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Creutzfeldt–Jakob disease: the Canadian situation

Creutzfeldt–Jakob disease (CJD) is a rare neurodegenerative disorder with a clinical presentation of dementia, myoclonus and progressive motor dysfunction leading to death, usually within a year of symptom onset. The clinical and pathologic picture varies, and there is no definitive clinical diagnostic test aside from neuropathologic investigation.[1]

CJD belongs to a group of diseases, termed transmissible spongiform encephalopathies, whose cause is not fully understood but is believed to be related to misfolding of an endogenous cellular protein ("prion protein," or PrP^{C}) into a pathogenic conformation (PrP^{Sc}).[2]

Three epidemiologic types of CJD are recognized. Sporadic CJD, which accounts for almost 90% of cases, may result from spontaneous somatic mutation of a prion protein gene within the host. Familial disease results from inheritance of a defective form of the prion protein gene and accounts for 5% to 10% of cases. Iatrogenic or infectious CJD accounts for less than 1% of cases. Transmission has occurred via donated tissue (corneal transplants and dura mater grafts), cadaveric pituitary gland extract (human growth hormone and gonadotrophin) and contaminated neurosurgical instruments.[3–5] Recent findings in the UK indicate that a new variant of CJD, vCJD, characterized by a young age of onset and a relatively long clinical course, is linked to the consumption of beef contaminated with the causative agent of bovine spongiform encephalopathy (BSE).[6–8]

Worldwide, the incidence of CJD is about 1 per million population per year. Most cases occur in people in their sixties. There is little or no sex bias, temporal clustering or geographic variation in incidence, although familial-type disease can show local ethnic and familial clustering.[3]

In Canada, 390 deaths attributed to CJD were recorded from 1979 to 1995. The number of deaths ranged from 14 to 34 per year, with a 1:1 male-to-female ratio. Forty-three percent of the deaths occurred among people in their sixties, which corresponds with the peak age of onset for sporadic CJD.[1–3] The crude annual mortality rates for the period 1979 to 1995 ranged from 0.6 to 1.1 cases per million population; this is consistent with estimates of incidence worldwide.

Most CJD in Canada is probably sporadic, although familial disease has been reported. No cases of CJD related to the use of human growth hormone have been reported in Canada, although cadaver-derived growth hormone was used in Canada from 1965 until 1985.[7]

Currently, blood is considered to pose a *theoretical* risk of transmitting CJD.[9] The Laboratory Centre for Disease Control (LCDC), Health Canada, is coordinating intensive active surveillance for CJD in Canada through the Creutzfeldt–Jakob Disease Surveillance System (CJD-SS Canada). This program is designed to determine the risk of developing CJD as a result of receiving blood, blood products or tissue transplants and to detect any cases of vCJD that might arise in relation to the consumption of beef. Neither BSE nor vCJD has been detected in Canada, but any such finding could be of considerable public-health significance as a "sentinel" observation.

CJD-SS Canada has enlisted the participation of clinicians in relevant specialties who will notify LCDC of any cases of CJD seen in their practices. Field investigators will conduct interviews to collect exposure information; this data will then be used in a case–control study. A blood donation history will also be taken to ensure that any stored blood products connected with the patient are recalled. Patient records will be reviewed to determine the date and location of any transfusions; if possible, it will also be determined whether CJD developed in any of the donors. Where possible, neuropathologic investigation will be conducted to confirm cases, and sequencing of the prion protein gene will be carried out for each confirmed case.

There is a national registry for those who received growth hormone as children in Canada. Physicians who note unexplained neurologic changes in patients previously treated with human growth hormone are asked to notify Dr. Heather Dean, chair of the Canadian Growth Hormone Advisory Committee, University of Manitoba at (204) 787-7435.

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Queries regarding the Creutzfeldt–Jakob Disease Surveillance System project may be directed to the Blood-borne Pathogens Division, Bureau of Infectious Diseases, LCDC, Health Canada (toll-free line 888 489-2999; fax 613 952-6668).

References

1. Brown P. Infectious cerebral amyloidosis: clinical spectrum, risks and remedies. *Dev Biol Stand* 1993;80:91-101.
2. Prusiner SB. Biology and genetics of prion diseases. *Ann Rev Microbiol* 1994;48:655-86.
3. Will RG. Surveillance of prion diseases in humans. In: Baker HF, Ridley RM, editors. *Methods in molecular medicine: prion diseases*. Totawa (NJ): Humana Press; 1996. p. 119-37.
4. Brown P. Environmental causes of human spongiform encephalopathy. In: Baker HF, Ridley RM, editors. *Methods in molecular medicine: prion diseases*. Totawa (NJ): Humana Press; 1996. p. 139-54.
5. Brown P, Preece MA, Will RG. "Friendly fire" in medicine: hormones, homografts and CJD. *Lancet* 1992;340:24-7.
6. Will RG, Ironside JW, Zeidler M, Cousens SN, Estibeiro K, Alperovitch A, et al. A new variant of Creutzfeldt–Jakob disease in the UK. *Lancet* 1996;347:921-5.
7. Bruce ME, Will RG, Ironside JW, McConnell I, Drummond D, Suttie A, et al. Transmissions to mice indicate that "new variant" CJD is caused by the BSE agent. *Nature* 1997;390(6650):498-501.
8. Hill AF, Desbruslais M, Joiner S, Sidle KCL, Gowland I, Collinge J. The same prion strain causes vCJD and BSE. *Nature* 1997;390(6650):448-50.
9. Dean HJ, Friesen HG. Growth hormone therapy in Canada: end of one era and beginning of another. *Can Med Assoc J* 1986;135:297-301.
10. Ricketts MN, Cashman NR, Stratton EE, El Saadany S. Is Creutzfeldt–Jakob disease transmitted in blood? *Emerg Infect Dis* 1997;3:155-63.