

103RD MEETING OF THE SPONGIFORM ENCEPHALOPATHY ADVISORY COMMITTEE

The Spongiform Encephalopathy Advisory Committee held its 103rd Meeting in London on 24th November 2009, and discussed the following:

CURRENT ISSUES

SEAC was informed about:

- Confirmation of BSE in a Scottish goat originally diagnosed with scrapie in 1990; the goat culled in 1990 was born prior to the introduction of the ruminant feed ban and could have been exposed to contaminated feed.
- The first case of atypical scrapie discovered in a sheep from New Zealand.
- A recommendation to Health Ministers by the Advisory Committee on the Safety of Blood Tissues and Organs that prion filtration should be introduced for blood used to treat patients who were not exposed to BSE through their diet.
- Recently published research¹ on the discovery in Papua New Guinea of a novel protective prion protein variant.

CJD UPDATE

SEAC was updated on the number of clinical cases of variant Creutzfeldt-Jakob Disease (vCJD) and sporadic CJD (sCJD).

To date there have been 170 definite or probable UK cases of clinical vCJD. 167 of these are associated with probable dietary exposure to BSE and three arose after transfusions of blood from donors who later developed vCJD. The mean age of death was 30 years. There have been no cases of vCJD in individuals born after 1989.

¹ A Novel Protective Prion Protein Variant that Colocalizes with Kuru Exposure by S Mead, J Whitfield, M Poulter, P Shah, J Uphill, T Campbell, H Al-Dujaily, H Hummerich, J Beck, C A Mein, C Verzilli, J Whittaker, M P Alpers & J Collinge. *New Eng J Med.*, 2009, **361**, 2056-65.

Elsewhere in the world, 47 vCJD cases have been reported: 25 in France, five in Spain, four in the Republic of Ireland, three in both the USA and the Netherlands, two in Portugal and Italy and single cases in Canada, Saudi Arabia and Japan.

A brief report was provided on the novel human disease known as Protease-Sensitive Prionopathy (PSP_r). Eleven cases have been reported with a mean age of onset of 62 years and disease duration of 20 months. The committee thought it was important that the unique pathology of this disease be more widely recognised to enable future diagnosis and tissue collection during autopsy procedures. SEAC will keep a watching brief on emerging data which may characterise this disease further.

EFFECT OF AGE ON THE PATHOGENESIS OF TSEs

Most clinical cases of variant CJD have occurred in young adults, the median age at onset of disease being 26 years and the median age at death 28 years (the comparable ages for sporadic CJD are both 67). The reasons behind this apparent age-related susceptibility are uncertain. A recent paper² reports the findings of a study in mice, which suggest that the age related decline in the functioning of follicular dendritic cells might impair TSE pathogenesis. The Committee agreed that a competent immune system was required for efficient replication of TSEs in the host. It was noted that older people are more immuno-compromised than the young, but there is very little data on the effect of ageing on the human immune system. The Committee concluded that there are insufficient data to suggest that this might provide an explanation for the young age of vCJD patients.

UP-DATE ON vCJD PREVALENCE STUDIES

The committee received an update on progress with the vCJD prevalence studies being conducted by the Health Protection Agency. This covered the National Anonymous Tonsil Archive (NATA), the post mortem archive and a new study of appendices. To date, NATA has tested approximately 80,000 pairs of tonsils and none were positive. A pilot for a study of spleens obtained

² The effects of host age on follicular dendritic cell status dramatically impair scrapie agent neuroinvasion in aged mice. K L Brown, G J Wathne, J Sales, M E Bruce, and N A Mabbott, the Journal of Immunology 2009, doi:10.4049/jimmunol.0802695.

from post mortems, that will assess a number of potential methodologies, will start in early 2010, and report on the efficacy of these methodologies in June 2010. The new study of 30,000 appendices, to be tested by immunohistochemistry, will also start in early 2010. The Committee noted progress, and commented that in order to have sufficient power, the spleen study would have to test approximately 50,000 samples.

CATEGORY 3 ANIMAL PRODUCTS IN FERTILISERS

Building on previous work in 2005, Defra has commissioned a full risk assessment, which SEAC was invited to consider, to evaluate the amount of potential infectivity available in the soil of non-pasture land following the application of Category 3-derived³ fertiliser.

The committee considered that the methodology used in the risk assessment was scientifically valid, but felt that too many worst case assumptions had been made in addressing the paucity of experimental data, that certain temporal aspects could have been addressed differently and that the possibility of regulatory failure had not been sufficiently considered.

FUTURE OPERATION OF SEAC

The Committee discussed the future organisation of SEAC business and the desirability of streamlining the decision making process, so that Ministers could obtain advice more quickly. In particular, SEAC considered a proposal that in future much of its business could be dealt with by correspondence, rather than in face-to-face meetings.

In discussion the committee recognised that some SEAC business could be carried out more efficiently by e-mail correspondence. The process would need to be structured, perhaps at fixed times of the year and the discussion would be summarised and recorded on the SEAC Website so that the decision-making process was transparent and available for public scrutiny. However, it was important that the committee still aimed to meet in person twice or three times per year.

³ Category 3 is low risk material, most of which is fit for human consumption, but not intended for human consumption.