Creutzfeldt-Jakob Disease Question

Creutzfeldt-Jakob disease (CJD) is a rare, degenerative, invariably fatal brain disorder. It affects about one person in every one million people worldwide.

ATLANTA, GEORGIA, UNITED STATES, January 3, 2015 /EINPresswire.com/ -- Creutzfeldt-Jakob disease (CJD)in the United States there are about 300 cases per year. CJD usually appears in later life and runs a rapid course. Typically, onset of symptoms occurs about age 60, and about 90 percent of individuals die within 1 year. In the early stages of disease, people 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:

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.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.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. Since CJD was first described in 1920, fewer than 1 percent of cases have been acquired CJD.CJD belongs to a family of human and animal diseases known as the transmissible spongiform encephalopathies (TSEs). Spongiform refers to the characteristic appearance of infected brains, which become filled with holes until they resemble sponges under a microscope. CJD is the most common of the known human TSEs. Other human TSEs include kuru, fatal familial insomnia (FFI), and Gerstmann-Straussler-Scheinker disease (GSS). Kuru was identified in people of an isolated tribe in Papua New Guinea and has now almost disappeared. FFI and GSS are extremely rare hereditary diseases, found in just a few families around the world. Other TSEs are found in specific kinds of animals. These include bovine spongiform encephalopathy (BSE), which is found in cows and is often referred to as “mad cow” disease; scrapie, which affects sheep and goats; mink encephalopathy; and feline encephalopathy. Similar diseases have occurred in elk, deer, and exotic zoo animals.

What are the Symptoms of the Disease?

CJD is characterized by rapidly progressive dementia. Initially, individuals experience problems with muscular coordination; personality changes, including impaired memory, judgment, and thinking; and impaired vision. 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, mental impairment becomes severe. Individuals often develop involuntary muscle jerks called myoclonus, and they may go blind. They eventually lose the ability to move and speak and enter a coma. Pneumonia and other infections often occur in these individuals and can lead to death.

There are several known variants of CJD. These variants differ somewhat in the symptoms and course of the disease. For example, a variant form of the disease-called new variant or variant (nv-CJD, v-CJD), described in Great Britain and France-begins primarily with psychiatric symptoms,

affects younger individuals than other types of CJD, and has a longer than usual duration from onset of symptoms to death. Another variant, called the panencephalopathic form, occurs primarily in Japan and has a relatively long course, with symptoms often progressing for several years. Scientists are trying to learn what causes these variations in the symptoms and course of the disease.

Some symptoms of CJD can be similar to symptoms of other progressive neurological disorders, such as Alzheimer's or Huntington's disease. However, CJD 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 and the doctor may seek consultation with other physicians. Standard diagnostic tests will include a spinal tap to rule out more common causes of dementia and an electroencephalogram (EEG) to record the brain's electrical pattern, which can be particularly valuable because it shows a specific type of abnormality in CJD. Computerized tomography of the brain can help rule out the possibility that the symptoms result from other problems such as stroke or a brain tumor. Magnetic resonance imaging (MRI) brain scans also can reveal characteristic patterns of brain degeneration that can help diagnose CJD.

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. This procedure may be dangerous for the individual, and the operation does not always obtain tissue from the affected part of the brain. Because a correct diagnosis of CJD does not help the person, a brain biopsy is discouraged unless it is needed to rule out a treatable disorder. In an autopsy, the whole brain is examined after death. Both brain biopsy and autopsy pose a small, but definite, risk that the surgeon or others who handle the brain tissue may become accidentally infected by self-inoculation. Special surgical and disinfection procedures can minimize this risk. A fact sheet with guidance on these procedures is available from the NINDS and the World Health Organization.

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