

# The National Prion Disease Pathology Surveillance Center

Supported by the Centers for Disease Control and Prevention (CDC)  
Sponsored by the American Association of Neuropathologists (AANP)

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Dear Clinician,

The National Prion Disease Pathology Surveillance Center (NPDPS) is funded by the Centers for Disease Control and Prevention (CDC) to assist in the surveillance of human prion diseases (e.g., Creutzfeldt-Jakob disease, CJD) in the U.S. The NPDPS provide services to assist you in diagnosing living patients as well as obtaining a definitive diagnosis of prion disease and its cause following death. The following resources are available through the NPDPS:

- Coordinating autopsies for suspected cases of prion disease from around the country for free. NPDPS autopsy coordinators work with local providers who are willing to perform brain only autopsies and will coordinate transportation of the body to the autopsy site and funeral home.
- Neuropathological analyses of brain tissue. The NPDPS will perform a variety of tests on brain tissue (Western blot and immunohistochemistry) that can provide a definitive diagnosis of prion disease and determine its etiology.
- Cerebrospinal fluid testing. The NPDPS will conduct a variety of CSF tests to assist with the diagnosis of prion disease. Two markers of neurodegeneration (14-3-3 and tau) are performed. Additionally, we are able to detect minute amounts of prion proteins in the CSF using a technique called real time quaking induced conversion (RT-QuIC), which approaches 100% specificity. The NPDPS is the only clinical lab in the country that performs RT-QuIC for the diagnosis of prion disease.
- Brain MRI consultation program. Brain MRI images can be sent to the NPDPS to be evaluated for free by a neuroradiologist who is an expert in prion disease (Dr. Alberto Bizzi).
- Genetic testing. Genetic testing can be performed on brain tissue or blood to detect mutations of the prion protein gene (*PRNP*) to assess the possibility of genetic prion disease (i.e., genetic CJD, Gerstmann-Straussler-Scheinker Syndrome, and fatal familial insomnia)
- Education. The NPDPS routinely speaks with clinicians and other providers to offer assistance with questions regarding prion disease

Prion disease surveillance is important. The only way to definitely diagnosis prion disease as well as its etiology is through neuropathological examination. The NPDPS offers autopsy coordination and neuropathological evaluation to monitor for changes in prion disease incidence as well as to detect acquired cases of prion disease (e.g., variant CJD due to bovine spongiform encephalopathy and iatrogenic CJD). Importantly, the NPDPS also monitors for the presence of novel human prion diseases such as assessing the risk of chronic wasting disease (CWD), a prion disease of deer and elk, transmitting to humans. However, we cannot do our work without receiving referrals from clinicians. Thank you for your help and support.

For assistance in coordinating an autopsy of a case of suspected prion disease, please call the NPDPS at 216-368-0587.

Sincerely yours,

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