Wisconsin Department of Health Services
Division of Public Health
P-01913 (Rev 08/2017)

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.

Heymann, published by the American Public Health Association, and the American Academy of Pediatrics' *Red Book: Report of the Committee on Infectious Diseases*, unless otherwise specified by the state epidemiologist.

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

A. Heymann DL, ed. Prion Diseases. In: *Control of Communicable Diseases Manual*. 20th ed. Washington, DC: American Public Health Association, 2015: 484-490.

B. Pickering LK, ed. Prion Diseases. In: *Red Book*: *2015 Report of the Committee on Infectious Diseases*. 30th ed. Elk Grove Village, IL: American Academy of Pediatrics, 2015: 653-656.

C. Centers for Disease Control and Prevention website: https://www.cdc.gov/prions/index.html

D. DHS Prion Disease Information for Wisconsin Medical Providers: https://www.dhs.wisconsin.gov/publications/p01262.pdf

E. DHS Criteria for Reporting a Suspect Case of Prion Disease: https://www.dhs.wisconsin.gov/publications/p01610.pdf

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:
        - Myoclonus
        - Visual or cerebellar signs
        - Pyramidal/extrapyramidal signs
        - Akinetic mutism

        AND both of the following:
        - 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
        - Routine investigations should not suggest an alternative diagnosis
      - **Possible:** Progressive dementia and at least two out of the following four clinical features:
        - Myoclonus
        - Visual or cerebellar signs
        - Pyramidal/extrapyramidal signs
        - Akinetic mutism

        AND
        - 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.