

Transmissible Spongiform Encephalopathy (TSE)

(Creutzfeldt-Jakob disease)

Last revised June 27, 2011

I. IDENTIFICATION

- A. **BACKGROUND:** TSE is the general term for any prion disease occurring in any species. Human TSEs encompass sporadic Creutzfeldt - Jakob disease (CJD), iatrogenic CJD, familial CJD, and variant CJD.
- B. **CLINICAL DESCRIPTION:** All of these entities 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. **REPORTING CRITERIA / CASE DEFINITION:** Because the actual surveillance case definitions are relatively complex and lengthy, the DPH, in conjunction with an expert advisory panel, has developed reporting criteria that should be used by clinicians to determine whether a potential case should be reported. The case definition needs to be applied only by DPH investigators and appears at the end of this section after the references.

Reporting Criteria – 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 CONFIRMATION:** Diagnosed by neuropathological techniques as outlined below in the case definitions.

E. **CASE CLASSIFICATION:** Confirmed, probable, and possible case definitions are listed below.

II. ACTIONS REQUIRED / PREVENTION MEASURES

A. WISCONSIN DISEASE SURVEILLANCE CATEGORY II:

Report to the patient's local health department either electronically through the Wisconsin Electronic Disease Surveillance System (WEDSS), by mail or fax using an Acute and Communicable Disease Case Report ([F-44151](#)), or by other means within 72 hours upon recognition of a case or suspected case.

B. EPIDEMIOLOGY REPORTS REQUESTED:

- *Electronically* – Report through WEDSS, including appropriate disease-specific tabs
OR
- *Paper Copy* – Acute and Communicable Diseases Case Report ([F-44151](#))

C. **PUBLIC HEALTH INTERVENTIONS:** None for local health departments. Investigations are conducted by DPH staff.

III. CONTACTS FOR CONSULTATION

BCD / COMMUNICABLE DISEASE EPIDEMIOLOGY SECTION: (608) 266-2154 or (608) 267-0401.

IV. RELATED REFERENCES

1. "Creutzfeldt-Jakob Disease" DPH Disease Fact Sheet Series: [View a list of all current Communicable Disease Fact Sheets](#)

2. Heymann DL, ed. Transmissible Spongiform Encephalopathies. In: *Control of Communicable Diseases Manual*. 19th ed. Washington, DC: American Public Health Association, 2008: 216-222.

V. ATTACHMENT – CASE DEFINITIONS (see section I. c. above re. reporting criteria)

WHO Diagnostic Criteria for Creutzfeldt-Jakob Disease (CJD)

1. 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

- 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

2. 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).

3. 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.

WHO Diagnostic Criteria for Variant Creutzfeldt-Jakob Disease in the United States

I. 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.

II. 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 6 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

1. 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, d, e, f and g of the above criteria, 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.

2. 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.