



CREUTZFELDT-JAKOB DISEASE (CJD)

What is Creutzfeldt-Jakob Disease?

Creutzfeldt-Jakob disease (CJD) is a rapidly progressive, fatal disorder that affects the brain and nervous system. It is caused by an infectious agent (organisms that can produce infectious disease) called a prion. Prion diseases are a group of rare, fatal brain diseases that occur in human and animals. There are three forms of CJD: sporadic, genetic/familial, and acquired (iatrogenic and variant).

Who gets CJD?

Prion diseases are rare with about 300 cases reported each year in the United States. The risk of CJD increases with age. Most cases of CJD are sporadic and tend to strike people around age 60. Variant CJD can occur in people in their twenties. Since genetic CJD is inherited, if one parent carries the mutation, there is a 50/50 chance for each child to inherit the gene.

How is CJD spread?

Sporadic CJD develops suddenly without any known risk factors or transmission pattern. Genetic/familial CJD runs in families with inherited mutations of the prion protein gene. Acquired iatrogenic CJD is caused by exposure to infected tissue or neurosurgical medical instruments during a surgical procedure such as a cornea transplant, pituitary hormone injections, and dura mater grafts.

Variant CJD can be transmitted by consuming beef infected with bovine spongiform encephalopathy (Mad Cow disease) or blood or blood transfusion. Individuals who were born with the CJD gene have a higher risk of developing genetic/familial CJD.

What are the symptoms of CJD?

CJD symptoms are rapidly developing dementia, confusion, difficulty walking, hallucinations, muscle stiffness, difficulty speaking, and defects in memory and higher brain functions.

How soon do symptoms appear?

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

Should an infected person be excluded from work or school?

No exclusions are required for a person diagnosed with CJD because there is no known human-to-human transmission at this time. For toddlers and school age children, staff can determine exclusion if the child is unwilling or unable to participate in activities or if they cannot care for the child without compromising their ability to care for the health and safety of the other children.



Frequently Asked Questions

What is the treatment for CJD?

Infection with this disease leads to death usually one year of onset of illness. There is no treatment for CJD. Only supportive care to manage symptoms is available.

What can a person or community do to prevent the spread of CJD?

Properly cleaning and sterilizing medical equipment may prevent the spread of the disease. Advocacy to prioritize prion disease research and continue CJD surveillance funding can help improve efforts to prevent and find a cure for CJD. Genetic testing is available. More information on genetic testing and counseling can be found at www.cjdfoundation.org.

Resources

Centers for Disease Control and Prevention, <https://www.cdc.gov/prions/>

Heymann DL. *Control of Communicable Diseases Manual*. Washington, DC: APHA Press, an imprint of American Public Health Association; 2022.