

## What is Creutzfeldt-Jakob disease?

Creutzfeldt-Jakob disease (CJD) is a rare, degenerative fatal brain disorder. It affects about one person in every one million people worldwide and about 200 people in the United States annually. CJD usually appears later in life and runs a rapid course. Typically, onset of symptoms occurs about age 60, and about 90% of patients die within one year. There are three major categories of CJD:

- In *sporadic CJD*, the disease appears even though the person has no known risk factors for the disease. This is by far the most common type of CJD and accounts for at least 85% of cases.
- In *hereditary CJD*, the person has a family history of the disease and/or tests positive for a genetic mutation associated with CJD. About five to ten percent of cases of CJD in the U.S. are hereditary.
- In *acquired CJD*, the disease is transmitted by exposure to brain or nervous system tissue, usually through certain medical procedures. There is no evidence that CJD is contagious through casual contact with a CJD patient.

## What are the symptoms of the disease?

The first symptoms of Creutzfeldt-Jakob disease typically include *dementia* – personality changes together with impaired memory, judgment, and thinking – and problems with muscular coordination, behavior changes and visual disturbances. As the illness progresses, persons quickly deteriorate mentally along with developing involuntary movements, blindness and weakness of extremities. They eventually lose the ability to move and speak and enter a coma. Pneumonia and other infections often occur in these patients and can lead to death. People with the disease also may experience insomnia, depression, or unusual sensations. CJD does not cause a fever or other flu-like symptoms. Some symptoms of CJD can be similar to symptoms of other progressive neurological disorders, such as Alzheimer's or Huntington's disease. It also tends to cause more rapid deterioration of a person's abilities than Alzheimer's disease or most other types of dementia.

## How long does it take to become ill with CJD once a person has been exposed?

The incubation period is usually very long - ranging from 15 months to several decades.

## How is CJD diagnosed?

There is currently no single diagnostic test for CJD. A neurological examination will be performed or your healthcare provider may seek consultation with other physicians. Some standard diagnostic tests, such as a spinal tap and an electroencephalogram (EEG), will also be done. The only way to confirm a diagnosis of CJD is by *brain biopsy* or *autopsy*. In a *brain biopsy*, a neurosurgeon removes a small piece of tissue from the patient's brain so that it can be examined by a neuropathologist. In an *autopsy*, the whole brain is examined after death.

## How is the disease treated?

There is no treatment that can cure or control Creutzfeldt-Jakob disease. Current treatment for CJD is aimed at helping symptoms and making the patient as comfortable as possible.

## What causes Creutzfeldt-Jakob disease?

The cause of CJD is believed to be a prion, which is a self-replicating protein. CJD is **not** caused by a bacteria, virus, or parasite. The normal form of the prion is found in all people, but sometimes is converted into an abnormal form. The abnormal CJD prion produces the brain lesions that result in the disease. Sporadic CJD may develop because some of a person's normal prions spontaneously change into abnormal prions. About five to ten percent of all CJD cases are inherited. These cases arise from a mutation, or change, in the gene that controls formation of the normal prion protein. Exposure to abnormal prions from an external source can sometimes result in the disease. It is also known that ingestion of human CJD prions can result in CJD.

### **How is CJD transmitted?**

While CJD can be transmitted to other people, it cannot be transmitted through the air, through touching or most other forms of casual contact. Spouses and other household members of sporadic CJD patients are at no higher risk of getting the disease than the general population. However, direct or indirect contact with brain tissue or spinal cord fluid from infected patients should be avoided to prevent transmission of the disease through these materials.

In about 85% of patients, CJD occurs as a sporadic disease with no recognizable pattern of transmission. Some patients develop CJD as a result of inherited mutations of the prion protein gene. This occurs among persons with a family history of CJD. The remaining cases of CJD are the result of medical procedures using infected human matter or surgical instruments from an infected individual.

### **Can CJD be acquired from animals?**

No. There is no current evidence of a case of CJD ever resulting from eating the meat of an infected animal. New variant CJD (nv-CJD or v-CJD) or “Mad Cow Disease” in people is a similar but distinctly different disease from CJD. This disease tends to cause illness in individuals who are much younger than people with CJD. There have been thoughts that human cases of “Mad Cow Disease” can be caused by eating beef contaminated with the animal form of the disease (BSE—Bovine Spongiform Encephalopathy). The majority of human “Mad Cow Disease” cases have occurred in Great Britain and France. One case was identified in the U.S. but this person had lived in the United Kingdom for several years. Importation of cattle and beef from countries with BSE has been banned in the United States since 1989 to reduce the risk that it will occur in the U.S. Chronic wasting disease (CWD) of deer and elk, and scrapie in sheep and goats, are also caused by prions .

### **What precautions should family members, health workers or other caregivers take?**

Normal sterilization procedures such as cooking, washing, and boiling do not destroy prions. The tissues and fluids considered infectious for CJD are corneas, brain and spinal cord tissue, and cerebrospinal fluid. For the routine daily care of a CJD patient, universal precautions are considered adequate. Caregivers, healthcare workers, and undertakers should take the following precautions when they are working with a person with CJD:

- Wash hands and exposed skin before eating, drinking, or smoking.
- Cover cuts and abrasions with waterproof dressings.
- Wear surgical gloves when handling a patient’s tissues and fluids or dressing the patient’s wounds.
- Avoid cutting or sticking themselves with instruments contaminated by the patient’s blood or other tissues.
- Use face protection if there is a risk of splashing contaminated material such as blood or cerebrospinal fluid.
- Soak instruments that have come in contact with the patient in undiluted chlorine bleach for an hour or more, then use an autoclave (pressure cooker) to sterilize them in distilled water for at least one hour at 132° - 134° Centigrade.

