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Variant Creutzfeldt-Jakob Disease (vCJD)

What is vCJD?

Variant Creutzfeldt-Jakob Disease, also called human Bovine Spongiform Encephalopathy (BSE) or human mad cow disease, was first discovered in the United Kingdom (UK) in 1996. It is a rare, degenerative and fatal brain disease that can occur in humans.

The disease damages brain cells and the spinal cord. Early symptoms include mood swings and memory loss. The disease also causes problems with movement, and advances quickly to a vegetative state and finally death.

It can occur at any age, but it is most commonly seen in people under 30 years old. It has a long incubation period of 5 to15 years.

How common is vCJD?

vCJD is extremely rare. In 2002, one case was diagnosed in Canada in a person who often travelled to the UK. Most cases have occurred in Britain. Since the discovery of vCJD in 1996 up to December 2003, there have been 153 cases worldwide. Between 1996 and March 2004, there have been 146 cases and 139 deaths in Britain.

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. These attack the brain, killing cells and creating gaps in tissue or sponge-like patches. The vCJD prion is the same prion found in cows with Bovine Spongiform Encephalopathy (BSE).

How does a person get vCJD?

The BSE prion may be passed onto humans who may eat infected cow parts. The BSE prion concentrates in the brain, skull, spine, nerve tissue and gut lining of cows. It is passed from cow to cow through feeding these waste materials back to the cattle. This process is now outlawed in most countries including Canada, the US, and Britain. Waste material is not fed to cows used for human consumption.

The BSE prion is not passed from human to human. Although human infection is extremely rare, vCJD is a devastating disease.

Precautionary efforts are being taken to prevent new human infections. Infected cows are usually more than 30 months old. An infected cow can be aggressive, usually moves in an uncoordinated fashion until it can no longer stand up, and often loses weight before dying 2 to 6 months later. In Britain and other parts of Europe, millions of cows have been killed to prevent the spread of BSE among the cattle.

In Canada, there have been two reported cases of BSE in cows. The first case was found in a beef cow in 1993 and was traced back to imported cattle from the UK. The second case was found in Alberta in May 2003. In both

cases, the herd and other cattle that came into contact with this cow were killed as soon as BSE was confirmed.

How do you test for vCJD?

There are blood tests for the abnormal prions, but they 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, the gaps in tissues or sponge-like patches 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 vCJD?

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 it related to classical CJD?

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 CJD usually occurs in adults between the ages of 45 and 75.

For more information

For more information on CJD, see BC HealthFile #55a Creutzfeldt-Jakob Disease (CJD).
For more information about Bovine Spongiform Encephalopathy (BSE), check the Web site for the Canadian Food Inspection Agency at www.inspection.gc.ca/english/toce.shtml

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