ECFSPR

2022
Annual Data Report

Table of contents

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Message from the ECFSPR Director

We are delighted to present the 2022 Annual Report from the European Cystic Fibrosis Society Patient Registry (ECFSPR).

This report features longitudinal data analyses of several critical variables such as lung function, microbiology and drug utilisation in Europe. To make the report easier to read the data tables are included in Appendix 1. This year we have also made a Highlights Report that summarises the main results, and a slide deck containing graphs of the most significant data from 2022 which is available for use in presentations and in the public domain.

The report indicates that more countries than ever before now have access to highly effective CFTR modulators; the health of people with cystic fibrosis (CF) in Europe has improved in most countries since their introduction. There are, however, socioeconomic disparities and other differences that influence these outcomes. The increased availability of CFTR modulators observed in 2022 fuels the hope of the CF community that these highly effective medications will become universally available.

One of our goals is to increase our geographical coverage and eventually include all 50 countries defined as European by the World Health Organisation (WHO). In this report there is data from 39 countries and 54,546 consenting individuals with CF. The epidemiological data comes from national CF registries and individual CF centres across Europe and neighbouring countries; the ECFSPR collaborates closely with these centres and registries to ensure that the data is as complete and high-quality as possible.

The ECFSPR serves as an invaluable resource for the CF community, facilitating quality improvement initiatives and research. We offer essential population-level data that is easily accessible to people with CF, clinicians, and researchers. The use of registry data helps us comprehend changes in population demographics and predict clinical outcomes. Additionally, the ECFSPR enables research on questions that may not be feasible to address through controlled trials.

Data quality continues to be a primary focus of the ECFSPR, and not just so that we can provide a dependable and comprehensive overview of clinical outcomes in CF across Europe, but also because the CF community is now represented in registry-based pharmacovigilance (PMV) studies through this data. These PMV studies provide crucial real-world evidence about the safety and efficacy of new drugs introduced for people with CF.

Our ongoing collaboration with CF Europe and national patient organisations ensures that registry data is used to benefit the community. We are grateful to all the people with CF and their families throughout Europe and beyond who participate in the European CF Patient Registry. I extend my thanks to the ECFSPR staff, the Executive and Scientific Committees, the volunteers who contribute to our working groups and various projects, and the sponsors for their financial 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.

This year we have also published a Highlights report containing key information from the Annual Report, specifically for the people with CF and their families and anyone wishing to know a little more about the disease. You will see that this report is different in content and style from the At-a-Glance reports which were published in previous years.

News, updates, and other interesting information are regularly posted on social media. You can find us on Facebook, Instagram, LinkedIn and X.

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 ecfs-pr@uzleuven.be.

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.

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 is collected using a common set of variables and definitions and is 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 is pseudonymised, and only year/ month of birth and randomised centre and patient codes are used as identifiers.

Data is available for scientific purposes on application. All requests 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 request; this decision is final. Requests 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. Where major discrepancies between the definitions occur those variables have been omitted from the annual report for that country; in the case of minor discrepancies an explanatory footnote has been added to the graphs and tables. For example, the ECFSPR collects information on chronic Pseudomonas aeruginosa infection according to the modified Leeds criteria and/or the presence of elevated anti-Pseudomonas antibodies (see Appendix 4). If a national registry collects such information as "at least one positive Pseudomonas aeruginosa culture this year", this information would be too different from the ECFSPR definition of chronic Pseudomonas aeruginosa, and we would set this variable to "missing" for that particular country. If a country defines chronic Pseudomonas aeruginosa as "the presence of more than four positive cultures in 6 months", the data of this variable would be included in the annual report since the definition is much closer to the ECFSPR definition and a footnote would be 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; all of the data, however, is 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 (Appendix 1). These differences can influence how the data is 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 28 February 2024.

Summary of data report

Outcome		Females		Males		Total	
PwCF registered in the ECFSPR	n (%)	25964	(47.6)	28582	(52.4)	54546	(100)
Age at follow-up (years) (PwCF alive on 31/12/2022)	median (25th pctl-75th pctl)	19.4	(10.2-31.8)	20.5	(10.5-33.3)	20.0	(10.4-32.5)
PwCF ≥ 18 years (PwCF alive on 31/12/2022)	n (%)	13787	(53.4)	15757	(55.4)	29544	(54.5)
Age at diagnosis (months)*	median (25th pctl-75th pctl)	3.6	(1.2-31.3)	3.6	(1.2-31.0)	3.6	(1.2-31.2)
PwCF with at least one F508del allele recorded*	n (%)	19483	(80.3)	21226	(80.3)	40749	(80.3)
PwCF living with lung transplant**	n (%)	1285	(5.3)	1262	(4.8)	2547	(5.0)
PwCF living with liver transplant**	n (%)	103	(0.4)	224	(0.9)	327	(0.6)
PwCF deceased in 2022***	n (%)	150	(0.6)	159	(0.6)	309	(0.6)
Age at death (years)***	median (25th pctl-75th pctl)	30.5	(21.0-42.0)	36.0	(25.0-46.0)	33.0	(23.0-45.0)

*   Only people with CF seen during the year by clinical staff. The total number presented is 51,022.

**   Only people with CF alive at 31/12/2022. The total number of the CF population presented is 50,744.

***   Only people with CF seen during the year. For the United Kingdom, all individuals with a confirmed diagnosis of CF were included (N=11,148). The total number presented is 51,919.

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

Data report

1. Demographics

The ECFSPR has continuously increased its coverage over the recent years. Only a few countries in Europe have not yet contributed data to the ECFSPR and we are in contact with the remaining countries to welcome them into the European registry. In some countries not everyone with CF has had the opportunity to join the ECFSPR. We invite all CF centres to participate in the registry and we are confident that over the next few years several more centres will join us. The National Coordinators that have been appointed by their country are involved in this process, encouraging their centres to become a new ECFSPR member.

National registries, as well as countries with centres that enter their data directly to our data collection software, called ECFSTracker, contribute to the ECFSPR. This chapter gives information on coverage (i.e. the proportion of the estimated total number of PwCF included in the registry) 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 2022.

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

The countries that contributed 2022 data are in turquoise.

1. Demographics

Table 1.1   Number of people with CF in 2022, by country.

Country	People with CF registered, not lost to follow-up	People with CF seen	Estimated coverage
Albania	103	69	>80%
Armenia	26	25	>70%
Austria	888	862	>90%
Belarus*	152	152	68%
Bulgaria	220	212	91%
Croatia**	150	145	>98%
Cyprus	34	24	>80%
Czech Republic*	695	673	99%
Denmark*	570	545	99%
Finland	65	64	64%
France*	7288	7288	>90%
Georgia	86	43	>80%
Germany*	7254	6980	80%
Greece*	646	597	80%
Hungary*	550	547	98%
Iceland	15	15	>90%
Ireland*	1378	1345	89%
Israel**	564	516	>95%
Italy*	6083	6077	98%
Latvia	47	46	90%
Lithuania	46	43	>55%
Luxembourg	26	26	60%
Republic of Moldova	56	48	>90%
The Netherlands*	1634	1622	95%
North Macedonia	142	125	>90%
Norway*	366	362	>90%
Poland	1620	1524	91%
Portugal**	393	383	>95%
Romania	315	287	45%
Russian Federation*	3960	2404	88%
Serbia	223	204	>90%
Slovak Republic**	308	280	>90%
Slovenia	121	118	>95%
Spain	2578	2476	85%
Sweden*	780	737	>95%
Switzerland**	1054	1026	>99%
Turkey	2554	2537	>60%
Ukraine	408	344	41-70%
United Kingdom*	11148	10251	99%
Total	54546	51022	

*   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.

The column "People with CF registered, not lost to follow-up" displays the individuals with CF that attended centres and 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 have one individual centre that includes almost all people with CF, such as Latvia and Serbia.

1. Demographics

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

Number of people with CF registered in the ECFSPR in 2022.
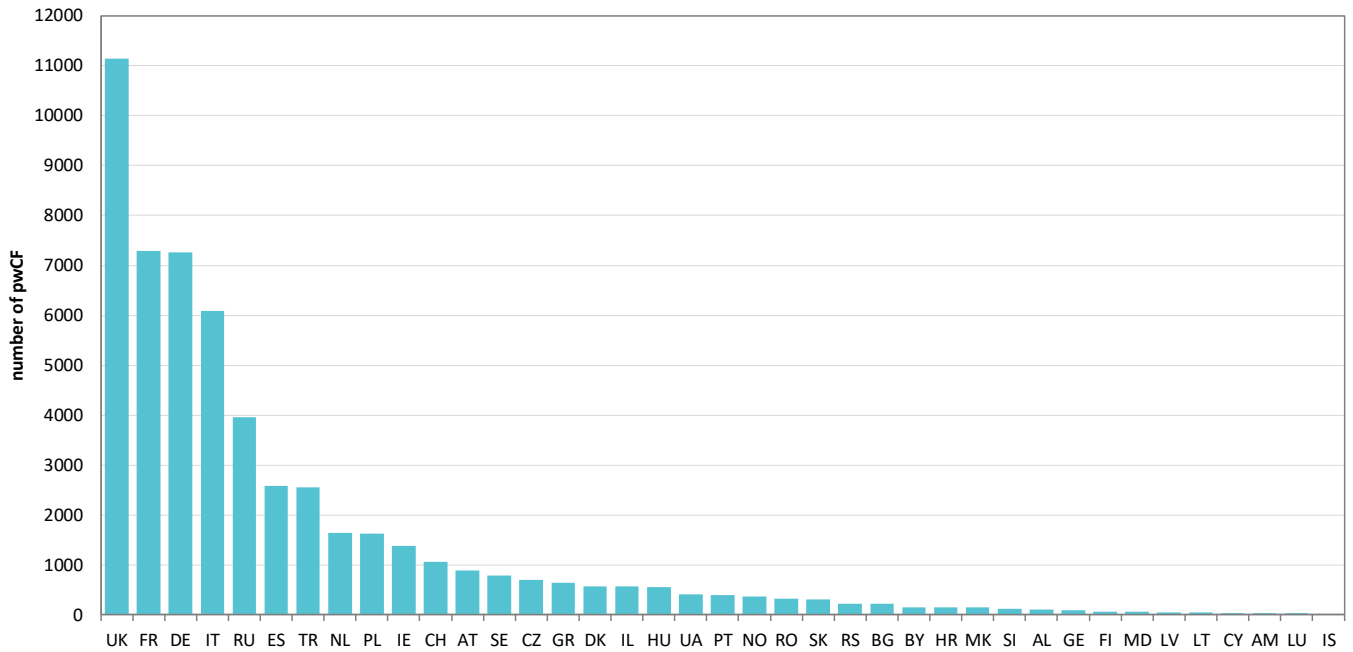
Each vertical bar shows the number of registered pwCF (excluding lost to follow-up) living in that country in 2022. Please refer to table 1.1 for the coverage in each country.

1. Demographics

Figure 1.3   The number of countries and people with CF in the ECFSPR has risen continuously over the years.

Number   of   people   with   CF   and   number   of   countries   from   2008   to   2022.
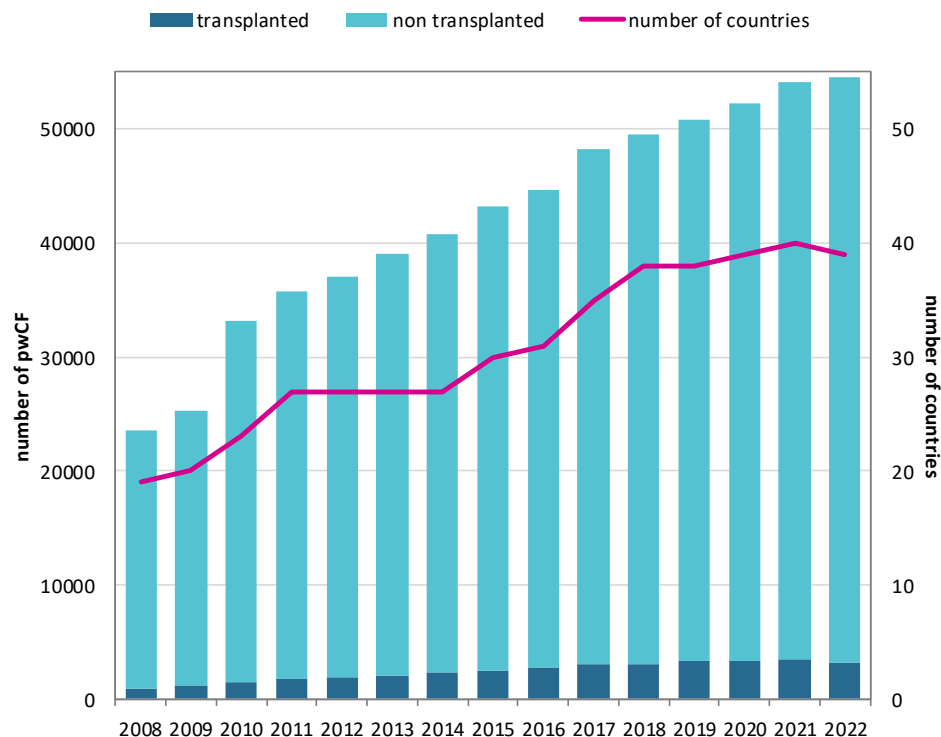
Figure 1.3 presents data over time using cross sectional data per year of people with a confirmed CF diagnosis. All people with CF alive, deceased, or not seen but alive during the year of follow-up were included. When computing the yearly prevalence we excluded people with CF with missing values and people with CF who were lost to follow-up.

1. Demographics

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

Distribution of age at follow-up. People with CF alive on 31/12/2022.
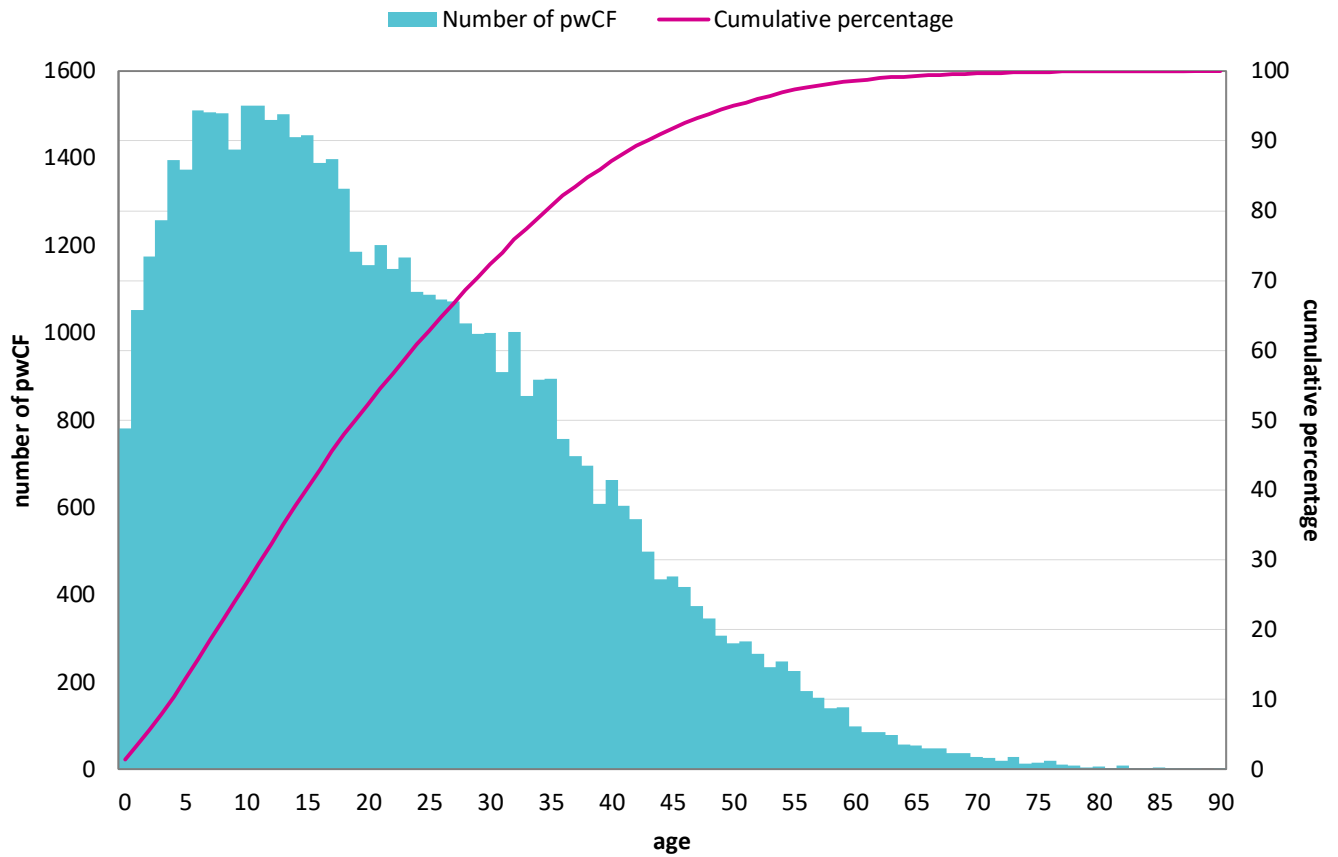
Each vertical bar shows the number of people with CF of that age alive in 2022. The cumulative percentage (the pink line) describes how many people with CF, as a percentage, are below a certain age. For example, 50% are younger than 19 years old.

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.
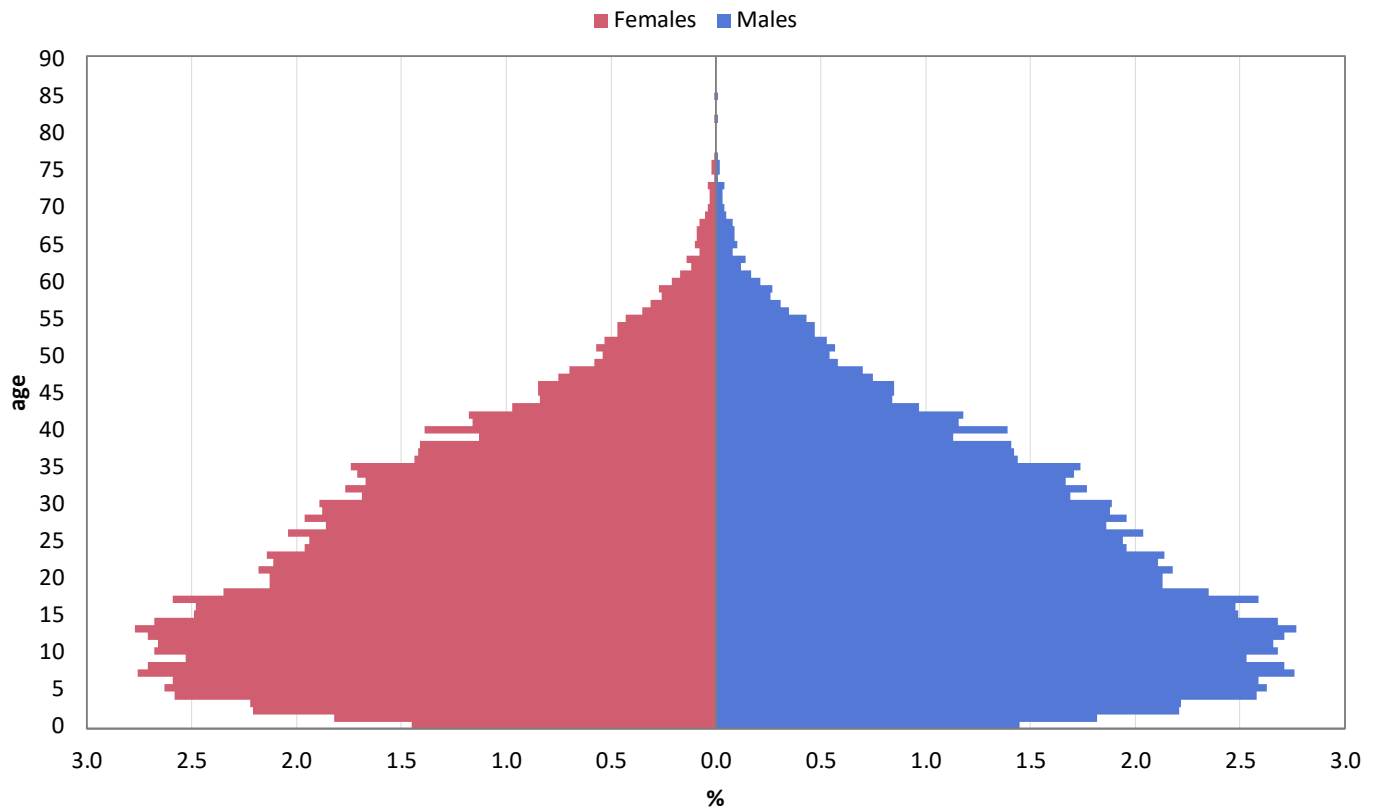
The pyramid shows the percentage of people with CF of different ages as horizontal bars. The left side of the pyramid (red) shows, how many females with CF, as a percentage, are of a certain age, and the right side (blue) shows the same for males. The lower percentage of children with CF at the bottom of the pyramid is a result of the fact that some children are not diagnosed early in life. In 2022 the mean age at diagnosis was 0.8 years (see table A 2.1, Appendix 1).

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. People with CF alive on 31/12/2022 (table A1.1, Appendix 1).
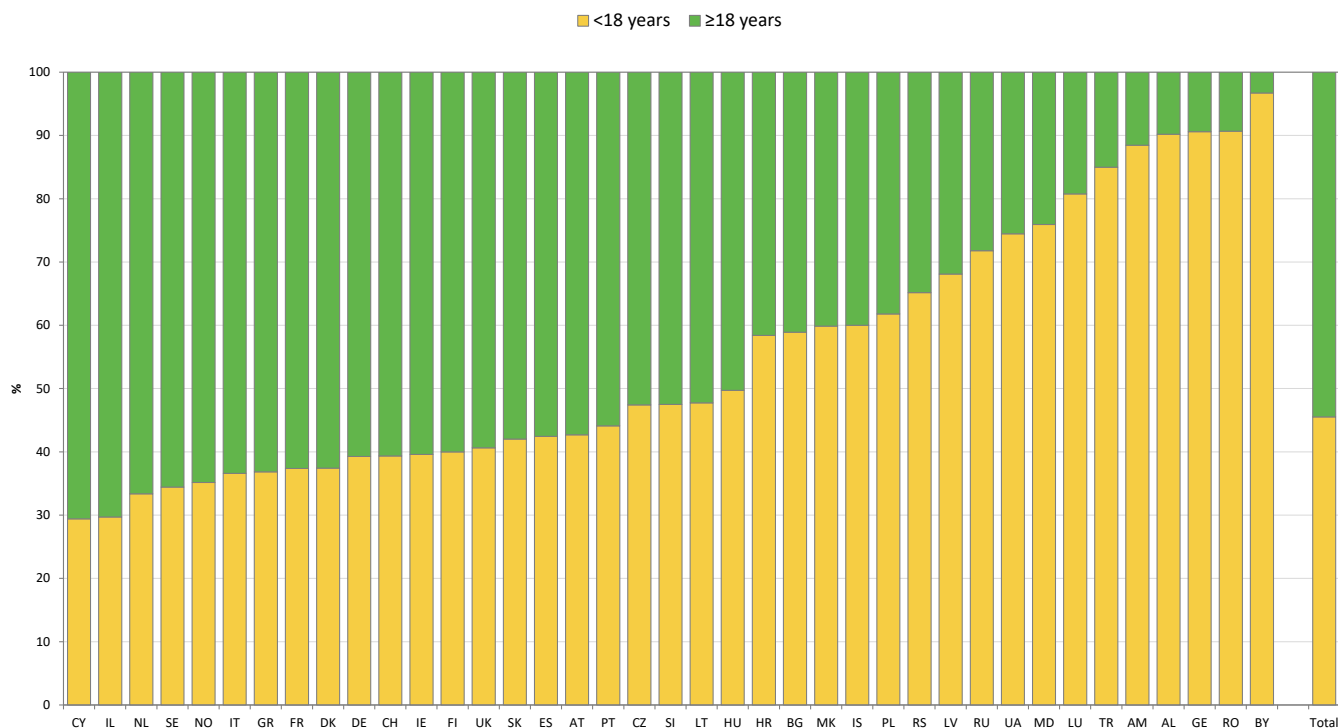
The yellow vertical bar shows the percentage of children and adolescents with CF living in that country in 2022, the green vertical bar shows the percentage of adults. Overall (see "Total") in the ECFSPR there are more adults than children.

Figure 1.7   In recent years the proportion of adults with CF in Europe has risen significantly; as of 2022, adults made up more than >50% of the total.

Number of people with CF and percentage of adults and children from 2008 to 2022.
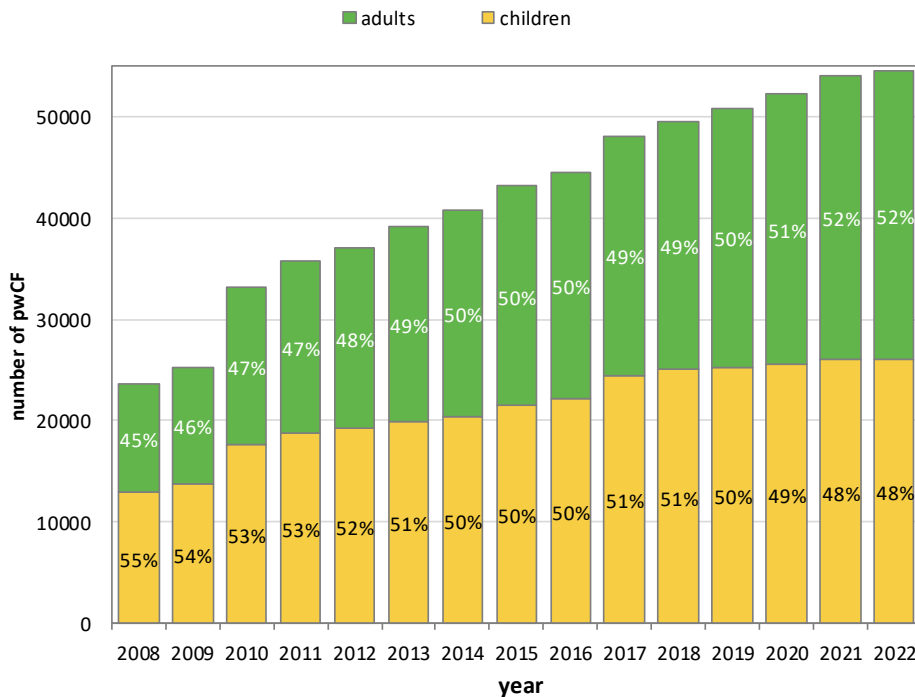
This graph presents data over time using cross sectional data per year of people with a confirmed CF diagnosis. All people with CF alive, deceased, or not seen but alive during the year of follow-up were included. When computing the yearly prevalence we excluded people with CF with missing values and people with CF who were lost to follow-up.

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. People with CF alive on 31/12/2022 (table A1.2, Appendix 1).
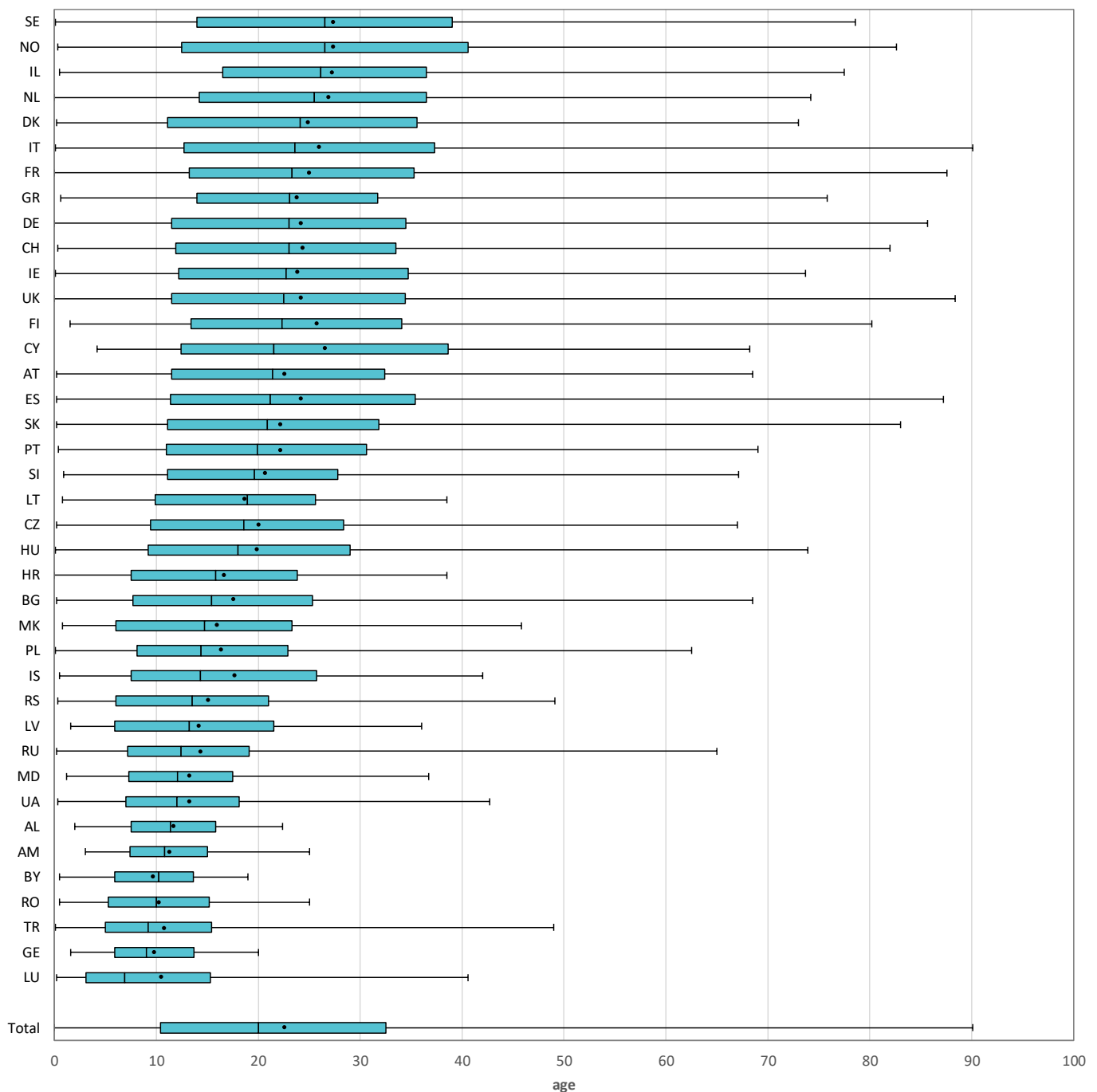
This box plot is a graphic representation of the age at follow-up detailed in table A1.2, Appendix 1. For each country the vertical borders of the box are the first and third quartile, the dash (vertical black line crossing the box) is the median, the black dot is the mean and the whiskers (lines with a T-shaped end) are the minimum and the maximum.

1. Demographics

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

Sex distribution, by country and overall. People with CF alive on 31/12/2022.
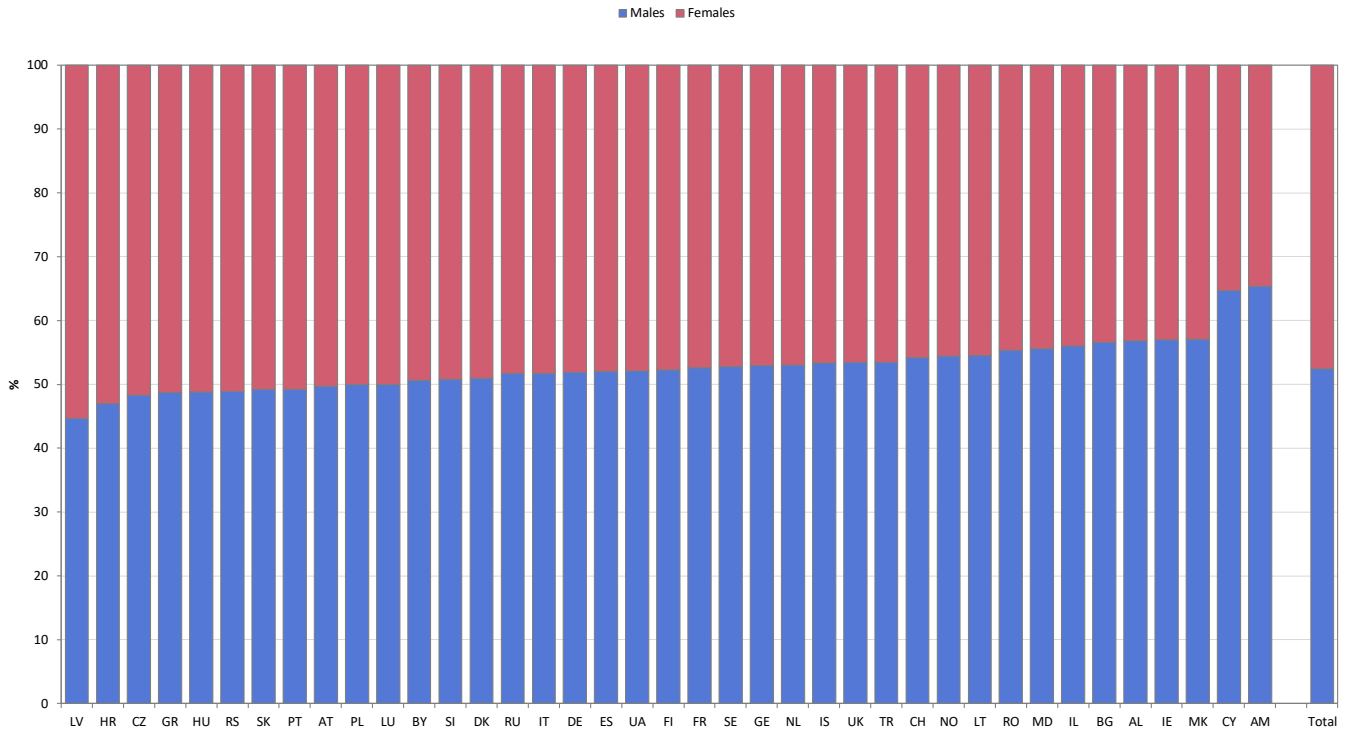
Sex distribution of the CF population. Overall (see "Total") in the ECFSPR there are slightly more males than females with CF.

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 presence or absence 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 tables below.

In this chapter and the following ones, only the data for people with CF seen during the year is 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) seen in 2022 (table A2.1, Appendix 1).
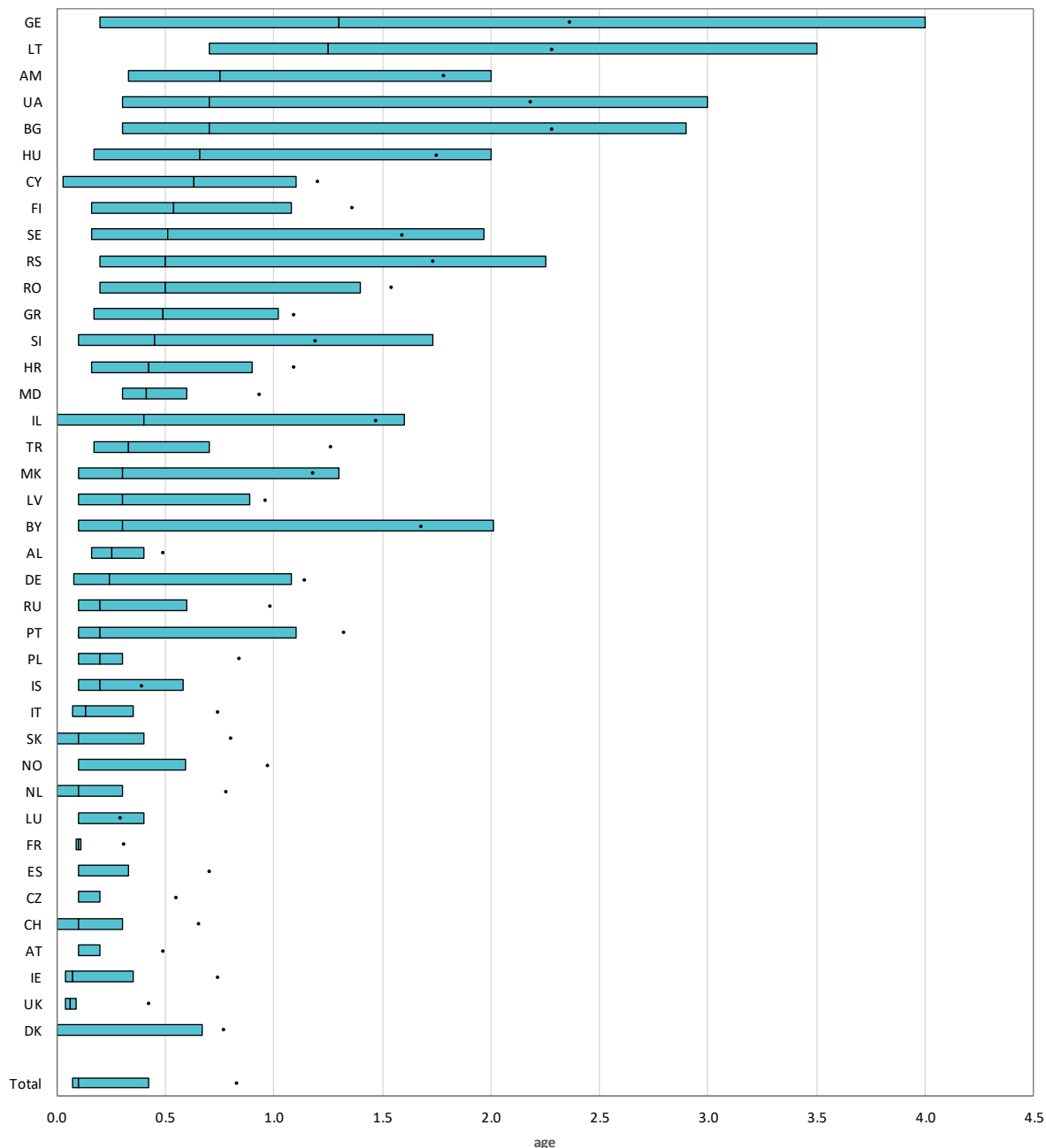
Note: For Cyprus, Greece, Lithuania and the Slovak Republic the information on age at diagnosis is missing for more than 10% of the children.

This boxplot is a graphic representation of the age at diagnosis detailed in table A2.1 (Appendix 1). For each country the vertical borders of the box are the first and third quartiles, the dash (vertical black line crossing the 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 really high for some countries and this would have shrunk the boxes at the left side 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) seen in 2022 (table A2.2, Appendix 1).
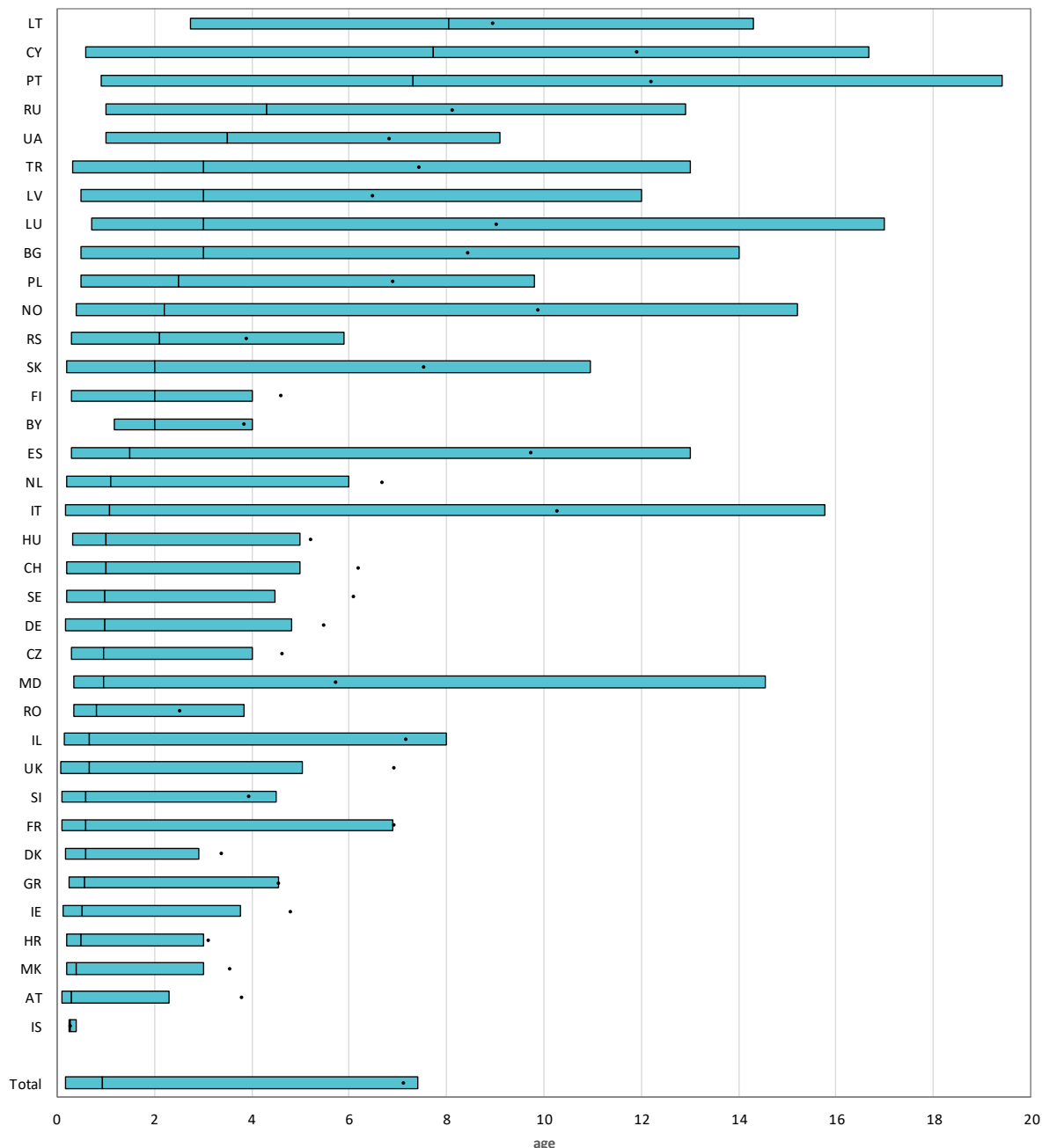
Note: Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the table, but the people are included in the total number.

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

This boxplot is a graphic representation of the age at diagnosis detailed in table A2.2 (Appendix 1). For each country the vertical borders of the box are the first and third quartiles, the dash (vertical black line crossing the 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 really high for some countries and this would have shrunk the boxes at the left side of the graph.

2. Diagnosis

Figure 2.3   With 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 2022.
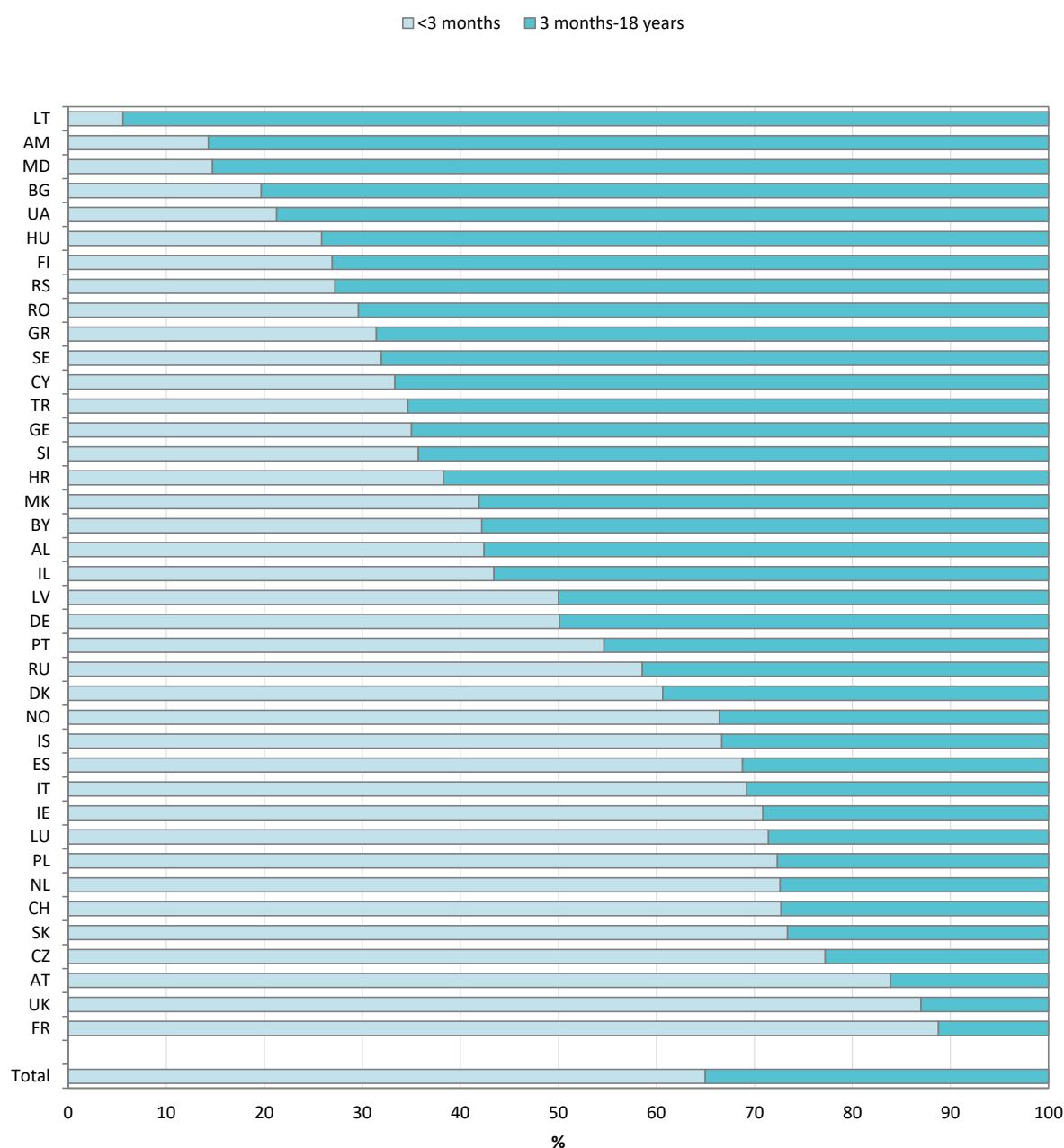
Note: For Cyprus, Greece, Lithuania and the Slovak Republic the information on age at diagnosis is missing for more than 10% of the children.

This graph shows the percentage of children and adolescents with CF by age at diagnosis. Light turquoise represents individuals diagnosed at younger than 3 months, turquoise shows those diagnosed between 3 months and 18 years. The bars sum to 100%.

2. Diagnosis

Figure 2.4   Age at diagnosis has shifted to the first 3 months of life in many countries, due to early recognition of symptoms or longer running neonatal screening programmes.

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 2022.
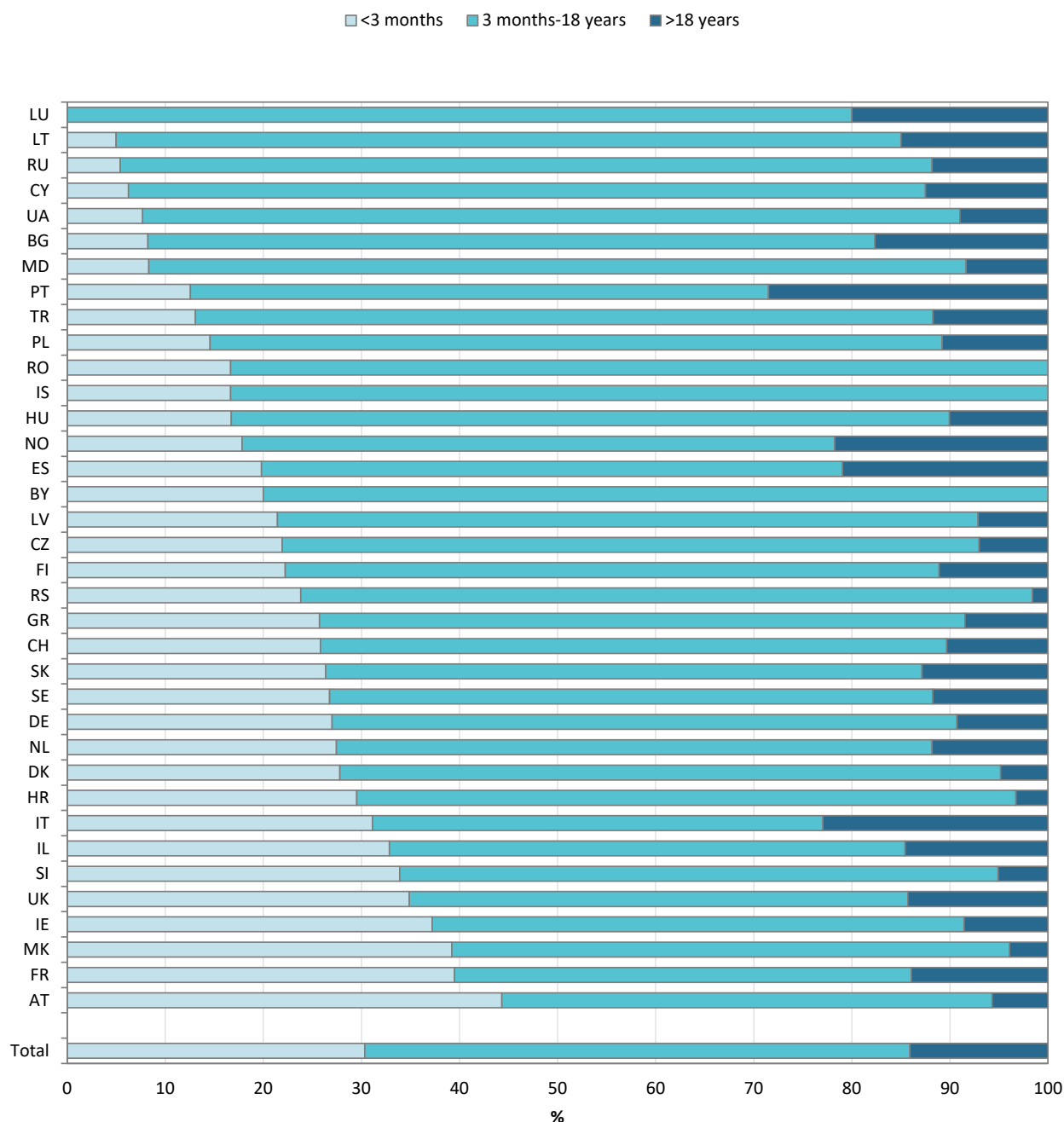
Note: Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the table, but the people are included in the total number.

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

This graph shows the percentage of adults with CF by age at diagnosis. Light turquoise represents individuals diagnosed at younger than 3 months, turquoise shows those diagnosed between 3 months and 18 years, and the dark blue represents individuals diagnosed at older than 18 years. The bars sum to 100%.

2. Diagnosis

Figure 2.5   *The proportion of young children with CF diagnosed through newborn screening has increased in many countries over the years.*

Proportion of children with CF who underwent neonatal screening, by country and overall. Children 5 years old or younger seen in 2022.
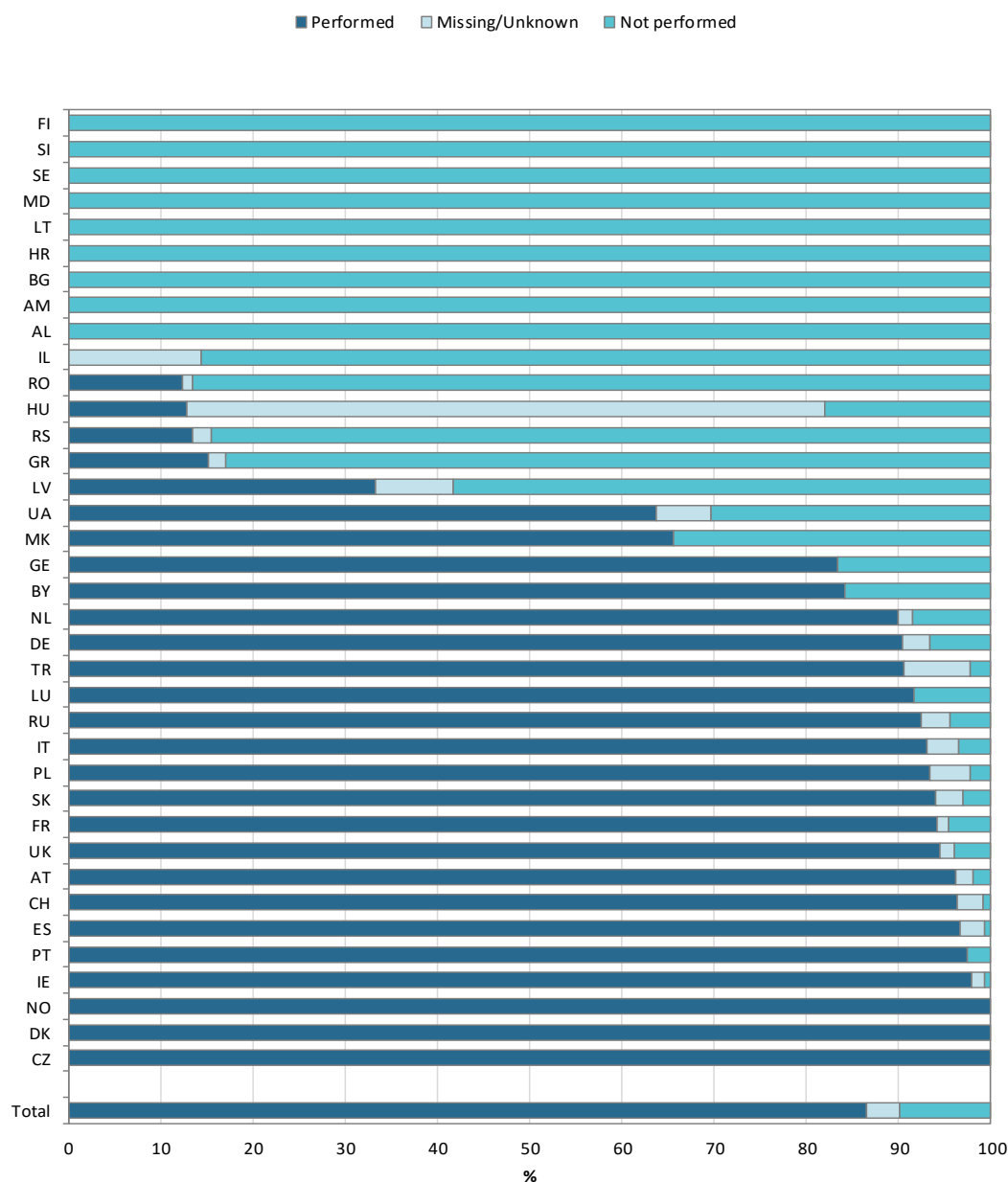
Note: Cyprus and Iceland have <5 children 5 years old or younger seen in 2022 and are excluded from the graph.

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

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

This graph shows the percentage of children with CF aged 5 years or younger in 2022 who were screened at birth. Dark blue represents neonatal screening "done" and turquoise "not done". This graph shows that, in the five years before 2022, in many countries people with CF underwent newborn screening, and that in some countries there is no neonatal screening programme. In total, 87% of all children 5 years old or younger registered in the ECFSPR in 2022 were screened at birth.

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.
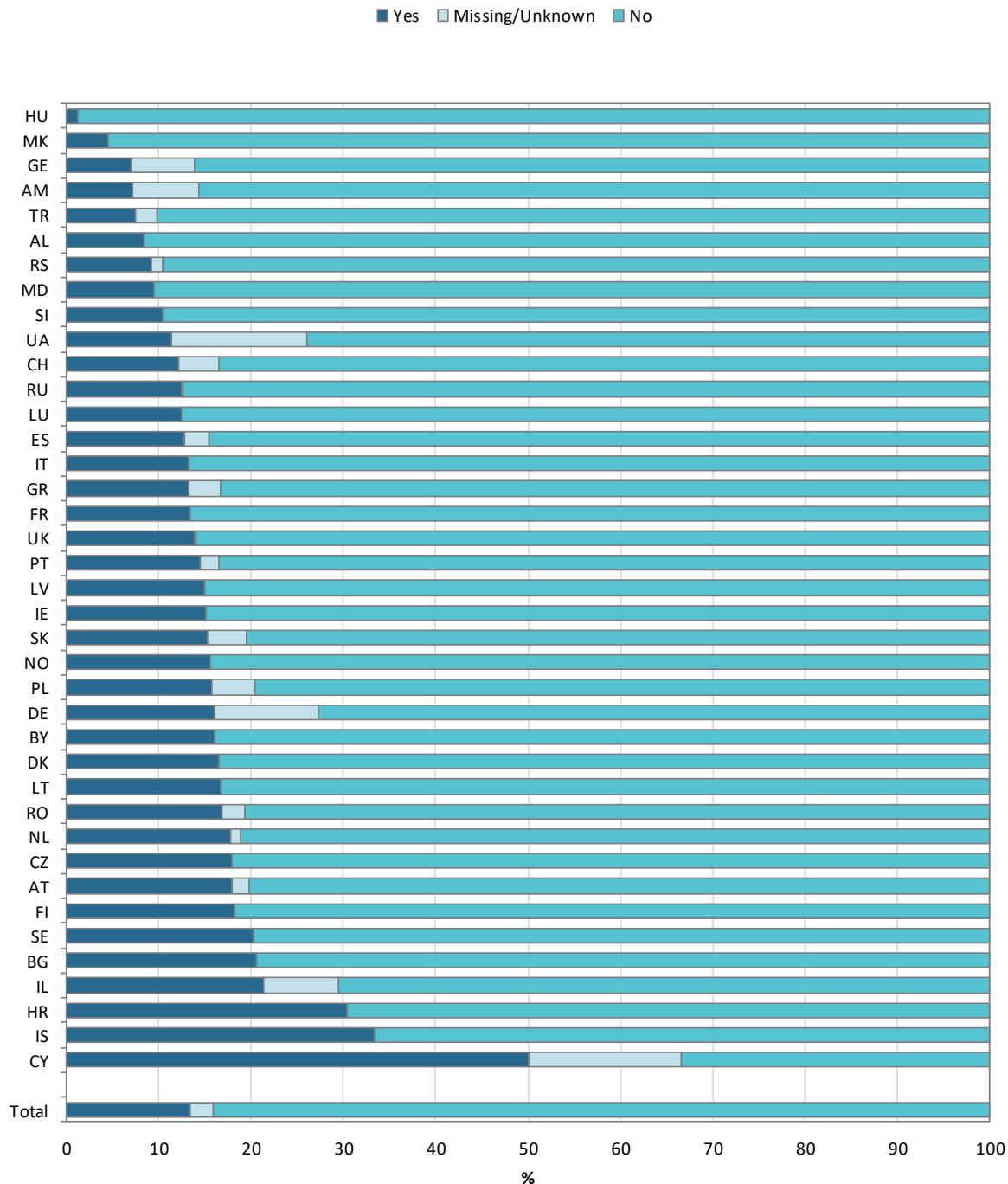
This graph shows the percentage of children with CF aged 10 years or younger in 2022 who had meconium ileus at birth. Dark blue represents meconium ileus "yes", turquoise is "no".

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 2012 to 2022.
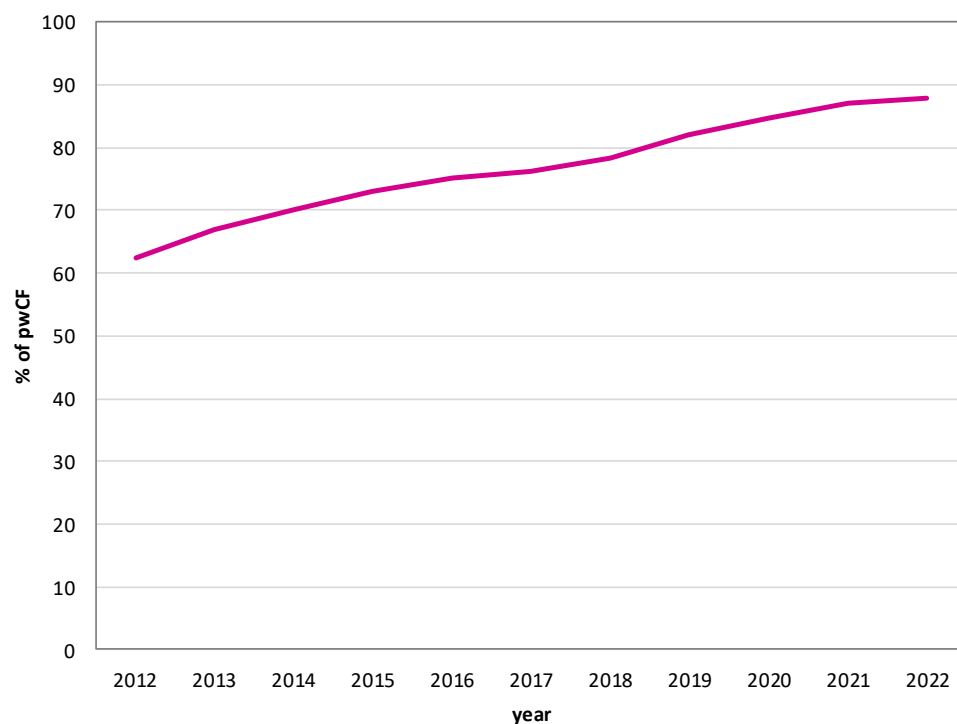
In this graph data over time is presented using cross sectional data per year of children with a confirmed CF diagnosis. Children with CF who are alive, deceased, or not seen during the year of follow-up were included and those who were lost to follow-up and/or transplanted (lung and/or liver) were excluded. When computing the yearly prevalence, where the information was missing for a child it was excluded from the total number.

3. Genetics

Cystic fibrosis is caused by pathogenic variants of the *CFTR* gene. At least one variant on each copy or allele of the gene is inherited from the mother and from 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 was not present in this database, the name of the variant can be 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 and 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.

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

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.

3. Genetics

Table 3.1   Proportion of people with CF with DNA analysis and the result of this, by country and overall. All people with CF seen in 2022.

Country	Genotyping				Among genotyping done			
	Not done		Done		At least one variant unknown		Two variants identified	
	Number	%	Number	%	Number	%	Number	%
Albania	5	7.25	64	92.75	3	4.69	61	95.31
Armenia	0	0	25	100	4	16.00	21	84.00
Austria	1	0.12	861	99.88	45	5.23	816	94.77
Belarus	0	0	152	100	46	30.26	106	69.74
Bulgaria	0	0	212	100	12	5.66	200	94.34
Croatia	0	0	145	100	4	2.76	141	97.24
Cyprus	0	0	24	100	1	4.17	23	95.83
Czech Republic	1	0.15	672	99.85	7	1.04	665	98.96
Denmark	0	0	545	100	0	0	545	100
Finland	2	3.13	62	96.88	2	3.23	60	96.77
France	0	0	7288	100	128	1.76	7160	98.24
Georgia	5	11.63	38	88.37	2	5.26	36	94.74
Germany	15	0.21	6965	99.79	180	2.58	6785	97.42
Greece	2	0.34	595	99.66	22	3.70	573	96.30
Hungary	0	0	547	100	104	19.01	443	80.99
Iceland	0	0	15	100	0	0	15	100
Ireland	1	0.07	1344	99.93	31	2.31	1313	97.69
Israel	2	0.39	514	99.61	35	6.81	479	93.19
Italy	1	0.02	6076	99.98	152	2.50	5924	97.50
Latvia	0	0	46	100	0	0	46	100
Lithuania	0	0	43	100	2	4.65	41	95.35
Luxembourg	0	0	26	100	0	0	26	100
Rep of Moldova	0	0	48	100	6	12.50	42	87.50
The Netherlands	9	0.55	1613	99.45	15	0.93	1598	99.07
North Macedonia	1	0.80	124	99.20	0	0	124	100
Norway	0	0	362	100	3	0.83	359	99.17
Poland	1	0.07	1523	99.93	53	3.48	1470	96.52
Portugal	0	0	383	100	3	0.78	380	99.22
Romania	1	0.35	286	99.65	23	8.04	263	91.96
Russian Federation	77	3.20	2327	96.80	282	12.12	2045	87.88
Serbia	3	1.47	201	98.53	12	5.97	189	94.03
Slovak Republic	0	0	280	100	28	10.00	252	90.00
Slovenia	1	0.85	117	99.15	3	2.56	114	97.44
Spain	1	0.04	2475	99.96	124	5.01	2351	94.99
Sweden	0	0	737	100	1	0.14	736	99.86
Switzerland	5	0.49	1021	99.51	35	3.43	986	96.57
Turkey	105	4.14	2432	95.86	362	14.88	2070	85.12
Ukraine	3	0.87	341	99.13	72	21.11	269	78.89
United Kingdom	24	0.23	10227	99.77	262	2.56	9965	97.44
Total	266	0.52	50756	99.48	2064	4.07	48692	95.93

3. Genetics

Figure 3.1   Most of the people with CF in Europe have two CFTR variants identified.

Proportion of variants identified and not identified, by country and overall. Only people with CF for whom DNA analysis has been done.
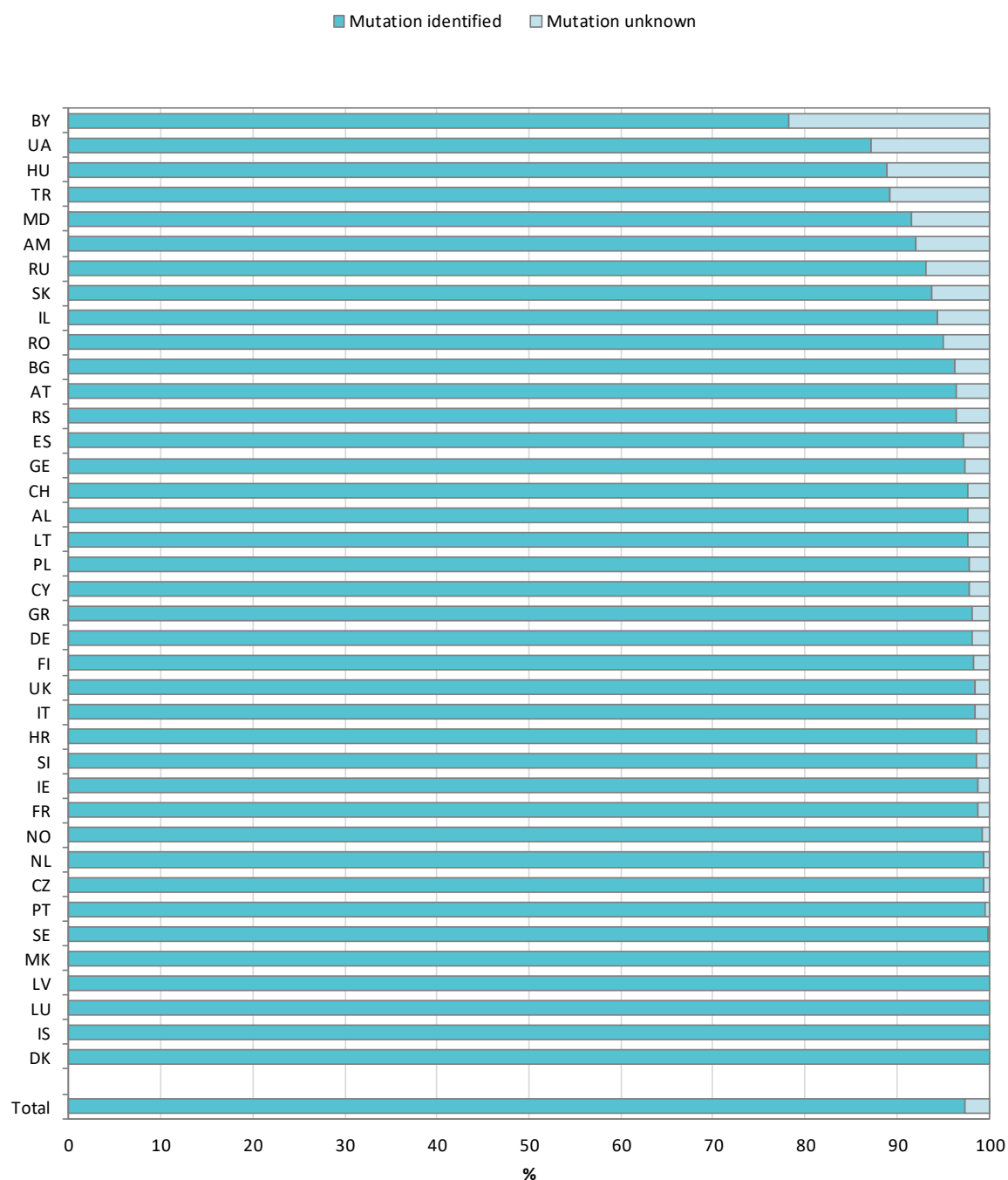
This graph shows the percentage of variants identified (dark turquoise) and variants not identified (light turquoise) through DNA analysis by country and overall. The number of variants not identified on one of the 2 alleles varies greatly from country to country. This is partly due to the different approaches to DNA testing. Overall, more than 2.6% of variants remain unidentified after DNA analysis, leaving 4.1% 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 F508del homozygous and heterozygous people with CF, by country and overall. All people with CF seen in 2022.
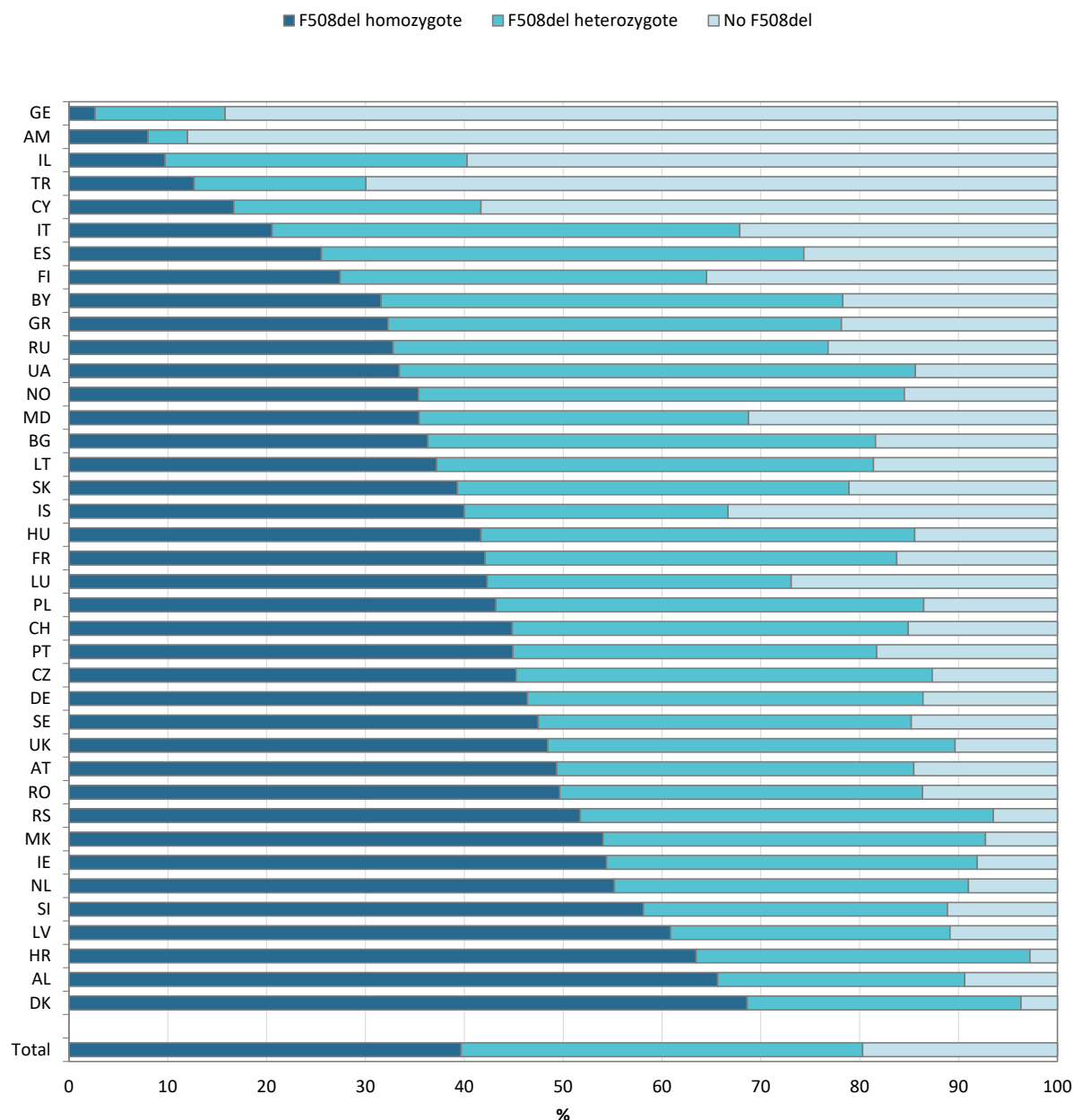
F508del is the name of the most commonly occurring CFTR variant in the world. People with CF who carry two F508del variants are often described as having "classic CF", but other variant combinations can cause the same degree of disease. We have grouped the people with CF into F508del homozygous (with two F508del variants), F508del heterozygous (with one F508del variant and another that is not F508del), and people with CF who do not haveF508del. 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 the 17 most common 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	60705	59.87	Denmark (82.5%)
G542X	2794	2.76	Armenia (8.0%)
N1303K	2213	2.18	Iceland (40.0%)
G551D	1266	1.25	Ireland (8.3%)
2789+5G->A	1131	1.12	Turkey (3.1%)
W1282X	1096	1.08	Israel (23.1%)
3849+10kbC->T	1024	1.01	Lithuania (8.1%)
R117H	1009	1.00	Ireland (3.1%)
CFTRdele2,3	985	0.97	Belarus (9.5%)
1717-1G->A	853	0.84	Switzerland (2.7%)
R553X	822	0.81	Lithuania (5.8%)
D1152H	733	0.72	Israel (5.5%)
2183AA->G	725	0.72	Armenia (10.0%)
621+1G->T	688	0.68	Greece (6.9%)
R347P	613	0.60	Luxembourg (5.8%)
G85E	575	0.57	Israel (2.4%)
1677delTA	527	0.52	Georgia (46.1%)

This table presents the 17 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   Geographical distribution of the F508del variant.
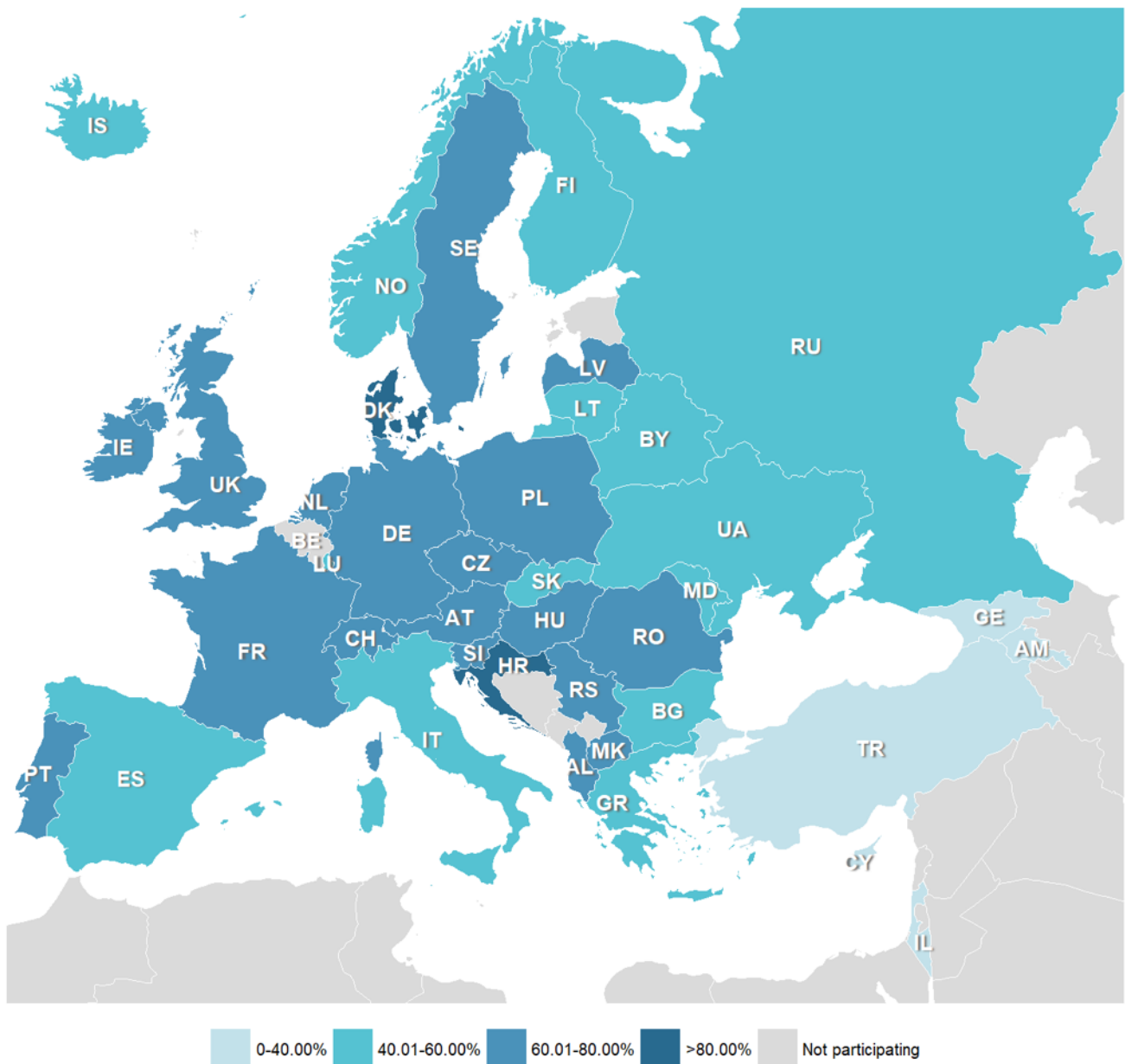
F508del is the most common variant in all countries; the highest frequency occurs in Denmark (82.5%) and Croatia (80.3%).

3. Genetics

Figure 3.4   Geographical distribution of the G542X variant.
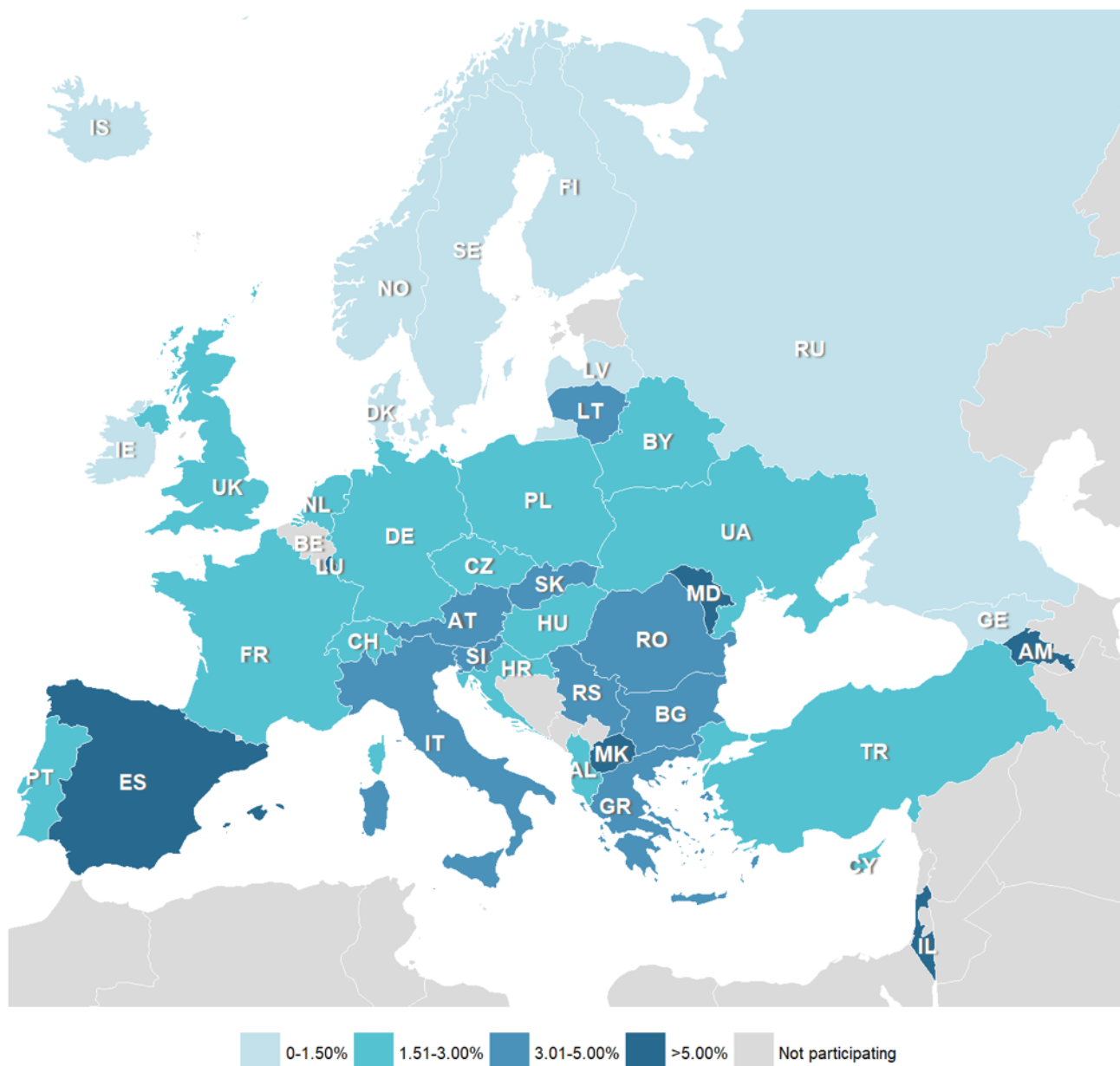
The *G542X* variant is most frequent in Southern Europe, with the highest allele frequency in Armenia (8.0%), whereas it is very rarely found in Ireland, the Scandinavian countries and Russia.

3. Genetics

Figure 3.5   Geographical distribution of the N1303K variant.
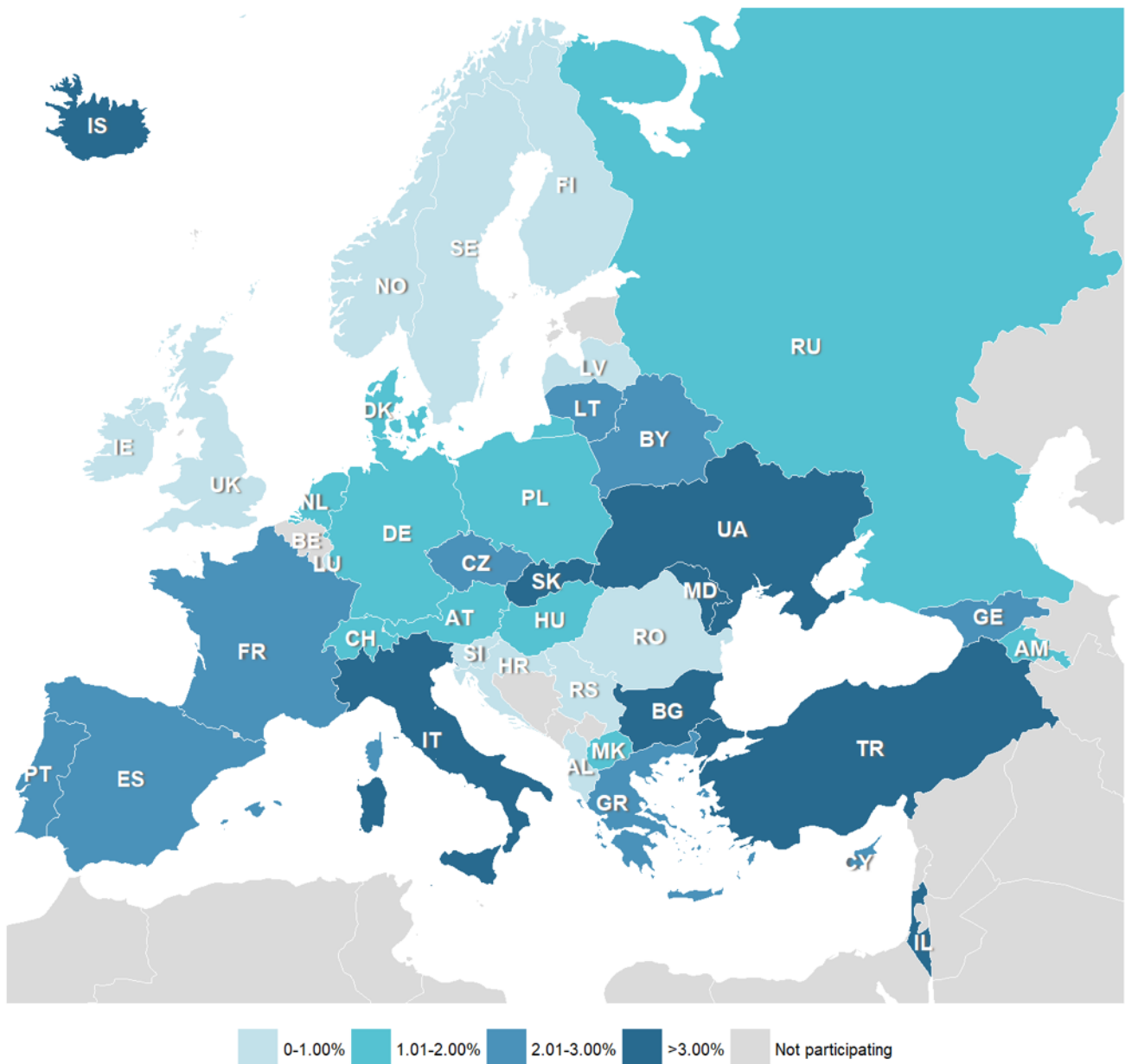
The *N1303K* variant is most frequent in Iceland (40.0%). This is an exception in Northern Europe where it is otherwise rare; it is much more frequent in the countries of Southern and Eastern Europe.

3. Genetics

Figure 3.6   Geographical distribution of the G551D variant.
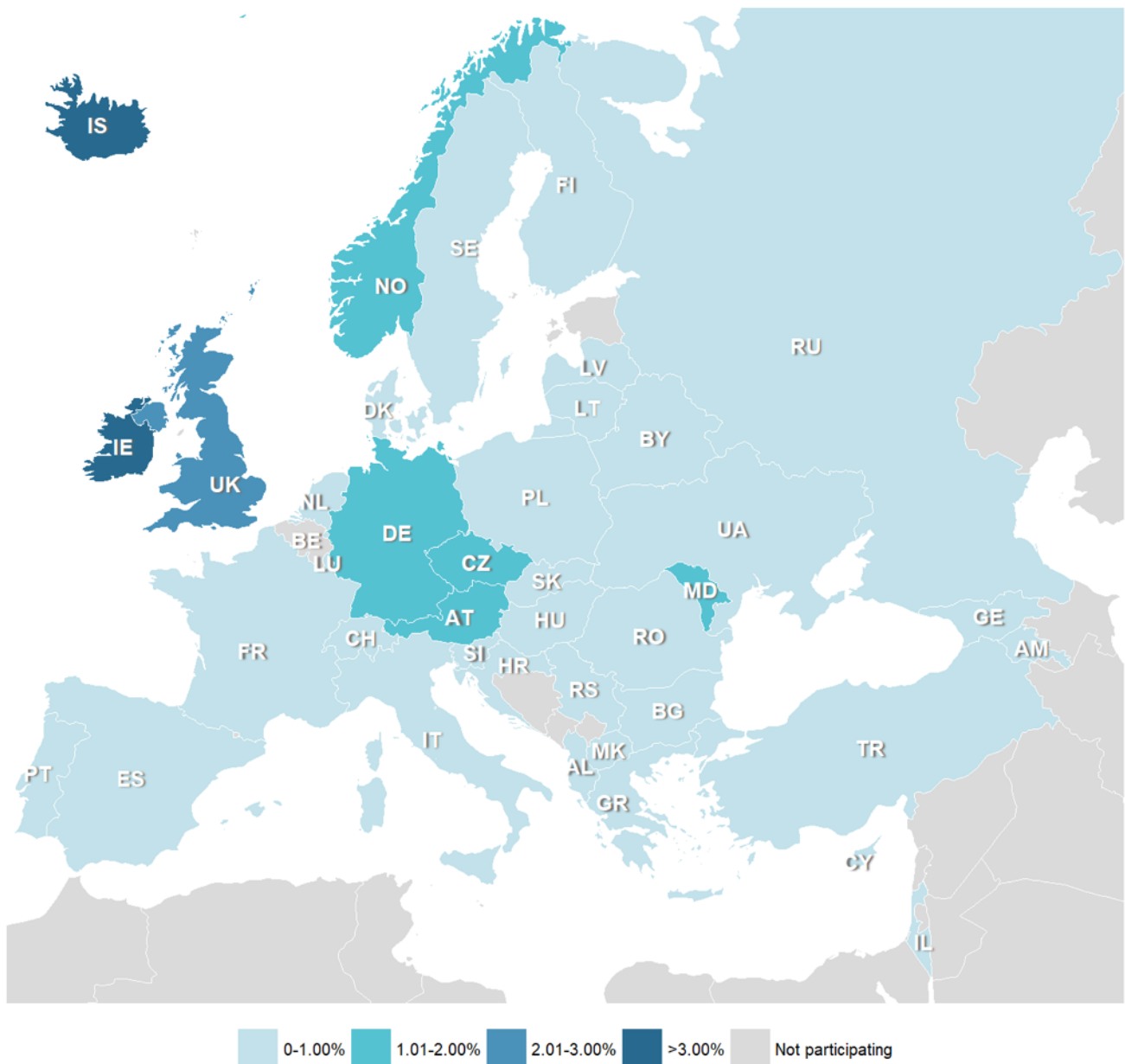
The *G551D* variant is most frequent in Ireland (8.3%) and in the North and Central region of Europe whereas it is rare in the East and South of Europe.

3. Genetics

Figure 3.7   Geographical distribution of the 2789+5G->A variant.
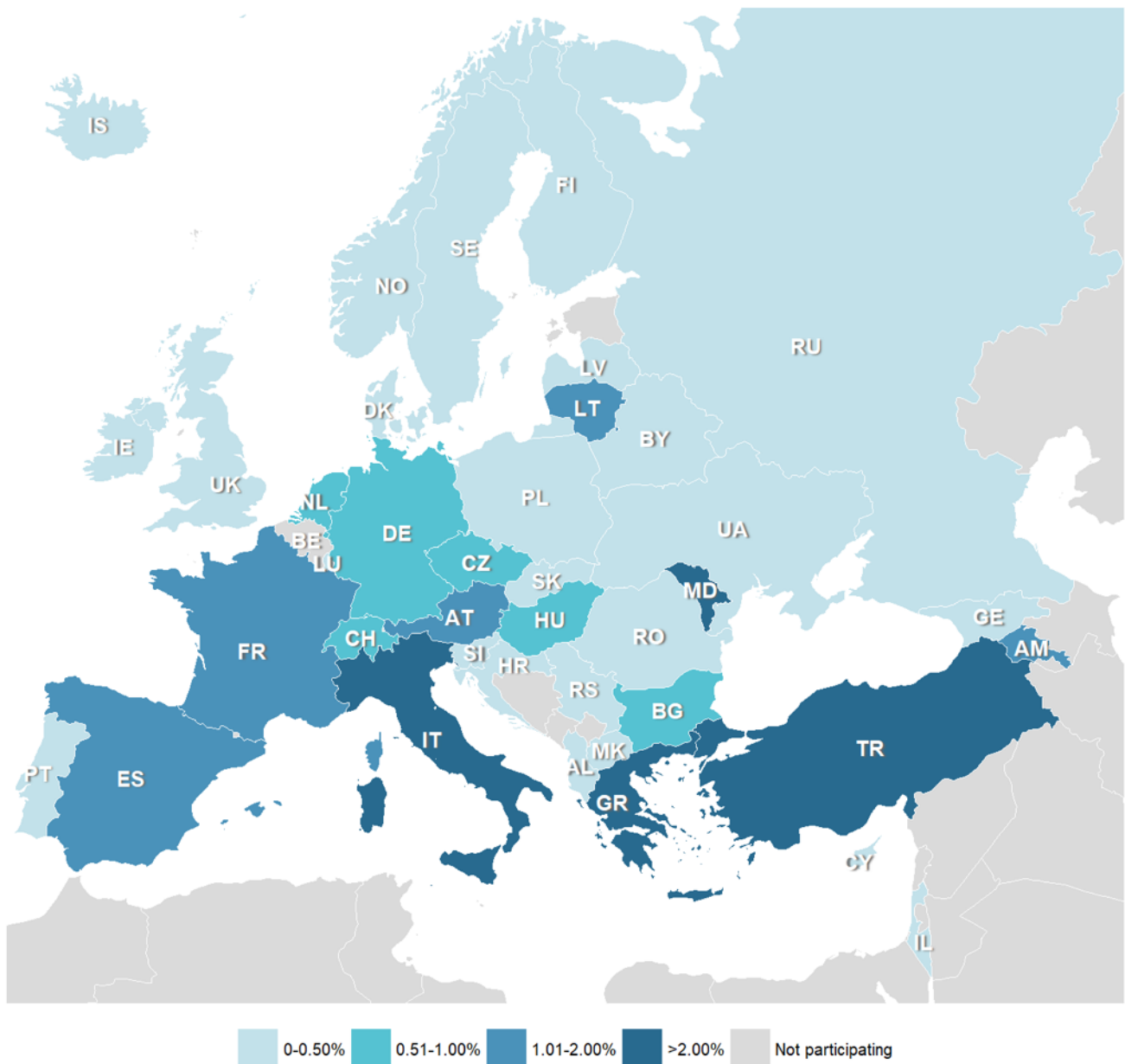
The *2789+5G->A* variant is most frequent in Turkey (3.1%), and in the Southern region of Europe, whereas it is less common in Eastern Europe.

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 FEV_1 (Forced Expiratory Volume$_1$) is measured in litres but the lung capacity is normally expressed as a percentage of the expected (or predicted) value (FEV_1% of predicted). The predicted value is determined from healthy individuals of the same age, sex, ethnicity, height and is called the 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). This is the global reference for spirometry and it has been agreed, as part of the CF global harmonisation project, that this is the best way to present lung function.

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.

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

We asked the countries to report the best FEV_1 recorded throughout the year (from the best FEV_1% of predicted computed at the CF centres).

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 had a liver transplantation since follow-up data for them is sometimes missing.

4. Lung function

Figure 4.1   Median FEV1% of predicted of young children and adolescents with CF <18 years of age is >80% in almost all countries in Europe.

FEV1% of predicted: boxplot by country. Children and adolescents with CF aged 6-17 years who have never had an organ transplant, seen in 2022 (table A4.1, Appendix 1).
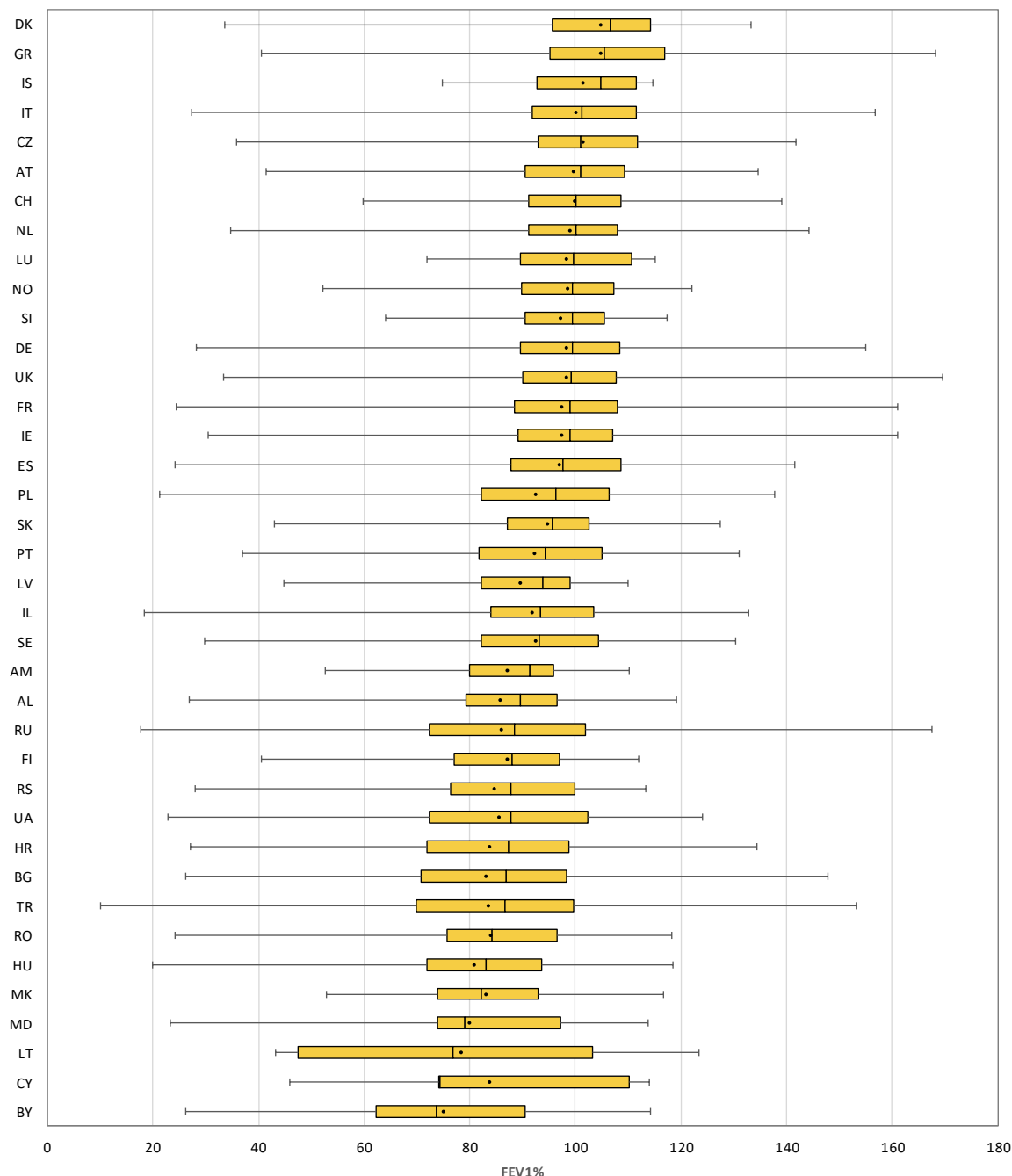
Note: Georgia has <5 individuals aged 6-17 years at the date of FEV$_1$ measurement and is excluded from the graph.

Sweden and the United Kingdom report FEV$_1$ from the annual review, which might not be the FEV1 of the best FEV$_1$% of the year, and, in some cases, the FEV$_1$ measurement could 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 of adults with CF varies between <60% and >90% depending on the country.

FEV1% of predicted: boxplot by country. Adults with CF who have never had a transplant, seen in 2022 (table A4.2, Appendix 1).

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

Note   Sweden and the United Kingdom report FEV_1 from the annual review, which might not be the FEV1 of the best FEV1% of predicted of the year and, in some cases, the FEV_1 measurement could 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   FEV₁% 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	7386	999	97.7	17.9	88.7	99.3	108.4	169.5
12-17	7577	435	93.5	10.0	83.8	96.0	106.2	168.2
18-29	11131	439	82.3	12.1	66.8	86.0	99.5	193.3
30-39	6412	234	73.4	10.2	55.1	74.4	91.4	138.9
40-49	3373	78	70.1	13.5	51.0	69.5	88.5	164.4
50-59	1626	37	69.0	17.8	49.8	69.3	86.6	138.3
60+	665	13	70.3	17.5	51.4	70.2	87.8	147.8

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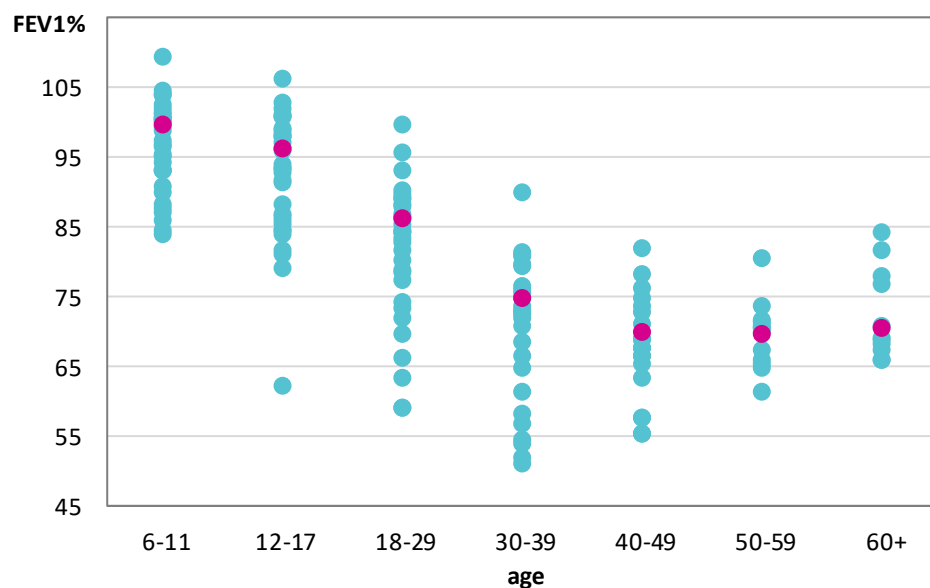
Note: We excluded from the graph those age groups where the number of individuals was <10.

This graph shows the median (the value that separates the highest and lowest half of the people with CF) 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, Georgia, Iceland and Luxembourg from the graphs because none of the age groups had more than 10 people with CF.

Quartiles of FEV_1%: Albania
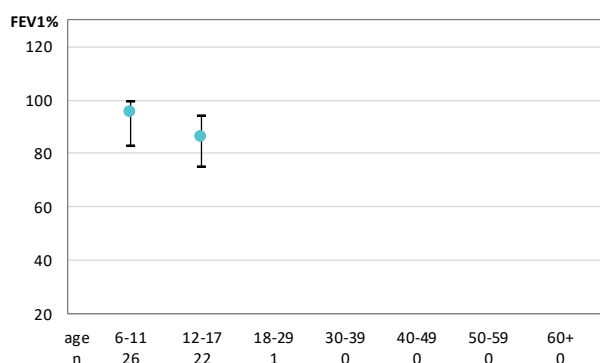
Quartiles of FEV_1%: Armenia
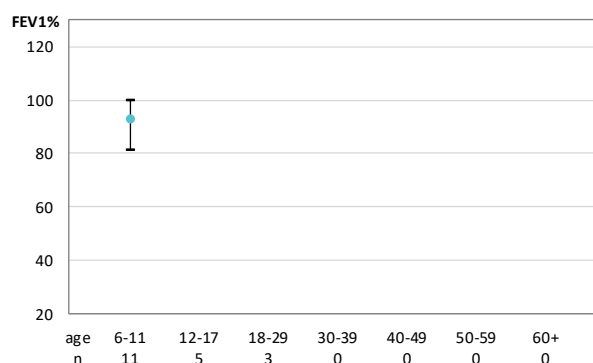
Quartiles of FEV_1%: Austria
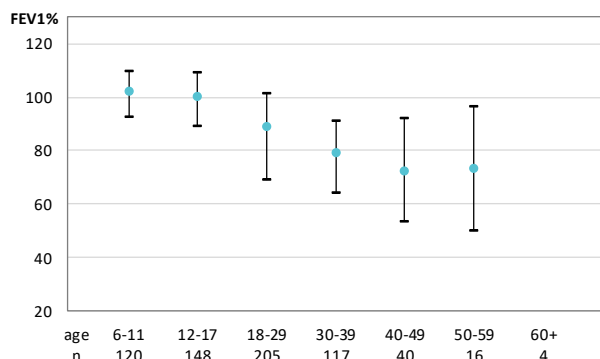
Quartiles of FEV_1%: Belarus
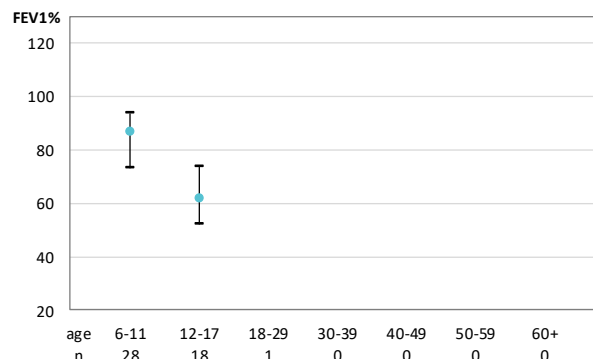
Quartiles of FEV_1%: Bulgaria
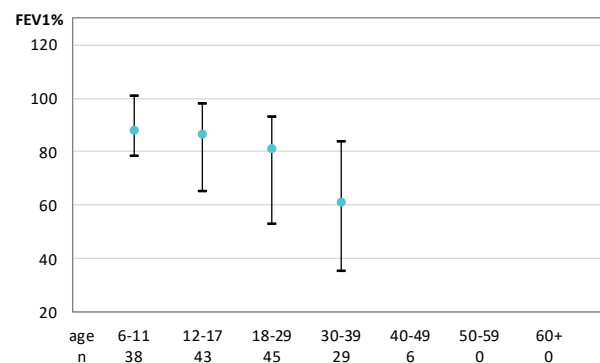
Quartiles of FEV_1%: Croatia
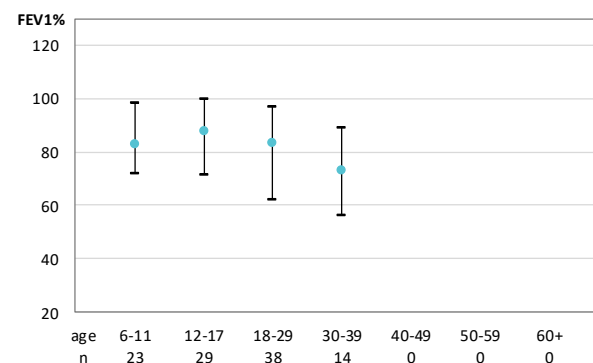
4. Lung function

[figure 4.4 continued]

Quartiles of FEV₁%: Czech Republic
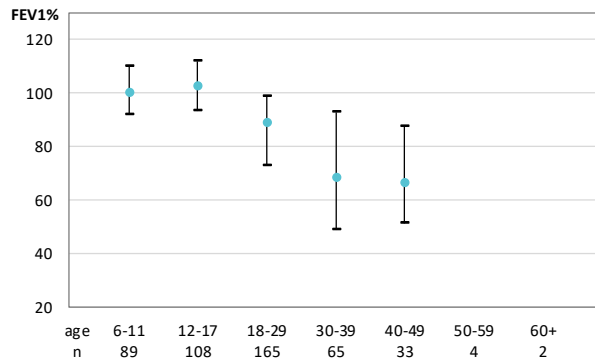
Quartiles of FEV₁%: Denmark
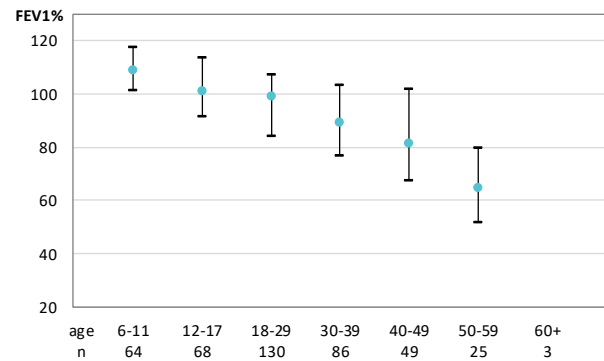
Quartiles of FEV₁%: Finland
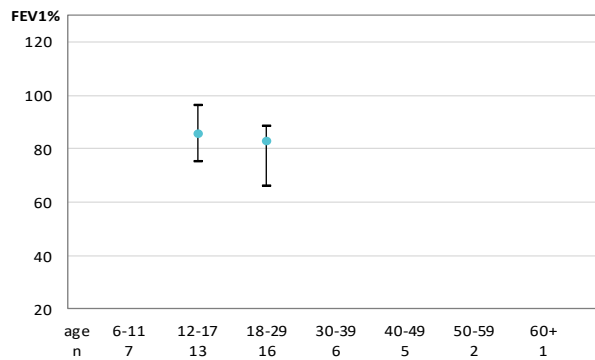
Quartiles of FEV₁%: France
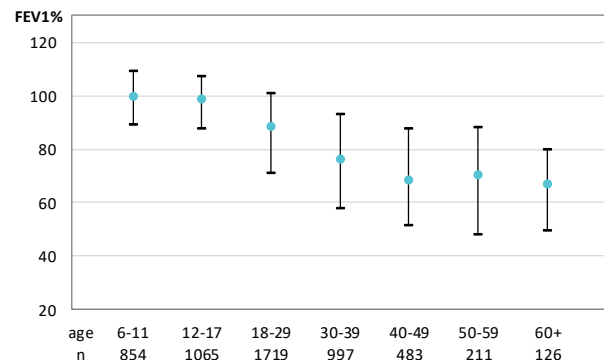
Quartiles of FEV₁%: Germany
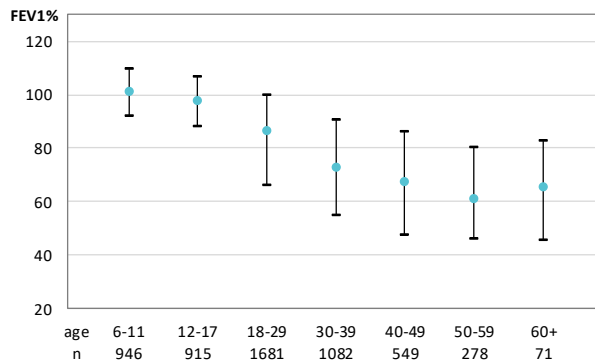
Quartiles of FEV₁%: Greece
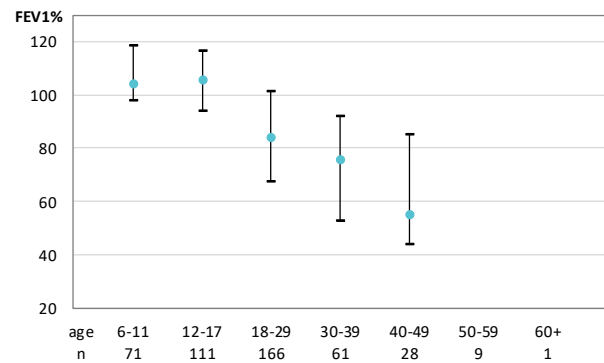
Quartiles of FEV₁%: Hungary
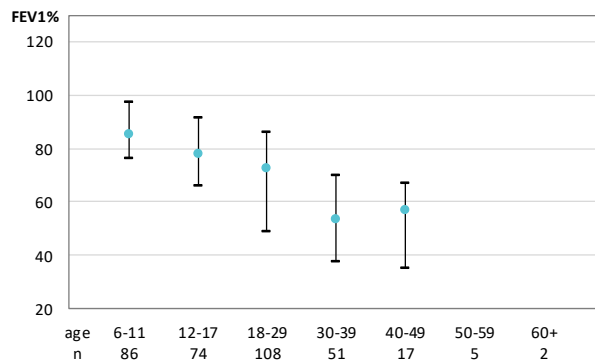
Quartiles of FEV₁%: Ireland
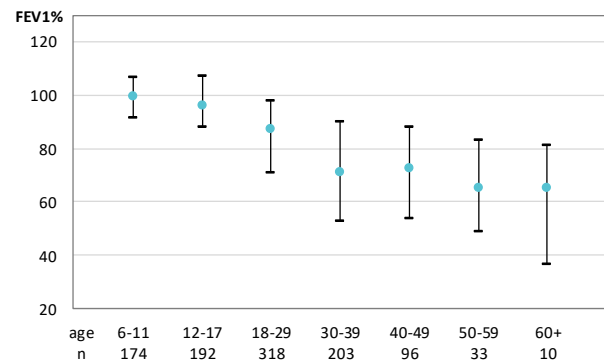
4. Lung function

[figure 4.4 continued]

Quartiles of FEV₁%: Israel

age	6-11	12-17	18-29	30-39	40-49	50-59	60+
n	53	84	156	92	51	15	12

Quartiles of FEV₁%: Italy

age	6-11	12-17	18-29	30-39	40-49	50-59	60+
n	754	781	1406	849	595	343	110
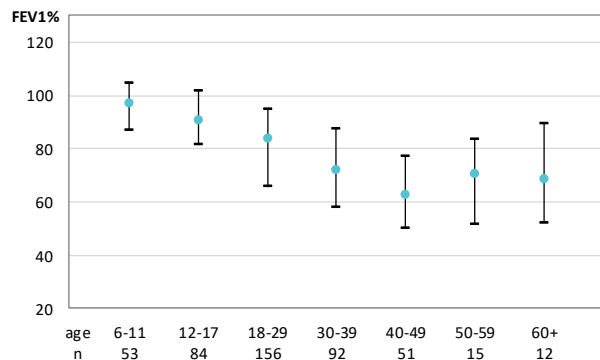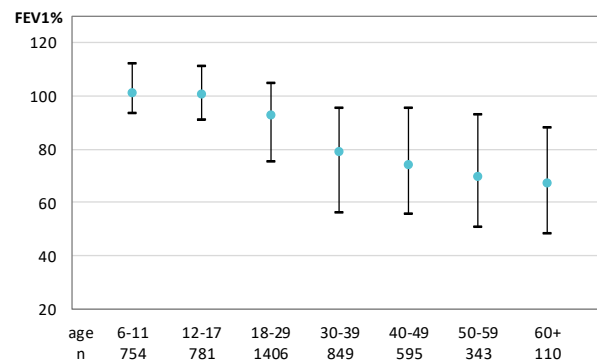
Quartiles of FEV₁%: Lithuania

age	6-11	12-17	18-29	30-39	40-49	50-59	60+
n	3	7	14	8	0	0	0

Quartiles of FEV₁%: Latvia

age	6-11	12-17	18-29	30-39	40-49	50-59	60+
n	8	10	12	2	0	0	0
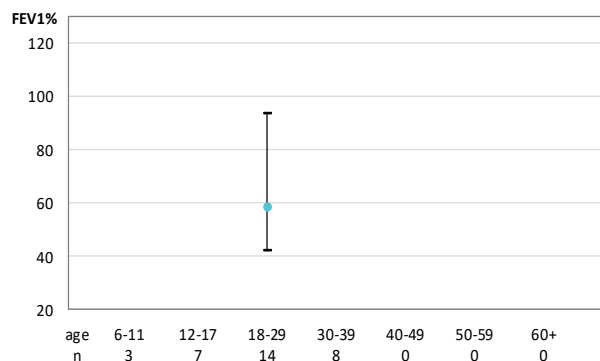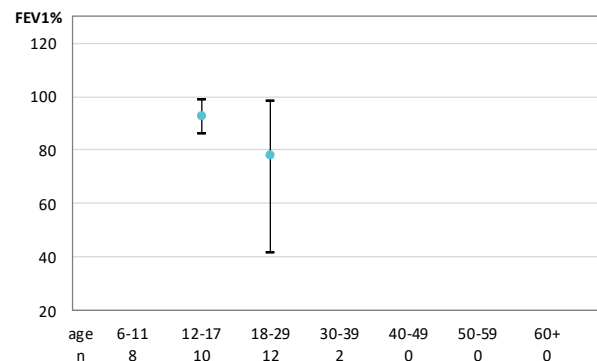
Quartiles of FEV₁%: North Macedonia

age	6-11	12-17	18-29	30-39	40-49	50-59	60+
n	15	28	31	11	3	0	0
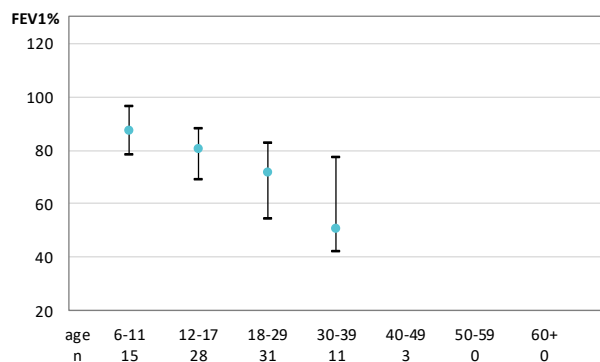
Quartiles of FEV₁%: Rep of Moldova

age	6-11	12-17	18-29	30-39	40-49	50-59	60+
n	10	9	4	3	0	0	0
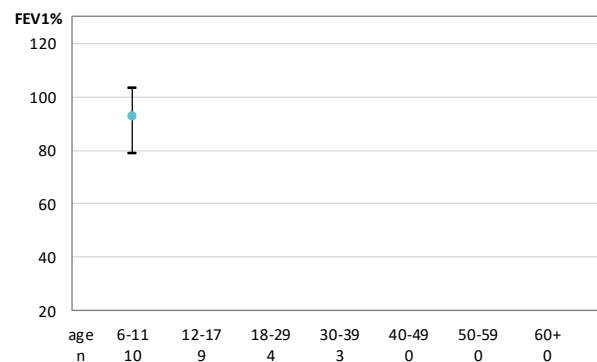
Quartiles of FEV₁%: The Netherlands
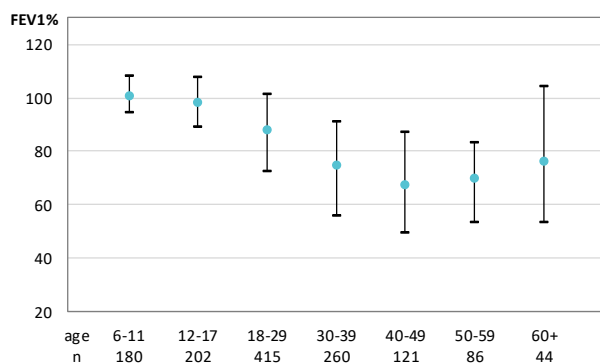
age	6-11	12-17	18-29	30-39	40-49	50-59	60+
n	180	202	415	260	121	86	44

Quartiles of FEV₁%: Norway

age	6-11	12-17	18-29	30-39	40-49	50-59	60+
n	38	45	67	51	36	19	14

4. Lung function

[figure 4.4 continued]

Quartiles of FEV$_1$%: Poland

age	6-11	12-17	18-29	30-39	40-49	50-59	60+
n	282	332	334	95	35	8	1
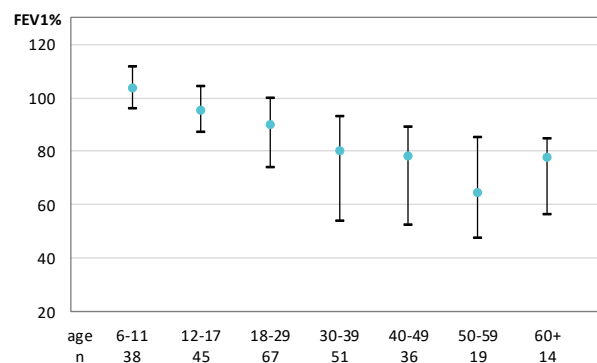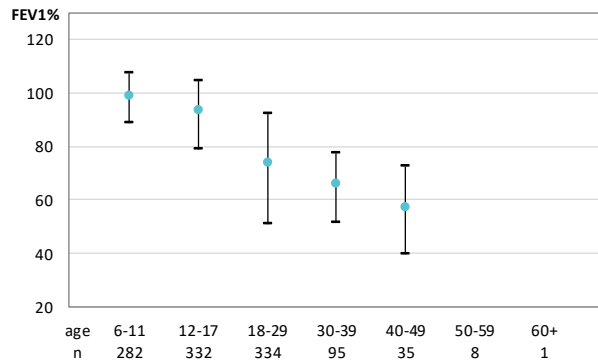
Quartiles of FEV$_1$%: Portugal

age	6-11	12-17	18-29	30-39	40-49	50-59	60+
n	57	57	91	37	28	6	3
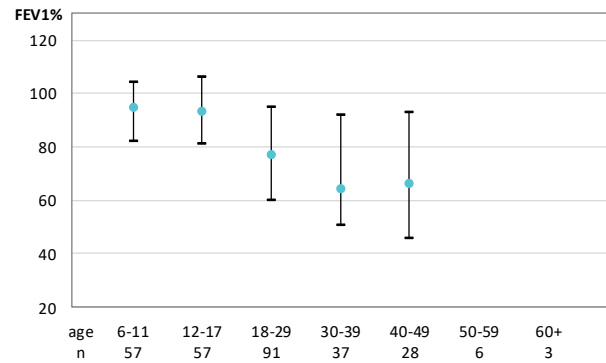
Quartiles of FEV$_1$%: Romania

age	6-11	12-17	18-29	30-39	40-49	50-59	60+
n	59	84	13	0	0	0	0
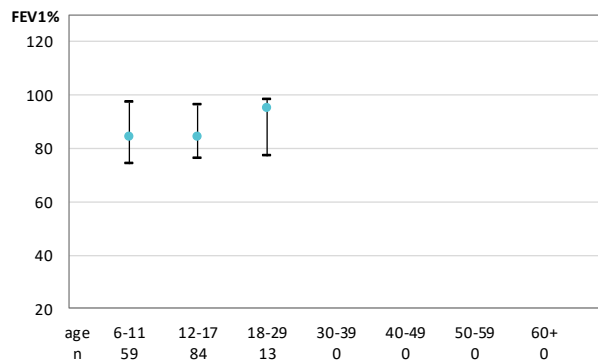
Quartiles of FEV1%: Russian Federation

age	6-11	12-17	18-29	30-39	40-49	50-59	60+
n	583	463	147	54	9	1	0

Quartiles of FEV$_1$%: Serbia

age	6-11	12-17	18-29	30-39	40-49	50-59	60+
n	33	46	43	12	5	0	0
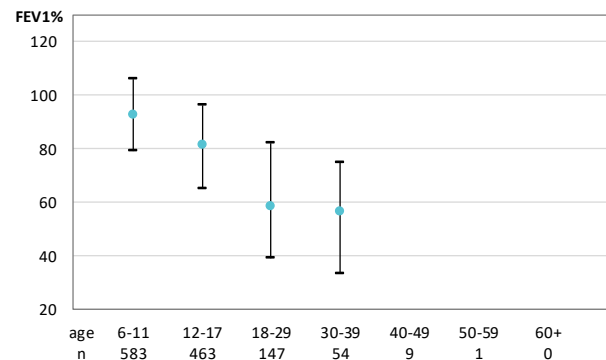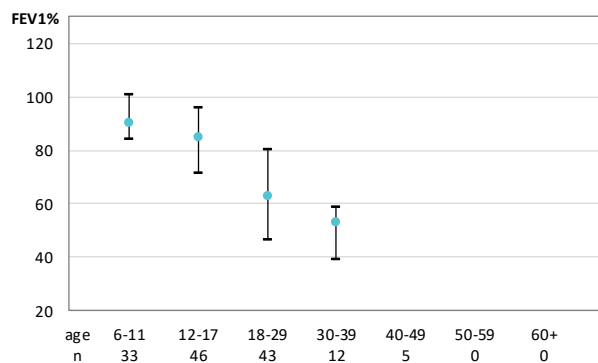
Quartiles of FEV$_1$%: Slovak Republic
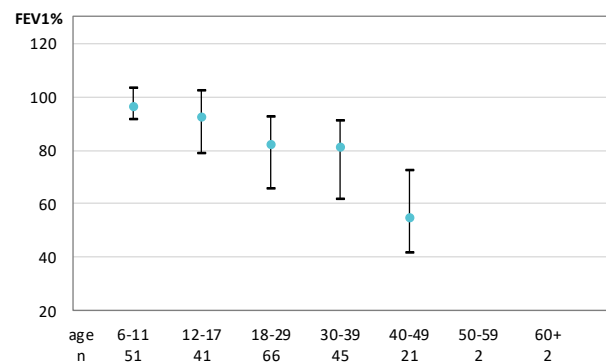
age	6-11	12-17	18-29	30-39	40-49	50-59	60+
n	51	41	66	45	21	2	2

Quartiles of FEV$_1$%: Slovenia

age	6-11	12-17	18-29	30-39	40-49	50-59	60+
n	28	19	30	8	3	3	0

Quartiles of FEV$_1$%: Spain

age	6-11	12-17	18-29	30-39	40-49	50-59	60+
n	378	394	507	289	205	93	42

4. Lung function

[figure 4.4 continued]

Quartiles of FEV$_1$%: Sweden

age	6-11	12-17	18-29	30-39	40-49	50-59	60+
n	87	115	153	103	64	37	11

Quartiles of FEV$_1$%: Switzerland

age	6-11	12-17	18-29	30-39	40-49	50-59	60+
n	154	145	268	123	79	33	18

Quartiles of FEV$_1$%: Turkey

age	6-11	12-17	18-29	30-39	40-49	50-59	60+
n	471	430	240	35	5	0	0
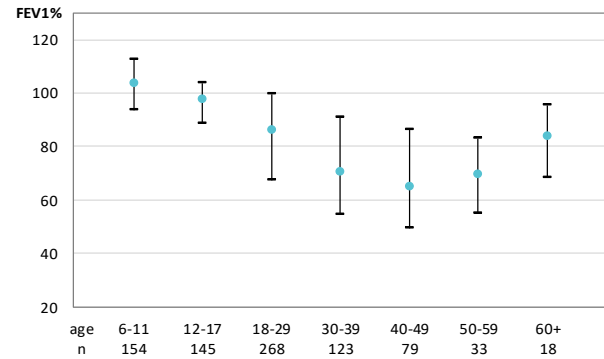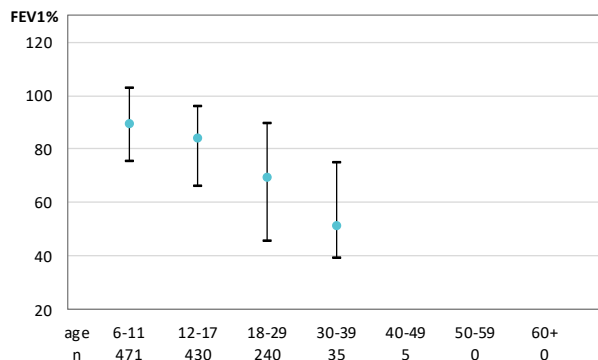
Quartiles of FEV$_1$%: Ukraine

age	6-11	12-17	18-29	30-39	40-49	50-59	60+
n	55	49	48	14	3	0	0
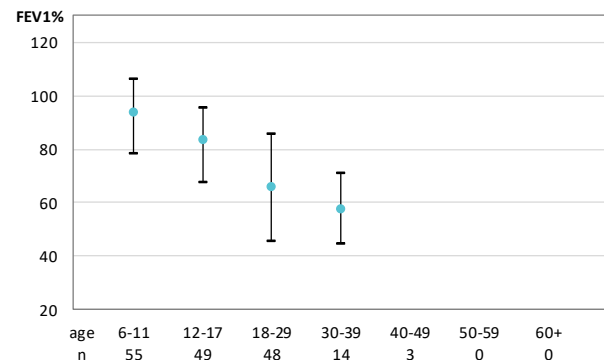
Quartiles of FEV$_1$%: United Kingdom
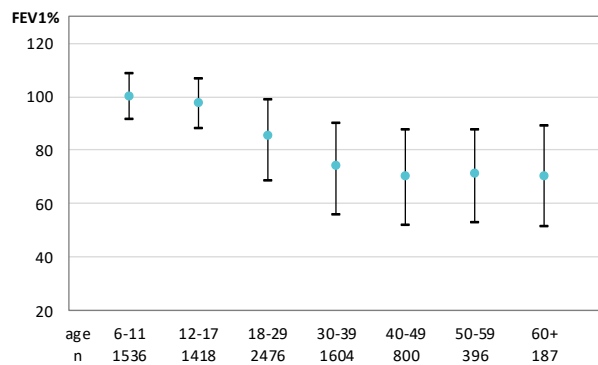
age	6-11	12-17	18-29	30-39	40-49	50-59	60+
n	1536	1418	2476	1604	800	396	187

Note: Sweden and the United Kingdom report FEV$_1$ from the annual review, which might not be the FEV1 of the best FEV$_1$% of predicted of the year and, in some cases, the FEV$_1$ measurement could 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 transplant.
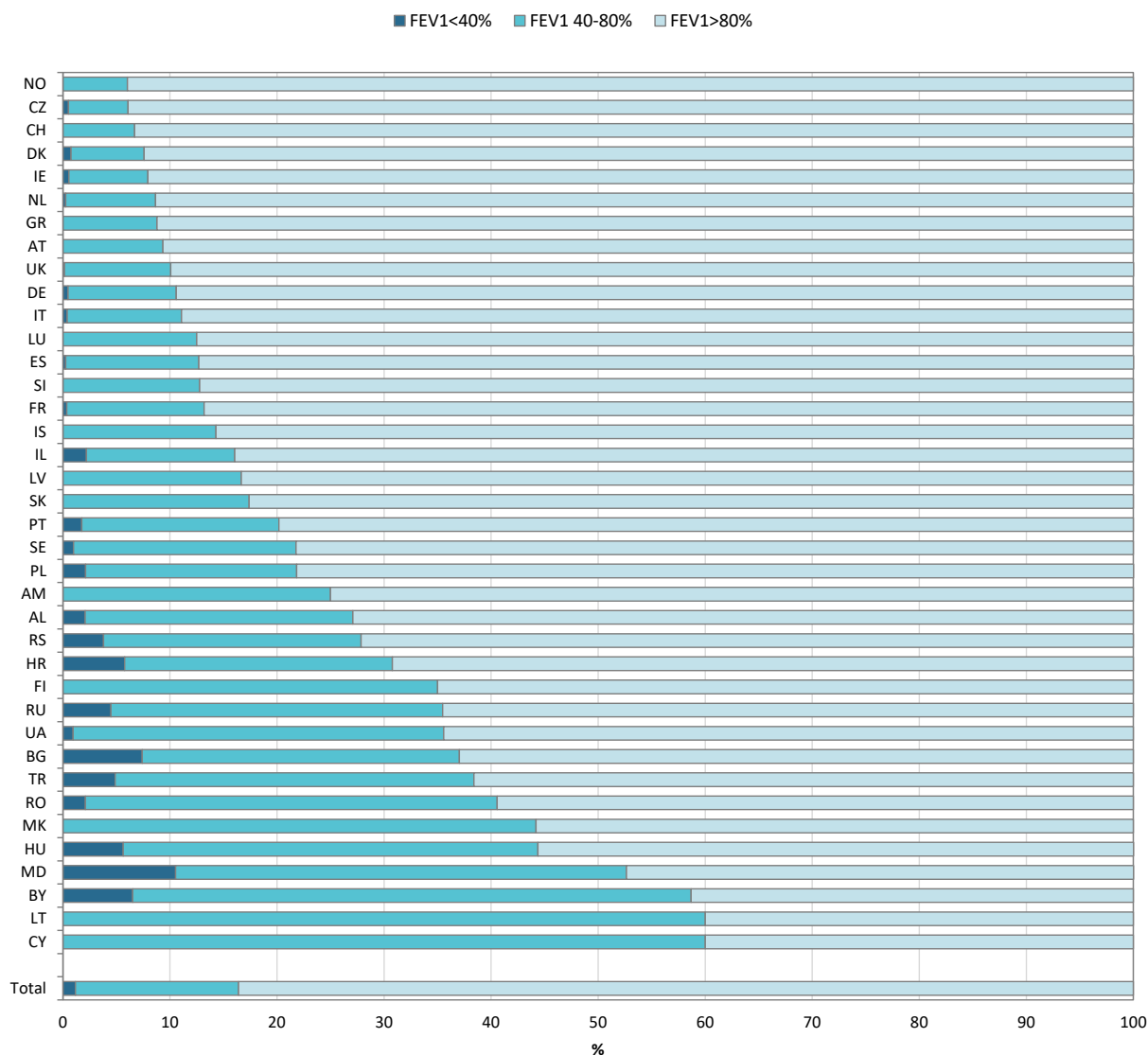
Note: Georgia has <5 people with CF aged 6-17 years at FEV1 measurement and is excluded from the graph.
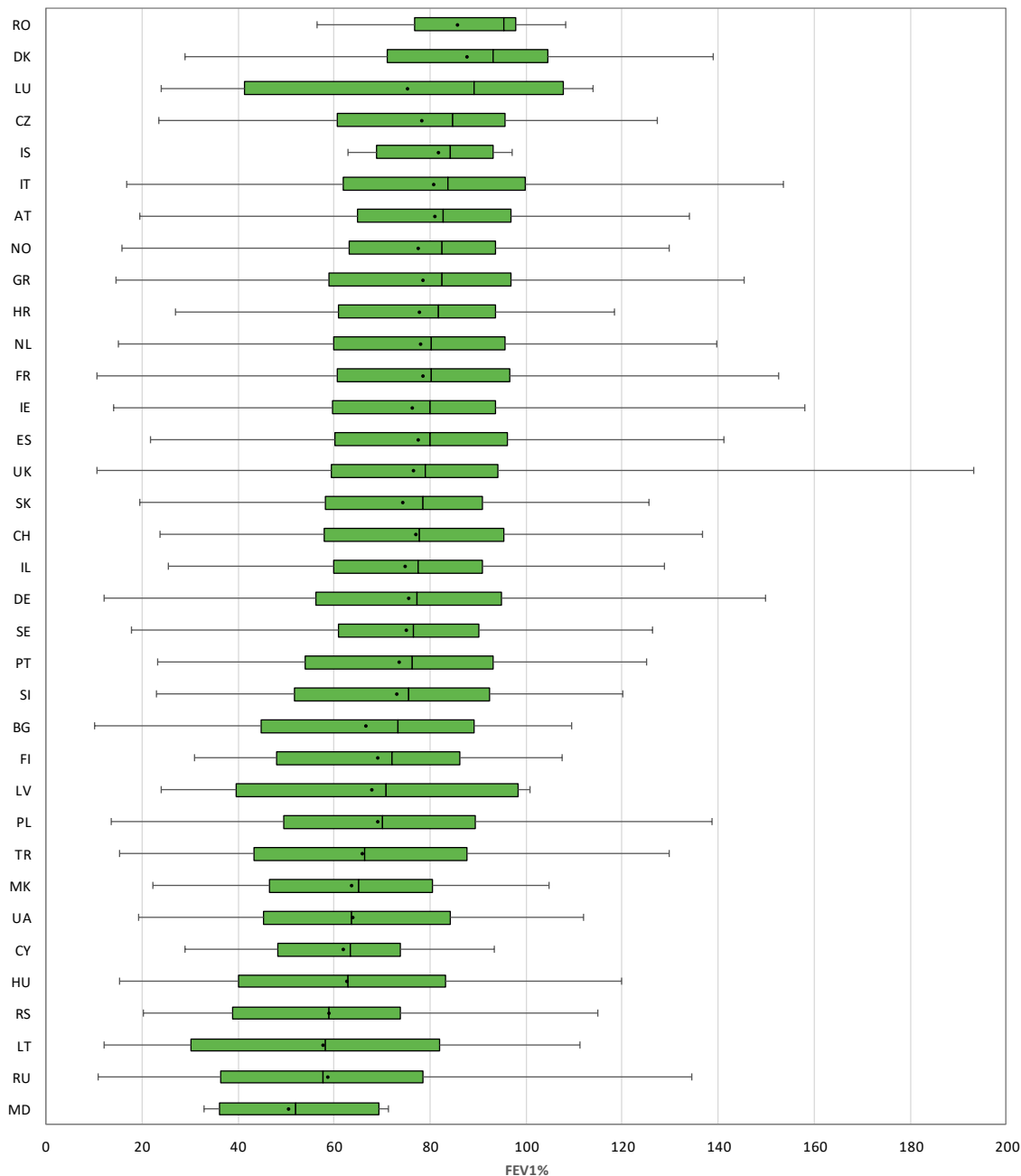
Note: Sweden and the United Kingdom reports FEV$_1$ from the annual review which might not be the FEV1 of the best FEV$_1$% of the year and, in some cases, the FEV$_1$ measurement could be from the previous calendar year.

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 10-20%.

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.
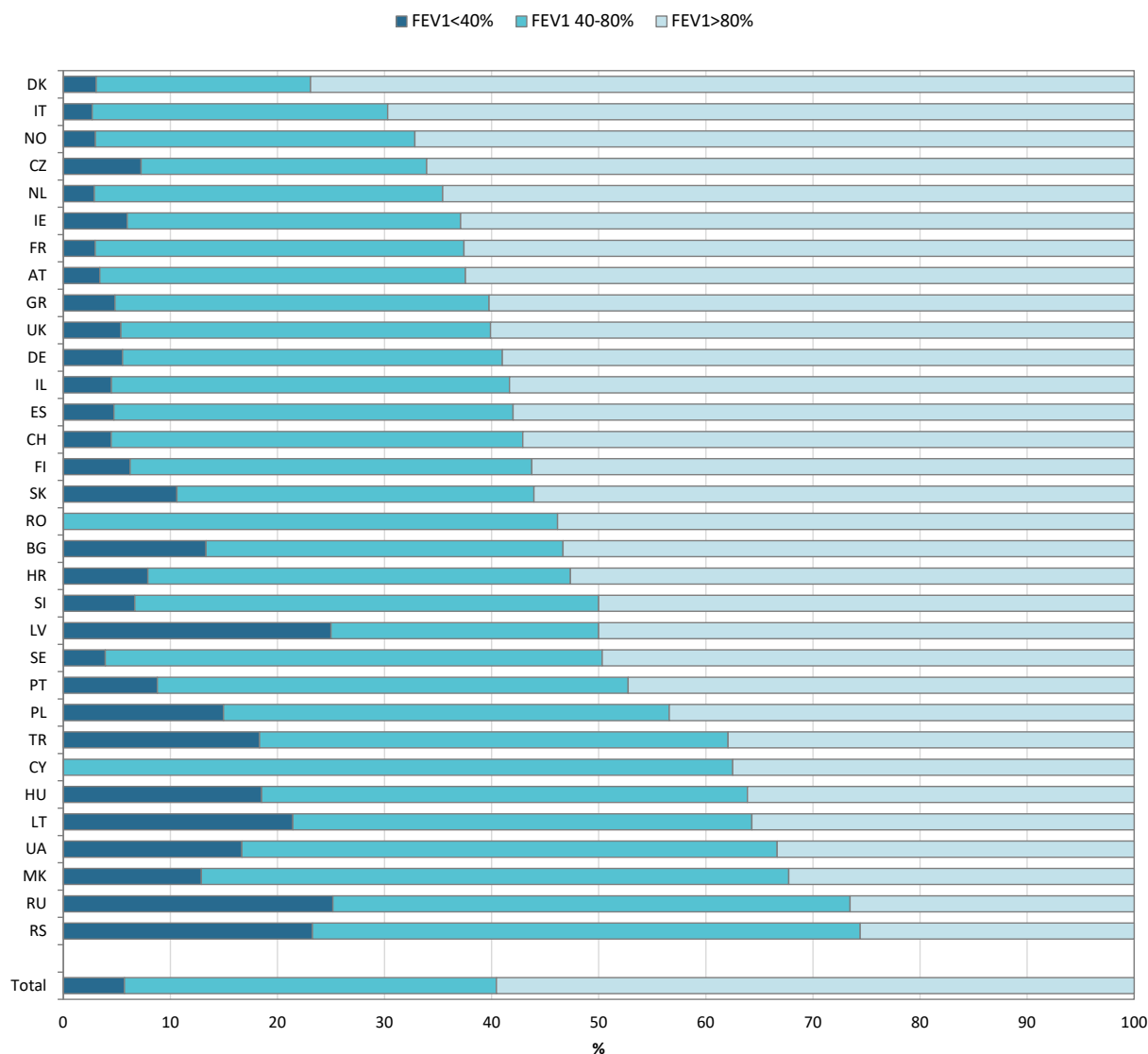
Note: Albania, Armenia, Belarus, Georgia, Iceland, Luxembourg and Rep of Moldova have <5 people aged 18-29 years with FEV$_1$ measurement and are excluded from the graph.

Note: Sweden and the United Kingdom report FEV$_1$ from the annual review, which might not be the FEV1 of the best FEV$_1$% of predicted of the year, and, in some cases, the FEV$_1$ measurement could 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.
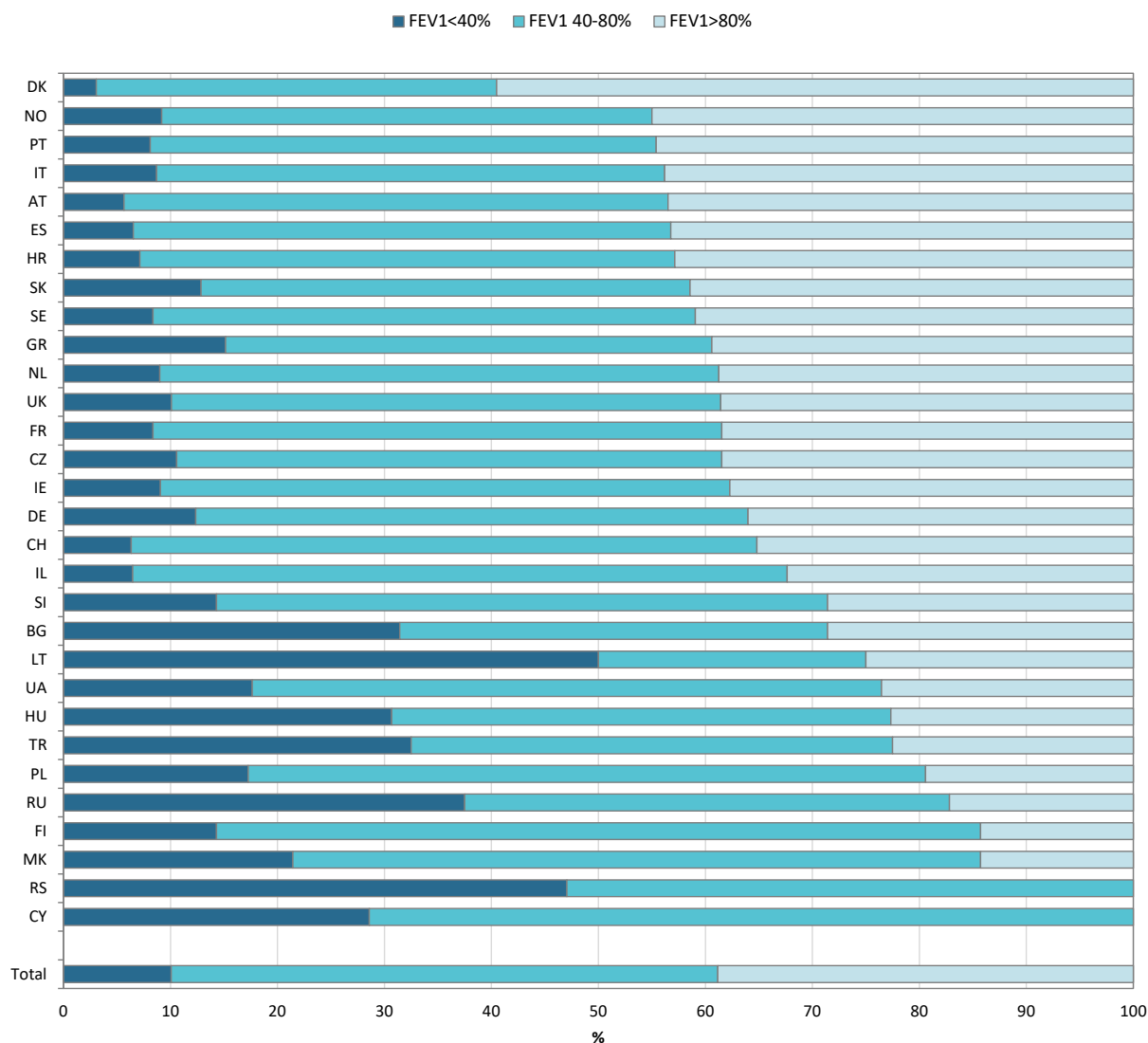
Note: Albania, Armenia, Belarus, Georgia, Iceland, Latvia, Luxembourg, Rep of Moldova, and Romania have <5 people aged 30 years or more with FEV$_1$ measurement and are excluded from the graph.

Note: Sweden and the United Kingdom report FEV$_1$ from the annual review, which might not be the FEV1 of the best FEV$_1$% of predicted of the year and, in some cases, the FEV$_1$ measurement could 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 2012, 2017 and 2022.

age	6-11	12-17	18-29	30-39	40-49	50-59	60+
2022	7386	7578	11134	6419	3376	1627	665
2017	6991	6567	10151	4932	2371	923	354
2012	5138	5495	8372	3366	1580	416	175
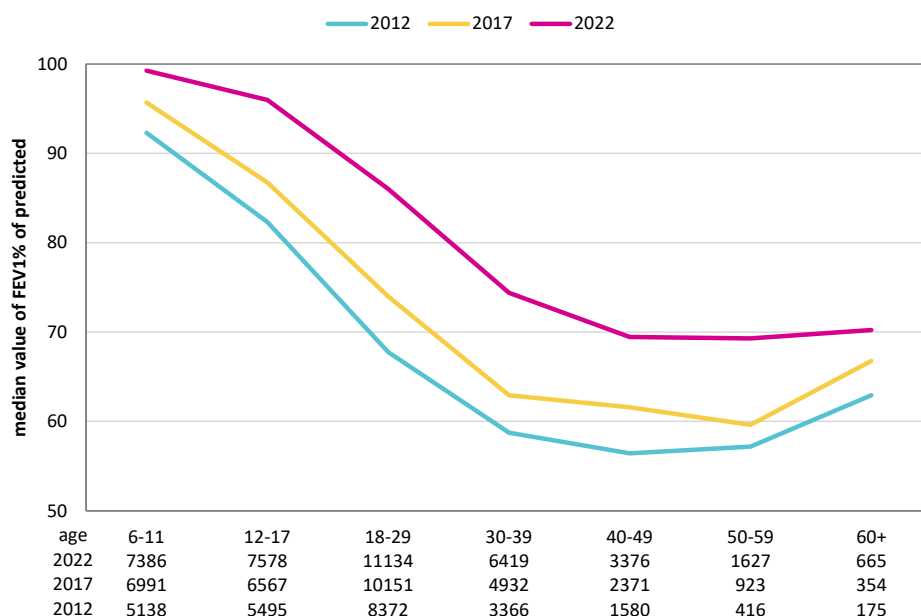
Note: People with CF aged 6 years or more at lung function measurement, who have never had a lung or liver transplant.

In this graph we present data over time using cross sectional data per year of people with a confirmed CF diagnosis. The years 2012, 2017 and 2022 were used for the analysis. All people with CF alive, deceased, or not seen during the year of follow-up were included. Exclusion criteria were people who were lost to follow-up and/or transplanted (lung and/or liver), people with missing values, 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 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 are positive; at least 4 samples were collected during that period;

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 2022 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, Armenia and Georgia have <5 adults seen in 2022 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 2022.

           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 the bars 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 increasing prevalence in some countries.

Prevalence of chronic *Burkholderia cepacia complex* species in people with CF seen in 2022 who have never had a transplant, by country (table A5.2, Appendix 1).

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. People with CF are less frequently chronically infected with *Burkholderia cepacia* complex species than with *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* in people with CF seen in 2022 who have never had a transplant, by country (table A5.3, Appendix 1).
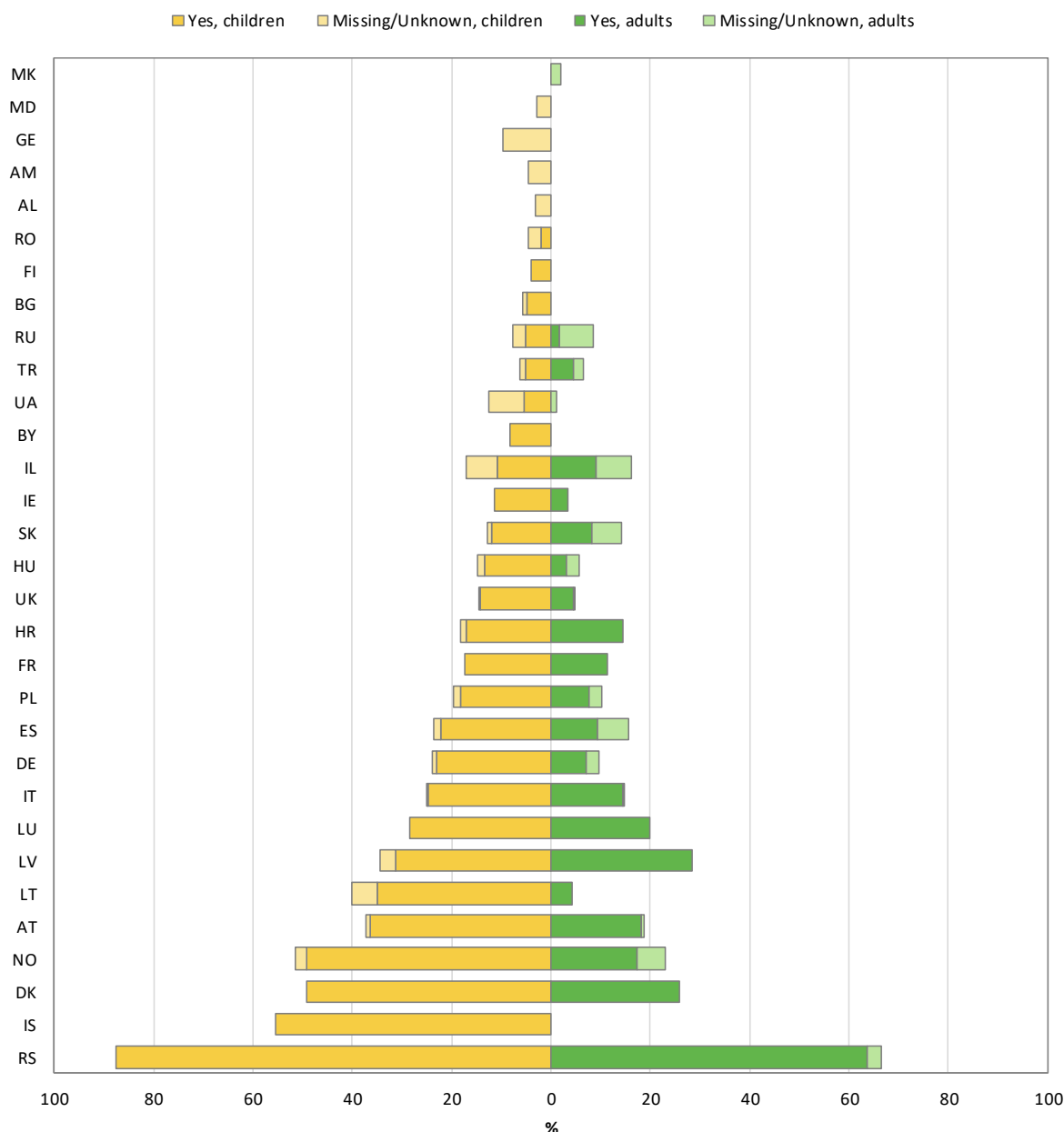
Note: We excluded from the graph the countries for which the information is missing for more than 10% of the children/adults. Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the graph for adults.

Note: Belgium, France, Germany 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 2022.

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 2022 who have never had a transplant, by country (table A5.4, Appendix 1).
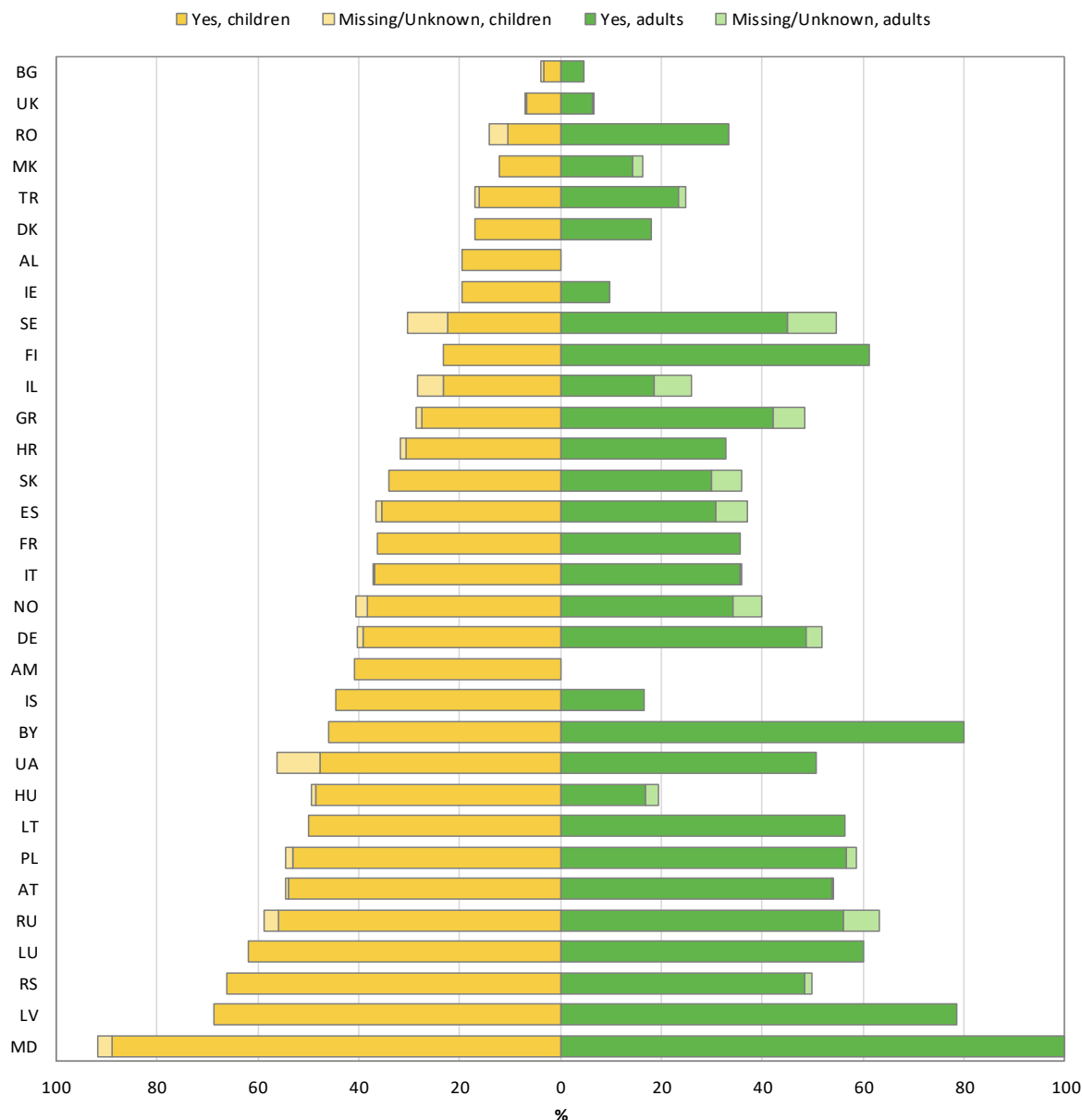
Note: We excluded from the graph the countries for which the information is missing for more than 10% of the children/adults. Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the graph for adults.

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 2022.

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) in people with CF seen in 2022 who have never had a transplant, by country (table A5.5, Appendix 1).
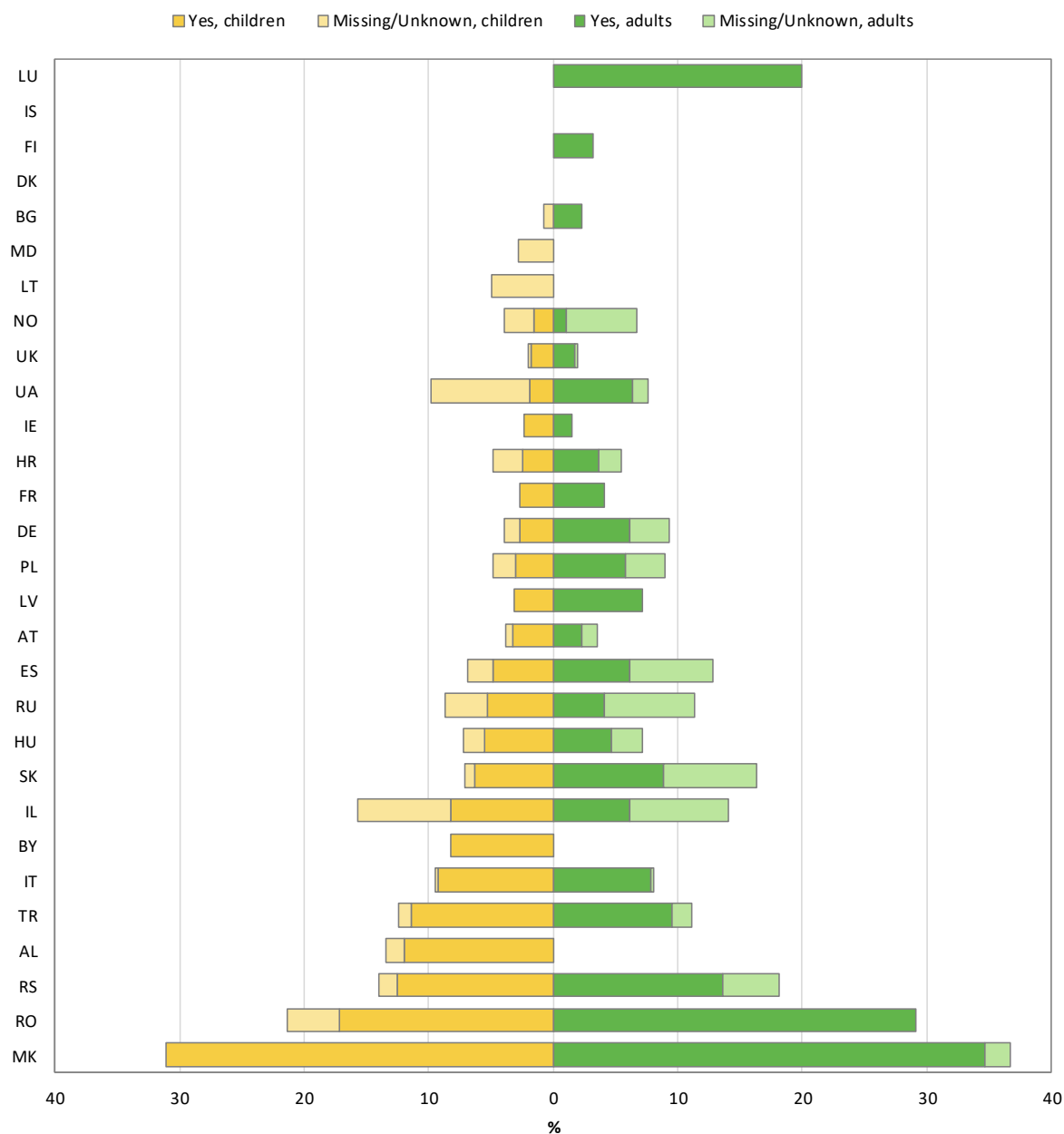
This graph represents the percentage of people with methicillin-resistant *Staphylococcus aureus* (MRSA) infection (in dark colours) and the percentage of people where information on methicillin-resistant *Staphylococcus aureus* infection is missing/unknown (in light colours). The horizontal bars on 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 a significant number of airway samples in children and adults with CF.

Prevalence of *Stenotrophomonas maltophilia* in people with CF seen in 2022 who have never had a transplant, by country (table A5.6, Appendix 1).
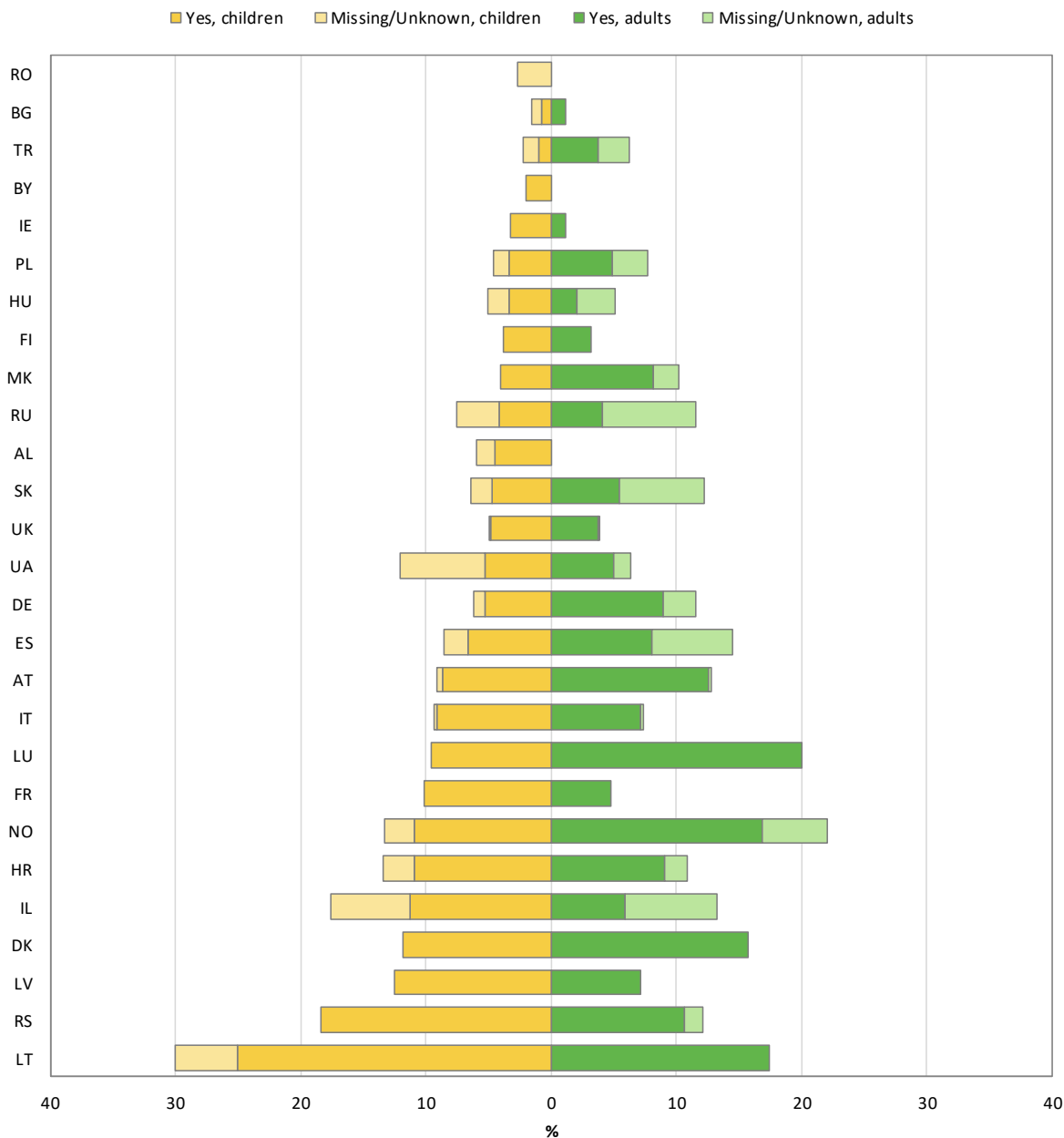
Note: We excluded from the graph the countries for which the information is missing for more than 10% of the children/adults. Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the graph for adults.

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 2022.

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 in people with CF seen in 2022 who have never had a transplant, by country (table A5.7, Appendix 1).
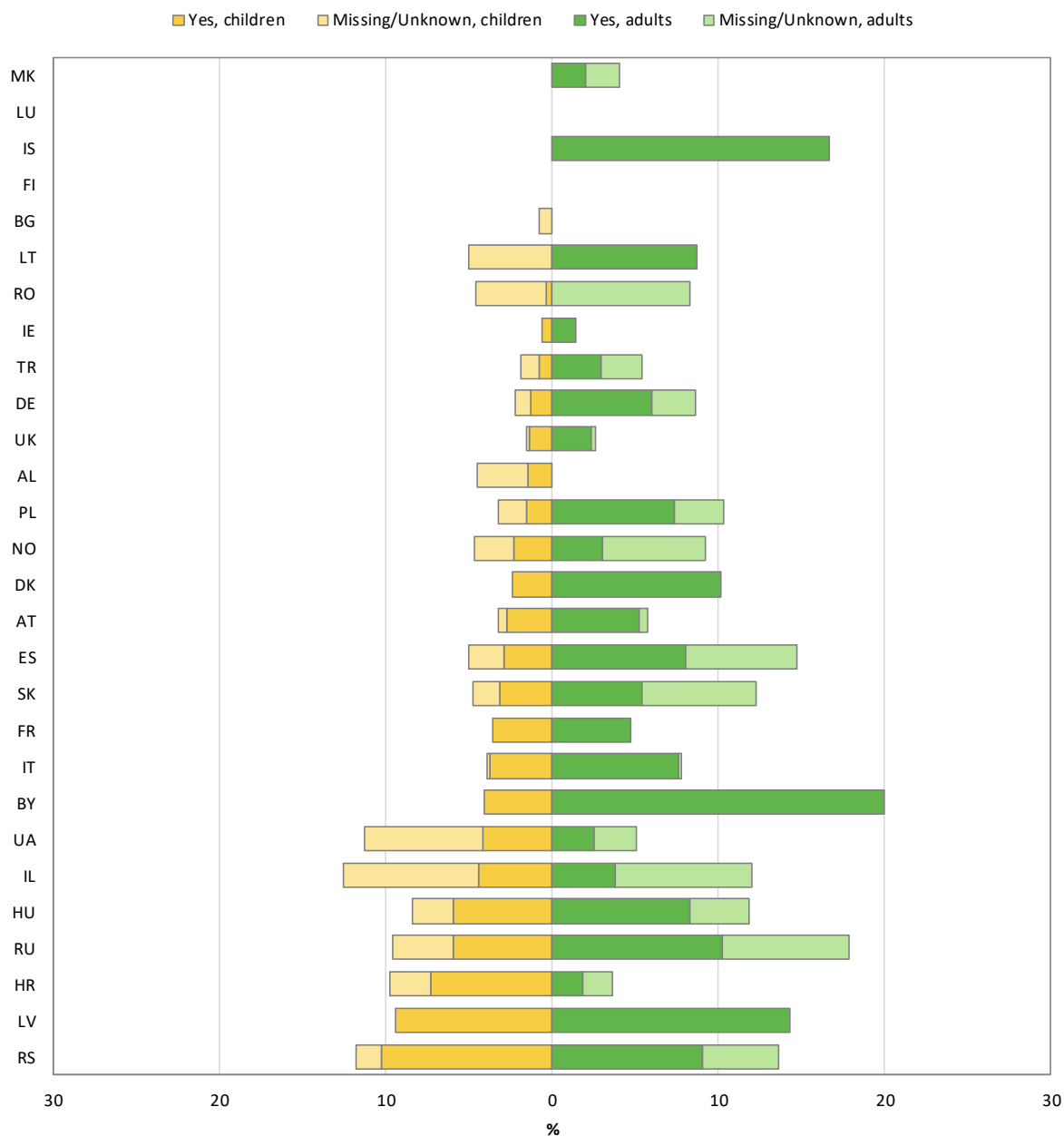
Note: We excluded from the graph the countries for which the information is missing for more than 10% of the children/adults. Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the graph for adults.

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 2022.

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 2022 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	1	1.5	66	98.5	0	0.0	0	0.0	0	0.0
Armenia	0	0.0	17	77.3	5	22.7	5	100	0	0.0
Austria	8	2.2	214	57.7	149	40.2	136	91.3	13	8.7
Belarus	0	0.0	146	100	0	0.0	0	0.0	0	0.0
Bulgaria	1	0.8	124	98.4	1	0.8	0	0.0	1	100
Croatia	9	11.0	32	39.0	41	50.0	41	100	0	0.0
Cyprus	0	0.0	7	100	0	0.0	0	0.0	0	0.0
Czech Republic	40	12.4	270	83.3	14	4.3	14	100	0	0.0
Denmark	0	0.0	0	0.0	211	100	210	99.5	1	0.5
Finland	25	96.2	0	0.0	1	3.9	0	0.0	1	100
France	456	17.0	1025	38.1	1210	45.0	1166	96.4	44	3.6
Georgia	14	34.2	27	65.9	0	0.0	0	0.0	0	0.0
Germany	36	1.3	2325	84.4	395	14.3	385	97.5	10	2.5
Greece	237	100	--	-	-	-	-	-	-	-
Hungary	5	2.1	156	65.8	76	32.1	75	98.7	1	1.3
Iceland	0	0.0	7	77.8	2	22.2	2	100	0	0.0
Ireland	0	0.0	83	15.8	442	84.2	441	99.8	1	0.2
Israel	4	2.5	79	49.7	76	47.8	66	86.8	10	13.2
Italy	23	1.0	1456	66.1	725	32.9	709	97.8	16	2.2
Latvia	0	0.0	14	43.8	18	56.3	18	100	0	0.0
Lithuania	1	5.0	14	70.0	5	25.0	4	80.0	1	20.0
Luxembourg	0	0.0	7	33.3	14	66.7	14	100	0	0.0
Rep of Moldova	3	8.3	33	91.7	0	0.0	0	0.0	0	0.0
The Netherlands	203	37.7	0	0.0	336	62.3	331	98.5	5	1.5
North Macedonia	0	0.0	50	67.6	24	32.4	23	95.8	1	4.2
Norway	3	2.3	34	26.6	91	71.1	87	95.6	4	4.4
Poland	39	4.1	836	87.7	78	8.2	74	94.9	4	5.1
Portugal	5	2.9	83	48.5	83	48.5	78	94.0	5	6.0
Romania	13	5.0	232	88.6	17	6.5	14	82.4	3	17.7
Russian Fed.	274	14.6	1359	72.5	241	12.9	235	97.5	6	2.5
Serbia	0	0.0	78	57.4	58	42.7	56	96.6	2	3.5
Slovak Republic	0	0.0	77	61.1	49	38.9	49	100	0	0.0
Slovenia	0	0.0	36	65.5	19	34.6	19	100	0	0.0
Spain	54	5.1	514	48.5	491	46.4	480	97.8	11	2.2
Sweden	0	0.0	77	29.1	188	70.9	184	97.9	4	2.1
Switzerland	10	2.5	381	93.6	16	3.9	15	93.8	1	6.3
Turkey	17	0.8	1768	82.6	356	16.6	340	95.5	16	4.5
Ukraine	57	21.5	202	76.2	6	2.3	6	100	0	0.0
United Kingdom	206	5.0	2656	64.2	1276	30.8	1211	94.9	65	5.1
Total	1744	7.6	14485	63.1	6714	29.3	6488	96.6	226	3.4

5. Microbiology

Table 5.2   Prevalence of non-tuberculous mycobacteria in adults (≥ 18 years) with CF seen in 2022 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	%
Austria	11	2.8	144	36.1	244	61.2	220	90.2	24	9.8
Belarus	0	0.0	5	100	0	0.0	0	0.0	0	0.0
Bulgaria	0	0.0	81	95.3	4	4.7	4	100	0	0.0
Croatia	0	0.0	5	9.1	50	90.9	50	100	0	0.0
Cyprus	0	0.0	13	81.3	3	18.8	2	66.7	1	33.3
Czech Republic	40	13.3	175	58.0	87	28.8	79	90.8	8	9.2
Denmark	0	0.0	0	0.0	305	100	297	97.4	8	2.6
Finland	28	90.3	0	0.0	3	9.7	2	66.7	1	33.3
France	937	25.4	943	25.6	1804	49.0	1697	94.1	107	5.9
Germany	126	3.3	2498	65.5	1192	31.2	1095	91.9	97	8.1
Greece	354	100	-	-	-	-	-	-	-	-
Hungary	3	1.6	16	8.3	175	90.2	161	92.0	14	8.0
Iceland	0	0.0	3	50.0	3	50.0	3	100	0	0.0
Ireland	0	0.0	319	44.4	400	55.6	398	99.5	2	0.5
Israel	9	2.7	157	46.2	174	51.2	138	79.3	36	20.7
Italy	57	1.6	2477	69.3	1039	29.1	961	92.5	78	7.5
Latvia	0	0.0	0	0.0	14	100	14	100	0	0.0
Lithuania	4	17.4	19	82.6	0	0.0	0	0.0	0	0.0
Luxembourg	1	20.0	0	0.0	4	80.0	3	75.0	1	25.0
Rep of Moldova	0	0.0	12	100	0	0.0	0	0.0	0	0.0
The Netherlands	205	21.1	0	0.0	765	78.9	747	97.7	18	2.4
North Macedonia	1	2.0	16	32.7	32	65.3	31	96.9	1	3.1
Norway	2	1.0	61	31.3	132	67.7	120	90.9	12	9.1
Poland	61	11.5	352	66.2	119	22.4	110	92.4	9	7.6
Portugal	20	10.6	28	14.8	141	74.6	135	95.7	6	4.3
Romania	0	0.0	18	75.0	6	25.0	6	100	0	0.0
Russian Fed.	81	15.9	360	70.6	69	13.5	68	98.6	1	1.5
Serbia	1	1.5	4	6.1	61	92.4	59	96.7	2	3.3
Slovak Republic	9	6.1	56	38.1	82	55.8	80	97.6	2	2.4
Slovenia	7	15.2	23	50.0	16	34.8	14	87.5	2	12.5
Spain	105	8.7	240	19.9	862	71.4	796	92.3	66	7.7
Sweden	0	0.0	0	0.0	389	100	361	92.8	28	7.2
Switzerland	100	18.4	247	45.4	197	36.2	188	95.4	9	4.6
Turkey	9	2.4	189	51.2	171	46.3	160	93.6	11	6.4
Ukraine	5	6.3	66	83.5	8	10.1	2	25.0	6	75.0
United Kingdom	121	2.1	2519	43.5	3146	54.4	2928	93.1	218	6.9
Total	2297	9.2	11052	44.1	11697	46.7	10929	93.4	768	6.6

Note: Albania, Armenia and Georgia have <5 adults seen in 2022 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 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 2012, 2017 and 2022.
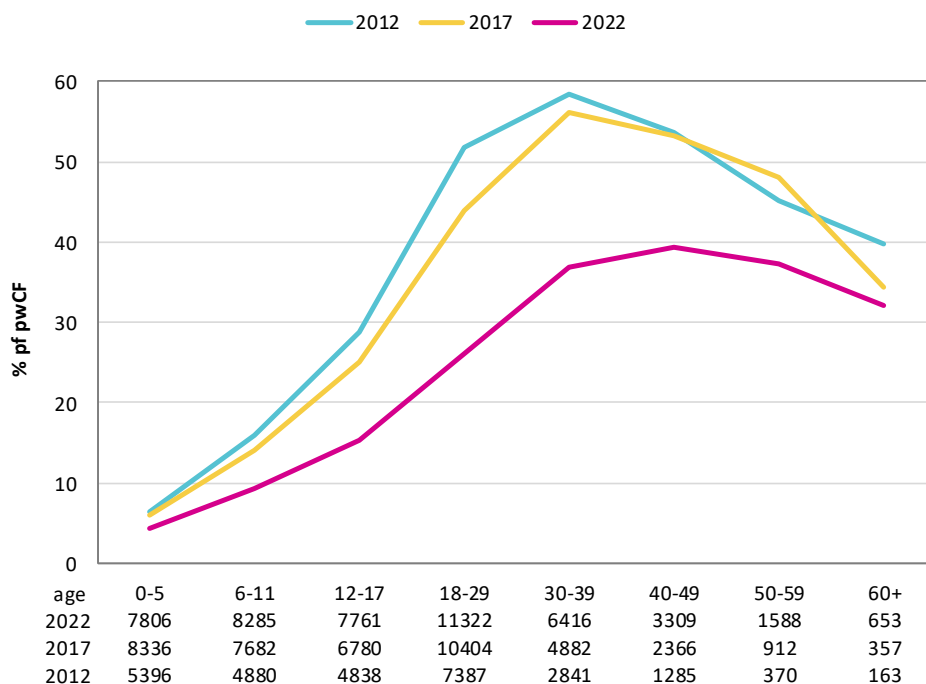
age	0-5	6-11	12-17	18-29	30-39	40-49	50-59	60+
2022	7806	8285	7761	11322	6416	3309	1588	653
2017	8336	7682	6780	10404	4882	2366	912	357
2012	5396	4880	4838	7387	2841	1285	370	163

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. Exclusion criteria were people who were lost to follow-up and/or transplanted individuals (lung and/or liver), and people with missing values.

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 have applied the information on the use of pancreatic enzymes as an indicator of pancreatic insufficiency.

We collected weight and height measured on the date of the 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/m^2 for adults; for older children and adolescents, that they achieve the 50th percentile for BMI; for infants and children up to 2 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 more than 80% of the people with CF are pancreatic insufficient.

Use of pancreatic enzymes in 2022 for all people with CF who have never had a transplant, by country.
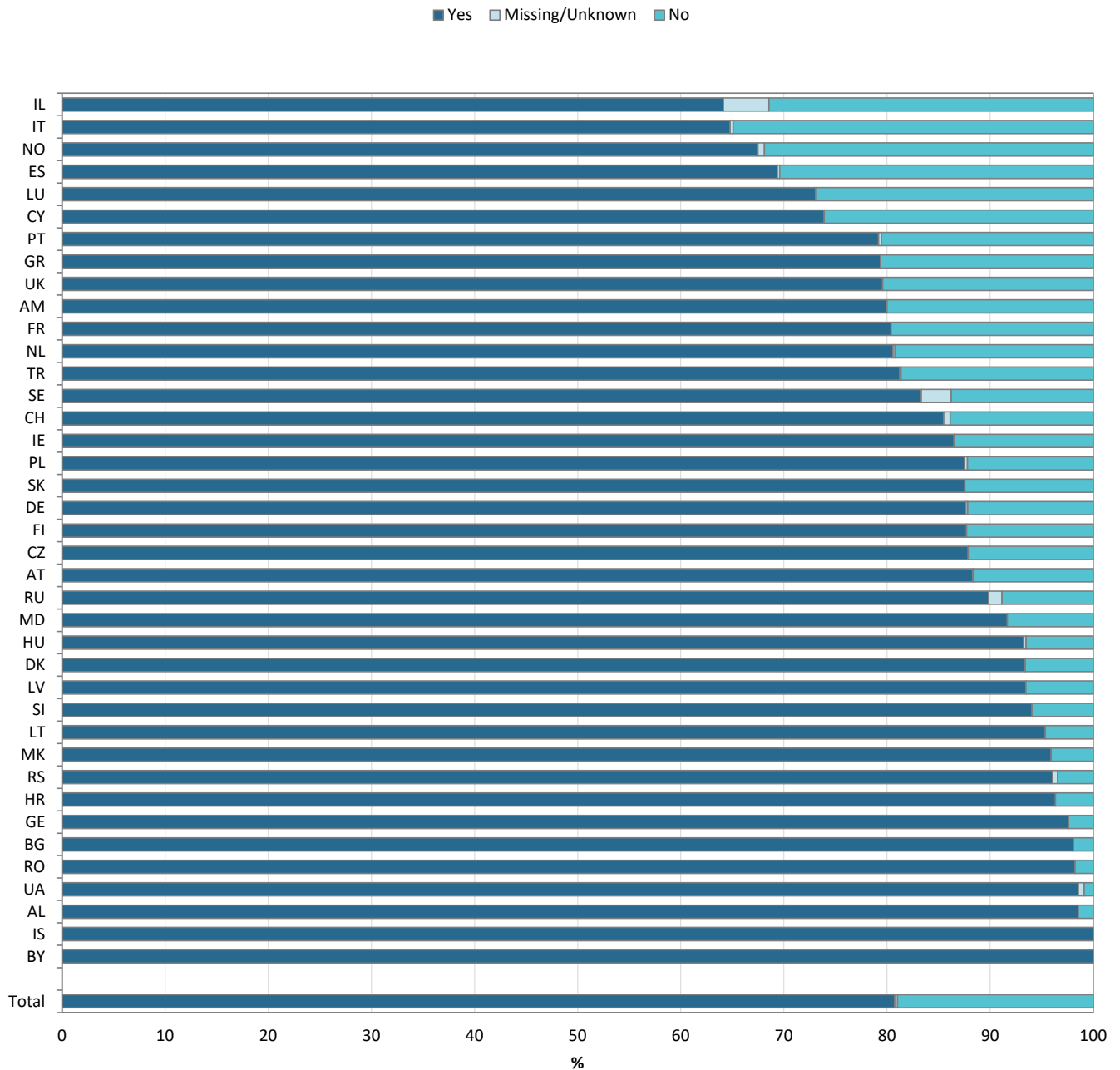
This graph shows the use of pancreatic enzymes by country. This can be seen as an indication of pancreatic insufficiency.

6. Nutrition

Table 6.1   *Z-score for BMI: descriptive statistics by country and overall. All children and adolescents with CF seen in 2022 aged 2-17 years who have never had a transplant.*

Country	Number	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	57	-0.3	-3.6	-0.7	-0.3	0.2	2.5
Armenia	22	-0.8	-7.0	-1.3	-0.6	0.5	2.6
Austria	351	-0.1	-3.7	-0.7	-0.1	0.4	2.1
Belarus	102	-0.7	-5.6	-1.5	-0.5	0.2	2.3
Bulgaria	117	-1.0	-5.2	-1.7	-0.9	0.0	1.7
Croatia	71	-0.6	-5.5	-1.4	-0.3	0.5	2.7
Cyprus	7	-1.5	-6.4	-1.8	-1.5	-0.3	1.7
Czech Republic	252	-0.2	-3.8	-0.8	-0.1	0.5	2.2
Denmark	193	-0.1	-3.1	-0.7	0.0	0.5	2.2
Finland	25	-0.1	-1.7	-0.8	-0.1	0.8	1.3
France	2497	-0.3	-4.9	-0.9	-0.3	0.4	3.5
Germany	2533	-0.2	-16.9	-0.8	-0.2	0.5	3.0
Greece	226	0.2	-5.2	-0.5	0.3	0.9	2.2
Hungary	224	-0.6	-7.0	-1.3	-0.5	0.2	2.4
Iceland	8	0.2	-0.9	0.0	0.4	0.7	0.7
Ireland	501	0.2	-3.4	-0.3	0.3	0.9	2.5
Israel	157	-0.2	-4.0	-0.9	-0.2	0.4	2.4
Italy	2074	0.0	-12	-0.7	0.0	0.7	3.9
Latvia	31	-0.6	-2.5	-1.0	-0.6	0.2	0.8
Lithuania	18	-0.8	-2.5	-1.5	-0.7	-0.2	1.0
Luxembourg	17	-0.2	-1.7	-0.8	0.1	0.3	2.1
Rep of Moldova	27	-1.8	-5.5	-2.8	-1.4	-0.4	0.6
The Netherlands	487	-0.1	-2.6	-0.7	-0.1	0.4	2.0
North Macedonia	70	-0.5	-5.7	-1.1	-0.2	0.3	1.9
Norway	121	-0.1	-2.7	-0.6	0.0	0.5	1.8
Poland	872	-0.3	-4.5	-1.0	-0.2	0.4	2.9
Portugal	159	-0.2	-3.2	-0.8	-0.1	0.4	1.6
Romania	215	-0.7	-5.6	-1.6	-0.5	0.2	2.8
Russian Fed.	1736	-0.7	-9.3	-1.4	-0.6	0.2	2.8
Serbia	125	-0.5	-4.4	-1.2	-0.5	0.3	2.2
Slovak Republic	115	-0.3	-6.1	-0.9	-0.2	0.5	2.3
Slovenia	52	-0.3	-2.3	-0.8	-0.4	0.2	1.7
Spain	990	-0.2	-5.8	-0.7	-0.1	0.5	4.6
Sweden	261	-0.1	-3.4	-0.7	-0.1	0.5	2.7
Switzerland	392	-0.2	-2.9	-0.8	-0.1	0.5	3.2
Turkey	1945	-0.6	-10.1	-1.5	-0.4	0.4	3.9
Ukraine	242	-0.8	-6.4	-1.5	-0.7	0.1	2.3
United Kingdom	3927	0.2	-6.0	-0.4	0.2	0.8	5.3

Note: Georgia has <5 children seen in 2022 with information on height and weight and are excluded from the table, but the people are included in the total number.

Note: 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 FEV1% of predicted. If no lung function measurement is reported, the date of the last visit is 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 2022 aged 18 years or older who have never had a transplant.

Country	Number	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
Austria	385	22.9	14.9	20.5	22.2	24.7	39.0
Bulgaria	82	20.9	15.1	18.4	20.2	22.3	39.8
Croatia	52	22.9	17.0	20.5	22.5	25.0	30.4
Cyprus	15	22.2	18.8	19.4	22.0	24.3	27.0
Czech Republic	273	23.0	14.9	20.6	22.5	24.8	36.1
Denmark	294	23.8	16.5	21.0	23.4	25.7	42.2
Finland	30	23.4	17.1	21.7	23.0	24.5	35.2
France	3556	22.6	12.5	20.2	21.9	24.2	48.1
Germany	3710	22.8	13.7	20.3	22.3	24.6	52.3
Greece	280	23.1	14.8	20.5	22.5	25.2	35.2
Hungary	186	21.2	13.8	18.9	20.8	23.3	31.9
Iceland	6	23.8	18.2	21.7	23.5	27.2	28.7
Ireland	617	24.2	16.9	21.4	23.6	26.2	46.4
Israel	327	23.4	14.6	20.7	22.9	25.6	41.8
Italy	3369	23.1	14.2	20.6	22.5	24.8	53.7
Latvia	14	20.4	15.6	18.0	20.2	22.8	26.6
Lithuania	22	20.4	15.9	18.1	19.8	21.2	27.7
Luxembourg	5	19.2	17.0	18.6	18.8	19.8	22.1
Rep of Moldova	10	18.8	15.8	17.2	18.4	20.8	23.7
The Netherlands	933	23.2	13.7	20.9	22.7	24.7	47.3
North Macedonia	45	22.5	16.4	20.4	22.5	24.9	32.1
Norway	187	23.6	16.0	20.7	22.9	25.6	40.8
Poland	494	21.9	12.9	19.5	21.5	23.7	38.6
Portugal	173	23.1	17.2	20.8	22.4	24.4	45.8
Romania	14	20.9	13.4	18.3	20.9	22.8	28.9
Russian Fed.	442	20.0	12.9	17.8	19.4	21.6	39.0
Serbia	60	20.2	15.2	18.5	20.1	22.1	26.2
Slovak Republic	139	21.9	15.0	19.6	21.2	23.8	33.8
Slovenia	44	21.2	15.4	18.8	21.4	23.0	27.0
Spain	1152	23.2	14.8	20.7	22.7	25.0	49.6
Sweden	369	23.0	14.9	21.0	22.5	24.8	41.7
Switzerland	521	22.4	14.7	20.2	21.9	24.1	40.7
Turkey	340	21.1	12.6	18.4	20.8	23.6	33.0
Ukraine	72	19.8	13.6	18.0	19.9	21.4	28.3
United Kingdom	5322	24.5	13.6	21.5	23.8	26.6	57.6
Total	23545	23.1	12.5	20.5	22.6	25.0	57.6

Note: Albania, Armenia, Belarus and Georgia have <5 adults seen in 2022 with information on height and weight and are excluded from the table, but the people are included in the total number.

Note: 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 $FEV1_{\%}$ of predicted. 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 2022 who have never had a transplant.
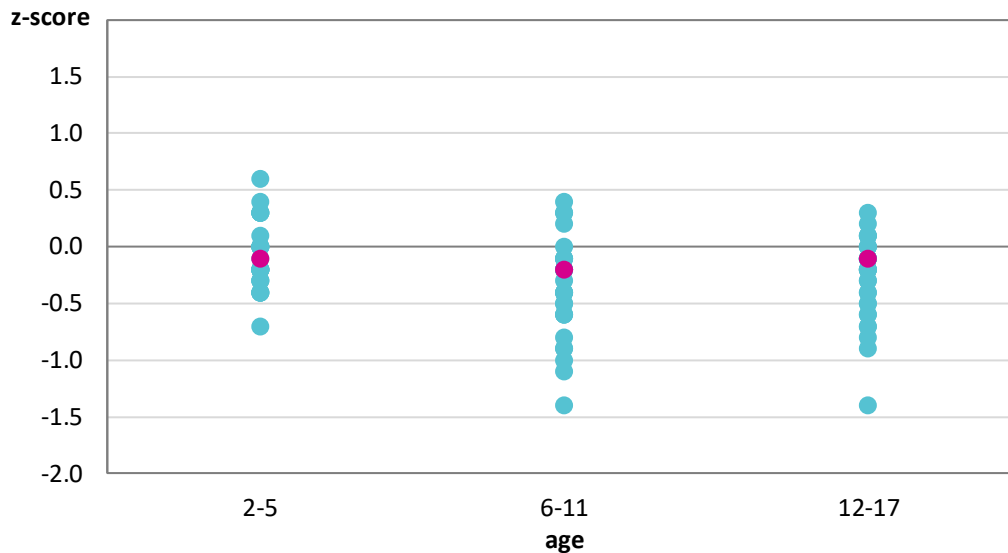
Note: We excluded from the graph those age groups where the number of individuals was <10.

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 2022 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, Georgia, 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
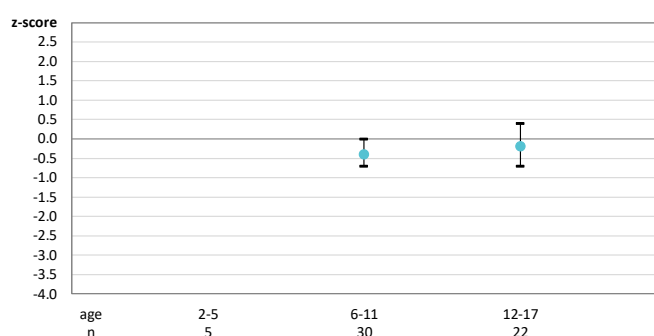
Quartiles of z-scores for BMI: Armenia
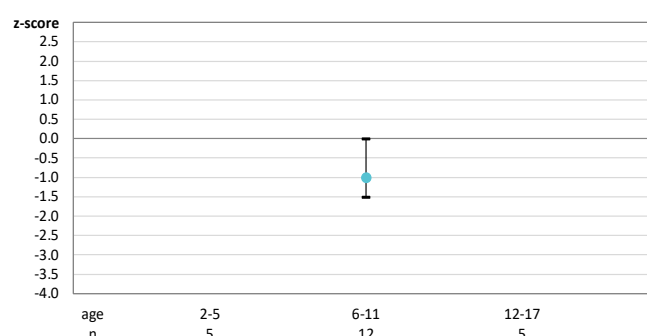
Quartiles of z-scores for BMI: Austria
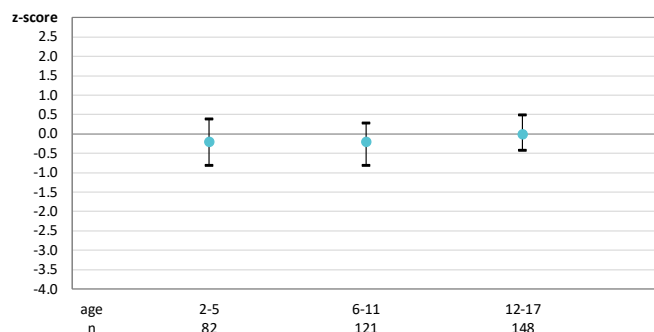
Quartiles of z-scores for BMI: Belarus
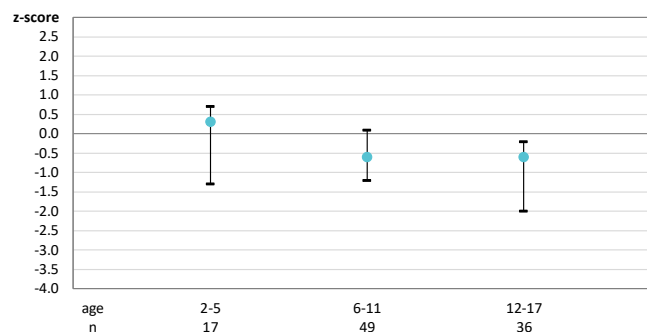
Quartiles of z-scores for BMI: Bulgaria
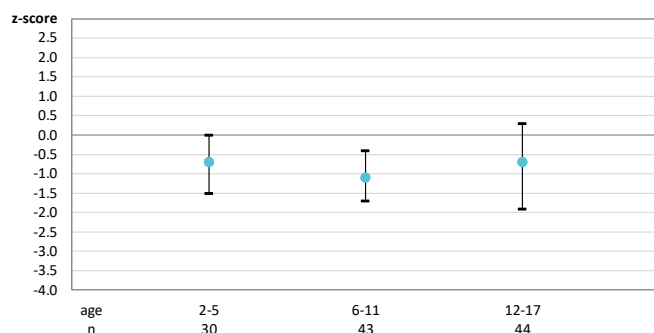
Quartiles of z-scores for BMI: Croatia
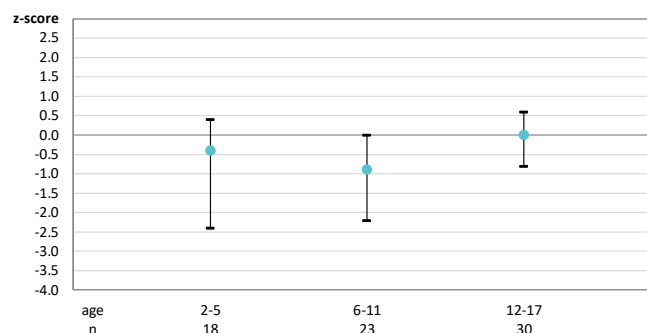
6. Nutrition

[figure 6.3 continued]

Quartiles of z-scores for BMI: Czech Republic

age	2-5	6-11	12-17
n	50	93	109
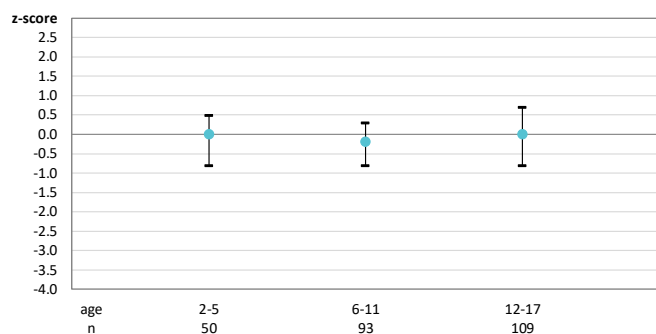
Quartiles of z-scores for BMI: Denmark

age	2-5	6-11	12-17
n	61	64	68
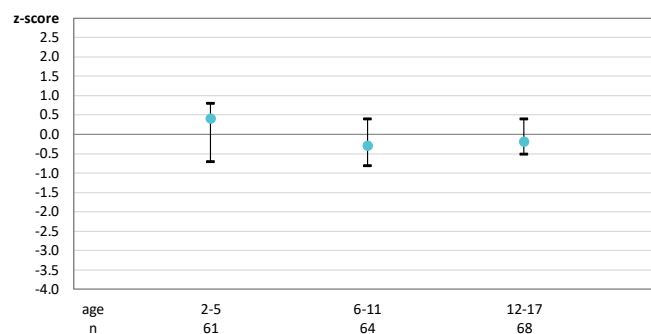
Quartiles of z-scores for BMI: Finland

age	2-5	6-11	12-17
n	4	8	13

Quartiles of z-scores for BMI: France

age	2-5	6-11	12-17
n	516	902	1079
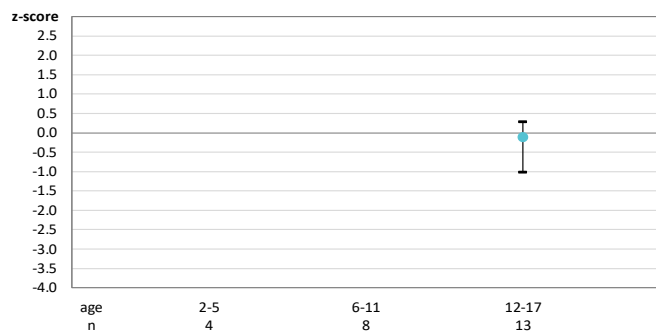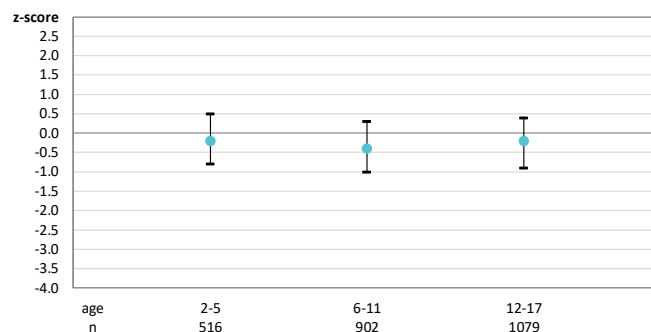
Quartiles of z-scores for BMI: Germany

age	2-5	6-11	12-17
n	647	966	920
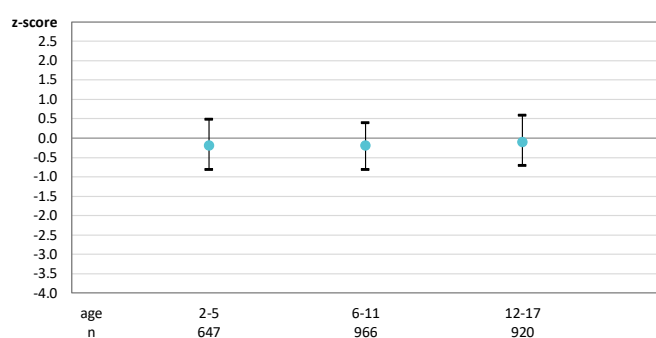
Quartiles of z-scores for BMI: Greece

age	2-5	6-11	12-17
n	37	76	113
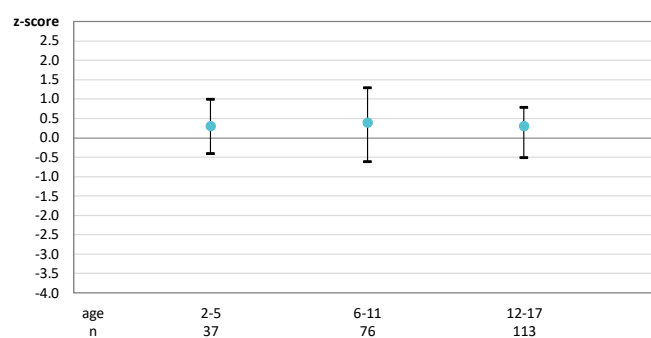
Quartiles of z-scores for BMI: Hungary

age	2-5	6-11	12-17
n	51	97	76
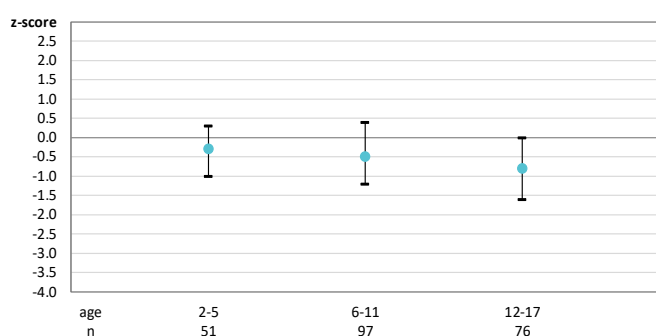
Quartiles of z-scores for BMI: Ireland

age	2-5	6-11	12-17
n	110	194	197
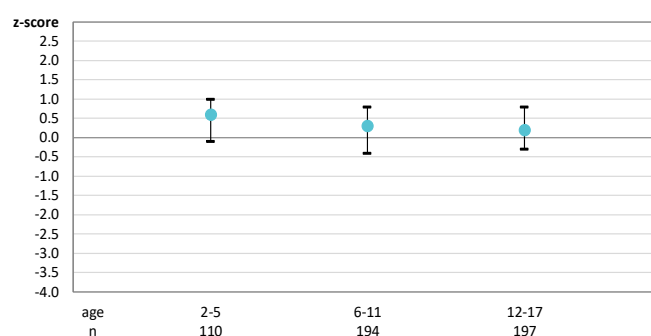
6. Nutrition

[figure 6.3 continued]

Quartiles of z-scores for BMI: Israel

age	2-5	6-11	12-17
n	17	56	84

Quartiles of z-scores for BMI: Italy

age	2-5	6-11	12-17
n	454	824	796
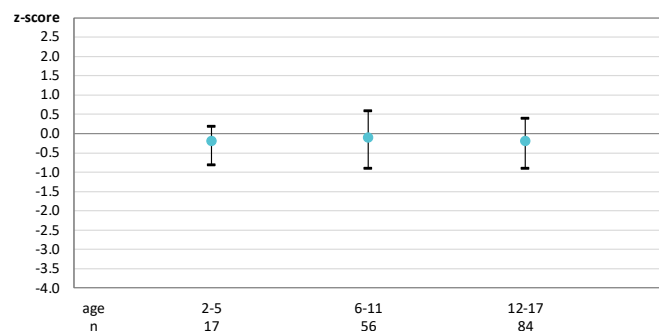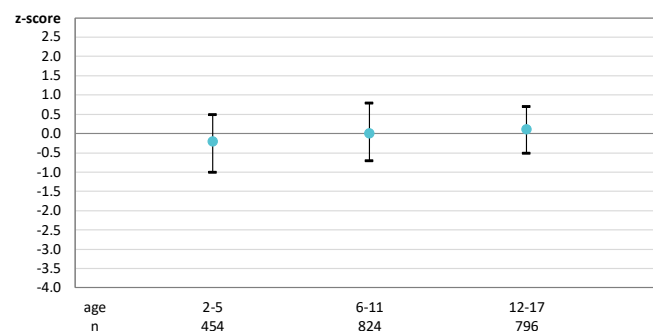
Quartiles of z-scores for BMI: Latvia

age	2-5	6-11	12-17
n	12	9	10
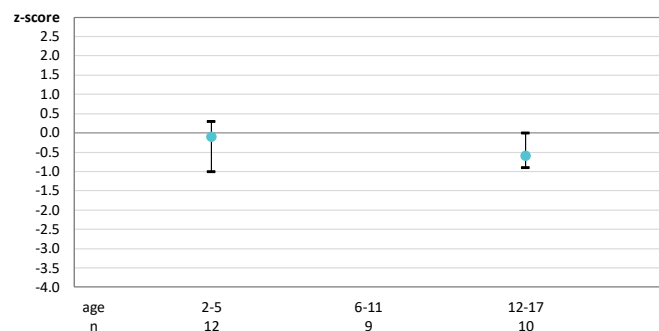
Quartiles of z-scores for BMI: Rep. of Moldova

age	2-5	6-11	12-17
n	3	13	11
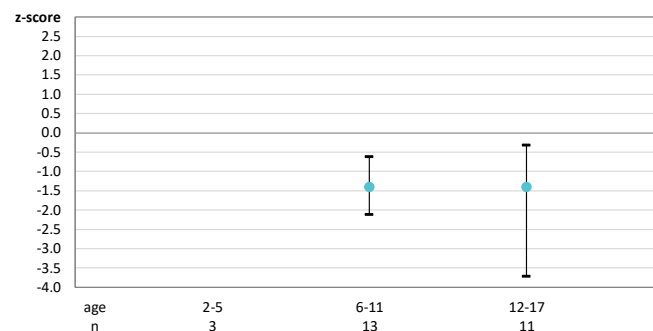
Quartiles of z-scores for BMI: The Netherlands

age	2-5	6-11	12-17
n	95	190	202
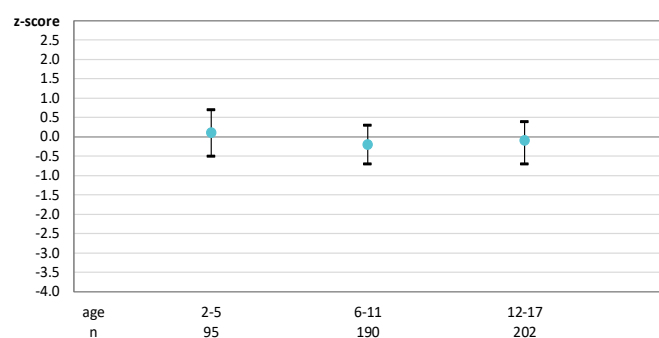
Quartiles of z-scores for BMI: North Macedonia

age	2-5	6-11	12-17
n	26	15	29
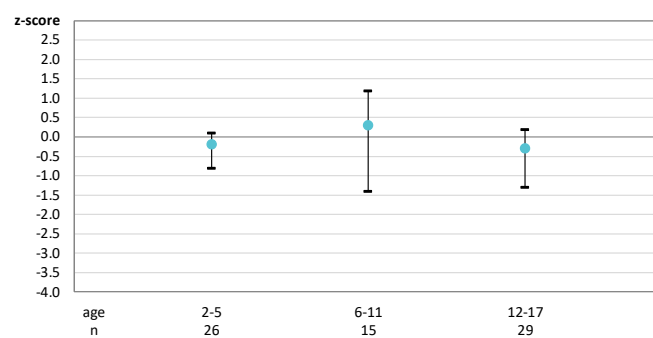
Quartiles of z-scores for BMI: Norway
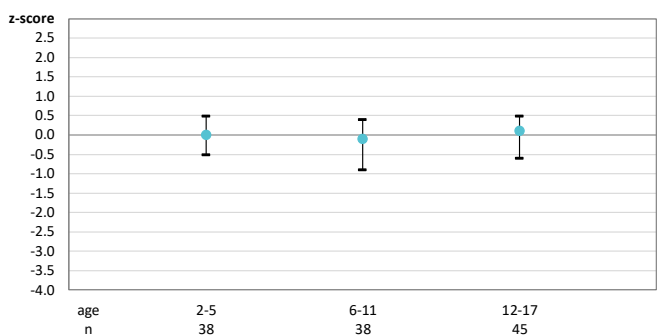
age	2-5	6-11	12-17
n	38	38	45

Quartiles of z-scores for BMI: Poland

age	2-5	6-11	12-17
n	203	331	338
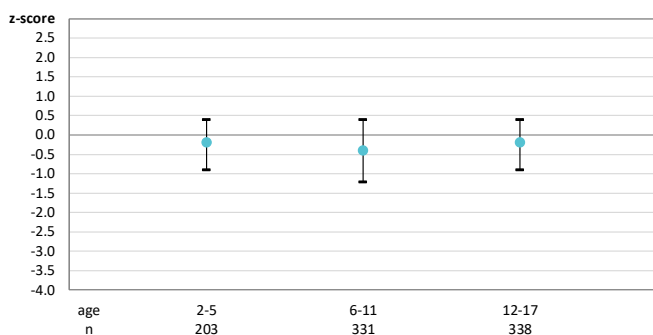
6. Nutrition

[figure 6.3 continued]

Quartiles of z-scores for BMI: Portugal

age	2-5	6-11	12-17
n	33	63	63
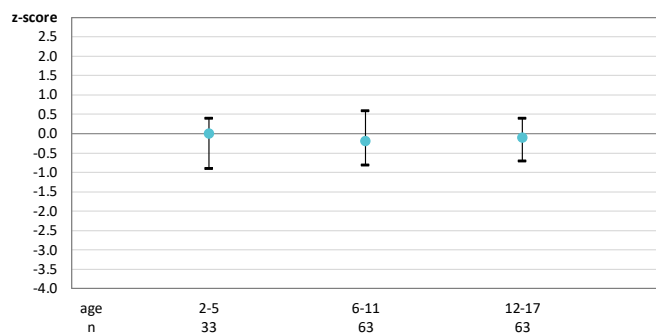
Quartiles of z-scores for BMI: Romania

age	2-5	6-11	12-17
n	49	73	93
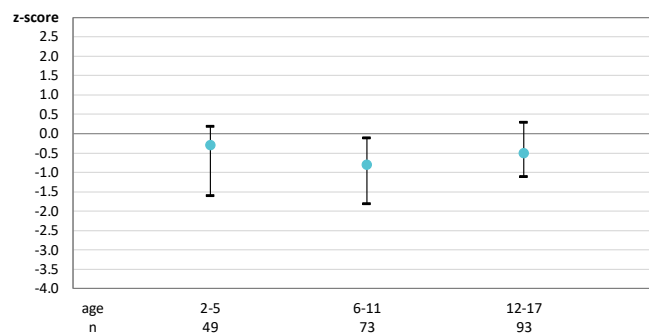
Quartiles of z-scores for BMI: Russian federation

age	2-5	6-11	12-17
n	371	786	579
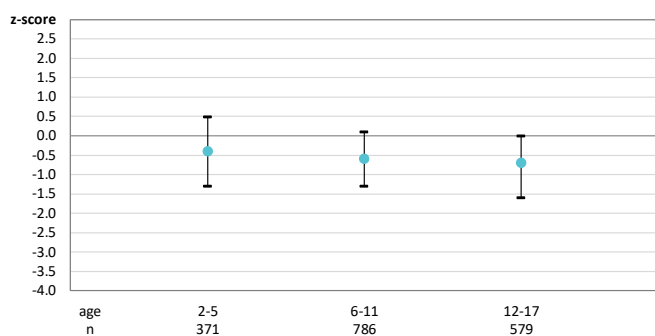
Quartiles of z-scores for BMI: Serbia

age	2-5	6-11	12-17
n	44	35	46
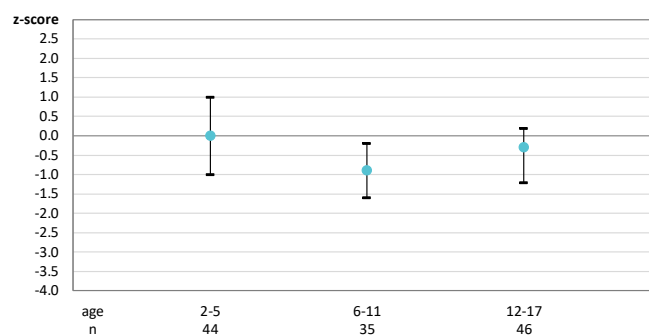
Quartiles of z-scores for BMI: Slovakia

age	2-5	6-11	12-17
n	23	51	41
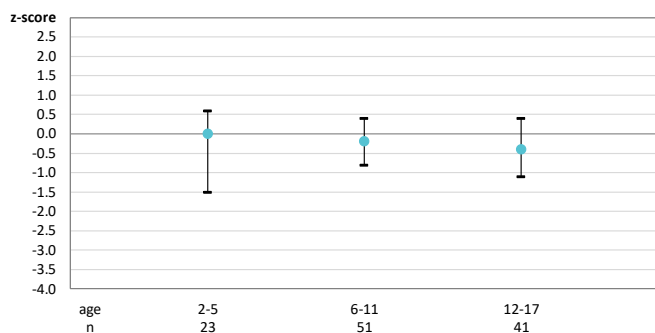
Quartiles of z-scores for BMI: Slovenia

age	2-5	6-11	12-17
n	5	28	19

Quartiles of z-scores for BMI: Spain

age	2-5	6-11	12-17
n	198	393	399
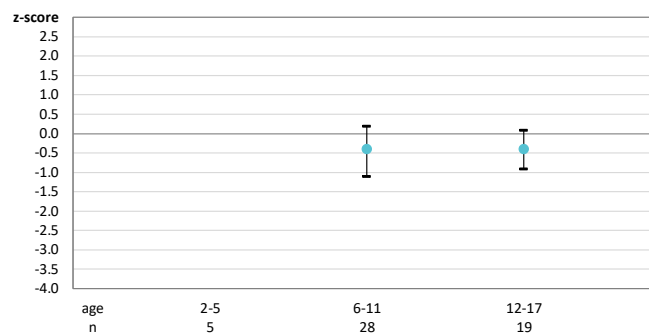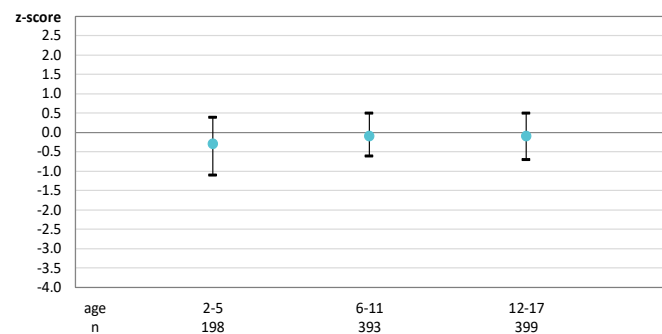
Quartiles of z-scores for BMI: Sweden

age	2-5	6-11	12-17
n	54	90	117
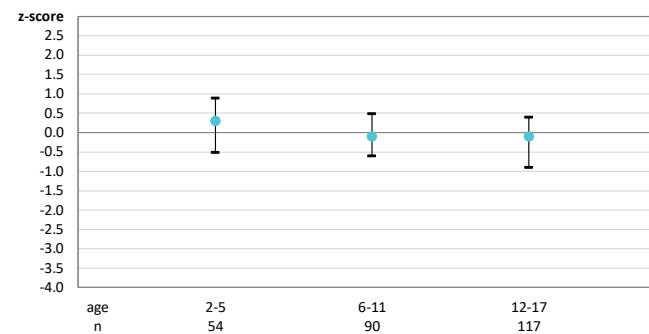
6. Nutrition

[figure 6.3 continued]

Quartiles of z-scores for BMI: Switzerland

age	2-5	6-11	12-17
n	90	155	147

Quartiles of z-scores for BMI: Turkey

age	2-5	6-11	12-17
n	595	790	560
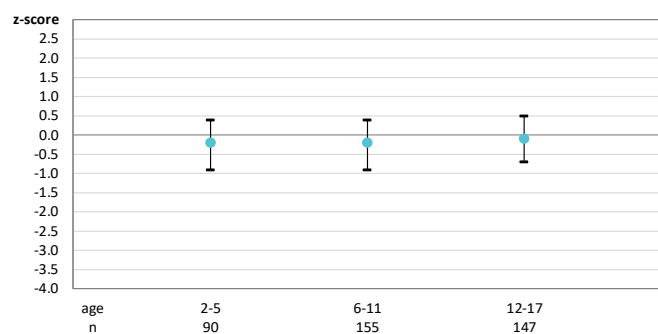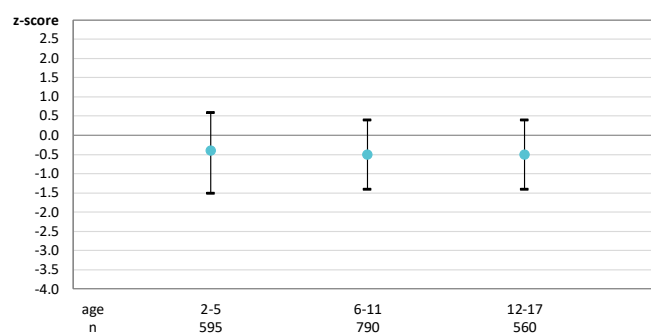
Quartiles of z-scores for BMI: Ukraine

age	2-5	6-11	12-17
n	54	107	81
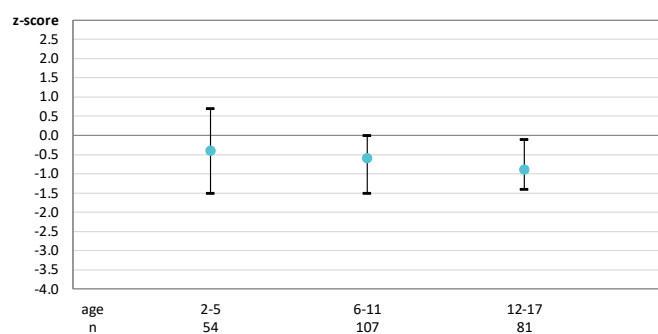
Quartiles of z-scores for BMI: United Kingdom

age	2-5	6-11	12-17
n	918	1583	1426
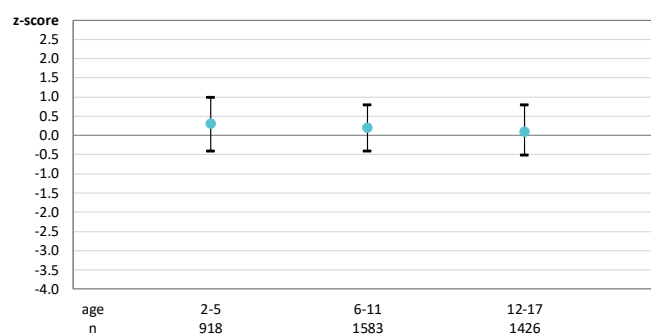
6. Nutrition

Figure 6.4   Being underweight is a hallmark 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 2022 who have never had a transplant.
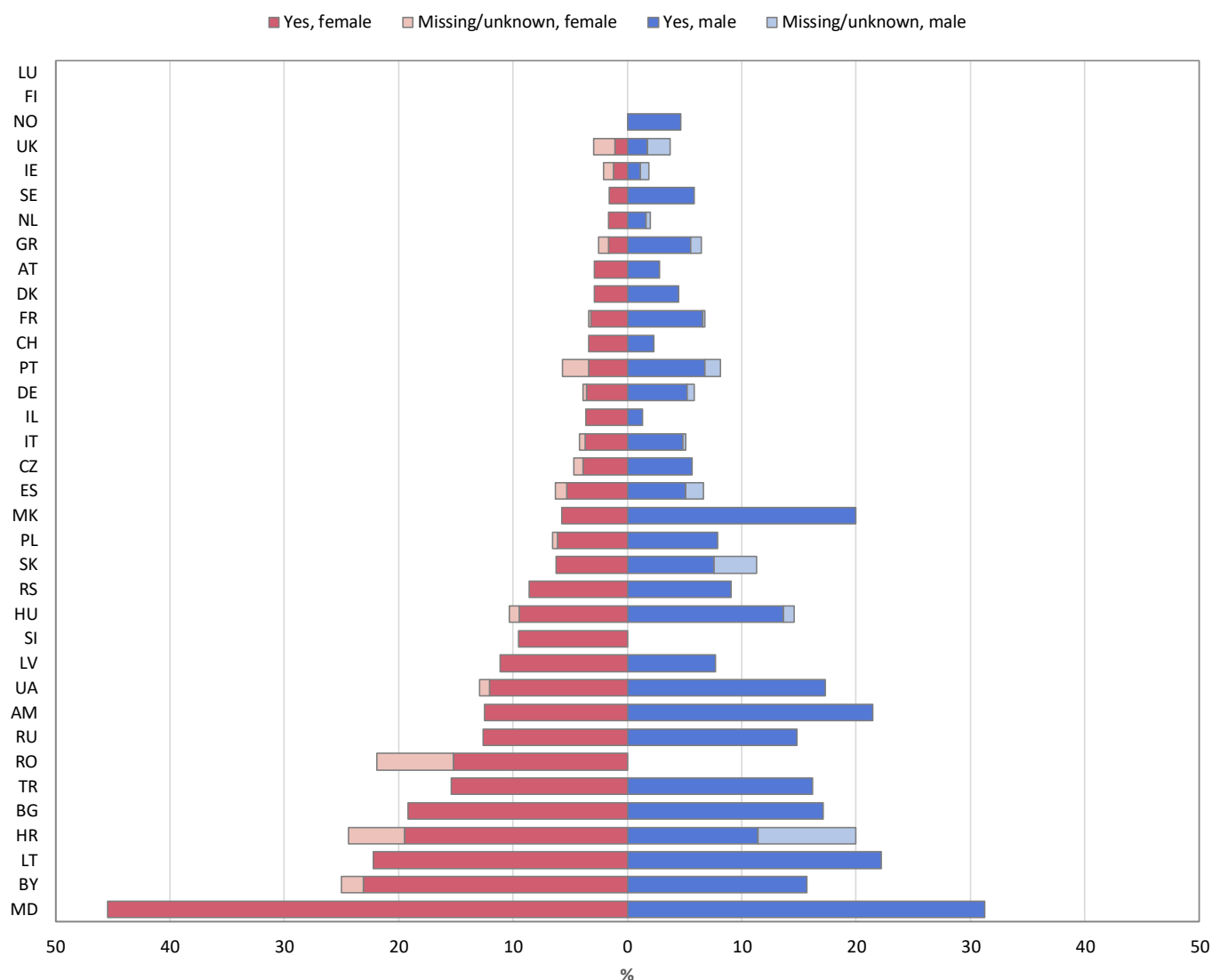
Note: We excluded from the graph the countries for which the information on underweight children and young people is missing for more than 10% of the individuals.

Cyprus, Georgia 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.

6. Nutrition

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

Proportion of adults with BMI <20 by sex and by country; aged 18 years or older in 2022 who never had a transplant.
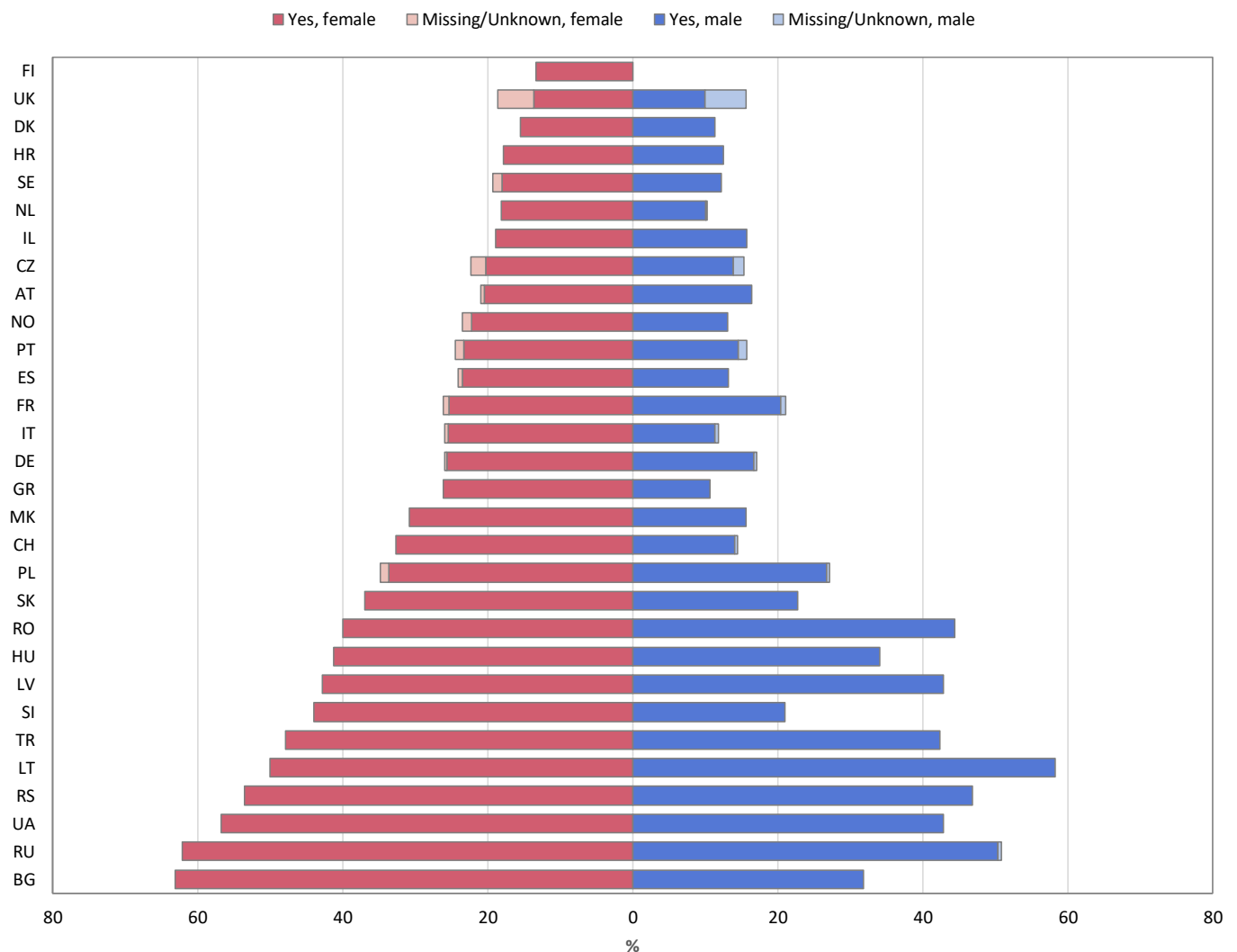
Note: We excluded from the graph the countries for which the information on underweight adults is missing for more than 10% of the individuals.

Albania, Armenia, Belarus, Cyprus, Iceland, Luxembourg and Republic of Moldova 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.

6. Nutrition

Figure 6.6   A significant improvement in BMI in 2022 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 2012, 2017 and 2022.

age	2-5	6-11	12-17	18-29	30-39	40-49	50-59	60+
2022	4933	8328	7962	11375	6516	3379	1623	668
2017	5213	7756	6952	10502	5039	2408	940	361
2012	3567	5560	5716	8590	3422	1587	422	179

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. Exclusion criteria were people who were lost to follow-up, and transplanted individuals (lung and/or liver). Also, people with missing values are excluded when computing the yearly prevalence for each variable.

7. Complications

Common respiratory complications in CF include allergic bronchopulmonary aspergillosis, haemoptysis and pneumothorax. For pulmonary exacerbations, we present data on the use of intravenous antibiotics, considered a surrogate marker. 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 is 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 is also reported in this section.

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 is 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 2022, 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	1	1.5	42	62.7	24	35.8						
Armenia	0	0.0	16	72.7	6	27.3						
Austria	0	0.0	291	78.4	80	21.6	0	0.0	333	83.5	66	16.5
Belarus	0	0.0	73	50.0	73	50.0	0	0.0	3	60.0	2	40.0
Bulgaria	6	4.8	88	69.8	32	25.4	1	1.2	72	84.7	12	14.1
Croatia	0	0.0	68	82.9	14	17.1	1	1.8	46	83.6	8	14.6
Cyprus	0	0.0	6	85.7	1	14.3	0	0.0	12	75.0	4	25.0
Czech Republic	10	3.1	291	89.8	23	7.1	20	6.6	247	81.8	35	11.6
Denmark	0	0.0	193	91.5	18	8.5	0	0.0	226	74.1	79	25.9
Finland	0	0.0	24	92.3	2	7.7	0	0.0	25	80.7	6	19.4
France	3	0.1	2428	90.2	260	9.7	20	0.5	3144	85.3	520	14.1
Georgia	7	17.1	22	53.7	12	29.3						
Germany	3	0.1	2615	94.9	138	5.0	10	0.3	3409	89.3	397	10.4
Greece	1	0.4	199	84.0	37	15.6	6	1.7	286	80.8	62	17.5
Hungary	237	100	-	-	-	-	194	100	-	-	-	-
Iceland	0	0.0	7	77.8	2	22.2	0	0.0	4	66.7	2	33.3
Ireland	0	0.0	489	93.1	36	6.9	0	0.0	599	83.3	120	16.7
Israel	0	0.0	136	85.5	23	14.5	0	0.0	252	74.1	88	25.9
Italy	176	8.0	1631	74.0	397	18.0	485	13.6	2497	69.9	591	16.5
Latvia	0	0.0	25	78.1	7	21.9	0	0.0	7	50.0	7	50.0
Lithuania	3	15.0	6	30.0	11	55.0	0	0.0	7	30.4	16	69.6
Luxembourg	1	4.8	17	81.0	3	14.3	0	0.0	2	40.0	3	60.0
Rep of Moldova	1	2.8	17	47.2	18	50.0	0	0.0	0	0.0	12	100
The Netherlands	4	0.7	484	89.8	51	9.5	19	2.0	842	86.8	109	11.2
North Macedonia	1	1.4	26	35.1	47	63.5	0	0.0	13	26.5	36	73.5
Norway	2	1.6	110	85.9	16	12.5	3	1.5	134	68.7	58	29.7
Poland	11	1.2	682	71.6	260	27.3	21	4.0	276	51.9	235	44.2
Portugal	0	0.0	159	93.0	12	7.0	2	1.1	171	90.5	16	8.5
Romania	10	3.8	160	61.1	92	35.1	0	0.0	14	58.3	10	41.7
Russian Fed.	49	2.6	1055	56.3	770	41.1	53	10.4	142	27.8	315	61.8
Serbia	0	0.0	108	79.4	28	20.6	1	1.5	46	69.7	19	28.8
Slovak Republic	0	0.0	108	85.7	18	14.3	0	0.0	123	83.7	24	16.3
Slovenia	0	0.0	40	72.7	15	27.3	5	10.9	39	84.8	2	4.4
Spain	26	2.5	961	90.8	72	6.8	3	0.3	1077	89.2	127	10.5
Sweden	15	5.7	198	74.7	52	19.6	7	1.8	182	46.8	200	51.4
Switzerland	56	13.8	338	83.1	13	3.2	16	2.9	465	85.5	63	11.6
Turkey	0	0.0	1693	79.1	448	20.9	1	0.3	253	68.6	115	31.2
Ukraine	4	1.5	88	33.2	173	65.3	1	1.3	21	26.6	57	72.2
United Kingdom	0	0.0	3366	81.3	772	18.7	0	0.0	4368	75.5	1418	24.5
Total	627	2.7	18260	79.6	4056	17.7	870	3.5	19340	77.2	4836	19.3

Note: Albania, Armenia and Georgia have <5 adults seen in 2022 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 2022, 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	1	1.5	42	62.7	24	35.8						
Armenia	0	0.0	16	72.7	6	27.3						
Austria	0	0.0	291	78.4	80	21.6	0	0.0	340	85.2	59	14.8
Belarus	0	0.0	73	50.0	73	50.0	0	0.0	3	60.0	2	40.0
Bulgaria	6	4.8	88	69.8	32	25.4	1	1.2	74	87.1	10	11.8
Croatia	0	0.0	68	82.9	14	17.1	1	1.8	46	83.6	8	14.6
Cyprus	0	0.0	6	85.7	1	14.3	0	0.0	12	75.0	4	25.0
Czech Republic	10	3.1	291	89.8	23	7.1	20	6.6	248	82.1	34	11.3
Denmark	181	85.8	30	14.2	0	0.0	305	100	-	-	-	-
Finland	0	0.0	24	92.3	2	7.7	0	0.0	25	80.7	6	19.4
France	50	1.9	2441	90.7	200	7.4	268	7.3	3196	86.8	220	6.0
Georgia	7	17.1	23	56.1	11	26.8						
Germany	8	0.3	2625	95.3	123	4.5	14	0.4	3517	92.2	285	7.5
Greece	1	0.4	202	85.2	34	14.4	6	1.7	296	83.6	52	14.7
Hungary	157	66.2	0	0.0	80	33.8	110	56.7	0	0.0	84	43.3
Iceland	0	0.0	7	77.8	2	22.2	0	0.0	4	66.7	2	33.3
Ireland	0	0.0	493	93.9	32	6.1	0	0.0	612	85.1	107	14.9
Israel	0	0.0	142	89.3	17	10.7	0	0.0	301	88.5	39	11.5
Italy	172	7.8	1651	74.9	381	17.3	485	13.6	2525	70.7	563	15.8
Latvia	0	0.0	25	78.1	7	21.9	1	7.1	7	50.0	6	42.9
Lithuania	3	15.0	6	30.0	11	55.0	0	0.0	7	30.4	16	69.6
Luxembourg	1	4.8	18	85.7	2	9.5	0	0.0	2	40.0	3	60.0
Rep of Moldova	1	2.8	18	50.0	17	47.2	0	0.0	6	50.0	6	50.0
The Netherlands	3	0.6	491	91.1	45	8.4	19	2.0	868	89.5	83	8.6
North Macedonia	1	1.4	26	35.1	47	63.5	0	0.0	13	26.5	36	73.5
Norway	2	1.6	110	85.9	16	12.5	3	1.5	164	84.1	28	14.4
Poland	12	1.3	681	71.5	260	27.3	20	3.8	277	52.1	235	44.2
Portugal	0	0.0	159	93.0	12	7.0	2	1.1	172	91.0	15	7.9
Romania	9	3.4	161	61.5	92	35.1	0	0.0	14	58.3	10	41.7
Russian Fed.	38	2.0	1096	58.5	740	39.5	32	6.3	217	42.6	261	51.2
Serbia	0	0.0	108	79.4	28	20.6	1	1.5	46	69.7	19	28.8
Slovak Republic	0	0.0	108	85.7	18	14.3	0	0.0	124	84.4	23	15.7
Slovenia	0	0.0	40	72.7	15	27.3	5	10.9	39	84.8	2	4.4
Spain	26	2.5	969	91.5	64	6.0	3	0.3	1119	92.7	85	7.0
Sweden	15	5.7	237	89.4	13	4.9	8	2.1	357	91.8	24	6.2
Switzerland	53	13.0	341	83.8	13	3.2	15	2.8	486	89.3	43	7.9
Turkey	0	0.0	1697	79.3	444	20.7	1	0.3	254	68.8	114	30.9
Ukraine	3	1.1	90	34.0	172	64.9	0	0.0	26	32.9	53	67.1
United Kingdom	0	0.0	3426	82.8	712	17.2	0	0.0	4680	80.9	1106	19.1
Total	760	3.3	18320	79.9	3863	16.8	1321	5.3	20080	80.2	3645	14.6

Note: Albania, Armenia and Georgia have <5 adults seen in 2022 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 2022, 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	1	1.5	42	62.7	24	35.8						
Armenia	0	0.0	12	54.6	10	45.5						
Austria	0	0.0	252	67.9	119	32.1	0	0.0	328	82.2	71	17.8
Belarus	0	0.0	73	50.0	73	50.0	0	0.0	3	60.0	2	40.0
Bulgaria	6	4.8	33	26.2	87	69.1	1	1.2	64	75.3	20	23.5
Croatia	0	0.0	56	68.3	26	31.7	1	1.8	44	80.0	10	18.2
Cyprus	0	0.0	6	85.7	1	14.3	0	0.0	12	75.0	4	25.0
Czech Republic	8	2.5	267	82.4	49	15.1	20	6.6	240	79.5	42	13.9
Denmark	153	72.5	53	25.1	5	2.4	304	99.7	1	0.3	0	0.0
Finland	0	0.0	24	92.3	2	7.7	0	0.0	24	77.4	7	22.6
France	180	6.7	2069	76.9	442	16.4	266	7.2	2906	78.9	512	13.9
Georgia	7	17.1	23	56.1	11	26.8						
Germany	60	2.2	2119	76.9	577	20.9	47	1.2	3043	79.7	726	19.0
Greece	1	0.4	198	83.5	38	16.0	7	2.0	285	80.5	62	17.5
Hungary	142	59.9	0	0.0	95	40.1	101	52.1	0	0.0	93	47.9
Iceland	0	0.0	6	66.7	3	33.3	0	0.0	4	66.7	2	33.3
Ireland	0	0.0	475	90.5	50	9.5	0	0.0	587	81.6	132	18.4
Israel	0	0.0	134	84.3	25	15.7	0	0.0	268	78.8	72	21.2
Italy	0	0.0	1513	68.7	691	31.4	3	0.1	2640	73.9	930	26.0
Latvia	0	0.0	25	78.1	7	21.9	1	7.1	7	50.0	6	42.9
Lithuania	3	15.0	4	20.0	13	65.0	0	0.0	7	30.4	16	69.6
Luxembourg	1	4.8	17	81.0	3	14.3	0	0.0	1	20.0	4	80.0
Rep of Moldova	1	2.8	16	44.4	19	52.8	0	0.0	6	50.0	6	50.0
The Netherlands	3	0.6	463	85.9	73	13.5	18	1.9	783	80.7	169	17.4
North Macedonia	1	1.4	25	33.8	48	64.9	0	0.0	13	26.5	36	73.5
Norway	3	2.3	100	78.1	25	19.5	4	2.1	135	69.2	56	28.7
Poland	10	1.1	301	31.6	642	67.4	21	4.0	211	39.7	300	56.4
Portugal	0	0.0	155	90.6	16	9.4	2	1.1	170	90.0	17	9.0
Romania	0	0.0	72	27.5	190	72.5	0	0.0	8	33.3	16	66.7
Russian Fed.	35	1.9	1032	55.1	807	43.1	30	5.9	212	41.6	268	52.6
Serbia	0	0.0	106	77.9	30	22.1	1	1.5	46	69.7	19	28.8
Slovak Republic	0	0.0	95	75.4	31	24.6	0	0.0	122	83.0	25	17.0
Slovenia	0	0.0	34	61.8	21	38.2	5	10.9	38	82.6	3	6.5
Spain	26	2.5	930	87.8	103	9.7	5	0.4	1104	91.5	98	8.1
Sweden	0	0.0	215	81.1	50	18.9	0	0.0	332	85.4	57	14.7
Switzerland	50	12.3	327	80.3	30	7.4	16	2.9	465	85.5	63	11.6
Turkey	0	0.0	1529	71.4	612	28.6	1	0.3	234	63.4	134	36.3
Ukraine	2	0.8	82	30.9	181	68.3	0	0.0	24	30.4	55	69.6
United Kingdom	0	0.0	2943	71.1	1195	28.9	0	0.0	4403	76.1	1383	23.9
Total	693	3.0	15826	69.0	6424	28.0	855	3.4	18773	75.0	5418	21.6

Note: Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the table for adults, but the people are included in the total number.

7. Complications

Figure 7.1   ABPA may be difficult to diagnose and report. In the ECFSPR the registered ABPA prevalence is lower in children than in adults..

Prevalence of allergic bronchopulmonary aspergillosis in children and adults seen in 2022 who have never had a transplant, by country (table A7.1, Appendix 1).
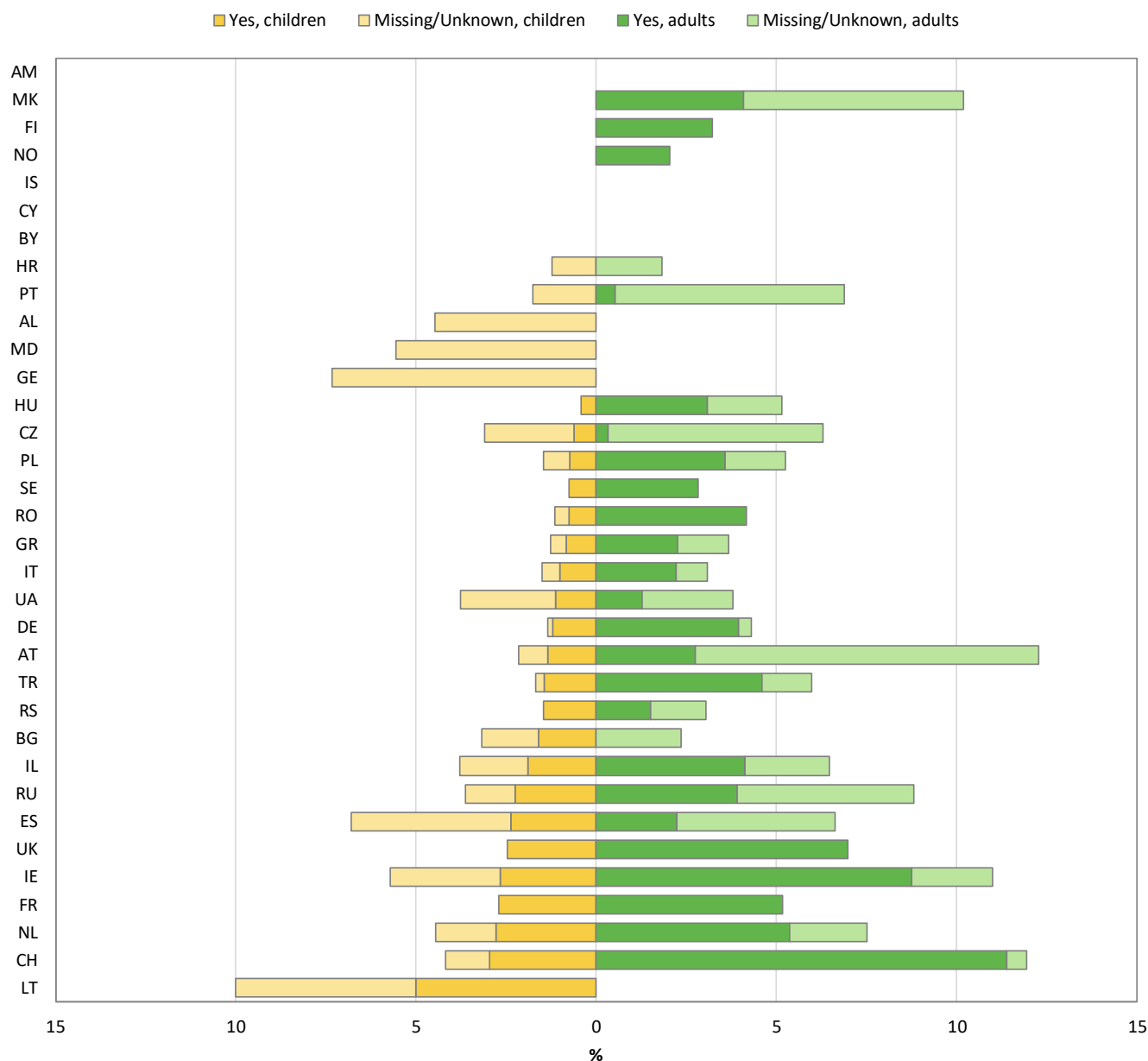
Note: We excluded from the graph the countries for which the information on allergic bronchopulmonary aspergillosis (ABPA) is missing for more than 10% of the children/adults with CF.

Albania, Armenia and Georgia have <5 adults seen in 2022 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.

7. Complications

Table 7.4   Prevalence of pneumothorax in all people with CF seen in 2022 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	3	4.5	64	95.5	0	0.0						
Armenia	0	0.0	22	100	0	0.0						
Austria	4	1.1	367	98.9	0	0.0	4	1.0	395	99.0	0	0.0
Belarus	0	0.0	145	99.3	1	0.7	0	0.0	5	100	0	0.0
Bulgaria	3	2.4	123	97.6	0	0.0	2	2.4	83	97.7	0	0.0
Croatia	1	1.2	81	98.8	0	0.0	0	0.0	55	100	0	0.0
Cyprus	0	0.0	7	100	0	0.0	0	0.0	16	100	0	0.0
Czech Republic	16	4.9	308	95.1	0	0.0	23	7.6	278	92.1	1	0.3
Denmark	0	0.0	211	100	0	0.0	0	0.0	305	100	0	0.0
Finland	0	0.0	26	100	0	0.0	0	0.0	31	100	0	0.0
France	0	0.0	2690	100	1	0.0	0	0.0	3678	99.8	6	0.2
Georgia	3	7.3	38	92.7	0	0.0						
Germany	4	0.2	2751	99.8	1	0.0	13	0.3	3787	99.2	16	0.4
Greece	0	0.0	237	100	0	0.0	0	0.0	354	100	0	0.0
Hungary	2	0.8	235	99.2	0	0.0	4	2.1	188	96.9	2	1.0
Iceland	0	0.0	9	100	0	0.0	0	0.0	6	100	0	0.0
Ireland	9	1.7	516	98.3	0	0.0	8	1.1	711	98.9	0	0.0
Israel	3	1.9	156	98.1	0	0.0	10	2.9	330	97.1	0	0.0
Italy	17	0.8	2186	99.2	1	0.1	38	1.1	3530	98.8	5	0.1
Latvia	0	0.0	32	100	0	0.0	1	7.1	13	92.9	0	0.0
Lithuania	0	0.0	20	100	0	0.0	0	0.0	21	91.3	2	8.7
Luxembourg	0	0.0	21	100	0	0.0	0	0.0	5	100	0	0.0
Rep of Moldova	0	0.0	36	100	0	0.0	0	0.0	12	100	0	0.0
The Netherlands	7	1.3	532	98.7	0	0.0	25	2.6	942	97.1	3	0.3
North Macedonia	0	0.0	74	100	0	0.0	0	0.0	49	100	0	0.0
Norway	2	1.6	126	98.4	0	0.0	1	0.5	194	99.5	0	0.0
Poland	10	1.1	943	99.0	0	0.0	11	2.1	518	97.4	3	0.6
Portugal	0	0.0	171	100	0	0.0	5	2.7	184	97.4	0	0.0
Romania	5	1.9	256	97.7	1	0.4	0	0.0	23	95.8	1	4.2
Russian Fed.	2	0.1	1864	99.5	8	0.4	21	4.1	480	94.1	9	1.8
Serbia	1	0.7	135	99.3	0	0.0	1	1.5	65	98.5	0	0.0
Slovak Republic	0	0.0	125	99.2	1	0.8	5	3.4	141	95.9	1	0.7
Slovenia	0	0.0	55	100	0	0.0	4	8.7	42	91.3	0	0.0
Spain	13	1.2	1046	98.8	0	0.0	28	2.3	1178	97.6	1	0.1
Sweden	15	5.7	250	94.3	0	0.0	4	1.0	385	99.0	0	0.0
Switzerland	6	1.5	401	98.5	0	0.0	6	1.1	538	98.9	0	0.0
Turkey	1	0.1	2136	99.8	4	0.2	1	0.3	362	98.1	6	1.6
Ukraine	2	0.8	263	99.3	0	0.0	0	0.0	78	98.7	1	1.3
United Kingdom	0	0.0	4138	100	0	0.0	0	0.0	5774	99.8	12	0.2
Total	129	0.6	22796	99.4	18	0.1	215	0.9	24762	98.9	69	0.3

Note: Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the table for adults, but the people are included in the total number.

7. Complications

Table 7.5   Prevalence of haemoptysis major (≥250 ml over the course of a day) in all people with CF seen in 2022 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	3	4.5	64	95.5	0	0.0						
Armenia	0	0.0	21	95.5	1	4.6						
Austria	1	0.3	370	99.7	0	0.0	6	1.5	392	98.3	1	0.3
Belarus	0	0.0	145	99.3	1	0.7	0	0.0	5	100	0	0.0
Bulgaria	4	3.2	120	95.2	2	1.6	5	5.9	78	91.8	2	2.4
Croatia	1	1.2	81	98.8	0	0.0	0	0.0	53	96.4	2	3.6
Cyprus	0	0.0	7	100	0	0.0	0	0.0	16	100	0	0.0
Czech Republic	11	3.4	313	96.6	0	0.0	22	7.3	279	92.4	1	0.3
Denmark	0	0.0	211	100	0	0.0	0	0.0	302	99.0	3	1.0
Finland	0	0.0	26	100	0	0.0	0	0.0	29	93.6	2	6.5
France	0	0.0	2689	99.9	2	0.1	0	0.0	3655	99.2	29	0.8
Georgia	3	7.3	38	92.7	0	0.0						
Germany	9	0.3	2747	99.7	0	0.0	58	1.5	3753	98.4	5	0.1
Greece	0	0.0	235	99.2	2	0.8	0	0.0	340	96.1	14	4.0
Hungary	0	0.0	235	99.2	2	0.8	4	2.1	180	92.8	10	5.2
Iceland	0	0.0	9	100	0	0.0	0	0.0	6	100	0	0.0
Ireland	10	1.9	510	97.1	5	1.0	7	1.0	710	98.8	2	0.3
Israel	2	1.3	156	98.1	1	0.6	5	1.5	330	97.1	5	1.5
Italy	13	0.6	2178	98.8	13	0.6	37	1.0	3406	95.3	130	3.6
Latvia	0	0.0	32	100	0	0.0	0	0.0	14	100	0	0.0
Lithuania	0	0.0	20	100	0	0.0	1	4.4	21	91.3	1	4.4
Luxembourg	0	0.0	21	100	0	0.0	0	0.0	5	100	0	0.0
Rep of Moldova	0	0.0	36	100	0	0.0	0	0.0	12	100	0	0.0
The Netherlands	8	1.5	531	98.5	0	0.0	21	2.2	928	95.7	21	2.2
North Macedonia	0	0.0	74	100	0	0.0	0	0.0	49	100	0	0.0
Norway	2	1.6	126	98.4	0	0.0	1	0.5	193	99.0	1	0.5
Poland	12	1.3	938	98.4	3	0.3	5	0.9	508	95.5	19	3.6
Portugal	1	0.6	169	98.8	1	0.6	5	2.7	179	94.7	5	2.7
Romania	1	0.4	261	99.6	0	0.0	1	4.2	23	95.8	0	0.0
Russian Fed.	13	0.7	1855	99.0	6	0.3	23	4.5	474	92.9	13	2.6
Serbia	0	0.0	136	100	0	0.0	1	1.5	65	98.5	0	0.0
Slovak Republic	0	0.0	126	100	0	0.0	8	5.4	136	92.5	3	2.0
Slovenia	0	0.0	55	100	0	0.0	3	6.5	43	93.5	0	0.0
Spain	23	2.2	1033	97.5	3	0.3	37	3.1	1161	96.2	9	0.8
Sweden	15	5.7	250	94.3	0	0.0	4	1.0	381	97.9	4	1.0
Switzerland	8	2.0	399	98.0	0	0.0	12	2.2	526	96.7	6	1.1
Turkey	1	0.1	2132	99.6	8	0.4	1	0.3	352	95.4	16	4.3
Ukraine	8	3.0	252	95.1	5	1.9	1	1.3	76	96.2	2	2.5
United Kingdom	0	0.0	4138	100	0	0.0	0	0.0	5776	99.8	10	0.2
Total	149	0.7	22739	99.1	55	0.2	268	1.1	24462	97.7	316	1.3

Note: Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the table for adults, but the people are included in the total number.

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.

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 2022 aged 18 years or older who have never had a transplant (table A7.2, Appendix 1).
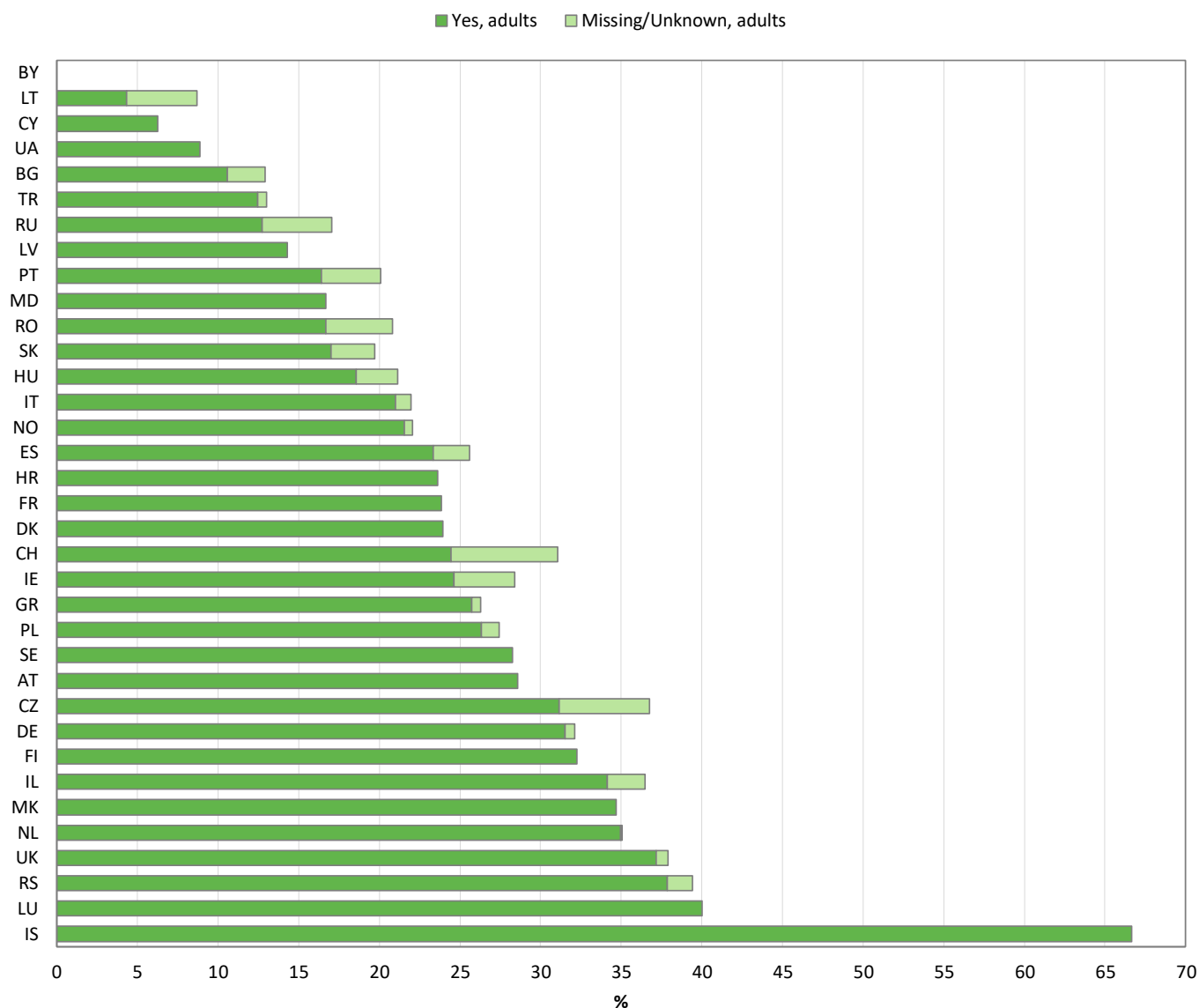
Note: We excluded from the graph the countries for which the information on CFRD is missing for more than 10% of the adults.

Albania, Armenia and Georgia have <5 adults seen in 2022 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 in Europe.

Prevalence and severity of liver disease in children (<18 years) and adults (≥ 18 years) with CF seen in 2022 who have never had a transplant, by country (table A7.3 and table A7.4, 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.

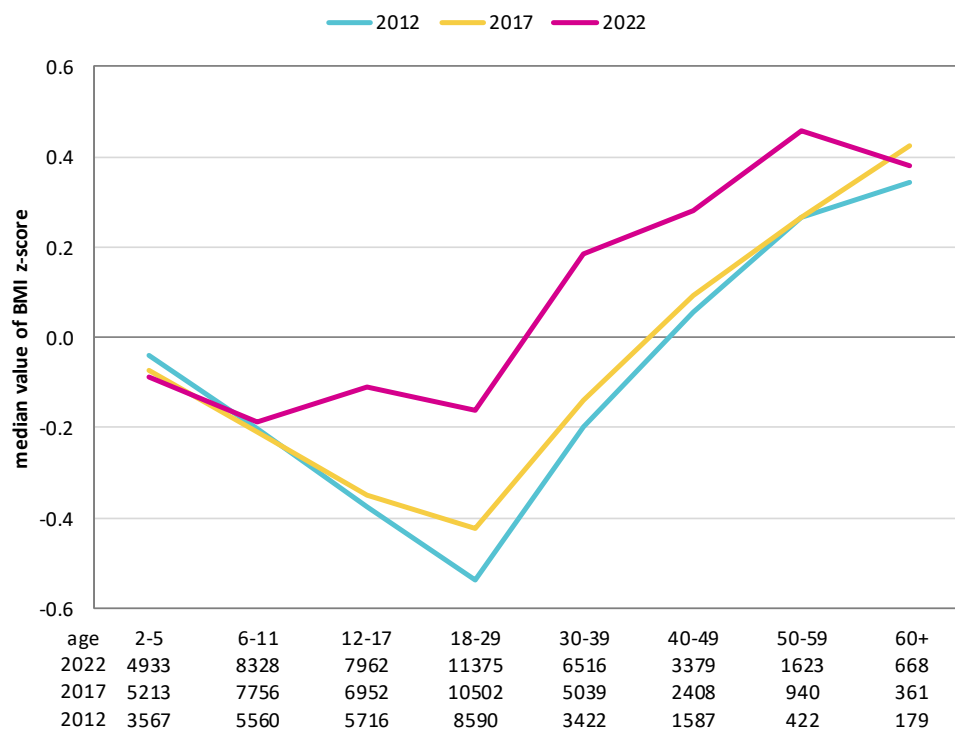Albania, Armenia and Georgia have <5 adults seen in 2021 and are excluded from the graph for adults.

Note: 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 veins, 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 table 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   Malignancy newly diagnosed this year in all people with CF seen in 2022 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	3	4.5	64	95.5	0	0.0						
Armenia	0	0.0	22	100	0	0.0						
Austria	2	0.5	369	99.5	0	0.0	18	4.5	380	95.2	1	0.3
Belarus	0	0.0	145	99.3	1	0.7	0	0.0	5	100	0	0.0
Bulgaria	2	1.6	124	98.4	0	0.0	2	2.4	82	96.5	1	1.2
Croatia	2	2.4	80	97.6	0	0.0	0	0.0	54	98.2	1	1.8
Cyprus	0	0.0	7	100	0	0.0	0	0.0	16	100	0	0.0
Czech Republic	8	2.5	316	97.5	0	0.0	46	15.2	254	84.1	2	0.7
Denmark	0	0.0	211	100	0	0.0	0	0.0	304	99.7	1	0.3
Finland	0	0.0	26	100	0	0.0	0	0.0	31	100	0	0.0
France	0	0.0	2690	100	1	0.0	5	0.1	3655	99.2	24	0.7
Georgia	3	7.3	38	92.7	0	0.0						
Germany	6	0.2	2745	99.6	5	0.2	17	0.5	3755	98.4	44	1.2
Greece	0	0.0	237	100	0	0.0	1	0.3	350	98.9	3	0.9
Hungary	1	0.4	236	99.6	0	0.0	4	2.1	190	97.9	0	0.0
Iceland	0	0.0	9	100	0	0.0	0	0.0	6	100	0	0.0
Ireland	12	2.3	513	97.7	0	0.0	6	0.8	713	99.2	0	0.0
Israel	2	1.3	157	98.7	0	0.0	7	2.1	332	97.7	1	0.3
Italy	14	0.6	2182	99.0	8	0.4	37	1.0	3510	98.2	26	0.7
Latvia	0	0.0	32	100	0	0.0	0	0.0	14	100	0	0.0
Lithuania	0	0.0	20	100	0	0.0	0	0.0	23	100	0	0.0
Luxembourg	0	0.0	20	95.2	1	4.8	0	0.0	5	100	0	0.0
Rep of Moldova	0	0.0	36	100	0	0.0	0	0.0	12	100	0	0.0
The Netherlands	6	1.1	533	98.9	0	0.0	0	0.0	965	99.5	5	0.5
North Macedonia	0	0.0	74	100	0	0.0	0	0.0	49	100	0	0.0
Norway	1	0.8	127	99.2	0	0.0	1	0.5	189	96.9	5	2.6
Poland	10	1.1	943	99.0	0	0.0	12	2.3	519	97.6	1	0.2
Portugal	1	0.6	170	99.4	0	0.0	6	3.2	183	96.8	0	0.0
Romania	0	0.0	262	100	0	0.0	0	0.0	24	100	0	0.0
Russian Fed.	9	0.5	1865	99.5	0	0.0	26	5.1	483	94.7	1	0.2
Serbia	0	0.0	136	100	0	0.0	1	1.5	65	98.5	0	0.0
Slovak Republic	0	0.0	126	100	0	0.0	4	2.7	142	96.6	1	0.7
Slovenia	1	1.8	54	98.2	0	0.0	3	6.5	43	93.5	0	0.0
Spain	16	1.5	1043	98.5	0	0.0	22	1.8	1179	97.7	6	0.5
Sweden	15	5.7	250	94.3	0	0.0	4	1.0	385	99.0	0	0.0
Switzerland	5	1.2	401	98.5	1	0.3	8	1.5	535	98.4	1	0.2
Turkey	1	0.1	2139	99.9	1	0.1	1	0.3	366	99.2	2	0.5
Ukraine	1	0.4	263	99.3	1	0.4	0	0.0	79	100	0	0.0
United Kingdom	6	0.1	4131	99.8	1	0.0	13	0.2	5755	99.5	18	0.3
Total	127	0.6	22796	99.4	20	0.1	244	1.0	24658	98.5	144	0.6

Note: Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the table for adults, but the people are included in the total number.

7. Complications

Table 7.7 Type of malignancy newly diagnosed this year in children with CF seen in 2022 who have never had a transplant, by country and overall.

Country	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	%
Albania	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Armenia	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Austria	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Belarus	1	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Bulgaria	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Croatia	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Cyprus	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Czech Republic	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Denmark	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Finland	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
France	0	0.0	0	0.0	1	100	0	0.0	0	0.0	0	0.0	0	0.0
Georgia	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Germany	0	0.0	0	0.0	1	20.0	0	0.0	0	0.0	0	0.0	4	80.0
Greece	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Hungary	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Iceland	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Ireland	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Israel	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Italy	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	8	100
Latvia	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Lithuania	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Luxembourg	0	0.0	0	0.0	1	100	0	0.0	0	0.0	0	0.0	0	0.0
Rep of Moldova	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
The Netherlands	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
N. Macedonia	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Norway	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Poland	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Portugal	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Romania	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Russian Fed.	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Serbia	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Slovak Republic	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Slovenia	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Spain	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Sweden	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Switzerland	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	1	100
Turkey	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	1	100
Ukraine	0	0.0	0	0.0	0	0.0	1	100	0	0.0	0	0.0	0	0.0
United Kingdom	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	1	100
Total	1	5.0	0	0.0	3	15.0	1	5.0	0	0.0	0	0.0	15	75.0

7. Complications

Table 7.8   Type of malignancy newly diagnosed this year in adults with CF seen in 2022 who have never had a transplant, by country and overall.

Country	Type of malignancy													
	Colorectal cancer		Small cancer	bowel	Lymphoid leukaemia		Testicular cancer		Breast cancer		Thyroid gland cancer		Other or type unknown	
	N	%	N	%	N	%	N	%	N	%	N	%	N	%
Austria	0	0.0	0	0.0	1	100	0	0.0	0	0.0	0	0.0	0	0.0
Belarus	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Bulgaria	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	1	100
Croatia	1	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Cyprus	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Czech Republic	0	0.0	1	50.0	0	0.0	0	0.0	0	0.0	0	0.0	1	50.0
Denmark	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	1	100
Finland	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
France	3	12.5	0	0.0	3	12.5	1	4.2	4	16.7	0	0.0	13	54.2
Germany	8	18.2	0	0.0	0	0.0	5	11.4	7	15.9	2	4.6	22	50.0
Greece	0	0.0	0	0.0	0	0.0	0	0.0	2	66.7	1	33.3	0	0.0
Hungary	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Iceland	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Ireland	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Israel	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	1	100
Italy	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	26	100
Latvia	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Lithuania	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Luxembourg	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Rep of Moldova	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Netherlands	2	40.0	0	0.0	0	0.0	1	20.0	0	0.0	0	0.0	2	40.0
N. Macedonia	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Norway	0	0.0	0	0.0	0	0.0	0	0.0	2	40.0	0	0.0	3	60.0
Poland	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	1	100
Portugal	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Romania	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Russian Fed.	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	1	100
Serbia	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Slovak Republic	0	0.0	0	0.0	0	0.0	0	0.0	1	100	0	0.0	0	0.0
Slovenia	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Spain	2	33.3	0	0.0	0	0.0	1	16.7	1	16.7	0	0.0	2	33.3
Sweden	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Switzerland	0	0.0	0	0.0	1	100	0	0.0	0	0.0	0	0.0	0	0.0
Turkey	0	0.0	0	0.0	0	0.0	0	0.0	1	50.0	0	0.0	1	50.0
Ukraine	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
United Kingdom	0	0.0	4	22.2	1	5.6	1	5.6	1	5.6	0	0.0	11	61.1
Total	16	11.1	5	3.5	6	4.2	9	6.3	19	13.2	3	2.1	86	59.7

Note: Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the table for adults, but the people are included in the total number.

7. Complications

Table 7.9 Prevalence of distal intestinal obstruction syndrome (DIOS) in all people with CF seen in 2022 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	3	4.5	61	91.0	3	4.5						
Armenia	0	0.0	19	86.4	3	13.6						
Austria	2	0.5	357	96.2	12	3.2	0	0.0	390	97.7	9	2.3
Belarus	0	0.0	145	99.3	1	0.7	0	0.0	5	100	0	0.0
Bulgaria	2	1.6	123	97.6	1	0.8	2	2.4	82	96.5	1	1.2
Croatia	1	1.2	81	98.8	0	0.0	0	0.0	50	90.9	5	9.1
Cyprus	0	0.0	7	100	0	0.0	0	0.0	16	100	0	0.0
Czech Republic	12	3.7	308	95.1	4	1.2	24	8.0	278	92.1	0	0.0
Denmark	0	0.0	206	97.6	5	2.4	0	0.0	299	98.0	6	2.0
Finland	0	0.0	24	92.3	2	7.7	0	0.0	31	100	0	0.0
France	0	0.0	2624	97.5	67	2.5	0	0.0	3592	97.5	92	2.5
Georgia	3	7.3	37	90.2	1	2.4						
Germany	8	0.3	2690	97.6	58	2.1	25	0.7	3663	96.0	128	3.4
Greece	0	0.0	234	98.7	3	1.3	3	0.9	345	97.5	6	1.7
Hungary	0	0.0	236	99.6	1	0.4	6	3.1	185	95.4	3	1.6
Iceland	0	0.0	9	100	0	0.0	0	0.0	6	100	0	0.0
Ireland	10	1.9	509	97.0	6	1.1	8	1.1	709	98.6	2	0.3
Israel	2	1.3	156	98.1	1	0.6	6	1.8	327	96.2	7	2.1
Italy	13	0.6	2144	97.3	47	2.1	36	1.0	3494	97.8	43	1.2
Latvia	0	0.0	32	100	0	0.0	1	7.1	13	92.9	0	0.0
Lithuania	0	0.0	20	100	0	0.0	0	0.0	23	100	0	0.0
Luxembourg	0	0.0	21	100	0	0.0	0	0.0	5	100	0	0.0
Rep of Moldova	0	0.0	35	97.2	1	2.8	0	0.0	12	100	0	0.0
The Netherlands	8	1.5	525	97.4	6	1.1	18	1.9	940	96.9	12	1.2
North Macedonia	0	0.0	72	97.3	2	2.7	0	0.0	49	100	0	0.0
Norway	2	1.6	124	96.9	2	1.6	4	2.1	189	96.9	2	1.0
Poland	7	0.7	938	98.4	8	0.8	7	1.3	520	97.7	5	0.9
Portugal	0	0.0	167	97.7	4	2.3	5	2.7	183	96.8	1	0.5
Romania	2	0.8	252	96.2	8	3.1	0	0.0	23	95.8	1	4.2
Russian Fed.	6	0.3	1839	98.1	29	1.6	22	4.3	482	94.5	6	1.2
Serbia	0	0.0	136	100	0	0.0	1	1.5	65	98.5	0	0.0
Slovak Republic	0	0.0	125	99.2	1	0.8	4	2.7	142	96.6	1	0.7
Slovenia	0	0.0	51	92.7	4	7.3	3	6.5	42	91.3	1	2.2
Spain	7	0.7	1041	98.3	11	1.0	20	1.7	1181	97.9	6	0.5
Sweden	15	5.7	237	89.4	13	4.9	4	1.0	377	96.9	8	2.1
Switzerland	5	1.2	397	97.5	5	1.2	4	0.7	532	97.8	8	1.5
Turkey	1	0.1	2132	99.6	8	0.4	1	0.3	367	99.5	1	0.3
Ukraine	2	0.8	261	98.5	2	0.8	1	1.3	77	97.5	1	1.3
United Kingdom	0	0.0	4046	97.8	92	2.2	0	0.0	5450	94.2	336	5.8
Total	111	0.5	22421	97.7	411	1.8	205	0.8	24150	96.4	691	2.8

Note: Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the table for adults, but the people are included in the total number.

Note: Denmark only reported DIOS requiring hospitalisation.

7. Complications

Table 7.10   Prevalence of salt loss syndrome in all people with CF seen in 2022 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	3	4.5	64	95.5	0	0.0						
Armenia	0	0.0	17	77.3	5	22.7						
Austria	1	0.3	368	99.2	2	0.5	1	0.3	397	99.5	1	0.3
Belarus	0	0.0	142	97.3	4	2.7	0	0.0	5	100	0	0.0
Bulgaria	2	1.6	122	96.8	2	1.6	2	2.4	82	96.5	1	1.2
Croatia	1	1.2	77	93.9	4	4.9	0	0.0	55	100	0	0.0
Cyprus	0	0.0	7	100	0	0.0	0	0.0	16	100	0	0.0
Czech Republic	8	2.5	313	96.6	3	0.9	18	6.0	284	94.0	0	0.0
Denmark	211	100	-	-	-	-	305	100	-	-	-	-
Finland	0	0.0	25	96.2	1	3.9	0	0.0	31	100	0	0.0
France	2691	100	-	-	-	-	3684	100	-	-	-	-
Georgia	3	7.3	38	92.7	0	0.0						
Germany	7	0.3	2723	98.8	26	0.9	17	0.5	3795	99.5	4	0.1
Greece	0	0.0	235	99.2	2	0.8	0	0.0	354	100	0	0.0
Hungary	237	100	-	-	-	-	194	100	-	-	-	-
Iceland	0	0.0	9	100	0	0.0	0	0.0	6	100	0	0.0
Ireland	173	33.0	352	67.1	0	0.0	708	98.5	11	1.5	0	0.0
Israel	2	1.3	157	98.7	0	0.0	7	2.1	332	97.7	1	0.3
Italy	11	0.5	2150	97.6	43	2.0	40	1.1	3458	96.8	75	2.1
Latvia	1	3.1	31	96.9	0	0.0	0	0.0	14	100	0	0.0
Lithuania	2	10.0	18	90.0	0	0.0	1	4.4	22	95.7	0	0.0
Luxembourg	0	0.0	21	100	0	0.0	0	0.0	5	100	0	0.0
Rep of Moldova	0	0.0	33	91.7	3	8.3	0	0.0	12	100	0	0.0
The Netherlands	539	100	-	-	-	-	970	100	-	-	-	-
North Macedonia	0	0.0	73	98.7	1	1.4	0	0.0	49	100	0	0.0
Norway	1	0.8	127	99.2	0	0.0	2	1.0	193	99.0	0	0.0
Poland	7	0.7	942	98.9	4	0.4	9	1.7	519	97.6	4	0.8
Portugal	1	0.6	170	99.4	0	0.0	5	2.7	184	97.4	0	0.0
Romania	0	0.0	260	99.2	2	0.8	0	0.0	24	100	0	0.0
Russian Fed.	22	1.2	1792	95.6	60	3.2	30	5.9	478	93.7	2	0.4
Serbia	0	0.0	135	99.3	1	0.7	1	1.5	65	98.5	0	0.0
Slovak Republic	2	1.6	123	97.6	1	0.8	5	3.4	142	96.6	0	0.0
Slovenia	0	0.0	53	96.4	2	3.6	3	6.5	43	93.5	0	0.0
Spain	7	0.7	1044	98.6	8	0.8	19	1.6	1187	98.3	1	0.1
Sweden	15	5.7	249	94.0	1	0.4	4	1.0	385	99.0	0	0.0
Switzerland	4	1.0	403	99.0	0	0.0	5	0.9	539	99.1	0	0.0
Turkey	1	0.1	2074	96.9	66	3.1	1	0.3	364	98.6	4	1.1
Ukraine	6	2.3	255	96.2	4	1.5	0	0.0	73	92.4	6	7.6
United Kingdom	4138	100	-	-	-	-	5786	100	-	-	-	-
Total	8096	35.3	14602	63.6	245	1.1	11817	47.2	13130	52.4	99	0.4

Note: Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the table for adults, but the people are included in the total number.

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.

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 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 is 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 2022 who have never had a transplant, by country (table A8.1, Appendix 1).
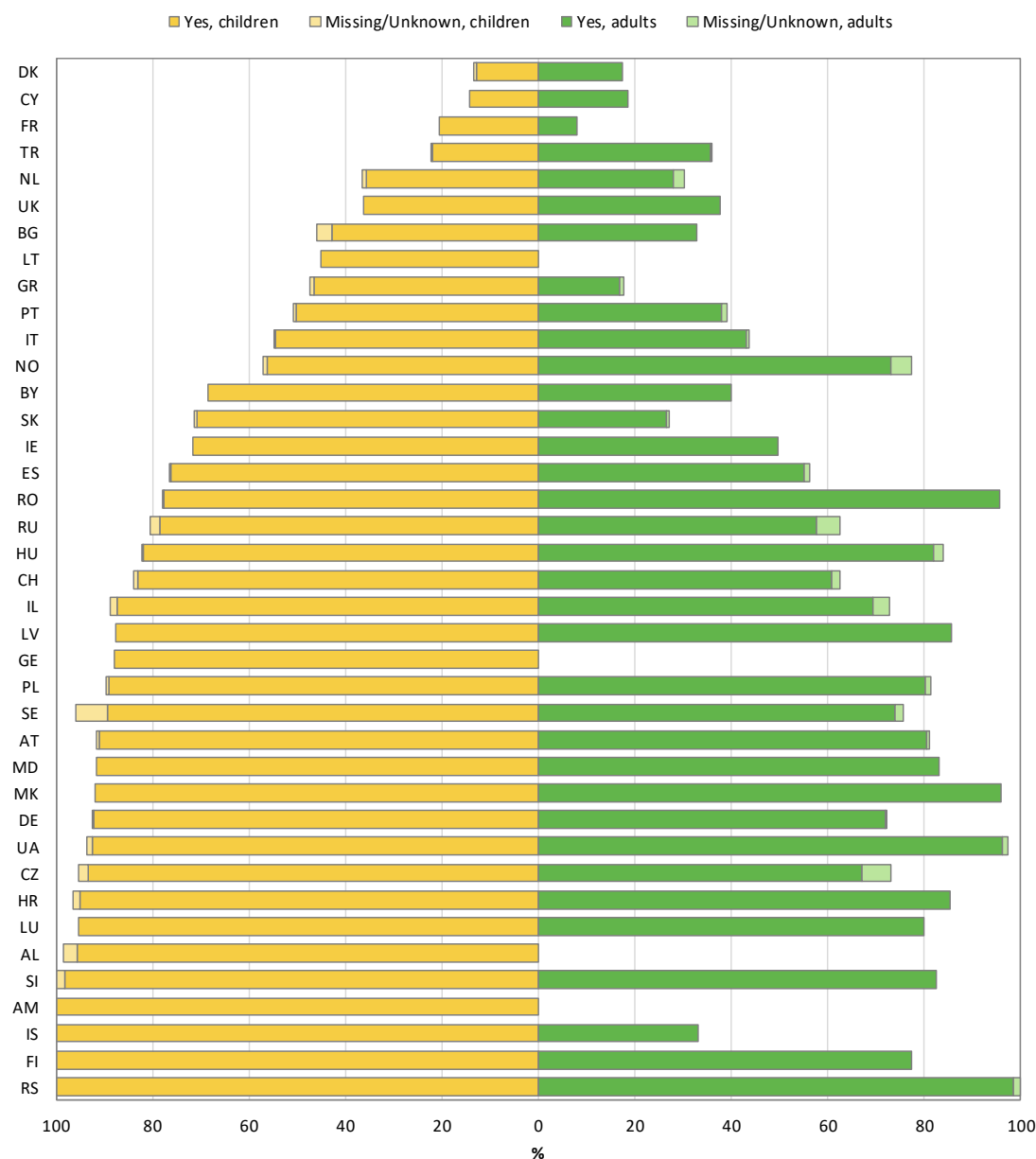
Note: We excluded from the graph the countries for which the information on inhaled hypertonic saline is missing for more than 10% of the children/adults with CF. Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the graph for adults.

Note: Inhaled hypertonic saline is reimbursed in most countries except in Albania, Armenia, Bulgaria, Georgia, Lithuania, the Republic of Moldova, Poland, Romania and Ukraine. In Turkey 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 2022 who have never had a transplant, by country (table A8.2, Appendix 1).

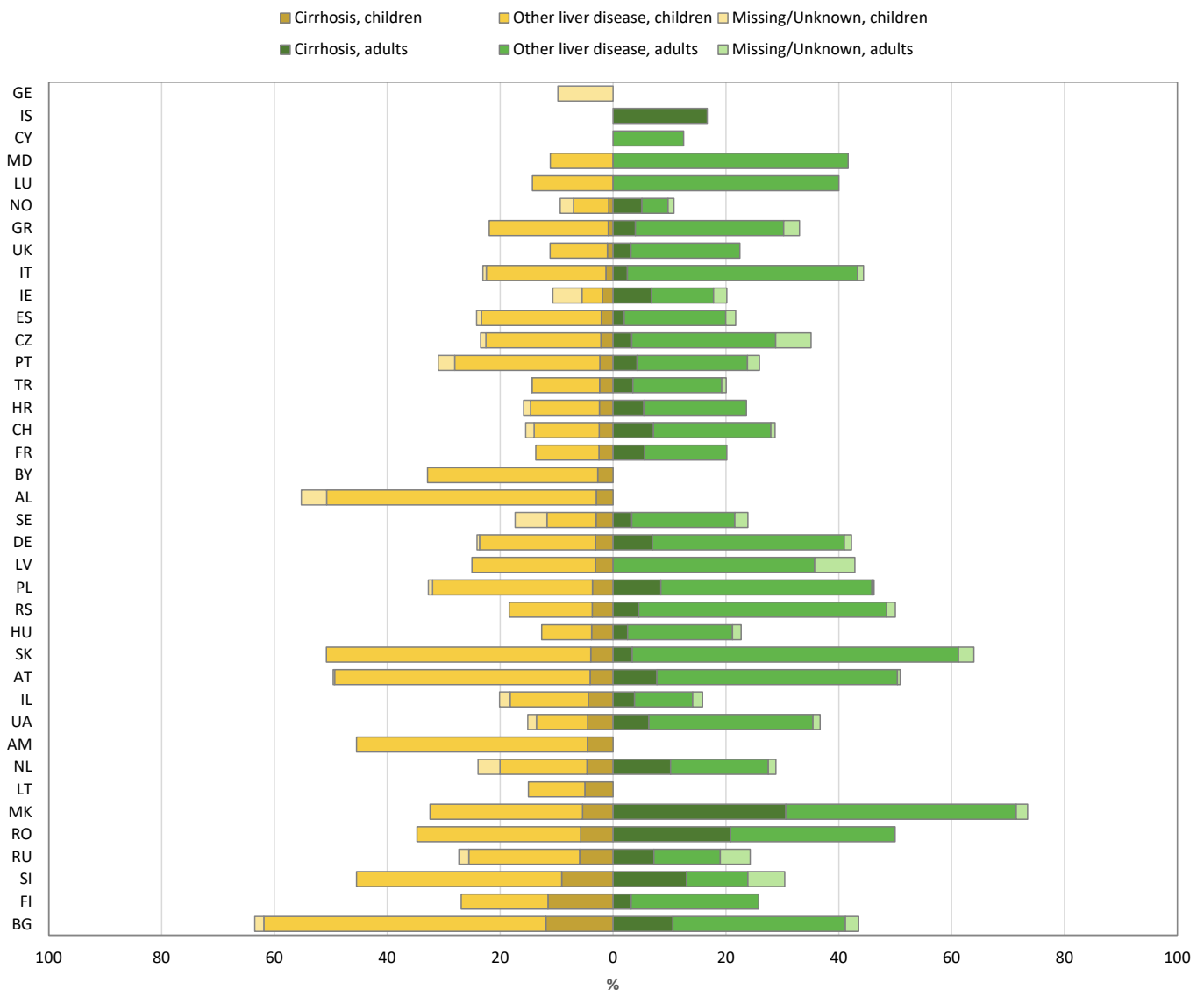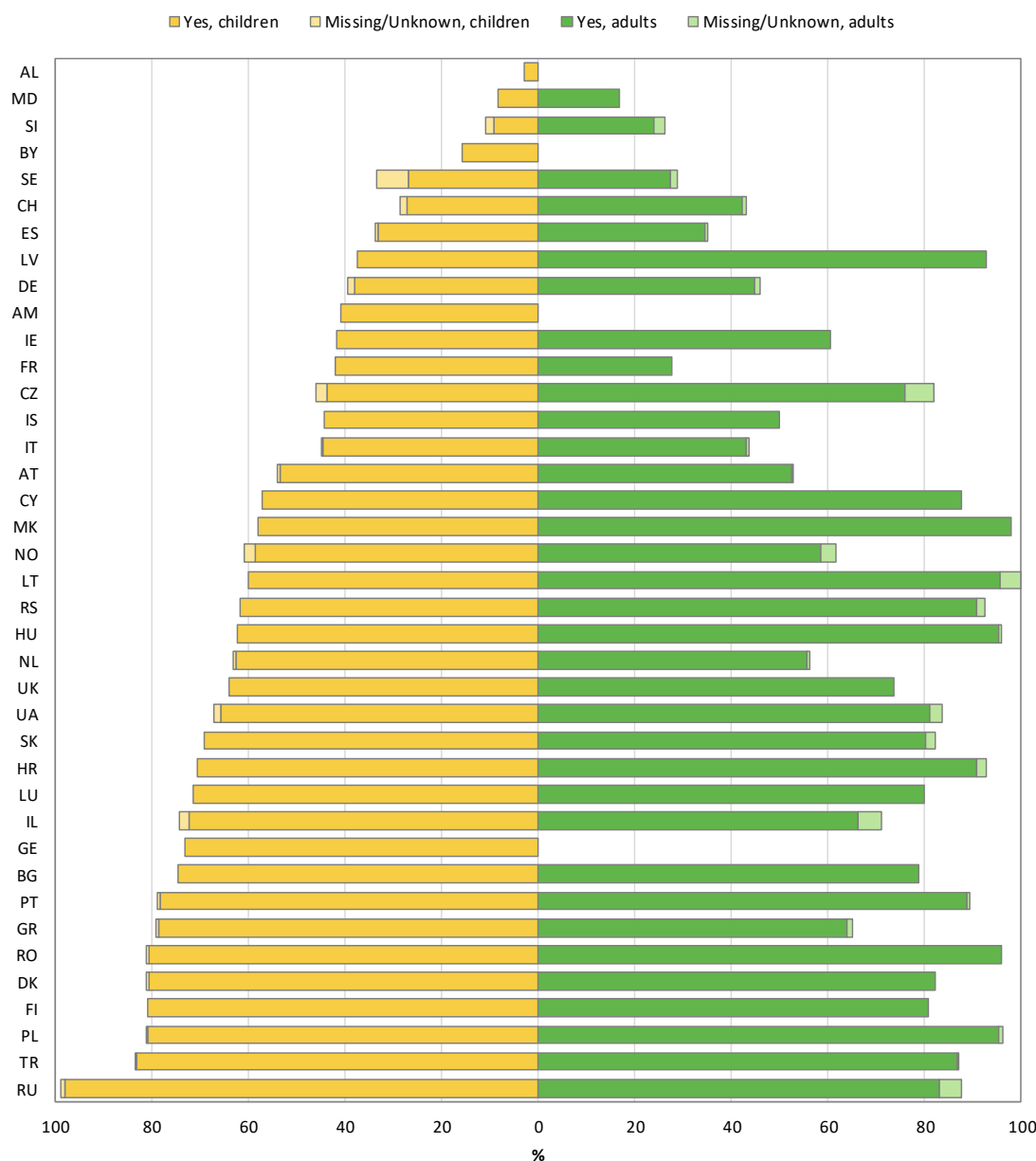Note: We excluded from the graph the countries for which the information on rhDNase is missing for more than 10% of the children/adults with CF. Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the graph for adults.

Note: Inhaled rhDNase is reimbursed in most countries except in Albania, Armenia, Belarus and the Republic of Moldova. It is reimbursed in Georgia for people with CF ≥ 2 years; in Bulgaria, Germany, Luxembourg, Macedonia, Norway, Romania, Spain, and the United Kingdom for individuals ≥ 5 years; 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 2022 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	67	100	0	0.0						
Armenia	0	0.0	18	81.8	4	18.2						
Austria	2	0.5	369	99.5	0	0.0	1	0.3	391	98.0	7	1.8
Belarus	0	0.0	135	92.5	11	7.5	0	0.0	4	80.0	1	20.0
Bulgaria	2	1.6	124	98.4	0	0.0	0	0.0	85	100	0	0.0
Croatia	0	0.0	82	100	0	0.0	0	0.0	55	100	0	0.0
Cyprus	0	0.0	7	100	0	0.0	0	0.0	16	100	0	0.0
Czech Republic	4	1.2	320	98.8	0	0.0	18	6.0	280	92.7	4	1.3
Denmark	1	0.5	206	97.6	4	1.9	0	0.0	296	97.1	9	3.0
Finland	0	0.0	26	100	0	0.0	0	0.0	31	100	0	0.0
France	2691	100	-	-	-	-	3684	100	-	-	-	-
Georgia	2	4.9	39	95.1	0	0.0						
Germany	55	2.0	2698	97.9	3	0.1	83	2.2	3593	94.2	140	3.7
Greece	3	1.3	233	98.3	1	0.4	3	0.9	344	97.2	7	2.0
Hungary	1	0.4	235	99.2	1	0.4	5	2.6	176	90.7	13	6.7
Iceland	0	0.0	9	100	0	0.0	0	0.0	6	100	0	0.0
Ireland	525	100	-	-	-	-	719	100	-	-	-	-
Israel	1	0.6	157	98.7	1	0.6	10	2.9	323	95.0	7	2.1
Italy	2	0.1	2189	99.3	13	0.6	14	0.4	3484	97.5	75	2.1
Latvia	0	0.0	32	100	0	0.0	0	0.0	14	100	0	0.0
Lithuania	0	0.0	20	100	0	0.0	0	0.0	23	100	0	0.0
Luxembourg	0	0.0	21	100	0	0.0	0	0.0	5	100	0	0.0
Rep of Moldova	0	0.0	36	100	0	0.0	0	0.0	12	100	0	0.0
The Netherlands	539	100	-	-	-	-	970	100	-	-	-	-
North Macedonia	0	0.0	74	100	0	0.0	0	0.0	49	100	0	0.0
Norway	1	0.8	127	99.2	0	0.0	1	0.5	192	98.5	2	1.0
Poland	3	0.3	949	99.6	1	0.1	7	1.3	524	98.5	1	0.2
Portugal	1	0.6	170	99.4	0	0.0	2	1.1	187	98.9	0	0.0
Romania	2	0.8	260	99.2	0	0.0	1	4.2	23	95.8	0	0.0
Russian Fed.	21	1.1	1724	92.0	129	6.9	23	4.5	413	81.0	74	14.5
Serbia	0	0.0	136	100	0	0.0	1	1.5	65	98.5	0	0.0
Slovak Republic	1	0.8	125	99.2	0	0.0	1	0.7	145	98.6	1	0.7
Slovenia	1	1.8	54	98.2	0	0.0	1	2.2	45	97.8	0	0.0
Spain	4	0.4	1053	99.4	2	0.2	9	0.8	1184	98.1	14	1.2
Sweden	17	6.4	247	93.2	1	0.4	7	1.8	382	98.2	0	0.0
Switzerland	4	1.0	403	99.0	0	0.0	8	1.5	536	98.5	0	0.0
Turkey	2	0.1	2103	98.2	36	1.7	1	0.3	349	94.6	19	5.2
Ukraine	3	1.1	261	98.5	1	0.4	5	6.3	74	93.7	0	0.0
United Kingdom	0	0.0	4133	99.9	5	0.1	0	0.0	5523	95.5	263	4.6

Note: For inhaled mannitol the total percentage of missing information is higher than 10%, therefore the totals are excluded from the table.

Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the table for adults, but the people are included in the total number.

Note: Inhaled mannitol is reimbursed for all patients with CF in Austria, Czech Republic, Denmark, Norway, the Russian Federation (depending on the region of residence), and Slovenia. It is reimbursed for people with CF ≥ 18 years in Germany, Greece, Italy, and the United Kingdom and it is reimbursed for people with CF ≥ 6 years in Turkey. 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 2022 who have never had a transplant, by country (table A8.3, Appendix 1).
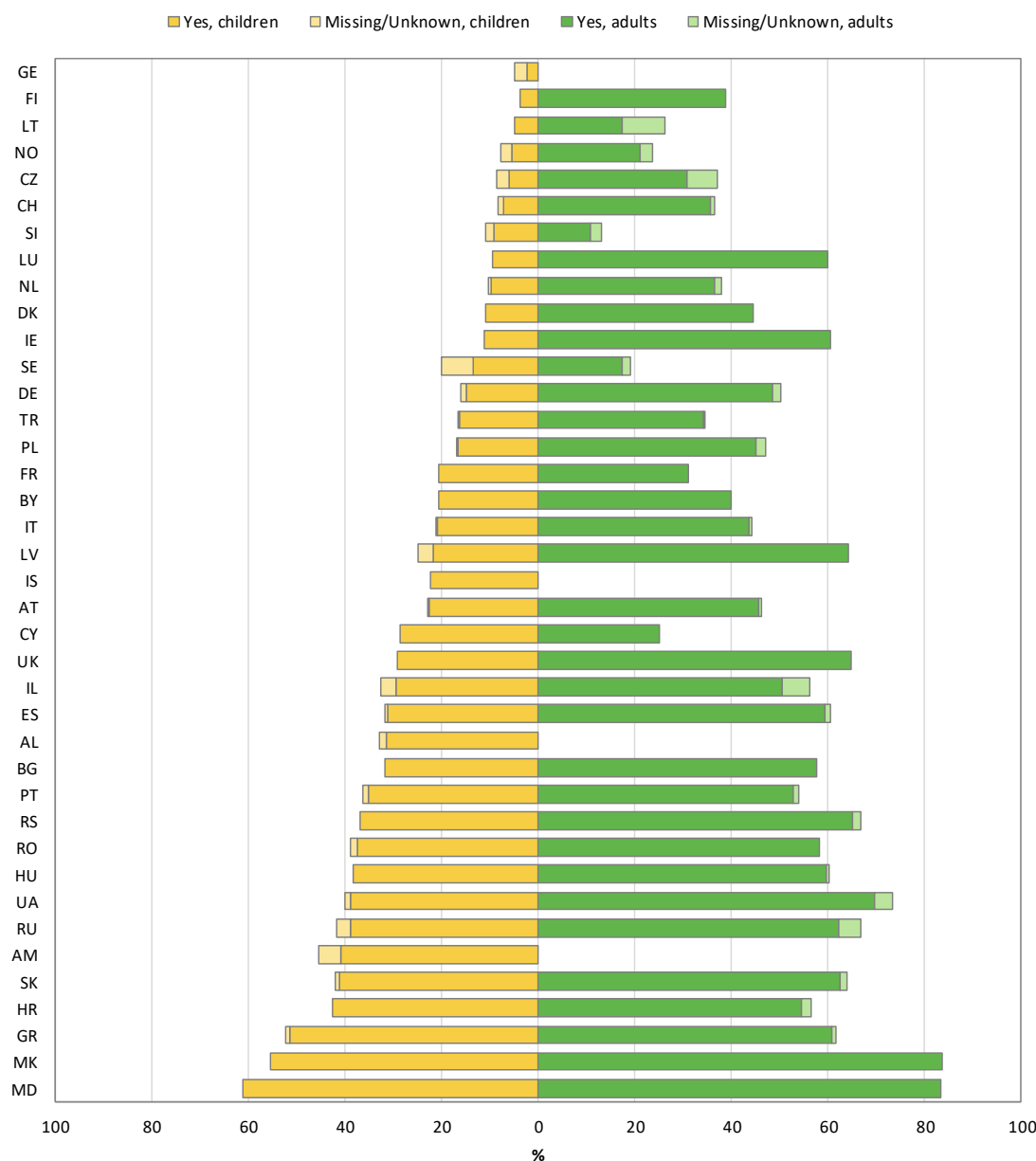
Note: We excluded from the graph the countries for which the information on inhaled antibiotics is missing for more than 10% of the children/adults with CF. Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the graph for adults.

Note: Inhaled antibiotics are reimbursed in all countries. In Armenia only Gentamycin is reimbursed, in Romania only Tobramycin solution and Colistin dry powder are reimbursed for children ≥ 6 years old, in Ukraine only Colistin dry powder is reimbursed.

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 2022 who have never had a transplant, by country (table A8.4 and A8.5, Appendix 1).
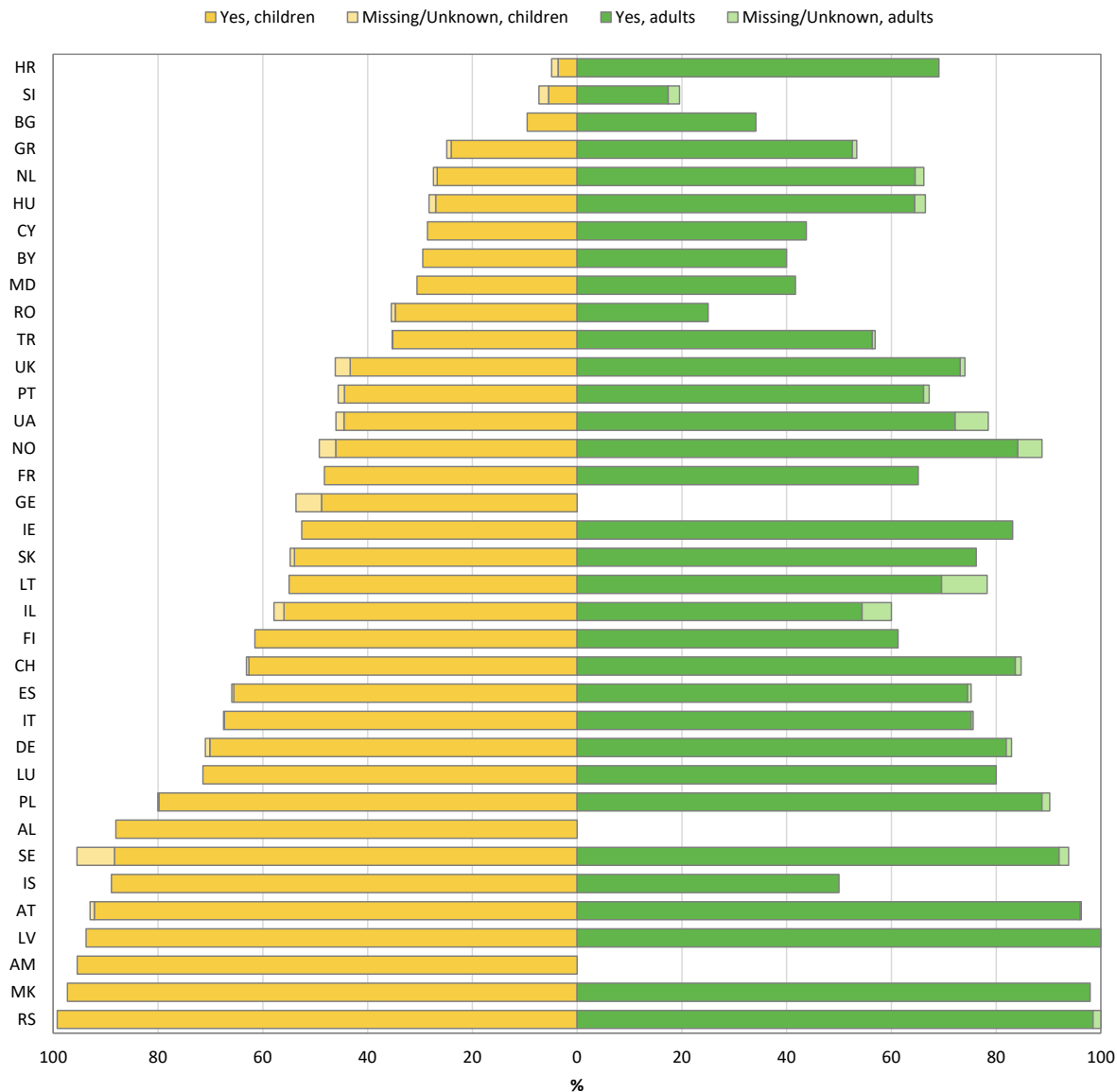
Note: We excluded from the graph the countries for which the information on the use of bronchodilators is missing for more than 10% of the children/adults with CF. Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the graph for adults.

Note: Inhaled bronchodilators are reimbursed in most countries except in Bulgaria, Georgia, Poland, Serbia and Ukraine.

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 widely used as an antibiotic and anti-inflammatory mediator throughout Europe, mostly by adults with CF.

Use of macrolides in children and adults seen in 2022 who have never had a transplant, by country (table A8.6, Appendix 1).
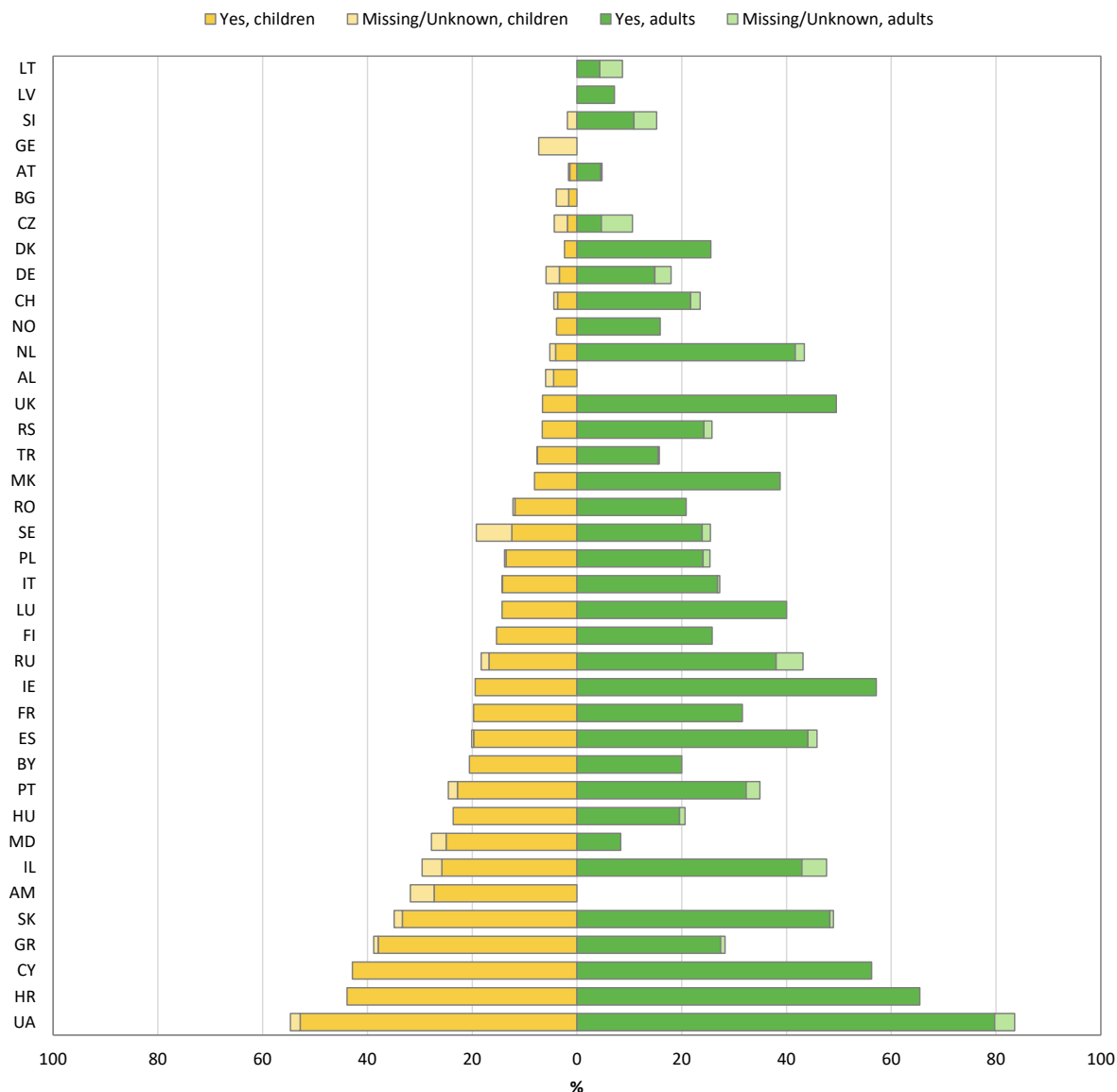
Note: We excluded from the graph the countries for which the information on the use of macrolides is missing for more than 10% of the children/adults with CF. Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the graph for adults.

Note: Oral macrolides are reimbursed in most countries except in Bulgaria, Georgia, Serbia and Ukraine. In the Republic of Moldova they are reimbursed for children. Inhaled macrolides are reimbursed in Slovenia, Sweden (≥ 18 years) and the UK.

This graph shows the use of macrolides (e.g. azithromycin or another macrolide) for at least 3 consecutive months during 2022. 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, an indicator of severe lung disease, is prescribed in up to 22% of people with CF, mostly in the adult population.

Use of oxygen in children and adults seen in 2022 who have never had a transplant, by country (table A8.7, Appendix 1).
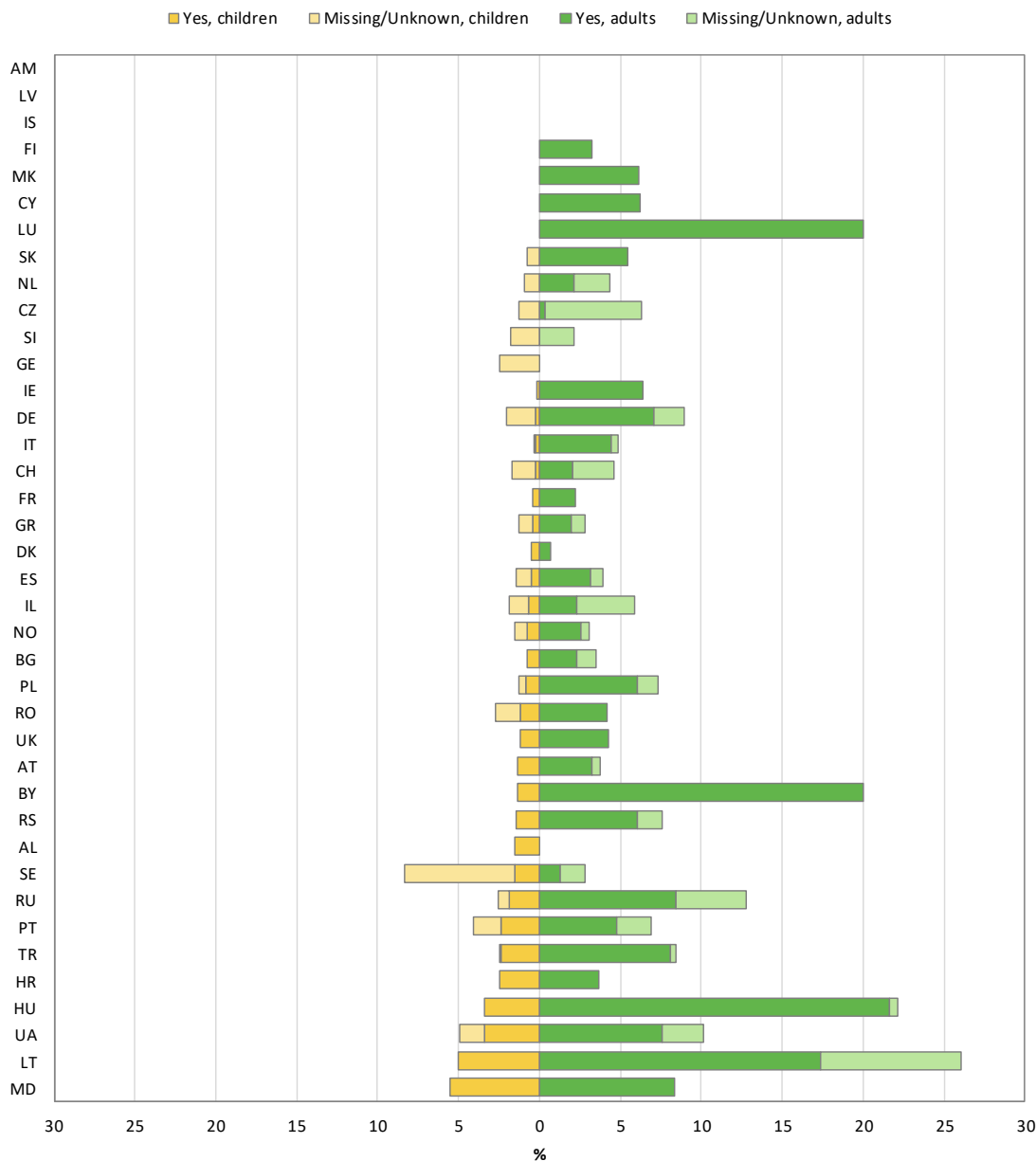
Note: We excluded from the graph the countries for which the information on the use of oxygen is missing for more than 10% of the children/adults with CF.

Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the graph for adults.

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 therapy at home is reimbursed.

This graph shows the use of oxygen for at least 3 months during 2022. 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 2022 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	66	98.5	0	0.0	1	1.5								
Armenia	1	4.6	21	95.5	0	0.0	0	0.0								
Austria	0	0.0	371	100	0	0.0	0	0.0	1	0.3	397	99.5	1	0.3	0	0.0
Belarus	0	0.0	143	98.0	0	0.0	3	2.1	0	0.0	5	100	0	0.0	0	0.0
Bulgaria	0	0.0	126	100	0	0.0	0	0.0	0	0.0	84	98.8	1	1.2	0	0.0
Croatia	0	0.0	81	98.8	1	1.2	0	0.0	0	0.0	55	100	0	0.0	0	0.0
Cyprus	0	0.0	7	100	0	0.0	0	0.0	0	0.0	16	100	0	0.0	0	0.0
Czech Rep.	4	1.2	320	98.8	0	0.0	0	0.0	18	6.0	284	94.0	0	0.0	0	0.0
Denmark	0	0.0	209	99.1	0	0.0	2	1.0	1	0.3	300	98.4	0	0.0	4	1.3
Finland	0	0.0	26	100	0	0.0	0	0.0	0	0.0	30	96.8	0	0.0	1	3.2
France	2691	100	-	-	-	-	-	-	3684	100	-	-	-	-	-	-
Georgia	1	2.4	39	95.1	0	0.0	1	2.4								
Germany	52	1.9	2701	98.0	2	0.1	1	0.0	106	2.8	3674	96.3	19	0.5	17	0.5
Greece	2	0.8	234	98.7	1	0.4	0	0.0	3	0.9	348	98.3	3	0.9	0	0.0
Hungary	2	0.8	233	98.3	1	0.4	1	0.4	3	1.6	186	95.9	2	1.0	3	1.6
Iceland	0	0.0	9	100	0	0.0	0	0.0	0	0.0	6	100	0	0.0	0	0.0
Ireland	0	0.0	518	98.7	4	0.8	3	0.6	0	0.0	667	92.8	50	7.0	2	0.3
Israel	2	1.3	155	97.5	2	1.3	0	0.0	11	3.2	318	93.5	10	2.9	1	0.3
Italy	179	8.1	1988	90.2	5	0.2	32	1.5	497	13.9	2976	83.3	36	1.0	64	1.8
Latvia	0	0.0	32	100	0	0.0	0	0.0	0	0.0	14	100	0	0.0	0	0.0
Lithuania	1	5.0	19	95.0	0	0.0	0	0.0	2	8.7	20	87.0	1	4.4	0	0.0
Luxembourg	0	0.0	21	100	0	0.0	0	0.0	0	0.0	4	80.0	1	20.0	0	0.0
Rep. Moldova	0	0.0	36	100	0	0.0	0	0.0	0	0.0	12	100	0	0.0	0	0.0
Netherlands	6	1.1	533	98.9	0	0.0	0	0.0	16	1.7	953	98.3	1	0.1	0	0.0
N. Macedonia	0	0.0	74	100	0	0.0	0	0.0	0	0.0	49	100	0	0.0	0	0.0
Norway	1	0.8	127	99.2	0	0.0	0	0.0	4	2.1	190	97.4	0	0.0	1	0.5
Poland	5	0.5	947	99.4	1	0.1	0	0.0	6	1.1	513	96.4	11	2.1	2	0.4
Portugal	1	0.6	165	96.5	5	2.9	0	0.0	2	1.1	183	96.8	3	1.6	1	0.5
Romania	6	2.3	256	97.7	0	0.0	0	0.0	0	0.0	24	100	0	0.0	0	0.0
Russian Fed.	10	0.5	1858	99.2	0	0.0	6	0.3	23	4.5	478	93.7	7	1.4	2	0.4
Serbia	0	0.0	133	97.8	3	2.2	0	0.0	1	1.5	63	95.5	2	3.0	0	0.0
Slovak Rep.	1	0.8	125	99.2	0	0.0	0	0.0	0	0.0	143	97.3	4	2.7	0	0.0
Slovenia	1	1.8	54	98.2	0	0.0	0	0.0	1	2.2	45	97.8	0	0.0	0	0.0
Spain	5	0.5	1053	99.4	1	0.1	0	0.0	9	0.8	1187	98.3	4	0.3	7	0.6
Sweden	19	7.2	245	92.5	1	0.4	0	0.0	11	2.8	373	95.9	5	1.3	0	0.0
Switzerland	4	1.0	402	98.8	1	0.3	0	0.0	8	1.5	531	97.6	0	0.0	5	0.9
Turkey	2	0.1	2095	97.9	44	2.1	0	0.0	1	0.3	349	94.6	19	5.2	0	0.0
Ukraine	6	2.3	258	97.4	0	0.0	1	0.4	2	2.5	76	96.2	0	0.0	1	1.3
United Kingdom	0	0.0	4138	100	0	0.0	0	0.0	0	0.0	5781	99.9	0	0.0	5	0.1

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, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the table for adults, but they are included in the total number.

Note: NIPPV is reimbursed in most countries except in Albania, Armenia, Belarus, Bulgaria, the Republic of Moldova, Serbia 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 2022 who have never had a transplant, by country (table A8.8, Appendix 1).
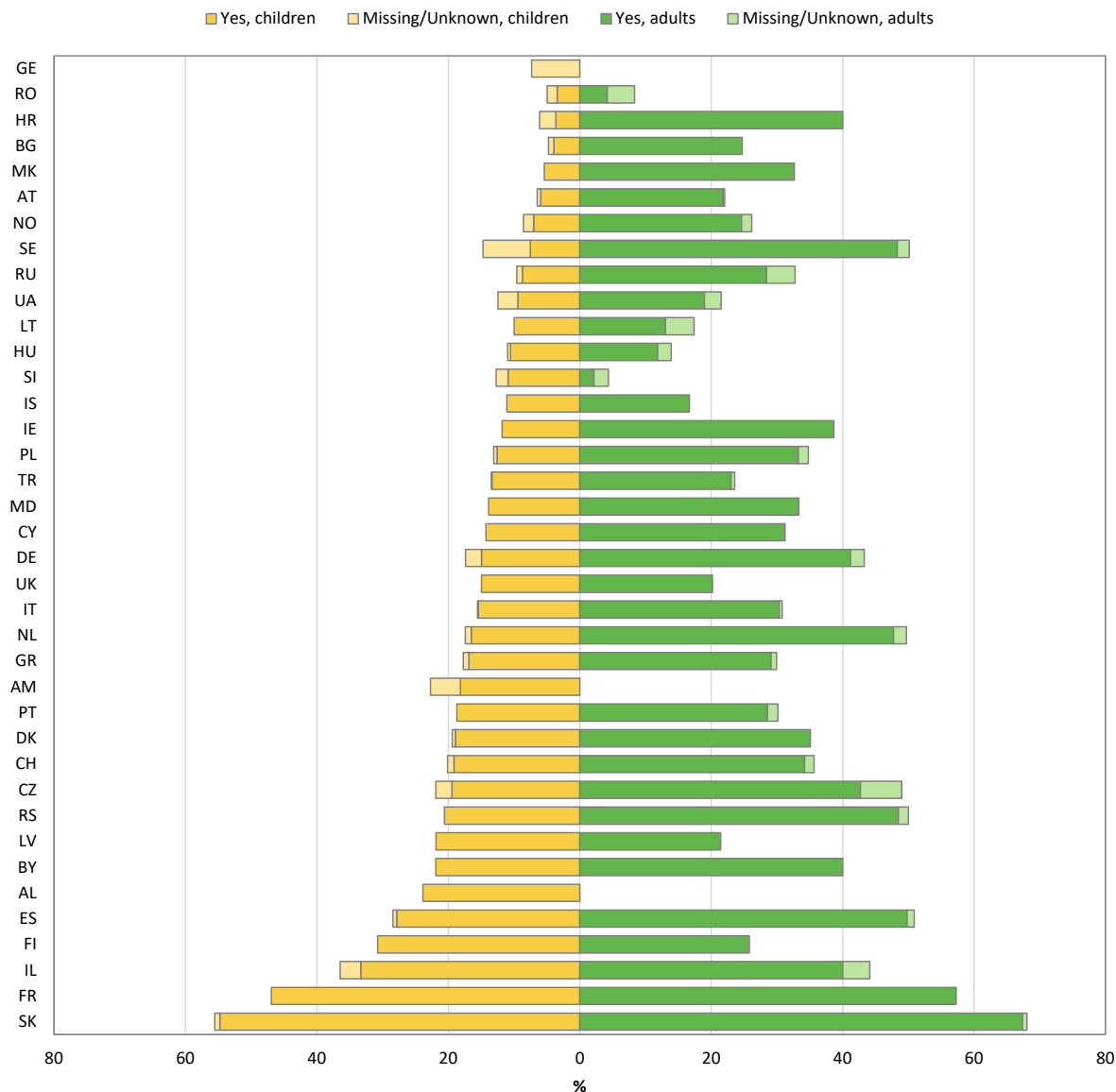
Note: We excluded from the graph the countries for which the information on use of inhaled steroids is missing for more than 10% of the children/adults with CF. Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the graph for adults.

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

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

Use of oral steroids in children and adults seen in 2022 who have never had a transplant, by country (table A8.9, Appendix 1).
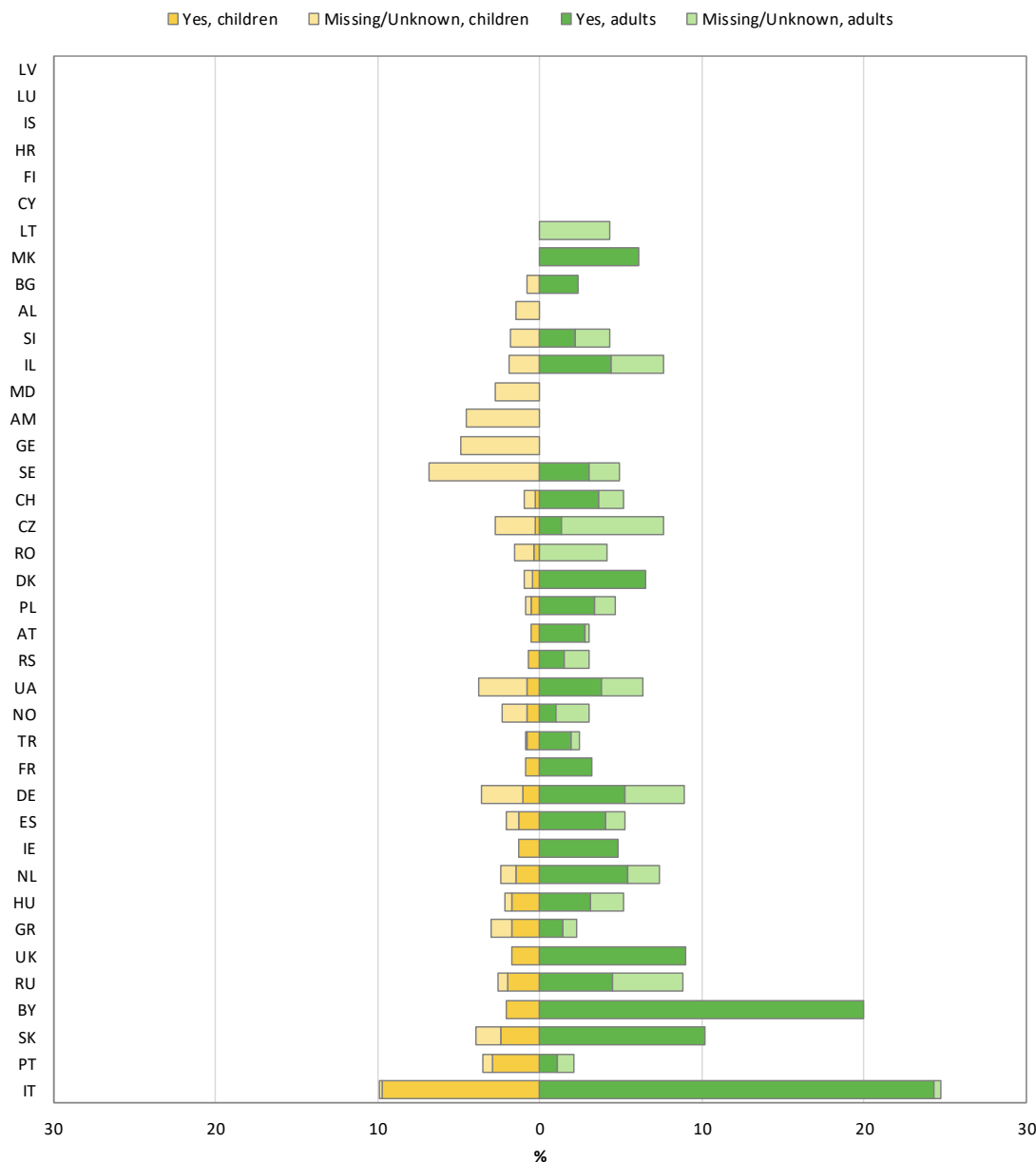
Note: We excluded from the graph the countries for which the information on use of oral steroids is missing for more than 10% of the children/adults with CF.

Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the graph for adults.

Note: Oral steroids are reimbursed in most countries except in Bulgaria, Georgia, 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 2022 who have never had a transplant, by country (table A8.10, Appendix 1).
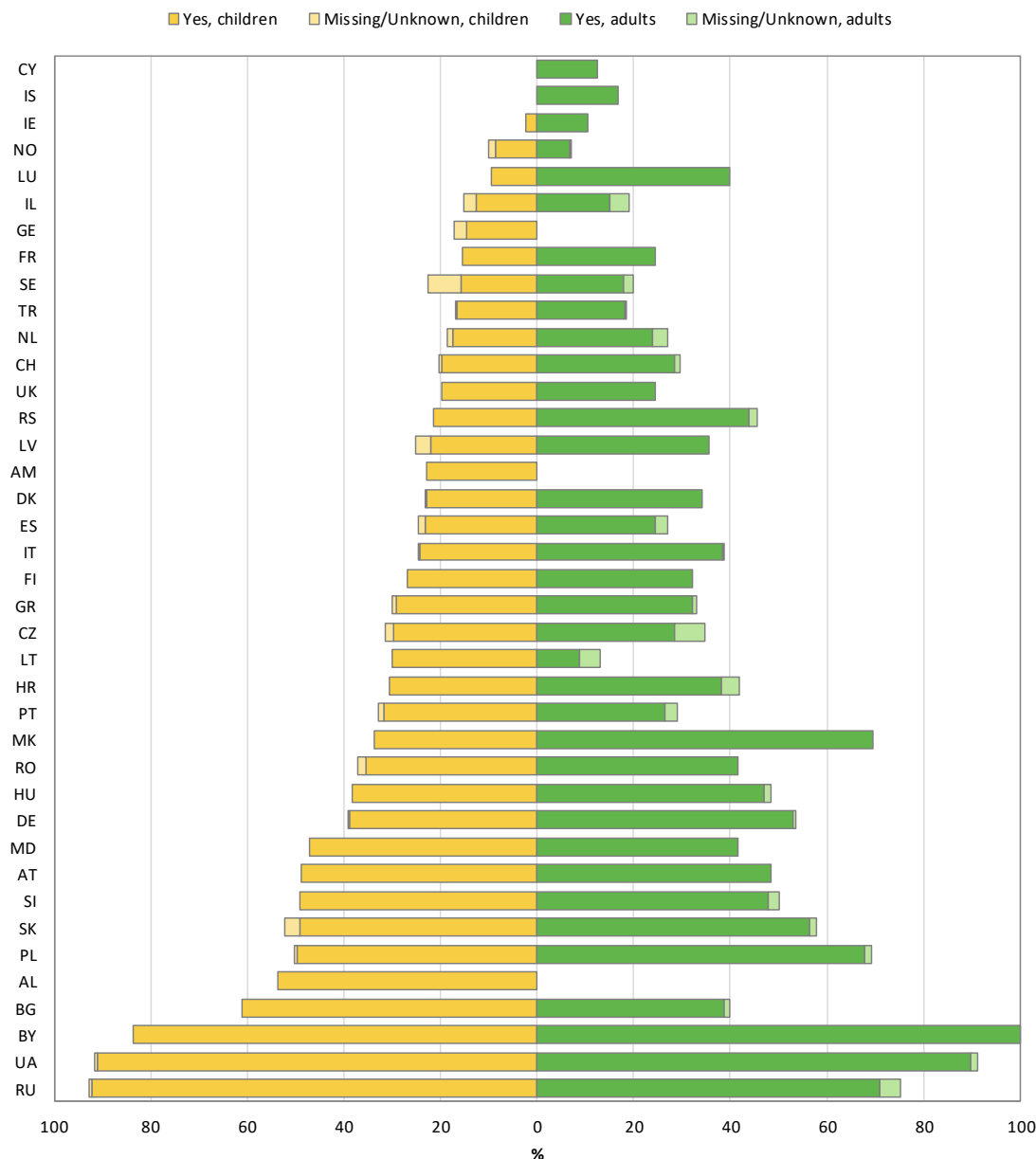
Note: We excluded from the graph the countries for which the information on oral ursodeoxycholic acid use is missing for more than 10% of the children/adults with CF. Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the graph for adults

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. In Ukraine it is reimbursed for children.

This graph shows how many people with CF used ursodeoxycholic acid for at least three consecutive months during 2022. 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, or to enhance pancreatic enzyme efficacy.

Use of proton pump inhibitors (PPI) in children and adults seen in 2022 who have never had a transplant, by country (table A8.11, Appendix 1).
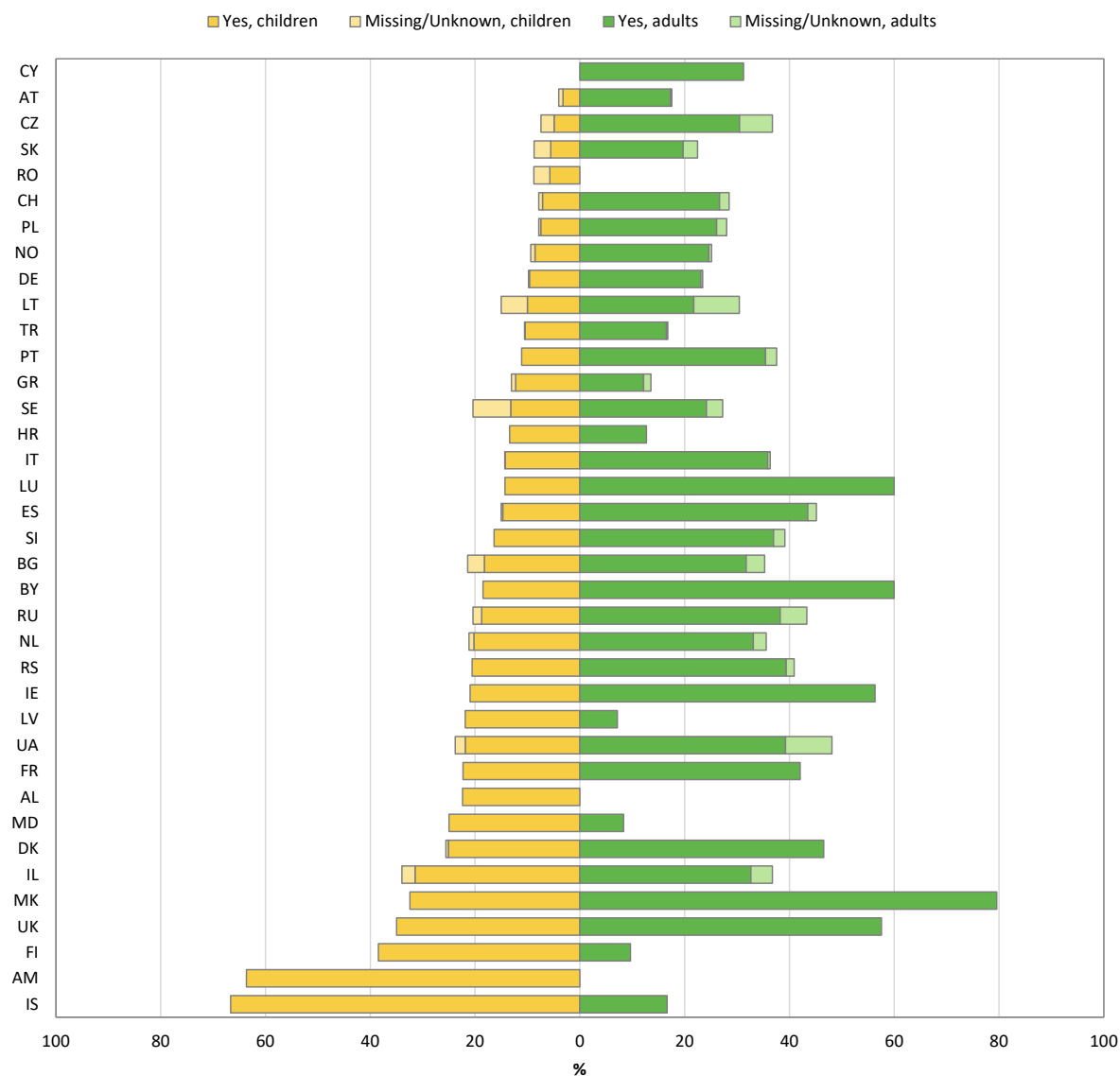
Note: We excluded from the graph the countries for which the information on the use of PPI is missing for more than 10% of the children/adults with CF.

Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the graph for adults.

Note: Oral proton pump inhibitors are reimbursed in most countries except in Bulgaria, Georgia, Lithuania, the Republic of Moldova and Serbia.

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 2012 and 2022
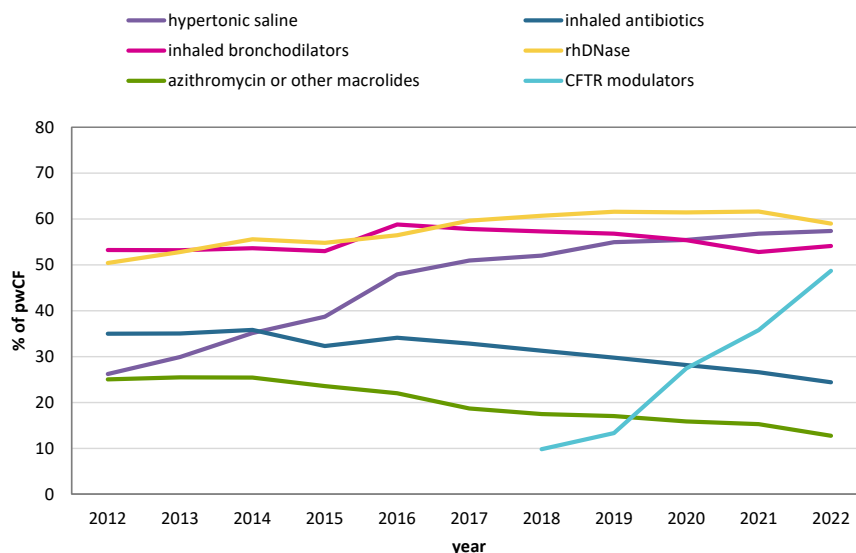
Figure 8.12   The increased use of CFTR modulators in adults with CF in Europe goes hand in hand with a decrease in the prescription of azithromycin and inhaled antibiotics, while that of inhaled mucolytics remains mostly unchanged.

Therapy use in adults between 2012 and 2022.
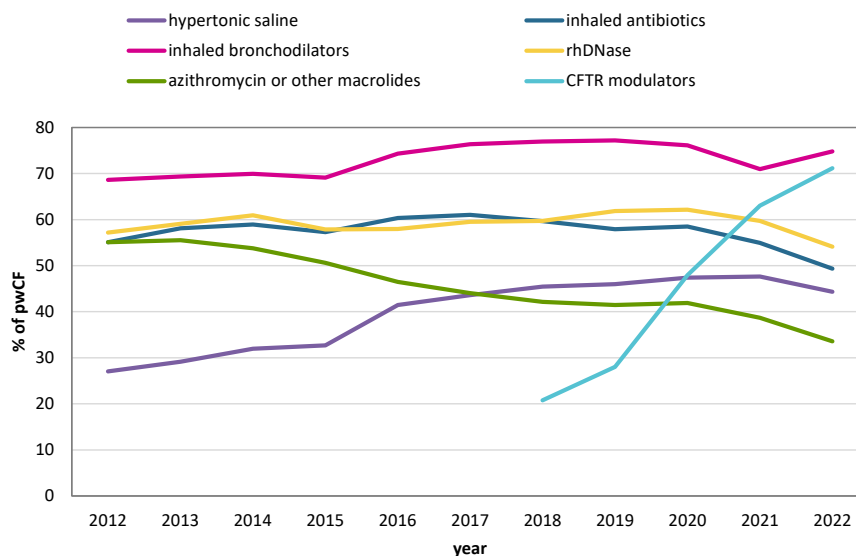
Figures 8.11 and 8.12 present data over time using cross sectional data per year of people with a confirmed CF. All people with CF alive, deceased, or not seen during the year of follow-up were included. When computing the yearly prevalence for each variable people who were lost to follow-up, transplanted (lung and/or liver) or with missing values 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 protein. However, CFTR modulators are only effective in people with specific variant classes since different variants cause different defects in the protein.

In this chapter we present information about the use of the different CFTR modulators for people with CF. Maps show the availability of these therapies and whether they were reimbursed or not by national health services in the countries participating in the ECFSPR in 2022. These maps help with the interpretation of the results in the tables and illustrate the country-specific variations in therapy use.

We adopted the eligibility criteria of the European Medicines Agency (EMA) for all ECFSPR countries for 2022, except for Israel, the Russian Federation and Switzerland where country-specific eligibility criteria laid down by the national regulatory authorities have been applied. Where these are different from EMA criteria, it has been stated.

The eligibility criteria for the CFTR modulators in 2022 were:

ivacaftor:

at least 4 months old (1 year in Israel) 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).

lumacaftor/ivacaftor:

at least 2 years old (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 6 years old and have at least one F508del variant.

In countries where a therapy was licensed in 2022 but not reimbursed, or not licensed, people may sometimes have had access to the therapy through a clinical trial or a compassionate use programme.

9. CFTR modulator therapies

Figure 9.1   Countries where ivacaftor was licensed and reimbursed in 2022.

Legend:
- Licensed and reimbursed ≥ 4 months
- Licensed and reimbursed ≥ 6 months
- Licensed and reimbursed ≥ 1 year
- Licensed and reimbursed ≥ 2 years
- Licensed and reimbursed ≥ 6 years
- Licensed and reimbursed ≥ 12 years
- No access
- NA
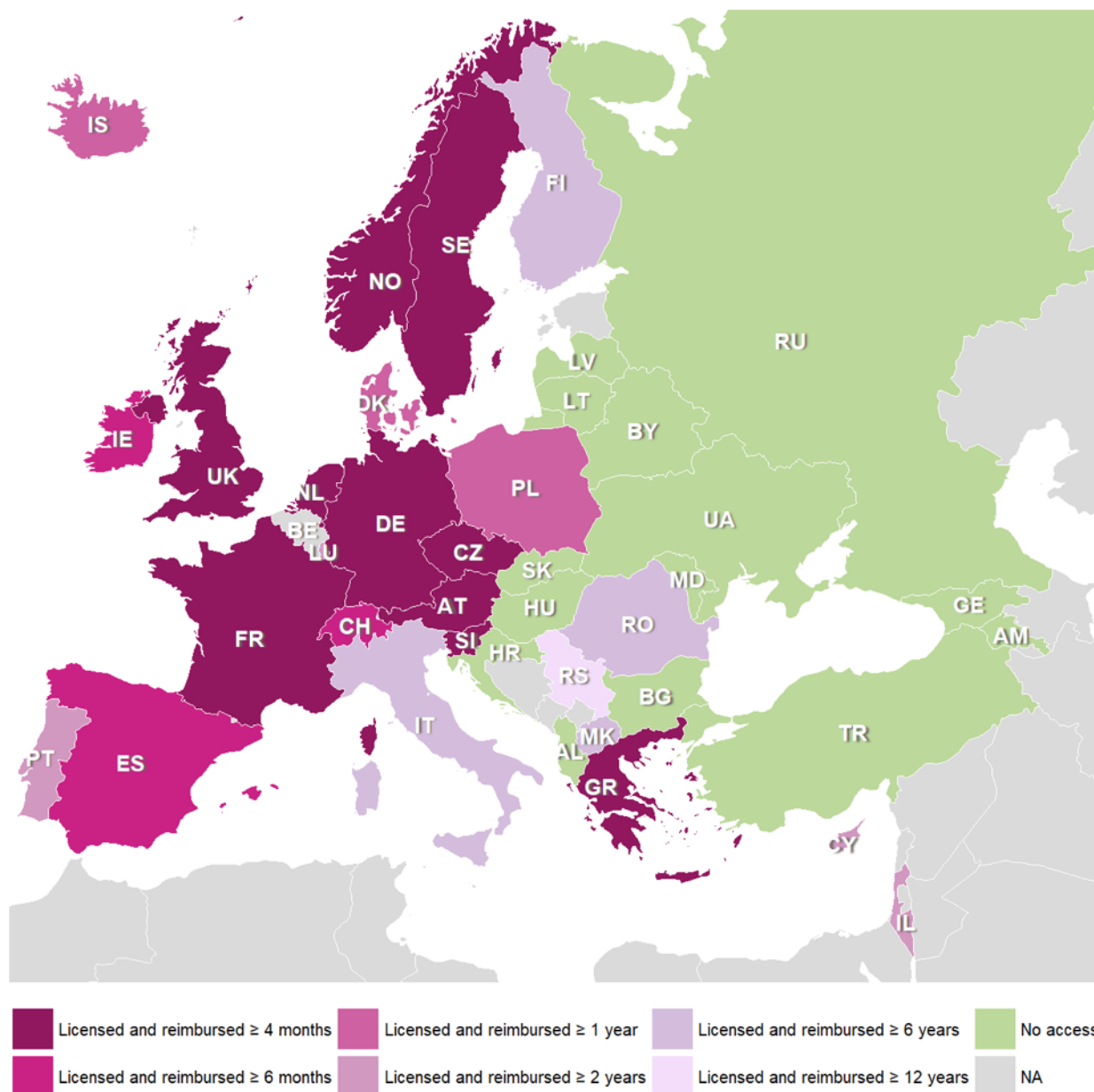
Note: Iceland: ivacaftor was reimbursed for people with CF with the variant F508del and/or G551D.

Norway: ivacaftor was reimbursed for children with CF if ≥ 5 kg.

Spain: ivacaftor was reimbursed for children with CF ≥ 6 months from 1 July 2022.

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 2022.

Legend: Licensed and reimbursed ≥ 2 year | Licensed and reimbursed ≥ 6 years | No access | NA
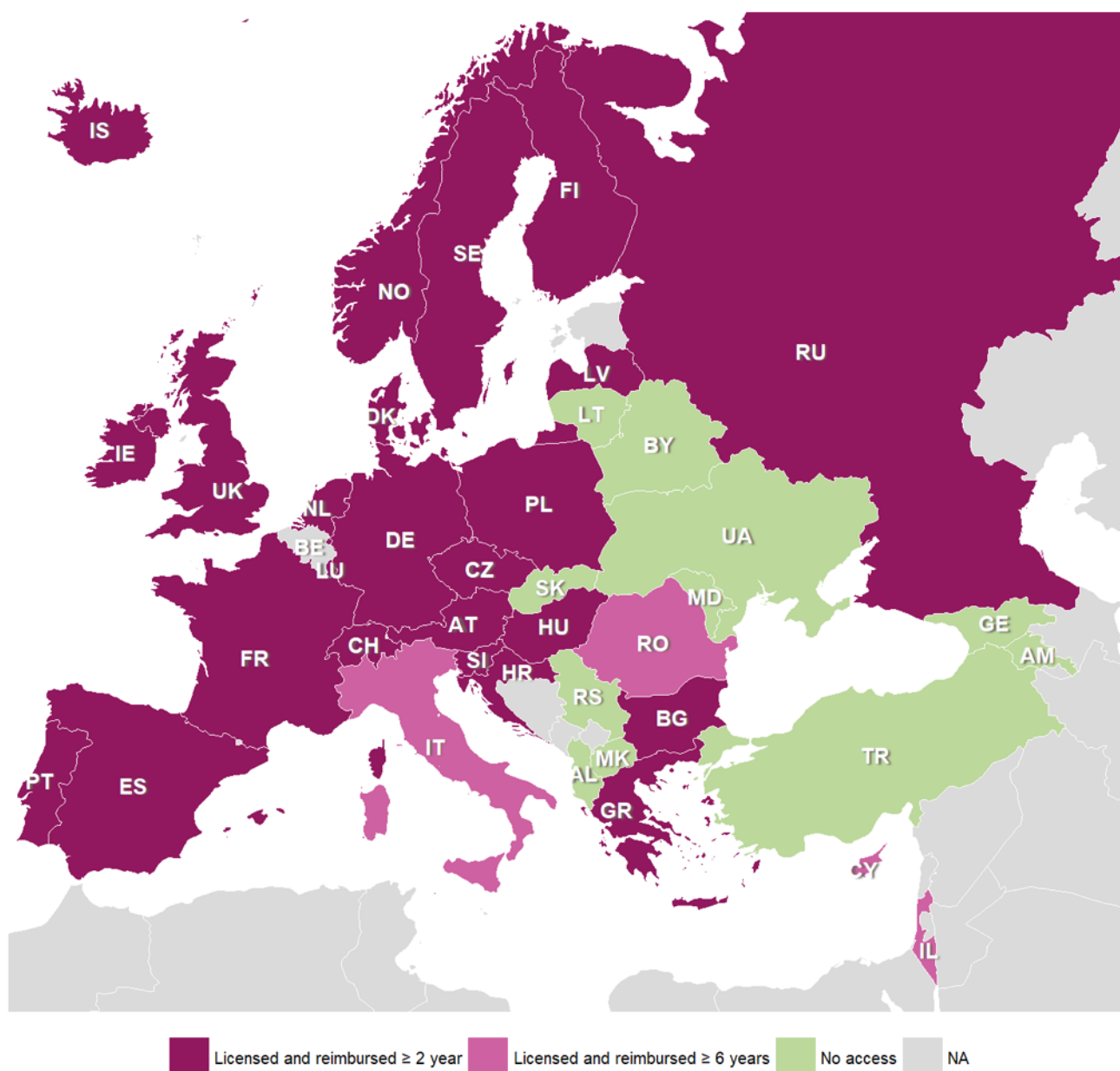
Note: Spain: lumacaftor/ivacaftor was reimbursed for children with CF between 2 and 5 years old.

Russian Federation: lumacaftor/ivacaftor was reimbursed for people with CF between 2 and 18 years old.

9. CFTR modulator therapies

Figure 9.3   Countries where tezacaftor/ivacaftor was licensed and reimbursed in 2022.

Legend: Licensed and reimbursed ≥ 6 years | Licensed and reimbursed ≥ 12 years | No access | NA
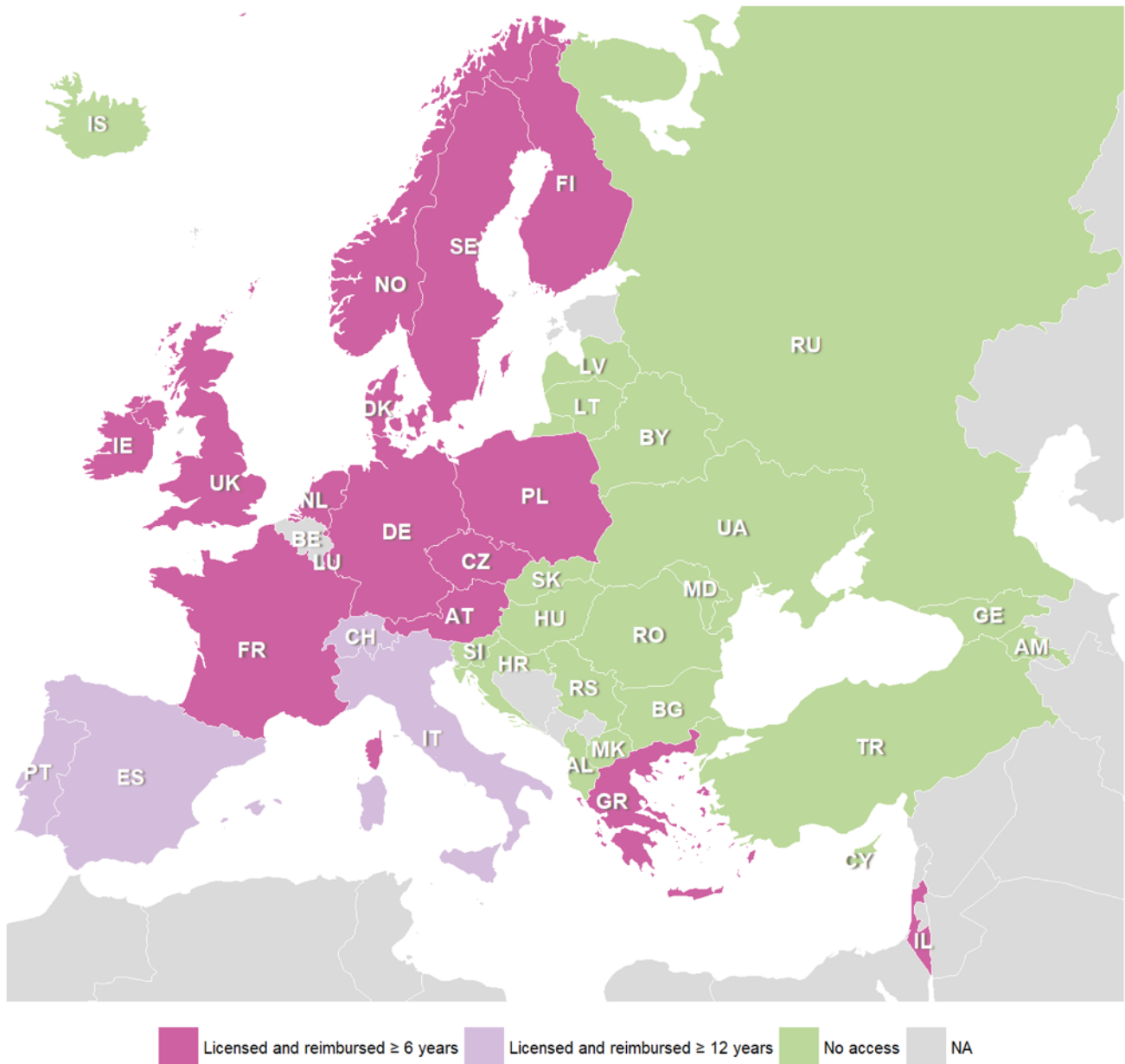
Note: Sweden: tezacaftor/ivacaftor was reimbursed from December 2022.

9. CFTR modulator therapies

Figure 9.4   Countries where elexacaftor/tezacaftor/ivacaftor was is licensed and reimbursed in 2022.

■ Licensed and reimbursed ≥ 6 years	■ Licensed and reimbursed ≥ 12 years	■ No access	■ NA
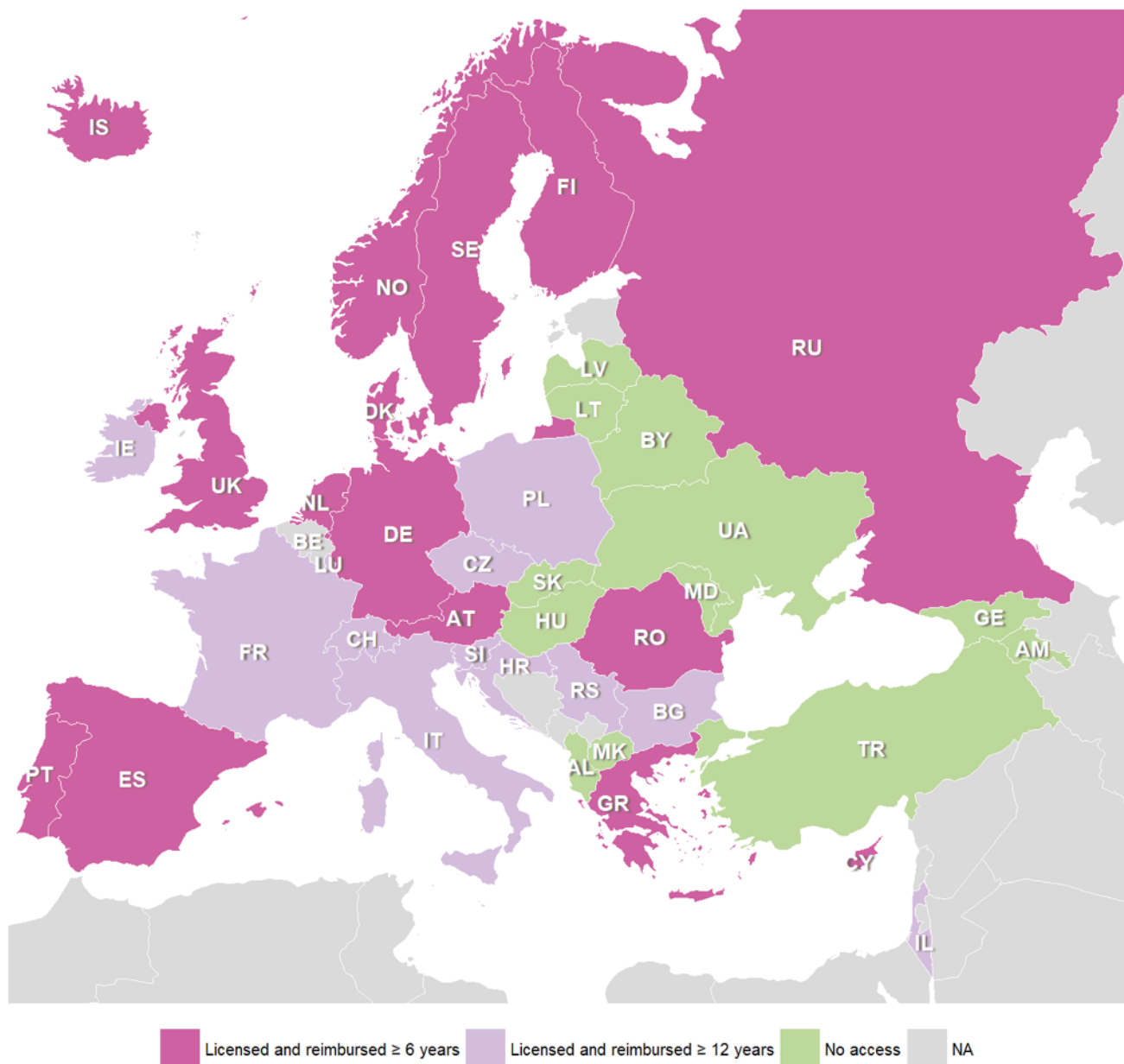
Note: Czech Republic: from September 2022, elexacaftor/tezacaftor/ivacaftor was reimbursed for people with CF ≥ 6 years.

France: from December 2022 elexacaftor/tezacaftor/ivacaftor was reimbursed for people with CF ≥ 6 years and for some people (early access programme), from March 2022)

Russian Federation: elexacaftor/tezacaftor/ivacaftor was reimbursed for people with CF between 6 and 18 years old.

Sweden: elexacaftor/tezacaftor/ivacaftor was reimbursed from December 2022.

9. CFTR modulator therapies

Table 9.1   People with CF, by country, eligible for at least one modulator, by age and genotype. People with CF seen in 2022 who have never had a transplant.

| Country | | Not eligible | | | | | | Eligible | | | |
| | | Genotyping not done | | At least one F508del | | Not F508del | | At least one F508del | | Not F508del | |
		N	%	N	%	N	%	N	%	N	%
Albania	0-1	0	0.0	1	100.0	0	0.0	0	0.0	0	0.0
	2-5	2	28.6	2	28.6	1	14.3	2	28.6	0	0.0
	6-11	3	8.3	0	0.0	1	2.8	32	88.9	0	0.0
	12-17	0	0.0	0	0.0	3	13.0	19	82.6	1	4.4
	18+	0	0.0	0	0.0	0	0.0	2	100.0	0	0.0
Armenia	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	1	20.0	3	60.0	1	20.0	0	0.0
	6-11	0	0.0	0	0.0	10	90.9	1	9.1	0	0.0
	12-17	0	0.0	0	0.0	6	100.0	0	0.0	0	0.0
	18+	0	0.0	0	0.0	3	100.0	0	0.0	0	0.0
Austria	0-1	0	0.0	25	92.6	2	7.4	0	0.0	0	0.0
	2-5	0	0.0	18	23.4	15	19.5	40	52.0	4	5.2
	6-11	0	0.0	0	0.0	11	8.7	113	89.7	2	1.6
	12-17	0	0.0	0	0.0	25	17.7	116	82.3	0	0.0
	18+	0	0.0	0	0.0	45	11.3	350	87.7	4	1.0
Belarus	0-1	0	0.0	9	81.8	2	18.2	0	0.0	0	0.0
	2-5	0	0.0	17	63.0	6	22.2	4	14.8	0	0.0
	6-11	0	0.0	0	0.0	10	16.1	52	83.9	0	0.0
	12-17	0	0.0	0	0.0	13	28.3	33	71.7	0	0.0
	18+	0	0.0	0	0.0	2	40.0	3	60.0	0	0.0
Bulgaria	0-1	0	0.0	4	57.1	3	42.9	0	0.0	0	0.0
	2-5	0	0.0	10	35.7	2	7.1	16	57.1	0	0.0
	6-11	0	0.0	0	0.0	4	8.3	43	89.6	1	2.1
	12-17	0	0.0	0	0.0	6	14.0	37	86.1	0	0.0
	18+	0	0.0	0	0.0	22	25.9	62	72.9	1	1.2
Croatia	0-1	0	0.0	5	83.3	1	16.7	0	0.0	0	0.0
	2-5	0	0.0	8	36.4	1	4.6	13	59.1	0	0.0
	6-11	0	0.0	0	0.0	0	0.0	23	100.0	0	0.0
	12-17	0	0.0	0	0.0	1	3.2	30	96.8	0	0.0
	18+	0	0.0	0	0.0	1	1.8	54	98.2	0	0.0
Cyprus	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	2	100.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	1	25.0	3	75.0	0	0.0
	12-17	0	0.0	0	0.0	1	100.0	0	0.0	0	0.0
	18+	0	0.0	0	0.0	8	50.0	7	43.8	1	6.3
Czech Rep.	0-1	0	0.0	28	84.9	5	15.2	0	0.0	0	0.0
	2-5	0	0.0	32	42.1	4	5.3	39	51.3	1	1.3
	6-11	0	0.0	0	0.0	13	12.3	91	85.9	2	1.9
	12-17	0	0.0	0	0.0	12	11.0	96	88.1	1	0.9
	18+	1	0.3	0	0.0	33	10.9	259	85.8	9	3.0
Denmark	0-1	0	0.0	24	100.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	17	27.9	0	0.0	44	72.1	0	0.0
	6-11	0	0.0	0	0.0	4	6.6	57	93.4	0	0.0
	12-17	0	0.0	0	0.0	1	1.5	64	98.5	0	0.0
	18+	0	0.0	0	0.0	12	3.9	291	95.4	2	0.7
Finland	0-1	0	0.0	0	0.0	1	100.0	0	0.0	0	0.0
	2-5	0	0.0	1	25.0	1	25.0	2	50.0	0	0.0
	6-11	0	0.0	0	0.0	3	37.5	5	62.5	0	0.0
	12-17	0	0.0	0	0.0	5	38.5	8	61.5	0	0.0
	18+	1	3.2	0	0.0	9	29.0	21	67.7	0	0.0
France	0-1	0	0.0	189	79.8	43	18.1	4	1.7	1	0.4
	2-5	0	0.0	187	36.6	76	14.9	243	47.6	5	1.0
	6-11	0	0.0	0	0.0	148	16.6	736	82.5	8	0.9
	12-17	0	0.0	0	0.0	143	13.6	889	84.6	19	1.8
	18+	0	0.0	0	0.0	557	15.1	3066	83.2	61	1.7
Georgia	0-1	0	0.0	0	0.0	1	100.0	0	0.0	0	0.0
	2-5	3	27.3	2	18.2	5	45.5	1	9.1	0	0.0
	6-11	1	4.8	0	0.0	18	85.7	2	9.5	0	0.0

| Country | | Not eligible | | | | | | Eligible | | | |
| | | Genotyping not done | | At least one F508del | | Not F508del | | At least one F508del | | Not F508del | |
		N	%	N	%	N	%	N	%	N	%
	12-17	0	0.0	0	0.0	7	87.5	1	12.5	0	0.0
	18+	1	100.0	0	0.0	0	0.0	0	0.0	0	0.0
Germany	0-1	2	0.8	197	83.1	32	13.5	4	1.7	2	0.8
	2-5	11	1.8	248	39.4	75	11.9	290	46.1	5	0.8
	6-11	0	0.0	0	0.0	130	13.4	827	85.4	11	1.1
	12-17	0	0.0	0	0.0	101	11.0	806	87.4	15	1.6
	18+	2	0.1	0	0.0	441	11.6	3314	86.8	59	1.6
Greece	0-1	0	0.0	15	83.3	3	16.7	0	0.0	0	0.0
	2-5	0	0.0	10	28.6	11	31.4	14	40.0	0	0.0
	6-11	0	0.0	0	0.0	12	15.6	64	83.1	1	1.3
	12-17	0	0.0	0	0.0	23	21.5	84	78.5	0	0.0
	18+	2	0.6	0	0.0	76	21.5	274	77.4	2	0.6
Hungary	0-1	0	0.0	15	83.3	3	16.7	0	0.0	0	0.0
	2-5	0	0.0	19	38.8	6	12.2	24	49.0	0	0.0
	6-11	0	0.0	0	0.0	11	11.2	86	87.8	1	1.0
	12-17	0	0.0	0	0.0	9	12.5	63	87.5	0	0.0
	18+	0	0.0	0	0.0	32	16.5	162	83.5	0	0.0
Iceland	0-1	0	0.0	1	100.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	1	100.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	2	50.0	2	50.0	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	2	66.7	1	33.3
	18+	0	0.0	0	0.0	1	16.7	5	83.3	0	0.0
Ireland	0-1	0	0.0	22	71.0	2	6.5	7	22.6	0	0.0
	2-5	0	0.0	29	25.9	7	6.3	67	59.8	9	8.0
	6-11	0	0.0	0	0.0	9	5.1	161	90.5	8	4.5
	12-17	0	0.0	0	0.0	6	2.9	192	94.1	6	2.9
	18+	0	0.0	0	0.0	17	2.4	665	92.5	37	5.2
Israel	0-1	0	0.0	1	16.7	5	83.3	0	0.0	0	0.0
	2-5	1	6.7	4	26.7	10	66.7	0	0.0	0	0.0
	6-11	0	0.0	9	17.0	34	64.2	10	18.9	0	0.0
	12-17	0	0.0	0	0.0	51	60.0	34	40.0	0	0.0
	18+	1	0.3	0	0.0	187	55.0	144	42.4	8	2.4
Italy	0-1	0	0.0	92	70.8	36	27.7	0	0.0	2	1.5
	2-5	0	0.0	194	44.8	128	29.6	99	22.9	12	2.8
	6-11	0	0.0	0	0.0	245	29.0	579	68.6	20	2.4
	12-17	0	0.0	0	0.0	257	32.3	524	65.8	16	2.0
	18+	1	0.0	0	0.0	1070	30.0	2430	68.0	72	2.0
Latvia	0-1	0	0.0	1	100.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	1	9.1	3	27.3	7	63.6	0	0.0
	6-11	0	0.0	0	0.0	2	20.0	8	80.0	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	10	100.0	0	0.0
	18+	0	0.0	0	0.0	0	0.0	14	100.0	0	0.0
Lithuania	0-1	0	0.0	1	100.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	4	57.1	0	0.0	3	42.9	0	0.0
	6-11	0	0.0	0	0.0	0	0.0	5	100.0	0	0.0
	12-17	0	0.0	0	0.0	3	42.9	4	57.1	0	0.0
	18+	0	0.0	0	0.0	5	21.7	18	78.3	0	0.0
Luxemburg	0-1	0	0.0	4	100.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	5	62.5	1	12.5	2	25.0	0	0.0
	6-11	0	0.0	0	0.0	2	40.0	3	60.0	0	0.0
	12-17	0	0.0	0	0.0	1	25.0	3	75.0	0	0.0
	18+	0	0.0	0	0.0	3	60.0	2	40.0	0	0.0
Moldova	0-1	0	0.0	5	100.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	2	50.0	2	50.0	0	0.0
	6-11	0	0.0	0	0.0	2	14.3	12	85.7	0	0.0
	12-17	0	0.0	0	0.0	4	30.8	8	61.5	1	7.7
	18+	0	0.0	0	0.0	6	50.0	6	50.0	0	0.0
Netherlands	0-1	0	0.0	34	89.5	4	10.5	0	0.0	0	0.0
	2-5	0	0.0	27	26.5	5	4.9	69	67.7	1	1.0
	6-11	0	0.0	0	0.0	19	10.0	168	88.4	3	1.6
	12-17	0	0.0	0	0.0	14	6.7	192	91.9	3	1.4
	18+	0	0.0	0	0.0	77	7.9	884	91.1	9	0.9
N. Macedonia	0-1	0	0.0	5	83.3	0	0.0	1	16.7	0	0.0

Country		Not eligible						Eligible			
		Genotyping not done		At least one F508del		Not F508del		At least one F508del		Not F508del	
		N	%	N	%	N	%	N	%	N	%
	2-5	0	0.0	9	34.6	2	7.7	15	57.7	0	0.0
	6-11	0	0.0	0	0.0	0	0.0	15	100.0	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	27	100.0	0	0.0
	18+	1	2.0	0	0.0	7	14.3	41	83.7	0	0.0
Norway	0-1	0	0.0	10	83.3	2	16.7	0	0.0	0	0.0
	2-5	0	0.0	20	58.8	4	11.8	10	29.4	0	0.0
	6-11	0	0.0	0	0.0	10	27.0	27	73.0	0	0.0
	12-17	0	0.0	0	0.0	8	17.8	37	82.2	0	0.0
	18+	0	0.0	0	0.0	22	11.3	166	85.1	7	3.6
Poland	0-1	0	0.0	62	89.9	5	7.3	1	1.5	1	1.5
	2-5	0	0.0	80	39.6	37	18.3	84	41.6	1	0.5
	6-11	0	0.0	0	0.0	47	13.8	290	85.3	3	0.9
	12-17	0	0.0	0	0.0	42	12.3	300	87.7	0	0.0
	18+	1	0.2	0	0.0	64	12.0	465	87.4	2	0.4
Portugal	0-1	0	0.0	8	88.9	1	11.1	0	0.0	0	0.0
	2-5	0	0.0	16	51.6	6	19.4	8	25.8	1	3.2
	6-11	0	0.0	0	0.0	9	13.9	56	86.2	0	0.0
	12-17	0	0.0	0	0.0	9	13.6	57	86.4	0	0.0
	18+	0	0.0	0	0.0	37	19.6	150	79.4	2	1.1
Romania	0-1	0	0.0	13	86.7	2	13.3	0	0.0	0	0.0
	2-5	1	1.4	28	37.8	9	12.2	36	48.7	0	0.0
	6-11	0	0.0	0	0.0	10	13.0	66	85.7	1	1.3
	12-17	0	0.0	0	0.0	12	12.5	84	87.5	0	0.0
	18+	0	0.0	0	0.0	5	20.8	19	79.2	0	0.0
Russian Fed.	0-1	25	17.5	88	61.5	30	21.0	0	0.0	0	0.0
	2-5	18	4.8	138	36.6	102	27.1	119	31.6	0	0.0
	6-11	12	1.6	0	0.0	164	21.2	596	77.2	0	0.0
	12-17	8	1.4	0	0.0	127	21.8	447	76.8	0	0.0
	18+	14	2.8	0	0.0	111	21.8	385	75.5	0	0.0
Serbia	0-1	0	0.0	13	100.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	19	48.7	2	5.1	18	46.2	0	0.0
	6-11	0	0.0	0	0.0	2	4.9	39	95.1	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	43	100.0	0	0.0
	18+	3	4.6	0	0.0	9	13.6	54	81.8	0	0.0
Slovak Rep.	0-1	0	0.0	9	90.0	1	10.0	0	0.0	0	0.0
	2-5	0	0.0	4	17.4	4	17.4	15	65.2	0	0.0
	6-11	0	0.0	0	0.0	10	19.2	42	80.8	0	0.0
	12-17	0	0.0	0	0.0	8	19.5	31	75.6	2	4.9
	18+	0	0.0	0	0.0	29	19.7	113	76.9	5	3.4
Slovenia	0-1	0	0.0	2	100.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	3	50.0	0	0.0	3	50.0	0	0.0
	6-11	0	0.0	0	0.0	1	3.9	25	96.2	0	0.0
	12-17	0	0.0	0	0.0	3	14.3	18	85.7	0	0.0
	18+	0	0.0	0	0.0	7	15.2	39	84.8	0	0.0
Spain	0-1	0	0.0	53	69.7	22	29.0	0	0.0	1	1.3
	2-5	0	0.0	87	46.0	49	25.9	51	27.0	2	1.1
	6-11	0	0.0	0	0.0	98	24.6	299	74.9	2	0.5
	12-17	0	0.0	0	0.0	71	18.0	321	81.3	3	0.8
	18+	0	0.0	0	0.0	315	26.1	875	72.5	17	1.4
Sweden	0-1	0	0.0	16	100.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	9	17.0	13	24.5	31	58.5	0	0.0
	6-11	0	0.0	0	0.0	10	11.8	74	87.1	1	1.2
	12-17	0	0.0	0	0.0	16	14.4	93	83.8	2	1.8
	18+	0	0.0	0	0.0	47	12.1	334	85.9	8	2.1
Switzerland	0-1	0	0.0	21	84.0	2	8.0	2	8.0	0	0.0
	2-5	0	0.0	35	41.7	14	16.7	35	41.7	0	0.0
	6-11	0	0.0	0	0.0	23	15.0	130	85.0	0	0.0
	12-17	0	0.0	0	0.0	16	11.0	128	88.3	1	0.7
	18+	0	0.0	0	0.0	77	14.2	461	84.7	6	1.1
Turkey	0-1	33	17.2	49	25.5	108	56.3	0	0.0	2	1.0
	2-5	19	3.3	95	16.3	394	67.6	65	11.2	10	1.7
	6-11	22	2.8	0	0.0	533	67.2	228	28.8	10	1.3
	12-17	17	3.0	0	0.0	388	67.7	163	28.5	5	0.9

Country		Not eligible						Eligible			
		Genotyping not done		At least one F508del		Not F508del		At least one F508del		Not F508del	
		N	%	N	%	N	%	N	%	N	%
	18+	10	2.7	0	0.0	231	62.6	122	33.1	6	1.6
Ukraine	0-1	0	0.0	12	85.7	2	14.3	0	0.0	0	0.0
	2-5	1	1.9	28	53.9	3	5.8	20	38.5	0	0.0
	6-11	2	1.8	0	0.0	12	10.5	100	87.7	0	0.0
	12-17	0	0.0	0	0.0	15	17.7	70	82.4	0	0.0
	18+	0	0.0	0	0.0	17	21.5	62	78.5	0	0.0
United Kingdom	0-1	1	0.6	149	83.7	8	4.5	13	7.3	7	3.9
	2-5	9	1.0	271	31.3	61	7.1	507	58.6	17	2.0
	6-11	4	0.3	0	0.0	91	5.8	1451	91.8	35	2.2
	12-17	0	0.0	0	0.0	132	8.7	1342	88.6	40	2.6
	18+	8	0.1	0	0.0	458	7.9	5135	88.8	185	3.2
Total	0-1	61	3.8	1183	73.1	326	20.2	32	2.0	16	1.0
	2-5	65	1.3	1678	34.4	1065	21.9	1999	41.0	68	1.4
	6-11	44	0.5	9	0.1	1711	20.4	6521	77.7	109	1.3
	12-17	25	0.3	0	0.0	1539	19.1	6376	79.2	116	1.4
	18+	46	0.2	0	0.0	4043	16.1	20454	81.7	503	2.0

9. CFTR modulator therapies

Table 9.2   People with CF with at least one F508del variant and eligible for at least one modulator, by country, by age and last CFTR modulator prescribed. People with CF seen in 2022 who have never had a transplant.

Country		Missing/ Unknown		No		Ivacaftor		Lumacaftor/ Ivacaftor		Tezacaftor/ Ivacaftor		Elexacaftor/ Tezacaftor/ Ivacaftor		Other CFTR modulator	
		N	%	N	%	N	%	N	%	N	%	N	%	N	%
Albania	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	2	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	32	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	19	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	2	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Armenia	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	1	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	1	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Austria	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	16	40.0	3	7.5	20	50.0	0	0.0	1	2.5	0	0.0
	6-11	0	0.0	35	31.0	2	1.8	11	9.7	1	0.9	64	56.6	0	0.0
	12-17	0	0.0	28	24.1	1	0.9	5	4.3	0	0.0	81	69.8	1	0.9
	18+	0	0.0	81	23.1	9	2.6	2	0.6	8	2.3	246	70.3	4	1.1
Belarus	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	4	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	52	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	33	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	3	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Bulgaria	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	8	50.0	0	0.0	8	50.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	25	58.1	1	2.3	17	39.5	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	9	24.3	2	5.4	13	35.1	1	2.7	12	32.4	0	0.0
	18+	0	0.0	10	16.1	0	0.0	10	16.1	0	0.0	42	67.7	0	0.0
Croatia	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	5	38.5	0	0.0	8	61.5	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	15	65.2	0	0.0	7	30.4	0	0.0	1	4.4	0	0.0
	12-17	0	0.0	5	16.7	0	0.0	0	0.0	0	0.0	25	83.3	0	0.0
	18+	0	0.0	7	13.0	0	0.0	0	0.0	1	1.9	46	85.2	0	0.0
Cyprus	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	0	0.0	1	33.3	0	0.0	2	66.7	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	7	100	0	0.0
Czech Rep.	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	6	15.4	1	2.6	32	82.1	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	18	19.8	0	0.0	15	16.5	1	1.1	57	62.6	0	0.0
	12-17	0	0.0	3	3.1	2	2.1	2	2.1	0	0.0	89	92.7	0	0.0
	18+	0	0.0	23	8.9	3	1.1	0	0.0	7	2.7	226	87.2	0	0.0
Denmark	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	3	6.8	0	0.0	41	93.2	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	2	3.5	0	0.0	0	0.0	0	0.0	55	96.5	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	64	100	0	0.0
	18+	0	0.0	15	5.2	0	0.0	0	0.0	2	0.7	264	90.7	10	3.4
Finland	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	2	100	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	2	40.0	0	0.0	3	60.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	8	100	0	0.0
	18+	0	0.0	2	9.5	0	0.0	0	0.0	0	0.0	19	90.5	0	0.0
France	0-1	2	50.0	0	0.0	2	50.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	21	8.6	0	0.0	7	2.9	212	87.2	0	0.0	3	1.2	0	0.0
	6-11	175	23.8	0	0.0	11	1.5	264	35.9	49	6.7	237	32.2	0	0.0
	12-17	80	9.0	0	0.0	7	0.8	18	2.0	6	0.7	778	87.5	0	0.0
	18+	367	12.0	0	0.0	30	1.0	16	0.5	72	2.4	2581	84.2	0	0.0

Country		Missing/ Unknown		No		Ivacaftor		Lumacaftor/ Ivacaftor		Tezacaftor/ Ivacaftor		Elexacaftor/ Tezacaftor/ Ivacaftor		Other CFTR modulator	
		N	%	N	%	N	%	N	%	N	%	N	%	N	%
Georgia	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	1	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	2	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	1	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Germany	0-1	0	0.0	3	75.0	1	25.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	97	33.5	10	3.5	181	62.4	0	0.0	2	0.7	0	0.0
	6-11	1	0.1	181	21.9	15	1.8	46	5.6	4	0.5	578	69.9	2	0.3
	12-17	1	0.1	94	11.7	19	2.4	13	1.6	10	1.2	669	83.0	0	0.0
	18+	4	0.1	332	10.0	45	1.4	22	0.7	53	1.6	2848	85.9	10	0.3
Greece	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	1	7.1	1	7.1	12	85.7	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	22	34.4	1	1.6	15	23.4	0	0.0	26	40.6	0	0.0
	12-17	0	0.0	15	17.9	0	0.0	8	9.5	2	2.4	59	70.2	0	0.0
	18+	0	0.0	28	10.2	1	0.4	6	2.2	4	1.5	235	85.8	0	0.0
Hungary	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	4	16.7	0	0.0	0	0.0	20	83.3	0	0.0	0	0.0	0	0.0
	6-11	33	38.4	0	0.0	0	0.0	42	48.8	0	0.0	11	12.8	0	0.0
	12-17	27	42.9	0	0.0	0	0.0	24	38.1	0	0.0	12	19.1	0	0.0
	18+	67	41.4	0	0.0	0	0.0	60	37.0	0	0.0	35	21.6	0	0.0
Iceland	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	2	100	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	2	100	0	0.0
	18+	0	0.0	1	20.0	0	0.0	0	0.0	0	0.0	4	80.0	0	0.0
Ireland	0-1	0	0.0	2	28.6	5	71.4	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	6	9.0	10	14.9	50	74.6	0	0.0	1	1.5	0	0.0
	6-11	0	0.0	32	19.9	10	6.2	1	0.6	0	0.0	118	73.3	0	0.0
	12-17	0	0.0	3	1.6	10	5.2	1	0.5	0	0.0	178	92.7	0	0.0
	18+	0	0.0	39	5.9	50	7.5	7	1.1	10	1.5	559	84.1	0	0.0
Israel	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	1	10.0	0	0.0	0	0.0	0	0.0	9	90.0	0	0.0
	12-17	0	0.0	4	11.8	1	2.9	0	0.0	1	2.9	28	82.4	0	0.0
	18+	0	0.0	12	8.3	0	0.0	1	0.7	1	0.7	123	85.4	7	4.9
Italy	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	49	49.5	4	4.0	46	46.5	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	312	53.9	14	2.4	66	11.4	1	0.3	186	32.1	0	0.0
	12-17	0	0.0	107	20.4	8	1.5	10	1.9	10	1.9	389	74.2	0	0.0
	18+	0	0.0	499	20.5	39	1.6	35	1.4	68	2.8	1788	73.6	1	0.0
Latvia	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	7	100	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	1	12.5	0	0.0	7	87.5	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	4	40.0	0	0.0	6	60.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	9	64.3	0	0.0	5	35.7	0	0.0	0	0.0	0	0.0
Lithuania	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	3	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	5	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	4	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	17	94.4	0	0.0	0	0.0	0	0.0	1	5.6	0	0.0
Luxemburg	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	2	100	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	3	100	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	3	100	0	0.0
	18+	0	0.0	1	50.0	0	0.0	0	0.0	0	0.0	1	50.0	0	0.0
Moldova	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	2	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	12	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	8	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	6	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0

Country		Missing/ Unknown		No		Ivacaftor		Lumacaftor/ Ivacaftor		Tezacaftor/ Ivacaftor		Elexacaftor/ Tezacaftor/ Ivacaftor		Other CFTR modulator	
		N	%	N	%	N	%	N	%	N	%	N	%	N	%
Netherlands	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	7	10.1	2	2.9	60	87.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	10	6.0	2	1.2	1	0.6	0	0.0	155	92.3	0	0.0
	12-17	0	0.0	9	4.7	1	0.5	0	0.0	1	0.5	181	94.3	0	0.0
	18+	1	0.1	47	5.3	12	1.4	6	0.7	15	1.7	800	90.5	3	0.3
N. Macedonia	0-1	0	0.0	1	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	15	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	15	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	26	96.3	1	3.7	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	39	95.1	2	4.9	0	0.0	0	0.0	0	0.0	0	0.0
Norway	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	5	50.0	0	0.0	5	50.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	2	7.4	0	0.0	0	0.0	0	0.0	25	92.6	0	0.0
	12-17	2	5.4	0	0.0	1	2.7	0	0.0	0	0.0	34	91.9	0	0.0
	18+	2	1.2	8	4.8	1	0.6	0	0.0	0	0.0	155	93.4	0	0.0
Poland	0-1	0	0.0	1	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	50	59.5	0	0.0	34	40.5	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	159	54.8	0	0.0	0	0.0	128	44.1	3	1.0	0	0.0
	12-17	1	0.3	45	15.0	0	0.0	0	0.0	16	5.3	234	78.0	4	1.3
	18+	14	3.0	57	12.3	1	0.2	1	0.2	35	7.5	354	76.1	3	0.7
Portugal	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	7	87.5	0	0.0	1	12.5	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	21	37.5	0	0.0	32	57.1	0	0.0	3	5.4	0	0.0
	12-17	0	0.0	6	10.5	0	0.0	6	10.5	0	0.0	45	78.9	0	0.0
	18+	0	0.0	26	17.3	0	0.0	1	0.7	0	0.0	122	81.3	1	0.7
Romania	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	36	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	36	54.5	0	0.0	14	21.2	0	0.0	16	24.2	0	0.0
	12-17	0	0.0	30	35.7	0	0.0	3	3.6	0	0.0	51	60.7	0	0.0
	18+	0	0.0	2	10.5	1	5.3	2	10.5	0	0.0	14	73.7	0	0.0
Russian Fed.	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	4	3.4	45	37.8	0	0.0	70	58.8	0	0.0	0	0.0	0	0.0
	6-11	32	5.4	226	37.9	0	0.0	165	27.7	1	0.2	172	28.9	0	0.0
	12-17	19	4.3	137	30.7	0	0.0	126	28.2	1	0.2	164	36.7	0	0.0
	18+	18	4.7	217	56.4	6	1.4	56	14.6	5	1.3	83	21.6	0	0.0
Serbia	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	18	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	35	89.7	0	0.0	1	2.6	0	0.0	3	7.7	0	0.0
	12-17	0	0.0	37	86.1	0	0.0	0	0.0	0	0.0	6	14.0	0	0.0
	18+	0	0.0	47	87.0	0	0.0	0	0.0	0	0.0	7	13.0	0	0.0
Slovak Rep.	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	4	26.7	0	0.0	11	73.3	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	24	57.1	0	0.0	18	42.9	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	3	9.7	0	0.0	4	12.9	0	0.0	24	77.4	0	0.0
	18+	0	0.0	31	27.4	0	0.0	13	11.5	0	0.0	69	61.1	0	0.0
Slovenia	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	2	66.7	0	0.0	1	33.3	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	5	20.0	0	0.0	0	0.0	0	0.0	20	80.0	0	0.0
	12-17	0	0.0	4	22.2	0	0.0	0	0.0	0	0.0	14	77.8	0	0.0
	18+	1	2.6	5	12.8	0	0.0	0	0.0	0	0.0	33	84.6	0	0.0
Spain	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	13	25.5	1	2.0	37	72.5	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	94	31.4	0	0.0	33	11.0	2	0.7	170	56.9	0	0.0
	12-17	0	0.0	25	7.8	0	0.0	0	0.0	2	0.6	293	91.3	1	0.3
	18+	0	0.0	95	10.9	2	0.2	0	0.0	15	1.7	756	86.4	7	0.8
Sweden	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	31	100	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	26	35.1	0	0.0	35	47.3	0	0.0	13	17.6	0	0.0
	12-17	0	0.0	29	31.2	1	1.1	23	24.7	0	0.0	38	40.9	2	2.2
	18+	0	0.0	84	25.2	3	0.9	59	17.7	3	0.9	154	46.1	31	9.3

Country		Missing/ Unknown		No		Ivacaftor		Lumacaftor/ Ivacaftor		Tezacaftor/ Ivacaftor		Elexacaftor/ Tezacaftor/ Ivacaftor		Other CFTR modulator	
		N	%	N	%	N	%	N	%	N	%	N	%	N	%
Switzerland	0-1	0	0.0	1	50.0	0	0.0	0	0.0	0	0.0	1	50.0	0	0.0
	2-5	0	0.0	25	71.3	0	0.0	10	28.6	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	35	26.2	1	0.8	3	2.3	0	0.0	91	70.0	0	0.0
	12-17	0	0.0	10	7.1	3	2.3	2	1.6	1	0.8	112	87.5	0	0.0
	18+	1	0.2	47	10.2	4	0.9	1	0.2	3	0.6	405	87.9	0	0.0
Turkey	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	58	89.2	0	0.0	7	10.8	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	182	79.8	1	0.4	2	0.9	1	0.4	42	18.4	0	0.0
	12-17	1	0.6	117	71.8	0	0.0	0	0.0	1	0.6	44	27.0	0	0.0
	18+	2	1.6	57	46.7	0	0.0	0	0.0	0	0.0	63	51.6	0	0.0
Ukraine	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	20	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	99	99.0	0	0.0	0	0.0	0	0.0	1	1.0	0	0.0
	12-17	1	1.4	69	98.6	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	60	96.8	0	0.0	0	0.0	1	1.6	1	1.6	0	0.0
United Kingdom	0-1	0	0.0	6	46.2	7	53.8	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	107	21.1	53	10.5	347	68.4	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	214	14.7	52	3.6	96	6.2	27	1.9	1062	73.2	0	0.0
	12-17	0	0.0	91	6.8	31	2.3	18	1.4	30	2.2	1172	87.3	0	0.0
	18+	0	0.0	411	8.0	123	2.4	4	0.1	121	2.4	4476	87.2	0	0.0
Total	0-1	2	6.3	14	43.8	15	46.9	0	0.0	0	0.0	1	3.1	0	0.0
	2-5	29	1.5	616	30.8	92	4.6	1255	62.8	0	0.0	7	0.4	0	0.0
	6-11	241	3.7	1933	29.6	110	1.7	895	13.7	215	3.3	3125	47.9	2	0.0
	12-17	132	2.1	975	15.3	88	1.4	282	4.4	82	1.3	4809	75.4	8	0.1
	18+	477	2.3	2320	11.3	332	1.6	307	1.5	424	2.1	16517	80.8	77	0.4

9. CFTR modulator therapies

Table 9.3   People with CF without F508del, eligible for at least one modulator, by country, by age and by last CFTR modulator prescribed. People with CF seen in 2022 who have never had a transplant.

Country		Missing/ Unknown		No		Ivacaftor		Lumacaftor/ Ivacaftor		Tezacaftor/ Ivacaftor		Elexacaftor/ Tezacaftor/ Ivacaftor	
		N	%	N	%	N	%	N	%	N	%	N	%
Albania	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	1	100.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Armenia	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Austria	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	1	25.0	3	75.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	1	50.0	1	50.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	1	25.0	3	75.0	0	0.0	0	0.0	0	0.0
Belarus	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Bulgaria	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	1	100.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	1	100.0	0	0.0	0	0.0	0	0.0	0	0.0
Croatia	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Cyprus	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	0	0.0	1	100.0	0	0.0	0	0.0	0	0.0
Czech Rep.	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	1	100.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	2	100.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	0	0.0	1	100.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	0	0.0	9	100.0	0	0.0	0	0.0	0	0.0
Denmark	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	1	50.0	0	0.0	0	0.0	0	0.0	1	50.0
Finland	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
France	0-1	1	100.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	1	20.0	0	0.0	4	80.0	0	0.0	0	0.0	0	0.0
	6-11	3	37.5	0	0.0	5	62.5	0	0.0	0	0.0	0	0.0
	12-17	9	47.4	0	0.0	9	47.4	0	0.0	0	0.0	1	5.3
	18+	18	29.5	0	0.0	37	60.7	0	0.0	0	0.0	6	9.8

Country		Missing/ Unknown		No		Ivacaftor		Lumacaftor/ Ivacaftor		Tezacaftor/ Ivacaftor		Elexacaftor/ Tezacaftor/ Ivacaftor	
		N	%	N	%	N	%	N	%	N	%	N	%
Georgia	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Germany	0-1	0	0.0	0	0.0	2	100.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	2	40.0	3	60.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	2	18.2	9	81.8	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	5	33.3	10	66.7	0	0.0	0	0.0	0	0.0
	18+	0	0.0	16	27.1	37	62.7	0	0.0	0	0.0	6	10.2
Greece	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	1	100.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	1	50.0	1	50.0	0	0.0	0	0.0	0	0.0
Hungary	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	1	100.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Iceland	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	0	0.0	1	100.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Ireland	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	2	22.2	7	77.8	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	1	12.5	7	87.5	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	0	0.0	6	100.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	3	8.1	34	91.9	0	0.0	0	0.0	0	0.0
Israel	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	0	0.0	5	62.5	0	0.0	0	0.0	3	37.5
Italy	0-1	0	0.0	2	100.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	8	66.7	4	33.3	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	11	55.0	9	45.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	3	18.8	12	75.0	0	0.0	0	0.0	1	6.3
	18+	0	0.0	26	36.1	45	62.5	0	0.0	0	0.0	1	1.4
Latvia	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Lithuania	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Luxemburg	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Moldova	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	1	100.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0

Country		Missing/ Unknown		No		Ivacaftor		Lumacaftor/ Ivacaftor		Tezacaftor/ Ivacaftor		Elexacaftor/ Tezacaftor/ Ivacaftor	
		N	%	N	%	N	%	N	%	N	%	N	%
Netherlands	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	1	100.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	2	66.7	1	33.3	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	2	66.7	1	33.3	0	0.0	0	0.0	0	0.0
	18+	0	0.0	3	33.3	6	66.7	0	0.0	0	0.0	0	0.0
N. Macedonia	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Norway	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	2	28.6	5	71.4	0	0.0	0	0.0	0	0.0
Poland	0-1	0	0.0	1	100.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	1	100.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	2	66.7	1	33.3	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	0	0.0	2	100.0	0	0.0	0	0.0	0	0.0
Portugal	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	1	100.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	0	0.0	1	50.0	0	0.0	0	0.0	1	50.0
Romania	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	1	100.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Russian Fed.	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Serbia	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Slovak Rep.	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	2	100.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	5	100.0	0	0.0	0	0.0	0	0.0	0	0.0
Slovenia	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Spain	0-1	0	0.0	0	0.0	1	100.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	2	100.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	1	50.0	1	50.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	1	33.3	2	66.7	0	0.0	0	0.0	0	0.0
	18+	0	0.0	8	47.1	9	52.9	0	0.0	0	0.0	0	0.0

Country		Missing/Unknown		No		Ivacaftor		Lumacaftor/Ivacaftor		Tezacaftor/Ivacaftor		Elexacaftor/Tezacaftor/Ivacaftor	
		N	%	N	%	N	%	N	%	N	%	N	%
Sweden	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	1	100.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	0	0.0	2	100.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	4	50.0	4	50.0	0	0.0	0	0.0	0	0.0
Switzerland	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	0	0.0	1	100.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	3	50.0	2	33.3	0	0.0	0	0.0	1	16.7
Turkey	0-1	0	0.0	2	100.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	9	90.0	1	10.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	8	80.0	1	10.0	0	0.0	0	0.0	1	10.0
	12-17	0	0.0	3	60.0	2	40.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	3	50.0	2	33.3	0	0.0	0	0.0	1	16.7
Ukraine	0-1	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	12-17	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
	18+	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
United Kingdom	0-1	0	0.0	3	42.9	4	57.1	0	0.0	0	0.0	0	0.0
	2-5	0	0.0	5	29.4	12	70.6	0	0.0	0	0.0	0	0.0
	6-11	0	0.0	7	20.0	20	57.1	0	0.0	1	2.9	7	20.0
	12-17	0	0.0	6	15.0	25	62.5	0	0.0	0	0.0	9	22.5
	18+	0	0.0	49	26.5	70	37.8	1	0.5	0	0.0	65	35.1
Total	0-1	1	6.3	8	50.0	7	43.8	0	0.0	0	0.0	0	0.0
	2-5	1	1.5	28	41.2	39	57.4	0	0.0	0	0.0	0	0.0
	6-11	4	3.7	35	32.1	61	56.0	0	0.0	1	0.9	8	7.3
	12-17	9	7.8	24	20.7	72	62.1	0	0.0	0	0.0	11	9.5
	18+	18	3.6	126	25.1	273	54.3	1	0.2	0	0.0	85	16.9

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 2022 and who have never had a transplant.
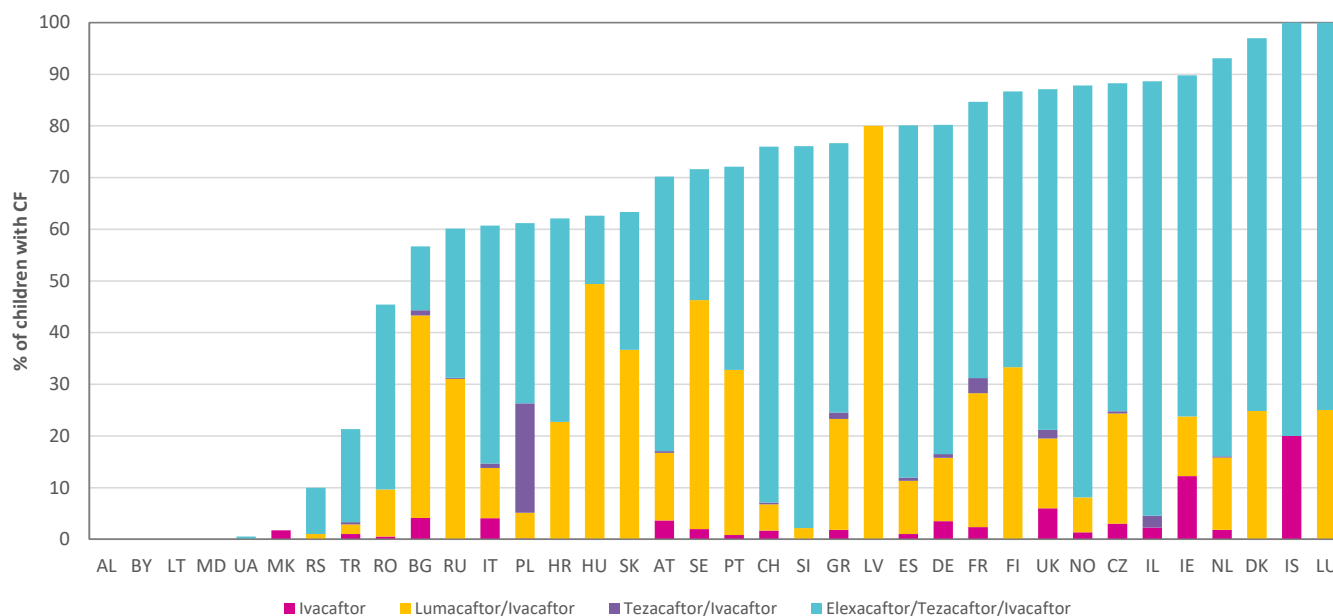
Note: Armenia, Cyprus and Georgia have <5 eligible children seen in 2022 and are excluded from the graph.

Figure 9.6   In the majority of 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 2022 and who have never had a transplant.
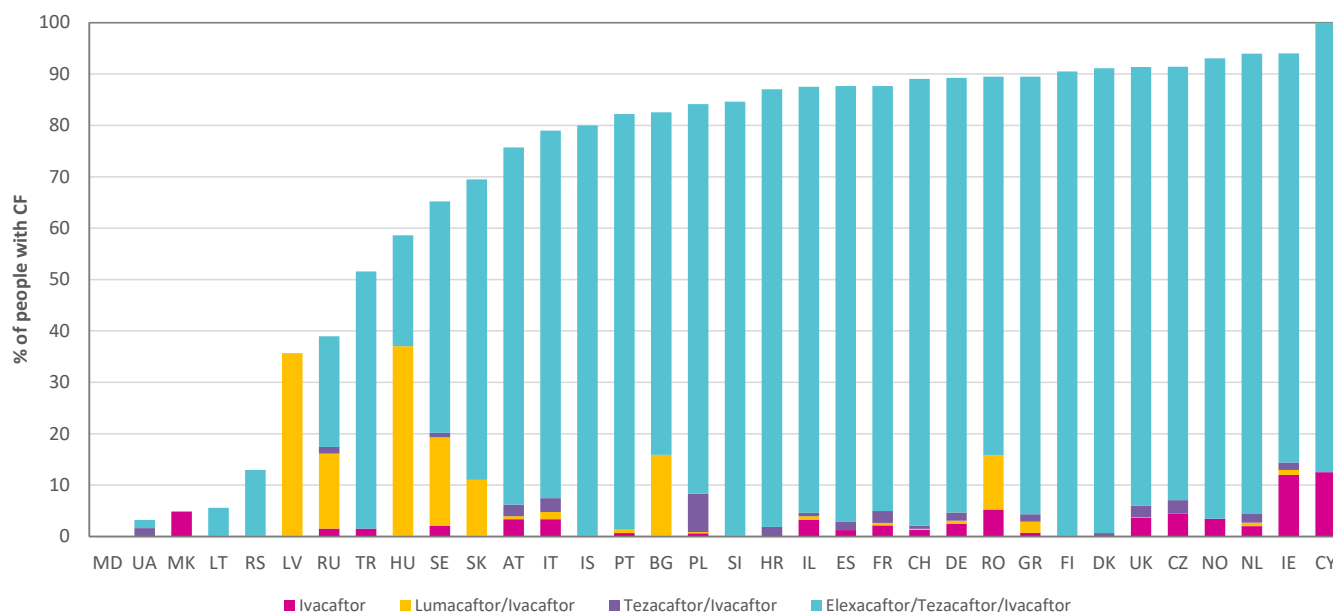
Note: Albania, Armenia, Belarus, Georgia and Luxembourg have <5 eligible adults seen in 2022 and are excluded from the graph.

9. CFTR modulator therapies

Figure 9.7   *A widening of the eligibility criteria and increased reimbursement for CFTR modulators in Europe means a considerable increase in their use from 2020 onwards.*

Use of CFTR modulator therapy from 2018 to 2022.
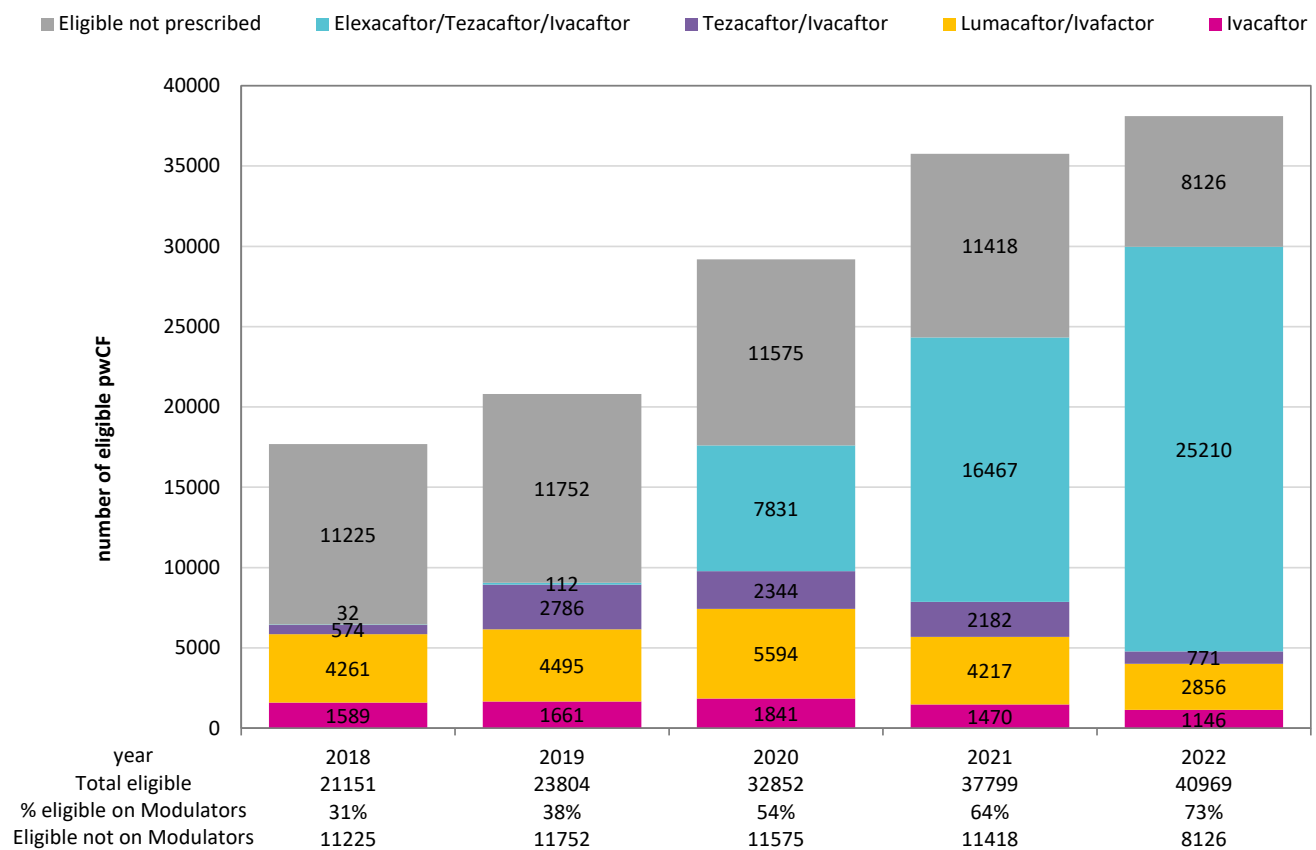
year	2018	2019	2020	2021	2022
Total eligible	21151	23804	32852	37799	40969
% eligible on Modulators	31%	38%	54%	64%	73%
Eligible not on Modulators	11225	11752	11575	11418	8126

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. Exclusion criteria were people who were lost to follow-up and/or transplanted (lung and/or liver), and people with missing values.

10. Transplantation

Despite the advent of highly effective CFTR modulators lung transplant is still a realistic scenario for some people with CF. The availability of lung transplantation differs amongst the countries participating in the ECFSPR and 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 transplantation 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 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.

10. Transplantation

Table 10.1   Number of people with CF alive in 2022 with transplanted lung(s), by age and sex.

Age	Males	Females	Total	Transplants carried out in 2022
0-5	0	0	0	0
6-11	1	2	3	0
12-17	10	18	28	5
18-29	186	279	465	18
30-39	469	448	917	17
40-49	396	381	777	12
50-59	168	130	298	0
60+	32	27	59	0
Total	1262	1285	2547	52

This table shows the number of people with CF alive in 2022 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 transplanted during 2022.

Figure 10.1   The number and proportion of people with CF living with a lung transplant is extremely heterogenous across Europe.

Number of people with CF living in 2022 with transplanted lungs, by country.
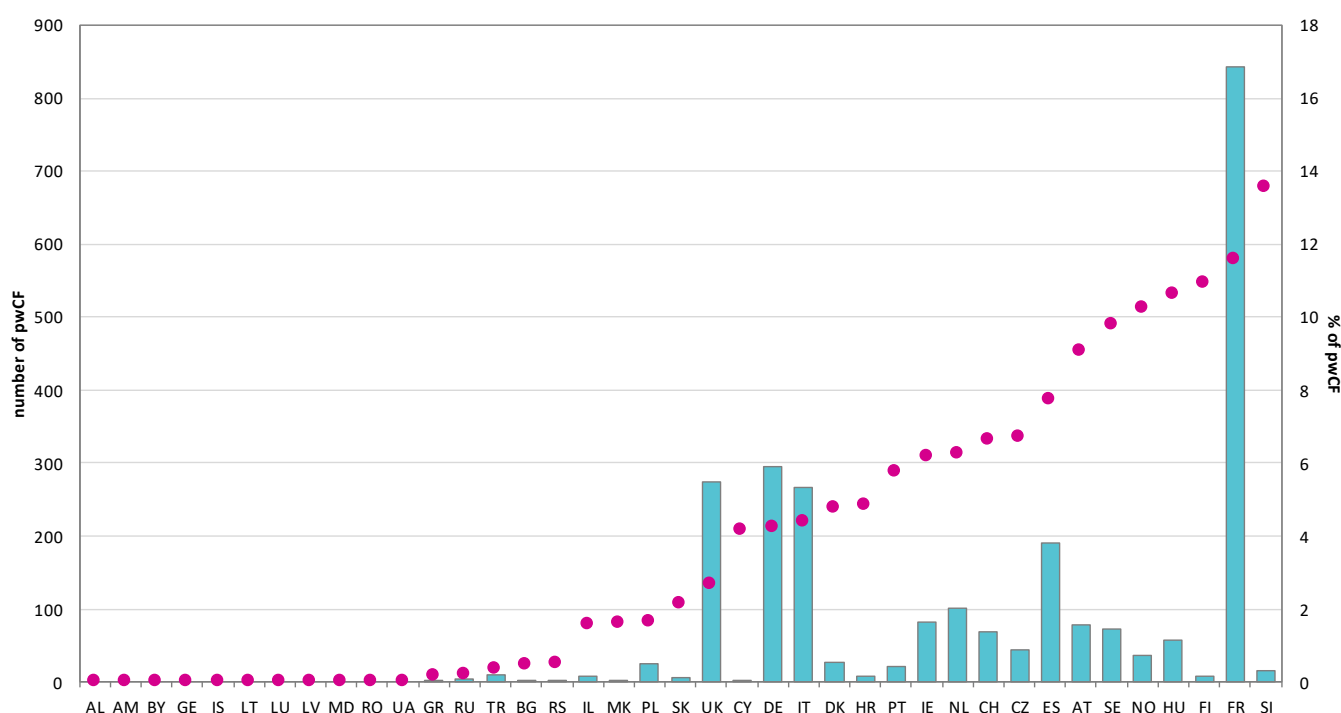
This graph shows the number of people with CF alive at 31/12/2022 who have had a lung transplant (light turquoise bars) at some point in their life. The pink dots (right axis) show the percentage of people that are living with transplanted lung in 2022 out of all people with CF that were seen in 2022.

10. Transplantation

Table 10.2   Number of people with CF living in 2022 with transplanted liver, by age and sex.

Age	Males	Females	Total	Transplants carried out in 2022
0-5	2	0	2	0
6-11	6	0	6	2
12-17	22	13	35	9
18-29	88	39	127	2
30-39	64	32	96	2
40-49	31	14	45	0
50-59	10	5	15	1
60+	1	0	1	0
Total	224	103	327	16

This table shows the number of people with CF alive in 2022 who have had a liver transplant at some time in their life, by age group, as well as the number of people transplanted during 2022.

Figure 10.2   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 2022 with transplanted liver, by country.
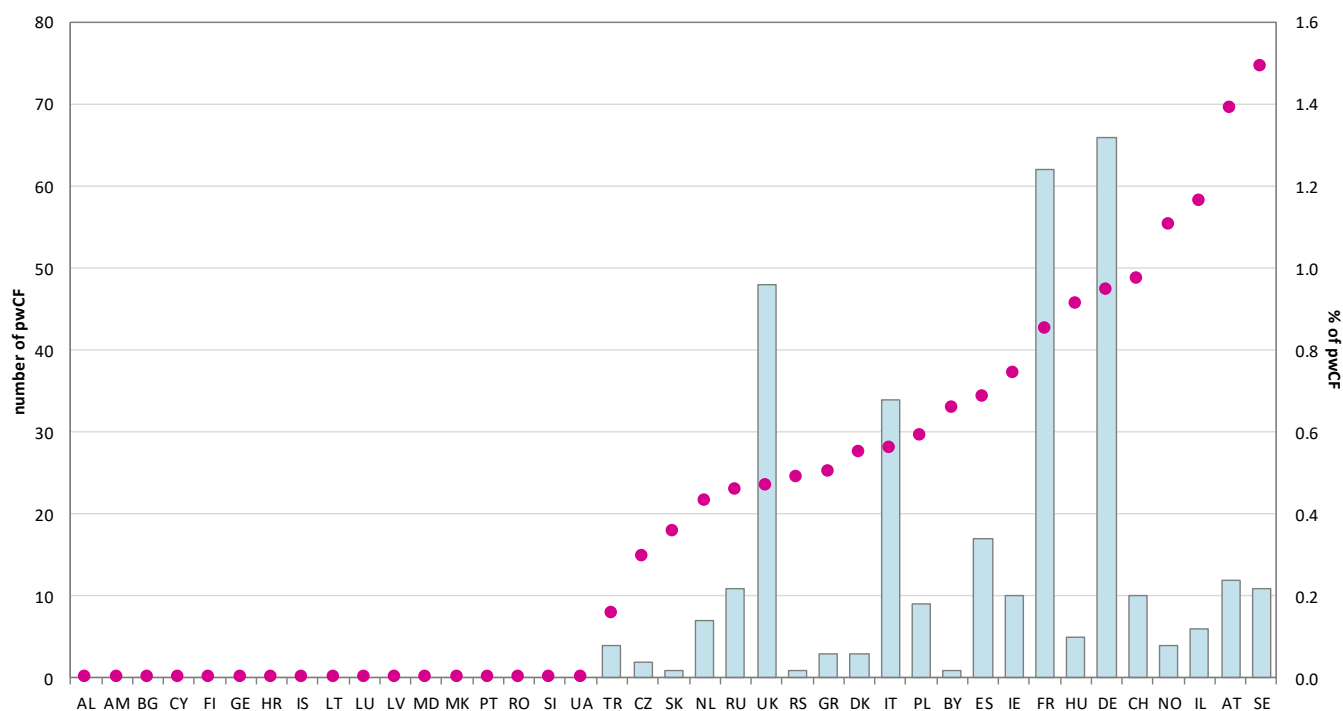
This graph shows the number of people with CF alive at 31/12/2022 who have had a liver transplant (light turquoise bars) at some point in their life. The pink dots (right axis) show the percentage of people that are living with transplanted liver in 2022 out of all people with CF that were seen in 2022. Note that (left vertical axis) the number of people who had a liver transplant is much lower than the number of lung transplanted people. 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.

10. Transplantation

Table 10.3   Number of people with CF living in 2022 with transplanted kidney(s), by age and sex.

Age	Males	Females	Total	Transplants carried out in 2022
0-5	0	0	0	0
6-11	1	0	1	0
12-17	1	0	1	0
18-29	10	8	18	7
30-39	20	32	52	5
40-49	43	33	76	8
50-59	17	14	31	1
60+	4	2	6	0
Total	96	89	185	21

Note: Hungary does not collect information on kidney transplant.

This table shows the number of people with CF alive in 2022 who have had a kidney transplant at some time in their life, by age group, as well as the number of people transplanted during 2022.

Table 10.4   Number of people with CF living in 2022 with other transplanted organs (not lung, liver, kidney), by age and sex.

Age	Males	Females	Total	Transplants carried out in 2022
0-5	0	1	1	0
6-11	0	0	0	0
12-17	2	2	4	1
18-29	7	6	13	0
30-39	6	10	16	0
40-49	9	11	20	0
50-59	5	4	9	0
60+	0	0	0	0
Total	29	34	63	1

This table shows the number of people with CF alive in 2022 who have had an organ transplant that is not lung, liver or kidney at some time in their life, by age group, as well as the number of people transplanted during 2022.

11. 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 per age group, divided by females and males. Respiratory disease remains the predominant cause of death.

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.

11. Mortality

Table 11.1   Number of deaths in 2022, 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	4	2.5	5	3.3	9	2.9
6-11	7	4.4	5	3.3	12	3.9
12-17	6	3.8	9	6.0	15	4.8
18-29	39	24.5	52	34.7	91	29.5
30-39	38	23.9	37	24.7	75	24.3
40-49	32	20.1	23	15.3	55	17.8
50-59	23	14.5	12	8.0	35	11.3
60+	10	6.3	7	4.7	17	5.5
Total	159		150		309	

Note: For the United Kingdom, all people with a confirmed diagnosis of CF were included (N=11,148). The total number of the CF population presented is 51,919.

This table shows the number of deaths in 2022 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 centre during the year, and therefore the information may not have been recorded.

Figure 11.1   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 2022, by sex.
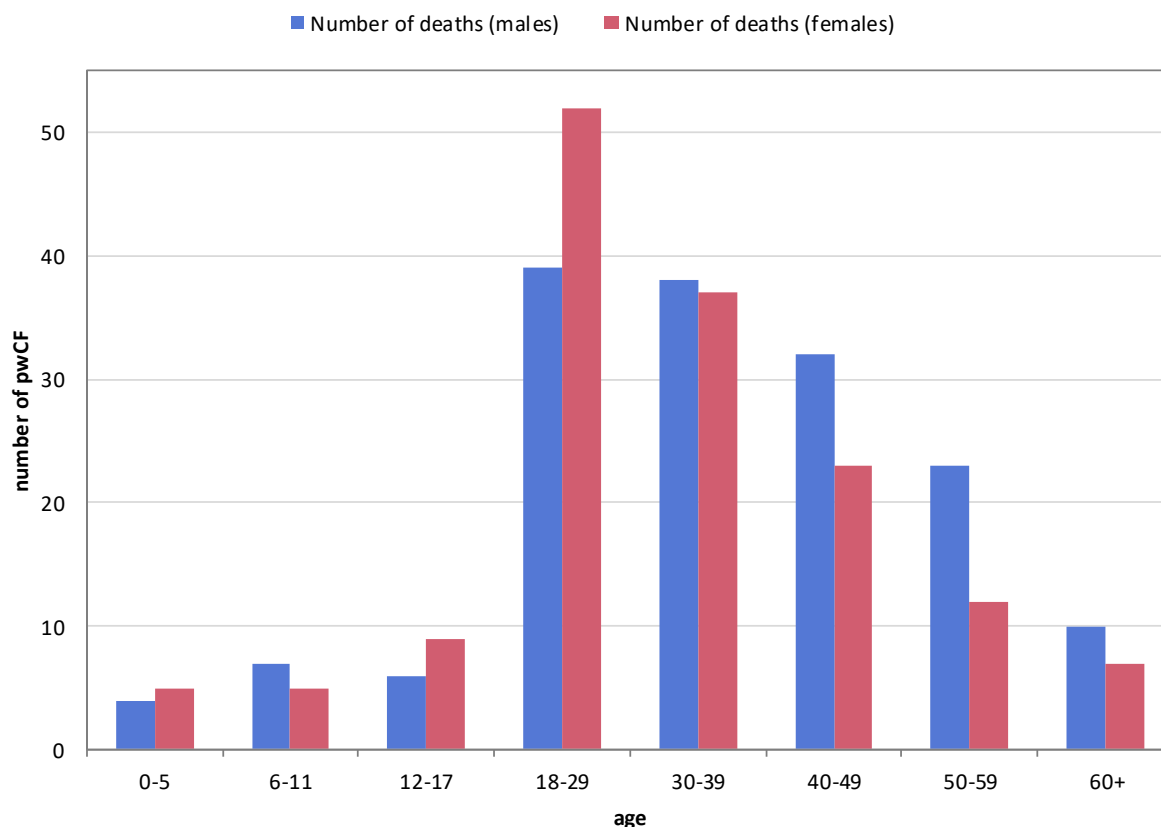
This graph shows the distribution of age at death of people with CF who died in 2022, separated by males (blue) and females (red).

11. Mortality

Table 11.2   Cause of death distribution of deaths in 2022.

Cause of death in 2022	Number of deaths	Percentage of all deaths
Respiratory	149	48.22
Transplantation	51	16.50
Non-CF related	30	9.71
Unknown	23	7.44
Other CF related	21	6.80
Cancer	19	6.15
Liver-GI	12	3.88
Suicide	4	1.29
Total	309	

Note: For the United Kingdom, all people with a confirmed diagnosis of CF were included (N=11,148). The total number of the CF population presented is 51,919.

Note: 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 2022. The most frequent cause of death was respiratory disease.

12. Data Quality

Data that will be used in vital research and pharmacovigilance studies and to inform public health planning, and as an instrument to monitor and review a range of patient outcomes needs to be of demonstrably high quality.

Data quality measures deployed by the ECFSPR

Several measures are in place in the ECFSPR that demonstrate our ongoing commitment to quality and our support for the contributing centres and national registries, namely:
- clear guidance documentation including variable descriptions, parameters, options and references;
- training and ongoing expert assistance for participants (live & recorded webinars, ad-hoc sessions, ticketing system, constant monitoring and communication before deadlines);
- a secure, custom-built data collection platform that highlights errors and inconsistencies during data input / upload;
- additional rigorous checks applied by the statisticians and final data checks and pre-publication approval by the Registry Director and the country representatives.

To enhance and complement these efforts we launched a new initiative in 2018, whereby members of the ECFSPR staff visit participant centres to verify and validate data at source.

Aims of the on-site data validation visits

Our objectives are to quantify how complete the data is, the consistency and accuracy of data-input and to verify that the centre has obtained the informed consent of individuals with CF to include his/her data in the Registry in accordance with current local and European legislation. The visits are also an invaluable opportunity for the Registry and its participants to collaborate on improving further the quality, relevance, and reliability of the ECFSPR data.

Consistency: adherence by the centre to the variable definitions, options, and parameters used by the Registry.
Accuracy of data-input: the proportion of values recorded in the ECFSPR software that match the medical records.

In a selected country, the aim is to visit at least 10% of the centres, and visits are limited to centres with ≥50 individuals with CF. In each centre, a subset of variables is checked for 15-20% of the total number of people with CF in a given follow-up year (this is always the most recent data approved by the ECFSPR).

For each visit, the ECFSPR statisticians generated a random list of people with CF registered in the centre, in 3 age categories, to be checked on-site. Of the selected population 50% were aged 18 or older, 40% were aged 6-17 years and 10% were younger than 6.

What data is checked?

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 or misrepresentation (based on ECFSPR experience and participant feedback); they are significant because they are used in reported Registry outcomes. They cover demographics, diagnosis, transplantation, anthropometric and lung function measurements, bacterial infections, selected medications and some complications.

Results from the on-site data validation visits

In 22 countries, clinical data from 2204 individuals from a total of 46,309 people with CF were validated for the follow-up year 2021 or 2022. Of these, the ECFSPR visited 16 during 2023 and early 2024: *Albania, Armenia, Bulgaria, Cyprus, Georgia, Iceland, Israel, Italy, Lithuania, Moldova, North Macedonia, Poland, Romania, Spain, Sweden* and Turkey. Seven countries with national CF registries – *Belgium, Czech Republic, France, Germany, Ireland, Netherlands* and *United Kingdom* – provided results from their own internal Data Validation Programmes. Overall, the validated clinical data amounts to 5% of the total number of people with CF in these countries. In this report we present a synopsis of the overall quality of the ECFPSR data based on completeness and accuracy. The overall results are shown and areas where improvement has been demonstrated since the data quality programme was introduced can be clearly seen.

12. Data Quality

Figure 12.1　Data completeness in the follow-up year 2022 for all non-transplanted people with CF seen in all participating countries, as overall percentages by variable.
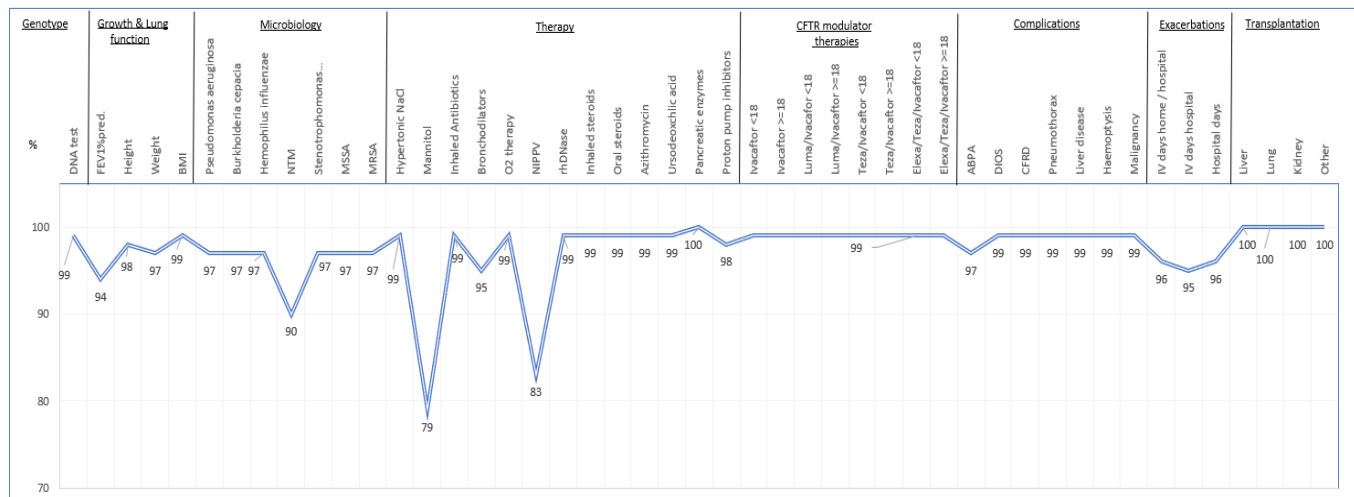
Note:　Completeness for FEV1 is evaluated only for people of at least 6 years old.

Note:　Completeness for BMI is evaluated for people of at least 2 years old.

The overall completeness of data, 97%, for all variables from all countries participating in the Registry in 2022 is high. For the variables Mannitol and NIPPV (non-invasive positive pressure ventilation) however, the completion rates are below 90%. This is a reflection of the fact that Mannitol and NIPPV are not preferred therapies for CF in some countries or are not reimbursed or unavailable in others.

Accuracy

The exactness of the values and the data consistency were determined by comparing the ECFSPR data and the data in the patient medical record at source; source data was checked only for a selection of variables.

Figure 12.2　Data accuracy for the follow-up years 2021 and 2022 from countries visited, overall results by variable.
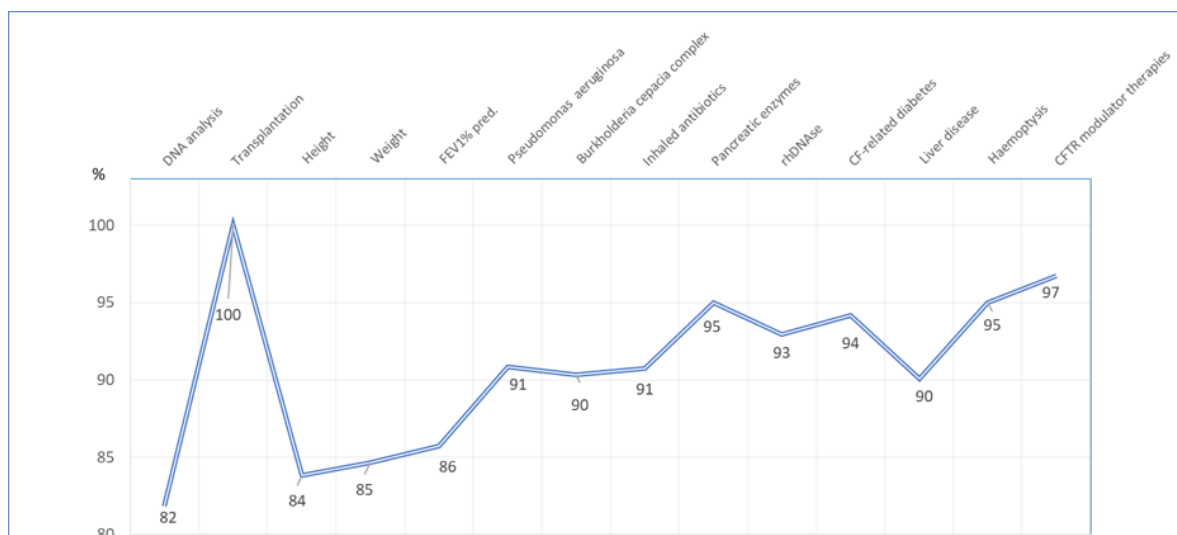
The accuracy results of the validated data from onsite visits for the follow-up years 2021 and 2022 are presented in Table 12.1 as percentages.

Table 12.1   Quartiles of validated data accuracies for people with CF, from the follow-up years 2021 and 2022 for the countries visited.

Variable	DNA analysis	Transplantation	Height	Weight	FEV1% pred.	Pseudomonas aeruginosa	Burkholderia cepacia complex	Inhaled antibiotics	Pancreatic enzymes	rhDNAse	CF-related diabetes	Liver disease	Haemoptysis	CFTR modulator therapies
25th percentile	74	100	83	83	84	88	92	87	93	88	93	87	95	98
50th percentile	84	100	89	88	92	95	98	91	98	96	97	91	97	100
75th percentile	96	100	94	95	95	96	99	98	99	98	100	96	100	100

Data accuracy is directly influenced by the availability of the original patient medical records at the centres and by local interpretation and understanding of the ECFSPR variable definitions.

Overall, for most variables, accuracy of the data validated in 2021-2022 averages 91%. Variables such as genotype, height, weight and lung function are more challenging for data providers, but the overall anomalies were minimal (less than 7%). The accuracy of genetic data cannot be verified without the original genetic report and the percentages of DNA-testing data validated as accurate varied across centres; this can be because genotyping reports are not forwarded when an individual moves for reasons such as transplantation and follow-up in another centre, transfer to another centre or country or to adult care and shared care. Genetic retesting is currently being carried out in many centres however genotype must be known before a CFTR modulator can be prescribed.

To minimise errors and improve adherence to ECFSPR definitions for height, weight and lung function, the centres have agreed to review and revise where necessary their procedures and the organisation of documentation across hospital departments.

Conclusion

The overall results present a positive picture of the completeness and accuracy of the data submitted to the ECFSPR. The areas for improvement that have been identified are already helping us to focus efforts on where they have the highest impact. We are confident that the measures deployed in partnership with the participating countries certify our collective efforts to further optimise the quality of data in the ECFSPR.

National registries continue to apply data quality measures in their own countries that mirror the ECFSPR system and to share the results with us.

Publications

The ECFSPR database is a useful source for research and the data is actively used. Applications for data are conscientiously handled in accordance with the ECFSPR guidelines. You will find more information on the data application process on the website here: Data Request Application.

From 2011 to March 2024 (time of writing) we had received a total of 124 applications to use Registry data. The majority of these requests, 81%, originated from researchers from the European Cystic Fibrosis Society and other institutes, and 19% 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

- Kim C, Higgins M, Liu L, Volkova N, Zolin A, Naehrlich L, on behalf of the ECFS Patient Registry Study Group. Effectiveness of lumacaftor/ivacaftor initiation in children with CF aged 2 through 5 years on disease progression: interim results from an ongoing registry-based study. J Cyst Fibros 2024;1569-1993. doi: 10.1016/j.jcf.2024.02.004

- Kerem E , Orenti A , Adamoli A, Hatziagorou E , Naehrlich L, Sermet-Gaudelus I, on behalf of the ECFS Patient Registry Steering Group. Cystic fibrosis in Europe: improved lung function & longevity: reasons for cautious optimism, but challenges remain. Eur Respir J. 2004;63(3): 2301241. doi: 10.1183/13993003.01241-2023

- Bradbury JD, Chesshyre E, Orenti A, Jung A, Warris A, European Cystic Fibrosis COVID project group. A multinational report on SARS-COV-2 infection outcomes in people with CF and Aspergillus infection or ABPA. J Cyst Fibros. 2023; S1569-1993(23)01666-1. doi: 10.1016/j.jcf.2023.10.017.

- Orenti A, Pranke I, Faucon C, et al, on behalf of the ECFS Patient Registry Steering Group. Nonsense mutations accelerate lung disease and decrease survival of cystic fibrosis children. J Cyst Fibros. 2023;22(6):1070-1079. doi: 10.1016/j.jcf.2023.06.005.

Abstracts

- Gambazza S, Orenti A, Zolin A, Ambrogi F. A multi-state model evaluating the association of oxygen therapy with the course of Cystic Fibrosis in Europe. 44th Annual Conference of the International Society for Clinical Biostatistics, Milan, Italy, 28-31 August 2023. Poster.

- Orenti, A, Adamoli A. Kerem E, Hatziagorou E, Zolin, A, Ambrogi F. 10-years changes in lung function of cystic fibrosis patients in Europe: different statistical methods at work. 44th Annual Conference of the International Society for Clinical Biostatistics, Milan, Italy, 28-31 August 2023. Poster.

- Adamoli A, Orenti A, Kerem E, Hatziagorou E, Zolin A, Ambrogi F. 10-years changes in lung function of cystic fibrosis patients in Europe: different statistical methods at work. XII Congresso Nazionale SISMEC, Jesi (AN), Italy, 20-23 September 2023. Oral presentation.

- Orenti A, Zolin A, Naehrlich L, Sermet-Gaudelus I. Evolution of disease in people with cystic fibrosis carrying 2 nonsense mutations in Europe. XII Congresso Nazionale SISMEC, Jesi (AN), Italy, 20-23 September 2023. Oral presentation.

- Orenti A, Hatziagorou E, Kerem E, De Boeck C, Burgel PR. Evolution of cystic fibrosis adult population over the last 10-years in Europe. Adult Cystic Fibrosis Conference. Milan, Italy, 1-2 December 2023. Oral presentation.

- Tomarelli I, De Petro C, Orenti A, Annicchiarico L, Misuraca S, Contarini M, Gramegna A, Blasi F. Patients not eligible for CFTR-modulators (CFTRm) in Europe: characteristics and distribution. Adult Cystic Fibrosis Conference. Milan, Italy, 1-2 December 2023. Oral presentation.

- V. Prasad, A. Fox, M. Krasynk, A. Zolin, J. van Rens, L. Naehrlich. Assessment of the quality of data in the European Cystic Fibrosis Society Patient Registry. Journal of Cystic Fibrosis, Vol. 22, Supplement, S106, P137, Published in issue: June 2023.

- M. Mei-Zahav, A. Orenti, A. Jung, J. van Rens, E. Kerem. Differences in disease severity among different residual function mutations: data from the ECFS Patient Registry. Journal of Cystic Fibrosis, Vol. 22, Supplement, S5–S6, WS03.01, Published in issue: June 2023.

- A.C.S. Wathne, T.L. Katzenstein, C. Krantz, A. Lindblad, I. de Monestrol, H.V. Olesen, and others. Scandinavian cystic fibrosis registry study - comparing care and clinical outcome. Journal of Cystic Fibrosis, Vol. 22, Supplement, S107, P139, Published in issue: June 2023.

A complete overview of publications is available on our website.

Sponsors

The ECFSPR is grateful to the patient organisations for the support of our work by means of an unrestricted grant:
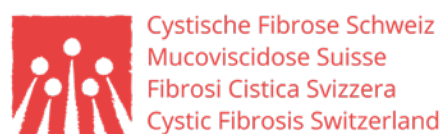
Appendix 1 Tables

1. Demographics

Table 1.1   Proportion of children (<18 years) and adults (≥18 years), by country and overall. People with CF alive on 31/12/2022.

Country	Children (<18 years)		Adults (≥18 years)	
	Number	%	Number	%
Albania	92	90.20	10	9.80
Armenia	23	88.46	3	11.54
Austria	377	42.70	506	57.30
Belarus	147	96.71	5	3.29
Bulgaria	129	58.90	90	41.10
Croatia	87	58.39	62	41.61
Cyprus	10	29.41	24	70.59
Czech Republic	329	47.41	365	52.59
Denmark	213	37.43	356	62.57
Finland	26	40.00	39	60.00
France	2703	37.37	4531	62.63
Georgia	77	90.59	8	9.41
Germany	2838	39.27	4389	60.73
Greece	238	36.84	408	63.16
Hungary	271	49.72	274	50.28
Iceland	9	60.00	6	40.00
Ireland	540	39.59	824	60.41
Israel	166	29.70	393	70.30
Italy	2219	36.59	3845	63.41
Latvia	32	68.09	15	31.91
Lithuania	21	47.73	23	52.27
Luxembourg	21	80.77	5	19.23
Rep of Moldova	41	75.93	13	24.07
The Netherlands	541	33.37	1080	66.63
North Macedonia	85	59.86	57	40.14
Norway	128	35.16	236	64.84
Poland	995	61.80	615	38.20
Portugal	172	44.10	218	55.90
Romania	282	90.68	29	9.32
Russian Federation	2821	71.78	1109	28.22
Serbia	144	65.16	77	34.84
Slovak Republic	129	42.02	178	57.98
Slovenia	57	47.50	63	52.50
Spain	1089	42.46	1476	57.54
Sweden	267	34.45	508	65.55
Switzerland	414	39.35	638	60.65
Turkey	2154	84.97	381	15.03
Ukraine	303	74.45	104	25.55
United Kingdom	4503	40.63	6581	59.37
Total	24693	45.50	29544	54.50

1. Demographics

Table 1.2   Age at follow-up: descriptive statistics, by country and overall. People with CF alive on 31/12/2022.

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	102	11.7	2.0	7.5	11.4	15.8	22.4
Armenia	26	11.3	3.0	7.4	10.8	15.0	25.0
Austria	883	22.6	0.2	11.5	21.4	32.4	68.5
Belarus	152	9.7	0.5	5.9	10.2	13.6	19.0
Bulgaria	219	17.6	0.2	7.7	15.4	25.3	68.5
Croatia	149	16.7	0.0	7.5	15.8	23.8	38.5
Cyprus	34	26.6	4.2	12.4	21.5	38.6	68.2
Czech Republic	694	20.1	0.2	9.4	18.6	28.4	67.0
Denmark	569	24.9	0.2	11.1	24.1	35.6	73.0
Finland	65	25.8	1.5	13.4	22.3	34.1	80.2
France	7234	25.0	0.0	13.2	23.3	35.3	87.6
Georgia	85	9.8	1.6	5.9	9.0	13.7	20.0
Germany	7227	24.2	0.0	11.5	23.0	34.5	85.7
Greece	646	23.8	0.6	14.0	23.1	31.7	75.8
Hungary	545	19.9	0.1	9.2	18.0	29.0	73.9
Iceland	15	17.7	0.5	7.5	14.3	25.7	42.0
Ireland	1364	23.9	0.1	12.2	22.7	34.7	73.7
Israel	559	27.3	0.5	16.5	26.1	36.5	77.5
Italy	6064	26.0	0.1	12.7	23.6	37.3	90.1
Latvia	47	14.2	1.6	5.9	13.2	21.5	36.0
Lithuania	44	18.7	0.8	9.9	18.9	25.6	38.5
Luxembourg	26	10.5	0.2	3.1	6.9	15.3	40.6
Rep of Moldova	54	13.3	1.2	7.3	12.1	17.5	36.7
The Netherlands	1621	26.9	0.0	14.2	25.5	36.5	74.2
North Macedonia	142	16.0	0.8	6.0	14.7	23.3	45.8
Norway	364	27.4	0.3	12.5	26.5	40.6	82.6
Poland	1610	16.4	0.1	8.1	14.4	22.9	62.5
Portugal	390	22.2	0.4	11.0	19.9	30.6	69.0
Romania	311	10.3	0.5	5.3	10.0	15.2	25.0
Russian Fed.	3930	14.4	0.2	7.2	12.4	19.1	65.0
Serbia	221	15.1	0.3	6.0	13.5	21.0	49.1
Slovak Republic	307	22.2	0.2	11.1	20.9	31.8	83.0
Slovenia	120	20.7	0.9	11.1	19.6	27.8	67.1
Spain	2565	24.2	0.2	11.4	21.2	35.4	87.2
Sweden	775	27.4	0.1	14.0	26.5	39.0	78.6
Switzerland	1052	24.4	0.3	11.9	23.0	33.5	82.0
Turkey	2535	10.8	0.1	5.0	9.2	15.4	49.0
Ukraine	407	13.3	0.3	7.0	12.0	18.1	42.7
United Kingdom	11084	24.2	0.0	11.5	22.5	34.4	88.4
Total	54237	22.6	0.0	10.4	20	32.5	90.1

2. Diagnosis

Table 2.1　Age at diagnosis (in years): descriptive statistics, by country and overall. All children and adolescents (<18 years) seen in 2022.

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	66	0	0.49	0	0.16	0.25	0.40	4.00
Armenia	21	1	1.78	0.10	0.33	0.75	2.00	6.17
Austria	366	9	0.49	0	0.10	0.10	0.20	14.7
Belarus	147	0	1.68	0.01	0.10	0.30	2.01	11.00
Bulgaria	122	3	2.28	0	0.30	0.70	2.90	17.30
Croatia	81	2	1.09	0	0.16	0.42	0.90	7.10
Cyprus	6	1	1.20	0.01	0.03	0.63	1.10	4.80
Czech Republic	325	0	0.55	0	0.10	0.10	0.20	12.30
Denmark	211	0	0.77	0	0	0	0.67	8.20
Finland	26	0	1.36	0	0.16	0.54	1.08	7.50
France	2664	39	0.31	0	0.10	0.10	0.10	17.20
Georgia	40	1	2.36	0	0.20	1.30	4.00	10.70
Germany	2734	47	1.14	0	0.08	0.24	1.08	16.85
Greece	210	28	1.09	0	0.17	0.49	1.02	12.20
Hungary	267	2	1.75	0	0.17	0.66	2.00	14.65
Iceland	9	0	0.39	0	0.10	0.20	0.58	1.20
Ireland	525	0	0.74	0	0.04	0.07	0.35	12.73
Israel	152	7	1.47	0	0	0.40	1.60	13.00
Italy	2179	38	0.74	0	0.07	0.13	0.35	16.76
Latvia	32	0	0.96	0	0.10	0.30	0.89	6.10
Lithuania	18	2	2.28	0	0.70	1.25	3.50	7.50
Luxembourg	21	0	0.29	0	0.10	0.10	0.40	1.30
Rep of Moldova	34	0	0.93	0.10	0.30	0.41	0.60	6.00
The Netherlands	530	8	0.78	0	0	0.10	0.30	14.17
North Macedonia	74	0	1.18	0	0.10	0.30	1.30	7.90
Norway	128	0	0.97	0	0.10	0.10	0.59	11.60
Poland	944	10	0.84	0	0.10	0.20	0.30	16.90
Portugal	172	0	1.32	0	0.10	0.20	1.10	13.60
Romania	250	9	1.54	0	0.20	0.50	1.40	14.90
Russian Fed.	1873	2	0.98	0	0.10	0.20	0.60	16.40
Serbia	136	0	1.73	0	0.20	0.50	2.25	15.80
Slovak Republic	109	17	0.80	0	0	0.10	0.40	11.00
Slovenia	56	0	1.19	0	0.10	0.45	1.73	5.60
Spain	1054	5	0.70	0	0.10	0.10	0.33	14.00
Sweden	263	3	1.59	0	0.16	0.51	1.97	13.87
Switzerland	396	12	0.65	0	0	0.10	0.30	11.20
Turkey	2122	21	1.26	0	0.17	0.33	0.70	17.00
Ukraine	259	6	2.18	0	0.30	0.70	3.00	14.80
United Kingdom	4142	0	0.42	0	0.04	0.06	0.09	17.12
Total	22764	273	0.83	0	0.07	0.10	0.42	17.30

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 2.2   Age at diagnosis (in years): descriptive statistics, by country and overall. All adults (≥18 years) seen in 2022.

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)
Austria	422	60	3.78	0	0.10	0.30	2.30	58.60
Belarus	5	0	3.84	0.05	1.17	2.00	4.00	12.00
Bulgaria	85	1	8.44	0.10	0.50	3.00	14.00	37.70
Croatia	61	0	3.11	0.08	0.20	0.50	3.00	31.00
Cyprus	16	1	11.90	0.11	0.60	7.72	16.67	66.10
Czech Republic	342	5	4.62	0	0.30	0.96	4.00	53.90
Denmark	331	2	3.38	0	0.17	0.58	2.92	42.67
Finland	27	11	4.61	0	0.30	2.00	4.00	30.00
France	4478	53	6.91	0	0.10	0.60	6.90	81.20
Germany	3998	174	5.47	0	0.17	0.98	4.83	72.67
Greece	307	52	4.55	0	0.24	0.56	4.54	54.88
Hungary	269	4	5.21	0.08	0.33	1.00	5.00	40.03
Iceland	6	0	0.28	0.10	0.25	0.28	0.40	0.40
Ireland	806	0	4.79	0	0.13	0.51	3.76	65.61
Israel	350	2	7.17	0	0.15	0.67	8.00	62.00
Italy	3756	85	10.27	0	0.17	1.09	15.78	77.62
Latvia	14	0	6.49	0.10	0.50	3.00	12.00	25.60
Lithuania	20	1	8.94	0	2.75	8.05	14.30	24.00
Luxembourg	5	0	9.01	0.33	0.70	3.00	17.00	24.00
Rep of Moldova	12	0	5.72	0.20	0.35	0.95	14.55	19.00
The Netherlands	973	98	6.67	0	0.20	1.10	6.00	68.00
North Macedonia	51	0	3.55	0	0.20	0.40	3.00	29.20
Norway	230	2	9.88	0	0.40	2.20	15.20	69.00
Poland	556	4	6.90	0	0.50	2.50	9.80	54.10
Portugal	207	1	12.19	0	0.90	7.30	19.40	58.00
Romania	24	0	2.53	0.10	0.35	0.80	3.85	12.00
Russian Fed.	499	0	8.11	0	1.00	4.30	12.90	59.50
Serbia	63	3	3.89	0.10	0.30	2.10	5.90	18.60
Slovak Republic	148	5	7.52	0	0.20	2.00	10.95	59.00
Slovenia	59	2	3.94	0	0.10	0.60	4.50	37.40
Spain	1379	25	9.73	0	0.30	1.50	13.00	75.00
Sweden	460	6	6.10	0	0.21	0.99	4.47	70.57
Switzerland	523	93	6.18	0	0.20	1.00	5.00	75.20
Turkey	375	0	7.43	0	0.33	3.00	13.00	43.60
Ukraine	78	0	6.82	0	1.00	3.50	9.10	38.50
United Kingdom	6076	0	6.92	0	0.08	0.66	5.05	81.35
Total	27017	690	7.12	0	0.17	0.93	7.42	81.35

Note: Albania, Armenia and Georgia have <5 adults seen in 2022 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 4.1   FEV₁% of predicted of children and adolescents: descriptive statistics, by country and overall. People with CF aged 6-17 years who have never had an organ 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	48	10	85.8	26.9	79.3	89.6	96.6	119.2
Armenia	16	1	87.2	52.6	79.9	91.4	95.8	110.2
Austria	268	1	99.7	41.5	90.5	101.0	109.4	134.6
Belarus	46	40	75.0	26.3	62.2	73.7	90.6	114.3
Bulgaria	81	6	83.2	26.3	70.9	87.0	98.3	147.8
Croatia	52	2	83.7	27.1	71.8	87.4	98.8	134.5
Cyprus	5	0	83.7	46.0	74.1	74.4	110.2	114.0
Czech Republic	197	5	101.5	35.9	93.0	101.1	111.7	141.9
Denmark	132	0	104.8	33.7	95.6	106.7	114.3	133.3
Finland	20	1	87.1	40.6	77.1	88.1	96.9	111.9
France	1919	65	97.5	24.5	88.4	99.1	108.0	161.0
Germany	1861	29	98.4	28.3	89.7	99.4	108.4	155.1
Greece	182	9	104.9	40.6	95.3	105.4	116.9	168.2
Hungary	160	15	80.8	20.0	71.9	83.1	93.7	118.5
Iceland	7	0	101.5	74.9	92.8	104.9	111.5	114.8
Ireland	366	28	97.4	30.5	89.2	99.0	107.0	161.1
Israel	137	3	91.8	18.3	84.1	93.4	103.6	132.9
Italy	1535	92	100.2	27.3	91.8	101.2	111.6	156.7
Latvia	18	1	89.7	44.8	82.3	93.9	99.0	110.1
Lithuania	10	2	78.5	43.3	47.6	76.9	103.2	123.5
Luxembourg	8	0	98.4	72.0	89.7	99.6	110.6	115.2
Rep of Moldova	19	5	79.9	23.4	73.9	79.0	97.3	113.8
The Netherlands	382	11	99.1	34.7	91.2	100.1	108.0	144.3
North Macedonia	43	1	83.1	52.9	74.0	82.2	92.9	116.7
Norway	83	0	98.6	52.1	89.9	99.5	107.3	122.0
Poland	614	55	92.5	21.2	82.3	96.3	106.3	137.8
Portugal	114	13	92.2	37.0	81.7	94.2	105.1	131.0
Romania	143	31	84.1	24.3	75.7	84.3	96.5	118.2
Russian Fed.	1046	319	86.1	17.8	72.4	88.5	102.0	167.5
Serbia	79	2	84.7	27.9	76.4	87.9	100.0	113.4
Slovak Republic	92	0	94.7	43.0	87.1	95.6	102.7	127.4
Slovenia	47	0	97.2	64.0	90.5	99.4	105.5	117.3
Spain	772	31	97.0	24.1	87.8	97.7	108.7	141.5
Sweden	202	5	92.5	29.9	82.3	93.3	104.3	130.3
Switzerland	299	3	99.9	59.8	91.1	100.2	108.7	139.2
Turkey	901	449	83.6	10.0	69.9	86.6	99.6	153.3
Ukraine	104	84	85.6	22.9	72.4	87.8	102.3	124.0
United Kingdom	2954	113	98.3	33.3	90.1	99.2	107.7	169.5
Total	14963	1434	95.5	10.0	86.2	97.6	107.3	169.5

Note: Georgia has <5 individuals aged 6-17 years with FEV1 measurement and is excluded from the table.

Note: Sweden and the United Kingdom report FEV₁ from the annual review, which might not be the best FEV₁ of the year, and, in some cases, the FEV₁ measurement could be from the previous calendar year.

This table shows some descriptive statistics for FEV₁ in children and young people, expressed as % of predicted. Note that people with CF who have had an organ transplant and children below 6 years of age have been excluded from the analyses.

4. Lung function

Table 4.2   FEV$_1$% of predicted of adults: descriptive statistics, by country. 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)
Austria	382	4	81.0	19.6	64.9	82.6	96.9	134.1
Bulgaria	80	2	66.6	10.2	44.7	73.3	89.1	109.4
Croatia	52	0	77.7	27.0	60.8	81.6	93.6	118.4
Cyprus	15	0	61.8	29.0	48.2	63.4	73.7	93.3
Czech Republic	269	9	78.3	23.5	60.6	84.6	95.5	127.2
Denmark	293	1	87.7	28.9	71.0	93.1	104.6	138.9
Finland	30	0	69.2	30.9	48.1	72.0	86.1	107.6
France	3536	45	78.6	10.6	60.6	80.2	96.5	152.7
Germany	3661	62	75.5	12.1	56.2	77.3	94.9	149.8
Greece	265	15	78.6	14.7	59.0	82.4	96.8	145.4
Hungary	183	3	62.7	15.3	40.2	62.9	83.2	120.0
Iceland	6	0	81.7	63.0	68.8	84.2	93.0	97.2
Ireland	660	42	76.2	14.1	59.7	79.9	93.5	158.1
Israel	326	1	74.9	25.4	59.9	77.6	90.9	128.9
Italy	3303	83	80.8	16.8	61.8	83.8	99.9	153.6
Latvia	14	0	67.9	24.0	39.5	70.8	98.4	100.8
Lithuania	22	0	57.8	12.2	30.3	58.1	82.0	111.3
Luxembourg	5	0	75.3	24.1	41.4	89.2	107.8	114.0
Rep of Moldova	7	3	50.5	32.8	36.2	52.0	69.3	71.2
The Netherlands	926	8	78.0	15.0	60.0	80.2	95.5	139.7
North Macedonia	45	0	63.7	22.3	46.6	65.1	80.5	104.7
Norway	187	1	77.4	15.7	63.2	82.5	93.7	129.8
Poland	473	25	69.1	13.6	49.4	70.1	89.3	138.7
Portugal	165	10	73.6	23.2	54.0	76.2	93.1	125.0
Romania	13	1	85.8	56.4	76.8	95.3	97.9	108.2
Russian Fed.	211	232	58.7	10.8	36.5	57.7	78.6	134.5
Serbia	60	0	59.0	20.3	38.9	59.0	73.9	114.9
Slovak Republic	136	3	74.4	19.6	58.1	78.6	90.9	125.6
Slovenia	44	0	73.1	23.1	51.7	75.5	92.4	120.1
Spain	1136	19	77.6	21.7	60.1	79.9	96.0	141.2
Sweden	368	3	75.1	17.8	60.9	76.5	90.2	126.3
Switzerland	521	1	77.0	23.7	57.9	77.7	95.3	136.7
Turkey	280	60	65.9	15.4	43.2	66.4	87.7	129.9
Ukraine	65	7	63.9	19.3	45.2	63.6	84.3	111.9
United Kingdom	5463	161	76.4	10.5	59.3	78.9	94.1	193.3
Total	23207	801	76.8	10.2	58.6	79.1	95.4	193.3

Note: Albania, Armenia, Belarus and Georgia have <5 adults with FEV$_1$ measurement and are excluded from the table, but the people are included in the total number.

Note: Sweden and the United Kingdom report FEV$_1$ from the annual review, which might not be the best FEV$_1$ of the year and in some cases, the FEV$_1$ measurement could be from the previous calendar year.

This table shows some descriptive statistics for FEV$_1$ in adults with CF, expressed as the % of predicted. Note that adults who have had a transplant have been excluded from the analyses.

5. Microbiology

Table 5.1   Prevalence of Pseudomonas aeruginosa in people with CF seen in 2022 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	37	55.2	12	17.9	18	26.9								
Armenia	1	4.6	15	68.2	4	18.2	2	9.1								
Austria	3	0.8	302	81.4	23	6.2	43	11.6	1	0.3	235	58.9	142	35.6	21	5.3
Belarus	0	0.0	111	76.0	35	24.0	0	0.0	0	0.0	4	80.0	1	20.0	0	0.0
Bulgaria	1	0.8	90	71.4	24	19.1	11	8.7	0	0.0	44	51.8	24	28.2	17	20.0
Croatia	1	1.2	53	64.6	12	14.6	16	19.5	0	0.0	28	50.9	20	36.4	7	12.7
Cyprus	0	0.0	5	71.4	2	28.6	0	0.0	1	6.3	7	43.8	5	31.3	3	18.8
Czech Rep.	21	6.5	277	85.5	8	2.5	18	5.6	37	12.3	173	57.3	71	23.5	21	7.0
Denmark	0	0.0	180	85.3	8	3.8	23	10.9	0	0.0	194	63.6	76	24.9	35	11.5
Finland	0	0.0	24	92.3	1	3.9	1	3.9	0	0.0	20	64.5	11	35.5	0	0.0
France	0	0.0	2192	81.5	135	5.0	364	13.5	0	0.0	2395	65.0	966	26.2	323	8.8
Georgia	3	7.3	25	61.0	8	19.5	5	12.2								
Germany	33	1.2	2303	83.6	225	8.2	195	7.1	126	3.3	1723	45.2	1820	47.7	147	3.9
Greece	4	1.7	175	73.8	15	6.3	43	18.1	18	5.1	107	30.2	215	60.7	14	4.0
Hungary	2	0.8	148	62.5	36	15.2	51	21.5	6	3.1	82	42.3	29	15.0	77	39.7
Iceland	0	0.0	8	88.9	1	11.1	0	0.0	0	0.0	5	83.3	0	0.0	1	16.7
Ireland	0	0.0	497	94.7	6	1.1	22	4.2	0	0.0	579	80.5	86	12.0	54	7.5
Israel	8	5.0	110	69.2	21	13.2	20	12.6	23	6.8	154	45.3	115	33.8	48	14.1
Italy	4	0.2	1687	76.5	138	6.3	375	17.0	8	0.2	1817	50.9	1278	35.8	470	13.2
Latvia	0	0.0	23	71.9	5	15.6	4	12.5	0	0.0	6	42.9	7	50.0	1	7.1
Lithuania	0	0.0	13	65.0	1	5.0	6	30.0	0	0.0	10	43.5	8	34.8	5	21.7
Luxembourg	0	0.0	19	90.5	2	9.5	0	0.0	0	0.0	2	40.0	3	60.0	0	0.0
Rep. Moldova	1	2.8	17	47.2	15	41.7	3	8.3	0	0.0	1	8.3	10	83.3	1	8.3
Netherlands	16	3.0	460	85.3	28	5.2	35	6.5	103	10.6	507	52.3	261	26.9	99	10.2
N. Macedonia	0	0.0	47	63.5	9	12.2	18	24.3	1	2.0	12	24.5	29	59.2	7	14.3
Norway	3	2.3	108	84.4	1	0.8	16	12.5	11	5.6	125	64.1	50	25.6	9	4.6
Poland	13	1.4	721	75.7	105	11.0	114	12.0	7	1.3	209	39.3	267	50.2	49	9.2
Portugal	6	3.5	120	70.2	20	11.7	25	14.6	31	16.4	95	50.3	49	25.9	14	7.4
Romania	6	2.3	159	60.7	72	27.5	25	9.5	0	0.0	14	58.3	9	37.5	1	4.2
Russian Fed.	55	2.9	1068	57.0	489	26.1	262	14.0	37	7.3	168	32.9	271	53.1	34	6.7
Serbia	0	0.0	75	55.2	26	19.1	35	25.7	1	1.5	22	33.3	37	56.1	6	9.1
Slovak Rep.	0	0.0	111	88.1	6	4.8	9	7.1	9	6.1	75	51.0	48	32.7	15	10.2
Slovenia	0	0.0	42	76.4	0	0.0	13	23.6	12	26.1	23	50.0	8	17.4	3	6.5
Spain	18	1.7	813	76.8	82	7.7	146	13.8	74	6.1	659	54.6	357	29.6	117	9.7
Sweden	18	6.8	192	72.5	23	8.7	32	12.1	10	2.6	163	41.9	189	48.6	27	6.9
Switzerland	8	2.0	355	87.2	19	4.7	25	6.1	96	17.7	286	52.6	120	22.1	42	7.7
Turkey	21	1.0	1630	76.1	283	13.2	207	9.7	6	1.6	185	50.1	134	36.3	44	11.9
Ukraine	18	6.8	143	54.0	69	26.0	35	13.2	0	0.0	17	21.5	54	68.4	8	10.1
United Kingdom	8	0.2	3434	83.0	125	3.0	571	13.8	11	0.2	3971	68.6	854	14.8	950	16.4
Total	272	1.2	17789	77.5	2094	9.1	2788	12.2	629	2.5	14122	56.4	7624	30.4	2671	10.7

Note: Albania, Armenia and Georgia have <5 adults seen in 2022 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 2022.

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 5.2   Prevalence of Burkholderia cepacia complex species in people with CF seen in 2022 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	1.5	65	97.0	0	0.0	1	1.5								
Armenia	2	9.1	20	90.9	0	0.0	0	0.0								
Austria	2	0.5	368	99.2	0	0.0	1	0.3	2	0.5	377	94.5	17	4.3	3	0.8
Belarus	0	0.0	144	98.6	2	1.4	0	0.0	0	0.0	5	100	0	0.0	0	0.0
Bulgaria	1	0.8	124	98.4	0	0.0	1	0.8	0	0.0	84	98.8	1	1.2	0	0.0
Croatia	1	1.2	80	97.6	0	0.0	1	1.2	0	0.0	55	100	0	0.0	0	0.0
Cyprus	0	0.0	7	100	0	0.0	0	0.0	2	12.5	13	81.3	0	0.0	1	6.3
Czech Rep.	19	5.9	302	93.2	3	0.9	0	0.0	35	11.6	238	78.8	29	9.6	0	0.0
Denmark	0	0.0	210	99.5	0	0.0	1	0.5	0	0.0	286	93.8	12	3.9	7	2.3
Finland	0	0.0	26	100	0	0.0	0	0.0	0	0.0	31	100	0	0.0	0	0.0
France	0	0.0	2660	98.9	7	0.3	24	0.9	0	0.0	3607	97.9	52	1.4	25	0.7
Georgia	11	26.8	30	73.2	0	0.0	0	0.0								
Germany	24	0.9	2712	98.4	11	0.4	9	0.3	102	2.7	3590	94.1	109	2.9	15	0.4
Greece	1	0.4	236	99.6	0	0.0	0	0.0	55	15.5	298	84.2	0	0.0	1	0.3
Hungary	2	0.8	227	95.8	2	0.8	6	2.5	5	2.6	175	90.2	2	1.0	12	6.2
Iceland	0	0.0	8	88.9	0	0.0	1	11.1	0	0.0	6	100	0	0.0	0	0.0
Ireland	0	0.0	523	99.6	1	0.2	1	0.2	0	0.0	707	98.3	7	1.0	5	0.7
Israel	10	6.3	147	92.5	1	0.6	1	0.6	26	7.7	308	90.6	4	1.2	2	0.6
Italy	4	0.2	2192	99.5	6	0.3	2	0.1	8	0.2	3465	97.0	87	2.4	13	0.4
Latvia	0	0.0	32	100	0	0.0	0	0.0	0	0.0	13	92.9	1	7.1	0	0.0
Lithuania	2	10.0	17	85.0	1	5.0	0	0.0	0	0.0	20	87.0	3	13.0	0	0.0
Luxembourg	0	0.0	21	100	0	0.0	0	0.0	0	0.0	5	100	0	0.0	0	0.0
Rep. Moldova	36	100	0	0.0	0	0.0	0	0.0	12	100	0	0.0	0	0.0	0	0.0
Netherlands	32	5.9	500	92.8	4	0.7	3	0.6	103	10.6	842	86.8	18	1.9	7	0.7
N. Macedonia	0	0.0	72	97.3	2	2.7	0	0.0	1	2.0	47	95.9	0	0.0	1	2.0
Norway	3	2.3	124	96.9	1	0.8	0	0.0	11	5.6	176	90.3	5	2.6	3	1.5
Poland	13	1.4	932	97.8	4	0.4	4	0.4	14	2.6	496	93.2	13	2.4	9	1.7
Portugal	3	1.8	164	95.9	1	0.6	3	1.8	31	16.4	145	76.7	13	6.9	0	0.0
Romania	7	2.7	253	96.6	0	0.0	2	0.8	0	0.0	24	100	0	0.0	0	0.0
Russian Fed.	62	3.3	1764	94.1	21	1.1	27	1.4	39	7.7	398	78.0	68	13.3	5	1.0
Serbia	0	0.0	128	94.1	4	2.9	4	2.9	1	1.5	54	81.8	11	16.7	0	0.0
Slovak Rep.	1	0.8	124	98.4	0	0.0	1	0.8	9	6.1	127	86.4	8	5.4	3	2.0
Slovenia	0	0.0	55	100	0	0.0	0	0.0	12	26.1	34	73.9	0	0.0	0	0.0
Spain	22	2.1	1025	96.8	4	0.4	8	0.8	82	6.8	1051	87.1	64	5.3	10	0.8
Sweden	27	10.2	236	89.1	2	0.8	0	0.0	20	5.1	355	91.3	11	2.8	3	0.8
Switzerland	11	2.7	394	96.8	2	0.5	0	0.0	94	17.3	433	79.6	11	2.0	6	1.1
Turkey	22	1.0	2112	98.7	6	0.3	1	0.1	6	1.6	360	97.6	3	0.8	0	0.0
Ukraine	18	6.8	243	91.7	1	0.4	3	1.1	2	2.5	76	96.2	0	0.0	1	1.3
United Kingdom	8	0.2	4079	98.6	0	0.0	51	1.2	11	0.2	5590	96.6	0	0.0	185	3.2
Total	345	1.5	22356	97.4	86	0.4	156	0.7	683	2.7	23497	93.82	549	2.2	317	1.3

Note: Albania, Armenia and Georgia have <5 adults seen in 2022 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 2022.

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 5.3   Prevalence of Haemophilus influenzae in people with CF seen in 2022 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	2	3.0	65	97.0	0	0.0	0	0.0								
Armenia	1	4.6	21	95.5	0	0.0	0	0.0								
Austria	3	0.8	233	62.8	38	10.2	97	26.2	2	0.5	324	81.2	30	7.5	43	10.8
Belarus	0	0.0	134	91.8	12	8.2	0	0.0	0	0.0	5	100	0	0.0	0	0.0
Bulgaria	1	0.8	119	94.4	0	0.0	6	4.8	0	0.0	85	100	0	0.0	0	0.0
Croatia	1	1.2	67	81.7	0	0.0	14	17.1	0	0.0	47	85.5	0	0.0	8	14.6
Cyprus	0	0.0	6	85.7	0	0.0	1	14.3	2	12.5	12	75.0	0	0.0	2	12.5
Czech Rep.	34	10.5	283	87.4	2	0.6	5	1.5	40	13.3	258	85.4	0	0.0	4	1.3
Denmark	0	0.0	107	50.7	36	17.1	68	32.2	0	0.0	226	74.1	8	2.6	71	23.3
Finland	0	0.0	25	96.2	1	3.9	0	0.0	0	0.0	31	100	0	0.0	0	0.0
France	0	0.0	2223	82.6	0	0.0	468	17.4	0	0.0	3267	88.7	0	0.0	417	11.3
Georgia	4	9.8	37	90.2	0	0.0	0	0.0								
Germany	24	0.9	2097	76.1	0	0.0	635	23.0	103	2.7	3443	90.2	0	0.0	270	7.1
Greece	2	0.8	226	95.4	0	0.0	9	3.8	56	15.8	277	78.3	2	0.6	19	5.4
Hungary	3	1.3	202	85.2	6	2.5	26	11.0	5	2.6	183	94.3	1	0.5	5	2.6
Iceland	0	0.0	4	44.4	1	11.1	4	44.4	0	0.0	6	100	0	0.0	0	0.0
Ireland	0	0.0	465	88.6	9	1.7	51	9.7	0	0.0	695	96.7	2	0.3	22	3.1
Israel	10	6.3	132	83.0	4	2.5	13	8.2	24	7.1	285	83.8	10	2.9	21	6.2
Italy	4	0.2	1653	75.0	202	9.2	345	15.7	9	0.3	3049	85.3	345	9.7	170	4.8
Latvia	1	3.1	21	65.6	3	9.4	7	21.9	0	0.0	10	71.4	0	0.0	4	28.6
Lithuania	1	5.0	12	60.0	0	0.0	7	35.0	0	0.0	22	95.7	0	0.0	1	4.4
Luxembourg	0	0.0	15	71.4	1	4.8	5	23.8	0	0.0	4	80.0	1	20.0	0	0.0
Rep. Moldova	1	2.8	35	97.2	0	0.0	0	0.0	0	0.0	12	100	0	0.0	0	0.0
Netherlands	16	3.0	417	77.4	16	3.0	90	16.7	104	10.7	766	79.0	50	5.2	50	5.2
N. Macedonia	0	0.0	74	100	0	0.0	0	0.0	1	2.0	48	98.0	0	0.0	0	0.0
Norway	3	2.3	62	48.4	5	3.9	58	45.3	11	5.6	150	76.9	4	2.1	30	15.4
Poland	13	1.4	767	80.5	14	1.5	159	16.7	14	2.6	477	89.7	5	0.9	36	6.8
Portugal	3	1.8	131	76.6	3	1.8	34	19.9	32	16.9	134	70.9	4	2.1	19	10.1
Romania	7	2.7	250	95.4	0	0.0	5	1.9	0	0.0	24	100	0	0.0	0	0.0
Russian Fed.	52	2.8	1728	92.2	8	0.4	86	4.6	35	6.9	467	91.6	0	0.0	8	1.6
Serbia	0	0.0	17	12.5	77	56.6	42	30.9	2	3.0	22	33.3	24	36.4	18	27.3
Slovak Rep.	1	0.8	110	87.3	1	0.8	14	11.1	9	6.1	126	85.7	1	0.7	11	7.5
Slovenia	0	0.0	33	60.0	7	12.7	15	27.3	12	26.1	30	65.2	0	0.0	4	8.7
Spain	16	1.5	808	76.3	23	2.2	212	20.0	78	6.5	1017	84.3	22	1.8	90	7.5
Sweden	27	10.2	200	75.5	2	0.8	36	13.6	30	7.7	308	79.2	5	1.3	46	11.8
Switzerland	5	1.2	307	75.4	20	4.9	75	18.4	89	16.4	386	71.0	20	3.7	49	9.0
Turkey	22	1.0	2008	93.8	24	1.1	87	4.1	7	1.9	345	93.5	2	0.5	15	4.1
Ukraine	19	7.2	232	87.6	0	0.0	14	5.3	1	1.3	78	98.7	0	0.0	0	0.0
United Kingdom	8	0.2	3537	85.5	0	0.0	593	14.3	11	0.2	5516	95.3	0	0.0	259	4.5
Total	284	1.2	18863	82.2	515	2.2	3281	14.3	677	2.7	22140	88.4	536	2.1	1693	6.8

Note: Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the table for adults, but the people are included in the total number.

Note: Belgium, France, Germany 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 2022.

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

5. Microbiology

Table 5.4 Prevalence of Staphylococcus aureus in people with CF seen in 2022 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	18	26.9	13	19.4	36	53.7								
Armenia	0	0.0	0	0.0	9	40.9	13	59.1								
Austria	2	0.5	67	18.1	200	53.9	102	27.5	1	0.3	119	29.8	215	53.9	64	16.0
Belarus	0	0.0	79	54.1	67	45.9	0	0.0	0	0.0	1	20.0	4	80.0	0	0.0
Bulgaria	1	0.8	91	72.2	4	3.2	30	23.8	0	0.0	68	80.0	4	4.7	13	15.3
Croatia	1	1.2	15	18.3	25	30.5	41	50.0	0	0.0	24	43.6	18	32.7	13	23.6
Cyprus	0	0.0	3	42.9	1	14.3	3	42.9	2	12.5	9	56.3	3	18.8	2	12.5
Czech Rep.	17	5.3	68	21.0	106	32.7	133	41.1	35	11.6	128	42.4	72	23.8	67	22.2
Denmark	0	0.0	86	40.8	36	17.1	89	42.2	0	0.0	144	47.2	55	18.0	106	34.8
Finland	0	0.0	20	76.9	6	23.1	0	0.0	0	0.0	12	38.7	19	61.3	0	0.0
France	0	0.0	732	27.2	980	36.4	979	36.4	0	0.0	1883	51.1	1310	35.6	491	13.3
Georgia	5	12.2	19	46.3	8	19.5	9	22.0								
Germany	26	0.9	740	26.9	1080	39.2	910	33.0	117	3.1	1205	31.6	1865	48.9	629	16.5
Greece	3	1.3	133	56.1	65	27.4	36	15.2	23	6.5	147	41.5	149	42.1	35	9.9
Hungary	2	0.8	49	20.7	115	48.5	71	30.0	5	2.6	78	40.2	33	17.0	78	40.2
Iceland	0	0.0	2	22.2	4	44.4	3	33.3	0	0.0	4	66.7	1	16.7	1	16.7
Ireland	0	0.0	353	67.2	103	19.6	69	13.1	0	0.0	574	79.8	70	9.7	75	10.4
Israel	8	5.0	57	35.9	37	23.3	57	35.9	25	7.4	192	56.5	63	18.5	60	17.7
Italy	4	0.2	716	32.5	811	36.8	673	30.5	10	0.3	1497	41.9	1274	35.7	792	22.2
Latvia	0	0.0	2	6.3	22	68.8	8	25.0	0	0.0	3	21.4	11	78.6	0	0.0
Lithuania	0	0.0	4	20.0	10	50.0	6	30.0	0	0.0	6	26.1	13	56.5	4	17.4
Luxembourg	0	0.0	5	23.8	13	61.9	3	14.3	0	0.0	2	40.0	3	60.0	0	0.0
Rep. Moldova	1	2.8	1	2.8	32	88.9	2	5.6	0	0.0	0	0.0	12	100	0	0.0
Netherlands	15	2.8	207	38.4	135	25.1	182	33.8	102	10.5	446	46.0	276	28.5	146	15.1
N. Macedonia	0	0.0	48	64.9	9	12.2	17	23.0	1	2.0	24	49.0	7	14.3	17	34.7
Norway	3	2.3	33	25.8	49	38.3	43	33.6	11	5.6	60	30.8	67	34.4	57	29.2
Poland	13	1.4	195	20.5	506	53.1	239	25.1	12	2.3	137	25.8	301	56.6	82	15.4
Portugal	5	2.9	74	43.3	52	30.4	40	23.4	30	15.9	71	37.6	49	25.9	39	20.6
Romania	10	3.8	199	76.0	27	10.3	26	9.9	0	0.0	10	41.7	8	33.3	6	25.0
Russian Fed.	56	3.0	507	27.1	1046	55.8	265	14.1	35	6.9	163	32.0	287	56.3	25	4.9
Serbia	0	0.0	20	14.7	90	66.2	26	19.1	1	1.5	16	24.2	32	48.5	17	25.8
Slovak Rep.	0	0.0	48	38.1	43	34.1	35	27.8	9	6.1	69	46.9	44	29.9	25	17.0
Slovenia	0	0.0	3	5.5	46	83.6	6	10.9	11	23.9	12	26.1	14	30.4	9	19.6
Spain	13	1.2	355	33.5	374	35.3	317	29.9	75	6.2	563	46.6	372	30.8	197	16.3
Sweden	21	7.9	85	32.1	59	22.3	100	37.7	38	9.8	120	30.9	175	45.0	56	14.4
Switzerland	10	2.5	94	23.1	188	46.2	115	28.3	94	17.3	231	42.5	151	27.8	68	12.5
Turkey	22	1.0	1520	71.0	342	16.0	257	12.0	6	1.6	217	58.8	86	23.3	60	16.3
Ukraine	23	8.7	69	26.0	126	47.6	47	17.7	0	0.0	30	38.0	40	50.6	9	11.4
United Kingdom	7	0.2	3279	79.2	281	6.8	571	13.8	11	0.2	4460	77.1	365	6.3	950	16.4
Total	268	1.2	9996	43.6	7120	31.0	5559	24.2	654	2.6	12725	50.8	7470	29.8	4197	16.8

Note: Albania, Armenia and Georgia have <5 adults seen in 2022 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 2022.

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 *Staphylococcus aureus* in children and adults, by country and overall.

5. Microbiology

Table 5.5   Prevalence of methicillin-resistant Staphylococcus aureus (MRSA) in people with CF seen in 2022 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	1.5	58	86.6	0	0.0	8	11.9								
Armenia	7	31.8	15	68.2	0	0.0	0	0.0								
Austria	2	0.5	357	96.2	4	1.1	8	2.2	5	1.3	385	96.5	8	2.0	1	0.3
Belarus	0	0.0	134	91.8	12	8.2	0	0.0	0	0.0	5	100	0	0.0	0	0.0
Bulgaria	1	0.8	125	99.2	0	0.0	0	0.0	0	0.0	83	97.7	1	1.2	1	1.2
Croatia	2	2.4	78	95.1	0	0.0	2	2.4	1	1.8	52	94.6	0	0.0	2	3.6
Cyprus	0	0.0	5	71.4	1	14.3	1	14.3	2	12.5	12	75.0	0	0.0	2	12.5
Czech Rep.	20	6.2	304	93.8	0	0.0	0	0.0	37	12.3	257	85.1	0	0.0	8	2.7
Denmark	0	0.0	211	100	0	0.0	0	0.0	0	0.0	305	100	0	0.0	0	0.0
Finland	0	0.0	26	100	0	0.0	0	0.0	0	0.0	30	96.8	1	3.2	0	0.0
France	0	0.0	2619	97.3	29	1.1	43	1.6	0	0.0	3534	95.9	101	2.7	49	1.3
Georgia	7	17.1	34	82.9	0	0.0	0	0.0								
Germany	33	1.2	2648	96.1	52	1.9	23	0.8	121	3.2	3460	90.7	196	5.1	39	1.0
Greece	2	0.8	209	88.2	10	4.2	16	6.8	50	14.1	274	77.4	10	2.8	20	5.7
Hungary	4	1.7	220	92.8	6	2.5	7	3.0	5	2.6	180	92.8	6	3.1	3	1.6
Iceland	0	0.0	9	100	0	0.0	0	0.0	0	0.0	6	100	0	0.0	0	0.0
Ireland	0	0.0	513	97.7	5	1.0	7	1.3	0	0.0	708	98.5	3	0.4	8	1.1
Israel	12	7.6	134	84.3	0	0.0	13	8.2	27	7.9	292	85.9	7	2.1	14	4.1
Italy	4	0.2	1996	90.6	76	3.5	128	5.8	8	0.2	3283	91.9	116	3.3	166	4.7
Latvia	0	0.0	31	96.9	1	3.1	0	0.0	0	0.0	13	92.9	0	0.0	1	7.1
Lithuania	1	5.0	19	95.0	0	0.0	0	0.0	0	0.0	23	100	0	0.0	0	0.0
Luxembourg	0	0.0	21	100	0	0.0	0	0.0	0	0.0	4	80.0	1	20.0	0	0.0
Rep. Moldova	1	2.8	35	97.2	0	0.0	0	0.0	0	0.0	12	100	0	0.0	0	0.0
Netherlands	123	22.8	410	76.1	3	0.6	3	0.6	109	11.2	848	87.4	11	1.1	2	0.2
N. Macedonia	0	0.0	51	68.9	5	6.8	18	24.3	1	2.0	31	63.3	9	18.4	8	16.3
Norway	3	2.3	123	96.1	0	0.0	2	1.6	11	5.6	182	93.3	2	1.0	0	0.0
Poland	17	1.8	907	95.2	19	2.0	10	1.1	17	3.2	484	91.0	21	4.0	10	1.9
Portugal	3	1.8	157	91.8	3	1.8	8	4.7	32	16.9	144	76.2	6	3.2	7	3.7
Romania	11	4.2	206	78.6	24	9.2	21	8.0	0	0.0	17	70.8	3	12.5	4	16.7
Russian Fed.	62	3.3	1712	91.4	22	1.2	78	4.2	37	7.3	452	88.6	16	3.1	5	1.0
Serbia	2	1.5	117	86.0	11	8.1	6	4.4	3	4.6	54	81.8	7	10.6	2	3.0
Slovak Rep.	1	0.8	117	92.9	3	2.4	5	4.0	11	7.5	123	83.7	7	4.8	6	4.1
Slovenia	0	0.0	51	92.7	3	5.5	1	1.8	13	28.3	30	65.2	1	2.2	2	4.4
Spain	22	2.1	986	93.1	15	1.4	36	3.4	81	6.7	1052	87.2	39	3.2	35	2.9
Sweden	28	10.6	230	86.8	1	0.4	6	2.3	21	5.4	362	93.1	5	1.3	1	0.3
Switzerland	8	2.0	392	96.3	3	0.7	4	1.0	90	16.5	450	82.7	1	0.2	3	0.6
Turkey	23	1.1	1875	87.6	99	4.6	144	6.7	6	1.6	328	88.9	14	3.8	21	5.7
Ukraine	21	7.9	239	90.2	0	0.0	5	1.9	1	1.3	73	92.4	5	6.3	0	0.0
United Kingdom	8	0.2	4055	98.0	0	0.0	75	1.8	11	0.2	5672	98.0	0	0.0	103	1.8
Total	429	1.9	21429	93.4	407	1.8	678	3.0	700	2.8	23226	92.7	597	2.4	523	2.1

Note: Albania, Armenia and Georgia have <5 adults seen in 2022 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 2022.

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

This table shows the frequency of methicillin-resistant *Staphylococcus aureus* in children and adults, by country and overall.

5. Microbiology

Table 5.6   Prevalence of Stenotrophomonas maltophilia in people with CF seen in 2022 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	1.5	63	94.0	0	0.0	3	4.5								
Armenia	8	36.4	14	63.6	0	0.0	0	0.0								
Austria	2	0.5	337	90.8	10	2.7	22	5.9	1	0.3	348	87.2	17	4.3	33	8.3
Belarus	0	0.0	143	98.0	3	2.1	0	0.0	0	0.0	5	100	0	0.0	0	0.0
Bulgaria	1	0.8	124	98.4	0	0.0	1	0.8	0	0.0	84	98.8	0	0.0	1	1.2
Croatia	2	2.4	71	86.6	0	0.0	9	11.0	1	1.8	49	89.1	0	0.0	5	9.1
Cyprus	0	0.0	7	100	0	0.0	0	0.0	2	12.5	14	87.5	0	0.0	0	0.0
Czech Rep.	20	6.2	303	93.5	0	0.0	1	0.3	38	12.6	263	87.1	0	0.0	1	0.3
Denmark	0	0.0	186	88.2	5	2.4	20	9.5	0	0.0	257	84.3	19	6.2	29	9.5
Finland	0	0.0	25	96.2	1	3.9	0	0.0	0	0.0	30	96.8	1	3.2	0	0.0
France	0	0.0	2418	89.9	31	1.2	242	9.0	0	0.0	3506	95.2	47	1.3	131	3.6
Georgia	10	24.4	31	75.6	0	0.0	0	0.0								
Germany	24	0.9	2586	93.8	26	0.9	120	4.4	102	2.7	3374	88.4	192	5.0	148	3.9
Greece	2	0.8	215	90.7	2	0.8	18	7.6	49	13.8	287	81.1	4	1.1	14	4.0
Hungary	4	1.7	225	94.9	0	0.0	8	3.4	6	3.1	184	94.9	3	1.6	1	0.5
Iceland	1	11.1	7	77.8	0	0.0	1	11.1	0	0.0	5	83.3	0	0.0	1	16.7
Ireland	0	0.0	508	96.8	2	0.4	15	2.9	0	0.0	711	98.9	1	0.1	7	1.0
Israel	10	6.3	131	82.4	1	0.6	17	10.7	25	7.4	295	86.8	3	0.9	17	5.0
Italy	4	0.2	1998	90.7	15	0.7	187	8.5	8	0.2	3309	92.6	63	1.8	193	5.4
Latvia	0	0.0	28	87.5	1	3.1	3	9.4	0	0.0	13	92.9	1	7.1	0	0.0
Lithuania	1	5.0	14	70.0	0	0.0	5	25.0	0	0.0	19	82.6	1	4.4	3	13.0
Luxembourg	0	0.0	19	90.5	1	4.8	1	4.8	0	0.0	4	80.0	0	0.0	1	20.0
Rep. Moldova	34	94.4	1	2.8	0	0.0	1	2.8	12	100	0	0.0	0	0.0	0	0.0
Netherlands	18	3.3	493	91.5	7	1.3	21	3.9	104	10.7	817	84.2	16	1.7	33	3.4
N. Macedonia	0	0.0	71	96.0	0	0.0	3	4.1	1	2.0	44	89.8	0	0.0	4	8.2
Norway	3	2.3	111	86.7	1	0.8	13	10.2	10	5.1	152	78.0	8	4.1	25	12.8
Poland	12	1.3	909	95.4	1	0.1	31	3.3	15	2.8	491	92.3	6	1.1	20	3.8
Portugal	5	2.9	156	91.2	0	0.0	10	5.9	32	16.9	146	77.3	3	1.6	8	4.2
Romania	7	2.7	255	97.3	0	0.0	0	0.0	0	0.0	24	100	0	0.0	0	0.0
Russian Fed.	65	3.5	1732	92.4	17	0.9	60	3.2	38	7.5	451	88.4	8	1.6	13	2.6
Serbia	0	0.0	111	81.6	1	0.7	24	17.7	1	1.5	58	87.9	0	0.0	7	10.6
Slovak Rep.	2	1.6	118	93.7	0	0.0	6	4.8	10	6.8	129	87.8	3	2.0	5	3.4
Slovenia	0	0.0	49	89.1	1	1.8	5	9.1	11	23.9	31	67.4	1	2.2	3	6.5
Spain	20	1.9	969	91.5	4	0.4	66	6.2	78	6.5	1032	85.5	29	2.4	68	5.6
Sweden	27	10.2	213	80.4	3	1.1	22	8.3	27	6.9	327	84.1	10	2.6	25	6.4
Switzerland	7	1.7	386	94.8	5	1.2	9	2.2	90	16.5	431	79.2	12	2.2	11	2.0
Turkey	26	1.2	2094	97.8	6	0.3	15	0.7	9	2.4	346	93.8	4	1.1	10	2.7
Ukraine	18	6.8	233	87.9	4	1.5	10	3.8	1	1.3	74	93.7	0	0.0	4	5.1
United Kingdom	8	0.2	3932	95.0	0	0.0	198	4.8	11	0.2	5559	96.1	0	0.0	216	3.7
Total	342	1.5	21286	92.8	148	0.7	1167	5.1	682	2.7	22875	91.3	452	1.8	1037	4.1

Note: Albania, Armenia and Georgia have <5 adults seen in 2022 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 2022.

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 5.7   Prevalence of Achromobacter species infection in people with CF seen in 2022 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	2	3.0	64	95.5	0	0.0	1	1.5								
Armenia	9	40.9	13	59.1	0	0.0	0	0.0								
Austria	2	0.5	359	96.8	3	0.8	7	1.9	2	0.5	376	94.2	13	3.3	8	2.0
Belarus	0	0.0	140	95.9	6	4.1	0	0.0	0	0.0	4	80.0	1	20.0	0	0.0
Bulgaria	1	0.8	125	99.2	0	0.0	0	0.0	0	0.0	85	100	0	0.0	0	0.0
Croatia	2	2.4	74	90.2	0	0.0	6	7.3	1	1.8	53	96.4	0	0.0	1	1.8
Cyprus	0	0.0	7	100	0	0.0	0	0.0	2	12.5	12	75.0	0	0.0	2	12.5
Czech Rep.	18	5.6	306	94.4	0	0.0	0	0.0	37	12.3	259	85.8	0	0.0	6	2.0
Denmark	0	0.0	206	97.6	4	1.9	1	0.5	0	0.0	274	89.8	13	4.3	18	5.9
Finland	0	0.0	26	100	0	0.0	0	0.0	0	0.0	31	100	0	0.0	0	0.0
France	0	0.0	2595	96.4	33	1.2	63	2.3	0	0.0	3509	95.3	109	3.0	66	1.8
Georgia	7	17.1	34	82.9	0	0.0	0	0.0								
Germany	24	0.9	2696	97.8	17	0.6	19	0.7	102	2.7	3485	91.3	190	5.0	39	1.0
Greece	2	0.8	230	97.1	0	0.0	5	2.1	50	14.1	287	81.1	7	2.0	10	2.8
Hungary	6	2.5	217	91.6	6	2.5	8	3.4	7	3.6	171	88.1	8	4.1	8	4.1
Iceland	0	0.0	9	100	0	0.0	0	0.0	0	0.0	5	83.3	0	0.0	1	16.7
Ireland	0	0.0	522	99.4	1	0.2	2	0.4	0	0.0	709	98.6	1	0.1	9	1.3
Israel	13	8.2	139	87.4	1	0.6	6	3.8	28	8.2	299	87.9	5	1.5	8	2.4
Italy	4	0.2	2118	96.1	32	1.5	50	2.3	8	0.2	3294	92.2	175	4.9	96	2.7
Latvia	0	0.0	29	90.6	0	0.0	3	9.4	0	0.0	12	85.7	1	7.1	1	7.1
Lithuania	1	5.0	19	95.0	0	0.0	0	0.0	0	0.0	21	91.3	1	4.4	1	4.4
Luxembourg	0	0.0	21	100	0	0.0	0	0.0	0	0.0	5	100	0	0.0	0	0.0
Rep. Moldova	34	94.4	2	5.6	0	0.0	0	0.0	12	100	0	0.0	0	0.0	0	0.0
Netherlands	131	24.3	404	75.0	0	0.0	4	0.7	375	38.7	572	59.0	16	1.7	7	0.7
N. Macedonia	0	0.0	74	100	0	0.0	0	0.0	1	2.0	47	95.9	0	0.0	1	2.0
Norway	3	2.3	122	95.3	1	0.8	2	1.6	12	6.2	177	90.8	1	0.5	5	2.6
Poland	16	1.7	922	96.8	4	0.4	11	1.2	16	3.0	477	89.7	21	4.0	18	3.4
Portugal	5	2.9	159	93.0	3	1.8	4	2.3	33	17.5	150	79.4	3	1.6	3	1.6
Romania	11	4.2	250	95.4	0	0.0	1	0.4	2	8.3	22	91.7	0	0.0	0	0.0
Russian Fed.	68	3.6	1695	90.5	52	2.8	59	3.2	39	7.7	419	82.2	36	7.1	16	3.1
Serbia	2	1.5	120	88.2	3	2.2	11	8.1	3	4.6	57	86.4	2	3.0	4	6.1
Slovak Rep.	2	1.6	120	95.2	1	0.8	3	2.4	10	6.8	129	87.8	5	3.4	3	2.0
Slovenia	0	0.0	54	98.2	0	0.0	1	1.8	12	26.1	33	71.7	1	2.2	0	0.0
Spain	22	2.1	1006	95.0	5	0.5	26	2.5	81	6.7	1029	85.3	58	4.8	39	3.2
Sweden	27	10.2	232	87.6	4	1.5	2	0.8	29	7.5	336	86.4	13	3.3	11	2.8
Switzerland	7	1.7	393	96.6	1	0.3	6	1.5	89	16.4	445	81.8	7	1.3	3	0.6
Turkey	24	1.1	2100	98.1	6	0.3	11	0.5	9	2.4	349	94.6	3	0.8	8	2.2
Ukraine	19	7.2	235	88.7	4	1.5	7	2.6	2	2.5	75	94.9	1	1.3	1	1.3
United Kingdom	8	0.2	4074	98.5	0	0.0	56	1.4	11	0.2	5636	97.4	0	0.0	139	2.4
Total	470	2.1	21911	95.5	187	0.8	375	1.6	973	3.9	22850	91.2	691	2.8	532	2.1

Note: Albania, Armenia and Georgia have <5 adults seen in 2022 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 2022.

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 7.1   Prevalence of allergic bronchopulmonary aspergillosis (ABPA) in all people with CF seen in 2022 who have never had a transplant, by country and overall.

Country	Children (<18 years)						Adults (≥18 years)					
	Missing/ Unknown		No ABPA		Yes, current ABPA		Missing/ Unknown		No ABPA		Yes, current ABPA	
	N	%	N	%	N	%	N	%	N	%	N	%
Albania	3	4.5	64	95.5	0	0.0						
Armenia	0	0.0	22	100	0	0.0						
Austria	3	0.8	363	97.8	5	1.4	38	9.5	350	87.7	11	2.8
Belarus	0	0.0	146	100	0	0.0	0	0.0	5	100	0	0.0
Bulgaria	2	1.6	122	96.8	2	1.6	2	2.4	83	97.7	0	0.0
Croatia	1	1.2	81	98.8	0	0.0	1	1.8	54	98.2	0	0.0
Cyprus	0	0.0	7	100	0	0.0	0	0.0	16	100	0	0.0
Czech Republic	8	2.5	314	96.9	2	0.6	18	6.0	283	93.7	1	0.3
Denmark	211	100	0	0.0	0	0.0	305	100	0	0.0	0	0.0
Finland	0	0.0	26	100	0	0.0	0	0.0	30	96.8	1	3.2
France	0	0.0	2618	97.3	73	2.7	0	0.0	3493	94.8	191	5.2
Georgia	3	7.3	38	92.7	0	0.0						
Germany	4	0.2	2719	98.7	33	1.2	13	0.3	3652	95.7	151	4.0
Greece	1	0.4	234	98.7	2	0.8	5	1.4	341	96.3	8	2.3
Hungary	0	0.0	236	99.6	1	0.4	4	2.1	184	94.9	6	3.1
Iceland	0	0.0	9	100	0	0.0	0	0.0	6	100	0	0.0
Ireland	16	3.1	495	94.3	14	2.7	16	2.2	640	89.0	63	8.8
Israel	3	1.9	153	96.2	3	1.9	8	2.4	318	93.5	14	4.1
Italy	11	0.5	2171	98.5	22	1.0	31	0.9	3463	96.9	79	2.2
Latvia	4	12.5	28	87.5	0	0.0	0	0.0	14	100	0	0.0
Lithuania	1	5.0	18	90.0	1	5.0	0	0.0	23	100	0	0.0
Luxembourg	0	0.0	21	100	0	0.0	4	80.0	1	20.0	0	0.0
Rep of Moldova	2	5.6	34	94.4	0	0.0	0	0.0	12	100	0	0.0
The Netherlands	9	1.7	515	95.6	15	2.8	21	2.2	897	92.5	52	5.4
North Macedonia	0	0.0	74	100	0	0.0	3	6.1	44	89.8	2	4.1
Norway	0	0.0	128	100	0	0.0	0	0.0	191	98.0	4	2.1
Poland	7	0.7	939	98.5	7	0.7	9	1.7	504	94.7	19	3.6
Portugal	3	1.8	168	98.3	0	0.0	12	6.4	176	93.1	1	0.5
Romania	1	0.4	259	98.9	2	0.8	0	0.0	23	95.8	1	4.2
Russian Fed.	26	1.4	1806	96.4	42	2.2	25	4.9	465	91.2	20	3.9
Serbia	0	0.0	134	98.5	2	1.5	1	1.5	64	97.0	1	1.5
Slovak Republic	3	2.4	120	95.2	3	2.4	18	12.2	125	85.0	4	2.7
Slovenia	0	0.0	52	94.6	3	5.5	8	17.4	38	82.6	0	0.0
Spain	47	4.4	987	93.2	25	2.4	53	4.4	1127	93.4	27	2.2
Sweden	0	0.0	263	99.3	2	0.8	0	0.0	378	97.2	11	2.8
Switzerland	5	1.2	390	95.8	12	3.0	3	0.6	479	88.1	62	11.4
Turkey	5	0.2	2105	98.3	31	1.5	5	1.4	347	94.0	17	4.6
Ukraine	7	2.6	255	96.2	3	1.1	2	2.5	76	96.2	1	1.3
United Kingdom	0	0.0	4036	97.5	102	2.5	0	0.0	5382	93.0	404	7.0
Total	386	1.7	22150	96.5	407	1.8	605	2.4	23290	93.0	1151	4.6

Note: Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the table for adults, but the people are included in the total number.

7. Complications

Table 7.2   Prevalence of CF-related diabetes (CFRD) in adults with CF seen in 2022 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	%
Austria	0	0.0	285	71.4	90	22.6	2	0.5	21	5.3	1	0.3
Belarus	0	0.0	5	100	0	0.0	0	0.0	0	0.0	0	0.0
Bulgaria	2	2.4	74	87.1	9	10.6	0	0.0	0	0.0	0	0.0
Croatia	0	0.0	42	76.4	11	20.0	0	0.0	2	3.6	0	0.0
Cyprus	0	0.0	15	93.8	1	6.3	0	0.0	0	0.0	0	0.0
Czech Republic	17	5.6	191	63.3	77	25.5	0	0.0	9	3.0	8	2.7
Denmark	0	0.0	232	76.1	73	23.9	0	0.0	0	0.0	0	0.0
Finland	0	0.0	21	67.7	10	32.3	0	0.0	0	0.0	0	0.0
France	0	0.0	2805	76.1	633	17.2	51	1.4	195	5.3	0	0.0
Germany	22	0.6	2591	67.9	832	21.8	60	1.6	69	1.8	242	6.3
Greece	2	0.6	261	73.7	76	21.5	3	0.9	11	3.1	1	0.3
Hungary	5	2.6	153	78.9	34	17.5	0	0.0	2	1.0	0	0.0
Iceland	0	0.0	2	33.3	3	50.0	0	0.0	1	16.7	0	0.0
Ireland	27	3.8	515	71.6	138	19.2	0	0.0	39	5.4	0	0.0
Israel	8	2.4	216	63.5	86	25.3	7	2.1	12	3.5	11	3.2
Italy	34	1.0	2788	78.0	743	20.8	1	0.0	7	0.2	0	0.0
Latvia	0	0.0	12	85.7	0	0.0	0	0.0	2	14.3	0	0.0
Lithuania	1	4.4	21	91.3	1	4.4	0	0.0	0	0.0	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	10	83.3	2	16.7	0	0.0	0	0.0	0	0.0
The Netherlands	1	0.1	630	65.0	267	27.5	18	1.9	50	5.2	4	0.4
North Macedonia	0	0.0	32	65.3	17	34.7	0	0.0	0	0.0	0	0.0
Norway	1	0.5	152	78.0	34	17.4	3	1.5	4	2.1	1	0.5
Poland	6	1.1	386	72.6	92	17.3	3	0.6	41	7.7	4	0.8
Portugal	7	3.7	151	79.9	22	11.6	0	0.0	9	4.8	0	0.0
Romania	1	4.2	19	79.2	3	12.5	0	0.0	1	4.2	0	0.0
Russian Fed.	22	4.3	423	82.9	48	9.4	1	0.2	16	3.1	0	0.0
Serbia	1	1.5	40	60.6	25	37.9	0	0.0	0	0.0	0	0.0
Slovak Republic	4	2.7	118	80.3	16	10.9	1	0.7	8	5.4	0	0.0
Slovenia	5	10.9	31	67.4	8	17.4	0	0.0	2	4.4	0	0.0
Spain	27	2.2	898	74.4	197	16.3	23	1.9	61	5.1	1	0.1
Sweden	0	0.0	279	71.7	81	20.8	8	2.1	0	0.0	21	5.4
Switzerland	36	6.6	375	68.9	122	22.4	2	0.4	9	1.7	0	0.0
Turkey	2	0.5	321	87.0	33	8.9	2	0.5	8	2.2	3	0.8
Ukraine	0	0.0	72	91.1	7	8.9	0	0.0	0	0.0	0	0.0
United Kingdom	41	0.7	3593	62.1	1545	26.7	173	3.0	165	2.9	269	4.7
Total	272	1.1	17767	70.9	5338	21.3	358	1.4	745	3.0	566	2.3

Note: Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the table for adults, but the people are included in the total number.

7. Complications

Table 7.3   Prevalence of liver disease in children and adolescents with CF (<18 years) seen in 2022 who have never had a transplant, by country and overall.

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	%
Albania	3	4.5	30	44.8	2	3.0	0	0.0	0	0.0	32	47.8	0	0.0
Armenia	0	0.0	12	54.6	0	0.0	1	4.6	0	0.0	9	40.9	0	0.0
Austria	1	0.3	187	50.4	4	1.1	9	2.4	2	0.5	168	45.3	0	0.0
Belarus	0	0.0	98	67.1	3	2.1	1	0.7	0	0.0	44	30.1	0	0.0
Bulgaria	2	1.6	46	36.5	6	4.8	9	7.1	0	0.0	63	50.0	0	0.0
Croatia	1	1.2	69	84.2	2	2.4	0	0.0	0	0.0	10	12.2	0	0.0
Cyprus	0	0.0	7	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Czech Republic	3	0.9	248	76.5	4	1.2	2	0.6	1	0.3	66	20.4	0	0.0
Denmark	52	24.6	131	62.1	2	1.0	1	0.5	0	0.0	24	11.4	1	0.5
Finland	0	0.0	19	73.1	2	7.7	1	3.9	0	0.0	4	15.4	0	0.0
France	0	0.0	2323	86.3	32	1.2	35	1.3	0	0.0	301	11.2	0	0.0
Georgia	4	9.8	37	90.2	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Germany	13	0.5	2091	75.9	31	1.1	43	1.6	11	0.4	567	20.6	0	0.0
Greece	0	0.0	185	78.1	1	0.4	1	0.4	0	0.0	50	21.1	0	0.0
Hungary	0	0.0	207	87.3	5	2.1	3	1.3	1	0.4	19	8.0	2	0.8
Iceland	0	0.0	9	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Ireland	27	5.1	469	89.3	4	0.8	3	0.6	3	0.6	19	3.6	0	0.0
Israel	3	1.9	127	79.9	2	1.3	5	3.1	0	0.0	22	13.8	0	0.0
Italy	13	0.6	1696	77.0	13	0.6	15	0.7	0	0.0	467	21.2	0	0.0
Latvia	0	0.0	24	75.0	1	3.1	0	0.0	0	0.0	7	21.9	0	0.0
Lithuania	0	0.0	17	85.0	0	0.0	1	5.0	0	0.0	2	10.0	0	0.0
Luxembourg	0	0.0	18	85.7	0	0.0	0	0.0	0	0.0	3	14.3	0	0.0
Rep of Moldova	0	0.0	32	88.9	0	0.0	0	0.0	0	0.0	4	11.1	0	0.0
The Netherlands	21	3.9	410	76.1	12	2.2	11	2.0	2	0.4	83	15.4	0	0.0
North Macedonia	0	0.0	50	67.6	2	2.7	2	2.7	0	0.0	20	27.0	0	0.0
Norway	3	2.3	116	90.6	1	0.8	0	0.0	0	0.0	8	6.3	0	0.0
Poland	7	0.7	641	67.3	25	2.6	8	0.8	2	0.2	269	28.2	1	0.1
Portugal	5	2.9	118	69.0	4	2.3	0	0.0	0	0.0	44	25.7	0	0.0
Romania	0	0.0	171	65.3	10	3.8	2	0.8	3	1.2	76	29.0	0	0.0
Russian Fed.	33	1.8	1362	72.7	56	3.0	52	2.8	3	0.2	368	19.6	0	0.0
Serbia	0	0.0	111	81.6	5	3.7	0	0.0	0	0.0	20	14.7	0	0.0
Slovak Republic	0	0.0	62	49.2	0	0.0	5	4.0	0	0.0	59	46.8	0	0.0
Slovenia	0	0.0	30	54.6	3	5.5	2	3.6	0	0.0	20	36.4	0	0.0
Spain	9	0.9	803	75.8	8	0.8	14	1.3	0	0.0	225	21.3	0	0.0
Sweden	15	5.7	219	82.6	3	1.1	5	1.9	0	0.0	23	8.7	0	0.0
Switzerland	6	1.5	344	84.5	4	1.0	1	0.3	5	1.2	47	11.6	0	0.0
Turkey	1	0.1	1832	85.6	20	0.9	17	0.8	14	0.7	257	12.0	0	0.0
Ukraine	4	1.5	225	84.9	6	2.3	5	1.9	1	0.4	24	9.1	0	0.0
United Kingdom	0	0.0	3676	88.8	32	0.8	10	0.2	0	0.0	420	10.2	0	0.0
Total	226	1.0	18252	79.6	305	1.3	264	1.2	48	0.2	3844	16.8	4	0.0

Note: Germany: oesophageal variceal bleeding is reported.

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

7. Complications

Table 7.4   Prevalence of liver disease in adults with CF (≥18 years) seen in 2022 who have never had a transplant, by country and overall.

| 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	%
Austria	2	0.5	196	49.1	15	3.8	14	3.5	2	0.5	170	42.6	0	0.0
Belarus	0	0.0	5	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Bulgaria	2	2.4	48	56.5	3	3.5	6	7.1	0	0.0	25	29.4	1	1.2
Croatia	0	0.0	42	76.4	3	5.5	0	0.0	0	0.0	10	18.2	0	0.0
Cyprus	0	0.0	14	87.5	0	0.0	0	0.0	0	0.0	2	12.5	0	0.0
Czech Republic	19	6.3	196	64.9	6	2.0	1	0.3	3	1.0	77	25.5	0	0.0
Denmark	97	31.8	143	46.9	11	3.6	2	0.7	1	0.3	51	16.7	0	0.0
Finland	0	0.0	23	74.2	1	3.2	0	0.0	0	0.0	7	22.6	0	0.0
France	0	0.0	2941	79.8	97	2.6	110	3.0	0	0.0	536	14.6	0	0.0
Germany	49	1.3	2203	57.7	125	3.3	84	2.2	57	1.5	1298	34.0	0	0.0
Greece	10	2.8	237	67.0	10	2.8	4	1.1	0	0.0	93	26.3	0	0.0
Hungary	3	1.6	150	77.3	1	0.5	1	0.5	3	1.6	35	18.0	1	0.5
Iceland	0	0.0	5	83.3	1	16.7	0	0.0	0	0.0	0	0.0	0	0.0
Ireland	17	2.4	574	79.8	31	4.3	6	0.8	12	1.7	79	11.0	0	0.0
Israel	6	1.8	286	84.1	11	3.2	1	0.3	1	0.3	35	10.3	0	0.0
Italy	39	1.1	1987	55.6	56	1.6	32	0.9	3	0.1	1456	40.8	0	0.0
Latvia	1	7.1	8	57.1	0	0.0	0	0.0	0	0.0	5	35.7	0	0.0
Lithuania	0	0.0	23	100	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
Luxembourg	0	0.0	3	60.0	0	0.0	0	0.0	0	0.0	2	40.0	0	0.0
Rep of Moldova	0	0.0	7	58.3	0	0.0	0	0.0	0	0.0	5	41.7	0	0.0
The Netherlands	13	1.3	690	71.1	76	7.8	21	2.2	2	0.2	168	17.3	0	0.0
North Macedonia	1	2.0	13	26.5	5	10.2	9	18.4	1	2.0	20	40.8	0	0.0
Norway	2	1.0	174	89.2	6	3.1	3	1.5	1	0.5	9	4.6	0	0.0
Poland	2	0.4	286	53.8	35	6.6	4	0.8	6	1.1	198	37.2	1	0.2
Portugal	4	2.1	140	74.1	3	1.6	5	2.7	0	0.0	37	19.6	0	0.0
Romania	0	0.0	12	50.0	2	8.3	3	12.5	0	0.0	7	29.2	0	0.0
Russian Fed.	27	5.3	386	75.7	17	3.3	19	3.7	1	0.2	60	11.8	0	0.0
Serbia	1	1.5	33	50.0	1	1.5	2	3.0	0	0.0	29	43.9	0	0.0
Slovak Republic	4	2.7	53	36.1	4	2.7	1	0.7	0	0.0	85	57.8	0	0.0
Slovenia	3	6.5	32	69.6	5	10.9	1	2.2	0	0.0	5	10.9	0	0.0
Spain	21	1.7	945	78.3	15	1.2	7	0.6	2	0.2	217	18.0	0	0.0
Sweden	9	2.3	296	76.1	10	2.6	3	0.8	0	0.0	71	18.3	0	0.0
Switzerland	4	0.7	388	71.3	28	5.2	4	0.7	7	1.3	113	20.8	0	0.0
Turkey	3	0.8	295	80.0	6	1.6	5	1.4	2	0.5	58	15.7	0	0.0
Ukraine	1	1.3	50	63.3	0	0.0	4	5.1	1	1.3	23	29.1	0	0.0
United Kingdom	0	0.0	4486	77.5	109	1.9	75	1.3	0	0.0	1116	19.3	0	0.0
Total	340	1.4	17374	69.4	693	2.8	427	1.7	105	0.4	6104	24.4	3	0.0

Note: Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the table for adults, but the people are included in the total number.

Note: Germany: oesophageal variceal bleeding is reported.

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

8. Therapies

Table 8.1   Use of inhaled hypertonic saline (NaCl) >3 consecutive months this year in all people with CF seen in 2022 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	2	3.0	1	1.5	64	95.5						
Armenia	0	0.0	0	0.0	22	100						
Austria	2	0.5	31	8.4	338	91.1	2	0.5	75	18.8	322	80.7
Belarus	0	0.0	46	31.5	100	68.5	0	0.0	3	60.0	2	40.0
Bulgaria	4	3.2	68	54.0	54	42.9	0	0.0	57	67.1	28	32.9
Croatia	1	1.2	3	3.7	78	95.1	0	0.0	8	14.6	47	85.5
Cyprus	0	0.0	6	85.7	1	14.3	0	0.0	13	81.3	3	18.8
Czech Republic	7	2.2	15	4.6	302	93.2	18	6.0	81	26.8	203	67.2
Denmark	1	0.5	183	86.7	27	12.8	0	0.0	252	82.6	53	17.4
Finland	0	0.0	0	0.0	26	100	0	0.0	7	22.6	24	77.4
France	0	0.0	2136	79.4	555	20.6	0	0.0	3388	92.0	296	8.0
Georgia	0	0.0	5	12.2	36	87.8						
Germany	5	0.2	213	7.7	2538	92.1	20	0.5	1050	27.5	2746	72.0
Greece	2	0.8	125	52.7	110	46.4	3	0.9	291	82.2	60	17.0
Hungary	1	0.4	42	17.7	194	81.9	4	2.1	31	16.0	159	82.0
Iceland	0	0.0	0	0.0	9	100	0	0.0	4	66.7	2	33.3
Ireland	0	0.0	149	28.4	376	71.6	0	0.0	362	50.4	357	49.7
Israel	2	1.3	18	11.3	139	87.4	12	3.5	92	27.1	236	69.4
Italy	4	0.2	1002	45.5	1198	54.4	15	0.4	2010	56.3	1548	43.3
Latvia	0	0.0	4	12.5	28	87.5	0	0.0	2	14.3	12	85.7
Lithuania	0	0.0	11	55.0	9	45.0	0	0.0	23	100	0	0.0
Luxembourg	0	0.0	1	4.8	20	95.2	0	0.0	1	20.0	4	80.0
Rep of Moldova	0	0.0	3	8.3	33	91.7	0	0.0	2	16.7	10	83.3
The Netherlands	4	0.7	343	63.6	192	35.6	22	2.3	676	69.7	272	28.0
North Macedonia	0	0.0	6	8.1	68	91.9	0	0.0	2	4.1	47	95.9
Norway	1	0.8	55	43.0	72	56.3	8	4.1	44	22.6	143	73.3
Poland	5	0.5	98	10.3	850	89.2	6	1.1	98	18.4	428	80.5
Portugal	1	0.6	84	49.1	86	50.3	2	1.1	115	60.9	72	38.1
Romania	1	0.4	58	22.1	203	77.5	0	0.0	1	4.2	23	95.8
Russian Fed.	33	1.8	368	19.6	1473	78.6	25	4.9	190	37.3	295	57.8
Serbia	0	0.0	0	0.0	136	100	1	1.5	0	0.0	65	98.5
Slovak Republic	1	0.8	36	28.6	89	70.6	1	0.7	107	72.8	39	26.5
Slovenia	1	1.8	0	0.0	54	98.2	0	0.0	8	17.4	38	82.6
Spain	2	0.2	251	23.7	806	76.1	14	1.2	526	43.6	667	55.3
Sweden	17	6.4	11	4.2	237	89.4	7	1.8	94	24.2	288	74.0
Switzerland	3	0.7	66	16.2	338	83.1	9	1.7	203	37.3	332	61.0
Turkey	3	0.1	1670	78.0	468	21.9	1	0.3	236	64.0	132	35.8
Ukraine	3	1.1	17	6.4	245	92.5	1	1.3	2	2.5	76	96.2
United Kingdom	0	0.0	2643	63.9	1495	36.1	0	0.0	3592	62.1	2194	37.9
Total	106	0.5	9768	42.6	13069	57.0	171	0.7	13646	54.5	11229	44.8

Note: Albania, Armenia and Georgia have <5 adults seen in 2022 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, Georgia, Lithuania, the Republic of Moldova, Poland and Romania. In Ukraine it is reimbursed for children, and in Turkey for children ≥ 6 years.

8. Therapies

Table 8.2   Use of inhaled rhDNase ≥3 months in all people with CF seen in 2022 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	65	97.0	2	3.0						
Armenia	0	0.0	13	59.1	9	40.9						
Austria	2	0.5	171	46.1	198	53.4	1	0.3	188	47.1	210	52.6
Belarus	0	0.0	123	84.3	23	15.8	0	0.0	5	100	0	0.0
Bulgaria	0	0.0	32	25.4	94	74.6	0	0.0	18	21.2	67	78.8
Croatia	0	0.0	24	29.3	58	70.7	1	1.8	4	7.3	50	90.9
Cyprus	0	0.0	3	42.9	4	57.1	0	0.0	2	12.5	14	87.5
Czech Republic	7	2.2	175	54.0	142	43.8	18	6.0	55	18.2	229	75.8
Denmark	1	0.5	40	19.0	170	80.6	0	0.0	54	17.7	251	82.3
Finland	0	0.0	5	19.2	21	80.8	0	0.0	6	19.4	25	80.7
France	0	0.0	1559	57.9	1132	42.1	0	0.0	2664	72.3	1020	27.7
Georgia	0	0.0	11	26.8	30	73.2						
Germany	36	1.3	1671	60.6	1049	38.1	44	1.2	2061	54.0	1711	44.8
Greece	2	0.8	49	20.7	186	78.5	4	1.1	124	35.0	226	63.8
Hungary	0	0.0	89	37.6	148	62.5	1	0.5	8	4.1	185	95.4
Iceland	0	0.0	5	55.6	4	44.4	0	0.0	3	50.0	3	50.0
Ireland	0	0.0	305	58.1	220	41.9	0	0.0	284	39.5	435	60.5
Israel	3	1.9	41	25.8	115	72.3	17	5.0	98	28.8	225	66.2
Italy	4	0.2	1216	55.2	984	44.7	15	0.4	2015	56.4	1543	43.2
Latvia	0	0.0	20	62.5	12	37.5	0	0.0	1	7.1	13	92.9
Lithuania	0	0.0	8	40.0	12	60.0	1	4.4	0	0.0	22	95.7
Luxembourg	0	0.0	6	28.6	15	71.4	0	0.0	1	20.0	4	80.0
Rep of Moldova	0	0.0	33	91.7	3	8.3	0	0.0	10	83.3	2	16.7
The Netherlands	3	0.6	199	36.9	337	62.5	5	0.5	424	43.7	541	55.8
North Macedonia	0	0.0	31	41.9	43	58.1	0	0.0	1	2.0	48	98.0
Norway	3	2.3	50	39.1	75	58.6	6	3.1	75	38.5	114	58.5
Poland	4	0.4	178	18.7	771	80.9	5	0.9	20	3.8	507	95.3
Portugal	1	0.6	36	21.1	134	78.4	1	0.5	20	10.6	168	88.9
Romania	2	0.8	49	18.7	211	80.5	0	0.0	1	4.2	23	95.8
Russian Fed.	15	0.8	20	1.1	1839	98.1	23	4.5	63	12.4	424	83.1
Serbia	0	0.0	52	38.2	84	61.8	1	1.5	5	7.6	60	90.9
Slovak Republic	0	0.0	39	31.0	87	69.1	3	2.0	26	17.7	118	80.3
Slovenia	1	1.8	49	89.1	5	9.1	1	2.2	34	73.9	11	23.9
Spain	4	0.4	702	66.3	353	33.3	8	0.7	782	64.8	417	34.6
Sweden	18	6.8	176	66.4	71	26.8	6	1.5	277	71.2	106	27.3
Switzerland	5	1.2	291	71.5	111	27.3	6	1.1	309	56.8	229	42.1
Turkey	4	0.2	355	16.6	1782	83.2	1	0.3	48	13.0	320	86.7
Ukraine	4	1.5	87	32.8	174	65.7	2	2.5	13	16.5	64	81.0
United Kingdom	0	0.0	1489	36.0	2649	64.0	0	0.0	1529	26.4	4257	73.6
Total	119	0.5	9467	41.3	13357	58.2	170	0.7	11232	44.9	13644	54.5

Note: Albania, Armenia and Georgia have <5 adults seen in 2022 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, Belarus, the Republic of Moldova. In Georgia it is reimbursed for people with CF ≥ 2 years; in Bulgaria, Germany, Luxembourg, North Macedonia, Norway, Romania, Spain, Ukraine and the United Kingdom it is reimbursed for people with CF ≥ 5 years; in Latvia it is reimbursed for ≥ 6 years.

8. Therapies

Table 8.3   *Use of inhaled antibiotics ≥3 months in all people with CF seen in 2022 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	1.5	45	67.2	21	31.3						
Armenia	1	4.6	12	54.6	9	40.9						
Austria	1	0.3	286	77.1	84	22.6	2	0.5	215	53.9	182	45.6
Belarus	0	0.0	116	79.5	30	20.6	0	0.0	3	60.0	2	40.0
Bulgaria	0	0.0	86	68.3	40	31.8	0	0.0	36	42.4	49	57.7
Croatia	0	0.0	47	57.3	35	42.7	1	1.8	24	43.6	30	54.6
Cyprus	0	0.0	5	71.4	2	28.6	0	0.0	12	75.0	4	25.0
Czech Republic	8	2.5	296	91.4	20	6.2	19	6.3	190	62.9	93	30.8
Denmark	0	0.0	188	89.1	23	10.9	0	0.0	169	55.4	136	44.6
Finland	0	0.0	25	96.2	1	3.9	0	0.0	19	61.3	12	38.7
France	0	0.0	2140	79.5	551	20.5	0	0.0	2538	68.9	1146	31.1
Georgia	1	2.4	39	95.1	1	2.4						
Germany	29	1.1	2316	84.0	411	14.9	67	1.8	1899	49.8	1850	48.5
Greece	2	0.8	113	47.7	122	51.5	3	0.9	136	38.4	215	60.7
Hungary	0	0.0	146	61.6	91	38.4	1	0.5	77	39.7	116	59.8
Iceland	0	0.0	7	77.8	2	22.2	0	0.0	6	100	0	0.0
Ireland	0	0.0	466	88.8	59	11.2	0	0.0	283	39.4	436	60.6
Israel	5	3.1	107	67.3	47	29.6	19	5.6	149	43.8	172	50.6
Italy	2	0.1	1741	79.0	461	20.9	15	0.4	1994	55.8	1564	43.8
Latvia	1	3.1	24	75.0	7	21.9	0	0.0	5	35.7	9	64.3
Lithuania	0	0.0	19	95.0	1	5.0	2	8.7	17	73.9	4	17.4
Luxembourg	0	0.0	19	90.5	2	9.5	0	0.0	2	40.0	3	60.0
Rep of Moldova	0	0.0	14	38.9	22	61.1	0	0.0	2	16.7	10	83.3
The Netherlands	3	0.6	483	89.6	53	9.8	15	1.6	601	62.0	354	36.5
North Macedonia	0	0.0	33	44.6	41	55.4	0	0.0	8	16.3	41	83.7
Norway	3	2.3	118	92.2	7	5.5	5	2.6	149	76.4	41	21.0
Poland	2	0.2	792	83.1	159	16.7	10	1.9	282	53.0	240	45.1
Portugal	2	1.2	109	63.7	60	35.1	2	1.1	87	46.0	100	52.9
Romania	4	1.5	160	61.1	98	37.4	0	0.0	10	41.7	14	58.3
Russian Fed.	51	2.7	1094	58.4	729	38.9	23	4.5	169	33.1	318	62.4
Serbia	0	0.0	86	63.2	50	36.8	1	1.5	22	33.3	43	65.2
Slovak Republic	1	0.8	73	57.9	52	41.3	2	1.4	53	36.1	92	62.6
Slovenia	1	1.8	49	89.1	5	9.1	1	2.2	40	87.0	5	10.9
Spain	6	0.6	724	68.4	329	31.1	11	0.9	478	39.6	718	59.5
Sweden	17	6.4	212	80.0	36	13.6	6	1.5	315	81.0	68	17.5
Switzerland	5	1.2	373	91.7	29	7.1	5	0.9	345	63.4	194	35.7
Turkey	3	0.1	1790	83.6	348	16.3	1	0.3	242	65.6	126	34.2
Ukraine	3	1.1	159	60.0	103	38.9	3	3.8	21	26.6	55	69.6
United Kingdom	0	0.0	2932	70.9	1206	29.1	0	0.0	2038	35.2	3748	64.8
Total	152	0.7	17444	76.0	5347	23.3	215	0.9	12640	50.5	12191	48.7

Note: Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the table for adults, but the individuals are included in the total number.

Note: Inhaled antibiotics are reimbursed in all countries. In Armenia only Gentamycin and in Romania only Tobramycin solution and Colistin dry powder are reimbursed.

8. Therapies

Table 8.4   Use of inhaled bronchodilators >3 months in children with CF seen in 2022 who have never had a transplant, by country and overall.

Country	Children (<18 years)									
	Missing/ Unknown		No		Yes, long-acting		Yes, short-acting		Yes, both short & long acting	
	N	%	N	%	N	%	N	%	N	%
Albania	0	0.0	8	11.9	57	85.1	2	3.0	0	0.0
Armenia	0	0.0	1	4.6	20	90.9	0	0.0	1	4.6
Austria	3	0.8	26	7.0	265	71.4	69	18.6	8	2.2
Belarus	0	0.0	103	70.6	41	28.1	0	0.0	2	1.4
Bulgaria	0	0.0	114	90.5	9	7.1	3	2.4	0	0.0
Croatia	1	1.2	78	95.1	0	0.0	0	0.0	3	3.7
Cyprus	0	0.0	5	71.4	0	0.0	0	0.0	2	28.6
Czech Republic	129	39.8	94	29.0	22	6.8	67	20.7	12	3.7
Denmark	211	100	0	0.0	0	0.0	0	0.0	0	0.0
Finland	0	0.0	10	38.5	0	0.0	16	61.5	0	0.0
France	0	0.0	1393	51.8	0	0.0	1298	48.2	0	0.0
Georgia	2	4.9	19	46.3	11	26.8	6	14.6	3	7.3
Germany	23	0.8	800	29.0	104	3.8	1561	56.6	268	9.7
Greece	2	0.8	178	75.1	57	24.1	0	0.0	0	0.0
Hungary	3	1.3	170	71.7	7	3.0	46	19.4	11	4.6
Iceland	0	0.0	1	11.1	8	88.9	0	0.0	0	0.0
Ireland	0	0.0	249	47.4	276	52.6	0	0.0	0	0.0
Israel	3	1.9	67	42.1	59	37.1	25	15.7	5	3.1
Italy	3	0.1	716	32.5	213	9.7	1127	51.1	145	6.6
Latvia	0	0.0	2	6.3	24	75.0	6	18.8	0	0.0
Lithuania	0	0.0	9	45.0	9	45.0	0	0.0	2	10.0
Luxembourg	0	0.0	6	28.6	9	42.9	2	9.5	4	19.1
Rep of Moldova	0	0.0	25	69.4	9	25.0	0	0.0	2	5.6
The Netherlands	4	0.7	391	72.5	102	18.9	26	4.8	16	3.0
North Macedonia	0	0.0	2	2.7	69	93.2	0	0.0	3	4.1
Norway	4	3.1	65	50.8	2	1.6	50	39.1	7	5.5
Poland	2	0.2	190	19.9	660	69.3	43	4.5	58	6.1
Portugal	2	1.2	93	54.4	50	29.2	15	8.8	11	6.4
Romania	2	0.8	169	64.5	87	33.2	4	1.5	0	0.0
Russian Fed.	1049	56.0	544	29.0	281	15.0	0	0.0	0	0.0
Serbia	0	0.0	1	0.7	114	83.8	0	0.0	21	15.4
Slovak Republic	1	0.8	57	45.2	46	36.5	12	9.5	10	7.9
Slovenia	1	1.8	51	92.7	1	1.8	2	3.6	0	0.0
Spain	4	0.4	361	34.1	528	49.9	99	9.4	67	6.3
Sweden	19	7.2	12	4.5	1	0.4	214	80.8	19	7.2
Switzerland	2	0.5	150	36.9	228	56.0	15	3.7	12	3.0
Turkey	2	0.1	1385	64.7	377	17.6	365	17.1	12	0.6
Ukraine	4	1.5	143	54.0	89	33.6	27	10.2	2	0.8
United Kingdom	117	2.8	2227	53.8	4	0.1	1784	43.1	6	0.1
Total	1593	6.9	9915	43.2	3839	16.7	6884	30	712	3.1

Note: Inhaled bronchodilators are reimbursed in most countries except in Bulgaria, Georgia, Poland and Serbia.

8. Therapies

Table 8.5   Use of inhaled bronchodilators >3 months in adults with CF seen in 2022 who have never had a transplant, by country and overall.

Country	Adults (≥18 years)									
	Missing/ Unknown		No		Yes, long-acting		Yes, short-acting		Yes, both short & long acting	
	N	%	N	%	N	%	N	%	N	%
Austria	1	0.3	15	3.8	193	48.4	90	22.6	100	25.1
Belarus	0	0.0	3	60.0	1	20.0	0	0.0	1	20.0
Bulgaria	0	0.0	56	65.9	3	3.5	24	28.2	2	2.4
Croatia	0	0.0	17	30.9	15	27.3	22	40.0	1	1.8
Cyprus	0	0.0	9	56.3	0	0.0	2	12.5	5	31.3
Czech Republic	48	15.9	57	18.9	42	13.9	111	36.8	44	14.6
Denmark	305	100	0	0.0	0	0.0	0	0.0	0	0.0
Finland	0	0.0	12	38.7	0	0.0	19	61.3	0	0.0
France	0	0.0	1284	34.9	0	0.0	2400	65.2	0	0.0
Germany	38	1.0	652	17.1	630	16.5	936	24.5	1560	40.9
Greece	3	0.9	165	46.6	185	52.3	0	0.0	1	0.3
Hungary	4	2.1	65	33.5	5	2.6	12	6.2	108	55.7
Iceland	0	0.0	3	50.0	2	33.3	1	16.7	0	0.0
Ireland	0	0.0	121	16.8	598	83.2	0	0.0	0	0.0
Israel	19	5.6	136	40.0	50	14.7	98	28.8	37	10.9
Italy	15	0.4	871	24.4	1295	36.2	853	23.9	539	15.1
Latvia	0	0.0	0	0.0	12	85.7	2	14.3	0	0.0
Lithuania	2	8.7	5	21.7	10	43.5	1	4.4	5	21.7
Luxembourg	0	0.0	1	20.0	0	0.0	2	40.0	2	40.0
Rep of Moldova	0	0.0	7	58.3	3	25.0	0	0.0	2	16.7
The Netherlands	16	1.7	328	33.8	285	29.4	147	15.2	194	20.0
North Macedonia	0	0.0	1	2.0	32	65.3	0	0.0	16	32.7
Norway	9	4.6	22	11.3	10	5.1	99	50.8	55	28.2
Poland	8	1.5	52	9.8	243	45.7	105	19.7	124	23.3
Portugal	2	1.1	62	32.8	24	12.7	84	44.4	17	9.0
Romania	0	0.0	18	75.0	6	25.0	0	0.0	0	0.0
Russian Fed.	315	61.8	61	12.0	134	26.3	0	0.0	0	0.0
Serbia	1	1.5	0	0.0	35	53.0	0	0.0	30	45.5
Slovak Republic	0	0.0	35	23.8	12	8.2	61	41.5	39	26.5
Slovenia	1	2.2	37	80.4	4	8.7	3	6.5	1	2.2
Spain	8	0.7	299	24.8	285	23.6	256	21.2	359	29.7
Sweden	7	1.8	24	6.2	3	0.8	170	43.7	185	47.6
Switzerland	6	1.1	83	15.3	235	43.2	102	18.8	118	21.7
Turkey	2	0.5	159	43.1	108	29.3	97	26.3	3	0.8
Ukraine	5	6.3	17	21.5	33	41.8	17	21.5	7	8.9
United Kingdom	52	0.9	1501	25.9	168	2.9	3970	68.6	95	1.6
Total	867	3.5	6178	24.7	4665	18.6	9685	38.7	3651	14.6

Note: Albania, Armenia and Georgia have <5 adults seen in 2022 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, Poland and Serbia.

8. Therapies

Table 8.6   Use of macrolides ≥3 months in all people with CF seen in 2022 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	1.5	63	94.0	3	4.5						
Armenia	1	4.6	15	68.2	6	27.3						
Austria	1	0.3	365	98.4	5	1.4	1	0.3	380	95.2	18	4.5
Belarus	0	0.0	116	79.5	30	20.6	0	0.0	4	80.0	1	20.0
Bulgaria	3	2.4	121	96.0	2	1.6	0	0.0	85	100	0	0.0
Croatia	0	0.0	46	56.1	36	43.9	0	0.0	19	34.6	36	65.5
Cyprus	0	0.0	4	57.1	3	42.9	0	0.0	7	43.8	9	56.3
Czech Republic	8	2.5	310	95.7	6	1.9	18	6.0	270	89.4	14	4.6
Denmark	0	0.0	206	97.6	5	2.4	0	0.0	227	74.4	78	25.6
Finland	0	0.0	22	84.6	4	15.4	0	0.0	23	74.2	8	25.8
France	0	0.0	2160	80.3	531	19.7	0	0.0	2521	68.4	1163	31.6
Georgia	3	7.3	38	92.7	0	0.0						
Germany	70	2.5	2593	94.1	93	3.4	119	3.1	3130	82.0	567	14.9
Greece	2	0.8	145	61.2	90	38.0	3	0.9	254	71.8	97	27.4
Hungary	0	0.0	181	76.4	56	23.6	2	1.0	154	79.4	38	19.6
Iceland	1	11.1	5	55.6	3	33.3	0	0.0	4	66.7	2	33.3
Ireland	0	0.0	423	80.6	102	19.4	0	0.0	308	42.8	411	57.2
Israel	6	3.8	112	70.4	41	25.8	16	4.7	178	52.4	146	42.9
Italy	2	0.1	1889	85.7	313	14.2	15	0.4	2599	72.7	959	26.8
Latvia	0	0.0	32	100	0	0.0	0	0.0	13	92.9	1	7.1
Lithuania	0	0.0	20	100	0	0.0	1	4.4	21	91.3	1	4.4
Luxembourg	0	0.0	18	85.7	3	14.3	0	0.0	3	60.0	2	40.0
Rep of Moldova	1	2.8	26	72.2	9	25.0	0	0.0	11	91.7	1	8.3
The Netherlands	6	1.1	511	94.8	22	4.1	17	1.8	549	56.6	404	41.7
North Macedonia	0	0.0	68	91.9	6	8.1	0	0.0	30	61.2	19	38.8
Norway	0	0.0	123	96.1	5	3.9	0	0.0	164	84.1	31	15.9
Poland	3	0.3	821	86.2	129	13.5	7	1.3	397	74.6	128	24.1
Portugal	3	1.8	129	75.4	39	22.8	5	2.7	123	65.1	61	32.3
Romania	1	0.4	230	87.8	31	11.8	0	0.0	19	79.2	5	20.8
Russian Fed.	28	1.5	1531	81.7	315	16.8	26	5.1	290	56.9	194	38.0
Serbia	0	0.0	127	93.4	9	6.6	1	1.5	49	74.2	16	24.2
Slovak Republic	2	1.6	82	65.1	42	33.3	1	0.7	75	51.0	71	48.3
Slovenia	1	1.8	54	98.2	0	0.0	2	4.4	39	84.8	5	10.9
Spain	4	0.4	846	79.9	209	19.7	21	1.7	654	54.2	532	44.1
Sweden	18	6.8	214	80.8	33	12.5	6	1.5	290	74.6	93	23.9
Switzerland	3	0.7	389	95.6	15	3.7	10	1.8	416	76.5	118	21.7
Turkey	2	0.1	1977	92.3	162	7.6	1	0.3	311	84.3	57	15.5
Ukraine	5	1.9	120	45.3	140	52.8	3	3.8	13	16.5	63	79.8
United Kingdom	0	0.0	3866	93.4	272	6.6	0	0.0	2922	50.5	2864	49.5
Total	175	0.8	19998	87.2	2770	12.1	275	1.1	16558	66.1	8213	32.8

Note: Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the table for adults, but they are included in the total number.

Note: Oral macrolides are reimbursed in most countries except in Bulgaria, Georgia and Serbia. In the Republic of Moldova, they are reimbursed for children. Inhaled macrolides are reimbursed in Germany, Slovenia, Sweden (for people with CF ≥ 18 years) and the UK.

8. Therapies

Table 8.7 — Use of oxygen ≥3 months in all people with CF seen in 2022 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	66	98.5	1	1.5						
Armenia	0	0.0	22	100	0	0.0						
Austria	0	0.0	366	98.7	5	1.4	2	0.5	384	96.2	13	3.3
Belarus	0	0.0	144	98.6	2	1.4	0	0.0	4	80.0	1	20.0
Bulgaria	0	0.0	125	99.2	1	0.8	1	1.2	82	96.5	2	2.4
Croatia	0	0.0	80	97.6	2	2.4	0	0.0	53	96.4	2	3.6
Cyprus	0	0.0	7	100	0	0.0	0	0.0	15	93.8	1	6.3
Czech Republic	4	1.2	320	98.8	0	0.0	18	6.0	283	93.7	1	0.3
Denmark	0	0.0	210	99.5	1	0.5	0	0.0	303	99.3	2	0.7
Finland	0	0.0	26	100	0	0.0	0	0.0	30	96.8	1	3.2
France	0	0.0	2680	99.6	11	0.4	0	0.0	3601	97.8	83	2.3
Georgia	1	2.4	40	97.6	0	0.0						
Germany	51	1.9	2699	97.9	6	0.2	72	1.9	3473	91.0	271	7.1
Greece	2	0.8	234	98.7	1	0.4	3	0.9	344	97.2	7	2.0
Hungary	0	0.0	229	96.6	8	3.4	1	0.5	151	77.8	42	21.7
Iceland	0	0.0	9	100	0	0.0	0	0.0	6	100	0	0.0
Ireland	0	0.0	524	99.8	1	0.2	0	0.0	673	93.6	46	6.4
Israel	2	1.3	156	98.1	1	0.6	12	3.5	320	94.1	8	2.4
Italy	2	0.1	2197	99.7	5	0.2	15	0.4	3399	95.1	159	4.5
Latvia	0	0.0	32	100	0	0.0	0	0.0	14	100	0	0.0
Lithuania	0	0.0	19	95.0	1	5.0	2	8.7	17	73.9	4	17.4
Luxembourg	0	0.0	21	100	0	0.0	0	0.0	4	80.0	1	20.0
Rep of Moldova	0	0.0	34	94.4	2	5.6	0	0.0	11	91.7	1	8.3
The Netherlands	5	0.9	534	99.1	0	0.0	21	2.2	928	95.7	21	2.2
North Macedonia	0	0.0	74	100	0	0.0	0	0.0	46	93.9	3	6.1
Norway	1	0.8	126	98.4	1	0.8	1	0.5	189	96.9	5	2.6
Poland	4	0.4	941	98.7	8	0.8	7	1.3	493	92.7	32	6.0
Portugal	3	1.8	164	95.9	4	2.3	4	2.1	176	93.1	9	4.8
Romania	4	1.5	255	97.3	3	1.2	0	0.0	23	95.8	1	4.2
Russian Fed.	12	0.6	1827	97.5	35	1.9	22	4.3	445	87.3	43	8.4
Serbia	0	0.0	134	98.5	2	1.5	1	1.5	61	92.4	4	6.1
Slovak Republic	1	0.8	125	99.2	0	0.0	0	0.0	139	94.6	8	5.4
Slovenia	1	1.8	54	98.2	0	0.0	1	2.2	45	97.8	0	0.0
Spain	10	0.9	1044	98.6	5	0.5	9	0.8	1160	96.1	38	3.2
Sweden	18	6.8	243	91.7	4	1.5	6	1.5	378	97.2	5	1.3
Switzerland	6	1.5	400	98.3	1	0.3	14	2.6	519	95.4	11	2.0
Turkey	2	0.1	2088	97.5	51	2.4	1	0.3	338	91.6	30	8.1
Ukraine	4	1.5	252	95.1	9	3.4	2	2.5	71	89.9	6	7.6
United Kingdom	0	0.0	4089	98.8	49	1.2	0	0.0	5537	95.7	249	4.3
Total	133	0.6	22590	98.5	220	1.0	215	0.9	23721	94.7	1110	4.4

Note: Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the table for adults, but they 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 therapy at home is reimbursed.

8. Therapies

Table 8.8   Use of inhaled steroids >3 months in all people with CF seen in 2022 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	51	76.1	16	23.9						
Armenia	1	4.6	17	77.3	4	18.2						
Austria	2	0.5	347	93.5	22	5.9	1	0.3	311	77.9	87	21.8
Belarus	0	0.0	114	78.1	32	21.9	0	0.0	3	60.0	2	40.0
Bulgaria	1	0.8	120	95.2	5	4.0	0	0.0	64	75.3	21	24.7
Croatia	2	2.4	77	93.9	3	3.7	0	0.0	33	60.0	22	40.0
Cyprus	0	0.0	6	85.7	1	14.3	0	0.0	11	68.8	5	31.3
Czech Republic	8	2.5	253	78.1	63	19.4	19	6.3	154	51.0	129	42.7
Denmark	1	0.5	170	80.6	40	19.0	0	0.0	198	64.9	107	35.1
Finland	0	0.0	18	69.2	8	30.8	0	0.0	23	74.2	8	25.8
France	0	0.0	1428	53.1	1263	46.9	0	0.0	1574	42.7	2110	57.3
Georgia	3	7.3	38	92.7	0	0.0						
Germany	67	2.4	2277	82.6	412	15.0	80	2.1	2164	56.7	1572	41.2
Greece	2	0.8	195	82.3	40	16.9	3	0.9	248	70.1	103	29.1
Hungary	1	0.4	211	89.0	25	10.6	4	2.1	167	86.1	23	11.9
Iceland	0	0.0	8	88.9	1	11.1	0	0.0	5	83.3	1	16.7
Ireland	0	0.0	463	88.2	62	11.8	0	0.0	441	61.3	278	38.7
Israel	5	3.1	101	63.5	53	33.3	14	4.1	190	55.9	136	40.0
Italy	3	0.1	1861	84.4	340	15.4	16	0.5	2473	69.2	1084	30.3
Latvia	0	0.0	25	78.1	7	21.9	0	0.0	11	78.6	3	21.4
Lithuania	0	0.0	18	90.0	2	10.0	1	4.4	19	82.6	3	13.0
Luxembourg	0	0.0	14	66.7	7	33.3	1	20.0	1	20.0	3	60.0
Rep of Moldova	0	0.0	31	86.1	5	13.9	0	0.0	8	66.7	4	33.3
The Netherlands	5	0.9	445	82.6	89	16.5	19	2.0	488	50.3	463	47.7
North Macedonia	0	0.0	70	94.6	4	5.4	0	0.0	33	67.4	16	32.7
Norway	2	1.6	117	91.4	9	7.0	3	1.5	144	73.9	48	24.6
Poland	5	0.5	828	86.9	120	12.6	8	1.5	347	65.2	177	33.3
Portugal	0	0.0	139	81.3	32	18.7	3	1.6	132	69.8	54	28.6
Romania	4	1.5	249	95.0	9	3.4	1	4.2	22	91.7	1	4.2
Russian Fed.	17	0.9	1694	90.4	163	8.7	22	4.3	343	67.3	145	28.4
Serbia	0	0.0	108	79.4	28	20.6	1	1.5	33	50.0	32	48.5
Slovak Republic	1	0.8	56	44.4	69	54.8	1	0.7	47	32.0	99	67.4
Slovenia	1	1.8	48	87.3	6	10.9	1	2.2	44	95.7	1	2.2
Spain	6	0.6	758	71.6	295	27.9	13	1.1	593	49.1	601	49.8
Sweden	19	7.2	226	85.3	20	7.6	7	1.8	194	49.9	188	48.3
Switzerland	4	1.0	325	79.9	78	19.2	8	1.5	350	64.3	186	34.2
Turkey	2	0.1	1853	86.6	286	13.4	2	0.5	282	76.4	85	23.0
Ukraine	8	3.0	232	87.6	25	9.4	2	2.5	62	78.5	15	19.0
United Kingdom	0	0.0	3519	85.0	619	15.0	0	0.0	4617	79.8	1169	20.2
Total	170	0.7	18510	80.7	4263	18.6	230	0.9	15835	63.2	8981	35.9

Note: Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the table for adults, but they are included in the total number.

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

8. Therapies

Table 8.9   Use of oral steroids ≥3 months in all people with CF seen in 2022 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	1.5	66	98.5	0	0.0						
Armenia	1	4.6	21	95.5	0	0.0						
Austria	0	0.0	369	99.5	2	0.5	1	0.3	387	97.0	11	2.8
Belarus	0	0.0	143	98.0	3	2.1	0	0.0	4	80.0	1	20.0
Bulgaria	1	0.8	125	99.2	0	0.0	0	0.0	83	97.7	2	2.4
Croatia	0	0.0	82	100	0	0.0	0	0.0	55	100	0	0.0
Cyprus	0	0.0	7	100	0	0.0	0	0.0	16	100	0	0.0
Czech Republic	8	2.5	315	97.2	1	0.3	19	6.3	279	92.4	4	1.3
Denmark	1	0.5	209	99.1	1	0.5	0	0.0	285	93.4	20	6.6
Finland	0	0.0	26	100	0	0.0	0	0.0	31	100	0	0.0
France	0	0.0	2667	99.1	24	0.9	0	0.0	3565	96.8	119	3.2
Georgia	2	4.9	39	95.1	0	0.0						
Germany	70	2.5	2657	96.4	29	1.1	140	3.7	3474	91.0	202	5.3
Greece	3	1.3	230	97.1	4	1.7	3	0.9	346	97.7	5	1.4
Hungary	1	0.4	232	97.9	4	1.7	4	2.1	184	94.9	6	3.1
Iceland	0	0.0	9	100	0	0.0	0	0.0	6	100	0	0.0
Ireland	0	0.0	518	98.7	7	1.3	0	0.0	684	95.1	35	4.9
Israel	3	1.9	156	98.1	0	0.0	11	3.2	314	92.4	15	4.4
Italy	4	0.2	1985	90.1	215	9.8	16	0.5	2688	75.2	869	24.3
Latvia	0	0.0	32	100	0	0.0	0	0.0	14	100	0	0.0
Lithuania	0	0.0	20	100	0	0.0	1	4.4	22	95.7	0	0.0
Luxembourg	0	0.0	21	100	0	0.0	0	0.0	5	100	0	0.0
Rep of Moldova	1	2.8	35	97.2	0	0.0	0	0.0	12	100	0	0.0
The Netherlands	5	0.9	526	97.6	8	1.5	19	2.0	898	92.6	53	5.5
North Macedonia	0	0.0	74	100	0	0.0	0	0.0	46	93.9	3	6.1
Norway	2	1.6	125	97.7	1	0.8	4	2.1	189	96.9	2	1.0
Poland	3	0.3	945	99.2	5	0.5	7	1.3	507	95.3	18	3.4
Portugal	1	0.6	165	96.5	5	2.9	2	1.1	185	97.9	2	1.1
Romania	3	1.2	258	98.5	1	0.4	1	4.2	23	95.8	0	0.0
Russian Fed.	12	0.6	1825	97.4	37	2.0	22	4.3	465	91.2	23	4.5
Serbia	0	0.0	135	99.3	1	0.7	1	1.5	64	97.0	1	1.5
Slovak Republic	2	1.6	121	96.0	3	2.4	0	0.0	132	89.8	15	10.2
Slovenia	1	1.8	54	98.2	0	0.0	1	2.2	44	95.7	1	2.2
Spain	8	0.8	1037	97.9	14	1.3	15	1.2	1143	94.7	49	4.1
Sweden	18	6.8	247	93.2	0	0.0	7	1.8	370	95.1	12	3.1
Switzerland	3	0.7	403	99.0	1	0.3	8	1.5	516	94.9	20	3.7
Turkey	2	0.1	2122	99.1	17	0.8	2	0.5	360	97.6	7	1.9
Ukraine	8	3.0	255	96.2	2	0.8	2	2.5	74	93.7	3	3.8
United Kingdom	0	0.0	4066	98.3	72	1.7	0	0.0	5266	91.0	520	9.0
Total	164	0.7	22322	97.3	457	2.0	286	1.1	22742	90.8	2018	8.1

Note: Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the table for adults, but they are included in the total number.

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

8. Therapies

Table 8.10   Prevalence of the use of ursodeoxycholic acid for ≥ 3 months in all people with CF seen in 2022 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	31	46.3	36	53.7						
Armenia	0	0.0	17	77.3	5	22.7						
Austria	0	0.0	190	51.2	181	48.8	0	0.0	206	51.6	193	48.4
Belarus	0	0.0	24	16.4	122	83.6	0	0.0	0	0.0	5	100
Bulgaria	0	0.0	49	38.9	77	61.1	1	1.2	51	60.0	33	38.8
Croatia	0	0.0	57	69.5	25	30.5	2	3.6	32	58.2	21	38.2
Cyprus	0	0.0	7	100	0	0.0	0	0.0	14	87.5	2	12.5
Czech Republic	6	1.9	222	68.5	96	29.6	19	6.3	197	65.2	86	28.5
Denmark	1	0.5	162	76.8	48	22.8	0	0.0	201	65.9	104	34.1
Finland	0	0.0	19	73.1	7	26.9	0	0.0	21	67.7	10	32.3
France	0	0.0	2274	84.5	417	15.5	0	0.0	2787	75.7	897	24.4
Georgia	1	2.4	34	82.9	6	14.6						
Germany	4	0.2	1680	61.0	1072	38.9	14	0.4	1778	46.6	2024	53.0
Greece	2	0.8	166	70.0	69	29.1	3	0.9	237	67.0	114	32.2
Hungary	0	0.0	146	61.6	91	38.4	3	1.6	100	51.6	91	46.9
Iceland	0	0.0	9	100	0	0.0	0	0.0	5	83.3	1	16.7
Ireland	0	0.0	512	97.5	13	2.5	0	0.0	643	89.4	76	10.6
Israel	4	2.5	135	84.9	20	12.6	14	4.1	275	80.9	51	15.0
Italy	2	0.1	1669	75.7	533	24.2	15	0.4	2185	61.2	1373	38.4
Latvia	1	3.1	24	75.0	7	21.9	0	0.0	9	64.3	5	35.7
Lithuania	0	0.0	14	70.0	6	30.0	1	4.4	20	87.0	2	8.7
Luxembourg	0	0.0	19	90.5	2	9.5	0	0.0	3	60.0	2	40.0
Rep of Moldova	0	0.0	19	52.8	17	47.2	0	0.0	7	58.3	5	41.7
The Netherlands	6	1.1	439	81.5	94	17.4	28	2.9	709	73.1	233	24.0
North Macedonia	0	0.0	49	66.2	25	33.8	0	0.0	15	30.6	34	69.4
Norway	2	1.6	115	89.8	11	8.6	1	0.5	181	92.8	13	6.7
Poland	5	0.5	475	49.8	473	49.6	7	1.3	164	30.8	361	67.9
Portugal	2	1.2	115	67.3	54	31.6	5	2.7	134	70.9	50	26.5
Romania	4	1.5	165	63.0	93	35.5	0	0.0	14	58.3	10	41.7
Russian Fed.	11	0.6	136	7.3	1727	92.2	22	4.3	126	24.7	362	71.0
Serbia	0	0.0	107	78.7	29	21.3	1	1.5	36	54.6	29	43.9
Slovak Republic	4	3.2	60	47.6	62	49.2	2	1.4	62	42.2	83	56.5
Slovenia	0	0.0	28	50.9	27	49.1	1	2.2	23	50.0	22	47.8
Spain	15	1.4	799	75.5	245	23.1	31	2.6	882	73.1	294	24.4
Sweden	18	6.8	205	77.4	42	15.9	7	1.8	312	80.2	70	18.0
Switzerland	3	0.7	324	79.6	80	19.7	6	1.1	383	70.4	155	28.5
Turkey	3	0.1	1783	83.3	355	16.6	1	0.3	301	81.6	67	18.2
Ukraine	2	0.8	22	8.3	241	90.9	1	1.3	7	8.9	71	89.9
United Kingdom	0	0.0	3320	80.2	818	19.8	0	0.0	4366	75.5	1420	24.5
Total	96	0.4	15621	68.1	7226	31.5	186	0.7	16490	65.8	8370	33.4

Note: Albania, Armenia and Georgia have <5 adults seen in 2022 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 for children and 70% for adults.

8. Therapies

Table 8.11   Prevalence of the use of proton pump inhibitors (PPI) for ≥ 3 months in all people with CF seen in 2022 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	52	77.6	15	22.4						
Armenia	0	0.0	8	36.4	14	63.6						
Austria	3	0.8	356	96.0	12	3.2	1	0.3	329	82.5	69	17.3
Belarus	0	0.0	119	81.5	27	18.5	0	0.0	2	40.0	3	60.0
Bulgaria	4	3.2	99	78.6	23	18.3	3	3.5	55	64.7	27	31.8
Croatia	0	0.0	71	86.6	11	13.4	0	0.0	48	87.3	7	12.7
Cyprus	0	0.0	7	100	0	0.0	0	0.0	11	68.8	5	31.3
Czech Republic	8	2.5	300	92.6	16	4.9	19	6.3	191	63.3	92	30.5
Denmark	1	0.5	157	74.4	53	25.1	0	0.0	163	53.4	142	46.6
Finland	0	0.0	16	61.5	10	38.5	0	0.0	28	90.3	3	9.7
France	0	0.0	2091	77.7	600	22.3	0	0.0	2135	58.0	1549	42.1
Georgia	7	17.1	34	82.9	0	0.0						
Germany	5	0.2	2486	90.2	265	9.6	13	0.3	2922	76.6	881	23.1
Greece	2	0.8	206	86.9	29	12.2	5	1.4	306	86.4	43	12.2
Hungary	237	100	0	0.0	0	0.0	194	100	0	0.0	0	0.0
Iceland	0	0.0	3	33.3	6	66.7	0	0.0	5	83.3	1	16.7
Ireland	0	0.0	415	79.1	110	21.0	0	0.0	314	43.7	405	56.3
Israel	4	2.5	105	66.0	50	31.5	14	4.1	215	63.2	111	32.7
Italy	2	0.1	1888	85.7	314	14.3	16	0.5	2275	63.7	1282	35.9
Latvia	0	0.0	25	78.1	7	21.9	0	0.0	13	92.9	1	7.1
Lithuania	1	5.0	17	85.0	2	10.0	2	8.7	16	69.6	5	21.7
Luxembourg	0	0.0	18	85.7	3	14.3	0	0.0	2	40.0	3	60.0
Rep of Moldova	0	0.0	27	75.0	9	25.0	0	0.0	11	91.7	1	8.3
The Netherlands	5	0.9	425	78.9	109	20.2	24	2.5	625	64.4	321	33.1
North Macedonia	0	0.0	50	67.6	24	32.4	0	0.0	10	20.4	39	79.6
Norway	1	0.8	116	90.6	11	8.6	1	0.5	146	74.9	48	24.6
Poland	4	0.4	878	92.1	71	7.5	10	1.9	383	72.0	139	26.1
Portugal	0	0.0	152	88.9	19	11.1	4	2.1	118	62.4	67	35.5
Romania	8	3.1	239	91.2	15	5.7	0	0.0	24	100	0	0.0
Russian Fed.	31	1.7	1492	79.6	351	18.7	26	5.1	289	56.7	195	38.2
Serbia	0	0.0	108	79.4	28	20.6	1	1.5	39	59.1	26	39.4
Slovak Republic	4	3.2	115	91.3	7	5.6	4	2.7	114	77.6	29	19.7
Slovenia	0	0.0	46	83.6	9	16.4	1	2.2	28	60.9	17	37.0
Spain	3	0.3	900	85.0	156	14.7	20	1.7	662	54.9	525	43.5
Sweden	19	7.2	211	79.6	35	13.2	12	3.1	283	72.8	94	24.2
Switzerland	3	0.7	375	92.1	29	7.1	10	1.8	389	71.5	145	26.7
Turkey	3	0.1	1915	89.4	223	10.4	1	0.3	307	83.2	61	16.5
Ukraine	5	1.9	202	76.2	58	21.9	7	8.9	41	51.9	31	39.2
United Kingdom	0	0.0	2690	65.0	1448	35.0	0	0.0	2455	42.4	3331	57.6
Total	360	1.6	18414	80.3	4169	18.2	388	1.6	14958	59.7	9700	38.7

Note: Albania, Armenia and Georgia have <5 adults seen in 2022 and are excluded from the table for adults but are included in the total number.

Note: Oral proton pump inhibitors are reimbursed in most countries except in Bulgaria, Georgia, Lithuania, the Republic of Moldova and Serbia.

Appendix 2 List of contributing centres and national registries

List of individual centres and national registries that contributed data to the ECFSPR in 2022.
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 2022 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
Austria	14 individual centres:	Andreas Pfleger
	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	Adrienne Molnar Claudia Altmann
	Kepler Universitätsklinikum, Klinik für Lungenheilkunde/ Pneumologie, Linz	Katrin Scheich Viktoria Reinelt
	Kardinal Schwarzenberg Klinikum, Abteilung für Kinder- und Jugendmedizin, Schwarzach im Pongau	Josef Riedler Christoph Seelbach
	Salzburger Landeskliniken, Universitätsklinik für Pneumologie, Salzburg	Michael Studnicka Natalie Firlei-Fleischmann
	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

Country	Centre/National Registry name	Contact
Belarus	1 individual centre: Belarusian Republic Children's Centre of Pulmonology and Cystic Fibrosis, Pulmonary Department, 3rd City Children's Clinical Hospital, Minsk	Sviatlana Keegan Vladimir Bobrovnichiy Sviatlana Keegan Katsiaryna Chyrkun
Bulgaria	2 individual centres: Alexandrovska University Hospital, Pediatric 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 Andreas Matthaiou 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
Finland	Cystic Fibrosis in Finland	Varpu Elenius Katriina Pihlajamaa Aleksi Kemppainen
France	Registre Français de la Mucoviscidose	Antoine Bessou Clémence Dehillotte
Germany	German Cystic Fibrosis Registry	Lutz Naehrlich Julia Wosniok
Georgia	1 individual centre: LTD, Medical Genetics and Laboratory Diagnostic Centre, Tblisi	Dodo Agladze Dodo Agladze Ia Khurtsilava
Greece	Cystic Fibrosis Registry of Greece	Elpis Hatziagorou John Tsanakas Panagiota Mitrou Kostas Mathioudakis Anastasios Tsolakidis
Hungary	Cystic Fibrosis Registry of Hungary	Andrea Párniczky Géza Marsal
Iceland	1 individual centre: Children´s Medical Center 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 Center, 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 Center of Israel, Petah Tikva, Israel; Faculty of Medical and Health Sciences, Tel Aviv University, Tel Aviv,	Meir Mei-Zahav
	Safra Children's Hospital, Sheba Medical Centre, Ramat Gan	Ori Efrati
Italy	Italian Cystic Fibrosis Registry	Rita Padoan
		Marco Salvatore
		Annalisa Amato
		Gianluca Ferrari
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	Stojka Fustik
		Andriana Andeevska
Rep. of Moldova	1 individual centre:	Oxana Turcu
	Outpatient Centre for Cystic Fibrosis and Other Rare Diseases, Chisinau	Oxana Turcu
Netherlands	Dutch Cystic Fibrosis Registry	Domenique Zomer
		Renate Kos
Norway	Norwegian Cystic Fibrosis Patient Registry	Egil Bakkeheim
		Anita C.S. Wathne
Poland	18 individual centres:	Łukasz Woźniacki
	Voivodeship Children's Hospital, Dept. of Paediatric Pneumology and Allergology, Bydgoszcz	Radoslawa Staszak – Kowalska
		Mikolaj Kowalski
		Anna Olszewska
	Cystic Fibrosis Centre, Polanki Paediatric Hospital, Gdansk	Maria Trawinska-Bartnicka
		Ewa Sapiejka
	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

Country	Centre/National Registry name	Contact
	Paediatric Clinic Holy Cross Paediatric Centre Provincial Integrated Hospital in Kielce, Kielce	*Elzbieta Kolodziej* *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
	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 Agata Nowicka
	Karol Jonscher University Hospital of Poznan University of Medical Sciences, Poznan	Irena Wojsyk-Banaszak
	Institute of Tuberculosis and Lung Diseases, Rabka-Zdrój Branch, Dept. of Pneumology and Cystic Fibrosis, Rabka Zdroj	Henryk Mazurek 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	7 individual centres: Regional Cystic Fibrosis Centre, Clinical Emergency Children's Hospital of Brasov, Brasov	Liviu Pop Laura Larisa Dracea
	Clinical Children's Hospital "Grigore Alexandrescu", Bucharest	Simona Mosescu Livia Brezeanu
	Emergency Hospital for Children Marie Curie" – Paediatrics 3, Bucharest	Maria Iulia Brustan Ioana Gradinaru
	Mother & Child Health Institute, Bucharest	Iustina Stan Valentina Comanici
	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 Timisoara - County Emergency Clinical Hospital "Pius Branzeu" Timisoara	Liviu Pop Ioana Ciuca
Russian Federation	Cystic Fibrosis Registry of the Russian Federation	Elena Kondratyeva Elena Amelina Marina Starinova Stanislav Krasovskiy Anna Voronkova Nataliya Kashirskaya

Country	Centre/National Registry name	Contact
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: Childrens CF Centre, DFN Banská Bystrica, Banská Bystrica Centrum Cystickej Fibrozy pre dospelych FNSP FDR, Banská Bystrica Centrum Cystickej Fibrozy pre dospelych, Klinika Pneumologie l.SZU a Univerzitna Nemocnica, Bratislava Klinika detskej Pneumologie SZU UN Bratislava, pracovisko Podunajské Biskupice, Bratislava CF Adult centre, University Hospital L Pasteura, Košice Centrum cystickej fibrozy detí, Detská fakultná nemocnica Košice, Košice	Hana Kayserova Branko Takáč Ivana Gondová Eva Bérešova Marta Hajkova Hana Kayserova Nina Bližňáková Lenka Kopčová Anna Fetekeova Zuzana Hribíková
Slovenia	3 individual centres: University Clinic of Pulmonary and Allergic Diseases, Golnik University Medical Centre Ljubljana, Department of Pulmonology and Allergy, Ljubljana University Medical Centre Ljubljana, University Children`s Hospital, Department of Paediatric Pulmonology, Ljubljana	Uroš Krivec Matjaž Fležar Julij Šelb Barbara Salobir 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 Hospital Sant Joan de Déu, Unitat de Pneumologia Pediàtrica i Fibrosi Quística, Barcelona Hospital Universitari Vall d'Hebron, Unidad de Fibrosis Quística del Adulto, Barcelona Hospital Universitari Vall d'Hebron, Unidad Fibrosis Quística y Neumología Pediátrica, Barcelona Hospital Universitario Cruces, Unidad de Fibrosis Quística, Bizkaia 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 Complejo Hospitalario Universitario Insular Materno Infantil, Las Palmas de Gran Canaria Hospital Universitario La Paz, Unidad de Fibrosis Quística Adultos, Servicio de Neumología, Madrid Hospital Universitario La Paz, Sección de Neumología Pediátrica, Unidad de Fibrosis Quística Pediátrica, Madrid Hospital Universitario La Princesa, Neumología Adultos, Madrid	M. Dolores Pastor Vivero Oscar Asensio de la Cruz Miguel Garcia Gonzàlez Xavier Pomares Amigó Concepción Montón Soler Maria Cols i Roig Jordi Costa i Colomer Antonio Alvarez Fernández Eva Polverino Silvia Gartner Sandra Rovira Amigo M. Dolores Pastor Vivero Ainhoa Gómez Bonilla Beatriz Gómez Crespo Javier Torres Borrego José Manuel Vaquero Barrios Antonio José Aguilar Fernández Concha Prados Marta Ruiz de Valbuena Maiz Cristina de Manuel Gómez Rosa María Girón Rosa Mar Gómez-Punter

Country	Centre/National Registry name	Contact
Spain (cont.)	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
	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: Kinderspital Aarau, Kantonsspital Aarau AG, Abteilung pädiatrische Pneumologie, Allergologie und Immunologie, Aarau	Andreas Jung Dominik Müller-Suter Peter Eng Rachel Kusche
	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 Michael Tamm

Country	Centre/National Registry name	Contact
Switzerland (cont.)	UKBB Universitäts-Kinderspital beider Basel, Abteilung Intensivmedizin & Pneumologie, Basel	Daniel Trachsel Anja Jochmann Diana Reppucci Jakob Usemann
	Inselspital Bern, Universitätsklinik für Pneumologie, Adulte Cystische Fibrose, Bern	Dagmar Lin Thomas Geiser Michaela Semmler
	Lindenhofspital Quartier Bleu, Bern	Bernhard Schwizer Reta Fischer Iris Schmid
	Universitätsklinik für Kinderheilkunde, Zentrum für Cystische Fibrose und Pulmonologie, Inselspital, Bern	Philipp Latzin Carmen Casaulta Romy Rodriguez
	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 Zisis Balmpouzis Georgia Mitropoulou Isabelle Huart Bellavere
	Luzerner Kantonsspital, Zentrum für Zystische Fibrose für Kinder und Jugendliche, Luzern	Nicolas Regamey Michael Hitzler Marco Lurà 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	Sidikka Ozturk-Beungies Alain Sauty Jean Marc Fellrath
	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 Otto Schoch Rebekka Kleiner
	Kantonsspital Winterthur, Klinik für Pneumologie und Klinik für Innere Medizin, Adulte Cystische Fibrose, Winterthur	Markus Hofer Sieghart Filippi

Country	Centre/National Registry name	Contact
Switzerland (cont.)	Universitäts-Kinderspital Zürich, Abteilung für Pneumologie, Zürich	Andreas Jung Alexander Möller Demet Inci Eugènie Collaud
	Universitätsspital Zürich, Klinik für Pneumologie, Adultes CF Zentrum, Zürich	Macé Schuurmans Carolin Steinack Dominik Damm Christian Clarenbach Maurice Roeder Thomas Kurowski
Turkey	Cystic Fibrosis Registry of Turkey	Deniz Dogru
Turkey	Cystic Fibrosis Registry of Turkey, Ankara	Deniz Dogru
	Marmara University Faculty of Medicine, Division of Pediatric Pulmonology, Istanbul	Bülent Karadağ Yasemin Gökdemir Ela Erdem Eralp
	Medipol University Faculty of Medicine, Division of Pediatric Pulmonology, Istanbul	Füsun Ünal
	Medeniyet University, Faculty of Medicine, Division of Pediatric Pulmonology, Istanbul	Saniye Girit Zeynep Reyhan Onay
Ukraine	14 individual centres:	Halyna Makukh
Ukraine	Dnipro Children's Clinical Hospital, 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*
	Volyn Regional Childrens Hospital, Paediatric Department; 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*
	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*

Country	Centre/National Registry name	Contact
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

Patient 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 patient's 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.

Data of people without a CF diagnosis according to the agreed definitions are accepted in the database but not included in the analyses.

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 is anonymous, the ECFS collects only year and month of birth and the day of birth was 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.
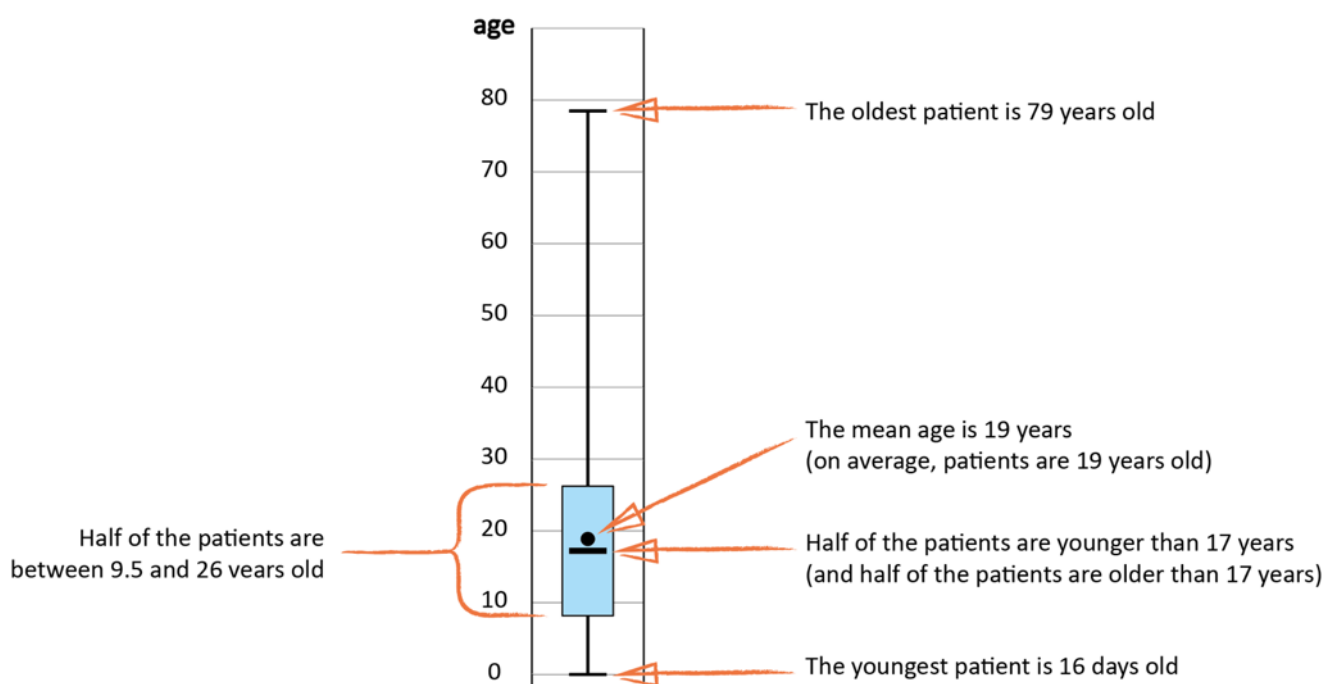
Note: This is an example of how to read a boxplot. The numbers used in this figure are not real.

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

Originally removed from the variables collected from 2022 onwards but reinstated in January 2023 at start of the 2022 data collection period (as requested by various members of the ECFSPR Steering Committee.

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 (previously Staphylococcus aureus, no specification)
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

Note: Changes valid from 2022 are in turquoise.

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 millimols 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 can be 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 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** <u>**ATS/ERS guidelines.**</u>

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.

* Direct Data Entry Hospitals (not National Registries): if a patient is transferred to a different hospital for transplant and that hospital submits data to the ECFSPR, the transplant centre must not re-register the patient. The Core Data will be transferred through the data collection software to the transplant centre.

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 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: a range of CFTR modulators have been approved for use. They are 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. Since the CFTR modulator therapies work specifically for certain variant classes, those currently available are effective only in people with those variants.

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 get CF you need to have two CFTR variants, one on each allele in chromosome 7 (where the CFTR-gene is located). If both variants are 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 at the same allele it is called a complex allele.

DIOS: distal intestinal obstruction syndrome is a condition, unique to people with CF. In DIOS, the intestines are blocked by thickened stool due to sticky mucus and other mechanisms, which leads to reduced stool flow through the intestines and abdominal pain and can result in an emergency.

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 are two different variants, 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 a strong anti-inflammatory property. The types that are prescribed to people with CF are the group known as 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
ES   Spain
FI   Finland
FR   France
GE   Georgia
GR   Greece
HR   Croatia
HU   Hungary
IE   Ireland
IL   Israel
IS   Iceland
IT   Italy
LT   Lithuania
LU   Luxembourg
LV   Latvia
MD   Republic of Moldova
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   Turkey
UA   Ukraine
UK   United Kingdom of Great Britain and Northern Ireland

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