

vCJD case definition

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- 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
- E     No evidence of a familial form of TSE
  
- II    A     Early psychiatric symptoms <sup>a</sup>
- B     Persistent painful sensory symptoms <sup>b</sup>
- C     Ataxia
- D     Myoclonus or chorea or dystonia
- E     Dementia
  
- III   A     EEG does not show the typical appearance of sporadic CJD <sup>c</sup>  
       (or no EEG performed)
- B     MRI brain scan shows bilateral symmetrical pulvinar high signal<sup>d</sup>
  
- IV    A     Positive tonsil biopsy <sup>e</sup>

DEFINITE:            I A **and** neuropathological confirmation of vCJD <sup>f</sup>

PROBABLE:           I **and** 4/5 of II **and** III A **and** III B

**OR**

**I and IV A<sup>d</sup>**

POSSIBLE:           I and 4/5 of II and III A

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a depression, anxiety, apathy, withdrawal, delusions.

b this includes both frank pain and/or dysaesthesia.

c generalised triphasic periodic complexes at approximately one per second.

d relative to the signal intensity of other deep grey matter nuclei and cortical grey matter.

e tonsil biopsy is not recommended routinely, nor in cases with EEG appearances typical of sporadic CJD, but may be useful in suspect cases in which the clinical features are compatible with vCJD and where MRI does not show bilateral pulvinar high signal.

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

# クロイツフェルト・ヤコブ病診療マニュアル(改訂版)

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