



CREUTZFELDT – JAKOB DISEASE (CJD) AND MAD COW DISEASE

What is CJD?

CJD is a group of rare diseases called “transmissible spongiform encephalopathies (TSE).” These diseases attack the central nervous system in people and some animals, and invade the brain. The disease progresses rapidly and is always fatal. Death usually occurs within one year of onset. CJD is very rare; there is about one reported case per one million people worldwide, each year.

What is Mad Cow Disease (Bovine Spongiform Encephalopathy or BSE)?

Mad Cow Disease is a progressive neurological (brain) disorder of cattle. The disorder causes the cow to act strangely and lose its ability to do normal things, such as walk. Infected cows act “mad,” which sometimes means mentally ill. There is strong evidence that Mad Cow results from feeding cows BSE-contaminated feed.

Because BSE was a problem in the United Kingdom, the United States enacted rules to prevent live cows and some cow products from entering this country. The United States has had three cases of BSE in cows: one each in 2003, 2005 and 2006. The government assured that these cattle would never be sold for human consumption.

Is CJD the same as Mad Cow Disease?

No. CJD is not related to Mad Cow Disease (BSE). Although they are both considered TSE's, only people get CJD and only cattle get Mad Cow disease.

What causes CJD?

CJD is caused by a protein called a prion. A prion is a disease-causing agent that is not a germ like a bacteria or virus. Prions can cause other proteins in the brain to fold into unusual shapes, causing the death of brain cells. When this occurs, it makes the brain look spongy under a microscope. There are four forms of CJD:

1. Sporadic (sCJD) – Sporadic CJD accounts for about 85-90 percent of all cases for which the cause is unknown. It usually occurs in persons over 50 years of age. The duration of illness is short.
2. Familial or Inherited – Familial or Inherited CJD accounts for 5-15 percent of all cases. These cases occur at an earlier age than Sporadic CJD. The length of illness is longer.
3. Acquired or Iatrogenic – Acquired or Iatrogenic CJD accounts for about 1 percent of all cases in which there has been contamination with tissue from an infected person. This has occurred from the use of contaminated neurosurgical instruments, transplant of contaminated tissues, or administration of human hormones extracted from the organs of human cadavers.
4. Variant (vCJD) – Variant CJD is thought to be transmitted through the consumption of contaminated meat (i.e., beef from cattle infected with Mad Cow Disease). This form of CJD is very rare. Since it was first reported in 1996, there are no known cases of vCJD that were acquired in the United States.



Frequently Asked Questions

What are the symptoms of CJD?

Symptoms of CJD vary from person to person. Early symptoms may include memory loss, mental status changes, impaired coordination and visual disturbances. As the disease progresses, there may be pronounced mental decline, involuntary movements, profound weakness and ultimately, coma.

How is CJD diagnosed?

Diagnosis is very difficult. Doctors make diagnoses based on clinical observation and progression of the disease. Doctors can test electrical activity of the brain and examine spinal fluid to aid in the diagnosis. However, diagnosis can be **confirmed only through a brain biopsy** (examination of the brain before death) or **autopsy** (examination of the brain after death). In addition to providing a confirmatory diagnosis, autopsies help medical professionals and researchers learn more about CJD.

How is CJD treated?

There is no specific treatment or cure for CJD. The disease progresses rapidly and is always fatal. Death usually occurs within one year of onset of illness. Health providers and caregivers can ease patients' symptoms and limit their discomfort.

References and Sources of Additional Information:

- Centers for Disease Control and Prevention: <http://www.cdc.gov/ncidod/dvrd/cjd/>
- [Control of Communicable Diseases Manual](#), 18th Edition, 2004, American Public Health Association.
- Creutzfeldt-Jakob Disease Foundation, Inc., <http://www.cjdfoundation.org/>
- National Prion Disease Pathology Surveillance Center: <http://www.cjdsurveillance.com/>
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http://www.who.int/csr/resources/publications/bse/WHO_CDS_CSRAPH_2000_3/en/