

Creutzfeldt-Jakob Disease - Classic and Variant

Revision Dates

Case Definition	January 2013
Reporting Requirements	May 2018
Remainder of the Guideline (i.e., Etiology to References sections inclusive)	January 2011

A. Sporadic Creutzfeldt-Jakob Disease (sCJD)

Confirmed sCJD

Neuropathologically and/or immunocytochemically and/or biochemically confirmed, via observation of one or more neuropathologic features (see Box 1) and no evidence of iatrogenic CJD or genetic human prion disease (see Sections B and C).

Box 1

I	Spongiform encephalopathy in cerebral and/or cerebellar cortex and/or subcortical grey matter
II	Encephalopathy with prion protein (PrP) immunoreactivity in plaque-like and/or diffuse synaptic and/or patchy/perivacuolar patterns, by examination of tissue either directly or with assistance of capillary transfer from paraffin-embedded tissue (PET) to secondary support (PET blot)
III	Presence of scrapie-associated fibrils (SAF) by electron microscopy
IV	Presence of protease-resistant PrP by Western blot

Probable sCJD

Routine investigation should not suggest an alternative diagnosis.

Rapidly progressive dementia **AND** at least two features from list I and the feature in list II (see Box 2)

OR

Possible CJD **AND** duration < 2 years **AND** cerebrospinal fluid positive for:

- 14-3-3 protein^a by Western Blot

OR

- ≥ 976 pg/ml for the tau protein by ELISA

OR

- ≥ 2.5 µg/l (2.5ng/ml) for the S100B protein by ELISA^b

^a 14-3-3 protein positivity or presence of increased levels of tau protein or S100B protein must be present with other criteria **AND** only for probable case definition because of limited usefulness due to poor specificity.

^b link to ProvLab bulletin regarding the use and interpretation of these tests: [ProvLab Partner Updates - Laboratory Bulletins](#)

Possible sCJD

Rapidly progressive dementia **AND** two of list I (see Box 2) **AND** duration < 2 years **AND** no electroencephalography (EEG) or Atypical EEG.

Box 2

I	A	Myoclonus
	B	Visual disturbances or cerebellar dysfunction (ataxia)
	C	Pyramidal or extrapyramidal features
	D	Akinetic mutism
II	Typical electroencephalographic pattern: periodic sharp-wave complexes ca. 1 Hz	

B. Iatrogenic CJD (iCJD)

Confirmed iCJD

Definite CJD (see Section A for diagnostic criteria) with a recognized risk factor for iatrogenic transmission (see Box 3).

Probable iCJD

Progressive predominant cerebellar syndrome in a recipient of cadaverically derived human pituitary growth hormone

OR

Probable CJD (see Section A for diagnostic criteria) with a recognized risk factor for iatrogenic transmission (see Box 3).

Box 3

Note: Assessment of the relevance of any proposed risk factor to disease causation should take into account the timing of the putative exposure in relation to disease onset, especially where the putative exposure is recent. This list is also provisional, as the risks of iatrogenic transmission of prion disease by other routes are currently incompletely understood.

I	Treatment with human cadaveric pituitary growth hormone, human pituitary gonadotrophin or human dura mater graft
II	Corneal graft in which the corneal donor has been classified as having a definite or probable prion disease
II	Neurosurgical exposure to instruments previously used on a patient classified as having definite or probable prion disease

C. Genetic Prion Diseases

Confirmed Genetic Prion Disease

Definite (pathologically confirmed) prion disease **AND** definite or probable prion disease in a first-degree relative

OR

Definite prion disease **AND** pathogenic mutation in prion protein gene (*PRNP*) (see Box 4)

OR

Typical neuropathologic phenotype of Gerstmann-Sträussler-Scheinker disease (GSS).^a

Probable Genetic Prion Disease

Progressive neuropsychiatric disorder **AND** definite or probable prion disease in a first-degree relative

OR

Progressive neuropsychiatric disorder **AND** pathogenic mutation in *PRNP* (see Box 4).

^a Presence of multicentric PrP-immunoreactive plaques in cerebral and/or cerebellar cortex, with neuron loss and spongiosis. Other large amorphous plaques or neurofibrillary tangles immunoreactive for PrP have been described in subsets of GSS but these are associated with less-frequent *PRNP* mutations (A117V and F198S). Florid or Kuru plaques are not considered diagnostic for GSS.

Box 4

I	<i>PRNP</i> mutations associated with a neuropathologic phenotype of CJD*: P105T; G114V; R148H; D178N; V180I; V180I+M232R; T183A; T188A; T193I; E196K; E200K; V203I; R208H; V210I; E211Q; M232R; octapeptide repeat insertions (various lengths) and deletion (48 bp)
II	<i>PRNP</i> mutations associated with a neuropathologic phenotype of GSS (see note ^b above): P102L; P105L; A117V; G131V; A133V; Y145Stop; H187R; F198S; D202N; Q212P; Q217R; M232T; octapeptide repeat insertions (various lengths)
III	<i>PRNP</i> mutations associated with a neuropathologic phenotype of Familial Fatal Insomnia (FFI): D178N
IV	<i>PRNP</i> mutations associated with other neuropathologic phenotypes: I138M; G142S; Q160Stop; T188K; T188R; P238S; M232R; octapeptide repeat insertions (various lengths)
* See Section A	

D. Variant Creutzfeldt-Jakob Disease (vCJD)

Confirmed vCJD

IA and neuropathologic confirmation as per pathologic features (see Box 5 ^a)

Probable vCJD

I AND 4 or 5 criteria of II + IIIA + IIIB (see Box 5)

OR

I AND IVA

Possible vCJD

I AND 4 or 5 criteria of II + IIIA (see Box 5)

Box 5

I	A	Progressive neuropsychiatric disorder
	B	Duration > 6 months
	C	Routine investigations do not suggest alternative diagnosis
	D	No history of potential iatrogenic exposure
	E	No evidence of genetic prion disease
II	A	Early psychiatric symptoms ^b
	B	Persistent painful sensory symptoms ^c
	C	Ataxia
	D	Myoclonus or chorea or dystonia
	E	Dementia
III	A	EEG does not show typical appearance of sporadic CJD ^d (or no EEG performed) in the early stages of the illness
	B	Bilateral pulvinar high signal on magnetic resonance imaging (MRI) scan ^e
IV	A	Tonsil biopsy positive for prion protein immunoreactivity ^f
^a Spongiform change, extensive PrP deposition, florid plaques throughout cerebrum & cerebellum ^b Depression, anxiety, apathy, withdrawal, delusions ^c Frank pain and/or dysaesthesia ^d Generalized triphasic periodic complexes at ca. 1 Hz. Rarely, these may occur in the late stages of vCJD. ^e Relative to the signal intensity of other deep grey matter nuclei and cortical grey matter ^f Tonsil biopsy is not recommended routinely, or 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 where MRI does not show bilateral pulvinar high signal.		

NOTE: Genetic analysis is required in every suspected case to exclude familial CJD; patients should have no history of exposure to human pituitary-derived products or any other source of iatrogenic CJD. Surveillance case definitions and diagnostic criteria for CJD are subject to change due to the introduction of new diagnostic testing procedures and increased surveillance efforts.

Reporting Requirements

1. Physicians, Health Practitioners and others

Physicians, health practitioners and others shall notify the Medical Officer of Health (MOH) (or designate) of the zone, in the prescribed form by mail, fax or electronic transfer within 48 hours (two business days) about the following:

- All confirmed, probable, and possible cases of CJD and vCJD.

2. Laboratories

All laboratories shall report all positive laboratory results (diagnostic and pathology* lab results) indicative of CJD and vCJD by mail, fax or electronic transfer within 48 hours (two business days) to the:

- Chief Medical Officer of Health (CMOH) (or designate), and
- MOH (or designate) of the zone.

* Testing currently performed at Centre for Research in Neurodegenerative Diseases in Toronto, Ontario.

3. Alberta Health Services and First Nations and Inuit Health Branch

- The MOH (or designate) of the zone where the case currently resides shall forward the initial Notifiable Disease Report (NDR) of all confirmed, probable and possible cases of CJD and vCJD to the CMOH (or designate) within two weeks of notification and the final NDR (amendment) within four weeks of notification.
- For out-of-province and out-of-country reports, the following information should be forwarded to the CMOH (or designate) by phone, fax or electronic transfer within 48 hours (two business days):
 - name,
 - date of birth,
 - out-of-province health care number,
 - out-of-province address and phone number,
 - positive laboratory report, and
 - other relevant clinical / epidemiological information.

4. Additional Reporting Requirements

- Beginning in 1998, Health Canada launched an intensive active surveillance program. The Creutzfeldt-Jakob Disease Surveillance System (CJD-SS) is a research project that relies on direct CJD reporting by all neurologists, neurosurgeons, neuropathologists, geriatricians, and infectious disease physicians to the Public Health Agency of Canada.(1)
- Canadian Blood Services (CBS): All persons with confirmed, probable or possible vCJD must be reported by the zone MOH (or designate) to CBS within two working days if they have ever had a history of donating or receiving blood in Canada.
- Funeral Directors: A funeral director, embalmer, or other person who knows or has reason to believe that a person was infected with a specified disease (including CJD) at the time of death must, within 12 hours, notify the zone MOH of the zone in which the person died by telephone, email or facsimile. Refer to [Bodies of Deceased Persons Regulation](#).

Etiology

Transmissible spongiform encephalopathies (TSEs), also known as prion diseases, are a unique group of infectious fatal neurodegenerative disorders caused by a toxic gain of function in a normal host cell protein (the prion protein PrP).(2) These disorders are characterized pathologically by microscopic vacuoles (spongiform change) and the deposition of amyloid (prion) protein (PrP) in the grey matter of the brain. It is thought that these prions are rogue forms of a normal protein found in the brain.(3) The TSE agents are hardy, remain infectious for years in dried state(4), and resist all routine sterilization and disinfection procedures commonly used.(5) Human TSE includes CJD, which is classified as sporadic CJD (sCJD), genetic (familial or inherited), and acquired, which includes iatrogenic (iCJD) and variant CJD (vCJD).(3) sCJD occurs worldwide and is of unknown etiology.(6) Genetic forms of CJD are linked to mutations of the prion protein gene (PRNP). iCJD is caused by the transmission of infection from person to person in the course of medical treatment. vCJD is a novel form of human TSE, which has been linked to transmission of BSE to humans, likely representing the first zoonotic transmission of a TSE to the human population.(6;7)

Clinical Presentation

TSE refers to a group of diseases, inherited or infectious, that may occur in animals and humans, which share common characteristics related to the deterioration of the central nervous system.(8) Non-human TSE includes scrapie in sheep and goats, bovine spongiform encephalopathy (BSE) in cattle, encephalopathies in mink, cats, and chronic wasting disease in mule deer and elk.(2)

The clinical presentation of CJD varies by presumed etiology, however, CJD in all forms is invariably fatal, and there is no treatment. In sCJD, neurodegeneration is rapidly progressive, and signs and symptoms include progressive dementia and cognitive decline, myoclonus, pyramidal or extrapyramidal symptoms (such as spasticity, hyperreflexia, or tremor and rigor), visual disturbances, cerebellar ataxia, and akinetic mutism.(3;6;7) The electroencephalography (EEG) often shows a disease-specific pattern of periodic synchronous discharge (PSD) consisting of periodic triphasic waves at 1–2 cycles, which is not normally seen in the genetic or acquired forms of CJD.(9) The clinical progression is typically weeks long, progressing to death in a matter of a few months.(10) The median duration of illness is 4 months with a range of 1 – 18 months.(11) Median age of onset is between 55–65 years of age, but it has been reported to have occurred in persons as young as 14 years, and in a person 92 years of age.(3)

Genetic CJD illness, which is associated with a variety of mutations of the PrP gene (PRNP) on chromosome 20 (6;11), occurs earlier than sCJD (mean age at onset is 58 years, range 33 – 84 years of age)(12), with variability in the clinical and pathological findings, the age of onset, and the duration of illness depending on the particular PrP mutation involvement.(9) Higher-order brain activities are affected at a much later stage of disease, with manifestations ranging from forgetfulness, reduced intellectual performance, to severe dementia. Individuals with genetic forms of CJD also show signs of motor and sensory peripheral neuropathy rarely found in those with sCJD.(12) The duration of illness in genetic CJD is longer than sCJD, and the patient may survive for several years after the onset of symptoms.(13)

Persons diagnosed with vCJD have an earlier age of onset (mean age of 26 years)(3;14) and present with behavioural changes or psychiatric symptoms such as agitation, aggression, anxiety, depression, and poor concentration.(3) In addition, sensory disturbances such as pain and paresthesias may also appear early.(3) The median duration of illness is about 14 months.(3;7) Neurological signs normally begin to appear about 6 months after the onset of illness.(3) Cerebellar ataxia often appears early, and may be followed by cognitive impairment, involuntary movements, and incontinence of urine. As neurological deterioration progresses, the patient may become mute, immobile, and unresponsive.(3) With vCJD, in addition to PrP deposits in the brain, a distinctive pattern of spongiform change occurs,

where the holes are particularly numerous around dense microscopic protein deposits called florid plaques. These structures in the brain are used by the pathologist to help distinguish vCJD from other forms of CJD. These plaques are due to the abnormal PrP molecular strain type 4 which is unique to vCJD. It has commonly been detected in lymphoreticular tissues of patients with vCJD, but not in such tissues of patients with other forms of CJD.(13) vCJD can be diagnosed in the appropriate clinical context by a tonsil biopsy.(15) The duration of illness is longer than classical CJD, lasting a median of 14 months (range 8 – 38 months).

The clinical presentation of iCJD cases varies, as the route of inoculation of the infectious agent appears to be an important determination of clinical presentation.(16) Early onset of CJD should prompt a thorough search for possible iatrogenic source of infection.(17) All CJD types present with cerebellar or gait disturbances, followed by dementia/cognitive decline/memory impairment. Cerebellar/gait disturbance, myoclonus, and visual/oculomotor disturbance were the most common symptoms throughout the course of illness.(18)

Diagnosis

A definitive diagnosis of any form of CJD requires neuropathological examination of brain tissue. (4;13;17) This would usually be undertaken at post mortem examination.(15) Rarely, a biopsy of the brain may be taken in life but this is not usually necessary in the investigation of cases of possible CJD. (17) A protein assay that detects the 14-3-3 protein in cerebrospinal fluid (CSF) is used as a diagnostic test during clinical illness, and when the test is performed with due regard to the clinical picture, a positive result has approximately 90% sensitivity and specificity for sCJD,(19) but the sensitivity is lower in patients with other forms of CJD.(20) Sensitivity tends to increase as the illness progresses, so a negative test early in the illness should always be repeated at a later date.(21) The limitation of 14-3-3 protein detection in sCJD is non-specificity and so this test should be reserved for confirmation and not for screening.(2) In addition, false positives occur in a range of diseases.(17) Other investigations that may be useful include EEG (presence of periodic, 1-2 Hz sharp wave discharges) and MRI (typical findings, if present, include high signal in the caudate nucleus and putamen).(2) In vCJD, the EEG does not show the typical finding seen in patients with sCJD, and CSF 14-3-3 protein sensitivities are much lower (25 to 60%). The MRI shows the “pulvinar sign” (high T2 MRI signal in the posterior thalamus) in about 75% of cases.(2) The recognition that patients with vCJD have PrP in extraneural sites, including lymphoreticular tissues, means that diagnosis can be made antemortem by tonsil biopsy.(2;13;14)

Epidemiology

Reservoir

Humans are the only proven reservoir of CJD. Bovine spongiform encephalopathy (BSE) infected cattle are suspected to be the reservoir of vCJD.(17)

Transmission

CJD, first described in the 1920s, was recognized as a transmissible disease in the mid-1960s.(14) CJD and other human TSEs are not known to spread by direct contact from person to person or by the airborne or respiratory route. Spouses and other household contacts of sCJD patients have no higher risk of contracting the disease than the general population.(22) Iatrogenic spread is exceedingly rare. Since the first evidence of iatrogenic transmission of CJD in 1974, via a corneal transplant, other mechanisms of iatrogenic transmission have been identified.(16) The following describes persons at risk of iCJD:

- Recipients of human tissue derived pituitary hormone treatment (either growth hormone or gonadotrophin).
- Recipients of dura matter graft (until 1992 for Lyodura grafts, until 1997 for Tutoplast Dura grafts).

- Recipients of 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.(23)

With vCJD, infection has been linked to consuming contaminated beef products from animals infected with BSE(5) or “mad-cow disease”. Until recently the risk of developing CJD as a consequence of a blood transfusion was a theoretical concern. However, there have been four highly probable instances of vCJD transmission by non-leucocyte depleted red blood cell concentrates. In December 2003 a patient died from vCJD after receiving a blood transfusion from a donor who subsequently also had vCJD. Since then, three further patients have been identified.(24) There continues to be no evidence of transmission of other forms of CJD via transfusion.(25) Current information on vCJD indicates that lymphoreticular tissues are involved and that the infectivity is different from that of other forms of CJD. Patients with vCJD might therefore pose a greater risk of transmitting iatrogenic infections than those with sCJD.(4)

CJD is not known to be transmitted from mother to child during pregnancy and/or childbirth.(11) There have been no proven association of CJD with dentistry, however, the risk of transmission through dentistry is unclear, as there is theoretical risk of transmission of prion disease through dental treatment.(26) This theoretical risk of transmission emphasizes the need to maintain optimal standards of infection control and decontamination procedures.(27) Guidelines produced by the Alberta Dental Association and College offer that dental instruments and devices touching pulpal tissue must be discarded in sharp containers after each patient use, and not reprocessed when CJD or vCJD is diagnosed or suspected.(28)

Distribution of Infectivity in the Human Body

Using evidence from animal studies and reports of infection through iatrogenic exposure, human tissue is classified into three categories, according to its risk of transmitting CJD. The Public Health Agency of Canada (PHAC) [Infection Control Guidelines: Classic Creutzfeldt-Jakob Disease in Canada, Quick Reference Guide 2007](#) and the World Health Organization (WHO) [Guidelines on Tissue Infectivity Distribution in Transmissible Spongiform Encephalopathies 2006](#) provide detailed information on this issue.

Preventing Iatrogenic Transmission(23)

The most effective, safe, and efficient means of preventing iatrogenic transmission of CJD are to identify high-risk patients prior to any invasive procedures, in order to implement the required infection prevention and control measures, and to have a system for instrument tracking.

Patients considered to be at high risk of transmitting CJD iatrogenically are those diagnosed, prospectively or retrospectively, with:

- CJD – confirmed, probable, or possible CJD (all forms), depending on pathological, laboratory, and clinical evidence and following the case definitions (included at the front of this document).
- Suspected CJD – undiagnosed, rapidly progressive dementia and CJD not ruled out.
- Asymptomatic carrier of genetic TSE – a person who displays no symptoms or signs of TSE, but meets one or more of the following criteria:
 - The person has been confirmed by genetic testing to carry a genetic mutation causative of genetic CJD;
 - 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;
 - 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.

The Health Canada [Infection control guideline: Classic Creutzfeldt-Jakob disease in Canada 2002](#) and PHAC [Infection Control Guidelines: Classic Creutzfeldt-Jakob Disease in Canada, Quick Reference Guide 2007](#) guidelines outline appropriate procedures for managing instruments that have been in contact with high-risk patients, and should be referred to for detailed information.

Incubation Period

The incubation period for naturally occurring sCJD is not known. In iCJD, the incubation period and clinical presentation are determined by the titre of inoculum and site of inoculation.(2) With iCJD, the mean incubation period is 1.5 – 1.6 years following direct intracerebral contact of infectious material by neurosurgery of depth electrodes.(7) As the inoculation site moves further away from the brain to other tissues, the incubation period is extended. For example, incubation periods ranged from 1.5 – 18 years after exposure to contaminated dura mater, while transmission from exposure through a peripheral route (as with human growth hormone [hGH] injections) is associated with an incubation period ranging from 5 – 30 years.(15) The incubation period for vCJD is unknown, however, it is likely the incubation period will be measured in terms of many years or decades.(30) The concept of incubation period is not applicable to genetic forms of CJD.(31)

Period of Communicability

In prion diseases, generally the highest levels of infectivity are associated with the central nervous system (CNS) and related tissues (e.g., parts of the eye) during, and throughout clinical illness. In sCJD, non-CNS tissues may be infective, but at much lower levels, probably during the period of clinical illness. In vCJD, infection is present in lymphoid tissues and blood during the incubation period and during clinical illness. Late in the incubation period, the level of infectivity in the CNS rises, and high levels of infectivity occur in the CNS throughout symptomatic illness.(29)

Host Susceptibility

Mutations of the prion protein gene (PRNP) are associated with genetic forms of human prion disease. Polymorphic regions of the PRNP influence susceptibility to infection and incubation period in animal species. In human disease, the genotype at codon 129 of the PRNP influences susceptibility to sCJD, vCJD, and iCJD, and also has potential effects on the incubation period in acquired forms of CJD (i.e. iCJD and potentially vCJD).(31)

Occurrence

General

Globally, the majority of cases (approximately 85 – 90%) of CJD occur as a sporadic disease, with genetic CJD accounting for approximately 10 – 15%(3;15), and iCJD and vCJD representing a very small proportion of cases (less than 1%).(15) CJD is considered a rare disease, affecting about one person in every one million people per year worldwide(20), with equal incidence in men and women.(12)

Iatrogenic CJD is very rare and is a result of transmission during invasive medical interventions where the source patient had CJD.(15) Worldwide, since the first report in 1974 – 2006, 405 documented iatrogenic transmissions of CJD have been reported. New cases continue to appear, however, since 2000 there has been a downward trend in the number of newly diagnosed cases. In particular, cases due to contaminated human growth hormone (hGH) and dura mater grafts have subsided, and new cases that occur each year are the result of longer and longer incubation periods following infections acquired during the 1980s.(32) As of 2006, two confirmed cases and one possible case of corneal graft-related CJD have been reported worldwide. Cadaver derived dura mater transplant infections were first reported in 1987. As of 2006, 196 cases have been reported worldwide. By 1985, it was

recognized that injected human cadaver extracted pituitary growth hormone and human pituitary gonadotropin administered by injection could be transmitted between humans. As of 2006, 198 cases of CJD worldwide have been related to growth hormone (194) and gonadotropic hormone(4) of human origin.(32) The use of cadaver-extracted hGH has been replaced with a recombinant form of the hormone.

vCJD was first diagnosed in the United Kingdom in 1996. As of June 2010, a total of 217 patients with this disease from 11 countries have been identified. vCJD cases have been reported from the following countries: 170 from the United Kingdom, 25 from France, 5 from Spain, 4 from Ireland, 3 from the United States, 3 in the Netherlands, 2 in Portugal, 2 in Italy, and one each from Canada, Japan, and Saudi Arabia.(33) Two of the three U.S. cases, two of the four cases from Ireland and the single cases from Canada and Japan were likely exposed to the BSE agent while residing in the United Kingdom. One of the 25 French cases may also have been infected in the United Kingdom.(30)

Canada

In Canada, 390 deaths attributed to CJD were recorded from 1979 – 1995. Forty-three per cent of the deaths occurred among people in their sixties, which corresponds with the peak age of onset for sCJD, and the male-to-female ratio was 1:1. The number of deaths ranged from 14 – 34 per year. The crude annual mortality rates for the period 1979 – 1995 ranged from 0.6 – 1.1 cases per million population; this is consistent with estimates of incidence worldwide.(34)

The Public Health Agency of Canada (PHAC), through the Creutzfeldt-Jakob Disease Surveillance System (CJD-SS), investigates 70 – 100 reports of suspected cases each year. In 2008, there were 100 referrals of suspected cases of CJD to the CJD-SS; in 2009 there were 96 referrals. There are between 30 – 40 deaths attributed to CJD annually in Canada, with the vast majority being cases of sCJD. In 2007, there were 35 CJD deaths (definite and probable cases) reported by the CJD-SS, with 32 classified as sCJD, and 3 classified as genetic CJD. In 2008 (provisional data), there were 38 CJD deaths; all classified as cases of sCJD.(35) There have been four cases involving dura mater grafts and there have been no cases of corneal graft-related CJD(4) or hGH-associated CJD reported in Canada.(5)

The first confirmed case of vCJD was reported in April 2002. It was reported as being acquired while the individual was visiting in the United Kingdom.(30;36;37) There have been no cases of vCJD linked to eating Canadian beef(37) although BSE has been reported in Canadian cattle. A case of BSE was reported in Canada in 1993 in a cow that had been imported from the U.K. in 1987.(38) In 2003, a case of BSE was reported in a cow from northern Alberta. Since then additional cases have been identified in Canada, however, the risk of exposure to BSE in Canada is considered extremely low due to safeguards put in place to identify and reduce the potential for the spread of BSE. (39;40)

Alberta

Reporting for CJD began in 1983 and for vCJD in 1998. From 1983 to mid 1998, 9 cases of CJD were reported in the province (range of 0 – 4 cases annually). From July 1998 to 2004, 10 cases of CJD were reported; six were confirmed, one probable and one suspect. For two cases, the final diagnosis is unknown. From 2005 – 2009, 16 cases of CJD were reported, with 5 of these cases being confirmed, 9 probable, and 2 cases from 2009 are classified as possible. No cases of vCJD have been reported in the province.(41)

Key Investigation

- Identify the potential source of infection (relevant exposure).
 - Invasive neurological or neurosurgical procedures including dura mater grafting.
 - Corneal transplant.
 - Exposure to human growth hormone.
 - Living in or extended travel to an area of high incidence (vCJD) for a cumulative period of longer than three months from 1980 to 1996.
 - Family history of dementia.
- History of blood or blood product donation.
- History of blood or blood product receipt.
- Obtain a medical history.

Control

Management of a Case (4;42)

- As most patients become cognitively impaired, it may be necessary to obtain information from a family member.
- Care of Client in the Home or Health Care Setting.
 - Current evidence suggests that normal social or routine contact does not present a risk to health care workers, families or others.
 - Patients with CJD and vCJD may receive care on a regular hospital unit or at home with no special precautions other than routine practices that would apply to any other patients with the exception of invasive procedures involving high and low infectivity tissues.
 - CJD is not thought to present a risk through normal social or routine contact. Families caring for patients at home should be advised of the routine infection control practice that would apply to them. They should have gloves, paper towels, bags, and sharps containers as appropriate.
 - A private room is not necessary.
 - Feeding utensils, feeding tubes, suction tubes, razors or personal care items do not require special precautions.
 - Care settings are responsible for notifying funeral homes or other organizations that may be handling high or low infectivity tissues from a high-risk client or high infectivity tissues and CSF from a high-risk client e.g., mortuary procedures, laboratory investigations.
 - Clients at high-risk or at-risk for transmission of CJD, or their responsible caregiver should notify their doctors, dentists or other health care workers of their status so that precautions can be implemented as required.
 - Local infection control personnel should be consulted for more detailed information on infection control policies in acute care facilities regarding CJD.

Treatment of a Case

- Supportive therapy as no treatment is available.

Management of Contacts (4;23)

- In cases of potential exposure (i.e., confirmed CJD case diagnosed after undergoing surgery with re-usable instruments) decisions regarding whether or not to inform contacts of equipment should be made with the involvement of various stakeholders.
- As no specific diagnostic test for CJD or the presence of prions is available, contact notification must be considered on a case-by-case basis.

Preventive Measures

- Handling of specimens(4)
 - Special precautions are only required for handling high or low infectivity tissues from a high-risk client or high infectivity tissues and CSF from an at-risk client.
 - The specimens should be collected in a sealed, leak-proof, puncture resistant container and clearly labelled “high-risk for CJD”.
 - Decontamination process is determined based upon the infectivity level of the material/tissue involved.
- Special Infection Prevention Measures - Surgical instruments and decontamination procedures
 - Effective decontamination procedures of re-usable surgical instruments used on confirmed, probable, or possible cases of CJD should be followed. These decontamination procedures require special measures, which are described in the Canadian national guideline documents.(4;23)
- Occupational injury/exposure
 - There have been no confirmed cases of occupational transmission of CJD to humans.(4;17)
 - Cases of CJD in health care workers have been reported in which a link to occupational exposure is suggested, however, the incidence of CJD is not higher in this occupational group than in the general population.(4)
- Post-exposure counseling of a HCW who has been exposed to high infectivity tissue from a high-risk patient/client should include the fact that no case of human TSE is known to have occurred through occupational accident or injury. For the present, a common-sense approach to post-exposure is recommended. For example:
 - Wash unbroken skin thoroughly with soap and water,
 - Gently encourage bleeding of needlestick injuries and other lacerations,
 - Wash broken skin with soap and water; irrigate the eye or mouth with saline or tap water, and report all exposures according to normal procedures for your hospital or health care facility.(4)
- Persons with CJD, vCJD or other forms of dementia are not eligible to donate blood, organs or other body tissues or fluids.
- Individuals who have lived in or visited high-risk countries for BSE may have been exposed to meat or meat products from cattle infected with BSE and are excluded from donating blood in Canada. This includes individuals who have:
 - Spent a cumulative period of three months or more in the United Kingdom or France between the years 1980 and 1996, or
 - Spent a cumulative period of five years or more in countries in Western Europe from 1980 to present, or
 - Received a transfusion of whole blood or blood components in the U.K., France, or elsewhere in Europe from 1980 to the present.(43)
- People who are at risk (see Transmission - Client Risk of CJD) of developing CJD and blood relatives (parent, child, sibling) of people with familial CJD are not eligible to donate blood or blood products.(43)
- After death:
 - Refer to the Alberta [Bodies of Deceased Persons Regulation](#) for procedures that describe how to label, transport, and embalm the body.
 - On the death of a patient with confirmed, probable or possible CJD or vCJD, the removal of the body from the ward, community setting, morgue or hospice, should be carried out using routine infection control measures. All bodies should be transported inside an

impervious body bag and, if CSF is leaking, a sealed body bag lined with absorbent materials should be used.

Archived

References

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