

Creutzfeldt-Jakob Disease (CJD)

Agent: Believed to be caused by a prion protein

Mode of Transmission: The majority of CJD cases (80%-90%) are sporadic CJD, with no known source. A small percentage of cases (5%-15%) may be due to heredity (familial CJD) or exposure to organ tissue contaminated with the prion (iatrogenic CJD). Classic CJD includes sporadic CJD, familial CJD, and iatrogenic CJD. A form of the disease, variant Creutzfeldt-Jakob disease (vCJD), is thought to be transmitted through ingestion of beef from cattle with bovine spongiform encephalopathy (BSE, commonly referred to as mad cow disease).

Signs/Symptoms: Symptoms may begin with confusion, and they rapidly progress to a wide range of neurological signs and symptoms, including loss of coordination and dementia.

Prevention: Organ and tissue transplants from infected individuals should be avoided. For protection against vCJD, the federal government has regulations in place to prevent the spread of BSE in the United States.

Other Important Information: vCJD occurs in younger individuals, while classic CJD occurs more often in older individuals and has a slower progression. In Virginia, CJD is reportable when it occurs in persons under 55 years of age.

Two cases of Creutzfeldt-Jakob disease in persons less than 55 years of age were reported in Virginia during 2014. Both cases were diagnosed with classic CJD with no known exposures for either case. The two cases occurred in white females between the ages of 30 and 55 years. Prior to 2014, the last reported case occurred in 2007 in a white male in the 30-39 year age group, and the infection was determined to be classic CJD. Ten cases of classic CJD infection have been reported in Virginia residents less than 55 years of age since 1995.

The only case of vCJD ever diagnosed in a Virginia resident occurred in 2006. Based on the patient's history, it was determined that the infection most likely occurred from consumption of contaminated cattle products while living as a child in Saudi Arabia. This was one of four cases of vCJD reported in U.S. residents. The most recent vCJD case in the U.S. occurred in 2014 in a Texas resident, who is thought to have acquired the infection while residing overseas.