



Creutzfeldt-Jakob Disease (CJD) **Frequently Asked Questions**

What is Creutzfeldt-Jakob disease (CJD)?

Creutzfeldt-Jakob disease (CJD) is the most common human prion disease. Prion diseases are a group of rare brain and nervous system diseases that affect humans and some kinds of animals. Prion diseases are not caused by germs (i.e., viruses, bacteria). Instead, they are thought to result if normal brain proteins fold into abnormal forms. In affected people, clumps of these prion proteins cause brain damage and death.

CJD occurs worldwide in about one to two persons per million each year. In Washington State it affects about 7–8 people annually.

What causes CJD?

Most cases of CJD (~85%) occur in persons over 55 years old for unknown reasons. This type of CJD is called “sporadic” CJD. In some cases (10–15%), there is a genetic cause for CJD. Very rarely, CJD has been spread through contaminated neurosurgical instruments, dura mater or corneal grafts, or human-derived hormone therapies (no longer in use).

Another form of CJD, called variant CJD or vCJD, has affected more than 200 people worldwide. Most of those affected were 15–55 years old and lived in the United Kingdom during an epidemic of “mad cow disease” that occurred from 1980–1996. Mad cow disease is a cattle prion disease also known as bovine spongiform encephalopathy (BSE). vCJD is associated with eating products from BSE-affected cattle. There have not been any vCJD patients who got the disease in the United States. There are now many food safety measures in place to prevent BSE in cattle and to protect the food supply.

CJD is not spread from person to person by kissing, hugging or other close contact.

What are the symptoms of CJD?

People with CJD develop dementia and quickly deteriorate mentally. Involuntary twitching, rigid or spasming muscles, lack of coordination and balance, and visual problems are also common symptoms. CJD always leads to death, usually within one year of first symptoms.

How is CJD diagnosed?

Symptoms, family history, a lab test, and brain tests such as EEGs and MRIs can suggest to healthcare providers that a patient may have CJD. However, the definite diagnosis of CJD requires testing of brain tissue. When a person has an illness that could be CJD, it is important

that the person's family talk to their healthcare provider about getting a clear diagnosis after their loved one's death. There is no charge for the autopsy or specialized lab testing. Lab tests are done at the National Prion Disease Pathology Surveillance Center www.cjdsurveillance.com

How is CJD treated?

There is no treatment for CJD. The goal of care is to make the patient comfortable before death.

To support patients' families, the CJD Foundation operates a national toll-free line at (800) 659-1991 and a Web site: <http://www.cjdfoundation.org/>