SURVEILLANCE OF INFECTIOUS DISEASES IN ANIMALS AND HUMANS IN SWEDEN 2019

Chapter excerpt -

Scrapie

SVA NATIONAL VETERINARY INSTITUTE
Jordbruksverket Swedish Board of Agriculture
Folkhälsomyndigheten PUBLIC HEALTH AGENCY OF SWEDEN
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Editor: Karl Ståhl
Department of Disease Control and Epidemiology, National Veterinary Institute (SVA), SE-751 89 Uppsala, Sweden.

Authors: Charlotte Axén, Mia Brytting, Ioana Bujila, Erika Chenais, Rikard Dryselius, Helena Eriksson, Pernille Etterlin, Eva Forsgren, Malin Grant, Gittan Gröndahl, Gunilla Hallgren, Anette Hansen, Gete Hestvik, Marika Hjertqvist, Mia Holmberg, Cecilia Hultén, Helena Höök, Cecilia Jernberg, Jerker Jonsson, Ulrika König, Elina Lahti, Emelie Larsdotter, Mats Lindblad, Anna Lundén, Emma Löf, Hans-Olof Nilsson, Oskar Nilsson, Maria Nöremark, Anna Ohlson, Ylva Persson, Karin Persson-Waller, Thomas Rosendal, Karl Ståhl, Robert Söderlund, Kaisa Sörén, Magnus Thelander, Karin Troell, Anders Wallensten, Per Wallgren, Stefan Widgren, Ulrika Windahl, Beth Young, Nabil Yousef, Siamak Zohari, Erik Ågren, Estelle Ågren, Elina Åsbjer

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Reporting guidelines: Reporting guidelines were introduced in 2018 for those 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, produced by authors, 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 using pandoc and R to the LaTeX typesetting language. Most figures and maps are produced using the R software for statistical computing. Development for 2019 has further improved the importing of content from Word to LaTeX. The method can now import text, tables and figure captions from Word, as well as the newly designed 'IN FOCUS' sections of some chapters. The tool is available as an R-package at GitHub (https://github.com/SVA-SE/mill/). This year the report was also built with a continuous integration pipeline on Microsoft's Azure DevOps platform, allowing every committed change to the content to be built and tested automatically. The report generation R-package and process was designed by Thomas Rosendal and Stefan Widgren. In 2019, figures and the final typsetting were done by Wiktor Gustafsson and Thomas Rosendal with contributions from the report authors.

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SVA NATIONAL VETERINARY INSTITUTE

visiting address. Ulls väg 2B **address.** 751 89 Uppsala
telephone. +46 18-67 40 00 **fax.** +46 18-30 91 62
e-mail. sva@sva.se **web.** www.sva.se

Scrapie

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: Whiptop/Shutterstock.

BACKGROUND

Scrapie, which affects sheep and goats, belongs to a group of diseases called Transmissible Spongiform Encephalopathies (TSE) 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 misfolded prions (small proteins) induce the same misfolded and pathological structure in normal prion-proteins of the host, resulting in accumulation of prions and cellular damage without involvement of any microorganism. Susceptibility to scrapie is genetically related and some countries has chosen to control the disease through specific breeding programs.

Scrapie occurs in different variants; classical and atypical scrapie. Classical scrapie, which is clearly transmissible within flocks, has been detected in Sweden 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 also 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, epidemiological studies on the European level indicate that atypical scrapie probably is a spontaneously occurring disease which does not seem to spread within or between flocks.

After classical BSE in cattle became a disease of public health concern (see chapter on BSE), and the existence of BSE in small ruminants was suspected, both surveillance and control of TSE in small ruminants was increased within the European Union in 2002. Since the start of the increased 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 country with negligible risk 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 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 skin lesions. The disease is progressive and always fatal. All routes of transmission of classical scrapie have not been established, however, transmission of classical scrapie occurs horizontally within flocks and at lambing, and pastures can be contaminated for long periods of time. Scrapie has, based on epidemiological data, not been considered a zoonotic disease; however, the question is regularly raised.

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 program.

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. This legislation cover compensation to farmers for financial losses due to eradication measures. Sampling at the national level is regulated by SJVFS 2010:9, last amended through SJVFS 2013:3.

SURVEILLANCE

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

Passive surveillance

All suspicions of scrapie, i.e. sheep or goats showing clinical signs or post mortem findings where scrapie cannot be excluded, must be reported to the authorities. The obligation to report applies to animal owners, veterinarians and everyone else who is responsible for the animals. If the animal is still alive it is examined by a veterinarian who is in close contact with disease experts. If scrapie can still not be excluded the animals is euthanized. Brainstem samples from animals with clinical suspicion of scrapie are examined with Bio-Rad TeSeE short assay protocol (SAP). If inconclusive or positive the results are confirmed with Bio-Rad TeSeE Western Blot.

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 is 1500 fallen sheep and 100 fallen goats above the age of 18 months. The samples should be representative for the population.

The current national purpose of the surveillance is to maintain freedom (negligible risk) and to detect introduction. Regulation (EC) 999/2001 requires that for the preceding 7 years, sufficient numbers 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%.

Except for the northern parts of Sweden, where animal density is low (less than 10% of the sheep population are in this area), it is mandatory to send fallen animals for rendering. 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 sent for post mortem examinations are sampled. This is done by veterinarians or veterinary assistants.

Prior to 2017 sampling was based on an EU-approved national control program, which included sampling of all dead sheep and goats over 18 months of age that were not slaughtered for human consumption.

Farms with confirmed cases of atypical scrapie are obligated to have increased surveillance in the herd for two years (Regulation (EC) 999/2001). In addition to fallen stock, healthy slaughtered animals above 18 months of age are examined from these flocks. These animals are sampled at slaughterhouses by trained employees or inspectors employed by the National Food Agency.

The samples from active surveillance were examined with Bio-Rad TeSeE short assay protocol (SAP) at the National Veterinary Institute in accordance with Regulation (EC) 999/2001. In case of positive or inconclusive results the material was examined by Bio-Rad TeSeE Western Blot.

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

RESULTS

Passive surveillance

In 2019, one sheep was examined due to clinical suspicion of scrapie and tested with negative results.

Active surveillance

Sheep

In 2019, the National Veterinary Institute examined 1285 sheep from fallen stock and 32 sheep from flocks under increased surveillance due to Nor98 or under restrictions due to imports, sampled at slaughter. Out of these, all samples were negative for scrapie. The northern part of the country is

under-represented in the sampling and due to problems with rapid decomposition of carcasses during summertime, sampling is not evenly distributed throughout the year. Apart from this, sampling seems fairly representative.

Goats

In 2019, the National Veterinary Institute examined 84 goats from fallen stock for scrapie. All were negative both for classical scrapie and for atypical scrapie.

DISCUSSION

Classical scrapie

Classical scrapie is a challenging disease both to detect and 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. This means that introduction of the disease would potentially have negative consequences for the sheep industry. Imports of sheep and goats to Sweden have for many years been limited and in combination with trade requirements this has kept the risk for introduction at a low level. Within the European union, relaxation of current trade rules is being discussed. For Sweden, and other countries with a susceptible population and negligible risk, it is important that trade rules that minimise risk for introduction of classical scrapie to the country are kept in place.

Regarding the active surveillance, no positive cases have been detected. Continued efforts need to be made to increase samples from the northern parts of the country. From a surveillance point of view, a seasonal variation with less samples during summer is not deemed to have a systematic effect. There was only one reported clinical suspicion of scrapie and efforts are needed to improve passive surveillance.

Atypical scrapie

Since the first case of atypical scrapie was detected in Sweden in 2003, more than 50 cases have been detected. Out of these, two were detected through passive surveillance and the rest through active surveillance. Currently, the flocks are put under intensified monitoring in accordance with Regulation (EC) 999/2001. 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 contagious disease are normally distributed in the 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.

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