The European Union summary report on surveillance for the presence of transmissible spongiform encephalopathies (TSE) in 2021

European Food Safety Authority (EFSA)

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 2021 by 27 Member States (MS, EU27), the United Kingdom (in respect of Northern Ireland) (XI), and eight other non-EU reporting countries: Bosnia and Herzegovina, Iceland, Montenegro, North Macedonia, Norway, Serbia, Switzerland and Turkey. In total, 1,021,252 cattle were tested by EU27 and XI (−9%, compared with 2020 when data from the United Kingdom were not restricted to Northern Ireland), and 66,121 cattle by eight non-EU reporting countries, with two cases of H-BSE in France and Spain, and four L-BSE in France (2), Germany and Spain. In total, 311,174 sheep and 118,457 goats were tested in the EU27 and XI (−6.4% and −1.8%, respectively, compared to 2020 when data from the whole United Kingdom were considered). In sheep, 551 cases of scrapie were reported by 17 MS and XI: 448 classical scrapie (CS) by six MS [80 index cases (IC) with genotypes of susceptible groups in 97% of the cases], 103 atypical scrapie (AS) (96 IC) by 13 MS and XI. In the other non-EU reporting countries, 27,594 sheep were tested with 55 CS and 1 AS in Iceland and 8 AS in Norway. Ovine random genotyping was reported by nine MS and genotypes of susceptible groups accounted for 7.9%. In goats, 224 cases of scrapie were reported by six EU MS: 219 CS (30 IC) by six MS, and five AS (5 IC) by three MS. In total, 5,854 cervids were tested for chronic wasting disease by eight MS; all resulted negative. Norway tested 21,670 cervids with two moose and one red deer positive. In total, 149 animals from four other species tested negative in Finland and Turkey.

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

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Summary

This report of the European Food Safety Authority (EFSA) presents the detailed results of surveillance activities on animal transmissible spongiform encephalopathies (TSE) carried out during 2021 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, i.e. Bosnia and Herzegovina, Iceland, Montenegro, North Macedonia, Norway, Serbia, Switzerland and Turkey, as well as genotyping data in sheep and goats. Turkey submitted data for the first time and Albania confirmed no surveillance on TSE was conducted in 2021.

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 mainly submitted through the EFSA TSE data reporting tool. Six 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), whereas the rest of the reporting countries submitted XML files by the EFSA TSE data collection tool. The electronically submitted data were extracted from the EFSA database and further processed and validated to summarise the information and to draft the summary tables presented in the current EU summary report (EUSR).

Despite the new arrangements for the data from the United Kingdom, the 2021 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 2021 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 Turkey) 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, 1,021,252 cattle were tested in 2021 in the EU27 and XI, a 9% reduction compared with the previous year, which cannot be considered a decrease given the throughput reported by the United Kingdom in 2020 (134,042). Over 85.2% 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 805,512 cattle tested in 2021 (92.6% of all cattle in the risk group). Additional 66,121 cattle were tested by the other eight non-EU reporting countries. Serbia (the main contributor with 19,578 cattle tested), Turkey (with 12,098) and Bosnia and Herzegovina (with 8,902) reported mostly cattle from the healthy slaughtered (HS) target group.

In the EU, six atypical cases of BSE were confirmed in 2021 by France (one H-BSE and two L-BSE), Germany (one L-BSE) and Spain (one H-BSE and one L-BSE), all born between 2006 and 2009 and referring to the FS (5) and ES (1) target groups. Four additional cases of BSE were reported in the rest of the world in 2021: two by Brazil, one by Canada and one by the United Kingdom. The case from the United Kingdom was a classic BSE case, detected in a homebred dairy cow born in February 2015, fallen stock cattle aged over 6.5 years (79 months) euthanised on farm (fallen stock).

In total, 429,631 small ruminants were tested in 2021 in the EU27 and XI: 311,174 sheep (a 6.4% decrease from 2020) and 118,457 goats (a 1.8% decrease), which cannot be considered a decrease given the throughput reported by the United Kingdom in 2020 (21,982 and 1,358, respectively). In addition, 27,594 sheep were tested by five of the eight other non-EU reporting countries, namely Iceland, North Macedonia, Norway, Serbia and Turkey, and 595 goats tested by Iceland, North Macedonia, Norway, Serbia and Turkey.

In sheep, 551 scrapie cases were reported in the EU27 and XI in 2021, 137 fewer cases than in 2020, due to the reduction in cases of Greece (−58.2%) and Spain (−25.8%). In total, 64 cases of scrapie in sheep were reported by two (Iceland and Norway) of the five other non-EU reporting countries that tested sheep. In total, 448 ovine cases in the EU27 and XI were AS (81.3%), 103 were AS (18.7%). Among the other non-EU reporting countries, 55 CS cases and one AS case were reported by Iceland and eight AS cases by Norway. Classical scrapie (CS) was reported by six MS, namely Bulgaria, Cyprus, Greece, Italy, Romania, Spain and one non-EU reporting country, Iceland. Atypical scrapie (AS) was reported by 13 MS (Croatia, Finland, France, Germany, Greece, Hungary, Ireland, Italy, Poland, Slovakia, Slovenia, Spain, Sweden), XI, and by two non-EU reporting countries (Iceland...
and Norway). A total of 13 cases in sheep were reported as inconclusive by Italy, which are not included in the total scrapie caseload for this country. Most of the ovine cases in the EU27 and XI (87.8%) were reported by five countries: Greece, Italy, Portugal, Romania and Spain, as it was the case in previous years.

In sheep, 176 (31.9%) of all cases in the EU27 and XI reported in 2021 were index cases (IC), with a much higher proportion in AS cases (93.2%) compared with CS cases (17.9%). In total, 98.6% of the CS cases in sheep reported in 2021 with known National Scrapie Plan (NSP) genotypes belonged to animals holding genotypes of the susceptible groups (NSP3, NSP3O, NSP4 or NSP5).

In 2021, the random genotyping of the national EU sheep populations was carried out by nine MS: Belgium, Cyprus (where genotyping is conducted systematically in the breeding sheep population), France, Germany, Greece, Italy, Latvia, the Netherlands and Poland. After excluding Cyprus, 7.9% of the randomly genotyped sheep still carried genotypes of the susceptible groups, lower than the 8.8% in 2020. The exception is Italy, with 21.2% of sheep with the susceptible genotypes and the highest case load in 2021.

In goats, in total, 224 scrapie cases were reported in the EU27 and XI: five were AS cases and 219 CS (97.8%, with Cyprus accounting for 60.3% of these). Compared with 2020, when 319 CS cases were reported, there was a 31.7% reduction (\( \frac{319 - 224}{319} \)) mainly due to the situation in Cyprus that has improved continuously over the last years. Six MS (Bulgaria, Cyprus, Greece, Italy, Romania and Spain) reported CS, whereas three MS (France, Italy and Spain) reported AS. The five other non-EU reporting countries (Iceland, North Macedonia, Norway, Serbia and Turkey) that reported tested goats did not report any scrapie cases. One additional scrapie case was reported as inconclusive by Italy, which is not included in the previous total scrapie caseload.

In goats, 15.6% (35) of all cases reported in the EU27 and XI in 2021 was IC, down from 2020 (18.6%), with a higher proportion in AS (100%) than in CS (13.7%). For the first time, data on genotyping of goat cases were reported. At least one polymorphism in codons 146 or 222 was reported from 152 cases (2 AS, 149 CS and 1 inconclusive) by Cyprus (115), Greece (11) and Italy (26). Of those, 48 CS cases (21.9%) and 2 AS cases (40%) were characterised both for the 146 and 222. No goat cases carried the alleles considered resistant.

With regard to the long-term trends (cases per 10,000 tests), the analysis up to 2021 confirmed the 10-year statistically significant decrease in sheep (5% annually for both CS and AS) and no detectable trends in goats for either CS or AS.

In 2021 5,854 cervids were tested by eight MS, 82.1% of them tested by Sweden and Romania, and with no cases reported. In 2021, the hunted/slaughtered fit for human consumption (HSHC) animals was the most frequently tested target group with 70.2% of all tested cervids. In 2021, Norway continued its intensified testing programme in wild and captive cervids and tested 21,670 animals, leading to the detection of three cases in wild animals (two moose and one red deer). Additionally, Iceland and Serbia reported testing of 3 and 185 cervids, respectively, which were all negative.

In total, 149 animals of other species were TSE tested by Finland and Turkey: 48 cats (Felis catus), 12 raccoon dogs (Nyctereutes procyonoides), 56 American minks (Neovison vison) and 33 foxes (genus Vulpes). None of them tested positive.

Two interactive communication tools on TSE – a story map (click this link), providing general information on TSEs, and three dashboards ( ), 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.
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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) 999/2001\(^1\) (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.

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

According to Part I.B, Chapter B of the same Annex III:

‘The compilation of reports containing the information referred to in Section A and submitted to the Commission (which shall send it to the European Food Safety Authority) on a monthly basis in the electronic format agreed between the Member States, the Commission and the European Food Safety Authority or, with regard to the information referred to in point 8 on a quarterly basis, may constitute the annual report as required by Article 6(4), provided that the information is updated whenever additional information becomes available.’\(^3\)

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

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

1.2.1. **Legal basis**

Animals suspected of 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

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\(^1\) Regulation (EC) No 999/2001 of the European Parliament and of the Council of 22 May 2001 laying down rules for the prevention, control and eradication of certain transmissible spongiform encephalopathies. OJ L 147, 31.5.2001, pp. 1–40.

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

\(^3\) Since 2018, TSE data are submitted by reporting countries directly to the European Food Safety Authority (EFSA) with different frequency and periodicity.
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/EC⁴ allowed MS to apply a revised BSE monitoring programme. Commission Implementing Decision 2013/76/EU⁵ 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 Decision (EU) 2016/851⁶, 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. 36/2005⁷] also provides for mandatory discriminatory testing for BSE of TSE cases detected in small ruminants.

Commission Regulation (EU) 2021/1176 of 16 July 2021 amended Annex III to Regulation (EC) No 999/2001 by requiring RCs to genotype prion protein for codons 146 and 222 for every positive TSE case in goats.

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 2021, 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 rules 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 2021 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 Turkey) are shown in Table 2.

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⁴ Commission Decision 2009/719/EC of 28 September 2009 authorising certain Member States to revise their annual BSE monitoring programmes. OJ L 256, 29.9.2009, pp. 35-37.
⁵ Commission Implementing Decision of 4 February 2013 amending Decision 2009/719/EC authorising certain Member States to revise their annual BSE monitoring programmes.
⁶ Commission Implementing Decision (EU) 2016/851 of 26 May 2016 amending the Annex to Decision 2009/719/EC as regards the authorisation for Croatia to revise its BSE annual monitoring programme. OJ L 141, 28.5.2016, pp. 131–132.
⁷ Commission Regulation (EC) No 36/2005 of 12 January 2005 amending Annexes III and X to Regulation (EC) No 999/2001 of the European Parliament and of the Council as regards epidemi-surveillance for transmissible spongiform encephalopathies in bovine, ovine and caprine animals. OJ L 10, 13.1.2005, pp. 9–17.
Table 1: Criteria for BSE surveillance in bovine animals as applied in 2021 by country, age limit and surveillance target group, based on the TSE Regulation (EC) as last amended, Commission Implementing Decision 2013/76/EU of 4 February 2013 and Commission Implementing Decision (EU) 2016/851 of 26 May 2016

| Surveillance target group | EU 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) |          |                          |

\(^{(a)}\): Different criteria were applied in 2021 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 2021 by reporting country and surveillance target group

| Country | Surveillance target group |
|---------|---------------------------|
|         | ES | AM | 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 |
| CY      | > 48 | > 48 | > 48 | No testing | No age limit | > 48 |
| CZ      | > 24 | > 24 | > 24 | No testing | No age limit | No age limit |
| DE      | > 48 | > 24 | > 48 | No testing | No age limit | No age limit |
| DK      | > 48 | > 48 | > 48 | No testing | No age limit | > 48 |
| EE      | > 48 | > 48 | > 48 | No testing | No age limit | No age limit |
| EL      | > 48 | > 48 | > 48 | > 72 | No age limit | No age limit |
| ES      | > 48 | > 48 | > 48 | Born before 2001 and coming from herds with BSE positive cases | No age limit | No age limit |
| FI      | > 48 | > 48 | > 48 | No testing | No age limit | No age limit |
| FR      | > 48 | > 48 | > 48 | Born before 1 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 |
| IT      | > 48 | > 48 | > 48 | No testing | No age limit | No age limit |
| LT      | > 48 | > 48 | > 48 | No testing | No age limit | No age limit |
| LU      | > 48 | > 48 | > 48 | No testing | No age limit | > 48 |
| LV      | > 48 | > 48 | > 48 | No testing | No age limit | No age limit |
| MT      | > 48 | > 48 | > 48 | No testing | No age limit | No age limit |
| NL      | > 48 | > 48 | > 48 | No testing | No age limit | No age limit |
| PL      | > 48 | > 48 | > 48 | No testing | No age limit | No age limit |
| PT      | > 48 | > 48 | > 48 | No testing | No age limit | No age limit |
| RO      | > 24 | > 24 | > 24 | > 30 | No age limit | No age limit |
| SE      | > 48 | > 48 | > 48 | No testing | No age limit | No age limit |
| SI      | > 48 | > 48 | > 48 | No testing | No age limit | No age limit |
| SK      | > 48 | > 48 | > 48 | No testing | No age limit | No age limit |
| XT\(^{(c)}\) | > 48 | > 48 | > 48 | No testing | No age limit | No age limit |
| BA      |          |          |          |              |          |          |
| CH      | > 48 | > 48 | > 48 | > 48 | No age limit | > 48 |

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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 TSE 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 2021 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 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) 2021/1176, 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 will no longer need to be conducted for a period of 2 years on holdings where an atypical scrapie case has been confirmed. A reduction in the number of tests on TSE-infected flocks is expected in the second half of the 2021 and subsequent years.

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.

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Table 3: Surveillance target groups in small ruminants to be reported for surveillance for TSE in 2021

| Country | Surveillance target group |
|---------|---------------------------|
| ES AM FS HS SU EM |
| IS | > 48 | > 48 | > 48 | No testing | No age limit | No age limit |
| ME | > 24 | > 24 | > 24 | > 30 | No age limit | No age limit |
| MK | > 24 | > 24 | > 24 | > 30 | No age limit | No age limit |
| NO | > 48 | > 48 | > 48 | No testing | No age limit | No age limit |
| 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 |

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.

The nine non-EU reporting countries (United Kingdom from 1 February 2020 and Turkey from 1 January 2021) are included in the table for information. The TSE Regulation does not apply to the nine non-EU reporting countries.

(a): If 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 01.01.2021), then the age limit is > 24 months.
(b): If 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 01.01.2021), then the age limit is > 30 months.
(c): Data from XI, i.e. United Kingdom (in respect of Northern Ireland), are available from 2021 onwards.

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

9 Commission 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.
As described in the 2018 EUSR on TSE (EFSA, 2019), the Commission Regulation (EC) 2017/894\textsuperscript{10} 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 for MS with an adult sheep population of \(> 750,000\) animals, and for other MS, to genotype a minimum sample of at least 100 animals. The new requirements establish to genotype once every 3 years with a minimum sample of at least 1,560 ovine animals; or at a frequency and with a sample size determined by the MS based on compliance with a set of criteria.

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’.

\textsuperscript{10} Commission Regulation (EU) 2017/894 of 24 May 2017 amending Annexes III and VII to Regulation (EC) No 999/2001 of the European Parliament and of the Council as regards the genotyping of ovine animals. OJ L 138, 25.5.2017, pp. 117–119.
Table 3: Target surveillance groups in small ruminants to be reported for surveillance for TSE based on the infection status of flock/herd/holding, the case type detected and the control measures taken according to the TSE Regulation

| Reported flock/herd status | Index case | Case type | Control measures taken | Sampled population | Surveillance target group to be reported |
|---------------------------|------------|-----------|------------------------|---------------------|-----------------------------------------|
| Non-infected flock/herd(b) | Yes        | CS or AS  | n/a                    | Slaughtered for human consumption. Annex III, Chapter A, Part II, point 2 | SHC                      |
|                           |            |           |                        | Not slaughtered for human consumption. Annex III, Chapter A, Part II, point 3 | NSHC                     |
|                           |            |           |                        | TSE suspects        | SU                                      |
| TSE infected flock/herd under official control at sampling(c) | No         | CS        | Killing and complete destruction of all animals (option 1), TSE Regulation, Annex VII, Chapter B, point 2.2.2(b) or killing and complete destruction of the susceptible animals only (option 2(o)) Annex VII, Chapter B, point 2.2.2(c) | Culled and destroyed under options 1 or 2 | EM                                      |
|                           |            |           |                        | Slaughtered for human consumption after application of option 1 or option 2(o) | SHC                      |
|                           |            |           |                        | TSE clinical suspects Chapter 4, Article 12, points 1 and 2 | SU                                      |
| TSE infected flock/herd under official control at sampling(c) | No         | CS        | Follow-up after implementation of control measures according to Annex VII, point 2. Intensified TSE monitoring protocol (Annex VII, point 3) after option 1 or option 2, or if derogation of option 2 was established, after complete destruction or slaughtering for human consumption of identified animals. | Slaughtered for human consumption point 3.1(a) | SHC                      |
|                           |            |           |                        | Not slaughtered for human consumption point 3.1(b) | NSHC                     |
|                           |            |           |                        | TSE clinical suspects Chapter 4, Article 12, points 1 and 2 | SU                                      |
| TSE infected flock/herd under official control at sampling(c) | No         | CS        | Follow-up after implementation of control measures according to Annex VII, point 2. Intensified TSE monitoring protocol (Annex VII, point 4) after option 3. | Slaughtered for human consumption point 4.1(a) | SHC                      |
|                           |            |           |                        | Not slaughtered for human consumption point 4.1(b) | NSHC                     |
|                           |            |           |                        | TSE clinical suspects Chapter 4, Article 12, points 1 and 2 | SU                                      |
| TSE infected flock/herd under official control sampling(c) | No         | CS        | Intensified TSE monitoring protocol pending the implementation of control measures according to the derogation in point 2.2.2(c)(iii) and after the implementation of the control measures | Slaughtered for human consumption. Points 4.1.(a) and 3.1.(a) | SHC                      |
|                           |            |           |                        | Not Slaughtered for human consumption. Points 4.1.(b) and 3.1.(b) | NSHC                     |
|                           |            |           |                        | TSE clinical suspects Chapter 4, Article 12, points 1 and 2 | SU                                      |
| Reported flock/herd status | Index case | Case type | Control measures taken                                                                                                                                                                                                 | Sampled population                                                                                           | Surveillance target group to be reported |
|----------------------------|------------|-----------|------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|-------------------------------------------------------------------------------------------------------------|----------------------------------------|
| TSE infected flock/herd under official control sampling\(^c\) | No         | AS        | Until July 2021, intensified TSE monitoring protocol after the detection of an Atypical Scrapie case (Annex VII point 2.2.3); the active monitoring (i.e. targeting SHC and NSHC) has been lifted on the basis of Commission Regulation (EU) 2021/1176 | Slaughtered for human consumption point 2.2.3 (discontinued from July 2021)                                | SHC                                    |
|                            |            |           |                                                                                                                                                                                                                       | Not slaughtered for human consumption point 2.2.3 (discontinued from July 2021)                            | NSHC                                   |
|                            |            |           |                                                                                                                                                                                                                       | TSE clinical suspects Chapter 4, Article 12, points 1 and 2                                              | SU                                     |

TSE: transmissible spongiform encephalopathy; CS: classical scrapie; AS: atypical scrapie; EM: animals culled under TSE eradication measures; NSHC: animals not slaughtered for human consumption; SHC: animals slaughtered for human consumption SU: animals clinically suspected of being infected by TSE.

(a): Option 2 can be applied both to sheep and goats (genotyping and culling).
(b): Sheep flocks or goat herds that are not under control measures or intensified TSE protocols or a sheep flock or goat herd that has never been infected with scrapie and for which every new detected case will be an index case.
(c): Sheep flocks or goat herds that are under control measures or intensified TSE protocols or a sheep flock or goat herd that has been infected during reporting year.
### Table 4: Minimum sample size for the TSE surveillance in small ruminants by reporting country in 2021

| Country | Sheep Population size\(^{(a)}\) | Surveillance target group | Goats Population size\(^{(a)}\) | Surveillance target group |
|---------|---------------------------------|---------------------------|-------------------------------|---------------------------|
|         | SHC | NSHC | SHC | NSHC | SHC | NSHC |
| AT      | 100–750 | 0 | 1,500 | 40–250 | 0 | 100% up to 500 |
| BE      | 100–750 | 0 | 1,500 | 40–250 | 0 | 100% up to 500 |
| BG      | > 750 | 10,000 | 10,000 | > 750 | 10,000 | 10,000 |
| CY      | 100–750 | 0 | 1,500 | 40–250 | 0 | 100% up to 500 |
| CZ      | 100–750 | 0 | 1,500 | < 40 | 0 | 100% up to 100 |
| DE      | > 750 | 10,000 | 10,000 | 40–250 | 0 | 100% up to 500 |
| DK      | 40–100 | 0 | 100% up to 500 | < 40 | 0 | 100% up to 100 |
| EE      | 40–100 | 0 | 100% up to 500 | < 40 | 0 | 100% up to 100 |
| EL      | > 750 | 10,000 | 10,000 | > 750 | 10,000 | 10,000 |
| ES      | > 750 | 10,000 | 10,000 | > 750 | 10,000 | 10,000 |
| FI      | 40–100 | 0 | 100% up to 500 | < 40 | 0 | 100% up to 100 |
| FR      | > 750 | 10,000 | 10,000 | > 750 | 10,000 | 10,000 |
| HR      | 100–750 | 0 | 1,500 | 40–250 | 0 | 100% up to 500 |
| HU      | > 750 | 10,000 | 10,000 | < 40 | 0 | 100% up to 100 |
| IE      | > 750 | 10,000 | 10,000 | < 40 | 0 | 100% up to 100 |
| IT      | > 750 | 10,000 | 10,000 | > 750 | 10,000 | 10,000 |
| LT      | 100–750 | 0 | 1,500 | < 40 | 0 | 100% up to 100 |
| LU      | < 40 | 0 | 100% up to 100 | < 40 | 0 | 100% up to 100 |
| LV      | 40–100 | 0 | 100% up to 500 | < 40 | 0 | 100% up to 100 |
| MT      | < 40 | 0 | 100% up to 100 | < 40 | 0 | 100% up to 100 |
| NL      | 100–750 | 0 | 1,500 | 250–750 | 0 | 1,500 |
| PL      | 100–750 | 0 | 1,500 | 40–250 | 0 | 100% up to 500 |
| PT      | > 750 | 10,000 | 10,000 | 250–750 | 0 | 1,500 |
| RO      | > 750 | 10,000 | 10,000 | > 750 | 10,000 | 10,000 |
| SE      | 100–750 | 0 | 1,500 | < 40 | 0 | 100% up to 100 |
| SI      | 40–100 | 0 | 100% up to 500 | < 40 | 0 | 100% up to 100 |
| SK      | 100–750 | 0 | 1,500 | < 40 | 0 | 100% up to 100 |
| XI\(^{(d)}\) | > 750 | 10,000 | 10,000 | 40–250 | 0 | 100% up to 500 |
| BA      | > 750 | 0 | 1,500 | 40–250 | 0 | 1,500 |
| CH      | – | – | – | – | – | – |
| IS      | 100–750 | < 40 | < 40 | < 40 | < 40 | < 40 |
| ME      | 100–750 | < 40 | < 40 | < 40 | < 40 | < 40 |
| MK      | 100–750 | 40–250 | 40–250 | 40–250 | 40–250 | 40–250 |
| NO      | > 750 | 0 | 1,500 | 40–250 | 0 | 1,500 |
| RS      | > 750 | 1,000\(^{(b)}\) | 1,000\(^{(b)}\) | 40–250 | 1,000\(^{(c)}\) | 1,000\(^{(c)}\) |
| TR      | > 750 | 0 | 1,500 | 40–250 | 0 | 1,500 |

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

The eight non-EU reporting countries (Turkey from 1 January 2021) are included in the table for information. The TSE Regulation does not apply to the eight non-EU reporting countries.

\(^{(a)}\): Thousand heads.
\(^{(b)}, (c)\): The 1,000 animals targeted are split between SHC and NSHC.
\(^{(d)}\): Data from XI – United Kingdom (in respect of Northern Ireland) are available from 2021 onwards.

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

Live sheep population in 2021 (or latest available) extracted from: [http://appsso.eurostat.ec.europa.eu/nui/show.do?dataset=apro_mt_lssheep&lang=en](http://appsso.eurostat.ec.europa.eu/nui/show.do?dataset=apro_mt_lssheep&lang=en) [Ewes and ewe-lambs put to the ram].
1.2.4. TSE surveillance in cervids and other species

By Commission Regulation (EU) 2017/1972\(^1\) requirements for a 3-year surveillance programme for chronic wasting disease (CWD) in cervids in Estonia, Finland, Latvia, Lithuania, Poland and Sweden were established for the period 2018–2020.

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

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, 2021).

2. Data and methods

2.1. Origin of the data

The 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 were mainly submitted through the EFSA TSE data reporting tool for the reporting of surveillance data on TSE as required by the TSE Regulation. The tool allows reporting countries to edit and automatically upload the data to the EFSA Data Collection Framework (DCF) for inclusion in the EFSA Scientific Data Warehouse (DWH). The tool has been used for the first time during the 2018 TSE data collection period. Six reporting countries (CZ, ES, FI, FR, IT and SE) transmitted data directly as eXtensible Markup Language (XML) files in 2021 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 collection 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 2021 data visualisation.

Finally, information on the population of bovines in 2021 were obtained from Eurostat annual data\(^1\) (Bovine animals, 2 years or over), while information on the population of small ruminants in 2021 as presented in Table 4 were obtained from the 2021 or latest available Eurostat annual data\(^1\). 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 from WOAH staff. During validation of the data with the reporting countries, additional information was asked with relation to the reporting according to (i) Annex III, Chapter B, Section 1.A, point 1 of the TSE Regulation: the number of suspected cases placed under official movement

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\(^1\) Commission Regulation (EU) 2017/1972 of 30 October 2017 amending Annexes I and III to Regulation (EC) No 999/2001 of the European Parliament and of the Council as regards a surveillance programme for chronic wasting disease in cervids in Estonia, Finland, Latvia, Lithuania, Poland and Sweden and repealing Commission Decision 2007/182/EC. OJ L 281, 31.10.2017, pp. 14–20.

\(^2\) https://appsso.eurostat.ec.europa.eu/nui/show.do?dataset=apro_mt_lscatl&lang=en.

\(^3\) http://appsso.eurostat.ec.europa.eu/nui/show.do?dataset=apro_mt_lssheep&lang=en.
restrictions in accordance with Article 12(1), per animal species; (ii) Annex III, Chapter B, Section 1.A, point 3 of the TSE Regulation: The number of flocks for which suspected cases in ovine and caprine animals have been reported and investigated pursuant to Article 12(1) and (2); and (iii) the number of ovine and caprine flocks tested within each subpopulation referred to in 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 2021 and 31 December 2021 in the EU (27 Member States and United Kingdom in respect of Northern Ireland, referred to in the report as ‘EU27 and XI’) and other eight additional non-EU reporting countries: Bosnia and Herzegovina, Iceland, Montenegro, North Macedonia, Norway, Serbia, Switzerland and Turkey. Turkey (non-EU and non-EFTA reporting country) submitted TSE data to EFSA for the first time and accepted their data to appear in the EU summary report. Albania did not contribute with TSE surveillance data in 2021.

EFSA validated the 2021 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 2022 and was finalised on 10 June 2022. The results and tables presented in the current EUSR are based on the data retrieved from the EFSA Scientific Data Warehouse on 13 July 2021. An additional consultation with reporting countries was conducted between 15 September 2022 and 5 October 2022. 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.

Historical data (data between 2001 and 2021 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 2021. 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 2021 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 Turkey). 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 1059/200314 (see section Country codes).

Since 1 January 2021 United Kingdom data are no longer submitted; they are partially replaced by XI (United Kingdom in respect of Northern Ireland) data. In the tables in 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.

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

14 Regulation (EC) No 1059/2003 of the European Parliament and of the Council of 26 May 2003 on the establishment of a common classification of territorial units for statistics (NUTS). OJ L 154 21.6.2003, p. 1.
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 story map (click this link), providing general information on TSEs, and three dashboards ( ), 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 2021, 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 were available for the period 2001–2021 for bovine animals, for 2002–2021 for small ruminants and for the 2018–2021 period for cervids and other species.

Despite the new provisions for the data from United Kingdom, the 2021 EU and XI data have been compared with data for the EU and the United Kingdom of previous years; 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 2021 by reporting country and surveillance target group. In addition, historical data on confirmed cases since 2017 are presented in detail whereas those on the 2001–2016 period have been summed up. This 5-year period has been selected as during the period 2017–2021 a harmonised EU-wide, active BSE surveillance was applied and restricted to at-risk animals of $\geq 48$ months of age, even though few exceptions are still in place in some countries as shown in Table 2.

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

To obtain relevant epidemiological information about the BSE cases detected in 2021, EFSA asked for additional information from the individual concerned reporting countries by a small questionnaire.

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 (2012–2021) 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.
An 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 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 non-infected 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 2021 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 2021.

A number of tested animals in species other than cattle, sheep, goats and cervids tested for TSE in reporting countries in 2021 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 2012–2021) 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. 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.

TSE data of small ruminants from the last 10 years (period 2012–2021) 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. The RR obtained by exponentiating the beta coefficient associated with the ‘year’ variable was used as a measurement of the annual variation in the probability of detection, i.e. the temporal trend for the entire period. In the model, the RR indicates the average annual change in the proportion of cases per animals tested corresponding with the annual probability of detecting the disease: 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 ≤ 0.05 was considered statistically significant for all the above-described statistical analyses.

3. Assessment

3.1. BSE surveillance in bovine animals

Since 2001 approximately 120.9 million bovine animals have been tested for BSE in the EU, including the United Kingdom until 2020 and XI instead in 2021. In 2021, there was a 9% reduction in the number of tested bovine animals in the EU27 and XI, from 1,122,671 in 2020 to 1,021,252 in 2021, which cannot be considered a real decrease given the throughput reported by the United Kingdom in 2020 (134,042). This reduction is mainly due to the exclusion of data from the United Kingdom (excluding Northern Ireland) (XU). If only EU27 data are considered, the number of tested animals presented a small increase of 0.8% since there was a decrease of 7.1% and 7.9% in the total number of cattle tested by Ireland and Poland, respectively, partially offset by the increase of 24.2% in the number of cattle tested in Romania and 13.5% in Hungary. Romania and Bulgaria are the main contributors to the HS testing group with 137,437 (91.5%) of all HS tested cattle in the EU27 and XI.

The other eight non-EU reporting countries (Bosnia and Herzegovina, Iceland, Montenegro, Norway, North Macedonia, Serbia, Switzerland and Turkey) tested 66,121 cattle in 2021. Serbia was the main contributor with 19,578 cattle tested (an increase of 27.7% compared to 2020), followed by Turkey, a country reporting data for the first time, that tested 12,098 cattle, all of them except seven in the HS target group. 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 993,190 in 2020 in EU and the United Kingdom to 870,328 in 2021 (−12.4%), in EU27 and XI. As stated above, this reduction is mainly due to the absence of data from XU (i.e. the United Kingdom excluding Northern Ireland). In previous years, the United Kingdom reported almost all cattle in the risk group. Similar to the previous year, cattle in the risk group accounted for 85.2% of all tested cattle in the EU27 and XI and cattle tested in the FS target group accounted for 92.6% of all risk cattle tested. The number of cattle tested for BSE per reporting country for each target group in 2021 is shown in Table 5.
Table 5: Number of bovine animals tested for BSE by reporting country and surveillance target group in 2021 in the EU and other reporting countries

| Country | Surveillance target group | Risk animals | Other target groups | Total |
|---------|---------------------------|--------------|---------------------|-------|
|         | FS | AM | ES | Subtotal risk animals | HS | EM | SU | Subtotal other target groups |       |
| AT      | 15,472 | 15 | 3,075 | 18,562 | 42 | 10 | 52 | 18,614 |
| BE      | 24,918 | 3 | 866 | 25,787 | 2 | 9 | 11 | 25,798 |
| BG      | 3,153 | 6 | 661 | 3,820 | 32,434 | 32,434 | 36,254 |
| CY      | 1,893 | 113 | 2,006 | 72 | 72 | 2,078 |
| CZ      | 20,884 | 3,956 | 24,840 | 31 | 1 | 32 | 24,872 |
| DE      | 166,223 | 10,806 | 177,029 | 368 | 599 | 967 | 177,996 |
| DK      | 21,534 | 1,336 | 22,870 |       |       |       | 22,870 |
| EE      | 3,140 | 88 | 164 | 3,392 |       |       |       | 3,392 |
| EL      | 2,029 | 1 | 2,030 | 8,759 | 12 | 8,771 | 10,801 |
| ES      | 61,492 | 34 | 1,117 | 62,643 | 285 | 7 | 4 | 296 | 62,939 |
| FI      | 9,552 | 3 | 9,555 |       |       |       |       | 9,555 |
| FR      | 194,021 | 1,734 | 195,755 | 2,445 | 1 | 2,446 | 198,201 |
| HR      | 5,240 | 5 | 13 | 5,258 | 107 | 107 | 5,365 |
| HU      | 12,116 | 83 | 78 | 12,277 | 13 | 13 | 26 | 12,303 |
| IE      | 59,121 | 299 | 59,420 | 43 | 6 | 49 | 59,469 |
| IT      | 32,759 | 221 | 14,630 | 47,610 | 239 |       | 47,849 |
| LT      | 3,717 | 1 | 14 | 3,732 |       |       |       | 3,732 |
| LU      | 2,766 |       | 2,766 | 2 | 2 | 2,768 |
| LV      | 3,157 | 80 | 136 | 3,373 | 14 | 14 | 3,387 |
| MT      | 119 |       | 213 |       |       |       |       | 213 |
| NL      | 54,091 | 6,808 | 60,899 | 4 | 4 | 60,903 |
| PL      | 40,538 | 1,095 | 6,926 | 48,559 | 149 | 7 | 156 | 48,715 |
| PT      | 16,127 | 1,083 | 1,718 | 18,928 | 6 | 6 | 18,934 |
| RO      | 3,674 | 3,111 | 3,352 | 10,137 | 105,003 | 45 | 105,048 | 115,185 |
| SE      | 8,377 | 14 | 159 | 8,550 | 14 | 14 | 8,564 |
| SI      | 5,964 | 63 | 397 | 6,424 | 21 | 17 | 38 | 6,462 |
| SK      | 9,101 | 5 | 9,106 | 2 | 2 | 9,108 |
| Total EU27 | 781,178 | 6,202 | 58,161 | 845,541 | 150,019 | 7 | 760 | 150,786 | 996,327 |
| XI(5) | 24,334 | 312 | 141 | 24,787 | 138 |       | 138 | 24,925 |
| Total EU27 + XI | 805,512 | 6,514 | 58,302 | 870,328 | 150,157 | 7 | 760 | 150,924 | 1,021,252 |
| BA      | 1 |       | 8,901 |       |       | 8,901 | 8,902 |
| CH      | 6,796 | 4,254 | 11,050 | 21 | 21 | 11,071 |
| IS      | 10 |       | 10 |       |       |       | 10 |
| ME      |       |       |       | 5,731 | 5,731 |       | 5,731 |
| MK      | 8 |       | 1,686 | 1,686 |       |       | 1,694 |
| NO      | 1,637 | 65 | 5,295 | 6,997 | 40 | 40 | 7,037 |
| RS      | 4,381 | 100 | 4,481 | 15,081 | 16 | 15,097 | 19,578 |
| TR      | 7 |       | 12,091 |       |       | 12,091 | 12,098 |
| Total other non-EU | 12,840 | 65 | 9,649 | 22,554 | 43,530 | 37 | 43,567 | 66,121 |
| Total | 818,352 | 6,579 | 67,951 | 892,882 | 193,687 | 7 | 797 | 194,491 | 1,087,373 |
The distribution of the number of bovine animals tested for BSE by age group, surveillance target group and reporting country in 2021 can be found in the following link https://doi.org/10.5281/zenodo.7282822 distributed as follows:

- **Table 6**: Number of bovine animals tested by age group in the EU27 and XI and non-EU reporting countries in 2021.
- **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 2021.
- **Table 8**: Number of tested healthy slaughtered bovine animals by age group in EU27 and XI and non-EU reporting countries in 2021.
- **Table 9**: Number of BSE suspected bovine animals, by age group, tested in EU27 and XI MS and non-EU-reporting countries in 2021.
- **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 2020. The table is empty because no animals in this category were tested for the year 2021.

In the EU27 and XI, six BSE cases were reported in 2021, all atypical BSE. Five of them were submitted to the FS testing group and one, in Germany, as emergency slaughtered (ES). Two were of the H-type (one from Spain and one from France) and four of the L-type (one from Germany, one from Spain and two from France). Table 11 reports the main clinical and epidemiological data of all the positive cases. Two L-type cases reported by France in the FS target group showed clinical signs before death. Four additional cases of BSE were reported in the rest of the world in 2021: two by Brazil, one by Canada and one by the United Kingdom (https://wahis.oie.int/#/home). The case from the United Kingdom was classic BSE, detected in a homebred dairy cow born in February 2015, aged 6.5 years (79 months) euthanised on farm (fallen stock). The other three cases were H-type BSE and were found in a 8.5-year-old symptomatic beef cow in Canada, and in a 10- and 11-year-old beef cows during emergency slaughter in Brazil.

In general, the number of H-type and L-type BSE cases 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 2016 to 2021. Most of the atypical cases reported in 2021 was detected in beef cattle older than 12 years (only in France, one L-type case cattle was younger than the rest, i.e. 150 months).

Based on 125 atypical BSE cases with known age since 2001, the average age at detection was 12.05 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.

The number of BSE cases by reporting country, type and year (up to 2021, 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 2012–2021) shows a significant decreasing trend in the occurrence of C-BSE (annual RR = 0.5, i.e. an annual decrease of 40% in the proportion of cases per tested animals; p < 0.0001), whereas no significant trend for the two atypical BSE forms was found (H-BSE: RR: 1.10 p = 0.18; L-BSE: RR:1.06 p = 0.4).

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 2001–2021 are shown in Appendix B.

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15 https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/1041282/Report_on_the_epidemiological_investigation_of_a_single_BSE_case_in_Somerset.pdf.
Table 11: Clinical and epidemiological description of the BSE cases detected in 2021

| Country | DE – atypical | ES – atypical | FR – atypical | FR – atypical | FR – atypical |
|---------|---------------|---------------|---------------|---------------|---------------|
| Surveillance target group | Emergency slaughtered | Fallen stock | Fallen stock | Fallen stock | Fallen stock |
| Case type | L-BSE | H-BSE | L-BSE | L-BSE | H-BSE |
| Month and year of birth | Mar-07 | Jan-07 | Apr-08 | Mar-09 | Mar-06 |
| Age at detection (in months) | 175 | 170 | 160 | 150 | 181 |
| BARB status | None | None | None | behavioural and locomotor disorders the day before the death of the animal | decubitus, left hemiparalysis |
| Clinical signs | None | None | None | Limousine | Aubrac |
| Cattle type | Suckler cow | Beef | Beef | Beef | Beef |
| Breed | Pinzgauer | Mixed | Mixed | Limousine | Charolais |
| Was the case confirmed at herd/holding where the animal was born? | Yes | No | Yes | Yes | Yes |
| Location (NUTS3) of natal herd or herd where case found | Mühldorf am Inn (Oberbayern) | Zahinos | Villar de Plasencia | Hautes-Pyrenees | Jura |
| Herd size | 56 | 26 | 88 | 123 | 79 |
| Herd type | Suckler herd | Beef | Beef | Beef | Beef |
| Feeding system during first year of life | Pasture, farm own compound feed | Mixed | Mixed | Mother’s milk fed | Mother’s milk fed |
| Feed cohorts? Tested? If Yes: Results (number tested; number positives) | 251 cattle; No Deferred sacrifice | No | Yes, 12 tested/all negative | Yes, 1 cattle tested and negative | No |
| Birth cohorts? Tested? If Yes: Results (number tested; number positives) | 121 cattle, 2 cattle tested, both negative | Deferred sacrifice | YES (2/0/2) | Yes, 12 tested/all negative | No |
| Offspring? Tested? If Yes: Results (number tested; number Positives) | 3 cattle, No Deferred sacrifice | YES (1/0/1) | Yes, 1 tested/negative | No, a single offspring (less than 2 years old) | No, 1 cattle waiting for slaughtering |
| Country | DE – atypical | ES – atypical | ES – atypical | FR – atypical | FR – atypical | FR – atypical |
|---------|---------------|---------------|---------------|---------------|---------------|---------------|
| Sire? Tested? (Yes/No). If Yes: Results (positive? Negative?) | Unknown | No | No | No | No | No |
| Dam? Tested (Yes/No). If Yes: Results (positive? Negative?) | DE 0936291579 Date of birth 11.03.2002, Slaughter 10.04.2010, yes, negative | No | No | No | No | No |

N/A: not available.
Table 12: Total number of reported BSE cases (classical-BSE + atypical H-BSE + atypical L-BSE) in reporting countries and worldwide by year (period 1991–2021) and country

| Country | Year | Total |
|---------|------|-------|
|         | Up to 2016 | 2017 | 2018 | 2019 | 2020 | 2021 |
| AT      | 8  | 8     |
| BE      | 133 | 133  |
| CZ      | 30 | 30    |
| DE<sup>a</sup> | 421 | 1 | 422 |
| DK<sup>a</sup> | 16 | 16 |
| EL      | 1  | 1     |
| ES      | 814 | 3 | 2 | 1 | 2 | 822 |
| FI      | 1  | 1     |
| FR<sup>a</sup> | 1,004 | 2 | 3 | 4 | 2 | 3 | 1,018 |
| IE<sup>a</sup> | 1,660 | 1 | 0 | 1 | 1,662 |
| IT<sup>a</sup> | 147 | 0 | 147 |
| LU      | 3  | 0     |
| NL      | 88 | 0 | 88 |
| PL      | 74 | 1 | 75 |
| PT<sup>a</sup> | 1,086 | 1,086 |
| RO      | 2  | 2     |
| SE<sup>b</sup> | 1 | 1 |
| SI<sup>a</sup> | 9 | 9 |
| SK      | 27 | 27    |
| Total EU27 | 5,525 | 6 | 3 | 7 | 4 | 6 | 5,551 |
| BRA     | 2  | 1 | 2 | 5 |
| CAN<sup>a</sup> | 20 | 1 | 21 |
| CH<sup>a</sup> | 465 | 1 | 466 |
| ISR     | 1  | 1     |
| JPN     | 36 | 36    |
| LI      | 2  | 2     |
| NO      | 1  | 1     |
| USA<sup>a</sup> | 4 | 1 | 1 | 6 |
| United Kingdom<sup>c</sup> | 184,594 | 1 | 184,595 |
| XU<sup>c</sup> | 1 | 1 |
| Total other non-EU | 185,125 | 7 | 5 | 8 | 5 | 10 | 190,685 |
| Total  | 190,650 | 7 | 5 | 8 | 5 | 10 | 190,650 |

BSE: bovine spongiform encephalopathy; H-BSE: H-type BSE; L-BSE: L-type BSE; BRA: Brazil; CAN: Canada; ISR: Israel; JPN: Japan; USA: The United States of America.

Each cell reports the total number of BSE cases (C-BSE + H-BSE + L-BSE). Grey-shaded cells indicate the year(s) and Member State where at least one BARB case was detected (EFSA BIOHAZ Panel, 2017a). EU countries without BSE cases (Bulgaria, Cyprus, Estonia, Croatia, Hungary, Lithuania, Latvia and Malta) are not included in the table.

(a): Included 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.

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

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

(c): United Kingdom was member of European Union until 2020. XI (United Kingdom in respect of Northern Ireland) and XU (the United Kingdom excluding Northern Ireland) are presented from thereon.
Table 13: Number of reported classical BSE cases in the EU and non-EU reporting countries by year (period 2001–2021) and country

| Country code | Year | Total |
|--------------|------|-------|
|              | Up to 2016 | 2017 | 2018 | 2019 | 2020 | 2021 |
| AT           | 5      |      |      |      |      |      |
| BE           | 133    |      |      |      |      |      |
| CZ           | 29     |      |      |      |      |      |
| DE           | 416    |      |      |      |      |      |
| DK           | 15     |      |      |      |      |      |
| EL           | 1      |      |      |      |      |      |
| ES           | 798    |      |      |      |      |      |
| FI           | 1      |      |      |      |      |      |
| FR           | 969    |      |      |      |      |      |
| IE           | 1,656  |      |      |      |      |      |
| IT           | 142    |      |      |      |      |      |
| LU           | 3      |      |      |      |      |      |
| NL           | 84     |      |      |      |      |      |
| PL           | 60     |      |      |      |      |      |
| PT           | 1,079  |      |      |      |      |      |
| SI           | 8      |      |      |      |      |      |
| SK           | 27     |      |      |      |      |      |
| Total EU27   | 5,426  | 0    | 0    | 0    | 0    | 0    |
| CH           | 464    |      |      |      |      |      |
| United Kingdom(a) | 184,578 | 1    |      |      |      |      |
| XU(a)        |        |      |      |      |      | 1    |
| Total other non-EU | 185,042 | 0    | 1    | 0    | 0    | 1    |
| Total        | 190,468| 0    | 1    | 0    | 0    | 1    |

BSE: bovine spongiform encephalopathy.
Each cell reports the total number of C-BSE cases.
Reporting countries that have never reported classical cases are not included in the table.
Grey-shaded cells indicate the year and Member State where at least one BARB case was detected (EFSA BIOHAZ Panel, 2017a).
Source: data were retrieved from the EU TSE Database and from the OIE website for CH.
(a): United 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 2001–2021), type and country

| Country | Year | Total |
|---------|------|-------|
|         | up to 2016 | 2017 | 2018 | 2019 | 2020 | 2021 |
| H       | L     | H     | L     | H     | L     | H     |
| AT      | 1     | 2     |      |      |      |      |
| CZ      | 1     |      |      |      |      |      |
| DE      | 2     | 3     |      |      |      |      |
| DK      | 1     |      |      |      |      |      |
| ES      | 8     | 8     | 1     | 2     | 2     | 1     | 1     | 1     | 13    | 11    |
| FR      | 18    | 17    | 1     | 1     | 4     | 1     | 1     | 1     | 2     | 26    | 23    |
| IE      | 4     | 1     |      |      |      |      |      | 5     | 1     |      |      |
| IT      | 5     |      |      |      |      |      |      | 0     | 5     |      |      |
| NL      | 1     | 3     |      |      |      |      |      | 1     | 3     |      |      |
| PL(a)   | 2     | 12    |      |      |      |      |      | 2     | 13    |      |      |
| PT      | 7     |      |      |      |      |      |      | 7     | 0     |      |      |
The number of historical reported BSE cases can be found in the following link [https://doi.org/10.5281/zenodo.7282881](https://doi.org/10.5281/zenodo.7282881), 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.

### Table 15

| Country | up to 2016 | 2017 | 2018 | 2019 | 2020 | 2021 | Total |
|---------|-----------|------|------|------|------|------|-------|
|         | H | L | H | L | H | L | H | L | H | L |
| RO      | 2 | 0 | 2 |     |     |     |     |     |     |     |
| SE      | 1 | 0 |    |    |     |     |     |     |     |     |
| SI      | 1 | 1 |    |    |     |     |     |     |     |     |
| Total EU27 | 46 | 53 | 2 | 4 | 1 | 2 | 6 | 1 | 3 | 2 | 4 | 60 | 65 |

### Table 16

| Country | 2012 | 2013 | 2014 | 2015 | 2016 | 2017 | 2018 | 2019 | 2020 | 2021 | Total |
|---------|------|------|------|------|------|------|------|------|------|------|-------|
| CH      | 1 | 0 |    |     |     |     |     |     |     |     | 1 |
| NO      | 1 |    |    |     |     |     |     |     |     |     | 1 |
| United Kingdom | 7 | 9 |    |    |    |    |    |    |    |    | 7 |

### Table 17

| Country | 2012 | 2013 | 2014 | 2015 | 2016 | 2017 | 2018 | 2019 | 2020 | 2021 | Total |
|---------|------|------|------|------|------|------|------|------|------|------|-------|
| CH      | 1 | 0 |    |     |     |     |     |     |     |     | 1 |
| NO      | 1 |    |    |     |     |     |     |     |     |     | 1 |
| United Kingdom | 7 | 9 |    |    |    |    |    |    |    |    | 7 |

### Table 18

| Country | 2012 | 2013 | 2014 | 2015 | 2016 | 2017 | 2018 | 2019 | 2020 | 2021 | Total |
|---------|------|------|------|------|------|------|------|------|------|------|-------|
| CH      | 1 | 0 |    |     |     |     |     |     |     |     | 1 |
| NO      | 1 |    |    |     |     |     |     |     |     |     | 1 |
| United Kingdom | 7 | 9 |    |    |    |    |    |    |    |    | 7 |

Each cell reports the total number of H-BSE and L-BSE cases. Reporting countries that have never reported atypical cases are not included in the table.

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

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

The number of historical reported BSE cases can be found in the following link [https://doi.org/10.5281/zenodo.7282881](https://doi.org/10.5281/zenodo.7282881), 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.
(a) C-BSE

(b) H-BSE

(c) L-BSE

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.

**Figure 1:** Cases per million tested bovine animals by surveillance target group and case type for the period 2016–2021 in the EU and the United Kingdom (until 2020) and in the EU and XI in 2021
3.2. TSE surveillance in small ruminants

Since 2002, more than 10.4 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 from 2021. In 2021, 429,631 small ruminants were tested by the EU27 and XI: 311,174 sheep (61.9%) and 118,457 goats (38.1%), which represents an overall 5.2% decrease (23,563) in the number of tested small ruminants, compared to 2020; however, it cannot be considered a real decrease given the throughput reported by the United Kingdom in 2020 (21,982 and 1,358 for sheep and goats, respectively).

In five of the eight non-EU reporting countries (Iceland, North Macedonia, Norway, Serbia, Turkey), a total of 28,289 small ruminants were tested: 27,594 sheep (97.8%) and 595 goats (2.2%), an increase of 1,524 (+5.7%) from 2020, mostly due to the increase in testing by Norway. Bosnia and Herzegovina, Montenegro and Switzerland did not report data on small ruminants. The new reporting country, Turkey, reported 29 small ruminants tested.

In sheep, the decrease in the total tested in the EU27 and XI was 6.4% (311,174 tested in 2021 compared with 332,579 in 2020), due to the decrease of testing in TSE-infected flocks with a 28.5% decrease in 2021 (from 29,094 in 2020 to 20,793 in 2021), and a 3.6% decrease in the non-TSE infected flocks (from 301,089 in 2020 to 290,381 in 2021). The absence of the data from the United Kingdom (excluding Northern Ireland) (XU) has also contributed to the absolute decrease in non-infected flocks, since in 2020, the United Kingdom reported 21,982 sheep tested, 21,548 of them in non-infected flocks.

In goats, there was a 1.8% decrease in the tested animals in the EU27 and XI (118,457 in 2021 compared with 120,615 in 2020), partially due to the absence of the data from the United Kingdom (excluding Northern Ireland) (XU). The testing in TSE-infected herds suffered a 23.8% decrease in 2021 (from 8,063 in 2020 to 6,145 in 2021), again partially due to the absence of XU data, whereas the testing in non-infected herds remained almost the same.

The numbers of sheep and goats tested for TSE by reporting country, surveillance target group and flock/herd status in 2021 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, 22 countries in the group EU27 and XI fulfilled the requirements for sheep testing. In goat surveillance, 23 countries in the group EU27 and XI fulfilled the requirements for goat testing.

In 2021, the pattern of ovine testing by country and flock status was different from that of 2020. For each sheep tested in a TSE-infected flock in the EU27 and XI, there were about 14 (13.96) sheep tested in non-TSE-infected flocks. The decrease in the overall testing of sheep is due to the balance between the exclusion of data from XU and the lower level of testing in Czechia, Bulgaria and Spain with an overall decrease of 526 (−22%), 6,675 (−17.7%), 4,353 (−11.8%) sheep tested, respectively, compensated partially by a higher level of testing in Hungary, Italy and Romania with an overall increase of 4,859 (+22.5%), 3,961 (+14.5%) and 4,298 (+13.7%) sheep tested, respectively.

Also, in goats, the 2021 pattern of testing by country and flock status was different from that of 2020. In 2021, for each goat tested in a TSE-infected herd in the EU27 and the XI, there were about 19.3 goats tested in non-TSE-infected herds. The decrease in the overall testing of goats is due to the balance between the exclusion of data from XU and the lower level of testing in Cyprus, Bulgaria and Italy of 1,942 (−41.4%), 642 (−18.4%) and 2,948 (−8.9%), respectively, compensated partially by the increase of tested goats in Romania of 2,397 (+11.9%), 367 in Germany (+18.6%) and 359 in the Netherlands (+22.9%).

Table 19: Number of sheep tested for TSE by reporting country, surveillance target group and flock status in 2021 in the EU and other reporting countries

| Flock status | TSE infected flocks | Non-infected flocks | Subtotal | Total |
|--------------|---------------------|---------------------|----------|-------|
| Surveillance Target group | EM | NSHC | SHC | SU | EM | NSHC | SHC | SU | Subtotal | Non-infected flocks | Subtotal | Non-infected flocks |
| AT | 2,809 | 115 | 3 | 2,927 | 2,927 |
| BE | 589 | 589 | 915 | 920 | 1,509 |
| BG | 3,868 | 27,203 | 25 | 31,096 | 31,096 |

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**Flock status**
- TSE infected flocks
- Non-infected flocks

| Surveillance Target group | EM | NSHC | SHC | SU | Subtotal TSE infected flocks | EM | NSHC | SHC | SU | Subtotal Non infected flocks | Total |
|---------------------------|----|------|-----|----|-------------------------------|----|------|-----|----|-------------------------------|-------|
| CY                        | 535| 362  | 897 |    | 1,626                        | 6  | 1,632 |    |    |                               | 2,529 |
| CZ                        | 1,870| 1,870|     |    |                               | 1,870|       |    |    |                               | 1,870 |
| DE                        | 24 | 24   | 10,652| 8,972| 41                            | 19,665| 19,689 |    |    |                               |       |
| DK                        | 455|      | 455  |    |                               | 455 |       |    |    |                               | 455   |
| EE                        | 185|      | 185  |    |                               | 185 |       |    |    |                               | 185   |
| EL                        | 653| 518  | 11   | 1,182| 1,924                        | 1,805| 2      | 3,731|    |                               | 4,913 |
| ES                        | 10,053| 10,053| 12,643| 9,717| 2                            | 22,362| 32,415 |    |    |                               |       |
| FI                        | 29 | 4    | 33   | 1,492| 6                            | 1,498| 1,531 |    |    |                               |       |
| FR                        | 20 | 20   | 17,124| 5,158| 4                            | 22,286| 22,306 |    |    |                               |       |
| HR                        | 1,497|     |      | 7   | 1,504                        | 1,504|       |    |    |                               |       |
| HU                        | 1,401| 256  | 1,657| 11,005| 13,768                       | 24,773| 26,430 |    |    |                               |       |
| IE                        | 24 | 24   | 48   | 10,712| 10,327                       | 21,039| 21,087 |    |    |                               |       |
| IT                        | 3,727| 637  | 392  | 4,756| 14,768                       | 11,817| 31,343 |    |    |                               |       |
| LT                        | 764|      |      | 764 |                               | 764 |       |    |    |                               |       |
| LU                        | 101|      |      | 101 |                               | 101 |       |    |    |                               |       |
| LV                        | 288|      |      | 288 |                               | 288 |       |    |    |                               |       |
| MT                        | 95 |      |      | 95 |                               | 95 |       |    |    |                               |       |
| NL                        | 1,888|     |      | 1,888| 1,888                        | 1,888|       |    |    |                               |       |
| PL                        | 10,142| 19,993|      | 30,135| 30,135                       | 30,135|       |    |    |                               |       |
| PT                        | 134| 17   | 151  | 15,452| 5,134                        | 20,586| 20,737 |    |    |                               |       |
| RO                        | 5  | 724  | 18   | 747 | 15,264                       | 19,564| 35,584 |    |    |                               |       |
| SE                        | 2  |      | 2    | 1,687| 1,687                        | 1,687|       |    |    |                               |       |
| SI                        | 2,548| 181  | 13   | 2,742| 2,742                        | 2,742|       |    |    |                               |       |
| SK                        | 37 | 597  | 634  | 12,189| 12,189                       | 12,189| 12,823 |    |    |                               |       |
| Total EU27                | 14,514| 4,471| 1,790| 20,793| 153,963                      | 133,766| 113   | 287,842| 308,635 |                               |       |
| XI(a)                     | 2,539|     |      | 2,539|                               | 2,539|       |    |    |                               |       |
| Total EU27 + XI           | 14,514| 4,471| 1,790| 20,793| 156,502                      | 133,766| 113   | 290,381| 311,174 |                               |       |
| BA                        | 0  |      |      | 0   |                               | 0   |       |    |    |                               |       |
| CH                        | 0  |      |      | 0   |                               | 0   |       |    |    |                               |       |
| IS                        | 1,594| 4    | 1,598| 96  | 5,037                        | 7   | 5,140 | 6,738|    |                               |       |
| ME                        | 0  |      |      | 0   |                               | 0   |       |    |    |                               |       |
| MK                        | 223 | 10   | 233  |      |                               | 233 |       |    |    |                               |       |
| NO                        | 10,167| 10,087| 9    | 20,263| 20,263                       | 20,263|       |    |    |                               |       |
| RS                        | 124 | 210  | 334  |      |                               | 334 |       |    |    |                               |       |
| TR                        | 26 | 26   | 26   |    |                               | 26 |       |    |    |                               |       |
| Total other non-EU       | 1,594| 0    | 0    | 1,598| 0                            | 10,387| 15,583| 26   | 25,996| 27,594                       |       |
| Total                    | 16,108| 4,471| 1,790| 22,391| 166,889                      | 149,349| 139   | 316,377| 338,768 |                               |       |

**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).

(a): data 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 2021 in the EU and other reporting countries

| Herd status | Surveillance Target group | Infected herds | Non-infected herds | Total |
|-------------|---------------------------|----------------|-------------------|-------|
|             | EM | NSHC | SHC | SU | Subtotal infected herds | EM | NSHC | SHC | SU | Subtotal non-infected herds | |
| AT          | 836 | 31   |     |    |                      | 867 |       |     |    |                                | |
| BE          | 2   | 2    |     |    |                      | 696 | 6     |    | 702 |                                | |
| BG          | 508 | 2,328| 7   |    |                      | 2,843 | 2,843 |   | 2,843 |                                | |
| CY          | 1,349 | 767 | 83  | 2,199 | 547 | 5 | 552 | 2,751 | |
| CZ          | 650 |       |     |    |                      | 650 |       |     |    |                                | |
| DE          | 2,080 | 255 | 9   | 2,344 | 2,344 | 2,344 |   | 2,344 |                                | |
| DK          | 107 | 30   |     |    |                      | 137 | 137 |     |    |                                | |
| EE          | 6   | 6    |     |    |                      | 6   | 6    |     |    |                                | |
| EL          | 422 | 223 | 645 | 639 | 537 | 1,766 | 1,821 | |
| ES          | 1,774 | 1,774 | 10,858 | 8,593 | 19,451 | 21,225 | |
| FI          | 229 |       |     |    |                      | 229 |       |     |    |                                | |
| FR          | 2   | 2    | 16,460 | 4,678 | 21,138 | 21,140 | |
| HR          | 514 |       |     |    |                      | 514 |       |     |    |                                | |
| HU          | 164 | 167 |     |    |                      | 331 | 331 |     |    |                                | |
| IE          | 116 |       |     |    |                      | 116 |       |     |    |                                | |
| IT          | 1,129 | 51 | 337 | 1,517 | 6,541 | 22,083 | 28,624 | 30,141 | |
| LT          | 34  |       |     |    |                      | 34  |       |     |    |                                | |
| LU          | 101 |       |     |    |                      | 101 |       |     |    |                                | |
| LV          | 15  |       |     |    |                      | 15  |       |     |    |                                | |
| MT          | 70  |       |     |    |                      | 70  |       |     |    |                                | |
| NL          | 1,927 |     |     |    | 1,927 | 1,927 | |
| PL          | 3,735 | 1,419 | 5,154 | 5,154 | 5,154 | 5,154 | |
| PT          | 1   | 1    | 1,582 | 5 | 1,587 | 1,588 | |
| RO          | 8,236 | 14,284 | 3 | 22,523 | 22,523 | |
| SE          | 170 |       |     |    |                      | 170 |       |     |    |                                | |
| SI          | 528 | 55 | 588 | 588 | |
| SK          | 5   | 5    | 458 | 458 | 463 | |
| Total EU27  | 3,327 | 1,629 | 1,104 | 85 | 6,145 | 57,807 | 54,470 | 30 | 112,307 | 118,452 | |
| XI(a)       | 5   |       |     |    |                      | 5   |       |     |    |                                | |
| Total EU27 + XI | 3,327 | 1,629 | 1,104 | 85 | 6,145 | 57,812 | 54,470 | 30 | 112,312 | 118,457 | |
| BA          | 0   |       |     |    |                      | 0   |       |     |    |                                | |
| CH          | 0   |       |     |    |                      | 0   |       |     |    |                                | |
| IS          | 16  |       | 16  | 16 |                                | |
| ME          | 0   |       |     |    |                      | 0   |       |     |    |                                | |
| MK          | 14  | 14   |     |    |                      | 14  | 14   |     |    |                                | |
| NO          | 522 | 20   | 542 | 542 |                                | |
| RS          | 20  |       | 20  | 20 |                                | |
| TR          | 3   | 3    | 3   |   |                                | |
| Total other non-EU | 0 | 0 | 0 | 0 | 0 | 542 | 53 | 0 | 595 | 595 | |
| Total       | 3,327 | 1,629 | 1,104 | 85 | 6,145 | 58,354 | 54,523 | 30 | 112,907 | 119,052 | |

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).

(a): Data from XI, United Kingdom (in respect of Northern Ireland) are available from 2021 onwards.
In total, 551 scrapie cases in sheep were reported in the EU27 and XI in 2021, 137 (19.8%) less than in 2020, due to the reduction in cases of Greece (−58.2%) and Spain (−25.8%). They were reported by 17 MS and XI (one country more than in 2020). All countries that reported cases in 2020, except for Belgium, also reported cases in 2021 plus Croatia and Slovenia that reported AS cases only. In addition, 64 scrapie cases in sheep were reported by two other non-EU reporting countries: Iceland and Norway.

CS was reported by six MS: Bulgaria, Cyprus, Greece, Italy, Romania, Spain, and one non-EU reporting country Iceland. AS was reported by 13 MS: the United Kingdom in respect of Northern Ireland and by two other non-EU reporting countries: Croatia, Finland, France, Germany, Hungary, Ireland, Italy, Poland, Portugal, Slovakia, Slovenia, Spain, Sweden, the United Kingdom in respect of Northern Ireland, and two non-EU reporting countries: Iceland and Norway. Most of the ovine CS cases in the EU27 and XI (98.2%) were reported by four countries, namely Greece, Italy, Romania and Spain, as in the previous years.

Out of the 551 sheep scrapie cases reported in the EU27 and XI in 2021, 448 were CS cases (81.3%), 103 were AS cases (18.7%). Among the non-EU reporting countries, 55 CS cases and 1 AS case were reported by Iceland, whereas Norway reported eight AS cases. 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 2021 in sheep is shown in Appendix C. An additional total of 13 cases in sheep were reported as inconclusive by Italy (see Table 31) which are not included in the caseload for this country in Table 21.

In sheep, 176 (31.9%) of all cases in the EU27 and XI reported in 2021 were index cases (IC): 96 AS and 80 CS. This percentage is higher than the previous year (24.6% in 2020) and so is the absolute number of all cases (169 in 2020) that increased by 4.7% in the reporting year. There was a much higher proportion of IC in AS cases (96/103: 93.2%) than in CS cases (80/448: 17.9%), 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 2020 with 2021, there was no change in the number of CS IC (from 81 in 2020 to 80 in 2021, −1.2%) and an increase in the number of AS IC (from 88 to 96, +9.1%). Three CS cases out of 55 (5.5%) and the AS case reported by Iceland were IC, as well as the eight AS cases reported by Norway (100%).

In total, 22 scrapie cases in goats were reported in the EU27 and XI in 2021, a 31.7% reduction (−104) compared with 2020 when 328 cases were reported. This change is due mainly to the decrease in the number of cases in goats in Cyprus of 42.8% (from 236 to 135 cases). One additional scrapie case was reported as inconclusive by Italy (see Table 33) which is not included in the caseload for this country in Table 22. Only Italy and Spain reported both CS and AS. Bulgaria, Cyprus, Greece and Romania reported only CS cases whereas France reported only one AS case. Most of the CS cases were reported by Cyprus with a slight decrease in the contribution of this country to the CS caseload in 2021 (from 72% in 2020 to 60.3% in 2021). The three other non-EU reporting countries that reported tested goats did not report any scrapie cases.

In goats, 15.6% (35) of all cases reported in the EU27 and XI in 2021 were IC: 5 AS and 30 CS. This percentage is lower than the 18.6% (61) observed in 2020, with a higher proportion in AS (5/5: 100%) than in CS (30/219: 13.7%). Italy accounted for 34.3% (12/35) 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 2021 with 2020, there was a decrease in the number of CS IC (from 52 to 30, 42.3%) and there was nearly half number of AS IC (from 9 to 5).

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 was reported only one case of AS scrapie in sheep. In 2021, the CS/AS ratio was 4.4:1 in sheep (lower than in 2020: 6:1) and 43.8:1 in goats (higher than in 2020: 35.4:1). If, for goats, Cyprus is excluded, the CS/AS ratio was 16.8:1 in 2021 compared with 9.2:1 in 2020.
Table 21: Number of confirmed scrapie cases in sheep by country, case type, index case status, surveillance target group in 2021 in the EU and other reporting countries

| Case type       | Atypical scrapie (AS) | Classical scrapie (CS) | Total confirmed cases |
|-----------------|-----------------------|------------------------|-----------------------|
| Index case      | No                    | Yes                    |                        |
| Surveillance target group | EM | NSHC | SHC | Subtotal | EM | NSHC | SHC | Subtotal | Total AS | EM | NSHC | SHC | SU | Subtotal | Total CS |                        |
| BG(a)           | 3                      | 3                      | 6                      | 6                      | 6                      |          |          |
| CY              | 2                      | 2                      |                        |                        |                        |          |          |
| DE              | 4                      | 1                      | 5                      | 5                      |                        |          |          |
| EL              | 2                      | 7                      | 11                     | 45                     | 7                      | 2        | 2         | 74        | 74        |
| ES              | 3                      | 3                      | 6                      | 6                      |                        |          |          |
| FI              | 1                      | 1                      |                        |                        |                        |          |          |
| FR              | 7                      | 7                      |                        |                        |                        |          |          |
| HR              | 2                      | 2                      | 2                      |                        |                        |          |          |
| HU              | 1                      | 1                      | 7                      | 8                      | 15                     | 16       |          |
| IE              | 1                      | 1                      |                        |                        |                        |          |          |
| IT(b)           | 1                      | 1                      | 4                      | 1                      | 5                      | 11       | 11        |
| PL              | 1                      | 5                      | 11                     |                        |                        |          |          |
| PT              | 2                      | 2                      | 32                     |                        |                        |          |          |
| RO              | 3                      | 9                      | 5                      | 17                     | 8                      | 9        | 17        | 34        | 34        |
| SE              | 1                      | 1                      |                        |                        |                        |          |          |
| SI              | 2                      | 2                      |                        |                        |                        |          |          |
| SK              | 5                      | 7                      | 7                      | 12                     |                        |          |          |
| Total EU27      | 0                      | 7                      | 0                      | 7                      | 65                     | 28        | 2         | 95        | 102       | 300     | 41        | 22        | 5       | 368        | 48     | 30        | 2         | 80       | 448        | 550     |          |
| XI(c)           | 1                      | 1                      |                        |                        |                        |          |          |
| Total EU27 + XI | 0                      | 7                      | 0                      | 7                      | 66                     | 28        | 2         | 96        | 103       | 300     | 41        | 22        | 5       | 368        | 48     | 30        | 2         | 80       | 448        | 551     |          |
| IS              | 1                      | 1                      | 1                      | 50                     |                        | 2        | 52        | 1         | 2        | 3         | 55       | 56        |
| NO              | 6                      | 2                      | 8                      | 8                      |                        |          |          |          |          |            |          |            |
| Total other non-EU | 0                      | 0                      | 0                      | 0                      | 7                      | 2        | 0         | 9         | 9        | 50       | 0        | 0         | 7        | 52       | 0         | 1       | 2         | 3       | 55       | 64       |          |
| Total           | 0                      | 7                      | 0                      | 7                      | 73                     | 30        | 2         | 105       | 112       | 350     | 41        | 22        | 0       | 420        | 48     | 31        | 4         | 83       | 503        | 615     |          |
EM: animals culled under TSE eradication measures; NSHC: Animals not slaughtered for human consumption; SHC: animals slaughtered for human consumption; SU: animals clinically suspected of being infected by a TSE.

Only the reporting countries in which scrapie cases in sheep were detected in 2021 are mentioned in the table.

(a): Bulgaria reported Scrapie cases by result date. Cases before which the date of testing occurred in the last months of 2021 might be reported at the beginning of 2022. Same applies to cases sampled at the end of 2020.

(b): 13 inconclusive cases from Italy are not reported in this table.

(c): Data from United Kingdom (in respect of Northern Ireland) (XI) are available from 2021 onwards.
Table 22: Number of confirmed scrapie cases in goats by country, case type, index case status, surveillance target group in 2021 in the EU and other reporting countries

| Surveillance target group | Case type | Atypical scrapie (AS) | Classical scrapie (CS) | Total confirmed cases |
|---------------------------|-----------|-----------------------|-----------------------|----------------------|
|                           | Index case | No | Yes | Subtotal | Total AS | No | Yes | Subtotal | No | Yes | Subtotal | No | Yes | Subtotal | Total CS |
|                           | EM | NSHC | SHC | Subtotal | NSHC | SHC | Subtotal | EM | NSHC | SHC | Subtotal | NSHC | SHC | Subtotal | |
| BG(a)                     | No | Yes | Subtotal | NSHC | SHC | Subtotal | NSHC | SHC | Subtotal | NSHC | SHC | Subtotal | 3 | 1 | 4 | 4 |
| CY                        | No | Yes | Subtotal | NSHC | SHC | Subtotal | NSHC | SHC | Subtotal | NSHC | SHC | Subtotal | 39 | 27 | 64 | 135 |
| EL                        | No | Yes | Subtotal | NSHC | SHC | Subtotal | NSHC | SHC | Subtotal | NSHC | SHC | Subtotal | 1 | 7 | 8 | 3 |
| ES                        | No | Yes | Subtotal | NSHC | SHC | Subtotal | NSHC | SHC | Subtotal | NSHC | SHC | Subtotal | 1 | 1 | 2 | 2 |
| FR                        | No | Yes | Subtotal | NSHC | SHC | Subtotal | NSHC | SHC | Subtotal | NSHC | SHC | Subtotal | 38 | 1 | 4 | 5 |
| IT(b)                     | No | Yes | Subtotal | NSHC | SHC | Subtotal | NSHC | SHC | Subtotal | NSHC | SHC | Subtotal | 13 | 1 | 9 | 10 |
| RO                        | No | Yes | Subtotal | NSHC | SHC | Subtotal | NSHC | SHC | Subtotal | NSHC | SHC | Subtotal | 3 | 3 | 3 | 3 |
| Total EU27 + XI(c)        | No | Yes | Subtotal | NSHC | SHC | Subtotal | NSHC | SHC | Subtotal | NSHC | SHC | Subtotal | 52 | 46 | 27 | 64 |
| Total other non-EU        | No | Yes | Subtotal | NSHC | SHC | Subtotal | NSHC | SHC | Subtotal | NSHC | SHC | Subtotal | 189 | 10 | 19 | 1 |
| Total                     | No | Yes | Subtotal | NSHC | SHC | Subtotal | NSHC | SHC | Subtotal | NSHC | SHC | Subtotal | 30 | 219 | 224 |

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).

Only the reporting countries in which scrapie cases in goats were detected in 2021 are included in the table.

(a): Bulgaria reported Scrapie cases by result date. Cases that should be in the last months of 2020 might end up in 2021.

(b): One inconclusive case from Italy has not been included in the total number of classical scrapie cases in this table.

(c): Data from United Kingdom (in respect of Northern Ireland) (XI) are available from 2021 onwards.
Focusing on the last 10 years, the evolution in the number of scrapie cases detected at EU27 and the United Kingdom 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 3,142 (when the number of tests also peaked), CS cases have decreased from 1,416 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. A further decrease is clear in 2021 to 448 cases, primarily due to the lower number of CS cases in Greece (−58%), Romania (−34.6%) and Spain (−24.6%). This trend is likely to be associated with the decrease in the testing of TSE-infected flocks due to the lower number of flocks under restrictions and intensified surveillance and the reduction of the number of IC in Greece, with 14 fewer IC in 2021.

In goats, the decreasing trend in the absolute number of CS cases continued in 2021. The evolution is mainly associated with one single MS (Cyprus), where the number of detected cases has consistently declined since the peak in 2013 when 1,792 cases were reported by the country and 1,799 in total including all other MS. Since then, the total number of CS cases in EU27 and XI has consistently decreased to 219 in 2021. In 2021 compared with 2020, there was also a small reduction in the number of CS in goats in Greece from 27 to 11. Whereas Italy and Spain reported an increase in the number of CS cases in 2021 by 10 (23.1%) and 14 (+48.3%) cases, respectively. Cyprus and Greece reported a decrease in index CS cases, from 12 to 5 and from 21 to 3, respectively, whereas Italy reported an increase from 6 to 10.

Based on the 39,113 cases of scrapie with known type, species and age between 2002 and 2021 in sheep, the average age of AS cases (82.6 months) is significantly higher (p < 0.001) than that of CS cases (48 months). Similarly, in goats, the average age of AS cases (84.3 months) is significantly higher (p < 0.001) than that of CS cases (51.7 months). When comparing sheep with goats, there was no significant difference in the average age for AS (p = 0.6) whereas the average age for CS in sheep is lower than in goats (p < 0.001).
Tables 23 and 24 show the cases of CS and AS, respectively, in sheep for the period 2002–2021, 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 2002–2021, with a focus on the last 5 years.

In sheep, in 2021, 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: 3.1 in NSHC and 2.2 in SHC; (2) for AS: 4.3 in NSHC and 2.1 in SHC.

In goats, in 2021, 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: 1.7 in NSHC and 3.5 in SHC; (2) for AS: 0.5 in NSHC and 0.4 in SHC.

### Table 23: Number of classical scrapie cases in sheep by year and reporting country between 2002 and 2021

| Country | Up to 2016 | 2017 | 2018 | 2019 | 2020 | 2021 | Total CS |
|---------|------------|------|------|------|------|------|----------|
| BE      | 38         |      |      |      |      |      | 38       |
| BG      | 11         | 1    | 5    | 8    | 6    | 31    |          |
| CY      | 3,204      | 2    | 4    | 1    | 2    | 2    | 3,215    |
| CZ      | 56         |      |      |      |      |      | 56       |
| DE      | 116        |      |      |      |      |      | 116      |
| EL      | 5,744      | 247  | 178  | 276  | 176  | 74    | 6,695    |
| ES      | 1,139      | 247  | 279  | 312  | 244  | 184   | 2,405    |
| FR      | 1,534      |      |      |      |      |      | 1,534    |
| HU      | 10         |      |      |      |      |      | 10       |
| IE      | 575        | 11   | 1    |      |      |      | 587      |
| IT      | 2,659      | 240  | 150  | 171  | 102  | 148   | 3,470    |
| NL      | 401        |      |      |      |      |      | 401      |
| PT      | 33         | 1    | 5    |      |      |      | 39       |
| RO      | 733        | 76   | 203  | 141  | 52   | 34    | 1,239    |
| SI      | 174        |      |      |      |      |      | 174      |
| SK      | 117        | 15   |      |      |      |      | 132      |
| Total EU27 | 16,544   | 839  | 820  | 902  | 589  | 448   | 20,142   |
| Total EU27 + XI (a) | 16,544 | 839  | 820  | 902  | 589  | 448   | 20,142   |
| IS      | 206        | 1    | 21   | 21   | 53   | 55    | 357      |
| NO      | 17         |      |      |      |      |      | 17       |
| United Kingdom | 1,995 | 9    |      |      |      |      | 2,004    |
| Total other non-EU | 2,218 | 1    | 21   | 30   | 53   | 55    | 2,378    |
| Total   | 18,762     | 840  | 841  | 932  | 642  | 503   | 22,520   |

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.7282881.

(a): Data 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 between 2002 and 2021 in the reporting countries

| Country | Up to 2016 | 2017 | 2018 | 2019 | 2020 | 2021 | Total AS |
|---------|------------|------|------|------|------|------|----------|
| AT      | 13         | 1    | 1    |      |      |      | 15       |
| BE      | 8          |      |      | 2    |      |      | 10       |
| BG      | 6          |      |      |      |      |      | 6        |
| CZ      | 7          | 1    |      |      |      |      | 8        |
| DE      | 120        | 4    | 4    | 4    | 14   | 5    | 151      |
| DK      | 13         |      |      |      |      |      | 14       |
| EE      | 2          |      |      |      |      |      | 2        |
| EL      | 32         |      |      | 1    |      |      | 33       |
| ES      | 211        | 12   | 9    | 7    | 12   | 6    | 257      |
### Table 24: Number of classical scrapie cases in sheep by year and country between 2002 and 2021 in the reporting countries

| Country | Up to 2016 | 2017 | 2018 | 2019 | 2020 | 2021 | Total AS |
|---------|-----------|------|------|------|------|------|----------|
| FI      | 13        | 2    | 3    | 1    | 1    | 1    | 20       |
| FR      | 554       | 6    | 8    | 3    | 7    | 581   |
| HR      | 2         | 2    | 4    |      |      |       |
| HU      | 129       | 14   | 13   | 17   | 14   | 16   | 203      |
| IE      | 35        | 1    | 8    | 6    | 1    | 1    | 52       |
| IT      | 91        | 3    | 8    | 6    | 6    | 121   |
| NL      | 18        |      |      |      |      |      | 18       |
| PL      | 47        | 7    | 6    | 4    | 5    | 11   | 80       |
| PT(a)   | 617       | 29   | 30   | 20   | 18   | 32   | 746      |
| RO      |           | 1    |      |      |      |      | 1        |
| SE      | 45        | 2    | 2    | 1    | 1    |      | 51       |
| SI      | 10        |      |      |      |      |      | 12       |
| SK      | 27        | 5    | 6    | 4    | 5    | 12   | 59       |
| **Total EU27** | **2,000** | **82** | **96** | **80** | **84** | **102** | **2,444** |
| XI(b)   |           | 1    |      |      |      |      | 1        |
| **Total EU27 + XI** | **2,000** | **83** | **97** | **80** | **84** | **103** | **2,445** |
| IS      | 8         |      |      |      |      |      | 9        |
| NO      | 138       | 13   | 8    | 10   | 12   | 8    | 189      |
| **United Kingdom** | **333** | **12** | **17** | **6** | **14** |      | **382** |
| **Total other non-EU** | **479** | **25** | **25** | **16** | **26** | **9** | **580** |
| **Total** | **2,479** | **107** | **121** | **96** | **110** | **112** | **3,025** |

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

(a): In 2018, it includes one imported case: an animal imported for slaughter from Spain which died at lairage/resting area of the slaughterhouse and was tested as NSHC.

(b): Data from United Kingdom (in respect of Northern Ireland) (XI) are available from 2021 onwards.

### Table 25: Number of classical scrapie cases in goats by year and country between 2002 and 2021 in the reporting countries

| Country | Up to 2016 | 2017 | 2018 | 2019 | 2020 | 2021 | Total CS |
|---------|-----------|------|------|------|------|------|----------|
| BG      | 7         | 2    | 5    | 7    | 4    | 25   |
| CY      | 10,286    | 484  | 381  | 308  | 236  | 135  | 11,830   |
| EL      | 550       | 25   | 19   | 7    | 27   | 11   | 639      |
| ES      | 106       | 34   | 89   | 35   | 29   | 43   | 336      |
| FI      | 8         |      |      |      |      |      | 8        |
| FR      | 168       |      |      |      |      |      | 173      |
| HU      | 0         | 1    |      |      |      |      | 1        |
| IT      | 101       | 8    | 15   | 26   | 13   | 23   | 186      |
| RO      | 11        | 2    | 3    | 5    | 3    | 24   |
| SI      | 4         |      |      |      |      |      | 4        |
| **Total EU27** | **11,241** | **555** | **517** | **377** | **317** | **219** | **13,226** |
| XI(a)   |           |      |      |      |      |      |          |
| **Total EU27 + XI** | **11,241** | **555** | **517** | **377** | **317** | **219** | **13,226** |
| United Kingdom | 226 | 3 | 2 | 2 | 2 | 233 |
| **Total other non-EU** | **226** | **3** | **0** | **2** | **2** | **0** | **233** |
| **Total** | **11,467** | **558** | **517** | **379** | **319** | **219** | **13,459** |

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

(a): Data 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.7282881](https://doi.org/10.5281/zenodo.7282881), 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.

Figure 3 shows the 10-year trend (2012–2021): 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 2012–2020, and EU27 and XI for 2021. The results of the Poisson regression models, in sheep, showed a statistically significant decreasing trend for CS (annual RR: 0.97, p < 0.01) and AS (annual RR = 0.963, p < 0.001), i.e. 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 = 0.49 and 0.14, respectively).

The number of historical reported scrapie cases can be found in the following [https://doi.org/10.5281/zenodo.7282881](https://doi.org/10.5281/zenodo.7282881), 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.

Table 26: Number of atypical scrapie cases in goats by year and country between 2002 and 2021 in the EU and other reporting countries

| Country | Up to 2016 | 2017 | 2018 | 2019 | 2020 | 2021 | Total AS |
|---------|------------|------|------|------|------|------|----------|
| AT      | 1          |      |      |      |      |      | 1        |
| CY      | 1          | 1    | 1    | 1    |      |      | 4        |
| DE      | 1          |      |      |      |      | 1    | 2        |
| DK      | 0          |      |      |      | 1    |      | 1        |
| EL      | 4          | 2    | 2    | 2    | 3    | 2    | 5        |
| ES      | 49         | 2    | 2    | 2    | 3    | 2    | 60       |
| FI      | 1          |      |      |      |      |      | 1        |
| FR      | 56         | 2    | 3    | 1    | 1    |      | 63       |
| IT      | 21         | 3    | 3    | 3    | 2    |      | 34       |
| PL      | 0          | 1    |      |      |      |      | 1        |
| PT      | 13         | 1    | 1    |      |      |      | 15       |
| SI      | 1          |      |      |      |      |      | 1        |
| Total EU27 | 148    | 9    | 6    | 11   | 9    | 5    | 188      |
| Total EU27+ XI (a) | 148     | 9    | 6    | 11   | 9    | 5    | 188      |
| NO      | 1          |      |      |      |      |      | 1        |
| Total other non-EU | 1       |      |      |      |      |      | 1        |
| Total   | 149        | 9    | 6    | 11   | 9    | 5    | 189      |

EU and reporting countries without atypical scrapie cases in goats are not included in the table. (a): Data from United Kingdom (in respect of Northern Ireland) (XI) are available from 2021 onwards.
Tables 31 and 32 summarise the number of discriminatory tests performed by country in 2021 for CS and AS in sheep. Tables 33 and 34 summarise the number of discriminatory tests performed by country in 2021 for CS and AS in goats. In sheep, 459 (99.6%) of the CS and all the inconclusive cases reported in the EU27 and XI were submitted for discriminatory testing and so were 29 of the AS cases (28.1%). The 55 cases of CS reported by Iceland and the eight cases of AS reported by Norway were also submitted to discriminatory testing. All sheep scrapie cases submitted for discriminatory testing were confirmed as ‘BSE excluded’, except 13 cases reported by Italy, that were declared ‘inconclusive’. In goats, 90 (40.9%) of the CS cases reported in the EU27 and XI were submitted for discriminatory testing as well as four of the AS cases (80%) and one inconclusive case. All goat cases subjected to discriminatory testing were confirmed as ‘BSE excluded’.

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

**Figure 3:** Number of scrapie (index) cases per 10,000 tests in the EU27 and the United Kingdom until 2020 and the EU27 and XI from 2021 in (a) sheep and (b) goats in non-TSE-infected flocks/herds, reported by case type and target group in the period 2012–2021.
Table 31: Number of discriminatory tests and results in classical and inconclusive scrapie cases in sheep by reporting country

| Country | No. of classical scrapie, and inconclusive cases | Cases submitted for discriminatory testing | % of total classical scrapie and inconclusive case(b) |
|---------|-----------------------------------------------|------------------------------------------|--------------------------------------------------|
|         | No. of classical scrapie, and inconclusive cases | BSE-not-excluded | BSE-excluded | Inconclusive(a) | Total | |
| BG      | 6 | 6 | 6 | 6 | 100% |
| CY      | 2 | 0 | 0 | 0 | 0% |
| EL      | 74 | 74 | 74 | 74 | 100% |
| ES      | 184 | 184 | 184 | 184 | 100% |
| IT      | 161 | 148 | 13 | 161 | 100% |
| RO      | 34 | 34 | 34 | 34 | 100% |
| **Total EU27** | **461** | 0 | **446** | **13** | **459** | 99.6% |
| IS      | 55 | 55 | 55 | 55 | 100% |
| **Total other non-EU** | **55** | 0 | **55** | 0 | **55** | 100% |
| **Total** | **516** | 0 | **501** | **13** | **514** | 99.6% |

(a): The inconclusive cases have not been included in the total number of sheep scrapie cases in Table 21; these are non-index cases whose discrimination between BSE and scrapie has not been concluded due to technical problems.

(b): It indicates the proportion of classical TSE and inconclusive cases that are submitted to discriminatory testing by each reporting country.

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

Table 32: Number of discriminatory tests and results in atypical scrapie cases in sheep by reporting country

| Country | No. of atypical scrapie cases | Cases submitted for discriminatory testing | % of total atypical scrapie cases(a) |
|---------|-------------------------------|------------------------------------------|-----------------------------------|
|         | No. of atypical scrapie cases | BSE-not-excluded | BSE-excluded | Total | |
| DE      | 5 | 0 | 0 | 0% |
| ES      | 6 | 6 | 6 | 100% |
| FI      | 1 | 0 | 0 | 0% |
| FR      | 7 | 0 | 0 | 0% |
| HR      | 2 | 0 | 0 | 0% |
| HU      | 16 | 16 | 16 | 100% |
| IE      | 1 | 0 | 0 | 0% |
| IT      | 6 | 6 | 6 | 100% |
| PL      | 11 | 0 | 0 | 0% |
| PT      | 32 | 0 | 0 | 0% |
| SE      | 1 | 0 | 0 | 0% |
| SI      | 2 | 0 | 0 | 0% |
| SK      | 12 | 0 | 0 | 0% |
| **Total EU27** | **102** | 0 | **28** | **28** | **27.5%** |
| XI(b)   | 1 | 1 | 1 | 100% |
| **Total EU27 + XI** | **103** | 0 | **29** | **29** | **28.2%** |
| IS      | 1 | 0 | 0 | 0% |
| NO      | 8 | 8 | 8 | 100% |
| **Total other non-EU** | **9** | 0 | **8** | **8** | **89%** |
| **Total** | **112** | 0 | **37** | **37** | **33%** |
3.2.1. Genotyping

3.2.1.1. Sheep

The classification of genotypes of the sheep prion protein PRNP gene used in this report and based on to the Great Britain’s NSP is summarised in Table 35.

| NSP group | Genotype  | Comment                       | Report group |
|-----------|-----------|-------------------------------|--------------|
| NSP1      | ARR/ARR   | Genetically most resistant    | Resistant    |
| NSP2      | ARR/ARQ; ARR/ARH; ARR/AHQ | Genetically resistant      | Semi-resistant |
| NSP3      | ARQ/ARQ   | Genetically little resistant (ARQ/ARQ may be scientifically reviewed) | Susceptible  |

Table 33: Number of discriminatory tests and results in classical and inconclusive scrapie cases in goats in 2021 by reporting country

| Country | No. of classical scrapie and inconclusive cases | BSE-not-excluded | BSE-excluded | Inconclusive | Total | % of total classical scrapie and inconclusive cases |
|---------|-----------------------------------------------|------------------|--------------|--------------|-------|-----------------------------------------------|
| BG      | 4                                             | 4                |              |              | 4     | 100%                                          |
| CY      | 135                                           | 5                |              |              | 5     | 3.7%                                         |
| EL      | 11                                            | 11               |              |              | 11    | 100%                                         |
| ES      | 43                                            | 43               |              |              | 43    | 100%                                         |
| IT      | 24                                            | 23               | 1            |              | 24    | 100%                                         |
| RO      | 3                                             | 3                |              |              | 3     | 100%                                         |
| **Total** | **220**                                     | **0**           | **89**       | **1**        | **90** | **40.9%**                                      |
| **EU27** |                                                |                  |              |              |       |                                               |
| **Total** | **220**                                     | **0**           | **89**       | **1**        | **90** | **40.9%**                                      |

Table 34: Number of discriminatory tests and results in atypical scrapie cases in goats by reporting country

| Country | No. of atypical scrapie cases | BSE-not-excluded | BSE-excluded | Total | % of total atypical scrapie |
|---------|-------------------------------|------------------|--------------|-------|-----------------------------|
| ES      | 2                             | 2                | 2            | 4     | 100%                        |
| FR      | 1                             | 0                |              | 1     | 0%                          |
| IT      | 2                             | 2                | 2            | 4     | 100%                        |
| **Total EU27** | **5**                     | **0**           | **4**        | **4** | **80%**                     |
| **Total** | **5**                        | **0**           | **4**        | **4** | **80%**                     |

BSE: bovine spongiform encephalopathy, TSE: transmissible spongiform encephalopathies.
EU and reporting countries without atypical scrapie cases in sheep are not included in the table.
(a): The inconclusive case has not been included in the total number of goats’ scrapie cases in Table 21.
(b): Indicates the proportion of atypical TSE cases that are submitted to discriminatory testing by each reporting country.

Table 35: Classification of the genotypes of the sheep prion protein PRNP gene according to Great Britain’s National Scrapie Plan (NSP) and the three tiers of report groups

| NSP group | Genotype | Comment                       | Report group |
|-----------|----------|-------------------------------|--------------|
| NSP1      | ARR/ARR  | Genetically most resistant    | Resistant    |
| NSP2      | ARR/ARQ; ARR/ARH; ARR/AHQ | Genetically resistant | Semi-resistant |
| NSP3      | ARQ/ARQ  | Genetically little resistant (ARQ/ARQ may be scientifically reviewed) | Susceptible |

BSE: bovine spongiform encephalopathy, TSE: transmissible spongiform encephalopathies.
EU and reporting countries without atypical scrapie cases in sheep are not included in the table.
(a): Indicates the proportion of atypical TSE cases that are submitted to discriminatory testing by each reporting country.
Table 36 shows the genotypes of sheep scrapie cases in 2021 in the EU and other reporting countries.

In total, 423 (98.6%) of the 429 cases of CS in sheep with NSP genotype reported in the EU27 and XI in 2021 (94.4% 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% of all CS cases with known genotypes were from the susceptible groups. In the other non-EU reporting countries, 50 CS cases reported by Iceland (90.9%) were from the susceptible genotype groups. In the current year, no country has reported cases of CS in a sheep with the ARR/ARR genotype (NSP1), a very rare event last reported by Spain in 2019 and by Romania in 2020.

Among ovine AS cases, 45 (48.4%) of the 93 cases of AS in sheep with NSP genotype reported in the EU27 and XI in 2021 (90.3% the total AS caseload) were from the genotype groups NSP3, NSP3O NSP4 or NSP5, very similar to 2020. The additional 48 AS case showed an NSP1 or NSP2 that are often associated with this case type. Figure 4 shows the frequency distribution of genotypes of sheep scrapie cases by case type, year and NSP group in the period 2012–2021 in the reporting countries.

Table 37 shows the genotypes obtained in 2021 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, nine MS conducted the genotype on a random sample of sheep: Belgium, Cyprus where genotyping is conducted systematically in the breeding sheep population, France, Germany, Greece, Italy, Latvia, the Netherlands and Poland. One of the eight other non-EU reporting countries, Iceland, also reported the number of genotyped sheep: Data have not been included in the table as they were obtained from scrapie outbreaks rather than form a random sample of the sheep population. The subset of EU27 and XI that carried out the activity in 2021 reported a total of 5,411 known genotypes. Excluding data from Cyprus, 7.9% of the sheep population (with known NSP genotype) were susceptible to CS (NSP3, NSP3O, NSP4 and NSP5), lower than the 8.8% in 2020 and lower than the 15.7% in 2019. This percentage stands at 21.2% in Italy, one of the countries with the highest case load in 2021, whereas it was between 0.9% and 75% in the remaining seven MS.

Considering the past 10 years of random genotyping and 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 66.8% in 2021 (in which eight MS contributed), even though lower than the 70.1% in 2020.
Table 36: Distribution of genotypes of confirmed scrapie cases in sheep by reporting country and National Scrapie Plan (NSP) group

| Country/NSP types | Atypical scrapie | Classical scrapie | Inconclusive (c) | Total scrapie cases |
|-------------------|-----------------|------------------|-----------------|--------------------|
|                   | NSP1 | NSP2 | NSP3 | NSP3O | NSP4 | NSP5 | Unknown N/G | Total AS | NSP1 | NSP2 | NSP3 | NSP3O | NSP4 | NSP5 | Unknown N/G | Total CS | NSP3 | NSP3O | Unknown N/G | Total Inconclusive | Total cases |
| BG                |      |      |      |       |      |      |            | 6        | 6    |      |      |      |      |      |            |          |      |      |      |            | 6              |
| CY                |      |      |      |       |      |      |            | 1        | 1    |      |      |      |      |      |            |          |      |      |      |            | 2              |
| DE                | 2    | 1    | 2    |       |      |      |            | 5        |      |      |      |      |      |      |            |          |      |      |      |            | 5              |
| EL                |      |      |      |       |      |      |            | 1        | 56   | 14   |      |      |      |      |            |          |      |      |      |            | 74             |
| ES                | 2    | 1    | 2    |       | 1    | 6    |            | 171      | 4    | 9    | 184  |      |      |      |            |          |      |      |      |            | 190            |
| FI                | 1    |      |      |       |      |      |            | 1        |      |      |      |      |      |      |            |          |      |      |      |            | 1              |
| FR                | 2    | 1    |      |       | 4    | 7    |            |          |      |      |      |      |      |      |            |          |      |      |      |            | 7              |
| HR                | 1    | 1    |      |       |      |      |            | 2        |      |      |      |      |      |      |            |          |      |      |      |            | 2              |
| HU                | 4    | 3    | 4    | 5     |      |      |            | 16       |      |      |      |      |      |      |            |          |      |      |      |            | 16             |
| IE                | 1    |      |      |       |      |      |            | 1        |      |      |      |      |      |      |            |          |      |      |      |            | 1              |
| IT                | 2    | 1    | 2    |       | 1    | 6    |            | 134      | 10   |      | 4    | 148  | 12   | 1    |            |          |      |      |      |            | 167            |
| PL                | 9    | 2    |      |       |      |      |            | 11       |      |      |      |      |      |      |            |          |      |      |      |            | 11             |
| PT                | 6    | 5    | 11   | 7     |      |      |            | 3        | 32   |      |      |      |      |      |            |          |      |      |      |            | 32             |
| RO                |      |      |      |       | 4    | 18   | 2          | 10       |      |      |      |      |      |      |            |          |      |      |      |            | 34             |
| SE                | 1    |      |      |       |      |      |            | 1        |      |      |      |      |      |      |            |          |      |      |      |            | 1              |
| SI                | 1    | 1    |      |       |      |      |            | 1        |      |      |      |      |      |      |            |          |      |      |      |            | 1              |
| SK                | 1    | 9    | 2    |       |      |      |            | 12       |      |      |      |      |      |      |            |          |      |      |      |            | 12             |
| Total EU27        | 11   | 36   | 24   | 21    |      |      |            | 10       | 102  | 6    | 379  | 27   |      |      |            |          |      |      |      |            | 13             |
| XI(d)             |      |      |      |       |      |      |            | 1        |      |      |      |      |      |      |            |          |      |      |      |            | 1              |
| Total EU27 + XI   | 11   | 37   | 24   | 21    |      |      |            | 10       | 103  | 6    | 379  | 27   |      |      |            |          |      |      |      |            | 13             |
| IS                | 1    | 1    |      |       | 38   |      |            |          |      |      |      |      |      |      |            |          |      |      |      |            | 56             |
| NO                | 1    | 2    | 5    |       |      |      |            | 8        |      |      |      |      |      |      |            |          |      |      |      |            | 8              |
### EUSR on transmissible spongiform encephalopathies in 2021

Table showing the distribution of scrapie cases by NSP types and inconclusive cases:

| Country/NSP types | Atypical scrapie | Classical scrapie | Inconclusive<sup>(c)</sup> | Total scrapie cases |
|-------------------|------------------|-------------------|---------------------------|--------------------|
| NSP1              | 1                | 38                | 12                        | 55                 |
| NSP2              | 2                | 26                | 26                        | 51                 |
| NSP3              | 5                | 1                 | 12                        | 38                 |
| NSP3O             | 9                | 2                 | 12                        | 64                 |
| Total AS           | 38               | 12                | 12                        | 64                 |
| Total CS           | 27               | 24                | 55                        | 226                |
| Total              | 61               | 60                | 60                        | 226                |
| Total N/G          | 55               | 24                | 55                        | 29                 |
| Total Unknown      | 12               | 1                 | 12                        | 13                 |
| Total other non-EU| 64               | 628               |                           | 692                |

(a): Unknown: genotype other than those included in the NSP list.
(b): N/G: not genotyped.
(c): The inconclusive cases have not been included in the total number of sheep scrapie cases in Table 21.
(d): Data from United Kingdom (in respect of Northern Ireland) (XI) are available from 2021 onwards.
(a) Atypical scrapie. (b) Classical scrapie. NSP1: resistant (black); NSP2: semi-resistant (grey); NSP3 (orange) + NSP3O (yellow) + NSP4 (red) + P5 (blue): susceptible as referred in Table 35.

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 2012–2021 in the reporting countries
Table 37: Number of genotyped animals (% of sample within country) in randomly selected sheep in the EU and other reporting countries in 2021 by reporting country and National Scrapie Plan (NSP) group, in accordance with Regulation (EC) 999/2001 Annex VII, Chapter C, Part I, point 8

| Country | NSP1 | NSP2 | NSP3 | NSP3O | NSP4 | NSP5 | Other | Total |
|---------|------|------|------|-------|------|------|-------|-------|
| AT      | n/a  | n/a  | n/a  | n/a   | n/a  | n/a  | n/a   | n/a   |
| BE      | 410  | 128  | 24   | 18    | 12   | 6    | 0     | 598   |
|         | (68.6%) | (21.4%) | (4%) | (3%) | (2%) | (1%) | (0%) |       |
| BG      | n/a  | n/a  | n/a  | n/a   | n/a  | n/a  | n/a   | n/a   |
| CY(a)   | 56,645 | 3,994 | 107  | 72    | 88   | 9    | 188   | 61,103 |
|         | (92.7%) | (6.5%) | (0.2%) | (0.1%) | (0.1%) | (0.0%) | (0.3%) |       |
| CZ      | n/a  | n/a  | n/a  | n/a   | n/a  | n/a  | n/a   | n/a   |
| DE      | 3,475 | 1,172 | 226  | 44    | 9    | 0    | 1     | 4,927 |
|         | (70.5%) | (23.8%) | (4.6%) | (0.9%) | (0.2%) | (0.0%) | (0%) |       |
| DK      | n/a  | 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   | n/a   |
| EL      | 0    | 2    | 3    | 1     | 1    | 1    | 0     | 8     |
|         | (0%) | (25%) | (37.5%) | (12.5%) | (12.5%) | (12.5%) | (0%) |       |
| ES      | n/a  | 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   | n/a   |
| FR      | 246  | 104  | 24   | 5     | 6    | 2    | 0     | 387   |
|         | (63.6%) | (26.9%) | (6.2%) | (1.3%) | (1.6%) | (0.5%) | (0%) |       |
| HR      | n/a  | 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   | n/a   |
| IE      | n/a  | n/a  | n/a  | n/a   | n/a  | n/a  | n/a   | n/a   |
| IT      | 250  | 297  | 104  | 29    | 5    | 9    | 2     | 696   |
|         | (35.9%) | (42.7%) | (14.9%) | (4.2%) | (0.7%) | (1.3%) | (0.3%) |       |
| LT      | n/a  | 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   | n/a   |
| LV      | 102  | 5    | 0    | 1     | 0    | 0    | 0     | 108   |
|         | (94.4%) | (4.6%) | (0%) | (0.9%) | (0%) | (0%) | (0%) |       |
| MT      | n/a  | n/a  | n/a  | n/a   | n/a  | n/a  | n/a   | n/a   |
| NL      | 889  | 266  | 32   | 28    | 26   | 6    | 33    | 1,280 |
|         | (69.5%) | (20.8%) | (2.5%) | (2.2%) | (2%) | (0.5%) | (2.6%) |       |
| PL      | 39   | 41   | 17   | 3     | 0    | 0    | 0     | 100   |
|         | (39%) | (41%) | (17%) | (3%) | (0%) | (0%) | (0%) |       |
| PT      | n/a  | 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   | n/a   |
| SE      | n/a  | 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   | n/a   |
| SK      | n/a  | n/a  | n/a  | n/a   | n/a  | n/a  | n/a   | n/a   |
| Total EU27 | 62,056 | 6,009 | 537 | 201 | 147 | 33 | 224 | 69,207 |
|           | (89.7%) | (8.7%) | (0.8%) | (0.3%) | (0.2%) | (0%) | (0.3%) |       |
| XI(b)   | n/a  | n/a  | n/a  | n/a   | n/a  | n/a  | n/a   | n/a   |
| Total EU27 + XI | 62,056 | 6,009 | 537 | 201 | 147 | 33 | 224 | 69,207 |
|           | (89.7%) | (8.7%) | (0.8%) | (0.3%) | (0.2%) | (0.04%) | (0.3%) |       |
| Total(c) | 62,056 | 6,009 | 537 | 201 | 147 | 33 | 224 | 69,207 |
|           | (89.7%) | (8.7%) | (0.8%) | (0.3%) | (0.2%) | (0.04%) | (0.3%) | (100%) |

n/a: genotyping not performed.
(a): The Cyprus data are different from those of other reporting countries since Cyprus systematically genotypes the breeding sheep population.
(b): Data from XI, United Kingdom (in respect of Northern Ireland) are available from 2021 onwards.
(c): In previous years, Iceland reported genotypes from scrapie culled flocks rather than from random genotyping: as those data are not complying with the EU requirements, the data provided by Iceland have not been included in the table.
3.2.1.2. Goats

In 2021, at least one polymorphism in codons 146 or 222 was reported from 152 cases (2 AS, 149 CS and 1 inconclusive) by Cyprus, Greece and Italy, three of the seven countries in the EU27 and XI that reported cases. Forty-eight (33.2%) samples from CS cases and 2 from AS cases and 1 inconclusive were characterised both for the 146 and 222 codons. The reported polymorphisms in atypical, classical and inconclusive cases were N at the 146 codon and Q at the 222 codon, when available. Table 38 shows the genotypes of goat scrapie cases in 2021 in the EU. None of the non-EU reporting countries reported any case.

| 2012 | 2013 | 2014 | 2015 | 2016 | 2017 | 2018 | 2019 | 2020 | 2021 |
|------|------|------|------|------|------|------|------|------|------|
| 22   | 24   | 25   | 25   | 25   | 20   | 7    | 7    | 8    | 8    |
| Total genotyped | 8,390 | 8,340 | 9,437 | 9,823 | 9,413 | 8,871 | 2,713 | 3,180 | 7,985 | 5,411 |

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 in Table 36.

**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 2012–2021 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.

3.2.1.2. Goats

In 2021, at least one polymorphism in codons 146 or 222 was reported from 152 cases (2 AS, 149 CS and 1 inconclusive) by Cyprus, Greece and Italy, three of the seven countries in the EU27 and XI that reported cases. Forty-eight (33.2%) samples from CS cases and 2 from AS cases and 1 inconclusive were characterised both for the 146 and 222 codons. The reported polymorphisms in atypical, classical and inconclusive cases were N at the 146 codon and Q at the 222 codon, when available. Table 38 shows the genotypes of goat scrapie cases in 2021 in the EU. None of the non-EU reporting countries reported any case.

**Table 38:** Distribution of genotypes of confirmed scrapie cases in goats

| Country | Atypical scrapie | Classical scrapie | Inconclusive |
|---------|------------------|-------------------|--------------|
|         | NQ/ NQ | ZZ/ZZ | Total AS | NQ/ NQ | NZ/NZ NZ/ZZ ZZ/ZZ | Total CS | NQ/ NQ | Total inconclusive | Total |
| **BG**  |         |       |         |        |                 |           |       |                  |       |
| **CY**  | 14      | 99    | 2       | 20     | 135             |           |       |                  | 135   |
| **EL**  | 11      |       |         | 11     |                 |           |       |                  | 11    |
| **ES**  | 2       | 2     |         | 43     | 43              |           |       |                  | 45    |
3.3. TSE surveillance in cervids

In 2021, 5,854 cervids were tested for TSE in the EU27 and XI. The eight MS that contributed to the monitoring were Estonia, Finland, Hungary, Italy, Latvia, Romania, Spain and Sweden. Two countries, Sweden and Romania, contributed to 82.1% of the total number of tested cervids in the EU and XI, with 56.7% (3,316) and 25.4% (1,485), respectively.

Out of the 5,854 cervids tested by the MS, 2,967 (50.7%) were captive, farmed or semi-domesticated animals, mostly reindeer (2,656), followed by red deer (286). Among the 2,887 (49.3%) wild cervids tested by MS, 516 (17.9%) were roe deer and 521 European moose (18.1%). Romania reported 1,485 tested wild deer (species not specified), 51.4% of all wild cervids.

When considering the target groups, the most common target group tested by the MS was the ‘Hunted/slaughtered fit for human consumption’ (HSHC) that accounted for 4,110 (70.2% of all tested cervids, being the main target group for Romania and Sweden). A total of 1,744 cervids were tested in the risk groups: 830 (14.2% of 5,854) road/predator killed (RK); 806 (13.8%) fallen/culled (FC); 64 (1.1%) clinical suspect animals (SUS); 44 (0.8%) hunted/slaughtered not fit for human consumption (HSNHC). The numbers of tested cervids by reporting country, management system and target group in 2021 are displayed in Table 39.

All tested cervids by the EU MS resulted negative.

Norway continued its intensified testing programme in wild and captive cervids and tested 21,670 animals in 2021, mostly semi-domesticated reindeer (28.3%), followed by wild red deer (21.3%) and wild moose (20.9%). During the 2021, two cases of CWD in wild European moose and one in a red deer were reported by Norway. The number of cervids tested in 2021 by management system, species and reporting country is displayed in Table 39. The description of the three CWD cases in 2021 is shown in Table 40. Iceland and Serbia also reported three (unspecified semi-domesticated/farmed deer) and 185 (60% farmed roe deer) cervids tested in 2021, respectively; they were all negative.

| Case type                  | Atypical scrapie | Classical scrapie | Inconclusive | Total |
|----------------------------|------------------|-------------------|--------------|-------|
| Country                    | NQ/NQ ZZ/ZZ Total | NQ/NQ NZ/NZ ZZ/ZZ Total | NQ/NQ ZZ/ZZ Total |
| FR                         | 1/1              |                   |              | 1     |
| IT                         | 2/2              | 23/23             | 1/1          | 26    |
| RO                         | 3/3              |                   |              | 3     |
| Total                      | 2/3              | 48/99             | 2/70         | 225   |

N: wild-type codon 142; Q: wild-type codon 222; options ZK, ZQ, NZ, SZ, DZ: one of the codons could not be determined; ZZ: the genotype could not be determined or was different from the available options or genotyping was not performed.
Table 39: Number of tested cervids in the EU and reporting countries by management system, species, country and target group in 2021

| Management system species(a) Country(b) and target group(c) | Semi-domesticated/farmed deer species | Wild deer species | Total |
|-------------------------------------------------------------|---------------------------------------|------------------|-------|
|                                                             | Deer European Moose Fallow deer Reindeer Roe deer Red deer White-tailed deer Sub-Total | Deer European Moose Fallow deer Reindeer Roe deer Red deer Sila deer White-tailed deer Subtotal |
| EE                                                          | FC 4 4 1 3 2 | 6 10 |
| HSHC                                                        | 1 1 1 1 1 1 |
| Total                                                       | 4 1 3 3 7 11 |
| ES                                                          | SUS 77 7 7 7 7 7 |
| RK                                                          | 13 6 19 19 |
| HSNHC                                                       | 2 2 2 2 |
| HSNC                                                       | 20 12 14 114 154 154 |
| Total                                                       | 35 12 14 121 182 182 |
| FI                                                          | RK 1 90 91 23 10 60 20 113 204 |
| FC                                                          | 3 33 1 4 41 45 14 28 23 110 151 |
| HSNHC                                                       | 5 5 10 4 14 19 |
| HSHC                                                       | 1 1 9 12 3 24 25 |
| Total                                                       | 3 1 128 1 5 138 87 24 100 50 261 399 |
| HU                                                          | SUS 1 1 1 3 4 5 |
| FC                                                          | 1 1 1 2 2 |
| Total                                                       | 1 1 2 4 1 6 7 |
| IT                                                          | SUS 1 3 4 1 19 9 29 33 |
| RK                                                          | 25 228 37 290 290 |
| FC                                                          | 3 1 3 7 2 34 119 126 |
| Total                                                       | 3 1 4 3 11 28 330 80 438 449 |
| LV                                                          | SUS 3 1 2 1 2 2 3 3 |
| RK                                                          | 1 1 1 1 1 1 1 1 |
| FC                                                          | 1 1 |
| Total                                                       | 1 1 2 2 5 5 |
| RO                                                          | RK 65 65 |
| FC                                                          | 41 41 |
| HSHC                                                        | 1,379 1,379 |
| Total                                                       | 1,485 1,485 1,485 1,485 |
### Management system species (a) and target group (c) for cervids in Europe in 2021

| Country | Semi-domesticated/farmed deer species | Wild deer species | Total |
|---------|--------------------------------------|------------------|-------|
|         | Deer | European Moose | Follow deer | Reindeer | Roe deer | Red deer | White-tailed deer | Sub-Total | Deer | European Moose | Follow deer | Reindeer | Roe deer | Red deer | Silka deer | White-tailed deer | Subtotal |
| SE      | SUS  | 11              | 5            | 16        | 16       | 244      | 244       | 197      | 51       | 6       | 187      | 475       |
|         | RK   | 244             | 1            | 1         | 5        | 1        | 1         | 1        | 2        | 198     | 251      | 251       |
|         | FC   | 247             | 38           | 288       | 129      | 51       | 6         | 187      | 51       | 6       | 187      | 475       |
|         | HSNHC| 16              | 2            | 18        | 4        | 1        | 1         | 288      | 51       | 6       | 187      | 475       |
|         | HSHC | 2,020           | 243          | 2,263     | 285      | 2        | 1         | 288      | 2        | 198     | 2,551    | 2551      |
| Total   | SUS  | 2,527           | 283          | 2,813     | 430      | 3        | 63       | 7        | 503      | 3,316   | 3,316     | 3,316     |
| Total EU|      |                 |              |           |          |          |          |          |          |         |           |           |
| IS      | FC   | 3               | 3            | 3         | 3        | 3        | 3         | 3        | 3        | 3       | 3         | 3         |
| NO      | RK   | 3               | 20           | 1         | 24       | 172      | 370       | 17       | 1,336    | 298     | 2,193     | 2,217     |
|         | FC   | 1               | 112          | 10        | 123      | 31       | 413       | 103      | 431      | 185     | 1,163     | 1,286     |
|         | HSNHC| 91              | 21           | 6,000     | 311      | 6,425    | 353       | 3,738    | 3,397    | 113     | 4,414     | 11,742    |
|         | HSHC | 91              | 21           | 6,132     | 1,321    | 6,572    | 556       | 4,521    | 3,517    | 1,880   | 4,624     | 15,098    |
| Total   | SUS  | 95              | 21           | 6,132     | 1,321    | 6,572    | 556       | 4,521    | 3,517    | 1,880   | 4,624     | 15,098    |
| Total EU|      |                 |              |           |          |          |          |          |          |         |           |           |
| RS      | SUS  | 1               | 1            | 1         | 1        | 11       | 11        | 1        | 1        | 11      | 1         | 1         |
|         | RK   | 11              | 11           | 11        | 11       | 11        | 11        | 11       | 11       | 11      | 11        | 11        |
|         | FC   | 4               | 6            | 10        | 10       | 10        | 10        | 10       | 10       | 10      | 10        | 10        |
|         | HSNHC| 20              | 20           | 20        | 20       | 20        | 20        | 20       | 20       | 20      | 20        | 20        |
|         | HSHC | 58              | 2            | 73        | 10       | 143       | 143       | 143      | 143      | 143     | 143       | 143       |
| Total   | SUS  | 62              | 21           | 111       | 10       | 185       | 185       | 185      | 185      | 185     | 185       | 185       |
| Total EU|      |                 |              |           |          |          |          |          |          |         |           |           |
| Total non-EU | 160 | 23 | 6,132 | 1123 | 331 | 6,760 | 556 | 4,521 | 3,517 | 1,880 | 4,624 | 15,098 | 21,858 |
| Total | 164 | 8 | 27 | 8,788 | 118 | 617 | 5 | 9,727 | 2,077 | 5,042 | 3,541 | 2,396 | 4,834 | 1 | 50 | 17,985 | 27,712 |

(a): Deer: not specified. Moose (or Eurasian/European elk) (*Alces alces alces*); fallow deer (*Dama dama*); Reindeer: Eurasian tundra reindeer (*Rangifer tarandus tarandus*) in Sweden; Finnish (Eurasian) forest reindeer (*Rangifer tarandus fennicus*) in Finland; roe deer (*Capreolus capreolus*); red deer (*Cervus elaphus*); white-tailed deer (*Odocoileus virginianus*); sika deer (*Cervus nippon*).

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

(c): SUS: clinical suspect animals; RK: road/predator killed; FC: fallen/culled; HSNHC: hunted/slaughtered not fit for human consumption; HSHC: hunted/slaughtered fit for human consumption.
### Table 40: Description of the CWD cases in 2021

| Country | Progressive numbering of cases throughout the year | Management system | Species | Sex | Age group | Target group\(^{(a)}\) | Part sampled | Analytical method type | Analytical method | Result |
|---------|----------------------------------------------------|-------------------|---------|-----|-----------|----------------|--------------|----------------------|------------------|--------|
| NO      | 1                                                  | Wild deer         | European moose | Male | ≥ 12 months | FC             | Obex (as part-nature) | Screening          | Bio-Rad TeSeE SAP rapid test (using the CWD protocol) | Positive          |
|         | 2                                                  | Wild deer         | European moose | Female | ≥ 12 months | FC             | Obex (as part-nature) | Screening          | Bio-Rad TeSeE SAP rapid test (using the CWD protocol) | Positive          |
|         | 3                                                  | Wild deer         | Red Deer      | Female | ≥ 12 months | HSHC           | Obex (as part-nature) | Screening          | Bio-Rad TeSeE SAP rapid test (using the CWD protocol) | Positive          |

\(^{(a)}\): Target groups: FC: fallen/culled; HSHC: hunted/slaughtered fit for human consumption.
3.4. Other species

Only one MS (Finland) and one non-EU country (Turkey) reported results of samples tested for TSE in species other than cattle, domestic sheep and goats and cervids. In total, 148 samples were collected from Finland and tested from the following species: cat (*Felis catus*), American mink (*Neovison vison*), raccoon dog (*Nyctereutes procyonoides*) and fox (genus *Vulpes*). Turkey reported results for one sample collected from one cat. None of them tested positive (Table 41).

**Table 41:** Numbers of animals in species other than cattle, sheep, goats and cervids tested for TSE in reporting countries in 2021

| Country        | Cat (*Felis catus*) | American mink (*Neovison vison*) | Fox (genus *Vulpes*) | Raccoon dog (*Nyctereutes procyonoides*) | Total |
|----------------|---------------------|----------------------------------|----------------------|-----------------------------------------|-------|
| FI             | 47                  | 56                               | 33                   | 12                                      | 148   |
| Total EU27     | 47                  | 56                               | 33                   | 12                                      | 148   |
| TR             | 1                   | 1                                |                      |                                         | 1     |
| Total other non-EU | 1                  |                                   |                      |                                         | 1     |
| Total          | 48                  | 56                               | 33                   | 12                                      | 149   |

4. Conclusions

As part of the BSE surveillance system in cattle in the EU, the EU27 and XI tested 1,021,252 cattle in 2021, 9% less than in the previous year, which cannot be considered a real decrease given the throughput reported by the United Kingdom in 2020 (134,042). The testing throughput combined with a risk-based strategy (85.2% 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 the EU27 and XI, six atypical cases of BSE (four L-BSE and two H-BSE cases) were confirmed in 2021 by three reporting countries: France (one H-BSE and two L-BSE), Germany (one L-BSE) and Spain (one H-BSE and one L-BSE). In total, 66,121 cattle were tested by eight other non-EU reporting countries, with no additional cases reported. Turkey, a country reporting data to EFSA for the first time, tested 12,098 cattle.

The situation of BSE remains similar to the one of previous years in terms of tested animals and caseload, despite the addition of Turkey and the absence of the data from the United Kingdom. The detection of H- and L-forms in both fallen stock and emergency slaughtered animals continue at a stable rate. The finding of a classical BSE case in the United Kingdom (Somerset, England) is a matter of concern. The report on the epidemiological investigation of this single BSE case (RBSE 2021:00002) concluded that ‘a number of potential source risk pathways have been identified and thoroughly investigated. The most likely source is assessed to be residual material remaining in this silo, contaminating the cattle rearer ration that was fed to the index case during the first year of its life’. The atypical cases reported by Brazil and Canada drew temporary consequences for the trade of beef last year.

In total 429,631 small ruminants were tested in 2021 in the EU27 and XI, as part of the TSE surveillance system, leading to an overall testing of more than 10.4 million tests since 2002. Twenty-two countries in the group EU27 and XI complied with the EU monitoring requirements for sheep and 23 countries for goats. Compared with 2020, there was a slight increase in the detection of the ovine IC in both CS and AS, from 169 to 176 and this occurred despite the 4.2% decrease in the level of testing in non-infected flocks. This contrasts with the trend in previous years when a continuous decrease in the overall incidence of the disease (newly infected flocks) in this species was noticed. Over the same period, the number of caprine IC (CS and AS) decreased by 57.4%, from 61 to 35, despite the stable number of animals tested in non-infected herds. The new reporting country, Turkey, did not report any tests in small ruminants.

For CS in sheep in 2021 in the EU27 and XI and compared with 2020, the caseload decreased by 23.9% as a result, partially, of a decrease in testing in TSE-infected flocks by 28.5%. The discontinuation of the 2-year intensified monitoring in scrapie affected flocks/herds with atypical scrapie may have had an impact on the testing throughput of infected flocks in the second half of the year and in the number of cases detected in this subpopulation. Three of the largest contributors reduced their caseload: Greece by 66%, Romania by 58.5% and Spain by 25.7% while Italy increased the caseloads by 45.3%. CS is reported by a minority of MS, six, less than in 2020.
In goats, in total of 219 of 224 scrapie cases reported in the EU27 and XI in 2021 were CS (97.8%). They were reported by six MS. Compared with 2020 when 319 CS cases were reported, there was a 31.3% reduction (−100) mainly due to the situation in Cyprus that has improved continuously over the last years.

When looking at the long-term trends of CS in terms of cases per 10,000 tests, the situation up to 2021 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. Genetic-based culling and outbreak management in herds based on goats carrying at least one of the known resistant alleles (i.e. K222, D146 and S146) are now applicable, following the amendment of the TSE regulation. If fully applied, the trend in CS in goats could be aligned with that of the CS in sheep in the next few years.

With regard to AS in sheep in EU27 and XI, compared with 2020, testing activity resulted in a 5.1% increase in reported cases, from 98 in 2020 to 103 in 2021, which was similar for the proportion of cases per 10,000 tested animals (active surveillance) (from 2.6 to 3.3). In goats, the AS situation has improved compared to the previous year in terms of caseload and proportion of cases per 10,000 tested animals and number of IC. 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 2021 from ovine CS cases consistently confirmed the association between the occurrence of the disease and the susceptible genotypes (NSP3, NSP3O, NSP4 or NSP5), with 98.6% of the cases with known NSP genotype carrying them. The 2021 genotyping data from random samples of the EU sheep population (data from eight MS after excluding Cyprus) showed a marginal improvement (7.9% of the genotyped sheep carrying genotypes of the susceptible group) compared with the previous years (8.8%). The NSP1 group (i.e. ARR/ARR) accounted for 66.8% 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 still showed a high proportion of susceptible sheep.

For the first time, data on genotyping of goat cases have been collected. At least one polymorphism in codons 146 or 222 was reported from 152 cases reported by three countries in the EU27 and XI. All the reported polymorphisms in 152 scrapie cases (2 atypical, 149 classical, 1 inconclusive) were N at the 146 codon and Q at the 222 codons. Thus, no goat cases carried the alleles considered resistant.

The CWD surveillance was conducted by eight MS resulting in 5,854 cervids tested and no additional cases detected. Two countries, Sweden and Romania, contributed to 82.1% of the total number of tested cervids in the EU and XI, with 56.7% (3,316) and 25.4% (1,485), respectively. Iceland and Serbia also reported 3 and 185 cervids tested in 2021, all negative. Norway continued its intensified testing programme in wild and captive cervids and tested 21,670 cervids in 2021. This activity led to the detection of three cases: two moose and one red deer.

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### Country codes

| Country                        | Code |
|-------------------------------|------|
| Austria                       | AT   |
| Bosnia and Herzegovina        | BA   |
| Belgium                       | BE   |
| Bulgaria                      | BG   |
| Croatia                       | HR   |
| Cyprus                        | CY   |
| Czechia                       | CZ   |
| Denmark                       | DK   |
| Estonia                       | EE   |
| Finland                       | FI   |
| France                        | FR   |
| Germany                       | DE   |
| Greece                        | EL   |
| Hungary                       | HU   |
| Iceland                       | IS   |
| Ireland                       | IE   |
| Italy                         | IT   |
| Latvia                        | LV   |
| Lithuania                     | LT   |
| Luxembourg                    | LU   |
| Malta                         | MT   |
| Montenegro                    | ME   |
| The Netherlands               | NL   |
| North Macedonia               | MK   |
| Norway                        | NO   |
| Portugal                      | PT   |
| Romania                       | RO   |
| Serbia                        | RS   |
| Slovakia                      | SK   |
| Slovenia                      | SI   |
| Spain                         | ES   |
| Sweden                        | SE   |
| Switzerland                   | CH   |
| Turkey                        | TR   |
| United Kingdom (in respect of Northern Ireland) | XI |

**EU27 MS countries:** AT; BE; BG; HR; CY; CZ; DK; EE; FI; FR; DE; EL; HU; IE; IT; LV; LT; LU; MT; NL; PL; PT; RO; SK; SI; ES; SE.

**Non-EU reporting countries:** BA, CH (including Lichtenstein); IS; ME; MK; NO; RS; TR; XI; XU.
## Appendix A – Additional surveillance data (Table A.1)

**Table A.1:** BSE active monitoring in relation to the adult bovine population (age >2 years) in 2021

| EU/non-EU groups | Country code | Adult cattle (>2 years)<sup>(a)</sup> | Number of tested bovine animals at risk<sup>(b)</sup> | Proportion (%) of tested bovine animals at risk<sup>(b)</sup> |
|------------------|--------------|-------------------------------------|-------------------------------------------------|-------------------------------------------------|
| **EU**           |              |                                     |                                                 |                                                 |
| AT               | 838,020      | 18,562                              | 2.2%                                            |                                                 |
| BE               | 1,143,440    | 25,787                              | 2.3%                                            |                                                 |
| BG               | 422,750      | 3,820                               | 0.9%                                            |                                                 |
| CY               | 42,700       | 2,006                               | 4.7%                                            |                                                 |
| CZ               | 654,820      | 24,840                              | 3.8%                                            |                                                 |
| DE               | 5,115,940    | 177,029                             | 3.5%                                            |                                                 |
| DK               | 684,000      | 22,870                              | 3.3%                                            |                                                 |
| EE               | 131,100      | 3,392                               | 2.6%                                            |                                                 |
| EL               | 298,000      | 2,030                               | 0.7%                                            |                                                 |
| ES               | 3,240,410    | 62,643                              | 1.9%                                            |                                                 |
| FI               | 337,830      | 9,555                               | 2.8%                                            |                                                 |
| FR               | 9,364,040    | 195,755                             | 2.1%                                            |                                                 |
| HR               | 165,000      | 5,258                               | 3.2%                                            |                                                 |
| HU               | 487,400      | 12,277                              | 2.5%                                            |                                                 |
| IE               | 2,766,380    | 59,420                              | 2.1%                                            |                                                 |
| IT               | 2,996,820    | 47,610                              | 1.6%                                            |                                                 |
| LT               | 328,400      | 3,732                               | 1.1%                                            |                                                 |
| LU               | 98,130       | 2,766                               | 2.8%                                            |                                                 |
| LV               | 218,840      | 3,373                               | 1.5%                                            |                                                 |
| MT               | 6,670        | 213                                 | 3.2%                                            |                                                 |
| NL               | 1,704,000    | 60,899                              | 3.6%                                            |                                                 |
| PL               | 2,662,100    | 48,559                              | 1.8%                                            |                                                 |
| PT               | 898,740      | 18,928                              | 2.1%                                            |                                                 |
| RO               | 1,261,000    | 10,137                              | 0.8%                                            |                                                 |
| SE               | 595,710      | 8,550                               | 1.4%                                            |                                                 |
| SI               | 195,310      | 6,424                               | 3.3%                                            |                                                 |
| SK               | 227,580      | 9,106                               | 4.0%                                            |                                                 |
| **Total EU27**   | **36,885,130** | **845,541**                          | **2.3%**                                        |                                                 |
| **XI<sup>(c)</sup>** | **nd**       | **24,787**                          |                                                 |                                                 |
| **Total EU27 + XI** | **36,885,130** | **870,328**                          | **2.36%**                                       |                                                 |
| **Other non-EU** |              |                                     |                                                 |                                                 |
| BA               | nd           | 1                                   |                                                 |                                                 |
| CH               | 771,320      | 11,050                              |                                                 |                                                 |
| IS               | 37,000       | 10                                  | 0.0%                                            |                                                 |
| ME               | nd           | 0                                   |                                                 |                                                 |
| MK               | 120,000      | 8                                   | 0.0%                                            |                                                 |
| NO<sup>(d)</sup> | 353,700      | 6,997                               | 2.0%                                            |                                                 |
| RS               | 472,000      | 4,481                               | 0.9%                                            |                                                 |
| TR               | 8,803,750    | 7                                   |                                                 |                                                 |
| **Total other non-EU** | **1,0557,770** | **22,554**                          | **0.21%**                                       |                                                 |
| **TOTAL**        | **47,442,900** | **892,882**                          | **2.57%**                                       |                                                 |

(a): Population data obtained at: https://appsso.eurostat.ec.europa.eu/nui/show.do?dataset=apro_mt_lscatl&lang=en [Bovine animals, 2 years or over]; nd = not reported by the cited source.

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

(c): Data from XI, United Kingdom (in respect of Northern Ireland) are available from 2021 onwards.

(d): Norway’s and Iceland’s cattle population taken form the TSE EUSR report 2020 (EFSA, 2021).
Appendix B – Geographical distribution of BSE in the period 2001–2021 (Figures B.1 and B.2)

The size of the circles is proportional to the measurements and are only comparable within the map but not between maps. With regard to the United Kingdom and 2021, only data from United Kingdom (in respect of Northern Ireland) (XI) have been considered.

**Figure B.1:** Geographical distribution of cumulative number of cases of C-BSE (A), BARB cases (B), H-BSE (C), and L-BSE (D) in the period 2001–2021
The size of the circles is proportional to the measurements and is only comparable within the map but not between maps. With regard to the United Kingdom and 2021, 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 2001–2021 in the EU. (A) Total classical cases per million tests; (B) Total classical BARB cases per million tests; (C) Total atypical H-type cases per million tests; (D) Total atypical L-type cases per million tests.
Appendix C – Geographical distribution of scrapie in 2021 (Figure C.1 and C.2)

The size of the circles is proportional to the measurements and is only comparable within the map but not between maps.

**Figure C.1:** Geographical distribution of numbers of cases of ovine CS (A), caprine CS (B), ovine AS (C) and caprine AS (D) in 2021 in the reporting countries.
The size of the circles is proportional to the measurements and is 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 2021
Appendix D – Additional information asked by EFSA in relation to reporting, according to Annex III of Regulation 999/2001 (Tables D.1–D.3)

Table D.1: The number of suspected cases placed under official movement restrictions in accordance with Article 12(1) in 2021

| Country | Cattle | Sheep | Goats |
|---------|--------|-------|-------|
| AT      | 11     | 3     |       |
| BE      | 242    |       |       |
| BG      |        | 6     | 35    |
| CY      |        | 8,079 | 15,665|
| CZ      | 1      |       |       |
| DE      | 2      |       |       |
| DK      |        |       |       |
| EE      |        |       |       |
| EL      | 790    | 6,818 | 676   |
| ES      | 3      | 3     |       |
| FI      |        |       |       |
| FR      | 5      | 11    | 1     |
| HR      | 64     | 532   |       |
| HU      | 13     |       |       |
| IE      | 8      |       |       |
| IT      |        | 2     |       |
| LT      |        |       |       |
| LU      | 2      |       |       |
| LV      | 14     |       |       |
| MT      |        |       |       |
| NL      |        |       |       |
| PL      |        |       |       |
| PT      |        |       |       |
| RO      | 44     | 26    | 3     |
| SE      |        |       |       |
| SI      |        |       |       |
| SK      |        |       |       |
| Total EU27 | 1,197 | 15,482 | 16,380 |
| XI(a)   | 0      | 1     | 0     |
| Total EU27 + XI | 1,197 | 15,483 | 16,380 |
| BA      |        |       |       |
| CH      | 21     | 1     | 1     |
| IS      |        | 2     |       |
| ME      |        |       |       |
| MK      |        |       |       |
| NO      | 2      |       |       |
| RS      | 16     |       |       |
| TR      |        |       |       |
| Total other non-EU | 37 | 5 | 1 |
| Total | 1,234 | 15,488 | 16,381 |

(a): Data from XI, United Kingdom (in respect of Northern Ireland) are available from 2021 onwards.
### Table D.2: Number of flocks where suspected cases in ovine and caprine animals have been reported and investigated pursuant to Article 12(1) and (2) in 2021

| Country | Sheep | Goats |
|---------|-------|-------|
| AT      | 2     |       |
| BE      |       |       |
| BG      | 8     | 4     |
| CY<sup>(a)</sup> | 5 | 17 |
| CZ      | 22    | 4     |
| DK      |       |       |
| EE      |       |       |
| EL      | 24    | 3     |
| ES      | 4     | 1     |
| FI      | 11    | 1     |
| FR      | 2     |       |
| HR      |       |       |
| HU      |       |       |
| IE      |       |       |
| IT      | 2     |       |
| LT      |       |       |
| LU      |       |       |
| LV      |       |       |
| MT      |       |       |
| NL      |       |       |
| PL      | 3     |       |
| PT      |       |       |
| RO      | 21    | 7     |
| SE      |       |       |
| SI      |       |       |
| SK      |       |       |
| **Total EU27** | **104** | **37** |
| **XI<sup>(b)</sup>** | 1 | 0 |
| **Total EU27 + XI** | **105** | **37** |
| BA      |       |       |
| CH      |       |       |
| IS      | 2     |       |
| ME      |       |       |
| MK      | 2     |       |
| NO      | 2     |       |
| RS      |       |       |
| TR      |       |       |
| **Total other non-EU** | **6** | **0** |
| **Total** | **111** | **37** |

<sup>(a)</sup>: In addition, 26 mixed flocks (sheep and goats).

<sup>(b)</sup>: Data 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 2021

| Country | Sheep SHC | Sheep NSHC | Sheep EM | Goats SHC | Goats NSHC | Goats EM | Other (a) |
|---------|-----------|------------|---------|-----------|------------|---------|-----------|
| AT      | 103       | 1,762      | 22      | 540       | n/a        | n/a     |           |
| BE(b)   | n/a       | n/a        | n/a     | n/a       | n/a        | n/a     |           |
| BG(c)   | n/a       | n/a        | n/a     | n/a       | n/a        | n/a     |           |
| CY(d)   | 563       | 324        | 104     |           |            |         |           |
| CZ      | 723       | 199        |         |           |            |         |           |
| DE      | 3,919     | 4,326      | 24      | 103       | 767        | 49      |           |
| DK(e)   |           |            |         |           |            |         |           |
| EE      | 57        | 3          |         |           |            |         |           |
| EL      | 174       | 412        | 24      | 87        | 151        | 4       |           |
| ES      | 956       | 4,475      | 68      | 922       | 1,966      | 8       |           |
| FI      | 3         | 492        |         |           |            | 55      |           |
| FR      | 5,346     | 17,728     | 21      | 4,696     | 16,764     | 2       |           |
| HR      | 994       |            |         |           |            | 243     | n/a       |
| HU      | 1,589     | 1,615      |         | 37        | 151        | 65      |           |
| IE      | 5,055     | 6,985      |         |           |            | 39      |           |
| IT      | 4,383     | 6,189      | 46      | 3,851     | 3,868      | 28      |           |
| LT(f)   |           |            |         | 139       | 22         | n/a     |           |
| LU(f)   | n/a       | n/a        |         |           |            |         |           |
| LV      | 288       |            |         |           |            | 15      |           |
| MT      | 95        |            |         |           |            |         |           |
| NL      | 1,888     |            |         |           |            |         |           |
| PL      | 1,074     | 3,687      | 132     | 1,548     |            |         |           |
| PT      | 926       | 5,668      | 2       | 856       |            |         |           |
| RO      | 1,068     | 33         |         |           |            |         |           |
| SE      |            | 1,615      |         |           |            |         |           |
| SI      | 45        | 1,332      | 18      | 374       |            |         |           |
| SK      | 459       | 2          |         |           |            |         |           |
| XI(g)   |           |            |         |           |            |         |           |

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

(a): Monitoring 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).
(b): The Belgian competent authority informed that in the central database for sheep and goat identification, there is no direct link between the official ear tag 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.
(c): The Bulgarian competent authority informed that no data were available for sheep and goats.
(d): The 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).
(e): The Danish competent authority informed that in the central database for sheep and goat identification, there is no direct link between the official ear tag number and the last holding where the sheep or goat was kept.
(f): The Luxembourg competent authority informed that for NSHC sheep and NSHC goats, the number of flocks is unknown.
(g): Data 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 link to the data sets. The countries that submitted data sets on the 2021 monitoring data year are the 27 EU Member States, XI and eight other non-EU reporting countries (Table E.1).

Table E.1: Links to the TSE data sets for 2021 by reporting country

| Country       | Link to the data set                              |
|---------------|--------------------------------------------------|
| EU27 Member States |                                               |
| AT            | https://doi.org/10.5281/zenodo.4091585            |
| BE            | https://doi.org/10.5281/zenodo.4091591            |
| BG            | https://doi.org/10.5281/zenodo.4091602            |
| CY            | https://doi.org/10.5281/zenodo.4091604            |
| 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            |
| IT            | 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            |
| MT            | https://doi.org/10.5281/zenodo.4091678            |
| NL            | https://doi.org/10.5281/zenodo.4091684            |
| PL            | https://doi.org/10.5281/zenodo.4091686            |
| PT            | 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-EU  |                                               |
| BA            | https://doi.org/10.5281/zenodo.5652824            |
| CH            | https://doi.org/10.5281/zenodo.4091716            |
| IS            | https://doi.org/10.5281/zenodo.4091719            |
| ME            | https://doi.org/10.5281/zenodo.4091723            |
| MK            | https://doi.org/10.5281/zenodo.4091725            |
| NO            | https://doi.org/10.5281/zenodo.4091727            |
| RS            | https://doi.org/10.5281/zenodo.4091729            |
| TR            | https://doi.org/10.5281/zenodo.7257583            |