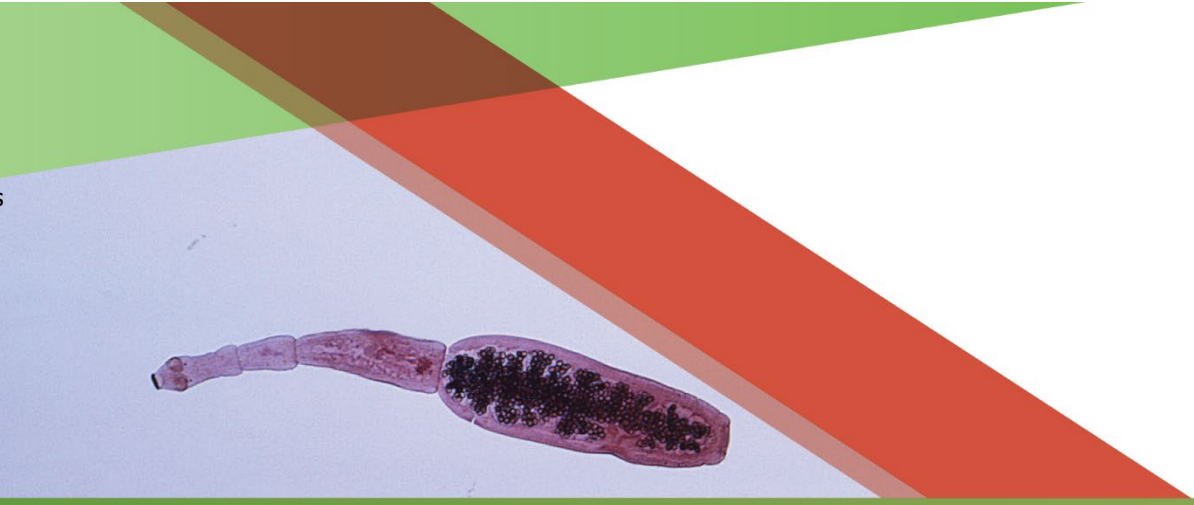


SURVEILLANCE REPORT

Echinococcosis

Annual Epidemiological Report for 2020

Key facts

- In 2020, 529 confirmed echinococcosis cases were reported in the European Union/European Economic Area (EU/EEA). Of these, 243 cases were reported as *Echinococcus granulosus*, 114 as *Echinococcus multilocularis*, and 172 were reported with unknown species.
- The EU/EEA notification rate for 2020 was 0.15 per 100 000 population, which represents a further decrease compared to the rate of 0.17 for 2019, and 0.21 for 2018. The 2020 notification rate is the lowest since EU surveillance of echinococcosis began in 2007.
- The highest notification rate in males was reported in those aged ≥65 years, and in females among those 25-44 years.

Methods

This report is based on data for 2020 retrieved from The European Surveillance System (TESSy) on 19 January 2022. 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-four countries reported echinococcosis cases using the 2008, 2012 or 2018 EU case definitions, which are identical. One country (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, and under voluntary surveillance in Belgium, France, and the Netherlands [2]. Denmark and Italy have no surveillance system for echinococcosis. For 2020, Spain did not receive data from all its regions due to the COVID-19 pandemic, so the case numbers might not be complete. Most reporting countries provided case-based data except for Bulgaria and the Netherlands, which reported aggregate data. Nineteen countries had surveillance systems that integrate laboratory and epidemiological data from physicians or hospitals. No data for 2020 were reported by the United Kingdom (UK) due to its withdrawal from the EU on 1 February 2020.

Suggested citation: European Centre for Disease Prevention and Control. Echinococcosis. In: ECDC. Annual epidemiological report for 2020. Stockholm: ECDC; 2022.

Stockholm, May 2022

Epidemiology

For 2020, 27 EU/EEA countries reported data on echinococcosis; 23 countries reported 624 cases, of which 529 (85%) were confirmed cases. Four countries (Ireland, Malta, Romania, and Iceland) reported zero cases (Table 1). The EU/EEA notification rate for 2020 was 0.15 per 100 000 population, which represents a further decrease compared to the rate of 0.17 for 2019, and 0.21 for 2018. The 2020 notification rate is the lowest since EU surveillance of *Echinococcus* spp. began in 2007. In 2020, a considerable reduction in the reported number of confirmed cases was observed in Bulgaria, Spain, and Poland in comparison to 2019. For Bulgaria, the overall number of reported cases was consistent with previous years, but approximately half were reported with probable case classification in 2020. The reasons for this are unclear. For Spain, reporting from some regions was incomplete due to a lack of resources resulting from the COVID-19 pandemic [4]. The reasons for the reduction in the reported number of confirmed cases observed in Poland is unknown.

In 2020, the highest notification rates were observed in Bulgaria (1.37 per 100 000 population) and Lithuania (1.32 per 100 000 population) (Table 1, Figure 1). The increased notification rate reported by Lithuania in recent years is attributed to improved diagnosis of echinococcosis [5]. Germany and Bulgaria reported the highest numbers of confirmed cases, with 152 (29%) and 95 (18%) cases out of 529 cases, respectively.

Fourteen countries provided information on hospitalisation, covering 16% (n=84) of all confirmed cases of echinococcosis in the EU/EEA in 2020. Among these, almost two thirds (57%) were hospitalised. Four countries (Cyprus, Estonia, Greece, and Portugal) reported that all cases were hospitalised, while 78% of cases in Poland were hospitalised. Among cases with known information, more than three quarters (n=11; 79%) of human *E. multilocularis* cases were hospitalised compared to approximately two thirds (n=25; 58%) of human *E. granulosus s.l.* cases based on reporting by five and 13 countries, respectively.

Information on the outcome of 206 cases was provided by 15 countries; no deaths were reported.

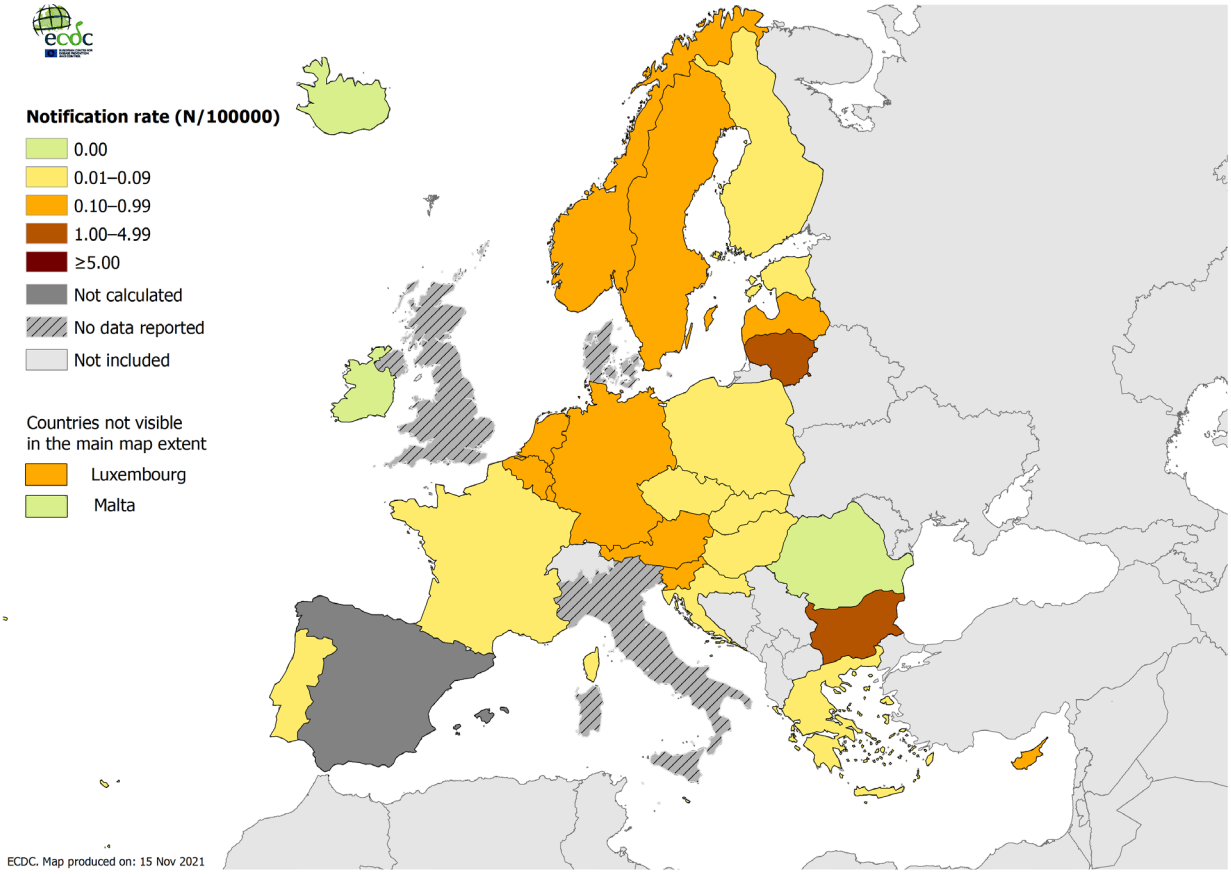
Table 1. Number of confirmed echinococcosis cases and rates per 100 000 population by country and year, EU/EEA, 2016–2020

Country	2016		2017		2018		2019		2020			
	Number	Rate	Number	Rate	Number	Rate	Number	Rate	Confirmed cases	Rate	ASR	Reported cases
Austria	26	0.30	50	0.57	46	0.52	36	0.41	34	0.38	0.37	34
Belgium	17	0.15	13	0.11	15	0.13	22	0.19	19	0.16	-	19
Bulgaria	269	3.76	218	3.07	206	2.92	193	2.76	95	1.37	1.46	190
Croatia	9	0.21	15	0.36	4	0.10	3	0.07	3	0.07	0.08	3
Cyprus	0	0.00	0	0.00	0	0.00	0	0.00	1	0.11	0.12	1
Czechia	4	0.04	1	0.01	4	0.04	1	0.01	4	0.04	0.03	4
Denmark	ND	NR	ND	NR	ND	NR	ND	NR	ND	NR	NR	ND
Estonia	0	0.00	1	0.08	0	0.00	2	0.15	1	0.08	0.10	1
Finland	4	0.07	5	0.09	1	0.02	8	0.14	4	0.07	0.07	4
France	38	0.06	53	0.08	62	0.09	45	0.07	53	0.08	0.08	53
Germany	181	0.22	141	0.17	176	0.21	149	0.18	152	0.18	0.18	152
Greece	18	0.17	15	0.14	11	0.10	7	0.07	7	0.07	0.07	7
Hungary	5	0.05	14	0.14	9	0.09	10	0.10	4	0.04	0.04	4
Iceland	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00	0.00	0
Ireland	2	0.04	0	0.00	2	0.04	0	0.00	0	0.00	0.00	0

Country	2016		2017		2018		2019		2020			
	Number	Rate	Number	Rate	Number	Rate	Number	Rate	Confirmed cases	Rate	ASR	Reported cases
Italy	ND	NR	ND	NR	ND	NR	ND	NR	ND	NR	NR	ND
Latvia	11	0.56	6	0.31	10	0.52	6	0.31	2	0.10	0.08	2
Liechtenstein	ND	NR	ND	NR	ND	NR	ND	NR	ND	NR	NR	ND
Lithuania	26	0.90	53	1.86	50	1.78	81	2.90	37	1.32	-	37
Luxembourg	0	0.00	2	0.34	0	0.00	1	0.16	3	0.48	0.48	3
Malta	1	0.22	0	0.00	0	0.00	0	0.00	0	0.00	0.00	0
Netherlands	33	0.19	38	0.22	42	0.24	48	0.28	48	0.28	0.27	48
Norway	3	0.06	6	0.11	7	0.13	7	0.13	6	0.11	0.11	6
Poland	64	0.17	75	0.20	51	0.13	70	0.18	18	0.05	0.05	18
Portugal	2	0.02	2	0.02	9	0.09	5	0.05	1	0.01	0.01	1
Romania	13	0.07	14	0.07	4	0.02	1	0.01	0	0.00	0.00	0
Slovakia	4	0.07	7	0.13	10	0.18	11	0.20	3	0.05	0.05	3
Slovenia	3	0.15	7	0.34	6	0.29	6	0.29	3	0.14	0.15	3
Spain	87	0.19	83	0.18	68	0.15	34	0.07	8	-	-	8
Sweden	27	0.27	34	0.34	29	0.29	26	0.25	23	0.22	0.23	23
United Kingdom	ND	NR	4	0.01	ND	NR	3	0.00	ND	NR	NR	ND
EU-EEA	**847**	**0.22**	**857**	**0.19**	**822**	**0.21**	**775**	**0.17**	**529**	**0.15**	**0.14**	**624**

ND: no data reported, NR: no rate calculated, ASR: age-standardised rate
For 2020, Spain did not receive data from all its regions due to the COVID-19 pandemic. Denmark and Italy have no surveillance system for echinococcosis. Data were not collected from the UK in 2020, as the country left the EU on 31 January 2020. 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, 2020

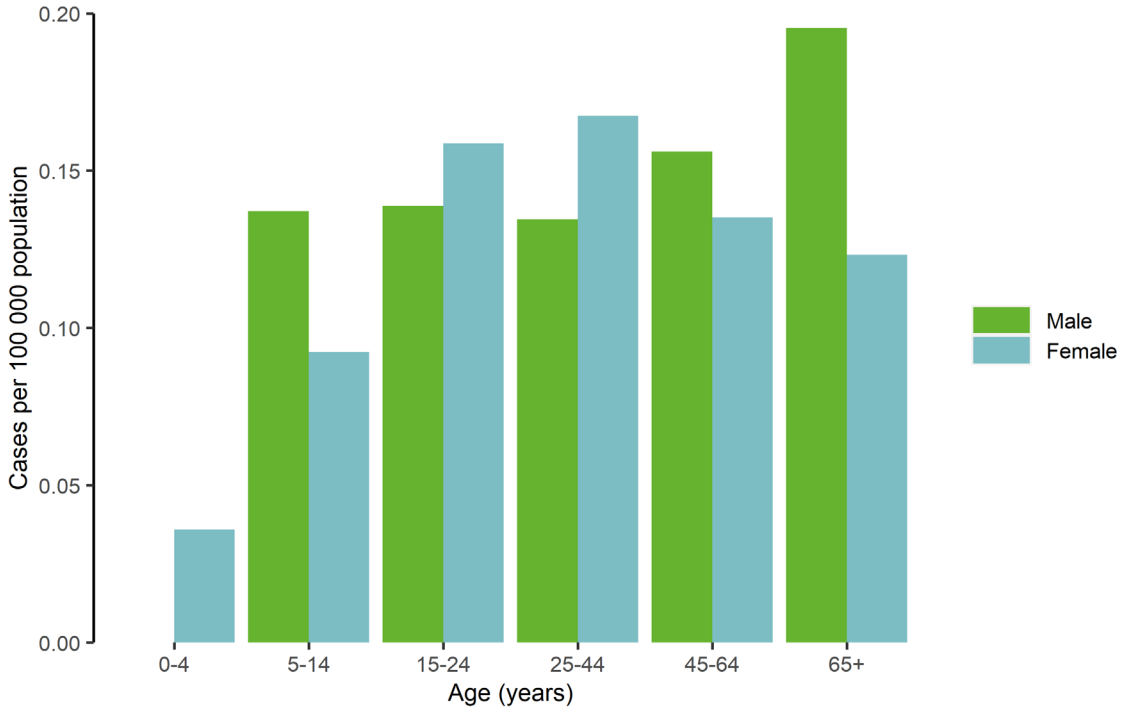


Sources: Country reports from Austria, Belgium, Bulgaria, Croatia, Czechia, Estonia, Finland, France, Germany, Greece, Hungary, Iceland, Ireland, Latvia, Lithuania, Luxembourg, Malta, the Netherlands, Norway, Poland, Portugal, Romania, Slovakia, Slovenia, and Sweden.

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

Most echinococcosis cases (74%) were reported without importation status. Of cases with known status (n=138), 65% were reported to have been infected outside of the reporting country in 2020. Four of the 13 countries (Czechia, Estonia, Hungary, and Portugal) reporting information on importation status in 2020 notified all *Echinococcus* spp. infections as being domestically acquired. The highest proportions of imported cases were reported by Sweden (74%) and Norway (50%).

Figure 2. Distribution of confirmed echinococcosis cases per 100 000 population, by age and sex, EU/EEA, 2020



Echinococcosis by species

Species information was known for 357 of 440 (81%) confirmed cases from 19 countries in 2020 (four countries did not report on species or form of disease).

Echinococcus granulosus

Nineteen countries reported 243 confirmed cases of *E. granulosus* sensu lato (s.l.) (cystic echinococcosis) in 2020 (Table 2). The number of reported cystic echinococcosis cases in 2020 was considerably lower than the average annual number of 439 cases reported between 2016 and 2019. Bulgaria accounted for 39% of all cystic echinococcosis cases in 2020 and Germany for 29%. The majority (34%) of cystic echinococcosis cases was observed in the age group 25–44 years, followed by the age group 45–64 years (21%). Of the cystic echinococcosis cases with known sex (n=232), 51% were female. Although Bulgaria accounted for the majority of cystic echinococcosis cases in 2020, cases in Bulgaria decreased by 65%, from 269 cases in 2016 to 95 cases in 2020. Among cases with known importation status (n=74), 81% of cystic echinococcosis cases were reported to have been infected outside of the reporting country in 2020, compared to 68% in 2019 (aggregate data from Bulgaria not included).

Echinococcus multilocularis

Nine countries reported 114 cases of *E. multilocularis* (alveolar echinococcosis) in 2020 (Table 2). Human infections caused by *E. multilocularis* in 2020 were fewer than the numbers reported between 2017 and 2019 and slightly less than in 2016. Germany and France accounted for 77% of the reported *E. multilocularis* cases in the EU/EEA in 2020. Most cases of alveolar echinococcosis were reported in the age groups 45-64 years (39%) and ≥65 years (40%). Of the alveolar echinococcosis cases with known sex (n=112), there were slightly more cases reported in males (53%) than in females. Poland reported a sharp decrease of cases caused by *E. multilocularis* compared to 2016–2019, but also reported a drop in all echinococcosis cases. Among cases with known importation status (n=17), 71% of cases with *E. multilocularis* were reported to have been infected within the reporting country in 2020.

Table 2. Reported confirmed echinococcosis cases, by species, EU/EEA, 2016–2020

Country	Total Cases					*E. granulosus*					*E. multilocularis*				
	2016	2017	2018	2019	2020	2016	2017	2018	2019	2020	2016	2017	2018	2019	2020
Austria	26	50	46	36	34	22	37	29	16	18	4	8	12	13	4
Belgium	17	13	15	22	19	UNK	9	10	12	10	UNK	4	5	10	8
Bulgaria	269	218	206	193	95	269	218	206	193	95	0	0	0	0	0
Croatia	9	15	4	3	3	UNK	UNK	UNK	UNK	UNK	UNK	UNK	UNK	UNK	UNK
Cyprus	0	0	0	0	1	0	0	0	0	UNK	0	0	0	0	UNK
Czechia	4	1	4	1	4	UNK	UNK	1	UNK	1	UNK	UNK	2	UNK	2
Estonia	0	1	0	2	1	0	0	0	UNK	1	0	1	0	UNK	0
Finland	4	5	1	8	4	4	5	1	8	3	0	0	0	0	UNK
France	38	53	62	45	53	0	5	21	10	11	38	48	41	35	42
Germany	181	141	176	149	152	122	86	93	87	70	40	35	59	40	46
Greece	18	15	11	7	7	UNK	UNK	UNK	UNK	7	UNK	UNK	UNK	UNK	0
Hungary	5	14	9	10	4	UNK	1	UNK	UNK	1	UNK	1	UNK	UNK	UNK
Iceland	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
Ireland	2	0	2	0	0	1	0	UNK	0	0	UNK	0	UNK	0	0
Latvia	11	6	10	6	2	1	4	5	4	2	1	UNK	1	UNK	0
Lithuania	26	53	50	81	37	5	19	11	30	UNK	10	20	17	21	UNK
Luxembourg	0	2	0	1	3	0	2	0	1	3	0	0	0	0	0
Malta	1	0	0	0	0	1	0	0	0	0	0	0	0	0	0
Netherlands	33	38	42	48	48	UNK	UNK	UNK	UNK	UNK	UNK	UNK	UNK	UNK	UNK
Norway	3	6	7	7	6	1	3	5	2	1	UNK	1	UNK	UNK	1
Poland	64	75	51	70	18	18	27	17	21	8	22	31	19	25	6
Portugal	2	2	9	5	1	2	UNK	9	5	1	0	UNK	0	0	0
Romania	13	14	4	1	0	UNK	UNK	UNK	UNK	0	UNK	UNK	UNK	UNK	0
Slovakia	4	7	10	11	3	1	2	3	3	1	2	3	3	8	2
Slovenia	3	7	6	6	3	UNK	UNK	3	1	1	UNK	UNK	UNK	UNK	UNK
Spain	87	83	68	34	8	1	4	12	6	1	UNK	UNK	UNK	UNK	UNK
Sweden	27	34	29	26	23	20	11	5	17	8	1	4	2	2	3
United Kingdom	ND	4	ND	3	ND	ND	4	ND	3	ND	ND	0	ND	0	ND
EU/EEA	847	857	822	775	529	468	437	431	419	243	118	156	161	154	114

UNK: Species unknown
ND: No data reported

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 2020. Additionally, since 2019 (2018 data), it has also been possible to report clinical presentation data, which help differentiate the two forms of the disease, to ECDC's TESSy database. 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.

The EU notification rate of confirmed human echinococcosis cases decreased in 2020 compared to the previous four years. There was no apparent impact of the COVID-19 pandemic on the reported case numbers in 2020. In addition, the withdrawal of the UK from the EU is likely to have had a negligible impact on the surveillance data for echinococcosis in 2020; the UK only marginally contributed to seven (0.21%) of the 3 301 confirmed cases reported between 2016 and 2020.

In a small number of countries, an increase in case notification rates has been observed in the recent years. This may be due to intensified surveillance and/or improved notification systems for echinococcosis. The increased awareness of the disease among clinicians and influx of migrants (people from endemic countries) may have also influenced the number of diagnosed cases in some countries [7].

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 [6,8] 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 [6,9], 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, and Switzerland [6]. 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 [10], or due to the expansion of fox habitats into urban areas [12] or may reflect an increased surveillance effort [6]. However, the lack of baseline data and standardised detection methods make it difficult to explain the geographical expansion of *E. multilocularis* in Europe [6]. 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 2020, *E. granulosus s.l.* infections in animals were mainly detected in sheep in Spain, Greece, and Italy.

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 [15].

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

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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