Washington State Department of Health

Human Prion Disease

(Rare Disease of Public Health Significance)

LHJ Use
ID _____

Please enter the case in PHIMS as a Rare Disease of Public Health Significance and send/fax this form to Communicable Diseases Epidemiology

Investigator
_____

Phone/email
_____
_____

Investigation start date
___/___/_____

Investigation complete date
___/___/_____

Disease:
- **Creutzfeldt Jakob Disease (sporadic, familial, iatrogenic)** ☐
- **Variant Creutzfeldt Jakob Disease** ☐
- **Fatal Familial Insomnia** ☐
- **Gerstmann-Straussler-Scheinker Syndrome** ☐
- **Other**_____ ☐

County:

PHIMS ID:

REPORT SOURCE

Initial report date: ___/___/_____

Reporter (check all that apply)
- ☐ NPDPSC lab ☐ Hospital
- ☐ Public health agency
- ☐ Health Care Provider
- ☐ Other_____

OK to talk to surviving relative? ☐ Yes ☐ No ☐ DK

Date of interview ___/___/___

Reporter name _____ Phone_____

Primary Health Care provider _____ Phone_____

Neurologist_____ Phone_____

Infection control practitioner_____ Phone_____

Other _____

PATIENT INFORMATION

Name (last, first) _____

Address _____ ☐ Homeless

City/State/Zip _____

Next of kin: Name _____

Next of kin: Phone _____

☐ Spouse ☐ Parent ☐ Child ☐ Sibling ☐ Legal guardian

☐ Other relative _____ ☐ Other _____

Patient's primary occupation _____

Employer/worksite _____

Birth date ___/___/___ Age _____

Birthplace _____

Gender ☐ F ☐ M ☐ Other ☐ Unk

Ethnicity ☐ Hispanic or Latino
☐ Not Hispanic or Latino ☐ Unk

Race (check all that apply)
- ☐ Amer Ind/AK Native ☐ Asian
- ☐ Native HI/other PI ☐ Black/Afr Amer
- ☐ White ☐ Other ☐ Unk

CLINICAL INFORMATION

Onset date (mm/yy) ___/___ ☐ Derived Diagnosis date ___/___/___ Illness duration _____ days/months/years

Clinical Findings

First symptom(s) _____
_____

Y	N	DK	NA	
☐	☐	☐	☐	**Rapidly progressive dementia**
☐	☐	☐	☐	**Myoclonus**
☐	☐	☐	☐	**Visual abnormality**
				Hallucinations☐ Hemianopsia☐ Opsoclonus☐
				Blindness☐ Visual field cut/deficit☐ Diplopia☐
☐	☐	☐	☐	**Cerebellar signs**
				Ataxia☐ Movement tremor☐ Nystagmus☐
☐	☐	☐	☐	**Pyramidal signs**
				Spasticity☐ Hyperreflexia☐ Clonus☐
				Spastic paralysis☐ Babinksi's sign☐
				Upper motor neuron weakness☐ Hemiplegia☐
☐	☐	☐	☐	**Akinetic mutism**

Y	N	DK	NA	
☐	☐	☐	☐	**Extrapyramidal signs**
				Chorea☐ Dystonia☐ Bradykinesia/hypokinesia☐
				Tremor☐ Hypomimia☐ Shuffling gait ☐
				Rigidity☐ Ballismus/hemiballismus☐
				Choreoathetosis☐ Postural instability☐
☐	☐	☐	☐	**Progressive neuropsychiatric disorder**
☐	☐	☐	☐	**Early psychiatric symptom(s)**
				Delusions☐ Apathy☐ Anxiety☐
				Depression☐ Withdrawal☐
☐	☐	☐	☐	**Persistent painful sensory symptom(s)**
				Frank pain☐ Dysesthesia☐
☐	☐	☐	☐	**Neurodegenerative disease**

See Appendix A in the Guideline for signs/symptoms definition

Notes on clinical symptoms _____
_____

Answers are: Yes, No, Unknown to case, Not asked /Not answered

Case Name: _____

CSF laboratory findings

Test date ___/___/_____
CSF condition: Clear ☐ Bloody ☐ Other ☐_____

Y N DK NA
☐ ☐ ☐ ☐ **14-3-3 protein test**
☐ Positive ☐ Negative ☐ Ambiguous

☐ ☐ ☐ ☐ Tau protein test
☐ Positive ☐ Negative Amount _____

☐ ☐ ☐ ☐ RT-Quic
☐ Positive ☐ Negative ☐ Not performed

CSF estimated probability of prion disease: _____ %

Laboratory where CSF sent:
☐ Nat Prion Disease Path Surv Center (NPDPSC)
NPDPSC ID _____
☐ Other laboratory:_____

Brain Imaging Findings

Y N DK NA
☐ ☐ ☐ ☐ **EEG** performed Date___/___/_____
☐ ☐ ☐ ☐ EEG with **periodic sharp wave complexes**
EEG result notes_____
_____
_____

Y N DK NA
☐ ☐ ☐ ☐ **MRI** performed Date___/___/_____
☐ ☐ ☐ ☐ FLAIR/DWI* hyperintensities within the basal ganglia (caudated nucleus and/or putamen)
☐ ☐ ☐ ☐ Bilateral FLAIR hyperintensities involving the pulvinar thalamic nuclei (hockey stick sign)
MRI result notes _____
_____
_____
_____

**FLAIR: Fluid attenuated inversion recovery DWI: diffusion-weighted imaging.*

Brain Biopsy Findings

Y N DK NA
☐ ☐ ☐ ☐ Brain biopsy performed
Date of procedure___/___/_____

☐ ☐ ☐ ☐ Western Blot reveals PrPsc (PrP 27-30)
☐ ☐ ☐ ☐ Immunostaining with 3F4 reveals granular deposits
☐ ☐ ☐ ☐ Numerous widespread kuru-type amyloid plaques surrounded by vacuoles in both the cerebellum and cerebrum - florid plaques.
☐ ☐ ☐ ☐ Spongiform change and extensive prion protein deposition shown by immunohistochemistry throughout the cerebellum and cerebrum.

If sporadic CJD, subtype classification:
MM1/MV1 ☐ VV2 ☐ MV2 ☐
MM2 ☐ VV1 ☐ sFI ☐

If familial CJD, results of gene sequencing analysis:
_____

Laboratory where tissue sent:
☐ Nat Prion Disease Path Surv Center (NPDPSC)
NPDPSC ID _____
☐ Other laboratory:_____

Autopsy findings

Y N DK NA
☐ ☐ ☐ ☐ Brain autopsy performed
Date of autopsy___/___/___

☐ ☐ ☐ ☐ Western Blot reveals PrPsc (PrP 27-30)
☐ ☐ ☐ ☐ Immunostaining with 3F4 reveals granular deposits
☐ ☐ ☐ ☐ Numerous widespread kuru-type amyloid plaques surrounded by vacuoles in both the cerebellum and cerebrum - florid plaques.
☐ ☐ ☐ ☐ Spongiform change and extensive prion protein deposition shown by immunohistochemistry throughout the cerebellum and cerebrum.

If sporadic CJD, subtype classification:
MM1/MV1 ☐ VV2 ☐ MV2 ☐
MM2 ☐ VV1 ☐ sFI ☐

If familial CJD, results of gene sequencing analysis:
_____

Laboratory where tissue sent:
☐ Nat Prion Disease Path Surv Center (NPDPSC)
NPDPSC ID _____
☐ Other laboratory:_____

Alternative diagnosis

Y N DK NA
☐ ☐ ☐ ☐ Indication of an alternative, non-prion disease diagnosis (i.e., subarachnoid hemorrhage, encephalitis, stroke with acute infarction, multi-infarct dementia with acute infarction, brain neoplasm, paraneoplastic neurological disorder)

Notes
_____
_____
_____
_____
_____
_____
_____
_____

Hospitalization

Y N DK NA
☐ ☐ ☐ ☐ Hospitalized at least overnight for this illness

Hospital name/City _____
Admit date ___/___/___ Discharge date ___/___/___

Death Status

Y N DK
☐ ☐ ☐ Died from illness Date of death ___/___/_____
Age at death _____
County of death _____

☐ ☐ ☐ Autopsy Date of autopsy ___/___/_____

Answers are: Yes, No, Unknown to case, Not asked/Not answered

Case Name: _____

POSSIBLE SOURCES

Predisposing Factors

Y N DK NA
☐ ☐ ☐ ☐ Family history of prion disease in a first degree relative
☐ ☐ ☐ ☐ PrP gene mutation known

Exposures

Y N DK NA **Has the patient ever…**
☐ ☐ ☐ ☐ Spent 3 months or more in the U.K. since 1980?
☐ ☐ ☐ ☐ Lived outside the United States

Country_____ Specific Months/Year_____
Country_____ Specific Months/Year_____
Country_____ Specific Months/Year_____
Country_____ Specific Months/Year_____

Notes_____
_____
_____

Y N DK NA **Has the patient ever…**
☐ ☐ ☐ ☐ Received human-derived pituitary hormones (e.g., growth hormone)
Dates ___/___/_____ to ___/___/_____

☐ ☐ ☐ ☐ Received a dura mater or corneal allograft
Date ___/___/_____

☐ ☐ ☐ ☐ Had neurosurgery (on brain, spinal cord or eyes)
Date ___/___/_____
Anatomic Site_____
Hospital name/city _____
_____

☐ ☐ ☐ ☐ Consumed venison from deer/elk
Dates of consumption, if known:___/___/___ ___/___/____
_____
_____
_____

CASE CLASSIFICATION

Sporadic CJD: ☐ Definite ☐ Probable ☐ Possible
Iatrogenic CJD: ☐ Definite ☐ Probable
Familial CJD: ☐ Definite ☐ Probable
Variant CJD: ☐ Definite ☐ Probable ☐ Possible

☐ Other prion disease
_____
☐ Prion disease unlikely
☐ Cannot be determined/provider report only

Source(s) of Patient History

☐ Chart review ☐ Health care provider interview ☐ Patient interview
☐ Relative/friend interview Name _____ Relationship _____

PUBLIC HEALTH ISSUES

Y N DK NA
☐ ☐ ☐ ☐ Case donated organs or tissues
Date___/___/_____
Facility _____
Organs/tissues donated_____
☐ ☐ ☐ ☐ Case underwent neurosurgery or eye surgery
Date___/___/_____
Facility _____
Procedure_____

PUBLIC HEALTH ACTIONS

Y N DK NA
☐ ☐ ☐ ☐ Autopsy/Biopsy discussed with medical provider (if notification occurred before patient's death)
☐ ☐ ☐ ☐ Infection control measures discussed with facility(s) ICP (to be done in all cases)
☐ ☐ ☐ ☐ Blood/tissue/organ program notified
Date___/___/_____
☐ ☐ ☐ ☐ Surgical facility notified
Date___/___/_____
☐ ☐ ☐ ☐ Report sent to CDC
Date___/___/_____

NOTES

Answers are: Yes, No, Unknown to case, Not asked/Not answered

Annex 1. Human Prion Diseases - Local Health Jurisdictions Investigation Checklist Year: _____

LHJ: _____ Investigator:_____ Phone number:_____

Date received report: / / Date follow-up started: / / Date closed: / /

Case status: ☐ Definite ☐ Probable ☐ Possible ☐ Not a case ☐ Died

Classification: ☐Sporadic ☐Iatrogenic ☐Familial ☐Variant ☐ Unknown ☐ < 55 years old

Name: _____DOB: _____ / _____ / _____ Age: _____ PHIMS ID: _____

	Name	Agency or Facility	Phone	Fax
Ordering Physician				
Neurologist				
Infection Control				
Other:				

Initial record-keeping

☐ Enter in PHIMS ☐ Start case report form

☐ *If suspect variant or iatrogenic CJD, novel prion disease, cluster of cases, or age < 55: Immediately notify DOH.*

Date DOH notified: ___ / ____/ _____ ☐ If age <55, fill CDC form

Investigation

☐ Verify patient's county of residence

☐ Collect clinical information (with ICP and/or neurologist and/or primary health care provider)

☐Clinical presentation ☐Predisposing factors ☐Infection risk to others

☐Ask ICP/provider/facility to fax medical records (Include Neurology/Psychiatry consultations, EEGs, and MRIs within one year)

☐Request to be notified when patient dies

☐Determine if case meets criteria for possible, probable, or confirmed case

☐ Provide information to medical provider

Recommended biopsy/autopsy (confirmatory diagnosis) to provider?

☐Yes ☐No → why not? _____

Infection control information provided?

☐Yes, discussed on phone ☐Yes, provided print materials via email/letter

☐No → why not? _____

☐ Determine whether patient died, and:

☐Confirm whether autopsy performed ☐Obtain death certificate and verify that prion disease is listed as a cause of death

☐Obtain autopsy results (final results can take up to 8 weeks)

☐ Notify DOH (possible, probable, or definite case) within 7 days of investigation

Final record-keeping

☐Complete CRF and this form and fax to DOH ☐Complete PHIMS entry ☐Complete CDC form and fax to DOH, if applicable