

# Appendix A: Disease-Specific Chapters

**Chapter: Creutzfeldt-Jakob Disease, all types**

Effective: February 2019

# Creutzfeldt-Jakob Disease, all types

Communicable

Virulent

**Health Protection and Promotion Act:  
O. Reg. 135/18 (Designation of Diseases)**

## 1.0 Aetiologic Agent

The infectious agents associated with Creutzfeldt-Jakob Disease (CJD) are abnormally folded, unique proteins called prions which potentially become a template causing the further conversion of normal proteins.<sup>1</sup>

## 2.0 Case Definition

### 2.1 Surveillance Case Definition

Refer to [Appendix B](#) for Case Definitions.

### 2.2 Outbreak Case Definition

The outbreak case definition varies with the outbreak under investigation. Please refer to the *Infectious Diseases Protocol, 2018* (or as current) for guidance in developing an outbreak case definition as needed.

The outbreak case definitions are established to reflect the disease and circumstances of the outbreak under investigation. The outbreak case definitions should be developed for each individual outbreak based on its characteristics, reviewed during the course of the outbreak, and modified if necessary, to ensure that the majority of cases are captured by the definition. The case definitions should be created in consideration of the outbreak definitions.

Outbreak cases may be classified by levels of probability (*i.e.* confirmed and/or probable).

## 3.0 Identification

### 3.1 Clinical Presentation

CJD is a prion disease and part of a group of rare, rapidly progressive, universally fatal neuro-degenerative syndromes that are characterized by neuronal degeneration, spongiform vacuolation in the cerebral gray matter, reactive proliferation of astrocytes and microglia, and accumulation of abnormal misfolded protease-resistant prion protein.<sup>1</sup>

Clinical presentation most commonly manifests as a rapidly progressive syndrome with confusion, behavioural and cognitive abnormalities, dementia, and variable other symptoms such as ataxia and myoclonus.<sup>1</sup>

Classic CJD can be sporadic (sCJD), familial or iatrogenic. It typically presents as a subacute illness in the middle-aged and elderly.

Variante CJD (vCJD) is another category, first described in 1996 and associated with ingesting meat from bovine spongiform encephalopathy (BSE) infected cattle.<sup>1</sup> vCJD has a longer clinical course than sCJD and usually presents with psychiatric or behavioural abnormalities, followed by signs of neurologic dysfunction, usually delayed by several months after the onset of illness.<sup>2</sup>

## 3.2 Diagnosis

See [Appendix B](#) for diagnostic criteria relevant to the Case Definitions.

# 4.0 Epidemiology

## 4.1 Occurrence

Worldwide, the highest age specific average mortality rate associated with classic CJD occurs in the 65-79 age group.<sup>2</sup> There are several classic CJD cases reported annually in Ontario. As of June 2016, approximately 231 cases of vCJD had been reported globally since 1995, most of which occurred in the United Kingdom.<sup>1</sup> Two cases were reported in Canada; one in 2002 and one case, which occurred in Ontario, in 2011.<sup>3</sup>

Please refer to Public Health Ontario's (PHO) Reportable Disease Trends in Ontario reporting tool and other reports for the most up-to-date information on infectious disease trends in Ontario.

<http://www.publichealthontario.ca/en/DataAndAnalytics/Pages/DataReports.aspx>

For additional national and international epidemiological information, please refer to the Public Health Agency of Canada and the World Health Organization.

## 4.2 Reservoir

Human cases constitute the only known reservoir for classic CJD. BSE-infected cattle were the original main 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-infected cattle significantly. Currently, subclinical (ongoing) infections in humans are considered to be a potential reservoir for secondary, human-to-human transmission of vCJD by blood transfusion, organ transplantation or surgery.<sup>2</sup>

## 4.3 Modes of Transmission

The mode of transmission for sporadic disease is unknown; some cases of CJD have occurred iatrogenically and some have a genetic component. vCJD is believed to be

transmitted by consumption of specific risk materials from BSE-infected cattle.<sup>2</sup> Three cases of vCJD have also been transmitted by blood transfusion.<sup>2</sup>

## 4.4 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 of infection. In iatrogenic cases, the route of exposure influences the length of the incubation period: direct 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.<sup>2</sup> It is estimated that the incubation period for vCJD cases related to exposure of BSE-infected cattle is from 10 to 20 years.<sup>2</sup> vCJD contracted via a transfusion of red cells has an incubation period estimated to be from 6.6 to 8.5 years.<sup>2</sup>

## 4.5 Period of Communicability

Transmissibility and period of communicability varies with disease, tissue involved and stage of disease. CNS and other tissues are infectious throughout symptomatic illness; lymphoid and other organs are probably infectious before signs of illness appear. Blood has been proven infectious in the preclinical phase of vCJD.<sup>2</sup>

For further information regarding the infectivity of various tissues please refer to Classic Creutzfeldt-Jakob Disease in Canada: Quick Reference Guide 2007.<sup>4</sup>

## 4.6 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.<sup>1,2</sup>

## 5.0 Reporting Requirements

As per Requirement #3 of the “Reporting of Infectious Diseases” section of the *Infectious Diseases Protocol, 2018* (or as current), the minimum data elements to be reported for each case are specified in the following:

- *Ontario Regulation 569* (Reports) under the *Health Protection and Promotion Act* (HPPA);<sup>5</sup>
- The iPHIS User Guides published by PHO; and
- Bulletins and directives issued by PHO.

## 6.0 Prevention and Control Measures

### 6.1 Personal Prevention Measures

Preventive measures include:<sup>2</sup>

- Excluding infected persons as well as their family members from donating blood, organs, and other body tissues;
- Avoiding iatrogenic exposures; and

- Avoiding exposures to the BSE-causing agent in food of bovine origin.

For further information regarding preventing iatrogenic transmission please refer to Classic Creutzfeldt-Jakob Disease in Canada: Quick Reference Guide 2007.<sup>4</sup>

## 6.2 Infection Prevention and Control Strategies

Surgical instruments that have been in contact with high risk tissue from infected persons, such as the brain, spinal cord, cornea, retina, pituitary gland, *dura mater*, and CSF should be considered contaminated and must be discarded or decontaminated and quarantined until the diagnosis is confirmed. Any surgical instruments that have contacted high risk tissue in a confirmed case of CJD should be discarded.<sup>4</sup>

Single use cardiac catheters, pacemakers, and other single use devices should not be re-used after being used on an infected person.

For further information regarding infection prevention and control please refer to Classic Creutzfeldt-Jakob Disease in Canada: Quick Reference Guide 2007.<sup>4</sup>

## 6.3 Management of Cases

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, 2018* (or as current), the board of health shall investigate cases to determine the source of infection. Refer to Section 5: Reporting Requirements above for relevant data to be collected during case investigation.

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

- History of invasive neurological or neuro-surgical procedures, corneal transplants;
- Any possible exposure to human growth hormone or transplacental tissue; and
- A family history of dementia.

Investigation of cases occurs in collaboration with the Ministry of Health and Long-Term Care, PHO and the Public Health Agency of Canada.

There is no specific treatment available.<sup>1</sup>

For further information please refer to Classic Creutzfeldt-Jakob Disease in Canada: Quick Reference Guide 2007.<sup>4</sup>

## 6.4 Management of Contacts

No public health action required.

## 6.5 Management of Outbreaks

Please see the *Infectious Diseases Protocol, 2018* (or as current) for the public health management of outbreaks or clusters in order to identify the source of illness, manage

the outbreak and limit secondary spread. Review case for potential IPC issues for follow up in institutional settings.

## 7.0 References

1. Committee on Infectious Diseases, American Academy of Pediatrics. Section 3: Summaries of Infectious Diseases: Prion Diseases - Transmissible Spongiform Encephalopathies. In: Kimberlin DW, Brady MT, Jackson MA, Long SS, editors. Red Book: 2018 Report of the Committee on Infectious Diseases. 31 ed. Itasca, IL: American Academy of Pediatrics; 2018.
2. Heymann DL, editor. Control of Communicable Diseases Manual. 20 ed. Washington, D.C: American Public Health Association; 2015.
3. Public Health Agency of Canada. Creutzfeldt-Jakob Disease Surveillance System Report [Internet]. Ottawa, ON: Her Majesty the Queen in Right of Canada; 2018 [updated May 14, 2018; cited June 11, 2018]. Available from: <https://www.canada.ca/en/public-health/services/surveillance/blood-safety-contribution-program/creutzfeldt-jakob-disease/cjd-surveillance-system.html#cases>
4. Public Health Agency of Canada. Classic Creutzfeldt-Jakob Disease in Canada: Quick Reference Guide 2007. Ottawa, ON: Her Majesty the Queen in Right of Canada; 2007. Available from: <https://www.canada.ca/en/public-health/services/infectious-diseases/nosocomial-occupational-infections/creutzfeldt-jakob-disease/infection-control-guidelines.html>
5. Health Protection and Promotion Act, R.S.O. 1990, Reg. 569, Reports, (2018). Available from: <https://www.ontario.ca/laws/regulation/900569>

## 8.0 Document History

**Table 1: History of Revisions**

<b>Revision Date</b>	<b>Document Section</b>	<b>Description of Revisions</b>
March 2017	Aetiologic Agent	Terminology revised to provide a better explanation of the aetiology of CJD

Revision Date	Document Section	Description of Revisions
March 2017	Epidemiology	<p>4.1: Whole section revised</p> <p>4.2: Whole section revised</p> <p>4.3: Last sentence updated to “Three cases of vCJD have also been transmitted by blood transfusion”</p> <p>4.4: Whole section revised</p> <p>4.5:</p> <ul style="list-style-type: none"> <li>- Paragraph 1, Delete “(up to 40 months before onset of clinical symptoms, ref 51)”</li> <li>- Paragraph 2, At the beginning, add “For further information regarding the infectivity of various tissue please” and at the end, add “Quick reference guide 2007 (<a href="http://www.phac-aspc.gc.ca/nois-sinp/cjd/cjd-eng.php">http://www.phac-aspc.gc.ca/nois-sinp/cjd/cjd-eng.php</a>)”</li> </ul> <p>4.6: Delete “Codon 129 of the prion protein gene (PRNP) is a polymorphic locus; homozygosity for methionine has been associated with all cases of vCJD so far.”</p>
March 2017	Reporting Requirements	5.1: add “, probable” after “confirmed” and before “and suspect...”

Revision Date	Document Section	Description of Revisions
March 2017	Prevention and Control Measures	<p>6.1: change “Preventative” to “Preventive”</p> <ul style="list-style-type: none"> <li>- Bullet 1: At the beginning, add “Excluding”, delete “should be excluded” after “member” and delete “and” after “...tissue;”</li> </ul> <p>Add bullets two and three respectively:</p> <ul style="list-style-type: none"> <li>- “Avoiding iatrogenic exposures”</li> <li>- “Avoiding exposures to the BSE-causing agent in food of bovine origin”</li> </ul> <p>Delete bullet 4 “Persons diagnosed with this infection who have made donations as listed above are to be reported to Canadian Blood Services (CBS) to enable look back and recall procedures.”</p> <p>Update last paragraph to, “Refer to Public Health Agency of Canada. Classic Creutzfeldt-Jakob disease in Canada: quick reference guide. Ottawa, ON: Her Majesty the Queen in Right of Canada; 2007 [cited 2017 Mar 8]. Available from: <a href="http://www.phac-aspc.gc.ca/nois-sinp/cjd/cjd-eng.php">http://www.phac-aspc.gc.ca/nois-sinp/cjd/cjd-eng.php</a>”</p> <p>6.2:</p> <p>Paragraph 1: Add “the” after “...such as” and before “brain, spinal cord...”.</p> <p>Add “gland” after “...pituitary” and before “dura mater and CFS,...”.</p> <p>Add “discarded or decontaminated and quarantined until the diagnosis is confirmed. Any surgical instruments that have contacted high risk tissue in a confirmed case of CJD should be discarded.” after “...contaminated and must be”</p> <p>Delete “inactivated and followed with appropriate disinfection and sterilization procedures.”</p>



<b>Revision Date</b>	<b>Document Section</b>	<b>Description of Revisions</b>
March 2017	Prevention and Control Measures	<p>6.2: Paragraph 3: Update to “Refer to Public Health Agency of Canada. Classic Creutzfeldt-Jakob disease in Canada: quick reference guide. Ottawa, ON: Her Majesty the Queen in Right of Canada; 2007 [cited 2017 Mar 8]. Available from: <a href="http://www.phac-aspc.gc.ca/nois-sinp/cjd/cjd-eng.php">http://www.phac-aspc.gc.ca/nois-sinp/cjd/cjd-eng.php</a></p> <p>6.3: - Delete “As per Ontario Regulation 338/96, report past blood donations / transfusions to Canadian Blood Services.” - Add “Refer to Public Health Agency of Canada. Classic Creutzfeldt-Jakob disease in Canada: quick reference guide. Ottawa, ON: Her Majesty the Queen in Right of Canada; 2007 [cited 2017 Mar 8]. Available from: <a href="http://www.phac-aspc.gc.ca/nois-sinp/cjd/cjd-eng.php">http://www.phac-aspc.gc.ca/nois-sinp/cjd/cjd-eng.php</a>”</p>
March 2017	References	Revised
March 2017	Document History	Updated
February 2019	General	Minor revisions were made to support the regulation change to Diseases of Public Health Significance. Common text included in all Disease Specific chapters: Surveillance Case Definition, Outbreak Case Definition, Diagnosis, Reporting Requirements, Management of Cases, and Management of Outbreaks. The epidemiology section and references were updated and Section 8.0 Additional Resources was deleted.
February 2019	3.1 Clinical Presentation	Minor revisions to entire section.
February 2019	4.2 Reservoir	Added “blood transfusion, organ transplantation or surgery” to source of secondary infection in last sentence.

<b>Revision Date</b>	<b>Document Section</b>	<b>Description of Revisions</b>
February 2019	4.4 Incubation Period	Iatrogenic cases incubation period updated.

