### Purpose

- To describe best practice for the prevention of transmission of Prion diseases including Creutzfeldt-Jacob disease (CJD), sporadic CJD, Gerstmann-Straussler-Scheinker syndrome, and fatal familial insomnia. All these diseases are classified as Transmissible Spongiform Encephalopathies.

- To minimize the risk of transmission of Prion diseases from High-Risk Patients (both adults and children) to other patients and/or staff within Alberta Health Services Settings.

Note: Variant CJD (vCJD) is a rare disease in humans, with differing epidemiology from classic CJD and as such, infection prevention and control measures to prevent all potential exposures to vCJD are beyond the scope of this policy. To date there has been no documented case of surgical transmission of vCJD.

### Policy Statement

Alberta Health Services shall have procedures and processes in place to reduce the risk of exposure to patients and staff from the agents that cause Prion diseases as per the Alberta Health and Wellness *IPC Standards for Cleaning, Disinfection and Sterilization of Reusable Medical Devices for all Health Care Facilities and Settings* (2008) and the Public Health Agency of Canada *Infection Control Guidelines for Classic Creutzfeldt-Jacob Disease in Canada* (2002) and *Quick Reference Guide* (2007).
APPLICABILITY
Compliance with this policy is required by all AHS employees, members of the medical and midwifery staffs, students, volunteers, and other persons acting on behalf of AHS (including contracted services providers as necessary). This policy does not limit any legal rights to which you may otherwise be entitled.

POLICY ELEMENTS

1. The surgeon, or medical delegate, shall complete the Alberta Health Services Creutzfeldt-Jacob Disease (CJD) Risk Assessment Tool prior to performing elective or emergent:

1.1 surgery, invasive investigations, or procedures involving the:
   a) brain,
   b) spinal cord and spinal ganglia,
   c) dura mater,
   d) pituitary gland,
   e) retina or optic nerve,
   f) trigeminal ganglia;

1.2 all spine surgeries; and,

1.3 procedures to access the spinal canal or sample cerebrospinal fluid.

1.4 Exception: Lumbar/spinal taps using disposable instruments.

2. If on completion of the CJD risk assessment, implementation of CJD precautions is indicated, the surgeon/medical delegate is responsible for notifying relevant stakeholders including operating room nursing and Infection Prevention & Control to ensure that CJD precautions are implemented. Designated CJD precautions shall be implemented as outlined in the Alberta Health Services Procedure: Prion Disease (Creutzfeldt-Jacob Disease) Precautions for the Surgical Patient (Adult or Child).

3. If a High-Risk Patient is identified during surgery/investigations or procedures outlined in Section 1 of this policy, designated intra-operative and post-operative CJD precautions for clinical management, quarantine and decontamination of instruments and equipment shall be followed as outlined in the Alberta Health Services Procedure: Prion Disease (Creutzfeldt-Jacob Disease) Precautions for the Surgical Patient (Adult or Child).

4. In the event of emergent surgery/investigations or procedures identified in Section 1 above, the surgeon/medical delegate shall complete the CJD risk assessment prior to commencement of the procedure, except where this causes a delay that may create a risk
to patient safety. In the case of the latter, the surgeon/medical delegate shall complete the Alberta Health Services Creutzfeldt-Jacob Disease (CJD) Risk Assessment Tool immediately post procedure and the surgical instruments will be handled in accordance with CJD precautions defined in the Alberta Health Services Procedure: Prion Disease (Creutzfeldt-Jacob Disease) Precautions for the Surgical Patient (Adult or Child). This policy also applies to all surgery/investigations/procedures identified in Section 1 above that are to be performed at the bedside in an intensive care/emergency room or other Alberta Health Services Setting. The completed Alberta Health Services Creutzfeldt-Jacob Disease (CJD) Risk Assessment Tool shall be held in the patient’s Health Record.

5. Surgery/investigations/procedures (outlined in Section 1 of this policy) proposed for identified High-Risk Patients may proceed, subject to:

   5.1 all reasonable alternatives being exhausted; and,
   5.2 CJD precautions being implemented as outlined in the Alberta Health Services Procedure: Prion Disease (Creutzfeldt-Jacob Disease) Precautions for the Surgical Patient.

6. Single use disposable items shall be used for identified High-Risk Patients whenever possible during surgery/investigations/procedures outlined in Section 1 of this policy.

7. All sites will develop, and adhere to, approved procedures for the quarantine and decontamination of instruments and equipment used for High-Risk Patients.

8. Completed Alberta Health Services Creutzfeldt-Jacob Disease (CJD) Risk Assessment Tools shall be included in the surgical booking package, and then held in the patient’s Health Record. If on receipt of the surgical booking package, the completed Alberta Health Services Creutzfeldt-Jacob Disease (CJD) Risk Assessment Tool is not included or is incomplete, the surgical booking package will be returned to the surgeon’s office.

9. All staff involved in surgery/investigations/procedures undertaken on High-Risk Patients shall be educated and trained regarding CJD precautions, the occupational risks involved, and the procedures and processes to be followed.

DEFINITIONS

Alberta Health Services setting means any environment where treatment, procedures and other health care are delivered by, on behalf of or in conjunction with Alberta Health Services.

Creutzfeldt-Jacob Disease/Transmissible Spongiform Encephalopathies (CJD/TSE) means a rare neuro-degenerative disease caused by an abnormal Prion protein that causes surrounding proteins to change their shape resulting in pre-senile dementia, myoclonus and progressive motor dysfunction.

Health record means the Alberta Health Services legal record of the patient's diagnostic, treatment and care information.
High-risk patient(s) means an individual with a confirmed, probable or familial CJD, or with clinically suspected CJD, or is an asymptomatic carrier, but meets one or more of the following criteria:

- a) the person has been confirmed by genetic testing to carry a genetic mutation causative of familial CJD, GSS, or FFI;
- b) the person has at least one first-degree relative who has been confirmed by genetic testing to carry such a mutation, with or without pathologic confirmation of TSE; and/or
- c) the person has two or more first-degree relatives who have been diagnosed with either confirmed or probable TSE, with or without confirmation by genetic testing.

Prion means an abnormal form of a normal cellular protein that is the causative agent of CJD and other associated diseases. These non-cellular structures are hardy and resist all routine inactivation procedures commonly used by health care facilities to reprocess instrumentation between patients.

REFERENCES

- Alberta Health Services Procedure: Prion Disease (Creutzfeldt-Jacob Disease) Precautions for the Surgical Patient
- Alberta Health and Wellness IPC Standards for Cleaning, Disinfection and Sterilization of Reusable Medical Devices for all Health Care Facilities and Settings (2008)
- Public Health Agency of Canada Infection Control Guidelines for Classic Creutzfeldt-Jacob Disease in Canada (2002)
- Alberta Health Services Creutzfeldt-Jacob Disease (CJD) Risk Assessment Tool

REVISIONS

N/A