BCHealthFiles

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Creutzfeldt-Jakob Disease (CJD)

What is CJD?

Creutzfeldt-Jakob Disease or classical CJD is a rare, degenerative, fatal disease of the brain in humans. It is most common in adults 45 to 75 years old. Early symptoms include mood swings and memory loss. The disease advances quickly over 2 to 12 months and shows symptoms of balance problems, difficulty with speech and movement, and finally death.

How common is CJD?

CJD is very rare. In Canada there are about 30 cases each year. It occurs at a rate of about one case per million people. It 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 85-90 per cent of cases occur spontaneously, while 10-15 per cent of the time CJD may run in the family. Less than one per cent of the time, CJD is passed onto a person on instruments or transplanted tissue used in eye, brain or spine surgery. CJD is not contagious.

How do you test for CJD?

There are blood tests for CJD. However, these may not always be accurate. The diagnosis is made by a neurologist who looks at symptoms and brain images, using CT and MRI scans. Unfortunately, it can be difficult to detect gaps in tissues or sponge-like patches as these only show up late in 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 supporting the patient with physical and occupational therapies. A person with CJD eventually becomes confined to bed and must be fed by a tube.

Is it 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. The 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.

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

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