

What is Creutzfeldt-Jakob disease?

Creutzfeldt-Jakob disease (CJD) is a rare disease that causes brain lesions in humans. It is believed to be caused by an infectious particle called a prion.

Who is at risk for Creutzfeldt-Jakob?

CJD is a rare disease occurring sporadically at a rate of one case per million each year. The risk of CJD increases with a person's age. The onset of classic CJD peaks in the 60 to 74 year old age group.

What are the symptoms of Creutzfeldt-Jakob?

CJD progresses rapidly once neurological symptoms appear. Symptoms include dementia, confusion, and defects in memory and other higher brain functions. Patients may not be able to coordinate voluntary muscle movements, and may have difficulty speaking and have involuntary muscle contractions. CJD is fatal. Once symptoms appear, infected people usually die within weeks to three to five months after symptoms appear. Only 10 to 15 percent of patients with CJD have survived longer than a year.

How soon do symptoms appear?

It is unknown how soon symptoms appear after infection in CJD cases.

How is Creutzfeldt-Jakob spread?

It is unknown how CJD is spread. Blood, milk, saliva, urine and feces do not appear to be involved in person-to-person transmission. Some CJD cases acquired their infection from another CJD case accidentally by medical or surgical treatments and procedures, such as pituitary hormone injections, dura mater grafts, cornea transplants, and use of some neurosurgical instruments. There is no documentation of transmission to infants born to infected mothers. There is some evidence of CJD disease running in families.

When and for how long is a person able to spread the disease?

Brain, eyes, spinal cord and spinal fluid are tissues associated with infectivity. Prolonged intimate contact with infected people does not transmit the disease.

How is a person diagnosed?

CJD is diagnosed by clinical symptoms along with diagnostic tests. However, diagnosis with CJD can be made certain only during an autopsy examination of the brain tissue.

What is the treatment?

There is no treatment for CJD. Only supportive treatment to manage symptoms is available.

Does past infection make a person immune?

No. There is no evidence of a protective immune response to CJD infection and a vaccine is not available. CJD is always fatal.

Should children or others be excluded from child care, school, work or other activities if they have Creutzfeldt-Jakob disease?

No. Infants, toddlers and school-age children should not be excluded unless the staff determines the child is unwilling or unable to participate in activities. They also should be excluded if the staff determines that they cannot care for the child without compromising their ability to care for the health and safety of the other children in the group.

What can be done to prevent the spread of Creutzfeldt-Jakob?

Making sure that organs or tissues from infected people are not used as transplants, along with strict donor selection criteria and protocols, can help prevent the spread of CJD cases associated with medical procedures. The proper cleaning of contaminated surgical instruments is also a preventative measure.

Variant Information:

In 1996, an outbreak of variant CJD (vCJD) occurred in people who had exposure to cattle tissue that had died of Mad Cow Disease (also known as Bovine Spongiform Encephalopathy or BSE). vCJD differs from classic CJD in that the disease onset occurs in younger people. Additionally, once infected, people usually die within 13 to 14 months. See the chart below for a few comparisons between classic and variant CJD:

Characteristic	Classic CJD	Variant CJD
Median age at death	68 years	28 years
Median duration of illness	4-5 months	13-14 months
Clinical signs and symptoms	Dementia; early neurologic signs	Prominent psychiatric/behavioral symptoms; painful dyesthesias; delayed neurologic signs
Periodic sharp waves on electroencephalogram	Often present	Often absent
"Pulvinar sign" on MRI*	Not reported	Present in >75% of cases
Presence of "florid plaques" on neuropathology	Rare or absent	Present in large numbers

Additional Information:

Additional information is available by calling the North Dakota Department of Health at 800.472.2180.

Resources:

1. American Academy of Pediatrics. [Children in Out-Of-Home Care]. In: Kimberlin DW, Brady MT, Jackson MA, Long SS, eds. *Red Book: 2015 Report of the Committee on Infectious Diseases*. 30th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2015: 132-151
2. American Academy of Pediatrics. [Prion Disease]. In: Kimberlin DW, Brady MT, Jackson MA, Long SS, eds. *Red Book: 2015 Report of the Committee on Infectious Diseases*. 30th ed. Elk Grove Village, IL: American Academy of Pediatrics; 2015: 653-656.
3. Centers for Disease Control and Prevention. (2015). Creutzfeldt-Jakob Disease, Classic (CJD). www.cdc.gov/ncidod/dvrd/vcjd/.
4. Centers for Disease Control and Prevention. (2015). Variant Creutzfeldt-Jakob Disease (vCJD). <http://www.cdc.gov/prions/vcjd/index.html>.