State of California—Health and Human Services Agency
California Department of Public Health

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TO: Local Health Officers
Local Communicable Disease Controllers

FROM: Infectious Diseases Branch
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Center for Infectious Diseases
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SUBJECT: Surveillance for Transmissible Spongiform Encephalopathies (TSEs) and Creutzfeldt-Jakob Disease (CJD)

As of June 2007, the California Department of Public Health (CDPH) added Transmissible Spongiform Encephalopathies (TSEs), which include Creutzfeldt-Jakob Disease (CJD), to the Title 17, California Code of Regulations, Section 2500's list of reportable diseases and conditions.

Background

TSEs are a group of rare, fatal, progressive neurological disorders that occur in humans and animals. TSEs are prion diseases. CJD is the most common of these diseases, but is still rare with an incidence of one case per million people per year. In California, approximately 30 cases of CJD per year have been noted.

CJD and the other TSEs come in a variety of forms:

1. CJD includes sporadic CJD (sCJD), the classic and most common form (> 80% of CJD), which is of unknown etiology; familial CJD (fCJD) which is genetically inherited; and iatrogenic CJD (iCJD) which has been transmitted through contaminated human pituitary-derived growth hormone, human dura mater allografts, corneal transplants, and neurosurgical instruments.
2. Variant CJD (vCJD) transmitted through consumption of beef products contaminated with the agent of Bovine Spongiform Encephalopathy (BSE or "mad cow disease").

3. Gerstmann-Straussler-Scheinker Syndrome (GSS), a genetically inheritable disease.
4. Fatal Familial Insomnia (FFI), which is also a genetically inheritable disease.
5. Kuru, a TSE described in New Guinea tribal members transmitted through previously practiced ritual cannibalism at funerals.

Transmission of CJD and other TSEs from infected individuals is very rare. TSEs are not transmitted through the air or through touching or most other forms of casual contact. However, iatrogenic person-to-person transmission has occurred previously as mentioned above, and infection control guidelines are available for handling potentially infectious materials and contaminated surgical instruments (http://www.cdc.gov/ncidod/dvrd/cjd/qa_cjd_infection_control.htm).

Variant CJD, though often confused with other forms of CJD, is a distinctly different clinical entity. In 1996, the first cases of vCJD were reported in the United Kingdom. Transmission is believed to occur primarily through the consumption of tissue from cattle infected with BSE, commonly known as Mad Cow Disease. To date, there have been only three cases of vCJD in the United States, all linked to previous exposures abroad (two while in the U.K. and one while in Saudi Arabia) (http://www.cdc.gov/ncidod/dvrd/vcjd/other/News_06122008.htm). There have not been any domestically acquired vCJD cases in the United States. vCJD, but not sCJD, may be transmissible through blood transfusion from blood donated by an infected individual. For this reason, individuals who lived in the U.K. between 1980 and 1996 are currently banned from donating blood in the U.S.

There are currently no effective treatments for TSEs or CJD. The only clinical treatment trial for CJD in the U.S. is currently being conducted through the University of California, San Francisco (UCSF) Memory and Aging Center. A website by this Center (www.memory.ucsf.edu/cjd) provides medical information and related resources on CJD for the public and medical community.

For several years, the CDPH Infectious Diseases Branch has worked closely with the California Emerging Infections Program (CEIP) on a CJD surveillance project in California, particularly for patients under 55 years of age. Information on the CEIP CJD Surveillance Project can be found at http://ceip.us/cjd.htm. CDPH and CEIP will continue to collaborate on this surveillance as CJD and TSEs are now reportable.

Surveillance

We implemented surveillance for CJD and TSEs in California to assess the burden of prion disease, monitor trends, and possibly detect any novel forms of disease or transmission, such as with vCJD. TSEs are difficult to diagnose. Their diagnosis is generally based on a number of clinical features and confirmed by pre- or post-mortem neuropathological examination when possible. Currently, examination of brain tissue

obtained from a biopsy or an autopsy is the only way to absolutely confirm the diagnosis of CJD or the type of CJD.

We have previously sent a letter to neurologists and neuropathologists in CA to encourage them to obtain brain biopsy or autopsy in all clinically diagnosed or suspected CJD cases regardless of age, to significantly increase the number of cases identified and confirmed by neuropathologic testing. The National Prion Disease Pathology Surveillance Center (NPDPSC) at the Case Western Reserve University in Cleveland, Ohio is the national reference laboratory for human prion diseases. The NPDPSC provides free diagnostic laboratory testing for prion related disease and can arrange for autopsy and shipping of tissue samples through their autopsy network. For details regarding the collection and shipment of clinical specimens, see the NPDPSC's website (http://www.cjdsurveillance.com) or call (216) 368-0587. Alerting clinicians and pathologists to the free services offered by the NPDPSC increases the accuracy of CJD/TSE diagnoses and provides opportunities to learn more about these complex diseases.

We have recently submitted a brief article to the Association of California Neurologists for inclusion in their quarterly newsletter. In this article, we gave basic information on CJD, notified neurologist members that CJD and TSEs are now reportable in CA, and asked them to report cases to their local health department. Again, though the disease may be considered more often in the differential diagnosis and tested for, given the incidence of one case per million population, we expect only approximately 30 cases per year in all of California.

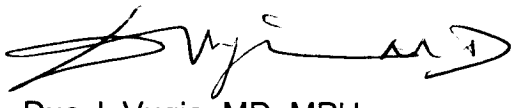
Case Reporting

Local Health Departments may receive notice of TSEs—most likely CJD—from clinicians or may receive abnormal 14-3-3 CSF prion protein laboratory reports or biopsy and autopsy reports from the NPDPSC. Reflecting the complex nature of the disease and its transmission, CDPH's surveillance case definition requires that the diagnosis of CJD be made either by a neurologist or confirmed by biopsy or autopsy tissue. A case report form should be filled out for each pathologically confirmed case or a case diagnosed as CJD or TSE by a neurologist. The new case report form can be found at this weblink
http://www.cdph.ca.gov/pubsforms/forms/CtrldForms/cdph9002.pdf, or at the CDPH Division of Communicable Disease Control (DCDC) Forms Site
[http://www.cdph.ca.gov/pubsforms/forms/Pages/CD-Report-Forms.aspx] under "Creutzfeldt-Jakob Disease (CJD) and Transmissible Spongiform Encephalopathy (TSE)."

Please forward copies of completed forms, as usual, to the Surveillance and Statistics Section of the Infectious Diseases Branch in Sacramento.

As this surveillance is new and some requested information may be difficult to get, we are open to comments and feedbacks when you do get to use the form for a rare case of CJD. If you have any questions about reporting or investigating a case of CJD or TSE, please contact Dr. Margaret Wheeler or Susan Brooks at the California Emerging Infections Program-CJD Surveillance Program at (510) 451-1344.

Thank you,



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