

VARIANT CREUTZFELDT-JAKOB DISEASE (VCJD)

Case definitions

CONFIRMED CASE

IA [see [Box 5](#)] and neuropathologic confirmation as per pathologic features [see [Footnote a, Box 5](#)].

PROBABLE CASE

I + four or five criteria of II, IIIA and IIIB [see [Box 5](#)].

OR

I + IVA [see [Box 5](#)].

SUSPECT CASE

I + four or five criteria of II + IIIA [see [Box 5](#)].

Box 5

- I. A – Progressive neuropsychiatric disorder
B – Duration > 6 months
C – Routine investigations do not suggest alternative diagnosis
D – No history of potential iatrogenic exposure
E – No evidence of genetic prion disease
- II. A – Early psychiatric symptoms ^b
B – Persistent painful sensory symptoms ^c
C – Ataxia
D – Myoclonus or chorea or dystonia
E – Dementia
- III. A – EEG does not show typical appearance of sporadic CJD ^d [or no EEG performed] in the early stages of the illness.
B – Bilateral pulvinar high signal on magnetic resonance imaging (MRI) scan ^e
- IV. A – Tonsil biopsy positive for prion protein immunoreactivity ^f

Footnotes

- a) Spongiform change, extensive PrP deposition, florid plaques throughout the cerebrum & cerebellum.
- b) Depression, anxiety, apathy, withdrawal, delusions.
- c) Frank pain and/or dysaesthesia.
- d) Generalized triphasic period complexes at ca. 1 Hz. Rarely, these may occur in the late stages of vCJD.
- e) Relative to the signal intensity of other deep grey matter nuclei & cortical grey matter.
- f) Tonsil biopsy is not recommended routinely or in cases with EEG appearance typical of sporadic CJD, but may be useful in suspect cases in which the clinical features are compatible with vCJD and MRI does not show bilateral pulvinar high signal.

Causative agent

Thought to be a unique, self-replicating protein called a prion that replicates by a poorly understood mechanism

Source

Humans. Variant CJD is believed to be associated with a disease in cattle called bovine spongiform encephalopathy (BSE), more commonly known as 'mad cow disease'.

Incubation

Fifteen months to possibly more than 30 years

Transmission

Although there is strong evidence that the agent responsible for human cases of vCJD is the same agent responsible for BSE in cattle, the specific foods that may be associated with the transmission of this agent from cattle to humans are unknown.

Communicability

Central nervous system (CNS) tissues are infectious throughout symptomatic illness. Other tissues and cerebral spinal fluid (CSF) are sometimes infectious.

Symptoms

Persons with vCJD usually experience psychiatric symptoms, early in illness, which most commonly take the form of depression, or a "schizophrenic-like" psychosis. As the illness progresses, neurological signs include: unsteadiness, difficulty walking and involuntary muscle movements.

Variant CJD typically affects younger patients (average 28 years) and has a relatively long duration of illness – 14 months compared to 4.5 months for classic CJD.

Diagnostic testing

Consult your local laboratory

Treatment

There is no known effective treatment available to cure or control vCJD and the disease appears to be uniformly fatal. Current treatment is therefore aimed at controlling symptoms and making the person as comfortable as possible.

PUBLIC HEALTH MANAGEMENT & CONTROL

Case management

- Obtain the client's medical history including symptoms, date of onset, treatment, surgical procedures of concern, and/or hospitalization and any potential sources of exposure, particularly a history of any receipt or donation of blood, blood products, cells, tissues or organs.
- If necessary, contact the client's physician to obtain further information and clarification of the client's history, especially with respect to past surgical procedures and blood/tissue/organ receipt and/or donation.
- If the client has a history of receipt or donation of blood, cells, tissues or organs, inform the Medical Officer of Health immediately and fax the "CJD Case Report Form" to the Department of Health & Wellness so that appropriate 'lookback' and 'traceback' procedures may be initiated immediately.
- If client has a history of surgical procedures of concern when symptomatic, inform infection control program in hospital where procedure occurred.
- Discuss the role of public health and provide information to the individual or family [i.e., general information sheets].
- Complete the "CJD/vCJD Case Report Form" and update as new information becomes available.
- If client is deceased, ensure that attending physician has notified the funeral director of vCJD diagnosis so appropriate infection control precautions can be taken.

Education

People who spent six or cumulative months between January 2, 1968 and December 31, 1996 in the United Kingdom should not donate blood, organs or other body tissues or fluids.

Surveillance forms

novascotia.ca/dhw/populationhealth/surveillanceguidelines/NS_Notifiable_Disease_Surveillance_Case_Report_Form.pdf

novascotia.ca/dhw/populationhealth/surveillanceguidelines/CJD_Case_Report_Form.pdf

[General Information Sheet](#)