Ministry of Health

Ontario Public Health Standards:
Requirements for Programs, Services and Accountability

Infectious Diseases Protocol

Appendix 1: Case Definitions and Disease-Specific Information

Disease: Creutzfeldt-Jakob Disease, all types

Effective: April 2026

Ontario

Creutzfeldt-Jakob Disease (CJD), all types

☒ Communicable

☐ Virulent

Health Protection and Promotion Act **(HPPA)**[1]
Ontario Regulation (O. Reg.) 135/18 **(Designation of Diseases)**[2]

Provincial Reporting Requirements

☒ Confirmed case

☒ Probable case

☒ Suspect case

As per Requirement #3 of the "Reporting of Infectious Diseases" section of the *Infectious Diseases Protocol, 2023* (or as current)[3], the minimum data elements to be reported for each case are specified in the following:

- *Reports*, R.R.O.1990, Reg. 569 made under the HPPA;[4]
- The iPHIS User Guides published by Public Health Ontario (PHO); and
- Bulletins and directives issued by PHO.

Type of Surveillance

Case-by-case

Case Definition

Sporadic CJD

Confirmed Case

Progressive neurological syndrome **AND**

Neuropathologically **or** immunohistochemically **or** biochemically confirmed.

Probable Case

Progressive neurological syndrome and positive end-point quaking-induced conversion (EP-QuIC) in cerebrospinal fluid (CSF) or other tissues.

OR

Rapidly progressive cognitive impairment.

AND

Two of the following: myoclonus; visual or cerebellar problems; pyramidal or extrapyramidal features; akinetic mutism.

AND

Typical electroencephalography* (EEG) **or** typical magnetic resonance imaging** (MRI) brain scan **or** positive EP-QuIC.

Interpretation of EEG and/or MRI results

*Typical EEG results:

- Generalized triphasic periodic complexes at approximately one per second.

**Typical MRI brain scan results:

- High signal in caudate/putamen or at least two cortical regions (temporal, parietal, occipital) either on diffusion-weighted imaging (DWI) or fluid-attenuated inversion recovery (FLAIR).
- Equivalent terms used to describe MRI findings for CJD include:
    - High signal = hyperintensity/hyperintensities or restricted diffusion on DWI
    - High signal in at least two cortical regions = cortical ribboning

Additional note:

If the criteria above have not been noted but the clinical impression in the EEG and/or MRI report indicates that CJD is on the differential diagnosis, the PHU may take the following steps:

- Connect with the CJDSS in PHAC to discuss the findings; or
- Follow up with the clinician (e.g., neurologist and/or radiologist) for further confirmation; and
- Document the discussion: the decisions made (e.g., criteria met or not met); the date of discussion(s); and the name of the individual(s) consulted with.

Suspect Case

Rapidly progressive cognitive impairment.

AND

Two of the following: myoclonus; visual or cerebellar problems; pyramidal or extra-pyramidal features; akinetic mutism.

AND

Duration < 2 years.

Iatrogenic CJD

Confirmed Case

Confirmed CJD with a recognized iatrogenic risk factor (see below).

Probable Case

Probable CJD with recognized iatrogenic risk factor (see below).

OR

Progressive predominant cerebellar syndrome in human pituitary hormone recipients.

Relevant exposure risks for classification as iatrogenic CJD:

- Treatment with human pituitary growth hormone, human pituitary gonadotropin, or human dura mater graft.
- Corneal graft in which the corneal donor has been classified as confirmed or probable human prion disease.
- Exposure to neurosurgical instruments used in a case of confirmed or probable human prion disease.

Notes:

- The above list is provisional as unknown mechanisms of exposure risk to human prion disease may occur.
- The relevance of any exposure should consider the timing of the exposure in relation to disease onset.

Genetic CJD

Confirmed Case

Confirmed CJD and confirmed or probable CJD in a first-degree relative.

OR

Confirmed CJD with a pathogenic prion protein gene (*PRNP)* mutation.

Probable Case

Progressive neuropsychiatric disorder and confirmed or probable CJD in a first-degree relative.

OR

Progressive neuropsychiatric disorder and pathogenic *PRNP* mutation.

Variant CJD (vCJD)

Confirmed Case

Progressive neuropsychiatric disorder **and** neuropathological confirmation of vCJD (i.e., spongiform change and extensive PrP deposition with florid plaques throughout the cerebrum and cerebellum).

Probable Case

Progressive neuropsychiatric disorder **and** duration of illness > 6 months[a] **and** positive tonsil biopsy.[b]

OR

Progressive neuropsychiatric disorder **and** duration of illness > 6 months.[a]

AND

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[a] Routine investigations do not suggest an alternative diagnosis, no history of potential iatrogenic exposure, and no evidence of a familial form of CJD.

[b] Tonsil biopsy is not recommended routinely, nor in cases with EEG appearances typical of sporadic CJD but may be useful in suspect cases in which the clinical features are compatible with vCJD and MRI does not show bilateral pulvinar high signal.

Four of the following: early psychiatric symptoms[c]; persistent painful sensory symptoms[d]; ataxia; myoclonus or chorea or dystonia; dementia.

AND

EEG does not show the typical appearance of sporadic CJD[e] in the early stages of illness.

AND

Bilateral pulvinar high signal on an MRI scan.

Suspect Case

Progressive neuropsychiatric disorder **and** duration of illness > 6 months.[a]

AND

Four of the following: early psychiatric symptoms[c]; persistent painful sensory symptoms[d]; ataxia; myoclonus or chorea or dystonia; dementia.

AND

EEG does not show the typical appearance of sporadic CJD[e] in the early stages of illness

Outbreak Case Definition

An outbreak case definition is established to reflect the context-specific circumstances (i.e., person, place, and time) of the cases under investigation and need to be created for each outbreak.

The case definition should be reviewed and modified, as necessary, throughout the course of the outbreak investigation, to ensure that the majority of cases are captured.

For guidance on developing an outbreak case definition refer to the *Infectious Diseases Protocol, 2023* (or as current).[3]

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[c] Depression, anxiety, apathy, withdrawal, delusions.

[d] This includes both frank pain and/or dysaesthesia.

[e] The typical appearance of the EEG in sporadic CJD consists of generalized triphasic periodic complexes at approximately one per second. These may occasionally be seen in the late stages of vCJD.

Clinical Information

Clinical Evidence

Clinically compatible signs and symptoms vary according to the type of CJD (i.e., classic [sporadic, genetic, iatrogenic] or variant). Refer to the clinical criteria outlined in the above case definitions for further details.

Clinical Presentation

CJD is a type of human prion disease (also known as transmissible spongiform encephalopathies), which are a group of rare, rapidly progressive, universally fatal neuro-degenerative syndromes.[5,6] They are characterized by neuronal degeneration, spongiform degeneration in parts of the brain (cerebral cortical, subcortical, and cerebellar gray matter), and the accumulation of abnormally misfolded partially protease-resistant prion proteins which are the transmissible agents.[5,6]

There are two main types of CJD: classic and variant. Classic CJD can be further characterized by the mode of transmission as either sporadic (~85% of cases), iatrogenic (~15% of cases), or genetic (~1% of cases).[7]

Classic CJD typically presents as a subacute illness in the middle-aged and elderly but has occasionally been reported in adolescents and younger adults. Its clinical presentation most commonly manifests as a rapidly progressive syndrome with confusion, behavioural and cognitive abnormalities (e.g., memory loss, personality changes), dementia, and variable other symptoms such as ataxia (poor gross motor coordination, including difficulty walking) and myoclonus (involuntary muscle twitching).[5,6] Death often occurs within weeks to months after the onset of illness.[5,6]

Variant CJD (vCJD) is associated with the ingestion of beef and/or beef products derived from cattle infected with bovine spongiform encephalopathy (BSE). It was first described in 1996.[5,6] vCJD typically has an earlier age of onset, compared to classic CJD, along with a longer clinical course. The disease then presents with psychiatric and/or behavioural abnormalities, followed by signs of neurological dysfunction, usually delayed by several months after the onset of illness.[5,6]

For further details on the differences between classic CJD and vCJD, refer to the summary table in the United States' Centers for Disease Control and Prevention's Clinical Overview of Variant Creutzfeldt-Jakob Disease.[6]

Laboratory Evidence

Laboratory Confirmation

The following constitutes laboratory confirmation of CJD:

- Confirmation of protease-resistant prion protein accumulation in brain tissue (e.g., by immunohistochemistry, paraffin-embedded tissue [PET] blot, or Western Blot), with histopathology demonstrating spongiform changes.

Additional Laboratory Evidence

The following may be used to support a probable case classification:

- Positive EP-QuIC in CSF (Sporadic, Iatrogenic CJD cases)
- *PRNP* gene sequencing (Genetic CJD cases)
- Confirmation of protease-resistant prion protein accumulation in tonsillar tissue (e.g., by immunochemistry, PET blot, or Western blot) [vCJD cases]

Indications and Limitations

- In Canada, EP-QuIC is performed by the National Microbiology Laboratory. A similar assay (the RT-QuIC) is used in other jurisdictions such as the US and European countries with similar performance characteristics.
- EP-QuIC is highly sensitive and specific for classic CJD with positive and negative predictive values of 96% and 99%, respectively.[8]
- *PRNP* gene sequencing should be considered if a genetic form of CJD is suspected, or if unable to determine the type of CJD based on clinical presentation and case history.
- Tonsil biopsy is not recommended routinely; however, it may be useful if vCJD is suspected on the basis of compatible clinical features in the absence of typical vCJD findings on a brain MRI.
- Demonstration of scrapie-associated fibrils by electron microscopy was historically part of the diagnostic criteria. Although historically important, this technique is no longer used for diagnostic purposes.

For further information about diagnostic testing, refer to PHO's Test Information Sheet for CJD.[9]

Case Management

In addition to the requirements set out in the Requirement #2 of the "Management of Infectious Diseases – Sporadic Cases" and "Investigation and Management of Infectious Diseases Outbreaks" sections of the *Infectious Diseases Protocol, 2023* (or as current)[3], the board of health shall investigate cases, including those that meet the probable case definition, to determine the source of infection. Refer to Provincial Reporting Requirements section above for relevant data to be collected during case investigation.

The following disease-specific exposure information should also be obtained during case management:

- History of invasive neurological or neurosurgical procedures, corneal transplants, or dura mater grafts; and
- History of treatment with human growth hormone or pituitary gonadotropin; and
- A family history of dementia.

Public health unit (PHU) investigation of cases occurs in collaboration with the attending clinician, the health care setting(s) where the individual is/was admitted (including their Infection Prevention and Control [IPAC] team), the Ministry of Health, PHO (available for consultation and advice, if needed), and the Public Health Agency of Canada (PHAC, see below).

For further guidance on the investigation and management of individuals considered to be at high risk of transmitting CJD iatrogenically, including patients suspected of having CJD (i.e., with rapidly progressive dementia that has not yet been diagnosed and CJD has not been ruled out), refer to PHAC's Classic Creutzfeldt-Jakob Disease in Canada: Quick Reference Guide 2007.[10]

There is no specific treatment available.[5-7]

Note: PHAC's CJD Surveillance System (CJDSS)[11] offers support to collaborating health professionals via consultation, laboratory investigations, logistic support, and education. With informed consent from patients or their representatives, a detailed medical chart review and family interview are also completed. The resulting diagnostic and epidemiologic information are assembled and used to support epidemiological analysis and public health decision-making. Attending clinicians and/or PHUs are strongly encouraged to contact PHAC's CJDSS when CJD is suspected. PHAC's CJDSS can be reached at 1-888-489-2999 or by email (cjdsurveillance@phac-aspc.gc.ca).

Contact Management

No public health action required.

Outbreak Management

Please see the *Infectious Diseases Protocol, 2023* (or as current)[3] for the public health management of outbreaks, or clusters, in order to identify the source of illness, manage the outbreak, and limit secondary spread (where applicable). Review each case for potential IPAC issues for follow up in institutional settings.

Prevention and Control Measures

Personal Prevention Measures

Measures that can be taken to prevent transmission and acquisition of CJD include:

- Excluding affected persons as well as their family members from donating blood, organs, and other body tissues;
- Where feasible, avoiding medical treatments and/or surgical procedures that carry potential risk for iatrogenic exposures (see Classic Creutzfeldt-Jakob Disease in Canada: Quick Reference Guide 2007;[10] and

Avoiding consumption of beef and/or beef products in countries with a non-negligible risk of BSE in cattle.

Infection Prevention and Control Strategies

Surgical instruments that have been in contact with high-infectivity tissue from affected persons (i.e., brain, spinal cord, pituitary gland, dura mater, posterior eye [optic nerve and retina], and CSF*) should be considered contaminated and must be appropriately discarded following the guidance outlined in PHAC's Classic Creutzfeldt-Jakob Disease in Canada: Quick Reference Guide 2007.[10]

*Note: While CSF itself is considered a low-infectivity tissue, contact with CSF implies contact with high-infectivity tissues (e.g., brain, spinal cord, dura mater). As a result, exposures to CSF should be managed in the same manner as high-infectivity tissues.

Single use cardiac catheters, pacemakers, and other single use devices are intended for use on a single patient, during a single procedure, and are not meant to be reprocessed or reused.

For further guidance on iatrogenic risks and IPAC measures, refer to Classic Creutzfeldt-Jakob Disease in Canada: Quick Reference Guide 2007.[10]

Disease Characteristics

Aetiologic Agent- The transmissible agent associated with CJD is an abnormally folded, unique protein called a prion. These misfolded prions can become a template, resulting in the conversion of normal proteins.[5-7] vCJD is associated with a particular prion agent that causes BSE in cattle (i.e., 'mad cow disease').[5-7]

Modes of Transmission- The mode of transmission for sporadic CJD is unknown; some cases of CJD have occurred iatrogenically, while others have a genetic component. vCJD is believed to be transmitted by consumption of specific risk materials from BSE-affected cattle; however, a small number of vCJD cases have also been transmitted by blood transfusion.[5-7]

Incubation Period- Incubation periods in prion diseases can be extremely long and are not applicable to naturally occurring sporadic and genetic cases, since these do not involve exposure to an external source.

In iatrogenic cases, the route of exposure determines the length of the incubation period: direct central nervous system (CNS) exposure results in an incubation period from 1.3 to 30 years, while peripheral exposure results in an incubation period of 5 to 42 years.[7]

It is estimated that the incubation period for vCJD cases related to exposure of BSE-affected cattle is from 10 to 20 years.[5] vCJD contracted via a blood transfusion has an incubation period estimated from 6.6 to 8.5 years.[7]

Period of Communicability- Transmissibility and period of communicability varies with the disease, the affected tissue, and the stage of disease. CNS and other tissues can cause transmission throughout symptomatic illness; lymphoid and other organs may cause transmission before signs of illness appear. Blood has been proven to cause transmission in the preclinical phase of vCJD.[7]

For further information regarding the infectivity of various tissues, refer to Classic Creutzfeldt-Jakob Disease in Canada: Quick Reference Guide 2007.[10]

Reservoir- Human cases constitute the only known reservoir for classic CJD. BSE-affected cattle were the original reservoir for vCJD; however, changes made in the management of livestock feeding and the introduction of specific risk material management processes during slaughter since the early 2000s have reduced the number of BSE-affected cattle significantly. Currently, subclinical (ongoing) cases in humans are considered to be a potential reservoir for secondary, human-to-human transmission of vCJD by blood transfusion, organ transplantation, or surgery.[5,7]

Host Susceptibility and Resistance- Genetic differences in susceptibility, resembling those of autosomal dominant traits, have been shown to explain patterns of occurrence of the disease in families.[5,7]

Please refer to Infectious Disease Trends in Ontario interactive tool for the most up-to-date information on CJD trends in Ontario.[12]

For information on national and international trends in the epidemiology of CJD, refer to the PHAC's CJDSS[11] and the World Health Organization.[13]

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Document History

Revision Date	Document Section	Description of Revisions
April 2022	Entire Document	New template. Appendix A and B merged. No material content changes.
April 2022	Epidemiology: Occurrence section	Removed.
April 2022	ICD Codes	Removed.
March 2026	Entire Document	Revision, edit and update.
March 2026	Case Definition	Expanded on the case definition section by specifying Sporadic, Iatrogenic, Genetic, and Variant case definitions. Further clarification on case definition for Classic/Sporadic CJD.