Creutzfeldt-Jakob Disease (CJD)

Agent: Believed to be caused by a protein called a prion
Mode of Transmission: The disease is not spread through contact with a person who has CJD except that it can be spread if surgical instruments used on a CJD patient are not thoroughly sterilized and then are used on someone else. The majority of CJD cases (about 85%) are sporadic CJD, with no recognizable source. A small percentage of cases (5-15%) develop due to inheritance of a mutated prion protein gene (familial CJD). Very rarely, exposure to organ tissue contaminated with the prion can result in transmission (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: Classic CJD symptoms may begin with confusion, and they rapidly progress to a wide range of neurological signs and symptoms, including loss of coordination and dementia. The symptoms of vCJD begin with prominent psychiatric and behavioral symptoms, impairment of senses, and delayed neurological signs. The disease is always fatal, with death usually occurring within one year of onset of illness.
Prevention: Organ and tissue transplants from infected individuals should be avoided. Hospitals should take extra precautions to dispose of or sterilize surgical instruments used on CJD patients. 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.

One case of classic CJD was diagnosed in Virginia in 2016 in a person less than 55 years of age with no known exposures. The person died as a result of this condition. There were no cases of classic CJD reported in Virginia in 2015. In 2014, two classic CJD cases were identified in Virginia, both had no known exposures. Eleven cases of CJD have been reported in Virginia residents less than 55 years of age since 1995. All but one case was diagnosed with classic CJD.

The only case of vCJD diagnosed in a Virginia resident less than 55 years of age occurred in 2006. Based on the patient's history, the infection most likely occurred from consumption of contaminated cattle products as a child while living in Saudi Arabia. This is 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. This person is thought to have acquired the infection while living overseas.