



PREVENTION OPPORTUNITIES UNDER THE BIG SKY

Creutzfeldt-Jakob Disease in Montana

Creutzfeldt-Jakob disease (CJD), a form of transmissible spongiform encephalopathy (TSE), is a rare degenerative brain disease that is always fatal. In the United States, approximately 200 cases of CJD are reported each year. CJD is believed to be caused by abnormal prion proteins (PrP). CJD is subdivided into three major categories: sporadic CJD, hereditary CJD, and variant CJD. Approximately 85% of CJD cases are **sporadic CJD (sCJD)** in which the patient typically lacks risk factors for other categories of CJD. Five to 15% of the cases are **hereditary CJD**, an autosomal dominant condition related to one of several possible mutations of the PRNP gene that codes PrP. Patients with hereditary CJD will have a family history of the disease and/or positive tests for the genetic mutation associated with abnormal PrP. **Variant CJD (vCJD)** represents less than 1% of the cases of CJD and occurs following exposure to brain or nervous system tissue. Rarely, iatrogenic transmission of CJD has been reported following exposure to neurosurgical equipment previously used on patients that died of CJD, pituitary-derived human growth hormone, or dura mater grafts. The clinical and pathological characteristics of sCJD and vCJD differ considerably (Table). No curative treatment exists for CJD.

Diagnosis of CJD

The diagnosis of CJD is based upon clinical symptoms and signs, radiological imaging, electroencephalogram (EEG) patterns, laboratory tests, and neuropathological analysis. The diagnostic criteria for CJD can be found at <http://www.cdc.gov/ncidod/dvrd/cjd>.

More specifically, in patients for whom a more likely diagnosis has been excluded, the use of EEG and magnetic resonance imaging (MRI), and the evaluation of cerebrospinal fluid for protein 14-3-3 and Tau protein can provide supportive evidence of CJD. Importantly, these results cannot confirm the diagnosis of CJD. At present, confirmation of CJD requires postmortem examination of brain tissue. Furthermore, an autopsy of the brain is the only method that can exclude vCJD as a diagnosis and can potentially detect other types of TSEs. The autopsy should be performed as soon after death as possible, but the tissue can be examined successfully within 48–72 hours after death if refrigeration is used.

The National Prion Disease Pathology Surveillance Center (NPDPS) provides autopsy and laboratory support services for the testing of patients suspected of having CJD. Healthcare providers who suspect a patient of having CJD are encouraged to pursue laboratory and autopsy testing with the NPDPS. More information about the laboratory and autopsy services offered by NPDPS, and important shipping and processing instructions, can be found at <http://www.cjdsurveillance.com/> or by calling (206) 368-0587.

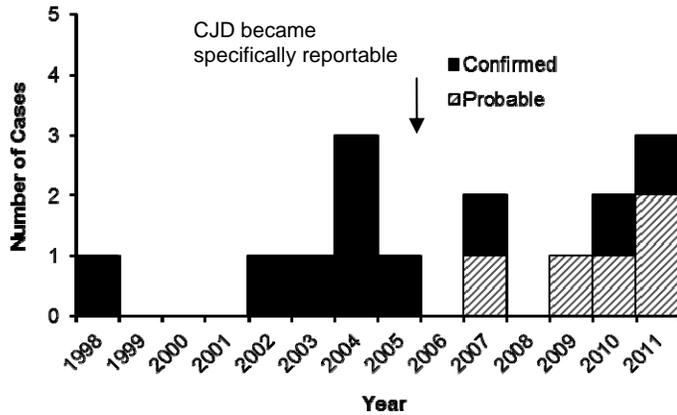
Table. Clinical and pathological characteristics of sporadic CJD (sCJD) and variant CJD (vCJD)

Characteristic	sCJD	vCJD
Median age at death	68 years	28 years
Median duration of illness	4–5 months	13–14 months
Clinical signs/ symptoms	Dementia; early neurologic signs	Prominent psychiatric/behavioral symptoms; painful dysesthesias; delayed neurological signs
“Pulvinar sign” on MRI*	Not reported	Present in >75% of cases
Presence of ‘florid plaques’ on neuropathology	Rare or absent	Present in large numbers
Immunohistochemical analysis of brain tissue	Variable accumulation	Marked accumulation of protease-resistance prion protein
Presence of agent in lymphoid tissue	Not readily detected	Readily detected
Increased glycoform ratio on immunoblot analysis of protease-resistance prion protein	Not reported	Marked accumulation of protease-resistance prion protein

Source: <http://www.cdc.gov/ncidod/dvrd/cjd/>

*An abnormal signal in the posterior thalami on T2- and diffusion weighted images and fluid attenuated inversion recovery sequences on brain magnetic resonance imaging (MRI); in the appropriate clinical context this signal is highly specific for vCJD.

Figure. Reported confirmed and probable Creutzfeldt-Jakob Disease (CJD) cases — Montana, 1998–2011



Montana CJD Cases, 1998–2011

In Montana, a case of suspected TSE (including CJD) is a reportable condition (ARM 37.114.203). A case of TSE first became specifically reportable in 2006. Since 1998, 15 confirmed or probable cases of CJD have been reported in Montana (Figure). Of the 15 cases, 11 (73%) occurred among males and the median age was 68 years (range: 49–84 years). In 2011, a cluster of three sCJD cases (one confirmed and two probable) occurred in a single Montana county, which most likely represented a statistical coincidence of three sCJD cases clustering in time and place. To improve surveillance of CJD, healthcare providers should immediately report a case of suspected CJD to the local health officer and report the ICD-9 code 046.1 (CJD) on the death certificate.

General Information and Recommendations for Creutzfeldt-Jacob Disease (CJD)

- Over 85% of the cases of CJD are sporadic and not related to any known risk factors for acquiring CJD.
- To confirm a CJD diagnosis, clinicians should arrange for autopsies for patients suspected of having CJD.
- The National Prion Disease Pathology Surveillance Center (NPDPSC) provides autopsy and laboratory services for patients suspected of having CJD. Contact the NPDPSC at (216) 368-0587 or visit <http://www.cjdsurveillance.com/> to make arrangements for laboratory and autopsy testing.
- Physicians should use ICD-9 code 046.1 on the death certificate for any patient diagnosed with CJD.
- Healthcare providers should report any suspected case of CJD immediately to their local health officer.
- For more information, contact the Communicable Disease Epidemiology section at (406) 444-0273.

References:

1. CDC. Creutzfeldt-Jacob Disease. <http://www.cdc.gov/ncidod/dvrd/cjd/> [accessed February 22, 2012].
2. Belay ED and Schonberger LB. The public health impact of prion diseases. *Ann Rev Public Health.* 2005;26:191–212.
3. CJD National Prion Disease Pathology Surveillance Center. <http://www.cjdsurveillance.com/> [accessed February 22, 2012].
4. CDC. National, State, and Local Area Vaccination Coverage Among Children Aged 19–35 Months — United States, 2009. *MMWR.* 2010;59:1171–77.

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