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The European Union summary report on surveillance for the presence of transmissible spongiform encephalopathies (TSE) in 2022

European Food Safety Authority (EFSA)

Correspondence: zoonoses@efsa.europa.eu

Abstract

This report presents the results of surveillance on transmissible spongiform encephalopathies (TSE) in cattle, sheep, goats, cervids and other species, and genotyping in sheep and goats, carried out in 2022 by 27 Member States (MS, EU27), the United Kingdom (in respect of Northern Ireland [XI]) and other eight non-EU reporting countries: Bosnia and Herzegovina, Iceland, Montenegro, North Macedonia, Norway, Serbia, Switzerland and Türkiye. In total, 977,008 cattle were tested by EU27 and XI (-4.3%, compared with 2021), and 52,395 cattle by eight non-EU reporting countries, with one case of H-BSE in France. In total, 295,145 sheep and 109,074 goats were tested in the EU27 and XI (-5.2% and -7.9%, respectively, compared to 2021). In the other non-EU reporting countries, 25,535 sheep and 633 goats were tested. In sheep, 557 cases of scrapie were reported by 17 MS and XI: 480 classical scrapie (CS) by five MS (93 index cases [IC] with genotypes of susceptible groups in 97.6% of the cases), 77 atypical scrapie (AS) (76 IC) by 14 MS and XI. In the other non-EU reporting countries, Norway reported 16 cases of ovine AS. Ovine random genotyping was reported by eight MS and genotypes of susceptible groups accounted for 7.3%. In goats, 224 cases of scrapie were reported, all from EU MS: 216 CS (42 IC) by six MS, and 8 AS (8 IC) by four MS. In Cyprus, two cases of CS were reported in goats carrying the heterozygous DN146 allele. In total, 3202 cervids were tested for chronic wasting disease by 10 MS. One wild European moose tested positive in Finland. Norway tested 17,583 cervids with two European moose, one reindeer and one red deer positive. In total, 154 animals from four other species tested negative in Finland.

K E Y W O R D S

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

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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 2022 in the European Union (EU) Member States (MS), in the United Kingdom (in respect of Northern Ireland) (hereafter: 'XI') and in other eight non-EU reporting countries: Bosnia and Herzegovina, Iceland, Montenegro, North Macedonia, Norway, Serbia, Switzerland and Türkiye, as well as geno-typing data in sheep and goats. Albania and Kosovo* confirmed no TSE surveillance was conducted in 2022.

TSE monitoring data for cattle, sheep, goats, cervids and species other than domestic ruminants are reported by country according to Regulation (EC) 999/2001 (the TSE Regulation) and consist of testing and case data. Surveillance data were submitted through the EFSA TSE data reporting tool by 29 reporting countries. Seven MS submitted data directly as eXtensible Markup Language (XML) files by using their own system for the automatic upload of data into the EFSA Data Collection Framework (DCF). The electronically submitted data from the EFSA database were further processed, validated and extracted to draft the summary tables presented in the current EU summary report (EUSR).

As in 2021, the 2022 data of EU and XI have been compared with those of the previous years for the EU and the United Kingdom. That might introduce some bias, mainly in the 10-year trend analysis. In this report, the 2022 EU27 data (i.e. data from the current 27 EU MS, referred to in the report as 'EU27') have been summed up with those provided by XI. However, all tables present separately the EU27 totals and those including EU27 data plus XI. Totals obtained from the three European Free Trade Association (EFTA) countries (Iceland, Norway and Switzerland), the five non-EFTA IPA (Instrument for Pre-Accession Countries) (Bosnia and Herzegovina, Montenegro, North Macedonia, Serbia and Türkiye) were referred to as 'non-EU reporting countries' in the text and shortened in the Tables to 'other non-EU' for brevity of expression.

In total, 977,008 cattle were tested in 2022 in the EU27 and XI, a 4.3% reduction compared with the previous year. The 84% of all cattle tested in the EU27 and XI was reported to the group of risk animals (emergency slaughtered animals [ES], animals with clinical signs at *ante mortem* inspection [AM] and fallen stock [FS]), with FS being the largest contributor with 752,310 cattle tested in 2022 (91.7% of all cattle in the risk group). An additional 52,395 cattle were tested by the eight other non-EU reporting countries. Serbia (the main contributor with 16,913 cattle tested) and Türkiye (with 8108) reported mostly cattle from the healthy slaughtered (HS) target group while Switzerland (with 10,074) reported mostly cattle from animals in the risk group.

In the EU27 and XI, one atypical BSE case was confirmed in 2022 by France; it was a case of H-BSE in a 154-month-old beef bovine animal reported as FS. No other BSE cases were reported worldwide in 2022.

In total, 404,219 small ruminants were tested in 2022 in the EU27 and XI: 295,145 sheep (a 5.2% decrease compared to 2021) and 109,074 goats (a 7.9% decrease). In addition, 25,535 sheep were tested by five of the eight other non-EU reporting countries: Iceland, North Macedonia, Norway, Serbia and Türkiye, and 633 goats were tested by Iceland, Norway, Serbia and Türkiye.

In sheep, 557 scrapie cases were reported in the EU27 and XI in 2022, six more cases than in 2021. Among the five other non-EU reporting countries that tested sheep, Norway reported 16 cases of atypical scrapie (AS). In total, 480 ovine cases in the EU27 and XI were classical scrapie (CS) (86.2%), 77 were AS (13.8%). CS was reported only by five MS: Cyprus, Greece, Italy, Romania and Spain. AS was reported by 14 EU reporting countries (Austria, Croatia, Denmark, France, Germany, Hungary, Ireland, Italy, Poland, Portugal, Slovakia, Slovenia, Spain and Sweden) and the United Kingdom in respect of Northern Ireland. Among non-EU reporting countries, only Norway reported AS. Most of the ovine cases in the EU27 and XI (92.3%) were reported by five countries: Greece, Italy, Romania and Spain for CS and Portugal for AS, as it was the case in previous years.

In sheep, 169 (30.3%) of all cases in the EU27 and XI reported in 2022 were index cases (IC), with a much higher proportion in AS cases (98.7%) compared with CS cases (19.4%). In total, 97.3% of the CS cases in sheep reported in 2022 with known genotypes belonged to animals holding genotypes of the susceptible groups (NSP3, NSP3O, NSP4 or NSP5).

In 2022, the random genotyping of the national EU sheep populations was carried out by eight MS: Belgium, Cyprus (where genotyping is conducted systematically in the breeding sheep population), France, Germany, Italy, Latvia, the Netherlands and Poland. After excluding Cyprus, 7.3% of the randomly genotyped sheep with known genotypes still carried those of the susceptible groups, lower than the 7.9% in 2021. The exception is Italy, with 23.3% of sheep with the susceptible genotypes and the highest case load in 2022.

In goats, in total 224 scrapie cases were reported in the EU27 and XI: 216 CS (96.4%, with Cyprus accounting for 62% of these) and eight were AS cases (3.6%). Six MS (Bulgaria, Cyprus, Greece, Italy, Romania and Spain) reported CS, whereas four MS (France, Germany, Italy and Spain) reported AS. The four other non-EU reporting countries (Iceland, Norway, Serbia and Türkiye) that reported tested goats did not report any scrapie cases. In goats, 22.3% (50) of all cases reported in the EU27 and XI in 2022 was IC, higher than in 2021 (15.6%), with a higher proportion in AS (100%) than in CS (19.4%).

The genotyping of goat cases resulted in at least one polymorphism at codons 146 or 222: 184 cases (5 AS, 179 CS) have been reported by Cyprus (108), Greece (43), Italy (11) and Spain (22). Of these, 118 CS cases (64.1%) and 5 AS (100%) were genotyped at both codon 146 and 222. Two cases of CS reported by Cyprus were heterozygous goats at codon 146: DQ/ NQ and DZ/NZ.

With regard to long-term trends (cases per 10,000 tests), the analysis up to 2022 confirmed the 10-year statistically significant decrease in sheep (3% annually for both CS and AS) and no detectable trends in goats for either CS or AS.

^{*}This designation is without prejudice to positions on status and is in line with UNSCR 1244 and the ICJ Opinion on the Kosovo Declaration of Independence.

In 2022, 3202 cervids where tested by 10 MS (59.1% of them tested by Sweden and Romania). Only one case was reported by Finland involving a female European moose over 12 months old from the fallen/culled (FC) group. In 2022, the hunted/ slaughtered fit for human consumption (HSHC) target group was the most commonly tested group with 63.2% of all tested cervids. In 2022, Norway tested 17,583 animals, leading to the detection of four cases in wild animals (two moose, one reindeer and one red deer). Additionally, Iceland and Serbia reported testing of 79 and 281 cervids, respectively, which were all negative. In total, 154 animals of other species were TSE tested by Finland: 54 domestic cats, 54 American minks, 34 foxes and 12 raccoon dogs. None of them tested positive.

Two interactive communication tools on TSE – a storymap (click this link), providing general information on TSEs, and a dashboard (click this link), to search and visualise the surveillance data from EU MS and other reporting countries, have been updated with the data of the reporting year.

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1 | INTRODUCTION

1.1 | Background and terms of reference

According to Part I.A, Chapter B, 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 shall be indicated. For scrapie cases, the results of the primary and secondary molecular testing, referred to in Annex X, Chapter C, point 3.2(c), shall be reported, when appropriate.
- 7. In animals other than bovine, ovine and caprine animals, the number of samples and confirmed TSE cases per species.
- 8. The genotype, and, where possible, the breed, of each ovine and caprine animal found positive to TSE and sampled in accordance with Chapter A, Part II, point 8.

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.²

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. From 1 January 2016, the European Food Safety Authority shall analyse the information referred to in Part I and publish by the end of November a summary report on the trends and sources of TSE in the Union'.

1.2 | Surveillance of TSE in the European Union

1.2.1 | Legal basis

Animals suspected of a TSE should be examined in accordance with Article 12.2 of the TSE Regulation. The legal framework for the active 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.

Commission Decision 2009/719/EC³ allowed MS to apply a revised BSE monitoring programme. Commission Implementing Decision 2013/76/EU⁴ of 4 February 2013, amending Commission Decision 2009/719/EC, authorised 25 MS to decide to stop testing slaughtered bovine animals for human consumption. Following the EFSA scientific report on the evaluation of the revision of the BSE monitoring regime in Croatia (EFSA, 2016a) and the Commission Implementing

¹Regulation (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.

²Since 2018, TSE data are submitted by reporting countries directly to the European Food Safety Authority (EFSA) with different frequency and periodicity.

³Commission 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. ⁴Commission Implementing Decision of 4 February 2013 amending Decision 2009/719/EC authorising certain Member States to revise their annual BSE monitoring programmes.

Decision (EU) 2016/851⁵, Croatia was allowed to discontinue the testing of slaughtered bovine animals for human consumption, that is still required for Bulgaria and Romania.

With regard to the United Kingdom, the transition period agreed as part of the Agreement on the Withdrawal of the United Kingdom from the EU ended on 31 December 2020 and the United Kingdom is considered a third country. However, the Article 5(4) and Section 24 of Annex 2 of the Protocol on Ireland/Northern Ireland contemplates the EU requirements on data sampling are also applicable to Northern Ireland; so, for the purpose of this report, references to Member States are read as including the United Kingdom in respect of Northern Ireland.

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 36/2005⁶) also provides for mandatory discriminatory testing for BSE of TSE cases detected in small ruminants.

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 animals (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).

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 in EU, applied in 2022, 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 on TSE (EFSA, 2016b).

However, there are still some differences in the application of these general rules due to specific national provisions that provide some residual testing of HS or the testing of at-risk animals (AM, ES and FS) at younger age. The age limits (in months) of bovine animals tested for BSE surveillance applied in 2022 by Member States (MS), and United Kingdom (in respect of Northern Ireland) or the other non-EU reporting countries (Bosnia and Herzegovina, Iceland, Montenegro, North Macedonia, Norway, Serbia, Switzerland and Türkiye) are shown in Table 2.

TABLE 1	Criteria for BSE surveillance in bovine animals as applied in 2022 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 2	26 May 2016.

Surveillance target group	EU 25 + XI	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
Animals culled under BSE eradication measures (EM)		

^aDifferent criteria were applied in 2022 because Bulgaria and Romania were not in the list of the 25 MS and XI authorised to revise their BSE annual surveillance programmes.

TABLE 2 Age limits (in months) of bovine animals tested for BSE surveillance applied in 2022 by reporting country and surveillance target group.

	Surveilla	Surveillance target group						
Country	ES	АМ	FS	HS	SU	EM		
AT	>24	>24	>48 ^a	No testing ^b	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		
СҮ	>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		

⁵Commission Implementing Decision (EU) 2016/851 of 26 May 2016 amending the Annex to Decision 2009/719/EC as regards the authorisation for Croatia to revise its BSE annual monitoring programme. OJ L 141, 28.5.2016, p. 131–132.

⁶Commission 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.

TABLE 2 (Continued)

	Surveillanc	e target group				
Country	ES	AM	FS	HS	SU	EM
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 January 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
ІТ	>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	>48	>48	>48	No testing	No age limit	No age limit
МТ	>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
РТ	>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
XI ^c	>48	>48	>48	No testing	No age limit	No age limit
ВА	n/a	n/a	n/a	n/a	n/a	n/a
СН	>48	>48	>48	No testing	No age limit	>48
IS	>48	>48	>48	No testing	No age limit	No age limit
ME	>24	>24	>24	>30	No age limit	No age limit
МК	>24	>24	>24	>30	No age limit	No age limit
NO	>48	>48	>48	No testing	No age limit	No age limit
RS	>24	>24	>24	>72 for domestic animals> 30 for imported animals	No age limit	No age limit
TR	>30	> 30	> 30	>36	> 30	No age limit

Notes: The TSE Regulation does not apply to the eight non-EU reporting countries.

Abbreviations: ES, emergency slaughtered; AM, animals with clinical signs ante mortem; FS, fallen stock; HS, healthy slaughtered; SU, animals clinically suspected of being infected with BSE; EM, animals culled under BSE eradication measures; n/a, not available.

^alf surveillance target group is FS and animals are born in Romania, Bulgaria or Switzerland, or the United Kingdom (with the exception of Northern Ireland and if the movement to the European Union took place since 1.1.2021), then the age limit is > 24 months.

^bIf surveillance target group is HS and animals are born in Romania, Bulgaria, Switzerland or the United Kingdom (with the exception of Northern Ireland and if the movement to the European Union took place since 1.1.2021), then the age limit is > 30 months.

^cData from XI, i.e. United Kingdom (in respect of Northern Ireland), are available from 2021 onwards.

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 TSE⁷ is performed based on testing samples obtained from the following surveillance target groups: animals culled under TSE eradication measures (EM); animals not slaughtered for human consumption (NSHC); healthy animals slaughtered for human consumption (SHC) and animals clinically suspected of being infected by TSE (SU).

Target surveillance groups in small ruminants to be reported for surveillance for TSE in 2022 based on the infection status of flock/herd/holding, the case type detected and the control measures taken according to the TSE Regulation, have been summarised in Table 3.

The minimal sample sizes for NSHC and SHC are set out in Tables A and B of Annex III, Chapter A, Section II, point 3 and point 2(a) and (b), 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

⁷The 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.

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.

According to Commission Regulation (EU) 2021/1176⁸, point 4.6, Chapter B, Annex VII is replaced by the following: 'the restrictions set out in points 4.1 to 4.5 shall apply for a period of two years following the detection of the last TSE case, other than atypical scrapie, on the holdings where option 3 laid down in point 2.2.2(d) has been implemented'. This means, among other things, intensive surveillance no longer needs to be conducted for a period of 2 years on holdings where an atypical scrapie case has been confirmed.

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. Where the positive TSE case is an atypical scrapie case, the prion protein genotype for the codon 141 shall also be determined.

As described in the 2018 EUSR on TSE (EFSA, 2019), the Commission Regulation (EC) 2017/894⁹ amended the TSE Regulation with regard to representative genotyping activities in the ovine populations. The changes in the TSE Regulation no longer require genotyping a minimum sample of at least 600 animals per 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 a minimum sample of at least 1560 ovine animals once every 3 years; or at a frequency and with a sample size determined by the MS based on compliance with a set of criteria.

1.2.3.2 | Genotyping in goats

Point 8.2 Part II Chapter A of Annex III of Commission Regulation (EU) 2021/1176, amending the TSE Regulation, established that 'the prion protein genotype for the codons 146 and 222 shall be determined for each positive TSE case in goats. TSE cases found in goats of genotypes which encode serine (S) or aspartic acid (D) on at least one allele at codon 146 and/or lysine (K) on at least one allele at codon 222, shall immediately be reported to the Commission'.

⁸Commission regulation (EU) 2021/1176 of 16 July 2021 amending Annexes III, V, VII and IX to Regulation (EC) No 999/2001 of the European Parliament and of the Council as regards genotyping of positive TSE cases in goats, the determination of age in ovine and caprine animals, the measures applicable in a herd or flock with atypical scrapie and the conditions for imports of products of bovine, ovine and caprine origin.

⁹Commission 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.

TABLE 3 the TSE Regul	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	ation.
2	Target su	ie TSE Regulation.

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EUSR ON TRANSMISSIBLE SPONGIFORM ENCEPHALOPATHIES IN 2022

Minimum sample size for the TSE surveillance in small ruminants by reporting country in 2022. TABLE 4

	Sheep			Goats		
		Surveillanc	e target group		Surveillance	target group
Country	Population size ^a	SHC	NSHC	Population size ^a	SHC	NSHC
AT	100–750	0	1500	40–250	0	100% up to 500
BE	100–750	0	1500	40–250	0	100% up to 500
BG	>750	10,000	10,000	40–250	0	100% up to 500
СҮ	100–750	0	1500	40–250	0	100% up to 500
CZ	100–750	0	1500	<40	0	100% up to 100
DE	>750	10,000	10,000	40–250	0	100% up to 500
DK	40–100	0	100% up to 500	<40	0	100% up to 100
EE	40–100	0	100% up to 500	<40	0	100% up to 100
EL	>750	10,000	10,000	>750	10,000	10,000
ES	> 750	10,000	10,000	> 750	10,000	10,000
FI	40-100	0	100% up to 500	< 40	0	100% up to 100
FR	>750	10,000	10,000	>750	10,000	10,000
HR	100–750	0	1500	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	1500	<40	0	100% up to 100
LU	<40	0	100% up to 100	<40	0	100% up to 100
LV	40-100	0	100% up to 500	<40	0	100% up to 100
МТ	<40	0	100% up to 100	<40	0	100% up to 100
NL	100–750	0	1500	250–750	0	1500
PL	100–750	0	1500	40–250	0	100% up to 500
РТ	>750	10,000	10,000	250–750	0	1500
RO	>750	10,000	10,000	>750	10,000	10,000
SE	100–750	0	1500	<40	0	100% up to 100
SI	40–100	0	100% up to 500	<40	0	100% up to 100
SK	100–750	0	1500	<40	0	100% up to 100
XI	>750	10,000	10,000	40–250	0	100% up to 500
BA	>750			40–250		
СН	-			-		
IS	100–750			<40		
ME	100–750			<40		
мк	100–750			40–250		
NO	>750			40-250		
RS	>750	1000 ^b	1000 ^b	40-250	1000 ^c	1000 ^c
TR	>750			>750		

Abbreviations: TSE, transmissible spongiform encephalopathy; NSHC, animals not slaughtered for human consumption; SHC, animals slaughtered for human consumption. The TSE regulation does not apply to the eight non-MS countries. ^aThousand heads.

^{b,c}The 1000 animals targeted are split between SHC and NSHC.

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

Live sheep population in 2022 (or latest available) extracted from: https://ec.europa.eu/eurostat/databrowser/view/APRO_MT_LSSHEEP/default/table?lang=en Live goat population in 2022 (or latest available) extracted from: https://ec.europa.eu/eurostat/databrowser/T_LSGOAT/default/table?lang=en

TSE surveillance in cervids and other species 1.2.4

Since 2021 MS and non-EU reporting countries may carry out monitoring for CWD in cervids only on a voluntary basis.

8314732, 2023, 11, Downloaded from https://efsa.onlinelibrary.wiley.com/doi/10.2903/j.efsa.2023.8384 by CochraneItalia, Wiley Online Library on [30/11/2023]. See the Terms and Conditions (https://onlinelibrary.wiley.con/terms-and-conditions) on Wiley Online Library for rules of use; OA articles are governed by the applicable Creative Commons License

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). The testing protocols were updated in 2020 and came into force in 2021; a reference to the updated protocols is included in the Guidance for reporting 2021 surveillance data on TSE (EFSA et al., 2021).

2 | DATA AND METHODS

2.1 | Origin of the data

Raw data are electronically submitted by EU MS and non-EU reporting countries. 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 on TSE were submitted to EFSA as required by the TSE Regulation. The EFSA data reporting 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 (DWH). The tool has been used for the first time during the 2018 TSE data collection period. Seven reporting countries (CZ, ES, FI, FR, IT, NL and SE) transmitted data directly as eXtensible Markup Language (XML) files in 2022 by using their own system for the XML file generation and the upload of data into the DCF, whereas the rest of the reporting countries transmitted XML files to the DCF by the EFSA TSE data reporting tool. All data were then submitted to the EFSA DWH and confirmed by the reporting countries. The electronically submitted data were extracted from the EFSA DWH and further processed and validated by EFSA to summarise the information and to draft the summary tables presented in the current EUSR. The validation dashboard, available to all reporting countries to visualise the data since 2018, has been updated for 2022 data visualisation.

Finally, information on the population of bovine animals in 2022 were obtained from Eurostat annual data¹⁰ (Bovine animals, 2 years or over), while information on the population of small ruminants in 2022 as presented in Table 4 were obtained from the 2022 or latest available Eurostat annual data.¹¹ The number of BSE cases worldwide (Table 12) was 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 World Animal Health Information System (WOAH-WAHIS; https://wahis.woah.org/#/home) a final check regarding the number of BSE cases outside Europe was made by requesting confirmation of WAHIS data to WOAH staff.

During validation of the data, the following additional information was asked to the reporting countries:

- the number of suspected cases placed under official movement restrictions in accordance with Article 12(1), per animal species (according to (i) Annex III, Chapter B, Section 1.A, point 1 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) according to (ii) Annex III, Chapter B, Section 1.A, point 3 of the TSE Regulation; and
- the number of ovine and caprine flocks tested within each subpopulation referred to in Annex III, Chapter A, Part II, points 2, 3, 5 and 6.

The results of this questionnaire are summarised in Appendix D.

In accordance with the Agreement on the Withdrawal of the United Kingdom from the EU, and in particular with the Protocol on Ireland/Northern Ireland, the EU requirements on data sampling are also applicable to Northern Ireland. Therefore, pursuant to Article 5(4) and Section 24 of Annex 2 of the Protocol on Ireland/Northern Ireland, which is an integral part of the Agreement on the withdrawal of the United Kingdom of Great Britain and Northern Ireland from the European Union and the European Atomic Energy Community, for the purpose of this report, references to Member States are read as including the United Kingdom in respect of Northern Ireland.

The data in this report refer only to the samples collected and cases confirmed between 1 January 2022 and 31 December 2022 in the EU (27 Member States and United Kingdom in respect of Northern Ireland, referred to in the report as 'EU27 + XI') and other eight additional non-EU reporting countries: Bosnia and Herzegovina, Iceland, Montenegro, North Macedonia, Norway, Serbia, Switzerland and Türkiye. Upon request, Albania and Kosovo* informed that they did not conduct TSE surveillance data in 2022.

EFSA validated the 2022 data by checking for inconsistencies in the electronically extracted data, and by comparing the reported data with previous years. Members of the TSE subgroup of the Scientific Network for Zoonoses Monitoring Data in the reporting countries were consulted during this validation. The data validation started on 1 May 2023 and was finalised on 20 July 2023. The results and tables presented in the current EUSR are based on the data retrieved from the EFSA

¹⁰Live bovine animals 2 years or over in 2022 (or latest available) extracted from: https://ec.europa.eu/eurostat/databrowser/view/APRO_MT_LSSHEEP/default/table?lang=en ¹¹Live sheep population in 2022 (or latest available) extracted from: https://ec.europa.eu/eurostat/databrowser/view/APRO_MT_LSSHEEP/default/table?lang=en Live goat population in 2022 (or latest available) extracted from: https://ec.europa.eu/eurostat/databrowser/view/APRO_MT_LSGHEEP/default/table?lang=en Live goat *This designation is without prejudice to positions on status and is in line with UNSCR 1244 and the ICJ Opinion on the Kosovo Declaration of Independence.

Scientific Data Warehouse on 20 July 2023. An additional consultation with reporting countries was conducted between 15 September 2023 and 5 October 2023. Data submitted from 2018 onwards can be corrected in the EFSA DWH. However, if data were corrected by the reporting countries in the report but not updated in the EFSA Scientific Data Warehouse, the corrections will only be mentioned by means of footnotes in the current or future EUSR.

Data between 2001 and 2022 with focus on the last 5 years in cattle and sheep are presented in tables and figures. As certain MS and non-EU reporting countries 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 reporting countries for 2022. In addition, subsequent submissions of updated/ amended historical data by reporting countries may have resulted in differences in the figures included in this report when compared with the same data presented in previous EUSR.

2.2 Presentation of the data

The current report should be considered the EU summary report for 2022 in compliance with Section II, Chapter B, Annex III of the TSE Regulation.

The reporting countries in this report are the 27 EU MS or EU27, the United Kingdom (in respect of Northern Ireland), three EFTA members (Iceland, Norway and Switzerland) and five non-EFTA IPA (Instrument for Pre-Accession Countries) countries (Bosnia and Herzegovina, Montenegro, North Macedonia, Serbia and Türkiye). 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 1059/2003¹² (see section Country codes).

Since 1 January 2021, complete United Kingdom data are no longer submitted to EFSA. Instead, data from Northern Ireland (XI) (United Kingdom in respect of Northern Ireland) are submitted and included in this report. In the tables of the report, EU27 data are shown individually and summed to those provided by the XI (EU27 + XI). Totals obtained from the three EFTA countries and the five non-EFTA IPA are referred as to 'Other non-EU'. Tables 12–18, 23–30 include historical data in which full data from United Kingdom are displayed until 2020 and XI for 2021–2022.

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 group 'risk animals' is used here to indicate those animals in which the probability of detecting 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.

Two interactive communication tools on TSE – a storymap (click this link), providing general information on TSEs, and a dashboard (click this link), to search and visualise the surveillance data from EU Member States and other reporting countries, have been updated with the data of the reporting year.

2.3 Methods

2.3.1 | Descriptive methods

To describe the results of the TSE surveillance programme in the EU in 2022, figures and tables have been produced along with a short narrative text to describe the main findings. The report is split into four sections: bovine animals (cattle), small ruminants (sheep and goats), cervids and species other than bovine, ovine and caprine animals and cervids. Both EU aggregated data and data at the national level are presented. When it was considered relevant, multiyear and historical data are shown. Surveillance data covered the period 2001–2022 for bovine animals, and the period 2002–2022 for small ruminants.

Despite the new provisions for the data from United Kingdom, the 2022 EU and XI data have been compared with data for the EU and the United Kingdom until 2021; hence, some caution must be applied when interpreting the results.

For bovine animals, summary statistics were obtained based on the total number of tests performed in 2022 by reporting country and surveillance target group. In addition, historical data on confirmed cases between 2018 and 2022 (a 5-year period) are presented in detail whereas those on the 2001–2017 period have been summed up. Over this period data of reporting countries are comparable: as of 2017 in the EU as a whole, the surveillance system has been harmonised with active surveillance limiting monitoring to at-risk animals older than 48 months; 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 2022, EFSA asked for additional information from the individual concerned reporting countries by a small questionnaire.

¹²Regulation (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.

For small ruminants, summary statistics are presented in this report, and when 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 (2013–2022) 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 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, e.g. 350, the MS was categorised as compliant as the total subpopulation of NSHC in the country is not known.

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 (IC), 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 sheep genotyping while allele nomenclature was used to summarise and describe the data on goats genotyping.

To describe and plot the data, some assumptions were made to report the results of bovine animals and small ruminants (sheep and goats):

- To present the temporal change in evolution of BSE cases (C-BSE, L-BSE and H-BSE) in tables or graphs, 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 change in evolution of the total number of scrapie IC, it was assumed that all IC were confirmed in noninfected flocks/herds. If a case was reported as non-index or unknown index status, 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 2022 by target group, reporting country, species and management system (wild and semi-domesticated/farmed).
- For all reporting countries, the number of cases in cervids in 2022.

Number of tested animals in species other than cattle, sheep, goats and cervids tested for TSE in reporting countries in 2022 are presented in tabular format by species and reporting country.

2.3.2 | Data analysis 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 2013–2022) has 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.

TSE data of small ruminants from the last 10 years (period 2013–2022) have been used to check if any significant temporal trend was detectable. As per BSE, a Poisson regression model has been fitted for each case type (CS and AS) and for each species (ovine and caprine) 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.

For both species, 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: An RR > 1 indicates an average annual increase in the number of cases per million whereas an RR << 1 indicates an average annual decrease.

Over the same 10-year period, and considering cases from all reporting countries, the mean age of the AS cases has been compared with that of CS cases in sheep and goats by applying a two-sample *t*-test with unequal variances.

A *p*-value \leq 0.05 was considered statistically significant for all the above-described statistical analyses.

3 | ASSESSMENT

3.1 | BSE surveillance in bovine animals

Since 2001 ~ 121.9 million bovine animals have been tested for BSE in the EU, including the United Kingdom until 2020 and XI instead since 2021. In 2022, there was a 4.3% reduction in the number of tested bovine animals in the EU27 and XI, from 1,021,252 in 2021 to 977,008 in 2022. This reduction is due to a sharp decrease in the total number of cattle tested by 44,244 animals, mainly by Bulgaria, France, Germany and Italy, partially compensated by an increase in the number of cattle tested by 15,386 in Greece and Romania. Romania and Bulgaria are the main contributors to the HS testing group with 139,264 (89.5%) of all HS tested cattle in the EU27 and XI.

The other eight non-EU reporting countries (Bosnia and Herzegovina, Iceland, Montenegro, North Macedonia, Norway, Serbia, Switzerland and Türkiye) tested 52,395 cattle in 2022. Serbia was the main contributor with 16,913 cattle tested (showing a decrease of 13.6% compared to 2021), followed by Switzerland that tested 10,074 cattle. All the non-EFTA IPA countries reported mostly cattle tested in the HS target group. Switzerland and Norway tested mainly FS and ES.

The number of animals tested in the risk group (ES + AM + FS) decreased from 870,328 in 2021 in EU and XI to 820,561 in 2022 (-5.7%). This reduction is mainly due to the decrease in testing in the risk group in Germany and France, with a reduction of 7% and 9.8%, respectively. Similarly, to the previous year, cattle in the risk group accounted for 84% of all tested cattle in the EU27 and XI and cattle tested in the FS target group accounted for 91.7% of all risk cattle tested. The number of cattle tested for BSE per reporting country for each target group in 2022 is shown in Table 5.

	Surveillance target group									
	Risk anima	als			Other target groups					
Country	FS	АМ	ES	Subtotal risk animals	нѕ	ЕМ	SU	Subtotal other target groups	Total	
AT	15,075	26	3360	18,461	33		8	41	18,502	
BE	22,697		1126	23,823	5		1	6	23,829	
BG	1145	2	238	1385	27,601		1	27,602	28,987	
СҮ	1847		111	1958	80			80	2038	
cz	19,361	1	4583	23,945	30			30	23,975	
DE	152,711		11,949	164,660	136		648	784	165,444	
DK	22,868		1607	24,475	1			1	24,476	
EE	2862	45	126	3033					3033	
EL	1959			1959	13,546		20	13,566	15,525	
ES	57,560	35	1540	59,135	379			379	59,514	
FI	9815		2	9817					9817	
FR	174,790		1867	176,657	1591		1	1592	178,249	
HR	4734		66	4800	195		3	198	4998	
HU	11,600	249	34	11,883	82		10	92	11,975	
IE	59,510	227		59,737	115		5	120	59,857	

TABLE 5 Number of bovine animals tested for BSE by reporting country and surveillance target group in 2022 in the EU and other reporting countries.

TABLE 5 (Continued)

	Surveillance target group									
	Risk anima	als			Other targ	Other target groups				
Country	FS	АМ	ES	Subtotal risk animals	нѕ	EM SU	Subtotal other target groups	Total		
ІТ	31,017	154	10,795	41,966	182		182	42,148		
LT	3969	20	9	3998				3998		
LU	2668			2668		3	3	2671		
LV	3287	68	77	3432		6	6	3438		
мт	114		121	235				235		
NL	49,793		7588	57,381	4		4	57,385		
PL	37,964	1060	7183	46,207	2	6	8	46,215		
РТ	15,101	741	1790	17,632	1	7	8	17,640		
RO	3966	5800	4365	14,131	111,663	53	111,716	125,847		
SE	7420	26	200	7646		8	8	7654		
SI	5795	45	538	6378	11	10	21	6399		
SK	8610		3	8613				8613		
Total EU27	728,238	8499	59,278	796,015	155,657	790	156,447	952,462		
XI ^a	24,072	377	97	24,546				24,546		
Total EU27 + XI	752,310	8876	59,375	820,561	155,657	790	156,447	977,008		
BA					3653		3653	3653		
СН	5594		4458	10,052		22	22	10,074		
IS	58			58	161		161	219		
ME	1			1	4714		4714	4715		
МК					1726		1726	1726		
NO	1696	31	5194	6921	66		66	6987		
RS	3737		37	3774	13,131	8	13,139	16,913		
TR	8			8	8100		8100	8108		
Total other non-EU	11,094	31	9689	20,814	31,551	30	31,581	52,395		
Total	763,404	8907	69,064	841,375	187,208	820	188,028	1,029,403		

Abbreviations: 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.

^aData from XI, United Kingdom (in respect of Northern Ireland) are available from 2021 onwards.

The distribution of the number of bovine animals tested for BSE by age group, surveillance target group and reporting country in 2022 can be found in the following link https://doi.org/10.5281/zenodo.5602748 distributed as follows:

TABLE 6 Number of bovine animals tested by age group in the EU27 and XI and non-EU reporting countries in 2022.

TABLE 7 Number of bovine animals in the risk group (animals with clinical signs at *ante mortem*, emergency slaughtered and fallen stock), by age group, tested in EU27 and XI and non-EU reporting countries in 2022.

TABLE 8 Number of tested healthy slaughtered bovine animals by age group in EU27 and XI and non-EU reporting countries in 2022.

TABLE 9 Number of BSE suspected bovine animals, by age group, tested in EU27 and XI MS and non-EU-reporting countries in 2022.

TABLE 10 Number of bovine animals culled under BSE eradication measures, by age group, tested in EU27 and XI and non-EU reporting countries in 2022. The table is empty because no animals in this category were tested for the year 2022.

In the EU27 and XI, one BSE case was reported in 2022. The case, an H-type, was found in the FS testing group in France in a beef cow older than 12 years of age. Table 11 reports the main clinical and epidemiological data of the case. The reported case showed clinical signs before death. In 2022, no additional cases of BSE were reported in the rest of the world.

In general, considering the low annual incidence of H-type and L-type BSE cases, this single case found was in line with the number of cases reported in the last few years. Figure 1 shows the proportion of cases per million tests from 2017 to 2022.

Based on 126 atypical BSE cases with known age since 2001, the average age at detection was 12.1 years (range: 5.5–18.5 years). The FS target group accounts, as mentioned above, for most of the tested animals and hence the cases.

TABLE 11 Clinical and epidemiological description of the BSE cases detected in 2022.

CountryFR - atypicalSurveillance target groupFallen stockCase typeH-BSEMonth and year of birth4/2010Age at detection (in months)154BARB statusNoClinical signsDifficulty to get up and stand up, always turning 'in circles' in the same direction, walking 'crab-wise'. Clinical signsCattle typeBeefBreedBlonde d'AquitaineWas the case confirmed at herd/holding where the animal was born?YesLocation (NUTS3) of natal herd or herd where case foundTarn et Garonne, FRANCEHerd size8Herd typeBeefFeeding system during first year of lifeNoFeed cohorts? Tested? If Yes: Results (number tested; number positives)NoOffspring? Tested? If Yes: Results (number tested; number positives)NoSire? Tested? (Yes/No). If Yes: Results (positive? Negative?)NoDam? Tested (Yes/No). If Yes: Results (positive? Negative?)No		
Case typeH-BSEMonth and year of birth4/2010Age at detection (in months)154BARB statusNoClinical signsDifficulty to get up and stand up, always turning 'in circles' in the same direction, walking 'crab-wise'. Clinical signs started 1 month before deathCattle typeBeefBreedBlonde d'AquitaineWas the case confirmed at herd/holding where the animal was born?YesLocation (NUTS3) of natal herd or herd where case foundTarn et Garonne, FRANCEHerd size8Herd typeBeefFeeding system during first year of lifeBeefFeed cohorts? Tested? If Yes: Results (number tested; number positives)NoBirth cohorts? Tested? If Yes: Results (number tested; number positives)NoSire? Tested? If Yes: Results (number tested; number Positives)8 offspring / not testedSire? Tested? If Yes: Results (number tested; number Positives)No	Country	FR – atypical
Month and year of birth4/2010Age at detection (in months)154BARB statusNoClinical signsDifficulty to get up and stand up, always turning 'in circles' in the same direction, walking 'crab-wise'. Clinical signs started 1 month before deathCattle typeBeefBreedBlonde d'AquitaineWas the case confirmed at herd/holding where the animal was born?YesLocation (NUT53) of natal herd or herd where case foundTarn et Garonne, FRANCEHerd size8Herd typeBeefFeeding system during first year of lifeBeefFeed cohorts? Tested? If Yes: Results (number tested; number positives)NoBirth cohorts? Tested? If Yes: Results (number tested; number positives)NoOffspring? Tested? If Yes: Results (positive? Negative?)No	Surveillance target group	Fallen stock
Age at detection (in months)154BARB statusNoClinical signsDifficulty to get up and stand up, always turning 'in circles' in the same direction, walking 'crab-wise'. Clinical signs started 1 month before deathCattle typeBeefBreedBlonde d 'AquitaineWas the case confirmed at herd/holding where the animal was born?YesLocation (NUTS3) of natal herd or herd where case foundTarn et Garonne, FRANCEHerd size8Herd typeBeefFeeding system during first year of lifeFeed cohorts? Tested? If Yes: Results (number tested; number positives)NoBirth cohorts? Tested? If Yes: Results (number tested; number positives)NoOffspring? Tested? If Yes: Results (number tested; number Positives)NoSire? Tested? (Yes/No). If Yes: Results (positive? Negative?)No	Case type	H-BSE
BARB statusNoClinical signsDifficulty to get up and stand up, always turning 'in circles' in the same direction, walking 'crab-wise'. Clinical signs started 1 month before deathCattle typeBeefBreedBlonde d 'AquitaineWas the case confirmed at herd/holding where the animal was born?YesLocation (NUTS3) of natal herd or herd where case foundTarn et Garonne, FRANCEHerd size8Herd typeBeefFeeding system during first year of lifeFeed cohorts? Tested? If Yes: Results (number tested; number positives)NoBirth cohorts? Tested? If Yes: Results (number tested; number positives)NoOffspring? Tested? If Yes: Results (number tested; number Positives)8 offspring / not testedSire? Tested? (Yes/No). If Yes: Results (positive?) Negative?)No	Month and year of birth	4/2010
Clinical signsDifficulty to get up and stand up, always turning 'in circles' in the same direction, walking 'crab-wise'. Clinical signs started 1 month before deathCattle typeBeefBreedBlonde d 'AquitaineWas the case confirmed at herd/holding where the animal was born?YesLocation (NUTS3) of natal herd or herd where case foundTarn et Garonne, FRANCEHerd size8Herd typeBeefFeeding system during first year of lifeNoFeed cohorts? Tested? If Yes: Results (number tested; number positives)NoBirth cohorts? Tested? If Yes: Results (number tested; number positives)NoSire? Tested? If Yes: Results (number tested; number Positives)NoSire? Tested? If Yes: Results (positive? Negative?)No	Age at detection (in months)	154
in the same direction, walking 'crab-wise'. Clinical signs started 1 month before deathCattle typeBeefBreedBlonde d 'AquitaineWas the case confirmed at herd/holding where the animal was born?YesLocation (NUTS3) of natal herd or herd where case foundTarn et Garonne, FRANCEHerd size8Herd typeBeefFeeding system during first year of lifeNoFeed cohorts? Tested? If Yes: Results (number tested; number positives)NoBirth cohorts? Tested? If Yes: Results (number tested; number positives)NoOffspring? Tested? If Yes: Results (number tested; number Positives)NoSire? Tested? (Yes/No). If Yes: Results (positive? Negative?)No	BARB status	No
BreedBlonde d'AquitaineWas the case confirmed at herd/holding where the animal was born?YesLocation (NUTS3) of natal herd or herd where case foundTarn et Garonne, FRANCEHerd size8Herd typeBeefFeeding system during first year of lifeEefFeed cohorts? Tested? If Yes: Results (number tested; number positives)NoBirth cohorts? Tested? If Yes: Results (number tested; number positives)NoOffspring? Tested? If Yes: Results (number tested; number positives)8 offspring / not testedSire? Tested? If Yes: Results (positive? Negative?)No	Clinical signs	in the same direction, walking 'crab-wise'. Clinical signs
Was the case confirmed at herd/holding where the animal was born?YesLocation (NUTS3) of natal herd or herd where case foundTarn et Garonne, FRANCEHerd size8Herd typeBeefFeeding system during first year of lifeNoFeed cohorts? Tested? If Yes: Results (number tested; number positives)NoBirth cohorts? Tested? If Yes: Results (number tested; number positives)NoOffspring? Tested? If Yes: Results (number tested; number Positives)NoSire? Tested? If Yes: Results (positive? Negative?)No	Cattle type	Beef
Location (NUTS3) of natal herd or herd where case foundTarn et Garonne, FRANCEHerd size8Herd typeBeefFeeding system during first year of lifeNoFeed cohorts? Tested? If Yes: Results (number tested; number positives)NoBirth cohorts? Tested? If Yes: Results (number tested; number positives)NoOffspring? Tested? If Yes: Results (number tested; number Positives)8 offspring / not testedSire? Tested? If Yes: Results (positive? Negative?)No	Breed	Blonde d 'Aquitaine
Herd size8Herd typeBeefFeeding system during first year of lifeNoFeed cohorts? Tested? If Yes: Results (number tested; number positives)NoBirth cohorts? Tested? If Yes: Results (number tested; number positives)NoOffspring? Tested? If Yes: Results (number tested; number Positives)8 offspring / not testedSire? Tested? (Yes/No). If Yes: Results (positive? Negative?)No	Was the case confirmed at herd/holding where the animal was born?	Yes
Herd type Beef Feeding system during first year of life No Feed cohorts? Tested? If Yes: Results (number tested; number positives) No Birth cohorts? Tested? If Yes: Results (number tested; number positives) No Offspring? Tested? If Yes: Results (number tested; number Positives) 8 offspring / not tested Sire? Tested? If Yes: Results (positive? Negative?) No	Location (NUTS3) of natal herd or herd where case found	Tarn et Garonne, FRANCE
Feeding system during first year of life Feed cohorts? Tested? If Yes: Results (number tested; number positives) No Birth cohorts? Tested? If Yes: Results (number tested; number positives) No Offspring? Tested? If Yes: Results (number tested; number Positives) 8 offspring / not tested Sire? Tested? (Yes/No). If Yes: Results (positive? Negative?) No	Herd size	8
Feed cohorts? Tested? If Yes: Results (number tested; number positives)NoBirth cohorts? Tested? If Yes: Results (number tested; number positives)NoOffspring? Tested? If Yes: Results (number tested; number Positives)8 offspring / not testedSire? Tested? (Yes/No). If Yes: Results (positive? Negative?)No	Herd type	Beef
Birth cohorts? Tested? If Yes: Results (number tested; number positives)NoOffspring? Tested? If Yes: Results (number tested; number Positives)8 offspring / not testedSire? Tested? (Yes/No). If Yes: Results (positive? Negative?)No	Feeding system during first year of life	
Offspring? Tested? If Yes: Results (number tested; number Positives)8 offspring / not testedSire? Tested? (Yes/No). If Yes: Results (positive? Negative?)No	Feed cohorts? Tested? If Yes: Results (number tested; number positives)	No
Sire? Tested? (Yes/No). If Yes: Results (positive? Negative?) No	Birth cohorts? Tested? If Yes: Results (number tested; number positives)	No
	Offspring? Tested? If Yes: Results (number tested; number Positives)	8 offspring / not tested
Dam? Tested (Yes/No). If Yes: Results (positive? Negative?) No	Sire? Tested? (Yes/No). If Yes: Results (positive? Negative?)	No
	Dam? Tested (Yes/No). If Yes: Results (positive? Negative?)	No

The number of BSE cases by reporting country, type and year (up to 2022, with a focus on the last 5 years) is shown in Tables 12–14 for total cases, classical BSE and atypical BSE, respectively.

Time-series analysis carried out over the last 10-year period (period 2013–2022) shows a significant decreasing trend in the occurrence of C-BSE (annual RR=0.64, i.e. an annual decrease of 36% in the proportion of cases per tested animals; p=0.01), whereas no significant trend for the two atypical BSE forms was found (H-BSE: RR=0.99 p=0.95; L-BSE: RR =1.05 p=0.52).

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 2001–2022 are shown in Appendix B.

 TABLE 12
 Total number of reported BSE cases (classical-BSE + atypical H-BSE + atypical L-BSE) in reporting countries and worldwide by year (period 1991–2022) and country.

	Year						
Country	Up to 2016	2018	2019	2020	2021	2022	Total
AT	8						8
BE	133						133
CZ	30						30
DE ^a	421				1		422
DK ^a	16						16
EL	1						1
ES	817		2	1	2		822
FI	1						1
FR ^a	1006	3	4	2	3	1	1019
IE ^a	1661			1			1662
IT ^a	147						147
LU	3						3
NL	88						88
PL	74		1				75
PT ^a	1086						1086
RO	2						2

TABLE 12 (Continued)

	Year						
Country	Up to 2016	2018	2019	2020	2021	2022	Total
SE ^b	1						1
Sl ^a	9						9
SK	27						27
Total EU27	5531	3	7	4	6	1	5552
BRA	2		1		2		5
CAN ^a	20				1		21
CH ^a	465			1			466
ISR	1						1
JPN	36						36
LI	2						2
NO	1						1
USA ^a	5	1					6
United Kingdom ^c	184,594	1					184,595
XU ^c					1		1
Total other non-EU	185,126	2	1	1	4	0	185,134
Total	190,657	5	8	5	10	1	190,686

Notes: Grey-shaded cells indicate the data availability due to the transition from UK to XI+XU due to Brexit. EU countries without BSE cases (Bulgaria, Cyprus, Estonia, Croatia, Hungary, Lithuania, Latvia and Malta) are not included in the table.

Abbreviations: BSE, bovine spongiform encephalopathy; H-BSE, H-type BSE; L-BSE, L-type BSE; BRA, Brazil; CAN, Canada; ISR, Israel; JPN, Japan; LI, Liechtenstein; USA, The United States of America. Each cell reports the total number of BSE cases (C-BSE + H-BSE + L-BSE).

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

^bGavier-Widén et al. (2008).

^cUnited Kingdom was member of European Union until 1 February 2020. XI (United Kingdom in respect of Northern Ireland) and XU (the rest of the United Kingdom) are presented from there on. 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 (https://www.oie.int/wahis). Source: data regarding non-EU cases from 2003 collected from https://wahis.woah.org/#/event-management.

TABLE 13 Number of reported classical BSE cases in the EU and non-EU reporting countries by year and country.

	Year						
Country code	Up to 2017	2018	2019	2020	2021	2022	Total
AT	5						5
BE	133						133
CZ	29						29
DE	416						416
DK	15						15
EL	1						1
ES	798						798
FI	1						1
FR	969						969
IE	1656						1656
ІТ	142						142
LU	3						3
NL	84						84
PL	60						60
РТ	1079						1079
SI	8						8
SK	27						27

(Continues)

TABLE 13 (Continued)

	Year						
Country code	Up to 2017	2018	2019	2020	2021	2022	Total
Total EU27	5426	0	0	0	0	0	5426
СН	464						464
United Kingdom ^a	184,578	1					184,579
XU ^a					1		1
Total other non-EU	185,042	1	0	0	1	0	185,044
Total	190,468	1	0	0	1	0	190,470

Notes: 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 data availability due to the transition from UK to XI+XU due to Brexit.

Abbreviations: BSE, bovine spongiform encephalopathy.

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

^aUnited Kingdom was member of European Union until 2020. XI (United Kingdom in respect of Northern Ireland) and XU (the rest of the United Kingdom) are presented from there on.

TABLE 14 Number of reported BSE atypical cases in EU and non-EU reporting countries by year (period 2001–2022), type and country.

	Year													
	Up to 20)17	2018	;	2019)	2020)	2021		2022	2	Total	
Country code	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	2	3								1			2	4
DK	0	1											0	1
ES	9	10			2		1		1	1			13	11
FR	19	18	1	2	4		1	1	1	2	1		27	23
IE	4	1					1						5	1
ІТ	0	5											0	5
NL	1	3											1	3
PL ^a	2	12				1							2	13
РТ	7	0											7	0
RO	0	2											0	2
SE	1	0											1	0
SI	1	0											1	0
Total EU27	48	57	1	2	6	1	3	1	2	4	1	0	61	65
СН	1	0						1					1	1
NO	1	0											1	0
United Kingdom ^b	7	9											7	9
Total other non-EU	9	9	0	0	0	0	0	1	0	0	0	0	9	10
Total	57	66	1	2	6	1	3	2	2	4	1	0	70	75

Notes: Each cell reports the total number of H-BSE and L-BSE cases. EU countries without atypical cases are not included in the table.

^aIn 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.

^bUnited Kingdom was member of European Union until 2020. XI (United Kingdom in respect of Northern Ireland) and XU (the rest of the United Kingdom) are presented from there on.

The number of historical reported BSE cases can be found in the following link https://doi.org/10.5281/zenodo.4252321, as follows:

TABLE 15	Number of BSE cases per country and year until 2000 (included) in the EU and non-EU reporting countries.
TABLE 16	Number of classical BSE cases per country and year from 2001 in the EU and non-EU reporting countries.
TABLE 17	Number of atypical H-BSE cases per country and year from 2001 in the EU and non-EU reporting countries.

 TABLE 18
 Number of atypical L-BSE cases per country and year from 2001 in the EU and non-EU reporting countries.

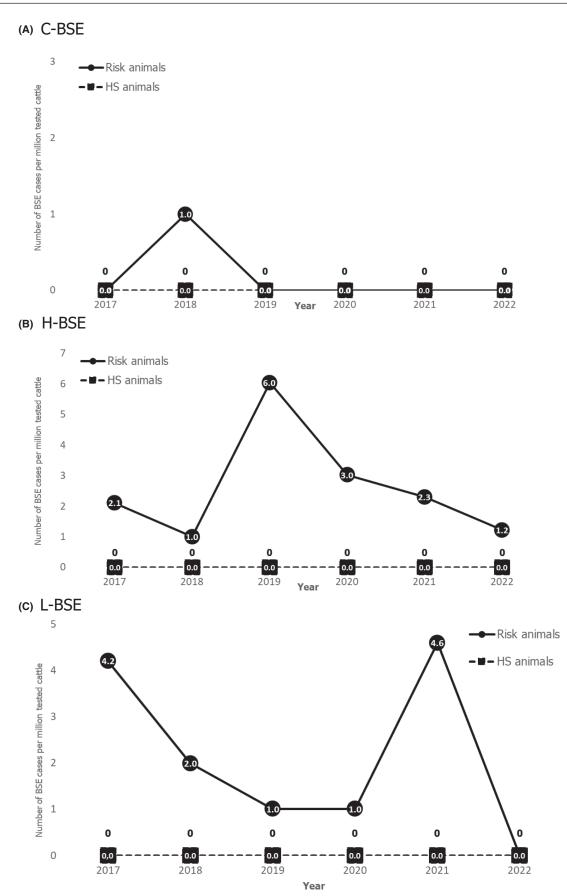


FIGURE 1 Cases per million tested bovine animals by surveillance target group and case type for the period 2017–2022 in the EU and the United Kingdom (until 2020) and in the EU and XI in 2021–2022. BSE, bovine spongiform encephalopathy; C-BSE, classical BSE; H-BSE, H-type BSE; L-BSE, L-type BSE. Black numbers in white background: number of cases.

3.2 | TSE surveillance in small ruminants

Since 2002, more than 10.8 million small ruminants have been tested as part of the official EU TSE surveillance in the EU, including the United Kingdom until 2020 and XI instead since 2021. In 2022, 404,219 small ruminants were tested by the EU27 and XI: 295,145 sheep (73%) and 109,074 goats (27%), which represents an overall 5.9% decrease (25,442) in the number of tested small ruminants, compared to 2021.

In five of the eight non-EU reporting countries (Iceland, North Macedonia, Norway, Serbia, Türkiye), a total of 26,168 small ruminants were tested: 25,535 sheep (97.6%) and 633 goats (2.4%), a decrease of 2021 (7.2%) compared with 2021, mostly due to a 30.7% decrease in testing by Iceland compared to the previous year. Bosnia and Herzegovina, Montenegro and Switzerland did not report data on small ruminants.

In sheep, the decrease in the animals tested in the EU27 and XI was 5.2% (295,145 tested in 2022 compared with 311,174 in 2021), due to the decrease of both the testing in TSE-infected flocks of 27.4% in 2022 (from 20,793 in 2021 to 15,102 in 2022), and a 3.6% in the non-TSE-infected flocks (from 290,381 in 2021 to 280,013 in 2022).

In goats, there was a 7.9% decrease in the animals tested in the EU27 and XI (109,074 in 2022 compared with 118,457 in 2021). The testing showed a 32.6% decrease in TSE-infected herds (from 6145 in 2021 to 4141 in 2022). and a 6.6% decrease in non-TSE-infected herds (from 112,312 in 2021 to 104,933 in 2022).

The numbers of sheep and goats tested for TSE by reporting country, surveillance target group and flock/herd status in 2022 are summarised in Tables 19 and 20, 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) and the criteria described in Section 2.3.1, 24 countries in the group EU27 + XI fulfilled the requirements for sheep testing. In goat surveillance, 24 countries in the group EU27 and XI fulfilled the requirements for goat testing.

In 2022, the pattern of ovine testing by country and flock status was different from that of 2021. For each sheep tested in a TSE-infected flock in the EU27 and XI, there were about 18.5 sheep tested in non-TSE-infected flocks, higher than in 2021 (14). The decrease in the overall testing of sheep is due to the balance between the lower level of testing in Bulgaria, Italy and Hungary with a decrease of 15,069 (–48.5%), 7208 (–23%), 3830 (–14.5%) sheep tested, respectively, compensated partially by a higher level of testing in Romania and Poland with an overall increase of 8924 (+25.1%) and 3111 (+10.3%) sheep tested, respectively.

Also, in goats, the 2022 pattern of testing by country and flock status was different from that of 2021. In 2022, for each goat tested in a TSE-infected herd in the EU27 and the XI, there were 25.3 goats tested in non-TSE-infected herds, higher than that in 2020 and 2021 (with a value of approximately 14 and 18.3, respectively), but much lower than in 2017 with nearly 31 goats tested in non-TSE-infected herds in 2017. The decrease in testing of 9383 goats is mainly by France, Italy and Bulgaria. That decrease is not compensated by the increase in the number of goats tested by mainly Cyprus (25%) and Poland (23.6%).

Flock status	Infected f	locks				Non-infect	ed flocks			
Country/ Surveillance Target group	ЕМ	NSHC	SHC	SU	Subtotal infected flocks	NSHC	SHC	SU	Subtotal non-infected flocks	Total
AT						2435	123		2558	2558
BE						1483		1	1484	1484
BG						2180	13,845	2	16,027	16,027
СҮ		530	418		948	1482	23		1505	2453
cz						1874	1		1875	1875
DE						11,147	9792	62	21,001	21,001
DK						505			505	505
EE						140			140	140
EL	929	33	175		1137	2796	1157	16	3969	5106
ES	8177		1		8178	11,878	10,139		22,017	30,195
FI						1685	8		1693	1693
FR	32				32	15,117	6701	1	21,819	21,851
HR						1503		2	1505	1505
HU						10,925	11,675		22,600	22,600
IE						10,578	11,129		21,707	21,707
ІТ	2405	250	23		2678	10,559	10,898		21,457	24,135

TABLE 19 Number of sheep tested for TSE by reporting country, surveillance target group and flock status in 2022 in the EU and other reporting countries.

Flock status	Infected f	locks				Non-infect	ted flocks			
Country/ Surveillance Target group	EM	NSHC	SHC	SU	Subtotal infected flocks	NSHC	SHC	SU	Subtotal non-infected flocks	Total
LT						1000			1000	1000
LU						112			112	112
LV						455			455	455
МТ						115	1		116	116
NL						1588			1588	1588
PL						10,044	23,202		33,246	33,246
РТ						15,107	5130		20,237	20,237
RO		670	1439	9	2118	17,729	24,643	18	42,390	44,508
SE						1579			1579	1579
SI						2244	198	2	2444	2444
SK		11			11	12,371			12,371	12,382
Total EU27	11,543	1494	2056	9	15,102	148,631	128,665	104	277,400	292,502
XI ^a						2035	608		2643	2643
Total EU27 + XI	11,543	1494	2056	9	15,102	150,666	129,273	104	280,043	295,145
IS						49	4617	6	4672	4672
МК							559		559	559
NO	23				23	9841	10,237	3	20,081	20,104
RS						157	36		193	193
TR							7		7	7
Total other non-EU	23	0	0	0	23	10,047	15,456	9	25,512	25,535
Total	11,566	1494	2056	9	15,125	160,713	144,729	113	305,555	320,680

Abbreviations: 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 (transmissible spongiform encephalopathies). ^aData from XI, United Kingdom (in respect of Northern Ireland) are available from 2021 onwards.

TABLE 20 Number of goats tested for TSE by reporting country, surveillance target group and herd status in 2022 in the EU and other reporting countries.

Country/ surveillance target groupEMNSHCSHCSubtotal infected herdsSubtotal infected herdsSubtotal infected herdsTotalATSHCSU721721721BE15842586586BG1125310343513221323CY11201680762876533295623438CZ125310341323372337DE1680762876533295623438CZ1680762876533295623438DE1841323372337DK16807628761841323372337DK168016621031407814461582ES68212120,28920,97116,22920,971FI122,23273273273273FR3405405405HU112,22396716,22916,22920,816HU <t< th=""><th>Herd status</th><th>Infected</th><th>herds</th><th></th><th></th><th></th><th>Non-infec</th><th>ted herds</th><th></th><th></th><th></th></t<>	Herd status	Infected	herds				Non-infec	ted herds			
BE 2 586 586 BG 1 253 1034 35 1322 1323 CY 1120 1680 76 2876 533 29 562 3438 CZ 1120 1680 76 2876 533 29 562 3438 CZ - - 713 713 713 713 DE - - 103 1 2337 2337 DK - - 103 1 2337 2337 EE 121 13 2 136 1031 407 8 1446 1582 ES 682 - 2 273 273 273 273 FR - - 12,262 3967 16,229 16,229 16,229 HU - - 402 - 3 405 405 HU - - 111 97 2088 2088 IE - 153 153 153	Surveillance	EM	NSHC	SHC	SU	infected	NSHC	SHC	SU	non-infected	Total
BG 1 253 1034 35 1322 1323 CY 1120 1680 76 2876 533 29 562 3438 CZ 713 713 713 713 713 713 DE	AT						690	31		721	721
CY 1120 1680 76 2876 533 29 562 3438 CZ 713 713 713 713 713 713 DE 1 13 2337 2337 2337 DK 1 13 2337 103 103 EE 121 13 2 136 1031 407 8 1446 1582 ES 682 682 682 10,814 9,475 20,289 20,971 FI 12 13 2 136 1031 407 8 1446 1582 FR 121 13 2 136 1031 9,475 20,289 20,971 FR 12 13 2 1682 3967 16,229 16,229 HR 13 208 405 405 405 405 405 HU 111 97 208 208 208 208 IE 153 153 153 153 153 153	BE						584		2	586	586
CZ 713 713 713 DE 2140 184 13 2337 2337 DK 103 103 103 103 103 EE 2 2 2 2 2 EL 121 13 2 136 1031 407 8 1446 1582 ES 682 682 10,814 9,475 20,289 20,971 FI 2 2 273 273 273 273 FR 2 2 16,229 16,229 16,229 16,229 HR 402 3 907 16,229 3087 405 HU 111 97 208 208 208 IE 153 153 153 153 153	BG		1			1	253	1034	35	1322	1323
DE 2140 184 13 2337 2337 DK 103 103 103 103 103 EE 2 2 2 2 2 EL 121 13 2 136 1031 407 8 1446 1582 ES 682 682 10,814 9,475 20,289 20,971 FI 2 273 273 273 273 FR 2 2 3967 16,229 16,229 HR 405 405 405 405 HU 111 97 208 208 IE 153 153 153 153	СҮ		1120	1680	76	2876	533	29		562	3438
DK 103 103 103 EE 2 2 2 EL 121 13 2 136 1031 407 8 1446 1582 ES 682 682 10,814 9,475 20,289 20,971 FI 2 2 273 273 273 273 FR 2 2 12,262 3967 16,229 16,229 HR 405 405 405 405 405 HU 111 97 208 208 IE 153 153 153 153	cz						713			713	713
EE 2 2 EL 121 13 2 136 1031 407 8 1446 1582 ES 682 682 10,814 9,475 20,289 20,971 FI - - 273 273 273 FR - - 12,262 3967 16,229 16,229 HR - - 402 3 405 405 HU - - 111 97 208 208 IE - - 153 153 153 153	DE						2140	184	13	2337	2337
EL1211321361031407814461582ES68268210,8149,47520,28920,971FI273273273FR12,262396716,22916,229HR4023405405HU11197208208IE-153153153153	DK						103			103	103
ES 682 10,814 9,475 20,289 20,971 FI 273 273 273 273 FR 12,262 3967 16,229 16,229 HR 402 3 405 405 HU 111 97 208 208 IE 153 153 153 153	EE						2			2	2
FI273273273FR12,262396716,22916,229HR4023405405HU11197208208IE153153153153	EL	121	13	2		136	1031	407	8	1446	1582
FR12,262396716,22916,229HR4023405405HU11197208208IE153153153	ES	682				682	10,814	9,475		20,289	20,971
HR4023405405HU11197208208IE153153153153	FI						273			273	273
HU11197208208IE153153153	FR						12,262	3967		16,229	16,229
IE 153 153 153	HR						402		3	405	405
	HU						111	97		208	208
IT 397 45 2 444 5870 20,616 26,486 26,930	IE						153			153	153
	ІТ	397	45	2		444	5870	20,616		26,486	26,930

(Continues)

TABLE 20

(Continued)

Herd status	Infected	herds				Non-infec	ted herds			
Country/ Surveillance target group	EM	NSHC	SHC	SU	Subtotal infected herds	NSHC	SHC	SU	Subtotal non-infected herds	Total
LT						30			30	30
LU						100			100	100
LV						22			22	22
МТ						59			59	59
NL						1702			1702	1702
PL						3673	2,696		6369	6369
РТ						1540			1540	1540
RO		1	1		2	8145	13,971	2	22,118	22,120
SE						109		1	110	110
SI						557	102	9	668	668
SK						375			375	375
Total EU27	1200	1180	1685	76	4141	52,246	52,609	73	104,928	109,069
XI ^a						5			5	5
Total EU27 + XI	1200	1180	1685	76	4141	52,251	52,609	73	104,933	109,074
IS							7		7	7
NO						522	31		553	553
RS						31	41		72	72
TR							1		1	1
Total other non-EU	0	0	0	0	0	553	80	0	633	633
Total	1200	1180	1685	76	4141	52,804	52,689	73	105,566	109,707

Abbreviations: 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 (transmissible spongiform encephalopathies).

^aData from XI, United Kingdom (in respect of Northern Ireland) are available from 2021 onwards.

In total, 557 scrapie cases in sheep were reported in the EU27 and XI in 2022, 6 (1.1%) more than in 2021, a comparable number to the previous year. They were reported by 17 MS and XI (the same number of countries as in 2021). Except for Bulgaria and Finland, all countries that had reported cases in 2021 did so again in 2022 with the addition of Austria and Denmark. In addition, 16 scrapie cases in sheep were reported by one non-EU reporting country: Norway.

CS was reported by five MS: Cyprus, Greece, Italy, Romania, Spain. AS was reported by 14 EU reporting countries: Austria, Croatia, Denmark, France, Germany, Hungary, Ireland, Italy, Poland, Portugal, Slovakia, Slovenia, Spain and Sweden; the United Kingdom in respect of Northern Ireland plus one non-EU reporting country: Norway. Most of the ovine CS cases in the EU27 and XI (99.6%) were reported by four countries, namely Greece, Italy, Romania and Spain.

Out of the 557 sheep scrapie cases reported in the EU27 and XI in 2022, 480 were CS cases (86.2%), 77 were AS cases (13.8%). Among the non-EU reporting countries, no CS cases were reported and 16 AS cases were reported by Norway. Table 21 shows the number of scrapie cases in sheep by reporting country, case type, index case status and surveillance target group. The geographical distribution of AS and CS in 2022 in sheep is shown in Appendix C.

In sheep, 169 (30.3%) of all cases in the EU27 and XI reported in 2022 were index cases (IC): 76 AS and 93 CS. This percentage is slightly lower than the previous year (31.9% in 2021) and so is the absolute number of all cases (176 in 2021) that decreased by 4% in the reporting year. There was a much higher proportion of IC in AS cases (76/77: 98.7%) than in CS cases (93/480: 19.4%), reflecting the within-flock spread of CS. Using the absolute number of IC as a proxy for the flock-level incidence in sheep and comparing 2021 with 2022, there was an increase in the number of CS IC (from 80 in 2021 (17.9%) to 93 in 2022 (19.4%)) and a decrease in the number of AS IC (from 96 to 77, -19.8%). All the AS cases reported by Norway were IC (100%).

In total, 224 scrapie cases in goats were reported in the EU27 and XI in 2022, exactly the same number of cases as in 2021. The increase in cases in Greece from 11 to 43 cases was balanced by the decreased number of cases in Spain from 45 to 22. Only Italy and Spain reported both CS and AS. Bulgaria, Cyprus, Greece and Romania reported only CS cases whereas France and Germany reported only AS cases, two and one cases, respectively. Most of the CS cases were reported from Cyprus with this country's contribution to the 2022 CS caseload similar to that of the previous year (60.3% in 2021 and 59.8% in 2022). The four other non-EU reporting countries that tested goats did not report any scrapie cases.

In total, 216 caprine cases in the EU27 and XI in 2022 were CS cases (96.4%) and 8 were AS (3.6%). Table 22 shows the number of scrapie cases in goats by reporting country, case type, index case status and surveillance target group in 2022. The geographical distribution of AS and CS in 2022 in goats is shown in Appendix C.

In goats, 22.3% (50) of all cases reported in the EU27 and XI in 2022 were IC: 8 AS and 42 CS. This percentage is higher than the 15.6% (35 observed in 2021, with a higher proportion in AS (8/8: 100%) than in CS (42/216: 19.4%). Greece accounted for 46% (23/50) of all IC in goats. Using the absolute number of IC in goats as a proxy for the herd-level incidence in goats and comparing 2022 with 2021, there was an increase in the number of CS IC (from 30 to 42, 40%) and there was a slightly larger number of AS IC (from 5 to 8).

In general, considering the total number of cases by type and without restricting the calculation to IC only, CS is still the most frequently reported type of scrapie in the EU27 in both species of small ruminants while in XI only one case of AS scrapie in sheep was reported. In 2022, the CS/AS ratio was 6.2:1 in sheep (higher than in 2021: 4.4:1) and 27:1 in goats (lower than in 2021: 43.8:1). If, for goats, Cyprus is excluded, the CS/AS ratio was 10.2:1 in 2022 compared with 16.8:1 in 2021.

untry, c	ntry,
	in sh
Number of scrapie	

ie type <u>Atypical</u> ex case <u>No</u> untry/Surveillance EM get group	crapie (AS)							assical	Classical scrapie (CS)	(SS)								
ex case No Intry/Surveillance EM																		
ıntry/Surveillance get group EM			Yes				No	0					Yes					
AT	NSHC SH	SHC Subtotal	NSHC	SHC	su s	T Subtotal <i>µ</i>	Total AS El	EM	NSHC 5	SHC	sU Su	Subtotal N	NSHC	SHC	SU	Subtotal	Total CS	Total
2			-			-	-											-
۲									2			2					2	2
DE			٦			1	-											-
DK			-			-	-											-
EL								18 3	33	38	8	89 4	43	5	8	56	145	145
ES	-	1	9	m		9	10 10	106			106		10	-		11	117	127
FR			9	-		7	7											7
HR					2	2	2											2
Н			m	16	-	19 1	19											19
E				-		1	-											-
IT ^a			m			n	3	118 1	15	23	156	6	8	9		14	170	173
PL			-	m		4	4											4
РТ			21	2	2	23 23	23											23
RO								-	11	20	ю́ Э	34	4	9	2	12	46	46
SE			2			2	2											2
SI			-			1	1											
SK			1			1	1											-
Total EU27 0 0	-	۲	47	26	2 7	75 7	76 24	242 6	61 8	18	3 387		65	18	10	93	480	556
XI ^b			1			1	1											-
Total EU27 + XI 0 0	-	٢	48	26	2 7	76 7	77 24	242 6	61 8	8	3 387		65	18	10	93	480	557
NO			11	5	-	16 1	16											16
Total other non-EU 0 0	0	0	11	2	0	16 1	16	0	0	0	0	0	0	0	0	0	0	16
Total 0 0	1	1	59	31	2 9	92 92	93 24	242 6	61 8	81	3 387		65	18	10	93	480	573
Note: Only the reporting countries in which scrapie cases in sheep were detected in 2022 are mentioned in the table.	crapie cases in	n sheep were detec	ted in 2022 a	re mention	ied in the table.	table.		-			-	-	:					

Abbreviations: EM, Animals culled under TSE eradication measures; NSHC, Animals not slaughtered for human consumption; SHC, animals slaughtered for human consumption; SU, animals culled under TSE eradication measures; NSHC, Animals not slaughtered for human consumption; SU, animals culled under TSE eradication measures; NSHC, Animals not slaughtered for human consumption; SU, animals culled under TSE eradication measures; NSHC, Animals not slaughtered for human consumption; SU, animals culled under TSE eradication measures; NSHC, Animals not slaughtered for human consumption; SU, animals culled under TSE eradication measures; NSHC, Animals not slaughtered for human consumption; SU, animals culled and being infected by a TSE. ^a13 inconclusive cases from Italy are not reported in this table.

^bData from United Kingdom (in respect of Northern Ireland) (XI), are available from 2021 onwards.

IABLE 22 Number of scrapte cases in goats by country, case type, index case status, surveniance target group in 2022 in the EU and other reporting countries.	Der of SC	crapie case	es in goars	by country, c	ase type, II	ndex case	status, survei	llance target	group ir	1 2022 IN U	ie EU and		sporting cour	irries.				
Case type	Atypi	Atypical scrapie (AS)	ie (AS)						Classic	Classical scrapie (CS)	(CS)							
Index case	No				Yes			Total	No					Yes			Total	Total
Surveillance target group	EM	NSHC	SHC	Subtotal	NSHC	SHC	Subtotal	AS	EM	NSHC	SHC	SU	Subtotal	NSHC	SHC	SU Sub	CS Subtotal	
BG										-			1		8	8	6	6
CY										24	44	62	130	4		4	134	134
DE					-		-	-										1
EL									5	13	2		20	21	2	23	43	43
ES					-	-	2	2	17				17	ŝ		m	20	22
FR					2		2	2										2
F					-	2	ŝ	ŝ	2		2		4		4	4	8	11
RO										-			2				2	2
Total EU27 + XI ^a	0	0	0	0	'n	m	8	8	24	39	49	62	174	28	14	0 42	216	224
Total other non-EU	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
Total	0	0	0	0	5	e	8	8	24	39	49	62	174	28	14	0 42	216	224
	40100 00	للم تطريب من مم ت	a see of monor	or in contract	10404040		مطغمة لممام ما ممنا	ماطمه										

Number of scrapie cases in goats by country, case type, index case status, surveillance target group in 2022 in the EU and other reporting countries. TABLE 22

Notes: Only the reporting countries in which scrapie cases in goats were detected in 2022 are included in the table.

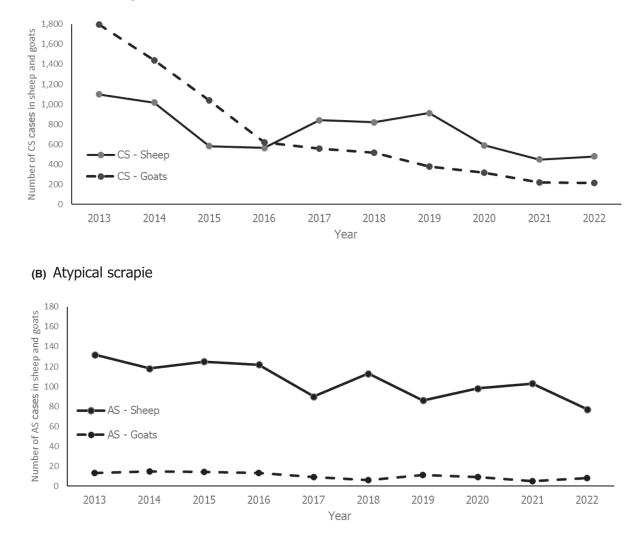
Abbreviations: EM, animals culled under TSE eradication measures; NSHC, Animals not slaughtered for human consumption; SHC, animals slaughtered for human consumption; SU, animals clinically suspected of being infected by a TSE (transmissible spongiform encephalopathies).

T

^aData from United Kingdom (in respect of Northern Ireland) (XI) are available from 2021 onwards.

Focusing on the last 10 years (2013–2022), the evolution in the number of scrapie cases detected at EU27 and the United Kingdom level for the period 2013–2020 and at EU27 and XI level for the period 2021–2022 level is shown for each species and by case type in Figure 2. After the 2006 peak in the number of reported CS scrapie cases in sheep in the EU with 3142 (when the number of tests also peaked), CS cases have decreased from 1416 in 2011 to 554 in 2016. An increase was observed in 2017 (839 CS and unknown cases), and a similar order of magnitude was observed in 2018 (820 cases). A new increase to 911 was observed in 2019, mainly due to CS cases from TSE-infected flocks reported by Greece, Italy and Spain. A decrease to 589 CS cases was reported in EU27 and the UK in 2020, largely as a result of the decrease in cases in Greece, Spain, Italy and Romania. Another decrease occurred in 2021 with 448 cases, primarily due to the lower number of CS cases in Greece, Romania and Spain. Differently in 2022 there was an increase with 480 cases, primarily due to the higher number of CS cases in Greece (+95.9%) and Italy (+14.9%) despite the decrease in cases in Spain (-36.4%). This change in trend is associated with an approximately a twofold increase in cases in both TSE-infected and uninfected flocks in Greece and TSE-infected flocks in Italy.

In goats, although to a limited extent, the decreasing trend in the absolute number of CS cases (from 219 to 216) continued in 2022. The long-term evolution is mainly associated with one single MS (Cyprus), where the number of detected cases has consistently declined since the peak in 2013 when 1792 cases were reported by the country and 1799 in total including all other MS. However, in 2022, Cyprus remained stable with 134 cases (4 IC) compared to 135 (5 IC) in 2021. In 2022, compared with 2021, there was a reduction in the number of CS in goats in Spain from 43 to 20 (–53.5%) and Italy from 23 to 8 (–65.2%), but an increase in the number of CS in goats in Greece by about four times, from 11 to 43.



(A) Classical scrapie

FIGURE 2 Number of reported scrapie cases in sheep and goats in the EU and the UK by case type in the period 2012–2022 in (A) CS and (B) AS.

Focusing on the last 10 years, based on the cases of scrapie with known type, species and age in sheep (8630 cases), the average age of AS cases (89.9 months) is significantly higher (p < 0.001) than that of CS cases (54.1 months). Similarly, in goats (6784 cases), the average age of AS cases (91.4 months) is significantly higher (p < 0.001) than that of CS cases (54 months). Similarly, in goats (6784 cases), the average age of AS cases (91.4 months) is significantly higher (p < 0.001) than that of CS cases (54 months). Comparing sheep and goats, no significant difference in mean age was found for either AS (p = 0.75) or CS (p = 0.8).

Tables 23 and 24 show the cases of CS and AS, respectively, in sheep for the period 2002–2022, with a focus on the last 5 years. Tables 25 and 26 show the cases of CS and AS, respectively, in goats for the period 2002–2022, with a focus on the last 5 years.

Country	Up to 2017	2018	2019	2020	2021	2022	Total CS
BE	38						38
BG	12	5		8	6		31
СҮ	3206	4	1	2	2	2	3217
CZ	56						56
DE	116						116
EL	5991	178	276	176	74	145	6840
ES	1386	279	312	244	184	117	2522
FR	1534						1534
HU	10						10
IE	586	1					587
ΙТ	2899	150	171	102	148	170	3640
NL	401						401
РТ	33		1	5			39
RO	809	203	141	52	34	46	1285
SI	174						174
SK	132						132
Total EU27	17,383	820	902	589	448	480	20,622
XI ^a							
Total EU27 + XI	17,383	820	902	589	448	480	20,622
IS	207	21	21	53	55		357
NO	17						17
United Kingdom	1995		9				2004
Total other non-EU	2219	21	30	53	55	0	2378
Total	19,602	841	932	642	503	480	23,000

Note: Only the reporting countries in which classical scrapie cases in sheep were detected are included in the table. The table with all historical cases can be found on https://doi.org/10.5281/zenodo.4106089.

^aData from United Kingdom (in respect of Northern Ireland) (XI) are available from 2021 onwards.

TABLE 24	Number of atypical scrapie	cases in sheep by year and country	y between 2002 and 2022 in the report	ina countries.

Country	Up to 2017	2018	2019	2020	2021	2022	Total AS
AT	14	1				1	16
BE	8			2			10
BG	6						6
CZ	8						8
DE	124	4	4	14	5	1	152
DK	13	1				1	15
EE	2						2
EL	32			1			33
ES	223	9	7	12	6	10	267
FI	13	2	3	1	1		20
FR	557	6	8	3	7	7	588
HR	2				2	2	6
HU	143	13	17	14	16	19	222
IE	36	8	6	1	1	1	53
ІТ	94	8	7	6	6	3	124
NL	18						18
PL	54	6	4	5	11	4	84
PT ^a	646	30	20	18	32	23	769
RO	0			1			1

(Continues)

TABLE 24 (Continued)

Country	Up to 2017	2018	2019	2020	2021	2022	Total AS
SE	47	2		1	1	2	53
SI	10				2	1	13
SK	32	6	4	5	12	1	60
Total EU27	2082	96	80	84	102	76	2520
ХІ ^b					1	1	2
Total EU27+ XI	2082	96	80	84	103	77	2522
IS	8				1		9
NO	151	8	10	12	8	16	205
United Kingdom	345	17	6	14			382
Total other non-EU	504	25	16	26	9	16	596
Total	2586	121	96	110	112	93	3118

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

^aln 2018, it includes 1 one imported case: an animal imported for slaughter from Spain which died at lairage/resting area of the slaughterhouse and was tested as NSHC. ^bData from United Kingdom (in respect of Northern Ireland) (XI) are available from 2021 onwards.

Country	Up to 2017	2018	2019	2020	2021	2022	Total CS
BG	9	5		7	4	9	34
СҮ	10,770	381	308	236	135	134	11,964
EL	575	19	7	27	11	43	682
ES	140	89	35	29	43	20	356
FI	8						8
FR	168	5					173
HU	-		1				1
ІТ	109	15	26	13	23	8	194
RO	13	3		5	3	2	26
SI	4						4
Total EU27	11,796	517	377	317	219	216	13,442
XI ^a	0						0
Total EU27 + XI	11,796	517	377	317	219	216	13,442
United Kingdom	229		2	2			233
Total other non-EU	229	0	2	2	0	0	233
Total	12,025	517	379	319	219	216	13,675

TABLE 25 Number of classical scrapie cases in goats by year and country between 2002 and 2022 in the reporting countries.

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

^aData from United Kingdom (in respect of Northern Ireland) (XI) are available from 2021 onwards.

TABLE 26 Number of atypical scrapie cases in goats by year and country between 2002 and 2022 in the EU and other reporting countries.

Country	Up to 2017	2018	2019	2020	2021	2022	Total AS
AT	1						1
СҮ	2	1	1				4
DE	2					1	3
DK	0			1			1
EL	4	1					5
ES	51	2	2	3	2	2	62
FI	1						1
FR	58		3	1	1	2	65
IT	24	2	3	3	2	3	37
PL	0		1				1
РТ	13		1	1			15

TABLE 26 (Continued)							
Country	Up to 2017	2018	2019	2020	2021	2022	Total AS
SI	1						1
Total EU27	157	6	11	9	5	8	196
XI ^a	0						0
Total EU27+ XI	157	6	11	9	5	8	196
NO	1						1
Total other non-EU	1						1
Total	158	6	11	9	5	8	197

TABLE 26 (Continued)

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

^aData from United Kingdom (in respect of Northern Ireland) (XI) are available from 2021 onwards.

The number of historical reported scrapie cases can be found in the following https://doi.org/10.5281/zenodo.4252321, as follows:

TABLE 27 Number of classical scrapie cases in sheep per country and year from 2002 in the EU and non-EU reporting countries.

 TABLE 28
 Number of atypical scrapie cases in sheep per country and year from 2002 in the EU and non-EU reporting countries.

 TABLE 29
 Number of classical scrapie cases in goats per country and year from 2002 in the EU and non-EU reporting countries.

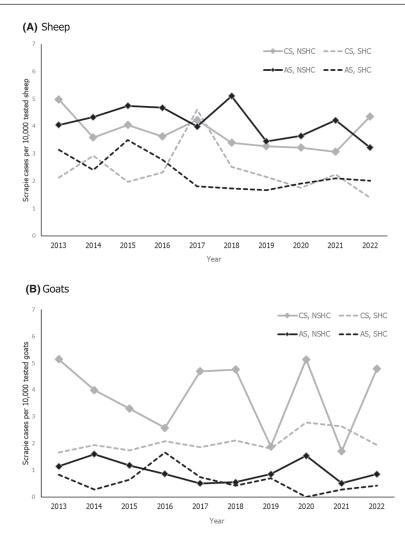
TABLE 30 Number of atypical scrapie cases in goats per country and year from 2002 in the EU and non-EU reporting countries.

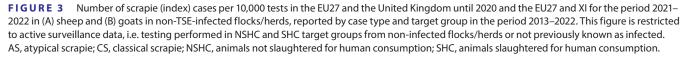
In sheep, in 2022, the number of IC of CS and AS per 10,000 tests carried out by target group at EU27 and XI level was: (1) for CS: 4.3 in NSHC and 1.4 in SHC; (2) for AS: 3.2 in NSHC and 2 in SHC.

In goats, in 2022, the number of IC of CS and AS per 10,000 tests carried out by target group at EU27 and XI level was: (1) for CS: 5.4 in NSHC and 2.7 in SHC; (2) for AS: 1 in NSHC and 0.6 in SHC.

Figure 3 shows the 10-year trend (2013–2022): by target group of the number of scrapie cases per 10,000 tests of sheep and goats in TSE non-infected flocks/herds and separately per case type. The data consist of those for EU 27 and the United Kingdom for the period 2013–2020, and EU27 and XI for the period 2021–2022. The results of the Poisson regression model showed a statistically significant decreasing trend for CS (annual RR: 0.97, p < 0.01) and AS (annual RR=0.966, p < 0.01) in sheep. In both cases the average decrease was about 3% per year. In goats, the model did not show any statistically significant trend for either CS or AS (p=0.53 and 0.08, respectively), although for AS it was borderline.

Based on the same model, the probability of detecting CS in the NSHC surveillance target group was higher than that in SHC one in both sheep (RR: 1.58, p < 0.0001) and goats (RR: 1.71, p < 0.0001). The same was true for AS, where a statistically significant higher probability was observed in both sheep and goats (respectively RR=1.8, p < 0.0001, and RR=1.59, p < 0.05).





Tables 31 and 32 summarise the number of discriminatory tests performed by country in 2022 for CS and AS in sheep. Tables 33 and 34 summarise the number of discriminatory tests performed by country in 2022 for CS and AS in goats. In sheep, 478 (99.6%) of the CS reported in the EU27 and XI were submitted for discriminatory testing and so were 34 of the AS cases (44.7%). The 16 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'. In goats, 86 (39.8%) of the CS reported in the EU and XI were submitted for discriminatory testing as well as 5 of the AS cases (62.5%). All goat cases subjected to discriminatory testing were confirmed as 'BSE excluded'.

TABLE 31 Number of discriminatory tests and results in classical scrapie cases in sheep by reporting country.

		Cases submi	tted for discriminator	ry testing	
Country	No. of classical scrapie, CH1641-like and inconclusive cases	BSE-not- excluded	BSE-excluded	Total	% of total classical scrapie and inconclusive case ^a
СҮ	2		0	0	0%
EL	145		145	145	100%
ES	117		117	117	100%
ІТ	170		170	170	100%
RO	46		46	46	100%
Total EU27	480	0	478	478	99.6%
Total other non-EU	0	0	0	0	-
Total	480	0	478	478	99.6%

Note: Reporting countries without classical scrapie cases in sheep are not included in the table.

^aIndicates the proportion of classical TSE cases that are submitted to discriminatory testing by each reporting country.

TABLE 32	Number of discriminatory tests and results in atypical scrapie cases in sheep by reporting country.
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		Cases submitted for discriminatory testing					
Country	No. of atypical scrapie	BSE-not- excluded	BSE-excluded	Total	% of total atypical scrapie cases ^a		
AT	1		0	0	0%		
DE	1		0	0	0%		
DK	1		0	0	0%		
ES	10		10	10	100%		
FR	7		0	0	0%		
HR	2		0	0	0%		
HU	19		19	19	100%		
IE	1		0	0	0%		
ІТ	3		3	3	100%		
PL	4		0	0	0%		
РТ	23		0	0	0%		
SE	2		0	0	0%		
SI	1		1	1	100%		
SK	1		1	1	100%		
Total EU27	76	0	34	34	44.7%		
ХІ ^b	1		1	1	100%		
Total EU27 + XI	77	0	35	35	45.4%		
NO	16		16	16	100%		
Total other non-EU	16	0	16	16	100%		
Total	93	0	51	51	54.8%		

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

Abbreviations: BSE, bovine spongiform encephalopathy; TSE, transmissible spongiform encephalopathies.

^aIndicates the proportion of atypical TSE cases that are submitted to discriminatory testing by each reporting country.

^bData from XI, United Kingdom (in respect of Northern Ireland) (XI) are available from 2021 onwards.

		Cases submitt	ed for discriminatory tes	sting	
Country	No. of classical scrapie cases	BSE-not- excluded	BSE-excluded	Total	% of total classical scrapie cases ^a
BG	9		9	9	100%
СҮ	134		4	4	3%
EL	43		43	43	100%
ES	20		20	20	100%
ΙТ	8		8	8	100%
RO	2		2	2	100%
Total EU27	216	0	86	86	39.8 %
Total	216	0	86	86	39.8%

TABLE 33 Number of discriminatory tests and results in classical scrapie cases in goats in 2022 by reporting country.

Note: EU and reporting countries without atypical scrapie cases in sheep are not included in the table. Abbreviations: BSE, bovine spongiform encephalopathy; TSE, transmissible spongiform encephalopathies.

^aIndicates the proportion of classical TSE cases that are submitted to discriminatory testing by each reporting country.

TABLE 34	Number of discriminatory tests and results in atypical scrapie cases in goats by reporting country.
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		Cases submitted for discriminatory testing								
Country	No. of atypical scrapie cases	BSE-not-excluded	BSE-excluded	Total	% of total atypical scrapie cases ^a					
DE	1		0	0	0%					
ES	2		2	2	100%					
FR	2		0	0	0%					

TABLE 34 (Continued)

		Cases submitted for d	Cases submitted for discriminatory testing								
Country	No. of atypical scrapie cases	BSE-not-excluded	BSE-excluded	Total	% of total atypical scrapie cases ^a						
ІТ	3		3	3	100%						
Total EU27	8	0	5	5	62.5%						
Total	8	0	5	5	62.5%						

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

Abbreviations: BSE, Bovine spongiform encephalopathy; TSE, Transmissible spongiform encephalopathies.

^aIndicates the proportion of atypical TSE cases that are submitted to discriminatory testing by each reporting country.

3.2.1 | Genotyping

3.2.1.1 | Sheep

The classification of genotypes of the sheep prion protein *PRNP* gene used in this report, based on an adaptation from the Great Britain's National Scrapie Plan (NSP), is summarised in Table 35 (the risk is relative to classical scrapie only).

TABLE 35 Classification of the genotypes of the sheep prion protein PRNP gene according to Great Britain's National Scrapie Plan (NSP).

NSP group	Genotype	Risk of classical scrapie (Hunter, 2003) ^a
NSP1	ARR/ARR	Most resistant to scrapie
NSP2	ARR/ARQ; ARR/ARH; ARR/AHQ	Resistant to scrapie but offspring may be susceptible depending on genotype of the other parent
NSP3	ARQ/ARQ	Higher risk of scrapie in these sheep and in offspring
NSP3/Other (NSP3O)	AHQ/AHQ; ARH/ARH; ARH/ARQ; AHQ/ARH; AHQ/ARQ	Higher risk of scrapie in these sheep and in offspring
NSP4	ARR/VRQ	Susceptible to scrapie but could be used as a breeding source of the ARR allele associated with resistance
NSP5	ARQ/VRQ; ARH/VRQ; AHQ/VRQ; VRQ/VRQ	Sheep of highest susceptibility to scrapie in self and offspring

^aInformation extracted from the DEFRA National Scrapie Plan for Great Britain, Ram genotyping scheme.

Table 36 shows the genotypes of sheep scrapie cases in 2022 in the EU and other reporting countries.

In total, 439 (97.3%) of the 451 cases of CS in sheep with genotype reported in the EU27 and XI in 2022 (91.4% of the total CS caseload) were from the susceptible genotype groups (NSP3, NSP3O NSP4 or NSP5). This is similar to previous years in which over 97.5% of all CS cases with known genotypes were from the susceptible groups. In the other non-EU reporting countries, no CS cases were reported. In the current year, no country has reported cases of CS in a sheep with the ARR/ARR genotype (NSP1), a very rare occurrence which has been reported in the past (e.g. by Spain in 2019 and by Romania in 2020).

Among ovine AS cases, 38 (52.8%) of the 70 cases of AS in sheep with NSP genotype reported in the EU27 and XI in 2022 (50% of the total AS caseload) were from the genotype groups NSP3, NSP3O NSP4 or NSP5), very similar to 2021. The additional 33 AS cases had genotypes in the NSP1 or NSP2.

 TABLE 36
 Distribution of genotypes of confirmed scrapie cases in sheep by reporting country and National Scrapie Plan (NSP) group.

	Atyp	Atypical scrapie							Classical scrapie								
Country/NSP types	NSP1	NSP2	NSP3	NSP3O	NSP4	NSP5	Unknown ^a N/G ^b	Total AS	NSP1	NSP2	NSP3	NSP3O	NSP4	NSP5	Unknown N/G	Total CS	Total scrapie cases
AT				1				1									1
СҮ															2	2	2
DE				1				1									1
DK				1				1									1
EL											94	38	8	5		145	145
ES		3	5				2	10		1	85			4	27	117	127
FR	1	1	2				3	7									7
HR		1	1					2									2
HU	1	11	2	5				19									19

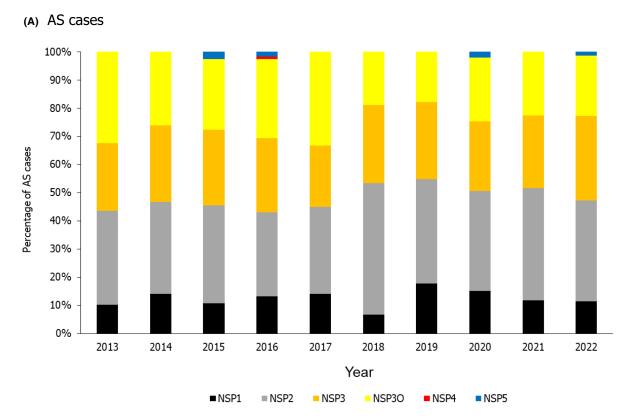
	Atyp	oical scr	apie						Class	sical scr	apie						
Country/NSP types	NSP1	NSP2	NSP3	NSP3O	NSP4	NSP5	Unknown ^a N/G ^b	Total AS	NSP1	NSP2	NSP3	NSP3O	NSP4	NSP5	Unknown N/G	Total CS	Total scrapie cases
IE				1				1									1
ІТ				3				3			153	12		5		170	173
PL	1	2		1				4									4
РТ	3	7	10	2		1		23									23
RO										11	18	3		14		46	46
SE			1				1	2									2
SI	1							1									1
SK	1							1									1
Total EU27	8	25	21	15		1	6	76		12	350	53	8	28	29	480	556
XI ^c				1				1									1
Total EU27 + XI	8	25	21	16		1	6	77		12	350	53	8	28	29	480	557
NO	1	4	5	5		1		16									16
Total other non-EU	1	4	5	5		1		16									16
Total	9	29	26	21		2	6	93		12	350	53	8	28	29	480	573

^aUnknown: genotype other than those included in the NSP list.

^bN/G: not genotyped.

^cData from XI, United Kingdom (in respect of Northern Ireland) are available from 2021 onwards.

Figure 4 shows the frequency distribution of genotypes of sheep scrapie cases by case type, year and NSP group in the period 2013–2022 in the reporting countries.





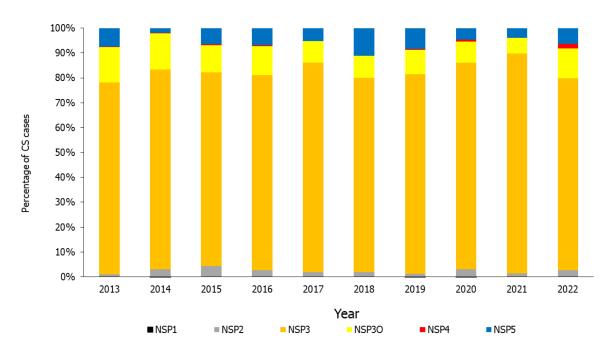


FIGURE 4 Frequency distribution of genotypes of sheep scrapie cases by case type (A) AS cases (B) CS cases, year and National Scrapie Plan (NSP) group in the period 2013–2022 in the reporting countries. (A) Atypical scrapie. (B) Classical scrapie. NSP1: resistant (black); NSP2: semi-resistant (grey); NSP3 (orange) + NSP3O (yellow) + NSP4 (red) + P5 (blue): susceptible as referred to in Table 35.

Table 37 shows the genotypes obtained in 2022 from the random samples of tested sheep in the reporting countries. In the EU27 and XI, following the changes in the legislation that entered into force in 2018, eight MS conducted the genotyping of a random sample of sheep: Belgium, Cyprus (where genotyping is conducted systematically in the breeding sheep population), France, Germany, Italy, Latvia, the Netherlands and Poland. One of the eight other non-EU reporting countries, Iceland, also reported a large number of genotypes, obtained from scrapie outbreaks rather than from a random sample of the sheep population. The subset of EU27 and XI (excluding data from Cyprus) that carried out the activity in 2022 reported a total of 7904 known genotypes: 7.3% (575) of the sheep population (with known genotype) were susceptible to CS (NSP3, NSP3O, NSP4 and NSP5), lower than the 7.9% in 2021 and the 8.8% in 2020. This percentage stands at 23.2% in Italy, one of the countries with the highest case load in 2022, whereas it was between 5.1% (Germany) and 7.9% (France) in the other six MS.

After excluding Cyprus, the proportion of sheep in the resistant genotype group (NSP1; black colour in the bars of Figure 5) shifted from 28.6% of the total number of genotyped sheep in 2011 (in which 23 MS contributed) to 71.8% in 2022 (to which seven MS contributed), the highest ever recorded, and 5.5% higher than the 66.8% of 2021, to which eight MS contributed.

TABLE 37Number of genotyped animals (% of sample within country) in randomly selected sheep in the EU and other reporting countries in2021 by reporting country and National Scrapie Plan (NSP) group, in accordance with Regulation (EC) 999/2001 Annex VII, Chapter C, Part I, point 8.

	Number of geno	otyped animals	(% of sample witl	hin country)				
Country	NSP1	NSP2	NSP3	NSP3O	NSP4	NSP5	Other	Total
AT	n/a ^d	n/a	n/a	n/a	n/a	n/a	n/a	
BE	443 (70.3%)	138 (21.9%)	18 (2.9%)	13 (2.1%)	14 (2.2%)	4 (0.6%)	0 (0%)	630
BG	n/a	n/a	n/a	n/a	n/a	n/a	n/a	
CY ^a	56,300 (92.9%)	3785 (6.2%)	140 (0.2%)	62 (0.1%)	74 (0.1%)	13 (0.02%)	232 (0.4%)	60,606
cz	n/a	n/a	n/a	n/a	n/a	n/a	n/a	
DE	3904 (78.6%)	806 (16.2%)	186 (3.7%)	61 (1.2%)	6 (0.1%)	1 (0.02%)	1 (0.02%)	4965
DK	n/a	n/a	n/a	n/a	n/a	n/a	n/a	
EE	n/a	n/a	n/a	n/a	n/a	n/a	n/a	
EL	n/a	n/a	n/a	n/a	n/a	n/a	n/a	
ES	n/a	n/a	n/a	n/a	n/a	n/a	n/a	
FI	n/a	n/a	n/a	n/a	n/a	n/a	n/a	
FR	224 (62%)	79 (21.9%)	15 (4.2%)	2 (0.6%)	9 (2.5%)	0 (0%)	32 (8.9%)	361
HR	n/a	n/a	n/a	n/a	n/a	n/a	n/a	
HU	n/a	n/a	n/a	n/a	n/a	n/a	n/a	
IE	n/a	n/a	n/a	n/a	n/a	n/a	n/a	
IT	225 (33.9%)	283 (42.7%)	122 (18.4%)	23 (3.5%)	4 (0.6%)	5 (0.8%)	1 (0.2%)	663
LT	n/a	n/a	n/a	n/a	n/a	n/a	n/a	
LU	n/a	n/a	n/a	n/a	n/a	n/a	n/a	
LV	31 (64.6%)	14 (29.2%)	3 (6.3%)	0 (0%)	0 (0%)	0 (0%)	0 (0%)	48
МТ	n/a	n/a	n/a	n/a	n/a	n/a	n/a	
NL	829 (69.1%)	261 (21.8%)	19 (1.6%)	34 (2.8%)	23 (1.9%)	7 (0.6%)	27 (2.3%)	1200
PL	61 (62.2%)	31 (31.6%)	5 (5.1%)	1 (1%)	0 (0%)	0 (0%)	0 (0%)	98
PT	n/a	n/a	n/a	n/a	n/a	n/a	n/a	
RO	n/a	n/a	n/a	n/a	n/a	n/a	n/a	
SE	n/a	n/a	n/a	n/a	n/a	n/a	n/a	
SI	n/a	n/a	n/a	n/a	n/a	n/a	n/a	
SK	n/a	n/a	n/a	n/a	n/a	n/a	n/a	
Total EU27	62,017 (90.4%)	5397 (7.9%)	508 (0.7%)	196 (0.3%)	130 (0.2%)	30 (0.04%)	293 (0.4%)	68,571
XI ^a	n/a	n/a	n/a	n/a	n/a	n/a	n/a	00,07
	62,017 (90.4%)	5397 (7.9%)	508 (0.7%)	196 (0.3%)	130 (0.2%)	30 (0.04%)	293 (0.4%)	68,571
BA	n/a	n/a	n/a	n/a	n/a	n/a	n/a	00,071
СН	n/a	n/a	n/a	n/a	n/a	n/a	n/a	
IS ^c	0 (0%)	54 (0.1%)	33,885 (82.1%)	4491 (10.9%)	0 (0%)	2368 (5.7%)	460 (1.1%)	41,258
ME	n/a	n/a	n/a	n/a	n/a	n/a	n/a	1,230
МК	n/a	n/a	n/a	n/a	n/a	n/a	n/a	
NO	n/a	n/a	n/a	n/a	n/a	n/a	n/a	
RS	n/a	n/a	n/a	n/a	n/a	n/a	n/a	
TR	n/a	n/a	n/a	n/a	n/a	n/a	n/a	
Total other	n/a 0 (0%)	n/a 54 (0.1%)	n/a 33,885 (82.1%)	n/a 4491 (10.9%)	n/a 0 (0%)	n/a 2368 (5.7%)	n/a 460 (1.1%)	41,258
non-EU					- (- · · · /			
Total	62,017 (56.5%)	5451 (5%)	34,393 (31.3%)	4687 (4.3%)	130 (0.1%)	2398 (2.2%)	753 (0.7%)	109,829

^aThe Cyprus data are different from those of other reporting countries since Cyprus systematically genotypes the breeding sheep population.

^bData from XI, United Kingdom (in respect of Northern Ireland) are available from 2021 onwards.

 c lceland reported genotypes obtained from scrapie outbreaks rather than from a random sample of the sheep population. d n/a: not applicable.

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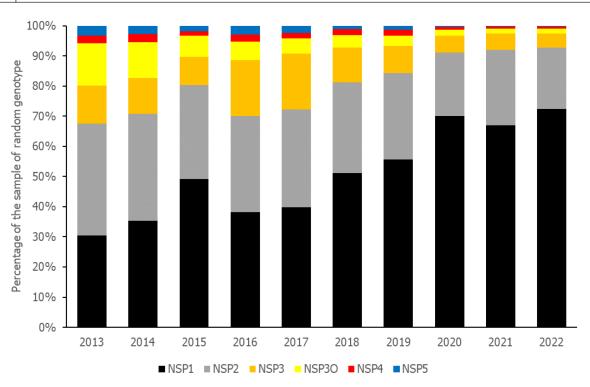


FIGURE 5 Frequency distribution of the six genotype National Scrapie Plan (NSP) groups in sheep randomly sampled for genotyping in the EU in the period 2013–2022 according to Regulation (EC) 999/2001, Annex III, Chapter A, Part II, point 8 until end of 2017 and Annex VII, in Chapter C, in Part 1, point 8 from 2018. The table below the figure describes the number of contributing MS. Data from Cyprus were excluded. NSP1: resistant (black); NSP2: semi-resistant (grey); NSP3 (orange) + NSP3O (yellow) + NSP4 (red) + NSP5 (blue): susceptible as referred to in Table 36.

	2013	2014	2015	2016	2017	2018	2019	2020	2021	2022
Number of contributing MS	24	25	25	25	20	7	7	8	8	7
Total genotyped	8340	9437	9823	9413	8871	2713	3180	7985	8068	7904

3.2.1.2 | Goats

In 2022, at least one polymorphism at either codon 146 or 222 was reported from 184 cases (5 AS, 179 CS) by Cyprus, Greece, Spain and Italy, four of the eight countries in the EU27 and XI that reported cases. A total of 118 (64.1%) CS cases and 5 AS (100%) cases were genotyped at both codon 146 and 222. Two cases of CS reported by Cyprus were heterozygous goats at codon 146: DQ/NQ and DZ/NZ. Table 38 shows the genotypes of goat scrapie cases in 2022 in the EU. None of the non-EU reporting countries reported any case.

Case type	Atypical S	Atypical Scrapie			Classical Scrapie							
Country	NQ/NQ	ZZ/ZZ	Total AS	DQ/NQ	DZ/NZ	NQ/NQ	NZ/NZ	ZZ/ZZ	Total CS	Total		
BG								9	9	9		
СҮ				1	1	54	52	26	134	134		
DE		1	1							1		
EL						43			43	43		
ES	2		2			12	8		20	22		
FR		2	2							2		
ІТ	3		3			8			8	11		
RO								2	2	2		
Total	5	3	8	1	1	117	60	37	216	224		

TABLE 38	Distribution of genotypes of confirmed scrapie cases in goats.
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Abbreviations: N, wild-type codon 146; D, mutated codon 146; Q, wild-type codon 222; options ZK, ZQ, NZ, SZ, DZ, used when one of the codons could not be or was not determined; ZZ, used if the genotype could not be determined, was not determined or was different from the available options.

3.3 | TSE surveillance in cervids

In 2022, 3202 cervids were tested for TSE in the EU27 and XI. The 10 MS that contributed to the monitoring were Austria, Denmark, Estonia, Finland, Hungary, Italy, Latvia, Romania, Spain and Sweden. Two countries, Sweden and Romania, contributed to 59.1% of the total number of tested cervids in the EU and XI, with 20.2% (646) and 38.9% (1246), respectively.

Out of the 3202 cervids tested by the MS, 649 (20.3%) were captive, farmed or semi-domesticated animals, mostly reindeer (518), followed by red deer (104). Among the 2553 (79.7%) wild cervids tested by MS, 1454 (57%) were roe deer and 588 red deer (23%).

When considering the target groups, the most commonly tested group by the MS was the 'Hunted/slaughtered fit for human consumption' (HSHC) with 2024 animals (63.2%); Romania and Sweden contributed particularly to this group. A total of 1178 cervids were tested in the risk groups: 785 (24.5% of the total tested) fallen/culled (FC); 304 (9.5%) road/ predator killed (RK); 66 (2.1%) clinical suspect animals (SUS); 23 (0.7%) hunted/slaughtered not fit for human consumption (HSNHC). The numbers of tested cervids by reporting country, management system and target group in 2022 are displayed in Table 39.

One cervid in Finland tested positive. It was a female European moose over 12 months from the FC group.

In 2022, Norway tested 17,583 animals, mostly semi-domesticated reindeer (37.9%), followed by wild European moose (17.9%) and wild reindeer (17.5%). HSHC animals accounted for 82% of total. Two cases of CWD in wild European moose, one in wild reindeer and one in a red deer were reported by Norway. All cases were female; three reported as FC and one as HSHC.

Iceland and Serbia also reported 79 (semi-domesticated/farmed reindeer) and 281 (80.4% wild roe deer) cervids tested in 2022, respectively. They were all negative.

The description of the five CWD cases detected in 2022 is shown in Table 40.

TABLE 39		Number of tested cervids in the EU and reporting countries by management system, species, country and target group in 2022.	in the E	U and re	porting c	ountries	by manag	ement sysi	tem, species	s, country an	nd target	group in 202	2.								
, ee.		Semi-domesticated/farmed deer species ^a	ticated	/farmed	deer spe	cies ^a					Wild de	Wild deer species ^a									
Managen species ^b a group ^c	Management system species ^b and target group ^c	European Deer Moose	an	Fallow deer R	Reindeer	Roe deer	Red deer	Sika r deer	White- tailed deer	Sub-Total Deer		European Moose	Fallow deer I	Reindeer	Roe I deer	Red deer	V tı Sika deer d	White- tailed deer	Sub- Total	Total	<u> </u>
АТ	FC	-		-			2	-		9									-	7	
	Total	-		-	-		2	-		9						-			-	7	
DK	FC												105		30	201	81		417	417	
	Total												105		30	201	81		417	417	
H	FC	£								m										m	
	HSHC											-							-	1	
	Total	3								e		٢							٢	4	
ES	SUS															-			-	۲	
	RK										-				4				5	5	
	FC	-									4					2			9	7	
	HSNHC												c			ŝ			9	9	
	HSHC										62		16		19	138			235	235	
	Total	-								۲	67		19		23	144			253	254	
H	RK				ŝ					ŝ		7			2		1	12	21	24	
	FC	2			11	2			2	17		32		2	10			6	53	70	
	HSNHC				۲					-		ŝ						2	Ŋ	9	
	HSHC											66							66	66	
	Total	7			15	7			2	21		141		2	12		7	23	178	199	
ЯH	SUS						-			-					£	2			5	9	
	FC									-	-					-			2	ß	
	HSNHC					-				-					-				-	2	
	Total					2	-			m	-				4	٣			∞	11	
F	SUS					-				-					18	8			26	27	
	RK												26		213	25			264	264	
	Ę			7		ŝ	4			14			2		86	20			111	125	
	Total			7		4	4			15			31		317	53			401	416	
۲۸	FC											-							2	2	
	Total											-				-			2	2	
RO	RK														2	2			4	4	
	FC														62	80			70	70	

Number of tested cervids in the EU and reporting countries by management system. species. country and target group in 2022. TABLE 39

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ountry /																				
Management syster species ^b and target group ⁶	F	Deer	European Moose	Fallow deer	Reindeer	Roe deer	Red deer	Sika deer	White- tailed deer	Sub-Total	Deer	European Moose	Fallow deer	Reindeer	Roe deer	Red deer	Sika deer	White- tailed deer	Sub- Total	Total
	HSNHC														∞				80	∞
	HSHC														992	172			1164	1164
	Total														1064	182			1246	1246
SE	SUS											29			c				32	32
	RK				7					7										7
	FC		-		59		6			69		6			-	2			12	81
	HSNHC											-							-	-
	HSHC				436		88			524						-			-	525
	Total		-		502		97			600		39			4	m			46	646
Total EU		ŝ	e	œ	518	ø	104	-	2	649	68	182	155	2	1454	588	81	23	2553	3202
IS	FC				m					m										ŝ
	HSHC				76					76										76
	Total				79					79										79
NO	RK	2			36					38	41	355		-	1443	232			2072	2110
	FC	5			71		11			87	56	302		44	351	244			667	1084
	HSHC	6		17	6552		317			6895	107	2494		3033	23	1837			7494	14,389
	Total	16		17	6659		328			7020	204	3151		3078	1817	2313			10,563	17,583
RS	RK										-				m				4	4
	FC										27				59				86	86
	HSNHC																		-	-
	HSHC										œ				163	19			190	190
	Total										36				226	19			281	281
Total non-EU		16		17	6738		328			7099	240	3151		3078	2043	2332			10,844	17,943
Total		21	e	25	7256	8	432	-	2	7748	308	3333	155	3080	3497	2920	81	23	13,397	21,145
Ily countries er: not spec	^a Only countries that reported tested cervids are included in the table. ^b Deer: not specified. Moose (or Eurasian/European elk) (<i>Alces alces</i>). Fallow deer (<i>Dama dama</i>). Reindeer: Eurasian tundra reindeer (<i>Rangifer tarandus</i>) in Sweden; Finnish (Eurasian) forest reindeer (<i>Rangifer tarandus</i>) in Finland. Roe	Eurasi	ærvids are inc¦ an/European €	luded in tl elk) (<i>Alces</i>	ne table. <i>alces alces</i>). Fal	low deer (I	Jama dama).	Reindeer:	Eurasian tur	dra reindeer (Rangifer t	arandus tarar.	i <i>dus</i>) in Swe	den: Finnish ((Eurasian)	forest rei	ndeer (<i>Rangif</i> e	er tarandus	fennicus) in	Finland.

Country	National case ID	Management system	Species	Sex	Age group	Target group	Part sampled	Analytical method type	Analytical method	Result
EU										
E	E20221025-100/ HVCW-150	Wild deer	European moose (as animal)	Female	≥12 months	FC	Brain (as part-nature)	Confirmatory	Western blot	POS
Non-EU										
ON	F	Wild deer	European moose (as animal)	Female	≥12 months	FC	Obex (as part-nature)	Screening	Bio-Rad TeSeE SAP rapid test (using the CWD protocol)	POS
		Wild deer	Reindeer (as animal)	Female	≥12 months	HSHC	Obex (as part-nature)	Screening	Bio-Rad TeSeE SAP rapid test (using the CWD protocol)	POS
							Retropharyngeal lymph node (as part-nature)	Screening	Bio-Rad TeSeE SAP rapid test (using the CWD protocol)	POS
	2	Wild deer	European moose (as animal)	Female	≥12 months	FC	Obex (as part-nature)	Screening	Bio-Rad TeSeE SAP rapid test (using the CWD protocol)	POS
	4	Wild deer	Red Deer (as animal)	Female	≥12 months	ĥ	Obex (as part-nature)	Screening	Bio-Rad TeSeE SAP rapid test (using the CWD protocol)	POS

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 TABLE 40
 Description of the CWD cases in 2022.

Only single MS (Finland) reported results of samples tested for TSE in species other than cattle, domestic sheep and goats and cervids. In total, 154 samples were collected from 54 American minks (*Neovison vison*), 12 raccoon dogs (*Nyctereutes procyonoides*), 34 foxes (genus *Vulpes*) and 54 domestic cats (*Felis catus*). None of them tested positive.

4 | CONCLUSIONS

As part of the BSE surveillance system in cattle in the EU, the EU27 and XI tested 977,008 cattle in 2022, 4.3% less than in the previous year. This drop in numbers largely resulted from reduced testing in Bulgaria, France, Germany and Italy. Nevertheless, the overall testing throughput combined with a risk-based strategy (84% of all tests were targeting risk animals) contributed to maximise the sensitivity of the BSE surveillance system considering the EU27 and XI as a single epidemiological unit. In 2022 in the EU27 and XI, only one atypical H-BSE case was confirmed by France. The situation of BSE was similar to the one of previous years in terms of tested animals and caseload. Considering the low annual incidence of H-type and L-type BSE cases, this single case found was in line with the expected variability of rare events. In total, 52,395 cattle were tested by eight other non-EU reporting countries, with no additional cases reported.

In total, 404,189 small ruminants were tested in 2022 in the EU27 and XI, as part of the TSE surveillance system, leading to an overall testing of more than 10.8 million tests since 2002. Twenty-four countries in the group EU27 complied with the EU monitoring requirements for sheep and 24 countries for goats.

Compared with 2021, there was an increase in the detection of the ovine CS IC (from 80 to 93) and a decrease in the detection of AS IC (from 96 to 76). The increase in CS IC occurred despite 3.6% decrease in the level of testing in non-infected flocks. As in 2021, this is in contrast to the trend in previous years, when a continuous decrease in the overall incidence of the disease (new infected herds) in this species was noted. Yet again, this could be due to normal variability between years. Over the same period, the number of caprine IC (CS and AS combined) increased by 42.9%, from 35 to 50, despite the lower number of animals tested in non-infected herds.

For CS in sheep in 2022 in the EU27 and XI and compared with 2021, the caseload increased by 13.8% despite a decrease in testing in TSE-infected flocks by 27.4%. It is worth emphasising the twofold increase in the number of CS cases reported by Greece, compared to the previous year, followed by increases of Romania by 35.3% and Italy by 14.9%, while Spain decreased the caseload by 36.4%. CS is reported by five MS, with four of them, Greece, Italy, Romania and Spain, accounting for more than 99% of all CS cases.

In goats, the total number of cases (224) remained unchanged compared to 2021. Of these, 216 scrapie cases reported in the EU-27 and XI in 2022 were CS (96.4%). They were reported by six MS. Cyprus, that experienced a steady decline in the incidence of CS over the last 10 years, remained stable in 2002 with 134 cases (4 IC) compared to 135 (5 IC) in 2021.

When looking at the long-term trends of CS in terms of cases per 10,000 tests, the situation up to 2022 confirmed the 10-year statistically significant decrease in sheep and no detectable trend in goats, respectively, as estimated by modelling of the available epidemiological data. In the coming years, caprine epidemiological trends may see improvements with the implementation of risk management strategies focused on breeding for resistance. This is now achievable following the recent amendment of the TSE Regulation.

With regard to AS in sheep in EU27 and XI, compared with 2021, testing activity resulted in a 25.2% decrease in reported cases, from 103 in 2021 to 77 in 2022, which is consistent with the decrease of the number of cases per 10,000 tested animals (active surveillance) (from 3.2 to 2.6). In goats, the AS situation was similar to the previous year in terms of caseload (5 in 2021 and 8 in 2022) while the proportion of cases per 10,000 tested animals and number of IC increased from 0.5 to 0.8. With regard to the long-term trends of AS, there was a 10-year statistically significant decreasing trend in sheep and no detectable trend in goats.

The genotyping data collected in 2022 from ovine CS cases consistently confirmed the association between the occurrence of the disease in animals with susceptible genotypes (NSP3, NSP3O, NSP4 or NSP5): 97.3% of the cases with known NSP genotype. The 2022 genotyping of random samples of the EU sheep population (data from seven MS after excluding Cyprus) showed a marginal improvement (7.3% of the genotyped sheep with known genotype carrying those of the susceptible groups) compared to the previous years (7.9%). The NSP1 group (i.e. ARR/ARR) accounted for 71.2% of all genotyped sheep. However, some caution is needed in interpreting this result as it could reflect the small number of MS contributing with data. Countries in which the caseload is large, like Italy, still showed a high proportion of susceptible sheep.

With regard to genotyping of goat cases, one polymorphism at codons 146 or 222 was reported from 184 cases by four reporting countries in the EU27 and XI. In Cyprus, two cases of CS were reported in goats heterozygous at codon 146: DQ-NQ and DZ/NZ.

Some caution is needed when interpreting the CWD surveillance data. In 2022, there was a substantial reduction of testing in both EU MS and Norway. The monitoring was mainly carried out on the category 'hunted/killed animals fit for human consumption' (78.9%), a target group with lower probability of disease, compared to the risk groups (FC, HSNHC, SUS) (EFSA BIOHAZ Panel et al., 2023). Ten MS tested 3202 cervids, compared to 5854 tested by eight MS in 2021 (–45.3%). This activity resulted in one case in Finland in a female European wild moose. Norway continued its surveillance programme in wild and captive cervids and tested 17,583 cervids in 2022, 4087 less than in 2021 (–18.9%). Four cases were confirmed by Norway in wild animals: two European moose, one reindeer and one red deer.

ABBREVIATIONS

ABBRE	EVIATIONS
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
DCF	Data Collection Framework
DWH	Data Warehouse
EFTA	European Free Trade Association
EM	Eradication measures
ES	Emergency slaughtered
EUSR	European Union summary report
FC	Fallen/culled
FS	Fallen stock
H-BSE	H-type bovine spongiform encephalopathy
HS	Healthy slaughtered
HSHC	Hunted/slaughtered fit for human consumption
HSNHC	· · · · · · · · · · · · · · · · · · ·
IC	Index case/s
IPA	Instrument for Pre-Accession Countries
L-BSE	L-type bovine spongiform encephalopathy
MS	Member State(s)
NSHC	Not slaughtered for human consumption
NSP	National Scrapie Plan
NUTS	Nomenclature of Units for Territorial Statistics
RK	Road/predator killed
RR	Relative risk
SHC	Slaughtered for human consumption
SU	Clinical suspect
SUS	Clinical suspect (cervids)

TSE Transmissible spongiform encephalopathies

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CONFLICT OF INTEREST

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COUNTRY CODES

Austria	AT	Finland	FI	Lithuania	LT	Romania	RO
Bosnia and Herzegovina	BA	France	FR	Luxembourg	LU	Serbia	RS
Belgium	BE	Germany	DE	Malta	MT	Slovakia	SK
Bulgaria	BG	Greece	EL	Montenegro	ME	Slovenia	SI
Croatia	HR	Hungary	HU	The Netherlands	NL	Spain	ES
Cyprus	CY	Iceland	IS	North Macedonia	МК	Sweden	SE
Czechia	CZ	Ireland	IE	Norway	NO	Switzerland	CH
Denmark	DK	Italy	IT	Poland	PL	Türkiye	TR
Estonia	EE	Latvia	LV	Portugal	PT	United Kingdom (in respect of Northern Ireland)	XI
United Kingdom (excluding Nor	thern Irelar	id)	XU				
EU27 MS countries and United H PL; PT; RO; SK; SI; ES; SE, XI. Non-EU reporting countries: BA	•	-			DK; EE; FI; F	⁻ R; DE; EL; HU; IE; IT; LV; LT; LU; MT	; NL;
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APPENDIX A

Additional surveillance data

TABLE A.1 BSE active monitoring in relation to the adult bovine population (age > 2 years) in 2022.

EU/non-EU groups	Country code	Adult cattle (>2 years) ^a	Number of tested bovine animals at risk ^b	Proportion (%) of tested bovine animals at risk ^b
EU	AT	833,950	18,461	2.2%
	BE	1,138,290	23,823	2.1%
	BG	402,250	1206	0.3%
	СҮ	41,940	1958	4.7%
	CZ	669,430	23,945	3.6%
	DE	5,089,050	164,660	3.2%
	DK	675,000	24,475	3.6%
	EE	131,550	3033	2.3%
	EL	318,900	1959	0.6%
	ES	3,181,440	59,135	1.9%
	FI	335,900	9817	2.9%
	FR	9,126,550	176,657	1.9%
	HR	161,000	4800	3.0%
	HU	477,300	11,883	2.5%
	IE	2,746,130	59,737	2.2%
	IT	2,966,000	41,966	1.4%
	LT	339,190	3998	1.2%
	LU	98,030	2668	2.7%
	LV	218,520	3432	1.6%
	МТ	6890	235	3.4%
	NL	1,719,000	57,381	3.3%
	PL	2,821,170	46,207	1.6%
	РТ	862,440	17,632	2.0%
	RO	1,259,200	14,131	1.1%
	SE	596,700	7646	1.3%
	SI	185,940	6378	3.4%
	SK	225,440	8613	3.8%
	Total EU27	36,627,200	795,836	2.2%
	XI ^c	746,000	24,546	3.3%
	Total EU27 + XI	37,373,200	820,382	2.24%
Other non-EU	BA	N/A ^e		
	СН	768,560	10,052	
	IS	37,490	58	0.2%
	ME	35,914	1	0.0%
	МК	118,000		0.0%
	NO ^d	353,700	6921	2.0%
	RS	423,580	3774	0.9%
	TR	8,371,060	8	
	Total other non-EU	10,108,304	20,814	0.21%
	TOTAL	47,481,504	841,196	1.77%

^aPopulation data obtained at: https://ec.europa.eu/eurostat/databrowser/view/APRO_MT_LSCATL__custom_6807810/default/table?lang=en

^bAt risk animals is the sum of animals with clinical signs at *ante mortem*, emergency slaughtered and fallen stock.

^cData from XI, United Kingdom (in respect of Northern Ireland) are available from 2021 onwards.

^dNorway's cattle population taken form the TSE EUSR report 2020 (EFSA et al., 2021).

^eN/A: not available.

APPENDIX B

Geographical distribution of BSE in the period 2001–2022

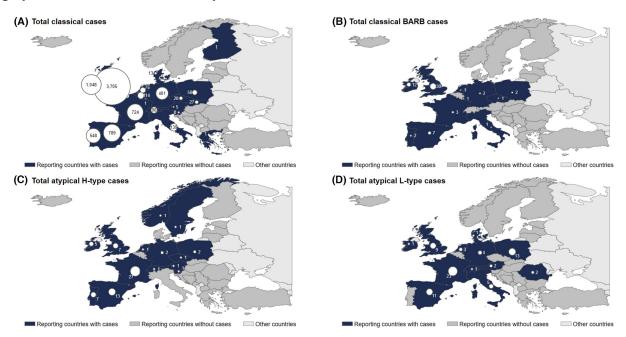


FIGURE B.1 Geographical distribution of cumulative number of cases of C-BSE (BARB cases not included) (A), BARB cases (B), H-BSE (C) and L-BSE (D) in the period 2001–2022. The size of the circles is proportional to the measurements and only comparable within the map but not between maps. With regard to the United Kingdom and 2022, only data from United Kingdom (in respect of Northern Ireland) (XI) have been considered.

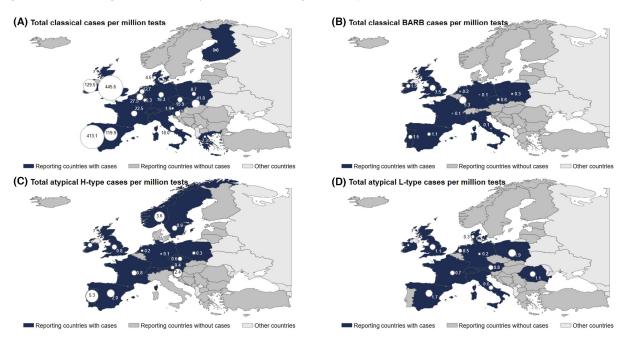


FIGURE B.2 Country-specific BSE cases per million tests by case type in the period 2001–2022 in the EU. The size of the circles is proportional to the measurements and only comparable within the map but not between maps. With regard to the United Kingdom and 2022, only data from United Kingdom (in respect of Northern Ireland) (XI) have been considered.

APPENDIX C

(A) Classical cases in sheep (B) Classical cases in goats France France Reporting countries with c Other countrie Reporting countries with cases Other countri (C) Atypical cases in sheep (D) Atypical cases in goats Con al Reporting countries without cases Other countries Reporting countries with cases Other countries Reporting countries with cases Reporting countries without cases

Geographical distribution of scrapie in 2022

FIGURE C.1 Geographical distribution of numbers of cases of ovine CS (A), caprine CS (B), ovine AS (C) and caprine AS (D) in 2022 in the reporting countries. The size of the circles is proportional to the measurements and only comparable within the map but not between maps.

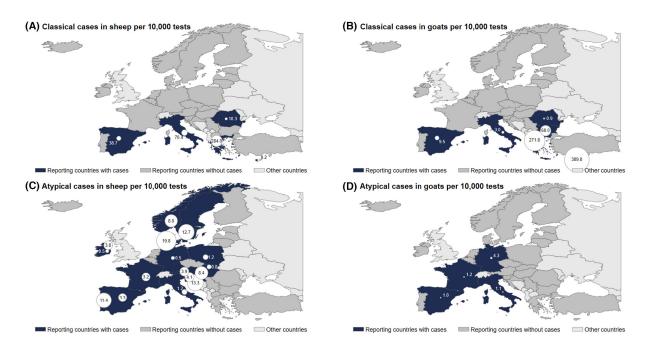


FIGURE C.2 Geographical distribution of proportion of cases per 10,000 tests of ovine CS (A), caprine CS (B), ovine AS (C) and caprine AS (D) in 2022. The size of the circles is proportional to the measurements and only comparable within the map but not between maps.

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APPENDIX D

Additional information, according to Annex III of Regulation 999/2001

TABLE D.1The number of suspected cases placed under official movementrestrictions in accordance with Article 12(1) in 2022.

Country	Cattle	Sheep	Goats
AT	8	1	0
BE	0	0	0
BG	0	0	3
СҮ	0	6446	8434
CZ	0	0	0
DE	0	1	1
DK	0	0	0
EE	0	0	0
EL	20	14	3
ES	1	9	1
FI	0	0	0
FR	2	1	0
HR	0	12	0
HU	10	0	0
IE	5	0	0
IT	0	0	0
LT	0	0	0
LU	3	0	0
LV	6	2	0
МТ	0	0	0
NL	0	0	0
PL	13	2	0
РТ	0	0	0
RO	57	52	4
SE	0	0	1
SI	0	1	0
SK	0	0	0
Total EU27	125	6541	8447
XI ^a	0	0	0
Total EU27 + XI	125	6541	8447
BA	3653	0	0
СН	22	0	0
IS	0	0	0
ME	0	0	0
МК	n/a	n/a	n/a
NO	0	0	0
RS	8	0	0
TR	0	0	0
Total other non-EU	3683	0	0
Total	3808	6541	8447

Abbreviations: n/a, not available.

^aData from XI, United Kingdom (in respect of Northern Ireland) are available from 2021 onwards.

TABLE D.2Number of flocks where suspected casesin ovine and caprine animals have been reported andinvestigated pursuant to Article 12(1) and (2) in 2022.

Country	Sheep	Goats
AT	1	0
BE	0	0
BG	0	3
CY ^a	2	16
cz	0	0
DE	27	8
DK	1	0
EE	0	0
EL	40	22
ES	0	0
FI	0	0
FR	14	3
HR	1	0
HU	0	0
IE	0	0
п	0	0
LT	0	0
LU	0	0
LV	2	0
МТ	0	0
NL	0	0
PL	3	0
РТ	0	0
RO	17	4
SE	0	1
SI	1	0
SK	0	0
Total EU27	109	57
XI ^b	0	0
Total EU27 + XI	109	57
ВА	0	0
СН	0	0
IS	0	0
ME	0	0
МК	n/a	n/a
NO	0	0
RS	0	0
TR	0	0
Total other non-EU	0	0
Total	109	57

Abbreviations: n/a, not available.

^aIn addition, 26 mixed flocks (sheep and goats)

^bData from XI, United Kingdom (in respect of Northern Ireland) are available from 2021 onwards.

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 in 2022.

Country	Sheep SHC	Sheep NSCH	Sheep EM	Goats SHC	Goats NSHC	Goats EM	Other ^a
AT	97	1625	0	25	482	0	0
BE ^b	0	n/a	0	0	n/a	0	0
BG ^c	n/a	n/a	n/a	n/a	n/a	n/a	0
CY ^d	0	548	0	0	363	0	76
cz	0	725	0	0	240	0	0
DE	4834	4398	0	80	782	0	60
DK ^e	n/a	n/a	n/a	n/a	n/a	n/a	0
EE	0	48	0	0	1	0	0
EL	151	503	22	41	228	10	0
ES	1043	3513	37	866	1549	9	0
FI	4	531	0	0	72	0	0
FR	1	12	0	0	3	0	0
HR	0	1035	0	0	255	0	n/a
HU	1579	1816	0	39	68	0	0
IE	3907	6627	0	0	49	0	0
п	4136	5242	42	3535	3714	11	0
LT	0	122	0	0	19	0	0
LU ^f	0	112	0	0	100	0	0
LV	0	43	0	0	22	0	0
МТ	1	115	0	0	59	0	0
NL	0	1584	0	0	1698	0	0
PL	3153	3616	0	259	1543	0	0
РТ	1121	5509	0	0	796	0	0
RO	1147	48	0	5	8	0	0
SE	0	1262	0	0	82	0	0
SI	48	1193	0	26	364	0	0
SK	0	475	0	0	78	0	0
ХI ^g	308	748	0	0	6	0	0
BA	0	0	0	0	0	0	0
СН	0	0	0	0	0	0	0
IS	410	52	0	0	0	0	0
ME	0	0	0	0	0	0	0
МК	0	0	0	0	0	0	0
NO	3705	4306	7	15	227	0	0
RS	8	88	0	1	10	0	0
TR	2	0	0	1	0	0	0

Abbreviations: EM, emergency slaughter; NSHC, not slaughtered for human consumption; SHC, slaughtered for human consumption; n/a, Not available.

^aMonitoring in other ovine and caprine categories (= 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).

^bThe 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. At the rendering plant sheep and goats are randomly sampled during the year.

^cThe Bulgarian competent authority informed that no data were available for sheep and goats.

^dThe Cypriot competent authority informed that in the category 'Other', the total number of animals born or derived from TSE infected holdings is 24,001 ovine and 30,386 caprine animals out of 104 holdings under restrictions (32 caprine, 8 ovine, mixed 64).

^eThe Danish 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.

^fThe Luxembourg competent authority informed that, for NSHC sheep and NSHC goats, the number of flocks is unknown.

^gData from XI, United Kingdom (in respect of Northern Ireland) are available from 2021 onwards.

APPENDIX E

Country data sets

All country data sets containing the tables on the occurrence of TSE per country are available on the EFSA Knowledge Junction community on ZENODO. Please see below the list and corresponding links to the data sets. The countries that submitted data sets on the 2022 monitoring data year are the 27 EU Member States, XI and eight other non-EU reporting countries.

Country	Link to the data set
EU27 Memb	
AT	
BE	https://doi.org/10.5281/zenodo.4091585
BG	https://doi.org/10.5281/zenodo.4091591 https://doi.org/10.5281/zenodo.4091602
СҮ	https://doi.org/10.5281/zenodo.4091602
cz	https://doi.org/10.5281/zenodo.4091616
DE	https://doi.org/10.5281/zenodo.4091621
DK	https://doi.org/10.5281/zenodo.4091627
EE	https://doi.org/10.5281/zenodo.4091635
EL	https://doi.org/10.5281/zenodo.4091641
ES	https://doi.org/10.5281/zenodo.4091643
FI	https://doi.org/10.5281/zenodo.4091647
FR	https://doi.org/10.5281/zenodo.4091649
HR	https://doi.org/10.5281/zenodo.4091653
HU	https://doi.org/10.5281/zenodo.4091655
IE	https://doi.org/10.5281/zenodo.4091659
ІТ	https://doi.org/10.5281/zenodo.4091663
LV	https://doi.org/10.5281/zenodo.4091665
LU	https://doi.org/10.5281/zenodo.4091671
LT	https://doi.org/10.5281/zenodo.4091673
мт	https://doi.org/10.5281/zenodo.4091678
NL	https://doi.org/10.5281/zenodo.4091684
PL	https://doi.org/10.5281/zenodo.4091686
РТ	https://doi.org/10.5281/zenodo.4091688
RO	https://doi.org/10.5281/zenodo.4091691
SE	https://doi.org/10.5281/zenodo.4091703
SI	https://doi.org/10.5281/zenodo.4091693
SK	https://doi.org/10.5281/zenodo.4091709
XI	https://doi.org/10.5281/zenodo.7257656
Other non-l	EU
BA	https://doi.org/10.5281/zenodo.5652824
СН	https://doi.org/10.5281/zenodo.4091716
IS	https://doi.org/10.5281/zenodo.4091719
ME	https://doi.org/10.5281/zenodo.4091723
МК	https://doi.org/10.5281/zenodo.4091725
NO	https://doi.org/10.5281/zenodo.4091727
RS	https://doi.org/10.5281/zenodo.4091729



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