SURVEILLANCE REPORT

Echinococcosis

Annual Epidemiological Report for 2019

Key facts

- In 2019, 775 confirmed echinococcosis cases were reported in the European Union/European Economic Area (EU/EEA). Of these, 419 cases (54%) were reported as *Echinococcus granulosus*, 154 (20%) as *Echinococcus multilocularis*, and 205 (26%) as unknown species.
- The EU/EEA notification rate of echinococcosis was 0.17 cases per 100 000 population, which was the lowest notification rate in the last 5 years.
- The highest notification rate in males was reported among those aged 25–44 years and ≥65 years, and in females among those aged 45-64 years.

Methods

This report is based on data for 2019 retrieved from The European Surveillance System (TESSy) on 4 November 2021. TESSy is a system for the collection, analysis and dissemination of data on communicable diseases.

For a detailed description of methods used to produce this report, please refer to the *Methods* chapter [1]. An overview of the national surveillance systems is available online [2].

A subset of the data used for this report is available through ECDC's online *Surveillance atlas of infectious diseases* [3].

Twenty-five countries reported echinococcosis cases using the 2008, 2012 or 2018 EU case definitions, which are identical. Luxembourg reported in accordance with the 2002 EU case definition, and two countries (France and Germany) used other/unspecified definitions. Echinococcosis is under mandatory surveillance in 24 EU/EEA countries. Surveillance is voluntary in four EU countries (Belgium, France, the Netherlands, and the United Kingdom (then a member)) [2]. Denmark and Italy have no surveillance system for echinococcosis. Most reporting countries provided case-based data except for Bulgaria and the Netherlands, which reported aggregate data. Twenty countries had surveillance systems that integrate laboratory and epidemiological data from physicians or hospitals.

Epidemiology

For 2019, 28 EU/EEA countries reported data on echinococcosis; 24 countries reported 776 cases, of which 775 were confirmed cases, and four countries (Cyprus, Ireland, Malta, and Iceland) reported zero cases (Table 1). The EU/EEA notification rate in 2019 was 0.17 confirmed echinococcosis cases per 100 000 population, which was the

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lowest notification rate in the previous 5 years. The highest notification rates were observed in Bulgaria (2.76 per 100 000) and Lithuania (2.90 per 100 000 population) (Table 1, Figure 1). The increased notification rate reported by Lithuania is attributed to improved diagnosis of echinococcosis [4]. The highest number of cases was reported by Bulgaria, accounting for 25% of all reported cases, and Germany, accounting for 19% of all reported cases.

Fifteen countries provided information on hospitalisation, covering 33% (n=253) of all confirmed cases of echinococcosis in the EU/EEA in 2019. Among these, 44% were hospitalised, representing a continuous decrease during the last 10 years from 100% in 2008, when only hospitalised cases were reported. The highest proportion of cases requiring hospitalisation were reported in Czechia (100%), Greece (100%), Poland (83%), Portugal (60%), Romania (100%), Slovakia (60%) and Slovenia (60%). Among cases with known information, more than half (n=35; 56%) of human *Echinococcus* (*E.*) *multilocularis* cases were hospitalised compared to approximately one-third (n=30; 36%) of human *E. granulosus sensu lato (s.l.)* cases based on reporting by four and ten countries, respectively.

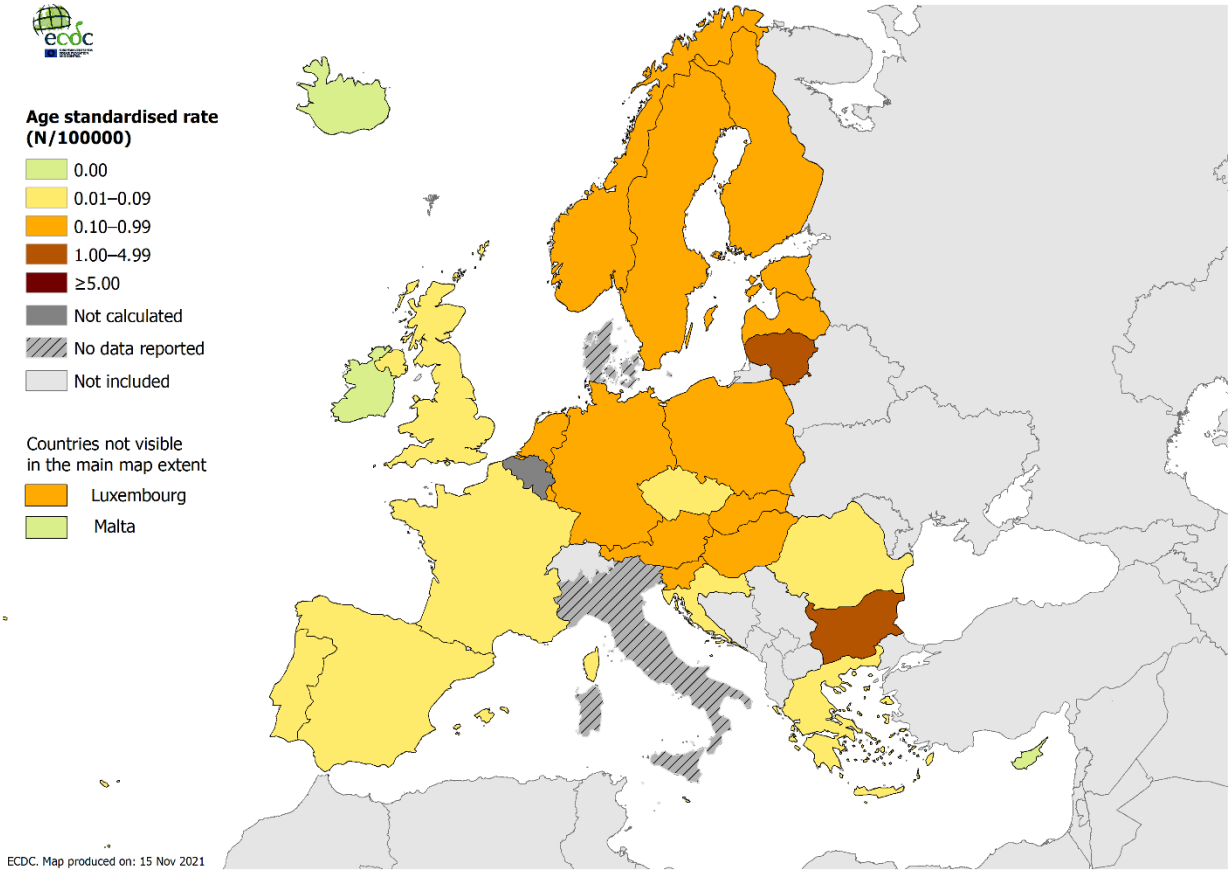
Information on the outcome of 248 cases was provided by 15 countries. One fatal case each due to infection with *E. granulosus s.l.* and infection with *E. multilocularis* was reported in Portugal and Poland, respectively. The fatal cases, one male and one female, were aged 44 and 77 years.

Table 1. Number of confirmed echinococcosis cases and rates per 100 000 population by country and year, EU/EEA, 2015–2019

Country	2015		2016		2017		2018		2019			
	Number	Rate	Number	Rate	Number	Rate	Number	Rate	Confirmed cases	Rate	ASR	Reported cases
Austria	8	0.1	26	0.3	50	0.6	46	0.5	36	0.41	0.40	36
Belgium	10	0.1	17	0.2	13	0.1	15	0.1	22	0.19	-	22
Bulgaria	313	4.3	269	3.8	218	3.1	206	2.9	193	2.76	2.78	193
Croatia	7	0.2	9	0.2	15	0.4	4	0.1	3	0.07	0.07	4
Cyprus	2	0.2	0	0.0	0	0.0	0	0.0	0	0.00	0.00	0
Czechia	3	0.0	4	0.0	1	0.0	4	0.0	1	0.01	0.01	1
Denmark	ND	NR	ND	NR	ND	NR	ND	NR	ND	NR	NR	ND
Estonia	0	0.0	0	0.0	1	0.1	0	0.0	2	0.15	0.13	2
Finland	2	0.0	4	0.1	5	0.1	1	0.0	8	0.14	0.16	8
France	48	0.1	38	0.1	53	0.1	62	0.1	45	0.07	0.07	45
Germany	157	0.2	181	0.2	141	0.2	176	0.2	149	0.18	0.18	149
Greece	13	0.1	18	0.2	15	0.1	11	0.1	7	0.07	0.06	7
Hungary	2	0.0	5	0.1	14	0.1	9	0.1	10	0.10	0.10	10
Iceland	0	0.0	0	0.0	0	0.0	0	0.0	0	0.00	0.00	0
Ireland	0	0.0	2	0.0	0	0.0	2	0.0	0	0.00	0.00	0
Italy	ND	NR	ND	NR	ND	NR	ND	NR	ND	NR	NR	ND
Latvia	10	0.5	11	0.6	6	0.3	10	0.5	6	0.31	0.29	6
Liechtenstein	ND	NR	ND	NR	ND	NR	ND	NR	ND	NR	NR	ND
Lithuania	33	1.1	26	0.9	53	1.9	50	1.8	81	2.90	2.63	81
Luxembourg	0	0.0	0	0.0	2	0.3	0	0.0	1	0.16	0.16	1
Malta	0	0.0	1	0.2	0	0.0	0	0.0	0	0.00	0.00	0
Netherlands	64	0.4	33	0.2	38	0.2	42	0.2	48	0.28	0.28	48
Norway	2	0.0	3	0.1	6	0.1	7	0.1	7	0.13	0.14	7
Poland	47	0.1	64	0.2	75	0.2	51	0.1	70	0.18	0.18	70
Portugal	4	0.0	2	0.0	2	0.0	9	0.1	5	0.05	0.04	5
Romania	18	0.1	13	0.1	14	0.1	4	0.0	1	0.01	0.01	1
Slovakia	5	0.1	4	0.1	7	0.1	10	0.2	11	0.20	0.20	11
Slovenia	7	0.3	3	0.1	7	0.3	6	0.3	6	0.29	0.28	6
Spain	83	0.2	87	0.2	83	0.2	68	0.1	34	0.07	0.07	34
Sweden	26	0.3	27	0.3	34	0.3	29	0.3	26	0.25	0.26	26
United Kingdom	26	0.0	ND	NR	4	0.0	ND	NR	3	0.00	0.00	3
EU/EEA	**890**	**0.2**	**847**	**0.2**	**857**	**0.2**	**822**	**0.2**	**775**	**0.17**	**0.17**	**776**

ND: no data reported, NR: no rate calculated, ASR: age-standardised rate
Denmark and Italy have no surveillance system for echinococcosis. Data were not reported by Liechtenstein (for all years) or the UK (in 2016 and 2018); the reasons for this are unclear.

Figure 1. Distribution of confirmed echinococcosis cases per 100 000 population by country, EU/EEA, 2019

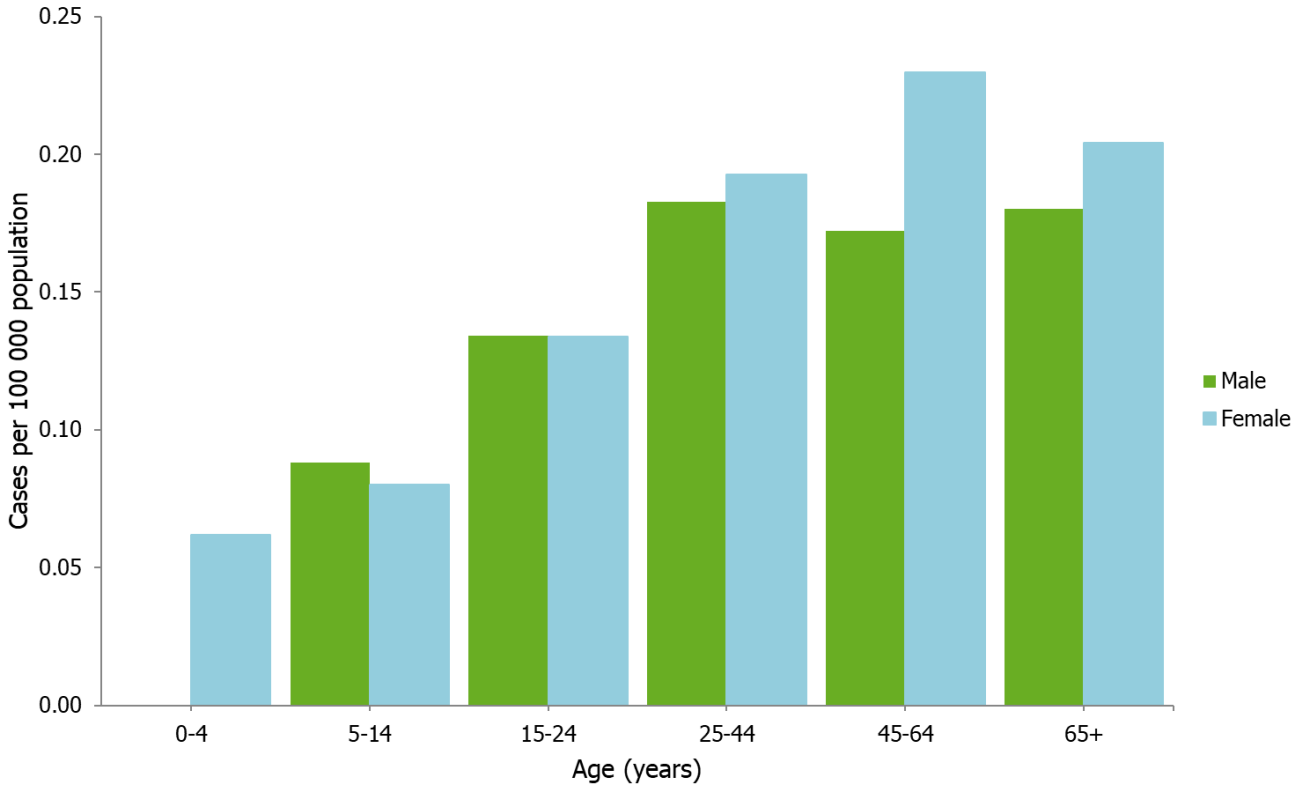


Sources: Country reports from Austria, Belgium, Bulgaria, Croatia, Cyprus, Czechia, Estonia, Finland, France, Germany, Greece, Hungary, Iceland, Ireland, Latvia, Lithuania, Luxembourg, Malta, the Netherlands, Norway, Poland, Portugal, Romania, Slovakia, Slovenia, Spain, Sweden, and the United Kingdom.

The male-to-female ratio was 0.8:1. The highest notification rate in males was reported among people aged 25–44 years and ≥65 years, and in females among people aged 45-64 years (Figure 2).

Most echinococcosis cases (64%) were reported without importation status. Of cases with known status (n = 281), 47% were reported to have been infected outside of the reporting country in 2019. Seven of the 16 countries (Czechia, Estonia, Hungary, Latvia, Lithuania, Romania, and Slovakia) reporting information on importation status in 2019 notified all *Echinococcus* spp. infections as being domestically acquired. The highest proportions of imported cases were reported by Finland (100%), Luxembourg (100%), Norway (100%) and Sweden (81%).

Figure 2. Distribution of confirmed echinococcosis cases per 100 000 population, by age and sex, EU/EEA, 2019



Echinococcosis by species

Species information was known for 573 of 703 (82%) confirmed cases from 17 countries (seven countries did not report on species or form of disease).

Echinococcus granulosus

Seventeen countries reported 419 confirmed cases of *E. granulosus* sensu lato (s.l.) (cystic echinococcosis) in 2019 (Table 2). Bulgaria accounted for 46% of cases in 2019 and Germany for 21%. The largest proportion (33%) of cystic echinococcosis cases was observed in the age group 25–44 years, followed by the age group 45–64 years (28%). Of the cystic echinococcosis cases with known sex (n=406), there were slightly more cases reported in females (53%) than in males (47%). In the five-year period 2015–2019, the trend of *E. granulosus* did not show any significant increase or decrease in the EU/EEA [5]. Bulgaria, which reported the majority of cystic echinococcosis cases, was not included in the trend calculations since no monthly data were available. However, cases in Bulgaria decreased by 38.3% from 313 cases in 2015 to 193 cases in 2019. There was strong evidence ($p \leq 0.05$) of an increasing trend in case numbers in Lithuania and Finland [5]. Among cases with known importation status (n=143), 68% were reported to have been infected outside of the reporting country, which is similar to that reported in 2018 (71%) (aggregate data from Bulgaria not included).

Echinococcus multilocularis

Eight countries reported 154 cases of *E. multilocularis* (alveolar echinococcosis) in 2019 (Table 2). Germany and France accounted for 49% of the reported *E. multilocularis* cases in the EU/EEA in 2019. Most cases were reported in the age groups 25-44 years (33%) and 45-64 years (28%). Of the alveolar echinococcosis cases with known sex (n = 153), there was slightly more cases reported in females (56%) than in males (44%). In the five-year period 2015–2019, the number of *E. multilocularis* infections was stable in the EU/EEA overall [5]. For 10 countries with available data for 2015 to 2019, Austria was the only country with an increasing trend, and none had a decreasing trend [5]. Among cases with known importation status (n=47), 85% of cases with *E. multilocularis* were reported to have been infected within the reporting country in 2019.

Table 2. Reported confirmed echinococcosis cases, by species, EU/EEA, 2015–2019

Country	Total cases					*E. granulosus*					*E. multilocularis*				
	2015	2016	2017	2018	2019	2015	2016	2017	2018	2019	2015	2016	2017	2018	2019
Austria	8	26	50	46	36	2	22	37	29	16	3	4	8	12	13
Belgium	10	17	13	15	22	5	UNK	9	10	12	5	UNK	4	5	10
Bulgaria	313	269	218	206	193	313	269	218	206	193	0	0	0	0	0
Croatia	7	9	15	4	3	UNK	UNK	UNK	UNK	UNK	UNK	UNK	UNK	UNK	UNK
Cyprus	2	0	0	0	0	UNK	0	0	0	0	UNK	0	0	0	0
Czechia	3	4	1	4	1	UNK	UNK	UNK	1	UNK	UNK	UNK	UNK	2	UNK
Estonia	0	0	1	0	2	0	0	0	0	UNK	0	0	1	0	UNK
Finland	2	4	5	1	8	UNK	4	5	1	8	UNK	0	0	0	0
France	48	38	53	62	45	0	0	5	21	10	48	38	48	41	35
Germany	157	181	141	176	149	85	122	86	93	87	48	40	35	59	40
Greece	13	18	15	11	7	UNK	UNK	UNK	UNK	UNK	UNK	UNK	UNK	UNK	UNK
Hungary	2	5	14	9	10	UNK	UNK	1	UNK	UNK	UNK	UNK	1	UNK	UNK
Iceland	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
Ireland	0	2	0	2	0	0	1	0	UNK	0	0	UNK	0	UNK	0
Latvia	10	11	6	10	6	2	1	4	5	4	UNK	1	UNK	1	UNK
Lithuania	33	26	53	50	81	9	5	19	11	30	11	10	20	17	21
Luxembourg	0	0	2	0	1	0	0	2	0	1	0	0	0	0	0
Malta	0	1	0	0	0	0	1	0	0	0	0	0	0	0	0
Netherlands	64	33	38	42	48	UNK	UNK	UNK	UNK	UNK	UNK	UNK	UNK	UNK	UNK
Norway	2	3	6	7	7	1	1	3	5	2	UNK	UNK	1	UNK	UNK
Poland	47	64	75	51	70	9	18	27	17	21	26	22	31	19	25
Portugal	4	2	2	9	5	1	2	UNK	9	5	UNK	0	UNK	0	0
Romania	18	13	14	4	1	2	UNK	UNK	UNK	UNK	UNK	UNK	UNK	UNK	UNK
Slovakia	5	4	7	10	11	2	1	2	3	3	3	2	3	3	8
Slovenia	7	3	7	6	6	UNK	UNK	UNK	3	1	UNK	UNK	UNK	UNK	UNK
Spain	83	87	83	68	34	4	1	4	12	6	UNK	UNK	UNK	UNK	UNK
Sweden	26	27	34	29	26	19	20	11	5	17	UNK	1	4	2	2
United Kingdom	26	ND	4	ND	3	26	ND	4	ND	3	0	ND	0	ND	0
EU/EEA	890	847	857	822	775	480	468	437	431	419	144	118	156	161	154

UNK: Species unknown
ND: No data reported

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Discussion

Cases of both alveolar echinococcosis caused by *E. multilocularis,* and cystic echinococcosis caused by *E. granulosus sensu lato (s.l.),* are listed with the common name 'echinococcosis' in the EU case definition, thus not distinguishing between these two diseases. Despite this, most countries reported *Echinococcus* species information between 2008 and 2019. Additionally, between 2018 and 2019, a small number of countries also the reported clinical presentation, which helps differentiate the two forms of the disease. Since the beginning of the surveillance of human echinococcosis in the EU in 2007, cystic echinococcosis has been more frequently reported than alveolar echinococcosis; this is consistent with the data reported in the scientific literature for Europe.

In the five years between 2015 and 2019, the EU/EEA notification rate of confirmed human echinococcosis cases was stable and the trends for infections caused by *E. granulosus s.l.* and *E. multilocularis* did not show any significant increase or decrease. In some countries, an increase in the number of cases in the recent years may be due to intensified surveillance and/or improved notification systems for echinococcosis. The raised awareness of the disease among clinicians and migrants (people from endemic countries) may have also influenced the number of diagnosed cases in some countries [7]. In Bulgaria, the notification rate has continued to decrease since reporting to ECDC began in 2007. This reduction is attributed to measures implemented as part of a national control programme for echinococcosis in humans and animals carried out between 2004 and 2008 and to the considerable reduction in the number of stray dogs [6].

The distribution of the two forms of disease varies in different parts of the EU/EEA. *E. multilocularis* is endemic in the fox population in central Europe [5,8] and human cases of alveolar echinococcosis are principally reported by these countries, with most cases considered to be infected within the reporting country. In contrast, *E. granulosus s.l.*, is mainly prevalent in intermediate hosts (e.g. sheep) in southern and south-eastern Europe and is an important public health issue in many countries in the Balkan region [5,9], while in northern and western Europe most cases are considered imported. In accordance with Regulation (EU) 2018/772, surveillance of *E. multilocularis* focuses mainly on red foxes as definitive hosts; in 2019, *E. multilocularis* infections were primarily detected in foxes in Czechia, France, Germany, Hungary, Luxembourg, Poland, Slovakia and Switzerland [5]. The surveillance of *E. multilocularis* in foxes is considered important to assess the prevalence of the disease in Europe. Indeed, the geographical distribution of *E. multilocularis* appears to have widened in recent decades [10]. The exact reasons for this are unclear but may be linked to the growth in the European fox population [11], due to the expansion of fox habitats into urban areas [12] or may reflect an increased surveillance effort [5]. However, the lack of baseline data and standardised detection methods makes it difficult to explain the geographical expansion of *E. multilocularis* in Europe [5]. Although Regulation (EU) 2018/772 is in force for *E. multilocularis*, no specific EU Regulation is in place for detecting *E. granulosus s.l.* in animals, so surveillance for the latter parasite depends on national regulations. In 2019, *E. granulosus s.l.* infections in animals were mainly detected in sheep in Bulgaria, Spain, Greece, Poland, Italy and UK [5].

Distinction between infection with *E. granulosus s.l.* and *E. multilocularis* is important because the two diseases require different clinical management and strategies for control. It is also important to note that the true prevalence of these diseases is difficult to estimate because of the long incubation period (in both alveolar echinococcosis and cystic echinococcosis), the high proportion of asymptomatic or paucisymptomatic carriers who never seek medical attention (cystic echinococcosis) and the underreporting/misdiagnosis of cases (alveolar echinococcosis and cystic echinococcosis), factors that contribute to the neglected status of these diseases [13]. For these reasons, the data reported by countries on the number of people with echinococcosis, are considered the 'tip of the iceberg' of the true prevalence of echinococcosis in Europe. The hidden (undetected and unreported) proportion of echinococcosis cases includes asymptomatic carriers of cystic echinococcosis and misdiagnosed cases of alveolar echinococcosis [14]. Indeed, a recent cross-sectional ultrasound-based survey, conducted in Romania and Bulgaria, estimated approximately 45 000 human cystic echinococcosis infections in rural areas of these two endemic European countries [25].

An attempt to collect harmonised clinical data in the EU on a voluntary basis is currently being undertaken by the European Register of Cystic Echinococcosis (ERCE) [16, 17; http://www.heracles-fp7.eu/erce.html) and in the past with the European (Alveolar) Echinococcosis Registry (EurEchinoReg) [14].

Public health implications

It is recommended that reporting of echinococcosis cases should include species information and preferably data collected at the NUTS-2 or NUTS-3 level. This would allow for a more complete monitoring of cases, foster a better understanding of the epidemiology of these diseases, improve monitoring of spatial and temporal trends, and ultimately enable the design and evaluation of targeted prevention and control actions.

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