Creutzfeldt-Jakob Disease (CJD)

Merlin disease code=04610
Case report form (CRF): Creutzfeldt-Jakob Disease Worksheet
PAPER CRF REQUIRED

Background
A progressive uniformly fatal dementia characterized by myoclonus, visual or cerebellar signs, akinetic mutism, and pyramidal or extrapyramidal signs.

Clinical criteria for case classification
Confirmatory:
A fatal outcome with a clinically compatible illness.

Presumptive:
All of the following:
• A fatal outcome, and
• Progressive dementia, and
• A clinical duration to death <2 years, and
• At least 2 of the following clinical features:
  o Myoclonus
  o Visual or cerebellar signs
  o Pyramidal or extrapyramidal signs
  o Akinetic mutism, and
• No alternative diagnosis suggested during routine investigation.

Laboratory criteria for case classification
Confirmatory:
One or more of the following:
• Standard neuropathological techniques, or
• Immunocytochemical testing, or
• Western blot confirmed protease-resistant prion protein, or
• Presence of scrapie-associated fibrils conducted on brain tissue.

Presumptive:
Either of the following:
• Analysis of tau or 14-3-3 proteins in CSF consistent with prion disease or
• Periodic sharp and slow wave complexes in electroencephalogram (EEG) (test suggestive but not specific for CJD).

Supportive:
No EEG or atypical EEG.

Epidemiological criteria for case classification
Not applicable.
**Case classification**

**Confirmed:**
A person with confirmatory clinical criteria and confirmatory laboratory evidence.

**Probable:**
A person with presumptive clinical criteria and presumptive laboratory evidence.

**Suspect:**
A person with presumptive clinical criteria and supportive laboratory evidence.

**Criteria to distinguish a new case from previous reports**
Not applicable.

**Comments**
Cases under the age of 55 years old should be evaluated for the variant form of CJD. Brain tissue for diagnosis and CSF for the tau and 14-3-3 protein should be sent to the National Prion Disease Pathology Surveillance Center at Case Western Reserve University. Information about the center and shipping instructions can be found on their web site: [www.cjdsurveillance.com](http://www.cjdsurveillance.com). Please notify Bureau of Epidemiology to assist with case evaluation and laboratory testing.

[Return to Table of Contents](#)