

CDC's Diagnostic Criteria for Creutzfeldt-Jakob Disease (CJD), 2010

Adapted from: a) Global Surveillance, diagnosis, and Therapy of Human Transmissible spongiform Encephalopathies: Report of WHO consultation, February 9-11, 1998, Geneva, Switzerland; and b) Zerr, I, Kallenberg K, Summers DM, et al. Brain 2009, 132; 2659-2668.

1. Sporadic CJD

Definite:

Diagnosed by standard neuropathological techniques; and/or immunocytochemically; and/or Western blot confirmed protease-resistant PrP; and /or presence of scrapie-associated fibrils.

Probable:

Rapidly progressive dementia; and at least two out of the following four clinical features:

- i. Myoclonus
- ii. Visual or cerebellar signs
- iii. Pyramidal/extrapyramidal signs
- iv. Akinetic mutism

AND a positive result on at least one of the following laboratory tests:

- a. a typical EEG (periodic sharp wave complexes) during an illness of any duration; and/or
- b. a positive 14-3-3 cerebrospinal fluid (CSF) assay in patients with a disease duration of less than 2 years
- c. Magnetic resonance imaging (MRI) high signal abnormalities in caudate nucleus and/or putamen on diffusion-weighted imaging (DWI) or fluid attenuated inversion recovery (FLAIR)

AND without routine investigations indicating an alternative diagnosis.

Possible:

Progressive dementia; and at least two out of the following four clinical features:

- i. Myoclonus
- ii. Visual or cerebellar signs
- iii. Pyramidal/extrapyramidal signs
- iv. Akinetic mutism

AND the absence of a positive result for any of the three laboratory tests that would classify a case as "probable" (see tests a-c above)

AND duration of illness less than two years

AND without routine investigations indicating an alternative diagnosis.

2. Iatrogenic CJD

Progressive cerebellar syndrome in a recipient of human cadaveric-derived pituitary hormone; or sporadic CJD with a recognized exposure risk, e.g., antecedent neurosurgery with dura mater implantation.

3. Familial CJD

Definite or probable CJD plus definite or probable CJD in a first degree relative; and/or Neuropsychiatric disorder plus disease-specific PrP gene mutation.

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

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.

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.

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Content source: [Centers for Disease Control and Prevention](#)

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