State of Alaska Epidemiology Bulletin

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# Human Prion Diseases Now Reportable in Alaska

## Background

Prion diseases (PDs) or transmissible spongiform encephalopathies (TSEs) are a family of rare progressive neurodegenerative disorders that affect both humans and animals. PDs are distinguished pathologically by spongiform changes associated with neuronal loss and the failure to produce an inflammatory response. A prion is a transmissible agent that is able to induce abnormal folding of cellular proteins in the brain, leading to brain damage and the characteristic signs and symptoms of the disease. Known prion diseases have long incubation periods, primarily involve the central nervous system, and are always fatal.

The prototypic human PD is kuru. Other examples include Creutzfeldt-Jakob disease (CJD) and variant Creutzfeldt-Jakob disease (vCJD). Examples of animal prion diseases include bovine spongiform encephalopathy (BSE) in cows, scrapie in sheep, and chronic wasting disease (CWD) in deer, moose, and elk. As of December 29, 2006, suspected or confirmed cases of human prion diseases are reportable by health care providers and laboratories to the Alaska Section of Epidemiology (SOE).

The estimated annual rate of PDs in the United States is one case per one million persons; this rate has remained stable for over 20 years. Globally, 200 patients have been diagnosed with vCJD, including three residents of the United States, all of whom had previously lived for extended periods in the United Kingdom or the Middle East.

## Historical Data on PDs in Alaska

From 1977-2005, Alaska death certificate data listed CJD as a primary or contributory case of death for 10 Alaskans. However, because PDs were not reportable until recently, it is unknown how many of these cases had autopsies or were laboratory-confirmed. Making PDs reportable will allow SOE to estimate rates of PDs for Alaska in the future.

While 12 cases of BSE have been identified in cattle in the United States (3) and Canada (9), to date, neither BSE nor CWD have been documented in Alaska's domestic or wild animal populations. Surveillance and testing for BSE and other PDs in domestic animals is coordinated through the Alaska Department of Environmental Conservation (ADEC). Surveillance and testing for CWD in wild animals is coordinated through the Alaska Department of Fish and Game (ADFG).[1]

## Modes of Transmission

Although some PDs can occur spontaneously in people, there are three distinct potential environmental sources that make PDs a public health concern: 1) iatrogenic transmission of CJD or vCJD, including via contaminated a) corneal or dural grafts, b) pituitary derived hormones, c) surgical equipment, or d) blood; 2) occurrence of vCJD from consumption of BSE-contaminated food products; and 3) the theoretically possible transmission of CWD to humans.[2] The likelihood of cases from the first two sources appear to be declining given safeguards that have been put into place, including decontamination of surgical instruments[3] and screening of blood donors. The third source is only theoretical at this point because no studies have shown that CWD has caused disease in humans.[4]

## Laboratory Diagnosis

Clinical evaluation of patients with a suspected PD may include an electroencephalogram, a magnetic resonance imagining study, and cerebrospinal fluid (CSF) 14-3-3 protein testing. Elevated levels of 14-3-3 protein have been found in CSF of some patients with CJD, but have also been found in patients with subarachnoid hemorrhage, stroke with acute infarction, brain neoplasm or paraneoplastic disorders. These tests may support the diagnosis of PD, but no test is currently available to confirm the diagnosis without neuropathologic confirmation. Post-mortem examination of brain tissue is the only definitive means of diagnosing PDs.

## Case Reporting Criteria

Health care providers should notify SOE of any patients with an otherwise unexplained, subacute, progressive dementia and at least one of the following neurologic features: myoclonus, visual or cerebellar signs, pyramidal or extrapyramidal signs, or akinetic mutism.

## Public Health Response

Public health follow-up for each suspected or confirmed PD case may vary. SOE will work with health care providers to assist with diagnostic testing. The National Prion Disease Pathology Surveillance Center (NPDPSC) at Case Western University in Ohio provides free testing to confirm the diagnosis of PDs. Additionally, SOE may interview the patients and their relatives or friends to obtain epidemiologic information that could assist with specific PD classification.

## Recommendations

1. Health care providers should contact SOE if they suspect a patient may have a PD. Call 907-269-8000, Monday through Friday 8AM-5PM. SOE will work with NPDPSC to arrange for autopsies.
2. Animals suspected to have PDs should be reported to the appropriate authority. For domestic animals, contact the ADEC State Veterinarian at 907-375-8214. For wildlife, contact the ADFG Wildlife Veterinarian at 907-459-7257.

## References

1. Alaska Dept of Fish & Game, Division of Wildlife Conservation, Chronic Wasting Disease Program. Available at: http://www.wildlife.alaska.gov/index.cfm?adfg=disease.cwd.
2. Belay ED, Schonberger LB. The public health impact of prion diseases. *Ann Rev Pub Health* 2005;26:191-212.
3. World Health Organization. WHO Infection Control Guidelines for Transmissible Spongiform Encephalopathies. 1999. Available at: http://www.who.int/csr/resources/publications/bse/whocdscsraph2003.pdf.
4. Belay ED, et al. Chronic wasting disease and potential transmission to humans. *Emerg Inf Dis* 2004;10:977-84.

## Web Resources

NPDSPC: http://www.cjdsurveillance.com
CJD Foundation: www.cjdfoundation.org
Centers for Disease Control and Prevention (CDC): http://www.cdc.gov/ncidod/dvrd/prions/index.htm

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