Variant Creutzfeldt-Jakob disease

Australian national notifiable diseases case definition

This document contains the surveillance case definition for variant Creutzfeldt-Jakob disease which is nationally notifiable within Australia. State and territory health departments use this definition to decide whether to notify the Australian Government Department of Health and Aged Care of a case.

Version	Status	Last reviewed	Implementation date
1.0	Initial CDNA case definition	CDWG November 2009	1 July 2010

Reporting

Confirmed and **probable** cases should be notified. (NB: a "confirmed" case is equivalent to the ANCJDR classification of "definite")

Confirmed case

A confirmed case requires **laboratory definitive evidence** AND **clinical evidence**

Laboratory definitive evidence

Neuropathological confirmation of vCJD

Clinical evidence

Progressive neuropsychiatric disorder

Probable case

A probable case requires **clinical definitive evidence**

OR

Clinical suggestive evidence AND **laboratory suggestive evidence.**

Clinical definitive evidence

1. Progressive neuropsychiatric disorder AND duration of illness greater than six months AND routine investigations do not suggest an alternative diagnosis AND no history of potential iatrogenic exposure AND no evidence of a familial form of TSE

AND

2. Four of the following symptoms:

- Early psychiatric symptoms
- Persistent painful sensory symptoms
- Ataxia
- Myoclonus or chorea or dystonia
- Dementia

AND

3. Bilateral pulvinar high signals on magnetic resonance imaging (MRI) scans

AND

4. Electroencephalogram (EEG) which does not exhibit the typical appearance of classic CJD

Clinical suggestive evidence

1. Progressive neuropsychiatric disorder AND duration of illness greater than six months AND routine investigations do not suggest an alternative diagnosis AND no history of potential iatrogenic exposure AND no evidence of a familial form of TSE

Laboratory suggestive evidence

1. A PrP^{sc} positive tonsil biopsy