

CREUTZFELDT-JAKOB DISEASE

Infectious Disease Fact Sheet

What is Creutzfeldt-Jakob Disease?

Creutzfeldt-Jakob Disease (CJD) is a very rare and deadly brain disease. There are two main types:

- **Classic CJD:** Happens in about one person per million each year. In Canada approximately 40 to 90 people living with CJD die each year. Most cases are random and not passed down or caught from others. The disease most common in older adults and has been reported in every country around the world.
- **Variante CJD (vCJD):** This form of CJD is even more rare. It was first found in the UK in 1996 and is linked to eating beef from cows with mad cow disease (also called BSE – bovine spongiform encephalopathy). vCJD has been seen in younger people, with an average age of death around 28 years old. As of 2025, there have been fewer than 250 cases of vCJD worldwide, most of them in Europe.

For more information about BSE, see the Canadian Food Inspection Agency (CFIA) at www.inspection.gc.ca

What causes Creutzfeldt-Jakob Disease?

CJD is caused by an abnormal prion – a protein found on the surface of cells. The abnormal prion attaches to other brain cell proteins and bends them out of shape. This damages the brain by killing cells and creating gaps or sponge-like patches in tissue.

Once these abnormal CJD prions appear in a person, it can take up to 30 years before symptoms begin. The age at which symptoms start has been reported to range from 12 to 74 years. After symptoms begin, people live an average of one year. The average age of death is 29 years. vCJD affects males and females equally.

What are the symptoms of Creutzfeldt-Jakob Disease?

Early symptoms may include:

- Confusion
- Depression
- Forgetfulness
- Difficulty sleeping
- Behavior changes
- Impaired vision

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- Abnormal physical sensations
- Difficulty with voluntary coordination

Once a person begins showing signs or symptoms of CJD, the disease advances quickly over 2 to 12 months.

More advanced symptoms include:

- Balance problems
- Difficulty with speech and movement
- Increased risk of pneumonia
- Dementia
- Coma and eventual death

How does Creutzfeldt-Jakob Disease spread?

CJD is not contagious, and most cases, about 93 percent, occur without a known reason. Six percent of cases run in families. In very rare cases, less than one percent of the time, CJD is passed to a person by instruments or transplanted tissue used in eye, brain or spine surgery.

How is Creutzfeldt-Jakob Disease treated?

There is currently no cure for CJD. Treatment focuses on supportive care such as physical and occupational therapies. A person with CJD eventually becomes confined to bed and must be fed by a tube.

Psychological support may also help families of affected people. Genetic counselling is indicated in familial disease.

How can Creutzfeldt-Jakob Disease be prevented?

There is no vaccine for CJD and no protective immune response to infection has been demonstrated.