SCIENTIFIC REPORT



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The European Union summary report on surveillance for the presence of transmissible spongiform encephalopathies (TSE) in 2018

European Food Safety Authority (EFSA)

Abstract

This report presents the results of surveillance on transmissible spongiform encephalopathies (TSE) in bovine animals, sheep, goats, cervids and other species, and genotyping in sheep, carried out in 2018 by 28 Member States (MS) according to legislation, and by Iceland, North Macedonia, Norway and Switzerland (non-MS). In total, 1,181,934 cattle were tested by MS, a 10% decrease from 2017 and 20,402 by the four non-MS. One case of classical bovine spongiform encephalopathy (BSE) was reported in 2018 by the UK, born after the enforcement of the total feed ban. Three atypical BSE cases (2 L-type/1 H-type) were reported by France. Over the year, a total of 325,386 sheep and 138,128 goats were tested in the EU, an increase of 3.4% and 17.8%, respectively, compared to 2017. In sheep, 934 cases of scrapie were reported: 821 classical (99 index cases and 97.8% with genotypes of susceptible groups) by 7 MS and 113 atypical (105 index cases) by 14 MS. Twenty-nine ovine scrapie cases were reported by Iceland and Norway. Random genotyping was only reported by eight MS and after excluding Cyprus showed that 18.7% of the genotyped sheep carried genotypes of the susceptible groups. In goats, 523 cases of scrapie were reported: 517 classical (38 index cases) by seven MS and six atypical (all index cases) by four MS. The heterogenous enforcement of a 3-year surveillance programme for chronic wasting disease (CWD) from 1/1/2018 in six MS (Estonia, Finland, Latvia, Lithuania, Poland and Sweden) resulted in the testing of 5,110 cervids and the confirmation of the first case of CWD in the EU: a wild moose in Finland. Other six MS tested 3,075 cervids with no positive results. Norway tested 33,037 cervids in 2018, resulting in seven new cases: six reindeer and one moose. In total, 105 animals from three other species were tested by two MS, with negative results.

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Keywords: TSE, BSE, CWD, scrapie, classical, atypical, surveillance

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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 2018 in the European Union (EU) and in four non-Member States (non-MS), i.e. Iceland, North Macedonia, Norway and Switzerland, as well as genotyping data in sheep. TSE monitoring data for bovine animals, small ruminants, cervids and species other than domestic ruminants are reported by country according to Regulation (EC) 999/2001 (the TSE Regulation) and consist of testing data as well as case data.

Surveillance data were mainly submitted through the newly developed EFSA TSE data reporting tool. The tool allows reporting countries to edit and automatically upload the data to the EFSA Data Collection Framework (DCF) for inclusion in the EFSA Scientific Data Warehouse. The tool has been applied for the first time during the 2018 TSE data collection period. Few MS submitted data directly as eXtensible Markup Language (XML) files by using their own system for the automatic upload of data into DCF. The electronically submitted data were extracted from the EFSA database and further processed and validated by EFSA to summarise the information and to elaborate the summary tables presented in the current EU summary report (EUSR).

A descriptive summary of the reported data is provided at MS level or aggregated at EU level and/or for all reporting countries. When possible, descriptions and calculations were stratified according to the available variables, 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/herd status (infected/non-infected), index case and age class.

In total, 1,181,934 cattle were tested in 2018 in the EU. BSE testing was concentrated in the group of risk animals (emergency slaughtered animals (ES), animals with clinical signs at *ante mortem* inspection (AM), and fallen stock (FS)) with almost 85% of all cattle tested, FS being the largest contributor with 932,049 cattle tested in 2018. An additional 20,402 cattle were tested by the four non-MS reporting countries, with no cases reported.

While no cases of C-BSE had been reported world-wide in 2017, one C-BSE case was confirmed in Scotland (UK) in 2018, a 65-month old Aberdeen angus beef cow, born after the enforcement of the total EU-wide feed ban in 2001 (BARB case). There were also three atypical cases of BSE confirmed in France: 2 L-BSE and 1 H-BSE, all born between 2001 and 2010.

In total, 463,514 small ruminants were tested in 2018 in the EU: 325,386 sheep and 138,128 goats. In addition, 21,616 and 334 sheep and goats, respectively, were tested by the four non-MS.

In sheep, 934 scrapie cases were reported in the EU in 2018, one more case than in 2017. A total 29 cases of scrapie in sheep were reported by two (Iceland and Norway) of the three non-MS reporting countries (Iceland, North Macedonia and Norway) that tested sheep (21,616). CS was reported by seven MS and one non-MS: Bulgaria, Cyprus, Greece, Ireland, Italy, Romania, Spain, and Iceland; whereas AS was reported by 14 MS and one non-MS: Austria, Denmark, Finland, France, Germany, Hungary, Ireland, Italy, Poland, Portugal, Slovakia, Spain, Sweden, the UK and Norway. Most of the ovine cases (88.6%) were reported by four countries, namely Greece, Italy, Romania and Spain, as it was the case in the previous year. In total, 821 ovine cases in the EU were CS (87.9%) and 113 were AS (12.1%). Among the non-MS, 21 CS cases were reported by Iceland and 8 AS cases by Norway. In sheep, 22% (204) of all cases in the EU reported in 2018 were index cases, with a much higher proportion in AS cases (105/113: 93%) compared to CS cases (99/821: 12%). In total, 98% of the CS cases in sheep reported in 2018 and with known genotypes belonged to animals holding genotypes of the susceptible groups (NSP3, NSP3O, NSP4 or NSP5).

In goats, 523 scrapie cases were reported in the EU in 2018, which is a reduction of 7.8% compared with 2017 when 567 cases were reported. This reduction is due mainly to the further decrease in the number of cases reported by Cyprus (from 485 in 2017 to 382 in 2018). CS was reported by seven MS, namely Bulgaria, Cyprus, France, Greece, Italy, Romania and Spain; whereas AS was reported by four MS: Cyprus, Greece, Italy and Spain. The 334 goats tested by two non-MS (Iceland and Norway) were all negative. Out of 523 caprine scrapie cases, 517 were CS (98.9%, with Cyprus accounting for 73.7% of these) and six were AS. In goats, only 8.4% of all cases reported in the EU in 2018 were index cases, similar to 2017 (8.6%), with a higher proportion in AS (6/6: 100%) than in CS (38/517: 7.3%). Currently, CS is still the most frequently reported type of scrapie in the EU in both species. Focusing on the last 10 years (2009–2018), there has been a significant average decrease of 5% per year in CS in sheep and a significant increase in CS in goats of 4%.



In 2018, the genotyping activity from random samples of the national EU sheep populations was carried out by eight MS. After excluding Cyprus, 18.7% of the randomly genotyped sheep still carried genotypes of the susceptible groups and that percentage rose to 42.4% in Greece and 31.1% in Italy, the countries with the third and the fourth highest caseloads in the EU, respectively.

The enforcement of a 3-year surveillance programme for chronic wasting disease (CWD) in six MS – Estonia, Finland, Latvia, Lithuania, Poland and Sweden – resulted in the testing of 5,110 cervids and the confirmation of the first case of CWD in the EU: a wild moose in Finland. The implementation of the mandatory surveillance in the six MS is quite heterogeneous in terms of the design (number and characteristics of the declared primary sampling units (PSU)), the number of cervids tested in general and per PSU and the distribution of testing by species and target groups. The low number of cervids tested per PSU and the targeting of mostly hunted/slaughtered fit for human consumption (HSHC) animals may have resulted in a lower than expected sensitivity of the surveillance system. Other six MS – Austria, Denmark, Hungary, Italy, Spain and Romania – tested 3,075 cervids, all with negative results and with Romania accounting for 77,6% of all cervids tested by them. Norway continued its intensified testing programme in wild and captive cervids and tested 33,037 animals in 2018, mostly semi-domesticated reindeer (36.4%), followed by wild red deer (23.5%) and wild moose (20.1%), mainly from the hunted/slaughtered for human consumption target group, leading to the detection of six reindeer cases and one moose case.

A total of 105 animals of other species were tested by Finland (99) and Estonia (6): 13 raccoon dogs (*Nyctereutes procyonoides*), 59 American minks (*Neovison vison*) and 33 foxes (genus *Vulpes*). None of them tested positive.



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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) 999/2001¹ (here referred to as the transmissible spongiform encephalopathy (TSE) Regulation), the information to be presented by MS 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 should 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 should be given for each TSE case in bovine, ovine and caprine animals. TSE cases that have been considered atypical should 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), should be reported, when appropriate.
- 7) In animals other than bovine, ovine and caprine animals, as well as in cervids other than those covered by the 3-year CWD monitoring programme referred to in Part III.A of Chapter A of this Annex, the number of samples and confirmed TSE cases per species.
- 8) The genotype, and, when 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.
- 9) For Member States covered by the 3-year CWD monitoring programme referred to in Part III.A of Chapter A of this Annex, the annual report for the years 2018, 2019 and 2020 shall include:
 - a) The number of cervid samples submitted for testing, by target group according to the following criteria:
 - primary Sampling Unit (PSU) identifier,
 - species,
 - management system (farmed, captive, wild or semi-domesticated),
 - target group,
 - sex.
 - b) The results of the rapid and confirmatory tests (number of positives and negatives) and, where applicable, of further isolate characterisation investigations, the tissue sampled and the rapid test and confirmatory technique used.
 - c) The geographical location, including the country of origin if not the same as the reporting Member State, of positive cases of TSE.
 - d) The genotype and species of each cervid found positive for TSE.
 - e) Where tested, the genotype of cervids tested and found negative for TSE.

¹ 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. 1–40.



Changes in points 7 and 9 are the result of the amendment following Commission Regulation (EU) $2017/1972^2$ – amending Annexes I and III of the TSE Regulation (see Section 1.2.4).

According to Part I.B, Chapter B of the same Annex III 'The compilation of reports containing the information referred to in Section A 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 Member States, 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.³

According to Part II of Chapter B, the Union summary shall be presented in a tabled format covering at least the information referred to in Part I.A for each Member State.

1.2. Surveillance of TSE in the European Union

1.2.1. Legal basis

Animals suspected of TSE should be examined in accordance with Article 12.2 of the TSE regulation. The legal framework for the active surveillance (i.e. the testing of animals not reported as suspected of being infected by a TSE) 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 Member States (MS) at that time, Commission Decision 2009/719/EC⁴, allowed 25 MS (all except Bulgaria and Romania) to apply a revised bovine spongiform encephalopathy (BSE) monitoring programme. Commission Implementing Decision 2013/76/EU⁵ of 4 February 2013, amending Commission Decision 2009/719/EC, authorised these 25 MS to decide to stop testing slaughtered bovine animals for human consumption. Within EU28, this monitoring programme is also applicable for Croatia.

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)⁶ also provides for mandatory discriminatory testing for BSE of TSE cases detected in small ruminants.

The recent amendments of the TSE Regulation affecting 2018 TSE surveillance, related to genotyping, classical scrapie (CS) national control programmes and chronic wasting disease (CWD) surveillance, are described below.

1.2.1.1. Genotyping

The changes following Commission Regulation (EU) 2017/894⁷ came into force on 1 January 2018. According to this amendment affecting Annex III:

a) In Chapter A, in Part II, point 8 is replaced by the following:

8. Genotyping

The prion protein genotype for the codons 136, 154 and 171 shall be determined for each positive TSE case in sheep. TSE cases found in sheep of genotypes which encode alanine on both alleles at codon 136, arginine on both alleles at codon 154 and arginine on both alleles at codon 171 shall immediately be reported to the Commission. Where the positive TSE case is an atypical scrapie case the prion protein genotype for the codon 141 shall also be determined.

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² Commission Regulation (EU) 2017/1972 of 30 October 2017 amending Annexes I and III to Regulation (EC) No 999/2001 of the European Parliament and of the Council as regards a surveillance programme for chronic wasting disease in cervids in Estonia, Finland, Latvia, Lithuania, Poland and Sweden and repealing Commission Decision 2007/182/EC. OJ L 281, 31.10.2017, p. 14–20.

³ Since 2018, TSE data are submitted by reporting countries directly to the European Food Safety Authority (EFSA) with different frequency and periodicity.

⁴ Commission Decision 2009/719/EC of 28 September 2009 authorising certain Member States to revise their annual BSE monitoring programmes. OJ L 256, 29.9.2009, p. 35–37.

⁵ Commission Implementing Decision 2013/76/EU of 4 February 2013 amending Decision 2009/719/EC authorising certain Member States to revise their annual BSE monitoring programme. OJ L 35, 6.2.2013, p. 6–7.

⁶ 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 epidemio-surveillance for transmissible spongiform encephalopathies in bovine, ovine and caprine animals. OJ L 10, 13.1.2005, p. 9–17.

Commission Regulation (EU) 2017/894 of 24 May 2017 amending Annexes III and VII to Regulation (EC) No 999/2001 of the European Parliament and of the Council as regards the genotyping of ovine animals. OJ L 138, 25.5.2017, p. 117–119.



- b) In Chapter B, in Part I(A), point 8 is replaced by the following:
 - 8. The genotype, and, where possible, the breed, of each ovine animal found positive to TSE and sampled in accordance with Chapter A, Part II, point 8.

In Annex VII, in Chapter C, in Part 1, the following point 8 is added:

- 8. Where the MS allows, in accordance with the second paragraph of point 1, the sampling and genotyping of breeding rams in flocks not participating in the breeding programme, the prion protein genotype for the codons 136, 141, 154 and 171 shall be determined for a minimum sample representative of the entire ovine population of the Member State, either:
- (a) once every 3 years with a minimum sample of at least 1,560 ovine animals; or (b) at a frequency and with a sample size determined by the Member State based on compliance with the following criteria:
 - (i) the sampling design takes into account relevant epidemiological data collected during previous surveys, including data on the prion protein genotype of sheep for the codons 136, 141, 154 and 171 by breed, region, age, sex and flock type;
 - (ii) the sampling design allows at a minimum to detect a change of 5% in genotype prevalence over a 3-year period, with an 80% power and 95% confidence level.

1.2.1.2. Classical scrapie national control programme

By Commission Implementing Regulation (EU) 2017/736,⁸ the national control programme for CS of Slovenia was approved.

1.2.1.3. CWD surveillance

By Commission Regulation (EU) 2017/1972 requirements for a 3-year surveillance programme for CWD in cervids in Estonia, Finland, Latvia, Lithuania, Poland and Sweden were established. Annexes I and III of Regulation (EC) No 999/2001 are amended in accordance with the Annex to this Regulation, which describes the surveillance programme.

The MS which have a wild and/or farmed and/or semi-domesticated population of moose and/or reindeer (Estonia, Finland, Latvia, Lithuania, Poland and Sweden) shall carry out a 3-year monitoring programme for CWD in cervids, from 1 January 2018 to 31 December 2020. The TSE tests performed for the purpose of this monitoring programme shall take place between 1 January 2018 and 31 December 2020; however, the collection of samples for the purpose of the monitoring programme may start in 2017. Details of the surveillance programme are described in Section 1.2.4.

1.2.1.4. The EU reference laboratory for TSE

Commission Regulation (EU) 2018/221⁹ of 15 February 2018 amending Regulation (EC) No 999/2001 of the European Parliament and of the Council and Regulation (EC) No 882/2004¹⁰ of the European Parliament and of the Council established that from 1 January 2019 the EU reference laboratory for TSE will be a consortium between the Istituto Zooprofilattico Sperimentale del Piemonte, Liguria e Valle d'Aosta (IZSPLVA) and the Istituto Superiore di Sanità (ISS), led by IZSPLVA.

1.2.2. BSE surveillance of bovine animals

As described in the 2016 European Union summary report (EUSR) (EFSA, 2017) on TSE, the BSE surveillance of bovine animals is based on the testing of samples from the following target groups: emergency slaughtered (ES); animals with clinical signs at *ante-mortem* (AM); fallen stock (FS); healthy slaughtered animals (HS); animals clinically suspected of being infected by BSE (SU), and animals culled under BSE eradication measures (EM).

Ommission Implementing Regulation (EU) 2017/736 of 26 April 2017 amending Annex VIII to Regulation (EC) No 999/2001 of the European Parliament and of the Council as regards the approval of Slovenia's national control programme for classical scrapie. OJ L 110, 27.4.2017, p. 2–3.

⁹ Commission Regulation (EU) 2018/221 of 15 February 2018 amending Regulation (EC) No 999/2001 of the European Parliament and of the Council and Regulation (EC) No 882/2004 of the European Parliament and of the Council as regards the European Union reference laboratory for transmissible spongiform encephalopathies. OJ L 43, 16.2.2018, p. 6–7.

Regulation (EC) No 882/2004 of the European Parliament and of the Council of 29 April 2004 on official controls performed to ensure the verification of compliance with feed and food law, animal health and animal welfare rules. OJ L 165, 30.4.2004, p. 1–141.



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 (age limits have been changed over time) and surveillance target groups. The general rules for BSE surveillance, applied in 2018, are summarised in Table 1. A table summarising the evolution of the changes (age limits for different target groups) was published in the 2015 EU Summary Report (EFSA, 2016).

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 at-risk animals at younger age. The age limits (in months) of bovine animals tested for BSE surveillance applied in 2018 by MS or non-MS reporting countries (Iceland, North Macedonia, Norway and Switzerland,) are shown in Table 2.

Table 1: Criteria for BSE surveillance in bovine animals as applied in 2018 by country, age limit and surveillance target group, based on the TSE Regulation (EC) as last amended, Commission Implementing Decision 2013/76/EU of 4 February 2013 and Commission Implementing Decision (EU) 2016/851 of 26 May 2016

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

⁽a): Different criteria were applied in 2018 because Bulgaria and Romania were not in the list of the 26 Member States authorised to revise their BSE annual surveillance programmes.

Table 2: Age limits (in months) of bovine animals tested for BSE surveillance applied in 2018 by reporting country and surveillance target group

				Surveillance target group		
Country	ES	AM	FS	HS	SU	EM
AT	> 24	> 24	> 48 ^(a)	No testing	No age limit	No age limit
BE	> 48	> 48	> 48	No testing	No age limit	> 24
BG	> 24	> 24	> 24	> 30	No age limit	No age limit
CY	> 48	> 48	> 48	No testing	No age limit	> 48
CZ	> 24	> 24	> 24	No testing	No age limit	No age limit
DE	> 48	> 24	> 48	No testing	No age limit	No age limit
DK	> 48	> 48	> 48	No testing	No age limit	> 48
EE	> 48	> 48	> 48	No testing	No age limit	No age limit
EL	> 48	> 48	> 48	> 72	No age limit	No age limit
ES	> 48	> 48	> 48	Born before 2001 and coming from herds with BSE positive cases	No age limit	No age limit
FI	> 48	> 48	> 48	No testing	No age limit	No age limit
FR	> 48	> 48	> 48	Born before 1/1/2002	No age limit	> 48
HR	> 48	> 48	> 48	No testing	No age limit	No age limit
HU	> 24	> 24	> 24	No testing	No age limit	No age limit
IE	> 48	> 48	> 48	No testing	No age limit	> 48
IT	> 48	> 48	> 48	No testing	No age limit	No age limit
LT	> 48	> 48	> 48	No testing	No age limit	No age limit
LU	> 48	> 48	> 48	No testing	No age limit	> 48
LV	> 24	> 24	> 24	No testing	No age limit	No age limit
MT	> 48	> 48	> 48	No testing	No age limit	No age limit
NL	> 48	> 48	> 48	No testing	No age limit	No age limit
PL	> 48	> 48	> 48	No testing	No age limit	No age limit



	Surveillance target group											
Country	ES	AM	FS	HS	SU	EM						
PT	> 48	> 48	> 48	No testing	No age limit	No age limit						
RO	> 24	> 24	> 24	> 30	No age limit	No age limit						
SE	> 48	> 48	> 48	No testing	No age limit	No age limit						
SI	> 48	> 48	> 48	No testing	No age limit	No age limit						
SK	> 24	> 24	> 24	No testing	No age limit	No age limit						
UK	> 48	> 48	> 48	No testing	No age limit	No age limit						
СН	> 48	> 48	> 48	> 48	No age limit	> 48						
IS	> 48	> 48	> 48	No testing	No age limit	No age limit						
MK	> 24	> 24	> 24	> 30	No age limit	No age limit						
NO	> 48	> 48	> 48	No testing	No age limit	No age limit						

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

1.2.3. TSE surveillance of small ruminants

As described in the 2016 EUSR on TSE (EFSA, 2017), the surveillance of ovine and caprine animals for the presence of TSE¹¹ is performed based on testing samples obtained from the following surveillance target groups: animals culled under TSE EM; animals not slaughtered for human consumption (NSHC); and healthy animals slaughtered for human consumption (SHC) and animals clinically suspected of being infected by TSE (SU).

Historical data and data for the reporting year have been summarised as shown in Table 3.

The minimal sample sizes for NSHC and SHC are set in Tables A and B of Annex III, chapter A, Section II, point 3 and point 2(a), respectively, of the TSE Regulation. The application of the quotas according to sheep and goat populations in each MS is displayed in Table 4. MS may choose to replace up to a maximum of 50% of their SHC ovine and caprine animals by animals obtained from NSHC, e.g. dead ovine and caprine animals over the age of 18 months and up to a maximum of 10% of their ovine and caprine animals tested in SHC and NSHC by animals (> 18 months of age) killed as part of disease eradication campaign(s) at a ratio of 1:1.

1.2.3.1. Genotyping in sheep

The prion protein genotype for the codons 136, 154 and 171 should be determined for each positive TSE case in sheep.

As described in Section 1.2.1, in 2017 the Regulation (EU) 2017/894 amended the TSE Regulation with regards to representative genotyping activities in the ovine populations. However, as it has come into force on 1 January 2018, the changes in the TSE regulation do not require to genotype a minimum sample of at least 600 for MS with an adult sheep population of > 750,000 animals, and for other MS, to genotype a minimum sample of at least 100 animals. The new requirements establish to genotype once every 3 years with a minimum sample of at least 1,560 ovine animals; or at a frequency and with a sample size determined by the MS based on compliance with a set of criteria.

⁽a): If surveillance target group is FS and animals are born in Romania, Bulgaria or Switzerland then the age limit is > 24 months.

¹¹ The term TSE surveillance is used in small ruminants as both scrapie and BSE have been detected naturally in small ruminants. When reporting TSE cases in small ruminants, the TSE type assigned is scrapie, unless stated otherwise.



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 to the TSE Regulation

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

TSE: transmissible spongiform encephalopathy; CS: classical scrapie; AS: atypical scrapie; 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): Option 2 can only be applied to sheep (genotyping and culling).

⁽b): 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.

⁽c): 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 sample size for the TSE surveillance in small ruminants by reporting country in 2018

		Sheep			Goats		
Country	Population	Surveilla	nce target group	Population	Surveilla	ance target group	
	size ^(a)	SHC	NSHC	size ^(a)	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	250–750	0	1,500	
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	100–750	0	1,500	< 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	100-750	0	1,500	< 40	0	100% up to 100	
LU	< 40	0	100% up to 100	< 40	0	100% up to 100	
LV	100–750	0	1,500	< 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	40–250	0	100% up to 500	
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	100–750	0	1,500	< 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	
СН	_			_			
IS	100–750			< 40			
MK	100–750			< 40			
NO	> 750			40–250			

TSE: transmissible spongiform encephalopathy; NSHC: animals not slaughtered for human consumption; SHC: animals. slaughtered for human consumption.

The four non-MS reporting countries are included in the Table for information. The TSE regulation does not apply to them.

Norway tested according a population (thousand heads) of > 750 and 40-250 in sheep and goats respectively.

Live sheep population in 2017 (or latest available) extracted from: https://ec.europa.eu/eurostat/databrowser/view/tag00017/default/table?lang=en

Live goat population in 2017 (or latest available) extracted from: http://appsso.eurostat.ec.europa.eu/nui/show.do?dataset=a pro_mt_lsgoat&lang=en

(-): No active surveillance system (in CH only suspect animals are tested).

1.2.4. TSE surveillance in cervids and other species

According to the Commission Regulation (EU) 2017/1972, amending Annexes I and III of the TSE Regulation, MS which have a wild and/or farmed and/or semi-domesticated population of moose and/or reindeer (Estonia, Finland, Latvia, Lithuania, Poland and Sweden) shall carry out a 3-year monitoring programme for CWD in cervids, from 1 January 2018 to 31 December 2020, although 'the collection of samples for the monitoring programme may, however, start in 2017'. The 3-year

⁽a): Thousand heads.



monitoring programme for CWD in cervids is described in detail in Annex III, chapter A, Part III of the TSE Regulation. The other MS may carry out monitoring for CWD in cervids on a voluntary basis.

Points 1.2. 1.3, 2.1, 2.2,2.3,2.4 and 2.5 of Section III, Chapter A, Annex III describe the monitoring for CWD, as follows (quoting):

- 1.2 The 3-year CWD monitoring programme shall cover the following cervid species:
 - Eurasian tundra reindeer (Rangifer tarandus tarandus);
 - Finnish forest reindeer (Rangifer tarandus fennicus);
 - Moose (Alces alces);
 - Roe deer (Capreolus capreolus);
 - White-tailed deer (Odocoileus virginianus);
 - Red deer (Cervus elaphus).
- 1.3 By way of derogation, a Member State may, based on a documented risk assessment submitted to the European Commission, select for the 3-year CWD monitoring programme a subset of the species listed above.
- 2 Sampling design
- 2.1 The Member States referred to in point 1.1 shall identify Primary Sampling Units (PSU), which shall cover all territories in which cervid populations are present, using at least the following elements:
 - (a) for farmed and captive cervids, each farm and each facility in which cervids are kept in an enclosed territory shall be considered as a PSU.
 - (b) for wild and semi-domesticated cervids, PSU shall be defined geographically based on the following criteria:
 - (i) the areas in which wild and semi-domesticated animals of a species covered by the monitoring programme gather in at least a certain period of the year;
 - (ii) if no gathering takes place for a species, the areas delimited by natural or artificial barriers in which animals of the species covered by the monitoring programme are present;
 - (iii) the areas in which animals of the species covered by the monitoring programme are hunted and areas connected to other relevant activities related to the species covered by the monitoring programme.
- 2.2 The Member States involved shall select farmed, captive, wild and semi-domesticated cervids for TSE testing using the following two-stage sampling approach:
 - (a) in the first stage, those Member States shall:
 - (i) for farmed and captive cervids:
 - select, on a random basis ensuring geographical representativeness, and if relevant taking into account relevant risk factors identified in a documented risk assessment carried out by the Member State, 100 PSU to be covered over the 3year period of the monitoring programme, or
 - if the Member State was unable to identify 100 PSU for farmed and captive cervids, select all PSU identified.
 - (ii) for wild and semi-domesticated cervids:
 - select, on a random basis ensuring geographical representativeness, and if relevant taking into account relevant risk factors identified in a documented risk assessment carried out by the Member State, 100 PSU to be covered over the 3year period of the monitoring programme, or
 - if the Member State was unable to identify 100 PSU for wild and semidomesticated cervids, select all PSU identified.
 - (b) in the second stage:
 - (i) for farmed and captive cervids:



- a Member State having selected 100 PSU shall, within every selected PSU, sample all animals belonging to the target groups listed under point 2.4.(a) over the 3-year period until a target of 30 animals tested per PSU is reached. If however certain PSU are not be able to reach the target of 30 animals tested over the 3-year period due to the limited size of their cervid population, the sampling of animals belonging to the target groups listed under point 2.4.(a) may continue in larger PSU even after having reached the target of 30 animals tested, with the objective of reaching a total number of up to 3 000 farmed and captive cervids, where possible, tested at national level over the 3-year period of the monitoring programme;
- a Member State having identified fewer than 100 PSU shall, within every PSU, sample all animals belonging to the target groups listed under point 2.4.(a) over the 3-year period, with the objective of approaching a total number of up to 3 000 farmed and captive cervids, where possible, tested at national level over the 3-year period of the monitoring programme.
- (ii) for wild and semi-domesticated cervids:
 - a Member State having selected 100 PSU shall, within every selected PSU, sample all animals belonging to the target groups listed under point 2.4.(b), over the 3-year period until a target of 30 animals tested per PSU is reached, with the objective of reaching up to 3 000 wild and semi-domesticated cervids tested at national level over the 3-year period;
 - a Member State having identified fewer than 100 PSU shall, within every PSU, sample all animals belonging to the target groups listed under point 2.4.(b) over the 3-year period, with the objective of approaching a total number of 3 000 wild and semi-domesticated cervids tested at national level over the 3-year period of the monitoring programme.
- 2.3 All cervids selected must be over 12 months of age. The age shall be estimated on the basis of dentition, obvious signs of maturity, or any other reliable information.
- 2.4 The cervids must be selected from the following target groups:
 - (a) for farmed and captive cervids:
 - (i) fallen/culled farmed or captive cervids, defined as farmed or captive cervids found dead on the enclosed territory in which they are kept, during transport or at slaughterhouse, as well as farmed or captive cervids killed for health/age reasons;
 - (ii) clinical/sick farmed or captive cervids, defined as farmed or captive cervids showing abnormal behavioural signs and/or locomotor disturbances and/or as being generally in poor condition;
 - (iii) slaughtered farmed cervids which have been declared unfit for human consumption;
 - (iv) slaughtered farmed cervids considered fit for human consumption if a Member State identifies fewer than 3 000 farmed and captive cervids from the groups (i) to (iii).
 - (b) for wild and semi-domesticated cervids:
 - (i) fallen/culled wild or semi-domesticated cervids, defined as cervids found dead in the wild as well as semi-domesticated cervids found dead or killed for health/age reasons;
 - (ii) road- or predator-injured or killed cervids, defined as wild or semi-domesticated cervids hit by road vehicles, by trains or attacked by predators;
 - (iii) clinical/sick wild and semi-domesticated cervids, defined as wild and semi-domesticated cervids which are observed as showing abnormal behavioural signs and/or locomotor disturbances and/or as being generally in poor health condition;
 - (iv) wild hunted cervids and slaughtered semi-domesticated cervids which have been declared unfit for human consumption;
 - (v) hunted wild game and slaughtered semi-domesticated cervids considered fit for human consumption if a Member State identifies fewer than 3 000 wild and semidomesticated cervids from the groups (i) to (iv).
- 2.5 In case of a positive finding of TSE in a cervid, the number of samples from cervids collected in the zone where the positive TSE case was found must be increased, based on an assessment carried out by the Member State concerned.



MS may, on a voluntary basis, carry out monitoring for TSE in animal species other than bovine, ovine, caprine and cervids according Annex III, Chapter A, Part IV of the TSE Regulation.

1.3. Testing protocols

The testing protocol for BSE surveillance in bovine animals is described in pages 8 and 9 of the 2016 EUSR on TSE (EFSA, 2017). The testing protocol for TSE surveillance in small ruminants is described in pages 13 and 14 of the 2016 EUSR on TSE (EFSA, 2017).

2. Data and methods

2.1. Origin of the data

The raw data are electronically submitted by MS and non-MS. The data to be submitted consist of testing data and case-based data for bovine animals, small ruminants, cervids and other species, according to the reporting periods (monthly basis) as described in Chapter B.I of Annex III of the TSE Regulation.

Surveillance data were mainly submitted through the newly developed EFSA TSE data reporting tool for the reporting of surveillance data on TSE as required by the TSE Regulation. The tool allows reporting countries to edit and automatically upload the data to the EFSA Data Collection Framework (DCF) for inclusion in the EFSA Scientific Data Warehouse. The tool has been applied for the first time during the 2018 TSE data collection period. Few MS submitted data directly as eXtensible Markup Language (XML) files by using their own system for the automatic upload of data into DCF. The electronically submitted data were extracted from the EFSA database and further processed and validated by EFSA to summarise the information and to elaborate the summary tables presented in the current EUSR.

Finally, information on the population of small ruminants in 2018 as presented in Table 4 were obtained from the 2017 Eurostat annual data (https://ec.europa.eu/eurostat/data/database). The number of BSE cases world-wide (Table 7) were obtained from the last available report on the monitoring and testing of ruminants for the presence of TSE in the EU (European Commission, 2016) and the OIE animal information system (http://www.oie.int/wahis).

During validation of the data with the reporting countries, additional information was asked with relation to the reporting according to (i) Annex III, Chapter B, Section 1.A, point 1 of the TSE Regulation: the number of suspected cases placed under official movement restrictions in accordance with Article 12(1), per animal species; (ii) Annex III, Chapter B, Section 1.A, point 3 of the TSE Regulation: the number of flocks for which suspected cases in ovine and caprine animals have been reported and investigated pursuant to Article 12(1) and (2)); and (iii) the number of ovine and caprine flocks tested within each subpopulation referred to in Chapter A, Part II, points 2, 3, 5 and 6. The results of this questionnaire are summarised in Appendix D.

The data in this report refer only to the samples collected and confirmed cases reported between 1 January 2018 and 31 December 2018 in the EU and the four additional non-EU reporting countries: Iceland, North Macedonia, Norway and Switzerland. North Macedonia (a non-MS and non-EFTA country) is the first time that submitted TSE data for the EU summary report.

EFSA validated the 2018 data by checking for inconsistencies in 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 started on 1 April 2019 and was finalised on 9 August 2019. The results and tables presented in the current EUSR are based on the data retrieved from the EFSA Scientific Data Warehouse on 10 July 2019. An additional consultation with reporting countries was conducted between 28 September 2019 and 12 October 2019. If data were corrected by the reporting countries in the report but not updated in the EFSA Scientific Data Warehouse, the corrections were only mentioned by means of footnotes in the current EUSR.

For some tables and figures, historical data (data between 2001 and 2018 with focus on the last 5 years in cattle and the last 10 years in sheep) were extracted from the EU TSE database. As certain MS and non-MS 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 MS for 2018. In addition, subsequent submissions of updated/amended data by MS 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 European Commission summary report for 2018 in compliance with Section II, Chapter B, Annex III of the TSE Regulation.

The 28 EU MS or EU28, the three non-MS EFTA members, Iceland, Norway and Switzerland, and the non-MS non-EFTA country, North Macedonia, are the reporting countries included in this report. 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 1059/2003.¹²

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 animals' is used here to indicate those animals whose probability of being detected with the disease is higher than in the surveillance target group of HS animals. However, this does not imply that the risk animals experienced 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/herds.

2.3. Methods

2.3.1. Descriptive methods

To describe the results of the TSE surveillance programme in the EU in 2018, 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 (e.g. cervids). Both EU aggregated data and data at the national level are presented. Where it was considered relevant, multiyear and historical data are shown. Surveillance data were available for the period 2001–2018 for bovine animals, for 2002–2018 for small ruminants, and only for the reporting year (2018) for cervids and other species.

For bovine animals, summary statistics were obtained based on the total number of tests performed in 2018 by MS and surveillance target group. In addition, historical data relating to the number of tested animals and confirmed cases since 2014 are presented in detail whereas those on the 2001–2014 period have been summed up. This 5-year period has been selected as during the period 2014-2018 a harmonised EU-wide, active BSE surveillance was applied and restricted to at risk animals of \geq 48 months of age, even though few exceptions are still in place in some countries as shown in Table 2.

Additional epidemiological parameters have been presented: number of cases by case type (e.g. C-BSE, H-BSE, L-BSE), target group, and proportions (cases per million tests) by case type and year. These have been used to describe the development of the BSE epidemic and to put into context the findings of the reporting year.

To obtain relevant epidemiological information about the BSE cases detected in 2018, EFSA asked for additional information from the individual MS 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/herd status (infected, non-infected, unknown/not available), surveillance type (passive surveillance restricted to SU vs active surveillance restricted to SHC and NSHC in non-infected flocks/herds), country, year (since 2002), case type (CS or AS), index case (yes/no). In particular, when historical data have been considered for trend analysis, the last 10-year period (2009–2018) has been included in the analysis.

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 compliance

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.



had been achieved by applying the derogation provided by the TSE Regulation (according to 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 1:1 and in addition to the minimum sample size for NSHC.

• If the MS is required to test 100% up to 500 of the NSHC in sheep, and the reported number of tested sheep was, for example 350 – the MS was categorised as compliant since it is not known the total subpopulation of NSHC in the country.

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

The reporting system of TSE surveillance data does not allow the collation of the number of newly infected flocks and herds during the reporting year but only the number of index cases, considered to be a proxy for the number of incident scrapie cases.

Finally, the classification originally developed by the Great Britain's National Scrapie Plan (NSP) was used to summarise and describe the data on genotyping.

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):

- 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.
- To plot the reported scrapie cases according to the flock/herd status, it was assumed that flocks/herds with status reported as 'unknown', 'other' or blank were considered for reporting purposes as 'non-infected flocks/herds'.
- 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/herds. If an index case was reported as 'no' or 'unknown', it was considered for reporting purposes as 'infected flocks/herds'.
- 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.

For cervids, summary statistics were extracted and presented in tabular format as follows:

- for all reporting countries, number of tested cervids in 2018 by reporting country, species and management system (wild and semi-domesticated/farmed);
- for the six MS subject to mandatory surveillance, number of PSU declared, proportion tested and median, minimum and maximum number of tested cervids in 2018 per PSU by MS and management system;
- for all reporting countries, the number of tested cervids in 2018 by target group, species, management system, and reporting country.

Number of tested animals in species other than cattle, sheep, goats and cervids tested for TSE in reporting countries in 2018 are presented in tabular format by species and reporting country.

2.3.2. Analytical methods

With regard to surveillance in cattle, the average number of cases detected per million tests at the EU level in both the risk animals and HS target groups (period 2009–2018) 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 taken into account in the model (offset).

The target group (risk animals vs HS), potentially affecting the probability of detecting the disease, was added to the model as covariate to adjust for any confounding effect. A $p \le 0.05$ was considered statistically significant. The relative risk (RR) obtained by exponentiating the beta coefficient associated with the 'year' variable, was used as a measurement of the annual variation in the probability of detection, i.e. the temporal trend for the entire period. In the model, the RR indicates the average annual change in the proportion of cases per animals tested corresponding with the annual probability of detecting the disease: a RR > 1 indicates an average annual increase in the number of cases per million whereas a RR < 1 indicates an average annual decrease.



TSE data of small ruminants from the last ten years (period 2009–2018) have been used to check if any significant temporal trend was detectable. As per BSE, a Poisson regression model has been fitted for ovine and caprine animals separately, using the number of cases as dependent variable and the year as a continuous independent variable. The number of tests was taken into account in the model (offset). The target group (NSHC vs SHC), potentially affecting the probability of detecting the disease, was added to the model as covariate to adjust for any confounding effect. A p \leq 0.05 was considered statistically significant for both the statistical analyses. The RR obtained by exponentiating the beta coefficient associated with the 'year' variable was used as a measurement of the annual variation in the probability of detection, i.e. the temporal trend for the entire period. In the model the RR indicates the average annual change in the proportion of cases per animals tested corresponding with the annual probability of detecting the disease: a RR > 1 indicates an average annual increase in the number of cases per million whereas a RR < 1 indicates an average annual decrease.

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 $p \le 0.05$ was considered statistically significant.

3. Assessment

3.1. BSE surveillance in bovine animals

About 117.7 million bovine animals have been tested for BSE in EU since 2001. In 2018, there was a 10% reduction in the number of tested bovine animals in the EU, from 1,312,714 in 2017 to 1,181,934 in 2018, mainly due to a reduction in the HS target group: from 359,137 to 177,536. Apart from Romania and Bulgaria, that tested a total of 150,613 HS animals over 30 months of age (84.8% of all tested in the HS group in the EU), Poland discontinued the testing of HS animals, from 124,492 tested in 2017 to none in the reporting year.

The four non-MS (Iceland, Norway, North Macedonia and Switzerland) tested 20,402 cattle in 2018. There was an increase of 5.5% in the number of animals tested in the risk group (ES + AM + FS), from 951,316 in 2017 to 1,003,707 in 2018. For the last 3 years, cattle in the risk group accounted for more than two-thirds of all cattle tested: 71%, 72.5% and 84.9% in 2016 and 2017 and 2018, respectively. As in previous years, cattle tested in the FS target group accounted for most of the risk animals. In 2018, it accounted for 92.9% of all risk cattle tested.

The number of cattle tested for BSE per reporting country for each target group in 2018 is shown in Table 5.

Table 5: Number of bovine animals tested for BSE by reporting country and surveillance target group in 2018 in the EU and other reporting countries

			Sur	veillance target group				
Country			Risk anir	nals	Other tar	Total		
	ES	AM	FS	Subtotal Risk animals	HS	SU	EM	
AT	3,830	32	14,588	18,450	49	17		18,516
BE	953	1	24,994	25,948	5	14		25,967
BG	3,211	1	667	3,879	27,074			30,953
CY	14		1,505	1,519	20			1,539
CZ	833	2	20,879	21,714	18			21,732
DE	10,409		161,046	171,455	260	515		172,230
DK	1,565		22,272	23,837	63	1		23,901
EE	159	55	3,390	3,604				3,604
EL	3	11	925	939	11,472			12,411
ES	300	20	64,852	65,172	210	3		65,385
FI	15	2	11,295	11,312	4			11,316
FR	5,135		205,741	210,876	13,615	3	12	224,506
HR	28		5,006	5,034	121			5,155
HU	93	2	11,892	11,987	587	2		12,576
IE		485	69,727	70,212		20		70,232



			Sur	veillance target group				
Country			Risk anir	mals	Other tar	get gro	oups	Total
	ES	AM	FS	Subtotal Risk animals	HS	SU	EM	
IT	18,641	415	35,646	54,702	345			55,047
LT	12	16	3,002	3,030		3,030		
LU			2,790	2,790		3		2,793
LV	534	644	3,831	5,009		5		5,014
MT	55		155	210				210
NL	5,111		45,290	50,401	32			50,433
PL	4,261	544	34,900	39,705	9,705 10		39,715	
PT	1,623	1,547	17,674	20,844	,844 43		20,887	
RO	2,573	3,042	3,364	8,979	123,539	40		132,558
SE	166	30	7,396	7,592		30		7,622
SI	441	99	6,092	6,632	55	11		6,698
SK	23	2	7,935	7,960	1			7,961
UK	4,274	446	145,195	149,915	23	1	4	149,943
Total EU	64,262	7,396	932,049	1,003,707	177,536	675	16	1,181,934
СН	4,398		6,331	10,729		37		10,766
IS	10		67	77	14			91
MK			2	2	3,215	1		3,218
NO	5,054	89	1,183	6,326		1		6,327
Total Non-EU	9,462	89	7,583	17,134	3,229	39	0	20,402
Total	73,724	7,485	939,632	1,020,841	180,765	714	16	1,202,336

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

The distribution of the number of bovine animals tested for BSE by age group, surveillance target group and reporting country in 2018 can be found in the following link https://doi.org/10.5281/zenodo. 3444249, distributed as follows:

- Table 6: Number of bovine animals tested by age group in the EU MS and non-MS reporting countries in 2018.
- Table 7: Number of bovine animals in the risk group (animals with clinical signs at antemortem, emergency slaughtered and fallen stock) and animals culled under BSE eradication measures, by age group, tested in EU MS and non-MS reporting countries in 2018.
- Table 8: Number of tested healthy slaughtered bovine animals by age group in EU MS and in non-MS reporting countries in 2018.
- Table 9: Number of BSE suspected bovine animals, by age group, tested in EU MS and in non-MS-reporting countries in 2018.

In the EU, four BSE cases were reported in 2018: one C-BSE detected in Scotland (UK) and three atypical BSE cases in France (2 L-BSE and 1 H-BSE). Table 10 reports the main clinical and epidemiological data of the positive cases. The C-BSE case was an Aberdeen angus beef cow, born in April 2013 (BARB case). The two L-type cases were confirmed in two dairy cows born in May 2010 and September 2008, respectively. The H-type case was found in a very old Limousin beef cow born in November 2001. One additional H-type case was reported outside Europe, in the state of Florida (United States of America) in a 6-year old mixed-breed-beef cow. ¹³

The number of atypical cases is similar to those found in the previous years. Focusing on the last five years, the proportion of cases per million tests ranged between 0 and 4, considering the three types of BSE (Figure 1). Most of the EU atypical cases were detected in old animals. Based on the known age of

¹³ http://www.oie.int/wahis_2/public/wahid.php/Reviewreport/Review?page_refer=MapFullEventReport&reportid=27683 https://www.cdc.gov/prions/bse/case-us.html



108 atypical BSE cases, the average age at detection was 11.7 years (range: 6–18 years) and tested in the FS target group that accounts, as mentioned above, for most of the tested animals.

The number of BSE cases by reporting country, type and year (period 1991-2018, with a focus on the last five years) is shown in Tables 11-13.

Time series analysis carried out over the last 10-year period (period 2009–2018) shows a significant decreasing trend in the occurrence of C-BSE (annual RR = 0.61 i.e. an annual decrease of 39% in the proportion of cases per tested animals; p < 0.0001), whereas no significant trend for the two atypical BSE forms was found (H-BSE: annual RR = 0.95, p = 0.46; L-BSE: annual RR = 1.05, p = 0.48).

Maps showing the geographical distribution of the cumulative number of cases and the cumulative proportion of cases per million tests of C-BSE cases born after the total (reinforced) feed ban (BARB), H-BSE and L-BSE for the period 2001–2018, are shown in Appendix B.

Table 10: Clinical and epidemiological description of the four BSE cases detected in 2018

Country	UK – classical 1	FR – atypical 1	FR – atypical 2	FR – atypical 3	
Surveillance target group	Fallen stock	Emergency slaughter	Fallen stock	Fallen stock	
Case type	Classical	L-BSE	H-BSE	L-BSE	
Month and year of birth	April 2013	May 2010	November 2001	September 2008	
Age at detection (in months)	65	92	194	123	
BARB status	Yes	No	No	No	
Clinical signs	Falling, recumbent	No clinical symptoms	No clinical symptoms	Aggressive animal	
Cattle type	Beef	Dairy	Beef	Dairy	
Breed	Aberdeen Angus	Prim'Holstein	Limousin	Limousin	
Was the case confirmed at herd/ holding where the animal was born?	Yes	Yes	Yes	Yes	
Location (NUTS3) of natal herd or herd where case found	UKM5	76 260 33540 ETALONDES SAUVETERRE DE GUYENNE Gironde		87 210 LA CROIX SUR GARTEMPE Haute-Vienne	
Herd size	22	119	30	296	
Herd type	Beef	Dairy	Suckling	Beef	
Feeding system during first year of life	Unknown		Milk (from its mother), hay and grass	Hay, pellets and grass	
Feed cohorts? Tested? If Yes: Results (number tested; number positives)	No	0	0	Not yet slaughtered	
Birth cohorts? Tested? If Yes: Results (number tested; number positives)	Yes, 3 tested all negative	13 all tested all negative	2 cows + 1 veal negatives	Not yet slaughtered	
Offspring? Tested? If Yes: Results (number tested; number Positives)	3 offspring 2 previously died not tested, 1 tested and negative	0	0	Not yet slaughtered	
Sire? Tested? (Yes/ No). If Yes: Results (positive? Negative?)	Previously died - Not tested	0	0	Yes, negative	
Dam? Tested (Yes/No). If Yes: Results (positive? Negative?)	Alive at time	0	0	Yes, negative	

BSE: bovine spongiform encephalopathy; H-BSE: H-type BSE; L-BSE: L-type BSE.



Table 11: Total number of reported BSE cases (classical-BSE + atypical H-BSE + atypical L-BSE) in reporting countries and worldwide by year (period 1991–2018) and country

_			Year					
Country	Up to 2013	2014	2015	2016	2017	2018	Total	
AT	8	0	0	0	0	0	8	
BE	133	0	0	0	0	0	133	
CZ	30	0	0	0	0	0	30	
DE ^(a)	419	2	0	0	0	0	421	
DK ^(a)	16	0	0	0	0	0	16	
EL	1	0	0	0	0	0	1	
ES	810	2	1	1	3	0	817	
FI	1	0	0	0	0	0	1	
FR ^(a)	997	3	0	4	2	3	1,009	
IE ^(a)	1,659	0	1	0	1	0	1,661	
IT ^(a)	147	0	0	0	0	0	147	
LU	3	0	0	0	0	0	3	
NL ^(b)	88	0	0	0	0	0	88	
PL	75	0	0	0	0	0	75	
PT	1,085	1	0	0	0	0	1,086	
RO	0	2	0	0	0	0	2	
SE ^(c)	1	0	0	0	0	0	1	
SI	8	0	1	0	0	0	9	
SK	27	0	0	0	0	0	27	
UK	184,591	1	2	0	0	1	184,595	
Total EU-28	190,099	11	5	5	6	4	190,130	
BRA	1	1	0	0	0	0	2	
CAN ^(a)	19	0	1	0	0	0	20	
ISR	1	0	0	0	0	0	1	
JPN	36	0	0	0	0	0	36	
LI	2	0	0	0	0	0	2	
NO	0	0	1	0	0	0	1	
CH ^(a)	467	0	0	0	0	0	467	
USA ^(a)	4	0	0	0	1	1	6	
Total non-EU	530	1	2	0	1	1	535	
Total	190,629	12	7	5	7	5	190,665	

BSE: bovine spongiform encephalopathy; H-BSE: H-type BSE; L-BSE: L-type BSE; BRA: Brazil; CAN: Canada; ISR: Israel; JPN: Japan; LI: Liechtenstein; NO: Norway; CH: Switzerland; USA: The United States of America.

Each cell reports the total number of BSE cases (C-BSE + H-BSE + L-BSE). Grey shaded cells indicate the year(s) and Member State where at least one BARB case was detected (EFSA, 2017).

EU countries without BSE cases are not included.

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 Commission, 2016). Data were retrieved from the EU TSE Database and the OIE website (http://www.oie.int/wahis).

⁽a): Included imported cases: CAN 1 case in 1993; Denmark 1 case in 1992; France 1 case in 1999; Germany 1 case in 1992, 3 cases in 1994, 2 cases in 1997; Ireland 5 cases in 1989, 1 case in 1990, 2 cases in 1991 and 1992, 1 case in 1994 and 1 case in 1995; Italy 2 cases in 1994, 2001 and 2002; Portugal 1 case in 1990, 1991, 1992, 2000 and 2004 and 3 cases in 1993; Slovenia 1 case in 2004; Switzerland 1 case in 2012; USA 1 case in 2003.

⁽b): NL: The number of classical BSE for NL should be updated in the database for 2001 from 18 cases to 19 cases. Therefore, the total of the Netherlands for the column up to 2013 is 88 instead of 87.

⁽c): Gavier-Widen et al. (2008).



Table 12: Number of reported classical BSE cases in the EU and non-EU reporting countries by year (period 2001–2018) and country

			Year					
Country code	up to 2013	2014	2015	2016	2017	2018	Total	
AT	5	0	0	0	0	0	5	
BE	133	0	0	0	0	0	133	
CZ	29	0	0	0	0	0	29	
DE	416	0	0	0	0	0	416	
DK	15	0	0	0	0	0	15	
EL	1	0	0	0	0	0	1	
ES	797	1	0	0	0	0	798	
FI	1	0	0	0	0	0	1	
FR	968	0	0	1	0	0	969	
IE	1,655	0	1	0	0	0	1,656	
IT	142	0	0	0	0	0	142	
LU	3	0	0	0	0	0	3	
NL	84	0	0	0	0	0	84	
PL	60	0	0	0	0	0	60	
PT	1,078	1	0	0	0	0	1,079	
SI	8	0	0	0	0	0	8	
SK	27	0	0	0	0	0	27	
UK	184,576	1	1	0	0	1	184,579	
Total EU	189,998	3	2	1	0	1	190,005	
СН	464	0	0	0	0	0	464	
Total non-EU	464	0	0	0	0	0	464	
Total	190,462	3	2	1	0	1	190,469	

BSE: bovine spongiform encephalopathy.

Each cell reports the total number of C-BSE cases.

Reporting countries that have never reported classical cases are not included in the table.

Grey shaded cells indicate the year and Member State where at least one BARB case was detected (EFSA BIOHAZ Panel, 2017). Source: data were retrieved from the EU TSE Database and from the OIE website for CH.

Table 13: Number of reported BSE atypical cases in EU and non-EU reporting countries by year (period 2001–2018), type and country

		Year												
Country	up to 2013		20	2014		15	2016		2017		2018			
	Н	L	Н	L	Н	L	Н	L	н	L	Н	L	Н	L
AT	1	2											1	2
CZ	1	0											1	0
DE	1	2	1	1									2	3
DK	0	1											0	1
ES	7	6		1		1	1		1	2			9	10
FR	14	15	1	2			3		1	1	1	2	20	20
IE	4	0								1			4	1
IT	0	5											0	5
NL	1	3											1	3
PL ^(a)	2	12											2	12
PT	7	0											7	0
RO	0	0		2									0	2
SE	1	0											1	0



						Yea	r						TO	TAL
Country	up to	2013	20	14	20	15	20	16	20	17	20	18		
	Н	L	Н	L	Н	L	Н	L	Н	L	Н	L	Н	L
SI	0	0			1								1	0
UK	6	9			1								7	9
Total EU	45	55	2	6	2	1	4	0	2	4	1	2	56	68
NO	0	0			1								1	0
CH	1	0											1	0
Total non-EU	1	0	0	0	1	0	0	0	0	0	0	0	2	0
Total	46	55	2	6	3	1	4	0	2	4	1	2	58	68

BSE: bovine spongiform encephalopathy.

Each cell reports the total number of H-BSE and L-BSE cases.

Reporting countries that have never reported atypical cases are not included in the table.

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

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

The number of historical reported BSE cases can be found in the following link https://doi.org/10.5281/zenodo.1436520, as follows:

- Number of BSE cases per country and year until 2000 (included) in the EU and EFTA.
- Number of classical BSE cases per country and year from 2001 in the EU and non-EU reporting countries.
- Number of atypical H BSE cases per country and year from 2001 in the EU and non-EU reporting countries.
- Number of atypical L BSE cases per country and year from 2001 in the EU and non-EU reporting countries.



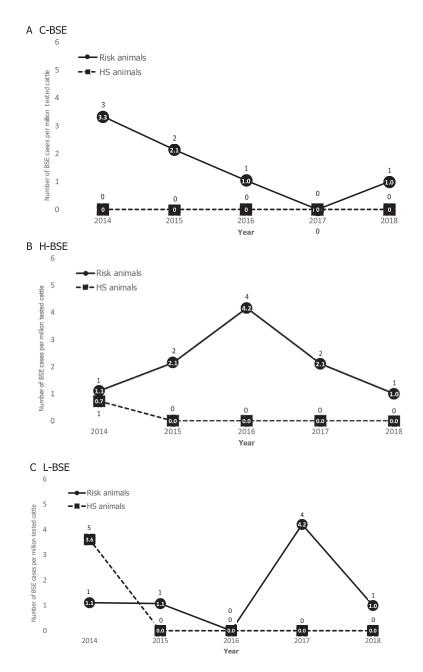


Figure 1: Cases per million tested bovine animals by surveillance target group and case type for the period 2014–2018 in the EU (black numbers in white background: number of cases)

3.2. TSE surveillance in small ruminants

Since 2002, nearly 9.5 million small ruminants have been tested as part of the official EU TSE surveillance. In 2018, 463,514 small ruminants were tested by the 28 MS: 325,386 sheep (70.2%) and 138,128 goats (29.8%), which represents a 7.3% increase (31,699) in the number of tested small ruminants in the EU, compared with that of 2017.

In the four non-MS reporting countries, a total 21,950 small ruminants were tested: 21,616 sheep (98.5%) and 334 goats (1.5%).

In sheep, the increase in the total tested in the EU was 3.4% (325,386 tested in 2018 compared with 314,547 in 2017), mostly due to testing in TSE-infected flocks with a 39.6% increase in 2018 (from 26,982 in 2017 to 37,662 in 2018), whereas testing in non TSE-infected flocks remained at the same level as in 2017.

In goats, there was a 17.8% increase in the tested animals in the EU (138,128 in 2018 compared with 117,268 in 2017), mostly due to testing in TSE-infected herds with more than a three-fold



increase (from 3,686 in 2017 to 12,375 in 2018), whereas testing in non TSE-infected herds had a 10.6% increase (from 113,582 in 2017 to 125,658 in 2018).

The numbers of sheep and goats tested for TSE by reporting country, surveillance target group and flock/herd status in 2018 are summarised in Tables 14 and 15, respectively. 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), 19 MS fulfilled the requirements for sheep testing. In goat surveillance, 22 MS fulfilled the requirements for goat testing.

In 2018, for each sheep tested in a TSE-infected flock, there were 7.6 sheep tested in non-TSE-infected flocks, compared with nearly 11 tested in non-TSE-infected flocks in 2017. This difference is due to the increase in the number of tested sheep from TSE-infected flocks under restrictions which has increased (+10,680) probably due to the number of cases confirmed in 2018 and previous years, mostly in Greece, Italy, Romania and Spain.

In 2018, for each goat tested in a TSE-infected herd, there were over 10 goats tested in non-TSE infected herds, compared with nearly 31 goats tested in non-TSE-infected herds in 2017. This is due to the increase in the number of tested goats from TSE-infected herds under restrictions (+8,689). The main contributors to goats tested from TSE-infected herds were Cyprus and Spain.



Table 14: Number of sheep tested for TSE by reporting country, surveillance target group and flock status in 2018 in the EU and other reporting countries

Flock status		TSE-i	nfected fl	ocks			Non-i	nfected fl	ocks			Unkn	own/N	ot av	ailable	
Surveillance Target group	ЕМ	NSHC	SHC	SU	Subtotal TSE infected flocks	EM	NSHC	SHC	SU	Subtotal Non- infected flocks	EM	NSHC	SHC	SU	Subtotal Unknown/ Not available	Total
AT		20			20		2,731	126		2,857					0	2,877
BE					0		1,570		2	1,572					0	1,572
BG					0		342	19,647		19,989					0	19,989
CY		2,839	993		3,832					0					0	3,832
CZ					0		2,918	3		2,921					0	2,921
DE	6				6		9,891	10,008	44	19,943					0	19,949
DK					0		437	1	1	439					0	439
EE					0		253			253					0	253
EL	1,653	2,006	698		4,357		287	2,473	11	2,771					0	7,128
ES	9,086	·			9,086		12,173	8,660	7	20,840					0	29,926
FI		46	23		69		1,521	3		1,524					0	1,593
FR	55				55		18,319	5,733		24,052					0	24,107
HR					0		1,374		1	1,375					0	1,375
HU		4,720	364		5,084		7,144	9,341	1	16,486					0	21,570
IE	57	68	455		580		10,734	10,580		21,314					0	21,894
IT	249	303	3,595		4,147		12,535	11,866	2	24,403					0	28,550
LT					0		1,831			1,831					0	1,831
LU					0		108			108					0	108
LV					0		201			201					0	201
MT					0		232	117		349					0	349
NL					0		1,487			1,487					0	1,487
PL					0		10,637	18,928		29,565					0	29,565
PT		281	108		389		15,062	5,048		20,110					0	20,499
RO		904	5,971	102	6,977		18,298	18,406	70	36,774					0	43,751
SE			,		0		,	,		0		1,740	87		1,827	1,827



Flock status		TSE-	infected fl	ocks			Non-	infected fl	ocks			Unkn	own/N	lot av	ailable	
Surveillance Target group	EM	NSHC	SHC	SU	Subtotal TSE infected flocks	EM	NSHC	SHC	SU	Subtotal Non- infected flocks	EM	NSHC	SHC	SU	Subtotal Unknown/ Not available	Total
SI					0		2,309	213	10	2,532					0	2,532
SK	4	1,533	1,021	1	2,559		10,608			10,608					0	13,167
UK		501			501		16,001	5,591	1	21,593					0	22,094
Total EU	11,110	13,221	13,228	103	37,662	0	159,003	126,744	150	285,897	0	1,740	87	0	1,827	325,386
СН					0					0					0	0
IS	467				467		50	3,081	32	3,163					0	3,630
MK					0			145	47	192					0	192
NO	159				159		6,730	10,901	4	17,635					0	17,794
Total non-EU	626	0	0	0	626	0	6,780	14,127	83	20,990	0	0	0	0	0	21,616
Total	11,736	13,221	13,228	103	38,288	0	165,783	140,871	233	306,887	0	1,740	87	0	1,827	347,002

EM: animals culled under TSE eradication measures; NSHC: animals not slaughtered for human consumption; SHC: animals slaughter

Sheep reported with clinical signs at ante-mortem (AM) have been included in this table as SU: 11 from EL, 3 from IS and 4 from NO.



Table 15: Number of goats tested for TSE by reporting country, surveillance target group and herd status in 2018 in the EU and other reporting countries

Herd status		TSE-i	nfected	herds			Non-	infected	herds			Unk	nown/I	Not ava	ailable	
Surveillance Target group	EM	NSHC	SHC	SU	Subtotal infected herds	EM	NSHC	SHC	SU	Subtotal non- infected herds	EM	NSHC	SHC	SU	Subtotal Unknown/Not available	Total
AT					0		821	29		850					0	850
BE					0		546			546					0	546
BG					0		113	1,529		1,642					0	1,642
CY	206	5,039	3,263		8,508		9	2		11					0	8,519
CZ					0		449			449					0	449
DE					0		1,842	290	9	2,141					0	2,141
DK					0		98	1		99					0	99
EE					0		11			11					0	11
EL	108	253			361		761	1380	2	2,143					0	2,504
ES	1,651				1,651		10,352	9,374		19,726					0	21,377
FI					0		282			282					0	282
FR	740				740		16,724	5,018	1	21,743					0	22,483
HR					0		320			320					0	320
HU			1		1		112	102		214					0	215
IE					0		151			151					0	151
IT	68	46	566		680		6,536	24,191		30,727					0	31,407
LT					0		5			5					0	5
LU					0		88			88					0	88
LV					0		23			23					0	23
MT					0		127	113		240					0	240
NL					0		1,487			1,487					0	1,487
PL					0		3,776	642		4,418					0	4,418
PT		13			13		1537	14		1,551					0	1,564
RO		18			18		7,094	28,327	9	35,430					0	35,448
SE					0					0		95			95	95



Herd status		TSE-i	nfected	herds			Non-	-infected	herds			Unk	nown/l	Not ava	ailable	
Surveillance Target group	EM	NSHC	SHC	SU	Subtotal infected herds	EM	NSHC	SHC	SU	Subtotal non- infected herds	EM	NSHC	SHC	SU	Subtotal Unknown/Not available	Total
SI					0		512	85	7	604					0	604
SK			5		5		251			251					0	256
UK	86	312			398		506			506					0	904
Total EU	2,859	5,681	3,835	0	12,375	0	54,533	71,097	28	125,658	0	95	0	0	95	138,128
СН					0					0					0	0
IS					0			4	2	6					0	6
MK					0					0					0	0
NO					0		296	32		328					0	328
Total non-EU	0	0	0	0	0	0	296	36	2	334	0	0	0	0	0	334
Total	2,895	5,681	3,835	0	12,375	0	54,829	71,113	30	125,992	0	95	0	0	95	138,462

EM: animals culled under TSE eradication measures; NSHC: animals not slaughtered for human consumption; SHC: animals slaughter

Goats reported with clinical signs at ante-mortem (AM) have been included in this table as SU: 2 from EL.



In total, 934 scrapie cases in sheep were reported in the EU in 2018, one more case than in 2017. They were reported by 18 MS. In addition, 29 scrapie cases in sheep were reported by two (Iceland and Norway) of the four non-MS reporting countries.

CS was reported by seven different MS and one non-MS: Bulgaria, Cyprus, Greece, Ireland, Italy, Romania, Spain and Iceland, whereas AS was reported by 14 MS and one non-MS: Austria, Denmark, Finland, France, Germany, Hungary, Ireland, Italy, Poland, Portugal, Slovakia, Spain, Sweden, the UK and Norway. Most of the ovine cases in the EU of both CS and AS (88.6%) were reported by four countries, namely Greece, Italy, Romania and Spain, as it was the case in the previous years.

In total, 821 sheep scrapie cases in the EU in 2018 were CS cases (87.9%) and 113 were AS cases (12.1%). Among the non-EU reporting countries, 21 CS cases were reported by Iceland and 8 AS cases by Norway. Table 16 shows the number of scrapie cases in sheep by reporting country, case type, index case status and surveillance target group in 2018. The geographical distribution of AS and CS in 2018 in sheep is shown in Appendix C.

In sheep, 21.8% (204) of all cases in the EU reported in 2018 were index cases, with a much higher proportion in AS cases (105/113: 93%) than in CS cases (99/821: 12%), probably due to the high within-flock spread of CS. Using the absolute number of index cases as a proxy for the flock-level incidence in sheep and comparing 2017 with 2018, there was a decrease in the number of CS index cases (from 145 to 99, -31.7%) and an increase in the number of AS index cases (from 89 to 105, +18%).

In total, 523 scrapie cases in goats were reported in the EU in 2018, 7.8% reduction (–44) compared with 2017 when 567 cases were reported. This change is due mainly to the further decrease in the number of cases in goats in a single reporting country, Cyprus, from 485 in 2017 to 382 in 2018. CS cases were reported by seven MS: Bulgaria, Cyprus, France, Greece, Italy, Romania and Spain, whereas AS was reported by four of them: Cyprus, Greece, Italy and Spain. As mentioned previously, most of the CS cases were reported by Cyprus, but the contribution of this country to the caseload continued decreasing (from 86.7% in 2017 to 73.7% in 2018). The four non-MS reporting countries did not report any scrapie cases in goats.

In total, 517 caprine cases in the EU in 2018 were CS cases (98.9%) and six were AS (1.1%). Table 17 shows the number of scrapie cases in goats by reporting country, case type, index case status and surveillance target group in 2018. The geographical distribution of AS and CS in 2018 in goats is shown in Appendix C.

In goats, 8.4% (44) of all cases reported in the EU in 2018 were index cases, similar to that in 2017 (8.6%), with a higher proportion in AS (6/6: 100%) than in CS (38/517: 7.3%). Cyprus and Italy accounted for 50% of all index cases in goats. Using the absolute number of index cases in goats as a proxy for the herd-level incidence in goats and comparing 2018 with 2017, there was a slight decrease in the number of CS index cases (from 42 to 38) and almost no change in the number of AS index cases (from 7 to 6).

In general, considering the total number of cases by type and without restricting the calculation to index cases only, CS is still the most frequently reported type of scrapie in the EU in both the species. In 2018 the CS/AS ratio was 7.3:1 in sheep (slightly lower than in 2017: 8.9:1) and 86.1:1 in goats (55.8:1 in 2017). If, for goats, Cyprus is excluded, the CS/AS ratio in goats was 27.2 in 2018, much higher than in 2017 (8.2:1).



Table 16: Number of scrapie cases in sheep by country, case type, index case status, surveillance target group in 2018 in the EU and other reporting countries

Case type			Atyp	ical sc	rapie (AS)							Classical	scrapie	(CS)				
Index case		No			,	Yes		Total			No				Y	es		Total	Total
Surveillance target group	NSHC	SHC	Subtotal	NSHC	SHC	SU	Subtotal	AS	EM	NSHC	SHC	SU	Subtotal	NSHC	SHC	SU	Subtotal	CS	
AT			0	1			1	1					0				0	0	1
BG			0				0	0					0		5		5	5	5
CY			0				0	0		3	1		4				0	4	4
DE		1	1	2		1	3	4					0				0	0	4
DK			0	1			1	1					0				0	0	1
EL			0				0	0	75	66	4		145	19	11	4	34	179	179
ES			0	8	1		9	9	266				266	9	4		13	279	288
FI			0	2			2	2					0				0	0	2
FR				6			6	6					0				0	0	6
HU	3		3	5	5		10	13					0				0	0	13
IE			0	7	1		8	8					0	1			1	1	9
IT			0	6	2		8	8	1	17	110		128	15	7		22	150	158
PL			0	3	3		6	6					0				0	0	6
PT	2		2	20	8		28	30					0				0	0	30
RO			0				0	0		27	140	12	179	11	5	8	24	203	203
SE		1	1	1			1	2					0				0	0	2
SK			0	6			6	6					0				0	0	6
UK	1		1	14	2		16	17					0				0	0	17
Total EU	6	2	8	82	22	1	105	113	342	113	255	12	722	55	32	12	99	821	934
IS			0				0	0	20				20		1		1	21	21
NO				5	3		8	8					0				0	0	8
Total non-EU	0	0	0	5	3	0	8	8	20	0	0	0	20	0	1	0	1	21	29
Total	6	2	8	87	25	1	113	121	362	113	255	12	742	55	33	12	100	842	963

^{&#}x27;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.

Sheep cases reported with clinical signs at ante-mortem (AM) have been included in this table as SU: 4 from EL.

Only the reporting countries in which scrapie cases in sheep were detected in 2017 are mentioned in the table.'



Table 17: Number of scrapie cases in goats by country, case type, index case status, surveillance target group in 2018 in the EU and other reporting countries

Case type				Atypical	scrapie	(AS)							Classical	scrapie	(CS)				
Index case			No			Yes	5	Total			No				Y	es		Total	Total
Surveillance target group	EM	NSHC	SHC	Subtotal	NSHC	SHC	Subtotal	AS	EM	NSHC	SHC	SU	Subtotal	NSHC	SHC	SU	Subtotal	CS	
BG				0			0	0					0		5		5	5	5
CY				0		1	1	1	162	146	63		371	9	1		10	381	382
EL				0		1	1	1	5	10			15	4			4	19	20
ES				0	2		2	2	83				83	6			6	89	91
FR				0			0	0	4						1		5	5	5
IT				0	1	1	2	2		1	5		6	1	8		9	15	17
RO				0			0	0					0			3	3	3	3
Total EU	0	0	0	0	3	3	6	6	254	157	68	0	479	20	15	3	38	517	523
Total non-EU	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
Total	0	0	0	0	3	3	6	6	254	157	68	0	479	20	15	3	38	517	523



Focusing on the last 10 years, the evolution in the number of scrapie cases detected at EU level is shown for each species and by case type in Figure 2. After the 2006 peak in the number of reported scrapie cases in sheep with 2,596 CS cases (when the number of tests also peaked), CS cases have decreased from 1,444 in 2011 to 554 in 2016. An increase was observed in 2017 (839 CS and unknown cases), mainly due to CS cases from TSE-infected flocks in Greece, Italy, Romania and Spain, and a similar order of magnitude has been observed in 2018 (821 cases).

Although Spain and Romania increased the overall number of CS scrapie cases by 13% and 267%, respectively, compared to the previous year, the increase was due to a 19.3% increase in the detection in TSE-infected flocks and a 8.3% in the index cases in Spain; and a 526.4% increase in the detection in TSE-infected flocks and a 42.8% decrease in the index cases in Romania. In goats, the decreasing trend in CS cases is less clear. The evolution is mainly affected by one single MS (Cyprus), where the number of detected cases has consistently declined since the peak in 2013 when 1,678 cases were reported by Cyprus and 1,799 in total including all other MS. Since then, the total number of cases has decreased consistently to 381 in the reporting year, which indicates a 4.4-fold reduction in the number of scrapie cases in goats reported by Cyprus in a 5-year period.

Based on the 18,929 cases of scrapie with known type, species and age between 2009 and 2018, in sheep, the average age of AS cases (83.1 months) is significantly higher (p < 0.001) than that of CS cases (50.1 months). Similarly, in goats the average age of AS cases (85.4 months) is significantly higher (p < 0.001) than that of CS cases (51.9 months). When comparing sheep with goats, there is no significant difference in the average age for AS (85.4 months in goats and 83.1 months in sheep) (p = 0.54), with the average age of CS cases in sheep (50.1 months) being very similar to that of goats (51.9 months).

Tables 18 and 19 show the cases of CS and AS, respectively, in sheep for the period 2002-2018, with a focus on the last seven years. Tables 20 and 21 show the cases of CS and AS, respectively, in goats for the period 2002-2018, with a focus on the last seven years.

In sheep, in 2018 the number of index cases of CS and AS per 10,000 tests carried out by target group at EU level was: (1) for CS: 2.9 in NSHC, 2.5 in SHC and 800 in SU; (2) for AS: 5.1 in NSHC, 1.7 in SHC and 66.6 in SU.

In goats, in 2018 the number of index cases of CS and AS per 10,000 tests carried out by target group at EU level was: (1) for CS: 3.7 in NSHC, 2.1 in SHC and 1,071.4 in SU; (2) for AS: 0.5 in NSHC and 0.4 in SHC.

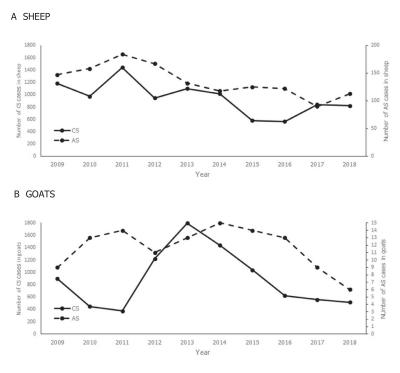


Figure 2: Number of reported scrapie cases in the EU by case type in the period 2009-2018 in (A) sheep and (B) goats



Table 18: Number of classical scrapie cases in sheep by year and reporting country between 2002 and 2018

Country code	up to 2011	2012	2013	2014	2015	2016	2017	2018	Total CS
BE	38								38
BG	6	2		3			1	5	17
CY	3,142	9	8	25	13	7	2	4	3,210
CZ	56								56
DE	115				1				116
EL	3,542	565	601	557	252	227	247	179	6,170
ES	862	33	48	36	69	91	247	279	1,665
FR	1,497	2	4	28	1	2			1,534
HU	8		1	1					10
IE	539	8	7	19	1	1	11	1	587
IT	1,682	192	260	241	141	143	240	150	3,049
NL	399		2						401
PT	19	1	6			7			33
RO	184	127	154	95	98	75	76	203	1,012
SI	176								176
SK	98			6	3	10	15		132
UK	1,981	6	6		2				1,995
Total EU	14,344	945	1,097	1,011	581	563	839	821	20,201
IS	173			2	29	11	1	21	237
NO	13								13
Total non-EU	186	0	0	2	29	11	1	21	250
Total	14,530	945	1,097	1,013	610	574	840	842	20,451

EU and reporting countries without classical scrapie cases in sheep are not included in the table. The table with all historical cases can be found on https://doi.org/10.5281/zenodo.1436520.

Table 19: Number of atypical scrapie cases in sheep by year and country between 2002 and 2018 in the reporting countries

Country	up to 2011	2012	2013	2014	2015	2016	2017	2018	Total AS
AT	4	3	2	2	1	1	1	1	15
BE	8								8
BG	2	2				2			6
CZ	1			1	3	2	1		8
DE	80	8	7	10	10	5	4	4	128
DK	12					1		1	14
EE	2								2
EL	15	5	3	5	2	2			32
ES	142	20	18	6	12	13	12	9	232
FI	8	1	1	1		2		2	15
FR	507	22	10	6	5	4	3	6	563
HR			1		1				2
HU	50	11	9	22	14	23	14	13	156
IE	12	4	4	7	7	1	1	8	44
IT	66	5	7	2	6	5	3	8	102
NL	12	5	1						18
PL	10	2	5	13	9	8	7	6	60
PT	461	44	37	20	30	28	29	30	679



Country	up to 2011	2012	2013	2014	2015	2016	2017	2018	Total AS
SE	26	3	3	7	3	3	2	2	49
SI	3		1	1	2	3			10
SK	9	3	4	3	3	5	5	6	38
UK	245	28	18	11	17	14	12	17	362
Total EU	1,675	166	131	117	125	122	94	113	2,543
IS	5		2		1				8
NO	87	6	12	9	10	14	13	8	159
Total non-EU	92	6	14	9	11	14	13	8	167
Total	1,767	172	145	126	136	136	107	121	2,710

EU and reporting countries without atypical scrapie cases in sheep are not included in the table.

Table 20: Number of classical scrapie cases in goats by year and country between 2002 and 2018 in the reporting countries

Country	up to 2011	2012	2013	2014	2015	2016	2017	2018	Total CS
BG	4				1	2	2	5	14
CY	4,635	1,116	1,678	1,364	923	570	484	381	11,151
EL	349	69	68	31	22	11	25	19	594
ES	58	3	2	8	16	19	34	89	229
FI	8								8
FR	98	5	25		40			5	173
IT	51	7	7	7	21	8	8	15	124
RO	2	1	3	1	1	3	2	3	16
SI	4								4
UK	139	21	16	26	16	8	3		229
Total EU	5,348	1,222	1,799	1,437	1,040	621	558	517	12,542
Total non-EU									0
Total	5,348	1,222	1,799	1,437	1,040	621	558	517	12,542

EU and reporting countries without classical scrapie cases in goats are not included in the table.

Table 21: Number of atypical scrapie cases in goats by year and country between 2002 and 2018 in the EU and other reporting countries

Country	up to 2011	2012	2013	2014	2015	2016	2017	2018	Total AS
AT				1					1
CY					1		1	1	3
DE				1			1		2
EL			1	1	1	1		1	5
ES	25	3	4	7	5	5	2	2	53
FI	1								1
FR	34	6	3	5	5	3	2		58
IT	14		3		1	3	3	2	26
PT	8	2	2			1			14
SI					1				1
UK									0
Total EU	82	11	13	15	14	13	9	6	163
NO	1								1
Total non-EU	1								1
Total	83	11	13	15	14	13	9	6	164

EU and reporting countries without atypical scrapie cases in goats are not included in the table.

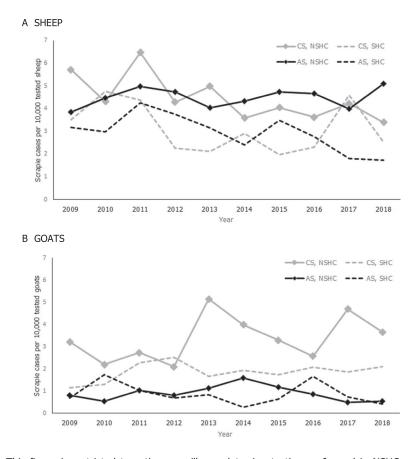


The number of historical reported scrapie cases can be found in the following link https://doi.org/10.5281/zenodo.1436520, as follows

- Number of classical scrapie cases in sheep per country and year from 2001 in the EU and non-EU reporting countries.
- Number of atypical scrapie cases in sheep per country and year from 2001 in the EU and non-EU reporting countries.
- Number of classical scrapie cases in goats per country and year from 2001 in the EU and non-EU reporting countries.
- Number of atypical scrapie cases in goats per country and year from 2001 in the EU and non-EU reporting countries.

Over the last 10 years (2009–2018), the proportion of cases per 10,000 tested animals for both CS and AS and for both species ranged between 1 and 6. Figure 3 shows the ten-year evolution by target group of the number of scrapie cases per 10,000 tests of sheep and goats in TSE non-infected flocks/ herds. Based on those data, the outputs of the Poisson regression model did not detect any statistically significant trend for both ovine and caprine AS (p = 0.12 and 0.52, respectively). For CS, there is a statistically significant decreasing trend in sheep (annual RR = 0.95, i.e. an average 5% annual decrease in the probability of detecting CS: p < 0.0001) and a statistically significant increasing trend in goats (RR = 1.04, i.e. an average 4% annual increase in the probability of detecting CS per year, p = 0.03).

Based on the same model, the probability of detecting CS in the NSHC surveillance target group was higher than in the SHC one in both sheep and goats (RR: 1.5, p < 0.001 in sheep and RR: 1.8, p < 0.0001 in goats). In AS, a statistically significant higher probability was only observed in sheep (RR = 1.4, p < 0.0001).



Note: This figure is restricted to active surveillance data, i.e. testing performed in NSHC and SHC target groups from non-infected flocks/herds or not previously known as infected. CS: classical scrapie; AS: Atypical scrapie; NSHC: animals not slaughtered for human consumption; SHC: animals slaughtered for human consumption.

Figure 3: Number of scrapie (index) cases per 10,000 tests in the EU in (A) sheep and (B) goats in non-TSE- infected flocks/herds, reported by case type and target group in the period 2009–2018



Tables 22 and 23 summarise the number of discriminatory tests performed by country in 2018 for both sheep and goats. In sheep, 526 (64.1%) of the CS cases reported in the EU were submitted for discriminatory testing and so were 17 of the AS cases (15%). The eight cases of AS reported by Norway were also submitted to discriminatory testing. All sheep scrapie cases submitted for discriminatory testing were confirmed as 'BSE excluded', except eight cases reported by Italy that were declared 'inconclusive'. In goats, 125 (24.2%) of the CS cases reported in the EU were submitted for discriminatory testing and 5 of the AS cases (83.3%). All goat cases subjected to discriminatory testing were confirmed as 'BSE excluded'.



Table 22: Number of discriminatory tests and results in sheep in 2018 by reporting country

					Ca	ses submitted 1	or discr	iminatory testing		
Country	Total cases	No. of classical scrapie	No. of atypical scrapie	BSE-not- excluded	BSE- excluded	Inconclusive	Total	% of total classical scrapie cases ^(a)	% of total atypical scrapie cases ^(b)	Cases not submitted for discriminatory testing or blank
AT	1		1				0	0%	0%	1
BG	5	5			1		1	20.0%	0%	4
CY	4	4					0	0%	0%	4
DE	4		4				0	0%	0%	4
DK	1		1				0	0%	0%	1
EL	179	179			96		96	53.6%	0%	83
ES	288	279	9		288		288	100%	100%	0
FI	2		2				0	0%	0%	2
FR	6		6				0	0%	0%	6
HU	13		13				0	0%	0%	13
IE	9	1	8				0	0%	0%	9
IT	158	150	8		158	8 ^(c)	166	100%	100%	0
PL	6		6				0	0%	0%	6
PT	30		30				0	0%	0%	30
RO	203	203					0	0%	0%	203
SE	2		2				0	0%	0%	2
SK	6		6				0	0%	0%	6
UK	17		17				0	0%	0%	17
Total EU	934	821	113	0	543	8	551	64.1%	15%	391
IS	21	21					0	0%	0%	21
NO	8		8		8		8	0%	100%	0
Total non-EU	29	21	8	0	8	0	8	0%	100%	21
Total	963	842	128	0	551	8	559	62.3%	20.7%	412

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

⁽b): Indicates the proportion of atypical TSE cases that are submitted to discriminatory testing by each reporting country.

⁽c): The eight inconclusive cases are not included in the EU total number of sheep scrapie cases of 2018 and in the calculation of the % of total classical/atypical scrapie cases. Note: only the reporting countries in which scrapie cases were detected in 2018 are mentioned in the table.



Table 23: Number of discriminatory tests and results in goats in 2018 by reporting country

					Cas	ses submitted	for disci	riminatory testing		
Country	Total cases	No. of classical scrapie	No. of atypical scrapie	BSE-not- excluded	BSE- excluded	Inconclusive	Total	% of total classical TSE cases ^(a)	% of total atypical TSE cases ^(b)	Cases not submitted for discriminatory testing or blank
BG	5	5					0	0%	0%	5
CY	382	381	1		11		11	2.6%	100%	371
EL	20	19	1		6		6	31.6%	0%	14
ES	91	89	2		91		91	100%	100%	0
FR	5	5			5		5	100%	0%	0
IT	17	15	2		17		17	100%	100%	0
RO	3	3					0	0%	0%	3
Total EU	523	517	6	0	130	0	130	24.2%	83.3%	393
Total non-EU								·		
Total	523	517	6	0	130	0	130	24.2%	83.3%	393

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

Note: only the reporting countries in which scrapie cases were detected in 2018 are mentioned in the table.

⁽b): Indicates the proportion of atypical TSE cases that are submitted to discriminatory testing by each MS reporting country.



3.2.1. Genotyping in sheep

The classification of genotypes of the sheep prion protein PRNP gene used in this report and based on to the Great Britain's National Scrapie Plan (NSP) is summarised in Table 24.

Table 24: 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 25 shows the genotypes of sheep scrapie cases in 2018 in the EU and other reporting countries.

In total, 777 (98%) of the 793 cases of CS in sheep with known genotype reported in the EU in 2018 (94.6% of the total CS caseload) were from the susceptible genotype groups (NSP3, NSP3O, NSP4 and NSP5). This is very similar to previous years in which over 95% of all CS cases with known genotypes were of the susceptible groups. In non-MS reporting countries, all CS cases reported by Iceland were of the susceptible genotype groups.

For AS, the same genotype groups (NSP3, NSP3O, NSP4 and NSP5) accounted for 46.7% of all cases with known genotype, lower than in 2017 (57.6%). Figure 4 shows the frequency distribution of genotypes of sheep scrapie cases by case type, year and NSP group in the period 2009–2018 in the reporting countries.

Table 26 shows the genotypes obtained in 2018 from the random samples of tested sheep in the reporting countries. The number of MS that have not reported any random genotypes has increased from 8 in 2017 to 20 in 2018, due to the changes in the legislation. The eight MS that conducted the genotype to a random sample of sheep were: Belgium, Cyprus (where genotyping is conducted systematically in the breeding sheep population), France, Greece, Italy, Latvia, The Netherlands and Poland. One of the four non-MS reporting country, Iceland, also reported random genotype results. In the subset of EU MS that carried out the activity in 2018, excluding data from Cyprus, 18.7% of the sheep population is susceptible to CS (NSP3, NSP3O, NSP4 and NSP5), lower than the 26.5% in 2017. This percentage rose to 42.4% in Greece and 31.1% in Italy, two of the countries with the highest caseload in 2018.

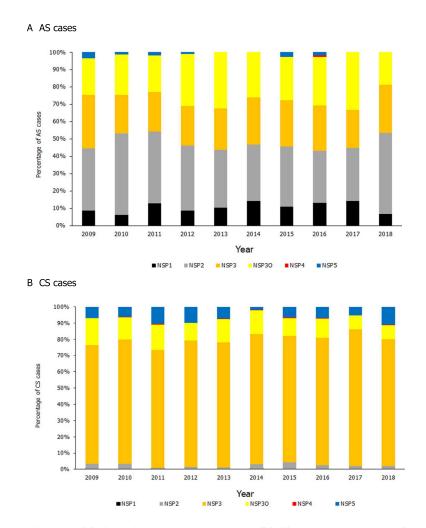
Considering the past 10 years of random sampling, the proportion of sheep in the resistant genotype group (NSP1; black colour in the bars of Figure 5) shifted from 27.8% of the total number of genotyped sheep in 2008 (in which 21 MS contributed) to 50.8% in 2018 (on which 7 MS contributed).



Table 25: Distribution of genotypes of confirmed scrapie cases in sheep in 2018 by reporting country and National Scrapie Plan (NSP) group

Type NSP				Atypic	al scrapi	e						Classica	al scrapi	е		
group/ Country	NSP1	NSP2	NSP3	NSP30	NSP4	NSP5	Unknown	Total	NSP1	NSP2	NSP3	NSP30	NSP4	NSP5	Unknown	Total
AT			1					1								0
BG								0							5	5
CY								0		3					1	4
DE	1	2					1	4								0
DK							1	1								0
EL								0		9	126	28		8	8	179
ES		3	2	3			1	9		3	255	8			13	279
FI			1				1	2								0
FR		3					3	6								0
HU		4	5	4				13								0
IE		4	2	1			1	8			1					1
IT	1	5		1			1	8			122	25		2	1	150
PL	1	3		2				6								0
PT		7	9	1			13	30								0
RO								0		1	115	8	1	78		203
SE	1	1						2								0
SK		3	1	1			1	6								0
UK	2	7	4	4				17								0
Total EU	6	42	25	17	0	0	23	113	0	16	619	69	1	88	28	821
IS								0			14	1		3	3	21
NO		2	1	5				8								0
Total non-EU	0	2	1	5	0	0	0	8	0	0	14	1	0	3	3	21
Total	6	44	26	22	0	0	23	121	0	16	633	70	1	91	31	842





(A) Atypical scrapie; (B) Classical scrapie. NSP1: Resistant (black); NSP2: Semi-resistant (grey); NSP3 (orange) + NSP3O (yellow) + NSP4 (red) + SNP5 (purple): susceptible as referred to in Table 24.

Figure 4: Frequency distribution of genotypes of sheep scrapie cases by case type, year and NSP group in the period 2009–2018 in the reporting countries

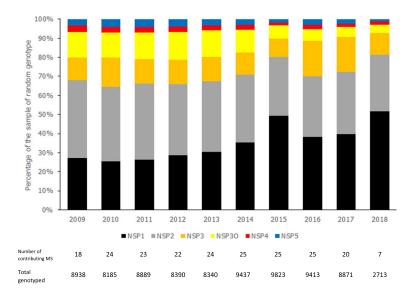


Table 26: Number of genotyped animals (% of sample within country) in randomly selected sheep in the EU and other reporting countries in 2018 by reporting country and National Scrapie Plan (NSP) group, in accordance with Regulation (EC) 999/2001 Annex III, Chapter A, Part I, point 8.2

	Nu	mber of ger	осурей анн	11415 (70 01 1	sample with	iiii Counti y		
Country	NSP1	NSP2	NSP3	NSP30	NSP4	NSP5	Other/ Unknown	Total
AT	not done	not done	not done	not done	not done	not done	not done	0
BE	159 (75.4%)	41 (19.4%)	3 (1.4%)	4 (1.9%)	2 (0.9%)	2 (0.9%)	0 (0%)	211
BU	not done	not done	not done	not done	not done	not done	not done	0
CY ^(a)	60,477 (89.4%)	6,083 (9%)	175 (0.3%)	126 (0.2%)	366 (0.5%)	45 (0.1%)	380 (0.6%)	67,652
CZ	not done	not done	not done	not done	not done	not done	not done	0
DE	not done	not done	not done	not done	not done	not done	not done	0
DK	not done	not done	not done	not done	not done	not done	not done	0
EE	not done	not done	not done	not done	not done	not done	not done	0
EL	26 (16.5%)	56 (35.4%)	41 (25.9%)	18 (11.4%)	5 (3.2%)	3 (1.9%)	9 (5.7%)	158
ES	not done	not done	not done	not done	not done	not done	not done	0
FI	not done	not done	not done	not done	not done	not done	not done	0
FR	204 (60.5%)	91 (27%)	29 (8.6%)	0 (0%)	9 (2.7%)	4 (1.2%)	0 (0%)	337
HR	not done	not done	not done	not done	not done	not done	not done	0
HU	not done	not done	not done	not done	not done	not done	not done	0
IE	not done	not done	not done	not done	not done	not done	not done	0
IT	154 (22.2%)	321 (46.3%)	142 (20.5%)	46 (6.6%)	15 (2.2%)	13 (1.9%)	3 (0.4%)	694
LT	not done	not done	not done	not done	not done	not done	not done	0
LU	not done	not done	not done	not done	not done	not done	not done	0
LV	63 (70%)	24 (26.7%)	2 (2.2%)	1 (1.1%)	0 (0%)	0 (0%)	0 (0%)	90
MT	not done	not done	not done	not done	not done	not done	not done	0
NL	736 (65.7%)	236 (21.1%)	82 (7.3%)	35 (3.1%)	23 (2.1%)	8 (0.7%)	0 (0%)	1,120
PL	36 (36%)	44 (44%)	10 (10%)	10 (10%)	0 (0%)	0 (0%)	0 (0%)	100
PT	not done	not done	not done	not done	not done	not done	not done	0
RO	not done	not done	not done	not done	not done	not done	not done	0
SE	not done	not done	not done	not done	not done	not done	not done	0
SI	not done	not done	not done	not done	not done	not done	not done	0
SK	not done	not done	not done	not done	not done	not done	not done	0
UK	not done	not done	not done	not done	not done	not done	not done	0
Total EU	61,855 (87.9%)	6,896 (9.8%)	484 (0.7%)	240 (0.3%)	420 (0.6%)	75 (0.1%)	392 (0.6%)	70,362
СН	not done	not done	not done	not done	not done	not done	not done	0
IS	0 (0%)	0 (0%)	147 (42.5%)	57 (16.5%)	0 (0%)	142 (41.0%)	0 (0%)	346
MK	not done	not done	not done	not done	not done	not done	not done	0
NO	not done	not done	not done	not done	not done	not done	not done	0
Total non-EU	0 (0%)	0 (0%)	147 (45.1%)	57 (17.5%)	0 (0%)	142 (37.4%)	0 (0%)	346
Total	61,855 (87.5%)	6,896 (9.8%)	631 (0.9%)	297 (0.4%)	420 (0.6%)	217 (0.3%)	392 (0.6%)	70,708

⁽a): Data of Cyprus are different from those of other reporting countries since Cyprus systematically genotypes the breeding sheep population.





Data from Cyprus were excluded. NSP1: Resistant (black); NSP2: Semi-resistant (grey); NSP3 (orange) + NSP3O (yellow) + NSP4 (red) + SNP5 (purple): susceptible as referred to in Table 24.

Figure 5: Frequency distribution of the six genotype groups in sheep sampled for genotyping in the EU in the period 2009–2018 according to Regulation (EC) 999/2001 Annex III, Chapter A, Part I, point 8

3.3. TSE surveillance in cervids

In 2018, 8,185 cervids were tested for TSE in the EU by 12 MS, 6 of them subject to mandatory surveillance (hereinafter referred to as the MS6) (Section 1.2.4): Estonia, Finland, Latvia, Lithuania, Poland and Sweden with 5,110 tested cervids (62.4% of the total), and 6 MS not subject to mandatory surveillance: Austria, Denmark, Hungary, Italy, Spain and Romania, with 3,075 tested cervids (37.6% of the total).

Out of the 5,110 tested cervids by the MS6, 4,674 (91.5%) were wild animals, mostly roe deer (2,856), followed by red deer (883) and moose (878). Among the 436 (8.5%) captive, farmed or semi-domesticated cervids tested by the MS6, 296 (67.9%) were semi-domesticated reindeer tested in Finland. The number of cervids tested and positive in 2018 by species, management system and reporting country are displayed in Table 27.

As a result of the mandatory surveillance in MS6, the first case of CWD in the UE was confirmed in 2018: a wild moose in Finland.

The criteria to identify the PSU by each of the MS6 are described below:

- Estonia: 15 PSU for both wild/semi-domesticated and captive/farmed cervids, corresponding to each of the Estonian counties.
- Finland: 54 PSU for semi-domesticated reindeer, based on administrative units of the reindeer herding cooperative (RHC); 295 PSU for wild cervids based on the local game administration units according to the game management association (GMA).
- Lithuania: each farm and each facility in which cervids are kept in an enclosed territory shall be considered as a PSU, approximately 655, based on permissions issued by the Ministry of the Environment (list not available); 51 PSU for wild cervids based on State Food and Veterinary Service (SFVS) territorial units.
- Latvia: 100 PSU based on territorial units for the determination of the number of animals continuous areas with defined natural boundaries, which include one or more hunting districts as undivided as possible, and which, according to the total hunting area, are not less than 20,000 ha for wolves and lynx, 5,000 ha for moose and red deer, 1,500 ha for wild boar and 500 ha for roe deer.
- Poland: 16 PSU for both wild/semi-domesticated and captive/farmed cervids based on voivodeships (administrative units).



 Sweden: 109 PSU for farmed (one for each farm), 51 PSU for semi-domesticated reindeer corresponding to the 51 Sami villages (administrative units where reindeer are kept), and 50 PSU for wild cervids, covering all the country. PSU of different management systems geographically overlap for reindeer.

In terms of testing at PSU level, the number and proportion of PSU for wild/semi-domesticated and farmed/captive cervids declared and tested by the MS6 is shown in Table 28. There is also large variability in the proportion of PSU from which samples have been collected, which is determined by the number of PSU included in the sampling programme. For example, Poland and Estonia sampled cervids from 100% and 66.7% of the 16 and 15 PSU, respectively. Sweden tested cervids from 41 out of the 50 PSU for wild (82%) and 13 (8.1%) of the 160 for farmed and semidomesticated cervids. Finland sampled from 37.2% of the wild PSU and 79.6% of the semi-domesticated PSU. Latvia declared 100 PSU but submitted samples from 145 different PSU codes. Lithuania did not submit PSU codes for the tested cervids. Median, minimum and maximum number of cervids tested in the different types of PSU and countries of the MS6 are also displayed in Table 28.

The most common target group tested by the MS6 was the 'Hunted/slaughtered fit for human consumption' (HSHC) that accounted for 3,039 (59.5% of all tested cervids). All the rest were risk animals in the different target groups: clinical suspect animals (SU): 34; road/predator killed (RK): 1,243; fallen/culled (FC): 719; hunted/slaughtered not fit for human consumption (HSNHC): 75. There is a large variability between MS6 countries in the proportion of cervids tested in the HSHC, ranging from 5% tested by Sweden to 94.2% by Latvia or 84.8% by Lithuania. The numbers of tested cervids by reporting country, management system and target group in 2018 are displayed in Table 29.

Among the non-MS6, Romania accounted for 77.6% of all tested cervids, followed by Italy with 585 (19%). The majority of the tested cervids were roe deer (75.6%), followed by red deer (22.1%). All tested cervids by the non-MS6 resulted negative.

Norway continued its intensified testing programme in wild and captive cervids and tested 33,037 animals in 2018, mostly semi-domesticated reindeer (36.4%), followed by red deer (23.5%) and wild moose (20.1%), mainly from the hunted/slaughtered for human consumption target group, leading to the detection of six reindeer cases and one moose case. Iceland also reported 100 tested cervids in 2018, all negative.



Table 27: Number of cervids tested in the reporting countries in 2018 by management system, species and country^(a)

			W	ILD DE	ER SPE	CIES ^(b)					SE	MI-DOM	ESTIC	CATED/I	FARMI	ED DEEI	R SPECIES	(b)	
Country ^{(c),(d)}	Eurasian tundra reindeer	Finnish (Eurasian) forest reindeer	Moose	Roe deer	White- tailed deer	Red deer		Other or unknown	Sun-Intai	Eurasian tundra reindeer	Finnish (Eurasian) forest reindeer	Moose	Roe deer	White- tailed deer	Red deer		Other or unknown	Sub- Total	Total
AT									0						4			4	4
DK				47		3	8		58									0	58
EE			84	127		1			212				1		4			5	217
ES				5		32			37									0	37
FI		12	238 (1)	61	44				355 (1)	296		4	2	6				308	663 (1)
HU				1		3			4									0	4
IT				446		96	20		562		6		3		3	11		23	585
LT			119	1268	1	388			1,776			3	19		35		2	59	1,835
LV			239	510		285			1,034						20			20	1,054
PL			42	875		201			1,118						23			23	1,141
RO				1,796		526	15	10	2,347				26		14			40	2,387
SE			156	15		8			179	15		1			5			21	200
TOTAL EU	0	12	878 (1)	5151	45	1543	43	10	7,682 (1)	311	6	8	51	6	108	11	2	503	8,185 (1)
IS	100								100										100
NO	3,624 (6)		6,643 (1)	2,106		7,762	4	164	20,403 (7)	12,043		1			639	42	9	12,734	33,037 (7)
TOTAL non-EU	3,724 (6)	0	6,643 (1)		0	7,762	4	164	20,403 (7)	12,043	0	1	0	0	639	42	9		33,137 (7)
TOTAL	3,724 (6)	12	7,521 (2)	7,257	45	9,305	47	174	28,085 (8)	12,354	6	9	51	6	747	53	11	13,237	41,322 (8)

⁽a): Number of positives for chronic wasting disease (CWD) are given in brackets.

⁽b): Eurasian tundra reindeer (Rangifer tarandus tarandus); Finnish (Eurasian) forest reindeer (Rangifer tarandus fennicus); Moose (or Eurasian/European elk) (Alces alces alces); Roe deer (Capreolus capreolus); White-tailed deer (Odocoileus virginianus); Red deer (Cervus elaphus); Fallow deer (Dama dama); Other or Unknown.

⁽c): Only countries that reported tested cervids are included in the table.

⁽d): Shaded in grey: MS conducting mandatory surveillance from 2018.



Table 28: Number of PSU by management type in the six MS with mandatory CWD surveillance in 2018

Management system		PSU (WI	LD)	PSU (CAP	TIVE/FARMED	DEER)		Total PSU	
Country	Number of PSU declared	Number of PSU tested (%)	Median number of cervids tested (min-max)	Number of PSU declared	Number of PSU tested (%)	Median number of cervids tested (min-max)	Number of PSU declared	Number of PSU tested (%)	Median number of cervids tested (min-max)
EE	15	9 (60%)	5 (1–78)	15	2 (13.3%)	3 (1–4)	15	10 ^(c) (66.7%)	4 (1–78)
FI	295	110 (37.3%)	1 (1–31)	54 ^(d)	43 (79.6%)	3 (1–44)	349	153 (43.8%)	2 (1–44)
LT	51		Not available	655 ^(a)	Not available			Not available	
LV		139	5 (1–20)		6		100	145 (145%)	4 (1–20)
PL	16	16 ^(b) (100%)	63 (8–197)	16	3 (18.75%)	8 (5–10)	16	16 (100%)	63 (8–197)
SE	50	41 (82%)	3 (1–15)	160 (109 (farmed) + 51 semi- domesticated)	13 (8.1%)	1 (1–4)	210	54 (25.7%)	2 (1–13)

⁽a): Each farm and each facility in which cervids are kept in an enclosed territory shall be considered as a PSU. There are approximately 655 permissions issued by the Ministry of the Environment for keeping wild animals, including cervids, but the active number (with cervids) is unknown.

Table 29: Number of tested cervids in the EU and reporting countries by management system, species, country and target group in 2018

				V	VILD D	EER SPE	CIES					SEMI-D	OMESTI	CATE	D/FARM	ED DE	ER SPE	CIES		
Manage System Countr	l v/Species	Eurasian tundra reindeer ^(a)	Finnish (Eurasian) forest reindeer	Moose	Roe deer	White- tailed deer	Red deer		Other or unknown	Sub- Total	Eurasian tundra reindeer	Finnish (Eurasian) forest reindeer	Moose	Roe deer	White- tailed deer	Red deer		Other or unknown		Total
AT ^(b)	FC ^(c)															4			4	4
	Total															4			4	4
DK	FC				47		3	8		58										58
	Total				47		3	8		58										58
EE	SU			1	1					2										2
	RK			5	66					71										71
	FC			9	22		1			32				1		4			5	37
	HSNHC			2	6					8										8
	HSHC			67	32					99										99
	Total			84	127	0	1	0	0	212				1		4			5	217

⁽b): the PSU are not split by management systems.

⁽c): In one PSU wild and farmed animals were both sampled.

⁽d): Semi-domesticated PSU.



				V	VILD D	EER SPE	CIES					SEMI-D	OMESTI	CATE	D/FARM	ED DE	ER SPE	CIES		
Syste	gement m try/Species	Eurasian tundra reindeer ^(a)	Finnish (Eurasian) forest reindeer	Moose	Roe deer	White- tailed deer	Red deer	Fallow deer	Other or unknown	Sub- Total	Eurasian tundra reindeer	Finnish (Eurasian) forest reindeer		Roe deer	White- tailed deer	Red deer		Other or unknown	Sub- Total	Total
ES	HSNHC				5		12			17										17
	HSHC						20			20										20
	Total				5		32			37										37
FI	RK		6	48	39	17				110	128								128	238
	FC		6	75	21	26				128	164		4	2	6				176	304
	HSNHC			16						16	4								4	20
	HSHC			99	1	1				101									0	101
	Total		12	238	61	44	0	0	0	355	296		4	2	6	0	0	0	308	663
HU	SU				1		1			2										2
	FC						2			2										2
	Total				1		3			4										4
IT	SU				30		12			42		1		1		2			4	46
	RK				321		43	18		382										382
	FC				95		41	2		138		5		2		1	11		19	157
	Total				446		96	20		562		6		3		3	11		23	585
LT	RK			8	110	1	23			142									0	142
	FC			6	85		9			100						7		2	9	109
	HSNHC				23		1			24						3			3	27
	HSHC			105	1,050		355			1,510			3	19		25			47	1,557
	Total			119	1,268	1	388			1,776	0		3	19	0	35	0	2	59	1,835
LV	SU			1	2		5			8										8
	RK			4	16		3			23										23
	FC			7	15		8			30										30
	HSHC			227	477		269			973						20			20	993
	Total			239	510	0	285	0	0	1,034						20			20	1,054
PL	SU				8		1			9										9
	RK			38	657		60			755										755
	FC			4	72		11			87						3			3	90
	HSNHC				2		1			3						5			5	8
	HSHC				136		128			264						15			15	279
	Total			42	875	0	201	0	0	1,118	0		0	0	0	23	0	0	23	1,141



				V	VILD D	EER SPE	CIES					SEMI-D	OMESTI	CATE	D/FARM	ED DE	ER SPE	CIES		
Manage System Country		Eurasian tundra reindeer ^(a)	Finnish (Eurasian) forest reindeer	Moose	Roe deer	White- tailed deer	Red deer	Fallow deer	Other or unknown	Sub- Total	Eurasian tundra reindeer	Finnish (Eurasian) forest reindeer	Moose	Roe deer	White- tailed deer	Red deer		Other or unknown	Sub- Total	Total
RO	RK				13		3			16						3			3	19
	FC				39		14			53				21		1			22	75
	HSHC				1,744		509	15	10	2,278				5		10			15	2,293
	Total				1,796		526	15	10	2,347				26		14			40	2,387
SE	SU			9	3		2			14			1						1	15
	RK			8						8	6								6	14
	FC			123	12		4			139	5					5			10	149
	HSNHC			6			2			8	4								4	12
	HSHC			10						10									0	10
	Total			156	15	0	8	0	0	179	15		1	0	0	5	0	0	21	200
TOTAL EU		0	12	878	5,151	45	1,543	43	10	7,682	311	6	8	51	6	108	11	2	503	8,185
IS	RK	2								2										2
	FC	3								3										3
	HSHC	95								95										95
	Total	100								100										100
NO	RK			463	1245		301		8	2,017	83								83	2,100
	FC	122		529	646		512		61	1,870	161		1			25	3		190	2,060
	HSNHC	446								446										446
	HSHC	3,056		5,651	215		6949	4	95	15,970	11,799					614	39	9	12,461	28,431
	Total	3,624		6,643	2,106		7,762	4	164	20,303	12,043		1			639	42	9	12,734	33,037
TOTAL non-EU		3,724	0	6,643	2,106	0	7,762	4	164	20,403	12,043	0	1	0	0	639	42	9	12,734	33,137
TOTAL		3,724	12	7,521	7,257	45	9,305	47	174	28,085	12,354	6	9	51	6	747	53	11	13,237	41,322

⁽a): Eurasian tundra reindeer (Rangifer tarandus tarandus); B: Finnish (Eurasian) forest reindeer (Rangifer tarandus fennicus); C: Moose (or Eurasian/European elk) (Alces alces alces); D: Roe deer (Capreolus capreolus); E: White-tailed deer (Odocoileus virginianus); F: Red deer (Cervus elaphus); G: Fallow deer (Dama dama); H: Other or Unknown.

⁽b): Only countries that reported tested cervids are included in the table.

⁽c): SU: clinical suspect animals; RK: Road/predator killed; FC: Fallen/culled; HSNHC: Hunted/slaughtered not fit for human consumption; HSHC: Hunted/slaughtered fit for human consumption.



3.4. Other species

Two MS, Estonia and Finland, reported results on samples tested for TSE in species other than cattle, small ruminants and cervids. In total, 105 samples were collected and tested from the following species: raccoon dog (*Nyctereutes procyonoides*), american mink (*Neovison vison*) and fox (genus *Vulpes*). None of them tested positive (Table 30).

Table 30: Numbers of animals in species other than cattle, sheep, goats and cervids tested for TSE in reporting countries in 2018

Country	Raccoon dog (Nyctereutes procyonoides)	Fox (genus Vulpes)	American mink (Neovison vison)	Total
Estonia			6	6
Finland	13	33	53	99
Total	13	33	59	105

4. Conclusions

As part of the BSE surveillance system in cattle in the EU, nearly 1.2 million cattle were tested in 2018, 10% less than in the previous year. The testing throughput combined with a risk-based strategy (about 84.9% of all tests were targeting risk animals) contributed to maintain the sensitivity of the BSE surveillance system considering the EU as a single epidemiological unit. That allowed the consolidation of the current situation in which even few cases are emerging they remain detectable. Three atypical cases of BSE (2 L-BSE and 1 H-BSE cases) were confirmed in 2018 by one single reporting country: France. From an epidemiological point of view, no statistical departure from known trends has been detected in 2018 with the atypical cases being similar to those recently reported both in number and epidemiological features, except one of the H-BSE that was confirmed in a 92-month old cow, 3 years younger than the average age for atypical BSE cases. One C-BSE case was confirmed in a 65-month Aberdeen angus beef cow, born in April 2013 (BARB case). An additional H-BSE case was reported by the USA

Over 460,000 small ruminants were tested in 2018 in the EU, as part of the TSE surveillance system, leading to an overall testing of more than 9.5 million tests since 2002. Nineteen MS complied with the EU monitoring requirements in sheep and 22 MS in goats. Compared with 2017, there was a decrease in the detection of the disease in non-infected flocks (from 8.1 to 7 cases per 10,000 - 13.6% in sheep) despite the same level of testing in non-infected flocks as in 2017. This may be consistent with a decrease in the overall incidence of the disease. In the same period in goats, there was a similar decrease in the detection of the disease in non-infected herds (from 4.3 to 3.5 cases per 10,000 tests: -18.6%), despite the increase in the testing in non TSE-infected herds, due to the low number of index cases reported by Cyprus (11).

For CS in sheep, compared with 2017, the increase in the testing activity has not resulted in an increase in the EU caseload, in the proportion of cases per 10,000 tested animals or in the number of index cases. Overall, the four countries with the largest number of CS cases, namely Greece, Italy, Romania and Spain, have reported less cases compared to 2017, and index cases in particular. However, Spain and Romania increased the number of CS scrapie cases by 13% and 167%, respectively. From a geographical point of view, the disease is reported by a minority of the MS (seven in 2018, compared to eight and nine in 2017 and 2016, respectively) and, as in 2017, only a very small proportion (1.2%) of the CS caseload is from MS other than Greece, Spain, Italy and Romania.

In goats, compared with 2017, the EU caseload of CS showed a 7.3% decrease mainly due to the improved situation in Cyprus whose caseload accounted for 73.7% of the total.

When looking at the long-term trends of CS in terms of cases per 10,000 tests in both species, the situation in 2018 confirmed the 10-year statistically significant decreasing trend in sheep and increasing trend in goats, respectively, as estimated through modelling of the available epidemiological data. The disadvantage of the control of the disease in goat herds, due to the lack of both breeding for resistance programmes and genetic-based culling and restocking of infected herds, may explain to a certain extent this long-term trend.

For AS in sheep, compared with 2017 the above described testing activity resulted in an increase of the caseload, in the proportion of cases per 10,000 tested animals, in the number of index cases and the number of MS reporting cases. The opposite occurred in goats where the increase in the testing



activity resulted in the decrease of the caseload, in the proportion of cases per 10,000 tested animals and in the number of index cases. These changes could be due to the annual variability and there is no indication of changes in the long-term trends.

The genotyping data collected in 2018 from ovine CS cases consistently confirms the association between the occurrence of the disease and the susceptible genotypes (NSP3, NSP3O, NSP4 or NSP5), with 98% of the cases carrying them. The 2018 genotyping data from random samples of the EU sheep population (after excluding Cyprus) did show an improvement compared with the previous years with an average 19.2% of the genotyped sheep carrying genotypes of the susceptible group. Some caution is needed in interpreting this result as it could reflect the implementation of the amendment of the TSE Regulation with the subsequent reduction in the number of contributing MS.

The enforcement of the 3-year surveillance programme for CWD in six MS - Estonia, Finland, Latvia, Lithuania, Poland and Sweden – resulted in the testing of 5,110 cervids and the confirmation of the first case of CWD in the EU: a wild moose in Finland. The surveillance was complemented by the additional 3,075 cervids tested by other six MS (77.6% of them tested by Romania) with no additional cases detected. The implementation of the mandatory surveillance in the six MS is quite heterogeneous in terms of the design (number and characteristics of the declared PSU), the number of cervids tested in general and per PSU in particular and the distribution of testing by species and target groups. The low average number of cervids tested per PSU and the targeting of mostly hunted/slaughtered fit for human consumption (HSHC) animals may have resulted in a lower than expected sensitivity of the surveillance system. No conclusions can be drawn about the presence of the disease in other MS and about the prevalence in the affected MS.

Norway continued its intensified testing programme in wild and captive cervids and tested 33,037 animals in 2018, leading to the detection of six reindeer cases and one moose case. At the end of the reporting year and since 2016, Norway had confirmed in total 19 cases in reindeer, 4 in moose and 1 in red deer. Iceland also reported 100 tested cervids in 2018, all negative.

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Abbreviations

AM *ante-mortem*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

EM eradication measures ES emergency slaughtered

EURL European Union Reference Laboratory
EUSR European Union summary report

FS fallen stock

GMA game management association

H-BSE H-type bovine spongiform encephalopathy

HS healthy slaughtered

HSHC hunted/slaughtered fit for human consumption hunted/slaughtered not fit for human consumption

L-BSE L-type bovine spongiform encephalopathy

MS Member State/s

NSHC not slaughtered for human consumption

NSP National Scrapie Plan

RHC reindeer herding cooperative

RK road/predator killed

RR relative risk

SFVS State Food and Veterinary Service SHC slaughtered for human consumption

SU clinical suspect

TSE transmissible spongiform encephalopathies

AT

Country codes

Austria

BE Belgium Bulgaria BG Croatia HR Cyprus CY The Czech Republic CZ DK Denmark Estonia EE Finland FΙ France FR Germany DE Greece EL Hungary HU Iceland IS Ireland ΙE Italy ΙT Latvia LV Lithuania LT Luxembourg LU Malta MT The Netherlands NL North Macedonia MK Norway NO Poland PLPortugal PT Romania RO Slovakia SK Slovenia SI Spain ES Sweden SE



Switzerland CH The United Kingdom UK

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.

Non-MS countries: CH (including Lichtenstein); IS; MK, NO.



Appendix A - Additional surveillance data

Table A.1: BSE active monitoring in relation to the adult bovine population (age > 2 years) in 2018

EU/non-EU groups	Country code	Adult cattle (> 2 year) ^(a)	Number of tested bovine animals at risk ^(b)	Proportion (%) of tested bovine animals at risk ^(b)
EU ^(a)	AT	866,210	18,450	2.1%
	BE	1,209,740	25,948	2.1%
	BG	375,230	3,879	1.0%
	CY	34,870	1,519	4.4%
	CZ	657,340	21,714	3.3%
	DE	5,536,320	171,455	3.1%
	DK	712,000	23,837	3.3%
	EE	132,100	3,604	2.7%
	EL	305,000	939	0.3%
	ES	3,169,740	65,172	2.1%
	FI	353,930	11,312	3.2%
	FR	10,151,640	210,876	2.1%
	HR	165,000	5,034	3.1%
	HU	451,000	11,987	2.7%
	IE	2,803,930	70,212	2.5%
	IT	3,070,800	54,702	1.8%
	LT	347,600	3,030	0.9%
	LU	102,630	2,790	2.7%
	LV	224,530	5,009	2.2%
	MT	6,970	210	3.0%
	NL	1,715,000	50,401	2.9%
	PL	2,742,300	39,705	1.4%
	PT	899,440	20,844	2.3%
	RO	1,322,600	8,979	0.7%
	SE	625,010	7,592	1.2%
	SI	195,850	6,632	3.4%
	SK	232,420	7,960	3.4%
	UK	4,336,000	149,915	3.5%
	Total EU	42,745,200	1,003,707	2.3%
lon-EU	СН	794,980	10,729	1.3%
	IS	38,500	77	0.2%
	MK	138,674 ^(c)	2	0.0%
	NO	353,700	6,326	1.8%
	Total non-EU	1,355,180	17,134	1.3%
TOTAL		44,100,380	1,020,841	2.3%

⁽a): Taken from https://ec.europa.eu/eurostat/documents/3217494/9455154/KS-FK-18-001-EN-N.pdf/a9ddd7db-c40c-48c9-8ed5-a8a90f4faa3f

Norway's cattle population taken from the TSE EUSR report 2017 (EFSA, 2017).

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

⁽c): Data provided by the Food and Veterinary Agency of the Republic of North Macedonia.



Appendix B – Geographical distribution of BSE in the period 2001–2018

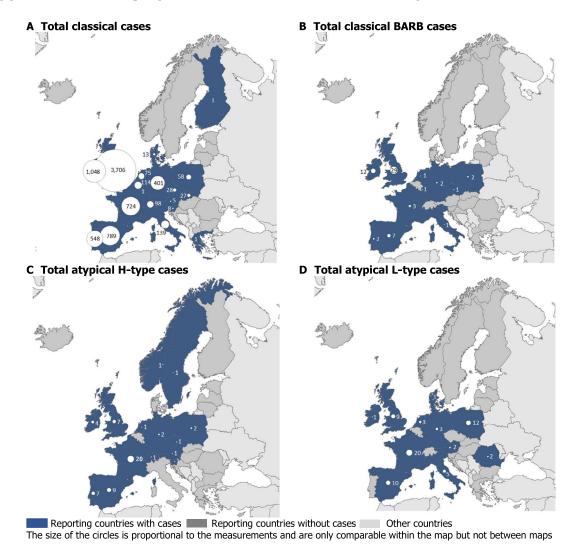


Figure B.1: Geographical distribution of cumulative number of cases of C-BSE (A); BARB cases (B); H-BSE (C); and L-BSE (D) in the period 2001–2018



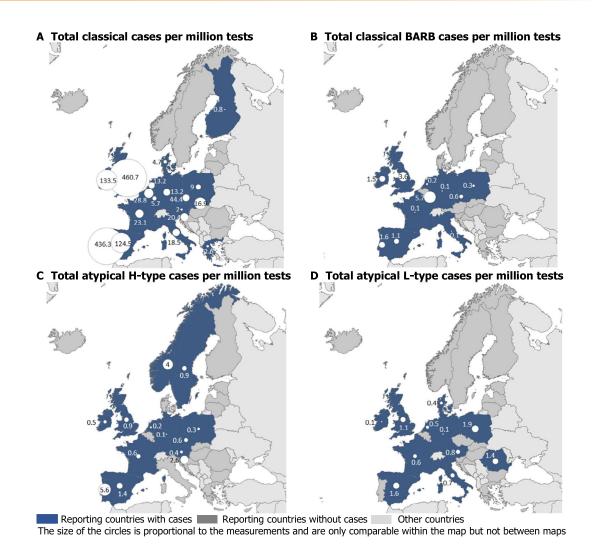


Figure B.2: Country-specific BSE cases per million tests by case type in the period 2001–2018 in the EU



Appendix C – Geographical distribution of scrapie in 2018

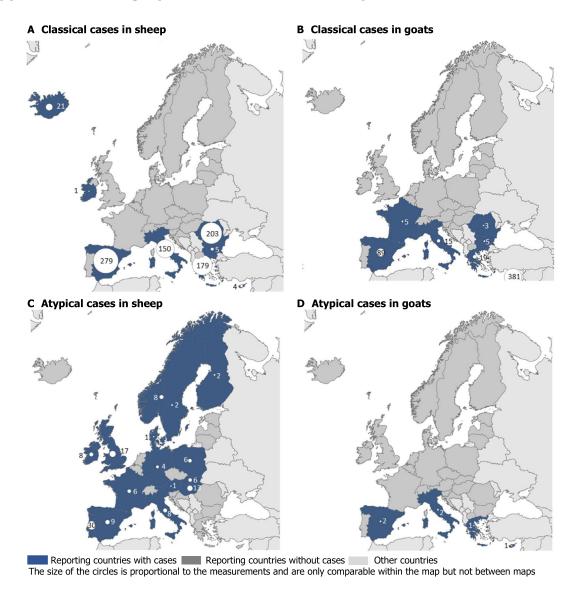


Figure C.1: Geographical distribution of numbers of cases of ovine CS (A); caprine CS (B); ovine AS (C); and caprine AS (D) in 2018 in the reporting countries



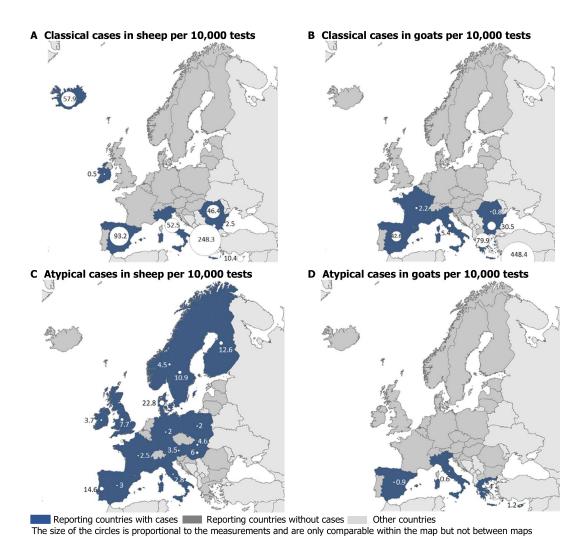


Figure C.2: Geographical distribution of proportion of cases per 10,000 tests of ovine CS (A); caprine CS (B); ovine AS (C); and caprine AS (D) in 2018



Appendix D - Additional information asked by EFSA in relation to reporting, according to Annex III of Regulation 999/2001

Table D.1: The number of suspected cases placed under official movement restrictions in accordance with Article 12(1)

Country	Cattle	Sheep	Goats	
AT	18	1	0	
BE	402	0	0	
BG	0	0	0	
CY	0	13	17	
CZ	0	0	0	
DE	0	145	0	
DK	2	2	0	
EE	0	0	0	
EL	831	3,634	1,036	
ES	348	2,847	1,805	
FI	0	0	0	
FR	4	3	1	
HR	0	0	0	
HU	2	1	0	
Œ	20	0	0	
IT	0	462	18(*)	
LV	5	0	0	
LU	3	0	0	
LT	0	0	0 0 0	
МТ	0	0		
NL	0	0		
PL	72	4	0	
PT	0	0	0	
RO	696	11,115	677	
SI	11*	10*	7*	
SE	0	0	0	
SK	0	2,627	40	
UK	1	1	0	
Total EU	2,404	20,855	3,576	
СН	0	0	0	
IS	0	0	0	
MK	3	47	0	
NO	0	0	0	
Total non-EU	3	47	0	
Total	2,407	20,902	3,576	

^{(*):} These goats were present in one of the two sheep flocks reported in Table D.2.
*: Animals with neurological clinical symptoms (differential diagnosis TSE, Rabies, Listeriosis).



Table D.2: Number of flocks where suspected cases in ovine and caprine animals have been reported and investigated pursuant to Article 12(1) and (2)

Country	Sheep	Goats	
AT	1		
BE	1	0	
BG	0	0	
CY	13	22*	
CZ	0	0	
DE	19	7	
DK	2	0	
EE	0	0	
EL	18	7	
ES	6	0	
FI	0	0	
FR	3	0	
HR	0	0	
HU	1	0	
IE	0	0	
IT	2	0	
LV	0	0	
LU	0	0	
LT	0	0	
MT	0	0	
NL	0	0	
PL	4	0	
PT	0	0	
RO	36	6	
SI	8	4	
SE	0	0	
SK	25	4	
UK	1	0	
Total EU	140	28	
СН	0	0	
IS	2	0	
МК	5	0	
NO	0	0	
Total non-EU	7	0	
Total	147	28	

^{*:} In addition, 49 mixed flocks (sheep and goats).



Table D.3: Number of ovine and caprine 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

Country	Sheep SHC	Sheep NSHC	Sheep EM	Goats SHC	Goats NSHC	Goats EM	Other ^(d)
AT	108	1,692	2	18	465	0	0
BE	0	*	0	0	*	0	
BG	1,766	105	0	417	26	0	
CY	108	706	1	220	399	12 ^(a)	0
CZ	3	1,030	0	0	162	0	no
DE	4,875	4,652	1	150	719	0	Cattle: 1,029; Sheep: 110; Goat: 17
DK	1	N/A	0	1	N/A	0	0
EE	N/A	N/A	N/A	N/A	N/A	N/A available	
EL	538	250	55	197	112	7	
ES	955	5,031	68	1,025	2,394	16	
FI	4	533	0	0	57	0	
FR	2,647	8,641	3	1,263	4,205	8 ^(b)	
HR	0	1,374	0	0	320	0	
HU	1,304	1,556	0	34	61	0	
IE	11,035	10,802	57	0	151	0	
IT	4,206	5,717	82	4,370	3,801	31	
LV	0	66	0	0	16	0	0
LU	0	103	0	0	88	0	
LT	0	186	0	0	5	0	
MT	76	138	0	65	68	0	0
NL	0	N/A	0	0	N/A	0	
PL	986	2,039	0	131	1,103	0	0
PT	1,202	10,325	120 ^(c)	2	1,138	7 ^(c)	
RO	11,035	6,044	0	3,380	2,298	0	
SI	45	1,160	0	22	355	0	
SE	N/A	N/A	0	N/A	N/A	0	
SK	0	505	11	0	55	1	0
UK	> 167	> 12,266	0	0	> 326	4	
СН	0	0	0	0	0	0	
IS	247	37	0	4	2	0	0
MK	11	0	0	0	0	0	
NO	4,084	3,051	19	14	137	0	0

EM: emergency slaughter; NSHC: not slaughtered for human consumption; SHC: slaughtered for human consumption; N/A: Not available.

^{*:} The Belgian competent authority informed that in the central database for sheep and goat identification, there is no direct link between the official Eartag number and the last holding where the sheep or goat was kept. Only the herd of birth is registered in the central database. For this reason, it is impossible to answer question 3. At the rendering plant sheep and goats are randomly sampled during the year.

⁽a): In addition, 10 mixed flocks (sheep and goats).

⁽b): Excluding flocks without number.

⁽c): Atypical Scrapie intensified surveillance.

⁽d): Monitoring in other animals (= for dairy production, OR from countries with indigenous TSE, OR animals that have consumed potentially contaminated feeding stuffs, OR animals born or derived from TSE-infected dams).