QUINTE HEALTHCARE CORPORATION

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Creutzfeldt- Jakob Disease (CJD)

Title: Creutzfeldt-Jakob Disease (CJD)		**Policy No:**	**5-20**
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Manual:	**Infection Prevention and Control**	**Last Review Date:**	December 2019
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Section 5:	**Significant Organisms and Ectoparasites**	**Policy Lead:**	Infection Control Practitioner
Approved By:	**Medical Advisory Committee October 2012 Infection Control Program Advisory Committee**		

1. POLICY

Infection prevention and control will be informed of all cases of confirmed or suspect Creutzfeldt-Jakob Disease

2. PURPOSE

1. To provide guidelines for the management of patients with proven or suspected prion diseases.
2. To provide guidelines for the collection, handling, decontamination and/or disposal of contaminated equipment, tissues or body fluids from patients with proven or suspected prion diseases.

3. BACKGROUND:

Creutzfeldt-Jakob belongs to a family of human and animal diseases known as transmissible spongiform encephalopathies. The disease is a rare, degenerative, fatal brain disorder affecting about one person in one million people per year worldwide. As the illness progresses, mental deterioration becomes pronounced and involuntary movements, blindness, weakness of extremities, and coma may occur. There are three main categories of CJD. Sporadic is the most common with at least 85 percent of cases falling into this category. Hereditary consists of approximately 5 to 10% of cases. The final category of acquired CJD makes up less than 1% of cases. See Appendix A.

4. DIAGNOSIS

CJD is not readily diagnosed. A spinal tap, electroencephalogram (EEG), computerized tomography (CT), and magnetic resonance imaging (MTI), brain scans can help reveal characteristic features of CJD and rule out other treatable forms of dementia such as encephalitis or chronic meningitis.

4.1 Confirmed Case

Neuropathologically confirmed, with confirmation of protease-resistant prion protein (immunohistochemistry, PET blot, or Western Blot).

4.2 Probable Case

- Rapidly progressive dementia **AND**
- At least two additional neurological manifestations (See Section 5.0 – Clinical Evidence) **AND**
- One of three clinical tests:
  - Typical electroencephalography (EEG): generalized bilateral or unilateral triphasic periodic complexes at approximately one per second, lasting continuously for at least 10 seconds.
  - MRI with caudate nucleus and/or (anterior) putamen attenuation (preferred sequence DWI or FLAIR).
  - Positive assay for 14-3-3 protein in cerebrospinal fluid (CSF) AND total disease duration less than 24 months.

4.3 Suspect Case

- Rapidly progressive dementia **AND**
- At least two additional neurological manifestations (See Section 5.0 – Clinical Evidence) **AND**
- Duration of illness less than 2 years in the absence of a conclusive MRI and 14- 3-3 protein assay.

MODE OF TRANSMISSION(see Appendix A)

CJD cannot be transmitted through air or casual contact.

5. PATIENT RISK ASSESSMENT FOR CJD

5.1 High-Risk Patients for iatrogenically transmitting CJD are those diagnosed, prospectively or retrospectively, with:

- CJD – confirmed, probable or possible CJD, familial CJD, Gerstmann-Straussler – Scheinker diseases (GSS), or fatal familial insomnia (FFI) depending on pathological, laboratory and clinical evidence.
- Suspected CJD – undiagnosed, rapidly progressive dementia and CJD not ruled out.
- Asymptomatic carrier of genetic transmissible spongiform encephalopathy (TSE) – a person who displays no symptoms or signs of TSE, but meets one or more of the following criteria:

1. The person has been confirmed by genetic testing to carry a genetic mutation causative of familial CJD, GSS, or FFI;

2. 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;

3. 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.

*See appendix C for appropriate procedures for managing the instruments that have been in contact with high risk patients, depending on the potential infectiousness of the tissue contacted.

5.2 At-Risk Patients of iatrogenic (caused by medical exam or treatment) **CJD**

- Recipients of human tissue derived pituitary hormone treatment (either growth hormone or gonadotropin). All human growth hormone has been made synthetically since 1985. There is no longer risk to patients who receive these products after 1985.
- Recipients of dura mater graft (until 1992 for Lyodura grafts, until 1997 for Tutoplast Dura grafts)

- Recipients of a corneal graft originating in a jurisdiction that does not require graft donors to be screened for neurological disease.
- Patients who have been exposed, via contact with instruments, to high infectivity tissue of a confirmed CJD patient.
- The risk of transmission via instruments used on at-risk, asymptomatic patients is negligible low, and therefore, such instruments may be routinely decontaminated and then reused.

Source: Public Health Agency of Canada 2007

5.3 TISSUE RISK CATEGORIES

Level of Infectivity	Tissues, Secretions, and Excretions
High Infectivity	brain (including dura mater), cerebrospinal fluid (CSF), pituitary gland, Posterior eye (optic nerve and retina), Spinal cord and spinal ganglia, & trigeminal ganglia
Low Infectivity	corneal, kidney, liver, lung, lymph nodes, spleen and placenta
No Detectable Infectivity **	adipose tissue, adrenal gland, appendix, blood, bone marrow, breast milk, dental pulp, epididymis, esophagus, feces, gingival tissue, ileum, jejunum, large intestine, nasal mucosa, nasal mucous, ovary, pancreas, pericardium, peripheral nerve, placenta fluid, prostate, saliva, semen, seminal vesicle, skeletal muscle, skin, sweat, tears, testis, thymus, thyroid gland, tongue, tonsil and trachea, urine and uterus

- CJD testing is an ever evolving process with some organizations changing the label of infectivity for certain tissues. However, the Public Health Agency of Canada continues to label tissues' infectivity per the table above.

Source: Public Health Agency of Canada 2007

6 NOTIFICATION

6.1 Attending Physician or Surgeon will provide advance notification if a High Risk or an At Risk patient is to be admitted.

Advance Notification Should Include:

a) Infection Control and Chair of Infection Control Program
b) Administration
c) Operating room, laboratory, histology, microbiology and Central Processing prior to any surgery or procedure that involves a possibly infectious tissue/fluid:
   i. Patient care areas in order to implement CJD precautions when contact with possibly infectious tissues of a High Risk patient or high infectivity tissue and CSF of an At Risk patient are possible.
   ii. Pathology Services.

6.2 The facility is responsible for notifying funeral home or other organizations that will be handling body or tissues/fluids (crematorium, laboratories, and memorial societies).

6.3 Patients who are at high risk for transmission of CJD or their responsible persons should be advised to notify their doctors, dentists and other health care providers of their status so that precautions can be taken.

7 GENERAL PATIENT CARE

7.1 Use Routine Practices for all procedures except for invasive procedures involving high or low infectivity tissue of a high risk patient or high infectivity tissues and CSF of an at risk patient.
7.2 A single room is *not* required for Infection Prevention and Control purposes.
7.3 No special precautions are required for feeding utensils, feeding tubes, suction tubes, razors or items used in the care of skin or bed sores.
7.4 Routine practices apply to most situations involving linen. Linen including PPE that are exposed to high or low infectivity tissues and CSF of an at risk patient should be sealed in a leak proof container, labeled "biohazardous" and incinerated.

8 MEDICAL PROCEDURES

8.1 High risk or at risk CJD patients may need to undergo diagnostic procedures unrelated to CJD. These procedures may be conducted without any special precautions, as long as procedures only involve tissues containing no detected infectivity.
8.2 For medical and surgical procedures that involve contact high and low infectivity tissues see **Appendix C** for action to be taken.

8.3 CJD Precautions for Surgical/ Invasive Procedures

a) The number of health care workers involved in the surgery should be limited to the minimum required.
b) Health care workers should wear single-use personal protective equipment, such as gloves, liquid repellent gowns, fluid-resistant aprons, head and foot coverings, face shields, and masks.
c) Protective clothing should be sealed in a red plastic bag and sent for incineration at the end of the procedure.
d) Single use items and instruments should be used whenever possible.
e) Limit supplies and equipment in the Operating Suite or procedure room to only what is necessary for the procedure.
f) Temporary covers, shields, or guards made of disposable, liquid-resistant materials that can be removed and incinerated should be used wherever surfaces, such as surgical tables, counter tops, instruments (e.g. drills) or equipment have a possibility of being exposed to high or low infectivity tissues from a High Risk patient or high infectivity tissues and CSF from an At Risk patient. The covers, shields or guards must then be placed in a red plastic bag and sent for incineration.
g) Maintain a one-way flow for instruments used in procedure. Pass sharps in trays, not from hand to hand.
h) Separate instruments exposed to high or low infectivity tissues from instruments exposed to no detectable infectivity tissues.
i) All disposable equipment, supplies, body tissues, and fluids must be sent for incineration.
j) Stereotactic CNS procedures are not recommended for High Risk and at risk patients.
k) High or low infectivity specimens of a High Risk patient, or high infectivity specimens of an At Risk patient that are sent to the laboratory should be sealed in a leak proof, puncture- resistant container, bagged in a biohazard pouch, and clearly labeled as "high risk for CJD".
l) Schedule surgery at the end of the day if possible, to facilitate cleaning of the Operating Suite.
m) Clean and decontaminate all hard surfaces that are exposed to high or low infectivity tissues from High Risk patient or high infectivity tissues from an At Risk patient. See 7.2

9 **Process for CJD decontamination:**

9.1 Routine sterilization is inadequate and does not destroy the CJD agent. Flash and routine autoclaving are ineffective to destroy the CJD agent and must never be considered as a processing method for reusable instruments exposed to the CJD agent.

9.2 Any instruments that are used on a patient who have suspected CJD must be placed in a red biohazard bag/red biohazard container and quarantined in the patient room until the diagnosis of CJD is confirmed or ruled out.

9.3 If CJD is ruled out, instruments may then be cleaned per protocol for said instrument.

9.4 If CJD is confirmed, all instruments quarantined must then be sent for incineration.

9.5 Hard Surface Decontamination

Remove visible soil with a disposable cloth. Place cloth in red plastic bag for incineration. Then flood with undiluted sodium hypochlorite (bleach); let stand for 1 hour; then mop up and rinse with water. Staff member should wear chemical specific respirator, eye protection, gloves, and protective clothing as per material safety data sheet.

9.6 Quarantine

Quarantine instruments under suitable storage conditions (dry, protected, secure environment, clearly identified as quarantined). Do not reuse, unless a diagnosis is made that eliminates the possibility that the patient on whom the instruments were used had CJD. A confirmed diagnosis other than CJD, either clinical, pathological, or a post-mortem examination excluding CJD is required to take instruments out of quarantine. A brain biopsy that is negative for CJD, in the absence of a confirmed alternate diagnosis, does not suffice to take instruments out of quarantine.

10 **Pregnancy/Childbirth**

a) CJD is not known to be transmitted from mother to child during pregnancy and childbirth.
b) In general, childbirth for women at high risk or at risk for CJD should be managed using Routine Practices. Because the placenta is considered low infectivity tissue, CJD precautions should be taken for high-risk women to reduce the risk of exposure to placenta and other products of conception.
c) Health Care workers should wear appropriate single-use personal protective equipment such as gloves, liquid repellant gowns, fluid-resistant aprons, head and foot coverings, face shields and masks. Protective eyewear (visors or goggles) or other protective devices must be worn when it is necessary to protect the eyes and face from splashes and particles.
d) The placenta and products of conception of women at high risk for CJD must be sealed in red plastic bag and incinerated. The placenta and products of conception must not be used for any other purpose.
e) If the placenta or products of conception are to be sent to the laboratory for investigation, they should be sealed in a leak proof, puncture resistant container and clearly labeled as high risk for CJD.
f) Linen, solid waste, personal protective equipment exposed to the placenta or products of conception of women at high risk for CJD must be sealed in a red plastic bag and sent for incineration.
g) Environmental surfaces exposed to the placenta or products of conception of women at high risk for CJD should undergo CJD decontamination.

11 **Specimen Collecting and Handling**

a) All health care workers should wear single use protective clothing (gloves, gowns and masks) when collecting and handling high infectivity tissues or low infectivity tissues from a high risk patient or high infectivity tissues and CSF from an at risk patient. Protective eyewear (visors or goggles) must be worn when necessary, to protect the eyes and the face from splashes and particles. Exposed protective equipment must be placed in a red plastic bag and sent for incineration.
b) High or low infectivity specimens from a high risk patient or high infectivity specimens and CSF from an at risk patient should be sent to laboratory in a sealed leak proof puncture resistant container which is clearly labeled as at high risk for CJD.

For high or low infectivity tissue from a high risk patient or high infectivity tissue or CSF from an at risk patient:

i. A sealed container and closed centrifuge bucket must be used when mixing or centrifuging is to be done.
ii. The use of a biological safety cabinet is recommended for opening the container. Specimens should be sealed in a leak proof, puncture resistant container and placed in a red plastic bag and sent for incineration.
iii. All single use laboratory instruments or equipment should be sealed in a red plastic bag and sent for incineration.
iv. All non-disposable laboratory instruments or equipment and laboratory work surfaces may be cleaned and disinfected as per Appendix B.
v. Waste should be sealed in a red plastic bag and sent for incineration.
vi. No special measures are required for cleaning or decontaminating auto analyzers.
vii. CJD precautions are not necessary for collecting, handling or processing of blood, urine or fecal specimens.

12 Post Mortem Exam

If any patient with suspected CJD passes away prior ruling out said diagnosis a post mortem exam is indicated to determine if the patient truly had CJD. This will allow for proper disposal of any contaminated items associated with that patient's care.

APPENDICES AND REFERENCES

Appendices: Appendix A − Human Prion Diseases
Appendix B − CJD Risk Assessment Tool/Pre-op Check List Appendix C − High Risk CJD Patients Managed Prospectively

References:

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Related Policies:

2-05 Communicable and Reportable Diseases
3-45 Personal Protective Equipment − General Standards and Requirements
3-20 Hand Hygiene
3-50 Routine Practices and Additional Precautions
3-55 Sharps Handling and Disposal