Department
for Environment
Food & Rural Affairs

What is the risk of a cervid TSE being introduced from Norway into Great Britain?

Qualitative Risk Assessment

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Contents

Summary

This document is an update of a previous risk assessment carried out in April 2016 to account for the risk of incursion of CWD from the USA and Canada and the assessment of a new pathway, deer urine lures. In April 2016, a report of CWD-like disease was reported in Norway in wild reindeer and later in moose, and this assessment is to review the risk pathways for this new geographic area. Therefore any updates which differ from the assessment of April will appear in red throughout the main text.

The Norwegian Veterinary Institute reported a wild reindeer (*Rangifer tarandus tarandus)* found moribund and which later died in March 2016, had tested positive for the presence of prions. This was the first case of TSE found in a cervid in Europe and the first ever TSE case in a reindeer or caribou. In regions where chronic wasting disease is commonly found in native deer, there have been no reported cases in caribou (related to reindeer), but this is due to the isolation of the populations, rather than the refractivity of the species. The following month, two further cases were reported in wild moose (*Alces alces*), in a different region and again testing positive for TSE prion protein. In the last month, a fourth case was detected, again in a reindeer in Nordfjell region, this time in a healthy bull, shot for disease surveillance.

The new assessment focuses on the potential routes of entry for a cervid TSE (whether confirmed as chronic wasting disease or another related prion) from Norway. The main conclusions from this assessment are:

- The likelihood of further cases being found in wild reindeer in Norway is high, if confirmed as CWD as opposed to a spontaneous mutation event.

- The likelihood of further cases being found in moose is dependent on whether these are confirmed as a familial case or if related to the TSE in the reindeer. Moose are generally solitary animals so the risk of spread will depend on the level of wider environmental contamination, rather than direct contact with other infected cervids.

- The likelihood of spread of a CWD type disease into the farmed reindeer herd or into other farmed cervids in Norway is difficult to assess, and depends on the level of contact between migratory wild reindeer and the semi herded populations in the north or with other farmed cervid species. In the USA, new foci are often detected first in farmed herds, before detection in local wild cervids, so the contact between these two discrete populations is clearly sufficiently high to facilitate transmission in North America. This should be assessed for Norwegian / Scandinavian populations.

- The likelihood of spread into other (wild) deer populations in Norway is medium.

- The possible routes of spread of TSE from Norway to the UK include movements of live animals, imports of deer-related products (urine lures, meat used as pet food),

contaminated equipment, including clothing and hunting or skiing equipment and soil surrounding plant imports.

- The likelihood of a reindeer imported from Norway to the UK being infected with TSE is very low as they are imported from farmed herds, but there is uncertainty around this level of risk as it is not known if there is disease in the farmed herds.

- Other cervid species are not generally imported from Norway to the UK and therefore this is a lower risk pathway. If the pattern of trade changes, or if disease is detected over a wider area, the risk will also change.

- For other animals, the movement of pet dogs used for hunting or sledging competitions should be assessed for whether meat of cervid origin is fed to the animals. Where this occurs, this is considered a low risk of disease introduction.

- For other pathways, these are a non-negligible risk which is difficult to evaluate without understanding more on the extent of disease in Norway, but is likely to be between very low or low, depending on the pathway. For lures made from natural deer urine, where the provenance of the animal is unknown, the risk is medium for North American origin urine and Norwegian.

- As a result of the cases in Norway not all the risk levels have increased in comparison to the same risk pathways with an origin in North America.

- Reindeer in the UK are not commonly kept – there are small herds present including in the Cairngorms and Staffordshire and some seasonal imports. However the poor outcome of reindeer kept in captivity in the UK means it is difficult to ascertain whether any may have been infected with prion disease – a fallen stock programme does not exist for such animals at present.

- Our previous assessment suggested that of the cervid British species, red deer (*Cervus elaphus elaphus*) are susceptible to CWD, fallow deer (*Dama dama*) may be less susceptible and the roe deer (*Capreolus capreolus*) prion gene codes for susceptibility (and are the most closely related to white-tailed deer). More recent experimental data suggest Sika deer, Chinese Water deer and Muntjac deer may also be susceptible. Therefore, it is likely that given exposure to an infectious dose of CWD or a related prion, deer in GB could become infected.

Overall, the probability of importing a TSE into the GB deer herds from Norway and causing infection in British deer is uncertain but likely to be **no greater than very low** via movement of deer hunters, other tourists and British service personnel; **at most, low** via live animal imports or imported (non-ruminant) animal feed; **very low** for the use of lures specifically sourced in Norway and **negligible** for plant imports. However, if it was imported and (a) deer did become infected with CWD, the consequences would be severe as eradication of the disease is unfeasible, it is clinically indistinguishable from BSE infection in deer and populations of wild and farmed deer would be under threat.

Acknowledgements

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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 very unlikely.

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*) carried out several years ago (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. The public health risk of CWD is not known but current assessments suggest the risk is very low.

In 2015, the British Deer Society (BDS) carried out an online survey of BDS and BASC members to gather evidence about the use of deer urine as a lure. Fifteen percent of respondents (~1,800) answered yes about knowing that deer urine was used as a lure. Of the respondents, less than 2% responded yes to using such a product themselves. Of those that use the product, 50% had sourced the product from the USA, while 20% use more than a litre in volume a year and ~70% is natural (as opposed to synthetic).

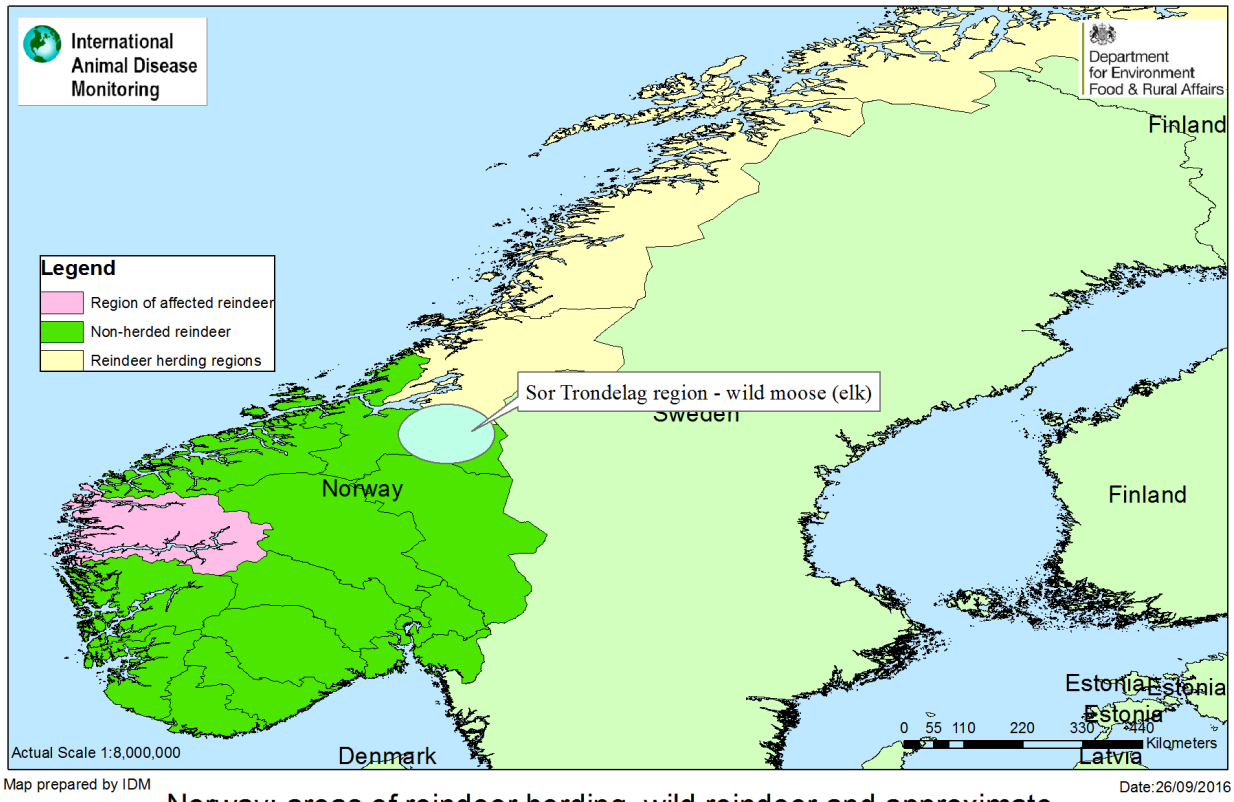
UPDATE: The report of TSE (CWD like) infection in Norwegian free ranging reindeer and then in European moose has increased the risk to the UK because of the trade in live animals and the different levels of activity for certain pathways, therefore we are reviewing those risk pathways. In addition, a further pathway was identified by a veterinary colleague overseas, which is that of plant and soil imports as well as for movement of hunting dogs so we have included these in the review.

Hazard identification

The hazard is identified as **TSE in wild European Reindeer and Moose**

The current geographic range of the cervid TSE, Chronic Wasting Disease (CWD) is the USA and Canada (and occasional outbreaks in South Korea following imports of infected animals). A new TSE type infection was identified in Norway, in a wild reindeer in March 2016 (NVI, 2016). Until the cases in Norway are confirmed to have a source in USA or Canada, at present we will identify the hazard as a transmissible spongiform encephalopathy (TSE) affecting wild European reindeer and moose.

The (found dead) adult reindeer cow showed signs of below-average body condition and detected in connection with capture for GPS-collaring, when it died. It was tested as a routine sample for the national surveillance programme for CWD at the Norwegian Veterinary Institute. Prion disease was confirmed in mid-March by both biochemical and immunohistochemical tests. According to the EURL, most of the samples from different organs were strongly positive for TSE prion protein. Samples were sent to the OIE reference laboratory (Canada) for confirmation as Chronic Wasting Disease. However, based on the widespread distribution of PrPCWD in the brain and the case history, the conclusion has been made that the animal had a spongiform encephalopathy compatible with CWD in the an early clinical stage (Benestad *et al.,* 2016). The animal was found in the Nordfjell region (see map below). In a second event, an adult (pregnant) female moose, (*Alces alces*) in the Sør-Trøndelag region was found with signs of poor body condition and lack of response to stimuli. It was culled and samples tested by both ELISA and Western blot tests. A third case was detected in another moose, found dead in a river near by a few days later. It also tested positive. There is approximately 300 km distance between the reindeer and the two elk cases. In a fourth case, in August 2016, a reindeer bull was shot and tested positive as part of a surveillance programme, also in the Nordfjell region (Sogn og Fjordane) and a fifth case, again in a reindeer (cow) shot by hunters in the same region was detected in September 2016. The North American moose (also *Alces alces*) is susceptible to CWD and cases have been found across the moose populations in both the USA and Canada. *Alces alces* is different to the North American elk (*Cervus canadensis*) which is also susceptible to CWD.



Norway: areas of reindeer herding, wild reindeer and approximate location of TSE-infected reindeer (04/16, 08/16 & 09/16) and moose (05/16)

http://wwweng.vetinst.no/eng/Highlights/Detection-of-Chronic-Wasting-Disease-in-two-Norwegian-moose.html

http://www.mattilsynet.no/dyr_og_dyrehold/dyrehelse/dyresykdommer/skrantesjuke__cwd_/ny_paavisning_av_skrantesjuke.23759

The passive surveillance system in Norway has been running since 2003 and involves testing samples from wild native cervid species of which there are four, red deer (*Cervus elaphus*), roe deer (*Capreolus capreolus*), moose (*Alces alces*) and reindeer (*Rangifer tarandus*) and from captive deer (Sviland et al, 2015). Red deer predominate along the west coast, wild reindeer live in high mountain areas in southern Norway (see map above). In 2013, the numbers of hunted cervids were nearly 35,000 moose, over 36,000 red deer, over 25,000 roe deer and nearly 8,000 reindeer. There is also a semi-domestic (herded) reindeer population of 250,000 which are located in north Norway and managed by the Sami people, and some of these animals will also be tested. There are 90 deer farms which mainly keep red deer and some keep fallow deer (*Dama dama*). Scrapie is present in sheep in areas where there are free ranging red deer populations. The number tested each year is very small; in 2014, only 10 deer were tested (all negative), and none of them reindeer. In 2013, again, just ten animals were tested (all negative) (Sviland et al, 2014) and in 2012, 21 animals were tested (all negative) and none were reindeer (Vikoren et al., 2013). This level of surveillance means that when a single positive sample is recorded, it suggests a high prevalence level is likely but the statistical confidence in such sampling is very low. It is not known how many animals have been tested in 2015/2016 for CWD under

the Norwegian programme. The Norwegian Authorities are proposing a large surveillance programme to start in the autumn of 2016, to test around 15,000 animals (moose, roe deer, red deer and reindeer) for fallen stock, hunted animals and at game slaughter houses and approved locations.

In Europe and North America, moose or elk (*A.alces*) are solitary animals, coming together primarily in the mating season, although young stay with their mothers for several months until the next offspring is born. There is a wide level of variation in their movement behaviour with some undertaking very long range migrations, and others being more sedentary. These movements can be categorised as migration, dispersal, nomadism or residence. In Scandinavia, seasonal migration is more likely in northerly populations (regions north of 66°N) than those in the southern regions (regions between 56°N and 66°N) and mean distances decline from ~100 km to 5 km. Seasonal migration can also change with time, depending on the environmental changes, climate or urbanisation. A recent study into the population genetics of *Alces alces* in Europe suggests there are genetically distinct populations, with the Scandinavian cluster showing low genetic diversity and separate to the other European populations (Niedziałkowska et al. 2016). Nevertheless, the low genetic mixing does not preclude mixing of animals at common grazing areas and therefore having access to contaminated land.

The genetic sub-structuring of the *A.alces* population in Scandinavia could be partly due to geographic barriers, such as the Scandes mountain range which separates Sweden and Norway. This supports the understanding that there is a lower risk of direct disease transmission to other populations of cervids, even of the same species, which are separated by semi-permeable geographic boundaries. However, if there has been widespread environmental contamination over time from a common source of prion, then the risk to other populations will be more difficult to assess.

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 the USA and Canada 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).

Caribou (*Rangifer tarandus caribou*, *R.t. granti* and *R.t. goenlandicus*) are a subspecies of the Eurasian reindeer, *Rangifer tarandus* and several populations overlap with the current CWD distribution in Canada. The disease has not been reported in the scientific literature in caribou as natural infections. However, experimental infection of six reindeer resulted in TSE in two of the six animals via oral inoculation (Mitchell et al, 2012). In this study on experimental infection in reindeer, Mitchell and colleagues showed that the two out of three reindeer infected with CWD prion protein (PrP) from brain homogenates of infected white-tailed deer started to show clinical signs between 17 and 18 months after oral inoculation. The same infection route using PrP from infected elk brains did not result in

clinical infection in three further reindeer. Results from histopathology showed PrP present in peripheral lymphoid tissue, in the kidney, the pituitary and adrenal glands, in nerves associated with the gastro-intestinal tract and of course the brain and central nervous system.

The widespread distribution in the USA and two Canadian provinces may be detected because of enhanced surveillance but may have increased because of natural movements 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). However the separation between caribou populations in Canada and affected cervids is probably the main reason for disease not being detected.

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 American 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 or Prion Protein (P_rP^{CWD}) 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 1). The distribution and levels of PrP^{CWD} in tissues differ between species (e.g. American elk versus white tailed or mule deer).

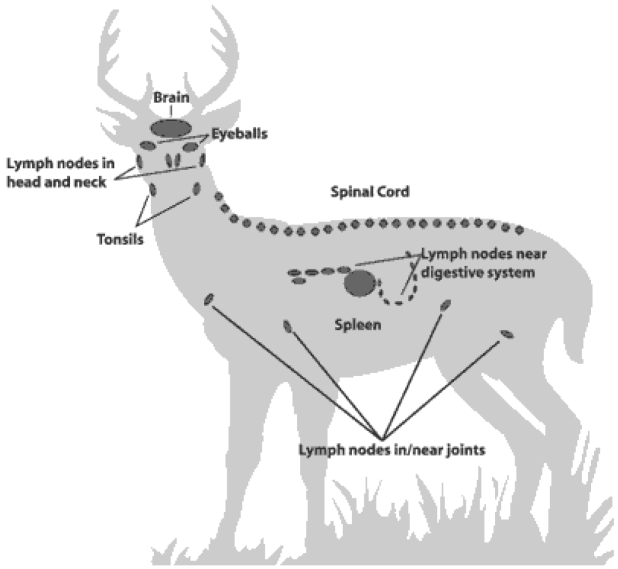


Figure 1: Diagram displaying the main organs affected by CWD in infected cervids (http://www.dnr.state.mn.us/mammals/deer/cwd/index.html)

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; Nalls *et al*., 2013). 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.

The hypothesis that disease can be transmitted between cervid species has been supported by recent experimental studies that have demonstrated that European red deer become infected with CWD after oral inoculation with brain tissue from infected Rocky Mountain elk (Balachandran *et al*., 2010). Specifically, two of the four 2-month old red deer challenged, 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 with 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). However it should be noted that these animals were all sourced from a single breeder therefore genetic diversity would be low and it cannot be ruled out that other fallow deer sourced from other breeders with greater heterogeneity would behave differently.

Initial studies into the PRioN Protein (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.

Recent 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 also suggests variability in susceptibility for these species (Robinson et al., 2012; Nalls et al., 2013). 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, most deer species in GB could become infected with CWD.

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 suggests that human susceptibility to CWD is low and there may be a robust species barrier for CWD transmission to humans (Sigurdson, 2008), however the risk appetite for a public health threat may still find this level unacceptable. It is apparent, though, that CWD is affecting wild and farmed cervid populations in endemic areas in the USA 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 was concluded that the prevalence of CWD in the EU is less than 0.5%.

Risk Question

This risk assessment considers the risk posed to the Great Britain (GB) deer population if chronic wasting disease (CWD) is confirmed in Norway. The specific risk question addressed is:

What is the risk of TSE being introduced into Great Britain (GB) from Norway 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 TSE into GB from Norway? [Entry assessment]
2. What is the probability of a deer species in GB being exposed to the TSE prion? [Exposure assessment]
3. What is the probability of a GB deer species becoming infected with TSE upon exposure to the prion? [Consequence assessment]

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 Norway include:

- Importation of live deer (including reindeer, other cervids, other animals)
- Importation of deer urine lures

- 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 (skiers and walkers) and British servicemen travelling from affected areas to GB with contaminated equipment (e.g. boots, clothing, knives)
- Importation of plants, shrubs and trees with root balls where the soil could be contaminated with prion protein

The previous assessments which we carried out concluded the risk of CWD being imported from the USA in meat or other products of animal origin (skins, antlers, semen) was very low and for animal feed as non-negligible (but likely to be very low) because of the requirement for deer entering the food or feed chain to be tested. For deer urine lures, the risk was considered medium. For areas where CWD had only recently been detected there was a very low risk of soil contamination so the risk from movement of equipment of people is considered to be non-negligible but would depend on the frequency of movement. The risk from live animals was not evaluated as live deer imports from the USA and Canada are not allowed under EU Regulation 206/2010. *A new pathway is also considered: for plant imports where root balls may be associated with soil contaminated with prion protein.*

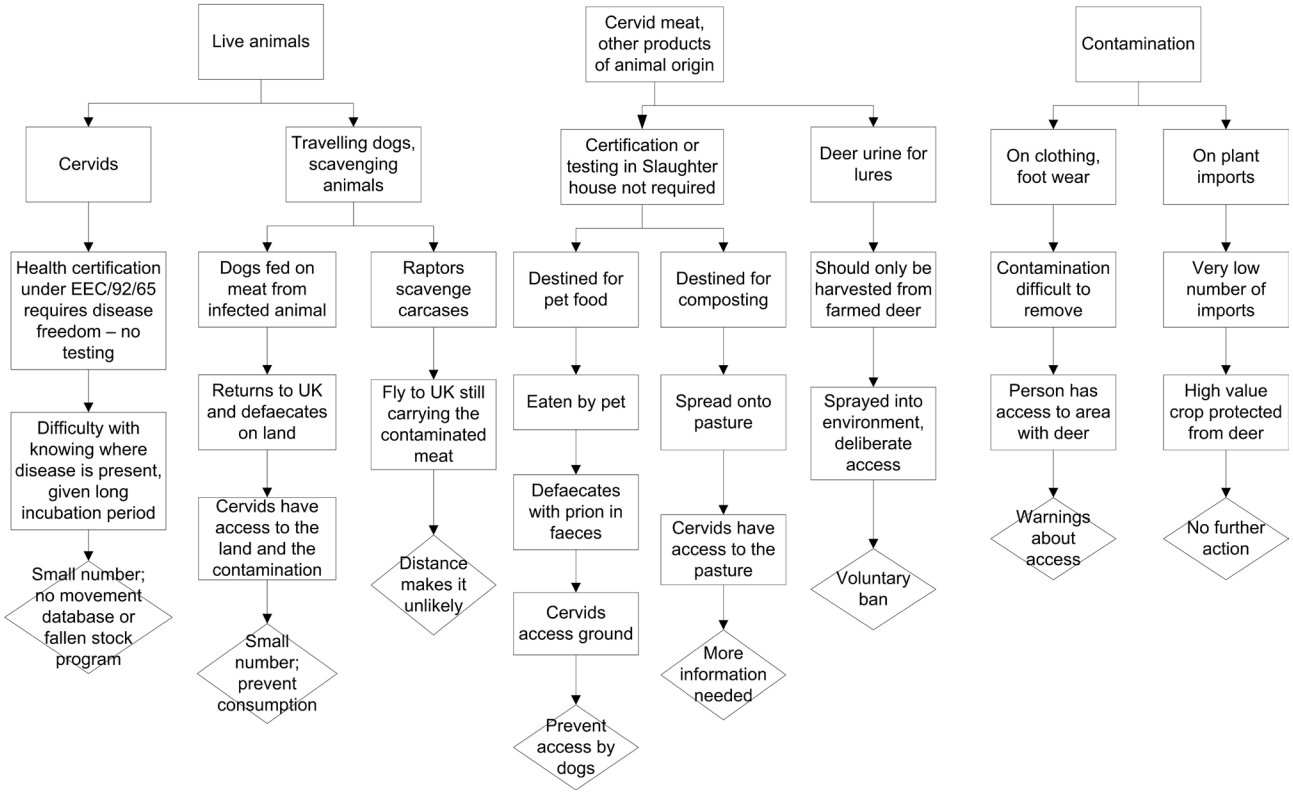
These same controls for meat, products and live animals are not applied to other countries as CWD was geographically restricted. Therefore, this assessment focuses on the following:

1) Imports of live animals
2) Imports of reindeer meat or products
3) Importation of animal feed
4) Importation of deer urine lures
5) Importation of CWD prion on contaminated equipment and clothing/footwear of hunters or other tourists and British servicemen
6) Importation of plants, shrubs and trees

Below is a schematic of the different pathways which could potentially lead to entry of CWD to the UK. Note, the human exposure pathway is not included here.

For each pathway, the end point is the possible action which could be put in place.

Schematic pathway for introduction of Chronic Wasting Disease through movement of live animals, products or contamination



Trade in live animals

Cervids

Assuming a maximum incubation period of 2-4 years, the EU Electronic Trade Notification System (TRACES) was interrogated for consignments of reindeer (*Rangifer tarandus*), moose (*Alces alces*) and other cervids since 2012. During this period, there have been just two consignments of 28 individual captive reindeer from Norway, as well as 13 consignments of 347 animals from Sweden and 15 consignments of 266 animals from Finland, all from approved holdings. In the case of the Norwegian imports, these are from the north of the country, significantly far from the wild reindeer case and are from semi-domestic herds. There were no recorded reindeer imports from Norway to the UK between 2011 and 2013 or since 2015. In terms of other deer species, there was a single consignment of a zoo species (Pudu) from Sweden, but no other deer species were recorded from the other Nordic or Baltic States. Multiple consignments of deer were traded from other countries in the EU. The importer of the reindeer from Norway has been in contact to assure us his animals are still in a healthy condition.

There is no requirement under EEC/92/65 for the animals to be pre-movement tested for CWD or for CWD-herd freedom for movement of any cervid species around the EU or EEA countries, as Europe was believed to be disease free. Nevertheless, the animals must be moved with a veterinary inspection and health certificate and TSE infection of cervidae is listed under Annex A as a notifiable disease of which approved bodies must be free in

order to trade. There is no CWD test approved for live animals, therefore the certifying veterinarian must be confident in signing that the herd is disease free.

On the basis of the small number of animals legally imported from Norway and the origin of the reindeer, we consider there is a **very low risk** of introduction via this route, but this is based on the **high level of uncertainty** around the possible geographic distribution of disease in Norway and the potential for mixing of wild and captive reindeer and low level of sensitivity in the surveillance programme. Once more surveillance is undertaken to ascertain the level of disease in the wild and semi-domestic herds, this risk level and the uncertainty associated with it can be refined. The level of illegal movement of reindeer or other cervids is not known and cannot be entirely ruled out, if there is deliberate attempt to move the animals without declaring them to the veterinary authorities.

Other animals

Working dogs may travel to Norway for hunting or sledging competitions. They travel as accompanied animals (under the pet travel scheme) and not with animal health certification (under EEC/92/65). The risk pathway is these dogs being fed raw deer meat and defaecating when they return to the UK. The prion protein would survive the intestine conditions and therefore lead to environmental contamination. It is difficult to assess the likelihood of this pathway, and there are no controls over the use of hunted meat for pet food which is used on farm as opposed to commercially produced. Around 300 dogs are moved under the pet travel scheme every year from Norway to the UK [1], which is less than 1% of all pets moving to the UK. Not all will be dogs involved in hunting or sledging competitions. Therefore we consider this to be a **low risk pathway** with **high level of uncertainty**.

Other animals which could potentially spread prion proteins to the UK would be from the movements of raptors or other scavenging birds which has fed on a carcase of an infected animal. We consider this risk pathway to be unlikely, given the distance between Norway and the UK.

Importation of reindeer meat or products

Products of animal origin destined for human consumption are traded from Norway to the EU and UK under the same rules as intra-community trade, under the EEA agreement and are subject to EU Food Law, Regulation (EC) 178/2002. This means there are no health certification requirements or border checks. However, there are still rules under the ABP regulations about not allowing the swill feeding of ruminant material to other ruminants and Food Law covers the general requirements for only safe food to be placed onto the market. Therefore this type of product should not be diverted to animal feed for ruminants. However, poultry and fish feed could contain ruminant material, and this is a pathway

[1] These numbers are based on figures provided voluntarily to the APHA from pet travel companies and therefore may not be accurate, but are representative.

which may need some further consideration if disease becomes more widely established or detected in farmed cervids.

At present, we therefore consider this is **no greater than a very low risk pathway**, as not only will the product originate in domestic livestock and go through slaughter house checks, but it will be destined for human consumption and not animal feed. In the USA, cervids over 12 months old entering a slaughter house must be tested for CWD, under the USA APHIS Herd Certification Programme, but this is not applied in Norway, therefore there is uncertainty associated with this risk estimate for Norway given the lack of data on the amount of deer or reindeer protein possibly being consigned for human consumption and the lack of surveillance data for the farmed herds in Norway. Food imported for human consumption can be reclassified as category 3 material and be composted or spread as fertiliser, but to understand this pathway better, we consider it would be beneficial to carry out a quantitative risk assessment on the infectious dose and availability of prions through this pathway, should disease be reported more widely.

This assessment is only to cover the risk to animal health if the food product were to be diverted to animal feed or mistakenly fed to cervids. It is not the role of this assessment to consider the public health risk associated with meat or other products from cervids infected with CWD. The European Food Safety Authority will be looking into this issue, but the current assessment from the Norwegian Authorities is that it poses a very low risk.

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 (i.e. feed for non-ruminants) containing material of animal origin, according to EU Regulation (EC) No. 1069/2009 and its implementing Regulation (EC) 142/2011 on Animal By-Products, must be derived from animals inspected and passed as fit for human consumption prior to slaughter (Category 3 – lowest risk - material). Category 1 material (eg from animals in which the presence of TSE has been confirmed or suspected, or which are derived from animals killed as part of a TSE eradication programme) must not be used for animal feed. Furthermore, the category 3 products are subject to strict microbiological criteria for *Enterobacteriaceae* and *Salmonella*. Under the EU Regulation Category 3 processed animal proteins (PAP) must also adhere to a set of standards. More specifically, the pet food must satisfy the following criteria:

- The PAP must have been produced in accordance with requirements 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

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

These regulations do cover entire bodies or parts of wild animals which are carcases and suspected of being diseased, such that they must be disposed of as Category 1 – high risk – material via rendering and incineration and not enter the food or feed chain. It will not cover wild animal carcases or parts from animals not suspected of being diseased or which have been hunted and not collected for disposal. Indeed, as this is considered EU trade there is no requirement for veterinary health certificates or TRACES information as this is part of the European Single Market. Deer antlers are a popular dog chew and deer velvet may contain a low level of prion (Angers *et al.,* 2009) therefore if the antlers are collected from wild deer and imported with the velvet still in place, this could represent a low risk pathway for the pet to consume and defaecate prion proteins.

CWD-like TSE disease in Norway has only been detected to date in the non-farmed, wild reindeer and moose populations, but there is still uncertainty over whether there could be undetected disease in the farmed herds in the North if they had been in contact with diseased animals in the non-herding areas.

Overall, therefore, it is considered there is a **very low** risk that (non-ruminant) animal feed and pet food containing TSE-infected cervid protein is consigned to GB. There is high uncertainty associated with this estimate given the lack of data on the amount of deer or reindeer protein possibly being imported in these products and the lack of surveillance data for the farmed herds in Norway.

Importation of deer urine lures

In our previous assessment, the import of deer urine from the USA as lures from the hunting industry was recognised as a potential route for introduction of disease to the UK / EU (Defra, 2016). It is well established that urine from CWD-infected deer contains CWD infectivity. The urine collected for deer lures has no processing and is immediately refrigerated and bottled (Anon 2015b). There is no inactivation of the CWD agent in the urine and thus all infectivity present at the point of bottling in the USA will still be present at the point of use in the UK. However, under EU rules, (EC Regulation 1069/2009), urine from farmed deer should fall under the definition of “manure” and therefore the import of such a product, if unprocessed, is not allowed, according to Regulation (EC) 142/2011. Nevertheless, the processing required for bottling prior to retail is unlikely to affect the CWD prion and if it did fulfil the requirements in 142/2011, the active ingredient in the urine would no longer be effective. It is therefore presumed that the urine is considered “unprocessed” under EU law. The import and transit of urine from wild deer is not covered by 1069/2009.

A survey conducted by the British Deer Society (BDS) in July 2015 suggested small number of hunters in GB were aware of and used deer urine lures (http://www.bds.org.uk/index.php/news-events/135-deer-urine-lure-survey-july-2015).

It is not known whether Norway is a source of imported deer urine for lures. No information has been found to suggest this. Therefore this route for introduction, while it cannot be discounted, is of no greater risk than for the import of deer lures from countries like the USA where disease is well established in certain deer species.

Animal urine is not considered a commodity which is subject to animal by-products legislation for imports. Internet sales are common and although a license would be required, there are no conditions for the safe sourcing of such products. Deer urine lures are available in Europe and may be produced from carcases of hunted deer. The use of deer urine produced from a species such as reindeer would be questioned for its value in attracting our native GB deer.

This risk is **very low,** but with a high level of uncertainty, based on the unknown distribution of disease.

Movement of hunters, other tourists and British servicemen

Probability that the environment in Norway is contaminated with TSE prions

At present there are just four animals of two different species and in two different regions which have tested positive for TSE prion proteins in Norway and it is not known what the wider geographic distribution may be or for how long the animals had been infected. However, given the incubation period in experimental infections, the infection was likely to be present for several months, possibly years. Without knowing the original source of disease it is difficult to make an assessment of what the environmental contamination would be. If these were each down to spontaneous mutation events, it may be limited, but it is unlikely for two unrelated species to each have spontaneous events within a few weeks of another. The animals in each case may have been ill for some time, most likely weeks rather than days given the poor condition of the carcases, therefore it is likely the animals were excreting prion protein in various bodily fluids, including urine and faeces which can occur during the pre-clinical phase. This can lead to leaching of prions into the soil and contamination. Animals with clinical signs may show erratic behaviour which can include wandering behaviour, so contamination may be wider than normal host range.

CWD (TSE) 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). There is no reason to expect TSE in reindeer or moose to behave differently, given the experimental infection data and the testing carried out at the EURL and NVI.

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.

The extent of the contaminated area is unknown at present as it would depend on the range of the infected, clinically ill animals and the presence of undetected infection. However, if there are a larger number of undetected cases of TSE and if animal carcases have not been picked up as fallen stock, but allowed to decompose *in situ*, or been predated on by other animals, the likelihood of contamination of the land is greater and the risk level increases. Carcases are only one possible source of contamination, as animals may be excreting infection in pre-clinical stages which could be for months or even years. Data from the USA confirm that infection is shed in urine and saliva in particular. The Norwegians are now withdrawing all salt licks because of this (http://www.mattilsynet.no/language/english/animals/additional_legal_measures_to_limit_the_spread_of_chronic_wasting_disease_cwd_in_cervids.23275). Large carnivores, such as lynx, wolves and wolverines may follow herds and prey on sick and weak members of the herd. Foxes, raptors and crows may also contribute to the dissemination.

In summary, in areas where TSE in cervids has only very recently been reported, it is certain that the soil would be contaminated with CWD prion, but the extent and persistence of that contamination is uncertain but we consider is represents a **medium risk in the immediate affected regions** and **very low to low risk for other areas in Norway**.

Movement of deer hunters, other outdoor tourists and British service personnel between Norway and GB

The probability a person comes into contact with TSE 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 TSE affected areas travelling to GB (particularly the countryside) and British tourists travelling to TSE affected areas
- Hunters travelling between Norway and GB to hunt/stalk deer
- British service personnel training in and/or near TSE affected areas

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

The region where the infected reindeer was found is a relatively poorly populated area, but nevertheless is an area where walkers and skiers will visit. Hunting in Norway is a popular sport with many thousand deer shot each year. In order for a foreign national 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. The actual number of known hunters either visiting Norway or coming from Norway to the UK is highlighted as a significant data gap. Some of these hunters may also take their own dogs to Norway as highlighted in previous section on live animal imports.

As well as tourists, British service personnel frequently move between Norway and GB for winter sports and for alpine training. Consequently, the service personnel have the potential to be in close contact with areas where TSE is present. However data provided by MoD confirm that these affected areas are not used by the British service personnel and therefore there is very low risk via this pathway, with a degree of uncertainty around how widespread is the contamination.

In summary, given the volume of tourists, hunters and service personnel moving between GB and Norway, the probability of at least one person travelling to/from a TSE affected area and, in doing so, contaminating their clothing, footwear and/or equipment prior to arriving in GB is considered **no greater than low**. For deer hunters, specifically, the risk is likely to be greater given the increased contact with deer and their environment. However, there is **high uncertainty** associated with these estimates, mainly around the level of environmental contamination in Norway.

Probable amount of CWD prions on contaminated boots and equipment

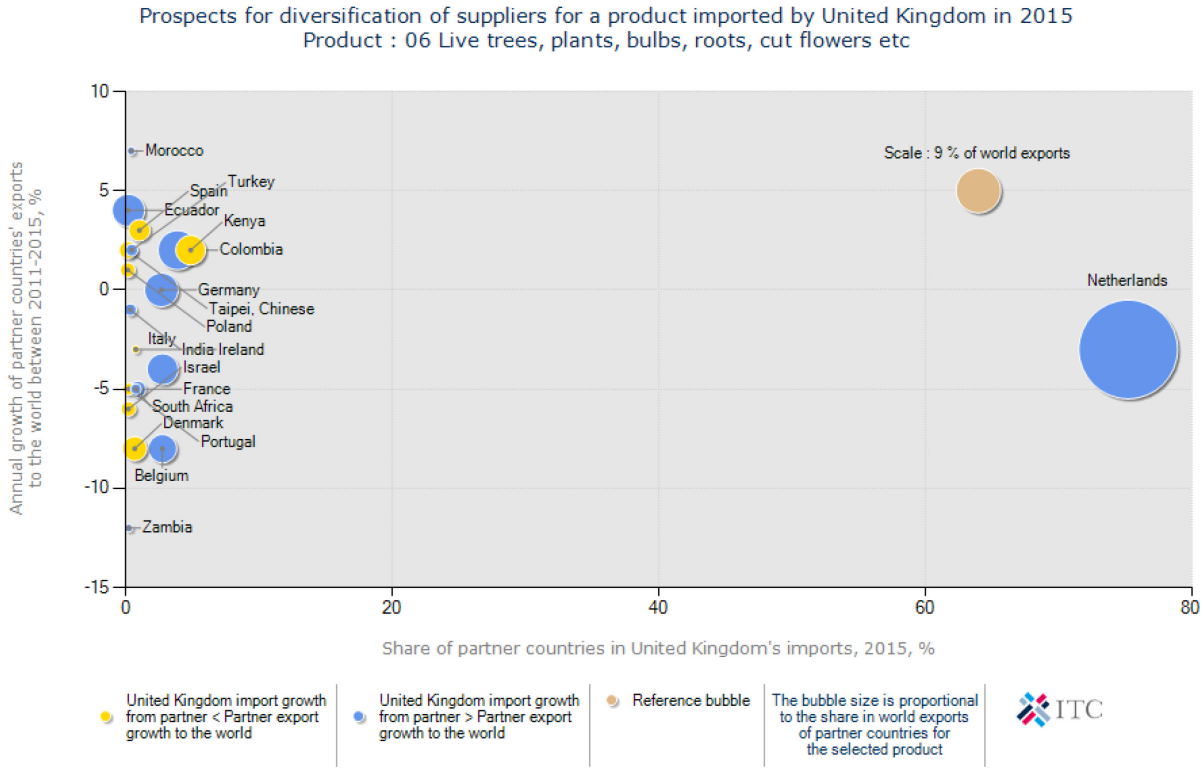
Given that a hunter or tourist walks in areas which are contaminated with TSE, it is possible that they will collect soil on their boots and other equipment. This likelihood will increase if the hunter has shot and handled a TSE infected carcase resulting in contamination of the hunting equipment (e.g. knives) and their clothing and they subsequently arrive in GB with this equipment, footwear and clothing. Furthermore, 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 TSE prion in this amount of soil will depend upon the density of TSE-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 TSE prion given the highly infectious nervous system matter entering into the environment and soil (Saunders *et al*., 2010).

Trade in plants, shrubs and trees

Live plants (trees, shrubs etc) are moved around the World in considerable quantities and many, including large full-grown trees will have intact root balls which will be covered in soil. As mentioned in the preceding section, prion proteins will be present in soil for many months / years as a result of contamination (Johnson *et al*., 2007).

There are several different categories for imports of stocks of plants, shrubs and trees. According to Eurostat data for codes beginning 0602 since January 2014, there have been only two consignments from Norway of “Live trees and other plants; bulbs, roots and the like; cut flowers and ornamental foliage // Other live plants (including their roots), cuttings and slips; mushroom spawn. // -Other”. Further possible commodities would be “plants for planting” which are generally smaller nursery plants, but we consider these are a very low risk of being in contact with cervids where they are grown, as there would be measures to exclude deer from these high value crops. Further interrogation of the UN COM trade database shows the greatest proportion of imports to the UK of Live trees, plants, bulbs, roots, flowers etc is from Netherlands, and Norway is only a tiny proportion of the total value of our trade. The following figure is a graphical presentation of the imports to the UK of these products. It is also noted that USA and Canada are similarly low exporters to the UK, with less than 50 tons a year combined (ITC, 2016).



Therefore although this is a potential route for introducing prion proteins into a new area, for this particular situation (Norway to the UK) we consider this **is a very low, if not negligible,** risk.

Exposure assessment

Imports of live animals

Reindeer management by the Sami in the north of the Scandinavian countries provides only two opportunities a year to handle the reindeer: early summer and autumn. It is these periods which provide many of the non-farmed reindeer for trade. Farmed reindeer can be drawn off and traded at any time, but it is the autumn draw which provides many of the reindeer for export across Europe for the seasonal trade for Christmas markets. In the past they have not thrived particularly in the UK and often succumb to welfare issues when brought in for this seasonal trade. There are a few semi-enclosed herds in the UK – including one in the Cairngorms and one in Staffordshire and there will be plenty of reindeer kept in small numbers in parks and zoos. The exact number is not known as there is no requirement for registration of such animals as there would be for farmed domestic livestock. It is however, estimated that there are around 1,200 – 1,500 reindeer in the UK and there is an intention to start a stud book for the UK animals in the future (British Reindeer Herders Association, pers. comm.). Using a combination of Intra Community Health Certification and performance licencing we have identified several other premises which keep low numbers of reindeer.

Should an infected animal be imported into the resident herds in GB, there is a high probability disease would spread to other in-contact conspecifics, but the level of contact with wild deer may not be as high as for other species (as these are valuable animals). It is nevertheless, difficult to prevent contact between wild and farmed deer and in the USA, disease is often detected first in farmed deer where there are more regular inspections. As there is no official programme of checks on the health status of the reindeer, it is not possible to ascertain whether disease is already present and CWD has not featured as a differential diagnosis for any fallen stock which is reported to APHA, because this was not considered a likely cause of death.

Importation of animal feed or products of animal origin

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 contain 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. fed) imported animal feed containing any PAP.

The likelihood of non-ruminant feed or cervid origin POAO being fed accidentally to reindeer or other captive deer in GB is very low.

Therefore, assuming this ban is adhered to correctly the risk of farmed deer being exposed to animal feed containing deer protein from Norway is considered **negligible** but with associated uncertainty. However, given that non-ruminant feed produced elsewhere (eg the USA) may contain deer and moose 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 TSE prion in the faeces of pets that have consumed and digested imported, contaminated pet feed. Also to be considered is that food may be diverted to composting and the resulting effluate spread on pasture. 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 or other affected region 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.

Consequence assessment

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*). In the UK, enclosed deer herds are principally red deer (*Cervus elaphus*) on farms and fallow deer (*Dama dama*) within parks: currently, only one commercial fallow deer farm is known to exist in Great Britain. The number of farmed deer in the UK has grown substantially to about 31,000. The numbers of park deer are unknown but the most recent assessment in 2005 gave an annual cull of around 8,000 deer suggesting about 40,000 park deer. The number of wild deer will be vastly more numerous than the enclosed or captive cervid species, estimated at around 2 million, with an annual cull of over 300,000.

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 TSE 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 TSE prion is transferred to the environment, there is an uncertain probability that a deer will come into contact with the TSE prion.

Farmed and park deer

The Deer Initiative recently carried out an informal review of enclosed deer herds in GB (Deer Initiative, pers comm. Data published in 2015). Current estimates give the number of extant deer parks or collections as 259. The wild roe deer population is the most numerous (see maps in Annex 1), while for farmed deer, fallow deer represent the highest number. Recent estimates provided by the Deer Initiative are:

Expected number (by species) held in parks and wild population			
Species	# expected parks	Expected population	Wild population
Sika spp. (*C.nippon*)	27	2160	~35,000
Red (*C. elaphus*)	111	12386	>350,000
Fallow (*D. dama*)	196	31849	150-200,000
Roe (*C.capreolus*)	-	-	~500,000

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). It is less likely that tourists, deer hunters and British service personnel will come into contact with conventionally farmed deer compared to park deer.

Reindeer were first introduced in the 1950's into the Cairngorms, where there is now an established herd. In the 1990's the imports of live reindeer increased with the popularity of Christmas markets. The imports referred to earlier, from Norway are to one importer where a small herd is maintained for breeding with occasional imported animals from the Northern farmed / semi-domestic herds to improve the bloodlines. Reindeer in this particular herd are fed deer food manufactured in the UK to an adapted Scandinavian recipe. Some feeds are available for other deer that are used and there are a number of manufacturers of pellet feed available in the UK now. Reindeer moss may be imported from a production facility in Norway. This is a dry product which is rehydrated with tap water prior to feeding. This tends to be harvested from close to the Finnish Border away from heavy populations of Reindeer otherwise it wouldn't be available to pick. A number of people use moss as a treat, training and supplementary feed. Reindeer moss doesn't store well wet unless frozen so UK imports are dried for longevity. Reindeer in parks in England are generally high fenced and this can reduce contact with local wild deer, particularly nose-to-nose even across single fencing, although the Cairngorm herd is more likely to have some level of interaction with local wildlife. However, it may not be possible to prevent contact with contaminated environment. Reindeer life expectancy is around 10-12 years and can be as long as 18 years in captivity, but is more commonly 14-16 years. There is no requirement for registration of cervidae and their movements, as there is for livestock species.

There are therefore several locations in GB where tourists and returning residents may come into contact with park deer. Nevertheless, there is a relatively low volume of tourists and other travellers moving between Norway and GB, so there are considerably fewer opportunities in comparison to the risk from the USA and Canada for CWD / TSE 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. Nevertheless, **the risk is not increased** by this case in Norway, as to that posed by visitors from the USA and Canada.

Exposure of UK deer to soil, feed or infected urine

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 **no greater than 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.

Although our previous risk assessment considered the risk of importing TSE through imports of deer urine for lures as medium entry assessment, and the likely exposure assessment as very low, the risk is not increased as a result of this case in Norway. The import of unprocessed deer urine is still an illegal product and should not be imported into the EU, no matter what the source of the animals.

The CWD agent is relatively dilute in deer urine compared to brain and spinal cord material with 1 ic LD_{50} per 10 ml. The LD_{50} determined by Henderson et al. (2015) is an intracerebral ID_{50} in cervidized transgenic mice. The oral ID_{50} in deer would be in a much larger volume of urine, because the oral route may be 100,000-fold less efficient than the intracerebral route in terms of TSE transmission (Gale et al 1998). Thus, in terms of oral LD_{50}, there may be one in a 1,000,000 ml i.e. 1 m^3 or 1,000 litre volume of deer urine. Therefore a deer would have to ingest 1,000 litres of urine to have a 50% chance of being infected through the oral route. Of course the CWD agent would be concentrated as the urine evaporated from the tree trunk.

The main sources of uncertainty are:-

1. The amount of urine ingested by the deer. A bottle of Tink's "69 Doe-in-Rut Buck" is about 30 ml and boasts 100% natural doe oestrous urine (Anon 2105d). A bottle from an infected doe would thus contain about 3 i.c. LD_{50} units. The bottles come with an easy to use squirt top, so it seems relatively small amounts are used. There are also gel forms which do not freeze as fast, last longer in the rain, and do not dry out so quickly.

2. The magnitude of oral/i.c. barrier

Assuming a deer drank 10 ml of urine from each 30 ml bottle, then that deer would ingest 10^{-5} LD_5. It is generally assumed that there is no threshold dose for TSEs (Gale 2006) and the risk of CWD infection in GB deer per 30 ml bottle imported is therefore 0.69 x 10^{-5} (Gale 1998). This is **very low**.

Risk of infection of deer in GB per 30 ml bottle of deer urine lure imported from the USA

Step in pathway	Risk	Uncertainty
Entry (probability a 30-ml bottle contains urine from an infected deer)	Medium	Low
Exposure (probability some of 30-ml bottle is ingested by UK deer)	Medium	High

Consequence (probability that deer is infected given exposure)	Very low	Medium
Overall risk	Very low	High

Overall the risk of a deer in GB being infected per 30 ml bottle of urine imported from the USA is **very low**, albeit with **high** uncertainty.

The next question is how much deer urine is imported into GB from the USA per year. If there are n 30-ml bottles, then the risk is calculated as:-

$$p_{n\_bottle} = 1 - (1 - p_{one\_bottle})^n$$

Hundreds of gallons of urine are sold every year just by one company in the USA (Anon 2015b). It is difficult, however, to estimate the volume of deer urine imported into the UK per year from the USA. That some respondents to the BDS survey reported they used upwards of 1litre per year, is consistent with importation of a high number of 30-ml bottles. Assuming that the number, n, of 30-ml bottles imported annually is high, then the risk of CWD infection in at least one deer in the UK per year will be **medium** (given the probability, of infection of a UK deer from one 30-ml bottle is very low (see Table 1)).

Overall it is concluded 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**. This assumes a high number of 30 ml bottles imported per year from all areas of the USA.

The case in Norway does not increase this risk at present. A quick internet search continues to suggest the main source of deer urine lures is the USA, nevertheless, in the BDS survey, half of respondents sourced deer urine from North America and a quarter from Europe.

Control and risk management options

The highest level of uncertainty is around the source and level of disease in Norway and whether it is limited to the wild reindeer herds or if it has spread into either other species of wild deer or the herder reindeer in the North. As there is so little by way of surveillance in the EU and surrounding countries, it is not possible to say whether this is a wider problem.

If these were spontaneous mutations, a form of “atypical CWD”, then these may be rare and isolated cases. However the animals concerned were in two regions, of two species and not particularly old, so it may be more likely the source is an imported prion infection. If this has an origin in the USA or Canada, it is important to try to understand the source and therefore whether there are likely to be more cases over a wider area, where the import event may also have taken place.

If it were to be found in the farmed or herded cervid populations, it will be necessary to put in place some import controls along the lines of those in place for imports of live animals and cervid meat from the USA and Canada. As it is, Norway voluntarily stopped signing trade certificates for all cervid species and authorisation is required to move cervids

between counties within Norway (http://www.mattilsynet.no/language/english/animals/additional_legal_measures_to_limit_the_spread_of_chronic_wasting_disease_cwd_in_cervids.23275).

If the geographic distribution is wider and the environment heavily contaminated, to reduce the potential amount of TSE prion entering GB on boots and clothing, it is important to meticulously clean off all adherent material prior to departing from an affected area and 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.

For deer urine lures, the import of unprocessed natural deer urine is not allowed under the EU Animal By-Product legislation. However the import of such commodities including internet sales would still require licenses, but it would be a voluntary action on behalf of the importer to ensure the urine is sourced from safe herds. One option is to ensure the sourcing is from herds in the USA which are currently registered under the National CWD Herd Certification Programme which has recently been introduced in the USA. Otherwise the use of synthetic or domestic-produced lures should be promoted by the industry and stakeholder groups.

Areas identified which are not fully understood are those of pets eating meat from infected animals and of food diverted to Category 3 for composting. Both would benefit from further investigation if the results of the Norwegian surveillance suggest wider distribution.

Diagnosis of CWD / TSE in cervids

Conventional immunohistochemistry (IHC) testing for TSE's involves using cadavers to remove the brainstem as the most sensitive organ to show the distribution of prion protein in the nervous and associated tissue is typical of the pathology associated with the species-specific TSE. The ELISA for CWD has similar sensitivity and specificity as the IHC test. Both tests use a protease step which can reduce sensitivity. This means that surveillance cannot be carried out on live animals. There is a considerable need for a fast, reliable antemortem test.

There is no current fallen stock scheme for testing deer species in the UK and there is no European Reference Lab for CWD (although the EURL for TSE's is APHA, Weybridge).

Conclusions

There is significant uncertainty associated with estimating the risk of TSE entering the UK from Norway via imports of live animals, movement of people (tourists, hunters and British servicemen) and importation of animal feed or urine lures. This stems from the lack of data on the current distribution of disease, not only in wild reindeer and moose in Norway, but also in herded animals and more widely in other wild cervids across the region and

neighbouring counties. Notwithstanding this uncertainty, the probability of importing TSE into GB from Norway and causing infection in British deer is likely to be **greater than negligible** via movement of deer hunters, other tourists and British servicemen and **low** via live animals, **very low** via products of animal origin or imported (non-ruminant) animal feed. However the risk of natural deer urine lures from Norway is lower than for those products sourced from the USA, where the risk of containing CWD PrP is considered **medium** (reflected by some US States banning the use) and the probability of such a commodity, if used in significant volumes, leading to CWD infection in GB populations is considered to be **very low** (potentially reduced susceptibility in certain species and limited use by hunters and stalkers in GB) but with a high level of uncertainty. Nevertheless, the voluntary restrictions on sourcing urine lures from USA and Canada should be extended to Norwegian sources, as a precautionary measure.

The consequences of CWD, however, are severe with the minimal possibility of eradicating the disease from a wild cervid population and populations of both wild and farmed deer in the UK would be under threat.

Current research indicates that of the six free-ranging deer species in the UK, red deer, and muntjac are susceptible to CWD, while roe deer, which is the closest related to white-tailed deer, Japanese sika and Chinese water deer are likely to also be susceptible. Farmed fallow deer are numerous in the UK and while those studied to date have lacked the PRNP polymorphisms associated with higher susceptibility to CWD, our populations are genetically heterogenous so the risk of infection cannot be ruled out. Wild roe deer are even more numerous, so again, understanding the susceptibility of this species will be important. The new case in reindeer in Norway taken alongside existing experimental data suggests that this species is also at risk.

It is important, therefore, that the risk of these species being exposed to TSE is minimised by taking appropriate precautionary measures.

Change in risk level for entry assessment (and level of uncertainty) from previous risk assessment (Defra, 2016)

Pathway	USA and Canada	Norway
Live animals - cervids	Not assessed as not possible	Very low (high)
Live animal - other	Not assessed as new pathway	Low (high)
Products of Animal Origin	Very low (low)	Very low (medium)
Animal Feed	Greater than negligible (medium)	Very low (high)
Urine lures	Medium (low)	Medium (medium)
Equipment and people	Greater than negligible (medium)	Greater than negligible (medium)
Plants	Not assessed as new pathway	Negligible (low)

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