

Creutzfeldt-Jakob Disease (CJD)

Prepared by the Centre for Neurodegenerative Diseases,
University of Toronto

Is vCJD in Canada?

To date, there has been one case of variant CJD reported in Canada. Health Canada has confirmed that this person lived and traveled extensively throughout the United Kingdom during the height of the BSE (Mad Cow) epidemic and had eaten beef in the U.K. during that period. There is no indication the individual contracted the disease anywhere other than the United Kingdom.

To ensure that any cases of vCJD are detected immediately Health Canada has set up a CJD Surveillance System, which began in 1998. The Canadian surveillance unit screens approximately 90 samples that are suspected of having sCJD each year. Of these between 30 and 35 cases are diagnosed as sporadic CJD. These numbers are consistent with the one person per million worldwide occurrence of sCJD per year.

What is CJD?

CJD is a rare and fatal brain disease in humans. The symptoms include progressive loss of mental functioning and loss of coordination.

What causes CJD?

CJD is a member of a disease group known as transmissible spongiform encephalopathies (TSE). TSEs are fatal diseases of the central nervous system affecting both humans and animals. Scrapie in sheep, chronic wasting disease and BSE (mad cow disease) are examples of TSEs in the animal population.

Although it is not known what causes these diseases, the presence of an infectious agent known as the 'prion' (proteinaceous infectious particle) is believed to be responsible for a conversion of normal host proteins in the central nervous system. This conversion is the likely cause for a loss of brain cells resulting in the 'spongy' appearance of the brain tissue.

Are prion diseases in humans linked to 'mad cow disease' (BSE)?

There are actually several different forms of prion diseases affecting humans. The different types are categorized according to how someone would acquire the disease.

Sporadic CJD: The most common form is sporadic CJD (sCJD), which accounts for more than 85% of all identified cases of CJD. Although the cause for sCJD is unknown, it is unrelated to BSE exposure.

Creutzfeldt-Jakob Disease (CJD)

Hereditary: Prion diseases can also be genetically inherited. Hereditary forms of CJD include Gerstmann-Straussler-Scheinker syndrome (GSS) and fatal familial insomnia (FFI) and are caused by a mutation of the prion gene. This form is also unrelated to BSE exposure.

Acquired: The acquired prion diseases are due to transmissibility of the prion protein through ingestion or inoculation. The transmission of kuru disease in Papua, New Guinea was due to a cannibalistic burial ritual, which involved consumption of the deceased kin's brain. The discovery that the transmission of kuru was occurring in this manner ended this ritual and as a result kuru is virtually non-existent today.

Variant CJD: The form of CJD that is likely acquired through exposure to BSE is known as variant CJD (vCJD). Although this disease is very similar to sCJD, it affects a much younger age group. The sporadic form is most commonly seen in those between 60-65 years of age, whereas vCJD patients are typically younger in age (average 28 years). The clinical course of the disease is also generally prolonged (1-2 years), in comparison to sCJD (2-12 months).