Council of State and Territorial Epidemiologists
Position Statement

04-ID-09

Committee: **Infectious Disease**

Title: **Enhancing Surveillance and confirmatory testing of Creutzfeldt-Jakob and other Human Prion Diseases**

Statement of the Problem:
National surveillance for Creutzfeldt-Jakob disease (CJD) in the United States has been performed by periodic review of multiple cause-of-death data obtained from the National Center for Health Statistics, Centers for Disease Control and Prevention (CDC). In 1996, surveillance for variant CJD (vCJD) cases was conducted in five Emerging Infections Program sites (MN, OR, CN, Atlanta, San Francisco), covering a population of 16.3 million. Subsequently, CDC enhanced vCJD surveillance nationally by initiating more extensive follow-up investigations with state and local health departments of CJD cases <55 years of age. Currently, approximately 50% of states mandate reporting of human prion diseases including CJD; some states collect data by mandated reporting of rare diseases of public health significance or by routine review of death certificate information. Although review of death certificate data has been shown to capture approximately 80% of CJD deaths in the United States, more than half of the cases lack documentation of histopathologic confirmation of the diagnosis. The challenge of obtaining better data and surveillance for prion diseases depends on successful promotion of the importance of collection of autopsy tissues for laboratory confirmation.

During 1996-1997, the CDC and the American Association of Neuropathologists established the National Prion Disease Pathology Surveillance Center (NPDPSC) as a national reference center for advanced neuropathologic and biochemical diagnosis of human prion diseases. Although testing at NPDPSC is free of charge, many clinicians and public health agencies are unaware of this service.

The emergence of vCJD and its zoonotic relationship to bovine spongiform encephalopathy (BSE) in Europe and other countries, the recent identification of BSE in North America, and the expanding range of chronic wasting disease in North American deer and elk highlight the importance of animal prion diseases and their potential impact on humans. While most prion diseases seem to have species barriers, emphasis on disease surveillance and laboratory confirmation are needed to enhance understanding of the pathology and epidemiology of human prion diseases and to implement a system of detecting emerging human prion diseases.

To enhance surveillance of human prion diseases, public health should work to educate health care providers about the NPDPSC and enlist their cooperation in educating family members of patients suspected to have CJD, about the importance of confirming suspected human prion disease by pathologic analysis.

Statement of Desired Action to be Taken:
1) CSTE/NASPHV believes that any case of confirmed vCJD or newly described human prion disease acquired in North America has significant public health importance.

2) CSTE/NASPHV recommends that state and local public health officials facilitate arrangement of brain autopsy on suspected or clinically diagnosed cases of prion disease in humans and submission of the tissues to the NPDPSC for analysis.

3) CSTE/NASPHV endorses periodic (e.g. annual, biannual) review by state, territorial or local public health agencies of collected mortality data and/or other comparable surveillance data to identify cases of, and monitor trends in CJD and other human prion disease.

4) CSTE/NASPHV recommends that suspected, probable and confirmed cases of human prion diseases (particularly among persons less than 55 years of age) be investigated by state, territorial, local or federal public health agencies to collect epidemiologic, laboratory and clinical data.

5) CSTE/NASPHV encourages educational efforts directed at clinicians involved in the care or evaluation of patients with suspected or confirmed CJD or other human prion diseases (neurologists, neuropathologists, pathologists, medical examiners, etc.) to:

a) Increase clinician awareness of the clinical features of human TSE, including variant CJD.

b) Increase the proportion of brain autopsies performed on decedents with suspected or confirmed prion disease

c) Increasing awareness and use of prion disease diagnostic services available from the NPDPSC,

d) Increase the accuracy of data entered on death certificates of CJD decedents

Goals of Surveillance:

1. Describe more accurately the burden of endemic prion diseases of humans.
2. Develop capacity to identify zoonotic prion disease cases in humans such as vCJD
3. Develop capacity to identify the emergence of novel prion diseases in humans

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Methods for Surveillance: See 2-5 above

Case Definition:

Period of Surveillance: Indefinite

Background:

Transmissible spongiform encephalopathies (TSE) are a group of fatal neurodegenerative disorders that affect both humans and animals. TSEs are believed to be caused by an abnormal isoform of a cellular protein, known as a prion. Creutzfeldt-Jakob disease (CJD) is the most common TSE of humans and occurs at a worldwide rate of approximately one case per million population annually. About 85% of CJD cases are sporadic, and the remaining 15% are familial, or the result of iatrogenic exposure through contaminated neurosurgical instruments or receipt of contaminated cornea, dura mater grafts, or pituitary-derived human growth hormone. Other human TSEs include kuru, Gerstman-Sträussler-Scheinker syndrome, fatal familial insomnia and variant CJD (vCJD), associated with BSE.

CJD has recently received increased attention because of the association between vCJD and the outbreak of BSE. More than 150 cases of vCJD have been reported primarily from Europe. Variant CJD has been associated with human consumption of BSE-contaminated beef products. The only case of vCJD identified to date in the US was in a former resident of the United Kingdom.

Animal TSEs include scrapie in sheep, BSE in cattle, and chronic wasting disease (CWD) in cervids. The detection of cases of BSE in North America in 2003 resulted in global trade restrictions, large economic losses, and widespread concern about food safety. Scrapie has been recognized for many years and is present in sheep flocks in North America and most of the world. Chronic wasting disease has been found only in North America, however, the geographic range

of the disease in cervids has expanded significantly in the past decade. No human disease has been epidemiologically associated with either CWD or scrapie.

Agencies for Response:

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