

Diagnostic criteria for variant CJD
Will, RG. et al. Ann. Neurol, 2000; 47: 575-82

- I. A Progressive neuropsychiatric disorder
- B Duration of illness > 6 months
- C Routine investigations do not suggest an alternative diagnosis
- D No history of potential iatrogenic exposure
- II. A Early psychiatric symptoms*
- B Persistent painful sensory symptoms**
- C Ataxia***
- D Myoclonus or chorea dystonia
- E Dementia
- III. A Electroencephalogram (EEG) does not show the typical appearance of classical CJD (after review by CJDSU staff)*** or no EEG performed
- B Posterior thalamic high signal on MRI scan (after review by CJDSU staff)

Definite variant CJD: IA and neuropathological confirmation of vCJD****

Probable variant CJD: I and 4/5 of II and III.A and III.B

Possible variant CJD: I and 4/5 of II and III.A

* depression, anxiety, apathy, withdrawal, delusions

** including both frank pain and/or unpleasant dysaesthesia

*** generalised triphasic periodic complexes at approximately one per second

**** spongiform change and extensive PrP deposition with florid plaques, throughout the cerebrum and cerebellum

In addition, there are three additional sub-categories for those referrals that do not meet the criteria of possible CJD, which are;

- Diagnosis unclear – when the diagnostic criteria for possible, probable or definite CJD are not met nor is there a reasonable alternative diagnosis and, therefore, CJD remains a possibility;
- CJD thought unlikely – when information indicates that a clinical diagnosis of CJD is very unlikely because of atypical disease features, and/or an atypical course, and/or atypical clinical investigation results, and/or a reasonable alternative diagnosis is made, but is not confirmed. This category includes cases which improve clinically without another firm diagnosis being made;
- Definitely not CJD – when information indicates that CJD is not the diagnosis and there is another definite diagnosis proven by clinical examination, clinical investigations, or pathology.
- Since publication, in the UK, a probable case also includes someone with a positive tonsil biopsy and a progressive neuropsychiatric disorder of greater than six months' duration, no alternative diagnosis and no history of iatrogenic exposure.