



# Human Prion Disease in Washington, 2004–2009

## 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 usually characterized by rapidly progressing dementia, poor balance, visual changes and/or muscle jerks. Sporadic CJD (sCJD) has no known cause and accounts for about 85% of CJD cases. It occurs worldwide at a rate of approximately 1 case per million population per year<sup>1</sup>, however, higher rates have been reported<sup>2</sup>. Most persons affected with sCJD are over 55 years old. Familial CJD (fCJD) results from an inherited mutation and accounts for 10–15% of cases. Rarely, CJD has been transmitted through certain surgical procedures or human-derived growth hormone. In 1996, a new variant of CJD (vCJD) 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 vCJD are thought to have been acquired in Washington or the United States.

## Surveillance Strategies in Washington State

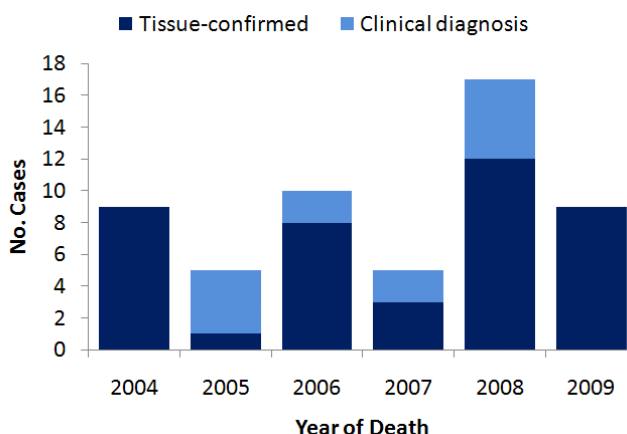
In December 2003, the first known case of bovine spongiform encephalopathy (i.e., “mad cow disease”) in the United States was diagnosed in an adult Holstein cow from Washington State. Prior to the identification of this cow, surveillance for human prion disease in Washington was performed through death 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 and the National Prion Disease Pathology Surveillance Center (NPDPSC), and investigating Washington residents with elevated 14-3-3 protein prior to death. In addition, healthcare providers in Washington are required to report suspect human prion disease as a rare disease of public health significance.

## Human Prion Disease in Washington State

During 2004–2009, 55 cases of CJD were detected in Washington; 42 (76%) were tissue-confirmed by NPDPSC and 13 (24%) were clinically diagnosed. The average yearly incidence rate over this time period is 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 year of death and the characteristics of CJD cases diagnosed in Washington during 2004–2009.

## Number of CJD Cases by Year of Death, Washington, 2004–2009



## Characteristics of Probable or Confirmed CJD Cases, Washington, 2004–2009 (n=55)

Characteristic	No. Cases (%)
Male	26 (47)
Median age [range]	67 years [38–87 years old]
Western WA residents*	41 (75)
Median duration of illness** [range]	3 months [2–20 months]
Laboratory-confirmed	42 (76)
Sporadic CJD (sCJD)	37 (88)
Familial CJD (fCJD)	1 (2)
Sporadic or familial CJD	4 (10)
Variant CJD	0 (0)

\*Western Washington is defined as living west of the Cascade Mountains. In 2009, 78% of the population lived in western Washington.

\*\*Data available for 2007–2009 only. Onset date is missing for one case.

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

### Summary

The incidence of CJD in Washington State is consistent with reported rates worldwide. During 2004–2009, 76% of cases were tissue-confirmed. No cases of vCJD were diagnosed.

### 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.