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Creutzfeldt-Jakob Disease (CJD)

What is Creutzfeldt-Jakob Disease (CJD)?

Creutzfeldt-Jakob Disease (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, coma and eventual death. CJD is very rare, but approximately 30 to 40 people living in Canada die from it each year. 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?

Most cases of CJD, about 85 to 90 per cent, occur without a known reason. 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 is CJD diagnosed?

A tentative diagnosis of CJD is made based on:

- symptoms,
- tests on the fluid surrounding the spinal cord,
- brain images using CT and MRI scans,
- and recordings of the brain's electrical activity using EEG.

The diagnosis can only be confirmed through an autopsy.

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

For more information on vCJD, see HealthLinkBC File #55b Variant Creutzfeldt-Jakob Disease(vCJD).

For more information on CJD, visit the Canadian Alzheimer Society web page on Creutzfeldt-Jakob disease at https://alzheimer.ca/en/Home/About-dementia/Dementias/Creutzfeld-Jakob-Disease.

BC Centre for Disease Control
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