Creutzfeldt-Jakob Disease

Creutzfeldt-Jakob disease (CJD) is a rare, degenerative, invariably fatal brain disorder. Typically, onset of *symptoms* (problems) occurs at about age 60. There are three major categories of CJD: sporadic CJD, *hereditary* (passed from parents) CJD, and acquired CJD. There is currently no single diagnostic test for CJD. The first concern is to rule out treatable forms of dementia such as encephalitis or *chronic* (longstanding) meningitis. The only way to confirm a *diagnosis* (learning what is wrong) of CJD is by brain *biopsy* (tissue sample) or autopsy. In a brain biopsy, a neurosurgeon removes a small piece of tissue from the patient's brain so that is can be examined by a neurologist. Because a correct diagnosis of CJD does not help the patient, a brain biopsy is discouraged unless it is need to rule out a treatable disorder. While CJD can be transmitted to other people, the risk of this happening is extremely small.

TREATMENT

There is no treatment that can cure or control CJD. Current treatment is aimed at alleviating symptoms and making the patient as comfortable as possible. Opiate drugs can help relieve pain, and the drugs clonazepam and sodium valproate may help relieve involuntary muscle jerks.

WHAT WILL HAPPEN

About 90 percent of patients die within 1 year of diagnosis. 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* (arms/hands, legs/feet), and coma may occur.

RESEARCH BEING DONE

The leading scientific theory at this time maintains that CJD is caused by a type of protein called a prion. The harmless and the infectious forms of the prion protein are nearly identical, but the infectious form takes a different folded shape than the normal protein. Researchers are examining whether the transmissible agent is, in fact, a prion and trying to discover factors that influence prion infectivity and how the disorder damages the brain. Using rodent models of the disease and brain tissue from autopsies, they are also trying to identify factors that influence the susceptibility to the disease and that govern when in life the disease appears.

Research is continually being done to learn more about this problem.
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