

Prions and Public Health Office (PPHO)

PPHO's Work

PPHO monitors human prion diseases in the US and coordinates programs related to these and other selected neurological diseases and illnesses of unknown causes such as Nodding syndrome and Kawasaki syndrome. PPHO conducts epidemiological research and provides expert consultation on prion diseases and Kawasaki syndrome. Its statisticians provide consultation on a variety of studies related to high-consequence pathogens.

- Why we care: Prion diseases, which can occur in humans and animals, are relatively rare but always fatal. Surveillance and research are essential to define how prion diseases spread and to identify opportunities for preventing and controlling these diseases.
- Prions are abnormal protein particles that are infectious. When they multiply, they damage different parts of the brain. Prions are believed to be the cause of a class of diseases referred to as prion diseases or transmissible spongiform encephalopathies (TSEs).
- Kawasaki syndrome is an acute febrile illness of unknown etiology that primarily affects children younger than 5 years of age. It is the leading cause of acquired heart disease in children in the US.

Human Prion Diseases

- Creutzfeldt-Jakob disease (CJD, three types: sporadic, familial, iatrogenic)
- Variant Creutzfeldt-Jakob disease (vCJD)
- Gerstmann-Straussler-Scheinker syndrome (GSS)
- Fatal familial insomnia (FFI)
- Variably Protease Sensitive Prionopathy (VPSPr)
- Kuru

Animal Prion Diseases

- Bovine spongiform encephalopathy (BSE)
- Chronic wasting disease (CWD)
- Scrapie
- Transmissible mink encephalopathy
- Feline spongiform encephalopathy
- Ungulate spongiform encephalopathy



Surveillance:

To detect and monitor the occurrence and type of prion diseases in the US, including new forms of emerging human prion diseases, PPHO:

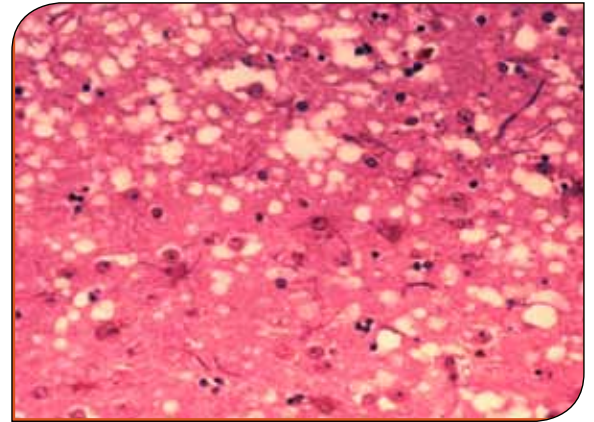
- Supports the National Prion Disease Pathology Surveillance Center (NPDPSC), which serves to confirm clinically diagnosed and suspected cases of prion disease.
- Works with state and local health departments to investigate patients of interest, including prion disease patients younger than 55 years of age and those with a suspected prion exposure.
- Monitors prion diseases among recipients of potentially contaminated pituitary-derived human growth hormone or blood from a donor known to have developed a non-vCJD prion disease; these are known or potential sources respectively of prion transmission.

- Investigates new or emerging prion diseases, such as VCJD, linked to consumption of products from cows with bovine spongiform encephalopathy (BSE, or “mad cow” disease).

Research:

PPHO statisticians analyze data related to prion and other neurological diseases. To conduct these analyses, PPHO:

- Collaborates on research on potential links between vaccines and neurological disorders, including an ongoing research on the potential link between childhood vaccines and Kawasaki syndrome, and between influenza vaccines and a neurological disorder, Guillain-Barré syndrome.
- Provides bioinformatics and statistical consultations, particularly on the use of large administrative datasets on causes of deaths and hospitalizations in the US.



Expert Consultation:

PPHO provides consultations and support to patients’ families and physicians, infection control specialists, and other partners in responding to outbreaks and infectious diseases with neurological manifestations worldwide. To provide this support, PPHO:

- Analyzes large data sets such as national death and hospitalization records to gather information on infectious diseases.
- Educates families and physicians working with patients who have confirmed or suspected cases of prion disease.
- Provides expert guidance on human prion disease outbreaks related to possible exposure sources.
- Informs blood safety policies and healthcare-associated infection control policies.

PPHO in Action: Investigating Nodding Syndrome in Africa

PPHO co-leads an ongoing CDC investigation of nodding syndrome (NS), a severe neurological seizure condition of unknown origin affecting children in Uganda, Tanzania, and South Sudan. This condition impairs physical growth and cognitive development, rendering children almost entirely dependent on caregivers for life.

Since December 2009, CDC’s multi-disciplinary teams have conducted studies in several African countries in an effort to identify the cause of NS, assess disease burden, test treatments, conduct laboratory analysis, and determine the natural history of the illness.

PPHO in Action: National Prion Disease Pathology Surveillance Center Confirms Fourth Case of Variant Creutzfeldt-Jakob Disease (vCJD) in the US

In 2014, PPHO staff members coordinated an investigation in response to reports of a patient with suspected vCJD in Texas. The National Prion Disease Pathology Surveillance Center confirmed the diagnosis of vCJD through analyses of the patient’s brain. The investigation indicated that the patient most likely contracted vCJD overseas in a geographic area not previously recognized as posing a risk for vCJD, underscoring that healthcare providers should not dismiss this diagnosis based solely on a patient’s residential and travel history. Furthermore, the investigation highlighted the importance of continued national surveillance for vCJD and the critical role that post-mortem brain examination plays.

Visit Online at http://www.cdc.gov/nceid/dhcpp/prion_public/index.html