WINTER 2011

CJD IN CANADA
FAMILY EDITION

IN THIS ISSUE

Mission Statement

The mission of the Prion Diseases Program of the Public Health Agency of Canada is to continually assess, mitigate, and ultimately eliminate risks to human health posed by infectious prion diseases in Canada, through surveillance, laboratory services, research and education.

INTRODUCTION

The **Canadian Creutzfeldt-Jakob disease Surveillance System** (CJDSS) was established in 1998 as a national surveillance system for Creutzfeldt-Jakob disease (CJD). The main role of the CJDSS, which is maintained by the Public Health Agency of Canada, is to help protect Canadians from health risks posed by CJD.

The CJDSS also works more directly to support patients, families and healthcare providers dealing with this difficult disease through education and sharing of information. The *CJD in Canada* newsletter is one way in which this is done.

This issue of the newsletter provides an update on the number of CJD cases identified in Canada, a feature article on blood transfusions and prion diseases, and a "You Asked Us" section where we strive to provide you with answers to sometimes-difficult questions. It is our hope that this newsletter provides you with helpful information about CJD in Canada.

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CJD UPDATE

From 1998 to August 1, 2010, the CJDSS has investigated 1082 patient referrals. Of the 1082 referrals, 482 cases of CJD were confirmed by pathology or diagnosed on a clinical basis.

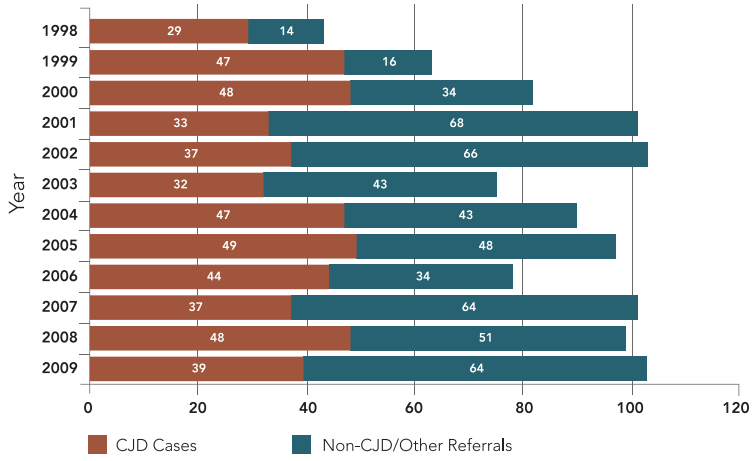
The only way to confirm a diagnosis of CJD is through a brain autopsy. Of all cases of CJD identified by the CJDSS to date, 84.1% have been confirmed in this way. This is one of the highest percentages reached by any country that carries out national CJD surveillance. This is because people (including physicians) remain aware of the disease, and because Canada maintains high-quality laboratory testing services.

The CJDSS uses rules set by the World Health Organization (WHO) when deciding a diagnosis of CJD. As shown in the graph, case referrals to the CJDSS have been relatively steady over time. Each year since 1999 about 80-100 patients have been referred to the CJDSS. From these referrals the CJDSS has recorded an average of 37.3 Canadian CJD cases per year from 1999 to 2009, which means an average of 1.17 cases per million people per year. This rate is consistent with worldwide rates of occurrence.

The success of CJDSS surveillance activities also depends on support and cooperation from physicians and other healthcare professionals caring for patients with CJD, as well as those affected by this disease. The CJDSS will continue to help physicians and other healthcare professionals to identify and understand all suspected cases of CJD in Canada, and to help affected families to better understand the disease. For more information please visit the CJDSS website at http://www.nml-lnm.gc.ca/cjd-mcj/index-eng.htm or contact our toll-free number: 1-888-489-2999.

Cases & Referrals to CJDSS by Year of Reporting

Note: 2008 and 2009 figures are provisional as information may be incomplete. For CJD incidence by year of death, please refer to our website.



The graph above shows the total number of CJD patients (brown bars) and the total number of patients that did not have CJD (blue bars).

BLOOD TRANSFUSION AND PRION DISEASES (CJD)

By Dr. Mindy Goldman, Executive Medical Director, Donor & Transplantation Services, Canadian Blood Services

The issue of blood transfusion and CJD is of the utmost importance. In our feature article, Dr. Goldman explains how this issue affects families who have been touched by CJD.

Because of prion diseases (CJD), some people are not allowed to give blood. There have not been any cases of CJD from blood transfusion in Canada. However, because there is no test to detect this disease in blood donors, all donors must answer a list of questions about risks for CJD before giving blood. There are two main kinds of CJD (classic and variant). These two diseases are treated differently because they have different risks for blood transfusion.

Classic CJD

Classic CJD usually arises without warning and for no specific reason. No cases of classic CJD transmission have been linked to a blood transfusion. However, this disease can also be caused by genetic changes or, very rarely, by infection during certain medical procedures. Potential blood donors are therefore screened out (deferred) if any close relatives (parent, child, brother or sister) have had CJD.

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Potential blood donors are also screened out if they have received dura mater (a material used in some kinds of surgery) or treatment with human pituitary hormones, since these treatments have been linked with classic CJD.

Variant CJD

Variant CJD is a human prion disease caused by infection with a prion disease of cattle (BSE, sometimes called "mad cow disease"). Between 1996 and 2010, approximately 220 people have been found to have variant CJD, mainly in the United Kingdom and Europe. Four of these people, all in the United Kingdom, are believed to have been infected by blood transfusion. Potential blood donors are therefore screened out if they have spent enough time in countries that have or may have BSE, or if they had a transfusion in one of these countries. This period of time is a total of three months or more in the United Kingdom or France from January 1, 1980 to December 31, 1996, or five years or more in the rest of Europe since January 1, 1980. Deferral policies are based on the magnitude of exposure to BSE in various countries.

For more information about Canadian Blood Services and our donor criteria, visit our website at www.blood.ca. For more information about Héma-Québec, visit the Héma-Québec website at www.hema-quebec.qc.ca.

YOU ASKED US

This section gives us a chance to answer questions you may have when dealing with CJD. The CJDSS also invites you to contact us directly if you have any questions, comments or concerns - toll free at 1-888-489-2999 or via email at CJDSS@phac-aspc.gc.ca.

Q. I've recently been caring for someone with CJD. Am I at risk of catching this disease?

A. No, physicians and scientists who study CJD do not believe you can catch it by caring for someone with the disease. You cannot catch the disease by social or sexual contact either. However, special care is taken when contact with the person's tissues is possible, such as during specific kinds of surgery, when handling samples in laboratories, and during an autopsy. Funeral service workers must also take special care when handling a person's remains.

Q. Why does a nurse from the CJDSS need to speak with me?

A. The CJDSS employs nurse investigators across Canada to visit and interview families affected by CJD. Families are always asked for their consent before the actual interview and chart review take place. Interviews are conducted because we need to be vigilant in identifying potential public health risks. During this interview, the nurse completes a questionnaire to collect information about a patient's medical history and possible risks. The nurse also collects medical information by reviewing the CJD patient's medical charts.

Q. Why is an autopsy required?

A. Firstly, post-mortem examination is not compulsory when CJD is suspected - the doctor always 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. During the autopsy, only the brain is removed and examined because prions (which are believed to be the cause of CJD) mostly accumulate here. The procedure takes place in a facility that has specialized equipment and staff are trained to ensure respect for the patient is always maintained and safety standards are followed.

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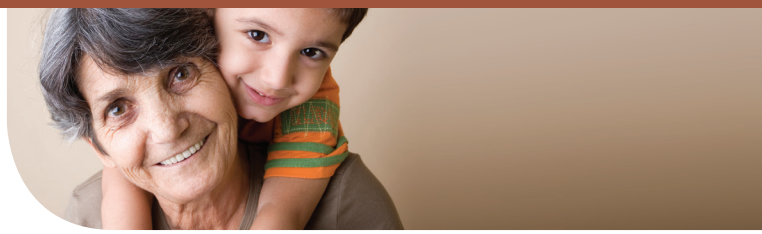

CONSENT FORM FOR DONATION OF BIOLOGICAL MATERIALS

Continued research is the key to finding answers to the many questions around CJD. This research may lead to improved diagnostic tests, better understanding of the causes of CJD, and better ways to deal with public health risks. Unused portions of the specimens of blood, brain tissue and spinal fluid that are received by the CJDSS for laboratory testing can also help us make new discoveries and further advance our knowledge of this difficult disease.

All participants or their representatives are asked whether or not these specimens may be used in future research, and to sign the Donation of Biological Materials for Research consent form. Please call 1-888-489-2999 if you have not completed this form but wish to do so, or would like more information. The CJDSS is very grateful for your participation.

EMAIL SUBSCRIPTION

Attention Readers:

This issue will be the final one that is automatically sent out by mail to all of our readers. If you wish to continue receiving our newsletter, you may do one of the following:

- Send us an e-mail at: CJDSS@phac-aspc.gc.ca. Please tell us whether you prefer to receive future issues by regular mail or e-mail.
- Read and download future newsletters online from: http://www.nml-lnm.gc.ca/cjd-mcj/index-eng.htm
- If you do not have access to the Internet, please call 1-888-489-2999. If you provide us with your mailing address, a member of our team will ensure you continue to receive a copy of our newsletter by mail.

CONTACT US

What would you like to see in the next newsletter? Was this newsletter helpful? Please let us know your thoughts or submit questions by contacting us at:

Toll free: 1-888-489-2999
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