Canadian Creutzfeldt-Jakob Disease Surveillance System

What are prion diseases?

Prion diseases are rare, neurodegenerative and fatal brain disorders. They occur worldwide in both humans and animals. The prion protein in the brain does not cause disease in its normal form. However, for unknown reasons and very rarely, the prion protein can become abnormal. This causes brain damage and loss of function over time.

Human prion diseases

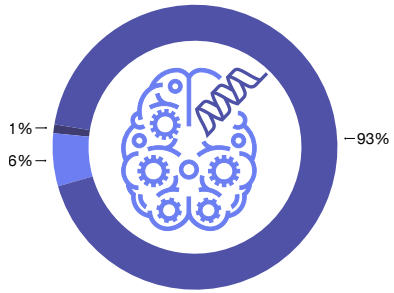
The most common human prion disease is Creutzfeldt-Jakob disease (CJD).

Animal prion diseases

Prion diseases that affect animals include:

- scrapie in sheep and goats
- chronic wasting disease in:
  - elk
  - deer
  - moose
  - reindeer
- bovine spongiform encephalopathy in cattle

CREUTZFELDT-JAKOB DISEASE SUBTYPES



- **Sporadic:** cause unknown.
- **Inherited:** passed down through genetics from previous generations.
- **Acquired:** extremely rare;
  - **Iatrogenic:** accidental infectious transmission during medical procedures involving human tissues.
  - **Variant:** human exposure to a prion disease of cattle.

Common signs and symptoms in humans

Unstable movement

Change in behaviour

Rapidly progressive dementia

In **2022, 90 people in Canada** were diagnosed with sporadic Creutzfeldt-Jakob disease.

- Around 90% of people diagnosed with Creutzfeldt-Jakob disease are aged **45 to 85**.
- **Most people with Creutzfeldt-Jakob disease pass away within 1 year** of showing symptoms.
- Every year, Creutzfeldt-Jakob disease is found in around **1 to 2 people per million** among the general population in Canada and worldwide.

Treatment

Medical care for people with Creutzfeldt-Jakob disease is limited to supportive therapy. There is no cure to slow down human prion disease progression.

Creutzfeldt-Jakob Disease Surveillance System offers

Lab & pathology testing

Coordination of case investigation

Public health research

Education

The Public Health Agency of Canada operates the Creutzfeldt-Jakob Disease Surveillance System with a focus to:

- improve rapid and accurate diagnosis
- better understand all types of human prion diseases in Canada
- protect the health of people in Canada by reducing risks of human prion disease transmission

Notifications of Suspect Cases of Creutzfeldt-Jakob disease

If you are a health care professional in Canada, you must inform your local public health authority. We strongly encourage you to notify the Creutzfeldt-Jakob Disease Surveillance System as well.

Telephone: 1-888-489-2999
Email: cjdsurveillance@phac-aspc.gc.ca
Website: Creutzfeldt-Jakob Disease Surveillance System

Pub.: 230551 Cat.: HP40-349/2023E-PDF ISBN: 978-0-660-68725-4

Public Health Agency of Canada | Agence de la santé publique du Canada

Canada