Creutzfeldt-Jakob Disease

Alzheimer Society

Other Dementias

Introduction

Alzheimer's disease is the most common of a large group of disorders known as "dementias." It is an irreversible disease of the brain in which the progressive degeneration of brain cells causes thinking ability and memory to deteriorate. Alzheimer's disease also affects behaviour, mood and emotions, and the ability to perform daily living activities.

Other forms of dementia resemble Alzheimer's disease in that they also involve a progressive degeneration of brain cells that is currently irreversible. They include the dementia associated with Vascular Dementia (the second most common dementia after Alzheimer's disease), Frontotemporal Dementia, Creutzfeldt-Jakob Disease (CJD), Lewy body Dementia, Huntington disease, and Parkinson's disease.

Sometimes a person may have different symptoms in the early stages of the disease, such as memory loss, behaviour changes, or difficulties with speech and movement. These symptoms may suggest a form of dementia other than Alzheimer's disease. In any event a person should always seek a thorough medical assessment if any of these symptoms are present.

Regardless of the type of dementia, individuals are encouraged to obtain information and support from the Alzheimer Society.

What is Creutzfeldt-Jakob Disease?

Creutzfeldt-Jakob Disease (CJD), often referred to as prion disease, is a rare, rapidly developing form of dementia caused by infectious proteins called prions. Prions are proteins that occur naturally in the brain and are normally harmless. However, prions cause disease when they take on a misshapen form, accumulate in the brain and kill off brain cells. Prion diseases affect both humans and animals.

Prion diseases started to attract public attention during the mid 1980s with the bovine spongiform encephalopathy (BSE) epidemic, a prion disease of cattle (also called “mad cow disease”). The best known prion disease in humans is Creutzfeldt-Jakob Disease. It affects about one to two persons in a million worldwide each year, with about 35 new cases being diagnosed in Canada every year.[1]

[1] Public Health Agency of Canada (2007). *CJD and human prion diseases*. [Accessed June 9, 2011] at: http://www.phac-aspc.gc.ca/hcai-iamss/pdf/cjd_prion_disease-eng.pdf

There are two types of CJD: classic CJD and variant CJD (vCJD).

Classic CJD

The three types of classic CJD include:

1) **Sporadic:** This type of CJD accounts for 90% of cases in Canada. It affects people between 45-75 years old. The cause is unknown and the disease appears without warning. Most persons with sporadic CJD die within one year.
2) **Genetic:** This type of CJD appears in families with an abnormal gene. It accounts for 7% of cases. Genetic testing can be done by a blood test or brain tissue examination after death. A person who has this abnormal gene has a 50% chance of passing it on to each of his or her children. Familial, Gerstmann-Straussler-Scheinker (GSS) and Fatal Familial Insomnia (FFI) are very rare forms of genetic CJD.
3) **Iatrogenic:** Very few people around the world got CJD from accidental transmission during a medical procedure such as: human pituitary hormone therapy, human dura-mater grafts, corneal grafts or instruments used in neurosurgery. Less than 1% of people with CJD have this type of the disease. There are now infection control guidelines and screening of tissue/organ donors in place to help prevent further transmission.

Variant CJD (vCJD)

VariantCJD affects younger people at an average age of 28 years. This form of CJD is related to eating beef infected with bovine spongiform encephalopathy (BSE) or, as it is commonly called, "mad cow disease". Also, variant CJD has been reported to be transmitted by a blood transfusion from a person with variant CJD.[2]

How does Creutzfeldt-Jakob Disease affect the person?

CJD can mimic many other dementias. However, it has a rapid onset and decline once symptoms appear. Depending on the type of CJD, the person may have memory problems, a lack of interest and not act like him or herself, difficulty with balance when walking, clumsiness,vision problems, muscle jerks or twitching, difficulty with speech and swallowing. People with CJD generally live less than 12 months after the signs and symptoms appear, although some people may live longer. Most people lapse into coma before death. Death is usually a result of complications such as heart failure, respiratory failure or pneumonia.

How is Creutzfeldt-Jakob Disease assessed?

Creutzfeldt-Jakob Disease is very difficult to diagnose, especially in the beginning of the disease. There is no test to confirm the diagnosis of CJD in a living person. The best way to confirm if a person has CJD is to examine brain tissue after death during an autopsy. However, doctors will do a detailed exam and many tests to help diagnose this disease.

[2] Brown, P. (2007). Creutzfeldt-Jakob Disease: reflections on the risk of blood product therapy. Haemophilia, 13, 5. p. 33-40. [Accessed June 9, 2011 at: http://onlinelibrary.wiley.com/doi/10.1111/j.1365-2516.2007.01572.x/pdf

The following steps may be taken:

- Detailed medical history: This will help the doctor learn when the person's signs and symptoms started, as the length of CJD is of a short duration.
- CT scan (computerized tomography) produces a picture of the brain. It can be used to diagnose other diseases, as well.
- MRI (magnetic resonance imaging): An MRI also produces a picture of the brain. It helps to distinguish sporadic CJD from variant CJD. It can also be used to find other diseases.
- EEG (electroencephalogram) measures the electrical activity of the brain. Sometimes, but not always, there is a specific pattern on the EEG that helps to diagnose CJD.
- Lumbar puncture: Fluid can be taken from the person's spine and examined to exclude other infections of the brain. One of the tests is called the 14-3-3 protein test. If the 14-3-3 is positive it means that there has been some brain cell death but not necessarily due to CJD. However, combined with other tests it helps in the diagnosis of probable CJD.
- Blood test: A blood test can detect a mutation in the person's genes and determine a genetic form of CJD.
- Brain autopsy: The best way to confirm CJD is by looking at brain tissue with a microscope after death. Brain autopsies are performed only in certain large hospitals in Canada. The Canadian CJD Surveillance System (see contact information at end of this sheet) can help make arrangements and pays for the brain autopsy if CJD is suspected and if the next of kin gives their consent.

What are the risk factors for Creutzfeldt-Jakob Disease?

The true risk of developing either form of CJD is largely unknown. At this time, there is no specific way to protect a person from getting sporadic or familial CJD.

However, certain factors may increase one's risk:

Pituitary hormone treatment:

If pituitary hormone treatment derived from human tissue was used prior to the genetically engineered form of the hormone becoming available in the 1980s, there is an increased risk. Since 1985, human growth hormones are made synthetically and, as a result, there is no longer any risk of transmission from this source.

Family history of CJD:

A few people have a genetic mutation that increases their likelihood of developing the disease. A blood test or brain tissue examination after death can be done to find out if a person has the mutation.

Contaminated surgical instruments:

A few people have been infected from contaminated instruments used during brain surgery. Today, instruments used on the brain of someone with possible CJD are destroyed.

CJD precautions:

These are infection control precautions required only for certain medical procedures involving specific tissue. When embalming the remains of a person who has died of a possible prion disease, the World Health Organization and the Public Health Agency of Canada recommend that funeral services workers use CJD precautions.

You cannot catch CJD by touching, feeding or taking care of a person with CJD at home. CJD is not a contagious disease transmitted by social or sexual contact, or by air.

Blood transfusions:

The Canadian Blood Services and Héma-Quebec screen for and exclude persons with CJD from giving blood.

Is there treatment?

At present, there is no known cure for CJD and no effective way to slow its progression. It is important to relieve pain, discomfort and other symptoms such as jerking movements and unsteadiness. Supportive nursing care is focused on keeping the person as comfortable as possible and helping the family members understand this rare and devastating disease.

New behavioural therapeutic strategies are also helping people living with the disease. Therapeutic techniques like physical activity and music are being used as viable and useful treatments. Research shows that the quality of life of people with dementia, and also their caregivers, is significantly improved by activities that emphasize their strengths and abilities. By understanding the person's personality, life experiences, support systems and ways of coping, a person-centred approach to care can be created that preserves and improves quality of life.

For more information:

The Public Health Agency of Canada's Creutzfeldt-Jakob Disease Surveillance System (CJDSS) study team. The Public Health Agency of Canada CJD Surveillance System conducts active monitoring of CJD in Canada. Their main purpose is to study human prion diseases in Canada and to protect public health by reducing the risk of prion disease transmission.

More information about CJD and the CJD Surveillance System can be found on the website: www.phac-aspc.gc.ca/hcai-iamss/cid-mci/cidss-eng.php or call: toll free 1-888-489-2999

Alzheimer Society

Alzheimer Society of Canada
20 Eglinton Avenue West, Suite 1600, Toronto, Ontario, M4R 1K8
Tel: 416-488-8772 • 1-800-616-8816 • Fax: 416-488-3778
E-mail: info@alzheimer.ca • Website: www.alzheimer.ca

R300-05E