Creutzfeldt–Jakob Disease (CJD)

Creutzfeldt–Jakob Disease (CJD) is an extremely rare disease in humans that affects the nervous system. There is no cure. It is one of a family of diseases called transmissible spongiform encephalopathies (TSEs) or prion diseases. These diseases lead to a build-up of abnormal proteins in the brain. They cause nerve damage and result in neurological conditions such as dementia, coordination issues, cognitive decline, and death. Animals can suffer from other types of TSEs. Examples include bovine sponge encephalopathy (BSE) (otherwise known as "mad cow disease") in cattle, scrapie in sheep, and chronic wasting disease (CWD) in deer or elk. CJD occurs most in people who are over 65 years old.

What causes CJD?

- CJD occurs when normal proteins in the brain misfold and become abnormal. These misfolded, abnormal proteins are called prions. When these proteins misfold, they cause other similar proteins to become abnormal as well. This leads to a harmful buildup of the abnormal proteins.
- The disease can take a long time to develop before symptoms are noticeable. Their onset can range from 15 months to decades after the introduction of the abnormal form of prion protein.

Types of CJD?

- **Sporadic CJD** occurs when normal proteins in the body become abnormal for an unknown reason. This type of CJD makes up nearly 90% of all CJD cases.
- **Familial (or genetic) CJD** occurs when a person inherits a gene from a parent that leads to the development of abnormal proteins. This form of CJD makes up approximately 10% of cases.
- **Acquired (or iatrogenic) CJD** is a form of the disease that occurs when a person comes in contact with the abnormal protein. The contact occurs during a surgical procedure from contaminated medical equipment. Due to advances in sterilization methods, donor screening, and surgical techniques, cases of acquired CJD are now very rare.
- **Variant CJD** is a rare type of CJD that is caused by eating beef from cattle infected with BSE. The majority of reported cases were seen in the United Kingdom (U.K.) as a result of an outbreak the 1980s. While there have been four cases of vCJD in the United States, none of them were due to an exposure within the country.

What are the signs and symptoms?

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

What are the testing and treatment options?

- There are no current treatment options for CJD; it is always fatal. Health care providers can offer services to keep people with CJD comfortable.
- Advances in diagnostic testing now can provide a likely diagnosis through electroencephalogram (EEG) testing, cerebrospinal fluid analysis, and magnetic resonance imagery (MRI). The only way to confirm a diagnosis and definitively distinguish between sporadic CJD and familial CJD is through an autopsy.

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 **familial CJD**.
- You cannot get CJD through normal, everyday contact with an affected person. **Acquired CJD** only occurs under very specific circumstances that are very rarely present.
- **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 still occur. The U.S. has taken steps to prevent sick cattle from entering the country's food chain.

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

- As of now, the Centers for Disease Control and Prevention (CDC) and World Health Organization (WHO) have not identified CWD in humans. The only known time an animal prion disease affected humans is vCJD by eating beef from BSE-infected cattle.
- As a precautionary measure, people should not eat any part of a deer or elk that are acting strangely, appearing thin, or have other signs of CWD. CWD-causing prions can be found at low levels in the muscles of affected animals.
- Generally, it's recommended that brain, spinal cord, eyes, spleen, tonsils, and lymph nodes from any deer or elk not be eaten. The CWD prion is found in these locations at higher levels in infected animals. Testing of harvested deer or elk can be done though the Wisconsin Department of Natural Resources.

Wisconsin Department of Health Services | Division of Public Health
BUREAU OF COMMUNICABLE DISEASES
https://www.dhs.wisconsin.gov/dph/bcd.htm | DHSDPHBCD@dhs.wi.gov