ECFSPR 2023 Annual Data Report

European Cystic Fibrosis Society
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Message from the ECFSPR Director

We are pleased to present the 2023 Annual Report from the European Cystic Fibrosis Society Patient Registry (ECFSPR).

As in previous years, this report includes longitudinal data analyses of key variables such as lung function, microbiology, and drug utilization across Europe. To enhance readability, detailed data tables are in Appendix 1. The full 2022 annual report, and a Highlights report, were published in June 2024. For the 2023 annual report, the Highlights were already published 5 months earlier in January 2025. As for the 2022 annual report, a PowerPoint slide deck featuring key maps, graphs and charts from the 2023 data can be found on our website for presentations and public use.

Access to highly effective CFTR modulators, which significantly impact the health of individuals with cystic fibrosis (CF) in Europe, has steadily improved but disparities remain. In many European countries, compared to other regions, such as the United States, regulatory restrictions on CFTR modulators limit access.

The ECFSPR aims to eventually cover all 53 countries in the World Health Organisation's (WHO) definition of Europe. This year's report includes data from 42 countries and 56,144 consenting individuals with CF. The epidemiological data are sourced from national CF registries and individual CF centres across Europe. Through close collaboration with these centres and registries we strive to ensure data accuracy and completeness.

The ECFSPR serves as a vital resource for the CF community, supporting quality improvement initiatives and research. Our registry provides essential population-level data, accessible to people with CF, clinicians, and researchers. By analysing this data, we gain insights into demographic shifts and clinical outcomes. Additionally, the registry facilitates research into questions that may not be easily addressed through controlled trials.

Ensuring data quality remains a top priority, not only for delivering a reliable and comprehensive overview of CF outcomes across Europe but also for supporting registry-based pharmacovigilance (PMV) studies. These studies provide critical real-world evidence on the safety and efficacy of newly introduced CF treatments.

Our continued partnership with CF Europe and national patient organisations ensures that registry data are used to benefit the broader CF community. We are deeply grateful to the individuals with CF and their families across Europe who participate in the ECFSPR. I also extend my sincere thanks to the ECFSPR staff, the Executive and Scientific Committees, the volunteers contributing to our working groups and projects, and our sponsors for their invaluable support.

Sincerely,

Egil Bakkeheim

ECFSPR Director

To the people with cystic fibrosis

This report is about you and how cystic fibrosis (CF) affects you and other people all over Europe. The report is based on information collected by individual CF centres and the national CF registries that participate in the European Cystic Fibrosis Society Patient Registry (ECFSPR). We have tried to make the presentation of this data as clear as possible and hope that you will find the report interesting and easy to understand.
Also this year we have published a Highlights report containing key information from the Annual Report, specifically for people with CF and their families and anyone wishing to know a little more about the disease.
News, updates, and other interesting information are regularly posted on social media. You can find us on Facebook, Instagram and LinkedIn.
We will continue to work with patient organisations to increase awareness of the Registry among people with CF and their families. If you have suggestions on how we can improve or if anything is unclear you are welcome to contact us by email at info@ecfregistry.eu.
To discuss the results from your country in this report we encourage you to contact your CF centre. For more information about the Registry please visit the dedicated page for people with CF on our website.
Information on how we handle your data and how you can exercise your rights is available in the Privacy Notice.

We thank the people with CF, and their families, for consenting to their data being included in the European CF Society Patient Registry.

Introduction

The European Cystic Fibrosis Society Patient Registry (ECFSPR)

The ECFSPR collects demographic and clinical data of consenting people with cystic fibrosis from Europe and neighbouring countries. Data are collected using a common set of variables and definitions and are sent to the ECFSPR in one of the following ways:

- National CF registries (or individual centres with local databases) extract data from their own database and import the data into the secure, online ECFSPR data-collection software.
- Individual centres enter data directly into the ECFSPR software.

Collection of data at a local level must be approved by local data protection authorities in accordance with European data protection legislation. Data stored in the central database are pseudonymised, and only year/ month of birth and randomised centre and patient codes are used as identifiers.
ECFSPR data can be analysed for scientific purposes on application. All projects are reviewed by the ECFSPR Scientific Committee and, based on their recommendation, the country coordinators in the Steering Group (composed of national representatives of the countries that contribute data to the ECFSPR) decide if the data from their country can be used for a project; this decision is final. Projects originating from Industry are also reviewed by the ECFS Clinical Trials Network. All applications must meet the European and individual country data protection legislation regarding patient anonymity.
For more information, please visit our website.

General Considerations

It is possible that some national registries use data definitions and parameters that do not fully correspond to those employed by the ECFSPR, either because some types of information are not collected, or they are collected by the national registry using a different method. When the national registries upload their data, they are asked to state whether their variable definitions meet those of the ECFSPR or not. If there is a significant difference between the definitions, that variable has been omitted from the annual report for that country; if the difference is minor, we included the variable but added an explanatory footnote to the graphs and tables.
For example, the ECFSPR collects information on chronic Pseudomonas aeruginosa infection and uses the modified Leeds criteria and/or the presence of elevated anti-Pseudomonas antibodies (see Appendix 4). If a national registry collected such information as “at least one positive Pseudomonas aeruginosa culture this year”, this information would be significantly different from the ECFSPR definition of chronic Pseudomonas aeruginosa, and we would set this variable to “missing” for that particular country. If a country defined chronic Pseudomonas aeruginosa as “the presence of more than four positive cultures in 6 months”, this is much closer to the ECFSPR definition and we would include the variable and data in the annual, with a footnote added to the relevant tables and graphs.
If a country does not collect a certain variable we have omitted that country from the relevant graphs in the report but all of the data are presented in the tables. The same applies for countries where the information for a variable is missing for more than 10% of the people with CF. The countries with less than 5 individuals in an age group (e.g. less than 5 adults) are excluded from both the graphs and the tables. The number of missing values is important when interpreting the results, since it is impossible to know if a person with CF with a missing value for a given complication has this complication or not, meaning given frequencies are less accurate. For example, in a country where 7% of the people with CF have liver disease but for 20% the information on liver disease is unknown/missing, the true frequency of liver disease will be anything between 7 and 27%.
You will find some differences between the findings of the national registries’ own reports and the ECFSPR report. This is because some variable values are recoded or computed in different ways. For example, some national registries compute the age of the individual at the date of the annual visit and consider 16 years as the cut-off for adult age. The ECFSPR computes the age at FEV1/height/weight measurement and the age at follow-up (the end of the year) and considers 18 years as the cut-off for adult age. Another example: for lung function values such as FEV1 the raw data values, reported in litres, are not informative unless they are expressed in relation to the age, sex, and height of the individual. We therefore needed to transform the raw values into new variables to compare lung

function between people with CF in different countries. We used common reference populations for all data when calculating the values as a percentage of predicted from the raw data. Slightly different values can be obtained when using another reference population on the same raw data. It is important to use a common method of calculation when comparing different countries, just as the national registries choose a common method of calculation when they compare the individual centres in that country.

The estimated coverage, i.e. the percentage of people with CF included in the national registry or national data presented by the country, varies; see table 1.1. These differences can influence how the data are interpreted and we therefore advise comparisons to be made only between countries with similar coverage.

The date of the database that was used to create the tables and graphs in this report is 7 February 2025.

Summary of data report

Outcome		Females		Males		Total	
PwCF registered in the ECFSPR	n (%)	26714	(47.6)	29430	(52.4)	56144	(100)
Age at follow-up (years) (PwCF alive on 31/12/2023)	median (25^{th} pctl-75^{th} pctl)	19.8	(10.5-32.5)	20.8	(10.9-33.9)	20.3	(10.7-33.2)
PwCF ≥ 18 years (PwCF alive on 31/12/2023)	n (%)	14472	(54.5)	16532	(56.4)	31004	(55.5)
Age at diagnosis (months)*	median (25^{th} pctl-75^{th} pctl)	3.6	(1.2-32.4)	3.6	(1.2-31.9)	3.6	(1.2-32.0)
PwCF with at least one F508del allele recorded*	n (%)	20160	(80.1)	22055	(80.0)	42215	(80.0)
PwCF living with lung transplant**	n (%)	1245	(5.0)	1226	(4.5)	2471	(4.7)
PwCF living with liver transplant**	n (%)	106	(0.4)	230	(0.8)	336	(0.6)
PwCF deceased in 2023***	n (%)	138	(0.5)	145	(0.5)	283	(0.5)
Age at death (years)***	median (25^{th} pctl-75^{th} pctl)	31.0	(19.0-44.0)	35.0	(27.0-48.0)	34.0	(23.0-47.0)

* Only people with CF seen during the year by clinical staff. The total number presented is 52,952.

** Only people with CF alive at 31/12/2023. The total number of the CF population presented is 52,694.

*** Only people with CF seen during the year. For the United Kingdom, all individuals with a confirmed diagnosis of CF were included (N=11,318). The total number presented is 53,926.

Note: PwCF is an abbreviation for people with Cystic Fibrosis.

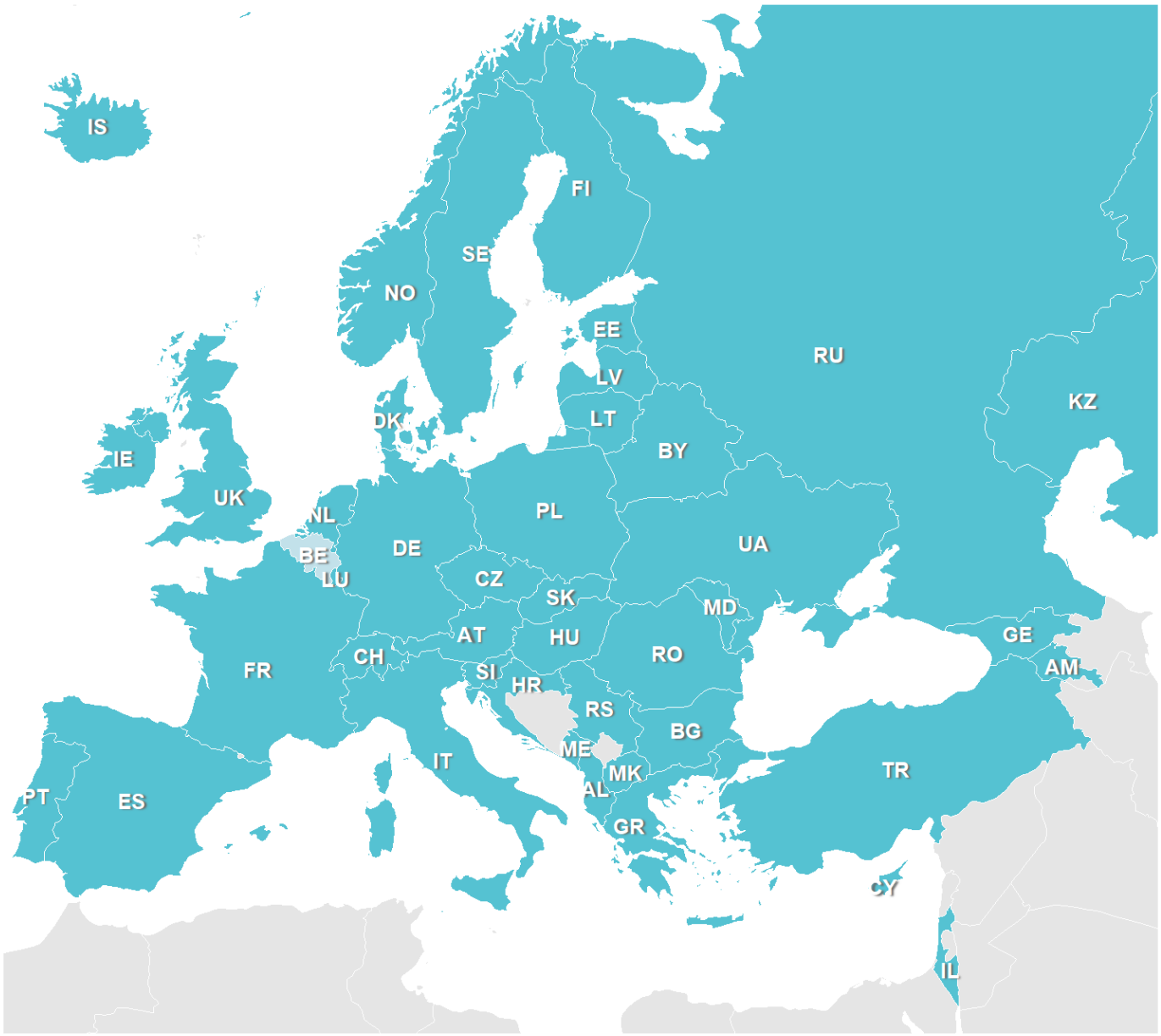
Data report

1. Demographics

The ECFSPR coverage (i.e. the proportion of the estimated total number of people with CF in a country included in the registry) continues to grow. Data come from national registries with their own collection systems or from centres in other countries that input data directly to our custom-designed data collection platform.
Only a few countries in Europe have not yet contributed data to the ECFSPR and we are in contact with those remaining to welcome them. In some countries not everyone with CF has had the opportunity to contribute to the ECFSPR and we invite all CF centres to join. We are confident that over the next few years several more centres and countries will do just that. National Coordinators that have been appointed by their country assist in this process by encouraging local centres to become new ECFSPR contributors.
This chapter gives information on coverage as well as on age and sex distribution in Europe and in the participating countries. In countries with a lower coverage, age distribution and mean age of the people with CF might be skewed since not all CF centres for children and adults in the country have contributed data. For more details, please refer to the information given in the footnotes of the tables and graphs.

1. Demographics

Figure 1.1 Map of countries that contributed data to the ECFSPR for the year 2023.



Note: BE could not provide data due to internal technical software issues and is marked in light blue.

The countries that contributed 2023 data are in turquoise.

1. Demographics

Table 1.1 Number of people with CF in 2023, by country.

Country	People with CF, registered, not lost to follow-up	People with CF seen	Estimated coverage
Albania	96	53	>80%
Armenia	32	32	>70%
Austria	877	855	90%
Belarus*	152	152	68%
Bulgaria	237	230	>90%
Croatia**	150	144	>98%
Cyprus	31	22	>80%
Czech Republic*	738	713	99%
Denmark*	581	578	99%
Estonia	45	45	75%
Finland	76	76	>60%
France*	7329	7329	>90%
Georgia	82	54	>90%
Germany*	7547	7179	>80%
Greece*	684	621	90%
Hungary*	507	507	98%
Iceland	16	16	>90%
Ireland*	1385	1359	92%
Israel**	587	541	>95%
Italy*	6138	6127	98%
Kazakhstan[1]	27	27	15%
Latvia	53	53	90%
Lithuania	48	46	75%
Luxembourg[2]	27	25	60%
Republic of Moldova	51	46	>90%
Montenegro	40	40	>80%
The Netherlands*	1644	1621	95%
North Macedonia**	167	139	>80%
Norway*	388	382	>90%
Poland	1705	1609	95%
Portugal**	410	397	>95%
Romania	416	396	50%
Russian Federation*	4102	3122	90%
Serbia	201	172	>90%
Slovak Republic**	309	280	>90%
Slovenia**	119	118	>95%
Spain	2670	2542	85%
Sweden*	808	785	>98%
Switzerland**	1082	997	>99%
Türkiye*	2761	2746	>70%
Ukraine***	508	432	>50%
United Kingdom*	11318	10344	99%
Total	56144	52952	

* Countries with an established national CF registry.

** These countries are defined as a national registry since all centres in the country participate in the ECFSPR.

*** It is unknown how many people with CF left the country after the start of the war.

1 Kazakhstan sent data on people with CF aged 16 and older.

2 For Luxembourg the coverage for children with CF is >80% and for all people with CF it is 60%.

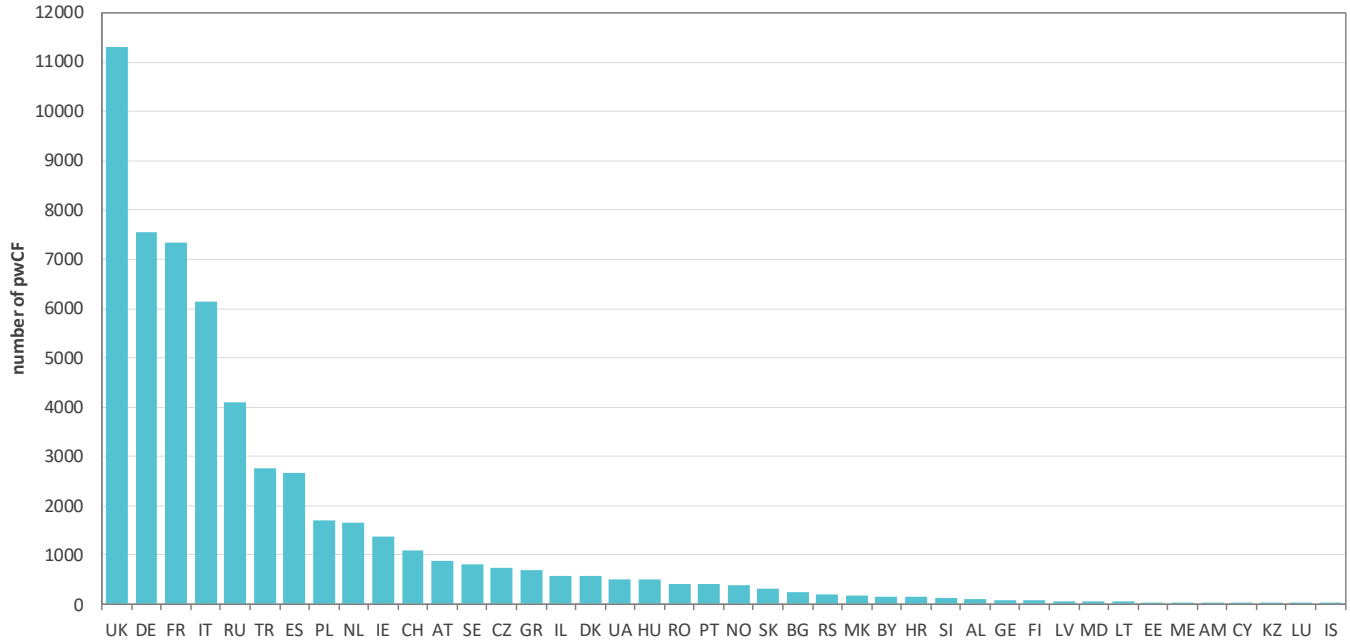
The column "People with CF, registered, not lost to follow-up" displays the individuals with CF that attended clinic and also those who were not seen by clinical staff during the year but were known to be alive that year.

The column “People with CF seen” presents only the individuals with CF who have attended the clinic during the year. The column “Estimated coverage” shows the estimated percentage of people with CF living in that country who are included in the national registry / national data collection as reported by the country. Some countries may only have one individual centre that includes almost all people with CF. From chapter 4 onwards, only people with CF seen during the year who have never had a liver/lung transplant are considered for the analyses (total number 50083).

1. Demographics

Figure 1.2 The number of people with CF registered in the ECFSPR varies across countries.

Number of people with CF registered in the ECFSPR in 2023.



Each vertical bar shows the number of registered people with CF (excluding lost to follow-up) living in that country in 2023. Please refer to table 1.1 for the coverage in each country.

1. Demographics

Figure 1.3 The number of countries and people with CF in the ECFSPR has risen continuously over the years.
Number of registered people with CF and number of countries from 2008 to 2023.

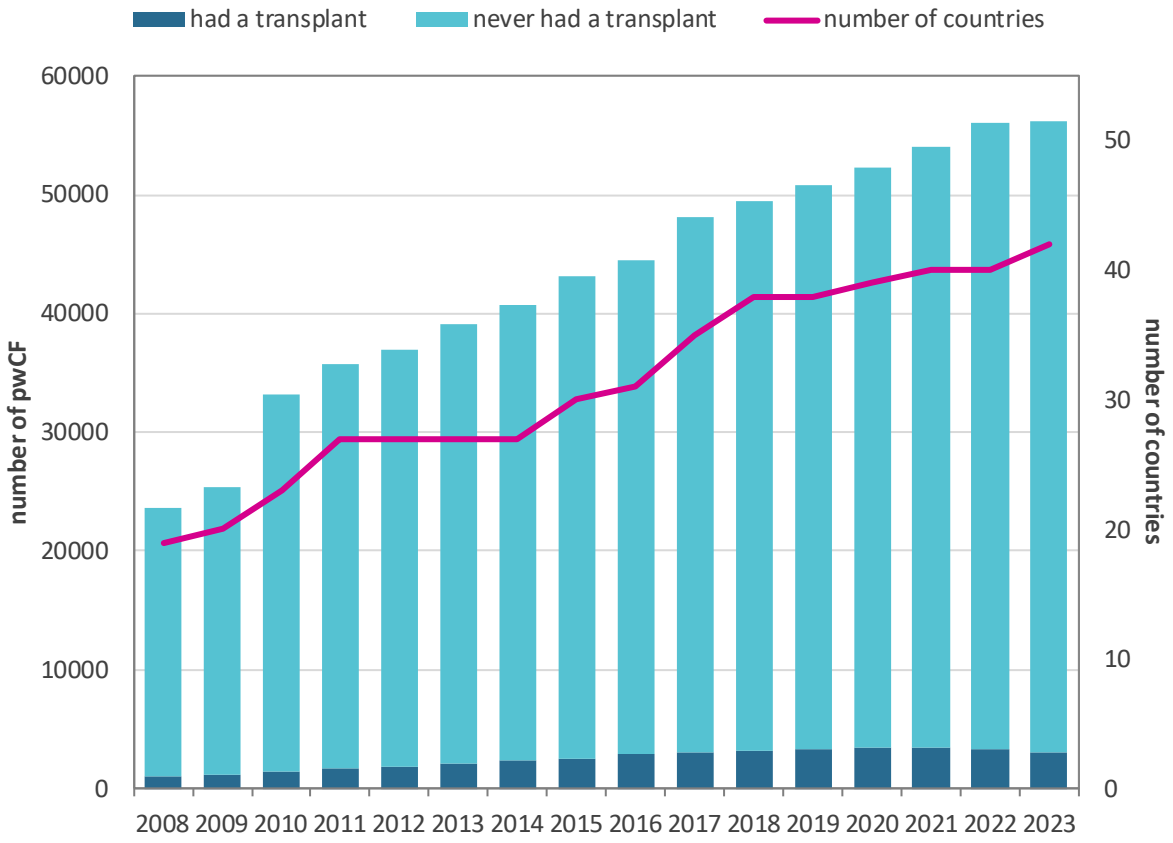
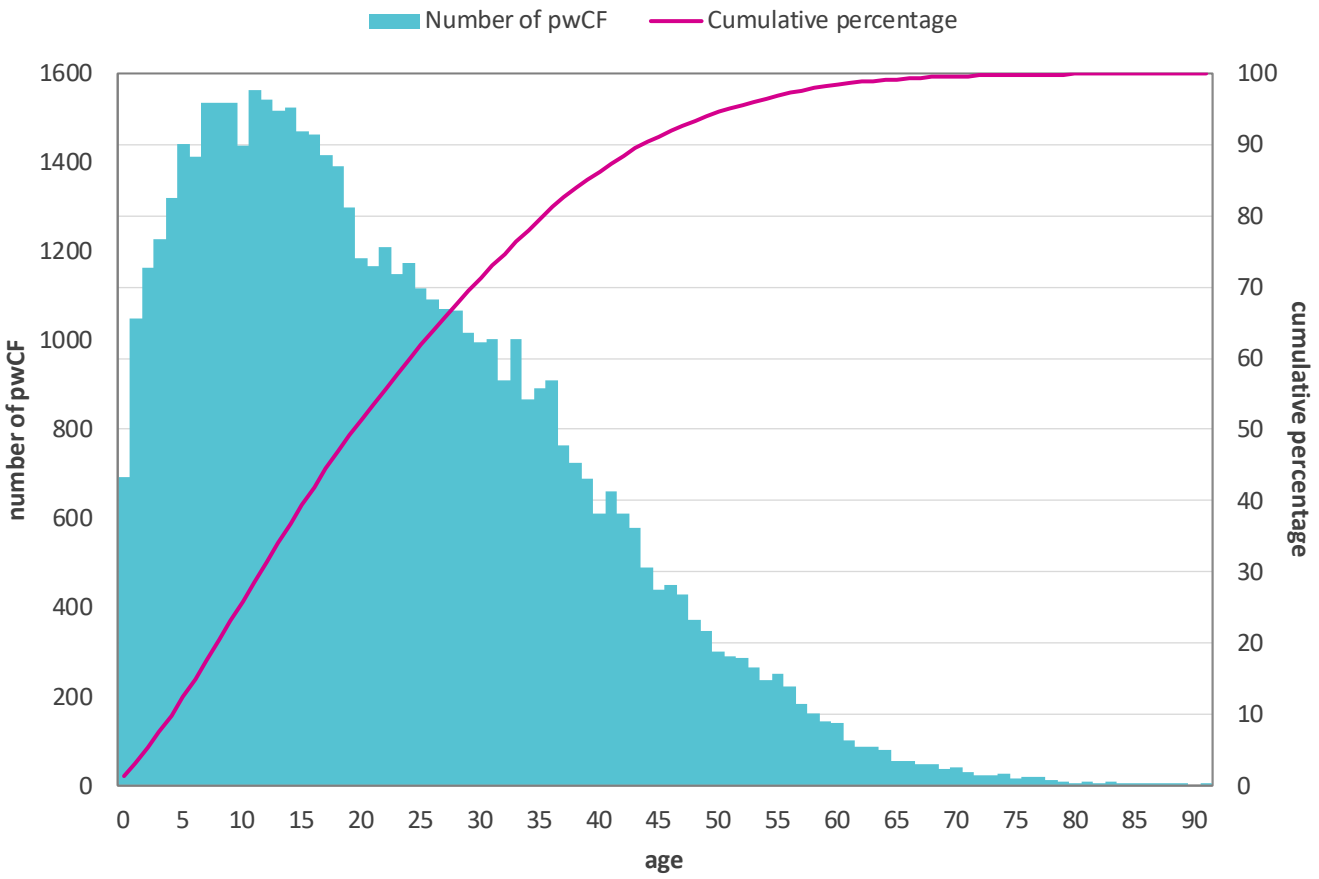


Figure 1.3 presents data over time using cross sectional data per year of people with a confirmed CF diagnosis. All people with CF alive, deceased, or not seen but alive during the year of follow-up were included.

1. Demographics

Figure 1.4 Age distribution demonstrates a sharp decline from the third decade of life.

Distribution of age at follow-up. Registered people with CF alive on 31/12/2023.

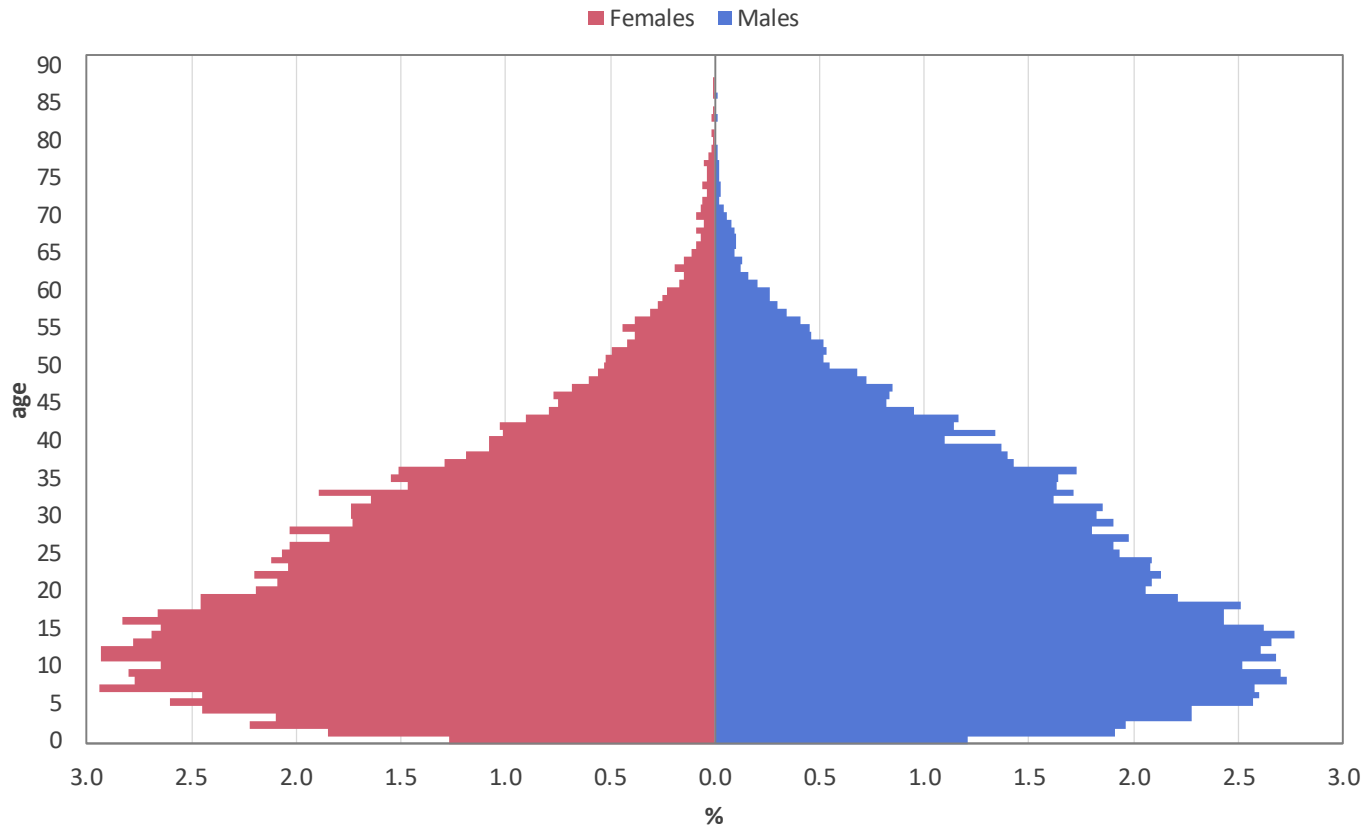


Each vertical bar shows the number of people with CF of that age alive in 2023. The cumulative percentage (the pink line) describes how many people with CF, as a percentage, are below a certain age. For example, 50% are younger than 19 years old.

1. Demographics

Figure 1.5 Age distribution is significantly skewed towards childhood and adolescence in CF.

Distribution of age at follow-up (in years) by sex. Registered people with CF alive on 31/12/2023.

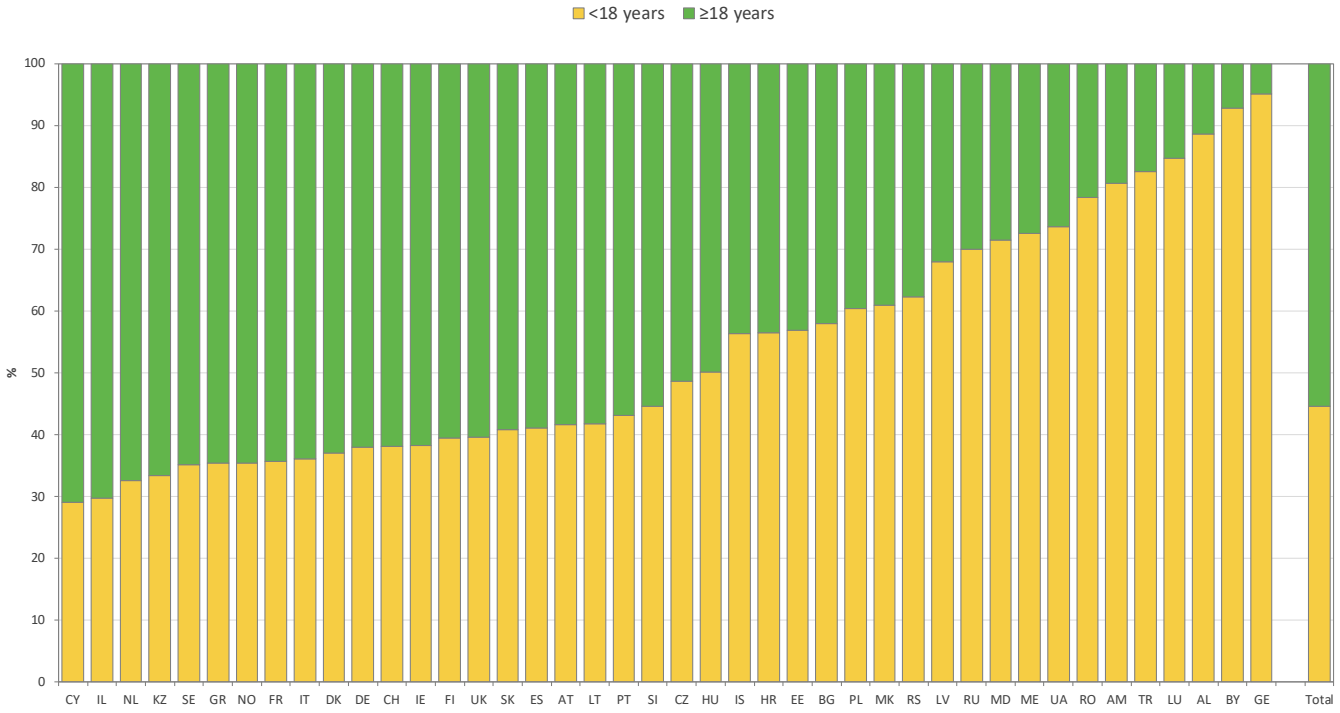


The pyramid shows the percentage of people with CF of different ages as horizontal bars. The left side of the pyramid (red) shows how many females with CF, as a percentage, are of a certain age; the right side (blue) shows the same for males. The lower percentage of children with CF at the bottom of the pyramid is because some children are not diagnosed early in life.

1. Demographics

Figure 1.6 The proportion of adults with CF varies considerably between European countries.

Proportion of children (<18 years) and adults (≥18 years), by country and overall. Registered people with CF alive on 31/12/2023 (table A1.1, Appendix 1).

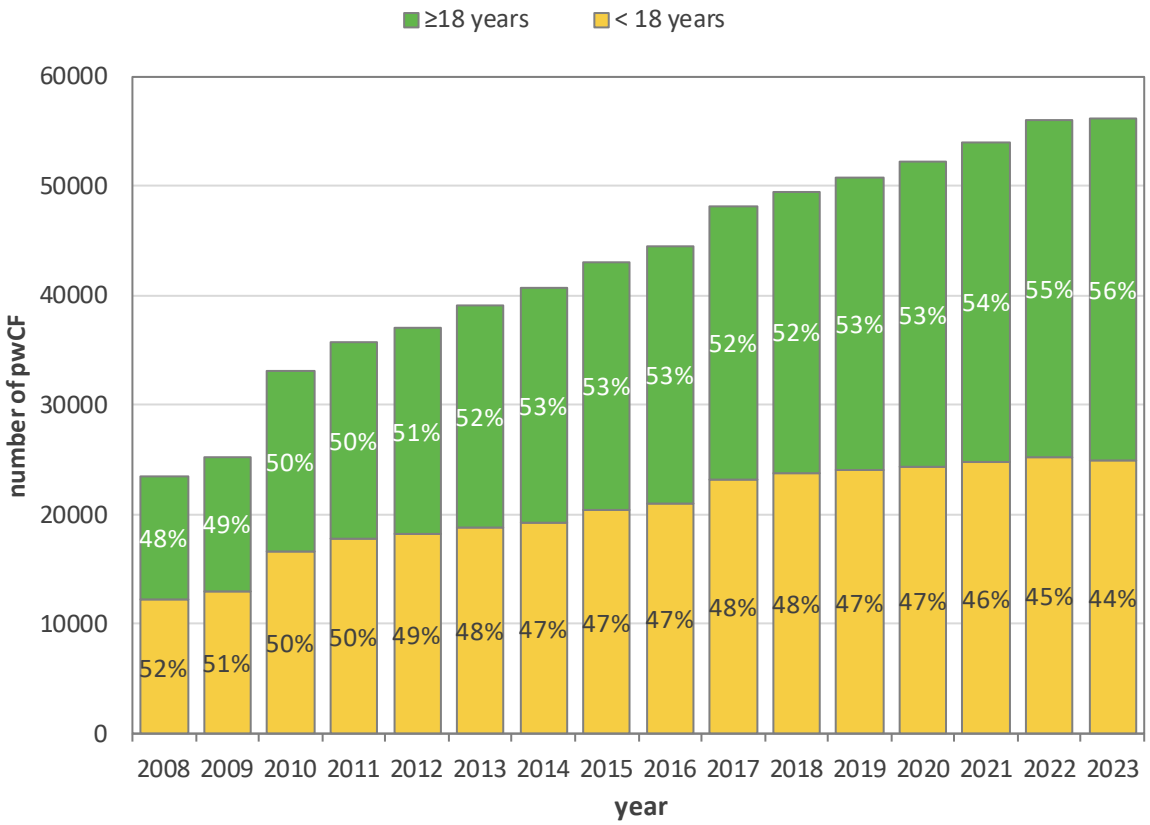


The yellow vertical bar shows the percentage of children and adolescents with CF living in that country in 2023, the green vertical bar shows the percentage of adults. Overall (see "Total") in the ECFSPR there are more adults than children.

1. Demographics

Figure 1.7 In recent years the proportion of adults with CF in Europe has risen significantly; as of 2023, adults made up 56% of the total.

Number of registered people with CF and percentage of adults and children from 2008 to 2023.

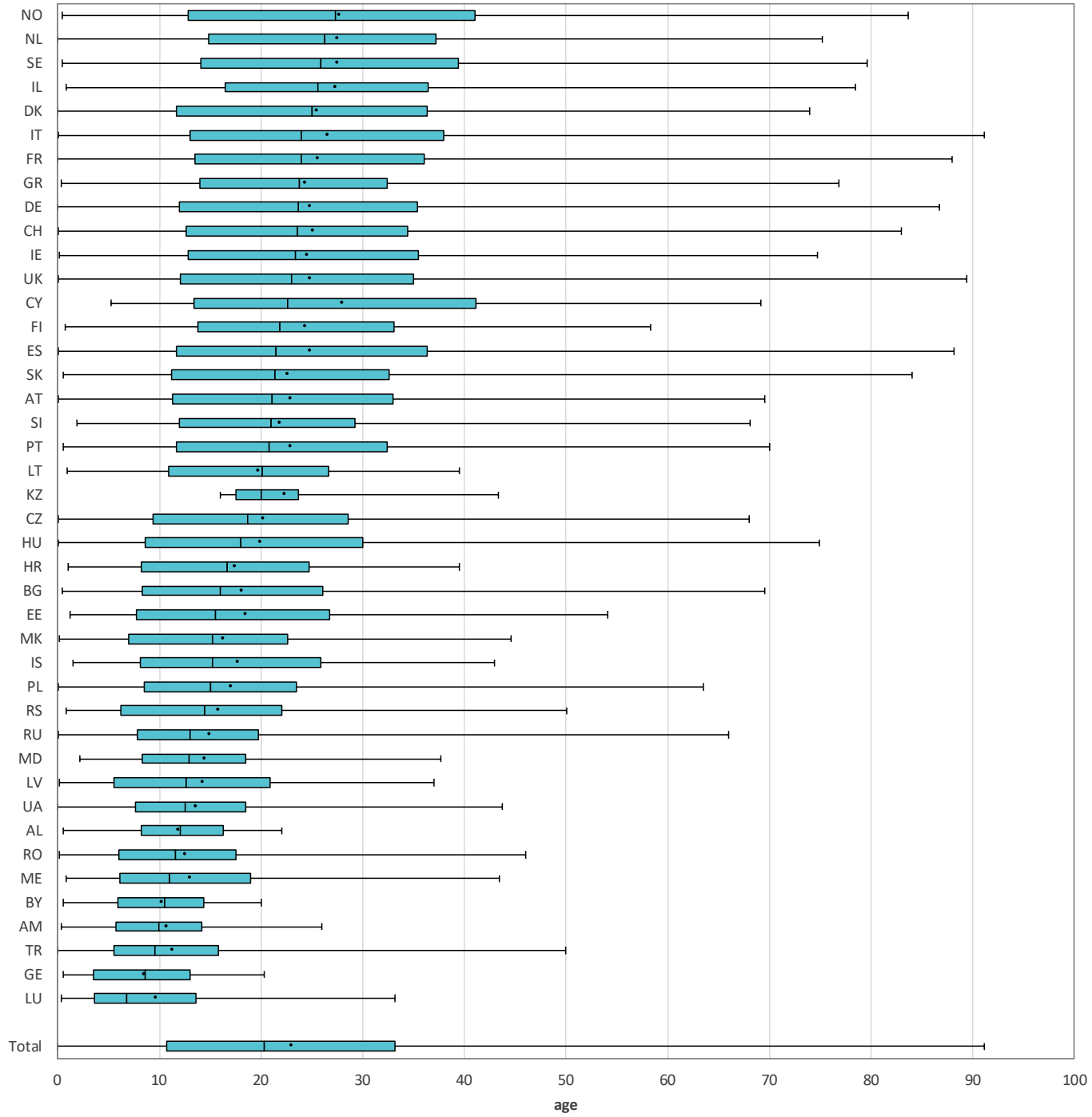


This graph presents data over time using cross sectional data per year of people with a confirmed CF diagnosis. All people with CF alive, deceased, or not seen but alive during the year of follow-up were included.

1. Demographics

Figure 1.8 The mean age of the CF population is not homogenous in Europe and depends on the country or region of residence.

Age at follow-up: box plot, by country and overall. Registered people with CF alive on 31/12/2023 (table A1.2, Appendix 1).

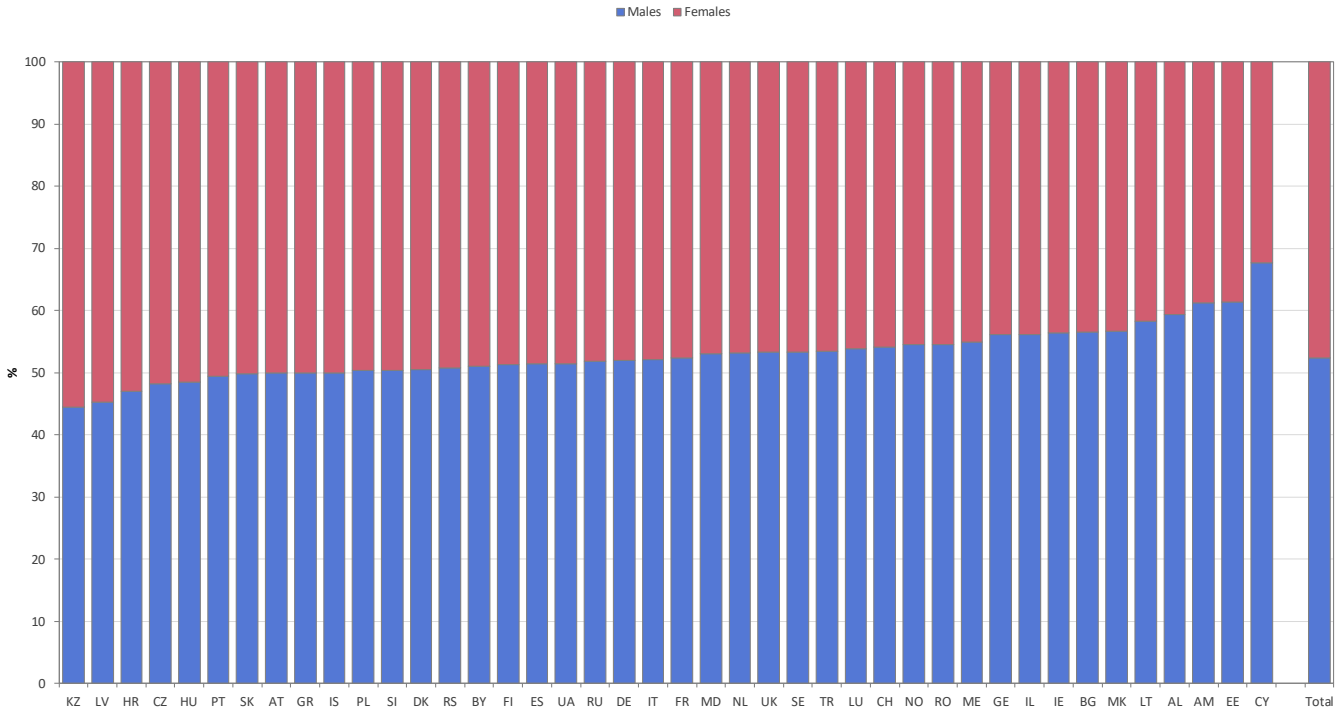


This box plot is a graphic representation of the age at follow-up detailed in table A1.2, Appendix 1. For each country the vertical borders of the box are the first and third quartile, the dash (vertical black line crossing the box) is the median, the black dot is the mean and the whiskers (lines with a T-shaped end) are the minimum and the maximum.

1. Demographics

Figure 1.9 Sex distribution is comparatively homogenous throughout Europe except for in a few countries.

Sex distribution, by country and overall. Registered people with CF alive on 31/12/2023.



Sex distribution of the CF population. Overall (see “Total”) in the ECFSPR there are slightly more males than females with CF.

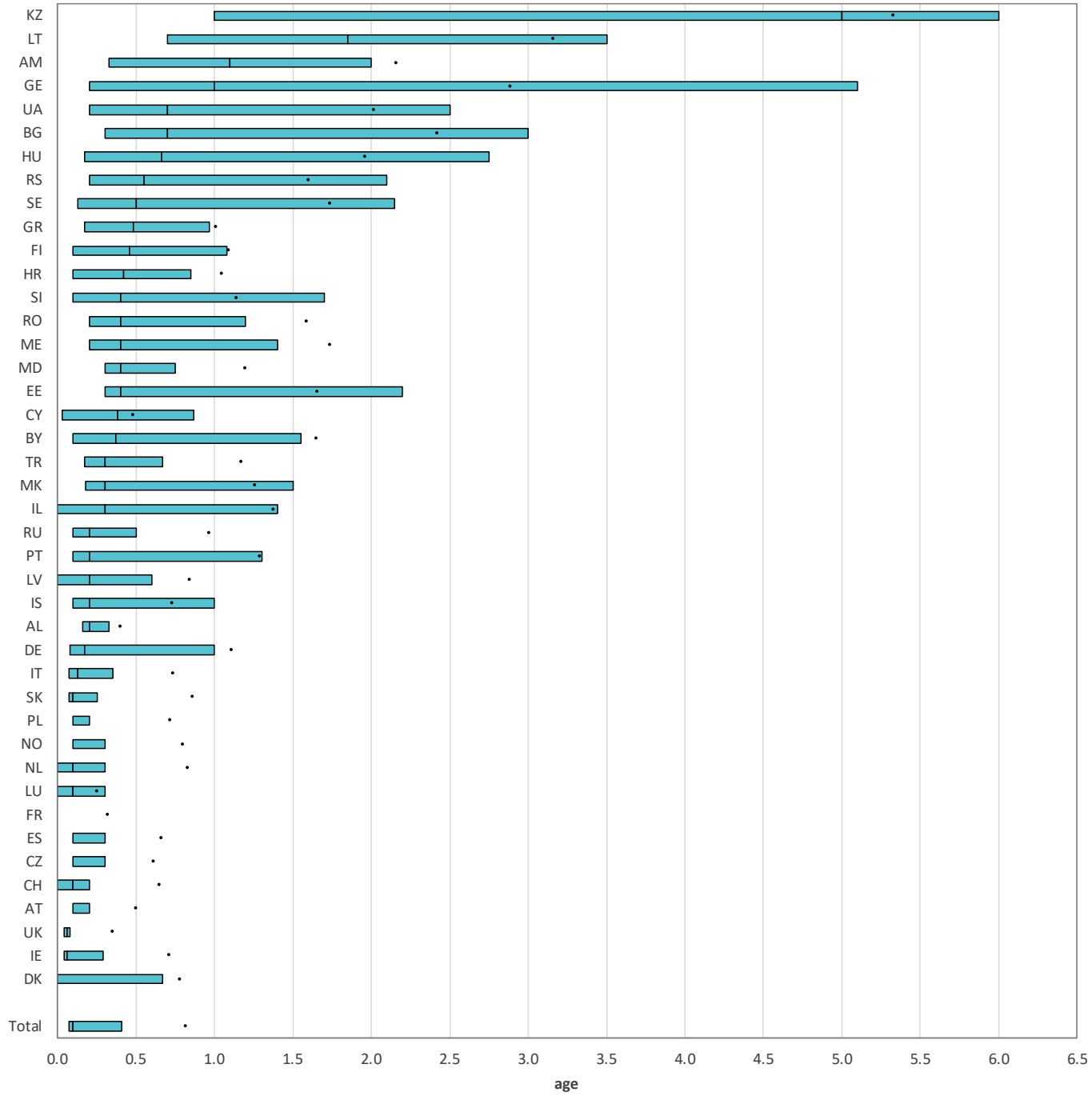
2. Diagnosis

In the following tables and figures the age at diagnosis and information on newborn screening are shown. The age at diagnosis, particularly in children and adolescents, is strongly influenced by the existence of a national CF newborn screening program. Information on the proportion of people with CF per country diagnosed by newborn screening is therefore also depicted. In some cases, meconium ileus at birth might trigger further investigations to exclude or diagnose CF, even though its prevalence differs considerably between the countries, as highlighted in one of the graphs below.
In this chapter and the following ones, only data for people with CF seen during the year are presented.

2. Diagnosis

Figure 2.1 Age at diagnosis in children and adolescents depends on various factors, including the existence or not of a newborn screening programme in the country.

Age at diagnosis (in years): boxplot, by country and overall. All children and adolescents (<18 years) alive and seen in 2023 (table A2.1, Appendix 1).



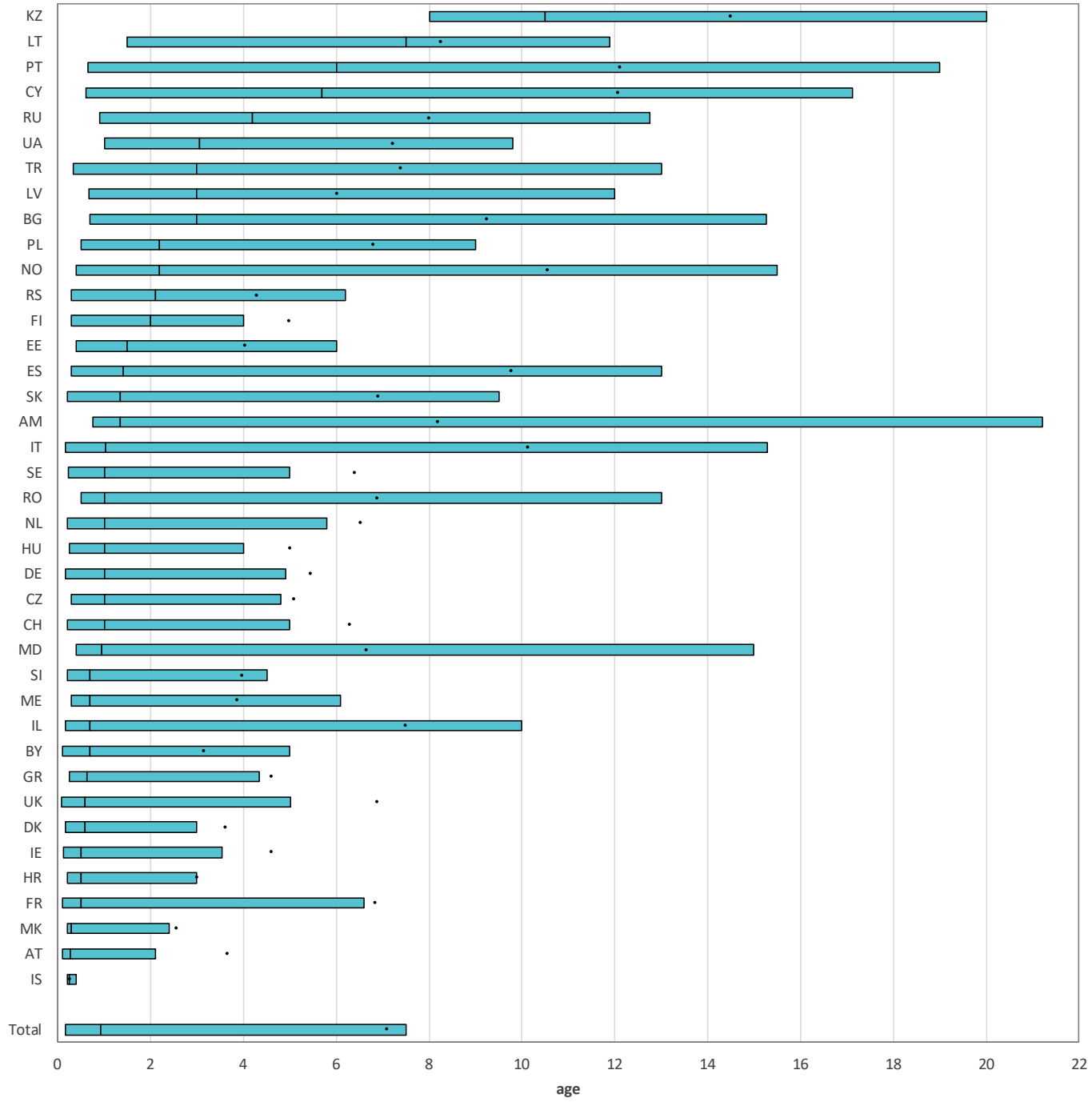
Note: For Cyprus, Greece, Lithuania and the Slovak Republic the information on age at diagnosis is missing for more than 10% of the children.

For each country the vertical borders of the box are the first and third quartiles, the dash (vertical black line crossing the box) is the median, the black dot is the mean. The whiskers that are the minimum and the maximum values are not shown because the maximum values are very high for some countries and this would have shrunk the boxes to the left of the graph.

2. Diagnosis

Figure 2.2 For adults the age at diagnosis reflects national differences in the diagnostic approach over the last decades.

Age at diagnosis (in years): boxplot, by country and overall. All adults (≥ 18 years) alive and seen in 2023 (table A2.2, Appendix 1).



Note: Albania, Georgia and Luxembourg have <5 adults seen in 2023 and are excluded from the table, but the individuals are included in the total number.

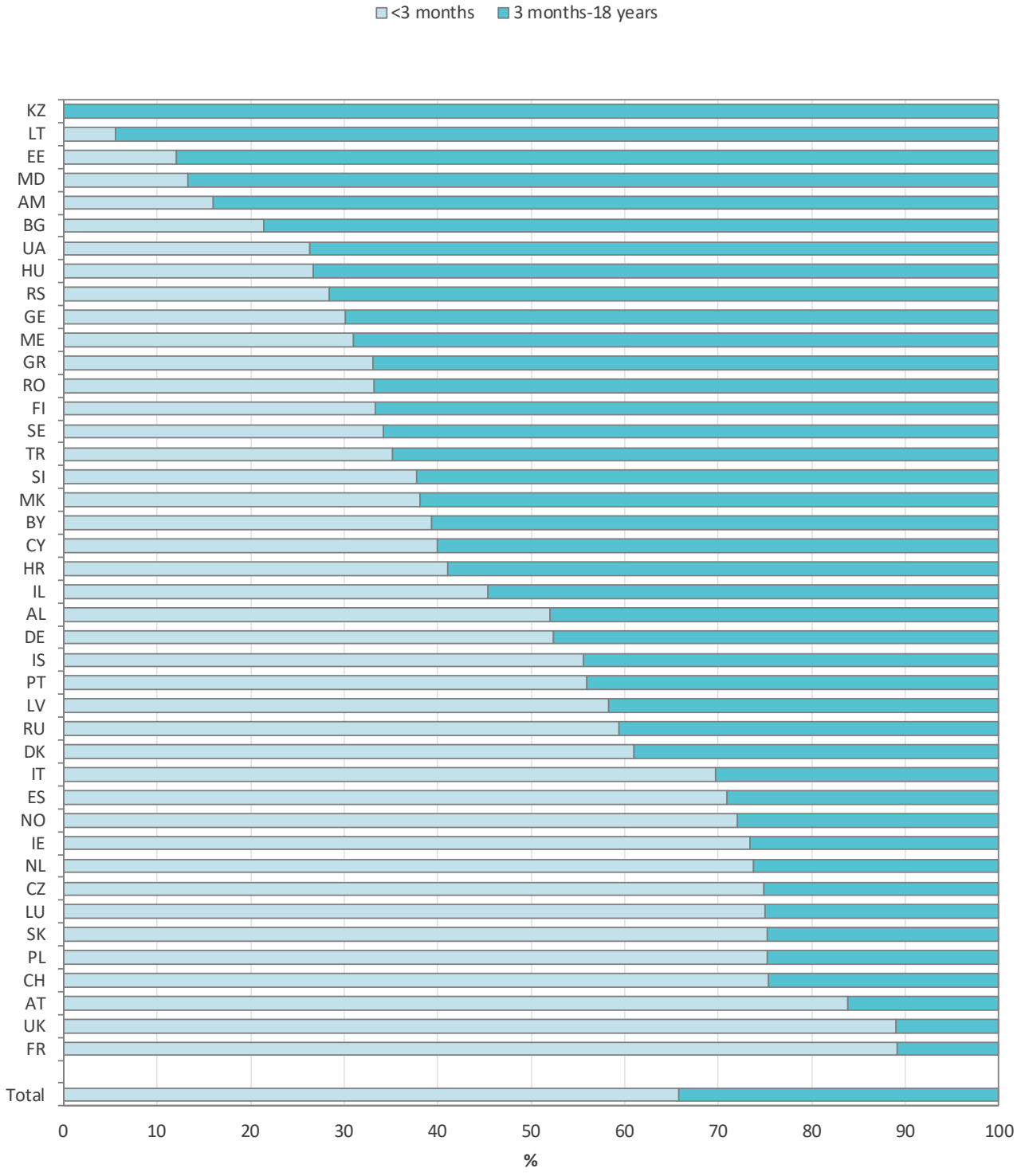
For Austria, Finland, Greece and Switzerland the information on age at diagnosis is missing for more than 10% of the people with CF.

For each country the vertical borders of the box are the first and third quartiles, the dash (vertical black line crossing the box) is the median, the black dot is the mean. The whiskers that are the minimum and the maximum values are not shown because the maximum values are very high for some countries and this would have shrunk the boxes to the left side of the graph.

2. Diagnosis

Figure 2.3 After the implementation of newborn screening programmes, age at diagnosis has shifted to the first 3 months of life in many countries.

Proportion of children with CF diagnosed at younger than 3 months, between 3 months and 18 years, by country and overall. All children and adolescents with CF seen in 2023.

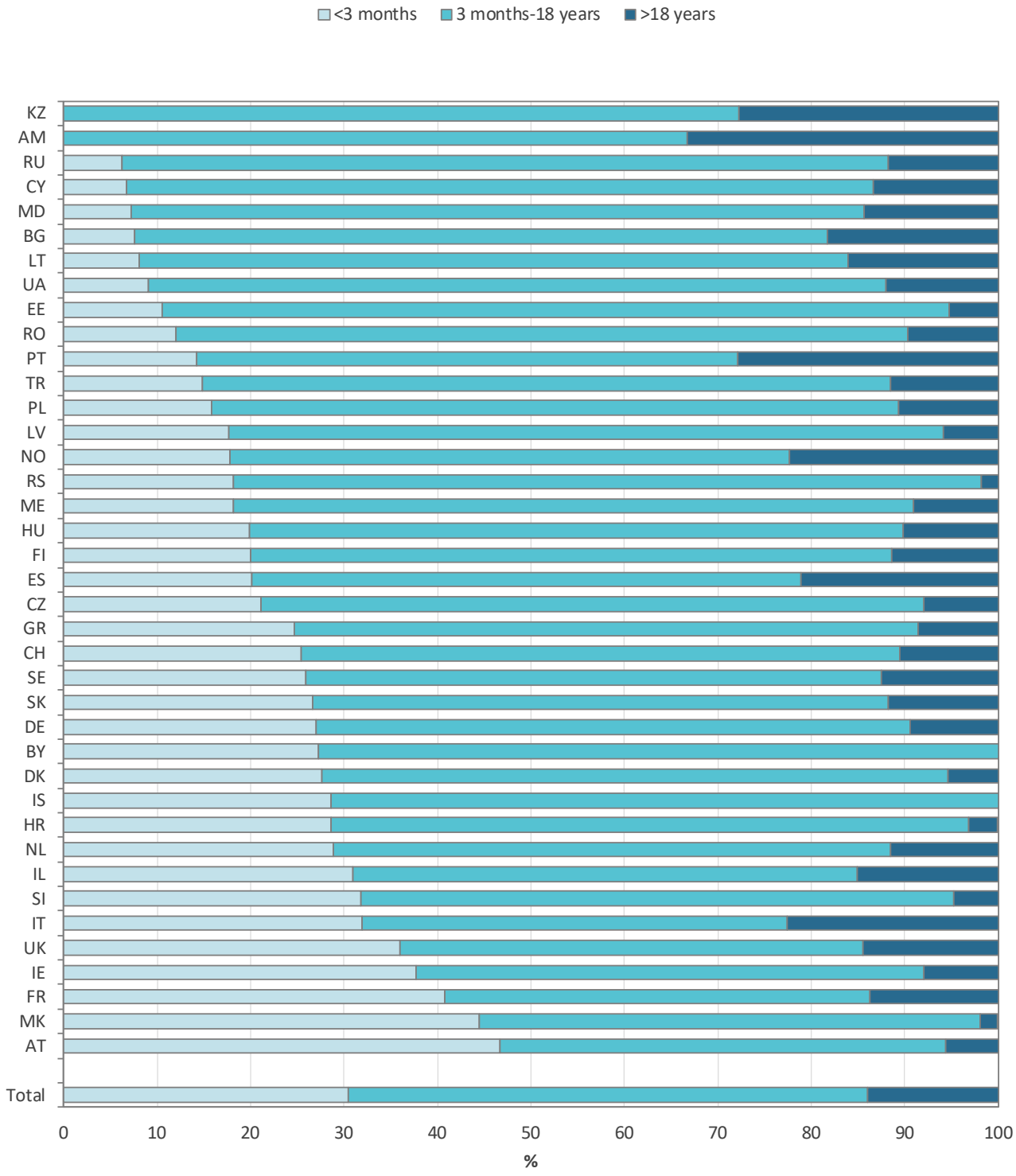


Note: For Cyprus, Greece, Lithuania and the Slovak Republic the information on age at diagnosis is missing for more than 10% of the children.

2. Diagnosis

Figure 2.4 Age at diagnosis has shifted to the first 3 months of life in many countries.

Proportion of adults with CF diagnosed at younger than 3 months, between 3 months and 18 years, and older than 18 years, by country and overall. All adults with CF seen in 2023.



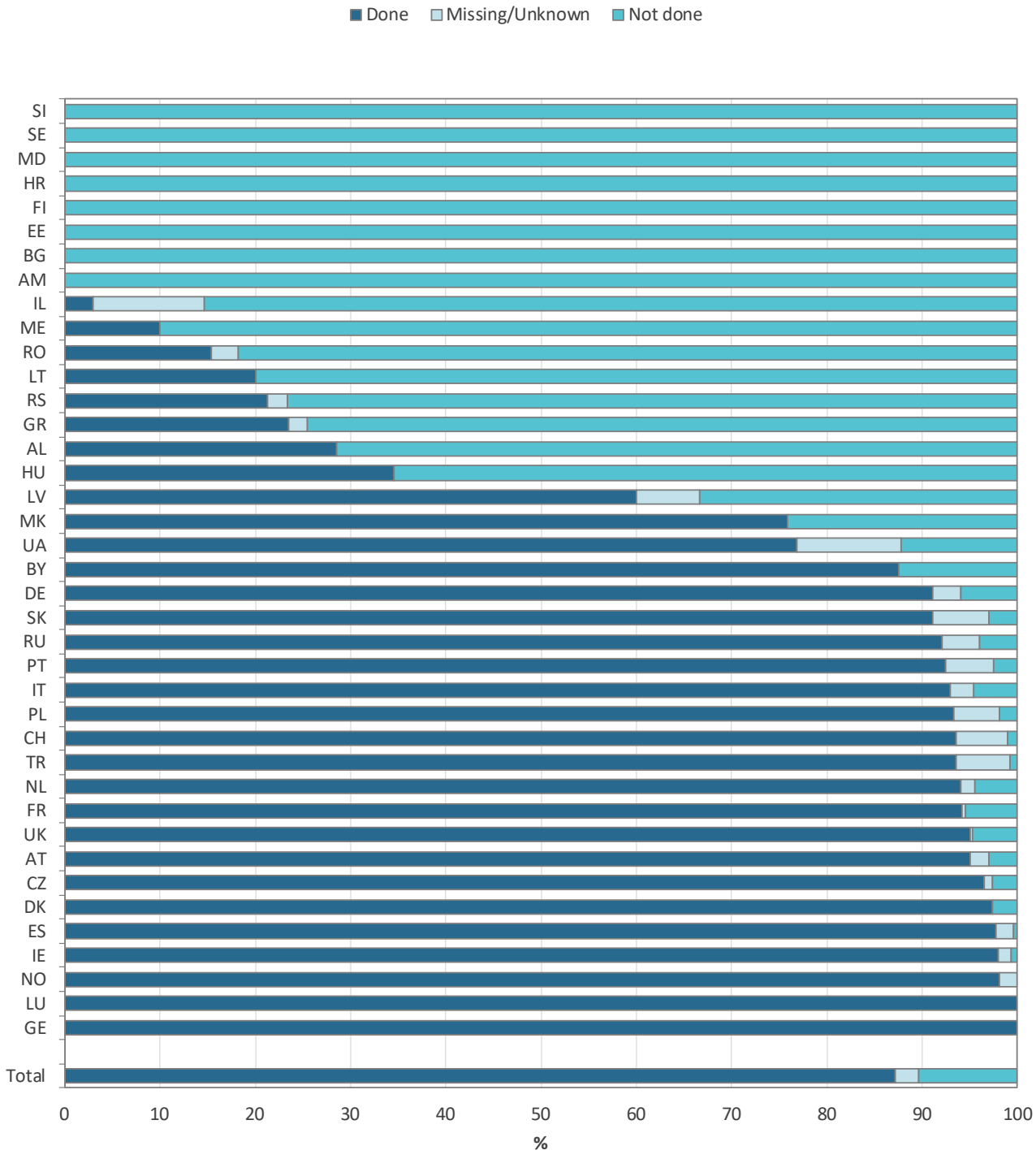
Note: Albania, Georgia and Luxembourg have <5 adults seen in 2023 and are excluded from the table, but the individuals are included in the total number.

Note: For Austria, Finland, Greece and Switzerland the information on age at diagnosis is missing for more than 10% of the people with CF.

2. Diagnosis

Figure 2.5 More than 85% of young children with CF are diagnosed through newborn screening.

Proportion of children with CF who underwent neonatal screening, by country and overall. Children 5 years old or younger seen in 2023.



Note: Cyprus and Iceland have <5 children 5 years old or younger seen in 2023 and are excluded from the graph.

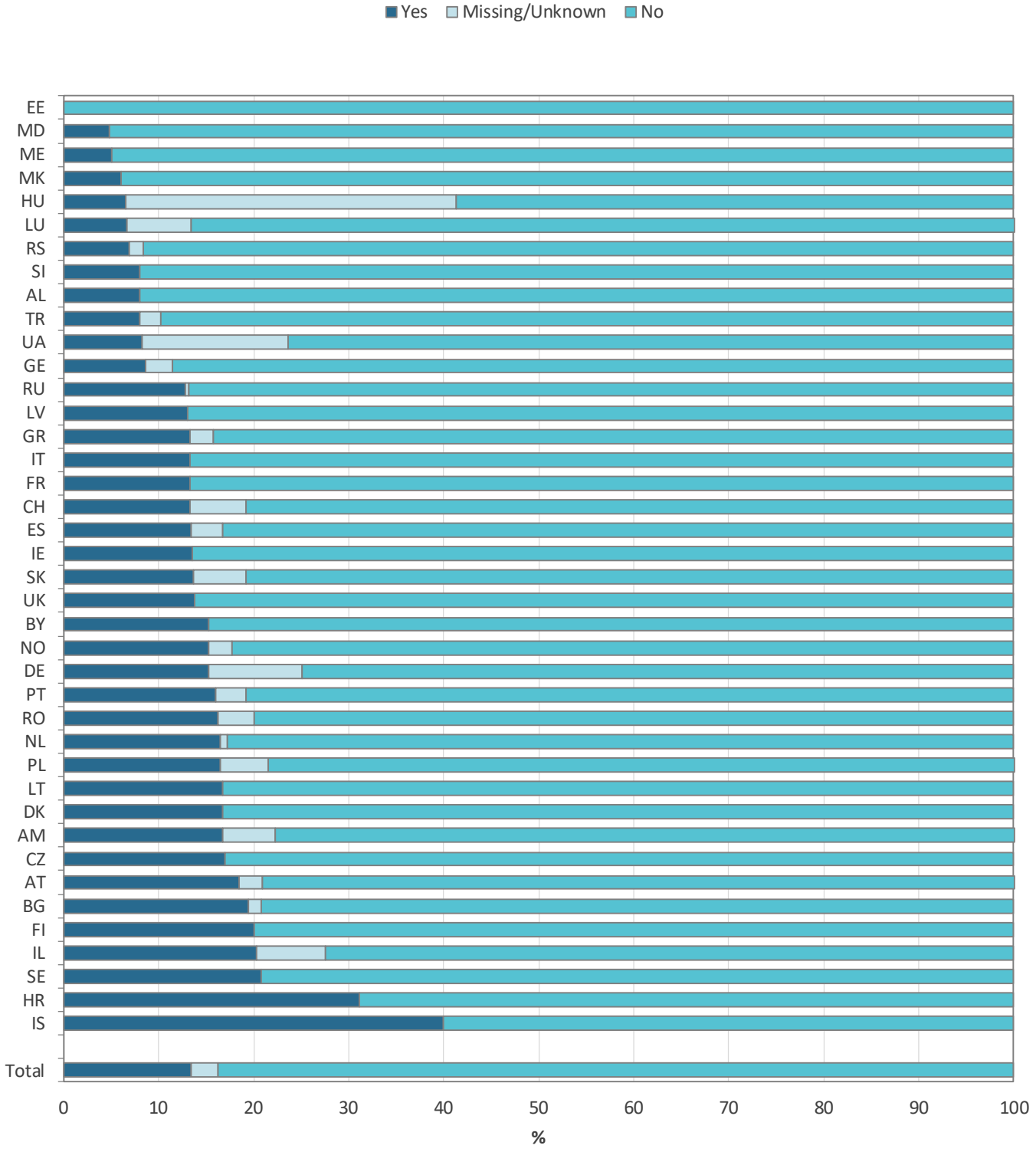
Note: For Israel and Ukraine the information on neonatal screening is missing for more than 10% of the children ≤5 years old.

Note: For France and the United Kingdom positive answers ("neonatal screening done") are reported only when neonatal screening is one of the factors that led to CF diagnosis.

2. Diagnosis

Figure 2.6 Meconium ileus at birth is not rare and may be the first symptom of CF detected in newborns.

People with CF with meconium ileus, by country and overall. People with CF aged 10 years or younger.



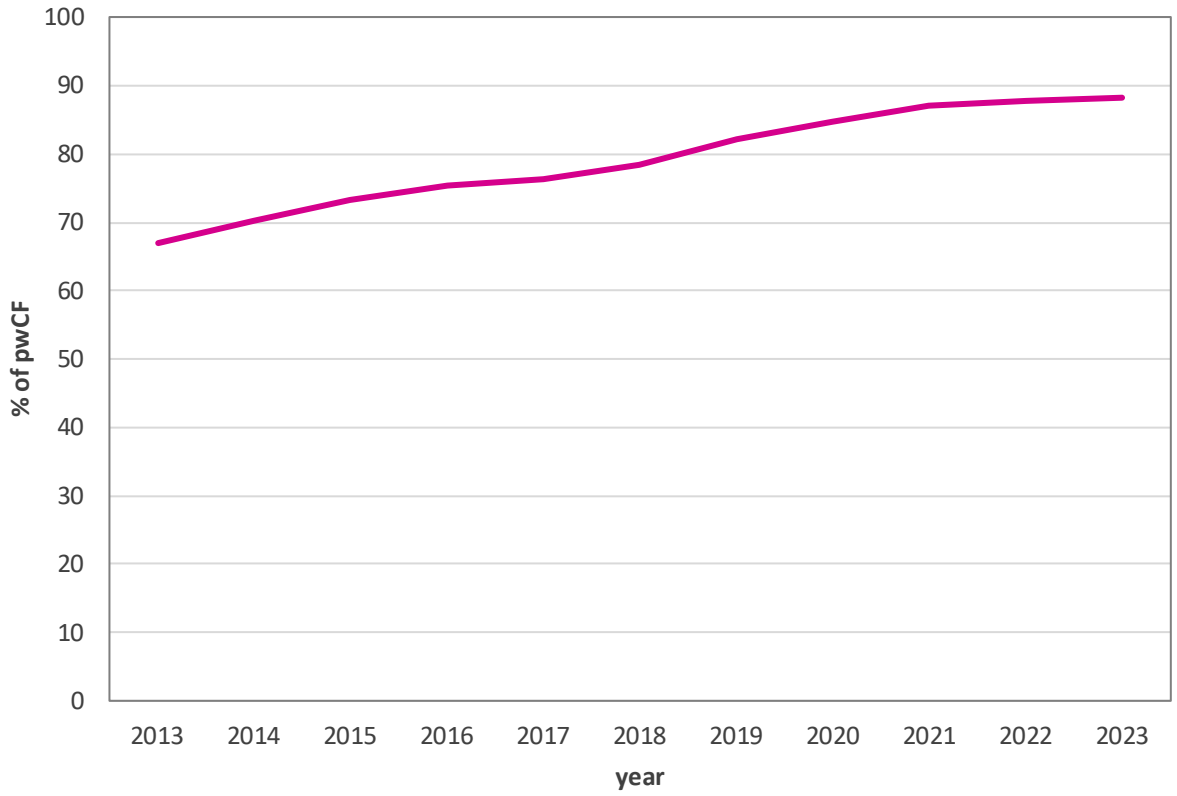
Note: Cyprus has <5 children 10 years old or younger seen in 2023 and are excluded from the graph.

Note: For Hungary and Ukraine, the information on meconium ileus is missing for more than 10% of the children ≤10 years old.

2. Diagnosis

Figure 2.7 In the last decade the proportion of children diagnosed with CF through newborn screening has increased to almost 90% throughout Europe.

Neonatal screening in children with CF aged 5 years or younger in the years from 2013 to 2023.



In this graph data over time are presented using cross sectional data per year of children with a confirmed CF diagnosis. Children with CF who are alive, deceased, or not seen during the year of follow-up were included and those who were lost to follow-up and/or had transplanted lung/s and/or liver were excluded.

3. Genetics

Cystic fibrosis is caused by pathogenic variants, also called mutations, of the *CFTR* (cystic fibrosis transmembrane conductance regulator) gene. At least one variant on each copy, or allele, of the gene is inherited from both the mother and the father. If the variants on both alleles are the same, the person is said to be homozygous for this variant; if these are two different variants, the person is considered to be heterozygous.
We supplied the countries with a list of the 1600 most common variants based on the Cystic Fibrosis Mutation Database (CFTR1). If an individual with CF has a variant that is not present in this database, the name of the variant is provided as free text. During the data cleaning process, genotypes not on our list were checked for obvious misspellings or alternative names and, if identified as a known variant, they were renamed. There are different naming conventions for variants; in this report we use the original variant name (legacy name), if it exists, since more than 90% of the variants in the CFTR1 database have this nomenclature.
If DNA analysis to look for variants on the *CFTR* gene has not been carried out we asked the countries to report "Not done". If DNA analysis has been done, but only one or no variants were found, we asked the countries to report this as "Unknown" for the unidentified variants.
How DNA testing is carried out differs from country to country; some use standard kits to test only a limited number of the most common variants (e.g. 28), while other countries perform DNA-analyses of the whole gene until the variant is detected.

Please note that, although not presented in this report, information on complex alleles is also captured and available.

3. Genetics

Table 3.1 Genotyping has been done for more than 99% of people with CF in the ECFSPR; for 96% of those tested, at least two variants have been identified.

Proportion of people with CF with DNA analysis and the result, by country and overall. People with CF seen in 2023.

Country	Genotyping				Among genotyping done			
	Not done		Done		At least one variant unknown		Two variants identified	
	Number	%	Number	%	Number	%	Number	%
Albania	0	0.0	53	100.0	1	1.9	52	98.1
Armenia	0	0.0	32	100.0	4	12.5	28	87.5
Austria	0	0.0	855	100.0	43	5.0	812	95.0
Belarus	0	0.0	152	100.0	48	31.6	104	68.4
Bulgaria	0	0.0	230	100.0	11	4.8	219	95.2
Croatia	0	0.0	144	100.0	4	2.8	140	97.2
Cyprus	0	0.0	22	100.0	2	9.1	20	90.9
Czech Republic	1	0.1	712	99.9	8	1.1	704	98.9
Denmark	0	0.0	578	100.0	0	0.0	578	100.0
Estonia	0	0.0	45	100.0	0	0.0	45	100.0
Finland	0	0.0	76	100.0	1	1.3	75	98.7
France	0	0.0	7329	100.0	121	1.7	7208	98.4
Georgia	3	5.6	51	94.4	1	2.0	50	98.0
Germany	3	0.0	7176	100.0	117	1.6	7059	98.4
Greece	3	0.5	618	99.5	22	3.6	596	96.4
Hungary	0	0.0	507	100.0	41	8.1	466	91.9
Iceland	0	0.0	16	100.0	0	0.0	16	100.0
Ireland	2	0.2	1357	99.9	20	1.5	1337	98.5
Israel	1	0.2	540	99.8	39	7.2	501	92.8
Italy	2	0.0	6125	100.0	147	2.4	5978	97.6
Kazakhstan	0	0.0	27	100.0	4	14.8	23	85.2
Latvia	0	0.0	53	100.0	0	0.0	53	100.0
Lithuania	0	0.0	46	100.0	2	4.4	44	95.7
Luxembourg	0	0.0	25	100.0	0	0.0	25	100.0
Rep of Moldova	0	0.0	46	100.0	6	13.0	40	87.0
Montenegro	0	0.0	40	100.0	1	2.5	39	97.5
The Netherlands	6	0.4	1615	99.6	14	0.9	1601	99.1
North Macedonia	0	0.0	139	100.0	0	0.0	139	100.0
Norway	0	0.0	382	100.0	3	0.8	379	99.2
Poland	0	0.0	1609	100.0	36	2.2	1573	97.8
Portugal	1	0.3	396	99.8	5	1.3	391	98.7
Romania	0	0.0	396	100.0	45	11.4	351	88.6
Russian Federation	90	2.9	3032	97.1	238	7.9	2794	92.2
Serbia	0	0.0	172	100.0	12	7.0	160	93.0
Slovak Republic	0	0.0	280	100.0	24	8.6	256	91.4
Slovenia	1	0.9	117	99.2	3	2.6	114	97.4
Spain	1	0.0	2541	100.0	110	4.3	2431	95.7
Sweden	0	0.0	785	100.0	1	0.1	784	99.9
Switzerland	5	0.5	992	99.5	35	3.5	957	96.5
Türkiye	65	2.4	2681	97.6	286	10.7	2395	89.3
Ukraine	2	0.5	430	99.5	91	21.2	339	78.8
United Kingdom	20	0.2	10324	99.8	268	2.6	10056	97.4
Total	206	0.4	52746	99.6	1814	3.4	50932	96.6

3. Genetics

Figure 3.1 Almost 98% of CFTR variants have been identified.

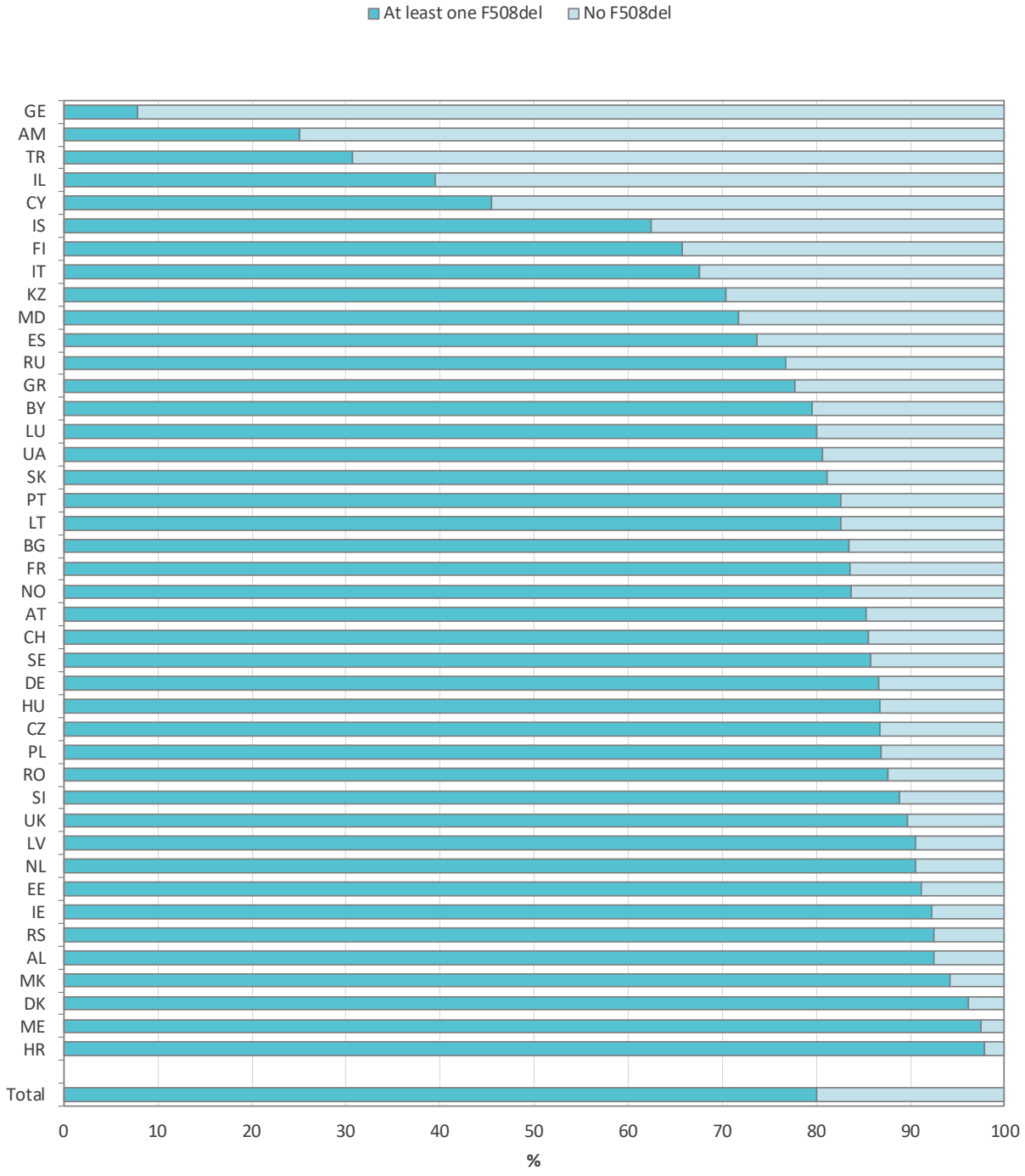
Proportion of variants identified and not identified, by country and overall. Only people with CF for whom DNA analysis has been done.

This graph shows the percentage of variants identified (dark turquoise) and variants not identified (light turquoise) through DNA analysis by country and overall. The number of variants not identified on one of the 2 alleles varies greatly from country to country; this is partly due to different approaches to DNA testing. Overall, 2.1 % of variants remain unidentified after DNA analysis, leaving 3.4% of the people with CF with at least one unidentified variant.

3. Genetics

Figure 3.2 The prevalence of the F508del variant varies considerably between the countries in Europe; this has a major impact on CFTR modulator eligibility.

Prevalence of the F508del variant in people with CF, by country and overall. All people with CF seen in 2023.



F508del is the name of the most commonly occurring CFTR variant in the world. People with CF who carry two F508del variants are often described as having "classic CF" but other variant combinations can cause similar degree of disease. Only people with CF for whom the genotype is known have been included in this graph. "Unknown" variants have been classified as "other", since F508del is included in all genotyping kits and would have been identified. Please note that the genotype grouping in this graph does not reflect the severity of the disease in the countries.

3. Genetics

Table 3.2 Allelic frequencies of most common (≥0.50%) variants in the ECFSPR database.

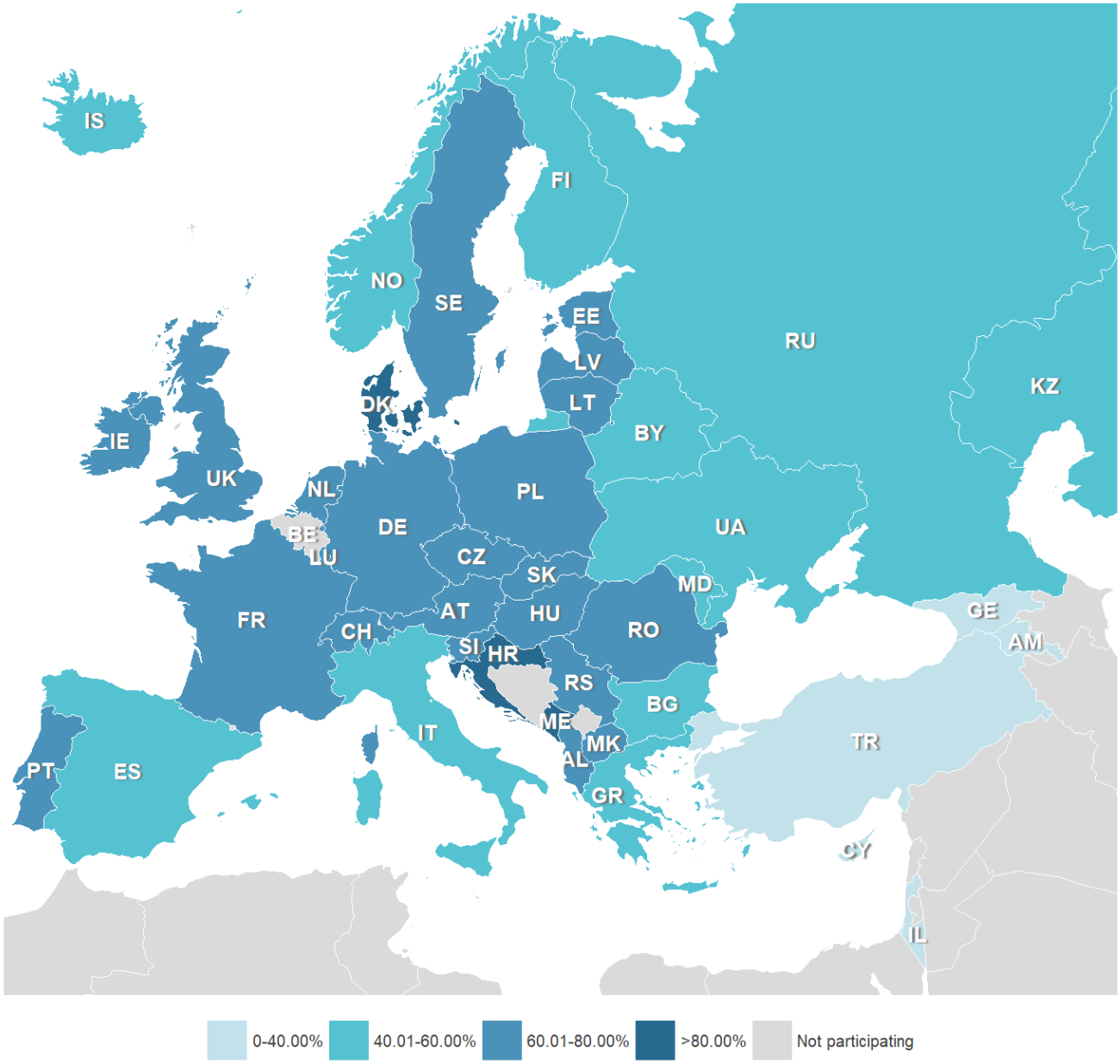
Variant name	Number of alleles with the variant	Percentage of those tested	Country with highest allele frequency for the variant
F508del	62754	59.49	Denmark (81.9%)
G542X	2906	2.75	Montenegro (8.7%)
N1303K	2300	2.18	Iceland (40.6%)
G551D	1284	1.22	Ireland (8.3%)
2789+5G->A	1167	1.11	Türkiye (3.2%)
3849+10kbC->T	1150	1.09	Lithuania (7.6%)
W1282X	1146	1.09	Israel (22.1%)
CFTRdele2,3	1137	1.08	Belarus (8.5%)
R117H	994	0.94	United Kingdom (3.0%)
1717-1G->A	855	0.81	Switzerland (2.3%)
R553X	836	0.79	Lithuania (6.5%)
D1152H	780	0.74	Israel (6.0%)
2183AA->G	736	0.70	Armenia (12.5%)
621+1G->T	701	0.66	Greece (6.9%)
R347P	656	0.62	Luxembourg (6.0%)
G85E	612	0.58	Israel (2.6%)
1677delTA	611	0.58	Georgia (52.0%)
2184insA	534	0.51	Belarus (7.6%)
3272-26A->G	528	0.50	Portugal (2.3%)

This table presents the most common variants found in the ECFSPR database. The last column indicates in which country this particular variant is most frequent. F508del is, by far, the most common variant.

3. Genetics

Figure 3.3 The allelic frequency of F508del variant is higher in Central Europe

Geographical distribution of the F508del variant. All people with CF seen in 2023.

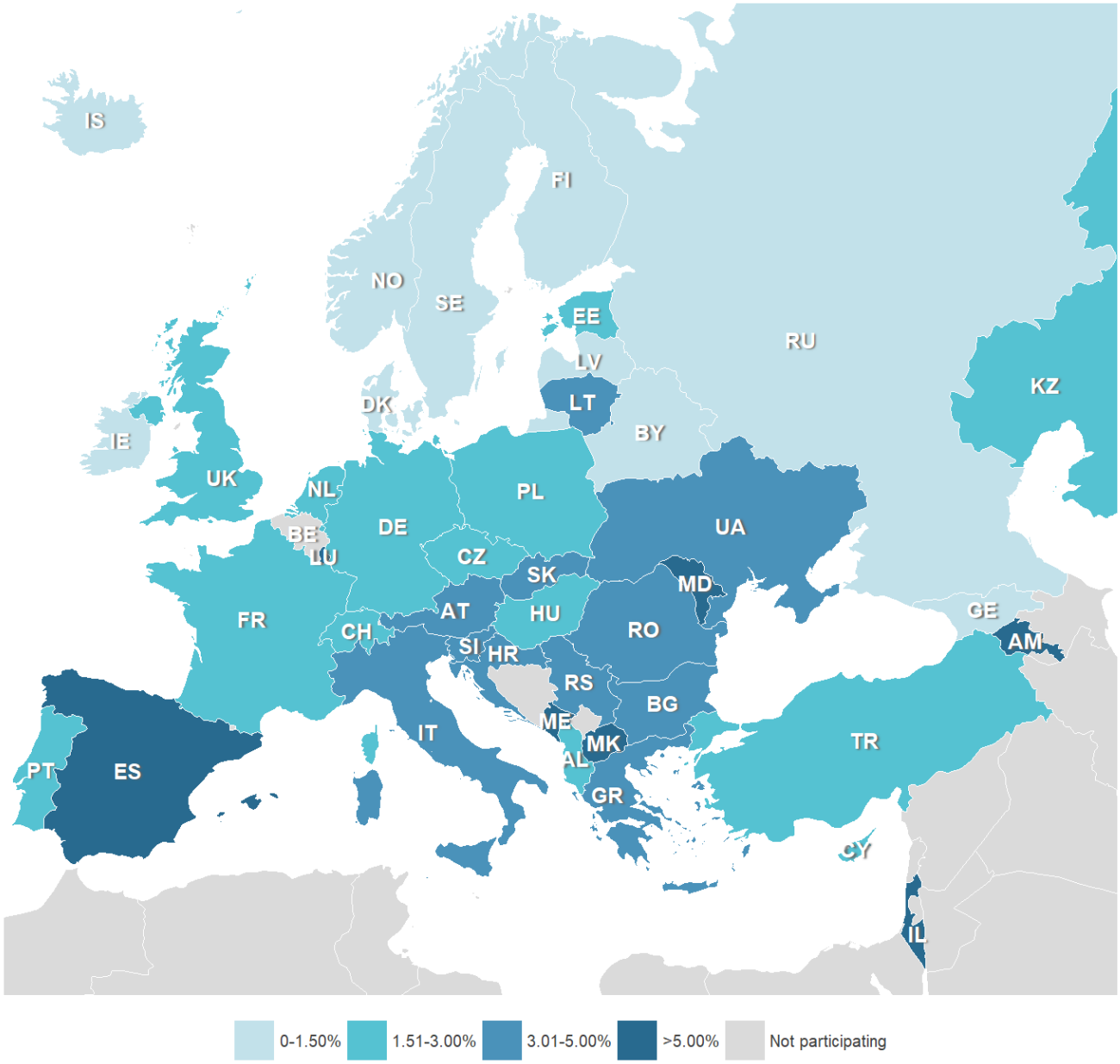


F508del is the most common variant in all countries; the highest frequency occurs in Denmark (81.9%) followed by Montenegro (81.2%) and Croatia (80.9%).

3. Genetics

Figure 3.4 The G542X variant is more prevalent in Southern Europe.

Geographical distribution of the G542X variant. All people with CF seen in 2023.

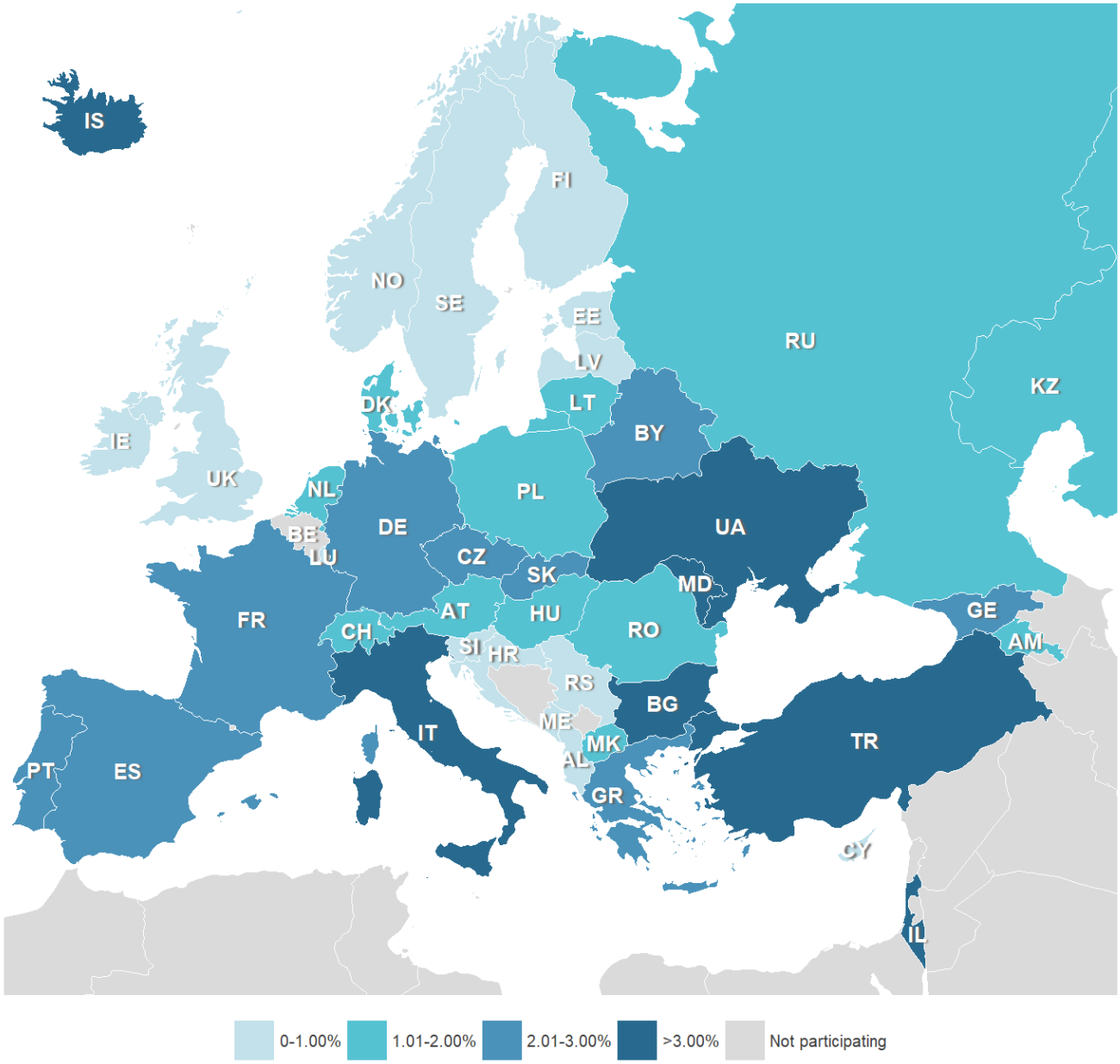


The *G542X* variant is most frequent in Southern Europe, with the highest allele frequency in Montenegro (8.7%), whereas it is very rarely found in Ireland, the Scandinavian countries or the Russian Federation.

3. Genetics

Figure 3.5 The N1303K variant is more prevalent in Southern and Eastern Europe

Geographical distribution of the N1303K variant. All people with CF seen in 2023.

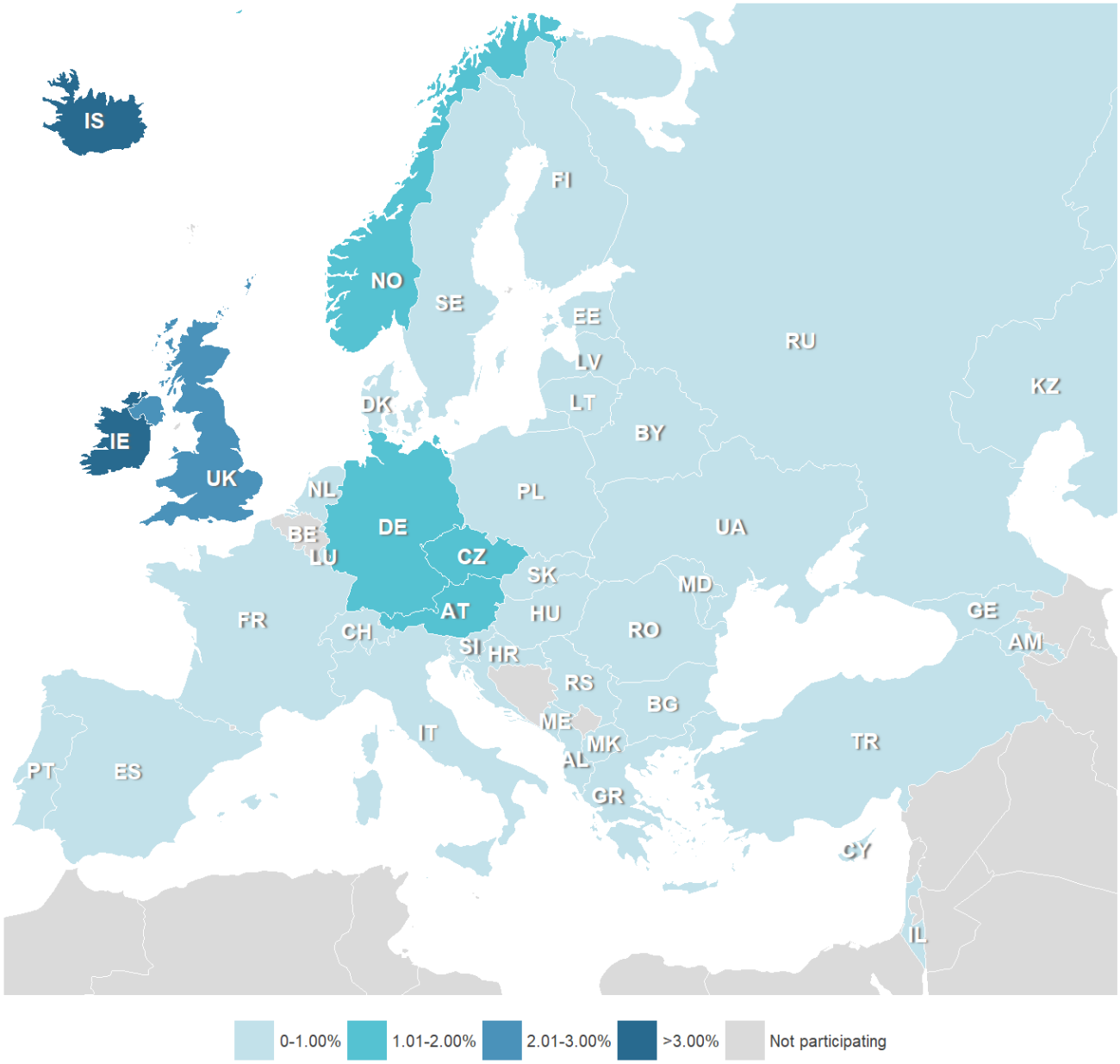


The *N1303K* variant is most frequent in Iceland (40.6%). This is an exception in Northern Europe where it is otherwise rare; it is much more frequent in the countries of Southern and Eastern Europe.

3. Genetics

Figure 3.6 The G551D variant is more frequent in the northern and central regions of Europe.

Geographical distribution of the G551D variant. All people with CF seen in 2023.

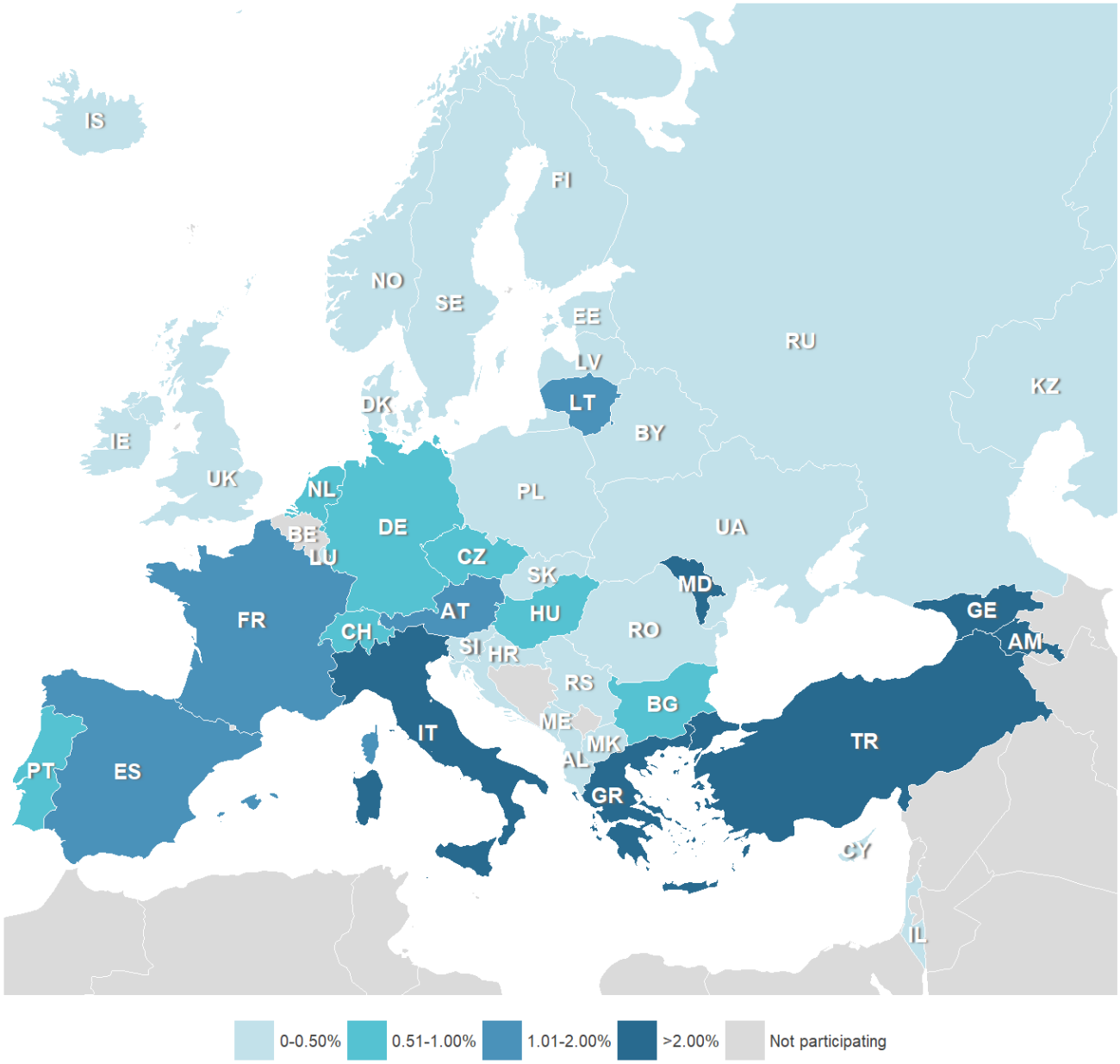


The *G551D* variant is most frequent in Ireland (8.3%) and in northern and central regions of Europe whereas it is rare in the east and south of Europe.

3. Genetics

Figure 3.7 The 2789+5G->A variant is more common in Türkiye and the southern regions of Europe.

Geographical distribution of the 2789+5G->A variant. All people with CF seen in 2023.



The *2789+5G->A* variant is most frequent in Türkiye (3.2%) and in Southern Europe, and less common in Eastern Europe.

4. Lung function

Lung function, or lung capacity, is measured by spirometry, a test which calculates how much air can be forced out of the lungs in one breath. The measurement is called FEV_1 (Forced Expiratory $Volume_1$) and it is measured in litres, but the lung capacity is normally expressed as a percentage of the expected (or predicted) value (FEV_1% of predicted or FEV1pp). The predicted value is determined from healthy individuals of the same age, sex, ethnicity and height called the reference population. A FEV_1% of predicted of 100 means that the lung function measurement is equal to the mean lung function measurement of people of the same age, sex, ethnicity and height of the healthy reference population.

To calculate the FEV_1% of predicted for this report we used the Global Lung Function Initiative equations and the ethnicity categories described by Quanjer PH et al. (for the full reference refer to Appendix 4).

Spirometry testing requires a certain amount of coordination and usually cannot be performed reliably and consistently until a child is about five to six years of age; we therefore computed FEV_1% of predicted values only for people with CF who are aged 6 or older.

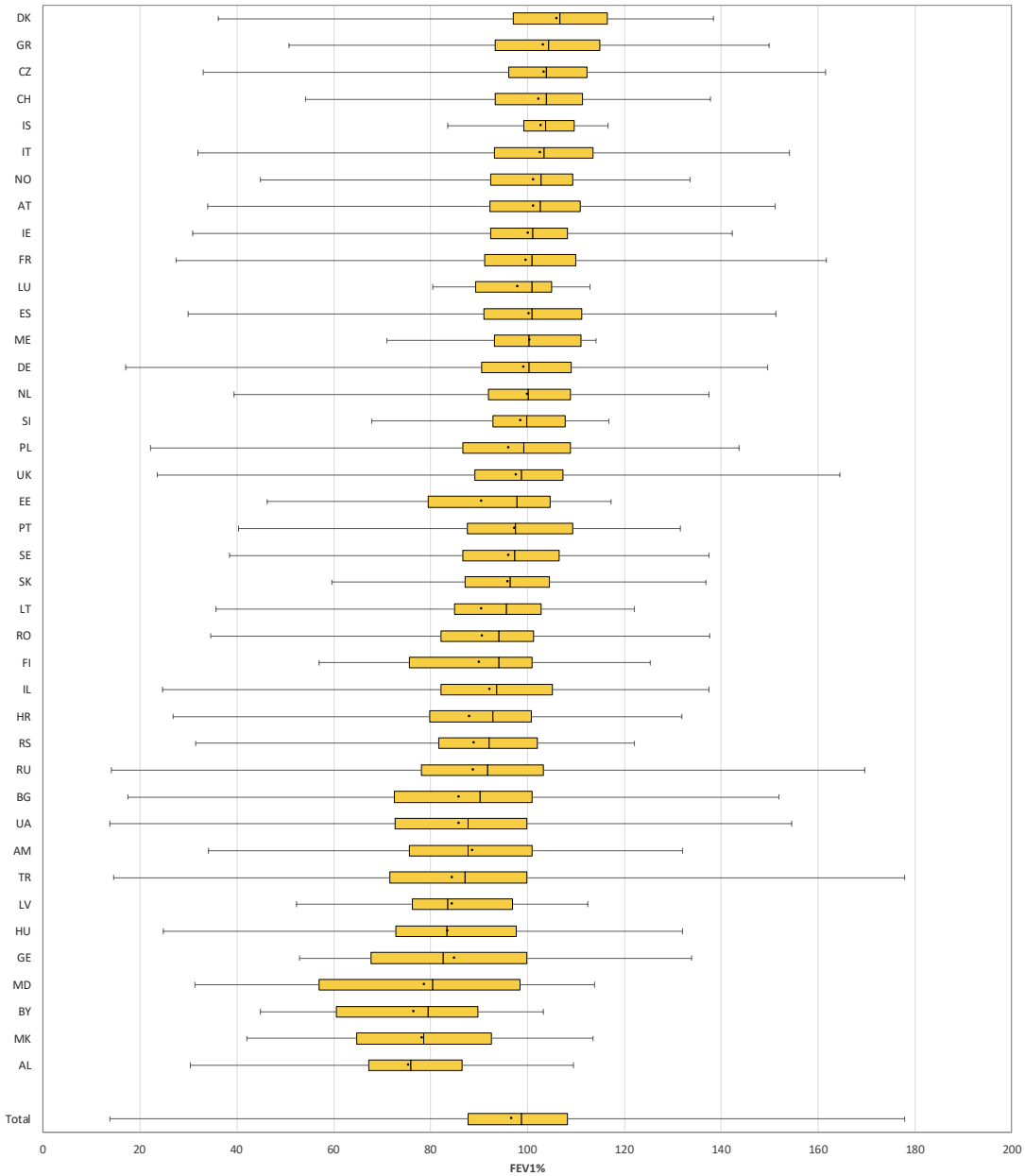
We asked the countries to report the FEV_1 measured in litres from the best FEV_1% of predicted (computed at the centres during spirometry testing) recorded throughout the year.

We excluded people from the analyses of FEV_1 who have had one or more lung transplants since their lung function does not reflect the severity of their CF lung disease. Moreover, we also excluded people with CF who have had a liver transplant since follow-up data for them is sometimes missing.

4. Lung function

Figure 4.1 The median FEV1% of predicted is >80% for children and adolescents with CF (between 6 and 17 years of age) in nearly all countries in Europe.

FEV1% of predicted: boxplot by country. Children and adolescents with CF aged 6-17 years who have never had a lung/liver transplant, seen in 2023 (table A4.1, Appendix 1).



Note: Cyprus and Kazakhstan have <5 individuals aged 6-17 years at the date of FEV_1 measurement and are excluded from the graph.

Finland and Sweden report the best FEV1pp in the 12 months prior to the annual review which means, in some cases, that the value can be from the previous calendar year.

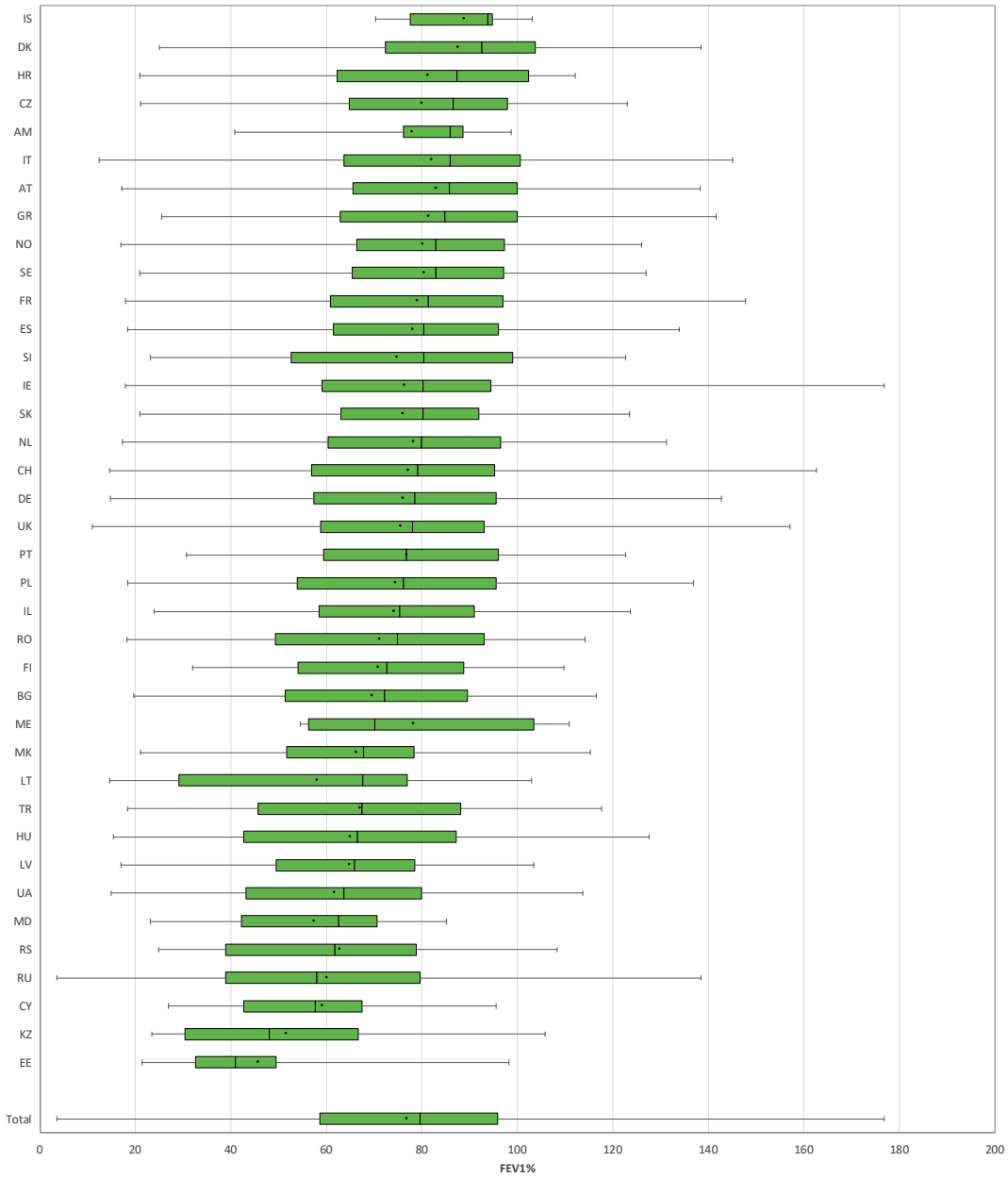
The UK reports the best FEV1pp from the period between annual reviews which means, in some cases, that the value can be from the previous calendar year.

This boxplot is a graphic representation of the FEV_1 in children and adolescents with CF, expressed as a % of predicted, detailed in table A4.1 (Appendix 1). For each country, the vertical borders of the box are the first and third quartiles, the dash (vertical black line crossing the yellow box) is the median, the black dot is the mean, and the whiskers (vertical lines with a T-shaped end) are the minimum and the maximum.

4. Lung function

Figure 4.2 Median FEV1% of predicted for adults with CF varies between 59% and 96% depending on the country.

FEV1% of predicted: boxplot by country. Adults with CF who have never had a transplant, seen in 2023 (table A4.2, Appendix 1).



Note: Albania, Belarus, Georgia and Luxembourg have <5 adults with FEV_1 measurement and are excluded from the table but the individuals are included in the total number.

Finland and Sweden report the best FEV1pp in the 12 months prior to the annual review which means, in some cases, that the value can be from the previous calendar year.

The UK reports the best FEV1pp from the period between annual reviews which means, in some cases, that the value can be from the previous calendar year.

This boxplot is a graphic representation of the FEV_1 in adults, expressed as the % of predicted detailed in table A4.2 (Appendix 1). For each country the vertical borders of the box are the first and third quartiles, the dash (vertical black line crossing the green box) is the median, the black dot is the mean, and the whiskers (vertical lines with a T-shaped end) are the minimum and the maximum.

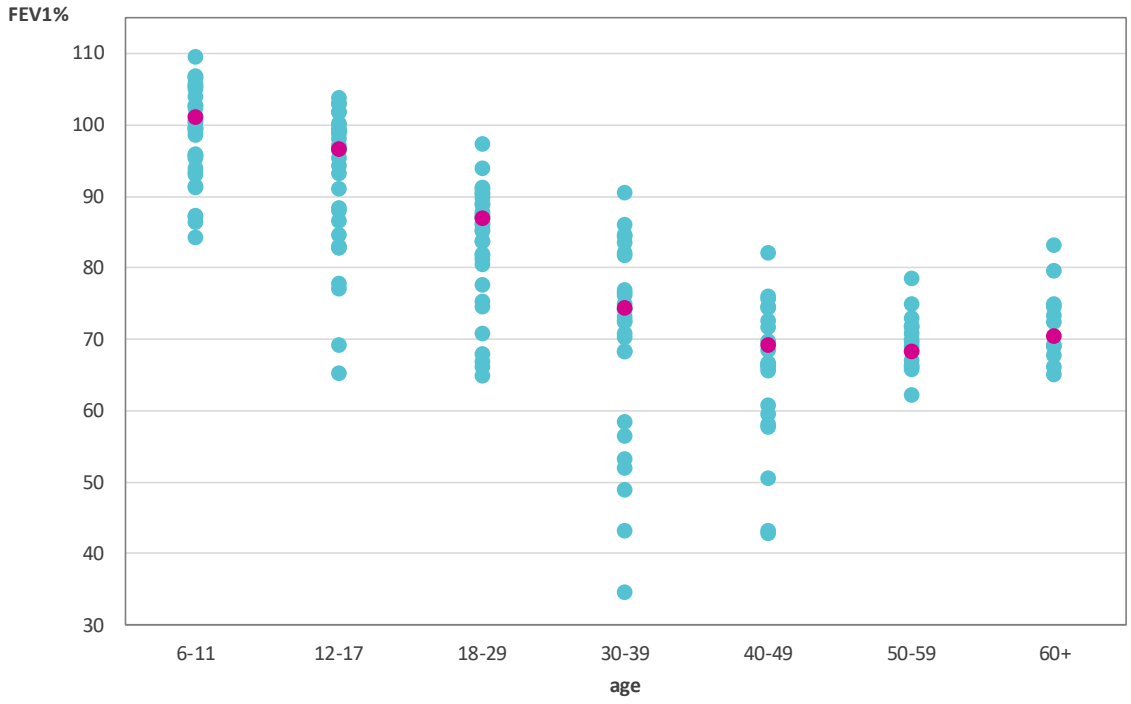
4. Lung function

Table 4.1 FEV_1% of predicted: descriptive statistics by age group (people with CF aged 6 years or older) who have never had a transplant.

Age at FEV_1 measurement	Number	Number of missing	Mean (average FEV1% value)	Min (lowest FEV1% value)	25th pctl (25% of the pwCF have a FEV1% lower than the value)	Median (half the pwCF have a FEV1% lower than the value)	75th pctl (75% of the pwCF have a FEV1% lower than the value)	Max (highest FEV1% value)
6-11	7612	953	99.8	14.1	90.7	101.1	110.4	177.8
12-17	7828	517	94.0	13.8	84.8	96.5	106.2	169.7
18-29	11675	539	83.0	3.5	68.4	86.9	100.0	176.8
30-39	6849	275	73.5	15.4	54.9	74.4	91.8	157.2
40-49	3590	150	69.8	11.0	50.7	69.2	88.6	162.6
50-59	1733	88	68.8	14.7	49.9	68.2	86.6	145.1
60+	763	30	70.5	20.0	51.2	70.4	88.8	146.2

This table shows FEV1% of predicted by age group for the total dataset. The median values reported in this table are shown as pink dots in Figure 4.3.

Figure 4.3 Lung function declines between the third and fifth decade of life but stabilises in older people with CF.
Median FEV_1% of predicted by age group and by country.



Note: We excluded from the graph those age groups where the number of individuals was <10.

This graph shows the median (the value that separates the highest and lowest half) FEV_1% of predicted by age group. Each country median is represented by a dot (in turquoise) and the overall median is in pink. The general pattern shows that the FEV_1% of predicted slowly decreases until the age of 40-49 and then levels out. The people in the older age groups may have a less severe form of the disease and therefore live longer. There is considerable variation amongst the countries.

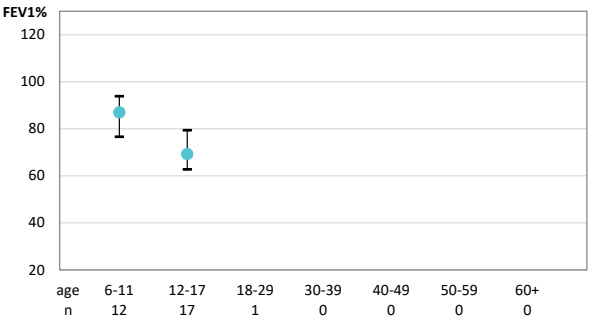
4. Lung function

Figure 4.4 Decline in lung function over time in adults with CF still poses a challenge in Europe.

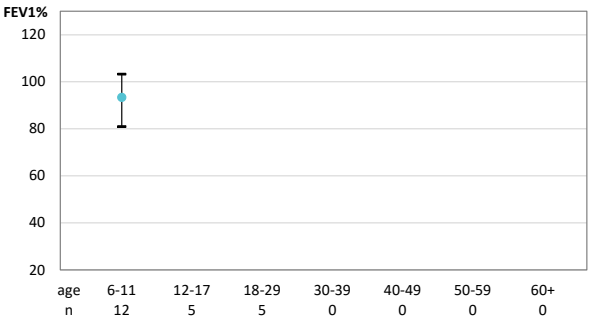
Quartiles of FEV1% of predicted by age group and by country. People with CF aged 6 years or older and who have never had a transplant.

The figures below show the FEV_1% of predicted in different age groups for each country. The dot shows the median, and the whiskers show the 25th and 75th percentiles (the median, the 25th percentile and the 75th percentile are collectively named "quartiles"). We did not calculate quartiles where the number of people with CF is <10 in a given age group, so there are no dots for those age groups (the number of people with CF in each age group is shown below the horizontal axis); we therefore excluded Cyprus, Estonia, Georgia, Iceland, Kazakhstan and Luxembourg from the graphs because none of the age groups had more than 10 people with CF.

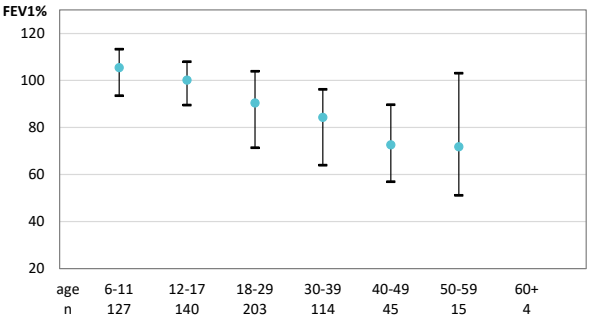
Quartiles of FEV_1%: Albania



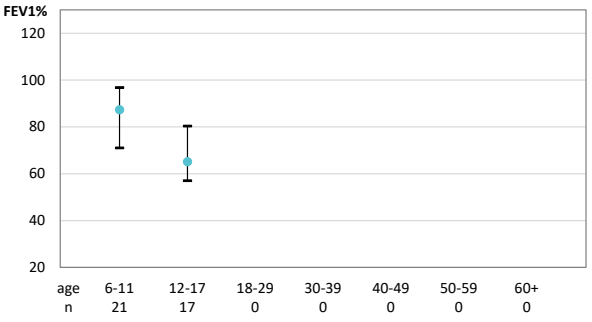
Quartiles of FEV_1%: Armenia



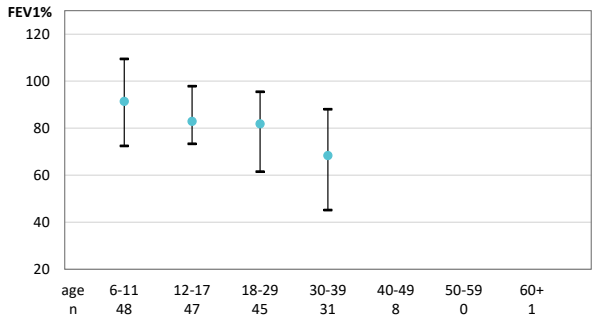
Quartiles of FEV_1%: Austria



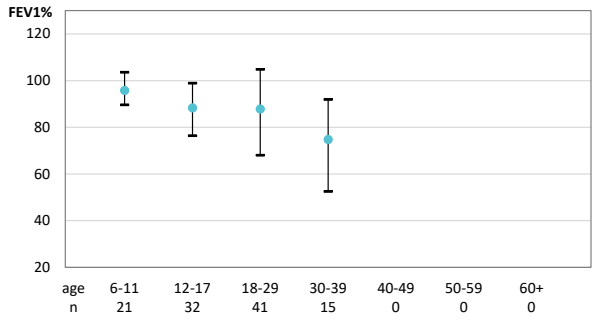
Quartiles of FEV_1%: Belarus



Quartiles of FEV_1%: Bulgaria



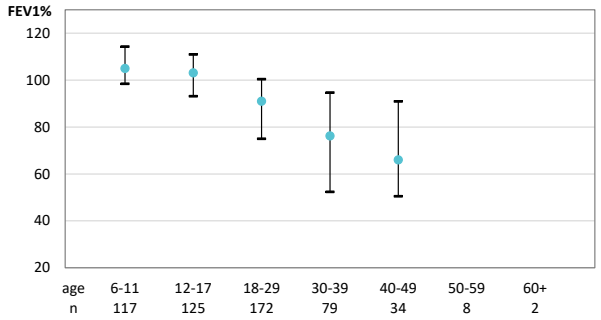
Quartiles of FEV_1%: Croatia



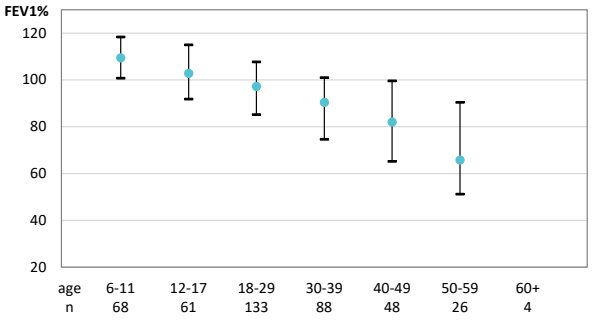
4. Lung function

[figure 4.4 continued]

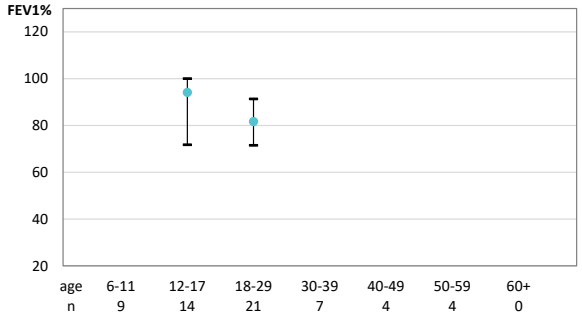
Quartiles of FEV_1%: Czech Republic



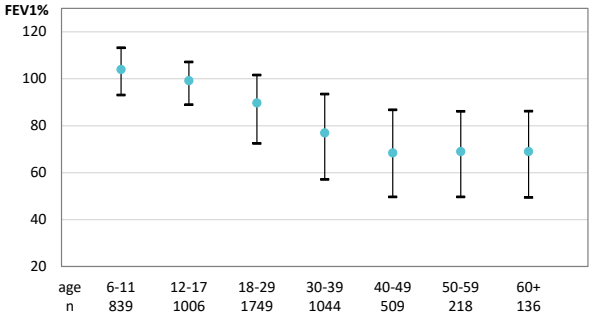
Quartiles of FEV_1%: Denmark



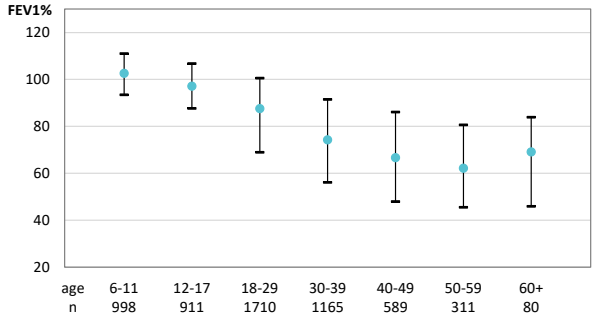
Quartiles of FEV_1%: Finland



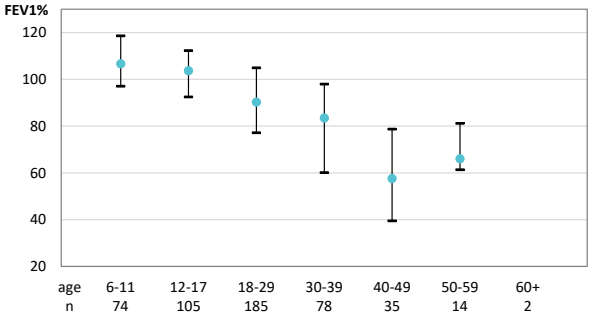
Quartiles of FEV_1%: France



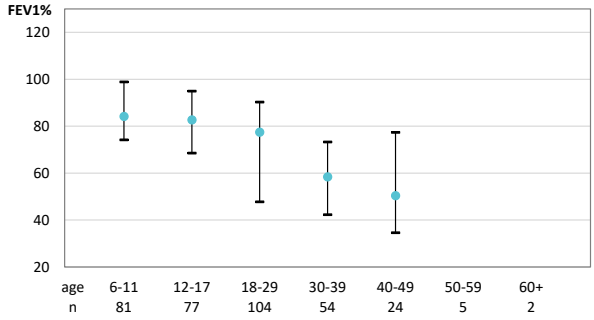
Quartiles of FEV_1%: Germany



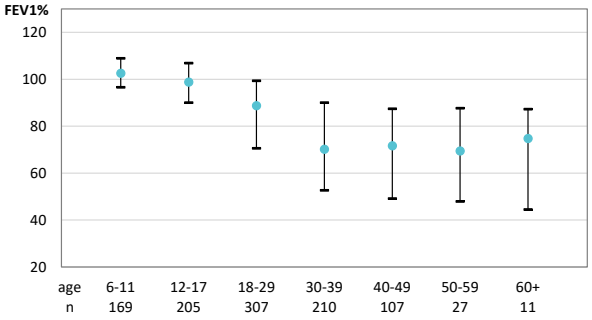
Quartiles of FEV_1%: Greece



Quartiles of FEV_1%: Hungary



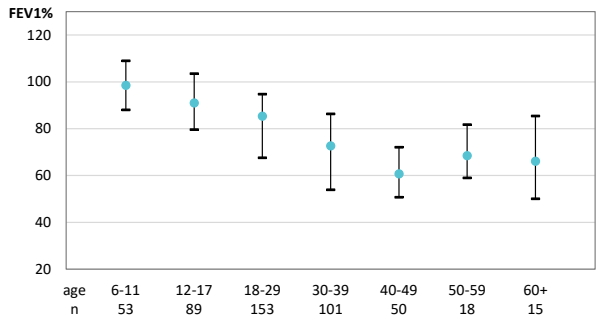
Quartiles of FEV_1%: Ireland



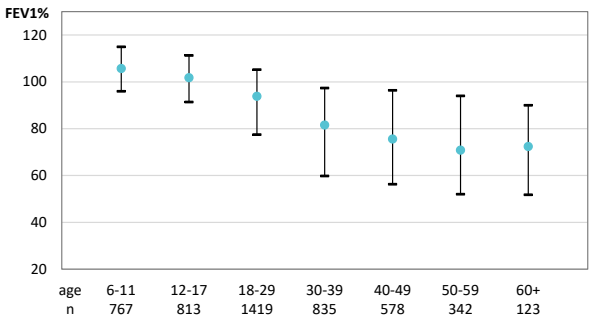
4. Lung function

[figure 4.4 continued]

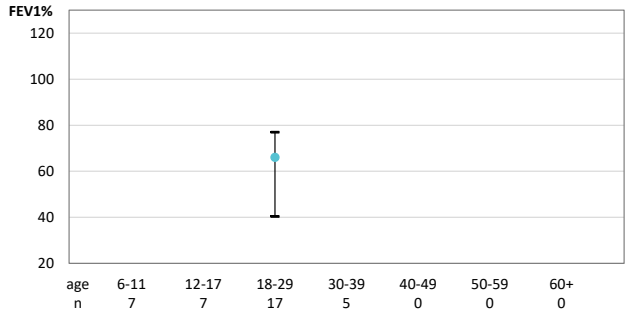
Quartiles of FEV_1%: Israel



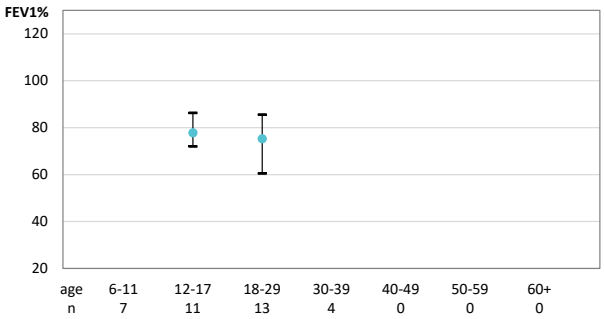
Quartiles of FEV_1%: Italy



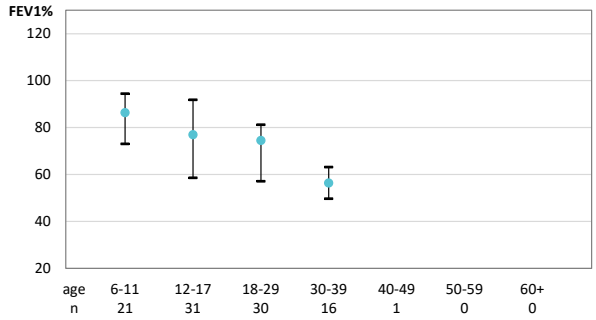
Quartiles of FEV_1%: Lithuania



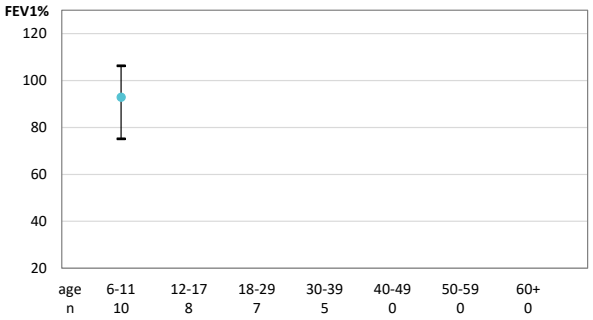
Quartiles of FEV_1%: Latvia



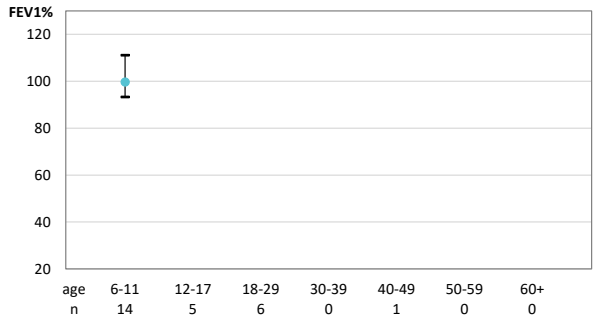
Quartiles of FEV_1%: North Macedonia



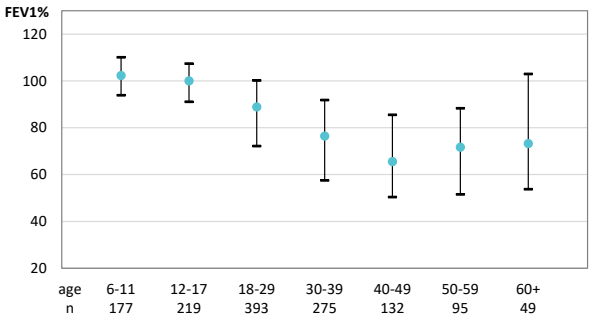
Quartiles of FEV_1%: Rep of Moldova



Quartiles of FEV_1%: Montenegro



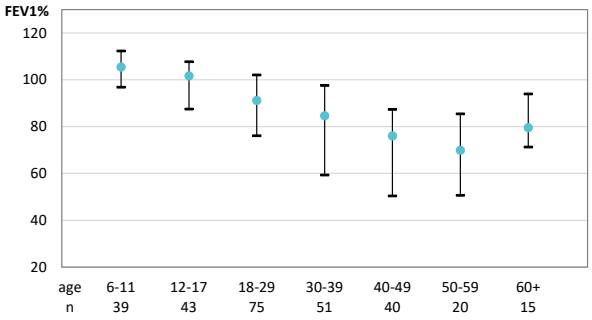
Quartiles of FEV_1%: The Netherlands



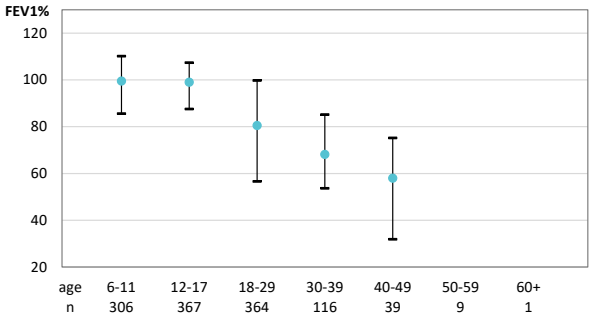
4. Lung function

[figure 4.4 continued]

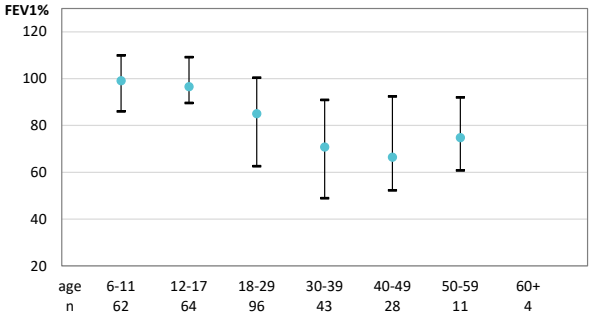
Quartiles of FEV_1%: Norway



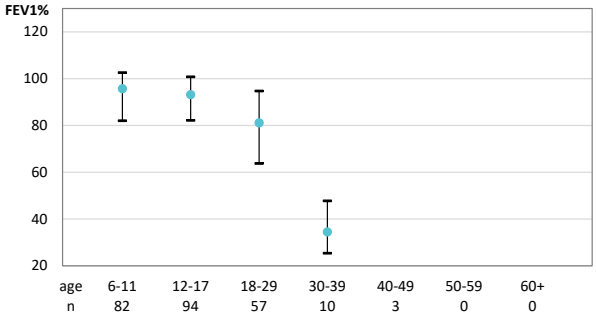
Quartiles of FEV_1%: Poland



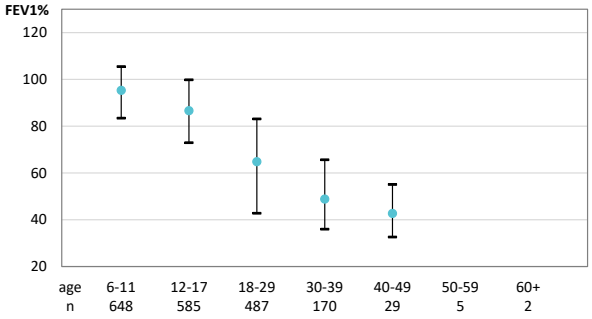
Quartiles of FEV_1%: Portugal



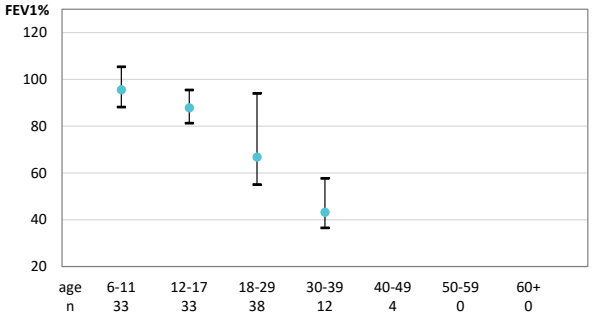
Quartiles of FEV_1%: Romania



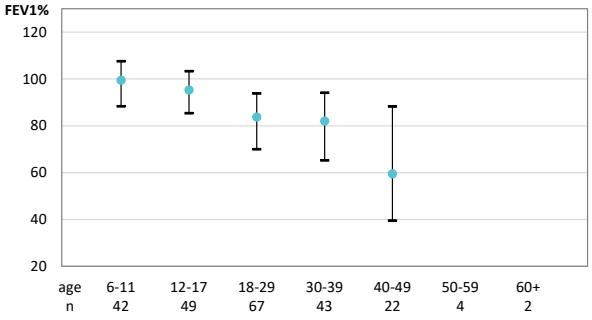
Quartiles of FEV1%: Russian Federation



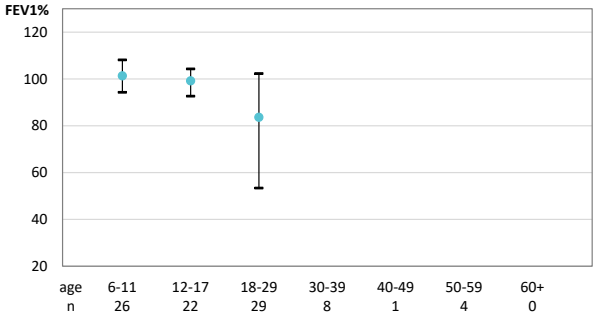
Quartiles of FEV_1%: Serbia



Quartiles of FEV_1%: Slovak Republic



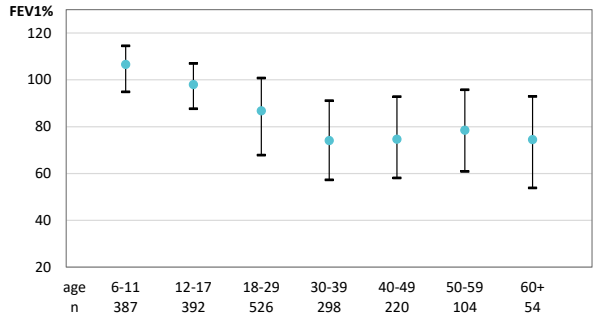
Quartiles of FEV_1%: Slovenia



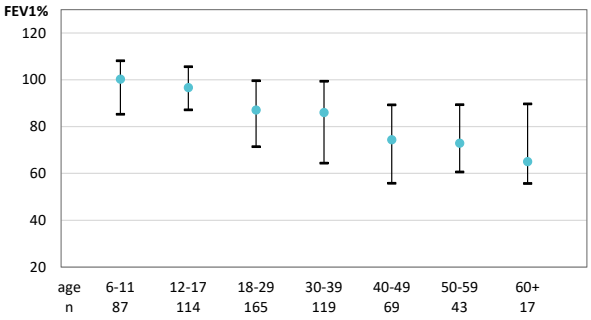
4. Lung function

[figure 4.4 continued]

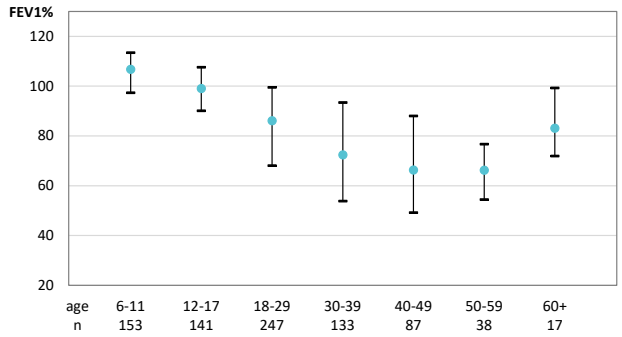
Quartiles of FEV_1%: Spain



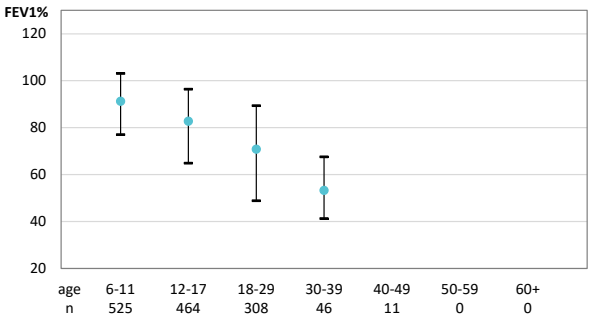
Quartiles of FEV_1%: Sweden



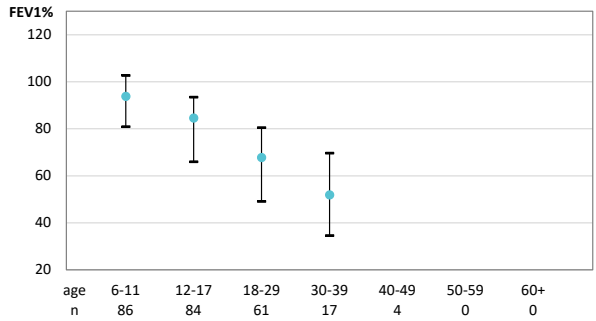
Quartiles of FEV_1%: Switzerland



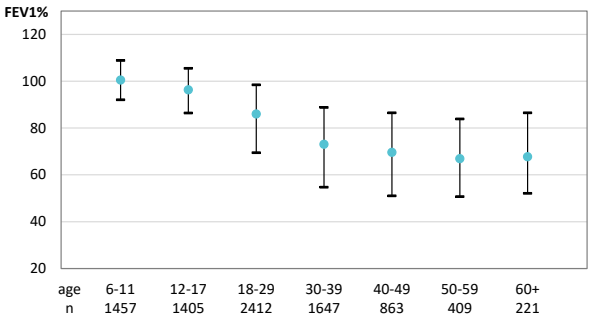
Quartiles of FEV_1%: Türkiye



Quartiles of FEV_1%: Ukraine



Quartiles of FEV_1%: United Kingdom



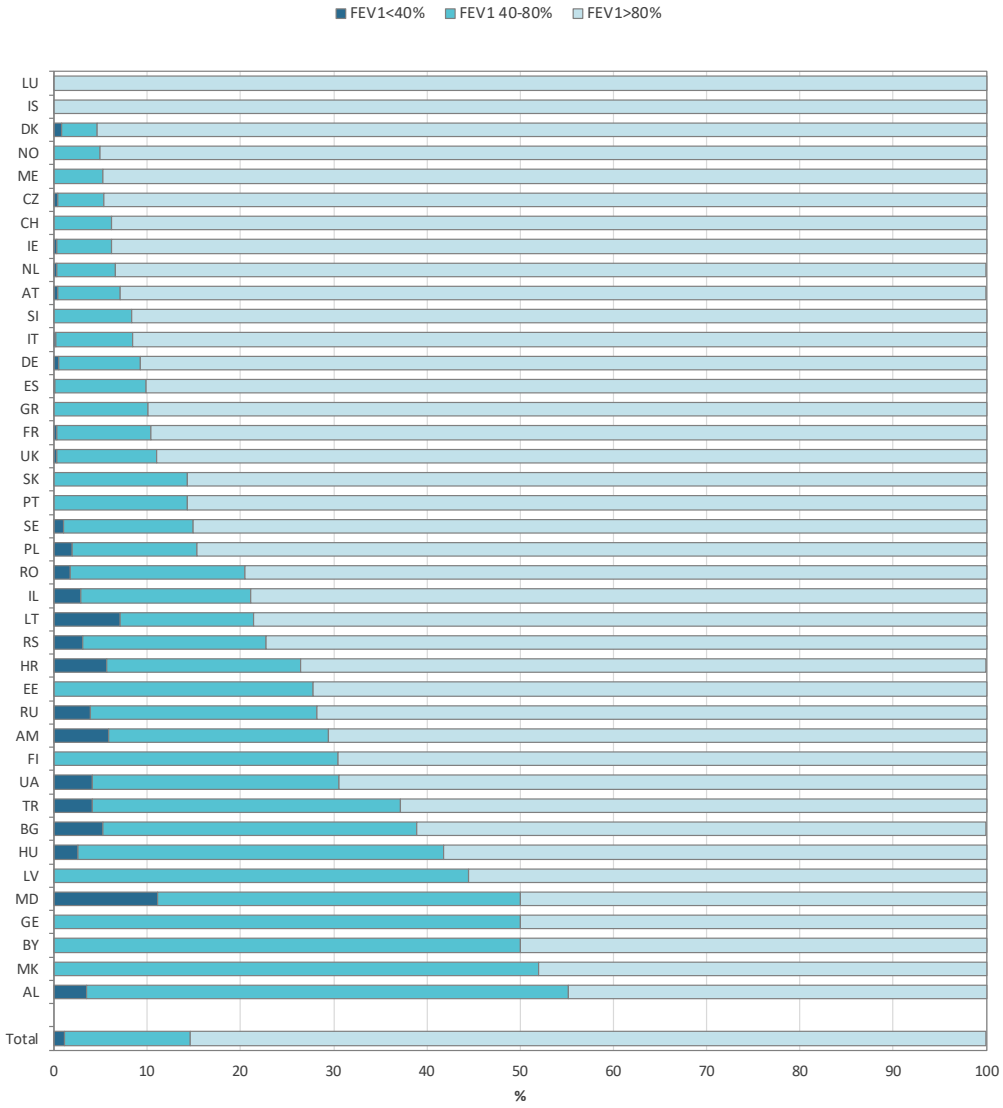
Note: Finland and Sweden report the best FEV1pp in the 12 months prior to the annual review which means, in some cases, that the value can be from the previous calendar year.

The UK reports the best FEV1pp from the period between annual reviews which means, in some cases, that the value can be from the previous calendar year.

4. Lung function

Figure 4.5 The majority of all children and adolescents with CF in Europe have a FEV_1 of >80% predicted.

FEV_1% of predicted according to severity group and age group, by country. Children and adolescents with CF aged 6–17 years who have never had a lung/liver transplant.



Note: Cyprus and Kazakhstan have <5 people with CF aged 6-17 years at FEV1 measurement and are excluded from the graph.

Finland and Sweden report the best FEV1pp in the 12 months prior to the annual review which means, in some cases, that the value can be from the previous calendar year.

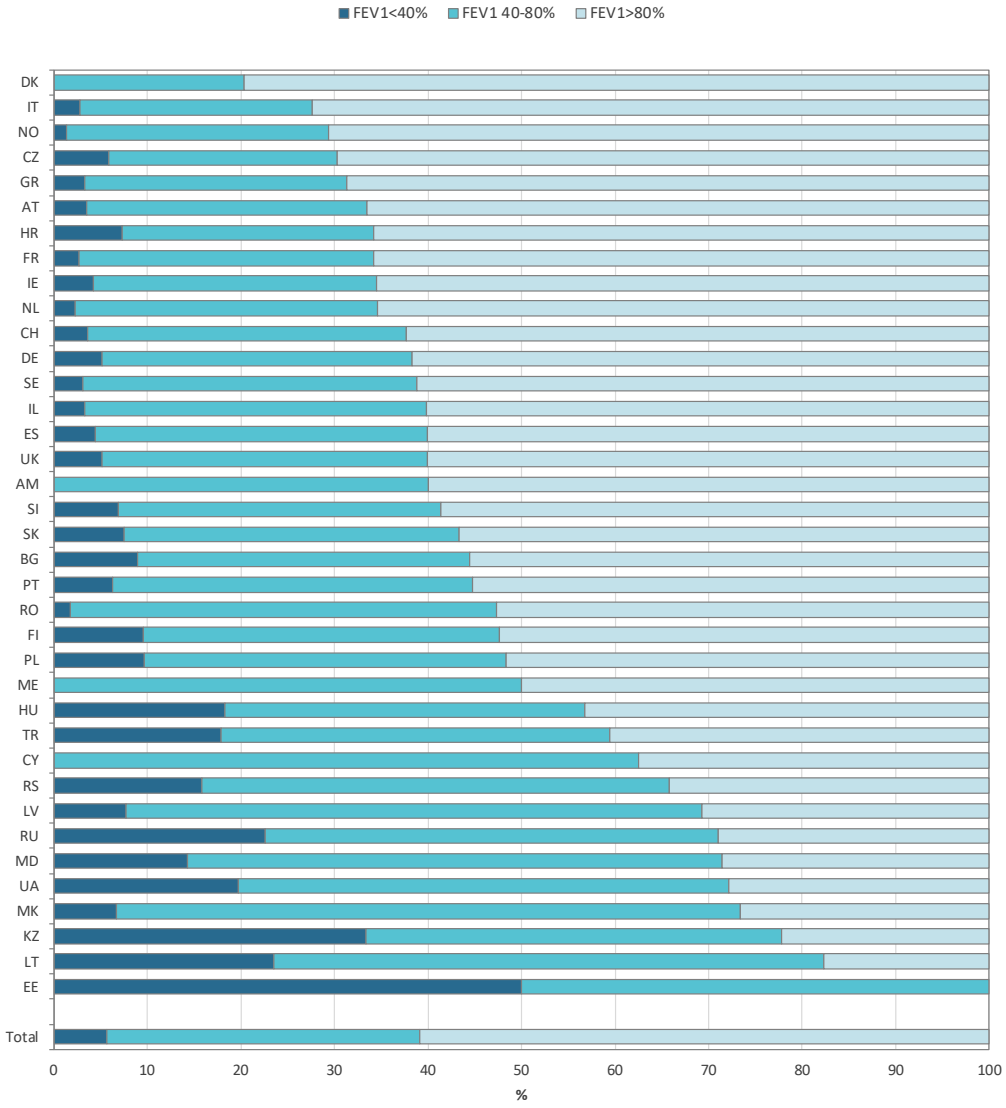
The UK reports the best FEV1pp from the period between annual reviews which means, in some cases, that the value can be from the previous calendar year.

Figures 4.5, 4.6 and 4.7 show the FEV_1% of predicted by severity group, by country and overall. People with CF with a FEV_1% of predicted higher than 80% are generally considered to have mild lung disease, those with a value between 80% and 40% to have moderate lung disease, and those with a FEV_1% of predicted lower than 40% to have severe lung disease. However, since a 10-year-old child with a lung function of 50% of predicted has considerably worse lung disease than a 50-year-old with the same value, and the age distribution is not the same in all countries, we have chosen to present children (Figure 4.5) and adults (Figure 4.6 and 4.7) separately.

4. Lung function

Figure 4.6 In the majority of countries, the proportion of young adults with CF with a FEV_1% of predicted below 40% is less than 10%.

FEV_1% of predicted according to severity group and age group, by country. Adults with CF aged 18 – 29 years who have never had a transplant.



Note: Albania, Belarus, Georgia, Iceland and Luxembourg have <5 people aged 18-29 years with FEV_1 measurement and are excluded from the graph.

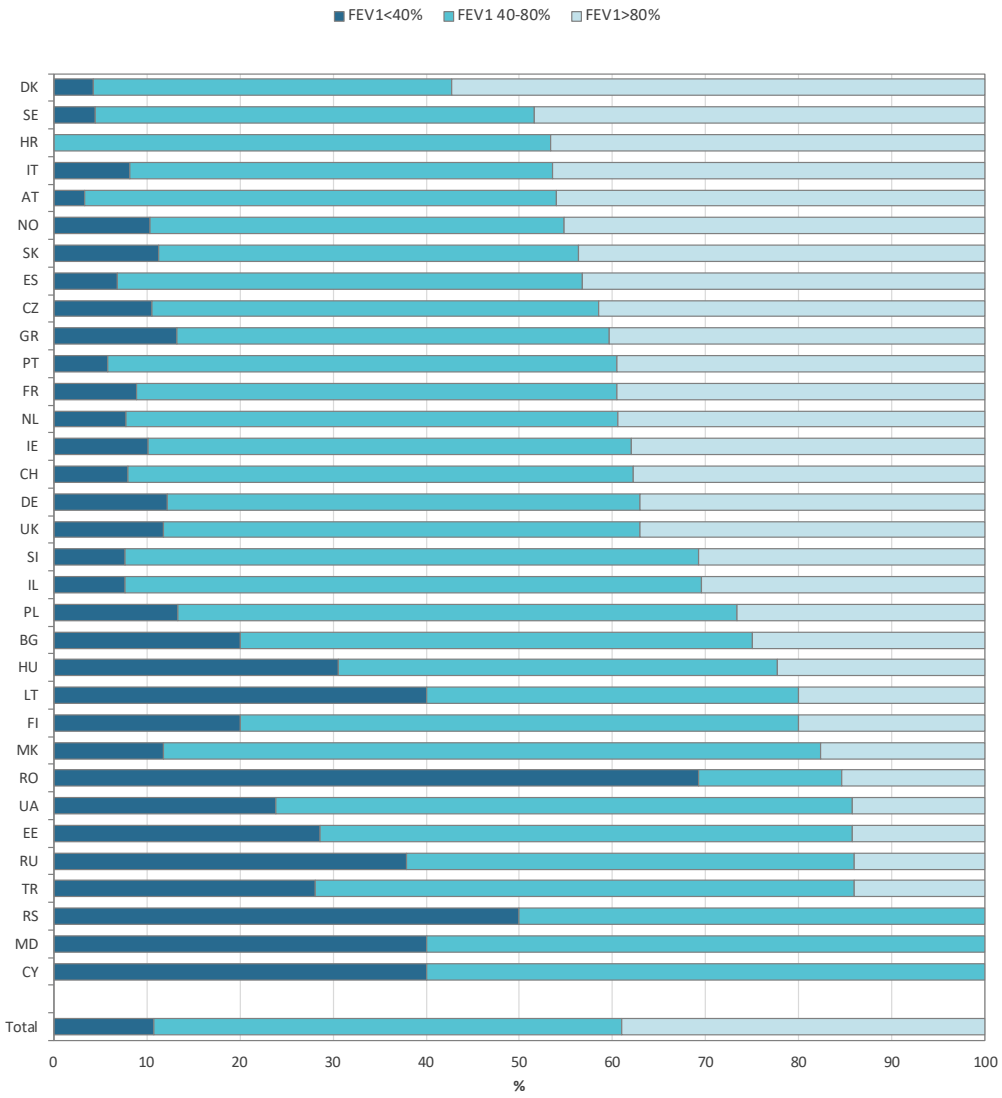
Finland and Sweden report the best FEV1pp in the 12 months prior to the annual review which means, in some cases, that the value can be from the previous calendar year.

The UK reports the best FEV1pp from the period between annual reviews which means, in some cases, that the value can be from the previous calendar year.

4. Lung function

Figure 4.7 In the majority of countries, most adults with CF aged 30 years or older have a FEV_1% of predicted between 40% and 80%.

FEV_1% of predicted according to severity group and age group, by country and overall. Adults with CF aged 30 years or older who have never had a transplant.



Note: Albania, Armenia, Belarus, Georgia, Iceland, Kazakhstan, Latvia, Luxembourg and Montenegro have <5 people aged 30 years or more with FEV_1 measurement and are excluded from the graph.

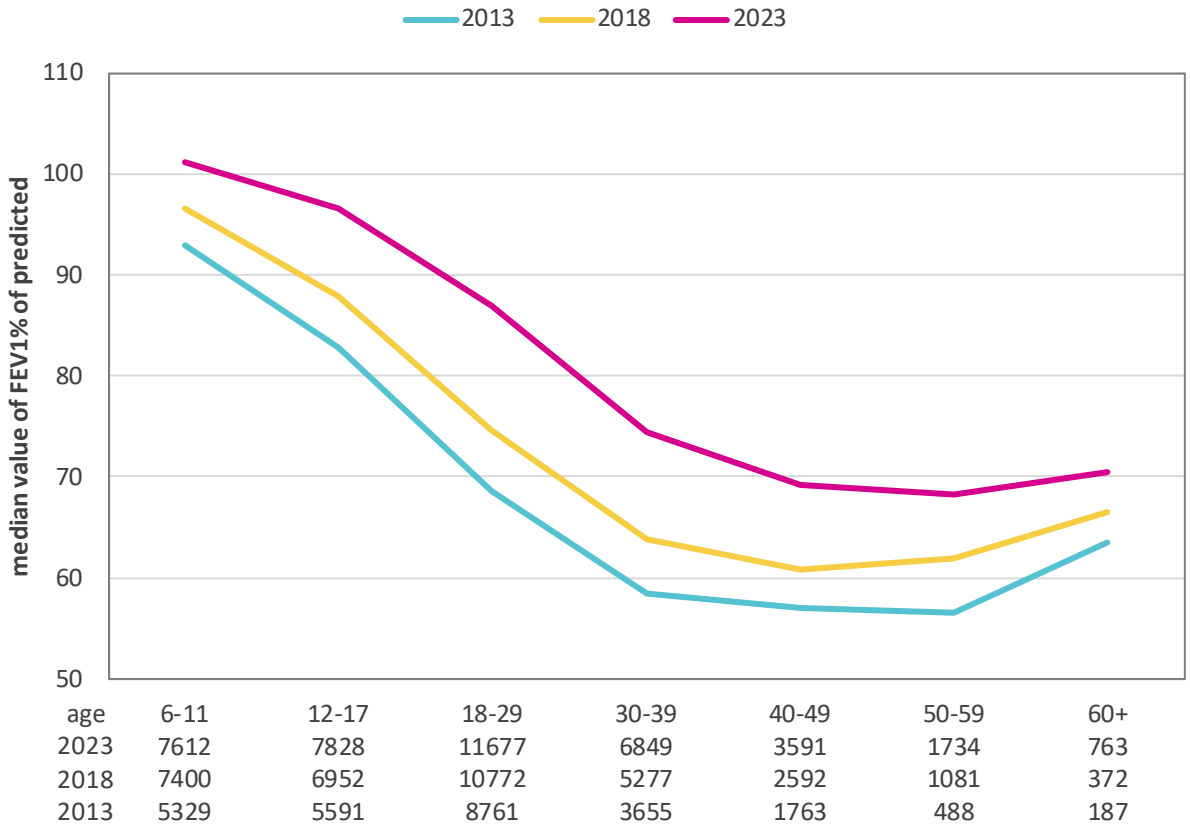
Finland and Sweden report the best FEV1pp in the 12 months prior to the annual review which means, in some cases, that the value can be from the previous calendar year.

The UK reports the best FEV1pp from the period between annual reviews which means, in some cases, that the value can be from the previous calendar year.

4. Lung function

Figure 4.8 Pulmonary function, expressed as FEV_1% of predicted, has been increasing over the years in all age groups, with a clear improvement since the introduction of CFTR modulators.

Median FEV_1% of predicted by age group in 2013, 2018 and 2023.



age	6-11	12-17	18-29	30-39	40-49	50-59	60+
2023	7612	7828	11677	6849	3591	1734	763
2018	7400	6952	10772	5277	2592	1081	372
2013	5329	5591	8761	3655	1763	488	187

Note: People with CF aged 6 years or more at lung function measurement, who have never had a lung or liver transplant.

In this graph we present data over time using cross sectional data per year of people with a confirmed CF diagnosis. The years 2013, 2018 and 2023 were used for the analysis. All people with CF, alive and deceased, were included. Exclusion criteria were people who have had a lung or liver transplant at any time during their and children younger than 6 years old at the time of the lung function measurement.

5. Microbiology

We collected data on a number of pulmonary infections common in CF and whether the infection is either chronic or not chronic/intermittent (with the exception of certain types of non-tuberculous mycobacteria where we asked only if the pathogen was found at any time during the follow-up year).
In the microbiology category there are discrepancies between the ECFSPR definition of chronicity and those of some national registries. The ECFSPR definition of chronic infection (modified Leeds criteria for chronic infection, applied also to gram negative bacteria) (see Appendix 4) is as follows:
The individual should be defined as chronically infected if he/she fulfils the criteria now or has done in recent years and the physician has no reason to believe the status has changed, when:

- >50% of respiratory samples collected during the last 12 months (at least 4 samples)are positive;

and/or

- significantly raised bacteria-specific antibodies, according to local laboratories, are present.

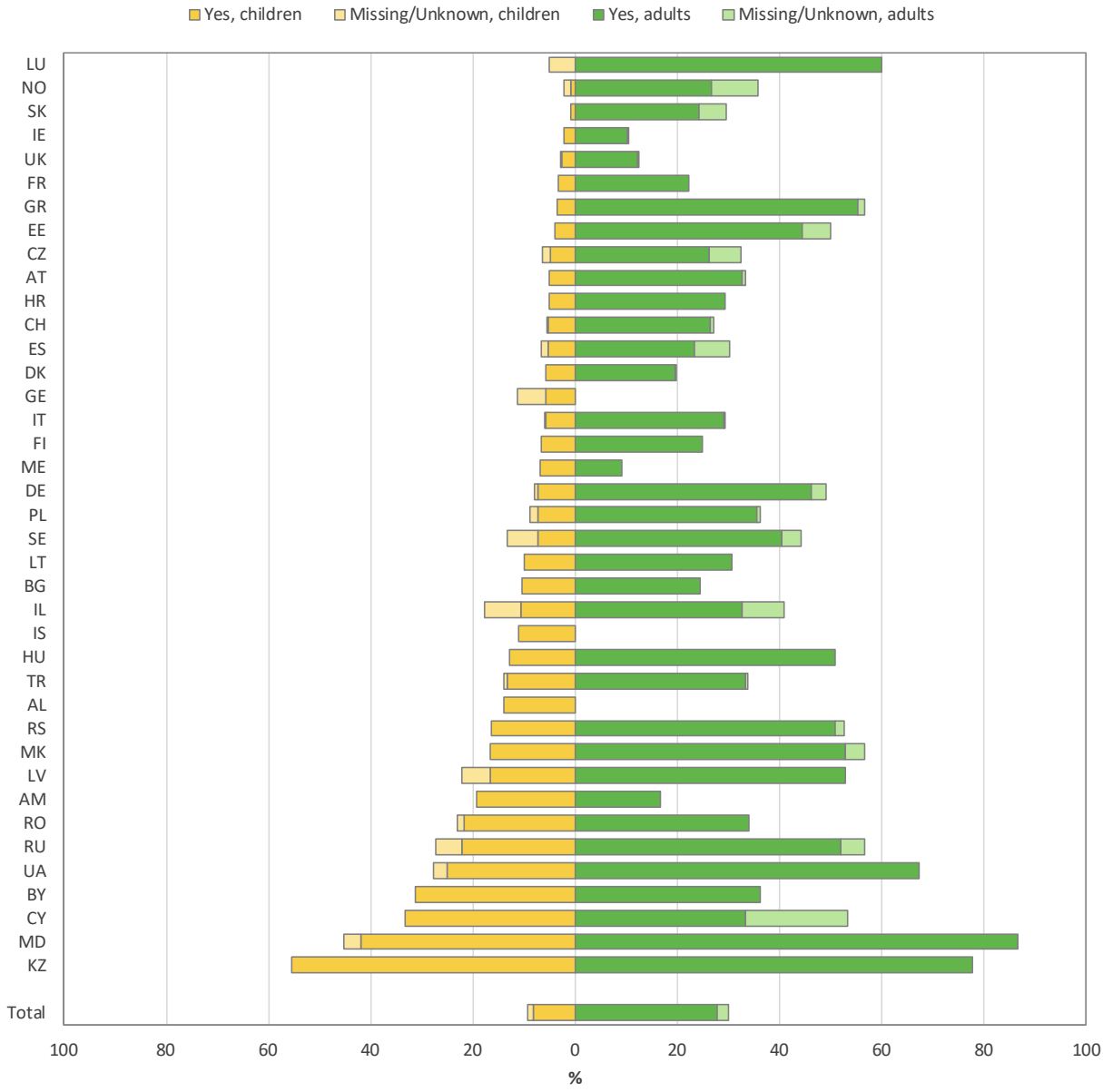
When minor differences exist, the alternative definition is in a footnote; when differences are major, or if the variable is not collected at all, the variable has been set to missing for that country.

In the following graphs and tables data from people with CF who have had a lung and/or liver transplant were excluded.

5. Microbiology

Figure 5.1 Pseudomonas aeruginosa, together with Staphylococcus aureus and Haemophilus influenzae, is the predominant respiratory pathogen in people with CF, though prevalence varies between age and countries.

Prevalence of chronic *Pseudomonas aeruginosa* in people with CF seen in 2023 who have never had a transplant, by country (table A5.1, Appendix 1).



Note: We excluded from the graph the countries for which the information is missing for more than 10% of the children/adults. Albania and Georgia have <5 adults seen in 2023 and are excluded from the graph for adults.

Note: Ireland and Italy: chronicity for *Pseudomonas aeruginosa* is defined as: at least 3 or more positive isolates during the last 12 months preceding the last reported culture in 2023.

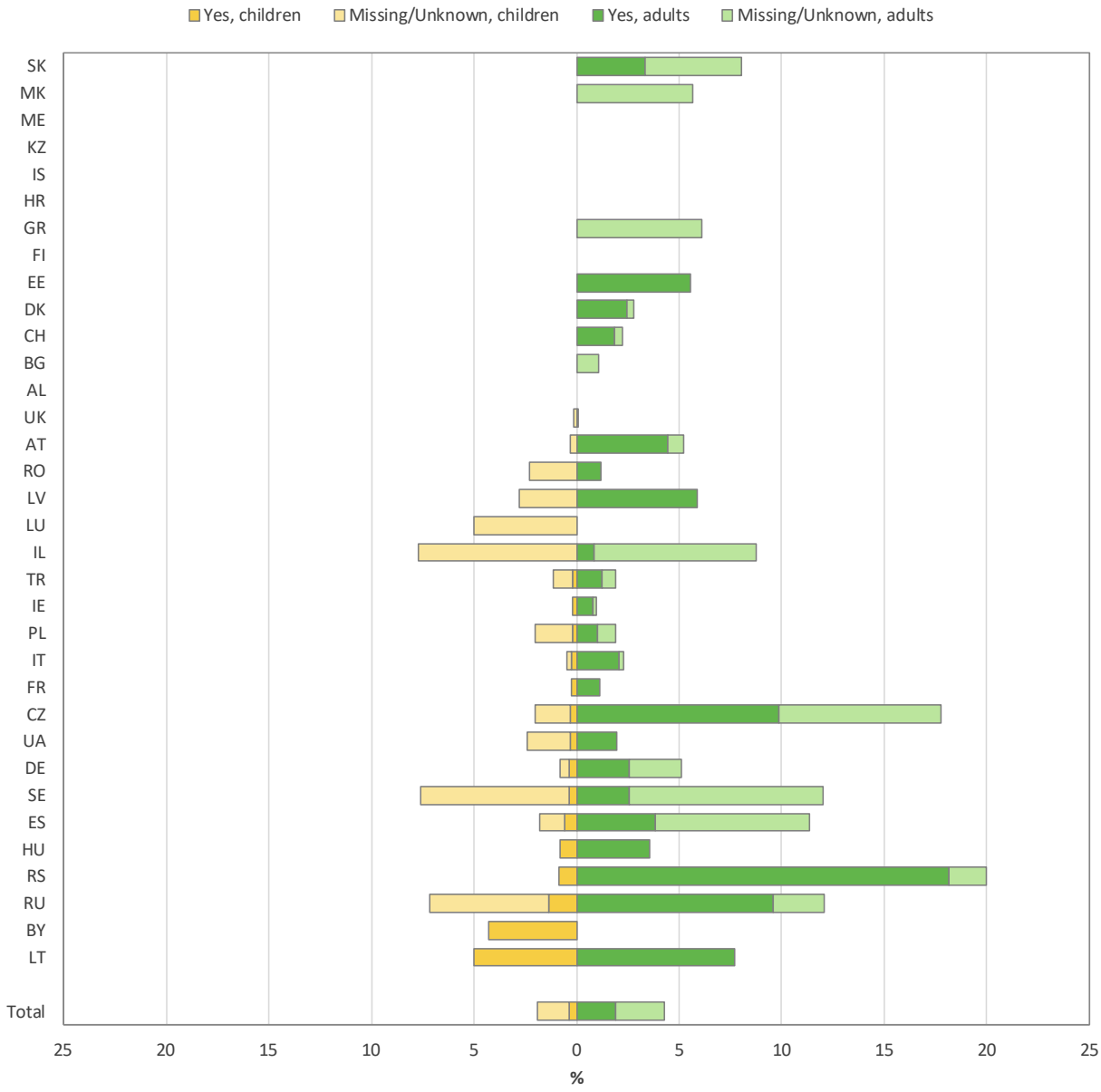
The United Kingdom: chronicity for *Pseudomonas aeruginosa* is defined as: 3 or more positive isolates during the 12 months preceding the last annual review.

This graph represents the percentage of people with chronic *Pseudomonas aeruginosa* infection (in dark colours) and the percentage of people where information on chronic *Pseudomonas aeruginosa* infection is missing/ unknown (in light colours). The bars on the left of the graph represent children and those on the right represent adults. *Pseudomonas aeruginosa* is a frequent infection, but prevalence varies considerably between countries.

5. Microbiology

Figure 5.2 Burkholderia cepacia complex species belong to the emerging respiratory pathogens, with a prevalence of >5% in some countries.

Prevalence of chronic *Burkholderia cepacia complex* species in people with CF seen in 2023 who have never had a transplant, by country (table A5.2, Appendix 1).



Note: We excluded from the graph the countries for which the information is missing for more than 10% of the children/adults. Albania and Georgia have <5 adults seen in 2023 and are excluded from the graph for adults.

Note: Ireland and Italy: chronicity for *Burkholderia cepacia complex* is defined as: at least 3 or more positive isolates during the last 12 months preceding the last reported culture in 2023.

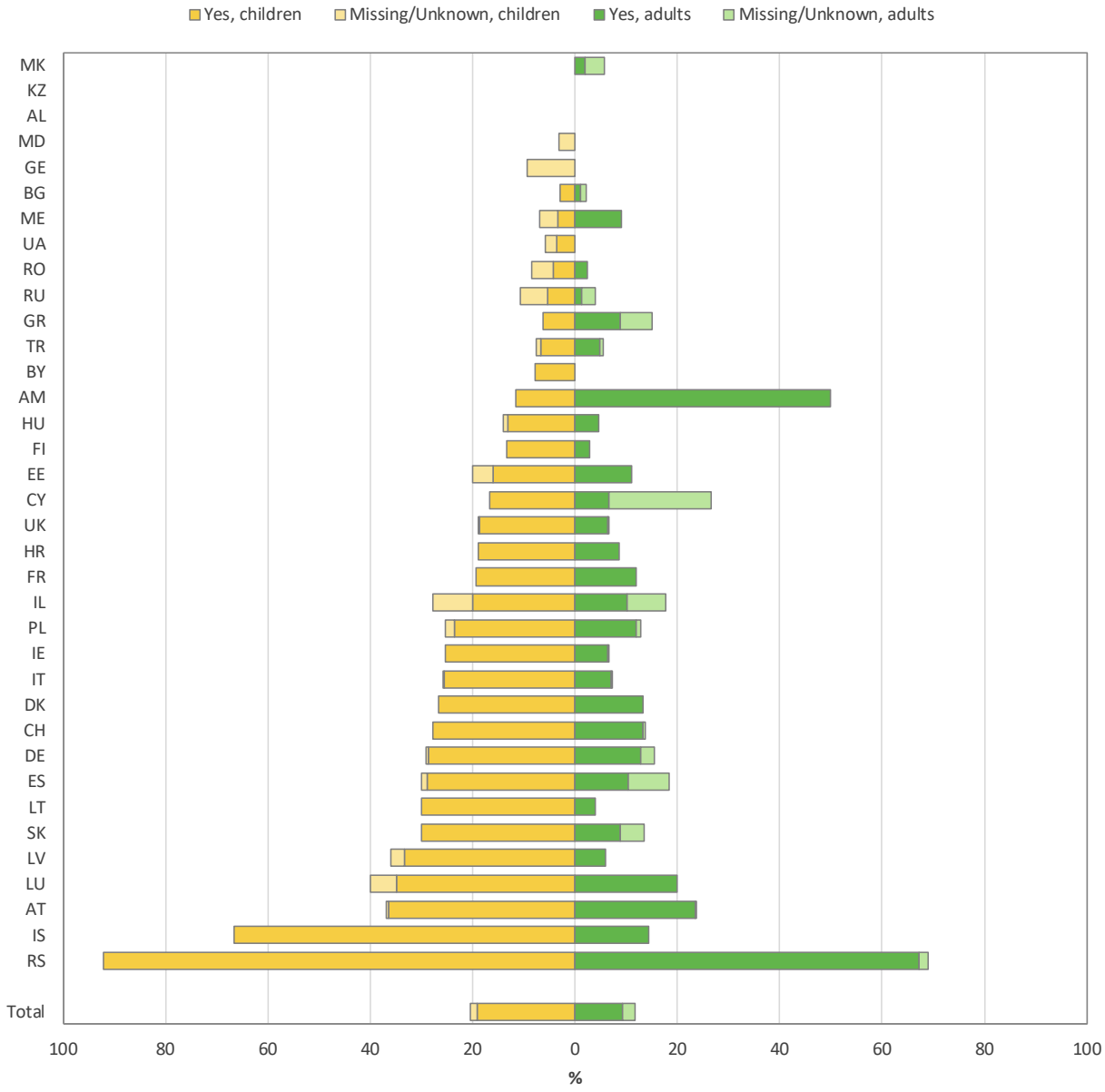
The United Kingdom: chronicity for *Burkholderia cepacia complex* is not collected.

This graph represents the percentage of people with chronic *Burkholderia cepacia* complex species infection (in dark colours) and the percentage of people where information on *Burkholderia cepacia* complex species infection is missing/unknown (in light colours). The bars on the left of the graph represent children, and the bars on the right the adults. Chronic infection by *Burkholderia cepacia* complex species in people with CF is less frequent than chronic infection by *Pseudomonas aeruginosa* (note the different scale on the horizontal axis). There is some variation among countries.

5. Microbiology

Figure 5.3 Haemophilus influenzae, together with Pseudomonas aeruginosa and Staphylococcus aureus, is the predominant respiratory pathogen in people with CF, though prevalence varies between age and countries.

Prevalence of *Haemophilus influenzae* (detected at least once a year) in people with CF seen in 2023 who have never had a transplant, by country (table A5.3, Appendix 1).



Note: We excluded from the graph the countries for which the information is missing for more than 10% of the children/adults. Albania and Georgia have <5 adults seen in 2023 and are excluded from the graph for adults.

Note: France and United Kingdom: chronicity for *Haemophilus influenza* is not collected.

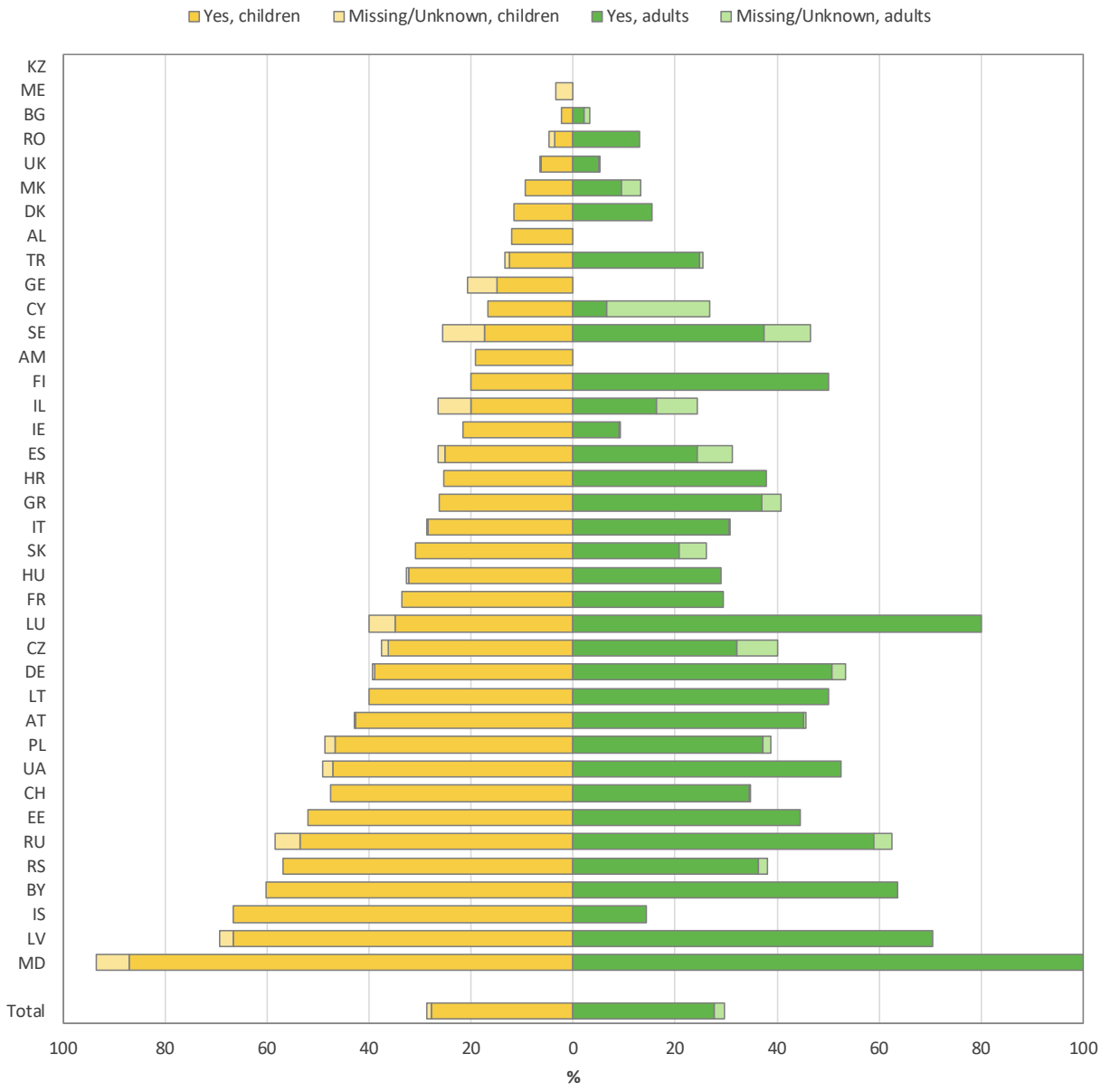
Ireland and Italy: chronicity for *Haemophilus influenzae* is defined as: at least 3 or more positive isolates during the last 12 months preceding the last reported culture in 2023.

This graph represents the percentage of people with *Haemophilus influenzae* infection (in dark colours) and the percentage of people where information on *Haemophilus influenzae* infection is missing/unknown (in light colours). The horizontal bars on the left of the graph represent children, and the bars on the right adults. This infection is as frequent as *Pseudomonas aeruginosa* infection and there is a similar degree of variation between the countries.

5. Microbiology

Figure 5.4 Staphylococcus aureus, together with Pseudomonas aeruginosa and Haemophilus influenzae, is the predominant respiratory pathogen in people with CF, though prevalence varies by age and between countries.

Prevalence of chronic methicillin-sensitive *Staphylococcus aureus* (MSSA) in people with CF seen in 2023 who have never had a transplant, by country (table A5.4, Appendix 1).



Note: We excluded from the graph the countries for which the information is missing for more than 10% of the children/adults. Albania and Georgia have <5 adults seen in 2023 and are excluded from the graph for adults.

Note: Ireland and Italy: chronicity for *Staphylococcus aureus* is defined as: at least 3 or more positive isolates during the last 12 months preceding the last reported culture in 2023.

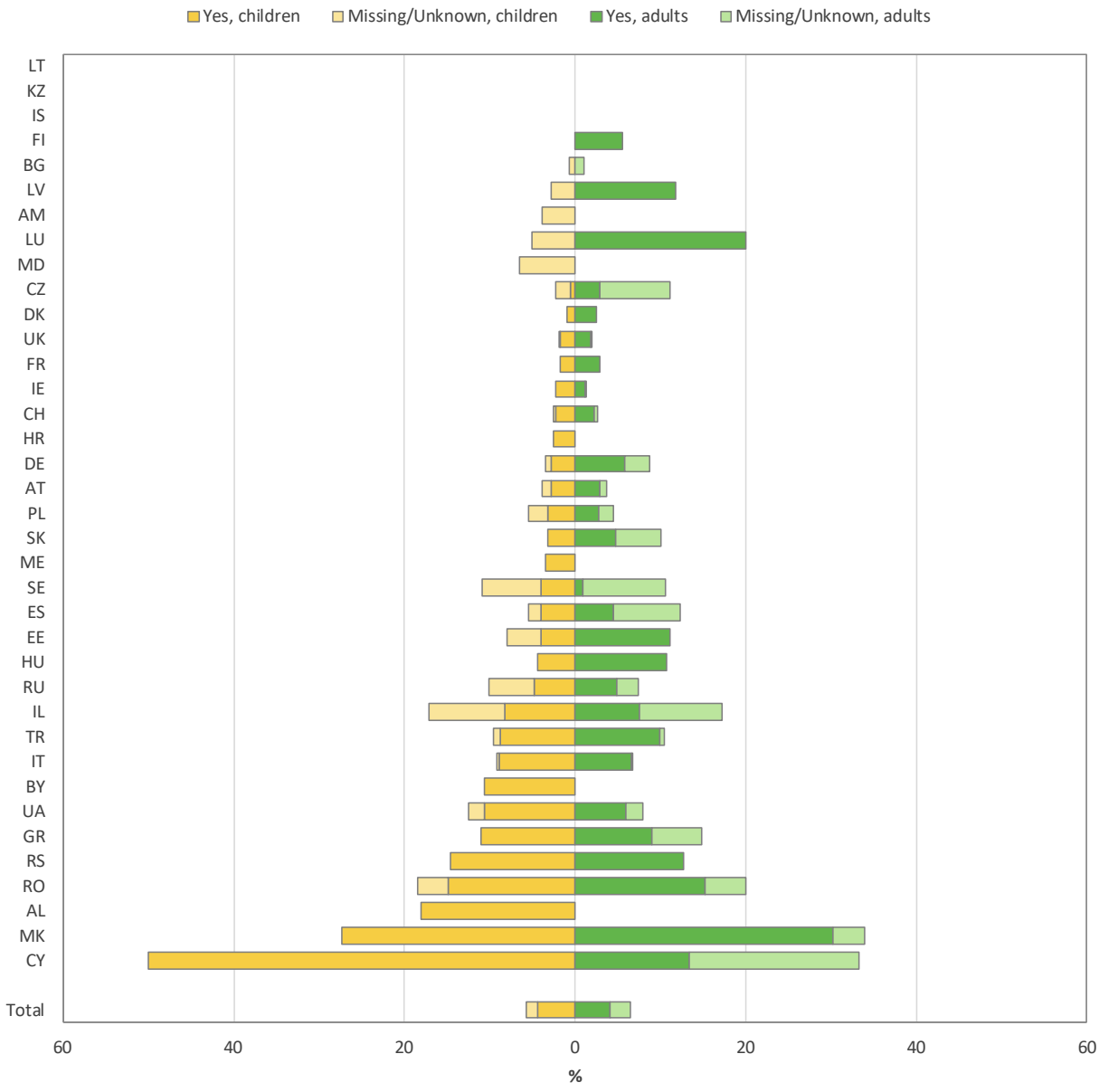
The United Kingdom: chronicity for *Staphylococcus aureus* is defined as: 3 or more positive isolates during the 12 months preceding the last annual review.

This graph represents the percentage of people with chronic methicillin-sensitive *Staphylococcus aureus* infection (dark colours) and the percentage of people where information on MSSA infection is missing/unknown (light colours). The horizontal bars on the left of the graph refer to children, while the horizontal bars on the right refer to adults. This infection is as frequent as *Pseudomonas aeruginosa* infection with a similar degree of variation between the countries.

5. Microbiology

Figure 5.5 Prevalence of methicillin-resistant Staphylococcus aureus (MRSA) in the airways is very heterogeneous in people with CF throughout Europe.

Prevalence of methicillin-resistant *Staphylococcus aureus* (MRSA) (detected at least once a year) in people with CF seen in 2023 who have never had a transplant, by country (table A5.5, Appendix 1).



Note: We excluded from the graph the countries for which the information is missing for more than 10% of the children/adults. Albania and Georgia have <5 adults seen in 2023 and are excluded from the graph for adults.

Note: Ireland and Italy: chronicity for methicillin-resistant *Staphylococcus aureus* is defined as: at least 3 or more positive isolates during the last 12 months preceding the last reported culture in 2023.

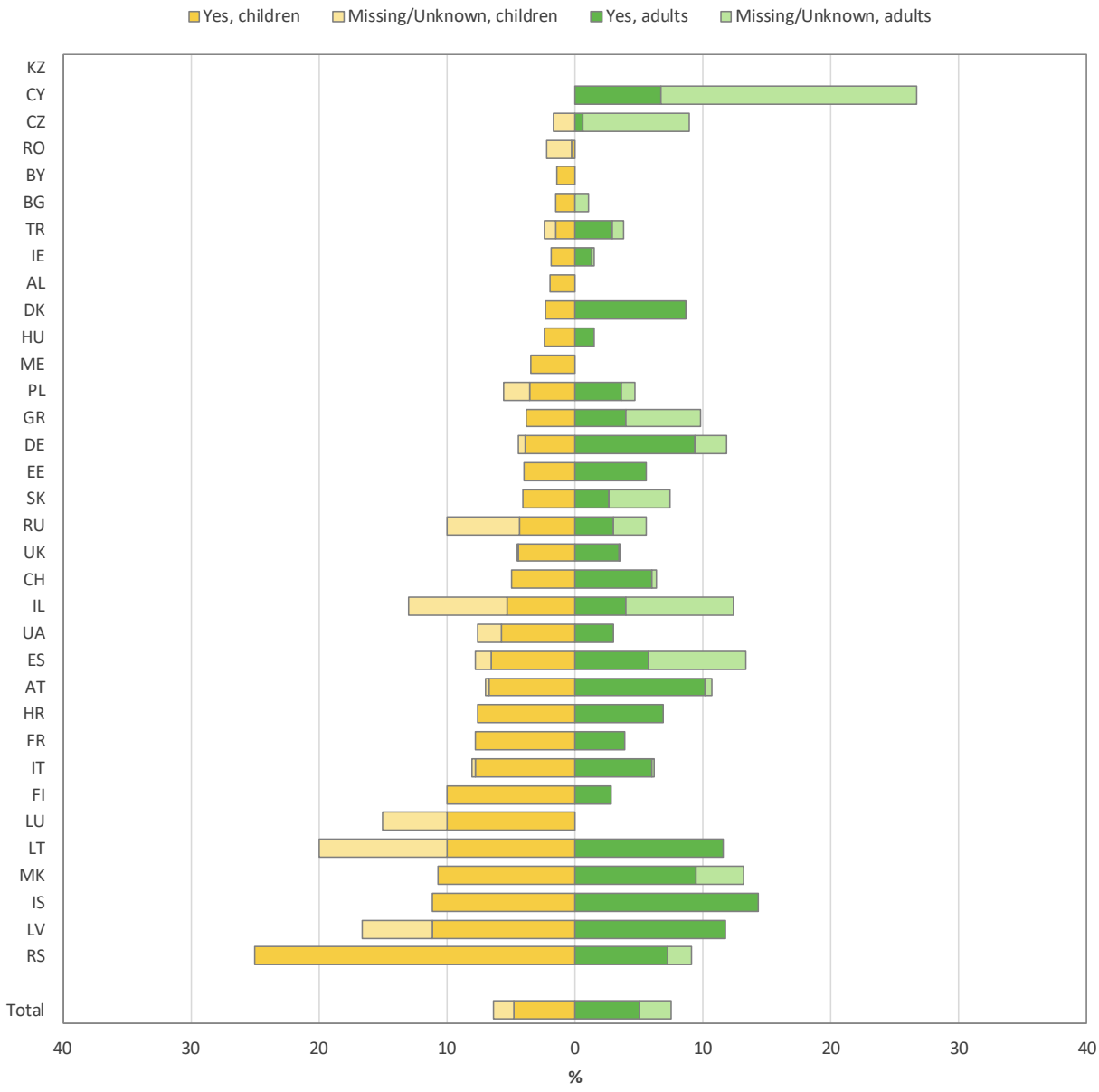
The United Kingdom: chronicity for methicillin-resistant *Staphylococcus aureus* is not collected.

This graph represents the percentage of people with methicillin-resistant *Staphylococcus aureus* (MRSA) infection (in dark colours) and the percentage of people where information on methicillin-resistant *Staphylococcus aureus* infection is missing/unknown (in light colours). The horizontal bars on the left of the graph refer to children, while the horizontal bars on the right refer to adults.

5. Microbiology

Figure 5.6 In the majority of countries, Stenotrophomonas maltophilia is found in less than 10% in children and adults with CF.

Prevalence of *Stenotrophomonas maltophilia* (detected at least once a year) in people with CF seen in 2023 who have never had a transplant, by country (table A5.6, Appendix 1).



Note: We excluded from the graph the countries for which the information is missing for more than 10% of the children/adults. Albania and Georgia have <5 adults seen in 2023 and are excluded from the graph for adults.

Note: Ireland and Italy: chronicity for *Stenotrophomonas maltophilia* is defined as: at least 3 or more positive isolates during the last 12 months preceding the last reported culture in 2023.

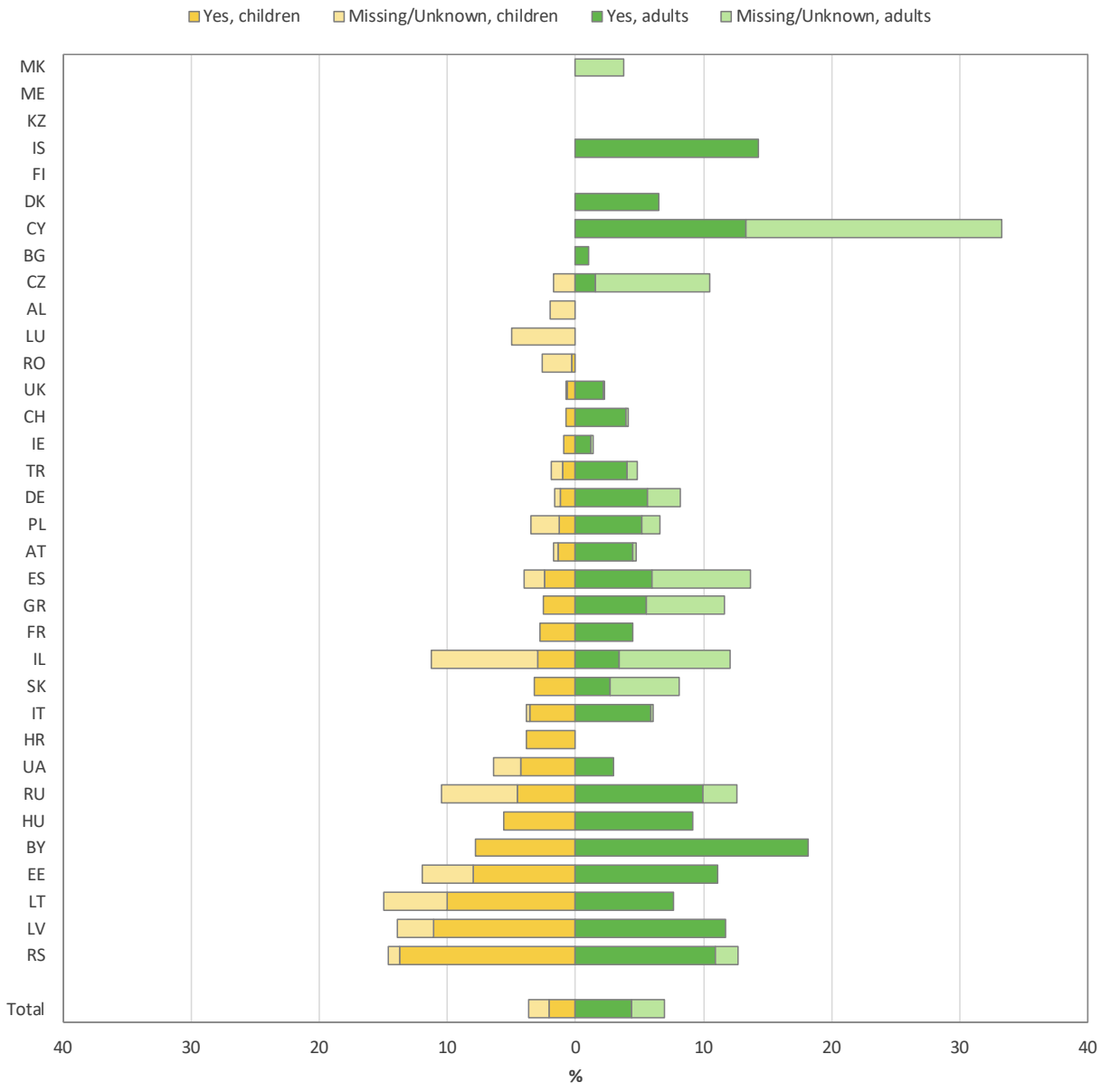
The United Kingdom: chronicity for *Stenotrophomonas maltophilia* is not collected.

This graph represents the percentage of people with *Stenotrophomonas maltophilia* infection (dark colours) and the percentage of people where information on *Stenotrophomonas maltophilia* infection is missing/unknown (in light colours). The horizontal bars on the left of the graph refer to children, while the horizontal bars on the right refer to adults.

5. Microbiology

Figure 5.7 Achromobacter species can be found in up to 20% of the airways of people with CF, with a higher prevalence in adults.

Prevalence of *Achromobacter species* infection (detected at least once a year) in people with CF seen in 2023 who have never had a transplant, by country (table A5.7, Appendix 1).



Note: We excluded from the graph the countries for which the information is missing for more than 10% of the children/adults. Albania and Georgia have <5 adults seen in 2023 and are excluded from the graph for adults.

Note: Ireland and Italy: chronicity for *Achromobacter species* is defined as: at least 3 or more positive isolates during the last 12 months preceding the last reported culture in 2023.

The United Kingdom: chronicity for *Achromobacter species* is not collected.

This graph represents the percentage of people with *Achromobacter species* infection (in dark colours) and the percentage of people where information on *Achromobacter species* infection is missing/unknown (in light colours). The horizontal bars on the left of the graph represent children, while those on the right represent adults.

5. Microbiology

Table 5.1 Prevalence of non-tuberculous mycobacteria in children (<18 years) with CF seen in 2023 who have never had a transplant, by country and overall.

Country	Sputum/BAL investigated for non-tuberculous mycobacteria infection						If yes, investigated			
	Missing/Unknown		No, not investigated		Yes, investigated		only negative cultures		at least one positive culture	
	N	%	N	%	N	%	N	%	N	%
Albania	0	0.0	50	100.0	0	0.0				
Armenia	0	0.0	20	76.9	6	23.1	6	100.0	0	0.0
Austria	0	0.0	236	65.9	122	34.1	121	99.2	1	0.8
Belarus	141	100.0	-	-	-	-				
Bulgaria	1	0.7	132	97.8	2	1.5	2	100.0	0	0.0
Croatia	5	6.3	63	79.8	11	13.9	11	100.0	0	0.0
Cyprus	0	0.0	5	83.3	1	16.7	1	100.0	0	0.0
Czech Republic	11	3.1	294	84.0	45	12.9	45	100.0	0	0.0
Denmark	0	0.0	0	0.0	214	100.0	212	99.1	2	0.9
Estonia	2	8.0	16	64.0	7	28.0	7	100.0	0	0.0
Finland	0	0.0	15	50.0	15	50.0	13	86.7	2	13.3
France	456	17.6	994	38.4	1139	44.0	1106	97.1	33	2.9
Georgia	3	5.7	50	94.3	0	0.0				
Germany	31	1.1	2388	85.8	363	13.1	349	96.1	14	3.9
Greece	236	100.0	-	-	-	-				
Hungary	7	2.8	138	55.0	106	42.2	102	96.2	4	3.8
Iceland	0	0.0	6	66.7	3	33.3	3	100.0	0	0.0
Ireland	0	0.0	293	56.0	230	44.0	227	98.7	3	1.3
Israel	2	1.2	66	39.1	101	59.8	88	87.1	13	12.9
Italy	0	0.0	1700	77.7	488	22.3	479	98.2	9	1.8
Kazakhstan	0	0.0	9	100.0	0	0.0				
Latvia	1	2.8	24	66.7	11	30.6	11	100.0	0	0.0
Lithuania	0	0.0	18	90.0	2	10.0	2	100.0	0	0.0
Luxembourg	0	0.0	8	40.0	12	60.0	12	100.0	0	0.0
Rep of Moldova	1	3.2	30	96.8	0	0.0				
Montenegro	0	0.0	2	6.9	27	93.1	26	96.3	1	3.7
The Netherlands	2	0.4	358	67.6	170	32.1	168	98.8	2	1.2
North Macedonia	0	0.0	72	85.7	12	14.3	12	100.0	0	0.0
Norway	1	0.7	34	24.8	102	74.5	102	100.0	0	0.0
Poland	63	6.4	859	87.0	65	6.6	63	96.9	2	3.1
Portugal	3	1.7	81	47.1	88	51.2	85	96.6	3	3.4
Romania	21	6.8	263	85.1	25	8.1	25	100.0	0	0.0
Russian Fed.	319	15.3	1642	78.6	129	6.2	122	94.6	7	5.4
Serbia	0	0.0	80	69.0	36	31.0	35	97.2	1	2.8
Slovak Republic	0	0.0	44	35.8	79	64.2	79	100.0	0	0.0
Slovenia	0	0.0	31	59.6	21	40.4	19	90.5	2	9.5
Spain	20	1.9	693	65.9	339	32.2	331	97.6	8	2.4
Sweden	101	36.5	71	25.6	105	37.9	102	97.1	3	2.9
Switzerland	0	0.0	351	90.9	35	9.1	34	97.1	1	2.9
Türkiye	19	0.8	1847	81.9	388	17.2	376	96.9	12	3.1
Ukraine	87	26.5	235	71.7	6	1.8	6	100.0	0	0.0
United Kingdom	282	6.9	2622	64.0	1196	29.2	1148	96.0	48	4.0
Total	1815	7.8	15840	67.8	5701	24.4	5530	97.0	171	3.0

5. Microbiology

Table 5.2 Prevalence of non-tuberculous mycobacteria in adults (≥ 18 years) with CF seen in 2023 who have never had a transplant, by country and overall.

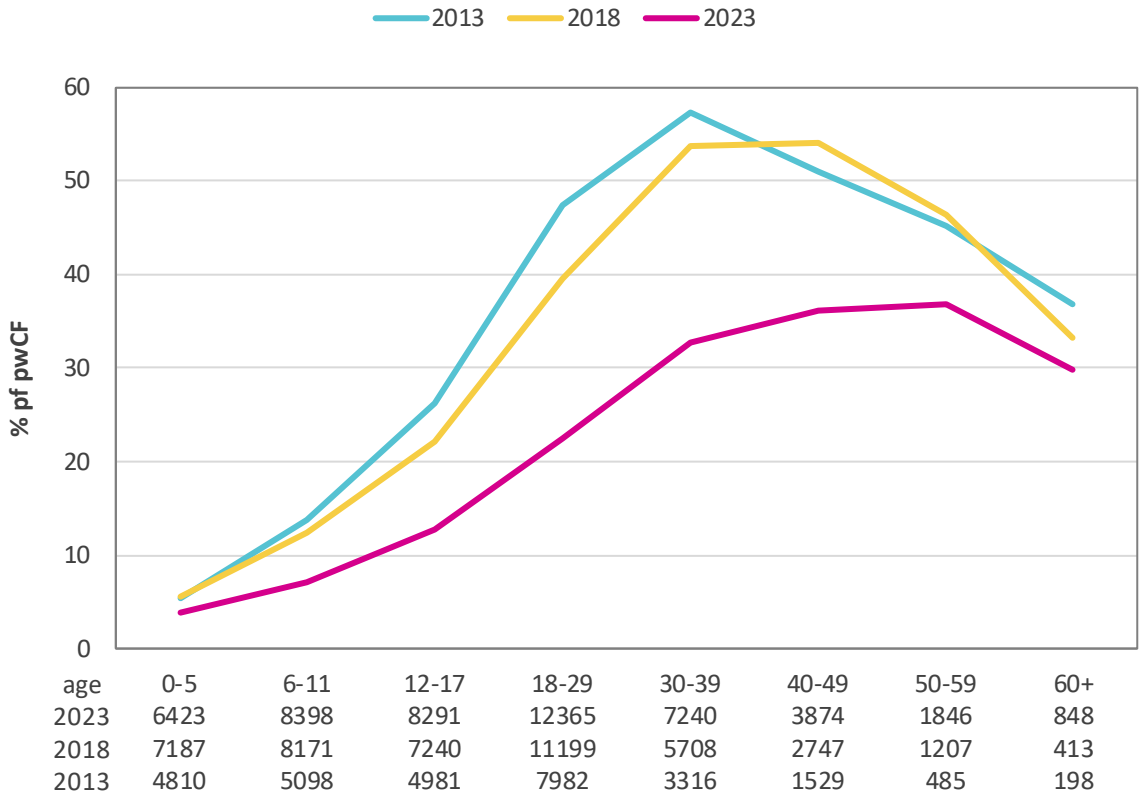
Country	Sputum/BAL investigated for non-tuberculous mycobacteria infection						If yes, investigated			
	Missing/Unknown		No, not investigated		Yes, investigated		only negative cultures		at least one positive culture	
	N	%	N	%	N	%	N	%	N	%
Armenia	0	0.0	3	50.0	3	50.0	3	100.0	0	0.0
Austria	0	0.0	205	50.7	199	49.3	179	90.0	20	10.1
Belarus	11	100.0	-	-	-	-				
Bulgaria	0	0.0	93	98.9	1	1.1	1	100.0	0	0.0
Croatia	0	0.0	58	100.0	0	0.0				
Cyprus	1	6.7	6	40.0	8	53.3	6	75.0	2	25.0
Czech Republic	14	4.4	188	59.7	113	35.9	100	88.5	13	11.5
Denmark	0	0.0	0	0.0	323	100.0	318	98.5	5	1.6
Estonia	0	0.0	4	22.2	14	77.8	14	100.0	0	0.0
Finland	2	5.6	14	38.9	20	55.6	18	90.0	2	10.0
France	1042	26.7	1152	29.5	1712	43.8	1581	92.4	131	7.7
Germany	124	3.1	2764	68.7	1134	28.2	1044	92.1	90	7.9
Greece	378	100.0	-	-	-	-				
Hungary	0	0.0	17	8.7	179	91.3	168	93.9	11	6.2
Iceland	0	0.0	3	42.9	4	57.1	2	50.0	2	50.0
Ireland	0	0.0	420	56.5	323	43.5	318	98.5	5	1.6
Israel	7	2.0	159	44.8	189	53.2	153	81.0	36	19.1
Italy	3	0.1	2624	72.0	1018	27.9	946	92.9	72	7.1
Kazakhstan	0	0.0	18	100.0	0	0.0				
Latvia	0	0.0	5	29.4	12	70.6	12	100.0	0	0.0
Lithuania	0	0.0	25	96.2	1	3.9	0	0.0	1	100.0
Luxembourg	1	20.0	3	60.0	1	20.0	1	100.0	0	0.0
Rep of Moldova	1	6.7	14	93.3	0	0.0				
Montenegro	0	0.0	0	0.0	11	100.0	11	100.0	0	0.0
The Netherlands	24	2.4	415	42.3	543	55.3	534	98.3	9	1.7
North Macedonia	2	3.8	26	49.1	25	47.2	25	100.0	0	0.0
Norway	2	1.0	73	35.4	131	63.6	119	90.8	12	9.2
Poland	27	4.7	406	70.1	146	25.2	140	95.9	6	4.1
Portugal	17	8.6	45	22.7	136	68.7	131	96.3	5	3.7
Romania	1	1.2	74	87.1	10	11.8	10	100.0	0	0.0
Russian Fed.	213	21.8	686	70.1	79	8.1	70	88.6	9	11.4
Serbia	0	0.0	6	10.9	49	89.1	49	100.0	0	0.0
Slovak Republic	0	0.0	62	41.6	87	58.4	87	100.0	0	0.0
Slovenia	8	16.3	18	36.7	23	46.9	21	91.3	2	8.7
Spain	69	5.4	366	28.5	849	66.1	800	94.2	49	5.8
Sweden	166	39.1	0	0.0	259	60.9	241	93.1	18	7.0
Switzerland	0	0.0	358	67.0	176	33.0	166	94.3	10	5.7
Türkiye	2	0.4	270	56.8	203	42.7	197	97.0	6	3.0
Ukraine	24	23.8	75	74.3	2	2.0	1	50.0	1	50.0
United Kingdom	128	2.2	2516	42.3	3302	55.5	3082	93.3	220	6.7
Total	2267	8.5	13175	49.3	11285	42.2	10548	93.5	737	6.5

Note: Albania and Georgia have <5 adults seen in 2023 and are excluded from the table for adults, but the people are included in the total number.

5. Microbiology

Figure 5.8 The prevalence of chronic Pseudomonas aeruginosa infection has decreased in the CF population in Europe since increased availability of CFTR modulators.

Prevalence of chronic *Pseudomonas aeruginosa* infection in people with CF, by age group, in 2013, 2018 and 2023.



age	0-5	6-11	12-17	18-29	30-39	40-49	50-59	60+
2023	6423	8398	8291	12365	7240	3874	1846	848
2018	7187	8171	7240	11199	5708	2747	1207	413
2013	4810	5098	4981	7982	3316	1529	485	198

In this graph we present data over time using cross sectional data per year of people with a confirmed CF diagnosis. All people with CF alive, deceased, or not seen during the year of follow-up were included. Individuals who have had a lung or/and liver transplant were excluded.

6. Nutrition

Pancreatic insufficiency is usually defined as the absence of pancreatic enzymes in two stool samples or elevated levels of fat in stools (faecal fat). Since information on faecal fat is rarely collected by the national registries we consider the use of pancreatic enzymes as an indicator of pancreatic insufficiency.

We collected weight and height measured on the date of the reported FEV_1 value (the FEV_1 of the highest FEV_1% predicted of the year). Where no FEV1 value was reported (for children under 6 or because spirometry was not done) latest weight and height measurements of the year were considered. From these raw values we calculated body mass index (BMI). BMI is an effective measurement to illustrate the nutritional status of a person because it describes the weight/height relationship; an individual with a low weight is not necessarily underweight if the height is also low. The ECFS Standards of Care guidelines recommend:

- a BMI of above 20 kg/m2 for adults;
- for older children and adolescents, that they achieve the 50th percentile for BMI;
- for infants and children up to two years of age, weight and height percentiles similar to those for the non-
- CF population.[1]

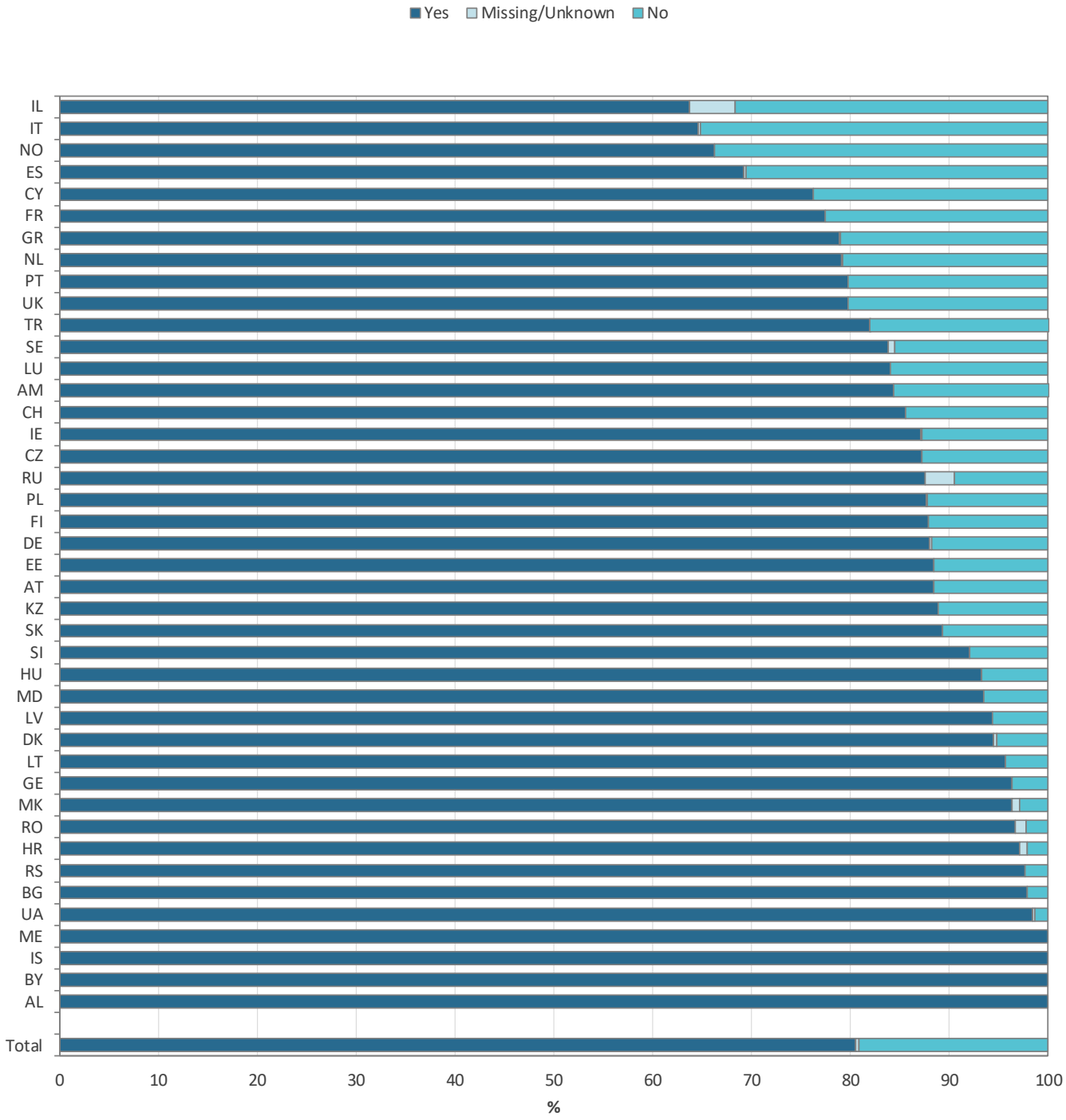
Weight, height and BMI were then expressed in terms of z-scores using a reference population of healthy individuals (in this case the US population with reference values issued by the Centre for Disease Control, USA, see Appendix 4 for details). A z-score of 0 means that the height/weight/BMI is equal to the mean height/weight/BMI of people of the same age and sex in the reference population. A z-score of-2 indicates that the height/weight/BMI value is 2 standard deviations below the mean height/weight/BMI of people of the same age and sex in the reference population; a z-score of +2 means that the value is 2 standard deviations above that mean. In the reference population, 95% of all individuals have a z-score for weight between-2 and +2 (the same for height) and it is expected that the same happens for approximately 95% of individuals in a population without conditions that affect weight (or height). The average z-score for a largely healthy population should be very close to zero.

[1] A.R. Smyth et al, JCF 2014;13, S23–S42.

6. Nutrition

Figure 6.1 In the majority of countries in the ECFSPR more than 80% of the people with CF are pancreatic insufficient.

Use of pancreatic enzymes in 2023 for all people with CF who have never had a transplant, by country.



This graph shows the use of pancreatic enzymes by country. This can be seen as an indication of pancreatic insufficiency.

6. Nutrition

Table 6.1 Z-score for BMI: descriptive statistics by country and overall. All children and adolescents with CF seen in 2023 aged 2-17 years who have never had a transplant.

Country	Number	Number of missing	Mean	Min	25th pctl (25% of the pwCF are below this z-score for BMI)	Median (half the pwCF are below this z-score for BMI)	75th pctl (75% of the pwCF are below this z-score for BMI)	Max
Albania	41	1	-0.6	-2.9	-1.4	-0.3	0.0	2.4
Armenia	22	0	-0.6	-3.8	-1.8	-0.5	1.1	2.3
Austria	346	0	-0.1	-4.2	-0.7	0.0	0.5	2.4
Belarus	74	0	-0.9	-6.8	-1.5	-0.8	0.1	1.9
Bulgaria	127	0	-0.7	-8.4	-1.5	-0.5	0.4	1.9
Croatia	75	0	-0.1	-2.7	-0.8	0.0	0.7	2.3
Cyprus	6	0	-1.0	-1.5	-1.4	-0.9	-0.8	-0.6
Czech Republic	323	0	-0.2	-3.3	-0.8	-0.1	0.5	2.3
Denmark	197	0	-0.1	-3.5	-0.7	-0.1	0.6	2.5
Estonia	25	0	-0.3	-2.4	-1.0	-0.3	0.6	1.1
Finland	29	0	-0.1	-1.6	-0.8	0.0	0.7	1.3
France	2406	3	-0.3	-5.5	-0.9	-0.2	0.4	3.1
Georgia	38	0	-0.9	-3.3	-1.7	-1.0	0.1	1.1
Germany	2596	11	-0.2	-5.2	-0.8	-0.1	0.5	2.9
Greece	228	4	0.2	-3.9	-0.4	0.2	0.8	3.0
Hungary	221	1	-0.5	-4.5	-1.2	-0.5	0.3	2.5
Iceland	8	0	0.3	-0.4	-0.1	0.3	0.5	1.1
Ireland	512	2	0.3	-3.8	-0.3	0.3	0.9	2.7
Israel	166	0	-0.1	-4.1	-0.9	-0.1	0.7	3.1
Italy	2068	4	0.0	-7.9	-0.7	0.0	0.7	2.9
Kazakhstan	10	0	-1.0	-2.1	-1.5	-1.0	-0.6	0.8
Latvia	32	0	-0.5	-2.7	-1.0	-0.3	0.1	1.0
Lithuania	18	0	-1.1	-2.7	-1.8	-1.1	-0.4	0.7
Luxembourg	15	0	-0.1	-1.7	-1.1	0.2	0.9	1.3
Rep of Moldova	25	1	-1.6	-6.9	-2.0	-1.0	-0.4	0.6
Montenegro	27	0	-0.2	-3.3	-1.0	-0.2	1.0	2.4
Netherlands	500	0	-0.1	-2.9	-0.7	-0.1	0.5	2.1
N. Macedonia	77	1	-0.5	-5.5	-1.3	-0.2	0.2	1.9
Norway	119	0	0.1	-3.4	-0.5	0.1	0.6	2.3
Poland	919	3	-0.3	-6.9	-0.9	-0.2	0.4	3.1
Portugal	163	0	-0.3	-4.2	-0.8	-0.2	0.3	2.1
Romania	276	15	-0.8	-7.2	-1.7	-0.6	0.2	1.9
Russian Fed.	1957	1	-0.6	-7.7	-1.2	-0.5	0.3	3.1
Serbia	106	1	-0.3	-3.8	-1.0	-0.2	0.5	2.4
Slovak Republic	116	0	-0.4	-6.4	-1.0	-0.3	0.4	2.3
Slovenia	53	0	-0.2	-2.0	-0.8	-0.3	0.3	1.7
Spain	1000	4	-0.2	-5.3	-0.7	-0.1	0.5	3.3
Sweden	270	0	0.0	-3.0	-0.7	0.1	0.6	2.5
Switzerland	373	0	-0.1	-2.9	-0.7	-0.1	0.6	2.1
Türkiye	2095	0	-0.5	-6.8	-1.3	-0.4	0.5	4.0
Ukraine	301	2	-0.9	-7.8	-1.6	-0.7	0.0	2.5
United Kingdom	3888	28	0.2	-5.0	-0.4	0.2	0.8	5.6
Total	21848	82	-0.2	-8.4	-0.8	-0.1	0.6	5.6

Note: Finland, Sweden and the United Kingdom: height and weight at the date of the annual review are used. This may not be the date of the best FEV1pp.

Note: If no lung function measurement is reported, height and weight at the date of the last visit are used.

This table reports the mean z-score for BMI, the median z-score for BMI and other descriptive statistics for children and adolescents aged 2 to 17 years, by country.

6. Nutrition

Table 6.2 BMI: descriptive statistics by country and overall. All adults with CF seen in 2023 who have never had a transplant.

Country	Number	Number of missing	Mean	Min	25th pctl (25% of the pwCF are below this BMI)	Median (half the pwCF are below this BMI)	75th pctl (75% of the pwCF are below this BMI)	Max
Armenia	6	0	21.0	18.3	19.1	19.4	21.0	29.1
Austria	383	0	23.1	14.9	20.6	22.4	25.1	42.7
Bulgaria	89	0	21.3	15.9	19.5	20.7	22.9	34.5
Croatia	56	0	22.7	14.5	20.6	22.9	24.5	29.7
Cyprus	14	0	24.2	17.9	20.6	23.4	24.7	43.1
Czech Republic	299	0	22.9	15.1	20.5	22.5	25.1	40.0
Denmark	299	0	23.7	16.3	20.9	23.3	25.4	43.2
Estonia	14	0	19.9	15.4	17.5	19.0	21.3	27.1
Finland	36	0	23.3	15.2	21.2	23.3	24.2	34.9
France	3669	41	22.7	11.5	20.2	22.0	24.4	49.1
Germany	3898	23	22.9	13.1	20.4	22.3	24.7	52.3
Greece	326	22	23.4	16.2	21.1	23.0	25.6	37.9
Hungary	189	0	21.6	14.3	19.7	21.2	23.2	34.7
Iceland	7	0	24.8	22.0	23.1	23.9	27.7	28.7
Ireland	627	98	24.3	16.0	21.6	23.8	26.4	46.1
Israel	339	0	23.5	14.5	20.9	23.1	25.8	42.8
Italy	3358	118	23.2	13.9	20.7	22.6	24.9	54.1
Kazakhstan	16	0	18.0	13.9	16.4	17.8	19.7	22.5
Latvia	17	0	20.8	16.5	18.7	20.8	21.9	26.9
Lithuania	22	0	20.5	14.3	18.6	20.1	22.5	25.8
Rep of Moldova	13	0	19.5	17.2	18.0	18.5	20.2	23.7
Montenegro	7	0	21.7	17.4	18.3	22.1	22.8	27.5
Netherlands	949	4	23.4	13.7	21.1	22.9	25.0	51.0
N. Macedonia	47	0	22.3	17.0	20.2	21.4	24.5	32.2
Norway	202	0	23.6	15.1	21.1	23.2	25.5	40.4
Poland	541	4	22.3	14.8	19.9	21.9	24.1	39.5
Portugal	187	4	23.2	16.9	20.6	22.7	25.0	44.6
Romania	70	0	20.6	13.5	18.4	20.6	22.3	31.2
Russian Fed.	923	0	20.4	11.9	18.1	19.8	21.8	38.7
Serbia	54	0	20.5	15.7	18.4	20.0	22.9	28.9
Slovak Republic	138	0	22.4	15.4	19.7	21.9	24.9	34.7
Slovenia	42	3	21.5	16.0	19.8	21.4	23.4	27.7
Spain	1211	21	23.4	15.1	20.7	22.8	25.3	50.7
Sweden	413	3	23.5	14.9	21.2	22.7	25.2	50.6
Switzerland	523	0	22.4	13.6	20.4	22.0	24.1	42.2
Türkiye	442	0	21.6	12.5	18.8	21.0	24.1	37.7
Ukraine	94	0	19.7	13.1	17.6	19.5	21.3	30.5
United Kingdom	5710	115	24.5	13.2	21.5	23.8	26.7	57.9
Total	25236	456	23.2	11.5	20.5	22.6	25.1	57.9

Note: Albania and Luxembourg have <5 adults seen in 2023 with information on height and weight and are excluded from the table, but the individuals are included in the total number.

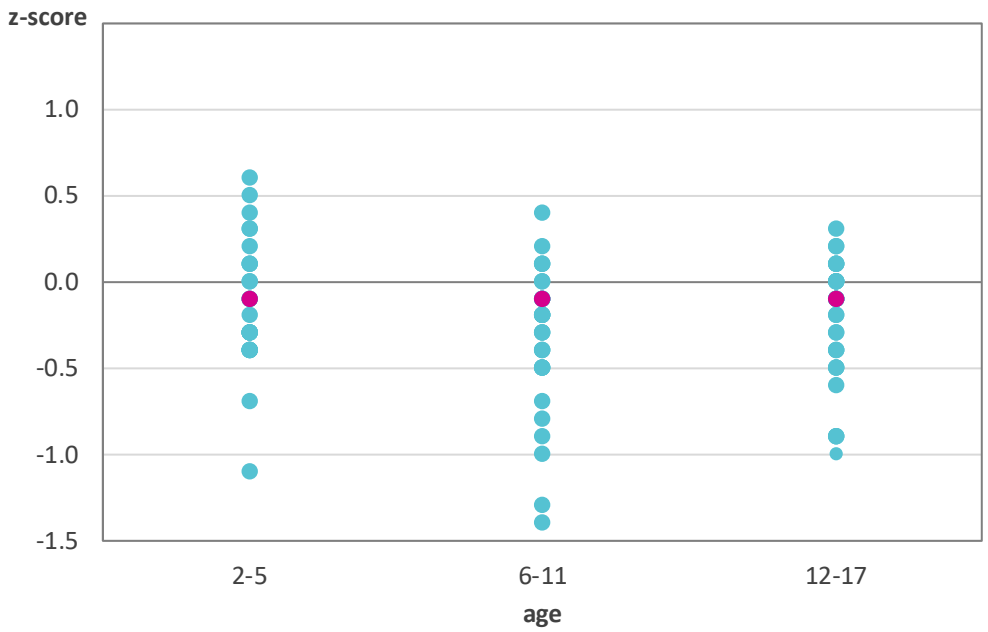
Note: Finland, Sweden and the United Kingdom: height and weight at the date of the annual review are used instead of at the date of FEV1 of the best FEV1pp. If no lung function measurement is reported, the date of the last visit is used.

This table reports the mean BMI (expressed as absolute values, not as z-scores), the median BMI and other descriptive statistics for all adults aged 18 years or older, by country and overall.

6. Nutrition

Figure 6.2 While the median BMI z-score for children and adolescents with CF in Europe is close to normal for all age groups, a lot of variation amongst the countries can be observed.

Median z-score for BMI by age group and by country. Children and adolescents with CF aged 2-17 years in 2023 who have never had a transplant.



Note: We excluded from the graph those age groups where the number of individuals was <10.

This graph shows the median z-score for BMI (the value that separates the highest and lowest half of the people with CF) by age group. Each country median is represented by a turquoise dot and the median overall for the age group by a pink dot. There is a lot of variation between countries.

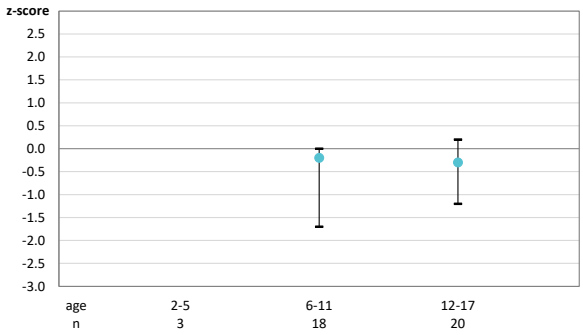
6. Nutrition

Figure 6.3 The median BMI of children and adolescents with CF is influenced by age and country of residence.

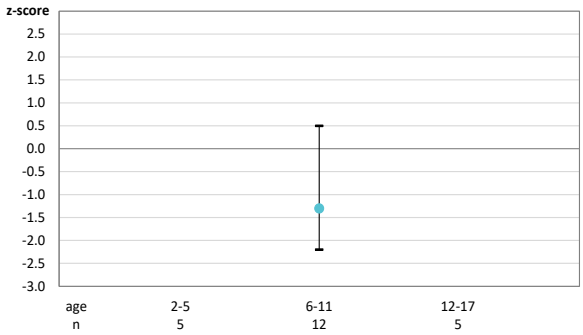
Quartiles of z-scores for BMI by age group and country. Children and adolescents with CF aged 2-17 years in 2023 who have never had a transplant.

The figures below show the z-scores for BMI by country. The dot is the median, and the whiskers show the 25th and 75th percentiles. We did not calculate quartiles where the number of individuals in the age group is <10 and therefore there are no blue dots for those age groups (the number of people in each age group is shown underneath the horizontal axis). We therefore excluded Cyprus, Iceland, Lithuania and Luxembourg from the graphs because none of the age groups in these countries had more than 10 individuals.

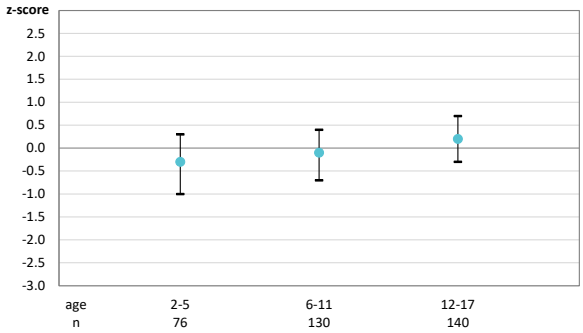
Quartiles of z-scores for BMI: Albania



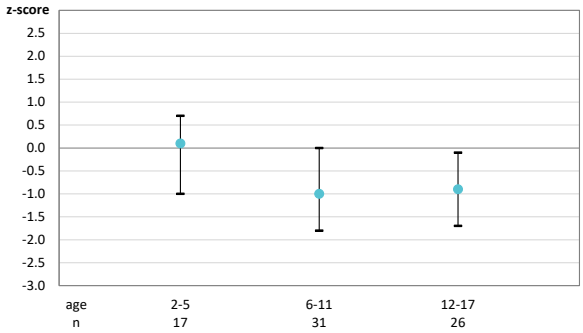
Quartiles of z-scores for BMI: Armenia



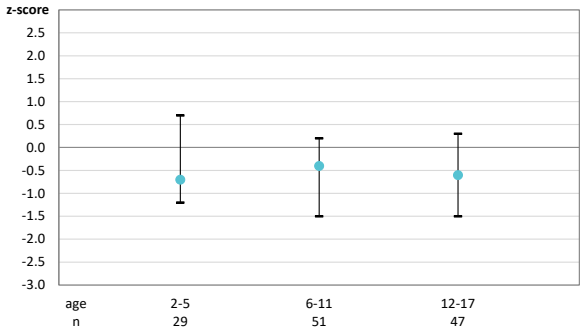
Quartiles of z-scores for BMI: Austria



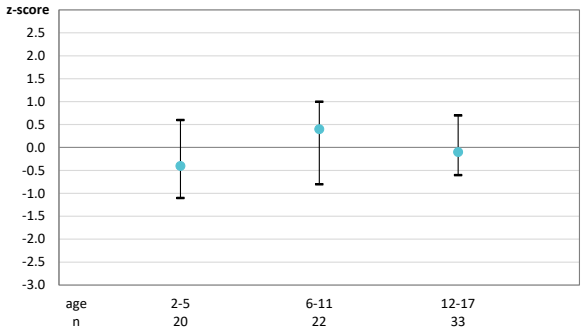
Quartiles of z-scores for BMI: Belarus



Quartiles of z-scores for BMI: Bulgaria



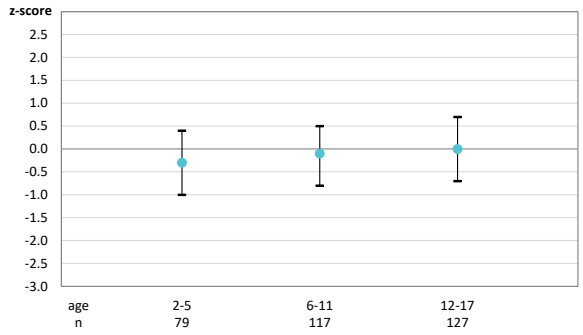
Quartiles of z-scores for BMI: Croatia



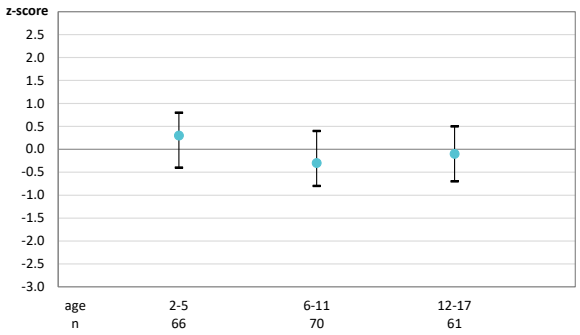
6. Nutrition

[figure 6.3 continued]

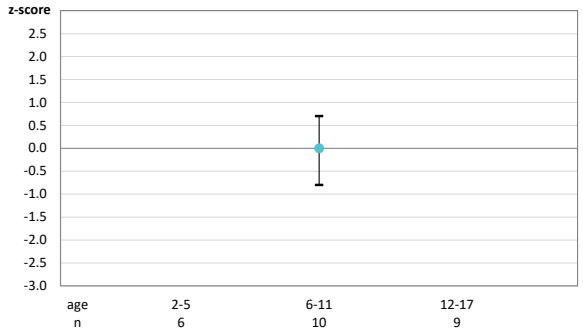
Quartiles of z-scores for BMI: Czech Republic



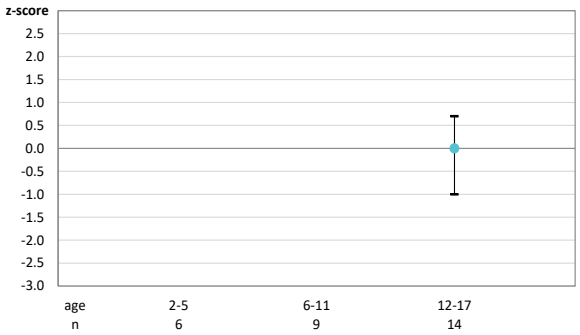
Quartiles of z-scores for BMI: Denmark



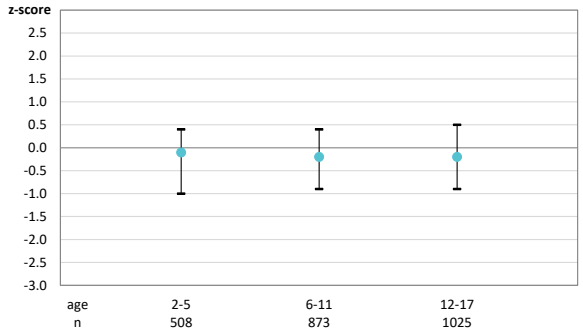
Quartiles of z-scores for BMI: Estonia



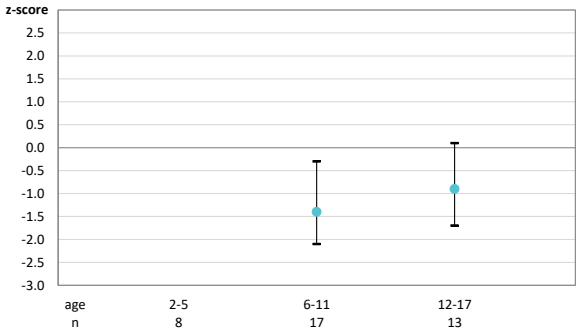
Quartiles of z-scores for BMI: Finland



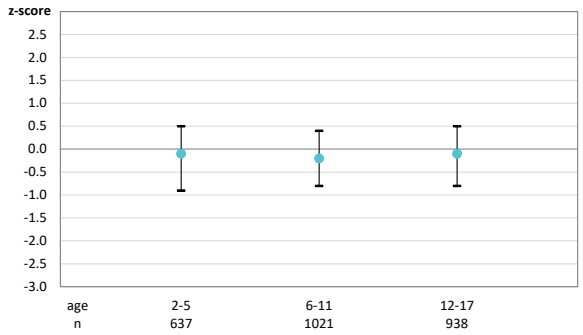
Quartiles of z-scores for BMI: France



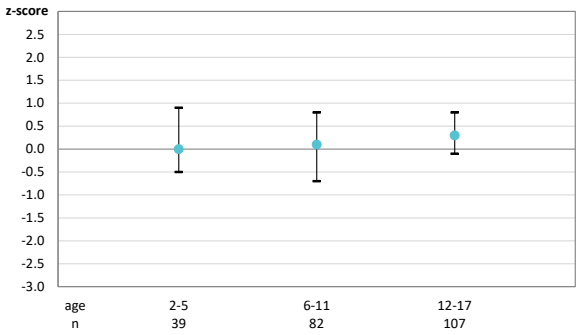
Quartiles of z-scores for BMI: Georgia



Quartiles of z-scores for BMI: Germany



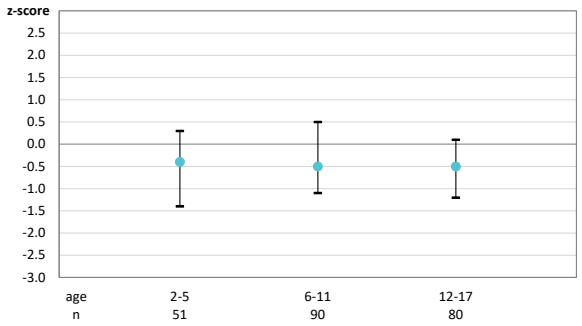
Quartiles of z-scores for BMI: Greece



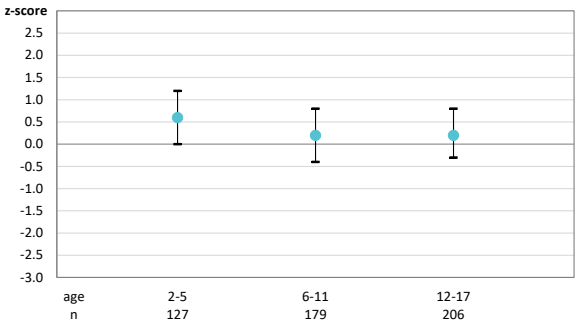
6. Nutrition

[figure 6.3 continued]

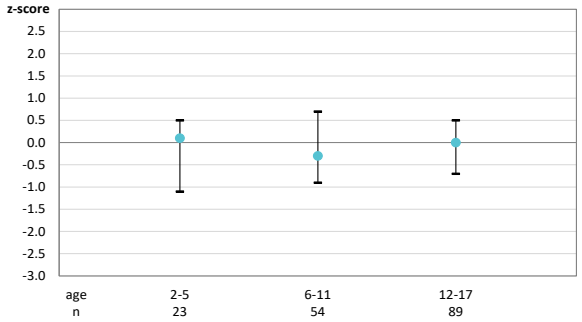
Quartiles of z-scores for BMI: Hungary



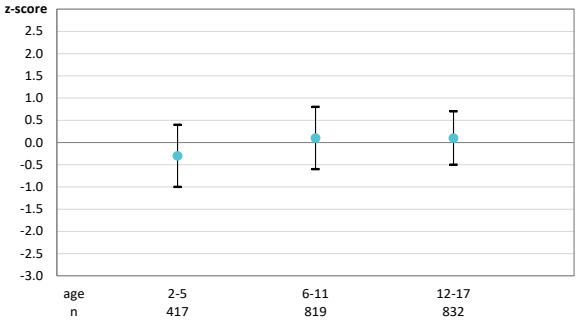
Quartiles of z-scores for BMI: Ireland



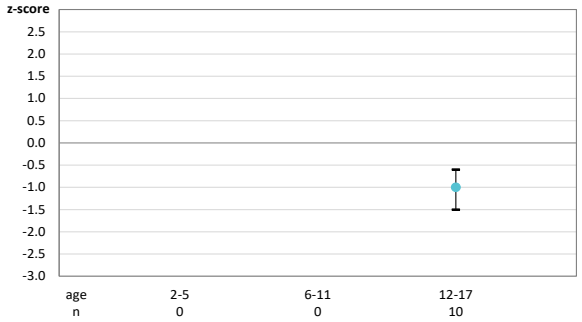
Quartiles of z-scores for BMI: Israel



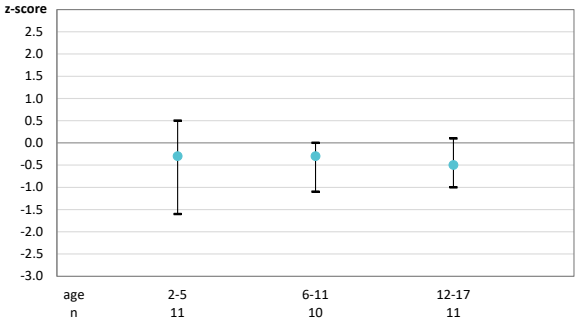
Quartiles of z-scores for BMI: Italy



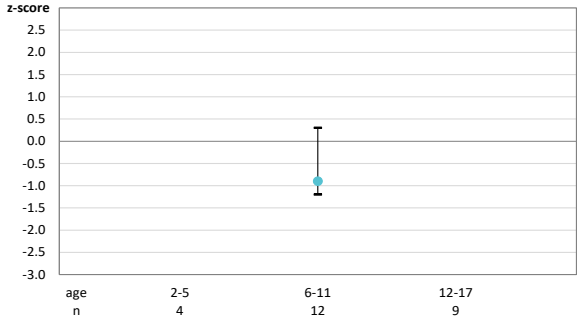
Quartiles of z-scores for BMI: Kazakhstan



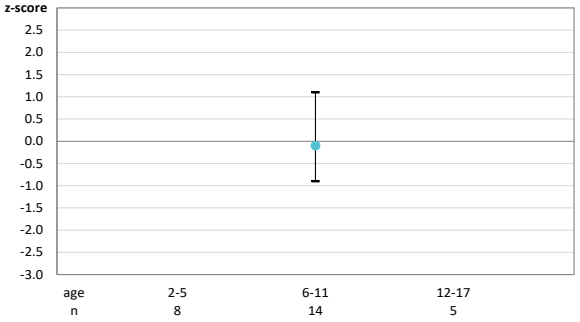
Quartiles of z-scores for BMI: Latvia



Quartiles of z-scores for BMI: Rep. of Moldova



Quartiles of z-scores for BMI: Montenegro



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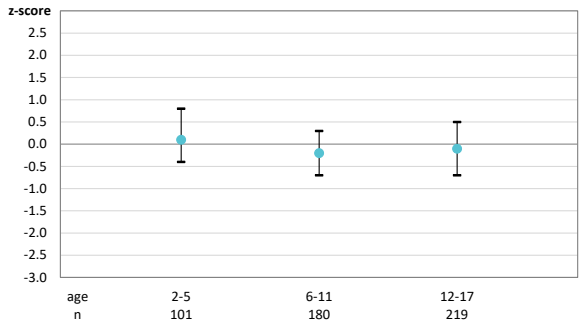
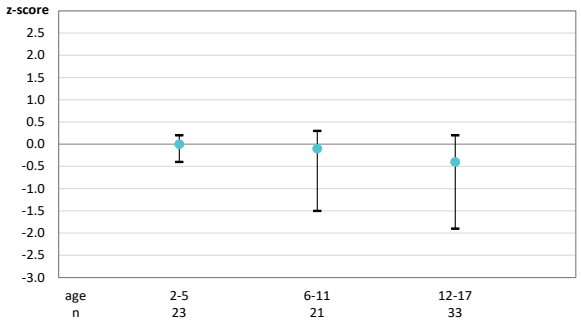
[figure 6.3 continued]

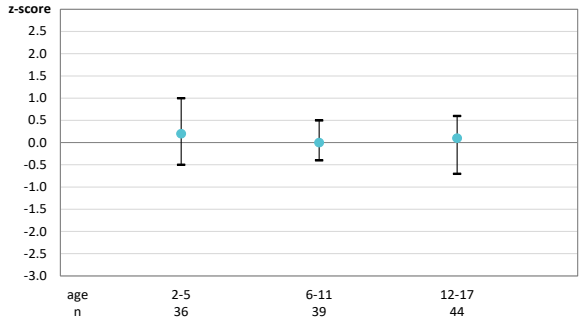
Quartiles of z-scores for BMI: The Netherlands



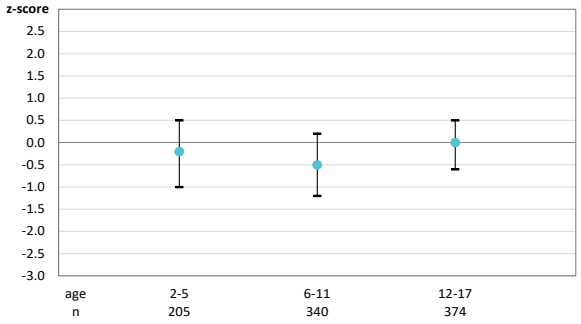
Quartiles of z-scores for BMI: North Macedonia



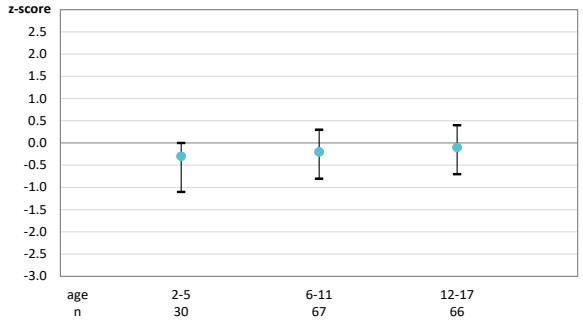
Quartiles of z-scores for BMI: Norway



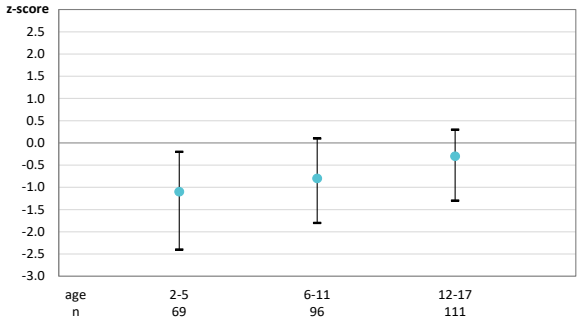
Quartiles of z-scores for BMI: Poland



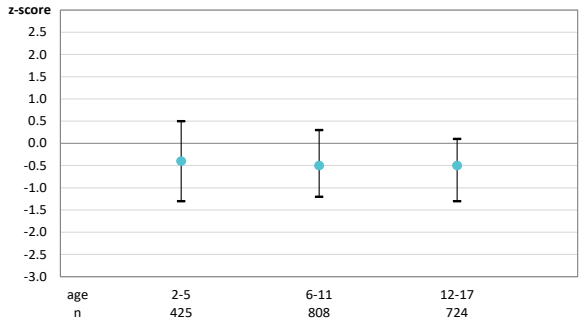
Quartiles of z-scores for BMI: Portugal



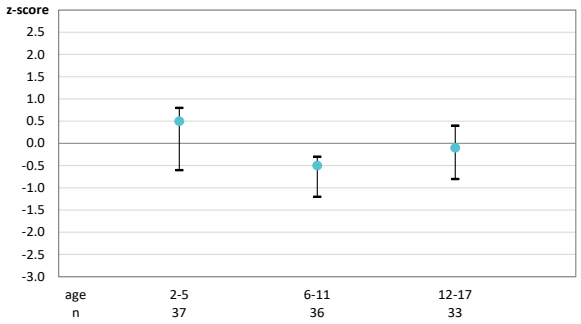
Quartiles of z-scores for BMI: Romania



Quartiles of z-scores for BMI: Russian federation



Quartiles of z-scores for BMI: Serbia



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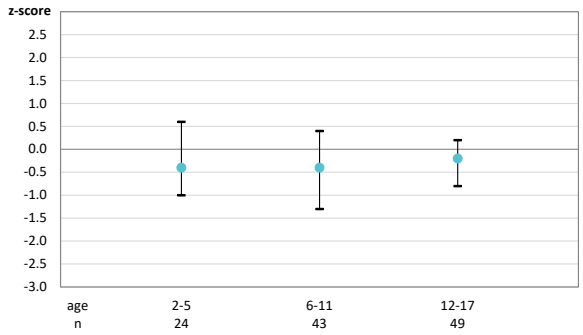
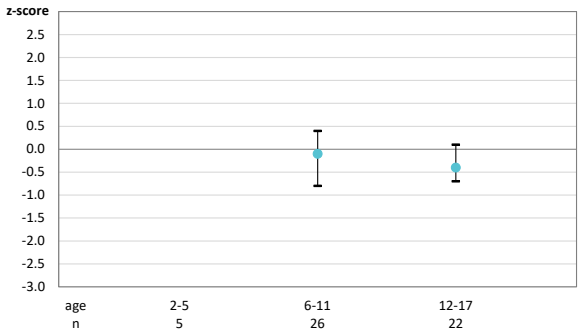
[figure 6.3 continued]

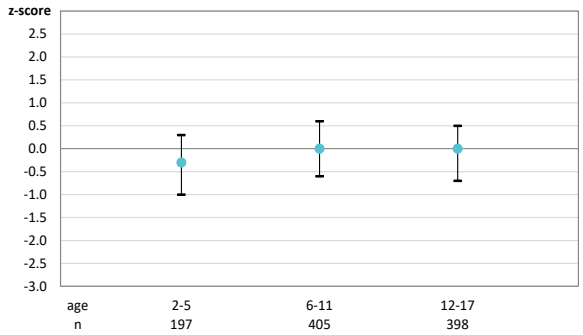
Quartiles of z-scores for BMI: Slovakia



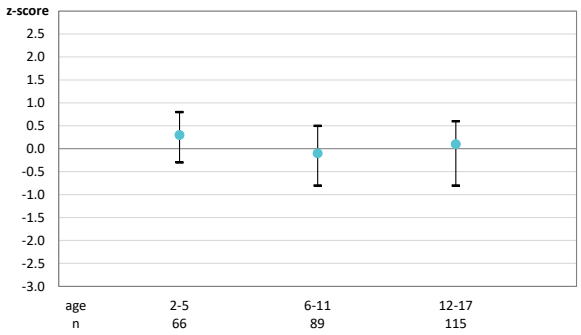
Quartiles of z-scores for BMI: Slovenia



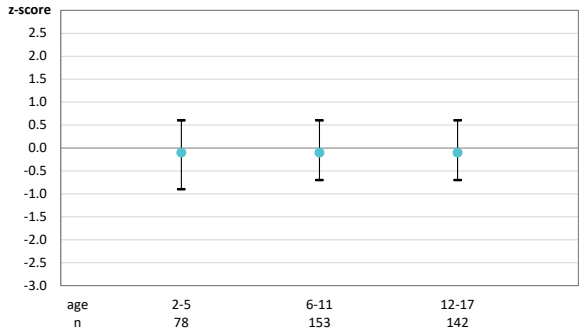
Quartiles of z-scores for BMI: Spain



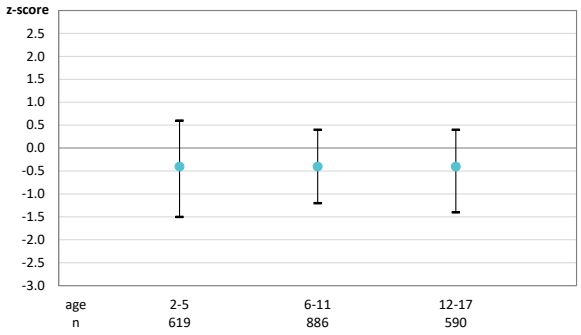
Quartiles of z-scores for BMI: Sweden



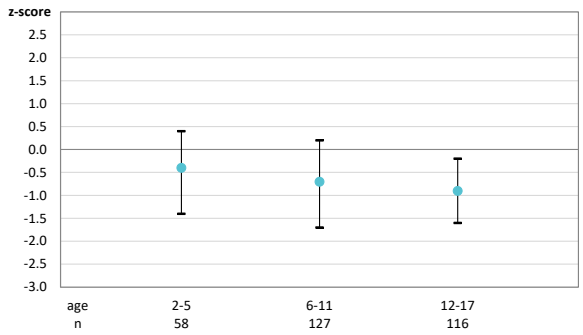
Quartiles of z-scores for BMI: Switzerland



Quartiles of z-scores for BMI: Türkiye



Quartiles of z-scores for BMI: Ukraine



Quartiles of z-scores for BMI: United Kingdom

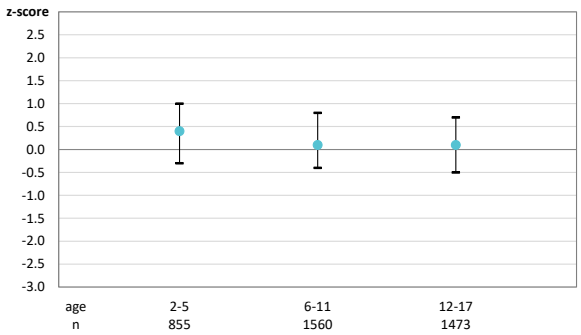


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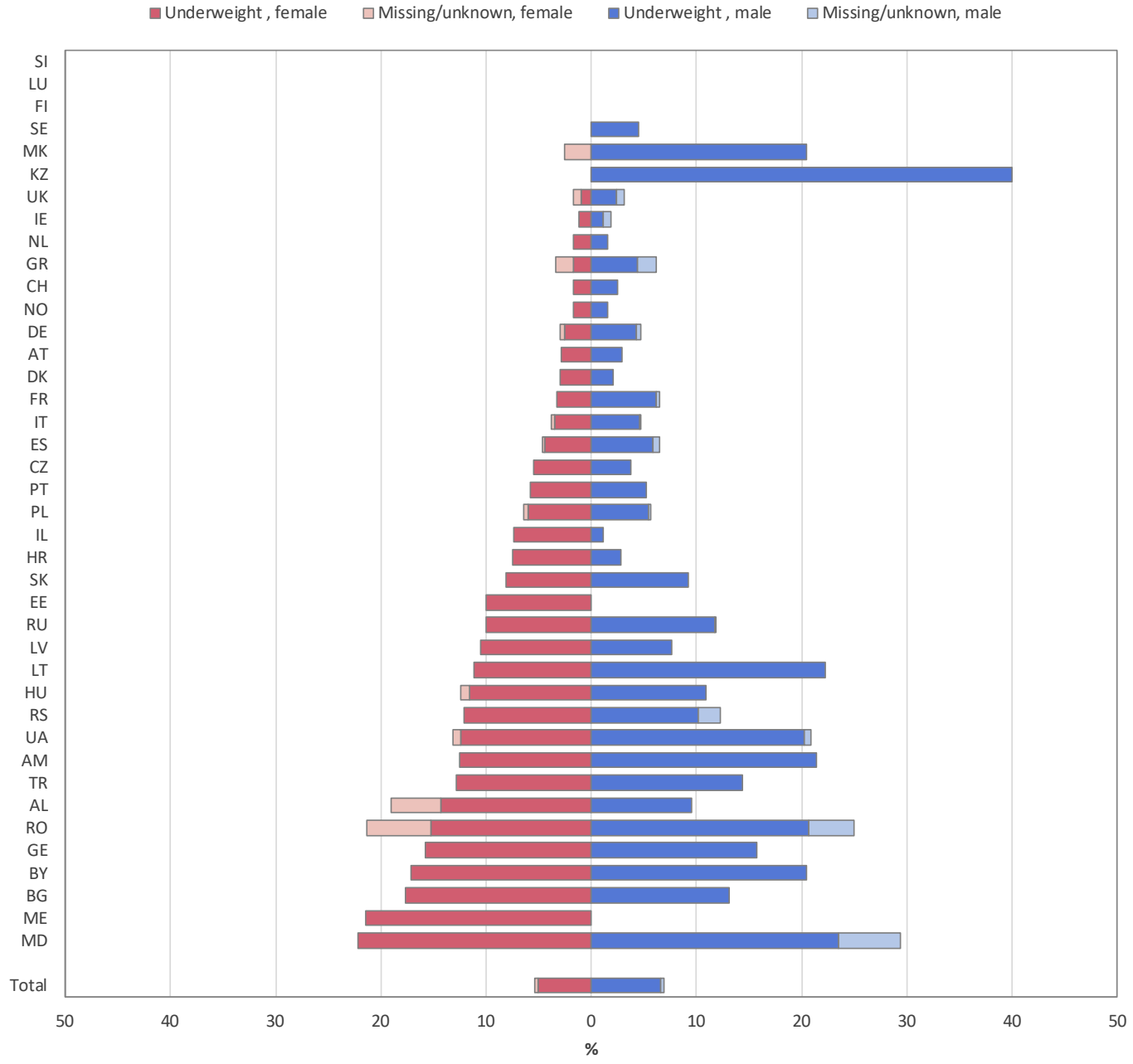
Figure 6.4 Being underweight is a clinical feature in children and adolescents with CF. There are considerable differences amongst the countries.

Proportion of children and adolescents with CF who are underweight (z-score of BMI <-2) by sex and by country, aged 2-17 years in 2023 who have never had a transplant.



Note: Cyprus and Iceland have been excluded from this graph because the number of children in one of the sex groups is less than 5.

The dark coloured bars (red for females, blue for males) represent the percentage of underweight children in each country. The lighter-coloured bars (light red for females, light blue for males) represent the percentage of missing values on BMI for children and adolescents in each country.

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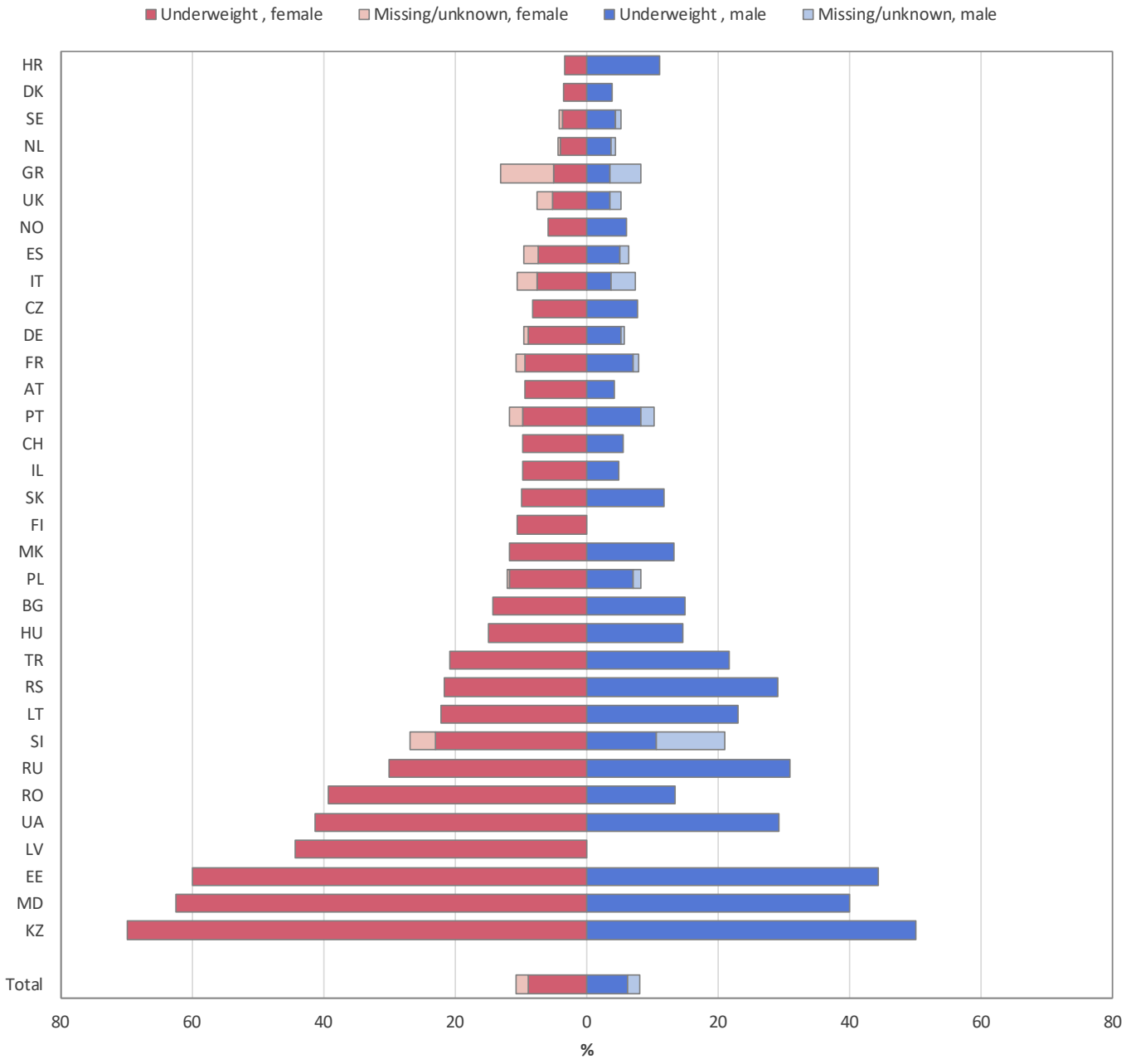
Figure 6.5 Being underweight is a clinical feature in adults with CF and more common in females. There are considerable differences amongst the countries.

Proportion of adults with BMI <18.5 by sex and by country, aged 18 years or older in 2023 who never had a transplant.



Note: We excluded from the graph the countries for which the information on underweight adults is missing for more than 10% of the individuals.

Albania, Armenia, Cyprus, Iceland, Luxembourg and Montenegro have been excluded from this graph because the number of adults in one of the sex groups is less than 5.

The dark coloured bars (red for females, blue for males) represent the percentage of underweight adults in each country. The light-coloured bars (light red for females, light blue for males) represent the percentage of missing values on BMI for adults in each country.

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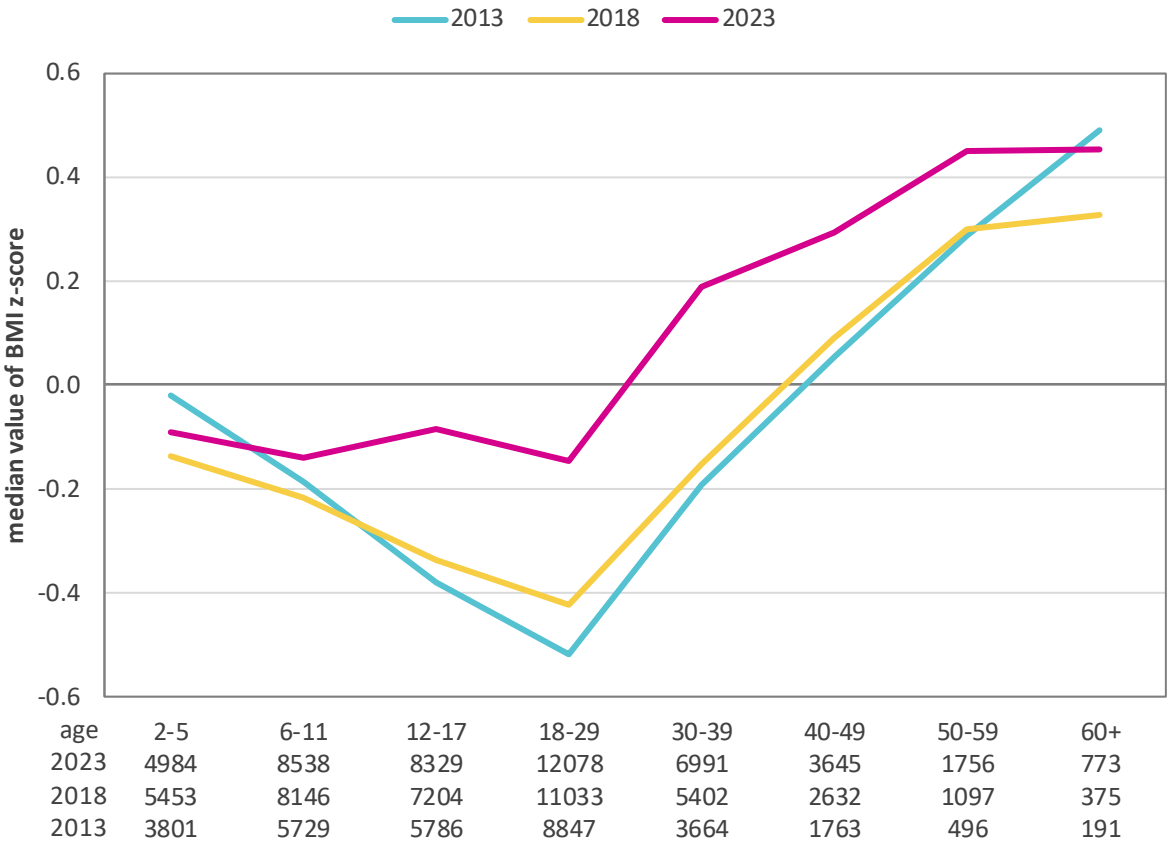
Figure 6.6 A significant improvement in BMI in 2023 from the age of 6 years is a reflection of the efficacy of CFTR modulator therapy in Europe.

Median z-score for BMI by age group in 2013, 2018 and 2023.



age	2-5	6-11	12-17	18-29	30-39	40-49	50-59	60+
2023	4984	8538	8329	12078	6991	3645	1756	773
2018	5453	8146	7204	11033	5402	2632	1097	375
2013	3801	5729	5786	8847	3664	1763	496	191

Note: Only people with CF aged 2 years or more at measurements and who have never had a lung or liver transplant.

In this graph we present data over time using cross sectional data per year of people with a confirmed CF. All people with CF alive, deceased, or not seen during the year of follow-up were included. Individuals who have had a transplant in their lifetime (lung and/or liver) were excluded.

7. Complications

Respiratory complications in CF include allergic bronchopulmonary aspergillosis, haemoptysis and pneumothorax. For pulmonary exacerbations, we present data on the number of people with CF who have had at least one pulmonary exacerbation treated with intravenous antibiotics, the number of exacerbation episodes and the number of days on IV antibiotics. In this chapter we also present statistics on gastro-intestinal complications such as distal intestinal obstruction syndrome (DIOS), salt loss syndrome (Pseudo Bartter Syndrome) and CF-related diabetes (CFRD). Data on liver disease are also included, despite the observation that the definitions for the types of liver disease may be interpreted differently from country to country and even from centre to centre within a country. Data on newly diagnosed malignancy are also reported in this section. Some of the tables do not show numbers by country, because for some countries the number of people with a particular complication is very low and could lead to people being identified.

The information in this section should not be considered complete for a number of reasons: national CF registries may use a different definition or different parameters for a complication; data about one or more of the complications are not collected; the status of a given complication is unknown.

In the tables we show the number of missing values for the various complications, whereas in the graphs we have included only countries where less than 10% of the data are missing. For a full list of complications and the definitions used by the ECFSPR please see Appendix 4.

7. Complications

Table 7.1 Prevalence in people with CF of at least 1 day on intravenous (IV) antibiotics (for CF-related reasons) at home and/or in hospital. People with CF seen in 2023, who have never had a transplant, by country and overall.

Country	Children (<18 years)						Adults (≥18 years)					
	Missing/ Unknown		No days on IV		Yes, at least one day on IV		Missing/ Unknown		No days on IV		Yes, at least one day on IV	
	N	%	N	%	N	%	N	%	N	%	N	%
Albania	0	0.0	35	70.0	15	30.0						
Armenia	0	0.0	16	61.5	10	38.5	0	0.0	5	83.3	1	16.7
Austria	0	0.0	304	84.9	54	15.1	0	0.0	350	86.6	54	13.4
Belarus	0	0.0	65	46.1	76	53.9	0	0.0	4	36.4	7	63.6
Bulgaria	0	0.0	103	76.3	32	23.7	0	0.0	83	88.3	11	11.7
Croatia	0	0.0	65	82.3	14	17.7	0	0.0	54	93.1	4	6.9
Cyprus	0	0.0	4	66.7	2	33.3	0	0.0	11	73.3	4	26.7
Czech Republic	1	0.3	323	92.3	26	7.4	8	2.5	273	86.7	34	10.8
Denmark	0	0.0	196	91.6	18	8.4	0	0.0	266	82.4	57	17.7
Estonia	0	0.0	17	68.0	8	32.0	4	22.2	7	38.9	7	38.9
Finland	0	0.0	27	90.0	3	10.0	0	0.0	31	86.1	5	13.9
France	6	0.2	2373	91.7	210	8.1	23	0.6	3374	86.4	509	13.0
Georgia	4	7.6	41	77.4	8	15.1						
Germany	0	0.0	2656	95.5	126	4.5	9	0.2	3644	90.6	369	9.2
Greece	9	3.8	198	83.9	29	12.3	13	3.4	298	78.8	67	17.7
Hungary	251	100.0	-	-	-	-	196	100.0	-	-	-	-
Iceland	0	0.0	5	55.6	4	44.4	0	0.0	5	71.4	2	28.6
Ireland	0	0.0	478	91.4	45	8.6	1	0.1	608	81.8	134	18.0
Israel	0	0.0	147	87.0	22	13.0	0	0.0	274	77.2	81	22.8
Italy	164	7.5	1681	76.8	343	15.7	489	13.4	2567	70.4	589	16.2
Kazakhstan	2	22.2	1	11.1	6	66.7	0	0.0	1	5.6	17	94.4
Latvia	0	0.0	29	80.6	7	19.4	0	0.0	9	52.9	8	47.1
Lithuania	0	0.0	10	50.0	10	50.0	4	15.4	7	26.9	15	57.7
Luxembourg	0	0.0	15	75.0	5	25.0	0	0.0	1	20.0	4	80.0
Rep of Moldova	1	3.2	13	41.9	17	54.8	0	0.0	3	20.0	12	80.0
Montenegro	0	0.0	23	79.3	6	20.7	0	0.0	10	90.9	1	9.1
The Netherlands	1	0.2	499	94.2	30	5.7	8	0.8	879	89.5	95	9.7
North Macedonia	5	6.0	33	39.3	46	54.8	2	3.8	14	26.4	37	69.8
Norway	0	0.0	131	95.6	6	4.4	1	0.5	178	86.4	27	13.1
Poland	44	4.5	746	75.6	197	20.0	5	0.9	422	72.9	152	26.3
Portugal	0	0.0	154	89.5	18	10.5	5	2.5	181	91.4	12	6.1
Romania	1	0.3	217	70.2	91	29.5	10	11.8	70	82.4	5	5.9
Russian Fed.	153	7.3	1226	58.7	711	34.0	192	19.6	238	24.3	548	56.0
Serbia	0	0.0	99	85.3	17	14.7	2	3.6	43	78.2	10	18.2
Slovak Republic	0	0.0	113	91.9	10	8.1	1	0.7	129	86.6	19	12.8
Slovenia	0	0.0	43	82.7	9	17.3	0	0.0	45	91.8	4	8.2
Spain	1	0.1	980	93.2	71	6.8	4	0.3	1162	90.5	118	9.2
Sweden	9	3.3	218	78.7	50	18.1	13	3.1	271	63.8	141	33.2
Switzerland	2	0.5	372	96.4	12	3.1	24	4.5	469	87.8	41	7.7
Türkiye	0	0.0	1824	80.9	430	19.1	1	0.2	351	73.9	123	25.9
Ukraine	4	1.2	81	24.7	243	74.1	5	5.0	12	11.9	84	83.2
United Kingdom	0	0.0	3425	83.5	675	16.5	0	0.0	4436	74.6	1510	25.4
Total	658	2.8	18986	81.3	3712	15.9	1020	3.8	20787	77.8	4920	18.4

Note: Albania and Georgia have <5 adults seen in 2023 and are excluded from the table for adults, but the people are included in the total number.

7. Complications

Table 7.2 Prevalence in people with CF of at least 1 day on IV antibiotics (for CF-related reasons) in hospital only. People with CF seen in 2023, who have never had a transplant, by country and overall.

Country	Children (<18 years)						Adults (≥18 years)					
	Missing/ Unknown		No days on IV		Yes, at least one day on IV		Missing/ Unknown		No days on IV		Yes, at least one day on IV	
	N	%	N	%	N	%	N	%	N	%	N	%
Albania	0	0.0	35	70.0	15	30.0						
Armenia	0	0.0	16	61.5	10	38.5	0	0.0	5	83.3	1	16.7
Austria	0	0.0	304	84.9	54	15.1	0	0.0	359	88.9	45	11.1
Belarus	0	0.0	65	46.1	76	53.9	0	0.0	4	36.4	7	63.6
Bulgaria	0	0.0	103	76.3	32	23.7	0	0.0	87	92.6	7	7.5
Croatia	0	0.0	65	82.3	14	17.7	0	0.0	54	93.1	4	6.9
Cyprus	0	0.0	4	66.7	2	33.3	0	0.0	11	73.3	4	26.7
Czech Republic	1	0.3	323	92.3	26	7.4	8	2.5	273	86.7	34	10.8
Denmark	65	30.4	145	67.8	4	1.9	323	100.0	0	0.0	0	0.0
Estonia	0	0.0	17	68.0	8	32.0	4	22.2	7	38.9	7	38.9
Finland	0	0.0	27	90.0	3	10.0	0	0.0	31	86.1	5	13.9
France	45	1.7	2379	91.9	165	6.4	242	6.2	3418	87.5	246	6.3
Georgia	4	7.6	42	79.3	7	13.2						
Germany	0	0.0	2664	95.8	118	4.2	13	0.3	3725	92.6	284	7.1
Greece	9	3.8	198	83.9	29	12.3	13	3.4	304	80.4	61	16.1
Hungary	0	0.0	227	90.4	24	9.6	0	0.0	114	58.2	82	41.8
Iceland	0	0.0	5	55.6	4	44.4	0	0.0	5	71.4	2	28.6
Ireland	0	0.0	486	92.9	37	7.1	1	0.1	624	84.0	118	15.9
Israel	0	0.0	150	88.8	19	11.2	0	0.0	309	87.0	46	13.0
Italy	165	7.5	1687	77.1	336	15.4	489	13.4	2601	71.4	555	15.2
Kazakhstan	2	22.2	1	11.1	6	66.7	0	0.0	4	22.2	14	77.8
Latvia	0	0.0	29	80.6	7	19.4	0	0.0	10	58.8	7	41.2
Lithuania	0	0.0	10	50.0	10	50.0	4	15.4	7	26.9	15	57.7
Luxembourg	0	0.0	16	80.0	4	20.0	0	0.0	1	20.0	4	80.0
Rep of Moldova	1	3.2	14	45.2	16	51.6	0	0.0	7	46.7	8	53.3
Montenegro	0	0.0	23	79.3	6	20.7	0	0.0	10	90.9	1	9.1
The Netherlands	0	0.0	505	95.3	25	4.7	1	0.1	893	90.9	88	9.0
North Macedonia	5	6.0	33	39.3	46	54.8	2	3.8	14	26.4	37	69.8
Norway	0	0.0	131	95.6	6	4.4	1	0.5	185	89.8	20	9.7
Poland	33	3.3	753	76.3	201	20.4	5	0.9	423	73.1	151	26.1
Portugal	0	0.0	155	90.1	17	9.9	5	2.5	182	91.9	11	5.6
Romania	1	0.3	219	70.9	89	28.8	10	11.8	71	83.5	4	4.7
Russian Fed.	159	7.6	1241	59.4	690	33.0	193	19.7	322	32.9	463	47.3
Serbia	0	0.0	99	85.3	17	14.7	2	3.6	43	78.2	10	18.2
Slovak Republic	0	0.0	113	91.9	10	8.1	1	0.7	130	87.3	18	12.1
Slovenia	0	0.0	43	82.7	9	17.3	0	0.0	45	91.8	4	8.2
Spain	1	0.1	982	93.4	69	6.6	4	0.3	1200	93.5	80	6.2
Sweden	9	3.3	250	90.3	18	6.5	12	2.8	383	90.1	30	7.1
Switzerland	2	0.5	372	96.4	12	3.1	24	4.5	481	90.1	29	5.4
Türkiye	0	0.0	1827	81.1	427	18.9	1	0.2	351	73.9	123	25.9
Ukraine	4	1.2	83	25.3	241	73.5	5	5.0	13	12.9	83	82.2
United Kingdom	0	0.0	3461	84.4	639	15.6	0	0.0	4768	80.2	1178	19.8
Total	506	2.2	19302	82.6	3548	15.2	1363	5.1	21476	80.4	3888	14.6

Note: Albania and Georgia have <5 adults seen in 2023 and are excluded from the table for adults, but the people are included in the total number.

7. Complications

Table 7.3 Prevalence in people with CF of at least 1 day in hospital, for any reason (routine check-up days not included). People with CF seen in 2023, who have never had a transplant, by country and overall.

Country	Children (<18 years)						Adults (≥18 years)					
	Missing/ Unknown		No days in hospital		Yes, at least one day in hospital		Missing/ Unknown		No days in hospital		Yes, at least one day in hospital	
	N	%	N	%	N	%	N	%	N	%	N	%
Albania	0	0.0	35	70.0	15	30.0						
Armenia	1	3.9	15	57.7	10	38.5	0	0.0	5	83.3	1	16.7
Austria	0	0.0	253	70.7	105	29.3	0	0.0	342	84.7	62	15.4
Belarus	0	0.0	65	46.1	76	53.9	0	0.0	4	36.4	7	63.6
Bulgaria	0	0.0	34	25.2	101	74.8	0	0.0	79	84.0	15	16.0
Croatia	0	0.0	60	76.0	19	24.1	0	0.0	54	93.1	4	6.9
Cyprus	0	0.0	4	66.7	2	33.3	0	0.0	10	66.7	5	33.3
Czech Republic	1	0.3	281	80.3	68	19.4	8	2.5	259	82.2	48	15.2
Denmark	0	0.0	198	92.5	16	7.5	0	0.0	260	80.5	63	19.5
Estonia	0	0.0	15	60.0	10	40.0	0	0.0	7	38.9	11	61.1
Finland	0	0.0	27	90.0	3	10.0	0	0.0	30	83.3	6	16.7
France	157	6.1	2046	79.0	386	14.9	344	8.8	3113	79.7	449	11.5
Georgia	4	7.6	42	79.3	7	13.2						
Germany	18	0.7	2229	80.1	535	19.2	48	1.2	3280	81.6	694	17.3
Greece	9	3.8	187	79.2	40	17.0	13	3.4	296	78.3	69	18.3
Hungary	0	0.0	169	67.3	82	32.7	0	0.0	109	55.6	87	44.4
Iceland	0	0.0	5	55.6	4	44.4	0	0.0	5	71.4	2	28.6
Ireland	0	0.0	470	89.9	53	10.1	1	0.1	584	78.6	158	21.3
Israel	0	0.0	142	84.0	27	16.0	2	0.6	292	82.3	61	17.2
Italy	0	0.0	1550	70.8	638	29.2	0	0.0	2703	74.2	942	25.8
Kazakhstan	2	22.2	1	11.1	6	66.7	0	0.0	4	22.2	14	77.8
Latvia	0	0.0	29	80.6	7	19.4	0	0.0	10	58.8	7	41.2
Lithuania	0	0.0	6	30.0	14	70.0	4	15.4	7	26.9	15	57.7
Luxembourg	0	0.0	15	75.0	5	25.0	0	0.0	1	20.0	4	80.0
Rep of Moldova	1	3.2	14	45.2	16	51.6	0	0.0	7	46.7	8	53.3
Montenegro	0	0.0	23	79.3	6	20.7	0	0.0	10	90.9	1	9.1
The Netherlands	0	0.0	463	87.4	67	12.6	0	0.0	816	83.1	166	16.9
North Macedonia	5	6.0	31	36.9	48	57.1	2	3.8	12	22.6	39	73.6
Norway	1	0.7	106	77.4	30	21.9	1	0.5	171	83.0	34	16.5
Poland	34	3.4	522	52.9	431	43.7	14	2.4	305	52.7	260	44.9
Portugal	0	0.0	155	90.1	17	9.9	5	2.5	175	88.4	18	9.1
Romania	0	0.0	109	35.3	200	64.7	10	11.8	61	71.8	14	16.5
Russian Fed.	87	4.2	1237	59.2	766	36.7	173	17.7	334	34.2	471	48.2
Serbia	0	0.0	97	83.6	19	16.4	1	1.8	43	78.2	11	20.0
Slovak Republic	0	0.0	109	88.6	14	11.4	1	0.7	126	84.6	22	14.8
Slovenia	0	0.0	38	73.1	14	26.9	0	0.0	43	87.8	6	12.2
Spain	2	0.2	962	91.4	88	8.4	4	0.3	1191	92.8	89	6.9
Sweden	0	0.0	243	87.7	34	12.3	1	0.2	368	86.6	56	13.2
Switzerland	2	0.5	363	94.0	21	5.4	24	4.5	475	89.0	35	6.6
Türkiye	0	0.0	1631	72.4	623	27.6	1	0.2	335	70.5	139	29.3
Ukraine	2	0.6	69	21.0	257	78.4	5	5.0	11	10.9	85	84.2
United Kingdom	0	0.0	3004	73.3	1096	26.7	0	0.0	4517	76.0	1429	24.0
Total	326	1.4	17054	73.0	5976	25.6	662	2.5	20456	76.5	5609	21.0

Note: Albania and Georgia have <5 adults seen in 2023 and are excluded from the table for adults, but the people are included in the total number.

7. Complications

Table 7.4 Prevalence of at least one pulmonary exacerbation treated with IV antibiotics in children (<18 years) with CF seen in 2023 who have never had a transplant, by country and overall.

Country	Missing/Unknown		No		Yes, at least one PEx treated with IV antibiotics	
	N	%	N	%	N	%
Albania	0	0.0	35	70.0	15	30.0
Armenia	2	7.7	16	61.5	8	30.8
Austria	8	2.2	329	91.9	21	5.9
Belarus	0	0.0	65	46.1	76	53.9
Bulgaria	0	0.0	101	74.8	34	25.2
Croatia	0	0.0	69	87.3	10	12.7
Cyprus	0	0.0	4	66.7	2	33.3
Czech Republic	0	0.0	335	95.7	15	4.3
Denmark	0	0.0	201	93.9	13	6.1
Estonia	0	0.0	17	68.0	8	32.0
Finland	0	0.0	27	90.0	3	10.0
France	2589	100.0	-	-	-	-
Georgia	11	20.8	33	62.3	9	17.0
Germany	5	0.2	2646	95.1	131	4.7
Greece	236	100.0	-	-	-	-
Hungary	0	0.0	194	77.3	57	22.7
Iceland	0	0.0	5	55.6	4	44.4
Ireland	0	0.0	485	92.7	38	7.3
Israel	7	4.1	143	84.6	19	11.2
Italy	165	7.5	1732	79.2	291	13.3
Kazakhstan	2	22.2	1	11.1	6	66.7
Latvia	1	2.8	28	77.8	7	19.4
Lithuania	0	0.0	10	50.0	10	50.0
Luxembourg	2	10.0	15	75.0	3	15.0
Rep of Moldova	1	3.2	11	35.5	19	61.3
Montenegro	0	0.0	23	79.3	6	20.7
The Netherlands	3	0.6	503	94.9	24	4.5
North Macedonia	3	3.6	34	40.5	47	56.0
Norway	0	0.0	131	95.6	6	4.4
Poland	45	4.6	751	76.1	191	19.4
Portugal	11	6.4	143	83.1	18	10.5
Romania	25	8.1	194	62.8	90	29.1
Russian Fed.	104	5.0	1249	59.8	737	35.3
Serbia	3	2.6	96	82.8	17	14.7
Slovak Republic	1	0.8	117	95.1	5	4.1
Slovenia	0	0.0	43	82.7	9	17.3
Spain	32	3.0	953	90.6	67	6.4
Sweden	0	0.0	226	81.6	51	18.4
Switzerland	0	0.0	378	97.9	8	2.1
Türkiye	0	0.0	1826	81.0	428	19.0
Ukraine	7	2.1	87	26.5	234	71.3
United Kingdom	0	0.0	3750	91.5	350	8.5
Total	3263	14.0	17006	72.8	3087	13.2

7. Complications

Table 7.5 Prevalence of at least one pulmonary exacerbation in adults (≥18 years) with CF seen in 2023 who have never had a transplant, by country and overall.

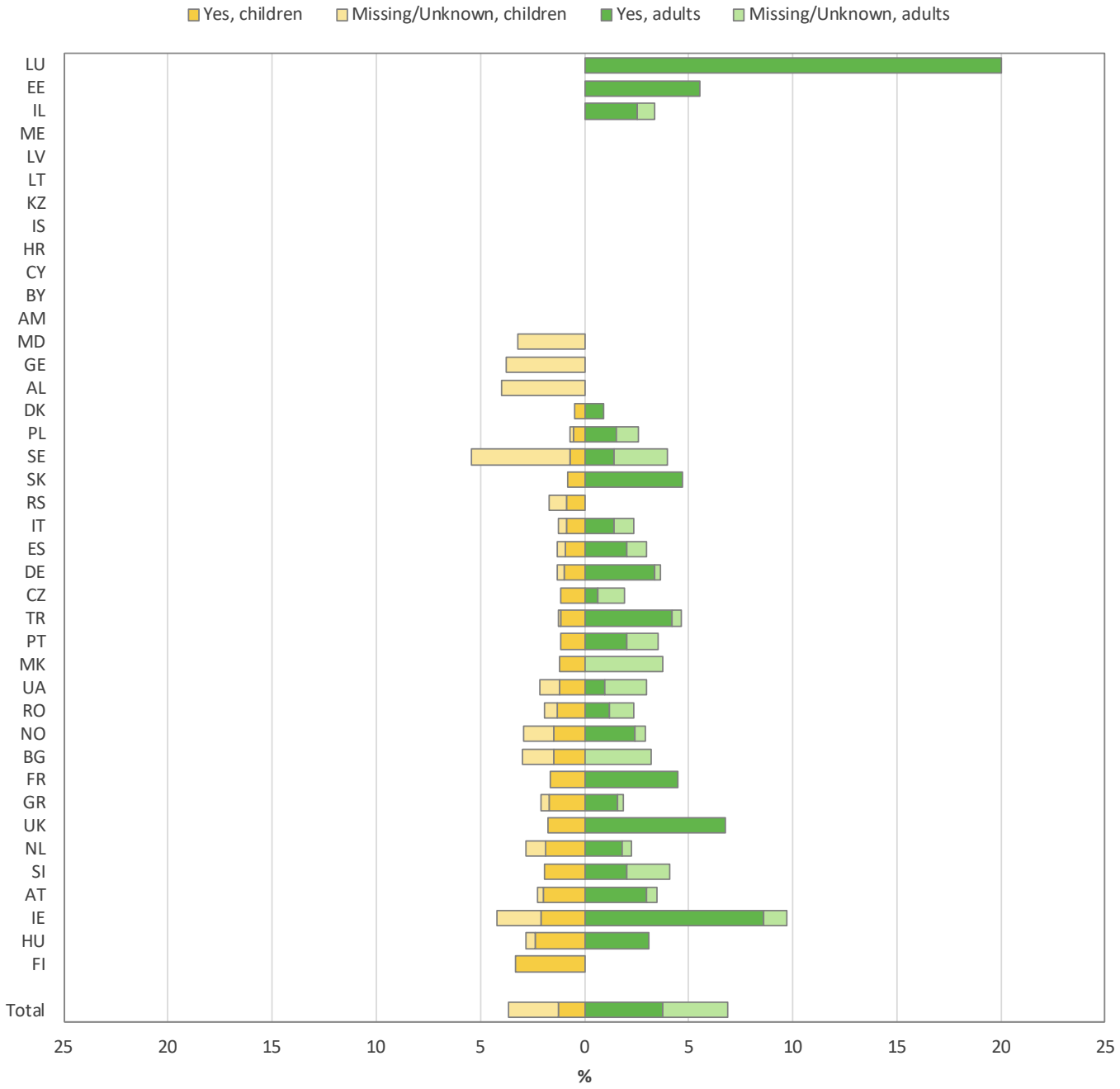
Country	Missing/Unknown		No		Yes, at least one PEx treated with IV antibiotics	
	N	%	N	%	N	%
Armenia	1	16.7	4	66.7	1	16.7
Austria	35	8.7	335	82.9	34	8.4
Belarus	0	0.0	4	36.4	7	63.6
Bulgaria	0	0.0	84	89.4	10	10.6
Croatia	1	1.7	53	91.4	4	6.9
Cyprus	0	0.0	11	73.3	4	26.7
Czech Republic	0	0.0	289	91.8	26	8.3
Denmark	0	0.0	272	84.2	51	15.8
Estonia	0	0.0	7	38.9	11	61.1
Finland	0	0.0	31	86.1	5	13.9
France	3906	100.0	-	-	-	-
Germany	14	0.4	3640	90.5	368	9.2
Greece	378	100.0	-	-	-	-
Hungary	0	0.0	109	55.6	87	44.4
Iceland	0	0.0	5	71.4	2	28.6
Ireland	0	0.0	616	82.9	127	17.1
Israel	10	2.8	266	74.9	79	22.3
Italy	489	13.4	2667	73.2	489	13.4
Kazakhstan	0	0.0	1	5.6	17	94.4
Latvia	0	0.0	9	52.9	8	47.1
Lithuania	0	0.0	11	42.3	15	57.7
Luxembourg	0	0.0	1	20.0	4	80.0
Rep of Moldova	0	0.0	3	20.0	12	80.0
Montenegro	0	0.0	10	90.9	1	9.1
The Netherlands	32	3.3	860	87.6	90	9.2
North Macedonia	2	3.8	14	26.4	37	69.8
Norway	0	0.0	179	86.9	27	13.1
Poland	26	4.5	404	69.8	149	25.7
Portugal	16	8.1	170	85.9	12	6.1
Romania	9	10.6	71	83.5	5	5.9
Russian Fed.	32	3.3	332	34.0	614	62.8
Serbia	1	1.8	44	80.0	10	18.2
Slovak Republic	8	5.4	123	82.6	18	12.1
Slovenia	1	2.0	44	89.8	4	8.2
Spain	22	1.7	1144	89.1	118	9.2
Sweden	1	0.2	288	67.8	136	32.0
Switzerland	0	0.0	494	92.5	40	7.5
Türkiye	1	0.2	351	73.9	123	25.9
Ukraine	1	1.0	17	16.8	83	82.2
United Kingdom	0	0.0	4563	76.7	1383	23.3
Total	4986	18.7	17528	65.6	4213	15.8

Note: Albania and Georgia have <5 adults seen in 2023 and are excluded from the table for adults, but the people are included in the total number.

7. Complications

Figure 7.1 ABPA prevalence is lower in children than in adults.

Prevalence of allergic bronchopulmonary aspergillosis in children and adults seen in 2023 who have never had a transplant, by country.



Note: We excluded from the graph the countries for which the information on allergic bronchopulmonary aspergillosis (ABPA) is missing for more than 10% of the children/adults with CF.

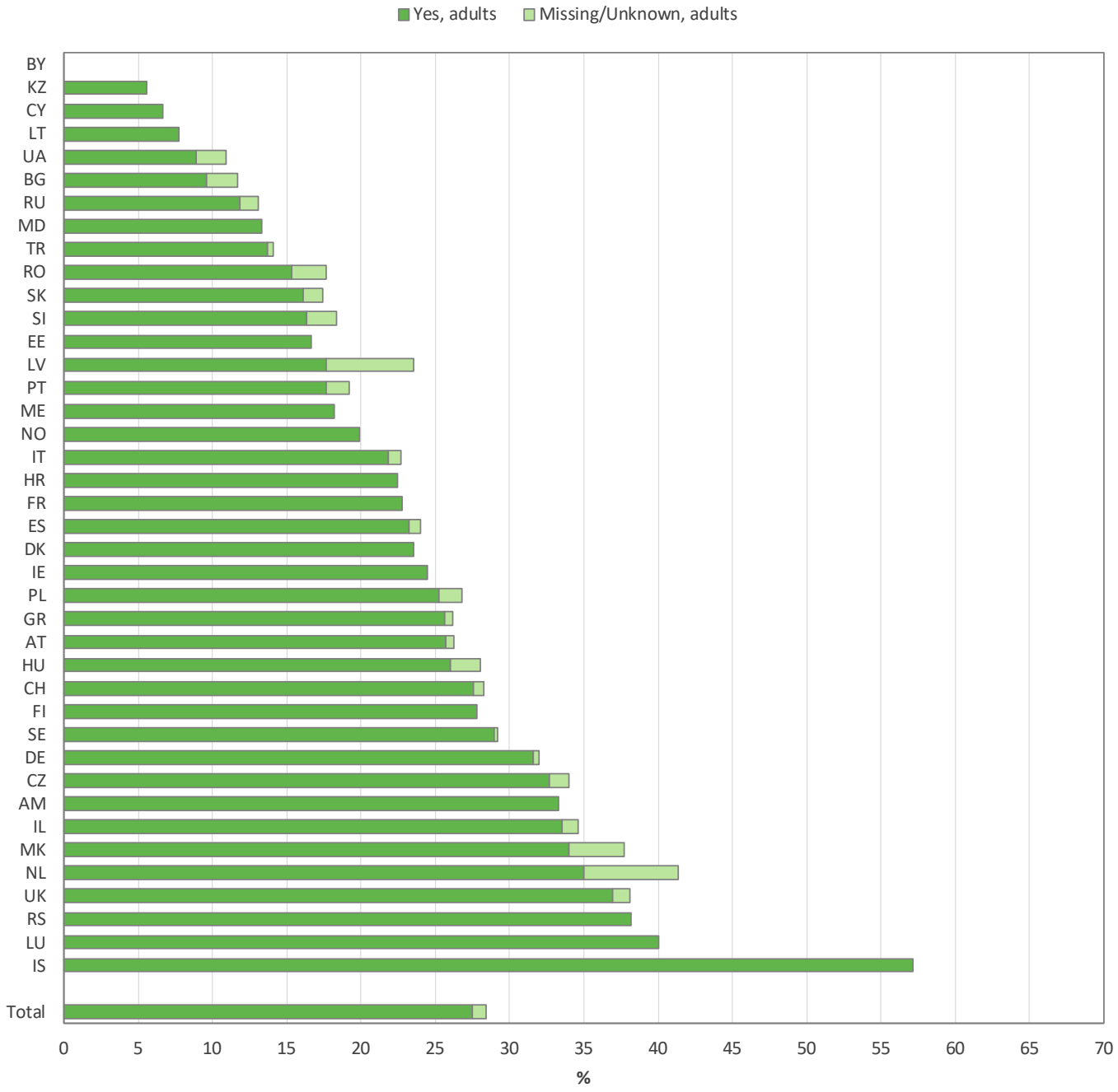
Albania and Georgia have <5 adults seen in 2023 and are excluded from the graph for adults.

This graph shows the frequency of allergic bronchopulmonary aspergillosis (ABPA) by country. For the definition of ABPA see Appendix 4. The dark colour shows the percentage of people with CF with ABPA, the light colours show the percentage of people with CF for whom this information is missing. ABPA is difficult to diagnose.

7. Complications

Figure 7.2 Important differences in the prevalence of CF-related diabetes in adults with CF throughout Europe might reflect genetic backgrounds but could also be linked to life expectancy.

Prevalence of CFRD, by country. All adults with CF seen in 2023 aged 18 years or older who have never had a transplant (table A7.1, Appendix 1).



Note: We excluded from the graph the countries for which the information on CFRD is missing for more than 10% of the adults.

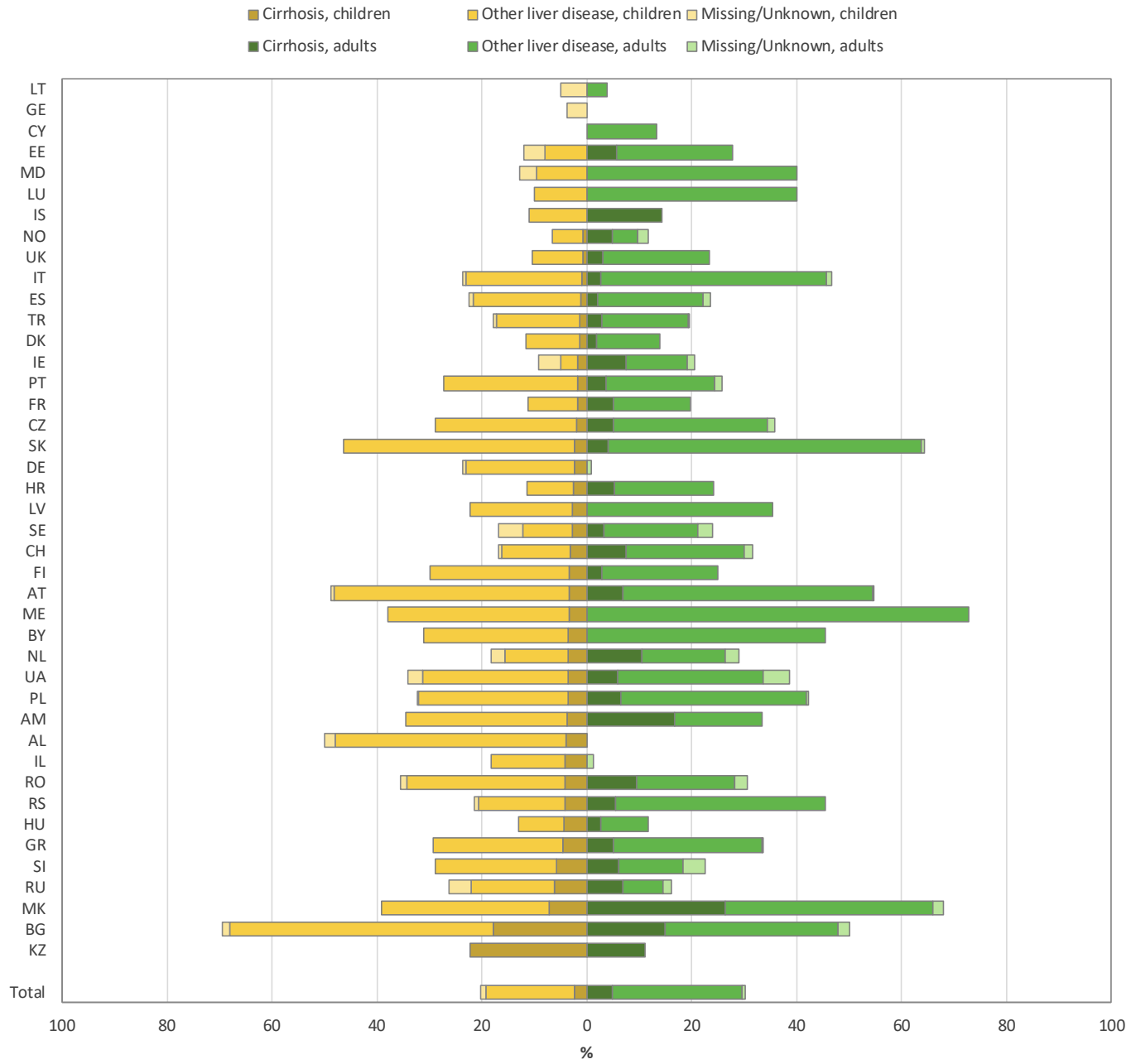
Albania and Georgia have <5 adults seen in 2023 and are excluded from the graph for adults.

This graph shows the prevalence of CF-related diabetes (CFRD), by country. The dark area of the bar shows the percentage of adults with CF who have CFRD, the lighter area shows the percentage of adults for whom this information is missing. Only people aged 18 years or older were included in this graph.

7. Complications

Figure 7.3 The prevalence of liver disease with or without cirrhosis is heterogenous across the countries.

Prevalence and severity of liver disease in children (<18 years) and adults (≥ 18 years) with CF seen in 2023 who have never had a transplant, by country (table A7.2, Appendix 1).



Note: We excluded from the graph the countries for which the information on liver disease is missing for more than 10% of the children/adults with CF. Albania and Georgia have <5 adults seen in 2023 and are excluded from the graph for adults.

Serbia: cirrhosis without portal hypertension/hypersplenism means ultrasound changes in liver tissue and/or abnormal liver function tests.

Figure 7.3 shows the frequency of liver disease by country. Liver disease is defined according to severity of portal hypertension (increased blood pressure in the liver vein, often resulting in blood shunting past the cirrhotic liver) divided into five categories, including no liver disease (see Appendix 4). This graph emphasises better than the tables (table A7.3 and table A7.4, Appendix 1) the vast differences in frequency and severity, which may be due to different interpretations of diagnostic results and differences in definitions.

7. Complications

Table 7.6 Prevalence of rare complications in all people with CF seen in 2023 who have never had a transplant.

Complication	Children (<18 years)						Adults (≥18 years)					
	Missing/ Unknown		No		Yes		Missing/ Unknown		No		Yes	
	N	%	N	%	N	%	N	%	N	%	N	%
Allergic bronchopulmonary aspergillosis	558	2.4	22503	96.4	295	1.3	826	3.1	24893	93.1	1008	3.8
Pneumothorax	155	0.7	23178	99.2	23	0.1	146	0.6	26523	99.2	58	0.2
Major haemoptysis (≥250 ml over the course of a day)	185	0.8	23127	99.0	44	0.2	210	0.8	26256	98.2	261	1.0
Malignancy newly diagnosed this year	186	0.8	23151	99.1	19	0.1	195	0.7	26356	98.6	176	0.7
Distal intestinal obstruction syndrome (DIOS)	139	0.6	22815	97.7	402	1.7	296	1.1	25752	96.4	679	2.5

Note: Germany and the United Kingdom define haemoptysis major > 240 ml.

Ireland: haemoptysis major is defined as haemoptysis massive > 240ml/day or > 100ml/day for several days.

Denmark only reported DIOS requiring hospitalisation.

Table 7.7 Type of malignancy newly diagnosed this year in people with CF seen in 2023 who have never had a transplant.

Country	Type of malignancy													
	Colorectal cancer		Small bowel cancer		Lymphoid leukaemia		Testicular cancer		Breast cancer		Thyroid gland cancer		Other or type unknown	
	N	%	N	%	N	%	N	%	N	%	N	%	N	%
Children	2	10.5	0	0.0	5	26.3	0	0.0	0	0.0	0	0.0	12	63.2
Adults	23	13.1	7	4.0	4	2.3	8	4.6	34	19.3	6	3.4	94	53.4

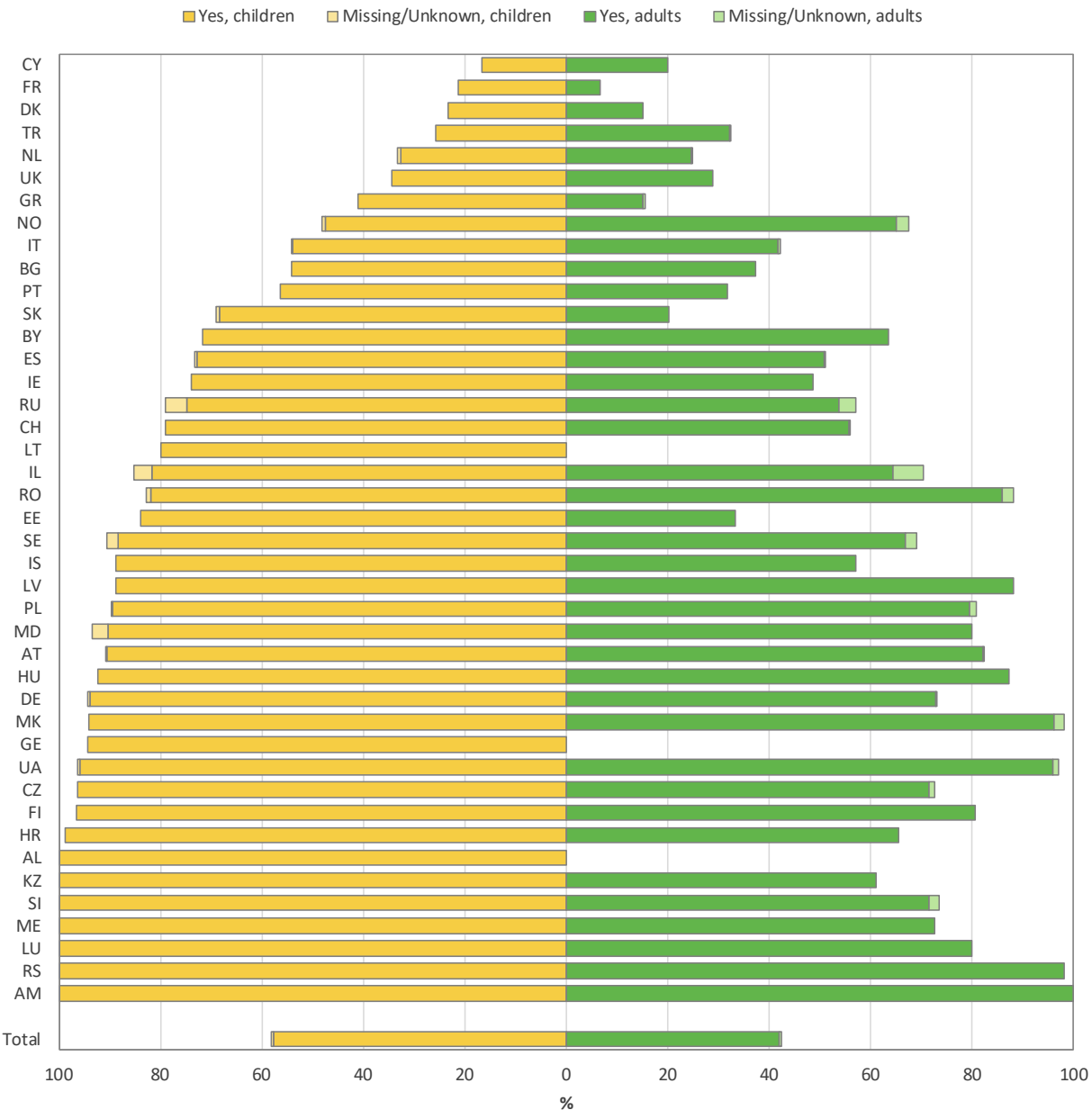
8. Therapies

In this chapter we report on the use of mucolytics (hypertonic saline, rhDNAse and mannitol), inhaled antibiotics, macrolides, bronchodilators and anti-inflammatories (inhaled and oral steroids). We also present data on the use of oxygen and non-invasive positive pressure ventilation. We collected information using the generic name of the medication, not the brand name.
The therapeutic options for the treatment of gastro-intestinal complications are limited; here we show the data on the use of ursodeoxycholic acid and proton pump inhibitors (PPI). We collected information using the generic name of the medications, not the brand name.
For a number or reasons, the information in this section should not be considered complete: national CF registries may use a different definition or different parameters for data about a therapy; data about one or more of the therapies are not collected; the use of a given therapy is unknown. In the tables we show the number of missing values for the various therapies, whereas in the graphs we have included only countries where less than 10% of the data are missing. For a full list of therapies and the definitions used by the ECFSPR about the data presented in this section please see Appendix 4.

8. Therapies

Figure 8.1 Variation in the use of inhaled hypertonic saline indicates both inequalities in availability and different therapeutic approaches.

Use of inhaled hypertonic saline in children and adults seen in 2023 who have never had a transplant, by country (table A8.1, Appendix 1).



Note: We excluded from the graph the countries for which the information on inhaled hypertonic saline is missing for more than 10% of the children/adults with CF. Albania and Georgia have <5 adults seen in 2023 and are excluded from the graph for adults.

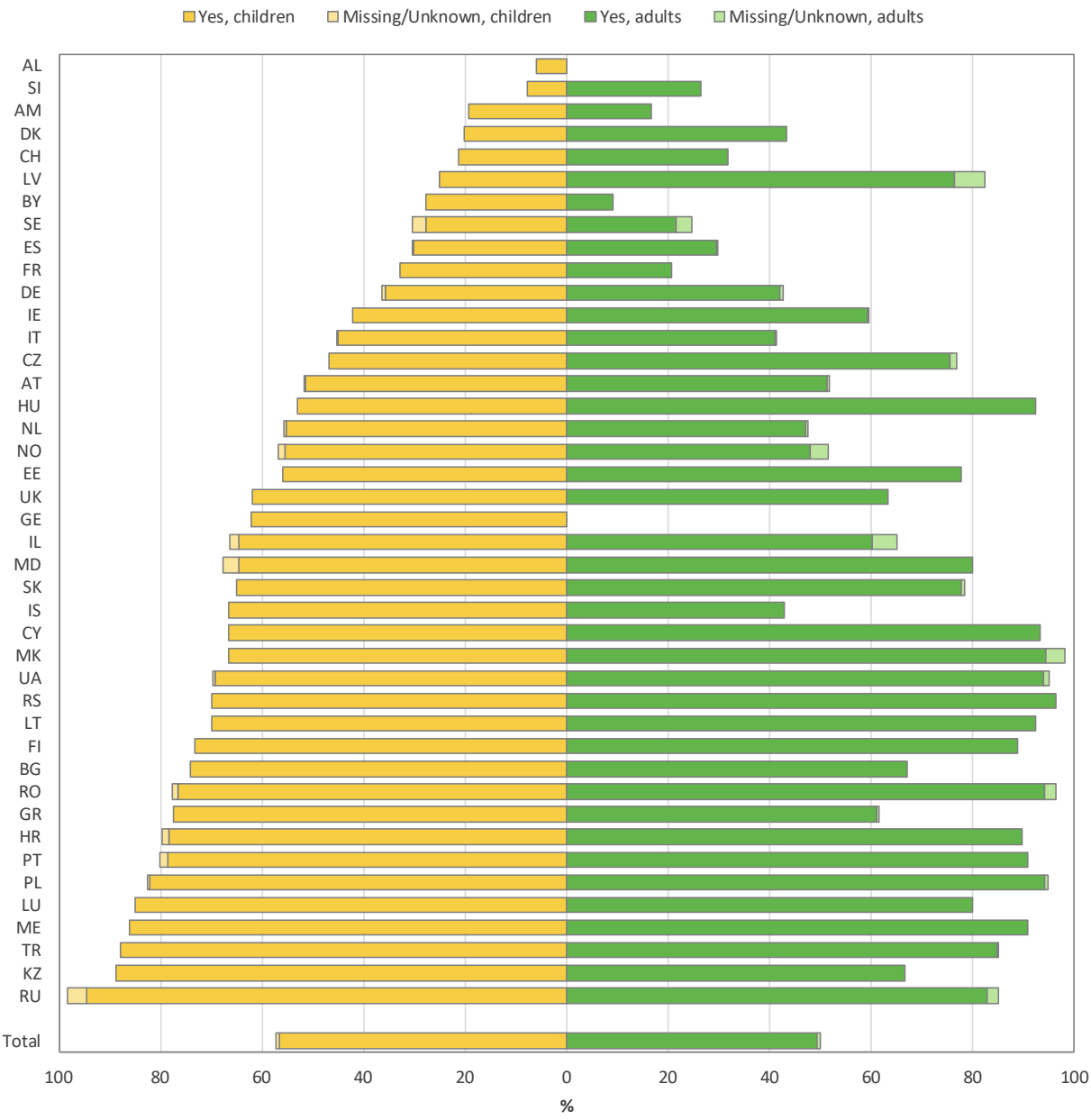
Inhaled hypertonic saline is reimbursed in most countries except in Albania, Armenia, Bulgaria, Estonia, Georgia, Kazakhstan, Lithuania, the Republic of Moldova, Poland, Romania. In Türkiye it is reimbursed for children ≥ 6 years.

This graph shows the use of inhaled hypertonic saline (≥ 3%) for at least three consecutive months during the survey year. The dark colours indicate the percentage of people with CF who took the medication, the lighter colours show the percentage of people with CF for whom this information is missing.

8. Therapies

Figure 8.2 Variation in the use of rhDNAse indicates both inequalities in availability and different therapeutic approaches.

Use of rhDNase in children and adults seen in 2023 who have never had a transplant, by country (table A8.2, Appendix 1).



Note: We excluded from the graph the countries for which the information on rhDNase is missing for more than 10% of the children/adults with CF. Albania and Georgia have <5 adults seen in 2023 and are excluded from the graph for adults.

Inhaled rhDNase is reimbursed in most countries except in Albania, Armenia and Belarus. It is reimbursed in Georgia for people with CF ≥ 2 years, in Bulgaria, Germany, Luxembourg, Macedonia, the Republic of Moldova, Norway, Romania, Spain, and the United Kingdom for individuals ≥ 5 years and in Latvia and Hungary for individuals ≥ 6 years.

This graph shows the use of rhDNase as inhalations for at least 3 consecutive months during the survey year. The dark coloured areas of the bar indicate the percentage of individuals with CF who took this medication, the lighter coloured areas show the percentage of individuals for whom this information is missing.

8. Therapies

Table 8.1 Use of inhaled mannitol for ≥3 consecutive months in all people with CF seen in 2023 who have never had a transplant, by country.

Country	Children (<18 years)						Adults (≥18 years)					
	Missing/ Unknown		No		Yes		Missing/ Unknown		No		Yes	
	N	%	N	%	N	%	N	%	N	%	N	%
Albania	0	0.0	50	100	0	0.0						
Armenia	0	0.0	22	84.6	4	15.4	0	0.0	6	100	0	0.0
Austria	5	1.4	353	98.6	0	0.0	1	0.3	397	98.3	6	1.5
Belarus	0	0.0	130	92.2	11	7.8	0	0.0	10	90.9	1	9.1
Bulgaria	0	0.0	135	100	0	0.0	0	0.0	94	100	0	0.0
Croatia	1	1.3	78	98.7	0	0.0	0	0.0	58	100	0	0.0
Cyprus	0	0.0	6	100	0	0.0	0	0.0	15	100	0	0.0
Czech Republic	0	0.0	350	100	0	0.0	4	1.3	307	97.5	4	1.3
Denmark	0	0.0	214	100	0	0.0	0	0.0	316	97.8	7	2.2
Estonia	0	0.0	25	100	0	0.0	0	0.0	18	100	0	0.0
Finland	0	0.0	30	100	0	0.0	0	0.0	36	100	0	0.0
France	2589	100	-	-	-	-	3906	100	-	-	-	-
Georgia	0	0.0	53	100	0	0.0						
Germany	21	0.8	2757	99.1	4	0.1	46	1.1	3832	95.3	144	3.6
Greece	1	0.4	234	99.2	1	0.4	2	0.5	368	97.4	8	2.1
Hungary	2	0.8	247	98.4	2	0.8	1	0.5	187	95.4	8	4.1
Iceland	0	0.0	9	100	0	0.0	0	0.0	7	100	0	0.0
Ireland	523	100	-	-	-	-	743	100	-	-	-	-
Israel	2	1.2	167	98.8	0	0.0	2	0.6	351	98.9	2	0.6
Italy	2	0.1	2176	99.5	10	0.5	10	0.3	3563	97.8	72	2.0
Kazakhstan	0	0.0	9	100	0	0.0	0	0.0	18	100	0	0.0
Latvia	0	0.0	36	100	0	0.0	0	0.0	17	100	0	0.0
Lithuania	0	0.0	20	100	0	0.0	0	0.0	26	100	0	0.0
Luxembourg	0	0.0	20	100	0	0.0	0	0.0	5	100	0	0.0
Rep of Moldova	1	3.2	30	96.8	0	0.0	0	0.0	15	100	0	0.0
Montenegro	0	0.0	29	100	0	0.0	0	0.0	11	100	0	0.0
The Netherlands	530	100	-	-	-	-	982	100	-	-	-	-
North Macedonia	0	0.0	84	100	0	0.0	1	1.9	52	98.1	0	0.0
Norway	0	0.0	136	99.3	1	0.7	2	1.0	202	98.1	2	1.0
Poland	1	0.1	986	99.9	0	0.0	7	1.2	572	98.8	0	0.0
Portugal	0	0.0	172	100	0	0.0	0	0.0	198	100	0	0.0
Romania	0	0.0	309	100	0	0.0	1	1.2	84	98.8	0	0.0
Russian Fed.	102	4.9	1772	84.8	216	10.3	17	1.7	843	86.2	118	12.1
Serbia	0	0.0	116	100	0	0.0	0	0.0	55	100	0	0.0
Slovak Republic	0	0.0	123	100	0	0.0	0	0.0	149	100	0	0.0
Slovenia	0	0.0	52	100	0	0.0	0	0.0	48	98.0	1	2.0
Spain	5	0.5	1046	99.4	1	0.1	4	0.3	1266	98.6	14	1.1
Sweden	7	2.5	269	97.1	1	0.4	11	2.6	413	97.2	1	0.2
Switzerland	0	0.0	386	100	0	0.0	0	0.0	534	100	0	0.0
Türkiye	0	0.0	2211	98.1	43	1.9	1	0.2	455	95.8	19	4.0
Ukraine	3	0.9	323	98.5	2	0.6	2	2.0	99	98.0	0	0.0
United Kingdom	0	0.0	4096	99.9	4	0.1	0	0.0	5726	96.3	220	3.7

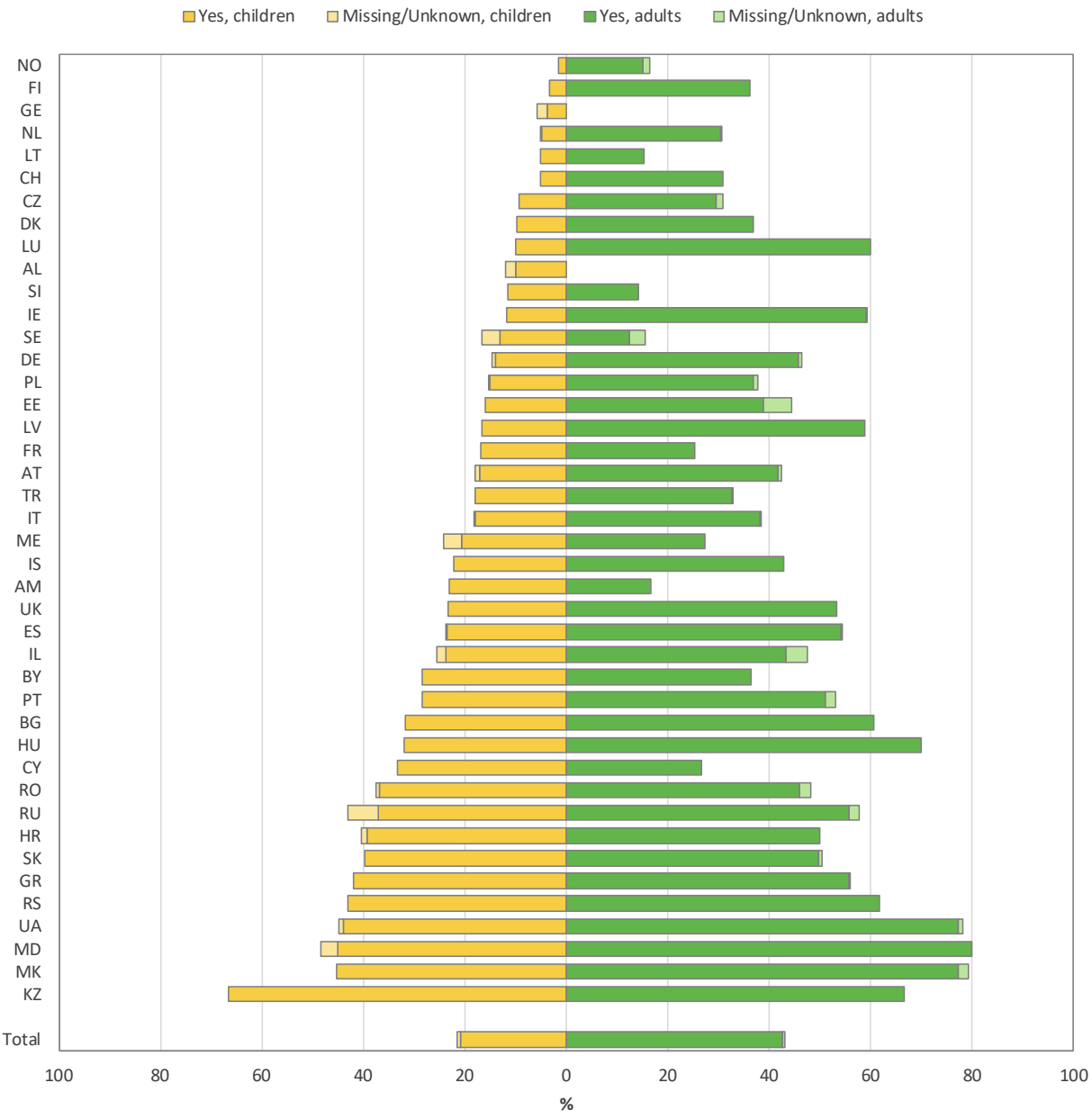
Note: For inhaled mannitol the total percentage of missing information is higher than 10% therefore the totals are excluded from the table. Albania and Georgia have <5 adults seen in 2023 and are excluded from the table for adults.

Inhaled mannitol is reimbursed for all people with CF in Austria, Czech Republic, Denmark, Norway, the Russian Federation (depending on the region of residence), Slovenia and Spain. It is reimbursed for people with CF ≥ 18 years in France, Germany, Greece, Italy, and the United Kingdom and it is reimbursed for people with CF ≥ 6 years in Switzerland and for people with CF > 6 years in Türkiye. It is not reimbursed in the other countries.

8. Therapies

Figure 8.3 Inhaled antibiotics are still an important therapeutic strategy in the prevention of pulmonary exacerbations, especially in adults with CF.

Use of inhaled antibiotics in children and adults seen in 2023 who have never had a transplant, by country (table A8.3, Appendix 1).



Note: We excluded from the graph the countries for which the information on inhaled antibiotics is missing for more than 10% of the children/adults with CF. Albania and Georgia have <5 adults seen in 2023 and are excluded from the graph for adults.

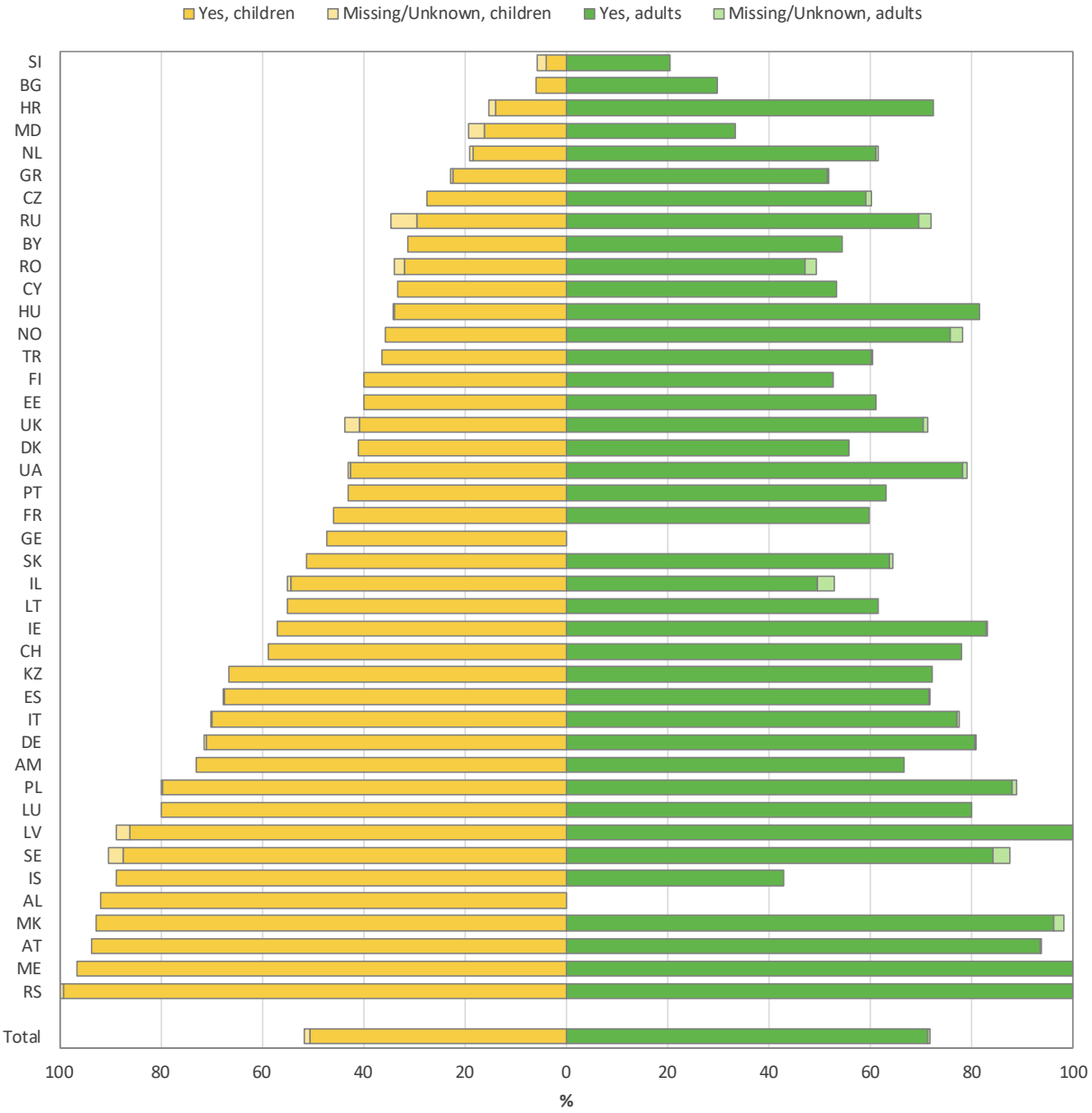
Inhaled antibiotics are reimbursed in all countries except Armenia and Georgia. In Bulgaria, colistin is reimbursed for all, tobramycin for > 7years, and levofloxacin for > 18 years. In Estonia, tobramycin and colistin are reimbursed and in Romania,only tobramycin solution and colistin dry powder are reimbursed for ≥ 6 years.

This graph shows the use of inhaled antibiotics (of any kind) for at least three months (consecutively or cyclic therapy) during the survey year. The dark area of the bar shows the percentage of people with CF who took inhaled antibiotics, the lighter area shows the percentage of people with CF for whom this information is missing.

8. Therapies

Figure 8.4 Bronchodilators (both short and long acting) are used as widespread supportive treatment in many countries in Europe.

Use of bronchodilators (short- or long-acting) in children and adults seen in 2023 who have never had a transplant, by country (table A8.4 and A8.5, Appendix 1).



Note: We excluded from the graph the countries for which the information on the use of bronchodilators is missing for more than 10% of the children/adults with CF. Albania and Georgia have <5 adults seen in 2023 and are excluded from the graph for adults.

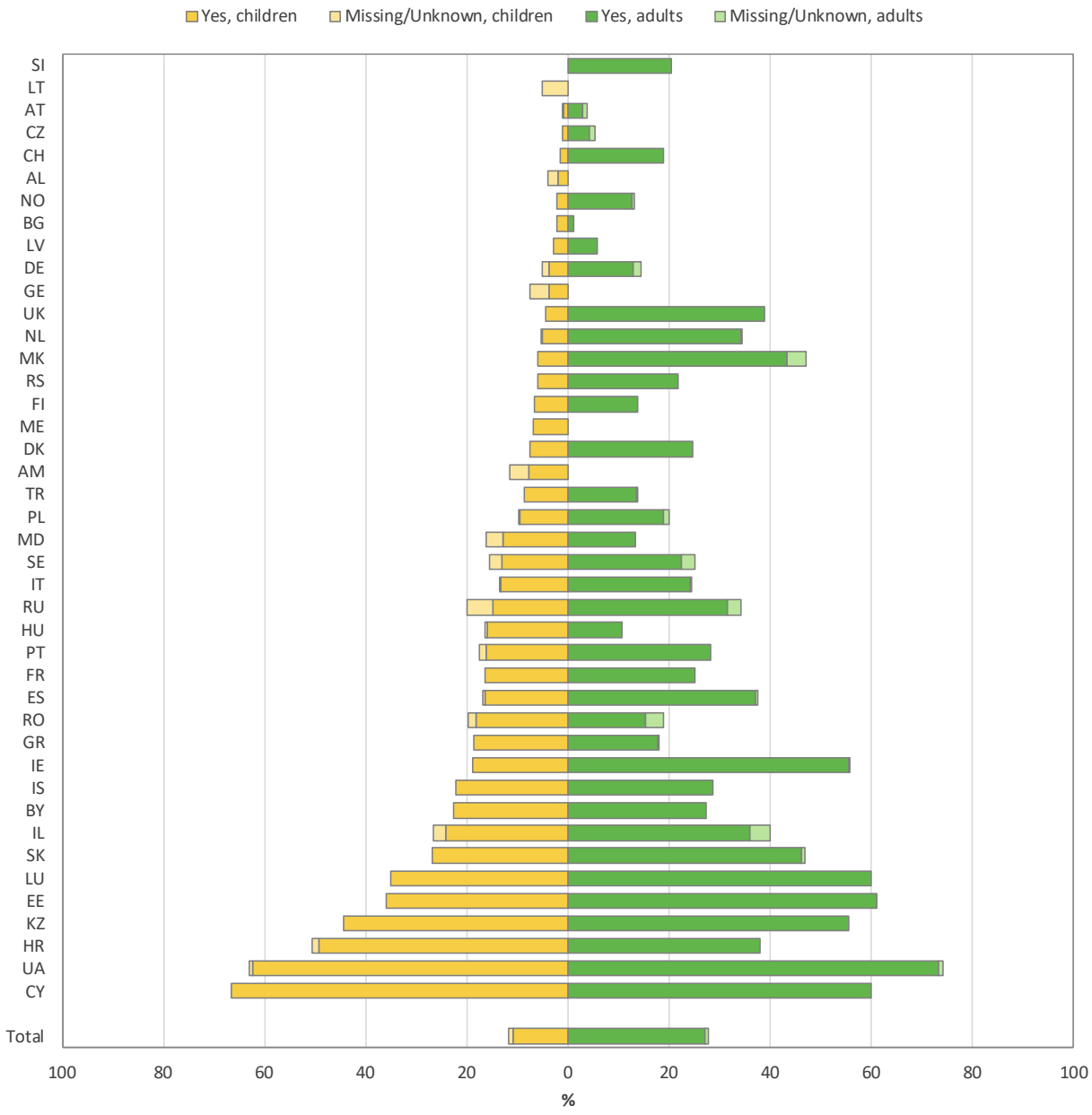
Inhaled bronchodilators are reimbursed in most countries except in Bulgaria, Georgia, the Republic of Moldova (reimbursed for people diagnosed with asthma), Poland and Serbia. In Kazakhstan it is subject to the availability of the regional budget. In Estonia long- and short-acting bronchodilators are reimbursed for people with CF diagnosed with asthma.

This graph shows the use of bronchodilators, both long-acting and short-acting, for at least three consecutive months during the survey year. This is the most widely used inhaled medication but there are significant differences in frequency of use in the countries. The dark area of the bars indicates the percentage of people with CF who took bronchodilators, the lighter area shows the percentage of people with CF for whom this information is missing.

8. Therapies

Figure 8.5 Azithromycin is used as an antibiotic and anti-inflammatory mediator throughout Europe, mostly by adults with CF.

Use of macrolides in children and adults seen in 2023 who have never had a transplant, by country (table A8.6, Appendix 1).



Note: We excluded from the graph the countries for which the information on the use of macrolides is missing for more than 10% of the children/adults with CF. Albania and Georgia have <5 adults seen in 2023 and are excluded from the graph for adults.

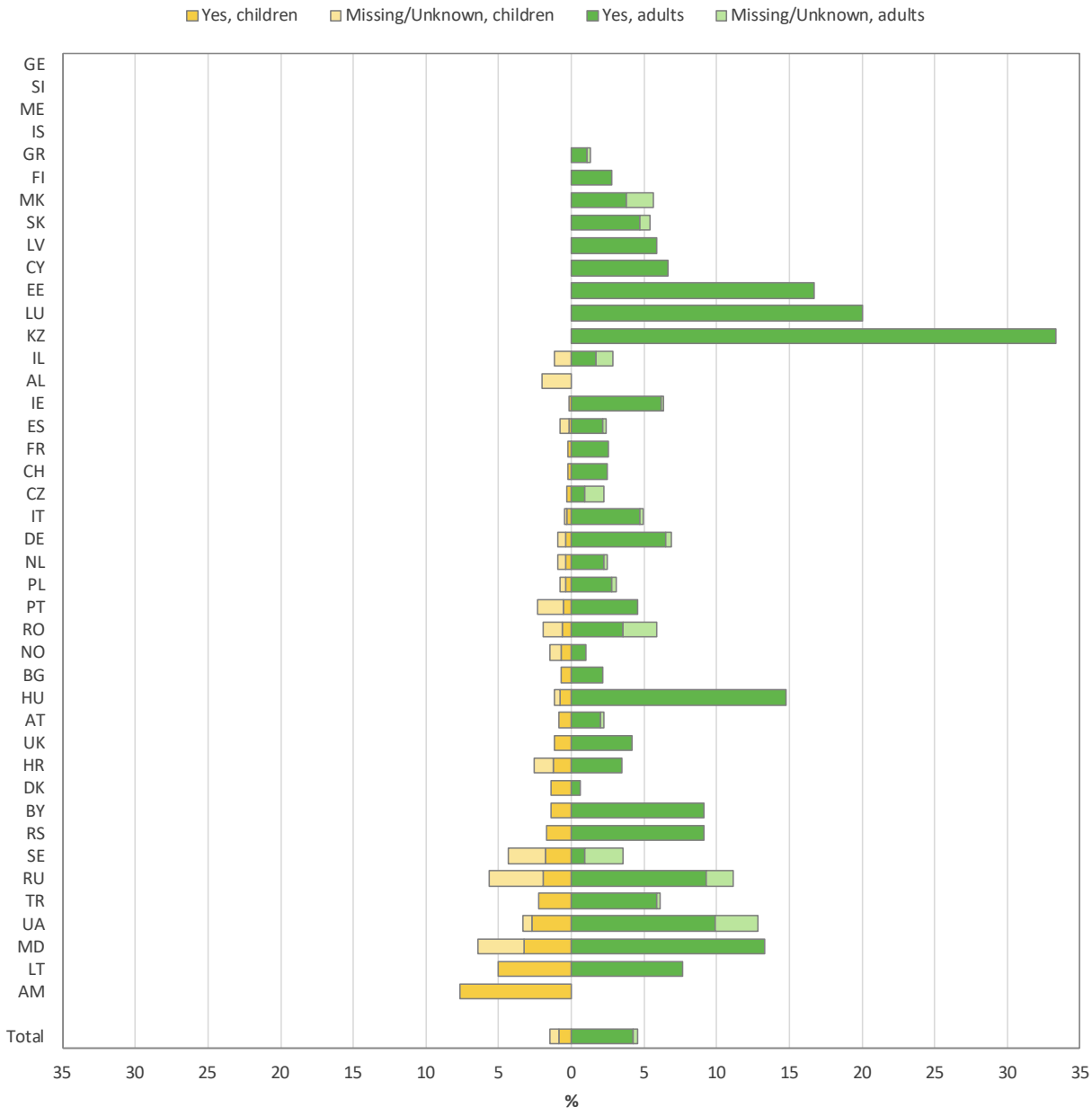
Oral macrolides are reimbursed in most countries except in Bulgaria, Georgia, Kazakhstan and Serbia. In Armenia, they are reimbursed for some outpatients.

This graph shows the use of macrolides (e.g. azithromycin or another macrolide) for at least 3 consecutive months during 2023. Macrolides are antibiotics but when taken continuously, they can also modulate the immune system, probably due to their anti-inflammatory properties. Clinical studies have shown that people with chronic *Pseudomonas aeruginosa* infection benefit from continuous azithromycin treatment with regard to lung function and pulmonary exacerbation rates. The dark area of the bar indicates the percentage of people with CF taking this medication, the lighter area shows the percentage of people with CF for whom this information is missing.

8. Therapies

Figure 8.6 Oxygen treatment is an indicator of severe lung disease, mostly seen in the adult population.

Use of oxygen in children and adults seen in 2023 who have never had a transplant, by country (table A8.7, Appendix 1).



Note: We excluded from the graph the countries for which the information on the use of oxygen is missing for more than 10% of the children/adults with CF. Albania and Georgia have <5 adults seen in 2023 and are excluded from the graph for adults.

Oxygen therapy is reimbursed in most countries except in Bulgaria and the Republic of Moldova. In Armenia and Georgia it is only reimbursed if the individual is hospitalised; in Serbia therapy at home is reimbursed.

This graph shows the use of oxygen for at least 3 months, not necessarily consecutive, during 2023. Oxygen therapy is used for severe lung disease. The dark area of the bar indicates the percentage of people with CF who used supplementary oxygen, the lighter area shows the percentage of people for whom this information is missing.

8. Therapies

Table 8.2 Use of non-invasive positive pressure ventilation (NIPPV) for ≥3 consecutive months in all people with CF seen in 2023 who have never had a transplant, by country.

Country	Children (<18 years)								Adults (≥18 years)							
	Missing/ Unknown		No		Yes, BiPAP (Bilevel Positive Airways Pressure)		Yes, CPAP (Continuous Positive Airways Pressure)		Missing/ Unknown		No		Yes, BiPAP (Bilevel Positive Airways Pressure)		Yes, CPAP (Continuous Positive Airways Pressure)	
	N	%	N	%	N	%	N	%	N	%	N	%	N	%	N	%
Albania	0	0.0	50	100	0	0.0	0	0								
Armenia	0	0.0	26	100	0	0.0	0	0.0	0	0	6	100	0	0.0	0	0.0
Austria	0	0.0	358	100	0	0.0	0	0.0	4	1.0	400	99.0	0	0.0	0	0.0
Belarus	0	0.0	140	99.3	0	0.0	1	0.7	0	0.0	10	90.9	0	0.0	1	9.1
Bulgaria	0	0.0	135	100	0	0.0	0	0.0	2	2.1	91	96.8	0	0.0	1	1.1
Croatia	1	1.3	77	97.5	1	1.3	0	0.0	0	0.0	58	100	0	0.0	0	0.0
Cyprus	0	0.0	6	100	0	0.0	0	0.0	0	0.0	15	100	0	0.0	0	0.0
Czech Rep.	0	0.0	349	99.7	0	0.0	1	0.3	4	1.3	311	98.7	0	0.0	0	0.0
Denmark	0	0.0	214	100	0	0.0	0	0.0	0	0.0	321	99.4	0	0.0	2	0.6
Estonia	0	0.0	25	100	0	0.0	0	0.0	0	0.0	16	88.9	2	11.1	0	0.0
Finland	0	0.0	30	100	0	0.0	0	0.0	0	0.0	35	97.2	0	0.0	1	2.8
France	2589	100	-	-	-	-	-	-	3906	100	-	-	-	-	-	-
Georgia	0	0.0	53	100	0	0.0	0	0.0								
Germany	16	0.6	2763	99.3	2	0.1	1	0.0	37	0.9	3944	98.1	25	0.6	16	0.4
Greece	0	0.0	236	100	0	0.0	0	0.0	1	0.3	377	99.7	0	0.0	0	0.0
Hungary	0	0.0	250	99.6	1	0.4	0	0.0	2	1.0	188	95.9	6	3.1	0	0.0
Iceland	0	0.0	9	100	0	0.0	0	0.0	0	0.0	6	85.7	1	14.3	0	0.0
Ireland	0	0.0	519	99.2	2	0.4	2	0.4	1	0.1	690	92.9	52	7.0	0	0.0
Israel	1	0.6	167	98.8	1	0.6	0	0.0	2	0.6	346	97.5	6	1.7	1	0.3
Italy	168	7.7	1984	90.7	5	0.2	31	1.4	500	13.7	3078	84.4	34	0.9	33	0.9
Kazakhstan	0	0.0	9	100	0	0.0	0	0.0	0	0.0	18	100	0	0.0	0	0.0
Latvia	0	0.0	36	100	0	0.0	0	0.0	1	5.9	16	94.1	0	0.0	0	0.0
Lithuania	0	0.0	20	100	0	0.0	0	0.0	0	0.0	25	96.2	1	3.9	0	0.0
Luxembourg	0	0.0	20	100	0	0.0	0	0.0	0	0.0	4	80.0	1	20.0	0	0.0
Rep. Moldova	1	3.2	30	96.8	0	0.0	0	0.0	0	0.0	15	100	0	0.0	0	0.0
Montenegro	0	0.0	29	100	0	0.0	0	0.0	0	0.0	11	100	0	0.0	0	0.0
Netherlands	1	0.2	528	99.6	0	0.0	1	0.2	6	0.6	972	99.0	3	0.3	1	0.1
N. Macedonia	0	0.0	84	100	0	0.0	0	0.0	1	1.9	52	98.1	0	0.0	0	0.0
Norway	0	0.0	137	100	0	0.0	0	0.0	1	0.5	205	99.5	0	0.0	0	0.0
Poland	4	0.4	981	99.4	2	0.2	0	0.0	5	0.9	566	97.8	8	1.4	0	0.0
Portugal	0	0.0	171	99.4	1	0.6	0	0.0	2	1.0	192	97.0	3	1.5	1	0.5
Romania	6	1.9	303	98.1	0	0.0	0	0.0	3	3.5	82	96.5	0	0.0	0	0.0
Russian Fed.	76	3.6	2005	95.9	0	0.0	9	0.4	12	1.2	947	96.8	10	1.0	9	0.9
Serbia	0	0.0	114	98.3	2	1.7	0	0.0	0	0.0	55	100	0	0.0	0	0.0
Slovak Rep.	0	0.0	123	100	0	0.0	0	0.0	0	0.0	147	98.7	2	1.3	0	0.0
Slovenia	0	0.0	52	100	0	0.0	0	0.0	1	2.0	48	98.0	0	0.0	0	0.0
Spain	6	0.6	1046	99.4	0	0.0	0	0.0	4	0.3	1273	99.1	3	0.2	4	0.3
Sweden	9	3.3	267	96.4	1	0.4	0	0.0	16	3.8	406	95.5	3	0.7	0	0.0
Switzerland	0	0.0	385	99.7	1	0.3	0	0.0	0	0.0	533	99.8	0	0.0	1	0.2
Türkiye	0	0.0	2208	98.0	45	2.0	1	0.0	1	0.2	454	95.6	20	4.2	0	0.0
Ukraine	2	0.6	325	99.1	0	0.0	1	0.3	1	1.0	100	99.0	0	0.0	0	0.0
United Kingdom	0	0.0	4080	99.5	0	0.0	20	0.5	0	0.0	5848	98.4	0	0.0	98	1.7

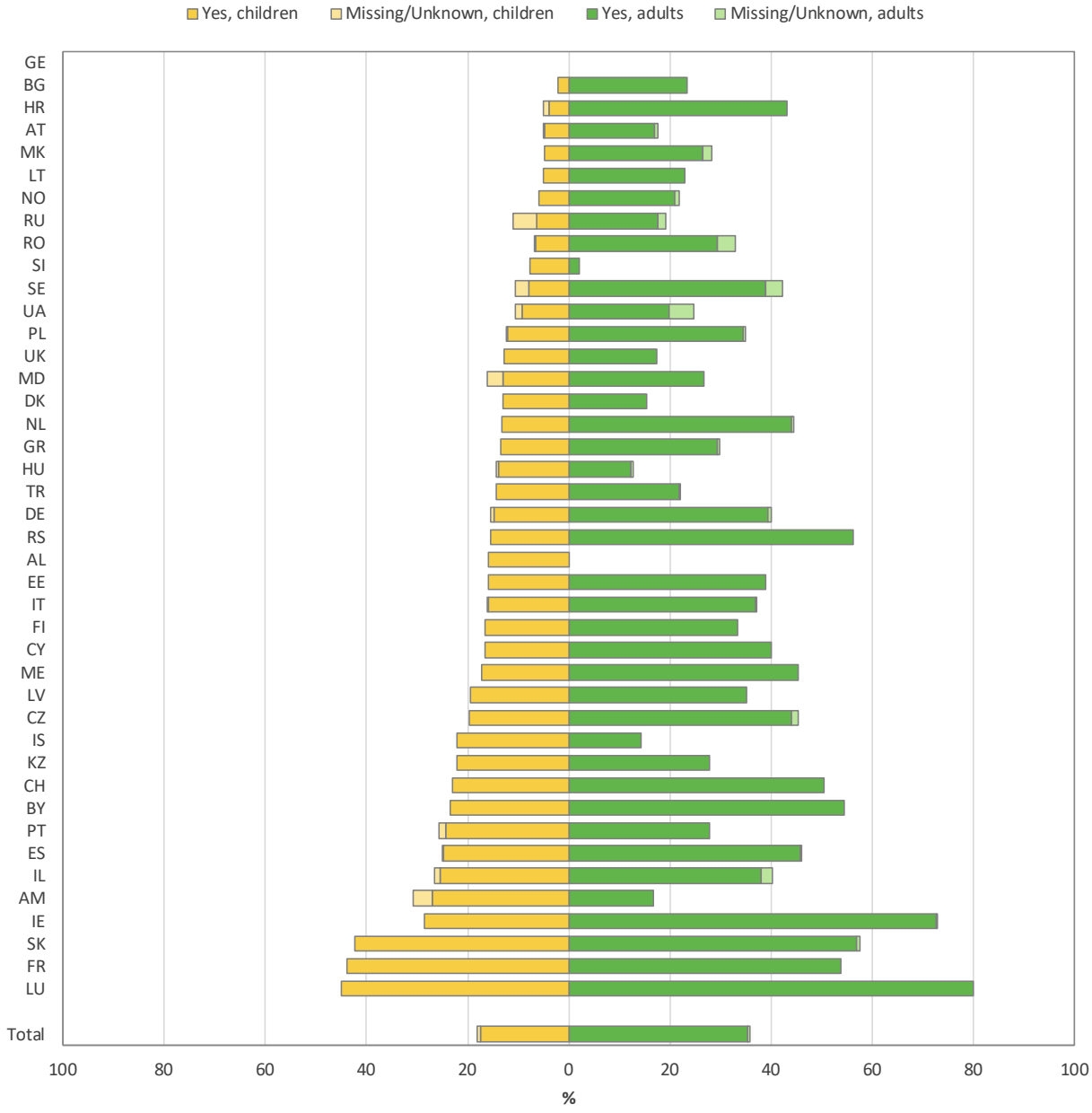
Note: For non-invasive positive pressure ventilation (NIPPV) the total percentage of missing information is higher than 10%, therefore the totals are excluded from the table. Albania and Georgia have <5 adults seen in 2023 and are excluded from the table for adults.

NIPPV is reimbursed in most countries except in Albania, Armenia, Belarus, Bulgaria, Kazakhstan, the Republic of Moldova and Ukraine. In Georgia it is reimbursed if the individual is hospitalised.

8. Therapies

Figure 8.7 Pulmonary inflammation, including obstructive symptoms, is often treated with corticosteroids.

Use of inhaled steroids in children and adults seen in 2023 who have never had a transplant, by country (table A8.8, Appendix 1).



Note: We excluded from the graph the countries for which the information on use of inhaled steroids is missing for more than 10% of the children/adults with CF. Albania and Georgia have <5 adults seen in 2023 and are excluded from the graph for adults.

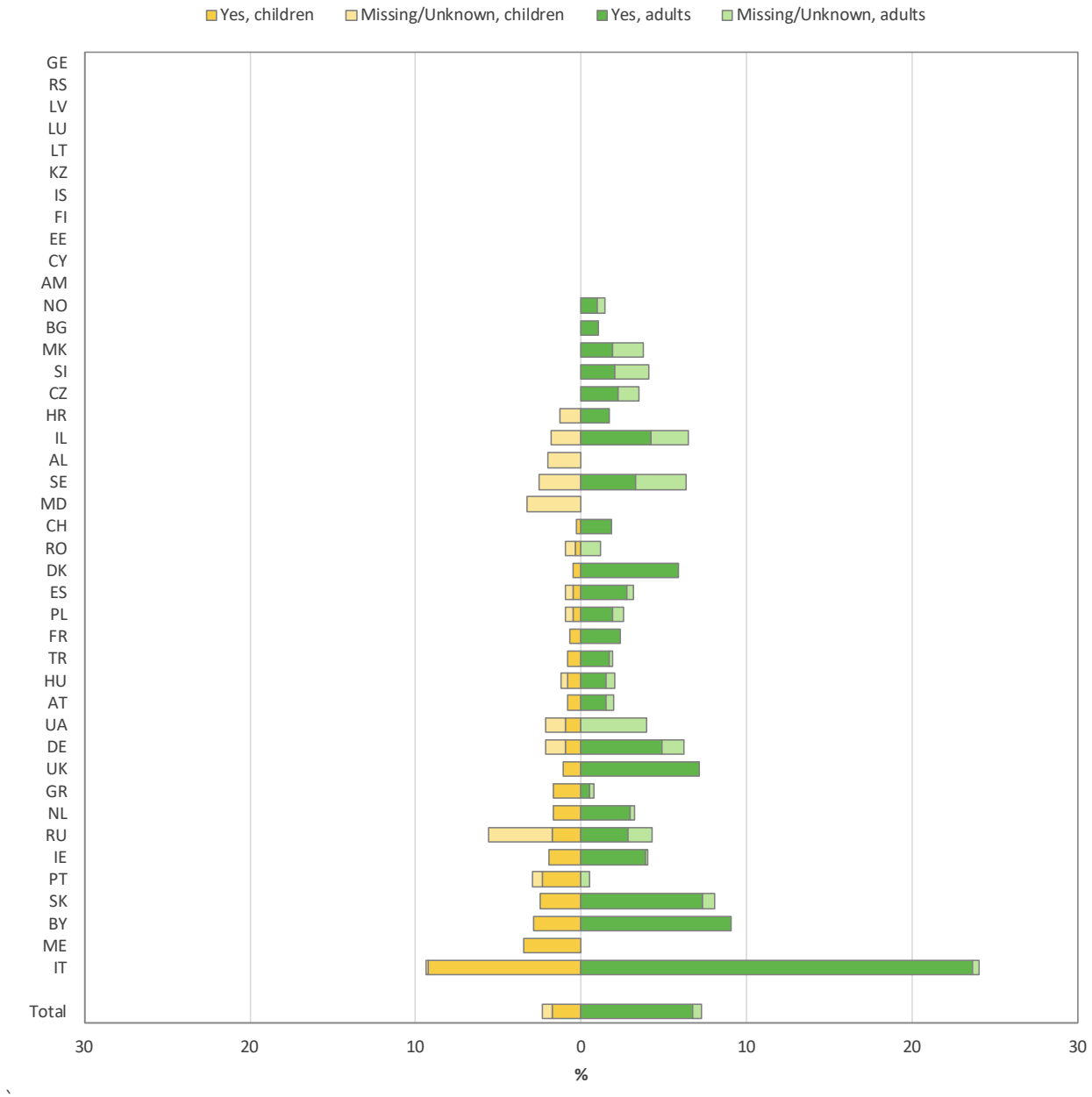
Inhaled steroids are reimbursed in most countries except Armenia, Georgia, Kazakhstan, Lithuania, Poland, Serbia and the Republic of Moldova (in the latter they are reimbursed for pwCF also diagnosed with asthma). In Bulgaria they are reimbursed for pwCF who are also diagnosed with asthma or chronic obstructive pulmonary disease (COPD). In Estonia and Romania inhaled steroids are reimbursed for pwCF who are also diagnosed with asthma.

This graph shows the use of inhaled steroids for at least 3 consecutive months during the survey year. The dark area of the bar indicates the percentage of people who took inhaled steroids, the lighter area shows the percentage of people for whom this information is missing.

8. Therapies

Figure 8.8 Oral steroids are less prescribed than inhaled steroids.

Use of oral steroids in children and adults seen in 2023 who have never had a transplant, by country (table A8.9, Appendix 1).



Note: We excluded from the graph the countries for which the information on use of oral steroids is missing for more than 10% of the children/adults with CF. Albania and Georgia have <5 adults seen in 2023 and are excluded from the graph for adults.

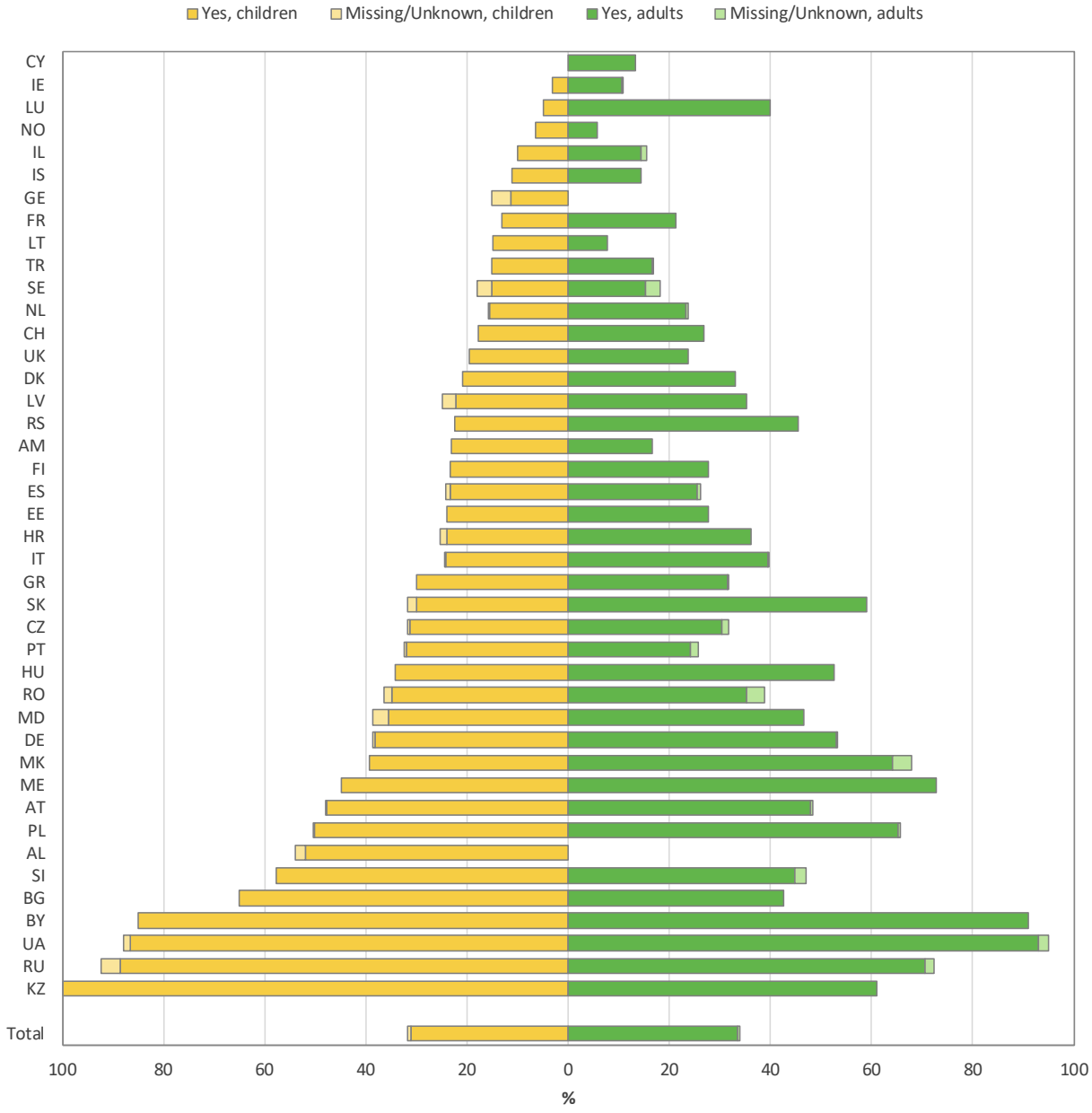
Oral steroids are reimbursed in most countries except in Armenia, Bulgaria, Georgia, Kazakhstan, Lithuania, the Republic of Moldova and Serbia.

This graph shows the use of oral steroids for at least three consecutive months during the survey year. The dark part of the bar indicates the percentage of people who took oral steroids, the light area shows the percentage of people for whom this information is missing.

8. Therapies

Figure 8.9 Ursodeoxycholic acid is often prescribed to treat cholestasis or liver disease in people with CF.

Use of ursodeoxycholic acid in children and adults seen in 2023 who have never had a transplant, by country (table A8.10, Appendix 1).



Note: We excluded from the graph the countries for which the information on oral ursodeoxycholic acid use is missing for more than 10% of the children/adults with CF. Albania and Georgia have <5 adults seen in 2023 and are excluded from the graph for adults.

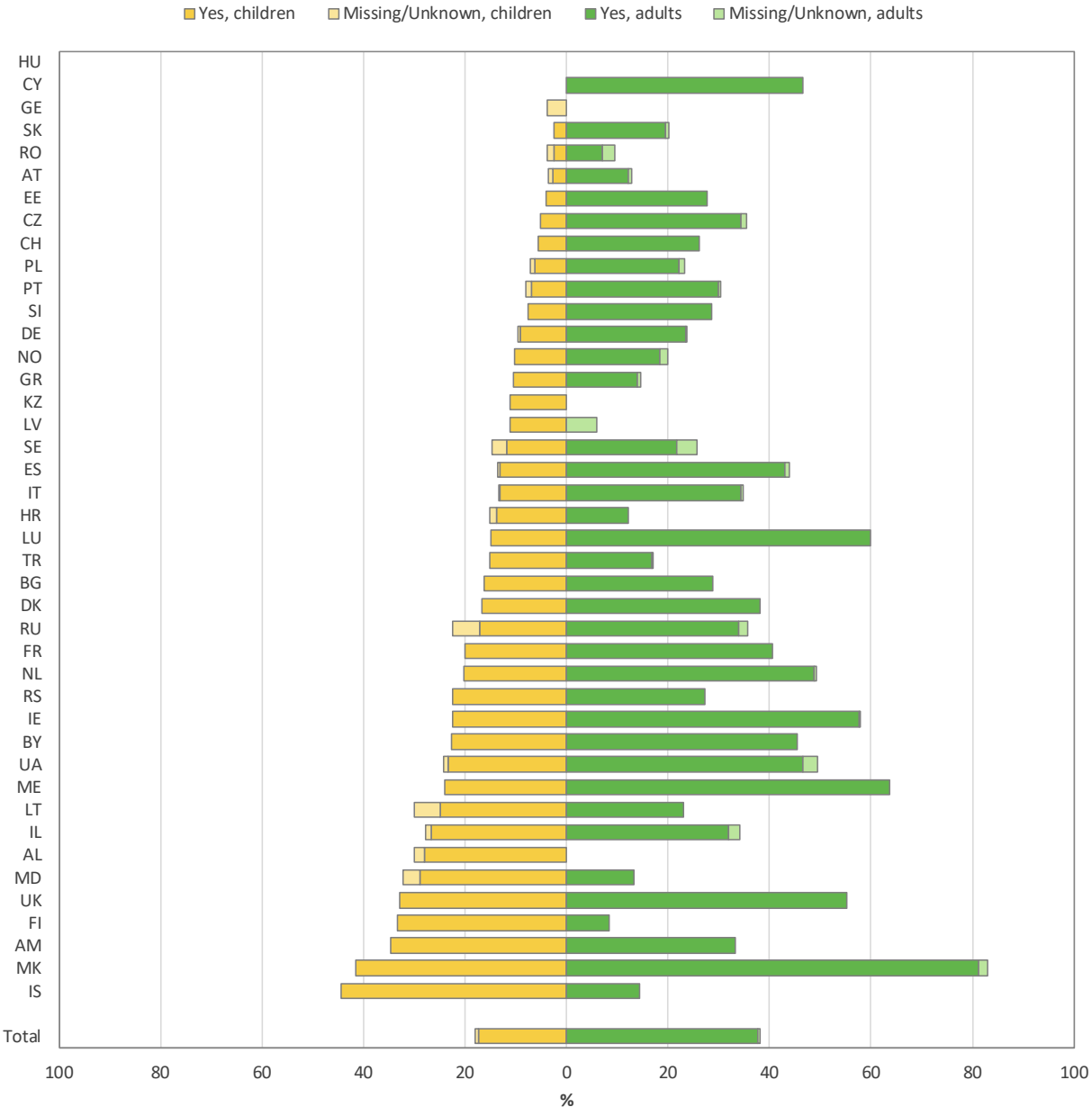
Oral ursodeoxycholic acid is reimbursed in most countries in Europe, except in Armenia, Bulgaria, Georgia, Lithuania and Serbia. In the Republic of Moldova it is reimbursed at 100% for children and at 70% for adults.

This graph shows how many people with CF used ursodeoxycholic acid for at least three consecutive months during 2023. Ursodeoxycholic acid is used as a treatment for CF liver disease. The dark area of the bar indicates the percentage of people who took the medication, the lighter area shows the percentage of people for whom this information is missing.

8. Therapies

Figure 8.10 Proton Pump Inhibitors are used to treat gastroesophageal reflux and gastritis, both common complications in CF, and to enhance pancreatic enzyme efficacy.

Use of proton pump inhibitors (PPI) in children and adults seen in 2023 who have never had a transplant, by country (table A8.11, Appendix 1).



Note: We excluded from the graph the countries for which the information on the use of PPI is missing for more than 10% of the children/adults with CF. Albania and Georgia have <5 adults seen in 2023 and are excluded from the graph for adults.

Oral proton pump inhibitors are reimbursed in most countries except in Bulgaria, Georgia, Kazakhstan, Lithuania and Serbia. In Armenia they are reimbursed for some outpatients.

This graph shows the use of proton pump inhibitors (PPI) for at least three consecutive months during the survey year. The dark area of the bar indicates the percentage of people with CF who used PPI, the lighter part shows the percentage of people for whom this information is missing.

8. Therapies

Figure 8.11 The increased use of CFTR modulators in children with CF in Europe goes hand in hand with a decrease in the prescription of azithromycin and inhaled antibiotics, while that of inhaled mucolytics remains mostly unchanged.

Therapy use in children between 2013 and 2023.

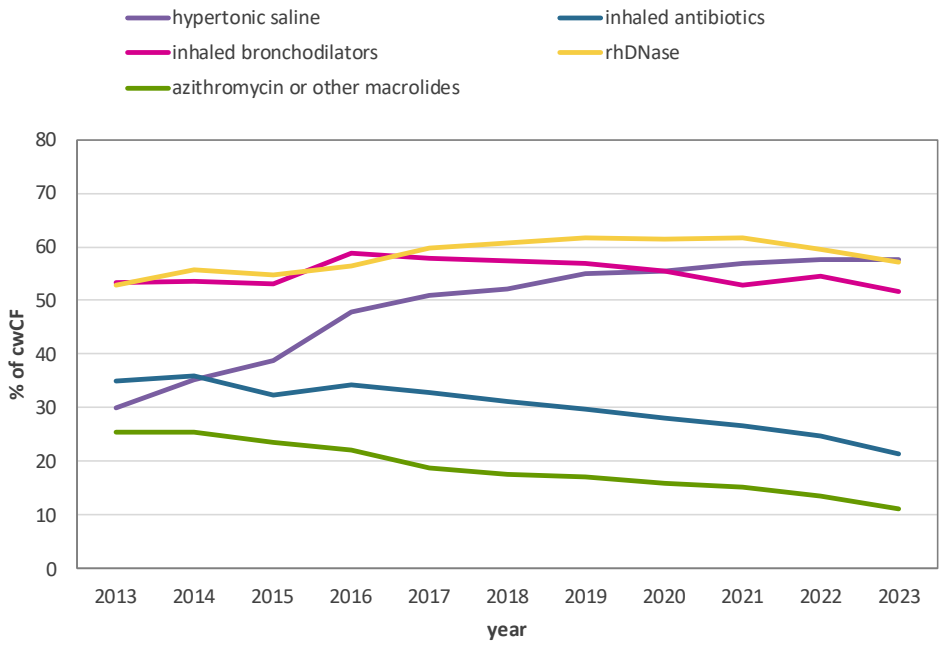
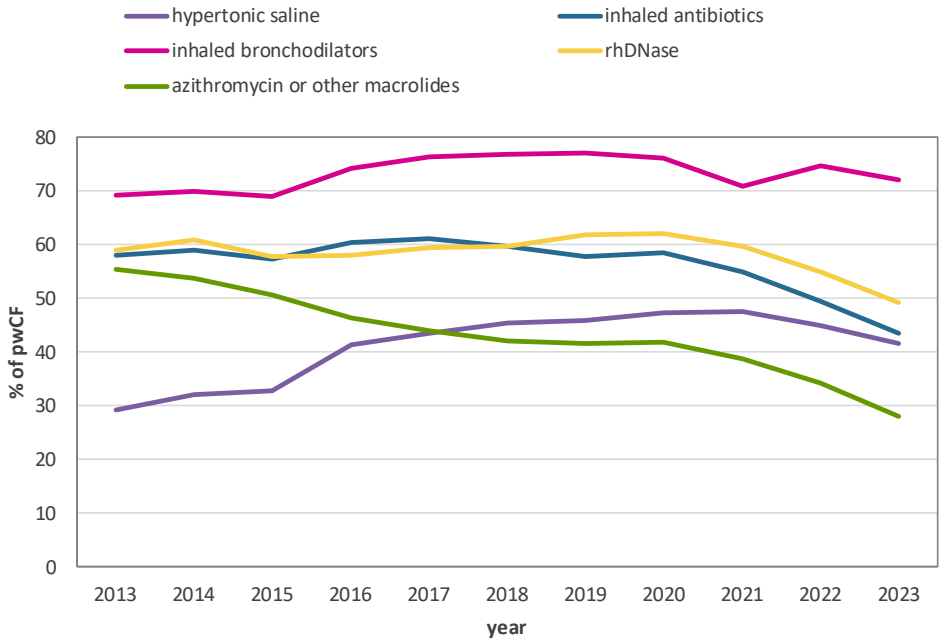


Figure 8.12 The increased use of CFTR modulators in adults with CF in Europe goes hand in hand with a decrease in the prescription of azithromycin, inhaled antibiotics and inhaled mucolytics.

Therapy use in adults between 2013 and 2023.



Figures 8.11 and 8.12 present data over time using cross sectional data per year of people with a confirmed CF. All people with CF alive, deceased or not seen during the year of follow-up were included. Individuals who have had a lung or/and liver transplant were excluded.

9. CFTR modulator therapies

The introduction of CFTR modulator therapies has had a significant impact on the health and quality of life of people with CF and also on CF care. These therapies target defects in the structure and function of the cystic fibrosis transmembrane conductance regulator (CFTR) protein. CFTR modulators are not effective in all people with CF since different variants cause different defects in the protein and/or its function.
In this chapter we present information about the use of the different CFTR modulators for people with CF.
We adopted the 2023 eligibility criteria of the European Medicines Agency (EMA) for all countries in the ECFSPR except Israel, the Russian Federation and Switzerland, where country-specific eligibility criteria laid down by the national regulatory authorities were applied. Where these are different from EMA criteria it is stated as such.

The country-specific eligibility criteria for the CFTR modulators in 2023 were provided by the medical Authorisation Holder:

Ivacaftor:
- at least 4 months old (1 month in UK) with at least one of the following variants: G551D, G1244E, G1349D, G178R, G551S, S1251N, S1255P, S549N, S549R, R117H (for R117H: in Switzerland people must be at least 18 years old);
- at least 2 years in Israel with at least one of the following variants: G551D, G1244E, G1349D, G178R, G551S, S1251N, S1255P, S549N, S549R.

Lumacaftor/ivacaftor:
- at least 1 year old (2 years in Switzerland and Russia, 6 years old in Israel) and F508del homozygous.

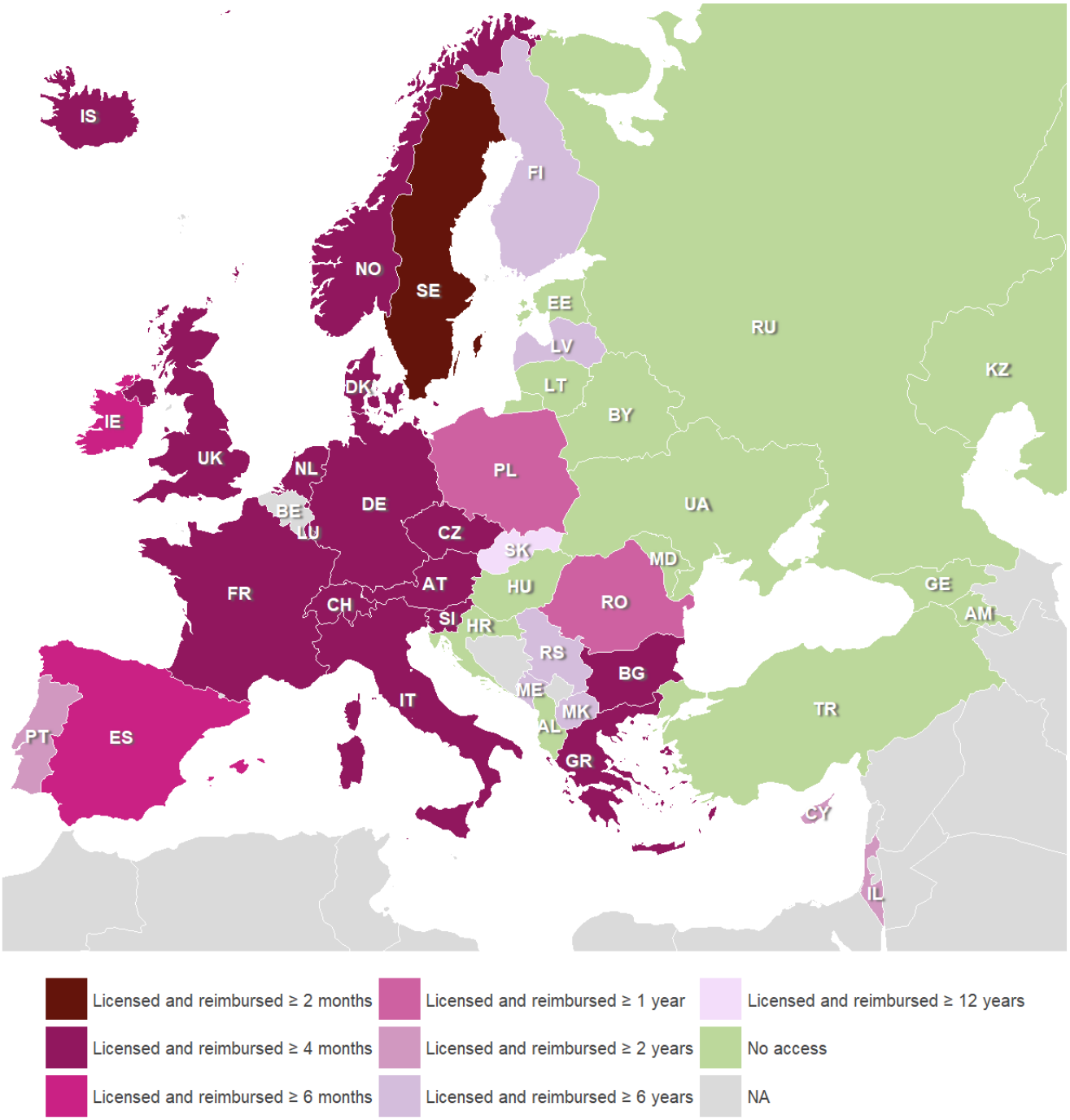
Tezacaftor/ivacaftor:
- at least 6 years old and F508del homozygous, or F508del heterozygous with one of the following variants: P67L, R117C, L206W, R352Q, A455E, D579G, 711+3A→G, S945L, S977F, R1070W, D1152H, 2789+5G→A, 327226A→G, or 3849+10kbC→T (also R347H in Israel).

Elexacaftor/tezacaftor/ivacaftor:
- at least 2 years old (6 years old in Switzerland and in Russia, 12 years old in Israel) and have at least one F508del variant (or a mutation in the CFTR gene that is responsive based on in vitro data in Israel and Russia).

The maps in this chapter show whether the CFTR modulators were licensed and reimbursed or not by national health services (information provided by the nominated country representative) in the countries participating in the ECFSPR in 2023 (Figure 9.1-9.4). These maps help with the interpretation of the country-specific variations in therapy use in people with CF who are eligible in each country (Figure 9.5, 9.6).

9. CFTR modulator therapies

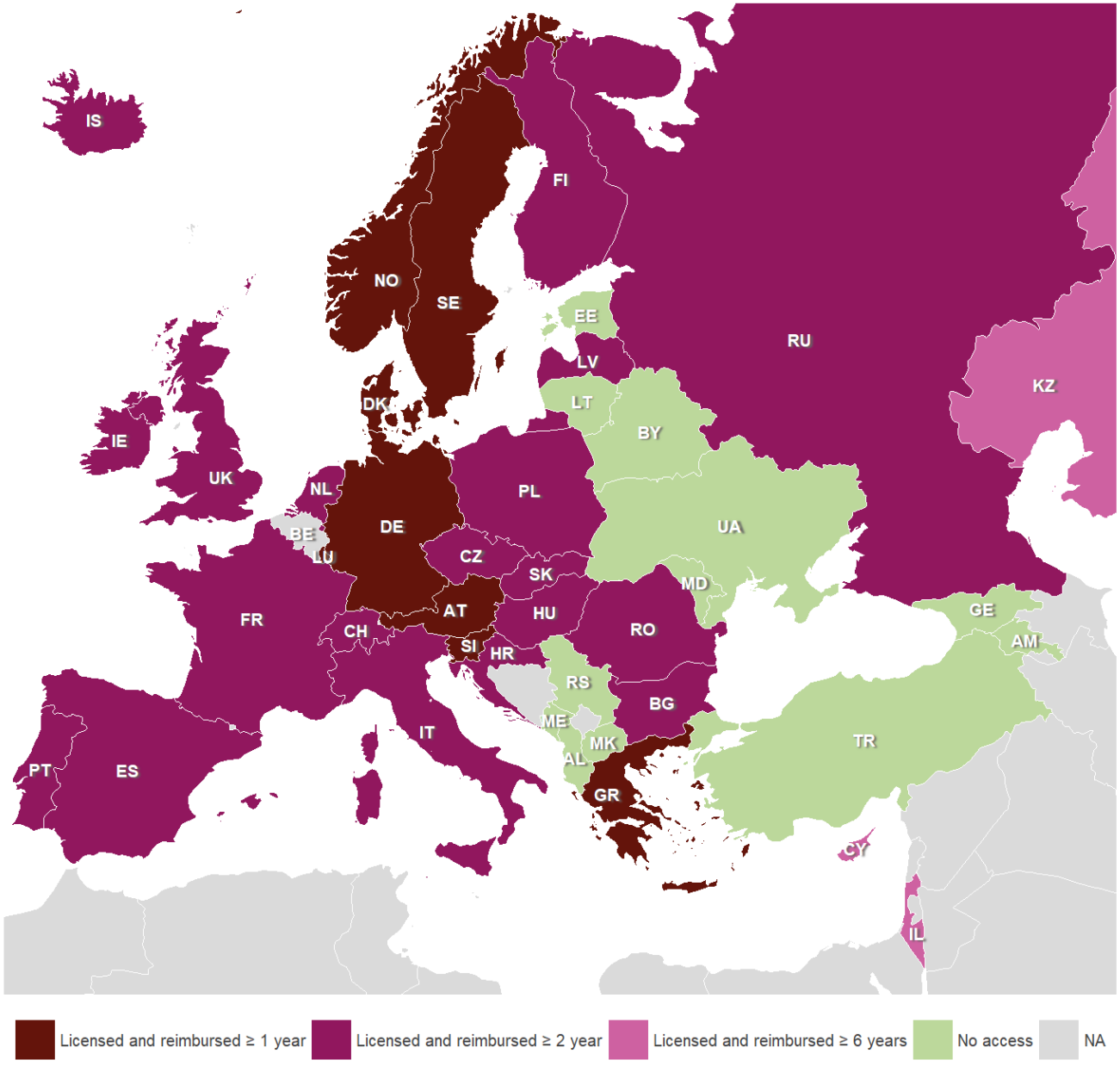
Figure 9.1 Countries where ivacaftor was licensed and reimbursed in 2023.



Note: Norway: ivacaftor was reimbursed for children with CF if weight ≥ 5 kg.
United Kingdom: ivacaftor was reimbursed for people with CF with the variant R117H who were ≥ 6 months old.

9. CFTR modulator therapies

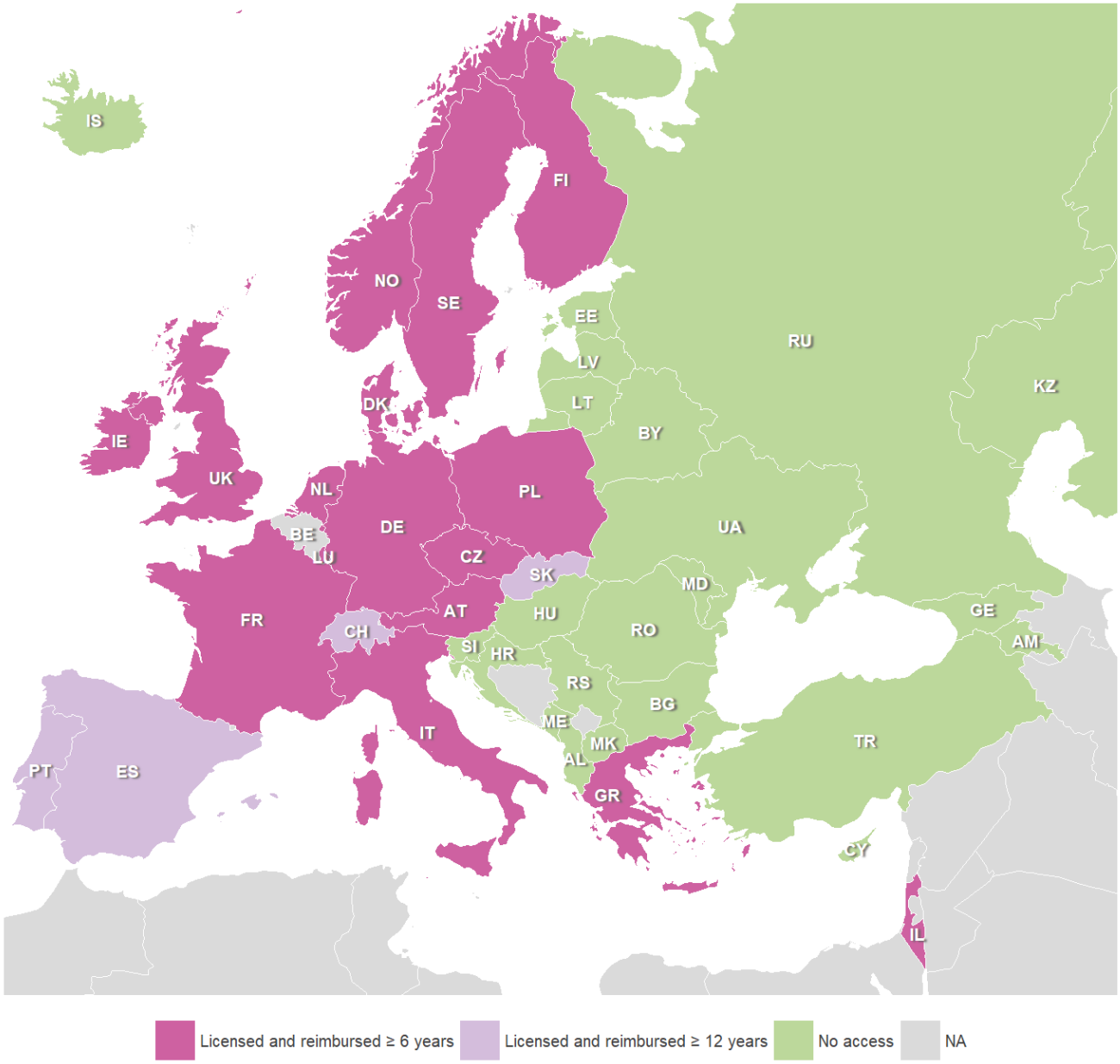
Figure 9.2 Countries where lumacaftor/ivacaftor was licensed and reimbursed in 2023.



Note: Denmark: ≥ 1 year from June 2023.
Finland and Hungary: ≥ 2 years (F508 homozygous).
Iceland: ≥ 1 year from December 2023.
Norway: ≥ 1 year from October 2023.
Russian Federation: > 2 years and <18 years.
Spain: ≥ 2 years and < 11 years.
Sweden: ≥ 1 year from summer 2023.

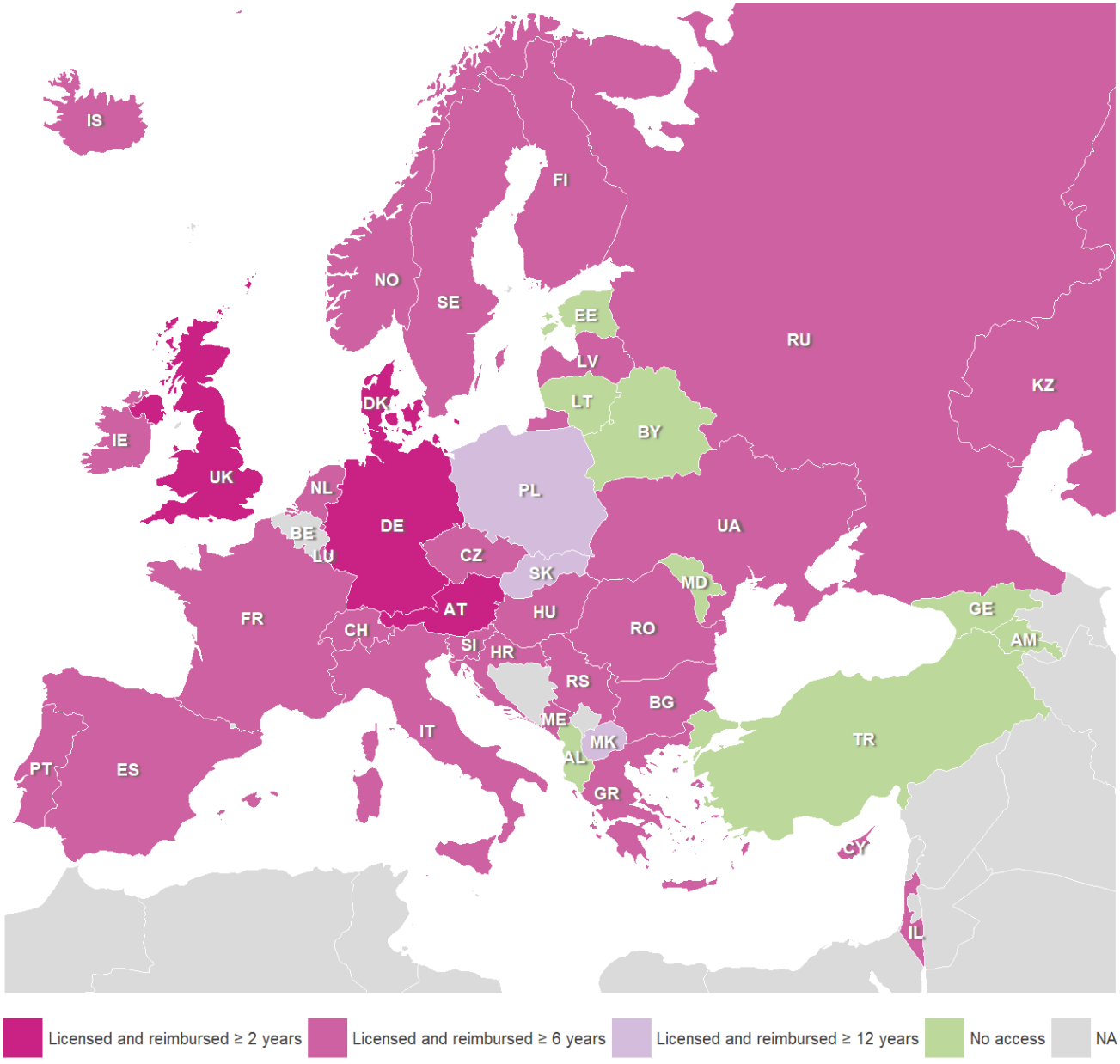
9. CFTR modulator therapies

Figure 9.3 Countries where tezacaftor/ivacaftor was licensed and reimbursed in 2023.



9. CFTR modulator therapies

Figure 9.4 Countries where elexacaftor/tezacaftor/ivacaftor was licensed and reimbursed in 2023.

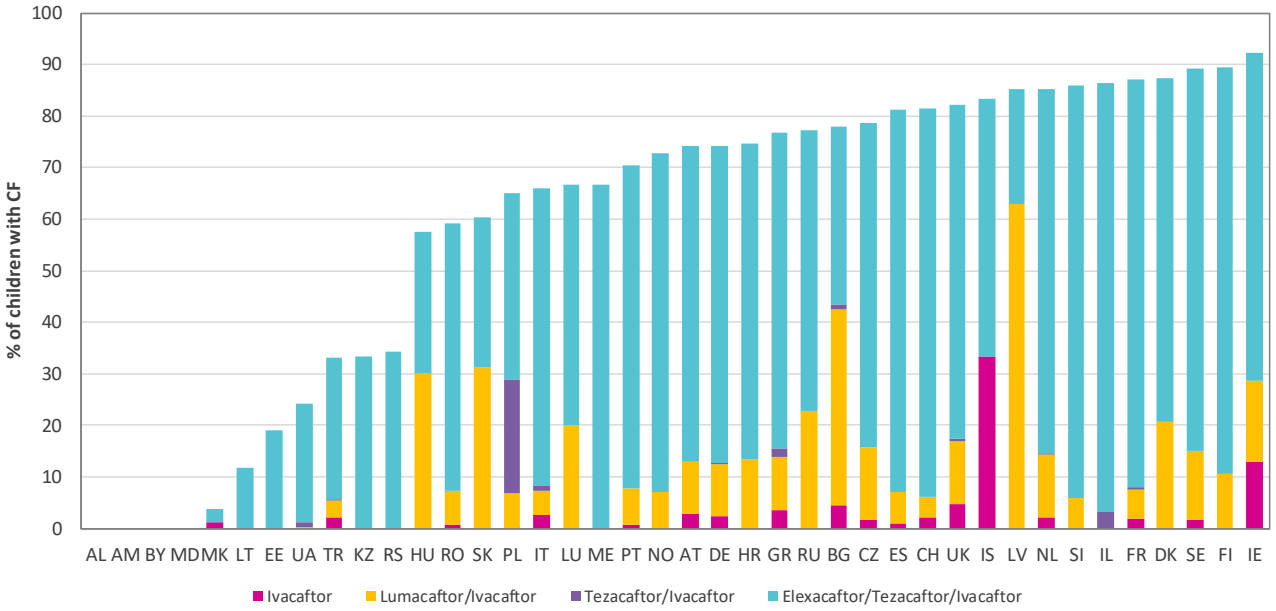


Note: Denmark: ≥ 2 years from November 2023.
Finland and Hungary: ≥ 6 years with at least one F508 mutation.
France: from September 2023 elexacaftor/tezacaftor/ivacaftor was reimbursed for people with CF 2-5 years (early access programme)
Ukraine: ≥ 6 years from August 2023.
United Kingdom: ≥ 2 years from November 2023.

9. CFTR modulator therapies

Figure 9.5 Elexacaftor/tezacaftor/ivacaftor is the CFTR modulator most commonly used in children, followed by lumacaftor/ivacaftor.

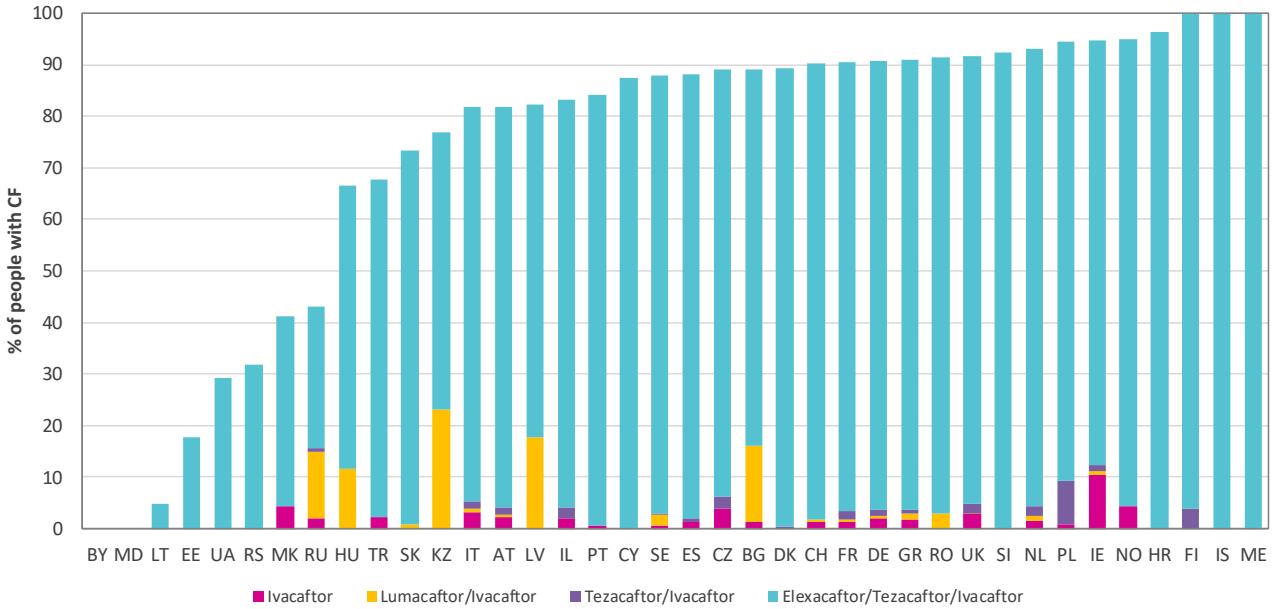
Children and adolescents with CF (<18 years), eligible for and treated with at least one modulator, by country and last CFTR modulator prescribed, seen in 2023 and who have never had a transplant.



Note: Cyprus and Georgia have <5 eligible children seen in 2023 and are excluded from the graph.

Figure 9.6 In most countries in Europe the majority of all adults with CF are eligible for CFTR modulator treatment.

Adults with CF (≥18 years), eligible for and treated with at least one modulator, by country and last CFTR modulator prescribed, seen in 2023 and who have never had a transplant.

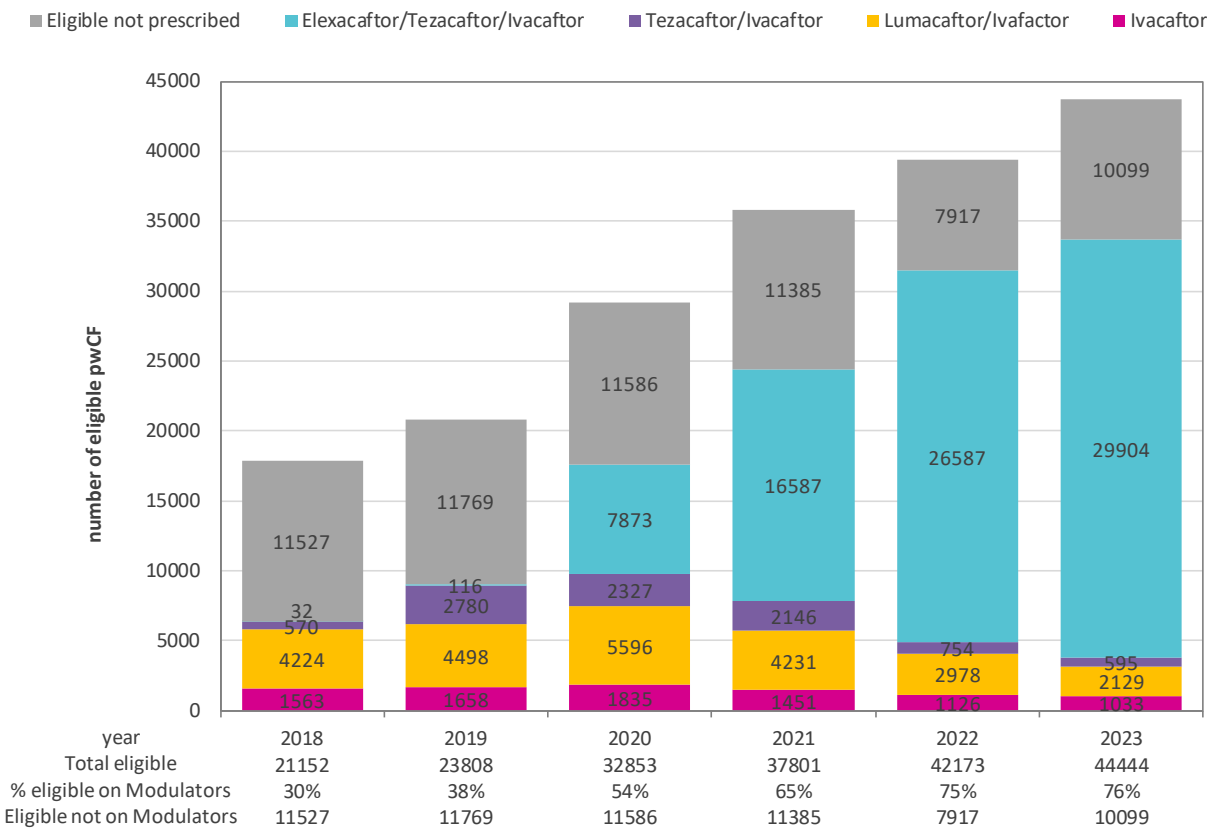


Note: Albania, Armenia, Georgia and Luxembourg have <5 eligible adults seen in 2023 and are excluded from the graph.

9. CFTR modulator therapies

Figure 9.7 With expansion of the eligibility criteria and a more extensive reimbursement programme for CFTR modulators in Europe there has been a considerable increase in their use since 2020.

Use of CFTR modulator therapy from 2018 to 2023.



In this graph we present data over time using cross sectional data per year of people with a confirmed CF diagnosis. All people with CF alive, deceased or not seen during the year of follow-up were included.

10. Pregnancy

Previous patient registry data demonstrated no difference in survival for women with CF who have had one or more pregnancies compared with those who have never been pregnant[1]. However, there is a higher risk of pregnancy-related complications in women with CF (e.g. gestational diabetes, need for caesarean section, premature birth and congenital anomalies).
The introduction of CFTR modulators has significantly improved lung function, nutritional status and overall health in individuals with cystic fibrosis. Before modulators, many women with CF experienced reduced fertility due to thick cervical mucus and overall poor health, however, with improved airway hydration and nutritional status and better systemic health, more women are conceiving naturally.
While pregnancy is now more achievable it presents unique challenges. The safety of CFTR modulator therapy during pregnancy is still to be ascertained and is under study, with some evidence suggesting potential benefits for both mother and baby, though long-term effects remain unknown. Increased metabolic demands and the need for optimised respiratory and nutritional care make close medical supervision essential. Careful planning and multidisciplinary management are crucial to ensure the best outcome for both mother and child.
We collected information about pregnancies and pregnancy outcome during the year of follow-up.

[1] C.H. Goss at al., Chest 2003;124(4):1460-8.

10. Pregnancy

Table 10.1 Pregnancy and pregnancy outcomes in women with CF (≥16 years) seen in 2023 who have never had a transplant.

Reason	Number	Percentage
Missing/Unknown	495	3.6
No	12566	91.0
Yes, ongoing at the end of 2023	247	1.8
Yes, stopped during the year	501	3.6
Pregnancy outcome:		
Live birth	410	81.8
Spontaneous abortion (<28weeks)	47	9.4
Still birth (>=28weeks)	1	0.2
Therapeutic abortion for medical reasons	26	5.2
Other	8	1.6
Missing/Unknown	9	1.8

11. Lung and other organ transplants

Despite the advent of highly effective CFTR modulators lung transplant is still a realistic scenario for some people with CF. The availability of a lung transplant programme differs amongst the countries participating in the ECFSPR and transplant eligibility criteria also vary. The numbers presented in the tables and figures that follow should be considered an indication of accessibility to a regional or national lung transplant programme rather than a reflection of the standards of patient care and health status of the people with CF in a specific country. The same applies to liver transplant, the second most common organ that is transplanted in CF. In this chapter, we also give information on kidney transplant and other (unspecified) organ transplant.

We asked if people have had a transplant or not and, if yes, the year of their (latest) transplant. In some countries people who have had a transplant are no longer registered in the CF centres or the national CF registry because they have been transferred to a transplant centre. For this reason the figures may report a lower number than the reality but it was not possible to acquire more accurate data.

11. Lung and other organ transplants

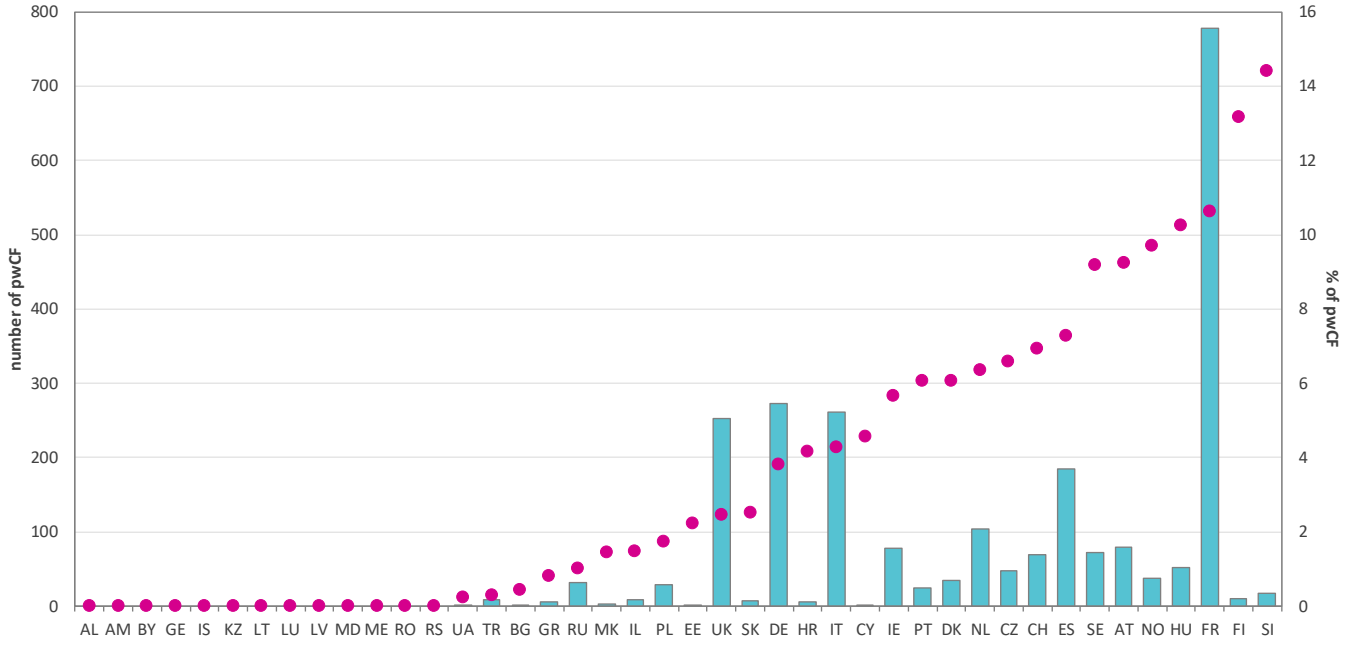
Table 11.1 Number of people with CF alive in 2023 with transplanted lung(s), by age and sex.

Age	Males	Females	Total	Transplants carried out in 2023
0-5	0	0	0	0
6-11	2	1	3	1
12-17	6	17	23	7
18-29	159	240	399	23
30-39	439	427	866	15
40-49	406	387	793	8
50-59	180	145	325	3
60+	34	28	62	1
Total	1226	1245	2471	58

This table shows the number of people with CF alive in 2023 who have had one or more lung transplant(s) at some time in their life, by age group, as well as the number of people who had a lung transplant in 2023.

Figure 11.1 The number and proportion of people with CF living with transplanted lung/s is heterogenous across Europe.

Number of people with CF living in 2023 with transplanted lungs, by country.



This graph shows the number of people with CF alive at 31/12/2023 who have had a lung transplant (light turquoise bars) at some point in their life. The pink dots (right axis) show the percentage of people that are living with transplanted lung in 2023 out of all people with CF that were seen in 2023.

11. Lung and other organ transplants

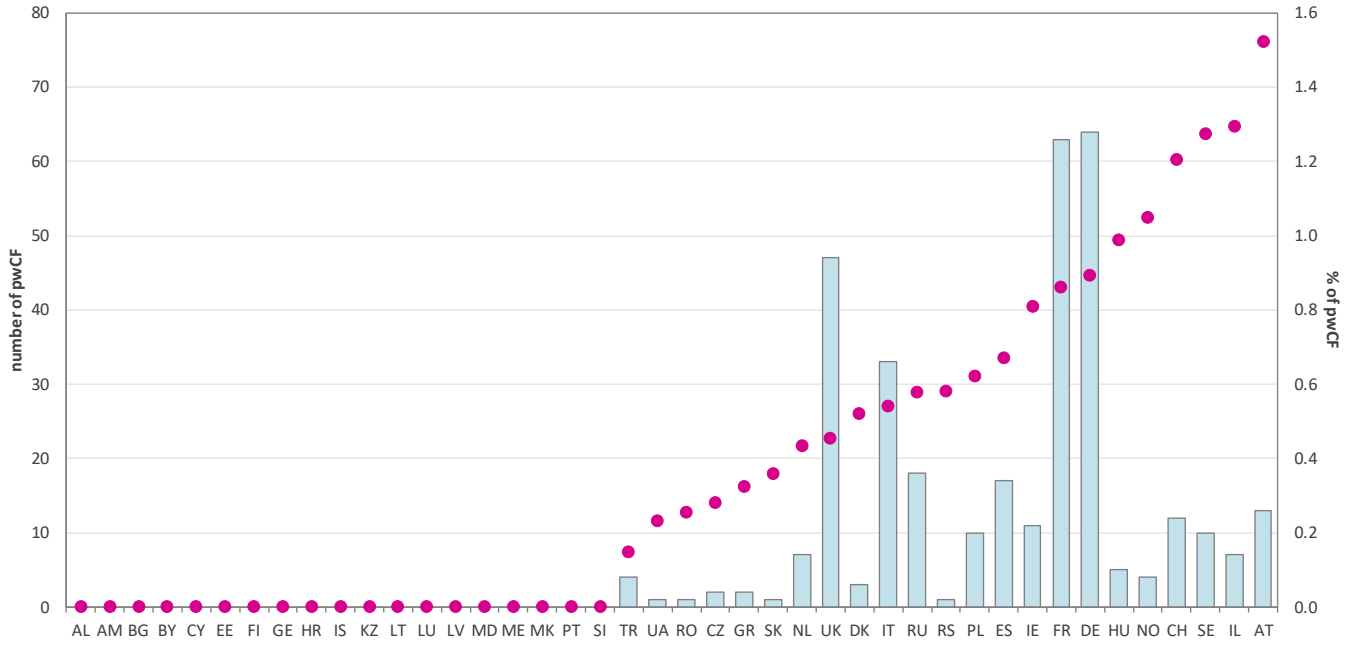
Table 11.2 Number of people with CF living in 2023 with transplanted liver, by age and sex.

Age	Males	Females	Total	Transplants carried out in 2023
0-5	2	0	2	0
6-11	8	0	8	2
12-17	26	8	34	5
18-29	82	46	128	6
30-39	69	33	102	3
40-49	34	10	44	0
50-59	9	9	18	1
60+	0	0	0	0
Total	230	106	336	17

This table shows the number of people with CF alive in 2023 who have had a liver transplant at some time in their life, by age group, as well as the number of people who had a transplant in 2023.

Figure 11.2 The number and proportion of people with CF living with a liver transplant is extremely heterogenous throughout Europe.

Number of people with CF living in 2023 with transplanted liver, by country.



This graph shows the number of people with CF alive at 31/12/2023 who have had a liver transplant (light turquoise bars) at some point in their life. The pink dots (right axis) show the percentage of people living with transplanted liver in 2023 out of all people with CF that were seen in 2023. Note that (left vertical axis) the number of people who have had a liver transplant is much lower than the number of people with transplanted lungs. The main reason for this is that liver disease is only found in a subset of people with CF, whereas lung disease affects almost all people with CF.

11. Lung and other organ transplants

Table 11.3 Number of people with CF living in 2023 with transplanted kidney(s), by age and sex.

Age	Males	Females	Total	Transplants carried out in 2023
0-5	0	0	0	0
6-11	1	0	1	0
12-17	1	0	1	0
18-29	10	9	19	2
30-39	19	39	58	8
40-49	41	33	74	4
50-59	18	13	31	0
60+	6	2	8	1
Total	96	96	192	15

This table shows the number of people with CF alive in 2023 who have had a kidney transplant at some time in their life, by age group, as well as the number of people who had a transplant during 2023.

Table 11.4 Number of people with CF living in 2023 with other transplanted organs (not lung, liver, kidney), by age and sex.

Age	Males	Females	Total	Transplants carried out in 2023
0-5	0	0	0	0
6-11	1	1	2	1
12-17	2	2	4	0
18-29	7	7	14	1
30-39	6	9	15	1
40-49	9	9	18	0
50-59	5	2	7	0
60+	0	0	0	0
Total	30	30	60	3

This table shows the number of people with CF alive in 2023 who have had an organ transplant that is not lung, liver or kidney (other) at some time in their life, by age group, as well as the number of people who had a transplant during 2023.

12. Mortality

Information on mortality and survival in the era of highly effective CFTR modulators is currently a major area of focus in CF. Although we can speculate that these drugs will increase life expectancy in people with CF, the effects can only be monitored in the long-term. In this chapter we present the number of deaths by age group, for males and females. Respiratory disease continues to be the predominant cause of death in people with CF.
We do not present data on survival prediction in this report since mortality data are heterogenous and may be incomplete in the participating countries. However, information on survival is collected and reported on by selected National Registries and we refer you to their country-specific annual reports for further reading.

12. Mortality

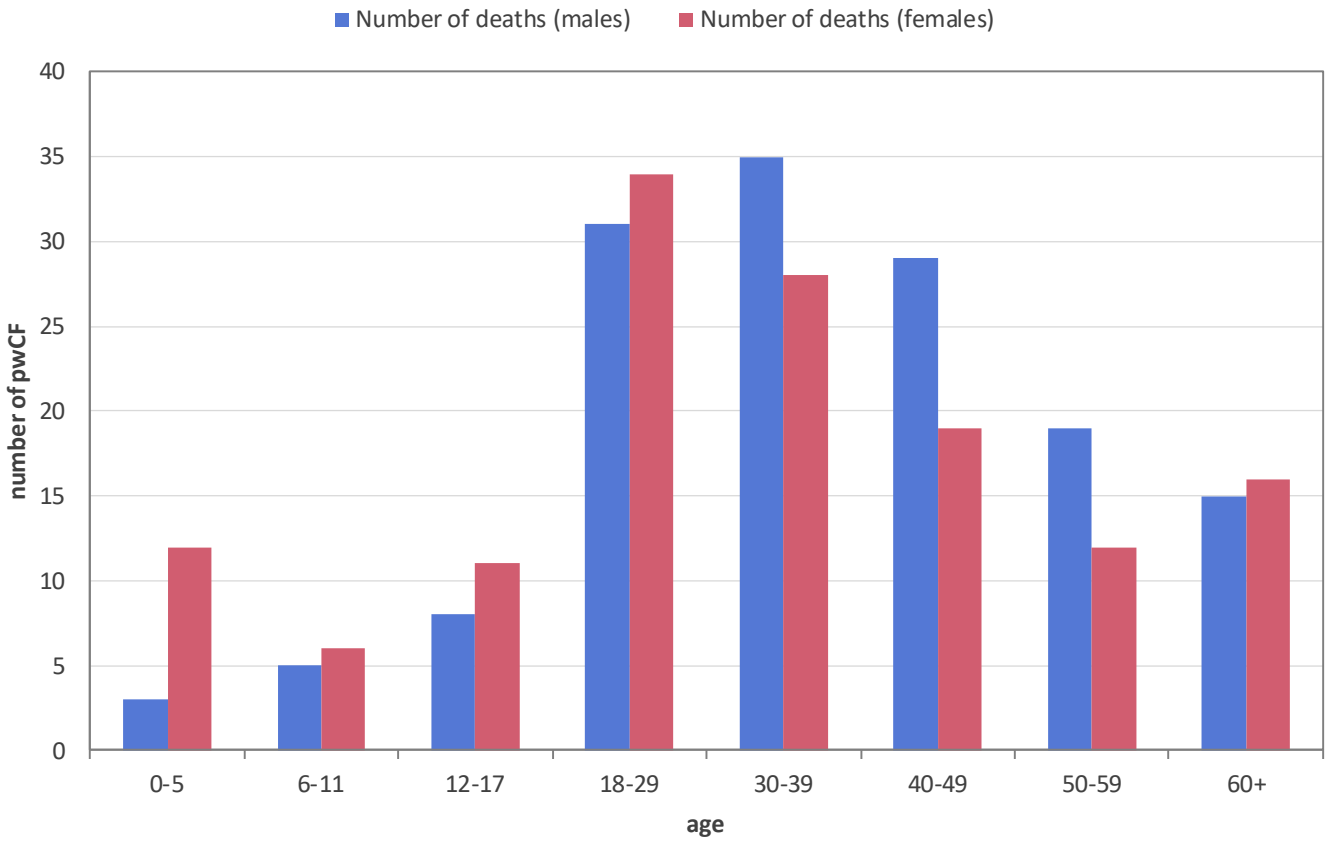
Table 12.1 Number of deaths in 2023, by age and sex.

Age at death	Number of male pwCF	% of deaths in this age group (of all male deaths)	Number of female pwCF	% of deaths in this age group (of all female deaths)	Total	% Total
0-5	3	2.07	12	8.70	15	5.30
6-11	5	3.45	6	4.35	11	3.89
12-17	8	5.52	11	7.97	19	6.71
18-29	31	21.38	34	24.64	65	22.97
30-39	35	24.14	28	20.29	63	22.26
40-49	29	20.00	19	13.77	48	16.96
50-59	19	13.10	12	8.70	31	10.95
60+	15	10.34	16	11.59	31	10.95
Total	145		138		283	

Note: For the United Kingdom, all people with a confirmed diagnosis of CF were included (N=11,318). The total number of the CF population presented is 53,926.

This table shows the number of deaths in 2023 by age group and sex. Death in small children is very rare, and the most frequent range of age at death for both sexes is 18-29 years. It is possible that the numbers are under reported because some of the people who died may not have been seen at the CF centre during the year, and therefore the information may not have been recorded.

Figure 12.1 Most of the deaths occur between the third and the fifth decade of life in people with CF in Europe.
Age at death distribution of people with CF deceased in 2023, by sex.



This graph shows the distribution of age at death of people with CF who died in 2023, separated by males (blue) and females (red).

12. Mortality

Table 12.2 Cause of death distribution of deaths in 2023.

Cause of death in 2023	Number of deaths	Percentage of all deaths
Respiratory	139	49.12
Transplantation	35	12.37
Cancer	26	9.19
Non-CF related	23	8.13
Other CF related	23	8.13
Unknown	14	4.95
Liver-GI	13	4.59
Suicide	6	2.12
Trauma	4	1.41
Total	283	

Note: For the United Kingdom, all people with a confirmed diagnosis of CF were included (N=11,318). The total number of the CF population presented is 53,926.

Ireland, Germany and the United Kingdom record cause of death as "cardio/respiratory".

The table shows cause of death for people with CF who died in 2023. The most frequent cause of death was respiratory disease.

13. Data quality

Data that will be employed in vital research and pharmacovigilance studies, inform public health planning and serve as an instrument to monitor and review a range of patient outcomes need to be of demonstrably high quality. Several measures are in place in the ECFSPR that demonstrate both our ongoing commitment to quality and our support for the contributing centres and national registries, namely: guidance documentation, training, on-demand assistance to participants on all aspects of the data cycle, a customised software with inbuilt controls and other rigorous checks.

In addition, in 2018 we launched a new initiative to verify and validate data at source in participating centres; objectives are to quantify data completeness, consistency and the accuracy of data. The visits also offer an invaluable opportunity to discuss quality, relevance and reliability in the ECFSPR data.

“Consistency” means adherence by the centre to the variable definitions, options, and parameters used by the Registry. “Accuracy” of data-input is the proportion of values recorded in the ECFSPR software that matches the medical records and is consistent with the ECFSPR definitions and limits.

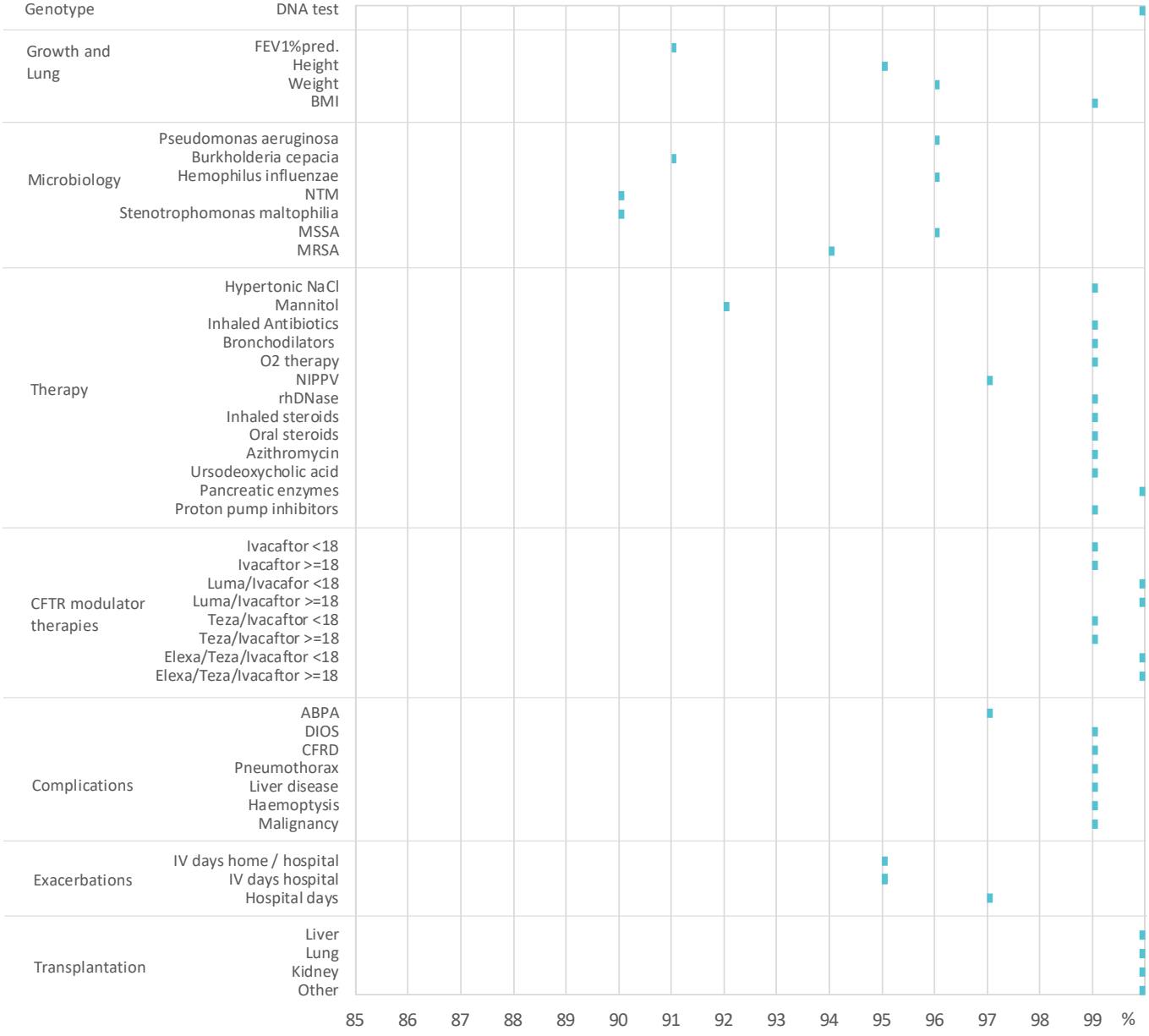
Consistency of data was determined for a selection of variables by comparing the ECFSPR data and the data in the patient medical health record at the centres. The variables checked are the same for each centre and they were chosen because they are more challenging to collect and/or more open to misinterpretation (based on ECFSPR experience and participant feedback), and they are statistically significant.

In this chapter we present a synopsis of the overall quality of the ECFPSR data based on the factors of completeness and accuracy. Unlike the rest of this report data quality results refer to both 2022 and 2023; the overall results have been shown in to illustrate areas where improvement has been demonstrated.

13. Data quality

Figure 13.1 The overall completeness of data for all variables from the countries participating in the Registry in 2023 is 98%.

Completeness of the data in 2023, for all people with CF who have never had a lung/liver transplant seen in all participating countries, overall percentages by variable.



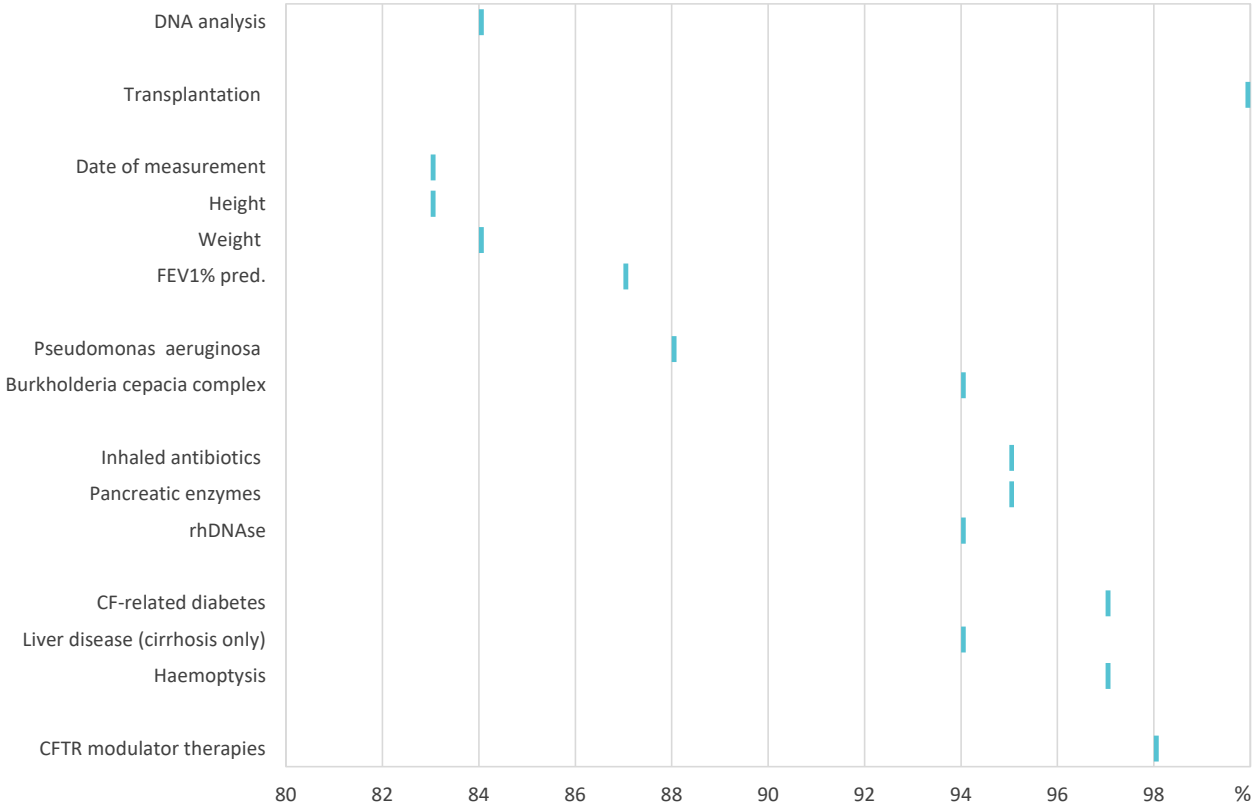
Note: Completeness for FEV1 is evaluated only for individuals with CF ≥ 6 years.
Completeness for BMI is evaluated only for individuals with CF ≥ 2 years.

For lung function and some microbiology data completion is in the range 90-91%. Lung function data are dependent on the ability of a person with CF to perform spirometry correctly. Likewise, data completeness for microbiology depends on sample availability and access to testing laboratories.

13. Data quality

Figure 13.2 The validated clinical data amounts to 4% of the people with CF in these countries for whom we have 2022 and 2023 data.

Data accuracy for the follow-up years 2022 and 2023 from countries visited, overall percentages by variable.



We validated data (from 2022 and 2023) for 1651 people with CF from 17 countries. 10 countries with established national CF registries (Czech Republic, France, Germany, Greece, Ireland, Italy, the Netherlands, Sweden, Türkiye and United Kingdom) have their own internal data validation programme and have provided us with the results presented in this report.

Table 13.1 Accuracy of data (quartiles) for people with CF, from the follow-up years 2022 and 2023 for the countries visited.

Variable	DNA analysis	Transplantation	Date of measurement	Height	Weight	FEV1% pred.	Pseudomonas aeruginosa	Burkholderia cepacia complex	Inhaled antibiotics	Pancreatic enzymes	rhDNase	CF-related diabetes	Liver disease (cirrhosis only)	Haemoptysis	CFTR modulator therapies
25th percentile	81	100	76	76	77	80	81	91	93	95	91	96	91	97	99
50th percentile	89	100	82	84	84	88	91	99	96	98	98	98	95	100	100
75th percentile	95	100	91	93	93	95	96	100	100	99	99	100	100	100	100

Overall, for most variables, median accuracy of validated data in the year 2022-2023 is 92%. Genotype, date of measurements, height, weight and lung function are the variables that proved most challenging for data providers. The accuracy of genetic data cannot be verified without the original genetic report. For the other four variables, to minimise errors and improve adherence to ECFSPR definitions, the centres have agreed to review procedures and the organisation of documentation across hospital departments.

Publications

The ECFSPR database is an excellent and unique resource for researchers and the data are actively used by many individuals, institutions and companies. Applications for projects are handled in accordance with the ECFSPR guidelines. You will find more information on the rigorous research project application process on our website here: Research Project Application.
From 2011 to March 2025 (time of writing) we had received a total of 131 applications to use Registry data for projects. The majority of these requests, 83%, originated from researchers from the European Cystic Fibrosis Society members and other institutes; 17% of the applications came from Industry. Many of these research projects resulted in publications and other publications are in the pipeline.

From April 2023 to April 2024 the following publications and abstracts were accepted:

Publications

- Variability in disease severity among cystic fibrosis patients carrying residual-function variants: data from the European Cystic Fibrosis Society Patient Registry. Mei-Zahav M, Orenti A, Jung A, et al. ERJ Open Res 2025; 11: 00587-2024. doi: 10.1183/23120541.00587-2024.

- Cystic fibrosis in Europe: improved lung function & longevity: reasons for cautious optimism, but challenges remain. Kerem E, Orenti A, Adamoli A, Hatziagorou E, Naehrlich L, Sermet-Gaudelus I, on behalf of the ECFS Patient Registry Steering Group. Eur Respir J 2024; 63(3): 2301241. doi: 10.1183/13993003.01241-2023.

Abstracts

- Assessment of respiratory infection following initiation of elexacaftor/tezacaftor/ivacaftor using the European Cystic Fibrosis Society patient registry. M. Pollak, S. Gambazza, A. Orenti, J. van Rens, V. De Rose, M. Mei Zahav, on behalf of ECFSPR Steering Group. Journal of Cystic Fibrosis, Volume 23, Supplement 1, S4, June 2024.

- Methicillin-Resistant Staphylococcus aureus and pulmonary outcome in people with cystic fibrosis: a European cystic fibrosis patient registry data analysis. M. Mei-Zahav, M. Dotan, L. Annicchiarico, A. Orenti, D. Prais, on behalf of ECFSPR Steering Group. Journal of Cystic Fibrosis, Volume 23, Supplement 1, S116, June 2024.

- Improved outcome in the adult cystic fibrosis population in Europe from 2012 to 2022: analysis of the European Cystic Fibrosis Society Patient Registry. A. Orenti, E. Hatziagorou, E. Kerem, C. De Boeck, P.-R. Burgel, on behalf of ECFSPR Steering Group. Journal of Cystic Fibrosis, Volume 23, Supplement 1, S50, June 2024.

- A retrospective analysis of the ECFSPR to characterise the pulmonary phenotype and use of intravenous antibiotics in people with cystic fibrosis harbouring Bi-allelic CFTR class 1 mutations. A. Orenti, W.K. Mountford, J.C. Davies, D. Polineni, A. Adamoli, E. Bakkeheim, I. Isabelle, M.A. Mall, C.P. Alves, W. Seibold, R. Sigmund, S.B. Carr, on behalf of ECFSPR Steering Group. Journal of Cystic Fibrosis, Volume 23, Supplement 1, S120-S121, June 2024.

- Factors associated with more frequent and severe pulmonary exacerbations in patients with Cystic Fibrosis: data from the ECFS Patient Registry. V. De Rose, A. Orenti, A. Adamoli, J. Van Rens, M. Pollak, M. Mei Zahav, on behalf of ECFSPR Steering Group. Journal of Cystic Fibrosis, Volume 23, Supplement 1, S50, June 2024.

- Disease burden in people with cystic fibrosis according to CFTR genotype and eligibility to CFTR modulator therapy: a ECFS Patient Registry analysis. I. Tomarelli, A. Orenti, P.-R. Burgel, A. Gramegna, F. Blasi, on behalf of ECFSPR Steering Group. Journal of Cystic Fibrosis, Volume 23, Supplement 1, S3-S4, June 2024.

- The quality of the data in the European Cystic Fibrosis Society Patient Registry as assessed through source data verification from 2018-2024. V. Prasad, A. Fox, M. Krasnyk, A. Zolin, J. van Rens, L. Naehrlich, on behalf of ECFSPR Steering Group. Journal of Cystic Fibrosis, Volume 23, Supplement 1, S123, June 2024.

- Utilisation of multiple breath washout in Europe differs substantially between age groups and countries. L. Naehrlich, A. Zolin, A. Lindblad, P. Gkolia, on behalf of the ECFSPR Steering Group. Journal of Cystic Fibrosis, Volume 23, Supplement 1, S122, June 2024.

- Association of change in Pseudomonas aeruginosa infection with clinical outcomes after Elexacaftor/Tezacaftor/Ivacaftor treatment, data from the European Cystic. Pollak M, Gambazza S, Orenti A, van Rens J, De Rose V, Mei-Zahav M, on behalf of the ECFSPR Steering Committee. North American Cystic Fibrosis Conference (NACFC) 2024 - Boston (US). September 2024.

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Appendix 1 Tables

1. Demographics

Table A1.1 Proportion of children (<18 years) and adults (≥18 years), by country and overall. People with CF alive on 31/12/2023.

Country	Children (<18 years)		Adults (≥18 years)	
	Number	%	Number	%
Albania	85	88.54	11	11.46
Armenia	25	80.65	6	19.35
Austria	364	41.60	511	58.40
Belarus	140	92.72	11	7.28
Bulgaria	136	57.87	99	42.13
Croatia	84	56.38	65	43.62
Cyprus	9	29.03	22	70.97
Czech Republic	357	48.57	378	51.43
Denmark	213	36.98	363	63.02
Estonia	25	56.82	19	43.18
Finland	30	39.47	46	60.53
France	2595	35.58	4698	64.42
Georgia	78	95.12	4	4.88
Germany	2850	37.87	4675	62.13
Greece	242	35.38	442	64.62
Hungary	251	50.10	250	49.90
Iceland	9	56.25	7	43.75
Ireland	526	38.25	849	61.75
Israel	173	29.73	409	70.27
Italy	2206	36.05	3913	63.95
Kazakhstan	9	33.33	18	66.67
Latvia	36	67.92	17	32.08
Lithuania	20	41.67	28	58.33
Luxembourg	22	84.62	4	15.38
Rep of Moldova	35	71.43	14	28.57
Montenegro	29	72.50	11	27.50
The Netherlands	533	32.58	1103	67.42
North Macedonia	101	60.84	65	39.16
Norway	137	35.40	250	64.60
Poland	1028	60.36	675	39.64
Portugal	175	43.00	232	57.00
Romania	324	78.26	90	21.74
Russian Federation	2840	69.93	1221	30.07
Serbia	125	62.19	76	37.81
Slovak Republic	126	40.78	183	59.22
Slovenia	53	44.54	66	55.46
Spain	1088	40.98	1567	59.02
Sweden	281	35.08	520	64.92
Switzerland	410	38.07	667	61.93
Türkiye	2255	82.54	477	17.46
Ukraine	372	73.66	133	26.34
United Kingdom	4460	39.58	6809	60.42
Total	24857	44.50	31004	55.50

1. Demographics

Table A1.2 Age at follow-up: descriptive statistics, by country and overall. People with CF alive on 31/12/2023.

Country	Number	Mean (average age)	Min (age of the youngest pwCF)	25th pctl (25% of the pwCF are younger than this age)	Median (half the pwCF are younger than this age)	75th pctl (75% of the pwCF are younger than this age)	Max (age of the oldest pwCF)
Albania	96	11.9	0.5	8.2	12.1	16.3	22.0
Armenia	31	10.7	0.3	5.7	9.9	14.2	26.0
Austria	875	22.9	0.1	11.3	21.1	33.0	69.5
Belarus	151	10.2	0.5	5.9	10.5	14.4	20.0
Bulgaria	235	18.1	0.4	8.3	16.0	26.1	69.5
Croatia	149	17.4	1.0	8.2	16.7	24.7	39.5
Cyprus	31	28.0	5.2	13.4	22.6	41.1	69.2
Czech Republic	735	20.2	0.1	9.4	18.7	28.6	68.0
Denmark	576	25.5	0.0	11.7	25.0	36.3	74.0
Estonia	44	18.5	1.2	7.7	15.5	26.7	54.1
Finland	76	24.3	0.7	13.8	21.8	33.1	58.3
France	7293	25.6	0.0	13.5	24.0	36.0	88.0
Georgia	82	8.5	0.5	3.5	8.6	13.0	20.3
Germany	7525	24.8	0.0	12.0	23.7	35.4	86.7
Greece	684	24.3	0.3	14.0	23.8	32.4	76.8
Hungary	501	19.9	0.1	8.6	18.0	30.0	74.9
Iceland	16	17.7	1.5	8.1	15.2	25.9	43.0
Ireland	1375	24.5	0.2	12.8	23.4	35.5	74.7
Israel	582	27.3	0.8	16.5	25.6	36.4	78.5
Italy	6119	26.5	0.1	13.0	24.0	38.0	91.1
Kazakhstan	27	22.3	16.0	17.5	20.0	23.7	43.3
Latvia	53	14.3	0.2	5.5	12.6	20.9	37.0
Lithuania	48	19.7	0.9	10.9	20.1	26.6	39.5
Luxembourg	26	9.7	0.3	3.6	6.8	13.6	33.2
Rep of Moldova	49	14.5	2.2	8.3	12.9	18.5	37.7
Montenegro	40	13.0	0.8	6.1	11.0	19.0	43.4
The Netherlands	1636	27.5	0.0	14.8	26.3	37.2	75.2
North Macedonia	166	16.3	0.2	7.0	15.2	22.6	44.6
Norway	387	27.7	0.4	12.8	27.3	41.0	83.6
Poland	1703	17.0	0.1	8.5	15.0	23.5	63.5
Portugal	407	22.9	0.5	11.7	20.8	32.4	70.0
Romania	414	12.5	0.2	6.0	11.6	17.5	46.0
Russian Fed.	4061	14.9	0.1	7.8	13.0	19.7	66.0
Serbia	201	15.8	0.8	6.2	14.5	22.0	50.1
Slovak Republic	309	22.6	0.5	11.2	21.4	32.6	84.0
Slovenia	119	21.8	1.9	12.0	21.0	29.2	68.1
Spain	2655	24.8	0.1	11.7	21.5	36.3	88.2
Sweden	801	27.5	0.4	14.1	25.9	39.4	79.6
Switzerland	1077	25.1	0.1	12.6	23.6	34.4	83.0
Türkiye	2732	11.3	0.0	5.5	9.6	15.8	50.0
Ukraine	505	13.6	0.0	7.6	12.5	18.5	43.7
United Kingdom	11269	24.8	0.1	12.1	23.0	35.0	89.4
Total	55861	23.0	0.0	10.7	20.3	33.2	91.1

2. Diagnosis

Table A2.1 Age at diagnosis (in years): descriptive statistics, by country and overall. All children and adolescents (<18 years) alive and seen in 2023.

Country	Number	Number of missing	Mean (average age at diagnosis)	Min (lowest age at diagnosis)	25th pctl (25% of the pwCF were diagnosed before this age)	Median (half the pwCF were diagnosed before this age)	75th pctl (75% of the pwCF were diagnosed before this age)	Max (highest age at diagnosis)
Albania	50	0	0.40	0	0.16	0.20	0.33	4.00
Armenia	25	0	2.16	0.10	0.33	1.10	2.00	10.0
Austria	354	8	0.50	0	0.10	0.10	0.20	14.8
Belarus	140	0	1.65	0.01	0.10	0.37	1.55	11.0
Bulgaria	131	3	2.42	0	0.30	0.70	3.00	17.2
Croatia	78	2	1.05	0	0.10	0.42	0.85	7.10
Cyprus	5	1	0.48	0.01	0.03	0.38	0.87	1.10
Czech Republic	351	1	0.61	0	0.10	0.10	0.30	12.2
Denmark	213	0	0.78	0	0	0	0.67	14.9
Estonia	25	0	1.66	0	0.30	0.40	2.20	9.10
Finland	30	0	1.09	0	0.10	0.46	1.08	5.00
France	2550	45	0.32	0	0.10	0.10	0.10	14.9
Georgia	53	0	2.89	0	0.20	1	5.10	13.0
Germany	2749	50	1.11	0	0.08	0.17	1.00	16.5
Greece	212	25	1.01	0	0.17	0.48	0.97	9.97
Hungary	251	0	1.96	0	0.17	0.66	2.75	17.0
Iceland	9	0	0.73	0	0.10	0.20	1.00	3.20
Ireland	523	0	0.71	0	0.04	0.06	0.29	12.7
Israel	161	8	1.38	0	0	0.30	1.40	13.0
Italy	2163	37	0.74	0	0.07	0.13	0.35	17.3
Kazakhstan	9	0	5.33	0.50	1	5	6.00	15.0
Latvia	36	0	0.84	0	0	0.20	0.60	6.10
Lithuania	18	2	3.16	0	0.70	1.85	3.50	16.6
Luxembourg	20	0	0.25	0	0	0.10	0.30	1.30
Rep of Moldova	30	0	1.20	0.10	0.30	0.40	0.75	8.10
Montenegro	29	0	1.74	0	0.20	0.40	1.40	10.1
The Netherlands	527	3	0.83	0	0	0.10	0.30	17.0
North Macedonia	84	0	1.26	0	0.18	0.30	1.50	7.90
Norway	136	1	0.80	0	0.10	0.10	0.30	11.6
Poland	977	17	0.72	0	0.10	0.10	0.20	16.9
Portugal	175	0	1.29	0	0.10	0.20	1.30	12.5
Romania	298	11	1.59	0	0.20	0.40	1.20	17.0
Russian Fed.	2085	0	0.97	0	0.10	0.20	0.50	17.3
Serbia	116	0	1.60	0	0.20	0.55	2.10	15.1
Slovak Republic	109	14	0.86	0	0.07	0.10	0.25	11.0
Slovenia	53	0	1.14	0	0.10	0.40	1.70	5.60
Spain	1046	6	0.66	0	0.10	0.10	0.30	14.7
Sweden	275	3	1.74	0	0.13	0.50	2.15	14.2
Switzerland	378	9	0.65	0	0	0.10	0.20	12.4
Türkiye	2228	16	1.17	0	0.17	0.30	0.67	17.0
Ukraine	323	5	2.02	0	0.20	0.70	2.50	16.0
United Kingdom	4105	0	0.35	0	0.04	0.06	0.08	16.1
Total	23130	267	0.82	0	0.07	0.1	0.41	17.3

Note: For Cyprus, Greece, Lithuania and the Slovak Republic the information on age at diagnosis is missing for more than 10% of the people with CF.

2. Diagnosis

Table A2.2 Age at diagnosis (in years): descriptive statistics, by country and overall. All adults (≥18 years) alive and seen in 2023.

Country	Number	Number of missing	Mean (average age at diagnosis)	Min (lowest age at diagnosis)	25th pctl (25% of the pwCF were diagnosed before this age)	Median (half the pwCF were diagnosed before this age)	75th pctl (75% of the pwCF were diagnosed before this age)	Max (highest age at diagnosis)
Armenia	6	0	8.19	0.50	0.75	1.34	21.2	24.0
Austria	443	48	3.66	0	0.10	0.28	2.11	58.6
Belarus	11	0	3.17	0.03	0.10	0.70	5.00	12.0
Bulgaria	93	1	9.26	0.10	0.70	3.00	15.3	64.3
Croatia	63	0	3.01	0.08	0.20	0.50	3.00	31.0
Cyprus	15	1	12.1	0.11	0.61	5.69	17.1	66.1
Czech Republic	351	7	5.09	0	0.30	1.00	4.80	53.9
Denmark	358	2	3.63	0	0.17	0.59	3.00	42.7
Estonia	19	0	4.05	0.10	0.40	1.50	6.00	25.0
Finland	35	11	5.00	0	0.30	2.00	4.00	37.0
France	4646	52	6.85	0	0.10	0.50	6.60	81.2
Germany	4180	178	5.46	0	0.17	1.00	4.92	72.7
Greece	329	55	4.62	0	0.25	0.62	4.35	48.7
Hungary	247	3	5.02	0	0.25	1.00	4.00	42.7
Iceland	7	0	0.27	0.10	0.20	0.25	0.40	0.40
Ireland	826	0	4.61	0	0.12	0.50	3.54	71.8
Israel	365	2	7.50	0	0.17	0.70	10.0	62.0
Italy	3823	85	10.1	0	0.16	1.04	15.3	77.6
Kazakhstan	18	0	14.5	3.00	8.00	10.5	20.0	43.0
Latvia	17	0	6.02	0.10	0.67	3.00	12.0	25.6
Lithuania	25	1	8.26	0	1.50	7.50	11.9	27.1
Rep of Moldova	14	0	6.66	0.20	0.40	0.95	15.0	23.0
Montenegro	11	0	3.88	0.20	0.30	0.70	6.10	20.5
The Netherlands	995	88	6.53	0	0.20	1.00	5.80	68.0
North Macedonia	54	0	2.57	0	0.20	0.30	2.40	29.2
Norway	242	2	10.6	0	0.40	2.20	15.5	77.2
Poland	608	5	6.80	0	0.50	2.20	9.00	54.1
Portugal	219	0	12.1	0	0.66	6.00	19.0	58.0
Romania	83	2	6.90	0.08	0.50	1.00	13.0	43.0
Russian Fed.	996	0	8.01	0	0.90	4.20	12.8	59.5
Serbia	55	1	4.30	0.10	0.30	2.10	6.20	18.6
Slovak Republic	154	3	6.91	0	0.20	1.35	9.50	59.0
Slovenia	63	2	3.98	0	0.20	0.70	4.50	37.4
Spain	1451	24	9.78	0	0.30	1.40	13.0	75.0
Sweden	495	6	6.41	0	0.22	1.00	5.00	70.6
Switzerland	511	94	6.31	0	0.20	1.00	5.00	75.2
Türkiye	472	1	7.39	0	0.33	3.00	13.0	43.6
Ukraine	100	1	7.23	0	1.00	3.05	9.80	38.5
United Kingdom	6211	3	6.90	0	0.08	0.59	5.02	81.4
Total	28619	678	7.11	0	0.17	0.92	7.50	81.4

Note: Albania, Georgia and Luxembourg have <5 adults seen in 2023 and are excluded from the table, but the people are included in the total number.

For Austria, Finland, Greece and Switzerland the information on age at diagnosis is missing for more than 10% of the people with CF.

4. Lung function

Table A4.1 FEV_1% of predicted for children and adolescents: descriptive statistics, by country and overall. People with CF aged 6-17 years who have never had an lung/liver transplant.

Country	Number	Number of missing	Mean (average FEV1% value)	Min (lowest FEV1% value)	25th pctl (25% of the pwCF have a FEV1% lower than the value)	Median (half the pwCF have a FEV1% lower than the value)	75th pctl (75% of the pwCF have a FEV1% lower than the value)	Max (highest FEV1% value)
Albania	29	10	75.5	30.4	67.2	76.0	86.5	109.5
Armenia	17	0	88.7	34.1	75.6	87.7	100.9	132.0
Austria	267	3	101.2	34.0	92.3	102.6	110.9	151.1
Belarus	38	19	76.5	44.8	60.5	79.5	89.8	103.2
Bulgaria	95	3	85.9	17.5	72.5	90.3	101	151.9
Croatia	53	2	88.0	26.8	79.8	92.9	100.8	131.9
Czech Republic	242	2	103.4	33.1	96.1	103.9	112.3	161.5
Denmark	129	2	106.1	36.2	97.0	106.7	116.5	138.4
Estonia	18	1	90.5	46.3	79.5	97.8	104.6	117.3
Finland	23	0	90.1	56.9	75.6	94.1	101	125.3
France	1845	55	99.7	27.5	91.1	101.0	110	161.7
Georgia	16	14	84.9	53.0	67.7	82.6	99.8	133.9
Germany	1909	55	99.3	17.0	90.6	100.3	109.1	149.6
Greece	179	14	103.3	50.7	93.4	104.4	114.9	149.9
Hungary	158	13	83.5	24.8	72.8	83.4	97.7	132.0
Iceland	6	0	102.8	83.6	99.3	103.7	109.7	116.6
Ireland	374	11	100.1	30.9	92.4	101.1	108.2	142.3
Israel	142	1	92.3	24.7	82.2	93.7	105.2	137.5
Italy	1580	74	102.6	31.9	93.1	103.5	113.5	154.1
Latvia	18	3	84.4	52.3	76.2	83.5	96.9	112.4
Lithuania	14	0	90.6	35.7	84.9	95.6	102.8	122.0
Luxembourg	8	0	98.0	80.4	89.3	100.9	105	112.9
Rep of Moldova	18	4	78.8	31.3	56.9	80.4	98.4	113.8
Montenegro	19	0	100.5	70.9	93.2	100.4	111.1	114.2
The Netherlands	396	3	100.0	39.4	91.9	100.2	108.9	137.4
North Macedonia	52	2	78.3	42.0	64.8	78.6	92.6	113.5
Norway	82	1	101.3	44.8	92.4	102.8	109.4	133.6
Poland	673	44	96.2	22.1	86.6	99.2	108.8	143.7
Portugal	126	7	97.4	40.3	87.6	97.6	109.3	131.5
Romania	176	35	90.7	34.6	82.2	94.1	101.3	137.6
Russian Fed.	1233	300	88.9	14.1	78.1	91.7	103.3	169.7
Serbia	66	3	89.0	31.5	81.7	92.1	102.1	122.0
Slovak Republic	91	1	95.9	59.6	87.1	96.4	104.5	136.8
Slovenia	48	0	98.6	67.9	92.8	99.9	107.8	116.8
Spain	779	26	100.4	30.0	91.0	100.9	111.2	151.3
Sweden	201	3	96.2	38.5	86.7	97.4	106.6	137.5
Switzerland	294	1	102.3	54.1	93.3	103.9	111.4	137.8
Türkiye	989	487	84.4	14.5	71.5	87.1	99.9	177.8
Ukraine	170	74	85.8	13.8	72.7	87.7	99.8	154.5
United Kingdom	2862	187	97.7	23.5	89.1	98.8	107.3	164.5
Total	15440	1470	96.8	13.8	87.7	98.8	108.3	177.8

Note: Cyprus and Kazakhstan have <5 individuals aged 6-17 years with FEV1 measurement and are excluded from the table.

Note: Finland and Sweden report the best FEV1pp in the 12 months prior to the annual review which means, in some cases, that the value can be from the previous calendar year.

The UK reports the best FEV1pp from the period between annual reviews which means, in some cases, that the value can be from the previous calendar year.

This table shows some descriptive statistics for FEV_1 in children and young people, expressed as a percentage of predicted. Note that people with CF who have had an lung/liver transplant and children below 6 years of age have been excluded from the analyses.

4. Lung function

Table A4.2 FEV_1% of predicted for adults: descriptive statistics, by country for adults with CF (>18 years) who have never had a transplant.

Country	Number	Number of missing	Mean (average FEV1% value)	Min (lowest FEV1% value)	25th pctl (25% of the pwCF have a FEV1% lower than the value)	Median (half the pwCF have a FEV1% lower than the value)	75th pctl (75% of the pwCF have a FEV1% lower than the value)	Max (highest FEV1% value)
Armenia	5	1	78.0	40.7	76.2	85.9	88.6	98.7
Austria	381	2	83.1	17.1	65.6	85.8	100.0	138.3
Bulgaria	85	4	69.6	19.6	51.4	72.2	89.6	116.5
Croatia	56	0	81.3	20.9	62.3	87.3	102.3	112.2
Cyprus	13	1	59.2	26.9	42.7	57.7	67.5	95.5
Czech Republic	295	4	80.1	21.1	64.7	86.6	97.9	123.1
Denmark	299	0	87.6	25.0	72.4	92.5	103.7	138.5
Estonia	13	1	45.8	21.4	32.6	41.0	49.5	98.3
Finland	36	0	70.9	32.0	54.0	72.7	88.7	109.8
France	3656	54	79.1	17.9	60.9	81.4	97.0	147.8
Germany	3855	66	76.2	14.7	57.3	78.5	95.5	142.7
Greece	314	34	81.5	25.4	62.8	84.8	99.9	141.7
Hungary	189	0	65.1	15.3	42.6	66.5	87.2	127.6
Iceland	7	0	88.9	70.3	77.5	93.8	94.8	103.2
Ireland	662	63	76.5	17.8	59.1	80.3	94.4	176.8
Israel	337	2	74.2	23.9	58.4	75.3	90.9	123.6
Italy	3297	179	82.1	12.4	63.7	85.9	100.6	145.1
Kazakhstan	11	5	51.6	23.4	30.4	48.1	66.7	105.8
Latvia	17	0	65.0	16.9	49.5	65.8	78.5	103.5
Lithuania	22	0	58.2	14.5	29.1	67.6	77.0	103.0
Rep of Moldova	12	1	57.5	23.1	42.2	62.5	70.6	85.2
Montenegro	7	0	78.3	54.5	56.2	70.1	103.4	110.9
The Netherlands	944	9	78.3	17.2	60.4	80.0	96.5	131.2
North Macedonia	47	0	66.4	21.1	51.7	67.8	78.4	115.3
Norway	201	1	80.2	16.9	66.3	82.9	97.3	126.0
Poland	529	16	74.5	18.4	53.9	76.1	95.6	137.0
Portugal	182	9	76.9	30.6	59.4	76.7	96.0	122.7
Romania	70	0	71.2	18.2	49.3	74.9	93.0	114.2
Russian Fed.	693	230	60.2	3.5	38.9	58.0	79.6	138.5
Serbia	54	0	62.8	24.9	38.9	61.8	78.8	108.4
Slovak Republic	138	0	76.2	20.8	63.0	80.3	91.9	123.5
Slovenia	42	3	74.9	23.1	52.6	80.4	99.0	122.7
Spain	1202	30	78.2	18.4	61.5	80.4	96.1	134.0
Sweden	413	3	80.5	20.8	65.4	82.9	97.2	126.9
Switzerland	522	1	77.2	14.5	56.8	79.2	95.2	162.6
Türkiye	365	77	67.1	18.3	45.6	67.4	88.2	117.6
Ukraine	82	12	61.8	14.9	43.1	63.6	79.9	113.7
United Kingdom	5552	273	75.6	11.0	58.7	78.1	93.0	157.2
Total	24610	1082	77.0	3.5	58.6	79.6	95.8	176.8

Note: Albania, Belarus, Georgia and Luxembourg have <5 adults with FEV_1 measurement and are excluded from the table, but the people are included in the total number.

Note: Finland and Sweden report the best FEV1pp in the 12 months prior to the annual review which means, in some cases, that the value can be from the previous calendar year.

The UK reports the best FEV1pp from the period between annual reviews which means, in some cases, that the value can be from the previous calendar year.

This table shows some descriptive statistics for FEV_1 in adults with CF, expressed as a percentage of predicted. Note that adults who have had a transplant have been excluded from the analyses.

5. Microbiology

Table A5.1 Prevalence of Pseudomonas aeruginosa in people with CF seen in 2023 who have never had a transplant, by country and overall.

Country	Children (<18 years)								Adults (≥18 years)							
	Missing/ Unknown		No		Yes, chronic		Yes, not chronic/ intermittent		Missing/ Unknown		No		Yes, chronic		Yes, not chronic/ intermittent	
	N	%	N	%	N	%	N	%	N	%	N	%	N	%	N	%
Albania	0	0.0	28	56.0	7	14.0	15	30.0								
Armenia	0	0.0	17	65.4	5	19.2	4	15.4	0	0.0	5	83.3	1	16.7	0	0.0
Austria	0	0.0	308	86.0	18	5.0	32	8.9	3	0.7	241	59.7	132	32.7	28	6.9
Belarus	0	0.0	97	68.8	44	31.2	0	0.0	0	0.0	7	63.6	4	36.4	0	0.0
Bulgaria	0	0.0	95	70.4	14	10.4	26	19.3	0	0.0	60	63.8	23	24.5	11	11.7
Croatia	0	0.0	59	74.7	4	5.1	16	20.3	0	0.0	34	58.6	17	29.3	7	12.1
Cyprus	0	0.0	4	66.7	2	33.3	0	0.0	3	20.0	5	33.3	5	33.3	2	13.3
Czech Rep.	5	1.4	301	86.0	17	4.9	27	7.7	19	6.0	193	61.3	83	26.4	20	6.4
Denmark	0	0.0	198	92.5	12	5.6	4	1.9	1	0.3	242	74.9	63	19.5	17	5.3
Estonia	0	0.0	21	84.0	1	4.0	3	12.0	1	5.6	8	44.4	8	44.4	1	5.6
Finland	0	0.0	27	90.0	2	6.7	1	3.3	0	0.0	26	72.2	9	25.0	1	2.8
France	0	0.0	2203	85.1	87	3.4	299	11.6	0	0.0	2739	70.1	872	22.3	295	7.6
Georgia	3	5.7	39	73.6	3	5.7	8	15.1								
Germany	19	0.7	2419	87.0	200	7.2	144	5.2	112	2.8	1897	47.2	1860	46.3	153	3.8
Greece	0	0.0	195	82.6	8	3.4	33	14.0	5	1.3	150	39.7	209	55.3	14	3.7
Hungary	0	0.0	159	63.4	32	12.8	60	23.9	0	0.0	67	34.2	100	51.0	29	14.8
Iceland	0	0.0	8	88.9	1	11.1	0	0.0	0	0.0	5	71.4	0	0.0	2	28.6
Ireland	0	0.0	488	93.3	11	2.1	24	4.6	1	0.1	600	80.8	76	10.2	66	8.9
Israel	12	7.1	111	65.7	18	10.7	28	16.6	29	8.2	159	44.8	116	32.7	51	14.4
Italy	5	0.2	1724	78.8	127	5.8	332	15.2	7	0.2	2049	56.2	1064	29.2	525	14.4
Kazakhstan	0	0.0	4	44.4	5	55.6	0	0.0	0	0.0	4	22.2	14	77.8	0	0.0
Latvia	2	5.6	27	75.0	6	16.7	1	2.8	0	0.0	6	35.3	9	52.9	2	11.8
Lithuania	0	0.0	17	85.0	2	10.0	1	5.0	0	0.0	17	65.4	8	30.8	1	3.9
Luxembourg	1	5.0	17	85.0	0	0.0	2	10.0	0	0.0	2	40.0	3	60.0	0	0.0
Rep. Moldova	1	3.2	15	48.4	13	41.9	2	6.5	0	0.0	2	13.3	13	86.7	0	0.0
Montenegro	0	0.0	25	86.2	2	6.9	2	6.9	0	0.0	8	72.7	1	9.1	2	18.2
Netherlands	10	1.9	485	91.5	17	3.2	18	3.4	149	15.2	523	53.3	249	25.4	61	6.2
N. Macedonia	0	0.0	52	61.9	14	16.7	18	21.4	2	3.8	17	32.1	28	52.8	6	11.3
Norway	2	1.5	132	96.4	1	0.7	2	1.5	19	9.2	130	63.1	55	26.7	2	1.0
Poland	17	1.7	800	81.1	71	7.2	99	10.0	4	0.7	316	54.6	206	35.6	53	9.2
Portugal	4	2.3	126	73.3	18	10.5	24	14.0	41	20.7	91	46.0	53	26.8	13	6.6
Romania	4	1.3	209	67.6	67	21.7	29	9.4	0	0.0	48	56.5	29	34.1	8	9.4
Russian Fed.	105	5.0	1263	60.4	465	22.3	257	12.3	46	4.7	376	38.5	508	51.9	48	4.9
Serbia	0	0.0	74	63.8	19	16.4	23	19.8	1	1.8	20	36.4	28	50.9	6	10.9
Slovak Rep.	0	0.0	111	90.2	1	0.8	11	8.9	8	5.4	96	64.4	36	24.2	9	6.0
Slovenia	0	0.0	47	90.4	0	0.0	5	9.6	12	24.5	21	42.9	10	20.4	6	12.2
Spain	13	1.2	843	80.1	56	5.3	140	13.3	89	6.9	795	61.9	299	23.3	101	7.9
Sweden	17	6.1	212	76.5	20	7.2	28	10.1	16	3.8	213	50.1	172	40.5	24	5.7
Switzerland	1	0.3	347	89.9	20	5.2	18	4.7	4	0.8	348	65.2	141	26.4	41	7.7
Türkiye	19	0.8	1673	74.2	297	13.2	265	11.8	3	0.6	250	52.6	158	33.3	64	13.5
Ukraine	9	2.7	207	63.1	82	25.0	30	9.2	0	0.0	29	28.7	68	67.3	4	4.0
United Kingdom	5	0.1	3543	86.4	111	2.7	441	10.8	5	0.1	4193	70.5	728	12.2	1020	17.2
Total	254	1.1	18730	80.2	1900	8.1	2472	10.6	580	2.2	15993	59.8	7458	27.9	2696	10.1

Note: Albania and Georgia have <5 adults seen in 2023 and are excluded from the table for adults, but the people are included in the total number.

Note: Ireland, Italy: chronicity for Pseudomonas aeruginosa is defined as: at least 3 or more positive isolates during the last 12 months preceding the last reported culture in 2023.

The United Kingdom: chronicity for Pseudomonas aeruginosa is defined as: 3 or more positive isolates during the 12 months preceding the last annual review.

This table shows, separately by country and overall, the frequency of *Pseudomonas aeruginosa* in children and adults.

5. Microbiology

Table A5.2 Prevalence of Burkholderia cepacia complex species in people with CF seen in 2023 who have never had a transplant, by country and overall.

Country	Children (<18 years)								Adults (≥18 years)							
	Missing/ Unknown		No		Yes, chronic		Yes, not chronic/ intermittent		Missing/ Unknown		No		Yes, chronic		Yes, not chronic/ intermittent	
	N	%	N	%	N	%	N	%	N	%	N	%	N	%	N	%
Albania	0	0.0	48	96.0	0	0.0	2	4.0								
Armenia	13	50.0	13	50.0	0	0.0	0	0.0	2	33.3	4	66.7	0	0.0	0	0.0
Austria	1	0.3	356	99.4	0	0.0	1	0.3	3	0.7	382	94.6	18	4.5	1	0.3
Belarus	0	0.0	135	95.7	6	4.3	0	0.0	0	0.0	11	100.0	0	0.0	0	0.0
Bulgaria	0	0.0	134	99.3	0	0.0	1	0.7	1	1.1	92	97.9	0	0.0	1	1.1
Croatia	0	0.0	79	100.0	0	0.0	0	0.0	0	0.0	58	100.0	0	0.0	0	0.0
Cyprus	0	0.0	6	100.0	0	0.0	0	0.0	3	20.0	11	73.3	0	0.0	1	6.7
Czech Rep.	6	1.7	343	98.0	1	0.3	0	0.0	25	7.9	259	82.2	31	9.8	0	0.0
Denmark	0	0.0	213	99.5	0	0.0	1	0.5	1	0.3	309	95.7	8	2.5	5	1.6
Estonia	0	0.0	25	100.0	0	0.0	0	0.0	0	0.0	17	94.4	1	5.6	0	0.0
Finland	0	0.0	30	100.0	0	0.0	0	0.0	0	0.0	36	100.0	0	0.0	0	0.0
France	0	0.0	2570	99.3	7	0.3	12	0.5	0	0.0	3845	98.4	44	1.1	17	0.4
Georgia	51	96.2	2	3.8	0	0.0	0	0.0								
Germany	13	0.5	2755	99.0	10	0.4	4	0.1	102	2.5	3802	94.5	103	2.6	15	0.4
Greece	0	0.0	236	100.0	0	0.0	0	0.0	23	6.1	354	93.7	0	0.0	1	0.3
Hungary	0	0.0	245	97.6	2	0.8	4	1.6	0	0.0	185	94.4	7	3.6	4	2.0
Iceland	0	0.0	8	88.9	0	0.0	1	11.1	0	0.0	7	100.0	0	0.0	0	0.0
Ireland	0	0.0	521	99.6	1	0.2	1	0.2	1	0.1	727	97.9	6	0.8	9	1.2
Israel	13	7.7	155	91.7	0	0.0	1	0.6	28	7.9	323	91.0	3	0.9	1	0.3
Italy	5	0.2	2172	99.3	5	0.2	6	0.3	8	0.2	3548	97.3	75	2.1	14	0.4
Kazakhstan	0	0.0	9	100.0	0	0.0	0	0.0	0	0.0	18	100.0	0	0.0	0	0.0
Latvia	1	2.8	35	97.2	0	0.0	0	0.0	0	0.0	16	94.1	1	5.9	0	0.0
Lithuania	0	0.0	19	95.0	1	5.0	0	0.0	0	0.0	24	92.3	2	7.7	0	0.0
Luxembourg	1	5.0	19	95.0	0	0.0	0	0.0	0	0.0	5	100.0	0	0.0	0	0.0
Rep. Moldova	31	100.0	-	-	-	-	-	-	15	100.0	-	-	-	-	-	-
Montenegro	0	0.0	28	96.6	0	0.0	1	3.5	0	0.0	11	100.0	0	0.0	0	0.0
Netherlands	7	1.3	520	98.1	1	0.2	2	0.4	138	14.1	827	84.2	11	1.1	6	0.6
N. Macedonia	0	0.0	84	100.0	0	0.0	0	0.0	3	5.7	49	92.5	0	0.0	1	1.9
Norway	3	2.2	133	97.1	1	0.7	0	0.0	27	13.1	176	85.4	3	1.5	0	0.0
Poland	18	1.8	962	97.5	2	0.2	5	0.5	5	0.9	565	97.6	6	1.0	3	0.5
Portugal	4	2.3	164	95.4	2	1.2	2	1.2	42	21.2	148	74.8	8	4.0	0	0.0
Romania	7	2.3	302	97.7	0	0.0	0	0.0	0	0.0	84	98.8	1	1.2	0	0.0
Russian Fed.	122	5.8	1916	91.7	28	1.3	24	1.2	24	2.5	855	87.4	94	9.6	5	0.5
Serbia	0	0.0	113	97.4	1	0.9	2	1.7	1	1.8	43	78.2	10	18.2	1	1.8
Slovak Rep.	0	0.0	123	100.0	0	0.0	0	0.0	7	4.7	135	90.6	5	3.4	2	1.3
Slovenia	0	0.0	52	100.0	0	0.0	0	0.0	15	30.6	34	69.4	0	0.0	0	0.0
Spain	13	1.2	1024	97.3	6	0.6	9	0.9	97	7.6	1127	87.8	49	3.8	11	0.9
Sweden	20	7.2	253	91.3	1	0.4	3	1.1	40	9.4	372	87.5	11	2.6	2	0.5
Switzerland	0	0.0	386	100.0	0	0.0	0	0.0	2	0.4	519	97.2	10	1.9	3	0.6
Türkiye	21	0.9	2227	98.8	4	0.2	2	0.1	3	0.6	463	97.5	6	1.3	3	0.6
Ukraine	7	2.1	314	95.7	1	0.3	6	1.8	0	0.0	97	96.0	2	2.0	2	2.0
United Kingdom	6	0.2	4056	98.9	0	0.0	38	0.9	5	0.1	5776	97.1	0	0.0	165	2.8
Total	363	1.6	22785	97.6	80	0.3	128	0.6	622	2.3	25317	94.7	515	1.9	273	1.0

Note: Albania and Georgia have <5 adults seen in 2023 and are excluded from the table for adults, but the people are included in the total number.

Note: Ireland and Italy: chronicity for *Burkholderia cepacia complex* is defined as: at least 3 or more positive isolates during the last 12 months preceding the last reported culture in 2023.

The United Kingdom: chronicity for *Burkholderia cepacia complex* is not collected.

This table shows, separately by country, and overall, the frequency of *Burkholderia cepacia complex species* in children and adults. The identification rate may be influenced by differences in culture techniques employed.

5. Microbiology

Table A5.3 Prevalence of Haemophilus influenzae in people with CF seen in 2023 who have never had a transplant, by country and overall.

Country	Children (<18 years)								Adults (≥18 years)							
	Missing/ Unknown		No		Yes, chronic		Yes, not chronic/ intermittent		Missing/ Unknown		No		Yes, chronic		Yes, not chronic/ intermittent	
	N	%	N	%	N	%	N	%	N	%	N	%	N	%	N	%
Albania	0	0.0	50	100.0	0	0.0	0	0.0								
Armenia	0	0.0	23	88.5	0	0.0	3	11.5	0	0.0	3	50.0	0	0.0	3	50.0
Austria	1	0.3	226	63.1	18	5.0	113	31.6	1	0.3	308	76.2	30	7.4	65	16.1
Belarus	0	0.0	130	92.2	11	7.8	0	0.0	0	0.0	11	100.0	0	0.0	0	0.0
Bulgaria	0	0.0	131	97.0	0	0.0	4	3.0	1	1.1	92	97.9	0	0.0	1	1.1
Croatia	0	0.0	64	81.0	0	0.0	15	19.0	0	0.0	53	91.4	0	0.0	5	8.6
Cyprus	0	0.0	5	83.3	0	0.0	1	16.7	3	20.0	11	73.3	0	0.0	1	6.7
Czech Rep.	14	4.0	324	92.6	1	0.3	11	3.1	32	10.2	279	88.6	0	0.0	4	1.3
Denmark	0	0.0	157	73.4	26	12.2	31	14.5	0	0.0	280	86.7	5	1.6	38	11.8
Estonia	1	4.0	20	80.0	1	4.0	3	12.0	0	0.0	16	88.9	0	0.0	2	11.1
Finland	0	0.0	26	86.7	3	10.0	1	3.3	0	0.0	35	97.2	0	0.0	1	2.8
France	0	0.0	2086	80.6	0	0.0	503	19.4	0	0.0	3437	88.0	0	0.0	469	12.0
Georgia	5	9.4	48	90.6	0	0.0	0	0.0								
Germany	13	0.5	1970	70.8	80	2.9	719	25.8	102	2.5	3402	84.6	105	2.6	413	10.3
Greece	0	0.0	221	93.6	0	0.0	15	6.4	24	6.4	321	84.9	1	0.3	32	8.5
Hungary	2	0.8	216	86.1	1	0.4	32	12.8	0	0.0	187	95.4	1	0.5	8	4.1
Iceland	0	0.0	3	33.3	2	22.2	4	44.4	0	0.0	6	85.7	0	0.0	1	14.3
Ireland	0	0.0	390	74.6	20	3.8	113	21.6	1	0.1	695	93.5	2	0.3	45	6.1
Israel	13	7.7	122	72.2	6	3.6	28	16.6	27	7.6	292	82.3	6	1.7	30	8.5
Italy	5	0.2	1625	74.3	77	3.5	481	22.0	7	0.2	3378	92.7	30	0.8	230	6.3
Kazakhstan	0	0.0	9	100.0	0	0.0	0	0.0	0	0.0	18	100.0	0	0.0	0	0.0
Latvia	1	2.8	23	63.9	4	11.1	8	22.2	0	0.0	16	94.1	0	0.0	1	5.9
Lithuania	0	0.0	14	70.0	0	0.0	6	30.0	0	0.0	25	96.2	0	0.0	1	3.9
Luxembourg	1	5.0	12	60.0	1	5.0	6	30.0	0	0.0	4	80.0	0	0.0	1	20.0
Rep. Moldova	1	3.2	30	96.8	0	0.0	0	0.0	0	0.0	15	100.0	0	0.0	0	0.0
Montenegro	1	3.5	27	93.1	0	0.0	1	3.5	0	0.0	10	90.9	0	0.0	1	9.1
Netherlands	8	1.5	436	82.3	5	0.9	81	15.3	134	13.7	742	75.6	27	2.8	79	8.0
N. Macedonia	0	0.0	84	100.0	0	0.0	0	0.0	2	3.8	50	94.3	0	0.0	1	1.9
Norway	2	1.5	71	51.8	8	5.8	56	40.9	27	13.1	153	74.3	5	2.4	21	10.2
Poland	18	1.8	736	74.6	16	1.6	217	22.0	5	0.9	505	87.2	3	0.5	66	11.4
Portugal	5	2.9	116	67.4	8	4.7	43	25.0	40	20.2	136	68.7	3	1.5	19	9.6
Romania	13	4.2	283	91.6	1	0.3	12	3.9	0	0.0	83	97.7	0	0.0	2	2.4
Russian Fed.	107	5.1	1868	89.4	7	0.3	108	5.2	26	2.7	940	96.1	2	0.2	10	1.0
Serbia	0	0.0	9	7.8	78	67.2	29	25.0	1	1.8	17	30.9	16	29.1	21	38.2
Slovak Rep.	0	0.0	86	69.9	4	3.3	33	26.8	7	4.7	129	86.6	1	0.7	12	8.1
Slovenia	0	0.0	25	48.1	3	5.8	24	46.2	15	30.6	30	61.2	1	2.0	3	6.1
Spain	12	1.1	736	70.0	46	4.4	258	24.5	102	7.9	1049	81.7	22	1.7	111	8.6
Sweden	24	8.7	177	63.9	2	0.7	74	26.7	45	10.6	308	72.5	8	1.9	64	15.1
Switzerland	0	0.0	279	72.3	25	6.5	82	21.2	2	0.4	461	86.3	12	2.3	59	11.1
Türkiye	22	1.0	2082	92.4	24	1.1	126	5.6	3	0.6	449	94.5	5	1.1	18	3.8
Ukraine	7	2.1	309	94.2	1	0.3	11	3.4	0	0.0	101	100.0	0	0.0	0	0.0
United Kingdom	6	0.2	3326	81.1	0	0.0	768	18.7	5	0.1	5568	93.6	0	0.0	373	6.3
Total	282	1.2	18575	79.5	479	2.1	4020	17.2	612	2.3	23619	88.4	285	1.1	2211	8.3

Note: Albania and Georgia have <5 adults seen in 2023 and are excluded from the table for adults, but the people are included in the total number.

Note: France and UK: chronicity for Haemophilus influenza is not collected.

Ireland and Italy: chronicity for Haemophilus influenzae is defined as: at least 3 or more positive isolates during the last 12 months preceding the last reported culture in 2023.

This table shows, separately by country, and overall, the frequency of *Haemophilus influenzae* in children and adults.

5. Microbiology

Table A5.4 Prevalence of methicillin sensitive Staphylococcus aureus (MSSA) in people with CF seen in 2023 who have never had a transplant, by country and overall.

Country	Children (<18 years)								Adults (≥18 years)							
	Missing/ Unknown		No		Yes, chronic		Yes, not chronic/ intermittent		Missing/ Unknown		No		Yes, chronic		Yes, not chronic/ intermittent	
	N	%	N	%	N	%	N	%	N	%	N	%	N	%	N	%
Albania	0	0.0	24	48.0	6	12.0	20	40.0								
Armenia	0	0.0	2	7.7	5	19.2	19	73.1	0	0.0	0	0.0	0	0.0	6	100.0
Austria	1	0.3	94	26.3	153	42.7	110	30.7	1	0.3	134	33.2	183	45.3	86	21.3
Belarus	0	0.0	56	39.7	85	60.3	0	0.0	0	0.0	4	36.4	7	63.6	0	0.0
Bulgaria	0	0.0	100	74.1	3	2.2	32	23.7	1	1.1	77	81.9	2	2.1	14	14.9
Croatia	0	0.0	26	32.9	20	25.3	33	41.8	0	0.0	16	27.6	22	37.9	20	34.5
Cyprus	0	0.0	3	50.0	1	16.7	2	33.3	3	20.0	7	46.7	1	6.7	4	26.7
Czech Rep.	5	1.4	118	33.7	127	36.3	100	28.6	25	7.9	132	41.9	101	32.1	57	18.1
Denmark	0	0.0	146	68.2	25	11.7	43	20.1	0	0.0	226	70.0	50	15.5	47	14.6
Estonia	0	0.0	9	36.0	13	52.0	3	12.0	0	0.0	9	50.0	8	44.4	1	5.6
Finland	0	0.0	19	63.3	6	20.0	5	16.7	0	0.0	17	47.2	18	50.0	1	2.8
France	0	0.0	787	30.4	869	33.6	933	36.0	0	0.0	2174	55.7	1151	29.5	581	14.9
Georgia	3	5.7	23	43.4	8	15.1	19	35.9								
Germany	15	0.5	840	30.2	1082	38.9	845	30.4	109	2.7	1212	30.1	2043	50.8	658	16.4
Greece	0	0.0	127	53.8	62	26.3	47	19.9	14	3.7	175	46.3	140	37.0	49	13.0
Hungary	1	0.4	58	23.1	81	32.3	111	44.2	0	0.0	96	49.0	57	29.1	43	21.9
Iceland	0	0.0	1	11.1	6	66.7	2	22.2	0	0.0	2	28.6	1	14.3	4	57.1
Ireland	0	0.0	246	47.0	113	21.6	164	31.4	1	0.1	563	75.8	67	9.0	112	15.1
Israel	11	6.5	74	43.8	34	20.1	50	29.6	28	7.9	203	57.2	58	16.3	66	18.6
Italy	4	0.2	798	36.5	626	28.6	760	34.7	8	0.2	1673	45.9	1117	30.6	847	23.2
Kazakhstan	0	0.0	9	100.0	0	0.0	0	0.0	0	0.0	18	100.0	0	0.0	0	0.0
Latvia	1	2.8	5	13.9	24	66.7	6	16.7	0	0.0	2	11.8	12	70.6	3	17.7
Lithuania	0	0.0	3	15.0	8	40.0	9	45.0	0	0.0	10	38.5	13	50.0	3	11.5
Luxembourg	1	5.0	9	45.0	7	35.0	3	15.0	0	0.0	1	20.0	4	80.0	0	0.0
Rep. Moldova	2	6.5	0	0.0	27	87.1	2	6.5	0	0.0	0	0.0	15	100.0	0	0.0
Montenegro	1	3.5	22	75.9	0	0.0	6	20.7	0	0.0	10	90.9	0	0.0	1	9.1
Netherlands	6	1.1	273	51.5	62	11.7	189	35.7	129	13.1	547	55.7	182	18.5	124	12.6
N. Macedonia	0	0.0	40	47.6	8	9.5	36	42.9	2	3.8	40	75.5	5	9.4	6	11.3
Norway	2	1.5	35	25.6	40	29.2	60	43.8	22	10.7	65	31.6	80	38.8	39	18.9
Poland	21	2.1	212	21.5	461	46.7	293	29.7	9	1.6	198	34.2	216	37.3	156	26.9
Portugal	4	2.3	72	41.9	58	33.7	38	22.1	41	20.7	79	39.9	37	18.7	41	20.7
Romania	4	1.3	262	84.8	11	3.6	32	10.4	0	0.0	61	71.8	11	12.9	13	15.3
Russian Fed.	99	4.7	550	26.3	1123	53.7	318	15.2	35	3.6	332	34.0	576	58.9	35	3.6
Serbia	0	0.0	17	14.7	66	56.9	33	28.5	1	1.8	17	30.9	20	36.4	17	30.9
Slovak Rep.	0	0.0	37	30.1	38	30.9	48	39.0	8	5.4	78	52.4	31	20.8	32	21.5
Slovenia	0	0.0	8	15.4	30	57.7	14	26.9	14	28.6	10	20.4	14	28.6	11	22.5
Spain	14	1.3	382	36.3	266	25.3	390	37.1	88	6.9	617	48.1	313	24.4	266	20.7
Sweden	23	8.3	98	35.4	48	17.3	108	39.0	39	9.2	165	38.8	159	37.4	62	14.6
Switzerland	0	0.0	98	25.4	184	47.7	104	26.9	1	0.2	243	45.5	185	34.6	105	19.7
Türkiye	20	0.9	1587	70.4	284	12.6	363	16.1	3	0.6	272	57.3	118	24.8	82	17.3
Ukraine	6	1.8	115	35.1	155	47.3	52	15.9	0	0.0	40	39.6	53	52.5	8	7.9
United Kingdom	6	0.2	3394	82.8	259	6.3	441	10.8	5	0.1	4622	77.7	299	5.0	1020	17.2
Total	250	1.1	10779	46.2	6484	27.8	5843	25.0	587	2.2	14150	52.9	7369	27.6	4621	17.3

Note: Albania and Georgia have <5 adults seen in 2023 and are excluded from the table for adults, but the people are included in the total number.

Note: Ireland and Italy: chronicity for Staphylococcus Aureus is defined as: at least 3 or more positive isolates during the last 12 months preceding the last reported culture in 2023.

The United Kingdom: chronicity for Staphylococcus Aureus is defined as: 3 or more positive isolates during the 12 months preceding last annual review.

This table shows the frequency of MSSA in children and adults, by country and overall.

5. Microbiology

Table A5.5 Prevalence of methicillin-resistant Staphylococcus aureus (MRSA) in people with CF seen in 2023 who have never had a transplant, by country and overall.

Country	Children (<18 years)								Adults (≥18 years)							
	Missing/ Unknown		No		Yes, chronic		Yes, not chronic/ intermittent		Missing/ Unknown		No		Yes, chronic		Yes, not chronic/ intermittent	
	N	%	N	%	N	%	N	%	N	%	N	%	N	%	N	%
Albania	0	0.0	41	82.0	2	4.0	7	14.0								
Armenia	1	3.9	25	96.2	0	0.0	0	0.0	0	0.0	6	100.0	0	0.0	0	0.0
Austria	4	1.1	344	96.1	3	0.8	7	2.0	3	0.7	389	96.3	9	2.2	3	0.7
Belarus	0	0.0	126	89.4	15	10.6	0	0.0	0	0.0	11	100.0	0	0.0	0	0.0
Bulgaria	1	0.7	134	99.3	0	0.0	0	0.0	1	1.1	93	98.9	0	0.0	0	0.0
Croatia	0	0.0	77	97.5	0	0.0	2	2.5	0	0.0	58	100.0	0	0.0	0	0.0
Cyprus	0	0.0	3	50.0	1	16.7	2	33.3	3	20.0	10	66.7	1	6.7	1	6.7
Czech Rep.	6	1.7	342	97.7	0	0.0	2	0.6	26	8.3	280	88.9	0	0.0	9	2.9
Denmark	0	0.0	212	99.1	1	0.5	1	0.5	0	0.0	315	97.5	1	0.3	7	2.2
Estonia	1	4.0	23	92.0	0	0.0	1	4.0	0	0.0	16	88.9	2	11.1	0	0.0
Finland	0	0.0	30	100.0	0	0.0	0	0.0	0	0.0	34	94.4	2	5.6	0	0.0
France	0	0.0	2543	98.2	20	0.8	26	1.0	0	0.0	3794	97.1	74	1.9	38	1.0
Georgia	34	64.2	19	35.9	0	0.0	0	0.0								
Germany	18	0.7	2687	96.6	46	1.7	31	1.1	119	3.0	3671	91.3	195	4.9	37	0.9
Greece	0	0.0	210	89.0	9	3.8	17	7.2	22	5.8	322	85.2	14	3.7	20	5.3
Hungary	0	0.0	240	95.6	4	1.6	7	2.8	0	0.0	175	89.3	10	5.1	11	5.6
Iceland	0	0.0	9	100.0	0	0.0	0	0.0	0	0.0	7	100.0	0	0.0	0	0.0
Ireland	0	0.0	511	97.7	5	1.0	7	1.3	1	0.1	733	98.7	4	0.5	5	0.7
Israel	15	8.9	140	82.8	3	1.8	11	6.5	34	9.6	294	82.8	5	1.4	22	6.2
Italy	5	0.2	1989	90.9	58	2.7	136	6.2	7	0.2	3398	93.2	94	2.6	146	4.0
Kazakhstan	0	0.0	9	100.0	0	0.0	0	0.0	0	0.0	18	100.0	0	0.0	0	0.0
Latvia	1	2.8	35	97.2	0	0.0	0	0.0	0	0.0	15	88.2	1	5.9	1	5.9
Lithuania	0	0.0	20	100.0	0	0.0	0	0.0	0	0.0	26	100.0	0	0.0	0	0.0
Luxembourg	1	5.0	19	95.0	0	0.0	0	0.0	0	0.0	4	80.0	1	20.0	0	0.0
Rep. Moldova	2	6.5	29	93.6	0	0.0	0	0.0	0	0.0	15	100.0	0	0.0	0	0.0
Montenegro	0	0.0	28	96.6	0	0.0	1	3.5	0	0.0	11	100.0	0	0.0	0	0.0
Netherlands	9	1.7	516	97.4	2	0.4	3	0.6	134	13.7	823	83.8	21	2.1	4	0.4
N. Macedonia	0	0.0	61	72.6	7	8.3	16	19.1	2	3.8	35	66.0	6	11.3	10	18.9
Norway	3	2.2	133	97.1	1	0.7	0	0.0	27	13.1	179	86.9	0	0.0	0	0.0
Poland	22	2.2	933	94.5	15	1.5	17	1.7	10	1.7	553	95.5	11	1.9	5	0.9
Portugal	5	2.9	161	93.6	3	1.7	3	1.7	44	22.2	141	71.2	7	3.5	6	3.0
Romania	11	3.6	252	81.6	15	4.9	31	10.0	4	4.7	68	80.0	7	8.2	6	7.1
Russian Fed.	110	5.3	1879	89.9	30	1.4	71	3.4	25	2.6	905	92.5	31	3.2	17	1.7
Serbia	0	0.0	99	85.3	7	6.0	10	8.6	0	0.0	48	87.3	2	3.6	5	9.1
Slovak Rep.	0	0.0	119	96.8	0	0.0	4	3.3	8	5.4	134	89.9	3	2.0	4	2.7
Slovenia	0	0.0	48	92.3	2	3.9	2	3.9	15	30.6	32	65.3	2	4.1	0	0.0
Spain	15	1.4	995	94.6	10	1.0	32	3.0	102	7.9	1125	87.6	27	2.1	30	2.3
Sweden	19	6.9	247	89.2	2	0.7	9	3.3	41	9.7	380	89.4	2	0.5	2	0.5
Switzerland	1	0.3	376	97.4	6	1.6	3	0.8	2	0.4	520	97.4	8	1.5	4	0.8
Türkiye	20	0.9	2038	90.4	80	3.6	116	5.2	3	0.6	425	89.5	23	4.8	24	5.1
Ukraine	6	1.8	287	87.5	25	7.6	10	3.1	2	2.0	93	92.1	2	2.0	4	4.0
United Kingdom	5	0.1	4024	98.2	0	0.0	71	1.7	5	0.1	5835	98.1	0	0.0	106	1.8
Total	315	1.4	22013	94.3	372	1.6	656	2.8	641	2.4	24994	93.5	565	2.1	527	2.0

Note: Albania and Georgia have <5 adults seen in 2023 and are excluded from the table for adults, but the people are included in the total number.

Note: Ireland and Italy: chronicity for methicillin-resistant Staphylococcus Aureus is defined as: at least 3 or more positive isolates during the last 12 months preceding the last reported culture in 20223.

The United Kingdom: chronicity for methicillin-resistant Staphylococcus Aureus is not collected.

This table shows the frequency of MSSA in children and adults, by country and overall.

5. Microbiology

Table A5.6 Prevalence of Stenotrophomonas maltophilia in people with CF seen in 2023 who have never had a transplant, by country and overall.

Country	Children (<18 years)								Adults (≥18 years)							
	Missing/ Unknown		No		Yes, chronic		Yes, not chronic/ intermittent		Missing/ Unknown		No		Yes, chronic		Yes, not chronic/ intermittent	
	N	%	N	%	N	%	N	%	N	%	N	%	N	%	N	%
Albania	0	0.0	49	98.0	0	0.0	1	2.0								
Armenia	14	53.9	12	46.2	0	0.0	0	0.0	2	33.3	4	66.7	0	0.0	0	0.0
Austria	1	0.3	333	93.0	2	0.6	22	6.2	2	0.5	361	89.4	10	2.5	31	7.7
Belarus	0	0.0	139	98.6	2	1.4	0	0.0	0	0.0	11	100.0	0	0.0	0	0.0
Bulgaria	0	0.0	133	98.5	0	0.0	2	1.5	1	1.1	93	98.9	0	0.0	0	0.0
Croatia	0	0.0	73	92.4	1	1.3	5	6.3	0	0.0	54	93.1	0	0.0	4	6.9
Cyprus	0	0.0	6	100.0	0	0.0	0	0.0	3	20.0	11	73.3	0	0.0	1	6.7
Czech Rep.	6	1.7	344	98.3	0	0.0	0	0.0	26	8.3	287	91.1	0	0.0	2	0.6
Denmark	0	0.0	209	97.7	1	0.5	4	1.9	0	0.0	295	91.3	8	2.5	20	6.2
Estonia	0	0.0	24	96.0	0	0.0	1	4.0	0	0.0	17	94.4	0	0.0	1	5.6
Finland	0	0.0	27	90.0	2	6.7	1	3.3	0	0.0	35	97.2	1	2.8	0	0.0
France	0	0.0	2387	92.2	20	0.8	182	7.0	0	0.0	3755	96.1	37	1.0	114	2.9
Georgia	53	100.0	-	-	-	-	-	-								
Germany	13	0.5	2660	95.6	20	0.7	89	3.2	102	2.5	3545	88.1	241	6.0	134	3.3
Greece	0	0.0	227	96.2	0	0.0	9	3.8	22	5.8	341	90.2	5	1.3	10	2.7
Hungary	0	0.0	245	97.6	0	0.0	6	2.4	0	0.0	193	98.5	2	1.0	1	0.5
Iceland	0	0.0	8	88.9	1	11.1	0	0.0	0	0.0	6	85.7	1	14.3	0	0.0
Ireland	0	0.0	513	98.1	2	0.4	8	1.5	1	0.1	732	98.5	1	0.1	9	1.2
Israel	13	7.7	147	87.0	2	1.2	7	4.1	30	8.5	311	87.6	5	1.4	9	2.5
Italy	5	0.2	2012	92.0	12	0.6	159	7.3	7	0.2	3420	93.8	39	1.1	179	4.9
Kazakhstan	0	0.0	9	100.0	0	0.0	0	0.0	0	0.0	18	100.0	0	0.0	0	0.0
Latvia	2	5.6	30	83.3	1	2.8	3	8.3	0	0.0	15	88.2	0	0.0	2	11.8
Lithuania	2	10.0	16	80.0	1	5.0	1	5.0	0	0.0	23	88.5	1	3.9	2	7.7
Luxembourg	1	5.0	17	85.0	0	0.0	2	10.0	0	0.0	5	100.0	0	0.0	0	0.0
Rep. Moldova	31	100.0	-	-	-	-	-	-	15	100.0	-	-	-	-	-	-
Montenegro	0	0.0	28	96.6	0	0.0	1	3.5	0	0.0	11	100.0	0	0.0	0	0.0
Netherlands	10	1.9	507	95.7	0	0.0	13	2.5	183	18.6	770	78.4	12	1.2	17	1.7
N. Macedonia	0	0.0	75	89.3	0	0.0	9	10.7	2	3.8	46	86.8	0	0.0	5	9.4
Norway	2	1.5	126	92.0	1	0.7	8	5.8	28	13.6	158	76.7	8	3.9	12	5.8
Poland	20	2.0	932	94.4	2	0.2	33	3.3	6	1.0	552	95.3	6	1.0	15	2.6
Portugal	4	2.3	151	87.8	0	0.0	17	9.9	41	20.7	150	75.8	0	0.0	7	3.5
Romania	6	1.9	302	97.7	0	0.0	1	0.3	0	0.0	85	100.0	0	0.0	0	0.0
Russian Fed.	118	5.7	1881	90.0	18	0.9	73	3.5	25	2.6	924	94.5	7	0.7	22	2.3
Serbia	0	0.0	87	75.0	1	0.9	28	24.1	1	1.8	50	90.9	0	0.0	4	7.3
Slovak Rep.	0	0.0	118	95.9	0	0.0	5	4.1	7	4.7	138	92.6	3	2.0	1	0.7
Slovenia	0	0.0	49	94.2	1	1.9	2	3.9	15	30.6	33	67.4	1	2.0	0	0.0
Spain	13	1.2	970	92.2	4	0.4	65	6.2	97	7.6	1113	86.7	11	0.9	63	4.9
Sweden	21	7.6	237	85.6	2	0.7	17	6.1	44	10.4	357	84.0	5	1.2	19	4.5
Switzerland	0	0.0	367	95.1	5	1.3	14	3.6	2	0.4	500	93.6	19	3.6	13	2.4
Türkiye	20	0.9	2200	97.6	4	0.2	30	1.3	4	0.8	457	96.2	1	0.2	13	2.7
Ukraine	6	1.8	303	92.4	1	0.3	18	5.5	0	0.0	98	97.0	1	1.0	2	2.0
United Kingdom	5	0.1	3913	95.4	0	0.0	182	4.4	5	0.1	5737	96.5	0	0.0	204	3.4
Total	366	1.6	21866	93.6	106	0.5	1018	4.4	672	2.5	24714	92.5	425	1.6	916	3.4

Note: Albania and Georgia have <5 adults seen in 2023 and are excluded from the table for adults, but the people are included in the total number.

Note: Ireland and Italy: chronicity for Stenotrophomonas maltophilia is defined as: at least 3 or more positive isolates during the last 12 months preceding the last reported culture in 2023.

The United Kingdom: chronicity for Stenotrophomonas maltophilia is not collected.

This table shows the frequency of *Stenotrophomonas maltophilia* infection in children and adults, by country and overall.

5. Microbiology

Table A5.7 Prevalence of Achromobacter species infection in people with CF seen in 2023 who have never had a transplant, by country and overall.

Country	Children (<18 years)								Adults (≥18 years)							
	Missing/ Unknown		No		Yes, chronic		Yes, not chronic/ intermittent		Missing/ Unknown		No		Yes, chronic		Yes, not chronic/ intermittent	
	N	%	N	%	N	%	N	%	N	%	N	%	N	%	N	%
Albania	1	2.0	49	98.0	0	0.0	0	0.0								
Armenia	13	50.0	13	50.0	0	0.0	0	0.0	2	33.3	4	66.7	0	0.0	0	0.0
Austria	1	0.3	352	98.3	2	0.6	3	0.8	1	0.3	385	95.3	11	2.7	7	1.7
Belarus	0	0.0	130	92.2	11	7.8	0	0.0	0	0.0	9	81.8	2	18.2	0	0.0
Bulgaria	0	0.0	135	100.0	0	0.0	0	0.0	0	0.0	93	98.9	0	0.0	1	1.1
Croatia	0	0.0	76	96.2	0	0.0	3	3.8	0	0.0	58	100.0	0	0.0	0	0.0
Cyprus	0	0.0	6	100.0	0	0.0	0	0.0	3	20.0	10	66.7	1	6.7	1	6.7
Czech Rep.	6	1.7	344	98.3	0	0.0	0	0.0	28	8.9	282	89.5	0	0.0	5	1.6
Denmark	0	0.0	214	100.0	0	0.0	0	0.0	0	0.0	302	93.5	15	4.6	6	1.9
Estonia	1	4.0	22	88.0	1	4.0	1	4.0	0	0.0	16	88.9	0	0.0	2	11.1
Finland	0	0.0	30	100.0	0	0.0	0	0.0	0	0.0	36	100.0	0	0.0	0	0.0
France	0	0.0	2518	97.3	17	0.7	54	2.1	0	0.0	3730	95.5	102	2.6	74	1.9
Georgia	53	100.0	-	-	-	-	-	-								
Germany	13	0.5	2736	98.4	18	0.7	15	0.5	102	2.5	3693	91.8	193	4.8	34	0.9
Greece	0	0.0	230	97.5	0	0.0	6	2.5	23	6.1	334	88.4	7	1.9	14	3.7
Hungary	0	0.0	237	94.4	1	0.4	13	5.2	0	0.0	178	90.8	11	5.6	7	3.6
Iceland	0	0.0	9	100.0	0	0.0	0	0.0	0	0.0	6	85.7	0	0.0	1	14.3
Ireland	0	0.0	518	99.0	0	0.0	5	1.0	1	0.1	733	98.7	2	0.3	7	0.9
Israel	14	8.3	150	88.8	0	0.0	5	3.0	31	8.7	312	87.9	8	2.3	4	1.1
Italy	5	0.2	2104	96.2	18	0.8	61	2.8	7	0.2	3425	94.0	103	2.8	110	3.0
Kazakhstan	0	0.0	9	100.0	0	0.0	0	0.0	0	0.0	18	100.0	0	0.0	0	0.0
Latvia	1	2.8	31	86.1	1	2.8	3	8.3	0	0.0	15	88.2	1	5.9	1	5.9
Lithuania	1	5.0	17	85.0	0	0.0	2	10.0	0	0.0	24	92.3	2	7.7	0	0.0
Luxembourg	1	5.0	19	95.0	0	0.0	0	0.0	0	0.0	5	100.0	0	0.0	0	0.0
Rep. Moldova	31	100.0	-	-	-	-	-	-	15	100.0	-	-	-	-	-	-
Montenegro	0	0.0	29	100.0	0	0.0	0	0.0	0	0.0	11	100.0	0	0.0	0	0.0
Netherlands	12	2.3	513	96.8	0	0.0	5	0.9	181	18.4	778	79.2	11	1.1	12	1.2
N. Macedonia	0	0.0	84	100.0	0	0.0	0	0.0	2	3.8	51	96.2	0	0.0	0	0.0
Norway	2	1.5	133	97.1	1	0.7	1	0.7	29	14.1	174	84.5	1	0.5	2	1.0
Poland	21	2.1	953	96.6	4	0.4	9	0.9	8	1.4	541	93.4	19	3.3	11	1.9
Portugal	4	2.3	164	95.4	0	0.0	4	2.3	45	22.7	148	74.8	2	1.0	3	1.5
Romania	7	2.3	301	97.4	0	0.0	1	0.3	0	0.0	85	100.0	0	0.0	0	0.0
Russian Fed.	123	5.9	1872	89.6	43	2.1	52	2.5	26	2.7	855	87.4	71	7.3	26	2.7
Serbia	1	0.9	99	85.3	5	4.3	11	9.5	1	1.8	48	87.3	3	5.5	3	5.5
Slovak Rep.	0	0.0	119	96.8	1	0.8	3	2.4	8	5.4	137	92.0	2	1.3	2	1.3
Slovenia	0	0.0	51	98.1	0	0.0	1	1.9	15	30.6	31	63.3	1	2.0	2	4.1
Spain	16	1.5	1010	96.0	5	0.5	21	2.0	98	7.6	1109	86.4	50	3.9	27	2.1
Sweden	21	7.6	251	90.6	1	0.4	4	1.4	43	10.1	366	86.1	9	2.1	7	1.7
Switzerland	0	0.0	383	99.2	1	0.3	2	0.5	1	0.2	512	95.9	12	2.3	9	1.7
Türkiye	20	0.9	2211	98.1	6	0.3	17	0.8	4	0.8	452	95.2	6	1.3	13	2.7
Ukraine	7	2.1	307	93.6	1	0.3	13	4.0	0	0.0	98	97.0	1	1.0	2	2.0
United Kingdom	6	0.2	4068	99.2	0	0.0	26	0.6	5	0.1	5811	97.7	0	0.0	130	2.2
Total	381	1.6	22497	96.3	137	0.6	341	1.5	680	2.5	24878	93.1	646	2.4	523	2.0

Note: Albania and Georgia have <5 adults seen in 2023 and are excluded from the table for adults, but the people are included in the total number.

Note: Ireland and Italy: chronicity for Achromobacter species is defined as: at least 3 or more positive isolates during the last 12 months preceding the last reported culture in 2023.

The United Kingdom: chronicity for Achromobacter species is not collected.

This table shows the frequency of *Achromobacter species* infection in children and adults, by country and overall.

7. Complications

Table A7.1 Prevalence of CF-related diabetes (CFRD) in adults with CF seen in 2023 who have never had a transplant, by country and overall.

Country	CFRD this year											
	Missing/ Unknown		No		Yes, treated with daily insulin		Yes, treated with oral hypo-glycaemic agents		Yes, dietary advice only		Yes, therapy unknown	
	N	%	N	%	N	%	N	%	N	%	N	%
Armenia	0	0.0	4	66.7	0	0.0	0	0.0	2	33.3	0	0.0
Austria	2	0.5	298	73.8	83	20.5	0	0.0	21	5.2	0	0.0
Belarus	0	0.0	11	100.0	0	0.0	0	0.0	0	0.0	0	0.0
Bulgaria	2	2.1	83	88.3	9	9.6	0	0.0	0	0.0	0	0.0
Croatia	0	0.0	45	77.6	10	17.2	0	0.0	3	5.2	0	0.0
Cyprus	0	0.0	14	93.3	1	6.7	0	0.0	0	0.0	0	0.0
Czech Republic	4	1.3	208	66.0	80	25.4	0	0.0	18	5.7	5	1.6
Denmark	0	0.0	247	76.5	76	23.5	0	0.0	0	0.0	0	0.0
Estonia	0	0.0	15	83.3	3	16.7	0	0.0	0	0.0	0	0.0
Finland	0	0.0	26	72.2	9	25.0	1	2.8	0	0.0	0	0.0
France	0	0.0	3018	77.3	627	16.1	53	1.4	208	5.3	0	0.0
Germany	14	0.4	2737	68.1	864	21.5	59	1.5	101	2.5	247	6.1
Greece	2	0.5	279	73.8	79	20.9	6	1.6	12	3.2	0	0.0
Hungary	4	2.0	141	71.9	47	24.0	0	0.0	4	2.0	0	0.0
Iceland	0	0.0	3	42.9	4	57.1	0	0.0	0	0.0	0	0.0
Ireland	0	0.0	561	75.5	141	19.0	0	0.0	41	5.5	0	0.0
Israel	4	1.1	232	65.4	96	27.0	8	2.3	14	3.9	1	0.3
Italy	31	0.9	2818	77.3	712	19.5	21	0.6	54	1.5	9	0.3
Kazakhstan	0	0.0	17	94.4	1	5.6	0	0.0	0	0.0	0	0.0
Latvia	1	5.9	13	76.5	0	0.0	0	0.0	3	17.7	0	0.0
Lithuania	0	0.0	24	92.3	1	3.9	0	0.0	1	3.9	0	0.0
Luxembourg	0	0.0	3	60.0	2	40.0	0	0.0	0	0.0	0	0.0
Rep of Moldova	0	0.0	13	86.7	2	13.3	0	0.0	0	0.0	0	0.0
Montenegro	0	0.0	9	81.8	2	18.2	0	0.0	0	0.0	0	0.0
The Netherlands	62	6.3	576	58.7	234	23.8	27	2.8	73	7.4	10	1.0
North Macedonia	2	3.8	33	62.3	18	34.0	0	0.0	0	0.0	0	0.0
Norway	0	0.0	165	80.1	34	16.5	2	1.0	3	1.5	2	1.0
Poland	9	1.6	424	73.2	92	15.9	1	0.2	53	9.2	0	0.0
Portugal	3	1.5	160	80.8	22	11.1	2	1.0	11	5.6	0	0.0
Romania	2	2.4	70	82.4	13	15.3	0	0.0	0	0.0	0	0.0
Russian Fed.	12	1.2	850	86.9	96	9.8	0	0.0	19	1.9	1	0.1
Serbia	0	0.0	34	61.8	20	36.4	0	0.0	1	1.8	0	0.0
Slovak Republic	2	1.3	123	82.6	16	10.7	1	0.7	7	4.7	0	0.0
Slovenia	1	2.0	40	81.6	8	16.3	0	0.0	0	0.0	0	0.0
Spain	10	0.8	976	76.0	205	16.0	28	2.2	64	5.0	1	0.1
Sweden	1	0.2	301	70.8	86	20.2	9	2.1	0	0.0	28	6.6
Switzerland	4	0.8	383	71.7	127	23.8	2	0.4	14	2.6	4	0.8
Türkiye	2	0.4	408	85.9	48	10.1	6	1.3	8	1.7	3	0.6
Ukraine	2	2.0	90	89.1	9	8.9	0	0.0	0	0.0	0	0.0
United Kingdom	71	1.2	3680	61.9	1539	25.9	200	3.4	189	3.2	267	4.5
Total	247	0.9	19135	71.6	5417	20.3	426	1.6	924	3.5	578	2.2

Note: Albania and Georgia have <5 adults seen in 2023 and are excluded from the table for adults, but the people are included in the total number.

7. Complications

Table A7.2 Prevalence of liver disease in people with CF seen in 2023 who have never had a transplant.

Country	Liver disease this year													
	Missing/ Unknown		No		Yes, cirrhosis with portal hypertension/ hypersplenism		Yes, cirrhosis no portal hypertension/ hypersplenism		Yes, cirrhosis, portal hypertension unknown		Yes, liver disease without cirrhosis		Yes, variceal bleeding	
	N	%	N	%	N	%	N	%	N	%	N	%	N	%
Children	219	0.9	18631	79.8	309	1.3	220	0.9	36	0.2	3938	16.9	3	0.0
Adults	198	0.7	18649	69.8	721	2.7	450	1.7	107	0.4	6601	24.7	1	0.0

Note: Serbia: cirrhosis without portal hypertension/hypersplenism is reported when there are abnormal liver function tests and/or ultrasound changes in liver tissue

8. Therapies

Table A8.1 Use of inhaled hypertonic saline >3% (NaCl) for at least 3 consecutive months this year in all people with CF seen in 2023 who have never had a transplant, by country and overall.

Country	Children (<18 years)						Adults (≥18 years)					
	Missing/ Unknown		No		Yes		Missing/ Unknown		No		Yes	
	N	%	N	%	N	%	N	%	N	%	N	%
Albania	0	0.0	0	0.0	50	100						
Armenia	0	0.0	0	0.0	26	100	0	0.0	0	0	6	100
Austria	1	0.3	33	9.2	324	90.5	1	0.3	71	17.6	332	82.2
Belarus	0	0.0	40	28.4	101	71.6	0	0.0	4	36.4	7	63.6
Bulgaria	0	0.0	62	45.9	73	54.1	0	0.0	59	62.8	35	37.2
Croatia	0	0.0	1	1.3	78	98.7	0	0.0	20	34.5	38	65.5
Cyprus	0	0.0	5	83.3	1	16.7	0	0.0	12	80.0	3	20.0
Czech Republic	0	0.0	13	3.7	337	96.3	4	1.3	86	27.3	225	71.4
Denmark	0	0.0	164	76.6	50	23.4	0	0.0	274	84.8	49	15.2
Estonia	0	0.0	4	16.0	21	84.0	0	0.0	12	66.7	6	33.3
Finland	0	0.0	1	3.3	29	96.7	0	0.0	7	19.4	29	80.6
France	0	0.0	2035	78.6	554	21.4	0	0.0	3643	93.3	263	6.7
Georgia	0	0.0	3	5.7	50	94.3						
Germany	8	0.3	158	5.7	2616	94.0	8	0.2	1085	27.0	2929	72.8
Greece	0	0.0	139	58.9	97	41.1	2	0.5	319	84.4	57	15.1
Hungary	0	0.0	19	7.6	232	92.4	0	0.0	25	12.8	171	87.2
Iceland	0	0.0	1	11.1	8	88.9	0	0.0	3	42.9	4	57.1
Ireland	0	0.0	136	26.0	387	74.0	0	0.0	381	51.3	362	48.7
Israel	6	3.6	25	14.8	138	81.7	21	5.9	105	29.6	229	64.5
Italy	4	0.2	1004	45.9	1180	53.9	12	0.3	2109	57.9	1524	41.8
Kazakhstan	0	0.0	0	0.0	9	100	0	0.0	7	38.9	11	61.1
Latvia	0	0.0	4	11.1	32	88.9	0	0.0	2	11.8	15	88.2
Lithuania	0	0.0	4	20.0	16	80.0	0	0.0	26	100	0	0.0
Luxembourg	0	0.0	0	0.0	20	100	0	0.0	1	20.0	4	80.0
Rep of Moldova	1	3.2	2	6.5	28	90.3	0	0.0	3	20.0	12	80.0
Montenegro	0	0.0	0	0.0	29	100	0	0.0	3	27.3	8	72.7
The Netherlands	4	0.8	353	66.6	173	32.6	3	0.3	737	75.1	242	24.6
North Macedonia	0	0.0	5	6.0	79	94.1	1	1.9	1	1.9	51	96.2
Norway	1	0.7	71	51.8	65	47.5	5	2.4	67	32.5	134	65.1
Poland	1	0.1	103	10.4	883	89.5	7	1.2	111	19.2	461	79.6
Portugal	0	0.0	75	43.6	97	56.4	0	0.0	135	68.2	63	31.8
Romania	3	1.0	53	17.2	253	81.9	2	2.4	10	11.8	73	85.9
Russian Fed.	88	4.2	437	20.9	1565	74.9	33	3.4	420	42.9	525	53.7
Serbia	0	0.0	0	0.0	116	100	0	0.0	1	1.8	54	98.2
Slovak Republic	1	0.8	38	30.9	84	68.3	0	0.0	119	79.9	30	20.1
Slovenia	0	0.0	0	0.0	52	100	1	2.0	13	26.5	35	71.4
Spain	4	0.4	282	26.8	766	72.8	2	0.2	628	48.9	654	50.9
Sweden	6	2.2	26	9.4	245	88.5	10	2.4	131	30.8	284	66.8
Switzerland	0	0.0	81	21.0	305	79.0	1	0.2	235	44.0	298	55.8
Türkiye	0	0.0	1676	74.4	578	25.6	1	0.2	321	67.6	153	32.2
Ukraine	1	0.3	12	3.7	315	96.0	1	1.0	3	3.0	97	96.0
United Kingdom	0	0.0	2692	65.7	1408	34.3	0	0.0	4223	71.0	1723	29.0
Total	129	0.6	9757	41.8	13470	57.7	115	0.4	15412	57.7	11200	41.9

Note: Albania and Georgia have <5 adults seen in 2023 and are excluded from the table for adults, but the people are included in the total number.

Note: Inhaled hypertonic saline is reimbursed in most countries except in Albania, Armenia, Bulgaria, Estonia, Georgia, Kazakhstan, Lithuania, the Republic of Moldova, Poland and Romania. In Türkiye it is reimbursed for children ≥ 6 years.

8. Therapies

Table A8.2 Use of inhaled rhDNase for ≥3 months in all people with CF seen in 2023 who have never had a transplant, by country and overall.

Country	Children (<18 years)						Adults (≥18 years)					
	Missing/ Unknown		No		Yes		Missing/ Unknown		No		Yes	
	N	%	N	%	N	%	N	%	N	%	N	%
Albania	0	0.0	47	94.0	3	6.0						
Armenia	0	0.0	21	80.8	5	19.2	0	0.0	5	83.3	1	16.7
Austria	1	0.3	173	48.3	184	51.4	2	0.5	195	48.3	207	51.2
Belarus	0	0.0	102	72.3	39	27.7	0	0.0	10	90.9	1	9.1
Bulgaria	0	0.0	35	25.9	100	74.1	0	0.0	31	33.0	63	67.0
Croatia	1	1.3	16	20.3	62	78.5	0	0.0	6	10.3	52	89.7
Cyprus	0	0.0	2	33.3	4	66.7	0	0.0	1	6.7	14	93.3
Czech Republic	0	0.0	186	53.1	164	46.9	4	1.3	73	23.2	238	75.6
Denmark	0	0.0	171	79.9	43	20.1	0	0.0	183	56.7	140	43.3
Estonia	0	0.0	11	44.0	14	56.0	0	0.0	4	22.2	14	77.8
Finland	0	0.0	8	26.7	22	73.3	0	0.0	4	11.1	32	88.9
France	0	0.0	1738	67.1	851	32.9	0	0.0	3095	79.2	811	20.8
Georgia	0	0.0	20	37.7	33	62.3						
Germany	18	0.7	1768	63.6	996	35.8	27	0.7	2303	57.3	1692	42.1
Greece	0	0.0	53	22.5	183	77.5	2	0.5	145	38.4	231	61.1
Hungary	0	0.0	118	47.0	133	53.0	0	0.0	15	7.7	181	92.4
Iceland	0	0.0	3	33.3	6	66.7	0	0.0	4	57.1	3	42.9
Ireland	0	0.0	303	57.9	220	42.1	1	0.1	301	40.5	441	59.4
Israel	3	1.8	57	33.7	109	64.5	17	4.8	124	34.9	214	60.3
Italy	5	0.2	1199	54.8	984	45.0	11	0.3	2139	58.7	1495	41.0
Kazakhstan	0	0.0	1	11.1	8	88.9	0	0.0	6	33.3	12	66.7
Latvia	0	0.0	27	75.0	9	25.0	1	5.9	3	17.7	13	76.5
Lithuania	0	0.0	6	30.0	14	70.0	0	0.0	2	7.7	24	92.3
Luxembourg	0	0.0	3	15.0	17	85.0	0	0.0	1	20.0	4	80.0
Rep of Moldova	1	3.2	10	32.3	20	64.5	0	0.0	3	20.0	12	80.0
Montenegro	0	0.0	4	13.8	25	86.2	0	0.0	1	9.1	10	90.9
The Netherlands	2	0.4	235	44.3	293	55.3	5	0.5	515	52.4	462	47.1
North Macedonia	0	0.0	28	33.3	56	66.7	2	3.8	1	1.9	50	94.3
Norway	2	1.5	59	43.1	76	55.5	7	3.4	100	48.5	99	48.1
Poland	4	0.4	172	17.4	811	82.2	4	0.7	30	5.2	545	94.1
Portugal	3	1.7	34	19.8	135	78.5	0	0.0	18	9.1	180	90.9
Romania	3	1.0	69	22.3	237	76.7	2	2.4	3	3.5	80	94.1
Russian Fed.	80	3.8	35	1.7	1975	94.5	22	2.3	145	14.8	811	82.9
Serbia	0	0.0	35	30.2	81	69.8	0	0.0	2	3.6	53	96.4
Slovak Republic	0	0.0	43	35.0	80	65.0	1	0.7	32	21.5	116	77.9
Slovenia	0	0.0	48	92.3	4	7.7	0	0.0	36	73.5	13	26.5
Spain	2	0.2	732	69.6	318	30.2	2	0.2	903	70.3	379	29.5
Sweden	7	2.5	193	69.7	77	27.8	13	3.1	320	75.3	92	21.7
Switzerland	0	0.0	304	78.8	82	21.2	0	0.0	364	68.2	170	31.8
Türkiye	0	0.0	271	12.0	1983	88.0	1	0.2	71	15.0	403	84.8
Ukraine	2	0.6	99	30.2	227	69.2	1	1.0	5	5.0	95	94.1
United Kingdom	0	0.0	1558	38.0	2542	62.0	0	0.0	2187	36.8	3759	63.2
Total	134	0.6	9997	42.8	13225	56.6	125	0.5	13389	50.1	13213	49.4

Note: Albania and Georgia have <5 adults seen in 2023 and are excluded from the table for adults, but the people are included in the total number.

Note: Inhaled rhDNase is reimbursed in most countries except in Albania, Armenia and Belarus. It is reimbursed in Georgia for people with CF ≥ 2 years; in Bulgaria, Germany, Luxembourg, Macedonia, the Republic of Moldova, Norway, Romania, Spain, and the United Kingdom for individuals ≥ 5 years; in Latvia and Hungary for individuals ≥ 6 years.

8. Therapies

Table A8.3 Use of inhaled antibiotics for at least 3 months in all people with CF seen in 2023 who have never had a transplant, by country and overall.

Country	Children (<18 years)						Adults (≥18 years)					
	Missing/ Unknown		No		Yes		Missing/ Unknown		No		Yes	
	N	%	N	%	N	%	N	%	N	%	N	%
Albania	1	2.0	44	88.0	5	10.0						
Armenia	0	0.0	20	76.9	6	23.1	0	0.0	5	83.3	1	16.7
Austria	3	0.8	294	82.1	61	17.0	2	0.5	233	57.7	169	41.8
Belarus	0	0.0	101	71.6	40	28.4	0	0.0	7	63.6	4	36.4
Bulgaria	0	0.0	92	68.2	43	31.9	0	0.0	37	39.4	57	60.6
Croatia	1	1.3	47	59.5	31	39.2	0	0.0	29	50.0	29	50.0
Cyprus	0	0.0	4	66.7	2	33.3	0	0.0	11	73.3	4	26.7
Czech Republic	0	0.0	317	90.6	33	9.4	4	1.3	218	69.2	93	29.5
Denmark	0	0.0	193	90.2	21	9.8	0	0.0	204	63.2	119	36.8
Estonia	0	0.0	21	84.0	4	16.0	1	5.6	10	55.6	7	38.9
Finland	0	0.0	29	96.7	1	3.3	0	0.0	23	63.9	13	36.1
France	0	0.0	2150	83.0	439	17.0	0	0.0	2918	74.7	988	25.3
Georgia	1	1.9	50	94.3	2	3.8						
Germany	18	0.7	2376	85.4	388	14.0	30	0.8	2152	53.5	1840	45.8
Greece	0	0.0	137	58.1	99	42.0	1	0.3	166	43.9	211	55.8
Hungary	0	0.0	171	68.1	80	31.9	0	0.0	59	30.1	137	69.9
Iceland	0	0.0	7	77.8	2	22.2	0	0.0	4	57.1	3	42.9
Ireland	0	0.0	461	88.2	62	11.9	1	0.1	303	40.8	439	59.1
Israel	3	1.8	126	74.6	40	23.7	15	4.2	186	52.4	154	43.4
Italy	4	0.2	1790	81.8	394	18.0	13	0.4	2243	61.5	1389	38.1
Kazakhstan	0	0.0	3	33.3	6	66.7	0	0.0	6	33.3	12	66.7
Latvia	0	0.0	30	83.3	6	16.7	0	0.0	7	41.2	10	58.8
Lithuania	0	0.0	19	95.0	1	5.0	0	0.0	22	84.6	4	15.4
Luxembourg	0	0.0	18	90.0	2	10.0	0	0.0	2	40.0	3	60.0
Rep of Moldova	1	3.2	16	51.6	14	45.2	0	0.0	3	20.0	12	80.0
Montenegro	1	3.5	22	75.9	6	20.7	0	0.0	8	72.7	3	27.3
The Netherlands	1	0.2	503	94.9	26	4.9	2	0.2	681	69.4	299	30.5
North Macedonia	0	0.0	46	54.8	38	45.2	1	1.9	11	20.8	41	77.4
Norway	0	0.0	135	98.5	2	1.5	3	1.5	172	83.5	31	15.1
Poland	2	0.2	837	84.8	148	15.0	5	0.9	360	62.2	214	37.0
Portugal	0	0.0	123	71.5	49	28.5	4	2.0	93	47.0	101	51.0
Romania	2	0.7	193	62.5	114	36.9	2	2.4	44	51.8	39	45.9
Russian Fed.	125	6.0	1191	57.0	774	37.0	18	1.8	414	42.3	546	55.8
Serbia	0	0.0	66	56.9	50	43.1	0	0.0	21	38.2	34	61.8
Slovak Republic	0	0.0	74	60.2	49	39.8	1	0.7	74	49.7	74	49.7
Slovenia	0	0.0	46	88.5	6	11.5	0	0.0	42	85.7	7	14.3
Spain	3	0.3	801	76.1	248	23.6	2	0.2	585	45.6	697	54.3
Sweden	10	3.6	231	83.4	36	13.0	13	3.1	359	84.5	53	12.5
Switzerland	0	0.0	366	94.8	20	5.2	0	0.0	369	69.1	165	30.9
Türkiye	0	0.0	1849	82.0	405	18.0	1	0.2	319	67.2	155	32.6
Ukraine	3	0.9	181	55.2	144	43.9	1	1.0	22	21.8	78	77.2
United Kingdom	0	0.0	3146	76.7	954	23.3	0	0.0	2780	46.8	3166	53.3
Total	179	0.8	18326	78.5	4851	20.8	120	0.5	15204	56.9	11403	42.7

Note: Albania and Georgia have <5 adults seen in 2023 and are excluded from the table for adults, but the people are included in the total number.

Note: Inhaled antibiotics are reimbursed in all countries except Armenia and Georgia. In Bulgaria, colistin is reimbursed for all, tobramycin for > 7years, and levofloxacin for > 18 years. In Estonia, tobramycin and colistin are reimbursed and in Romania, only tobramycin solution and colistin dry powder are reimbursed for ≥ 6 years.

8. Therapies

Table A8.4 Use of inhaled bronchodilators for at least 3 months in children with CF seen in 2023 who have never had a transplant, by country and overall.

Country	Children (<18 years)					
	Missing/ Unknown		No		Yes	
	N	%	N	%	N	%
Albania	0	0.0	4	8.0	46	92.0
Armenia	0	0.0	7	26.9	19	73.1
Austria	0	0.0	23	6.4	335	93.6
Belarus	0	0.0	97	68.8	44	31.2
Bulgaria	0	0.0	127	94.1	8	5.9
Croatia	1	1.3	67	84.8	11	13.9
Cyprus	0	0.0	4	66.7	2	33.3
Czech Republic	0	0.0	254	72.6	96	27.4
Denmark	0	0.0	126	58.9	88	41.1
Estonia	0	0.0	15	60.0	10	40.0
Finland	0	0.0	18	60.0	12	40.0
France	0	0.0	1402	54.2	1187	45.9
Georgia	0	0.0	28	52.8	25	47.2
Germany	10	0.4	793	28.5	1979	71.1
Greece	1	0.4	182	77.1	53	22.5
Hungary	1	0.4	165	65.7	85	33.9
Iceland	0	0.0	1	11.1	8	88.9
Ireland	0	0.0	225	43.0	298	57.0
Israel	1	0.6	76	45.0	92	54.4
Italy	4	0.2	652	29.8	1532	70.0
Kazakhstan	0	0.0	3	33.3	6	66.7
Latvia	1	2.8	4	11.1	31	86.1
Lithuania	0	0.0	9	45.0	11	55.0
Luxembourg	0	0.0	4	20.0	16	80.0
Rep of Moldova	1	3.2	25	80.7	5	16.1
Montenegro	0	0.0	1	3.5	28	96.6
The Netherlands	3	0.6	429	80.9	98	18.5
North Macedonia	0	0.0	6	7.1	78	92.9
Norway	0	0.0	88	64.2	49	35.8
Poland	1	0.1	200	20.3	786	79.6
Portugal	0	0.0	98	57.0	74	43.0
Romania	6	1.9	204	66.0	99	32.0
Russian Fed.	109	5.2	1366	65.4	615	29.4
Serbia	1	0.9	0	0.0	115	99.1
Slovak Republic	0	0.0	60	48.8	63	51.2
Slovenia	1	1.9	49	94.2	2	3.9
Spain	1	0.1	342	32.5	709	67.4
Sweden	8	2.9	27	9.8	242	87.4
Switzerland	0	0.0	159	41.2	227	58.8
Türkiye	0	0.0	1433	63.6	821	36.4
Ukraine	1	0.3	187	57.0	140	42.7
United Kingdom	118	2.9	2304	56.2	1678	40.9
Total	269	1.2	11264	48.2	11823	50.6

Note: Inhaled bronchodilators are reimbursed in most countries except in Bulgaria, Georgia, the Republic of Moldova (reimbursed for people diagnosed with asthma), Poland and Serbia. In Kazakhstan, it is subject to the availability of the regional budget. In Estonia long- and short-acting bronchodilators are reimbursed for pwCF diagnosed with asthma.

8. Therapies

Table A8.5 Use of inhaled bronchodilators for at least3 months in adults with CF seen in 2023 who have never had a transplant, by country and overall.

Country	Adults (≥18 years)					
	Missing/ Unknown		No		Yes	
	N	%	N	%	N	%
Armenia	0	0.0	2	33.3	4	66.7
Austria	1	0.3	25	6.2	378	93.6
Belarus	0	0.0	5	45.5	6	54.5
Bulgaria	0	0.0	66	70.2	28	29.8
Croatia	0	0.0	16	27.6	42	72.4
Cyprus	0	0.0	7	46.7	8	53.3
Czech Republic	4	1.3	125	39.7	186	59.0
Denmark	0	0.0	143	44.3	180	55.7
Estonia	0	0.0	7	38.9	11	61.1
Finland	0	0.0	17	47.2	19	52.8
France	0	0.0	1574	40.3	2332	59.7
Germany	6	0.2	773	19.2	3243	80.6
Greece	1	0.3	182	48.2	195	51.6
Hungary	0	0.0	36	18.4	160	81.6
Iceland	0	0.0	4	57.1	3	42.9
Ireland	1	0.1	126	17.0	616	82.9
Israel	12	3.4	167	47.0	176	49.6
Italy	13	0.4	818	22.4	2814	77.2
Kazakhstan	0	0.0	5	27.8	13	72.2
Latvia	0	0.0	0	0.0	17	100.0
Lithuania	0	0.0	10	38.5	16	61.5
Luxembourg	0	0.0	1	20.0	4	80.0
Rep of Moldova	0	0.0	10	66.7	5	33.3
Montenegro	0	0.0	0	0.0	11	100.0
The Netherlands	5	0.5	377	38.4	600	61.1
North Macedonia	1	1.9	1	1.9	51	96.2
Norway	5	2.4	45	21.8	156	75.7
Poland	4	0.7	65	11.2	510	88.1
Portugal	0	0.0	73	36.9	125	63.1
Romania	2	2.4	43	50.6	40	47.1
Russian Fed.	24	2.5	273	27.9	681	69.6
Serbia	0	0.0	0	0.0	55	100.0
Slovak Republic	1	0.7	53	35.6	95	63.8
Slovenia	0	0.0	39	79.6	10	20.4
Spain	2	0.2	363	28.3	919	71.6
Sweden	14	3.3	53	12.5	358	84.2
Switzerland	0	0.0	117	21.9	417	78.1
Türkiye	1	0.2	188	39.6	286	60.2
Ukraine	1	1.0	21	20.8	79	78.2
United Kingdom	51	0.9	1704	28.7	4191	70.5
Total	149	0.6	7534	28.2	19044	71.2

Note: Albania and Georgia have <5 adults seen in 2023 and are excluded from the table for adults, but the people are included in the total number.

Note: Inhaled bronchodilators are reimbursed in most countries except in Bulgaria, Georgia, the Republic of Moldova (reimbursed for people diagnosed with asthma), Poland and Serbia. In Kazakhstan it is subject to the availability of the regional budget. In Estonia long- and short-acting bronchodilators are reimbursed for people with CF diagnosed with asthma.

8. Therapies

Table A8.6 Use of macrolides for at least3 months in all people with CF seen in 2023 who have never had a transplant, by country and overall.

Country	Children (<18 years)						Adults (≥18 years)					
	Missing/ Unknown		No		Yes		Missing/ Unknown		No		Yes	
	N	%	N	%	N	%	N	%	N	%	N	%
Albania	1	2.0	48	96.0	1	2.0						
Armenia	1	3.9	23	88.5	2	7.7	0	0.0	6	100	0	0.0
Austria	1	0.3	354	98.9	3	0.8	3	0.7	389	96.3	12	3.0
Belarus	0	0.0	109	77.3	32	22.7	0	0.0	8	72.7	3	27.3
Bulgaria	0	0.0	132	97.8	3	2.2	0	0.0	93	98.9	1	1.1
Croatia	1	1.3	39	49.4	39	49.4	0	0.0	36	62.1	22	37.9
Cyprus	0	0.0	2	33.3	4	66.7	0	0.0	6	40.0	9	60.0
Czech Republic	0	0.0	346	98.9	4	1.1	4	1.3	298	94.6	13	4.1
Denmark	0	0.0	198	92.5	16	7.5	0	0.0	243	75.2	80	24.8
Estonia	0	0.0	16	64.0	9	36.0	0	0.0	7	38.9	11	61.1
Finland	0	0.0	28	93.3	2	6.7	0	0.0	31	86.1	5	13.9
France	0	0.0	2166	83.7	423	16.3	0	0.0	2923	74.8	983	25.2
Georgia	2	3.8	49	92.5	2	3.8						
Germany	38	1.4	2641	94.9	103	3.7	66	1.6	3440	85.5	516	12.8
Greece	0	0.0	192	81.4	44	18.6	1	0.3	310	82.0	67	17.7
Hungary	1	0.4	210	83.7	40	15.9	0	0.0	175	89.3	21	10.7
Iceland	0	0.0	7	77.8	2	22.2	0	0.0	5	71.4	2	28.6
Ireland	0	0.0	424	81.1	99	18.9	1	0.1	329	44.3	413	55.6
Israel	4	2.4	124	73.4	41	24.3	14	3.9	213	60.0	128	36.1
Italy	4	0.2	1892	86.5	292	13.4	11	0.3	2754	75.6	880	24.1
Kazakhstan	0	0.0	5	55.6	4	44.4	0	0.0	8	44.4	10	55.6
Latvia	0	0.0	35	97.2	1	2.8	0	0.0	16	94.1	1	5.9
Lithuania	1	5.0	19	95.0	0	0.0	0	0.0	26	100	0	0.0
Luxembourg	0	0.0	13	65.0	7	35.0	0	0.0	2	40.0	3	60.0
Rep of Moldova	1	3.2	26	83.9	4	12.9	0	0.0	13	86.7	2	13.3
Montenegro	0	0.0	27	93.1	2	6.9	0	0.0	11	100	0	0.0
The Netherlands	1	0.2	502	94.7	27	5.1	3	0.3	643	65.5	336	34.2
North Macedonia	0	0.0	79	94.1	5	6.0	2	3.8	28	52.8	23	43.4
Norway	0	0.0	134	97.8	3	2.2	1	0.5	179	86.9	26	12.6
Poland	2	0.2	890	90.2	95	9.6	6	1.0	463	80.0	110	19.0
Portugal	2	1.2	142	82.6	28	16.3	0	0.0	142	71.7	56	28.3
Romania	5	1.6	248	80.3	56	18.1	3	3.5	69	81.2	13	15.3
Russian Fed.	106	5.1	1673	80.1	311	14.9	25	2.6	644	65.9	309	31.6
Serbia	0	0.0	109	94.0	7	6.0	0	0.0	43	78.2	12	21.8
Slovak Republic	0	0.0	90	73.2	33	26.8	1	0.7	79	53.0	69	46.3
Slovenia	0	0.0	52	100	0	0.0	0	0.0	39	79.6	10	20.4
Spain	5	0.5	874	83.1	173	16.4	5	0.4	803	62.5	476	37.1
Sweden	7	2.5	234	84.5	36	13.0	12	2.8	318	74.8	95	22.4
Switzerland	0	0.0	380	98.5	6	1.6	0	0.0	433	81.1	101	18.9
Türkiye	0	0.0	2060	91.4	194	8.6	1	0.2	410	86.3	64	13.5
Ukraine	2	0.6	121	36.9	205	62.5	1	1.0	26	25.7	74	73.3
United Kingdom	0	0.0	3923	95.7	177	4.3	0	0.0	3631	61.1	2315	38.9
Total	185	0.8	20636	88.4	2535	10.9	160	0.6	19296	72.2	7271	27.2

Note: Albania and Georgia have <5 adults seen in 2023 and are excluded from the table for adults, but the people are included in the total number.

Note: Oral macrolides are reimbursed in most countries except in Bulgaria, Georgia, Kazakhstan and Serbia. In Armenia, they are reimbursed for some outpatients.

8. Therapies

Table A8.7 Use of oxygen for at least 3 months in all people with CF seen in 2023 who have never had a transplant, by country and overall.

Country	Children (<18 years)						Adults (≥18 years)					
	Missing/ Unknown		No		Yes		Missing/ Unknown		No		Yes	
	N	%	N	%	N	%	N	%	N	%	N	%
Albania	1	2.0	49	98.0	0	0.0						
Armenia	0	0.0	24	92.3	2	7.7	0	0.0	6	100	0	0.0
Austria	0	0.0	355	99.2	3	0.8	1	0.3	395	97.8	8	2.0
Belarus	0	0.0	139	98.6	2	1.4	0	0.0	10	90.9	1	9.1
Bulgaria	0	0.0	134	99.3	1	0.7	0	0.0	92	97.9	2	2.1
Croatia	1	1.3	77	97.5	1	1.3	0	0.0	56	96.6	2	3.5
Cyprus	0	0.0	6	100	0	0.0	0	0.0	14	93.3	1	6.7
Czech Republic	0	0.0	349	99.7	1	0.3	4	1.3	308	97.8	3	1.0
Denmark	0	0.0	211	98.6	3	1.4	0	0.0	321	99.4	2	0.6
Estonia	0	0.0	25	100	0	0.0	0	0.0	15	83.3	3	16.7
Finland	0	0.0	30	100	0	0.0	0	0.0	35	97.2	1	2.8
France	0	0.0	2583	99.8	6	0.2	0	0.0	3806	97.4	100	2.6
Georgia	0	0.0	53	100	0	0.0						
Germany	15	0.5	2757	99.1	10	0.4	14	0.4	3747	93.2	261	6.5
Greece	0	0.0	236	100	0	0.0	1	0.3	373	98.7	4	1.1
Hungary	1	0.4	248	98.8	2	0.8	0	0.0	167	85.2	29	14.8
Iceland	0	0.0	9	100	0	0.0	0	0.0	7	100	0	0.0
Ireland	0	0.0	522	99.8	1	0.2	1	0.1	696	93.7	46	6.2
Israel	2	1.2	167	98.8	0	0.0	4	1.1	345	97.2	6	1.7
Italy	3	0.1	2178	99.5	7	0.3	10	0.3	3464	95.0	171	4.7
Kazakhstan	0	0.0	9	100	0	0.0	0	0.0	12	66.7	6	33.3
Latvia	0	0.0	36	100	0	0.0	0	0.0	16	94.1	1	5.9
Lithuania	0	0.0	19	95.0	1	5.0	0	0.0	24	92.3	2	7.7
Luxembourg	0	0.0	20	100	0	0.0	0	0.0	4	80.0	1	20.0
Rep of Moldova	1	3.2	29	93.6	1	3.2	0	0.0	13	86.7	2	13.3
Montenegro	0	0.0	29	100	0	0.0	0	0.0	11	100	0	0.0
The Netherlands	3	0.6	525	99.1	2	0.4	2	0.2	958	97.6	22	2.2
North Macedonia	0	0.0	84	100	0	0.0	1	1.9	50	94.3	2	3.8
Norway	1	0.7	135	98.5	1	0.7	0	0.0	204	99.0	2	1.0
Poland	4	0.4	979	99.2	4	0.4	2	0.4	561	96.9	16	2.8
Portugal	3	1.7	168	97.7	1	0.6	0	0.0	189	95.5	9	4.6
Romania	4	1.3	303	98.1	2	0.7	2	2.4	80	94.1	3	3.5
Russian Fed.	78	3.7	1972	94.4	40	1.9	18	1.8	869	88.9	91	9.3
Serbia	0	0.0	114	98.3	2	1.7	0	0.0	50	90.9	5	9.1
Slovak Republic	0	0.0	123	100	0	0.0	1	0.7	141	94.6	7	4.7
Slovenia	0	0.0	52	100	0	0.0	0	0.0	49	100	0	0.0
Spain	6	0.6	1044	99.2	2	0.2	3	0.2	1253	97.6	28	2.2
Sweden	7	2.5	265	95.7	5	1.8	11	2.6	410	96.5	4	0.9
Switzerland	0	0.0	385	99.7	1	0.3	0	0.0	521	97.6	13	2.4
Türkiye	0	0.0	2204	97.8	50	2.2	1	0.2	446	93.9	28	5.9
Ukraine	2	0.6	317	96.7	9	2.7	3	3.0	88	87.1	10	9.9
United Kingdom	0	0.0	4052	98.8	48	1.2	0	0.0	5696	95.8	250	4.2
Total	132	0.6	23016	98.5	208	0.9	79	0.3	25506	95.4	1142	4.3

Note: Albania and Georgia have <5 adults seen in 2023 and are excluded from the table for adults, but the people are included in the total number.

Note: Oxygen therapy is reimbursed in most countries except in Bulgaria and the Republic of Moldova. In Armenia and Georgia it is only reimbursed if the individual is hospitalised; in Serbia oxygen therapy at home is reimbursed.

8. Therapies

Table A8.8 Use of inhaled steroids >3 months in all people with CF seen in 2023 who have never had a transplant, by country and overall.

Country	Children (<18 years)						Adults (≥18 years)					
	Missing/ Unknown		No		Yes		Missing/ Unknown		No		Yes	
	N	%	N	%	N	%	N	%	N	%	N	%
Albania	0	0.0	42	84.0	8	16.0						
Armenia	1	3.9	18	69.2	7	26.9	0	0.0	5	83.3	1	16.7
Austria	1	0.3	340	95.0	17	4.8	2	0.5	333	82.4	69	17.1
Belarus	0	0.0	108	76.6	33	23.4	0	0.0	5	45.5	6	54.6
Bulgaria	0	0.0	132	97.8	3	2.2	0	0.0	72	76.6	22	23.4
Croatia	1	1.3	75	94.9	3	3.8	0	0.0	33	56.9	25	43.1
Cyprus	0	0.0	5	83.3	1	16.7	0	0.0	9	60.0	6	40.0
Czech Republic	0	0.0	281	80.3	69	19.7	4	1.3	172	54.6	139	44.1
Denmark	0	0.0	186	86.9	28	13.1	0	0.0	273	84.5	50	15.5
Estonia	0	0.0	21	84.0	4	16.0	0	0.0	11	61.1	7	38.9
Finland	0	0.0	25	83.3	5	16.7	0	0.0	24	66.7	12	33.3
France	0	0.0	1455	56.2	1134	43.8	0	0.0	1807	46.3	2099	53.7
Georgia	0	0.0	53	100	0	0.0						
Germany	24	0.9	2349	84.4	409	14.7	26	0.7	2415	60.0	1581	39.3
Greece	0	0.0	204	86.4	32	13.6	2	0.5	265	70.1	111	29.4
Hungary	1	0.4	215	85.7	35	13.9	1	0.5	171	87.2	24	12.2
Iceland	0	0.0	7	77.8	2	22.2	0	0.0	6	85.7	1	14.3
Ireland	0	0.0	374	71.5	149	28.5	1	0.1	202	27.2	540	72.7
Israel	2	1.2	124	73.4	43	25.4	8	2.3	212	59.7	135	38.0
Italy	3	0.1	1835	83.9	350	16.0	11	0.3	2286	62.7	1348	37.0
Kazakhstan	0	0.0	7	77.8	2	22.2	0	0.0	13	72.2	5	27.8
Latvia	0	0.0	29	80.6	7	19.4	0	0.0	11	64.7	6	35.3
Lithuania	0	0.0	19	95.0	1	5.0	0	0.0	20	76.9	6	23.1
Luxembourg	0	0.0	11	55.0	9	45.0	0	0.0	1	20.0	4	80.0
Rep of Moldova	1	3.2	26	83.9	4	12.9	0	0.0	11	73.3	4	26.7
Montenegro	0	0.0	24	82.8	5	17.2	0	0.0	6	54.6	5	45.5
The Netherlands	0	0.0	460	86.8	70	13.2	4	0.4	545	55.5	433	44.1
North Macedonia	0	0.0	80	95.2	4	4.8	1	1.9	38	71.7	14	26.4
Norway	0	0.0	129	94.2	8	5.8	2	1.0	161	78.2	43	20.9
Poland	2	0.2	865	87.6	120	12.2	2	0.4	377	65.1	200	34.5
Portugal	2	1.2	128	74.4	42	24.4	0	0.0	143	72.2	55	27.8
Romania	1	0.3	288	93.2	20	6.5	3	3.5	57	67.1	25	29.4
Russian Fed.	96	4.6	1861	89.0	133	6.4	15	1.5	791	80.9	172	17.6
Serbia	0	0.0	98	84.5	18	15.5	0	0.0	24	43.6	31	56.4
Slovak Republic	0	0.0	71	57.7	52	42.3	1	0.7	63	42.3	85	57.1
Slovenia	0	0.0	48	92.3	4	7.7	0	0.0	48	98.0	1	2.0
Spain	3	0.3	788	74.9	261	24.8	3	0.2	692	53.9	589	45.9
Sweden	7	2.5	248	89.5	22	7.9	14	3.3	245	57.7	166	39.1
Switzerland	0	0.0	297	76.9	89	23.1	0	0.0	264	49.4	270	50.6
Türkiye	0	0.0	1931	85.7	323	14.3	1	0.2	370	77.9	104	21.9
Ukraine	5	1.5	293	89.3	30	9.2	5	5.0	76	75.3	20	19.8
United Kingdom	0	0.0	3580	87.3	520	12.7	0	0.0	4914	82.6	1032	17.4
Total	150	0.6	19130	81.9	4076	17.5	106	0.4	17174	64.3	9447	35.4

Note: Albania and Georgia have <5 adults seen in 2023 and are excluded from the table for adults, but the people are included in the total number.

Note: Inhaled steroids are reimbursed in most countries except Armenia, Georgia, Kazakhstan, Lithuania, the Republic of Moldova (reimbursed for people diagnosed with asthma), Poland and Serbia. In Bulgaria they are reimbursed for people with CF who are also diagnosed with asthma or chronic obstructive pulmonary disease (COPD). In Estonia and Romania inhaled steroids are reimbursed for people with CF diagnosed with asthma.

8. Therapies

Table A8.9 Use of oral steroids for at least3 months in all people with CF seen in 2023 who have never had a transplant, by country and overall.

Country	Children (<18 years)						Adults (≥18 years)					
	Missing/ Unknown		No		Yes		Missing/ Unknown		No		Yes	
	N	%	N	%	N	%	N	%	N	%	N	%
Albania	1	2.0	49	98.0	0	0.0						
Armenia	0	0.0	26	100	0	0.0	0	0.0	6	100	0	0.0
Austria	0	0.0	355	99.2	3	0.8	2	0.5	396	98.0	6	1.5
Belarus	0	0.0	137	97.2	4	2.8	0	0.0	10	90.9	1	9.1
Bulgaria	0	0.0	135	100	0	0.0	0	0.0	93	98.9	1	1.1
Croatia	1	1.3	78	98.7	0	0.0	0	0.0	57	98.3	1	1.7
Cyprus	0	0.0	6	100	0	0.0	0	0.0	15	100	0	0.0
Czech Republic	0	0.0	350	100	0	0.0	4	1.3	304	96.5	7	2.2
Denmark	0	0.0	213	99.5	1	0.5	0	0.0	304	94.1	19	5.9
Estonia	0	0.0	25	100	0	0.0	0	0.0	18	100	0	0.0
Finland	0	0.0	30	100	0	0.0	0	0.0	36	100	0	0.0
France	0	0.0	2572	99.3	17	0.7	0	0.0	3814	97.6	92	2.4
Georgia	0	0.0	53	100	0	0.0						
Germany	32	1.2	2723	97.9	27	1.0	53	1.3	3771	93.8	198	4.9
Greece	0	0.0	232	98.3	4	1.7	1	0.3	375	99.2	2	0.5
Hungary	1	0.4	248	98.8	2	0.8	1	0.5	192	98.0	3	1.5
Iceland	0	0.0	9	100	0	0.0	0	0.0	7	100	0	0.0
Ireland	0	0.0	513	98.1	10	1.9	1	0.1	713	96.0	29	3.9
Israel	3	1.8	166	98.2	0	0.0	8	2.3	332	93.5	15	4.2
Italy	3	0.1	1983	90.6	202	9.2	14	0.4	2768	75.9	863	23.7
Kazakhstan	0	0.0	9	100	0	0.0	0	0.0	18	100	0	0.0
Latvia	0	0.0	36	100	0	0.0	0	0.0	17	100	0	0.0
Lithuania	0	0.0	20	100	0	0.0	0	0.0	26	100	0	0.0
Luxembourg	0	0.0	20	100	0	0.0	0	0.0	5	100	0	0.0
Rep of Moldova	1	3.2	30	96.8	0	0.0	0	0.0	15	100	0	0.0
Montenegro	0	0.0	28	96.6	1	3.5	0	0.0	11	100	0	0.0
The Netherlands	0	0.0	521	98.3	9	1.7	3	0.3	950	96.7	29	3.0
North Macedonia	0	0.0	84	100	0	0.0	1	1.9	51	96.2	1	1.9
Norway	0	0.0	137	100	0	0.0	1	0.5	203	98.5	2	1.0
Poland	4	0.4	978	99.1	5	0.5	4	0.7	564	97.4	11	1.9
Portugal	1	0.6	167	97.1	4	2.3	1	0.5	197	99.5	0	0.0
Romania	2	0.7	306	99.0	1	0.3	1	1.2	84	98.8	0	0.0
Russian Fed.	81	3.9	1973	94.4	36	1.7	14	1.4	936	95.7	28	2.9
Serbia	0	0.0	116	100	0	0.0	0	0.0	55	100	0	0.0
Slovak Republic	0	0.0	120	97.6	3	2.4	1	0.7	137	92.0	11	7.4
Slovenia	0	0.0	52	100	0	0.0	1	2.0	47	95.9	1	2.0
Spain	5	0.5	1042	99.1	5	0.5	5	0.4	1243	96.8	36	2.8
Sweden	7	2.5	270	97.5	0	0.0	13	3.1	398	93.7	14	3.3
Switzerland	0	0.0	385	99.7	1	0.3	0	0.0	524	98.1	10	1.9
Türkiye	0	0.0	2236	99.2	18	0.8	1	0.2	466	98.1	8	1.7
Ukraine	4	1.2	321	97.9	3	0.9	4	4.0	97	96.0	0	0.0
United Kingdom	0	0.0	4057	99.0	43	1.1	0	0.0	5522	92.9	424	7.1
Total	146	0.6	22811	97.7	399	1.7	134	0.5	24781	92.7	1812	6.8

Note: Albania and Georgia have <5 adults seen in 2023 and are excluded from the table for adults, but the people are included in the total number.

Note: Oral steroids are reimbursed in most countries except in Armenia, Bulgaria, Georgia, Kazakhstan, Lithuania, the Republic of Moldova and Serbia.

8. Therapies

Table A8.10 Use of ursodeoxycholic acid for ≥ 3 months in all people with CF seen in 2023 who have never had a transplant, by country and overall.

Country	Children (<18 years)						Adults (≥18 years)					
	Missing/ Unknown		No		Yes		Missing/ Unknown		No		Yes	
	N	%	N	%	N	%	N	%	N	%	N	%
Albania	1	2.0	23	46.0	26	52.0						
Armenia	0	0.0	20	76.9	6	23.1	0	0.0	5	83.3	1	16.7
Austria	1	0.3	186	52.0	171	47.8	1	0.3	209	51.7	194	48.0
Belarus	0	0.0	21	14.9	120	85.1	0	0.0	1	9.1	10	90.9
Bulgaria	0	0.0	47	34.8	88	65.2	0	0.0	54	57.5	40	42.6
Croatia	1	1.3	59	74.7	19	24.1	0	0.0	37	63.8	21	36.2
Cyprus	0	0.0	6	100	0	0.0	0	0.0	13	86.7	2	13.3
Czech Republic	1	0.3	239	68.3	110	31.4	4	1.3	215	68.3	96	30.5
Denmark	0	0.0	169	79.0	45	21.0	0	0.0	216	66.9	107	33.1
Estonia	0	0.0	19	76.0	6	24.0	0	0.0	13	72.2	5	27.8
Finland	0	0.0	23	76.7	7	23.3	0	0.0	26	72.2	10	27.8
France	0	0.0	2249	86.9	340	13.1	0	0.0	3078	78.8	828	21.2
Georgia	2	3.8	45	84.9	6	11.3						
Germany	10	0.4	1706	61.3	1066	38.3	9	0.2	1877	46.7	2136	53.1
Greece	0	0.0	165	69.9	71	30.1	1	0.3	258	68.3	119	31.5
Hungary	0	0.0	165	65.7	86	34.3	0	0.0	93	47.5	103	52.6
Iceland	0	0.0	8	88.9	1	11.1	0	0.0	6	85.7	1	14.3
Ireland	0	0.0	506	96.8	17	3.3	1	0.1	663	89.2	79	10.6
Israel	0	0.0	152	89.9	17	10.1	4	1.1	300	84.5	51	14.4
Italy	4	0.2	1653	75.6	531	24.3	11	0.3	2194	60.2	1440	39.5
Kazakhstan	0	0.0	0	0.0	9	100	0	0.0	7	38.9	11	61.1
Latvia	1	2.8	27	75.0	8	22.2	0	0.0	11	64.7	6	35.3
Lithuania	0	0.0	17	85.0	3	15.0	0	0.0	24	92.3	2	7.7
Luxembourg	0	0.0	19	95.0	1	5.0	0	0.0	3	60.0	2	40.0
Rep of Moldova	1	3.2	19	61.3	11	35.5	0	0.0	8	53.3	7	46.7
Montenegro	0	0.0	16	55.2	13	44.8	0	0.0	3	27.3	8	72.7
The Netherlands	1	0.2	446	84.2	83	15.7	3	0.3	750	76.4	229	23.3
North Macedonia	0	0.0	51	60.7	33	39.3	2	3.8	17	32.1	34	64.2
Norway	0	0.0	128	93.4	9	6.6	0	0.0	194	94.2	12	5.8
Poland	3	0.3	489	49.5	495	50.2	2	0.4	199	34.4	378	65.3
Portugal	1	0.6	116	67.4	55	32.0	3	1.5	147	74.2	48	24.2
Romania	5	1.6	196	63.4	108	35.0	3	3.5	52	61.2	30	35.3
Russian Fed.	79	3.8	156	7.5	1855	88.8	18	1.8	270	27.6	690	70.6
Serbia	0	0.0	90	77.6	26	22.4	0	0.0	30	54.6	25	45.5
Slovak Republic	2	1.6	84	68.3	37	30.1	0	0.0	61	40.9	88	59.1
Slovenia	0	0.0	22	42.3	30	57.7	1	2.0	26	53.1	22	44.9
Spain	9	0.9	797	75.8	246	23.4	10	0.8	947	73.8	327	25.5
Sweden	8	2.9	227	82.0	42	15.2	12	2.8	348	81.9	65	15.3
Switzerland	0	0.0	317	82.1	69	17.9	0	0.0	391	73.2	143	26.8
Türkiye	0	0.0	1913	84.9	341	15.1	1	0.2	395	83.2	79	16.6
Ukraine	5	1.5	39	11.9	284	86.6	2	2.0	5	5.0	94	93.1
United Kingdom	0	0.0	3293	80.3	807	19.7	0	0.0	4535	76.3	1411	23.7
Total	135	0.6	15923	68.2	7298	31.3	88	0.3	17683	66.2	8956	33.5

Note: Albania and Georgia have <5 adults seen in 2023 and are excluded from the table for adults, but the people are included in the total number.

Note: Oral ursodeoxycholic acid is reimbursed in most countries in Europe, except in Armenia, Bulgaria, Georgia, Lithuania and Serbia. In the Republic of Moldova, it is reimbursed at 100% for children and at 70% for adults.

8. Therapies

Table A8.11 Prevalence of the use of proton pump inhibitors (PPI) for ≥ 3 months in all people with CF seen in 2023 who have never had a transplant, by country and overall.

Country	Children (<18 years)						Adults (≥18 years)					
	Missing/ Unknown		No		Yes		Missing/ Unknown		No		Yes	
	N	%	N	%	N	%	N	%	N	%	N	%
Albania	1	2.0	35	70.0	14	28.0						
Armenia	0	0.0	17	65.4	9	34.6	0	0.0	4	66.7	2	33.3
Austria	3	0.8	345	96.4	10	2.8	3	0.7	352	87.1	49	12.1
Belarus	0	0.0	109	77.3	32	22.7	0	0.0	6	54.6	5	45.5
Bulgaria	0	0.0	113	83.7	22	16.3	0	0.0	67	71.3	27	28.7
Croatia	1	1.3	67	84.8	11	13.9	0	0.0	51	87.9	7	12.1
Cyprus	0	0.0	6	100	0	0.0	0	0.0	8	53.3	7	46.7
Czech Republic	0	0.0	332	94.9	18	5.1	4	1.3	203	64.4	108	34.3
Denmark	0	0.0	178	83.2	36	16.8	0	0.0	200	61.9	123	38.1
Estonia	0	0.0	24	96.0	1	4.0	0	0.0	13	72.2	5	27.8
Finland	0	0.0	20	66.7	10	33.3	0	0.0	33	91.7	3	8.3
France	0	0.0	2067	79.8	522	20.2	0	0.0	2325	59.5	1581	40.5
Georgia	2	3.8	51	96.2	0	0.0						
Germany	10	0.4	2514	90.4	258	9.3	13	0.3	3067	76.3	942	23.4
Greece	0	0.0	211	89.4	25	10.6	2	0.5	323	85.5	53	14.0
Hungary	0	0.0	251	100	0	0.0	0	0.0	196	100	0	0.0
Iceland	0	0.0	5	55.6	4	44.4	0	0.0	6	85.7	1	14.3
Ireland	0	0.0	405	77.4	118	22.6	1	0.1	313	42.1	429	57.7
Israel	2	1.2	122	72.2	45	26.6	8	2.3	234	65.9	113	31.8
Italy	5	0.2	1894	86.6	289	13.2	10	0.3	2380	65.3	1255	34.4
Kazakhstan	0	0.0	8	88.9	1	11.1	0	0.0	18	100	0	0.0
Latvia	0	0.0	32	88.9	4	11.1	1	5.9	16	94.1	0	0.0
Lithuania	1	5.0	14	70.0	5	25.0	0	0.0	20	76.9	6	23.1
Luxembourg	0	0.0	17	85.0	3	15.0	0	0.0	2	40.0	3	60.0
Rep of Moldova	1	3.2	21	67.7	9	29.0	0	0.0	13	86.7	2	13.3
Montenegro	0	0.0	22	75.9	7	24.1	0	0.0	4	36.4	7	63.6
The Netherlands	0	0.0	422	79.6	108	20.4	5	0.5	498	50.7	479	48.8
North Macedonia	0	0.0	49	58.3	35	41.7	1	1.9	9	17.0	43	81.1
Norway	0	0.0	123	89.8	14	10.2	3	1.5	165	80.1	38	18.5
Poland	7	0.7	917	92.9	63	6.4	7	1.2	444	76.7	128	22.1
Portugal	2	1.2	158	91.9	12	7.0	1	0.5	138	69.7	59	29.8
Romania	4	1.3	297	96.1	8	2.6	2	2.4	77	90.6	6	7.1
Russian Fed.	112	5.4	1620	77.5	358	17.1	19	1.9	628	64.2	331	33.8
Serbia	0	0.0	90	77.6	26	22.4	0	0.0	40	72.7	15	27.3
Slovak Republic	0	0.0	120	97.6	3	2.4	1	0.7	119	79.9	29	19.5
Slovenia	0	0.0	48	92.3	4	7.7	0	0.0	35	71.4	14	28.6
Spain	5	0.5	909	86.4	138	13.1	10	0.8	721	56.2	553	43.1
Sweden	8	2.9	236	85.2	33	11.9	17	4.0	316	74.4	92	21.7
Switzerland	0	0.0	364	94.3	22	5.7	0	0.0	395	74.0	139	26.0
Türkiye	0	0.0	1911	84.8	343	15.2	1	0.2	394	83.0	80	16.8
Ukraine	3	0.9	248	75.6	77	23.5	3	3.0	51	50.5	47	46.5
United Kingdom	0	0.0	2750	67.1	1350	32.9	0	0.0	2666	44.8	3280	55.2
Total	167	0.7	19142	82.0	4047	17.3	113	0.4	16553	61.9	10061	37.6

Note: Albania and Georgia have <5 adults seen in 2023 and are excluded from the table for adults, but the people are included in the total number.

Note: Oral proton pump inhibitors are reimbursed in most countries except in Bulgaria, Georgia, Kazakhstan, Lithuania and Serbia. In Armenia, it is reimbursed for some outpatients.

Appendix 2 List of contributing centres and national registries

This is a list of the individual centres and the national registries, with their representatives, that contributed data to the ECFSPR in 2023. In turquoise: the name of the country representative in the ECFSPR Steering Group; underlined: the name of the database manager for the national registry; in italics: new participants with 2023 data.

Country	Centre/National Registry name	Contact
Albania	1 individual centre:	Irena Kasmi
	"Mother Thereza" Hospital Centre, Department of Paediatrics, Tirana	Irena Kasmi Evda Vevecka
Armenia	2 individual centres:	Satenik Harutyunyan
	Yerevan State Medical University, Muratsan University Hospital, Cystic Fibrosis Centre, Yerevan	Satenik Harutyunyan Vachagan Baghdasaryan
	Arabkir Medical Centre-Institute of Child and Adolescent Health, Yerevan	*Aghavni Sararyan*
Austria	13 individual centres:	Andreas Pfleger Dorothea Appelt
	Medizinische Universität Graz, Universitätsklinik für Kinder- und Jugendheilkunde, Klinische Abteilung für Pädiatrische Pulmonologie und Allergologie und CF Zentrum für Kinder, Jugendliche und Erwachsene, Graz	Ernst Eber Andreas Pfleger Maria Gaber Manfred Modl Doris Malle-Scheid
	Medizinische Universität Innsbruck, Zertifiziertes CF Zentrum für Kinder, Jugendliche und Erwachsene, Innsbruck	Dorothea Appelt Johannes Eder Helmut Ellemunter
	Klinikum Klagenfurt am Wörthersee, Abteilung für Kinder- und Jugendheilkunde, Pädiatrische Pulmologie/ Allergologie, Klagenfurt	Franz Hubert Wadlegger Marc Schlapschy
	Kepler Universitätsklinikum, Universitätsklinik für Kinder- und Jugendheilkunde, Linz	Claudia Altmann
	Kepler Universitätsklinikum, Klinik für Lungenheilkunde/ Pneumologie, Linz	Katrin Scheich
	Kardinal Schwarzenberg Klinikum, Abteilung für Kinder- und Jugendmedizin, Schwarzach im Pongau	Josef Riedler Christoph Seelbach
	PEK Klinikum Steyr, Abteilung für Kinder- und Jugendheilkunde und Abteilung für Lungenheilkunde, Steyr	Alexander Ebner Margit Kallinger Monika Pell
	Medizinische Universität Wien, Allgemeines Krankenhaus Wien für Thoraxchirurgie, Vienna	Peter Jaksch Dagmar Liebhart
	Medizinische Universität, Allgemeines Krankenhaus Wien, Universitätsklinik für Kinder-und Jugendheilkunde, Klinische Abteilung für Pädiatrische Pneumologie, Allergologie und Endokrinologie, Zentrum für Cystische Fibrose, Vienna	Sabine Renner Saskia Gruber Brigitte Mersi
	Klinik Ottakring, Abteilung für Kinder- und Jugendheilkunde mit Ambulanz, Vienna	Mehtap Schmidt
	Klinik Hietzing, Abteilung für Atmungs- und Lungenkrankheiten, Vienna	Andrea Lakatos–Krepcik
	Klinikum Wels-Grieskirchen, Abteilung für Kinder- und Jugendheilkunde, Wels	Beatrix Wintersteiger Vera Karin Bauer
	Klinikum Wels-Grieskirchen, Abteilung für Lungenkrankheiten, Wels	Alexander Leitner Thomas Tempelmayer
Belarus	1 individual centre:	Sviatlana Keegan
	Belarusian Republic Children's Centre of Pulmonology and Cystic Fibrosis, Pulmonary Department, 3rd City Children's Clinical Hospital, Minsk	Vladimir Bobrovnichiy Sviatlana Keegan Katsiaryna Chyrkun

Country	Centre/National Registry name	Contact
Bulgaria	2 individual centres: Alexandrovska University Hospital, Paediatric Clinic, Sofia University Hospital St. Marina, 2nd Paediatric Clinic, Varna	Guergana Petrova Guergana Petrova Miglena Georgieva Margarita Nikolova
Croatia	1 individual centre: University Hospital Centre Zagreb, Cystic Fibrosis Centre – Paediatrics and Adults, Zagreb On behalf of the Croatian people with CF Database	Duska Tješić-Drinković Andrea Vukić Dugac Duska Tješić-Drinković Andrea Vukić Dugac Ivan Bambir Ivona Markelić
Cyprus	1 individual centre: Medical School, University of Cyprus, children and adults, Nicosia	Panayiotis Yiallouros Panayiotis Yiallouros Panayiotis Kouis Pinelopi Anagnostopoulou
Czech Republic	Cystic Fibrosis Registry of the Czech Republic	Pavel Drevinek Alena Bilkova Milan Macek Marek Turnovec
Denmark	Cystic Fibrosis Registry Denmark	Hanne Vebert Olesen Tacjana Pressler
Estonia	*4 individual centres:* *North Estonia Medical Centre, Internal Medicine, Clinical Department of Pulmonology, Tallin* *Tallin Children's Hospital, Paediatric Allergology and Pulmonology Unit, Tallin* *Lung Clinic of Tartu University Hospital* *Children's Clinic of Tartu University Hospital, Tartu*	*Maire Vasar* *Liina Viks* *Silvi Plado* *Viktoria Ivanova* *Maire Vasar*
Finland	Cystic Fibrosis in Finland	Varpu Elenius Katriina Pihlajamaa Aleksi Kemppainen
France	Registre Français de la Mucoviscidose	Antoine Bessou Kadiatou Kaba
Georgia	1 individual centre: LTD, Medical Genetics and Laboratory Diagnostic Centre, Tbilisi	Dodo Agladze Dodo Agladze
Germany	German Cystic Fibrosis Registry	Lutz Naehrlich Julia Wosniok
Greece	Cystic Fibrosis Registry of Greece	Elpis Hatziagorou John Tsanakas Panagiota Mitrou Kostas Mathioudakis Maria Davis
Hungary	Cystic Fibrosis Registry of Hungary	Andrea Párniczky Géza Marsal
Iceland	1 individual centre: Children´s Medical Centre Landspitali – The National University Hospital of Iceland, Reykjavik	Helga Elidottir Helga Elidottir Olafur Baldursson
Ireland	Cystic Fibrosis Registry of Ireland	Godfrey Fletcher Laura Kirwan

Country	Centre/National Registry name	Contact
Israel	6 individual centres:	Meir Mei-Zahav
	Soroka University Medical Centre, Ben Gurion University of the Negev, Beer Sheva	Inbal Golan-Tripto
	Carmel Medical Centre, Haifa	Galit Livnat
	Ruth Rappaport Children's Hospital, Rambam Medical Centre, Haifa	Michal Gur
	Hadassah Medical Centre, Mount Scopus, Jerusalem	Malena Cohen-Cymberknoh
	Schneider Children's Medical Centre of Israel, Petah Tikva, Israel; Faculty of Medical and Health Sciences, Tel Aviv University, Tel Aviv	Meir Mei-Zahav Moshe Heching
	Safra Children's Hospital, Sheba Medical Centre, Ramat Gan	Ori Efrati
Italy	Italian Cystic Fibrosis Registry	Rita Padoan Marco Salvatore Annalisa Amato Gianluca Ferrari
Kazakhstan	*1 individual centre:*	*Elena Amelina*
	Multidisciplinary City Hospital No. 1, Astana	*Irina Mukatova* *Elena Amelina*
Latvia	1 individual centre:	Elina Aleksejeva
	Rīga Stradiņš University, Children's Clinical University Hospital, Department of Pneumology, Riga	Elina Aleksejeva Dita Gaidule-Logina
Lithuania	2 individual centres:	Kęstutis Malakauskas
	Hospital of Lithuanian University of Health Sciences Kauno Klinikos, Adult Cystic Fibrosis Centre, Kaunas	Kęstutis Malakauskas Virginija Kalinauskaitė-Žukauskė
	Hospital of Lithuanian University of Health Sciences Kauno Klinikos, Centre of Paediatric Chronic Respiratory Diseases, Kaunas	Valdonė Misevičienė
Luxembourg	1 individual centre:	Anna-Maria Charatsi
	Centre Hospitalier de Luxembourg, Department of Paediatrics and Department of Pulmonology, Luxembourg	Anna-Maria Charatsi Michael Sieren
Rep. of North Macedonia	2 individual centres:	Tatjana Jakovska-Maretti Stojka Fustik
	Centre for Cystic Fibrosis - Children and Adults, University Clinic for Respiratory Diseases in Children-Kozle, Skopje	Tatjana Jakovska-Maretti Ivana Arnaudova Danevska
	University Children's Hospital, Centre for Cystic Fibrosis, Skopje	Valentina Cveovska Cholakovska Stojka Fustik Andrijana Andreevska
Rep. of Moldova	1 individual centre:	Oxana Turcu
	Outpatient Centre for Cystic Fibrosis and Other Rare Diseases, Chisinau	Oxana Turcu
Montenegro	*1 individual centre:*	*Tomo Plamenach*
	Institute for Children's Diseases, Clinical Centre of Montenegro, Podgorica	*Tomo Plamenach*
Netherlands	Dutch Cystic Fibrosis Registry	Domenique Zomer Renate Kos
Norway	Norwegian Cystic Fibrosis Patient Registry	Egil Bakkeheim Magnhild Louise Pollestad Kolsgaard

Country	Centre/National Registry name	Contact
Poland	19 individual centres:	Łukasz Woźniacki
	2nd Department of Lung Diseases and Tuberculosis, Medical University of Bialystok, Bialystok	*Łukasz Minarowski*
	Voivodeship Children's Hospital, Dept. of Paediatric Pneumology and Allergology, Bydgoszcz	Radoslawa Staszak–Kowalska Mikolaj Kowalski
	Cystic Fibrosis Centre, Polanki Paediatric Hospital, Gdansk	Maria Trawinska-Bartnicka Ewa Sapiejka Anna Steinert-Dymecki
	Centrum Medyczne Karpacz, Children/Adults' Hospital, Karpacz	Grzegorz Gaszczyk Monika Rams
	John Paul II Upper Silesian Child Health Centre, The Independent Public Clinical Hospital no. 6 of the Medical University of Silesian in Katowice, Katowice	Urszula Grzybowska-Chlebowczyk Bozena Kordys-Darmolinska
	Paediatric Clinic Holy Cross Paediatric Centre, Provincial Integrated Hospital in Kielce, Kielce	Elżbieta Kołodziej Maciej Szczukocki
	St. Louis Regional Specialised Children's Hospital, Krakow	Stanislaw Stepniewski Daria Dziecichowicz-Latala
	The University Hospital in Krakow, Pulmonology and Allergology Clinical Department, Krakow	Krysztof Sladek Iwona Gross-Sondej
	Barlicki Hospital, Medical University of Lodz, Department of General and Oncological Pulmonology, Lodz	Małgorzata Pietrusinska
	Wojewódzkie Wielospecjalistyczne centrum Onkologii i Traumatologii im. M. Kopernika w Lodzi, Ośrodek Pediatryczny im. J. Korczak, Lodz	Agnieszka Brzozowska Agnieszka Koniarek-Maniecka Katarzyna Kapszewicz
	Cystic Fibrosis Centre for Adults, Independent Hospital No. 4, Lublin	Irena Węgrzyn-Szkutnik Adam Krusiński
	University Hospital of Lords Transfiguration, Dept. of Pulmonology, Allergology and Pulmonary Oncology, Poznan	Szczepan Cofta Daria Springer Hanna Winiarska
	Karol Jonscher University Hospital of Poznan University of Medical Sciences, Poznan	Irena Wojsyk-Banaszak Agnieszka Korytowska-Niklas
	Institute of Tuberculosis and Lung Diseases, Rabka-Zdrój Branch, Dept. of Pneumology and Cystic Fibrosis, Rabka Zdroj	Henryk Mazurek Andrzej Pogorzelski Lidia Pawlik
	Provincial Clinical Hospital no. 2, St. Queen Jadwiga, Dept of Allergology and Cystic Fibrosis, St Jadwigi Krolowej in Rzeszów, Rzeszów	Marta Rachel
	Szczecin Hospital "Zdroje" Dep. Of Pediatrics, Allergology and Pulmonology, Szczecin	Pawel Gonerko Pawel Fabisiak
	Lubuski Institute of Pulmonary Medicine, Adult Cystic Fibrosis Treatment Centre, Torzym	Michal Karolak Agnieszka Szklarska
	Dziekanow Paediatric Hospital, Cystic Fibrosis Centre, Institute of Mother and Child, Warsaw	Dorota Sands Łukasz Woźniacki
	Institute of Tuberculosis and Lung Diseases, Adult CF Centre, Warsaw	Wojciech Skorupa Sylwia Ziernik
Portugal	Cystic Fibrosis Registry of Portugal	Luisa Pereira
Romania	10 individual centres:	Liviu Pop
	Regional Cystic Fibrosis Centre, Clinical Emergency Children's Hospital of Brasov, Brasov	Laura Larisa Dracea
	Clinical Children's Hospital "Grigore Alexandrescu", Bucharest	Simona Mosescu Livia Brezeanu

Country	Centre/National Registry name	Contact
	Emergency Hospital for Children Marie Curie" – Paediatrics 3, Bucharest	Maria Iulia Brustan Ioana Gradinaru
	Institute of Pneumology "Marius Nasta", Adult Centre, Bucharest	*Cristi Popa*
	Mother & Child Health Institute, Bucharest	Iustina Stan Valentina Comanici
	Sf. Apostol Andrei County Emergency Clinical Hospital, Department. of Paediatrics, Regional CF Centre, Constanta	*Cristina Andrei*
	Regional Cystic Fibrosis Centre Cluj, Clinical Emergency Hospital for Children of Cluj-Napoca, Cluj-Napoca	Radu Sorin Șerban Szabo Csilla-Enikő
	Regional Cystic Fibrosis Centre, "Sf. Maria" Children Emergency Hospital Iasi, Iasi	Dana-Teodora Anton-Paduraru
	National Cystic Fibrosis Centre- County Emergency Clinical Hospital "Pius Branzeu" Timisoara	Liviu Pop Ioana Ciuca
	Pulmonology Clinic, Adult CF Centre, Clinical Hospital of Infectious Diseases and Pulmonology "Victor Babes" University of Medicine and Pharmacy Timisoara,	*Camelia Corina Pescaru* *Adelina Maritescu*
Russian Federation	Cystic Fibrosis Registry of the Russian Federation	Elena Kondratyeva Marina Starinova Stanislav Krasovskiy Anna Voronkova Nataliya Kashirskaya Elena Amelina
Serbia	1 individual centre: National Centre for Cystic Fibrosis, Mother and Child Health Institute of Serbia "Dr Vukan Čupić", Belgrade	Milan Rodić Predrag Minić Milan Rodić Aleksandar Sovtić
Slovakia	6 individual centres: Children's CF Centre, DFN Banská Bystrica, Banská Bystrica	Hana Kayserova Branko Takáč Ivana Gondová
	Centrum Cystickej Fibrozy Pre Dospelych FNSP FDR, Banská Bystrica	Eva Bérešova
	Centrum Cystickej Fibrozy Pre Dospelych, Klinika Pneumologie l.SZU a Univerzitna Nemocnica, Bratislava	Marta Hajkova
	Klinika detskej Pneumologie SZU UN Bratislava, Pracovisko Podunajské Biskupice, Bratislava	Hana Kayserova Nina Bližňáková
	CF Adult centre, University Hospital L Pasteura, Košice	Lenka Kopčová
	Centrum Cystickej Fibrozy Detí, Detská Fakultná Nemocnica Košice, Košice	Anna Fetekeova Zuzana Hribíková
Slovenia	3 individual centres: University Clinic of Pulmonary and Allergic Diseases, Golnik	Uroš Krivec Matjaž Fležar Julij Šelb
	University Medical Centre Ljubljana, Department of Pulmonology and Allergy, Ljubljana	Barbara Salobir Maja Badinovac
	University Medical Centre Ljubljana, University Children`s Hospital, Department of Paediatric Pulmonology, Ljubljana	Uroš Krivec Jasna Rodman Berlot
Spain	25 individual centres Parc Taulí Hospital Universitario, Hospital de Sabadell, Unitat de Pneumologia Pediátrica i Unitat de Fibrosi Quística, Sabadell, Barcelona	M. Dolores Pastor Vivero Oscar Asensio de la Cruz Miguel Garcia Gonzàlez Xavier Pomares Amigó Concepción Montón Soler
	Hospital Sant Joan de Déu, Unitat de Pneumologia Pediàtrica i Fibrosi Quística, Barcelona	Maria Cols i Roig Jordi Costa i Colomer
	Hospital Universitari Vall d'Hebron, Unidad de Fibrosis Quística del Adulto, Barcelona	Antonio Alvarez Fernández Eva Polverino

Country	Centre/National Registry name	Contact
Spain (cont.)	Hospital Universitari Vall d'Hebron, Unidad Fibrosis Quística y Neumología Pediátrica, Barcelona	Silvia Gartner Sandra Rovira Amigo
	Hospital Universitari Vall d'Hebron, Unidad de Fibrosis Quística del Adulto, Barcelona	Antonio Alvarez Fernández Eva Polverino
	Hospital Universitari Vall d'Hebron, Unidad Fibrosis Quística y Neumología Pediátrica, Barcelona	Silvia Gartner Sandra Rovira Amigo
	Hospital Universitario Cruces, Unidad de Fibrosis Quística, Bizkaia	M. Dolores Pastor Vivero Ainhoa Gómez Bonilla Beatriz Gómez Crespo
	Hospital Universitario Reina Sofia, Unidad de Alergia y Neumología Pediátricas y UGC Neumología, Facultad de Medicina e Instituto Maimónides de Investigación Biomédica de Córdoba (IMIBIC), Cordoba	Javier Torres Borrego José Manuel Vaquero Barrios
	Complejo Hospitalario Universitario Insular Materno Infantil, Las Palmas de Gran Canaria	Antonio José Aguilar Fernández
	Hospital Universitario La Paz, Unidad de Fibrosis Quística Adultos, Servicio de Neumología, Madrid	Concha Prados
	Hospital Universitario La Paz, Sección de Neumología Pediátrica, Unidad de Fibrosis Quística Pediátrica, Madrid	Marta Ruiz de Valbuena Maiz, Cristina de Manuel Gómez
	Hospital Universitario La Princesa, Neumología Adultos, Madrid	Rosa María Girón Rosa Mar Gómez-Punter
	Hospital Niño Jesús, Sección de Neumología Pediátrica, Unidad de Fibrosis Quística, Madrid	Alejandro López Neyra Verónica Sanz Santiago José R. Villa Asensi
	Hospital Universitario La Paz, Sección de Neumología Pediátrica, Unidad de Fibrosis Quística Pediátrica, Madrid	Marta Ruiz de Valbuena Maiz, Cristina de Manuel Gómez
	Hospital Universitario La Princesa, Neumología Adultos, Madrid	Rosa María Girón Rosa Mar Gómez-Punter
	Hospital Niño Jesús, Sección de Neumología Pediátrica, Unidad de Fibrosis Quística, Madrid	Alejandro López Neyra Verónica Sanz Santiago José R. Villa Asensi
	Hospital Universitario Ramón y Cajal, Unidad de Fibrosis Quística, Madrid	Luis Maiz Carro Saioa Vicente Santamaria Enrique Blitz Castro Rosa Maria Nieto Royo Ana Morales Tirado
	Hospital Universitario 12 de Octubre, Unidad de Fibrosis Quística Pediátrica, Madrid	Carmen Luna Paredes Enrique Salcedo Lobato
	Hospital Universitario 12 de Octubre, Unidad de Fibrosis Quística Adultos, Madrid	Layla Diab Cáceres
	Hospital Regional Universitario de Málaga, Unidad Fibrosis Quística Adultos de Andalucía Oriental, Málaga	Casilda Olveira Fuster Gabriel María Olveira Fuster
	Hospital Regional Universitario de Málaga, Unidad de Fibrosis Quística Pediátrica, Málaga	Estela Pèrez-Ruiz Pilar Caro-Aguilera Juan Carlos Ramos Díaz
	Hospital Clínico Universitario Virgen de la Arrixaca, Unidad de Fibrosis Quística, Murcia	Pedro Mondéjar-López Silvia Lorca Mayor
	Hospital Universitario Central de Asturias, Unidad de Fibrosis Quística, Oviedo	José Ramón Gutiérrez Martínez David González Jimenez Marta Garcia Clemente
	Hospital Universitario Son Espases, Servicio de Neumología y Servicio de Pediatría, Unidad de Neumología y Alergia Pediátrica, Palma de Mallorca	Alexandre Palou-Rotger Catalina Bover-Bauza Joan Figuerola Mulet Leticia Rubia de Azevedo

Country	Centre/National Registry name	Contact
Spain (cont.)	Hospital Universitario Virgen del Rocío, Unidad de Fibrosis Quística, Sevilla	Isabel Delgado Pecellín Esther Quintana Gallego Laura Carrasco Hernández
	Hospital Universitario Nuestra Señora de Candelaria, Santa Cruz de Tenerife, Tenerife	Alicia Callejón Orlando Mesa Medina
	Hospital Clínico Universitario de Valencia, Unidad de Fibrosis Quística Pediátrica, Valencia	Silvia Castillo Corullón
	Hospital Universitario y Politécnico La Fe, Unidad de Trasplante Pulmonar y Fibrosis Quística, Valencia	Amparo Solé Jover Carmen Inés Perez Munoz
	Hospital Álvaro Cunqueiro, Servicio de Neumología y Servicio de Pediatría, Vigo	Cristina Ramos Hernández María Jesús Rodriguez Sáez
	Hospital Universitario Miguel Servet, Unidad de Neumología Pediátrica y Fibrosis Quística, Zaragoza	Carlos Martín de Vicente
Sweden	Cystic Fibrosis Registry of Sweden	Christina Krantz Anders Lindblad
Switzerland	20 individual centres:	Andreas Jung
	Kinderspital Aarau, Kantonsspital Aarau AG, Abteilung pädiatrische Pneumologie, Allergologie und Immunologie, Aarau	Dominik Müller-Suter
	Kantonsspital Aarau AG, Klinik für Pneumologie und Schlafmedizin, Aarau	G. Mauro Tini Lydia Eisenmann
	Universitätsspital Basel, Klinik für Pneumologie, Adulte Cystische Fibrose, Basel	Kathleen Jahn
	UKBB Universitäts-Kinderspital beider Basel, Abteilung Intensivmedizin & Pneumologie, Basel	Daniel Trachsel Anja Jochmann Jakob Usemann
	Inselspital Bern, Universitätsklinik für Pneumologie, Adulte Cystische Fibrose, Bern	Dagmar Lin Michaela Semmler
	Lindenhofspital Quartier Bleu, Bern	Bernhard Schwizer Iris Schmid
	Universitätsklinik für Kinderheilkunde, Zentrum für Cystische Fibrose und Pulmonologie, Inselspital, Bern	Philipp Latzin Carmen Casaulta
	Hôpital Cantonal Fribourg, Pädiatrie, Fribourg	Maxime Hensen Johannes Wildhaber
	Hôpitaux Universitaires de Genève, Département de la Femme, de l'Enfant et de l'Adolescent, Unité de Pneumologie Pédiatrique, Genève	Anne Mornand Nadège Gabent
	Hôpitaux Universitaires de Genève, Département de Médecine, Service de Pneumologie, Consultation de Mucoviscidose Adulte, Genève	Jérôme Plojoux Valerie Durand
	Centre Hospitalier Universitaire Vaudois (CHUV), Département femme-mère-enfant, Service de pédiatrie, Unité de pneumologie et mucoviscidose pédiatrique, Lausanne	Isabelle Rochat Laurence Mioranza
	Consultation de Mucoviscidose Adulte et de CFTR-related Disorders, Service de Pneumologie, Département de Médecine, Centre Hospitalier Universitaire Vaudois (CHUV), Lausanne	Angela Koutsokera Marie-France Derkenne Isabelle Huart Bellavere
	Luzerner Kantonsspital, Zentrum für Zystische Fibrose für Kinder und Jugendliche, Luzern	Nicolas Regamey Michael Hitzler Lucia Eichhorn Sonja Ettlin
	Luzerner Kantonsspital, Abteilung für Pneumologie, Zentrum für Cystische Fibrose für Erwachsene, Luzern	Christian Murer Gabriele Riedener Luzia Rytz
	Hôpital Neuchâtelois – Pourtales, Consultation de Mucoviscidose Adulte, Neuchâtel	Jean Marc Fellrath Sidikka Ozturk-Beungies

Country	Centre/National Registry name	Contact
Switzerland (cont.)	Children's Hospital of Eastern Switzerland, Division of Paediatric Pulmonology & CF Centre, St Gallen	Jürg Barben Katharina Hartog Christine Baumgartner
	Kantonsspital St. Gallen, Lungenzentrum, Zentrum für Cystische Fibrose für Erwachsene, St. Gallen	Anna-Lena Walter Martin Brutsche Rebekka Kleiner
	Kantonsspital Winterthur, Klinik für Pneumologie und Klinik für Innere Medizin, Adulte Cystische Fibrose, Winterthur	Markus Hofer
	Universitäts-Kinderspital Zürich, Abteilung für Pneumologie, Zürich	Andreas Jung Alexander Möller Eugènie Collaud Rachel Kusche Djamila Laban
	Universitätsspital Zürich, Klinik für Pneumologie, Adultes CF Zentrum, Zürich	Macé Schuurmans Carolin Steinack Thomas Kurowski
Türkiye	Cystic Fibrosis Registry of Türkiye	Deniz Dogru
	Cystic Fibrosis Registry of Türkiye, Ankara	Deniz Dogru
	Marmara University, Faculty of Medicine, Division of Paediatric Pulmonology, Istanbul	Bülent Karadağ Yasemin Gökdemir Ela Erdem Eralp
	Medipol University, Faculty of Medicine, Division of Paediatric Pulmonology, Istanbul	Füsun Ünal Sedat Oktem
	Medeniyet University, Faculty of Medicine, Division of Paediatric Pulmonology, Istanbul	Saniye Girit Zeynep Reyhan Onay Özge Ülgen
Ukraine	18 individual centres:	Halyna Makukh
	KNP "City Multidisciplinary Hospital of Mother and Child - Prof. Rudnev", Dnipro	Olga Lacinska-Prykhodko Anastasiia Fialkovska
	Ivano-Frankivsk Regional Children's Clinical Hospital of Ivano-Frankivsk Regional Council, Department of Pulmonology, Ivano-Frankivsk	Sirun Makian Olha Fedynska
	Municipal non-profit enterprise «Khmelnytskyi Regional Children's Hospital» of Khmelnytskyi Regional Council, Khmelnytskyi	Liliya Brukhnova Olga Yevchuk
	Regional Clinical Children's Hospital of the Kirovohrad Region, Kropyvnytskyi	Yuriy Chornyi Vasil Khoroshchak
	Municipal Non-Profit Enterprise "Kyiv Regional Children's Hospital", Boyarka, Kyiv	*Anna Mykytiuk*
	Okhmatdyt Specialised Children's Hospital, Centre for Orphan Diseases & Gene Therapy, Kyiv	*Yuliia Ostapyshena*
	Volyn Regional Children's Hospital, Paediatric Department and Volyn Regional Clinical Hospital, Pulmonology Department, Lutsk	Miroslava Melnyk Oleh Yakovenko
	Cystic Fibrosis Centre of Western Ukrainian Specialised Children's Medical Centre, Lviv	Lyudmyla Bober Halyna Makukh
	Municipal non-profit enterprise «Regional Children's Hospital» of the Transcarpathian Regional Council, Mukachevo	Elizaveta Birov Khrystyna Petrychko
	Mykolaiv Children's Regional Clinical Hospital, Mykolaiv	Natalia Lesnycha Olexandr Plytkin
	Odesa Regional Children's Clinical Hospital, Department of Specialised Care for Older Children, Odesa	Iryna Holovenko Pavlo Heorhiiev
	Odesa Regional Clinical Hospital, Odesa	Iryna Gonta Yuri Gulchencko

Country	Centre/National Registry name	Contact
Ukraine (cont.)	*Regional Centre of Medical Genetics, Poltava*	*Olena Tul*
	Regional Specialized Dispensary Cystic Fibrosis Centre, Rivne	*Oksana Yarmoiyuk*
	Communal non-commercial enterprise Sumy Regional Council «Regional Children's Clinical Hospital», Sumy	Olga Kolomiets Ihor Zmyslya
	Ternopil Regional Children's Hospital, II Children's Department / I. Horbachevsky Ternopil National Medical University of the Ministry of Health of Ukraine, Department of Children's Diseases with Children's Surgery, Ternopil	Iryna Shostak Oksana Boyarchuk Lesia Dobrovolska
	Communal non-profit enterprise «Vinnytsia Regional Children's Clinical Hospital Vinnytsia Regional Council», Department of Paediatrics #2, Vinnytsia	Valeriia Demianyshyna Oksana Moravska
	Municipal non-profit enterprise «Zaporizhzhya Regional Clinical Children's Hospital» Zaporizhzhya Regional Council, Pulmonary Department, Zaporizhzhya	Tetyana Okul Irina Kolman
United Kingdom	UK Cystic Fibrosis Registry, Cystic Fibrosis Trust, London	Sarah Clarke Susan Charman Elaine Gunn Jamie Duckers

Appendix 3 Inclusion criteria and technical notes

People with CF: inclusion criteria
The ECFSPR registers people diagnosed with CF in accordance with agreed definitions:
Two sweat tests value > 59 mmol/L chloride: CF diagnosis accepted.
One sweat test value > 59 mmol/L chloride and DNA Analysis/Genotyping – two identified disease-causing CF variants: CF diagnosis accepted.

Sweat value ≤ 59 mmol/L chloride:
If the sweat value is less than or equal to 59 mmol/L chloride or not reported, then at least 2 of these must be fulfilled:
DNA Analysis/Genotyping: two identified disease-causing CF variants;
Transepithelial (Nasal) Potential Difference or Intestinal Current Measurement: result consistent with a diagnosis of CF;
Clinical Presentation: typical features of CF.

Diagnosis reversal:
If the CF diagnosis was reversed during the year, one of the options must be true:
DNA Analysis: unable to identify two disease causing CF variants;
Transepithelial (Nasal) Potential Difference and/or Intestinal Current Measurement: result not consistent with a diagnosis of CF;
Repeated normal values from sweat tests and confirmed by the clinical team.

References

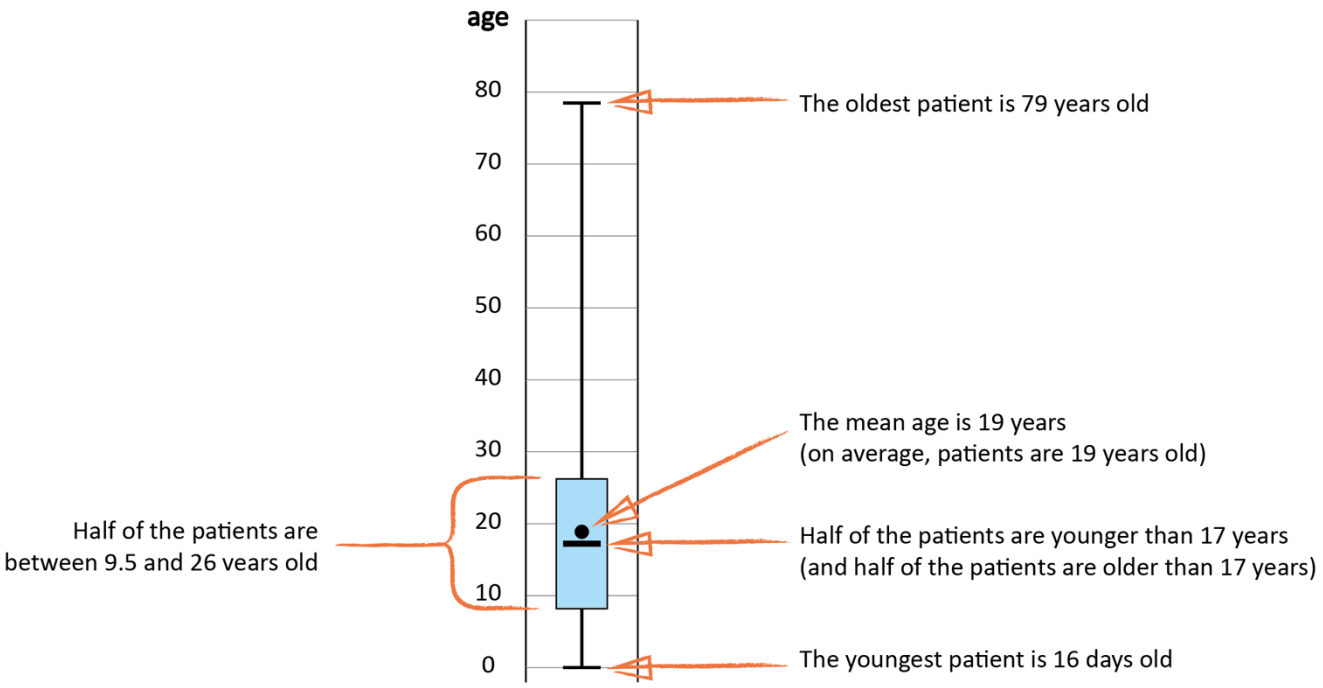
1) ECFS best practice guidelines: the 2018 revision

2) European Cystic Fibrosis Society Standards of Care: Best Practice guidelines (2013)

Data manipulation
To ensure that data are anonymous, the ECFS collects only year and month of birth and the day of birth is set to the 15th of the month.
For prenatal diagnoses, we set age at diagnosis equal to 0.
We checked for outliers and, whenever possible, we corrected the values according to the instructions of the national registries / individual centres. If, after the data quality controls, aberrant values were still present in the database, we set them to missing.
Software used for data management and statistical analyses: SAS software, Version 9.4. Copyright, SAS Institute Inc. SAS and all other SAS Institute Inc. product or service names are registered trademarks or trademarks of SAS Institute Inc., Cary, NC, USA.

Explanation of statistical terms
Max: maximum. It is the highest value.
Mean: it is the average value of a set of measurements. For example, if the mean age at diagnosis is 3 years, it means that, on average, the people are diagnosed when they are 3 years old.
Median: the value that separates the set of measurements in two halves, so that 50% of measurements are below the median value and the other 50% of measurements are above the median value. For example, if median age at diagnosis is 5 months, it means that half of the people are diagnosed before 5 months of age, and the other half of the people are diagnosed after 5 months of age.
Min: minimum. It is the lowest value.
N: the number of people in a group for whom the information is not missing.
N miss: number of missing values. It is the number of people for whom the information is missing.

Quartiles: the 25th percentile, the median (the 50th percentile) and the 75th percentile are collectively called quartiles, because they divide the set of measurements into quarters.
25th Pctl: 25th percentile, also called first quartile. It is the value that separates the set of measurements in two parts, so that one quarter (25%) of the measurements is below it and the other three quarters are above it. For example, if the 25th percentile for age at diagnosis is 1 month, it means that a quarter of the people were diagnosed before they were a month old, and the other three quarters were diagnosed after they were a month old.
50th Pctl: 50th percentile, also called second quartile or median (please refer to definition of Median).
75th Pctl: 75th percentile, also called third quartile. It is the value that separates the set of measurements in two parts, so that three quarters (75%) are below it and the other quarter is above it. For example, if the 75th percentile for age at diagnosis is 3 years, it means that three quarters of the people are diagnosed before they were 3 years old, and the remaining quarter was diagnosed after they reached 3 years of age.



Note: This is an example of how to read a boxplot. The numbers used in this figure are not real.

Appendix 4 Variables and definitions used by the ECFSPR

Demographics

CF centre code
Centre Patient code (optional)
Year of follow-up
Year and month of birth
Sex (Previously "Gender")
Ethnicity
Vital Status of patient
Cause of death
Date of death

Diagnosis

Age at diagnosis
Sweat test type and values (x2)
First & second variants (possible to record complex variants in cis)
Meconium Ileus
Neonatal screening
Nasal Potential Difference Measured? (NPD)
CF-typical NPD measurement Yes/No
Date of NPD measurement
Intestinal current value measured? (ICM)
CF-typical IC measurement Yes/No
Date of IC measurement

Complications

ABPA (Allergic bronchopulmonary aspergillosis)
Distal intestinal obstruction syndrome (DIOS) this year
Salt loss syndrome this year
Diabetes this year
Pneumothorax this year
Liver disease this year
Haemoptysis major volume of expectorate > 250ml in a day
Occurrence of malignancy - diagnosed this year
Pancreatic status: faecal elastase
Pancreatic status: faecal fat

Pregnancy

Pregnancy this year
Pregnancy stopped this year - reason for stop
Pregnancy ongoing at 31/12

Maintenance Therapy

Inhaled continuous (≥ 3 months) hypertonic saline ≥3%
Inhaled continuous (≥ 3 months) Mannitol
Inhaled antibiotic this year - continuous (≥ 3 months) or on/off for a total of (≥ 6months)
Inhaled continuous (≥ 3 months) bronchodilators, long-acting or short-acting or both
Oxygen therapy ≥ 3 months during the year of follow-up (inc. 24h/day, nighttime, exercise).
Does not need to be continuously but should be from a single prescription)
Use of continuous (≥ 3 months) non-invasive positive pressure ventilation (NIPPV)
Use of continuous (≥ 3 months) rhDNase this year
Use of continuous (≥ 3 months) Inhaled steroids
Use of continuous (≥ 3 months) Oral steroids
Use of continuous (≥ 3 months) azithromycin (or other macrolide) this year
Use of continuous (≥ 3 months) ursodeoxycholic acid this year
Use of continuous (≥ 3 months) pancreatic enzymes this year
Use of continuous (≥ 3 months) proton pump inhibitors (PPI)
Use of CFTR Modulator Therapy (data for each of the following are collected:
Ivacaftor, Lumacaftor /Ivacaftor, Tezacaftor/Ivacaftor,
Elexacaftor/Tezacaftor/Ivacaftor,
Other CFTR Modulator
Start and stop dates x 2 for each CFTR modulator
Sweat chloride values - before start and during (lowest of the year) for each CFTR modulator

Lung function and nutrition follow-up

Value of FEV1 in litres of highest FEV1% predicted of the year
Value of FVC in litres (from same spirometry as recorded FEV1)
Height measured at date of best FEV1 (or if no available FEV1, last height of the year)
Weight measured at date of best FEV1 (or if no available FEV1, last weight of the year)
Date of recorded FEV1 or if no FEV1 recorded, date of recorded height and weight
Lowest LCI 2.5% of the year
Type of device used for LCI measurement
Date of lowest LCI 2.5% this year
Value of lowest LCI 2.5% this year

Microbiology (*positive-chronic* or *positive-at least once/not chronic* for all pathogens)

Pseudomonas aeruginosa
MSSA
MRSA
Chronic Burkholderia cepacia complex
Stenotrophomonas maltophilia
Achromobacter spp
Haemophilus influenza
Nontuberculous mycobacteria cultured
Mycobacterium abscessus complex
Mycobacterium avium complex
Other mycobacteria
NTMB treated this year
Fungi investigated
Aspergillus fumigatus
Scedosporium spp

Hospitalisation, Pulmonary Exacerbations, IV Antibiotics

Total days on iv antibiotics at home and in hospital this year (CF-related reasons)
Total days on iv antibiotics in hospital this year (CF-related reasons)
Total days in hospital this year (any reason)
Number of PExs treated with intravenous antibiotics during the year

Transplant

Liver transplant at any time
Year of latest liver transplant (before or during this year)
Lung transplant at any time
Year of latest lung transplant (before or during this year)
Kidney transplant at any time
Year of latest lung transplant (before or during this year)
Other transplant at any time
Year of latest other transplant (before or during this year)

Definitions and References

1 Sweat Test: Parameters, Values to be reported, References

i. Diagnostic standards: the quantity of sweat should indicate an adequate rate of sweat production;
ii. a. The sweat sample should be processed immediately after sweat collection;
b. Chloride concentration measurement is the preferred analysis for Diagnostic sweat tests. For sweat tests in relation to CFTR modulator therapy, Chloride is the only accepted value;
c. Chloride value: report the Chloride value in millimoles per litre (mmol/L). If duplicate tests were completed on the same day, for Diagnostic sweat tests, **report the highest positive value;**
d. A sweat chloride value >59 mmol/L is consistent with a diagnosis of CF;
e. A sweat chloride value <30 mmol/L makes the diagnosis of CF unlikely (However, specific CF-causing mutations are associated with a sweat test below 30 mmol/L).
n.b. *The acceptable range for Chloride values is 1-160 mmol/L.* ***Anyone who has a Chloride value above 160 mmol/L should be re-tested;***
iii. As already mentioned above, the ECFSPR will consider only Titration/Chloride values in analyses.

References:

1) ECFS best practice guidelines: the 2018 revision
2) European Cystic Fibrosis Society Standards of Care: Best Practice guidelines (2013)

2 Nutrition: Method, Values and Dates to be reported, References

i. The height and weight reported to the ECFSPR should be from the same day as the reported FEV1 (of the highest FEV1% predicted of the year);

ii. If spirometry was not done, the last weight and height measurements of the year, and the date they were measured, should be recorded;

iii. Height and weight should be measured in accordance with EuroCareCF guidelines:
Weight: removal of outer clothing, shoes and socks;
Height: removal of shoes and socks, stadiometer - top of head in contact with headboard, slight pressure.

iv. Z-scores for height, weight & BMI are calculated with the CDC reference values [Kuczmarski et al (2002)].

References:

1) Kromeyer-Hauschild K, Wabitsch M, Kunze D, Geller F, Geiss HC, Hesse V et al. Percentiles of body mass index in children and adolescents evaluated from different regional German studies. Monatsschr Kinderheilkd 2001; 149:807-818.
2) Lai H-C, Corey M, FitzSimmons S, Kosorok MR, Farrell M. Comparison of growth status of people with cystic fibrosis between the United States and Canada. Am J Clin Nutr 1999; 69:531-538.
3) Public Use File BGS98, German National Health Interview and Examination Survey 1998, Robert-Koch-Institut, Berlin, Germany, 2000.
4) Wiedemann B, Paul KD, Stern M, Wagner TO, Hirche TO, on behalf of the German CFQA Group. Evaluation of body mass index percentiles for assessment of malnutrition in children with cystic fibrosis. Eur J Clin Nutr 2007; 61, 759-768.
5) Kuczmarski RJ, Ogden CL, Guo SS et al. 2000 CDC Growth Charts for the United States: methods and development. Vital Health Stat 2002; 11(246): 1-190.

3 Spirometry: Criteria, Method, Values to be reported, References

The ECFSPR collects data on spirometry values to obtain standardised data for comparison with other centres/countries and for use in specific epidemiological studies. n.b. Some of the conditions for this (see below) may not be met at every clinical visit for all people and, for the ECFSPR, only spirometry tests fulfilling the criteria should be recorded by centres/submitted by the National Registries. **All spirometry tests should be carried out in accordance with the ATS/ERS guidelines.**

For the spirometry values reported to the ECFSPR the following criteria should be met:

i. **Pre-test preparation**

a. All recorded spirometry tests should be pre-bronchodilator* values:
   i. short-acting bronchodilators: at least 4 hours pre-test;
   ii. long-acting bronchodilators: at least 12 hours pre-test.

b. Date of birth, gender and height should be recorded for calculation of predicted values. In addition, the ECFSPR asks for the weight to be measured at the same time and recorded.

*****_In accordance with the official criteria of PortCF._

ii. **Values to report:**

a. FEV1 in litres: must be the FEV1 in litres (to max 2 decimals) of the **highest FEV1% predicted of the year**, in accordance with local reference values;

b. FVC in litres ((to max 2 decimals): must be the FVC measured at the same time as the FEV1 of the highest FEV1% predicted of the year and it must be greater than or equal to the FEV1 measurement.

c. For the reported spirometry values, the date of the test and the patient's height and weight at that date should also be recorded in order to calculate the percent of predicted values and other values;

d. Only tests deemed valid according to ATS/ERS guidelines to be reported.

iii. **Calculation of percent of predicted values:**

a. A common set of reference values - the Global Lung Function Initiative equations (See (1) below) - is used for calculations;

References:

1) Global Lung Function Initiative equations described by Quanjer PH et al. (Multi-ethnic reference values for spirometry for the 3-95-yr age range: the global lung function 2012 equations. Eur Respir J 2012; 40: 1324–1343).
2) Miller et al. Standardisation of spirometry. Eur Respir J 2005; 26: 319–338.
3) Miller et al. General considerations for lung function testing. Eur Respir J 2005; 26: 153–161.
4) Cystic Fibrosis Foundation Patient Registry User Guide, Version 4.0. 2006.
5) Rosenfeld et al. Task Force to Evaluate Choice of Spirometric Reference Equations for the National Patient Registry: Summary and Recommendations. Cystic Fibrosis Foundation Registry Committee; 2005.

4 Chronic infection in the lower airways: Definition, References

i. Chronic Pseudomonas aeruginosa infection: A patient should be considered chronically infected if the modified Leeds criteria are met - (a) below - and/or anti-pseudomonas antibodies are detected - (b)below. A patient should be defined as chronically infected if he/she fulfils the criteria now, or has done so in recent years, and the physician has no reason to think that the status has changed.

a. Modified Leeds criteria - chronic infection: >50% of the samples (sputum/other) collected during the last 12 months should be positive; at least 4 samples collected.

b. Significantly raised levels of anti-pseudomonas antibodies according to local laboratories.

ii. Chronic infection with other gram-negative (and also gram-positive) bacteria should be defined using the same criteria as described above.

References:

1) Lee TWR, Brownlee KG, Conway SP, Denton M, Littlewood JM. Evaluation of a new definition for chronic Pseudomonas aeruginosa in cystic fibrosis patients. J Cystic Fibrosis.
2) Proesmans M, Balinska-Miskiewiscz, Dupont L et al. Evaluating the “Leeds criteria” for Pseudomonas aeruginosa infection in a cystic fibrosis centre. Eur Resp J 2006;27:937-943.
3) Doring G, Conway SP, Heijerman HG, et al. Antibiotic therapy against Pseudomonas aeruginosa in cystic fibrosis: a European consensus. Eur Respir J 2000;16:749-767.

5 Liver Disease: Definitions

The ECFSPR has adopted the definitions for Liver Disease used by the Cystic Fibrosis Registry in the UNITED KINGDOM. These definitions discriminate people with severe liver disease (with portal hypertension) from milder cases (cirrhosis without portal hypertension).

- **Cirrhosis with Hypertension**: scarring of the liver related to underlying CF, typically in a biliary pattern. Severe liver disease may include portal hypertension and/or hypersplenism;
- **Cirrhosis without Hypertension**: scarring of the liver related to underlying CF;
- **Liver disease without cirrhosis**: this includes fatty liver or viral hepatitis but not biliary cirrhosis.

6 Pancreatic Status: Pancreatic Insufficiency, References

i. **Indicator of Pancreatic Insufficiency - Faecal Fat** (2 determinations are mandatory)

a. Young children: Stool fat (van de Kamer) > 4-5 g/d;

b. Children older than 10 years and adults: Stool fat (van de Kamer) >7g/d and/or faecal pancreatic elastase-1 < 200 ug/g.

Please note:

- Faecal fat excretion values of infants below 3 months are contradictory.
- Other than pancreatic causes of steatorrhoea must have been excluded.

ii. **For the ECFSPR, pancreatic status will be assessed as follows:**

- Pancreatic insufficiency: Faecal elastase <200 µg/g (twice), and faecal fat high* (twice);
- Pancreatic sufficiency: Faecal elastase ≥200 µg/g (twice) and faecal fat normal* (twice).

** Refer to 6.i.a and 6.i.b above*

References:

1) Sinaasappel M, Stern M, Littlewood J, Wolfe S, Steinkamp G, Heijerman HGM, Robberecht E, Döring G. Nutrition in patients with cystic fibrosis. A European consensus. J Cystic Fibrosis 2002; 1:51-75.
2) Walkowiak J, Nousia-Arvanitakis S, Henker J, Stern M, Sinaasappel M, Dodge JA. Invited review: Indirect pancreatic function tests in children. J Pediatr Gastroenterol Nutr 2005; 40:107-114.

7 Salt Loss Syndrome: Definition and Reference

Primary metabolic alkalosis with blood pH > 7.45, serum sodium < 130 mmol/l and serum chloride < 90 mmol/l (all 3 of these to be manifest).

Reference:

1) Fustik S, Pop-Jordanova N, Slaveska N, Koceva S, Efremov G. Metabolic alkalosis with hypoelectrolytemia in infants with cystic fibrosis. Pediatr int 2002; 44: 289-92.

8 Transplantation: Indications

i. For people with CF who had a transplant during the year of follow up:

a. Use the best FEV1 before transplantation;
b. Record therapy, complications, and microbiology from before transplantation.

ii. For people with CF who had a transplant before the current follow-up year:

a. Record all available information.

Appendix 5 Explanation of terms / Abbreviations

ABPA: allergic bronchopulmonary aspergillosis is an allergic lung disease characterised by an excessive response to the mould *Aspergillus fumigatus.*
BMI: body mass index, weight (kg) / [height (m)]^2.
Bronchodilator: medication that relaxes the muscles of the airways, used also for asthma.
CFRD: CF related diabetes.
CFTR: CF transmembrane conductance regulator. This is a protein at the cell surface that controls the salt and water balance across a cell. The gene that causes CF is the blueprint for the CFTR protein. Everyone has two copies of the gene for CFTR, but to be born with CF both CFTR genes must be affected by a CF-causing variant.
CFTR modulator therapy: Medication designed to correct the malfunctioning CFTR protein: different variants cause different defects in the structure of the protein and its functionality and the different CFTR modulators either correct or potentiate CFTR assembly or function; they can also be combined to become more efficient. CFTR modulator therapies work specifically for certain variant classes and those currently available are effective only in people with those variants. Not all known variants respond to CFTR modulator therapy.
Compassionate use: is a treatment option that allows the use of an unauthorised medicine for people with CF who have no alternative treatment options and no access to clinical trials.
Complex allele: To have CF you need two CFTR variants, one on each allele in chromosome 7 (where the CFTR-gene is located). If there is a variant on each allele they are considered to be in trans; if both variants are on the same allele they are in cis. Sometimes three (or even more variants) are found. It could be two variants in cis (and they are often known to be combined, e.g.F508 del with another variant) and one variant in trans. If there are two or more variants on the same allele it may be referred to as a complex allele.
DIOS: distal intestinal obstruction syndrome is a condition that is unique to people with CF. In DIOS, the intestines are blocked by thickened stool caused by sticky mucus and other mechanisms; this leads to reduced stool flow through the intestines and abdominal pain and may require emergency treatment.
FEV_1: the Forced Expiratory Volume of air in the first second of a forced exhaled breath.
FEV_1%: the FEV_1 as a percentage of the average value for healthy people of the same age, height, and sex.
Haemoptysis: coughing up blood. This happens frequently in small amounts in CF, so the complication we asked for is major bleeding (major meaning when the volume of expectorate is more than 250 ml over the course of the day).
Homozygous: CF is caused by variants of the CFTR gene, one on each allele. One is inherited from the mother and one from the father. If both variants are the same, the person is said to be homozygous for this variant.
Heterozygous: CF is caused by variants of the CFTR gene, one on each allele. One is inherited from the mother and one from the father. If these two variants are different the person is considered to be heterozygous.
ICM: Intestinal current measurement is a method to diagnose or exclude CF in difficult situations (e.g. unclear relevance of CFTR variants). CF is caused by abnormalities in the mechanism that carries salt into and out of cells. With ICM, the rate of salt transport is measured in tissue samples taken from the person (rectal biopsy) and measured against reference values of a healthy population. ICM can be carried out at any age.
LCI: Lung clearance index, measured by multiple breath washout (MBW); this is a test that measures non-homogeneity of lung ventilation. A tracer gas is inhaled, and the time to exhale a defined proportion of the gas is determined. MBW is very sensitive and particularly useful to measure lung function in children and people with milder forms of CF.
Macrolides: a type of antibiotic with anti-inflammatory properties. Azithromycin is a macrolide often used in people with CF who have chronic Pseudomonas aeruginosa lung infection.
Meconium ileus: small-bowel obstruction caused by unusual thick, sticky faeces (i.e. meconium, which is the first stool of newborn babies).
NaCl: sodium chloride. Here: inhaled hypertonic saline.
NIPPV: Non-invasive positive pressure ventilation; this refers to mechanical ventilation that helps people with CF with breathing difficulties. It is done with the help of a face mask and does not require the insertion of an artificial airway (tube). It can be one of two types: BiPaP (Bi-level positive air pressure) or CpaP (continuous positive air pressure).
NPD: Nasal Potential Difference; this is a method to diagnose or exclude CF in unclear cases and involves placing an electrode on the surface of the inside of the nose to measure the electrical potential difference across the nasal

epithelium. The NPD is a result of the transport of ions such as sodium and chloride in and out of the cells, a mechanism that is affected by defects in the CFTR protein.
Pancreatic insufficiency: the absence of pancreatic enzymes in the gut leading to malnutrition if not treated (in the ECFSPR pancreatic insufficiency is therefore defined as the use of pancreatic enzyme supplementation).
Pneumothorax: collapsed lung. In CF usually because of severe lung damage.
PPI: Proton Pump Inhibitors (medication that reduces the level of stomach acids).
pwCF: People with Cystic Fibrosis
rhDNase: recombinant human DNase (marketed as Pulmozyme®).
Steroids: are a group of medicines with strong anti-inflammatory properties. The type that are prescribed to people with CF are corticosteroids or glucocorticoids.
Z-score (or standardised scores): a way to compare results with a "normal" population, the reference population. Negative z-scores mean that the value is below the mean of values in the reference population, whereas positive z-scores mean that the value is above the mean. Z-score equal to 0 means that the value is equal to the mean of values in the reference population. For example, a z-score for weight of-2 means that the weight is 2 standard deviations below the mean of subjects of the same age and sex of the reference population. For example, if the z-score for BMI of a 10-year-old boy is-2, it means that the BMI for that boy is 2 standard deviations below the mean BMI of 10-year-old boys of the reference population.

Appendix 6 Country Codes

AL	Albania
AM	Armenia
AT	Austria
BE	Belgium
BG	Bulgaria
BY	Belarus
CH	Switzerland
CY	Cyprus
CZ	Czech Republic
DE	Germany
DK	Denmark
EE	Estonia
ES	Spain
FI	Finland
FR	France
GE	Georgia
GR	Greece
HR	Croatia
HU	Hungary
IE	Ireland
IL	Israel
IS	Iceland
IT	Italy
KZ	Kazakhstan
LT	Lithuania
LU	Luxembourg
LV	Latvia
MD	Republic of Moldova
ME	Montenegro
MK	North Macedonia
NL	The Netherlands
NO	Norway
PL	Poland
PT	Portugal
RO	Romania
RS	Serbia
RU	Russian Federation
SE	Sweden
SI	Slovenia
SK	Slovak Republic
TR	Türkiye
UA	Ukraine
UK	United Kingdom of Great Britain and Northern Ireland

Reference: www.iso.org/iso-3166-country-codes.html