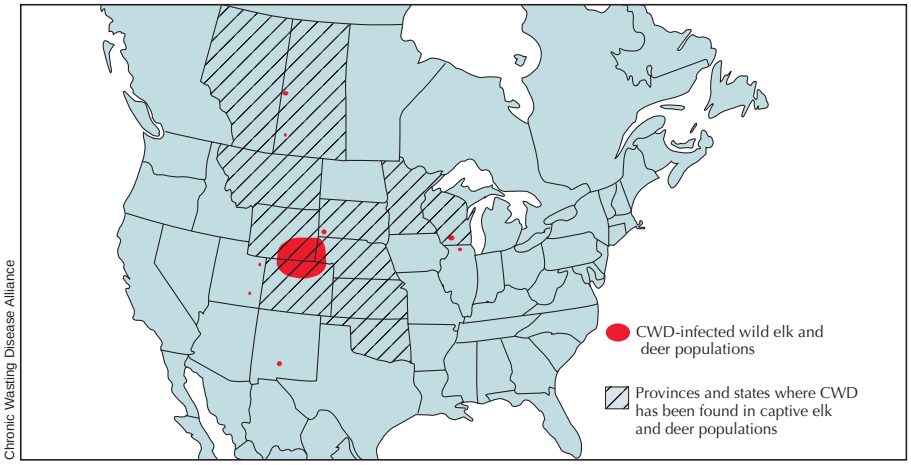
Wild game feasts and fatal degenerative neurologic illness

Background and epidemiology: Creutzfeldt–Jakob disease (CJD) is one of a group of conditions known as prion diseases that are caused by abnormally configured host-encoded prion proteins that accumulate in central nervous system tissue. There are 3 forms: sporadic, genetically determined and, more recently, variant CJD, acquired by infection when the disease crosses the species barrier between humans and cattle.[1] A similar prion disease in deer and elk known as chronic wasting disease (CWD) has raised concerns that this prion could also cross species barriers in people who eat wild game.



A report was recently published describing 3 cases of CWD in men who had attended wild game feasts.[2] In the first case, a 66-year-old man who had been a lifelong hunter and who ate venison frequently had hosted wild game feasts at his cabin in Montana for 26 years before he died in 1992 of an autopsy-confirmed subacute spongiform encephalopathy compatible with CJD. He had a history of seizures since the age of 43 and had sought treatment for recurring seizures, increasing forgetfulness and worsening hand tremors about a month before he died.

The 2 other cases involved men aged 55 and 65 who died of neurologic illness. These men had histories of eating venison and participating in 1 or more of the first patient's wild game feasts. The younger man presented in 1999 with a 3-month history of difficulty in writing and unsteadiness of gait, followed by dementia, speech abnormalities and myoclonic jerking. Pathologic examination of the brain at autopsy 3 months later revealed widespread subcortical spongiform lesions consistent with CJD. He had visited the first patient's cabin 12 times and had participated in 1 wild game feast.

The older man sought treatment in 1992 for progressive slowing of speech, worsening memory and personality changes. His speech became reduced to single-word utterances, and he died 10 months after initial presentation. He had been a lifelong hunter and had hunted mostly in Wisconsin but also in Wyoming and British Columbia. He had participated regularly in the first patient's wild game feasts. No game had been brought to the wild game feasts from his hunting trips outside of Wisconsin.

Testing of fixed brain tissue from all 3 men revealed no evidence on immunostaining for prions using antibodies to the prion protein in the first and third patients. In the second patient prion disease was confirmed by means of immunohistochemical and Western blot testing. The blot characteristics and prion disease phenotype were consistent with the common form of sporadic CJD, classified as M/M (M/V) 1. Subsequent genetic typing revealed methionine homozygosity (M/M) at codon 129 of the patient's prion protein gene, making it likely that the patient had the most common form of sporadic CJD.

Information obtained for 45 people of 53 identified as possibly participating in the wild game feasts revealed that only 34 had actually attended them. Of the 34, 7 were dead, including the 3 described here; none of the other 4 had died of a degenerative neurologic disorder. None of the living participants had any signs or symptoms consistent with a degenerative neurologic disorder.

Clinical management: Because of the continuing possibility that CWD in deer and elk might be transmissible to humans, physicians should attempt to confirm cases of illness compatible with a transmissible spongiform encephalopathy through pathological examination of brain tissue and by alerting the Canadian CJD surveillance unit (www.hc-sc.gc.ca/pphb-dgspsp/hcai-iamss/cjd-mcj; tel 888 489-2999) or the US National Prion Disease Pathology Surveillance Center (www.cjdsurveillance.com). There is no known treatment for the condition.

Prevention: CWD was first identified in the United States in 1967. In Canada, only 8 cases of CWD have been reported in wild deer, all in Saskatchewan. In the United States, CWD has been found in free-ranging deer and elk in Illinois, Nebraska, New Mexico, South Dakota, Wisconsin, Colorado and Wyoming. Limited epidemiologic investigations to date cannot exclude the possibility that CWD may cause human disease. Because of the severity of the spongiform encephalopathies in humans and the absence of treatment, animals with evidence of CWD should be excluded from human and animal food chains.

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References

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2. Fatal degenerative neurologic illnesses in men who participated in wild game feasts — Wisconsin, 2002. *MMWR Morb Mortal Wkly Rep* 2003;52(7):125-7.