

## ***Extension Responds: BSE***

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### **Chronic Wasting Disease and Potential Transmission to Humans**

From an article in the June 2004 issue of Emerging Infectious Diseases.

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Chronic wasting disease (CWD) is a transmissible spongiform encephalopathy (TSE), or prion disease, of the same category as BSE. While cattle are the natural hosts for BSE or Mad Cow Disease, deer and Rocky Mountain elk are the only known natural hosts for CWD. CWD and other TSEs are believed to be caused by a pathogenic effect on neurons caused by prion protein. The pathogenic form of this protein appears to be devoid of nucleic acids (genetic material) and supports its own amplification in the host.

CWD was first identified as a fatal wasting syndrome of captive mule deer in the late 1960s in research facilities in Colorado and was recognized as a TSE in 1978. Subsequently, this wasting disease was identified in captive deer and elk in both Colorado and Wyoming. The disease was first recognized in the wild in 1981, and by the mid-1990s, CWD had been diagnosed among free-ranging deer and elk in northeastern Colorado and southeastern Wyoming, and has since spread to adjacent areas in Nebraska. Surveillance data indicate that the overall prevalence of the disease in this area is approximately 5 percent in mule deer, 2 percent in white-tailed deer, and less than 1 percent in elk. The disease can be highly transmissible within captive deer and elk populations, but the mode of transmission is not clearly understood. Evidence supports lateral transmission through direct animal-to-animal contact or as a result of indirect exposure to prions in the environments, including contaminated food and water sources.

Concerns have been raised about the possible transmission of the CWD agent to domestic animals, such as cattle and sheep, which may come in contact with infected deer and elk or CWD-contaminated environments. If such transmissions were to occur, they would potentially increase the extent and frequency of human exposure to the CWD agent. In ongoing experimental studies, after more than 6 years of observation, no prion disease has developed in 11 cattle orally challenged with the CWD agent or 24 cattle living with infected deer herds. In addition, domestic cattle, sheep, and goat residing in research facilities in close contact with infected cervids did not develop a prion disease.

The increasing detection of CWD in a wider geographic area and the presumed foodborne transmission of BSE to humans, resulting in cases of Variant Creutzfeldt-Jakob Disease (vCJD) have raised concerns about the possible

transmission of CWD to humans. In the late 1990s, such concerns were heightened by the occurrence of CJD among three individuals who were deer hunters or ate deer and elk meat harvested by family members. However, an investigation into these illnesses indicated no strong evidence for a causal link between CWD and their CJD illnesses. Other cases have also been investigated. In 2001, the case of a 25-year-old man who reportedly died of a prion disease after an illness lasting 22 months was investigated. Although this man had hunted deer only rarely, his grandfather hunted deer and elk throughout much of the 1980s and 1990s and regularly shared the venison with the case-patient's family. The grandfather primarily hunted in southeastern Wyoming, around the known CWD-endemic area. The case-patient's illness began with a seizure and progressed to fatigue, poor concentration, and depression and worsening symptoms. A brain autopsy confirmed a prion disease diagnosis, but unrelated to CWD. It remains unknown whether the possible exposure of the case-patient to CWD-infected venison potentially contributed to the early onset of his prion disease. Other cases of CJD have also been investigated for potential links to CWD, but no direct links have been found. Recently, rare neurologic disorders resulting in the deaths of three men who participated in "wild game feasts" in Wisconsin in a cabin owned by one of the decedents created concern about the possible relationship of their illnesses with CWD. Two of the patients reportedly died of CJD, and the third died from Pick's disease. More than 50 persons were identified as possibly participating in these feasts; the three patients were the only participants reported to have died of a degenerative neurologic disorder. Reanalysis of autopsy brain tissues from the three patients at the National Prion Disease Pathology Surveillance Center indicated that two of them had no evidence of a prion disease by immunohistochemical analysis. CJD was confirmed in the third patient, who had participated in the feasts only once, perhaps in the mid-1980s. In addition, the investigation found no evidence that the deer and elk meat served during the feasts originated from the known CWD-endemic areas of Colorado and Wyoming.

Overall, despite the decades-long endemicity of CWD in Colorado and Wyoming, the incidence of CJD and the age distribution of CJD case-patients in these two states are similar to those seen in other parts of the United States. The age of disease onset for infected individuals in the two states is similar to the national average.

In conclusion, the lack of evidence of a link between CWD transmission and unusual cases of CJD, despite several epidemiologic investigations, and the absence of an increase in CJD incidence in Colorado and Wyoming suggest that the risk, if any, of transmission of CWD to humans is low. Although in vitro studies raise the possibility of low-level transmission of CWD to humans, no human cases of prion disease with strong evidence of a link with CWD have been identified. However, the transmission of BSE to humans and the resulting vCJD indicate that, provided sufficient exposure, the species barrier may not completely protect humans from animal prion diseases. Because CWD has

occurred in a limited geographic area for decades, an adequate number of people may not have been exposed to the CWD agent to result in a clinically recognizable human disease. The level and frequency of human exposure to the CWD agent may increase with the spread of CWD in the United States. Because the number of studies seeking evidence for CWD transmission to humans is limited, more epidemiologic and laboratory studies should be conducted to monitor the possibility of such transmissions. In the meantime, to minimize the risk for exposure to the CWD agent, hunters should avoid eating meat from deer and elk that look sick or test positive for CWD. They should wear gloves when field-dressing carcasses, bone-out the meat from the animal, and minimize handling of brain and spinal cord tissues. As a precaution, hunters should avoid eating deer and elk tissues known to harbor the CWD agent (e.g., brain, spinal cord, eyes, spleen, tonsils, lymph nodes) from areas where CWD has been identified.

For a full text of the article: Belay ED, Maddox RA, Williams ES, Miller MW, Gambetti P, Schonberger LB. Chronic wasting disease and potential transmission to humans. *Emerg Infect Dis.* 2004 June. Available from: <http://www.cdc.gov/ncidod/EID/vol10no6/03-1082.htm>

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