Fact sheet

Classic Creutzfeldt-Jakob disease (CJD)

What is classic Creutzfeldt-Jakob disease (CJD)?

Classic Creutzfeldt-Jakob disease (CJD) is a rare, deadly brain disease caused by abnormal proteins called prions. There are three types of classic CJD:

- Sporadic CJD (sometimes occurs with no known cause)
- Familial CJD (an inherited type of CJD that occurs in families)
- Iatrogenic or acquired CJD (occurs in a patient who was infected during a medical or surgical procedure)

How is classic CJD spread?

There is no known method of transmission for sporadic CJD. Those with familial CJD inherit an abnormal protein, making them more likely to develop the disease. Acquired CJD is spread through exposing brain tissue to infected material, and has been documented in the following circumstances:

- Persons receiving injections of pituitary hormones from the remains of infected bodies
- Persons receiving corneal (eye) transplants from infected donors
- Persons exposed to dirty surgical tools used on the brain during surgery
- Persons exposed to dirty equipment used during a test to monitor brain activity (EEG)
- Persons receiving grafts for the brain from infected donors
- Persons receiving transfusion of blood or blood products from a person with another type of CJD: variant CJD

Direct transmission of CJD from one person to another person, other than by the means listed above, has not been documented.

What are the signs and symptoms of classic CJD?

Initial symptoms differ from person to person. Symptoms include mental decline or dementia, memory loss, difficulty with balance and walking, dizziness, behavioral changes, visual troubles, and involuntary movements. In the advanced stages of the disease, people commonly develop severe difficulties moving and become unable to talk and swallow.

How long after infection do symptoms appear?

The exact time between infection and symptoms (incubation period) appearing is unknown for those infected with sporadic or familial CJD since there is no way to tell when the prion protein first formed. For those with acquired CJD, symptoms can appear anywhere from 1 year to 30 years or longer after infection. This time period varies depending on the way the infection was transmitted. Incubation periods tend to be longer for exposures that occur through the bloodstream, such as with pituitary-derived hormones.

Who is most at risk?

Most cases of classic CJD occur for unknown reasons, and no risk factors can be identified. However, some factors seem to be associated with the different types of classic CJD. Sporadic CJD usually develops later in life around age 60. Familial CJD comes from a genetic mutation, so family members of someone infected with familial CJD are at a slightly higher risk of CJD. Use of human growth hormone that is made from human pituitary glands has been linked with acquired CJD.

What type of health problems are caused by classic CJD?

Classic CJD is 100% fatal. Most people die within a few months of onset of symptoms. There is currently no cure or vaccine available.

How is classic CJD diagnosed?

After symptoms appear, doctors can often make an early diagnosis based on medical history, physical examination of the nervous system, and certain tests including:

- Electroencephalogram (EEG), a test that monitors brain activity
- Magnetic resonance imaging (MRI)
- Spinal fluid tests

A confirmed diagnosis of CJD requires sampling brain tissue, and can usually only be made after the patient's death.

How is classic CJD treated?

Symptoms of the disease are treated, but there is no treatment available that slows or stops the disease

How can classic CJD be prevented?

There is currently no known way to prevent cases of Sporadic and Familial CJD. There has been a large amount of work done to help prevent cases of Acquired CJD, such as:

- Using strict guidelines for sterilizing tools used in brain surgery
- Replacing growth hormone from human pituitary glands with a synthetic growth hormone
- Replacing human grafts of membranes that cover the brain and spinal cord with a synthetic substitute
- Denying organ and tissue donations from people who are suspected or known to have CJD

How common is classic CJD?

Classic CJD occurs in about 1 person per million people each year. Between 0 and 6 cases of CJD per year are expected in Utah residents. The majority (85-90%) of CJD cases are sporadic.

Is classic CJD reportable in Utah?

Both variant CJD and classic CJD are reportable and should be reported to your local health department or the Utah Department of Health and Human Services.

Where can I get more information?

- Your personal healthcare provider
- Utah Department of Health and Human Services
- Centers for Disease Control and Prevention (CDC)
- Creutzfeldt-Jakob Disease Foundation
- National Prion Disease Pathology Surveillance Center

Utah Department of Health & Human Services