Creutzfeldt-Jakob Disease (CJD)

CJD is a Class C Disease and must be reported to the state within five business days.

Creutzfeldt-Jakob disease (CJD) was first described by Creutzfeldt and Jakob in the early 1920s. This disease occurs worldwide at a very low rate, though advances in laboratory detection technology have led to an increase in reporting. It is one of a group of sub-acute degenerative diseases of the brain caused by prions, and part of a group of conditions known as Transmissible Spongiform Encephalopathies (TSE's).

TSEs include; bovine spongiform encephalopathy in cattle; scrapie in sheep and goats; chronic wasting disease in cervids; and Creutzfeldt-Jacob disease and other human prion diseases, including variant Creutzfeldt-Jacob disease (vCJD) the human form of "mad cow disease") in people. The infection is believed to be caused by abnormal proteins called prions, which are thought to cause damage to other normal prion proteins that can be found in tissues throughout the body, but most often in the brain and spinal cord, leading to brain damage and development of prion diseases.

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 U.S., but each of these infections was acquired overseas (two in the UK, one in Saudi Arabia).

The U.S. 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.

CJD Symptoms

Dementia (memory loss, mood changes, judgment 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 Alzheimer's 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.

CJD Epidemiology

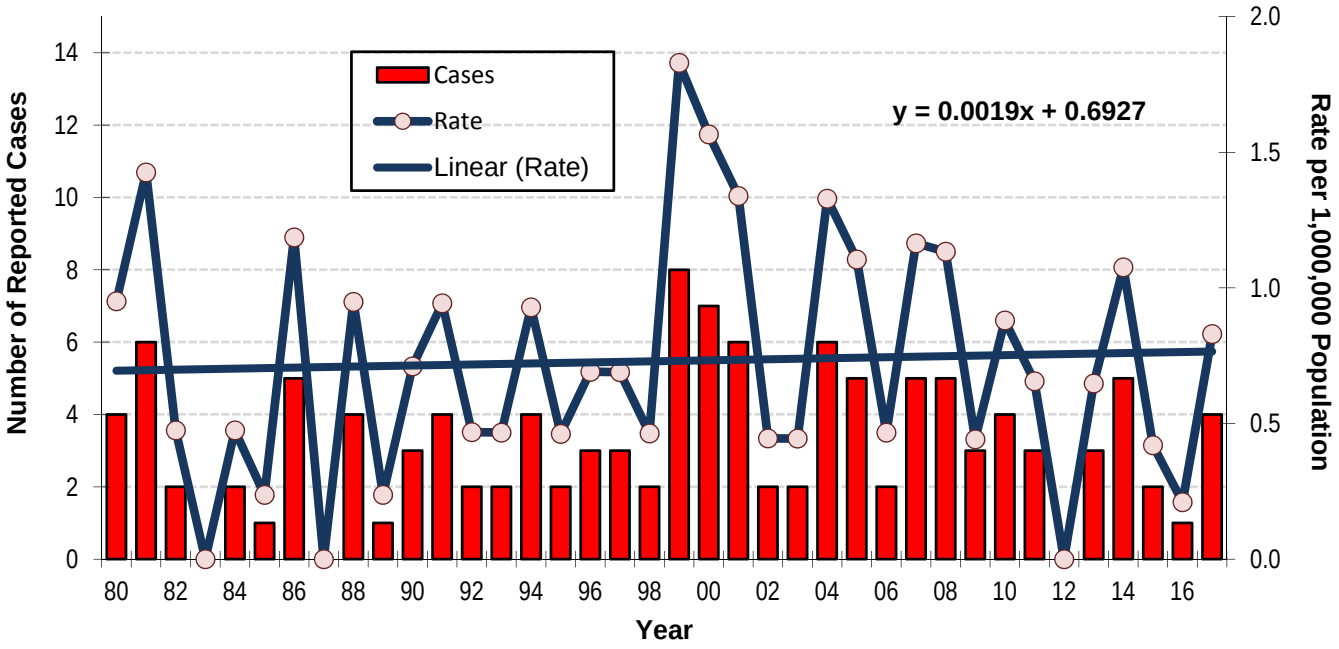
Some populations seem to have a higher incidence than others: incidence rates calculated in a few countries show a range from 0.3 to one per million per year, with an average of 0.9 per million in the United States.

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 (through the National Prion Disease Pathology Surveillance Center) which 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 (Figure 1). The incidence rates remained stable from 1980 to 2003 (Cochran Armitage trend test χ^2 =1.50, p=0.22). After 2003, rates started to increase steadily. Overall the trend is increasing (Cochran Armitage trend test χ^2 =6.10, p=0.0.01). This increase is probably the consequence of better laboratory test reporting and the triggering of case investigations.

Figure 1: Number of reported cases of CJD and new report rates per million population
Louisiana, 1980-2017



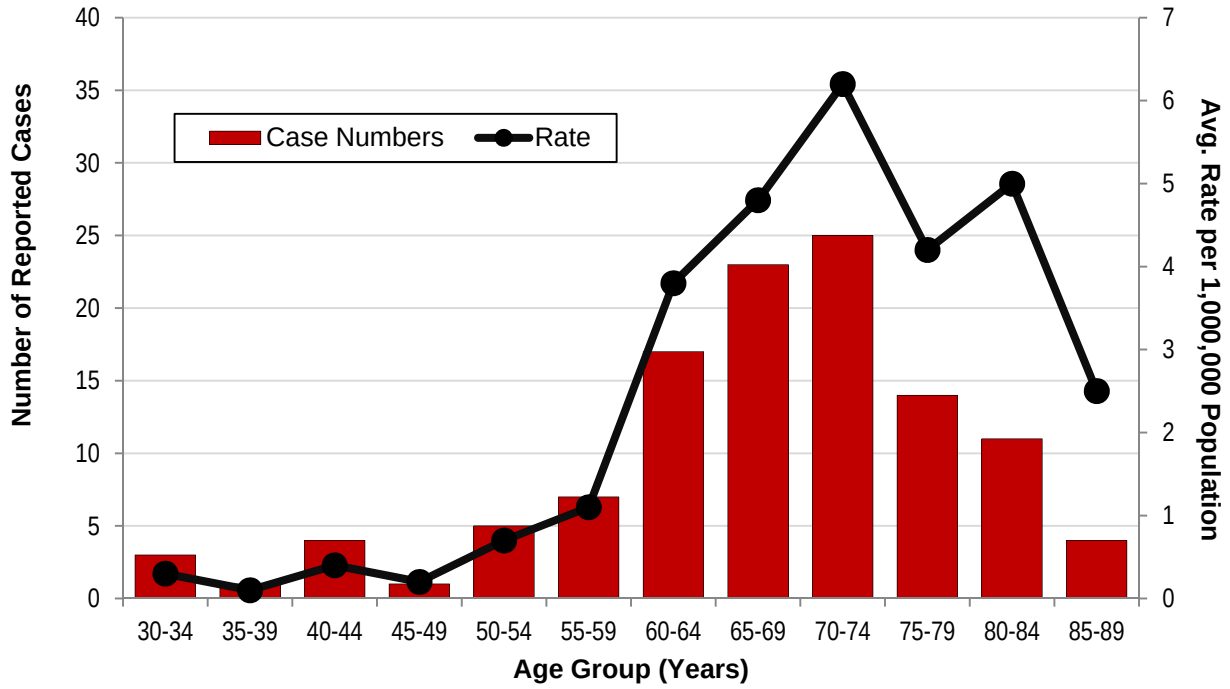
Sex

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 have ranged from 34 to 88 years. Incidence rates remained low until age 59 (less than 1.0 per million). From age 60 to 74 the rates increase to a high of six per million population, then the rates decreased back to 2.5 per million population. 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, 1980-2016



Geographical Distribution

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

Table: Number of CJD cases per parish – Louisiana, 1980-2017 by decade**

Parish	1980-89	1990-99	2000-09	2010-17	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	2	5
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	5	10
Ouachita	1	0	1	1	3
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	1	4
Winn	0	1	0	0	1
Louisiana	**29**	**37**	**24**	**17**	**107**

***Only parishes with cases are listed here*

Figure 3: Number of CJD cases – Louisiana, 1980-2016

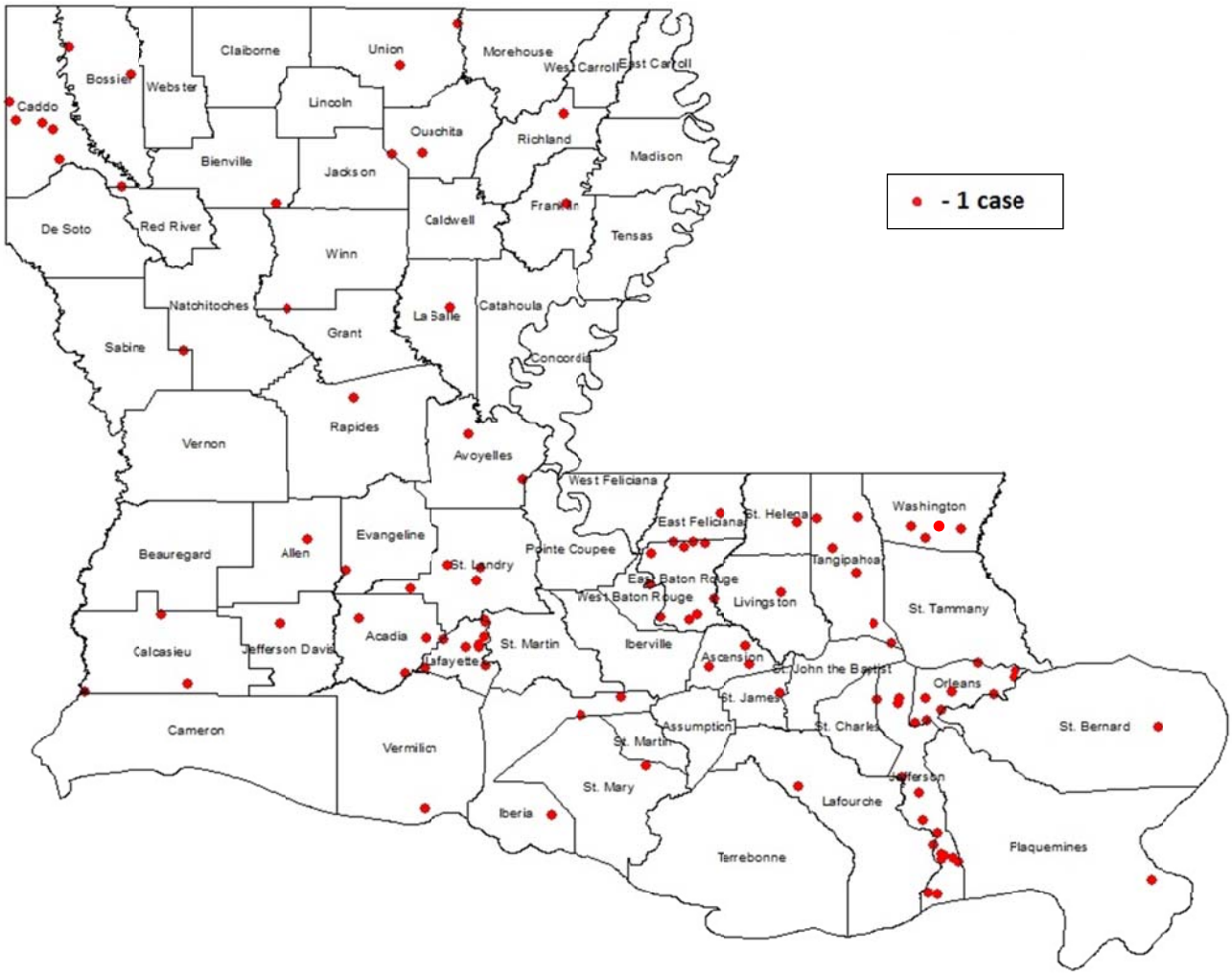
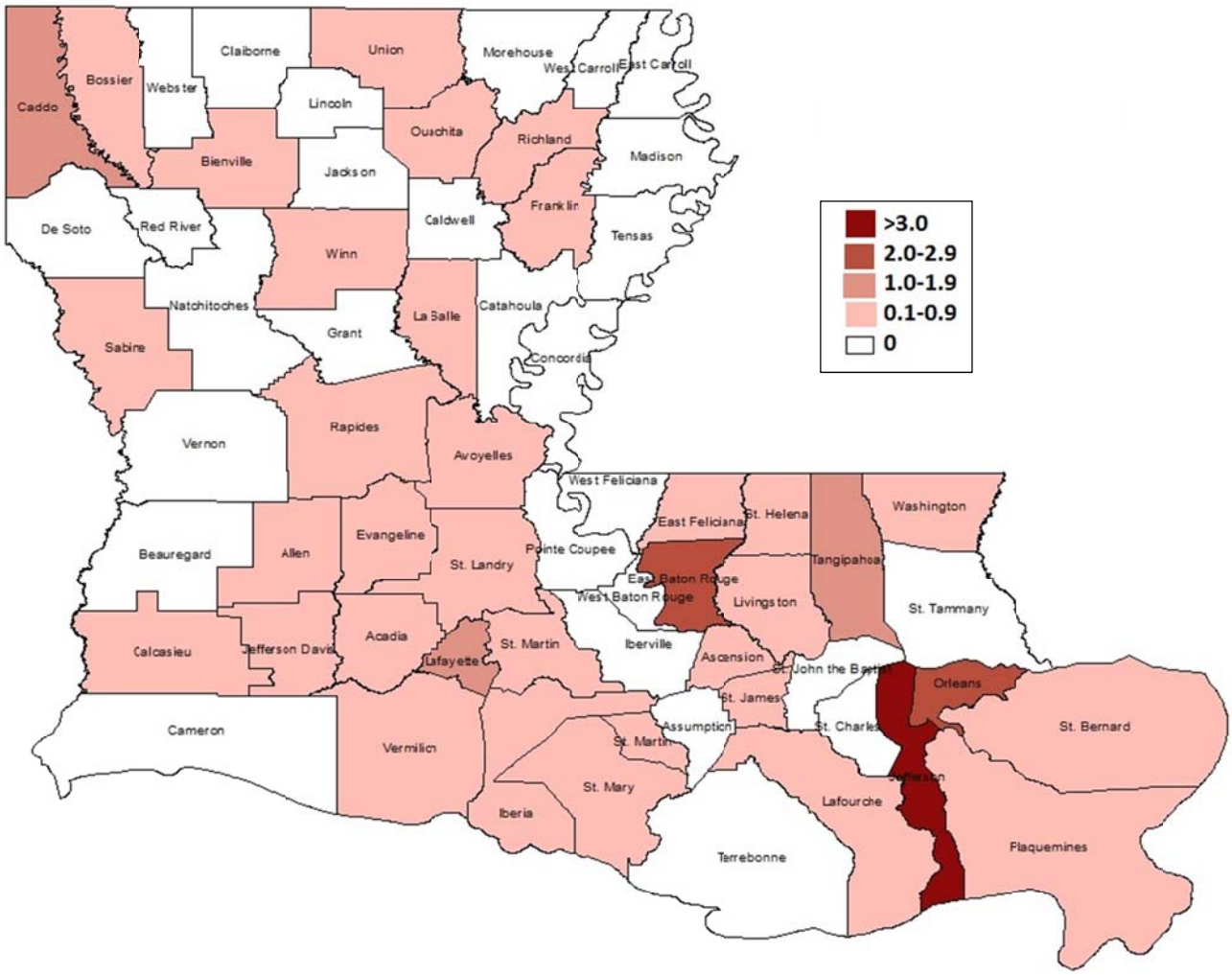


Figure 4: Incidence of CJD cases per million population per year – Louisiana, 1980-2016



Chronic Wasting Disease (CWD)

There has been a growing concern in the potential for chronic wasting disease (a prion disease in deer, elk, moose) to cause Creutzfeldt-Jakob disease in humans. Though there has **not** been any scientific evidence to support a claim of zoonotic transmission to humans, to date, CWD is of public health importance given its impact on the animal population and surveillance efforts have been ramped up.

CWD was first identified in captive deer in the late 1960s in Colorado and in wild deer in 1981. By the 1990s, it had been reported in surrounding areas in northern Colorado and southern Wyoming. Since 2000, the area known to be affected by CWD in free-ranging animals has increased to at least 22 states, including states in the Midwest, Southwest, and limited areas on the East Coast. It is possible that CWD may also occur in other states without strong animal surveillance systems, but that cases haven't been detected yet. Once CWD is established in an area, the risk can remain for a long time in the environment. The affected areas are likely to continue to expand.

Nationwide, the overall occurrence of CWD in free-ranging deer and elk is relatively low. However, in several locations where the disease is established, infection rates may exceed 10 % (one in ten), and localized infection rates of more than 25% (one in four) have been reported. The infection rates among some captive deer can be much higher, with a rate of 79% (nearly four in five) reported from at least one captive herd.

As of January 2018, CWD in free-ranging deer, elk and/or moose has been reported in at least 22 states in the continental United States, as well as two provinces in Canada. In addition, CWD has been reported in reindeer and moose in Norway, and a small number of imported cases have been reported in South Korea. The disease has also been found in farmed deer and elk.