Public Health England

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Qualitative assessment of the risk of Chronic Wasting Disease (CWD) to human health through non-food exposures

June 2018

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Published August 2018
PHE publications
gateway number: 2018337

PHE supports the UN
Sustainable Development Goals

Contents

Date of this assessment	24 May 2018
Version	1.1
Reason for assessment/update	24 May 2018 Update of additional cases of CWD in Norway and the first detection of CWD in Finland 17 July 2017 First detection of CWD in Europe during 2016 involving moose (*Alces alces)* and reindeer (*Rangifer tarandus)* in two different localities in Norway. This assessment was conducted to consider non-food risks to human health should it also be detected in the UK. This assessment is one of three parts. The other parts consider: • the risk of a cervid TSE being introduced from Norway into Great Britain • risk of infection from CWD prions and atypical CWD prions via consumption of cervid meat
Assessment	Public Health England CJD section and Emerging Infections and

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Date of previous assessment	17 July 2017

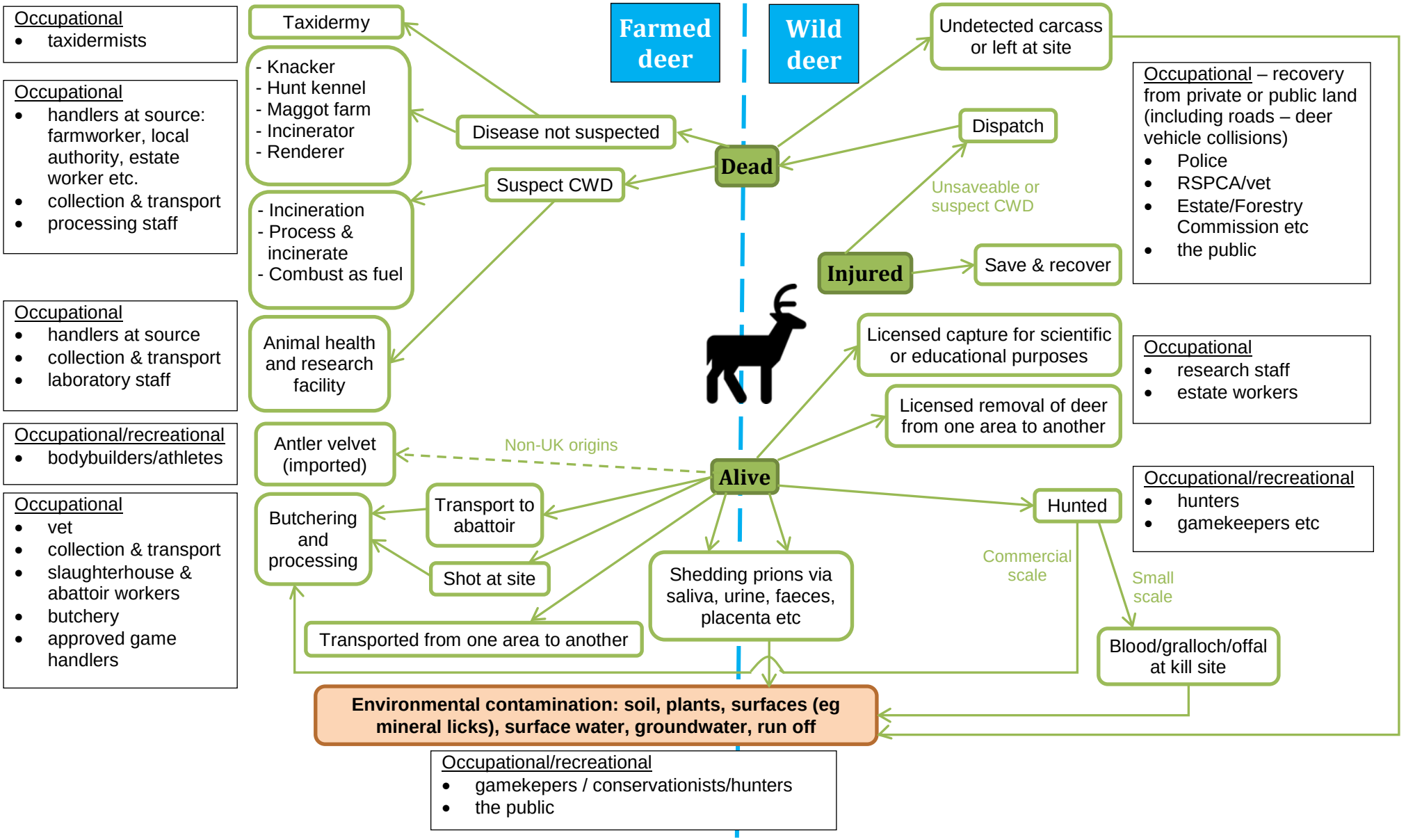
Overview

Qualitative assessment of the risk that CWD infected deer present to human health through non-food exposures	
Overview	The recent detection of Chronic Wasting Disease (CWD) for the first time in Europe raises concerns about domestic exposure to CWD prions and risks to human health. This assessment considers non-food exposure pathways in the UK. The risk of exposure to CWD prions through non-food routes in the UK is largely dependent on the level of the infection within UK deer species. The exception to this are any risks to travellers to parts of the world where the infection is endemic (eg areas of the US and Canada) and risks from exposure to imported deer-based products sourced from these endemic areas (such as deer antler velvet supplements or pet food) CWD has not been detected within UK cervids to date. However, data to support its absence is limited currently and it remains necessary to accumulate evidence to determine our confidence in the absence of this disease. The risk of introduction of CWD into Great Britain from Norway has been considered elsewhere. This risk assessment assumes a pessimistic scenario where CWD is present in the UK.
Assessment of risk	Negligible if CWD is not present in the UK. Very low if CWD is present in the UK (accounting for uncertainties in the scientific understanding of the disease, its aetiological agent and its zoonotic potential)
Level of confidence in assessment of risk based on the quality of evidence available	Medium

Summary of Action(s) and Recommendation(s)	Previous UK risk assessments for CWD as a risk to human health (SEAC[1], 2005) concluded that: 1. There is no evidence that CWD (or BSE) is present in the UK cervid population. However, because only limited surveillance is conducted in the cervid population, a low level prevalence of CWD cannot be ruled out. It is recommended that further surveillance of TSEs in UK cervids is conducted. 2. There is no evidence of transmission of CWD to humans from consumption of meat from infected cervids. Although epidemiological and experimental data on potential transmission of CWD are extremely limited, they suggest that there may be a significant species barrier. It would be helpful if further studies were available assessing the potential species barrier for transmission to humans. 3. Although limited, there is no evidence CWD can be transmitted to cattle, sheep or goats by natural means. 4. In summary, it appears that CWD currently poses relatively little risk to human health, or to the health of cattle, sheep or goats in the UK. Nevertheless, as a risk cannot be excluded a watching brief should be maintained. These conclusions remain valid. It is however further recommended that, should CWD be detected in the UK: • occupations at increased risk of exposure are aware of the appropriate guidance on measures to reduce the risk of exposure to and transmission of prions • epidemiological investigation of newly diagnosed human CJD cases includes relevant occupations and exposure routes • ACDP continues to review evidence of zoonoses • awareness of the disease is raised to prevent exposure

[1] Spongiform Encephalopathy Advisory Committee 1990 - 2011

Figure 1: Non-food pathways for human exposure to CWD prions from deer in the UK



Risk assessment

Epidemiology of Chronic Wasting Disease

Chronic wasting disease (CWD) is a transmissible spongiform encephalopathy (TSE) affecting farmed and wild cervid species. The build-up of abnormal prion protein in the brain of affected animals leads to progressive neurodegeneration. The clinical signs are weight loss and behavioural changes, typically over a period of weeks or months, leading to death. CWD was first recognised in 1967 in North America. The disease has since spread and intensified among and between deer species across the USA and Canada and is considered endemic in the central part of the USA (Wyoming, Colorado and Nebraska). It has also been found in South Korea, introduced via infected animals imported from Canada in the 1990s.

In 2016, the first case of CWD in Europe was detected in a reindeer in Norway (origin unknown) (Benestad et al., 2016). By June 2018, 19 reindeer, 3 moose and one red deer with CWD have now been reported in Norway. The affected reindeer are found in the region of Nordfjella whilst the affected moose and red deer are found in separate geographic regions in Norway. In early 2018, CWD was detected in a moose in Finland. Investigations indicate that the strain of CWD is different between the species, with the form found in reindeer appearing similar to that seen in North American deer and elk species (VKM, 2017).

CWD is transmitted directly from animal to animal and indirectly from the contaminated environment. The disease has now been observed in several deer species within four genera (see table 1).

There is no evidence to suggest that CWD is present in UK cervids. However, supporting data are limited and the most common UK deer species (red deer and roe deer), if not all UK species, are expected to be susceptible (Balachandran et al., 2010; EFSA, 2017).

Table 1. Captive and wild ranging species infected by CWD (to date)

North America	South Korea	Norway	Finland
Mule deer (*Odocoileus hemionus*) Black-tailed deer (*O. h.columbianus*) White-tailed deer (*O. virginianus*)			
Wapiti/elk (*Cervus canadensis*) Rocky Mountain elk (*C. elaphus nelsoni*)	Elk (*C. elaphus nelsoni*) Red deer (*C.elaphus*) Sika deer (*C.nippon*)	Red deer (*C. elaphus*)	
Shira's moose (*Alces alces shirasi)*		Moose (*Alces alces*)	Moose (*Alces alces*)
		Reindeer (*Rangifer tarandus*)	

Note the term Elk refers to different species in north America and Europe.

Hazard Characterisation – non-food routes of exposure to CWD prions

The non-food routes of human exposure to CWD prion infectivity are:

- direct contact with infected tissues of deer
- indirect exposure through other species, that have consumed infected deer tissues
- indirect exposure through contact with contaminated environments

Distribution of CWD infectivity in deer tissues

CWD prion infectivity is widely distributed in the tissues of infected animals and can be detected from early after infection, through to death. As with other TSEs, the highest infectivity and most extensive deposition of abnormal prion is found in the central nervous tissues such as brain and spinal cord (WHO, 2010; Race et al., 2007). There are differences between North American deer and elk in the distribution and level of infectivity in peripheral tissues, with greater involvement of lymphoid tissues in deer (Race et al., 2007). CWD infectivity has been observed in a wide range of other tissues, including musculoskeletal tissues (Angers et al., 2006) and also in secretions and excretions (saliva, urine and faeces).

The incubation period is typically longer than a year (on average 2-4 years in white tailed deer, 1.5 to 3 years in elk) (Williams, 2005; Miller et al, 1998). After symptoms appear, the course of clinical disease to death usually occurs within 12 months.

CWD infectivity in the environment

CWD prions are shed into the environment by infected deer (via saliva, urine, faeces, placenta etc). These may accumulate in places where deer congregate such as around mineral licks, deer rubs and scrapes, and supplementary feeding points. Discarded gralloch (viscera) +/- offal from gutting deer at hunt sites includes infectious peripheral tissues. Many wild deer die from natural causes (starvation, disease, exposure, predation) and may seek out secluded places to die, and are not found. Carcases and remains of dead animals, including tissues with high concentrations of prions may be further spread into the environment by scavengers, although this will not increase the net loading of infectivity in the environment.
Soil, water and plants have all been proposed as potential environmental reservoirs for CWD prions. Previous risk assessments have examined risks from environmental contamination with infectivity from BSE infected cattle carcasses, the persistence of prions in soil and the risks to surface and ground water sources (Environment Agency 1997; Gale et al., 1998).

Prion proteins are extremely resistant and degrade very slowly in ambient conditions. Experimentally it is presumed that the hydrophobic properties of prions cause them to bind to solids and particulates. Then either dispersing as suspended solids via surface and ground water and becoming greatly diluted within the aqueous environment, or partitioning into the solid fractions of processes, such as slaughterhouse wastewater treatment, landfill leachate filtering, and sewage sludge treatment, which capture particulates. Further processes such as drinking water treatment or groundwater chalk aquifer barriers which remove particulates are also likely to remove the majority of infectivity from water.

The risk posed is likely to be higher for the solid waste fractions from any treatment plants or other wastewater processes. This material may be spread as soil improvers on pasture or on crops with sufficient time delays to decrease the viability of other pathogen types that may be present but which would not significantly affect any levels of prion proteins present.

Research into the duration of prion persistence in soil, using scrapie prions, has found that infected material buried for 3 years showed some reduction in infectivity over the period but remained infectious, demonstrated by intracerebral inoculation of Syrian hamsters (Brown and Gajdusek, 1991). A similar experiment, also using scrapie infected material, showed that after 29 months all hamsters exposed orally to soil became infected, as did 4/11 of those which were exposed to the aqueous soil solution (Seidel et al., 2007). CWD prions have also been shown to bind strongly to clay soil minerals in particular, where they remain infectious to experimentally inoculated animals (Johnson et al., 2006). There is thus the potential for prion infectivity to remain in soil for extended periods.

CWD has been detected in and on plant surfaces and in very low concentration in environmental surface water samples and water sampled from treatment sites in CWD endemic areas (Miedema et al. 2017; Nichols et al., 2009; Zabel et al., 2017).

Effect of cumulative exposures to low doses of prion infectivity

Prion transmissibility is affected by host susceptibility; prion strain characteristics; route of exposure and dose. Non-food exposure pathways involve infection at peripheral sites through inoculation, or ingestion. Free-ranging of deer allows for the possibility of a low level of prion infectivity spread over large geographical areas. The probability of transmission decreases at low doses following a single exposure (Fryer and Mclean, 2011). The effects of repeat exposure to low doses of prion infectivity have been examined using scrapie prions in hamsters and mice (Diringer et al., 1998; Jacquemot et al.,2005). These experiments have shown that repeated exposure to low doses, which singly would be assessed as sub-infectious, can cause disease. The effect was reduced with longer intervals between exposures. Statistical modelling concluded that although the risk of oral infection increases with repeated doses, it does so to a lesser degree than would be expected if challenges combine independently or in a cumulative manner (Gravenor et al., 2003).

CWD susceptibility of non-cervid animals which contact humans (and companion animals)

CWD has been transmitted experimentally by intra-cerebral inoculation to a range of other species including: sheep, goats, cattle, pigs, ferrets and domestic cats. Cats were also susceptible to oral inoculation of brain homogenate that had first been passaged through intracerebrally infected cats (Mathiason et al., 2013), pigs are susceptible to direct oral inoculation (Moore et al., 2017). Horses, dogs and rabbits are considered to have low susceptibility to TSEs. Although there is now experimental evidence, using transgenic mice overexpressing rabbit prion protein, that the species barrier for rabbits can be breached by some prion species, CWD prions did not transmit (Vidal et al., 2015).

The potential risk from companion animals was suggested by the development of feline spongiform encephalopathy (FSE) in dozens of domestic cats in the UK, linked to BSE contaminated cat food. FSE is a notifiable disease in the UK but has not been reported since 2001. It has not been associated with onward transmission to humans. Dogs appear to be resistant to TSEs. However, pets and domesticated animals could potentially be exposed to new prion strains, should these become established and undetected, via several possible routes. For example, venison is used as an ingredient in both cat and dog food (including products manufactured in Canada and the USA), pet supplements containing deer antler velvet are sold, and fallen stock can be fed to fox hounds.

Zoonotic potential of CWD to infect humans

A species barrier is evident (Waddell et al., 2017). Experimental attempts to transmit mule deer CWD prion to transgenic mice overexpressing human prion protein did not succeed (Sandberg et al., 2010). However, there is some preliminary evidence emerging from ongoing studies by the University of Calgary that some macaques developed prion disease after being fed meat containing CWD prions (Czub, 2017). In vitro experiments have also shown that it is possible to convert human prion protein following exposure to CWD prions (Barria et al., 2014 & 2017) so the species barrier is not absolute. There is no epidemiological evidence of human prion disease associated with consumption or exposure to deer in CWD endemic parts of the USA, although it is highly likely that there has been human exposure to infected tissues and environments via food and non-food routes, particularly among hunters (Belay et al,. 2001 & 2004). An apparent increase in overall reporting of detected CJD cases within the endemic states in the USA has been attributed to increased detection and strengthened surveillance of prion disease over the same period.

Surveillance and detection of human prion disease

Detecting CWD associated human prion disease, should it occur, may be difficult. Diagnoses of human prion disease are made on a clinical basis and confirmed post mortem by pathology. The experience of variant CJD suggests that any associated illness could present years or decades following exposure and the link to CWD exposure may not be apparent. The UK CJD surveillance system, which follows up every suspected case of CJD with detailed assessments, would be well placed to detect any unusual patterns in CJD diagnoses or risk factors reported, including occupational or dietary risks. It would also be sensitised to the possible risk, should CWD exposure become a reality in the UK (NCJDRSU, 2017).

Subgroups potentially exposed to CWD prions through non-food exposures (see figure 1)

Figure 1 illustrates pathways through which individuals handling deer within the UK could come into direct contact with CWD infected tissues.

Direct contact with infected tissues of deer

Occupational

There are a number of occupations that routinely or occasionally have direct contact with cervid tissues which could therefore have a theoretical risk of transmission. If CWD can be transmitted like this, then it is most likely to occur via inoculation or introduction of contamination in the following ways:

- contaminating wounds and open lesions on the skin
- contaminating an inoculation injury of intact skin (ie via new cuts from knives, sharp instruments or bone fragments)
- splashing into mucous membranes (eyes and mouth)
- ingestion

The following occupations which could involve handling TSE infected animals, their carcasses or tissues, are identified in the HSE BSE occupational guidance (HSE, 2007):

- abattoir staff/slaughterhouse workers
- farmers
- veterinary surgeons
- hauliers
- knackermen
- boning plant operators
- renderers
- butchers
- workers in zoos and circuses
- workers handling meat and bonemeal
- incinerator operators
- landfill site workers
- cleaning and waste disposable workers
- maintenance engineers (eg in abattoirs, rendering plants, incinerators)

Additional occupations (not already covered by the guidance)

- taxidermists
- (various) handling injured deer/carcases following vehicle collisions
- gamekeepers
- conservationists/wildlife researchers
- hunters

Hunters

Hunters are a group considered at particular risk of exposure in endemic areas of the USA and Canada, and would be subject to similar risks of exposure if CWD were to become established in the UK. They may be repeatedly exposed through direct handling of carcases while gralloching/field dressing and undertaking butchery, as well as consumption of venison and offal and lower level environmental exposure.

Deer vehicle collisions

Several thousand deer vehicle collisions occur in the UK each year (Langbein, 2010) and there is evidence that CWD infection can increase the risk of deer being involved in collisions (Krumm et al., 2005). There is the potential for direct exposure to infected

tissues of injured animals/carcases by members of the public and those dealing with and removing injured or dead deer.

Laboratory staff working in diagnostic and research facilities

These staff may be regularly exposed to the highest infectivity tissues through diagnostic testing and research work.

Bodybuilders

Supplements containing deer antler velvet are readily available to UK consumers on the internet. Provenance is not always stated but some are sourced from north American farmed deer. Although largely targeted at body builders and athletes, other health and cosmetic/beauty benefits are claimed. The amount of deer antler velvet in these products is likely to be very small although some products are described as concentrated. They are usually formulated as oral sprays or sub lingual drops. Injectable preparations have been available in the past, but appear not to be available currently.

Indirect exposure through other species that have consumed infected deer tissues

Risks to farmers: Goats, sheep and cattle are not readily susceptible to natural infection with CWD. Pigs have been infected through oral exposure. The risk of CWD through intermediate contact via these animals remains unlikely. Other species specific TSEs are infrequently detected in ruminant animals in the UK and remain notifiable diseases.

Risks to pet owners: Evidence to date is that CWD transmits experimentally to cats but less efficiently than BSE, and that dogs, horses and rabbits will have low susceptibility.

Indirect exposure through contact with contaminated environments

Potential pathways of exposure to humans are via contaminated soil, surface water, ground water and run off, plant surfaces and other surfaces (such as mineral licks, deer rubs and scrapes). Accidental inoculation with or ingestion of these materials and surfaces are theoretical, but unlikely, routes of exposure. The effects of dispersal and dilution of prions within the environment suggest that dose exposures would be very low, at least for an extended period after initial incursion and spread within the UK deer population.
Other theoretical pathways to humans are through vegetables grown on soils spread with sewage sludge, including abattoir waste water or processed carcase material used as soil improver/fertiliser. Existing barriers include dilution of prion infectivity within the

soil and removal of the soil before consumption (Gale and Stanfield 2001). Most vegetable crop growers do not grow ready-to-eat crops on soil treated with sludge.

Mitigation

Acquired prion disease in humans (variant CJD from BSE contaminated beef, kuru and iatrogenic CJD via human to human routes) has occurred after inoculation, implantation or ingestion of contaminated tissues. Transmission in this way is rare and the conditions under which it will occur are not fully understood. Steps can be taken to minimise exposure to known or potential sources of prion contamination.

Guidance exists for occupations handling BSE infected livestock, carcases and laboratory samples, and can be extended to other occupations identified as at risk. The guidance aims to reduce the risk of inoculation, contamination of existing wounds and exposure of mucous membranes. (HSE 2007)

Guidance on laboratory safety when handling TSEs has been produced as part of the ACDP guidance (Department of Health, 2003), in which CWD is already included.

Advice on safe handling of carcases is already available from several sources (such as The Deer Initiative best practice guides) to ensure food safety and hygiene practices and protect hunters from exposure to other known pathogens carried by deer, for example TB. In CWD endemic areas in North America, hunters' CWD specific advice includes wearing gloves when handling carcases, avoid contacting with the brain and spinal cord, and washing knives and equipment thoroughly following use.

Given any indication of CWD within the UK deer population, Defra would be likely to bring in changes to the deer carcase tissues permitted in Category 3 materials (which may enter the environment as processed soil improver/fertiliser) and those categorised as Category 1 and therefore immediately incinerated.

Risk characterisation of human exposure to CWD deer via non-food exposures, should CWD become established in UK

Occupations with repeated contact with infected animals and direct handling of CWD infected or potentially infected tissues would have a higher risk of exposure than the general public. The latter could be exposed to CWD prions dispersed in the environment. Measures can be taken to prevent exposure to contaminated tissues and thus minimise the risk of transmission.

Level of uncertainty around the evidence in this risk assessment

Medium

The quality of the evidence is mainly good. However, there remains uncertainty in interpretation and fundamental gaps in our understanding of prion diseases and their properties. This suggests that new evidence will emerge that may change aspects of this risk assessment.

Guidance

Department of Health (2003). Transmissible Spongiform Encephalopathy Agents: Safe Working and the Prevention of Infection. Part 3. Laboratory Containment and Control Measures. Advisory Committee on Dangerous Pathogens. https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/209757/Part_3_-_Laboratory_containment_and_control_measures.pdf

HSE (2007) BSE – Occupational Guidance. Advisory Committee on Dangerous Pathogens: http://www.hse.gov.uk/pubns/web22.pdf

The Deer initiative England and Wales best practice guides http://www.thedeerinitiative.co.uk/best_practice/meat_hygiene.php
These include:

- carcass inspection
- carcass basic hygiene
- gralloching
- larder hygiene and safety

Risk assessments/position statements

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