



## Chronic Wasting Disease: Frequently Asked Questions

### What Is Chronic Wasting Disease?

Chronic wasting disease (CWD) is a contagious neurological disease affecting deer, elk, and moose. It causes a characteristic spongy degeneration of the brain in infected animals, resulting in emaciation, abnormal behavior, loss of bodily functions, and death.

CWD belongs to a group of diseases known as transmissible spongiform encephalopathies (TSEs). Within this family of diseases, there are several that affect domestic animals, such as scrapie, which has been identified in domestic sheep and goats for more than 200 years, bovine spongiform encephalopathy (BSE) in cattle (also known as "mad cow disease"), and transmissible mink encephalopathy in farmed mink.

Several rare human diseases are TSEs. Creutzfeldt-Jakob disease (CJD) occurs naturally in about one out of every one million people worldwide. Variant Creutzfeldt-Jakob disease (v-CJD) has been associated with the large-scale outbreak of BSE in cattle herds in Great Britain.

### What Wildlife Species Are Affected by CWD?

Four species of the deer family (cervids) are known to be naturally susceptible to CWD: elk, mule deer, white-tailed deer, and moose. Susceptibility of other cervids and other wildlife species is not completely understood, although ongoing research is further exploring this question.

### Can Humans Get CWD?

Though many observers try to compare CWD with "mad cow disease," the diseases are distinctly different despite both being TSEs. Currently, there is no evidence that CWD poses a risk for humans. Public health officials recommend that human exposure to the CWD infectious agent be avoided as they continue to evaluate any potential health risks, however. The World Health Organization has reviewed available scientific information and concluded that currently there is no evidence that CWD can be transmitted to humans.

### Can Livestock Get CWD?

Cattle and other domestic livestock appear to be resistant to natural infection. There are no reported cases of natural transmission of CWD from infected cervids to domestic livestock. The disease has been experimentally reproduced in cattle by the direct injection of the infectious agent into their brains, however. Several investigations are underway to further study this question.

### What Causes CWD?

The most widely accepted theory is that the agent is an abnormal form of cellular protein (prion) that is most commonly found in the central nervous system and in lymphoid tissue. The abnormal prion "infects" the host animal by promoting conversion of normal cellular protein to the abnormal form.

The CWD infectious agent is quite small and does not evoke any detectable immune response or inflammatory reaction in the host animal. Based on experience with other TSE agents, the CWD infectious agent is assumed to be resistant to enzymes and chemicals that normally break down proteins, as well as resistant to heat and normal disinfecting procedures. Special disinfection procedures are necessary to denature CWD prion proteins.

## **Where and How Did CWD Originate?**

The origin of CWD is unknown, and it may never be possible to definitively determine how or when CWD arose. It was first recognized as a syndrome in captive mule deer held in wildlife research facilities in Colorado in the late 1960s, but it was not identified as a TSE until the 1970s. Computer modeling suggests the disease may have been present in free-ranging populations of mule deer for more than 40 years.

Scrapie, a TSE of domestic sheep, has been recognized in the United States since 1947, and it is possible that CWD was derived from scrapie. It is possible, though never proven, that deer came into contact with scrapie-infected sheep either on shared pastures or in captivity somewhere along the front range of the Rocky Mountains.

It may be possible that CWD is a spontaneous TSE that arose in deer in the wild or in captivity and has infectious qualities promoting transmission to other deer and elk.

## **How Does CWD Spread?**

It is not known exactly how CWD is naturally transmitted. The infectious agent may be transmitted by blood, feces, urine, and saliva. Transmission can occur from the environment in highly contaminated settings as well as from direct animal contact. Environmental contamination likely plays a role in perpetuating epidemic transmission when animal densities are high.

Because CWD infectious agents are extremely resistant in the environment, transmission may be both direct (animal – animal) and indirect (animal – environment – animal), concentrating deer and elk in captivity or by artificial feeding probably increases the likelihood of both direct and indirect transmission between individuals. The apparent persistence of the infectious agents in contaminated environments represents a significant obstacle to eradication of CWD from either captive or free-ranging cervid populations.

The movement of live animals is one of the greatest risk factors in spreading the disease into new areas. Natural movements of wild deer and elk contribute to the spread of the disease, and human-aided transportation of both captive and wild animals greatly amplifies this risk factor.

## **What Are the Clinical Signs of CWD?**

Cases of CWD occur most commonly in adult animals, but may occur in yearlings. The disease is progressive and fatal. The most obvious and consistent clinical sign is progressive weight loss over time. Excessive drinking and urination are common in the late stages. Behavioral changes also occur in the majority of cases, including decreased interactions with other animals, listlessness, lowering of the head, blank facial expression and repetitive walking in set patterns. In elk, behavioral changes may also include hyper-excitability and nervousness. Excessive salivation can also be observed.

Clinical signs of CWD alone are not conclusive. There are other maladies that have symptoms that mimic those of CWD. Currently, the only conclusive diagnosis involves an examination of the brain, tonsils or lymph nodes performed after death.

## **How Is CWD Detected?**

Because clinical signs of CWD alone are not conclusive, a definitive diagnosis is based on examination of the brain for the characteristic microscopic spongiform lesions and/or accumulation of the CWD associated prion protein in brain and lymphoid tissues using a technique called immunohistochemistry (IHC). Gross lesions seen at necropsy reflect the clinical signs of CWD and aspiration pneumonia, which may be the actual cause of death, is also common in diseased animals. A live-animals test is being researched using tonsil tissue. This test may be viable for deer but has so far been ineffective at detecting the disease in elk.

## **Has CWD been found in Valley Forge National Historical Park?**

CWD has not been detected in Valley Forge NHP. In 2012, the first confirmed case of CWD was detected in Pennsylvania from a captive animal in Adams County, approximately 87 miles from Valley Forge. Following the detection of CWD in both captive and free-ranging deer in Pennsylvania, the Game commission establishes Disease Management Areas (DMAs) to minimize the effects of CWD on free ranging deer. As of October 2018, CWD has been detected in the following Pennsylvania DMAs: DMA 1 on a captive deer farm in Adams County (2012) and DMA 1 has since been eliminated; DMA 2 in multiple free-ranging deer in Bedford, Blair, Cambria, and Fulton counties, as well as captive deer farms in Bedford, Franklin, and Fulton counties; DMA 3 in two captive deer farms in Jefferson County and a free-ranging deer in Clearfield County; DMA 4 in a captive deer farm in Lancaster County. More information on the DMAs can be found at <https://www.pgc.pa.gov/Wildlife/Wildlife-RelatedDiseases/Pages/ChronicWastingDisease.aspx>. The closest case of CWD in free-ranging deer to VAFO is 130 miles west of the park in Fulton County Pa and the closest case of CWD in a captive population is 40 miles away in Lancaster County (PA Game Commission 2018).

## **What is Valley Forge National Historical Park Doing with Regard to CWD Detection and Response?**

National Park Service policy directs park units within a 60-mile radius of a known CWD case to conduct opportunistic and targeted surveillance. Although the closest wild case of CWD is 130 miles from the park boundary, the entire state of Pennsylvania is considered to be at high risk for CWD. In 2007, the park completed a CWD risk assessment and implementation plan for opportunistic CWD surveillance<sup>a</sup>. Park staff completed a protocol for the collection and handling of tissue samples, obtained funding and purchased supplies, and obtained necessary surveillance and sample collection training. By Spring, 2018, over 953 animals in the park were tested and all animals tested negative for the disease.

A full CWD Response Plan was developed cooperatively with the Pennsylvania Game Commission (PGC) and is presented in the White-tailed Deer Management Plan/Environmental Impact Statement (available at <http://www.nps.gov/vafo/parkmgmt/index.htm>). Response includes disease surveillance (detection) actions as well as actions to assess disease prevalence and distribution, minimize the likelihood of spread to surrounding communities and amplification within local deer populations, and if possible, promote elimination of CWD. All response actions are closely coordinated with the PGC and Pennsylvania Department of Agriculture due to the scale of the management area identified as necessary to address CWD (minimum 79 mi<sup>2</sup>) relative to park size (5.3 mi<sup>2</sup>).

## **What Are States in the Northeast doing with Regard to CWD Detection and Management?**

Since the September 2005 detections in West Virginia and New York, state agencies in the Northeast have increased CWD surveillance and development of CWD action plans. Many of these plans go beyond targeted and opportunistic surveillance and include lethally removing deer that appear to be healthy to look and/or manage for CWD. In October 2012, CWD was detected for the first time in a captive animal in Pennsylvania, triggering the PA Game Commission to establish disease management zones around the case, increase disease surveillance, and initiate herd reduction in captive animals near the site. To link to state wildlife agency CWD information, including the Pennsylvania Chronic Wasting Disease Response Plan, please visit: <http://www.cwd-info.org/index.php/fuseaction/links.main>.

<sup>a</sup>Opportunistic surveillance involves taking diagnostic samples for CWD testing from cervids found dead or removed through a lethal management action. Cause of death may be culling, disease, trauma (hit by car), or undetermined.