State of Alaska Epidemiology **Bulletin**

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Creutzfeldt-Jakob Disease and Other Prion Diseases

Background
Creutzfeldt-Jakob disease (CJD) is a rare, fatal, neurologic condition of humans that is categorized as a prion disease. In the U.S., around 250 cases occur annually; approximately one person out of a million dies from CJD each year. In the early stages of CJD, patients may have behavioral changes, lack of coordination, and visual disturbances. As illness progresses, mental deterioration becomes pronounced. Although typical clinical and EEG (electroencephalogram) features and a positive 14-3-3- test of cerebrospinal fluid can suggest a diagnosis of CJD antemortem, definitive diagnosis is made by postmortem examination of the brain.

What are prion diseases?
Prion diseases (also known as transmissible spongiform encephalopathies) result in microscopic vacuolization, or spongiform degeneration of brain tissue and accumulation of an abnormal form of a protein called prion protein. Prions are poorly characterized infectious protein-like agents that lack detectable nucleic acid, are resistant to procedures that inactivate viruses, and do not elicit an immune response.

There are three general categories of human prion diseases within each category of prion disease, there are different phenotypes of CJD.

Sporadic: five phenotypes of CJD are recognized in this category; the vast majority of all CJD cases are sporadic.

Familial: includes several inherited prion diseases, e.g., fatal familial insomnia, Gerstmann-Sträussler-Scheinker syndrome, and familial CJD.

Acquired: includes cases of iatrogenic CJD, kuru and variant CJD (vCJD). Iatrogenic CJD cases occur in persons receiving CJD-infected tissue implants, such as dura mater or corneal grafts, or contaminated human growth hormone.

Is CJD a "communicable" disease?
CJD is not communicable to other persons through casual contact. Decades of experience and several studies indicated that classic CJD may not be transmitted via blood and blood products. However, because vCJD is a new disease, the U.S. Food and Drug Administration (FDA) enacted stringent guidelines for deferring donors who may be at risk for developing vCJD, e.g., persons who lived 3+ cumulative months in the United Kingdom (U.K.) from 1980-1996, or 5+ years in other European countries from 1980-present.

What is variant CJD (vCJD)?
vCJD has well-defined, consistent clinical and pathological features that distinguish it from sporadic CJD. In 1996, the first cases of vCJD were reported in the U.K.; since then, the U.K. has reported 127 deaths. There is strong evidence that vCJD was acquired by persons who consumed cattle affected by bovine spongiform encephalopathy (BSE). Since the 1980s, BSE has occurred in epidemic proportions among cattle in the U.K., and the BSE outbreak has spread to many other European countries, Japan, and Israel. **No locally-acquired vCJD cases have occurred in the U.S.** The one probable case of vCJD in the U.S. reported in 2002, is in a 22-year-old Florida resident who was born in and spent her first 13 years of life in the U.K. There have been no vCJD cases in the world that did not have a history of exposure within a country where BSE has been documented.

What is BSE or "Mad Cow" disease?
BSE is a cattle prion disease that causes a fatal, progressive, neurologic condition. Because of the former practice in the U.K. whereby cattle were fed rendered sheep carcasses, BSE is hypothesized to have developed in cattle that had ingested sheep affected with scrapie, a sheep prion disease.

Have U.S. cattle been affected by BSE?
There has **never** been a case of BSE in the U.S. The U.S. Department of Agriculture (USDA) has developed strict guidelines to prevent BSE from coming into the U.S., as well as a response plan to manage a suspect case. In addition, some former practices that may have contributed to the development of BSE in the U.K. were not common in the U.S.

What is Chronic Wasting Disease (CWD)?
Chronic wasting disease (CWD) is another animal prion disease documented among elk and certain deer populations in the western U.S. and Canada. There is currently no evidence that CWD can infect moose, cows, reindeer, or sheep. There is also no evidence to suggest that CWD can cause disease in humans; however, it is important to continue CJD surveillance in CWD-infected areas to assess any possible association between these two diseases.

Has CWD been found in Alaska?
To date, CWD has not been detected among animals in Alaska. The Alaska Department of Fish and Game (ADFG), in conjunction with USDA, have developed a testing program for some hoofstock. Of the 60+ moose and 10+ reindeer tested so far, none have had evidence of CWD. Surveillance for CWD in hunter-harvested deer and elk will begin this fall.

How many deaths from CJD have occurred in Alaska?
From 1977-2001, CJD was listed as a primary or contributory cause of death on the death certificates of seven Alaska residents. CJD is not explicitly listed as a disease reportable to the Section of Epidemiology; therefore current statistics on the number of CJD cases in Alaska are not available.

Where can human prion diseases be diagnosed?
To facilitate neuropathologic studies of suspected prion diseases in humans, the U.S. Centers for Disease Control and Prevention (CDC), in collaboration with the American Association of Neuropathologists, established the National Prion Disease Pathology Surveillance Center (NPDPSC) during 1996/1997 at the Division of Neuropathology of Case Western Reserve University. Healthcare providers are encouraged to use the free services at NPDPSC to confirm a diagnosis of CJD. Contact the Section of Epidemiology (907-269-8000) for more information and assistance in arranging a NPDPSC consult.

Additional resources:
NPDPSC http://www.cjdsurveillance.com/
USCDC http://www.cdc.gov/ncidod/diseases/cjd/cjd.htm
UK CJD Policy Unit http://www.doh.gov.uk/cjd/index.htm
USFDA http://www.fda.gov/cber/gdlns/cjdvcjdq&a.htm
USDA http://www.aphis.usda.gov/lpa/issues/bse/bse.htm1
ADFG
http://www.state.ak.us/adfg/wildlife/geninfo/disease/cwd.htm
or e-mail mailto:cwdinfo@fishgame.state.ak.us

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