

# Creutzfeldt-Jakob Disease (CJD, Classic)

## What is Creutzfeldt-Jakob disease?

Creutzfeldt-Jakob disease, or CJD, is a rare degenerative brain disease that is incurable and fatal. It is caused by prions, infectious, misshapen versions of normal proteins, which build up in the brain, damaging brain cells. CJD is the most common type of transmissible spongiform encephalopathy (TSE) found in humans.

## What are the symptoms?

Initial symptoms of CJD may include personality and behavioral changes, like anxiety and depression, memory loss, difficulty sleeping, and impaired thinking.

Symptoms usually progress to include blindness or blurred vision, weakness, loss of balance and coordination, difficulty walking and talking, and involuntary muscle jerking (myoclonus). Confusion rapidly progresses to dementia. People eventually lose the ability to move and speak. Most people with CJD die within one year.

## Who is at risk?

CJD occurs worldwide. Each year, about one in every million people in the United States develops the disease. Since 1996, the number of Minnesotans diagnosed with CJD has been between 3 and 13 a year.

The risk of developing CJD increases with age, with most cases occurring around the age of 60. It is extremely rare for people under the age of 40 to develop CJD.

## How is it spread?

There is no evidence to suggest that CJD can be transmitted through the air, sharing food or drink, or from direct person-to-person contact.

In about 85% of cases, CJD occurs sporadically, in persons with no known risk factors or inherited genetic mutations. This is known as *sporadic CJD*.

A smaller proportion of cases (5 to 15%) are hereditary and occur among persons with a family history of CJD (*familial CJD*).

In a very small number of reported cases, CJD has been transmitted to patients receiving dura mater grafts, corneal grafts or human pituitary hormone from donors with CJD (*iatrogenic CJD*). Today, infection control precautions and donor screening programs have made the likelihood of developing iatrogenic CJD through a medical procedure extremely rare.

## How is it diagnosed?

Only a brain biopsy or examination of brain tissue after death can confirm the diagnosis of CJD; this is usually done after the patient has died. An autopsy is recommended to confirm the diagnosis.

Clinical symptoms, a neurological exam, and other diagnostic tests may be used to help diagnose (but not confirm) CJD.

## How is it treated?

Currently there is no known cure for patients with CJD, however there are ways to relieve symptoms and to provide comfort and support.