

SURVEILLANCE REPORT

Echinococcosis

Annual Epidemiological Report for 2022

Key facts

- In 2022, 27 European Union/European Economic Area (EU/EEA) countries reported 731 echinococcosis cases. Of these, 299 (41%) cases were reported as *Echinococcus granulosus sensu lato*, 185 (25%) as *E. multilocularis* and 247 (34%) as an unknown species.
- The EU/EEA notification rate for 2022 was 0.19 cases per 100 000 population, which is higher than the rates reported in 2020 and 2021, but consistent with the rates reported in 2018 and 2019.
- In 2022, the highest notification rates were reported in males aged 65 years and above (0.24 cases per 100 000 population) and in males aged 25–44 years (0.22 cases per 100 000 population).
- Among echinococcosis cases with available information for country of infection, 68% were domestic or infected within the EU/EEA, while 32% were linked to travel outside of the EU/EEA.

Introduction

Echinococcosis is a parasitic zoonotic disease (transmitted from animals to humans) caused by *Echinococcus* tapeworms in their larval stage. The most common mode of transmission to humans is accidental consumption of water or food that has been contaminated with tapeworm eggs. There are two main forms of the disease: cystic echinococcosis and alveolar echinococcosis. Both forms of the disease can be expensive and complicated to treat, sometimes requiring extensive surgery and/or prolonged drug therapy.

Cystic echinococcosis, also known as 'hydatid disease', is caused by infection with *Echinococcus granulosus sensu lato* tapeworms. Dogs are the definitive hosts for this tapeworm, while sheep, cattle, goats and pigs are intermediate hosts. Humans are accidental hosts. Infections in humans are often asymptomatic, but cystic echinococcosis can cause harmful, slowly enlarging cysts in the liver, lungs and other organs that often go undetected and untreated for years.

Alveolar echinococcosis is caused by infection with *Echinococcus multilocularis* tapeworms. Foxes, dogs and coyotes are the definitive hosts for this tapeworm, while small rodents are intermediate hosts. Although cases of alveolar echinococcosis in animals in endemic areas are relatively common, human cases are rare. The burden of disease in individuals with alveolar echinococcosis is much greater than for cystic echinococcosis. Alveolar echinococcosis is characterised by parasitic tumours in the liver, lungs, brain and other organs. If left untreated, it can be fatal.

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Suggested citation: European Centre for Disease Prevention and Control. Echinococcosis. In: ECDC. Annual Epidemiological Report for 2022. Stockholm: ECDC; 2024.

Methods

This report is based on data for 2022 retrieved from The European Surveillance System (TESSy) on 31 January 2024. TESSy is a system for the collection, analysis and dissemination of data on communicable diseases.

For a detailed description of the methods used to produce this report, refer to the Methods chapter of the 'ECDC Annual Epidemiological Report' [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].

In 2022, echinococcosis was under mandatory surveillance in 24 EU/EEA countries, and under voluntary surveillance in Belgium, France and the Netherlands [2]. Denmark and Italy did not have surveillance systems for echinococcosis. Most countries (26/27 countries) reported echinococcosis cases using the 2008, 2012 or 2018 EU case definitions, which are identical. France and Germany used other/unspecified definitions.

Most reporting countries provided case-based data, except for Bulgaria and the Netherlands, who reported aggregated data. Twenty countries had surveillance systems that integrated laboratory and epidemiological data from physicians or hospitals.

For 2020 and 2021, Spain did not receive data from all of its regions due to diverting resources to COVID-19 pandemic response; therefore, case numbers for this country were lower than expected and notification rates were not calculated. Data were not reported by Liechtenstein (for any year) or the United Kingdom (2018); the reasons for this are unknown.

Epidemiology

For 2022, 27 EU/EEA countries reported data on echinococcosis. Among these, 24 countries reported 731 confirmed cases. Three countries (Cyprus, Malta and Iceland) reported zero cases (Table 1). The EU/EEA notification rate for 2022 was 0.19 cases per 100 000 population, which is slightly higher than in 2021, but less than in 2018 (0.21 cases) and 2019 (0.20 cases). The notification rate in 2020 (0.16 cases) represents the lowest notification rate ever reported since surveillance of *Echinococcus* species began in the EU/EEA in 2007. In 2022, the highest notification rates were observed in Bulgaria (1.30 cases per 100 000 population), and Lithuania (2.64 cases per 100 000 population) (Table 1, Figure 1).

Sixteen countries provided information on hospitalisation, covering 39% (286 cases) of all echinococcosis cases in the EU/EEA in 2022. Just under half (136 cases; 47%) of these cases were hospitalised. Most (51%) of the hospitalised cases were reported by Poland (37 cases) and Spain (32 cases).

Sixteen countries provided information on the outcome of 408 cases. One death was reported.

Most echinococcosis cases (384/731 cases; 53%) were reported without information on importation and probable country of infection. Of the 347 cases with travel information available, 68% (237 cases) were domestic or infected within the EU/EEA, while 32% (110 cases) were linked to travel outside of the EU/EEA.

For further information by *Echinococcus* spp., see the detailed sections below.

Table 1. Confirmed echinococcosis cases and rates per 100 000 population by country and year, EU/EEA, 2018–2022

	2018		2019		202	0	202	1	2022		
Country	Number	Rate									
Austria	46	0.52	36	0.41	34	0.38	42	0.47	54	0.60	
Belgium	15	0.13	22	0.19	19	0.16	17	0.15	23	0.20	
Bulgaria	206	2.92	193	2.76	95	1.37	89	1.29	89	1.30	
Croatia	4	0.10	3	0.07	3	0.07	3	0.07	5	0.13	
Cyprus	0	0.00	0	0.00	1	0.11	0	0.00	0	0.00	
Czechia	4	0.04	1	0.01	4	0.04	1	0.01	10	0.10	
Denmark	NDR	NRC									
Estonia	0	0.00	2	0.15	1	0.08	4	0.30	1	0.08	
Finland	1	0.02	8	0.14	4	0.07	6	0.11	2	0.04	
France	62	0.09	55	0.08	55	0.08	75	0.11	79	0.12	
Germany	176	0.21	150	0.18	171	0.21	160	0.19	163	0.20	
Greece	11	0.10	7	0.07	7	0.07	4	0.04	5	0.05	
Hungary	9	0.09	10	0.10	4	0.04	7	0.07	9	0.09	
Iceland	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00	
Ireland	2	0.04	0	0.00	0	0.00	1	0.02	1	0.02	
Italy	NDR	NRC									
Latvia	10	0.52	6	0.31	5	0.26	6	0.32	4	0.21	
Liechtenstein	NDR	NRC									
Lithuania	50	1.78	81	2.90	37	1.32	20	0.72	74	2.64	
Luxembourg	0	0.00	1	0.16	3	0.48	1	0.16	1	0.15	
Malta	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00	
Netherlands	42	0.24	48	0.28	48	0.28	53	0.30	45	0.26	
Norway	7	0.13	7	0.13	6	0.11	11	0.20	9	0.17	
Poland	51	0.13	70	0.18	18	0.05	26	0.07	46	0.12	
Portugal	9	0.09	5	0.05	1	0.01	2	0.02	2	0.02	
Romania	4	0.02	1	0.01	0	0.00	1	0.01	4	0.02	
Slovakia	10	0.18	11	0.20	3	0.05	2	0.04	6	0.11	
Slovenia	6	0.29	6	0.29	3	0.14	11	0.52	5	0.24	
Spain	68	0.15	34	0.07	8	NRC	33	NRC	72	0.15	
Sweden	29	0.29	26	0.25	23	0.22	25	0.24	22	0.21	
EU/EEA (30 countries)	822	0.21	783	0.20	553	0.16	600	0.17	731	0.19	
United Kingdom	NDR	NRC	3	0.00	NDR	NRC	NA	NA	NA	NA	
EU/EEA (31 countries)	822	0.21	786	0.17	553	0.16	NA	NA	NA	NA	

Source: Country reports.

NA: Not applicable; NDR: No data reported; NRC: No rate calculated.

No data from 2020 onwards were reported by the United Kingdom, due to its withdrawal from the EU on 31 January 2020.



Figure 1. Number of confirmed echinococcosis cases per 100 000 population by country, EU/EEA, 2022

Source: Country reports.

Age and gender

The male-to-female ratio in 2022 was 1.1:1 (Figure 2). The highest notification rates were reported in males aged 65 years and above (0.24 cases per 100 000 population) and 25–44 years (0.22 cases per 100 000 population).





Source: 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 and Sweden.

Echinococcosis by species

Among 24 countries reporting echinococcosis cases in 2022, species information was known for 484 of 731 (66%) cases from 19 countries. Five countries (Greece, Ireland, Latvia, the Netherlands and Romania) that reported echinococcosis cases did not report information on species or form of disease (Table 2).

Echinococcus granulosus sensu lato

Sixteen countries reported 299 confirmed cases of cystic echinococcosis caused by *E. granulosus sensu lato* in 2022 (Table 2). Similar to in 2020 and 2021, the number of reported cystic echinococcosis cases in 2022 was considerably lower than the average annual number of cases reported in 2018 and 2019 (299 cases in 2022 vs an average of 430 cases in 2018–2019). Bulgaria and Germany reported the greatest number of cases (89 and 96 cases, respectively), accounting for 62% of all cystic echinococcosis cases reported in 2022. Among the 208 cases with known age, 35% of cystic echinococcosis cases were reported in those aged 25–44 years, followed by 29% in those aged 45–64 years. Of the 205 cystic echinococcosis cases with known gender, 59% were female. Among the 127 cases with known importation status, 59% were reported to have been infected outside of the EU/EEA in 2022. Among the 59 cases with known hospitalisation information, just under half (28 cases; 47%) were hospitalised.

Echinococcus multilocularis

Thirteen countries reported 185 cases of alveolar echinococcosis caused by *E. multilocularis* in 2022 (Table 2). Similar to previous years, Germany and France accounted for most of reported *E. multilocularis* cases in the EU/EEA in 2022 (51%). Among the 185 cases with known age, most cases of alveolar echinococcosis were reported in the age groups 65 years and above (42%) and 45–64 years (30%). Of the 184 alveolar echinococcosis cases with known gender, there were slightly more cases reported in females than in males (54% in females vs 46% in males). Among the 83 cases with known importation status, 98% were reported to have been infected within the reporting country in 2022. Among the 82 cases with known information on hospitalisation, just under two-thirds (52 cases; 63%) were hospitalised.

Country	E. granulosus					E. multilocularis					Unknown Species					
	2018	2019	2020	2021	2022	2018	2019	2020	2021	2022	2018	2019	2020	2021	2022	
Austria	29	16	18	24	24	12	13	4	7	8	5	7	12	11	22	
Belgium	10	12	10	11	11	5	10	8	6	12	0	0	1	0	0	
Bulgaria	206	193	95	89	89	0	0	0	0	0	0	0	0	0	0	
Croatia	0	0	0	0	0	0	0	0	0	3	4	3	3	3	2	
Cyprus	0	0	0	0	0	0	0	0	0	0	0	0	1	0	0	
Czechia	1	0	1	0	3	2	0	2	1	4	1	1	1	0	3	
Estonia	0	0	1	0	0	0	0	0	0	1	0	2	0	4	0	
Finland	1	8	3	5	2	0	0	0	0	0	0	0	1	1	0	
France	21	20	13	22	25	41	35	42	53	54	0	0	0	0	0	
Germany	93	87	81	95	96	59	41	54	42	41	24	22	36	23	26	
Greece	0	0	7	0	0	0	0	0	0	0	11	7	0	4	5	
Hungary	0	0	1	0	7	0	0	0	0	1	9	10	3	7	1	
Iceland	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	
Ireland	0	0	0	1	0	0	0	0	0	0	2	0	0	0	1	
Latvia	5	4	5	6	0	1	0	0	0	0	4	2	0	0	4	
Lithuania	11	30	0	2	5	17	21	0	14	28	22	30	37	4	41	
Luxembourg	0	1	3	0	1	0	0	0	0	0	0	0	0	1	0	
Malta	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0	
Netherlands	0	0	0	0	0	0	0	0	0	0	42	48	48	53	45	
Norway	5	2	1	5	0	0	0	1	0	1	2	5	4	6	8	
Poland	17	21	8	12	11	19	25	6	11	26	15	24	4	3	9	
Portugal	9	5	1	2	1	0	0	0	0	0	0	0	0	0	1	
Romania	0	0	0	0	0	0	0	0	0	0	4	1	0	1	4	
Slovakia	3	3	1	0	1	3	8	2	2	4	4	0	0	0	1	
Slovenia	3	1	1	2	1	0	0	0	2	0	3	5	2	7	4	
Spain	12	6	1	1	11	0	0	0	0	0	56	28	7	32	61	
Sweden	5	17	8	12	11	2	2	3	0	2	22	7	12	13	9	
United Kingdom	NDR	3	NDR	NDR	NDR	NDR	0	NDR	NDR	NDR	NDR	0	NDR	NDR	NDR	
Total	431	429	259	289	299	161	155	122	138	185	230	202	172	173	247	

Table 2. Reported confirmed echinococcosis cases, by species, EU/EEA, 2018–2022

NDR: No data reported.

Denmark and Italy are excluded, as they did not have surveillance systems for echinococcosis. No data from 2020 onwards were reported by the United Kingdom, due to its withdrawal from the EU on 31 January 2020.

Outbreaks and other threats

No national or multi-county echinococcosis outbreaks were reported through EpiPulse in 2022.

Discussion

In 2022, the EU/EEA notification rate of confirmed human echinococcosis cases showed evidence of a progressive resurgence of cases following the COVID-19 pandemic.

The rise in echinococcosis cases in a number of countries over the last five years may be attributed to the increased surveillance activities and improved notification systems for echinococcosis in humans. Other factors include increased awareness about the disease among clinicians, as well as immigration from endemic countries [4]. The recent discovery of alveolar echinococcosis in countries such as Croatia and Serbia also indicates a geographical spread of the parasite [5-6].

Distinction between infection with *E. granulosus s.l.* and *E. multilocularis* is important because the two diseases they cause (cystic echinococcosis and alveolar echinococcosis, respectively) 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 their years-long incubation periods and the high proportion of asymptomatic or pauci-symptomatic carriers who never seek medical attention (cystic echinococcosis), as well as the underreporting/misdiagnosis of cases (alveolar echinococcosis and cystic echinococcosis) – factors that contribute to the neglected status of these diseases [7, 8].

For these reasons, the data on the number of people with echinococcosis are considered to represent only a small proportion of the true prevalence of echinococcosis in Europe. Indeed, a cross-sectional, ultrasound-based survey conducted in Romania and Bulgaria estimated approximately 45 000 human cystic echinococcosis cases in rural areas of these two endemic European countries [9].

Since the beginning of the surveillance of human echinococcosis in the EU/EEA 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 distribution of the two forms of disease varies in different parts of the EU/EEA. *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 [10, 12-13]. In contrast, *E. multilocularis* is endemic in the fox population in central Europe [10, 11] and human cases of alveolar echinococcosis are principally reported by France, Germany and Poland, with most cases considered to be infected within the reporting country.

In accordance with Regulation (EU) 2018/772 regarding preventive health measures for the control of *E. multilocularis* infection in dogs, surveillance of this pathogen focuses mainly on red foxes as definitive hosts. In 2022, *E. multilocularis* infections were primarily detected in foxes in Czechia, Germany and Poland [10]. Surveillance 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 [14]. The exact reasons for this are unclear but may be linked to growth in the European fox population [14] or expansion of fox habitats into urban areas [14]. It could also reflect an increased surveillance effort [10]. However, the lack of baseline data and standardised detection methods make it difficult to explain the geographical expansion of *E. multilocularis* in Europe [10]. 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 2021, *E. granulosus s.l.* infections in animals were mainly detected in small ruminants (sheep and goats) in Spain, Greece, Italy and Slovakia [10]. Currently, the European Register of Cystic Echinococcosis (ERCE) attempts to collect harmonised clinical data in the EU on a voluntary basis [16-19]. In the past, this was done by the European (Alveolar) Echinococcosis Registry (EurEchinoReg) [8].

Public health implications

The most common mode of transmission to humans is accidental consumption of water or food that has been contaminated with tapeworm eggs. It is important to wash hands with soap and warm water after handling dogs and before handling food. Prevention measures for cystic echinococcosis involve preventing transmission of the parasite, such as limiting the areas where dogs are allowed and preventing animals from consuming meat infected with cysts. Food or water that may have been contaminated by faecal matter from dogs should not be consumed by humans. Prevention measures for alveolar echinococcosis include avoiding contact with wild animals such as foxes and coyotes, as well as dogs and their faecal matter, and limiting the interactions between dogs and rodents.

Reporting of echinococcosis cases should include species information and preferably data collected at the NUTS-2 or NUTS-3 level. This would allow for 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.

References

- 1. European Centre for Disease Prevention and Control (ECDC). Introduction to the Annual Epidemiological Report. In: ECDC. Annual epidemiological report. Stockholm: ECDC; 2022. Available at:
- <u>https://www.ecdc.europa.eu/en/surveillance-and-disease-data/annual-epidemiological-reports/introduction-annual</u>
 European Centre for Disease Prevention and Control (ECDC). Surveillance systems overview [downloadable
- spreadsheet]. Stockholm: ECDC; 2022. Available at: <u>https://www.ecdc.europa.eu/en/publications-data/surveillance-systems-overview-2022</u>
- 3. European Centre for Disease Prevention and Control (ECDC). Surveillance Atlas of Infectious Diseases. Stockholm: ECDC; 2022. Available at: <u>https://atlas.ecdc.europa.eu/public/index.aspx?Dataset=27&HealthTopic=18</u>
- 4. Casulli A, Abela-Ridder B, Petrone D, Fabiani M, Bobić B, Carmena D, et al. Unveiling the incidences and trends of the neglected zoonosis cystic echinococcosis in Europe: a systematic review from the MEmE project. The Lancet Infectious Diseases. 2023;23(3):e95-e107.
- Richter J, Esmann L, Lindner AK, Trebesch I, Equihua-Martinez G, Niebank M, et al. Cystic echinococcosis in unaccompanied minor refugees from Afghanistan and the Middle East to Germany, July 2016 through June 2017. European Journal of Epidemiology. 2019;34(6):611–612. Available at: <u>https://doi.org/10.1007/s10654-019-00492-8</u>
- Balen Topić M, Papić N, Višković K, Sviben M, Filipec Kanižaj T, Jadrijević S, et al. Emergence of Echinococcus multilocularisin central continental Croatia: A human case series and update on prevalence in foxes. Life (Basel). 2023;13(6): 1402. Available at: <u>https://doi.org/10.3390/life13061402</u>
- Lalošević D, Živojinov M, Isaković V, Ivanov D, Trivunović V, Ružić M. The first human case of multilocular echinococcosis recognized in Serbia. Serbian Archives of Medicine. 2023;151(7–8):453–456. Available at: <u>https://doi.org/10.2298/SARH230112060L</u>.
- 8. Casulli A. Recognising the substantial burden of neglected pandemics cystic and alveolar echinococcosis. Lancet Glob Health. 2020 Apr;8(4):e470-e471.
- 9. Kern P, Bardonnet K, Renner E, Auer H, Pawlowski Z, Ammann RW, et al. European echinococcosis registry: human alveolar echinococcosis, Europe, 1982-2000. Emerg Infect Dis. 2003 Mar;9(3):343-9.
- Tamarozzi F, Legnardi M, Fittipaldo A, Drigo M, Cassini R. Epidemiological distribution of *Echinococcus granulosus s.l.* infection in human and domestic animal hosts in European Mediterranean and Balkan countries: A systematic review. PLoS Negl Trop Dis. 2020;14(8):e0008519.
- 11. European Food Safety Authority (EFSA) and European Centre for Disease Prevention and Control (ECDC). The European Union One Health 2021 Zoonoses Report. EFSA Journal. 2022;20(12):7666. Available at: https://efsa.onlinelibrary.wiley.com/doi/full/10.2903/j.efsa.2022.7666
- 12. Deplazes P, Rinaldi L, Alvarez Rojas CA, Torgerson PR, Harandi MF, Romig T, et al. Global Distribution of Alveolar and Cystic Echinococcosis. Adv Parasitol. 2017;95:315-493.
- 13. Tamarozzi F, Akhan O, Cretu CM, Vutova K, Akinci D, Chipeva R, et al. Prevalence of abdominal cystic echinococcosis in rural Bulgaria, Romania, and Turkey: a cross-sectional, ultrasound-based, population study from the HERACLES project. Lancet Infect Dis. 2018;18(7):769-78.
- 14. Casulli A, Massolo A, Saarma U, Umhang G, Santolamazza F, Santoro A. Species and genotypes belonging to Echinococcus granulosus sensu lato complex causing human cystic echinococcosis in Europe (2000-2021): a systematic review. Parasit Vectors. 2022 Mar 28;15(1):109. Available at: https://doi.org/10.1186/s13071-022-05197-8
- 15. Davidson RK, Romig T, Jenkins E, Tryland M, Robertson LJ. The impact of globalisation on the distribution of *Echinococcus multilocularis*. Trends Parasitol. 2012 Jun;28(6):239-47.
- 16. Deplazes P, Hegglin D, Gloor S, Romig T. Wilderness in the city: the urbanization of *Echinococcus* multilocularis. Trends Parasitol. 2004 Feb;20(2):77-84.
- 17. Rossi P, Tamarozzi F, Galati F, Pozio E, Akhan O, Cretu CM, et al. The first meeting of the European Register of Cystic Echinococcosis (ERCE). Parasit Vectors. 2016 Apr;9:243.
- 18. Rossi P, Tamarozzi F, Galati F, Akhan O, Cretu CM, Vutova K, et al. The European Register of Cystic Echinococcosis, ERCE: state-of-the-art five years after its launch. Parasit Vectors. 2020 May;13(1):236.
- 19. Rossi P, Tamarozzi F, Galati F, Pozio E, Akhan O, Cretu CM, et al. The first meeting of the European Register of Cystic Echinococcosis (ERCE). Parasit Vectors. 2016;9:243.