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REVIEW

Chronic Wasting Disease in Deer and Elk: Scientific Facts and Findings

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ABSTRACT. Chronic wasting disease (CWD) is a prion disease of cervids such as deer and elk in North America. Unlike other transmissible spongiform encephalopathy (TSE) such as scrapie, CWD occurs in both captive and wild ranging animals, but not in domestic ruminants such as sheep and cattle. In this paper, the history of the disease, pathogenesis of CWD, susceptibility of animals, its transmission mechanisms, potential origins of the disease, diagnostic methods in the field and laboratory tests, surveillance and survey systems in the USA and Canada, control strategies, economic impact of the disease, food and feed safety, and the risks in human and animals are reviewed and summarized. Although there is no evidence that CWD has been transmitted to humans, it may have the potential to infect humans.
KEY WORDS: CWD, diagnostic method, food and feed safety, pathogenesis, surveillance

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Chronic wasting disease (CWD) is a transmissible spongiform encephalopathy (TSE) that can affect specific species of native North American deer, including mule deer (*Odocoileus hemionus*) and white-tailed deer (*Odocoileus virginianus*) as well as Rocky Mountain elk (*Cervus elaphus nelsoni*). The disease is found in both captive (farmed) and free-living populations of these species. The purpose of this paper is to present the current scientific knowledge about this disease.

HISTORY OF THE DISEASES

CWD was first identified in the late 1960s in captive mule deer in a Colorado wildlife research facility. Researchers working on natural history and nutritional studies with captive mule deer observed the clinical signs and called the syndrome chronic wasting disease (CWD). It was initially thought to be associated with the stresses of captivity, nutritional deficiencies, or intoxication. Later, the disease was recognized as a spongiform encephalopathy-forming disease through histological studies [27]. The disease was also recognized in Rocky Mountain elk [26]. Its neuropathology included the "daisy plaques" which are also a unique abnormality of the new variant Creutzfeldt-Jakob disease (vCJD) in humans. The occurrence of CWD remained limited to captive mule deer until 1981, but in the 1990s the disease was found in free-ranging mule deer, white tail deer, and elk in Colorado and Wyoming. This is the only TSE known to affect free-ranging wildlife species. Little attention was paid to this disease in its early discovery, however, it received much more attention after the potential link between vCJD and Bovine Spongiform Encephalopathy (BSE) was identified, and now many researchers and regulators in public health, wildlife, and animal health have intensified their interest in this disease.

By the year 2000, CWD had been identified in both farmed and free-ranging animals in several states neighboring the first reported case, as well as in contiguous regions of Canada. Intensified recent surveillance has identified what appears to be an ever-expanding geographic range. Cases have been identified in the western portion of Colorado, in Wisconsin, Minnesota, New Mexico, and Utah and some imported cases have been reported in South Korea [17].

PATHOGENESIS

The pathogenesis of CWD in its natural setting shares several similarities with its related diseases, mainly scrapie and BSE. The pathogenesis consists of early involvement of the lympho-reticular system, including gut-associated lymphoid tissue with incubation periods ranging between 15 and 36 months, depending on the species and conditions of infection. Minor differences in the amount and distribution of abnormal protein in different body tissues have been observed in deer and elk. It has not been detected, however, in either muscle or "antler velvet" - two products consumed by humans. Spongiform changes are present in the medulla oblongata, especially the parasympathetic vagal nucleus and in the thalamus, hypothalamus and olfactory cortex and are often severe. The disease specific abnormal prion protein, PrP^{CWD}, as demonstrated by immunohistochemistry (IHC) is found in the brain, palatine tonsils, visceral and regional lymph nodes, Peyers patches and other lymphoid tissue of the small and large intestine and also in the spleen of affected deer [16]. In the brain, the disease specific PrP accumulation and spongiform change is seen initially in the dorsal motor nucleus of the vagus nerve [15, 23, 24] detected PrP^{CWD} in the brain stem, spinal cord, pituitary (*pars intermedia and pars nervosa*), vagosympathetic trunk, sympathetic trunk, nodose ganglion, myenteric plexus, adrenal medulla, pancreatic islets, brachial plexus, sciatic nerve, but not in the trigeminal (gasserian) ganglion, coeliac ganglion, cranial cervical ganglion or spinal nerve roots. These findings suggest that there is, at least in the clinical disease, extensive involvement of multiple organ systems, including

central and peripheral nervous tissues, endocrine organs and the alimentary tract, the latter suggesting a possible means of agent shedding. Immunohistochemical evidence of disease specific PrP has not been found in the mucosa of the abomasum and intestines, thymus, bone marrow, skeletal muscle, liver, lungs, myocardium, walls of vessels, kidney, bladder, ovary, endometrium, testis, epididymis, sebaceous and sweat glands, and epidermis of skin of affected deer. In elk, PrP^{CWD} has been detected by IHC in the myenteric plexus, the vagosympathetic trunk, the cell column of the spinal cord and endocrine glands. PrP^{CWD} accumulates first in the dorsal motor nuclei of the vagus nerve at the level of the obex of the medulla and this accumulation precedes the development of lesions [15].

Brain lesions, associated with clinical disease in deer, have been found 16 months after experimental infection and in elk from the age of 12 months, whereas immunohistochemical demonstration of PrP^{CWD} is achieved much earlier, sometimes several months, or up to a year, in both lymphoid tissues and CNS.

There are no reported studies of tissue infectivity bioassays in CWD because there are no adequate biological models available to detect CWD infectivity and because the substantial resources necessary to conduct bioassays in deer and elk have not been allocated. This is an important omission in the research, which prevents any quantification of infection relative to tissue/organ.

SPECIES SUSCEPTIBILITY AND CROSS SPECIES

A major determinant of susceptibility to the TSE diseases is the host PrP gene. Genetic homology between species confers similarities and divergence in the spatial configuration of the respective protein, and is an important element of the structural basis of the species barrier. Only three species of cervidae are known to be naturally susceptible to CWD: mule deer, white-tailed deer and Rocky Mountain elk. One case was originally reported in black tailed deer (*Odocoileus hemionus columbianus*) [27], a subspecies of mule deer. Hybrid animals of mule deer and white-tailed deer have also been affected. Other non-domestic ruminants, including moose (*Alces alces*), pronghorn antelope (*Antilocapra americana*), Rocky Mountain bighorn sheep (*Ovis canadensis canadensis*), mouflon (*Ovis musimon*), mountain goats (*Oreamnos americanus*), and a blackbuck (*Antilope cervicapra*), have been in contact with CWD-affected deer and elk or have resided in premises in which CWD had occurred and have not developed the disease [25]. Cattle, sheep and goats that have resided in research facilities together with CWD-affected animals for prolonged periods or under field conditions did not develop the disease. These observations of apparent cross-species resistance are supported by molecular studies of [13] and *in vivo* studies of [8].

Several experimental studies to transmit CWD have been conducted, most by intra-cerebral (IC) inoculation. While such studies provide information on susceptibility to the most efficient means of interspecies transmission, they do not inform on interspecies susceptibility by natural routes of transmission. For the latter oral or other possible natural exposure route studies are considered the most appropriate. On-going research on the species barrier is indicating that there is a substantial biological barrier to transmission of CWD from deer to cattle. Preliminary data from experiments in progress in the USA indicate that only a few calves develop disease after challenge with CWD pathogen from affected mule deer using the intra-cerebral(IC) inoculation route of transmission. Cattle have been inoculated orally with a brain tissue pool from CWD-affected mule deer at the University of Wyoming and have not developed any evidence of transmission more than five years following exposure. These studies are scheduled to run for ten years. In addition, bovine calves have been orally inoculated with CWD brain tissue pools from mule deer and from elk; these calves are being sequentially necropsied and results are not yet available (Williams, *pers comm*).

Cattle living in close contact with infected deer and elk have not developed the disease during the first five years of a ten-year study. Twenty-four cattle were housed with resident deer and elk with endemic CWD, in two wildlife research facilities in Wyoming and Colorado. These studies started in 1997 and to date there is no evidence of transmission of CWD to cattle through contact. Control deer have all succumbed to CWD. Brains from cattle over five years of age and from different ranches within an enzootic area of CWD were examined with H&E and IHC stains and all were found to be negative (Gould, *pers. comm*).

Kaluz *et al.* [4] and O'Rourke *et al.* [10], indicated that the sequences of the prion protein gene are very similar between certain cervidae. Thus, it is possible to derive a conclusion from a specific study on one of these species.

Polymorphisms of the normal PrP gene influence susceptibility to infection and disease phenotype. In Rocky Mountain elk, sequence analysis of the PrP gene showed only a single polymorphism; one amino acid change (Met to Leu) at codon 132. It was found among 43 genotyped free-ranging and farmed Rocky Mountain elk that were positive for CWD, homozygous for PrP codon 132-Met (M/M) were over-represented when compared to unaffected control groups. In the same group, several heterozygous M/L were positive. Positive elk with the homozygous codon 132 L/L were not found [10]. Research is continuing into the influence of genetics on susceptibility; there may be an association between PrP genotype and resistance in elk but this has not been recognised [10]. A phylogenetic analysis suggested that cattle and mule deer have converged with great apes including humans in key areas of their prion protein [6]. It is, therefore, difficult to draw specific inferences from these data but such studies provide indications as to species in which the PrP gene should be examined in more detail.

A recent report described CJD in "unusually young patients who consumed venison", and although epidemiological and molecular biological investigation failed to show a convincing link between exposure and disease, the conclusion that these patients were most likely cases of spo-

radic CJD must be weighed against the fact that we do not know what CWD in humans would look like - it might look like sporadic CJD, or vCJD, or might have distinguishing characteristics unlike either form of disease.

There is epidemiological and biological strain typing evidence that the occurrence of spongiform encephalopathies in closely related wild ungulate species held in British zoological collections contemporaneously with the epidemic of BSE were due to food borne exposure to the BSE agent via contaminated proprietary ruminant feedstuffs. Such cases occurred only in species within the family Bovidae (subfamilies *bovinae* and *hippotraginae*) [5] and a considerably greater range of species, not only within the order Artiodactyla, but across several other orders, was exposed to feeds containing animal proteins. Within the Artiodactyla, an estimated 62 species were held in British zoos in 1989 [5] and undoubtedly this included members of the family *Cervidae*. The extent to which such species were exposed to commercial feedstuffs or supplements at the time is not known, but the practice was commonplace.

DISEASE TRANSMISSION

There is considerable evidence that CWD is both infectious and contagious but specific details of its transmission remain as yet to be determined. However, historically the epidemiology of CWD does not support its being a feed-borne disease like BSE, associated with rendered ruminant meat and bone meal (MBM). Evidence for this includes (1) the observations that captive cervidae without records of being fed with animal-protein also succumbed to the disease and (2) free-ranging animals are unlikely to have access to compound feed stuffs.

Lateral transmission, compounded by animal movements, is the most important factor in spread of CWD. Indirect transmission via environmental contamination may play a role in natural dynamics and persistence of the disease and thus exacerbate the spread of the disease, and may present an obstacle to eradicating CWD from infected premises.

Observational studies suggested that lateral transmission, similar to that experienced in scrapie epidemics, occur in CWD and is the most important factor impacting the spread of the disease [9, 25]. The presence of the CWD agent in lymphoid tissues of the alimentary tract suggests that the agent may be shed through the alimentary tract (feces and saliva). Contaminated pastures used by captive cervidae appear to have served as sources of infection in some CWD outbreaks. The potential role of invertebrate and/or vertebrate reservoirs in the spread of CWD warrants further study, as does the influence of weather conditions on disease persistence, especially in free-ranging populations. Rapid increases in prevalence within captive herds suggest transmission may be quite efficient, at least at a local level. Recently four Saskatchewan elk farmers were advised not to grow grain or raise livestock on certain parts of their land since it may harbor CWD. Restocking pastures after leaving them clear for more than ten years was no guarantee for complete removal of possible contamination and sentinel programs were initiated to test these pastures.

There is less evidence for the existence of maternal transmission but because this cannot be distinguished from the high component of lateral transmission, it is not possible to exclude it. Placentomes, ovaries and fetal tissues from two mule deer in term pregnancy were examined with IHC and PrP^{CWD} was not detected [17], in contrast to the finding of PrP^{Sc} in pregnant domestic sheep with scrapie [19]. Tuo *et al.* [19] demonstrated that accumulation of PrP^{Sc} in uterine-placental epithelial cells in the placentome was determined by the pregnancy status of scrapie-infected ewes. The distribution of PrP^{Sc} plaques in placentomes showed a tendency toward increased size and number of placentomal PrP^{Sc} plaques from the endometrial stalk (maternal side) to chorionic plate (fetal side). In any case, maternal transmission alone is unlikely to sustain epidemics of CWD [7].

Both sexes and a wide range of age classes of animals can be affected, underscoring the likely importance of animal-to-animal (lateral) transmission in sustaining epidemics. Both intra- and inter-specific transmission (e.g., mule deer/white-tailed deer, elk/white-tailed deer) probably occurs. The infectious period is unknown but it appears likely that PrP^{CWD} shedding is progressive through the disease course. The presence of PrP^{CWD} at the beginning of the incubation time in alimentary tract associated lymphoid tissues suggests that shedding may take place early on [16].

THE ORIGIN OF CWD

There is no epidemiological evidence that would suggest the origin of CWD. As indicated above, there is no evidence to support a feed-borne common source origin of CWD. Hypotheses as to the origin of the disease might include:

1) Infection of deer by a strain of scrapie that has adapted to cervidae [23].
2) A genetic form of TSE arising in deer, with subsequent natural transmission.
3) Exposure to a currently unknown TSE, expressing the possibility, borne particularly out of the infancy of the study of diseases of wildlife, that there could be undetected TSE or prion diseases in other species.
4) A spontaneous conformational change of the prion protein occurring in mule deer, with subsequent transmission to other deer and to elk.

None of these hypotheses provide a particularly plausible explanation but further consideration of the evidence against a sheep scrapie origin is necessary. Given the endemic occurrence of scrapie in North America, a scrapie origin might be considered the commonly accepted theory, but even this has substantial counter arguments. Scrapie in sheep has an almost world-wide distribution and is present in many countries that harbor free-ranging deer but CWD has not been reported in deer populations of countries outside of North America. Although CWD transmits to goats [25] and to sheep [3] by IC inoculation, the incubation

period (more than six years in goats) produced suggests a large species barrier and this is not what might be expected if the agent were originally a sheep scrapie agent strain. In addition, biological strain-typing in inbred mouse strains has shown that the CWD agent differs from the BSE agent and from strains of scrapie tested thus far)[2]. Lastly, comparisons of abnormal PrP glycoform patterns from CWD-affected deer and elk, and scrapie-affected sheep and cattle did not provide reliable indications of TSE infections of common origin among the species studied [12].

DIAGNOSIS

Clinical signs of CWD are not specific. A consistent clinical sign of CWD in deer and elk is progressive weight loss. Behavioral changes also occur in the majority of cases, including decreased interactions with other animals, listlessness, lowering of the head, drooping ears, blank facial expression and repetitive walking in set patterns. In elk, behavioral changes may also include hyper-excitability, nervousness, ataxia and head pressing. Free-ranging, CWD-affected elk may lose the fear of humans. Affected animals continue to eat grain but may show decreased interest in hay. In deer and elk polydipsia and polyuria also commonly occur. Excessive salivation and grinding of the teeth are also observed. The clinical disease is progressive and always fatal.

In captive herds experiencing a new outbreak of CWD, there is frequently a history that includes sporadic cases of prime-aged animals losing condition, being unresponsive to symptomatic treatment and dying from aspiration pneumonia. This pneumonia, presumably caused by difficulty in swallowing and by ptyalism, may lead to misdiagnosis of the condition if there is not histological and/or immunohistochemical examination of nervous or/and lymphoid tissues. "Sudden deaths" following handling also have been reported as the index cases in some situations as have unusual traumatic losses.

Most cases of CWD occur in adult animals. The majority of CWD-affected animals are 3–5 years of age. The oldest elk with CWD was >15 years old. The clinical course of CWD varies from a few days to approximately a year, with most of animals surviving from a few weeks to three or four months. Caretakers familiar with individual animals often recognize subtle changes in behavior well before serious weight loss occurs.

Differential diagnoses include mineral deficiencies that lead to neurological symptoms in deer and elk (e.g. fading elk syndrome, listeriosis, and copper deficiency).

Evidence of non-clinical CWD infection has been seen in deer fawns and elk calves by about six months of age (Spraker, Pers comm). The youngest naturally-infected mule deer diagnosed with clinical disease was 17 months of age. CWD has been diagnosed in a 24-month-old Rocky Mountain elk [1].

Gross lesions seen at necropsy reflect the clinical signs, primarily emaciation. Aspiration pneumonia, which may be the actual cause of death, is also a common post-mortem finding in animals affected with CWD.

LABORATORY TESTING

On microscopic examination, spongiform lesions of CWD in the central nervous system resemble those of other TSE's. Lesions are usually found in several nuclei in the medulla oblongata, pons, mesencephalon and telencephalon in clinically-affected animals [24, 18]. The parasympathetic vagal nucleus in the dorsal portion of the medulla oblongata at the obex is the most important site to be examined for diagnosis of CWD, especially in apparently clinically normal animals [11, 17].

Immunostaining of tissues using PrP antibodies can demonstrate disease specific prion protein in the brain, palatine tonsils, visceral and regional lymph nodes, Peyers patches of the small intestine, lymphoid tissue of the large intestine, and the spleen of affected deer. Immunohistochemistry (IHC) currently used as the 'gold standard' in testing for different TSEs, is also used to test brain tissue for the presence and accumulation of PrP^{CWD}, the protein marker used to diagnose CWD. The area of the brain used for testing (parasympathetic vagal nucleus of the medulla at the obex) is critical and if the correct area of the brain is not tested, this must be considered. Testing of both brain and lymphoid tissue is preferred.

The current rapid tests used for BSE in Europe are being evaluated for their usefulness as screening tests for CWD [14]. The Bio-Rad CWD ELISA test used on lymph node tissue has recently been licensed in the US for mule deer, elk and white-tailed deer. The IHC and Bio-Rad ELISA both provide reliable results in testing for CWD. The latter test was used in some veterinary diagnostic laboratories on samples from Colorado and Wyoming. To date, slightly over 27,000 tests in 25,000 animals with approximately 200 positive animals (mule deer, elk and white-tailed deer) have been run using the Bio Rad ELISA for free-ranging cervidae surveillance.

Tonsillar biopsies have been assessed for the diagnosis of CWD in live animals [20, 28]. This technique is useful for the pre-clinical diagnosis of CWD in farmed live mule deer and white-tailed deer. PrP^{CWD} accumulates in tonsillar and lympoid tissues in an early stage of the infection and can be detected with IHC 2 to 20 months before a CWD-related death and up to 14 months before the onset of clinical signs of CWD. These studies suggest that tonsillar biopsy is a valid method for detecting CWD in live deer during incubation stage, and may be used as an ante-mortem and pre-clinical diagnosis and as an adjunct management tool. This technique is currently being evaluated as a practical management tool under field conditions (i.e. involving the capture, anaesthetic and biopsy of wild deer) [28].

A third eyelid test used in sheep for the diagnosis of scrapie was examined for the pre-clinical identification of infected animals [10]. This approach, however, does not seem feasible in deer and elk due to the very limited amount

of lymphoid tissue associated with the third eyelid in these species (Miller and Spraker, unpublished data).

SURVEILLANCE AND SURVEY SYSTEMS

During the last 10–15 years, several wildlife and animal health agencies have initiated a series of surveys that include hunter-killed and -targeted sampling areas as well as deer and elk farms for the purpose of determining the extent of the infection in free-ranging and farmed cervidae. These surveys were mainly focused in the states of Colorado and Wyoming and to some extent on selected elk and deer farms across the USA and Canada. Most of these surveys, however, were initiated as a reaction to a reported case with the focus on determining the prevalence instead of a being part of a planned surveillance system.

These surveys have identified CWD cases in free-ranging mule deer in Wyoming, Colorado, Nebraska, South Dakota, and New Mexico. The disease has also been found in free-ranging elk in Wyoming, Colorado, and South Dakota. Similarly it has been found in free-ranging white-tailed deer in Wyoming, Colorado, South Dakota, Nebraska, Wisconsin, Illinois, and Utah. With continuing and planned levels of these surveys the distribution and level of prevalence may change over a period of only a few months.

In addition, CWD has been diagnosed in farmed elk herds in a number of states in the United States and in two Canadian provinces. The current US national surveillance plan for farmed cervidae herds includes:

1) Mandatory death reporting.
2) CWD testing of all animals, except calves, which are slaughtered or die on the affected premises.
3) Individual animal identification and annual census.

Surveillance for CWD in US farmed elk began in 1996 and has been a cooperative effort involving state agriculture and wildlife agencies, the U.S. Department of Agriculture's (USDA), and Animal and Plant Health Inspection Service (APHIS). Farmed cervidae surveillance has been increasing each year since 1997 and will become an integral part of the USDA program to eliminate CWD from farmed elk. The farmed cervidae surveillance program and the surveillance program for wildlife are interdependent. Particular aspects of surveillance programs depend upon conditions in each state. For areas with known CWD infections, estimates of disease prevalence can be used to judge the effectiveness of management actions and to evaluate disease dynamics in the context of ecological research questions. Surveillance activities are also needed to satisfy public and management information needs. The CWD-positive elk herds in the United States include South Dakota, Nebraska, Colorado, Oklahoma, Kansas, Montana, and Minnesota. CWD has been also diagnosed in farmed white-tailed deer in Wisconsin.

In late 2002, the Colorado Division of Wildlife in cooperation with the Colorado Department of Agriculture and Colorado State University initiated a planned surveillance as a model for hunted cervidae in Colorado for CWD. The rapid screening test (BioRad ELISA) was applied on a volunteer basis to screen more than 25,000 samples from Colorado elk and deer. The IHC was used as a confirmatory test for those samples testing positive by screening. Findings from this survey will be available soon.

In Canada, CWD has been diagnosed in deer or elk on at least 40 game ranches in Saskatchewan and in farmed white-tailed deer on one ranch in Alberta (since 1996). Of these, 95% of infected elk herds had only a few (1-3) infected animals as diagnosed by IHC on the brain. Most (91%) elk diagnosed with CWD were at a pre-clinical stage. Approximately 65% of infected herds in Saskatchewan had a prevalence of infection less than 5%. While animals under 12 months of age have been diagnosed with pre-clinical infection by IHC, the youngest elk diagnosed with clinical CWD was 17 months old. Canadian veterinary services consider that the incubation period for CWD is 16-36 months, with a mean of 22 months. With elk, as with deer, animals of all ages and both sexes have been found infected with CWD and no bias has been evident.

Until 2000, there was no active surveillance for CWD in Canada. The government is in the process of conducting retrospective inspections of all farms that have imported animals from the United States, with emphasis on those farms where imported animals died within three years of importation. Provincial Government surveillance has provided valuable information on CWD. A voluntary national CWD certification program was recently introduced to provide access to herd replacements of known ('certified') CWD status and to meet the requirements of trading partners. Subject to conditions, herds that have been enrolled in voluntary CWD certification programs can enter the federal program at higher entry level status.

There is no published information on the possible occurrence of, or surveillance for, TSEs in cervidae species on the European continent. Throughout the world (particularly in Europe, North America and Australia), pathological examinations will have been carried out on numerous species of deer that have died in, or have been culled from zoological collections. In many cases, this will have included histopathological examination of the brain. None of the cases from Europe, Australia, and New Zealand have indicated such disease.

Several zoological gardens and wildlife research institutes were contacted for further information on surveillance of cervidae. From data received, it is concluded that currently minor surveillance activity is on-going or planned for CWD in cervidae.

CONTROL STRATEGIES

Control measures in general include prevention of introduction, notification of the disease, control or ban on movements, quarantine, eradication of affected herds, and compensation, and measures to prevent/stop the spread from free-range to farmed animals (and vice versa). Because of the commercial aspect of game ranching, animals were commonly moved across the US and Canada. Recently,

laws have been passed to prevent the movement of these captive animals across state lines. Some states will not allow any parts of animals into their state, if the origin of the meat/tissue is from an area in which CWD is known to occur.

There is also some natural movement of deer and elk across state lines. Knowledge of herd management, prevalence of CWD, and susceptibility factors may provide additional support for efficient controls. For example, it can be predicted that a hierarchy of prevalence is likely among the species (white-tailed deer>mule deer>elk) given that white-tailed deer are more social and found at higher densities.

Several states in the USA have recently banned or restricted the importation of deer species, including North Carolina, Michigan, Vermont, Tennessee, Texas (March 2002), Nebraska, Wisconsin, New York, Colorado, and Arizona. In New Mexico, upon recognition of the disease in free-ranging mule deer, the state immediately stopped any importation of deer or elk. Following the screening of herds, herd certification may be an option. However, given the limited knowledge on the incubation of the disease and its variation in clinical presentation, it is likely to take as long as five years of surveillance of all juvenile and adult mortality before a farmed herd may be certified as being free from CWD. The United States FDA Center for Veterinary Medicine announced in November 2002 a proposed policy on rendering tissues from cervidae from CWD-positive areas or herds.

Key elements of the Canadian eradication for farmed and captive cervidae are as follows:

1) CWD is reportable under the Federal Health of Animals Act (since 2001).
2) The finding of an infected animal (confirmed by IHC in the government laboratory) triggers a series of events :
   a) Imposition of quarantine on all animals and animal products at the affected farm.
   b) Slaughter of all cervidae.
   c) Testing of all adult cervidae in a government laboratory.

Since the eradication program commenced in February 2000, the Canadian government has slaughtered approximately 8,300 farmed elk on the affected farms (40 in Saskatchewan and 1 in Alberta) and tested 7,153 adult animals (99 % elk) and has detected a total of 230 elk infected with CWD to date. It has cost the federal government 33 million (Canadian) to compensate the farmers (Peart, pers comm).

ECONOMIC IMPACT

It is obvious that there has been a significant impact on the North American farmed cervidae industry from CWD but the total effect is difficult to quantify. There has been some influence, bearing, consequence, and repercussions on the sale of hunting licenses in different US states (e.g. Wisconsin). Public awareness has been raised by multiple forms of outreach by many agencies. A huge cost is involved in the compensation of Canadian farmers where animals were eradicated on CWD-positive farms. The cost of quarantine of farm and grassland in an attempt to reduce the environmental contamination following CWD in a farmed herd is difficult to quantify. CWD has also had a major impact on the deer and elk farming industry. Elk are raised for the production of antler velvet and meat and for trophy hunting. About 70% of velvet antler was formerly exported to South Korea. In the course of Canadian eradication activities and the detection of an increasing number of cases in 2000-2001, some trading partners closed their markets to Canadian cervids and cervid products, including semen, embryos and velvet. It is difficult to determine the total economic impact of this market closure.

FOOD AND FEED SAFETY AND HUMAN AND ANIMAL RISKS

There is no evidence that CWD can be transmitted to humans consuming meat or handling infected cervids or their products, however this possibility cannot be ruled out. The World Health Organization recommends that people not consume animal products from any animal infected with a TSE disease and public health policies in Canada and the US are consistent with this directive. In North America, some health officials advise hunters not to consume meat from animals known to be infected with the disease. In addition, they suggest hunters take simple precautions when field dressing deer or elk taken in areas where the disease is found. In the USA, the consumption of meat from CWD-affected animals is discouraged; however, there is no ban. So, affected meat probably has been consumed for decades in Colorado and Wyoming. In Canada, all adult cervidae slaughtered under commercial arrangements in the provinces of Saskatchewan, Manitoba, and Alberta are tested for CWD and carcasses are only released upon receipt of a negative result. Offal may be disposed off by incineration or deep burial before test results are known. Once a farmed cervidae is diagnosed with CWD, the infected animal and all cervidae exposed to positive animals are destroyed and the carcasses disposed of by incineration or deep burial. Antler velvet from test negative animals in the herd is released from official control.

Recently, the United States' Center for Disease Control (CDC) issued a new statement concerning CWD and possible human infection: “Although it is generally prudent to avoid consuming food derived from any animal with evidence of a TSE, to date, there is no evidence that CWD has been transmitted or can be transmitted to humans under natural conditions”. However, the CDC has renewed surveillance efforts in order to rule out a link between CWD and vCJD. While to date there has been one case of vCJD reported in US (contracted in the UK), the CDC is working with ongoing investigations in Wyoming and Colorado to track cases of CJD or suspected CJD.

CONCLUSION

CWD is spreading and may have the potential to infect humans. It is not known whether CWD exists undetected outside North America. Its unique and troubling feature is that unlike scrapie and BSE, it occurs in both captive and wild ranging animals, which poses enigmas both for understanding the means by which it is transmitted from animal to animal, and for devising strategies to prevent its spread. When diagnosed in captive animals, herds can be culled or entirely destroyed, but this strategy cannot be used for animals in the wild.

Although CWD presents more of a problem to individuals (hunters, for example) than to general public health, individual infections could have public health consequences similar to those of vCJD: clinically healthy individuals harboring the infection during its incubation period could possibly transmit disease via cross-contamination of surgical instruments or blood donations, and after death from unsuspected disease, their bodies could be harvested for organ donations. Without the ability to establish a diagnosis of human CWD infection, or knowledge of the presence or absence of infectivity in peripheral body tissues and blood, the potential for human risk will continue to depend solely on epidemiological inference.

Another potentially dangerous situation would arise if CWD were to find its way into non-cervid animal species. In particular, if CWD were to be introduced and become endemic in livestock species such as sheep and cattle, the animal and human food chains could be put at the same kind of risk as what occurred with BSE. We know that sheep and cattle can be experimentally infected with CWD by intracerebral inoculation, and tests are ongoing to determine if oral dosing with CWD brain tissue, or close contact with CWD-infected deer, can transmit disease to cattle.

Although food chain infections would require a series of breakdowns in the system of precautionary measures already taken to prevent a BSE outbreak, including the banning of most mammalian protein for use in ruminant feed, the potential for human error is a real and unpredictable factor.

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