Chronic Wasting Disease

Chronic wasting disease (CWD) has been affecting North American wild ruminant populations for 50 years. It was detected in Korea in 2000 in a red deer (*Cervus elaphus*) and a sika deer (*Cervus nippon*) and currently may represent a serious challenge also for European wildlife, thus suggesting the European scientific community to make the point about the available knowledge on this disease. What is actually known about it? Are we ready to cope?

The disease

Chronic Wasting Disease (CWD) is a contagious neurological disease affecting white tailed deer (*Odocoileus virginianus*), Rocky Mountain elk (*Cervus elaphus nelsoni)* Shiras moose (*Alces alces shirasi*) and likely other subspecies of *Cervus elaphus*. Other cervid susceptibility at CWD is unknown. It causes a characteristic spongy degeneration of the brains of infected animals resulting in emaciation, abnormal behavior, loss of bodily functions and always fatal outcome.

CWD belongs to a group of diseases known as transmissible spongiform encephalopathies (TSEs). Within this group, there are several other variants that affect domestic animals: scrapie, which has been identified in domestic sheep and goats for more than 200 years, bovine spongiform encephalopathy (BSE) in cattle (also known as "mad cow disease"), and transmissible mink encephalopathy in farmed mink.

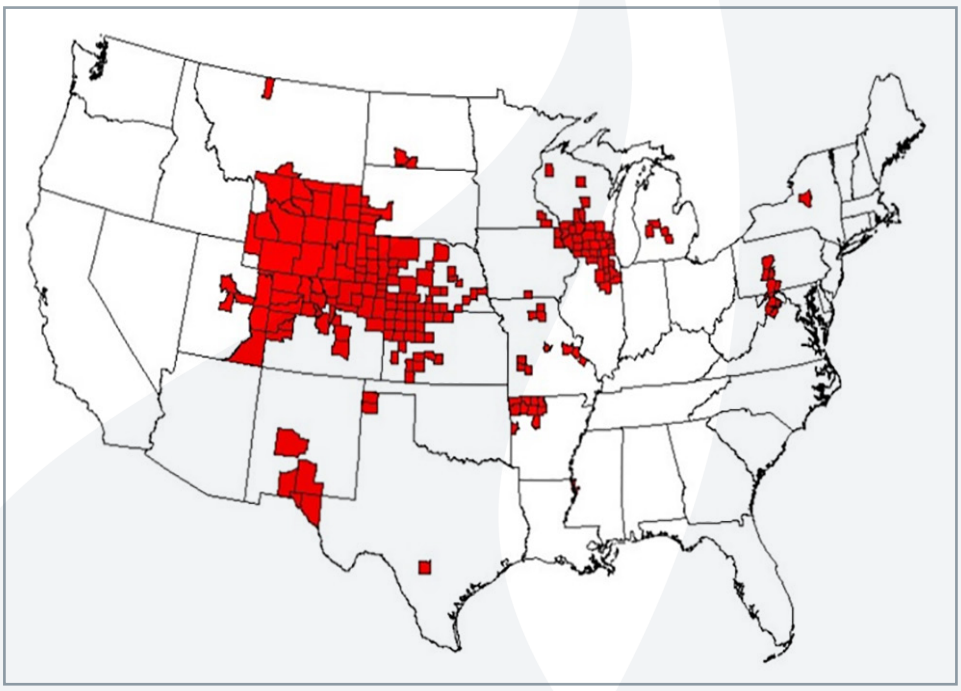
Several rare human diseases are also included in the TSEs. Creutzfeldt-Jakob disease (CJD) occurs naturally in about one out of every one million people worldwide. Variant Creutzfeldt-Jakob disease (v-CJD) has been associated with the large-scale outbreak of BSE in cattle herds in Great Britain. (http://cwd-info.org/faq/).

Distribution

As of March 27 2018, CWD in free-ranging deer, elk and/or moose has been reported in at least 23 states in the continental United States, as well as two provinces in Canada. In addition, CWD was detected in tundra reindeer (*Rangifer tarandus tarandus*) and moose (*Alces alces*) in Norway, and a small number of imported cases have been reported in South Korea. The disease has also been found in farmed deer and elk.

Nationwide, the overall occurrence of CWD in free-ranging deer and elk is relatively low. However, in several locations where the disease occurred, infection rates may exceed 10 percent (1 in 10), and localized infection rates of more than 25 percent (1 in 4) have been reported. The infection rates among some captive deer can be much higher, with a rate of 79% (nearly 4 in 5) reported from at least one captive herd.

Figure 1.
In March 2018, there were 215 counties in 23 states which reported CWD in free-ranging cervids.



This map is based on the best-available information from multiple sources, including state wildlife agencies and the United States Geological Survey (USGS).

Epidemiology

TSE agents are extremely resistant in the environment and because of it, the infection can be transmitted both directly and indirectly.

Modality of transmission still needs to be defined for certain aspects.

What we actually know is that in contrast to BSE, CWD is not a foodborne disease associated with rendered ruminant meat and bonemeal. Instead, observations of CWD among captive deer and elk provide strong evidence of lateral transmission, more similarly to scrapie.

Studies confirmed that prions are shed in the environment through body excretes and secretes, as saliva (Haley et al., 2011) (Mathianson et al., 2006), urine (Haley *et al.*, 2009) (Hamir *et al.*, 2006), feces (Tamguney *et al.*, 2009) (Tamguney *et al.*, 2010) and velvet (Angers *et al.*, 2009), but for now infection transmission has been only reported via saliva and blood (Mathiason *et al.*, in 2006). Mathiason and colleagues provided that under controlled indoor conditions CWD-naive deer can acquire infection by exposure to fomites from the environment of CWD-infected deer.

Concentrating deer and elk in captivity or by artificial feeding probably increases the likelihood of direct and indirect transmission between individuals. Contaminated pastures appear to have served as sources of infection in some CWD epidemics. The apparent persistence of PrPCWD in contaminated environments represents a significant obstacle to eradication of CWD from either farmed or free-ranging cervid populations.

Europe

LThe disease appeared for the first time in Europe in April 2016. A young reindeer specimen (*Rengifer tarandus*) was sighted by chance by a biologist who worked in the rocky mountains of Nordfjella.

Up to now, thanks to the surveillance implemented since 2016, 18 cases of CWD similar to the American type have been confirmed in Norway, and all of them come from living reindeer in the mountains of Nordfjella. While in other parts of Norway,

CWD has been ascertained in three other elks and a deer (*Cervus elaphus*). These four cases, however, differ from those of Nordfjella; they were older animals in which the disease is supposed to occur spontaneously and sporadically. The first Finnish case of CWD recognized in March 2018, in an European elk (*Alces alces*), is very similar to the latter cases. - See more at: https://www.vetinst.no/en/news/cwd-in-finland-is-different-from-the-nordfjella-cwd- type # sthash.6jSV9Yi3.dpuf

Clinical signs

Cases of CWD occur most commonly in adult animals, but also in yearlings. The disease is progressive and always fatal.

The most obvious and consistent clinical sign is weight loss over time. CWD affected animals continue to eat, but the amounts of feed consumed are reduced, leading to gradual loss of body condition. Excessive drinking and urination are common in the terminal stages.

Behavioral changes also occur in the majority of cases, including decreased interactions with other animals, listlessness, lowering of the head, blank facial expression and repetitive walking in set patterns. In elk, behavioral changes may also include hyper-excitability and nervousness. Excessive salivation, drooling and grinding of the teeth also are observed.

Clinical signs of CWD alone are not conclusive. There are other diseases with symptoms that mimic those of CWD. Currently there is no practical diagnostic test in live animals. The sole conclusive diagnosis is a post mortem examination of the brain, tonsils or lymph nodes.

A definitive diagnosis is based on examination of the brain to find the characteristic microscopic spongiform lesions and/or accumulation of the CWD associated prion protein in brain and lymphoid tissues using a immunohistochemistry technique. Gross lesions seen at necropsy reflect the clinical signs of CWD, primarily emaciation. Aspiration pneumonia, which may be the actual cause of death, also is a common finding in animals affected with CWD.

Researches ongoing to develop live-animal diagnostic tests for CWD. Early results indicate that a new live-test utilizing tissues from an animal's tonsils may be viable in deer, but so far has been ineffective in elk.

Risks for humans

To date, we do not know with certain if CWD can be transmitted to human as a consequence of the ingestion of meat infected with prions. Neither we have evidence that human cases of prion-like diseases increase between hunters and people who consume meat of animal hunted in endemic territories for CWD. The most interesting study, which is still ongoing, concern non-human primates. The study begun in 2009 by Canadian and German scientists(Brent Race et al. 2009) to evaluate whether CWD can be transmitted to macaque (*Macaca fascicularis*), a type of monkey that is genetically closer to human than any other animal that has been infected with CWD previously. At this point of the research they showed that after oral exposure, 2 squirrel monkeys (*Saimiri sciureus*) had PrPres in brain, spleen, and lymph nodes at 69 months post-infection. In contrast, cynomolgus macaques have not shown evidence of clinical disease as of 70 months post-infection. Thus, these 2 species differed in susceptibility to CWD. Because humans are evolutionarily closer to macaques than to squirrel monkeys, they may also be resistant to CWD. Because of the long time it takes before any symptom of disease appears, scientists expect the study to take many years before they will determine what the risk, if any, of CWD for people.

Studies made on the matter demonstrated that the barrier between cervids and humans is notable; however, prion diseases are dynamic and interspecies passage can result in prion adaptation to new host species.

As a consequence of these aspects to consume meat comes from an infected animal is discouraged. Good practices for hunters are widely explained on the CWD alliance website.

Eradication and control measures

No treatment is available for animals affected with CWD. Once clinical signs develop, CWD is invariably fatal. Similarly, no vaccine is available to prevent CWD infection in deer or elk. It follows that controlling CWD is problematic. Long incubation periods, subtle early clinical signs, absence of a reliable ante mortem diagnostic tests, extremely resistant infectious agent, possible environmental contamination, and incomplete understanding of transmission all constrain options for controlling or eradicating CWD.

One option for managing CWD in wild populations is to reduce the density of animals in the infected area to slow the transmission of the disease. This is made by selective culling of animals suspected to have been exposed to the disease. In Colorado, Nebraska, Wisconsin and Saskatchewan, efforts are underway to drastically reduce local wild cervid populations in an effort to eliminate CWD in areas where it recently was found.

The killing of an entire deer herd in a defined area where CWD was detected followed by two years quarantine was carried out in the region of Nordfjella.

Localized culling even its unpopularity actually is the most effective method to maintain low prevalence of the disease and to control it (Manjerovica *et al.*, 2013).

Sitography

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3. Norvegian Veterinary Institute

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