

Diagnostic Criteria for Variant Creutzfeldt-Jakob Disease in the United States

- I. **Definite Variant CJD:** Neuropathologic examination of brain tissue is required to confirm a diagnosis of variant CJD. The following confirmatory features should be present.
 - a. Numerous widespread kuru-type amyloid plaques surrounded by vacuoles in both the cerebellum and cerebrum - florid plaques.
 - b. Spongiform change and extensive prion protein deposition shown by immunohistochemistry throughout the cerebellum and cerebrum.
- II. **Suspected Variant CJD**
 - a. Current age or 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 neurologic signs: poor coordination, myoclonus, chorea, hyperreflexia, or 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 EEG changes often seen in classic CJD.
 - e. Duration of illness of 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

1. 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: 1) early psychiatric symptoms (anxiety, apathy, delusions, depression, withdrawal); 2) persistent painful sensory symptoms (frank pain and/or dysesthesia); 3) ataxia; 4) myoclonus or chorea or dystonia; and 5) dementia.
2. A history of possible exposure to bovine spongiform encephalopathy (BSE) such as residence or travel to a BSE-affected country after 1980 increases the index of suspicion for a variant CJD diagnosis.