SHEEP AND GOATS SCRAPIE SURVEILLANCE 2013
JOINT DESCRIPTIVE REPORT FOR GREAT BRITAIN

Animal Health and Veterinary Laboratories Agency

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List of acronyms and abbreviations

AC	Annual Cull
AH	Animal Health
AHVLA	Animal Health and Veterinary Laboratories Agency
ASM	Atypical Scrapie Monitoring
AS	Abattoir Survey
BSE	Bovine Spongiform Encephalopathy
CSFS	Compulsory Scrapie Flocks Scheme
DA	Devolved Administrations
Defra	Department of Environment, Food and Rural Affairs
DIT	Dead In Transit
EC	European Commission
EFSA	European Food Safety Authority
EU	European Union
EURL	European Union Reference Laboratory
FS	Fallen Stock
GB	Great Britain
IC	Initial Cull
IHC	Immunohistochemistry
LRT	Lymphoreticular tissue
NSPAC	National Scrapie Plan Administration Centre
OIE	World Organisation for Animal Health
SEAC	Spongiform Encephalopathy Advisory Committee
SND	Scrapie Notification Database
TSE	Transmissible Spongiform Encephalopathy
VLA	Veterinary Laboratories Agency

1. Executive summary

Sheep scrapie surveillance in 2013

- This was the third successive year where no clinical cases of classical scrapie had been confirmed since the disease became notifiable in 1993. However there was a clinical case of atypical scrapie confirmed, the ninth case ever and the first since 2007. It was a 10-year old female Ryeland of the ARR/ARQ genotype, reported by a farm owner in England. The number of clinical suspects remained very low with four notifications from four different holdings.
- No cases of classical scrapie were confirmed in the Abattoir survey (AS) in 2013. Although the estimated prevalence of infection of classical scrapie in the GB sheep population in 2013 using AS data was 0% (95% CI: 0% – 0.13%), the uncertainty of the parameter estimated up to 0.13% infection in the sheep population, which was not significantly different than that of previous years.
- The quota allocated in 2013 by the EU for the number of samples to be tested by active surveillance (FS and AS) was the same as in the previous year, 9300 in the FS and 9400 in the AS. The compensation of the diminishing number of abattoir survey samples with fallen sheep for cost-saving purposes continued in 2013.
- Only six cases of classical scrapie were confirmed in 2013: three cases by the Fallen Stock Survey which tested 31.7% in excess of the required quota, two fallen stock cases by the Compulsory Scrapie Flocks Scheme (CSFS) and one fallen stock case by the Atypical Scrapie Monitoring (ASM). A holding in the ASM had one case of classical and one case of atypical scrapie confirmed within a year.
- The number of cases of atypical scrapie reached 17 in 2013, confirmed in sheep submitted to the Fallen Stock survey (12), the Abattoir survey (3), dead in transit (1) and clinical cases (passive surveillance (1). The FS remained the main contributor to the detection of atypical scrapie but probably due to sampling variability rather than significant changes in the prevalence of infection in each of the populations tested by these two surveys.
- Despite the apparent decreasing trend observed in the proportion of atypical cases confirmed by both the AS and FS in 2013 compared to the previous year, there were no significant changes observed year by year and overall since 2003, with an average proportion of 0.1% of positive cases in both routes. At the current scenario of low prevalence, the level of testing precludes the detection of significant changes in the trend year by year.
- Changes to the operation of the FS survey introduced in 2011 including the elimination of the free collection of carcasses and the increase in the number of sampling sites continued to deliver beneficial effects in 2012 in terms of coverage and representativeness. It reduced

the proportion of holdings contributing very large number of samples and the unsuitability rate. The number of holdings contributing a single carcase was 52.5%, slightly lower than in the previous two years. England and Wales were over-represented in this survey, with Scotland being severely under-represented with the lowest contribution of samples and holdings ever.

- The contribution of the Atypical Scrapie Monitoring (ASM) to the testing throughput of sheep holdings under restrictions by any kind of scrapie increased substantially in 2013. Twenty-three holdings joined the existing 18 monitored due to confirmation of atypical scrapie. Thirty-two of the 41 holdings under restrictions submitted 1013 samples to the various testing routes of the scheme. No additional cases of atypical scrapie were detected but one case of classical scrapie was confirmed in one of the holding members.
- Four new holdings joined the existing 10 monitored due to confirmation of classical scrapie in the Compulsory Scrapie Flocks Scheme (CSFS) in 2013. Six of 14 holdings under restrictions submitted 1135 samples to the various testing routes of the scheme, with two classical cases confirmed in the same holding.
- There were no significant changes in the genotype profile of the classical and atypical sheep scrapie cases confirmed in 2013.

Goat scrapie surveillance in 2013

- Two purchased goats were reported as clinical suspects by an unregistered holding in England. The two goats were confirmed positive to classical scrapie with no results of tracing available at the time of writing this report. Thus no statutory action was taken in 2013 on that or any other holding/s due to passive surveillance.
- Fifteen other cases were confirmed in goats submitted under the different testing routes of the CSFS: Fallen Stock (8), the Annual Cull (7). All cases came from two holdings under restrictions, one of them with a high proportion of positive cases (2.3% of tested samples). Despite the difference in population sizes of the sheep and goats in GB, cases confirmed in goats outnumbered those confirmed in sheep.
- The only survey of active surveillance (FS) did not detect any case of scrapie. England was over-represented in the FS survey, with significantly greater numbers of samples and holdings submitting goats for testing than expected, Wales and Scotland were under-represented in 2013. No cases were confirmed by this survey.
- To date there has still not been any case of atypical scrapie confirmed in goats in Great Britain. A case of co-infection with BSE was suspected on the basis of molecular and pathology characteristics. The bioassay results did not confirm the suspicion of BSE.

2. Sheep scrapie surveillance

2.1 Background

Scrapie became a notifiable disease on January 1st 1993, as required by Council Directive 91/68/EEC. With the implementation in July 1998 of the Sheep and Goat Spongiform Encephalopathy Order and the Sheep and Goat Spongiform Encephalopathy Regulations, epidemiological investigations started on all premises in Great Britain (GB) where a suspect case of scrapie was reported. Recording of data started in a new database, the Scrapie Notifications Database (SND), held at the Veterinary Laboratories Agency (VLA), since 1 April 2011 known as the Animal Health and Veterinary Laboratories Agency (AHVLA).

In January 2002 Defra, on behalf of England and Wales, and Scotland began a programme of active surveillance for sheep scrapie. This arose as a result of Regulation (EC) No. 999/2001 and the recommendation of the Spongiform Encephalopathy Advisory Committee (SEAC) to estimate the prevalence of sheep scrapie in the British sheep flock. Surveys on the slaughtered population, i.e. the abattoir survey (AS), and the fallen stock on farm, i.e. the Fallen Stock survey (FS), were conducted throughout each year as per required EU quotas. Following Commission Regulation (EC) 1915/2003 amending Regulation (EC) 999/2001, Defra and the Devolved Administrations (DAs) in Scotland and Wales launched the Compulsory Scrapie Flocks Scheme (CSFS) respectively in England and Scotland on July 20th 2004 and in Wales on November 1st 2004. The scheme introduced compulsory eradication measures in sheep flocks and goat herds in which classical scrapie was confirmed, to meet European law requirements.

Since 2005, the AHVLA implemented one of two options in scrapie flocks where a scrapie case could be traced or linked to that flock: genotyping with killing and destruction of Type 3 and Type 5 genotypes in rams and ewes, with Type 2 and Type 4 genotype rams allowed/directed to be slaughtered for human consumption[1], ruling out the second option, except in exceptional circumstances: the killing and destruction of the entire flock without official genotyping. In both options, a sample of the culled sheep over 12 months of age was tested for Transmissible Spongiform Encephalopathies (TSEs) (Initial Cull or CSFS-IC). Flocks then entered a three, and later two year restriction period during which all fallen stock (CSFS-FS) over 18 months of age had to be submitted for TSE rapid testing, and re-stocking was only permitted with animals of resistant genotypes. A sample of all animals over 18 months of age slaughtered for human consumption was tested annually for TSEs during the restriction period (CSFS-AC). In the course of 2011, control measures in CSFS flocks were relaxed: the whole flock cull or the whole flock genotype and selective cull options were replaced, in most instances, by a monitoring option whereby movement restrictions are imposed and surveillance is maintained on the flock for two years after the last recorded case. Animals over 18 months of age can be

[1] There is no compulsion to slaughter Type 2 ewes immediately: they can be retained for breeding before going to slaughter, or sold to other CSFS flocks.

sent for slaughter and human consumption, subject to a negative TSE test and all fallen stock over 18 months of age has to be TSE tested.

Since October 10th 2011 Defra implemented, through the AHVLA, an enhanced monitoring programme on farms where only cases of atypical scrapie had been confirmed, known as Atypical Scrapie Monitoring (ASM). This programme was not applied retrospectively and only holdings where cases were confirmed from that date will be subject to TSE monitoring. The controls are based on paragraph 5(b) of Chapter A of Annex VII to Commission Regulation (EC) No. 999/2001, as amended by Commission Regulation (EC) No. 746/2008.

Owners of Atypical Scrapie Monitoring (ASM) flocks/herds are required to report all fallen stock over 18 months (ASM-FS) during the two year restriction period to be TSE tested and to submit for testing any culls over 18 months of age which are moved directly from the farm to an abattoir (ASM-AC). Sheep or goats sent to slaughter via a market, dealer or third party are exempt from the testing requirement. The results of the tests conducted under the ASM programme are reported separately after the CSFS section.

In July 2013 Commission Regulation (EU) No 630/2013 amending Annexes VII and VIII of EU Regulation 999/2001 came into force. This new piece of legislation a) clarifies the conditions under which the three control measures are applied, namely, the whole flock cull, the genotype & cull and the enhanced surveillance; b) removes the requirement for movement restrictions where a case of atypical scrapie has been confirmed; c) brings requirements for Intra-Community trade in line with those recommended by the World Organisation for Animal Health (OIE); d) and lays out a framework enabling the Member States to establish an official scheme for the recognition of classical scrapie status.

This 2013 report includes the individual descriptive analysis of the available testing routes of scrapie in sheep in GB (Section 2): passive surveillance (via the Scrapie Notification Database; SND), Fallen Stock (FS), Dead in Transit (DIT), Abattoir survey (AS) and the Compulsory Scrapie Flocks Scheme (CSFS) with its four testing options (clinical suspects, over 12 months from the initial cull, over 18 month annual cull and fallen stock) and the Atypical Monitoring Scheme (ASM) with its three testing routes (clinical suspects and over 18 month annual cull and fallen stock) . Classical and atypical scrapie figures are provided at the individual sheep level for the passive and three active surveillance sources, namely, SND, FS, DIT and AS, and for the testing routes of the CSFS (Clinical, IC, FS and AC) and ASM (Clinical, FS and AC). The results of the 2013 data are presented for each source including samples, and where possible, accounting for the lack of traceability of some active cases, holdings in GB, and by country and the description of cases by type and genotype. When possible, figures for 2013 have been compared with those of previous years, with a focus on the period 2002-2013 for comparative analysis and detection of trends. A further section describes jointly the results of all sources where comparisons allow.

The report includes for the fourth year a descriptive analysis of the testing routes in goats (Section 3). The executive summary includes the main conclusions of the descriptive analysis

and evolution of scrapie in GB based on surveillance data, including passive surveillance, the FS and the different routes of the CSFS. Since no cases of atypical scrapie have been confirmed in goats, there is no ASM scheme in this species.

2.2 Passive surveillance

The total number of clinical suspects reported during 2013 was four from four different identified holdings, two in England and two in Scotland. All of them were declared clinical suspect and tested with the result of one being confirmed as atypical scrapie. The numbers of reported, tested and confirmed cases in sheep from 2002 to 2013 are shown in Table 1. The counts per month of reported and confirmed cases in GB for the period 2002-2013 are shown in Figure 1. The six-month moving average series has also been plotted for a better visualization of the general trends.

Table 1 Number of cases reported, tested and confirmed by passive surveillance in sheep from 2002 to 2013

Year	Reported	Tested	Confirmed	% Reported not confirmed (95% CI)	% Tested not confirmed (95% CI)	Classical	% Classical (95% CI)	Atypical	% Atypical (95% CI)
2002	549	508	403	26.59% (23.07 – 30.45)	20.67% (17.37 - 24.42)	403	79.33% (75.57 – 82.62)	0	0.00% (0.00 – 0.7)
2003	519	479	379	26.97% (23.33 – 30.96)	20.87% (17.47 - 24.75)	379	79.12% (75.2 4– 82.52)	0	0.00% (0.00 – 0.77)
2004	487	425	307	36.96% (32.79 – 41.33)	27.77% (23.72 - 32.21)	307	72.23% (67.78 – 76.27)	0	0.00% (0.00 – 0.86)
2005	412	331	179	56.55% (51.72 – 61.25)	45.92% (40.63 - 51.30)	177	53.47% (47.79 –58.47)	3	0.90% (0.19 – 2.79)
2006	264	205	100	62.12% (56.12 – 67.75)	51.22% (44.4 – 58)	97	47.31% (40.6 – 54.1)	3	1.46% (0.32 – 4.46)
2007	69	45	13	81.16% (70.18 – 88.72)	71.11% (56.5 – 82.3)	11	24.44% (9.16 – 38.9)	2	4.44% (0.5 – 15)
2008	15	8	1	93.3% (68.16-99.99)	87.5% (50.8 – 99.9)	1	12.5% (0.11-49.22)	0	0% (0.00- 37)
2009	9	9	3	78.5% (49.2 – 95.3)	66.6% (29.9 – 92.5)	3	33.3% (7.5 – 70)	0	0% (0.00 – 33.6)
2010	3	2	0	100% (29.2 – 100)	100% (15.8 – 100)	0	0% (0.0 – 84.2)	0	0% (0.00 – 84.2)
2011	7	4	3	57.9% (18.41 – 90.1)	25% (0.6 – 80.6)	3	75% (19.4 – 99.4)	0	0% (0.00 – 60.2)
2012	5	5	0	100 (47.8 – 100)	100% (0 – 52.1)	0	0% (0 – 52.1)	0	0% (0 – 52.1)
2013	4	4	1	75% (19.4 – 99.4)	75% (19.4 – 99.4)	0	0% (0 – 60.2)	1	25% (0.6-80.6)

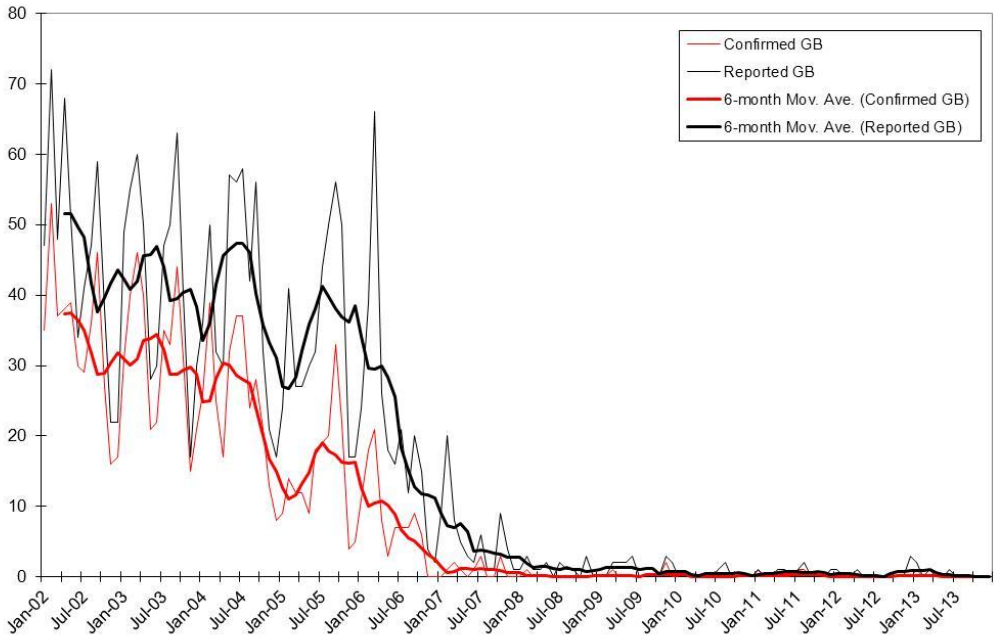


Figure 1 Counts per month of confirmed and reported cases in GB for the period Jan-2002 to Dec-2013. The 6-month moving average curves are shown to help to appreciate longer trends.

The pattern of reporting of clinical suspects remained consistent with that observed in the last five years. This was the third successive year where no clinical cases of classical scrapie had been confirmed since the disease became notifiable in 1993. However there was a case of atypical scrapie confirmed, the ninth case ever and the first since 2007. This can be considered a very rare event. The animal in question was a 10-year old female Ryeland of the ARR/ARQ genotype, reported by a farm owner in England where the animal had lambed four times. It was traced back to another holding in Wales. It is not clear whether this was the natal holding or not. The clinical signs observed during examination were: gait change, falling and loss of coordination. Additional reported neurological signs included: trembling, staring/vacant, repeated isolation from the flock and hiding behaviour.

The genotype distributions of classical scrapie cases confirmed in sheep by passive surveillance in the period 2002 – 2013 are displayed by individual genotype and NSP type[2] in Table 2 and Figure 2. Allelic variation at codons 136, 141, 154 and 171 gives rise to 21 different genotypes (ARQ denoting the wild type genotype ALRQ). The atypical case confirmed by passive surveillance 2013 was of the ARR/AFRQ genotype. A summary of the genotype distributions of atypical scrapie by individual genotype and NSP type over the period 2002-2013 is displayed in Table 3 and Figure 3.

[2] NSP types: Type 1 (ARR/ARR), Type 2 (ARR/AHQ, ARR/ARH, ARR/AFRQ, ARR/ARQ), Type 3 (AHQ/AHQ, AHQ/ARH, AHQ/AFRQ, AHQ/ARQ, ARH/ARH, ARH/AFRQ, ARH/ARQ, AFRQ/AFRQ, AFRQ/ARQ, ARQ/ARQ), Type 4 (ARR/VRQ) and Type 5 (AHQ/VRQ, ARH/VRQ, AFRQ/VRQ, ARQ/VRQ, VRQ/VRQ)

Table 2 Genotype distribution of the classical scrapie confirmed cases in sheep by passive surveillance from 2002 to 2013[3]

Genotype	2002-2012		2013	Total	
ARR/ARR					
ARR/AHQ					
ARR/ARH					
ARR/AFRQ					
ARR/ARQ	3	0.2%		3	0.2%
AHQ/AHQ	5	0.4%		5	0.4%
AHQ/ARH					
AHQ/AFRQ					
AHQ/ARQ	26	1.9%		26	1.9%
ARH/ARH	14	1.0%		14	1.0%
ARH/AFRQ					
ARH/ARQ	27	2.0%		27	2.0%
AFRQ/AFRQ					
AFRQ/ARQ	1	0.1%		1	0.1%
ARQ/ARQ	267	19.3%		267	19.3%
ARR/VRQ	56	4.1%		56	4.1%
AHQ/VRQ	2	0.1%		2	0.1%
ARH/VRQ	69	5.0%		69	5.0%
AFRQ/VRQ					
ARQ/VRQ	589	42.7%		589	42.7%
VRQ/VRQ	197	14.3%		197	14.3%
Unknown	125	9.1%		125	9.1%
Total	1381	100%	0	1381	100%

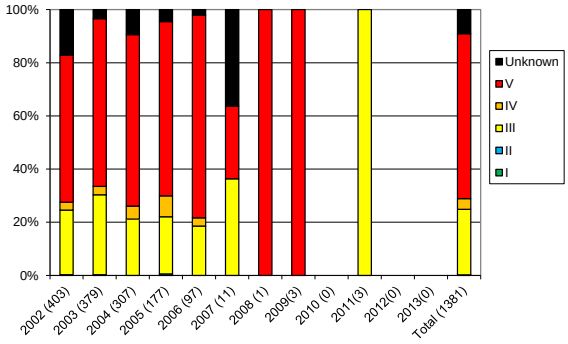


Figure 2 Percentage genotype distribution of NSP types of classical scrapie confirmed cases in sheep by passive surveillance from 2002 to 2013

Table 3 Genotype distribution of the atypical scrapie confirmed cases in sheep by passive surveillance from 2002 to 2013[4]

Genotype	2002-2012		2013		Total	
ARR/ARR						
ARR/AHQ	2	25%			2	22.2%
ARR/ARH						
ARR/AFRQ			1	100%	1	11.1%
ARR/ARQ						
AHQ/AHQ	1	12.5%			1	11.1%
AHQ/ARH						
AHQ/AFRQ						
AHQ/ARQ	4	50%			4	44.4%
ARH/ARH						
ARH/AFRQ						
ARH/ARQ						
AFRQ/AFRQ						
AFRQ/ARQ						
ARQ/ARQ						
ARR/VRQ						
AHQ/VRQ						
ARH/VRQ						
AFRQ/VRQ						
ARQ/VRQ						
VRQ/VRQ						
Unknown	1	12.5%			1	11.1%
Total	8	100%	1	100%	9	100%

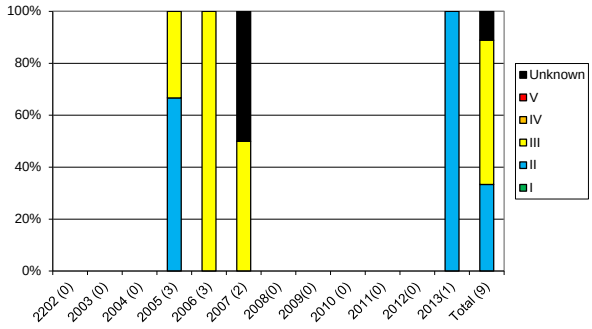


Figure 3 Percentage genotype distribution of NSP types of atypical scrapie cases in sheep confirmed by passive surveillance from 2002 to 2013

[3] Please note that ALRQ (ARQ) and AFRQ alleles were not discriminated in testing of all surveillance routes before 2007

[4] Please note that ALRQ (ARQ) and AFRQ alleles were not discriminated in testing of all surveillance routes before 2007

2.3 Fallen Stock Survey (FS)

In 2013 the EU requirement for testing fallen sheep over 18 months of age was set at 10,000 for the UK (with internal targets of 9,300 from GB and 700 from Northern Ireland). As with previous years, the quota was made up from fallen stock animals sampled at designated disposal sites. Twenty disposal sites participated in the survey and sample quality was monitored by AHVLA staff.

As the survey progressed, sampling rates (animals accepted per week) were regularly reviewed: volumes were typically at their highest in the first half of the year (up to 2,907 in March) coincident with lambing and the move of hill sheep to lower ground. Summer months and December are the least active for different reasons: the summer recess and the completion of the target before the end of the year. For example, 547 samples were submitted in August and only 31 in December.

A total of 12,246 samples from fallen sheep were submitted in GB during 2013, of which 12,227 (99.85%) were suitable for testing (Table 4). Thus the number suitable for testing exceeded the EU target for GB by 31.7%. The survey confirmed 12 atypical scrapie cases (0.1%) and three cases of classical scrapie (0.02%). The atypical cases came from eight holdings in England, three in Wales and one in Scotland. The classical cases originated from two holdings in England and one in Wales.

Table 4 FS data summary from sheep for the period 2002-2012

Year	Samples	Tested	Unsuitable	Clas.	% Clas. (95% CI)	Atyp.	% Atyp. (95%CI)	Ratio clas./aty.
2002	1066	903	163 (15.3%)	6	0.6 (0.24 – 1.4)	0	0 (0.00 – 0.41)	
2003	4282	3529	753 (17.6%)	8	0.22 (0.10 – 0.45)	3	0.08 (0.01 – 0.26)	2.6:1
2004	5018	3919	1099 (21.9%)	8	0.20 (0.09 – 0.41)	4	0.1 (0.03 – 0.27)	2:1
2005	9208	7644	1564 (17%)	25	0.32 (0.22 – 0.48)	5	0.06 (0.03 – 0.11)	5:1
2006	25804	17989	7815 (30.3%)	31	0.17 (0.12 – 0.24)	11	0.06 (0.03 – 0.11)	2.8:1
2007	18532	12670	5862 (31.6%)	17	0.13 (0.08 – 0.21)	10	0.08 (0.04 – 0.9)	1.7:1
2008	12377	10128	2249 (18.2%)	4[1]	0.04 (0.01 – 0.1)	4	0.04 (0.01 – 0.1)	1:1
2009	10478	9941	537 (5.1%)	2	0.02 (0.00 – 0.07)	8	0.08 (0.03 – 0.16)	1:4
2010	10268	10044	224 (2.2%)	0	0 (0.00 – 0.04)	6	0.08 (0.03 – 0.13)	-
2011	11973	11940	33 (0.28%)	3	0.03 (0.01 – 0.07)	11	0.09 (0.05 – 0.16)	1:3.7
2012	12916	12871	45 (0.35%)	2	0.02 (0.00 – 0.06)	17	0.13 (0.08 – 0.21)	1:8.5
2013	12246	12227	19 (0.15%)	3	0.02 (0.00 – 0.07)	12	0.1 (0.05 – 0.17)	1:4

[1]Two inconclusive results were not included

By country and taking into account only the submissions with known holding of origin, the annual incidence rate of classical scrapie in Wales and England was very similar and not statistical different than that of Scotland (Table 5). For atypical scrapie the incidence rate in Wales (0.06%) was half the incidence in England and much higher than in Scotland. However

the contribution of the samples in the FS from holding in Scotland was very low with resulting in a large degree of uncertainty in the estimates for this country,

Table 5 FS data summary from sheep by country in 2012

Country	Samples	Tested	Unsuitable	Clas.	% Clas. (95% CI)	Atyp.	% Atyp. (95%CI)
England	6523	6514	9	2	0.03 (0 – 0.14)	8	0.12 (0.05-0.24)
Scotland	411	408	3	0	0 (0 – 0.38)	1	0.01 (0 – 1.36)
Wales	5226	5219	7	1	0.02 (0 – 0.11)	3	0.06 (0.01 – 0.17)
Total	12160	12141	19	3	0.02 (0.001 – 0.07)	12	0.1 (0.05 – 0.17)

A total of 12,160 samples were supplied with CPH details which enabled the country of origin of the submitting holding to be readily ascertained. The 86 samples that could not be linked to a recognizable CPH tested negative for scrapie. By country the contribution of samples from holdings in England decreased substantially in 2013 to 53.6% of the total throughput, an absolute decrease of 18.7% on 2012 and of 12.2% on 2011 (Table 6). This was accompanied by a drastic increase of the proportion of samples from Welsh holdings, from 19.9% in 2012 to 43% the reporting year. This was the year with the lowest contribution of samples from holdings in England and with the largest contribution from holdings in Wales since the beginning of active surveillance in 2002. Scottish farms accounted for 3.4% of all submitted samples, the lowest ever. Comparisons with the national proportions of sheep in the three countries were made using data from the Sheep and Goat Inventory 2013 (49.9%, 23.8% and 26.3% of the sheep in England, Scotland and Wales, respectively). The one-way test for proportions showed that holdings in England and Wales were significantly over-represented and holdings in Scotland were under-represented at the 0.05 significance level. (http://www.defra.gov.uk/food-farm/animals/movements/sheep/).

Table 6 Summary of the FS sheep samples per country for the period 2002-2013

Year	Samples	England	%	Scotland	%	Wales	%
2002	1066	762	71.5%	172	16.1%	121	11.4%
2003	4282	3373	78.8%	623	9.5%	270	6.3%
2004	5018	3536	70.5%	388	7.7%	1083	21.6%
2005	9208	5390	58.5%	1751	19%	2065	22.4%
2006	25804	17100	66.3%	2496	9.7%	6153	23.8%
2007	18532	12304	66.4%	1791	9.7%	4384	23.7%
2008	12377*	8856	71.5%	1077	8.7%	2430	19.6%
2009	10478	6753	64.4%	945	9%	2780	26.5%
2010	10268	5555	54.1%	774	7.5%	3939	38.4%
2011	11973**	7211	65.8%	1132	10.3%	2612	23.8%
2012	12916***	9219	72.3%	988	7.7%	2544	19.9%
2013	12160****	6523	53.6%	411	3.4%	5226	43%

*CPHs for 9 samples could not be ascertained in 2008
**CPHs for 1018 samples could not be ascertained in 2011
***CPHs for 165 samples could not be ascertained in 2012
****CPHS for 86 samples could not be ascertained in 2013

The trend observed in 2011 and 2012 when the largest number of holdings submitting samples to this survey were recorded has reverted in 2013 to the levels observed in 2007, although still nearly three-fold higher than in 2009 and 2010 (Table 7). The contribution of holdings in England and Scotland was lower than in the previous year: 62.8% and 4.9%, respectively (It is important to note that this was the lowest contribution of holdings in Scotland to the FS survey ever), whereas Wales increased its contribution from 18.5% to 32.2%. Comparisons with the national proportions of sheep holdings in the three countries were made using data from the Sheep and Goat Inventory 2013 (60.7%, 19.1% and 20.2% of the active sheep holdings in England, Scotland and Wales, respectively). The one-way test for proportions showed that holdings in England and Wales were significantly over-represented and holdings in Scotland were under-represented at the 0.05 significance level.

Table 7 FS holdings summary per country for the period 2002-2013

Year	Holdings	England	%	Scotland	%	Wales	%
2002	785	557	71.0%	136	17.3%	92	11.7%
2003	1455	1100	75.6%	241	16.6%	114	7.8%
2004	1501	1035	69.0%	191	12.7%	275	18.3%
2005	2441	1643	67.3%	368	15.1%	427	17.5%
2006	5405	3808	70.5%	488	9.0%	1109	20.5%
2007	4580	3188	69.6%	454	9.9%	922	20.1%
2008	2853	2153	75.5%	228	8%	472	16.5%
2009	1843	1375	74.6%	146	7.9%	322	17.5%
2010	1765	1118	63.3%	121	6.9%	526	29.8%
2011	5671	3673	64.8%	683	12.0%	1315	23.2%
2012	6016	4304	70.4%	601	10%	1111	18.5%
2013	4965	3122	62.8%	245	4.9%	1598	32.2%

The average number of submitted samples per holding in England was 2.4 (median: 1, range: 1-63), lower than that in Wales (mean: 3.2, median: 2, range: 1-63) but higher than Scotland (mean: 1.6, median: 1, range: 1-9). The proportion of individually-identified holdings that submitted a single sheep to the FS survey decreased in 2013 (52.5%) when compared to 2012 (57.2%) and 2011 (59.2%), but still higher than in earlier years. At the other extreme, two holdings submitted more than 40 sheep and five submitted more than 30. The change already observed in 2011 and 2012 was brought about by the wider coverage of the larger number of sampling sites in terms of geographical areas and accessibility to submitting holdings. This rectifies a potential weakness of the survey in previous years when a few holdings contributing excessively to the FS survey risked impairing the ability of the survey to detect new cases. Table 8 shows the distribution of holdings by number of sheep submitted to the survey in the last six years.

Table 8 Distribution of holdings by number of sheep submitted to the FS survey for the period 2008-2013*

Number of sheep submitted	Number of holdings					
	2008	**2009**	**2010**	**2011**	**2012**	**2013**
1	1150	651	555	3356	3443	2607
2	487	257	249	1208	1246	1023
3	283	175	173	485	530	476
4	188	145	147	236	281	259
5	139	91	92	149	162	159
6	96	68	77	79	114	121
7	81	68	61	48	65	78
8	71	56	54	33	40	66
9	45	42	45	20	24	46
10	32	33	46	13	20	20
11-20	194	161	175	36	79	91
21-30	57	44	54	8	9	14
31-40	21	26	17	0	4	3
41-50	3	17	6	0	0	1
>50	7	9	14	0	0	1

* 4965 individually identified holdings contributed to FS survey in 2013

The numbers of positive cases of classical and atypical scrapie confirmed by the FS survey (as proportions of all tested samples) in the years 2002 – 2013 are shown in Figure 4. No significant difference was observable in the proportion of classical cases detected since 2006, despite the decreasing trend. The proportion of all tests that disclosed atypical cases was also unchanged despite the increasing trend observed in the last three years, probably due to the increase in the number of samples tested. At the current scenario of low prevalence, the level of testing precludes the detection of significant changes in the trend year by year, unlike the early years of implementation. Note that the 95% confidence intervals overlap between all consecutive years.

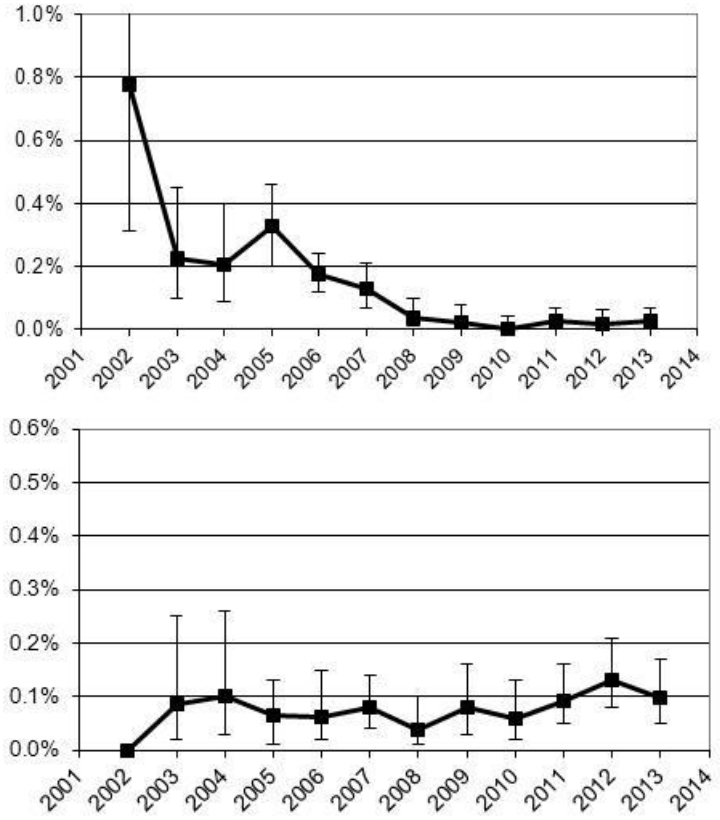


Figure 4 Prevalence estimates of scrapie in GB from the FS survey with 95% confidence intervals. Period 2002-2013.
Above: classical. Below: atypical. Note: the range in the Y-axis is different in the two graphs.

Of the three confirmed cases of classical scrapie, one was ARH/ARQ, the second ARQ/ARQ. A third case was of unknown genotype. Eleven out of the 12 confirmed atypical scrapie cases were suitable for genotype testing. Two of the eleven confirmed atypical cases in 2013 had the ARR/ARR genotype, three cases of ARR/AHQ and AHQ/AHQ each, and one case each of the AFRQ/AFRQ, ARR/AFRQ and AHQ/ARQ genotypes. The genotype distributions of classical and atypical scrapie in sheep by individual genotype and NSP type for the period 2002-2013 are shown in Tables 9 and 10 and Figures 5 and 6, respectively.

Table 9 Genotype distribution of the classical scrapie confirmed cases in sheep by the FS survey from 2002 to 2013

Genotype	2002-2012		2013		Total	
ARR/ARR						
ARR/AHQ						
ARR/ARH						
ARR/AFRQ						
ARR/ARQ						
AHQ/AHQ						
AHQ/ARH						
AHQ/AFRQ						
AHQ/ARQ	2	1.9%			2	1.8%
ARH/ARH	1	0.9%			1	0.9%
ARH/AFRQ						
ARH/ARQ	2	1.9%	1	33.3%	3	2.8%
AFRQ/AFRQ						
AFRQ/ARQ	2	1.9%			2	1.8%
ARQ/ARQ	20	18.9%	1	33.3%	21	19.3%
ARR/VRQ	10	9.4%			10	9.2%
AHQ/VRQ		0.0%				
ARH/VRQ	8	7.5%			8	7.3%
AFRQ/VRQ	1	0.9%			1	0.9%
ARQ/VRQ	48	45.3%			48	44.0%
VRQ/VRQ	10	9.4%			10	9.2%
Unknown	2	1.9%	1	33.3%	3	2.8%
Total	106	100%	3	100%	109	100%

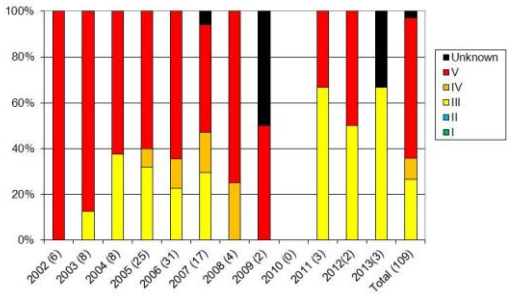


Figure 5 Percentage genotype distribution of NSP types of classical scrapie confirmed in sheep by the FS survey from 2002 to 2013

Table 10 Genotype distribution of the atypical scrapie confirmed cases in sheep by the FS survey from 2002 to 2013

Genotype	2002-12		2013		Total	
ARR/ARR	5	6.3%	2	16.6%	7	7.7%
ARR/AHQ	24	30.4%	3	25%	27	29.7%
ARR/ARH	1	1.3%			1	1.1%
ARR/AFRQ	2	2.5%	1	8.3%	3	3.3%
ARR/ARQ	2	2.5%			2	2.2%
AHQ/AHQ	15	19.0%	3	25%	18	19.8%
AHQ/ARH	1	1.3%			1	1.1%
AHQ/AFRQ	2	2.5%			2	2.2%
AHQ/ARQ	10	12.7%	1	8.3%	11	12.1%
ARH/ARH						
ARH/AFRQ	1	1.3%			1	1.1%
ARH/ARQ						
AFRQ/AFRQ	4	5.1%	1	8.3%	5	5.5%
AFRQ/ARQ	3	3.8%			3	3.3%
ARQ/ARQ	5	6.3%			5	5.5%
ARR/VRQ						
AHQ/VRQ	1	1.3%			1	1.1%
ARH/VRQ						
AFRQ/VRQ						
ARQ/VRQ						
VRQ/VRQ						
Unknown	3	3.8%	1	8.3%	4	4.4%
Total	79	100%	12	100%	91	100%

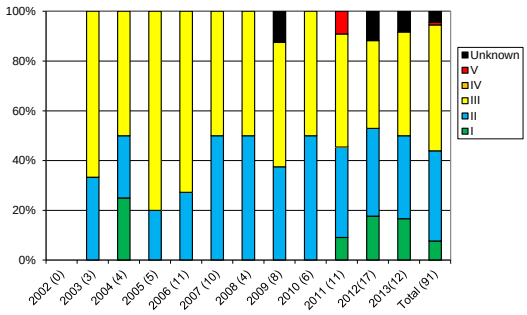


Figure 6 Percentage genotype distribution of NSP types of atypical scrapie confirmed in sheep by the FS survey from 2002 to 2013

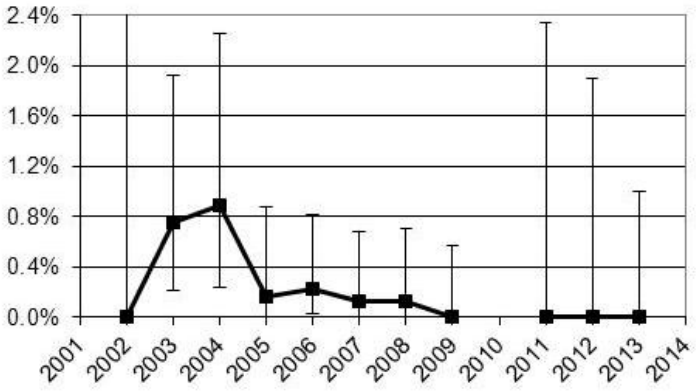
2.4 Dead in transit (DIT) survey

Sampling for the Dead in Transit (DIT) survey was discontinued in 2010 but put back in place in 2011. There were 340 DITs submitted and tested in 2013, 339 of them suitable for testing. The DIT survey confirmed one case of atypical scrapie of the ARH/AFRQ genotype. Numbers of samples tested, unsuitability rates, and the numbers classical and atypical scrapie cases for the period 2002-2013 are shown in Table 11.

Table 11 Summary of the DIT data in sheep for the period 2002-2013 (no sampling in DIT in 2010)

Year	Samples	Tested	Unsuitable	Clas.	% Clas. (95% CI)	Atyp.	% Atyp. (95%CI)
2002	10	10	0 (0.0%)	0	0 (0 – 32.6)	0	0 (0.00 – 32.6)
2003	550	530	20 (3.6%)	4	0.75 (0.22 – 2.01)	4	0.75 (0.22 – 2.01)
2004	480	452	28 (5.8%)	4	0.88 (0.26 – 2.36)	0	0 (0.00 – 1.04)
2005	676	634	42 (6.2%)	1	0.15 (0 – 1)	1	0.15 (0.00 – 1.00)
2006	941	892	49 (5.2%)	2	0.22 (0.01 – 0.88)	1	0.11 (0.00 – 0.71)
2007	884	822	62 (7.0%)	1	0.12 (0 – 0.68)	4	0.48 (0.9 – 1.30)
2008	876	792	84 (9.6%)	1	0.13 (0 – 0.7)	1	0.13 (0.00 – 0.7)
2009	663	650	13 (2%)	0	0 (0- 0.57)	1	0.15 (0.00 – 0.85)
2010	-	-	-	-	-	-	-
2011	156	156	0 (0%)	0	0 (0 – 2.34)	1	0.64 (0.02 – 3.52)
2012	193	193	0 (0%)	0	0 (0 – 1.9)	0	0 (0.00 – 1.9)
2013	340	339	1 (0.3%)	0	(0 – 1)	1	0.29 (0.1 – 1.6)

No significant changes in the prevalence of classical or atypical scrapie were detected in the DIT survey. Prevalence estimates of scrapie in GB from the DIT survey with 95% confidence intervals for the period 2002-2013 are shown in Figure 7.



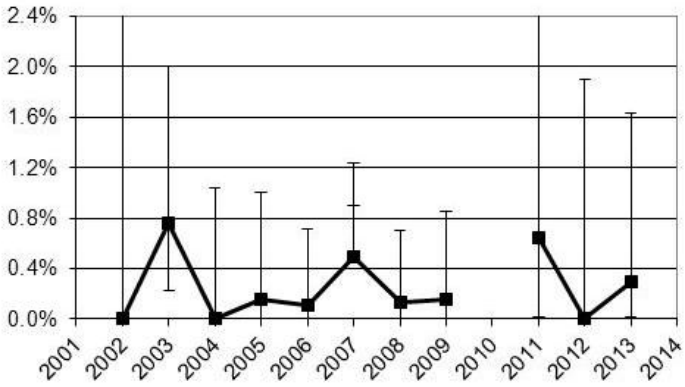


Figure 7 Prevalence estimates of scrapie in GB from the DIT survey with 95% confidence intervals. Period 2002-2013. Above: classical. Below: atypical

Table 12 and Figure 8 display the genotype distributions of the classical scrapie cases disclosed in the DIT for the period 2002-2013. Table 13 and Figure 9 show the corresponding genotype distributions for atypical scrapie cases.

Table 12 Genotype distributions of the classical scrapie confirmed cases in sheep by the DIT survey from 2002 to 2013

Genotype	2002-2012		2013	Total	
ARR/ARR					
ARR/AHQ					
ARR/ARH					
ARR/AFRQ					
ARR/ARQ					
AHQ/AHQ					
AHQ/ARH					
AHQ/AFRQ					
AHQ/ARQ					
ARH/ARH					
ARH/AFRQ					
ARH/ARQ					
AFRQ/AFRQ					
AFRQ/ARQ					
ARQ/ARQ					
ARR/VRQ	2	15.4%		2	15.4%
AHQ/VRQ					
ARH/VRQ	3	23.1%		3	23.1%
AFRQ/VRQ					
ARQ/VRQ	8	61.5%		8	61.5%
VRQ/VRQ					
Unknown					
Total	13	100%	0	13	100%

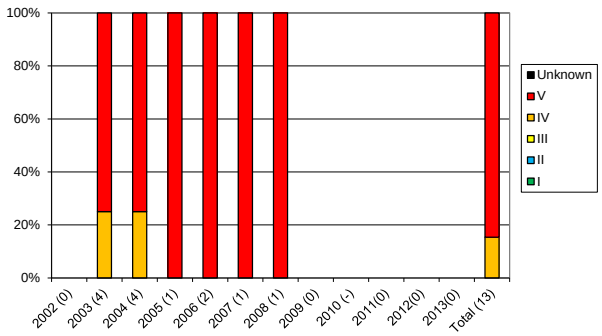


Figure 8 Percentage genotype distribution of NSP types of classical scrapie confirmed in sheep by the DIT survey from 2002 to 2013

Table 13 Genotype distributions of the atypical scrapie cases confirmed by the sheep DIT survey from 2002 to 2013

Genotype	2002-2012		2013		Total	
ARR/ARR	1	7.7%			1	7.1%
ARR/AHQ	7	53.8%			7	50.0%
ARR/ARH						
ARR/AFRQ						
ARR/ARQ						
AHQ/AHQ						
AHQ/ARH	1	7.7%			1	7.1%
AHQ/AFRQ						
AHQ/ARQ	2	15.4%			2	14.3%
ARH/ARH						
ARH/AFRQ			1	100%	1	7.1%
ARH/ARQ						
AFRQ/AFRQ						
AFRQ/ARQ						
ARQ/ARQ	2	15.4%			2	14.3%
ARR/VRQ						
AHQ/VRQ						
ARH/VRQ						
AFRQ/VRQ						
ARQ/VRQ						
VRQ/VRQ						
Unknown						
Total	13	100%	1	100%	14	100.0%

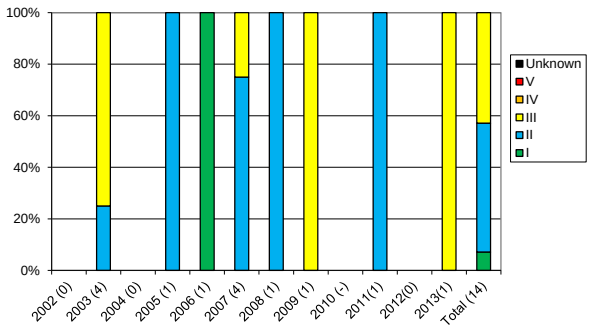


Figure 9 Percentage genotype distribution of NSP types of atypical scrapie confirmed in sheep by the DIT survey from 2002 to 2013

2.5 Abattoir survey (AS)

In 2013 the EU requirement for testing abattoir sheep over 18 months of age was set at 10,000 for the UK (with internal targets of 9,400 from GB and 600 from Northern Ireland). To achieve a seasonally adjusted sample, participating abattoirs were required to randomly select and sample a percentage of their total weekly throughput. The throughput was evenly distributed during the year with a sampling rate of 0.5% despite the fact that the quota was never intended to be met. Thirteen abattoirs were involved in the survey and one new plant was trained and became operational in July 2013 in England. Representation of abattoirs from Wales and Scotland was also maintained with one abattoir in each.

By the end of 2013, 6,896 samples had been submitted, of which 6,894 were suitable for testing, 73.4% of the allocated quota for GB. While short of the target for this survey, the reduced numbers were compensated for in part by the collection of more samples within the FS survey. The diminishing contribution of the Abattoir Survey to the active surveillance system continues compromising the only unbiased data source for the estimation of the prevalence of scrapie in GB (see section 2.5.1).

This was the third year in a row in which the AS survey did not confirm any case of classical scrapie. However there were three atypical cases confirmed (0.03% of suitable samples). the lowest since the establishment of this survey (Table 14).

Table 14 Summary of the AS data in sheep for the period 2002-2013

Year	Samples	Tested	Unsuitable	Clas.	% Clas. (95% CI)	Atyp.	% Atyp. (95%CI)	Ratio clas./aty.
2002	31847	30115	1732 (5.4%)	34	0.11 (0.08 – 0.15)	18	0.06 (0.03 – 0.09)	1.8:1
2003	76852	71250	5602 (7.2%)	46	0.06 (0.04 – 0.08)	45	0.06 (0.04 – 0.08)	1:1
2004	11481	10588	893 (7.7%)	9	0.08 (0.04 – 0.16)	12	0.11 (0.06 – 0.20)	1:1.3
2005	11629	11106	523 (4.5%)	12	0.1 (0.06 – 0.19)	16	0.14 (0.08 –0.23)	1:1.3
2006	48645	46259	2386 (4.9%)	8	0.017 (0.01 – 0.03)	35	0.075 (0.05 – 0.10)	1:4.3
2007	26059	24908	1151 (4.4%)	5	0.02 (0.01 – 0.04)	17	0.068 (0.04 – 0.11)	1:3.4
2008	10927	10158	769 (7%)	2	0.02 (0.00 – 0.07)	5	0.05 (0.02 – 0.12)	1:2.5
2009	10966	10652	314 (2.9%)	3	0.028 (0.01 – 0.08)	16	0.15 (0.09 – 0.24)	1:5.3
2010	7960	7823	137 (1.7%)	1	0.013 (0.00 – 0.07)	13	0.17 (0.09 – 0.28)	1:13
2011	6911	6813	98 (1.4%)	2	0.03 (0.00 – 0.11)	10	0.15 (0. 07 – 0.27)	1:5
2012	6429	6402	27 (0.4%)	0	0 (0.00 – 0.06)	11	0.17 (0.09 – 0.31)	
2013	6896	6894	2 (0.03%)	0	0 (0 - 0.05)	3	0.04 (0.01 – 0.13)	

There was no significant change in the prevalence of either classical or atypical scrapie in 2013 compared to the previous year. However the estimate of prevalence of atypical cases in 2013 was the lowest since this type of scrapie started to be discriminated from classical scrapie. At the current scenario of low prevalence, the level of testing precludes the detection

of significant changes in the trend year by year. It should be noted that the confidence intervals around annual estimates have overlapped in all consecutive years of the survey (Figure 10).

The number of animals tested in 2013 remained low when compared to 2010 and before, increasing the uncertainty of the estimated prevalence values for both classical and atypical scrapie.

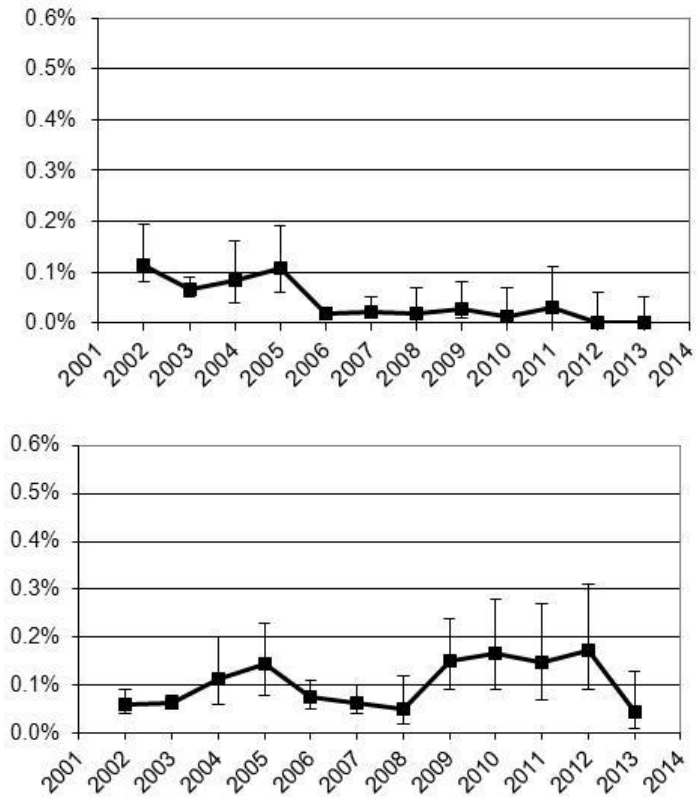


Figure 10 Prevalence estimates of scrapie in GB from the sheep AS with 95% confidence intervals. Period: 2002-2013. Above: classical. Below: atypical

Most of the AS cases cannot be traced back to the holdings of origin. They remain linked to the abattoir where they were sampled or the market where they were sourced, but not to the holding of origin, unless until further epidemiological investigations conclude with the confirmation of the cases in a particular holding/s.

The genotype distributions of classical scrapie cases in the AS, by individual genotype and NSP type, for the period 2002-2013 are displayed in Table 15 and Figure 11.

All three of the atypical scrapie cases confirmed in 2013 were of the following genotypes: ARR/AHQ, ARR/AFRQ and AHQ/AFRQ. The genotype and NSP type distributions of atypical scrapie cases detected by the AS between 2002 and 2013 are shown in Table 16 and Figure 12.

Table 15 Genotype distribution of classical scrapie cases confirmed in sheep by the AS survey from 2002 to 2013

Genotype	2002-2012		2013		Total	
ARR/ARR						
ARR/AHQ						
ARR/ARH						
ARR/AFRQ						
ARR/ARQ	1	0.8%			1	0.8%
AHQ/AHQ	1	0.8%			1	0.8%
AHQ/ARH						
AHQ/AFRQ						
AHQ/ARQ	2	1.6%			2	1.6%
ARH/ARH						
ARH/AFRQ						
ARH/ARQ						
AFRQ/AFRQ						
AFRQ/ARQ						
ARQ/ARQ	12	9.8%			12	9.8%
ARR/VRQ	39	32%			39	32%
AHQ/VRQ						
ARH/VRQ	8	6.6%			8	6.6%
AFRQ/VRQ						
ARQ/VRQ	50	41%			50	41%
VRQ/VRQ	8	6.6%			8	6.6%
Unknown	1	0.8%			1	0.8%
Total	122	100%	0		122	100%

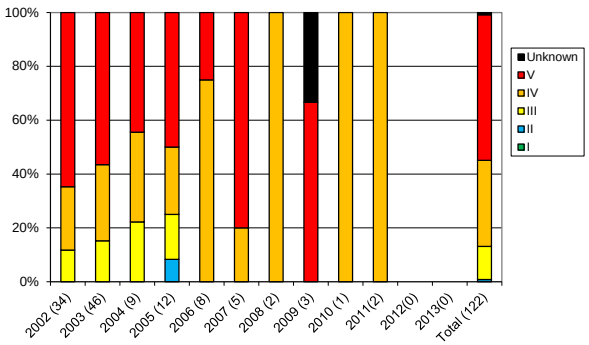


Figure 11 Percentage genotype distributions of NSP types of classical scrapie cases confirmed in sheep by the AS survey from 2002 to 2013

Table 16 Genotype distribution of the atypical scrapie cases confirmed in sheep by the AS survey from 2002 to 2013

Genotype	2002-2011		2012		Total	
ARR/ARR	18	9.6%			18	9.0%
ARR/AHQ	62	31.3%	1	33.3%	63	31.3%
ARR/ARH						
ARR/AFRQ	7	3.5%	1	33.3%	8	4.0%
ARR/ARQ	25	12.6%			25	12.4%
AHQ/AHQ	16	8.1%			16	8.0%
AHQ/ARH	4	2%			4	2.0%
AHQ/AFRQ	7	3.5%	1	33.3%	8	4.0%
AHQ/ARQ	35	17.7%			35	17.4%
ARH/ARH						
ARH/AFRQ						
ARH/ARQ	1	0.5%			1	0.5%
AFRQ/AFRQ	4	2%			4	2.0%
AFRQ/ARQ	3	1.5%			3	1.5%
ARQ/ARQ	15	7.6%			15	7.5%
ARR/VRQ						
AHQ/VRQ						
ARH/VRQ						
AFRQ/VRQ						
ARQ/VRQ	1	0.5%			1	0.5%
VRQ/VRQ						
Unknown						
Total	198	100.0%	3	100%	201	100.0%

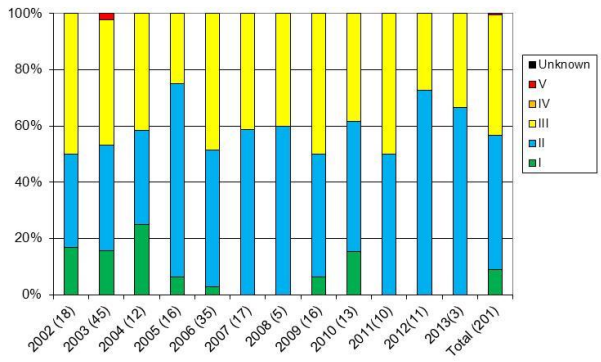


Figure 12 Percentage genotype distributions of NSP types of atypical scrapie cases confirmed in sheep by the AS survey from 2002 to 2013

2.5.1 Estimation of the prevalence of infection of classical scrapie in the GB national flock

The abattoir survey data provide the least biased estimation of the prevalence of scrapie since it is the closest to a random sampling of healthy animals over 18 months of age. The prevalence of classical scrapie was estimated applying the back-calculation model as described by Gubbins et al. (2003)[5] and using data from the 2013 AS data (total number of classical cases and total number of tested samples). Input parameters of the model include estimates of the incubation period distribution, the survivorship of sheep, and the sensitivity of the diagnostic tests. The output of the model is the number of infected sheep in the population required to produce the observed number of positives in the abattoir survey. According to the results of the model, the prevalence of infection in 2013 was 0% (95% CI: 0% – 0.13%). The estimate of no cases in the general population is due to the fact that the reporting year was the first in which the AS did not confirm any cases of classical scrapie. However the uncertainty of the parameter estimated up to 0.13% infection in the sheep population (Figure 13), which was not significantly different than that of previous years. In this situation of very low prevalence of classical scrapie it is advisable to integrate multiple data sources to estimate prevalence in a more robust way.

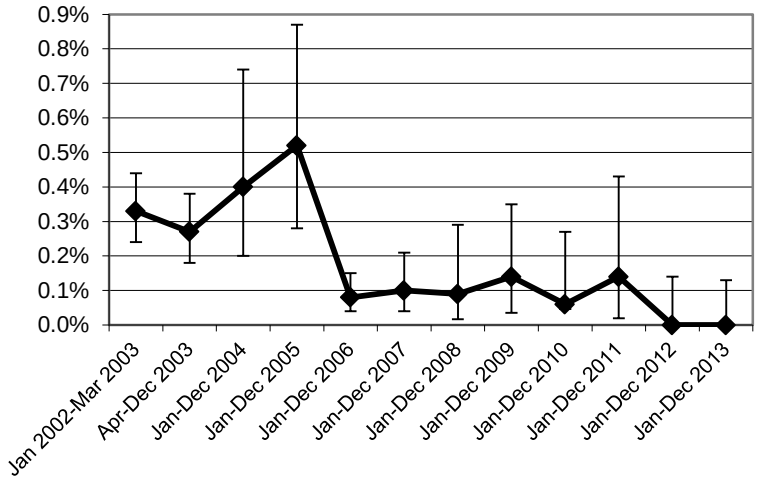


Figure 13 Estimates of prevalence of infection of classical scrapie in the sheep population for the period 2002-2013 with 95% confidence intervals

[5] Gubbins S, Simmons MM, Sivam K, Webb CR, Hoinville LJ. 2003. Prevalence of scrapie infection in Great Britain: interpreting the results of the 1997-1998-abattoir survey. Proc Biol Sci 270, 1919-1924.

2.6 Eradication measures: Classical scrapie. Compulsory Scrapie Flocks Scheme (CSFS)

There were four sheep holdings recruited to the scheme in 2013 (Figure 14), two of them located in Wales and two in England. Three of them were identified in the FS survey during the reporting year. The fourth case, confirmed in November 2012, joined the CSFS early in 2013. As part of the implementation of the new control measures, there is no more Initial Cull (IC) testing in CSFS holdings.

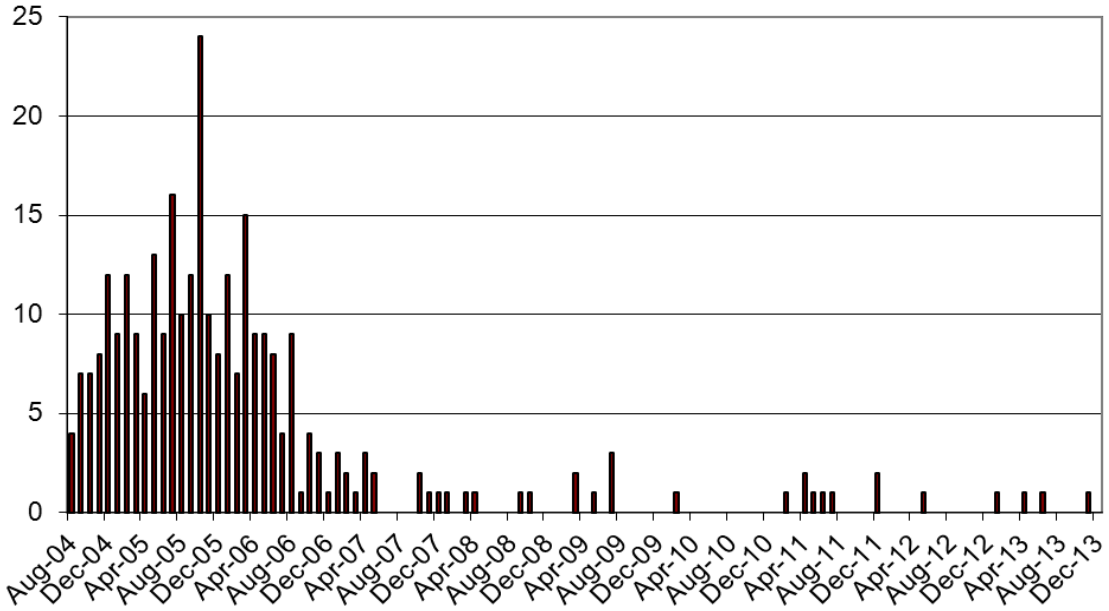


Figure 14 Number of declared sheep CSFS holdings per month for the period August 2004 to December 2013 [from 2011 figures based on date of service of NSP47].

A total of 10 holdings were under restrictions sometime during 2013: six still from previous years plus the new four CSFS holdings. Six of them (4 old and 2 new) submitted 244 samples for testing to the Fallen Stock (CSFS-FS) route, 231 of them suitable for testing. All the 13 unsuitable samples came from the same holding, located in Wales. The average number of submitted samples per holding was 40.6 (median: 25, range: 1-102). Two positive cases of classical scrapie were confirmed in the same CSFS holding in England under restrictions since April 2012.

Two holdings still under restrictions from previous years plus three of the new CSFS holdings submitted a total of 891 samples for testing to the Annual Cull (CSFS-AC) route, 888 of them suitable for testing. No positive cases of classical or atypical scrapie were confirmed by this testing route.

The total number of samples collected in 2013 under the various testing streams of the CSFS was 1135 from seven different holdings: 21.5% (244) were fallen sheep and 78.5%% (891) were healthy sheep slaughtered for human consumption (CSFS-AC). There were no clinical suspects submitted by CSFS holdings in 2013 (Table 17).

Table 17 Numbers of samples submitted, tested and confirmed scrapie positive under each of the CSFS testing routes in sheep for the period 2005 – 2013

Clinical suspects

Year	Sampled	Holdings	Tested (% unsuitable)	Classical	% Classical (95% CI)	Atypical	% Atypical (95% CI)
2005							
2006							
2007							
2008							
2009							
2010							
2011*	42	1	42 (0%)	41	97.6 (87.4-99.9%)	0	0(0.00-8.4%)
2012	-	-		-		-	
2013	-	-		-		-	

* All 41 scrapie cases identified in this route in 2011 originated from a multi-case holding

Initial Cull (CSFS-IC)

Year	Sampled	Holdings	Tested (% unsuitable)	Classical	% Classical (95% CI)	Atypical	% Atypical (95% CI)
2005*	15158		14963 (1%)	95[1]	0.63 (0.51-0.78%)	4	0.02 (0.01-0.07%)
2006	9364		9364 (0%)	76	0.81 (0.064-1.01%)	8	0.08 (0.04-0.17%)
2007	1353		1353 (0%)	3	0.22 (0.04-0.69%)	0	0 (0.00-0.35%)
2008	471		468 (0.34%)	0	0 (0.00-0.79%)	0	0 (0.00-0.79%)
2009	570		569 (0.2%)	1	0.18 (0.00-0.98%)	0	0 (0.00-0.65%)
2010	26	1	26 (0%)	0	0 (0.00-3.23%)	0	0 (0.00-3.23%)
2011	183	3	183 (0%)	0	0 (0.00-2%)	0	0 (0.00-2%)
2012	-	-	-	-		-	
2013	-	-	-	-		-	

*Three inconclusive results were not included in the 2005.

Fallen Stock (CSFS-FS)

Year	Sampled	Holdings	Tested (% unsuitable)	Classical	% Classical (95% CI)	Atypical	% Atypical (95% CI)
2005	383		288 (24.8%)	7	2.43 (0.98 – 4.94%)	1	0.35 (0.01-1.92%)
2006	2297		1611 (29.9%)	9	0.55 (0.28 -1.01%)	1	0.06 (0.0-0.39%)
2007	2428		1738 (28.4%)	2	0.11(0.05 - 0.45%)	3	0.17 (0.03-0.53%)
2008	1350		1085 (19.6%)	0	0 (0.00 - 0.34%)	2	0.18 (0.02-0.66%)
2009	293		270 (7.9%)	1	0.37(0.01 - 2.05%)	0	0 (0.00-1.36%)
2010	126	12	123 (2.4%)	0	0 (0.00 - 2.95%)	1	0.81 (0.02-4.45%)
2011*	291	8	286 (1.7%)	81	28.3 (23.2 - 33.9%)	0	0 (0.00– 1.28%)
2012	177	6	171 (3.4%)	2	1.7 (0.14 – 4.2%)	0	0 (0.00 – 2.1%)
2013	244	6	231 (4.5%)	2	0.87 (0.11 – 3.1%)	0	0 (0-1.6%)

* All 81 scrapie cases identified in this route in 2011 originate from a single holding.

Annual Cull (CSFS-AC)

Year	Sampled	Holdings	Tested (% unsuitable)	Classical	% Classical (95% CI)	Atypical	% Atypical (95% CI)
2005	-		-	-		-	
2006	351		350 (0.3%)	0	0 (0.0 –0.13%)	1	0.28 (0.0-0.18%)
2007	510		510 (0%)	0	0 (0.0 –0.92%)	0	0 (0.0 – 0.92%)
2008	390		390 (0%)	0	0 (0.00-0.94%)	0	0 (0.00-0.94%)
2009	73		73 (0%)	0	0 (0.00-4.93%)	0	0 (0.00-4.93%)
2010	12	2	12 (0%)	0	0 (0.00 26.47%)	0	0 (0.00-26.47%)
2011	17	1	17 (0%)	0	0 (0.00-19.51%)	0	0 (0.00-19.51%)
2012	444	5	440 (0.9%)	2	0.45 (0.06 – 1.6%)	0	0 (0.00 – 0.84%)
2013	891	3	888 (0.33%)	0	0 (0 – 0.41%)	0	0 (0 – 0.41%)

All active CSFS testing routes (IC, FS and AC)

Year	Sampled	Holdings	Tested (% unsuitable)	Classical	% Classical (95% CI)	Atypical	% Atypical (95% CI)
2005	15541		15251 (1.9%)	102	0.67 (0.55-0.81%)	5	0.03 (0.01-0.08%)
2006	12012		11325 (5.7%)	85	0.75 (0.6-0.93%)	10	0.09 (0.04-0.16%)
2007	4291		3601 (16.1%)	5	0.9 (0.05-0.32%)	3	0.08 (0.02-0.24%)
2008	2211		1943 (12.1%)	0	0 (0.00-0.19%)	2	0.1 (0.01-0.37%)
2009	936		912 (2.6%)	2	0.22 (0.03-0.79%)	0	0 (0-0.40%)
2010	164	13	161 (1.8%)	0	0 (0.00-2.27%)	1	0.6 (0.02-3.41%)
2011	491	12	486 (1.0%)	81	16.7 (13.5-30.3%)	0	0 (0-0.76%)
2012	621	7	611 (1.6%)	4	0.65 (0.18 - 1.6%)	0	0 (0 - 0.6%)
2013	1135	7	1122 (1.4%)	2	0.18 (0.02 – 0.64)	0	0 (0 – 0.33%)

Two classical scrapie cases were confirmed by the CSFS in 2013, both in a Welsh holding and by the CSFS-FS. The multi-case holding reported in the 2011 submitted 102 and 130 sheep to the FS and AC for TSE testing in 2013, respectively, with no more cases of classical scrapie confirmed. One of two classical scrapie cases was of the ARH/ARQ genotype and the other of unknown genotype. No cases of atypical scrapie were detected in any of the CSFS testing routes. The genotype distributions of classical and atypical scrapie cases confirmed in the CSFS in the period 2005-2013 are displayed in Tables 18 and Figure 15 (classical scrapie) and Table 19 and Figure 18 (atypical scrapie), respectively.

Table 18 Genotype distribution of classical scrapie cases confirmed in sheep by the CSFS from 2005 to 2013

Genotype	2005-2012		2013		Total	
ARR/ARR						
ARR/AHQ						
ARR/ARH						
ARR/AFRQ						
ARR/ARQ	1	0.3%			1	0.3%
AHQ/AHQ	2	0.6%			2	0.6%
AHQ/ARH					0	0.0%
AHQ/AFRQ					0	0.0%
AHQ/ARQ	6	1.9%			6	1.9%
ARH/ARH	3	0.9%			3	0.9%
ARH/AFRQ					0	0.0%
ARH/ARQ	10	3.2%	1	50%	11	3.4%
AFRQ/AFRQ					0	0.0%
AFRQ/ARQ					0	0.0%
ARQ/ARQ	74	23.1%			74	23.0%
ARR/VRQ	4	1.2%			4	1.2%
AHQ/VRQ					0	0.0%
ARH/VRQ	27	8.4%			27	8.4%
AFRQ/VRQ					0	0.0%
ARQ/VRQ	165	51.5%			165	51.2%
VRQ/VRQ	20	6.2%			20	6.2%
Unknown	8	2.5%	1	50%	9	2.8%
Total	320	100%	2	100%	322	100%

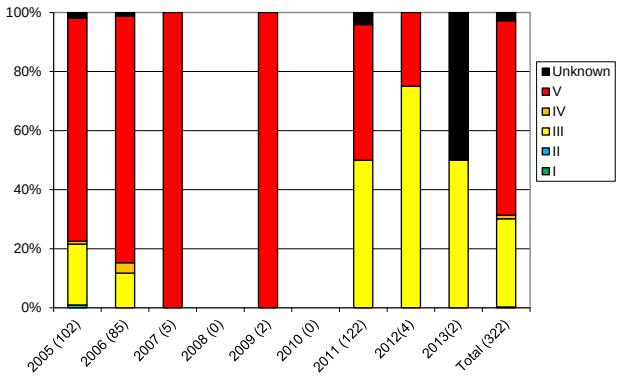


Figure 15 Percentage genotype distributions of NSP types of classical scrapie cases confirmed in sheep by the CSFS from 2005 to 2013 (totals include 3 originally inconclusive results in 2005).

Table 19 Genotype distribution of atypical scrapie cases confirmed in sheep by the CSFS from 2005 to 2013

Genotype	2005-2012		2013	Total	
ARR/ARR					
ARR/AHQ	8	38.1%		8	38.1%
ARR/ARH					
ARR/AFRQ	2	9.5%		2	9.5%
ARR/ARQ					
AHQ/AHQ	2	9.5%		2	9.5%
AHQ/ARH					
AHQ/AFRQ					
AHQ/ARQ	6	28.5%		6	28.5%
ARH/ARH					
ARH/AFRQ					
ARH/ARQ					
AFRQ/AFRQ					
AFRQ/ARQ					
ARQ/ARQ	3	14.3%		3	14.3%
ARR/VRQ					
AHQ/VRQ					
ARH/VRQ					
AFRQ/VRQ					
ARQ/VRQ					
VRQ/VRQ					
Unknown					
Total	21	100%	0	21	100%

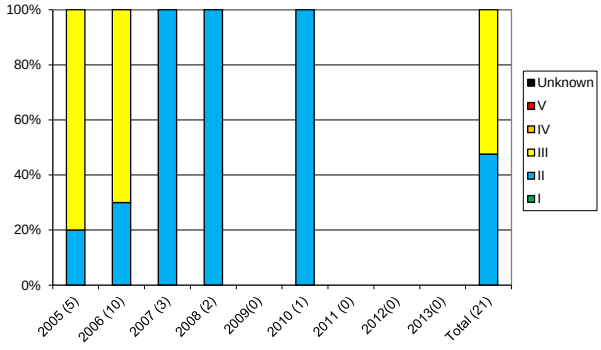


Figure 16 Percentage genotype distributions of NSP types of atypical scrapie cases confirmed in sheep by the CSFS from 2005 to 2013.

2.7 Eradication measures: Atypical Scrapie Monitoring (ASM)

Twenty-three holdings, thirteen in England, one in Scotland and nine in Wales joined the ASM in 2013. Some of them had the case confirmed in 2012 and not all the holdings with cases of atypical scrapie confirmed in 2013 joined the scheme. This is due to the time gap in the sequence of events between date of confirmation of the case and date of commencement of restrictions. The latter is the date used to include the holding in the scheme. Some if not all the holdings that reminded pending at the end of the 2013 will join the scheme during 2014 (Figure 17). These 23 holdings joined the 18 already in the scheme to a total of 41 holdings that were under the restrictions of the scheme sometime during 2013.

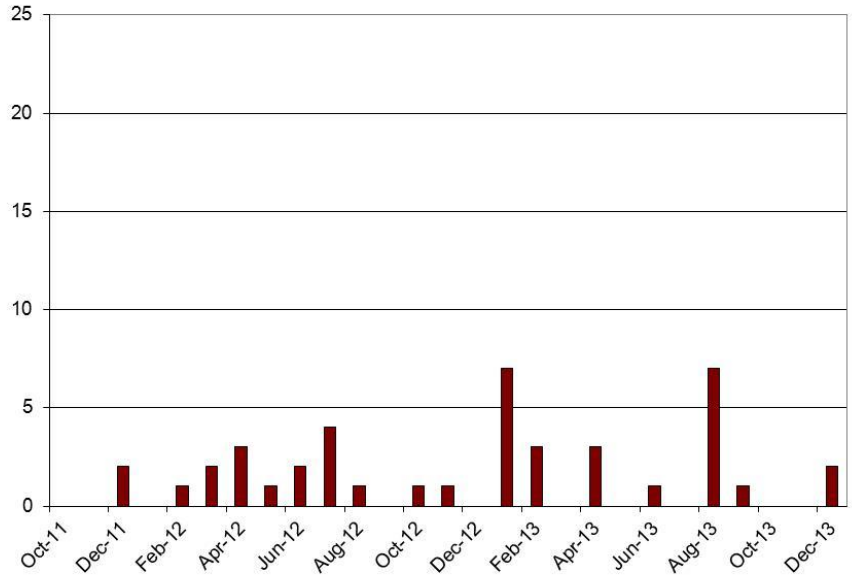


Figure 17 Number of declared sheep ASM holdings per month for the period October 2011 to December 2013 [based on date of start of restrictions].

Thirty-two of the 41 holdings under restrictions submitted 759 samples to the ASM-FS testing route during the reporting year, 743 of them suitable for testing. The average number of submitted samples per holding was 23.7 (median: 12, range: 1-95). One case of classical scrapie of the genotype ARH/VRQ was confirmed in a holding in Wales, in another example of a holding in which a case of classical and atypical scrapie each have been confirmed almost simultaneously (Table 20).

Six of the forty-one holdings under restrictions submitted 254 samples to the AC survey during the reporting year, 251 of them suitable for testing. The average number of submitted samples per holding was 42.3 (median: 26.5, range: 6-140). No cases were confirmed by this testing route of the ASM (Table 20).

Table 20 Numbers of samples submitted, tested and confirmed scrapie positive under each of the ASM testing routes in sheep for the period 2011 – 2013

Fallen Stock (ASM-FS)

Year	Sampled	Holdings	Tested (% unsuitable)	Classical	% Classical (95% CI)	Atypicals	% Atypical (95% CI)
2011	2	1	2 (0%)	0		0	
2012	177	10	174 (1.7%)	0	0 (0 - 2.1%)	0	0 (0 - 2.1%)
2013	759	32	743 (2.1%)	1	0.13 (0 – 0.75%)	0	0 (0– 0.5%)

Annual Cull (ASM-AC)

Year	Sampled	Holdings	Tested (% unsuitable)	Classical	% Classical (95% CI)	Atypicals	% Atypical (95% CI)
2011	-	-	-	-		-	
2012	-	-	-	-		-	
2013	254	6	253 (0.04%)	0	0 (0 – 1.4%)	0	0 (0 – 1.4%)

All active ASM testing routes (FS and AC)

Year	Sampled	Holdings	Tested (% unsuitable)	Classical	% Classical (95% CI)	Atypicals	% Atypical (95% CI)
2011	2	1	2 (0%)	0		0	
2012	177	10	174 (1.7%)	0	0 (0 - 2.1%)	0	0 (0 - 2.1%)
2013	1013	35	996 (1.7%)	1	0.1 (0 – 0.56%)	0	0 (0 – 0.37%)

The genotype distributions of classical and atypical scrapie cases confirmed in the ASM in the period 2011-2013 are displayed in Tables 21 and Figure 18 (classical scrapie) and Table 22 and Figure 19 (atypical scrapie), respectively.

Table 21 Genotype distribution of classical scrapie cases confirmed in sheep by the ASM from 2011 to 2013

Genotype	2011-2012	2013		Total	
ARR/ARR					
ARR/AHQ					
ARR/ARH					
ARR/AFRQ					
ARR/ARQ					
AHQ/AHQ					
AHQ/ARH					
AHQ/AFRQ					
AHQ/ARQ					
ARH/ARH					
ARH/AFRQ					
ARH/ARQ					
AFRQ/AFRQ					
AFRQ/ARQ					
ARQ/ARQ					
ARR/VRQ					
AHQ/VRQ					
ARH/VRQ		1	100%	1	100%
AFRQ/VRQ					
ARQ/VRQ					
VRQ/VRQ					
Unknown		1	100%	1	100%
Total00	0	1	100%	1	100%

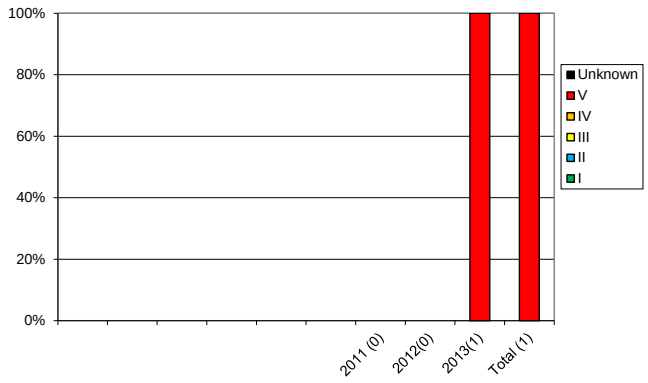


Figure 18 Percentage genotype distributions of NSP types of classical scrapie cases confirmed in sheep by the ASM from 2011 to 2013

Table 22 Genotype distribution of atypical scrapie cases confirmed in sheep by the ASM from 2011 to 2013

Genotype			
ARR/ARR			
ARR/AHQ			
ARR/ARH			
ARR/AFRQ			
ARR/ARQ			
AHQ/AHQ			
AHQ/ARH			
AHQ/AFRQ			
AHQ/ARQ			
ARH/ARH			
ARH/AFRQ			
ARH/ARQ			
AFRQ/AFRQ			
AFRQ/ARQ			
ARQ/ARQ			
ARR/VRQ			
AHQ/VRQ			
ARH/VRQ			
AFRQ/VRQ			
ARQ/VRQ			
VRQ/VRQ			
Unknown			
Total	0	0	0

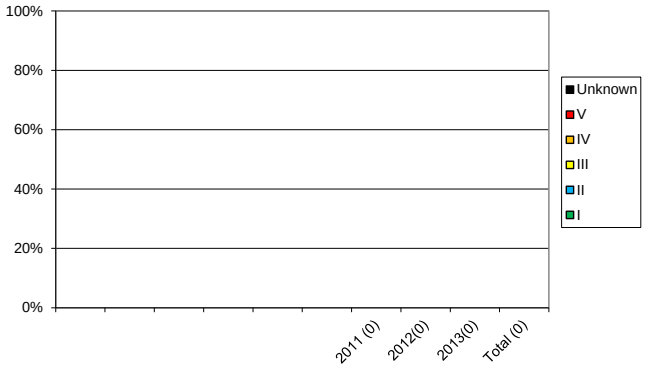


Figure 19 Percentage genotype distributions of NSP types of atypical scrapie cases confirmed in sheep by the ASM from 2011 to 2013.

2.8 All sources

2.8.1 Cases and genotypes

There was one holding that yielded cases from multiple active surveillance routes in 2013. A holding in Wales that joined the ASM scheme on the 24 July 2012 had a case of classical scrapie confirmed by the FS in 2013. Nineteen other holdings have had cases of both classical and atypical scrapie in animals that were born or had been kept, as per results of the investigation conducted under research project SE0260, funded by Defra (final report due on 31 October 2014). The multi-case holding identified in 2011 did not have any case confirmed in 2013 and will be under restrictions until February 2014, should it not have any new cases confirmed.

For active surveillance testing routes, the proportion of samples confirmed positive provides an estimate of the apparent prevalence of scrapie in the sampled population of each source. For example, in the FS survey in 2013, 0.02% (95% CI: 0 – 0.07%) of suitable samples were confirmed with classical scrapie and 0.1% (95% CI: 0.05 – 0.17%) with atypical scrapie, with a total of three and twelve cases, respectively. In the AS, three cases of atypical scrapie were confirmed in the abattoir survey yielding prevalence estimates in the population of sheep covered by the AS of 0.04% (0.01 - 0.13%). The only case confirmed in the DIT survey (atypical) yields a prevalence estimate of 0.29% for this source, the confidence interval (0.01–1.63%) overlaps those of the FS survey and AS, and so is not significantly different. The prevalence estimates of classical (0.18% and 0.1%) and atypical (0% and 0%) scrapie in the CSFS and ASM, respectively, should not be considered representative of the general population and result of the aggregation of FS and AC figures. The contribution of the different sources to the overall number of samples submitted, samples tested and to the number of cases of both types of scrapie in 2013 is shown in Figure 20. For the first time the ASM scheme has been included in the calculations.

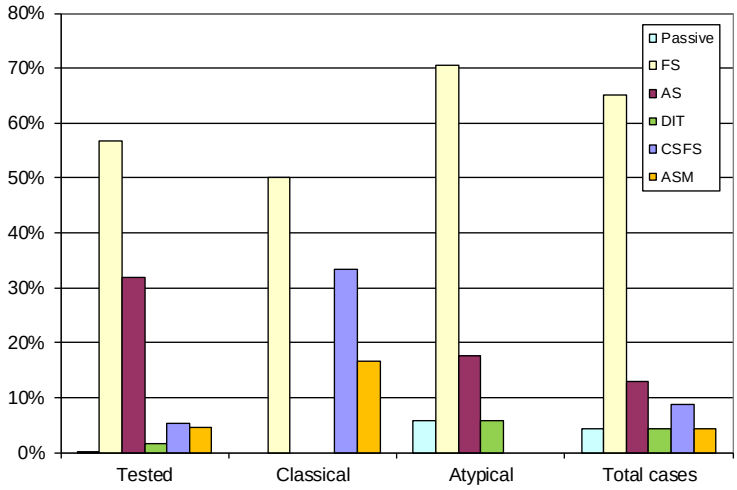


Figure 20 Percentage contribution of the six surveillance sources to the number of tested samples and cases of scrapie in sheep in 2013

The six cases of classical scrapie were detected by the FS (3), the CSFS (2) and the ASM (1). The contribution of the ASM to the total number of sheep testing throughput has increased substantially in 2013, with 996 tested samples (4.6%). The FS and the CSFS remain the main sources of classical scrapie cases with 50% and 33.3% of all cases, although the level of detection remained low. Figure 21 shows the changes in relative contribution to confirmed cases of classical scrapie provided by the different surveillance sources since 2002, including the ASM.

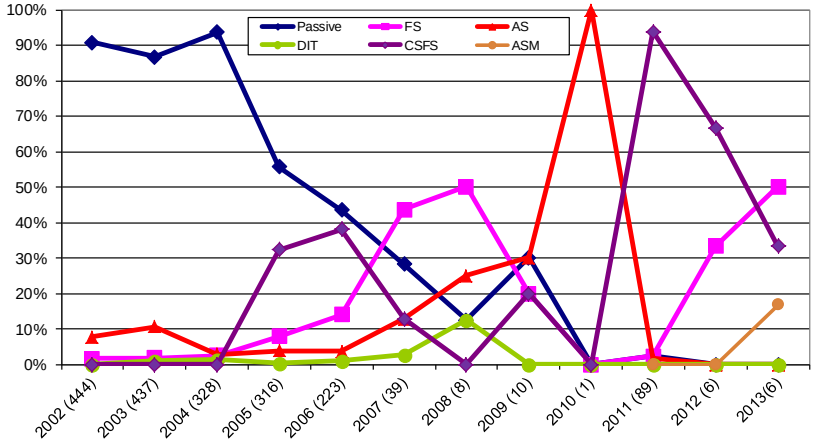


Figure 21 Percentage contribution of the six surveillance sources to the number of cases of classical scrapie in sheep for the period 2002-2013

Seventeen cases of atypical scrapie were confirmed in 2013 across all surveillance sources. Consolidating the feature of the 2011 distribution of cases, the FS remained the main contributor to the detection of atypical scrapie with 70.6% of the cases, followed by the AS survey with 17.6%. The AS continued decreasing its contribution to the detection of atypical scrapie to the lowest rate ever in 2013, in parallel with the diminishing throughput of this testing route. Passive surveillance and the DIT had one single case of atypical scrapie confirmed each. The prevalence estimates for atypical scrapie provided by the AS and FS: 0.04% and 0.1%, respectively, were not significantly different to values reported in 2012.

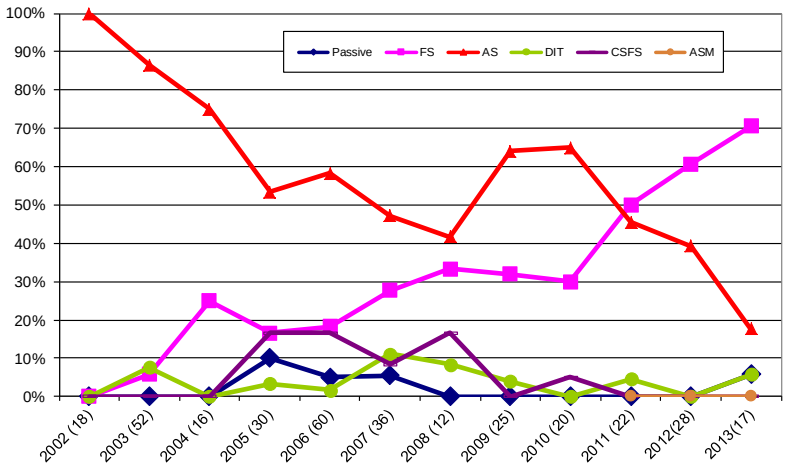


Figure 22 Percentage contribution of the six surveillance sources to the number of cases of atypical scrapie in sheep for the period 2002-2013

The prevalence estimates recorded by these sources cannot be used to make inferences on the true prevalence in the general population since the number of animals selected for sampling, and their farms of origin are not based on statistically sound probability sampling. Whereas the FS survey includes data from holdings of unknown scrapie status and depends on the willingness of farmers to report and be selected for testing (only a small proportion of all reported fallen sheep by farmers is selected for testing), the CSFS testing is driven by statutory requirements and only includes data from testing conducted at cull (IC) and post-cull (FS and AC) in scrapie affected holdings. The AS is supposed to represent a random and representative sample of the over 18 months healthy slaughter population. Passive surveillance is affected by the prevalence of the disease in the population and farmer's willingness to report disease whereas the FS survey and AS depends on quotas imposed by the EU. Despite these differences, the detection of variations and trends year by year within the same testing route can be informative, given the consistency of the selection and diagnostic methods applied in the last twelve years.

The most frequent genotypes in classical scrapie cases in GB since 2002 (excluding unknown/inconclusive results) have been ARQ/VRQ (47.5%), ARQ/ARQ (20%), VRQ/VRQ (13%), ARH/VRQ (6.4%) and ARR/VRQ (6.1%). Four of the six cases confirmed in 2013 had known genotypes: ARH/ARQ (2), ARQ/ARQ (1) and ARH/VRQ (1).

For atypical scrapie the most frequent genotypes in GB since 2002, again excluding any unknowns, were: ARR/AHQ (32.3%), AHQ/A(F/L)RQ (20.5%), A(F/L)RQ/A(F/L)RQ (12.1%), ARR/A(F/L)RQ (12.4%), and AHQ/AHQ (11.2%). The known genotypes (16) of the 17 atypical cases confirmed in 2013 were: ARR/AHQ (4), AHQ/AHQ (3), ARR/AFRQ (3), ARR/ARR (2), and ARQ/AHQ, AHQ/AFRQ, ARH/AFRQ and AFRQ/AFRQ with one case each. The genotypes were not significantly different in 2013 compared to frequencies observed in the period 2002 – 2012.

2.8.2. Genotypes of selected negatives

Following Commission Regulation (EC) 1139/2003 amending Regulation (EC) 999/2001, in addition to each positive TSE case in sheep, the prion protein genotype shall be determined for a random subsample of the ovine animals tested under active surveillance. Further amendment as per Commission Regulation (EC) No 727/2007 requires that the prion protein genotype for the codons 136, 141, 154 and 171 of at least 600 animals shall be determined for a member state with an adult sheep population of more than 750,000 animals, as is the case in the UK. The samples, representative of the entire ovine population, may be chosen from animals slaughtered for human consumption, from animals found dead-on-farm or from live animals.

The genotyping of selected negatives has been undertaken since 2004 and the number of sheep sampled every year has remained stable except in 2005 where only 19 negative sheep were genotyped. In 2004, 532 samples were selected for genotyping, and 423, 625, 723 and 579, 569, 600 in the years 2006-2011 respectively, and 615 in 2013. The proportion of

selected negative sheep testing for each NSP type in each year is displayed in Figure 23. In 2012-3, a research project (SE0259[6]) was conducted aiming at genotyping up to 10,000 samples from the two active surveillance routes. This amount exceeds the requirement of the EC Regulation. Results of 9,600 genotyped sheep are reported for 2012 and included in Figure 23.

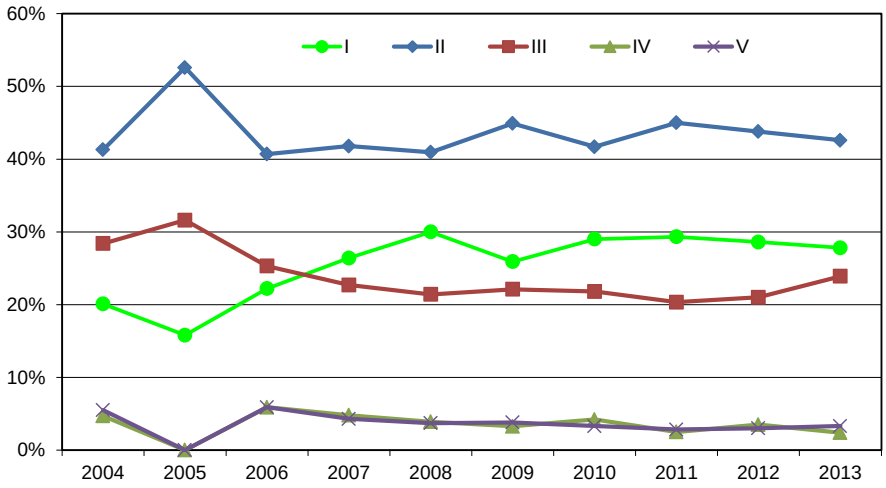


Figure 23 Percentage contribution of the five NSP types to the selected negatives for genotyping for the period 2004-2013

2.8.3 Unsuitable samples

The AS has shown low unsuitability rates historically, given the short time between death of the animal and sample collection, with 0.03% recorded in 2013. Equally the AC of the CSFS and more recently the ASM typically show low unsuitability rates (0.3% and 0.04%, respectively, in 2013) reflecting the controlled collection of samples and the short time between sampling and testing.

The FS survey reported the lowest unsuitability rate with 0.15% in 2013, a similar rate to that in the previous year and well below the 5% target set by Defra. The transfer of sample collection to the contractor's sites from the beginning of 2009 and the increase of sites in 2011 have proven to be a success in this respect. The unsuitability rate for individual months continued well below 2%, reaching the highest rate in August with 1.46%. As in the previous year, sample condition was the only cause of unsuitability.

The overall unsuitability rates in the CSFS and ASM, at 1.4% and 1.7% respectively, are higher than those of the FS and AS rates, but consolidating in all cases the trend observed in the last three years, with rates below the 5% target. The CSFS-FS was the exception with 4.5% of unsuitable samples due to the thirteen samples untestable out of a total of 76 submitted by a single holding in Wales. The ASM-FS had 2.1% of unsuitable samples. Finally

[6] Ortiz-Peláez A., Thompson C.E., Dawson M. The impact of the National Scrapie Plan on the PRNP genotype distribution of the British national flock, 2002-2012 (The Veterinary Record 24 May 2014, doi: 10.1136/vr.102255)

the AC of the CSFS-AC and the ASM-AC had both very low unsuitability rates: 0.33% and 0.04%, respectively.

3.Goat scrapie surveillance

3.1 Background

BSE has been confirmed in two goats in the EU. The first case was confirmed in January 2005 by the European Union Reference Laboratory (EURL) (formerly Community Reference Laboratory) for TSEs in a goat slaughtered in France in 2002. The goat was disposed of after slaughtering as well as its entire herd and did not enter the food chain. In addition, retrospective testing of a Scottish goat born in 1987 and culled in 1990 gave results indicative of BSE and in May 2009 the EURL concluded that the results were indistinguishable from BSE[7]. Samples from another goat slaughtered in 2008 in GB, in which initially BSE could not be excluded, underwent testing and bioassays. The results did not confirm suspicion of BSE.

In 2002 Defra and the Devolved Administrations (DAs) in Scotland and Wales began a programme of active surveillance for goat scrapie. Surveys of the slaughtered population, the abattoir survey (AS), and fallen stock on farm, the fallen stock survey (FS), have been conducted throughout each year as per EU quotas. In the AS, goats fit for human consumption and older than 18 months of age were sampled randomly at the abattoir. In the FS, dead-on-farm goats, older than 18 months and reported by the farmers, were sampled. The AS was discontinued after 2007.

Before the statutory CSFS came into force in 2004, there had been two confirmed cases of scrapie in goats: one clinical case in 2002 and one goat slaughtered for human consumption in 2003. Since then seven holdings are or have been under restrictions due to the confirmation of scrapie cases, all of them classical.

In March 2008, Defra killed two goat herds in England in which the annual incidences of cases of classical scrapie were increasing. The VLA collected a range of samples from different tissues from a number of animals from both herds for further investigation[8].

[7] http://vla.defra.gov.uk/science/docs/sci_tse_rl_steg0509.pdf

[8] For further information: González L., Martin S., Sisó S., Konold T., Ortiz-Peláez A., Phelan L., Goldmann W., Stuart P., Windl O., Jeffrey M., Hawkins S.A.C., Dawson M., Hope J. High prevalence of scrapie in a dairy goat herd. Veterinary Research 2009, 49:5; and

González L., Martin S., Hawkins S.A., Goldmann W., Jeffrey M., Sisó S. Pathogenesis of natural goat scrapie: modulation by host PRNP genotype and effect of co-existent conditions. Veterinary Research 2010 ;41:48

3.2 Passive surveillance

There were two reports of clinical suspects during 2013 from one unregistered holding in England. The two goats were not traced back yet hence no statutory action was applied on that holding or any other at the time of writing this report. Thus no goat holding was enrolled in the CSFS in 2013 due to passive surveillance.

The numbers of reported, tested and confirmed goat cases by SND from 2002 to 2013 are presented in Table 23. Please note that in the years 2006 – 2008, 49 positive cases were reported from two holdings previously confirmed as scrapie-affected. Subsequent cases from scrapie-affected herds were recorded and monitored by the Compulsory Scrapie Flocks Scheme (CSFS) (see section 2.5). These 49 cases are not included in Table 23.

Table 23 Number of cases reported, tested and confirmed by passive surveillance in goats from 2002 to 2013

Year	Reported	Tested	Confirmed	% Reported not confirmed	% Tested not confirmed	Classical	% Classical
2002	2	2	1	50%	50%	1	50%
2003	1	0	0	100%	0%	0	0%
2004	8	7	0	100%	100%	0	0%
2005	6	6	0	100%	100%	0	0%
2006	1	1	0	100%	100%	0	0%
2007	2	1	0	100%	100%	0	0%
2008	3	2	0	100%	100%	0	0%
2009	1	1	1	0%	100%	1	100%
2010	0	0	0	-	-	0	-
2011	0	0	0	-	-	0	-
2012	2	1	1	50%	0%	1	100%
2013	2	2	2	0%	0%	2	100%

3.3 Fallen stock

In 2013, the annual EU requirement for testing fallen goats over 18 months was set at 500 in the UK (with internal targets of 480 from Great Britain and 20 from Northern Ireland). A total of 506 samples from fallen goats were submitted during 2013, 5.4% more than required. All but one were suitable for testing, No submissions of goats Dead in Transit (DIT) were made in 2013.

No cases of classical scrapie were detected in the goat fallen stock survey in 2013. Summary data for the FS and DIT in goats in the period 2002-2013 are shown in Table 24.

Table 24 FS data summary in goats for the period 2002-2013. DIT figures are shown in brackets and not included in the FS figures

Year	Samples	Tested	Unsuitable	Classical	% (95% CI)	
2002	3 (0)	3 (0)	0 (0)	0	0	(0-70%)
2003	54 (2)	51 (2)	3 (0)	0 (0)	0	(0-7%)
2004	59 (0)	51 (0)	8 (0)	0	0	(0-7%)
2005	2001 (3)	1284 (2)	717 (1)	2 (0)	0.08	(0.02-0.56%)
2006	3893 (1)	2466 (1)	1427 (1)	1 (0)	0.04	(0-0.23%)
2007	1583 (1)	1194 (1)	389 (1)	1 (0)	0.08	(0-0.47%)
2008	870 (1)	715 (1)	155 (1)	1 (0)	0.9	(0.03-0.78%)
2009	710(0)	656 (0)	53 (0)	0	0	(0-0.56%)
2010	529 (0)	507 (0)	22 (0)	0	0	(0-0.73%)
2011	501 (0)	501 (0)	0 (0)	1	0.2	(0.01-1.11%)
2012	500	498	2 (0.4)	1	0.2	(0.01-1.11%)
2013	506	505	1 (0.2%)	0	0	(0 – 0.73%)

A total 497 of the 506 submissions (98.2%) were supplied with CPH details which permitted allocation by the country of the farm of origin. The contribution of submitted samples from holdings in England was 492 (99%), three from holdings in Scotland (0.6%) and two from holdings in Wales (0.4%) (Table 25). In terms of number of samples and using as reference the distribution of goat population by country as in the Sheep and Goat Inventory 2013 (89.2%, 3.7% and 7% in England, Scotland and Wales, respectively), England was significantly over-represented in the FS goat survey whereas Scotland and Wales were under-represented at the 0.05 level.

Table 25 Summary of the FS data in goats per country for the period 2002-2013

Year	Samples	England	%	Scotland	%	Wales	%
2002	3	3	100%	0	0.0%	0	0.0%
2003	54*	46	85.2%	3	5.6%	3	5.6%
2004	59	53	89.8%	1	1.7%	5	8.5%
2005	2001*	1847	92.3%	67	3.3%	63	3.1%
2006	3893*	3474	89.2%	171	4.4%	159	4.1%
2007	1583*	904	88.7%	92	5.8%	66	4.2%
2008	870*	792	91%	55	6.3%	22	2.5%
2009	710	652	91.8%	38	5.4%	20	2.8%
2010	529*	481	90.9%	31	5.9%	16	3.0%
2011	501*	440	87.8%	14	2.8%	12	2.4%
2012	500	483	99.1%	4	0.9%	0	0%
2013	497	492	99%	3	0.6%	2	0.4%

*CPHs from 2, 24, 89, 23, 1 , 1, 35, 13 and 1 samples submitted in 2003, 2005, 2006, 2007, 2008 , 2010, 2011, 2012 and 2103 respectively, could not be ascertained

The average number of submitted samples per goat holding in England in 2013 was 4.1 (median: 1 range: 1-104), higher than in Scotland and Wales (mean: 1, median: 1, range: 1-1). A total of 124 holdings submitted fallen goats in 2013 (Table 26), 119 from England (96%). three from Scotland (2.5%) and two from Wales (1.5%). In terms of number of holdings and using as reference the total number of goat holdings by country as in the Sheep and Goat Inventory 2013 (83.8%, 5.8% and 10.4%, respectively), England is significantly over-represented and Wales is under-represented while Scotland is fairly represented, all at the 5% significance level.

Table 26 Summary of the FS data on goat holdings per country for the period 2002-2013

Year	Holdings	England	%	Scotland	%	Wales	%
2002	1	1	100%	0	0.0%	0	0.0%
2003	40	35	87.5%	2	5.0%	3	7.5%
2004	41	37	90.2%	1	2.4%	3	7.3%
2005	628	566	90.1%	31	4.9%	31	4.9%
2006	834	726	87.1%	68	8.1%	40	4.8%
2007	406	338	83.3%	45	11%	23	5.7%
2008	278	237	85.3%	26	9.3%	15	5.4%
2009	189	166	87.8%	10	5.3%	13	6.9%
2010	171	148	86.5%	9	5.3%	8	4.7%
2011	164*	149	90.9%	5	3.%	10	6.1%
2012	160*	156	97.5%	4	2.5%	0	0%
2013	124*	119	96%	3	2.5%	2	1.5%

*holdings with confirmed CPH.

3.4 Abattoir survey

The requirement to test adult goats randomly selected at abattoirs was conducted for the last time in 2007. Summary data for the AS survey in goats from 2002 to 2007 are shown in Table 27.

Table 27 AS data summary in goats for the period 2002-2012

Year	Samples	Tested	Unsuitable	Clas.	% Clas. (95% CI)
2002	9	9	0	0	0 (0.00-33.6%)
2003	235	191	44	1	0.52 (0.01-2.88%)
2004	96	90	6	0	0 (0.00-4%)
2005	1344	1282	62	0	0 (0.00-0.29%)
2006	2563	2558	5	1	0.04 (0.00-0.22%)
2007	1467	1466	1	0	0 (0.00-0.25%)
2008	-	-	-	-	-
2009	-	-	-	-	-
2010	-	-	-	-	-
2011	-	-	-	-	-
2012	-	-	-	-	-
2013	-	-	-	-	-

3.5 Compulsory eradication measures (CSFS)

Since the CSFS came into force in 2004 seven goat holdings have been put under restrictions for having confirmed cases of classical scrapie: five holdings in England, one in Wales and one in Scotland. Six suspect cases were reported in 2013 in a holding under restrictions with a long history of scrapie. Although all of them were declared officially suspects and subsequently culled and tested, none resulted positive. Table 28 displays the number of clinical suspects notified from herds under CSFS restrictions during the period 2005-2012.

Table 28 Summary data of the clinical suspects notified by CSFS in goat holdings for the period 2005-2013 (allocated by slaughter date)

Year	Reported	Tested	Confirmed	Submitting holdings	% Tested not confirmed (95% CI)	Classical	% Classical (95% CI)	Atypical	% Atypical (95% CI)
2005/ 2008	49	49	49	2	0				
2009	-	-	-	-	-	-	-	-	-
2010	-	-	-	-	-	-	-	-	-
2011	5	5	1	1	80% (28.3 – 99.5%)	1	20% (0.5 – 71.6%)	-	-
2012	13	13	8	2	38.5% (13.9 – 68.4%)	8	61.5% (31.5 – 86.1%)	-	-
2013	6	6	0	1	100% (54.7 – 100%)	0	0% (0 – 45.9%)	-	-

Table 29 displays the number of samples submitted to the CSFS-FS and tested and numbers confirmed positive from goat holdings herds under restrictions during all or part of 2013. A total of 355 samples were submitted from two holdings and eight cases of classical scrapie were confirmed, all from the same holding. The proportion of the total samples suitable for testing in 2013 (2.3%), is not significantly different from previous years. The proportion of unsuitable samples in the fallen stock testing stream was 2.5%.

A total of 268 samples were submitted for testing to the CSFS-AC from the two holdings under restrictions and seven confirmed cases of classical scrapie were identified, six in one holding and one case in the other. All samples but one were suitable for testing (Table 29). As a proportion of the total samples suitable for testing in 2013 (267), seven confirmed cases (2.6%) is not significantly different from previous years, despite the difference in the number of cases confirmed. The proportion of unsuitable samples in the annual cull testing stream was very low: 0.3%.

The total number of samples collected in 2013 under the various testing streams of the CSFS from goat holdings was 623 from two different holdings: 57% (244) were fallen goats (CSFS-FS) and 43 % (891) were healthy sheep slaughtered for human consumption (CSFS-AC) (Table 29).

Table 29 Numbers of samples submitted, tested and confirmed scrapie positive under each of the CSFS testing routes in goats for the period 2005 – 2013

Initial Cull (CSFS-IC)

Year	Holdings	Samples	Tested	Unsuitable (%)	Clas.	% Clas. (95% CI)
2005	-	-	-	-	-	-
2006	-	-	-	-	-	-
2007	-	-	-	-	-	-
2008	2	366	366	0	80*	21.8% (17.7 – 26.4%)
2009	-	-	-	-	-	-
2010	-	-	-	-	-	-
2011	-	-	-	-	-	-
2012	-	-	-	-	-	-
2013	-	-	-	-	-	-

* An animal was considered positive if any of the Central Nervous System (CNS), or Lymphoreticular System (LRT) samples tested positive to Rapid test or to Immunohistochemistry.

Fallen Stock (CSFS-FS)

Year	Holdings	Samples	Tested	Unsuitable (%)	Clas.	% Clas. (95% CI)
2005	2	104	46	58 (55.8%)	3	6.5 (1.4 - 17.9%)
2006	3	298	159	139 (46.6%)	7	4.4 (1.8 - 8.9%)
2007	4	379	297	82 (21.6%)	31	10.4 (7.2 - 14.5%)
2008	4	194	168	26 (13.4%)	15	8.9 (5.1 - 14.3%)
2009	4	410	402	8 (2%)	4	1 (0.3 - 2.5%)
2010	5	530	505	25 (4.7%)	7	1.4 (0.6 - 2.8%)
2011	3	518	517	1 (0.2%)	5	1 (0.2 - 2.2%)
2012	3	249	239	10 (4%)	8	3.3 (1.5 - 6.5%)
2013	2	355	346	9 (2.5%)	8	2.3 (1 – 4.5%)

Annual Cull (CSFS-AC)

Year	Holdings	Samples	Tested	Unsuitable (%)	Clas.	% Clas. (95% CI)
2009	1	106	105	1 (0.9%)	1	0.9 (0.02 - 5.2%)
2010	3	227	227	0	0	0 (0 - 1.6%)
2011	2	249	246	3 (1.2%)	1	0.4 (0.01 - 2.2%)
2012	3	351	351	0 (0%)	4	1.1 (0.3 - 2.9%)
2013	2	268	267	1 (0.3%)	7	2.6 (1 – 5.3%)

All active CSFS testing routes (IC, FS and AC)

Year	Holdings	Samples	Tested	Unsuitable (%)	Clas.	% Clas. (95% CI)
2005	2	104	46	58 (55.8%)	3	6.5 (1.4 - 17.9%)
2006	3	298	159	139 (46.6%)	7	4.4 (1.8 - 8.9%)
2007	4	379	297	82 (21.6%)	31	10.4 (7.2 - 14.5%)
2008	4	560	534	26 (13.4%)	95	17.8 (14.6-21.3%)
2009	4	516	507	9 (1.7%)	5	1 (0.3 - 2.3%)
2010	5	757	732	25 (3.3%)	7	0.96 (0.39-1.96%)
2011	3	767	763	4 (0.5%)	6	0.79 (0.29-1.7%)
2012	3	600	590	10 (1.6%)	12	2 (1-3.5%)
2013	2	623	613	10 (1.6%)	15	2.4 (1.4-4%)

3.6 All sources

The contribution of the different sources to the overall number of samples tested and to the number of cases of both types of scrapie in goats in 2013 is shown in Figure 24.

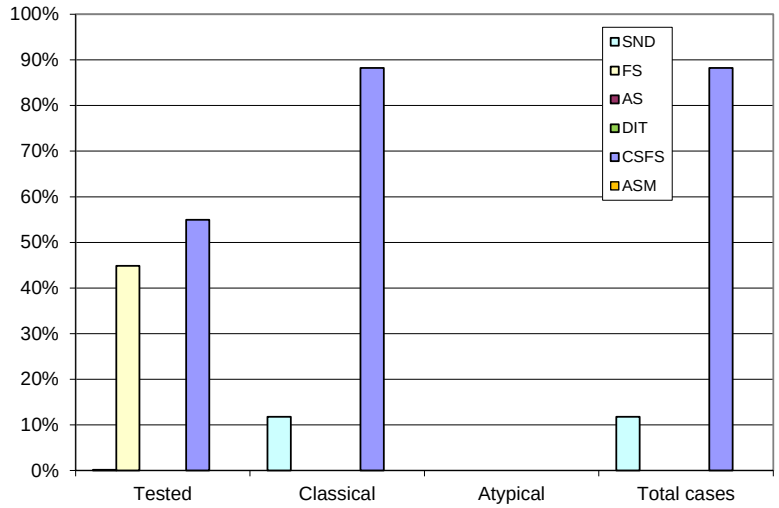


Figure 24 Percentage contribution of the six surveillance sources to the number of tested samples and scrapie in goats in 2013

The seventeen cases of classical scrapie were detected by passive surveillance (2), and the CSFS (15). The CSFS remains the main source of cases of scrapie in goats, due to the contribution of few but large and heavily infected herds. Figure 25 shows the changes in relative contribution to confirmed cases of classical scrapie in goats provided by the different surveillance sources since 2002.

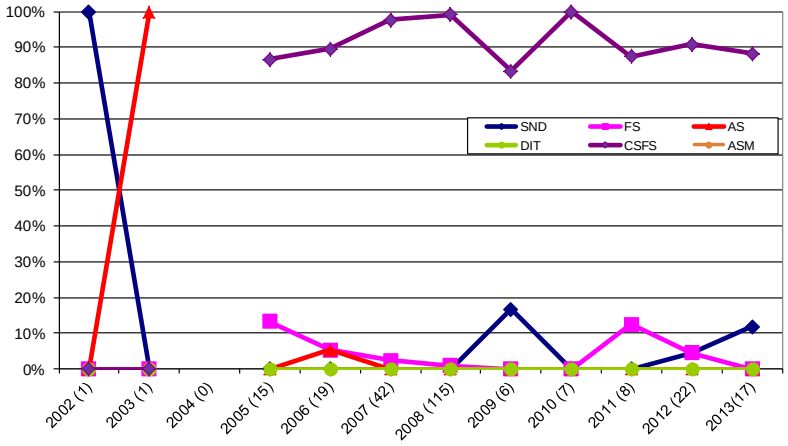


Figure 25 Percentage contribution of the six surveillance sources to the number of cases of classical scrapie in goats for the period 2002-2013