

SURVEILLANCE OF INFECTIOUS DISEASES

IN ANIMALS AND HUMANS IN SWEDEN 2022

*Chapter excerpt:
Echinococcosis*



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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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Echinococcosis



Figure 18: Through the national screening for *Echinococcus multilocularis* in red foxes, which started in 2021, 911 faecal samples were collected in 2022. Two of them were positive, both from the county of Dalarna. Photo: Ondrej Prosicky/iStock.

BACKGROUND

Echinococcosis is a common name for different diseases in humans caused by tapeworms belonging to the genus *Echinococcus*. The genus contains several species, of which *E. multilocularis* is the causative agent of alveolar echinococcosis, while cystic echinococcosis (hydatid disease) is caused by species within the *E. granulosus* sensu lato (s.l.) complex, mainly *E. granulosus* sensu stricto (s.s.), but also other species such as *E. canadensis* and *E. ortleppi*.

The life cycles of these parasites are similar with carnivorous definitive hosts and intermediate herbivorous/omnivorous intermediate hosts. However, host ranges vary between the different *Echinococcus* species. Humans are dead-end hosts and may become infected by accidental ingestion of eggs shed by the definitive host.

ALVEOLAR ECHINOCOCCOSIS

Background

Echinococcus multilocularis is endemic in large parts of Europe and has a reported increasing geographical range. Although a rare disease in humans, alveolar echinococcosis is of considerable public health concern due to its high mortality if untreated as well as high treatment costs. The definitive hosts of this parasite are mainly foxes, but raccoon dogs,

dogs, coyotes and wolves can also act as definitive hosts. Rodents, mainly voles, serve as intermediate hosts. Foxes contract *E. multilocularis* by eating infected rodents.

Prior to 2010, *E. multilocularis* had not been detected in any definitive host, and no case of alveolar echinococcosis had been reported in Sweden. As a response of finding *E. multilocularis* in foxes in Denmark in 2000 an active monitoring programme of red foxes (*Vulpes vulpes*) was implemented in Sweden. From 2000 to 2010 approximately 300 foxes per year were examined, all with negative results, until *E. multilocularis* was found in a fox shot in December 2010 in the county of Västra Götaland.

In the spring of 2011, a national screening was performed in which 2985 hunter-harvested foxes were analysed. Three foxes were found positive: one in Västra Götaland, one in Södermanland and one in the county of Dalarna. The national screening continued in 2012 – 2014 and 2779 fox scat samples were analysed. Three samples were positive: one from Gnesta, one from Katrineholm (both in Södermanland) and one from Västra Götaland. During this period the parasite was also detected in fox scats from an area near the town Växjö in the county of Kronoberg.

To obtain a better prevalence estimate in a known infected area, fox scats were collected in 2011, by a systematic sampling procedure from a circular area with a diameter of 25 km surrounding a positive finding in Södermanland. Six out of 790 (0.8%) faecal samples were positive. Also, to follow up the positive findings from the five areas where the parasite had been detected so far, hunters were asked to submit 30 foxes from an area with a diameter of 40 km. The sampling started in 2012 and continued to 2016. In Västra Götaland two foxes were positive, in Södermanland three foxes from Katrineholm and one from Gnesta were positive, whereas all tested foxes from Dalarna and Kronoberg were negative.

In 2020, fox scats were collected again from the areas in Uddevalla, Gnesta, and Katrineholm where the parasite has been previously found. The results revealed that it was still present in two of the three areas (12 of 109 fox scats from Uddevalla, Västra Götaland and 7 of 18 from Gnesta, Södermanland were positive), while none of 108 samples from the area in Katrineholm, Södermanland tested positive. In 2021 a follow-up targeted screening was conducted in Gnesta and Uddevalla to investigate potential spread from these hot spots. A circular area up to 5 km from each of the original finding sites was sampled. At both locations positive samples were found near the perimeter of the extended area. In total, 10 of 63 samples from Gnesta and 5 of 102 samples from Uddevalla were positive.

Potential intermediate hosts have also been examined and the parasite was first found in voles caught in the county of Södermanland in 2013 (Gnesta/Nyköping). One out of 187 field voles (*Microtus agrestis*) and eight out of 439 water voles (*Arvicola amphibius*) had metacystode lesions confirmed by PCR and sequencing. Protoscoleces were demonstrated in the *Microtus agrestis* and in three out of eight *Arvicola amphibius*. No lesions were found in bank voles (*Myodes glareolus*; $n=655$) or mice (*Apodemus* spp.; $n=285$).

In 2012, alveolar echinococcosis was diagnosed in humans in Sweden for the first time. There were two human cases with clinical symptoms, and both were considered to have been infected abroad. No human cases were diagnosed in 2013 to 2015. From 2016 and onwards, there have been zero to four cases reported yearly.

Disease

Animals

In the definitive animal host, the infection is asymptomatic. The main intermediate hosts, rodents, will usually die from the infection if not captured by a predator.

Humans

In humans, alveolar echinococcosis may develop into a serious, potentially fatal disease characterised by infiltrative tumour-like lesions in the affected organ. The incubation period for developing alveolar echinococcosis in humans is assumed to be between 5 and 15 years. Because of the long incubation period, the disease is most frequently seen in adults. The most common site of localisation is the liver, but other organs can also be affected. Symptoms depend on the site and size of the lesion.

Legislation

Animals

Detection of the parasite is notifiable according to Swedish legislation (SJVFS 2021:10).

Humans

Infection with *Echinococcus* spp. has been notifiable since 2004 according to the Communicable Disease Act (SFS 2004:168 with the amendments of SFS 2022:217). However, notification at the species level is not required. If cases of *E. multilocularis* occur in humans, the data will be presented in the annual report at the website of the Public Health Agency of Sweden (folkhalsomyndigheten.se). Before 2004, *Echinococcus* spp. was reported on a voluntary basis by the laboratories.

Surveillance

Animals

As *E. multilocularis* does not cause clinical signs in the definitive host, surveillance in these species must either be active or enhanced passive for example by collection of materials from animals submitted for other reasons. Since 2012, all free-living wolves submitted to necropsy at the National Veterinary Institute (SVA) have been tested with MC-PCR, without any positive finding. Furthermore, a second national screening funded by the Board of Agriculture was initiated in 2021. During this three-year project, up to 3000 samples (faecal samples from dead foxes and fox scats) will be collected and analysed with MC-PCR. Collaboration with field staff from the Swedish Association for Hunting and Wildlife Management and calls to the public to contribute with samples in this citizen science project is used to receive samples from the entire country.

Humans

Surveillance is passive and based on identification of the disease by a treating physician or by laboratory diagnosis. Both the physician and the laboratory are obligated to report identified cases to the regional and national level to enable further analyses and adequate intervention measures.

Results

Animals

Within the ongoing national screening that started in 2021 a total of 1089 fox scats or faecal samples from red foxes had been collected at the end of 2022. One of the 178 samples from 2021 tested positive with MC-PCR. Thereby a new infected area was identified in Kungsbacka in the county of Halland in south-west Sweden. By April 2023, 899 of the 911 samples collected in 2022 had been analysed, and two of these were positive. Both were from the county of Dalarna, one from Borlänge and one from Avesta (approximately 60 km apart). Of note is that in 2011 the parasite was found in one fox from Borlänge, and although around 30 foxes from the surrounding area were tested during the following years it has not been detected there again until 2022. Furthermore, 6 of 23 samples collected in the area around Gnesta, Södermanland, where the parasite has been detected repeatedly

since 2011, tested positive. In addition, 47 wolves (*Canis lupus lupus*) and two domestic dogs were tested with the MC-PCR and all were negative.

Humans

In 2022, molecular techniques confirmed infection of *E. multilocularis* (alveolar echinococcosis) in two women, median age 63 years.

It could not be determined if these cases were infected in Sweden or abroad. It is sometimes difficult to determine the exact country of infection, due to the long incubation period, from when the infection was acquired until the debut of symptoms.

Discussion

E. multilocularis occurs sporadically in Sweden. It is not known how and when the parasite was introduced into the country. The national screening finalised in 2014 can be used as a baseline estimate of the national prevalence, against which future trends can be assessed. It is well known from other countries that the prevalence of this parasite varies geographically. In Sweden, regional screenings have previously shown a prevalence of more than 1% in a part of the county of Södermanland, and within a research project 18 of 80 (22%) fox scats were found to be positive in one of four investigated small areas (Miller et al., 2016). However, the true geographical distribution is unknown but so far, no positive cases have been found north of the county of Dalarna. Up until 2020, the infection had been detected in five different areas. The repeated finding of positive fox scats in two of these areas shows that the parasite is still present in these locations (at least until 2021).

E. multilocularis has also been found in intermediate hosts, for the first time in 2013. This finding increases our knowledge about in which biotypes the life cycle of the parasite can be completed. It has been suggested that the absence of common voles (*Microtus arvalis*) in Sweden may be a contributing factor to the low prevalence of the parasite. However, in some small areas, prevalence has been reported to be higher and more research is needed to clarify which intermediate host(s) are most important.

Based on the knowledge available today, there is a risk for occasional cases of alveolar echinococcosis acquired in Sweden in the future, but the infection will most likely continue to be very rare in humans.

CYSTIC ECHINOCOCCOSIS

Background

Cystic echinococcosis is caused by *Echinococcus granulosus* s.l. and domestic dogs and wolves are the most frequent definitive hosts. Eggs of the parasite are excreted in faeces into the environment where they can infect intermediate hosts such as sheep, pigs, cattle, horses and wild ruminants. The eggs develop into the larval stage (hydatid cyst) mainly in the liver but also in other organs of the intermediate host. The definitive hosts become infected by consuming organs containing hydatid cysts.

Echinococcosis was quite common in reindeer in the

northern parts of Scandinavia in the first half of the 20th century. In the 1990s, single cases of *E. granulosus* s.l. were detected in moose and reindeer in Sweden. Since then, the parasite has not been detected in any intermediate host, except sporadic cases in horses imported from Great Britain or Ireland where they most likely had acquired the infection. However, in a retrospective study of biobank material from 116 wolves submitted to SVA during 2012–2020, faecal samples from two wolves culled in 2012 tested positive with a MC-PCR detecting *E. canadensis* genotype 8/10 as well as *E. ortleppi*.

Disease

Animals

In animals, the infection is usually asymptomatic.

Humans

In humans, the main site for cystic echinococcosis is the liver. However, it may also be located in the lungs, brain or other tissues. Infected patients may remain asymptomatic for years or permanently. Clinical signs of disease depend on the number of cysts, their size, localisation and pressure exerted on surrounding organs or tissues. The incubation period for developing cystic echinococcosis ranges from one to several years.

Legislation

Animals

Detection of the parasite is notifiable in all animals according to (SJVFS 2021:10).

Humans

Echinococcosis has been notifiable according to the Communicable Disease Act since 2004 (SFS 2004:168 with the amendments of SFS 2022:217). However, notification on species level is not required. If cases of *E. granulosus* occur in humans, the data will be presented in the annual report at the website of the Public Health Agency of Sweden (folkhalsomyndigheten.se). Before 2004 *Echinococcus* spp. was voluntarily reported by the laboratories.

Surveillance

Animals

At slaughter, all livestock are inspected for cysts during routine meat inspection. Semi-domesticated reindeer are inspected at slaughter, but not all free-ranging hunted cervids are inspected. If cysts, that could be hydatid cysts, are found in the liver or lung they should be sent to the SVA for diagnosis.

Since 2012, all free-living wolves submitted to necropsy at the SVA have been tested with MC-PCR, without any positive finding.

Humans

Surveillance is passive and based on identification of the disease by a treating physician or by laboratory diagnosis. Both the physician and the laboratory are obligated to report identified cases to the regional and national level to enable further analyses and adequate intervention measures.

Results

Animals

In addition to the routine inspection at slaughter, 47 wolves were tested by MC-PCR detecting *E. canadensis* genotype G8 and G10 as well as *E. ortleppi*, and all were negative.

Humans

During 2022, 20 human cases of infection with *Echinococcus* spp. were reported, most probably infected with *E. granulosus* s.l. (cystic echinococcosis), based on epidemiological data and the geographical distribution of the various *Echinococcus* species. Median age was 35 years (range 13–78 years) and slightly more men than women were infected (n = 11). All of these cases were deemed to have been infected abroad. The most common countries of infection were Iraq (n=5), Syrian Arab Republic (n=4) and Afghanistan (n=3).

Discussion

Echinococcus granulosus s.l. is very rarely detected in animal intermediate hosts in Sweden. In reindeer it has not been detected since the late 1990s, when it was reported in three reindeer in the northernmost regions of Sweden, bordering to Norway and Finland. However, retrospective analysis of biobank samples from 2012–2020 has revealed that two wolves culled in 2012 were infected with genotype G8/10 (or possibly G5). In Finland, the parasite is present at a low prevalence in wildlife (wolves, moose and reindeer) and has been genotyped as *E. canadensis* (G10). Retrospective analysis of one of the three above mentioned Swedish reindeer revealed the same genotype. This species is considered as less pathogenic, and possibly with a lower zoonotic potential, than *E. granulosus* s.s. that is prevalent in some other parts of Europe and identified mainly in a cycle between dogs and farm animals.

Hydatid cysts are also occasionally found in horses at slaughter with the latest case in 2021. The horse had been imported from Ireland in 2015, and parasite material was genotyped as *E. equinus* (G4, horse strain). Cystic equine echinococcosis is a very rare finding in Sweden with the latest previous case reported in 2011. Furthermore, there is no indication of any transmission in Sweden, since the infection has only been diagnosed in imported horses. The absolute majority of these have been from the United Kingdom or Ireland, where *E. equinus* is known to be endemic.

This species is regarded as specific for equids as intermediate hosts, and with a very low zoonotic potential.

In humans, cystic echinococcosis is a rare disease seen in immigrants or other people who have resided in endemic countries. In Sweden, no domestically acquired human cases have been reported since the infection became notifiable. In Finland, on the other hand, pulmonary cystic echinococcosis (*E. canadensis*) was confirmed in 2015 in a patient with no history of travelling abroad. The infection was presumably transmitted by hunting dogs.

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