Policy Applies to:
All staff, Credentialed Specialists and Allied Health Professionals involved in the care and management of patients with possible, probable or confirmed Creutzfeldt-Jakob disease (CJD).

Related Standard:

Rationale:
Creutzfeldt-Jacob Disease (CJD) is a rare and fatal neuro degenerative disease that is caused by a prion (i.e. an infectious particle smaller than a virus). CJD is a notifiable disease in New Zealand. Although transmission of CJD in the health care setting is very rare, health care staff should be aware of the potential for transmission by contaminated instruments or via contaminated higher-infectivity tissues. The infective agent of CJD (the prion) is resistant to routine reprocessing, making the additional procedures outlined in this document essential for the treatment of patients with an identified risk of CJD infection.

Objectives:
- To identify at risk patients prior to a surgical procedure
- To provide appropriate care and management for patients known or suspected to have CJD

Implementation:

Risk assessment tools
- Known or predicted high or medium infectivity of tissues and fluids
- Patient risk categories
- Risk assessment matrix

The application of transmission-based precautions to minimise the risk of transmission of CJD is based on a risk assessment. The tissues or body fluids likely to be exposed during a procedure should be classified according to Table 1 and the patient risk category should be identified according to Table 2. The additional procedures that may apply as a result of the risk assessment are outlined in Table 3.

Prior to booking a patient for any procedure involving any higher infectivity tissue surgery, the specialist must complete a risk assessment (Appendix One – CJD Risk Assessment Questionnaire) so precautions can be put into place if needed. Preadmission staff as part of the patient admission process, will review the patient CJD status and record this on the preadmission form.
Table 1. Known or predicted infectivity of human body tissue

<table>
<thead>
<tr>
<th>Infectivity category</th>
<th>Tissues</th>
<th>Secretions &amp; Excretions</th>
</tr>
</thead>
<tbody>
<tr>
<td>High-infectivity or medium</td>
<td>Brain</td>
<td>CSF</td>
</tr>
<tr>
<td>infectivity or medium</td>
<td>Dura mater</td>
<td>Amniotic fluid</td>
</tr>
<tr>
<td>infectivity</td>
<td>Pituitary Gland</td>
<td>Breast milk</td>
</tr>
<tr>
<td>or medium infectivity</td>
<td>Spinal cord</td>
<td>Nasal mucus</td>
</tr>
<tr>
<td>or high infectivity or</td>
<td>Posterior eye (includes: posterior hyaloid face; retina; retinal</td>
<td>Saliva</td>
</tr>
<tr>
<td>medium infectivity</td>
<td>pigment epithelium; choroid; sub-retinal fluid; optic nerve)</td>
<td>Semen</td>
</tr>
<tr>
<td>or high infectivity</td>
<td>Cranial or dorsal root ganglia</td>
<td>Serous exudate</td>
</tr>
<tr>
<td>or medium infectivity</td>
<td>Olfactory epithelium</td>
<td>Sweat</td>
</tr>
<tr>
<td>Low-infectivity or no</td>
<td>Cornea</td>
<td>Tears</td>
</tr>
<tr>
<td>detectable infectivity</td>
<td>Anterior segment of eye</td>
<td>Urine</td>
</tr>
<tr>
<td></td>
<td>Kidney</td>
<td>Faeces</td>
</tr>
<tr>
<td></td>
<td>Liver</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Lung</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Lymph nodes/spleen/tonsil</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Placenta/Uterus</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Adipose tissue</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Adrenal gland</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Blood and blood products</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Bone and bone marrow</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Oral tissue (teeth, gingival tissue, dental pulp)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Heart muscle</td>
<td></td>
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<tr>
<td></td>
<td>Intestine</td>
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<tr>
<td></td>
<td>Peripheral nerve</td>
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<tr>
<td></td>
<td>Prostate</td>
<td></td>
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<tr>
<td></td>
<td>Skeletal muscle</td>
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<tr>
<td></td>
<td>Ovaries</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Testes</td>
<td></td>
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<tr>
<td></td>
<td>Thyroid gland</td>
<td></td>
</tr>
</tbody>
</table>

**Patient Risk Categories**

- **High-risk** – people who represent a definite risk of CJD transmission. These patients typically report neurological symptoms and display neurological signs of disease (refer Australian Government Guidelines Appendix 1).
- **Low-risk** – people who represent a potential risk of CJD transmission. These patients may report neurological symptoms or be showing neurological signs or may have an identified risk factor (refer Australian Government Guidelines Appendix 2).
Table 2 Risk Assessment Matrix

<table>
<thead>
<tr>
<th>Patient risk category</th>
<th>Procedures involving high infectivity or medium-infectivity tissues (see Table 1)</th>
<th>Procedures involving exposure to low or no infectivity tissues (see Table 1)</th>
</tr>
</thead>
<tbody>
<tr>
<td>High risk patient</td>
<td>Use additional procedures</td>
<td>Use routine/standard precautions</td>
</tr>
<tr>
<td>Low risk patient</td>
<td>Use additional procedures</td>
<td>Use routine/standard precautions</td>
</tr>
</tbody>
</table>

High Risk Reprocessing Procedures; in addition to routine reprocessing procedures


Surgical Procedures

If surgery is planned on a patient who has or is suspected of having CJD, Infection Prevention & Control, Operating Theatre staff, Sterile Services staff and the Waste Officer must be advised prior to admission as the case must be managed using additional procedures (refer to Table 3)

Refer to the CSSD CJD Procedure for Management of Instruments and Equipment procedures work manual
### Table 3 Additional Procedures required ONLY for patients identified as high-or-low CJD risk and undergoing procedures involving higher-infectivity tissue

<table>
<thead>
<tr>
<th>Activity</th>
<th>Additional Procedures</th>
</tr>
</thead>
</table>
| **Operating Room Preparation and Setup**      | Schedule patients to allow for preparation and cleaning following the procedure.  
Where appropriate and it will not present a fire hazard, cover equipment not exposed to higher-infectivity tissue with plastic wrap to protect from splash. Wrap to be disposed of in hazardous waste bags after use for incineration. |
| **Personal Protective Equipment**             | Wear fluid repellent single use PPE including gloves, gowns and full face shields. Incinerate after use.                                                                                                               |
| **Anaesthetic Equipment**                     | Routine management and reprocessing.                                                                                                                                                                                   |
| **Surgical Drapes**                           | All drapes should be single use and incinerated after use.                                                                                                                                                            |
| **Tracking of Instruments**                   | HCFs performing procedures exposing higher-infectivity tissue and companies providing loan equipment should 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 individual patient. Reprocess separately and quarantine instruments pending determination of risk status. If determined high-or-low: incinerate or keep for the exclusive use of the patient and incinerate on completion of therapy OR place back in circulation if risk found to be background only. |
| **Intra-operative Handling of Instruments**   | Where possible, separate instruments used on higher-infectivity tissue from other instruments to reduce risk of contamination. Where possible, to prevent tissue residues drying on instruments during surgery, regularly wipe instruments with a moistened radio-opaque pack or keep in a tray/kidney dish covered with a moistened radio-opaque pack. |
| **Reprocessing Instruments (For quarantine or exclusive patient use)** | To prevent drying prior to reprocessing immerse instruments contaminated with higher-infectivity tissue in sterile water in a dedicated container after surgery. Reprocess separately. Do not mix with any other instruments or equipment at any stage. Instruments should not be exposed to chemical disinfectants prior to initial cleaning. Steam sterilise at 134oC for 3 minutes. Any item identified as difficult to clean should be destroyed or advice sought from the Medical Officer of Health. |
**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 should minimise the risk of accidental reintroduction of potentially infected equipment.

**Collection of Specimens**

Standard specimen collection, handling and transportation. The specimens should be clearly labelled including a CJD risk alert to laboratory HCWs.

**Environmental Cleaning**

Routine containment and cleaning procedures apply unless major contamination with higher-infectivity tissue has occurred. Expose area with freshly prepared sodium hypochlorite solution for 1 hour and then rinse with water.

**Waste Disposal**

The Waste Officer should be notified in advance of the surgery and waste is to be collected in specifically labelled hazardous waste bags. This includes specimens / operative tissue / fluids / covers / sharp containers involved in the case. Sharps should be disposed of in single-use sharps containers. The Waste Officer will arrange with the SDHB mortuary for waste collection and disposal by incineration.

**Endoscopes**

Any endoscope+ used in a procedure in a high-or-low risk patient where higher-infectivity tissue has been exposed (e.g. ventriculoscope) should be destroyed by incineration or kept exclusively for that patient. In all other situations, endoscopes may be reprocessed using routine processes. + Normal nasal endoscope procedures do not reach the olfactory epithelium.

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**General Management of Patients in Hospital**

**Patient Management:**

- Patients may be nursed using Standard Precautions
- There are no additional requirements for environmental decontamination at ward level
- CJD is a notifiable disease in New Zealand and must be reported immediately on suspicion by the credentialed specialist to the Medical Officer of Health.

**Waste Procedures**

- With the exception of Operating Theatres and Sterile Services, CJD waste is treated as normal waste and disposed of as per Waste policy
- For disposal of CJD waste in theatres refer to Table 3.

**Dental Procedures**

- The risks of transmission of infection from dental instruments is considered low, providing optimal standards of infection prevention & control are maintained.
- Instruments used in routine dental procedures that come in contact with low-infectivity tissues (Table 1) in high or low-risk patient categories (Table 2) may be reprocessed using routine procedures.
• Instruments or equipment used in oro-facio-maxillary surgical procedures that come into contact with high-infectivity tissues in patients of high or low risk (Table 2) should be reprocessed using additional reprocessing procedures (refer page 6, Section 3.2 Australian Government Guidelines) for handling and reprocessing of surgical instruments and diagnostic equipment.

• Sterile Services must be notified to enable the CJD protocol to be implemented.

Neurology Services
• Reusable EMG needles, EEG electrodes, sensory testing pins and lumbar puncture needles shall be reprocessed through normal procedures where they do not have contact with high infectivity tissues.

• A single use neurological endoscope should be used where a diagnosis CJD is unclear.

Endoscopy
• Any endoscope used in a procedure in a high risk-or-low risk patient where high-infectivity tissue has been exposed should be disposed of or kept exclusively for that patient.

• In all other situations, endoscopes may be reprocessed using routine processes.

• The advice of the consultant carrying out an endoscopic procedure in the nasal cavity should be sought to determine whether a risk of contamination of the scope with olfactory epithelium can be excluded with confidence (normal nasal endoscopes procedures do not reach the olfactory epithelium). If not, this scope will have to be treated as high risk and should be disposed of or kept exclusively for that patient.

Laboratory Procedures
• Label all relevant samples and their request forms with “CJD risk sample” and notify the laboratories in advance.

Occupational Exposure
There is no evidence of occupational transmission of CJD to a healthcare worker (SHEA 2010). The highest potential risk results from exposure to high infectivity tissues through needle stick injuries with inoculation.

Evaluation:
A debrief with all key stakeholders will be held to evaluate the policy and process following presentation of a CJD case with debrief and any actions required reported to the Infection Prevention and Control Committee.

Appendix One – Classical Creutzfeldt - Jakob disease (CJD) Risk Assessment Tool

Associated Documents
External
• Australian Government Department of Health CJD Infection Control Guidelines (2013)
• Government UK (2015) Minimise transmission of CJD and vCJD in healthcare settings
• Health Act, Notifiable Diseases List, Ministry of Health
• Management of Creutzfeldt-Jakob Disease (CJD) and related disorders, Lippincott, July 2012
• Creutzfeldt- Jacob Disease (CJD) Policy, Canterbury District Health Board, Document No 238533, March 2017

Internal

• Patient Assessment form
• CSSD CJD Procedure for Management of Instruments and Equipment Procedures, CSSD work manual
• Standard Precautions Policy
• Blood and Body Fluid Exposure Policy
• Waste Management Policy
• Linen Management Policy
• Personal Protective Equipment Policy - staff
Appendix One – Classical Creutzfeldt - Jakob disease (CJD) Risk Assessment Tool

The following questions should be asked of a patient prior to undergoing planned elective surgery involving any of the higher-infectivity tissues so precautions can be put in place if needed.

a) Brain, pituitary or dura mater
b) Cranial and dorsal root ganglia
c) Spinal cord
d) Eye (Retina / Optic Nerve)
e) Olfactory Epithelium

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

Specialist Questions to Determine Risk Status

Q. 1. Do you think the patient may have CJD?  

Yes  No

Q. 2. Has the patient had two or more first or second degree relatives with CJD? (It is important to know about relatives with CJD, but having a single affected relative with sporadic CJD does not place the patient in a high or low risk category)

Yes  No

Q. 3. Does the patient have an unexplained progressive neurological illness of less than 12 months?

Yes  No

Q.4. Does the patient have a history of receiving human pituitary hormone for infertility of human growth hormone for short stature (prior to 1986)?

Yes  No

Q.5. Has the patient previously had surgery on the brain or spinal cord that included a dura mater graft (prior to 1990)?

Yes  No

Q.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?

Yes  No

Action: If the patient answers yes to any of the above questions, please contact the Pre Admission Service and Infection Prevention and Control Nurse to discuss admission CJD action plans.