

F a c t s

CREUTZFELDT-JAKOB DISEASE (CJD)

What is CJD?

Creutzfeldt-Jakob disease (classical CJD) is a degenerative, fatal disease of the brain that occurs in humans. It is most common in adults 45-75 years old. It is rare with only about 30 cases per year in Canada, which is equal to 1 case for every million people. CJD is part of a group of diseases known as prion diseases. Prions are proteins found on the surface of cells. These diseases are also called Transmissible Spongiform Encephalopathies (TSEs). Prion diseases occur worldwide in both animals and humans.

What causes CJD?

CJD is caused by an abnormal prion. The abnormal prion attaches to other brain cell proteins and bends them out of shape. They attack the brain, killing cells and creating gaps or sponge-like patches in tissue. Once a person is exposed to the abnormal CJD prions, it can take up to thirty years before symptoms begin.

What are the symptoms?

Early symptoms include mood swings, memory loss, strange physical sensations, insomnia, depression, and confusion. Other symptoms may appear later and include rapidly progressive dementia as well as involuntary and irregular jerking movements.

How is it spread?

CJD occurs spontaneously 85-90% of the time, while 10-15% of the time it may run in families. CJD is passed onto a person by instruments or transplanted tissue used in eye, brain or spine surgery less than 1% of the time. CJD is not spread from person to person.

What is the incubation period?

The length of time between infection and the appearance of symptoms can be anywhere from 10-30 years. The length of time from symptom onset to death is usually one year; however, shorter duration periods of several months are common and longer duration periods of two or more years have been noted, usually in the familial form and with an earlier age of onset.

What is the treatment?

There is currently no cure for CJD. Treatment involves supporting the patient with physical and occupational therapies. A person with CJD eventually becomes dependent on others for care. CJD is inevitably fatal and the final diagnosis is made after death using a microscope to view brain cells.

—Health & Environment Facts —

Is CJD related to variant CJD (vCJD)?

CJD and vCJD are not the same disease. They are part of a family of diseases caused by abnormal prions. The symptoms are similar, although vCJD usually occurs before the age of 30. vCJD is sometimes called human BSE or mad cow disease. It is thought to be passed onto humans from eating cow parts infected with BSE prions.

Where can I get more information?

The following links provide more information on CJD as well as other related diseases:

World Health Organization

http://www.who.int/topics/creutzfeldtjakob_syndrome/en/

Public Health Agency of Canada

<http://www.phac-aspc.gc.ca/cjd-mcj/index.html>

For more information please call Public Health at 376-9420 or 1-800-263-3456.