death and duration of the disease were compared in familial patients from successive generations in the following groups: (1) All cases except from, 2009 (65); the data were also analyzed separately for Methionine/Methionine (M/M) and Methionine/Valine /M/V) cases; (2) Generation age difference was compared in patients from 2009. Statistical evaluation was performed by Kaplan-Meier and Mann-Whitney tests.

In Group I the mean age at death was: 62.20 ± 7,219 y in the 1st and 50.04 ± 9.52 y in the 2nd generation. The difference of 12.16 y was significant ($p < 0.001$). No significant difference in the age at death between Met/Met and Met/Val patients was observed.

The mean duration was 5.75 ± 7.52 mo in the 1st and 4.41 ± 3.21months in the 2nd generation. The difference 1.34 was not significant ($p = 0.773$). The difference of clinical duration between M/M and M/V cases in the 2nd generation 3.11 ± 1.84 was at the border of significance ($p = 0.041$).

In Group II the mean age at death was 63.87 ± 6.54 y in the 1st and 48.96 ± 9.45 y in the 2nd generation. The difference 14.91 ± 8.93 was significant ($p < 0.001$).

Significantly decreased mean age at death in patients from successive generation in Groups I and II confirmed the anticipation in the analyzed gCJDE200K subtype and in Group II it confirmed the suggested influence of anticipation on increased incidence of gCJD.[3] The polymorphism M129V had no effect on the age at death, but it showed a slight (small number of heterozygotes) influence upon the difference between the clinical duration in homo- and heterozygotes in the 2nd generation. Anticipation was described also in gCJDE200K in Israel.[4] The evidence of anticipation in two most frequent subtypes of gCJDE200K signalizes that carriers of the E200K mutation in successive generations will develop the disease and die in decreasing, significantly younger age. These data have not only prognostic value, but could be implemented in the prophylaxis. They also underline the importance of genetic testing in gCJD affected families.

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PO-108: The role of the European Food Safety Authority in the context of the revision of the risk management measures on animal TSEs in the European Union

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The European Food Safety Authority (EFSA) is the keystone of European Union (EU) risk assessment regarding food and feed safety. EFSA carries out independent scientific risk assessment in order to produce scientific opinions and advice for risk managers. The EFSA's Scientific Panel on Biological Hazards (BIOHAZ) issues scientific opinions on biological hazards in relation to food safety and food-borne diseases including Transmissible Spongiform Encephalopathies.

Most of EFSA's work in the TSE area is based on requests from the European Commission (EC), which are issued in the context of the strategic document The TSE Roadmap 2.[1] This document underlines that any amendment to the EU measures on animal TSEs should maintain the EU's high level of protection of human and animal health and of food safety and should be backed by solid science. Considering the current favorable evolution of the epidemic of Bovine Spongiform Encephalopathy (BSE) in the EU, the TSE Roadmap 2 outlines possible future changes to EU measures on BSE in the short, medium and long-term until the year 2015. Moreover, measures on TSEs in small ruminants are also reflected upon in that strategy paper. Such possible changes would be related to: definition of Specified Risk Material, review of the current feed-ban, consideration of BSE monitoring schemes, measures on cohort-culling in bovine animals, consideration of ante-mortem and post-mortem rapid tests and review of Scrapie eradication measures.

The work describes the risk assessment activities undertaken by EFSA and its BIOHAZ Panel in the framework of the TSE Roadmap 2. In particular, the activities related to the revision of (1) the feed ban, (2) the BSE surveillance and (3) the Specified Risk Material are summarized and their impact on EU TSE policy outlined.

The scientific opinions of the EFSA BIOHAZ Panel will proof to be key to provide sound scientific basis to the risk managers in relaxing TSE controls in the EU without compromising the high level of consumer protection.

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PO-109: Sharp decline in scrapie prevalence in the Netherlands after breeding for resistance: Are we close to achieving eradication?

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Introduction. In the Netherlands an ambitious scrapie control program was started at the national level in 1998, based on genetic selection of animals for breeding. From 2002 onwards EU regulations required intensive active scrapie surveillance as well as certain control measures in affected flocks.

Materials and Methods. Here we use standard statistical methods as well as mathematical modeling to analyze (1) data on genotype frequencies and scrapie prevalence in the Dutch sheep population obtained from both surveillance and affected flocks; (2) data on PrP genotype frequencies in a random sample of flocks; (3) postal survey results on between-flock differences in breeding strategy and flock management.

Results and Conclusions. Analyzing the data (1) we find that the breeding program has produced a steady increase in the level of genetic scrapie resistance in the Dutch sheep population. We also found that a few years later this was followed by a sharp decline in the prevalence of classical scrapie in tested animals. Notably, the estimated classical scrapie prevalence level per head of susceptible genotype declined significantly as well. This indicates that selective breeding has a disproportionate effect on infection prevalence, reminiscent of the well-known population effect of vaccination against a transmissible disease. The overall recent decline in classical scrapie prevalence in Dutch sheep suggests that eradication of the disease in The Netherlands may be within reach. However, a subset of farms may still continue to act as a core group for scrapie transmission for some time, as we show by analyzing between-flock heterogeneities using the data (2) and (3). In addition, genetic resistance levels may decline again in future as participation to the selective breeding program has recently become voluntary.

PO-110: The future trend of BSE in Europe

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Introduction. The trend of the BSE epidemic has been analyzed per country, using the active surveillance data from various EU countries.[1] Now that the surveillance becomes less intensive, and the incidence declines below the detection limit, it is again interesting to see whether we can predict the future behavior of the BSE prevalence.

Materials and Methods. We use a method previously developed to evaluate the impact of control measures for BSE[2] and an age structured model to extrapolate the BSE prevalence into the future.[3] Thus we can calculate the BSE prevalence using the historic BSE surveillance data, under the assumption that the control measures remain the same, but also under various scenario's with relaxed control measures.

Results. We find that under the first scenario (no change in control measures) classical BSE will probably not be found in the EU after 2020. If the control scenarios are relaxed, this does not change much, until the reproduction ratio of the infection increases above 1. If that is the case, the epidemic will return and slowly, the prevalence will again increase. Depending on the national situation different combinations of control measures can be considered sufficiently safe.

Conclusion. To eradicate BSE, continuation of the present control measures is effective. It may be more efficient to apply a reduced set of control measures, but enforcing of such relaxed measures can be difficult, depending on the national feed production and animal husbandry situation.

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PO-111: BSE monitoring in the Russian Federation in 2011

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Background. The objective of the BSE monitoring in the Russian Federation (RF) is the confirmation of the efficacy of previously taken measures aimed at the reduction of BSE agent introduction and recirculation risk in cattle population. BSE monitoring is one of the activities of the Federal Service for Veterinary and Phytosanitary Surveillance (FSVPS) of the RF.

The most important measures of BSE control in Russia (in accordance with the OIE Questionnaire for BSE-status recognition) are: (1) 'feed ban': prohibition on feeding of ruminant protein feeds to ruminants has been in effect in the RF since 1990, control of ruminants' feed production for absence of ruminant components was introduced in 1996; laboratory control for ruminant genome was put in place in 2001; (2) restrictions on cattle and feed import from the countries where BSE cases were registered have been applied since 1989; (3) permanent notification program used when cattle showing BSE clinical signs is detected has been in place since 1999; (4) cattle identification is performed at farm level at present; and (5) diagnostic tests of animals in the BSE risk groups, monitoring program in cattle have been in effect since 1999.

Material and Methods. BSE monitoring in 2011 was performed in 23 RF regions where more than a thousand of bovine animals from controlled BSE risk countries had been imported. The population of adult (>24 mo) cattle in these regions is 3.51 million animals or 38.9% of adult cattle population in the RF. Bio-Rad diagnostic kits and equipment were used for brain sample testing.

Results. 5,258 cattle brain samples were collected within monitoring program in 2011 that is 0.15% of adult cattle population in RF regions participating in the monitoring program. More than 50% of samples were delivered from the largest livestock regions of Russia, such as Republics of Bashkortostan and Tatarstan, Krasnodar Territory, Rostov and Chelyabinsk Regions. Out of 5,258 submitted cattle brain samples 5,243 (99.71%) samples were suitable for diagnostic testing (they contained brain stem). The age of 90% of animals was 3–8 y. One brain sample was additionally tested using immunohistochemistry due to exceeding OD value detected by ELISA. In none of tested samples BSE agent was detected.

Conclusion. About 8,600 brain samples of cattle from risk groups predominantly were tested for BSE in the RF before 2011.