



POSITION STATEMENT CHRONIC WASTING DISEASE IN UK DEER

Introduction

1. In 2004, the Food Standards Agency asked SEAC to consider the possible public and animal health implications of chronic wasting disease (CWD), in particular the level of risk posed to consumers of meat from infected animals. The committee also considered the possibility that BSE may be present in UK deer. The committee reconsidered these issues in 2006 in light of new information.

Background

2. CWD has emerged as an endemic transmissible spongiform encephalopathy (TSE) in certain captive and free-ranging species of cervid (deer) in some areas of North America. The disease is characterised by weight loss and behavioural changes in infected animals, usually over a period of weeks or months leading to death. CWD has not been found in the UK or elsewhere in Europe. No definitive or suspected cases of transmission of CWD to humans have been reported.
3. SEAC considered a review of the published, and some unpublished, research on CWD, together with surveillance data on TSEs in European cervids and information on UK cervid populations¹. A further review of the published literature was considered together with additional surveillance data on TSEs in European cervids².

Origins

4. The origins of CWD are unknown. On the basis of epidemiological data, it is highly improbable that CWD originated from the recycling of mammalian protein in processed feed. It has been suggested

¹ The information considered by the committee in 2004 is available at:
<http://www.seac.gov.uk/agenda/agen301104.htm>

² The information considered by the committee in 2006 is available at:
<http://www.seac.gov.uk/agenda/agen060706.htm>

that CWD may have arisen from transmission and adaptation of scrapie from sheep to cervids, as a result of a spontaneous change of endogenous prion protein (PrP) to an abnormal disease-associated form, or from an unknown source.

5. Data supporting any of these possible origins of CWD are either absent or equivocal. Although CWD could have originated from scrapie, the differing properties of the two prion diseases in strain typing bioassays, whilst limited, do not support this hypothesis. Evidence for multiple strains of CWD is equivocal. It seems most likely that CWD arose from a spontaneous change of endogenous PrP resulting in a disease-associated and laterally-transmissible form of PrP, although direct data to support this hypothesis are lacking.

Host range

6. The known natural hosts for CWD are mule deer (*Odocoileus hemionus hemionus*), black-tailed deer (*Odocoileus hemionus columbianus*), white-tailed deer (*Odocoileus virginianus*), Rocky Mountain elk (*Cervus elaphus nelsoni*) and moose (*Alces alces*). The prevalence and geographical distribution of CWD in these species appears to be increasing in North America in a manner which is unlikely to be due simply to increased surveillance.
7. There are no direct data relating to the transmissibility of CWD to UK cervid species. However, comparison of a limited number of PrP codons indicates some homology in the endogenous PrP gene of European and North American cervid species. Thus, the possibility that UK cervids may be susceptible to CWD cannot be excluded, in particular red deer (*Cervus elaphus elaphus*) which are closely related to Rocky Mountain elk.
8. There is no evidence to suggest that CWD is present in UK cervids. However, because surveillance in the UK is very limited, a low level prevalence of CWD cannot be ruled out. The committee endorsed the opinion of the European Food Safety Authority on CWD surveillance in the European Union (2004)³.
9. Transmission studies using parenteral routes of administration to cattle, sheep and a single goat, together with data from *in vitro* PrP conversion experiments, suggest that a significant barrier to CWD transmission to these species may exist. No transmission has been evident so far in an on-going oral transmission study in cattle

³ http://www.efsa.eu.int/science/biohaz/biohaz_opinions/501_en.html

after seven years. However, evidence from transmission experiments in cattle using the intracerebral route suggests that should cattle ever become infected with CWD, the barrier to transmission between cattle would be appreciably lower. In addition, these experiments show that the neuropathology of CWD is very different from BSE allowing CWD to be distinguished from BSE should natural transmission of CWD ever occur. Furthermore, no signs of infection have been observed from monitoring of cattle co-habiting areas with infected cervids, or in cattle, sheep or goats in close contact with infected cervids in research facilities. Thus, although the data are limited, there is currently no evidence to suggest that CWD can be transmitted naturally to cows, sheep or goats, and it is likely that there is a strong species barrier to such transmission.

Routes of transmission

10. Epidemiological data indicate that lateral transmission between infected and susceptible cervids occurring naturally is sufficiently effective to maintain epidemics in both captive and free-living populations. There is good evidence from studies of cervids inhabiting paddocks previously inhabited by infected animals or contaminated with infected carcasses, that CWD can be transmitted laterally between animals via the environment. The precise mechanism of transmission is unclear. It is possible that the infectious agent is shed in the saliva, faeces or urine or as a result of decomposition of infected carcasses and transferred to other cervids grazing the contaminated areas. It is also possible that some maternal transmission occurs.
11. There have also been suggestions that the lateral transmission of CWD may be influenced by environmental factors.

Pathogenesis

12. Information on the pathogenesis of CWD is limited. The data show that, following oral challenge, PrP^{CWD} is first detected in the oral and gut-associated lymphoid tissues before spreading more widely within the lymphoid system and then to the brain. However, involvement of the retropharyngeal lymph nodes or tonsils in the pathogenesis may not occur in some elk. At the microscopic level, the nature and distribution of the tissue lesions are similar to those found for scrapie. The available data suggest the pathogenesis of CWD is similar to scrapie. CWD infectivity has been detected in the muscle of mule deer.

BSE in UK deer

13. Both captive and free-ranging cervids in the UK may have been exposed to contaminated feed prior to the reinforced mammalian meat and bone meal ban instituted in 1996. A study to look at the potential susceptibility of red deer to BSE has shown no signs of transmission of the disease by the oral route, but this study is still ongoing. Although a theoretical possibility exists, there is no evidence from the very limited surveillance data to suggest that BSE is present in the UK cervid population.

Human health implications

14. Epidemiological data on possible CWD infection of humans are very limited. The possibility that clinical symptoms of CWD in humans differ from those of Creutzfeldt-Jakob Disease (CJD) cannot be excluded. There is no significant difference between the prevalence of CJD in CWD endemic areas and other areas of the world. However, because CJD surveillance in the USA is relatively recent, not all CJD cases may have been identified. Additionally, detection of a small increase in prevalence of such a rare disease is very difficult. Investigation of six cases of prion disease in young people (< 30 years of age) in the USA found no definite causal link with consumption of venison from known CWD endemic areas. The disease characteristics in these cases were indistinguishable from sporadic CJD or Gerstmann-Sträussler-Scheinker syndrome. Likewise, in a study of three hunters (> 54 years of age) diagnosed with sporadic CJD, no link with consumption of venison from CWD endemic areas was found. No causal link was found in an investigation of three men with neurological illnesses who were known to partake in "wild game feasts". Only one of these subjects was found to have a prion disease and this was also indistinguishable from sporadic CJD.
15. No study has examined the transmission of CWD to non-human primates by the oral route. However, CWD has been transmitted by intracranial inoculation to non-human primates. Transmission experiments using two strains of transgenic mice expressing human PrP, show that these animals do not develop CWD, suggesting a significant species barrier to the transmission of CWD to humans exists. However, these findings must be interpreted with caution as they may not accurately predict the human situation. Data from *in vitro* experiments on conversion of human PrP by disease-associated forms of PrP, including PrP^{CWD}, are equivocal.

16. The committee concluded there is no evidence of transmission of CWD to humans from consumption of venison, and that there may be significant barriers to transmission. Nevertheless, as the data are extremely limited a risk cannot be ruled out should CWD enter UK herds.

Conclusions

17. 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.
18. 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.
19. Although limited, there is no evidence CWD can be transmitted to cattle, sheep or goats by natural means.
20. 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.

SEAC

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