State of California—Health and Human Services Agency

Local ID Number: _____

California Department of Public Health
Center for Infectious Diseases
Division of Communicable Disease Control
Infectious Diseases Branch
Surveillance and Statistics Section
MS 7306, P.O. Box 997377
Sacramento, CA 95899-7377

CREUTZFELDT-JAKOB DISEASE AND PRION DISEASE CASE REPORT

PATIENT INFORMATION

Last Name	*First Name*	*Middle Name*	*Suffix*

Social Security Number (9 digits)	*DOB (mm/dd/yyyy)*	*Age*	
			☐ Years ☐ Months ☐ Days

Address Number & Street – Residence	*Apartment / Unit Number*

City / Town	*State*	*Zip Code*

Census Tract	*County of Residence*	*Country of Residence*

Country of Birth	*If not U.S. Born - Date of Arrival in U.S. (mm/dd/yyyy)*

Home Telephone	*Cellular Phone / Pager*	*Work / School Telephone*

E-mail Address	*Other Electronic Contact Information*

Work / School Location	*Work / School Contact*

Gender
☐ Female ☐ Trans female / transwoman ☐ Genderqueer or non-binary ☐ Unknown
☐ Male ☐ Trans male / transman ☐ Identity not listed ☐ Declined to answer

Pregnant? ☐ Yes ☐ No ☐ Unknown	*If Yes, Est. Delivery Date (mm/dd/yyyy)*

Medical Record Number	*Patient's Parent/Guardian Name*

Occupation Setting (see list on page 7)	*Other Describe/Specify*

Occupation (see list on page 7)	*Other Describe/Specify*

Primary Language
☐ English
☐ Spanish
☐ Other: _____

Ethnicity (check one)
☐ Hispanic/Latino
☐ Non-Hispanic/Non-Latino
☐ Unknown

Race(s)
(check all that apply, race descriptions on page 6)

The response to this item should be based on the patient's self-identity or self-reporting. Therefore, patients should be offered the option of selecting more than one racial designation.

☐ American Indian or Alaska Native

☐ Asian *(check all that apply, see list on page 6)*
- ☐ Asian Indian
- ☐ Bangladeshi
- ☐ Cambodian
- ☐ Chinese
- ☐ Filipino
- ☐ Hmong
- ☐ Indonesian
- ☐ Japanese
- ☐ Korean
- ☐ Laotian
- ☐ Malaysian
- ☐ Pakistani
- ☐ Sri Lankan
- ☐ Taiwanese
- ☐ Thai
- ☐ Vietnamese
- ☐ Other: _____

☐ Black or African-American

☐ Native Hawaiian or Other Pacific Islander *(check all that apply, see list on page 6)*
- ☐ Native Hawaiian
- ☐ Samoan
- ☐ Fijian
- ☐ Tongan
- ☐ Guamanian
- ☐ Other: _____

☐ White

☐ Other: _____

☐ Unknown

ADDITIONAL PATIENT DEMOGRAPHICS

Sex Assigned at Birth
☐ Female ☐ Unknown
☐ Male ☐ Declined to answer

Sexual Orientation
☐ Heterosexual or straight ☐ Questioning, unsure, or patient doesn't know ☐ Declined to answer
☐ Gay, lesbian, or same-gender loving ☐ Orientation not listed ☐ Unknown
☐ Bisexual

First three letters of patient's last name: | | | |

CLINICAL INFORMATION

RESIDENCE INFORMATION

Patient's Residence at Time of Diagnosis	*City*	*State*	*State in which Patient is Receiving Care*
Where is patient currently located? (e.g., facility name, family member living with, etc.)		*Known Date at This Location (mm/dd/yyyy)*	

DIAGNOSIS INFORMATION

Onset Date (mm/dd/yyyy)	*Date of CJD Diagnosis (mm/dd/yyyy)*		
Name of Hospital where CJD Diagnosis was Made	*Location*	*Diagnosing Physician's Name*	*Telephone Number*
Was the patient seen by a neurologist? ☐ Yes ☐ No ☐ Unknown	*Neurologist's Name*	*Address*	*Telephone Number*
Was diagnosis of CJD made by a neurologist? ☐ Yes ☐ No ☐ Unknown	*If No, Specialty of Diagnosing Physician*		
Other Significant Illnesses			

HOSPITALIZATION

Did the patient visit the emergency room for illness? ☐ Yes ☐ No ☐ Unknown

Was the patient hospitalized? ☐ Yes ☐ No ☐ Unknown	*If Yes, how many total hospital nights?*	*During any part of the hospitalization, did the patient stay in an intensive care unit (ICU) or a critical care unit (CCU)?* ☐ Yes ☐ No ☐ Unknown

If there were any ER visits or hospital stays related to this illness, specify details in the Hospitalization – Details section below.

HOSPITALIZATION – DETAILS

Hospital Name 1	*Street Address*			*Admit Date (mm/dd/yyyy)*	
	City			*Discharge / Transfer Date (mm/dd/yyyy)*	
	State	*Zip Code*	*Telephone Number*	*Medical Record Number*	*Discharge Diagnosis*
Hospital Name 2	*Street Address*			*Admit Date (mm/dd/yyyy)*	
	City			*Discharge / Transfer Date (mm/dd/yyyy)*	
	State	*Zip Code*	*Telephone Number*	*Medical Record Number*	*Discharge Diagnosis*

OUTCOME

Outcome? ☐ Survived ☐ Died ☐ Unknown	*If Survived,* Survived as of _____(mm/dd/yyyy)	*Date of Death (mm/dd/yyyy)*

If Died, specify:	*State in which Death Occurred*	
	Is CJD listed as a cause of death on the death certificate? ☐ Yes ☐ No ☐ Unknown	*If No, what was the primary cause of death on certificate?*

LABORATORY INFORMATION

LABORATORY RESULTS SUMMARY

EEG performed? ☐ Yes ☐ No ☐ Unknown	*If Yes, specify results*		
MRI performed? ☐ Yes ☐ No ☐ Unknown	*If Yes, specify results*		
CSF tests? ☐ Yes ☐ No ☐ Unknown	*If Yes, specify below.*		
	CSF Lab Report # 1 Date (mm/dd/yyyy)	*Was blood found in the sample?* ☐ Yes ☐ No ☐ Unknown	*CSF Results #1* 14-3-3 protein: ☐ Positive ☐ Negative ☐ Ambiguous; Tau protein: ☐ Positive ☐ Negative; RT-QuIC: ☐ Positive ☐ Negative
	CSF Lab Report # 2 Date (mm/dd/yyyy)	*Was blood found in the sample?* ☐ Yes ☐ No ☐ Unknown	*CSF Results #2* 14-3-3 protein: ☐ Positive ☐ Negative ☐ Ambiguous; Tau protein: ☐ Positive ☐ Negative; RT-QuIC: ☐ Positive ☐ Negative
	CSF specimens sent to the National Prion Disease Pathology Surveillance Center (NPDPSC)? ☐ Yes ☐ No ☐ Unknown		*If No, which laboratory?*
Brain biopsy performed? ☐ Yes ☐ No ☐ Unknown	*If Yes, specify below.*		
	Hospital where Biopsy Performed		*Date of Biopsy (mm/dd/yyyy)*
	Specimens sent to NPDPSC? ☐ Yes ☐ No ☐ Unknown	*Western Blot* ☐ Abnormal prion protein present ☐ Abnormal prion protein NOT present	*Immunohistochemistry* ☐ Positive ☐ Negative
	Diagnosis ☐ CJD ☐ Sporadic CJD ☐ Variant CJD ☐ Familial CJD ☐ Other (specify): _____		
Autopsy performed? ☐ Yes ☐ No ☐ Unknown	*If Yes, specify below.*		
	Date of Autopsy (mm/dd/yyyy)	*Hospital where Autopsy Performed*	*Autopsy Physician Name*
	Specimens sent to NPDPSC? ☐ Yes ☐ No ☐ Unknown	*Western Blot* ☐ Abnormal prion protein present ☐ Abnormal prion protein NOT present	*Immunohistochemistry* ☐ Positive ☐ Negative
	Diagnosis ☐ CJD ☐ Sporadic CJD ☐ Variant CJD ☐ Familial CJD ☐ Other (specify): _____		
Other Tests (e.g., CSF results, PCR results, etc.)			

EPIDEMIOLOGIC INFORMATION

SURVEILLANCE INSTRUCTIONS

Questions about reporting a case can be directed to the California Emerging Infections Program - CJD Surveillance Project at (510) 451-1344 or https://ceip.us/projects/cjd/.

EXPOSURES / RISK FACTORS

DID THE PATIENT UNDERGO ANY OF THE FOLLOWING PROCEDURES?

Procedure	Yes	No	Unk	If Yes, Specify as Noted	
Brain surgery				*Year(s)*	*Hospital / Location*
Spinal surgery				*Year(s)*	*Hospital / Location*
Eye surgery				*Year(s)*	*Hospital / Location*
Received dura mater allograft				*Year(s)*	*Hospital / Location*

(continued on page 4)

Procedure	Yes	No	Unk	If Yes, Specify as Noted	
Received corneal allograft				*Year(s)*	*Hospital / Location*
Received human derived pituitary growth hormone				*Year(s)*	*Hospital / Location*
RECEIVED a blood transfusion				*Date(s) (mm/dd/yyyy)*	*Hospital / Location*
DONATED blood				*Date(s) (mm/dd/yyyy)*	*Hospital / Location*
DONATED cells/tissues/organs				*Date(s) (mm/dd/yyyy)*	*Hospital / Location*
Other (specify)					

DID THE PATIENT HAVE ANY OF THE FOLLOWING EXPOSURES?

Exposure	Yes	No	Unk	If Yes, Specify as Noted	
HUNTED deer or elk				*Area(s) hunted*	*Year(s)*
Knowingly ATE deer or elk meat				*Source of meat*	*Year(s)*
History of definite or probable case of prion disease in a blood relative				*Relationship to patient*	*Name of disease*
Other (specify)					

TRAVEL HISTORY

Did patient live or travel outside of the U.S. (including military service) between 1980 - 1996? ☐ Yes ☐ No ☐ Unknown	*If Yes, specify all locations and dates below.*

TRAVEL HISTORY – DETAILS

Travel Type	State	Country	Other location details (city, resort, etc.)	Date Travel Started *(mm/dd/yyyy)*	Date Travel Ended *(mm/dd/yyyy)*
☐ Domestic ☐ Unknown ☐ International					
☐ Domestic ☐ Unknown ☐ International					
☐ Domestic ☐ Unknown ☐ International					

NOTES

REPORTING AGENCY

Investigator Name	*Local Health Jurisdiction*	*Telephone Number*	*Date (mm/dd/yyyy)*

First Reported By

☐ Clinician ☐ Laboratory ☐ Other (specify):_____

EPIDEMIOLOGICAL LINKAGE

Epi-linked to known case? ☐ Yes ☐ No ☐ Unknown	*Contact Name / Case Number*
	Link Type ☐ Family ☐ Surgery ☐ Other (specify): _____

DISEASE CASE CLASSIFICATION

Disease Type

☐ Sporadic CJD ☐ Variant CJD ☐ Other Prion Disease (specify):_____

☐ Iatrogenic CJD ☐ Familial Prion Disease (specify):_____

SUPPORTING DOCUMENTATION

Documentation Attached

☐ Hospital discharge summary ☐ MRI report ☐ CSF test results ☐ Brain biopsy report

☐ Autopsy report ☐ Neurologist report / notes ☐ EEG report

STATE USE ONLY

State Case Classification

☐ Definite ☐ Probable ☐ Possible ☐ Not a case ☐ Need additional information

CASE DEFINITION

CREUTZFELDT-JAKOB DISEASE (2018)

CASE CLASSIFICATION

1. **Sporadic CJD**

Definite:

Diagnosed by standard neuropathological techniques; and/or immunocytochemically; and/or Western blot confirmed protease-resistant PrP; and /or presence of scrapie-associated fibrils.

Probable:

- Neuropsychiatric disorder plus positive RT-QuIC in cerebrospinal fluid (CSF) or other tissues

OR

- Rapidly progressive dementia; **and** at least two out of the following four clinical features:
  1. Myoclonus
  2. Visual or cerebellar signs
  3. Pyramidal/extrapyramidal signs
  4. Akinetic mutism

AND a positive result on at least one of the following laboratory tests:

- A typical EEG (periodic sharp wave complexes) during an illness of any duration
- A positive 14-3-3 cerebrospinal fluid (CSF) assay in patients with a disease duration of less than 2 years
- High signal in caudate/putamen on magnetic resonance imaging (MRI) brain scan or at least two cortical regions (temporal, parietal, occipital) either on diffusion-weighted imaging (DWI) or fluid attenuated inversion recovery (FLAIR)

AND without routine investigations indicating an alternative diagnosis.

Possible:

- Progressive dementia; and at least two out of the following four clinical features:
  1. Myoclonus
  2. Visual or cerebellar signs
  3. Pyramidal/extrapyramidal signs
  4. Akinetic mutism

AND the absence of a positive result for any of the four laboratory tests that would classify a case as "probable"

AND duration of illness less than two years

AND without routine investigations indicating an alternative diagnosis.

2. **Iatrogenic CJD**

Progressive cerebellar syndrome in a recipient of human cadaveric-derived pituitary hormone; or sporadic CJD with a recognized exposure risk, e.g., antecedent neurosurgery with dura mater implantation.

3. **Familial CJD**

Definite or probable CJD **plus** definite or probable CJD in a first degree relative; and/or Neuropsychiatric disorder **plus** disease-specific PrP gene mutation.

(continued on page 6)

CASE DEFINITION (continued)

4. **Variant CJD (vCJD)**

Definite vCJD:

Neuropathologic examination of brain tissue is required to confirm a diagnosis of variant CJD. The following confirmatory features should be present.

a. Numerous widespread kuru-type amyloid plaques surrounded by vacuoles in both the cerebellum and cerebrum – florid plaques.
b. Spongiform change and extensive prion protein deposition shown by immunohistochemistry throughout the cerebellum and cerebrum.

Suspected vCJD:

a. Current age or age at death <55 years (a brain autopsy is recommended, however, for all physician-diagnosed CJD cases).
b. Psychiatric symptoms at illness onset and/or persistent painful sensory symptoms (frank pain and/or dysesthesia).
c. Dementia, and development ≥4 months after illness onset of at least two of the following five neurologic signs: poor coordination, myoclonus, chorea, hyperreflexia, or visual signs. (If persistent painful sensory symptoms exist, ≥4 months delay in the development of the neurologic signs is not required).
d. A normal or an abnormal EEG, but not the diagnostic EEG changes often seen in classic CJD.
e. Duration of illness of over 6 months.
f. Routine investigations of the patient do not suggest an alternative, non-CJD diagnosis.
g. No history of receipt of cadaveric human pituitary growth hormone or a dura mater graft.
h. No history of CJD in a first degree relative or prion protein gene mutation in the patient.

NOTE

1. If a patient has the typical bilateral pulvinar high signal on MRI scan, a suspected diagnosis of variant CJD requires the presence of a progressive neuropsychiatric disorder, d, e, f and g of the above criteria, and four of the following five criteria: 1) early psychiatric symptoms (anxiety, apathy, delusions, depression, withdrawal); 2) persistent painful sensory symptoms (frank pain and/or dysesthesia); 3) ataxia; 4) myoclonus or chorea or dystonia; and 5) dementia.
2. A history of possible exposure to bovine spongiform encephalopathy (BSE) such as residence or travel to a BSE-affected country after 1980 increases the index of suspicion for a variant CJD diagnosis.

RACE DESCRIPTIONS

Race	Description
American Indian or Alaska Native	Patient has origins in **any** of the original peoples of North and South America (including Central America).
Asian	Patient has origins in **any** of the original peoples of the Far East, Southeast Asia, or the Indian subcontinent (e.g., including Bangladesh, Cambodia, China, India, Indonesia, Japan, Korea, Malaysia, Nepal, Pakistan, the Philippine Islands, Thailand, and Vietnam).
Black or African American	Patient has origins in **any** of the black racial groups of Africa.
Native Hawaiian or Other Pacific Islander	Patient has origins in **any** of the original peoples of Hawaii, Guam, American Samoa, or other Pacific Islands.
White	Patient has origins in **any** of the original peoples of Europe, the Middle East, or North Africa.

ASIAN GROUPS

- Bangladeshi
- Bhutanese
- Burmese
- Cambodian
- Chinese
- Filipino
- Hmong
- Indian
- Indonesian
- Iwo Jiman
- Japanese
- Korean
- Laotian
- Madagascar
- Malaysian
- Maldivian
- Nepalese
- Okinawan
- Pakistani
- Singaporean
- Sri Lankan
- Taiwanese
- Thai
- Vietnamese

NATIVE HAWAIIAN AND OTHER PACIFIC ISLANDER GROUPS

- Carolinian
- Chamorro
- Chuukese
- Fijian
- Guamanian
- Kiribati
- Kosraean
- Mariana Islander
- Marshallese
- Melanesian
- Micronesian
- Native Hawaiian
- New Hebrides
- Palauan
- Papua New Guinean
- Pohnpeian
- Polynesian
- Saipanese
- Samoan
- Solomon Islander
- Tahitian
- Tokelauan
- Tongan
- Yapese

First three letters of patient's last name: | | | |

OCCUPATION SETTING

- Childcare/Preschool
- Correctional Facility
- Drug Treatment Center
- Food Service
- Health Care - Acute Care Facility
- Health Care - Long Term Care Facility
- Health Care - Other
- Homeless Shelter
- Laboratory
- Military Facility
- Other Residential Facility
- Place of Worship
- School
- Other

OCCUPATION

- Agriculture - farmworker or laborer (crop, nursery, or greenhouse)
- Agriculture - field worker
- Agriculture - migratory/seasonal worker
- Agriculture - other/unknown
- Animal - animal control worker
- Animal - farm worker or laborer (farm or ranch animals)
- Animal - veterinarian or other animal health practitioner
- Animal - other/unknown
- Clerical, office, or sales worker
- Correctional facility - employee
- Correctional facility - inmate
- Craftsman, foreman, or operative
- Daycare or child care attendee
- Daycare or child care worker
- Dentist or other dental health worker
- Drug dealer
- Fire fighting or prevention worker
- Flight attendant
- Food service - cook or food preparation worker
- Food service - host or hostess
- Food service - waiter or waitress
- Food service - other/unknown
- Homemaker
- Laboratory technologist or technician
- Laborer - private household or unskilled worker
- Manager, official, or proprietor
- Manicurist or pedicurist
- Medical - emergency medical technician or paramedic
- Medical - health care worker
- Medical - medical assistant
- Medical - pharmacist
- Medical - physician assistant or nurse practitioner
- Medical - physician or surgeon
- Medical - registered nurse
- Medical - other/unknown
- Military - officer
- Military - recruit or trainee
- Protective service - police officer
- Protective service - other
- Professional, technical, or related profession
- Retired
- Sex worker
- Student - preschool or kindergarten
- Student - elementary or middle school
- Student - high (secondary) school
- Student - college or university
- Student - other/unknown
- Teacher/employee - preschool or kindergarten
- Teacher/employee - elementary or middle school
- Teacher/employee - high (secondary) school
- Teacher/instructor/employee - college or university
- Teacher/instructor/employee - other/unknown
- Unemployed - seeking employment
- Unemployed - not seeking employment
- Unemployed - other/unknown
- Other
- Refused
- Unknown