FACTSHEET

ANIMAL SCIENCE ANIMAL DISEASES & PESTS

ORDER NO. 07-025

AUGUST 2007

AGDEX 400/663

Ontario Ministry of Agriculture, Food and Rural Affairs

CHRONIC WASTING DISEASE UPDATE

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Over the past 5 years, chronic wasting disease (CWD) has emerged as the most important disease to affect both wild and farmed cervid populations in North America. Since 2000, most jurisdictions in North America have developed surveillance programs to determine the presence or absence of CWD in both their farmed and wild cervid populations. In North America, more than 300,000 cervids have been tested for CWD in the past 5 years. Concerns over CWD transmission will affect the movement of live cervids and their products until better technology provides a live test with a high degree of sensitivity and specificity.

INTRODUCTION

CWD of elk and deer is an infectious disease causing animals to have increased salivation, polydypsia (increased drinking), polyuria (increased urination), excessive drooling, ataxia (incoordination) and progressive neurological signs. The incubation period can be up to 36 months, post exposure. CWD belongs to a group of diseases called transmissible spongiform encephalopathies (TSEs). The emergence of bovine spongiform encephalopathy (BSE) has raised the profile of CWD and other diseases in the TSE group. Hueston and Bryant have summarized the current knowledge on the TSE diseases[1].

Current research indicates that the agent associated with CWD is an abnormal infectious protein or prion. A prion is a normal cellular protein involved in synaptic (nerve junction) function at the neuron and coded by a single gene. The abnormal prion or proteinaceous infectious particle is protease resistant and abbreviated to PrP^{res}. The specific prion associated with CWD is abbreviated to PrP^{CWD}. PrP^{res} corrupt normal cellular prions and cause them to become protease resistant. Protease-resistant prions accumulate and cause vacuolation of neurons and loss of function. A recent theory is that a slow virus is the infectious agent and the abnormal prion is a manifestation of the infection[2]. There is no evidence that CWD can be transmitted naturally to other livestock or humans[3].

REGIONAL DISTRIBUTION

United States

CWD has been present in free-roaming elk, mule deer, white-tailed deer and black-tailed deer in an endemic area that includes northern Colorado, southern Wyoming and southwestern Nebraska since the late 1970s. The prevalence rate of CWD in the endemic areas of north-central Colorado and southeastern Wyoming can be as high as 17% in local populations of mule and white-tailed deer and about 1% in elk. Recently, a wild moose from the same area was identified as being infected with CWD.

Many states and provinces have increased their surveillance in both farmed and wild cervids. Wisconsin tested 69,000 samples from 2002 to 2005[4]. The common saying, "The more you test, the more you find," has unfortunately come true. Since 2001, the geographic distribution of CWD has expanded on farms and in the wild to include two Canadian provinces — Alberta and Saskatchewan — and at least 14 states — Colorado, Wyoming, Wisconsin, Kansas, New Mexico, New York, West Virginia, Minnesota, Illinois, Montana, Utah, South Dakota, Nebraska and Oklahoma. The increased geographic distribution may be due to heightened awareness and increased testing or to a true expansion of the range of the disease.

Canada

The first reported case of CWD in Canada was in a group of mule deer imported in 1974 via a shipment from the Denver Zoo to the Toronto Metropolitan Zoo. The last animal with CWD of the group died in 1981[5]. In 1996, CWD was diagnosed on 40 cervid farms in Saskatchewan. The source of this outbreak was traced to elk imported from South Dakota. This outbreak, coupled with a heightened awareness of the TSEs in the late 1990s, brought CWD to the forefront. In addition to the 40 Saskatchewan herds, two farms in Alberta in 2002 were also diagnosed with CWD. Approximately 9,000 farmed cervids were destroyed, with 233 being infected, 31 of them considered clinical[6].

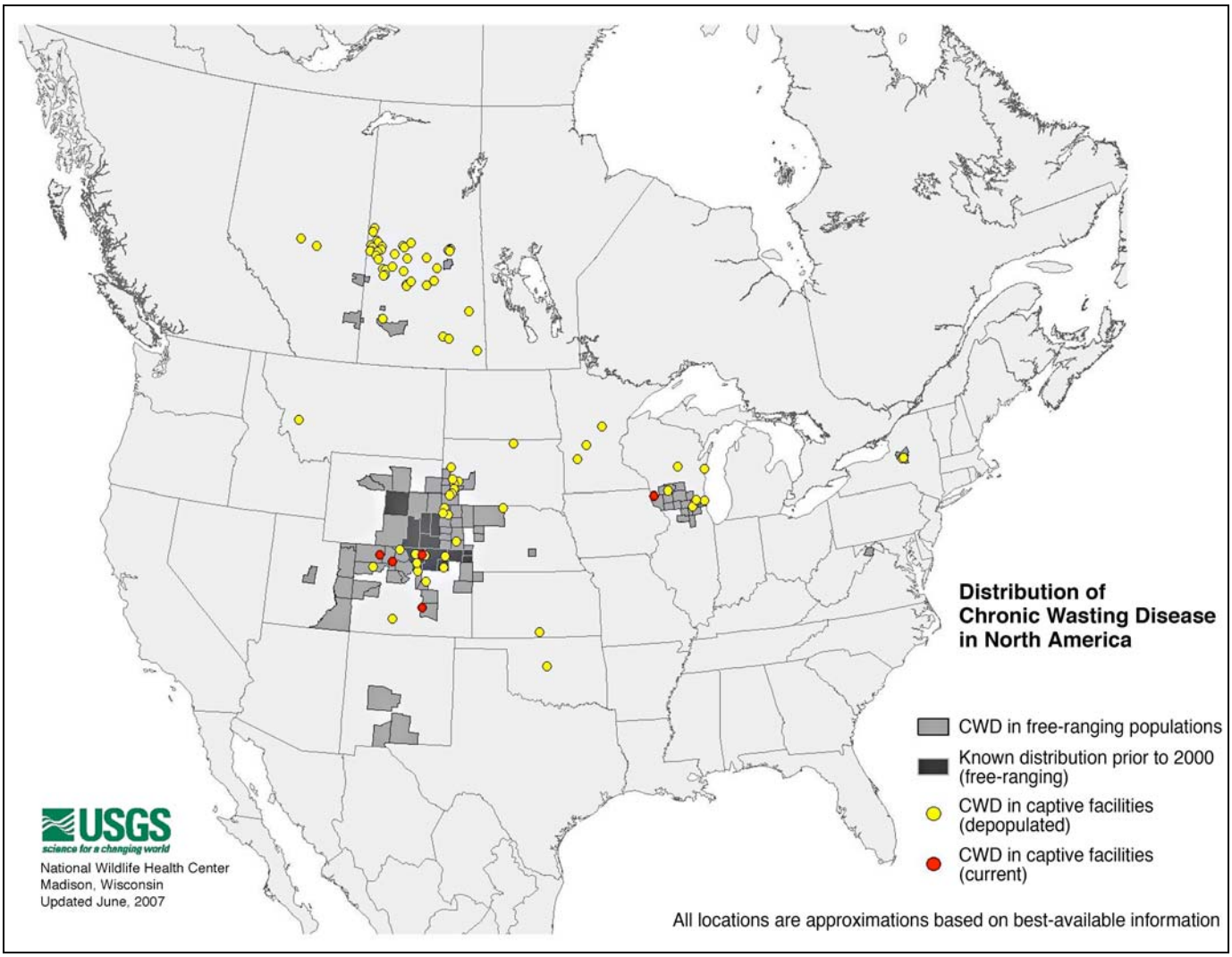

Figure 1. Distribution of Chronic Wasting Disease in North America current to June 2007.

Since the spring of 2000, wild mule deer and white-tailed deer in Saskatchewan and an adjacent area in Alberta have been confirmed positive for CWD. Since 2005, three more cervid herds in Saskatchewan have been found with CWD. CWD has not been diagnosed in the rest of Canada. Details of each state or province's experience with CWD are presented in the proceedings of the Second International CWD symposium[7]. The National Wildlife Health Centre summarizes CWD distribution in the map (see Figure 1)[8].

TRANSMISSION

Cervid Species

Epidemiology supports CWD as primarily being an infectious disease. Maternal transmission appears to be relatively rare[3]. During the eradication of the farmed herds in Saskatchewan, the infection rate in herds using stock tanks with large volumes of water (60 gal or more) was greater than in herds using small-volume water bowls[9]. This would be consistent with observations that cervids exhibiting clinical signs of CWD drool while drinking and spend more time around water sources than normal. These observations support the hypothesis that saliva and feces are important in the transmission of the agent[1, 3]. The PrP^{CWD} accumulates in gut-associated lymphoid tissue (e.g., tonsils, mesenteric lymph nodes). Saliva and feces are important in the shedding of the agent.

TSE infectivity can persist, when buried, for at least three years[10]. Practices that increase the concentration of cervids and, therefore, the environmental contamination (e.g., holding cervids in captivity or through artificial baiting of wild cervids) may be important in increasing the transmission of the agent. Casual fence-line contact is less likely to increase the possibility of transmission. Both direct (e.g., prolonged contact across the fence) and indirect transmission (e.g., from contaminated pastures) are possible. Shedding of the virus probably precedes clinical signs in both elk and deer[3].

In the Saskatchewan eradication of farmed elk, genetic analysis showed that the majority of CWD test-positive elk were homozygous for 132M. CWD was not detected in elk that were homozygous for 132L[6].

Wild Cervids

In free-roaming mule deer, adult males may play an important role in the spread of CWD. The prevalence of CWD peaks in 5–6-year-old males and shows no age-related pattern in females. Gender-related behaviour, such as the roaming of sexually mature males during breeding season with increased opportunities to interact with sources of infection and associate with numerous females, might explain the differences in infection rate (prevalence) between genders[11].

Domestic Livestock

Cattle, sheep and goats seem to be relatively resistant to infection. A long-term study intensively exposed cattle to CWD-infected deer and elk via oral inoculation or confinement with infected captive mule deer and elk. The cattle have remained healthy for over 5 years[3].

Humans

On occasion, media reports have indicated that hunters have died of Creutzfeldt-Jakob Disease (CJD). Investigations into these reports have not supported their claims[12]. Transgenic mice expressing the elk prion protein, "cervidized mice," were intracerebrally inoculated with elk CWD prion. Two genetic lines of "humanized" transgenic mice that are susceptible to human prions (PrP^{res}) were also intracerebrally inoculated with elk CWD prion. The humanized mice failed to develop CWD after more than 657 and more than 756 days respectively. In contrast, the "cervidized" transgenic mice became infected after 118–142 days. These data may indicate that there is a substantial species barrier for transmission of elk CWD to humans[13].

SURVEILLANCE

Wild Cervids

Many jurisdictions have developed surveillance programs for wild cervids using sampling frameworks based on geographic randomized sampling, using low prevalence rates (less than 1%) and probabilities of detecting the first case at or above the 95% confidence level. Various jurisdictions target their surveillance to areas based on perceived relative risk. Factors, such as proximity to provinces or states with CWD or areas where cervid farms are concentrated, are used to prioritize surveillance. Hunter-killed cervids are a common source of surveillance samples. In some areas, especially if there is no hunting season, road-kill submissions are used for CWD surveillance.

Farmed Cervids

Alberta, Saskatchewan and Manitoba currently have mandatory surveillance programs. All deaths and slaughter animals greater than 12 months of age must be tested for CWD. Quebec and Ontario have voluntary surveillance programs. The remaining provinces have very few or no farmed cervids. CWD is a reportable disease across Canada, where all suspect cases must be reported to the Canadian Food Inspection Agency (CFIA) under the *Health of Animals Act*.

In the U.S., a federal CWD herd certification program is in the development stages. Many states have placed a moratorium on the import of cervids, both farmed and wild, into their state. CWD surveillance programs vary from state to state; some programs are voluntary, and some are mandatory. The CFIA and U.S. Department of Agriculture are discussing harmonization of Canadian and U.S. import/export standards for CWD.

SAMPLE COLLECTION AND TESTING

From the late 1990s until the fall of 2005, diagnosis of CWD was made by the presence or absence of the abnormal protein PrP^{res} on immunohistochemistry (IHC); IHC is referred to as the gold standard for CWD diagnosis. The obex section of the brain stem (medulla oblongata) is the preferred site. It is collected and placed in 10% buffered formalin. Pathologists evaluate the staining in the region of the parasympathetic vagal nuclei. More recently, "rapid tests" have been developed and approved for use. Most of these "rapid tests" are ELISA based, while one is a commercial western blot assay. Fresh or frozen samples are submitted to the laboratory, and results are often available the same or next day. Bio-Rad ELISA is the most widely used test for CWD testing in Canada. In the last 3 years, the retropharyngeal lymph nodes (RPLN) have been the preferred tissue for testing in white-tailed and mule deer, while the obex remains the preferred tissue in elk and red deer[6]. Table 1 summarizes CWD surveillance in Ontario over the past 9 years. Biopsy and testing of tonsillar material has been tried[14]. Canadian researchers are currently evaluating the use of rectal mucosa as a live cervid test[15]. Researchers at the University of Guelph have been experimenting with an acoustic sensor for prion detection in blood and urine[16].

Because of the large expanse of Ontario, the impracticality of shipping whole carcasses or heads and the limited time span in which to collect suitable samples, training sessions and a training kit were developed to increase veterinarians' knowledge of CWD and to train them in the collection of tissues for testing.

Table 1. Ontario Sample Submissions for CWD — 1998 to July 26, 2007 (Farmed cervids, except where indicated).

Year	Submissions	Elk	Reds and Elk/Red Hybrids	White-tailed Deer	Fallow Deer	Other Reindeer, Moose	Unspecified
1998 Total submissions	51	15	18	10	6	2	
1999 Total submissions	47	16	15	9	3	4	
2000 Total submissions	50						
Histology on brain	19						
IHC	6	5	1				
2001 Total submissions	71						
IHC	25	19	3	3			
2002							
Wild (MNR) – IHC	155	4		151			
Farmed – IHC	121						
Total – IHC	**276**						
2003							
Wild (MNR) – IHC	613						
Farmed – IHC	241	167	16	47	1	10	
Total – IHC	**854**						
2004							
Wild (MNR) Bio-Rad	420						
Farmed – IHC	234	115	23	52		1	43
Total IHC & Bio-Rad	**654**						
2005							
Wild (MNR) Bio-Rad	1,436			1,436			
Farmed Bio-Rad	109	41	16	43		1	9
Farmed – IHC	1						
Total IHC & Bio-Rad	**1,544**						
2006							
Wild (MNR) Bio-Rad	1,451			1,451			
Farmed Bio-Rad	316	202	26	88			
Total	**1,767**						
2007 to July 26/07							
Wild (MNR) Bio-Rad	Not reported						
Farmed Bio-Rad	203	104	28	50	20	1	
Total	**203**						

IHC = The number of immunohistochemistries performed. Full necropsies may have been performed in addition to the IHC.
Wild = Data from wild deer, including hunter-killed white-tailed deer (WTD), elk from the elk restoration project and WTD from park culls.
MNR = Ministry of Natural Resources.

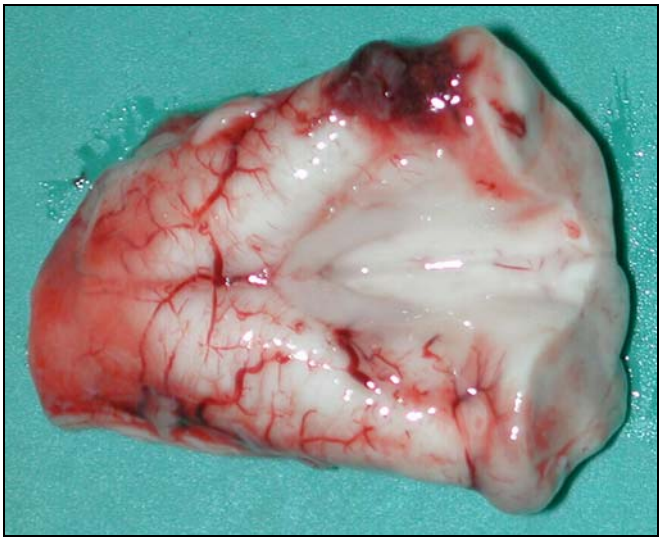

Figure 2. The brain stem is cut 1 cm above and below the obex (the "V" on the dorsal surface of the brain stem).

If Bio-Rad is being used, place the obex in a whirl-pack and keep it refrigerated or freeze it prior to submission to the laboratory. If immunohistochemistry is being used, place the obex in 10% buffered formalin. Some jurisdictions may require samples to be split, with half placed in buffered formalin and half fresh or frozen.

The Animal Health Laboratory (AHL), at the University of Guelph, is now using Bio-Rad to test for CWD and is no longer using IHC. All samples from Ontario to be tested using Bio-Rad must be shipped fresh or fresh/frozen. Ontario samples can be shipped to the AHL by Purolator at no cost by using "Purolator – University of Guelph incoming account #096691" on the waybill. Samples shipped in formalin can no longer be tested by AHL.

SAMPLING AT ABATTOIRS

When developing surveillance programs of slaughter animals, it is necessary to hold all carcasses until the samples have been declared free of CWD. The turn-around time between the collection, testing and release of carcasses can be a problem when cooler capacity in an abattoir is limited. With the new rapid tests having 24–36-hour turn-around times, this is less of a problem but requires the coordination of meat inspection and laboratory services.

SUMMARY

With the detection of CWD in at least 14 states and two provinces, its long incubation period, the public's heightened awareness and fear of the TSE diseases, it is critical that deer farmers and governments continue with surveillance to halt the spread of CWD.

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ISSN 1198-712X
Également disponible en français
(commande n° 07-026)