

## Creutzfeldt-Jakob Disease

### Maine CDC Reportable Condition (within 48 hours) 1-800-821-5821

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#### Definition

Creutzfeldt-Jakob disease (CJD) is a rare, worsening, fatal brain disorder. CJD is caused by a type of protein called a prion. Prion proteins occur in both a normal form, which is a harmless protein found in the body's cells, and in an infectious form, which causes disease. Prion diseases can be found in human and in animals.

CJD exists in four forms:

- **Sporadic CJD (sCJD):** also called spontaneous, for which the cause is not known. This is the most common type of CJD and accounts for at least 85 percent of cases.
- **Familial CJD (fCJD):** also called genetic or inherited. About 5 to 10 percent of cases of CJD in the United States are this type.
- **Acquired/Iatrogenic CJD:** is transmitted by exposure to brain or nervous system tissue, usually through certain medical procedures. Since CJD was first described in 1920, fewer than 1 percent of cases have been this type.
- **Variant Creutzfeldt-Jakob disease (vCJD):** is a new disease that was first described in March 1996. It is different from the classic forms of CJD. Variant CJD affects younger patients, has a longer course of illness, and is strongly linked to exposure, probably through food, to a disease known as "Mad Cow Disease."

#### Signs and symptoms

The symptoms of vCJD include noticeable social and behavioral changes, painful extremes of sense and touch, and slowing of brain function. CJD symptoms include dementia and early neurologic signs.

#### Transmission

CJD is usually sporadic with no known transmission. Occasionally transmission occurs through contact with infected neural tissue, either from animals such as cattle, or through neuro surgery or autopsy contact with human with CJD. There is no human to human transmission of CJD.

#### Diagnosis

Diagnosis is based on clinical features together with laboratory testing such as 14-3-3 tau protein tests. Definitive diagnosis cannot be completed until after death.

#### Treatment Recommendations

- There is no known treatment of CJD and all forms are fatal.

#### Role of the School Nurse

##### Prevention

- Educate students and staff on CJD

##### Exclusions

- None

##### Reporting Requirements

- Suspected cases of CJD should be reported within 48 hours – report to 1-800-821-5821

##### Resources

- Maine DOE website <http://www.maine.gov/education/sh/>
- Fact sheet can be found at: <http://www.maine.gov/dhhs/mecdc/infectious-disease/epi/shm/index.shtml>
- Federal CDC CJD website <http://www.cdc.gov/ncidod/dvrd/cjd/>