

## **Creutzfeldt-Jakob Disease (CJD)**

### **2007 Case Definition (North Carolina)**

#### **1. Sporadic CJD**

*Confirmed:*

A person who had clinically compatible illness diagnosed by one or more of the following:

- Standard neuropathological techniques
- Immunocytochemically
- Western blot confirmed protease-resistant PrP
- Presence of scrapie-associated fibrils

*Probable:*

A person with progressive dementia **and** at least two of the following four clinical features:

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

**and**

- Typical EEG during an illness of any duration, **or**
- Positive 14-3-3 CSF assay plus a clinical duration to death of <2 years

**and**

- Routine investigation does not suggest an alternative diagnosis

*Suspect:*

A person with progressive dementia **and** at least two of the following four clinical features:

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

**and**

- No EEG **or** an atypical EEG
- Duration to death of <2 years

#### **2. Iatrogenic CJD**

- A person with progressive cerebellar syndrome with a history of receiving human cadaveric-derived pituitary hormone, **or**
- A person with sporadic CJD with history of a recognized exposure risk such as antecedent neurosurgery with dura mater implantation

#### **3. Familial CJD**

A person with confirmed or probable CJD who has a first degree relative with a history of either:

- Confirmed or probable CJD, **or**
- Neuropsychiatric disorder and disease-specific PrP gene mutation.