Creutzfeldt-Jakob Disease
Surveillance System in Canada

Health Canada Santé Canada

Update: CJD in Canada

May 2003

Highlights

- *CJD-SS funding extended*
- *MRI-Useful tool in CJD diagnosis*
- *14-3-3 testing continues to play a crucial role in CJD diagnosis investigation and surveillance*
- *CJD-SS progress report*
- *Alzheimer's Society to provide support for CJD affected families*

For further Information
1-888-489-2999

Website
http://www.hc-sc.gc.ca/pphb-dgspsp/hcai-iamss/cjd-mcj/index.html

FUNDING EXTENDED

We are pleased to inform you that our funding has been extended. Please continue reporting any possible / probable cases of CJD to the CJD-SS by calling our toll free number:

1-888-489-2999

MRI very useful in CJD diagnosis

MRI changes have been described in patients with CJD in a number of recently published articles. In the case of sporadic CJD a hyperintensity of the putamen and caudate nuclei is frequently seen. In variant CJD a highly characteristic finding of bilateral hyperintensity of the pulvinar nuclei of the thalamus is described in the majority of patients. Because of these identified changes, MRI imaging (DWI, T2-weighted or FLAIR) is now considered an important non-invasive test for the diagnosis of both classical and variant CJD. See References on page 2.

14-3-3 Testing

Western-blot assays of 14-3-3 protein content of cerebrospinal fluid continue to play a crucial role in both diagnostic investigation and surveillance of Creutzfeldt-Jakob disease. Recent research investigations have shown that this family of proteins is markedly heterogeneous, consisting of at least seven genetically distinct isoforms. Research and international surveillance experience have also indicated that one specific member of this set of isoforms (gamma) is optimally informative for both medical and public-health purposes. **Health Canada is presently in the process of upgrading the previously offered version of the 14-3-3 western blot assay to a new version based on specific detection of the gamma isoform and plans to offer this testing service in the near future, once laboratory validation is complete.**

CJD-SS Progress Report

Objectives of CJD-SS

1. To gather information for all cases of Creutzfeldt Jakob Disease (CJD) identified to the Surveillance System in order to establish a database for research on CJD in Canada.
2. To participate in national and international studies to determine the epidemiology of CJD including variant CJD and to detect and characterize any cases occurring in Canada.

Methods

In April 1998, all Canadian physicians involved in the care of possible/probable cases of CJD were informed about our project and were asked to notify the CJD-SS, at a toll free number provided, to assist the Surveillance System in performing thorough case follow up. This involves ensuring the collection of a blood sample from the patient for genetic analysis, organizing an autopsy at time of death and obtaining consent from the patient's family to conduct a medical records review as well as an in depth interview. CJD-SS is involved with the European research community as an active participant within the EuroCJD group. This group usually meets once every 6 months to share data, information and knowledge and to further worldwide CJD research. Contact with Canadian physicians and patient's families is maintained by way of a yearly newsletter in which new information concerning CJD is disseminated.

Data and results obtained thus far

As of April 1, 2003 there have been 410 referrals to the Surveillance System: 44.6 % (183) of referrals are definite/probable CJD cases as defined by World Health Organization (WHO) criteria. Of these cases there have been 163 sporadic, 16 genetic (familial/GSS), 3 iatrogenic and one variant CJD. The annual incidence of CJD in Canada is as follows: for 1999, 1.02; for 2000, 1.14 and for 2001, 0.96 cases per million population.

The first case of variant CJD in Canada was diagnosed in August 2002 in a patient under the age of 50, who had multiple stays in the United Kingdom during the outbreak of bovine spongiform encephalopathy (BSE).

Conclusions

With the invaluable cooperation of both the physicians who care for patients with a CJD diagnosis and their families, CJD-SS has been able to detect, since 1999, CJD cases at a rate that reflects the expected worldwide incidence of CJD: approx. 1 case per million population per year. We have also identified the first case of variant CJD in Canada, carried out further study on familial cases of CJD as well as contributed to the ongoing worldwide research on CJD and variant CJD.

MRI References

Please see references below for more details or refer to our website links:

- DA Collie, RJ Sellar, M Zeidler, ACF Colchester, R Knight, R G Will. MRI of Creutzfeldt-Jakob Disease: Imaging features and recommended MRI protocol. Clin Radiol 2001; 56: 726-39.
- Schroter A, Zerr I, Henkel K, Tschampa HJ, Finkenstaedt M, Poser S. Magnetic resonance imaging in the clinical diagnosis of Creutzfeldt-Jakob disease. Arch Neurol 2000; 57: 1751-7.
- Taber KH, Cortelli P, Staffen W, Hurley RA. The expanding role of imaging in prion disease. Clin Neurosci 2002; 14: 371-6.

Support for families affected by CJD

The Alzheimer Society has agreed to assist in organizing support for families affected by CJD. Contact your local Alzheimer Society chapter for more information. A link to a list of these contact numbers can be found on our website.