

### **ONE HUNDRED AND FIRST MEETING OF THE SPONGIFORM ENCEPHALOPATHY ADVISORY COMMITTEE**

The Spongiform Encephalopathy Advisory Committee held its 101<sup>st</sup> meeting in London on 15<sup>th</sup> October 2008, and discussed the following:

#### **CURRENT ISSUES**

SEAC was informed about:

- A mother and son in Spain who had died of variant Creutzfeldt-Jakob Disease (vCJD). This is the first recorded instance of more than one case of vCJD within one family. Both the mother and son lived in a region of Spain with a history of BSE and had frequently shared meals of cattle brain. As no other risk factor has been identified, it seems most likely that both infections were acquired from dietary exposure.
- Results of tests on a single goat from a culled UK dairy herd with a large classical scrapie outbreak. On the basis of the results the presence of Bovine Spongiform Encephalopathy (BSE) cannot be excluded. Further testing by mouse bioassays, which may take at least two, if not more, years to complete, is required to make a definitive diagnosis.

#### **UPDATE ON vCJD PREVALENCE STUDIES**

SEAC was updated by the Health Protection Agency (HPA) about the progress of the National Anonymous Tonsil Archive (NATA), a proposed second retrospective survey of 30 000 stored appendix samples and a proposed post mortem tissue archive. These studies would provide data to estimate the prevalence of

subclinical vCJD (vCJD infections that have yet to develop, or may never develop, into clinical disease).

Around 62 500 tonsil samples collected by NATA have been tested with no positive samples found. An application for the second retrospective survey of appendix samples is currently under consideration by a Research Ethics Committee. SEAC learned that the establishment of a post mortem tissue archive, which is dependent on the collection of samples from Coroners' autopsies, does not have the support of Coroners needed to take it forward.

SEAC is extremely disappointed about the lack of support from Coroners for the post mortem tissue archive. As SEAC has repeatedly stated, the archive is key to obtaining better estimates of the prevalence of subclinical vCJD. These estimates are vital to make meaningful assessments of the risks to public health from vCJD and of the effectiveness of current, and the need for further, very costly public health protection measures. SEAC acknowledged the strenuous efforts made by the HPA, the Department of Health (DH) and National Health Service Blood and Tissue to devise a system to collect samples that would have the least impact on the work of Coroners. SEAC remains strongly in favour of establishing the archive.

## **PROPOSALS TO REDUCE TESTING OF CATTLE SLAUGHTERED FOR FOOD – IMPACT ON RISK TO HUMAN HEALTH**

The Food Standards Agency (FSA) asked SEAC to consider an analysis of the human health risk from increasing the minimum age at which cattle must be tested for BSE. SEAC agreed the following conclusion which was provided to the FSA Board:

*SEAC considered the results from a mathematical model that had been used to estimate the number of infected cattle that may be undetected as a result of raising the minimum age at which healthy slaughtered and fallen stock cattle must be tested for BSE. The model itself, produced by VLA, was previously reviewed by SEAC<sup>1</sup>.*

---

<sup>1</sup> Reviewed at SEAC 95, December 2006.

*The increased risks to human health estimated by the model from raising the age at which healthy slaughtered cattle are tested for BSE (up to 60 months, the highest age modelled) are very small. The model estimates that much less than one BSE case would be missed annually in the GB herd by increasing the age of testing to 60 months for the healthy slaughter surveillance stream. Although uncertainties are inherent in such modelling, the validation of the model that has been conducted provides assurances about the reliability of the results. Similar results from a different analysis by EFSA<sup>2,3</sup> provide additional confidence in the findings. The EFSA analysis estimates that less than two BSE cases would be missed annually in the whole of the EU15<sup>4</sup> by increasing the age of testing to 60 months for the healthy slaughter surveillance stream.*

*These risk assessments hold provided the incidence of BSE in cattle remains low. Therefore, regulations should not be modified unless effective surveillance remains in place. Surveillance is the only means of monitoring changes in the incidence and prevalence of BSE, the effectiveness of control measures in preventing an epidemic and the possible emergence of new prion diseases. As control measures to prevent cattle and human infection are modified, continued active and passive surveillance become increasingly important to ensure that the remaining controls are effective in minimising the risk to human and animal health.*

## **RISK ASSESSMENT OF SRM CONTROLS AT ABATTOIRS AND CUTTING PLANTS**

FSA asked SEAC to review a mathematical model produced by Det Norske Veritas Consulting to estimate the human exposure to BSE in the food supply under various enforcement regimes at abattoirs and cutting plants.

SEAC was content with the methodology, noting that some of the assumptions made in the modelling relied upon expert judgement rather than data, and that there is uncertainty about some

---

<sup>2</sup> Risk for Human and Animal Health related to the revision of the BSE Monitoring regime in some Member States. Scientific Opinion of the Panel on Biological Hazards. The EFSA Journal (2008) 762, 1-47.

<sup>3</sup> Further consideration of age-related parameters on the Risk for Human and Animal Health related to the revision of the BSE Monitoring regime in some Member States. Opinion of the Panel on Biological Hazards. The EFSA Journal (2008) 763, 1-8.

<sup>4</sup> Austria, Belgium, Denmark, Finland, France, Germany, Greece, Ireland, Italy, Luxembourg, Netherlands, Portugal, Spain, Sweden, United Kingdom.

parameters such as the infectivity of peripheral nervous tissues. SEAC considered that the risks to human health estimated for different enforcement regimes are very low provided the prevalence of BSE remains low. As with all modifications to disease control measures, it is important that risk managers consider any consequent, and potentially detrimental, impact on the effectiveness of other controls.

### **ATYPICAL SCRAPIE CASE-CONTROL STUDY**

SEAC considered a case-control study by the Veterinary Laboratories Agency to identify potential risk factors associated with atypical scrapie. The committee concluded that much more work on the study is needed before conclusions can be drawn from the data and the analysis presented.

### **DRAFT SEAC STATEMENT ON MODIFICATIONS TO THE TOTAL FEED BAN**

SEAC considered a draft statement of its assessment of the possible implications of various modifications to the total feed ban. These modifications include the introduction of tolerance levels for certain types of processed animal protein (PAP) in feed, the inclusion of fish meal in young ruminant diets and the feeding of non-ruminant PAP to non-ruminants of a different species. The statement was agreed with minor modifications for publication on the SEAC website

<http://www.seac.gov.uk/statements/feedban-oct08.pdf>

### **ALTERNATIVE HYPOTHESIS – AN AUTOIMMUNE ORIGIN FOR PRION DISEASES**

The Chief Medical Officer for England asked SEAC to consider the plausibility of a hypothesis proposed by Professor Alan Ebringer (King's College London) that prion diseases may be the result of an autoimmune reaction initiated by exposure to certain bacteria. The committee had considered the hypothesis in the past, concluding then that it was not supported by the evidence available.

SEAC noted that there are no new published data to support the hypothesis. Indeed, SEAC knew of at least one study involving

Professor Ebringer designed to test the hypothesis, which produced results that did not support his hypothesis and that has not been published.

## **PROTEASE SENSITIVE PRIONOPATHY**

SEAC discussed with Dr Pierluigi Gambetti (US National Prion Disease Pathology Surveillance Center) his recently published report<sup>5</sup> on the identification in the United States of America of a new human prion disease. SEAC agreed that there is considerable work to be done to characterise fully this new disease, its cause and whether it is infectious or not. As preliminary unpublished data were also presented, this issue was discussed in a reserved business session in accordance with the SEAC Code of Practice.

## **RESULTS ON HUMAN SCLERA**

SEAC considered preliminary results provided by the HPA and National CJD Surveillance Unit from tests on eye tissue (sclera) from a vCJD case. The results suggest the presence of infectivity and, in contrast with previous testing of samples from other vCJD cases, abnormal prion protein in this tissue. However, as the sclera is very difficult to remove from surrounding eye tissues, which are themselves known to carry vCJD infectivity, the findings may have arisen as a result of contamination at autopsy. Nevertheless, even if the data are reliable, they indicate that there may only be a relatively low level of infectivity present in sclera. As preliminary unpublished data were considered, this issue was discussed in a reserved business session in accordance with the SEAC Code of Practice.

---

<sup>5</sup> Gambetti *et al.* (2008) A novel human disease with abnormal prion protein sensitive to protease. *Ann. Neurol.* 63, 697-708.