The Challenge of CWD: Insidious and Dire

Only immediate action will avoid catastrophic outcomes

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We have a problem. A big problem. Chronic Wasting Disease (CWD), a sister to BSE or ‘mad cow,’ is threatening our deer and elk. Unfortunately, CWD has broad implications. Without immediate action, we are heading for worst cases outcomes that include severe population impacts, extinctions, crashing economies, and, although unlikely, potential transfers of CWD to people.

Chronic Wasting Disease is an incurable, always fatal degeneration of the brain. Technically, it’s a Transmissible Spongiform Encephalopathy (TSE), but there are a number of quite different versions, depending on species. They include in humans kuru and fatal familial insomnia, as well as some with even more unpronounceable names, such as the dreadful human Creutzfeldt-Jakob Disease (CJD) and Gerstmann–Sträussler–Scheinker Disease (GSS). The largest TSE epidemics have been in domestic or captive animals: such as Scrapie in domestic sheep, Bovine Spongiform Encephalopathy (BSE), or so-called ‘mad cow’ disease, Transmissible Mink Encephalopathy (TME) on mink farms, and CWD in captive deer and elk.

CWD emerged as a particular nasty variant, because it can be transmitted by body fluids of infected animals (urine, feces, and saliva). Unlike BSE, CWD is highly contagious and can spread to and through wild ungulate herds. The infective agents are mis-folded proteins called prions; they are virtually indestructible, can persist in the environment, and tiny quantities can transmit the disease. Prion diseases have repeatedly jumped species barriers—most alarmingly in the United Kingdom, when BSE-infected beef killed 229 people.

As CWD spread, naturally and through trade, the U.S. in 2001 officially declared a “State of Emergency.” Every factor has since gotten worse. It has now been confirmed in 24 US states, 3 Canadian provinces, South Korea, and recently in Norway. Field studies are confirming potentially severe impacts on wildlife populations. So far no transmission to humans has been documented, but the risk is not zero. Non-human primates and transgenic (humanized) mice have been infected. In many jurisdictions, a lack of awareness and availability of free, rapid, and convenient testing of harvested deer has led to significant level of human exposure. Estimates show 7,000 to 15,000 CWD-infected animals are being consumed by hunter families every year, and this number continuing to rise by as much as 20% per year.

The combination of threats is sobering. CWD has been shown to persist and remain infectious in the environment, including in clay-based soils that can dramatically increase infectivity (up to 680 times). Decomposing carcasses create contaminated “super-sites.” Prions are extremely resilient, known to resist disinfectants, alcohol, formaldehyde, detergents, protein enzymes, desiccation, radiation, freezing, and incineration >1100°F. Facilities infected with CWD have resisted all efforts at removing the infective agent. Canadian officials report that even on premises thought to be very low risk, restocking with healthy animals led to a 50% re-occurrence of CWD.

Transmission occurs animal to animal, soil to animal, mother-to-offspring, and from exposed plants or other surfaces including tools or surgical instruments (even autoclaving is ineffective). Now there is evidence the infective agent is taken up via the root systems of plants growing in contaminated soils, with transfer to stems and leaves. These were shown to be infective via inter-cerebral injection (oral tests are ongoing).
Left unchecked, the prospects for wildlife are bleak. CWD has clear population impacts; some models suggest extinction. Disproportionate impact on mature males carries implications for hunters and wildlife economies let alone populations. Still more bad news: Efforts for vaccines have failed, and evolutionary or adaptive salvation is unlikely and would be too late in any case. CWD is now deemed to be the largest-ever mass of infectious prions in global history, and experts sum up the threat (to wildlife, agriculture, our economies, and potentially to human health) in two words: “insidious and dire.” Current policy and apathy toward the levels of CWD consumption by people has been described as “one of the most outrageous human susceptibility experiments in history.”

The good news

There is, of course, much more—but we need to get to the good news: There is hope, beginning with the fact that CWD is relatively new—not a long-standing or indigenous disease of our wildlife. The vast majority of our herds are still disease-free. We have considerable expertise, leading-edge technologies, and the benefit of experience. We faced a crisis on this scale once before, almost exactly a century ago, when the very existence of wildlife on this continent was threatened by the severest of over-exploitation. Hunters and conservation organizations led the efforts to avert disaster. With the courage and foresight of presidents and prime ministers enlisting the best and ablest on both sides of the US/Canada border to enact science-based policies, they turned our greatest tragedy into a ‘triumph of the commons.’ Anchored in the public trust doctrine, and now recognized as the North American Model of Wildlife Conservation, it replenished an entire continent with wildlife.

We need, today, nothing less than a similar effort to manage the Chronic Wasting Disease crisis. We have the benefit of experience and principles for success. Following the Roosevelt Doctrine, the same concerned hunter and conservation organizations must once again be the standard-bearers of principled, science-and evidence-based leadership in wildlife conservation. We must be relentless in following the leading science and scholarship, tracking the evidence, and engaging in comprehensive analysis to foresee the implications. We understand how policies affect the spread of diseases, as documented in the scientific and historical record summarized below. This threat is dire, and immediate action is warranted.

While details and methods must be guided by science and evidence, there is significant agreement on critical needs; and we have assurances from leading experts and labs that we have the capacity to meet this challenge. We must secure mandate and funding to:

1. Contain the geographic spread of CWD by enacting and enforcing an immediate ban on the movement of all live cervids, all potentially CWD-infected carcasses, animal parts, products, exposed equipment, trailers, or other sources of infectious materials.
2. Mandate and implement for hunters, convenient, cost-free, rapid testing of all animals harvested from CWD-affected areas.
3. Ensure that no CWD-infected material reaches the food or feed chains, and that it is instead properly disposed of.
4. Establish and fund accountable research and science-based policy to protect public interest (health, wildlife and related industries, agriculture, our economies and communities).

The issues are numerous, serious, and complex, but complacency is not an option. The sooner we act, the greater the prospects to protect our greatest living legacy. Further details, discussion, citations, and scientific references follow.
Origin vs Novelty

While we want to know the details of CWD’s origin, a more important question is whether it is a new disease, or has been around longer. Ascertaining whether a disease is newly emerged, or has long been present is key to establishing epidemiologic characteristics, threat profiles and critical measures to protect public interest. Diseases that have long been present in indigenous North American wildlife have typically been documented with evidence of scale, impacts, cycles, and relative risk patterns. On the other hand, newly emerged or introduced diseases—or new, significantly evolved versions of old pathogens—present uncertain risk to populations, to other species, to ecosystems, and to economies.

After CWD had been confirmed as a TSE in 1978, three generalized origin or novelty scenarios seemed plausible. First, though considered highly improbable by even the early 1990s, was a possibility that CWD was an unrecognized, rare, but longstanding disease of North American wildlife, persisting at very low levels, without obvious or serious impacts. Second, that CWD recently and spontaneously emerged in deer, similar to sporadic variant Creutzfeldt-Jakob Disease (CJD) in people, where the cause or trigger is unknown. Third, that it may have been recently introduced from a non-indigenous prion disease like scrapie that, through unknown means transferred from domestic sheep or goats to establish as CWD in deer.

By 2005, based on extensive documentation of presence (or absence), growth, spread, and persistence, it had become clear that CWD could not possibly be a longstanding disease of North American deer.

Nevertheless, given the profound role of this question in public policy, and given some lingering advocacy presuming the contrary, further explanation is warranted. As detailed below, extensive evidence has shown CWD to be highly contagious and laterally transferable between living animals. CWD prions have been shown to persist and remain highly infectious in soils, on plants or other surfaces. Once established, CWD exhibited a consistent pattern of growth, spread, and persistence. By 2001, CWD became a major threat to wildlife, livestock, and human health.

“Chronic Wasting Disease,” a sister disease to Bovine Spongiform Encephalopathy (BSE), also known as ‘mad cow’ disease, is a misfolded protein or ‘prion’ disease first observed and documented in captive mule deer in the late 1960s. It is in the group of diseases known as Transmissible Spongiform Encephalopathy (TSE) and has now been confirmed in at least six species of deer. TSEs have repeatedly emerged and all of the largest epidemics have been documented in domestic or captive animals. These include scrapie in domestic sheep, Bovine Spongiform Encephalopathy (BSE) in domestic cattle, Transmissible Mink Encephalopathy (TME) on mink farms, and CWD in captive deer and elk. CWD seems unique in having established reservoirs and significant prevalence in wild species.

The origin of CWD is not definitively known, and may never be solved. It most likely is a conversion from the one prion disease known to have direct contact with deer and which it most closely resembles: scrapie—the similarly contagious version in domestic sheep.

Leading experts Beth Williams, Tom Thorne, and Michael Miller postulated that “It is possible, though never proven, that deer came into contact with scrapie infected sheep either on shared pastures or in captivity somewhere along the front range of the Rocky Mountains, where high levels of sheep grazing occurred in the early 1900s. In addition, laboratory tests suggest that there is less of a species barrier to TSE transmission between deer, elk, and sheep, than between these and either cattle or humans.” This is further supported by evidence that deer are susceptible to scrapie.

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Fundamentals

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consistent with the observed growth in prevalence and the invariably fatal nature of the disease, indications pointed to serious population impacts. Three of the leading causes of North American wildlife. All known pathology and epidemiology, and all available evidence is consistent with CWD being newly emerged (or introduced) in or around the early 1960s.

A resilient, persistent, contagious pathogen

The protein pathogens or ‘prions’ causing CWD are extremely resilient, known to resist disinfectants, alcohol, formaldehyde, detergents, protein enzymes, desiccation, radiation, freezing, and virtual incineration >1100 °C. 14

That temperature, nearly the melting point of prion, was sufficient to completely ‘ash’ the tissue; weights were reduced by 98-99%. Yet “when reconstituted with saline to their original weights, prions transmitted disease to 5 of 35 inoculated hamsters.” 15 Normal sewage treatments do not degrade or inactivate prions, offering a potential means of transporting CWD prions to new areas with CWD, and to ensure appropriate disposal of all materials known to be positive. 50, 51

The fact that species barriers for prion diseases can sometimes be breached via intermediate species shows the need for caution because of the risk of transfer to new species. And this includes potential risk to people. Studies of “species that may act as reservoir … support a potential role for native rodents in the infection cycle.” 16

“Studies have also demonstrated that prion diseases can be orally transmitted to many species; i.e., CWD to voles, mice, and ferrets, scrapie to squirrel monkeys and hamsters, BSE to sheep, goats, cynomolgus macaques, and lemurs, and CJD and Kuru to squirrel monkeys, with some requiring prior in vivo or in vitro adaptation.” 17

Further, “once a prion strain has been adapted to a new host species, the prions from this new host species propagate more efficiently in a third host.” 18

Health authorities advise precautionary measures

An absence of evidence of CWD having transferred to people is partially due to the fact that it cannot happen, which why health authorities universally advise against consumption of any suspected prion material: “Animals testing positive for any prion disease should not be consumed by humans or other animals.” 19 No tissues from infected cervids should be considered prion-free. 20

In addition to brain, spinal column, and various lymph tissues, infectious CWD prions have been confirmed in saliva, urine, feces, blood, velvet antler, milk, as well as in skeletal and cardiac muscle, and fat 21 “demonstrating that humans consuming or handling meat from CWD-infected deer are at risk to prion exposure.” 22

In any areas known to be positive for CWD, “hunters are advised to avoid harvesting deer and elk that appear ill, to debone during processing, to wear latex or rubber gloves when dressing the carcass, and to avoid contact with brain, spinal cord, and lymphoid tissues.” 23 To prevent geographic transfer, hunters are advised against moving remains of harvested animals from areas with CWD, and to ensure appropriate disposal of all materials known to be positive. 50, 51

“You’ll have to be aggressive; remove all sources and all potential movement. Cut wider and deeper and wider; but you’ll get one chance. If CWD gets widely established, you’ll have it for a very long time.”

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Followign confirmation of CWD on a Saskatchewan game farm, asked what Canada should do if it spills over into public wildlife.

Transfer

Transmission of CWD has been shown to occur: animal to animal, soil to animal, plants to animal, soil to plants, etc. (IC, oral tests ongoing), and mother-to-offspring. In addition, human caused (atrogenic) transmission of the CJD agent has been reported in over 250 patients worldwide 24 including via surgical instruments that cannot be sterilized. And this has implications regarding any tissues, products, or tools infected with CWD. 25, 26

Density and stress in the ‘captive wildlife’ industry (game farms) have been shown to exacerbate CWD risks. 27, 28

In the Expert Scientific Panel traced Canada’s CWD to game farm animals imported from South Dakota. It then spread to wildlife, including mule deer, white tailed deer, elk, and moose.

As of this writing (and further explained below), five separate field studies undertaken in multiple regions document significant population impacts in mule deer, white tailed deer, and elk. Meanwhile, there is neither evidence nor theory to support CWD as a longstanding, indigenous disease of North American wildlife. All known pathology and epidemiology, and all available evidence is consistent with CWD being newly emerged (or introduced) in or around the early 1960s.

Failure to trace the origin (until 2000), allowed the source herd to ship animals to 40 game farms, infecting at least 21. 3

CWD has been shown to persist and remain infectious in the environment. 23 CWD prions adhere to minerals such as montmorillonite (Mte) in clay-based soils that can dramatically increase infectivity, up to 680 times. 29 Interestingly, recent studies indicate that the high binding capacity of Mte could potentially be utilized to remove prions suspended in liquids, offering potential means of prevention, treatment, or decontamination. 30

Ultimate duration of CWD persistence has not been determined, but an epidemiological investigation of scrapie reoccurrence in Iceland "established with near certainty that the disease had not been introduced from the outside and it is concluded that the agent may have persisted in the old sheep-house for at least 16 years.” 31

Experiments on other species show how CWD can circumpotent species barriers. 32 Many rodents, including the little golden hamsters, effectively resist CWD. However, if CWD passes first through ferrets, it infects golden hamsters after all. 33

CWD is characterized by very high prion replication in the brain, spinal cord, and various lymph tissues. And this includes potential risk to people. Studies of “species that may act as reservoir … support a potential role for native rodents in the infection cycle.” 33

“Studies have also demonstrated that prion diseases can be orally transmitted to many species; i.e., CWD to voles, mice, and ferrets, scrapie to squirrel monkeys and hamsters, BSE to sheep, goats, cynomolgus macaques, and lemurs, and CJD and Kuru to squirrel monkeys, with some requiring prior in vivo or in vitro adaptation.” 35

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one infected research facility, more than 90% of mule deer resident for >2 years died or were euthanized while suffering from CWD.63

As with other biological agents capable of exponential growth and spread, risk of transfer and introduction can be dire. The Expert Scientific Panel traced all of Canada’s CWD to imports of game farm animals from South Dakota—perhaps even in a single animal. The disease repeatedly spilled beyond game farm fences, to public wildlife.13

A State of Emergency

CWD was declared a “State of Emergency” by the U.S. Secretary of Agriculture Anne Veneman in 2001.44 Since that declaration, every factor (growth, spread, persistence, adaptation, exposure) of CWD has only increased and threat profiles continue to rise.65

To date CWD has been confirmed in some 24 states, 3 provinces (including retrospective finding in a mule deer in the Toronto Zoo in 1971), in South Korea (from 3 provinces (including retrospective finding in a mule deer resident for >2 years died or were euthanized while suffering from CWD.63)

Environmental reservoirs

In addition to readily transfers between live animals, “mule deer were infected by contact with skeletal remains of CWD-infected deer and surrounding ground and vegetation.”44

On the landscape, CWD-infected carcasses can funnel the prions from decomposing brains into soil, where it will adhere to various minerals, creating a contaminated “super-site.”50

Carcasses provide easy, nutritious food sources for a spectrum of animals; and decomposition releases nutrients into the surrounding soils, stimulating a substantial flush of forage in carcass sites. This could lead to human exposure to CWD.50

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Potential role of plants

Excerpts from Dr. Christopher Johnson, USGS:

“Vegetation is ubiquitous in CWD-contaminated environments and plants are known to absorb a variety of substances from soil, ranging from nutrients to contaminants. The uptake of proteins from soil into plants has been documented for many years and we have been investigating the uptake of prions into plants in vitro. Using laser scanning confocal microscopy, we observed root uptake of fluorescently-tagged, abnormal prion protein in the model plant Arabidopsis thaliana, as well as the crop plants alfalfa (Medicago sativa), barley (Hordeum vulgare) and tomato (Solanum lycopersicum).”70

With microscopic evidence of root uptake, transfer to stems and leaves of those plants as well as corn was confirmed using PMCA. The work further confirmed that: “Both stems and leaves of A. thaliana grown in culture media containing prions are infectious when injected into mice, and oral bioassays are underway for A. thaliana and other plants. Our results suggest that prions are taken up by plants and that contaminated plants may represent a previously unrecognized risk of human, domestic species and wildlife exposure to CWD and scrapie agents.”70

The amplifying role of captivity

While CWD contamination varies widely across natural areas, and can be observed spreading slowly after its emergence,71 and while it is affected by local population densities, prevalence, conditions and co-factors47—by far the highest levels of CWD infections, persistence,14 and geographic transfer47, 48 have been found in commercial game farms.47 In such premises CWD was found to have extreme prevalence, persistence, transfer/geographic spread, and potentially irreversible contamination.49 “Healthy cervids can become infected solely from environmental exposure. No environmental decontamination procedures currently exist for application to prion-contaminated premises.”49

In Saskatchewan, “on premises with no evidence of environmental contamination, after the quarantine was lifted, of those that chose to re-stock…that is, to continue cervid farming, there was an alarming 50% re-occurrence rate of CWD.”49

While not yet undertaken for CWD, an analysis of “the effectiveness of recommended scrapie farm decontamination regimens was evaluated by a sheep bioassay using buildings naturally contaminated with scrapie.”49 Four separated pens were assessed by cumulatively adding (+) decontamination measures: from a control where only gross debris was brushed out; (+) pressure washing; (+) treatment with sodium hypochlorite solution containing 20,000 ppm free chlorine for one hour; (+) removal and replacement of metalwork (or treated by re-galvanization), and painting every item of immovable equipment, natural environmental surfaces such as stones, pieces of wood and small animals that live in contact with the soil and environment (e.g., earthworms).”79

In captivity, high density, stress, squalor, artificial feeding and transportation fosters and spreads all manner of diseases. (Photo: Pat Davison)
The presence of dust may explain such persisting contamination and re-infection. Gough et al demonstrated that CWD prions remain infectious in the feces.108, 109 Including crows and coyotes have been studied, showing that CWD prions remain infectious in the feces.108, 109

It has been demonstrated that scrapie can be efficiently transmitted via the nasal route in sheep,60 as is also the case for CWD in both murine models and in white tailed deer.61

Additionally, "naive" deer exposed to water, feed buckets, and bedding used by CWD-infected deer contracted the disease.62 Given these and other findings of potential CWD contamination of facilities, tools, equipment, and various surfaces, and the potential infectivity in or on plants,63 the protection of public wildlife demands closely enforced restrictions of any materials moving from contaminated sites, and to restrict wildlife’s access to such.

As had been repeatedly warned by scientists,102, 103 the direct relevance and risks of commercial game farming to public interest have been documented repeatedly, in both the U.S. and Canada. These include substantial costs, demonstrated disease transfer, persisting threat of CWD transfer to public wildlife, with corresponding harm to economies, and risks to agriculture and human health. In April 2011, because of extreme, ~80% CWD prevalence and probable site and wind direction, the Wisconsin Department of Natural Resources was compelled to purchase, double fence, and monitor the highly-infected premises known as the Buckhorn Flats Deer Farm in Portage County, WI.104 In Canada, the Canadian Food Inspection Agency (CFIA) “maintains any imposed declaration of infected place and associated quarantine for premises wherein there is evidence of environmental transmission.” Twenty such premises remain under indefinite quarantine in Saskatchewan.105 ‘"requiring the CFIA to erect an immediate fence of perimeter fencing for the exclusion of wild cervids."'106

Direct exposure of public wildlife to CWD-infected premises, excreta, carcasses, and ‘super-sites’ provide substantial, high-concentration opportunities not just for interspecies transfer, but also for ‘quiet carriers’ and long-distance transport.107 Resistance notwithstanding, prion passage through predator and scavenger species, including crows and coyotes have been studied, showing that CWD prions remain infectious in the feces.108, 109

Fences do not protect wildlife

History confirms that it is challenging to keep deer (let alone diseases) from crossing into or out of fenced facilities: “Game ranches form a bridge for the transmission of livestock diseases between captive and wild populations.”110 A CWD-positive elk pen in Minnesota, USA, was found to have >20 breaches within the fence and wild white-tailed deer were observed within the facility.111 Furthermore, given the persistence in soil, water and plants, natural erosion and vector passage present formidable containment challenges. Indeed, as had long been predicted by scientists, the inevitability of disease transfer, emergence, and spillover from commercial game farms, and the corresponding costs and threats to public wildlife have been well documented.112, 113, 114, 115 This highlights pivotal role of governance and public policy in wildlife conservation—and in protecting the resources and the rights, privileges, opportunities, and benefits they sustain. Publicly elected representatives to consider carefully all aspects of our interactions, impacts, and endeavours, whether direct or indirect, immediate or long term, and to protect public interest above all else.

Analyses accurately predicted severe threats to wildlife

The Wyoming Analysis cited above is revealing. In the late 1980s, John Dorrance III applied to establish a large commercial game farm in Wyoming. The facility was to import and contain many exotic as well as domestic species. Widespread concerns expressed by wildlife scientists, hunters, ranchers, and the general public led the state government to consider alternative sites and methods. Various about the proposal and its implications. Released in 1990, the Wyoming Analysis stands today as the most comprehensive investigation of game farming ever undertaken by a government. The state declined Mr. Dorrance’s application. The issues, science, evidence, and views of experts outlined in Wyoming’s Analysis subsequently withstood repeated court challenges. This is also included time-tested testimony after the 2000 election when Montana voters passed ballot initiative 143. In a written affidavit, after observing seven years of repeated epidemics and myriad problems with game farms across the continent, the following quote summarized Wyoming Game and Fish Department’s view of the analysis and decision: ‘"The issues raised in our report are as valid today as they were in 1990. The issues of game farming transport animals escaping into the wild, the spread of disease via escapes or movement of animals across jurisdictional boundaries in commerce, competition between native and exotic wildlife, the potential for hybridization and genetic pollution, the possibility of theft of public wildlife and an increase in poaching activity as a result of putting a monetary value on dead wildlife, and the damage penned shoots would have on the public's perception of sport hunting as a legitimate tool of wildlife management were issues that we thought made game farming an unacceptable risk to Wyoming's wildlife treasure. Events since 1990 have confirmed that the issues we raised in 1990 were real and reaffirmed the wisdom of the Commission’s decision to completely deny the applications."’116

"I declare under penalty of perjury that the foregoing statements are true and correct to the best of my knowledge." —Robert P. Lanka, Wildlife Management Coordinator, Wyoming Game and Fish Department June 28, 2001

Indeed, as part of the original CWD enzootic zone, Wyoming faced significant challenges that could only have been exacerbated by widespread commercial exploitation. The forewarnings detailed in their analysis emerged repeatedly. For example, just prior to and coincident with the acceleration of North America’s CWD epidemic in 1996, an epidemic of tuberculosis on North American game farms in the early 1990s spread to cattle, bison, pigs, and people.117 Investigation revealed that inadequate testing and false presumptions of safety enabled repeated outbreaks. Evidence suggests that Tb and CWD may have been transported into Canada simultaneously, in the same animals.118, 119

In 1989, Eileen Wellicome, Pulitzer Prize-winning journalist for The Albuquerque Tribune, authored a six-part series on game ranching documenting serious problems and warnings from scientists.126 Canadian officials found that the epidemiological tracing of outbreaks in captive animals was compromised, because farm records were often in error. Irregularities and illegal transport were widespread. Additionally, “over 120 elk from Tb infected game ranches in Alberta and Montana alone were known to have escaped or were inadvertently released into the wild.”121 CWD status of those animals remains unknown.

Further, as New Zealand’s National Tb Advisor had warned, cervids can become grossly infected and infectious with ovine Tb,122 explaining the unusual finding in Alberta where some 42 people (game farmers, veterinary technicians, and abattoir and rendering plant workers handling elk carcasses) tested (nearly) positive for Tb.123 Large numbers of animals were missing from infected and quarantined farms, causing “Alberta’s Director of Wildlife to issue an official warning to Alberta hunters that if they happen to shoot an animal with an ear tag, don’t even touch it.”124 “The (game farm Tb) outbreak not only cost taxpayers tens of millions for indemnification, staff, and administration, it cost all of Canada Tb-free status, valued by Agriculture Canada at $1 Billion.”125

Wisconsin deer farm with severe (80%) prevalence of CWD. The DNR purchased and double fenced it to prevent wild deer access. 20 such CWD farms in Canada are under permanent quarantine at taxpayer expense.
Principles and opportunities ignored

Early in Canada’s Tb epidemic on game farms, there were opportunities to test for CWD in depopulated Tb infected herds. This led to formal requests to secure such tests, as such were not possible prior to importation of large numbers of captive animals from the U.S. The Tb epidemic provided authorities with vital access to “animals already under the control of Agriculture Canada, they were already dead, already paid for, and there had been many suspicious or inexplicable deaths on game farms. Although the request was directed to Agriculture Canada, the Alberta government dismissed the need, and declined the request.135 The possibility of finding CWD early would almost certainly have prevented the massive spread of CWD in Canada, and may well have had similar benefits for the U.S. (outside of the CWD-endemic area).136

The disease epidemics highlight the role of the precautionary principle, and of comprehensive analysis in forming public policy. Without such consideration, public wildlife, public interest, and traditional agriculture are jeopardized. Yet despite its unique and direct threat, the “captive wildlife” industry has been all but immune from any accountability, or the “polluter pay” principle.

Unfortunately, the modelled predictions about CWD impacting the population are now being borne out in the field. In the last two years, a series of published and ongoing studies have confirmed significant and potentially severe population impacts for deer,137 138 139 which carry substantial population declines.133

ELK: Study of an endemic elk herd (prevalence 12.9%), in Rocky Mountain National Park showed that “CWD alone is capable of causing large declines in elk populations.”134 “CWD-caused mortality can exceed natural rates of mortality, reduce survival of adult females, and decrease population growth of elk herds.”134 Preventative efforts to minimize the risk of CWD into new geographic areas are therefore critical.

Early in Canada’s Tb epidemic on game farms, there were opportunities to test for CWD in depopulated Tb infected herds. This led to formal requests to secure such tests, as such were not possible prior to importation of large numbers of captive animals from the U.S. The Tb epidemic provided authorities with vital access to “animals already under the control of Agriculture Canada, they were already dead, already paid for, and there had been many suspicious or inexplicable deaths on game farms. Although the request was directed to Agriculture Canada, the Alberta government dismissed the need, and declined the request.135 The possibility of finding CWD early would almost certainly have prevented the massive spread of CWD in Canada, and may well have had similar benefits for the U.S. (outside of the CWD-endemic area).136

The disease epidemics highlight the role of the precautionary principle, and of comprehensive analysis in forming public policy. Without such consideration, public wildlife, public interest, and traditional agriculture are jeopardized. Yet despite its unique and direct threat, the “captive wildlife” industry has been all but immune from any accountability, or the “polluter pay” principle.

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Using advance testing and radio-collar monitoring, a study of 143 mule deer in the LaPrele Reservoir in southern Converse County, Wyoming showed that CWD-positive deer were 4.5 times more likely to die annually than CWD-negative deer.123 These results support concerns of wildlife managers, wildlife disease experts, and conservationists that this endemic (chronic) disease can diminish the sustainability of deer population at high disease prevalence.134

Activity analysis of deer in the Wyoming white-tailed deer study “suggested CWD-positive bucks did not participate in the rut at the same level as CWD-negative bucks,” and that “CWD positive bucks were less aware of the rut and the hunting season and were more susceptible to being shot by a hunter.”138 Further, “[o]ver-representation of CWD-positive deer in the hunter harvest suggests behavior is altered by CWD prior to clinically recognizable CWD infection. Rather than thinking of CWD as a strictly pre-clinical disease followed by a short, obvious clinical stage of disease, we believe CWD infection should be envisioned as a slow, progressive decline in health and alteration of normal behavior, which ends with clinically recognizable disease.”139

Potential for natural adaptation

Evolutionary adaptation based on genotypic resistant alleles remains questionable, and would likely be too late in any case. “Wild cervid populations are unlikely to evolve quickly enough for selection to influence disease management.”140 Even if hunting were stopped to accelerate the selection process through higher CWD prevalence—a prospect implying serious risks on many fronts—  achieving the hypothetical resistance could take some 50 years.141 Moreover, this assumes resistant genotypes have similar fitness as susceptible genotypes. In the interim, deer populations are likely to experience substantial population declines.142 Where hunting or other significant predation lowers prevalence, this timeline is extended beyond 200 years. But the health and suitability prospects remain suspect.

CWD resistant genotypes are weakly conserved, and attempts to breed deer with the resistant genotype have been unsuccessful. As in a 2016 CWD symposium Dr. Michael Miller said: “We actually tried breeding, for research purposes, 225 FF mule deer, tried for quite a few years, and there was something really wrong with them. It wasn’t that they didn’t look like mule deer, but they just weren’t quite right.”143 Dr. Tracy Nichols added: “…mother nature doesn’t like them. Ok, so in the wild, there aren’t very many, and there’s probably a very good reason for that. Are they immune-compromised, are they more susceptible to, perhaps, certain parasites or certain infectious diseases?”144

Prospects for vaccines

Potential development of vaccines for prion diseases are extremely difficult for a number of reasons, beginning with the challenge of initiating an immune response. Also, vaccinations are not without safety concerns, potential side effects, issues in other species, prospects of ameliorating the disease, and, in wildlife, significant delivery and monitoring challenges. The good news is that there have been some positive indications of an induced immune response to prion proteins. In a direct approach that targeted epitopes exposed by misfolding, a protective antibody response was shown in vitro. The group of authors note, however, that caution is advised on several fronts:

•“PrPc-reactive antibodies could have pathological consequences in otherwise healthy animals.” … “While any vaccine, with a therapeutic benefit is undoubtedly a scientific success, the use of a prion vaccine in wildlife populations will likely need to consider the mechanisms of protection, in particular as they relate to safety.”

•“Securing acceptance by regulatory authorities for wildlife vaccines where there is less opportunity to oversee, monitor, and regulate vaccinations.” 146

A partial therapeutic protective response to CWD was demonstrated for the first time in vivo in a Salmonella-delivering vaccine in white-tailed deer, including in a genotype with full sensitivity to CWD.147 One may note that this approach may allow oral delivery of the vaccine using food pellets, or some slow-release preparations to stimulate the entire gastrointestinal tract.

Additional challenges and concerns regarding vaccines remain on many fronts, in both targeted and additional species. Unfortunately, while not on the scale of evolving resistance shown with antibiotics, evidence shows that neither bacterial nor viral vaccines are evolution-proof. “Vaccines have the potential to change the spectrum of pathogens seen in several infectious diseases.”148 There is no obvious reason to expect that protein-targeted vaccines could not be vulnerable to similar effects. But neither that, nor the unfortunate result in a recent CWD vaccine trial in Wyoming that caused 7 times more disease, with faster onset,149 represents the greatest risk. The greatest risks are further delays in reassessing fundamental public policy. This also raises questions regarding the purpose and the beneficiaries of CWD vaccines. Even best case scenarios suggest that breakthrough disease management tools for wildlife would be decades away.

To be clear, given the importance and scale of the experimental opportunity, and potential zoonotic risk, CWD vaccine research should continue—but only (and cautiously) for potential application for human health, to assist in CWD control in wildlife (reducing prevalence and spreading), or in restoring wild populations in areas with potential environmental contamination.

Potential Risk of Transfer to People

There are few considerations which require greater foundational context than questions of zoonotic risk of infectious diseases, as the scientific debate and the potential public concern is very much a matter of addressing uncertain risks that are dynamic, evolving, and with complex, even profound consequences.

The reality is that most (~70%) emerging zoonotic diseases have come from animals.150 151 Each presented

Impacts and threats by species

MULE DEER: Using advanced testing and radio-collar monitoring, a study of 143 mule deer in the LaPrele Reservoir in southern Converse County, Wyoming showed “an annual population decline of 19%,” contrasted with “a stable population growth rate under CWD-free conditions.”134 The results suggest potential extinction of that mule deer population within 41 years.135

WHITE-TAILED DEER: A study of WT deer in east-central Wisconsin showed “substantial population-limiting and the strong population-level effects of CWD suggest affected populations are not sustainable at high disease prevalence under current harvest levels.”136

9
The Public Trust Doctrine
That government’s primary responsibility is to assess such risk is the essence of the Public Trust Doctrine, the contractual relationship at the core of all governance. It embodies the components, relationships, and legal obligations of a classic fiduciary trust. Components include:

- Trustees (governing representatives in various levels and branches: elected, executive, and the courts)
- Beneficiaries (constituents and future generations)
- Public interest related to property (tangible and intangible) and wellbeing (life and liberty)

Trustees accept and bear a burden of responsibility to protect and defend the interests of their constituents, and those of future generations. Where so-called hard sciences probe the vital questions of ‘what is’ and ‘what was,’ governance, or political science, must build from that foundation to confront the equally challenging questions of ‘what if.’

While often hidden by retail politics, the weight of this responsibility is profound. Fortunately, guidance has been further established in precautionary, accountability, and pollutor-pay principles that have saved untold billions of lives and immeasurable costs.

The Precautionary Principle
Where there is a potential for severe or irreversible harm, especially to public wellbeing and interest, an absence of scientific consensus or proof of harm cannot be used to delay or maintain policies or actions under the risk. In such cases, the burden to ‘prove safety’ falls on those advocating the potentially harmful policy or action.10

The standard of “severe or irreversible harm” is a very high bar; yet one CWD has long surpassed regarding public wildlife. It is only against that backdrop that the potential transfer of CWD to people can be reasonably considered. We must consider risk, consequences, and even worst case scenarios. The fact is that prion diseases are described by physicians and victim’s families as aggressive, horrific, and dreadful.

Faced, in his medical practice, with the reality of human prion and neurodegenerative diseases, a leading scientist Dr. Neil Cashman, former Scientific Director, PrionNet Canada, have long warned that CWD is “an emergency in slow motion.” At the “On The Horizon” PriNet Research Conference Dr. Cashman summarized the background and urgency as follows:

“CWD is spreading like wildfire. From a few foci in Saskatchewan, it has now come to involve deer and elk in Alberta and Saskatchewan and there are no geographical barriers. It will spread until it infects the entire continent. It also spreads across species… it can persist in soil; it’s spreading without check. It’s arguably the most contagious prion disease, and the human health impact is unknown. We just frankly do not know if humans are susceptible to chronic wasting disease. It’s an emergency in slow motion.”110

Assessing zoonotic risk of new, emerging, and especially fatal diseases, is challenged by the inability to experimentally test susceptibility in people. Indeed, the ethical challenges are formidable enough regarding potential treatments. Yet questions of susceptibility require new approaches combining epidemiological and laboratory analyses (both in vitro and in vivo), as well as considerations of known and probable human exposure. Questions of appropriate policy and regulatory responses must be weighed against all implied consequences (biological, social, and economic), and the entire range of outcomes. This must include potential worst case scenarios even if they are thought extremely unlikely, not just because of evolving risk, but because market, media, and societal responses are often based more on perception than on science or reality.

Epidemiological analyses
Epidemiological research regarding potential transfer of CWD to people has examined potential emergence as a zoonotic disease. It’s an emergency in slow motion.”

In vitro laboratory analyses
While no longer in its infancy, the science of prion, or protein-only pathogenicity, is still relatively new. Indeed, many considered it all but heretical, long after Prusiner, an American doctor who was awarded the Nobel Prize in 1997 for the theory of ‘prion,’ or self-replicating, protein-only etiology. Against the backdrop that protein folding, and the multitude of complex biological interactions (covariant / hydrophobic effects), it stands among the great remaining challenges in biology.

Prions multiply by protein fold-conversion—re-templating natively folded PrP( cellular protein) to amyloid forming, protease-resistant PrP( disease prion). While analytical approaches differ from the genetic replication and synthesis typical of bacterial/viral/fungal pathogens, prion self-propagation offers the unique opportunity for cell-free analyses. These studies offer valuable insights into the possibility and potential rates of cellular prP( interaction of prion and neurodegenerative disease, a leading scientists like Dr. Neil Cashman, former Scientific Director, PrionNet Canada, have long warned that CWD is “an emergency in slow motion.” At the “On The Horizon” PriNet Research Conference Dr. Cashman summarized the background and urgency as follows:

“The lack of evidence of a link between CWD transmission and unusual cases of CJD (i.e., vCJD), 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.”12

Still, the authors advise caution based on other indicators and limited exposure:

“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.”162

Zoonotic risks are neither static nor merely historic phenomena. “It is estimated that approximately 75 per cent of ‘new’ human pathogens reported in the past 25 years have originated in animals and the risk of zoonoses is predicted to continue to increase.110 As status quo matters of public policy they require consideration of known and potential consequences. “The Global Burden of Disease Study estimates that, in the year 2000, infectious diseases were responsible for 22% of all deaths and 27% of disability-adjusted life years worldwide.”110

Such risk profiles can only be considered as snapshots in dynamic, evolving landscapes, where observation and evidence of variability—indicating change or evolution—is a vital consideration. This underscores the very essence of the precautionary principle, and nowhere is it more requisite than with respect to infectious pathogens. Inadequate policy or regulatory failures can result in pandemics that kill thousands or even millions of people or other animals, causing enormous damage on economies and ecosystems.

Dr. Delwyn Keene at the Wisconsin Veterinary Diagnostic Laboratory examines deer samples. Pink areas signal CWD. Testing is vital to avoid infected deer from being consumed.
Early cell-free conversion experiments showed that CWD barriers that limit CWD transmission to humans. Collectively, these results help define the structural and S170 are significant inhibitors of CWD conversion, the maintained by the human-specific amino acids within the 2-α maintained by the human-specific amino acids within the... It also suggests that CWD prions have a dominant potential and raise new questions about the possible link between animal and human prions. In repeated studies, transgenic mice expressing human prion protein seemed to resist CWD infection. However, "a recent bioassay with a natural elk CWD isolate in a 167... transgenic mouse line led to clinical prion infection in 2 out of 5 mice (3 out of 5 if an infected animal that died is included)." These results indicate that CWD prion has the potential to infect human CNS or peripheral lymphoid tissues and that there might be asymptomatic human carriers of CWD infection. This follows the 2014 research demonstrating transmission of scrapie to transgenic humanized mice: "The serial transmission of different scrapie isolates in these mice led to the propagation of prions that are phenotypically identical to those causing sporadic CJD (sCJD) in humans. These results demonstrate that scrapie prions have a zoonotic potential and raise new questions about the possible link between animal and human prions."

Questions remain, however, as even more recent in vitro research (yet to be published) suggest potential for significant CWD adaptation and thus greater risk. This work showed that "CWD adapts to a new host more readily than BSE and that human prP was unexpectedly prone to misfolding by CWD prions." The analysis further determined that "human prP has a region that confers unusual susceptibility to conversion by CWD prions." Most concerning, where "BSE prions are essentially unaltered by infection in 2 out of 5 mice (3 out of 5 if an infected animal that died is included)." These results indicate that CWD prion has the potential to infect human CNS or peripheral lymphoid tissues and that there might be asymptomatic human carriers of CWD infection. This follows the 2014 research demonstrating transmission of scrapie to transgenic humanized mice: "The serial transmission of different scrapie isolates in these mice led to the propagation of prions that are phenotypically identical to those causing sporadic CJD (sCJD) in humans. These results demonstrate that scrapie prions have a zoonotic potential and raise new questions about the possible link between animal and human prions."

**Human CWD exposure, prion load, and threshold dose**

Despite universal warnings from health authorities advising against consumption of any infected prion products, levels will transfer, from the challenges of species barriers, to a transfer between cohorts.

**In vivo (animal) experiments**

Results of CWD laboratory challenges of non-human primates are mixed. CWD transferred readily to squirrel monkeys orally (92%), but macaques, which are genetically closer to humans than squirrel monkeys, have demonstrated significant resistance, even to direct intracerebral injection. Yet another recent study noted, however, that recently macaques were shown to be susceptible to scrapie, but only after an extended, silent incubation of ten years.

**Squirrel monkeys, susceptible to CWD**

**Macaques, robust species barrier**

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**Experts weigh in**

Given their own and the risk analyses of others, leading scientists are expressing concern.

"The increasing levels of CWD exposure are highly concerning." As a matter of policy, I believe all animals taken from CWD-infected areas should be tested before consumption and people should definitely not be consuming any infected material."

"The situation and increasing levels of CWD exposure is a concern for cervid and human public health. "CWD-testing should be conducted on animals harvested from CWD-infected areas prior to consumption."

"... The more opportunity to expose humans to this stuff, the more we're potentially playing with fire, in terms of these strain adaptations. It wouldn't take very many cases of human prion disease that were linked back to chronic wasting disease, to where this whole conversation could change, fairly dramatically, and pretty much overnight. So I think while we have the opportunity, to get out in front of this ... where we can as best we can, we should probably take advantage of that."

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**The full spectrum**

The scope of human exposure to CWD is broader than is generally appreciated. It includes some direct exposures that have been largely ignored, lessons from history notwithstanding. When early suspicions of BSE being spread through consumption of blood, bone, and nerve... were confirmed in 1988, it led to bans on feeding meat and bone meal (MBM) supplements. Yet the potential risk of CWD in velvet antlers (i.e., blood, bone, and nerve tissue) sold for human consumption continued to be ignored long after the confirmation of... cases of human CWD exposure have been steadily increasing. In Wisconsin alone, this number was over 4000 such carcasses... number was surpassed (219 positives) in 2010 with only 6,853 tests. By 2014, only 3,665 tests were conducted, but showed 327 positives.

As the prevalence rate for CWD increases, the number of undetected CWD positive deer carcasses entering the food chain is growing exponentially. By one estimate, in Wisconsin alone, this number was over 4000 such carcasses for 2015 alone. At the current rate of increase, the number of undetected carcasses being consumed will double every three years. Yet testing continues to decline.

While cutbacks in testing have compromised accuracy, North America wide, some 7,000—15,000 CWD-infected animals are now being consumed by hunter families and friends every year.

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In 2009, the University of Kentucky proved that CWD can be passed through velvet antler. (Photo: Pat Davison)
With its growth and spread, human exposure from all sources has been increasing exponentially, and we would do well to consider the implications of both known and unknown factors. For example, prion load has been shown to be relevant, but note the title of McLean and Fryer’s 2011 work “There is No Safe Dose of Prions.” Analysis of 4,338 mice showed “that infection is possible at the very low dose of a 1,000-fold dilution of the dose that infects half the challenged animals (ID50).”

After pointing out that bank voles (which are circumpolar) are described as the ‘universal acceptor for prions,’ Sigurdson asks if the sequence in the human β2-a2 loop creates a permissive host PrPf sequence that is converted by prions from other species, despite sequence mismatches. With multiple avenues of direct and indirect human exposure, potential bioaccumulation, the potential role of co-factors, stressors, and potentially consequential passage to or through intermediate species, caution remains prudent. Moreover, it’s becoming clear that our understanding will be well served by looking beyond mammals. Quite apart from the work documenting mammalian prions taken up or adhering to plants, Susan Lindquist’s team has recently shown the ‘first protein from the plant kingdom with bona fide prion attributes.’

**Science, our greatest ally**

These analyses outline more than risk: they offer hope. Lindquist has long been at the front of breakthrough prion research with yeast, and has not only documented many collaborative interactions, but key evolutionary and epigenetic analyses to help explain the phenotypic benefits that have conserved prion existence for 800 million years. These and other insights into prion function may well open opportunities to prevent, limit, or potentially even reverse prion disease. However hopeful those breakthroughs might be, they are distant, and the levels of human exposure to CWD are already into the UK’s range of 1,000—10,000 BSE-infected carcasses sufficient to result in BSE transferring to a person. North American hunter families are consuming some 7,000—15,000 CWD-infected animals per year, and the number is growing exponentially. Though deer are much smaller in mass than cattle, this is more than offset by the fact that CWD prions are spread far more broadly and are highly contagious in deer. It is not inconceivable that a possible transfer to people could result in similar prion shedding in urine, feces, and saliva. Such an occurrence is beyond any known, practical means of containment or treatment, or avenues to curtail the economic fallout.

Overall, the zoonotic risk profile of CWD is complex, uncertain and evolving. Without exception, dozens of experts consulted for this work concur with the current Director, Prion Diseases Program for the Public Health Agency of Canada, Dr. Michael Coulthart, who describes the risk of CWD transferring to people as “far from negligible.”

**Implications are broad, deep, and longterm**

What is clear to policy analysts is that even a single transfer of CWD to a person will carry catastrophic implications in reactions of the public, in markets, and in public policy and international trade, regardless of how the disease manifests. And we cannot ignore the reality that CWD is highly contagious in deer.

If less dramatic, the risk profile is broad, complex, and with potentially dire outcomes at every turn. As to one

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**Why worry?**

As evident from challenges in achieving a CWD vaccine for cervids, there is little hope that breakthrough treatments would soon emerge. CWD has been in the shadows; but the toll of other protein misfolding diseases in people (Alzheimer’s, Parkinson’s, Huntington’s, ALS, Creutzfeldt-Jakob, etc.) affects tens of millions of Americans and costs hundreds of billions per year. Yet the scale of complexity and level of difficulty is such, that, even after decades of research, there are no cures, and few effective treatments. For example, prion load has been shown to be relevant, but note the title of McLean and Fryer’s 2011 work “There is No Safe Dose of Prions.” Analysis of 4,338 mice showed “that infection is possible at the very low dose of a 1,000-fold dilution of the dose that infects half the challenged animals (ID50).”

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If less dramatic, the risk profile is broad, complex, and with potentially dire outcomes at every turn. As to one
minor example: consider the risk of CWD transfer through pet food. That was included in the UK's updated, March 2016 Qualitative Risk Assessment regarding chronic wasting disease being introduced into Great Britain. The assessment asks: What is the risk of CWD being introduced into Great Britain (GB) from North America and causing infection in deer?

The analysis focuses on three routes of potential CWD introduction:

1. importation of animal feed
2. importation of deer urine lures
3. importation of CWD prion on contaminated equipment and clothing/footwear of hunters or other tourists and British servicemen

The assessment cites the European Union Trade Control and Expert System (TRACES), which confirmed that “in November and December 2015, for example, GB imported 13.6112 tonnes of processed cat and dog food (including dog chews) containing products of ungulate origin from Canada and USA.”

An updated Qualitative Risk Assessment from the British press, the cat died from Feline Spongiform Encephalopathy (FSE), a toll eventually reaching 89 domestic cats in UK, one in Northern Ireland, one in Norway, one in Switzerland, and one in Liechtenstein.

Virtually all species of large cats in zoos were similarly infected, including: five cheetahs, three pumas, three ocelots, three tigers, five lions, and one Asian Leopard Cat. All were instances of simple oral ingestion of BSE-contaminated feed. The news of transfer across species barriers immediately rocked public confidence and affected markets, which were damaged further with the subsequent admission that people were dying of vCJD from consuming infected beef.

The UK assessment outlines a similarly “greater than negligible risk” of importation of CWD prion on contaminated equipment and clothing/footwear of hunters, and that “the annual risk of at least one infection of deer in the UK with CWD from deer urine lures imported from the USA is medium.” But whereas official assessments of CWD threats to European wildlife have been measured, largely unseen, and potentially understated, concerns raised by NGOs and the media have been blunt, as in an article in The Times headlined: “Disease from the U.S. could wipe out all the deer in Britain.”

Deer dead from CWD. Excessive salivation and thirst drives diseased animals to riparian areas. Prions persist in soil and water, presenting risk of further transfer. (Photo: Wisconsin DNR)

Articles from the UK, warning of the risk of CWD to their wildlife and landscapes.

Lessons from BSE

From a public policy perspective, the experience with BSE offers vital lessons—from the foundational frame to the conclusions and recommendations by the official inquiry (paraphrased for brevity):

- “At the heart of the BSE story lie questions of how to handle hazards, of how to manage an unknown hazard to cattle and an unknown hazard to humans.”
- “BSE developed into an epidemic as a consequence of intensive farming practice(s) … unchallenged over decades, (that) proved a recipe for disaster.”
- “Government was preoccupied with preventing an alarmist over-reaction … (they) believed that the risk was remote. It is now clear that this campaign of reassurance was a mistake.”
- “Public was repeatedly reassured that it was safe to eat beef.”
- “Repeated statements that ‘there is no evidence that BSE is transmissible to humans’ does not explain that such evidence would take many years to emerge.”
- “Even when risk to humans seems remote, all reasonable precautions must be taken.”
- “There should be more checks on possible pathways of transmission, and on occupational risks.”
- “Where there is uncertainty, government must not shrink from saying ‘we are not sure’.”

The analyses and recommendations of the UK experience with BSE are founded on failures of governments to
impacts may vary across areas, ecosystems, species, and
direct impacts on human-wildlife interactions. While CWD
harm will not be known for decades, but it will have
wildlife are overwhelming. The full extent of the ecological
Impacts on wildlife economies
governments to contain and limit the spread, growth,
uphold public trust and the precautionary principle.
It has come to my attention that during the legislative session the General
omnibus agriculture bills (House Bill 1326 and Senate Bill 506) that attempt to redefine the ter
"livestock" in Missouri statutes to include captive deer and put them into the same classification
Missouri Constitution gives the Conservation Commission authority over the management and
I understand that Governor Nixon vetoed the Bill from a constitutional perspective as the
squirrels.
Hunting, fishing and outdoor recreation have a significant positive impact on the state's economy
results in over $95 million dollars in state and local tax revenue each year
important family heritage for many generations of Missouri families.
I am very much opposed to the override of the Governor's veto.
We can't take that chance.

Market-based threats
While less obvious, CWD presents a threat to North America's livestock and agri-food economy,216 of which exports represent about
$150 billion per year.220 Where our previous descriptions focused on science in the transfer of the disease, regulatory responses and economic drivers operate on substantially different criteria. Markets are often
affected as much by perceptions as by science. The growth capacity of infectious diseases looms both real and
persuasive. As a result, international trade is routinely restricted as a precaution against potential transfer that
can harm people, industry, markets or public interest. One needs to note that this includes threats to wildlife or the
environment, and that reactive trade restrictions based on perceived threats can be initiated in a matter of days or even hours.221
Prior threats are comparatively rare, but the uncertainty (lack of adequate or live testing) coupled with extreme
resilience of prions and the invariably fatal outcome of CWD makes it comparatively rare, but the uncertainty (lack of adequate or live testing) coupled with extreme
resilience of prions and the invariably fatal outcome of CWD makes it
perceived threats can be initiated in a matter of days or even hours.221
At every turn, the sheepdog/pigpen quandary is significant—a perception is not just a perception and
and

Legal basis of perception over science
The substantial role of perception in both markets and public policy has been well documented in issues such as GMO food labeling.226 Specific to TSEs, the interaction of science, markets, perception, and public policy was examined
in the transfer of disease, but tariffs and other regulatory changes centered on the vital role, impact, and

Impacts on wildlife economies
The evidence regarding the presence, growth, spread, persistence, evolution/adaptation, and impacts of CWD on wildlife is overwhelming. The full extent of the ecological

Letter of concern regarding CWD from Johnny Morris, Founder/CEO, Bass Pro Shops
uphold public trust and the precautionary principle. The cases are directly applicable to CWD: Without immediate, science-based intervention by North American governments to contain and limit the spread, growth,
evolution, and exposure of CWD, the likelihood of 'worst-case' outcomes will continue to increase, and wildlife is in the
cross hairs in every scenario.

(1996 to 2006). It had devastating impacts on agricultural communities.
Canada's May 2003 confirmation of a single case of BSE in
Alberta initiated an immediate ban on exports of Canadian
beef. By the end of 2004, financial losses for Canadian beef producers as a result of BSE reached $5.3 billion.223

perform BSE testing, refused to sell them the test kits,
and intervened to stop Creekstone from purchasing (the

The necessity of depopulation of CWD-infected farms by state agencies becomes a taxpayer
burden, and is only one part of containment.
Immediate action is required to avoid worst-case outcomes. We require mandate and funding to:

1. Contain the geographic spread of CWD by enacting and enforcing an immediate ban on the movement of all live cervids, all potentially CWD-infected carcasses, animal parts, products, exposed equipment, trailers, or other sources of infectious materials.

2. Mandate and implement for hunters, convenient, cost-free, rapid testing of all animals harvested from CWD-affected areas.

3. Ensure that no CWD-infected material reaches the food or feed chains, and that it is instead properly disposed of.

4. Establish and fund accountable research and science-based policy to protect public interest (health, wildlife and related industries, agriculture, our economies and communities).

It is important to note that consensus regarding these needs extends to optimism regarding the efficacy, the practical efficiency, and cost-effectiveness of the actions. Comprehensive analysis with vital stakeholder engagement will foster understanding, re-connection, and genuine appreciation of interrelated systems on which we depend. While dire, we have a unique opportunity to realize enormous advantages of cooperative effort and precautionary approach.

Consensus on urgency and vital actions

The preceding are but glimpses into the various disciplines, aspects, and complex factors involved in analyzing CWD, the existing impacts, and dynamic risks. It is abundantly clear, however, that the CWD crisis is, indeed “insidious and dire.” While details and methods must be guided by science and evidence, there is significant agreement on critical needs; and we have assurances from leading experts that we have the labs and capacity to meet this challenge.
Endnotes


10. Michael Samuel, USGS Wisconsin Cooperative Wildlife Research Unit, University of Wisconsin, personal communications.

11. Williams, E. S. Chronic Wasting Disease. 2005. Veterinary Pathology 42(5) : 540. Note that by the time of writing (2004) Beth had dismissed any notion of CWD as a longstanding indigenous disease. The three possible hypotheses she outlines include: a transfer from scrapie, potential sporadic emergence, or from an alternative TSE, possibly anthroponotic.


15. Michael Samuel, USGS Wisconsin Cooperative Wildlife Research Unit University of Wisconsin, personal communications.


68. Bryan Richards, Chronic Wasting Disease Project Leader, National Wildlife Health Center, personal communications.


71. Claudio Soto, University of Texas Medical Center, personal communications 2016.


73. Johnson, C. 2013 Ibid.


75. Canadian Food Inspection Agency. 2014. ibid. See p. 5.

76. Williams, E. S. 2005. ibid. See p. 5.

77. Ingram, J. 2013. Fatal Flaws: How a Misfolded Protein Baffled Scientists and Changed the Way We Look at the Brain. New Haven: Yale University Press. USA.

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103. Wisconsin Natural Resources Board. 2011. Almond Deer Farm Update. Tom Hauge, Bureau Director, Cathy Stepp, Secretary, presented to DNR Board meeting, Dec. 2011.


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108. Claudio Soto, University of Texas Medical Center, personal communications 2016.


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146. Nichols, T., 2016. ibid.


171. Kong, Q. “Zoonotic Potential of CWD Prions.” Late-breaking Abstracts, Prion 2015. Fort Collins, amended in personal communications Sept. 2016. Liuting Qing1, Ignazio Cali1,2, Yue Yuan1, Shenghai Huang3, Diane Kofsky1, Pierluigi Gambetti1, Wenquan Zou1, Qingzhong Kong1 1Case Western Reserve University, Cleveland, Ohio, USA, 25econd University of Naples, Naples, Italy, 3Encore Health Resources, Houston, Texas, USA. Zoonotic Potential of CWD Prions. LATE-BREAKING ABSTRACTS PRION 2015 CONFERENCE PRION 2015 CONFERENCE FT. COLLINS CWD RISK FACTORS TO HUMANS. These results indicate that the CWD prion has the potential to infect human CNS and peripheral lymphoid tissues and that there might be asymptomatic human carriers of CWD infection. See also: Kristen Davenport, Davin Henderson, Candace Mathiason, and Edward Hoover Prion Research Center; Colorado State University; Fort Collins, CO USA . Conversely, FSE maintained sufficient BSE characteristics to more efficiently convert bovine rPrP than feline rPrP . Additionally, human rPrP was competent for conversion by CWD and FCD. This insinuates that, at the level of protein:protein interactions, the barrier preventing transmission of CWD to humans is less robust than previously estimated. http://chronic-wasting-diseases.blightcot.ca/2016/09/texas-parks-wildlife-chronic-wasting.html.


173. David Pedzeric, Past President, Saskatchewan Wildlife Federation, personal communications.

174. Katherine Mehl, Manager, Saskatchewan Environment, personal communications.


176. David Clausen, (former) Chair Wisconsin Natural Resources Board, personal communications

177. All the data are from the Wisconsin Department of Natural Resources.

178. David Clausen, (former) Chair Wisconsin Natural Resources Board, personal communications.

179. David Clausen, (former) Chair Wisconsin Natural Resources Board, personal communications.

180. Claussen, D., Rowledge, D., Richards, B. (unpublished)

181. Qingzhong Kong, PhD, Case Western Reserve University, personal communications 2016.

182. Candace Mathiason, PhD, Colorado State University, personal communications 2016.


186. The BSE Inquiry, ibid. p. 159

187. Rowledge, D. 2008. ibid. See p.87

188. Orr, Joan; Starodub, Mary Ellen; Risk Assessment of Transmissible Spongiform Encephalopathies in Canada, for the Health Canada, Science Team on TSEs, Draft Report. June 30, 2000

189. Rowledge, D. 2008. See pp. 87, 114


198. Range determined by dividing the number of confirmed victims (229). Data from The BSE Inquiry. ibid. p.87


200. This is the unanimous consensus of dozens of prion scientists and health authorities.

201. Dr. Michael Coulthart, current Director, Canadian CJD Surveillance System, and Director, Prion Diseases Program for the Public Health Agency of Canada, personal communications 2016.


204. David Clausen, (former) Chair Wisconsin Natural Resources Board, personal communication. 2016.


206. FDA Guidance for Industry #158. ibid.

207. Michael Hansen, Senior Staff Scientist, Policy and Advocacy Division, Consumers Union, Yonkers, NY. personal communications 2010.


210. Draney, N., “Disease from the U.S. could wipe out all the deer in Britain.” The Times Print edition Nov. 22, 2013 archived online http://www.thetimes.co.uk/tto/environment/wildlife/article3928334.ece under the title “Wasting disease is a threat to the entire UK deer population”.

Emerging Infectious Diseases. 8(12):2091-2092. doi:10.3201/1812.120528.


221. David Pezderic, HACCP Coordinator/Export Controller Prairie Pride Natural Foods Ltd. — Federally registered poultry processor, Canada.


229. Alastair Lucas, Director, Sustainable Energy Development, (former) Dean of Law, (past) Chair Natural Resources Law, University of Calgary, personal communications 2016.


233. “I should much regret to see grow up in this country a system of large private game-preserves kept for the enjoyment of the very rich. One of the chief attractions of the life of the wilderness is its rugged and stalwart democracy; there every man stands for what he actually is and can show himself to be.”

~ Theodore Roosevelt, 1893
“There are some who can live without wild things and some who cannot.”

~ Aldo Leopold