Creutzfeldt-Jakob Disease Fact Sheet

What is Creutzfeldt-Jakob Disease?

Cretuzfeldt-Jakob disease (CJD) is a rare, degenerative, invariably fatal brain disorder. It affects about one person in every one million people worldwide and about 200 people in the United States. CJD usually appears in later life and runs a rapid course. Typically, onset of symptoms occurs about age 60 and about 90 percent of patients die within 1 year. In the early stages of disease, patients may have failing memory, behavioral changes, lack of coordination and visual disturbances. AS the illness progresses, mental deterioration becomes pronounced and involuntary movements, blindness, weakness of extremities and coma may occur.

There are three major categories of CJD:

1. 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 percent of cases.
2. In hereditary CJD, the person has a family history of the disease and/or tests positive for a genetic mutation associated with CJD. About 5 to 10 percent of cases of CJD in the United States are hereditary.

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3. 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. People with the disease also may experience insomnia, depression or unusual sensations. CJD does not cause a fever or other flu like symptoms. As the illness progresses, the patient's mental impairment becomes severe. They often develop in-voluntary muscle jerks called myoclonus and they may go blind or lose bladder control. 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.

Some symptoms of CJD can be similar to symptoms of other progressive neurological disorders such as Alzheimer's or Huntington's disease. However, CJD

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causes unique changes in brain tissue, which can be seen at autopsy. It also tends to cause more rapid deterioration of a person's abilities than Alzheimer's disease or most other types of dementia.

How is CJD diagnosed?

There is currently no single diagnostic test for CJD. When a doctor suspects CJD the first concern is to rule out treatable forms of dementia such as encephalitis (inflammation of the brain) or chronic meningitis. A neurological examination will be performed or the doctor may seek consultation with other physicians. Some standard diagnostic tests, such as a spinal tap 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 a neuropathologist can examine it. In an autopsy, the whole brain is examined after death.

How is the disease treated?

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

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What causes Creutzfeldt-Jakob disease?

The leading scientific theory at this time maintains that CJD is caused not by an organism but by a type of protein called a prion. Prions occur in both a normal form, which is a harmless protein, found in the body's cells; and in an infectious form, which causes disease. The harmless and infectious forms of the prion protein are nearly identical, but the infectious form takes a different folded shape than the normal protein. Sporadic CJD may develop because some of a person's normal prions spontaneously change into the infectious form of the protein and then alter the prions in other cells in a chain reaction. Once they appear, abnormal prion proteins stick together and form fibers and/or clumps called plaques that can be seen with powerful microscopes.

Fibers and plaques may start to accumulate years before symptoms of CJD begin to appear. It is still unclear what role these abnormalities play in the disease or how they might affect symptoms. About 5 to 10 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.

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How is CJD transmitted?

While CJD can be transmitted to other people, the risk of this happening is extremely small. CJD cannot be transmitted through the air or through touching or most other forms of causal contact. Spouses and other household members of sporadic CJD patients have no higher risk of contracting the disease than the general population.

However, direct or indirect contact with brain tissue and spinal cord fluid from infected patients should be avoided to prevent transmission of the disease through these materials.

In a few very rare cases, CJD has spread to other people from grafts of dura mater (a tissue that covers the brain), transplanted corneas, implantation of inadequately sterilized electrodes in the brain, and injections of contaminated pituitary growth hormone derived from human pituitary glands taken from cadavers. Since 1985, all human growth hormone used in the United States has been synthesized by recombinant DNA procedures, which eliminates the risk of transmitting CJD by this route.

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Can CJD be acquired from animals?

The appearance of a new variant of CJD (nv-CJD or v-CJD) in several younger than average people in Europe has led to concern that BSE can be transmitted to humans through consumption of contaminated beef. Although laboratory tests have shown a strong similarity between the prions causing BSE and v-CJD, there is no direct proof to support this theory. Furthermore, **BSE has never been found in the United States** and importing 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 this country.

Chronic wasting disease (CWD) of deer and elk and scrapie in sheep and goats are also caused by prions in these animal species. There is no current evidence of a case of CJD ever resulting from eating the meat of an infected animal.

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 flu-ids 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.

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Caregivers, health care workers and undertakers should take the following precautions when they are working with a person with CJD:

1. Wash hands and exposed skin before eating, drinking or smoking.
2. Cover cuts and abrasions with waterproof dressings.
3. Wear surgical gloves when handling a patient's tissues and fluids or dressing the patient's wounds.
4. Avoid cutting or sticking themselves with instruments contaminated by the patient's blood or other tissues.
5. Uses face protection if there is a risk of splashing contaminated material such as blood or cerebrospinal fluid.
6. 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.

For further information, contact the
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