CHRONIC WASTING DISEASE IN WYOMING CERVIDAE

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Abstract: Chronic wasting disease (CWD) is a transmissible spongiform encephalopathy of elk (Cervus elaphus), mule deer (Odocoileus hemionus), and white-tailed deer (Odocoileus virginianus) in North America. The putative agent of CWD is an abnormal isoform (prion) of a host-encoded, protease sensitive protein. The minimum incubation period is 12 months in elk and 15 months in mule deer. Death can occur from a few days to several months after clinical signs appear. It is unknown how CWD is transmitted among animals. As the disease progresses, affected animals usually become reluctant to move, exhibit lowered head and ears, and gradually or quickly lose weight and coat condition. Increased drinking, salivation, and urination are usually seen in captive animals; free-ranging animals are frequently found in close proximity to water and are reluctant to leave such areas. As of May 2003, CWD has been found in free-ranging deer or elk in Saskatchewan, Wyoming, Colorado, Utah, New Mexico, South Dakota, Nebraska, Wisconsin, and Illinois and in farmed deer or elk in Alberta, Saskatchewan, Montana, Colorado, South Dakota, Nebraska, Kansas, Oklahoma, Minnesota, and Wisconsin. In Wyoming, most CWD cases have been found in deer (n = 246) as opposed to elk (n = 8) and prevalence rates are higher in white-tailed deer (17.6%) than in mule deer (9.5%). Thus far, CWD has been confined to the southeastern quarter of Wyoming. The management of CWD in free-ranging species is problematic. An overarching philosophy would be to stop the spread of CWD while reducing the prevalence within an endemic area. Although concerns exist, there is currently no evidence that CWD is transmissible to humans or livestock.

Key words: Cervus elaphus, chronic wasting disease, elk, mule deer, Odocoileus hemionus, Odocoileus virginianus, white-tailed deer, Wyoming

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(proteinaceous infectious particle) was proposed by Prusiner (1982) to distinguish the infectious pathogen from viruses or viroids.

There is no difference in the amino acid sequence between PrPC and PrPres (Stahl et al. 1993), yet the largely α-helical PrPC apparently can refold into the largely β-sheet PrPres (Pan et al. 1993). Although are several models are proposed to explain the conversion of PrPC to PrPres, the most coherent and general model suggests PrPres self associates to produce a stable supramolecular structure, or "seed." Once this seed is formed, additional PrPC can be recruited, leading to an explosive, autocatalytic formation of PrPres (Prusiner et al. 1990). The exact biochemical function of PrPC is unknown, but it appears to have a role in normal brain copper metabolism or transport (Brown 2001). Disturbance of this function by the conformational transition of PrPC to PrPres could be involved in prion-related neurotoxicity (Brown and Sassoon 2002).

Chronic wasting disease has been diagnosed only in elk, mule deer, and white-tailed deer. The pathology of the disease is approximately similar in all species. The minimum incubation period is 12 months in elk and 15 months in mule deer. The maximum course of CWD is unknown, but can exceed 25 months in mule deer and 34 months in elk (Williams et al. 2002). Death can occur from a few days to several months after clinical signs appear.

One of the earliest signs of CWD in captive cervids is hyperexcitability when the animal is restrained or confined. Such animals demonstrate trembling limbs, distended eyes, and persistent head bobbing. As the disease progresses, affected animals usually become reluctant to move, typically appearing with head and ears lowered. Free-ranging animals can often be approached quite closely (30 m) before "waking up" and running away in a normal manner. Affected animals can gradually or quickly lose weight and coat condition. The hair between the ears of elk is often erect (Fig. 1). Increased drinking, salivation, and urination are usually seen in captive animals; free-ranging animals are frequently found in close proximity to water and are reluctant to leave such areas. Although death from CWD is thought to be inevitable, animals may die from other causes, such as pneumonia, prior to succumbing to the actual effects of CWD neurotoxicity.

It is unknown how CWD is transmitted among animals. Transmission may be direct through contact with prion-infected saliva, feces, or urine. There is empirical evidence that prions can remain in the environment for some period of time. Soil has been treated in an attempt to rid it of prions and then allowed to lie fallow for years, only to have susceptible animals contract CWD or scrapie when reintroduced to the area (Brown and Gajdusek 1991, Williams and Young 1992).

Chronic wasting disease is usually diagnosed postmortem, although tonsillar biopsies of anesthetized mule deer have proven reliable indicators of disease (Wild et al. 2002). Diagnosis can be accomplished through histological examination for spongiform lesions in the parasympathetic vagal nucleus in the dorsal portion of the medulla oblongata at the obex (Williams and Young 1993), immunohistochemical staining for PrPres in the obex, tonsils, or retropharyngeal lymph nodes (Peters et al. 2000, Miller and Williams 2002), or enzyme-linked immunosorbent assay (ELISA) of lymphoid tissue (Deslys et al. 2001). Chronic wasting disease was first recognized as a syndrome in a wildlife research facility in Colorado in 1967. Subsequently, CWD was determined to be a spongiform
encephalopathy (Williams and Young 1980). In the decade following, CWD was found in both captive and free-ranging mule deer and elk in Colorado and Wyoming. In 1996, CWD was first diagnosed in farmed elk in Saskatchewan, although epidemiologic investigations traced the source of infection to a South Dakota game farm (Williams et al. 2002). As of May 2003, CWD has been found in free-ranging deer or elk in Saskatchewan, Wyoming, Colorado, Utah, New Mexico, South Dakota, Nebraska, Wisconsin, and Illinois. Chronic wasting disease has been found in farmed deer or elk in Alberta, Saskatchewan, Montana, Colorado, South Dakota, Nebraska, Kansas, Oklahoma, Minnesota, and Wisconsin (Fig. 2).

In Wyoming, the distribution of CWD is determined by using 2 surveillance methods. **Targeted surveillance** is used when a deer or elk showing clinical signs of CWD is harvested and examined or where an animal is found dead under suspicious circumstances (emaciated, found near water). Targeted surveillance is useful in locating new areas of CWD. **Hunter surveillance** is a systematic method of examining deer or elk killed by hunters. Hunter surveillance has been conducted in Wyoming since 1997 and is mostly accomplished by collecting samples at meat processors, taxidermists, or check stations. Prior to 2003, collected obexes were examined via immunohistochemistry and

Fig. 1. Captive elk in terminal stages of chronic wasting disease. Note gaunt condition, droopy ears, erect hair between ears, and salivation. This animal died abruptly 30 minutes after photo was taken.
histology. In 2003, only retropharyngeal lymph nodes will be collected and screened using ELISA. Suspect nodes will be confirmed by immunohistochemistry. Most CWD cases have been found in deer \((n = 246)\) as opposed to elk \((n = 8)\). Thus far, CWD has been confined to the southeastern quarter of Wyoming (Fig. 3).

The management of CWD in free-ranging species is problematic at best. An overarching philosophy would be to stop the spread of CWD while reducing the prevalence within an endemic area. To do this, states and provinces have attempted herd reductions, but their efforts produced ambiguous results. The single most important management strategy is to conduct the necessary research to determine means of transmission, ante-mortem diagnostics, genetic resistance, environmental disinfection, and treatment or prevention. Only when personnel are armed with such knowledge can they implement cogent management actions. Management actions that may help slow the spread of CWD include: harvesting any cervid showing clinical signs to prevent it from infecting susceptible animals; eliminating artificial feeding or other activities that concentrate cervids; and conducting intensive surveillance to identify "hot spots" of infection where herd reduction may be efficacious in decreasing spread of the disease.
Fig. 3. Location of animals examined for presence of chronic wasting disease in Wyoming since 1997. Light dots are positive deer and elk; dark dots are negative samples.

There are 3 major questions regarding CWD: 1) can humans contract CWD; 2) can domestic livestock contract CWD; and 3) what effect will CWD have on deer and elk populations? The possibility of humans contracting CWD is most likely the driving force behind public interest in this disease. If it was not for "mad cow" disease in Europe and subsequent development of variant Creutzfeldt-Jakob disease in people who ate infected beef (Bruce et al. 1997), there probably would be little or no interest in CWD in this country. There is no evidence, epidemiological or experimental, that suggests humans can contract CWD (Williams et al. 2002). Nonetheless, until fairly definitive research (such as exposure of chimpanzees, Pan troglodytes, to the CWD prion) demonstrates little or no human susceptibility to CWD, hunters are advised to take some basic precautions (Table 1; Williams et al. 2002).

Unlike on humans, susceptibility studies have been conducted on livestock. In the sixth year of ongoing research, there have been no signs of disease in cattle given a single oral dose of CWD-infected brain tissue or in cattle living amongst CWD-infected deer and elk (E. S. Williams, M. W. Miller, T. J. Kreeger, unpubl. data). However, 3 of 13
cattle became recumbent and were euthanized 24–27 months after being inoculated intracerebrally with the CWD prion. Microscopic lesions were found in 2 cattle and PrP\textsuperscript{res} was demonstrated in the brains of all 3 by immunohistochemistry (Hamir et al. 2001). Intracerebral inoculation is an artificial route of exposure and the Hamir et al. study was conducted to determine how CWD may present in cattle should they contract CWD naturally.

Table 1. Actions hunters can take to minimize exposure to the CWD agent and to decrease spread of the disease.

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<td>1.</td>
<td>Don't shoot any deer or elk showing clinical signs of CWD (emaciated, reluctant to move, excessive drinking, salivation, droopy ears).</td>
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<td>2.</td>
<td>Bone out the carcass; don't cut into the spinal cord if at all possible.</td>
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<td>3.</td>
<td>If you need to cut into the spinal cord or remove the head, make it your last step with the knife or saw. Then disinfect the saw or knife with household bleach (&gt;2% free chlorine, 280 ml in 720 ml water at room temperature for 1 hr).</td>
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<td>4.</td>
<td>Leave the skeleton, brain, spinal cord, lymph nodes, spleen, tonsils, and eyes in the field. These tissues appear to contain the greatest amount of CWD agent. Don't transport whole carcasses out of a CWD endemic area.</td>
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<td>5.</td>
<td>Wear rubber or latex gloves as a simple precaution against contact not only with the CWD agent, but with other bacteria and viruses as well.</td>
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<td>6.</td>
<td>Have your animal tested for CWD if such services are available.</td>
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Results of mathematical models suggested CWD could reduce cervid populations significantly after several decades (Miller et al. 2000). Although CWD has existed for more than 25 years in parts of Colorado and Wyoming, no decreases in deer or elk populations have been noted that could be attributed specifically to CWD. However, several obfuscating factors, such as drought, hinder accurate analysis of population fluctuations. There is concern that CWD may have more severe impacts on the dense populations of white-tailed deer found in the eastern U. S. In Wyoming, CWD prevalence is higher in white-tailed deer (17.6%) than in mule deer (9.5%). These data suggest white-tailed deer may be more susceptible to CWD or have behaviors that their increase exposure to the CWD agent. Mechanistic models indicated CWD epidemics do not stabilize if left unmanaged and populations of cervids could become extinct if CWD existed at high levels for an extended period of time (Gross and Miller 2001).

Chronic wasting disease will probably be one of the significant wildlife management challenges in the 21st century. The disease not only has serious wildlife ramifications, but economic, political, and social impacts as well. Although the "need" to blame someone for a problem seems to be inherent in humans, accusations and finger pointing do little to effect a solution. Nobody intentionally caused or spread CWD. But now that it is spreading throughout North America, we must all work together to minimize its impact on our natural resources.
LITERATURE CITED


Reviewer: J.C. deVos