

### **102<sup>ND</sup> MEETING OF THE SPONGIFORM ENCEPHALOPATHY ADVISORY COMMITTEE**

The Spongiform Encephalopathy Advisory Committee held its 102<sup>nd</sup> meeting in London on 4<sup>th</sup> March 2009, and discussed the following:

#### **UPDATE ON ANIMAL TSEs**

SEAC was updated on UK surveillance of transmissible spongiform encephalopathies (TSEs) in cattle, sheep and deer.

The Bovine Spongiform Encephalopathy (BSE) epidemic in cattle peaked in 1992, with over 37 000 confirmed cases, but has since declined with 37 cases confirmed by active and passive surveillance in the UK in 2008. Only four of these cases were found in healthy slaughter animals destined for human consumption.

Active surveillance of sheep over 18 months of age in Great Britain shows that the incidence rate of classical scrapie has declined from 0.2% in 2005 to below 0.05% in 2008. The incidence of atypical scrapie has remained relatively constant at around 0.07%, a similar level to several other European countries.

No TSEs have been found in relatively small surveys of deer in the UK and other European countries.

#### **NEW RESULTS ON IDIOPATHIC BRAINSTEM NEURONAL CHROMATOLYSIS**

SEAC considered recently published research<sup>1</sup> on Idiopathic Brainstem Neuronal Chromatolysis (IBNC), a neurodegenerative disease of cattle.

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<sup>1</sup> Jeffrey *et al.* (2008) Idiopathic Brainstem Neuronal Chromatolysis (IBNC): a novel prion protein related disorder of cattle? *BMC Vet. Res.* 4, 38.

SEAC noted that IBNC appeared to be a rare disease that occurred in older cattle, predominantly as single cases, although it is possible that surveillance may not detect all cases. Biochemical studies suggested that the prion protein may play a role in the disease. However, it is unclear whether the normal form of the protein or an abnormal form is involved. Studies are required to determine whether IBNC is transmissible or not.

SEAC concluded, noting that specified risk material controls are in place to prevent cattle brain from entering the food supply, that current data on IBNC do not suggest it presents a risk to human health.

## **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 168 definite or probable cases of clinical vCJD in the UK with 165 of those from dietary exposure to BSE and three from infection via transfusions of blood from donors who later developed vCJD. The mean age at death of the cases associated with dietary exposure to BSE was 28 years. There have been no cases of vCJD in individuals born after 1989.

Elsewhere in the world, 44 vCJD cases had been reported: 23 in France, five in Spain, four in the Republic of Ireland, three in both the USA and the Netherlands, two in Portugal and single cases in Canada, Saudi Arabia, Italy and Japan. In two Irish and two USA cases, one French, Japanese and Canadian cases, infection was presumed to have occurred during a period of residence in the UK.

From May 1990 to January 2009, 1027 cases of sCJD had been identified in the UK, with a mean age at death of 67 years.

## **COMPARING THE RELATIVE RISK OF vCJD TRANSMISSION VIA PLASMA**

The Department of Health asked SEAC for advice on a methodology for assessing the risks of using single unit plasma as opposed to pooled plasma, either sourced from the UK or non-UK source countries.

SEAC noted that there are many large uncertainties around the potential risk of transmission of vCJD via the use of plasma products. However, as the relative risks (as opposed to absolute risks) posed by plasma products were being estimated, uncertainties around the timing, level and distribution of infectivity in blood of an infected person would not appreciably affect the estimations. The best way to manage other major uncertainties, such as those around the prevalence of vCJD in the UK and other countries, would be to develop a range of scenarios incorporating reasonable high and low value estimates for such parameters.

### **vCJD INFECTION IN A HAEMOPHILIAC AT POST MORTEM**

SEAC considered data from investigations of a Haemophilia patient who had been shown on post mortem to have the abnormal prion protein associated with vCJD in his spleen (as reported recently by the Health Protection Agency<sup>2</sup>). In view of the fact that preliminary unpublished data were considered, this issue was discussed in a reserved business session in accordance with the SEAC Code of Practice.

SEAC agreed that, although the patient had not shown clinical signs of vCJD prior to death, this finding provides evidence of vCJD infection. It would appear more likely at this stage that the infection occurred from the administration of clotting factors prepared from the plasma of a donor who had later developed vCJD than from dietary exposure to BSE.

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<sup>2</sup> Asymptomatic vCJD abnormal prion protein in a haemophilia patient  
[http://www.hpa.org.uk/webw/HPAweb&HPAwebStandard/HPAweb\\_C/1195733818681?p=1225960597236](http://www.hpa.org.uk/webw/HPAweb&HPAwebStandard/HPAweb_C/1195733818681?p=1225960597236)