

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/extrapyrmidal 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/extrapyrmidal 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.