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Variant Creutzfeldt-Jakob disease (vCJD)

What is variant Creutzfeldt-Jakob Disease (vCJD)?

Variant Creutzfeldt-Jakob Disease (vCJD) is also called human mad cow disease or human Bovine Spongiform Encephalopathy (BSE). It is a very rare, degenerative and fatal brain disease that can occur in people. The disease damages brain cells and the spinal cord.

What are the symptoms of vCJD?

Early symptoms of vCJD include mood swings and memory loss. The disease also causes problems with movement and advances quickly to a vegetative state and death. It may take anywhere from 5 to15 years for symptoms of vCJD to develop from the time of being infected. vCJD can occur at any age, but the disease is most common in people under 30 years of age.

Since the discovery of vCJD in 1995, there have been more than 200 cases of vCJD reported worldwide. Most cases have occurred in the United Kingdom (UK). A few cases of vCJD have been reported in Canada but these people were infected in other countries.

What causes vCJD?

vCJD 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. The disease is caused when an abnormal prion attacks the brain, killing cells and creating gaps in tissue or sponge-like patches. The vCJD prion is the same prion found in cows with BSE.

How does a person get vCJD?

vCJD is not passed from person to person. However, it can be passed from someone with vCJD to others through blood transfusions, though this is extremely rare. It can also be passed onto people who eat BSE-infected cow parts or beef. In Canada to date, there have been 19 reported cases of BSE in cows. No part of these cattle has entered the human food supply. None of the reported cases of vCJD resulted from eating Canadian beef.

How does a cow get BSE?

The abnormal prions that cause BSE collect in the brain, skull, spine, nerve tissue and gut lining of cows. The prions can be passed to other cows when their feed contains contaminated material from infected cows. For more information on BSE, visit the Canadian Food Inspection Agency at https://inspection.canada.ca/en/animal-health/terrestrial-animals/diseases/reportable/bovine-spongiform-encephalopathy.

What precautions are taken to avoid the spread of BSE?

To prevent the spread of BSE, materials that are likely to contribute to the spread of BSE are not allowed to enter animal and human food chains.

For more information, visit Overview of Canada's BSE Safeguards at https://inspection.canada.ca/en/animal-health/terrestrial-animals/diseases/reportable/bovine-spongiform-encephalopathy/safeguards.

How is vCJD diagnosed?

A tentative diagnosis of vCJD is made based on:

- Symptoms
- Tests on the fluid surrounding the spinal cord
- Brain images using MRI scans
- Recordings of the brain's electrical activity using EEG
- Tonsil biopsy

The diagnosis can only be confirmed through an autopsy.

In individuals with vCJD, life-threatening complications tend to develop approximately 2 years after initial symptoms occur.

How is vCJD treated?

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

Is vCJD related to CJD?

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 CJD usually occurs in adults between the ages of 45 and 75. CJD is not connected to eating BSE-infected cow parts.

For more information

For more information on CJD, please visit HealthLinkBC File #55a Creutzfeldt-Jakob disease (CJD).

BC Centre for Disease Control
Provincial Health Services Authority