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## The European Union summary report on surveillance for the presence of transmissible spongiform encephalopathies (TSE) in 2016

European Food Safety Authority

### Abstract

This report presents the results of surveillance activities on transmissible spongiform encephalopathies (TSEs) in bovine animals, sheep, goats, cervids and other species, as well as genotyping data in sheep, carried out in 2016 in the European Union according to Regulation (EC) 999/2001, and in Iceland, Norway and Switzerland. In 2016, 1,352,585 bovine animals were tested in the European Union (5% less than in 2015). For the first time, the United Kingdom did not report any case of bovine spongiform encephalopathy (BSE), whereas France reported one classical and three atypical cases (H), and Spain one atypical case (H). The classical BSE case was born after the enforcement of the total EU-wide feed ban in 2001 (BARB case). In 2016, 286,351 sheep and 110,832 goats were tested (5% and 11% less than in 2015, respectively). Sheep scrapie was reported by 20 Member States (MSs) (685 cases) and goat scrapie by 9 MSs (634 cases). A total of 25 ovine scrapie cases were reported by Iceland and Norway. At the EU level, the occurrence of scrapie in small ruminants remains stable, with classical scrapie (1,175 cases) being reported more frequently than atypical scrapie (135 cases). A total of 97.2% of the classical scrapie cases in sheep occurred in animals with genotypes belonging to the susceptible group, and a random sampling showed that 26.6% of the genotyped sheep held genotypes of the susceptible group (excluding Cyprus). In 2016, five cases of chronic wasting disease were reported in cervids by Norway: three in wild reindeer and two in moose. It was the first time that this disease is reported in Europe. A total of 2,712 cervids were tested for TSEs in seven different member states, 90% of them in Romania, with negative results. A total of 490 animals from other non-ruminant species were tested in four different member states, with negative results.

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**Amendment:** An editorial correction was carried out that does not materially affect the contents or outcome of this scientific output. On page 36 numbers 112/154 were revised to show 112/554, and on page 41 in Table 15 the total number for 2005 has been corrected from 1,808 to 1,817. Table A.5 the total for EFSA countries has been corrected from 894,050 to 1,200,563 and from 2.19 to 1.54. To avoid confusion, the older version has been removed from the EFSA Journal, but is available on request.

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## Summary

This report of the European Food Safety Authority (EFSA) presents the detailed results of surveillance activities on animal transmissible spongiform encephalopathies (TSE) carried out during 2016 in the European Union (EU) and three non-Member States (non-MSs) as well as genotyping data in sheep. Summary statistics and trends are also presented covering the period between 2001 and 2016.

TSE monitoring data for bovine animals, small ruminants, cervids and species other than ruminants are reported by country according to Regulation (EC) 999/2001 (the TSE Regulation) and consist of testing data (reported monthly) as well as case data. Data regarding the genotyping of the ovine scrapie cases and of randomly selected sheep were retrieved from the annual reports submitted by the Member States (MSs) and non-MSs in accordance with Article 6.4 of, and as specified in Chapter B.I of Annex III of the TSE Regulation.

A descriptive summary of the reported data is provided at MS level. Where possible, descriptions and calculations were stratified according to the available variables in the database, such as surveillance target group (healthy slaughtered animals, animals culled under bovine spongiform encephalopathy (BSE)/TSE control and eradication measures, etc.) or surveillance type (passive vs active), country, sampling year (since 2001 for bovine animals and 2002 for small ruminants), case type (i.e. classical BSE (C-BSE), atypical BSE (H-BSE or L-BSE), classical scrapie (CS) or atypical scrapie (AS)), flock status (infected/non-infected) and age class/group.

Since 2001, approximately 115 million cattle have been tested for BSE in the EU, with a sustained decrease in the number of animals tested over time following amendments to the TSE Regulation. The total number of samples tested for BSE in the EU fell from 1,442,446 in 2015 to 1,352,585 in 2016, representing a reduction of 5%. This was due to the decrease in the testing in the HS category (20% reduction from 491,052 in 2015 to 390,586 in 2016), mainly in three MSs: Germany, France and Poland. However, the number of samples tested from risk animals (AM + ES + FS) over 48 months of age showed an increase (+ 28,118) compared with 2015. In 2016, over 71% of all samples tested in the 31 reporting countries were from fallen stock.

In 2016, five BSE cases were detected in the EU, all among fallen stock, from two different MSs: France and Spain. One French case was of C-BSE type, born after the reinforced EU-wide feed ban imposed on 1 January 2001 (BARB case). The remaining four cases were of H-BSE type: three in France and one in Spain. For the first time since BSE cases have been reported, no C-BSE or atypical BSE cases were reported by the United Kingdom in 2016.

Since 2002, approximately 8.8 million small ruminants have been tested for scrapie in the EU. In 2016, a total of 286,351 sheep were tested, representing a decrease of 5%, compared with 2015. A total of 685 cases of scrapie in sheep were reported in the EU MSs: 554 of them were CS, 122 were AS and 9 were unknown type. CS was reported by nine different MSs, whereas AS cases were reported by 18 MSs. Most of the CS cases in sheep were detected in four MS: Greece (227), followed by Italy (143), Spain (91) and Romania (75). Portugal and Hungary are the MS with the largest number of AS cases reported: 28 and 23, respectively. In addition, 25 scrapie cases in sheep were reported by two non-MSs: 11 CS cases in Iceland and 14 AS cases in Norway.

In 2016, a total of 110,832 goats were tested, representing a decrease of 11%, compared with 2015. A total of 634 cases of scrapie in goats were reported in the EU MSs: 621 of them were CS and 13 were AS. CS was reported by seven MSs with Cyprus accounting for 91.8% of all CS cases (570), followed by Spain (19 cases) and Greece (12 cases). AS was reported by five MSs, with Spain the country with the largest number of AS cases reported in goats: 5. Overall at the EU level, there is no consistent decline in the occurrence of scrapie in small ruminants, either in terms of the absolute number of cases or in the proportion of cases in tested animals.

A total of 97.2% of the CS cases in sheep reported in 2016 belonged to susceptible genotypes (NSP3, NSP30, NSP4 or NSP5), and the genotyping data from random samples of the EU sheep population (excluding CY) showed that 26.6% of the genotyped sheep held genotypes of the susceptible group.

Chronic Wasting Disease (CWD) was confirmed for the first time in Europe in 2016, in Norway, where out of a total of 10,139 tested cervids five cases were reported: three in wild reindeer and two in moose. During the same period, 2,712 cervids were tested in seven different MSs and all were found negative. Most of the tested cervids (> 90%) were reported by Romania. Four MSs tested for TSE 490 samples from animals of species other than ruminants and all were negative.

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## 1. Introduction

### 1.1. Background and Terms of Reference

According to Part I.A, Chapter B.I, Annex III of Regulation (EC) 999/2001<sup>1</sup> (hereinafter referred to as the transmissible spongiform encephalopathy (TSE) Regulation), the information to be presented by Member States (MSs) in their annual report, as provided for in Article 6(4), includes:

- 1) The number of suspected cases placed under official movement restrictions in accordance with Article 12(1), per animal species.
- 2) The number of suspected cases subject to laboratory examination in accordance with Article 12(2), per animal species, including the results of the rapid and confirmatory tests (number of positives and negatives) and, with regard to bovine animals, the age distribution of all tested animals. The age distribution should be grouped as follows: 'below 24 months', distribution per 12 months between 24 and 155 months, and 'above 155 months' of age.
- 3) The number of flocks where suspected cases in ovine and caprine animals have been reported and investigated pursuant to Article 12(1) and (2).
- 4) The number of bovine animals tested within each subpopulation referred to in Chapter A, Part I, points 2.1, 2.2, 3.1 and 5. The method of the sample selection, the results of the rapid and confirmatory tests and the age distribution of the tested animals grouped as set out in point 2 shall be provided.
- 5) The number of ovine and caprine animals and flocks tested within each subpopulation referred to in Chapter A, Part II, points 2, 3, 5 and 6 together with the method for sample selection and the results of the rapid and confirmatory tests.
- 6) The geographical distribution, including the country of origin if not the same as the reporting country, of positive cases of BSE and scrapie. The year, and where possible the month of birth shall be given for each TSE case in bovine, ovine and caprine animals. TSE cases that have been considered atypical shall be indicated. For scrapie cases, the results of the primary and secondary molecular testing, referred to in Annex X, Chapter C, point 3.2 (c), shall be reported, where appropriate.
- 7) In animals other than bovine, ovine and caprine animals, the number of samples and confirmed TSE cases per species.
- 8) The genotype, and, where possible, the breed, of each ovine animal either found positive for TSE and sampled in accordance with Chapter A, Part II, point 8.1, or sampled in accordance with Chapter A, Part II, point 8.2.

According to Chapter B.II, 'the compilation of reports containing the information referred to in B.I and submitted to the Commission (which shall send it to the European Food Safety Authority) on a monthly basis in the electronic format agreed between the MSs, the Commission and the European Food Safety Authority or, with regard to the information referred to in point 8 on a quarterly basis, may constitute the annual report as required by Article 6(4), provided that the information is updated whenever additional information becomes available'.

The Union summary shall be presented in a tabled format covering at least the information referred to in Part I.A Chapter B.I for each MS.

### 1.2. Surveillance of TSE in the European Union

#### 1.2.1. Legal basis

Animals suspected of a TSE should be examined in accordance with Article 12.2 of the TSE Regulation. The legal framework for the active monitoring of ruminants for the presence of TSE is laid down in Article 6 of the TSE Regulation, and specified in its Annex III, Chapter A.

Of the 27 MSs at that time, Commission Decision 2009/719/EC<sup>2</sup>, allowed 25 (all except Bulgaria and Romania) to apply a revised bovine spongiform encephalopathy (BSE) monitoring programme.

<sup>1</sup> 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.

<sup>2</sup> 2009/719/EC: Commission Decision of 28 September 2009 authorising certain Member States to revise their annual BSE monitoring programmes. OJ L 256, 29.9.2009, p. 35–37, as last amended.

Commission Implementing Decision 2013/76/EU<sup>3</sup> of 4 February 2013, amending Commission Decision 2009/719/EC, authorised these 25 MSs to decide to stop testing slaughtered bovine animals that had been healthy.

Commission Decision 2016/851 of 26 May 2016 authorised the revision of the Croatian BSE annual monitoring programme under the same conditions as granted to the 25 MSs currently listed in the Annex to Decision 2009/719/EC<sup>4</sup>.

The legal basis for the sample collection and for the test methods is laid down in Chapter C of Annex X of the TSE Regulation. From 2005, Annex X (as amended by Commission Regulation (EC) No 36/2005<sup>5</sup>) also provides for mandatory discriminatory testing for BSE of TSE cases detected in small ruminants.

The legal basis for the testing for the determination of the prion protein genotypes (genotype testing) is laid down in points 8.1 and 8.2 of Chapter A in Annex III of the TSE Regulation.

### 1.2.2. BSE surveillance of bovine animals

The BSE surveillance of bovine animals is based on the testing of samples from the following target groups:

- **Animals clinically suspected of being infected by BSE (SU):** live, slaughtered or dead animals that show or have shown neurological or behavioural disorders or a progressive deterioration of the general condition linked to impairment of the central nervous system and for which the information gathered on the basis of a clinical examination, response to treatment, a post-mortem examination or *ante-* or *post-mortem* laboratory analysis does not allow an alternative diagnosis to be established. BSE shall be suspected in bovine animals that have produced a positive result from a rapid test specifically for BSE as defined in Article 3.1 (h) of the TSE Regulation and subject to the measures described in Articles 12 and 13.
- **Animals culled under BSE eradication measures (EM):** birth cohorts (bovine animals born in the same herd as the affected bovine animal within the 12 months preceding or following the date of birth of the affected bovine animal), rearing cohorts (bovine animals that at any time during the first year of their lives were reared together with the affected bovine animal during the first year of its life) (point 2, Annex I) and, where the disease was confirmed in a female animal, its progeny born within a period of 2 years prior to or after the clinical onset of the disease (point 1(a) Chapter B, Annex VII).
- **Animals with clinical signs at ante-mortem (AM):** bovine animals with (abnormal) observations at *ante-mortem* inspections (Article 6 1a(a)).
- **Emergency slaughtered (ES):** bovine animals sent for emergency slaughter (Article 6.1a (a)).
- **Fallen stock (FS):** bovine animals not slaughtered for human consumption that have died or been killed on the farm, during transport or in an abattoir (Article 6 1a(c)). (MSs may decide to derogate from this provision in remote areas with low animal density, where no collection of dead animals is organised). The derogation shall not cover more than 10% of the bovine population in the MSs.
- **Healthy slaughtered (HS):** bovine animals slaughtered for human consumption (Article 6.1a (b)).

The categories of bovine animals to be submitted for BSE testing are defined in the TSE Regulation and are based on a combination of age (limits) and surveillance target groups. The general rules for BSE surveillance, applied in 2016, are summarised in Table 1. The age limits, to be applied for certain surveillance target groups, have been changed over time. In most of the countries, the testing age limit for the healthy slaughtered category has been shifted from 30 to 48 months in 2009, and to 72 months in 2011. Based on the revision of the testing regimes provided by Decision 2009/719/EC, the testing of HS has been discontinued by most of the MSs. Regarding the risk categories (i.e. FS, ES,

<sup>3</sup> 2013/76/EU: Commission Implementing Decision of 4 February 2013 amending Decision 2009/719/EC authorising certain Member States to revise their annual BSE monitoring programmes. OJ L 35, 6.2.2013, p. 6–7.

<sup>4</sup> Commission Implementing Decision (EU) 2016/851 of 26 May 2016 amending the Annex to Decision 2009/719/EC as regards the authorisation for Croatia to revise its BSE annual monitoring programme (notified under document C (2016) 3097). JOL\_2016\_141\_R\_0019.

<sup>5</sup> Commission Regulation (EC) No 36/2005 of 12 January 2005 amending Annexes III and X to Regulation (EC) No 999/2001 of the European Parliament and of the Council as regards epidemiological surveillance for transmissible spongiform encephalopathies in bovine, ovine and caprine animals. OJ L 10, 13.1.2005, p. 9–17.



AM), one shift from 24 to 48 months was enforced in 2009. A table summarising the evolution of the changes was published in the 2015 EU Summary Report.<sup>6</sup>

However, there are still some differences in the application of these general rules owing to specific national rules that provide some residual testing of HS or the testing of younger at-risk animals. These national rules, as applied in 2016, are shown in Table 2.

**Table 1:** Criteria for BSE surveillance in bovine animals as applied in 2016 by country, age limit and surveillance target group, based on the TSE Regulation (EC) as amended and Commission Implementing Decision 2013/76/EU of 4 February 2013

	EU 26	Romania, Bulgaria <sup>(a)</sup>
Animals with clinical signs at <i>ante-mortem</i> (AM); Emergency slaughtered animals (ES); Fallen stock (FS)	> 48 months	> 24 months
Healthy slaughtered animals (HS)	No mandatory testing required	> 30 months
Animal culled under BSE eradication measures (EM) BSE suspects (SU)	All	All

BSE: bovine spongiform encephalopathy.

(a): Different criteria were applied in 2016 because Bulgaria and Romania were not in the list of the 26 Member States authorised at that time to revise their BSE annual surveillance programmes according to Commission Implementing Decision 2013/76/EU of 4 February 2013.

<sup>6</sup> European Food Safety Authority (EFSA), Boelaert F, Hugas M, Ortiz Pelaez A, Rizzi V, Stella P and Van der Stede Y, 2016. The European Union summary report on data of the surveillance of ruminants for the presence of transmissible spongiform encephalopathies (TSEs) in 2015. EFSA Journal 2016;14(12):4643, 62 pp. <https://doi.org/10.2903/j.efsa.2016.4643>

**Table 2:** Age limits (in months) of bovine animals tested for BSE surveillance applied in 2016 by Member States and Norway and surveillance target group

Member state	Surveillance target group					
	SU	FS	ES	AM	HS	EM
AT	No age limit	> 48 <sup>(a)</sup>	> 24	> 24	No testing	No age limit
BE	No age limit	> 48	> 48	> 48	No testing	> 24
BG	No age limit	> 24	> 24	> 24	> 30	No age limit
CY	No age limit	> 48	> 48	> 48	No testing	> 48
CZ	No age limit	> 24	> 24	> 24	No testing	No age limit
DE	No age limit	> 48	> 48	> 48	No testing	No age limit
DK	No age limit	> 48	> 48	> 48	No testing	> 48
EE	No age limit	> 48	> 48	> 48	No testing	No age limit
EL	No age limit	> 48	> 48	> 48	> 72	No age limit
ES	No age limit	> 48	> 48	> 48	Born before 2001 and coming from flocks with BSE positive cases	No age limit
FI	No age limit	> 48	> 48	> 48	No testing	No age limit
FR	No age limit	> 24	> 24	> 24	Born before 1/1/2002	> 24
HR	No age limit	> 48 <sup>(b)</sup>	> 48	> 48	No testing	No age limit
HU	No age limit	> 24	> 24	> 24	No testing	No age limit
IE	No age limit	> 48	> 48	> 48	No testing	> 48
IT	No age limit	> 48	> 48	> 48	No testing	No age limit
LT	No age limit	> 48	> 48	> 48	No testing	No age limit
LU	No age limit	> 48	> 48	> 48	No testing	> 48
LV	No age limit	> 24	> 24	> 24	No testing	No age limit
MT	No age limit	> 48	> 48	> 48	No testing	No age limit
NL	No age limit	> 48	> 48	> 48	No testing	No age limit
PL	No age limit	> 48	> 48	> 48	> 108	No age limit
PT	No age limit	> 48	> 48	> 48	No testing	No age limit
RO	No age limit	> 24	> 24	> 24	> 30	No age limit
SE	No age limit	> 48	> 48	> 48	No testing	No age limit
SI	No age limit	> 24	> 24	> 24	No testing	No age limit
SK	No age limit	> 24	> 24	> 24	No testing	No age limit
UK	No age limit	> 48	> 48	> 48	No testing	No age limit
NO	No age limit	> 48	> 48	> 48	No testing	No age limit

SU: Animals clinically suspected of being infected with BSE; FS: fallen stock; ES: emergency slaughtered; AM: animals with clinical signs at *ante-mortem*; HS: healthy slaughtered; EM: animals culled under BSE eradication measures.

If surveillance target group is FS and animals are born in Romania, Bulgaria or Croatia the age limit is > 24. In particular for Croatia, the age limit of > 24 months for animals from FS was applied until the end of May 2016 as the Commission Implementing Decision (EU) 2016/851 changed this age limit to > 48 months from June 2016 onwards.

(a): Age limits are expressed in months.

(b): The limits have been revised after the publication the Commission Decision 2016/851 of 26 May 2016.

### 1.2.2.1. Testing protocol for BSE surveillance in bovine animals

All samples intended to be examined for the presence of TSE shall in general be collected using the methods and protocols laid down in the latest edition of the *Manual for Diagnostic Tests and Vaccines for Terrestrial Animals* of the World Organisation for Animal Health (OIE).

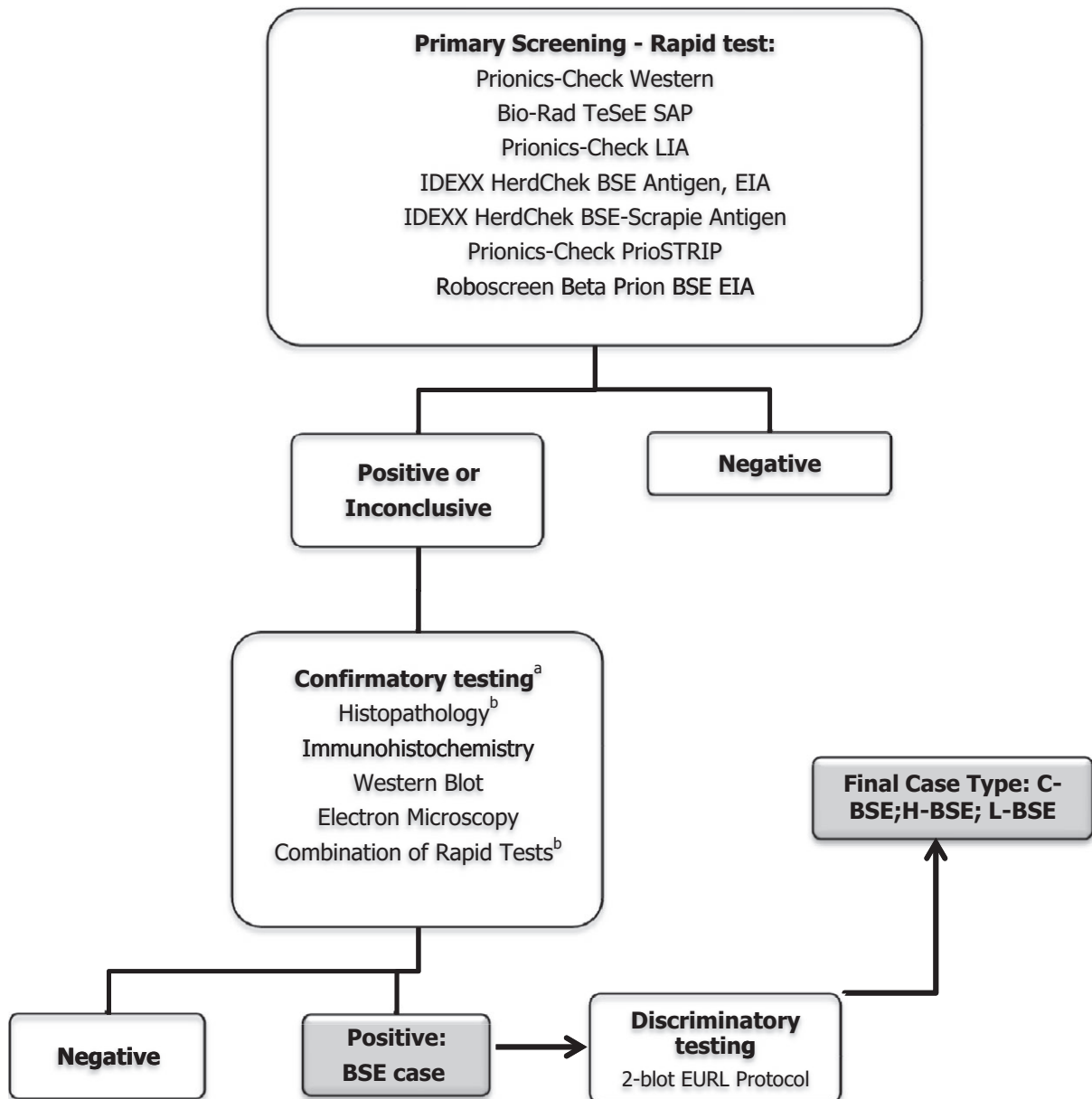
Samples from bovine animals sent for laboratory testing pursuant to the provisions of Annex III, Chapter A, Part I shall be examined with a rapid test. When the result of the rapid test is inconclusive or positive, the sample shall immediately be subjected to confirmatory examinations using at least one of the following methods and protocols laid down in the latest edition of the Manual (Figure 1).

Samples from bovine animals collected from suspect cases should be subjected to confirmatory examinations using at least one of the following methods: immunohistochemistry (IHC), western Blot (WB), demonstration of characteristic scrapie-associated fibrils (SAF) by electron microscopy, or



histopathological examination and a combination of rapid tests. Rapid tests may be used for both primary screening and, if inconclusive or positive, subsequent confirmation according to the guidelines from the European Union TSE Reference Laboratory (EURL).

According to point 3.1(b), Chapter C, Annex X, where the result of the primary screening is not confirmed by the subsequent rapid test, the sample must be subjected to an examination using one of the other confirmatory methods. Where histopathological examination is used for that purpose but proves to be inconclusive or negative, the tissues must be submitted for further examination using one of the other confirmatory methods and protocols.



(a): Animals clinically suspected of being infected by BSE should be immediately subjected to confirmatory examinations.

(b): If histopathology is negative or inconclusive or if there is a discrepancy between rapid tests, a new examination with another confirmatory method is needed.

**Figure 1:** TSE testing protocol for bovine animals

Samples from all positive BSE cases should be forwarded to a laboratory for discriminatory testing of confirmed BSE cases, for the classification of bovine TSE isolates in C-BSE, L-BSE and H-BSE; according to point 3.1(a)(c), Chapter C, Annex X of the TSE Regulation, as amended by Commission regulation 630/2013<sup>7</sup>.

### 1.2.3. TSE<sup>8</sup> surveillance of small ruminants

The surveillance of ovine and caprine animals for the presence of TSE is performed based on testing samples obtained from the following surveillance target groups:

- **Animals clinically suspected of being infected by TSE (SU):** ovine and caprine live, slaughtered or dead animals that show or have shown neurological or behavioural disorders or a progressive deterioration of their general condition linked to impairment of the central nervous system and for which the information gathered on the basis of a clinical examination, response to treatment, a post-mortem examination or an *ante-* or *post-mortem* laboratory analysis does not allow an alternative diagnosis to be established (Article 3.1(h) of the TSE Regulation) and subject to the measures described in Articles 12 and 13 of the TSE Regulation.
- **Animals culled under TSE eradication measures (EM):** in a TSE-infected flock or herd where BSE cannot be excluded, all animals, embryos and ova identified by the inquiry shall be killed and completely destroyed, without delay. When BSE and atypical scrapie (AS) can be excluded and option 1 (killing and complete destruction or slaughtered for human consumption of all animals) has been applied, animals over 18 months of age that are killed for destruction shall be tested for the presence of TSE based on the selection of a simple random sample, in accordance with the sample size set in Annex III, Chapter A, Section II, point 5. If derogations for killing and destruction are applied, all animals that are over 18 months of age slaughtered for human consumption shall be tested. When option 2 is applied (killing and complete destruction or slaughter for human consumption of the susceptible animals only), after the genotyping of all ovine animals, a simple random sample of the animals over 18 months of age selected for killing and destruction shall be tested for the presence of TSE in accordance with the sample size set in Annex III, Chapter A, Section II, point 5. If derogations for killing and destruction are applied, all animals that are over 18 months of age and slaughtered for human consumption shall be tested.
- **Animals not slaughtered for human consumption (NSHC):** ovine and caprine animals that have died or been killed, but which were not killed in the framework of a disease eradication campaign or slaughtered for human consumption. Minimum sample sizes are set (according Table A of Annex III, Chapter A, Section II, point 3) to take into account the size of the ovine populations in the individual MSs and are intended to provide achievable targets (Table 4).
- In the case of an infected flock or herd where either option 2 or 3 or derogations (points 2.2.2 (c) i) and ii)) have been applied, and during the 2-year intensified TSE testing programme or for a period of 2 years from the date of the detection of the last classical scrapie (CS) case, all animals (except ARR/ARR ovine animals) over the age of 18 months that have died or been killed on the holding but were not killed in the framework of a disease eradication campaign must be tested for the presence of TSE.
- In the case of an infected flock or herd where option 2 is applied but with a delayed killing and destruction or slaughtering for human consumption of susceptible animals (derogation point 2.2.2.(c)(iii)), during the period of delay (between the detection of the CS case and the complete killing and destruction or slaughtering of susceptible animals), all animals (except ARR/ARR ovine animals) over the age of 18 months which have died or been killed on the holding but which were not killed in the framework of a disease eradication campaign must be tested for the presence of TSE. Furthermore, after the killing and destruction or slaughtering of susceptible animals, a 2-year intensified TSE testing programme must be carried out or a programme for 2 years from the date of the detection of the last CS case, all animals (except ARR/ARR ovine animals) over the age of 18 months which have died or been killed on the holding but which were not killed in the framework of a disease eradication campaign must be tested for the presence of TSE.

<sup>7</sup> Commission Regulation (EU) No 630/2013 of 28 June 2013 amending the Annexes to Regulation (EC) No 999/2001 of the European Parliament and of the Council laying down rules for the prevention, control and eradication of certain transmissible spongiform encephalopathies. OJ L 179, 29.6.2013, p. 60–83.

<sup>8</sup> The term TSE surveillance in small ruminants is used since both scrapie and BSE have been detected in small ruminants as part of surveillance; when reporting the cases for TSE in small ruminants, the TSE type is assigned as scrapie.

- **Healthy animals slaughtered for human consumption (SHC):** MSs in which the population of ewes and ewe lambs put to the ram, in the case of sheep, or the population of goats that have already kidded and goats mated exceeds 750,000 animals, shall test a minimum annual sample of 10,000 ovine and/or caprine animals slaughtered for human consumption (point 2(a), Section II, Chapter A, Annex III of the TSE Regulation) (see Table 4). MSs may choose to replace a maximum of 50% of their minimum sample size of ovine and caprine animals slaughtered for human consumption by testing dead ovine or caprine animals over the age of 18 months and a maximum of 10% of their minimum sample size of SHC and NSHC by testing ovine or caprine animals killed in the framework of a disease eradication campaign over the age of 18 months at a ratio of one to one.
- In the case of an infected flock or herd where option 2 or derogations (point 2.2.2(c) i) and ii)) has been applied, and during the intensified TSE testing programme for a period of 2 years or from the date of the detection of the last CS case, animals that were kept in the holding at the time when the TSE case was confirmed and that have been slaughtered for human consumption shall be tested for TSE.
- In the case of an infected flock where option 3 (no mandatory killing and complete destruction of animals) have been applied, and during the intensified TSE testing programme for a period of 2 years from the date of the detection of the last CS case, all of the animals which are over the age of 18 months (except ARR/ARR animals) which have been slaughtered for human consumption shall be tested for the presence of TSE.
- In the case of an infected flock or herd where option 2 is applied but with a delayed killing and destruction or slaughtering for human consumption of susceptible animals (derogation 2.2.2.(c)(iii)), during the period of delay (between the detection of the CS case and the complete killing and destruction or slaughtering of susceptible animals), all animals (except ARR/ARR ovine animals) over the age of 18 months which are slaughtered for human consumption must be tested for the presence of TSE. Furthermore, after the killing and destruction or slaughtering of susceptible animals, a 2-year intensified TSE testing programme must be carried out or a programme for 2 years from the date of the detection of the last CS case, and all animals (except ARR/ARR ovine animals) over the age of 18 months which were kept at the holding at the time when the TSE case was confirmed, and which are slaughtered for human consumption, must be tested for the presence of TSE.

Where the TSE case confirmed in a holding is an AS case, the holding shall be subject to the following intensified TSE surveillance protocol for a period of 2 years from the date of detection of the last AS case: all ovine and caprine animals that are over the age of 18 months and slaughtered for human consumption, and all ovine and caprine animals over the age of 18 months that have died or been killed on the holding shall be tested for the presence of TSE. If a case of TSE, other than AS, is confirmed during the intensified TSE monitoring period of two years, the holding shall be subjected to measures laid down in points 2.2.1 and 2.2.2 of Annex VII, Chapter B, Part II.

The above classification of surveillance target groups has been used to outline the data in this report and to summarise historical data, as shown in Table 3.

**Table 3:** Target surveillance groups in small ruminants to be reported for surveillance for TSE based on the infection status of flock/herd/holding, the case type detected and the control measures taken according the TSE regulation

Reported flock status	Index case	Case type	Control measures taken	Sampled population	Surveillance target group to be reported
Non-infected flock <sup>(a)</sup>	Yes	CS or AS	n/a	Slaughtered for human consumption. Annex III, Chapter A, Part II, point 2	<b>SHC</b>
				Not slaughtered for human consumption. Annex III, Chapter A, Part II, point 3	<b>NSHC</b>
				TSE suspects	<b>SU</b>

Reported flock status	Index case	Case type	Control measures taken	Sampled population	Surveillance target group to be reported
TSE-infected flock under official control at sampling <sup>(b)</sup>	No	CS	Killing and complete destruction of all animals (option 1) Annex VII, Chapter B, point 2.2.2 (b) or killing and complete destruction of the susceptible animals only (option 2) Annex VII, Chapter B, point 2.2.2 (c)	Culled and destroyed under options 1 or 2	<b>EM</b>
				Slaughtered for human consumption after application of option 1 or option 2 (c)	<b>SHC</b>
				TSE clinical suspects Chapter 4, Article 12, points 1–2	<b>SU</b>
TSE-infected flock under official control at sampling <sup>(b)</sup>	No	CS	Follow-up after implementation of control measures according to Annex VII, point 2. Intensified TSE monitoring protocol (Annex VII, point 3) after option 1 or option 2, or in case of derogation of option 2 was established, after complete destruction or slaughtering for human consumption of identified animals	Slaughtered for human consumption point 3.1.(a)	<b>SHC</b>
				Not Slaughtered for human consumption point 3.1.(b)	<b>NSHC</b>
				TSE clinical suspects Chapter 4, Article 12, points 1–2	<b>SU</b>
TSE-infected flock under official control at sampling <sup>(b)</sup>	No	CS	Follow-up after implementation of control measures according to Annex VII, point 2 Intensified TSE monitoring protocol (Annex VII, point 4) after option 3	Slaughtered for human consumption point 4.1.(a)	<b>SHC</b>
				Not Slaughtered for human consumption point 4.1. (b)	<b>NSHC</b>
				TSE clinical suspects Chapter 4, Article 12, points 1–2	<b>SU</b>
TSE-infected flock under official control at sampling <sup>(b)</sup>	No	CS	Intensified TSE monitoring protocol pending the implementation of control measures according to the derogation in point 2.2.2.(c)(iii) and after the implementation of the control measures	Slaughtered for human consumption. Point 4.1.(a) and 3.1.(a)	<b>SHC</b>
				Not Slaughtered for human consumption. Point 4.1.(b) and 3.1.(a)	<b>NSHC</b>
				TSE clinical suspects. Chapter 4, Article 12, points 1–2	<b>SU</b>
TSE-infected flock under official control at sampling <sup>(b)</sup>	No	AS	Intensified TSE monitoring protocol after the detection of an Atypical Scrapie case (Annex VII point 2.2.3)	Slaughtered for human consumption point 2.2.3	<b>SHC</b>
				Not slaughtered for human consumption point 2.2.3	<b>NSHC</b>
				TSE clinical suspects Chapter 4, Article 12, points 1–2	<b>SU</b>

Option 2 can only be applied to sheep (genotyping and culling).

EM: Animals culled under TSE eradication measures; NSHC: Animals not slaughtered for human consumption; SHC: animals slaughtered for human consumption SU: Animals clinically suspected of being infected by TSE.

(a): Sheep flocks or goat herds that are not under control measures or intensified TSE protocols or a sheep flock or goat herd that has never been infected with scrapie and for which every new detected case will be an index case.

(b): Sheep flocks or goat herds that are under control measures or intensified TSE protocols or a sheep flock or goat herd that has been infected during reporting year.

**Table 4:** Minimum testing requirements for the TSE surveillance in small ruminants by Member State in 2016

Member State	Sheep			Goats		
	Population size <sup>(a)</sup>	Surveillance target group		Population size <sup>(a)</sup>	Surveillance target group	
		SHC	NSHC		SHC	NSHC
AT	100–750	0	1,500	40–250	0	100% up to 500
BE	100–750	0	1,500	40–250	0	100% up to 500
BG	> 750	10,000	10,000	40–250	0	100% up to 500
CY	100–750	0	1,500	40–250	0	100% up to 500
CZ	100–750	0	1,500	< 40	0	100% up to 100
DE	> 750	10,000	10,000	40–250	0	100% up to 500
DK	40–100	0	100% up to 500	< 40	0	100% up to 100
EE	40–100	0	100% up to 500	< 40	0	100% up to 100
EL	> 750	10,000	10,000	> 750	10,000	10,000
ES	> 750	10,000	10,000	> 750	10,000	10,000
FI	40–100	0	100% up to 500	< 40	0	100% up to 100
FR	> 750	10,000	10,000	> 750	10,000	10,000
HR	100–750	0	1,500	40–250	0	100% up to 500
HU	> 750	10,000	10,000	< 40	0	100% up to 100
IE	> 750	10,000	10,000	< 40	0	100% up to 100
IT	> 750	10,000	10,000	> 750	10,000	10,000
LT	40–100	0	100% up to 500	< 40	0	100% up to 100
LU	< 40	0	100% up to 100	< 40	0	100% up to 100
LV	< 40	0	100% up to 100	< 40	0	100% up to 100
MT	< 40	0	100% up to 100	< 40	0	100% up to 100
NL	100–750	0	1,500	250–750	0	1,500
PL	100–750	0	1,500	< 40	0	100% up to 100
PT	> 750	10,000	10,000	250–750	0	1,500
RO	> 750	10,000	10,000	> 750	10,000	10,000
SE	100–750	0	1,500	< 40	0	100% up to 100
SI	40–100	0	100% up to 500	< 40	0	100% up to 100
SK	100–750	0	1,500	< 40	0	100% up to 100
UK	> 750	10,000	10,000	40–250	0	100% up to 500
NO	> 750	10,000	10,000	40–250	0	100% up to 500

NSHC: Animals not slaughtered for human consumption; SHC: animals slaughtered for human consumption.

Norway tested according a population (thousand head) of > 750 and 40–250 in sheep and goats, respectively.

(a): Thousand head.

### 1.2.3.1. Testing protocols for TSE surveillance in small ruminants

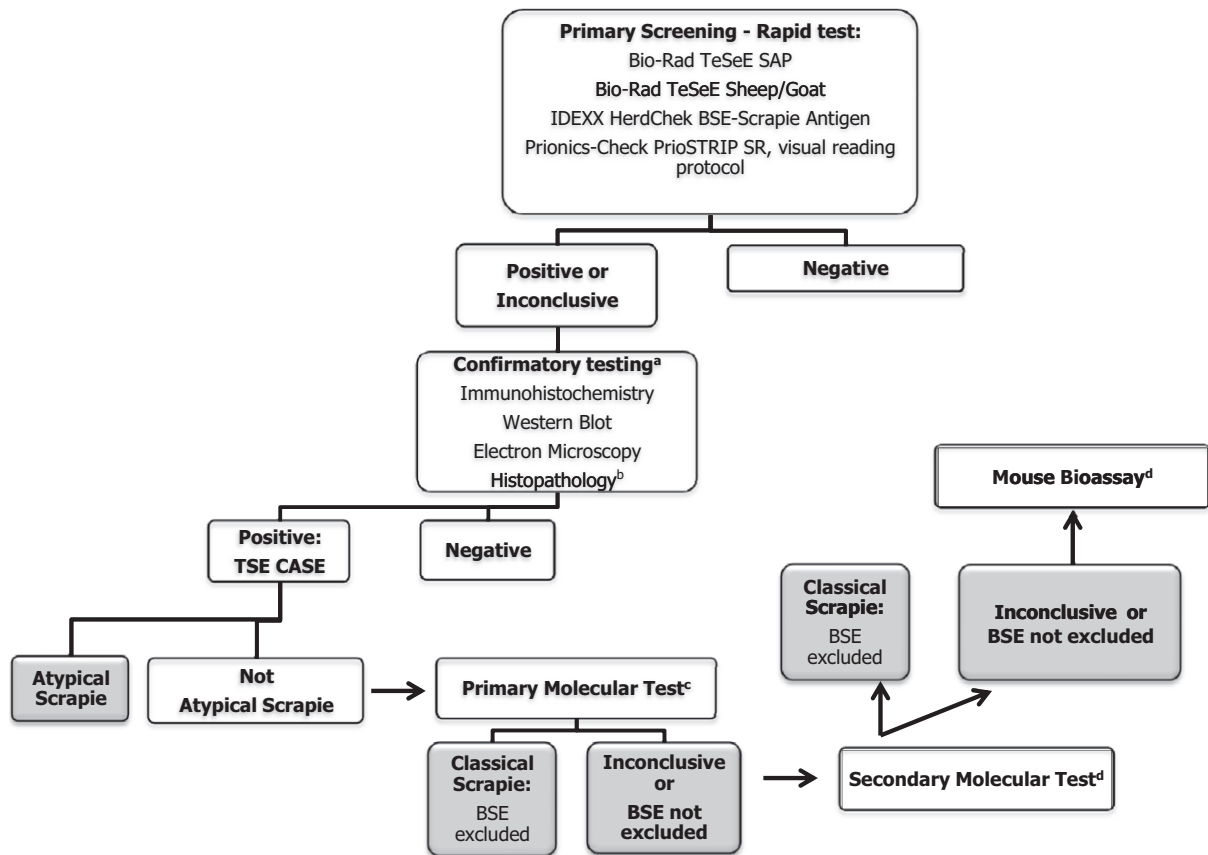
Samples from ovine and caprine animals sent for laboratory testing pursuant to the provisions of Annex III, Chapter A, Part II (Monitoring in ovine and caprine animals) shall be examined with a rapid test to ensure the detection of all known strains of TSE. When the result of the rapid test is inconclusive or positive, the sampled tissues shall immediately be sent to an official laboratory for confirmatory examinations by histopathology, IHC, WB or characterisation of SAF by electron microscopy, as referred to in point (a) of Annex X, Chapter C, Point 3.2. If the result of the confirmatory examination is negative or inconclusive, the tissues shall be submitted for further examination by IHC or WB. If the result of one of the confirmatory examinations is positive, the animal shall be regarded as a positive TSE case and further examination as referred to in point (c) of Annex X, Chapter C, Point 3.2 shall be performed (Figure 2).

Samples from ovine or caprine animals collected from suspect cases should immediately be subjected to confirmatory examinations using at least one of the following methods: IHC, WB, demonstration of characteristic SAF by electron microscopy or histopathological examination. Samples

from routine monitoring should be examined using a rapid test to ensure the detection of all known strains of TSE.

According to point 3.2(b), Chapter C, Annex X, when the result of the rapid test is inconclusive or positive, the sampled tissues should be immediately sent to an official laboratory for confirmatory examinations by histopathology, IHC, WB or SAF. If the result of the confirmatory examination is negative or inconclusive, the tissues should be submitted for further examination by IHC or WB.

Samples from clinical suspect cases and from animals tested in accordance with Annex III, Chapter A, Part II, points 2 and 3 which are regarded as positive TSE cases but which are not AS cases or that display characteristics deemed by the testing laboratory to merit investigation, shall be examined using a discriminatory WB method listed in the guidelines of the TSE EURL by an official diagnostic laboratory designated by the competent authority (point 3.2(c), Chapter C, Annex X). TSE cases in which the presence of BSE cannot be excluded by the primary molecular testing should be submitted for further investigation and confirmation using at least one alternative method differing immunochemically from the original primary molecular method.



(a): Suspect cases will be immediately subjected to confirmatory examinations.

(b): If histopathology is negative or inconclusive, further examination using another confirmatory method is needed.

(c): Primary molecular test using discriminatory WB assays performed in National Reference Laboratories (NRL) or the TSE EURL.

(d): Secondary molecular testing (repeat WB assays, discriminatory enzyme-linked immunosorbent assay (ELISA) and IHC) and possibly a mouse bioassay must be conducted in the EURL.

**Figure 2:** Testing protocol for small ruminants

### 1.2.3.2. Genotyping in sheep

The prion protein genotype for codons 136,154 and 171 shall be determined for each positive TSE case in sheep (point 8.1, Section II, Chapter A, Annex III). In addition, the prion protein genotype for the codons 136, 141, 154 and 171 of a minimum sample of ovine animals should be determined. In the case of MSs with an adult sheep population of > 750,000 animals, this minimum sample should be at least 600 animals. In the case of other MSs, the minimum sample should be at least 100 animals.



The samples may be chosen from animals slaughtered for human consumption, from dead-on-farm animals or from live animals. The sampling should be representative of the entire ovine population (point 8.2, Section II, Chapter A, Annex III).

#### 1.2.4. TSE surveillance in other species (CWD surveillance of cervids)

MSs are obliged according to Article 6(4) of the TSE Regulation to report the number of samples tested and the number of confirmed TSE cases for species other than bovine, ovine and caprine animals. For the reporting on chronic wasting Disease (CWD) in cervids, a new electronic data submission form was prepared in 2016. Separate summary data for surveillance of CWD in cervids is presented in this report.

## 2. Data and methods

### 2.1. Origin of the data

The raw data is electronically submitted by MSs and non-MSs. The data that must be submitted consist of testing data and case-based data for bovine animals and small ruminants according to the reporting periods (monthly basis) as described in Chapter B.I of Annex III.

Electronically submitted data are stored in the EU database and can be consulted using business intelligence tools (Business Objects). The electronically submitted data were extracted from the EU database and further processed and validated by the European Food Safety Authority (EFSA) to summarise the information and to elaborate the summary tables presented in the current European summary report (EUSR).

The remaining data (e.g. genotype data, bovine animals tested according to the required age classes, surveillance of TSE in animals other than bovine animals, ovine and caprine animals as well as CWD in cervids) were provided by the MSs in their annual reports submitted in accordance with Article 6.4 of, and as specified in Chapter B.I, Annex III to, the TSE Regulation. Genotype data for positive scrapie ovine cases and for national representative sampling were retrieved from annual reports submitted by the MSs. Data from both the cases and the sample of genotyped sheep were provided as described in the TSE Regulation Chapter A, Part II, point 8.1 or point 8.2.

Finally, information on the population of small ruminants in 2016 (as presented in Table 4) and the number of BSE cases worldwide (as presented in Table 7) was obtained from the last available report on the monitoring and testing of ruminants for the presence of TSE in the EU by the EC (European Union, 2016) and OIE (<http://www.oie.int/wahis>), respectively.

The data in this report refer only to the samples collected and confirmed cases reported between 1 January 2016 and 31 December 2016 in the EU and three additional non-MS European Free Trade Association (EFTA) reporting countries (Iceland, Norway and Switzerland). EFSA validated the 2016 data by checking for inconsistencies between the data in the annual reports and the electronically extracted data, and by comparing the reported data with previous years. Data providers in the reporting countries were consulted during this validation. The data validation was started in April 2017 and finalised on 20 June 2017. The results and tables presented in the current EUSR are based on the data retrieved from the EU database on 1 July 2017. An additional consultation period with MSs was conducted between 27 September 2017 and 13 October 2017. If historical data (Tables 7 to 9 for BSE, Tables 15–18 for Scrapie) were corrected by the MSs during consultation, EFSA made these changes directly in the tables or made this clear by means of footnotes. EFSA asked the MS to update these changes in the EU database.

For some tables and figures, historical data (period 2001–2016) were extracted from the EU TSE database. As certain MSs and non-MSs may calculate their annual statistics using different reporting criteria (e.g. based on the date of final test results rather than the date of sampling), the data summarised in this report may differ slightly from the national figures published by single MSs for 2016. In addition, subsequent submissions of updated/amended data by MSs may have resulted in differences in the figures included in this report when compared with the same data presented in previous EU summary reports.

### 2.2. Presentation of the data

The current report should be considered the EC summary report for 2016 in compliance with Section II, Chapter B, Annex III of the TSE Regulation.

The 28 EU Member States are referred to as MSs or EU28, and non-Member States as non-MSs when they belong to EFTA reporting countries. The non-MSs in this report are Iceland, Norway and Switzerland. The data reported by Switzerland include those of Lichtenstein. The countries are quoted in this report by using the country codes from the Nomenclature of Units for Territorial Statistics (NUTS) or the English name according to Regulation (EC) No 1059/2003<sup>9</sup>.

For some tables and figures, the surveillance target groups were combined: FS, ES and AM in bovine animals have been included in the group 'risk animals'. The word 'risk' is used hereinafter to indicate those animals whose probability of being detected with the disease is higher than in another surveillance target group (e.g. HS) but did not experience a higher level of exposure than normal (Doherr et al., 2001). The same holds for small ruminants from the NSHC target group (Bird, 2003) when tested from non-infected flocks.

## 2.3. Methods

### 2.3.1. Descriptive methods

To describe the results of the TSE surveillance programme in the EU in 2016, a number of figures and tables have been produced along with a short narrative text to describe the main findings. The report is split into three sections: bovine animals (cattle), small ruminants (sheep and goats) and species other than bovine, ovine and caprine animals. Both aggregated data and data at the national level are presented. Where it was considered relevant, multi-year and historical data are shown: surveillance data were available for the period 2001–2016 for bovine animals, for 2002–2016 for small ruminants, and only for the reporting year (2016) for other species.

In the case of bovine animals, summary statistics were obtained based on the total number of tests performed in 2016 by MS and surveillance target group. In addition, historical data relating to the number of tested animals and confirmed cases since 2001 are summarised. Additional epidemiological parameters have been estimated: for example, absolute number of cases or proportions (cases per million tests) by case type (e.g. C-BSE, H-BSE, L-BSE) and/or surveillance target group, and proportions (cases per million tests) by age-class and by year, among others. These have been used to describe the evolution of the BSE epidemic and to put into context the findings of the reporting year.

A comparison between the average number of cases detected per 1,000,000 tests at the EU level in risk animals and HS target group has been carried out by means of a t-test performed on data from the last 10 years (period 2007–2016). Moreover, the same data have been used to check if any significant temporal trend was detectable. For this purpose, a Poisson regression model has been fitted for each BSE type (C-BSE, H-BSE and L-BSE) separately, using the number of cases as dependent variable and the year as a continuous independent variable; the number of tests was used as offset. The target group (risk animals vs. HS) was added to the model as covariate to adjust for any potential confounding effect. A p-value  $\leq 0.05$  was considered statistically significant for both the statistical analyses.

In order to obtain relevant epidemiological information about the BSE cases detected in 2016, EFSA requested additional information from the individual MSs concerned via a small questionnaire.

For small ruminants, summary statistics are presented in this report, and where possible, stratified according to the relevant variables in the database such as: surveillance target group (SHC, NSHC, SU, EM), flock status (infected, non-infected, other, unknown), surveillance type (passive surveillance restricted to SU vs active surveillance restricted to SHC and NSHC in non-infected flocks), country, year (since 2002), case type (CS or AS), index case (yes/no).

Based on the minimum testing requirements for TSE surveillance in small ruminants (Table 4), a check has been carried out of the compliance of each MS. For assessing compliance, the following criteria have been applied:

- For testing in the NSHC surveillance target group: if the difference between observed testing and expected testing (minimum requirements) was positive, then the MS is compliant with the testing requirements.
- For testing in the SHC surveillance target group: if the difference between observed testing and expected testing (minimum requirements) was positive, the MS is compliant; when the difference was negative, a further calculation was performed to check if the MS's compliance had been achieved by applying the derogation provided by the TSE Regulation (According to

<sup>9</sup> Regulation (EC) No 1059/2003 of the European Parliament and of the Council of 26 May 2003 on the establishment of a common classification of territorial units for statistics (NUTS). OJ L 154, 21.6.2003, p. 1–41.

point II.2(c), Chapter A, Annex III of the TSE Regulation) i.e. replacing up to 50% of its minimum SHC sample size by testing dead ovine or caprine animals over the age of 18 months at the ratio of one to one and in addition to the minimum sample size for NSHC.

A MS has been considered to meet the minimum requirements when the above criteria have been met in both target groups.

Also in small ruminants, a comparison between the average number of cases per 10,000 tests detected at the EU level in NSHC and SHC surveillance groups has been carried out for animals tested on non-infected flocks: here, a t-test was used on data from the last 10 years (period 2007–2016). Moreover, the same data have been used to check if any significant temporal trend was detectable: a Poisson regression model has been fitted for ovine and caprine animals separately, using the number of cases as dependent variable, the year as a continuous independent variable; the number of tests was used as offset. The target group was added to the model as covariate to adjust for any potential confounding effect. A p-value  $\leq 0.05$  was considered statistically significant for both the statistical analyses.

To visualise a potential evolution (period 2002–2016) in the number of CS cases over the number of AS cases in sheep, an annual ratio (CS:AS) was plotted for each MS. A correction was applied for those MSs for which the numerator or denominator was equal to zero. In those cases, the ratio was replaced by a value calculated using the following formula:  $(CS + 0.5) / (AS + 0.5)$ . No calculation was carried out for years in which neither CS nor AS cases were detected. Finally, a log scale was used for the Y-axis to include all surveillance target groups in the same figure. The CS/AS plot can be interpreted by assuming that the occurrence of AS is homogeneously distributed in the EU and is not influenced by breeding programmes or the control measures. After a visual inspection of the national graphs, data from MSs with similar patterns have been combined to build separate graphs showing three main clusters: (1) a CS:AS ratio that is always below one; (2) a decreasing CS/AS ratio over time (crossing the ratio one at some point); and (3) a CS/AS ratio that is always greater than one (more CS than AS cases have been consistently detected).

The mean age of the AS cases has been compared with that of CS cases in sheep and goats by applying a two-sample t-test with unequal variances. A p-value  $\leq 0.05$  was considered statistically significant.

Finally, the classification originally developed by Great Britain's National Scrapie Plan (NSP) was used to summarise and describe the data on genotyping. Moreover, in order to identify the evolution of the effect of breeding against resistance in some MSs, the five NSP groups were recategorised into three groups, namely, resistant (homozygous ARR), semi-resistant (heterozygous ARR except ARR/VRQ) and susceptible (non-ARR except ARR/VRQ).

In order to describe and plot the reported data, some assumptions were made for reporting the results in bovine animals and small ruminants (sheep and goats):

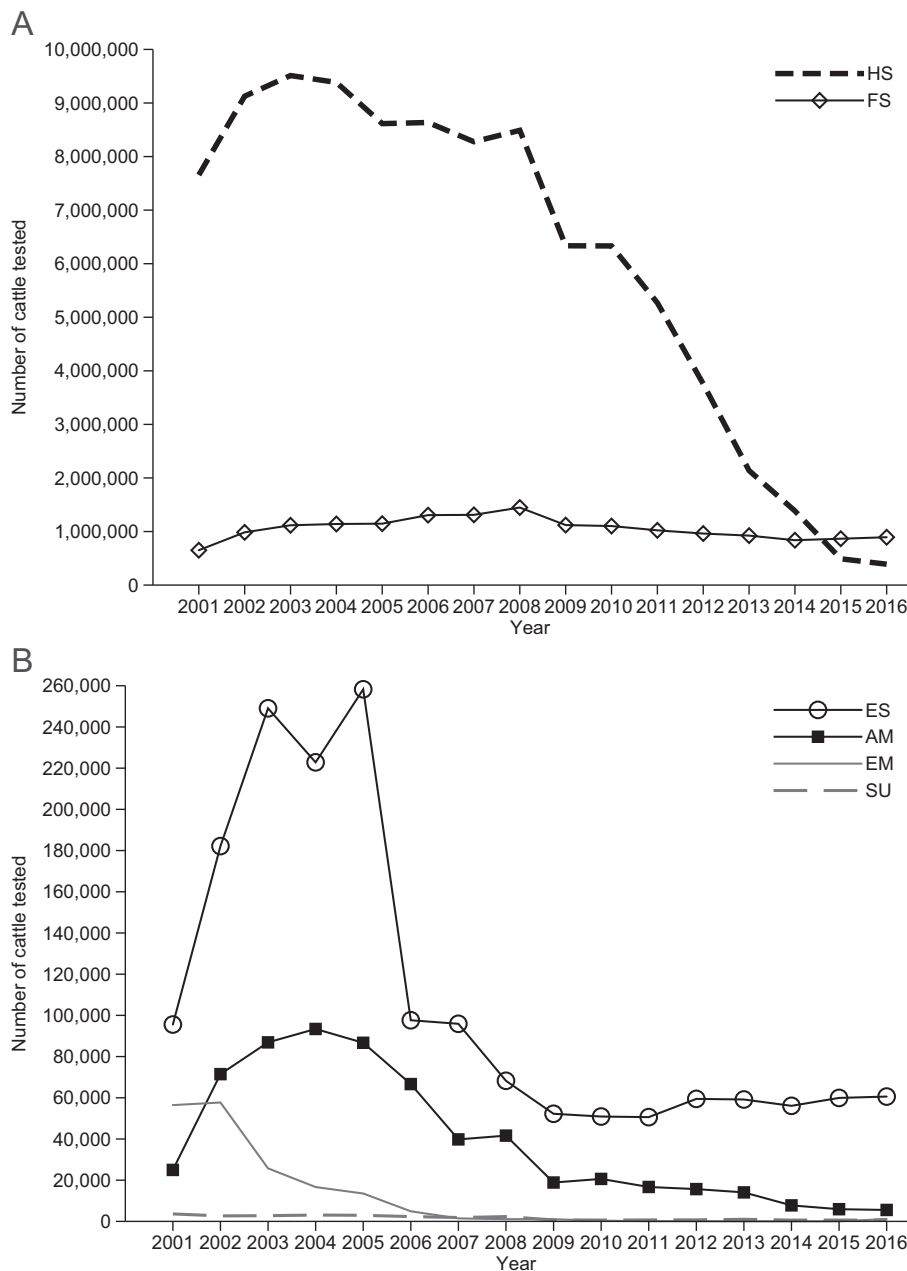
- With regards to bovine animals, to plot the temporal evolution of BSE cases (C-BSE, L-BSE and H-BSE), cases for which the type was reported as 'Unknown' or was missing were considered for reporting purposes as C-BSE, since most of these were reported before 2005.
- In small ruminants, the following assumptions were considered:
  - To plot the reported scrapie cases according to the flock status, it was assumed that flocks with status reported as 'unknown', 'other' or blank were considered for reporting purposes as 'non-infected flocks'.
  - To describe the evolution of the total number of scrapie index cases, it was assumed that all index cases ('yes') were confirmed in non-infected flocks. If an index case was reported as 'no' or 'unknown', it was considered for reporting purposes as 'infected flocks'.
  - To describe the results of the discriminatory TSE testing, it was assumed that all scrapie cases with 'BSE-like', 'non BSE-like' or 'inconclusive' results in the primary or secondary molecular tests have been submitted for discriminatory testing.

### 3. Assessment

#### 3.1. BSE surveillance in bovine animals

Circa 115 million bovine animals were tested for BSE between 2001 and 2016. As shown in Figure 3, the number of bovine samples tested for BSE decreased over the years. There was a 6% reduction in the total number of samples tested in the EU from 1,442,446 in 2015 to 1,352,585 in 2016. The target surveillance group most affected by this reduction was the HS, with a 20% reduction from 491,052 in 2015 to 390,586 in 2016 (mainly due to reductions in Germany, France and Poland),

as shown in Table 5. Compared with 2015, the number of samples tested from risk animals (AM + ES + FS) over 48 months of age showed an increase (+ 28,118), mainly attributable to a few countries (France, Ireland and the United Kingdom). Currently, over 71% of all samples tested in the 31 reporting countries is obtained solely from the FS surveillance target group.



The figure does not include: data from Norway, Switzerland and Iceland reporting ca. 270,000 tests between 2003 and 2016; 1,500 animals tested in 2003 and submitted for rapid testing without a clear indication of the target group. SU: Animals clinically suspected of being infected with BSE; FS: fallen stock; ES: emergency slaughtered; AM: animals with clinical signs at *ante-mortem*; HS: healthy slaughtered; EM: animals culled under BSE eradication measures.

**Figure 3:** Total number of bovine animals tested for BSE in the EU over the period 2001–2016 by surveillance target group. (A) surveillance target groups HS and FS; (B) surveillance target groups ES, AM, EM and SU

The number of cattle tested for BSE per reporting country for each surveillance target group in 2016 is shown in Table 5. The number of tested bovine animals per MS and per age category for each of the target surveillance groups is summarised in Appendix A (Tables A.1–A.5).

**Table 5:** Number of bovine animals tested for BSE by reporting country and surveillance target group in 2016 in the EU and other reporting countries

Country	Surveillance target group							Total
	Risk animals				Other			
	ES	AM	FS	Subtotal (ES + AM + FS)	HS	SU	EM	
AT	3,011	77	13,456	16,544	3,355	15	0	19,914
BE	810	2	24,955	25,767	42	27	0	25,836
BG	1,788	0	524	2,312	18,546	0	0	20,858
CY	0	0	916	916	0	0	0	916
CZ	909	0	14,569	15,478	36	2	0	15,516
DE	9,090	1	151,297	160,388	536	461	0	161,385
DK	1,307	0	19,367	20,674	21	1	0	20,696
EE	80	28	3,465	3,573	0	0	0	3,573
EL	26	28	1,701	1,755	9,253	6	1,091	12,105
ES	242	30	62,231	62,503	302	3	11	62,819
FI	39	1	11,190	11,230	4	0	0	11,234
FR	11,981	0	202,519	214,500	41,594	5	61	256,160
HR	402	0	5,881	6,283	12,327	1	0	18,611
HU	142	627	11,098	11,867	1,705	2	0	13,574
IE	0	706	58,659	59,365	0	0	0	59,365
IT	13,357	623	38,268	52,248	597	0	0	52,845
LT	53	22	3,095	3,170	0	0	0	3,170
LU	0	0	2,408	2,408	0	2	0	2,410
LV	74	212	2,695	2,981	0	3	0	2,984
MT	70	0	104	174	0	1	0	175
NL	5,411	0	49,456	54,867	21	0	0	54,888
PL	4,388	456	32,498	37,342	131,825	20	0	169,187
PT	1,589	1,555	15,931	19,075	603	1	0	19,679
RO	1,792	174	3,113	5,079	169,722	70	0	174,871
SE	190	20	8,750	8,960	1	2	0	8,963
SI	406	84	5,657	6,147	79	13	0	6,239
SK	51	5	7,618	7,674	0	0	0	7,674
UK	3,391	918	142,590	146,899	17	1	21	146,938
<b>Total EU</b>	<b>60,599</b>	<b>5,569</b>	<b>894,011</b>	<b>960,179</b>	<b>390,586</b>	<b>636</b>	<b>1,184</b>	<b>1,352,585</b>
CH	4,719	0	6,874	11,593	0	35	0	11,628
IS	1	0	12	13	127	0	0	140
NO	5,099	73	1,754	6,926	1	0	0	6,927
<b>Total EFTA</b>	<b>9,819</b>	<b>73</b>	<b>8,640</b>	<b>18,532</b>	<b>128</b>	<b>35</b>	<b>0</b>	<b>18,695</b>
<b>Total</b>	<b>70,418</b>	<b>5,642</b>	<b>902,651</b>	<b>978,711</b>	<b>390,714</b>	<b>671</b>	<b>1,184</b>	<b>1,371,280</b>

SU: Animals clinically suspected of being infected with BSE; FS: fallen stock; ES: emergency slaughtered; AM: animals with clinical signs at *ante-mortem*; HS: healthy slaughtered; EM: animals culled under BSE eradication measures.

In 2016, five BSE cases were reported by two different MSs (France and Spain). All cases were detected in the FS target surveillance group.

One of the four French cases was confirmed C-BSE and had been born in 2011: the animal was 59 months old and this is the first time since 2008 that a case has been detected in the age class < 60 months. It has been classified as a BARB case, i.e. born after 1 January 2001 when the 2001 EU-wide reinforcement of the European Union feed ban was implemented. Recently, an EFSA Opinion (EFSA BIOHAZ Panel, 2017a) has been dedicated to the investigation of the origin of the 60 BARB cases detected in the EU since 2001.

The remaining four cases were all of H-BSE type (three from France and one from Spain) and were detected in old animals (all were over 133 months of age).

Table 6 reports the main characteristics and epidemiological information relating to the positive cases, e.g. the presence of clinical symptoms, herd size, herd type, animal and feed type. This additional information was obtained from a small questionnaire sent in July 2017 to the TSE reporting officers of France and Spain.



**Table 6:** Clinical and epidemiological description of the five BSE cases detected in 2016 by Spain and France

Country	ES – atypical	FR – atypical1	FR – atypical2	FR - atypical3	FR – classical
<b>Surveillance target group</b>	Fallen stock	Fallen stock	Fallen stock	Fallen stock	Fallen stock
<b>Case type</b>	H-BSE	H-BSE	H-BSE	H-BSE	C-BSE
<b>Month and year of birth</b>	March 2000	March 2001	May 2002	December 2004	April 2011
<b>Age at detection (in months)</b>	193	186	165	133	59
<b>BARB status</b>	No	No	No	No	Yes
<b>Clinical symptoms</b>	Sialism	This cow was euthanised after an accident (slip on wet grass). It was found lying on its back and was unable to stand up	None	The pregnant cow had paresis. It was euthanised after a caesarean section resulted in paraplegia	Euthanasia following a fall
<b>Cattle type</b>	Beef	Beef	Beef	Beef	Beef
<b>Breed</b>	Morucha	Charolaise	Limousine	Charolaise	Salers
<b>Was the case confirmed at herd/holding where the animal was born?</b>	Yes	Yes	No	Yes	Yes
<b>Location (NUTS3) of natal herd or herd where case found</b>	Castilla y León Region/Salamanca province	Côte d’Or	Corrèze	Allier	Ardennes
<b>Herd size</b>	127	50	80	270	394
<b>Herd type</b>	Beef production	Beef	Beef	Beef	Beef/mixed (also sheep)
<b>Feeding system during first year of life</b>	Suckle, concentrate, pasture	NA	Unknown, herd closed in 2007	Hay, mash, mineral supplement, reconstituted milk for orphans and twins	Extensive livestock; calf under the mother, pasture, hay; few concentrate and mixed.
<b>Feed cohorts? Tested? If Yes: Results (number tested; number positives)</b>	No	No	Yes (1;0)	Yes (8;0)	Yes (1;0)
<b>Birth cohorts? Tested? If Yes: Results (number tested; number positives)</b>	Yes (10;0)	Yes (1;0)	Yes (1;0)	Yes; (8;0)	Yes (54;0)
<b>Offspring? Tested? If Yes: Results (number tested; number Positives)</b>	Yes (1;0)	Yes (1;0)	Not tested	Yes (NA)	Yes (1;0)
<b>Sire? Tested? (Yes/No). If Yes: Results (positive? negative?)</b>	NA <sup>(a)</sup>	Yes, negative	Not tested	Yes, negative	Not tested
<b>Dam? Tested (Yes/No). If Yes: Results (positive? negative?)</b>	Yes, negative	Yes, negative	NA <sup>(a)</sup>	Yes, negative	Yes, negative

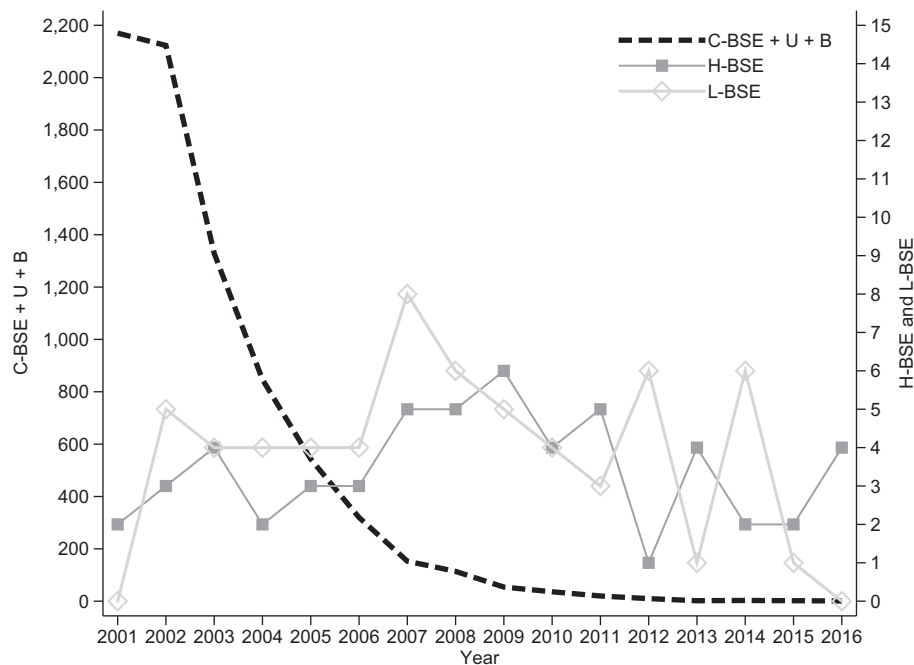
NA: Not available; BARB: Born after the reinforced feed ban.

The total number of BSE cases worldwide by year and country since the 1990 is displayed in Table 7. The occurrence of C-BSE since 2001 (highlighting the occurrence of BARB cases) and of the atypical cases are shown in Tables 8 and 9, respectively. In 2016, no BSE cases were detected outside the EU. The cumulative geographical distribution of C-BSE, H-BSE and L-BSE in the EU is shown in Appendix C.

Comparing C-BSE cases with the two previous years (three cases in 2014 and five in 2015), there was no apparent difference in the proportion of cases per million tests. Figure 5 shows the evolution of the proportion of BSE cases (per 1,000,000 tests) by case type. Over the last 10 years, the average number of C-BSE cases per 1,000,000 in the risk animals is higher ( $p$  value  $< 0.03$ ) than in HS animals (18.7 vs 1.6). The same is true for H-BSE (2.8 vs 0.2,  $p$  value  $< 0.0001$ ) and L-BSE (1.9 vs 0.6,  $p = 0.04$ ).

As described in the previous 2015 TSE EU summary report,<sup>6</sup> C-BSE in the EU has trended downwards over time, with only 5 cases reported in the EU28 in the last 3 years (Figure 4). For the first time since BSE cases have been reported, no C-BSE or atypical BSE cases were reported by the United Kingdom in 2016.

Since 2007, the EU trend in the proportion of C-BSE cases (per 1,000,000 tests), adjusted by target group, has been decreasing from 68.4 in 2007 to 1 in 2016 (Beta =  $-0.4$ ,  $p$  value  $< 0.0001$ ).



Left y-axis: C-BSE + U + B. Right y-axis: L- to H-BSE and L-BSE.

One case from Poland in 2012 that was reported as 'atypical' is not included in the figure.

Cases reported as unknown (U) or blank/missing (B) were combined with C-BSE cases.

The figure does not include data from non-MSs (Norway (1 case), Switzerland (101 cases)) over the same period.

Number of cases of C-BSE + U + B by year of detection: 2010:37; 2011:21; 2012:11; 2013:2; 2014:3; 2015:2; 2016:1.

**Figure 4:** Number of BSE cases by type over the period 2001–2016 in the EU



Country	Year																										Total		
	Up to 1990	1991	1992	1993	1994	1995	1996	1997	1998	1999	2000	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	2012	2013	2014	2015		2016	
<b>UK</b>	24,595	25,359	37,280	35,090	24,438	14,562	8,149	4,393	3,235	2,301	1,443	1,203	1,123	610	330	208 (1H/1L)	129 (1L)	65 (2H/2L)	42 (2L)	11 (1H)	11 (1L)	8 (1H/1L)	3 (1L)	3 (1H)	1	2 (1H)		<b>184,594</b>	
<b>Total EU-28</b>	<b>24,628</b>	<b>25,382</b>	<b>37,301</b>	<b>35,110</b>	<b>24,478</b>	<b>14,596</b>	<b>8,266</b>	<b>4,515</b>	<b>3,471</b>	<b>2,591</b>	<b>1,924</b>	<b>2,176</b>	<b>2,137</b>	<b>1,342</b>	<b>854</b>	<b>547</b>	<b>329</b>	<b>166</b>	<b>126</b>	<b>65</b>	<b>44</b>	<b>28</b>	<b>18</b>	<b>7</b>	<b>11</b>	<b>5</b>	<b>5</b>	<b>190,122</b>	
<b>BRA</b>																							1		1 (1H)			<b>2</b>	
<b>CAN</b>				1 <sup>(a)</sup>										1	1	1	5	3	4	1	1	1					1		<b>20</b>
<b>ISR</b>													1																<b>1</b>
<b>JPN</b>											3	2	4	5	7	10	3	1	1										<b>36</b>
<b>LI</b>									2																				<b>2</b>
<b>NO</b>																										1 (1H)			<b>1</b>
<b>CH</b>	2	8	15	29	64	68	45	38	14	50	33	42	24	21	3	3	5	0	0	0	0	2 (2A)	1 (1H) <sup>(a)</sup>						<b>467</b>
<b>USA</b>														1 <sup>(a)</sup>		1	1									1			<b>4</b>
<b>Total Non EU</b>	<b>2</b>	<b>8</b>	<b>15</b>	<b>30</b>	<b>64</b>	<b>68</b>	<b>45</b>	<b>38</b>	<b>16</b>	<b>50</b>	<b>33</b>	<b>45</b>	<b>27</b>	<b>27</b>	<b>9</b>	<b>12</b>	<b>21</b>	<b>6</b>	<b>5</b>	<b>2</b>	<b>1</b>	<b>3</b>	<b>3</b>	<b>0</b>	<b>1</b>	<b>2</b>	<b>0</b>	<b>533</b>	
<b>Total</b>	<b>24,630</b>	<b>25,390</b>	<b>37,316</b>	<b>35,140</b>	<b>24,542</b>	<b>14,664</b>	<b>8,311</b>	<b>4,553</b>	<b>3,487</b>	<b>2,641</b>	<b>1,957</b>	<b>2,221</b>	<b>2,164</b>	<b>1,369</b>	<b>863</b>	<b>559</b>	<b>350</b>	<b>172</b>	<b>131</b>	<b>67</b>	<b>45</b>	<b>31</b>	<b>21</b>	<b>7</b>	<b>12</b>	<b>7</b>	<b>5</b>	<b>190,655</b>	

BRA: Brazil; CAN: Canada; ISR: Israel; JPN: Japan; USA: The United States of America.

Each cell reports the total number of BSE cases (C-BSE (including cases reported as 'Unknown') + H-BSE + L-BSE). EU countries without BSE cases (Bulgaria, Cyprus, Estonia, Croatia, Hungary, Lithuania, Latvia and Malta) are not reported.

Source: data regarding non-EU cases and cases in EU Member States for the period 1987–2002 were made available by the European Commission (European Union, 2016). Data were retrieved from the EU TSE Database and the OIE website (<http://www.oie.int/wahis>).

(a): Included imported cases: CAN one case in 1993; Denmark one case in 1992; France one case in 1999; Germany one case in 1992, three cases in 1994, two cases in 1997; Ireland 5 cases in 1989, one case in 1990, 2 cases in 1991 and 1992, one case in 1994 and one case in 1995; Italy two cases in 1994 and one case in 2001 and 2002; Portugal one case in 1990, 1991, 1992, 2000 and 2004 and three cases in 1993; Slovenia one case in 2004; Switzerland one case in 2012; USA one case in 2003.

(b): Gavier-Widen et al. (2008).

**Table 8:** Number of reported Classical BSE cases in the EU and CH by year (period 2001–2016) and country

Country	Year																	Total
	Up to 2000	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	2012	2013	2014	2015	2016	
AT		1				2	2											5
BE	19	46	38	15	11	3	1											133
CZ		2	2	4	7	8	3	1		2								29
DE	13 <sup>(b)</sup>	125	105	53	64	32	16	4	2	2								416
DK	2 <sup>(a)</sup>	6	3	2		1				1								15
EL		1																1
ES	2	83	134	172	138	103	75	32	22	14	13	6	3		1			798
FI		1																1
FR	242 <sup>(b)</sup>	276	237	107	50	31	6	5	3	6	2	3					1	969
IE	596 <sup>(b)</sup>	246	332	182	126	69	41	25	23	9	1	2	3			1		1,656
IT	2 <sup>(b)</sup>	50	35	30	7	8	7	1	1	1								142
LU	1		1			1												3
NL	8	18	23	18	6	3	2	2	1		2							83
PL			3	5	9	18	8	7	5	3	2							60
PT	529 <sup>(b)</sup>	110	86	133	91	51	32	12	17	6	5	4	2		1			1,079
SI		1	1	1	2	1	1	1										8
SK		5	6	2	7	3		2	1		1							27
UK	180,845	1,203	1,123	610	330	206	128	61	40	10	10	6	2	2	1	1		184,578
<b>Total EU28</b>	<b>182,259</b>	<b>2,174</b>	<b>2,129</b>	<b>1,334</b>	<b>848</b>	<b>540</b>	<b>322</b>	<b>153</b>	<b>115</b>	<b>54</b>	<b>36</b>	<b>21</b>	<b>10</b>	<b>2</b>	<b>3</b>	<b>2</b>	<b>1</b>	<b>190,003</b>
CH	366	42	24	21	3	3	5											464
Total EFTA	366	42	24	21	3	3	5											464
<b>Total</b>	<b>182,625</b>	<b>2,216</b>	<b>2,153</b>	<b>1,355</b>	<b>851</b>	<b>543</b>	<b>327</b>	<b>153</b>	<b>115</b>	<b>54</b>	<b>36</b>	<b>21</b>	<b>10</b>	<b>2</b>	<b>3</b>	<b>2</b>	<b>1</b>	<b>190,467</b>

Each cell reports the total number of C-BSE cases. EU countries without C-BSE cases (Bulgaria, Cyprus, Estonia, Croatia, Hungary, Lithuania, Latvia, Malta, Romania and Sweden) are not reported.

Grey shaded cells indicate the year and Member State where at least one BARB case was detected (EFSA BIOHAZ Panel, 2017a).

Source: data were retrieved from the EU TSE Database except for CH.

(a): One of these cases was imported from the United Kingdom.

(b): Include imported cases.

**Table 9:** Number of reported BSE Atypical cases in EU and EFTA countries by year (period 2001–2016), type and country

Country code	Year																																Total						
	2001		2002		2003		2004		2005		2006		2007		2008		2009		2010		2011		2012		2013		2014		2015		2016		H	L					
	H	L	H	L	H	L	H	L	H	L	H	L	H	L	H	L	H	L	H	L	H	L	H	L	H	L	H	L	H	L									
AT														1						1	1													1	2				
CZ													1																						1				
DE				1		1	1																				1	1							2	3			
DK							1																													1			
ES					1						1			1	1	2	2			1			1	2				1			1	1		8	8				
FR	1		2	1	3	1		1		1		2	1	1	2	3	2	2	2	1			1	2		1	2				3		19	16					
IE			1																1		1				1										4				
IT				1		1								1					1			1														5			
NL	1			1		1													1																	1	3		
PL				1			2		2	1	1		2			1					1		2 <sup>(a)</sup>		1										2	12			
PT						1		2					1		2						1															7			
RO																												2									2		
SE											1																										1		
SI																																					1		
UK									1	1		1	2	2		2	1			1	1	1		1	1											1	9		
<b>Total EU-28</b>	<b>2</b>	<b>0</b>	<b>3</b>	<b>5</b>	<b>4</b>	<b>4</b>	<b>2</b>	<b>4</b>	<b>3</b>	<b>4</b>	<b>3</b>	<b>4</b>	<b>5</b>	<b>8</b>	<b>5</b>	<b>6</b>	<b>6</b>	<b>5</b>	<b>4</b>	<b>4</b>	<b>4</b>	<b>3</b>	<b>1</b>	<b>6</b>	<b>4</b>	<b>1</b>	<b>2</b>	<b>6</b>	<b>2</b>	<b>1</b>	<b>4</b>	<b>0</b>	<b>54</b>	<b>61</b>					
NO																																						1	
CH																								1														1	
<b>Total EFTA</b>																								<b>1</b>														<b>2</b>	
<b>Total</b>	<b>2</b>	<b>0</b>	<b>3</b>	<b>5</b>	<b>4</b>	<b>4</b>	<b>2</b>	<b>4</b>	<b>3</b>	<b>4</b>	<b>3</b>	<b>4</b>	<b>5</b>	<b>8</b>	<b>5</b>	<b>6</b>	<b>6</b>	<b>5</b>	<b>4</b>	<b>4</b>	<b>4</b>	<b>3</b>	<b>2</b>	<b>6</b>	<b>4</b>	<b>1</b>	<b>2</b>	<b>6</b>	<b>3</b>	<b>1</b>	<b>4</b>	<b>0</b>	<b>56</b>	<b>61</b>					

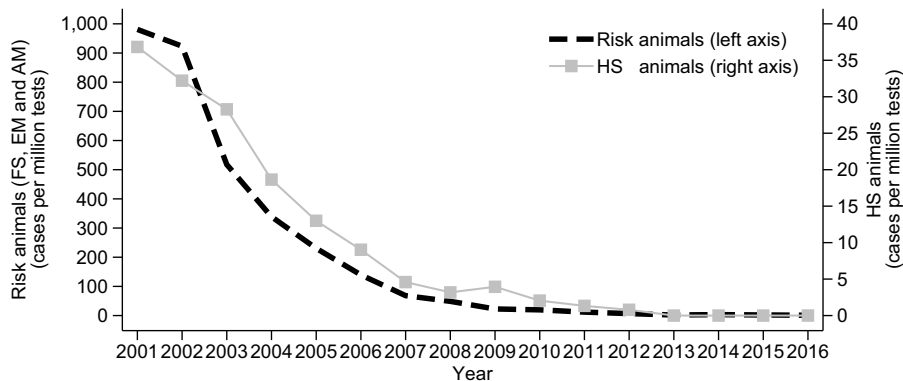
Each cell reports the total number of H-BSE and L-BSE cases. EU countries without atypical cases (Belgium, Greece, Finland, Luxembourg, Slovakia, Iceland) are not included in the table.

Source: data were retrieved from the EU TSE Database and from the OIE for CH.

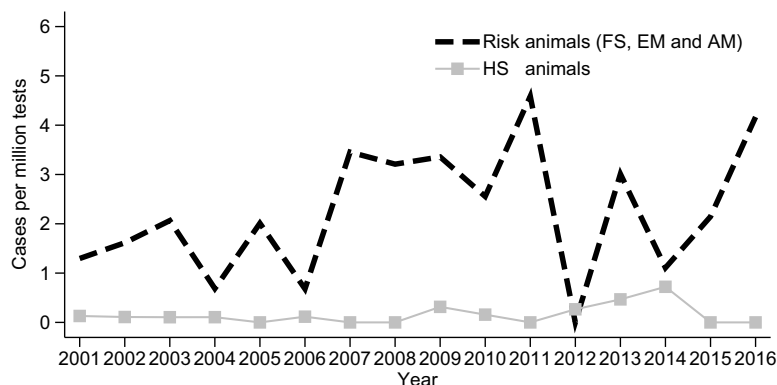
(a): In 2012, PL reported an atypical BSE case without specifying the type.



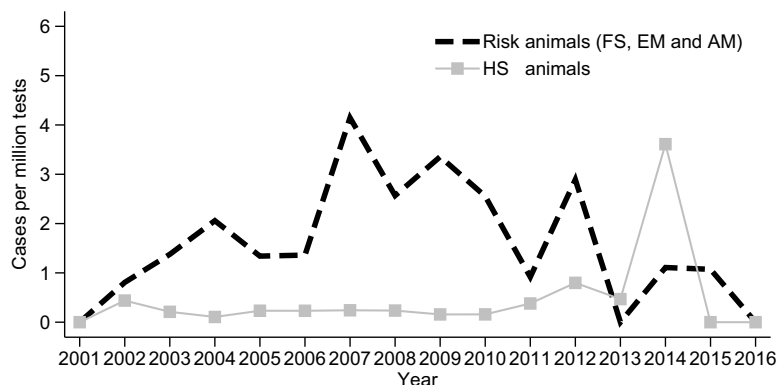
## A: C-BSE + U + B



## B: H-BSE



## C: L-BSE



A: C(Classical)-BSE + U(Unknown) + B(Blank).

**Figure 5:** Cases per million tested bovine animals by surveillance target group and by case type over the period 2001–2016 in the EU

Because of the introduction of mandatory discriminatory testing in 2003 and the retrospective discriminatory testing of BSE cases, 97% of all cases were subjected to confirmatory testing between 2003 and 2016. In total, 621 BSE cases (18%) remain with unknown type status (Table 10).

Based on the results from discriminatory testing and reporting, a total of 106 atypical BSE cases were reported by the 31 reporting countries during the period 2003–2016 (3% of all cases subject to discriminatory testing): 50 H-type and 56 L-Type (excluding one atypical case from Poland in 2012 that could not be defined as H- or L-type).

Focusing on the last 10 years (period 2007–2016), unlike C-BSE, no apparent trend is detectable for the two atypical BSE forms (H-BSE: Beta =  $-0.0003$ , p value = 0.99, L-BSE: Beta =  $-0.04$ , p value = 0.54; Figure 4).

**Table 10:** Number and proportion of BSE cases subject to discriminatory testing, by case type for the period 2003–2016

Country	Total BSE cases	Cases not submitted for discriminatory testing	Cases submitted for discriminatory testing by type							Total cases submitted for discriminatory testing	% of cases submitted for discriminatory testing
			Atypical (case type not defined)	H-BSE	L-BSE	C-BSE	% of BSE cases with known case type	Unknown			
AT	7	0	0	1	2	4	100.0%	0	7	100.0%	
BE	30	0	0	0	0	19	63.3%	11	30	100.0%	
CZ	26	0	0	1	0	21	84.6%	4	26	100.0%	
DE	177	1	0	2	2	167	96.6%	5	176	99.4%	
DK	5	0	0	0	1	2	60.0%	2	5	100.0%	
ES	595	0	0	8	8	533	92.3%	46	595	100.0%	
FR	245	0	0	16	15	201	94.7%	13	245	100.0%	
IE	486	0	0	4	0	144	30.5%	338	486	100.0%	
IT	59	0	0	0	4	55	100.0%	0	59	100.0%	
LU	1	0	0	0	0	1	100.0%	0	1	100.0%	
NL	36	1	0	0	2	33	97.2%	0	35	97.2%	
PL	71	0	1	2	11	57	98.6%	0	71	100.0%	
PT	361	97 <sup>(a)</sup>	0	7	0	249	70.9%	8	264	73.1%	
RO	2	0	0	0	2	0	100.0%	0	2	100.0%	
SE	1	0	0	1	0	0	100.0%	0	1	100.0%	
SI	7	0	0	1	0	6	100.0%	0	7	100.0%	
SK	16	0	0	0	0	15	93.8%	1	16	100.0%	
UK	1,423	0	0	7	9	1,214	86.4%	193	1,423	100.0%	
<b>Total EU</b>	<b>3,548</b>	<b>106</b>	<b>1</b>	<b>50</b>	<b>56</b>	<b>2,767</b>	<b>81.0%</b>	<b>621</b>	<b>3,442</b>	<b>97.0%</b>	
NO	1	0	0	1	0	0	100.0%	0	1	100.0%	
<b>Total EFTA</b>	<b>1</b>	<b>0</b>	<b>0</b>	<b>1</b>	<b>0</b>	<b>0</b>	<b>100.0%</b>	<b>0</b>	<b>1</b>	<b>100.0%</b>	
<b>Total</b>	<b>3,549</b>	<b>106</b>	<b>1</b>	<b>51</b>	<b>56</b>	<b>2,767</b>	<b>81.0%</b>	<b>621</b>	<b>3,443</b>	<b>97.0%</b>	

(a): The discriminatory testing in PT is still ongoing and database needs to be updated.

The table includes only cases for the period 2003–2016 and only for Member States that reported cases. Categories for case type: L-BSE, H-BSE, C-BSE, Unknown and Blank (missing data).

### 3.2. TSE surveillance in small ruminants

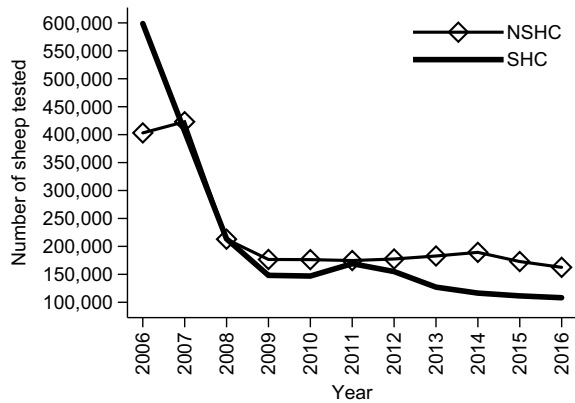
Since 2002, ca 8.8 million small ruminants have been tested for TSE in the EU. After a peak of testing in 2006 – due to changes in the TSE Regulation – the overall number of tested sheep and goats has decreased continuously over time (Figures 6 and 7). In 2016, a total of 286,351 sheep and 110,832 goats were tested in the EU, respectively, 5% and 11% less than in 2015, and, for both species, mainly restricted to NSHC from non-TSE-infected flocks in FR.

The numbers of sheep and goats tested for TSE by reporting country, surveillance target group and flock status in 2016 are summarised in Tables 11 and 12, respectively. Because of the differences in the size of the small ruminant population, national surveillance efforts differ across reporting countries. Taking into account the number of samples tested in the SHC and NSHC target groups, and those required according to the TSE regulation (Table 4), 23 MSs fulfilled the requirements for sheep testing and 17 fulfilled the requirements for goat testing.

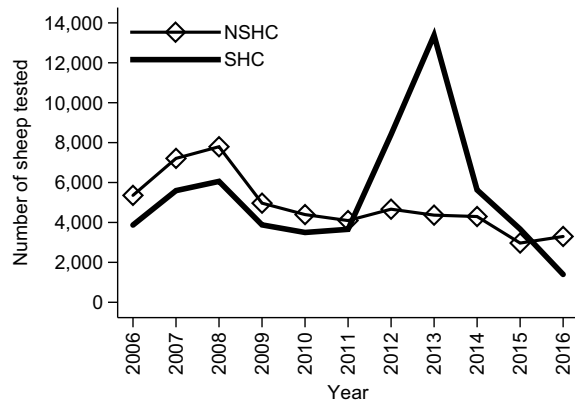
The ratio of tested sheep in TSE-infected vs non-TSE-infected flocks in the EU was 1:17.8 in 2016, which is slightly lower than in 2015 when the ratio was 1:18.5. The high ratio reveals that, in general, the number of sheep flocks under restrictions is very low. Only 15,230 sheep (5%) were tested from TSE-infected flocks, and most of those were derived from the culling (EM) of animals in flocks from the three MSs most affected by CS: EL, ES and IT. In goats, the pattern is similar, with ratios of tested goats in TSE-infected to non-TSE-infected flocks of 1:30.4 and 1:23.5 in 2015 and 2016, respectively. The main contributors to goats tested from TSE infected herds were RO, ES and IT even though the main contributor to the CS caseload in the EU is still CY.

Among non-infected flocks in both species, there is a decreasing trend in the number of sheep and goats notified as clinical suspects and tested as TSE suspects (SU target surveillance group). This may highlight either the decrease in the number of incident clinical cases or a decreasing awareness of scrapie among sheep and goat owners.

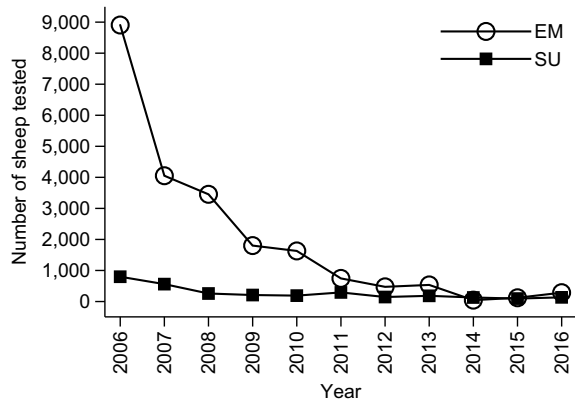
**A: Non-infected flocks**



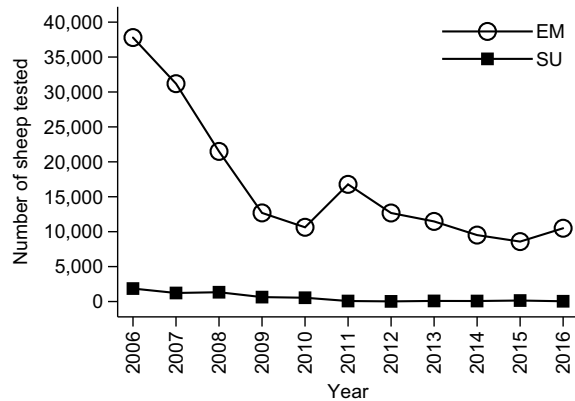
**B: TSE-infected flocks**



**C: Non-infected flocks**



**D: TSE-infected flocks**



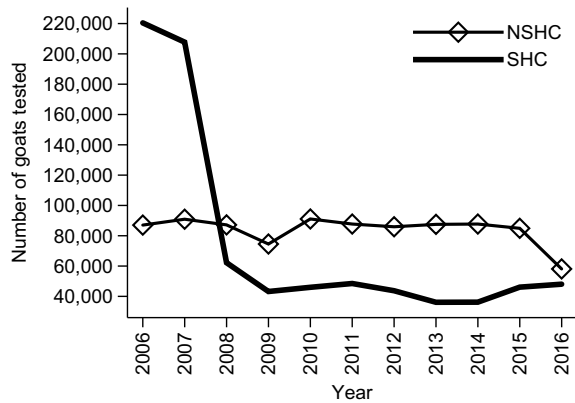
Data from Norway, Switzerland and Iceland are not included.

A and C: Non-infected flocks, including 'Other' or 'Unknown'; B and D: TSE-infected flocks.

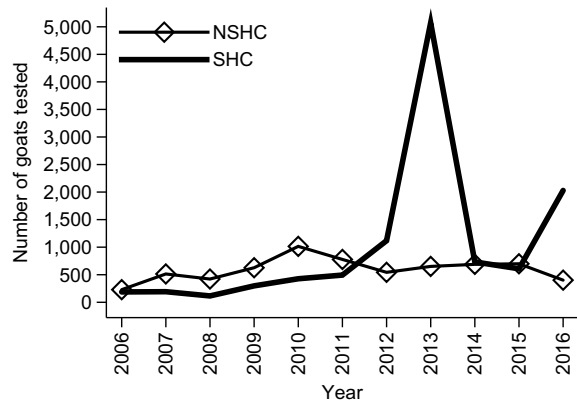
EM: animals culled under TSE eradication measures; NSHC: Animals not slaughtered for human consumption; SHC: animals slaughtered for human consumption; SU: animals clinically suspected of being infected by a TSE.

**Figure 6:** Number of sheep in the EU tested for TSE by surveillance target group (NSHC and SHC in A and B; EM and SU in C and D) and flock status in the period 2006–2016

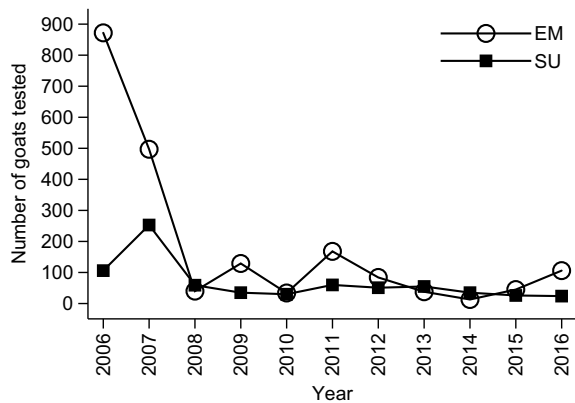
**A: Non-infected flocks**



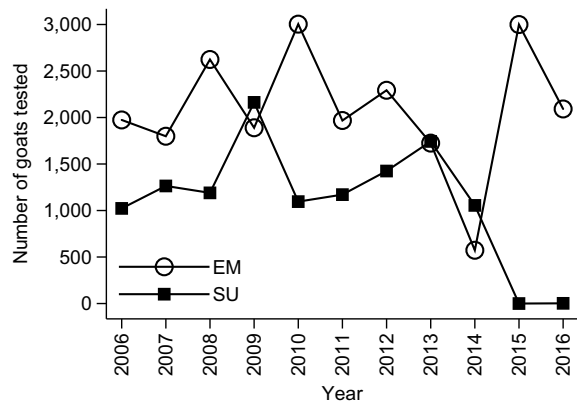
**B: TSE-infected flocks**



**C: Non-infected flocks**



**D: TSE-infected flocks**



Data from Norway, Switzerland and Iceland are not included.

A and C: Non-infected flocks, including 'Other' or 'Unknown'. B and D: TSE-infected flocks.

EM: animals culled under TSE eradication measures; NSHC: Animals not slaughtered for human consumption; SHC: animals slaughtered for human consumption; SU: animals clinically suspected of being infected by a TSE.

**Figure 7:** Number of goats in the EU tested for TSE by surveillance target group (NSHC and SHC in A and B; EM and SU in C and D) and flock status in the period 2006–2016

**Table 11:** Number of sheep tested for TSE by reporting country, surveillance target group and flock status in 2016 in the EU and other reporting countries

Flock status	Infected flocks					Other flocks <sup>(a)</sup>					Unknown <sup>(a)</sup>					Subtotal non-infected flocks	Total
	EM	NSHC	SHC	SU	Subtotal infected flocks	EM	NSHC	SHC	SU	Subtotal Other flocks	EM	NSHC	SHC	SU	Subtotal Unknown		
AT	0	13	9	0	22	0	2,532	136	2	2,670	0	0	0	0	0	2,670	2,692
BE	0	0	0	0	0	0	1,591	0	0	1,591	0	0	0	0	0	1,591	1,591
BG	0	0	0	0	0	0	457	10,737	0	11,194	0	0	0	0	0	11,194	11,194
CY	0	0	0	0	0	0	4,567	2,144	0	6,711	0	0	0	0	0	6,711	6,711
CZ	0	0	0	0	0	0	2,846	28	0	2,874	0	0	0	0	0	2,874	2,874
DE	0	0	0	0	0	0	0	0	0	0	70	12,322	7,026	36	19,454	19,454	19,454
DK	0	0	0	0	0	0	680	0	0	680	0	0	0	0	0	680	680
EE	0	0	0	0	0	0	0	0	0	0	0	300	0	0	300	300	300
EL	1,530	0	0	0	1,530	0	0	0	0	0	0	2,887	3,065	24	5,976	5,976	7,506
ES	5,585	0	0	0	5,585	0	12,681	9,678	1	22,360	0	0	0	0	0	22,360	27,945
FI	0	0	11	0	11	0	1,381	0	1	1,382	0	0	0	0	0	1,382	1,393
FR	168	9	98	0	275	0	19,622	5,713	1	25,336	0	0	0	0	0	25,336	25,611
HR	0	0	2	0	2	0	1,755	0	1	1,756	0	0	0	0	0	1,756	1,758
HU	0	1,171	200	0	1,371	0	6,380	6,234	0	12,614	0	0	0	0	0	12,614	13,985
IE	55	83	232	0	370	0	201	0	0	201	0	10,331	10,530	0	20,861	21,062	21,432
IT	3,150	326	402	3	3,881	2	11,523	10,375	3	21,903	0	0	0	0	0	21,903	25,784
LT	0	0	0	0	0	0	1,068	163	0	1,231	0	0	0	0	0	1,231	1,231
LU	0	0	0	0	0	0	101	0	0	101	0	0	0	0	0	101	101
LV	0	0	0	0	0	0	115	0	0	115	0	0	0	0	0	115	115
MT	0	0	0	0	0	0	125	67	0	192	0	5	0	0	5	197	197
NL	0	0	0	0	0	0	0	0	0	0	0	1,520	0	0	1,520	1,520	1,520
PL	0	0	0	0	0	0	8,638	7,385	0	16,023	0	0	0	0	0	16,023	16,023
PT	0	0	0	0	0	0	0	0	0	0	214	14,918	6,392	0	21,524	21,524	21,524
RO	0	86	234	40	360	0	9,275	20,190	43	29,508	0	1,047	1,148	7	2,202	31,710	32,070
SE	0	0	3	0	3	0	6,368	0	0	6,368	0	0	0	0	0	6,368	6,371
SI	0	6	0	0	6	0	2,186	202	14	2,402	0	0	0	0	0	2,402	2,408
SK	0	969	176	0	1,145	0	10,420	0	0	10,420	0	0	0	0	0	10,420	11,565

Flock status	Infected flocks					Other flocks <sup>(a)</sup>					Unknown <sup>(a)</sup>					Subtotal non-infected flocks	Total
	EM	NSHC	SHC	SU	Subtotal infected flocks	EM	NSHC	SHC	SU	Subtotal Other flocks	EM	NSHC	SHC	SU	Subtotal Unknown		
<b>UK</b>	6	635	28	0	669	0	14,729	6,916	2	21,647	0	0	0	0	0	21,647	22,316
<b>Total EU</b>	<b>10,494</b>	<b>3,298</b>	<b>1,395</b>	<b>43</b>	<b>15,230</b>	<b>2</b>	<b>119,241</b>	<b>79,968</b>	<b>68</b>	<b>199,279</b>	<b>284</b>	<b>43,330</b>	<b>28,161</b>	<b>67</b>	<b>71,842</b>	<b>271,121</b>	<b>286,351</b>
<b>CH</b>	0	0	0	0	0	0	0	0	1	1	0	0	0	0	0	1	1
<b>IS</b>	357	0	0	0	357	0	0	2	0	2	0	17	2,359	6	2,382	2,384	2,741
<b>NO</b>	0	0	0	0	0	168	6,324	9,859	29	16,380	0	0	0	0	0	16,380	16,380
<b>Total EFTA</b>	<b>357</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>357</b>	<b>168</b>	<b>6,324</b>	<b>9,861</b>	<b>30</b>	<b>16,383</b>	<b>0</b>	<b>17</b>	<b>2,359</b>	<b>6</b>	<b>2,382</b>	<b>18,765</b>	<b>19,122</b>
<b>Total sheep</b>	<b>10,851</b>	<b>3,298</b>	<b>1,395</b>	<b>43</b>	<b>15,587</b>	<b>170</b>	<b>125,565</b>	<b>89,829</b>	<b>98</b>	<b>215,662</b>	<b>284</b>	<b>43,347</b>	<b>30,520</b>	<b>73</b>	<b>74,224</b>	<b>289,886</b>	<b>305,473</b>

EM: animals culled under TSE eradication measures; NSHC: Animals not slaughtered for human consumption; SHC: animals slaughtered for human consumption; SU: animals clinically suspected of being infected by a TSE.

(a): The 'Other' and 'Unknown' are assumed to be non-infected flocks.



**Table 12:** Number of goats tested for TSE by reporting country, surveillance target group and herd status in 2016 in the EU and other reporting countries

Herd status	Infected herds					Other herds <sup>(a)</sup>					Unknown <sup>(a)</sup>					Subtotal non-infected herds	Total
	EM	NSHC	SHC	SU	Subtotal infected herds	EM	NSHC	SHC	SU	Subtotal Other herds	EM	NSHC	SHC	SU	Subtotal Unknown		
AT	0	1	0	0	1	0	849	35	0	884	0	0	0	0	0	884	885
BE	0	0	0	0	0	0	558	0	0	558	0	0	0	0	0	558	558
BG	0	0	0	0	0	0	92	1,760	0	1,852	0	0	0	0	0	1,852	1,852
CY	357	0	0	0	357	0	5,781	3,457	0	9,238	0	0	0	0	0	9,238	9,595
CZ	0	0	0	0	0	0	416	0	0	416	0	0	0	0	0	416	416
DE	0	0	0	0	0	0	0	0	0	0	1	1,668	253	8	1,930	1,930	1,930
DK	0	0	0	0	0	0	132	0	0	132	0	0	0	0	0	132	132
EE	0	0	0	0	0	0	0	0	0	0	0	22	0	0	22	22	22
EL	86	0	0	0	86	0	0	0	0	0	0	658	1,164	5	1,827	1,827	1,913
ES	853	0	0	0	853	0	10,128	9,547	0	19,675	0	0	0	0	0	19,675	20,528
FI	0	0	0	0	0	0	132	0	0	132	0	0	0	0	0	132	132
FR	1	0	713	0	714	0	18,003	5,389	0	23,392	0	0	0	0	0	23,392	24,106
HR	0	0	0	0	0	0	346	0	2	348	0	0	0	0	0	348	348
HU	0	0	0	0	0	0	105	56	0	161	0	0	0	0	0	161	161
IE	0	0	0	0	0	0	55	0	0	55	0	78	0	0	78	133	133
IT	559	20	154	0	733	97	7,096	16,785	0	23,978	0	0	0	0	0	23,978	24,711
LT	0	0	0	0	0	0	11	6	0	17	0	0	0	0	0	17	17
LU	0	0	0	0	0	0	95	0	0	95	0	0	0	0	0	95	95
LV	0	0	0	0	0	0	12	0	0	12	0	0	0	0	0	12	12
MT	0	0	0	0	0	0	116	56	0	172	0	0	0	0	0	172	172
NL	0	0	0	0	0	0	0	0	0	0	0	1,528	0	0	1,528	1,528	1,528
PL	0	0	0	0	0	0	3,074	278	0	3,352	0	0	0	0	0	3,352	3,352
PT	0	0	0	0	0	0	0	0	0	0	3	1,508	62	0	1,573	1,573	1,573
RO	0	34	1,113	2	1,149	0	3,602	8,515	1	12,118	0	633	645	0	1,278	13,396	14,545
SE	0	0	0	0	0	0	49	0	0	49	0	0	0	0	0	49	49
SI	0	0	0	0	0	0	657	48	6	711	0	0	0	0	0	711	711
SK	0	0	0	0	0	0	212	0	0	212	0	0	0	0	0	212	212

Herd status Surveillance target group	Infected herds					Other herds <sup>(a)</sup>					Unknown <sup>(a)</sup>					Subtotal non-infected herds	Total
	EM	NSHC	SHC	SU	Subtotal infected herds	EM	NSHC	SHC	SU	Subtotal Other herds	EM	NSHC	SHC	SU	Subtotal Unknown		
<b>UK</b>	241	347	50	0	638	0	504	0	2	506	0	0	0	0	0	506	1,144
<b>Total EU</b>	<b>2,097</b>	<b>402</b>	<b>2,030</b>	<b>2</b>	<b>4,531</b>	<b>97</b>	<b>52,025</b>	<b>45,932</b>	<b>11</b>	<b>98,065</b>	<b>4</b>	<b>6,095</b>	<b>2,124</b>	<b>13</b>	<b>8,236</b>	<b>106,301</b>	<b>110,832</b>
<b>CH</b>	0	0	0	0	0	0	0	0	2	2	0	0	0	0	0	2	2
<b>IS</b>	0	0	0	0	0	0	0	0	0	0	0	0	3	0	3	3	3
<b>NO</b>	0	0	0	0	0	0	449	13	1	463	0	0	0	0	0	463	463
<b>Total EFTA</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>449</b>	<b>13</b>	<b>3</b>	<b>465</b>	<b>0</b>	<b>0</b>	<b>3</b>	<b>0</b>	<b>3</b>	<b>468</b>	<b>468</b>
<b>Total</b>	<b>2,097</b>	<b>402</b>	<b>2,030</b>	<b>2</b>	<b>4,531</b>	<b>97</b>	<b>52,474</b>	<b>45,945</b>	<b>14</b>	<b>98,530</b>	<b>4</b>	<b>6,095</b>	<b>2,127</b>	<b>13</b>	<b>8,239</b>	<b>106,769</b>	<b>111,300</b>

EM: animals culled under TSE eradication measures; NSHC: Animals not slaughtered for human consumption; SHC: animals slaughtered for human consumption; SU: animals clinically suspected of being infected by a TSE.

(a): The 'Other' and 'Unknown' are assumed to be non-TSE-infected flocks.

A total of 685 scrapie cases in sheep was reported in the EU in 2016 (Table 13), similar to the previous year (641 cases). They were reported by 20 MSs. In particular, CS was reported by 9 different MSs and one non-MS (CY, EL, ES, FR, IE, IT, PT, RO and SK, and in IS), whereas AS was reported by 18 different MSs and one non-MS (the 9 that reported CS except CY and RO plus AT, BG, CZ, DE, DK, FI, HU, PL, SE, SI and the UK and in NO). About 81.2% of the ovine cases were reported by four countries, namely EL, ES, IT and RO. A total of 554 ovine cases were CS cases (81%), 122 AS (18%) and a few unknown (9 cases). An additional 25 cases of scrapie in sheep were reported by two non-MSs: 11 CS cases in Iceland and 14 AS cases in Norway. The geographical distribution of AS and CS in sheep is shown in Appendix D.

Currently, the reporting in the EU database does not include the number of newly infected flocks and herds during the reporting year. The number of index cases can therefore be considered a proxy for the incidence of scrapie at country level. In sheep in 2016, 32% of all cases were index cases, with a higher proportion (106/122) in atypical scrapie cases than in CS cases (112/554). Using these cases as a proxy for the flock-level incidence in sheep and comparing 2015 with 2016, there was a slight increase in CS cases (from 109 to 112) and a decrease in AS cases (from 120 to 106).

A total of 634 scrapie cases in goats was reported in the EU in 2016, a 40% reduction compared with 2015 when 1,052 cases were reported. This is due mainly to the decrease in the number of CS scrapie cases in goats in a single reporting country, Cyprus, from 923 in 2015 to 570 in 2016.

Scrapie in goats was reported by nine MSs. In particular, CS was reported by seven MSs (BG, CY, EL, ES, IT, RO and the UK), whereas AS was reported by 5 MSs (EL, ES, FR, IT and PT). As mentioned, most of the cases (89.9%) were reported by Cyprus.

Of the caprine scrapie cases, 621 were CS (98%) and 13 were atypical scrapie (2%) (Table 14). In goats, the proportion of index cases was 6.8% (13/13 atypical scrapie and 30/621 CS cases). The low proportion is due mainly to Cyprus where many cases in 2016 were still reported from TSE-infected herds. An improved epidemiological situation is suggested by the low number of new incident cases (only 12 index cases out of 570). Using the index cases in goats as a proxy for the herd-level incidence and comparing 2015 with 2016, there was a decrease in CS cases (from 37 to 30) and no change in AS cases (13). The geographical distribution of AS and CS in goats is shown in Appendix D.

In absolute numbers and without restricting the calculation to index cases, CS is still the most frequently reported type of scrapie in the EU: the CS/AS ratio was 4.5:1 in sheep and 47.8:1 in goats. If CY is excluded, the ratio in goats is similar to that in sheep, i.e. 3.9:1.

**Table 13:** Number of scrapie cases in sheep by country, case type, index case status, surveillance target group in 2016 in the EU and other reporting countries

Case type	Atypical scrapie (AS)									Classical scrapie (CS)									Unknown		Total				
	No				Yes				Total AS	No				Yes				Total CS	No EM	Total Unknown					
	EM	NSHC	SHC	Subtotal	NSHC	SHC	SU	Subtotal		EM	NSHC	SHC	SU	Subtotal	NSHC	SHC	SU					Subtotal			
AT	0	0	0	0	1	0	0	1	1	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	1
BG	0	0	0	0	1	1	0	2	2	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	2
CY	0	0	0	0	0	0	0	0	0	0	3	4	0	7	0	0	0	0	0	0	0	0	0	0	7
CZ	0	0	0	0	1	1	0	2	2	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	2
DE	0	0	0	0	5	0	0	5	5	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	5
DK	0	0	0	0	1	0	0	1	1	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	1
EL	0	0	0	0	2	0	0	2	2	78	88	2	5	173	32	10	12	54	227	0	0	0	0	0	229
ES	1	0	0	1	7	5	0	12	13	85	0	0	0	85	3	3	0	6	91	0	0	0	0	0	104
FI	0	0	0	0	1	0	1	2	2	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	2
FR	0	0	0	0	3	1	0	4	4	0	0	0	0	0	1	0	1	2	2	0	0	0	0	0	6
HU	0	1	3	4	13	6	0	19	23	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	23
IE	0	0	0	0	0	1	0	1	1	0	0	0	0	0	1	0	0	1	1	0	0	0	0	0	2
IT	0	0	0	0	1	4	0	5	5	111	0	0	0	111	12	9	2	23	134	9	9	0	0	0	148
PL	0	0	0	0	7	1	0	8	8	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	8
PT	0	5	5	10	12	6	0	18	28	4	0	0	0	4	3	0	0	3	7	0	0	0	0	0	35
RO	0	0	0	0	0	0	0	0	0	0	2	2	50	54	6	2	13	21	75	0	0	0	0	0	75
SE	0	0	0	0	3	0	0	3	3	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	3
SI	0	0	0	0	3	0	0	3	3	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	3
SK	0	0	0	0	5	0	0	5	5	0	1	7	0	8	1	1	0	2	10	0	0	0	0	0	15
UK	0	1	0	1	9	4	0	13	14	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	14
<b>Total EU 28</b>	<b>1</b>	<b>7</b>	<b>8</b>	<b>16</b>	<b>75</b>	<b>30</b>	<b>1</b>	<b>106</b>	<b>122</b>	<b>278</b>	<b>94</b>	<b>15</b>	<b>55</b>	<b>442</b>	<b>59</b>	<b>25</b>	<b>28</b>	<b>112</b>	<b>554</b>	<b>9</b>	<b>9</b>	<b>0</b>	<b>0</b>	<b>685</b>	
IS	0	0	0	0	0	0	0	0	0	8	0	0	1	9	0	0	2	2	11	0	0	0	0	0	11
NO	0	1	0	1	6	7	0	13	14	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	14
<b>Total EFTA</b>	<b>0</b>	<b>1</b>	<b>0</b>	<b>1</b>	<b>6</b>	<b>7</b>	<b>0</b>	<b>13</b>	<b>14</b>	<b>8</b>	<b>0</b>	<b>0</b>	<b>1</b>	<b>9</b>	<b>0</b>	<b>0</b>	<b>2</b>	<b>2</b>	<b>11</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>25</b>	
<b>Total</b>	<b>1</b>	<b>8</b>	<b>8</b>	<b>17</b>	<b>81</b>	<b>37</b>	<b>1</b>	<b>119</b>	<b>136</b>	<b>286</b>	<b>94</b>	<b>15</b>	<b>56</b>	<b>451</b>	<b>59</b>	<b>25</b>	<b>30</b>	<b>114</b>	<b>565</b>	<b>9</b>	<b>9</b>	<b>0</b>	<b>0</b>	<b>710</b>	

EM: animals culled under TSE eradication measures; NSHC: Animals not slaughtered for human consumption; SHC: animals slaughtered for human consumption; SU: animals clinically suspected of being infected by a TSE.

Only the reporting countries in which scrapie cases in sheep were detected in 2016 are mentioned in the table; no cases of scrapie were reported in non-MS (Switzerland, Iceland and Norway).

**Table 14:** Number of scrapie cases in goats by country, case type, index case status, surveillance target group in 2016 in the EU and other reporting countries

Case type Index case Surveillance target group	Atypical scrapie (AS)				Classical scrapie (CS)										Total
	Yes			Total AS	No					Yes				Total CS	
	NSHC	SHC	Subtotal		EM	NSHC	SHC	SU	Subtotal	NSHC	SHC	SU	Subtotal		
<b>BG</b>	0	0	0	<b>0</b>	0	0	0	0	0	0	2	0	2	<b>2</b>	<b>2</b>
<b>CY</b>	0	0	0	<b>0</b>	292	184	82	0	558	9	3	0	12	<b>570</b>	<b>570</b>
<b>EL</b>	0	1	1	<b>1</b>	2	3	0	1	6	1	1	3	5	<b>11</b>	<b>12</b>
<b>ES</b>	2	3	5	<b>5</b>	14	0	0	0	14	4	1	0	5	<b>19</b>	<b>24</b>
<b>FR</b>	2	1	3	<b>3</b>	0	0	0	0	0	0	0	0	0	<b>0</b>	<b>3</b>
<b>IT</b>	0	3	3	<b>3</b>	4	0	0	0	4	1	3	0	4	<b>8</b>	<b>11</b>
<b>PT</b>	1	0	1	<b>1</b>	0	0	0	0	0	0	0	0	0	<b>0</b>	<b>1</b>
<b>RO</b>	0	0	0	<b>0</b>	0	0	0	1	1	0	0	2	2	<b>3</b>	<b>3</b>
<b>UK</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>4</b>	<b>2</b>	<b>0</b>	<b>2</b>	<b>8</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>8</b>	<b>8</b>
<b>Total EU 28</b>	<b>5</b>	<b>8</b>	<b>13</b>	<b>13</b>	<b>316</b>	<b>189</b>	<b>82</b>	<b>4</b>	<b>591</b>	<b>15</b>	<b>10</b>	<b>5</b>	<b>30</b>	<b>621</b>	<b>634</b>

EM: animals culled under TSE eradication measures; NSHC: Animals not slaughtered for human consumption; SHC: animals slaughtered for human consumption; SU: animals clinically suspected of being infected by a TSE.

Only the reporting countries in which scrapie cases in sheep were detected in 2016 are mentioned in the table; no cases of scrapie were reported in non-MS (Switzerland, Iceland and Norway).

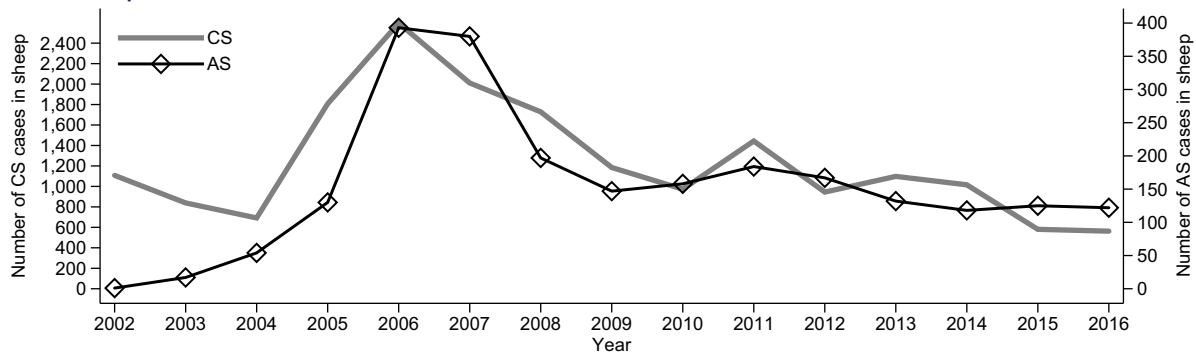
The evolution in the number of scrapie cases detected at EU level is shown for each species in Figure 8. After the 2006 peak in the number of reported scrapie cases in sheep with 3,507 cases (when the number of tests also peaked), CS cases have decreased from 1,444 in 2011 to 1,016 in 2014, 498 in 2015 and 554 in 2016.

In goats, this trend in CS cases is less clear but is mainly affected by one single MS (CY), where the number of detected cases has consistently declined since the peak in 2013 when 1,672 cases were reported by CY and 1,804 by the EU. However, the total number of cases has decreased consistently since 2013 to 621 in the reporting year.

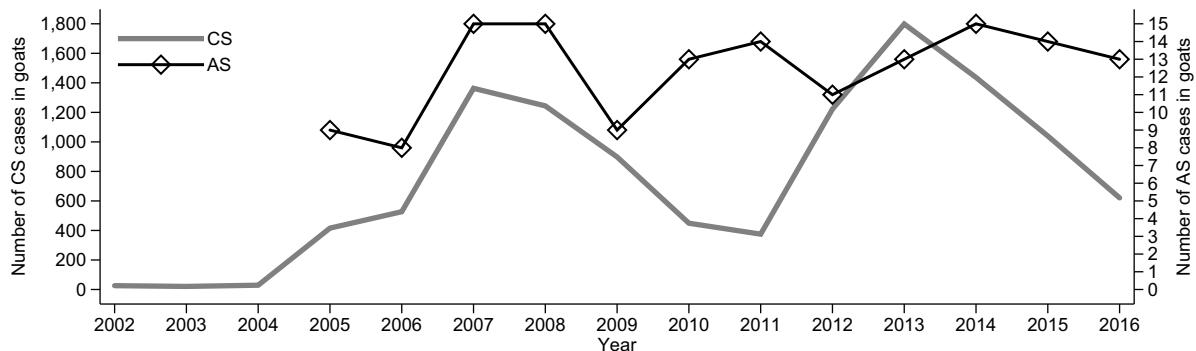
Figure 9 shows the evolution of the proportion of scrapie cases (per 10,000 tests) by target group in non-TSE-infected flocks of sheep and goats, i.e. the main output of the surveillance. As expected for CS, the average number of CS cases per 10,000 in NSHC, i.e. the risk animals, is higher (p-value < 0.001) than that in SHC animals both for sheep (4.7 vs 2.8) and goats (3.0 vs 1.7). Looking at AS, a significant difference is detectable only in sheep (4.4 vs 3.4, p-value < 0.001).

Focusing on the last 10 years, the EU trend has remained quite stable for both CS and AS, ranging between 2 and 6 cases per 10,000 tested animals. There is no significant trend for ovine CS and AS (p values = 0.2 and 0.51, respectively) and only a slight significant increase in both caprine CS (Beta = 0.06; p value = 0.001) and AS (Beta = 0.06; p value = 0.03). However, the individual national trends may show different patterns (EFSA BIOHAZ Panel, 2014). In goats, there was a decrease in the number of cases during the last four years.

### A: Sheep



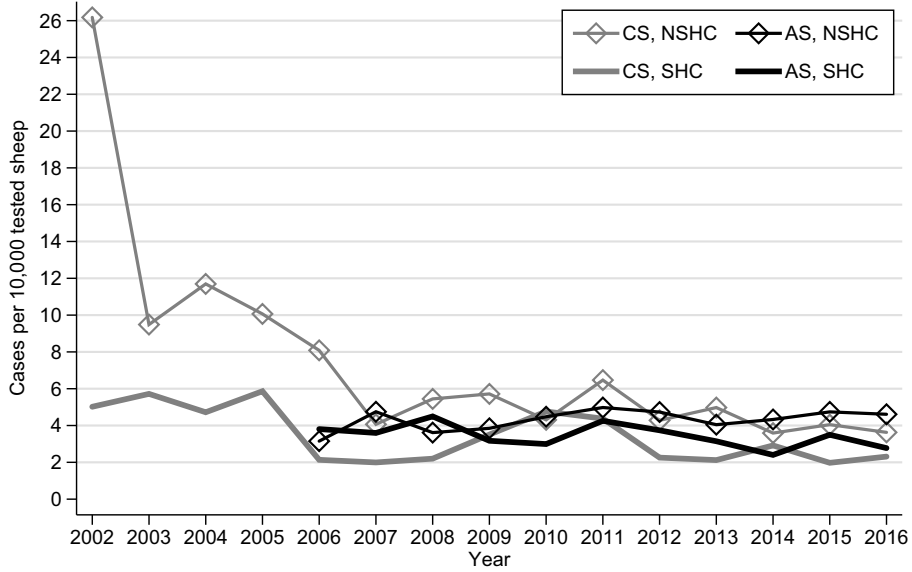
### B: Goats



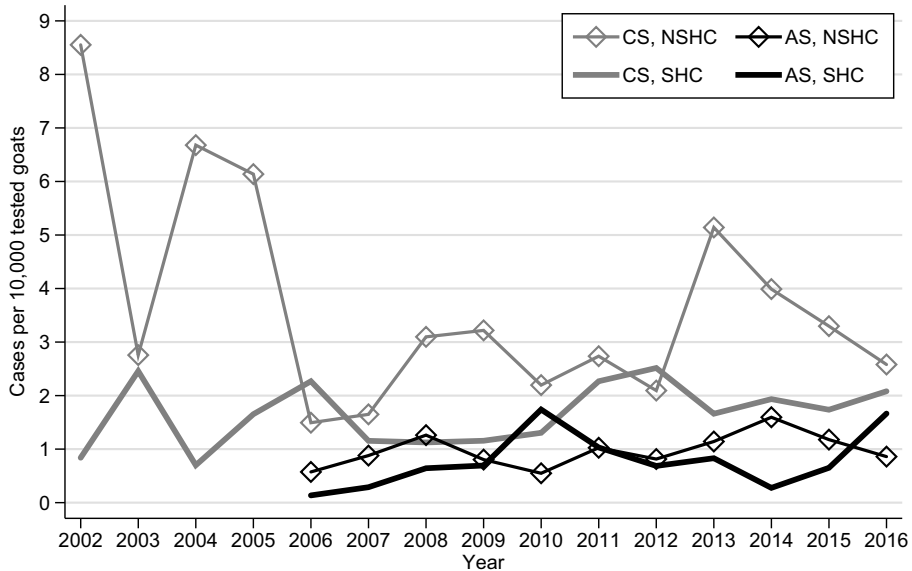
Right-hand y-axis: AS (Atypical scrapie); Left-hand y-axis: CS (Classical scrapie).

**Figure 8:** Number of scrapie TSE cases in the EU reported by case type in the period 2002–2016 in (A) sheep and (B) goats

### A: Sheep



### B: Goats



Note: This figure is restricted to testing performed in NSHC and SHC target groups from non-infected flocks or not previously known as infected.  
 NSHC: Animals not slaughtered for human consumption; SHC: animals slaughtered for human consumption; AS: Atypical scrapie; CS: Classical scrapie.

**Figure 9:** Proportion of scrapie cases (per 10,000 tests) in the EU in (A) sheep and (B) goats in the period 2002–2016



**Table 15:** Total number of classical scrapie cases in sheep by year and country between 2001 and 2016 in the EU and other reporting countries

Country	2001	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	2012	2013	2014	2015	2016	Total CS
<b>BE</b>	0	24	2	10	1	0	1	0	0	0	0	0	0	0	0	0	<b>38</b>
<b>BG</b>	0	0	0	0	0	0	0	2	1	3	0	2	0	3	0	0	<b>11</b>
<b>CY</b>	0	0	0	17	670	866	803	540	185	49	12	9	8	25	13	7	<b>3,204</b>
<b>CZ</b>	0	16	13	10	1	0	0	16	0	0	0	0	0	0	0	0	<b>56</b>
<b>DE</b>	0	12	23	43	27	3	6	0	0	1	0	0	0	0	1	0	<b>116</b>
<b>EL</b>	0	68	56	34	52	156	344	621	716	616	879	565	601	557	252	227	<b>5,744</b>
<b>ES</b>	0	21	29	14	125	57	148	215	87	126	40	33	48	36	69	91	<b>1,139</b>
<b>FR</b>	0	245	125	59	70	587	288	85	19	14	5	2	4	28	1	2	<b>1,534</b>
<b>HU</b>	0	0	0	0	0	2	4	1	1	0	0	0	1	1	0	0	<b>10</b>
<b>IE</b>	0	66	44	56	54	126	79	20	32	22	40	8	7	19	1	1	<b>575</b>
<b>IT</b>	0	135	43	24	329	383	197	148	108	104	211	192	260	241	141	143	<b>2,659</b>
<b>NL</b>	0	70	63	54	62	84	36	24	4	1	1	0	2	0	0	0	<b>401</b>
<b>PT</b>	0	0	0	0	0	0	0	12	0	3	4	1	6	0	0	7	<b>33</b>
<b>RO</b>	0	0	0	0	3	7	28	0	8	26	112	127	154	95	98	75	<b>733</b>
<b>SI</b>	0	0	0	12	101	42	12	2	3	4	0	0	0	0	0	0	<b>176</b>
<b>SK</b>	0	0	4	32	10	9	8	27	0	2	6	0	0	6	3	10	<b>117</b>
<b>UK</b>	0	451	436	331	303	252	50	12	12	1	133	6	6	0	2	0	<b>1,995</b>
<b>Total EU 28</b>	0	1,108	838	696	1,808	2,574	2,004	1,725	1,176	972	1,443	945	1,097	1,011	581	563	<b>18,541</b>
<b>IS</b>	9	12	19	19	9	21	15	57	7	5	0	0	0	2	29	11	<b>215</b>
<b>NO</b>	1	1	1	2	0	7	0	0	1	0	0	0	0	0	0	0	<b>13</b>
<b>Total EFTA</b>	10	13	20	21	9	28	15	57	8	5	0	0	0	2	29	11	<b>228</b>
<b>Total</b>	<b>10</b>	<b>1,121</b>	<b>858</b>	<b>717</b>	<b>1,817</b>	<b>2,602</b>	<b>2,019</b>	<b>1,782</b>	<b>1,184</b>	<b>977</b>	<b>1,443</b>	<b>945</b>	<b>1,097</b>	<b>1,013</b>	<b>610</b>	<b>574</b>	<b>18,769</b>

Note: Only the reporting countries in which classical scrapie cases in sheep were detected are mentioned in the table.

Since 1 July 2013, according to point 2 of Section A of Chapter A of Annex VIII to Regulation (EC) No 999/2001 as amended by Regulation (EU) 630/2013, a Member State (MS) can submit a request to the Commission to be recognised as a MS, or zone of a MS, with a negligible risk of classical scrapie (CS). By the end of 2016, MSS that can submit a request based on the data are Belgium, the Czech Republic, Croatia, Estonia, Latvia, Lithuania, Luxembourg, Malta and Poland. MSs with a negligible risk of classical scrapie (CS) are Austria, Denmark, Finland and Sweden.

**Table 16:** Total number of atypical scrapie cases in sheep by year and country between 2002 and 2016 in the EU and other reporting countries

Country	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	2012	2013	2014	2015	2016	Total AS
AT	0	0	0	0	0	0	0	0	0	4	3	2	2	1	1	<b>13</b>
BE	1	0	1	1	3	2	0	0	0	0	0	0	0	0	0	<b>8</b>
BG	0	0	0	0	0	0	2	0	0	0	2	0	0	0	2	<b>6</b>
CZ	0	0	0	0	0	1	0	0	0	0	0	0	1	3	2	<b>7</b>
DE	0	0	0	0	21	9	7	12	12	19	8	7	10	10	5	<b>120</b>
DK	0	0	0	0	3	0	2	0	2	5	0	0	0	0	1	<b>13</b>
EE	0	0	0	0	0	0	0	0	1	1	0	0	0	0	0	<b>2</b>
EL	0	0	0	0	2	1	3	4	1	4	5	3	5	2	2	<b>32</b>
ES	0	9	1	3	21	26	23	19	21	19	20	18	6	12	13	<b>211</b>
FI	0	0	1	1	2	1	0	0	3	0	1	1	1	0	2	<b>13</b>
FR	0	0	0	13	188	174	49	29	30	24	22	10	6	5	4	<b>554</b>
HR	0	0	0	0	0	0	0	0	0	0	0	1	0	1	0	<b>2</b>
HU	0	0	0	0	5	3	8	15	7	12	11	9	22	14	23	<b>129</b>
IE	0	0	2	0	0	2	0	5	2	1	4	4	7	7	1	<b>35</b>
IT	0	0	0	7	22	22	8	0	0	7	5	7	2	6	5	<b>91</b>
NL	0	0	0	2	0	2	0	0	1	7	5	1	0	0	0	<b>18</b>
PL	0	0	0	0	0	0	0	4	2	4	2	5	13	9	8	<b>47</b>
PT	0	1	28	71	70	91	78	36	46	40	44	37	20	30	28	<b>620</b>
SE	0	4	2	1	8	2	0	2	4	3	3	3	7	3	3	<b>45</b>
SI	0	0	0	0	0	0	0	0	2	1	0	1	1	2	3	<b>10</b>
SK	0	0	0	0	1	0	0	1	3	4	3	4	3	3	5	<b>27</b>
UK	0	0	17	30	68	44	17	26	19	24	28	18	11	17	14	<b>333</b>
<b>Total EU 28</b>	<b>1</b>	<b>14</b>	<b>52</b>	<b>129</b>	<b>414</b>	<b>380</b>	<b>197</b>	<b>153</b>	<b>156</b>	<b>179</b>	<b>166</b>	<b>131</b>	<b>117</b>	<b>125</b>	<b>122</b>	<b>2,336</b>
IS	0	0	2	0	0	1	1	0	0	1	0	2	0	1	0	<b>8</b>
NO	8	14	14	4	8	9	7	12	5	6	6	12	9	10	14	<b>138</b>
<b>Total EFTA</b>	<b>8</b>	<b>14</b>	<b>16</b>	<b>4</b>	<b>8</b>	<b>10</b>	<b>8</b>	<b>12</b>	<b>5</b>	<b>7</b>	<b>6</b>	<b>14</b>	<b>9</b>	<b>11</b>	<b>14</b>	<b>146</b>
<b>Total</b>	<b>9</b>	<b>28</b>	<b>68</b>	<b>133</b>	<b>422</b>	<b>390</b>	<b>205</b>	<b>165</b>	<b>161</b>	<b>186</b>	<b>172</b>	<b>145</b>	<b>126</b>	<b>136</b>	<b>136</b>	<b>2,482</b>

Only the reporting countries in which atypical scrapie cases in sheep were detected are mentioned in the table.

**Table 17:** Total number of classical scrapie cases in goats by year and country between 2001 and 2016 in the EU and other reporting countries

Country	2002	2003	2004	2005	2006	2007	2008	2009	2010	2011	2012	2013	2014	2015	2016	Total CS
<b>BG</b>	0	0	0	0	0	0	0	1	3	0	0	0	0	1	2	<b>7</b>
<b>CY</b>	0	0	11	369	470	1,219	1,121	808	342	295	1,102	1,672	1,364	923	570	<b>10,266</b>
<b>EL</b>	5	8	12	11	12	58	67	55	65	56	69	68	31	22	11	<b>550</b>
<b>ES</b>	1	1	0	10	6	15	4	6	5	10	3	2	8	16	19	<b>106</b>
<b>FI</b>	4	0	0	4	0	0	0	0	0	0	0	0	0	0	0	<b>8</b>
<b>FR</b>	15	9	4	8	14	3	14	9	22	0	5	25	0	40	0	<b>168</b>
<b>IT</b>	1	3	2	6	12	4	1	13	4	5	7	7	7	21	8	<b>101</b>
<b>RO</b>	0	0	0	0	0	2	0	0	0	0	1	3	1	1	3	<b>11</b>
<b>SI</b>	0	0	0	4	0	0	0	0	0	0	0	0	0	0	0	<b>4</b>
<b>UK</b>	0	0	0	4	13	62	37	6	8	9	21	16	26	16	8	<b>226</b>
<b>Total EU 28</b>	<b>26</b>	<b>21</b>	<b>29</b>	<b>416</b>	<b>527</b>	<b>1,363</b>	<b>1,244</b>	<b>898</b>	<b>449</b>	<b>375</b>	<b>1,208</b>	<b>1,793</b>	<b>1,437</b>	<b>1,040</b>	<b>621</b>	<b>11,447</b>
<b>IS</b>	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	<b>0</b>
<b>NO</b>	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	<b>0</b>
<b>Total EFTA</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>
<b>Total</b>	<b>26</b>	<b>21</b>	<b>29</b>	<b>416</b>	<b>527</b>	<b>1,363</b>	<b>1,244</b>	<b>898</b>	<b>449</b>	<b>375</b>	<b>1,208</b>	<b>1,793</b>	<b>1,437</b>	<b>1,040</b>	<b>621</b>	<b>11,447</b>

Only the reporting countries in which classical scrapie cases in goats were detected are mentioned in the table.

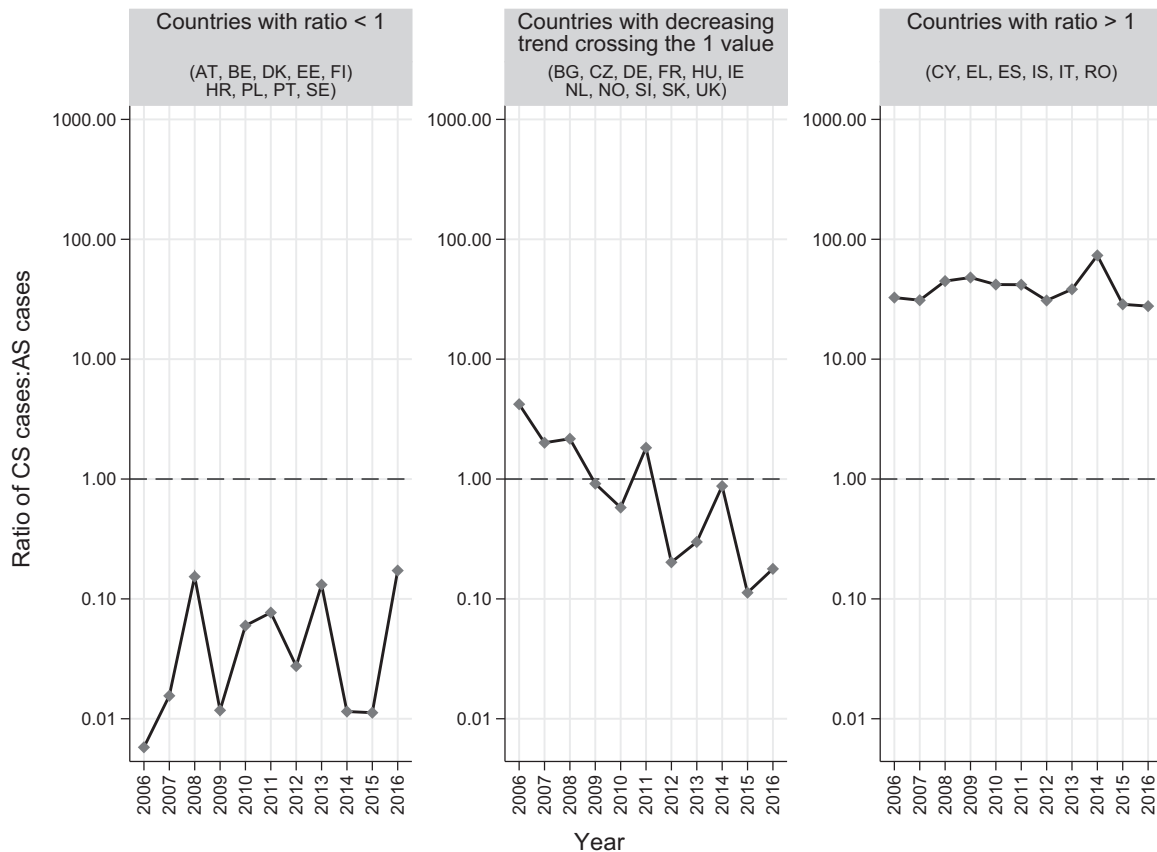
**Table 18:** Total number of atypical scrapie cases in goats by year and country between 2005 and 2016 in the EU and other reporting countries

Country	2005	2006	2007	2008	2009	2010	2011	2012	2013	2014	2015	2016	Total AS
AT	0	0	0	0	0	0	0	0	0	1	0	0	1
CY	0	0	0	0	0	0	0	0	0	0	1	0	1
DE	0	0	0	0	0	0	0	0	0	1	0	0	1
EL	0	0	0	0	0	0	0	0	1	1	1	1	4
ES	0	4	6	5	2	5	3	3	4	7	5	5	49
FI	0	0	0	0	1	0	0	0	0	0	0	0	1
FR	6	1	5	8	3	5	6	6	3	5	5	3	56
IT	3	3	3	1	0	0	4	0	3	0	1	3	21
PT	0	0	1	1	3	2	1	2	2	0	0	1	13
SI	0	0	0	0	0	0	0	0	0	0	1	0	1
<b>Total EU 28</b>	<b>9</b>	<b>8</b>	<b>15</b>	<b>15</b>	<b>9</b>	<b>12</b>	<b>14</b>	<b>11</b>	<b>13</b>	<b>15</b>	<b>14</b>	<b>13</b>	<b>148</b>
NO	0	1	0	0	0	0	0	0	0	0	0	0	1
<b>Total EFTA</b>	<b>0</b>	<b>1</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>1</b>
<b>Total</b>	<b>9</b>	<b>9</b>	<b>15</b>	<b>15</b>	<b>9</b>	<b>12</b>	<b>14</b>	<b>11</b>	<b>13</b>	<b>15</b>	<b>14</b>	<b>13</b>	<b>149</b>

Only the reporting countries in which atypical scrapie cases in goats were detected are mentioned in the table.

Tables 15 and 16 show the cases of classical and atypical scrapie, respectively, in sheep between 2002 and 2016. For this period, the annual CS:AS ratio was calculated and plotted for each reporting country (see Appendix B). Figure 10 shows the CS:AS ratio for three groups of countries aggregated based on the pattern of the CS:AS ratio: (1) countries with a CS:AS ratio that is always below one (more AS than CS cases have consistently been detected): AT, BE, DK, EE, FI, HR, PL, PT and SE; (2) countries with a decreasing CS:AS ratio over time (crossing the ratio of one at some point): BG, the CZ, DE, FR, HU, IE, the NL, NO, SK, SI, and the UK; (3) countries with a CS:AS ratio that is always greater than one (more CS than AS cases have consistently been detected): CY, EL, ES, IS, IT and RO.

Tables 17 and 18 show the cases of classical and atypical scrapie, respectively, in goats between 2002 and 2016.

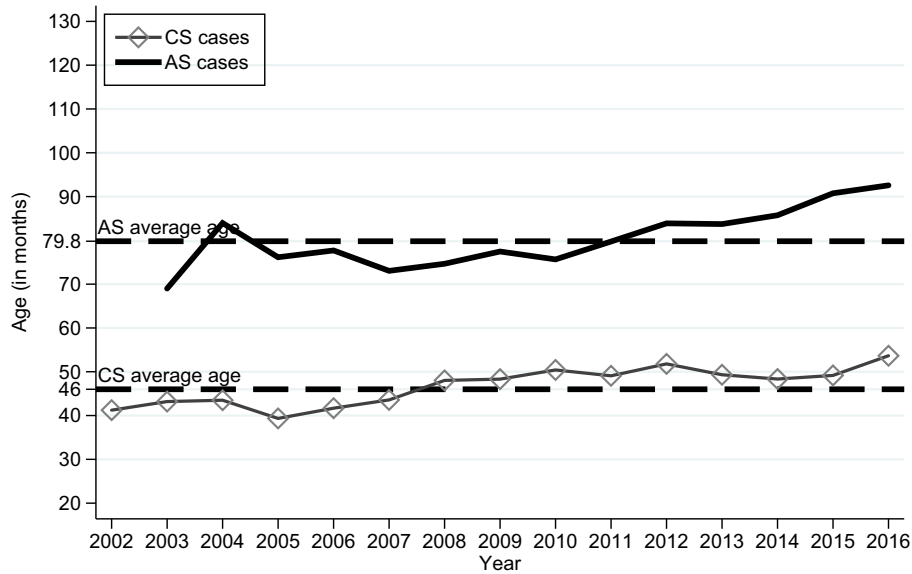


Only data from cases in sheep are considered. The CS:AS ratio's for the three groups of countries were calculated by aggregating the number of CS and AS cases for each year over the countries per group. Positive values of the CS:AS in a certain year show how many times CS cases were more frequently detected than AS cases. At CS:AS ratio equals to one, the number of CS and AS cases are equal in number.

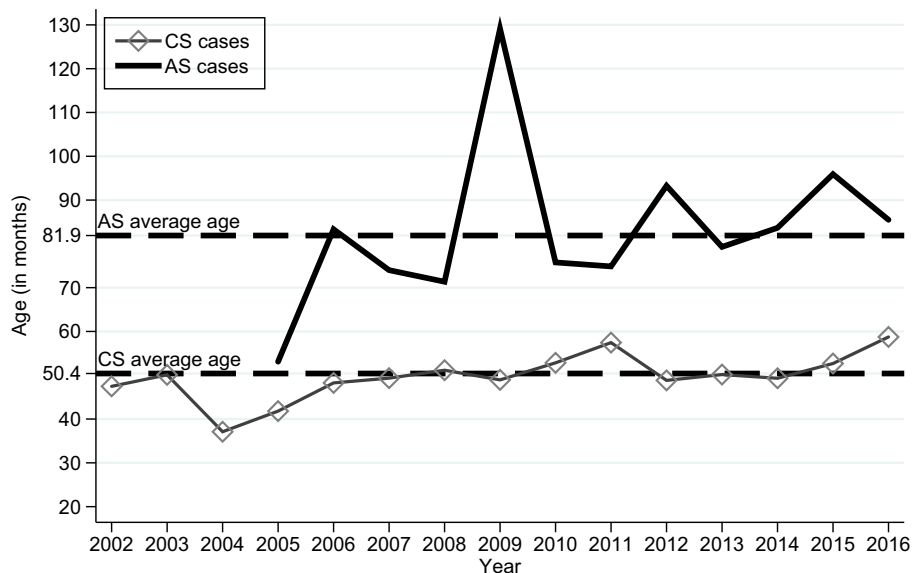
**Figure 10:** Evolution of the ratio of CS:AS cases in the period 2002–2016 in three distinct patterns

Figure 11 shows the average age of the TSE cases confirmed in sheep (A) and goats (B) by case type. In sheep, the average age of AS cases (79.9 months, standard error (SE): 0.83) is significantly higher ( $p$ -value < 0.001) than that of CS cases (46.0, SE: 0.17). Similarly, in goats the average age of AS cases (81.9 months, SE: 3.4) is statistically higher ( $p$ -value < 0.001) than in CS cases (50.4 months, SE: 0.21). When comparing sheep with goats, there is no difference in the average age for AS ( $p$ -value = 0.56), whereas the average age for CS in sheep was significantly lower than for goats ( $p$ -value < 0.01).

## A: Sheep



## B: Goats



AS: atypical scrapie; CS: classical scrapie.

**Figure 11:** Average age of TSE cases confirmed in sheep (A) and goats (B) by case type and year for the period 2002–2016

Tables 19 and 20 summarises by reporting country the number of cases subjected to discriminatory testing in 2016 for sheep and goats respectively. In sheep, 465 (82.6%) of the CS or unknown cases were submitted for discriminatory testing. In sheep, apart for a small proportion of inconclusive results (9 from IT), all scrapie cases submitted for discriminatory testing were confirmed as being CS cases (BSE excluded). In goats, 70 (11.3%) of the CS or unknown cases were submitted for discriminatory testing. All goat cases subjected to discriminatory testing (70) were CS cases.

**Table 19:** Number of cases subjected to discriminatory testing in sheep by reporting country, 2016

Sheep	Cases submitted for discriminatory testing										Cases not submitted for discriminatory testing or blank
	Country	Total cases	Nb of classical scrapie and unknown	Nb of atypical scrapie	BSE-not-excluded	BSE-excluded	Inconclusive	Total	% of Total classical TSE cases <sup>(a)</sup>	% of total atypical TSE cases <sup>(b)</sup>	
AT	1	0	1	0	0	0	0	0	100.0%	0.0%	1
BG	2	0	2	0	2	0	2	0	0.0%	100.0% <sup>(b)</sup>	0
CY	7	7	0	0	0	0	0	0	0.0%	0.0%	7
CZ	2	0	2	0	2	0	2	0	0.0%	100.0% <sup>(b)</sup>	0
DE	5	0	5	0	0	0	0	0	100.0%	0.0%	5
DK	1	0	1	0	0	0	0	0	100.0%	0.0%	1
EL	229	227	2	0	114	0	114	50.2%	0.0%	0.0%	115
ES	104	91	13	0	104	0	104	114.3%	100.0% <sup>(b)</sup>	0.0%	0
FI	2	0	2	0	0	0	0	0	100.0%	0.0%	2
FR	6	2	4	0	2	0	2	0	100.0%	0.0%	4
HU	23	0	23	0	0	0	0	0	100.0%	0.0%	23
IE	2	1	1	0	1	0	1	0	100.0%	0.0%	1
IT	148	143	5	0	139	9	148	103.4%	0.0%	0.0%	0
PL	8	0	8	0	0	0	0	0	100.0%	0.0%	8
PT	35	7	28	0	7	0	7	0	100.0%	0.0%	28
RO	75	75	0	0	75	0	75	0	100.0%	0.0%	0
SE	3	0	3	0	0	0	0	0	100.0%	0.0%	3
SI	3	0	3	0	0	0	0	0	100.0%	0.0%	3
SK	15	10	5	0	10	0	10	0	100.0%	0.0%	5
UK	14	0	14	0	0	0	0	0	100.0%	0.0%	14
<b>Total EU 28</b>	<b>685</b>	<b>563</b>	<b>122</b>	<b>0</b>	<b>456</b>	<b>9</b>	<b>465</b>	<b>82.6%</b>	<b>13.9%</b>	<b>0.0%</b>	<b>220</b>
IS	11	11	0	0	11	0	11	0	100.0%	0.0%	0
NO	14	0	14	0	0	0	0	0	100.0%	0.0%	14
<b>Total EFTA</b>	<b>25</b>	<b>11</b>	<b>14</b>	<b>0</b>	<b>11</b>	<b>0</b>	<b>11</b>	<b>100.0%</b>	<b>0.0%</b>	<b>0.0%</b>	<b>14</b>
<b>Total Sheep</b>	<b>710</b>	<b>574</b>	<b>136</b>	<b>0</b>	<b>467</b>	<b>9</b>	<b>476</b>	<b>82.9%</b>	<b>12.5%</b>	<b>0.0%</b>	<b>234</b>

Note: Only the reporting countries in which scrapie cases were detected in 2016 are mentioned in the table.

(a): Indicates the proportion of classical TSE cases that are submitted to discriminatory testing by each MS.

(b): Indicates the proportion of atypical TSE cases that are submitted to discriminatory testing by each MS; Bulgaria, the Czech Republic and Spain submitted, respectively, 2, 2 and 13 atypical scrapie cases for discriminatory testing.



**Table 20:** Number of cases subjected to discriminatory testing in goats by reporting country, 2016

Country	Total cases	Nb of classical scrapie and unknown	Nb of atypical scrapie	Cases submitted for discriminatory testing						Cases not submitted for discriminatory testing or blank
				BSE-not-excluded	BSE-excluded	Inconclusive	Total	% of Total classical TSE cases <sup>(a)</sup>	% of Total atypical TSE cases <sup>(b)</sup>	
<b>BG</b>	2	2	0	0	2	0	2	100.0%	0.0%	0
<b>CY</b>	570	570	0	0	13	0	13	2.3%	0.0%	557
<b>EL</b>	12	11	1	0	9	0	9	81.2%	0.0%	3
<b>ES</b>	24	19	5	0	24	0	24	126.3%	100.0%	0
<b>FR</b>	3	0	3	0	0	0	0	100.0%	0.0%	3
<b>IT</b>	11	8	3	0	11	0	11	137.5%	100.0%	0
<b>PT</b>	1	0	1	0	0	0	0	100.0%	0.0%	1
<b>RO</b>	3	3	0	0	3	0	3	100.0%	0.0%	0
<b>UK</b>	8	8	0	0	8	0	8	100.0%	0.0%	0
<b>Total EU 28</b>	<b>634</b>	<b>621</b>	<b>13</b>	<b>0</b>	<b>70</b>	<b>0</b>	<b>70</b>	<b>11.3%</b>	<b>61.5%</b>	<b>564</b>

Note: Only the reporting countries in which scrapie cases were detected in 2016 are mentioned in the table.

(a): Indicates the proportion of classical TSE cases that are submitted to discriminatory testing by each MS.

(b): Indicates the proportion of atypical TSE cases that are submitted to discriminatory testing by each MS; Spain and Italy submitted respectively 5 and 3 atypical scrapie cases for discriminatory testing.

### 3.2.1. Genotyping in sheep

In sheep, according to Regulation (EC) 999/2001 Annex III, Chapter A, Part I, point 8, the genotype, and, where possible, the breed of each positive case (according to point 8.1) should be determined. A random sample of ovine animals (according to point 8.2) should also be genotyped.

The classification of genotypes according to Great Britain's National Scrapie Plan (NSP) is summarised in Table 21. A three-tier classification is also reported, as described in Section 2, in order to show the evolution of the genotype distribution in sheep populations within the EU.

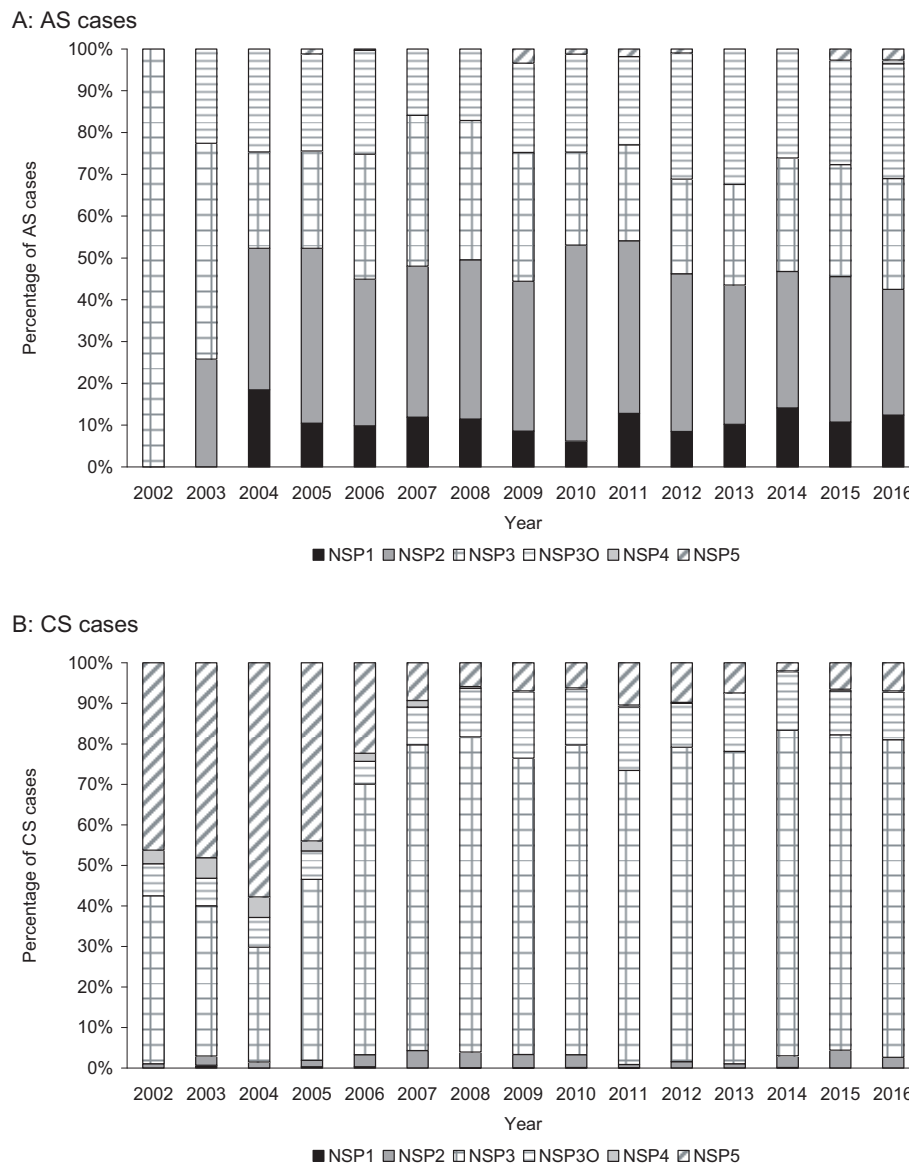
**Table 21:** Classification of the genotypes of the sheep *PRNP* gene according to Great Britain's National Scrapie Plan (NSP) and the three-tiers of report groups

NSP group	Genotype	Comment	Report group
NSP1	ARR/ARR	Genetically most resistant	Resistant
NSP2	ARR/ARQ; ARR/ARH; ARR/AHQ	Genetically resistant	Semi-resistant
NSP3	ARQ/ARQ	Genetically little resistant (ARQ/ARQ may be scientifically reviewed)	Susceptible
NSP3 Other (NSP3O)	AHQ/AHQ; ARH/ARH; ARH/ARQ; AHQ/ARH; AHQ/ARQ		Susceptible
NSP4	ARR/VRQ	Genetically susceptible	Susceptible
NSP5	ARQ/VRQ; ARH/VRQ; AHQ/VRQ; VRQ/VRQ	Genetically highly susceptible	Susceptible

Table 22 shows the genotypes of ovine scrapie cases in 2016 in the EU and other reporting countries.

**Table 22:** Distribution of genotypes of confirmed scrapie cases in sheep in 2016 by reporting country and National Scrapie Plan (NSP) group

NSP group	Atypical scrapie								Classical scrapie							
	NSP1	NSP2	NSP3	NSP30	NSP4	NSP5	Unknown	Total	NSP1	NSP2	NSP3	NSP30	NSP4	NSP5	Unknown	Total
AT	0	1	0	0	0	0	0	<b>1</b>	0	0	0	0	0	0	0	<b>0</b>
BG	0	0	0	0	0	0	2	<b>2</b>	0	0	0	0	0	0	0	<b>0</b>
CY	0	0	0	0	0	0	0	<b>0</b>	0	5	1	0	0	0	1	<b>7</b>
CZ	0	0	1	0	0	0	1	<b>2</b>	0	0	0	0	0	0	0	<b>0</b>
DE	0	1	0	0	0	0	2	<b>3</b>	0	0	0	0	0	0	0	<b>0</b>
DK	0	0	0	0	0	0	1	<b>1</b>	0	0	0	0	0	0	0	<b>0</b>
EL	0	1	0	1	0	0	0	<b>2</b>	0	3	165	42	1	13	3	<b>227</b>
ES	3	2	4	3	0	0	1	<b>13</b>	0	1	83	1	0	1	5	<b>91</b>
FI	0	0	1	1	0	0	0	<b>2</b>	0	0	0	0	0	0	0	<b>0</b>
FR	0	0	2	0	0	0	2	<b>4</b>	0	0	1	0	0	1	0	<b>2</b>
HU	3	13	3	4	0	0	0	<b>23</b>	0	0	0	0	0	0	0	<b>0</b>
IE	0	0	1	0	0	0	0	<b>1</b>	0	1	0	0	0	0	0	<b>1</b>
IT	0	1	2	2	0	0	0	<b>5</b>	0	0	117	16	0	1	0	<b>134</b>
PL	1	4	1	1	0	0	1	<b>8</b>	0	0	0	0	0	0	0	<b>0</b>
PT	2	3	10	4	1	2	6	<b>28</b>	0	0	5	0	0	1	1	<b>7</b>
RO	0	0	0	0	0	0	0	<b>0</b>	0	4	50	3	1	17	0	<b>75</b>
SE	0	0	0	0	0	0	3	<b>3</b>	0	0	0	0	0	0	0	<b>0</b>
SI	1	0	2	0	0	0	0	<b>3</b>	0	0	0	0	0	0	0	<b>0</b>
SK	1	2	1	1	0	0	0	<b>5</b>	0	1	5	1	0	3	0	<b>10</b>
UK	1	2	0	9	0	0	2	<b>14</b>	0	0	0	0	0	0	0	<b>0</b>
<b>Total EU 28</b>	<b>12</b>	<b>30</b>	<b>28</b>	<b>26</b>	<b>1</b>	<b>2</b>	<b>21</b>	<b>120</b>	<b>0</b>	<b>15</b>	<b>427</b>	<b>63</b>	<b>2</b>	<b>37</b>	<b>10</b>	<b>554</b>
CH	0	0	0	0	0	0	0	<b>0</b>	0	0	0	0	0	0	0	<b>0</b>
IS	0	0	0	0	0	0	0	<b>0</b>	0	0	9	0	0	2	0	<b>11</b>
NO	2	4	2	5	0	0	1	<b>14</b>	0	0	0	0	0	0	0	<b>0</b>
<b>Total EFTA</b>	<b>2</b>	<b>4</b>	<b>2</b>	<b>5</b>	<b>0</b>	<b>0</b>	<b>1</b>	<b>14</b>	<b>0</b>	<b>0</b>	<b>9</b>	<b>0</b>	<b>0</b>	<b>2</b>	<b>0</b>	<b>11</b>
<b>Total</b>	<b>14</b>	<b>34</b>	<b>30</b>	<b>31</b>	<b>1</b>	<b>2</b>	<b>22</b>	<b>134</b>	<b>0</b>	<b>15</b>	<b>436</b>	<b>63</b>	<b>2</b>	<b>39</b>	<b>10</b>	<b>565</b>



(A) Atypical scrapie. (B) Classical scrapie.

**Figure 12:** Frequency distribution of genotypes of sheep scrapie cases by case type, year and NSP group in the period 2002–2016 in the reporting countries

In total, 529 (97.2%) of the 544 cases of CS in sheep with known genotype reported in the EU in 2016 (Table 22) were from the susceptible genotype groups (NSP3, NSP3O, NSP4 and NSP5), consistent with the pattern observed in all cases reported since 2002 (Figure 12). For AS, the same genotype groups accounted for 57.6% of all cases.

Table 23 shows the genotypes obtained in 2016 from the random samples of tested sheep in the reporting countries. In the EU, excluding data from Cyprus (where genotyping is conducted systematically in the sheep population), 26.6% of the sheep population is susceptible to CS (i.e. belongs to groups NSP3, NSP3O, NSP4 or NSP5).

Considering the past 5 years of random sampling, the resistant genotype group (NSP1; black colour in the bars of Figure 13) accounts for between 29.8% (in 2012) and 50.6% (in 2015) of the total number of sheep genotyped (Table 22 and Figure 13).

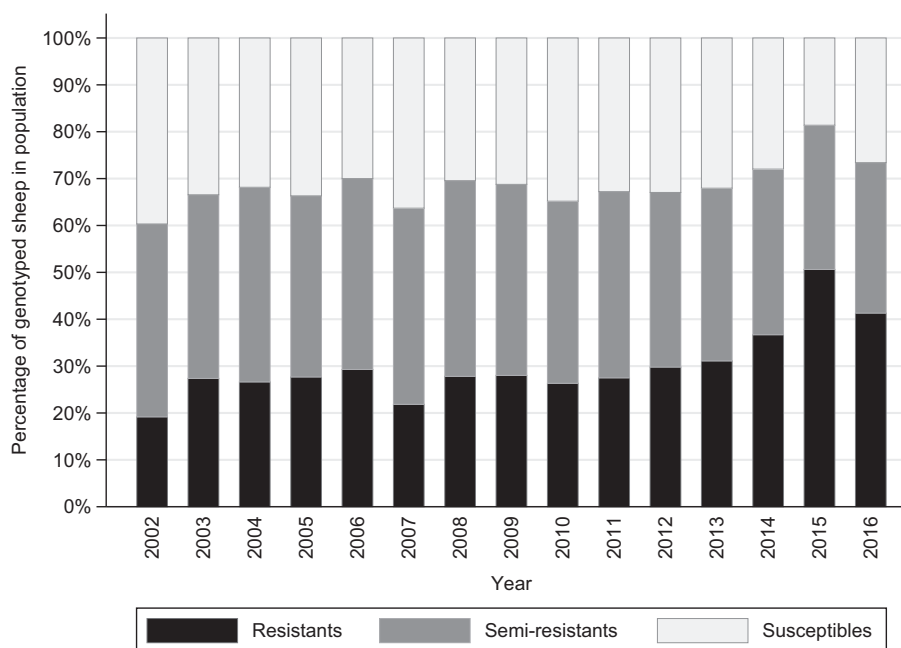
**Table 23:** Distribution of genotypes in randomly selected sheep in the EU and other reporting countries in 2016 by Member State and National Scrapie Plan (NSP) group, in accordance with Regulation (EC) 999/2001 Annex III, Chapter A, Part I, point 8.2

Country	Number of genotyped animals (% of sample within country)							Total
	NSP1	NSP2	NSP3	NSP30	NSP4	NSP5	Other	
AT	10 (9.4%)	39 (36.8%)	40 (37.7%)	11 (10.3%)	1 (1.0%)	3 (2.8%)	2 (1.8%)	106
BE	159 (63.9%)	57 (22.9%)	13 (5.2%)	7 (2.8%)	2 (0.8%)	4 (1.6%)	7 (2.8%)	249
BG	–	–	–	–	–	–	–	–
CY <sup>(a)</sup>	47,775 (86.5%)	6,099 (11.0%)	280 (0.5%)	118 (0.2%)	432 (0.8%)	45 (0.1%)	464 (0.8%)	55,213
CZ	6 (22.2%)	13 (48.1%)	6 (22.2%)	1 (3.7%)	0 (0.0%)	1 (3.7%)	0 (0.0%)	27
DE	1,268 (76.2%)	261 (15.7%)	75 (4.5%)	15 (0.9%)	41 (2.5%)	1 (0.1%)	2 (0.1%)	1,663
DK	36 (36.0%)	15 (15.0%)	28 (28.0%)	13 (13.0%)	0 (0.0%)	8 (8.0%)	0 (0.0%)	100
EE	30 (30.0%)	45 (45.0%)	10 (10.0%)	5 (5.0%)	5 (5.0%)	5 (5.0%)	0 (0.0%)	100
EL	47 (13.1%)	112 (31.3%)	121 (33.8%)	38 (10.6%)	8 (2.2%)	12 (3.4%)	20 (5.6%)	358
ES	76 (11.6%)	263 (40.1%)	218 (33.2%)	44 (6.7%)	7 (1.1%)	22 (3.4%)	26 (4.0%)	656
FI	2 (2.0%)	25 (25.0%)	45 (45.0%)	11 (11.0%)	4 (4.0%)	13 (13.0%)	0 (0.0%)	100
FR	315 (49.5%)	184 (28.9%)	55 (8.6%)	5 (0.8%)	8 (1.3%)	13 (2.0%)	57 (8.9%)	637
HR	13 (8.7%)	56 (37.3%)	59 (39.3%)	22 (14.7%)	0 (0.0%)	0 (0.0%)	0 (0.0%)	150
HU	357 (59.5%)	186 (31.0%)	24 (4.0%)	21 (3.5%)	6 (1.0%)	6 (1.0%)	0 (0.0%)	600
IE	249 (37.3%)	256 (38.3%)	65 (9.7%)	45 (6.7%)	31 (4.6%)	22 (3.3%)	0 (0.0%)	668
IT	119 (19.5%)	269 (44.1%)	175 (28.7%)	25 (4.1%)	8 (1.3%)	10 (1.6%)	4 (0.7%)	610
LT	29 (29.6%)	40 (40.8%)	23 (23.5%)	4 (4.1%)	0 (0.0%)	1 (1.0%)	1 (1.0%)	98
LU	63 (63.0%)	21 (21.0%)	4 (4.0%)	2 (2.0%)	5 (5.0%)	5 (5.0%)	0 (0.0%)	100
LV	18 (16.5%)	49 (45.0%)	38 (34.9%)	2 (1.8%)	0 (0.0%)	2 (1.8%)	0 (0.0%)	109
MT	–	–	–	–	–	–	–	–
NL	562 (64.2%)	216 (24.7%)	31 (3.5%)	40 (4.6%)	17 (1.9%)	10 (1.1%)	0 (0.0%)	876
PL	43 (38.1%)	46 (40.7%)	16 (14.2%)	3 (2.7%)	3 (2.7%)	2 (1.8%)	0 (0.0%)	113
PT	105 (16.9%)	244 (39.3%)	185 (29.8%)	48 (7.7%)	16 (2.6%)	23 (3.7%)	0 (0.0%)	621
RO	55 (10.2%)	225 (41.6%)	174 (32.2%)	40 (7.4%)	14 (2.6%)	33 (6.1%)	0 (0.0%)	541
SE	10 (10.0%)	13 (13.0%)	66 (66.0%)	3 (3.0%)	1 (1.0%)	7 (7.0%)	0 (0.0%)	100
SI	6 (4.6%)	45 (34.4%)	60 (45.8%)	13 (9.9%)	2 (1.5%)	5 (3.8%)	0 (0.0%)	131

Country	Number of genotyped animals (% of sample within country)							Total
	NSP1	NSP2	NSP3	NSP3O	NSP4	NSP5	Other	
<b>SK</b>	32 (32.0%)	42 (42.0%)	12 (12.0%)	4 (4.0%)	7 (7.0%)	3 (3.0%)	0 (0.0%)	100
<b>UK</b>	228 (38.0%)	261 (43.5%)	42 (7.0%)	54 (9.0%)	7 (1.2%)	8 (1.3%)	0 (0.0%)	600
<b>Total EU 28</b>	<b>51,613 (79.9%)</b>	<b>9,082 (14.1%)</b>	<b>1,865 (2.9%)</b>	<b>594 (0.9%)</b>	<b>625 (1.0%)</b>	<b>264 (0.4%)</b>	<b>583 (0.9%)</b>	<b>64,626</b>
<b>CH</b>	–	–	–	–	–	–	–	–
<b>IS</b>	0 (0.0%)	0 (0.0%)	212 (70.0%)	51 (16.8%)	0 (0.0%)	40 (13.2%)	0 (0.0%)	303
<b>NO</b>	69 (11.7%)	228 (38.6%)	106 (18.0%)	103 (17.5%)	37 (6.3%)	47 (8.0%)	0 (0.0%)	590
<b>Total EFTA</b>	<b>69 (7.7%)</b>	<b>228 (25.5%)</b>	<b>318 (35.6%)</b>	<b>154 (17.2%)</b>	<b>37 (4.1%)</b>	<b>87 (9.7%)</b>	<b>0 (0.0%)</b>	<b>893</b>
<b>Total</b>	<b>51,682 (78.9%)</b>	<b>9,310 (14.2%)</b>	<b>2,183 (3.3%)</b>	<b>748 (1.1%)</b>	<b>662 (1.0%)</b>	<b>351 (0.5%)</b>	<b>583 (0.9%)</b>	<b>65,516</b>

–: no genotyping performed in 2016.

(a): The Cyprus data are different from those of other Member States as CY systematically genotypes the breeding sheep population.



Data from Cyprus were excluded.

NSP1: Resistant (black); NSP2: Semi-Resistant (grey); NSP3+NSP3O+NSP4+NSP5: Susceptible (white). (As referred to in Table 21).

**Figure 13:** Frequency distribution of the three genotype groups in sheep sampled for genotyping in the EU in the period 2002–2016 according to Regulation (EC) 999/2001 Annex III, Chapter A, Part I, point 8.2

### 3.3. Other species

In 2016, 2,712 cervids (Table 24) were tested by seven MSs, with Romania accounting for more than 90% (2,500 wild cervids were tested). The animals were almost exclusively from wild populations even though no information is available on the design of the surveillance carried out. All animals tested negative.

In 2016, the first case of CWD in Europe was detected in a Norwegian free-ranging reindeer (Benestad et al., 2016). In April and May of 2016, Norway confirmed two additional cases of CWD, one in a wild reindeer and one in a wild moose. In light of this emerging issue, NO intensified its monitoring for CWD in wild and captive cervids and tested during 2016 10,139 deer. The total number of cases confirmed in 2016 was 5: 3 in wild Eurasian tundra reindeer and 2 in wild moose.

In parallel with the monitoring of cervids in 2016, four MSs (DK, EE, FI and HU) reported results on samples tested for TSE in species other than ruminants. In total, 490 samples were collected from the following species: cat (*Felis catus*) (76.9%), mink (*Mustela lutreola*) (13.9%), fox (genus *Vulpes*) (7.1%) and racoon dogs (*Nyctereutes procyonoides*) (2%). None tested positive (Table 25).

**Table 24:** Number of cervids tested in the reporting countries in 2016 by species, management system and country (number of positives for CWD in brackets)

Country	Wild deer									Domesticated/farmed deer								Total
	Eurasian tundra reindeer ( <i>Rangifer tarandus tarandus</i> )	Finnish (Eurasian) forest reindeer ( <i>Rangifer tarandus fennicus</i> )	Moose (or Eurasian / European elk) ( <i>Alces alces alces</i> )	Roe deer ( <i>Capreolus capreolus</i> )	White-tailed deer ( <i>Odocoileus virginianus</i> )	Red deer ( <i>Cervus elaphus</i> )	Fallow deer ( <i>Dama dama</i> )	Other or Unknown	Total	Eurasian tundra reindeer ( <i>Rangifer tarandus tarandus</i> )	Finnish (Eurasian) forest reindeer ( <i>Rangifer tarandus fennicus</i> )	Moose (or Eurasian / European elk) ( <i>Alces alces alces</i> )	Roe deer ( <i>Capreolus capreolus</i> )	White-tailed deer ( <i>Odocoileus virginianus</i> )	Red deer ( <i>Cervus elaphus</i> )	Fallow deer ( <i>Dama dama</i> )	Total	
AT	0	0	0	0	0	1	0	0	1	0	0	0	0	0	1	0	1	2
EE	0	0	1	0	0	0	0	0	1	0	0	0	0	0	0	0	0	1
FI	0	5	26	7	12	0	0	0	50	6	0	0	0	0	0	0	6	56
HU	0	0	0	6	0	1	1	1	9	0	0	0	0	0	0	0	0	9
IT	0	0	0	49	0	24	1	0	74	0	0	0	0	0	0	0	0	74
RO	0	0	0	0	0	459	7	2,034	2,500	0	0	0	0	0	0	0	0	2,500
SE	0	0	60	7	0	3	0	0	70	0	0	0	0	0	0	0	0	70
<b>Total EU</b>	<b>0</b>	<b>5</b>	<b>87</b>	<b>69</b>	<b>12</b>	<b>488</b>	<b>9</b>	<b>2,035</b>	<b>2,705</b>	<b>6</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>1</b>	<b>0</b>	<b>7</b>	<b>2,712</b>
NO	842 (3) <sup>(a)</sup>	0	4,403 (2) <sup>(a)</sup>	473	0	2,451	0	88	8,257	1,739	0	0	0	0	143	0	1,882	10,139
IS	13	0	0	0	0	0	0	0	13	1	0	0	0	0	0	0	1	14
<b>Total EFTA</b>	<b>855 (3)</b>	<b>0</b>	<b>4,403 (2)</b>	<b>473</b>	<b>0</b>	<b>2,451</b>	<b>0</b>	<b>0</b>	<b>8,270 (5)</b>	<b>1,740</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>143</b>	<b>0</b>	<b>1,883</b>	<b>10,153</b>
<b>Total</b>	<b>855 (3)</b>	<b>5</b>	<b>4,490 (2)</b>	<b>542</b>	<b>12</b>	<b>2,939</b>	<b>9</b>	<b>2,123</b>	<b>10,975 (5)</b>	<b>1,746</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>144</b>	<b>0</b>	<b>1,890</b>	<b>12,865</b>

(a): 3 out of 842 tested wild Eurasian tundra reindeer were found positive as well as 2 out of 4,403 tested wild moose.



**Table 25:** Numbers of tests<sup>(a)</sup> for TSE in species other than ruminants in EU Member States in 2016

Country	Wild ruminants	Raccoon dogs ( <i>Nyctereutes procyonoides</i> )	Fox (genus <i>Vulpes</i> )	Minks ( <i>Mustela lutreola</i> )	Cats ( <i>Felis catus</i> )	Total
DK	0	0	0	0	7	7
EE	0	0	0	14	0	14
FI	0	10	35	54	59	158
HU	0	0	0	0	311	311
<b>Total EU</b>	0	10	35	68	<b>377</b>	490

(a): All tests were negative.

## 4. Conclusions

Since 2001, ca.115 million cattle have been tested for BSE in the EU, with a sustained decrease in the number of animals tested over time following amendments to the TSE Regulation. In 2016, despite the 5% reduction in the number of cattle tested, the number of samples tested from risk animals (AM + ES + FS) over 48 months of age showed an increase (+ 28,118) compared with 2015.

The decline of C-BSE has been consistent over the past decade in terms of the absolute number of cases and the proportion of cases in tested animals. Focusing on the last 10 years, the decreasing trend for C-BSE is statistically significant, whereas no significant trend (either increasing or decreasing) is observed for H-BSE or L-BSE.

In 2016, only two MSs (France and Spain) reported cases of BSE: five in total. Of the five cases that were reported, four were atypical BSE (all H-type), three from FR and one from ES. The fifth case was a C-BSE detected in France in a beef cow born in 2011, i.e. 10 years after the EU-wide reinforced feed ban of 2001 (BARB case), reported as fallen stock. For the first time since BSE cases have been reported, no C-BSE or atypical BSE cases were reported by the United Kingdom in 2016.

Since 2002, ca. 8.8 million small ruminants have been tested as part of the EU-wide TSE surveillance. There have been a 5% and 11% reduction in the number of sheep and goats tested in 2016, respectively. The reduction was observed in reported 'non-infected flocks' category since the total number of small ruminants tested in the 'infected flocks' category remained stable at around 15,000 sheep and 4,500 goats. The requirements for testing a minimum number of samples from the SHC and NSHC surveillance target groups in the small ruminant populations were fulfilled by 23 MSs for sheep and by 17 MSs for goats.

In 2016, four countries (Greece, Spain, Italy and Romania) accounted for 95% of all CS sheep cases and 81.2% of all cases. CS was reported in sheep by 9 MSs and one non-MS, whereas AS was reported in sheep by 18 MSs and one non-MSs.

Cyprus accounted for 90% of all goat cases in 2016. Despite the large caseload from this MS, an improvement in the scrapie situation in Cyprus compared with 2015 is suggested by the 40% decrease in the total number of classical scrapie cases in 2016 (from 923 to 570) and in the number of index cases (from 18 to 12). CS was reported in goats by seven MSs and one non-MS, whereas AS was reported by five MSs and one non-MSs. The average age of AS cases is statistically significantly higher than that of CS cases in both sheep and goats ( $p < 0.001$ ). There is no significant difference in the average age for AS between sheep and goats ( $p = 0.56$ ) but the average age for CS in sheep was significantly lower than for goats.

In the EU, the occurrence of scrapie in 2016 was similar to 2015. Using the number of index cases as a proxy for incidence, there has not been a major change. In sheep, the number of CS index cases in 2016 was 112 compared with 109 in 2015, and the number of index cases of AS slightly decreased from 120 in 2015 to 106 in 2016. In goats, the number of CS index cases in 2016 was 30 compared with 37 in 2015, whereas the number of AS index cases was unchanged at 13. Focusing on the last 10 years in the EU, scrapie in sheep has remained quite stable overall for both CS and AS, ranging between 2 and 6 cases per 10,000 tested animals. Overall and for all MS, there has not been a significant trend in ovine CS and AS during that period ( $p$ -values of 0.2 and 0.51, respectively). Focusing on the last 10 years in the EU in goats, a slightly but significant increasing trend in both CS and AS ( $p$ -values of 0.001 and 0.03 respectively) has been observed. However, the last four years a decrease was observed in number of scrapie cases in goats. The individual national trends for scrapie in small ruminants may show different patterns compared with the overall trend in the EU. In absolute numbers, CS is still the most prevalent type of TSE in the European small ruminant population.

With regard to the occurrence of CS compared with that of AS in sheep at MS level in the period 2006–2016, three different patterns can be observed: (a) in nine MSs (Austria, Belgium, Croatia, Denmark, Estonia, Finland, Poland, Portugal and Sweden), the occurrence of CS was always lower than that of AS (the ratio CS:AS was always below one), either because there were more AS than CS cases, or because CS was never reported; (b) in 10 MSs (Bulgaria, the Czech Republic, Germany, France, Hungary, Ireland, the Netherlands, Slovenia, Slovakia and the United Kingdom), the ratio CS:AS was decreasing; (c) in five MSs (Cyprus, Greece, Spain, Italy and Romania), the ratio CS:AS was always above one because there were more CS than AS cases.

The genotyping data collected from the 2016 ovine CS cases consistently confirms the association between the occurrence of the disease and the susceptible genotypes, with 97.2% of the cases having susceptible genotypes (NSP3, NSP30, NSP4 or NSP5). In 2016, the genotyping data from random samples of the EU sheep population (excluding Cyprus) showed that 26.6% of the genotyped sheep held genotypes of the susceptible group.

In 2016, CWD was detected for the first time in Europe, in Norway (Benestad et al., 2016; EFSA BIOHAZ Panel, 2017b), where 10,139 cervids were tested and 5 cases confirmed in two different species: 3 in wild reindeer and 2 in moose. In 2016, 2,712 cervids were tested in seven MSs, but most of them in a single MS (2,500 in Romania), all with negative results. A total of 490 samples from non-ruminant species were also collected and tested as part of TSE surveillance, all of them with negative results.

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## Abbreviations

AM	<i>Ante-mortem</i>
AS	atypical scrapie
BARB	Born After the Revised feed Ban
BSE	bovine spongiform encephalopathy
C-BSE	classical bovine spongiform encephalopathy
CS	classical scrapie
CWD	chronic wasting disease
EFTA	European Free Trade Association
ELISA	enzyme-linked immunosorbent assay
EM	eradication MEASURES
ES	emergency slaughtered
EURL	European Union Reference Laboratory
EUSR	European summary report
FS	fallen stock

H-BSE	H-type bovine spongiform encephalopathy
IHC	immunohistochemistry
HS	healthy slaughtered
L-BSE	L-type bovine spongiform encephalopathy
MS	Member State
NRL	National Reference Laboratories
NSHC	not slaughtered for human consumption
NSP	National Scrapie Plan
NUTS	Nomenclature of Units for Territorial Statistics
OIE	World Organisation for Animal Health
SHC	slaughtered for human consumption
SAF	scrapie-associated fibrils
SU	BSE suspect
TSE	transmissible spongiform encephalopathy
WB	Western Blot

### Country codes

Austria	AT	Greece	EL	Poland	PL
Belgium	BE	Hungary	HU	Portugal	PT
Bulgaria	BG	Iceland	IS	Romania	RO
Croatia	HR	Ireland	IE	Slovakia	SK
Cyprus	CY	Italy	IT	Slovenia	SI
Czech Republic	CZ	Latvia	LV	Spain	ES
Denmark	DK	Lithuania	LT	Sweden	SE
Estonia	EE	Luxembourg	LU	Switzerland	CH
Finland	FI	Malta	MT	United Kingdom	UK
France	FR	Netherlands	NL		
Germany	DE	Norway	NO		

MS countries: AT;BE;BG;HR;CY;CZ;DK;EE;FI;FR;DE;EL;HU;IE;IT;LV;LT;LU;MT;NL;PL;PT;RO;SK;SI;ES;SE;UK.  
 EFTA Countries (non-MS countries): CH (including Lichtenstein); IS; NO.

## Appendix A – Number of tested bovine animals by country, age group and different surveillance target groups in EU Member States and EFTA-reporting countries during 2016

**Table A.1:** Bovine animals tested by age group in the EU Member States and EFTA reporting countries during 2016

EU/ EFTA group	Country code	Age group (months)														Total	
		< 24	24–29	30–35	36–47	48–59	60–71	72–83	84–95	96–107	108– 119	120– 131	132– 143	144–155	> 155		Unknown
EU	AT	0	303	316	632	2,986	3,179	2,922	2,533	1,995	1,461	1,097	776	571	1,143	0	19,914
	BE	4	6	6	18	7,424	6,043	4,609	3,076	1,819	1,100	580	364	205	321	261	25,836
	BG	0	287	4,335	2,583	2,391	2,046	1,624	1,557	1,157	1,023	1,139	805	598	1,313	0	20,858
	CY	0	0	0	1	217	213	168	121	95	43	20	20	10	8	0	916
	CZ	11	712	557	1,121	3,131	2,710	1,995	1,441	1,051	776	490	378	256	887	0	15,516
	DE	157	162	201	842	39,063	37,034	28,871	20,155	13,150	7,928	4,870	2,839	1,830	4,204	79	161,385
	DK	17	0	4	34	6,821	5,472	3,673	2,014	1,025	533	311	192	116	357	127	20,696
	EE	0	0	1	15	1,055	944	669	393	228	120	84	25	16	23	0	3,573
	EL	6	5	75	110	287	263	2,323	1,998	1,529	1,152	790	649	579	2,335	4	12,105
	ES	4	1	9	13	12,018	10,517	8,286	6,319	4,752	3,472	2,500	2,089	2,116	10,723	0	62,819
	FI	4	2	3	13	3,035	2,891	2,132	1,272	680	403	240	173	132	250	4	11,234
	FR	325	529	1,160	4,247	40,136	37,125	30,847	24,066	18,723	13,920	10,542	8,106	6,118	56,092	4,224	256,160
	HR	33	397	1,155	1,982	2,560	2,204	2,188	1,989	1,340	1,089	814	754	523	1,158	425	18,611
	HU	34	1,261	950	2,354	2,203	1,792	1,294	856	569	399	399	305	263	895	0	13,574
	IE	33	24	35	218	9,042	8,221	7,593	7,142	6,039	5,213	4,353	3,447	2,442	5,563	0	59,365
	IT	12	11	108	130	13,926	11,780	8,665	5,985	4,067	2,704	1,660	1,112	768	1,915	2	52,845
	LT	0	0	0	0	764	544	496	371	288	234	150	120	78	125	0	3,170
	LU	0	0	0	0	562	549	431	287	196	130	87	41	40	87	0	2,410
	LV	26	303	258	464	515	380	313	239	196	128	68	35	30	29	0	2,984
	MT	0	0	0	1	43	40	38	14	20	12	4	1	2	0	0	175
	NL	20	38	53	270	11,283	12,625	10,499	8,014	5,077	2,939	1,639	911	497	735	288	54,888
	PL	5	2	2	4	8,132	7,140	5,951	4,600	3,427	39,769	31,214	23,529	15,172	30,240	0	169,187
	PT	0	0	1	0	2,926	2,483	2,271	1,859	1,439	1,230	1,070	713	729	4,958	0	19,679
	RO	281	399	12,340	20,452	17,456	14,666	13,312	12,196	11,390	11,119	11,566	11,463	8,225	30,006	0	174,871
	SE	33	94	137	617	1,914	1,977	1,628	1,019	554	290	196	145	88	255	13	8,960
	SI	8	14	57	26	1,176	1,161	1,013	847	637	413	267	196	135	289	0	6,239

EU/ EFTA group	Country code	Age group (months)															Total
		< 24	24–29	30–35	36–47	48–59	60–71	72–83	84–95	96–107	108–119	120–131	132–143	144–155	> 155	Unknown	
	SK	1	616	643	1,300	1,358	1,037	817	602	363	300	212	149	77	199	0	7,674
	UK	2	78	161	797	25,605	24,389	21,223	17,935	13,850	10,361	7,899	5,904	4,779	13,720	235	146,938
	Total	1,016	5,244	22,567	38,244	218,029	199,425	165,851	128,900	95,656	108,261	84,261	65,241	46,395	167,830	5,662	1,352,582
EFTA	CH	12	30	50	384	1,607	1,977	1,966	1,743	1,301	882	597	392	319	368	0	11,628
	IS	1	0	0	1	22	4	6	28	16	49	8	4	0	0	1	140
	NO	14	27	26	137	2,004	1,783	1,203	713	404	207	92	67	43	62	145	6,927
	Total	27	57	76	522	3,633	3,764	3,175	2,484	1,721	1,138	697	463	362	430	146	18,695

**Table A.2:** Bovine animals at risk (animals with clinical signs at *ante-mortem*, animals culled under BSE eradication measures, emergency slaughtered and fallen stock) tested by age group in EU Member States and in EFTA-reporting countries during 2016

EU/ EFTA group	Country code	Age group (months)															Total
		< 24	24–29	30–35	36–47	48–59	60–71	72–83	84–95	96–107	108–119	120–131	132–143	144–155	> 155	Unknown	
EU	AT	0	302	177	276	2,534	2,745	2,499	2,136	1,730	1,238	903	607	457	940	0	16,544
	BE	2	2	5	15	7,414	6,036	4,607	3,070	1,813	1,076	579	363	205	321	259	25,767
	BG	0	287	278	217	215	175	177	177	159	146	147	93	86	155	0	2,312
	CY	0	0	0	1	217	213	168	121	95	43	20	20	10	8	0	916
	CZ	5	712	556	1,120	3,130	2,709	1,994	1,438	1,048	773	487	373	253	880	0	15,478
	DE	106	155	198	829	38,862	36,857	28,710	20,052	13,073	7,869	4,833	2,811	1,818	4,137	78	160,388
	DK	16	0	3	32	6,817	5,469	3,671	2,011	1,023	532	311	192	115	355	127	20,674
	EE	0	0	1	15	1,055	944	669	393	228	120	84	25	16	23	0	3,573
	EL	2	2	7	36	246	241	479	407	281	205	165	124	127	521	3	2,846
	ES	2	0	6	10	11,982	10,479	8,254	6,297	4,739	3,454	2,489	2,084	2,112	10,606	0	62,514
	FI	4	2	3	13	3,035	2,891	2,132	1,271	680	403	240	173	131	248	4	11,230
	FR	253	512	1,144	4,203	39,545	36,617	30,438	23,762	18,476	13,728	10,404	8,005	6,040	17,561	3,873	214,561
	HR	25	379	246	485	1,059	881	817	741	475	335	214	166	114	246	100	6,283
	HU	32	1,259	886	2,210	2,058	1,688	1,176	753	460	305	266	164	142	468	0	11,867
	IE	33	24	35	218	9,042	8,221	7,593	7,142	6,039	5,213	4,353	3,447	2,442	5,563	0	59,365
	IT	11	11	16	65	13,899	11,745	8,636	5,953	4,015	2,597	1,610	1,072	746	1,870	2	52,248

EU/ EFTA group	Country code	Age group (months)															Total
		< 24	24–29	30–35	36–47	48–59	60–71	72–83	84–95	96–107	108–119	120–131	132–143	144–155	> 155	Unknown	
EU	LT	0	0	0	0	764	544	496	371	288	234	150	120	78	125	0	3,170
	LU	0	0	0	0	561	549	431	287	195	130	87	41	40	87	0	2,408
	LV	25	303	258	464	515	380	312	238	196	128	68	35	30	29	0	2,981
	MT	0	0	0	1	43	39	38	14	20	12	4	1	2	0	0	174
	NL	20	38	53	270	11,278	12,622	10,498	8,013	5,075	2,939	1,638	911	496	734	282	54,867
	PL	1	0	0	1	8,129	7,139	5,950	4,600	3,425	2,553	1,862	1,263	859	1,560	0	37,342
	PT	0	0	0	0	2,926	2,483	2,154	1,766	1,366	1,174	1,029	682	699	4,796	0	19,075
	RO	60	376	362	624	608	447	456	348	294	259	242	238	168	597	0	5,079
	SE	33	94	137	616	1,914	1,977	1,628	1,019	554	289	196	145	88	255	12	8,957
	SI	5	13	5	14	1,172	1,159	1,013	842	634	411	265	195	134	285	0	6,147
	SK	1	616	643	1,300	1,358	1,037	817	602	363	300	212	149	77	199	0	7,674
	UK	2	78	161	796	25,598	24,383	21,223	17,935	13,847	10,361	7,899	5,904	4,779	13,720	235	146,920
Total		638	5,165	5,180	13,831	195,976	180,670	147,036	111,759	80,591	56,827	40,757	29,403	22,264	66,289	4,975	961,370
EFTA	CH	12	29	50	379	1,600	1,977	1,956	1,737	1,300	878	596	392	319	368	0	11,593
	IS	1	0	0	1	0	4	3	1	1	0	1	0	0	0	1	13
	NO	14	27	26	136	2,004	1,783	1,203	713	404	207	92	67	43	62	145	6,926
	Total	27	56	76	516	3,604	3,764	3,162	2,451	1,705	1,085	689	459	362	430	146	18,532

**Table A.3:** Healthy slaughtered bovine animals tested by age group in EU Member States and in EFTA-reporting countries during 2016

EU/ EFTA group	Country code	Age group (months)															Total		
		< 24	> 48	> 72	24–29	30–35	36–47	48–59	60–71	72–83	84–95	96–107	108–119	120–131	132–143	144–155		> 155	Unknown
EU	AT	0	0	0	0	138	355	450	431	422	395	263	223	194	169	114	201	0	3,355
	BE	0	0	0	0	1	1	4	2	1	2	4	24	1	0	0	0	2	42
	BG	0	0	0	0	4,057	2,366	2,176	1,871	1,447	1,380	998	877	992	712	512	1,158	0	18,546
	CY	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
	CZ	6	0	0	0	1	1	0	0	1	3	3	3	3	5	3	7	0	36
	DE	51	0	0	7	3	10	95	87	77	54	35	29	22	18	6	41	1	536
	DK	1	0	0	0	1	2	4	3	2	3	2	1	0	0	0	2	0	21

EU/ EFTA group	Country code	Age group (months)															Total		
		< 24	> 48	> 72	24-29	30-35	36-47	48-59	60-71	72-83	84-95	96-107	108-119	120-131	132-143	144-155		> 155	Unknown
	EE	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
	EL	4	0	0	3	68	74	40	21	1,844	1,590	1,247	945	625	525	452	1,814	1	9,253
	ES	1	0	0	1	3	3	36	38	32	22	13	18	11	4	4	116	0	302
	FI	0	0	0	0	0	0	0	0	0	1	0	0	0	0	1	2	0	4
	FR	72	0	0	17	15	44	591	507	408	304	247	191	137	101	78	38,531	351	41,594
	HR	7	0	0	18	909	1,497	1,501	1,323	1,371	1,248	865	754	600	588	409	912	325	12,327
	HU	2	0	0	2	64	144	145	104	118	103	109	93	133	141	120	427	0	1,705
	IE	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
	IT	1	0	92	65	27	35	29	32	52	107	50	40	22	45	0	0	0	597
	LT	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
	LU	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
	LV	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
	MT	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
	NL	0	0	0	0	0	0	5	3	1	1	2	0	1	0	1	1	6	21
	PL	0	0	0	0	0	0	0	0	0	0	1	37,216	29,350	22,266	14,312	28,680	0	131,825
	PT	0	0	0	0	0	0	0	0	117	93	73	56	41	31	30	162	0	603
	RO	213	0	0	19	11,876	19,824	16,842	14,217	12,852	11,846	11,090	10,857	11,322	11,220	8,052	29,392	0	169,722
	SE	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	1	1
	SI	0	0	0	0	51	9	2	2	0	4	2	1	2	1	1	4	0	79
	SK	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
	UK	0	0	0	0	0	1	8	6	0	0	2	0	0	0	0	0	0	17
	<b>Total</b>	<b>358</b>	<b>0</b>	<b>92</b>	<b>132</b>	<b>17,314</b>	<b>24,366</b>	<b>21,928</b>	<b>18,647</b>	<b>18,745</b>	<b>17,156</b>	<b>15,006</b>	<b>51,328</b>	<b>43,456</b>	<b>35,826</b>	<b>24,095</b>	<b>101,450</b>	<b>687</b>	<b>390,586</b>
<b>EFTA</b>	CH	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
	IS	0	0	0	0	0	0	22	0	3	27	15	49	7	4	0	0	0	127
	NO	0	0	0	0	0	0	1	0	0	0	0	0	0	0	0	0	0	1
	<b>Total</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>23</b>	<b>0</b>	<b>3</b>	<b>27</b>	<b>15</b>	<b>49</b>	<b>7</b>	<b>4</b>	<b>0</b>	<b>0</b>	<b>0</b>	<b>128</b>

**Table A.4:** BSE suspected bovine animals tested by age group in EU Member States and in EFTA-reporting countries during 2016

EU/EFTA group	Country code	Age group (months)														Total	
		< 24	> 48	> 72	24-29	30-35	36-47	48-59	60-71	72-83	84-95	96-107	108-119	120-131	132-143		144-155
EU	AT	0	1	1	1	2	3	1	2	2	0	0	0	0	2	0	15
	BE	2	4	0	2	6	5	1	4	2	0	0	1	0	0	0	27
	BG	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
	CY	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
	CZ	0	0	0	0	1	1	0	0	0	0	0	0	0	0	0	2
	DE	0	0	0	3	106	90	84	49	42	30	15	10	6	26	0	461
	DK	0	0	0	0	0	0	0	0	0	0	0	0	1	0	0	1
	EE	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
	EL	0	0	0	0	1	1	0	1	1	2	0	0	0	0	0	6
	ES	1	0	0	0	0	0	0	0	0	0	0	1	0	1	0	3
	FI	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
	FR	0	0	1	0	0	1	1	0	0	1	1	0	0	0	0	5
	HR	1	0	0	0	0	0	0	0	0	0	0	0	0	0	0	1
	HU	0	0	0	0	0	0	0	0	0	1	0	0	1	0	0	2
	IE	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
	IT	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
	LT	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
	LU	0	0	0	0	1	0	0	0	1	0	0	0	0	0	0	2
	LV	1	0	0	0	0	0	1	1	0	0	0	0	0	0	0	3
	MT	0	0	0	0	0	1	0	0	0	0	0	0	0	0	0	1
	NL	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
	PL	4	2	2	3	3	1	1	0	1	0	2	0	1	0	0	20
	PT	0	0	1	0	0	0	0	0	0	0	0	0	0	0	0	1
	RO	8	4	2	4	6	2	4	2	6	3	2	5	5	17	0	70
	SE	0	0	0	1	0	0	0	0	0	1	0	0	0	0	0	2
	SI	3	1	1	3	2	0	0	1	1	1	0	0	0	0	0	13
	SK	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
UK	0	0	0	0	0	0	0	0	1	0	0	0	0	0	0	1	
Total		20	12	8	17	128	105	93	60	57	39	20	17	14	46	0	636



EU/EFTA group	Country code	Age group (months)														Total	
		< 24	> 48	> 72	24-29	30-35	36-47	48-59	60-71	72-83	84-95	96-107	108-119	120-131	132-143		144-155
EFTA	CH	0	1	0	5	7	0	10	6	1	4	1	0	0	0	0	35
	IS	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
	NO	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
	Total	0	1	0	5	7	0	10	6	1	4	1	0	0	0	0	35

**Table A.5:** BSE active monitoring in relation to the adult bovine population (age > 2 years) during 2016

EU/EFTA group	Country code	Adult cattle (> 2 years) <sup>(a)</sup>	Number of tested bovine animals at risk <sup>(b)</sup>	Proportion (%) of tested bovine animals at risk <sup>(b)</sup>
EU	AT	894,050	16,544	1.85
	BE	1,261,350	25,767	2.04
	BG	388,240	2,312	0.60
	CY	28,720	916	3.19
	CZ	653,700	15,478	2.37
	DE	5,805,970	160,388	2.76
	DK	726,000	20,674	2.85
	EE	133,300	3,573	2.68
	EL	325,000	2,846	0.88
	ES	3,071,890	62,514	2.04
	FI	374,280	11,230	3.00
	FR	10,327,000	214,561	2.08
	HR	219,000	6,283	2.87
	HU	421,000	11,867	2.82
	IE	2,736,570	59,365	2.17
	IT	3,098,580	52,248	1.69
	LT	385,400	3,170	0.82
	LU	102,330	2,408	2.35
	LV	229,320	2,981	1.30
	MT	7,200	174	2.42
	NL	1,948,000	54,867	2.82
	PL	2,614,090	37,342	1.43
	PT	855,800	19,075	2.23
	RO	1,356,500	5,079	0.37
	SE	619,010	8,957	1.45
	SI	202,310	6,147	3.04
	SK	236,330	7,674	3.25
UK	4,477,000	146,920	3.28	
	Total	43,497,940	961,350	2.21
EFTA	CH	812,880	11,593	1.43
	IS	34,200	13	0.04
	NO <sup>(c)</sup>	353,483	6,926	1.96
	Total	1,200,563	18,532	1.54

NA: not applicable.

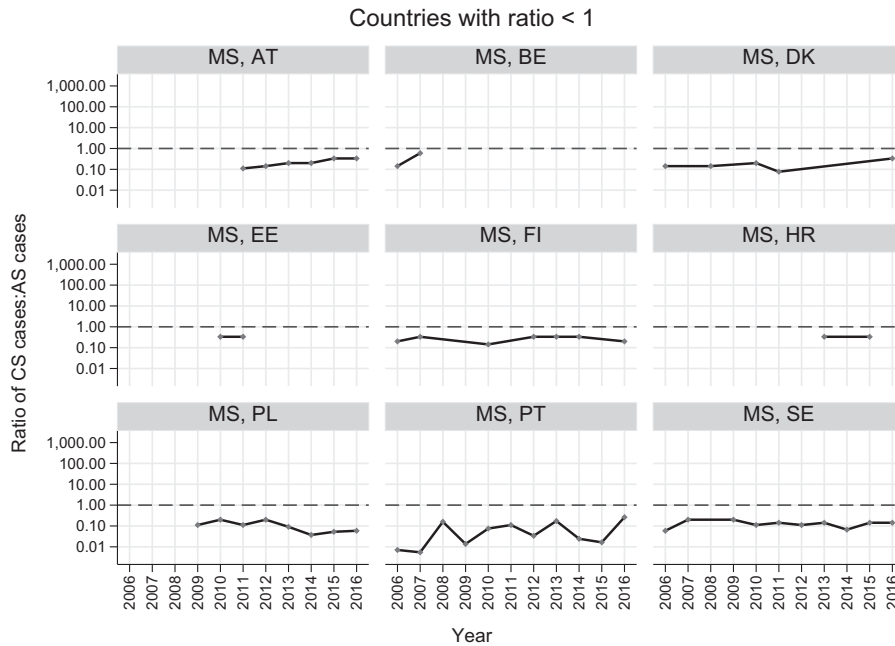
(a): Eurostat October 2016 refers to adult cattle population reported in 2015.

(b): At risk animals is the sum of animals with clinical signs at *ante-mortem*, animals culled under BSE eradication measures, emergency slaughtered and fallen stock.

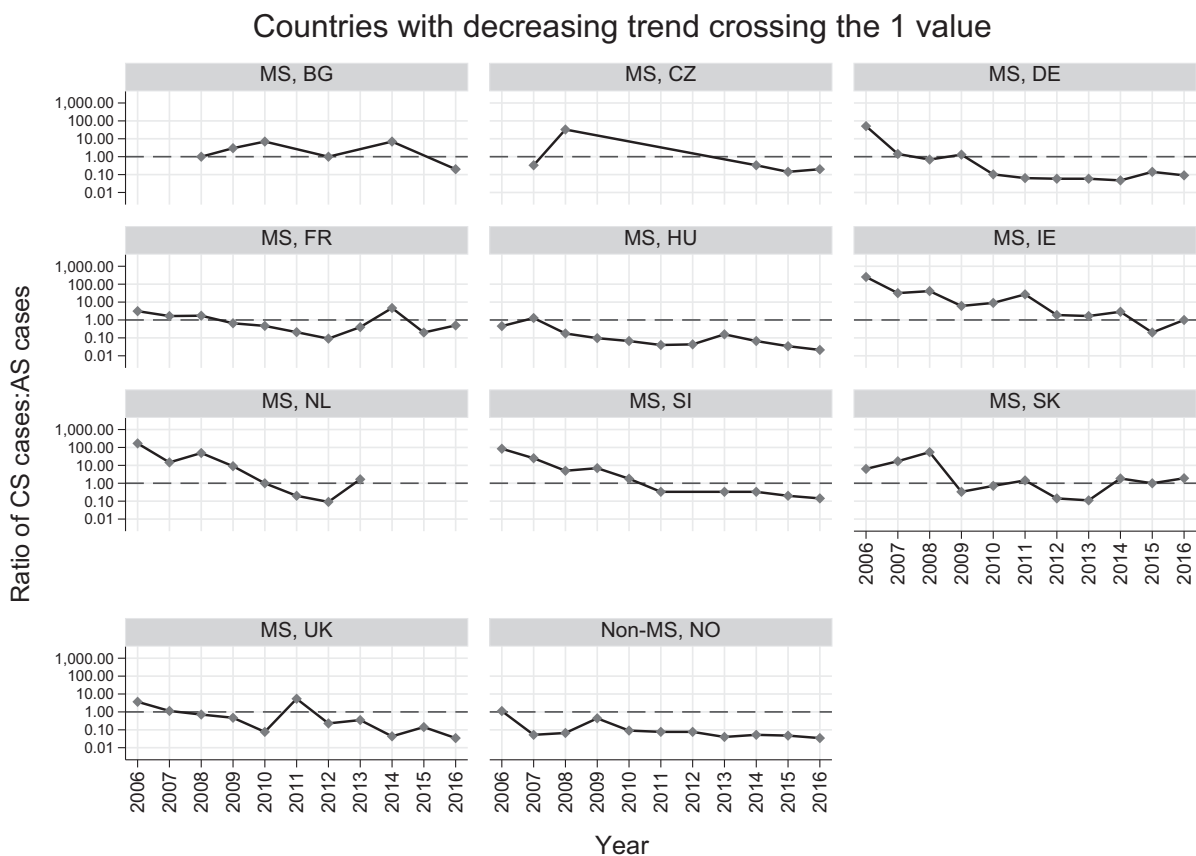
(c): Norwegian data were obtained from the central Cattle register in Norway.

## Appendix B – Evolution of the ratio of CS:AS cases in the period 2002–2016 by country

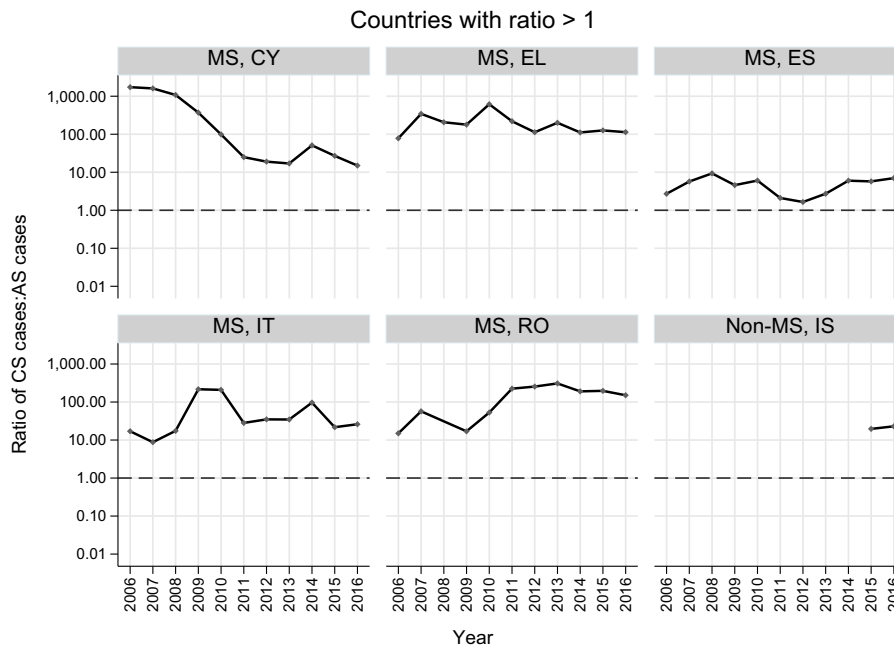
1) Countries with a CS:AS ratio < 1 (AT, BE, DK, EE, FI, HR, PL, PT and SE)



2) Countries with a CS:AS ratio crossing 1 at some time point (BG, the CZ, DE, FR, HU, IE, the NL, SI, SK, the UK, NO)

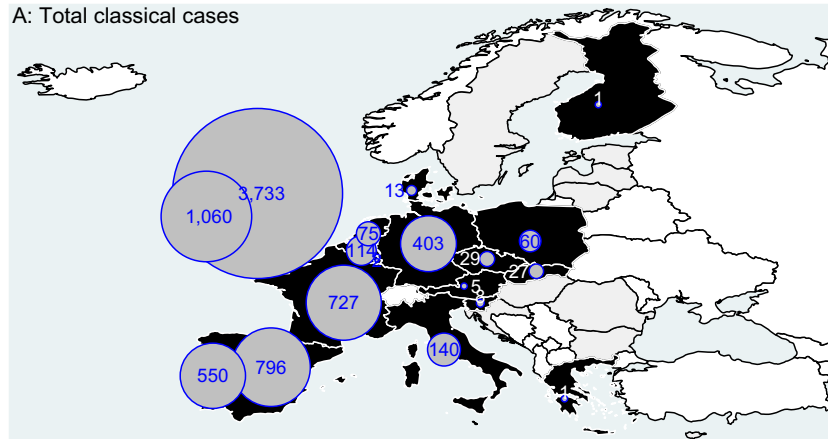


3) Countries with a CS:AS ratio > 1 (CY, EL, ES, IT, RO, IS)

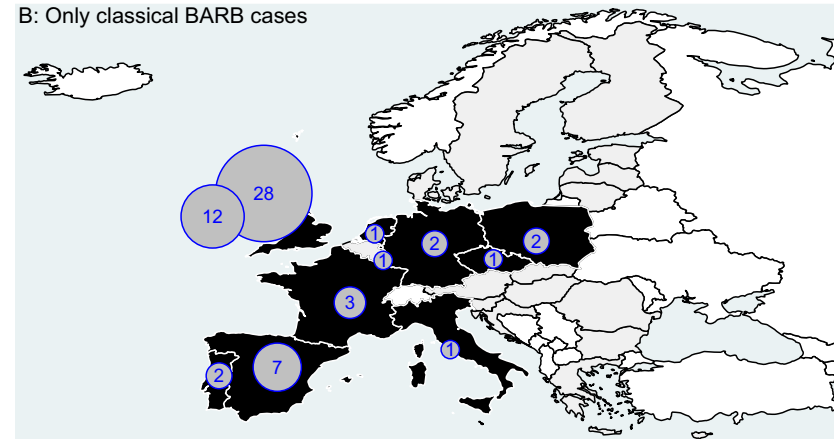


**Appendix C – Cumulative geographical distribution of numbers of cases of: C-BSE (A); C-BSE restricted to BARB cases (B); H-BSE (C); and L-BSE (D) for the period 2001–2016**

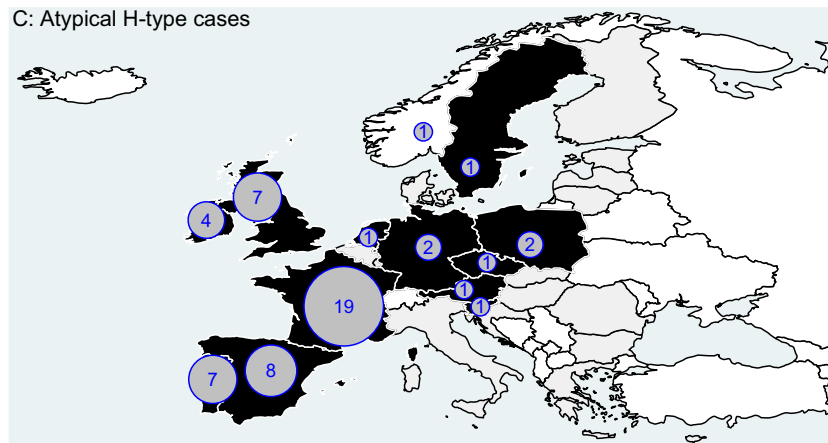
A: Total classical cases



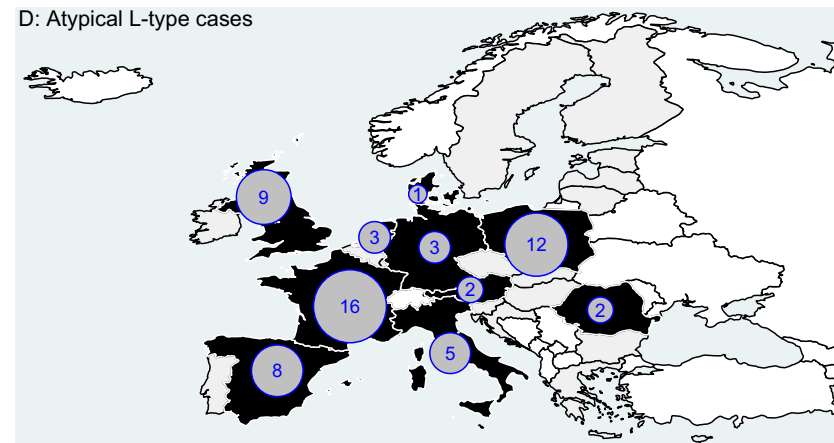
B: Only classical BARB cases



C: Atypical H-type cases

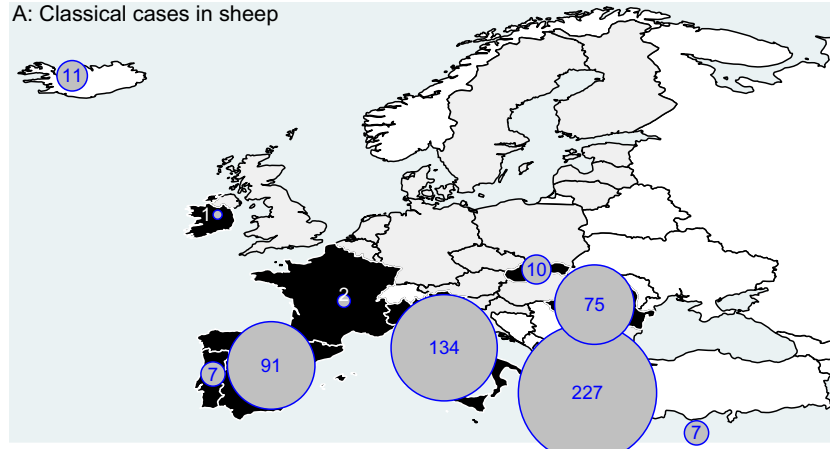


D: Atypical L-type cases



### Appendix D – Geographical distribution and numbers of cases of: ovine CS (A); caprine CS (B); ovine AS (C); and caprine AS (D) in 2016

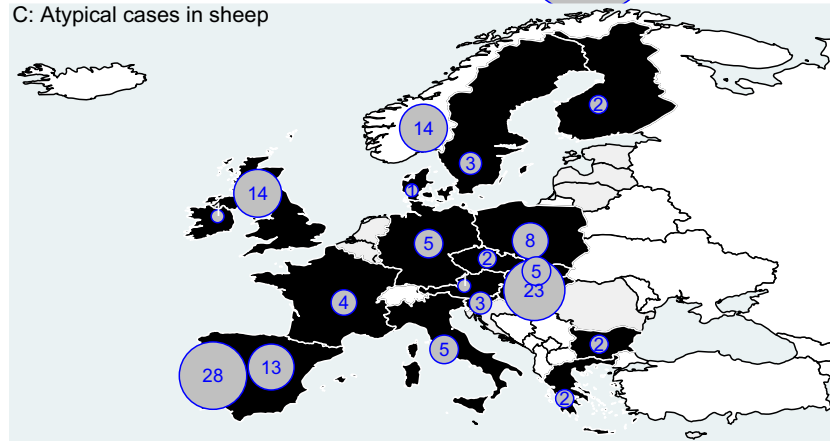
A: Classical cases in sheep



B: Classical cases in goats



C: Atypical cases in sheep



D: Atypical cases in goats

