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The surveillance programme for scrapie in Norway 2024

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Content

Summary

In 2024, 18,318 sheep and 629 goats were examined for prion protein scrapie (PrP^{Sc}). Nine sheep originating from nine flocks were positive for Nor98 scrapie, also called atypical scrapie. All the goats were negative for PrP^{Sc}. In the period between 1981 and 2024, scrapie was diagnosed in 287 sheep flocks, of which 230 was Nor98 scrapie. One goat herd was diagnosed with Nor98 scrapie.

Introduction

Scrapie was first diagnosed in indigenous Norwegian sheep in 1981. Increasing numbers of scrapie-infected flocks were identified in the 1990s, culminating with 31 detected flocks in 1996 (Figure 1).

Based on these findings the Norwegian scrapie surveillance programme was launched in 1997 (1).

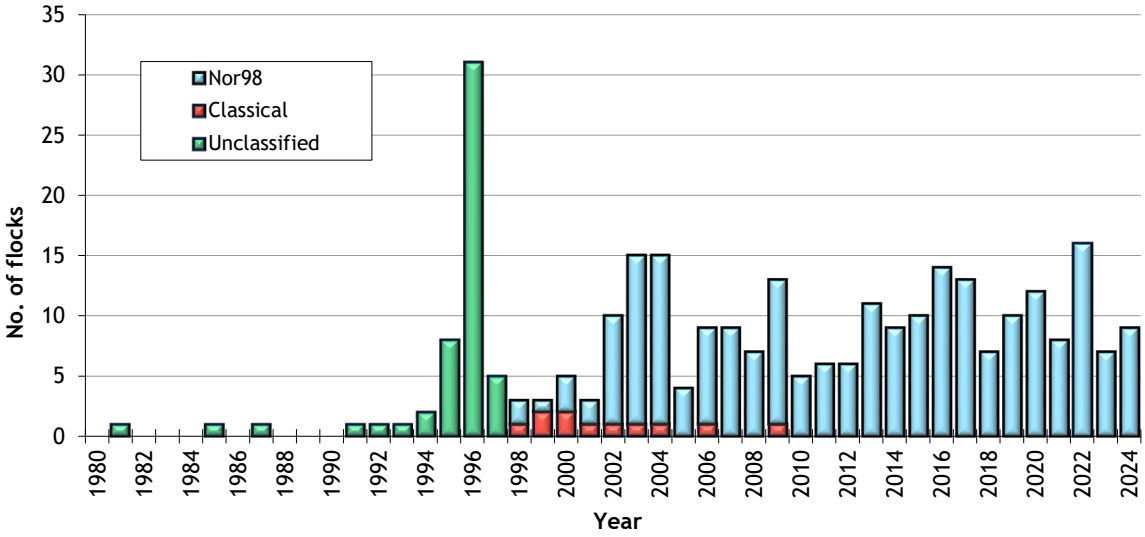

Figure 1. Annual number of sheep flocks and goat herds diagnosed with classical scrapie and Nor98 scrapie during the time period 1980-2024. Before 1998 the cases were not classified according to type of scrapie, but the majority of the scrapie cases, if not all, are considered to have been the classical type.

In 1998, a new type of scrapie, Nor98 scrapie, was identified in Norway. The diagnosis of Nor98 scrapie is verified by Western blot. Nor98 scrapie differs from classical scrapie in several aspects, including a higher age, the Western blot profile and distribution in the brain of the protease resistant prion protein (PrP^{Sc}), and particularly the absence of detectable PrP^{Sc} in lymphoid tissues (2). The main clinical sign observed in Nor98 scrapie cases is ataxia and abnormal behaviour. The PrP genotype distribution among Nor98 scrapie cases differs markedly from that of the cases with classical scrapie (3). Nor98 scrapie is considered to be not contagious or only contagious to a low degree between live animals under field conditions, and is hypothesised to have a spontaneous aetiology.

Scrapie has been a notifiable disease in Norway since 1965. The control measures have involved destruction of all sheep in affected flocks and in close contact flocks. Since 2004, the destruction of all sheep has only been carried out in flocks affected by classical scrapie.

The Norwegian Food Safety Authority (NFSA) is responsible for carrying out the surveillance programme for scrapie. The samples from small ruminants with clinical signs consistent with scrapie and sheep at the abattoirs are collected by inspectors from the NFSA. The NFSA carries out inspections of sheep flocks and goat herds. Since 2019, the company Biosirk, a company collecting and rendering fallen stock, performs the sampling of brains

from fallen stock from most parts of Norway on behalf of the NFSA. The employees involved, mainly truck drivers, have been trained in the sampling by attending courses arranged by the NFSA.

The Norwegian Veterinary Institute (NVI) performs the laboratory examinations and the reporting of the results.

Aim

The aim of the surveillance program is detection and eradication of the infective agent if present, documenting absence of disease in sheep slaughtered for human consumption and documenting the disease prevalence according to international regulations.

Materials and methods

In 2024, the surveillance programme was performed according to Regulation (EC) No. 999/2001 Annex III with amendments, and included examination of the following categories of small ruminants:

- all small ruminants with clinical signs consistent with scrapie, irrespective of age.
- imported small ruminants, irrespective of age.
- dead, killed or slaughtered small ruminants older than 18 months from herds with restrictions and their contact flocks.
- 10,000 sheep older than 18 months, which had died or been killed on the farm (fallen stock).
- 10,000 randomly sampled healthy sheep older than 18 months slaughtered for human consumption.
- 500 goats older than 18 months which had died or been killed on the farm (fallen stock).

Animals with clinical signs consistent with scrapie

When farmers identified clinical signs of scrapie in their sheep or goats, they were responsible for reporting the case to the NFSA.

If indicated, the animals were subject to either post mortem examination at a laboratory, or formalin-fixed and unfixed brain halves and medial retropharyngeal lymph nodes were submitted for laboratory examination. All the samples were examined at the NVI.

Surveillance of fallen stock

The sheep and goat farmers were responsible for reporting to the NFSA small ruminants older than 18 months that died or were killed on the farm due to disease. Employees from Biosirk or inspectors from the NFSA collected the samples, which consisted of unfixed medulla oblongata and part of the cerebellum obtained through the foramen magnum using a plastic spoon specially designed for the purpose. The samples were examined at the NVI.

Abattoir surveillance

Brain samples from healthy sheep older than 18 months were collected by the NFSA. The sheep samples were collected at 22 abattoirs, which slaughter 99% of all slaughtered adult sheep in Norway.

To ensure an appropriate distribution of the samples, the inspectors at the local NFSA were asked to organise a representative sampling for each region and season. The sample selection should be designed to avoid overrepresentation of any group as regards to the origin, species, age, breed, production type or any other characteristic.

The brain samples consisted of medulla oblongata, and often a small part of the cerebellum and midbrain, obtained through the foramen magnum using the specially designed plastic spoon. The samples were examined at the NVI.

Laboratory examination procedures

A rapid test (HerdChek BSE-Scrapie Ag test, IDEXX) for detection of prions (PrP^{Sc}) was performed for all submitted samples on a pooled brain tissue sample of obex and cerebellum when both structures were available or on the obex only when cerebellum was missing. In case of inconclusive or positive result, a western blot analysis (TeSeE® Western Blot, Bio-Rad) was used as confirmative test. The differentiation between classical scrapie and Nor98 scrapie was based on the Western blot profile.

PrP genotyping

PrP genotyping was performed on all scrapie positive sheep. Genotyping of scrapie positive sheep was performed on unfixed brain samples at NVI. Genomic DNA was isolated using the DNeasy Blood & Tissue Kit (QIAGEN). Polymorphisms in the PrP gene were detected through Sanger sequencing of a PCR-generated product covering the entire coding region of the PRNP (forward primer 5' CAGTCATTCATTATGCTGCAGAC; reverse primer 5' CTATCCTACTATGAGAAAAATGAG).

From 2004 to 2016, genotyping of approximately 600 brain samples from healthy slaughtered sheep were performed each year to get an estimate of the PrP genotypes in the normal Norwegian sheep population.

Prevalence

The scrapie prevalences in the fallen stock and abattoir populations were estimated assuming an exact binominal distribution.

Results

Sheep

Nor98 scrapie was diagnosed in nine sheep, each from a different flock. Four cases were identified in fallen stock, while five cases was an apparently healthy animal slaughtered for human consumption (Table 1).

The individual age was registered, and the prion protein genotype examined for all scrapie cases (Table 2). All sheep, except one, had PrP genotypes with at least one allele with polymorphisms at codon 141 (AF141RQ) and/or codon 154 (AHQ). One sheep had the genotype ARR/ARR, which is the most resistant PrP genotype for classical scrapie.

In total, 18,359 samples from sheep were received. Of these, 41 samples were rejected because they contained unsuitable material. The numbers of animals examined within each category are presented in Table 1. The prevalence of Nor98 scrapie in the fallen stock of sheep was estimated to 0.04% [0.01-0.10%], (95% confidence interval, CI), and the prevalence of Nor98 scrapie in sheep slaughtered for human consumption was estimated to 0.06% [0.02-0.14%], (95% CI) (Figure 2).

For 43 (0.2%) examined samples (40 healthy slaughtered and tree fallen stock), the flock of origin was not reported. All the samples with unidentified origin of the flock were negative. In the event of a positive sample from slaughtered animals, the flock identity can be traced using the carcass number. The remaining 18,316 samples were collected from carcasses originating in 5,995 different sheep flocks. The mean number of animals sampled per flock was 3.1 (range 1-77), sheep slaughtered for human consumption in restricted flocks due to scrapie are excluded. From 789 flocks, more than five animals were tested. There were 22 occasions were more

than nine animals were sampled from the same flock on the same date and on one occasion 67 animals were sampled from the same flock at the same date. This may indicate that not all samples are taken randomly.

Goat

Scrapie was not detected in goats in 2024. In total, 629 samples from goats were received. For 3 of these, the flock of origin was not reported (one healthy slaughtered and two fallen stock). The numbers of animals examined within each category are presented in Table 1.

The collected samples originated from 239 different herds. The mean number of animals sampled per herd was 2.6 (range 1-18). From 19 herds, more than five animals were tested.

Table 1. Brain samples from sheep and goats submitted for examination for scrapie in 2024.

Reason for submission to the laboratory	Number of samples			
	Total	Positive	Negative	Rejected
Sheep - total	**18,359**	**9**	**18,309**	**41**
Animals with clinical signs consistent with scrapie	5	0	5	0
Fallen stock	10,160	4	10,116	40
Healthy slaughtered animals	8,194	5	8,188	1
Animals examined due to scrapie restricted flocks	0	0	0	0
Imported animals	0	0	0	0
Goats - total	**629**	**0**	**629**	**0**
Animals with clinical signs consistent with scrapie	0	0	0	0
Fallen stock	614	0	614	0
Healthy slaughtered animals	15	0	15	0
Animals examined due to scrapie restricted flocks	0	0	0	0
Imported animals	0	0	0	0

Table 2. Year of birth, reason for submission to laboratory examination, breed, prion protein genotype and type of scrapie of the scrapie cases detected in 2024.

Case no	Year of birth	Reason for submission to laboratory examination[1]	Prion Protein Genotype	Scrapie type
1	2020	Fallen stock	$AF_{141}RQ$/AHQ	Nor98
2	2019	Fallen stock	$AF_{141}RQ$/A $F_{141}RQ$	Nor98
3	2017	Fallen stock	$AF_{141}RQ$/ARR	Nor98
4	2017	Healthy slaughtered animal	ARR/ARR	Nor98
5	2020	Healthy slaughtered animal	AHQ/AHQ	Nor98
6	2019	Healthy slaughtered animal	AHQ/ARR	Nor98
7	2017	Healthy slaughtered animal	AHQ/ARR	Nor98
8	2016	Healthy slaughtered animal	AHQ/ARQ	Nor98
9	2018	Fallen stock	$AF_{141}RQ$/ARR	Nor98

[1]The categories are: Healthy slaughtered animals, animals killed under scrapie eradication measures, suspect clinical signs consistent with scrapie including animals showing clinical signs at ante-mortem inspection, fallen stock (monitoring of fallen stock including animals examined because of other diseases than scrapie).

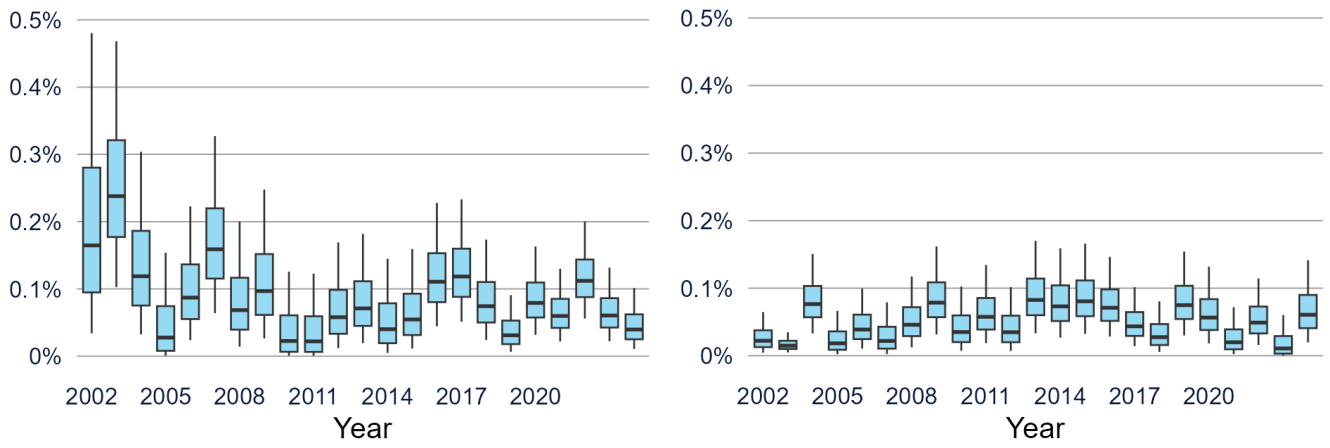

Figure 2. Box and whiskers plot of the prevalence of Nor98 scrapie from 2002 to 2024 in fallen stock (left) and slaughtered animals (right). The boxes represent the 25% to 75% quartiles and the whiskers the 2.5% and 97.5% exact binomial confidence intervals.

Discussion

Nor98 scrapie was diagnosed in nine sheep, originating from nine different flocks. The age and genotypes of these sheep were in accordance with the previous experience of Nor98 scrapie (5). All cases, except one, had at least one of the alleles AF141RQ or AHQ that previously have been found to be associated with Nor98 scrapie (3). The sheep positive for Nor98 scrapie were between four and eight years old, which are in agreement with the result from previous years with the mean age being seven years (Table 2).

The Nor98 scrapie cases detected in 2024 were located in five different counties. Nor98 scrapie cases have been found in most parts of Norway and in all counties except Oslo, the capital of Norway. In contrast, the classical form of scrapie, has been detected only in the western part of Norway (two counties) and in Nordland County. In 2024, the 11 Norwegian counties were separated into 15 counties, and all numbers refer to the counties as of 2024.

The prevalence estimates of Nor98 scrapie in fallen stock and in sheep slaughtered for human consumption have varied during 2002–2024; however most estimates have been within the confidence intervals (Figure 2) (4). The results from the surveillance programmes indicate that the prevalence of Nor98 scrapie in the sheep population has not changed since the start of the programme.

Scrapie was not detected in goats in 2024. The first and only Nor98 scrapie case in goats in Norway was diagnosed in 2006 and originated from a county with a large goat population. Both classical and atypical scrapie in goats has been diagnosed in several countries in Europe (5).

Acknowledgments

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