IS0900: Creutzfeldt - Jakob Disease (CJD)	**EFFECTIVE DATE:** September 2006 **REVISED DATE:** November 2010, December 2012, May, 2019 **REVIEWED DATE: May, 2019**

1.0 PURPOSE

To inform healthcare providers of their respective roles and responsibilities when involved in care and management of *suspected, probable* and *confirmed* cases of Creutzfeldt - Jakob disease (CJD) and other prion diseases to prevent iatrogenic transmission to patients and Interior Health (IH) staff including:

- **Notifying and reporting** requirements, and
- **Managing care delivery**,
- **Assessing risk** for performing lumbar punctures, nasal endoscopic procedures, and surgery on tissue/fluid of the eye and central nervous system,
- **Handling of the body.**

Note: In situations where an autopsy may be required or requested on a suspected or probable case of CJD, autopsies are ***not*** performed within an IH facility.

Background:

Creutzfeldt - Jakob disease (CJD) is a rare, progressive, untreatable condition of the central nervous system caused by abnormal prion. Iatrogenic transmission occurs by direct inoculation via corneal transplant, contaminated neurological electrodes, dura matter grafts, injections of hormone or human pituitary gland origin.

Incubation ranges from 15 months to approximately 30 years.

Tentative diagnosis of CJD is based on:

- Clinical presentation; and
- Diagnostic imaging (MRI/CT/EEG); and
- Laboratory testing (CSF for immunoassay of 14-3-3 protein).

Suspect CJD diagnosis is based on the following criteria:

1) Progressive dementia
2) At least two of the following:
   a. Myoclonus
   b. Visual or cerebellar disturbance
   c. Pyramidal or extra-pyramidal dysfunction
   d. Akinetic mutism

But, none of the EEG (Electroencephalogram), CSF (Cerebrospinal fluid) or MRI (Magnetic Resonance Imaging) results are indicative of CJD (BCCDC, 2017).

Probable CJD diagnosis is based on the following criteria:

1) Progressive dementia

2) At least two of the following:
   a. Myoclonus
   b. Visual or cerebellar disturbance
   c. Pyramidal or extra-pyramidal dysfunction
   d. Akinetic mutism
3) At least one of the following:
   a. Periodic sharp wave complexes on EEG
   b. Positive 14-3-3 assay on CSF
   c. MRI showing high abnormal signal in the caudate nucleus and putamen on fluid attenuated inversion recovery (FLAIR) or Diffusion Weighted Imaging (DWI).

Confirmed CJD diagnosis is based on positive brain tissue upon autopsy with neuropathological examination, immunocytochemical methods, or Western blot detection of disease-specific PrP. Note: This result will only be available after the patient is deceased.

2.0 DEFINITIONS and ABBREVIATIONS

At Risk Patients: Patients at risk of accidentally *acquiring* (iatrogenic) CJD including:

- recipients of human tissue derived pituitary hormone treatment (either growth hormone or gonadotrophin)
- recipients of a dura mater graft
- recipients of a corneal graft where donor was not screened for neurological disease
- patients who have been exposed to instruments having come into contact with high-infectivity tissue of a confirmed CJD patient

BCCDC: British Columbia Centre for Disease Control

CJD: Creutzfeldt - Jakob disease

CSF: Cerebrospinal fluid

High Risk Patients: Patients at high risk of *transmitting* CJD iatrogenically are those diagnosed, prospectively or retrospectively, with:

- CJD - confirmed, probable, or possible CJD, familial CJD, Gerstmann-Straussler-Scheinker disease (GSS) or fatal familial insomnia (FFI)
- 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 criterion:
  - The person has been confirmed by genetic testing to carry a genetic mutation causative of familial CJD, GSS, or FFI;
  - The person has at least one first-degree relative who has been confirmed by genetic testing to carry such a mutation, with or without pathological confirmation of

TSE;

- 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 Proteins (PrP): A protein found on the surface of cells that attack the brain, destroy cells and create gaps in tissue or form sponge-like patches. These proteins are resistant to most chemical and physical decontamination methods.

3.0 GUIDING PRINCIPLES

3.1 CJD precautions will be used when exposure to high infectivity tissues or CSF from a high risk or an at risk client is anticipated (see Appendix A).

3.2 Incineration must be followed without exception when instruments are exposed to high or low infectivity tissues of a high risk patient, or high infectivity tissue and CSF of an at-risk patient.

3.3 All cases of CJD (suspected, probable and confirmed) must be reported to IH Communicable Disease Unit for provincial reporting to BCCDC (see section 4.5).

4.0 PROCEDURE

4.1 Standard Process

For information on the roles and responsibilities for the management and reporting of CJD cases in Interior Health to prevent iatrogenic transmission, see *Creutsfeldt-Jakob disease (CJD) Standard Processes* in the CJD Toolkit.

4.2 Patient Care

4.21 For patients with suspect or probable CJD, Routine Practices should be followed when providing patient care, for collection of blood, urine, stool tests, and for noninvasive radiology examinations across all care settings including acute, long-term care, community,and Hospice/Palliative care.

4.2.2 For patient transport/transfer within and across facilities, apply Routine Practices.

4.2.3 For patient education see: CJD HealthLink BC File.

4.3 Notification

4.3.1 When a patient is suspected of having CJD, notification must be given to the ICP and/or Medical Microbiologist who will notify:

- Medical Health Officer (MHO) and Communicable Disease (CD) Unit of known/suspect cases of CJD preoperatively or intra-operatively or confirmed CJD post-operatively. See *SSI0125 Appendix D CJD Notification and Communication Algorithm* .
  - Medical Health Officer On-Call (1-866-457-5648) after hours
  - CD Unit (1-877-778-7736) Monday to Friday 0830-1630

4.3.2 When a patient is confirmed to have CJD the Most Responsible Physician (MRP) is required to report cases to the Medical Health Officer**.** See *Creutzfeldt-Jakob Disease (CJD): Reporting, Surveillance and Public Health Follow-Up Workflow*

4.3.3 For further information on the roles and responsibilities for the management and reporting of CJD cases in Interior Health to prevent iatrogenic transmission, see *Creutzfeldt-Jakob Disease (CDJ) Standard Processes.*

Note: Process Standards will be linked to this guideline as they become available electronically. Please contact the IH Professional Practice Office in the interim.

4.4 Reporting-Communicable Disease Unit/MHO

4.4.1 The Communicable Disease Unit will apply a CJD Special Indicator in Meditech which includes "discuss invasive surgical and LP procedures with Microbiologist".

4.4.2 Probable and confirmed cases of CJD are reportable in British Columbia and must be reported to the Medical Health Officer (MHO) or CD Unit. This is especially important when probable or confirmed cases of CJD have had invasive procedures or have donated any tissues or organs for transplantation.

Refer to *Creutzfeldt-Jakob Disease (CJD): Reporting, Surveillance and Public Health Follow-Up Workflow.*

4.4.3 The MHO will provide the physician with information to review with the patient's family related to blood, tissue and organ donation. Refer to Physician Letter.

4.5 Risk Assessment for Medical Procedures

4.5.1 **Performing Lumbar Puncture (LP) Procedure on Suspect /Probable CJD patient**

- A risk assessment (Appendix C) must be completed prior to any LPs being performed.
- LP will **NOT** be performed on patients with suspected or probable CJD without prior consultation with a neurologist.
- MRP screens for suspected CJD prior to LP using the *Non-Surgical Patient - Creutzfeldt Jacob Diesease (CJD) Risk Assessment Tool* (Appendix C) for all patients who fulfill the following criteria:
  - Between 45 and 75 years of age

    AND

  - Present with neurological symptoms with NO alternative diagnosis.

If any answer to screening is "YES"; MRP consults neurologist

- Neurologist to notify Medical Microbiologist and Infection Control Practitioner **prior** to the procedure.
- Notify unit manager prior to LP.
- Perform LP in a single bed room or a procedure room.
- Procedures are **ONLY TO BE DONE** at the following sites: Penticton Regional Hospital, Kelowna General Hospital, Vernon Jubilee Hospital, Royal Inland Hospital, East Kootenay Hospital, Kootenay Boundary Hospital.
- Refer to (Appendix B) Lumbar Puncture for Suspected CJD Algorithm

4.5.2 **Endoscopic Procedure on Suspect or Probable CJD patient**

- It is **NOT** recommended to do an endoscopic procedure via the **nasal route**.
- If an endoscopic procedure via the nasal route is not avoidable, MRP to consult with Medical Microbiologist prior to the procedure.

- Other types of endoscopic procedures are safe and require routine practice.

4.5.3 Perioperative Management of CJD

- Risk of iatrogenic transmission is higher in operating rooms. For information on perioperative procedures, please see SSI0125 *Perioperative Management of CJD.*
- Procedures are **ONLY TO BE DONE** at the following sites: Kelowna General Hospital, Royal Inland Hospital, East Kootenay Hospital, and Kootenay Boundary Hospital.

4.6 Cleaning of Spills, Decontamination Processes, and Waste Management

Patient Care Areas-Housekeeping is responsible for biological spill clean-up in patient care areas.

Operating Room –The most responsible person for cleaning can vary between sites. Refer to Surgical Services SSI0150 and SSI0125 for more information.

Laboratory –The lab is responsible for cleaning of spills in their area. Refer to laboratory process SA 0139.04

Refer to Housekeeping standard process *Biological Spill Clean Up Related To Suspected Creutzfeldt-Jackob Disease (CJD) Exposure* and Faucet-Mounted Eyewash AXION: product information and user instruction video.

4.6.1 Waste Management

- All items in contact with CSF must be contained and placed in the red biohazardous waste pail for incineration (this includes any linen if contaminated).
- All sharps and other disposables that are contaminated with CJD high or low risk tissue/fluid should be contained as CJD biomedical waste, labelled "**Biohazardous – For Incineration. DO NOT OPEN".**
    - Notify Housekeeping service.
    - Housekeeping service arranges the incineration.
- Other waste to be disposed according to routine practice.

4.7 Post-Exposure Procedures

Occupational exposure to CJD has been very rarely reported. See section 4.8, Occupational Safety, SSI0125 *Perioperative Management of CJD* for accidental exposure to:

- High risk infectivity tissues/fluid to unbroken skin.
- Needle-stick injury or laceration involving high infectivity tissues /fluid.
- Splashing into the eyes or mouth.

4.8 Deceased Body Management

4.8.1 Transport

The transportation of the body from a facility, community setting or hospice to a morgue or a funeral home should be carried out using Routine Practices.

- Place body in a sealable, impermeable plastic pouch.
- Absorbent pad should be added to the pouch to absorb any fluids.
- Complete two patient identifier tags with all required information and attach one tag to deceased's great toe with elastic.
- Second identifier tag is attached to zipper on body bag and includes the Biohazard sticker "Attention: Funeral Home – See Medical Certificate of Death prior to handling the body". Refer to Deceased Patient, Care of: Acute Care.

4.8.2 Pathology

No autopsies will be performed in IH on patients known to have, or suspected of having, CJD. Follow SA0139 *Creutzfeldt-Jakob Disease (CJD) Sample Handling Process*.

4.9 Laboratory

If a specimen (CSF or other high risk sample) requires CJD precautions, the surgeon, neurosurgeon, neurologist or MRP notifies the Medical Microbiologist of the suspected CJD case.

- For CJD testing order 14-3-3:
  - Minimum of 2ml CSF is required for testing.
  - Each tube must be labelled with required patient information.
  - Samples must be marked with "Suspect CJD".
  - CSF samples are placed in a biohazard bag and hand delivered to lab staff.
- Sample must be immediately delivered to the lab and handed to a staff member
  - Do not leave samples unattended.
  - Samples must not to be sent in the pneumatic tube system.

See the following links for further information regarding lab processes for CSF collection and sample management:

- Cerebrospinal Fluid – CSF Collection
- Ordering tests for Prion (CJD/14-3-3)

4.10 Retrospective Management of High Risk CJD patients

Mitigation against retrospective management of instruments that contact high risk tissue or fluid from a high or at risk CJD patient has been addressed through the use of disposable instrument sets for neurosurgical procedures and lumbar punctures, appropriate triage and screening by the Neurosurgery and Neurology teams; and protocols for management of waste.

- In the event of inadvertent exposure of patients or staff to contaminated equipment, a Risk Management Team should be called together to review the case and recommend necessary actions. Refer to *Creutzfeldt-Jakob Disease (CDJ)* Standard Processes.

APPENDIX:

Appendix A - Infectivity of CJD in Various Tissues Table

Appendix B - Lumbar Puncture for Suspected CJD Algorithm

Appendix C – CJD Risk Assessment Tool

5.0 REFERENCES

BC Centre for Disease Control. (2017). Cr*eutzfeldt-Jakob Disease: A Resource for Health Professionals*. Retrieved from http://www.bccdc.ca/resource-gallery/Documents/Guidelines%20and%20Forms/Guidelines%20and%20Manuals/Epid/Other/CJDwebsiteinfofinalv7.pdf

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Appendix A:
Infectivity of CJD in Various Tissues

High-infectivity	Low-infectivity
Brain Cerebrospinal fluid (CSF)[2] Dura mater Pituitary gland Posterior eye (optic nerve and retina) Spinal cord and spinal ganglia Trigeminal ganglia	Cornea[3] Kidney Liver Lung Lymph nodes Placenta Spleen
No detected infectivity	
Adipose tissue Adrenal gland Appendix[4] Blood (including cord blood)[4] Blood vessels[5] Bone marrow Breast milk (including colostrum)[4] Dental pulp Epididymis[4] Esophagus[4] Feces Gingival tissue[4] Heart Ileum[4] Jejunum[4] Large intestine[4] Nasal mucosa[5] Nasal mucous Ovary[4]	Pancreas[4] Pericardium[4] Peripheral nerves[5] Placental fluids[4] Prostate Saliva Semen Seminal vesicle[4] Skeletal muscle[5] Skin Sweat Tears Testis Thymus[4] Thyroid gland Tongue[4] Tonsil4 Trachea[4] Urine Uterus (non-gravid)[4]

In this update of the « 2002 CJD Infection Control Guideline », infectivity for cornea, optic nerve, and retina has been differentiated. Trigeminal and spinal ganglia have been classified as high-infectivity tissues. Dental pulp has been moved from low-infectivity to no detected infectivity based on recent experiments that did not detect the abnormal prion protein in dental pulp of patients with human TSEs. Serous exudates have been removed.

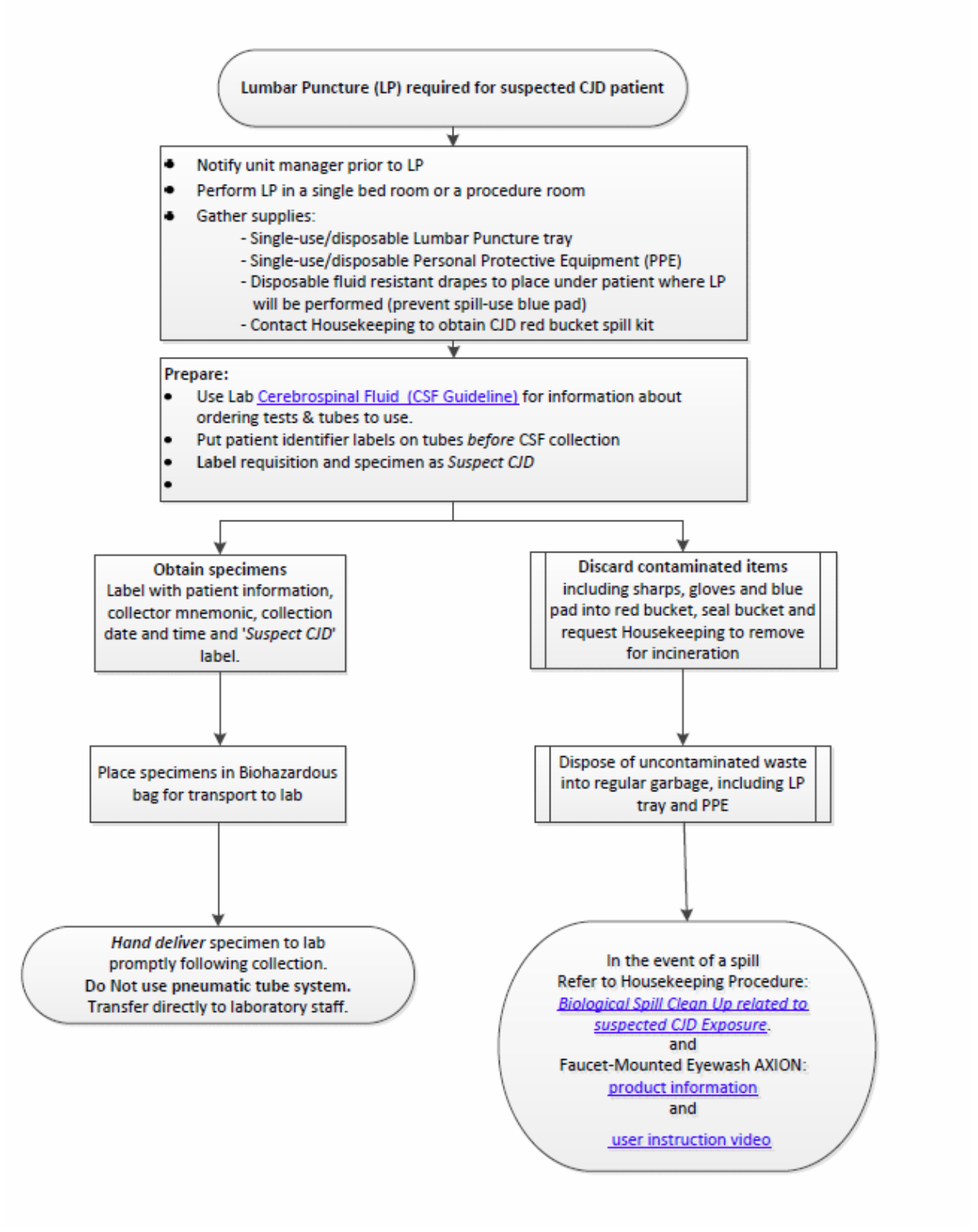
2 While CSF is a low-infectivity tissue, contact with CSF necessarily implies contact with high infectivity tissue and should be managed as a high infectivity tissue/fluid for infection prevention and control purposes.

3 Other anterior chamber tissues (lens, aqueous humor, iris, conjunctiva) have been tested with a negative result in human TSEs, and there is no epidemiological evidence that they have been associated with iatrogenic transmission.

4 A number of tissues have been examined for infectivity and/or the presence of abnormal prion protein with negative results in classic CJD.

5 Recent research findings in human TSEs have demonstrated the presence of the abnormal prion protein (PrPTSE) in several peripheral tissues (blood vessels, nasal mucosa, peripheral nerves, skeletal muscle). So far, no infectivity has been demonstrated with these tissues in classic CJD, and the precise relationship between the presence of PrPTSE and infectivity is not certain (e.g., detection of small amounts of PrPTSE in a tissue does not necessarily mean that it would transmit disease in all circumstances). For the purpose of infection control, they will be regarded as non-infectious.

Appendix B:
Lumbar Puncture for Suspect CJD



Appendix C:
Non-Surgical Patient - Creutzfeldt-Jacob (CJD) Risk Assessment Tool

For all lumbar puncture and endoscopy procedures via nasal route if patients are 1) Between 45 and 75 years of age **AND** 2) Present with neurological symptoms with NO alternative diagnosis.

a)	Does the patient have known or suspected CJD? (established diagnosis*)	Yes □	No □
b)	Is there a family history of CJD?	Yes □	No □
c)	Is there a family history of any other inheritable spongiform encephalopathy? **	Yes □	No □
d)	Has the patient ever received any human pituitary-growth hormone therapy?	Yes □	No □
e)	Does the patient have a history of receiving any human dural engraftment?	Yes □	No □
f)	Has the patient been investigated with depth electrode for epilepsy?	Yes □	No □
g)	Does the patient have a history of receiving a human corneal transplant?	Yes □	No □
h)	Does the patient have a rapidly progressive dementia not yet diagnosed?	Yes □	No □
i)	Has the patient received notification from their family physician that they have received blood products from a CJD donor?	Yes □	No □
j)	Has the patient been in surgical contact with a possible CJD donor?	Yes □	No □

If any answer for the above questions is “YES”, MRP consults Neurologist.

***** When the quartet of dementia, myoclonus, periodic EEG activity, and rapid progression is present, the diagnosis of CJD is almost certain. Diseases that must be ruled out are: Alzheimer’s, Parkinson’s, Familial Myoclonic Dementia or Gerstmann-Straussler-Scheinker (http://www.ninds.nih.gov/disorders/gss/gss.htm) and Fatal Familial Insomnia.