

## Creutzfeldt Jakob Disease (CJD)

*CJD is a Class B Disease and must be reported to the state within one business day.*

Creutzfeldt Jakob disease (CJD) was first described by Creutzfeldt and Jakob in the early 1920s. This disease occurs worldwide at a very low rate. It is one of a group of sub-acute degenerative diseases of the brain caused by prions.

In the mid-1980s an epidemic of bovine spongiform encephalopathy (BSE), another prion-related infection, killed nearly 200,000 heads of cattle in Great Britain. A human version of BSE causing an encephalopathy resembling CJD, was named new variant CJD or vCJD. This disease was initially limited to the United Kingdom, but has since been identified in 202 persons from 11 countries. Three cases have been identified in the United States, but each of these infections was acquired overseas (2 in the UK, 1 in Saudi Arabia).

The United States has been widely spared due to an early ban placed on the importation of British sheep and goats in the early 1950s and on British cattle in the late 1980s.

### Symptoms

Dementia (memory loss, mood changes, and judgement errors) is always present and is often the first manifestation of the disease. Patients lose interest, become apathetic or irritable, experience sleep disorders, intellectual decline and disorientation. They may also have tremors, disturbances of gait, stance and loss of motor control. As the disease progresses, the patient may experience hallucinations, delusional ideas and confusion. In some patients, the cerebellar and visual abnormalities (even cortical blindness), predominate. At the end of the illness, patients are mute, stuporous, spastic and rigid. The disease rapidly progresses to death within six months. Less than 10% of patients have an illness that lasts up to three years.

The generalized slowing of EEG waves observed at the beginning of the illness is replaced by distinctive repetitive sharp waves which become bilateral and synchronous. The regular rate of the waves found in CJD is not observed in dementia due to other causes such as Alzheimers or Binswanger sub-cortical encephalo-pathy. CT scans are, on the other hand, usually normal. As the disease progresses, CT scans and MRI show rapid development of bilateral cortical atrophy.

CJD may be mistaken for Alzheimer's disease with myoclonus, multi-infarct dementia, alcoholic or nutritional deficiency syndromes or brain tumors. However, the presence of cerebellar involvement, typical EEG changes and rapid deterioration over a few months, secures the diagnosis of CJD. Confirmation is made on the typical histological pattern of spongiform encephalopathy.

### Epidemiology

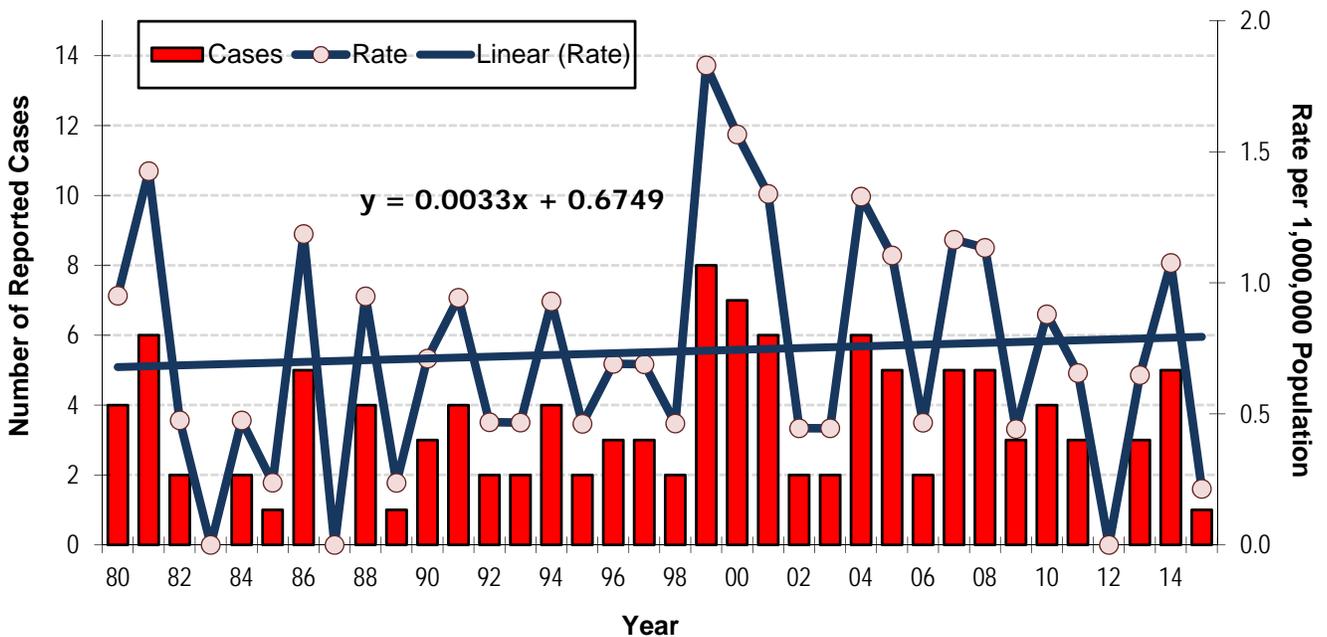
Some populations seem to have a higher incidence than others: incidence rates calculated in a few countries show a range from 0.3 to one per million per year, with an average of 0.9 per million in the United States.

### CJD in Louisiana

Surveillance of CJD in Louisiana is based on death certificates and hospital admission data since the disease is universally fatal in a few months. Recently, positive lab results became available; this reporting led to an increase in reported cases. A small proportion, (approximately 20%) of cases, are confirmed by an autopsy. Given the characteristic clinical picture, a pre-mortem clinical diagnosis would be reliable. In 75% of the cases, CJD is listed as the primary cause of death.

The overall incidence in Louisiana is 0.81 cases per million per year, very close to the worldwide average of one per million per year. Incidence rates remained stable from 1980 to 2003 (Cochran Armitage trend test  $\chi^2 = 1.50, p=0.22$ ). After 2003, rates started to increase steadily. Overall the trend is increasing (Cochran Armitage trend test  $\chi^2 = 6.10, p=0.001$ ). This increase is probably the consequence of better reporting due to laboratory tests being reported and triggering case investigations (Figure 1)

Figure 1: Number of reported cases of CJD and new report rates per million - Louisiana, 1980-2015



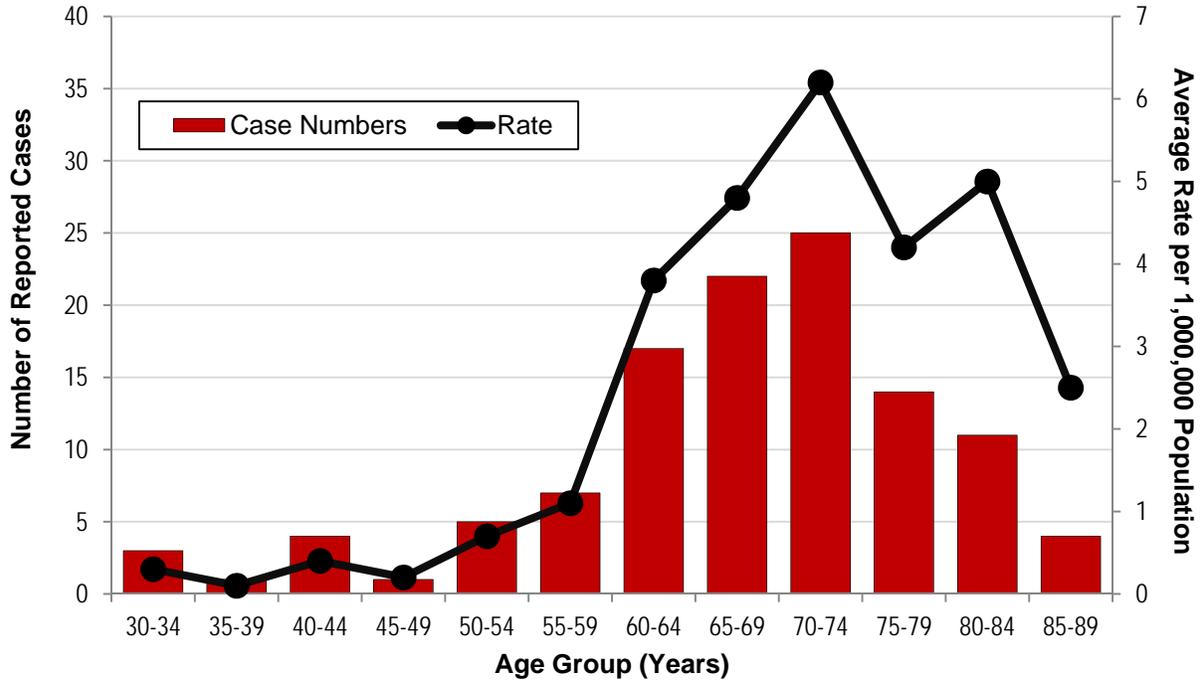
### Sex

There is a slight non-significant predominance of female cases: 55 females (56%) versus 51 males (44%) for a population where females represent 52% of the total population. This difference is similar by age group.

## Age Distribution

Ages ranged from 34 to 88 years. Incidence rates remained low until age 59 (less than 1.0 per million). From ages 60 to 74 years, rates increase to a high of 6.0 per million, then decrease back to 2.5 per million. It is possible that in the oldest age groups, due to the high prevalence of dementias, the specific diagnosis of CJD is not made (Figure 2).

Figure 2: Age group distribution of CJD Cases – Louisiana, 1980-2015



## Geographical Distribution

Cases are scattered throughout the state with no discernable pattern (Table and Figures 3 and 4)

Table: Number of cases of CJD per parish by decade\*- Louisiana, 1980 - 2015

Parish	1980-89	1990-99	2000-09	2010-15	Total
Acadia	0	1	2	0	3
Allen	0	1	0	0	1
Ascension	2	0	1	0	3
Avoyelles	0	1	0	1	2
Bienville	0	0	1	0	1
Bossier	0	1	1	0	2
Caddo	2	3	1	0	6
Calcasieu	1	1	1	0	3
East Baton Rouge	3	3	3	0	9
East Feliciana	0	1	0	0	1
Evangeline	1	0	0	0	1
Franklin	1	0	0	0	1
Iberia	0	0	2	1	3
Jefferson	4	9	1	1	15
Jefferson Davis	0	0	1	0	1
LaSalle	0	0	0	1	1
Lafayette	1	6	0	1	8
Lafourche	0	1	0	0	1
Livingston	0	1	1	0	2
Orleans	3	2	0	4	9
Ouachita	1	0	1	0	2
Plaquemines	1	0	0	0	1
Rapides	1	0	0	0	1
Red river	0	0	0	1	1
Richland	1	0	0	0	1
Sabine	1	0	0	0	1
St. Bernard	1	0	0	0	1
St. Helena	0	0	1	0	1
St. James	1	0	0	0	1
St. Landry	1	1	2	0	4
St. Martin	0	1	0	0	1
St. Mary	0	0	1	0	1
Tangipahoa	0	1	3	2	6
Union	1	0	1	0	2
Vermilion	1	0	0	0	1
Washington	1	2	0	0	3
Winn	0	1	0	0	1
<b>Total</b>	<b>29</b>	<b>37</b>	<b>24</b>	<b>12</b>	<b>102</b>

\*Only parishes with cases are listed here

Figure 3: Number of CJD cases – Louisiana, 1980 - 2015

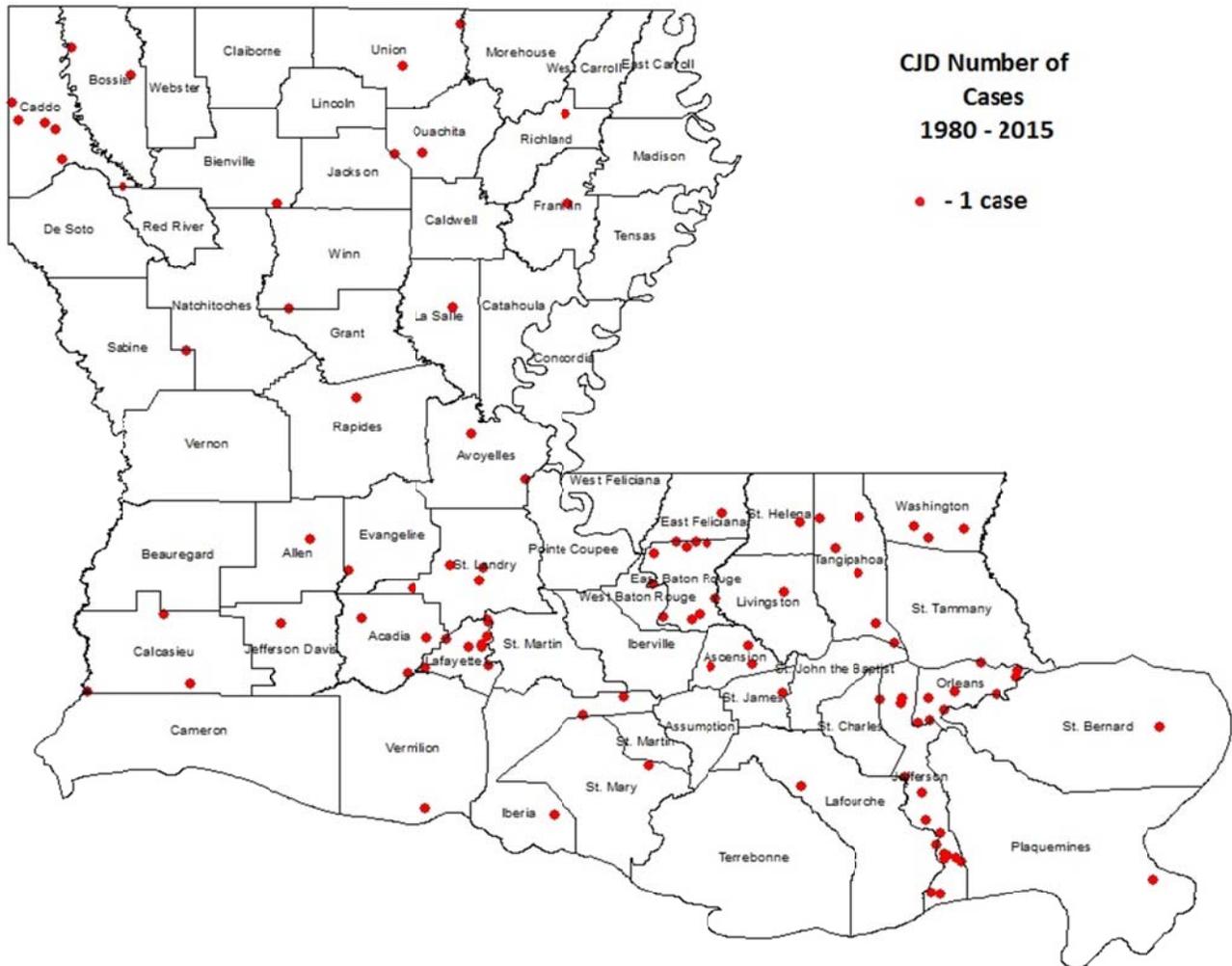


Figure 4: Incidence cases of CJD per million per year – Louisiana, 1980 - 2015

