

Chronic Wasting Disease (CWD) in Cervid Populations

INTRODUCTION

Chronic Wasting Disease. Chronic wasting disease (CWD) is a [prion disease](#) that affects deer, elk, reindeer, sika deer, and moose. It has been found in Canada, the United States, Norway, and South Korea. Symptoms, which may take over a year to develop, include drastic weight loss (wasting), stumbling, listlessness, and other neurologic symptoms. CWD affects animals of all ages; some infected animals may die without ever developing the disease. CWD is fatal to animals and there are no treatments or vaccines. (*Center for Disease Control and Prevention 2019*)

The potential spread of CWD to new areas is perhaps the greatest concern for managers of deer populations. Because of CWD's low prevalence and prolonged latent period makes detections difficult. The prion agent is viable in the environment for years. Infection may occur through contact with a contaminated environment or an infected animal. Infected animals can shed the prion before the onset of clinical signs, sometimes for months (*Mathiason et al. 2009*).

The tests for the presence of the prion in tissues of harvested animals are specific and sensitive (*Spraker et al. 2002, Hibler et al. 2003*). However, testing of live animals remains problematic due to the difficulty in obtaining diagnostic samples and the potential for false negative results early in the disease course. Several researchers have investigated the accuracy of two live animal tests; tonsil biopsy and rectal biopsy (*Wolfe et al. 2007, Geremia et al. 2015, Keane et al. 2009, Thomsen et al. 2012*). The sensitivity of these tests (portion of positives detected) has ranged from 63% to 80% in deer herds with a relatively high level of CWD prevalence (up to 67%). The detection of prions in tonsillar and rectal samples from infected animals is influenced by the quality of the sample, species, PRNP codon allele of the deer, and number of days post-exposure (*Thomsen et al. 2012*). Some infected animals will not test positive for more than 23 months post-exposure, but may excrete prions 7 to 11 months before developing clinical signs (*Mathiason et al. 2009, Tamgüney et al. 2009*). It is possible that an infected test-negative animal could be contagious for several months before succumbing to the disease and could represent a risk for disease introduction if it were to be translocated to a CWD negative area. Many states and provinces have tested for the presence of CWD in deer populations using samples from hunter-harvested deer, moose and elk at a level such that the disease would be detected if it was present in 1% or more of the animals and have identified CWD endemic and presumptive CWD free regions. The risk of introducing CWD with the translocation of a positive animal is significant for positive source populations, with a potentially significant impact to the herd into which it is moved. Therefore, accumulating a multi-year testing history on potential source populations is important and deer from CWD endemic areas should not be moved to areas where it has not been detected.

(*Source: Western Association of Fish & Wildlife Agencies Mule Deer Working Group*)

BACKGROUND

What is a prion?

Prion diseases or transmissible spongiform encephalopathies (TSEs) are a family of rare progressive neurodegenerative disorders that affect both humans and animals. They are distinguished by long incubation periods, characteristic spongiform changes associated with neuronal loss, and a failure to induce inflammatory response.

The causative agents of TSEs are believed to be prions. The term “prions” refers to abnormal, pathogenic agents that are transmissible and are able to induce abnormal folding of specific normal cellular proteins called prion proteins that are found most abundantly in the brain. The functions of these normal prion proteins are still not completely understood. The abnormal folding of the prion proteins leads to brain damage and the characteristic signs and symptoms of the disease. Prion diseases rapidly progress and are fatal.

To date, there have been no reported cases of CWD infection in humans. However, studies suggest CWD poses a risk to non-human primates, such as monkeys, that eat meat from [CWD-infected animals](#) or come in contact with brain or bodily fluids from infected deer or elk. These studies raise concerns that there may also be a risk to humans. Since 1997, the World Health Organization has recommended to keep the agents of all known prion diseases from entering the human food chain.

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 models suggest that 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 known to occur in the United States since 1947, and it is possible that CWD was derived from scrapie. Though never proven, deer may have come into contact with scrapie-infected sheep either on shared pastures or in captivity somewhere along the Front Range of the Rocky Mountains, where high levels of sheep grazing occurred in the early 1900s.

It may also be possible that CWD is a spontaneous TSE that arose in deer in the wild or in captivity.

Transmission

Scientists believe CWD proteins (prions) likely spread between animals through body fluids like feces, saliva, blood, or urine, either through direct contact or indirectly through environmental contamination of soil, food or water. Once introduced into an area or farm, the CWD protein is contagious within deer and elk populations and can spread quickly. Experts believe CWD prions can remain in the environment for a long time, so other animals can contract CWD from the environment even after an infected deer or elk has died.

The CWD prion has been shown to experimentally infect squirrel monkeys, and also laboratory mice that carry some human genes. In addition, a study begun in 2009 by Canadian and German scientists is evaluating whether CWD can be transmitted to macaques, a type of monkey that is genetically closer to people than any other animal that has been infected with CWD previously. On July 10, 2017, the scientists presented a summary of the study's progress (access the [recorded presentation External](#) and [slides Cdc-pdf\[PDF 3.88MB\]External](#)), in which they showed that CWD was transmitted to monkeys that were fed infected meat (muscle tissue) or brain tissue from CWD-infected deer and elk. Some of the meat came from asymptomatic deer that had CWD (i.e., deer that appeared healthy and had not begun to show signs of the illness yet). Meat from these asymptomatic deer was also able to infect the monkeys with CWD. CWD was also able to spread to macaques that had the infectious material placed directly into their brains.

This study showed different results than a previous study, which had not shown successful transmission of CWD to macaques. The reasons for the different experimental results are unknown. To date, there is no strong evidence for the occurrence of CWD in people, and it is not known if people can get infected with CWD prions. Nevertheless, these experimental studies raise the concern that CWD may pose a risk to people and suggest that it is important to prevent human exposures to CWD.

Additional studies are under way to identify if any prion diseases could be occurring at a higher rate in people who are at increased risk for contact with potentially CWD-infected deer or elk meat. Because of the long time it takes before any symptoms of disease appear, scientists expect the study to take many years before they will determine what the risk, if any, of CWD is to people.

Detection

The tests for CWD are specific and sensitive (*Spraker et al. 2002, Hibler et al. 2003*). However, testing of live animals remains problematic due to the difficulty in obtaining diagnostic samples and the potential for false negative results early in the disease course. Several researchers have investigated the accuracy of two live animal tests; tonsil biopsy and rectal biopsy (*Wolfe et al. 2007, Geremia et al. 2015, Keane et al. 2009, Thomsen et al. 2012*). The sensitivity of these tests (portion of positives detected) has ranged from 63% to 80% in deer herds with a relatively high level of CWD prevalence (up to 67%). The detection of prions in tonsillar and rectal samples from infected animals is influenced by the quality of the sample, species, PRNP codon allele of the deer, and number of days post-exposure (*Thomsen et al. 2012*). Some infected animals will not

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(Source: Western Association of Fish & Wildlife Agencies Mule Deer Working Group)

Prevention

If CWD could spread to humans, it would most likely be through eating of infected deer and elk. In a 2006-2007 [CDC surveyExternal](#) of U.S. residents, nearly 20 percent of those surveyed said they had hunted deer or elk and more than two-thirds said they had eaten venison or elk meat. However, to date, there is no strong evidence for the occurrence of CWD in people, and it is not known if people can get infected with CWD prions.

Hunters must consider many factors when determining whether to eat meat from deer and elk harvested from areas with CWD, including the level of risk they are willing to accept. Hunters harvesting wild deer and elk from areas with reported CWD should check state wildlife and public health guidance to see whether testing of animals is recommended or required in a given state or region. In areas where CWD is known to be present, CDC recommends that hunters strongly consider having those animals tested before eating the meat.

Tests for CWD are monitoring tools that some state wildlife officials use to look at the rates of CWD in certain animal populations. Testing may not be available in every state, and states may use these tests in different ways. A negative test result does not guarantee that an individual animal is not infected with CWD, but it does make it considerably less likely and may reduce your risk of exposure to CWD.

To be as safe as possible and decrease their potential risk of exposure to CWD, hunters should take the following steps when hunting in areas with CWD:

- Do not shoot, handle or eat meat from deer and elk that look sick or are acting strangely or are found dead (road-kill).
- When field-dressing a deer:
 - Wear latex or rubber gloves when dressing the animal or handling the meat.

- Minimize how much you handle the organs of the animal, particularly the brain or spinal cord tissues.
- Do not use household knives or other kitchen utensils for field dressing.
- Check state wildlife and public health guidance to see whether testing of animals is recommended or required. Recommendations vary by state, but information about testing is available from many state wildlife agencies.
- Strongly consider having the deer or elk tested for CWD before you eat the meat.
- If you have your deer or elk commercially processed, consider asking that your animal be processed individually to avoid mixing meat from multiple animals.
- If your animal tests positive for CWD, do not eat meat from that animal.

The U.S. Department of Agriculture’s Animal and Plant Health Inspection Service regulates commercially farmed deer and elk. The agency operates a [national CWD herd certification programExternal](#). As part of the voluntary program, states and individual herd owners agree to meet requirements meant to decrease the risk of CWD in their herds. Privately owned herds that do not participate in the herd certification program may be at increased risk for CWD.

Source: [Centers for Disease Control and Prevention](#) , [National Center for Emerging and Zoonotic Infectious Diseases \(NCEZID\)](#) , [Division of High-Consequence Pathogens and Pathology \(DHCPP\)](#)

Importation of Animal Parts:

It is unlawful for persons to bring deer, elk, moose or caribou carcasses, hides or antlers into -----
 - from any state, territory or province unless all meat has been deboned and skull plates and hides have been completely cleaned of all brain and spinal cord tissue.

CWD Positive States and Provinces

These states and provinces have been identified to be chronic wasting disease (CWD) positive by the Chronic Wasting Disease Alliance:

United States

- Arkansas
- Colorado
- Illinois
- Iowa
- Idaho
- Kansas
- Maryland
- Michigan
- Minnesota
- Mississippi
- Missouri
- Montana
- Nebraska
- New Mexico
- New York
- North Dakota
- Ohio
- Oklahoma
- Pennsylvania
- South Dakota
- Tennessee
- Texas
- Utah
- Virginia
- West Virginia
- Wisconsin
- Wyoming

Canada

- Alberta
- Saskatchewan
- Quebec

CWD has also been found in the Republic of Korea, Norway, and Finland.

[Centers for Disease Control and Prevention](#) , [National Center for Emerging and Zoonotic Infectious Diseases \(NCEZID\)](#) , [Division of High-Consequence Pathogens and Pathology \(DHCPP\)](#), February 2019

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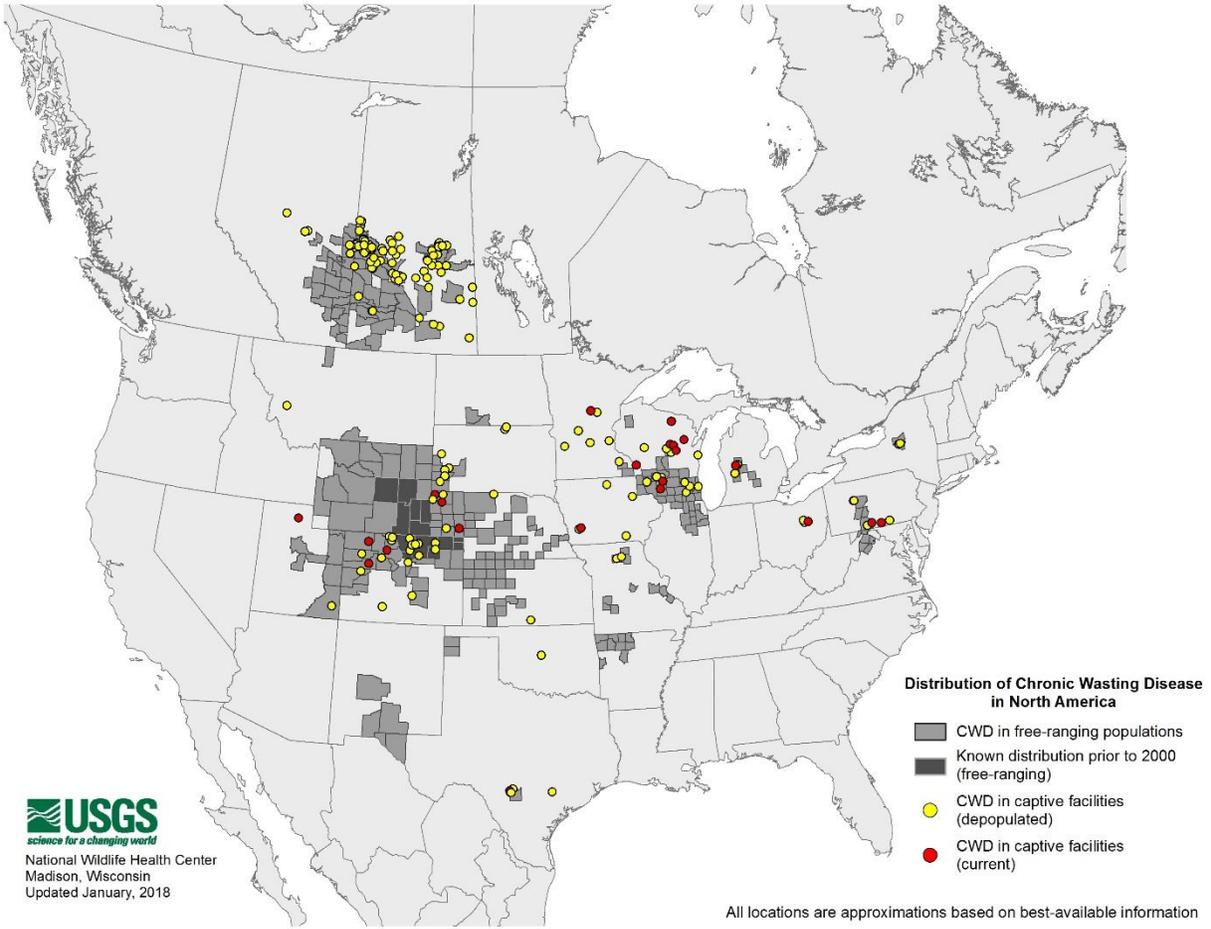
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