Creutzfeldt-Jakob Disease (CJD)

January 2015

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

Laboratory criteria for diagnosis
- Standard neuropathological techniques; and/or immunocytochemically; and/or Western blot confirmed protease-resistant PrP; and/or presence of scrapie-associated fibrils conducted on brain tissue.
- Analysis of tau or 14-3-3 proteins in CSF consistent with prion disease.
- Periodic sharp and slow wave complexes (PSWC) in EEG (Test suggestive but not specific for CJD).

Case classification
Confirmed:
A fatal outcome with a clinically compatible illness diagnosed by standard neuropathological techniques; and/or immunocytochemically; and/or Western blot confirmed protease-resistant PrP; and/or presence of scrapie-associated fibrils.

Probable:
A fatal outcome with a progressive dementia and ≥2 out of the following 4 clinical features:
- Myoclonus
- Visual or cerebellar signs
- Pyramidal/extrapyramidal signs
- Akinetic mutism
AND
- A clinical duration to death <2 years,
WITH
- Typical EEG during clinical illness or a tau or 14-3-3 CSF assay results consistent with prion disease, and
- No alternative diagnosis suggested during routine investigation.

Suspect:
A fatal outcome with a progressive dementia and ≥2 out of the following 4 clinical features:
- Myoclonus
- Visual or cerebellar signs
- Pyramidal/extrapyramidal signs
- Akinetic mutism
AND
- No EEG or atypical EEG and a clinical duration to death of <2 years.

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: http://www.cjdsurveillance.com. Please notify Bureau of Epidemiology to assist with case evaluation and laboratory testing.