

Creutzfeldt-Jakob Disease (CJD)

Disease Fact Sheet Series

What is Creutzfeldt-Jakob disease (CJD)?

CJD is a rare incurable disease of humans that affects the nervous system and results in rapidly progressive dementia, loss of motor control, paralysis, and death. It is one of several related diseases called transmissible spongiform encephalopathies or TSEs for short. The term "encephalopathy" means the brain is affected, and the term "spongiform" refers to the microscopic holes seen in the brain, giving it a sponge-like appearance.

Various animal species have distinct types of TSEs. In addition to CJD which affects humans, other TSEs include bovine spongiform encephalopathy (BSE, also known as "mad cow disease"), scrapie in sheep, and chronic wasting disease (CWD) in deer and elk.

What causes CJD?

A recently identified agent called a prion, that is a self-replicating protein, causes it. The current theory is that the normal form of the prion, found in all people, is converted into an abnormal form. This abnormal CJD prion produces the brain lesions that result in the disease.

What causes this conversion of normal protein into a disease producing prion?

In many cases, this is not known with certainty. In about 90% of cases, this conversion apparently occurs spontaneously, with no known cause. About 10% of cases are known to be familial – that is, these patients have inherited a genetic mutation that results in a tendency for their prion protein to change to the disease producing form. Exposure to abnormal prions from an external source can sometimes result in the disease. For example, some cases of CJD have resulted from medical procedures in which neural tissues or extracts, unknowingly taken from a CJD patient, have produced CJD in the person who received these products. It is also known that ingestion of human CJD prions can result in CJD.

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

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

Is CJD a new disease?

No. CJD may have always occurred, and has been recognized by scientists since the 1920s. Only recently, however, has the prion been generally accepted as the cause of the disease.

How common is CJD?

The Centers for Disease Control and Prevention estimates the incidence of CJD at one case per million of population. This means that, on average, about 5 cases of CJD should occur in Wisconsin each year. The highest incidence of CJD occurs in persons older than 65 years.

Is there a test to tell if a human has CJD?

There is no currently available test to determine if people are incubating CJD. Once symptoms occur, clinical signs make the CJD diagnosis, characteristic electroencephalogram (EEG) patterns detection of certain proteins in the cerebrospinal fluid, and by detection of spongiform changes in a brain biopsy.

Is CJD the same as mad cow disease and chronic wasting disease?

No. Although all three are TSEs, and therefore cause similar illnesses and similar lesions in the brain of the species they affect, these diseases are caused by distinct prions.

Isn't there a connection between CJD and mad cow disease (BSE)?

To answer that, an explanation is needed of the forms of CJD. There are actually two distinct types of CJD - classic Creutzfeldt-Jakob disease and new-variant Creutzfeldt-Jakob disease (nvCJD). The above discussion pertained to classic CJD that occurs worldwide, including the USA, and has been recognized for decades.

New-variant CJD is a recently described form of the disease found in Great Britain and some other European countries; it has never been found in the USA, with the exception of one person who had previously lived in Great Britain. New-variant CJD is quite similar to classic CJD, but tends to occur in younger people, under the age of 45 years, and differs subtly from classic CJD in clinical and neurological features.

There is strong evidence that the cases of nvCJD in Great Britain are related to the occurrence of BSE in their cattle. If true, this means that the BSE prion has been able to infect humans, probably through the consumption of beef products from infected cattle. Although hundreds of thousands of cases of BSE have occurred in British cattle, BSE has been detected in only one cow in the USA. American agriculture officials have taken measures to exclude any cattle potentially infected with BSE from entering our country.

Is there any evidence that chronic wasting disease (CWD), which does occur in deer and elk in the USA, poses a risk of causing CJD-like disease in humans?

There is currently no such evidence, even though CWD has occurred in the USA for at least four decades. Furthermore, the incidence of CJD in residents of Wyoming and Colorado where CWD has occurred for years is no higher than in states where CWD does not occur. The World Health Organization and the U.S. Centers for Disease Control and Prevention has found no scientific evidence that CWD can infect humans.

Although this is reassuring, but no one can predict with absolute certainty that CWD will *never* cause human disease. It is likely that at least once in the past, prions have crossed the "species barrier" when BSE prions presumably resulted in the outbreak of human nvCJD in Europe.

Because of this small degree of uncertainty, experts suggest that people should eat no part of a deer or elk with any evidence of CWD. Additionally people should avoid consuming brain, spinal cord, eyes, spleen, tonsils and lymph nodes of any harvested animals because the CWD prion occurs in these tissues if an animal is infected. Muscle tissue has not been shown to be infectious. Animals that test positive for CWD, or which appear to be emaciated, acting abnormally, or showing any other signs of CWD should not be harvested for food.