HUMAN PRION DISEASE ENHANCED SURVEILLANCE WASHINGTON STATE ANNUAL REPORT – 2023

DOH 420-161

Background

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.

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*.

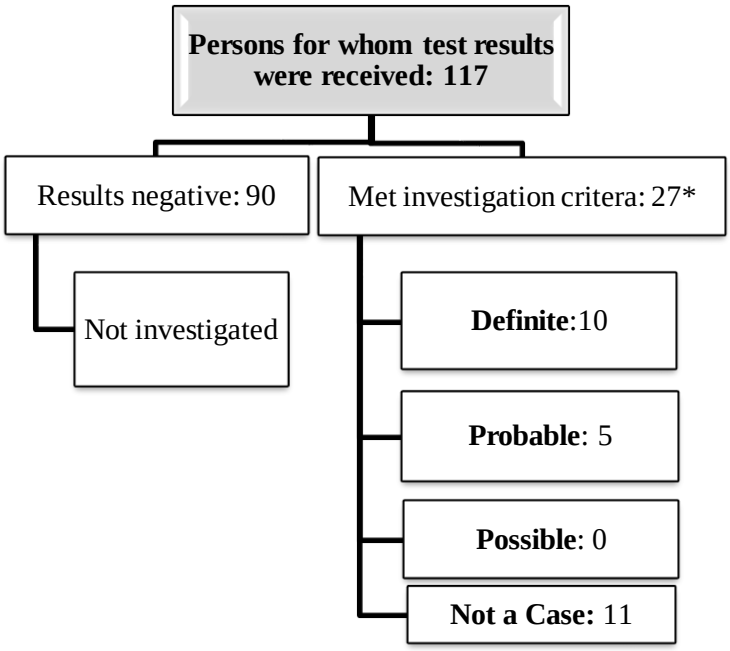
Iatrogenic transmission of the CJD agent (iCJD) has been linked to the use of contaminated human growth hormone (hGH), dura mater and corneal grafts, or neurosurgical equipment. All of the equipment-related cases occurred before the implementation of the routine sterilization procedures currently used 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 (RT-QuIC, 14-3-3 protein in cerebrospinal fluid, MRI, and EEG), can be used to make a clinical diagnosis of probable CJD.

Annual Summary

Prion disease test results for 117 individuals were received by DOH during 2023. Of these, 90 were not suggestive of prion disease. Follow-up investigation for 27 patients who met the criteria for further investigation revealed that 11 were diagnosed with a different condition, or on further investigation symptoms were inconsistent with CJD. The remaining 16 patients met the CDC criteria for definite or probable cases; one of these 16 cases died in 2024 and will be counted towards next year's report.

Washington prion disease surveillance overview, 2023



Prion deaths by county, 2023

County	N
Spokane	3
Benton-Franklin	1
Pacific	1
Pierce	4
King	1
Snohomish	1
Thurston	1
Whatcom	1
Clark	1
Yakima	1
Total	**15**

Characteristics of Washington CJD cases, 2023

Females comprised 53% of cases.

The median age at diagnosis was 69 years (range: 52-82).

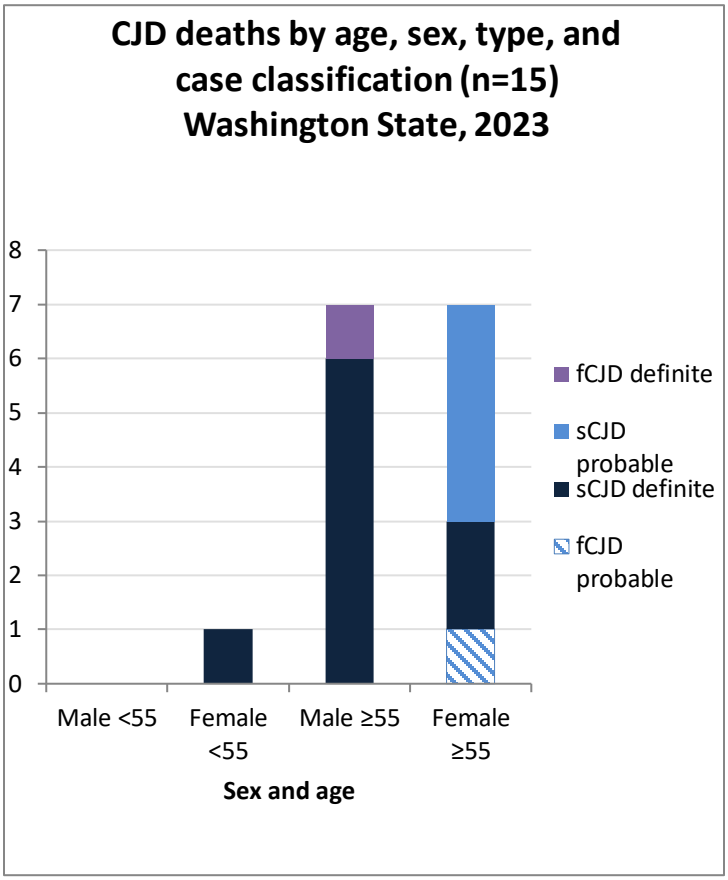
The median time between onset of symptoms and death was 2 months (range: 1-156 months).

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

No iatrogenic or variant CJD was reported.

The incidence of CJD in Washington State in 2022 was 1.89 cases/million population.

The average worldwide occurrence of CJD is approximately 1–2 cases/million population per year.



*A positive biopsy/autopsy result or a CSF test with a positive result for a RT-QuIC, or a highly elevated 14-3-3 or T-Tau, a positive test on anyone under 55 on any of the three diagnostic tests, or prion disease listed on their death certificate

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Table 1. CSF and confirmatory pathology testing in Washington State residents who died of prion disease, 2023

Case	CSF test	T-tau	14-3-3	RT-QuIC*	Autopsy	CJD type	Classification
1	NPDPSC	>20,000	124,692	Positive	No	Presumed sporadic	Probable
2	NPDPSC	>20,000	53,476	Positive	Yes	Sporadic	Definite
3	NPDPSC	>20,000	>160,000	positive	No	Presumed sporadic	Probable
4	NPDPSC	11,625	88,341	Positive	Yes	Sporadic	Definite
5	NPDPSC	27,111	151,437	Positive	No	Presumed sporadic	Probable
6	NPDPSC	>20,000	>160,000	Positive	Yes	Sporadic	Definite
7	NPDPSC	11,561	58,190	Positive	Yes	Familial	Definite
8	NPDPSC	2,821	26,658	Positive	Yes	Sporadic	Definite
9	NPDPSC	>20,000	>160,000	Positive	Yes	Sporadic	Definite
10	NPDPSC	14,414	47,858	Positive	Yes	Sporadic	Definite
11	None	N/A	N/A	N/A	Yes	Sporadic	Definite
12	NPDPSC	>20,000	115,296	Positive	No	Presumed sporadic	Probable
13	None	N/A	N/A	N/A	No	Presumed familial	Probable
14	NPDPSC	8,202	34,341	Positive	Yes	Sporadic	Definite
15	NPDPSC	>20,000	106,795	Positive	Yes	Sporadic	Definite

*RT-QuIC (Real Time Quaking-Induced Conversion) became available in April 2015 at NPDPSC. It was performed as a reflex test following a positive 14-3-3 protein or T-tau with a value of 500 pg/mL or higher 2015-2019. RT-QuIC is performed on all samples beginning January 2019.

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