

# PRION AND PUBLIC HEALTH OFFICE

Division of High Consequence Pathogens and Pathology



## Protecting Against Deadly Brain Diseases

CDC's **Prion and Public Health Office (PPHO)** tracks, investigates, and assists with diagnosis of prion diseases — a family of rare, always fatal neurologic diseases. Prion diseases occur when proteins normally found in the brain misfold, causing damage to the brain. These diseases can occur naturally in some people for unknown reasons (sporadic) or run in families (familial). In rare cases they may also be acquired through specific types of exposure to prion proteins. Most people die within a year from the time symptoms begin. Some prion diseases include:

In animals	In people
<ul style="list-style-type: none"><li>▶ <b>Bovine spongiform encephalopathy (BSE), or “mad cow” disease, which affects cattle</b></li><li>▶ <b>Chronic wasting disease (CWD), which affects deer, elk, and moose</b></li></ul>	<ul style="list-style-type: none"><li>▶ <b>Classic Creutzfeldt-Jakob disease (CJD)</b></li><li>▶ <b>Variant Creutzfeldt-Jakob disease (vCJD), the human form of bovine spongiform encephalopathy (BSE)</b></li></ul>

PPHO's scientists also study selected illnesses of unknown or multiple causes, such as

- ▶ Kawasaki disease, a sudden feverish illness that is a leading cause of acquired heart disease in U.S. children, and
- ▶ Guillain-Barré syndrome (GBS), a disorder caused by an unusual immune response that causes the body to attack its own nerves outside of the brain and spinal cord.

PPHO also houses a statistical unit that provides biostatistical advice and support for disease investigations across CDC. This unit uses large publicly available administrative datasets. It also maintains a unique dataset from Indian Health Service that allows CDC staff and other experts to investigate the impact and prevention of diseases affecting Native American populations.

## PPHO by the Numbers

- ▶ **As of January 2021, at least 339 counties in 25 states had reported cases of CWD in wild animals. Although there are no reported CWD infections in people, some lab studies suggest human infection could be a concern.**
- ▶ **More than 500 people in the US die annually from prion diseases, nearly all from sporadic or familial forms. PPHO expects the number to increase as the age of the U.S. population increases.**
- ▶ **About 5,500 specimens were submitted to a PPHO-supported laboratory that tests for prions in people in 2019.**



# Tracking, Researching, and Consulting on Brain Diseases



## Improve Laboratory-Based Prion Disease Monitoring

PPHO supports the National Prion Disease Pathology Surveillance Center (NPDPS), which uses state-of-the-art diagnostic testing to confirm prion disease cases, helping doctors diagnose prion diseases early and improving the tracking of new and known prion diseases. PPHO scientists have helped NPDPS assess a newer, more effective test that identifies prion diseases. Increased use of this test should lead to earlier, more accurate prion disease diagnoses, improved death certificate data, and better monitoring of prion disease in the United States.

## Track Prion Diseases and Assist in Their Diagnosis

PPHO scientists work with state and local health departments to identify and investigate suspected cases of prion disease. They fund enhanced prion monitoring activities in selected states to support studies to determine safety and risk of prion diseases in specific groups, such as those who received blood transfusions from donors who later developed CJD.

For CWD, PPHO collaborates with U.S. animal health experts who track the disease, which has been found in 25 states, and publishes maps of the areas where CWD has been found. PPHO provides recommendations to hunters and others who may come into contact with infected animals. If CWD could spread to people, it would most likely be through eating meat from infected animals or accidentally injecting the infectious agent during harvesting. PPHO experts monitor for the possibility of human cases of CWD by investigating unusual human prion disease cases and evaluating prion disease rates in groups more likely to be exposed to CWD, such as people who hunt in areas where the disease has been found.

## Research Debilitating Diseases

In addition to studying prion diseases, PPHO scientists specialize in researching other serious diseases. They study links between infectious illnesses and Guillain-Barré syndrome (GBS), including GBS following Zika infection and COVID-19. Recently, PPHO staff members and colleagues published strong evidence that treating certain higher-risk Kawasaki disease patients with corticosteroids in addition to the standard therapy reduced coronary artery abnormalities by almost half compared to standard therapy alone.

## Consult During Outbreaks

PPHO provides expert guidance when responding worldwide to outbreaks of infectious diseases with neurological symptoms. PPHO scientists have played an integral part in CDC's COVID-19 response during investigations of the neurological symptoms of COVID-19 and multisystem inflammatory syndrome in children (MIS-C). MIS-C is a rare but serious condition where different body parts can become inflamed. PPHO and other CDC scientists are collaborating with researchers, healthcare providers, and other partners to learn more about MIS-C and how it is related to COVID-19 infection.

