

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 eleven 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, 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 ten percent 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 encephalopathy. 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.

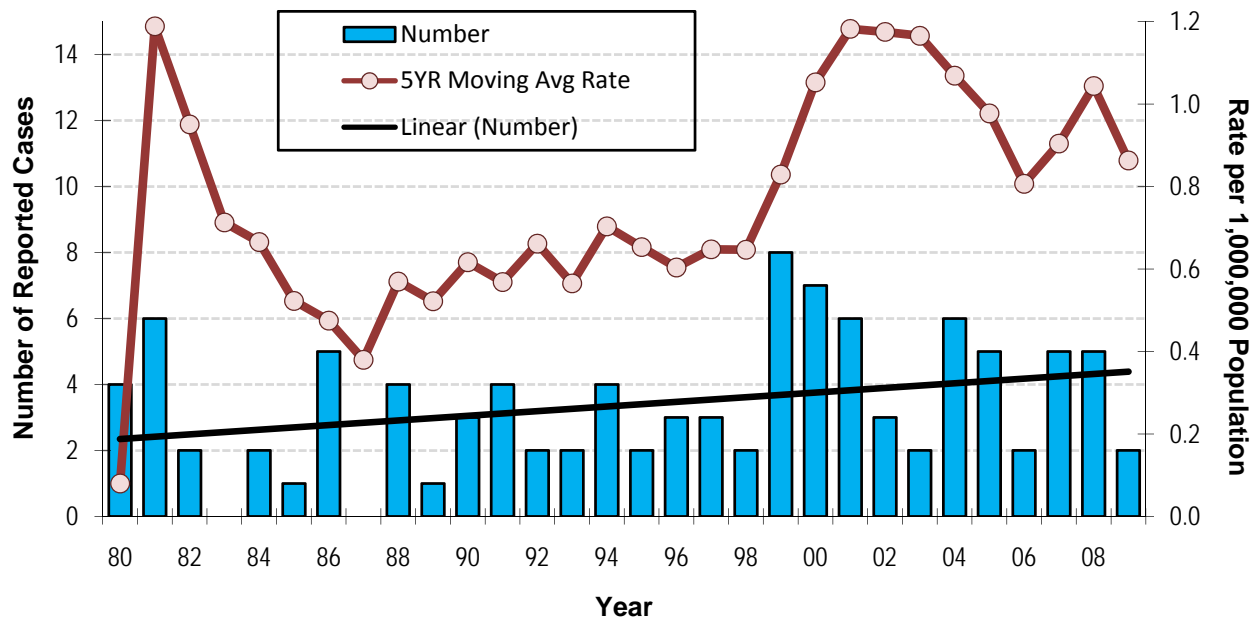
CJD in Louisiana

Surveillance of CJD in Louisiana is based on death certificates and hospital admission data since the disease is universally fatal in a few months. A small proportion (approximately 20%) of cases are confirmed by an autopsy. However, given the characteristic clinical picture, a pre-mortem clinical diagnosis would be reliable. In seventy-five percent of the cases, CJD is listed as the primary cause of death.

The overall incidence in Louisiana is 0.78 cases per million per year, very close to the worldwide average of one per million per year. The trend-line shows no increase with time. (Figure 1)

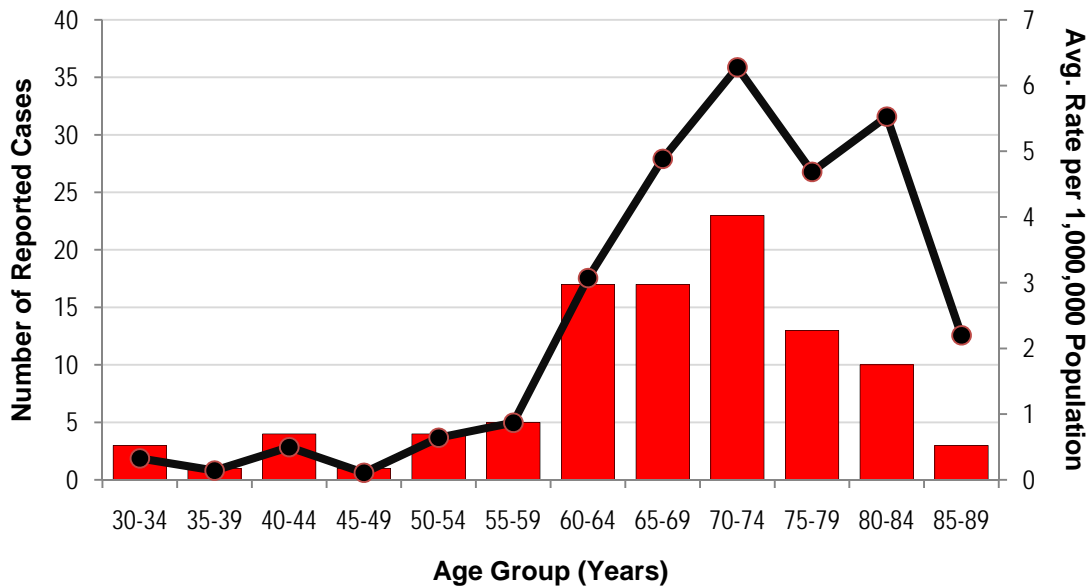
Figure 1: Number of reported cases and five year moving average per million of CJD

Louisiana, 1980-2009



Age Distribution

Ages ranged from thirty-four to eighty-eight years, with most cases occurring among ages seventy to seventy-four years. Age specific incidence is highest among those individuals sixty-five and older (3.4 cases per million). Only eight percent of cases occurred in persons under forty-five years of age (Figure 2).

Figure 2: Age group distribution of CJD Cases – Louisiana, 1980-2009

Gender and Race

Males and females were almost equally affected (46% male, 54% female).

There was distinct racial disparity with only eight percent of cases among African-Americans (with the African-American population representing only 26% of the population of the state). This disparity is likely to be due to lack of diagnosis and reporting among African-American cases.

As expected, the highest number of cases are reported in the large urban parishes. It is probable that number of cases from large urban centers are higher than those of the rural areas due to better reporting (Table).

Table: Incidence of CJD by parish per 1,000,000 - Louisiana, 1980-2009

Parish	2009		1980-2009		Parish	2009		1980-2009	
	Cases	Rates	Total Cases	Average Rate		Cases	Rates	Total Cases	Average Rate
Acadia	0	0.0	3	1.8	Madison	0	0	0	0.0
Allen	0	0.0	1	1.5	Morehouse	0	0	0	0.0
Ascension	0	0.0	3	1.7	Natchitoches	0	0	0	0.0
Assumption	0	0.0	0	0.0	Orleans	0	0	6	0.4
Avoyelles	0	0.0	1	0.9	Ouachita	0	0	3	0.7
Beauregard	0	0.0	0	0.0	Plaquemines	0	0	1	1.4
Bienville	1	63.3	1	2.3	Pointe Coupee	0	0	0	0.0
Bossier	0	0.0	2	0.4	Rapides	0	0	2	0.3
Caddo	0	0.0	6	0.9	Red river	0	0	0	0.0
Calcasieu	0	0.0	3	0.6	Richland	0	0	1	1.7
Caldwell	0	0.0	0	0.0	Sabine	0	0	2	1.6
Cameron	0	0.0	0	0.0	St Bernard	0	0	1	0.5
Catahoula	0	0.0	0	0.0	St Charles	0	0	0	0.0
Claiborne	0	0.0	0	0.0	St Helena	0	0	2	3.4
Concordia	0	0.0	0	0.0	St James	0	0	1	1.7
De Soto	0	0.0	0	0.0	St John	0	0	0	0.0
East Baton Rouge	1	2.2	10	0.8	St Landry	0	0	5	1.7
East Carroll	0	0.0	0	0.0	St Martin	0	0	1	0.8
East Feliciana	0	0.0	2	1.7	St Mary	0	0	1	0.7
Evangeline	0	0.0	1	1.1	St Tammany	0	0	3	0.0
Franklin	0	0.0	2	1.6	Tangipahoa	0	0	5	1.5
Grant	0	0.0	0	0.0	Tensas	0	0	0	0.0
Iberia	0	0.0	2	1.0	Terrebonne	0	0	0	0.0
Iberville	0	0.0	0	0.0	Union	0	0	2	3.3
Jackson	0	0.0	0	0.0	Vermilion	0	0	2	0.7
Jefferson	0	0.0	16	1.1	Vernon	0	0	0	0.0
Jefferson Davis	0	0.0	1	1.1	Washington	0	0	3	2.5
LaSalle	0	0.0	0	0.0	Webster	0	0	0	0.0
Lafayette	0	0.0	7	1.4	West Baton Rouge	0	0	0	0.0
Lafourche	0	0.0	1	0.4	West Carroll	0	0	0	0.0
Lincoln	0	0.0	1	0.0	West Feliciana	0	0	1	0.0
Livingston	0	0.0	3	1.1	Winn	0	0	1	2.0

In summary, the age rates and rural/urban distribution patterns of CJD in Louisiana fall within the range of observed rates and distribution in the United States. Therefore, it is reasonable to conclude that the surveillance based on death certificates represent fairly accurately, the pattern of CJD in the state.