

# SURVEILLANCE OF INFECTIOUS DISEASES IN ANIMALS AND HUMANS IN SWEDEN 2020

Chapter excerpt -  
Echinococcosis



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**Cover:** Juvenile mink in hand. Photo: Elina Kähkönen

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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, 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 and the LaTeX library pgfplots. Development for 2020 has further improved the importing of content from Excel files to automatically build figures in the pgfplots LaTeX library. The tool is available as an R-package on GitHub (<https://github.com/SVA-SE/mill/>). The report generation R-package and process was designed by Thomas Rosendal, Wiktor Gustafsson and Stefan Widgren. In 2020, final typesetting was done primarily by Wiktor Gustafsson with contributions from the report authors.

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# Echinococcosis

## 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.

### History

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 to the finding of *E. multilocularis* in foxes in Denmark, an active monitoring programme of red foxes (*Vulpes vulpes*) was implemented in Sweden in 2000. From 2000 to 2009, a total of 2962 red foxes, 68 raccoon dogs (*Nyctereutes procyonoides*) and 35 wolves (*Canis lupus*) were examined for *E. multilocularis*, all with negative results. Samples from the majority of foxes ( $n=2675$ ) were examined by ELISA (CoproAntigen ELISA) at the Institute for Parasitology, Zurich University, for the presence of the *E. multilocularis* coproantigen. The remaining samples and those that were ELISA positive, were examined using the sedimentation and counting technique (SCT) ( $n=726$ ). All samples from raccoon dogs and wolves were examined by SCT.



During 2020, fox scats were collected from three areas in Sweden where *Echinococcus multilocularis* previously has been found. The results show that the parasite is still present in two of the areas. Photo: SVA.

During 2010, 304 foxes were examined for *E. multilocularis*. A total of 103 were tested by SCT and 201 by egg PCR. One fox shot in south-west Sweden (county of Västra Götaland) and analysed in 2011 was found to be positive.

During the spring of 2011, a national surveillance programme was implemented where 2985 hunter-shot foxes were analysed with the segmental sedimentation and counting technique (SSCT). Three foxes were found positive: one in Västra Götaland, one in Södermanland and one in the county of Dalarna. In addition, 119 faecal samples from hunting dogs collected in the region of the first positive finding were analysed with egg PCR and all were negative. In the same area 236 rodents were necropsied and all potential lesions examined by an in-house PCR without any positive finding.

To obtain a better prevalence estimate in a known infected area, fox scats were collected, by a systematic sampling procedure, from a circular area with a diameter of 25 km surrounding a positive finding in the county of Södermanland. The samples were collected in 2011 and analysed in 2012, using semi-automated magnetic capture probe-based DNA extraction and real-time PCR method (MC-PCR). Six out of 790 (0.8%) faecal samples were positive.

A second national screening was initiated in 2012 and continued in 2013 and 2014. In all, a total of 2779 fox scat samples were analysed, and three positive fox scats were identified, one from Gnesta, one from Katrineholm (both in the county of Södermanland) and one from the county of Västra Götaland.

From the five known infected areas (including one area in the county of Kronoberg which was identified as infected in 2014, see below), hunters were asked to submit 30 foxes from each circular area with a diameter of 40 km. The aim was to follow up the positive findings, and to collect parasites from any positive cases, for further subtyping. Sampling was initiated in 2012 and finalized in 2016. In Västra Götaland two foxes were positive, in Södermanland three foxes from Katrineholm and one from Gnesta were positive, whereas no fox from Dalarna or Kronoberg was positive. In 2018, on one single occasion, fox scats were collected in Gnesta and 6 of 13 samples tested positive. This showed that the parasite was still present in this location.

Within the Emiro research project (finalized in 2016) and the FoMA Zoonosis monitoring programme ([www.slu.se/en/environment](http://www.slu.se/en/environment)) at the Swedish University of Agricultural Sciences (SLU), the parasite was found for the first time in intermediate hosts; 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 metacestode 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.;  $n=285$ ). Within this project, a new infected area was identified in 2014 near the town Växjö in the county of Kronoberg.

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 one 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 2013:23). Before 2012, all imported dogs and cats (except from certain countries) were required to be de-wormed with praziquantel before entering Sweden as a preventive measure. Because *E. multilocularis* has been detected in Sweden, there is presently no legal requirement to deworm pets entering the country. However, as the prevalence of the parasite in foxes is very low in Sweden compared to many European countries, dog owners are still encouraged to deworm their dogs prior to entry to Sweden.

### Humans

Infection with *Echinococcus* spp. has been notifiable since 2004 according to the Communicable Disease Act (SFS 2004:168) with the amendments of SFS 2013:634. 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 ([www.folkhalsomyndigheten.se](http://www.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. In 2020, all free-living wolves submitted to necropsy at the National Veterinary Institute were tested with MC-PCR. In addition, fox scats were collected from the areas in Uddevalla, Gnesta, and Katrineholm/Finspång where the parasite has been previously found.

## 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

During 2020, 29 wolves (*Canis lupus lupus*), four red foxes, three dogs and one cat were tested with the MC-PCR and all were negative. However, analysis of fox scats collected from areas where the parasite has previously been found 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, Sörmland were positive), while none of 108 samples from the area in Katrineholm, Sörmland/Finspång, Östergötland tested positive.

### Humans

In 2020, there were three cases of alveolar echinococcosis reported. One person had probably acquired the infection in his country of origin in Eastern Europe, while it cannot be ruled out for the remaining two cases that they had been infected in Sweden, but they could also have acquired the infection while travelling abroad.

## 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 the future trend can be assessed. It is well known from other countries that the prevalence of this parasite varies geographically. Regional screenings have previously shown a prevalence of more than 1% in a part of the county of Södermanland, and within the Emiro research project and FoMA Zoonosis monitoring programme 18 of 80 (20%) fox scats were found to be positive in one of four investigated small areas. However, the true geographical distribution is unknown but so far no positive cases have been found north of Dalarna county. Until now, the infection has been detected in five different areas. The recent finding of positive fox scats in two of these areas shows that the parasite is still present in these locations. In 2021 a new national screening to assess the present prevalence in foxes will be initiated, which is expected to run for three years.

*E. multilocularis* has also been found in intermediate hosts, for the first time in 2014 within the Emiro research project. 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 *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 get the infection when consuming organs containing hydatid cysts.

### History

Echinococcosis was quite common in reindeer in the northern parts of Scandinavia in the first half of the 20<sup>th</sup> 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. However, in a retrospective study of biobank material from 116 wolves submitted to the National Veterinary Institute during 2012–2020, two wolves shot 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 2013:23).

#### Humans

Echinococcosis has been notifiable according to the Communicable Disease Act since 2004 (SFS 2004:168) with the amendments of SFS 2013:634. 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 ([www.folkhalsomyndigheten.se](http://www.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 National Veterinary Institute for diagnosis.

### 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

*E. granulosus* s.l. was not detected in any animal sampled in 2020. In addition to the routine inspection at slaughter, 5 wolves and 2 dogs were tested by MC-PCR detecting *E. canadensis* genotype G8 and G10 as well as *E. ortleppi*.

### Humans

In 2020, 20 cases of cystic echinococcosis were reported. For eleven of these cases, it was not possible with available laboratory methods to determine which species they were infected with, but the epidemiology suggested that they also had cystic echinococcosis. Annually around 15–30 cases are reported in Sweden. In 2020, the reported cases ranged in age from 8 to 68 years (median 34 years). Eight cases were women and 12 were men. They were all considered to have been infected abroad in areas where the parasite is endemic. The most frequently specified countries of infection were Syria (6 cases) and Iraq (4 cases).

### Discussion

*E. granulosus* s.l. has not been detected in animal intermediate hosts in Sweden 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 shot 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*.

This species is considered as less pathogenic, and possibly with a lower zoonotic potential, than *E. granulosus* sensu stricto that is prevalent in other parts of Europe and identified mainly in a cycle between dogs and farm animals.

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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