SURVEILLANCE OF INFECTIOUS DISEASES

IN ANIMALS AND HUMANS IN SWEDEN 2022

Chapter excerpt:
Scrapie

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Cover: A cultivation of *Salmonella* at the Public Health Agency of Sweden.
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Reporting guidelines: Reporting guidelines were introduced in 2018 for those chapters related to purely animal pathogens. The guidelines build on experiences from several EU projects, and have been validated by a team of international experts in animal health surveillance. The aim is to develop these guidelines further in collaboration within the global surveillance community and they have therefore been made available in the form of a wiki on the collaborative platform GitHub (https://github.com/SVA-SE/AHSURED/wiki). Feel free to contribute!

Layout: The production of this report continues to be accomplished using a primarily open-source toolset. The method allows the source text to be edited independently of the template for the layout which can be modified and reused for future reports. Specifically, the chapter texts, tables and captions are authored in Microsoft Word and then converted to the LaTeX typesetting language using a custom package written in the R software for statistical computing. The package uses the pandoc document conversion software with a filter written in the lua language. Most figures and maps are produced using R and the LaTeX library pgfplots. Development for 2022 has focused on generalising the R package to accommodate conversion into formats other than LaTeX and PDF, with a focus on markdown files which can be published as HTML websites using the Quarto publishing system. The report generation R package and process was designed by Thomas Rosendal, Wiktor Gustafsson and Stefan Widgren.

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Scrapie

BACKGROUND

Scrapie, which affects sheep and goats, belongs to a group of diseases called Transmissible Spongiform Encephalopathies (TSEs) and was first described more than 250 years ago. The current theory about the causative agent is the protein-only hypothesis. This theory assumes that prions, which are misfolded and aggregated proteins, induce the same misfolded, aggregated, and pathological structure in normal prion-proteins of the host, resulting in accumulation of prions and cellular damage without the involvement of any microorganism. Susceptibility to scrapie is genetically related and some countries have chosen to control the disease through specific breeding programmes.

Scrapie occurs in different variants, classical and atypical scrapie/Nor98. Classical scrapie, which is clearly transmissible within flocks, has been detected in Sweden on one occasion, in a single flock in 1986. The whole flock was culled, and the farmer was not allowed to reintroduce sheep for seven years. The origin of the disease was never established.

In 1998, an atypical variant of scrapie was detected in Norway (Nor98), and this variant was detected in Sweden for the first time in 2003. Since then, several cases have been detected in Sweden and worldwide. Although atypical scrapie is experimentally transmissible, cases occur sporadically and epidemiological studies on the European level indicate that atypical scrapie probably is a spontaneously (without known cause) occurring disease which does not seem to spread within or between flocks.

In 2002, after classical bovine spongiform encephalopathy (BSE) in cattle became a disease of public health concern (see chapter on BSE, page 21), and the occurrence of BSE in small ruminants was suspected, both surveillance and control of TSE in small ruminants was intensified within the European Union. Since the start of this intensified surveillance, more than 75 000 sheep have been tested in Sweden without any positive cases of classical scrapie detected. In 2014, Sweden sent an application to the European Commission to obtain status as a country with negligible risk

Figure 48: Classical scrapie has not been detected in Sweden since 1986 and after several years of intensive surveillance, Sweden has been granted the status "negligible risk" for classical scrapie. Photo: Ylva Persson.

for classical scrapie. The dossier contained detailed information about the population, imports (which were limited), education about the disease, the EU-approved national control programme as well as results of estimates of the probability that Sweden is free from classical scrapie. The Commission evaluated the dossier and also asked the European Food Safety Authority (EFSA) for an opinion (doi: 10.2903/j.efsa.2015.4292). In August 2016, the application was approved, and Sweden was granted the status negligible risk for classical scrapie through Commission regulation (EC) 2016/1396.

DISEASE

The incubation period of scrapie is long, up to several years. Clinical signs of classical scrapie are related to the neurological system and include altered behaviour and sensation, affected movement and posture, as well as pruritus and subsequent skin lesions. The disease is progressive and always fatal. All routes of transmission of classical scrapie have not been established, but transmission occurs horizontally within flocks and especially at lambing, as foetal fluid and the placenta may contain large amounts of prions. Prions may remain on contaminated pastures for long periods of time. Scrapie has, based on epidemiological data, not been considered a zoonotic disease. However, the question is still regularly raised. The majority of cases of atypical scrapie/Nor98 are detected in the active surveillance and there are fewer reports of animals with clinical signs such as e.g., ataxia, loss of body condition and abnormal behaviour.

LEGISLATION

Surveillance and control of scrapie in sheep and goats is regulated through Regulation (EC) 999/2001 of the European Parliament and of the Council of 22 May 2001. At the national level, the surveillance scheme and control were, until 2016, also regulated by an EU-approved national scrapie control programme which from 2003 also formed the basis for additional guarantees related to trade within the union (Commission Regulation (EC) 546/2006).

Sweden was granted the status: "negligible risk" for classical scrapie through Commission regulation (EC) 2016/1396 amending Regulation (EC) 999/2001 and since then the rules in 999/2001 replace both the additional guarantees and previous surveillance scheme in the national programme.

Scrapie is a notifiable disease under the Swedish Act of Epizootic diseases (SFS 1999:657, with amendments) and should be notified already on clinical suspicion. The obligation to report applies to animal owners, veterinarians, and everyone else who is responsible for the animals. Sampling at the national level is regulated by SJVFS 2010:9, last amended through SJVFS 2013:3.

SURVEILLANCE

The Swedish Board of Agriculture oversees the surveillance programme which is carried out in cooperation with the National Veterinary Institute (SVA). Samples are analysed at SVA, which is also appointed the National Reference Laboratory (Regulation (EC) 999/2001). A majority of the samples are collected at rendering and hence there is a close collaboration with Svensk Lantbrukstjänst and Konvex, two companies that collect and render carcasses.

Passive surveillance

If scrapie is suspected or cannot be excluded based on clinical signs, the animal is euthanised and samples collected. Brainstem samples were for the first part of 2022 analysed using the Bio-Rad TeSeE ™ SAP rapid test (Bio-Rad Laboratories, Hercules, California, United States). During 2022 the analysis method at SVA was changed and for the second part of the year samples were analysed using the IDEXX HerdChek® BSE-Scrapie Antigen Test kit (IDEXX Laboratories, Westbrook, Maine, United States).

If the results are positive or inconclusive, the TeSeE™ Western Blot kit (Bio-Rad Laboratories, Hercules, California, United States) is used for confirmation.

Active surveillance

From 2017, the basis of the active surveillance is Regulation (EC), 999/2001 Annex III, which states a minimum number of animals to be sampled based on population size. The minimum number to be sampled in Sweden is 1500 fallen sheep and 100 fallen goats above the age of 18 months. The samples should be representative for the population. Prior to 2017 sampling was based on an EU-approved national control programme, which included sampling of all dead sheep and goats over 18 months of age that were not slaughtered for human consumption.

The current national purpose of the surveillance is to demonstrate freedom of the disease so that Sweden can maintain the official status of negligible risk as well as to detect any possible introduction. Regulation (EC) 999/2001 requires that for the preceding 7 years, a sufficient number of animals should have been tested annually to provide a 95% confidence of detecting classical scrapie if it is present in that population at a prevalence exceeding 0.1%.

It is mandatory to send fallen animals for rendering, except for in the northern parts of Sweden, where animal density is low (less than 10% of the sheep population is in this area). In the computerised system for collecting carcasses, roughly every second or every third (adjusted by season) animal is "flagged" for sampling. The carcasses sent for rendering are sampled by employees at the rendering plants. All sheep and goats above 18 months of age that are sent for post-mortem examinations are sampled by veterinarians or veterinary assistants.

Samples from the active surveillance were for the first part of 2022 analysed using the Bio-Rad TeSeE ™ SAP rapid test (Bio-Rad Laboratories, Hercules, California, United States). During 2022 the analysis method at SVA was changed, and the HerdChek® BSE-Scrapie Antigen Test kit (IDEXX Laboratories, Westbrook, Maine, United States) was used at SVA in accordance with Regulation (EC) 999/2001. If results are positive or inconclusive the Bio-Rad TeSeE ™ Western Blot kit was used for confirmation.

The number of samples and distribution between farms is followed up on a monthly basis.

RESULTS

Passive surveillance

In 2022, one sheep was tested due to clinical suspicion of scrapie with a negative result.

Active surveillance

Sheep

In 2022, SVA examined 1580 sheep from fallen stock. All samples were negative for classical scrapie and three samples were positive for atypical scrapie/Nor98. The northern part of the country is under-represented in the sampling and due to problems with rapid decomposition of carcasses during the summertime, sampling is not evenly distributed throughout the year. Apart from this, sampling is considered representative.

Goats

In 2022, SVA examined 109 goats from fallen stock for scrapie. All were negative both for classical scrapie and for atypical scrapie/Nor98.

DISCUSSION

Classical scrapie

Classical scrapie is a challenging disease to both detect and to eradicate, due to the long incubation period and persistence in the environment. Sweden has chosen not to breed for resistance and thus the sheep population is susceptible to classical scrapie. An introduction of the disease could therefore potentially have negative consequences for the sheep industry. The import of sheep and goats to Sweden has for many years been limited, and in combination with trade requirements, this has kept the risk of introduction at a low level.

In the active surveillance, no positive cases have been detected but continued efforts need to be made to increase the number of samples from the northern parts of the country. From a surveillance point of view, the seasonal variation with reduced sampling during the summer, is not deemed to have a systematic effect.

Atypical scrapie

Since the first case of atypical scrapie was confirmed in Sweden in 2003, more than 50 cases have been detected. Out of these, two were detected through passive surveillance and the remaining cases through active surveillance. At the European level, two epidemiological studies have concluded that the prevalence is similar in different countries, and that the prevalence in positive flocks does not differ from the prevalence in the rest of the sampled population. This pattern differs from the way a contagious disease normally is distributed in a population and supports the hypothesis that atypical scrapie is spontaneously occurring. Although within flock transmission between animals seems to be very low (if it exists) other routes of spread and the potential zoonotic aspect is regularly subject to discussion.

As a measure to further the knowledge of atypical scrapie/Nor98, farms with confirmed cases were for a number of years obligated to carry out increased surveillance in the herd for two years (Regulation (EC) 999/2001). In 2021 EFSA published a report on the analysis of this intensified surveillance which reached similar conclusions as the two previous publications. The increased surveillance was discontinued in 2021 (Commission Regulation (EU) 2021/1176).

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