

Creutzfeldt-Jakob Disease (CJD) Fact Sheet

What is Creutzfeldt-Jakob disease (CJD)?

Creutzfeldt-Jakob disease (CJD) is a rare, fatal brain disease caused by abnormal prions (microscopic protein particles). It is found worldwide, including in the United States (US), and usually affects older persons. CJD belongs to a class of prion diseases known as transmissible spongiform encephalopathies. CJD results when a brain prion protein mutates from a normal to an abnormal form causing other prion proteins to be transformed into abnormal forms leading to disease. Most prion mutations arise sporadically and have no known cause. CJD cannot be transmitted through the air or through touching or most other forms of casual contact.

How common is CJD?

In the US and worldwide, CJD occurs at an annual incidence of approximately one case per million people. During 2004-2008, approximately 300 cases of CJD were reported each year in the US. CJD is more common among older persons; the annual incidence averaged approximately 4.8 cases per million in persons aged 65 years or older. Rarely, CJD has been reported in a person younger than 40 years old.

In California, approximately 20-30 cases of laboratory-confirmed CJD have been reported annually in recent years.

What are the main types of CJD?

There are three main types of classic CJD:

1. Sporadic CJD (sCJD) occurs mostly in elderly persons (median age is 68 years) and occurs with no known cause. Most cases of CJD are sporadic. sCJD is not related to bovine spongiform encephalopathy (BSE).
2. Familial CJD (fCJD) is caused by a genetic mutation inherited from a parent. Approximately 5-10 percent of CJD cases in the US are familial. fCJD is not related to bovine spongiform encephalopathy (BSE).
3. Iatrogenic CJD (iCJD) represents less than one percent of CJD cases and is caused by unintended exposure to contaminated brain or nervous system tissue through uncommon medical procedures, including grafts of dura mater (a tissue that covers the brain) and injections of contaminated growth hormone derived from human pituitary glands taken from cadavers.

What is variant CJD?

Information on variant CJD is available on the [Variant CJD Fact Sheet](#).

What are the signs and symptoms of sCJD?

sCJD patients suffer from personality and behavioral changes, memory loss, and impaired thinking. As the illness progresses, dementia progresses and the patient may suffer muscle spasms, difficulty walking or speaking, blindness, and coma. Death occurs a median of 4-5 months following the onset of symptoms.

What are the signs and symptoms of fCJD?

The symptoms of fCJD can vary depending on the type of mutation involved. Symptoms can also vary greatly among affected family members. In general fCJD occurs at an earlier age than

sCJD. Symptoms may initially include depression, behavioral problems, and memory lapses. Other symptoms can include fatigue and visual disturbances, unsteadiness, lack of coordination, and difficulties with speech and/or swallowing.

What are the signs and symptoms of iCJD?

The signs and symptoms of iCJD are similar to those of sCJD.

How is CJD diagnosed?

It is hard to diagnose CJD. Clinical expertise from a neurologist (a medical specialist dealing with the nervous system) is usually needed, and often it is through the process of elimination of other diseases. Examination by a neurologist, laboratory tests, and other medical evaluations may help a doctor to suspect CJD, but the diagnosis can be confirmed only through a brain biopsy or autopsy.

How is CJD treated?

There are no effective treatments for CJD. The diseases are inevitably fatal. The few medications available for CJD patients focus only on easing their symptoms and discomfort.

Where can I get more information on CJD?

To get more information regarding CJD, please visit:

The U.S. Centers for Disease Control and Prevention website

<http://www.cdc.gov/ncidod/dvrd/prions/index.htm>

The National Institute of Health webpage

http://www.ninds.nih.gov/disorders/cjd/detail_cjd.htm#186503058

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