



Creutzfeldt-Jakob Disease (CJD)

What is Creutzfeldt-Jakob Disease (CJD)?

Creutzfeldt-Jakob Disease or CJD is a rare, degenerative, fatal disease of the brain.

Early symptoms may include confusion, depression, forgetfulness, difficulty sleeping, behavior changes, impaired vision, abnormal physical sensations, and 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, and eventual death. CJD is very rare, but approximately 30 to 40 people living in Canada die from it each year. There have been 61 cases of CJD reported in British Columbia between 1997 and 2010. CJD can be found in every country around the world.

What causes CJD?

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. They attack the brain, killing cells and creating gaps in tissue or sponge-like patches. Once these abnormal CJD prions appear in a person, it can take up to 30 years before symptoms begin.

How does a person get CJD?

Up to 85 to 90 per cent of cases of CJD are spontaneous. Ten to 15 per cent of cases run in families. Less than 1 per cent of the time, CJD is passed to a person by instruments or transplanted tissue used in eye, brain or spine surgery. CJD is not contagious.

How do you test for CJD?

Tests for CJD are done on the fluid from around the spine but they are not always accurate. The diagnosis is made by a neurologist who looks at symptoms and brain images, using CT and MRI scans. It can be

difficult to detect gaps in tissues or sponge-like patches because they only show up at a late stage of the disease. The final diagnosis is made after death, using a microscope to view brain cells.

How do you treat CJD?

There is currently no cure for CJD. Treatment involves physical and occupational therapies. A person with CJD eventually becomes confined to bed and must be fed by a tube.

Is CJD related to variant CJD (vCJD)?

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

For more information on vCJD, see HealthLink BC File [#55b Variant Creutzfeldt-Jakob Disease\(vCJD\)](#). Visit also the Canadian Alzheimer Society webpage on Alzheimer Disease Related Dementia (CJD) at www.alzheimer.ca/en/About-dementia/Dementias/Creutzfeld-Jakob-Disease



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