

Chronic Wasting Disease

Discovery

"Chronic wasting disease" was first recognized by biologists in the 1960s as a disease syndrome of captive deer held in wildlife research facilities in Ft. Collins, CO, but was not recognized as a transmissible spongiform encephalopathy until the late 1970s. This disease was subsequently recognized in captive deer, and later in captive elk, from wildlife research facilities near Ft. Collins, Kremmling, and Meeker, CO and Wheatland, WY, as well as at in least two zoological collections.

More recently, CWD has been diagnosed in privately-owned elk residing in game ranches in a few western states and provinces. Although CWD was first diagnosed in captive research cervids, the original source (or sources) of CWD in either captive cervids or free-ranging cervids is unknown; whether CWD in research animals really preceded CWD in the wild, or *vice versa*, is equally uncertain.

Occurrence

Chronic wasting disease is relatively rare, and its geographic distribution is quite limited. Fewer than 300 naturally occurring clinical cases, mostly in captive research and free-ranging mule deer, have been documented. Based on data from a combination of surveillance methods, CWD now appears in 11 contiguous counties in northeastern Colorado (5 counties), southeastern Wyoming, and the Nebraska Panhandle (1 county). Although the disease doesn't appear to be common, the number of cases detected has increased in recent years. This trend may be explained by increased vigilance by wildlife and animal health officials, the wildlife farming industry, and the public in reporting cases, but it may also reflect increased disease occurrence.

Based on random, preclinical testing of brain or tonsil tissues from animals harvested in specific management units, it appears that on average CWD probably infects about 5-15 percent of the deer in a small core endemic area of northcentral Colorado and southeastern Wyoming, and 1 percent or fewer of the deer in other surrounding mountain and plains areas. Testing of harvested animals indicates less than 1 percent of the elk in endemic areas are probably infected. To date, with the exception of the recent mule deer in Nebraska, no evidence of CWD has been detected in examinations of over 8,000 deer and elk from outside endemic areas in Colorado and Wyoming, or from other states and Canadian provinces where surveys have been conducted in recent years; additional surveys are ongoing.

In addition to cases in captive research and free-ranging deer and elk, CWD has been diagnosed in privately-owned elk on a small number of game farms in Colorado, Montana, Nebraska, Oklahoma, South Dakota, and Saskatchewan, Canada since 1996. Infection has been particularly severe in a group of interconnected facilities near Rapid City, South Dakota, that appear to be the original source of infection for other South Dakota game farms as well as the Saskatchewan epidemic. In contrast, infected elk in two of three Nebraska farms originated in Colorado, and infected elk in Oklahoma apparently originated in Montana; CWD has been confirmed in the Montana and Colorado source herds. Epidemiology of the Canadian cases has been under study, and South Dakota appears to be the likely source of CWD in Saskatchewan; it also appears that CWD was imported into Canada prior to 1990, and has spread among at least 18 farms via live animal sales over the last decade. The overall distribution and occurrence of CWD among farmed elk operations should become clearer as industry-wide surveillance programs are developed. There are no apparent epidemiological connections between the Colorado-Nebraska, South Dakota-Saskatchewan, and Montana-Oklahoma foci; moreover, there are no apparent epidemiological connections between any of the cases in farmed elk and cases in free-ranging or captive research deer and elk.

Transmission

Neither the agent causing chronic wasting disease nor its mode of transmission have been definitively identified, but clinical disease is associated with the accumulation of protease-resistant prion protein (PrP^{res}) in brain tissue (as in other transmissible spongiform encephalopathies). Experimental and circumstantial evidence suggests infected deer and elk probably transmit the disease laterally through animal-to-animal contact and/or contamination of feed or water sources with saliva, urine, and/or feces. Chronic wasting disease seems more likely to occur in areas where deer or elk are crowded or where they congregate at man-made feed and water stations. Although CWD does not appear to be transmitted via contaminated feed, artificial feeding of deer and elk may compound the problem. This may in part explain the intensity of infection in some cervid populations housed in farm or research settings.

According to public health (Centers for Disease Control, World Health Organization) and animal health officials, data available to date indicate that chronic wasting disease is not currently known to be naturally transmitted to humans, or to animals other than deer and elk; data from recent molecular studies provide quantitative evidence of the apparent inefficiency of cross-species transmission. As a general precaution, however, public health officials recommend that

people avoid contact with deer, elk, or any other wild animal that appears sick. Although there's no evidence that chronic wasting disease can be naturally transmitted to domestic livestock, chronic wasting disease is similar in some respects to two livestock diseases: scrapie, which affects domestic sheep and goats worldwide and has been recognized for over 200 years, and bovine spongiform encephalopathy (BSE), which is a more recent disease of cattle in the United Kingdom and Europe. Despite some similarities, there is no evidence suggesting either scrapie or BSE are caused by contact with wild deer or elk, or that wild deer or elk can contract either scrapie or BSE in countries where these diseases occur.

Clinical Signs

Deer and elk affected with chronic wasting disease show progressive loss of body condition accompanied by behavioral changes. In the later stages of disease, emaciation, excessive salivation, increased drinking and urination, stumbling, trembling, and depression may precede death. As with other TSEs, the clinical course of chronic wasting disease appears to be progressive and irreversible, ultimately leading to the death of affected animals. Because the clinical signs of chronic wasting disease are relatively nonspecific, laboratory examination of clinical suspects is essential for confirming this diagnosis.

Diagnosis

At present, the diagnosis of chronic wasting disease is based on microscopic examination of brain (specifically, the medulla oblongata at the obex) and tonsil tissues from suspected cases. Both histopathologic examination and immunohistochemistry (IHC) are used in routine diagnosis of clinical cases, and may also be used to detect preclinical cases in surveillance and monitoring programs; of these, IHC appears to offer greater sensitivity in detecting early preclinical cases. Western blots and negative-stain electron microscopy have also been used to further confirm diagnoses, and other diagnostic tests are being evaluated. There are currently no validated live-animal tests for diagnosing either clinical or preclinical chronic wasting disease in either deer or elk; however, research is underway to evaluate several promising avenues for ante mortem diagnosis.

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