

Related Dementia - Creutzfeldt-Jakob Disease

Introduction

Dementia is a syndrome consisting of a number of symptoms that include loss of memory, judgment and reasoning, and changes in mood, behaviour and communication abilities. These symptoms may affect a person's ability to function at work, in social relationships or in day-to-day activities. Sometimes symptoms of dementia can be caused by conditions that may be treatable, such as depression, thyroid disease, infections or drug interactions. However, if the symptoms are not treatable and progress over time, they may be due to damage to the nerve cells in the brain.

Alzheimer's disease is the most common form of dementia. It is a disease of the brain, characterized by deterioration of thinking ability and of memory, caused by the progressive death of brain cells. Alzheimer's disease accounts for approximately 64% of all dementias in Canada. The features of Alzheimer's disease include a gradual onset and continuing decline of memory, as well as changes in judgment or reasoning, and ability to perform familiar tasks. Other dementias include Vascular Dementia, Lewy body Dementia, Frontotemporal Dementia and Creutzfeldt-Jakob Disease.

Sometimes a person may have symptoms such as sudden onset of memory loss, behaviour changes, or difficulties with speech and movement. These symptoms may suggest a dementia other than Alzheimer's disease. Creutzfeldt-Jakob Disease is one of these dementias. A person should 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) is a rare, rapid and fatal form of dementia caused by infectious agents called prions. Prions are proteins that occur naturally in the brain and are normally harmless. When they are misshapen, however, they can cause devastating illness, attacking the brain, killing cells and creating gaps in tissue. There are two types of CJD: classical and variant. Classical CJD (also called sporadic CJD) occurs at random. Variant CJD (vCJD) is a disease linked to eating beef products from cattle infected with bovine spongiform encephalopathy (BSE) or "Mad Cow" disease. It attacks the central nervous system and is fatal if illness develops. Both types of CJD typically cause memory loss and behavioural changes. The rate of CJD is approximately one per million people per year worldwide.¹ The Canadian CJD surveillance system, considered one of the best in the world, involves national reporting through a toll-free line of all suspected cases of both classical and vCJD in Canada. As of May, 2007, there were 39 new cases of suspected CJD reported in Canada.² There has been one case of vCJD in Canada. However, this was related to this person's time living in the United Kingdom. There have been no cases linked to eating Canadian beef.³

How does Creutzfeldt-Jakob Disease affect the person?

CJD usually has rapid onset and decline once signs and symptoms appear. A number of subtypes of

1. Public Health Agency of Canada: www.phac-aspc.gc.ca/cjd-mcj/vcjd-faq_e.html
2. Ibid: www.phac-aspc.gc.ca/hcai-iamss/cjd-mcj/cjdss-ssmcj/stats_e.html#canada
3. Public Health Agency of Canada: www.phac-aspc.gc.ca/cjd-mcj/vcjd-faq_e.html

CJD exist, many with slight variations in signs and symptoms. The various forms are more alike than they are different, and everyone affected by CJD will have serious mental and physical problems. Classical and variant CJD share the following characteristics but they can present somewhat differently in each form of the disease.

Long incubation period

The incubation period is the time it takes a person to become sick after having contracted a disease. It often takes years or even decades after infection before someone with CJD develops signs and symptoms. Classical CJD has an average age onset of 60 whereas vCJD tends to occur in people with an average age under 30.

Personality changes

Anxiety, depression, memory loss and impaired thinking are often the first personality changes to be noted. As the diseases progress, people with both forms of CJD develop dementia. CJD usually progresses much more rapidly than other types of dementias.

Problems with balance and co-ordination

Both types of CJD affect balance and co-ordination, leading to stumbling, falls and difficulty walking. These problems typically occur earlier in vCJD than in classical CJD.

Coma and death

Most people lapse into coma before death. People with classical CJD generally live less than 12 months after the signs and symptoms appear, although some people may live as long as two years. Death is usually a result of complications such as heart failure, respiratory failure or pneumonia. People with vCJD tend to live slightly longer – about 12 to 14 months after signs and symptoms appear.

How is Creutzfeldt-Jakob Disease assessed?

Only a brain biopsy or an examination of brain tissue after death (autopsy) can definitely confirm CJD. However, doctors often use a detailed medical and personal history, a neurological exam and a variety of tests to diagnose this disease.

What are the risk factors for Creutzfeldt-Jakob Disease?

The true risk of developing either form of CJD is largely unknown. However, certain factors may increase one's risk:

Family or genetic history of CJD: a few people have a genetic mutation that increases their likelihood of developing the disease.

Human growth hormone injections: if a HGH was used prior to the genetically engineered form of the hormone becoming available in the 1980's, there is an increased risk.

Certain medical procedures: a small number of people worldwide have developed CJD from infected tissue used in cornea and skin transplants and from grafts used during brain or spinal cord surgery.

Contaminated surgical instruments: a few people have been infected from contaminated instruments used during brain surgery.

Blood transfusions: risk may increase if one received a blood transmission or used insulin derived from cow products after 1980 in a country with a high incidence of BSE such as Great Britain, Portugal, Spain, France or Germany.

Eating beef from certain countries: eating beef from countries with a relatively high rate of BSE may increase risk.

To help reduce the risk of contracting CJD from infected beef, consider the following options: be selective when eating beef in parts of the world where less stringent safeguards may exist; avoid eating the highest risk parts of cattle, such as eyes, brain, spinal cord and intestines; choose organic beef where cows have been fed on grains and grasses rather than on feed that contains parts of other animals. In order to reduce the risk of transmission, do not donate blood if you are at risk of exposure to CJD.

Is there treatment?

At present, there is no known cure for CJD and no effective way to slow its progression. Doctors focus on relieving pain and other symptoms such as jerking movements and unsteadiness.

For more information?

- The Creutzfeldt-Jakob Disease Surveillance System in Canada Study Team: 1-888-489-2999 or Web site: www.phac-aspc.gc.ca/hcai-iamss/cjd-mcj/index.html.
- Canadian Food Inspection Agency, BSE Fact Sheet or Web site: www.inspection.gc.ca/english/anim/hc/hasan/disemala/bseesb/bseesbfse.shtml.
- CJD Support Network (UK): www.cjdsupport.net.
- European Collaborative Study Group of CJD Web site: www.eurocjd.ed.ac.uk.
- Mayo Clinic: <http://www.mayoclinic.com/health/creutzfeldt-jakob-disease/DS00531/DSECTION=1>

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