

Questions and Answers About Chronic Wasting Disease

Q. What is chronic wasting disease?

A. Chronic wasting disease (CWD) is a fatal, neurological disease of farmed and wild deer and elk. The disease has been identified in wild and captive mule deer, white-tailed deer and North American elk, and in captive black-tailed deer. CWD belongs to the family of diseases known as transmissible spongiform encephalopathies (TSEs). TSEs include a number of different diseases affecting animals or humans including bovine spongiform encephalopathy (BSE) in cattle, scrapie in sheep and goats, and Creutzfeldt-Jacob disease (CJD) in humans. Although CWD shares certain features with other TSEs, it is a distinct disease affecting only deer and elk. CWD is a progressive, fatal, degenerative disease. Clinical signs in affected animals include loss of body condition, behavioral changes, excessive salivation, increased drinking and urination, depression, and eventual death. CWD is always fatal. There is no known treatment, vaccine, or live animal test for CWD.

Q. What causes this disease?

A. The agent that causes CWD and other TSEs has not been completely characterized. However, the theory supported by most scientists is that TSE diseases are caused by little understood proteins called prions. Prions are a form of protein normally found in the cells of the nervous system and other body tissues. Stanley Prusiner, a Nobel Prize winning neurologist, first described an abnormal form of prion resistant to enzymes that break down normal proteins. These abnormal, protease resistant prions are referred to as PrPres. PrPres have the ability to transform normal prions into this abnormal state. As the disease progresses, PrPres accumulate in the brain and lymphoid tissues (lymph nodes and tonsils). Accumulation of these abnormal PrPres produce tiny sponge-like holes in the brain that are visible microscopically. The word "spongiform" in TSEs describes the sponge-like condition of brain tissue found in infected animals. As the disease progresses, the affected animal loses its basic physical and mental abilities.

Q. What is the history of CWD?

- A.** The following is a brief chronology of CWD:
- CWD was first described clinically as a wasting syndrome in captive deer belonging to Colorado research facilities in 1967. A few years later it was described in a Wyoming research facility.
 - CWD was first determined to be a TSE in 1978 by Dr. Elizabeth Williams of the University of Wyoming.
 - The first cases of CWD in wild deer and elk were diagnosed in 1981 in Colorado and 1985 in Wyoming.
 - Beginning in the 1980s, the distribution of CWD in wild deer and elk in Colorado and Wyoming was determined through surveillance by wildlife agencies in those States. Through their efforts, an endemic area for the disease in wildlife in their States was described. This area includes much of northeastern Colorado and southeastern Wyoming.
 - In 2001, discovery of a positive wild mule deer in neighboring Kimball County, NE, extended the endemic area into southwestern Nebraska.
 - From 1996 to June 2002, CWD was diagnosed in farmed elk herds in Colorado, Kansas, Montana, Nebraska, Oklahoma, South Dakota, and the Canadian Provinces of Alberta and Saskatchewan.
 - From 2000 to June 2002, CWD has also been found in wild deer in northwestern Nebraska, southern New Mexico, southwestern South Dakota, south-central Wisconsin, northwestern Colorado, and the Canadian Province of Saskatchewan.

Q. What are the symptoms of CWD?

A. CWD is a slow and progressive disease. Because the disease has a long incubation period, elk and deer with CWD may not produce any visible symptoms of the disease for a number of years after they become infected. As the disease progresses, deer and elk with CWD show changes in behavior and appearance. These clinical signs may include progressive weight loss, stumbling, tremors, lack of coordination, blank facial expressions, excessive salivation, loss of appetite, excessive thirst and urination, listlessness, teeth grinding, abnormal head posture, and drooping ears. Because of effects on the central nervous system, animals can have difficulty in swallowing, resulting in pneumonia caused by aspiration of food or saliva. Clinical signs of CWD are usually present a few weeks to several months before the animal dies. Unfortunately, these

signs are not specific to CWD and can occur with other diseases or malnutrition.

Q. How is CWD transmitted?

A. The exact mechanism of transmission is unclear. Evidence suggests CWD is transmitted directly from one animal to another (lateral or horizontal transmission). The route by which the agent is shed from the animal's body is unknown. However, experimental and circumstantial evidence suggests that indirect transmission from an environment contaminated with the agent appears to be possible. Transmission of CWD has not been associated with any particular feeding practice or regimen in farmed elk or deer. Supplemental feeding of wild elk and deer, however, concentrates the animals and may contribute to disease spread.

Q. Is there any way to destroy the infectious agent?

A. A characteristic of all TSE agents is their resistance to conventional disinfectants, high temperatures, and enzymes that normally break down proteins. Recommendations for disinfection of areas in which infected animals have resided are still being developed.

Q. Is the disease transmissible to humans?

A. The Centers for Disease Control and Prevention has issued this statement: "It is generally prudent to avoid consuming food derived from any animal with evidence of a TSE. To date, there is no evidence that CWD has been transmitted or can be transmitted to humans under natural conditions. However, there is not yet strong evidence that such transmissions could not occur. To further assess the possibility that the CWD agent might occasionally cause disease in humans, additional epidemiologic and laboratory studies could be helpful. Such studies include molecular characterization and strain typing of the agents causing CWD in deer and elk and CJD in potentially exposed patients. Ongoing national surveillance for CJD and other neurological cases will remain important for continuing to assess the risk, if any, of CWD transmission to humans."

Q. Is the disease transmissible to domestic livestock?

A. During the approximately two decades of monitoring, researchers have not found any evidence that CWD can be transmitted to domestic cattle under natural conditions. Ongoing experiments involving oral exposure and contact exposure on heavily CWD contaminated sites have not resulted in infection of cattle. These experiments, however, require additional time before they are completed. CWD has been experimentally transmitted by artificial means to mice, ferrets, mink, goats, squirrel monkeys, and calves.

Q. How is CWD diagnosed?

A. Currently, CWD is diagnosed by examining brain and lymphoid tissue (lymph nodes and tonsils) from a dead animal. Tests to confirm CWD are performed in a laboratory, using brain tissue. Immunohistochemical (IHC) staining is the most commonly accepted method of detection and is the standard test used by USDA's National Veterinary Services Laboratories. IHC staining is an antibody-based test. Antibodies bind to abnormal PrPres in the tissue on a slide. Additional steps in the test allow a colored agent to be bound to the abnormal PrPres-antibody complex. Accumulations of color indicate the presence of the abnormal PrPres when the slide is examined microscopically. A CWD-positive animal is one in which the presence of abnormal PrPres has been confirmed in the brain or lymphoid tissues.

A research team in Colorado has recently developed a live animal test for CWD based on the collection of tonsil biopsies for microscopic examination. This test seems to work well in mule deer, but not in elk, and its application may be limited to special circumstances. Scientists are continuing to work on a number of approaches that may provide a rapid postmortem or live animal test for both deer and elk.

Q. Why is it so important that the sample collected for testing include the obex portion of the brainstem?

A. Studies on the distribution of abnormal PrPres in CWD-affected deer and elk have shown that the obex portion of the brainstem is the first place that the abnormal PrPres can be detected in the brain. As the disease progresses, the abnormal PrPres can be detected in multiple locations and, finally, throughout the brain. Because of this, it is necessary to test the obex to detect CWD in animals that are in the early stages of the disease. It is possible that other parts of the brain may test negative for the presence of disease while the obex would test positive. For white-tailed deer and mule deer (but not elk) some lymphoid tissues from the head (tonsils and retropharyngeal lymph nodes) become positive before the obex does, so these tissues will also be useful in surveillance and monitoring efforts in deer.

Q. What does a negative IHC test mean?

A. A negative test is one in which there is no detectable IHC staining of abnormal PrPres. The interpretation of a negative test depends on the species and the tissue tested. In elk, if the obex is negative, the animal is most likely not infected with the CWD agent. There is the possibility, however, that the animal is infected but the disease process is so early that the abnormal PrPres is not detectable with the current IHC test. Similarly, in white-tailed deer and mule deer if the obex and/or the lymphoid tissue

from the head are IHC–negative, the animal is most likely not infected with CWD. There is the possibility, however, that the disease process is so early that the abnormal PrPres is not detectable by the current IHC test.

Q. What is the USDA doing about CWD?

A. The USDA has taken the following steps to help control the spread of CWD:

- During 1997, USDA began supporting surveillance for CWD in farmed and wild elk and deer in cooperation with State agriculture and wildlife agencies. Farmed elk herds that tested positive for the disease were put under State quarantine.
- In September 2001, \$2.6 million in Commodity Credit Corporation (CCC) emergency funds were transferred to USDA to increase the effort to eliminate CWD in farmed elk. These funds paid for enhanced surveillance as well as depopulation of farmed CWD-positive, exposed, and suspect animals with compensation to the owners. In addition, the funds allowed USDA to provide assistance in cleaning and disinfecting premises where positive and exposed animals resided. Factsheets were developed to disseminate CWD information.
- In February 2002, an additional \$12.2 million of CCC funding was transferred to APHIS to continue this effort. These funds will also be used to support surveillance and diagnostics in wild elk and deer.
- In April 2002, USDA agreed to purchase farmed elk herds in the endemic area of Colorado where wild animals have tested positive for the disease. Sixteen ranches with about 1,350 animals accepted the offer to purchase their animals for 95 percent of their appraised value.
- USDA continues to provide diagnostic and surveillance support to States with active CWD surveillance and control programs for farmed elk and deer. USDA has proposed a Federal/State/industry certification program for farmed elk and deer.
- USDA has supported efforts to control CWD in wild populations through assistance with diagnostic testing and research.
- In May 2002, USDA and the U.S. Department of Interior formed a joint working group on CWD. The purpose of the group is to ensure a coordinated and cooperative Federal approach to assisting States, Tribes and Federal land management agencies with CWD response efforts. In June 2002, the working group presented a plan to the Congress.

Q. What can elk and deer farmers do?

A. Elk and deer farmers are strongly urged to enroll in State CWD surveillance and control programs. They should only purchase animals that have been enrolled in a State program or are otherwise known not to have been exposed to CWD.

Q. What precautions should hunters of deer and elk follow?

A. Hunters should be vigilant for identifying elk or deer displaying CWD symptoms. They should report suspected cases to authorities immediately. Several States have issued specific guidelines for hunters.

Q. What is the USDA's official position on CWD sample testing?

A. Official diagnosis of CWD should be performed exclusively by Federal and State regulatory agency laboratories. The international credibility of the U.S. animal health system is in large part predicated on having an established set of government laboratories with the expertise to accurately conduct diagnosis not only for CWD, but also BSE, avian influenza, foot-and-mouth disease, and a host of other diseases of concern. The system is designed not only to ensure consistency and accuracy but also to preserve domestic and international market confidence in U.S. agricultural commodities. Indeed, a "false positive" for any disease could result in unnecessary public concern and costly regulatory action. And in the case of a disease like BSE, a false positive could be devastating, costing the U.S. economy billions of dollars in unnecessary domestic and international market disruption from which it could take years to recover.

Q. Can this test be used to determine if an animal is safe for human consumption?

A. Because of the limitations of currently available tests for CWD, testing serves purely as a surveillance tool to determine the geographic parameters and prevalence of the disease in the United States. A positive test result can be used as reliable information that the disease has spread into a given area. However, a negative test result is not necessarily a reliable indicator that an animal is free of the disease. Indeed, at this time no test that can be used reliably on individual animals to determine whether that animal is free from CWD and whether the meat is safe to eat. This is because the disease has a very long incubation period, which leads to a high "false negative" rate during early infection. In addition, relatively little is known about the distribution of the CWD agent, so an animal whose brain and nervous system tissue tests negative might actually be carrying the infective agent in its muscle or other tissues.

Q. What test is used as the official test for CWD surveillance?

A. In order to ensure the integrity of the U.S. surveillance effort, USDA has designated an official test for CWD surveillance: the IHC assay as performed by APHIS's National Veterinary Services Laboratories (NVSL) and State/university laboratories with which NVSL has contracted. Employees at these laboratories, as part of a national network, are being trained, and the laboratories are being proficiency tested, and supplied with control samples to perform official tests. They will be linked through a reporting database. Currently, 10 laboratories with which APHIS has contracted perform CWD testing, and APHIS is working to bring another 5 on line by January 2003. This capacity is more than sufficient to handle the increased surveillance testing planned this fall to determine the geographic distribution and prevalence of CWD in the United States.

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