<table>
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<tr>
<th>CJD Type</th>
<th>Case Definitions and Confirmation</th>
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<td><strong>Confirmed</strong>: Diagnosed by standard neuropathological techniques; and/or immunocytochemically; and/or Western blot confirmed protease-resistant PrP; and/or presence of scrapie-associated fibrils.</td>
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|           | **Probable**: Progressive dementia  
And  
at least 2 of the following 4 clinical features:  
- Myoclonus  
- Visual or cerebellar disturbance  
- Pyramidal/extrapyramidal dysfunction  
- Akinetic mutism  
And  
A typical EEG during an illness of any duration; and/or a positive 14-3-3 CSF assay and a clinical duration to death of <2 years  
Routine investigations should not suggest an alternative diagnosis |
|           | **Possible**: Progressive dementia  
And  
at least 2 of the following 4 clinical features:  
- Myoclonus  
- Visual or cerebellar disturbance  
- Pyramidal/extrapyramidal dysfunction  
- Akinetic mutism  
And  
No EEG or atypical EEG and duration <2 years |
|           | **Confirmed**: Neuropathologic examination of brain tissue is required to confirm a diagnosis of variant CJD. The following confirmatory features should be present.  
- Numerous widespread kuru-type amyloid plaques surrounded by vacuoles in both the cerebellum and cerebrum–florid plaques.  
- Spongiform change and extensive prion protein desposition shown by immunohistochemistry throughout the cerebellum and cerebrum. |
|           | **Suspect**  
a. Current age at death <55 years (a brain autopsy is recommended, however, for all physician-diagnosed CJD cases).  
b. Psychiatric symptoms at illness onset and/or persistent painful sensory symptoms (frank pain and/or dysesthesia).  
c. Dementia, and development =4 months after illness onset of at least two of the following five neurological signs:  
   - Poor coordination  
   - Chorea  
   - Hyperreflexia  
   - Myoclonus  
   - Visual signs  
(If persistent painful sensory symptoms exist, =4 months delay in the development of the neurologic signs is not required).  
d. A normal or an abnormal EEG, but not the diagnostic changes often seen in sporadic CJD. |
Creutzfeldt-Jakob Disease Case Definitions

e. Duration of illness over 6 months.
f. Routine investigations of the patient do not suggest an alternative, non-CJD diagnosis.
g. No history of receipt of cadaveric human pituitary growth hormone or a dura mater graft.
h. No history of CJD in a first degree relative or prion protein gene mutation in the patient.

NOTE

a. If a patient has the typical bilateral pulvinar high signal on MRI scan, a suspected diagnosis of variant CJD requires the presence of a progressive neuropsychiatric disorder, d, e, f and g of the above criteria and four of the following five criteria
   - Early psychiatric symptoms (anxiety, apathy, delusions, depression, withdrawal)
   - Persistent painful sensory symptoms (frank pain and/or dysesthesia)
   - Ataxia
   - Myoclonus or chorea or dystonia
   - Dementia

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<th>Iatrogenic</th>
<th>Progressive cerebellar syndrome in a recipient of human cadaveric-derived pituitary hormone</th>
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<td>Or</td>
<td>Sporadic CJD with a recognized exposure risk, e.g. antecedent neurosurgery with dura mater implantation.</td>
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<th>Familial</th>
<th>Confirmed or probable CJD plus confirmed or probable CJD in a first degree relative</th>
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<td>And/or</td>
<td>Neuropsychiatric disorder plus disease-specific prion protein (PrP) gene mutation</td>
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