7.3 Creutzfeldt - Jakob disease

Case Definition

Case definitions for Creutzfeldt - Jakob disease (CJD) are available at the following website:

http://www.phac-aspc.gc.ca/publicat/ccdr-rmtc/09vol35/35s2/MCJ_vMCJ-eng.php

Creutzfeldt-Jakob Disease (CJD) is one of the forms of human prion diseases. Prion diseases are rare, fatal, degenerative brain disorders that occur worldwide in both humans and animals. They belong to a group of brain diseases called proteinopathies.

The brains of people or animals with prion disease undergo damage, called "spongiform change" or "spongiosis" because when the tissue is examined under a microscope it resembles a sponge, with many tiny holes. In addition, the brain tissue contains abnormal deposits of a specific protein called the prion protein (PrP). These pathological changes can be caused by genetic variations, or apparently arise spontaneously within a single individual. However, they can also be caused by infectious transmission between individuals of the same or different species.

CJD has been defined as either classic CJD or variant CJD (vCJD).

- Classic CJD includes sporadic CJD, genetic CJD and iatrogenic CJD:
  - Sporadic CJD occurs worldwide and is of unknown etiology
  - Genetic (familial or inherited) CJD is linked to mutations of the prion protein gene
  - Iatrogenic CJD is related to transmission from person to person in the course of medical treatment such as being exposed to contaminated neurosurgical equipment
- Variant CJD is a novel form of CJD which has been linked to transmission of bovine spongiform encephalopathy (BSE) from cattle to humans.

The Canadian Creutzfeldt-Jakob Disease Surveillance System (CJDSS) is operated by the Public Health Agency of Canada and conducts prospective national surveillance for all types of human prion disease in Canada. The main purposes of the CJDSS are to better understand the epidemiology of human prion diseases, to improve the options available for their rapid and accurate diagnosis, and ultimately to protect the health of Canadians by reducing risks of prion disease transmission.

Please note that all human prion diseases are provincially reportable and nationally notifiable in Canada.

For more information regarding the Canadian Creutzfeldt-Jakob Disease Surveillance System, please call toll-free: **1-888-489-2999.**

Creutzfeldt - Jakob disease is extremely rare in Newfoundland and Labrador, with one case in a million or one case every two years expected. Due to the complexity of this

condition healthcare professionals are referred to the PHAC's web site for further information. This is available at:
http://www.phac-aspc.gc.ca/hcai-iamss/cjd-mcj/index-eng.php

A fact sheet on CJD is available in Appendix A and more information at:
http://www.phac-aspc.gc.ca/cjd-mcj/vcjd-faq-eng.php

Infection Control Guidelines are available at:
http://www.phac-aspc.gc.ca/hcai-iamss/cjd-mcj/pub-eng.php

An additional source of information is available at:
http://www.health.alberta.ca/documents/Guidelines-Creutzfeldt-Jakob-Disease-2013.pdf

Reporting Requirements and Procedures

Most physicians are aware of CJD, although because the disease is so rare many have never directly observed a case. A prompt referral to a neurologist should follow reporting of any suspicious pattern of symptoms, where a number of investigations must be carried out.

- Since 1998 the PHAC of Canada has had an intensive active surveillance program for CJD. It relies on the direct reporting of all confirmed, probable and possible cases of CJD by all neurologists, neurosurgeons, neuropathologists, geriatricians and infection disease physicians. The surveillance system operates on a reference-services model by offering comprehensive support to referring physicians for laboratory investigations, clinical consultation and education. For more information regarding the Canadian Creutzfeldt - Jakob disease Surveillance System please contact the clinical coordinator 1-888-489-2999.

- The laboratory (hospital or public health laboratories) reports case/s to the attending physician, the Chief Medical Officer of Health and the Medical Officers of Health (MOH)
- MOH office will notify, as required, local physicians, nurse practitioners, environmental health officers, community health nurses, communicable disease control nurses (CDCNs) and Infection control practitioners (ICP), in the particular region as required for follow-up and case investigation
- The Canadian Blood Services screens donors for potential exposure to variant CJD in the donor pre-screening process
- The CDCN in collaboration with the ICP (if necessary) will collect case details
- The CDCN will enter the case details into the electronic reporting system and utilize the Canadian Network for Public Health Intelligence (CNPHI) tool, if indicated, for alerts or outbreak summaries

Provincial Disease Control

- Reports the aggregate case data to Public Health Agency of Canada
- Provides an analysis of the case/s with reports in the Quarterly Communicable Disease Report (CDR), also posted on the Public Health website http://www.health.gov.nl.ca/health/publichealth/cdc/informationandsurveillance.html
- Coordinates the response if an outbreak across RHAs.

Appendix A: Creutzfeldt-Jakob Disease (CJD) Fact Sheet

Frequently asked questions about CJD

Public Health Agency of Canada

1. **Can you catch CJD from someone?**
   CJD and other human prion diseases are not believed to spread by close or casual person-to-person contact or by the airborne/respiratory route. However, transmission can occur during invasive medical interventions. It is sensible for anyone who might be exposed to the blood of another person to wear gloves.

2. **How can we be sure that the diagnosis of CJD is the correct one?**
   It should be emphasized that a definite diagnosis of any form of CJD can only be given by brain tissue examination after death. Each individual case of CJD can be assigned to one of three subtypes: sporadic, genetic or acquired. The considerations for diagnosis vary depending on the subtype. In genetic prion diseases, the diagnosis depends on development of particular neurological symptoms and the identification of a PRNP gene mutation by genetic analysis. Iatrogenic CJD is diagnosed on the basis of a confirmed diagnosis of CJD in someone who had a relevant medical exposure. Variant CJD is diagnosed by distinctive features seen on post-mortem examination of the brain.

3. **Is the blood supply safe from CJD?**
   Since donors cannot, at present, be tested for early biological indicators of CJD, nor can blood donations be tested for the removal of the prion agent after processing, and considering vCJD has been shown to be transmissible through blood transfusion, the Canadian Blood Services has developed policies with regards to CJD and blood donation. Canadian Blood Services deferral policies are available by contacting Canadian Blood Services at 1-888-2donate.

4. **Is there a risk in contracting CJD from organ transplant surgery?**
   The risk of contracting CJD from organ transplants is uncertain, but believed to be small. Unfortunately, a transplant usually has to be done before a full post-mortem examination of the donor can be completed, so this risk cannot be completely eliminated. However, if a potential donor is suspected of having CJD their tissues and organs would not be used for transplantation. Note also that there is a risk of infection in any transplant.

5. **Is the person with CJD in pain?**
   Neurological examination and the EEG of people in the later stages of CJD indicate that they lose awareness of their condition as the disease progresses. In the early stages, however, patients with CJD can develop marked fear, which can be very distressing and is probably associated with visual hallucinations. They may feel discomfort and some of the symptoms of the disease - such as myoclonus, sudden jerking of the limbs - are distressing for caregivers to witness. There are medications which can relieve the symptoms and make the person more comfortable. In vCJD dysesthesia, an unpleasant abnormal sensations to normal stimuli, has been described.

6. **Is a post-mortem examination necessary in CJD?**
   Post-mortem examination is not compulsory when CJD is suspected - the doctor requires the permission of the next of kin. However, because it is the only way, at the moment, to definitively diagnose CJD, this knowledge is often very helpful for families. The autopsy findings and any donated tissues will also be very beneficial to support research into the disease.

7. **Will there be many more cases of variant CJD?**
   As of April 3, 2007, there were 202 cases of vCJD worldwide. If the disease comes from exposure to infected beef products prior to the ban on specified offal in human food in 1989, as is now widely accepted, then there could be more cases if the incubation period is very long. However, without knowing the exact circumstances of infection, or who is most at risk and why, it is currently impossible to predict how many more cases of vCJD there will be.

8. **What is being done to protect us from CJD?**
   At present there is no specific way of protecting people from developing sporadic or familial CJD. Destroying surgical instruments that have been used on certain tissues of people with CJD and not using their organs for transplant guards against iatrogenic CJD. There have also been recent measures taken by the Canadian Blood Services for safeguarding the blood supply from variant CJD.