

## Key Findings

Creutzfeldt-Jakob disease (CJD) is a rare disease of the brain and nervous system in humans. CJD is part of a group of diseases called prion diseases. In these diseases, prions (proteins that are naturally found in the body) change shape, build up in the brain, and destroy brain and nerve tissue, causing nervous system disorders. These disorders include rapid onset dementia, personality changes, memory problems, impaired vision, difficulty walking, moving, and speaking, all of which get worse over time and ultimately lead to death. There are different types of CJD based on the way the disease is caused.

### Creutzfeldt-Jakob Disease in California from 2013 through 2019

**Total Cases:** There were a total of 221 new CJD cases from 2013 through 2019, with 25 to 36 cases reported per year. Of these cases, 172 (78%) were reported to have died with CJD.

- **By County:** Cases of CJD were reported from 30 counties in California. About 1 out every 3 cases was reported in Los Angeles County (40 cases) and San Diego County (37 cases).
- **By Sex:** The number of CJD cases in males (112 cases) was similar to the number in females (108 cases).
- **By Age Group:** More cases of CJD were reported in people aged 65 to 74 years (82 cases) and 55 to 64 years (66 cases) than in other age groups.
- **By Race/Ethnicity:** For cases where race and ethnicity information was available, the highest percentage of cases was in people who reported non-Hispanic White race/ethnicity (about 68%).

CJD is a complicated and difficult disease to diagnose. A confirmed diagnosis of CJD can only be made by examining brain tissue during an autopsy. There is no cure for CJD, and treatment is used only to help manage symptoms or make the patient more comfortable. No specific therapy has been shown to stop the symptoms of CJD from getting worse over time.

For more information about CJD, please visit the [U.S. Centers for Disease Control and Prevention CJD website](#). For details about key infectious diseases in California, please visit the [CDPH Surveillance and Statistics Section webpage](#).

## Background

Transmissible spongiform encephalopathies (TSE) are a group of fatal, rare, progressive neurological disorders that occur in humans and animals. TSE are also referred to as prion diseases.<sup>1</sup> Prions are proteins that occur naturally in the body but have a higher concentration in the brain and nerve tissue. The most common TSE affecting humans is Creutzfeldt-Jakob disease (CJD). Disease occurs when a normal prion in the brain changes shape, causing other normal prions nearby to change. These abnormal prions build up, causing brain and nerve tissue destruction and damage. This leads to progressive dementia, neuromuscular disorders, and death.

CJD is classified into four subtypes: sporadic (also known as classic CJD), familial, iatrogenic, and variant.<sup>2</sup> All forms of CJD are characterized by neurological and psychiatric problems that become progressively worse. However, the different CJD subtypes have distinguishing features including: the age distribution of affected patients, duration of illness, clinical presentation, and the pathology of brain tissue. Sporadic CJD (sCJD) is the most common form of CJD. It is unknown what causes the spontaneous change of normal prions to abnormal prions leading to disease. It typically affects people aged 60 years or older. Once symptoms appear, sCJD progresses very quickly and is usually fatal within a few months of symptom onset. Familial CJD (fCJD) is an inherited condition and accounts for 10-15% of all cases. Symptoms are similar to those of sCJD. Often fCJD is diagnosed at an earlier age, around 50 years, and the course of illness is generally longer than sCJD. Iatrogenic CJD (iCJD) is the inadvertent transmission of CJD through medical or surgical procedures. The signs and symptoms of iCJD often look like sporadic CJD. The age at onset depends on the age at exposure and route of exposure. Variant CJD (vCJD) is believed to occur primarily through the consumption of beef infected with Bovine Spongiform Encephalopathy (BSE), commonly known as “Mad Cow Disease”. Initial symptoms of vCJD are typically characterized as psychiatric or behavioral changes, and painful sensory symptoms. For patients with vCJD, age of onset is usually less than 55 years of age, and the duration of illness is greater than one year.<sup>2</sup>

vCJD is the name of the disease in humans that have acquired it through eating beef that had BSE. Mad Cow Disease is the name of the prion disease that occurs in cattle. sCJD is the most common type of CJD and is not the same as vCJD or Mad Cow Disease.

There is no evidence of person-to-person transmission of CJD. In very rare circumstances CJD has been acquired iatrogenically. Documented transmission has been linked to the use of contaminated growth hormone prepared from human pituitary glands, dura mater, corneal grafts, and contaminated neurosurgical instruments.<sup>3</sup>

Symptoms of CJD can be similar to other rapid onset neurological disorders. There is currently no single diagnostic test for CJD. When a physician suspects CJD, the first concern is to rule out treatable forms of dementia. Physicians suspect CJD on the basis of the typical signs and symptoms and progression of the disease. CJD causes unique changes in brain tissue; the only way to confirm a diagnosis of CJD is by brain autopsy.<sup>4</sup>

This report describes the epidemiology of confirmed and probable CJD cases in California from 2013 through 2019. Due to multiple factors that can contribute to underreporting, data in this report are likely underestimates of actual disease incidence. For a complete

discussion of the definitions, methods, and limitations associated with this report, please refer to the *Technical Notes*.<sup>5</sup>

## California Reporting Requirements and Surveillance Case Definition

California Code of Regulations (CCR), Title 17, Section 2500 requires health care providers to report suspected cases of CJD to their local health department within seven calendar days of identification.<sup>6</sup>

California regulations require cases of CJD to be reported to the California Department of Public Health (CDPH). CDPH counted cases that satisfied the U.S. Centers for Disease Control and Prevention diagnostic criteria of a confirmed and probable case. During the surveillance period (2013-2019), a confirmed case of classic or sporadic CJD was defined as a case meeting at least one of the following criteria: diagnosed by standard neuropathological techniques; and/or immunocytochemically; and/or Western blot confirmed protease-resistant PrP; and/or presence of scrapie-associated fibrils. A probable case was defined as a case with one of the following scenarios: (1) neuropsychiatric disorder and positive RT-QuIC in CSF or other tissues, or (2) rapidly progressive dementia and at least two out of the following four clinical features:

- Myoclonus
- Visual or cerebellar signs
- Pyramidal/extrapyramidal signs
- Akinetic mutism

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

- Typical EEG (periodic sharp wave complexes) during an illness of any duration
- Positive 14-3-3 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.

A case of iatrogenic CJD was defined as 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.

A case of familial CJD was defined as definite or probable CJD plus definite or probable CJD in a first degree relative; and/or neuropsychiatric disorder plus disease-specific PrP gene mutation.<sup>7</sup>

A confirmed case of variant CJD was defined as a case with confirmation by brain biopsy or autopsy; confirmatory features include:

- 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.

A probable case of variant CJD was defined as:

- Age < 55 years at presentation/death
- Psychiatric symptoms at onset and/or persistent painful sensory symptoms
- Dementia, and development  $\geq 4$  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,  $\geq 4$  months delay in the development of the neurologic signs is not required)
- A normal or an abnormal EEG, but not characteristic of classic or sporadic CJD
- Duration of illness > 6 months
- Routine investigations of the patient do not suggest an alternative, non-CJD diagnosis
- No history of receipt of cadaveric human pituitary growth hormone or a dura mater graft
- No history of CJD in a first degree relative or prion protein gene mutation in the patient
- Bilateral pulvinar sign on MRI in the presence of above criteria
- History of residence or travel to a BSE-affected country after 1980 increases the index of suspicion.<sup>8</sup>

## Epidemiology of Creutzfeldt-Jakob Disease in California, 2013-2019

CDPH received reports of 221 total cases of CJD with estimated symptom onset dates from 2013 through 2019. The highest number of cases was reported in 2017 (36 cases) and the fewest number was reported in 2015 (25 cases). CJD case counts remained relatively stable throughout the surveillance period [Figure 1]. During the surveillance period, 172 (77.8%) case-patients were reported to have died with CJD.

Statewide from 2013 through 2019, cases of CJD were reported from 30 counties in California, and 5 counties reported at least 1 case of CJD for each year of the surveillance period: Los Angeles, Orange, San Bernardino, San Diego, and Santa Clara. Of all CJD cases in California, 34.8% occurred in two counties: Los Angeles County (40 cases) and San Diego County (37 cases) [Figure 2].

From 2013 through 2019, the number of cases among males (112 cases) was similar to the number of cases among females (108 cases); 50.9% of CJD case-patients were male and 49.1% were female.

By age group, case-patients were aged 25 to 34 years (1 case), 35 to 44 years (4), 45 to 54 years (21), 55 to 64 years (66), 65 to 74 (82), 75 and 85 years (38 years), and 85 years and older (9). No cases of CJD were reported in those aged less than 25 years.

For CJD cases with complete race/ethnicity data (see *Technical Notes*), cases reported non-Hispanic White race/ethnicity more frequently than would be expected compared to the percentage of this population in California during the same time period (67.8% vs. 38.0%, respectively) [Figure 3].

**Figure 1. Creutzfeldt-Jakob Disease Incidence Rates by Year of Estimated Illness Onset, California, 2013-2019**

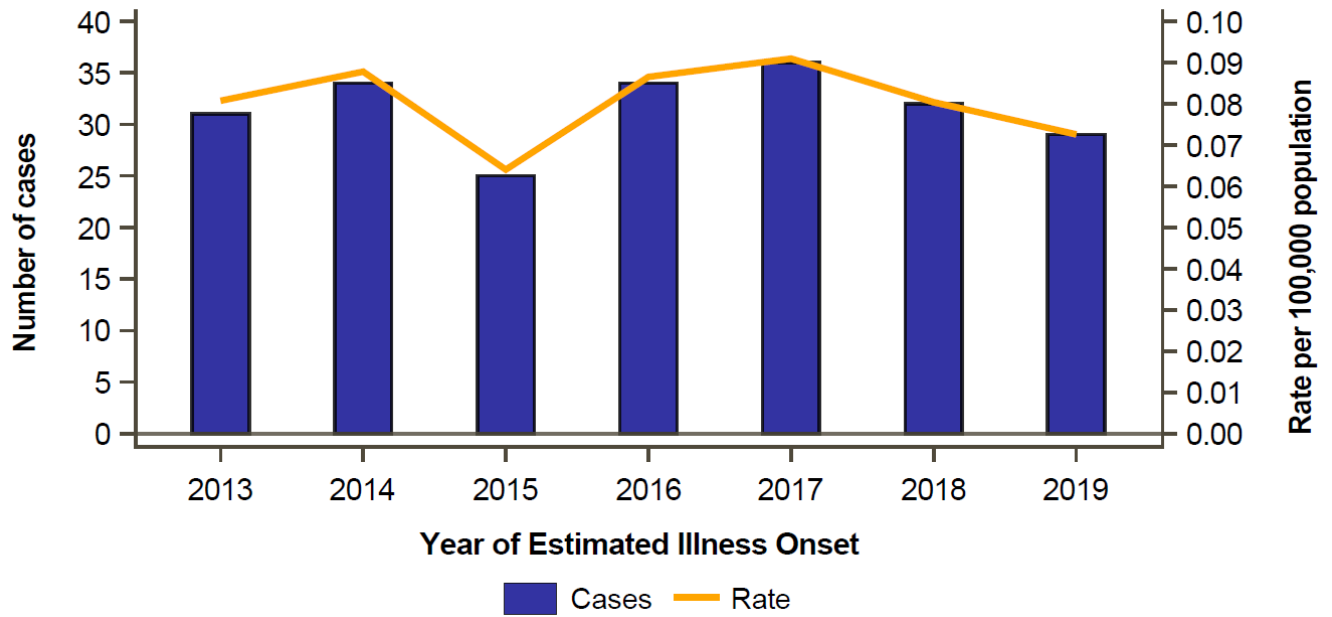
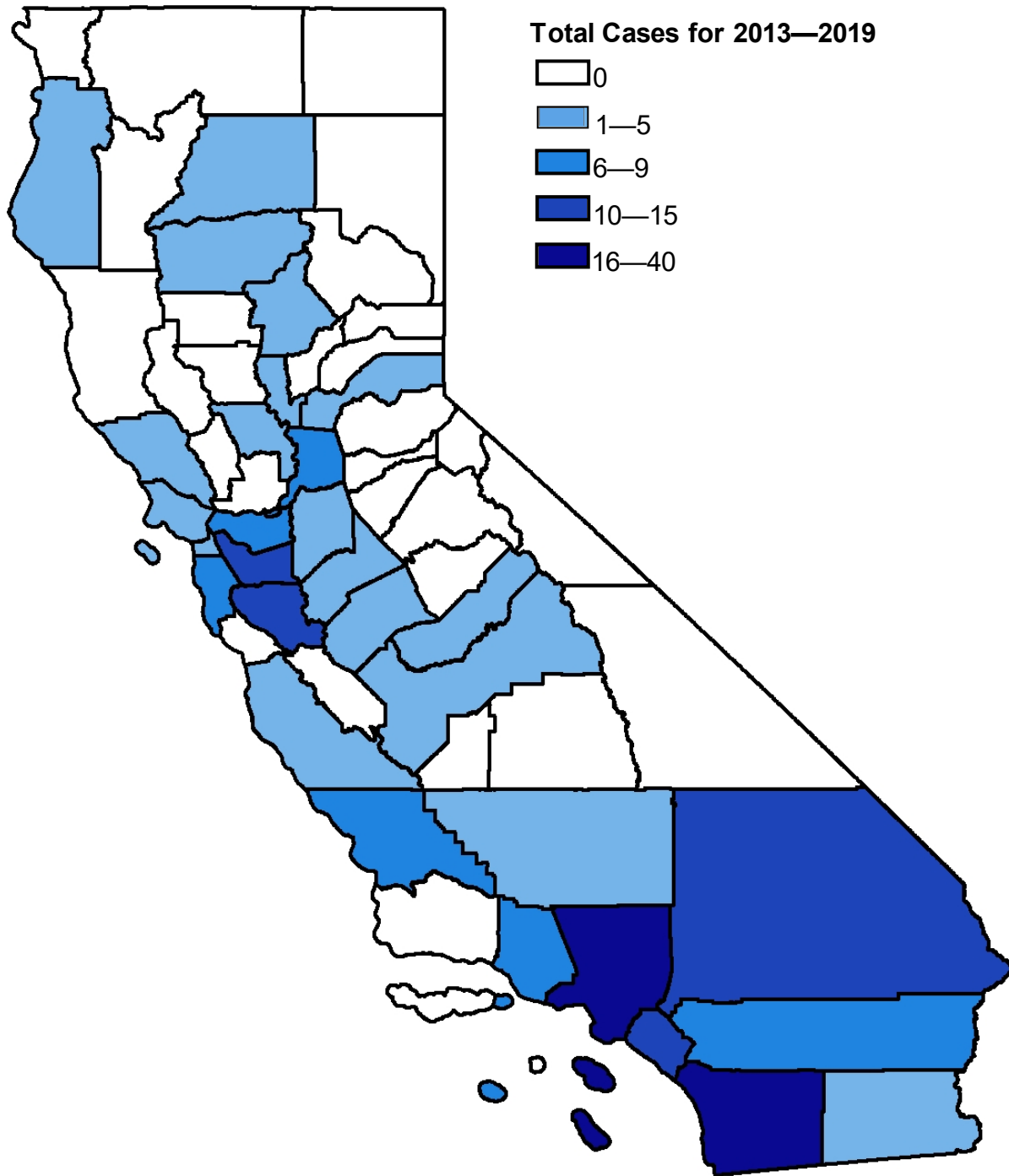
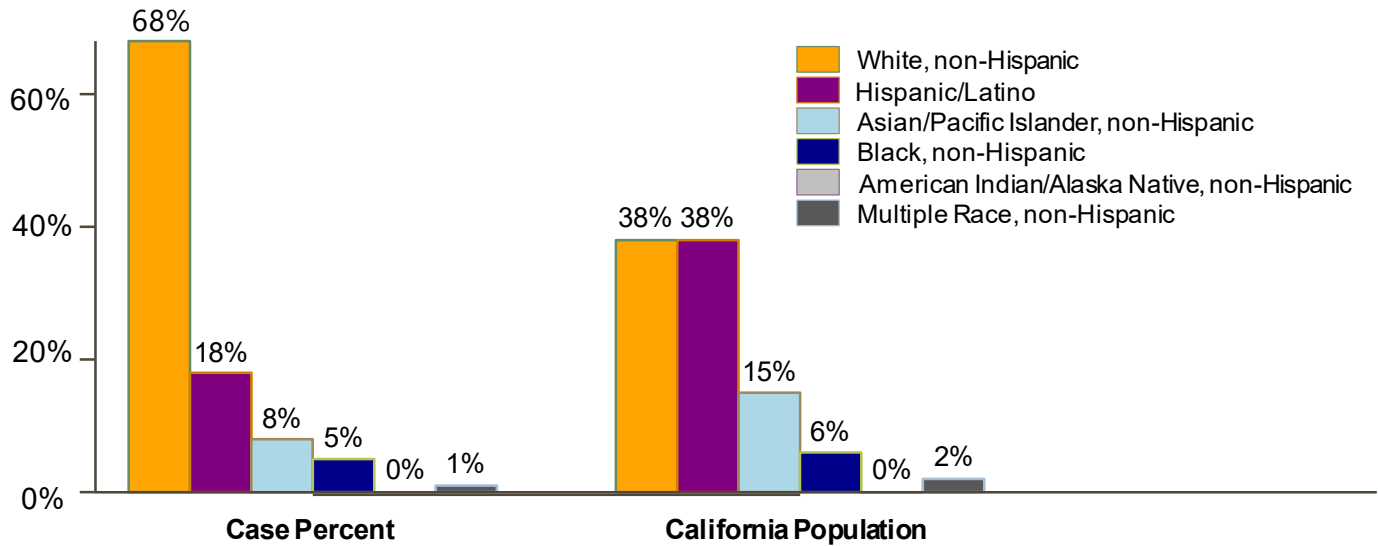


Figure 2. Creutzfeldt-Jakob Disease Total Case Count by County, California, 2013-2019



**Figure 3. Creutzfeldt-Jakob Disease Cases and Population by Race/Ethnicity, California, 2013-2019**



21.3% (n=47) of reported incidents of Creutzfeldt-Jakob Disease did not identify race/ethnicity and 1.4% (n=3) of incidents identified as 'Other' race/ethnicity and are not included in the Case Percent calculation. Information presented with a large percentage of missing data should be interpreted with caution.

## Comments

CJD is a complicated and difficult disease to diagnose. A confirmed diagnosis of CJD can only be made by examination of brain tissue on autopsy. There is no cure for CJD; treatment is aimed at alleviating symptoms. No specific therapy has been shown to stop disease progression. As such, CJD is always fatal. From 2013 through 2019 in California, 77.8% of case-patients (172 cases) were reported to have died with CJD.

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## References

<sup>1</sup> [Prion Diseases. U.S. Centers for Disease Control and Prevention website.](https://www.cdc.gov/prions/index.html) Updated November 17, 2021. Accessed December 8, 2021. <https://www.cdc.gov/prions/index.html>

<sup>2</sup> [Creutzfeldt-Jakob Disease \(CJD\) Background Information. California Emerging Infections Program website.](http://ceip.us/projects/cjd/background-information/) Accessed December 16, 2020. <http://ceip.us/projects/cjd/background-information/>

<sup>3</sup> [Creutzfeldt-Jakob Disease, Classic \(CJD\). Infection Control. U.S. Centers for Disease Control and Prevention website.](https://www.cdc.gov/prions/cjd/infection-control.html) Updated November 17, 2021. Access December 8, 2021. <https://www.cdc.gov/prions/cjd/infection-control.html>

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<sup>4</sup> [Creutzfeldt-Jakob Disease, Classic \(CJD\). About CJD. U.S. Centers for Disease Control and Prevention website.](https://www.cdc.gov/prions/cjd/about.html) Updated October 18, 2021. Accessed December 8, 2021.  
<https://www.cdc.gov/prions/cjd/about.html>

<sup>5</sup> State of California, Department of Public Health. [Technical notes. In: \*Epidemiologic Summaries of Selected Communicable Diseases in California, 2013-2019\*.](https://www.cdph.ca.gov/Programs/CID/DCDC/CDPH%20Document%20Library/EpiSummariesTechnicalNotes2013-2019.pdf) Sacramento, CA; 2021. Accessed December 30, 2021.  
<https://www.cdph.ca.gov/Programs/CID/DCDC/CDPH%20Document%20Library/EpiSummariesTechnicalNotes2013-2019.pdf>

<sup>6</sup> [Reportable Diseases and Conditions: Reporting to the Local Health Authority, 17 CCR § 2500 \(2021\).](https://govt.westlaw.com/calregs/Document/I5849DB60A9CD11E0AE80D7A8DD0B623B)  
<https://govt.westlaw.com/calregs/Document/I5849DB60A9CD11E0AE80D7A8DD0B623B>

<sup>7</sup> [Creutzfeldt-Jakob Disease, Classic \(CJD\). Diagnostic Criteria. U.S. Centers for Disease Control and Prevention website.](https://www.cdc.gov/prions/cjd/diagnostic-criteria.html) Updated January 24, 2019. Accessed December 16, 2020.  
<https://www.cdc.gov/prions/cjd/diagnostic-criteria.html>

<sup>8</sup> [Variant Creutzfeldt-Jakob Disease \(vCJD\). Diagnostic Criteria. U.S. Centers for Disease Control and Prevention website.](https://www.cdc.gov/prions/vcjd/diagnostic-criteria.html) Updated October 18, 2021. Accessed December 14, 2021.  
<https://www.cdc.gov/prions/vcjd/diagnostic-criteria.html>