



Human Prion Disease in Washington, 2006–2012

Human Prion Diseases

Prion diseases are a group of rare brain and nervous system diseases that affect humans and some kinds of animals. Creutzfeldt-Jakob disease (CJD) is by far the most common human prion disease. It is a rare, fatal neurodegenerative disease commonly characterized by rapidly progressing dementia, poor balance, visual changes and/or muscle jerks. **Sporadic CJD** has no known cause and accounts for about 85% of all CJD cases. It occurs worldwide at a rate of approximately 1 case per million population per year(1), however, higher rates have been reported(2). Most persons affected with sporadic CJD are over 55 years old. **Familial CJD** results from an inherited mutation and accounts for 10–15% of cases. Rarely, CJD is acquired. It has been transmitted through certain surgical procedures or human-derived growth hormone. In 1996, a new **variant CJD** recognized in the United Kingdom was associated with eating cattle products from cows affected with bovine spongiform encephalopathy (“mad cow disease”). To date, no cases of variant CJD are thought to have been acquired in Washington or the United States.

Surveillance Strategies in Washington State

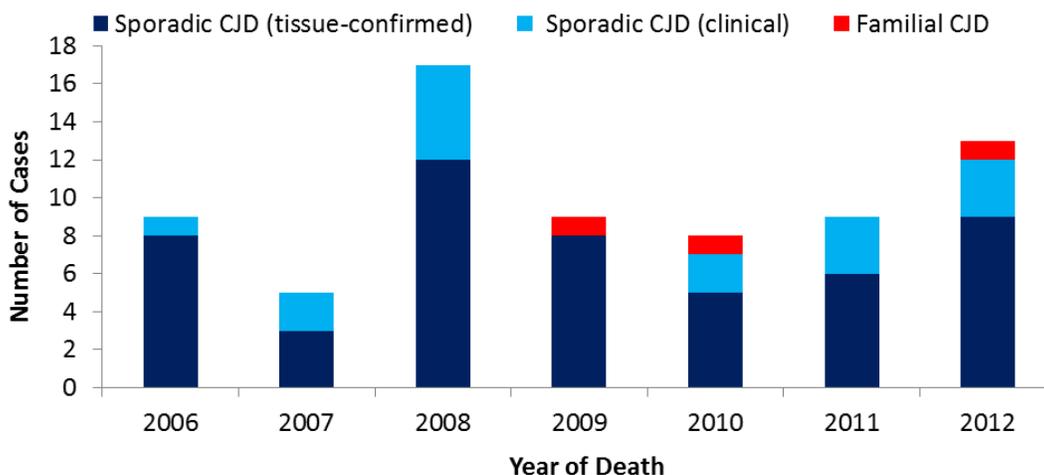
In December 2003, the first known case of bovine spongiform encephalopathy (i.e., “mad cow disease”) in the United States was diagnosed in an adult Holstein cow from Washington State. Prior to the identification of this cow, surveillance for human prion disease (including all types of CJD) in Washington was performed through death certificate review. Beginning in 2004, the Washington State Department of Health (DOH) enhanced efforts to identify and confirm human prion disease by collaborating with the Centers for Disease Control and Prevention (CDC) and the National Prion Disease Pathology Surveillance Center (NPDPSC). Healthcare providers in Washington are required to report suspected human prion disease to the local health jurisdiction where the patient resides.

Human Prion Disease in Washington State

During 2006–2012, 70 cases of CJD were detected in Washington (average 10 cases per year); 67 (96%) were sporadic CJD and 3 (4%) were familial CJD. No cases of variant CJD were diagnosed. Of the sporadic CJD cases, 51 (76%) were confirmed by examination of brain tissue and 16 (24%) were clinically diagnosed. The average yearly incidence rate of sporadic CJD over this time period was 1.4 cases per million population per year based on population estimates provided by the Office of Financial Management.

The following graph and table show the number of CJD cases by type and year of death and the characteristics of CJD cases diagnosed in Washington during 2006–2012.

Number of CJD Cases by Type and Year of Death, Washington State, 2006–2012 (n=70)



Characteristics of CJD Cases, Washington State, 2006–2012 (n=70)

Characteristic	No. Cases (%)
Male	34 (49)
Median age [range]	68 years [38–84 years old]
Western WA residents*	52 (74)
Median duration of illness	4 months

*Western Washington is defined as living west of the Cascade Mountains. In 2010, 78% of the population lived in western Washington.

All clinically diagnosed patients had a clinical presentation consistent with sporadic CJD. Five patients were less than 55 years old at the time of death; 3 were confirmed sporadic CJD, one was confirmed familial CJD, and one was diagnosed clinically. This last patient was 49 years old and developed symptoms 4 months prior to death, suggestive of sporadic CJD.

Summary

The incidence of sporadic CJD in Washington State is consistent with reported rates worldwide. During 2006–2012, 76% of sporadic CJD cases were confirmed by examination of brain tissue. No cases of variant CJD were diagnosed or suspected.

References

1. Holman RC, Belay ED, Christensen KY, et al. Human prion diseases in the United States. *PLoS One*. 2010;5(1):e8521.
2. Ladogana A, Puopolo M, Croes EA, et al. Mortality from Creutzfeldt-Jakob disease and related disorders in Europe, Australia, and Canada. *Neurology* 2005;64(9):1586-91.