Network on BSE-TSE
Minutes of the 13th meeting

Held on 15-16 October 2018, Parma

(Agreed on 7 December 2018)

Participants

- **Network Representatives of Member States (including EFTA Countries):**

Country	**Name**[1]
Austria	Hermann Schildorfer
Belgium/Luxemburg	Stefan Roels
Bulgaria	Sylvia Peeva
Croatia	Branko Sostaric, Karmen Branovic
Cyprus	Penelope Papasavva-Stylianou
Czech Republic	Pavel Vodrážka
Denmark	Tim Kaare Jensen
Estonia	Maarja Kristian
Finland	Maria Hautaniemi
France	Thomas Maignien
Greece	Vaia Palaska, Maria Christopanou
Hungary	Zsuzsanna Baraczka
Ireland	Justin Byrne
Italy	Giuseppe Ru, Romolo Nonno
Latvia	Madara Stinka
Lithuania	Kristina Stakyte
The Netherlands	O.F.J. (Olaf) Stenvers
Poland	Miroslaw Pavel Polak
Portugal	Renata Carvalho
Romania	Theodora Chesnoiu Vasile
Slovakia	Martin Mojzis
Slovenia	Ivan Ambroži, Polona Juntes
Spain	Soledad Collado Cortés
Sweden	Maria Nöremark
United Kingdom	John Spiropoulos
Norway	Michael A. Tranulis
Iceland	Sigrun Bjarndottir
Switzerland	Torsten Seuberlich

1 Indicate first full name and them surname (John Smith) all throughout the document

- **Hearing Experts**

Gabriele Vaccari (Department of Food Safety, Nutrition and Veterinary Public Health, Istituto Superiore di Sanità, Italy), for point 6.1.

Amie Adkin (Animal and Plant Health Agency, UK) for points 6.2 and 6.3.

European Commission:

Sara Perucho Martinez (DG-SANTE)

- **EFSA:**

BIOCONTAM Unit (Pietro Stella – chair; Angel Ortiz Pelaez – secretariat; Yves Van der Stede)

- **Others:**

Fernanda Mejia Salazar (OIE)

15 October 2018

1. Welcome and apologies for absence

Pietro Stella, the Chair, welcomed the participants. Apologies were received from Martin Groshup (Germany). Due to flight delays, the representative of Bulgaria (Sylvia Peeva) was not present at the beginning of the meeting but joined during day 1.

2. Adoption of agenda

The agenda was adopted without changes.

3. Review the minutes of the 12th meeting of the Network on BSE-TSE held on 16-17 October 2017, Parma.

The minutes were agreed by written procedure on 4 December 2017 and published on the EFSA website on 14 December 2017.

4. Topics for discussion

4.1. Round-the-table on activities of Network Members in the TSE field since the last meeting

The Members of the Network provided an update on TSE-related scientific activities, with special interest on Chronic Wasting Disease (CWD), including risk assessments and other initiatives, which had been undertaken in their respective countries since the previous Network meeting. The six members conducting compulsory surveillance on CWD were encouraged not to provide any update on this topic since there was a specific session for that. Switzerland reported the difficulty to score enough points in order to comply with the OIE requirements given the lack of clinical suspects. Extra efforts have been made to encourage farmers to report cases. Similar problem was reported by Sweden. Spain informed of studies conducted by the national reference laboratory (NRL) and

presented as posters in the last PRION conference. Slovakia, Romania and Portugal reported the number of cases of scrapie in their populations and the lack of cases of BSE or CWD. Poland is validating the rapid test for CWD using positive material from Canadian cases. Italy reported a decline in the number of cases in the first half of 2018 and the ongoing research studies on the zoonotic potential of scrapie and the impact of the genotype in the incidence of the disease in goats using the 222K resistant allele. Lithuania, Latvia and Estonia focused their efforts this year in the CWD surveillance plans while complying with the testing requirements for cattle and small ruminants. Ireland has confirmed one case of atypical scrapie and one case of classical scrapie, and informed of the publication of a paper in The Veterinary Record on the BSE cases detected in 2015. Iceland stressed the issue of the notification of clinical suspects in cattle for the OIE surveillance points, the lack of individuals in a local sheep breed holding resistant alleles and the cull of infected flocks following two classical scrapie cases, one in 2017 and one in 2018. Hungary detected 12 atypical cases so far this year out of approximately 15,000 tested. They intend to increase the number. Greece had 5 clinical suspects of BSE in 2017 and also highlighted the scarce notification by farmers of this type of animals. So far they have conformed 8 index cases of classical scrapie and one atypical. France has confirmed two atypical BSE cases so far in 2018 (L type). ANSES has published a report on the efficacy of cleaning and disinfecting products for TSE in relation to the environmental contamination of different strains for which there is no information (atypical BSE, atypical scrapie, BSE in small ruminants). Finland mentioned the positive case in one wild moose which is being characterised in laboratories of Italy and the USA. Denmark, Croatia, Slovenia and Austria reported routine surveillance with no cases and Belgium informed the awareness training for vets on TSE being conducted. The country obtained the OIE negligible risk status in 2012 and surveillance is based on risk animals and clinical suspects. Croatia is delivering a questionnaire to hunters to collect information on knowledge on CWD. The United Kingdom (UK) reported a new case of BSE detected in Scotland for which final classification was still undetermined. The UK member was also representative of the TSE EURL and reported the results of bioassays of two scrapie cases from Spain and the confirmation of the results: CH1641 strain was identified in both cases, excluding BSE. Despite the initial pattern undistinguishable from BSE of the first case in red deer, it has now been excluded following additional tests.

4.2. TSE activities of EFSA BIOHAZ Panel and BIOCONTAM Unit

Angel Ortiz Pelaez from the EFSA BIOCONTAM Secretariat presented the EFSA activities on TSE completed and ongoing since the 2017 Network meeting. Completed activities include two Scientific Opinions of the Scientific Panel on Biological Hazards (BIOHAZ) of EFSA: on CWD in cervids (Part II on diagnostic of CWD and occurrence of CWD in Europe) (EFSA Panel on Biological Hazards (BIOHAZ), 2018a) and on the request for a scientific opinion on an updated Quantitative Risk Assessment of the BSE risk posed by Processed Animal Proteins (PAP) (EFSA Panel on Biological Hazards (BIOHAZ), 2018b). Ongoing activities of the BIOHAZ Panel include one mandate for a Scientific Opinion on a request for an update of the scientific opinion on Chronic Wasting Disease (CWD) in cervids by reviewing the state of knowledge about the different strains

present in Europe, the zoonotic potential and identifying risk factors for the spread of the disease within the EU.

4.3. Update on CWD in Norway

Michael Tranulis, representative of Norway updated the Network on the epidemiological situation of CWD in the country. At the time of the Network meeting, 23 cases of disease in reindeer (19) and moose (3) and red deer (1) have been so far detected in Norway since April 2016. The cull of the entire population of wild reindeer in zone I of Nordjfella was conducted between November 2017 and May 2018 resulting in: 582 animals killed via enhanced regular haunting and 1442 via professional shooters. A five-year fallow period has started, including the installation of fences and physical barriers for cervids to access saltlick sites. An experimental study is being conducted whereby pre-colostrum lambs have been inoculated orally with NZ-Z1 strain (reindeer from Nordjfella). Multiple samples are being collected and clinical examination, rectal biopsy and full necropsy will be performed on the animals. Surveillance continues and some 19,470 cervids having been tested until mid-October 2018 although the target is to reach 30,000 now that the hunting season has started. The features of the CWD in the three species show differences in terms of lymph tropism, lesion profiles, age, PrP^{Sc} type and transmission to rodents. The representative stressed the importance of maintaining the vigilance and not lowering the guard in the fight against the disease.

4.4. CWD surveillance in Estonia, Finland, Latvia. Lithuania, Poland and Sweden: state of play

4.5.1 Estonia

Maarja Kristian, representative of Estonia, informed that the country has added the CWD surveillance into the National infectious animal disease control programme, informed zoos and places with captive cervids, signed contracts with hunters and have provided training for hunters using visual materials (video). They face logistical problems, financial to fund the surveillance and they need collaboration with Environmental inspectorate. In the next two years they will increase the budget and involve more hunters. A total of 75 cervids have been tested: 67 roe deer, 4 moose and 4 red deer.

4.5.2 Finland

Maria Hautaniemi, representative of Finland, informed that the country has defined 100 PSU: all reindeer herding association (54), 45 game management associations selected randomly and the Åland islands as the 100th PSU. The surrounding area where the moose case was found is subject intensified surveillance. So far 375 wild cervids have been tested: wild forest reindeer (10), moose (89, one positive), reindeer (197), roe deer (52) and white-tailed deer (27). In addition 7 farmed cervids have been tested. These animals have been tested in 28/54 herding associations, 16/45 selected and from non-selected 60 game management associations. Four samples from the Åland islands and 30 from the area of intensified surveillance have been also tested. The hunting season has just begun and more samples are expected during the next few months.

4.5.3 Latvia

Madara Stinka, representative of Latvia, informed that the CWD surveillance programme includes sampling in 100 PSU covering all the territory of the country a total of 1000 wild cervids (235 moose, 269 red deer, 496 roe deer) and 205 captive cervids (red deer) per year. Between January and August 2018 CWD passive surveillance was in place with 20 suspected wild cervids (2 moose, 9 red deer and 9 roe deer) and active surveillance has been in place between September 1st and October 10th, with 61 wild and 6 captive cervids having been tested, all negative. Out of the 61 wild cervids tested, 56 were hunted fit for human consumption. Current challenges include the mismatch between the territories of State Forest Service and the territories of Food and Veterinary Service; the small number of hunters clubs involved in the monitoring programme, the limited period for legal hunting and the lack of information among stakeholders.

4.5.4 Lithuania

Kristina Stakyte, representative of Lithuania, informed that the CWD surveillance programme requires samples to be collected by authorized veterinarians and/or official veterinarians and to be transported to the National State Food and Veterinary Risk Assessment Institute. Keepers, hunters, environment specialists and other persons must inform the veterinary services territorial unit about clinical suspects and /or found dead cervids. So far, a total of 345 wild cervids (13 moose, 65 red deer and 285 roe deer and 5 of other species) and 23 kept cervids have been tested.

4.5.5 Poland

Miroslaw P. Polak, representative of Poland, informed that the CWD surveillance programme includes the sampling of 100 PSU (corresponding with the number of farms keeping 6000 breeding cervids); 16 PSU corresponding to the administrative division of Poland into provinces in which 63 samples will be collected per year from each PSU. Samples collected on the farm, the rendering plant or at shooting sites are to be sent to the local veterinary surgeon or directly to the Veterinary Hygiene Unit for testing. So far a total of 461 cervids have been tested (428 roe deer, 16 red deer, 12 moose and 5 not identified). The moose population is under-represented whereas roe deer population is over-represented. The risk group prevails with 97% of all samples belonging to this group.

4.5.6 Sweden

Maria Nöremark, representative of Sweden, informed that the design of the CWD surveillance programme has defined each sami village as a PSU and a cervid farm a PSU. For free living cervids, PSU cover the entire country (not to waste fallen wild cervids dead outside a PSU). Sampling instructions have been issued, printed and online. Testing results aggregated at country level and can be followed via internet. Identified challenges are the surveillance sensitivity for different species, the farmed population is very small compared to the free living one, the logistics of collecting samples and the unpredictability of the occurrence of risk animals. Additional constraints are related to the stakeholders interest, the identification of animals sampling and the transport of samples to laboratories.

4.5. The zoonotic potential of TSE: the concept of species barrier. Latest research

Romolo Nonno, representative of Italy and senior scientist of the Istituto Superiore di Sanità, Rome (Italy) presented the basic concepts underpinning the zoonotic potential of TSE and the outputs of latest research with particular focus on European CWD. The species barrier can be defined as the inefficient transmission (or lack of transmission) on experimental challenge, followed by adaptation to the new species upon sub-passages and is determined by multiple factors like the recipient/host PrP homology or the strain. TSE strains are infectious isolates which cause specific phenotypes of disease, which persist upon serial transmission in the same host species and can be characterized using transmission in animal models since there are no molecular methods for direct identification of strains. The ability of a strain to cross the species barrier depends on the compatibility between the conformation of the aggregated PrP^{Sc} strain and the PrP of the recipient species. The zoonotic potential depends on the ability of the strain to convert human isoform prions in the host. The author developed an animal model to study the zoonotic potential of TSE and to characterise TSE strains, the bank vole (Myodes glareolus), susceptible to human, small ruminant, cervids and bovine prions. It has been applied to the study of the European CWD by inoculating seven CWD isolates from USA (Elk, Mule deer, White-tailed deer) and from Norway (reindeer, moose, red deer). Looking at the attack rates, incubation period, lesion profiles, PrP^{Sc} distribution in different anatomical areas and the Western blot (WB) pattern, the results show the presence of three different PrP^{Sc} types in Norwegian cervids, with further variation within the moose cases that are not due to the host effect. The complex WB patterns observed in Norwegian moose are reproduced in bank voles.

16 October 2018

5. Welcome and apologies for absence

Pietro Stella, the Chair, welcomed the participants. No apologies were received for day 2.

6. Topics for discussion

6.1 Camel prion disease

Gabriele Vaccari (Istituto Superiore di Sanità. Department of Food Safety, Nutrition and Veterinary Public Health, Italy) presented the newly described prion disease in dromedary camels. A recent publication by Babelhadj et al. (2018) jointly produced by researchers of the Istituto Superiore di Sanità and Algerian colleagues from the Ecole Normal Supérieure de Ouargla and the Laboratoire PPABIONUT Physiologie, Physiopathologie et Biochimie de la Nutrition, Université de Tlemcen, described a prion disease in camel slaughtered

at an abattoir in the city of Ouargla, in the centre of Algeria, a condition observed by a local veterinarian for the last five years. Three adult female camels showing clinical signs and one adult female apparently healthy were sampled and tested, confirming the diagnosis by detecting pathognomonic neurodegeneration and disease-specific PrPSc in brain tissues from dromedary camels and designate it as camel prion disease (CPD). A retrospective study of clinical suspects submitted for slaughter to the same abattoir indicated a 3.1% prevalence of animals showing neurological signs suggestive of the disease. The presentation of the disease suggests an acquired nature of CPD. The abundant extraneuronal pathogenesis may also suggest that CPD is a contagious disease. Molecular investigations show differences between CPD and BSE or scrapie however is not possible to exclude any potential link. Further studies are being conducted to characterize the prion strain of CPD and to ascertain the PRNP gene variability on dromedary populations. A case-control study in the region is also planned.

6.2 Recent studies on the analysis of BSE surveillance data

Amie Adkin (Animal and Plant Health Agency, UK) presented a summary of three recent publications analysing BSE surveillance data for the UK and the EU. In the first one, Adkin et al. (2016) applied the EFSA's Cattle TSE Monitoring Model (C-TSEMM) that allows the estimation of the underlying prevalence of BSE for each EU28 MS (detectable & infected). The application of the model to surveillance data for the period 2002 to 2011 showed that seven countries (Germany, Spain, France, Ireland, Italy, Poland and the UK) exceeded individually the required threshold the OIE for Type A surveillance. In the second study, Simons et al. (2017) estimated the time taken for a surveillance system to detect a re-emergence of BSE in cattle assuming a 10% rate of increase per year from 2012 onwards and one case required for detection testing different surveillance scenarios. The baseline of the current surveillance would require 15 years to detect the re-emergence, only two years less than the scenario in which only clinical were tested. Additional testing of younger animals does not improve the detection, nor does the additional testing of healthy slaughter animals. In the third study, Arnold et al. (2017), the C-TSEMM model was applied to compare the trend of classical BSE by birth cohort in different countries and to test the hypothesis of spontaneous occurrence. There were no significant difference in rate of decline between UK, Ireland and a group of other EU countries, with a decline per annual cohort of over 30% in all cases. There is no evidence of spontaneous occurrence since the applied model fits the data better without a constant rate different from zero. The predicted year of the final case of BSE in the UK would be around 2021.

6.3 Impact of changes in SRM on BSE infectivity in edible tissues (small ruminants and cattle)

Amie Adkin (Animal and Plant Health Agency, UK) presented a summary of a recent publication on the estimation of the impact on food and edible materials of changing BSE control measures. In Adkin et al. (2010) a new model (BSE control model) was described with the objective of comparing the level of infectivity in animal tissues and of estimating the total amount of infectivity

consumed in the food chain per year. The BSE control model was applied by Horigan et al. (2018) to estimate what effect any additional lingual tonsillar material from harvested tongues inadvertently entering the human food chain would have on the amount of infectivity being consumed. The results showed that with tonsils classed as SRM, a mean of 0.08 Bovine Oral (BO) ID_{50} was estimated to enter the food chain over one year for GB. Under negligible risk status this mean was estimated to increase to 0.83 $BOID_{50}$. This equates to a 10 fold increase in $BOID_{50}$ but the amount is still small due to the very low likelihood of infected animals being present in the 2017 healthy slaughter population. The same model was applied by Adkin et al. (2018) to assess the impact of the removal of Specified Risk Materials (SRM) from sheep on potential prion infectivity passed into the food and protein material chains. For classical scrapie, there is a mean estimate of infectivity of 30,000 Ovine Oral (OO)ID_{50} per year at abattoir, lower than for atypical scrapie (3,500,000 $OOID_{50}$ per year) due to the lower occurrence of this disease in Great Britain. However, more infectivity is destined to reach the food chain as the disease is peripherally distributed in the carcase. The highest contributor to the total amount of infectivity consumed per year is the intestines (duodenum and jejunum). If SRM removal is limited to the brain and spinal cord of sheep over 12 months of age, there is an approximate mean increase from 19,000 to 21,000 $OOID_{50}$ per year diverted to edible products. If the SRM list is restricted to brain only, this increases to over 23,000 $OOID_{50}$ per year.

6.4 The new TSE EURL: IZSPLVA & ISS

Giuseppe Ru, representative of Italy and TSE EURL director, presented the structure, plans and implementation of the new TSE EURL that will start operating from January 1st 2019. The new TSE EURL is an Italian consortium based in Torino and Rome. The coordinator institute is the Istituto Zooprofilattico Sperimentale del Piemonte, Liguria e Valle d'Aosta (IZSPLVA), which has been appointed as the Italian Reference Laboratory for TSEs since 1991 and OIE reference laboratory for BSE/scrapie since 2016. The Department of Food Safety, Nutrition and Veterinary Public Health (SANV) of the Istituto Superiore di Sanità (ISS) is responsible at the national level for molecular and biological strain typing since 2004 as well as genetics of animal TSE since 2006. A 3-year simulation programme has been provided to the EC outlining a number of annual activities and expected outputs to meet each of the 6 statutory functions as described in the legislation, namely, building up a collection of infected tissues, organizing >15 comparative exercises regarding the main diagnostic procedures (screening, confirmation, discrimination), epidemiological reporting, website maintenance, outbreaks assistance and training activities including an annual EURL meeting. The 2019 workplan has been designed including five main tasks: batch testing, active assistance for diagnostic and epidemiological investigations of TSE outbreaks, external quality assessments (EQAs), the dedicated website, and the maintenance of mouse breeding capabilities for experimental infections.

6.5 EU TSE annual report 2017: preliminary results and update on future data reporting

As per Regulation (EC) No 999/2001, as amended, EFSA is producing the European Union summary report on surveillance for the presence of TSE in 2017 (EFSA, 2018). Yves Van der Stede, member of the EFSA BIOCONTAM unit, presented the preliminary results of the on surveillance data collected through the testing of cattle, sheep, goats, cervids and species other than ruminants in the reporting countries (28 Member States plus Switzerland, Iceland and Norway) in 2017. The report will be published on November 28th, 2018. The reporting year, 2017, was the first year in which no cases of classical BSE have been reported world-wide (and the second year in the UK). Three MS reported six cases of Atypical BSE, four L-type and two H-type, out of 1,312,714 cattle tested in the EU. A total of 933 cases of scrapie were reported in 2017, 89% of them classical. In goats, 98% of the 567 scrapie cases were classical. Testing in cervids increased in 2017 with 3,585 in the EU and 25,736 in Norway, the only country that reported CWD cases: nine in reindeer, one in moose and one in red deer.

6.6 Update on the activities of the OIE in the TSE field

Fernanda Mejia Salazar, representative of OIE (Status Department), updated the Network on the recent TSE-related activities carried out at the OIE, and future ones. One country was recognised as having a negligible BSE risk status by the World Assembly of OIE Delegates in 2017: Nicaragua. At the moment, there are 35 territories registered with an OIE BSE risk status within Europe: 32 as countries 3 as zones. All member states have negligible risk status except Greece, Ireland and France. In the UK, Northern Ireland and Scotland have negligible risk. The maintenance of the official BSE risk status is conducted every year by the members with a recognised status reporting: changes in the epidemiological situation or other significant events, modification in legislation regarding BSE, imports of cattle and ruminant-derived meat-and-bone meal (MBM), audit findings in rendering plants and feed mills processing ruminant material and the results of the surveillance conducted in the previous 12 months. All countries or zones in Europe sample fallen stock, 26/35 sampled clinical suspects, 32/35 casualty slaughter and 21/35 healthy slaughtered cattle. In 2017, the OIE World Assembly of Delegates agreed on a scientific and technical review paper on the assessment of the current global BSE risk in support of the review of the standards. As a result two separate ad hoc groups were proposed to convene: on risk assessment and in surveillance.

6.7 Update on the activities of the European Commission in the TSE field

Sara Perucho Martinez, representative of the European Commission (DG SANTE – G), updated the Network on the recent TSE-related risk management activities in the European Commission, in relation to: i) specified risk material (SRM): the legislative proposal that removes tonsils, spleen and ileum from the SRM list for small ruminants and permits Member States to use an alternative method to determine the age of sheep at slaughter for the purposes of SRM removal were enforced via Commission Regulation (EU) 2018/969 of 9 July 2018; ii) feed ban,

i.e. relaxation of the total feed ban (lifting the ban for non-ruminants) provided that certain conditions are met, starting with pig PAP in poultry feed, insect PAP in poultry feed and the export of organic fertilisers and soil improvers; iii) the revision of BSE surveillance with the objective to reduce the numbers of tests of bovine animals and at the same time continue to measure the effectiveness of the measures in place with a better targeting of the surveillance activity; iv) scrapie eradication measures (i.e., proposal for a possible revision of Regulation (EC) Nº 999/2001 as regards TSE eradication measures in goats and endangered breeds will be presented for vote in the next Standing Committee on Plants, Animals, Food and Feed (PAFF) in October 2018); v) chronic wasting disease (i.e., new mandate to EFSA on zoonotic potential and transmission paths and possible next steps including ban of movements of live cervids from Finland to the rest of the EU, active surveillance expanded to all MS, the obligation of meat from cervids from Finland, the parts of Sweden and Norway being sourced from animals tested negative for CWD and the request to Non-EU countries exporting cervid meat to the EU (Russia and New Zealand) to increase surveillance regarding CWD if active surveillance for CWD to all Member States is implemented.

6.8 Round-the-table discussion on the EFSA Scientific Network on BSE-TSE

Network members were invited to provide, also after the meeting, suggestions for improvement of the functioning of the Network and for possible topics for future discussion in the Network. The chair invited all network members to share information with EFSA and the other Network members anytime during the year. The 14th meeting of the EFSA Network on BSE-TSE will be organised during the third quarter of 2019.

7. Any Other Business

No additional topics were discussed.

8. Closure of the meeting

The discussions held during the meeting will be summarised in the form of meeting minutes and an annual report, that will be circulated by EFSA Secretariat in due time to all participants for comments and agreement.

The next meeting of the EFSA Network on BSE-TSE will be organised during the third quarter of 2019.

The Chair thanked all speakers for their presentations, and all participants for attending the meeting and for their active participation in the discussions, and closed the meeting.