WHAT IS CJD?
Creutzfeldt-Jakob Disease (CJD) is a rare, fatal brain disorder within a group of illnesses called prion diseases. The incidence of CJD cases worldwide is one to two cases, per million individuals, per year. In the United States this statistic translates to nearly 500 new cases per year. There is one CJD death per every 6,000 to 10,000 deaths in the U.S. each year. Eighty-five percent of CJD cases are sporadic, meaning there is no known cause at present.

There are three types of prion disease:
- Sporadic (sCJD)
- Genetic (also known as familial)
- Acquired (variant CJD; vCJD and iatrogenic CJD; iCJD)

In the early stages of the disease, CJD patients may exhibit failing memory, behavior changes, impaired coordination, and/or visual disturbances. As the illness progresses, mental deterioration becomes more pronounced, and involuntary movements, blindness, weakness of extremities, and, ultimately, coma may occur. CJD usually occurs later in life and typically leads to death within a few months to one year following the onset of symptoms.

HELPLINE 1.800.659.1991

TYPES/CAUSES OF CJD

Sporadic CJD
- Cause unknown — the normal prion protein spontaneously misfolds into a disease-causing form and induces other normal prion proteins around it to misfold, in a similar pattern to other protein-related neurodegenerative diseases such as Alzheimer’s or Parkinson’s disease

Genetic Prion Disease
- Caused by a genetic mutation of the prion protein gene
- There are approximately 40 prion disease mutations, which cause varying symptoms, including the following diseases:
  - Genetic (familial) CJD (gCJD)
  - Fatal Familial Insomnia (FFI)
  - Gerstmann-Sträussler-Scheinker Disease (GSS)

Iatrogenic CJD has been transmitted by:
- Contaminated neurosurgical instruments
- Contaminated dura mater transplants obtained from cadavers
- Contaminated corneal transplants
- Contaminated human growth hormone obtained from cadavers

Variant CJD has been transmitted by:
- Beef contaminated by bovine spongiform encephalopathy (BSE) (or mad cow disease)
- Contaminated blood or blood plasma transfusion from individuals with vCJD

The sporadic form of CJD is the most prevalent form of CJD affecting nearly 500 new people in the United States each year. The genetic form of prion disease accounts for 10-15% of cases. Acquired CJD, which includes iatrogenic and variant CJD, accounts for less than 1% of all cases. As of 2018, there are no known cases of vCJD that have been acquired within the United States. More information can be found on www.cjdfoundation.org.

Diagnosis of prion disease is challenging and is often made from clinical observation and the process of elimination of other diseases. The diagnosis of CJD can only be definitively confirmed through a brain biopsy or autopsy. (If CJD is suspected, brain biopsy is not recommended.)

Autopsy determines the type of prion disease and whether there is a genetic mutation. Key diagnostics for prion disease include electroencephalogram (EEG), brain MRI, and cerebral spinal fluid (CSF), with the Real-Time Quaking Induced Conversion (RT-QuIC) test, as well as tests for 14-3-3 and Tau proteins. One of the most important parts of diagnosing prion disease, however, is ruling out other treatable conditions. For more information, call 800-659-1991.
PUBLIC HEALTH CONCERNS

CJD is NOT “Mad Cow Disease.” Bovine Spongiform Encephalopathy (BSE), the technical term for Mad Cow Disease, occurs only in cows. The first documented case of BSE found in the United States occurred in Washington State in December 2003 in a cow imported from Canada. The first endemic case found in cattle was announced in 2005 in Texas.

Eating beef contaminated with BSE can cause the variant form of CJD (vCJD) in humans. vCJD usually affects young people. No cases of vCJD have been thought to have been acquired within the United States.

A deeper understanding of all forms of CJD would provide vital direction to scientists in studying prion disease, improve public health, and support the ultimate goal of finding a cure. In particular, brain autopsy for suspected CJD patients is critical to learning where the disease originates, how it progresses, and how to treat it.

WWW.CJDFOUNDATION.ORG

BLOOD SAFETY

The American Red Cross (ARC) does not accept blood donations from individuals who have spent six months or more in Europe from 1980-96, have received a dura mater (brain covering) transplant or human pituitary growth hormone from cadavers, or have had a blood relative with prion disease. There is no epidemiological evidence that sporadic or genetic prion diseases are transmissible through blood, however, the ARC is taking strict precautions due to the theoretical risk. The first case of suspected blood transfusion transmission occurred in the UK in 2003 through blood donated from a patient with pre-symptomatic variant CJD. Studies by the American Red Cross and others have found no evidence for transmission of non-variant forms of prion disease by blood transfusion.

FUNERALS

There are no special interment, entombment, inurnment, or cremation requirements for patients with CJD. Interment of bodies in closed caskets does not present a significant risk of environmental contamination and cremated remains can be considered sterile, as the infectious agent does not survive incineration-range temperatures. Information for funeral and crematory practitioners is available on the CDC website and from the CJD Foundation and National Prion Disease Pathology Surveillance Center.

CHRONIC WASTING DISEASE (CWD)

Chronic Wasting Disease (CWD), a prion disease in deer and elk (i.e., cervids), appears to be environmentally contagious among members of its own species. First recognized in 1967 in Colorado, CWD has since spread from 2 states in 2000 to 24 states in 2018. Affected prions from the body fluids of deer and elk have been shown to be able to be taken up into the roots and leaves of plants as well as soil. Therefore, fields contaminated by a cervid herd can later spread CWD to other herds.

CWD has not been proven to cross the species barrier. However, studies in progress seem to demonstrate the potential for CWD from the brain and meat of affected cervids to be transmitted to swine and macaques (monkeys) through feeding. Health and wildlife authorities continue to monitor and study this possible threat to public health.

WWW.CJDFOUNDATION.ORG

THE CJD FOUNDATION

Our Contact Information
HelpLine: 1.800.659.1991
HelpLine Email: help@cjdfoundation.org
Website: www.cjdfoundation.org

We are available to help you with questions and concerns Monday through Friday, 9 am to 5 pm ET. Messages left on our voicemail after hours concerning patients will be returned evenings and weekends.