

# The National Prion Disease Pathology Surveillance Center

*Supported by the Centers for Disease Control and Prevention (CDC)  
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Dear Clinician:

The Centers for Disease Control and Prevention (CDC) mandate to the National Prion Disease Surveillance Center (NPDPS) is to monitor the occurrence of human prion diseases, such as Creutzfeldt-Jakob Disease (CJD). The indispensable method is investigating brain tissue pathology from clinically suspected cases. I wanted to make you aware of the following, which may be helpful to your work with current and future cases:

- Investigating brain tissue after autopsy provides the only method for establishing a definite diagnosis of CJD and pinpointing the disease origin – sporadic, genetic, iatrogenic, or variant (transmitted from “mad cow disease”).
- The 14-3-3 and tau protein tests performed on cerebrospinal fluid (CSF) do not definitively diagnose CJD or other prion diseases. A new CSF test – RT QuIC – with a higher specificity has been developed but will require an extensive validation to determine the ultimate diagnostic precision in clinical settings.
- It is important to note that autopsy allows detection of atypical cases and potential sources of zoonotic infection such as chronic wasting disease of deer and elk.
- Autopsies help to further our understanding of CJD; tissues acquired at autopsy are made available to a number of laboratories involved in the research of prions.
- Ideally, autopsies should be performed within 24 hours post mortem. However, the NPDPS has successfully diagnosed cases autopsied up to six days post mortem.
- ***To facilitate autopsies, the NPDPS offers a free coordination service.*** We make all of the arrangements, including locating the pathologist, arranging for transportation, coordinating with the funeral home, and covering all costs associated with the autopsy.
- Highly trained NPDPS professionals can alleviate some of the stress on the family, which results in more rapid and efficient coordination of the autopsy arrangements, any time after the diagnosis of suspected CJD has been made.
- Our autopsy network includes major health centers throughout the United States, and in most cases the autopsy and necessary transportation can be arranged and completed within 24 hours. In the event of delays (weekends, holidays), the family will be notified of the status.
- NPDPS staff members are available to consult with funeral homes to educate them on how to appropriately handle these cases and best meet the family’s needs and requests.
- All results are sent to the clinicians designated by the family to facilitate answering their questions and to discuss any appropriate steps which need to be taken.
- If the autopsy doesn’t confirm CJD, the NPDPS can help facilitate arrangements for a further workup to arrive at a final diagnosis.

To make autopsy arrangements, or for more information about the autopsy program, please contact our autopsy coordination team at 1-216-368-0587. Thank you for helping us to further our understanding of this significant health concern.

Sincerely,



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Director  
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