

Human Prion Disease

Prion diseases, also referred to as transmissible spongiform encephalopathies (TSE), are a rare group of progressive neurodegenerative disorders that can occur in humans and animals. Prion diseases can be sporadic, inherited, iatrogenic, or acquired. Creutzfeldt-Jakob disease (CJD) is the most common human prion disease. It is a rare, fatal disease commonly 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 all CJD cases. Familial CJD (fCJD) results from an inherited mutation and accounts for 10–15% of cases.

Very rarely, CJD is acquired. In 1996, a new variant CJD (vCJD) recognized in the United Kingdom was associated with eating cattle products from cows affected with bovine spongiform encephalopathy (“mad cow disease”).

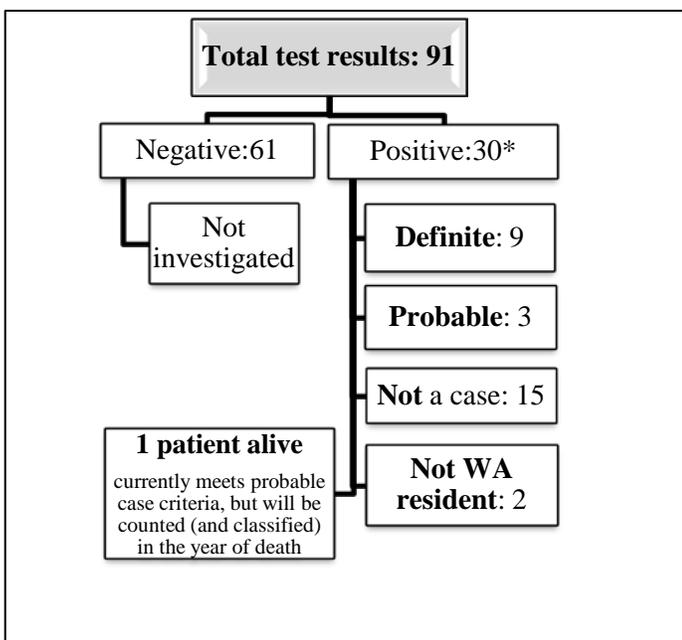
To date, no cases of variant CJD are thought to have been acquired in Washington or the United States.

Iatrogenic transmission of the CJD agent (iCJD) has been linked to the use of contaminated human growth hormone (hGH), dura mater and corneal grafts, or neurosurgical equipment. All of the equipment-related cases occurred before the implementation of the routine sterilization procedures currently used in health care facilities. No equipment-related cases have been reported since 1976. In the United States, 29 iCJD cases have been linked to the use of pituitary hGH in patients treated before 1977. The growth hormone currently used for treatment poses no threat of infection with CJD.

The only available method for confirming the diagnosis of prion diseases is the pathologic examination of brain tissue (autopsy or biopsy). Clinical symptoms, in conjunction with some non-confirmatory diagnostic tests (14-3-3 protein in cerebrospinal fluid, MRI, and EEG), can be used to make a clinical diagnosis of probable CJD.

Prion disease test results for 91 individuals were received by DOH during 2015. Of these, 61 were not suggestive of prion disease. Follow up investigation of positive* lab results from 30 patients revealed that 13 had a condition that met the CDC criteria for a definite or probable case (one of these patients was alive as Dec 31st 2015, and will be counted as a case in the year of death, according to CDC protocol). Of the remaining 17 patients, 15 were diagnosed with a different condition. Two were not WA State residents and follow up was done by other states.

Prion disease surveillance overview. WA, 2015



Prion disease deaths by county. WA, 2015

County	n
Chelan	1
Douglas	1
King	3
Kitsap	1
Pierce	1
Skagit	1
Snohomish	1
Spokane	2
Yakima	1
Total	12

* Includes a positive biopsy/autopsy or a CSF test result with at least one positive result for T-tau protein, 14-3-3 protein, or RT-QuIC.

Characteristics of CJD cases, 2015

Females accounted for 67% of cases.

The median age at diagnosis was 70.5 years (range: 52-78).

The median time between onset of symptoms and death was 4 months (range: 2-15 months).

The incidence of CJD was 1.7 cases/million population.

The average world-wide occurrence of CJD is approximately 1-2 cases/million population per year.

The incidence of CJD in Washington State is consistent with reported rates worldwide.

Confirmatory pathologic testing (biopsy and/or autopsy) was performed in 75% of the cases. Autopsy was performed for 67% of the cases

No iatrogenic or variant CJD was reported.

CJD deaths by age, sex, type, and case classification. WA State, 2015 (n=12)

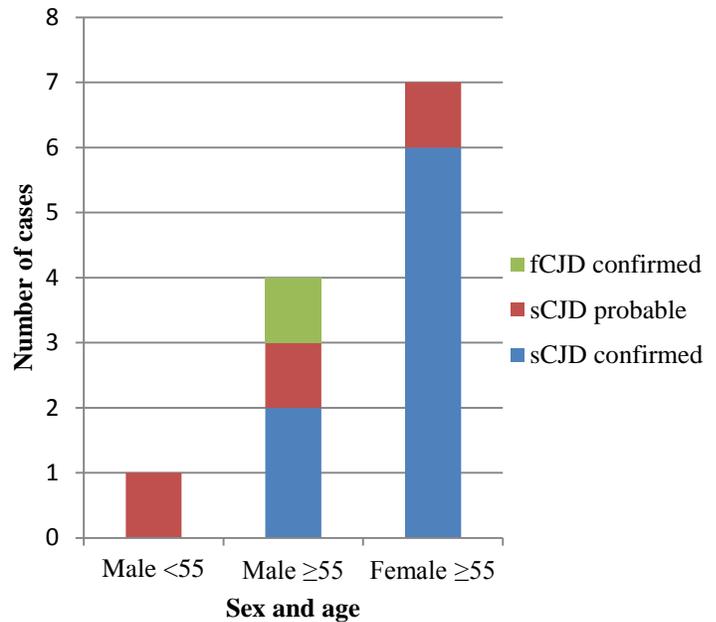


Table 1. CSF and confirmatory pathology testing in WA State prion disease deaths, 2015

Case	CSF test	Tau	14-3-3	RT-QuIC*	Biopsy	Autopsy	CJD type	Classification
1	Yes	Neg	Neg	Not done	No	Yes	Sporadic	Definite
2	Yes	Pos	Pos	Not done	No	Yes	Sporadic	Definite
3	Yes	Pos	Pos	Not done	No	Yes	Sporadic	Definite
4	Yes	Pos	Pos	Not done	No	Yes	Sporadic	Definite
5	Yes	Not done	Neg	Not done	Yes	Yes	Sporadic	Definite
6	Yes	Pos	Pos	Pos	No	Yes	Sporadic	Definite
7	Yes	Pos	Ambiguous	Not done	No	Yes	Familial	Definite
8	Yes	Neg	Pos	Not done	Yes	No	Sporadic	Definite
9	Yes	Pos	Pos	Pos	No	Yes	Sporadic	Definite
10	Yes	Not done	Pos	Not done	No	No	Sporadic	Probable
11	Yes	Pos	Pos	Pos	No	No	Sporadic	Probable
12	Yes	Pos	Pos	Pos	No	No	Sporadic	Probable

*RT-QuIC (Real Time Quaking-Induced Conversion) became available on April, 2015 at the NPDPC. It is performed as a reflex test following a positive 14-3-3 protein or Tau with a value of 500 pg/mL or higher.

CJD case definitions

For sporadic, familial, iatrogenic, and variant CJD case definitions please see:

<http://www.cdc.gov/prions/cjd/diagnostic-criteria.html>

<http://www.cdc.gov/prions/vcjd/diagnostic-criteria.html>