Communicable Disease Case Reporting and Investigation Protocol

TRANSMISSIBLE SPONGIFORM ENCEPHALOPATHY (TSE)
CREUTZFELDT-JAKOB DISEASE (CJD)

I. IDENTIFICATION AND DEFINITION OF CASES
A. Definitions: TSE is the general term for any prion disease occurring in any species. Human TSEs include sporadic Creutzfeldt-Jakob disease, iatrogenic CJD, familial CJD, variant CJD, Gertsmann-Straussler-Scheinker syndrome, and fatal familial insomnia.

B. Clinical Description: All TSEs are progressive neurodegenerative conditions, which are invariably fatal. All are characterized by progressive dementia, myoclonus, and ataxia, with death typically ensuing within 6-7 months after onset.

C. Laboratory Criteria: TSE is diagnosed by neuropathological techniques as outlined below in the case definitions (section VII).

D. Wisconsin Surveillance Case Definition: Case definitions are outlined below in section VII. These case definitions can only be applied by DPH investigators.

II. REPORTING
A. Wisconsin Disease Surveillance Category II – Methods for Reporting: This disease shall be reported to the patient’s local health officer or to the local health officer’s designee within 72 hours of recognition of a case or suspected case, per Wis. Admin. Code § DHS 145.04 (3) (b). Report electronically through the Wisconsin Electronic Disease Surveillance System (WEDSS), or mail or fax a completed Acute and Communicable Disease Case Report (F-44151) to the address on the form.

B. Responsibility for Reporting: According to Wis. Admin. Code § DHS 145.04(1), persons licensed under Wis. Stat. ch. 441 or 448, laboratories, health care facilities, teachers, principals, or nurses serving a school or day care center, and any person who knows or suspects that a person has a communicable disease identified in Appendix A.

C. Clinical Criteria for Reporting: A possible TSE should be suspected and reported in any patient with:
1. Dementia of early onset (<55 years of age) OR
2. Rapidly progressive dementia at any age, with one or more of the following:
   • Movement disorders (e.g., myoclonus, ataxia)
   • Painful sensory symptoms
   • Visual disturbances OR
3. Any case of physician-diagnosed TSE.

D. Laboratory Criteria for Reporting: Any laboratory evidence of a TSE, including positive tests on cerebrospinal fluid for Tau protein, 14-3-3 protein, or RT-QuIC analysis, as well as evidence of prion disease based on testing of brain tissue using IHC, western blot, and/or standard histopathology.

III. CASE INVESTIGATION
A. Responsibility for case investigation: Local health departments should notify the Bureau of Communicable Diseases (BCD) when notified about a suspect case. All TSE and CJD case investigations are conducted by BCD staff.

IV. PUBLIC HEALTH INTERVENTIONS AND PREVENTION MEASURES
A. In accordance with Wis. Admin. Code § DHS 145.05, local public health agencies should follow the methods of control recommended in the current editions of Control of Communicable Diseases Manual, edited by David L.

B. Local health departments **must** notify BCD when notified about a suspect case. Investigations are conducted by BCD staff.

V. CONTACTS FOR CONSULTATION
   A. Local health departments and tribal health agencies:  
      https://www.dhs.wisconsin.gov/lh-depts/index.htm
   
   B. Bureau of Communicable Diseases, Communicable Diseases Epidemiology Section: 608-267-9003

VI. RELATED REFERENCES
   
   
   C. Centers for Disease Control and Prevention website: https://www.cdc.gov/prions/index.html
   
   D. DHS Prion Disease Information for Wisconsin Medical Providers:  
   
   E. DHS Criteria for Reporting a Suspect Case of Prion Disease:  

VII. ATTACHMENTS
   A. Case Definitions
      1. **Creutzfeldt-Jakob Disease** – World Health Organization (WHO) Diagnostic Criteria (see section II [c] above for reporting criteria)
         a. **Sporadic CJD:**
            • **Definite:** Diagnosed by standard neuropathological techniques, and/or immunocytochemically, and/or Western blot confirmed protease-resistant PrP, and/or presence of scrapie-associated fibrils.
            • **Probable:** Progressive dementia and at least two out of the following four clinical features:
              o Myoclonus
              o Visual or cerebellar signs
              o Pyramidal/extrapyramidal signs
              o Akinetic mutism
              AND both of the following:
              o A typical EEG during an illness of any duration; and/or a positive 14-3-3 CSF assay and a clinical duration to death of <2 years
              o Routine investigations should not suggest an alternative diagnosis
            • **Possible:** Progressive dementia and at least two out of the following four clinical features:
              o Myoclonus
              o Visual or cerebellar signs
              o Pyramidal/extrapyramidal signs
              o Akinetic mutism
              AND
              o No EEG or atypical EEG and duration <2 years
         b. **Iatrogenic CJD:** Progressive cerebellar syndrome in a recipient of human cadaveric-derived pituitary hormone, or Sporadic CJD with a recognized exposure risk (e.g., antecedent neurosurgery with dura mater implantation).
         c. **Familial CJD:** Definite or probable CJD plus definite or probable CJD in a first degree relative; and/or neuropsychiatric disorder plus disease-specific PrP gene mutation.
2. Variant Creutzfeldt-Jakob Disease - WHO Diagnostic Criteria
   
a. **Definite Variant CJD:** Neuropathologic examination of brain tissue is required to confirm a diagnosis of variant CJD. The following confirmatory features should be present:
   - Numerous widespread kuru-type amyloid plaques surrounded by vacuoles in both the cerebellum and cerebrum-florid plaques.
   - Spongiform change and extensive prion protein deposition shown by immunohistochemistry throughout the cerebellum and cerebrum.

b. **Suspected Variant CJD:**
   - Current age or age at death <55 years (a brain autopsy is recommended, however, for all physician-diagnosed CJD cases).
   - Psychiatric symptoms at illness onset and/or persistent painful sensory symptoms (frank pain and/or dysesthesia).
   - Dementia, and development > 4 months after illness onset of at least two of the following five neurologic signs: poor coordination, myoclonus, chorea, hyperreflexia, or visual signs. (If persistent painful sensory symptoms exist, > 4 months delay in the development of the neurologic signs is not required).
   - A normal or an abnormal EEG, but not the diagnostic EEG changes often seen in classic CJD.
   - Duration of illness of over six months.
   - Routine investigations of the patient do not suggest an alternative, non-CJD diagnosis.
   - No history of receipt of cadaveric human pituitary growth hormone or a dura mater graft.
   - No history of CJD in a first degree relative or prion protein gene mutation in the patient.

**NOTE – Variant CJD:**

- If a patient has the typical bilateral pulvinar high signal on MRI scan, a suspected diagnosis of variant CJD requires the presence of a progressive neuropsychiatric disorder, and four of the following five criteria: 1) early psychiatric symptoms (anxiety, apathy, delusions, depression, withdrawal); 2) persistent painful sensory symptoms (frank pain and/or dysesthesia); 3) ataxia; 4) myoclonus or chorea or dystonia; and 5) dementia.

- A history of possible exposure to bovine spongiform encephalopathy (BSE) such as residence or travel to a BSE-affected country after 1980 increases the index of suspicion for a variant CJD diagnosis.