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SCIENTIFIC REPORT OF EFSA

Scientific and technical assistance on the minimum sample size to test should an annual BSE statistical testing regime be authorised in healthy slaughtered cattle[1]

European Food Safety Authority[2, 3]

European Food Safety Authority (EFSA), Parma, Italy

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ABSTRACT

This Scientific Report of EFSA provides scientific and technical assistance to the European Commission on (i) the minimum sample size to test, should an annual Bovine Spongiform Encephalopathy (BSE) statistical regime be authorised in healthy slaughtered cattle in certain EU Member States (MSs), and (ii) on the added value of that sample size for monitoring the trend of Classical BSE, Atypical BSE, and the emergence of a hypothetical new type of cattle Transmissible Spongiform Encephalopathy (TSE). Firstly, an evaluation of the epidemiological trends of BSE in 25 EU MSs was carried out in groups based on historical BSE monitoring data. Secondly, and with the aid of a purpose-built model called Cattle TSE Monitoring Model (C-TSEMM) developed by an EFSA contractor, both the assessment of the design prevalence and of the sensitivity of different BSE monitoring scenarios were carried out. Among the assumptions made in the C-TSEMM, a key one is that for those EU MSs with no, or few, BSE cases post-2001 an alternative estimate of cohort-based prevalence is required. This is estimated based on the average prevalence of the group of MSs with BSE cases under which they were placed in previous EFSA Opinions. Also, the model estimates presented are based on the demographics of the adult cattle population and on the number of adult cattle removed from the population via the different streams in 2011 (i.e. healthy slaughter, animals showing clinical signs of disease during *ante mortem* inspection, emergency slaughtered animals and fallen stock). Therefore, future fluctuations in those numbers at EU level and in each of the MSs will impact on the validity of the estimates presented in this report. A series of recommendations are made on sampling strategies for BSE monitoring and on the future use of the C-TSEMM.

KEY WORDS

cattle, TSE, BSE, monitoring, design prevalence

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[2] Correspondence: biohaz@efsa.europa.eu

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* The corrections made in this new version of the Scientific Output relate to changes in the C-TSEMM results provided by the contractor due to a model data input error (see Adkin et al., 2012). These changes impact on: (i) the quantitative estimates presented for the different scenarios modelled regarding BSE in general (i.e. when all strains including unknown strains were used in the calculations), and (ii) based on C-TSEMM estimates, the grouping of MSs where the estimated number of healthy slaughtered cattle tested may be reduced, whereby Finland is moved to the group where no sufficient numbers were tested based on the 2011 data. The correction made in the previous version of the Scientific Output (published on 24 January 2013) related to changes in Table 6, page 22, where the categories "Emergency slaughter" and "Fallen stock" were interchanged in Table 6 and in the text describing Table 6.

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SUMMARY

The European Commission has requested that the European Food Safety Authority (EFSA) provides scientific and technical assistance on the minimum sample size to test should an annual Bovine Spongiform Encephalopathy (BSE) statistical regime be authorised in healthy slaughtered cattle. In particular, in a scenario where the BSE testing of at-risk cattle would remain unchanged (i.e. testing of 100 % of at-risk cattle over 48 months), EFSA was asked: (i) to propose a minimum annual sample size in healthy slaughtered cattle above 72 months of age, that would allow the detection of BSE with a yearly design prevalence of at least 1 case per 100 000 in the adult population (i.e. older than 24 months of age) of the Member States (MSs), at a confidence level of 95% and both in the group of 25 EU MSs that are entitled for having the BSE monitoring system in healthy slaughtered cattle reviewed[4] as a whole and in each Member State individually; and (ii) to advise on the added value of this minimum sample to the overall surveillance programme in terms of monitoring the trend of Classical BSE, Atypical BSE and the emergence of a hypothetical new type of cattle Transmissible Spongiform Encephalopathy (TSE).

Firstly, an evaluation of the epidemiological trends of BSE in the 25 EU MSs is presented in this report based on BSE monitoring data provided by the European Commission. For this purpose, MSs are grouped following a similar approach taken in former EFSA Opinions: EU17 (Austria, Belgium, Cyprus, Denmark, Finland, France, Germany, Greece, Ireland, Italy, Luxembourg, Netherlands, Portugal, Slovenia, Spain, Sweden and United Kingdom) and EU8 (Czech Republic, Estonia, Hungary, Latvia, Lithuania, Malta, Poland and Slovakia). The EU8 group of MSs is further subdivided in EU5 (Estonia, Hungary, Latvia, Lithuania and Malta), where BSE has not been detected and in EU3 (Czech Republic, Poland and Slovakia), where BSE has been detected. It is to be noted that unless otherwise specified, the term BSE on its own refers to refers to all BSE types, including Classical BSE, Atypical BSE and "Unknown" type of BSE (i.e. a reported BSE case that has not been typed).

Based on that first evaluation, it was concluded that a constant decline in the total number of detected BSE cases (i.e. coming from both Active and Passive surveillance) has been recorded in the EU17 group from 2 157 cases in 2001 to 27 cases in 2011. In the EU3 group, the number of detected cases dropped down from 28 in 2005 (peak) to one in 2011. Moreover, the log10 transformed annual BSE prevalence and incidence (defined respectively as the number of positive BSE cases out of the tested population and out of the standing adult cattle population) in the EU17 and in the EU8 show a statistically significant decreasing trend. There has been a statistically significant increasing trend in the average age of the detected BSE cases per test year during the last 11 years and eight years in the EU17 and the EU8, respectively. At present, this average age exceeds 11 years in each of these MSs (where reported in 2011). Furthermore, and assuming that the age distribution of cattle within the EU25 has not changed substantially, the decreasing trend observed in the annual BSE occurrence and the increasing trend observed in the annual average age of the cases are the consequence of the implementation of the BSE control measures.

Regarding Atypical BSE, it is concluded that epidemiological data reported by the EU MSs indicate that over the last years the number of detected did not show any trend and that these cases were mainly identified in the fallen stock and healthy slaughtered animals older than eight years of age. However, it is also noted that the performance of the current BSE monitoring system, both in terms of its analytical sensitivity and earliness of the detection of animals infected with Atypical BSE is unknown.

Secondly, a model called Cattle TSE Monitoring Model (C-TSEMM) was developed by an EFSA contractor in order to provide a general frame for evaluating the design prevalence and the sensitivity

[4] The EU 25 MSs are: Austria, Belgium, Czech Republic, Cyprus, Denmark, Estonia, Finland, France, Germany, Greece, Hungary, Ireland, Italy, Latvia, Lithuania, Luxembourg, Malta, Netherlands, Poland, Portugal, Slovenia, Slovakia, Spain, Sweden and United Kingdom.

of cattle TSE monitoring systems[5]. The model was built considering available historical EU wide data on BSE monitoring, and contains assumptions, limitations and uncertainty that have to be considered when interpreting the different estimates that the model provides.

Among those assumptions, a key one is that for MSs with no, or few, BSE cases post-2001 an alternative estimate of cohort-based prevalence is required. This has been estimated for those MSs based on the average prevalence of the group of MSs with BSE cases under which they were placed in previous EFSA Opinions[6]: the EU17 or the EU8 group[7]. This results in an overestimate of prevalence for countries with no recorded cases as they are assumed to be a merged epidemiological unit with MSs where cases are observed.

Based on the estimates provided by the C-TSEMM model (that considered prevalence in the standing adult cattle population (i.e. period prevalence in a given year of detectable infected animals in the standing population) and the available historical EU wide data on BSE monitoring), it can be concluded that in the EU25 as a whole the current BSE monitoring regime enables the detection of one BSE case in 6 354 930 adult cattle with a confidence level of 95%. Moreover, if the current BSE monitoring regime would exclude testing of healthy slaughter cattle, it would be able to detect in the standing population one BSE case in 4 021 940 adult cattle with a confidence level of 95%. Therefore, no healthy slaughtered animals need to be tested in order to meet a design prevalence of 1 detectable case in 100 000 adult cattle, since testing of at risk animals (i.e. animals showing clinical signs during *ante mortem* inspection, emergency slaughter and fallen stock over 48 months of age and clinical suspects) is sufficient to meet the proposed design prevalence.

Furthermore and also based on C-TSEMM model estimates, it can also be concluded that at individual MS level, in eight MSs (Belgium, Denmark, France, Germany, Ireland, Netherlands, Spain and the UK) the testing of healthy slaughter animals is not needed in order to meet a 1 in 100 000 design prevalence with a confidence level of 95%, since testing of at risk animals is sufficient to meet the proposed design prevalence. On the other hand, in four MSs (Austria, Italy, Poland and Sweden) the testing of a fraction of healthy slaughtered animals older than 72 months of age (i.e. on the basis of the number tested in 2011) would be sufficient to meet a 1 in 100 000 design prevalence with a confidence level of 95%. Finally, in thirteen MSs (Cyprus, Czech Republic, Estonia, Finland, Greece, Hungary, Latvia, Lithuania, Luxembourg, Malta, Portugal, Slovakia and Slovenia) the number of tested animals in 2011 (i.e. including all the healthy slaughtered animals older than 72 months of age) did not allow to meet a 1 in 100 000 design prevalence with 95% confidence. However, fitting a sample size larger than the actually slaughtered cattle population of a MS is neither feasible nor realistic. Thus, the current testing of all animals of certain age categories that are slaughtered or dead may provide the most sensitive BSE monitoring system possible (i.e. that employs *post mortem* tests) under the current epidemiological scenario with the potential limitation on the impact of the age at testing as evaluated in former related EFSA Opinions[8].

[5] Amie Adkin, Robin Simmons and Mark Arnold; Model for evaluation of different options for the monitoring of Transmissible Spongiform Encephalopathies in cattle in the European Union (C-TSEMM). Supporting Publications 2012:EN-349. [55 pp.]. Available online: www.efsa.europa.eu/publications.

[6] Latest one: EFSA, 2010. Opinion of the Scientific Panel on Biological Hazards on a second update on the risk for human and animal health related to the revision of the BSE monitoring regime in some Member States. The EFSA Journal, 8(12), 1946.

[7] EU17: Austria, Belgium, Cyprus, Denmark, Finland, France, Germany, Greece, Ireland, Italy, Luxembourg, Netherlands, Portugal, Slovenia, Spain, Sweden and United Kingdom;
EU8: Czech Republic, Estonia, Hungary, Latvia, Lithuania, Malta, Poland and Slovakia

[8] EFSA (European Food Safety Authority), 2008a. Risk for Human and Animal Health related to the revision of the BSE Monitoring regime in some Member States. The EFSA Journal. 762, 1 - 47.
EFSA, 2008b. Further considerations 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. The EFSA Journal, 763, 1-8.
EFSA, 2009. Updated risk for human and animal health related to the revision of the BSE monitoring regime in some Member States. The EFSA Journal, 1059, 1-40.
EFSA, 2010. Opinion of the Scientific Panel on Biological Hazards on a second update on the risk for human and animal health related to the revision of the BSE monitoring regime in some Member States. The EFSA Journal, 8(12), 1946.

It is further concluded that in the event of a re-emergence of Classical BSE, stopping the testing of healthy slaughtered cattle would lower the sensitivity of its detection by the TSE monitoring system. As an example, based on a theoretical scenario of an annual 10% increase in detectable cases in the tested population (prevalence), the C-TSEMM model estimates that: (i) In the EU25 as a whole, where testing healthy slaughtered cattle above and age of 72 months is not needed in order to meet the proposed design prevalence, the time to detection of the supposed 10% yearly increase in detectable cases would increase from six to 11 years (i.e. five extra years to detect the supposed 10% yearly increase in prevalence of detectable cases) should testing of healthy slaughtered cattle be stopped compared to the current testing regime; (ii) In those MSs where testing healthy slaughtered cattle above the age of 72 months is not needed in order to meet the proposed design prevalence (Belgium, Denmark, France, Germany, France, Ireland, Netherlands, Spain and the UK), it would take between three and eight extra years (depending on the MS) to detect that yearly increase in prevalence should testing of healthy slaughtered cattle be stopped compared to the current testing regime; (iii) In those MSs where testing healthy slaughtered cattle could be reduced in order to meet the proposed design prevalence (Austria, Italy, Poland and Sweden), it would take between six and 16 extra years (depending on the MS) to detect that yearly increase in prevalence should testing of healthy slaughtered cattle older than 72 months of age be reduced to the number needed to meet the proposed design prevalence compared to the current testing regime; (iv) In those MSs where testing healthy slaughtered cattle older than 72 months of age as per the current BSE monitoring regime is not sufficient to meet the proposed design prevalence(Cyprus, Czech Republic, Estonia, Finland, Greece, Hungary, Latvia, Lithuania, Luxembourg, Malta, Portugal, Slovakia and Slovenia), it would take between three and 25 extra years (depending on the MS) to detect that yearly increase in prevalence should testing of healthy slaughtered cattle be stopped compared to the current testing regime.

When considering Atypical BSE, at EU25 as a whole there is not sufficient data (i.e. number of detected cases annually) to reliably estimate with the C-TSEMM model the impact of the stopping/continuation of testing healthy slaughtered animals older than 72 months. However, when using France as an example (i.e. country with a large population and sufficient number of detected Atypical cases) the C-TSEMM model indicates that, based on a theoretical scenario of an annual 10% increase of detectable prevalence of Atypical BSE in the tested population, it would take an extra 13 years to detect that yearly increase in prevalence should testing of healthy slaughtered cattle be stopped compared to the current testing regime.

Considering the timeframe available for this mandate, carrying out simulation studies for hypothetical new types of cattle TSEs was not possible. However, it was concluded that the C-TSEMM model can be considered as a useful tool in order to simulate future *ad hoc* epidemiological scenarios of hypothetical new types of cattle TSEs.

It is highlighted that when interpreting the estimates presented above or those obtained in future simulations performed with the C-TSEMM model, consideration has to be given to the assumptions, limitations and uncertainty in the model. Moreover, the models estimates presented in this report are based on the demographics of the adult cattle population in 2011 and on the number of adult cattle removed from the population via the different streams (i.e. healthy slaughter, animals showing clinical signs of disease during *ante mortem* inspection, emergency slaughtered animals and fallen stock). Therefore, future fluctuations in those numbers at EU level and in each MSs will impact the validity of current estimates.

A series of recommendations are made in this report including considerations on the sampling strategy should monitoring of BSE in healthy slaughtered cattle remain based on a sample of animals over certain age, and considerations on future potential needs for the assessment of the impact of changes to current EU BSE control measures in the sensitivity of the EU surveillance system.

It is finally recommended that if the C-TSEMM model will be employed in future years for the review of the BSE monitoring regime in the EU, updated yearly data including BSE testing data have to be considered as these drive the results estimated by the model.

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BACKGROUND AS PROVIDED BY THE EUROPEAN COMMISSION

According to TSE legislation[9], each Member State shall carry out an annual monitoring programme for BSE, including a screening procedure using rapid tests approved for that purpose. This programme shall cover, as a minimum, all bovine animals above 30 months of age slaughtered normally for human consumption (healthy slaughtered cattle) and all bovine animals above 24 months of age which have died/been killed or been sent for emergency slaughter (at risk cattle). However, a Member State which can demonstrate, based on epidemiological criteria, the improvement of the BSE situation on its territory may send an application to the Commission with a view to being authorised to revise its monitoring programme.

Since 2009, all Member States except Bulgaria and Romania have progressively been authorised, based on their favourable epidemiological situation and following positive EFSA opinions[10], to review their BSE monitoring programmes and to raise the age limit for testing to 72 months in healthy slaughtered cattle and 48 months in at risk cattle.

Furthermore, as laid down in Article 2, point 3 of Commission Decision 2009/719/EC as amended by Commission Implementing Decision 2011/358/EU[11], these 25 Member States will be allowed, as from 1st January 2013, to test only a minimum annual sample of the healthy slaughtered cattle above 72 months of age.

TERMS OF REFERENCE AS PROVIDED BY THE EUROPEAN COMMISSION

In accordance with Article 31 of (EC) Regulation 178/2002, EFSA is requested to provide scientific and technical assistance on the minimum sample size that the Member States that are listed in the Annex to Commission Decision 2009/719 as lastly amended by Commission Implementing Decision 2011/358/EU should test for BSE as from 1st January 2013, if they decide to opt after that date for testing only a minimum annual sample of the healthy slaughtered cattle population.

More specifically, in a scenario where the BSE testing of at-risk cattle would remain unchanged (i.e. testing of 100 % of at-risk cattle over 48 months), EFSA is asked to propose a minimum annual sample size in healthy slaughtered cattle above 72 months of age in order to:

- allow the detection of BSE (both Classical and Atypical strains) with a design prevalence of at least one case per 100 000 in the adult population of the Member States, at a confidence level of 95% (i.e. consistent with type A surveillance as described in article 11.5.22 of the OIE code);
- monitor the trend of BSE in cattle (both Classical and Atypical strains);
- detect the emergence of a hypothetical new type of TSE in cattle.

[9] Regulation (EC) No 999/2001 of the European Parliament and of the Council of 22 May 2001 laying down rules for the prevention, control and eradication of certain transmissible spongiform encephalopathies (OJ L 147, 31.5.2001, p. 1)

[10] 17 July 2008: 'Scientific Opinion of the Panel on Biological Hazards on a request from the European Commission on the risk for human and animal health related to the revision of the BSE monitoring regime in some Member States', The EFSA Journal (2008) 762, p. 1.
29 April 2009: 'Scientific Opinion of the Panel on Biological Hazards on a request from the European Commission on the updated risk for human and animal health related to the revision of the BSE monitoring regime in some Member States', The EFSA Journal (2009) 1059, p. 1.
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April 2011: "Scientific Opinion on the review on the risk for human and animal health related to the revision of the BSE monitoring regime in three EU Member States", The EFSA Journal 2011;9(4):2142

[11] Commission Implementing Decision 2011/358/EU of 17 June 2011 amending Decision 2009/719/EC authorising certain Member States to revise their annual BSE monitoring programmes (OJ L161, 21.6.2011, p.29)

In order to facilitate implementation by the Member States, according to their adult cattle population (>2 years of age - see Table 1), the sample size should be determined for each of the following adult cattle population groups: <25 000; [25 000 – 50 000]; [50 000 – 100 000]; [100 000 – 200 000]; [200 000 – 300 000]; [300 000 – 400 000]; [400 000 – 500 000]; [500 000 – 600 000]; [600 000 – 700 000]; [700 000 – 800 000]; [800 000 – 900 000]; [900 000 – 1 000 000]; ≥ 1 000 000.

The adult cattle population (>2 years of age) of the 25 Member States and an estimate of the number of cattle over 72 months old is provided in Table 1.

Table 1: Distribution of the adult cattle population in the 25 EU Member States.

Proposed adult cattle population group (>24 months)	**Member State**	**Adult cattle population (>24 months)[12]**	**Estimate of healthy cattle >72 months slaughtered each year[13]**
<25 000	Malta	7 200	635
	Cyprus	24 800	2 683
[25 000 – 50 000]			
[50 000 – 100 000]	Luxembourg	98 400	4 747
[100 000 – 200 000]	Estonia	125 900	10 804
[200 000 – 300 000]	Slovenia	201 000	14 384
	Latvia	206 300	20 145
	Slovakia	241 000	14 946
[300 000 – 400 000]	Hungary	357 000	33 048
	Greece	361 000	15 072
	Finland	380 200	31 202
[400 000 – 500 000]	Lithuania	420 000	49 078
[500 000 – 600 000]			
[600 000 – 700 000]	Czech Republic	628 600	52 511
	Sweden	650 500	53 717
[700 000 – 800 000]	Denmark	760 000	67 315
[800 000 – 900 000]	Portugal	810 200	62 016
[900 000 – 1 000 000]	Austria	935 600	118 175
≥ 1 000 000	Belgium	1 321 500	116 551
	Netherlands	1 766 000	197 673
	Italy	2 704 300	281 190
	Poland	2 948 300	335 689
	Ireland	2 957 400	292 424
	Spain	3 258 200	299 737
	United Kingdom	4 652 000	476 697
	Germany	5 822 600	606 562
	France	10 544 000	1 087 646

[12] Source: EUROSTAT July 2011
[13] Source: Number of healthy slaughtered cattle over 72 months tested in 2010 (TSE database)

Clarification on the Terms of Reference:

Following discussions with the European Commission, the above Terms of Reference were rephrased as follows:

More specifically, in a scenario where the BSE testing of at-risk cattle would remain unchanged (i.e. **testing of 100 % of at-risk cattle over 48 months**):

- EFSA is asked to propose a minimum annual sample size in healthy slaughtered cattle above 72 months of age in order to allow the detection of BSE with a **yearly** design prevalence of at least **one case per 100 000** in the adult population of the Member States, at a confidence level of 95% in the group of 25 EU Member States as a whole and in each Member State individually;

- What is the **added value** of this minimum sample to the overall surveillance programme in terms of monitoring the trend of Classical BSE, Atypical BSE and the emergence of a hypothetical new type of cattle TSE?

ANALYSIS

1. Introduction

The current Active monitoring of cattle TSEs in the European Union (EU) has been traditionally designed with the aim of ensuring that all bovines of certain age are tested. The age for testing depends on the health status of the animal (i.e. healthy slaughtered animals, emergency slaughtered, animals showing clinical signs during *ante mortem* inspection, fallen stock)[14].

In view of the BSE epidemiological trends in the EU, the European Commission has tasked in the past to EFSA the assessment of the age for TSE testing of cattle in some EU Member States (MSs) (EFSA, 2008a, 2008b, 2009, 2010, 2011). As a consequence, a derogation allows 25 EU MSs[15] to test healthy slaughtered cattle at 72 months and at risk cattle at 48 months of age. Nevertheless, MSs may continue testing at younger ages. Table 2 shows the age for testing BSE in cattle in the EU implemented by the MSs in 2011.

Table 2: BSE testing ages (in months) for bovine animals during 2011 in the EU MSs by testing stream. When the same testing age applies to different testing streams this is shown overarching those streams. Source: European Commission.

	Age limit in months					
	Fallen Stock	**Emergency slaughtered**	**Clinical signs at AM**	**Healthy slaughtered**	**BSE eradication**	**BSE suspects**
Austria	> 24	> 48		> 72*	No age limit	
Belgium	> 48			> 72*	> 24	No age limit
Bulgaria	> 24			> 30	No age limit	
Czech Republic	> 48*			> 72*	No age limit	
Cyprus*	> 24			> 72*	> 48	No age limit
Denmark	> 48			> 72*	> 24	No age limit
Estonia	> 48*			> 72*	No age limit	
Finland	> 48			> 72*	No age limit	
France	> 24			> 72*	> 24	No age limit
Germany	> 48			> 72*	No age limit	
Greece	> 48			> 72*	No age limit	
Hungary	> 24			> 72*	No age limit	
Ireland	> 48			> 72*		No age limit
Italy	> 48			> 72*	No age limit	
Latvia	> 48*			> 72*	No age limit	
Lithuania	> 48*			> 72*	No age limit	
Luxembourg	> 24	> 48		> 72*	> 24	No age limit
Malta	> 48*			> 72*	No age limit	
Netherlands	> 48			> 72*	No age limit	
Poland	> 48*			> 72*	No age limit	
Portugal	> 48*			> 72*	>48*	No age limit
Romania	> 24			> 30	No age limit	
Slovakia	> 48*			> 72*	No age limit	
Slovenia	> 48			> 72*	No age limit	
Spain	> 36			> 72*	No age limit	
Sweden	> 48			> 72*	No age limit	
United Kingdom	> 48			> 72*	No age limit	

* Since 1 July 2011

[14] As defined in Annex III, Chapter A of Reg. (EC) 999/2001 (as amended) of the European Parliament and of the Council of 22 May 2001 laying down rules for the prevention, control and eradication of certain transmissible spongiform encephalopathies (OJ L 147, 31.5.2001, p. 1).

[15] At present Bulgaria and Romania have not been yet considered for an increase in testing ages due to the six-year requirement of EU BSE monitoring and controls been fully implemented.

This Scientific Report of EFSA replies to the request of the European Commission that seeks advice on a minimum annual sample size in healthy slaughtered cattle above 72 months of age (i.e. move from systematic testing to annual sample-size testing) whereby testing of at-risk cattle would be maintained at 48 months of age (i.e. unchanged with current practices). This minimum annual sample size of healthy slaughtered cattle:

- Should allow the detection of BSE with a yearly design prevalence of at least one case per 100 000 in the adult cattle population of the MSs at a confidence level of 95% (it should be taken into account that testing of all the at risk cattle over 48 months of age also counts when meeting that design prevalence),

- Has to be examined for its added value to the overall surveillance programme for monitoring the trend of Classical BSE, Atypical BSE and the emergence of a hypothetical new type of cattle BSE.

2. Analysis of the trend of BSE in the 25 EU Member States

2.1. Approach, data sources and general assumptions

For the purposes of the review of the BSE monitoring regime in the 25 EU MSs considered in this Report, EU MSs are separated into two groups based on BSE monitoring data, resulting in:

- A group of 17 EU MSs (hereafter referred as EU17), for which the data taken into account goes back to 1st January 2001 (i.e. consistent with the approach taken in previous EFSA Opinions). The EU MSs forming this group are: Austria, Belgium, Cyprus, Denmark, Finland, France, Germany, Greece, Ireland, Italy, Luxembourg, Netherlands, Portugal, Slovenia, Spain, Sweden and United Kingdom.

- A group of 8 EU MSs (hereafter referred as EU8), for which data taken into account goes back to 1st January 2004[16] (i.e. consistent with the approach taken in previous EFSA Opinions). The EU MSs forming this group are: Czech Republic, Estonia, Hungary, Latvia, Lithuania, Malta, Poland and Slovakia. Within this group, two subgroups are identified: A subgroup of 5 EU MSs (hereafter referred as EU5) where BSE has not been reported (i.e. Estonia, Hungary, Latvia, Lithuania and Malta) and a subgroup of 3 EU MSs (hereafter referred as EU3) where BSE has been reported (i.e. Czech Republic, Poland and Slovakia).

The following data sources have been employed for the analysis presented in this Report:

- Data on BSE cases detected in the EU were received from the European Commission on 3 July 2012. Further clarifications on these data were made with the support of the European Commission. The following has to be taken into account when interpreting the data presented in this Scientific/Technical Report:

    - Unless otherwise specified, the term "BSE" on its own refers to all BSE types, including Classical BSE, Atypical BSE and Unknown type of BSE (i.e. a reported BSE case present in the data received from the European Commission that has not been typed).

    - Only reported BSE cases with known age category (i.e. age-group reported in the database from the European Commission) and year of testing are considered. Based on this, a total of 15 reported cases were excluded from the analysis, as described in Appendices A and B.

- Data on the number of rapid TSE tests performed in the EU in the frame of BSE monitoring

[16] When applicable, the number of BSE cases diagnosed before 1st May 2004 in the MSs of the EU8 group are addressed in the relevant tables in Appendix A.

were received from the European Commission (EC) on 11 January 2012 and 3 July 2012. When needed, clarifications on these data were made with the support of the European Commission.

- Data on the adult bovine population (over 24 months of age) from the year 2011 in the 25 MSs were received from the European Commission on 11 January 2012, and were also retrieved from EUROSTAT[17] on 5 and 26 April 2012 for the whole period 2001 to 2011.

Detailed epidemiological information on BSE monitoring in the EU can be found in the TSE annual reports released by the EC, available at:

http://ec.europa.eu/food/food/biosafety/tse_bse/monitoring_annual_reports_en.htm

Summary tables reviewing the trend of BSE in the EU 25 MSs grouped in EU17 and EU8 are presented in Appendix A. The number of BSE cases detected through the BSE surveillance (Active and Passive) between 2001/2004 and 2011 per EU MS, birth cohort and year of detection are presented in Appendix B.

Some minor differences may be found between the data presented in this EFSA Report and those presented in previous EFSA Opinions. This is due to ongoing updates/corrections that the MSs may make to the European Commission databases on BSE monitoring and to EUROSTAT databases. On the other hand, the few BSE cases arising from BSE eradication measures (i.e. cohort-culling) are not included in the calculations presented in section 2 of this Report, as they come from a stream other than the epidemiological surveillance. However, it has to be noted that their exclusion does not affect the trends of the BSE epidemic. Details on the number of these cases per MS are presented in Appendix B.

In line with previous EFSA Opinions dealing with requests on the BSE monitoring regime (EFSA, 2008a, 2008b and 2009), three key assumptions are made for each EU MS considered in this Report in order to render the analysis presented in this chapter valid:

- It is assumed that all 25 EU MSs considered for this mandate have implemented a BSE surveillance system and control measures as set out in Reg. (EC) 999/2001[18] for at least six years. If this assumption cannot be verified, the conclusions of this opinion will not apply to the respective MS.

- It is assumed that all 25 EU MSs considered for this mandate will continue to implement currently applied measures as set out in Reg. (EC) 999/2001 aimed at controlling and reducing BSE in the EU MSs.

- It is assumed that the rapid tests applied in the frame of the Reg. (EC) 999/2001 for BSE surveillance have a sensitivity of 100% in the very late stages of the incubation period**.** The likely point in the incubation period at which PrP^{res} is detectable with the rapid BSE tests depends on the infective dose. While the range of doses of exposure of field cases of BSE is not known, an oral attack rate study has shown that the mean incubation period arising from doses in the range 0.1-1g fits with that estimated for field cases. For a 1g dose, it was found that PrP^{res} was detectable only after 97% of the length of the incubation period. This degree of under-detection has to be taken into account when estimating infection prevalence from surveillance data.

[17] EUROSTAT data available at: http://epp.eurostat.ec.europa.eu/portal/page/portal/eurostat/home – Data tree: Statistics> Data Navigation Tree> Database by themes> Agriculture, forestry and fisheries> Food: From farm to fork statistics (food) > inputs to the food chain (food_in)> Livestock (1000 heads) (food_in_pagr2)

[18] Regulation (EC) 999/2001 of the European Parliament and of the Council laying down rules for the prevention, control and eradication of certain transmissible spongiform encephalopathies.

When interpreting the significance of the results presented in this section of the EFSA report, the following points should also be considered:

- The shape of the age distribution of BSE cases depends on two aspects: the age distribution of the cattle population and the level of BSE transmission in the past (de Koeijer et al., 2002).
- Out of the BSE cases found in the EU17, only 47 cases were related to animals born after the start of the total feed ban in 2001.
- The EU8 are all new EU MSs since 1 May 2004, since when the EU total feed ban has been implemented in these MSs. In the EU3 a total of 16 cases have been born since 2001, and 3 cases are born after 30th April 2004.
- The Geographical BSE Risk as well as the stage of the BSE epidemic can vary considerably between MSs.

Furthermore, this Scientific Report is supported by the modelling work of a contractor (Animal Health and Veterinary Laboratories Agency (AHVLA), United Kingdom) identified by EFSA through an open call for tender (Ref. CT/EFSA/BIOHAZ/2011/03[19]). Key background details and results of the modelling work are presented in section 5 of this EFSA Report. Also, the full Scientific Report submitted to EFSA by the contractor (Adkin et al., 2012) should be read as background and supporting information.

2.2. Trends of BSE in the EU17 during the period 2001 to 2011

Extensive epidemiological data on BSE has been collected via the BSE Active and Passive Surveillance over the last 11 years in the EU17. Detailed tables on the epidemiological trend of BSE are presented in Appendix A, while in this section summary tables and figures are presented.

From 2001 until the end of 2011, more than 92 million of tests were carried out in the framework of BSE Active Surveillance in the EU17. Of these, 5 220 animals were positive. These included 1 266 out of 79 277 027 million healthy slaughtered cattle tested (15.96 per million healthy cattle tested), and 3 954 out of 13 055 343 at risk cattle (302.86 per million), while testing schemes differed between MSs during this period of time. For example: Germany tested younger healthy stock than most MS. In the framework of BSE Passive Surveillance in EU17 during the period 2001 – 2011 a total 22 406 bovine animals were tested and 2 407 were positive.

Based on the available data, no BSE cases have been reported in EU17 in the framework of BSE Surveillance in 2011 in: Austria, Belgium, Cyprus, Denmark, Finland, Germany, Greece, Luxemburg, Netherlands, Slovenia and Sweden. Moreover, also in 2010 no cases have been reported in these same countries, except for Austria and Netherlands. Italy did not report any BSE case in 2010.

With respect to the number of BSE cases detected through the BSE Active and Passive Surveillance and in the frame of eradication measures in EU17 from 2001 to 2011, the data per target group are reported in Table 3.

[19] For further contract award details see http://ted.europa.eu/udl?uri=TED:NOTICE:393223-2011:TEXT:EN:HTML

Table 3: Number of BSE cases detected through the BSE Active and Passive Surveillance and eradication measures in EU17 during the period 2001 – 2011 per target group.

	No of detected BSE cases per testing year											
Target Group	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	**Total**
Active Surveillance												
Healthy slaughtered	280	292	264	162	97	72	30	25	23	13	8	1 266
At risk animals												
Emergency slaughter	321	509	316	167	121	31	7	5	3	0	0	1 480
Fallen stock	400	610	406	308	212	161	92	75	31	28	19	2 342
Presenting Clinical signs at *ante mortem* inspection	35	24	31	11	16	9	4	2	0	0	0	132
Total Active Surveillance	1 036	1 435	1 017	648	446	273	133	107	57	41	27	5 220
Passive Surveillance												
Suspects subject to lab	1121	674	304	172	74	37	15	8	2	0	0	2 407
Eradication Measures	9	10	3	5	13	1	1	3	0	0	0	45
Total	2 166	2 119	1 324	825	533	311	149	118	59	41	27	7 672

The trend observed in these data demonstrates that the control measures in place against BSE have been efficient because the prevalence of the disease (i.e. number of positive cases out of the total number of tested animals in a given year) is declining exponentially in the EU17. This can also be seen in Figure 1 below.

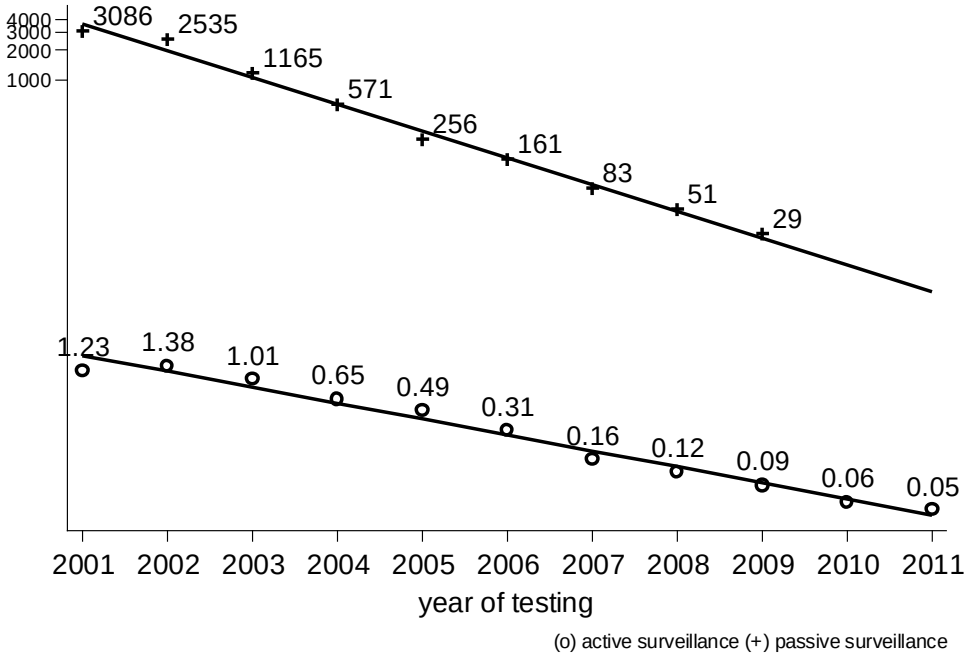

Figure 1: Prevalence (log10 scale) of BSE in the EU17 found by Active and Passive surveillance from 2001 to 2011. In 2010 and 2011 no cases were reported in Passive surveillance.

When modelling the log10 prevalence on the year, these declining trends on yearly prevalence in the EU17 are statistically significant for both the prevalence rates obtained within Active and Passive surveillance (beta values -0.16 and -0.27 for Active and Passive, respectively, with $p<0.001$ for both).

The total number of BSE cases per birth cohort detected through BSE Surveillance (both Active and Passive) in EU17 during the period 2001 – 2003 and 2004 to 2011 are presented in Figure 2. In the EU17 there were two apparent consecutive waves of infection, the first in the mid 90's and a second one (lower number of cases involved) between 1998 and 2000.

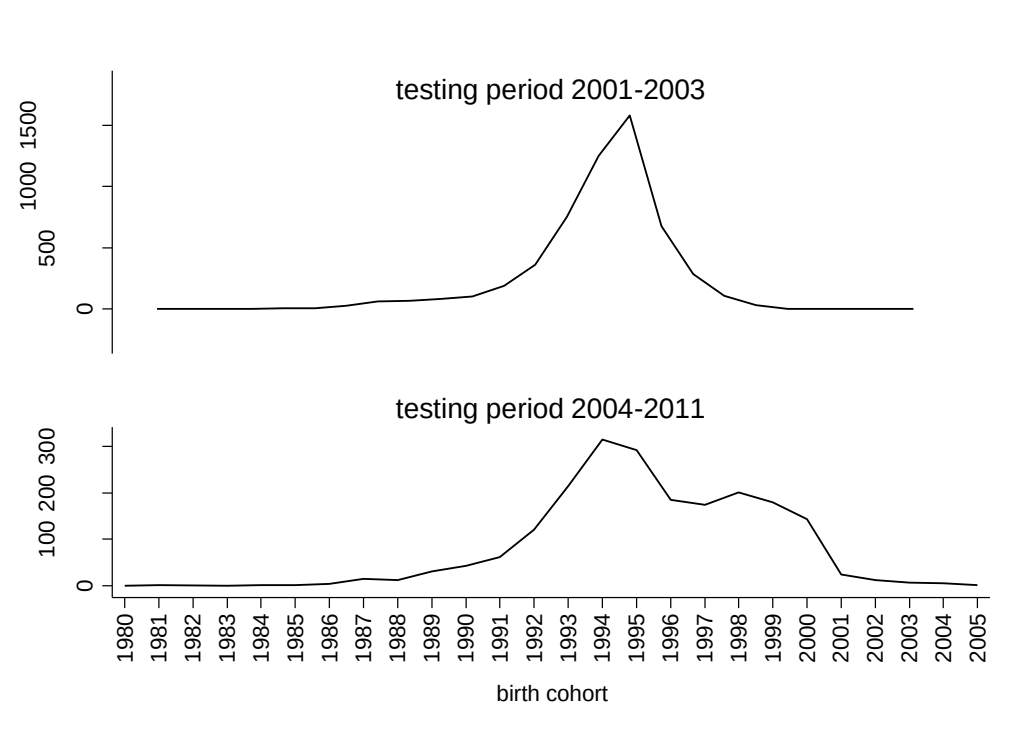

Figure 2: Number of BSE cases per birth cohort detected through BSE surveillance (both Active and Passive) in EU17 during the periods 2001 to 2003 and 2004 to 2011. Y-axis scales from 0 to 1 500 in the upper and form 0 to 300 cases in the lower figure.

The trend of the average age of BSE cases per year of detection and incidence (i.e. number of positive cases out of the total adult cattle population older than 24 months of age in a given year) in the EU17, considering both BSE Active and Passive Surveillance are shown in Figure 3.

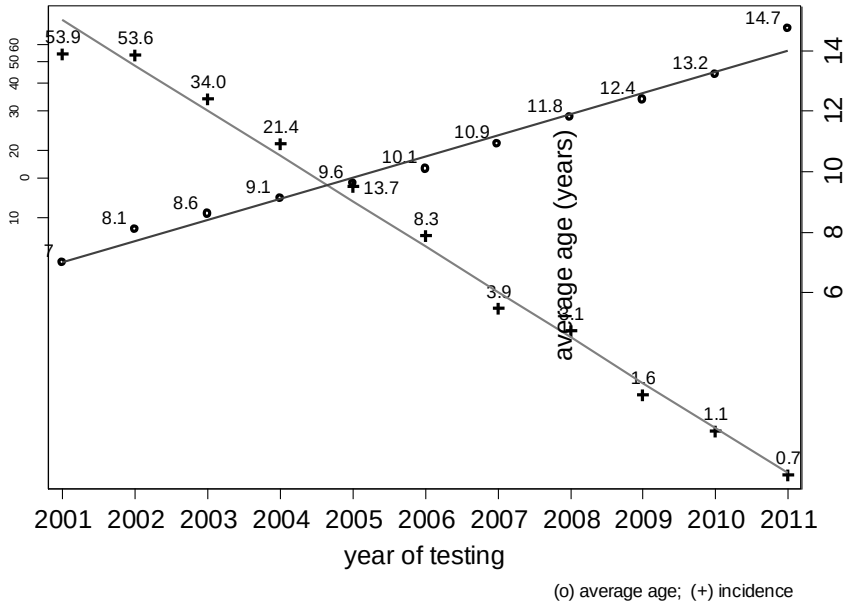

Figure 3: Incidence (log10 scale) of BSE in the EU17 per year of testing, considering both BSE Active and Passive Surveillance, and average age of those BSE cases for the given years.

When fitting regression models using year of testing as independent variable, and the log10 incidence and the average age at year of testing as dependent variables respectively, both the declining trend in incidence and the increasing trend in average age are statistically significant in the EU17 (beta values --0.20 and +0.70 for incidence and average age, respectively, with $p<0.001$ for both).

Concluding remarks on the trend of BSE in the EU 17

- A constant decline in the total number of BSE cases per year (coming from both Active and Passive surveillance) has been recorded in EU 17: from 2 157 cases in 2001 to 27 cases in 2011.
- The log10 transformed annual BSE prevalence and incidence within EU17 (defined respectively as the number of positive BSE cases out of the tested population and out of the standing adult cattle population) shows a statistically significant decreasing trend.
- Over the last 11 years, there has been a statistically significant increasing linear trend in the average age of the detected BSE cases in the EU17, which currently exceeds 11 years in each of these MSs (where reported in 2011).
- When considering the birth cohort of the BSE cases in the EU17 there were two apparent consecutive waves of infections, the first in the mid 90's and a second one (lower number of cases involved) between 1998 and 2000.
- Assuming that the age distribution of cattle tested for BSE within the EU17 MSs has not changed substantially over the considered period, the decreasing trend observed in the log10 transformed annual BSE incidence and the increasing trend observed in the annual average age of the cases indicates that the transmission of BSE has decreased in the EU17 as a consequence of the implementation of the control measures.
- Data on BSE surveillance from 2001 to 2011 indicate that in the EU17 the BSE epidemic is fading out.

2.3. Trend of BSE in the EU8 during the period 2004 to 2011

Extensive epidemiological data on BSE has been collected in the framework of EU regulations via the BSE Active and Passive Surveillance over the last eight years in the EU8. Detailed tables on the epidemiological description of the trend of BSE are presented in Appendix A, while in this section summary tables and figures are presented.

As addressed earlier on in this Report, it has to be noted that out of the eight MSs of interest, only three - referred to as EU3 - have reported positive BSE cases: Czech Republic, Poland and Slovakia. Thus, in five of the MSs of the EU8 group - referred to as EU5: Estonia, Hungary, Latvia, Lithuania and Malta - BSE cases have not been identified through the EU BSE monitoring regime.

In the EU5 group, where BSE has not been identified, more than 1.9 million tests have been carried out in the framework of BSE Active surveillance between 2004 and 2011. Of these tests, about 1.68 million were tests done in healthy slaughtered cattle, while approximately 240 000 at risk cattle were tested.

In the EU 3 group, more than 6.2 million of tests have been carried out in the framework of BSE Active surveillance since 2004. Of these, 95 animals were positive. These included 63 out of 5425242 healthy slaughtered cattle tested (11.61 per million healthy cattle tested), and 32 out of 862 751 at risk cattle tested (37.09 per million). In the framework of BSE Passive Surveillance in EU3 during the period from 2004 to 2011, a total of 169 bovine animals were tested and none was positive.

In the framework of BSE Surveillance in 2011, only one BSE case has been reported in the EU3 (in Poland), whereas in 2010 three cases were reported, 2 in Poland and 1 in Slovakia, respectively.

With respect to the number of BSE cases detected through the BSE Active and Passive Surveillance and in the frame of eradication measures in EU3 between 2004 and 2011, data per target group are presented in Table 4.

Table 4: Number of BSE cases detected through the BSE Active and Passive Surveillance and from eradication measures in EU3 during the period 2004 – 2011 per target group.

	No of detected BSE cases per testing year								
Target Group	2004[1]	2005	2006	2007	2008	2009	2010	2011	**Total**
Active Surveillance									
Healthy slaughtered	15	18	9	9	4	5	2	1	63
At risk animals									0
Emergency slaughter	5	2	0	1	2	0	0	0	10
Fallen stock	5	7	4	3	0	1	1	0	21
Presenting Clinical signs at *ante mortem* inspection	0	1	0	0	0	0	0	0	1
Total Active Surveillance	25	28	13	13	6	6	3	1	95
Passive Surveillance									0
Suspects subject to lab	0	0	0	0	0	0	0	0	0
Eradication Measures	0	3	0	0	0	0	0	0	3
Total	25	31	13	13	6	6	3	1	98

[1] In 2004, seven cases were diagnosed before 1 May.

The trend observed in these data demonstrates that the control measures in place against BSE have been efficient because the prevalence of the disease is declining exponentially in the EU3. This can also be seen in Figure 4 below.

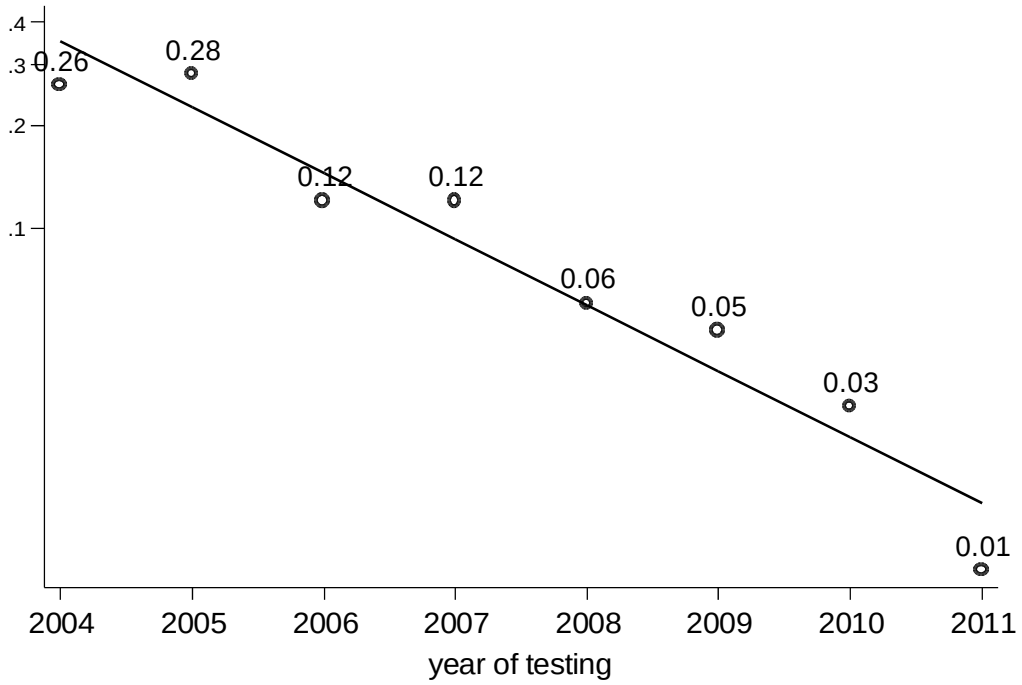

Figure 4: Prevalence (log10 scale) of BSE in the EU8 found by Active surveillance from 2004 to 2011. No cases were reported in EU8 by Passive surveillance.

When comparing the prevalence in the EU8 for the period 2004 to 2009 with the prevalence in the EU17 for the period 2001 to 2006 (i.e. 6 first years of the total feed ban), it can be noticed that the yearly prevalence in the EU8 is in the range of 7 to 4 times lower than that of the EU17.

As for the EU17, also in the EU8, when modelling the log10 prevalence on the year there is a significant continuous declining trend on the yearly prevalence (beta -0.19, $p<0.001$).

The total number of BSE cases per birth cohort detected through BSE Surveillance (both Active and Passive) in EU3 during the period 2004 to 2011 are presented in Figure 5. In the EU3 there were two apparent consecutive waves of infections, the first in the mid 90's and a second one (with larger number of cases involved) around the year 2000.

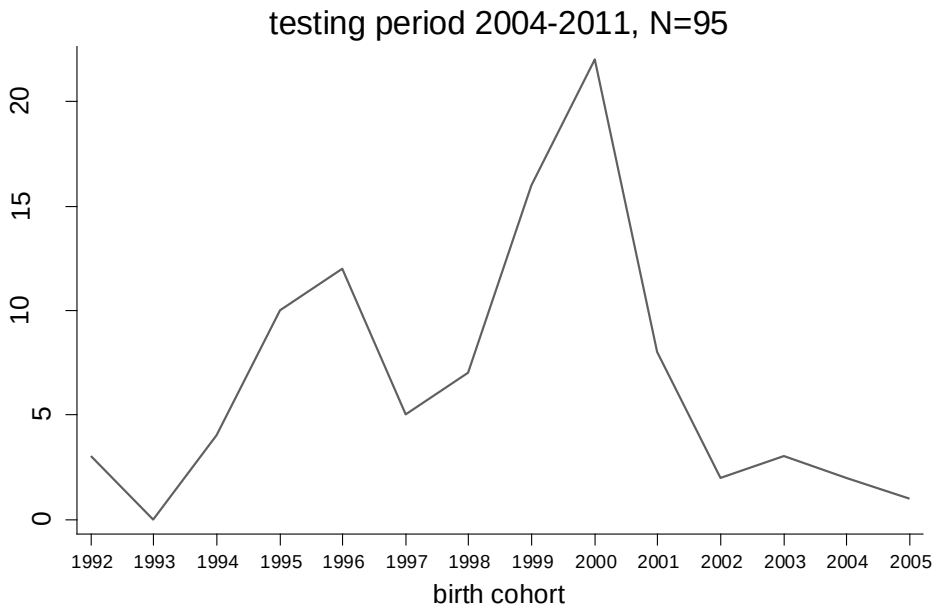

Figure 5: Number of BSE cases per birth cohort detected through BSE Active surveillance in EU3 during the period 2004 to 2011.

The trend of the average age of BSE cases per year of detection and incidence in the EU8, considering both BSE Active and Passive Surveillance are shown in Figure 6.

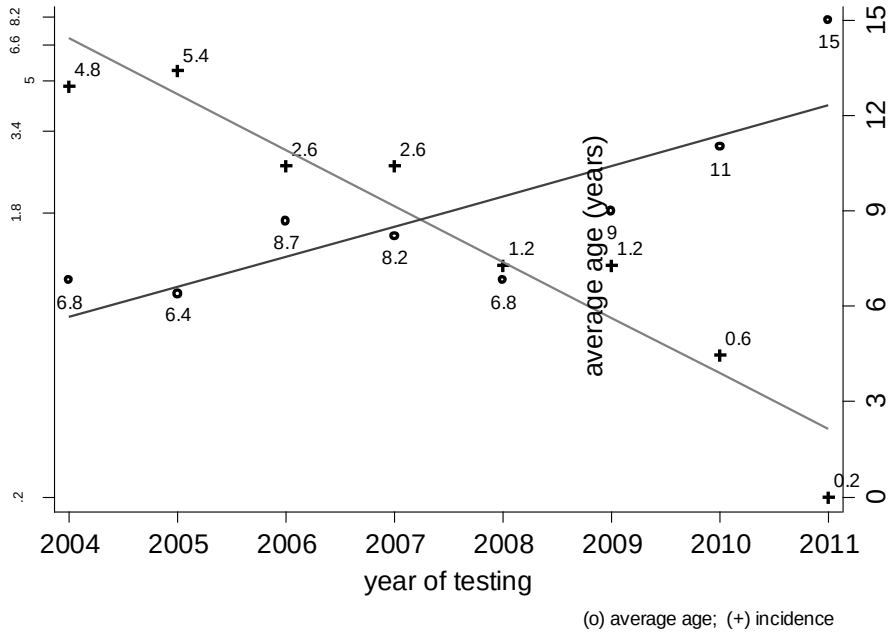

Figure 6: Incidence (log10 scale) of BSE in the EU8 per year of detection, considering both BSE Active and Passive Surveillance, and average age of those BSE cases for the given years.

When fitting regression models using year of testing as independent variable, and the log10 incidence or the average age at testing as dependent variables, both the declining trend in incidence and the

increasing trend in average age are statistically significant in the EU8 (beta values -0.18 and +0.95 for incidence and average age, respectively, with $p<0.001$ for both).

Concluding remarks on the trend of BSE in the EU8

- BSE has not been detected in five of the EU8 MSs (the EU5 group): Estonia, Hungary, Latvia, Lithuania and Malta. Three MSs (the EU3 group), these being Czech Republic, Poland and Slovakia, account for all the BSE cases detected in the EU8 group.

- The number of BSE cases in the EU3 decreased from 28 in 2005 (peak) to one in 2011.

- The log10 transformed annual BSE prevalence and incidence in the UE8 (defined respectively as the number of positive BSE cases out of the tested population and out of the standing adult cattle population) exhibits a statistically significant decreasing trend.

- There has been a statistically significant increasing trend in the average age of the detected BSE cases in the EU8 per test year over the last eight years, which currently exceeds 11 years in each of these MSs (where reported in 2011).

- When considering the birth cohort of the BSE cases in the EU3 there were two apparent consecutive waves of infections, the first in the mid 90's and a second one (with larger number of cases involved) around the year 2000.

- Assuming that the age distribution of cattle within the EU8 countries did not change substantially over the considered period, the decreasing trend observed in the log10 transformed annual BSE incidence and the increasing trend observed in the annual average age of the cases indicates that the transmission of BSE has decreased in the EU8 as a consequence of the implementation of the control measures.

3. Atypical BSE in cattle

Since its first report, Atypical BSE cases were described in a number of European and non-European countries. According to the data available in scientific literature or obtained through the EU active surveillance system, cases have been reported in several European countries (Jacobs et al., 2007; Stack et al., 2009), Japan (Masujin et al., 2008; Yamakawa et al., 2003), USA (Richt et al., 2007) and Canada (Dudas et al., 2010).

Atypical BSE cases reported by the different EU MSs since 2001 are presented in Table 5. These data have to be interpreted with caution as in the EU there is no legal requirement for typing BSE positive cases (i.e. all cases traditionally reported as BSE). Thus, the data presented here has been only reported in and *ad hoc* manner to the European Commission in the frame of the current and previously related mandates. Out of the 64 cases reported up to 2011, 37 were L-type and 27 H-type, respectively.

Table 5: Atypical BSE cases reported in the EU MSs since 2001. Source: European Commission.

Member State	Year of testing											
	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	**Total**
Austria							1			2		3
Denmark				1								1
France	1	3	4	1	1	2	2	5	4	3		26
Germany		1		1								2
Ireland		1								1	1	3
Italy		1	1				1		1		1	5
Netherlands	1	1	1							1		4
Poland		1[1]		2	2	2	2		1		1	11
Spain											1	1
Sweden						1						1
United Kingdom					1		2		1	1	2	7
Total	2	8	6	5	4	5	8	5	7	8	6	64

[1]Pre-May 2004 testing, before full implementation of EU regulations

Reported Atypical BSE cases were detected almost exclusively in animals over 8 years of age (except for one case in a 6 year old bovine reported in 2011). All these cases were identified by active surveillance testing. However, there is currently no data available on the performance of the validated rapid assays used for cattle TSE testing, for detection of Atypical BSE cases, both in terms of their analytical sensitivity and of the efficacy of detection of infected asymptomatic animals.

The number of Atypical BSE cases reported by EU MSs since 2001 by testing stream is included in table 6.

Table 6: Number of Atypical BSE cases reported by EU MSs since 2001 by testing stream. Source: European Commission.

	Year of testing											
Target group	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	**Total**
Fallen stock	1	3	4	2	2	2	7	4	4	6	4	39
Healthy slaughtered animals	1	5	2	2	2	3	1		2	2	2	22
Emergency slaughter				1				1	1			3
Total	2	8	6	5	4	5	8	5	7	8	6	64

Data presented in table 6 indicate that over the past decade the majority of the reported Atypical BSE cases are in the fallen stock and healthy slaughter target groups (around 60% and 35% respectively),

while only a few cases were detected in the emergency slaughter target group (around 5%). The number of Atypical BSE cases reported seems to be also rather stable throughout the years. In the EU17 group of MSs, where the majority of the BSE cases have been found, most of the cases identified over the past decade via active surveillance have been reported in the fallen stock target group.

Figure 7 shows the average age of the Atypical L-BSE and H-BSE cases reported in the EU since 2001

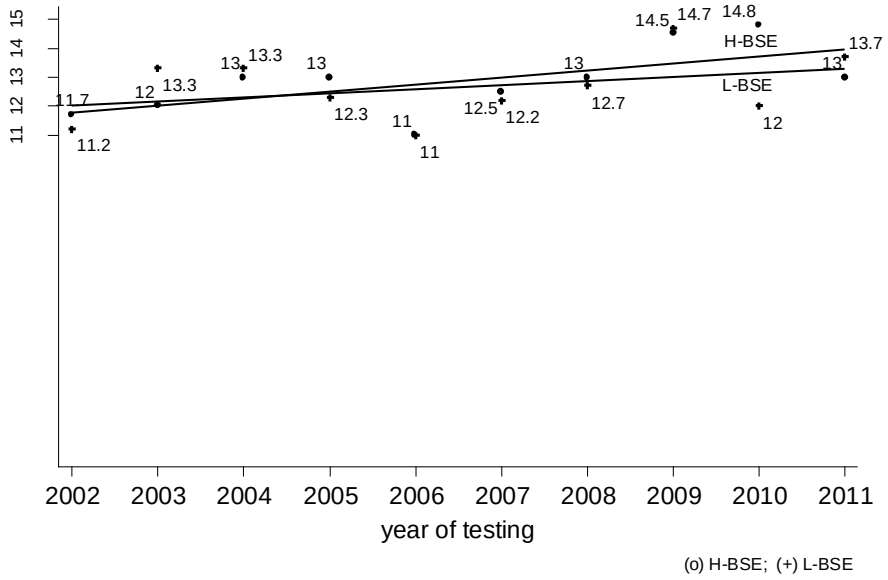

Figure 7: Average age of the Atypical L-BSE and H-BSE cases reported in the EU since 2001 to 2011.

There is a significant increasing trend of the average age of the Atypical BSE cases of the two types combined (i.e. H-BSE and L-BSE), (beta +0.24, p= 0.02) in the EU25.The slope of this trend is less pronounced that it is in the case of the increasing trend of the average age of the Classical BSE (C-BSE) cases. However, when assessing the trend individually for H-BSE and L-BSE, only a significant increasing trend in the yearly average age of the H-BSE cases is observed (beta +0.29, p=0.01), while for L-BSE the trend is not significant (beta=0.14, p=0.297).

3.1. Atypical BSE Type L (L-BSE)

Atypical L-BSE has been reported to be transmissible to different animal models. In particular, intra-cerebral (IC) inoculation of the L-BSE agent in cattle provokes a TSE which is both clinically and pathologically distinct from C-BSE (Lombardi et al., 2008). To date, there are no available results concerning the oral transmission of L-BSE in cattle.

Bioassay in transgenic (Tg) mice over-expressing the bovine PrP^{C} seems to indicate that infectivity might disseminate in skeletal muscle tissues in animals IC challenged with L-BSE (Suardi et al., 2012). In contrast no infectivity was detected in the kidney, in the spleen and in lymph nodes from the same animal. Both abnormal PrP^{Sc} deposits and infectivity (detected by bioassay in tg-Bov mice) was also observed in a naturally infected but asymptomatic BSE L case (Polak and Zmudzinski, 2012; Suardi et al., 2012).

In Classical BSE cases, pathogenesis studies have established that abnormal PrP deposition in the brainstem first occurs at the obex level, where substantial amount of this disease specific protein accumulates during the late incubation phase (Arnold et al., 2007; Simmons et al., 2010; Wells et al.,

2007). As a consequence, targeting obex for C-BSE rapid testing is considered as the most sensitive approach for detecting cases within the framework of the active surveillance system.

In Atypical BSE (both L and H type), the dynamics of the abnormal PrP deposition in the different brain areas is poorly documented, and the suitability of the obex as the target tissue for testing that would allow an early and sensitive detection of these conditions remain largely unknown. On one hand, all the Atypical BSE cases detected so far were identified through the active surveillance system, indicating that obex testing with currently validated tests allows the detection of at least a part of the Atypical cases. However, on other hand the distribution of abnormal PrP in L-BSE, as observed from a very limited number of samples, clearly indicates that brainstem deposition of abnormal PrP in the context of L-BSE is poor by comparison to other areas (Casalone et al., 2004; Konold et al.; Polak and Zmudzinski, 2012). This last finding strongly support the contention that active surveillance system as currently applied could have a more limited sensitivity to detect Atypical BSE cases than C-BSE cases in field cattle population.

Proof of principle of the L-BSE ability to propagate in sheep was brought by the IC propagation of a L-BSE isolate into ARQ/ARQ and in Tg mice expressing the ovine PrP^{C} variants. The propagation of L-BSE in sheep seemed to result in a TSE with a different profile to that of C-BSE (Nonno et al., 2008). Unexpectedly, L-BSE isolates transmitted to either Tg mice expressing ovine PrP^{C} (Beringue et al., 2007) or inbred wild-type mouse lines (Capobianco et al., 2007) resulted in a disease with similar phenotypic features to those of the C-BSE agent. However, the inoculation of tissues collected in mice over-expressing ovine PrP^{C} inoculated with C-BSE and L-BSE in bovine PrP^{C} transgenic mice, resulted into two different phenotypes specific of each agent indicating that the tg338 passaged agents, although producing a similar signature in the brain, were actually different (Beringue et al., 2010).

Results from several studies that focus on the potential human risk from Atypical L-BSE are available. Kong and colleagues (2008) investigated the infectivity and phenotype of L-BSE by IC inoculating Tg mice expressing the human PrP^{C} (M129M) with brain homogenates from two L-BSE affected cattle. Sixty percent of the inoculated Tg mice became infected after 20-22 months incubation, a transmission rate higher than those reported for C-BSE. A quarter of L-BSE infected Tg mice, but none of the Tg mice infected with sporadic CJD (sCJD), showed presence of PrP^{res} in the spleen, indicating that the L-BSE agent may be lymphotropic. The pathological prion protein isoforms in L-BSE infected humanized Tg mouse brains were different from those of the original cattle L-BSE or sCJD. Minimal brain spongiosis and long incubation time were observed in the L-BSE infected Tg mice. A similar study was performed in another Tg mice expressing the human PrP^{C} (M129M- Tg650) (Beringue et al., 2008). In contrast with C-BSE prions, L-BSE prions appeared to propagate in these mice with no obvious transmission barrier. Another study evaluated the transmission of L-BSE to a non-human primate (Comoy et al., 2008). Brain homogenates from cattle with C-BSE and L-BSE were IC inoculated into cynomolgus monkeys (*Macacca fascicularis*). The single monkey infected with L-BSE had a shorter survival, and a different clinical evolution, histopathology, and prion protein (PrP^{res}) pattern than what was observed for either C-BSE or vCJD-inoculated animals. These results were interpreted to suggest a possibly higher degree of pathogenicity of L-BSE than C-BSE in primates.

Taken together, these experimental studies may demonstrate that L-BSE or BASE is easily transmissible to both humanised mice and primates, and may be more virulent to humans than C-BSE.

More recently transmission of L-BSE into bank voles resulted in a TSE which phenotype (incubation period, PrP^{Sc} biochemical properties and vacuolar lesion profiles) were identical to the one observed after transmission of a VV2 s-CJD case in this rodent model (Nonno et al., 2009).

Finally, it has to be mentioned that there is no data available about the impact of the TSE inactivation process currently applied to processed animal proteins (133°C, 2 Bar pressure, 20 minutes) on the infectivity of the L-BSE agent.

3.2. Atypical BSE Type H (H-BSE)

There is currently no data available on the pathogenesis and the tissue infectivity distribution of H-BSE in ruminants.

H-BSE has been transmitted into a number of laboratory animal models. In most of the reported cases the transmission features obtained were distinct from those observed after inoculation with C-BSE (Beringue et al., 2006). However, a recently published work described the transmission of four French and one Polish H-BSE isolates into transgenic mice expressing bovine PrP^C (Tg110 mice) by IC challenge (Torres et al., 2011). Following these transmissions, two H-BSE isolates resulted in the propagation in some mice (respectively 3 and 2 out of 12) of a TSE displaying a C-BSE phenotype. Second passage of prions into TgBov mice confirmed that the TSE agent was C-BSE.

These results imply that C-BSE might emerge spontaneously from an H-BSE type isolate (in the absence of any interspecies passage), which could indicate that H-BSE might be a source of the C-BSE agent. This hypothesis is also consistent with the observed trend in the average age of cases.

Finally and equally to L-BSE, there is no data about the impact of the TSE inactivation process currently applied to processed animal proteins on the infectivity of the H-BSE Agent.

3.3. Concluding remarks on Atypical BSE

The following can be concluded regarding Atypical BSE:

- The origin and pathogenesis of Atypical forms of BSE in its natural host are unknown.
- Epidemiological data reported by the EU MSs indicate that over the last years the number of detected Atypical BSE cases did not show any trend and that these cases were mainly identified in the fallen stock and healthy slaughtered animals older than 8 years of age.
- The performance of the current TSE monitoring system, both in terms of its analytical sensitivity and earliness of the detection of animals infected with Atypical BSE is unknown.

4. A brief overview of existing BSE surveillance and risk models

Modelling has been applied quite intensively on BSE, with different aims and using different approaches to adapt to available data.

Existing BSE surveillance and risk models can be separated in two main categories: Models for data-rich situations and models for data-sparse situations. Data-sparse models are quite rare, and are generally risk models, that look at the risk of introduction or of transmission of BSE considering available test samples results. The data-rich models again separate into two categories: statistical models and predictive models, where the predictive models are generally an extension of a statistical data analysis model.

4.1. Data-sparse models

Data-sparse models (i.e. sparse case data) are rare. The EFSA Geographical BSE Risk Assessment is a good example of a reasonably good assessment of future risk was made based on rather limited information, (EFSA, 2007).

For parameter-sparse models, the OIE point system[20] for evaluation of surveillance is also available. This is not a model in the classical sense but to be considered as a criterion, which was based on evaluation of EU case data from the early active surveillance period, based on the BSurvE model (Prattley et al., 2007). Various risk categories were determined with relative risk, leading to the points

[20] For further details see: http://www.oie.int/fileadmin/Home/eng/Health_standards/tahc/2010/en_chapitre_1.11.5.htm

accredited to these categories. Although extrapolation of these risk estimates to other situations and non-randomized surveillance data is not scientifically sound, it surely offers a very simple evaluation method.

4.2. Statistical models

The statistical models analyse existing BSE case data and negative test data, aiming to provide 1.) a prediction of the prevalence in the selected population, 2.) to analyse the BSE trend in time, or 3.) to increase the sensitivity of the surveillance system. Most of these models include the age structure and an age-dependent case-risk functions.

Examples of these models are the BSurvE model (Prattley et al., 2007), models considered in former EFSA Opinions (EFSA, 2008a, 2009, 2010; Prattley et al., 2007) and that published in 2008 by de Koeijer (de Koeijer, 2008). A similar approach of analysing the trend of BSE has been the application of Age-Period-Cohort (APC) regression models (Ducrot et al., 2010; Sala and Ru, 2009). APC modelling, commonly used in the assessment of cancer trends, is based on the analysis of tested animals (in term of age at testing, date of testing i.e. period, and birth cohort): it may provide a calculation of the evolution of BSE risk over successive birth cohorts allowing the comparison of trends by country and the identification of difference in the probability of infection of subsequent cohorts.

Back calculation models are very detailed models offering more precision than more simple models. These models derive several transmission relations, like the age dependent susceptibility, age dependent mortality etc. simultaneously from the data, while also analysing (historical) prevalences.

4.3. Predictive models

Predictive extensions to the statistical models are made based either on linear extrapolation or on mechanistic analysis. Although back-calculation models are in theory less suitable for extrapolation, this can be solved as easily as with the less detailed categorizing models. However, the precision gained in the model will get lost with the extrapolation, while such a model remains more “black-box”- like, in the sense that all the computations performed within the modelling process are not easily evident.

5. Estimating sample sizes of healthy slaughtered cattle over 72 months of age for the monitoring and surveillance of TSEs

Following an EFSA open call for tender (CT/EFSA/BIOHAZ/2002), a model to estimate sample sizes of healthy slaughtered cattle to be tested in order to meet a given design prevalence has been provided by a contractor (Animal Health and Veterinary Laboratories Agency (AHVLA), UK). This model was called the Cattle-TSE Monitoring Model, to be referred to as C-TSEMM from now onwards.

The full report submitted to EFSA by the contractor should be read as part of this EFSA Scientific Report (Adkin et al., 2012).

5.1. Methodology, limitations and assumptions

The probabilistic statistical C-TSEMM model employs a framework developed in R software[21] with a Visual Basic[22] software interface. The detailed description of the structure of the model can be found in the contractor's Report (Adkin et al., 2012).

The model uses EU data on prevalence of infection by birth-cohort and demographic information, which is employed to estimate the design prevalence and the sensitivity of a monitoring system. European Commission and EU member states provided the relevant data. It has to be noticed that the models estimates presented in this report are based on the 2011 demographics of the adult cattle population and on the number of adult cattle removed from the population via the different streams (i.e. healthy slaughter, animals showing clinical signs of disease during *ante mortem* inspection, emergency slaughtered animals and fallen stock). Therefore, future fluctuations in those numbers at EU level and in each MSs will impact the validity of current estimates. Thus, should the C-TSEMM model be employed in future years for the review of the BSE monitoring regime in the EU, updated yearly data including BSE testing data have to be considered as these drive the results estimated by the model.

Based on the model approach, the number of animals of the healthy slaughter exit stream that need to be tested, in order that at least one animal is detected with a probability of τ, is given by using binomial formulae. This is an approximation to the hypergeometric distribution, which would be the correct distribution for a sampling scheme without replacement.

Testing for BSE is a testing procedure without replacement, but the approximation by the binomial distribution is acceptable is $n < 0{,}1N$. If we compare the means and the standard deviations of the binomial and the hypergeometric distributions we immediately see that the means are identical but the standard deviations differ by the factor $\mathrm{sqrt}((N-n)/(n-1))$. This factor is called the finite populations factor and is always less or equal to one. If $n<0{,}1N$, the corresponding binomial and hypergeometric probabilities correspond close enough (see C-TSEMM sensitivity analysis, section 5.4. of the contractor's Report, (Adkin et al., 2012)). In the simulation model the hypergeometric distribution is replaced by the binomial distribution because the model becomes computationally more tractable.

When dealing with the relationship between sample size and design prevalence in this report, calculations are presented taking into account both the standing adult cattle population and the tested adult cattle population, both of them older than 24 months of age. Thus, the definitions of prevalence in the contractor's Report (Adkin et al., 2012) and as presented in this section of the EFSA Report here are as follows:

- **Detectable prevalence in test population**: Period prevalence in a given year of detectable infected animals in the test population. Calculated by the model's predicted number of adult animals (>24 months), in the population of animals tested, that would test positive by a diagnostic test, divided by the total number of animals tested in one year.

[21] www.cran.r-project.org
[22] © Microsoft Cooperation www.micosoft.com

- **Infection prevalence in test population**: Period prevalence in a given year of infected animals in the test population. Calculated by the model's predicted number of adult animals (>24 months), in the population of animals tested, that are actually infected (i.e. animals that may or may not test positive or be showing clinical symptoms) divided by the total number of animals tested in year.

- **Detectable prevalence in standing population**: Period prevalence in a given year of detectable infected animals in the standing population. Calculated by the model's predicted number of adult animals (>24 months), in the standing population, that would test positive by a diagnostic test, divided by the total number of adult animals in the standing population.

In the context of BSE monitoring, addressing prevalence in the tested cattle population is usually considered of greater epidemiological value than addressing prevalence in the standing cattle population. This is because the population of study of BSE is the tested population, which is not a random representation of the standing population but part of the population that is slaughtered (or dead). Furthermore the predicted detectable prevalence in the standing population can never be validated. Nevertheless, results are presented considering also the standing population as this forms part of the technical request made by the Commission.

A series of assumptions are made in the C-TSEMM, which need to be considered when evaluating the outputs from the model (Adkin et al., 2012). These assumptions can be categorised in two groups:

a. **Assumptions made when transforming input data**. These include:

   - For MSs with no, or few, BSE cases post-2001 (i.e. Austria, Cyprus, Estonia, Finland, Greece, Hungary, Latvia, Lithuania, Luxembourg, Malta and Sweden) an alternative estimate of cohort-based prevalence is required. This has been estimated for those MSs based on the average prevalence of the group of MSs with BSE cases under which they were placed in previous related EFSA Opinions (EFSA, 2009, 2010), theEU17 or the EU8 group[23]. This results in an overestimate of prevalence for countries with no recorded cases as they are assumed to be a merged epidemiological unit with countries where cases are observed (see Appendix C of the contractor's report (Adkin et al., 2012) for further details).

   - Whilst the healthy slaughter, emergency slaughter, fallen stock and clinical suspects of BSE seem to be populated to a similar degree within European countries, clinical signs at *ante mortem* does not seem to be uniformly applied. When considering the definition of the emergency slaughter category there appears little to distinguish between the categories and therefore it has been agreed that the clinical signs at AM stream can be merged into the emergency slaughtered stream.

   - Animals culled under the eradication measures are traditionally difficult to include in modelling work as for most countries there are insufficient test positive data to estimate prevalence on a cohort basis. These were incorporated into the fallen stock category with the impact of this assumption investigated in section 5.1 of the contractor's Report (Adkin et al., 2012).

   - The proportion of animals > 155 months in the (i) slaughtered/dead, and (ii) standing population, by 12 month intervals to 204 months (17 years) is not known for most MSs. Therefore, the assumption was made that the proportions by 12 monthly intervals could be approximated by that recorded in (i) the UK slaughtered/dead population between 2008

[23] EU17: Austria, Belgium, Cyprus, Denmark, Finland, France, Germany, Greece, Ireland, Italy, Luxembourg, Netherlands, Portugal, Slovenia, Spain, Sweden and United Kingdom. EU8: Czech Republic, Estonia, Hungary, Latvia, Lithuania, Malta, Poland and Slovakia.

and 2010, and (ii) an average of that recorded in Austria and the UK standing population in 2010. Assumption (i) has been tested in the sensitivity analysis and could be replaced by assuming equal proportions by 12 monthly intervals without impacting results (refer to section 5.2 of the contractor's Report (Adkin et al., 2012)).

- Data are absent for the standing population for most MSs in 2011. It is assumed that the data for 2010 can be used as a proxy.

- Age data are transformed into cohort based testing data assuming an equal probability per month of birth and death.

- There are little experimental data to assess the sensitivity of the BSE test in cattle. It is assumed that we can use data based on 1g experimentally dosed cattle as detailed in Arnold et al. (2007) to approximate the sensitivity of the test for field cases.

b. **Model assumptions**. These include:

- Underpinning the estimate of true prevalence it is assumed that the use of an exponential distribution to model the true prevalence is appropriate. While other distributions could be fitted, analysis of alternative distributions has indicated that an exponential decay of prevalence over time is appropriate for the majority of European data (section 5.3 of the contractor's Report (Adkin et al., 2012)).

- Cases from the clinical suspect (CS) stream and fallen stock (FS) stream are assumed to be identified at the end of the incubation period, that is, death is as a result of the disease. Whereas healthy slaughter (HS) and emergency slaughter (ES) animals may be within a period of time before clinical onset depending on the distribution of the age at onset and test sensitivity. This assumption impacts the number of infected animals within these streams.

- Prevalence estimated for the combined streams (i.e. clinical suspects and fallen stock, healthy slaughter and emergency slaughter) can be divided into the individual exit streams according to the proportion of test positive animals observed in those streams. Where there are no test positives, it is assumed that the number of animals tested by birth cohort and testing year is an appropriate proxy.

- It is assumed that all cases of BSE are typed by strain such that the number tested for Classical and unknown strains is the same number as that tested for Atypical H and L type. This is not the case for all MSs and therefore only simulations from MSs where strain differentiation is routinely conducted will be valid.

- The design prevalence calculation is based on an infinite population (sampling with replacement) which is based on the binomial distribution. This method is straightforward to implement, however, for MSs with small slaughter populations, the use of the hypergeometric distribution produces lower estimates for the number to test to achieve a desired design prevalence. This has been investigated in the sensitivity analysis (section 5.4 of the contractor's Report (Adkin et al., 2012)). The conclusion is that for those countries with a small slaughter population, the number of animals needed to be tested is still greater than the number that are actually tested with the exception of Finland which has a marginal reduction in the number to test using the hypergeometric equation. For all other MSs that are not achieving a sufficient design prevalence to reduce current levels of testing, conclusions are not affected whether the hypergeometric or binomial based sample size formula is used. The impact of the use of the binomial could be further explored with the application in the field of a "sampling with replacement" strategy in BSE monitoring. However, that is not currently feasible under the current BSE testing

practices (i.e. *post mortem* testing) and may proof challenging even with a theoretical live animal testing BSE monitoring regime.

- In estimating either the re-emergence of an existing TSE, or emergence of a new TSE disease in cattle, it is assumed that the disease can be detected by current testing assays.

- For simulating the EU25 as a whole, it is assumed that it can be merged as an unique epidemiological unit or territory.

5.2. Results

5.2.1. Estimated design prevalence of the current BSE monitoring system

Table 7 shows the results for different design prevalence calculations that would be detected by the baseline monitoring regime in place in the EU25 as a whole and by the MSs individually with τ% confidence. Results are provided based on the detectable prevalence (prevalence of cases) for the adult standing population and adult tested population, together with results based on the infection prevalence (prevalence of infected animals) for the adult tested population. This considers all cases recorded in the Commission BSE database including the "unknown" strains.

The baseline monitoring regime is the testing of healthy slaughter animals > 72 months, emergency slaughter and fallen stock > 48 months and the testing of all clinical suspect animals. Results are expressed as 1 in X, so a result of 100 000 indicates that we would expect the current system to detect a prevalence in adult cattle >24 months of 1 in 100 000. For the main results τ=95%, to show the uncertainty surrounding these estimates we also present results for τ=92.5% and τ=97.5%. Design prevalence results are shaded where the estimated prevalence detected is greater than the threshold of 100 000. As the level of confidence is increased from τ=92.5% to τ=97.5%, it can be seen from the table that the estimated design prevalence reduces in sensitivity. N/A in Table 7 indicates that model has failed to converge, and thus not being able to provide estimates based on the country-specific data characteristics of those MSs

Table 7: Estimated design prevalence of baseline monitoring system for all strains, using detectable prevalence in the tested population and standing population, and infection prevalence in the tested population to a confidence (τ) of 95% (lower 92.5% and upper 97.5% confidence). Shaded results show that the estimated prevalence detected is greater than the threshold of 100 000.

MS	Estimated 'design prevalence' of baseline monitoring system considering all strains and unknown								
	Detectable prevalence in standing population (1 in X)			Detectable prevalence in tested population (1 in X)			Infection prevalence in tested population (1 in X)		
	τ=0.925	τ=0.95	τ=0.975	τ=0.925	τ=0.95	τ=0.975	τ=0.925	τ=0.95	τ=0.975
EU25	7 349 693	6 354 930	5 160 828	2 304 889	1 992 928	1 618 454	706 953	611 268	496 410
AT	142 490	123 205	100 056	55 885	48 321	39 242	14 318	12 380	10 054
BE	323 067	279 342	226 854	74 519	64 433	52 326	20 168	17 438	14 162
CY	6 136	5 306	4 310	N/A	N/A	N/A	461	N/A	N/A
CZ	60 099	51 965	42 201	31 586	27 311	22 181	5 766	4 985	4 049
DE	899 533	777 784	631 638	323 633	279 831	227 250	81 585	70 542	57 287
DK	N/A	N/A	N/A	44 659	N/A	31 359	15 408	13 323	10 819

MS	Estimated 'design prevalence' of baseline monitoring system considering all strains and unknown								
	Detectable prevalence in standing population (1 in X)			Detectable prevalence in tested population (1 in X)			Infection prevalence in tested population (1 in X)		
	τ=0.925	τ=0.95	τ=0.975	τ=0.925	τ=0.95	τ=0.975	τ=0.925	τ=0.95	τ=0.975
DK*	274 347	237 147	192 626	44 662	38 612	31 357	15 414	13 333	10 825
EE	16 443	14 220	11 548	6 239	5 395	4 381	1 319	1 140	N/A
EL	N/A	N/A	N/A	8 076	6 983	5 672	2 668	2 307	N/A
ES	294 174	254 359	206 565	141 902	122 696	99 624	43 943	37 995	30 856
FI	108 035	90 365	74 692	N/A	17 661	14 343	6 219	5 378	4 367
FR	2 005 412	1 733 985	1 408 168	603 303	521 647	423 629	236 882	204 821	166 335
HU	37 393	32 332	26 257	19 805	17 124	13 907	3 800	3 286	2 669
IE	527 100	455 760	370 123	127 587	110 318	89 590	56 919	49 215	39 968
IT	319 226	276 020	224 155	139 500	120 619	97 937	24 298	21 010	17 062
LT	28 060	N/A	19 703	21 459	18 555	15 069	3 301	N/A	2 318
LU	1 4 896	12 881	10 461	2 926	2 530	2 055	1 273	1 101	894
LV	18 833	16 284	13 224	11 155	9 646	7 833	1 689	1 460	1 186
MT	N/A	N/A	545	N/A	N/A	N/A	115	100	N/A
NL	336 577	290 828	236 340	110 685	94 162	77 722	29 881	25 836	N/A
PL	210 665	182 152	147 926	166 199	143 704	116 702	28 646	24 769	20 115
PT	73 615	63 652	51 692	N/A	N/A	N/A	9 720	8 405	6 825
SE	116 259	101 010	81 484	29 845	25 805	20 957	7 675	6 636	5 389
SI	17 683	15 290	12 417	8 115	7 017	5 699	2 355	2 036	1 654
SK	16 423	14 200	11 532	8 884	7 682	6 239	1 842	1 592	1 293
UK	785 476	679 164	551 548	246 037	212 736	172 763	62 482	54 025	43 874

N/A=the model has failed to find a viable value.
*Values for Denmark using alternative solver routine

From Table 7 it can be seen that the calculation using the **detectable prevalence in the standing population** produces the highest estimates for the design prevalence the baseline monitoring system is able to detect. Twelve MSs (i.e. Austria, Belgium, Denmark, Germany, Spain, France, Ireland, Italy, Netherlands, Poland, Sweden and the UK) have a design prevalence of at least 1 in 100 000 using the estimated detectable prevalence to a confidence level of 95%. The EU25 'design prevalence' is higher than for individual MSs as far more animals are tested, with an estimated design prevalence of 1 in 6 354 930 .

N/A in Table 7 indicates that model has failed to converge. This is due to the use of a solver to calculate the estimated 'design prevalence' a monitoring system is able to detect by the rearrangement of the design prevalence equation (see Equation 2 in Appendix B, contractor's Report (Adkin et al., 2012)), where the 'design prevalence' value is solved for a specified number of animals tested. The generic solver routine has been optimised to produce results for the majority of MSs. When considering the detectable prevalence in the standing population, a viable value has not been found at the 95th confidence value for Greece, Lithuania, and Malta. Based on other confidence values and the estimated number to test values provided in Table 8 below, the design prevalence of these countries is not meeting the 1 in 100 000 threshold. However, for Denmark it is likely that at the 95th confidence value, the monitoring system is detecting greater than 1 in 100 000. To investigate that value for Denmark, the generic solver routine was adapted specifically for Denmark. Results using the specific solver routine for Denmark are denoted in the table with an asterisk.

For the remaining MSs in Table 7 (i.e. Czech Republic, Cyprus, Estonia, Finland, Hungary, Luxembourg, Latvia, Portugal, Slovenia and Slovakia) the design prevalence the baseline monitoring system is able to detect ranges between 1 in 5 306 and 1 in 90 365 with a confidence level of 95%.

Using the estimated **detectable prevalence in the tested population** to a confidence level of 95%, the baseline monitoring regimes in seven MSs (Germany, Spain, France, Ireland, Italy, Poland and UK) have a design prevalence of at least 1 in 100 000. Additionally, France has a design prevalence greater than 1 in 100 000 estimated using the **prevalence of infection in the tested population**. The EU25 'design prevalence' is higher than for individual MSs as far more animals are tested, with an estimated design prevalence of 1 in 1 992 928 using the detectable prevalence in the tested population and 1 in 611 268 using the prevalence of infection in the tested population. The assumption is made that the EU25 can be estimated as a merged epidemiological unit or territory.

5.2.2. Estimated number to test in healthy slaughter stream to achieve a design prevalence of 1 in 100 000

Table 8 shows the number of healthy slaughter animals that would need to be tested, given the number of animals currently being tested in the other exit streams remains the number tested in those streams in 2011, in order to be τ(%) confident of detecting a positive animal if the overall prevalence in animals >24 months is 1 in 100 000. As in table 7, results are provided based on the detectable prevalence (prevalence of cases) for the adult standing population and adult tested population, together with results based on the infection prevalence (prevalence of infected animals) for the adult tested population. For the main results τ=95%, to show the uncertainty surrounding these estimates we also present results for τ=92.5% and τ=97.5%. Results for the number of healthy slaughter animals to be tested are shaded where the estimated number is less than current testing in this exit stream. This considers all cases recorded in the Commission BSE database including the "unknown" strains.

Table 8: Estimated number of health slaughtered animals required to be tested for all strains, given testing of emergency slaughter, fallen stock and clinical suspect animals, to achieve a design prevalence of 1 in 100 000 using detectable prevalence in the tested population and standing population, and infection prevalence in the tested population to a confidence (τ) of 95% (lower 92.5% and upper 97.5% confidence). Shaded results show that the estimated prevalence detected is greater than the threshold of 100 000.

		Number to test in healthy slaughter to detect prevalence of 1 in 100 000 considering all strains and "unknown"								
		Detectable prevalence in standing population			Detectable prevalence in tested population			Infection prevalence in tested population		
MS[1]	Actual number tested in HS >72 m (2011)	τ=0.925	τ=0.95	τ=0.975	τ=0.925	τ=0.95	τ=0.975	τ=0.925	τ=0.95	τ=0.975
EU25	3 730 778	0	0	0	0	0	0	0	0	0
AT	104 147	40 014	63 640	104 029	273 920	334 160	437 140	1 391 111	1 626 229	2 028 165
BE	112 059	0	0	0	206 255	264 120	363 041	1 202 488	1 416 297	1 781 805
CY	2 140	86 023	100 011	123 925	317 062	367 215	452 954	1 187 398	1 373 789	1 692 427
CZ	42 984	89 981	108 418	139 936	196 307	231 387	291 358	1 199 928	1 392 110	1 720 646
DE	513 746	0	0	0	0	0	0	786 379	1 018 122	1 414 290
DK	55 260	0	0	0	304 008	374 366	494 643	1 157 284	1 361 209	1 709 820
EE	7 739	80 328	93 927	117 189	222 438	258 282	319 557	1 076 700	1 246 265	1 536 138
EL	12 428	175 599	207 618	262 356	483 513	563 732	700 865	1 522 610	1 765 482	2 180 675
ES	255 669	0	0	0	13 615	104 038	258 615	1 301 366	1 593 365	2 092 539
FI	27 041	22 168	34 146[2]	54 623	344 146	406 524	513 161	1 254 421	1 459 289	1 809 512
FR	1 013 355	0	0	0	0	0	0	0	0	0
HU	24 700	94 256	111 647	141 376	192 920	225 755	281 887	1 076 287	1 247 399	1 539 917
IE	241 637	0	0	0	0	132 508	377 213	1 124 731	1 445 597	1 994 120

MS[1]	Number to test in healthy slaughter to detect prevalence of 1 in 100 000 considering all strains and "unknown"								
	Actual number tested in HS >72 m (2011)	Detectable prevalence in standing population			Detectable prevalence in tested population			Infection prevalence in tested population		
		τ=0.925	τ=0.95	τ=0.975	τ=0.925	τ=0.95	τ=0.975	τ=0.925	τ=0.95	τ=0.975
IT	255 135	28 898	45 052	72 667	161 855	198 821	262 014	1 281 486	1 493 712	1 856 516
LT	41 066	159 684	185 494	229 616	210 401	244 149	301 843	1 396 379	1 615 774	1 990 832
LU	3 738	107 523	126 612	159 243	606 397	703 576	869 704	1 412 466	1 635 822	2 017 652
LV	21 766	125 166	145 106	179 195	212 838	246 502	304 051	1 418 127	1 640 460	2 020 541
MT	416	95 368	110 347	135 954	115 067	133 130	164 008	647 505	748 912	922 270
NL	165 855	0	0	0	125 683	184 533	285 137	1 142 364	1 360 358	1 733 022
PL	310 559	103 168	132 503	182 651	153 308	190 491	254 057	1 293 962	1 509 697	1 878 497
PT	43 450	96 547	128 047	181 897	384 982	461 632	592 667	1 419 381	1 657 950	2 065 786
SE	45 963	31 314	45 417	69 525	292 182	347 119	441 035	1 305 964	1 519 593	1 884 793
SI	10 595	159 945	188 345	236 895	373 843	435 725	541 514	1 340 888	1 554 145	1 918 712
SK	9 721	114 198	133 766	167 217	220 263	256 433	318 266	1 103 870	1 278 355	1 576 639
UK	409 609	0	0	0	0	0	82 639	875 767	1 070 258	1 402 744

[1] See Appendix B for MS acronyms
[2] In the case of Finland the number of healthy slaughter cattle to test using the hypergeometric equation is estimated to be 16 2333, less than the number tested in 2011

Considering the estimated **detectable prevalence in the standing population,** in Table 8 we can see that when the EU25 is merged into one epidemiological unit, the area already tests sufficient animals in the ES, FS and CS streams such that they do not need to test any healthy slaughter animals (represented in the table by a value of 0). Thus, the C-TSEMM model estimates that, with the current BSE monitoring regime but excluding the testing of healthy slaughter cattle, the system is able to detect in the standing population one BSE case in 4 021 940 adult cattle with a confidence level of 95%.

Also considering the estimated detectable prevalence in the standing population, at a confidence level of 95%, eight MSs (i.e. Belgium, Germany, Denmark, Spain, France, Ireland, Netherlands, and the UK) do not require the testing of any healthy slaughter animals to meet a 1 in 100 000 design prevalence.

Also based on detectable prevalence in the standing population, four MSs (i.e. Austria, Italy, Poland and Sweden) do require testing less healthy slaughtered animals older than 72 months of age than the total number tested in those MSs in 2011 in order to meet a 1 in 100 000 design prevalence. In the remaining thirteen MSs (i.e. Cyprus, Czech Republic, Estonia, Finland, Greece, Hungary, Latvia, Lithuania, Luxembourg, Malta, Portugal, Slovenia and Slovakia) the number of healthy slaughtered animals older than 72 months of age that would need to be tested in order to meet a 1 in 100 000 design prevalence is higher than the actual number tested in 2011. In the case of Finland, the number of healthy slaughter cattle to test if the hypergeometric equation would be used instead of the binomial one is estimated to be less than the number tested in 2011.

However, it has to be noted that in the context of the current BSE monitoring system (i.e. *post mortem* test carried out in slaughtered cattle or dead cattle of certain age categories), fitting a sample size larger than the actually slaughtered cattle population is neither feasible nor realistic. Thus, the current testing of all animals of certain age categories that are slaughtered or dead, may provide the most sensitive BSE monitoring system possible (i.e. that employs *post mortem* tests) under the current epidemiological scenario with the potential limitation on the impact of the age at testing as evaluated in former EFSA Opinions (EFSA, 2008a, 2008b, 2009, 2010).

One potential consideration could be given to those MSs where the sample size is larger than the actual cattle population size. In that case, an alternative could be to consider extending (e.g. from one to more years) the time frame given to meet the design prevalence. However, this would not be in line with the technical aspects of the terms of reference (i.e. yearly design prevalence).

The calculations using the standing population prevalence show the lowest number of animals that need to be tested when compared to results using the detectable prevalence in the test population. The estimate of the numbers of animals needing to be sampled in order to detect a prevalence of 1 in 100 000 is lower (and thus the power of the surveillance in Table 8 is higher) when considering the standing population than when considering the test population. This is because the prevalence of BSE in the standing population is lower than the prevalence in the test population. As such, assuming a design prevalence of 1 in 100 000 in the standing population, as opposed to in the test population, will lead to higher stream prevalences in the test population after the appropriate scaling. In other words, a design prevalence of 1 in 100 000 in the standing population will lead to a greater than 1 in 100 000 prevalence in the test population (the design prevalence used when considering the test population). Therefore, the standing population prevalence calculations are effectively performed at a higher overall BSE prevalence than the test population calculations, leading to smaller sample sizes.

Using the estimated **detectable prevalence in the tested population**, we can see that when the EU25 is merged into one epidemiological unit, the area already tests sufficient animals in the ES, FS and CS streams such that they do not need to test any healthy slaughter animals (represented in the table by a value of 0). At a confidence level of 95%, three MSs (i.e. Germany, France, and the UK) do not require to test any healthy slaughter animals to meet 1 in 100000 design prevalence given the other exit streams are tested. Italy (IT), for example, with a confidence level of 95%, is required to test 198 821 HS animals. As Italy (IT) currently tests 255 135 there is a reduction in the animals required to be tested to achieve the desired design prevalence. Luxembourg (LU) is required to test 703 576 HS animals, but only test 3 738 so that MS will not achieve the design prevalence.

When using the estimated **infection prevalence in the tested population** only France and the EU25 as a whole achieve the required confidence with no testing of healthy slaughter animals required. The differences in the results between MSs are based on the estimated ratio of the prevalence in each of the four testing streams and how many animals are tested per year in those streams. France, for example, has a relatively high prevalence in FS and CS testing streams and tests a large number of animals within these streams. Therefore, for France the design prevalence is met without the requirement for testing in the healthy slaughter stream.

5.2.3. Estimated number of BSE cases missed should healthy slaughtered cattle testing stop

Table 9 displays, based on simulations done by the model, the predicted number of BSE cases (i.e. this considers all cases recorded in the Commission BSE database including the "unknown" strains) detected by the monitoring baseline and scenario and the total number of infected animals slaughtered/dead over one year. The baseline monitoring regime involves the testing of healthy slaughter animals > 72 months, emergency slaughter and fallen stock > 48 months and the testing of all clinical suspect animals. The scenario regime only affects the healthy slaughter stream in that no healthy slaughtered animals are tested. The mean values are presented, together with the 95% confidence intervals in brackets. For comparison purposes, the actual number of test positives for 2011 are shown in the second column by exit stream. The final column on the right hand side displays the total number of infected animals slaughtered/dead (for all four streams including all age groups) irrespective the testing scheme applied.

Table 9: Estimated mean number of cases missed and infected animals missed of all strains given a change in the monitoring regime from the baseline to a scenario where no healthy slaughter animals are tested with 95% confidence intervals (CI*).

		Number of animals missed between monitoring baseline and scenario in one year considering all strains and "unknown"			
MS[1]	**Actual cases in 2011 {HS,ES,FS,CS}**	Baseline detected [CI*]	Scenario detected[CI*]	Number detected missed [CI*] (baseline - scenario)	Number infected animals dead [CI*] (all streams, all age groups)
EU25	28 { 9, 0, 19, 0 }	28 [11 , 148]	18 [5.6 , 132]	10 [5.2 , 16]	100 [59 , 247]
AT	0	0.53 [0.74 , 101]	0.27 [0.71 , 100]	0.25 [0.03 , 0.52]	2.4 [1.1 , 104]
BE	0	0.22 [0.71 , 100]	0.13 [0.7 , 100]	0.09 [0.01 , 0.19]	0.99 [0.81 , 102]
CY	0	0.03 [0.66 , 96]	0.02 [0.66 , 96]	0.01 [0.001 , 0.02]	0.12 [0.67 , 96]
CZ	0	0.86 [0.77 , 98]	0.34 [0.7 , 97]	0.52 [0.07 , 1.05]	7.5 [2.6 , 110]
DE	0	3.6 [1.3 , 107]	2 [0.98 , 104]	1.5 [0.36 , 2.79]	20 [7.9 , 132]
DK	0	0.01 [0.69 , 100]	0.01 [0.68 , 100]	0.004 [0.0004 , 0.01]	0.05 [0.69 , 100]
EE	0	0.07 [0.67 , 96]	0.03 [0.66 , 96]	0.04 [0.004 , 0.08]	0.52 [0.72 , 97]
EL	0	0.09 [0.69 , 100]	0.06 [0.69 , 100]	0.03 [0.003 , 0.06]	0.30 [0.72 , 100]
ES	7 { 4, 0, 3, 0 }	16 [5.7 , 128]	11 [3.4 , 120]	5 [2.3 , 7.92]	60 [32 , 190]
FI	0	0.23 [0.71 , 100]	0.16 [0.7 , 100]	0.08 [0.009 , 0.16]	0.98 [0.81 , 102]
FR	3 { 0, 0, 3, 0 }	2.9 [1.1 , 105]	2.2 [0.98 , 104]	0.69 [0.14 , 1.32]	8.2 [2.6 , 115]
HU	0	0.26 [0.66 , 93]	0.11 [0.64 , 92]	0.15 [0.018 , 0.32]	2 [0.94 , 96]
IE	3 { 0, 0, 3, 0 }	3.9 [1.3 , 107]	3.1 [1.1 , 106]	0.81 [0.18 , 1.51]	9.5 [3 , 117]
IT	1 { 1, 0, 0, 0 }	0.31 [0.72 , 100]	0.07 [0.69 , 100]	0.24 [0.027 , 0.50]	2.2 [1 , 104]
LT	0	0.2 [0.68 , 96]	0.02 [0.66 , 96]	0.17 [0.019 , 0.36]	1.7 [0.89 , 99]
LU	0	0.04 [0.69 , 100]	0.03 [0.69 , 100]	0.009 [0.001 , 0.02]	0.11 [0.7 , 100]
LV	0	0.11 [0.67 , 96]	0.01 [0.66 , 96]	0.098 [0.011 , 0.21]	0.95 [0.78 , 98]
MT	0	0.004 [0.66 , 96]	0.002 [0.66 , 96]	0.0025 [0.00026 , 0.01]	0.043 [0.66 , 96]
NL	0	0.4 [0.73 , 100]	0.24 [0.71 , 100]	0.16 [0.019 , 0.33]	1.8 [0.95 , 103]

MS[1]	Actual cases in 2011 {HS,ES,FS,CS}	Number of animals missed between monitoring baseline and scenario in one year considering all strains and "unknown"			
		Baseline detected [CI*]	Scenario detected[CI*]	Number detected missed [CI*] (baseline - scenario)	Number infected animals dead [CI*] (all streams, all age groups)
PL	1 { 1, 0, 0, 0 }	1.2 [0.82 , 98]	0.26 [0.69 , 96]	0.96 [0.14 , 1.93]	8.9 [3 , 112]
PT	5 { 3 0 2 0 }	3.1 [1.1 , 106]	2.2 [0.97 , 104]	0.91 [0.17 , 1.74]	11 [3.5 , 119]
SE	0	0.31 [0.72 , 100]	0.18 [0.7 , 100]	0.13 [0.02 , 0.26]	1.5 [0.89 , 103]
SI	0	0.39 [0.7 , 97]	0.26 [0.69 , 96]	0.13 [0.016 , 0.27]	1.9 [0.94 , 100]
SK	0	0.56 [0.73 , 97]	0.3 [0.69 , 97]	0.27 [0.034 , 0.54]	4.6 [1.6 , 105]
UK	8 { 0, 0, 8, 0 }	11 [3.2 , 120]	5.2 [1.5 , 110]	5.8 [1.7 , 10.29]	44 [20 , 171]

[1] See Appendix B for MS acronyms

*Confidence intervals are results from using upper and lower 95% Poisson confidence interval values about the model predictions

If testing in the healthy slaughter stream were to cease, an estimated mean 10 cases (95% CI: 5.2, 16) across the EU25 (when the EU25 is merged into one epidemiological unit) would have been missed in 2011. The estimated number missed can be compared against the background estimated number of 100 infected animals slaughtered/dead which includes those animals which, if tested, would test negative.

The number of missed cases estimated by the C-TSEMM model based on detectable cases can be compared with those estimated by the model employed in the EFSA Scientific Opinion of 2010 on a second update of the risk to human and animal health on the review of the BSE monitoring regime in some MSs (EFSA, 2010). That Scientific Opinion estimated that should testing of healthy slaughtered cattle be completely stopped but maintaining testing of the at risk animal group in animals older than 48 months of age, less than three Classical BSE cases would have been missed in the EU17 in 2011 under a "more realistic"[24] scenario. When comparing these results, the following has to be noted:

- The C-TSEMM model employs new BSE data collected in 2011;
- The C-TSEMM model considers the EU25 MSs, while the former EFSA model considers the MSs in the EU17 group;
- The lower limit of the CI of the C-TSEMM model result is 5.2;
- The C-TSEMM model considers all cattle BSE strains (i.e. including Atypical BSE) while the former EFSA model considers only Classical BSE. Considering Atypical BSE would fit better under the "worst case" scenario of the former EFSA model, which may be consistent with a constant trend of BSE. The result based on the "worst case" scenario of the former EFSA model was that less than seven cases would be missed.

Comparison between these two models should be made with caution due to the differences in their methodology and in the scope of their use. Still, the estimates of the C-TSEMM model regarding number of detectable cases missed if testing of healthy slaughtered cattle were to be stopped in 2011 are in line with the estimates presented in the EFSA Opinion from 2010, taking account of the different scenario's and datasets used.

[24] The "more realistic" scenario of the model in the EFSA 2010 Opinion considers a declining Classical BSE trend based on the historical BSE data available at the time, while a "worst case scenario" was also presented based on a constant prevalence of Classical BSE in birth cohorts since 2004.

5.2.4. Estimated number of years to detect an hypothetical increase in the prevalence of Classical BSE

Table 10 displays the model predictions of the estimated number of years taken to detect a hypothetical annual 10% increase in Classical BSE cases (i.e. consider cases recorded as " Classical" and "unknown" strains in the Commission BSE database) given an emergence initiated in 2011. The assumption is made that detection will occur when the number of model predicted cases exceeds the upper confidence interval prediction of number of cases for 2011 (upper confidence interval prediction is calculated in the baseline model using the upper 95% Poisson confidence interval values for the input test positive data). Results for the number of cases (detectable) and the number of infected animals between 2011 and the year of detection are provided, based on an annual 10% increase in the number of test positive animals.

Table 10: Estimated number of years to detect a hypothetical increase in prevalence of 10% per year with "Classical" and "unknown" strains starting in 2011, together with estimates of the number of detectable cases and infected animals missed during that time interval between the scenario and baseline model.

	Number of years to detect an hypothetical annual 10% increase in prevalence of Classical and "unknown" strains and estimated number of positive cases missed						
MS[1]	Current number test positives (baseline, scenario)	Upper CI limit[+] (0.975)	Years to detection (i.e. cross upper CI limit) (baseline , scenario)	Total test positives at detection (baseline, scenario)	Number of extra cases under scenario before detection (scenario-baseline)	Total infected animals at detection (baseline, scenario)	Number of extra infected animals dead under scenario before detection (scenario-baseline)
EU25	26.85 , 16.98	40.34	6 , 11	207.14 , 314.70	107.56	694.87 , 1,668.93	974.06
AT[*]	0.49 , 0.25	0.73	6 , 13	3.79 , 6.21	2.41	15.47 , 49.15	33.69
BE	0.20 , 0.12	0.62	13 , 19	5.03 , 6.22	1.2	19.85 , 41.41	21.56
CY[*]	0.03 , 0.02	0.04	6 , 11	0.19 , 0.28	0.09	0.79 , 1.90	1.11
CZ	0.85 , 0.33	4.07	18 , 28	38.81 , 44.84	6.04	254.63 , 749.45	494.82
DE	3.64 , 2.11	10.9	13 , 19	89.31 , 108.02	18.72	385.01 , 803.20	418.19
DK	0.03 , 0.02	0.24	25 , 29	2.68 , 2.76	0.07	9.57 , 14.47	4.89
EE[**]	0.09 , 0.04	0.46	19 , 27	4.55 , 5.07	0.52	24.32 , 57.57	33.25
EL[*]	0.08 , 0.06	0.12	5 , 9	0.51 , 0.79	0.28	1.67 , 3.70	2.04
ES	15.37 , 10.57	27.2	7 , 11	145.77 , 195.88	50.11	510.50 , 997.16	486.66
FI[*]	0.22 , 0.15	0.33	6 , 10	1.71 , 2.36	0.65	5.97 , 12.33	6.36
FR	2.64 , 1.99	6.65	11 , 14	48.87 , 55.71	6.84	129.50 , 195.49	65.99
HU[**]	0.29 , 0.12	1.5	19 , 28	14.94 , 16.16	1.22	93.81 , 246.11	152.29
IE	3.54 , 2.81	6.7	8 , 11	40.50 , 52.05	11.55	92.02 , 149.12	57.09
IT	0.32 , 0.07	1.26	16 , 32	11.41 , 13.56	2.15	73.05 , 408.69	335.64
LT[**]	0.22 , 0.02	1.07	18 , 41	10.12 , 11.97	1.85	78.38 , 838.52	760.14

MS[1]	Number of years to detect an hypothetical annual 10% increase in prevalence of Classical and "unknown" strains and estimated number of positive cases missed						
	Current number test positives (baseline, scenario)	Upper CI limit[+] (0.975)	Years to detection (i.e. cross upper CI limit) (baseline , scenario)	Total test positives at detection (baseline, scenario)	Number of extra cases under scenario before detection (scenario-baseline)	Total infected animals at detection (baseline, scenario)	Number of extra infected animals dead under scenario before detection (scenario-baseline)
LU[*]	0.04 , 0.03	0.06	5 , 8	0.25 , 0.37	0.12	0.59 , 1.10	0.51
LV[**]	0.12 , 0.01	0.6	18 , 43	5.70 , 7.12	1.43	43.75 , 568.36	524.62
MT[**]	0.01 , 0.00	0.03	19 , 27	0.27 , 0.29	0.02	1.94 , 4.58	2.65
NL	0.46 , 0.28	2.76	20 , 26	26.31 , 30.18	3.87	106.48 , 202.98	96.5
PL	1.51 , 0.31	10.33	22 , 38	107.91 , 112.65	4.74	727.28 , 3,707.99	2,980.72
PT	2.82 , 2.00	6.38	10 , 14	44.90 , 55.87	10.98	143.20 , 251.36	108.16
SE[*]	0.27 , 0.15	0.4	6 , 12	2.10 , 3.26	1.16	8.80 , 24.38	15.58
SI	0.39 , 0.26	3.59	25 , 29	38.38 , 38.88	0.5	144.29 , 218.06	73.77
SK	0.56 , 0.29	3.82	22 , 28	39.78 , 39.38	-0.4	227.80 , 428.17	200.38
UK	10.61 , 5.00	21.13	9 , 17	144.05 , 202.55	58.5	571.95 , 1,707.70	1,135.75

[1] See Appendix B for MS acronyms
[*]Uses EU17 test positive data. [**]Uses EU8 test positive data, [+]Using model fit on 95[th] Poisson CI input values

From Table 10 it can be seen that across the EU25 (when the EU25 is merged into one epidemiological unit) detection of the emergence would take an estimated 6 years for the baseline monitoring system and 11 years for the scenario monitoring regime. In this intervening five years an additional estimated 108 test positives would be required for the number of cases to be greater than the threshold, and an estimated extra 974 infected animals would be slaughtered/die.

It has been noted that the countries that use the EU17 test positive data, as a proxy, in the absence of cases between 2002-2011, have a fairly short time to detection (i.e. until the model predicted number of cases is greater than the upper threshold value). This early detection is based on the relatively low level of uncertainty associated with the EU17 data and thus the upper CI limit is relatively close to the current number tested. The real level of uncertainty in the individual MSs is higher, due to a smaller sample size. Therefore the model underestimates the time to detection for these countries. A similar scenario exists for the countries using the EU8 test positive data, but to a lesser degree as it is a smaller sample size than the EU17. Therefore, the results for countries denoted with asterisk(s) in Table 10 could be considered to represent the combined MSs of the EU8 and EU17 rather than individual country time to detection.

For the remaining MSs, it can be seen that Spain has the shortest estimated time to detection of 7 years under the baseline monitoring regime, while both Spain and Ireland have the joint shortest estimated time to detection of 11 years under the scenario monitoring regime. The monitoring system of France and Ireland

are estimated to be least affected by the lifting of testing from healthy slaughtered animals, with only three additional years to detect the significant increase. Italy and Poland's monitoring regimes are estimated to be the most affected, with a difference of 16 years between the baseline and monitoring regimes.

The Upper CI limit for the current testing year (2011) in each MS was selected as a means to determine when a MS will 'detect' that there has been a significant increase in the annual number of cases. This approach may not be a realistic method that would be implemented to detect an emergence within the EU25, however, it provides a simple, comparative measure that can be generically applied across all MSs, strains and monitoring regimes without additional assumptions. The upper CI limit for the Slovakia (SK) in 2011 was 3.82, indicating that a significant increase in the number of cases would be detected when a year with greater than 3.82 cases occurs. For SK, the estimated number of cases, when considering a 10% from the current testing year, was greater than 3.79 at 22 years for the baseline regime, and 28 years after under the scenario regime. Over these time periods (22 and 28 years) the model estimates a total of approximately 39.78 observed cases under the baseline regime and 39.38 cases under the scenario regime. The negative results for SK for the number of additional cases are not intuitive, in that there is an estimated additional six years for detection for the scenario regime but this accounts for less cases in total. This is due to the lack of testing of HS animals in the scenario regime, for most other MS's estimates the scenario regime takes sufficiently longer to detect the increase that more cases are detected overall. It is useful to compare the difference in the number of cases with the difference in the number of infected animals dead/slaughtered between monitoring regimes. For SK it can be seen from the table that although the scenario regime detects the theoretical increase with less observed cases, due to the additional year of testing required, an additional 200 infected animals would be slaughtered/die when comparing the regimes.

Whilst Table 10 provides the time to detection for the baseline monitoring system and the scenario of no testing of healthy slaughtered animals, Table 11 below provides the results for the scenario where testing could be reduced by certain MSs to achieve a 1 in 100 000 design prevalence. For those countries shaded in Table 8, the testing of healthy slaughtered animals could be reduced to achieve a design prevalence of 1 in 100 000 at the 95% confidence value, using the detectable prevalence in the standing population. Given the estimated number of healthy slaughter animals to test, Table 11 shows the results for the number of years to detect for those countries with reduced testing. The first column on the left hand side of Table 8 refers to the percentage reduction calculated from the number to test for that MS divided by the total number of healthy slaughter animals tested. For example, the results from Table 8 suggest that Austria is required to test 63 640 HS animals to achieve a design prevalence of 1 in 100 000. Austria currently tests 104 147 HS animals, which suggests that they only need test 61% of their HS animals (63 640/104 147*100).

Table 11: Estimated number of years to detect a hypothetical increase in prevalence of 10% per year with "Classical" and "unknown" strains starting in 2011, together with estimates of the number of detectable cases and infected animals missed during that time interval between the scenario and baseline model, where the scenario is testing the proportion of HS slaughter suggested by the results in Table 8 for the standing population with τ=0.95.

	Number of years to detect an annual 10% increase in prevalence of Classical and "unknown" strains and estimated number of positives missed							
MS[1]	Proportion HS >72 months tested	Current number test positives (baseline, scenario)	Upper CI limit[+] (0.975)	Years to detection (i.e. cross upper CI limit) (baseline , scenario)	Total test positives at detection (baseline, scenario)	Number of extra cases under scenario before detection (scenario-baseline)	Total infected animals at detection (baseline, scenario)	Number of extra infected animals dead under scenario before detection (scenario-baseline)
EU25	0	26.85 , 16.98	40.34	6 , 11	207.14 , 314.70	107.56	694.87 , 1,668.93	974.06
AT[*]	0.61	0.49 , 0.40	0.73	6 , 8	3.79 , 4.56	0.76	15.47 , 22.92	7.46
BE	0	0.20 , 0.12	0.62	13 , 19	5.03 , 6.22	1.2	19.85 , 41.41	21.56
DE	0	3.64 , 2.11	10.9	13 , 19	89.31 , 108.02	18.72	385.01 , 803.20	418.19
DK	0	0.03 , 0.02	0.24	25 , 29	2.68 , 2.76	0.07	9.57 , 14.47	4.89
ES	0	15.37 , 10.57	27.2	7 , 11	145.77 , 195.88	50.11	510.50 , 997.16	486.66
FR	0	2.64 , 1.99	6.65	11 , 14	48.87 , 55.71	6.84	129.50 , 195.49	65.99
IE	0	3.54 , 2.81	6.7	8 , 11	40.50 , 52.05	11.55	92.02 , 149.12	57.09
IT	0.18	0.32 , 0.11	1.26	16 , 27	11.41 , 13.51	2.1	73.05 , 246.06	173.01
NL	0	0.46 , 0.28	2.76	20 , 26	26.31 , 30.18	3.87	106.48 , 202.98	96.5
PL	0.43	1.51 , 0.82	10.33	22 , 28	107.91 , 110.35	2.44	727.28 , 1,367.01	639.73
SE[*]	0.99	0.27 , 0.27	0.4	6 , 6	2.10 , 2.09	-0.01	8.80 , 8.80	0
UK	0	10.61 , 5.00	21.13	9 , 17	144.05 , 202.55	58.5	571.95 , 1,707.70	1,135.75

[1] See Appendix B for MS acronyms

[*]Uses EU17 test positive data. [+]Using model fit on 95th Poisson CI input values

From Table 11 it can be seen that for those MS where no healthy slaughter animals are required to be tested to achieve a 1 in 100 000 design prevalence in the standing population (i.e. those MSs with a 0 in the first column: EU25, BE, DE, DK, ES, FR, IE, NL and UK) the number of years to detect an increase in prevalence is the same between Table 10 and Table 11 where the scenario is no healthy slaughter testing. For those MSs where partial testing achieves the level of confidence required, results are between the baseline (100% testing of healthy slaughter > 72 months) and the scenario of no healthy slaughter testing results given in Table 10. For example, for Austria, under the scenario of no healthy slaughter testing, the number of years to cross the upper confidence interval is achieved at 13 years (Table 10), whereas with the random sampling of 61% of healthy slaughtered animals > 72 months, thus achieving an estimated 1 in 100 000 design prevalence, detection is achieved at 8 years (Table 11).

5.2.5. Estimated number of Atypical BSE cases missed should healthy slaughtered cattle testing stop

France was selected as an individual MS case study as the country has the highest number of L and H type strain typed within EU25 MS datasets.

Table 12 displays the comparison of the estimated number of Atypical cases detected by the baseline and scenario regimes and, as a comparison, the estimated number of infected animals slaughtered/dead for the case study France. The baseline monitoring regime is the testing of healthy slaughter animals > 72 months, emergency slaughter and fallen stock > 48 months and the testing of all clinical suspect animals. The scenario regime only affects the healthy slaughter stream in that no healthy slaughtered animals are tested. The mean values are presented, together with the 95% confidence intervals in brackets

Table 12: Estimated mean number of cases missed and infected animals missed comparing separate calculation of Atypical L and H type with all strains given a change in the monitoring regime from the baseline to a scenario where no healthy slaughter animals are tested with 95% confidence intervals (CI*).

MS	Actual cases 2011 {HS,ES,FS,CS}	Number of Atypical BSE cases missed between monitoring baseline and scenario			
		Baseline detected [CI*]	Scenario detected[CI*]	Number detected missed (baseline - scenario)	Number infected animals dead [CI*] (all streams, all age groups)
FR*-L&H strains	0	1.3 [0.84 , 102.20]	0.94 [0.8 , 101.55]	0.32 [0.045 , 0.64]	3.4 [1.2 , 106.44]
FR-All strains and "unknown"	3 { 0, 0, 3, 0 }	2.9 [1.1 , 105.30]	2.2 [0.98 , 103.98]	0.69 [0.14 , 1.32]	8.2 [2.6 , 114.81]

*FR=France

From Table 12 it can be seen that, for France, the estimated number of cases missed between the baseline and scenario monitoring regimes is approximately the same for both strain combinations, that is approximately 24%-25% of the cases currently detected would not be detected under a regime of no healthy slaughter testing.

When analysing results using only the Atypical data, it is assumed that all cases of BSE are typed by strain such that the number tested for Classical and unknown strains is the same number as that tested for Atypical H and L type. This is not the case for all MSs, and therefore only simulations from MSs where strain differentiation is routinely conducted will be valid.

5.2.6. Estimated number of years to detect an hypothetical increase in the prevalence of Atypical BSE

As per results presented in section 5.2.5, France was selected as an individual MS case study as the country has the highest number of L and H type strain typed within EU25 MS datasets.

Table 13 displays a comparison between the estimated number of years taken to detect a hypothetical 10% increase in French cases between Classical and unknown data and Atypical L and H type strain types. France was selected as the case study as the country has the highest number of L and H type strain typed within EU25 MS datasets. Results for the number of cases (detectable) and the number of infected animals between 2011 and the year of detection are provided, based on an annual 10% increase in the number of test positive animals. Note, when analysing results using only the Atypical data only simulations from MSs where strain differentiation is routinely conducted will be valid.

Table 13: Estimated number of years to detect a hypothetical increase in prevalence of 10% per year in France for different strains starting in 2011, together with estimates of the number of detectable cases and infected animals missed during that time interval for the baseline model

	Number of years to detect an hypothetical annual 10% increase in prevalence of Atypical BSE and estimated number of positive cases missed						
MS	Current number test positives (baseline, scenario)	Upper CI limit (0.975)	Years to detection (i.e. cross upper CI limit) (baseline , scenario)	Total test positives at detection (baseline, scenario)	Number of extra cases under scenario before detection (scenario-baseline)	Total infected animals at detection (baseline, scenario)	Number of extra infected animals dead under scenario before detection (scenario – baseline)
FR*-L&H strains	1.26 , 0.94	10.95	24 , 27	111.70 , 114.20	2.49	304.88 , 417.20	112.32
FR-Classical and "unknown"	2.64 , 1.99	6.65	11 , 14	48.87 , 55.71	6.84	129.50 , 195.49	65.99

*FR=France

From Table 13 it can be seen that, for France (FR), the upper CI limit in 2011 was an estimated 11 for Atypical strains and 6.7 for Classical and unknown strains, indicating that a significant increase in the number of cases would be detected when a year with greater than 11 or 6.7 cases occurs. For FR, the estimated number of Atypical cases was greater than 11 after 24 years for the baseline regime, and 27 years under the scenario regime. Over these time periods (24 and 27 years) the model estimates a total of approximately 112 observed cases under the baseline regime and 114 cases under the scenario regime. For Classical and unknown strains the results are similar, with the estimated number of cases greater that the upper threshold at 11 years for the baseline and 14 years for the scenario monitoring regime. Between these years an estimated 49 cases and 56 cases were observed. The results for France can be compared to those provided in Table 6 for all strains, where the estimated years to cross the upper CI limit were 11 and 14 years for the baseline

5.2.7. Concluding Remarks

The cattle TSE Monitoring Model (C-TSEMM) has a series of assumptions that have to be considered when interpreting the estimates provided by this. Among those assumptions, a key one is that for MSs with no, or few, BSE cases post-2001 an alternative estimate of cohort-based prevalence is required. This has been estimated for those MSs based on the average prevalence of the group of MSs with BSE cases under which they were placed in previous EFSA Opinions (EU17 or theEU8 group)[25]. This results in an overestimate of prevalence for countries with no recorded cases as they are assumed to be a merged epidemiological unit with countries where cases are observed.

Based on the estimates provided by the C-TSEMM model that considered **prevalence in the standing adult cattle population (i.e. Period prevalence in a given year of detectable infected animals in the standing population) and the available historical EU wide data on BSE monitoring**, the following provides support to the reply of the quantitative aspects of the request received from the European Commission:

- In the EU25 as a whole:
    - The current BSE monitoring regime enables the detection of one BSE case in 6 354 930 adult cattle (i.e. older than 24 months of age) with a confidence level of 95%.
    - If the current BSE monitoring regime would exclude testing of healthy slaughter cattle, it would be able to detect in the standing population one BSE case in 4 021 940 adult cattle with a confidence level of 95%. Therefore, no healthy slaughter animals need to be tested in order to meet a design prevalence of one detectable case in 100 000 adult cattle, since testing of at risk animals (i.e. animals showing clinical signs during *ante mortem* inspection, emergency slaughter animals and fallen stock over 48 months of age, and clinical suspects) is sufficient to meet the proposed design prevalence.
- At individual MS level:
    - In eight MSs (Belgium, Germany, Denmark, Spain, France, Ireland, Netherlands, and the UK) the testing of any healthy slaughter animals is not needed in order to meet a 1 in 100 000 design prevalence with a confidence level of 95%, since testing of at risk animals is sufficient to meet the proposed design prevalence.
    - In four MSs (Austria, Italy, Poland and Sweden) the testing of a fraction of healthy slaughtered animals older than 72 months of age (i.e. on the basis of the number tested in 2011) would be sufficient to meet a 1 in 100 000 design prevalence with a confidence level of 95%.
    - In thirteen MSs (Cyprus, Czech Republic, Estonia, Finland, Greece, Hungary, Latvia, Lithuania, Luxembourg, Malta, Portugal, Slovenia and Slovakia) the number of tested animals in 2011 (i.e. including all the healthy slaughtered animals older than 72 months of age) did not allow to meet a 1 in 100 000 design prevalence with 95% confidence. However, fitting a sample size larger than the actually slaughtered cattle population of a MS is neither feasible nor realistic. Thus, the current testing of all animals of certain age categories that are slaughtered or dead may provide the most sensitive BSE monitoring system possible (i.e. that employs *post mortem* tests) under the current epidemiological scenario with the potential limitation on the impact of the

[25] EU17: Austria, Belgium, Cyprus, Denmark, Finland, France, Germany, Greece, Ireland, Italy, Luxembourg, Netherlands, Portugal, Slovenia, Spain, Sweden and United Kingdom;
EU8: Czech Republic, Estonia, Hungary, Latvia, Lithuania, Malta, Poland and Slovakia

age at testing as evaluated in former related EFSA Opinions[26].

- Regarding Classical BSE:
  - Based on a theoretical scenario of an annual 10% increase in detectable cases in the tested population (prevalence), the C-TSEMM model estimates that:
    - In the EU25 as a whole, where testing healthy slaughtered cattle above and age of 72 months is not needed in order to meet the proposed design prevalence, the time to detection of the supposed 10% yearly increase in detectable cases would increase from six to 11 years (i.e. five extra years to detect the supposed 10% yearly increase in prevalence of detectable cases) should testing of healthy slaughtered cattle be stopped compared to the current testing regime.
    - In those MSs where testing healthy slaughtered cattle above the age of 72 months is not needed in order to meet the proposed design prevalence (Belgium, Denmark, France, Germany, Ireland, Netherlands, Spain and the UK), it would take between three and eight extra years (depending on the MS) to detect that yearly increase in prevalence should testing of healthy slaughtered cattle be stopped compared to the current testing regime.
    - In those MSs where testing healthy slaughtered cattle could be reduced in order to meet the proposed design prevalence (Austria, Italy, Poland and Sweden), it would take between six and 16 extra years (depending on the MS) to detect that yearly increase in prevalence should testing of healthy slaughtered cattle older than 72 months of age be reduced to the number needed to meet the proposed design prevalence compared to the current testing regime.
    - In those MSs where testing healthy slaughtered cattle older than 72 months of age as per the current BSE monitoring regime is not sufficient to meet the proposed design prevalence (Cyprus, Czech Republic, Estonia, Finland, Greece, Hungary, Latvia, Lithuania, Luxembourg, Malta, Portugal, Slovenia and Slovakia), it would take between three and 25 extra years (depending on the MS) to detect that yearly increase in prevalence should testing of healthy slaughtered cattle be stopped compared to the current testing regime.
- Regarding Atypical BSE:
  - Based on a theoretical scenario of an 10% annual increase in the number of Atypical BSE infected and detectable cattle in the tested population, the C-TSEMM model estimates that:
    - In the EU25 as a whole, there is not sufficient data (i.e. number of

[26] EFSA (European Food Safety Authority), 2008a. Risk for Human and Animal Health related to the revision of the BSE Monitoring regime in some Member States. The EFSA Journal. 762, 1 - 47.
EFSA, 2008b. Further considerations 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. The EFSA Journal, 763, 1-8.
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detected cases annually) to reliably estimate the impact of the stopping/continuation of testing healthy slaughtered animals older than 72 months.

- However, using France as an example (i.e. country with a large population and sufficient number of detected Atypical cases) the C-TSEMM model indicates that, based on a theoretical scenario of an annual 10% increase of detectable prevalence of Atypical BSE in the tested population, it would take an extra 13 years to detect that yearly increase in prevalence should testing of healthy slaughtered cattle be stopped compared to the current testing regime.

When interpreting the estimates presented above or those obtained in future simulations performed with the C-TSEMM model, consideration has to be given to the assumptions, limitations and uncertainty in the model. Moreover, the models estimates presented in this report are based on the demographics of the adult cattle population in 2011 and on the number of adult cattle removed from the population via the different streams (i.e. healthy slaughter, animals showing clinical signs of disease during *ante mortem* inspection, emergency slaughtered animals and fallen stock). Therefore, future fluctuations in those numbers at EU level and in each of the MSs will impact the validity of the current estimates.

Thus, should the C-TSEMM model be employed in future years for the review of the BSE monitoring regime in the EU, updated yearly data including BSE testing data have to be considered as these drive the results estimated by the model.

5.3. Estimating capacity of the BSE surveillance system to detect the emergence of an hypothetical new type of cattle TSE

The model developed to prepare this Scientific Report allows for future simulations via the modification of key parameters reflecting the epidemiological behaviour of a TSE in cattle (i.e. the age at onset, the age distribution and the sensitivity of current TSE rapid tests). It could therefore serve to estimate through simulation the performance of a defined surveillance system for detecting the emergence of a hypothetical new type of cattle TSE.

When elaborating on a possible new form of TSE in cattle, very different situations can be considered. Whereas certain situations would be purely hypothetical, other could correspond to features of already known TSE agents (e.g. chronic wasting disease (CWD), transmissible mink encephalopathy (TME), Classical and Atypical scrapie in small ruminants, Atypical BSE in cattle).

Amongst the possible scenario to be explored, the following could be included:

- A moderate to high contagiousness of the disease (e.g. similar to CWD, Classical scrapie), with efficient horizontal transmission during the asymptomatic incubation period.

- A limited capacity of the test to detect infected and incubating individuals (e.g. similar to Atypical Scrapie).

- A potentially long incubation asymptomatic period (e.g. similar Atypical BSE, Atypical scrapie) by comparison to life expectations of farmed animals.

Considering the timeframe available for this mandate, carrying out simulation studies was not possible. However, the C-TSEMM model developed in parallel to this Scientific Report (Adkin et al., 2012) can be considered as a useful tool in order to simulate *ad hoc* epidemiological scenarios of hypothetical new types of cattle TSEs.

6. The impact of a new TSE testing policy on TSE monitoring in healthy slaughtered cattle with unchanged testing in at risk cattle

As presented in two previously related EFSA Opinions concerning the first and the second update of the BSE monitoring regime (EFSA, 2008a, 2010), currently EU BSE surveillance aims at detecting:

- Any changes in the trend of the BSE epidemiology, like a decrease or an increase in the number of BSE cases per period in a given region, or in a specific cattle subpopulation (young animals, old animals).
- A hypothetical new emerging TSE in cattle, such as Atypical BSE.

Furthermore, it has to be noted that the main general conclusions of the first Opinion on the revision of the BSE monitoring regime in some MSs (EFSA, 2008a) do remain valid in the context of this Report:

- The purpose of the TSE surveillance in cattle in the EU is mainly to monitor the BSE epidemic.
- Prevention of human exposure to BSE Agent mainly relies on SRM removal.
- Prevention of animal exposure to and propagation of TSE Agents mainly relies on the Feed Ban.

The objective of the former EFSA Opinions was to assess the human and animal health consequences of modifications to the TSE monitoring system in cattle, including options in which TSE testing would be stopped in healthy slaughtered and /or at risk animals born after certain dates. Such scenarios would influence the capacity of the EU TSE monitoring system to fulfil these objectives.

It has to be noted that in the event of a re-emergence of Classical BSE, stopping the testing of healthy slaughtered cattle would lower the sensitivity of its detection by the TSE monitoring system.

The possible future relaxation of certain TSE control measures in cattle and the lack of knowledge related to Atypical BSE strongly plead in favour of a continued, although adapted TSE monitoring system in cattle, which is in balance with the relaxation of the control measures.

Other relevant conclusions of those former EFSA Opinions remain also valid, in particular it can be noted that:

- passive surveillance on its own cannot be considered an appropriate approach to TSE surveillance, since it leads to late detection, when clinical symptoms are not well known and is has a very low sensitivity;
- targeted testing of the at-risk population could represent an efficient early detection tool for the re-emergence of BSE and/or of a new TSE epidemics if it should occur in cattle in the future.

Assuming a new TSE monitoring system in cattle might be designed, aiming at detection of at least one TSE case per 100 000 at a confidence level of 95% over a period of 1 year in the adult cattle population, this could for example consist of the BSE testing of 100% at-risk cattle over 48 months and a minimum sample size in healthy slaughtered cattle above 72 months. In such a situation one may also consider the following:

- the (historic) scientific data and the uncertainties related to TSE in cattle, and
- the need to ensure a high level of protection towards TSE risks.

Apart from the sample size, another important point to consider in designing a sampling strategy for the monitoring and surveillance of TSE in healthy slaughtered cattle is the sampling method, in order to ensure the randomness of the sample (probability sampling) in the targeted population.

If monitoring of BSE in healthy slaughtered cattle remains based on a sample size of animals over certain age, then a sampling strategy should be designed in order to ensure randomness in the targeted population: in this way the sample drawn will better reflect the characteristics of the target population (e.g. in terms of age-, breed-, geographical, feeding regime-, time period-distribution). Stratification procedures may help in sampling each subpopulation (strata, in particular when they are thin) independently and therefore improving the representatives of the final sample.

In the frame of the active surveillance of BSE, the study population is mainly composed of healthy slaughter cattle and fallen stock; in this particular situation it is not possible to apply a simple random sampling which would require the selection of the sample units by drawing up them from a list, i.e. the sampling frame, that is not available by definition. The randomness may be obtained based on the animal ranking in the slaughtering chain through a systematic sampling where the first animal is selected randomly followed by selection at equal intervals (Duncan and Glen, 2006). This sampling strategy therefore does not require knowledge of total size of the study population.

If probability sampling is not ensured, the presence of sampling biases might modify the estimated prevalence of TSEs in the population, in any direction depending on the type of bias, and affect the surveillance ability to monitor the TSE trend.

Concluding remarks on the impact of the TSE testing policy on TSE monitoring in cattle

- BSE Passive Surveillance has been demonstrated to be a very insensitive detection system, when BSE is not expected or easily recognised.
- In contrast active surveillance has been demonstrated to be a far more effective and more sensitive method for BSE monitoring.
- In the event of a re-emergence of Classical BSE, stopping the testing of healthy slaughtered cattle would lower the sensitivity of its detection by the TSE monitoring system.
- If monitoring of BSE in healthy slaughtered cattle remains based on a sample of animals over certain age, then a sampling strategy should be designed in order to ensure randomness in the targeted population: in this way the sample drawn will better reflect the characteristics of the target population (e.g. in terms of age-, breed-, geographical, feeding regime-, time period-distribution). Stratification procedures may help in sampling each subpopulation (i.e. strata, in particular when they are thin) independently and therefore improving the representatives of the final sample.
- It is recommended to assess the sensitivity of the EU surveillance system for detecting the prevalence and trend of Atypical BSE, re-emergence of Classical BSE and the emergence of an hypothetical new type of TSE in cattle should changes be made to current EU BSE control measures and in particular to the total feed ban. This assessment should take into account that, at least for feed-borne transmitted TSEs like Classical BSE, the impact of potential changes to the current total feed ban, it will most likely take five or more years to become apparent due to the known incubation period for Classical BSE, whereas control measures related to the feed ban are likely not to have any influence on non-feed related TSE cases.

CONCLUSIONS AND RECOMMENDATIONS

CONCLUSIONS

General Conclusions

- A constant decline in the total number of detected BSE cases (i.e. coming from both Active and Passive surveillance) has been recorded in the EU 17 group of MSs (Austria, Belgium, Cyprus, Denmark, Finland, France, Germany, Greece, Ireland, Italy, Luxembourg, Netherlands, Portugal, Slovenia, Spain, Sweden and United Kingdom) from 2 157 cases in 2001 to 27 cases in 2011. BSE has not been detected in 5 MSs of the EU8 group of MSs (the EU5 group: Estonia, Hungary, Latvia, Lithuania and Malta). In 3 MSs of the EU8 group (the EU3 group: Czech Republic, Poland and Slovakia), the number of detected cases dropped down from 28 in 2005 (peak) to one in 2011.

- The log10 transformed annual BSE prevalence and incidence (defined respectively as the number of positive BSE cases out of the tested population and out of the standing adult cattle population) in the EU17 and in the EU8 show a statistically significant decreasing trend. There has been a statistically significant increasing trend in the average age of the detected BSE cases per test year during the last 11 years and eight years in the EU17 and the EU8, respectively. At present, this average age exceeds 11 years in each of these MSs (where reported in 2011).

- Assuming that the age distribution of cattle within the EU25 has not changed substantially, the decreasing trend observed in the annual BSE occurrence and the increasing trend observed in the annual average age of the cases are the consequence of the implementation of the BSE control measures.

- Epidemiological data reported by the EU MSs indicate that over the last years the number of detected Atypical BSE cases did not show any trend and that Atypical BSE cases were mainly identified in the fallen stock and healthy slaughtered animals older than eight years of age.

- The performance of the current BSE monitoring system, both in terms of its analytical sensitivity and earliness of the detection of animals infected with Atypical BSE is unknown.

Reply to the Terms of Reference

In order to support the quantitative reply to the terms of reference, a model called Cattle TSE Monitoring Model (C-TSEMM) was developed by an EFSA contractor in order to provide a general frame for evaluating the design prevalence and the sensitivity of cattle TSE monitoring systems[27].

This model, so called Cattle TSE Monitoring Model (C-TSEMM), has a series of assumptions that have to be considered when interpreting the estimates provided by this. Among those assumptions, a key one is that for MSs with no, or few, BSE cases post-2001 an alternative estimate of cohort-based prevalence is required. This has been estimated for those MSs based on the average prevalence of the group of MSs with BSE cases under which they were placed in previous EFSA Opinions[28]: the EU17

[27] Amie Adkin, Robin Simmons and Mark Arnold; Model for evaluation of different options for the monitoring of Transmissible Spongiform Encephalopathies in cattle in the European Union (C-TSEMM). Supporting Publications 2012:EN-349. [55 pp.]. Available online: www.efsa.euopa.eu/publications

[28] EFSA, 2010. Opinion of the Scientific Panel on Biological Hazards on a second update on the risk for human and animal health related to the revision of the BSE monitoring regime in some Member States. The EFSA Journal, 8(12), 1946.

or the EU8 group[29]. This results in an overestimate of prevalence for countries with no recorded cases as they are assumed to be a merged epidemiological unit with MSs where cases are observed.

Based on the estimates provided by the C-TSEMM model **that considered prevalence in the standing adult cattle population (i.e. period prevalence in a given year of detectable infected animals in the standing population) and the available historical EU wide data on BSE monitoring**, the following provides support to the reply of the quantitative aspects of the request received from the European Commission:

- With regards to the request for a proposal on a **minimum annual sample size in healthy slaughtered cattle above 72 months of age in order to allow the detection of BSE with a yearly design prevalence of at least 1 case per 100 000 in the adult population** (i.e. older than 24 months of age) of the Member States, at a confidence level of 95% in the group of 25 EU Member States as a whole and in each Member State individually.
    - In the EU25 as a whole and according to C-TSEMM model estimates:
        - The current BSE monitoring regime enables the detection of one BSE case in 6 354 930 adult cattle with a confidence level of 95%.
        - If the current BSE monitoring regime would exclude testing of healthy slaughter cattle, it would be able to detect in the standing population one BSE case in 4 021 940 adult cattle with a confidence level of 95%. Therefore, no healthy slaughter animals need to be tested in order to meet a design prevalence of 1 detectable case in 100 000 adult cattle, since testing of at risk animals (i.e. animals showing clinical signs during *ante mortem* inspection, emergency slaughter and fallen stock over 48 months of age, and clinical suspects) is sufficient to meet the proposed design prevalence.
    - At individual MS level, according to C-TSEMM model estimates:
        - In eight MSs (Belgium, Denmark, France, Germany, Ireland, Netherlands, Spain and the UK) the testing of healthy slaughter animals is not needed in order to meet a 1 in 100 000 design prevalence with a confidence level of 95%, since testing of at risk animals is sufficient to meet the proposed design prevalence.
        - In four MSs (Austria, Italy, Poland and Sweden) the testing of a fraction of healthy slaughtered animals older than 72 months of age (i.e. on the basis of the number tested in 2011) would be sufficient to meet a 1 in 100 000 design prevalence with a confidence level of 95%.
        - In thirteen MSs (Cyprus, Czech Republic, Estonia, Finland, Greece, Hungary, Latvia, Lithuania, Luxembourg, Malta, Portugal, Slovenia and Slovakia) the number of tested animals in 2011 (i.e. including all the healthy slaughtered animals older than 72 months of age) did not allow to meet a 1 in 100 000 design prevalence with 95% confidence. However, fitting a sample size larger than the actually slaughtered cattle population of a MS is neither feasible nor realistic. Thus, the current testing of all animals of certain age categories that are slaughtered or dead may provide the most sensitive BSE monitoring system possible (i.e. that employs *post mortem* tests) under the current epidemiological scenario with the potential limitation on the impact of the age

[29] EU17: Austria, Belgium, Cyprus, Denmark, Finland, France, Germany, Greece, Ireland, Italy, Luxembourg, Netherlands, Portugal, Slovenia, Spain, Sweden and United Kingdom; EU8: Czech Republic, Estonia, Hungary, Latvia, Lithuania, Malta, Poland and Slovakia

at testing as evaluated in former related EFSA Opinions[30].

- On the **added value** of this minimum sample to the overall surveillance programme in terms of monitoring the trend of Classical BSE, Atypical BSE and the emergence of an hypothetical new type of cattle TSE:
    - Regarding Classical BSE:
        - In the event of a re-emergence of Classical BSE, stopping the testing of healthy slaughtered cattle would lower the sensitivity of its detection by the TSE monitoring system.
        - Based on a theoretical scenario of an annual 10% increase in detectable cases in the tested population (prevalence), the C-TSEMM model estimates that:
            - In the EU25 as a whole, where testing healthy slaughtered cattle above and age of 72 months is not needed in order to meet the proposed design prevalence, the time to detection of the supposed 10% yearly increase in detectable cases would increase from six to 11 years (i.e. five extra years to detect the supposed 10% yearly increase in prevalence of detectable cases) should testing of healthy slaughtered cattle be stopped compared to the current testing regime.
            - In those MSs where testing healthy slaughtered cattle above the age of 72 months is not needed in order to meet the proposed design prevalence (Belgium, Denmark, France, Germany, Ireland, Netherlands, Spain and the UK), it would take between three and eight extra years (depending on the MS) to detect that yearly increase in prevalence should testing of healthy slaughtered cattle be stopped compared to the current testing regime.
            - In those MSs where testing healthy slaughtered cattle could be reduced in order to meet the proposed design prevalence (Austria, Italy, Poland and Sweden), it would take between six and 16 extra years (depending on the MS) to detect that yearly increase in prevalence should testing of healthy slaughtered cattle older than 72 months of age be reduced to the number needed to meet the proposed design prevalence compared to the current testing regime.
            - In those MSs where testing healthy slaughtered cattle older than 72 months of age as per the current BSE monitoring regime is not sufficient to meet the proposed design prevalence (Cyprus, Czech Republic, Estonia, Finland, Greece, Hungary, Latvia, Lithuania, Luxembourg, Malta, Portugal, Slovenia and Slovakia), it would take between three and 25 extra years (depending on the MS) to detect that yearly increase in prevalence should testing of healthy slaughtered cattle be stopped compared to the current testing regime.

[30] EFSA (European Food Safety Authority), 2008a. Risk for Human and Animal Health related to the revision of the BSE Monitoring regime in some Member States. The EFSA Journal. 762, 1 - 47.
EFSA, 2008b. Further considerations 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. The EFSA Journal, 763, 1-8.
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EFSA, 2010. Opinion of the Scientific Panel on Biological Hazards on a second update on the risk for human and animal health related to the revision of the BSE monitoring regime in some Member States. The EFSA Journal, 8(12), 1946.

- Regarding Atypical BSE:
  - Based on a theoretical scenario of an 10% annual increase in the number of Atypical BSE infected and detectable cattle in the tested population, the C-TSEMM model estimates that:
    - At EU25 as a whole, there is not sufficient data (i.e. number of detected cases annually) to reliably estimate the impact of the stopping/continuation of testing healthy slaughtered animals older than 72 months.
    - However, using France as an example (i.e. country with a large population and sufficient number of detected Atypical cases) the C-TSEMM model indicates that, based on a theoretical scenario of an annual 10% increase of detectable prevalence of Atypical BSE in the tested population, it would take an extra 13 years to detect that yearly increase in prevalence should testing of healthy slaughtered cattle be stopped compared to the current testing regime.
- Regarding an hypothetical new type of cattle TSE:
    - Considering the timeframe available for this mandate, carrying out simulation studies for hypothetical new types of cattle TSEs was not possible. However, the C-TSEMM model developed in parallel to this Scientific Report can be considered as a useful tool in order to simulate future *ad hoc* epidemiological scenarios of hypothetical new types of cattle TSEs.

When interpreting the estimates presented above or those obtained in future simulations performed with the C-TSEMM model, consideration has to be given to the assumptions, limitations and uncertainty in the model. Moreover, the models estimates presented in this report are based on the demographics of the adult cattle population in 2011 and on the number of adult cattle removed from the population via the different streams (i.e. healthy slaughter, animals showing clinical signs of disease during *ante mortem* inspection, emergency slaughtered animals and fallen stock). Therefore, future fluctuations in those numbers at EU level and in each of the MSs will impact the validity of current estimates.

RECOMMENDATIONS

The following is recommended:

- If monitoring of BSE in healthy slaughtered cattle remains based on a sample of animals over certain age, then a sampling strategy should be designed in order to ensure randomness in the targeted population: in this way the sample drawn will better reflect the characteristics of the target population (e.g. in terms of age-, breed-, geographical, feeding regime-, time period-distribution). Stratification procedures may help in sampling each subpopulation (i.e. strata, in particular when they are thin) independently and therefore improving the representatives of the final sample.
- To assess the sensitivity of the EU surveillance system for detecting the prevalence and trend of Atypical BSE, re-emergence of Classical BSE and the emergence of an hypothetical new type of TSE in cattle should changes be made to current EU BSE control measures and in particular to the total feed ban. This assessment should take into account that, at least for feed-borne transmitted TSEs like Classical BSE, the impact of potential changes to the current total feed ban will most likely take five or more years to become apparent due to the known

incubation period for Classical BSE, whereas control measures related to the feed ban are likely not to have any influence on non-feed related TSE cases.

- Should the C-TSEMM model be employed in future years for the review of the BSE monitoring regime in the EU, updated yearly data including BSE testing data have to be considered as these drive the results estimated by the model.

DOCUMENTATION PROVIDED TO EFSA

1. Mandate from the European Commission on a Request for scientific and technical assistance on the minimum sample size to test should an annual BSE statistical testing regime be authorised in healthy slaughtered cattle. Ref. SANCO/G4/FS/rz 2011. Received on 18 November 2011.

2. Data on BSE cases detected in the frame of BSE monitoring in the EU from 2001 to 2011. Provided by the European Commission on 3 July 2012.

3. Data on number of rapid TSE testes performed in the EU in the frame of BSE monitoring from 2001 to 2011. Provided by the European Commission on 11 January 2012 and 3 July 2012.

4. Additional data on details of BSE testing and cattle populations in the EU MSs requested to the EU MSs with the support of the European Commission, between 13 March 2012 and 6 July 2012.

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APPENDICES

A. SUMMARY TABLES ON THE REVIEW OF THE BSE TREND IN THE 25 EU MSS BY GROUP OF MSS

Data excludes cases with unidentified age category. Details are presented in Table 1 below.

Table 14: Reported BSE cases by MS that were excluded from the analysis due to incomplete data on age category or year of testing.

	Year of testing						
MSs	1991	2003	2004	2005	2006	2007	2008
Belgium	1*						
Portugal		1	1				1
United Kingdom			2	3	2	4	

*Reported year of testing 1991 is the reason for exclusion from the analysis.

1. Group of 17 EU MSs (EU17).

Table 15: Prevalence (number of BSE cases per ten thousand of animals tested) of BSE in the EU17 for passive and active surveillance from 2001 to 2011. Prevalence for some years presented in this table may be slightly different to those presented in the EFSA Scientific Opinion of 2010 (EFSA, 2010) due to changes made by MSs to the reported data hosted by the European Commission.

Type of testing	**Year of testing**										
	2001	**2002**	**2003**	**2004**	**2005**	**2006**	**2007**	**2008**	**2009**	**2010**	**2011**
Active Surveillance	1.23	1.38	1.01	0.64	0.49	0.31	0.16	0.12	0.09	0.06	0.05
Passive Surveillance	3086	2535	1165	571	256	161	83	51	29	0	0

Table 16: Number of BSE cases detected through the BSE Surveillance (Active and Passive) in EU17 during the period 2001 – 2011 per birth cohort and year of detection.

Birth cohort	N° of detected BSE cases per year											
	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	Grand Total
1980		1										1
1981	1									1		2
1982												0
1983			1									1
1984	1	3				1						5
1985	1	2	2		1							6
1986	13	10	3	3	1							30
1987	21	30	9	6	6	1		1				74
1988	20	28	21	8		1		2			1	81
1989	25	37	21	17	5	5	1	1	1			113
1990	28	54	22	22	9	7	1	3	1			147
1991	66	78	47	27	22	8	1	1		1	1	252
1992	120	156	84	55	37	15	10	1	2		1	481
1993	328	245	180	95	56	27	17	9	6	4	2	969
1994	577	457	218	123	91	48	25	16	6	3	3	1 567
1995	665	615	300	137	66	37	22	10	11	6	3	1 872
1996	243	269	163	79	37	25	9	23	5	5	2	860
1997	44	90	152	85	34	23	6	13	4	5	4	460
1998	4	29	73	94	40	32	17	7	5	2	4	307
1999		5	24	50	57	36	15	9	5	5	2	208
2000			1	19	49	35	19	8	6	4	3	144
2001					7	8	2	5	1	1		24
2002					2	1	3	3	2			11
2003								3	3			6
2004									1	4		5
2005											1	1
Grand Total	2 157	2 109	1 321	820	520	310	148	115	59	41	27	7 627

Table 17: Number of BSE cases, incidence per million adult cattle (i.e. over 24 months of age) and average age in years of cases during the period 2001 – 2011 per year of detection in the EU17 MS. The data consider both BSE Active and Passive Surveillance.

Member State		Year of testing											Total
		2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	
Austria	N° cases	1	0	0	0	2	2	1	0	0	2	0	8
	Incidence	1.0	0	0	0	2.1	2.1	1.1	0	0	2.1	0	
	Average age	5.0	NA	NA	NA	12.0	9.5	11.0	NA	NA	14.0	NA	10.9
Belgium	N° cases	45	38	15	11	3	1	0	0	0	0	0	113
	Incidence	29.7	26.1	10.6	7.8	2.2	0.7	0	0	0	0	0	
	Average age	6.0	6.7	7.4	7.5	10.0	12.0	NA	NA	NA	NA	NA	6.7
Cyprus	N° cases	0	0	0	0	0	0	0	0	0	0	0	0
	Incidence	0	0	0	0	0	0	0	0	0	0	0	
	Average age	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA
Denmark	N° cases	6	3	2	1	1	0	0	0	1	0	0	14
	Incidence	6.7	3.5	2.4	1.3	1.3	0	0	0	1.3	0	0	
	Average age	5.0	5.3	6.5	14.0	9.0	NA	NA	NA	14.0	NA	NA	6.9
Finland	N° cases	1	0	0	0	0	0	0	0	0	0	0	1
	Incidence	2.4	0	0	0	0	0	0	0	0	0	0	
	Average age	6	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA	6
France	N° cases	274	239	111	51	32	8	7	8	10	5	3	748
	Incidence	24.7	21.8	10.4	4.9	3.1	0.8	0.7	0.8	0.9	0.5	0.3	
	Average age	6.4	7.2	8.1	8.8	9.4	9.3	10.7	12.4	13.6	11.6	14.3	7.5
Germany	N° cases	121	103	53	63	32	16	4	2	2	0	0	396
	Incidence	18.8	16.5	8.6	10.5	5.5	2.8	0.7	0.3	0.3	0	0	
	Average age	5.5	6.4	5.9	6.2	6.2	7.0	7.8	8.0	11.0	NA	NA	6.1
Greece	N° cases	1	0	0	0	0	0	0	0	0	0	0	1
	Incidence	3.0	0	0	0	0	0	0	0	0	0	0	
	Average age	5.0	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA	5.0
Ireland	N° cases	242	329	182	125	73	38	25	20	9	2	3	1 048
	Incidence	79.5	109.4	60.8	41.0	23.8	12.6	8.5	7.3	3.2	0.7	1.1	
	Average age	6.6	7.8	8.7	9.8	10.1	11.1	11.6	11.8	11.2	15.0	16.7	8.4
Italy	N° cases	50	36	31	7	8	7	2	1	2	0	1	145
	Incidence	15.5	11.9	10.4	2.4	2.7	2.5	0.7	0.4	0.7	0	0.4	
	Average age	5.6	6.5	7.8	7.3	8.1	8.3	12.5	13.0	12.0	NA	14.0	6.9
Luxemburg	N° cases	0	1	0	0	1	0	0	0	0	0	0	2
	Incidence	0	10.3	0	0	10.8	0	0	0	0	0	0	
	Average age	NA[2]	6.0	NA	NA	4.0	NA	NA	NA	NA	NA	NA	5.0
Netherlands	N° cases	20	24	18	6	3	2	2	1	0	3	0	79
	Incidence	11.2	13.5	10.1	3.5	1.8	1.2	1.2	0.6	0	1.7	0	
	Average age	6.3	6.2	6.7	8.3	4.7	8.5	7.5	8.0		12.7		6.8
Portugal	N° cases	110	85	132	89	53	32	13	18	6	5	5	548
	Incidence	142.2	109.3	168.8	109.5	64.4	39.2	15.7	21.7	7.1	5.8	5.9	
	Average age	6.7	7.3	7.7	8.6	9.7	10.9	11.4	12.6	13.3	15.4	16.4	8.4
Slovenia	N° cases	1	1	1	2	1	1	1	0	0	0	0	8
	Incidence	4.7	4.6	4.8	9.9	5.1	5.1	4.9	0	0	0	0	
	Average age	5.0	7.0	4.0	5.0	5.0	6.0	7.0					5.5
Spain	N° cases	82	127	166	137	98	76	32	24	18	13	7	780
	Incidence	23.8	35.8	46.3	38.2	28.2	23.8	9.5	7.4	5.5	4.0	2.3	
	Average age	6.4	6.5	6.8	6.8	6.7	7.5	9.1	10.2	12.4	11.8	13.0	7.2
Sweden	N° cases	0	0	0	0	0	1	0	0	0	0	0	1
	Incidence	0	0	0	0	0	1.5	0	0	0	0	0	
	Average age	NA	NA	NA	NA	NA	12.0	NA	NA	NA	NA	NA	12.0
United Kingdom	N° cases	1 203	1 123	610	328	213	126	61	41	11	11	8	3 735
	Incidence	243.7	228.3	124.7	66.7	43.5	25.8	12.8	8.8	2.4	2.4	1.8	
	Average age	7.6	8.9	9.6	10.7	11.4	11.8	11.9	12.6	11.8	14.3	14.8	9.1
EU17	N° cases	2 157	2 109	1 321	820	520	310	148	115	59	41	27	7 627
	Incidence	53.9	53.6	34.0	21.4	13.7	8.3	3.9	3.1	1.6	1.1	0.7	
	Average age	7.0	8.1	8.6	9.1	9.6	10.1	10.9	11.8	12.4	13.2	14.7	8.4

[1]NA=Non Applicable

2. Group of 8 EU MSs (EU8).

Table 18: Prevalence (number of BSE cases per ten thousand of animals tested) of BSE in the EU3 (i.e. where BSE cases have been reported) and in the EU8 for active surveillance from 2004 to 2011. No cases have been identified through passive surveillance in the EU3.

Prevalence	Year of testing							
	2004	2005	2006	2007	2008	2009	2010	2011
EU3	0.33	0.37	0.15	0.16	0.07	0.07	0.04	0.02
EU8	0.26	0.28	0.12	0.12	0.06	0.05	0.03	0.01

Table 19: Number of BSE cases detected through the BSE Surveillance (Active and Passive) in EU3 (i.e. where BSE cases have been reported) during the period 2004 – 2011 per birth cohort and year of detection.

Birth Cohort	No of detected BSE cases per year								
	2004	2005	2006	2007	2008	2009	2010	2011	Total
1992	2	1							3
1993									0
1994	1	1	2						4
1995	3	1	1	3		1	1		10
1996	5	2	3	1				1	12
1997	2	2	1						5
1998	3	2	1	1					7
1999	2	5	2	3	2	2			16
2000	5	11	2	1	1	1	1		22
2001	2	2	1	2	1				8
2002		1					1		2
2003				1	1	1			3
2004				1		1			2
2005					1				1
Grand Total	25	28	13	13	6	6	3	1	95

[1] In 2004, seven cases were diagnosed before 1st May.

Table 20: Number of BSE cases, incidence per million cattle over 24 months and average age in years of cases during the period 2004 – 2011 per year of detection in the EU8 MS (the data consider both BSE Active and Passive Surveillance. Please note no cases identified in Passive Surveillance).

Member State		Year of testing								Total
		2004	2005	2006	2007	2008	2009	2010	2011	
Czech Republic	N° cases	7	7	3	2	0	2	0	0	21
	Incidence	10.7	10.9	4.6	3.1	0	3.1	0	0	
	Average age	5.9	5.1	6.3	10	NA	5.5	NA	NA	6.0
Estonia	N° cases	0	0	0	0	0	0	0	0	0
	Incidence	0	0	0	0	0	0	0	0	
	Average age	NA	NA	NA	NA	NA	NA	NA	NA	NA
Hungary	N° cases	0	0	0	0	0	0	0	0	0
	Incidence	0	0	0	0	0	0	0	0	
	Average age	NA	NA	NA	NA	NA	NA	NA	NA	NA
Latvia	N° cases	0	0	0	0	0	0	0	0	0
	Incidence	0	0	0	0	0	0	0	0	
	Average age	NA	NA	NA	NA	NA	NA	NA	NA	NA
Lithuania	N° cases	0	0	0	0	0	0	0	0	0
	Incidence	0	0	0	0	0	0	0	0	
	Average age	NA	NA	NA	NA	NA	NA	NA	NA	NA
Malta	N° cases	0	0	0	0	0	0	0	0	0
	Incidence	0	0	0	0	0	0	0	0	
	Average age	NA	NA	NA	NA	NA	NA	NA	NA	NA
Poland	N° cases	11	19	10	9	5	4	2	1	61
	Incidence	3.6	6.2	3.3	3.0	1.6	1.3	0.7	0.3	
	Average age	8.3	6.9	9.4	8.2	6.8	10.8	12.5	15.0	8.3
Slovakia	N° cases	7	2	0	2	1	0	1	0	13
	Incidence	25.9	7.4	0	7.9	4.0	0	4.1	0	
	Average age	5.3	5.0	NA	6.0	7.0	NA	8.0	NA	
EU8	N° cases	25	28	13	13	6	6	3	1	95
	Incidence	4.8	5.4	2.6	2.6	1.2	1.2	0.6	0.2	
	Average age	6.8	6.4	8.7	8.2	6.8	9.0	11.0	15.0	7.5

[1] In 2004, seven cases were diagnosed before 1st May.
[2] NA=Non applicable.

B. BSE CASES DETECTED THROUGH BSE MONITORING (ACTIVE AND PASSIVE) BETWEEN 2001/2004 AND 2011 PER MEMBER STATE, BIRTH COHORT AND YEAR OF DETECTION

The following has to be considered when interpreting the data provided in this Appendix:

- Data as provided by the European Commission on 3 July 2012.
- Data excludes cases identified in the frame of BSE eradication measures. Details are presented in Table 1 below.

Table 21: Reported BSE cases identified in the frame of BSE eradication measures in the EU from 2001 to 2011.

	N° of detected BSE cases per year[1]								
MSs	**2001**	**2002**	**2003**	**2004**	**2005**	**2006**	**2007**	**2008**	**Total**
Belgium	1								1
Czech Republic					1				1
France	3	1							4
Germany	4	3	1	2					10
Ireland		5	1	1				2	9
Poland					1				1
Portugal		1		2					3
Slovakia					1				1
Spain	1		1		5		1		8
United Kingdom					8	1		1	10
Total	9	10	3	5	16	1	1	3	48

[1]No cases identified in the frame of BSE eradication measures have been reported after 2008 up to 2011.

- Data excludes cases with unidentified age category. Details are presented in Table 2 below.

Table 22: Reported BSE cases by MS that were excluded from the analysis due to incomplete data on age category or year of testing.

	Year of testing						
MSs	**1991**	**2003**	**2004**	**2005**	**2006**	**2007**	**2008**
Belgium	1*						
Portugal		1	1				1
United Kingdom			2	3	2	4	

*Reported year of testing 1991 is the reason for exclusion from the analysis.

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Austria (AT)

Number of BSE cases detected through the BSE Surveillance (Active and Passive) in Austria since 2001 per birth cohort and year of detection.

	N° of detected BSE cases per year											
Birth cohort	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	Total
1980	-	-	-	-	-	-	-	-	-	-	-	0
1981	-	-	-	-	-	-	-	-	-	-	-	0
1982	-	-	-	-	-	-	-	-	-	-	-	0
1983	-	-	-	-	-	-	-	-	-	-	-	0
1984	-	-	-	-	-	-	-	-	-	-	-	0
1985	-	-	-	-	-	-	-	-	-	-	-	0
1986	-	-	-	-	-	-	-	-	-	-	-	0
1987	-	-	-	-	-	-	-	-	-	-	-	0
1988	-	-	-	-	-	-	-	-	-	-	-	0
1989	-	-	-	-	-	-	-	-	-	-	-	0
1990	-	-	-	-	-	-	-	-	-	-	-	0
1991	-	-	-	-	-	-	-	-	-	-	-	0
1992	-	-	-	-	1	-	-	-	-	-	-	1
1993	-	-	-	-	-	1	-	-	-	-	-	1
1994	-	-	-	-	1	-	-	-	-	-	-	1
1995	-	-	-	-	-	-	-	-	-	1	-	1
1996	1	-	-	-	-	-	1	-	-	-	-	2
1997	-	-	-	-	-	-	-	-	-	1	-	1
1998	-	-	-	-	-	-	-	-	-	-	-	0
1999	-	-	-	-	-	-	-	-	-	-	-	0
2000	-	-	-	-	-	1	-	-	-	-	-	1
2001	-	-	-	-	-	-	-	-	-	-	-	0
2002	-	-	-	-	-	-	-	-	-	-	-	0
2003	-	-	-	-	-	-	-	-	-	-	-	0
2004	-	-	-	-	-	-	-	-	-	-	-	0
2005	-	-	-	-	-	-	-	-	-	-	-	0
Total	1	0	0	0	2	2	1	0	0	2	0	8

Belgium (BE)

Number of BSE cases detected through the BSE Surveillance (Active and Passive) in Belgium since 2001 per birth cohort and year of detection.

	N° of detected BSE cases per year											
Birth cohort	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	Total
1980	-	-	-	-	-	-	-	-	-	-	-	0
1981	-	-	-	-	-	-	-	-	-	-	-	0
1982	-	-	-	-	-	-	-	-	-	-	-	0
1983	-	-	-	-	-	-	-	-	-	-	-	0
1984	-	-	-	-	-	-	-	-	-	-	-	0
1985	-	-	-	-	-	-	-	-	-	-	-	0
1986	-	-	-	-	-	-	-	-	-	-	-	0
1987	-	-	-	-	-	-	-	-	-	-	-	0
1988	-	-	-	-	-	-	-	-	-	-	-	0
1989	-	-	-	-	-	-	-	-	-	-	-	0
1990	-	-	-	-	-	-	-	-	-	-	-	0
1991	1	-	1	-	1	-	-	-	-	-	-	3
1992	1	3	-	-	-	-	-	-	-	-	-	4
1993	1	1	-	-	-	-	-	-	-	-	-	2
1994	7	5	2	1	-	1	-	-	-	-	-	16
1995	17	8	2	-	-	-	-	-	-	-	-	27
1996	18	13	6	4	-	-	-	-	-	-	-	41
1997	-	8	3	4	2	-	-	-	-	-	-	17
1998	-	-	1	2	-	-	-	-	-	-	-	3
1999	-	-	-	-	-	-	-	-	-	-	-	0
2000	-	-	-	-	-	-	-	-	-	-	-	0
2001	-	-	-	-	-	-	-	-	-	-	-	0
2002	-	-	-	-	-	-	-	-	-	-	-	0
2003	-	-	-	-	-	-	-	-	-	-	-	0
2004	-	-	-	-	-	-	-	-	-	-	-	0
2005	-	-	-	-	-	-	-	-	-	-	-	0
Total	45	38	15	11	3	1	0	0	0	0	0	113

Cyprus (CY)

Number of BSE cases detected through the BSE Surveillance (Active and Passive) in Cyprus since 2001 per birth cohort and year of detection.

	N° of detected BSE cases per year											
Birth cohort	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	Total
1980	-	-	-	-	-	-	-	-	-	-	-	0
1981	-	-	-	-	-	-	-	-	-	-	-	0
1982	-	-	-	-	-	-	-	-	-	-	-	0
1983	-	-	-	-	-	-	-	-	-	-	-	0
1984	-	-	-	-	-	-	-	-	-	-	-	0
1985	-	-	-	-	-	-	-	-	-	-	-	0
1986	-	-	-	-	-	-	-	-	-	-	-	0
1987	-	-	-	-	-	-	-	-	-	-	-	0
1988	-	-	-	-	-	-	-	-	-	-	-	0
1989	-	-	-	-	-	-	-	-	-	-	-	0
1990	-	-	-	-	-	-	-	-	-	-	-	0
1991	-	-	-	-	-	-	-	-	-	-	-	0
1992	-	-	-	-	-	-	-	-	-	-	-	0
1993	-	-	-	-	-	-	-	-	-	-	-	0
1994	-	-	-	-	-	-	-	-	-	-	-	0
1995	-	-	-	-	-	-	-	-	-	-	-	0
1996	-	-	-	-	-	-	-	-	-	-	-	0
1997	-	-	-	-	-	-	-	-	-	-	-	0
1998	-	-	-	-	-	-	-	-	-	-	-	0
1999	-	-	-	-	-	-	-	-	-	-	-	0
2000	-	-	-	-	-	-	-	-	-	-	-	0
2001	-	-	-	-	-	-	-	-	-	-	-	0
2002	-	-	-	-	-	-	-	-	-	-	-	0
2003	-	-	-	-	-	-	-	-	-	-	-	0
2004	-	-	-	-	-	-	-	-	-	-	-	0
2005	-	-	-	-	-	-	-	-	-	-	-	0
Total	0	0	0	0	0	0	0	0	0	0	0	0

Czech Republic (CZ)

Number of BSE cases detected through the BSE Surveillance (Active and Passive) in Czech Republic since 2004 per birth cohort and year of detection.

	N° of detected BSE cases per year								
Birth cohort	2004	2005	2006	2007	2008	2009	2010	2011	Total
1980	-	-	-	-	-	-	-	-	0
1981	-	-	-	-	-	-	-	-	0
1982	-	-	-	-	-	-	-	-	0
1983	-	-	-	-	-	-	-	-	0
1984	-	-	-	-	-	-	-	-	0
1985	-	-	-	-	-	-	-	-	0
1986	-	-	-	-	-	-	-	-	0
1987	-	-	-	-	-	-	-	-	0
1988	-	-	-	-	-	-	-	-	0
1989	-	-	-	-	-	-	-	-	0
1990	-	-	-	-	-	-	-	-	0
1991	-	-	-	-	-	-	-	-	0
1992	-	-	-	-	-	-	-	-	0
1993	-	-	-	-	-	-	-	-	0
1994	-	-	-	-	-	-	-	-	0
1995	-	-	-	-	-	-	-	-	0
1996	1	-	-	1	-	-	-	-	2
1997	2	-	-	-	-	-	-	-	2
1998	1	-	-	1	-	-	-	-	2
1999	1	1	1	-	-	-	-	-	3
2000	2	6	2	-	-	-	-	-	10
2001	-	-	-	-	-	-	-	-	0
2002	-	-	-	-	-	-	-	-	0
2003	-	-	-	-	-	1	-	-	1
2004	-	-	-	-	-	1	-	-	1
2005	-	-	-	-	-	-	-	-	0
Total	7	7	3	2	0	2	0	0	21

Denmark (DK)

Number of BSE cases detected through the BSE Surveillance (Active and Passive) in Denmark since 2001 per birth cohort and year of detection.

	N° of detected BSE cases per year											
Birth cohort	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	Total
1980	-	-	-	-	-	-	-	-	-	-	-	0
1981	-	-	-	-	-	-	-	-	-	-	-	0
1982	-	-	-	-	-	-	-	-	-	-	-	0
1983	-	-	-	-	-	-	-	-	-	-	-	0
1984	-	-	-	-	-	-	-	-	-	-	-	0
1985	-	-	-	-	-	-	-	-	-	-	-	0
1986	-	-	-	-	-	-	-	-	-	-	-	0
1987	-	-	-	-	-	-	-	-	-	-	-	0
1988	-	-	-	-	-	-	-	-	-	-	-	0
1989	-	-	-	-	-	-	-	-	-	-	-	0
1990	-	-	-	1	-	-	-	-	-	-	-	1
1991	-	-	-	-	-	-	-	-	-	-	-	0
1992	-	-	-	-	-	-	-	-	-	-	-	0
1993	1	-	-	-	-	-	-	-	-	-	-	1
1994	-	-	-	-	-	-	-	-	-	-	-	0
1995	-	-	-	-	-	-	-	-	1	-	-	1
1996	3	2	1	-	1	-	-	-	-	-	-	7
1997	1	-	1	-	-	-	-	-	-	-	-	2
1998	1	1	-	-	-	-	-	-	-	-	-	2
1999	-	-	-	-	-	-	-	-	-	-	-	0
2000	-	-	-	-	-	-	-	-	-	-	-	0
2001	-	-	-	-	-	-	-	-	-	-	-	0
2002	-	-	-	-	-	-	-	-	-	-	-	0
2003	-	-	-	-	-	-	-	-	-	-	-	0
2004	-	-	-	-	-	-	-	-	-	-	-	0
2005	-	-	-	-	-	-	-	-	-	-	-	0
Total	6	3	2	1	1	0	0	0	1	0	0	14

Estonia (EE)

Number of BSE cases detected through the BSE Surveillance (Active and Passive) in Estonia since 2004 per birth cohort and year of detection.

	N° of detected BSE cases per year								
Birth cohort	2004	2005	2006	2007	2008	2009	2010	2011	Total
1980	-	-	-	-	-	-	-	-	0
1981	-	-	-	-	-	-	-	-	0
1982	-	-	-	-	-	-	-	-	0
1983	-	-	-	-	-	-	-	-	0
1984	-	-	-	-	-	-	-	-	0
1985	-	-	-	-	-	-	-	-	0
1986	-	-	-	-	-	-	-	-	0
1987	-	-	-	-	-	-	-	-	0
1988	-	-	-	-	-	-	-	-	0
1989	-	-	-	-	-	-	-	-	0
1990	-	-	-	-	-	-	-	-	0
1991	-	-	-	-	-	-	-	-	0
1992	-	-	-	-	-	-	-	-	0
1993	-	-	-	-	-	-	-	-	0
1994	-	-	-	-	-	-	-	-	0
1995	-	-	-	-	-	-	-	-	0
1996	-	-	-	-	-	-	-	-	0
1997	-	-	-	-	-	-	-	-	0
1998	-	-	-	-	-	-	-	-	0
1999	-	-	-	-	-	-	-	-	0
2000	-	-	-	-	-	-	-	-	0
2001	-	-	-	-	-	-	-	-	0
2002	-	-	-	-	-	-	-	-	0
2003	-	-	-	-	-	-	-	-	0
2004	-	-	-	-	-	-	-	-	0
2005	-	-	-	-	-	-	-	-	0
Total	0	0	0	0	0	0	0	0	0

Finland (FI)

Number of BSE cases detected through the BSE Surveillance (Active and Passive) in Finland since 2001 per birth cohort and year of detection.

	N° of detected BSE cases per year											
Birth cohort	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	Total
1980	-	-	-	-	-	-	-	-	-	-	-	0
1981	-	-	-	-	-	-	-	-	-	-	-	0
1982	-	-	-	-	-	-	-	-	-	-	-	0
1983	-	-	-	-	-	-	-	-	-	-	-	0
1984	-	-	-	-	-	-	-	-	-	-	-	0
1985	-	-	-	-	-	-	-	-	-	-	-	0
1986	-	-	-	-	-	-	-	-	-	-	-	0
1987	-	-	-	-	-	-	-	-	-	-	-	0
1988	-	-	-	-	-	-	-	-	-	-	-	0
1989	-	-	-	-	-	-	-	-	-	-	-	0
1990	-	-	-	-	-	-	-	-	-	-	-	0
1991	-	-	-	-	-	-	-	-	-	-	-	0
1992	-	-	-	-	-	-	-	-	-	-	-	0
1993	-	-	-	-	-	-	-	-	-	-	-	0
1994	-	-	-	-	-	-	-	-	-	-	-	0
1995	1	-	-	-	-	-	-	-	-	-	-	1
1996	-	-	-	-	-	-	-	-	-	-	-	0
1997	-	-	-	-	-	-	-	-	-	-	-	0
1998	-	-	-	-	-	-	-	-	-	-	-	0
1999	-	-	-	-	-	-	-	-	-	-	-	0
2000	-	-	-	-	-	-	-	-	-	-	-	0
2001	-	-	-	-	-	-	-	-	-	-	-	0
2002	-	-	-	-	-	-	-	-	-	-	-	0
2003	-	-	-	-	-	-	-	-	-	-	-	0
2004	-	-	-	-	-	-	-	-	-	-	-	0
2005	-	-	-	-	-	-	-	-	-	-	-	0
Total	1	0	0	0	0	0	0	0	0	0	0	1

France (FR)

Number of BSE cases detected through the BSE Surveillance (Active and Passive) in France since 2001 per birth cohort and year of detection.

	N° of detected BSE cases per year											
Birth cohort	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	Total
1980	-	-	-	-	-	-	-	-	-	-	-	0
1981	-	-	-	-	-	-	-	-	-	-	-	0
1982	-	-	-	-	-	-	-	-	-	-	-	0
1983	-	-	-	-	-	-	-	-	-	-	-	0
1984	-	-	-	-	-	-	-	-	-	-	-	0
1985	-	-	-	-	-	-	-	-	-	-	-	0
1986	-	-	-	1	-	-	-	-	-	-	-	1
1987	-	-	1	-	-	-	-	-	-	-	-	1
1988	-	1	-	-	-	-	-	-	-	-	-	1
1989	-		1	-	-	-	-	-	-	-	-	1
1990	-	1	1	-	1	-	-	1	-	-	-	4
1991	-	3	2	-	-	-	-	-	-	-	-	5
1992	1	5	2	2	2	-	-	-	1	-	-	13
1993	29	17	7	4	3	1	-	2	2	-	-	65
1994	87	56	23	12	6	2	-	-	-	-	-	186
1995	132	102	41	10	7	-	3	-	2	2	1	300
1996	21	40	13	10	2	-	1	2	2	-	1	92
1997	4	10	16	4	3	2	1	1	1	1	-	43
1998	-	4	4	6	-	-	2	-	1	-	-	17
1999	-	-	-	2	5	2	-	-	1	-	1	11
2000	-	-	-	-	3	-	-	2	-	-	-	5
2001	-	-	-	-	-	1	-	-	-	1	-	2
2002	-	-	-	-	-	-	-	-	-	-	-	0
2003	-	-	-	-	-	-	-	-	-	-	-	0
2004	-	-	-	-	-	-	-	-	-	1	-	1
2005	-	-	-	-	-	-	-	-	-	-	-	0
Total	274	239	111	51	32	8	7	8	10	5	3	748

Germany (DE)

Number of BSE cases detected through the BSE Surveillance (Active and Passive) in Germany since 2001 per birth cohort and year of detection.

	N° of detected BSE cases per year											
Birth cohort	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	Total
1980	-	-	-	-	-	-	-	-	-	-	-	0
1981	-	-	-	-	-	-	-	-	-	-	-	0
1982	-	-	-	-	-	-	-	-	-	-	-	0
1983	-	-	-	-	-	-	-	-	-	-	-	0
1984	-	-	-	-	-	-	-	-	-	-	-	0
1985	-	-	-	-	-	-	-	-	-	-	-	0
1986	-	-	-	-	-	-	-	-	-	-	-	0
1987	-	1	-	-	-	-	-	-	-	-	-	1
1988	-	-	-	-	-	-	-	-	-	-	-	0
1989	-	-	-	-	-	-	-	-	-	-	-	0
1990	1	1	-	-	-	-	-	-	-	-	-	2
1991	1	-	-	1	-	-	-	-	-	-	-	2
1992	1	1	-	-	-	-	-	-	-	-	-	2
1993	-	3	-	-	-	-	-	-	-	-	-	3
1994	8	5	-	2	-	-	-	-	-	-	-	15
1995	40	32	8	2	1	1	-	-	-	-	-	84
1996	63	43	12	7	3	-	-	-	1	-	-	129
1997	5	10	13	14	1	-	-	-	-	-	-	43
1998	2	7	8	9	5	1	-	-	-	-	-	32
1999	-	-	12	18	11	9	3	-	-	-	-	53
2000	-	-	-	10	9	5	1	2	1	-	-	28
2001	-	-	-	-	2	-	-	-	-	-	-	2
2002	-	-	-	-	-	-	-	-	-	-	-	0
2003	-	-	-	-	-	-	-	-	-	-	-	0
2004	-	-	-	-	-	-	-	-	-	-	-	0
2005	-	-	-	-	-	-	-	-	-	-	-	0
Total	121	103	53	63	32	16	4	2	2	0	0	396

Greece (EL)

Number of BSE cases detected through the BSE Surveillance (Active and Passive) in Greece since 2001 per birth cohort and year of detection.

	N° of detected BSE cases per year											
Birth cohort	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	Total
1980	-	-	-	-	-	-	-	-	-	-	-	0
1981	-	-	-	-	-	-	-	-	-	-	-	0
1982	-	-	-	-	-	-	-	-	-	-	-	0
1983	-	-	-	-	-	-	-	-	-	-	-	0
1984	-	-	-	-	-	-	-	-	-	-	-	0
1985	-	-	-	-	-	-	-	-	-	-	-	0
1986	-	-	-	-	-	-	-	-	-	-	-	0
1987	-	-	-	-	-	-	-	-	-	-	-	0
1988	-	-	-	-	-	-	-	-	-	-	-	0
1989	-	-	-	-	-	-	-	-	-	-	-	0
1990	-	-	-	-	-	-	-	-	-	-	-	0
1991	-	-	-	-	-	-	-	-	-	-	-	0
1992	-	-	-	-	-	-	-	-	-	-	-	0
1993	-	-	-	-	-	-	-	-	-	-	-	0
1994	-	-	-	-	-	-	-	-	-	-	-	0
1995	-	-	-	-	-	-	-	-	-	-	-	0
1996	1	-	-	-	-	-	-	-	-	-	-	1
1997	-	-	-	-	-	-	-	-	-	-	-	0
1998	-	-	-	-	-	-	-	-	-	-	-	0
1999	-	-	-	-	-	-	-	-	-	-	-	0
2000	-	-	-	-	-	-	-	-	-	-	-	0
2001	-	-	-	-	-	-	-	-	-	-	-	0
2002	-	-	-	-	-	-	-	-	-	-	-	0
2003	-	-	-	-	-	-	-	-	-	-	-	0
2004	-	-	-	-	-	-	-	-	-	-	-	0
2005	-	-	-	-	-	-	-	-	-	-	-	0
Total	1	0	0	0	0	0	0	0	0	0	0	1

Hungary (HU)

Number of BSE cases detected through the BSE Surveillance (Active and Passive) in Hungary since 2004 per birth cohort and year of detection.

	N° of detected BSE cases per year								
Birth cohort	2004	2005	2006	2007	2008	2009	2010	2011	Total
1980	-	-	-	-	-	-	-	-	0
1981	-	-	-	-	-	-	-	-	0
1982	-	-	-	-	-	-	-	-	0
1983	-	-	-	-	-	-	-	-	0
1984	-	-	-	-	-	-	-	-	0
1985	-	-	-	-	-	-	-	-	0
1986	-	-	-	-	-	-	-	-	0
1987	-	-	-	-	-	-	-	-	0
1988	-	-	-	-	-	-	-	-	0
1989	-	-	-	-	-	-	-	-	0
1990	-	-	-	-	-	-	-	-	0
1991	-	-	-	-	-	-	-	-	0
1992	-	-	-	-	-	-	-	-	0
1993	-	-	-	-	-	-	-	-	0
1994	-	-	-	-	-	-	-	-	0
1995	-	-	-	-	-	-	-	-	0
1996	-	-	-	-	-	-	-	-	0
1997	-	-	-	-	-	-	-	-	0
1998	-	-	-	-	-	-	-	-	0
1999	-	-	-	-	-	-	-	-	0
2000	-	-	-	-	-	-	-	-	0
2001	-	-	-	-	-	-	-	-	0
2002	-	-	-	-	-	-	-	-	0
2003	-	-	-	-	-	-	-	-	0
2004	-	-	-	-	-	-	-	-	0
2005	-	-	-	-	-	-	-	-	0
Total	0	0	0	0	0	0	0	0	0

Ireland (IE)

Number of BSE cases detected through the BSE Surveillance (Active and Passive) in Ireland since 2001 per birth cohort and year of detection.

	N° of detected BSE cases per year											
Birth cohort	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	Total
1980	-	-	-	-	-	-	-	-	-	-	-	0
1981	-	-	-	-	-	-	-	-	-	-	-	0
1982	-	-	-	-	-	-	-	-	-	-	-	0
1983	-	-	-	-	-	-	-	-	-	-	-	0
1984	-	-	-	-	-	-	-	-	-	-	-	0
1985	-	-	-	-	-	-	-	-	-	-	-	0
1986	2	2	-	1	-	-	-	-	-	-	-	5
1987	-	2	-	-	-	-	-	-	-	-	-	2
1988	1	4	-	-	-	-	-	-	-	-	-	5
1989	2	1	4	3	-	1	-	-	-	-	-	11
1990	1	10	3	3	-	-	-	-	-	-	-	17
1991	6	10	6	3	1	2	1	-	-	-	-	29
1992	8	14	7	12	2	2	-	-	-	-	-	45
1993	21	40	25	16	11	1	2	-	-	-	2	118
1994	52	51	31	18	23	6	7	5	2	1	-	196
1995	110	132	73	43	19	11	6	2	3	-	-	399
1996	39	57	30	18	9	11	5	8	-	1	-	178
1997	-	5	3	3	-	2	-	2	-	-	1	16
1998	-	-	-	3	-	1	1	-	-	-	-	5
1999	-	1	-	2	3	-	-	1	-	-	-	7
2000	-	-	-	-	3	-	2	-	1	-	-	6
2001	-	-	-	-	2	1	-	1	1	-	-	5
2002	-	-	-	-	-	-	1	-	1	-	-	2
2003	-	-	-	-	-	-	-	1	-	-	-	1
2004	-	-	-	-	-	-	-	-	1	-	-	1
2005	-	-	-	-	-	-	-	-	-	-	-	0
Total	242	329	182	125	73	38	25	20	9	2	3	1 048

Italy (IT)

Number of BSE cases detected through the BSE Surveillance (Active and Passive) in Italy since 2001 per birth cohort and year of detection.

	N° of detected BSE cases per year											
Birth cohort	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	Total
1980	-	-	-	-	-	-	-	-	-	-	-	0
1981	-	-	-	-	-	-	-	-	-	-	-	0
1982	-	-	-	-	-	-	-	-	-	-	-	0
1983	-	-	-	-	-	-	-	-	-	-	-	0
1984	-	-	-	-	-	-	-	-	-	-	-	0
1985	-	-	-	-	-	-	-	-	-	-	-	0
1986	-	-	-	-	-	-	-	-	-	-	-	0
1987	1	-	-	-	-	-	-	-	-	-	-	1
1988	-	-	2	-	-	-	-	-	-	-	-	2
1989	-	-	-	-	-	-	-	-	-	-	-	0
1990	-	-	-	-	-	-	-	-	-	-	-	0
1991	-	1	-	-	-	-	-	-	-	-	-	1
1992	-	-	1	-	1	1	1	-	-	-	-	4
1993	1	-	3	-	-	-	-	-	-	-	-	4
1994	8	5	1	-	-	-	-	-	-	-	-	14
1995	12	10	4	-	-	-	-	1	-	-	-	27
1996	20	14	10	4	3	1	-	-	1	-	-	53
1997	8	4	9	2	1	1	1	-	-	-	1	27
1998	-	2	1	-	-	-	-	-	1	-	-	4
1999	-	-	-	1	2	2	-	-	-	-	-	5
2000	-	-	-	-	1	1	-	-	-	-	-	2
2001	-	-	-	-	-	1	-	-	-	-	-	1
2002	-	-	-	-	-	-	-	-	-	-	-	0
2003	-	-	-	-	-	-	-	-	-	-	-	0
2004	-	-	-	-	-	-	-	-	-	-	-	0
2005	-	-	-	-	-	-	-	-	-	-	-	0
Total	50	36	31	7	8	7	2	1	2	0	1	145

Latvia (LV)

Number of BSE cases detected through the BSE Surveillance (Active and Passive) in Latvia since 2004 per birth cohort and year of detection.

	N° of detected BSE cases per year								
Birth cohort	2004	2005	2006	2007	2008	2009	2010	2011	Total
1980	-	-	-	-	-	-	-	-	0
1981	-	-	-	-	-	-	-	-	0
1982	-	-	-	-	-	-	-	-	0
1983	-	-	-	-	-	-	-	-	0
1984	-	-	-	-	-	-	-	-	0
1985	-	-	-	-	-	-	-	-	0
1986	-	-	-	-	-	-	-	-	0
1987	-	-	-	-	-	-	-	-	0
1988	-	-	-	-	-	-	-	-	0
1989	-	-	-	-	-	-	-	-	0
1990	-	-	-	-	-	-	-	-	0
1991	-	-	-	-	-	-	-	-	0
1992	-	-	-	-	-	-	-	-	0
1993	-	-	-	-	-	-	-	-	0
1994	-	-	-	-	-	-	-	-	0
1995	-	-	-	-	-	-	-	-	0
1996	-	-	-	-	-	-	-	-	0
1997	-	-	-	-	-	-	-	-	0
1998	-	-	-	-	-	-	-	-	0
1999	-	-	-	-	-	-	-	-	0
2000	-	-	-	-	-	-	-	-	0
2001	-	-	-	-	-	-	-	-	0
2002	-	-	-	-	-	-	-	-	0
2003	-	-	-	-	-	-	-	-	0
2004	-	-	-	-	-	-	-	-	0
2005	-	-	-	-	-	-	-	-	0
Total	0	0	0	0	0	0	0	0	0

Lithuania (LT)

Number of BSE cases detected through the BSE Surveillance (Active and Passive) in Lithuania since 2004 per birth cohort and year of detection.

	N° of detected BSE cases per year								
Birth cohort	2004	2005	2006	2007	2008	2009	2010	2011	Total
1980	-	-	-	-	-	-	-	-	0
1981	-	-	-	-	-	-	-	-	0
1982	-	-	-	-	-	-	-	-	0
1983	-	-	-	-	-	-	-	-	0
1984	-	-	-	-	-	-	-	-	0
1985	-	-	-	-	-	-	-	-	0
1986	-	-	-	-	-	-	-	-	0
1987	-	-	-	-	-	-	-	-	0
1988	-	-	-	-	-	-	-	-	0
1989	-	-	-	-	-	-	-	-	0
1990	-	-	-	-	-	-	-	-	0
1991	-	-	-	-	-	-	-	-	0
1992	-	-	-	-	-	-	-	-	0
1993	-	-	-	-	-	-	-	-	0
1994	-	-	-	-	-	-	-	-	0
1995	-	-	-	-	-	-	-	-	0
1996	-	-	-	-	-	-	-	-	0
1997	-	-	-	-	-	-	-	-	0
1998	-	-	-	-	-	-	-	-	0
1999	-	-	-	-	-	-	-	-	0
2000	-	-	-	-	-	-	-	-	0
2001	-	-	-	-	-	-	-	-	0
2002	-	-	-	-	-	-	-	-	0
2003	-	-	-	-	-	-	-	-	0
2004	-	-	-	-	-	-	-	-	0
2005	-	-	-	-	-	-	-	-	0
Total	0	0	0	0	0	0	0	0	0

Luxemburg (LU)

Number of BSE cases detected through the BSE Surveillance (Active and Passive) in Luxemburg since 2001 per birth cohort and year of detection.

	N° of detected BSE cases per year											
Birth cohort	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	Total
1980	-	-	-	-	-	-	-	-	-	-	-	0
1981	-	-	-	-	-	-	-	-	-	-	-	0
1982	-	-	-	-	-	-	-	-	-	-	-	0
1983	-	-	-	-	-	-	-	-	-	-	-	0
1984	-	-	-	-	-	-	-	-	-	-	-	0
1985	-	-	-	-	-	-	-	-	-	-	-	0
1986	-	-	-	-	-	-	-	-	-	-	-	0
1987	-	-	-	-	-	-	-	-	-	-	-	0
1988	-	-	-	-	-	-	-	-	-	-	-	0
1989	-	-	-	-	-	-	-	-	-	-	-	0
1990	-	-	-	-	-	-	-	-	-	-	-	0
1991	-	-	-	-	-	-	-	-	-	-	-	0
1992	-	-	-	-	-	-	-	-	-	-	-	0
1993	-	-	-	-	-	-	-	-	-	-	-	0
1994	-	-	-	-	-	-	-	-	-	-	-	0
1995	-	-	-	-	-	-	-	-	-	-	-	0
1996	-	1	-	-	-	-	-	-	-	-	-	1
1997	-	-	-	-	-	-	-	-	-	-	-	0
1998	-	-	-	-	-	-	-	-	-	-	-	0
1999	-	-	-	-	-	-	-	-	-	-	-	0
2000	-	-	-	-	-	-	-	-	-	-	-	0
2001	-	-	-	-	1	-	-	-	-	-	-	1
2002	-	-	-	-	-	-	-	-	-	-	-	0
2003	-	-	-	-	-	-	-	-	-	-	-	0
2004	-	-	-	-	-	-	-	-	-	-	-	0
2005	-	-	-	-	-	-	-	-	-	-	-	0
Total	0	1	0	0	1	0	0	0	0	0	0	2

Malta (MT)

Number of BSE cases detected through the BSE Surveillance (Active and Passive) in Malta since 2004 per birth cohort and year of detection.

	N° of detected BSE cases per year								
Birth cohort	2004	2005	2006	2007	2008	2009	2010	2011	Total
1980	-	-	-	-	-	-	-	-	0
1981	-	-	-	-	-	-	-	-	0
1982	-	-	-	-	-	-	-	-	0
1983	-	-	-	-	-	-	-	-	0
1984	-	-	-	-	-	-	-	-	0
1985	-	-	-	-	-	-	-	-	0
1986	-	-	-	-	-	-	-	-	0
1987	-	-	-	-	-	-	-	-	0
1988	-	-	-	-	-	-	-	-	0
1989	-	-	-	-	-	-	-	-	0
1990	-	-	-	-	-	-	-	-	0
1991	-	-	-	-	-	-	-	-	0
1992	-	-	-	-	-	-	-	-	0
1993	-	-	-	-	-	-	-	-	0
1994	-	-	-	-	-	-	-	-	0
1995	-	-	-	-	-	-	-	-	0
1996	-	-	-	-	-	-	-	-	0
1997	-	-	-	-	-	-	-	-	0
1998	-	-	-	-	-	-	-	-	0
1999	-	-	-	-	-	-	-	-	0
2000	-	-	-	-	-	-	-	-	0
2001	-	-	-	-	-	-	-	-	0
2002	-	-	-	-	-	-	-	-	0
2003	-	-	-	-	-	-	-	-	0
2004	-	-	-	-	-	-	-	-	0
2005	-	-	-	-	-	-	-	-	0
Total	0	0	0	0	0	0	0	0	0

Netherlands (NL)

Number of BSE cases detected through the BSE Surveillance (Active and Passive) in the Netherlands since 2001 per birth cohort and year of detection.

	N° of detected BSE cases per year											
Birth cohort	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	Total
1980	-	-	-	-	-	-	-	-	-	-	-	0
1981	-	-	-	-	-	-	-	-	-	-	-	0
1982	-	-	-	-	-	-	-	-	-	-	-	0
1983	-	-	-	-	-	-	-	-	-	-	-	0
1984	-	-	-	-	-	-	-	-	-	-	-	0
1985	-	-	-	-	-	-	-	-	-	-	-	0
1986	-	-	-	-	-	-	-	-	-	-	-	0
1987	-	-	-	-	-	-	-	-	-	-	-	0
1988	1	-	-	-	-	-	-	-	-	-	-	1
1989	-	-	-	-	-	-	-	-	-	-	-	0
1990	-	-	-	-	-	-	-	-	-	-	-	0
1991	-	-	1	1	-	-	-	-	-	-	-	2
1992	1	1	-	-	-	-	-	-	-	-	-	2
1993	2	1	-	-	-	-	-	-	-	-	-	3
1994	2	2	-	-	-	-	-	-	-	-	-	4
1995	4	3	-	-	-	-	-	-	-	-	-	7
1996	9	10	10	3	-	-	-	-	-	1	-	33
1997	1	4	5	1	-	1	-	-	-	1	-	13
1998	-	3	1	1	-	1	-	-	-	-	-	6
1999	-	-	1	-	-	-	1	-	-	1	-	3
2000	-	-	-	-	2	-	1	1	-	-	-	4
2001	-	-	-	-	1	-	-	-	-	-	-	1
2002	-	-	-	-	-	-	-	-	-	-	-	0
2003	-	-	-	-	-	-	-	-	-	-	-	0
2004	-	-	-	-	-	-	-	-	-	-	-	0
2005	-	-	-	-	-	-	-	-	-	-	-	0
Total	20	24	18	6	3	2	2	1	0	3	0	79

Poland (PL)

Number of BSE cases detected through the BSE Surveillance (Active and Passive) in Poland since 2004 per birth cohort and year of detection.

	N° of detected BSE cases per year								
Birth cohort	2004	2005	2006	2007	2008	2009	2010	2011	Total
1980	-	-	-	-	-	-	-	-	0
1981	-	-	-	-	-	-	-	-	0
1982	-	-	-	-	-	-	-	-	0
1983	-	-	-	-	-	-	-	-	0
1984	-	-	-	-	-	-	-	-	0
1985	-	-	-	-	-	-	-	-	0
1986	-	-	-	-	-	-	-	-	0
1987	-	-	-	-	-	-	-	-	0
1988	-	-	-	-	-	-	-	-	0
1989	-	-	-	-	-	-	-	-	0
1990	-	-	-	-	-	-	-	-	0
1991	-	-	-	-	-	-	-	-	0
1992	2	1	-	-	-	-	-	-	3
1993	-	-	-	-	-	-	-	-	0
1994	1	1	2	-	-	-	-	-	4
1995	1	1	1	3	-	1	1	-	8
1996	4	2	3	-	-	-	-	1	10
1997	-	2	1	-	-	-	-	-	3
1998	2	2	1	-	-	-	-	-	5
1999	-	4	1	3	2	2	-	-	12
2000	1	3	-	1	1	1	1	-	8
2001	-	2	1	-	-	-	-	-	3
2002	-	1	-	-	-	-	-	-	1
2003	-	-	-	1	1	-	-	-	2
2004	-	-	-	1	-	-	-	-	1
2005	-	-	-	-	1	-	-	-	1
Total	11	19	10	9	5	4	2	1	61

Portugal (PT)

Number of BSE cases detected through the BSE Surveillance (Active and Passive) in Portugal since 2001 per birth cohort and year of detection.

	N° of detected BSE cases per year											
Birth cohort	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	Total
1980	-	-	-	-	-	-	-	-	-	-	-	0
1981	-	-	-	-	-	-	-	-	-	-	-	0
1982	-	-	-	-	-	-	-	-	-	-	-	0
1983	-	-	-	-	-	-	-	-	-	-	-	0
1984	-	1	-	-	-	-	-	-	-	-	-	1
1985	-	-	-	-	1	-	-	-	-	-	-	1
1986	-	-	-	-	-	-	-	-	-	-	-	0
1987	-	1	1	-	-	-	-	-	-	-	-	2
1988	-	-	-	-	-	1	-	-	-	-	1	2
1989	2	-	1	-	-	-	-	-	-	-	-	3
1990	1	-	1	3	2	3	-	-	-	-	-	10
1991	-	1	-	1	-	1	-	-	-	-	-	3
1992	3	1	4	3	2	-	1	-	-	-	-	14
1993	22	11	24	14	8	3	3	3	-	3	-	91
1994	38	21	19	13	7	7	2	4	1	1	1	114
1995	17	19	12	8	6	1	1	2	3	-	1	70
1996	22	19	23	9	7	2	1	3	1	-	-	87
1997	5	8	28	23	10	4	2	4	-	-	-	84
1998	-	1	18	12	7	8	-	1	-	-	2	49
1999	-	2	1	2	2	2	2	1	1	-	-	13
2000	-	-	-	1	-	-	-	-	-	1	-	2
2001	-	-	-	-	-	-	1	-	-	-	-	1
2002	-	-	-	-	1	-	-	-	-	-	-	1
2003	-	-	-	-	-	-	-	-	-	-	-	0
2004	-	-	-	-	-	-	-	-	-	-	-	0
2005	-	-	-	-	-	-	-	-	-	-	-	0
Total	110	85	132	89	53	32	13	18	6	5	5	548

Slovakia (SK)

Number of BSE cases detected through the BSE Surveillance (Active and Passive) in Slovakia since 2004 per birth cohort and year of detection.

	N° of detected BSE cases per year								
Birth cohort	2004	2005	2006	2007	2008	2009	2010	2011	Total
1980	-	-	-	-	-	-	-	-	0
1981	-	-	-	-	-	-	-	-	0
1982	-	-	-	-	-	-	-	-	0
1983	-	-	-	-	-	-	-	-	0
1984	-	-	-	-	-	-	-	-	0
1985	-	-	-	-	-	-	-	-	0
1986	-	-	-	-	-	-	-	-	0
1987	-	-	-	-	-	-	-	-	0
1988	-	-	-	-	-	-	-	-	0
1989	-	-	-	-	-	-	-	-	0
1990	-	-	-	-	-	-	-	-	0
1991	-	-	-	-	-	-	-	-	0
1992	-	-	-	-	-	-	-	-	0
1993	-	-	-	-	-	-	-	-	0
1994	-	-	-	-	-	-	-	-	0
1995	2	-	-	-	-	-	-	-	2
1996	-	-	-	-	-	-	-	-	0
1997	-	-	-	-	-	-	-	-	0
1998	-	-	-	-	-	-	-	-	0
1999	1	-	-	-	-	-	-	-	1
2000	2	2	-	-	-	-	-	-	4
2001	2	-	-	2	1	-	-	-	5
2002	-	-	-	-	-	-	1	-	1
2003	-	-	-	-	-	-	-	-	0
2004	-	-	-	-	-	-	-	-	0
2005	-	-	-	-	-	-	-	-	0
Total	7	2	0	2	1	0	1	0	13

Slovenia (SI)

Number of BSE cases detected through the BSE Surveillance (Active and Passive) in Slovenia since 2001 per birth cohort and year of detection.

	N° of detected BSE cases per year											
Birth cohort	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	Total
1980	-	-	-	-	-	-	-	-	-	-	-	0
1981	-	-	-	-	-	-	-	-	-	-	-	0
1982	-	-	-	-	-	-	-	-	-	-	-	0
1983	-	-	-	-	-	-	-	-	-	-	-	0
1984	-	-	-	-	-	-	-	-	-	-	-	0
1985	-	-	-	-	-	-	-	-	-	-	-	0
1986	-	-	-	-	-	-	-	-	-	-	-	0
1987	-	-	-	-	-	-	-	-	-	-	-	0
1988	-	-	-	-	-	-	-	-	-	-	-	0
1989	-	-	-	-	-	-	-	-	-	-	-	0
1990	-	-	-	-	-	-	-	-	-	-	-	0
1991	-	-	-	-	-	-	-	-	-	-	-	0
1992	-	-	-	-	-	-	-	-	-	-	-	0
1993	-	-	-	-	-	-	-	-	-	-	-	0
1994	-	-	-	-	-	-	-	-	-	-	-	0
1995	-	1	-	-	-	-	-	-	-	-	-	1
1996	1	-	-	-	-	-	-	-	-	-	-	1
1997	-	-	-	-	-	-	-	-	-	-	-	0
1998	-	-	-	1	-	-	-	-	-	-	-	1
1999	-	-	1	-	-	-	-	-	-	-	-	1
2000	-	-	-	1	1	1	1	-	-	-	-	4
2001	-	-	-	-	-	-	-	-	-	-	-	0
2002	-	-	-	-	-	-	-	-	-	-	-	0
2003	-	-	-	-	-	-	-	-	-	-	-	0
2004	-	-	-	-	-	-	-	-	-	-	-	0
2005	-	-	-	-	-	-	-	-	-	-	-	0
Total	1	1	1	2	1	1	1	0	0	0	0	8

Spain (ES)

Number of BSE cases detected through the BSE Surveillance (Active and Passive) in Spain since 2001 per birth cohort and year of detection.

	N° of detected BSE cases per year											
Birth cohort	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	Total
1980	-	-	-	-	-	-	-	-	-	-	-	0
1981	-	-	-	-	-	-	-	-	-	-	-	0
1982	-	-	-	-	-	-	-	-	-	-	-	0
1983	-	-	-	-	-	-	-	-	-	-	-	0
1984	-	-	-	-	-	-	-	-	-	-	-	0
1985	-	-	-	-	-	-	-	-	-	-	-	0
1986	1	-	-	-	-	-	-	-	-	-	-	1
1987	1	-	1	-	-	-	-	-	-	-	-	2
1988	1	1	1	-	-	-	-	-	-	-	-	3
1989	1	2	1	-	-	-	1	-	1	-	-	6
1990	1	-	1	1	-	-	-	1	1	-	-	5
1991	-	-	-	-	-	-	-	-	-	-	1	1
1992	1	1	2	1	-	-	1	1	-	-	-	7
1993	10	12	6	5	1	1	1	-	2	-	-	38
1994	13	9	9	4	3	1	-	1	1	-	-	41
1995	22	33	24	9	2	1	3	1	1	2	1	99
1996	20	33	34	14	7	4	1	2	-	2	-	117
1997	11	28	56	30	13	10	1	3	2	1	1	156
1998	-	7	26	49	22	18	7	4	3	1	1	138
1999	-	1	4	19	23	17	6	4	3	4	-	81
2000	-	-	1	5	26	23	11	3	4	2	2	77
2001	-	-	-	-	-	1	-	3	-	-	-	4
2002	-	-	-	-	1	-	-	1	-	-	-	2
2003	-	-	-	-	-	-	-	-	-	-	-	0
2004	-	-	-	-	-	-	-	-	-	1	-	1
2005	-	-	-	-	-	-	-	-	-	-	1	1
Total	82	127	166	137	98	76	32	24	18	13	7	780

Sweden (SE)

Number of BSE cases detected through the BSE Surveillance (Active and Passive) in Sweden since 2001 per birth cohort and year of detection.

	N° of detected BSE cases per year											
Birth cohort	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	Total
1980	-	-	-	-	-	-	-	-	-	-	-	0
1981	-	-	-	-	-	-	-	-	-	-	-	0
1982	-	-	-	-	-	-	-	-	-	-	-	0
1983	-	-	-	-	-	-	-	-	-	-	-	0
1984	-	-	-	-	-	-	-	-	-	-	-	0
1985	-	-	-	-	-	-	-	-	-	-	-	0
1986	-	-	-	-	-	-	-	-	-	-	-	0
1987	-	-	-	-	-	-	-	-	-	-	-	0
1988	-	-	-	-	-	-	-	-	-	-	-	0
1989	-	-	-	-	-	-	-	-	-	-	-	0
1990	-	-	-	-	-	-	-	-	-	-	-	0
1991	-	-	-	-	-	-	-	-	-	-	-	0
1992	-	-	-	-	-	-	-	-	-	-	-	0
1993	-	-	-	-	-	-	-	-	-	-	-	0
1994	-	-	-	-	-	1	-	-	-	-	-	1
1995	-	-	-	-	-	-	-	-	-	-	-	0
1996	-	-	-	-	-	-	-	-	-	-	-	0
1997	-	-	-	-	-	-	-	-	-	-	-	0
1998	-	-	-	-	-	-	-	-	-	-	-	0
1999	-	-	-	-	-	-	-	-	-	-	-	0
2000	-	-	-	-	-	-	-	-	-	-	-	0
2001	-	-	-	-	-	-	-	-	-	-	-	0
2002	-	-	-	-	-	-	-	-	-	-	-	0
2003	-	-	-	-	-	-	-	-	-	-	-	0
2004	-	-	-	-	-	-	-	-	-	-	-	0
2005	-	-	-	-	-	-	-	-	-	-	-	0
Total	0	0	0	0	0	1	0	0	0	0	0	1

United Kingdom (UK)

Number of BSE cases detected through the BSE Surveillance (Active and Passive) in United Kingdom since 2001 per birth cohort and year of detection.

	N° of detected BSE cases per year											
Birth cohort	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	Total
1980	-	1	-	-	-	-	-	-	-	-	-	1
1981	1	-	-	-	-	-	-	-	-	1	-	2
1982	-	-	-	-	-	-	-	-	-	-	-	
1983	-	-	1	-	-	-	-	-	-	-	-	1
1984	1	2	-	-	-	1	-	-	-	-	-	4
1985	1	2	2	-	-	-	-	-	-	-	-	5
1986	10	8	3	1	1	-	-	-	-	-	-	23
1987	19	26	6	6	6	1	-	1	-	-	-	65
1988	17	22	18	8	-	-	-	2	-	-	-	67
1989	20	34	14	14	5	4	-	1	-	-	-	92
1990	24	42	16	14	6	4	1	1	-	-	-	108
1991	58	63	37	21	20	5	-	1	-	1	-	206
1992	104	130	68	37	29	12	7	-	1	-	1	389
1993	241	160	115	56	33	20	11	4	2	1	-	643
1994	362	303	133	73	51	30	16	6	2	1	2	979
1995	310	275	136	65	31	23	9	4	1	1	-	855
1996	25	37	24	10	5	7	-	8	-	1	1	118
1997	9	13	18	4	4	3	1	3	1	1	1	58
1998	1	4	14	11	6	3	7	2	-	1	1	50
1999	-	1	5	6	11	4	3	3	-	-	1	34
2000	-	-	-	2	4	4	3	-	-	1	1	15
2001	-	-	-	-	1	4	1	1	-	-	-	7
2002	-	-	-	-	-	1	2	2	1	-	-	6
2003	-	-	-	-	-	-	-	2	3	-	-	5
2004	-	-	-	-	-	-	-	-	-	2	-	2
2005	-	-	-	-	-	-	-	-	-	-	-	
Total	1 203	1 123	610	328	213	126	61	41	11	11	8	3735

ABBREVIATIONS

EU25	The group of following EU MSs: Austria, Belgium, Czech Republic, Cyprus, Denmark, Estonia, Finland, France, Germany, Greece, Hungary, Ireland, Italy, Latvia, Lithuania, Luxembourg, Malta, Netherlands, Poland, Portugal, Slovenia, Slovakia, Spain, Sweden and United Kingdom.
EU17	The group of following EU MSs: Austria, Belgium, Cyprus, Denmark, Finland, France, Germany, Greece, Ireland, Italy, Luxembourg, Netherlands, Portugal, Slovenia, Spain, Sweden and United Kingdom.
EU8	The group of following EU MSs: Czech Republic, Estonia, Hungary, Latvia, Lithuania, Malta, Poland and Slovakia.
EU5	The group of following EU MSs: Estonia, Hungary, Latvia, Lithuania and Malta.
EU3	The group of following EU MSs: Czech Republic, Poland and Slovakia.
C-BSE	Classical BSE
C-TSEMM	Cattle TSE Monitoring Model
H-BSE	H-type BSE
L-BSE	L-type BSE
MS/MSs	Member State/Member States of the European Union
PrP^{res}	The PK resistant core of the the N-terminally truncated form of abnormal disease-associated isoforms (PrP^{Sc}) of the normal cellular prion protein (PrP^{C}), following digestion with proteinase K in the presence of detergents.