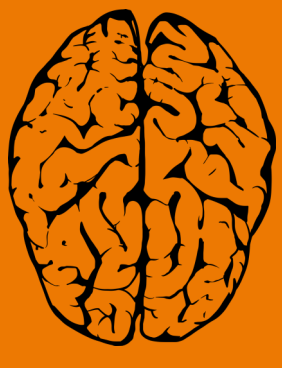
CREUTZFELDT-JAKOB DISEASE (CJD)



Creutzfeldt-Jakob Disease (CJD) is an extremely rare, incurable disease of humans that affects the nervous system. It is one of a family of diseases called transmissible spongiform encephalopathies (TSEs). Encephalopathy means that the brain is affected while spongiform refers to microscopic holes in the brain, making the brain look like a sponge. Other animals suffer from types of TSEs; cattle can have bovine sponge encephalopathy (BSE) also known as mad cow disease, sheep suffer from scrapie, and deer and elk can get chronic wasting (CWD). CJD occurs most in people who are over 65 years old.

What causes it?

- CJD is caused by a protein called a prion, which is self-reproducing. Everyone has these proteins in their body, but CJD occurs when the prion becomes abnormally shaped and causes lesions in the brain.
- CJD can take a long time to develop before symptoms are noticeable. Symptoms can take 15 months to decades to be seen.

What are the types of CJD?

- **Sporadic CJD** occurs when the protein spontaneously changes to the abnormal shape with no known cause. This type of CJD makes up approximately 90% of all CJD cases.
- **Inherited CJD** occurs when a person inherits a gene from a parent that makes the protein change shape. This type of CJD makes up approximately 10% of all CJD cases.
- **Acquired CJD** occurs very rarely and happens when a person comes into contact with outside sources of CJD contaminated tissue (e.g., from a tissue graft), which can cause their own prions to begin changing into an abnormal shape.
- **Variant CJD** is a rare type of human CJD that was discovered in the 1990s. Studies have shown that it resulted from people eating beef from British cattle that were infected with BSE prions during the 1980s. Most cases of variant CJD occurred in the United Kingdom (U.K.); none were acquired in the United States (U.S.).

What are the signs and symptoms?

- Rapidly progressive dementia
- Loss of muscle control
- Paralysis
- Death

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Is CJD the same as mad cow disease and CWD?

- CJD is not the same as mad cow disease or CWD. All three diseases are in the TSE family and can cause related illnesses and brain lesions. However, they are caused by three different prions that can be differentiated from one another in a laboratory.

Is it possible for CWD in deer and elk to cause CJD-like disease in humans?

- The U.S. Centers for Disease Control and Prevention (CDC) and World Health Organization (WHO) have found no evidence to date that CWD can infect humans. The only known instance of an animal prion disease affecting humans occurred during the 1990s in Great Britain. This occurred when a variant of CJD in humans was caused by human consumption of BSE prions in contaminated beef from cattle that had mad cow disease.
- People should not eat any part of deer or elk that have signs of CWD because CWD prions can be found at low levels in the muscles if the deer or elk are infected. It is recommended that brain, spinal cord, eyes, spleen, tonsils, and lymph node tissue from any deer or elk not be eaten because the CWD prion is found at higher levels in these tissues if the animal is infected.
- Deer that test positive for CWD, are acting strangely, look thin, or have any other signs of CWD should not be eaten.

What are the treatment options?

- There are no current treatment options for CJD, which is always fatal. Supportive care should be provided for complications.
- There is no test to tell if someone has CJD. Once symptoms begin, a diagnosis is made by clinical signs, electroencephalogram (EEG) testing, cerebrospinal fluid analysis, and finding lesions in brain tissue.

How can CJD be prevented?

- Unfortunately, there is no known way to prevent **sporadic CJD**.
- If you have a family history consistent with CJD, it may be helpful for you to meet with a genetic counselor to help you determine your specific risk for **inherited CJD**.
- To prevent **acquired CJD**, it is necessary to destroy or specially process any instruments that come into contact with suspected or confirmed CJD tissues.
- **Variant CJD** prevention was essentially accomplished when the outbreak of mad cow disease, or BSE, was controlled in the U.K. Rare, sporadic cases of BSE in cattle still occur, and the U.S. has taken steps to prevent diseased cattle from entering the U.S. food chain.