Creutzfeldt-Jakob Disease (CJD) Fact Sheet

What is CJD?

Creutzfeldt-Jakob disease (pronounced “kroits-felt-yah-cub”; CJD) is a rare, progressive brain disease that is incurable and fatal. Scientists believe that CJD is caused by a prion, which is an abnormal protein that builds up in the brain and causes brain damage.

Who gets CJD?

CJD occurs worldwide. In the United States, about one person in every one million people develop CJD. Most people with the disease are over 60 years of age. In about 85% of patients, CJD occurs for no known reason. A smaller percentage of patients (5%-15%) inherit the disease. Although very rare (<1%), people who received infected tissue during surgical procedures (corneal transplants, surgery on the brain or its coverings) have developed CJD. A different form of the disease, variant CJD or vCJD, is described at the end of this factsheet and is associated with eating meat from infected cows.

How is CJD spread?

There is no evidence that CJD prions are spread through the air, food or drink, or contact between people. The prions could be spread by certain medical or surgical procedures. Prions are very difficult to destroy by heat or chemicals, so patients could be infected if surgical instruments have not been sterilized adequately after use on a CJD patient. Hospitals are very aware of the need to use disposable equipment or take extra precautions to sterilize surgical instruments so all patients are protected against infection with CJD.

What are the symptoms of CJD?

Early symptoms include forgetfulness, behavior changes, and lack of coordination. Patients with CJD eventually lose the ability to talk, walk, and take care of themselves. The disease progresses rapidly and most patients with CJD die within one year of illness onset.

How soon after exposure do symptoms appear?

It can take 15 months–30 years or more for symptoms to appear. Most of the time, there is no exposure to pinpoint. Rather, it is thought that something happens to make a normal prion change to a form that can causes disease, with no environmental source of infection.

How is CJD diagnosed?

Although there are several tests that might provide clues about whether someone has CJD, the only definite way to diagnose CJD is to perform a brain biopsy (remove a small piece of tissue from the patient’s brain for testing) or exam the brain after death.

What is the treatment for CJD?

There is no specific medication or cure for CJD. Treatment involves relieving symptoms to make the patient as comfortable as possible.
How can CJD be prevented?

People with CJD and people who are at increased risk for getting CJD because of their family history of CJD should never donate blood, tissues, or organs.

Caregivers of CJD patients should use good hygiene, including:

- Wash hands and exposed skin before eating, drinking, or smoking. This is a good habit to practice, even though contact with intact skin of a CJD patient does not spread the disease.
- Cover cuts and abrasions with waterproof bandages.
- Wear surgical gloves when handling a patient’s tissues and fluids or dressing a patient’s wounds.
- Avoid getting cut or stuck with items (e.g., needles) that might be contaminated with body fluids.
- Use face protection if there is a risk of exposure to the patient’s blood or body fluids.
- If a CJD patient’s body fluids get on someone else’s skin, wash the affected area with detergent, rinse well with warm water (avoid scrubbing), rinse the site with a 1:10 dilution of bleach for 1 minute (for maximum safety), and then rinse again with water. If a CJD patient’s body fluids get into someone else’s eye or mouth, rinse well with saline (for eye) or tap water (for mouth).
- Dispose of all clinical waste (e.g., bandages) properly.

What is the relationship of CJD to “Mad Cow Disease”?

“Mad Cow Disease” or bovine spongiform encephalopathy (BSE) is a disease that occurs in cattle. Some people who have eaten meat from cattle infected with BSE have gotten a disease that is similar to CJD but is caused by a different prion. This disease is called ‘variant CJD’ (vCJD) and has been diagnosed in about 200 people worldwide. In the United States, only four cases of vCJD have been diagnosed; all in people who grew up and were exposed to BSE outside the United States. The U.S. Department of Agriculture has developed measures to screen cattle so BSE-infected animals are excluded from the food chain and their tissues cannot infect other animals or humans.

How can I learn more about CJD?

- If you have concerns about CJD, contact your healthcare provider.
- Call your local health department. A directory of local health departments is located at https://www.vdh.virginia.gov/local-health-districts/.
- Visit the Centers for Disease Control and Prevention website at https://www.cdc.gov/prions/cjd/.

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