Michigan Department Of Community Health
MDCH

Creutzfeldt-Jakob Disease (CJD) General Fact Sheet

What is Creutzfeldt-Jakob disease?

CJD is a rare, progressive, and incurable disease of the nervous system. The disease is caused by an abnormal form of a brain protein, which self-replicates and accumulates in the brain causing damage and eventually death. Physicians first recognized and described CJD in the 1920's.

What are the different types of CJD?

Sporadic CJD- is the most common human prion disease and accounts for approximately 85-90% of CJD cases. The disease results from the spontaneous conversion of the normal brain protein into the disease-causing abnormal prion protein.

Familial CJD- is an inherited form of CJD caused by a genetic mutation and accounts for 5-15% of cases. This mutation makes it more likely that the brain protein will convert to the abnormal disease causing form.

Iatrogenic CJD- is transmitted by direct exposure to abnormal prion proteins from an external source during a medical procedure. In rare situations, CJD has been spread by the re-use of contaminated surgical instruments or the transplantation of certain high-risk tissues from a CJD infected donor.

Variant CJD (vCJD)- the human form of bovine spongiform encephalopathy (BSE) or "Mad Cow Disease." Variant CJD is linked to the consumption of meat from an animal infected with BSE or by blood transfusion from a donor infected with vCJD. Variant CJD is a separate disease and is caused by a different prion than the sporadic or familial forms of CJD.

What are the symptoms of CJD?

CJD has a very long incubation (the time from infection to the onset of symptoms) ranging from 15 months to 30 years. Initial symptoms of CJD generally include progressive dementia, behavioral changes, and muscle incoordination. Other symptoms may include depression, insomnia, and problems with vision and speech. Once clinical symptoms begin, the progression of the disease is rapid; nearly all individuals diagnosed with CJD die within one year.

How is CJD diagnosed?

The use of computed tomography (CT) scans, magnetic resonance imaging (MRI), electroencephalogram (EEG), specific tests for certain proteins in cerebrospinal fluid, and clinical signs can provide evidence for the diagnosis of CJD. However, the only way a confirmed diagnosis of CJD can be made is by brain biopsy or autopsy. Brain tissue must be examined to determine if a person has a prion disease and also the type of prion disease. There is currently no cure for CJD and treatment relies on supportive care for the patient.

How many cases of sporadic CJD are seen in the US?

Current surveillance suggests that one to two cases of CJD occur per million people per year. However, the risk of CJD increases with age and the rate of disease is 3.4 cases per million people in individuals over the age of 50. In recent years, fewer than 300 cases of CJD have been reported per year in the U.S.

Are CJD and vCJD caused by BSE/"Mad Cow Disease" the same?

No, variant CJD (the human form of BSE or "mad cow disease") is caused by a different prion protein from the sporadic or familial forms of CJD. Individuals diagnosed with vCJD are generally younger, <55 years old, and have a longer course of illness. Beyond the marked differences in clinical signs and symptoms, the examination of brain tissue allows for differentiation between variant CJD and other forms of human prion disease. Three cases of variant CJD have been diagnosed in the United States. However, all of these cases were foreign born and grew up outside of the U.S. To date, no cases of variant CJD acquired in the U.S. have been found.

Does Chronic Wasting Disease (CWD) cause CJD disease in humans?

CWD is fatal neurological prion disease found in cervids (deer, elk and moose). To date, CWD is not known to cause or be associated with disease in humans. No increase in human prion disease has been observed in areas of the western United States where CWD has been endemic in cervid populations for decades. However, because much is still unknown about prion diseases, the Centers for Disease Control and Prevention and the World Health Organization advise that humans do not consume animals that are known to be infected with CWD. In general, people should not handle or consume wild animals that appear sick or act abnormally, regardless of the cause. The risk of acquiring a prion disease from an animal source is extremely remote. Research suggests that the biological differences between humans and animals prevent animal prion diseases from being easily transmitted to people. Scrapie is an animal prion disease in sheep that has been recognized since the 18th century and has never been shown to be transmitted to or cause disease in humans.

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