SURVEILLANCE REPORT

Echinococcosis

Annual Epidemiological Report for 2021

Key facts

- In 2021, 26 EU/EEA countries reported 540 echinococcosis cases. Of these, 283 (52%) cases were reported as *Echinococcus granulosus sensu lato*, 137 (25%) as *E. multilocularis* and 120 (22%) as an unknown species.
- The EU/EEA notification rate for 2021 was 0.16 cases per 100 000 population, which is the same as the rate reported in 2020.
- In 2021, the highest notification rates were reported in males aged 65 years and above (0.21 cases per 100 000 population) and in females aged 25–44 years (0.20 cases per 100 000 population).
- Among echinococcosis cases with available information for country of infection, 58% were domestic or infected within the EU/EEA, while 42% 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 soil, 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 host 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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Stockholm, February 2024

Methods

This report is based on data for 2021 retrieved from The European Surveillance System (TESSy) on 9 October 2022. 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 2021, echinococcosis was under mandatory surveillance in 24 EU/EEA countries, and under voluntary surveillance in Belgium and France [2]. Denmark and Italy did not have surveillance systems for echinococcosis. Most countries (24/26 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, who reported aggregated data. Twenty countries had surveillance systems that integrated laboratory and epidemiological data from physicians or hospitals.

The Netherlands did not report any data for 2021; the reasons for this are unknown. 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. The United Kingdom (UK) contributed surveillance data up to 2019. No data were reported by the UK for 2020 or 2021 due to its withdrawal from the EU on 31 January 2020.

Epidemiology

For 2021, 26 EU/EEA countries reported data on echinococcosis. Among these, 23 countries reported 540 confirmed cases. Three countries (Cyprus, Malta and Iceland) reported zero cases (Table 1). The EU/EEA notification rate for 2021 was 0.16 cases per 100 000 population, which is the same as in 2020, but less than in 2019 (0.17 cases). The notification rate in 2020 and 2021 represents the lowest notification rate ever reported since surveillance of *Echinococcus* species began in the EU/EEA in 2007. In 2021, the highest notification rates were observed in Bulgaria (1.27 cases per 100 000 population), Lithuania (0.72 cases per 100 000 population) and Slovenia (0.52 cases per 100 000 population) (Table 1, Figure 1).

Fifteen countries provided information on hospitalisation, covering 27% (145 cases) of all echinococcosis cases in the EU/EEA in 2021. Half (72 cases; 50%) of these cases were hospitalised. Most of the hospitalised cases were reported by Austria (11 cases), Belgium (12 cases) and Poland (14 cases).

Sixteen countries provided information on the outcome of 270 cases. No deaths were reported.

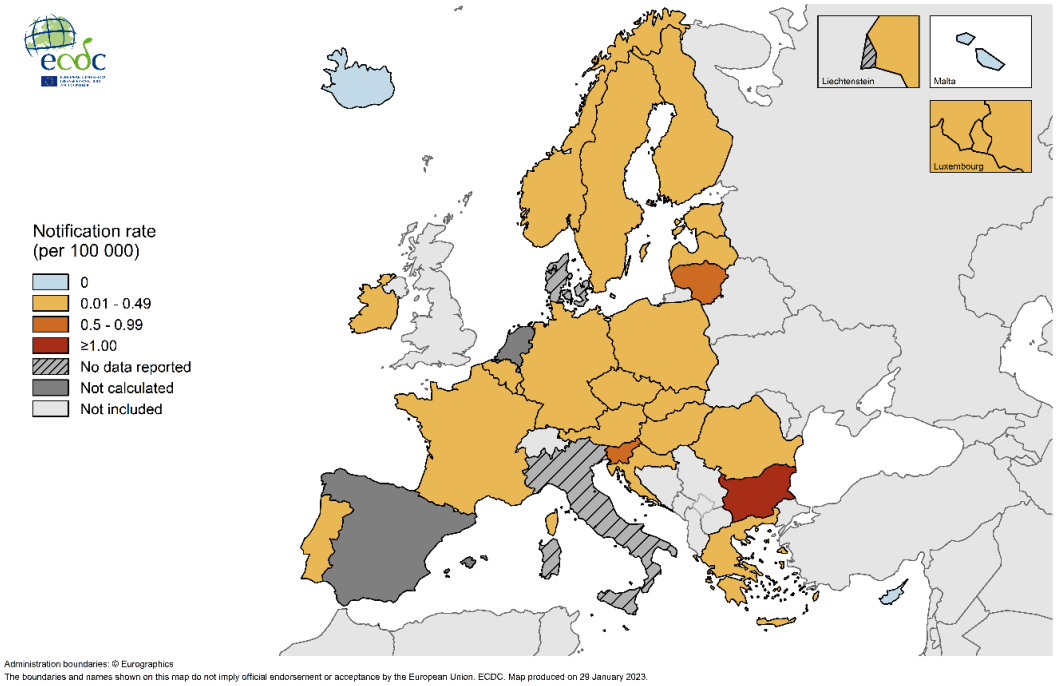
Most echinococcosis cases (316/540 cases; 59%) were reported without information on importation and probable country of infection. Of the cases with this information available, 57% (128/224 cases) were domestic or infected within the EU/EEA, while 41% (91/224 cases) were linked to travel outside of the EU/EEA. For further information by *Echinococcus* spp., see the detailed sections below.

Table 1. Number of confirmed echinococcosis cases and notification rates per 100 000 population, by country and year, EU/EEA, 2017–2021

Country	2017		2018		2019		2020		2021		
	Number	Rate	Number	Rate	Number	Rate	Number	Rate	Number	Rate	ASR
Austria	50	0.57	46	0.52	36	0.41	34	0.38	42	0.47	0.48
Belgium	13	0.11	15	0.13	22	0.19	19	0.16	18	0.16	0.15
Bulgaria	218	3.07	206	2.92	193	2.76	95	1.37	89	1.29	1.35
Croatia	15	0.36	4	0.10	3	0.07	3	0.07	3	0.07	0.08
Cyprus	0	0.00	0	0.00	0	0.00	1	0.11	0	0.00	0.00
Czechia	1	0.01	4	0.04	1	0.01	4	0.04	1	0.01	0.01
Denmark	NDR	NDR	NDR	NDR	NDR	NDR	NDR	NDR	NDR	NDR	NDR
Estonia	1	0.08	0	0.00	2	0.15	1	0.08	4	0.30	0.28
Finland	5	0.09	1	0.02	8	0.14	4	0.07	6	0.11	0.12
France	53	0.08	62	0.09	45	0.07	53	0.08	75	0.11	0.11
Germany	141	0.17	176	0.21	150	0.18	170	0.20	152	0.18	0.18
Greece	15	0.14	11	0.10	7	0.07	7	0.07	4	0.04	0.04
Hungary	14	0.14	9	0.09	10	0.10	4	0.04	7	0.07	0.07
Iceland	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00	0.00
Ireland	0	0.00	2	0.04	0	0.00	0	0.00	1	0.02	0.02
Italy	NDR	NDR	NDR	NDR	NDR	NDR	NDR	NDR	NDR	NDR	NDR
Latvia	6	0.31	10	0.52	6	0.31	5	0.26	6	0.32	0.28
Liechtenstein	NDR	NDR	NDR	NDR	NDR	NDR	NDR	NDR	NDR	NDR	NDR
Lithuania	53	1.86	50	1.78	81	2.90	37	1.32	20	0.72	0.70
Luxembourg	2	0.34	0	0.00	1	0.16	3	0.48	1	0.16	0.18
Malta	0	0.00	0	0.00	0	0.00	0	0.00	0	0.00	0.00
Netherlands	38	0.22	42	0.24	48	0.28	48	0.28	NDR	NDR	NDR
Norway	6	0.11	7	0.13	7	0.13	6	0.11	11	0.20	0.22
Poland	75	0.20	51	0.13	70	0.18	18	0.05	26	0.07	0.07
Portugal	2	0.02	9	0.09	5	0.05	1	0.01	2	0.02	0.01
Romania	14	0.07	4	0.02	1	0.01	0	0.00	1	0.01	0.01
Slovakia	7	0.13	10	0.18	11	0.20	3	0.05	2	0.04	0.04
Slovenia	7	0.34	6	0.29	6	0.29	3	0.14	11	0.52	0.49
Spain	83	0.18	68	0.15	34	0.07	8	NRC	33	NRC	NRC
Sweden	34	0.34	29	0.29	26	0.25	23	0.22	25	0.24	0.26
United Kingdom	4	0.01	NRC	NRC	3	0.00	NDR	NDR	NDR	NDR	NDR
EU/EEA	**857**	**0.19**	**822**	**0.21**	**776**	**0.17**	**550**	**0.16**	**540**	**0.16**	**0.16**

ASR: age-standardised rate; NDR: no data reported; NRC: no rate calculated.
Source: Country reports.
Denmark and Italy had no surveillance systems for echinococcosis from 2017 to 2021. For 2020 and 2021, Spain did not receive data from all of its regions due to diverting resources to COVID-19 pandemic response. The United Kingdom (UK) did not report data for 2020 or 2021 due to its withdrawal from the EU on 31 January 2020. Data were not reported by Liechtenstein (for any year), the Netherlands (2021) or the UK (2018); the reasons for this are unknown.

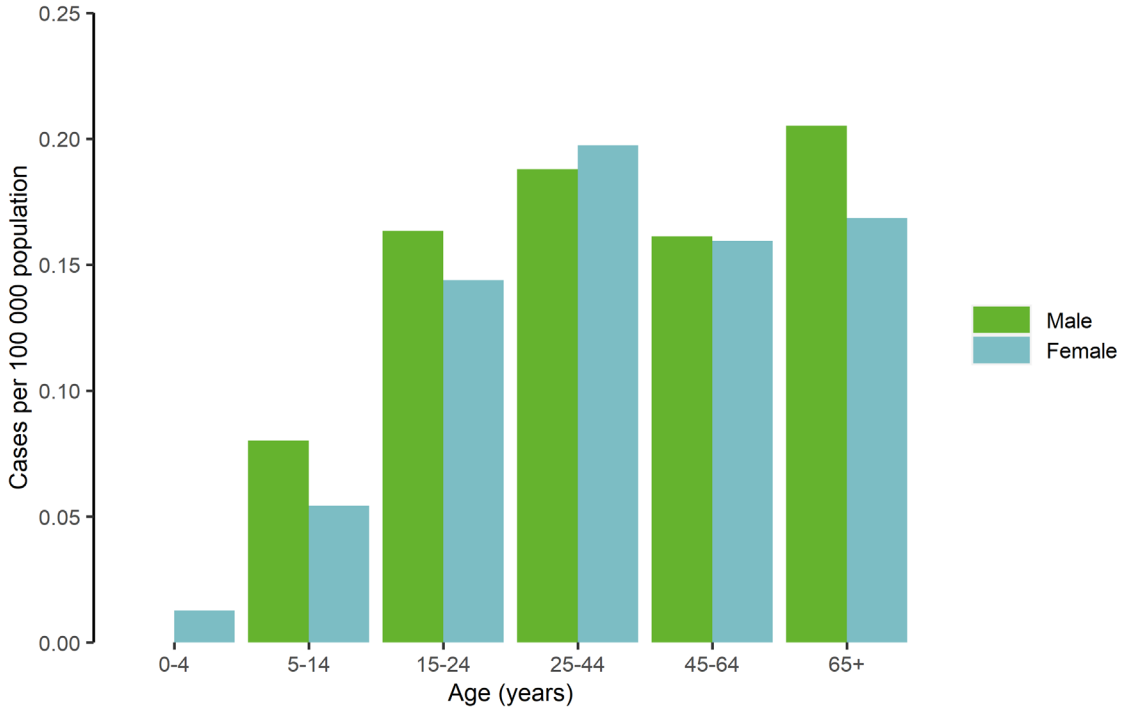
Figure 1. Number of confirmed echinococcosis cases per 100 000 population by country, EU/EEA, 2021



Source: Country reports.

The male-to-female ratio in 2021 was 1:1 (Figure 2). The highest notification rates were reported in males aged 65 years and above (0.21 cases per 100 000 population) and in females aged 25–44 years (0.20 cases per 100 000 population).

Figure 2. Number of confirmed echinococcosis cases per 100 000 population, by age and gender, EU/EEA, 2021



Source: country reports from Austria, Belgium, Bulgaria, Croatia, Cyprus, Estonia, Finland, France, Germany, Greece, Hungary, Iceland, Ireland, Latvia, Lithuania, Luxembourg, Malta, Norway, Poland, Portugal, Romania, Slovakia, Slovenia, Spain and Sweden.

Echinococcosis by species

Species information was known for 418 of 540 (77%) cases from 17 countries in 2021 (six countries who reported echinococcosis cases did not report information on species or form of disease) (Table 2).

Echinococcus granulosus sensu lato

Fifteen countries reported 283 confirmed cases of cystic echinococcosis caused by *E. granulosus s.l.* in 2021 (Table 2). Similar to in 2020, the number of reported cystic echinococcosis cases in 2021 was considerably lower than the average annual number of cases reported between 2017 and 2019 (281 cases in 2021 vs 429 cases between 2017–2019). Bulgaria and Germany reported the greatest number of cases (89 cases each), accounting for 63% of all cystic echinococcosis cases reported in 2021. Among the 281 cases with known age, 31% of cystic echinococcosis cases were reported in those aged 25–44 years, followed by 14% in those aged 45–64 years. Of the 276 cystic echinococcosis cases with known gender, 50% were female. Among the 109 cases with known importation status, 60% were reported to have been infected outside of the EU/EEA in 2021, compared with 68% in 2020. Among the 46 cases with known hospitalisation information, just over half (24 cases; 52%) were hospitalised.

Echinococcus multilocularis

Nine countries reported 137 cases of alveolar echinococcosis caused by *E. multilocularis* in 2021 (Table 2). Similar to in 2020, the number of reported alveolar echinococcosis cases in 2021 was lower than the average annual number of cases reported between 2017 and 2019 (137 cases in 2021 vs 157 cases between 2017–2019). Germany and France accounted for 68% of all reported *E. multilocularis* cases in the EU/EEA in 2021. Among the 135 cases with known age, most cases of alveolar echinococcosis were reported in the age groups 65 years and above (47%) and 45–64 years (33%). Of the 135 alveolar echinococcosis cases with known gender, there were slightly more cases reported in females than in males (51% in females vs 49% in males). Among the 41 cases with known importation status, 95% were reported to have been infected within the reporting country in 2021. Among the 40 cases with known information on hospitalisation, just under half (18 cases; 45%) were hospitalised.

Table 2. Reported confirmed echinococcosis cases, by species, EU/EEA, 2017–2021

Country	*E. granulosus s.l.*					*E. multilocularis*					Unknown Species				
	2017	2018	2019	2020	2021	2017	2018	2019	2020	2021	2017	2018	2019	2020	2021
Austria	37	29	16	18	24	8	12	13	4	7	5	5	7	12	11
Belgium	9	10	12	10	11	4	5	10	8	7	0	0	0	1	0
Bulgaria	218	206	193	95	89	0	0	0	0	0	0	0	0	0	0
Croatia	0	0	0	0	0	0	0	0	0	0	15	4	3	3	3
Cyprus	0	0	0	0	0	0	0		0	0	0	0	0	1	0
Czechia	0	1	0	1	0	0	2	0	2	1	1	1	1	1	0
Estonia	0	0	0	1	0	1	0	0	0	0	0	0	2	0	4
Finland	5	1	8	3	5	0	0	0	0	0	0	0	0	1	1
France	5	21	10	11	22	48	41	35	42	53	0	0	0	0	0
Germany	86	93	87	80	89	35	59	40	54	40	20	24	23	36	23
Greece	0	0	0	7	0	0	0	0	0	0	15	11	7	0	4
Hungary	1	0	0	1	0	1	0	0	0	0	12	9	10	3	7
Iceland	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
Ireland	0	0	0	0	1	0	0	0	0	0	0	2	0	0	0
Latvia	4	5	4	5	6	0	1	0	0	0	2	4	2	0	0
Lithuania	19	11	30	0	2	20	17	21	0	14	14	22	30	37	4
Luxembourg	2	0	1	3	0	0	0	0	0	0	0	0	0	0	1
Malta	0	0	0	0	0	0	0	0	0	0	0	0	0	0	0
Netherlands	0	0	0	0	NDR	0	0	0	0	NDR	38	42	48	48	NDR
Norway	3	5	2	1	5	1	0	0	1	0	2	2	5	4	6
Poland	27	17	21	8	12	31	19	25	6	11	17	15	24	4	3
Portugal	0	9	5	1	2	0	0	0	0	0	2	0	0	0	0
Romania	0	0	0	0	0	0	0	0	0	0	14	4	1	0	1
Slovakia	2	3	3	1	0	3	3	8	2	2	2	4	0	0	0
Slovenia	0	3	1	1	0	0	0	0	0	2	7	3	5	2	9
Spain	4	12	6	1	1	0	0	0	0	0	79	56	28	7	32
Sweden	11	5	17	8	12	4	2	2	3	0	19	22	7	12	13
United Kingdom	4	NDR	3	NDR	NDR	0	NDR	0	NDR	NDR	0	NDR	0	NDR	NDR
EU/EEA	**437**	**431**	**419**	**256**	**281**	**156**	**161**	**154**	**122**	**137**	**264**	**230**	**203**	**172**	**122**

NDR: No data reported.
Denmark and Italy are excluded due to having had no surveillance system for echinococcosis.

Outbreaks and other threats

In the summer of 2021, there was a transition from the Epidemic Intelligence Information System for Food and Waterborne Diseases (FWD-EPIS) to the new EpiPulse system for the reporting of outbreaks or unusual events of food- or waterborne diseases. No national or multicounty echinococcosis outbreaks were reported through EpiPulse in 2021.

Discussion

In 2020 and 2021, the EU/EEA notification rate and number of cases with infections caused by *Echinococcus* spp. was noticeably reduced compared with the previous three years. This is most likely due to the impact of the COVID-19 pandemic on the surveillance of echinococcosis in the EU/EEA. The withdrawal of the UK from the EU on 31 January 2020 likely had a negligible impact on the surveillance data for echinococcosis in 2020 and 2021, as the UK only marginally contributed to seven (0.21%) of the 3 302 cases reported between 2016 and 2019. Of note, in 2020 and 2021 – while the COVID-19 pandemic was ongoing – the EU/EEA notification rate was the same in both years.

Similar to previous years, in 2021, Bulgaria reported the highest notification rate in the EU/EEA (1.29 cases per 100 000 population). All Bulgarian cases were reported as cystic echinococcosis. These findings are consistent with the results of a recent review that identified south-eastern Europe as the epicentre of cystic echinococcosis in Europe [4]. The authors of the review also reported a general decrease in disease incidence in Europe, including in endemic southern and eastern European countries, where the disease has traditionally been highly prevalent. This decrease may be due to factors such as improved hygiene practices, migration of individuals from rural to urban areas at a national level, a decrease in sheep populations, an increase in intensive farming (often with higher biosecurity measures that help limit interactions between farmed animals and wildlife), and the implementation of national control programmes [5].

Distinction between infection with *E. granulosus s.l.* and *E. multilocularis* is important because the two diseases they cause (cystic echinococcosis and alveolar echinococcosis) 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 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 [6, 7].

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 [8].

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 [9, 11–12]. In contrast, *E. multilocularis* is endemic in the fox population in central Europe [9, 10] 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 2021, *E. multilocularis* infections were primarily detected in foxes in Czechia, France, Germany and Luxembourg [9]. 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 [13]. The exact reasons for this are unclear but may be linked to growth in the European fox population [13] or expansion of fox habitats into urban areas [14]. It could also be a reflection of an increased surveillance effort [9]. However, the lack of baseline data and standardised detection methods make it difficult to explain the geographical expansion of *E. multilocularis* in Europe [9]. 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 [9].

Currently, the European Register of Cystic Echinococcosis (ERCE) attempts to collect harmonised clinical data in the EU on a voluntary basis [15, 16, 17]. In the past, this was done by the European (Alveolar) Echinococcosis Registry (EurEchinoReg) [7].

Public health implications

The most common mode of transmission to humans is accidental consumption of soil, 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.

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