Creutzfeldt-Jakob disease (CJD)

Australian national notifiable diseases case definition

This document contains the surveillance case definition for Creutzfeldt-Jakob disease (CJD), 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. This includes sporadic, accidental and familial cases. (NB: a "confirmed" case is equivalent to the ANCJDR classification of "definite")

Confirmed case

A confirmed case requires **laboratory definitive evidence**

Laboratory definitive evidence

Neuropathological confirmation of CJD supplemented by immunochemical detection of protease-resistant PrP by western blot OR immunocytochemistry.

Probable case

A probable case requires **clinical evidence** AND either electroencephalogram (EEG) or laboratory suggestive evidence.

Laboratory suggestive evidence

Positive 14-3-3 protein CSF test.

Clinical evidence

1. Progressive dementia of less than two years duration; AND
2. At least 2 of the following clinical features:
    - myoclonus
    - visual or cerebellar signs
    - pyramidal/extrapyramidal signs
    - akinetic mutism.