- spinal cord
- posterior segment of the eye (includes retina and optic nerve)
- olfactory epithelium (not normally encountered in routine nasal or sinus surgery)

Note: for procedures of the anterior segment of the eye (includes cornea), which are lower-infectivity tissues, consider the use of single-use instruments in known high-risk patients.

Yes
- Proceed with procedure and routine reprocessing of instruments

No
- Is the patient classified as: High-risk (Appendix 2)?
  - or
  - Low-risk (Appendix 3)? for CJD transmission?

Yes
- Additional procedures required (Table 4)
  - Incinerate instruments immediately after use
  - OR
  - For those patients who are awaiting determination of risk status, reprocess reusable instruments separately and quarantine instruments pending determination of risk status of patient (then incinerate if deemed high or low-risk or reprocess and put back into circulation if risk is found to be background)
  - OR
  - Reprocess reusable instruments separately and keep for the exclusive use of an individual patient involved in a course of therapy (then incinerate when no longer required)

  Do not proceed with surgery/procedure until a management plan is initiated.

No
- Proceed with procedure and routine reprocessing of instruments