

Creutzfeldt-Jakob Disease (all types)

Reporting Obligations

All suspected and confirmed cases must be reported to the local Health Unit.

REPORTING FORM

Epidemiology

Aetiologic Agent:

Aetiology is not clear, but the infectious agents are thought to be unique proteins called prions which replicate by an unknown mechanism.

Clinical Presentation:

Creutzfeldt-Jakob Disease (CJD) is a prion disease and part of a group of rare, rapidly progressive, universally fatal neurodegenerative syndromes that are characterized by neuronal degeneration, spongiform change, gliosis, and accumulation of an abnormal protease-resistant amyloid protein or scrapie prion protein distributed throughout the grey matter structures of the brain.

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.

Classic CJD can be sporadic (sCJD), familial or iatrogenic. Variant CJD (vCJD) is another category described in 1996 and associated with ingesting meat from bovine spongiform encephalopathy (BSE) infected cattle.

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.

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. For vCJD cases related to exposure of BSE-infected cattle, incubation can be from 10 to 20 years. vCJD contracted via a blood transfusion has an incubation period from 6.6 to 8.5 years.

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.

Additional Resources

1. [Public Health Agency of Canada: Creutzfeldt-Jakob Disease](#)

References

1. Ministry of Health and Long-Term Care, *Infectious Diseases Protocol, 2017*.

Risk Factors/Susceptibility

Genetic differences in susceptibility, resembling those of autosomal dominant traits, have been shown to explain patterns of occurrence of the disease in families.

Diagnosis & Laboratory Testing

NOTE:

The CJD Surveillance System (phone 1-888-489-2999) provides support for pathological evaluation (autopsies and biopsies), CSF testing, and genetic testing. Sporadic Creutzfeldt-Jakob Disease (sCJD)

Confirmed case:

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

Probable case:

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

Suspect case:

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

Clinical Evidence:

Additional neurological manifestations include:

- Myoclonus
- Visual or cerebellar disturbances such as ataxia
- Pyramidal or extrapyramidal symptoms
- Akinetic mutism

Patient Information

PATIENT FACT SHEET

TESTING INFORMATION & REQUISITION

Treatment & Case Management

There is no specific treatment available. Infected persons as well as their family members should be excluded from donating blood, organs and other body tissues. Investigation of cases is in collaboration with Public Health Agency of Canada, PHO, and the PHD.