September 2011

Information sheet

Community and Health Services Department

York Region

Creutzfeldt-Jakob Disease (CJD)

What is Creutzfeldt-Jakob Disease?

CJD is a rare disease that affects the central nervous system. There are two types of CJD, classic and a new form, called variant CJD. Classic CJD is rare, occurring at a rate of approximately one case per million people annually. Variant CJD is even more rare and was first diagnosed in the United Kingdom in 1996. As of September 2008, there have been 206 definite or probable cases of variant CJD reported worldwide, mostly in European countries. It is linked to eating contaminated beef products from animals infected with bovine spongiform encephalopathy (BSE) or "mad-cow disease."

The age of onset of the disease in patients has been reported to range from 12 to 74 years of age, with patients living an average of one year after the onset of symptoms. The average age of death is 29 years. The incidence of variant CJD is the same in males and females.

The origin of BSE in cattle remains unknown, but the epidemic that occurred in the United Kingdom is believed to have resulted from feeding cattle meat-and-bone meal (rendered ruminant protein) containing the tissues of BSE-infected ruminants. For more information about BSE, see the Canadian Food Inspection Agency (CFIA) at www.inspection.gc.ca

What are the symptoms of Creutzfeldt-Jakob Disease?

In variant CJD, the period between exposure to the infection and the onset of symptoms is believed to be at least 10 to 15 years, but can be longer.

The symptoms of variant CJD differ somewhat from those of classical CJD. Variant CJD symptoms include early psychiatric symptoms such as anxiety, depression, withdrawal and behavioural changes. Persistent pain or odd sensations in the face or limbs often develop. The disease then progresses to include difficulties with motor skills, involuntary movements and mental deterioration, often ending in a persistent vegetative state before death.

Currently, the only way to diagnose variant CJD with certainty is by microscopic examination of the brain tissue, most often through autopsy. In variant CJD, the brain pathology is very characteristic and is different from classical CJD.

For further information, please call:
York Region *Health Connection* 1-800-361-5653
TTY 1-866-252-9933 or visit www.york.ca

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How does Creutzfeldt-Jakob Disease spread?

The potential for transmission of variant CJD between people is not well understood, and no cases of this kind of spread have been documented. Scientists do not believe that variant CJD can be transmitted through casual contact like touching or kissing, or even intimate (sexual) contact with a person with the disease. However, if the disease is transmitted in the same way as classical CJD, spread may theoretically be possible through contaminated medical devices and transplants. The tissues that are most infectious include the brain, spinal cord, pituitary gland and parts of the eye. In variant CJD, other tissues such as the tonsils and appendix have also been found to be infective, in contrast with classical CJD. There is extensive research around the world into the transmission of variant CJD and its detection and treatment.

How is Creutzfeldt-Jakob prevented?

There is no vaccine for CJD and no protective immune response to infection has been demonstrated.

What is the treatment for Creutzfeldt-Jakob?

No treatment had been shown in humans to slow or stop the progressive neurodegenerative syndromes of CJD. Experimental treatments are under study. Supportive therapy is necessary to manage dementia, spasticity, rigidity, and seizures arising during the course of the illness. Psychological support may help families of affected people. Genetic counselling is indicated in familial disease.

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