Department
for Environment
Food & Rural Affairs

www.defra.gov.uk

What is the risk of chronic wasting disease being introduced into Great Britain?

A Qualitative Risk Assessment

October 2012

Llywodraeth Cymru
Welsh Government

DOE
Department of
the Environment
www.doeni.gov.uk

The Scottish
Government

This document/publication is also available on our website at:

http://www.defra.gov.uk/animal-diseases/monitoring/risk-assessments/

Any enquiries regarding this document/publication should be sent to us at:

Rowena Kosmider [Rowena.kosmider@ahvla.gsi.gov.uk] or Helen Roberts [helen.roberts@ahvla.gsi.gov.uk]

Contents

Summary

Chronic wasting disease (CWD) is a highly infectious transmissible spongiform encephalopathy (TSE) that is circulating in the wild and farmed cervid populations in North America. It is the only TSE to be prevalent in free-ranging wild animal populations. A feature of CWD is its ability to spread both directly and indirectly via the contaminated environment where it is able to survive in a bio-available form for many years without any significant decrease in infectivity. Eradication of the disease from wild and farmed or managed cervid populations and the environment is extremely challenging and has not yet been successful.

Currently, there have been no reported cases of CWD or other TSE in deer in Great Britain (GB) or Europe. Given the consequences of CWD observed in North America, it is imperative that GB remains free of the disease. This risk assessment aims to assess the risk of CWD being imported into GB from North America and consequently, consider the risk of exposure and infection within the GB deer population. The assessment focuses on two main routes of entry including importation of animal feed and movement of contaminated clothing, footwear and equipment of tourists, deer hunters and British servicemen between affected areas of North America and GB. It is important to highlight that there are significant data gaps in this assessment. The main conclusions from this assessment are:

- Several different animal feed products are imported into GB from North America. These include processed pet foods and consignments of unfinished feed ingredients for use in animal feed. The amount of imported feed, including pet food, that contains cervid protein is unknown and identified as a significant data gap. As non-ruminant animal feed may be produced with cervid protein (but not from positive CWD animals) in the United States (US), there is a **greater than negligible** risk that feed with cervid protein is imported from North America into GB. There is, however, uncertainty associated with this estimate.
- In areas of North America where CWD has been reported, given that CWD is excreted in faeces, saliva, urine and blood, and survives in the environment for several years where it is able to bind to the soil, there is a **medium probability** that the environment (including soil) contains CWD.
- Given the volume of tourists, hunters and servicemen moving between GB and North America, the probability of at least one person travelling to/from a CWD affected area and, in doing so, contaminating their clothing and/or equipment prior to arriving in GB is **greater than negligible**. For deer hunters, specifically, the risk is likely to be greater given the increased contact with deer and their environment. However, there is significant uncertainty associated with these estimates.
- Once in GB, the use of animal feed is subject to the TSE Feed Ban and ABP Regulations. In accordance with the current ban, farmed deer should not be directly exposed to (i.e. feed) imported animal feed containing any PAP. Therefore, assuming this ban is strictly adhered to, the risk of farmed and wild deer being

exposed to ruminant animal feed containing deer protein from North America is considered **negligible** but with associated uncertainty. The probability of a (wild) deer being exposed to CWD infected deer protein in non-ruminant feed is considered to be **greater than negligible** but uncertain.

- The pathways by which naïve deer in GB may be exposed to CWD contaminated soil and prions on equipment and clothing from people arriving in GB from North America are variable and highly uncertain. Given associated uncertainty, there is a **greater than negligible** probability that a person could transfer CWD prions from their contaminated equipment and/or clothing into deer habitat/environment, particularly with respect to Roe deer (*Capreolus capreolus*) habitat but less so for Chinese Water deer (*Hydropotes inermis*) habitat. Further, given the volume of tourists and other travellers moving between North America and GB, there are potentially multiple opportunities for CWD prions to be transferred from equipment to the environment.
- None of the species affected by CWD in North America are present in GB. For a British species to become infected with CWD given exposure will depend on the dose and inherent susceptibility. Based on current scientific evidence Red deer (*Cervus elaphus elaphus*) are susceptible to CWD, Fallow deer (*Dama dama*) are likely to be less susceptible and Roe deer (*Capreolus capreolus*) have a gene conferring susceptibility. Therefore, it is likely that given exposure to an infectious dose of CWD, deer in GB could become infected with CWD.
- However, given that the amount of soil ingested is likely to be very small, the probability of ingesting an infectious dose via this route is considered **negligible to very low**. The probability of ingesting an infectious dose via consumption of non-ruminant feed is likely to be higher and may be **very low**, with associated uncertainty.

Overall, the probability of importing CWD into GB from North America and causing infection in British deer is uncertain but likely to be **negligible to very low** via movement of deer hunters, other tourists and British servicemen and **very low** via imported (non-ruminant) animal feed. However, if it was imported and (a) deer did become infected with CWD, the consequences would be severe as eradication of the disease is impossible, it is clinically indistinguishable from BSE infection in deer (Dagleish *et al.,* 2008) and populations of wild and farmed deer would be under threat.

Background

Chronic wasting disease (CWD) is a highly infectious transmissible spongiform encephalopathy (TSE) that is circulating in the wild and farmed cervid populations of North America. It is the only TSE maintained in free-ranging wild animal populations. A feature of CWD is that it is able to transmit both directly (animal-to-animal) and indirectly via the contaminated environment. In particular, CWD prions are able to bind to and survive in the soil in a bio-available form for many years without any decrease in infectivity. This makes eradication of the disease from a wild population increasingly challenging.

Thus far, there have been no reported cases of CWD or other TSE in deer in Great Britain (GB). This is based on surveys of wild and farmed red deer (*Cervus elaphus elaphus*) (EFSA, 2011). Given the consequences of CWD observed in North America, it is of high importance that GB remains free of the disease. Further, as the clinical signs of CWD in deer are similar to those of deer experimentally infected with bovine spongiform encephalopathy (BSE), all infected deer would need to be tested to differentiate if they were infected with CWD or BSE to minimise the risk of BSE entering the human food chain via affected venison.

Possible routes of entry of CWD into GB include importation of animal feed and the movement of tourists, hunters and British servicemen travelling between affected areas in North America and GB. In regards to the latter route, soil contaminated with CWD prions could be imported on people's boots, clothing and other equipment. This route of entry has been noted previously as a viable means for the global dispersal of free-living organisms (Wilkinson, 2010).

Hazard identification

The hazard is identified as **Chronic Wasting Disease**

Chronic wasting disease (CWD) is a transmissible spongiform encephalopathy (TSE) affecting cervids. It is the only TSE maintained in free-ranging wild animal populations; other TSE's are mostly restricted to captive domestic livestock populations or humans. Chronic wasting disease was first identified as a clinical disease of captive mule deer in Colorado in 1967 and later classified as a TSE in 1978 (Williams & Miller, 2003). The origin of the disease is unknown and may have been a spontaneous TSE that arose in deer. Currently, natural infections of CWD have been reported in mule deer (*Odocoileus hemionus hemionus*), black-tailed deer (*Odocoileus hemionus columbianus*), white-tailed deer (*Odocoileus virginianus*), Rocky Mountain elk (*Cervus elphus nelsoni*), Shira's moose (*Alces alces shirasi*) and mule deer and white-tailed deer hybrids (Hamir *et al*., 2008). Other species of elk may also be susceptible. The disease is restricted to North America except for isolated cases of infected elk being exported from Canada to South Korea (Williams & Miller, 2003). In the last decade, with increased CWD testing, a more widespread distribution of CWD within North America has been observed (Sigurdson, 2008) (Figure 1).

Figure 1: Current range of CWD in North America (map reproduced by permission of Chronic Wasting Disease Alliance; www.cwd-info.org)

This more widespread distribution may be due to enhanced surveillance but also to natural migration of cervids and translocation of infected animals by humans (EFSA, 2011). Within affected areas, the prevalence varies. In the endemic area of Wyoming, for example, the prevalence of CWD in mule deer has increased from approximately 11% in 1997 to 36% in 2007 (Almberg *et al*., 2011). In such areas, population declines of deer of up to 30 to 50% have been observed (Almberg *et al*., 2011). In areas of Colorado, the prevalence can be as high as 30% (EFSA, 2011).

The clinical signs of CWD in affected adults are weight loss and behavioural changes that can span weeks or months (Williams, 2005). In addition, signs might include excessive salivation, behavioural alterations including a fixed stare and changes in interaction with other animals in the herd, and an altered stance (Williams, 2005). These signs are indistinguishable from cervids experimentally infected with bovine spongiform encephalopathy (BSE). Given this, if CWD was to be introduced into countries with BSE such as GB, for example, infected deer populations would need to be tested to differentiate if they were infected with CWD or BSE to minimise the risk of BSE entering the human food-chain via affected venison.

The duration of clinical disease is highly variable and death can occur within 4 weeks but some infected animals may survive as long as a year (Williams, 2005). The incubation period is a minimum of approximately 16 months and is more likely to be between 2 and 4 years (Williams, 2005). In affected elk, the incubation period is between 1.5 and 3 years after which they become clinically affected and may succumb less than 12 months after initial clinical signs appear (Miller *et al.*, 1998). During the pre-clinical period, the animal is infectious (Almberg *et al*., 2011).

The CWD agent (P_rPCWD) in affected animals is distributed firstly in the gut associated lymphoid tissues, digestive tract (e.g. tonsils, Peyer's patches, mesenteric lymph nodes) and then in the brain and spinal cord as the disease progresses (Sigurdson, 2008). Prions of CWD have also been found in muscle tissue (Angers *et al.*, 2006) (see Figure 2). The distribution and levels of PrP^{CWD} in tissues differ between species (e.g. elk versus deer).

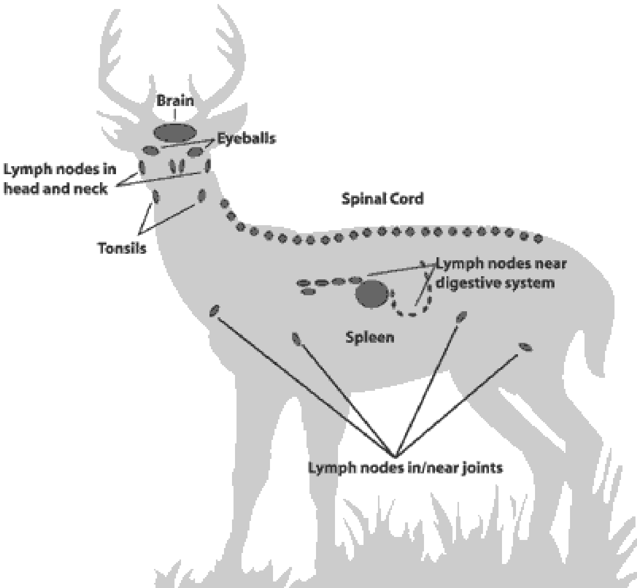


Figure 2: Diagram displaying the main organs affected by CWD in infected cervids (http://www.dnr.state.mn.us/mammals/deer/cwd/index.html)

Given its propensity to colonise the digestive tract, evidence suggests the prion is excreted in faeces (Safar *et al*., 2008), urine and saliva potentially leading to direct and indirect transmission between cervid species. Indeed, the disease is transmitted horizontally with high efficiency and circumstantial evidence suggests that environmental contamination with CWD prions contributes to the maintenance of CWD in affected areas (Safar *et al*., 2008). The rate of transmission of CWD has been reported to be as high as 30% and can approach 100% among captive animals in endemic areas (Safar *et al*., 2008). The efficiency of CWD transmission is unparalleled among TSE diseases (EFSA, 2011). Trifilo *et al*., (2007), using a murine tg mouse model, established that CWD can be transmitted via the oral route. Indeed, the distribution of PrPres in the orally infected mice (e.g. in the spleen and lymph nodes) mimicked what has been reported in deer developing CWD via natural infection (Trifilo *et al.,* 2007). Modelling studies also support the theory that transmission of CWD in deer herds is maintained by contact with a prion contaminated environment (Almberg *et al*., 2011). Scavenging of CWD-infected carcasses provides another route of releasing the prion into the environment and exposure of non-cervid species (Sigurdson, 2008). This indirect transmission route is problematic as it not only increases the basic reproductive number but also because there are very few effective mitigation strategies for reducing the risk from indirect transmission. This is due to the fact that the agent is extremely resistant in the environment and able to bind to soil particles making eradication and control of CWD a major obstacle in both farmed and free-ranging cervid populations.

Sheep and cattle may be exposed to CWD via common grazing areas with affected deer but so far, appear to be poorly susceptible to mule deer CWD (Sigurdson, 2008). In contrast, cattle are highly susceptible to white-tailed deer CWD and mule deer CWD in experimental conditions but no natural CWD infections in cattle have been reported (Sigurdson, 2008; Hamir *et al*., 2006). It is not known how susceptible humans are to CWD but given that the prion can be present in muscle, it is likely that humans have been exposed to the agent via consumption of venison (Sigurdson, 2008). Initial experimental research, however, suggests that human susceptibility to CWD is low and there may be a robust species barrier for CWD transmission to humans (Sigurdson, 2008). It is apparent, though, that CWD is affecting wild and farmed cervid populations in endemic areas with some deer populations decreasing as a result.

Thus far, CWD is restricted to North America with the exception of imported infected animals into South Korea from Canada. Surveys of wild and farmed cervid populations in the European Union between 2006 and 2010 did not identify any TSEs (EFSA, 2011). As part of this survey, 601 farmed and 598 wild red deer (*Cervus elaphus elaphus*) were tested (EFSA, 2010). These included clinical/sick animals, fallen stock, healthy shot/slaughtered animals and road killed animals. Based on the survey results, it can be concluded that the prevalence of CWD in the EU is less than 0.5%. It is important, therefore, to ensure that the disease is not introduced into Europe and establish within the EU wild and farmed cervid population as the probability of being able to eradicate the disease would be very small.

Risk Question

This risk assessment considers the risk posed to the Great Britain (GB) deer population if chronic wasting disease (CWD) was imported from North America (i.e. Canada and the United States). The specific risk question addressed is:

What is the risk of CWD being introduced into Great Britain (GB) from North America and causing infection in deer?

To answer the above question, the risk assessment follows the OIE framework of release (or entry), exposure and consequence assessment. Specifically, it is divided into the three key areas:

1. What is the probability of introducing CWD into GB from North America?
2. What is the probability of a deer species in GB being exposed to the CWD prion?
3. What is the probability of a GB deer species becoming infected with CWD upon exposure to the prion?

Risk Assessment

Terminology related to the assessed level of risk

For the purpose of the risk assessment, the following terminology will apply (OIE, 2004):

Negligible	So rare that it does not merit to be considered
Very low	Very rare but cannot be excluded
Low	Rare but does occur
Medium	Occurs regularly
High	Occurs often
Very high	Event occurs almost certainly

Entry assessment

The routes by which CWD may be introduced into GB from North America include:

- Importation of live deer
- Importation of meat and other products derived from cervid species (e.g. trophy items including antlers, semen)
- Importation of animal feed
- Hunters and other tourists and British servicemen travelling from affected areas to GB with contaminated equipment (e.g. boots, clothing, knives)

Currently, according to the European Union Trade Control and Expert System (TRACES) database, GB does not import live cervids, 'other' animal meat products or raw hides and skins. This was the same conclusion drawn by EFSA (2004) who stated GB does not import cervids or products from North America. Therefore, the two routes which this assessment focuses on are:

1) importation of animal feed
2) importation of CWD prion on contaminated equipment and clothing/footwear of hunters or other tourists and British servicemen

Importation of animal feed

Animal feed encompasses all feed fed to farmed livestock, horses, pets, farmed fish, zoo and circus animals and also animals living freely in the wild. Currently, legislation for animal feed relating to production, and labelling and composition is harmonised at the EU level and, in GB, is the responsibility of the Food Standards Agency (FSA). In addition, Defra is responsible for ABP Regulations which includes pet food manufacturing regulation.

Pet food containing material of animal origin, according to EU Regulation (EC) No. 142/2011 on Animal By-Products, must be derived from animals inspected and passed as fit for human consumption prior to slaughter. Further, the products are subject to strict microbiological criteria for *Enterobacteriaceae* and *Salmonella*. Under the EU Regulation, imported pet food produced using Category 3 processed animal proteins (PAP) must adhere to the same standards as that produced within the EU. More specifically, the imported pet food must satisfy the following criteria:

- The PAP must have been produced in accordance with the same requirements as PAP for placing on the market in the EU
- The PAP must have been sampled and tested to satisfy certain bacteriological criteria in accordance with the Regulations before release onto the EU market
- The product must enter the EU under correct Health Certification
- The Health Certification signed by the veterinarian or official inspector responsible for the rendering plant in the exporting country must verify that EU standards of sourcing of animal by-products, processing and sampling are met for each consignment.

These requirements apply to canned pet food, processed pet food other than canned pet food, dog chews, raw pet food and flavouring innards.

According to TRACES, GB imports processed pet food from Canada and the United States of America (USA). In November and December 2011, for example, GB imported 201.375 kg processed cat and dog food from Canada and USA. There are a limited number of processed pet food products made in the USA containing (roasted) venison for the cat and dog food market that are available in GB (e.g. Taste of the Wild pet food). Venison is high in iron content and considered a good alternative meat product for pets with intolerance to certain meat proteins. The specific amount of pet food products imported into GB from North America containing deer protein is unknown but is likely to be a small percentage of the overall amount of processed pet food imported.

Most other animal feeds imported are bulk consignments of ingredients, mainly of vegetable origin, that are then processed for final product use in GB. The TRACES database provides data on the varying and numerous animal feed products that are imported into GB. Two datasets have been extracted previously from TRACES providing information on animal feed importation. In January and February 2012, a survey of feed materials and feed products imported into GB from North America was conducted. All the products were exclusively of vegetable origin and included, for example, citrus pulp, corn gluten, sunflower, beet pulp, soil oil, and wheat feed. In a second survey, the amount of processed animal proteins including fishmeal imported from USA to GB in 2010 and 2011 was calculated. In 2010, 1359520 kilos of flours, meals and pellets of fish or meat and unfit for human consumption was imported into GB; 8716 kilos were of non-fish origin. In 2011, the amount of the same material imported decreased to 700818 kilos of which 8412 kilos were of non-fish origin.

In the USA, under the Food and Drug Administration's BSE Feed Regulation (21 CFR 589.2000) most material (exceptions include milk, tallow, and gelatin) from deer and elk is prohibited for use in feed for ruminant animals. With regards to feed for non-ruminant animals, under FDA law, CWD positive deer may not be used for any animal feed or feed ingredients. For elk and deer considered at high risk for CWD, the FDA recommends that these animals do not enter the animal feed system. However, this recommendation is guidance and not a requirement by law. Animals considered at high risk for CWD include: 1) animals from areas declared to be endemic for CWD and/or to be CWD eradication zones and 2) deer and elk that at some time during the 60-month period prior to slaughter were in a captive herd that contained a CWD-positive animal. Therefore, in the USA, materials from cervids other than CWD positive animals may be used in animal feed and feed ingredients for non-ruminants. The amount of animal PAP that is of deer and/or elk origin imported from the USA to GB can not be determined, however, as it is not specified in TRACES. It may constitute a small percentage of the 8412 kilos of non-fish origin processed animal proteins that were imported from US into GB in 2011.

Overall, therefore, it is considered there is a **greater than negligible** risk that (non-ruminant) animal feed and pet food containing deer and/or elk protein is imported into GB. There is uncertainty associated with this estimate given the lack of data on the amount of deer and/or elk protein possibly being imported in these products.

Movement of hunters, other tourists and British servicemen

Probability that the environment in North America is contaminated with CWD

As outlined in Figure 1, there are 20 states and provinces in the USA and Canada where CWD has been detected in farmed and wild cervids. These include: Alberta (Canada), Colorado, Illinois, Kansas, Maryland, Minnesota, Missouri, Nebraska, New Mexico, New York, North Dakota, Saskatchewan (Canada), South Dakota, Utah, Virginia, West Virginia, Wisconsin and Wyoming. In these areas, the environment is likely to be contaminated with CWD prions from direct excretion of the prion in various bodily fluids of infected animals, and leaching of prions into the soil from decaying carcasses of infected animals. A summary of the current studies on CWD in faeces, urine, blood and other bodily fluids or organs is summarised in Table 1.

Table 1: Summary of the studies on CWD prion excretion

Fluid/organ	Study summary	Reference
Faeces	• Faeces were the source of CWD infection in Syrian hamsters under experimental conditions. • Prions remain intact after being passed through the digestive tract and, therefore, are a viable	Safar *et al*., (2008)

	source of infectivity in the environment. • CWD prions are excreted in the faeces of infected mule deer 7 to 11 months prior to the onset of neurological signs (i.e. during the incubation period)	Tamguney *et al.* (2009)
Saliva	• During studies of oral transmission using a murine tg mouse model, it was observed that prior to and during clinical disease, serous and mucous glands contained PrPres. • Three naïve fawns were orally inoculated with 50ml of saliva from an infected deer in 3 doses over a 3 day period. Eighteen months post inoculation, CWD prions were detected in all 3 fawns during tonsil biopsy. • Pooled saliva from five terminally CWD infected white-tailed deer was inoculated into nine *tg1536* mice. Eight of the nine mice developed disease consistent with a TSE at 342 ± 109 days post inoculation suggesting infectious prions are present in saliva of infected cervids.	Trifilo *et al*., (2007) Mathiason *et al*., (2006) Haley *et al*., (2009)
Blood	• Two naïve white-tailed deer were inoculated intraperitoneally with 250 ml of frozen citrated blood and a further fawn with an intravenous transfusion of 250 ml freshly citrated whole blood. After 18 months post inoculation, all three fawns had CWD prions in their tonsils and retropharyngeal lymph node.	Mathiason *et al*., (2006)
Urine	• Pooled urine from five terminally CWD infected white-tailed deer was inoculated into nine *tg* mice. Two of the nine mice developed disease consistent with a TSE at 370 and 376 days post inoculation suggesting infectious prions are present in the urine of infected cervids but at a lower infectivity than other bodily fluids such as saliva.	Haley *et al*., (2009)
Antler velvet	• Antlers are covered by a layer of skin, velvet, which is shed after an increase in testosterone and ossification of antlers. CWD is present at low, but detectable, amounts in antler velvet from infected elk.	Angers *et al*., (2009)

It is clear from Table 1, that CWD is excreted in several different bodily fluids and, as demonstrated in experimental studies, can be a source for onward transmission to naïve animals. Infected carcasses decaying naturally in confined areas can also lead to new CWD infections in naïve deer (Sigurdson, 2008). This was proved, experimentally, by Miller *et al*., (2004) during a study of environmental transmission. Specifically, 3 naïve mule deer were stocked in a 800m^2 paddock in which a naturally infected mule deer had died and decomposed approximately 1.8 years prior. In a second paddock, a further 3 naïve mule deer were placed where infected mule deer had resided 2.2 years earlier and contaminated the environment with their faeces (Miller *et al*., 2004). The experiment was conducted in 3 replicates. In total, 3 out of 12 and 1 out of 9 deer were infected by being exposed to an infected decomposed carcass or residual excreta, respectively.

The CWD prion has also been detected in water. Specifically, very low levels (below infectious levels) were detected in a water sample from melting winter snow-pack from an endemic area (Nichols *et al*., 2009). The data showed persistence of CWD prions in water, accumulated levels of which, it is hypothesised, may promote transmission within deer herds.

Once in the environment, TSE prions can bind to soil particles and remain infectious (Saunder *et al*., 2010). Indeed, Johnson *et al.*, (2006) demonstrated that the disease-associated form of the prion protein can bind to all soil mineral surfaces and is preserved in a bioavailable form. Further, in a later study, Johnson *et al*., (2007) observed that prions bound to the soil mineral montmorillonite (Mte) significantly enhanced disease penetrance and reduced the incubation period compared to unbound prions. The reason why binding to Mte or other soil components enhances transmissibility is unclear but it may provide some protection for the prion in the gut against denaturation allowing more agent to be absorbed by the animal (Johnson *et al*., 2007). Further, binding to the soil particles maintains prions near the soil surface increasing the probability of animal exposure (Russo *et al*., 2009).

In addition to the enhanced infectivity, prions can remain in the soil for several years as the agents are resistant to inactivation by most chemical agents, radiation and heat (Johnson *et al*., 2006). Seidel *et al.* (2007), for example, demonstrated that scrapie agent (strain 263K) remains persistent in soil over a period of at least 29 months and remains highly infectious to Syrian hamsters in oral inoculation experiments. In Iceland during an epidemiological investigation of scrapie, a TSE of sheep and goats, Georgsson *et al*., (2006) reported that the scrapie agent survived on a farm for at least 16 years. However, Russo *et al*., (2009) demonstrated experimentally that reactive soil components such as manganese oxides may contribute to the inactivation process of TSE prions in soil. The authors did not study CWD prion specifically but the study highlights the complexity of the effect the inorganic and organic constituents in soil may have on prion survival and infectivity.

In summary, in endemic areas, there is a **medium** probability that the soil and surrounding environment is contaminated with CWD prions and in a bioavailable form. In rural areas where CWD has not been reported and deer are present, there is a **greater than negligible** risk the soil is contaminated with CWD prion.

Movement of deer hunters, other outdoor tourists and British servicemen between North America and GB

The probability a person comes into contact with CWD prions varies depending upon their place of residence and/or their involvement with outdoor pursuits (e.g. hunting). In this assessment, focus is given to the following groups of people:

- Residents in CWD affected areas travelling to GB (particularly the countryside) and British tourists travelling to CWD affected areas
- North American hunters travelling to GB to hunt/stalk deer and British hunters travelling to North America to hunt deer

- British servicemen training in and/or near CWD affected areas

All other people (e.g. city tourists and residents) are considered to pose a lower risk of being exposed to CWD in North America and, therefore, arriving in GB with contaminated clothing, footwear and/or equipment.

There are limited data on the number of North American tourists, stratified by location of residence, arriving in the UK. Therefore, it is not possible to ascertain of the 3.89 million visitors from the United States (US) to the United Kingdom (UK) in 2006, for example, how many were from CWD affected areas. Likewise, there is no breakdown of where in the USA the 4.5 million UK visitors travelled in 2008. This is a significant data gap in the assessment.

Hunting in the US is a popular sport with 4% of the population (10.1 million) involved in deer hunting in 2006 (USFWS, 2011). Of these people, 4.7 million only hunted deer whilst the remainder hunted deer and other species (e.g. small game, bears). Further, 58% of deer hunters were involved in wildlife watching activities and other outdoor pursuits (e.g. hiking, fishing); this is compared to 31% of the general public (USFWS, 2011; TAMS, 2007). Individuals that hunted whilst on a trip were likely to come from northern and western states (e.g. Alaska, Wyoming, North Dakota, South Dakota, Idaho and Montana) compared to highly urbanised states. In Canada, those individuals participating in hunting activities are most likely to do so within Canada with the majority taking a trip within their own province or region (TAMS, 2006). Canadian hunters were also more likely than other Canadian pleasure travellers to participate in wilderness activities and hiking (TAMS, 2006). There are no data collated on the number of hunters from North America travelling to the UK to stalk/hunt deer and, vice versa, there are no data on the number of UK residents hunting in North America. However, in order to hunt in GB with your own rifle, a visitor firearms permit has to be obtained from the police force in one of the devolved countries. In 2011, 123 licences were granted by the Scottish Police Force for non-EU residents (BASC, pers. Comm., 2012). This includes not only individuals from North America but also Norway and other non-EU countries (BASC, pers. Comm., 2012). The number of hunters arriving without their own rifle and participating in an organised hunting package/holiday is unknown. However, it is likely the total number of hunters from North America is in the low hundreds; the actual number, however, is highlighted as a significant data gap.

As well as tourists, British servicemen frequently move between North America and GB. In particular, British Army servicemen use the Canadian Forces base at Suffield, Alberta to take part in extensive training exercises. Suffield is located in the southwest of Alberta and comprises of 2,690km^2 of prairie landscape. The eastern boundary is designated as a wildlife management area and is south of a wildlife management area in which CWD was reported as recently as 2011 (www.srd.albert.ca). Consequently, the servicemen have the potential to be in close contact with areas where CWD is present.

In summary, given the volume of tourists, hunters and servicemen moving between GB and North America, the probability of at least one person travelling to/from a CWD affected area and, in doing so, contaminating their clothing, footwear and/or equipment prior to arriving in GB is **greater than negligible**. For deer hunters, specifically, the risk is likely to be greater given the increased contact with deer and their environment. However, there is significant uncertainty associated with these estimates.

Probable amount of CWD prions on contaminated boots and equipment

Given that a hunter or tourist walks in areas which are contaminated with CWD, it is possible that they will collect soil on their boots and other equipment. This likelihood will increase if the hunter has shot and handled a CWD infected deer resulting in contamination of the hunting equipment (e.g. knives) and their clothing and they subsequently arrive in GB with this equipment, footwear and clothing. Further, the soles of hiking boots tend to retain more soil than those of normal shoes. Wilkinson (2010), for example, removed 0.1 g of soil from hiking boots after returning to GB from a 2-month research visit to Canada. The amount of CWD prion in this amount of soil will depend upon the density of CWD infected animals excreting prions into the environment and the type of soil; CWD prion binds to clay soil, for example. Animal mortality sites could also be hotspots of CWD prion given the highly infectious nervous system matter entering into the environment and soil (Saunders *et al*., 2010).

Exposure assessment

Importation of animal feed

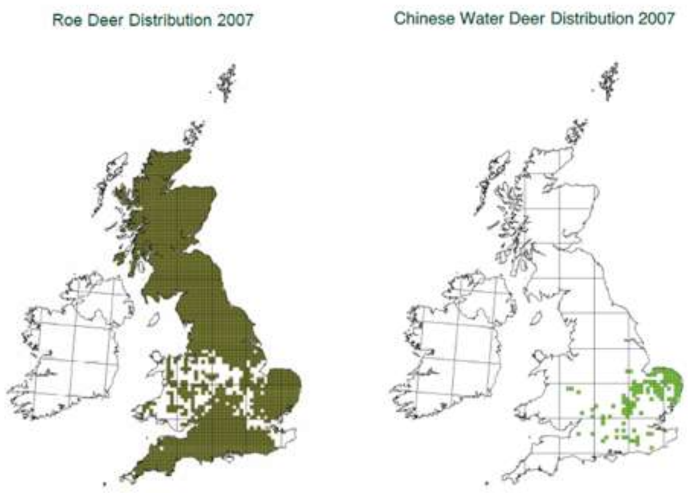
Once in GB, the use of animal feed is subject to the TSE Feed Ban and ABP Regulations. The BSE-related feed ban prohibits the feeding of PAP and gelatine from ruminants to ruminants (including farmed deer) or non-ruminant farmed animals. Further, ruminants must not be fed any animal protein or feedstuffs which contains animal protein except for milk, milk-based products and colostrum, eggs and egg products, gelatine from non-ruminants and hydrolysed proteins derived from non-ruminants or from ruminant hides and skins. Therefore, in accordance with the current ban, farmed deer should not be directly exposed to (i.e. feed) imported animal feed containing any PAP. Therefore, assuming this ban is adhered to correctly the risk of farmed deer being exposed to animal feed containing deer protein from North America is considered **negligible** but with associated uncertainty. However, given that non-ruminant feed produced in the USA may contain deer and elk PAP, it is theoretically possible that wild deer may be exposed to deer protein in legally imported non-ruminant feed. For this to occur, wild deer would need to access non-ruminant feed (e.g. pig, fish and chicken feed) on farms near their habitat. Alternatively, wild deer may be exposed to CWD prion in the faeces of pets that have consumed and digested imported, contaminated pet feed. The frequency in which these routes may occur is unknown and is considered to be a **greater than negligible** risk with associated uncertainty.

Movement of hunters, other tourists and British servicemen

The pathways by which naïve deer can be exposed to CWD contaminated soil and prions on equipment and clothing from people arriving to GB from North America are variable and highly uncertain. In principle, in order to expose a deer to CWD prions, the traveller (hunter, tourist or serviceman) would need to transfer the CWD prion from their clothing and/or equipment to the environment in which deer habit. The latter will depend upon the behaviours of returning GB residents or tourists and the probability of entering into and walking around deer territory. In GB, there are two main deer populations (wild and farmed or park deer) each of which will have differing risks of exposure given the type and frequency of contact with people. Each population type is considered in turn.

Wild deer

There are 6 species of wild deer residing in GB including: Red deer (*Cervus elaphus*), Roe deer (*Capreolus capreolus*), fallow deer (*Dama dama*), muntjac (*Muntiacus reevesi*), sika (*Cervus nippon*), and Chinese Water deer (*Hydropotes inermis*). The British Deer Society implemented a survey in 2007 to ascertain the distribution of these deer species across the UK. The survey provides the presence of deer on a standard template of 10km grid squares (www.bds.org.uk). A further survey was conducted in 2011 of which the results will be published later in 2012. The deer distribution as ascertained from the 2007 survey is summarised in Figure 3.



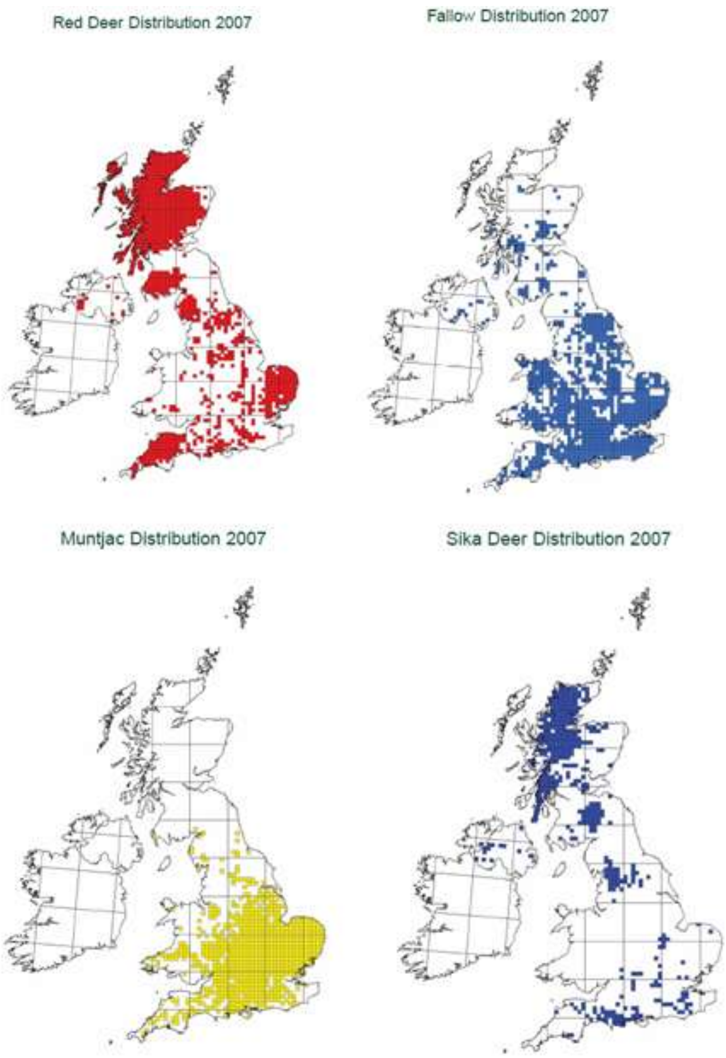


Figure 3: Distribution of the six deer species in the UK in 2007 (British Deer Society Deer Distribution Survey, http://www.bds.org.uk)

It can be seen from Figure 3 that deer are widely distributed across the UK with Roe deer being the most widespread. Chinese Water deer are the smallest deer population with approximately 700 deer.

Deer hunters, particularly, are most likely to be in direct contact with wild deer and their habitat compared to other tourists and returning GB residents. During the stalking and/or hunting of deer, there is opportunity for CWD prion on the hunter's boots, clothing and/or equipment to be transferred to the environment. The amount transferred will depend upon the measures taken to remove soil etc from the equipment prior to stalking. Assuming that CWD prion is transferred to the environment, there is an uncertain probability that a deer will come into contact with the CWD prion.

Farmed and park deer

Deer farming is a relatively recent enterprise. There are two systems currently used for managing enclosed deer: park and farm deer systems. In the park system, deer are raised in a park type setting and allowed to roam freely and may be provided with some supplemental feed. Farmed deer, in contrast, following conventional agricultural practices and may be housed in the winter and nutritional supplements are provided where necessary. In this farming system, there are several categories including calf rearers, calf finishers, breeder finishers and producer/processors (www.bdfpa.org). In 2011, according to the June Agricultural census, there were approximately 21,000 farmed deer on commercial agricultural holdings in England. The Economic Report on Scottish Agriculture (2011) cited that within Scotland, Wales, England and Northern Ireland there were 30,910 farmed deer. It is less likely that tourists, deer hunters and British servicemen will come into contact with conventionally farmed deer compared to park deer. The total park deer population in GB is unknown. However, in 2005, based on annual population control culling of about 8,000 animals, it was estimated that there were approximately 40,000 park deer. These deer are distributed across several parks (some of which are famous tourist sites) where wild and/or exotic species of deer can roam and be viewed. These are outlined in Table 2.

Table 2: Summary of parks in Great Britain where wild and/or exotic deer roam (The British Deer Society, www.bds.org.uk)

Country	Park	Species
England	Ashton Court, Bristol	Red, Fallow
	Bolderwood Deer Sanctuary, Minstead, Hampshire	Fallow,
	Bradgate Park, Charnwood Forest, Leicestershire	Fallow, Red
	Bushy Park, Hampton Hill, London	Fallow, Red
	Chatsworth Park, Chatsworth, Derbyshire	Fallow, Red
	Dunham Massey, Altrincham, Cheshire	
	Dyrham Park, Chippenham, Wiltshire	Fallow
	Grimsthorpe Castle Park and Gardens, Bourne, Lincolnshire	
	Helmington Hall, Stowmarket, Suffolk	Red

	Holkham Estate, Wells-next-the-Sea, Norfolk	Fallow
	Knole Park, Sevenoaks, Kent	Fallow, Sika
	Lodge Park & Sherborne Estate,	
	Lyme Park, Disley, Cheshire	Red, Fallow
	New Forest Wildlife Park, Longdown, Hampshire	European Bison, Red deer
	Petworth Park, Petworth, Sussex	Fallow
	Prinknash Deer and Bird Park, Cranham, Gloucester	
	Raby Castle, Staindrop, County Durham	Red, Fallow
	Richmond Park, Richmond, London	Red, Fallow
	Snettisham Park Farm, King's Lynn, Norfolk	Red
	South West Deer Rescue and Study Centre, Wayford	Red, White Red, Fallow, Axis, Roe, Japanese Sika
	Tatton Park, Knutsford, Cheshire	
	Wentworth Castle, Stainborough	Red, Fallow
	Wildwood Trust, Hern Bay, Kent	Roe, Fallow, Red
	Woburn Abbey, Woburn Park, Bedfordshire	Sika, Axis, Chital, Barasingha, Chinese Water, Rusa, Pere David
	Wollaton Park, Woolaton, Nottinghamshire	Red, Fallow
Wales	Abergavenny Priory Deer Park, Abergavenny	
	Dinefwr House, Kinefwr Park, Llandeilo, Carmarthenshire	Fallow

	Margam Country Park, Port Talbot	Red, Pere David, Chital, Hog, Barasingha, Roe, Muntjac, Chinese Water
Scotland	Beecraigs Country Park, Linlithgow, West Lothian	Red
	Glengoulandie Deer Park, Aberfeldy, Perthshire	Red
	Highland Wildlife Park, Kingussie, Inverness-shire	Red, reindeer
	Jedforest Deer and Farm Park, Jedburgh	
	The Scottish Deer Centre, Cupar, Fife	Nine species of deer

It is evident from Table 2 that there are several locations in GB where tourists and returning residents may come into contact with park deer and, in doing so, potentially expose the deer to CWD on their contaminated clothing and/or footwear. Further, given the volume of tourists and other travellers moving between North America and GB, there are potentially multiple opportunities for CWD prions to be transferred from clothing, boots and/or equipment to the environment. It has been observed that multiple exposures to low levels of CWD prions in the environment and increased infectivity of CWD when prions are bound to the soil are influential factors in transmission (Anger *et al*., 2009). Given the nature of their management, there is a restricted area (or environment) in which park deer inhabit enabling them to have a potentially higher probability of coming into contact with any CWD transferred to the environment by a tourist or returning GB resident compared to wild deer in a free-ranging environment. Therefore, it is considered that farmed and park deer may have a higher probability of exposure to CWD transferred to the environment than wild deer given the restricted habitat range and higher frequency of contact with tourists and returning GB residents.

Consequence assessment

Given that a deer within GB is exposed to CWD bio-available prions in the environment, the probability of becoming infected is dependent upon the infectious dose and the susceptibility of the animal to the prion. The majority of research into CWD has been conducted in North America where it has been shown that the following species are naturally infected with CWD (Hamir *et al*., 2008):

- Mule deer (*Odocoileus hemionus hemionus*)

- Black-tailed deer (*Odocoileus hemionus columbianus*)
- White-tailed deer (*Odocoileus virginianus*)
- Rocky Mountain elk (*Cervus elphus nelsoni*)
- Shira's moose (*Alces alces shirasi*)

None of these species are present in GB. However, EFSA (2010) considered that red deer (*Cervus elaphus*), a species present in GB (see Figure 3), is likely to be susceptible to CWD and was a species specifically targeted in the EU survey for CWD. This stems from the fact that red deer have a close genetic relationship to Rocky Mountain elk. This hypothesis has been supported by recent experimental studies that have demonstrated red deer become infected with CWD after oral inoculation of brain tissue from infected Rocky Mountain elk (Balachandran *et al*., 2010). Specifically, two of the four 2-month old red deer, showed clinical signs by 585 days p.i. and all deer had CWD prion in the brain, spinal cord and other organs at necropsy (Balachandran *et al*., 2010). Further, Martin *et al*., (2009) demonstrated in a similar study of four European red deer, that red deer can become infected upon inoculation of 5g of infected brain homogenate from four CWD elk and hence the species is susceptible to CWD.

Hamir *et al*., (2008) undertook a study to ascertain if fallow deer (*Dama dama*), another British deer species, could be experimentally infected with CWD brain suspension from infected elk or white-tailed deer. The authors concluded that it is possible to transmit CWD to fallow deer via the intracerebral route but the pathological features of CWD in the deer differs from those observed in white-tailed deer or elk (Hamir *et al*., 2008). It was further concluded that it might not be possible to transmit CWD via a more natural route or, alternatively, a higher dose of inoculum is required leading to a longer incubation period (Hamir *et al*., 2008).

Initial studies into the PRNP gene variability in European red deer and roe deer suggest that these species have a PRNP genetic background that is compatible with TSE susceptibility, including CWD (EFSA, 2011). It is important to note, however, that no experimental studies on roe deer have been conducted verifying this hypothesis.

There are no data on the susceptibility of the other free-ranging deer species present in Britain (muntjac (*Muntiacus reevesi*), sika (*Cervus nippon*), Chinese Water deer (*Hydropotes inermis*)) to CWD. Further experimental studies would be required to investigate the susceptibility of these species to CWD. Therefore, on the basis of current scientific understanding, it is likely that given exposure to an infectious dose to CWD, deer in GB could become infected with CWD. Whether the amount of CWD prion that could be transferred from clothing, boots and/or other equipment into the deer's environment is enough to induce infection given that the infectious dose is extremely small (Saunders *et al*., 2010) is uncertain. However, given that the amount of soil ingested is likely to be very small, the probability of ingesting an infectious dose via this route is considered **negligible to very low**. The probability of ingesting an infectious dose via consumption of non-ruminant feed is likely to be higher and may be **very low**, with associated uncertainty.

Control and risk management options

In order to reduce the potential amount of CWD prion entering GB on boots and clothing, it is important to meticulously clean off all adherent material prior to departing from North America. As CWD is a highly resistant agent, where possible, equipment should be soaked in a solution of bleach that has 20,000 parts per million of active chlorine for one hour. However, it is acknowledged it is impractical to soak hunting boots, clothing or firearms, for example, in strong concentrations of bleach.

Other risk management options could include:

- Landing cards could ask visitors whether they have visited CWD endemic areas and Border control officers could physically clean boots; Golfers returning to Australia have to declare their shoes for cleaning on entry.

Conclusions

There is significant uncertainty associated with estimating the risk of CWD entering the UK via movement of people (tourists, hunters and British servicemen) and importation of animal feed. This partly stems from the lack of data on these two importation routes. Given this uncertainty, the probability of importing CWD into GB from North America and causing infection in British deer is likely to be **negligible to very low** via movement of deer hunters, other tourists and British servicemen and **very low** via imported (non-ruminant) animal feed. The consequences of CWD, however, are severe with the minimal possibility of eradicating the disease from a wild cervid population and populations of wild and farmed deer would be under threat.

Current research indicates that of the six free-ranging deer species in the UK, red deer are susceptible to CWD. This deer species is concentrated in distinct areas of the country (e.g. North of Scotland) and one of the key species which hunters, in particular, stalk. It is important, therefore, that the risk of this species being exposed to CWD is minimised by taking appropriate precautionary measures.

References

Almberg, E.S., Cross, P.C., Johnson, C.J., Heisey, D.M. & Richards, B.J. (2011) Modeling routes of chronic wasting disease transmission: environmental prion persistence promotes deer population decline and extinction. *PLos ONE*, 6(5), e19896.

Angers, R.C, Browning, S.R, Seward, T.S., Sigurdson, C.J., Miller, M.W., Hoover, E.A., & Telling, G.C. (2006) Prions in skeletal muscles of deer with chronic wasting disease. *Science*, 311, 1117.

Angers, R.C., Seward, T.S., Napier, D., Green, M., Hoover, E., Spraker, T., O'Rourke, K., Balachandran, A., & Telling, G.C. (2009) Chronic wasting disease prions in elk antler velvet. *Emerging Infectious Diseases*, 15 (5), 696-703.

Balachandran, A., Harrington, N.P., Algire, J., Soutyrine, A., Spraker, T.R., Jeffrey, M., Gonzalez, L., & O'Rourke, K.I. (2010). Experimental oral transmission of chronic wasting disease to red deer (*Cervus elaphus elaphus*): Early detection and late stage distribution of protease-resistant prion protein. *Canadian Veterinary Journal-Revue Veterinaire Canadienne*, 51, 169-178.

Dagleish, M.P., Martin, S., Steele, P., Finlayson, J., Siso, S., Hamilton, S., Chianini, F., Reid, H.W., Gonzalez, L., & Jeffrey, M. (2008) Experimental transmission of bovine spongiform encephalopathy to European red deer (Cervus elaphus elaphus). *BMC Veterinary Research, 4,* 17.

EFSA (2004) Annex to the EFSA Journal (2004) 70 On the Opinion on a surveillance programme for Chronic Wasting Disease in the EU.

EFSA Panel on Biological Hazards. (2011) Joint Scientific Opinion on any possible epidemiological or molecular association between TSEs in animals and humans. *EFSA Journal*, 9(1), 1945.

EFSA Panel on Biological Hazards. (2010). Scientific Opinion on the results of the EU survey for Chronic Wasting Disease (CWD) in cervids. *EFSA Journal*, 8(10), 1861.

Georgsson, G., Sigurdarson, S., & Brown, P. (2006) Infectious agent of sheep scrapie may persist in the environment for at least 16 years. *Journal of General Virology*, 87, 3737-3740.

Gross, J.E. & Miller, m.W. (2001) Chronic wasting disease in mule deer: disease dynamics and control. *Journal of Wildlife Management*, 65, 2001.

Haley, N.J., Seelig, D.M., Zabel, M.D., Telling, G.C., & Hoover, E.A. (2009) Detection of CWD prions in urine and saliva of deer by transgenic mouse bioassay. *PLoS One*, 4(3), e4848.

Hamir, A.N., Kunkle, R.A., Nicholson, E.M., Miller, J.M., Hall, S.M., Schoenenbruecher, H., Brunelle, B.W., & Richt, J.A. (2008) Preliminary observations on the experimental transmission of chronic wasting disease (CWD) from elk and white-tailed deer to fallow deer. *Journal of Comparative Pathology*, 138, 121-30.

Hamir, A.N., Kunkle, R.A., Miller, J.M., Greenlee, J.J., & Richt, J.A. (2006) Experimental second passage of chronic wasting disease ($CWD^{mule\ deer}$) agent to cattle. *J. Comp. Path*, 134, 67-73.

Hartley, M., Voller, F., Murray, T. & Roberts, H. (2012) Qualitative veterinary risk assessment of the role of wild deer in the likelihood of incursion and the impact on effective disease control of selected exotic notifiable diseases in England. Eur J. Wildl Res DOI 10.1.1007/s10344-012-0647-7.

Johnson, C.J., Philips, K.E., Schramm, P.T., McKenzie, D., Aiken, J.M., & Pedersen, J.A. (2006) Prions adhere to soil minerals and remain infectious. *PLoS Pathogens*, 2 (4), e32.

Johnson, C.J., Pedersen, J.A., Chappell, R.J., McKenzie, D., & Aiken, J.M. (2007) Oral transmissibility of prion disease is enhanced by binding to soil particles. *PLoS Pathogens*, 3 (7), e93.

Martin, S., Jeffrey, M., Gonzalez, L., Siso, S., Reid, H.W., Steele, P., Dagleish, M.P., Stack, M.J., Chaplin, M.J., & Balachandran, A. (2009) Immunohistochemical and biochemical characteristics of BSE and CWD in experimentally infected European red deer (*Cervus elaphus elaphus*). *BMC Veterinary Research*, 5:26.

Mathison, C,K., et al., (2006) Infectious prions in the saliva and blood of deer with chronic wasting disease. *Science*, 314, 133-136.

Miller, M.W., Wild, M.A., & Williams, E.S. (1998) Epidemiology of chronic wasting disease in captive rocky mountain elk. *Journal of Wildlife Diseases*, 34 (3), 532-538.

Miller, M.W., Williams, E.S., Hobbs, N.T., & Wolfe, L.L. (2004) Environmental sources of prion transmission in mule deer. *Emerging Infectious Diseases*, 10(6), 1003-1006.

Nichols, T.A., Pulford, B., Wyckoff, A.C., Meyerett, C., Michel, B., Gertig, K., Hoover, E.A., Jewell, J.E., Telling, G.C., & Zabel, M.D. (2009) Detection of protease-resistant cervid protein in water from a CWD-endemic area. *Prion*, 3, 171-183.

Rural & Environmental Science & Analytical Services (2011). Economic Report on Scottish Agriculture 2011 Edition. Scottish Government: Edinburgh.

Russo, F., Johnson, C.J., Johnson, C.J., McKenzie, D., Aiken, J.M., & Pedersen, J.A. (2009) Pathogenic prion protein is degraded by manganese oxide material found in soils. *Journal of General Virology*, 90, 275-280.

Safar, J.G., Lessard, P., Tamguney, G., Freyman, V., Deering, C., Letessier, F., DeArmond, S.J., & Prusiner, S.B. (2008) Transmission and detection of prions in feces. *Journal of Infectious Diseases*, 198, 81-89.

Saunders, S.E., Bartz, J.C., Vercauteren, K.C., & Bartelt-Hunt, S.L. (2010) Enzymatic digestion of chronic wasting disease prions bound to soil. *Environ. Sci. Technol.*, 44, 4129-4135.

Seidel, B., Thomzig, A., Buschmann, A., Groschup, M.H., Peters, R., Beekes, M., & Terytze, K., (2007) Scrapie agent (strain 263K) can transmit disease via the oral route after persistence in soil over years. *PLoS Pathogens*, 5, e435.

Sigurdson, C.J. (2008) A prion disease of cervids: Chronic wasting disease. *Veterinary Research*, 39, 41.

Tamguney, G., Miller, M.W., Wolfe, L.L., Sirochman, T.M., Glidden, D.V., Palmer, C., Lemus, A., DeArmond, S.J. & Prusiner, S.B. (2009) Asymptomatic deer excrete infectious prions in faeces. *Nature*, 461, 529-590.

TAMS (2006) Travel activities and motivations of Canadian residents: An overview. Ministry of Tourism.

TAMS (2007) Travel activities and motivations of U.S. residents: An overview. Ministry of Tourism.

Trifilo, M.J., Ying, G., Teng, C. & Oldstone, M.B.A. (2007) Chronic wasting disease of deer and elk in transgenic mice: oral transmission and pathobiology. *Vaccine*, 365, 136-143.

U.S. Fish & Wildlife Service (2011). Deer hunting in the United States: Demographics and Trends. Addendum to the 2006 National Survey of Fishing, Hunting, and Wildlife-associated recreation. Report 2006–10. Wildlife and Sport Fish Restoration: Arlington, VA.

Williams, E.S. (2005) Chronic wasting disease. *Vet Pathol.*, 42, 530-549.

Williams, E.S., & Miller, M.W. (2003) Transmissible spongiform encephalopathies in non-domestic animals: origin, transmission and risk factors. *Rev. Sci. Tech. Off. Int Epiz.*, 22, 145-156.

Wilkinson, D.M. (2010) Have we underestimated the importance of humans in the biogeography of free-living terrestrial microorganisms? *Journal of Biogeography*, 37, 393-397.