2019

ECFS Patient Registry
Annual Data Report

European Cystic Fibrosis Society
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Annual Data Report

2019 data

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Preface

We are pleased to share with you the 2019 Annual Report from the European Cystic Fibrosis Society Patient Registry (ECFSPR). This 15th report contains demographic and clinical data of 50,902 consenting CF patients from 38 countries. The epidemiological data is provided by national cystic fibrosis (CF) registries and individual CF centres throughout Europe and neighbouring countries.

It is the ECFSPR's mission to provide a reliable and comprehensive picture of clinical outcomes in CF across Europe that will help to enhance quality of CF care in Europe and worldwide. The number of countries and centres that contribute data to the ECFSPR continues to grow and one of our aims is to achieve complete European-wide coverage. Our strong collaboration with CF Europe and national patient organisations guarantees that the ECFSPR data is used to benefit the community and we are grateful to all people with CF, and their families, throughout Europe, for their willingness to participate in the Registry.

The raw data submitted by participating countries is analysed by the ECFSPR statisticians and the outcomes are presented in this report. The results of analyses for some countries presented in the ECSFPR report may differ from the data published in their national annual registry report; these differences can originate from variations in patient inclusion criteria, the definitions used for disease complications and the employment of different reference values. Further details can be found in the report and in the List of ECFSPR Variables and Definitions in Appendix 3 (page 156). Over the next few years we will introduce changes in the design and content of the report.

The ECFSPR collaborates closely with CF centres and national CF registries to ensure that their data is as complete and high quality as possible. Since the European Medicines Agency (EMA) qualified the ECFSPR and its data collection software, ECFSTracker, as a platform and data source for post-authorisation safety (PASS) and efficacy (PAES) studies, we have begun conducting our first large pharmacovigilance studies. The results of these studies will provide important real world evidence for novel therapies such as CFTR modulators.

New in the ECFSPR is the collection of data from people with CF infected with SARS-CoV-2 throughout the pandemic, beginning early in 2020. Two manuscripts have been published on the outcomes of COVID-19 in CF (for more details visit page 145).

The countries and national registries that participate in the ECFSPR, represented by their country coordinators, patients, and their families, have dedicated their valuable time and resources to the Registry. They are supported by the patient organisations, the ECFSPR staff, the Executive and Scientific Committees and a vast number of voluntary working group members of the many projects undertaken by the ECFSPR. I would like to thank all these stakeholders for their huge efforts in making the Registry an important and invaluable database with regard to CF care. Finally, I would like to thank our sponsors and supporters who provide the financial background to make the ECFSPR possible.

Sincerely,

Andreas Jung, 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.

With each ECFSPR Annual Report we publish a separate At-a-Glance report containing key information from the report, specifically for the people with CF and their families, and anyone wishing to know a little more about the disease: www.ecfs.eu/projects/ecfs-patient-registry/annual-reports. Interactive maps with country-specific information are available on the homepage of our website: www.ecfs.eu/ecfspr.

We continue to develop country posters with information and basic statistics from the Registry for display in CF centres. The posters are published online at www.ecfs.eu/ecfspr/posters. The data in the posters will be regularly updated.

News, updates and other interesting information are regularly posted on social media. Find us on Facebook www.facebook.com/EuropeanCysticFibrosisPatientRegistry/ and Twitter @ECFSRegistry.

, we will continue to work with patient organisationson increasing 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 presented in this report we encourage you to contact your CF centre.

For more detailed information about the Registry please visit the patient-dedicated page on our website www.ecfs.eu/projects/ecfs-patient-registry/information-about-ecfspr-cf-patients. More information on how we handle your data and how you can exercise your rights is available in the Privacy Notice www.ecfs.eu/sites/default/files/general-content-files/working-groups/ecfs-patient-registry/Privacy%20notice_Update_ECFSPR_vs%205_0.pdf.

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 ECFSPR 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 www.ecfs.eu/ecfspr.

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 the presence of chronic *Pseudomonas aeruginosa* infection according to the modified Leeds criteria, and/or the presence of elevated *Pseudomonas* antibodies (see Appendix 3 on page 156). 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, instead, 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 patients. The countries with less than 5 patients 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 valuefor 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 20% have unknown/missing information on liver disease, 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 patient 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. Since clinical outcomes do not change very much over a 12-month period, we do not consider this to be a serious obstacle to interpretation. 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 patient. We therefore needed to transform the raw values into new variables in order to compare lung function between people with CF and 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, page 11. These differences can influence how the data is interpreted, and we therefore advise comparisons to be made only between countries with similar coverage.

Date of the database that is used to create the tables and graphs in this report is 20 July 2021.

Glossary and Abbreviations

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
FR:	France
GB:	United Kingdom
GE:	Georgia
GR:	Greece
HR:	Croatia
HU:	Hungary
IE:	Ireland
IL:	Israel
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

Explanation of terms:

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 CFboth CFTR genes must be affected by a CF-causing mutation.

CFTR modulator therapy: a range of CFTR modulators have been approved for use. They are designed to correct the malfunctioning CFTR protein: different mutations 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 mutation classes, those currently available are effective only in people with those mutations.

Compassionate use: is a treatment option that allows the use of an unauthorised medicine for patients who have no alternative treatment options and no access to clinical trials.

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 here is major bleeding (more than 250 ml).

Homozygous: CF is caused by mutations of the CFTR gene, one on each allele. One is inherited from the mother and one from the father. If both mutations are the same, the person is said to be homozygous for this mutation.

Heterozygous: CF is caused by mutations of the CFTR gene, one on each allele. One is inherited from the mother and one from the father. If these are two different mutations, 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 mutations). 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 (LCI). 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.

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 patients are diagnosed when they are 3 years old.

Meconium ileus: small-bowel obstruction caused by unusual thick, sticky faeces (i.e. meconium, which is the first stool of newborn babies).

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 patients are diagnosed before 5 months of age, and the other half of the patients are diagnosed after 5 months of age.

Min: minimum. It is the lowest value.

N: the number of patients in a group for whom the information is not missing.

N miss: number of missing values. It is the number of patients for whom the information is missing.

NaCl: sodium chloride. Here: inhaled hypertonic saline.

NIPPV: Non-invasive positive pressure ventilation; this refers to mechanical ventilation that helps patients 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).

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 patients 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 patients are diagnosed before they were 3 years old, and the remaining quarter was diagnosed after they reached 3 years of age.

Pneumothorax: collapsed lung. In CF usually because of severe lung damage.

PPI: Proton Pump Inhibitors, is medication that reduces stomach acid levels.

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.

rhDNase: recombinant human DNase (marketed as Pulmozyme®).

Z-score (or standardised scores): a way to compare results with a "normal" population, the reference population (see Appendix 2 for details). 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.

Summary of data report

Outcome		Females	Males	Total
Patients registered in the ECFSPR	n	24199	26703	50902
	(%)	(47.5%)	(52.5%)	
Age at follow-up (in years; patients alive on 31/12/2019)	mean	21.0	21.7	21.4
	median	18.5	19.5	19.0
Patients ≥ 18 years (patients alive on 31/12/2019)	n	12247	14200	26447
	(%)	(51.1%)	(53.6%)	(52.4%)
Age at diagnosis*	mean (years)	4.2	4.0	4.1
	median (months)	3.7	3.6	3.7
Patients with at least one F508del allele recorded*	n	18834	20736	39570
	(%)	(80.7%)	(80.8%)	(80.8%)
Patients living with lung transplant**	n	1440	1405	2845
	(%)	(6.2%)	(5.5%)	(5.8%)
Patients living with liver transplant**	n	101	196	297
	(%)	(0.4%)	(0.8%)	(0.6%)
Patients deceased in 2019***	n	212	202	414
	(%)	(0.9%)	(0.8%)	(0.8%)
Age at death (years)***	mean	28.7	34.2	31.4
	median	28.0	33.0	30.0

* Only patients seen during the year are presented. The total number of patients presented is 49,340.

** Only patients alive at 31/12/2019 are presented. The total number of patients presented is 48,978.

*** Only patients seen during the year are presented. For the United Kingdom, all patients with a confirmed diagnosis of CF were included (N=10,665). The total number of patients presented is 49,925.

Data report

1. Demographics

Figure 1.1 Map of countries that contributed to the ECFSPR in year 2019.

Marked in blue are the countries that contributed 2019 data.

Table 1.1 Number of patients in year 2019, by country.

Country	Patients registered, not lost to follow-up	Patients seen	Estimated coverage
Albania	133	106	80%
Armenia	30	25	>70%
Austria	833	816	>90%
Belarus	155	155	90%
Belgium*	1360	1294	>90%
Bulgaria	197	188	>70%
Croatia**	146	139	>95%
Cyprus	31	28	>80%
Czech Republic*	654	643	99%
Denmark*	530	514	99%
France*	6729	6729	>90%
Georgia[1]	80	72	>80%
Germany*	6481	6481	80%
Greece[2]	451	436	>70%
Hungary*	528	509	90%
Ireland*	1268	1251	90%
Israel**	588	529	>95%
Italy*	5618	5585	95%
Latvia	47	45	>90%
Lithuania	32	28	52%
Luxembourg	40	38	>95%
Rep of Moldova	59	53	>90%
The Netherlands*	1533	1501	95%
North Macedonia	141	131	>90%
Norway*	319	312	90%
Poland	1242	1194	>60%
Portugal**	368	347	>95%
Romania	246	230	50%
Russian Federation*	3433	3162	>95%
Serbia	202	183	>90%
Slovak Republic**	326	287	>90%
Slovenia	113	113	>95%
Spain	2417	2329	80%
Sweden*	704	704	95%
Switzerland**	999	974	>95%
Turkey	2002	1988	>60%
Ukraine	212	151	23%
United Kingdom[3]*	10655	10070	99%
Total	50902	49340	

* 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.

1 Georgia has 0% coverage for adults.

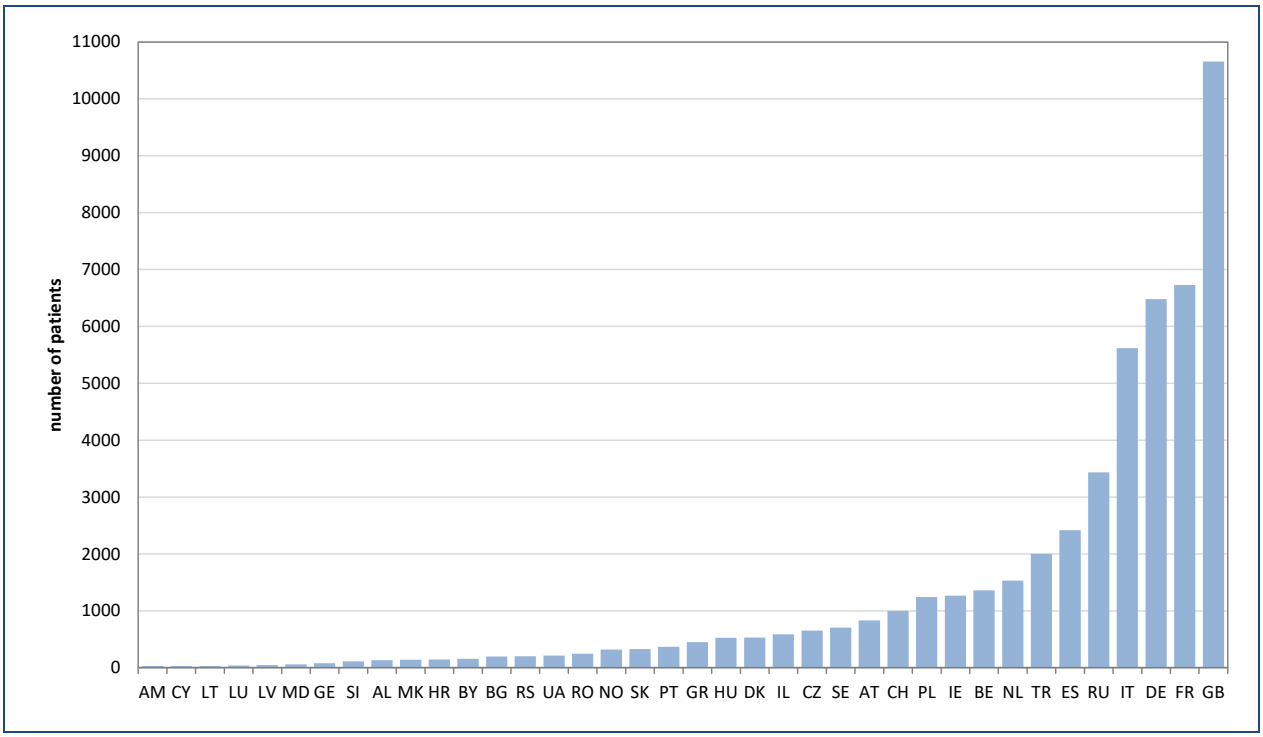
2 Greece: one centre didn't provide data for follow-up year 2019.

3 United Kingdom: in the tables and figures of this report we use GB as abbreviation for United Kingdom, reference www.iban.com/country-codes.

The column “Patients registered, not lost to follow-up” shows the patients that attend centres and includes patients that were not seen by clinical staff during the year but are known to be alive that year. NB: The ECFSPR recommends marking a patient as “lost to follow-up” if a patient has not been seen for the third consecutive year and nothing is known about his/her condition.

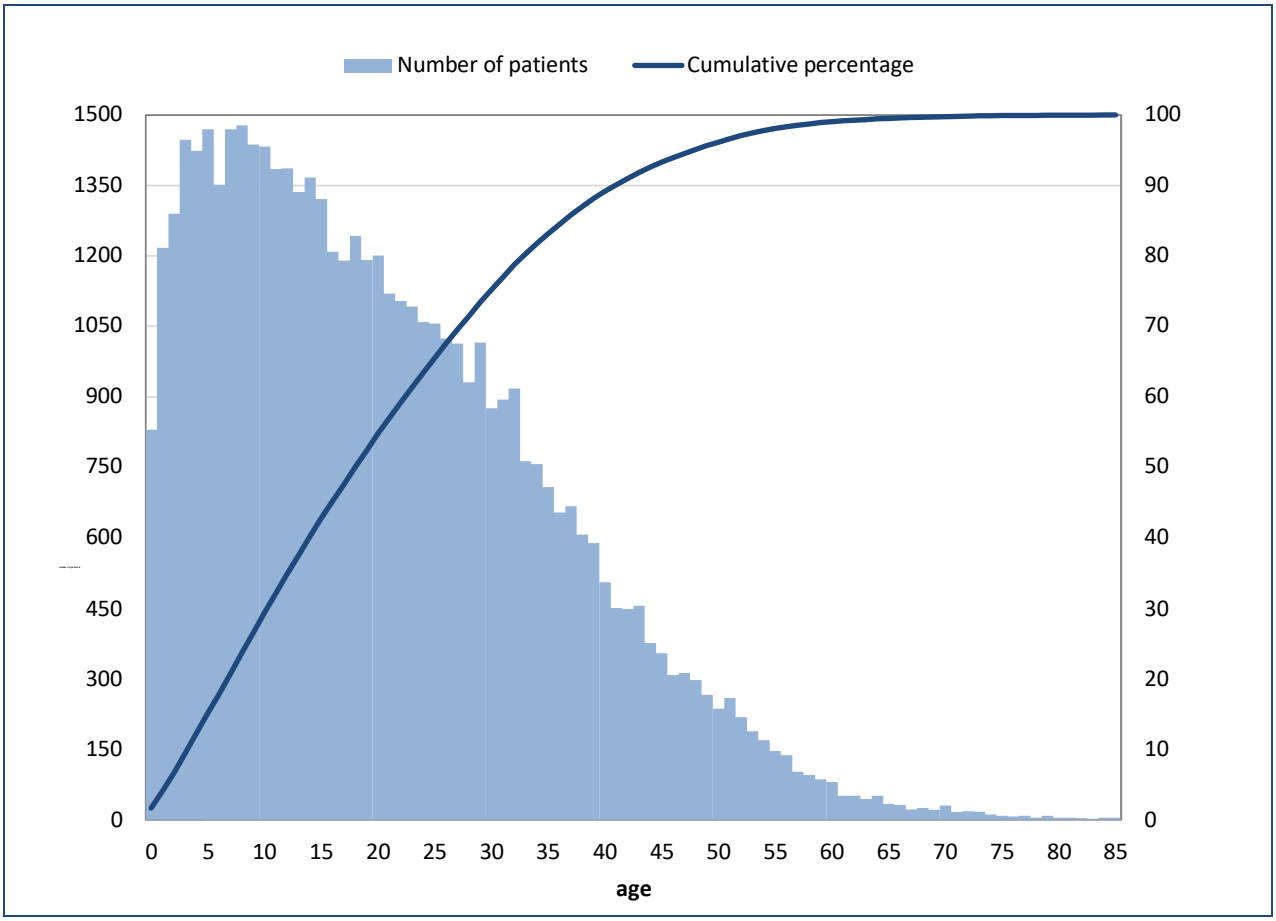
The column “patients seen” presents only the patients who have attended the clinic during the year. The column “Estimated coverage 2019” shows the estimated percentage of people with CF living in that country who are included in the national registry/national data collection as reported by the country. For some countries, one individual centre may include almost all patients, e.g. Latvia and Serbia.

Figure 1.2 Number of patients registered in the ECFSPR in year 2019, by country.



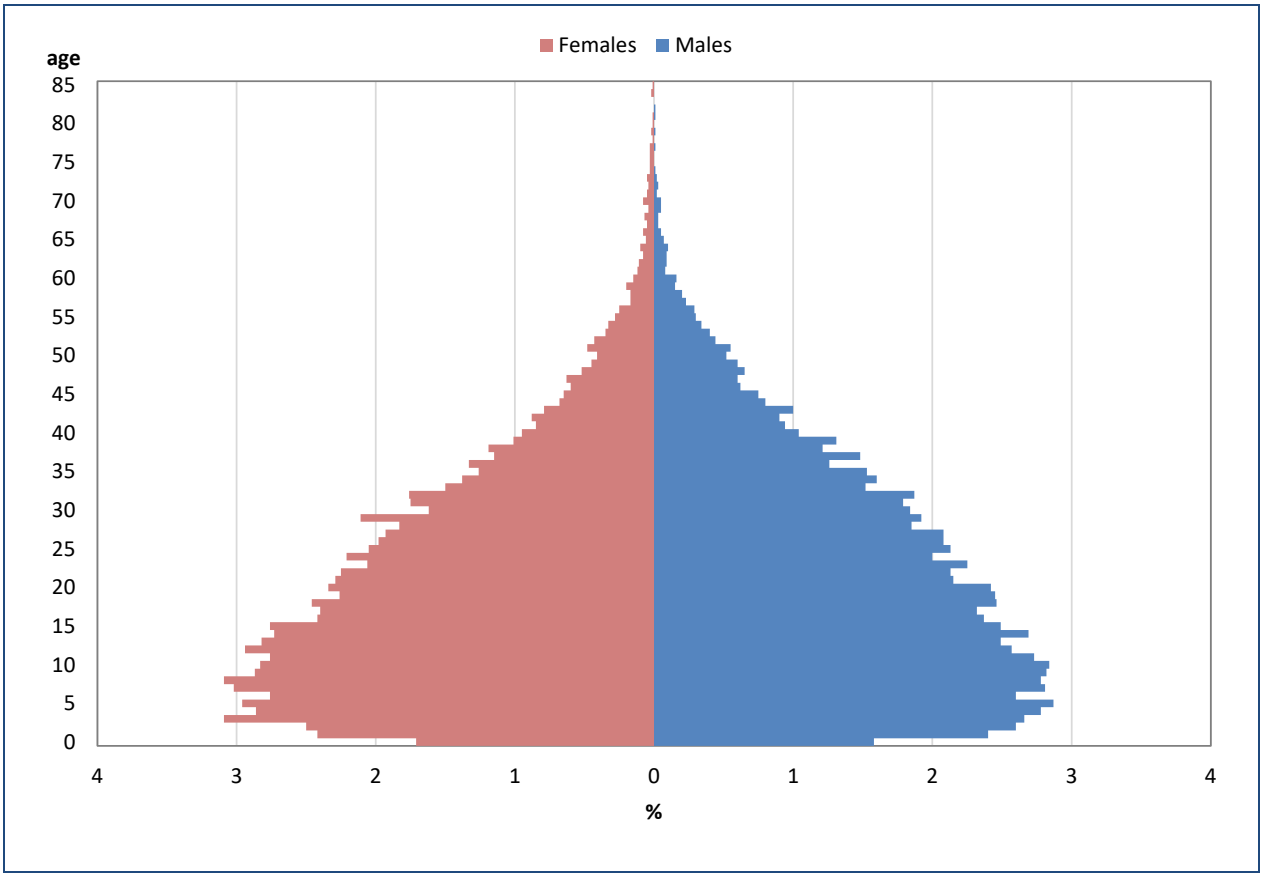
Each vertical bar shows the number of registered patients (excluding lost to follow-up) living in that country in 2019. Please refer to table 1.1 for the coverage in each country.

Figure 1.3 Age at follow-up distribution. Patients alive on 31/12/2019.



Each blue vertical bar represents the number of patients of that age alive in 2019. The cumulative percentage (the dark blue line) describes how many patients (as a percentage) are below a certain age (e.g. 50% of the patients are younger than 19 years of age).

Figure 1.4 Age at follow-up distribution by sex. Patients alive on 31/12/2019.



The pyramid shows the percentage of patients of different ages as horizontal bars. The right side of the pyramid (blue) shows, for males, how many patients (as a percentage) are a certain age, the left side (red) shows the same for females. The lower percentage of patients at the bottom of the pyramid is a result of the fact that some patients have not yet been diagnosed (mean age at diagnosis is 4.1 years, see table 2.1).

Table 1.2 Proportion of children (<18 years) and adults (≥18 years), by country. Patients alive on 31/12/2019.

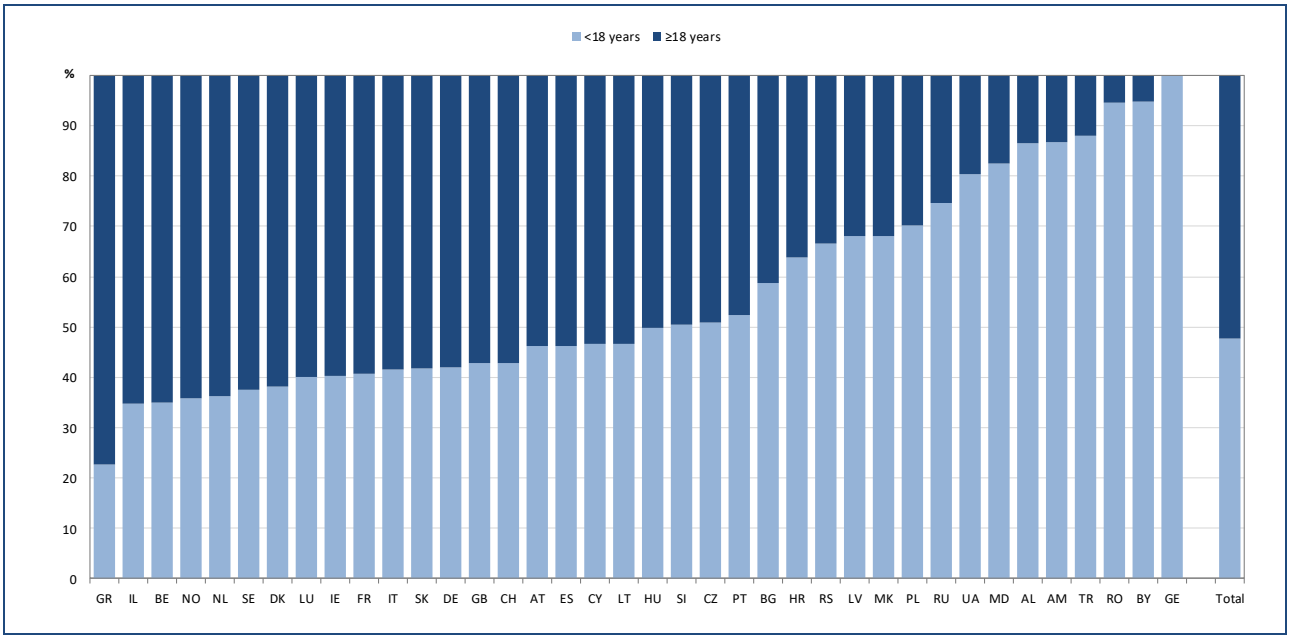
Country	Children (<18 years) number (%)	Adults (≥18 years) number (%)
Albania	115 (86.47)	18 (13.53)
Armenia	26 (86.67)	4 (13.33)
Austria	383 (46.26)	445 (53.74)
Belarus	147 (94.84)	8 (5.16)
Belgium	473 (34.93)	881 (65.07)
Bulgaria	114 (58.76)	80 (41.24)
Croatia	92 (63.89)	52 (36.11)
Cyprus	14 (46.67)	16 (53.33)
Czech Republic	327 (50.86)	316 (49.14)
Denmark	200 (38.17)	324 (61.83)
France	2731 (40.82)	3959 (59.18)
Georgia	79 (100)	0 (0)
Germany	2697 (41.92)	3737 (58.08)
Greece	101 (22.60)	346 (77.40)
Hungary	259 (49.81)	261 (50.19)
Ireland	508 (40.38)	750 (59.62)
Israel	204 (34.87)	381 (65.13)
Italy	2325 (41.64)	3258 (58.36)
Latvia	32 (68.09)	15 (31.91)
Lithuania	14 (46.67)	16 (53.33)
Luxembourg	16 (40.00)	24 (60.00)
Rep of Moldova	47 (82.46)	10 (17.54)
The Netherlands	554 (36.28)	973 (63.72)
North Macedonia	96 (68.09)	45 (31.91)

Note: Georgia has 0% coverage for adults.
Greece: The data of one of the paediatric centres is not included, which accounts for the high percentage of adult patients.

[table 1.2 continued]

Country	Children (<18 years) number (%)	Adults (≥18 years) number (%)
Norway	114 (35.85)	204 (64.15)
Poland	863 (70.11)	368 (29.89)
Portugal	191 (52.47)	173 (47.53)
Romania	229 (94.63)	13 (5.37)
Russian Federation	2526 (74.71)	855 (25.29)
Serbia	134 (66.67)	67 (33.33)
Slovak Republic	134 (41.74)	187 (58.26)
Slovenia	57 (50.44)	56 (49.56)
Spain	1112 (46.26)	1292 (53.74)
Sweden	264 (37.61)	438 (62.39)
Switzerland	427 (42.91)	568 (57.09)
Turkey	1752 (87.95)	240 (12.05)
Ukraine	169 (80.48)	41 (19.52)
United Kingdom	4515 (42.83)	6026 (57.17)
Total	24041 (47.62)	26447 (52.38)

Figure 1.5 Proportion of children (<18 years) and adults (≥18 years), by country and overall. Patients alive on 31/12/2019.



Note: Georgia has 0% coverage for adults.
Greece: The data of one of the paediatric centres is not included, which accounts for the high percentage of adult patients.

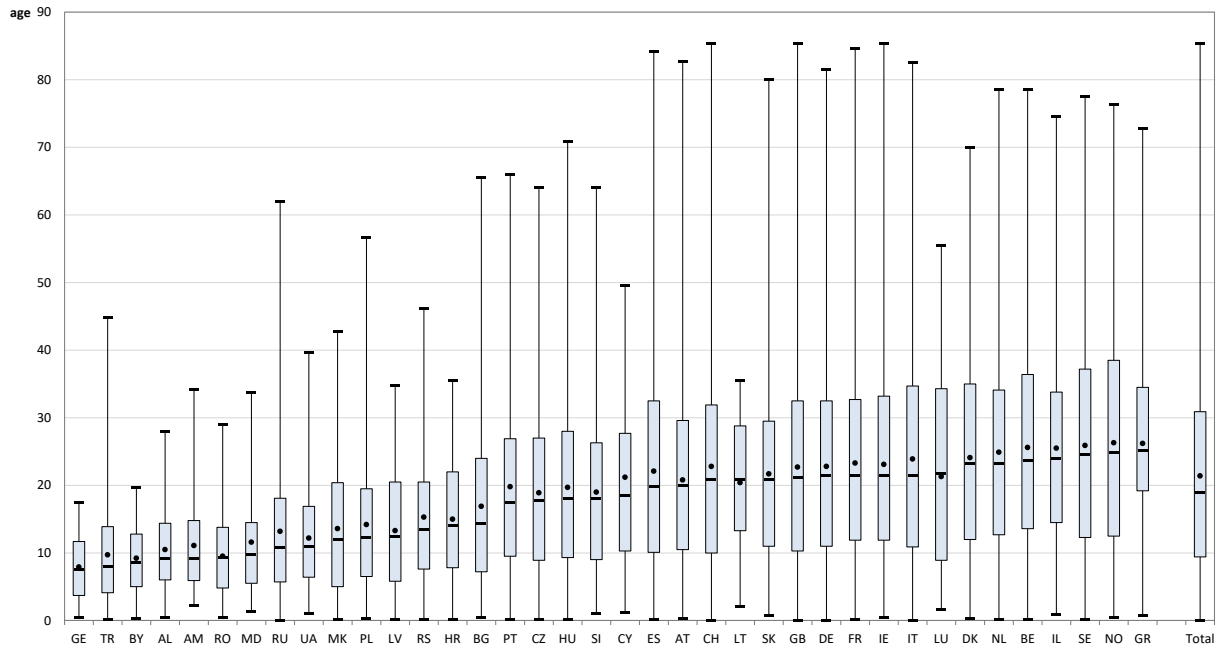
This graph shows the percentage of patients in each country who are adults (dark blue) or children (light blue). The percentage of adults varies considerably between the different countries, but this is partly an effect of the way the patients are included. For example, for some countries only a few individual centres send data to the ECFSPR, or the proportion of children and adults may reflect only the proportion of paediatric and adult centres in that country who participate in the ECFSPR and not the real proportion in the country. Please refer to table 1.1, page 12, for national coverage.

Table 1.3 Age at follow-up: descriptive statistics, by country and overall. Patients alive on 31/12/2019.

Country	N	Mean (average age)	Min (age of the youngest patient)	25th pctl (25% of the patients are younger than this age)	Median (half the patients are younger than this age)	75th pctl (75% of the patients are younger than this age)	Max (age of the oldest patient)
Albania	133	10.5	0.5	6.0	9.2	14.4	28.0
Armenia	30	11.1	2.2	5.9	9.2	14.8	34.2
Austria	828	20.8	0.3	10.5	20.0	29.6	82.7
Belarus	155	9.2	0.3	5.0	8.5	12.8	19.7
Belgium	1354	25.6	0.1	13.6	23.7	36.4	78.5
Bulgaria	194	16.9	0.5	7.2	14.4	24.0	65.5
Croatia	144	15.0	0.2	7.8	14.0	22.0	35.5
Cyprus	30	21.2	1.2	10.3	18.5	27.7	49.5
Czech Republic	643	18.9	0.1	8.9	17.7	27.0	64.0
Denmark	524	24.1	0.3	12.0	23.3	35.0	70.0
France	6690	23.3	0.1	11.9	21.5	32.7	84.6
Georgia	79	7.9	0.5	3.7	7.6	11.7	17.4
Germany	6434	22.8	0.0	11.0	21.4	32.5	81.5
Greece	447	26.2	0.8	19.2	25.2	34.5	72.8
Hungary	520	19.7	0.1	9.3	18.0	28.0	70.9
Ireland	1258	23.1	0.4	11.9	21.5	33.2	85.4
Israel	585	25.5	0.9	14.5	24.0	33.8	74.5
Italy	5583	23.9	0.0	10.9	21.5	34.7	82.5
Latvia	47	13.3	0.2	5.8	12.4	20.5	34.7
Lithuania	30	20.4	2.0	13.3	20.9	28.8	35.5
Luxembourg	40	21.3	1.6	8.9	21.7	34.3	55.5
Rep of Moldova	57	11.6	1.3	5.5	9.8	14.5	33.7
The Netherlands	1527	24.9	0.1	12.7	23.3	34.1	78.5
North Macedonia	141	13.6	0.1	5.0	12.0	20.4	42.8
Norway	318	26.3	0.4	12.5	24.8	38.5	76.3
Poland	1231	14.2	0.3	6.5	12.3	19.5	56.7
Portugal	364	19.8	0.2	9.5	17.5	26.9	66.0
Romania	242	9.5	0.4	4.8	9.3	13.8	29.0
Russian Federation	3381	13.2	0.0	5.7	10.8	18.1	62.0
Serbia	201	15.3	0.1	7.6	13.5	20.5	46.1
Slovak Republic	321	21.7	0.7	11.0	20.9	29.5	80.0
Slovenia	113	19.0	1.0	9.0	18.0	26.3	64.1
Spain	2404	22.1	0.1	10.1	19.8	32.5	84.2
Sweden	702	25.9	0.2	12.3	24.5	37.2	77.5
Switzerland	995	22.8	0.0	10.0	20.9	31.9	85.3
Turkey	1992	9.7	0.1	4.1	8.0	13.9	44.8
Ukraine	210	12.2	1.0	6.4	10.9	16.9	39.7
United Kingdom	10541	22.7	0.0	10.3	21.2	32.5	85.4
Total	50488	21.4	0.0	9.4	19.0	30.9	85.4

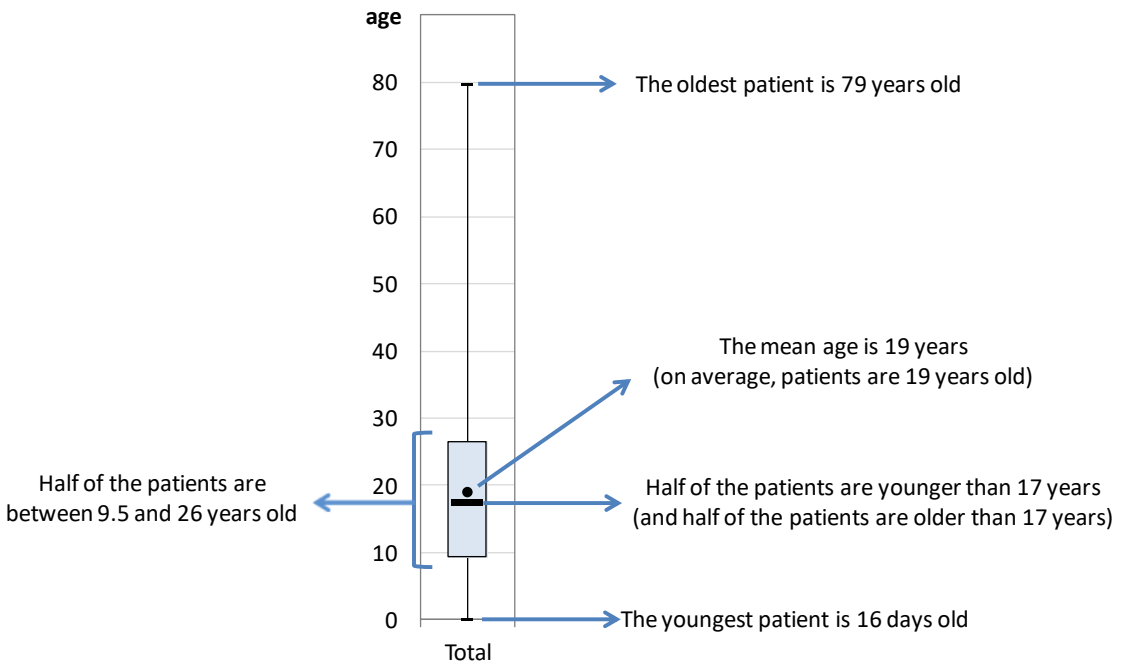
This table shows the descriptive statistics for age at follow-up of the patients by country and overall. Only patients who were alive on 31/12/2019 are included.

Figure 1.6 Age at follow-up: box-plot, by country and overall. Patients alive on 31/12/2019.



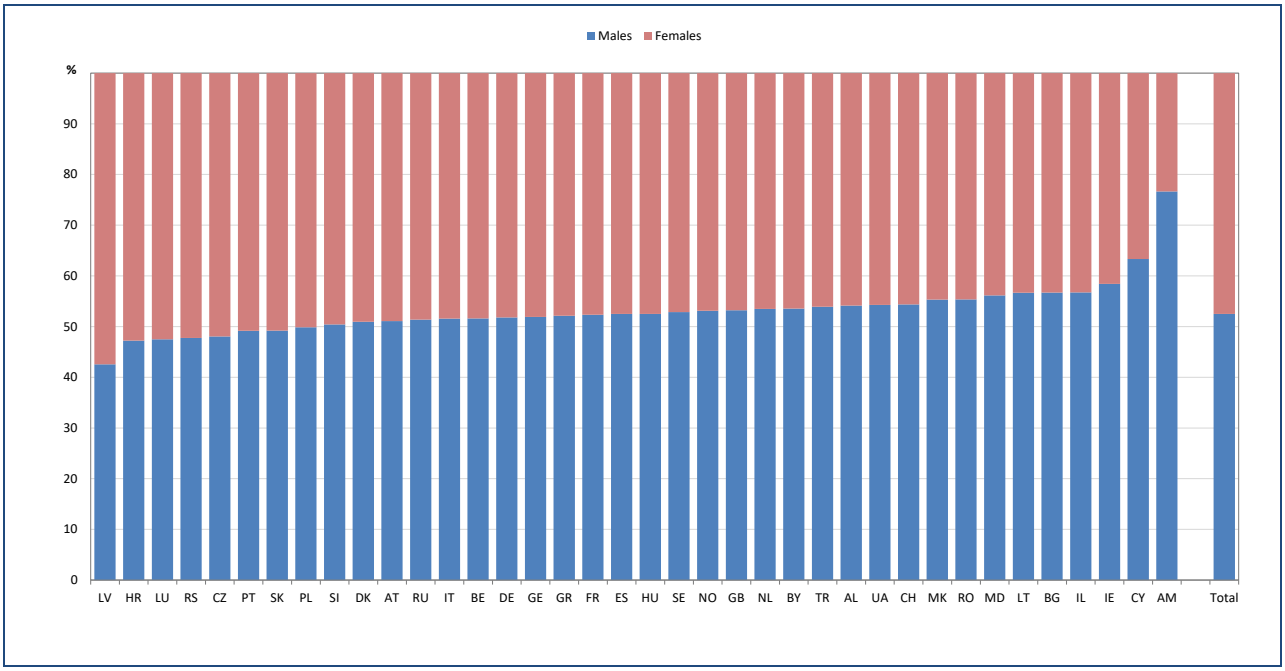
This box-plot is a graphic representation of the age detailed in table 1.3. For each country the dash (black line crossing the blue box) is the median, the black dot is the mean and the whiskers (vertical lines with a T-shaped end) are the minimum and the maximum.

The following figure explains how to read the box-plot.



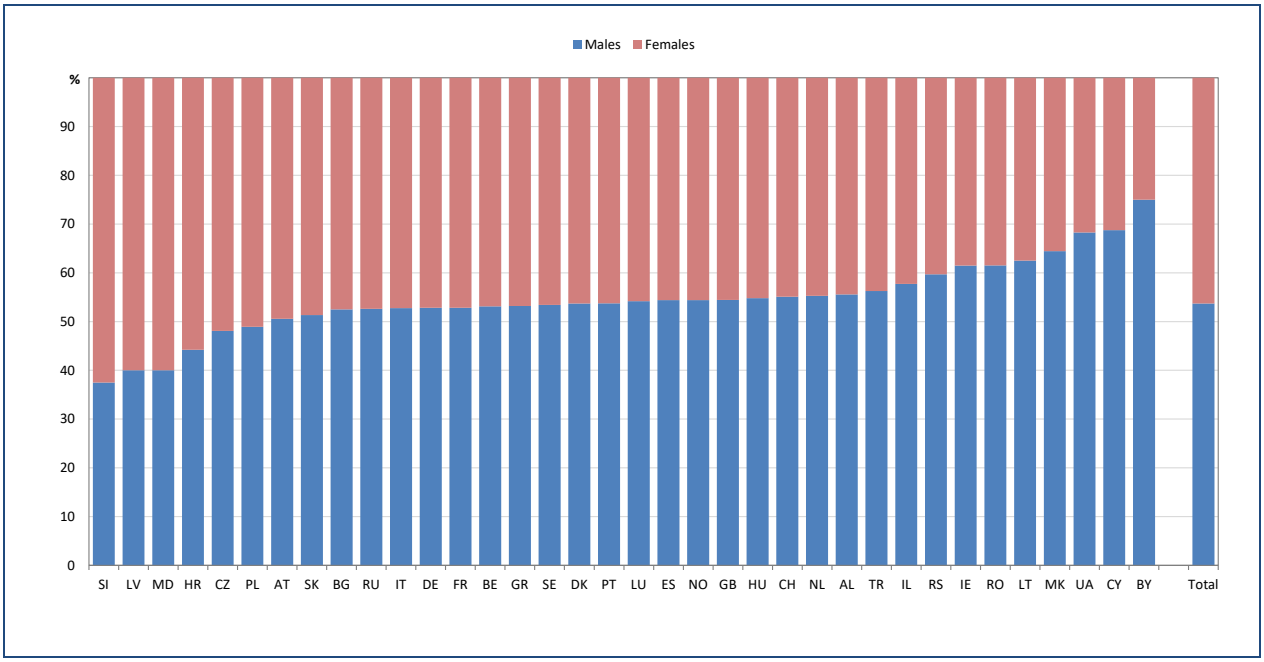
Note: This is an example of how to read a box-plot. The numbers used in this figure are not real and do not refer to figure 1.6.

Figure 1.7 Sex distribution, by country and overall. Patients alive on 31/12/2019.



Sex distribution of all patients. Overall (see "Total") in the ECFSPR there are slightly more male than female patients.

Figure 1.8 Sex distribution, by country and overall. Patients alive on 31/12/2019 and aged 18 years or more.



Note: Georgia has 0% coverage for adults and is excluded from the graph.
Armenia has <5 patients aged 18 years or more and is excluded from the graph.

Sex distribution for adults. The total proportion of females in the adult group is similar to the proportion of females in the total ECFSPR population (fig 1.7).

2. Diagnosis

Hereafter, only patients seen during the year are presented.

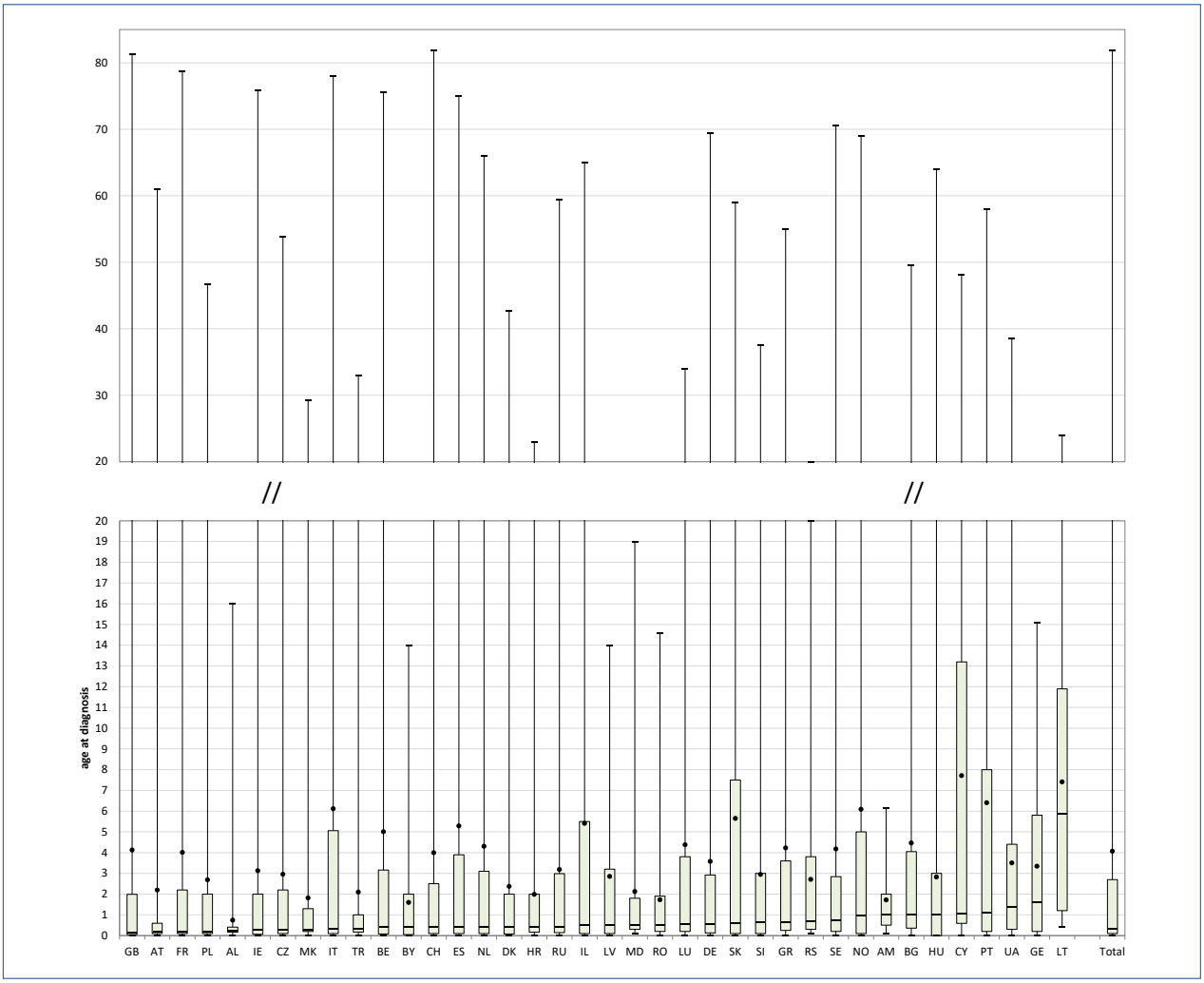
Table 2.1 Age at diagnosis (in years): descriptive statistics, by country and overall. All patients seen in 2019.

Country	N	N miss	Mean (average age at diagnosis)	Min (lowest age at diagnosis)	25th pctl (25 % of the patients were diagnosed before this age)	Median (half the patients were diagnosed before this age)	75th pctl (75% of the patients were diagnosed before this age)	Max (highest age at diagnosis)
Albania	106	0	0.75	0.00	0.16	0.25	0.41	16.00
Armenia	25	0	1.72	0.10	0.50	1.00	2.00	6.17
Austria	751	65	2.20	0.00	0.10	0.20	0.60	61.00
Belarus	155	0	1.60	0.01	0.05	0.40	2.00	14.00
Belgium	1293	1	5.01	0.00	0.08	0.40	3.16	75.53
Bulgaria	184	4	4.47	0.00	0.36	1.00	4.05	49.60
Croatia	131	8	1.99	0.00	0.16	0.42	2.00	23.00
Cyprus	26	2	7.72	0.00	0.59	1.05	13.20	48.09
Czech Republic	635	8	2.96	0.00	0.10	0.30	2.20	53.90
Denmark	514	0	2.37	0.00	0.08	0.42	2.00	42.67
France	6657	72	4.01	0.00	0.10	0.20	2.20	78.70
Georgia	70	2	3.35	0.00	0.20	1.60	5.80	15.10
Germany	6238	243	3.58	0.00	0.13	0.58	2.92	69.41
Greece	424	12	4.23	0.00	0.25	0.67	3.60	55.00
Hungary	453	56	2.83	0.00	0.00	1.00	3.00	64.00
Ireland	1246	5	3.13	0.00	0.06	0.27	2.00	75.83
Israel	524	5	5.41	0.00	0.10	0.50	5.50	65.00
Italy	5471	114	6.12	0.00	0.10	0.31	5.06	77.95
Latvia	45	0	2.86	0.00	0.10	0.50	3.20	14.00
Lithuania	27	1	7.41	0.40	1.20	5.90	11.90	24.00
Luxembourg	38	0	4.38	0.00	0.20	0.55	3.80	34.00
Rep of Moldova	53	0	2.13	0.10	0.30	0.50	1.80	19.00
The Netherlands	1438	63	4.31	0.00	0.10	0.40	3.10	66.00
North Macedonia	131	0	1.82	0.00	0.20	0.30	1.30	29.20
Norway	302	10	6.09	0.00	0.10	0.95	5.00	69.00
Poland	1191	3	2.69	0.00	0.10	0.20	2.00	46.70
Portugal	336	11	6.41	0.00	0.20	1.10	8.00	58.00
Romania	222	8	1.73	0.00	0.20	0.50	1.90	14.60
Russian Federation	3141	21	3.19	0.00	0.14	0.42	2.99	59.46
Serbia	180	3	2.71	0.10	0.30	0.70	3.80	20.00
Slovak Republic	258	29	5.65	0.00	0.10	0.59	7.50	59.00
Slovenia	111	2	2.95	0.00	0.10	0.63	3.00	37.50
Spain	2299	30	5.29	0.00	0.10	0.40	3.90	75.00
Sweden	692	12	4.18	0.00	0.20	0.74	2.84	70.57
Switzerland	864	110	4.00	0.00	0.10	0.40	2.50	81.90
Turkey	1978	10	2.10	0.00	0.17	0.33	1.00	33.00
Ukraine	150	1	3.51	0.00	0.30	1.40	4.40	38.50
United Kingdom	10052	18	4.13	0.00	0.05	0.16	1.99	81.35
Total	48411	929	4.07	0.00	0.10	0.31	2.70	81.90

Note: For Hungary, Slovak Republic and Switzerland the information on age at diagnosis is missing for more than 10% of the patients.

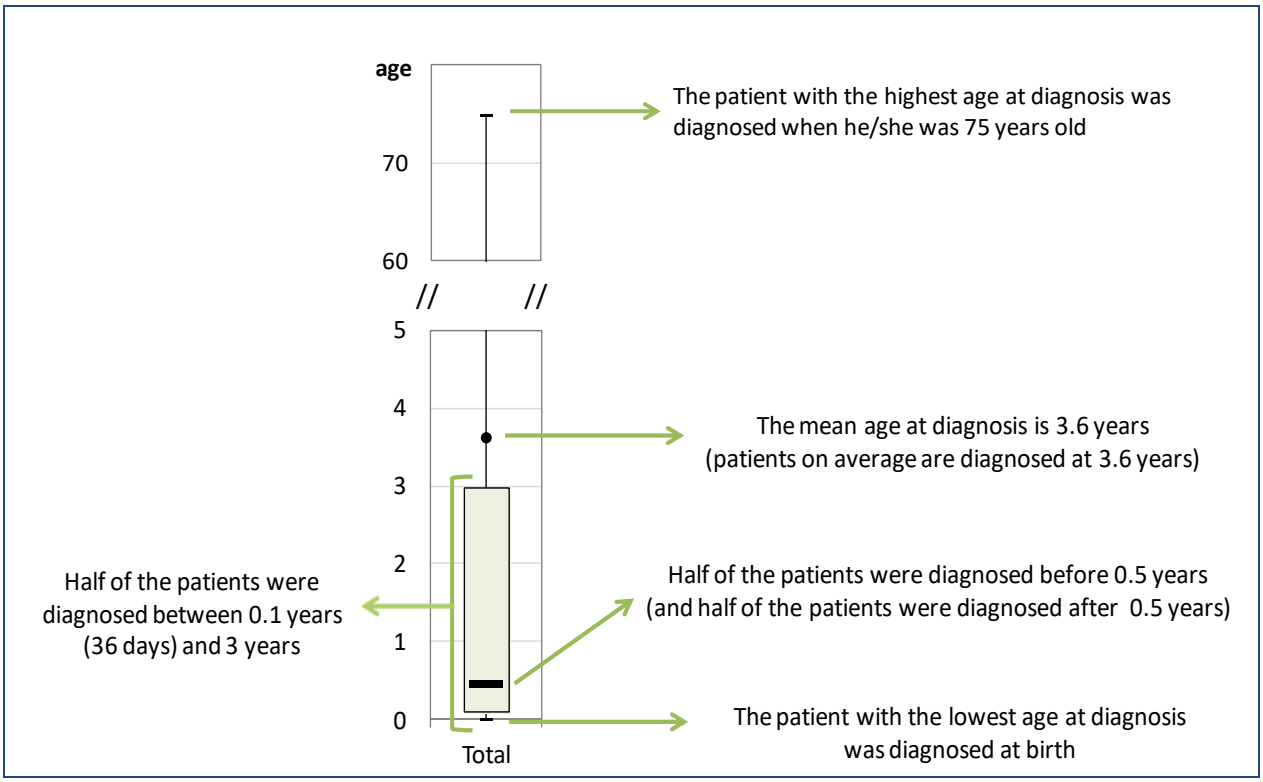
Table 2.1 shows the descriptive statistics for age at diagnosis by country and overall. For prenatal diagnoses (children diagnosed before birth), the age at diagnosis has been set to 0.

Figure 2.1 Age at diagnosis (in years): box-plot, by country and overall. All patients seen in 2019.



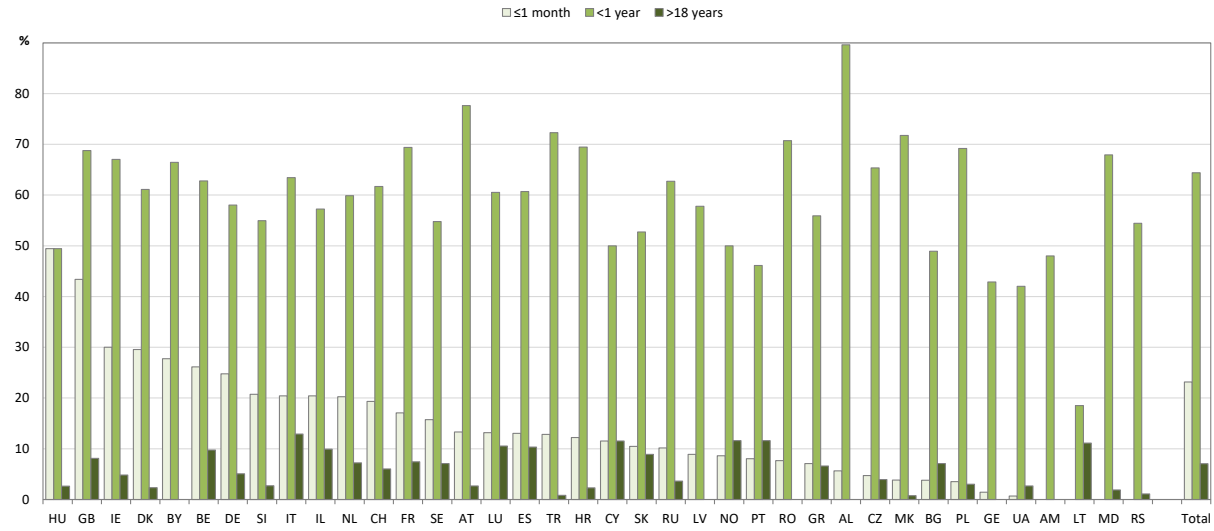
Note: For Hungary, Slovak Republic and Switzerland the information on age at diagnosis is missing for more than 10% of the patients.

This box-plot is a graphic representation of age at diagnosis as detailed in table 2.1. For each country the dash (black line crossing the blue 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. Please note that the vertical axis is interrupted to emphasise the change of scale in the upper part of the graph. The figure on the next page explains how to read the box-plot.



Note: This is an example of how to read a box-plot. The numbers used in this figure are not real and do not refer to figure 2.1.

Figure 2.2 Proportion of patients diagnosed at age 1 month or younger, younger than 1 year and older than 18 years, by country and overall. All patients seen in 2019.

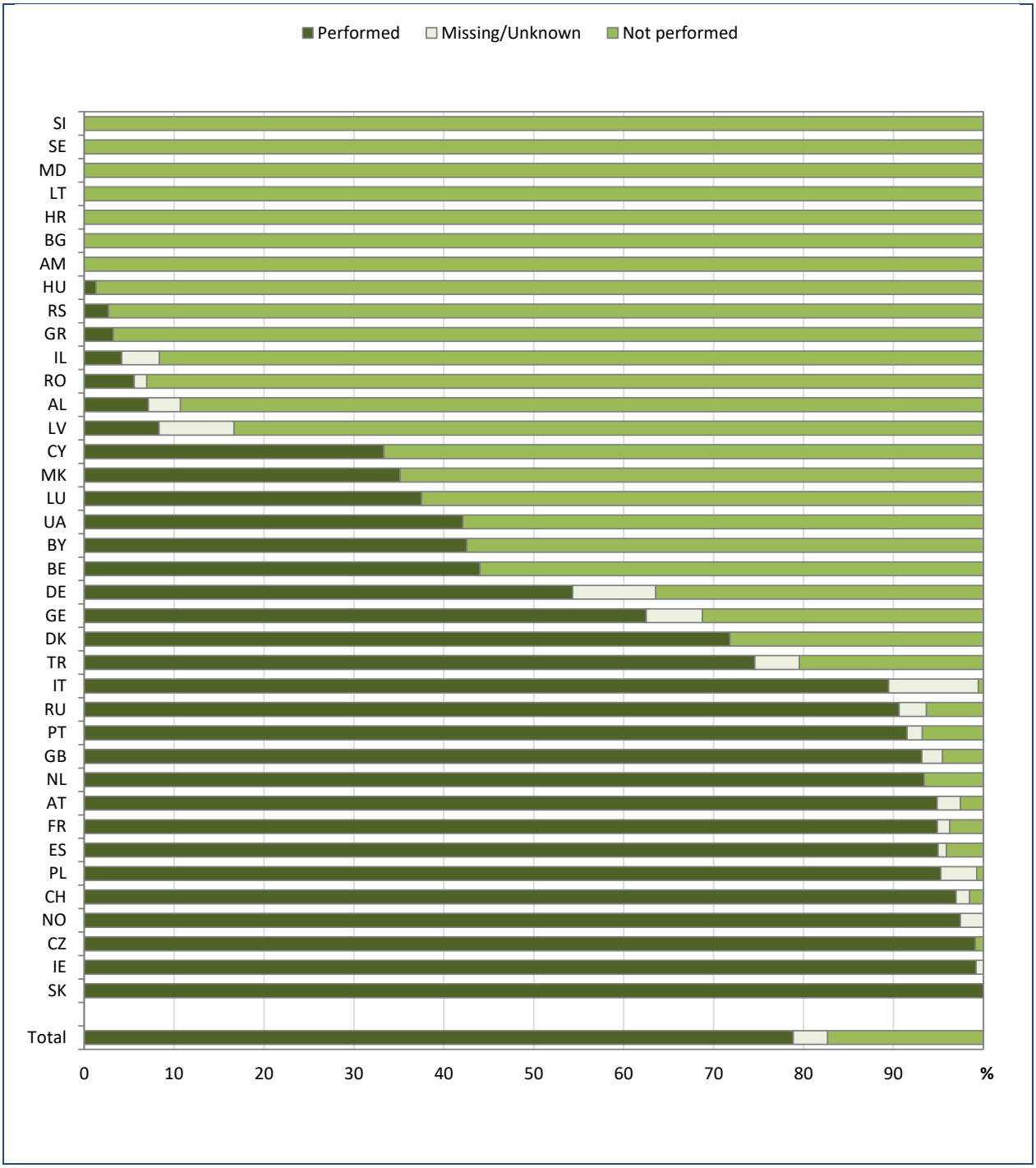


Note: For Hungary, Slovak Republic and Switzerland the information on age at diagnosis is missing for more than 10% of the patients.

Note: The high proportion of patients diagnosed at the age of ≤1 month for Hungary might be caused by an approximation of the age at diagnosis by the clinician.

This graph shows age at diagnosis in subgroups. The vertical bars represent how many patients (as a percentage) were diagnosed within the first month of life (grey), within the first year of life (light green), and after 18 years of age (dark green). Note that the number in the subgroup for diagnosis within 1 month are also part of the numbers in the subgroup for diagnoses within the first year, and that diagnoses between 1 year and 18 years are not shown in the graph; therefore, the bars do not sum to 100%.

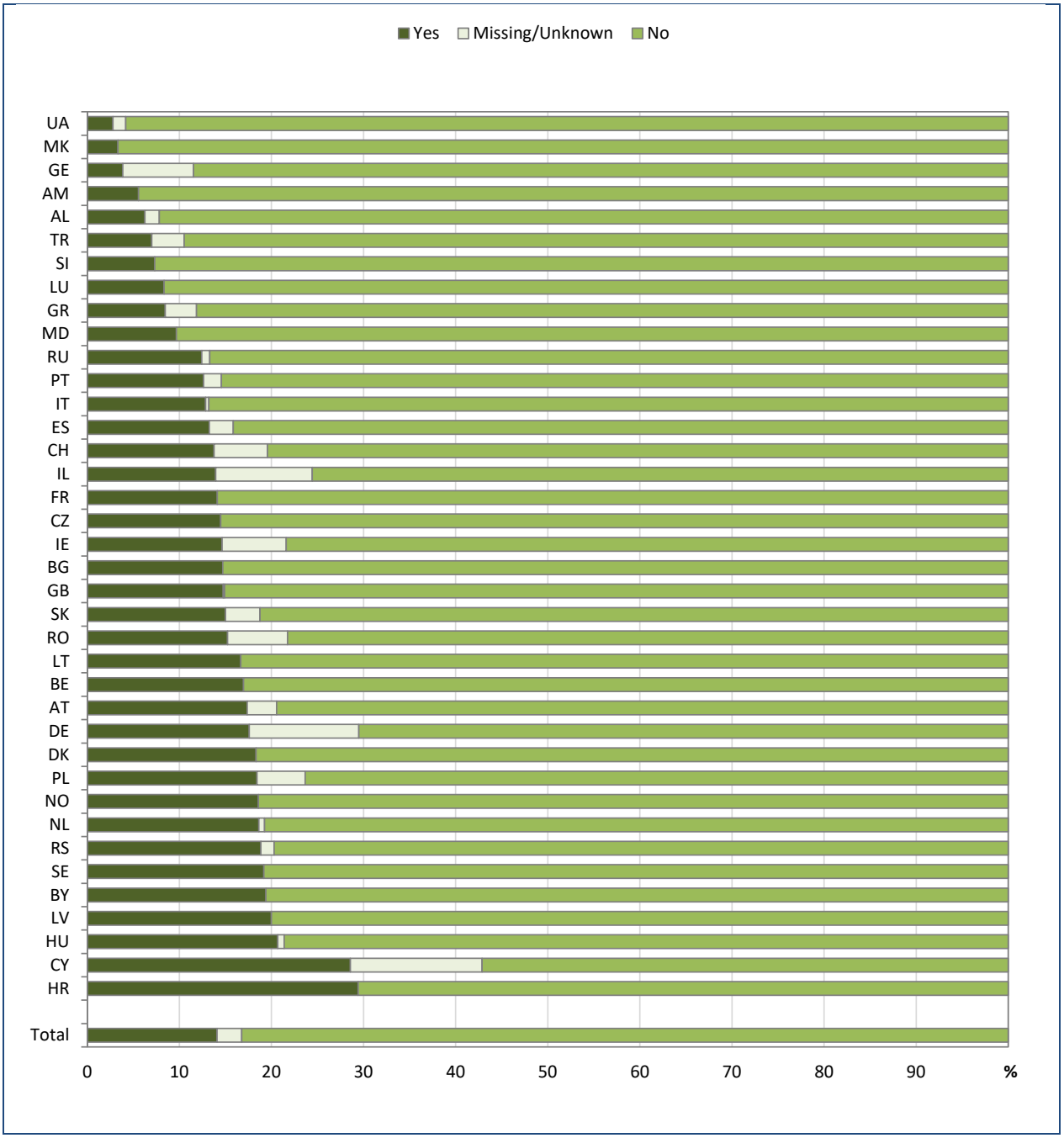
Figure 2.3 Proportion of patients who underwent neonatal screening, by country and overall. Patients 5 years old or younger seen in 2019.



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

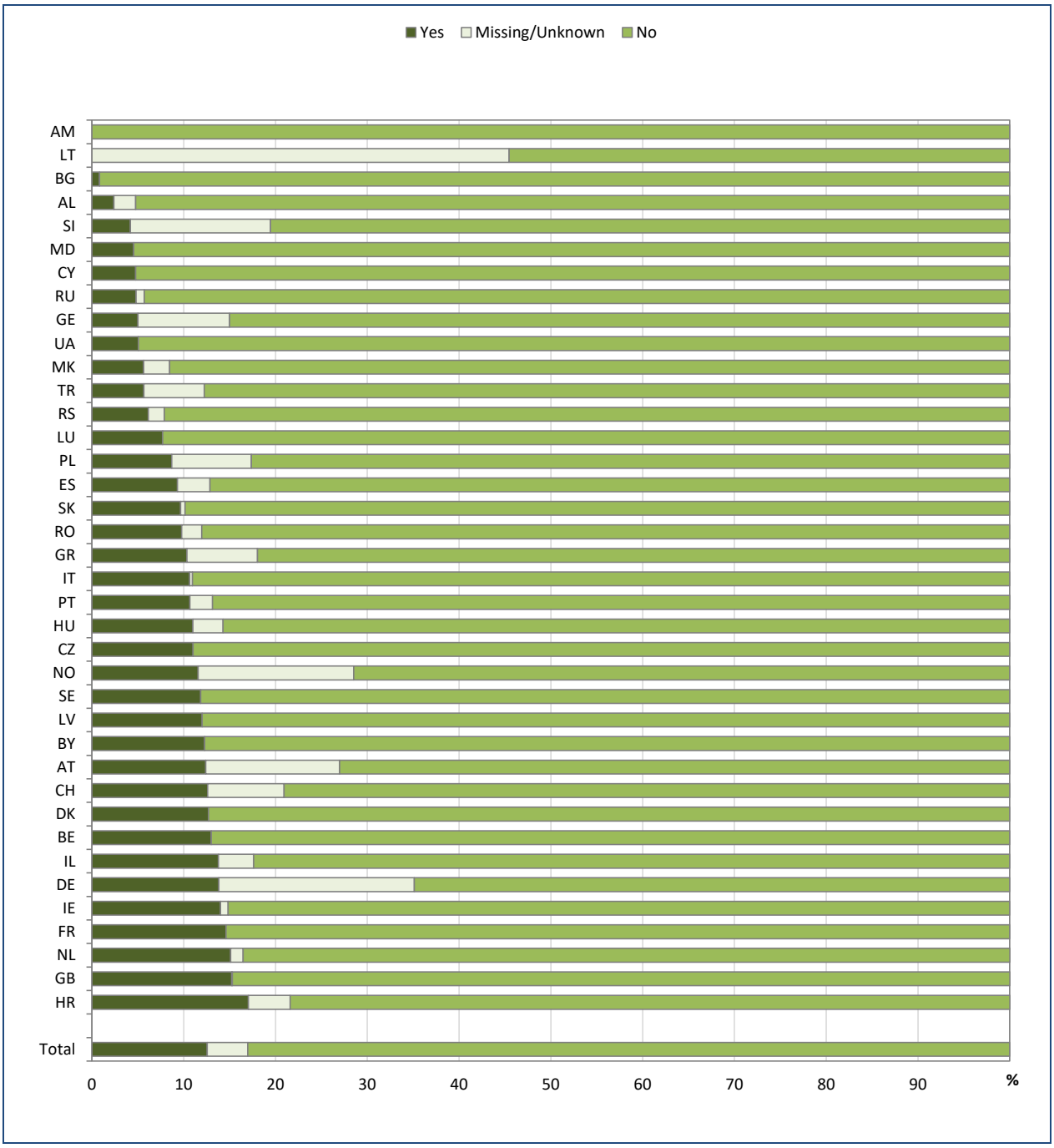
This graph shows the percentage of patients at the age of 5 years or younger in 2019 who were screened at birth. Dark green horizontal bars represent neonatal screening "performed", light green ones "not performed". This graph shows that, in the five years previous to 2019, in many countries the CF patients underwent neonatal, i.e. newborn screening, and that in some countries there is no neonatal screening programme. In total, 79% of all children of 5 years old or younger registered in the ECFSPR in 2019 were screened at birth. This estimate also reflects the fact that not all the countries carry out newborn screening.

Figure 2.4 Patients with meconium ileus, by country and overall. Patients aged 10 years or younger.



Note: For Cyprus, Germany and Israel the information on meconium ileus is missing for more than 10% of the patients ≤10 years.

Figure 2.5 Patients with meconium ileus, by country and overall. Patients aged 11 years or older.



Note: For Austria, Germany, Lithuania, Norway and Slovenia the information on meconium ileus is missing for more than 10% of the patients aged ≥11 years.

These two graphs show the prevalence of meconium ileus (with or without surgical repair) at birth in two age groups: 0 to 10 years (fig 2.4) and 11 years or older (fig 2.5). Overall, the proportion of younger patients (≤10 years) with meconium ileus is slightly higher compared to the older age group (>11 years). This difference is not because of an increase in the prevalence of meconium ileus in the younger generations but could be due to the fact that some older patients with meconium ileus have died and are therefore not present in the current data collection (which refers to patients seen in 2019). The graphs also show that the frequency of reported meconium ileus varies between countries.

3. Genetics

Cystic fibrosis is caused by mutations of the 'CFTR' gene; one on each allele. One mutation is inherited from the mother and one from the father. If both mutations are the same, the person is homozygous for this mutation. If these are two different mutations, the person is considered to be heterozygous.

We supplied the countries with a list of the 1600 most common mutations based on the Cystic Fibrosis Mutation database (CFTR1). If the patient had a mutation that was not present in the database, the country had the possibility to enter the name of the mutation as free text. During the data cleaning process, the genotypes not on our list were checked for obvious misspellings or alternative names and, if identified as a known mutation, renamed. Although there are different naming conventions for mutations, we use the original mutation name (legacy name) in this report, since more than 90% of the mutations in the database use this nomenclature.

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

If DNA analysis to look for CFTR mutations was never carried out, we asked the countries to report "Not done" in the genotype field. If DNA analysis was done, but only one or no mutations were found, we asked the countries to write "Unknown" for the unidentified mutations. Please note that there are differences from country to country in how DNA testing is carried out: some countries use standard kits that test only a limited number of common mutations (e.g. 28), and other countries perform DNA analyses of the whole gene until the mutation is identified.

Table 3.1 Proportion of patients with DNA analysis and the result of this, by country and overall. All patients seen in 2019.

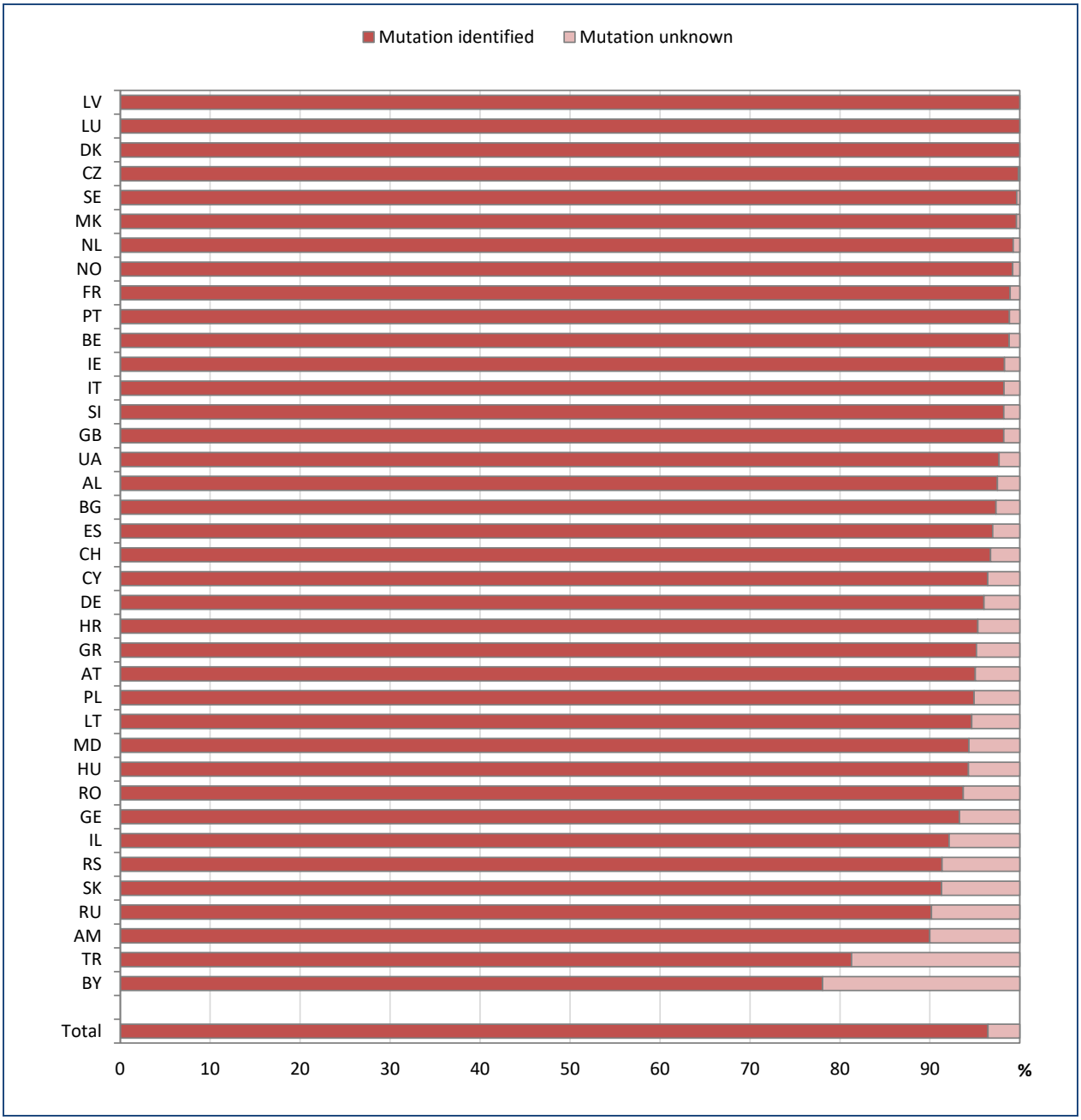
Country	Genotyping		Among genotyping done	
	not done	done	at least one mutation unknown	two mutations identified
	number (%)	number (%)	number (%)	number (%)
Albania	6 (5.66)	100 (94.34)	5 (5.00)	95 (95.00)
Armenia	0 (0.00)	25 (100.00)	5 (20.00)	20 (80.00)
Austria	1 (0.12)	815 (99.88)	56 (6.87)	759 (93.13)
Belarus	0 (0.00)	155 (100.00)	47 (30.32)	108 (69.68)
Belgium	0 (0.00)	1294 (100.00)	26 (2.01)	1268 (97.99)
Bulgaria	0 (0.00)	188 (100.00)	10 (5.32)	178 (94.68)
Croatia	0 (0.00)	139 (100.00)	10 (7.19)	129 (92.81)
Cyprus	0 (0.00)	28 (100.00)	2 (7.14)	26 (92.86)
Czech Republic	1 (0.16)	642 (99.84)	2 (0.31)	640 (99.69)
Denmark	0 (0.00)	514 (100.00)	0 (0.00)	514 (100.00)
France	0 (0.00)	6729 (100.00)	110 (1.63)	6619 (98.37)
Georgia	5 (6.94)	67 (93.06)	7 (10.45)	60 (89.55)
Germany	19 (0.29)	6462 (99.71)	406 (6.28)	6056 (93.72)
Greece	0 (0.00)	436 (100.00)	33 (7.57)	403 (92.43)
Hungary	1 (0.20)	508 (99.80)	46 (9.06)	462 (90.94)
Ireland	0 (0.00)	1251 (100.00)	35 (2.80)	1216 (97.20)
Israel	1 (0.19)	528 (99.81)	54 (10.23)	474 (89.77)
Italy	14 (0.25)	5571 (99.75)	170 (3.05)	5401 (96.95)
Latvia	0 (0.00)	45 (100.00)	0 (0.00)	45 1(00.00)
Lithuania	0 (0.00)	28 (100.00)	2 (7.14)	26 (92.86)
Luxembourg	0 (0.00)	38 (100.00)	0 (0.00)	38 (100.00)
Rep of Moldova	0 (0.00)	53 (100.00)	5 (9.43)	48 (90.57)
The Netherlands	8 (0.53)	1493 (99.47)	19 (1.27)	1474 (98.73)
North Macedonia	1 (0.76)	130 (99.24)	1 (0.77)	129 (99.23)

[table 3.1 continued]

Country	Genotyping		Among genotyping done	
	not done	done	at least one mutation unknown	two mutations identified
	number (%)	number (%)	number (%)	number (%)
Norway	0 (0.00)	312 (100.00)	3 (0.96)	309 (99.04)
Poland	2 (0.17)	1192 (99.83)	86 (7.21)	1106 (92.79)
Portugal	0 (0.00)	347 (100.00)	6 (1.73)	341 (98.27)
Romania	0 (0.00)	230 (100.00)	24 (10.43)	206 (89.57)
Russian Federation	156 (4.93)	3006 (95.07)	495 (16.47)	2511 (83.53)
Serbia	4 (2.19)	179 (97.81)	25 (13.97)	154 (86.03)
Slovak Republic	0 (0.00)	287 (100.00)	42 (14.63)	245 (85.37)
Slovenia	2 (1.77)	111 (98.23)	4 (3.60)	107 (96.40)
Spain	1 (0.04)	2328 (99.96)	126 (5.41)	2202 (94.59)
Sweden	0 (0.00)	704 (100.00)	4 (0.57)	700 (99.43)
Switzerland	4 (0.41)	970 (99.59)	45 (4.64)	925 (95.36)
Turkey	100 (5.03)	1888 (94.97)	456 (24.15)	1432 (75.85)
Ukraine	0 (0.00)	151 (100.00)	6 (3.97)	145 (96.03)
United Kingdom	35 (0.35)	10035 (99.65)	313 (3.12)	9722 (96.88)
Total	361 (0.73)	48979 (99.27)	2686 (5.48)	46293 (94.52)

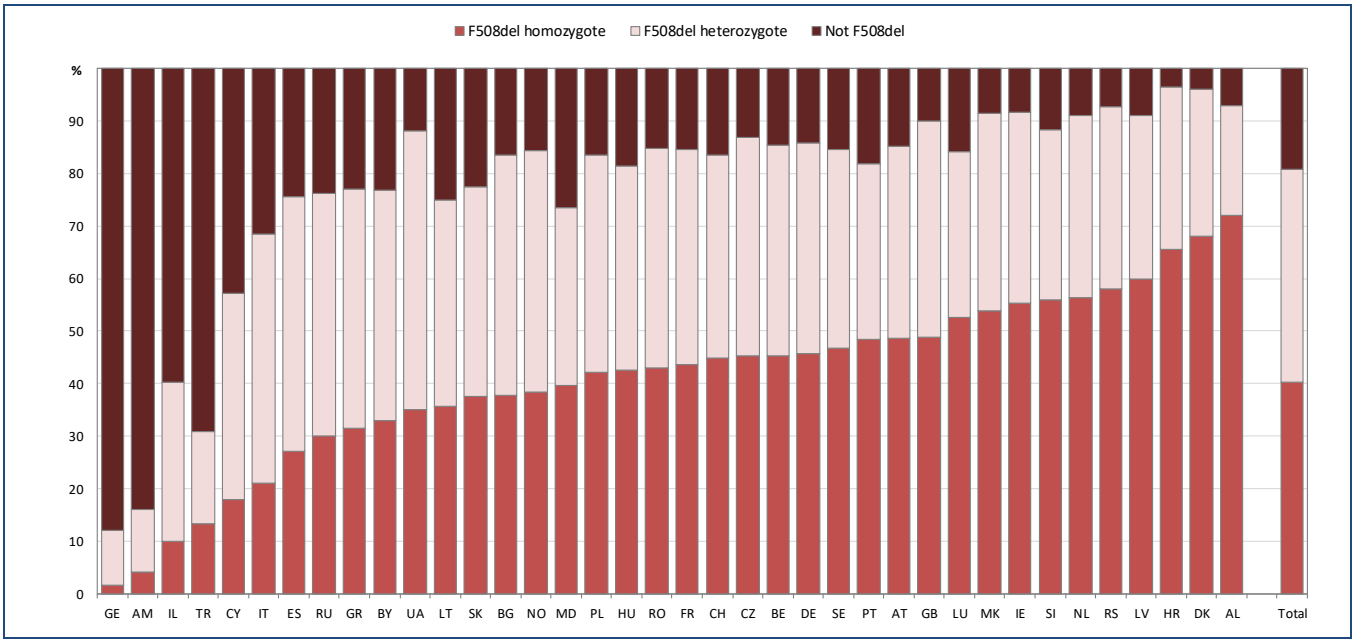
The table shows how many patients underwent DNA analysis to identify the CFTR mutations (column “genotyping done”) and, for those patients, how many patients had both mutations identified (column “two mutations identified”) and for how many one or both mutations remained unidentified (column “at least one mutation unknown”).

Figure 3.1 Proportion of identified mutations, by country and overall. Only patients with DNA analysis.



This graph shows the percentage of mutations that are not identified (in light pink) after DNA analysis, by country and overall. One "allele" means one of the two CFTR genes. The number of non-identified alleles varies greatly from country to country; this is partly due to the different approaches to DNA testing. Overall, more than 3% of mutations remain unidentified after DNA analysis, leaving 5.48% of the patients with at least one mutation unidentified.

Figure 3.2 Prevalence of F508del homozygous and heterozygous patients, by country and overall. All patients seen in 2019.



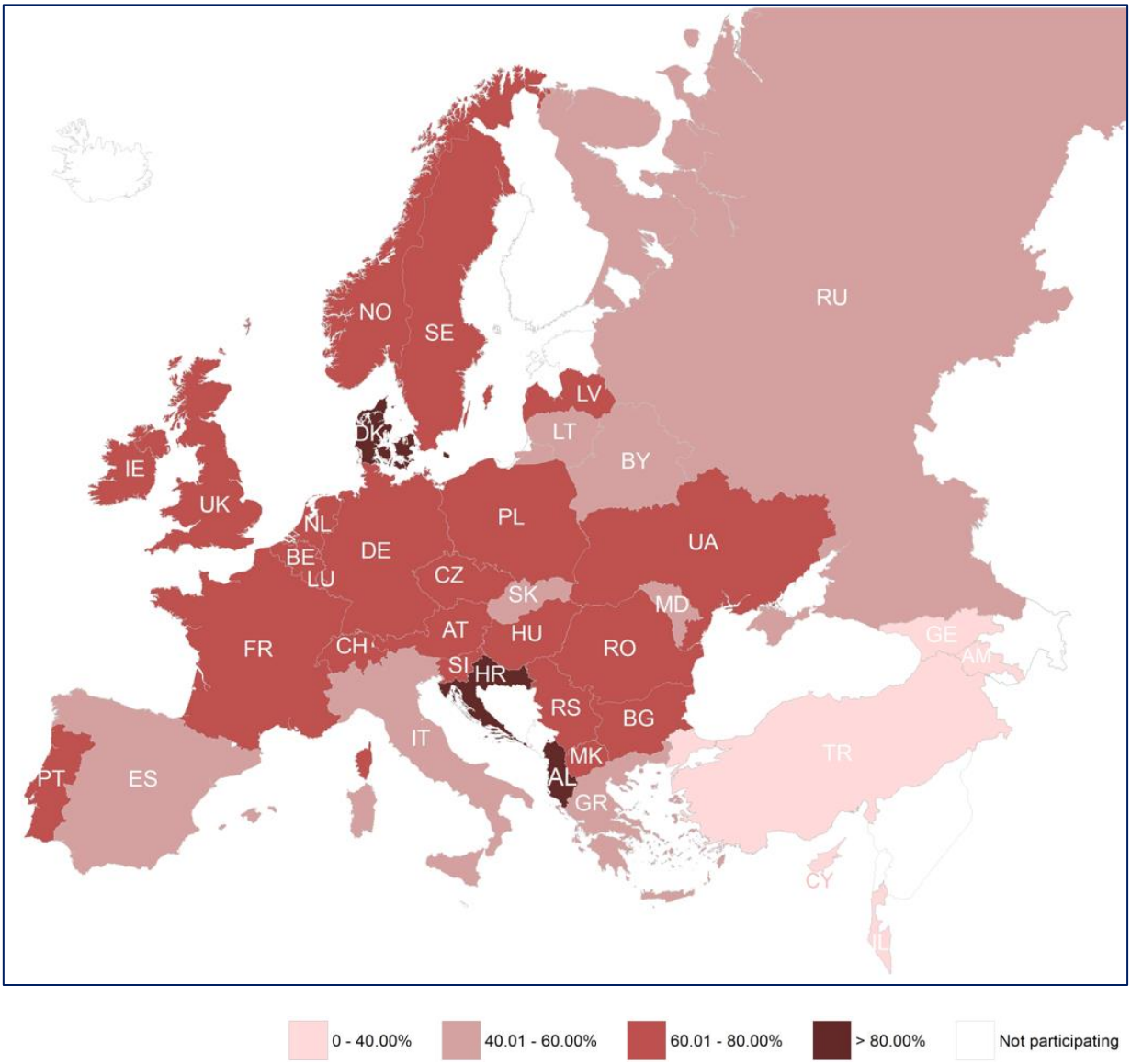
F508del is the name of the most commonly occurring CFTR mutation in the world. Patients who carry two F508del mutations are often described as having “classic CF”, but other combinations of mutations may cause the same degree of disease. We have grouped the patients in F508del homozygous (patients who have two F508del mutations), F508del heterozygous (patients who have one F508del mutation and another known mutation, different from F508del), and patients without F508del mutations. Only patients for whom the genotype is known have been included in this graph. “Unknown” mutations 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.

Table 3.2 Allelic frequencies of the 17 most common mutations in the ECFSPR database.

Mutation name	Number of alleles	Percentage among tested	Country with highest allele frequency
F508del	59205	60.44	Albania (82.5%)
G542X	2657	2.71	Armenia (8.0%)
N1303K	2104	2.15	Italy (5.5%)
G551D	1259	1.29	Ireland (8.3%)
W1282X	1041	1.06	Israel (23.0%)
2789+5G->A	1039	1.06	Turkey (3.1%)
3849+10kbC->T	986	1.01	Lithuania (12.5%)
CFTRdele2,3	983	1.00	Belarus (8.7%)
R117H	955	0.97	Ireland (3.1%)
1717-1G->A	844	0.86	Switzerland (3.1%)
R553X	821	0.84	Lithuania (3.6%)
2183AA->G	669	0.68	Armenia (10.0%)
621+1G->T	616	0.63	Greece (6.2%)
D1152H	611	0.62	Israel (5.5%)
R347P	541	0.55	Luxembourg (2.6%)
G85E	502	0.51	Israel (2.6%)
3272-26A->G	490	0.50	Belgium (2.1%)

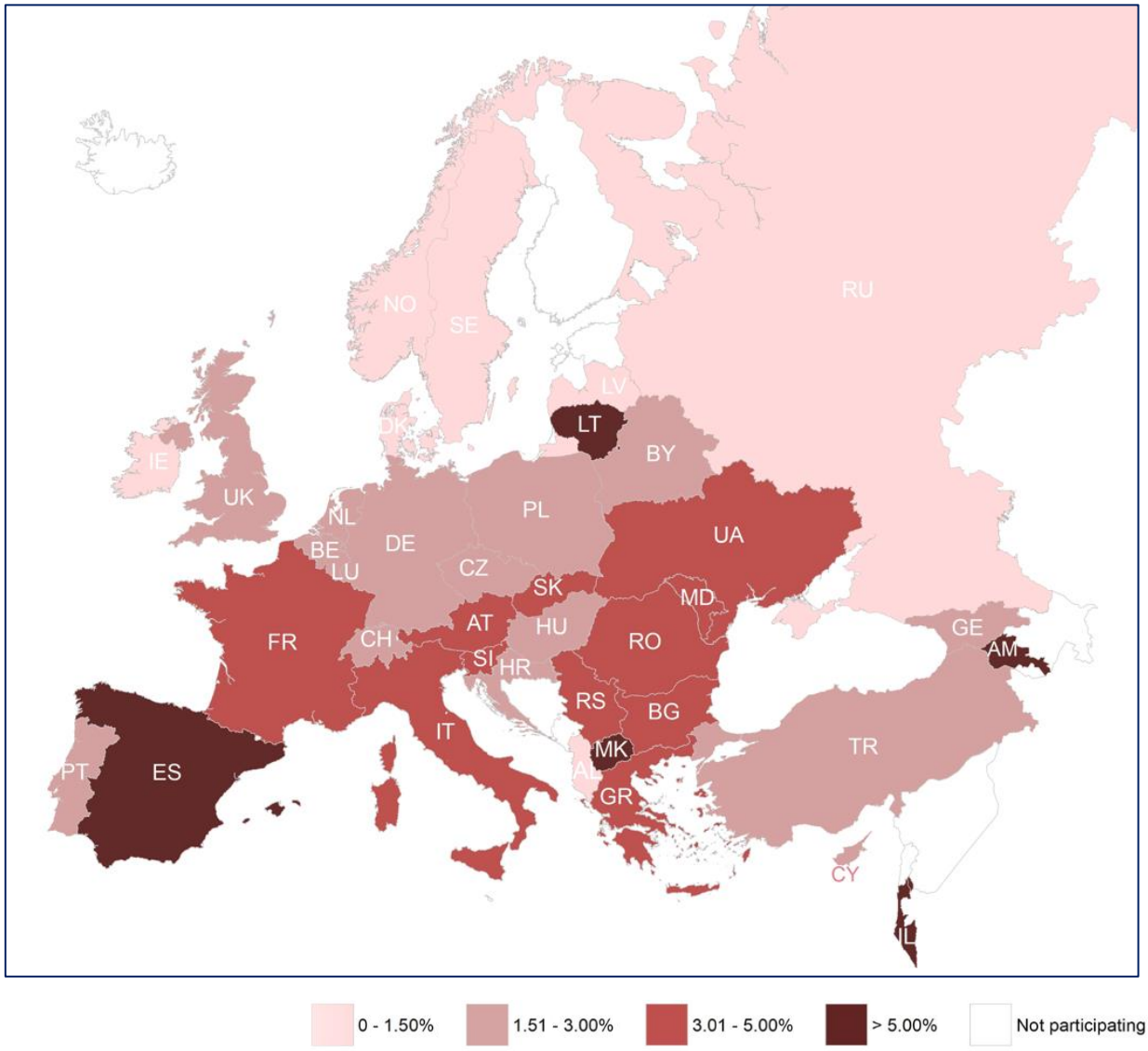
This table presents the allele frequency of the 17 most commonly occurring mutations found in the ECFSPR database. The last column reports in which country this particular mutation is most frequent. F508del is, by far, the most frequent mutation.

Figure 3.3 Geographical distribution of mutation F508del.



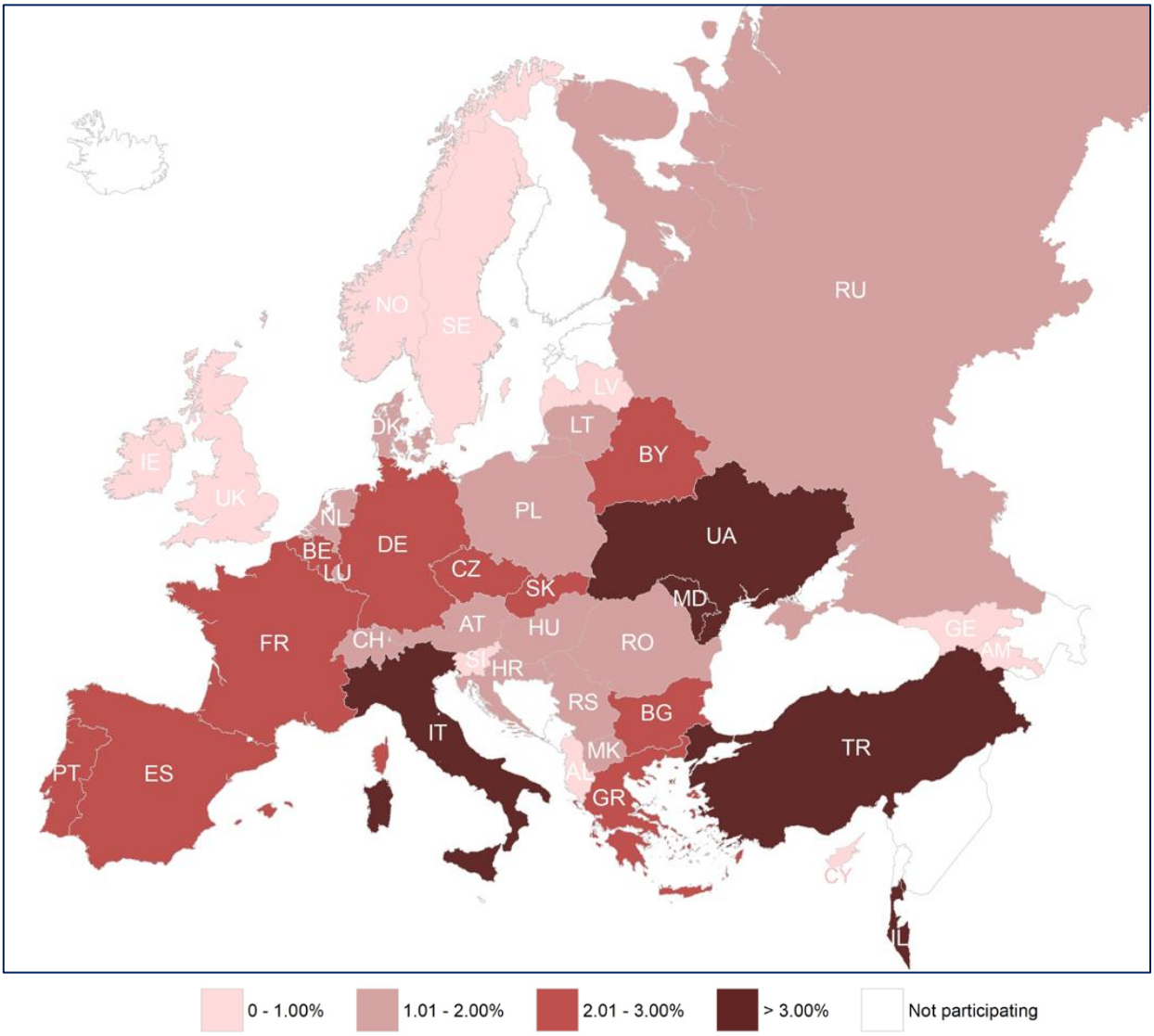
Although this mutation is the most common in all countries, the highest frequency occurs in Albania (82.5%) and Croatia (80.90%), and in the north of Europe, in Denmark (82.1%).

Figure 3.4 Geographical distribution of mutation G542X.



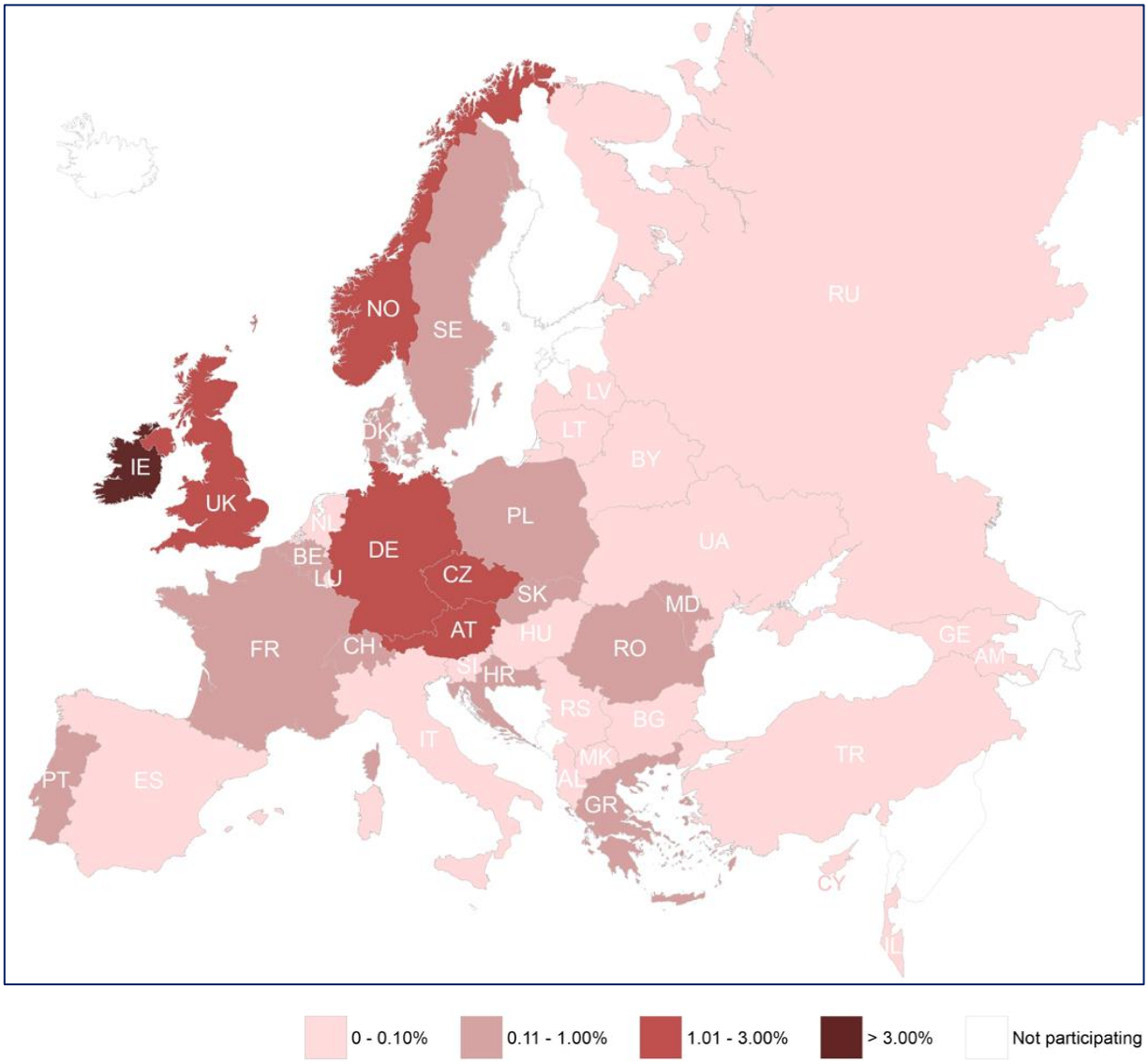
This mutation is most frequent in Southern Europe, with the highest allele frequency in Armenia (8.0%), whereas it is very rarely found in Ireland, the Scandinavian and the Russian Federation.

Figure 3.5 Geographical distribution of mutation N1303K.



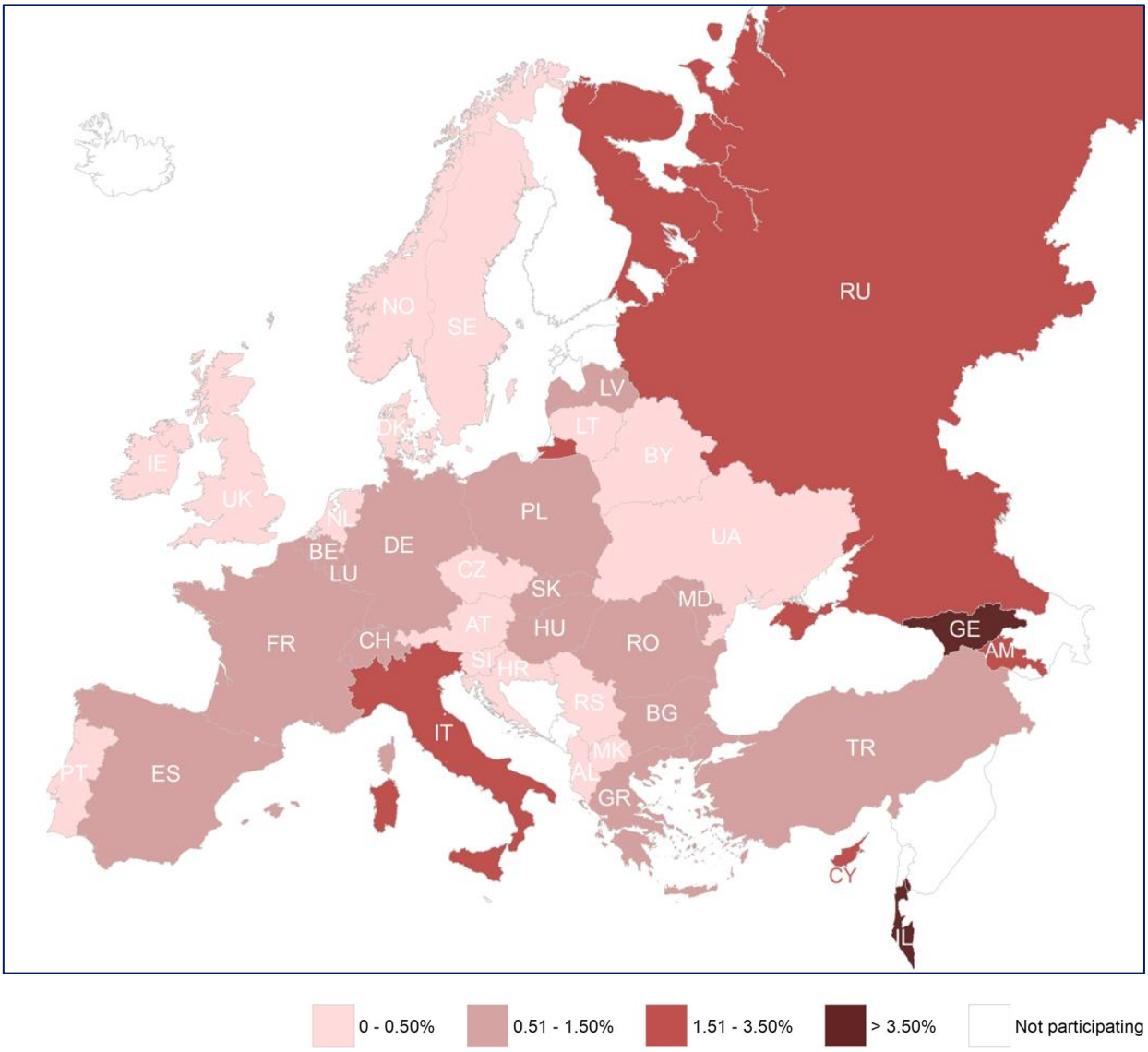
This mutation is most frequent in Italy (5.5%) and other countries in Southern and Eastern Europe, but rare in Northern Europe.

Figure 3.6 Geographical distribution of mutation G551D.



This mutation is most frequent in Ireland (8.3%) and in the north of Europe whereas it is rare in Southern and Eastern Europe.

Figure 3.7 Geographical distribution of mutation W1282X.



This mutation, of Middle-Eastern origin, is by far most frequent in Israel (23.0%) with a very high allele frequency in Ashkenazi Jews; and frequent in Georgia (9.0%).

4. Lung function

FEV_1 is measured in litres but it is normally expressed as a percentage of the expected (i.e. predicted) value (FEV_1%). The predicted value is computed from healthy individuals of the same sex, height and age and is termed the reference population.

We used the Global Lung Function Initiative equations described by Quanjer PH et al. for this report (for the full reference we refer you to Appendix 3, page 156). 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 100 means that the lung function measurement is equal to the mean lung function measurement of people of the same age, sex and height of the healthy reference population.

Spirometry, the test that measures FEV_1, requires a certain amount of coordination, and usually cannot be performed reliably until a person with CF is about four to six years of age. We have therefore computed FEV_1% values only for patients aged 6 years or older.

We asked the countries to report the best FEV_1 recorded throughout the year (relative to the best FEV_1% computed at the CF centres) to the ECFSPR.

We excluded patients from the analyses on 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 patients who had a liver or other transplantation since the follow-up data of those patients is sometimes missing.

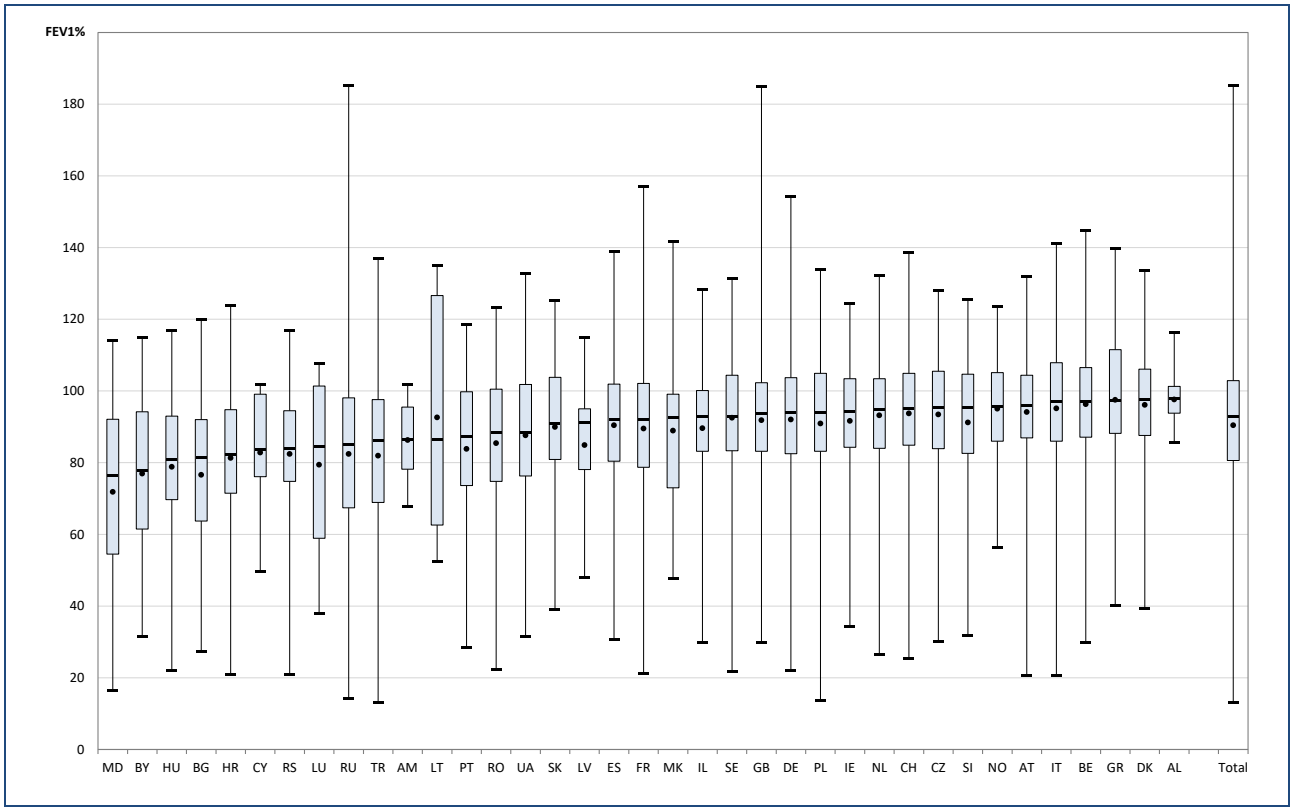
Table 4.1 FEV_1% of predicted: descriptive statistics, by country. Patients aged 6-17 years who have never had a transplant.

Country	N	N Miss	Mean (average FEV_1%)	Min	25th pctl (25% of patients have FEV_1% below this value)	Median (50% of patients have FEV_1% below this value)	75th pctl (75% of patients have FEV_1% below this value)	Max
Albania	53	15	97.6	85.7	93.8	97.8	101.3	116.2
Armenia	14	2	86.3	67.8	78.2	86.4	95.5	101.8
Austria	253	1	94.1	20.6	86.9	95.8	104.4	131.8
Belarus	49	22	76.9	31.4	61.5	77.7	94.2	115.0
Belgium	325	6	96.3	29.7	87.1	97.0	106.5	144.8
Bulgaria	65	7	76.6	27.4	63.7	81.3	92.0	120.0
Croatia	53	5	81.3	21.0	71.5	82.2	94.8	123.7
Cyprus	10	1	82.8	49.6	76.1	83.5	99.1	101.7
Czech Republic	213	6	93.4	30.0	83.9	95.5	105.5	128.0
Denmark	131	1	96.1	39.3	87.6	97.6	106.1	133.5
France	1896	86	89.5	21.1	78.7	92.0	102.1	156.9
Germany	1734	24	92.0	21.9	82.5	93.9	103.7	154.3
Greece	67	2	97.5	40.1	88.2	97.3	111.5	139.8
Hungary	137	36	78.8	21.9	69.7	80.8	93.0	116.9
Ireland	381	9	91.6	34.3	84.3	94.2	103.4	124.5
Israel	177	0	89.6	29.7	83.2	92.9	100.1	128.2
Italy	1511	59	95.1	20.5	86.0	96.9	107.9	141.2
Latvia	16	4	84.9	48.0	78.1	91.2	95.0	114.8
Lithuania	10	0	92.6	52.4	62.6	86.5	126.6	135.1
Luxembourg	8	0	79.4	37.9	58.9	84.4	101.4	107.6
Rep of Moldova	26	2	71.8	16.4	54.5	76.4	92.1	114.1
The Netherlands	411	4	93.2	26.5	84.0	94.8	103.4	132.2
North Macedonia	49	3	88.9	47.6	73.0	92.6	99.1	141.7
Norway	75	1	95.0	56.4	86.0	95.7	105.1	123.5
Poland	509	41	90.9	13.6	83.2	94.0	104.9	134.0
Portugal	125	6	83.8	28.4	73.6	87.4	99.8	118.4
Romania	100	41	85.4	22.3	74.8	88.3	100.5	123.2
Russian Federation	1052	436	82.4	14.3	67.4	85.0	98.1	185.1
Serbia	85	4	82.4	21.0	74.8	84.0	94.5	116.7
Slovak Republic	86	1	89.9	39.0	80.9	91.0	103.8	125.2
Slovenia	41	0	91.2	31.8	82.6	95.5	104.7	125.4
Spain	731	18	90.4	30.7	80.4	92.0	101.9	139.0
Sweden	190	3	92.5	21.8	83.3	92.9	104.4	131.3
Switzerland	286	9	93.7	25.4	84.9	95.2	104.9	138.6
Turkey	768	198	81.9	13.2	68.9	86.1	97.6	136.9
Ukraine	78	4	87.6	31.5	76.3	88.4	101.8	132.9
United Kingdom	2986	61	91.8	29.8	83.2	93.6	102.3	185.0
Total	14702	1149	90.4	13.2	80.6	92.8	102.9	185.1

Note: Georgia has <5 patients aged 6-17 years at FEV_1 measurement and is excluded from the table.
Note: The United Kingdom reports best FEV1 from the annual review, which is the time period between data sets and is not necessarily a calendar year. Therefore, in some cases measurement of FEV1 could be from the previous calendar year.

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

Figure 4.1 FEV_1% of predicted: box-plot, by country and overall. Patients aged 6-17 years who have never had a transplant.



Note: Georgia has <5 patients aged 6-17 years at FEV_1 measurement and is excluded from the graph.

Note: The United Kingdom reports best FEV1 from the annual review, which is the time period between data sets and is not necessarily a calendar year. Therefore, in some cases measurement of FEV1 could be from the previous calendar year.

This box-plot is a graphic representation of the FEV_1 in children, expressed as % of predicted, detailed in table 4.1. For each country, the dash (black line crossing the blue 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

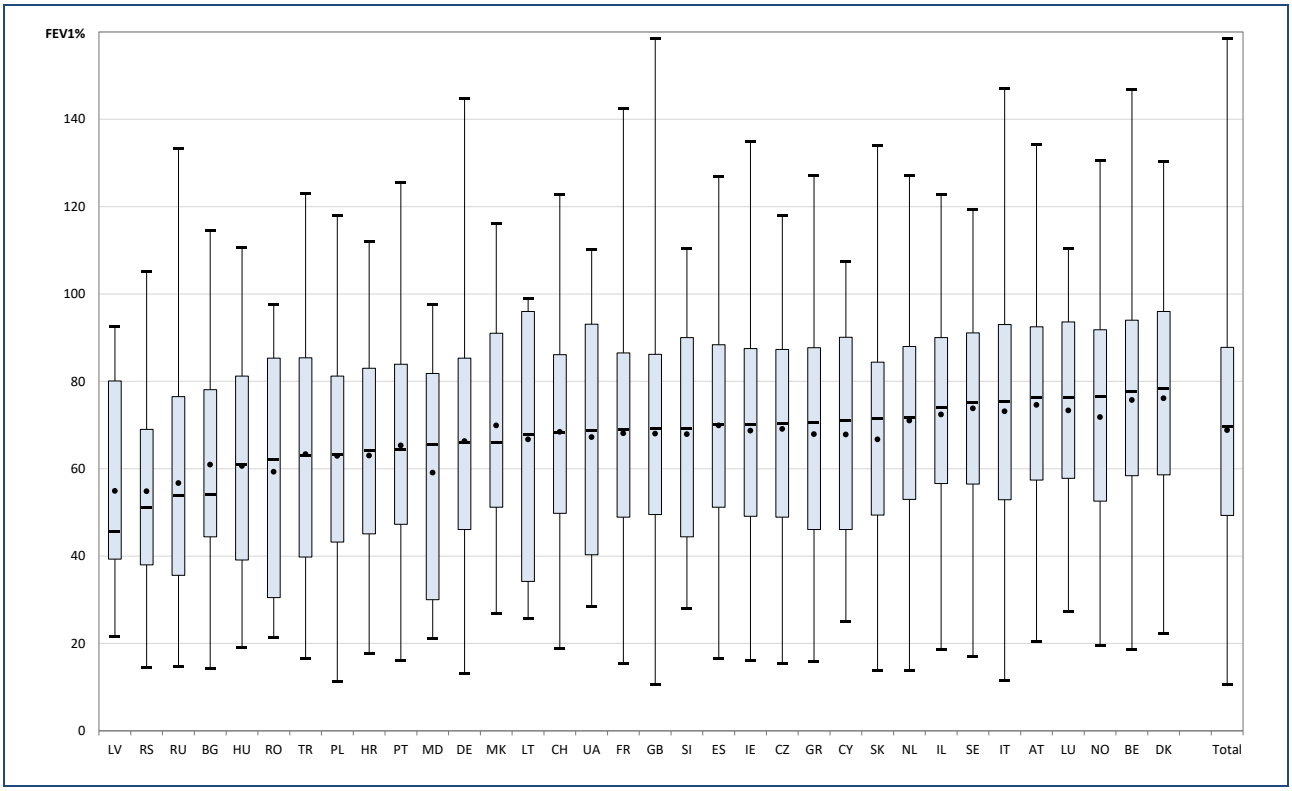
Table 4.2 FEV_1% of predicted: descriptive statistics, by country. Patients aged 18 years or older who have never had a transplant.

Country	N	N Miss	Mean (average FEV_1%)	Min	25th pctl (25% of patients have FEV_1% below this value)	Median (50% of patients have FEV_1% below this value)	75th pctl (75% of patients have FEV_1% below this value)	Max
Austria	333	2	74.6	20.5	57.4	76.2	92.5	134.2
Belgium	630	9	75.7	18.6	58.4	77.7	94.0	146.7
Bulgaria	66	1	60.9	14.3	44.4	54.0	78.1	114.4
Croatia	39	0	63.0	17.7	45.1	64.2	83.0	111.9
Cyprus	13	0	67.8	25.1	46.1	71.0	90.1	107.4
Czech Republic	247	6	69.1	15.5	48.9	70.3	87.3	118.0
Denmark	259	0	76.1	22.3	58.6	78.3	96.0	130.2
France	2923	48	68.1	15.3	48.9	69.0	86.5	142.4
Germany	3186	62	66.3	13.1	46.1	66.0	85.3	144.8
Greece	265	4	67.9	15.9	46.1	70.6	87.7	127.2
Hungary	167	28	60.6	19.1	39.1	61.0	81.2	110.6
Ireland	604	13	68.7	16.1	49.1	70.2	87.5	135.0
Israel	303	0	72.4	18.7	56.6	74.0	90.0	122.7
Italy	2764	80	73.1	11.5	52.9	75.3	93.0	147.1
Latvia	12	1	54.9	21.5	39.3	45.7	80.1	92.5
Lithuania	11	0	66.7	25.8	34.2	67.8	96.0	99.0
Luxembourg	20	0	73.3	27.2	57.8	76.2	93.6	110.3
Rep of Moldova	8	0	59.1	21.1	30.0	65.5	81.8	97.5
The Netherlands	808	3	71.0	13.8	53.0	71.8	88.0	127.1
North Macedonia	36	0	69.9	26.9	51.2	66.0	91.0	116.2
Norway	157	3	71.8	19.6	52.6	76.6	91.8	130.6
Poland	284	11	62.9	11.3	43.2	63.2	81.2	118.0
Portugal	116	8	65.3	16.2	47.3	64.4	83.9	125.4
Romania	8	0	59.3	21.3	30.5	62.0	85.3	97.6
Russian Federation	504	208	56.7	14.8	35.6	53.8	76.5	133.4
Serbia	54	0	54.8	14.4	38.0	51.1	69.0	105.1
Slovak Republic	139	1	66.7	13.7	49.4	71.4	84.4	133.9
Slovenia	38	0	67.9	27.9	44.4	69.2	90.0	110.4
Spain	965	16	69.9	16.5	51.2	70.2	88.4	126.9
Sweden	339	9	73.8	17.1	56.5	75.1	91.1	119.4
Switzerland	467	1	68.4	18.8	49.8	68.3	86.1	122.7
Turkey	193	27	63.3	16.5	39.8	63.1	85.4	123.0
Ukraine	24	3	67.2	28.4	40.3	68.8	93.1	110.2
United Kingdom	5215	47	68.0	10.5	49.5	69.2	86.2	158.5
Total	21201	594	68.8	10.5	49.3	69.7	87.8	158.5

Note: Georgia has 0% coverage for adults and is excluded from the table.
Note: Albania, Armenia, Belarus have <5 patients aged 18 years or more at FEV_1 measurement and are excluded from the table.
Note: The United Kingdom reports best FEV1 from the annual review, which is the time period between data sets and is not necessarily a calendar year. Therefore, in some cases measurement of FEV1 could be from the previous calendar year.

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

Figure 4.2 FEV_1% of predicted: box-plot, by country and overall. Patients aged 18 years or older who have never had a transplant.



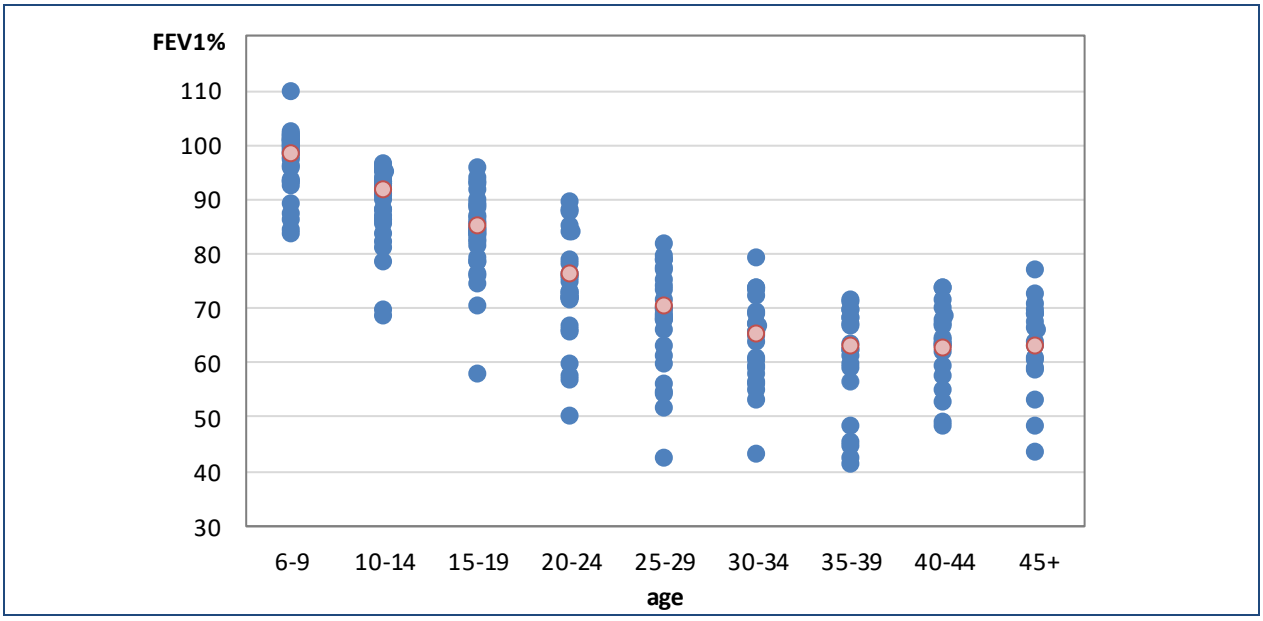
Note: Georgia has 0% coverage for adults and is excluded from the graph.

Note: Albania, Armenia and Belarus have <5 patients aged 18 years or more at FEV_1 measurement and are excluded from the graph.

Note: The United Kingdom reports best FEV1 from the annual review, which is the time period between data sets and is not necessarily a calendar year. Therefore, in some cases measurement of FEV1 could be from the previous calendar year.

This box-plot is a graphic representation of the FEV_1 in adults, expressed as % of predicted detailed in table 4.2. For each country the dash (black line crossing the blue 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.

Figure 4.3 Median FEV_1% of predicted by age group and by country. Patients aged 6 years or older who have never had a transplant.



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

This graph shows the median FEV_1% (the value that separates the highest and lowest half of the patients) by age group. Each country is represented by a dot (in blue) and the overall estimate is in red. The general pattern shows that the FEV_1% slowly decreases until the age of 35-39, and then levels out. The patients in the oldest age groups are patients that survived, and may therefore represent the patients with less disease severity. There is considerable variability between countries.

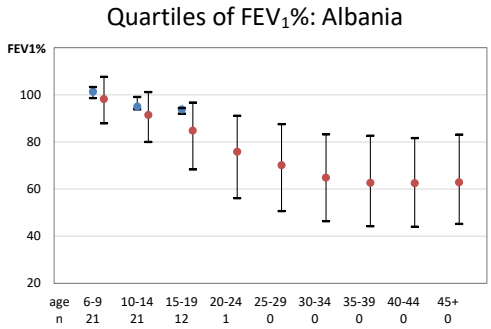
Table 4.3 FEV_1% of predicted: descriptive statistics by age group (patients aged 6 years or older) who have never had a transplant.

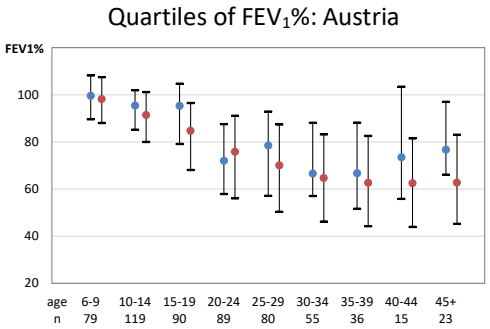
Age at FEV_1 measurement	N	N Miss	Mean	Min	25th pctl	Median	75th pctl	Max
6-9	5051	624	96.9	18.1	88.0	98.2	107.6	185.1
10-14	6235	371	89.0	13.6	80.1	91.5	101.1	157.5
15-19	5537	225	81.3	13.1	68.3	84.9	96.7	158.5
20-24	4825	149	73.2	10.5	56.1	75.8	91.1	142.4
25-29	4147	122	69.2	11.4	50.6	70.1	87.5	144.8
30-34	3298	92	65.3	13.8	46.3	64.8	83.2	136.3
35-39	2377	62	63.7	11.3	44.2	62.6	82.6	146.7
40-44	1620	39	63.8	12.2	44.0	62.5	81.6	147.1
45+	2813	59	64.5	11.8	45.2	62.8	83.1	141.2

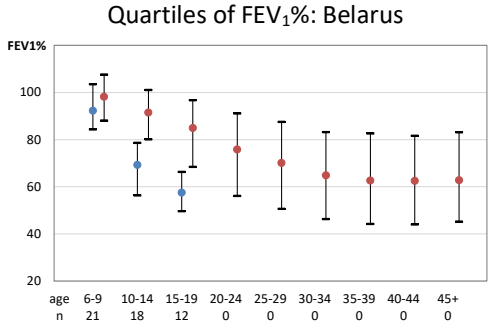
This table shows FEV_1% by age group for the total data-set. The median values reported in this table are shown as red dots in fig 4.3.

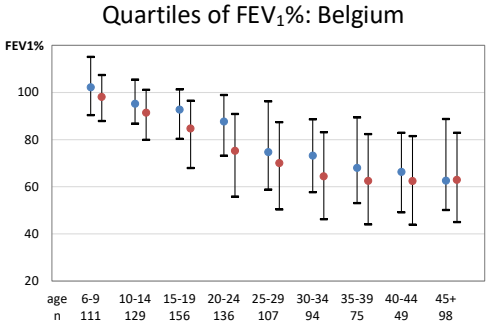
Figure 4.4 Quartiles of FEV$_1$% of predicted by age group and by country. Patients aged 6 years or older and who have never had a transplant.

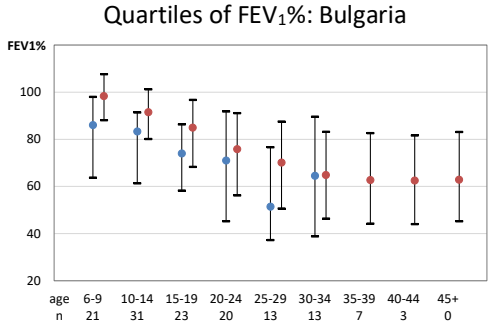
The figures below show the FEV$_1$% in different age groups, separately for each country. The dot shows the median, and the whiskers show the 25th and 75th percentiles (the median, the 25th percentile and the 75th percentile are collectively named "quartiles"). In blue are the quartiles for the country, in red are the pooled quartiles computed on all other countries (i.e. excluding that country). We did not compute quartiles where the number of patients is <10 in an age group, so there are no blue dots for those age groups (the number of patients in each age group is shown below the horizontal axis). We therefore excluded Armenia, Cyprus, Georgia, Latvia, Lithuania, and Luxembourg from the graphs because none of the age groups had more than 10 patients.

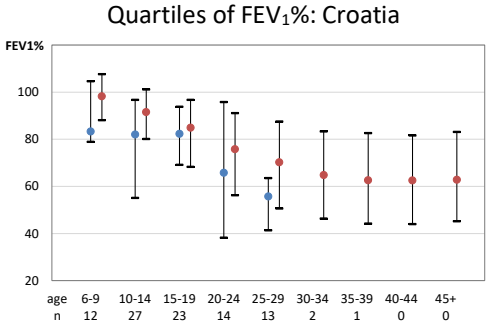


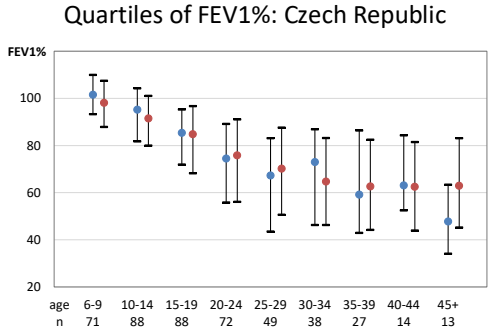


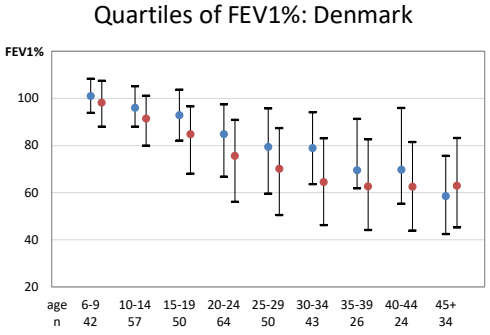






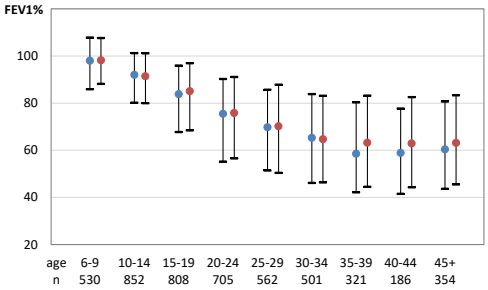




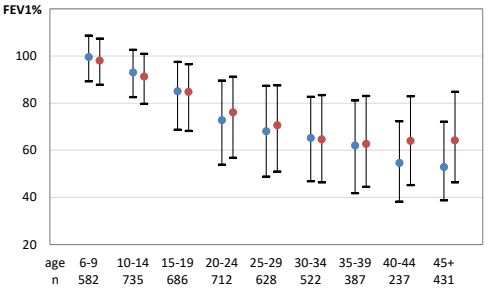


[figure 4.4 continued]

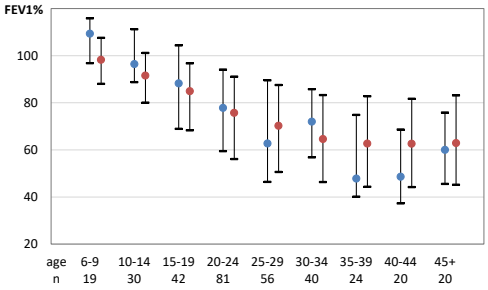
Quartiles of FEV_1%: France



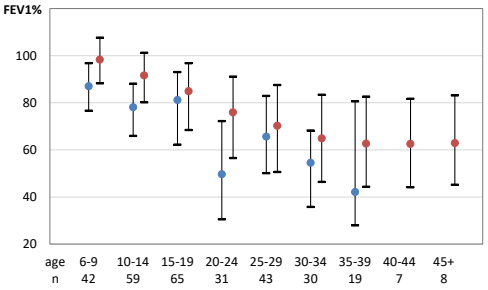
Quartiles of FEV_1%: Germany



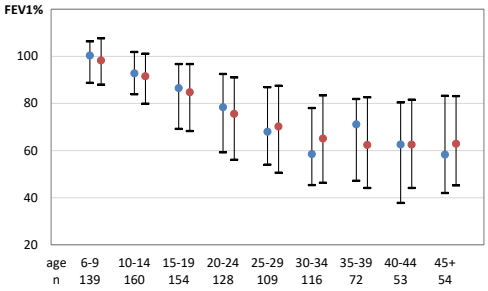
Quartiles of FEV_1%: Greece



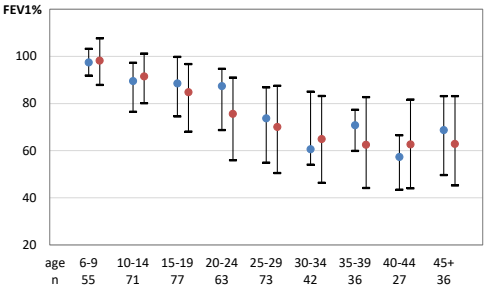
Quartiles of FEV_1%: Hungary



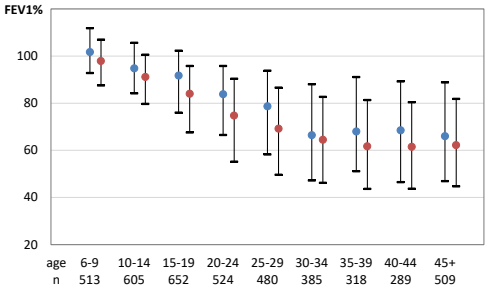
Quartiles of FEV_1%: Ireland



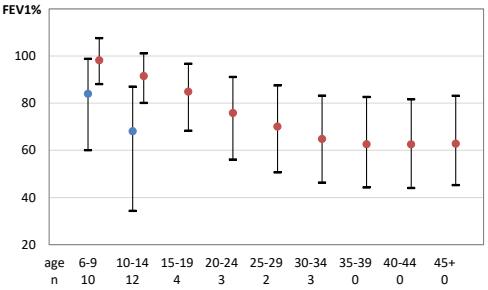
Quartiles of FEV_1%: Israel



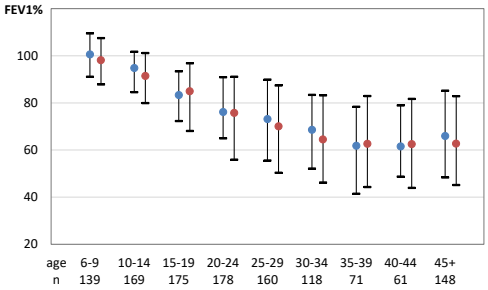
Quartiles of FEV_1%: Italy



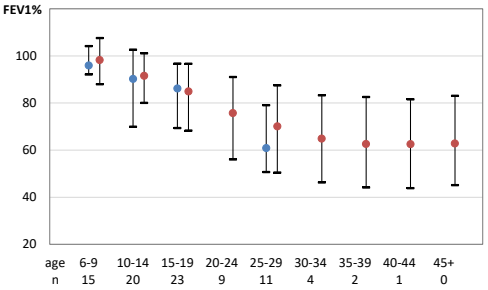
Quartiles of FEV_1%: Rep of Moldova



Quartiles of FEV_1%: The Netherlands

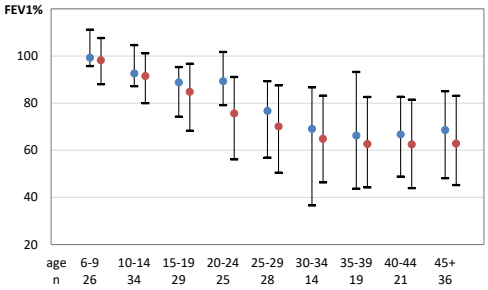


Quartiles of FEV_1%: North Macedonia

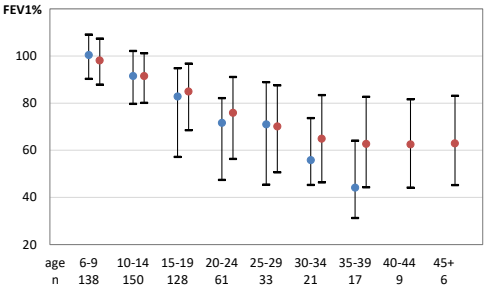


[figure 4.4 continued]

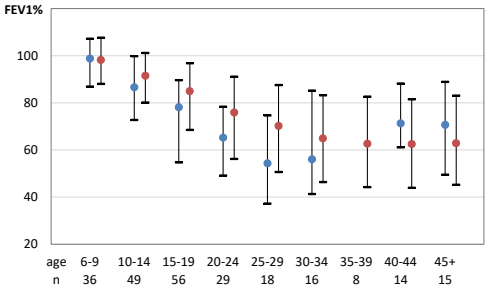
Quartiles of FEV_1%: Norway



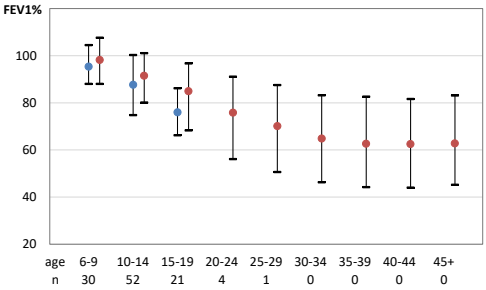
Quartiles of FEV_1%: Poland



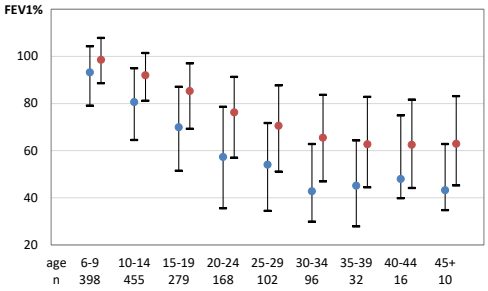
Quartiles of FEV_1%: Portugal



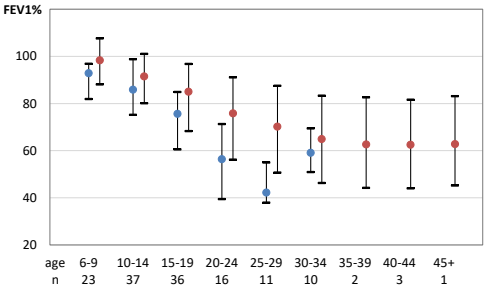
Quartiles of FEV_1%: Romania



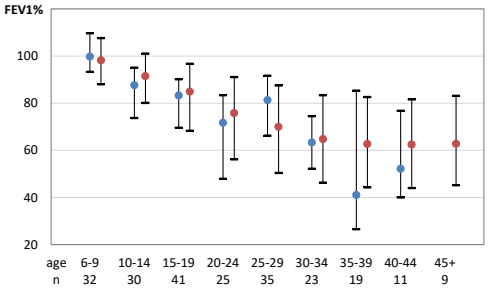
Quartiles of FEV_1%: Russian Federation



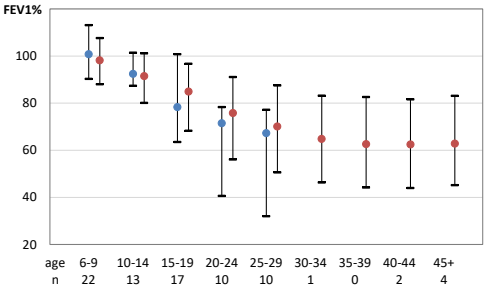
Quartiles of FEV_1%: Serbia



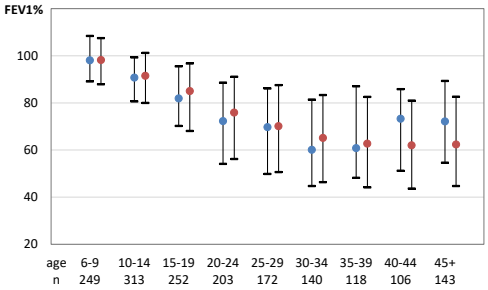
Quartiles of FEV_1%: Slovak Republic



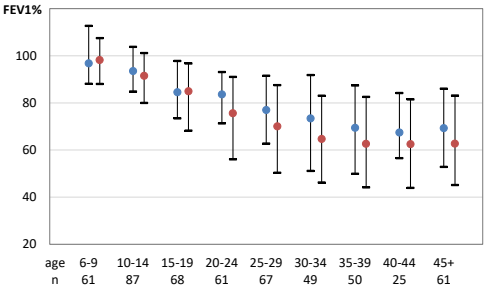
Quartiles of FEV_1%: Slovenia



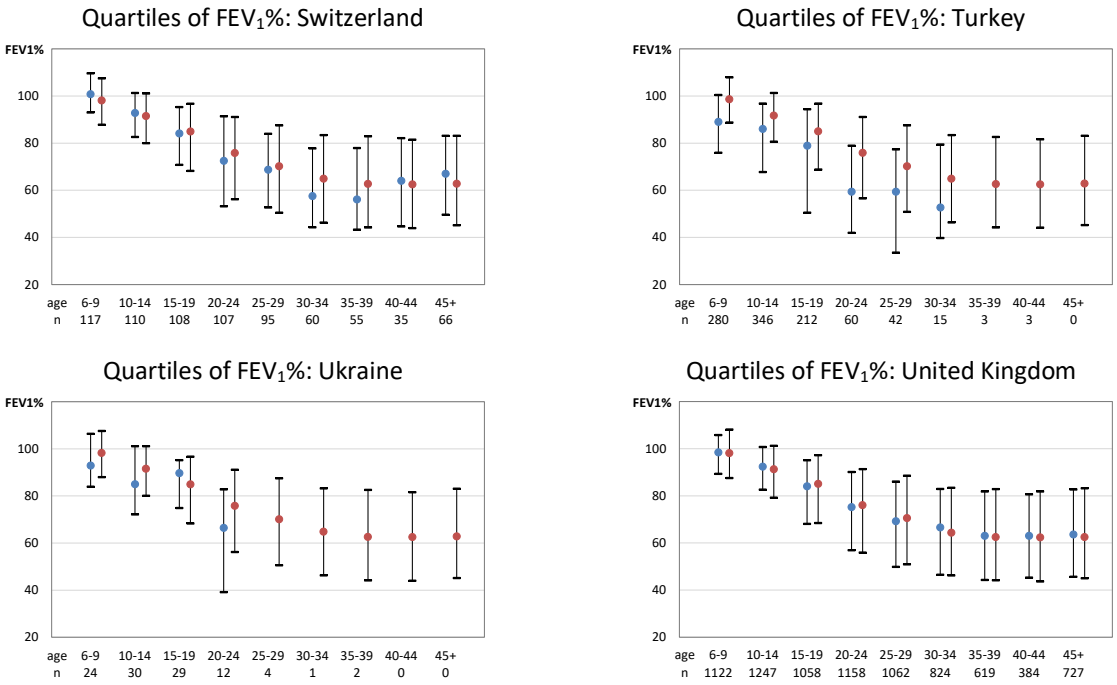
Quartiles of FEV_1%: Spain



Quartiles of FEV_1%: Sweden

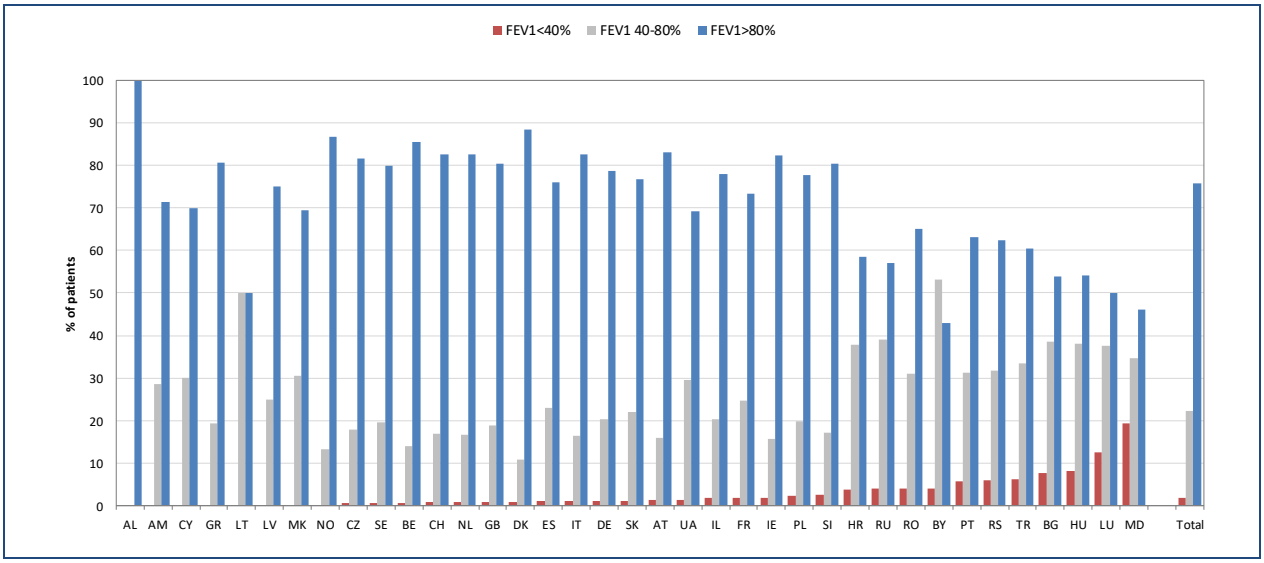


[figure 4.4 continued]



Note: The United Kingdom reports best FEV1 from the annual review, which is the time period between data sets and is not necessarily a calendar year. Therefore, in some cases measurement of FEV1 could be dated in the previous calendar year.

Figure 4.5 FEV_1% of predicted according to severity group and age group, by country and overall. Patients aged 6-17 years who have never had a transplant.

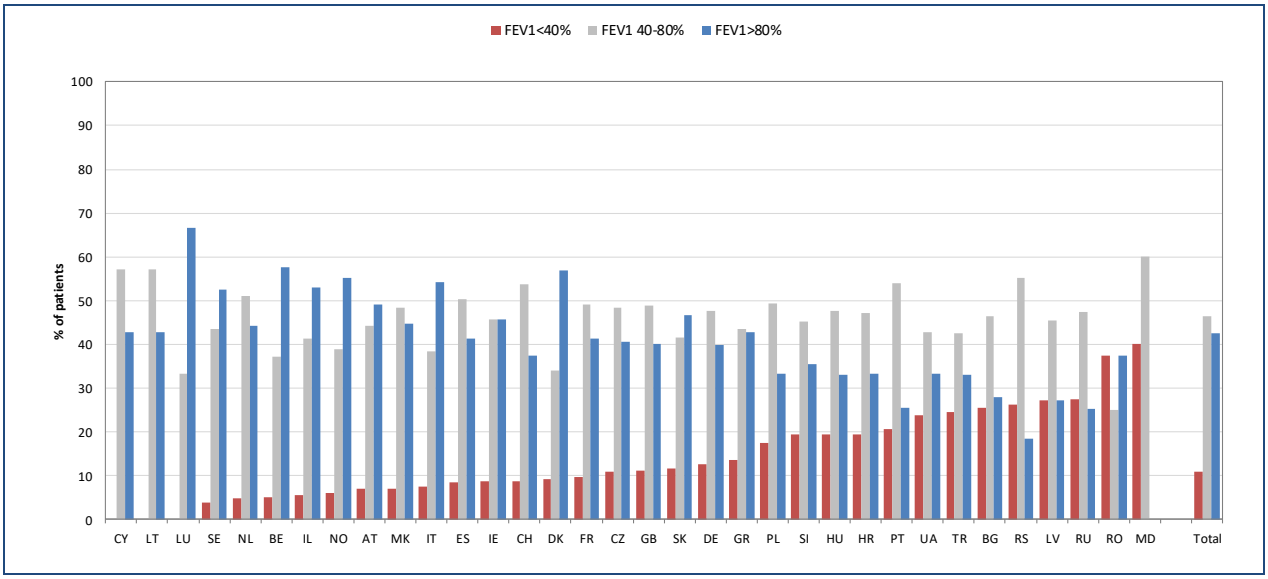


Note: Georgia has <5 patients aged 6-17 years at FEV_1 measurement and is excluded from the graph.

Note: The United Kingdom reports best FEV1 from the annual review, which is the time period between data sets and is not necessarily a calendar year. Therefore, in some cases measurement of FEV1 could be from the previous calendar year.

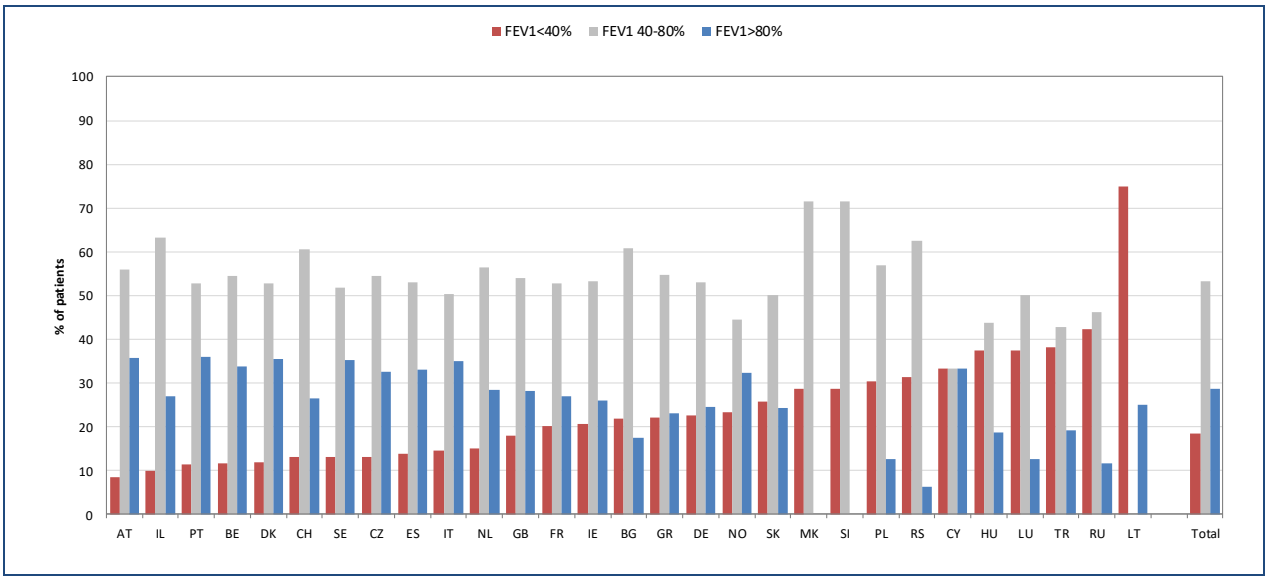
Figures 4.5, 4.6 and 4.7 show the FEV_1% by severity group, by country and overall. Patients with an FEV_1% higher than 80% are generally considered to have mild lung disease, patients with FEV_1% between 80% and 40% moderate lung disease, and patients with FEV_1 <40% severe lung disease. However, since a 10 year old child with a lung function of 50% has considerably worse lung disease than a 50 year old patient with the same FEV_1%, 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.

Figure 4.6 FEV_1% of predicted according to severity group and age group, by country and overall. Patients aged 18-29 years who have never had a transplant.



Note: Georgia has 0% coverage for adults and is excluded from the graph.
Note: Albania, Armenia and Belarus have <5 patients aged 18-29 years at FEV_1 measurement and are excluded from the graph.
Note: The United Kingdom reports best FEV1 from the annual review, which is the time period between data sets and is not necessarily a calendar year. Therefore, in some cases measurement of FEV1 could be from the previous calendar year.

Figure 4.7 FEV_1% of predicted according to severity group and age group, by country and overall. Patients aged 30 years or older who have never had a transplant.



Note: Georgia has 0% coverage for adults and is excluded from the graph.
Note: Albania, Armenia, Belarus, Croatia, Latvia, Rep of Moldova, Romania and Ukraine have <5 patients aged 30 years or more at FEV_1 measurement and are excluded from the graph.
Note: The United Kingdom reports best FEV1 from the annual review, which is the time period between data sets and is not necessarily a calendar year. Therefore, in some cases measurement of FEV1 could be from the previous calendar year.

5. Microbiology

We collect data on three chronic infections – *Pseudomonas aeruginosa*, *Burkholderia cepacia complex species* and *Staphylococcus aureus* – as well as the occurrence of *non-tuberculous mycobacteria* (NTM), *Stenotrophomonas maltophilia, Achromobacter species, Haemophilus Influenzae* and methicillin-resistant *Staphylococcus aureus* (MRSA).

In the microbiology category, discrepancies exist between the ECFSPR definition of chronicity and those of the national registries. The ECFSPR definition of chronic infection (see Appendix 3, page 156) is:

Patient 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 think the status has changed, when:

a. >50% of respiratory samples collected during the last 12 months are positive; at least 4 samples collected during that period (modified Leeds criteria for chronic infection);
and/or
b. 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.

Table 5.1 Prevalence of bacterial infection in children seen in 2019 who have never had a transplant, by country and overall.

Country	Chronic *Pseudomonas aeruginosa* number (%)			Chronic *Burkholderia cepacia complex species* number (%)			*Haemophilus influenzae* number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	3 (3.09)	64 (65.98)	30 (30.93)	2 (2.06)	95 (97.94)	0 (0.00)	4 (4.12)	93 (95.88)	0 (0.00)
Armenia	0 (0.00)	14 (58.33)	10 (41.67)	8 (33.33)	16 (66.67)	0 (0.00)	0 (0)	24 (100)	0 (0)
Austria	1 (0.27)	347 (92.28)	28 (7.45)	3 (0.80)	371 (98.67)	2 (0.53)	1 (0.27)	230 (61.17)	145 (38.56)
Belarus	0 (0.00)	109 (74.15)	38 (25.85)	0 (0.00)	143 (97.28)	4 (2.72)	0 (0.00)	138 (93.88)	9 (6.12)
Belgium	0 (0.00)	427 (92.83)	33 (7.17)	0 (0.00)	455 (98.91)	5 (1.09)	0 (0.00)	271 (58.91)	189 (41.09)
Bulgaria	0 (0.00)	74 (66.67)	37 (33.33)	2 (1.80)	109 (98.20)	0 (0.00)	1 (0.90)	105 (94.60)	5 (4.50)
Croatia	11 (12.22)	67 (74.45)	12 (13.33)	11 (12.22)	79 (87.78)	0 (0.00)	9 (10.00)	70 (77.78)	11 (12.22)
Cyprus	0 (0.00)	9 (69.23)	4 (30.77)	0 (0)	13 (100)	0 (0)	0 (0.00)	3 (23.08)	10 (76.92)
Czech Republic	4 (1.23)	301 (92.62)	20 (6.15)	4 (1.23)	317 (97.54)	4 (1.23)	4 (1.23)	248 (76.31)	73 (22.46)
Denmark	0 (0.00)	183 (91.96)	16 (8.04)	0 (0.00)	197 (98.99)	2 (1.01)	0 (0.00)	86 (43.22)	113 (56.78)
France	0 (0.00)	2501 (92.12)	214 (7.88)	0 (0.00)	2700 (99.45)	15 (0.55)	0 (0.00)	2035 (74.95)	680 (25.05)
Georgia	0 (0.00)	50 (69.44)	22 (30.56)	0 (0)	72 (100)	0 (0)	72 (100)	0 (0)	0 (0)
Germany	32 (1.24)	2301 (89.01)	252 (9.75)	28 (1.08)	2540 (98.26)	17 (0.66)	27 (1.04)	1788 (69.17)	770 (29.79)
Greece	1 (0.99)	86 (85.15)	14 (13.86)	0 (0)	101 (100)	0 (0)	0 (0)	101 (100)	0 (0)
Hungary	0 (0.00)	177 (70.80)	73 (29.20)	1 (0.40)	246 (98.40)	3 (1.20)	250 (100)	0 (0)	0 (0)
Ireland[1]	0 (0.00)	471 (94.01)	30 (5.99)	0 (0.00)	493 (98.40)	8 (1.60)	0 (0.00)	338 (67.47)	163 (32.53)
Israel	7 (3.59)	150 (76.92)	38 (19.49)	9 (4.62)	186 (95.38)	0 (0.00)	10 (5.13)	143 (73.33)	42 (21.54)
Italy[2]	141 (6.14)	1944 (84.59)	213 (9.27)	141 (6.14)	2157 (93.86)	0 (0.00)	5 (0.22)	1813 (78.89)	480 (20.89)
Latvia	20 (62.50)	8 (25.00)	4 (12.50)	23 (71.87)	9 (28.13)	0 (0)	7 (21.88)	15 (46.87)	10 (31.25)
Lithuania	1 (7.14)	10 (71.43)	3 (21.43)	0 (0.00)	13 (92.86)	1 (7.14)	0 (0.00)	13 (92.86)	1 (7.14)
Luxembourg	0 (0.00)	11 (73.33)	4 (26.67)	0 (0)	15 (100)	0 (0)	0 (0.00)	9 (60.00)	6 (40.00)
Moldova	0 (0.00)	18 (40.91)	26 (59.09)	44 (100)	0 (0)	0 (0)	0 (0)	43 (97.73)	1 (2.27)
The Netherlands	5 (0.91)	482 (87.64)	63 (11.45)	5 (0.91)	540 (98.18)	5 (0.91)	5 (0.91)	398 (72.36)	147 (26.73)

[1] Ireland: chronicity for *Pseudomonas aeruginosa* and *Burkholderia cepacia complex species* is defined as: at least 3 or more positive isolates during the last 12 months preceding the last reported culture in 2019.

[2] Italy: chronicity for *Pseudomonas aeruginosa* and *Burkholderia cepacia complex species* is defined as: at least 3 or more positive cultures during 2019.

[table 5.1 continued]

Country	Chronic *Pseudomonas aeruginosa* number (%)			Chronic *Burkholderia cepacia complex species* number (%)			*Haemophilus influenzae* number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
North Macedonia	1 (1.12)	64 (71.91)	24 (26.97)	0 (0.00)	87 (97.75)	2 (2.25)	0 (0.00)	87 (97.75)	2 (2.25)
Norway	3 (2.68)	106 (94.64)	3 (2.68)	1 (0.89)	110 (98.22)	1 (0.89)	0 (0.00)	55 (49.11)	57 (50.89)
Poland	8 (0.96)	721 (86.14)	108 (12.90)	6 (0.72)	826 (98.68)	5 (0.60)	6 (0.72)	663 (79.21)	168 (20.07)
Portugal	4 (2.13)	146 (77.66)	38 (20.21)	3 (1.60)	180 (95.74)	5 (2.66)	5 (2.66)	106 (56.38)	77 (40.96)
Romania	2 (0.93)	145 (67.44)	68 (31.63)	5 (2.33)	210 (97.67)	0 (0.00)	71 (33.02)	135 (62.79)	9 (4.19)
Russian Federation	71 (3.02)	1653 (70.31)	627 (26.67)	71 (3.02)	2210 (94.00)	70 (2.98)	133 (5.66)	2060 (87.62)	158 (6.72)
Serbia	1 (0.81)	85 (69.11)	37 (30.08)	2 (1.63)	113 (91.87)	8 (6.50)	2 (1.63)	48 (39.02)	73 (59.35)
Slovak Republic	0 (0.00)	82 (63.08)	48 (36.92)	2 (1.54)	127 (97.69)	1 (0.77)	4 (3.08)	116 (89.23)	10 (7.69)
Slovenia	1 (1.82)	42 (76.36)	12 (21.82)	0 (0)	55 (100)	0 (0)	0 (0.00)	24 (43.64)	31 (56.36)
Spain	12 (1.11)	937 (86.84)	130 (12.05)	17 (1.58)	1044 (96.75)	18 (1.67)	9 (0.83)	758 (70.25)	312 (28.92)
Sweden	5 (1.91)	227 (86.64)	30 (11.45)	0 (0.00)	260 (99.24)	2 (0.76)	0 (0.00)	186 (70.99)	76 (29.01)
Switzerland	7 (1.67)	365 (87.11)	47 (11.22)	8 (1.91)	409 (97.61)	2 (0.48)	6 (1.43)	270 (64.44)	143 (34.13)
Turkey	14 (0.80)	1421 (81.58)	307 (17.62)	15 (0.86)	1722 (98.85)	5 (0.29)	1 (0.06)	1556 (89.32)	185 (10.62)
Ukraine	2 (1.65)	71 (58.68)	48 (39.67)	4 (3.31)	117 (96.69)	0 (0.00)	3 (2.48)	111 (91.73)	7 (5.79)
United Kingdom[3]	5 (0.12)	3978 (93.82)	257 (6.06)	5 (0.12)	4173 (98.42)	62 (1.46)	4 (0.09)	3239 (76.40)	997 (23.51)
Total	362 (1.56)	19847 (85.63)	2968 (12.81)	420 (1.81)	22510 (97.12)	247 (1.07)	639 (2.76)	17378 (74.98)	5160 (22.26)

[3] United Kingdom: chronicity for *Pseudomonas aeruginosa* is defined as: 3 or more positive isolates during the last 12 months. Information on *Burkholderia cepacia complex species* is collected as: *Burkholderia cepacia complex species* grown since last annual review, not necessarily chronic.

Table 5.1 shows, separately by country, the frequency of chronic *Pseudomonas aeruginosa*, chronic *Burkholderia cepacia complex species* and *Haemophilus influenzae* in children. The number of missing values is also included. The identification rate of *Burkholderia cepacia complex species* in particular may also be influenced by differences in culture techniques employed.

Table 5.2 Prevalence of bacterial infection in adults seen in 2019 who have never had a transplant, by country and overall.

Country	Chronic *Pseudomonas aeruginosa* number (%)			Chronic *Burkholderia cepacia complex species* number (%)			*Haemophilus influenzae* number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	0 (0.00)	6 (66.67)	3 (33.33)	1 (11.11)	8 (88.89)	0 (0.00)	1 (11.11)	8 (88.89)	0 (0.00)
Austria	1 (0.29)	185 (54.10)	156 (45.61)	3 (0.88)	320 (93.56)	19 (5.56)	1 (0.29)	271 (79.24)	70 (20.47)
Belarus	0 (0.00)	5 (62.50)	3 (37.50)	0 (0)	8 (100)	0 (0)	0 (0)	8 (100)	0 (0)
Belgium	1 (0.15)	424 (64.83)	229 (35.02)	1 (0.15)	634 (96.94)	19 (2.91)	1 (0.15)	563 (86.09)	90 (13.76)
Bulgaria	0 (0.00)	23 (30.67)	52 (69.33)	0 (0.00)	74 (98.67)	1 (1.33)	0 (0.00)	71 (94.67)	4 (5.33)
Croatia	2 (4.88)	8 (19.51)	31 (75.61)	2 (4.88)	39 (95.12)	0 (0.00)	0 (0)	41 (100)	0 (0)
Cyprus	0 (0.00)	9 (64.29)	5 (35.71)	2 (14.29)	12 (85.71)	0 (0.00)	2 (14.29)	2 (14.29)	10 (71.42)
Czech Republic	7 (2.61)	181 (67.54)	80 (29.85)	8 (2.99)	225 (83.96)	35 (13.06)	10 (3.73)	242 (90.30)	16 (5.97)
Denmark	2 (0.74)	160 (59.26)	108 (40.00)	2 (0.74)	245 (90.74)	23 (8.52)	2 (0.74)	207 (76.67)	61 (22.59)
France	0 (0.00)	2008 (64.73)	1094 (35.27)	0 (0.00)	3030 (97.68)	72 (2.32)	0 (0.00)	2642 (85.17)	460 (14.83)
Germany	106 (3.17)	1526 (45.69)	1708 (51.14)	102 (3.05)	3140 (94.02)	98 (2.93)	93 (2.78)	2970 (88.93)	277 (8.29)
Greece	11 (3.69)	72 (24.16)	215 (72.15)	14 (4.70)	284 (95.30)	0 (0.00)	12 (4.03)	274 (91.94)	12 (4.03)
Hungary	4 (2.05)	91 (46.67)	100 (51.28)	6 (3.08)	182 (93.33)	7 (3.59)	195 (100)	0 (0)	0 (0)
Ireland[1]	2 (0.31)	371 (58.25)	264 (41.44)	2 (0.31)	618 (97.02)	17 (2.67)	2 (0.31)	560 (87.92)	75 (11.77)
Israel	11 (3.46)	128 (40.25)	179 (56.29)	15 (4.72)	298 (93.71)	5 (1.57)	13 (4.09)	275 (86.48)	30 (9.43)
Italy[2]	207 (6.95)	1538 (51.67)	1232 (41.38)	205 (6.89)	2693 (90.46)	79 (2.65)	16 (0.54)	2791 (93.75)	170 (5.71)
Latvia	4 (30.77)	3 (23.08)	6 (46.15)	7 (53.85)	6 (46.15)	0 (0.00)	0 (0.00)	11 (84.62)	2 (15.38)
Lithuania	0 (0.00)	11 (84.62)	2 (15.38)	0 (0)	13 (100)	0 (0)	0 (0.00)	11 (84.62)	2 (15.38)
Luxembourg	1 (4.76)	13 (61.91)	7 (33.33)	1 (4.76)	17 (80.95)	3 (14.29)	2 (9.52)	12 (57.15)	7 (33.33)
Moldova	1 (11.11)	1 (11.11)	7 (77.78)	9 (100)	0 (0)	0 (0)	1 (11.11)	8 (88.89)	0 (0.00)
The Netherlands	43 (5.09)	399 (47.28)	402 (47.63)	44 (5.21)	782 (92.66)	18 (2.13)	45 (5.33)	700 (82.94)	99 (11.73)

[1] Ireland: chronicity for *Pseudomonas aeruginosa* and *Burkholderia cepacia complex species* is defined as: at least 3 or more positive isolates during the last 12 months preceding the last reported culture in 2019.

[2] Italy: chronicity for *Pseudomonas aeruginosa* and *Burkholderia cepacia complex species* is defined as: at least 3 or more positive cultures during 2019.

[table 5.2 continued]

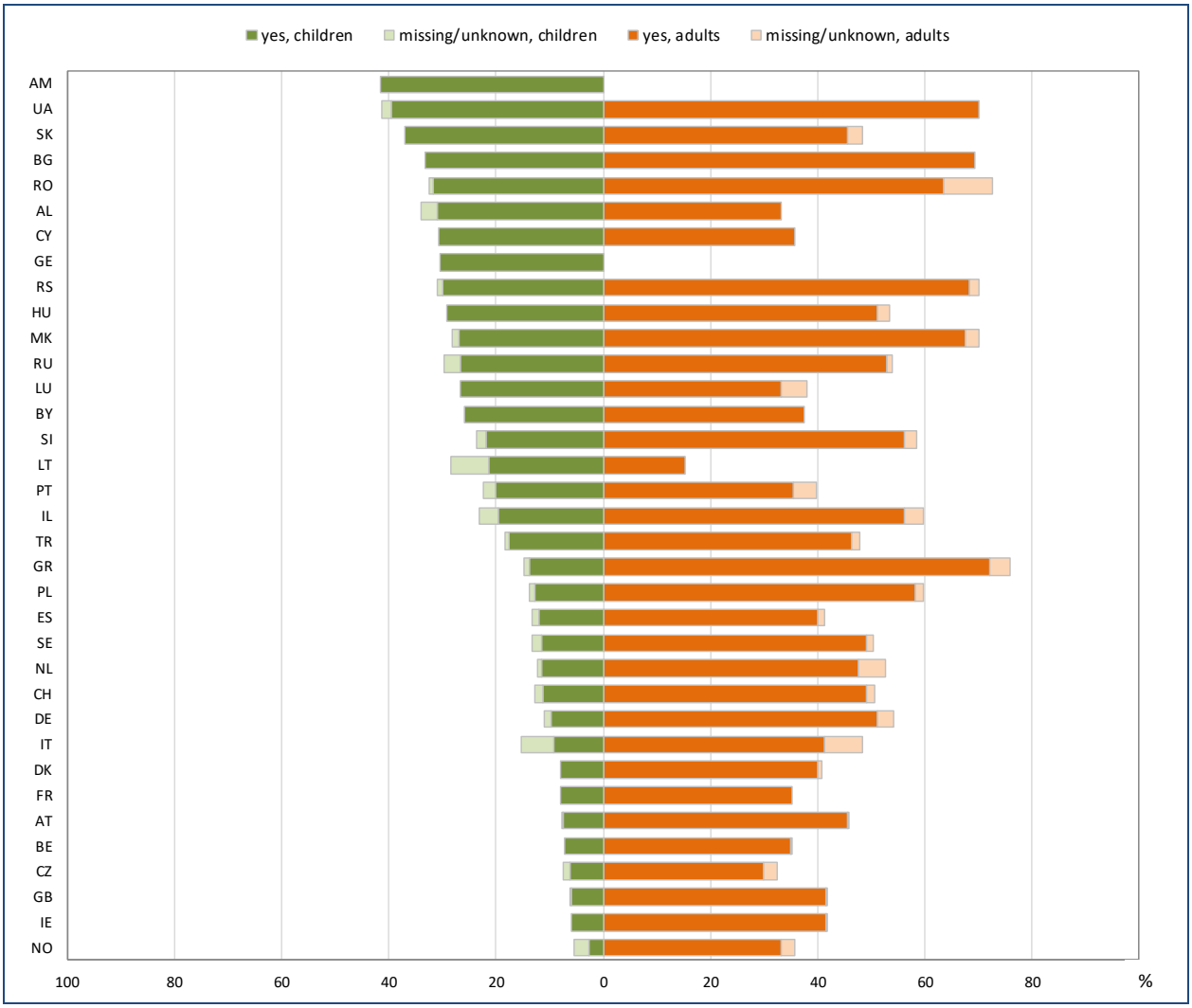
Country	Chronic *Pseudomonas aeruginosa* number (%)			Chronic *Burkholderia cepacia complex species* number (%)			*Haemophilus influenzae* number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
North Macedonia	1 (2.50)	12 (30.00)	27 (67.50)	1 (2.50)	39 (97.50)	0 (0.00)	0 (0)	40 (100)	0 (0)
Norway	4 (2.42)	106 (64.25)	55 (33.33)	7 (4.24)	154 (93.34)	4 (2.42)	4 (2.42)	127 (76.97)	34 (20.61)
Poland	5 (1.48)	136 (40.36)	196 (58.16)	6 (1.78)	314 (93.18)	17 (5.04)	6 (1.78)	311 (92.29)	20 (5.93)
Portugal	6 (4.35)	83 (60.14)	49 (35.51)	5 (3.62)	124 (89.86)	9 (6.52)	5 (3.62)	110 (79.71)	23 (16.67)
Romania	1 (9.09)	3 (27.27)	7 (63.64)	0 (0.00)	10 (90.91)	1 (9.09)	1 (9.09)	10 (90.91)	0 (0.00)
Russian Federation	7 (0.92)	353 (46.14)	405 (52.94)	10 (1.31)	669 (87.45)	86 (11.24)	37 (4.84)	716 (93.59)	12 (1.57)
Serbia	1 (1.67)	18 (30.00)	41 (68.33)	1 (1.67)	52 (86.66)	7 (11.67)	1 (1.67)	34 (56.66)	25 (41.67)
Slovak Republic	4 (2.68)	77 (51.68)	68 (45.64)	5 (3.36)	134 (89.93)	10 (6.71)	5 (3.36)	134 (89.93)	10 (6.71)
Slovenia	1 (2.44)	17 (41.46)	23 (56.10)	1 (2.44)	39 (95.12)	1 (2.44)	1 (2.44)	36 (87.80)	4 (9.76)
Spain	13 (1.26)	605 (58.62)	414 (40.12)	22 (2.13)	939 (90.99)	71 (6.88)	23 (2.23)	907 (87.89)	102 (9.88)
Sweden	4 (1.14)	175 (49.71)	173 (49.15)	0 (0.00)	343 (97.44)	9 (2.56)	0 (0.00)	308 (87.50)	44 (12.50)
Switzerland	7 (1.43)	241 (49.39)	240 (49.18)	11 (2.25)	457 (93.65)	20 (4.10)	11 (2.25)	392 (80.33)	85 (17.42)
Turkey	4 (1.67)	125 (52.08)	111 (46.25)	3 (1.25)	231 (96.25)	6 (2.50)	1 (0.42)	199 (82.91)	40 (16.67)
Ukraine	0 (0.00)	9 (30.00)	21 (70.00)	0 (0.00)	29 (96.67)	1 (3.33)	1 (3.33)	28 (93.34)	1 (3.33)
United Kingdom[3]	3 (0.05)	3194 (58.51)	2262 (41.44)	3 (0.05)	5180 (94.89)	276 (5.06)	3 (0.05)	5064 (92.77)	392 (7.18)
Total	464 (2.04)	12316 (54.12)	9976 (43.84)	499 (2.19)	21343 (93.79)	914 (4.02)	495 (2.18)	20084 (88.25)	2177 (9.57)

[3] United Kingdom: chronicity for *Pseudomonas aeruginosa* is defined as: 3 or more positive isolates during the last 12 months. Information on *Burkholderia cepacia complex species* is collected as follows: *Burkholderia cepacia complex species* grown since last annual review, not necessarily chronic.

Note: Georgia have 0% coverage for adults and is excluded from the table.
Note: Armenia has <5 patients aged 18 years or more at 31/12/2019 and is not shown in this table, but is considered in the total.

Table 5.2 shows, separately by country, the frequency of chronic *Pseudomonas aeruginosa*, chronic *Burkholderia cepacia complex species* and *Haemophilus influenza* in adults. The number of missing values is also included.

Figure 5.1 Prevalence of chronic Pseudomonas aeruginosa infection in children and adults seen in 2019 who have never had a transplant, by country.

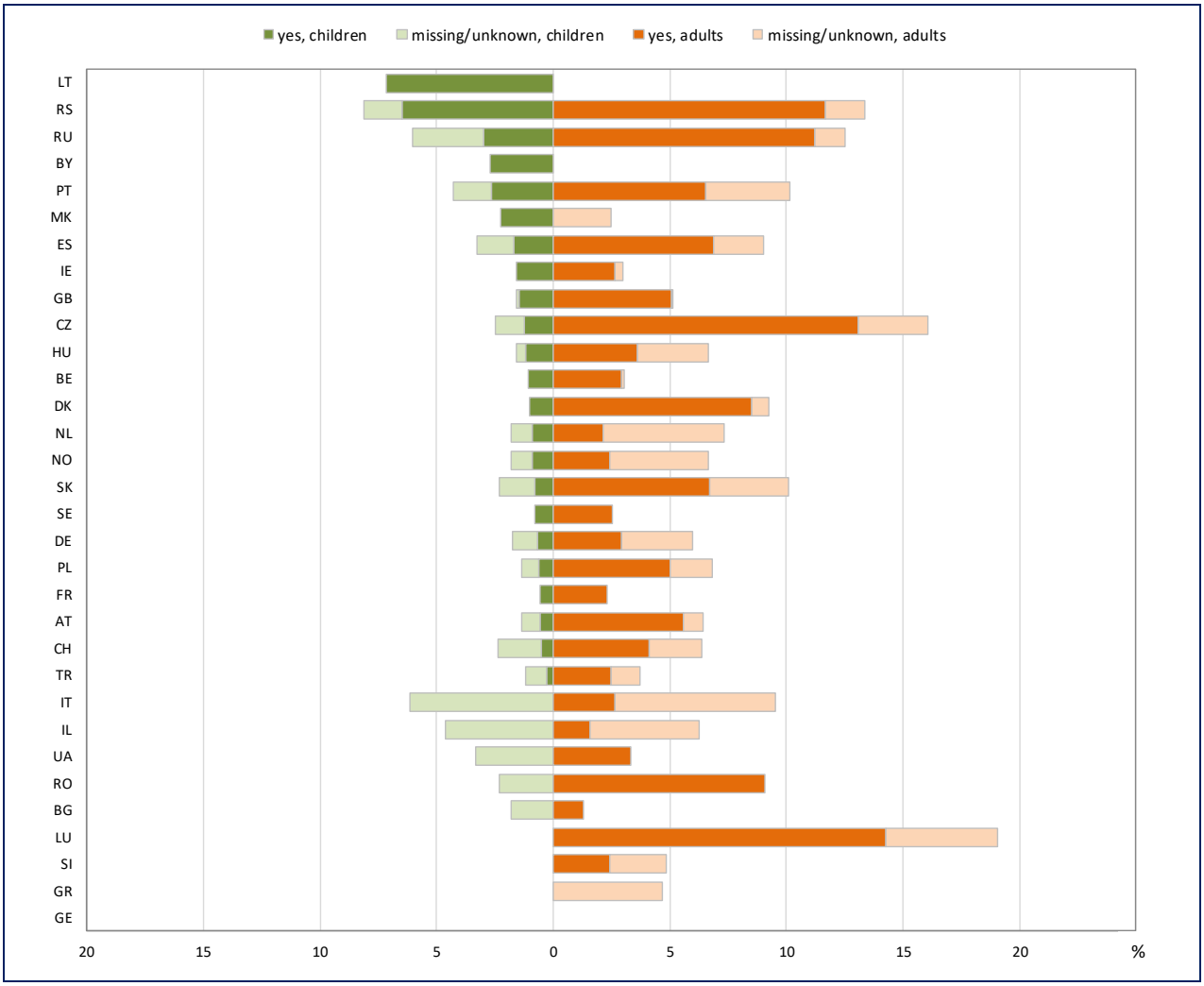


Note: We excluded from the graph the countries for which the information on *Pseudomonas aeruginosa* was missing for more than 10% of the children and/or adults.
Georgia has 0% coverage for adults and the adults bar is excluded from the graph.
Armenia has <5 patients aged 18 years or more at 31/12/2019 and the adults bar is excluded from the graph.

Note: Ireland: 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 2019.
Italy: chronicity for *Pseudomonas aeruginosa* is defined as: at least 3 or more positive cultures during 2019.
United Kingdom: for chronic *Pseudomonas aeruginosa* the definition is: 3 or more positive isolates during the last 12 months.

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 was 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. This is a frequent infection, but prevalence varies considerably between countries.

Figure 5.2 Prevalence of chronic Burkholderia cepacia complex species infection in children and adults seen in 2019 who have never had a transplant, by country.

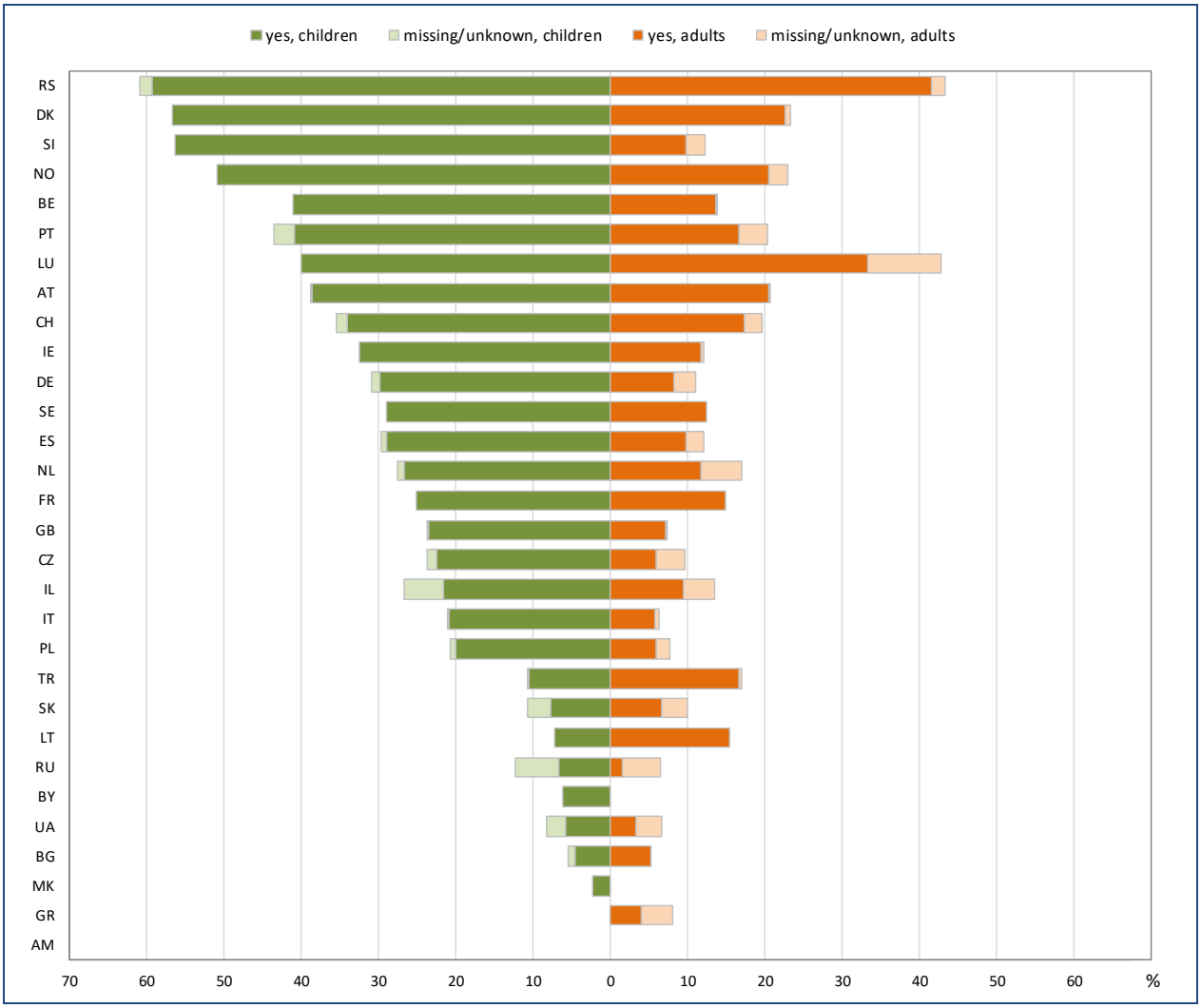


Note: We excluded from the graph the countries for which the information on *Burkholderia cepacia complex species* was missing for more than 10% of the children and/or adults.
Georgia have 0% coverage for adults and the adults bar is excluded from the graph.
Armenia has <5 patients aged 18 years or more at 31/12/2019 and the adults bar is excluded from the graph.

Note: Ireland: chronicity for *Burkholderia cepacia complex species* is defined as: at least 3 or more positive isolates during the last 12 months preceding the last reported culture in 2019.
Italy: chronicity for *Burkholderia cepacia complex species* is defined as: at least 3 or more positive cultures during 2019.
United Kingdom: information on *Burkholderia cepacia complex species* is collected as: *Burkholderia cepacia complex species* grown since last annual review, not necessarily chronic.

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 chronic *Burkholderia cepacia complex species* infection was 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. This infection is much less frequent than *Pseudomonas aeruginosa* (note the different scale on the horizontal axis), and there is also some variation among countries.

Figure 5.3 Prevalence of Haemophilus influenzae infection in children and adults seen in 2019 who have never had a transplant, by country.



Note: We excluded from the graph the countries for which the information on *Haemophilus influenzae* was missing for more than 10% of the children and/or adults.
Georgia has 0% coverage for adults and the adults bar is excluded from the graph.
Armenia has <5 patients aged 18 years or more at 31/12/2019 and the adults bar is excluded from the graph.

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 was 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. This infection is as frequent as chronic *Pseudomonas aeruginosa* infection and a similar degree of variation between the countries can be observed.

Table 5.3 Prevalence of chronic Staphylococcus aureus and methicillin-resistant Staphylococcus aureus (MRSA) in children seen in 2019 who have never had a transplant, by country and overall.

Country	Chronic *Staphylococcus aureus* number (%)			MRSA number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	5 (5.15)	59 (60.83)	33 (34.02)	4 (4.12)	93 (95.88)	0 (0.00)
Armenia	0 (0.00)	5 (20.83)	19 (79.17)	16 (66.67)	8 (33.33)	0 (0.00)
Austria	1 (0.27)	139 (36.97)	236 (62.76)	1 (0.27)	370 (98.40)	5 (1.33)
Belarus	0 (0.00)	72 (48.98)	75 (51.02)	0 (0.00)	146 (99.32)	1 (0.68)
Belgium	460 (100)	0 (0)	0 (0)	0 (0.00)	448 (97.39)	12 (2.61)
Bulgaria	0 (0.00)	86 (77.48)	25 (22.52)	0 (0.00)	109 (98.20)	2 (1.80)
Croatia	11 (12.22)	43 (47.78)	36 (40.00)	9 (10.00)	79 (87.78)	2 (2.22)
Cyprus	0 (0.00)	11 (84.62)	2 (15.38)	0 (0.00)	12 (92.31)	1 (7.69)
Czech Republic	3 (0.92)	169 (52.00)	153 (47.08)	4 (1.23)	318 (97.85)	3 (0.92)
Denmark	0 (0.00)	141 (70.85)	58 (29.15)	0 (0.00)	198 (99.50)	1 (0.50)
France	0 (0.00)	1554 (57.24)	1161 (42.76)	0 (0.00)	2600 (95.76)	115 (4.24)
Georgia	0 (0.00)	50 (69.44)	22 (30.56)	2 (2.78)	70 (97.22)	0 (0.00)
Germany	29 (1.12)	1591 (61.55)	965 (37.33)	27 (1.04)	2468 (95.47)	90 (3.48)
Greece	1 (0.99)	61 (60.40)	39 (38.61)	0 (0.00)	83 (82.18)	18 (17.82)
Hungary	0 (0.00)	108 (43.20)	142 (56.80)	1 (0.40)	232 (92.80)	17 (6.80)
Ireland[1]	0 (0.00)	303 (60.48)	198 (39.52)	0 (0.00)	479 (95.61)	22 (4.39)
Israel	7 (3.59)	99 (50.77)	89 (45.64)	9 (4.62)	180 (92.30)	6 (3.08)
Italy[2]	141 (6.14)	1385 (60.27)	772 (33.59)	5 (0.22)	2072 (90.16)	221 (9.62)
Latvia	16 (50.00)	1 (3.13)	15 (46.87)	6 (18.75)	26 (81.25)	0 (0.00)
Lithuania	1 (7.14)	3 (21.43)	10 (71.43)	0 (0)	14 (100)	0 (0)
Luxembourg	0 (0.00)	7 (46.67)	8 (53.33)	0 (0.00)	14 (93.33)	1 (6.67)
Moldova	0 (0.00)	4 (9.09)	40 (90.91)	0 (0)	44 (100)	0 (0)
The Netherlands	5 (0.91)	355 (64.54)	190 (34.55)	5 (0.91)	542 (98.54)	3 (0.55)

[1] Ireland: 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 2019.

[2] Italy: chronicity for *Staphylococcus aureus* is defined as: at least 3 or more positive cultures during 2019.

[table 5.3 continued]

Country	Chronic *Staphylococcus aureus* number (%)			MRSA number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
North Macedonia	0 (0.00)	70 (78.65)	19 (21.35)	0 (0.00)	71 (79.78)	18 (20.22)
Norway	1 (0.89)	55 (49.11)	56 (50.00)	0 (0)	112 (100)	0 (0)
Poland	5 (0.60)	289 (34.53)	543 (64.87)	8 (0.96)	797 (95.22)	32 (3.82)
Portugal	5 (2.66)	101 (53.72)	82 (43.62)	4 (2.13)	165 (87.76)	19 (10.11)
Romania	5 (2.33)	175 (81.39)	35 (16.28)	4 (1.86)	164 (76.28)	47 (21.86)
Russian Federation	57 (2.42)	824 (35.05)	1470 (62.53)	99 (4.21)	2174 (92.47)	78 (3.32)
Serbia	2 (1.63)	31 (25.20)	90 (73.17)	3 (2.44)	106 (86.18)	14 (11.38)
Slovak Republic	4 (3.08)	54 (41.54)	72 (55.38)	1 (0.77)	124 (95.38)	5 (3.85)
Slovenia	0 (0.00)	4 (7.27)	51 (92.73)	0 (0.00)	51 (92.73)	4 (7.27)
Spain	16 (1.48)	635 (58.85)	428 (39.67)	12 (1.11)	1013 (93.89)	54 (5.00)
Sweden	18 (6.87)	179 (68.32)	65 (24.81)	0 (0.00)	256 (97.71)	6 (2.29)
Switzerland	9 (2.15)	175 (41.77)	235 (56.08)	7 (1.67)	404 (96.42)	8 (1.91)
Turkey	13 (0.75)	1291 (74.11)	438 (25.14)	1 (0.06)	1552 (89.09)	189 (10.85)
Ukraine	2 (1.65)	48 (39.67)	71 (58.68)	2 (1.65)	115 (95.04)	4 (3.31)
United Kingdom[3]	5 (0.12)	3769 (88.89)	466 (10.99)	5 (0.12)	4131 (97.43)	104 (2.45)
Total	822 (3.55)	13946 (60.17)	8409 (36.28)	235 (1.01)	21840 (94.24)	1102 (4.75)

[3] United Kingdom: chronicity for *Staphylococcus aureus* is defined as: 3 or more positive isolates during the last 12 months.

Table 5.3 shows, separately by country, the frequency of chronic *Staphylococcus aureus* and methicillin-resistant *Staphylococcus aureus* (MRSA) in children. The number of missing values is also included.

Table 5.4 Prevalence of chronic Staphylococcus aureus, methicillin-resistant Staphylococcus aureus (MRSA) in adults seen in 2019 who have never had a transplant, by country and overall.

Country	Chronic *Staphylococcus aureus* number (%)			MRSA number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	1 (11.11)	2 (22.22)	6 (66.67)	1 (11.11)	8 (88.89)	0 (0.00)
Austria	1 (0.29)	122 (35.67)	219 (64.04)	1 (0.29)	335 (97.96)	6 (1.75)
Belarus	0 (0.00)	3 (37.50)	5 (62.50)	0 (0)	8 (100)	0 (0)
Belgium	654 (100)	0 (0)	0 (0)	1 (0.15)	600 (91.75)	53 (8.10)
Bulgaria	0 (0.00)	57 (76.00)	18 (24.00)	0 (0.00)	74 (98.67)	1 (1.33)
Croatia	2 (4.88)	17 (41.46)	22 (53.66)	0 (0.00)	38 (92.68)	3 (7.32)
Cyprus	2 (14.29)	8 (57.14)	4 (28.57)	2 (14.29)	12 (85.71)	0 (0.00)
Czech Republic	10 (3.73)	130 (48.51)	128 (47.76)	11 (4.10)	243 (90.68)	14 (5.22)
Denmark	2 (0.74)	165 (61.11)	103 (38.15)	0 (0.00)	269 (99.63)	1 (0.37)
France	0 (0.00)	1748 (56.35)	1354 (43.65)	0 (0.00)	2811 (90.62)	291 (9.38)
Germany	103 (3.08)	1652 (49.46)	1585 (47.46)	92 (2.75)	3037 (90.93)	211 (6.32)
Greece	13 (4.36)	158 (53.02)	127 (42.62)	13 (4.36)	245 (82.22)	40 (13.42)
Hungary	4 (2.05)	80 (41.03)	111 (56.92)	5 (2.56)	174 (89.23)	16 (8.21)
Ireland[1]	2 (0.31)	411 (64.53)	224 (35.16)	2 (0.31)	595 (93.41)	40 (6.28)
Israel	11 (3.46)	201 (63.21)	106 (33.33)	12 (3.77)	289 (90.88)	17 (5.35)
Italy[2]	206 (6.92)	1704 (57.24)	1067 (35.84)	16 (0.54)	2710 (91.03)	251 (8.43)
Latvia	7 (53.85)	0 (0.00)	6 (46.15)	0 (0)	13 (100)	0 (0)
Lithuania	0 (0.00)	3 (23.08)	10 (76.92)	0 (0)	13 (100)	0 (0)
Luxembourg	1 (4.76)	8 (38.10)	12 (57.14)	1 (4.76)	18 (85.72)	2 (9.52)
Moldova	1 (11.11)	3 (33.33)	5 (55.56)	1 (11.11)	8 (88.89)	0 (0.00)
The Netherlands	45 (5.33)	394 (46.68)	405 (47.99)	50 (5.92)	788 (93.37)	6 (0.71)

[1] Ireland: 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 2019.

[2] Italy: chronicity for *Staphylococcus aureus* is defined as: at least 3 or more positive cultures during 2019.

[table 5.4 continued]

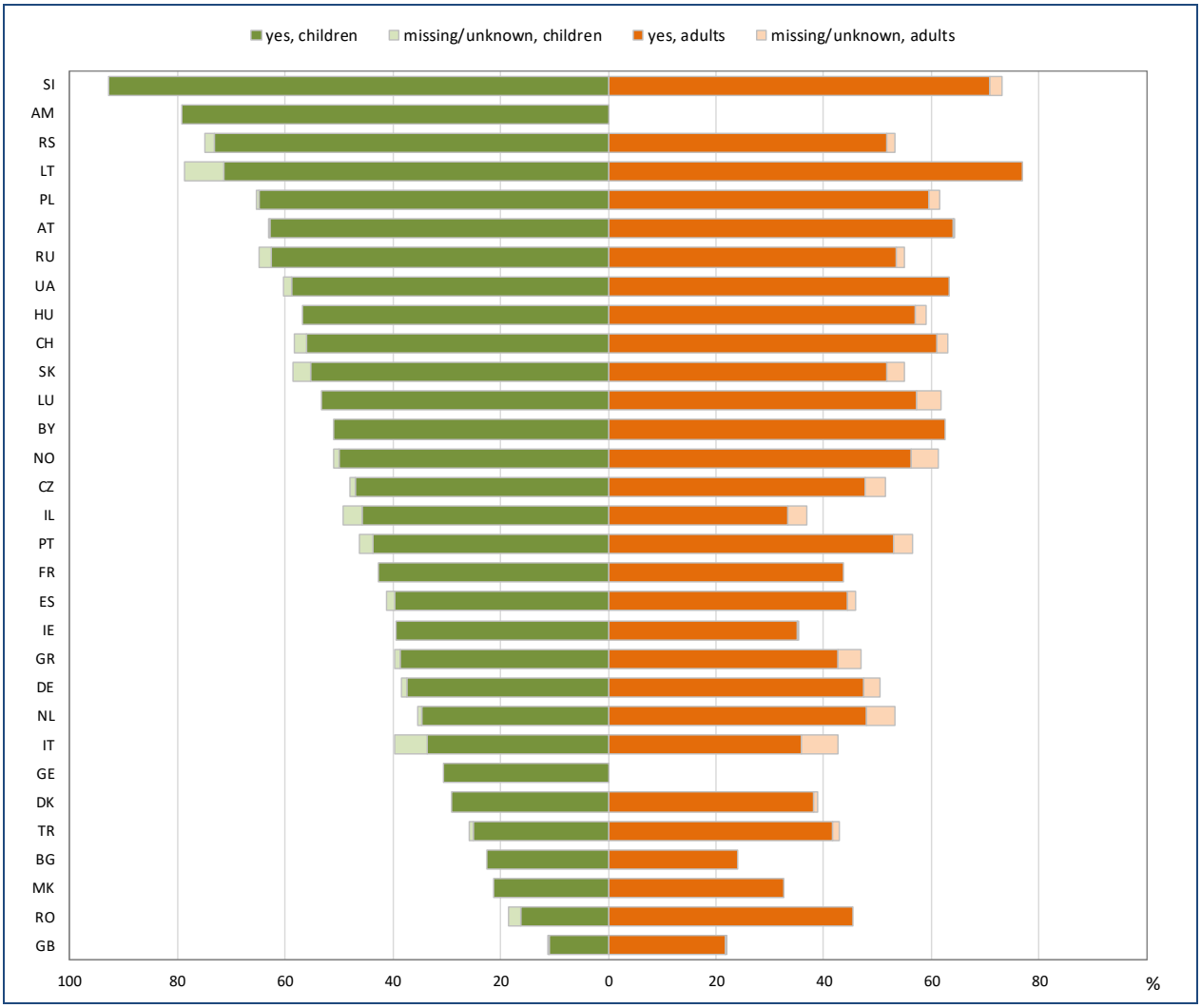
Country	Chronic *Staphylococcus aureus* number (%)			MRSA number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
North Macedonia	0 (0.00)	27 (67.50)	13 (32.50)	0 (0.00)	34 (85.00)	6 (15.00)
Norway	8 (4.85)	64 (38.79)	93 (56.36)	4 (2.42)	159 (96.37)	2 (1.21)
Poland	6 (1.78)	130 (38.58)	201 (59.64)	7 (2.08)	311 (92.28)	19 (5.64)
Portugal	5 (3.62)	60 (43.48)	73 (52.90)	5 (3.62)	121 (87.68)	12 (8.70)
Romania	0 (0.00)	6 (54.55)	5 (45.45)	0 (0.00)	10 (90.91)	1 (9.09)
Russian Federation	11 (1.44)	345 (45.10)	409 (53.46)	29 (3.79)	694 (90.72)	42 (5.49)
Serbia	1 (1.67)	28 (46.67)	31 (51.66)	1 (1.67)	51 (85.00)	8 (13.33)
Slovak Republic	5 (3.36)	67 (44.97)	77 (51.67)	7 (4.70)	132 (88.59)	10 (6.71)
Slovenia	1 (2.44)	11 (26.83)	29 (70.73)	3 (7.32)	36 (87.80)	2 (4.88)
Spain	15 (1.45)	558 (54.07)	459 (44.48)	16 (1.55)	940 (91.09)	76 (7.36)
Sweden	36 (10.23)	166 (47.16)	150 (42.61)	0 (0.00)	350 (99.43)	2 (0.57)
Switzerland	10 (2.05)	180 (36.88)	298 (61.07)	30 (6.15)	451 (92.42)	7 (1.43)
Turkey	3 (1.25)	137 (57.08)	100 (41.67)	1 (0.42)	211 (87.91)	28 (11.67)
Ukraine	0 (0.00)	11 (36.67)	19 (63.33)	1 (3.33)	24 (80.00)	5 (16.67)
United Kingdom[3]	3 (0.05)	4262 (78.08)	1194 (21.87)	3 (0.05)	5313 (97.33)	143 (2.62)
Total	1169 (5.14)	12918 (56.76)	8669 (38.10)	316 (1.39)	21125 (92.83)	1315 (5.78)

[3] United Kingdom: chronicity for *Staphylococcus aureus* is defined as: 3 or more positive isolates during the last 12 months.

Note: Georgia has 0% coverage for adults and is excluded from the table.
Note: Armenia has <5 patients aged 18 years or more at 31/12/2019 and is not shown in this table, but is considered in the total.

Table 5.4 shows, separately by country, the frequency of chronic *Staphylococcus aureus* and methicillin-resistant *Staphylococcus aureus* (MRSA) in adults. The number of missing values is also included.

Figure 5.4 Prevalence of chronic Staphylococcus aureus infection in children and adults seen in 2019 who have never had a transplant, by country.

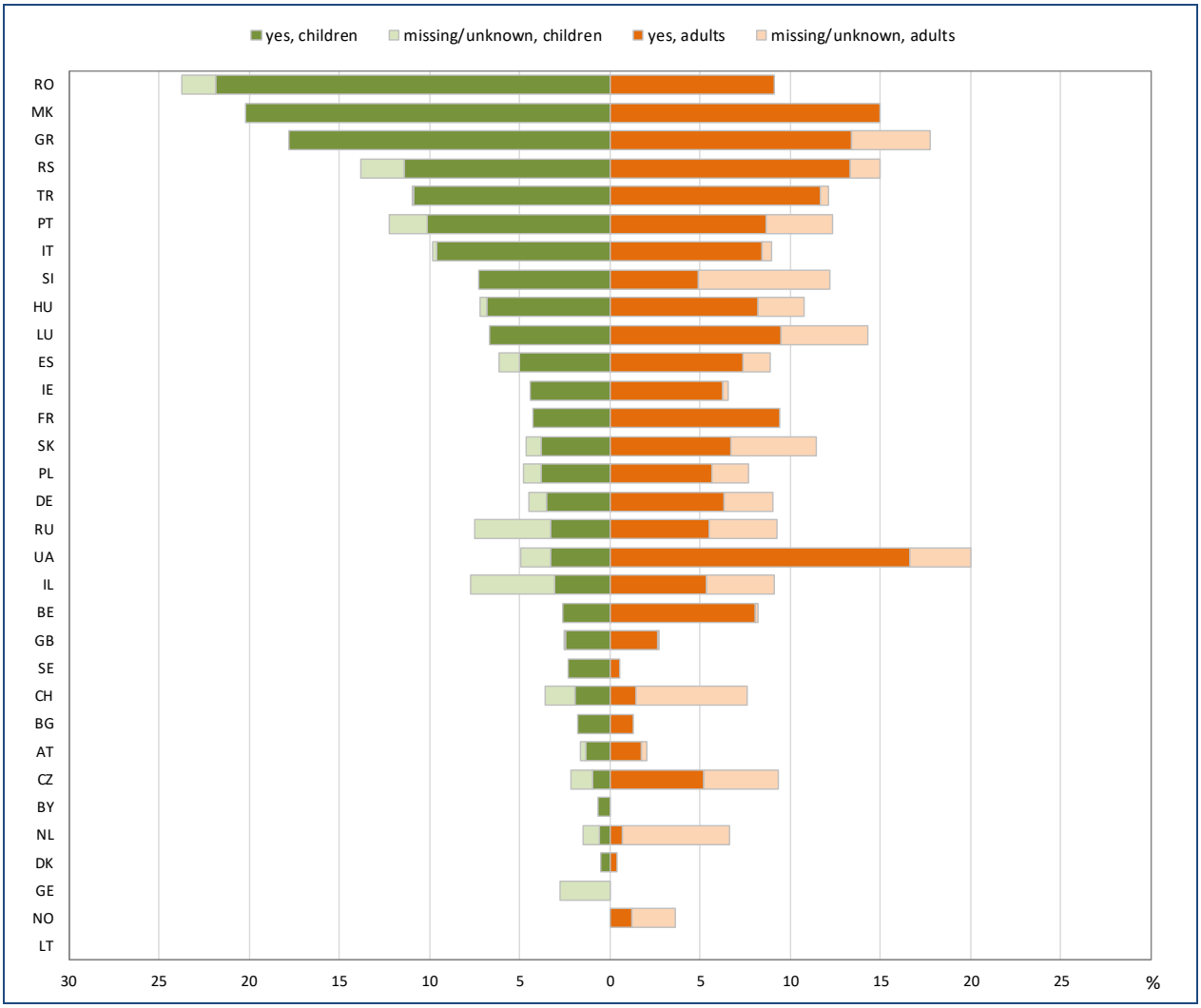


Note: We excluded from the graph the countries for which the information on *Staphylococcus aureus* was missing for more than 10% of the children and/or adults.
Georgia has 0% coverage for adults and the adults bar is excluded from the graph.
Armenia has <5 patients aged 18 years or more at 31/12/2019 and the adults bar is excluded from the graph.

Note: Ireland: 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 2019.
Italy: chronicity for *Staphylococcus aureus* is defined as: at least 3 or more positive cultures during 2019.
United Kingdom: for chronic Staphylococcus aureus the definition is: 3 or more positive isolates during the last 12 months.

This graph represents the percentage of people with chronic *Staphylococcus aureus* infection (in dark colours) and the percentage of people where information on chronic *Staphylococcus aureus* infection was 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. This infection is as frequent as chronic *Pseudomonas aeruginosa* infection and a similar degree of variation between the countries can be observed.

Figure 5.5 Prevalence of methicillin-resistant Staphylococcus aureus (MRSA) infection in children and adults seen in 2019 who have never had a transplant, by country.



Note: We excluded from the graph the countries for which the information on MRSA was missing for more than 10% of the children and/or adults.
Georgia has 0% coverage for adults and the adults bar is excluded from the graph.
Armenia has <5 patients aged 18 years or more at 31/12/2019 and the adults bar is excluded from the graph.

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 was 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. Prevalence of MRSA varies considerably between countries.

Table 5.5 Prevalence of non-tuberculous mycobacteria, Stenotrophomonas maltophilia and Achromobacter species infection in children seen in 2019 who have never had a transplant, by country and overall.

Country	Non-tuberculous mycobacteria (NTM) infection this year number (%)			*Stenotrophomonas maltophilia* infection this year number (%)			*Achromobacter species* infection this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	5 (5.15)	92 (94.85)	0 (0.00)	3 (3.09)	94 (96.91)	0 (0.00)	6 (6.19)	91 (93.81)	0 (0.00)
Armenia	12 (50.00)	12 (50.00)	0 (0.00)	20 (83.33)	4 (16.67)	0 (0.00)	20 (83.33)	4 (16.67)	0 (0.00)
Austria	7 (1.86)	364 (96.81)	5 (1.33)	2 (0.53)	329 (87.50)	45 (11.97)	3 (0.80)	363 (96.54)	10 (2.66)
Belarus	147 (100)	0 (0)	0 (0)	0 (0.00)	143 (97.28)	4 (2.72)	0 (0.00)	136 (92.52)	11 (7.48)
Belgium	0 (0.00)	453 (98.48)	7 (1.52)	0 (0.00)	404 (87.83)	56 (12.17)	0 (0.00)	434 (94.35)	26 (5.65)
Bulgaria	99 (89.19)	12 (10.81)	0 (0.00)	1 (0.99)	107 (96.40)	3 (2.70)	1 (0.90)	106 (95.50)	4 (3.60)
Croatia	55 (61.11)	35 (38.89)	0 (0.00)	10 (11.11)	75 (83.33)	5 (5.56)	9 (10.00)	81 (90.00)	0 (0.00)
Cyprus	0 (0.00)	12 (92.31)	1 (7.69)	0 (0.00)	12 (92.31)	1 (7.69)	0 (0)	13 (100)	0 (0)
Czech Republic	236 (72.61)	84 (25.85)	5 (1.54)	3 (0.92)	293 (90.16)	29 (8.92)	3 (0.92)	314 (96.62)	8 (2.46)
Denmark	0 (0.00)	197 (98.99)	2 (1.01)	0 (0.00)	164 (82.41)	35 (17.59)	0 (0.00)	183 (91.96)	16 (8.04)
France	0 (0.00)	2647 (97.50)	68 (2.50)	0 (0.00)	2376 (87.51)	339 (12.49)	0 (0.00)	2563 (94.40)	152 (5.60)
Georgia	72 (100)	0 (0)	0 (0)	2 (2.78)	70 (97.22)	0 (0.00)	1 (1.39)	71 (98.61)	0 (0.00)
Germany	1982 (76.67)	574 (22.21)	29 (1.12)	26 (1.01)	2364 (91.45)	195 (7.54)	26 (1.01)	2498 (96.63)	61 (2.36)
Greece	0 (0)	101 (100)	0 (0)	0 (0.00)	86 (85.15)	15 (14.85)	0 (0.00)	96 (95.05)	5 (4.95)
Hungary	8 (3.20)	240 (96.00)	2 (0.80)	0 (0.00)	236 (94.40)	14 (5.60)	250 (100)	0 (0)	0 (0)
Ireland	0 (0.00)	486 (97.01)	15 (2.99)	0 (0.00)	461 (92.02)	40 (7.98)	0 (0.00)	489 (97.60)	12 (2.40)
Israel	11 (5.64)	169 (86.67)	15 (7.69)	8 (4.10)	166 (85.13)	21 (10.77)	7 (3.59)	184 (94.36)	4 (2.05)
Italy	6 (0.26)	2268 (98.70)	24 (1.04)	6 (0.26)	2093 (91.08)	199 (8.66)	5 (0.22)	2175 (94.65)	118 (5.13)
Latvia	15 (46.88)	17 (53.12)	0 (0.00)	6 (18.75)	19 (59.37)	7 (21.88)	6 (18.75)	25 (78.12)	1 (3.13)
Lithuania	0 (0)	14 (100)	0 (0)	0 (0.00)	12 (85.71)	2 (14.29)	0 (0)	14 (100)	0 (0)
Luxembourg	0 (0)	15 (100)	0 (0)	0 (0.00)	13 (86.67)	2 (13.33)	0 (0)	15 (100)	0 (0)
Moldova	44 (100)	0 (0)	0 (0)	44 (100)	0 (0)	0 (0)	44 (100)	0 (0)	0 (0)
The Netherlands	10 (1.82)	522 (94.91)	18 (3.27)	5 (0.91)	487 (88.54)	58 (10.55)	6 (1.09)	532 (96.73)	12 (2.18)

[table 5.5 continued]

Country	Non-tuberculous mycobacteria (NTM) infection this year number (%)			*Stenotrophomonas maltophilia* infection this year number (%)			*Achromobacter species* infection this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
North Macedonia	1 (1.12)	88 (98.88)	0 (0.00)	0 (0.00)	85 (95.51)	4 (4.49)	0 (0)	89 (100)	0 (0)
Norway	13 (11.61)	93 (83.03)	6 (5.36)	0 (0.00)	91 (81.25)	21 (18.75)	0 (0.00)	110 (98.21)	2 (1.79)
Poland	246 (29.39)	590 (70.49)	1 (0.12)	9 (1.08)	789 (94.26)	39 (4.66)	8 (0.96)	814 (97.25)	15 (1.79)
Portugal	3 (1.60)	183 (97.34)	2 (1.06)	3 (1.60)	166 (88.29)	19 (10.11)	4 (2.13)	172 (91.49)	12 (6.38)
Romania	71 (33.02)	144 (66.98)	0 (0.00)	3 (1.40)	208 (96.74)	4 (1.86)	6 (2.79)	206 (95.81)	3 (1.40)
Russian Federation	334 (14.21)	2008 (85.41)	9 (0.38)	81 (3.45)	2129 (90.55)	141 (6.00)	91 (3.87)	2141 (91.07)	119 (5.06)
Serbia	2 (1.63)	120 (97.56)	1 (0.81)	2 (1.63)	103 (83.74)	18 (14.63)	3 (2.44)	112 (91.06)	8 (6.50)
Slovak Republic	1 (0.77)	128 (98.46)	1 (0.77)	1 (0.77)	127 (97.69)	2 (1.54)	1 (0.77)	129 (99.23)	0 (0.00)
Slovenia	0 (0.00)	51 (92.73)	4 (7.27)	0 (0.00)	49 (89.09)	6 (10.91)	0 (0.00)	53 (96.36)	2 (3.64)
Spain	200 (18.54)	858 (79.51)	21 (1.95)	8 (0.74)	981 (90.92)	90 (8.34)	13 (1.20)	1011 (93.70)	55 (5.10)
Sweden	8 (3.05)	245 (93.51)	9 (3.44)	0 (0.00)	235 (89.69)	27 (10.31)	0 (0.00)	257 (98.09)	5 (1.91)
Switzerland	7 (1.67)	406 (96.90)	6 (1.43)	5 (1.19)	377 (89.98)	37 (8.83)	6 (1.43)	403 (96.18)	10 (2.39)
Turkey	33 (1.89)	1702 (97.71)	7 (0.40)	15 (0.86)	1685 (96.73)	42 (2.41)	1 (0.06)	1701 (97.64)	40 (2.30)
Ukraine	115 (95.04)	5 (4.13)	1 (0.83)	2 (1.65)	105 (86.78)	14 (11.57)	2 (1.65)	119 (98.35)	0 (0.00)
United Kingdom	11 (0.26)	4061 (95.78)	168 (3.96)	5 (0.12)	3952 (93.21)	283 (6.67)	5 (0.12)	4158 (98.06)	77 (1.82)
Total				270 (1.16)	21090 (91.00)	1817 (7.84)	527 (2.27)	21862 (94.33)	788 (3.40)

Note: For non-tuberculous mycobacteria (NTM) the total percentage of missing information is higher than 10%, therefore the totals are excluded from the table.

Table 5.5 shows the frequency of three other infections, non-tuberculous mycobacteria (NTM), *Stenotrophomonas maltophilia* and *Achromobacter species* in children.

Table 5.6 Prevalence of non-tuberculous mycobacteria, Stenotrophomonas maltophilia and Achromobacter species infection in adults seen in 2019 who have never had a transplant, by country and overall.

Country	Non-tuberculous mycobacteria (NTM) infection this year number (%)			*Stenotrophomonas maltophilia* infection this year number (%)			*Achromobacter species* infection this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	0 (0)	9 (100)	0 (0)	0 (0)	9 (100)	0 (0)	0 (0)	9 (100)	0 (0)
Austria	34 (9.94)	279 (81.58)	29 (8.48)	1 (0.29)	273 (79.83)	68 (19.88)	3 (0.88)	312 (91.23)	27 (7.89)
Belarus	8 (100)	0 (0)	0 (0)	0 (0)	8 (100)	0 (0)	0 (0)	8 (100)	0 (0)
Belgium	1 (0.15)	631 (96.49)	22 (3.36)	1 (0.15)	575 (87.92)	78 (11.93)	1 (0.15)	581 (88.84)	72 (11.01)
Bulgaria	68 (90.67)	6 (8.00)	1 (1.33)	0 (0.00)	73 (97.33)	2 (2.67)	0 (0.00)	74 (98.67)	1 (1.33)
Croatia	4 (9.76)	35 (85.36)	2 (4.88)	0 (0.00)	38 (92.68)	3 (7.32)	0 (0)	41 (100)	0 (0)
Cyprus	2 (14.29)	12 (85.71)	0 (0.00)	2 (14.29)	12 (85.71)	0 (0.00)	2 (14.29)	11 (78.57)	1 (7.14)
Czech Republic	47 (17.54)	204 (76.12)	17 (6.34)	9 (3.36)	245 (91.42)	14 (5.22)	10 (3.73)	245 (91.42)	13 (4.85)
Denmark	2 (0.74)	250 (92.59)	18 (6.67)	2 (0.74)	200 (74.07)	68 (25.19)	2 (0.74)	226 (83.70)	42 (15.56)
France	0 (0.00)	2923 (94.23)	179 (5.77)	0 (0.00)	2730 (88.01)	372 (11.99)	0 (0.00)	2790 (89.94)	312 (10.06)
Germany	1729 (51.77)	1468 (43.95)	143 (4.28)	92 (2.75)	2869 (85.90)	379 (11.35)	89 (2.66)	3031 (90.75)	220 (6.59)
Greece	0 (0.00)	284 (95.30)	14 (4.70)	16 (5.37)	249 (83.56)	33 (11.07)	14 (4.70)	253 (84.90)	31 (10.40)
Hungary	5 (2.56)	179 (91.80)	11 (5.64)	5 (2.56)	177 (90.77)	13 (6.67)	195 (100)	0 (0)	0 (0)
Ireland	2 (0.31)	616 (96.71)	19 (2.98)	2 (0.31)	574 (90.11)	61 (9.58)	2 (0.31)	611 (95.92)	24 (3.77)
Israel	16 (5.03)	269 (84.59)	33 (10.38)	12 (3.77)	283 (89.00)	23 (7.23)	15 (4.72)	289 (90.88)	14 (4.40)
Italy	17 (0.57)	2927 (98.32)	33 (1.11)	17 (0.57)	2760 (92.71)	200 (6.72)	16 (0.54)	2693 (90.46)	268 (9.00)
Latvia	4 (30.77)	9 (69.23)	0 (0.00)	0 (0.00)	10 (76.92)	3 (23.08)	0 (0.00)	11 (84.62)	2 (15.38)
Lithuania	0 (0)	13 (100)	0 (0)	0 (0.00)	10 (76.92)	3 (23.08)	0 (0)	13 (100)	0 (0)
Luxembourg	2 (9.52)	18 (85.72)	1 (4.76)	2 (9.52)	19 (90.48)	0 (0.00)	1 (4.76)	20 (95.24)	0 (0.00)
Moldova	9 (100)	0 (0)	0 (0)	9 (100)	0 (0)	0 (0)	9 (100)	0 (0)	0 (0)
The Netherlands	101 (11.97)	711 (84.24)	32 (3.79)	51 (6.04)	684 (81.05)	109 (12.91)	55 (6.52)	742 (87.91)	47 (5.57)

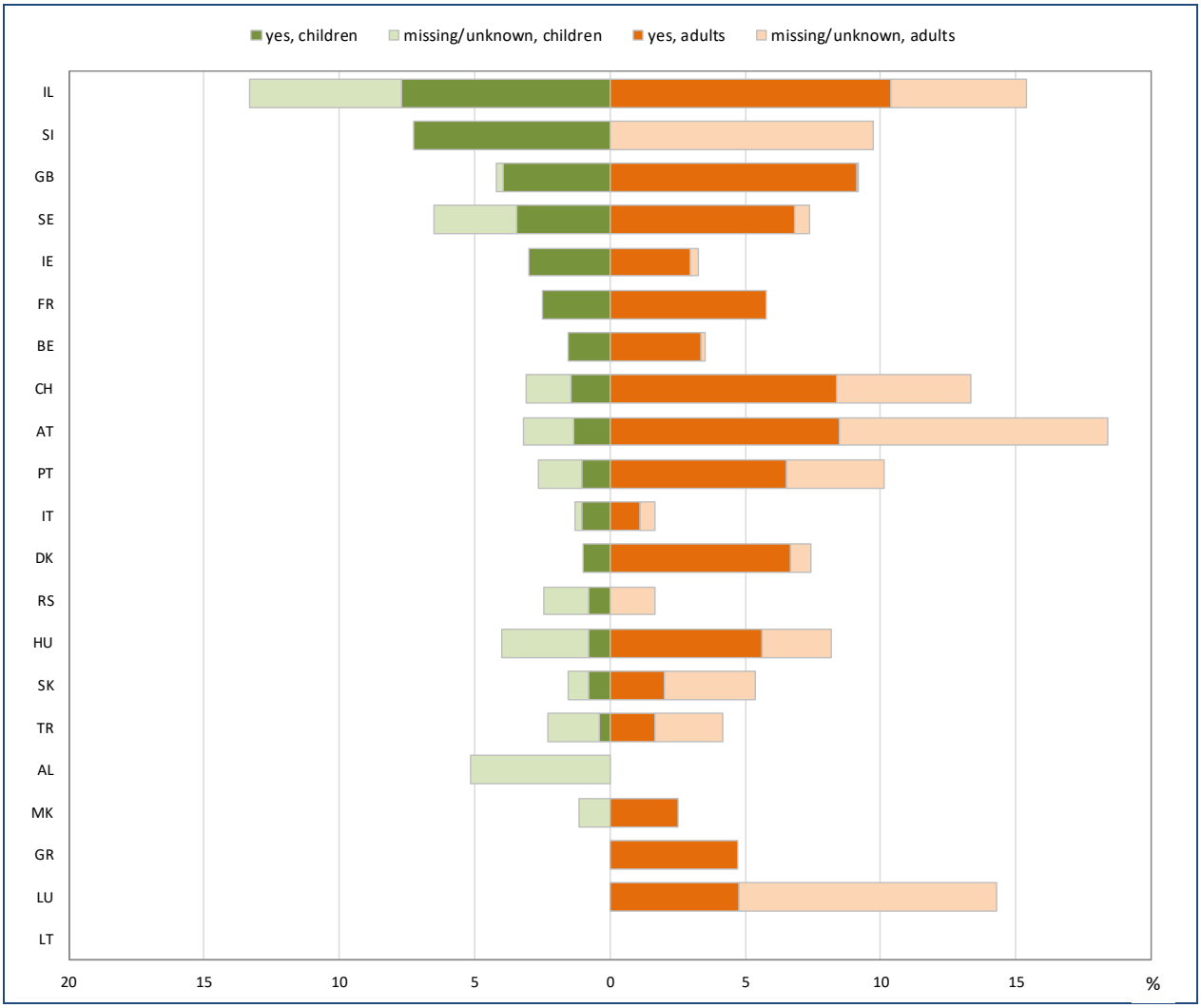
[table 5.6 continued]

Country	Non-tuberculous mycobacteria (NTM) infection this year number (%)			*Stenotrophomonas maltophilia* infection this year number (%)			*Achromobacter species* infection this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
North Macedonia	0 (0.00)	39 (97.50)	1 (2.50)	0 (0)	40 (100)	0 (0)	0 (0)	40 (100)	0 (0)
Norway	15 (9.09)	141 (85.46)	9 (5.45)	5 (3.03)	131 (79.39)	29 (17.58)	5 (3.03)	151 (91.52)	9 (5.45)
Poland	27 (8.01)	307 (91.10)	3 (0.89)	7 (2.08)	307 (91.10)	23 (6.82)	8 (2.37)	298 (88.43)	31 (9.20)
Portugal	5 (3.62)	124 (89.86)	9 (6.52)	5 (3.62)	120 (86.96)	13 (9.42)	5 (3.62)	121 (87.68)	12 (8.70)
Romania	1 (9.09)	10 (90.91)	0 (0.00)	0 (0)	11 (100)	0 (0)	0 (0)	11 (100)	0 (0)
Russian Federation	38 (4.97)	708 (92.55)	19 (2.48)	21 (2.75)	711 (92.94)	33 (4.31)	15 (1.96)	684 (89.41)	66 (8.63)
Serbia	1 (1.67)	59 (98.33)	0 (0.00)	1 (1.67)	56 (93.33)	3 (5.00)	2 (3.33)	53 (88.34)	5 (8.33)
Slovak Republic	5 (3.36)	141 (94.63)	3 (2.01)	6 (4.03)	130 (87.25)	13 (8.72)	7 (4.70)	132 (88.59)	10 (6.71)
Slovenia	4 (9.76)	37 (90.24)	0 (0.00)	6 (14.63)	29 (70.74)	6 (14.63)	3 (7.32)	37 (90.24)	1 (2.44)
Spain	22 (2.13)	933 (90.41)	77 (7.46)	13 (1.26)	928 (89.92)	91 (8.82)	23 (2.23)	905 (87.69)	104 (10.08)
Sweden	2 (0.57)	326 (92.61)	24 (6.82)	0 (0.00)	306 (86.93)	46 (13.07)	0 (0.00)	339 (96.31)	13 (3.69)
Switzerland	24 (4.92)	423 (86.68)	41 (8.40)	9 (1.84)	408 (83.61)	71 (14.55)	11 (2.25)	445 (91.19)	32 (6.56)
Turkey	6 (2.50)	230 (95.83)	4 (1.67)	4 (1.67)	221 (92.08)	15 (6.25)	1 (0.42)	222 (92.50)	17 (7.08)
Ukraine	26 (86.67)	3 (10.00)	1 (3.33)	6 (20.00)	23 (76.67)	1 (3.33)	1 (3.33)	29 (96.67)	0 (0.00)
United Kingdom	3 (0.05)	4958 (90.83)	498 (9.12)	3 (0.05)	4960 (90.86)	496 (9.09)	3 (0.05)	5162 (94.56)	294 (5.39)
Total	2230 (9.80)	19283 (84.74)	1243 (5.46)	308 (1.35)	20179 (88.68)	2269 (9.97)	499 (2.19)	20589 (90.48)	1668 (7.33)

Note: Georgia has 0% coverage for adults and is excluded from the table.
Note: Armenia has <5 patients aged 18 years or more at 31/12/2019 and is not shown in this table, but is considered in the total.

Table 5.6 shows the frequency of three other infections, non-tuberculous mycobacteria (NTM), *Stenotrophomonas maltophilia* and *Achromobacter species* in adults.

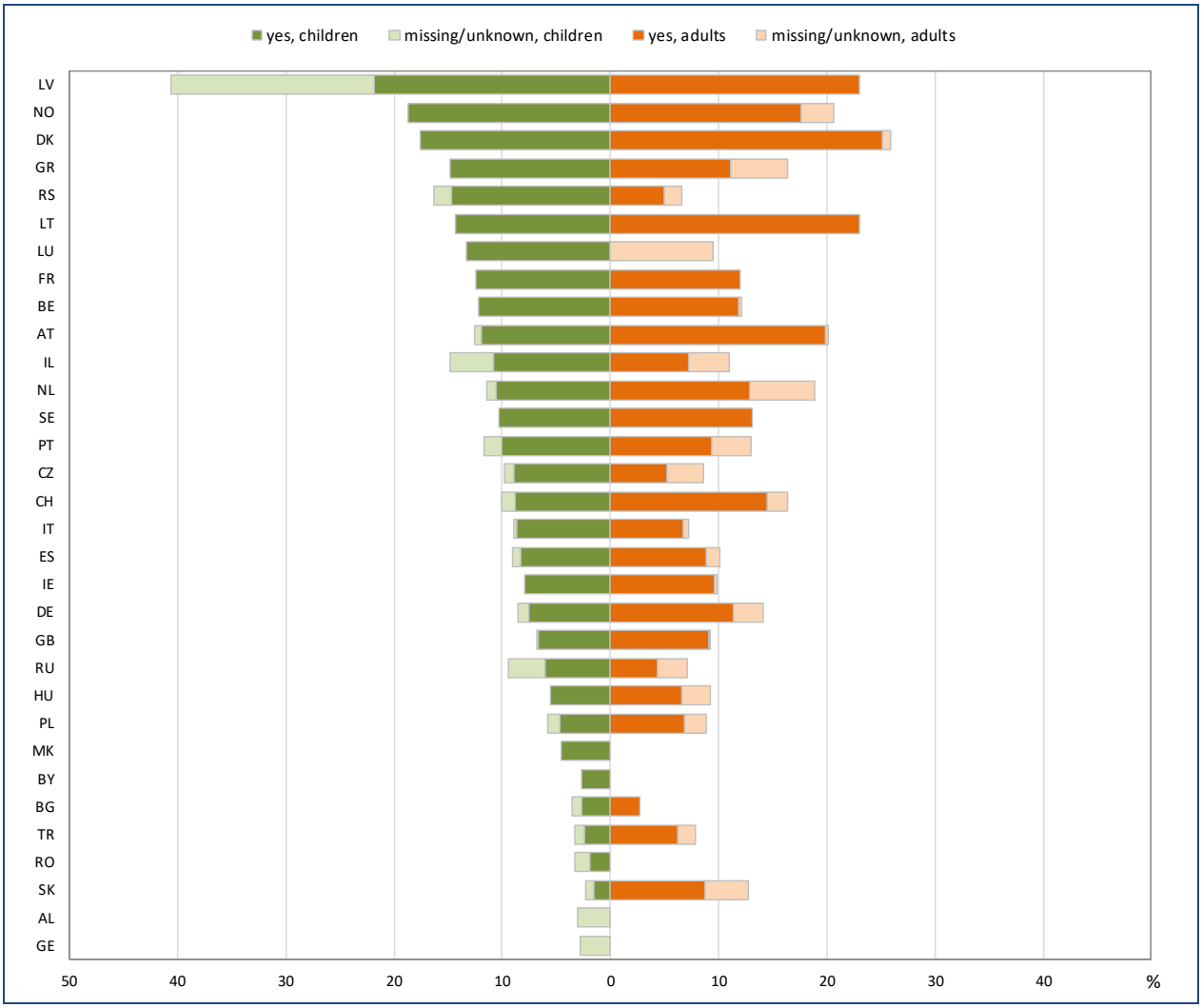
Figure 5.6 Prevalence of non-tuberculous mycobacteria in children and adults seen in 2019 who have never had a transplant, by country.



Note: We excluded from the graph the countries for which the information on non-tuberculous mycobacteria was missing for more than 10% of the children and/or adults.
Georgia has 0% coverage for adults and the adults bar is excluded from the graph.
Armenia has <5 patients aged 18 years or more at 31/12/2019 and the adults bar is excluded from the graph.

This graph represents the percentage of people with non-tuberculous mycobacteria infection (in dark colours) and the percentage of people where information on non-tuberculous mycobacteria infection was 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. Detection of non-tuberculous mycobacteria infection depends on sputum production, which is not always possible for all patients, especially younger patients. Generally, infections from these bacteria are not very frequent in any country.

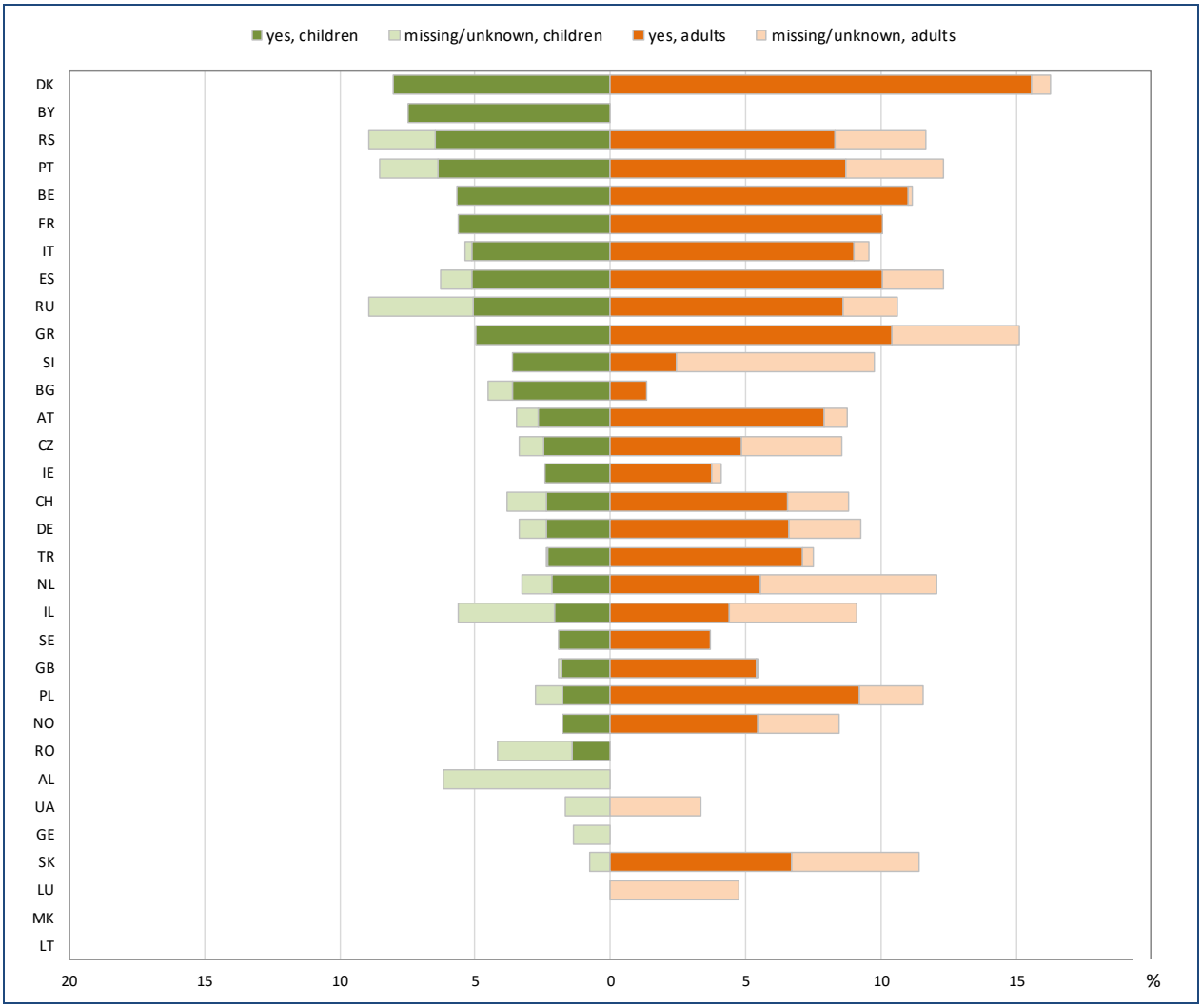
Figure 5.7 Prevalence of Stenotrophomonas maltophilia infection in children and adults seen in 2019 who have never had a transplant, by country.



Note: We excluded from the graph the countries for which the information on *Stenotrophomonas maltophilia* was missing for more than 10% of the children and/or adults.
Georgia has 0% coverage for adults and the adults bar is excluded from the graph.
Armenia has <5 patients aged 18 years or more at 31/12/2019 and the adults bar is excluded from the graph.

This graph represents the percentage of people with *Stenotrophomonas maltophilia* infection (in dark colours) and the percentage of people where information on *Stenotrophomonas maltophilia* infection was 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. The frequency varies considerably between countries.

Figure 5.8 Prevalence of Achromobacter species in children and adults seen in 2019 who have never had a transplant, by country.



Note: We excluded from the graph the countries for which the information on *Achromobacter species* was missing for more than 10% of the children and/or adults.
Georgia has 0% coverage for adults and the adults bar is excluded from the graph.
Armenia has <5 patients aged 18 years or more at 31/12/2019 and the adults bar is excluded from the graph.

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 was 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.

6. Nutrition

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

We collected weight and height measured on the date the best FEV1 value (of the highest FEV1% predicted of the year) was recorded. For patients that did not perform spirometry, the last weight and height measurements of the year were considered. From these raw values we calculated body mass index (BMI). A patient with a low weight is not necessarily underweight if the height is also low, therefore BMI may better illustrate the nutritional status because it describes the weight/height relationship. The ECFS Standards of Care guidelines recommend: for adults, a BMI of above 20 kg/m^2; for older children and adolescents, 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 by 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 3, page 156, 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 of the reference population. A z-score of -2 means that the height/weight/BMI value is 2 standard deviations below the mean height/weight/BMI of people of the same age and sex of 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 of 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.

Figure 6.1 Use of pancreatic enzymes in 2019 for all patients who have never had a transplant, by country and overall.

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

Table 6.1 Z-scores for height: descriptive statistics by country. Patients aged 17 years or younger who have never had a transplant.

Country	N	N miss	Mean	Min	25th pctl (25% of the patients are below this z-score for height)	Median (50% of the patients are below this z-score for height)	75th pctl (75% of the patients are below this z-score for height)	Max
Albania	83	15	-0.6	-3.8	-1.3	-0.6	0.0	1.9
Armenia	25	0	-1.0	-3.3	-1.9	-1.0	-0.2	1.5
Austria	380	1	0.1	-2.6	-0.6	0.0	0.7	4.0
Belarus	94	3	-0.5	-4.0	-1.7	-0.4	0.5	5.0
Belgium	475	0	-0.4	-4.8	-1.1	-0.4	0.3	2.5
Bulgaria	108	0	-0.7	-5.7	-1.6	-0.8	0.3	2.2
Croatia	87	5	0.0	-2.3	-0.7	0.1	0.8	2.6
Cyprus	14	0	-0.1	-1.7	-0.8	-0.1	0.5	1.2
Czech Republic	331	4	-0.1	-8.1	-0.8	-0.0	0.6	3.5
Denmark	207	0	0.1	-2.2	-0.5	0.2	0.7	2.5
France	2760	18	-0.4	-8.9	-1.1	-0.4	0.2	4.2
Georgia	59	0	-1.4	-5.5	-2.2	-1.4	-0.6	2.1
Germany	2654	4	-0.2	-4.5	-0.8	-0.2	0.5	5.5
Greece	99	4	-0.1	-2.5	-0.8	-0.1	0.6	2.8
Hungary	249	1	-0.5	-5.6	-1.3	-0.3	0.3	4.7
Ireland	486	33	-0.2	-3.4	-0.8	-0.2	0.4	3.2
Israel	198	0	-0.5	-4.7	-1.3	-0.5	0.3	2.1
Italy	2271	14	-0.1	-5.0	-0.8	-0.1	0.6	4.4
Latvia	31	0	-0.0	-2.6	-0.4	0.2	0.6	2.1
Lithuania	13	1	0.0	-1.3	-0.7	-0.4	0.5	2.7
Luxembourg	16	0	-0.1	-2.1	-0.4	-0.3	0.7	1.1
Rep of Moldova	45	0	-0.9	-3.8	-1.4	-0.9	-0.2	1.4
The Netherlands	567	5	0.2	-3.9	-0.4	0.2	0.9	3.5
North Macedonia	91	0	-0.2	-4.7	-1.0	-0.3	0.7	3.0
Norway[1]	115	0	0.1	-1.9	-0.5	0.1	0.8	2.6
Poland	817	8	-0.0	-3.9	-0.7	-0.0	0.7	3.3
Portugal	186	1	-0.5	-4.3	-1.2	-0.5	0.2	2.1
Romania	208	8	-0.4	-4.5	-1.2	-0.4	0.5	3.5
Russian Federation	2309	17	-0.5	-9.9	-1.3	-0.4	0.4	6.6
Serbia	127	1	-0.1	-3.1	-0.8	-0.1	0.5	2.5
Slovak Republic	126	5	0.1	-3.9	-0.7	0.1	1.0	2.8
Slovenia	57	0	0.2	-2.1	-0.5	0.2	0.7	3.4
Spain	1084	4	-0.2	-4.5	-0.9	-0.2	0.5	5.6
Sweden[2]	265	0	0.0	-3.0	-0.6	0.0	0.7	3.6
Switzerland	428	4	-0.2	-2.9	-0.8	-0.1	0.4	3.3
Turkey	1761	0	-0.5	-7.6	-1.3	-0.5	0.3	4.0
Ukraine	121	1	-0.3	-3.0	-0.9	-0.3	0.3	2.7
United Kingdom[3]	4395	26	-0.2	-4.9	-0.9	-0.2	0.4	4.7
Total	23342	183	-0.3	-9.9	-1.0	-0.2	0.5	6.6

[1] Norway: sometimes any value (instead of last of the year) for height is used when no lung function test was available.
[2] Sweden: if there is no lung function measurement, the height and weight reported are those measured at annual review.
[3] United Kingdom: height and weight at date of annual data is used instead of the date of best FEV1. If no lung function measurement is reported, the date of the last visit is used.

This table reports the median z-score for height (the value that separates the highest and lowest half of the patients), the mean z-score for height (the average) and other descriptive statistics for children (17 years or younger).

Table 6.2 Z-scores for height: descriptive statistics by country. Patients aged 18 years or older who have never had a transplant.

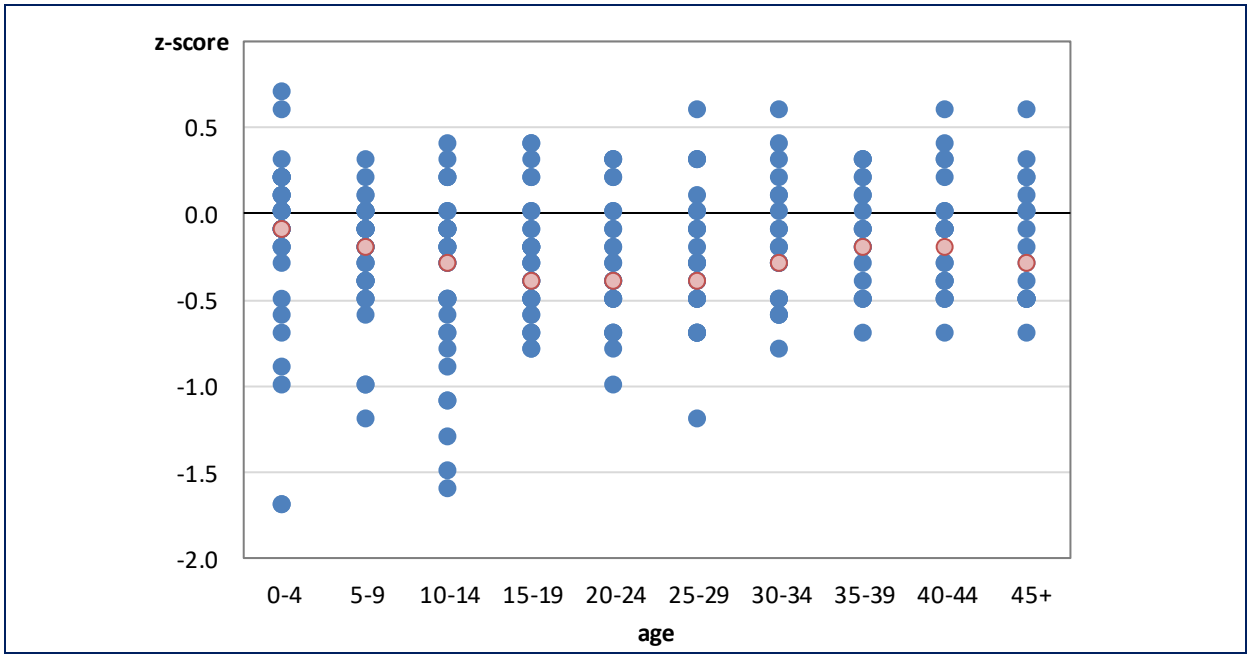
Country	N	N miss	Mean	Min	25th pctl (25% of the patients are below this z-score for height)	Median (50% of the patients are below this z-score for height)	75th pctl (75% of the patients are below this z-score for height)	Max
Austria	335	0	-0.2	-3.4	-0.8	-0.3	0.3	2.7
Belgium	637	2	-0.3	-3.9	-1.1	-0.3	0.4	3.2
Bulgaria	67	0	-0.0	-2.3	-0.8	-0.4	0.7	2.1
Croatia	39	0	0.1	-2.6	-0.4	0.1	0.7	1.9
Cyprus	13	0	-0.8	-2.4	-0.8	-0.7	-0.4	-0.2
Czech Republic	253	0	-0.1	-3.4	-0.7	-0.1	0.6	2.7
Denmark	259	0	0.2	-2.3	-0.5	0.1	0.9	3.3
France	2964	7	-0.5	-5.7	-1.1	-0.5	0.1	3.9
Germany	3246	2	-0.0	-4.5	-0.8	-0.1	0.7	3.6
Greece	265	4	-0.5	-2.8	-1.2	-0.5	0.2	2.0
Hungary	189	6	-0.2	-3.2	-1.0	-0.3	0.6	3.6
Ireland	617	0	-0.3	-5.1	-1.0	-0.3	0.3	2.1
Israel	303	0	-0.6	-3.6	-1.3	-0.5	0.2	2.4
Italy	2818	26	-0.5	-3.7	-1.2	-0.5	0.2	4.0
Latvia	13	0	0.5	-1.1	-0.1	0.7	1.0	1.7
Lithuania	11	0	1.1	-0.2	0.4	1.0	2.0	2.3
Luxembourg	20	0	-0.2	-2.6	-1.0	-0.1	0.2	2.4
Rep of Moldova	8	0	0.0	-1.6	-1.0	0.1	1.1	1.5
The Netherlands	811	0	0.3	-2.8	-0.4	0.3	1.0	4.0
North Macedonia	36	0	-0.7	-2.6	-1.2	-0.7	-0.2	2.3
Norway[1]	160	0	0.3	-2.5	-0.3	0.4	1.0	2.8
Poland	294	1	-0.2	-4.0	-1.0	-0.2	0.4	3.0
Portugal	119	5	-0.9	-2.7	-1.6	-0.8	-0.4	1.2
Romania	8	0	-0.5	-1.0	-0.9	-0.5	-0.4	0.6
Russian Federation	712	0	-0.3	-6.9	-0.9	-0.3	0.3	3.4
Serbia	54	0	0.1	-1.5	-0.5	0.1	0.7	2.3
Slovak Republic	140	0	0.1	-3.7	-0.5	0.1	0.8	2.4
Slovenia	38	0	0.2	-1.5	-0.5	0.2	0.8	2.9
Spain	976	5	-0.6	-4.8	-1.3	-0.5	0.1	2.3
Sweden[2]	348	0	0.1	-3.1	-0.5	0.2	0.7	3.3
Switzerland	468	0	-0.2	-3.7	-0.8	-0.2	0.4	2.7
Turkey	220	0	-0.7	-3.6	-1.3	-0.7	-0.1	2.3
Ukraine	27	0	-0.6	-3.0	-1.2	-0.5	0.0	0.9
United Kingdom[3]	5260	2	-0.3	-6.0	-1.0	-0.4	0.3	4.8
Total	21735	60	-0.3	-6.9	-1.0	-0.4	0.4	4.8

[1] Norway: sometimes any value (instead of last of the year) for height is used when no lung function test was available.
[2] Sweden: if there is no lung function measurement, the height and weight reported are those measured at annual review.
[3] United Kingdom: height and weight at date of annual data is used instead of the date of best FEV1. If no lung function measurement is reported, the date of the last visit is used.

Note: Georgia has 0% coverage for adults and is excluded from the table.
Note: Albania, Armenia, Belarus have <5 patients aged 18 years or more at height measurement and are excluded from the table.

This table reports the median z-score for height (the value that separates the highest and lowest half of the patients), the mean z-score for height (the average) and other descriptive statistics for adults (18 years or older).

Figure 6.2 Median z-scores for height by age group and by country. All patients seen in 2019 who never had a transplant.



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

This graph shows the median z-scores for height by age group. Each country is represented by a dot (in blue) and the overall median estimate is in red. The overall median z-scores for height tend to slowly decrease up to the teenage years and then rise again. The graph also shows that there is large variability between countries.

Table 6.3 Z-scores for height: descriptive statistics by age group. All patients seen in 2019 who never had a transplant.

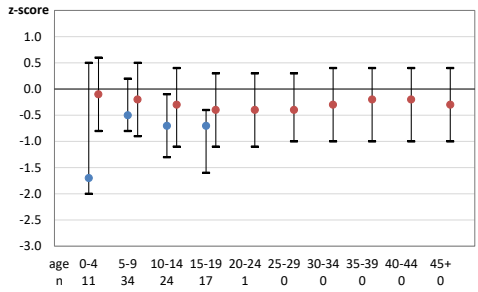
Age at height measurement	N	Mean	Min	25th pctl	Median	75th pctl	Max
0-4	6131	-0.2	-9.9	-0.9	-0.1	0.6	6.6
5-9	7069	-0.2	-5.8	-0.9	-0.2	0.5	5.9
10-14	6586	-0.3	-7.1	-1.1	-0.3	0.4	4.0
15-19	5740	-0.4	-7.6	-1.1	-0.4	0.3	4.2
20-24	4957	-0.4	-6.0	-1.1	-0.4	0.3	3.3
25-29	4260	-0.3	-4.8	-1.0	-0.4	0.3	4.8
30-34	3385	-0.3	-4.0	-1.0	-0.3	0.4	3.6
35-39	2430	-0.2	-5.7	-1.0	-0.2	0.4	3.6
40-44	1653	-0.2	-3.7	-1.0	-0.2	0.4	3.3
45+	2866	-0.3	-5.1	-1.0	-0.3	0.4	4.0

This table reports the median z-score for height and other descriptive statistics by age group for all the patients seen in 2019 who never had a transplant. The median values reported in this table are shown as red dots in fig 6.2.

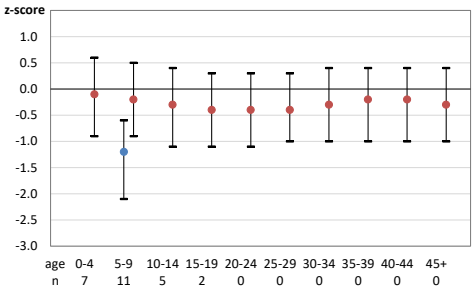
Figure 6.3 Quartiles of z-scores for height by age group and by country. All patients seen in 2019 who never had a transplant.

The figures below show the z-scores for height by country. The dot is the median and the whiskers show the 25th and 75th percentiles. In blue are the quartiles for the country, in red are the pooled quartiles computed on all other countries (i.e. excluding that country). We did not compute quartiles where the number of patients in the age group is <10, therefore there are no blue dots for those age groups (the number of patients in each age group is shown underneath the horizontal axis). We therefore excluded Cyprus, Lithuania and Luxembourg from the graphs because none of the age groups in these countries had more than 10 patients.

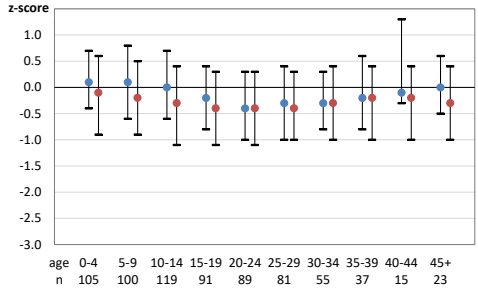
Quartiles of z-scores for height: Albania



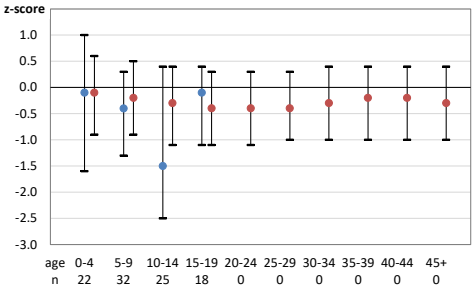
Quartiles of z-scores for height: Armenia



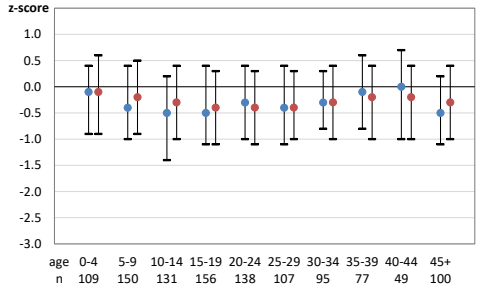
Quartiles of z-scores for height: Austria



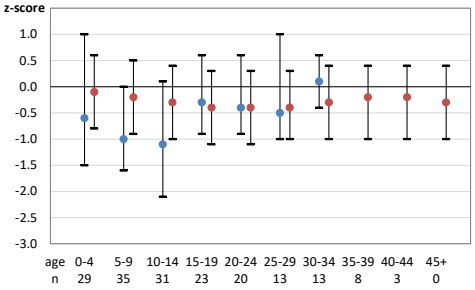
Quartiles of z-scores for height: Belarus



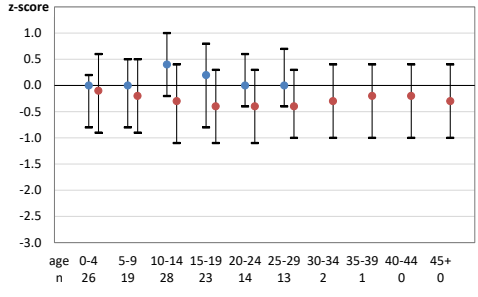
Quartiles of z-scores for height: Belgium



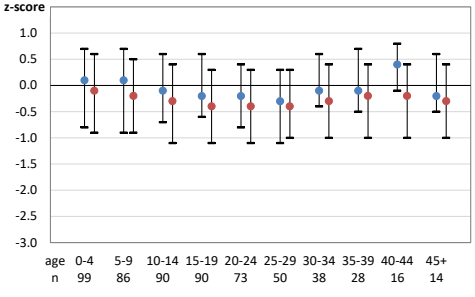
Quartiles of z-scores for height: Bulgaria



Quartiles of z-scores for height: Croatia

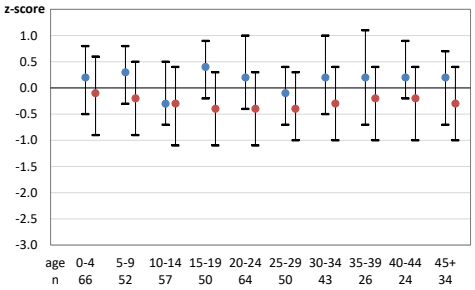


Quartiles of z-scores for height: Czech Republic

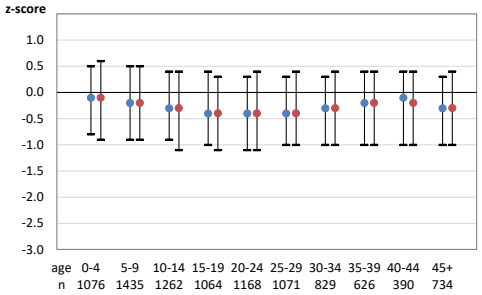


[figure 6.3 continued]

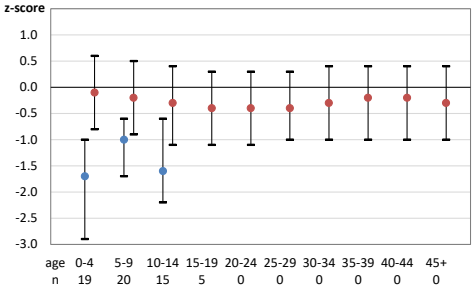
Quartiles of z-scores for height: Denmark



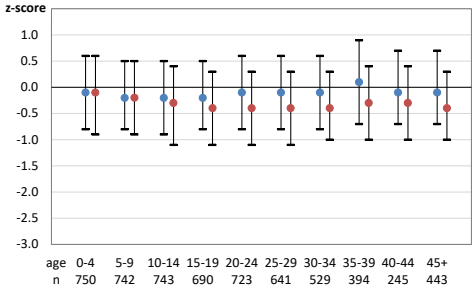
Quartiles of z-scores for height: France



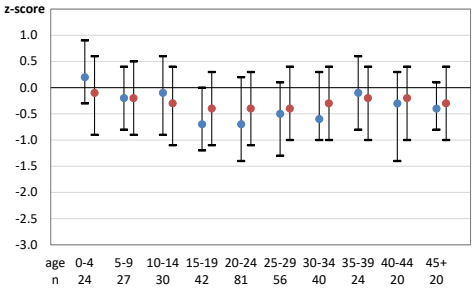
Quartiles of z-scores for height: Georgia



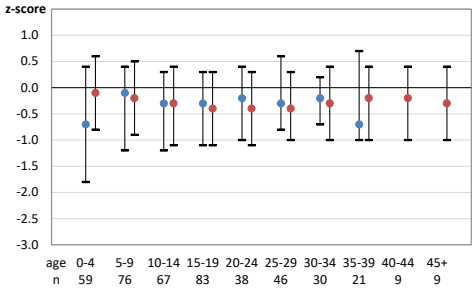
Quartiles of z-scores for height: Germany



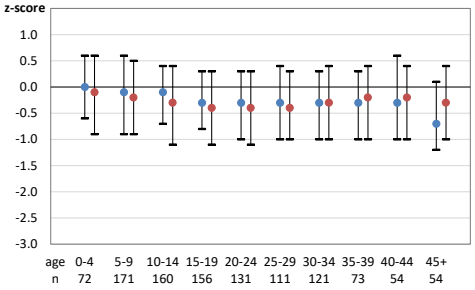
Quartiles of z-scores for height: Greece



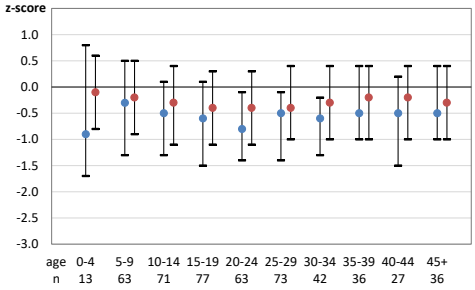
Quartiles of z-scores for height: Hungary



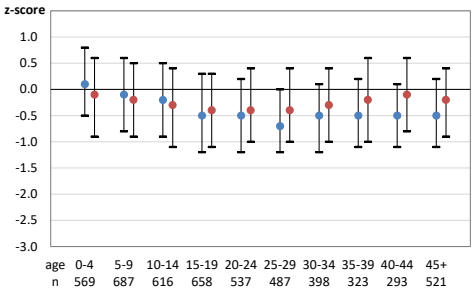
Quartiles of z-scores for height: Ireland



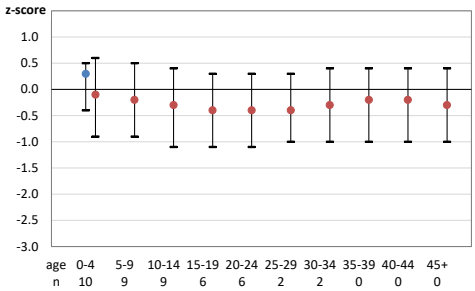
Quartiles of z-scores for height: Israel



Quartiles of z-scores for height: Italy

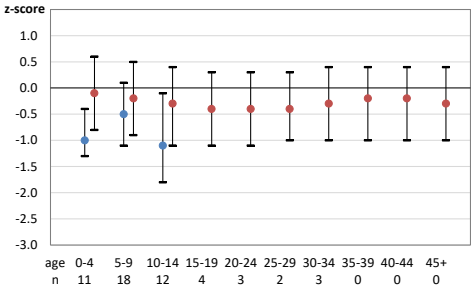


Quartiles of z-scores for height: Latvia

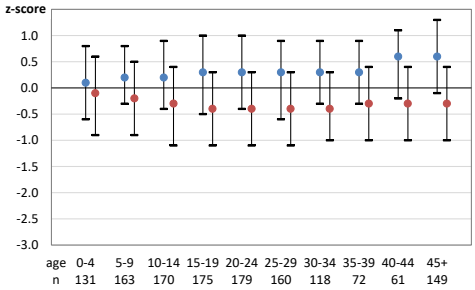


[figure 6.3 continued]

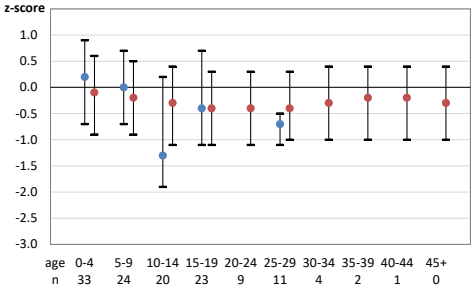
Quartiles of z-scores for height: Rep of Moldova



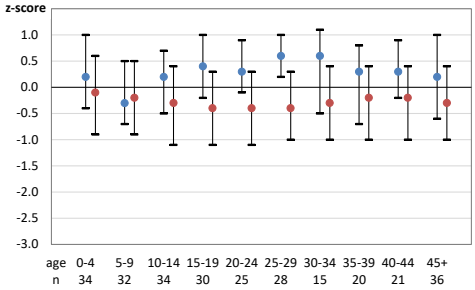
Quartiles of z-scores for height: The Netherlands



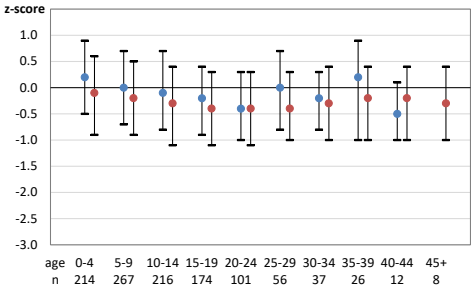
Quartiles of z-scores for height: North Macedonia



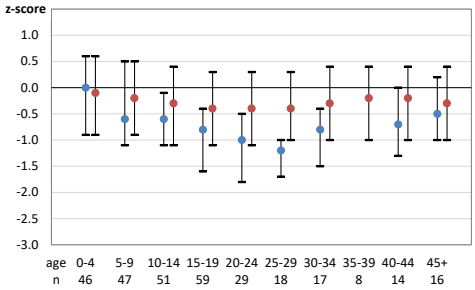
Quartiles of z-scores for height: Norway[1]



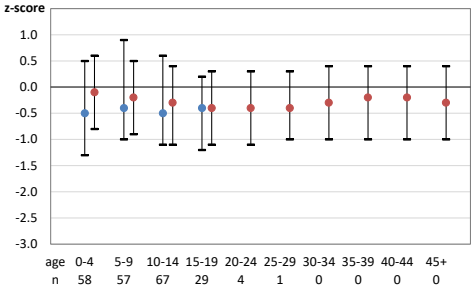
Quartiles of z-scores for height: Poland



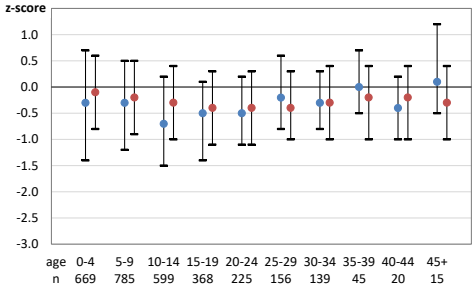
Quartiles of z-scores for height: Portugal



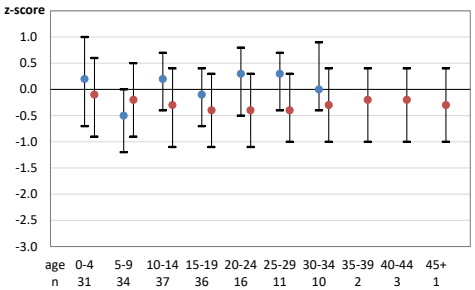
Quartiles of z-scores for height: Romania



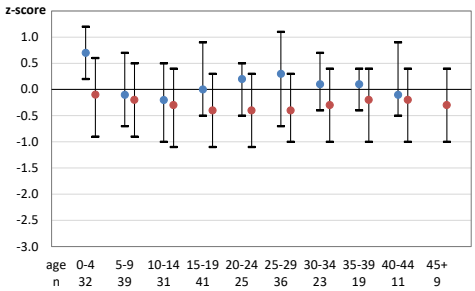
Quartiles of z-scores for height: Russian Federation



Quartiles of z-scores for height: Serbia

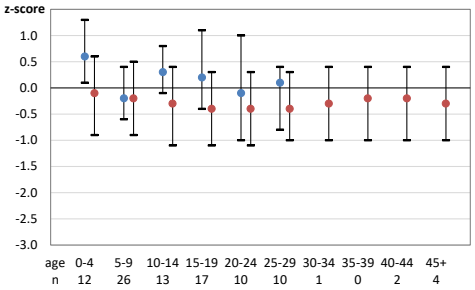


Quartiles of z-scores for height: Slovak Republic

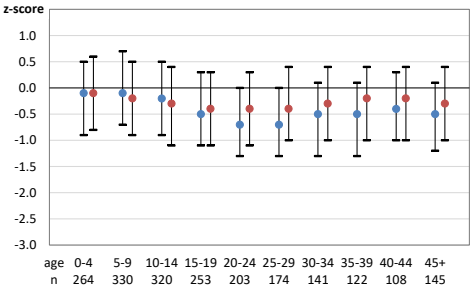


[figure 6.3 continued]

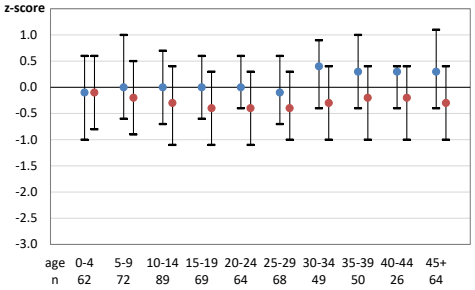
Quartiles of z-scores for height: Slovenia



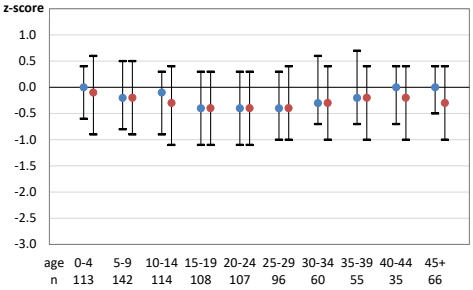
Quartiles of z-scores for height: Spain



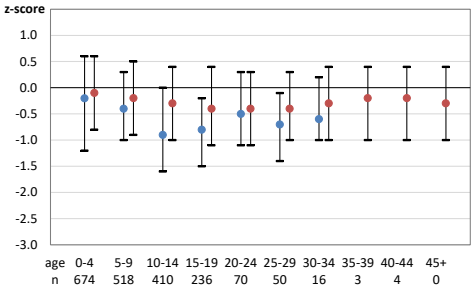
Quartiles of z-scores for height: Sweden[2]



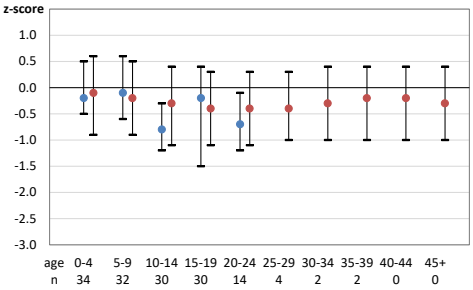
Quartiles of z-scores for height: Switzerland



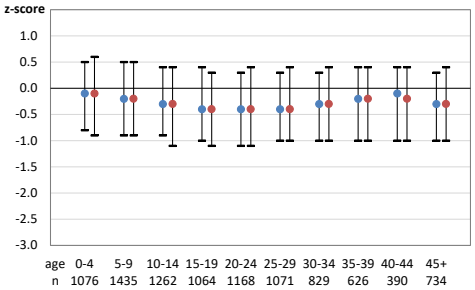
Quartiles of z-scores for height: Turkey



Quartiles of z-scores for height: Ukraine



Quartiles of z-scores for height: United Kingdom[3]



[1] Norway: sometimes any value (instead of last of the year) for height is used when no lung function test was available.
[2] Sweden: if there is no lung function measurement, the height and weight reported are those measured at annual review.
[3] United Kingdom: height and weight at date of annual data is used instead of the date of best FEV1. If no lung function measurement is reported, the date of the last visit is used.

Table 6.4 Z-scores for weight: descriptive statistics by country. Patients aged 17 years or younger who never had a transplant.

Country	N	N miss	Mean	Min	25th pctl (25% of the patients are below this z-score for weight)	Median (50% of the patients are below this z-score for weight)	75th pctl (75% of the patients are below this z-score for weight)	Max
Albania	98	0	-0.8	-5.9	-1.4	-0.5	0.0	2.8
Armenia	25	0	-1.0	-4.7	-1.9	-0.6	-0.2	1.5
Austria	381	0	-0.2	-3.4	-0.8	-0.1	0.5	2.8
Belarus	96	1	-0.9	-4.7	-1.5	-0.7	0.0	1.9
Belgium	475	0	-0.5	-3.9	-1.1	-0.5	0.2	2.7
Bulgaria	108	0	-1.1	-4.9	-1.9	-0.9	-0.1	2.4
Croatia	92	0	-0.3	-4.4	-1.0	-0.4	0.4	3.8
Cyprus	14	0	-0.4	-1.8	-1.1	-0.6	0.1	1.9
Czech Republic	335	0	-0.3	-7.1	-1.0	-0.2	0.4	2.4
Denmark	207	0	-0.1	-3.1	-0.9	-0.1	0.5	2.0
France	2761	17	-0.6	-7.9	-1.2	-0.6	0.1	3.6
Georgia	59	0	-0.7	-3.1	-1.7	-0.6	0.1	2.0
Germany	2657	1	-0.4	-6.7	-1.0	-0.3	0.3	7.4
Greece	103	0	0.1	-2.7	-0.7	0.2	0.9	2.4
Hungary	249	1	-0.9	-5.8	-1.6	-0.7	0.1	2.1
Ireland	496	23	0.0	-3.1	-0.5	0.0	0.6	2.7
Israel	198	0	-0.5	-6.8	-1.3	-0.4	0.3	2.2
Italy	2263	22	-0.2	-5.1	-0.9	-0.2	0.6	3.4
Latvia	31	0	-0.7	-2.9	-1.6	-0.6	0.2	1.3
Lithuania	14	0	-0.9	-5.5	-1.2	-1.0	-0.2	0.9
Luxembourg	16	0	-0.4	-2.2	-1.0	-0.4	-0.0	1.0
Rep of Moldova	45	0	-1.2	-5.4	-2.3	-1.0	-0.3	1.6
The Netherlands	572	0	-0.0	-3.9	-0.6	-0.0	0.6	2.1
North Macedonia	91	0	-0.3	-3.1	-1.2	-0.3	0.5	5.5
Norway[1]	115	0	-0.1	-2.2	-0.7	-0.1	0.6	1.9
Poland	824	1	-0.3	-4.7	-0.9	-0.2	0.4	2.8
Portugal	186	1	-0.6	-4.3	-1.3	-0.5	0.1	1.7
Romania	214	2	-1.0	-6.0	-1.6	-0.8	-0.1	2.0
Russian Federation	2319	7	-0.9	-9.1	-1.7	-0.7	0.2	8.7
Serbia	128	0	-0.5	-4.5	-1.2	-0.5	0.2	2.3
Slovak Republic	131	0	-0.3	-3.0	-1.1	-0.3	0.6	2.3
Slovenia	57	0	-0.2	-2.9	-0.7	-0.3	0.3	2.1
Spain	1086	2	-0.3	-5.7	-0.9	-0.2	0.4	3.1
Sweden[2]	265	0	-0.1	-2.9	-0.7	-0.1	0.5	2.5
Switzerland	432	0	-0.3	-4.1	-0.9	-0.2	0.3	4.1
Turkey	1761	0	-0.8	-9.2	-1.7	-0.7	0.1	3.8
Ukraine	121	1	-0.9	-7.2	-1.7	-0.9	-0.2	2.9
United Kingdom[3]	4404	17	-0.1	-5.1	-0.7	-0.1	0.6	4.6
Total	23429	96	-0.4	-9.2	-1.1	-0.3	0.4	8.7

[1] Norway: sometimes any value (instead of last of the year) for height is used when no lung function test was available.
[2] Sweden: if there is no lung function measurement, the height and weight reported are those measured at annual review.
[3] United Kingdom: height and weight at date of annual data is used instead of the date of best FEV1. If no lung function measurement is reported, the date of the last visit is used.

Table 6.4 reports the median z-score for weight (the value that separates the highest and lowest half of the patients), the mean z-score for weight (the average) and other descriptive statistics for children (17 years or younger).

Table 6.5 Z-scores for weight: descriptive statistics by country. Patients aged 18 years or older who never had a transplant.

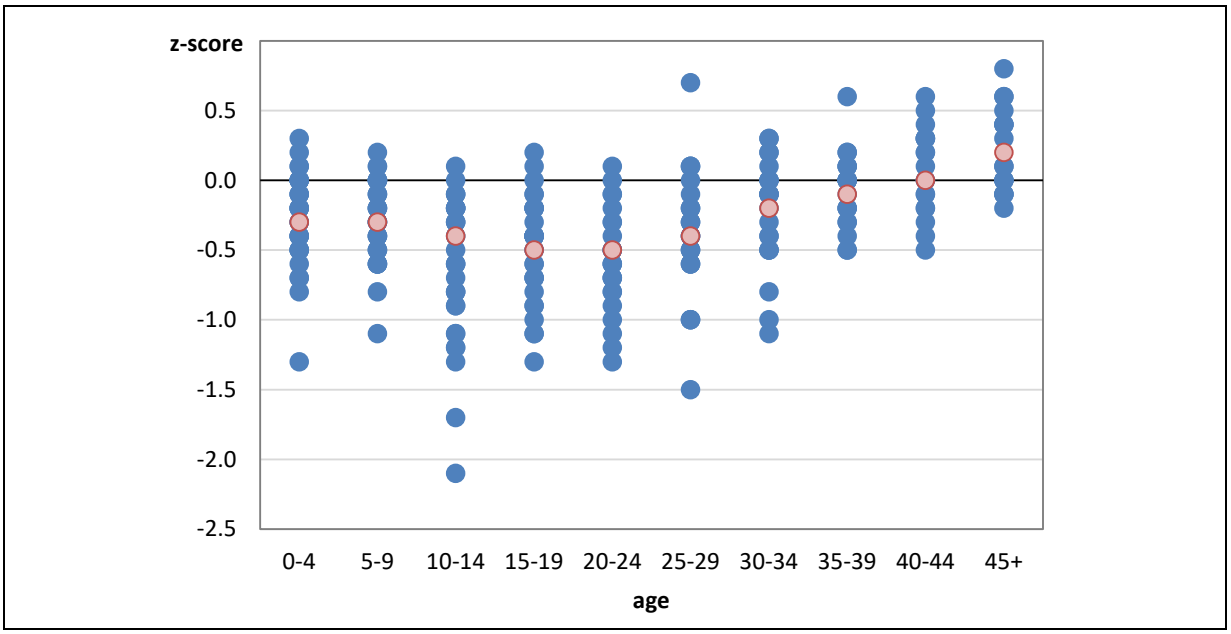
Country	N	N miss	Mean	Min	25th pctl (25% of the patients are below this z-score for weight)	Median (50% of the patients are below this z-score for weight)	75th pctl (75% of the patients are below this z-score for weight)	Max
Austria	335	0	-0.4	-3.4	-1.1	-0.3	0.3	2.5
Belgium	637	2	-0.3	-4.8	-0.9	-0.1	0.5	2.5
Bulgaria	67	0	-0.9	-4.9	-1.5	-0.9	-0.1	2.5
Croatia	39	0	-0.3	-2.9	-1.1	-0.0	0.4	1.3
Cyprus	13	0	-0.2	-1.3	-1.0	-0.5	0.5	2.0
Czech Republic	253	0	-0.3	-4.6	-1.0	-0.2	0.4	2.1
Denmark	259	0	-0.0	-5.2	-0.7	0.1	0.7	2.3
France	2925	46	-0.6	-6.8	-1.3	-0.6	0.1	3.0
Germany	3236	12	-0.3	-7.3	-1.0	-0.2	0.4	2.7
Greece	265	4	-0.3	-4.2	-1.0	-0.3	0.4	2.1
Hungary	191	4	-0.8	-5.5	-1.7	-0.6	0.1	2.0
Ireland	509	108	-0.2	-7.7	-0.7	-0.1	0.6	2.8
Israel	303	0	-0.3	-3.6	-1.1	-0.2	0.5	2.3
Italy	2802	42	-0.4	-6.0	-1.0	-0.4	0.3	3.5
Latvia	13	0	-0.8	-2.5	-1.3	-0.9	-0.4	0.9
Lithuania	11	0	0.3	-0.7	-0.2	0.1	0.7	1.8
Luxembourg	20	0	-0.2	-3.2	-0.7	0.0	0.4	2.4
Rep of Moldova	8	0	-1.3	-3.9	-2.2	-1.1	-0.3	0.3
The Netherlands	811	0	0.1	-3.4	-0.5	0.1	0.7	2.6
North Macedonia	36	0	-0.6	-3.1	-1.1	-0.4	0.2	1.4
Norway[1]	159	1	0.2	-2.7	-0.5	0.1	0.9	2.8
Poland	295	0	-0.6	-6.9	-1.3	-0.6	0.2	2.1
Portugal	124	0	-0.7	-7.6	-1.5	-0.7	0.2	2.5
Romania	8	0	-1.9	-3.8	-3.0	-1.7	-0.9	-0.1
Russian Federation	712	0	-1.2	-9.2	-2.0	-1.1	-0.2	2.3
Serbia	54	0	-0.9	-3.3	-1.5	-0.9	0.2	1.1
Slovak Republic	139	1	-0.4	-4.1	-1.2	-0.4	0.4	2.5
Slovenia	38	0	-0.3	-2.0	-0.7	-0.3	0.4	1.5
Spain	977	4	-0.4	-4.9	-1.0	-0.3	0.4	3.0
Sweden[2]	338	10	0.0	-3.8	-0.6	0.1	0.7	2.9
Switzerland	467	1	-0.4	-4.9	-1.0	-0.3	0.2	2.1
Turkey	220	0	-1.2	-6.3	-2.0	-0.9	-0.0	1.8
Ukraine	27	0	-1.3	-4.6	-1.8	-1.2	-0.6	0.8
United Kingdom[3]	5213	49	-0.1	-7.4	-0.8	-0.1	0.6	3.2
Total	21511	284	-0.4	-9.2	-1.0	-0.3	0.4	3.5

[1] Norway: sometimes any value (instead of last of the year) for height is used when no lung function test was available.
[2] Sweden: if there is no lung function measurement, the height and weight reported are those measured at annual review.
[3] United Kingdom: height and weight at date of annual data is used instead of the date of best FEV1. If no lung function measurement is reported, the date of the last visit is used.

Note: Georgia has 0% coverage for adults and is excluded from the table.
Note: Albania, Armenia, Belarus have <5 patients aged 18 years or more at height measurement and are excluded from the table.

Table 6.5 reports the median z-score for weight (the value that separates the highest and lowest half of the patients), the mean z-score for weight (the average) and other descriptive statistics for adults (18 years or older).

Figure 6.4 Median z-scores for weight by age group and by country. All patients seen in 2019 who never had a transplant.



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

This graph shows the median z-scores for weight by age group. Each country is represented by a dot (in blue) and the overall estimate is in red. Overall, the median z-scores for weight decrease from the third youngest age group to the 15-19 and 20-24 years age groups before they increase in the older age groups. Again, the patients in the oldest age groups are patients that survived, and may therefore represent the patients with less disease severity. There is considerable variability between countries.

Table 6.6 Z-scores for weight: descriptive statistics by age group. All patients seen in 2019 who never had a transplant.

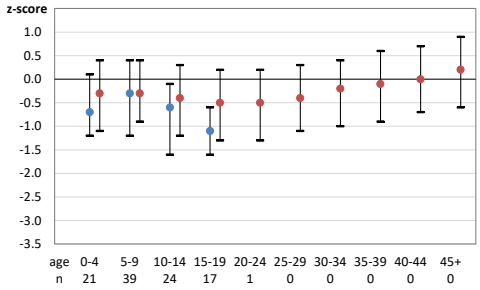
Age at weight measurement	N	Mean	Min	25th pctl	Median	75th pctl	Max
0-4	6210	-0.4	-9.1	-1.1	-0.3	0.4	8.7
5-9	7079	-0.3	-7.9	-0.9	-0.3	0.4	3.1
10-14	6590	-0.5	-7.1	-1.2	-0.4	0.3	3.1
15-19	5708	-0.6	-9.2	-1.3	-0.5	0.2	2.9
20-24	4910	-0.6	-7.6	-1.3	-0.5	0.2	3.0
25-29	4214	-0.4	-6.3	-1.1	-0.4	0.3	3.2
30-34	3346	-0.3	-6.8	-1.0	-0.2	0.4	3.4
35-39	2406	-0.2	-5.8	-0.9	-0.1	0.6	2.9
40-44	1634	-0.0	-4.9	-0.7	-0.0	0.7	3.1
45+	2843	0.1	-7.1	-0.6	0.2	0.9	3.5

This table reports the median z-score for weight and other descriptive statistics by age group for all the patients seen in 2019 who never had a transplant. The median values reported in this table are shown as red dots in fig 6.4.

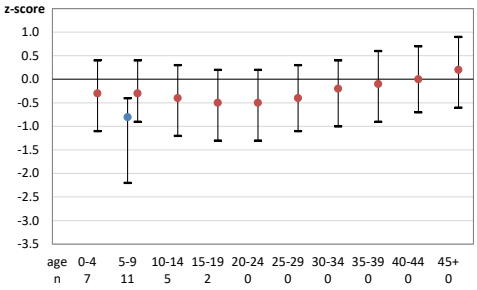
Figure 6.5 Quartiles of z-scores for weight by age group and by country. All patients seen in 2019 who never had a transplant.

The figures below show the z-scores for weight by country. The dot is the median, and the whiskers show the 25th and 75th percentiles. In blue are the quartiles for the country, in red are the pooled quartiles computed on all other countries (i.e. excluding that country). We did not compute quartiles where the number of patients in the age group is <10. Therefore, there are no blue dots for those age groups (the number of patients in each age group is shown underneath the horizontal axis). We therefore excluded Lithuania and Luxembourg from the graphs because none of the age groups in these countries had more than 10 patients.

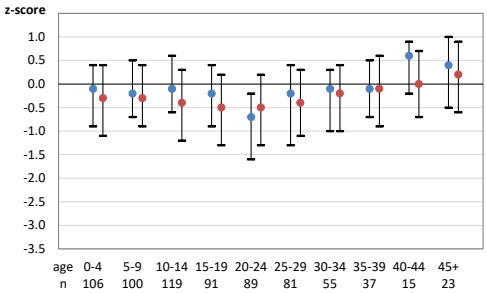
Quartiles of z-scores for weight: Albania



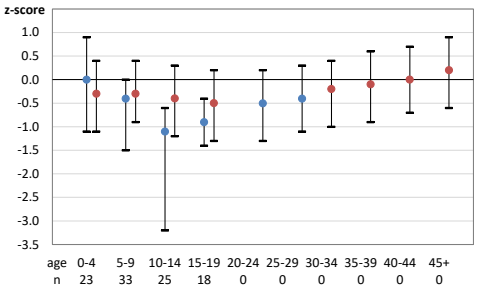
Quartiles of z-scores for weight: Armenia



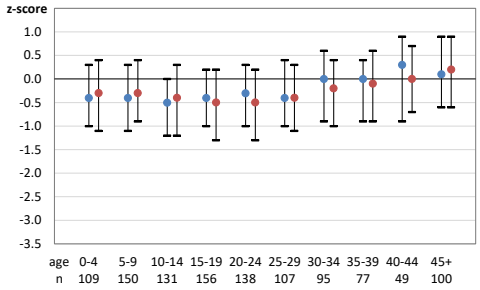
Quartiles of z-scores for weight: Austria



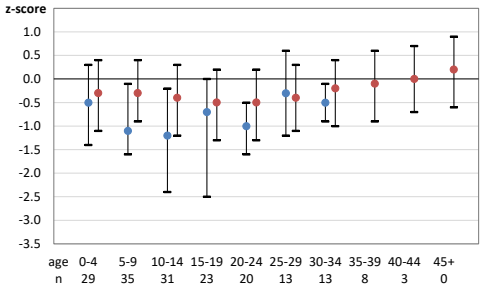
Quartiles of z-scores for weight: Belarus



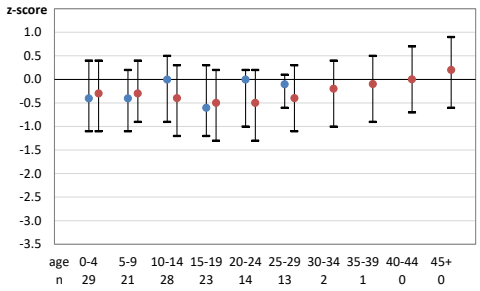
Quartiles of z-scores for weight: Belgium



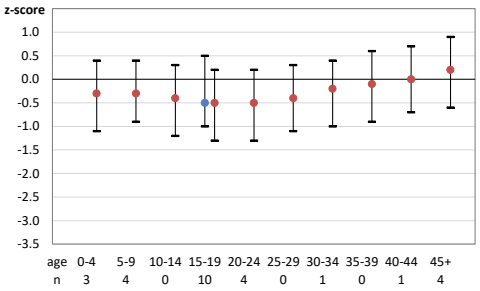
Quartiles of z-scores for weight: Bulgaria



Quartiles of z-scores for weight: Croatia

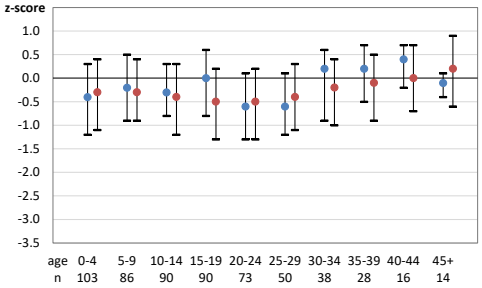


Quartiles of z-scores for weight: Cyprus

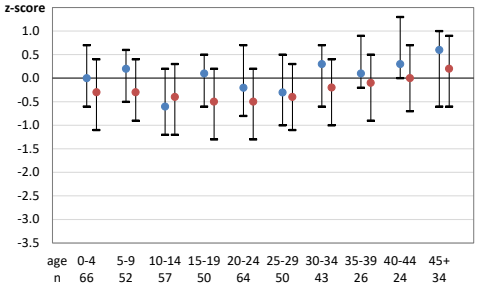


[figure 6.5 continued]

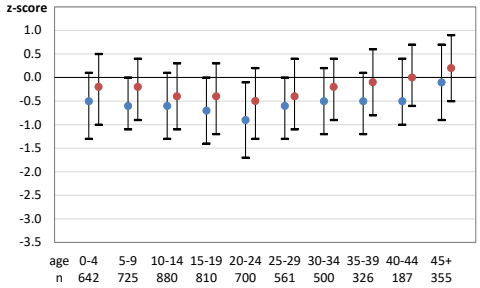
Quartiles of z-scores for weight: Czech Republic



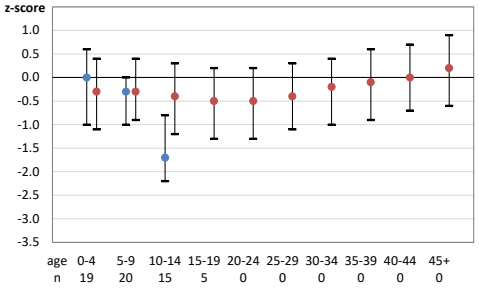
Quartiles of z-scores for weight: Denmark



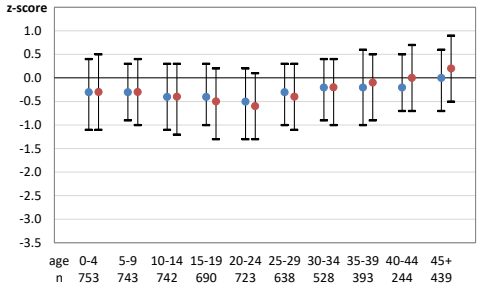
Quartiles of z-scores for weight: France



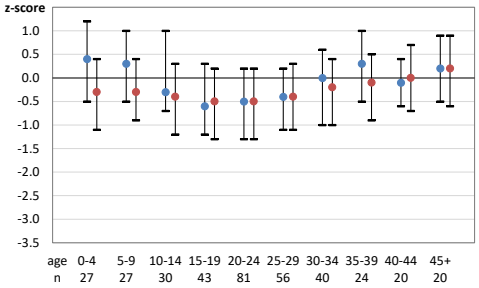
Quartiles of z-scores for weight: Georgia



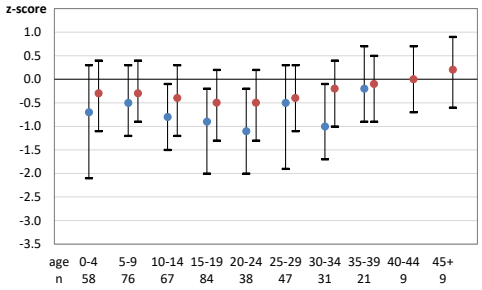
Quartiles of z-scores for weight: Germany



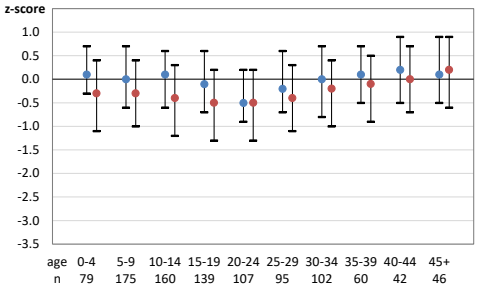
Quartiles of z-scores for weight: Greece



Quartiles of z-scores for weight: Hungary



Quartiles of z-scores for weight: Ireland



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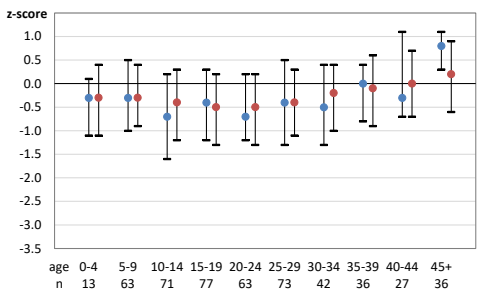
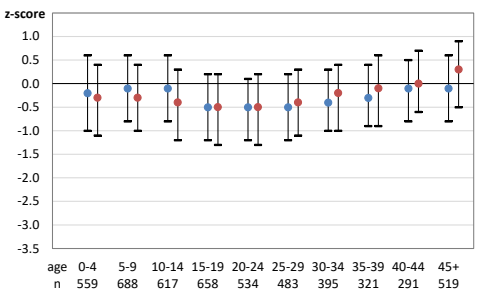
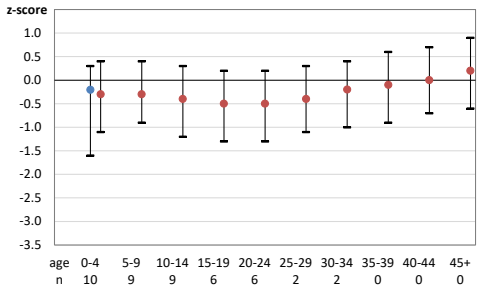


Quartiles of z-scores for weight: Italy

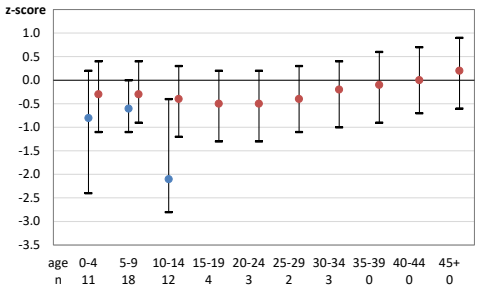


[figure 6.5 continued]

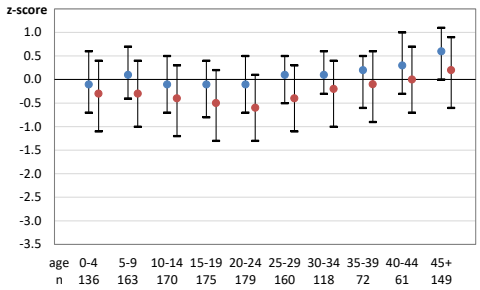
Quartiles of z-scores for weight: Latvia



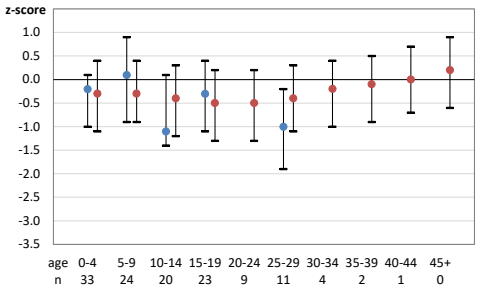
Quartiles of z-scores for weight: Rep of Moldova



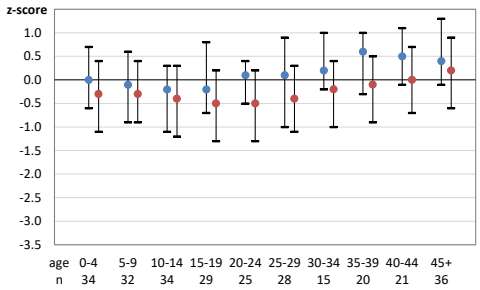
Quartiles of z-scores for weight: The Netherlands



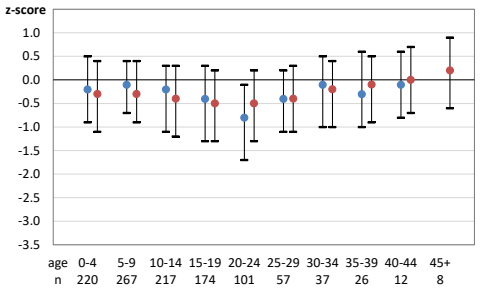
Quartiles of z-scores for weight: North Macedonia



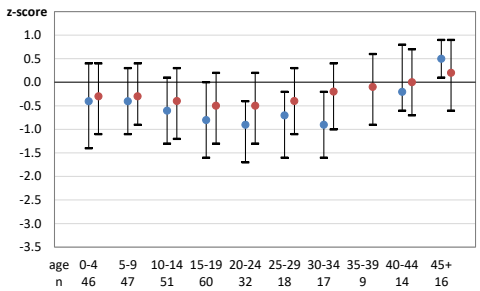
Quartiles of z-scores for weight: Norway[1]



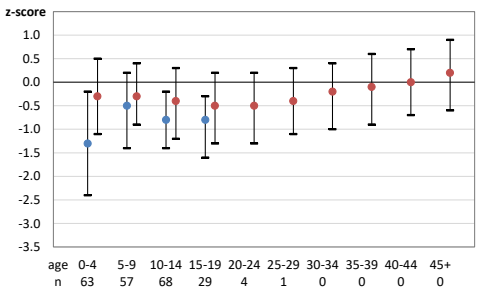
Quartiles of z-scores for weight: Poland



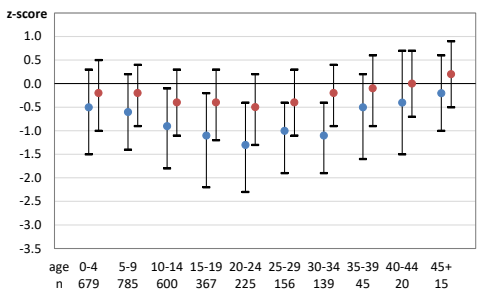
Quartiles of z-scores for weight: Portugal



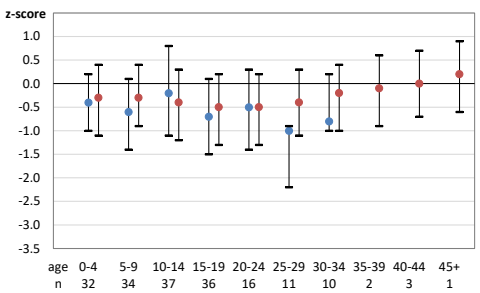
Quartiles of z-scores for weight: Romania



Quartiles of z-scores for weight: Russian Federation

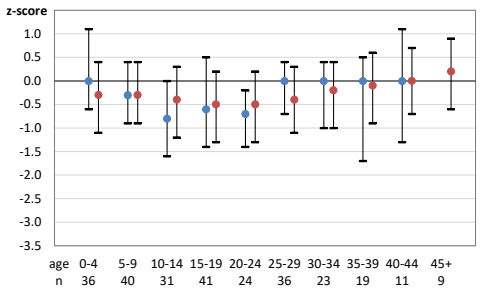


Quartiles of z-scores for weight: Serbia

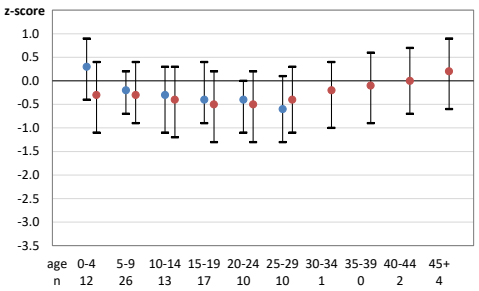


[figure 6.5 continued]

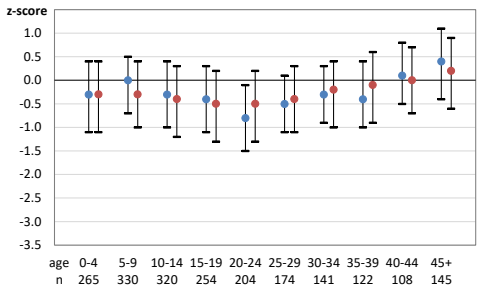
Quartiles of z-scores for weight: Slovak Republic



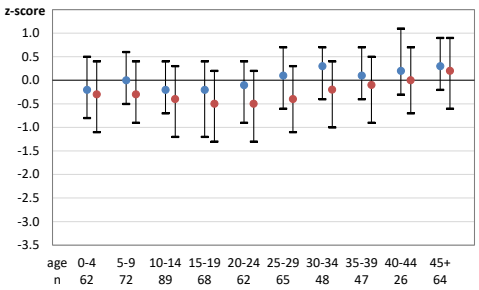
Quartiles of z-scores for weight: Slovenia



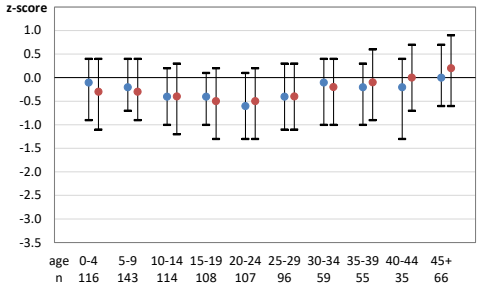
Quartiles of z-scores for weight: Spain



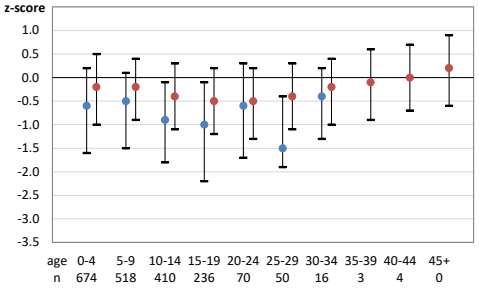
Quartiles of z-scores for weight: Sweden[2]



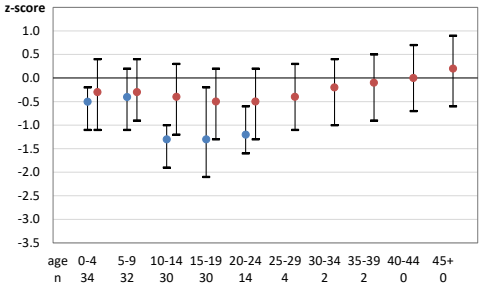
Quartiles of z-scores for weight: Switzerland



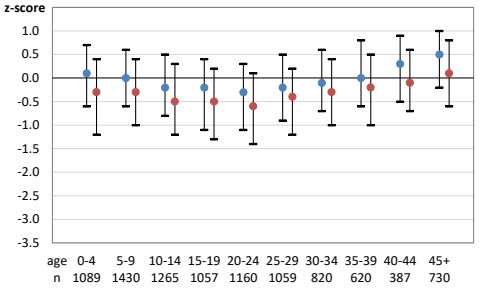
Quartiles of z-scores for weight: Turkey



Quartiles of z-scores for weight: Ukraine



Quartiles of z-scores for weight: United Kingdom[3]



[1] Norway: sometimes any value (instead of last of the year) for height is used when no lung function test was available.
[2] Sweden: if there is no lung function measurement, the height and weight reported are those measured at annual review.
[3] United Kingdom: height and weight at date of annual data is used instead of the date of best FEV1. If no lung function measurement is reported, the date of the last visit is used.

Table 6.7 Z-scores for BMI: descriptive statistics by country. All patients seen in 2019 aged 2-17 years who never had a transplant.

Country	N	N Miss	Mean	Min	25th pctl (25% of the patients are below this z-score for BMI)	Median (50% of the patients are below this z-score for BMI)	75th pctl (75% of the patients are below this z-score for BMI)	Max
Albania	81	13	-0.4	-5.0	-1.3	-0.2	0.4	2.8
Armenia	25	0	-0.7	-6.8	-1.1	-0.6	0.2	2.3
Austria	346	0	-0.2	-3.9	-0.8	-0.2	0.4	2.5
Belarus	84	3	-0.9	-6.8	-1.7	-0.7	0.0	1.7
Belgium	440	0	-0.3	-2.9	-0.9	-0.3	0.3	2.6
Bulgaria	97	0	-0.8	-6.7	-1.6	-0.8	0.1	2.3
Croatia	75	2	-0.5	-5.5	-1.2	-0.4	0.1	3.7
Cyprus	12	0	-0.6	-3.2	-1.1	-0.5	-0.3	1.7
Czech Republic	292	0	-0.3	-3.5	-0.9	-0.3	0.3	2.2
Denmark	179	0	-0.2	-3.3	-0.8	-0.2	0.3	1.9
France	2523	22	-0.5	-9.9	-1.1	-0.4	0.2	3.5
Georgia	50	0	0.1	-2.9	-0.6	0.1	0.7	2.9
Germany	2380	3	-0.3	-6.5	-0.9	-0.3	0.3	2.8
Greece	94	1	0.2	-3.0	-0.5	0.4	1.0	3.1
Hungary	228	1	-0.7	-5.3	-1.5	-0.6	0.1	2.6
Ireland	457	27	0.2	-2.4	-0.3	0.2	0.8	2.8
Israel	197	0	-0.3	-5.2	-0.9	-0.2	0.6	2.2
Italy	2086	25	-0.1	-4.3	-0.8	-0.1	0.6	3.9
Latvia	25	0	-0.9	-2.3	-1.6	-1.1	-0.2	0.8
Lithuania	12	0	-1.0	-2.6	-1.5	-1.0	-0.4	0.4
Luxembourg	12	0	-0.3	-1.2	-0.9	-0.3	0.2	0.9
Rep of Moldova	43	0	-0.9	-4.2	-1.9	-1.1	0.1	2.9
The Netherlands	526	0	-0.2	-3.6	-0.8	-0.1	0.4	1.8
North Macedonia	73	0	-0.2	-4.5	-1.2	-0.2	0.7	2.1
Norway[1]	102	0	-0.3	-2.7	-1.0	-0.3	0.4	1.7
Poland	737	5	-0.3	-5.6	-1.0	-0.2	0.4	2.6
Portugal	172	1	-0.3	-2.7	-0.9	-0.3	0.3	1.7
Romania	187	5	-1.0	-9.2	-1.7	-0.7	0.0	1.9
Russian Federation	2077	9	-0.7	-9.3	-1.5	-0.6	0.2	4.0
Serbia	117	0	-0.6	-5.0	-1.2	-0.6	0.2	2.4
Slovak Republic	118	4	-0.5	-3.7	-1.3	-0.5	0.2	2.3
Slovenia	56	0	-0.4	-3.0	-1.0	-0.3	0.2	1.9
Spain	995	3	-0.2	-4.6	-0.8	-0.1	0.5	2.6
Sweden[2]	244	0	-0.2	-3.0	-0.7	-0.1	0.5	2.0
Switzerland	395	2	-0.2	-3.1	-0.7	-0.2	0.4	3.9
Turkey	1532	0	-0.7	-9.3	-1.6	-0.6	0.3	4.4
Ukraine	114	1	-1.0	-6.5	-1.6	-1.0	-0.2	2.7
United Kingdom[3]	4091	26	0.1	-7.0	-0.5	0.1	0.7	3.7
Total	21274	153	-0.3	-9.9	-0.9	-0.2	0.4	4.4

[1] Norway: sometimes any value (instead of last of the year) for height is used when no lung function test was available.
[2] Sweden: if there is no lung function measurement, the height and weight reported are those measured at annual review.
[3] United Kingdom: height and weight at date of annual data is used instead of the date of best FEV1. If no lung function measurement is reported, the date of the last visit is used.

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

Table 6.8 BMI: descriptive statistics by country. All patients seen in 2019 aged 18 years or older who never had a transplant.

Country	N	N Miss	Mean	Min	25th pctl (25% of the patients are below this BMI)	Median (50% of the patients are below this BMI)	75th pctl (75% of the patients are below this BMI)	Max
Austria	335	0	21.8	14.9	19.5	21.3	23.6	38.5
Belgium	637	2	22.5	15.1	20.2	22.1	24.2	36.5
Bulgaria	67	0	20.2	12.3	17.9	19.7	21.8	40.1
Croatia	39	0	21.5	15.1	20.2	21.8	23.1	27.8
Cyprus	13	0	23.5	19.1	21.6	22.6	24.1	34.7
Czech Republic	253	0	21.9	13.7	19.7	21.3	23.9	34.4
Denmark	259	0	22.6	13.8	20.0	22.3	24.1	39.3
France	2921	50	21.6	12.1	19.4	21.1	23.1	46.7
Germany	3235	13	21.8	13.1	19.4	21.3	23.5	42.1
Greece	265	4	22.6	15.0	20.5	22.1	24.2	37.0
Hungary	189	6	20.5	14.4	18.6	20.1	22.3	30.8
Ireland	509	108	23.1	15.2	20.8	22.6	24.8	44.7
Israel	303	0	23.1	16.2	20.4	22.7	25.2	39.7
Italy	2792	52	22.5	14.2	20.1	21.9	24.3	49.5
Latvia	13	0	19.1	16.2	17.3	19.5	20.8	24.0
Lithuania	11	0	21.6	18.0	19.6	21.0	24.7	26.3
Luxembourg	20	0	23.0	17.3	21.3	22.6	24.0	40.8
Rep of Moldova	8	0	18.6	15.8	17.0	18.9	19.9	21.5
The Netherlands	811	0	22.6	14.3	20.5	22.1	24.1	46.7
North Macedonia	36	0	22.0	16.6	20.3	21.8	23.8	26.5
Norway[1]	159	1	23.1	17.0	19.9	22.5	25.7	37.7
Poland	294	1	21.1	13.3	18.6	20.6	23.0	37.3
Portugal	119	5	22.2	12.3	19.8	21.3	24.0	39.0
Romania	8	0	18.2	14.2	16.3	17.7	19.5	23.9
Russian Federation	712	0	19.7	12.0	17.5	19.2	21.2	35.4
Serbia	54	0	19.8	13.7	17.9	19.6	21.5	25.1
Slovak Republic	139	1	21.3	14.2	19.0	20.9	23.6	34.6
Slovenia	38	0	21.0	15.1	19.7	21.3	22.0	25.2
Spain	976	5	22.7	12.2	20.2	22.2	24.3	43.4
Sweden[2]	338	10	22.8	13.9	20.4	22.3	24.4	39.0
Switzerland	467	1	21.7	13.8	19.4	21.3	23.4	36.6
Turkey	220	0	20.5	12.9	18.0	19.9	23.0	31.8
Ukraine	27	0	19.8	16.4	17.5	19.9	21.3	25.9
United Kingdom[3]	5213	49	23.2	12.1	20.6	22.6	25.2	49.3
Total	21487	308	22.3	12.0	19.8	21.8	24.1	49.5

[1] Norway: sometimes any value (instead of last of the year) for height is used when no lung function test was available.
[2] Sweden: if there is no lung function measurement, the height and weight reported are those measured at annual review.
[3] United Kingdom: height and weight at date of annual data is used instead of the date of best FEV1. If no lung function measurement is reported, the date of the last visit is used.

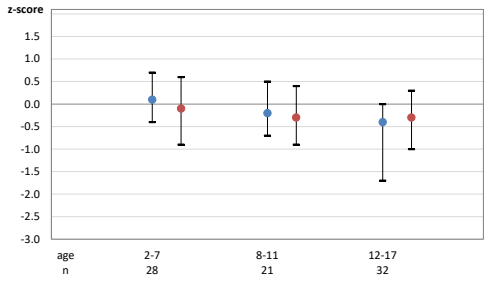
Note: Georgia has 0% coverage for adults and is excluded from the table.
Note: Albania, Armenia, Belarus have <5 patients aged 18 years or more at BMI measurement and are excluded from the table.

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

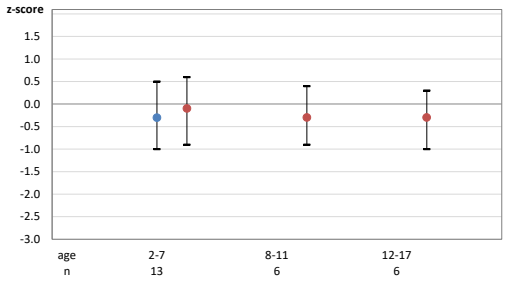
Figure 6.6 Quartiles of z-scores for BMI by age group and country. Patients aged 2-17 years in 2019 who never had a transplant.

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. In blue are the quartiles for the country, in red are the pooled quartiles computed on all other countries (i.e. excluding that country). We did not compute quartiles where the number of patients in the age group is <10. Therefore, there are no blue dots for those age groups (the number of patients in each age group is shown underneath the horizontal axis). We therefore excluded Cyprus, Latvia, Lithuania and Luxembourg from the graphs because none of the age groups in these countries had more than 10 patients.

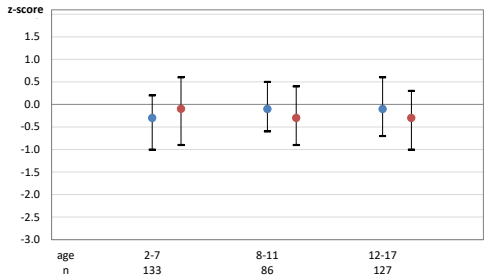
Quartiles of z-scores for BMI: Albania



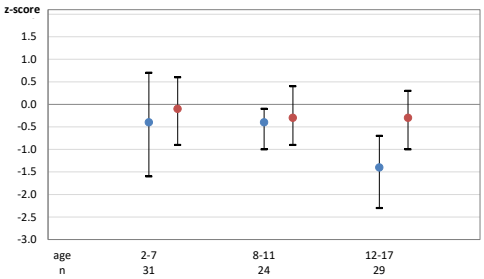
Quartiles of z-scores for BMI: Armenia



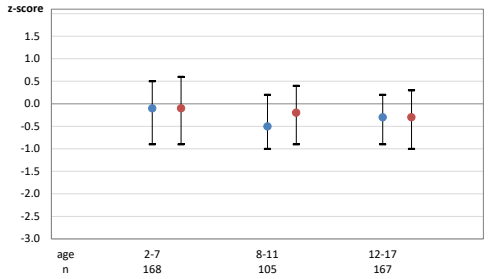
Quartiles of z-scores for BMI: Austria



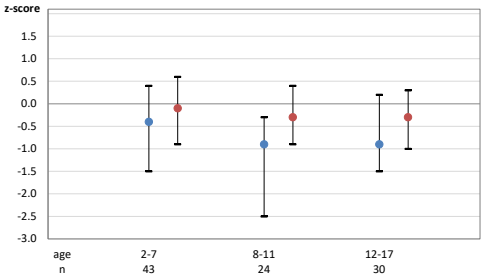
Quartiles of z-scores for BMI: Belarus



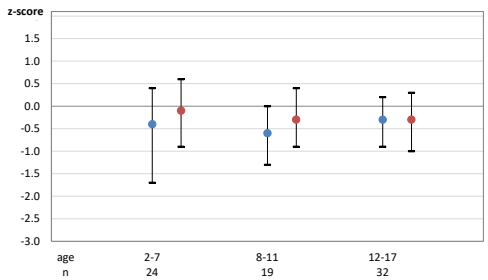
Quartiles of z-scores for BMI: Belgium



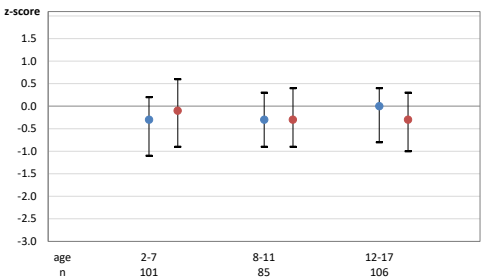
Quartiles of z-scores for BMI: Bulgaria



Quartiles of z-scores for BMI: Croatia

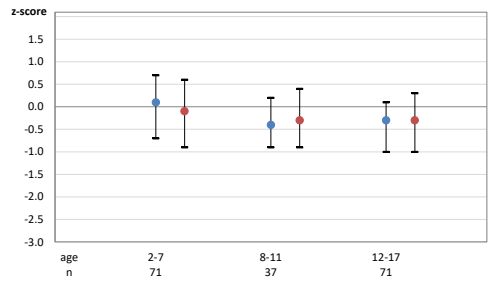


Quartiles of z-scores for BMI: Czech Republic

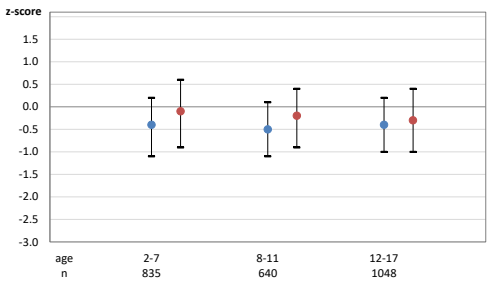


[figure 6.6 continued]

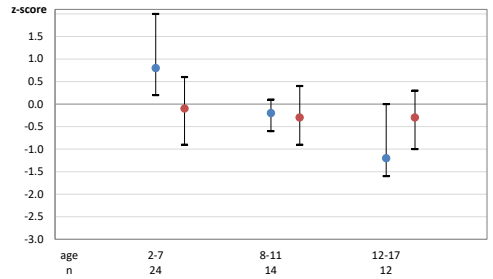
Quartiles of z-scores for BMI: Denmark



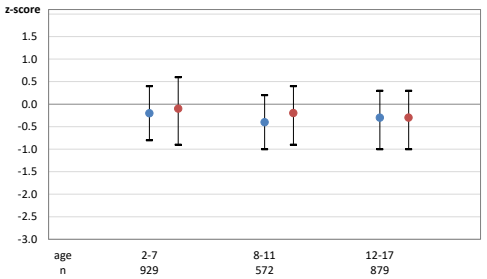
Quartiles of z-scores for BMI: France



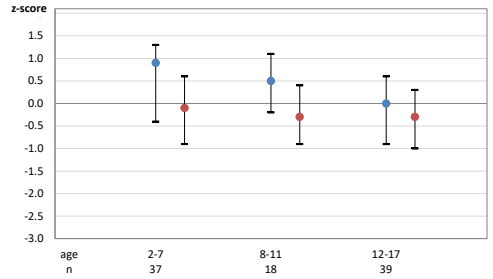
Quartiles of z-scores for BMI: Georgia



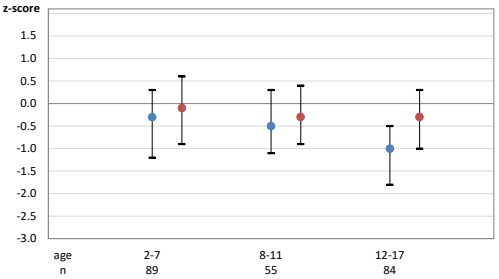
Quartiles of z-scores for BMI: Germany



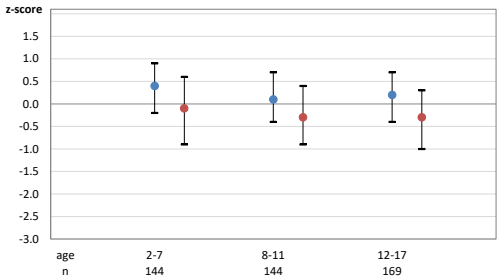
Quartiles of z-scores for BMI: Greece



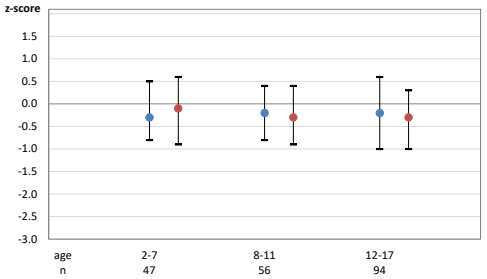
Quartiles of z-scores for BMI: Hungary



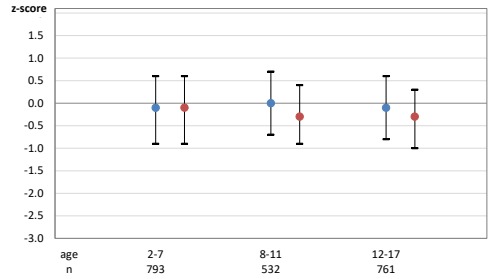
Quartiles of z-scores for BMI: Ireland



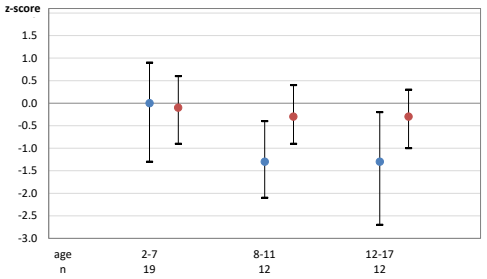
Quartiles of z-scores for BMI: Israel



Quartiles of z-scores for BMI: Italy

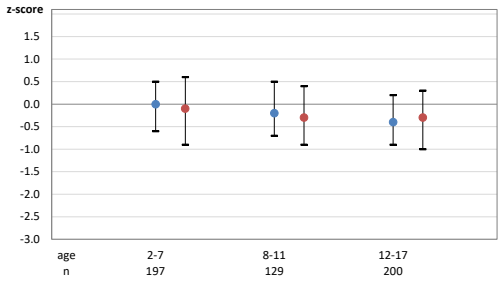


Quartiles of z-scores for BMI: Rep. of Moldova

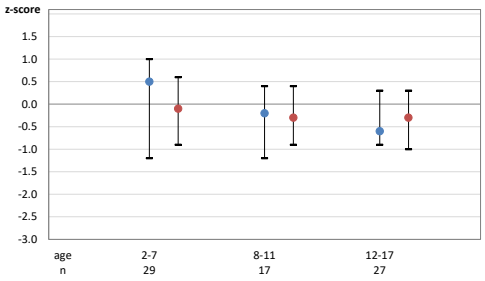


[figure 6.6 continued]

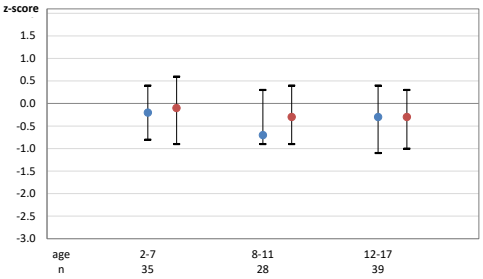
Quartiles of z-scores for BMI: The Netherlands



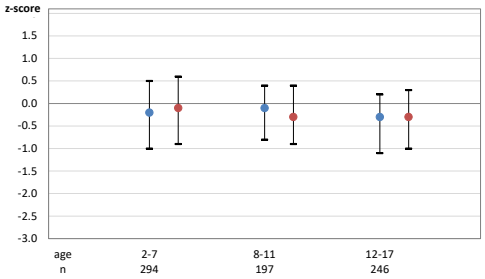
Quartiles of z-scores for BMI: North Macedonia



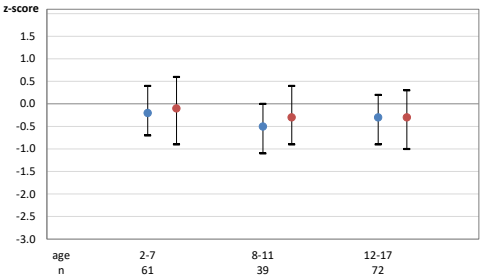
Quartiles of z-scores for BMI: Norway



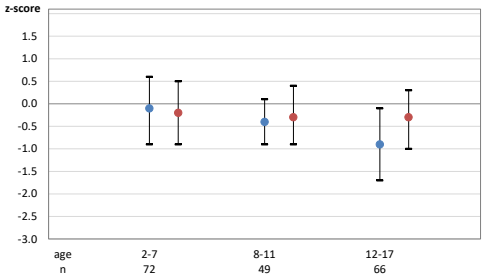
Quartiles of z-scores for BMI: Poland



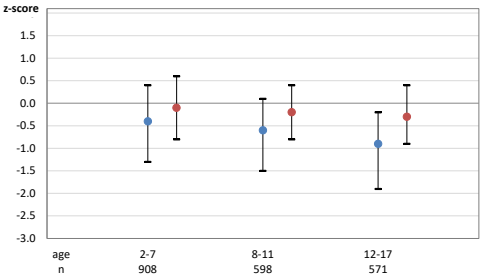
Quartiles of z-scores for BMI: Portugal



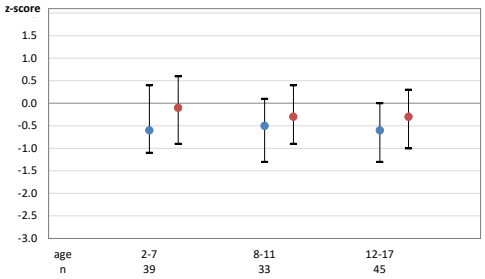
Quartiles of z-scores for BMI: Romania



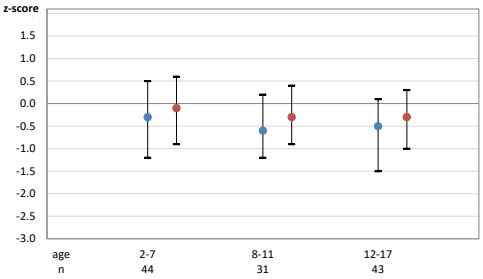
Quartiles of z-scores for BMI: Russian Federation



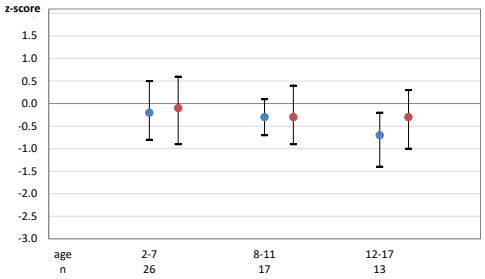
Quartiles of z-scores for BMI: Serbia



Quartiles of z-scores for BMI: Slovak Republic

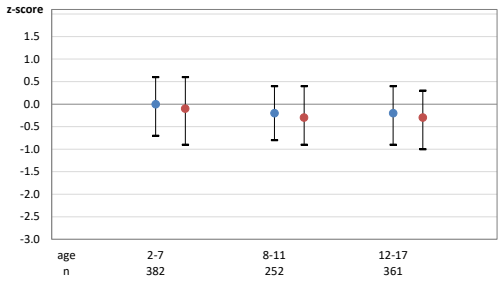


Quartiles of z-scores for BMI: Slovenia

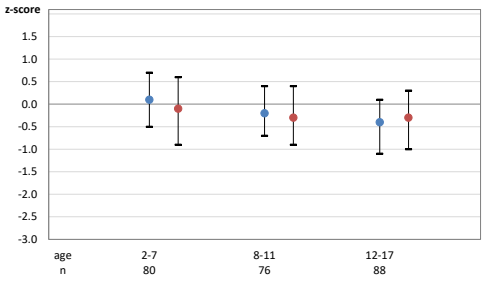


[figure 6.6 continued]

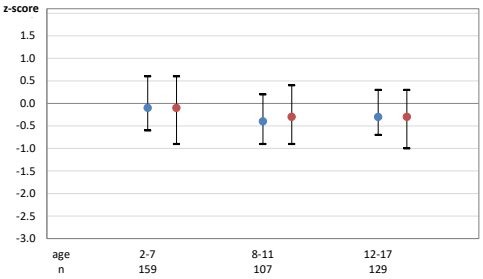
Quartiles of z-scores for BMI: Spain



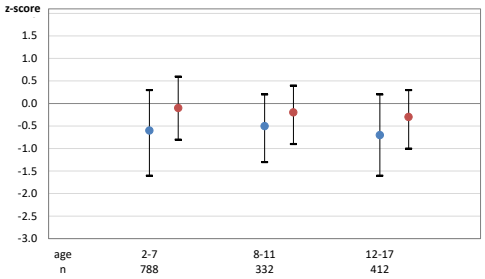
Quartiles of z-scores for BMI: Sweden



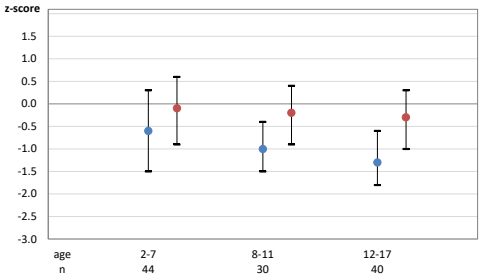
Quartiles of z-scores for BMI: Switzerland



Quartiles of z-scores for BMI: Turkey



Quartiles of z-scores for BMI: Ukraine



Quartiles of z-scores for BMI: United Kingdom

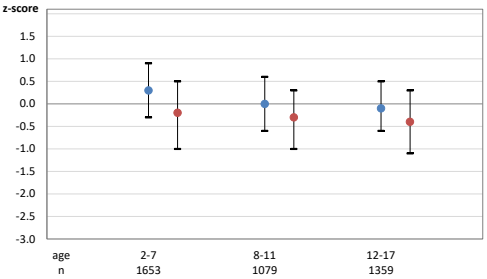
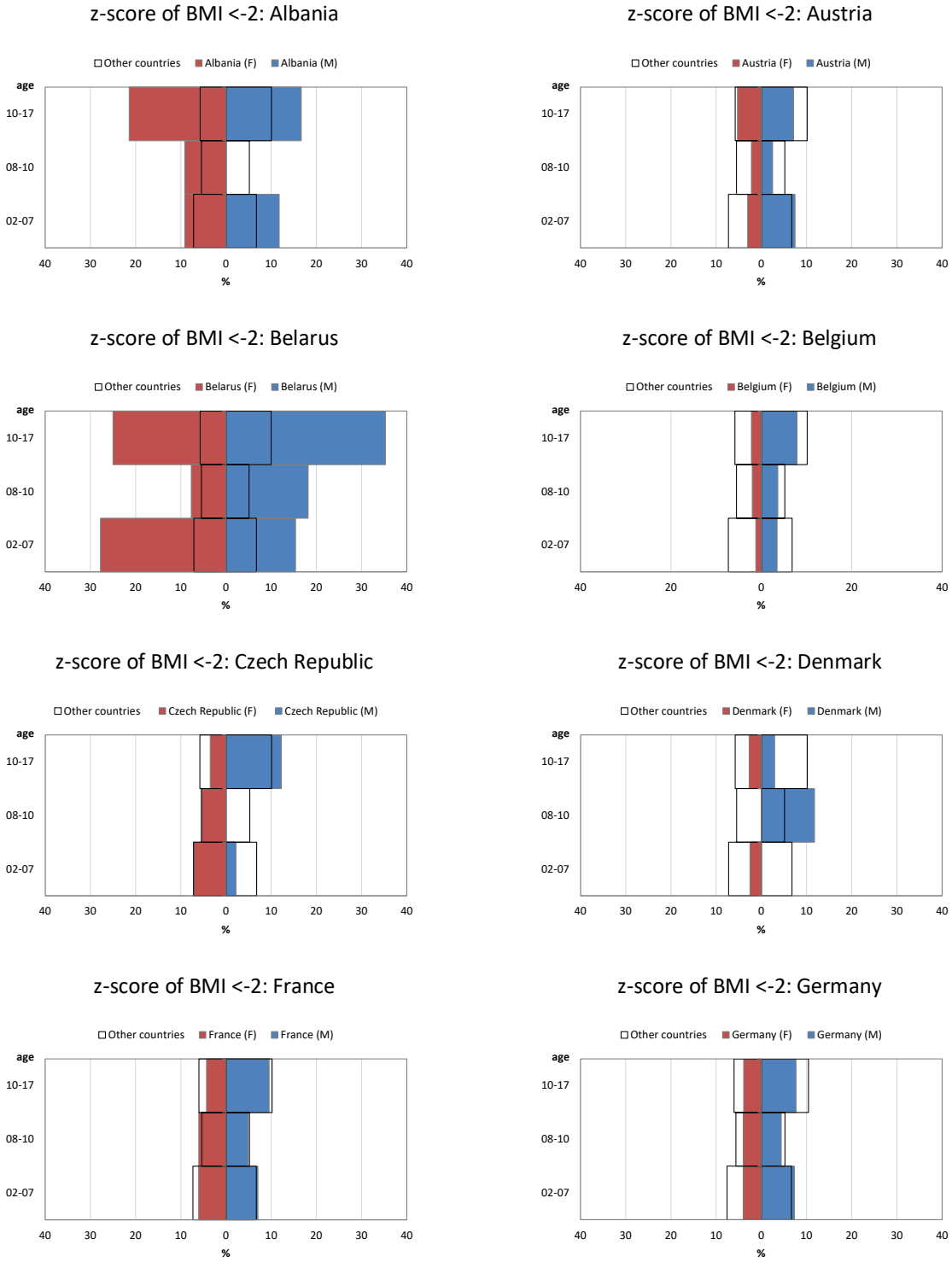


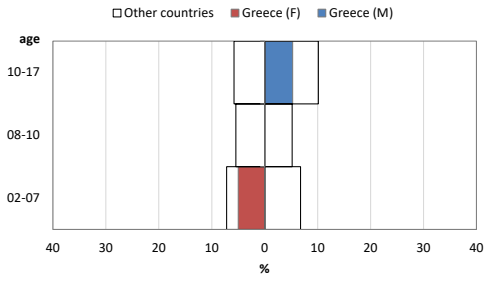
Figure 6.7 Proportion of children underweight (z-score of BMI<-2): age and sex pyramids, by country and overall. Patients aged 2-17 years in 2019 who never had a transplant.

The coloured bars (red for females, blue for males) represent the percentage of underweight patients in the selected country, whereas the non-coloured bars represent the percentage of underweight patients in all the remaining countries (i.e. excluding that country). We excluded the graphs of Armenia, Bulgaria, Croatia, Cyprus, Georgia, Latvia, Lithuania, Luxembourg, North Macedonia, Republic of Moldova and Slovenia because at least one group (age/sex) in these countries had fewer than 10 patients.

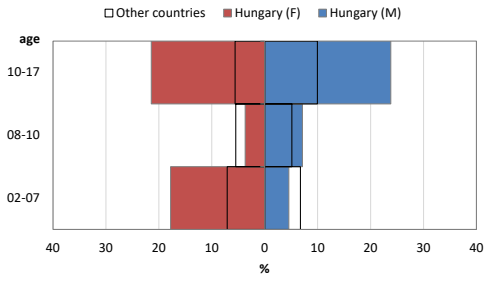


[figure 6.7 continued]

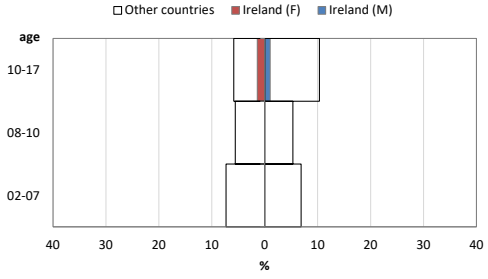
z-score of BMI <-2: Greece



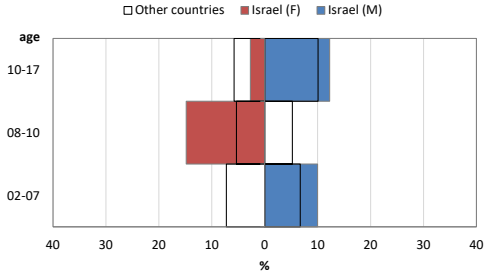
z-score of BMI <-2: Hungary



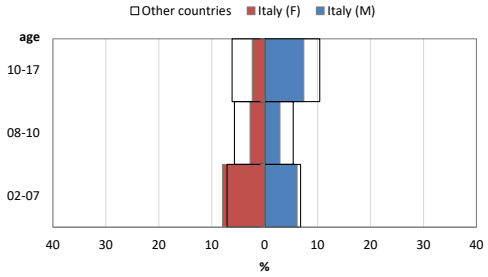
z-score of BMI <-2: Ireland



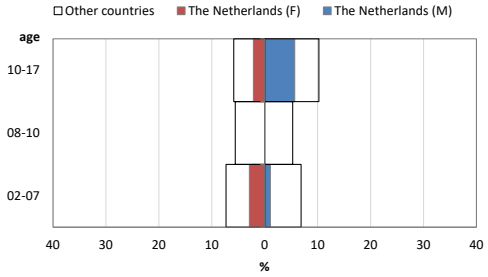
z-score of BMI <-2: Israel



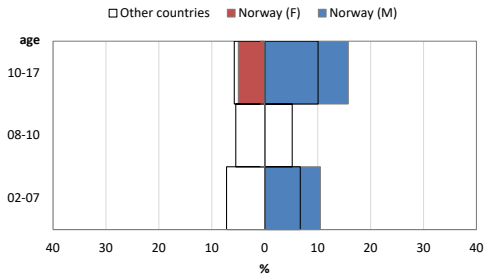
z-score of BMI <-2: Italy



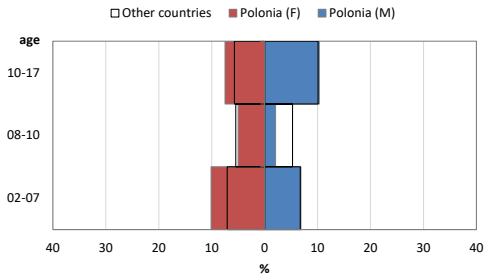
z-score of BMI <-2: The Netherlands



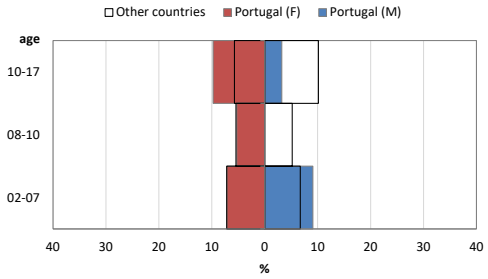
z-score of BMI <-2: Norway



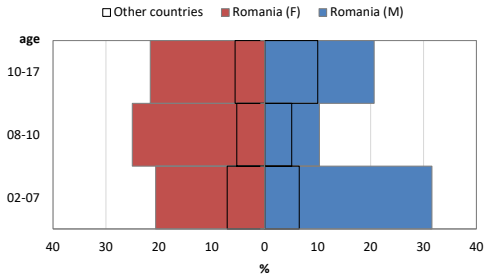
z-score of BMI <-2: Poland



z-score of BMI <-2: Portugal

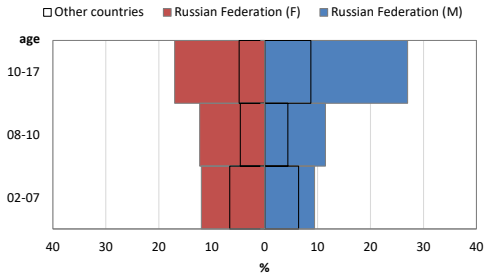


z-score of BMI <-2: Romania

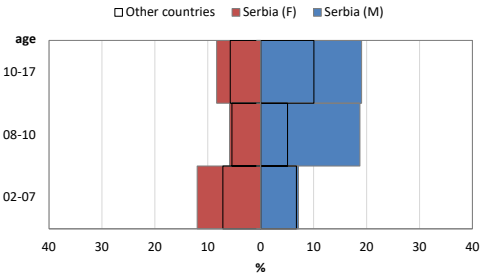


[figure 6.7 continued]

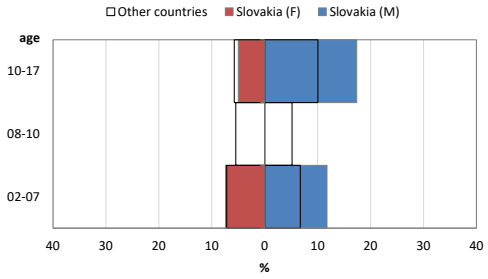
z-score of BMI <-2: Russian Federation



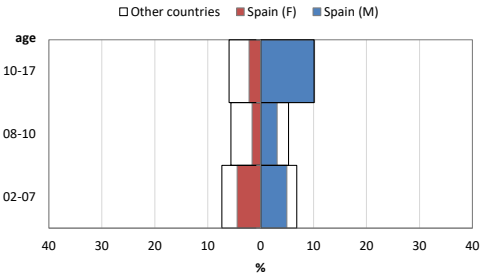
z-score of BMI <-2: Serbia



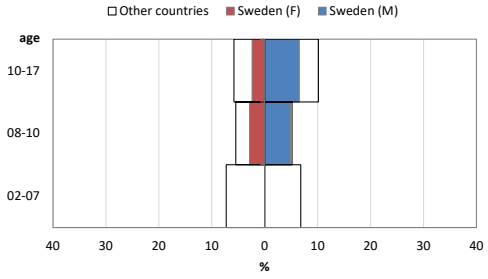
z-score of BMI <-2: Slovak Republic



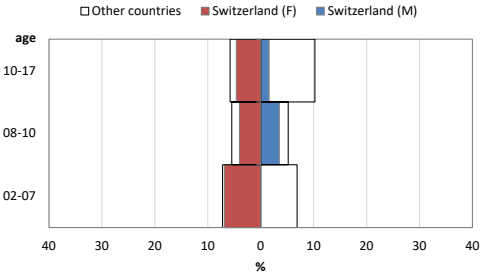
z-score of BMI <-2: Spain



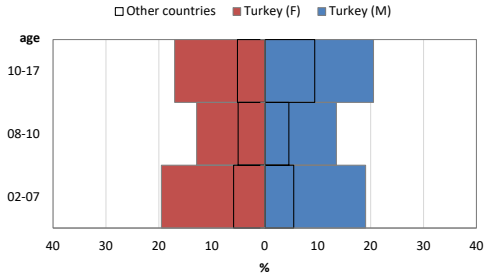
z-score of BMI <-2: Sweden



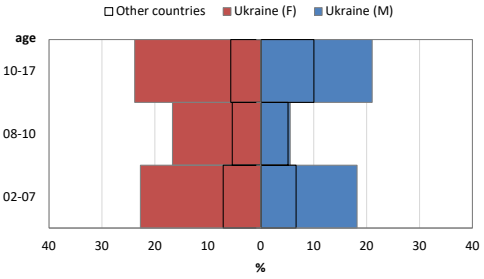
z-score of BMI <-2: Switzerland



z-score of BMI <-2: Turkey



z-score of BMI <-2: Ukraine



z-score of BMI <-2: United Kingdom

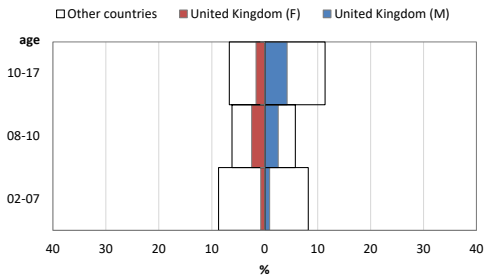
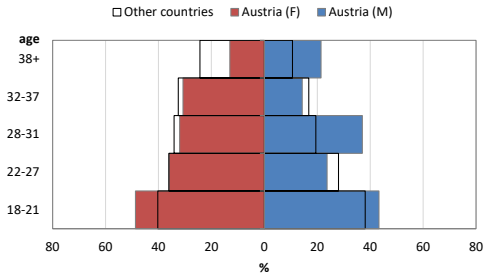


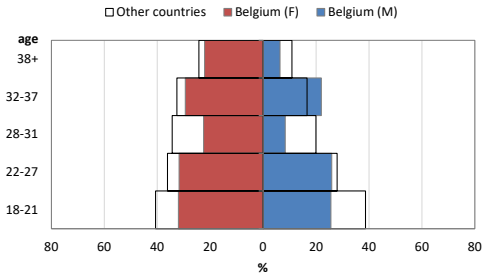
Figure 6.8 Proportion of adults with BMI<20: age and sex pyramids, by country and overall. Patients aged 18 years or older in 2019 who never had a transplant.

The coloured bars (red for females, blue for males) represent the percentage of underweight patients in the selected country, whereas the non-coloured bars represent the percentage of underweight patients in all the remaining countries (i.e. excluding that country). We therefore excluded from the graphs Albania, Armenia, Belarus, Bulgaria, Croatia, Cyprus, Georgia, Hungary, Latvia, Lithuania, Luxembourg, Republic of Moldova, North Macedonia, Norway, Poland, Portugal, Romania, Serbia, Slovak Republic, Slovenia, Turkey and Ukraine because at least one group (age/sex) in these countries had fewer than 10 patients.

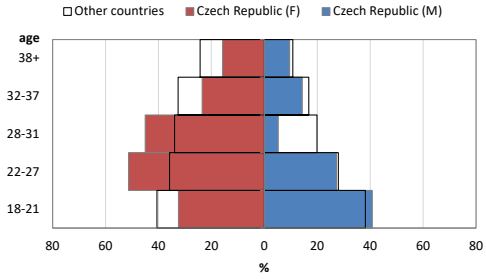
BMI<20 kg/m^2: Austria



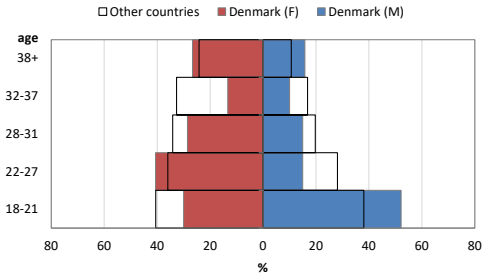
BMI<20 kg/m^2: Belgium



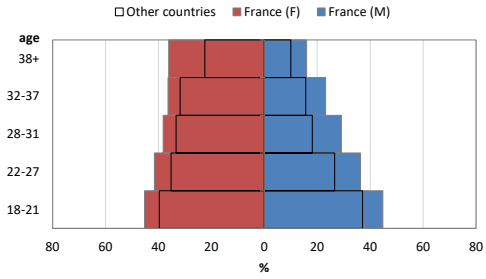
BMI<20 kg/m^2: Czech Republic



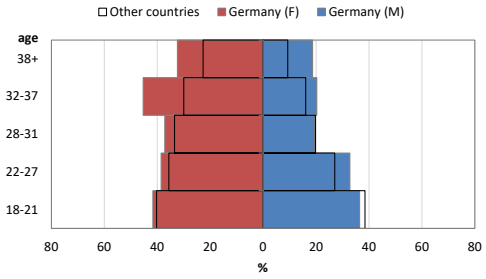
BMI<20 kg/m^2: Denmark



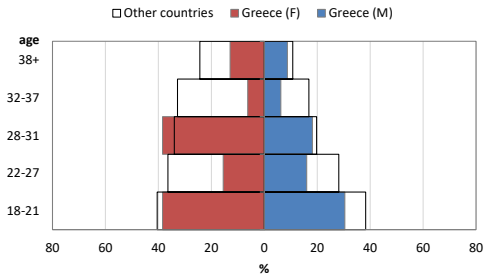
BMI<20 kg/m^2: France



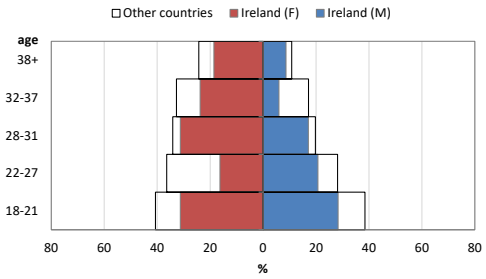
BMI<20 kg/m^2: Germany



BMI<20 kg/m^2: Greece

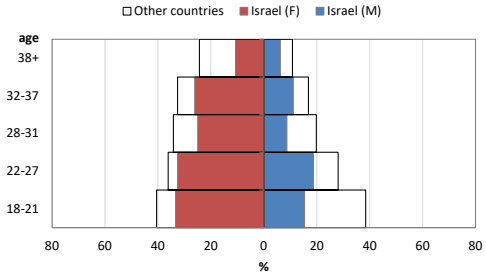


BMI<20 kg/m2: Ireland

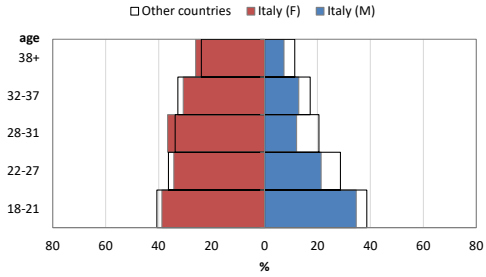


[figure 6.8 continued]

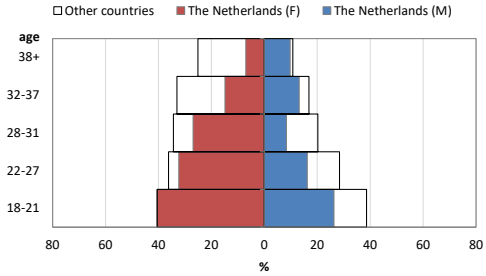
BMI<20 kg/m2: Israel



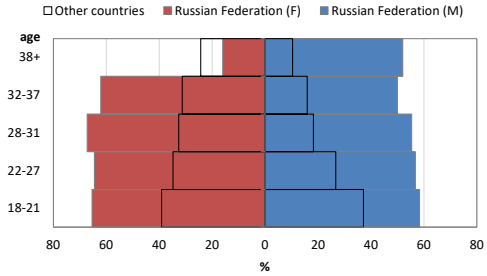
BMI<20 kg/m^2: Italy



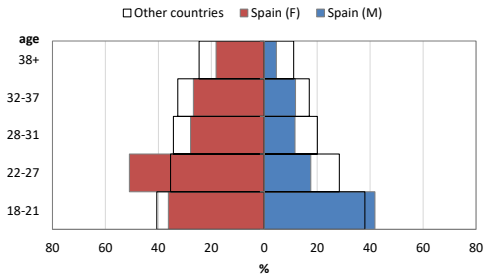
BMI<20 kg/m^2: The Netherlands



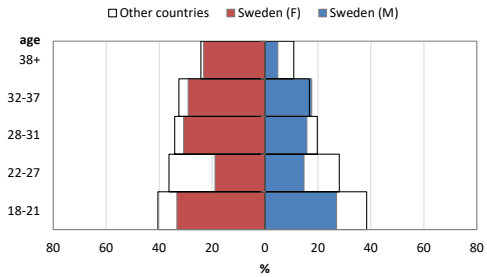
BMI<20 kg/m^2: Russian Federation



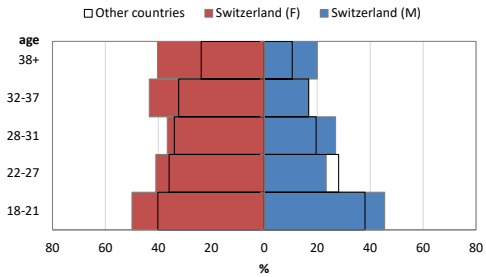
BMI<20 kg/m2: Spain



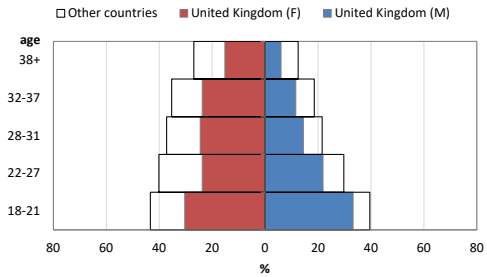
BMI<20 kg/m2: Sweden



BMI<20 kg/m2: Switzerland



BMI<20 kg/m2: United Kingdom



7. Complications and Therapy

The information in this section should not be considered complete, for several reasons: national registries may use a different definition or different parameters for a complication; data about one or more of the complications is not collected; the status of the complication is truly unknown (e.g. liver disease, where the definition requires ultrasound examination and this is not always done for a patient). In the tables, therefore, we show the number of missing values for the various complications, but in the graphs we have included only countries where less than 10% of the data was missing. For a full list of complications and definitions please see Appendix 3 on page 156.

In this section we also present data on selected therapies. We collected information on therapies using the generic name of the drug, not the brand name. For example, instead of naming individual antibiotics, we ask whether the patient has been taking “inhaled antibiotics for more than three months this year”.

We have included information about CFTR modulators, therapies that target defects in the structure and function of the cystic fibrosis transmembrane conductance regulator protein. We also present approval status and availability of these therapies in each country in order to aid interpretation of results and understand the country-specific variations in therapy use.

Table 7.1 Prevalence of allergic bronchopulmonary aspergillosis (ABPA) (all patients seen in 2019 who never had a transplant) and CF-related diabetes (CFRD) in 2019 (patients aged 18 years or older seen in 2019 who never had a transplant), by country and overall.

Country	ABPA this year number (%)			CFRD this year number (%)					
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes, treated with daily insulin	Yes, treated with oral hypo-glycaemic agents	Yes, dietary advice only	Yes, therapy unknown
Albania	1 (0.94)	105 (99.06)	0 (0.00)	0 (0.00)	7 (77.78)	2 (22.22)	0 (0.00)	0 (0.00)	0 (0.00)
Armenia	0 (0)	25 (100)	0 (0)	-	-	-	-	-	-
Austria	2 (0.28)	696 (96.93)	20 (2.79)	3 (0.88)	249 (72.81)	79 (23.10)	1 (0.29)	10 (2.92)	0 (0.00)
Belarus	0 (0.00)	154 (99.35)	1 (0.65)	0 (0)	8 (100)	0 (0)	0 (0)	0 (0)	0 (0)
Belgium	6 (0.54)	1004 (90.12)	104 (9.34)	0 (0.00)	473 (72.32)	125 (19.11)	19 (2.91)	0 (0.00)	37 (5.66)
Bulgaria	1 (0.54)	181 (97.31)	4 (2.15)	1 (1.33)	67 (89.33)	7 (9.33)	0 (0.00)	0 (0.00)	0 (0.00)
Croatia	1 (0.76)	129 (98.48)	1 (0.76)	0 (0.00)	32 (78.05)	9 (21.95)	0 (0.00)	0 (0.00)	0 (0.00)
Cyprus	0 (0)	27 (100)	0 (0)	0 (0.00)	13 (92.86)	0 (0.00)	0 (0.00)	0 (0.00)	1 (7.14)
Czech Republic	0 (0.00)	576 (97.13)	17 (2.87)	0 (0.00)	176 (65.67)	85 (31.72)	0 (0.00)	6 (2.24)	1 (0.37)
Denmark	279 (59.49)	186 (39.66)	4 (0.85)	0 (0.00)	197 (72.96)	73 (27.04)	0 (0.00)	0 (0.00)	0 (0.00)
France	0 (0.00)	5325 (91.54)	492 (8.46)	0 (0.00)	2505 (80.75)	597 (19.25)	0 (0.00)	0 (0.00)	0 (0.00)
Georgia	1 (1.39)	71 (98.61)	0 (0.00)	-	-	-	-	-	-
Germany	63 (1.06)	5585 (94.26)	277 (4.68)	52 (1.56)	2281 (68.29)	743 (22.25)	53 (1.59)	24 (0.72)	187 (5.60)
Greece	11 (2.76)	377 (94.48)	11 (2.76)	11 (3.69)	220 (73.83)	66 (22.15)	1 (0.34)	0 (0.00)	0 (0.00)
Hungary	6 (1.35)	426 (95.73)	13 (2.92)	6 (3.08)	152 (77.95)	37 (18.97)	0 (0.00)	0 (0.00)	0 (0.00)
Ireland	2 (0.18)	1070 (94.02)	66 (5.80)	2 (0.31)	512 (80.38)	123 (19.31)	0 (0.00)	0 (0.00)	0 (0.00)
Israel	17 (3.31)	462 (90.06)	34 (6.63)	8 (2.52)	203 (63.84)	92 (28.93)	3 (0.94)	11 (3.46)	1 (0.31)
Italy	34 (0.64)	5090 (96.50)	151 (2.86)	18 (0.60)	2363 (79.38)	593 (19.92)	0 (0.00)	2 (0.07)	1 (0.03)
Latvia	2 (4.44)	43 (95.56)	0 (0.00)	1 (7.69)	11 (84.62)	1 (7.69)	0 (0.00)	0 (0.00)	0 (0.00)
Lithuania	0 (0)	27 (100)	0 (0)	0 (0)	13 (100)	0 (0)	0 (0)	0 (0)	0 (0)

[table 7.1 continued]

Country	ABPA this year number (%)			CFRD this year number (%)					
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes, treated with daily insulin	Yes, treated with oral hypo-glycaemic agents	Yes, dietary advice only	Yes, therapy unknown
Luxembourg	0 (0.00)	32 (88.89)	4 (11.11)	0 (0.00)	12 (57.14)	9 (42.86)	0 (0.00)	0 (0.00)	0 (0.00)
Rep of Moldova	0 (0.00)	52 (98.11)	1 (1.89)	0 (0.00)	7 (77.78)	2 (22.22)	0 (0.00)	0 (0.00)	0 (0.00)
The Netherlands[1]	7 (0.50)	1266 (90.82)	121 (8.68)	2 (0.24)	569 (67.42)	273 (32.35)	0 (0.00)	0 (0.00)	0 (0.00)
North Macedonia	0 (0.00)	128 (99.22)	1 (0.78)	0 (0.00)	26 (65.00)	13 (32.50)	1 (2.50)	0 (0.00)	0 (0.00)
Norway	2 (0.72)	271 (97.84)	4 (1.44)	0 (0.00)	131 (79.39)	23 (13.94)	2 (1.21)	2 (1.21)	7 (4.24)
Poland	29 (2.47)	1122 (95.57)	23 (1.96)	3 (0.89)	263 (78.04)	49 (14.54)	1 (0.30)	20 (5.93)	1 (0.30)
Portugal	5 (1.53)	313 (96.02)	8 (2.45)	5 (3.62)	116 (84.06)	15 (10.87)	0 (0.00)	2 (1.45)	0 (0.00)
Romania	1 (0.44)	224 (99.12)	1 (0.44)	1 (9.09)	9 (81.82)	1 (9.09)	0 (0.00)	0 (0.00)	0 (0.00)
Russian Federation	62 (1.99)	2987 (95.86)	67 (2.15)	5 (0.65)	676 (88.37)	69 (9.02)	4 (0.52)	4 (0.52)	7 (0.92)
Serbia	1 (0.55)	177 (96.72)	5 (2.73)	0 (0.00)	34 (56.67)	26 (43.33)	0 (0.00)	0 (0.00)	0 (0.00)
Slovak Republic	3 (1.08)	262 (93.90)	14 (5.02)	3 (2.01)	126 (84.56)	12 (8.05)	0 (0.00)	8 (5.37)	0 (0.00)
Slovenia	2 (2.08)	92 (95.84)	2 (2.08)	2 (4.88)	27 (65.85)	10 (24.39)	0 (0.00)	2 (4.88)	0 (0.00)
Spain	24 (1.14)	2019 (95.64)	68 (3.22)	25 (2.42)	787 (76.26)	188 (18.22)	13 (1.26)	19 (1.84)	0 (0.00)
Sweden	11 (1.79)	585 (95.28)	18 (2.93)	0 (0.00)	261 (74.15)	65 (18.47)	7 (1.99)	0 (0.00)	19 (5.40)
Switzerland	6 (0.66)	868 (95.70)	33 (3.64)	0 (0.00)	341 (69.88)	122 (25.00)	4 (0.82)	20 (4.10)	1 (0.20)
Turkey	26 (1.31)	1913 (96.52)	43 (2.17)	3 (1.25)	208 (86.67)	26 (10.83)	0 (0.00)	2 (0.83)	1 (0.42)
Ukraine	2 (1.32)	147 (97.36)	2 (1.32)	0 (0.00)	28 (93.33)	1 (3.33)	1 (3.33)	0 (0.00)	0 (0.00)
United Kingdom[2]	0 (0.00)	8957 (92.35)	742 (7.65)	90 (1.65)	3424 (62.72)	1516 (27.77)	124 (2.27)	86 (1.58)	219 (4.01)
Total	607 (1.32)	42974 (93.56)	2352 (5.12)	241 (1.06)	16527 (72.63)	5052 (22.20)	234 (1.03)	219 (0.96)	483 (2.12)

[1] The Netherlands: only diabetes treated with daily insulin is recorded.

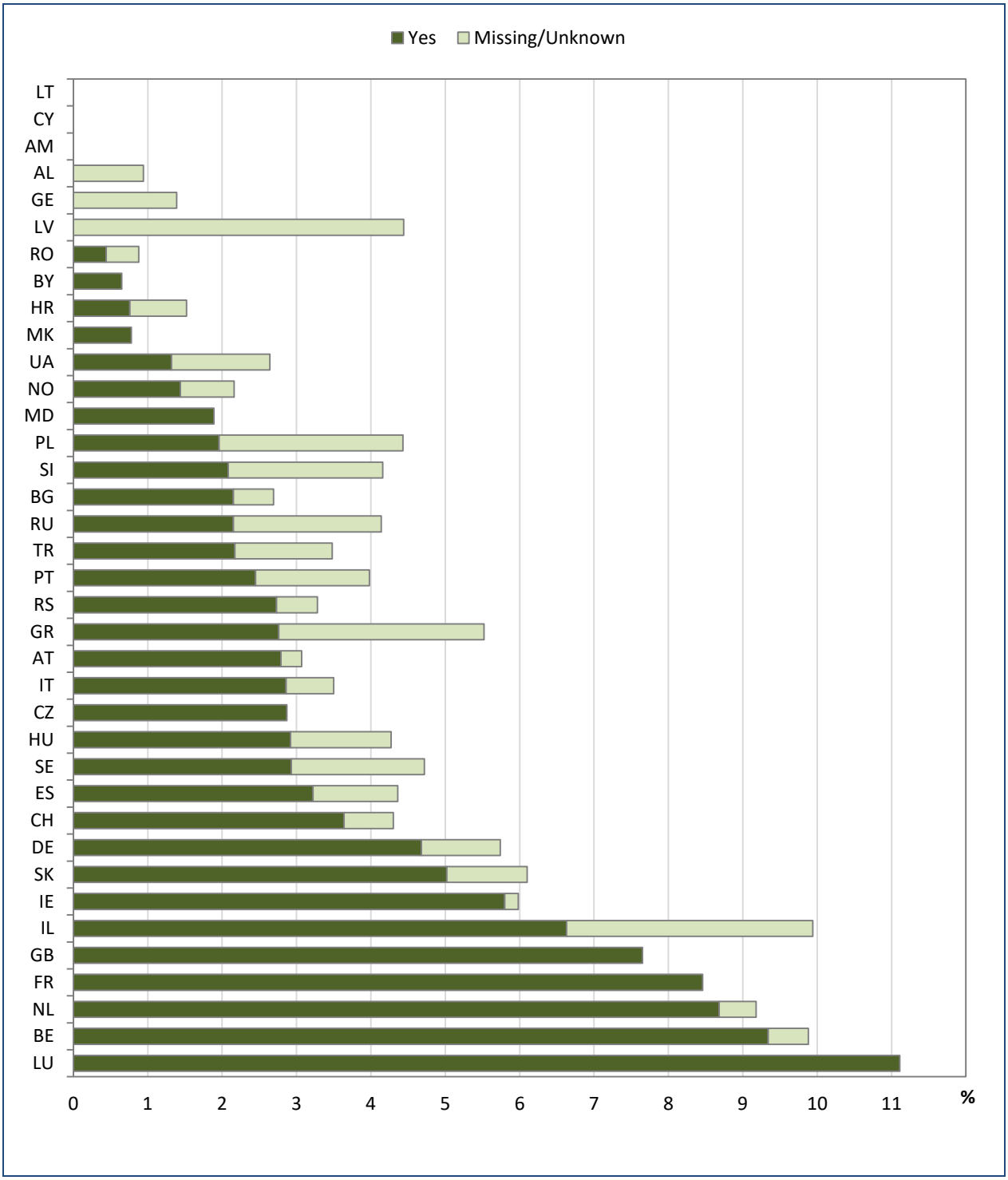
[2] United Kingdom: for ABPA clinician reported aspergillus.

Note: Georgia has 0% coverage for adults and is excluded from the CFRD table.

Note: Armenia has <5 patients aged 18 years or more on 31/12/2019 and are not shown in the CFRD table, but the patients are included in the total.

Table 7.1 shows the frequency of allergic bronchopulmonary aspergillosis (see Appendix 3, page 156, for ABPA definitions) and CF-related diabetes (CFRD) by country. For CFRD only patients 18 years and older are included.

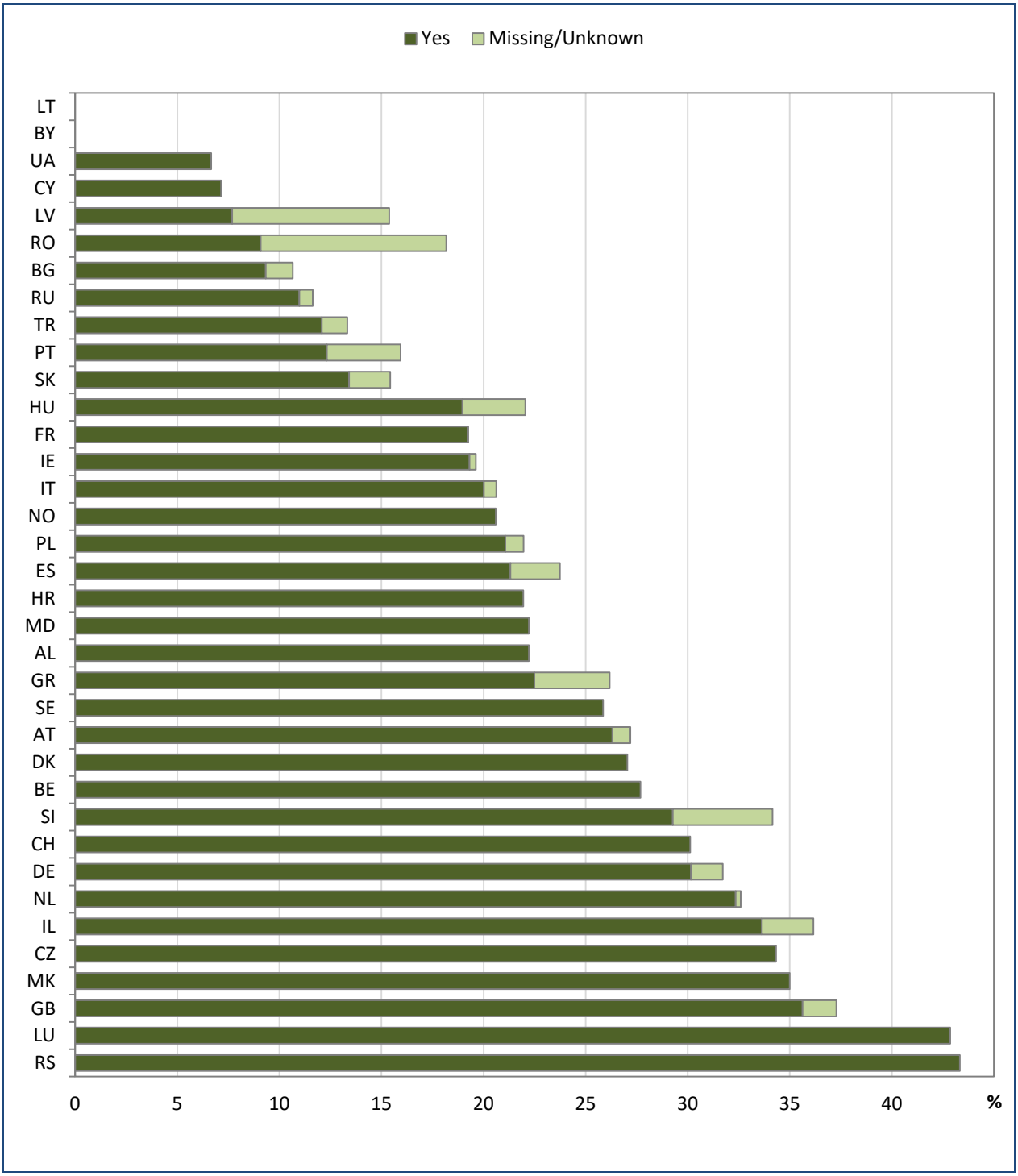
Figure 7.1 Prevalence of allergic bronchopulmonary aspergillosis in all patients seen in 2019 who have never had a transplant, by country.



Note: We excluded from the graph the countries for which the information on allergic bronchopulmonary aspergillosis (ABPA) was missing for more than 10% of the patients.
Note: United Kingdom: for ABPA clinician reported aspergillus.

This graph shows the frequency of allergic bronchopulmonary aspergillosis (ABPA) by country. For the definition of ABPA see Appendix 3 (page 156) the dark green part of the bar shows the percentage of patients with ABPA, the light green part shows the percentage of patients for which this information was missing.

Figure 7.2 Prevalence of CFRD, by country. All patients seen in 2019 aged 18 years or older who have never had a transplant.



Note: In the Netherlands only diabetes treated with daily insulin is recorded.
Note: Georgia has 0% coverage for adults and is excluded from the graph.
Note: Armenia has <5 patients aged 18 years or more on 31/12/2019 and is excluded from the graph.

This graph shows the prevalence of CF-related diabetes (CFRD), by country. The dark green part of the bar shows the percentage of patients who have CFRD, the light green part shows the percentage of patients for whom this information was missing. Only patients aged 18 years or older were included in this graph.

Table 7.2 Prevalence of pneumothorax and haemoptysis major in all patients seen in 2019 who have never had a transplant, by country and overall.

Country	Pneumothorax this year number (%)					Haemoptysis major over 250 ml this year number (%)		
	Missing/ unknown	No	Yes, chest drain	Yes, observation only	Yes, therapy unknown	Missing/ unknown	No	Yes
Albania	1 (0.94)	105 (99.06)	0 (0.00)	0 (0.00)	0 (0.00)	3 (2.83)	102 (96.23)	1 (0.94)
Armenia[1]	0 (0.00)	22 (88.00)	0 (0.00)	3 (12.00)	0 (0.00)	0 (0.00)	20 (80.00)	5 (20.00)
Austria	6 (0.84)	711 (99.02)	0 (0.00)	1 (0.14)	0 (0.00)	6 (0.84)	698 (97.21)	14 (1.95)
Belarus	0 (0)	155 (100)	0 (0)	0 (0)	0 (0)	0 (0.00)	153 (98.71)	2 (1.29)
Belgium	23 (2.06)	1087 (97.58)	0 (0.00)	0 (0.00)	4 (0.36)	124 (11.13)	981 (88.06)	9 (0.81)
Bulgaria	2 (1.08)	184 (98.92)	0 (0.00)	0 (0.00)	0 (0.00)	1 (0.54)	174 (93.55)	11 (5.91)
Croatia	1 (0.76)	129 (98.48)	1 (0.76)	0 (0.00)	0 (0.00)	1 (0.76)	127 (96.95)	3 (2.29)
Cyprus	1 (3.70)	26 (96.30)	0 (0.00)	0 (0.00)	0 (0.00)	0 (0)	27 (100)	0 (0)
Czech Republic	11 (1.85)	580 (97.81)	2 (0.34)	0 (0.00)	0 (0.00)	11 (1.85)	582 (98.15)	0 (0.00)
Denmark	1 (0.21)	467 (99.58)	1 (0.21)	0 (0.00)	0 (0.00)	279 (59.49)	188 (40.08)	2 (0.43)
France	0 (0.00)	5807 (99.83)	10 (0.17)	0 (0.00)	0 (0.00)	0 (0.00)	5764 (99.09)	53 (0.91)
Georgia	1 (1.39)	70 (97.22)	0 (0.00)	0 (0.00)	1 (1.39)	1 (1.39)	69 (95.83)	2 (2.78)
Germany[2]	60 (1.01)	5832 (98.44)	15 (0.25)	16 (0.27)	2 (0.03)	163 (2.75)	5755 (97.13)	7 (0.12)
Greece	12 (3.01)	385 (96.49)	1 (0.25)	0 (0.00)	1 (0.25)	18 (4.51)	377 (94.49)	4 (1.00)
Hungary	6 (1.35)	435 (97.75)	4 (0.90)	0 (0.00)	0 (0.00)	14 (3.15)	419 (94.15)	12 (2.70)
Ireland[3]	2 (0.18)	1136 (99.82)	0 (0.00)	0 (0.00)	0 (0.00)	2 (0.18)	1130 (99.29)	6 (0.53)
Israel	16 (3.12)	497 (96.88)	0 (0.00)	0 (0.00)	0 (0.00)	15 (2.92)	492 (95.91)	6 (1.17)
Italy	77 (1.46)	5192 (98.43)	5 (0.09)	0 (0.00)	1 (0.02)	672 (12.74)	4311 (81.72)	292 (5.54)
Latvia	2 (4.44)	43 (95.56)	0 (0.00)	0 (0.00)	0 (0.00)	2 (4.44)	42 (93.34)	1 (2.22)
Lithuania	0 (0)	27 (100)	0 (0)	0 (0)	0 (0)	0 (0.00)	26 (96.30)	1 (3.70)
Luxembourg	0 (0)	36 (100)	0 (0)	0 (0)	0 (0)	0 (0)	36 (100)	0 (0)
Rep of Moldova	0 (0)	53 (100)	0 (0)	0 (0)	0 (0)	0 (0)	53 (100)	0 (0)

[1] Armenia: considered also patients with haemoptysis <250ml.
[2] Germany: defines haemoptysis major over 240 ml.
[3] Ireland: haemoptysis major is defined as haemoptysis massive over 240ml/day or over 100ml/day for several days.

[table 7.2 continued]

Country	Pneumothorax this year number (%)					Haemoptysis major over 250 ml this year number (%)		
	Missing/ unknown	No	Yes, chest drain	Yes, observation only	Yes, therapy unknown	Missing/ unknown	No	Yes
The Netherlands	5 (0.36)	1385 (99.35)	4 (0.29)	0 (0.00)	0 (0.00)	1 (0.07)	1327 (95.20)	66 (4.73)
North Macedonia	0 (0)	129 (100)	0 (0)	0 (0)	0 (0)	1 (0.78)	125 (96.89)	3 (2.33)
Norway	1 (0.36)	274 (98.92)	1 (0.36)	0 (0.00)	1 (0.36)	3 (1.08)	273 (98.56)	1 (0.36)
Poland	31 (2.64)	1141 (97.19)	2 (0.17)	0 (0.00)	0 (0.00)	32 (2.73)	1097 (93.44)	45 (3.83)
Portugal	7 (2.15)	316 (96.93)	2 (0.61)	1 (0.31)	0 (0.00)	5 (1.53)	299 (91.72)	22 (6.75)
Romania	3 (1.33)	221 (97.79)	1 (0.44)	1 (0.44)	0 (0.00)	2 (0.88)	219 (96.91)	5 (2.21)
Russian Federation	17 (0.55)	3077 (98.74)	22 (0.71)	0 (0.00)	0 (0.00)	42 (1.35)	3046 (97.75)	28 (0.90)
Serbia	1 (0.55)	182 (99.45)	0 (0.00)	0 (0.00)	0 (0.00)	1 (0.55)	173 (94.53)	9 (4.92)
Slovak Republic	6 (2.15)	272 (97.49)	1 (0.36)	0 (0.00)	0 (0.00)	9 (3.23)	250 (89.61)	20 (7.17)
Slovenia	0 (0)	96 (100)	0 (0)	0 (0)	0 (0)	1 (1.04)	93 (96.88)	2 (2.08)
Spain	36 (1.71)	2074 (98.24)	1 (0.05)	0 (0.00)	0 (0.00)	29 (1.37)	2003 (94.89)	79 (3.74)
Sweden	11 (1.79)	603 (98.21)	0 (0.00)	0 (0.00)	0 (0.00)	11 (1.79)	600 (97.72)	3 (0.49)
Switzerland	6 (0.66)	899 (99.12)	2 (0.22)	0 (0.00)	0 (0.00)	5 (0.55)	886 (97.69)	16 (1.76)
Turkey	7 (0.35)	1969 (99.35)	6 (0.30)	0 (0.00)	0 (0.00)	6 (0.30)	1969 (99.35)	7 (0.35)
Ukraine	2 (1.32)	148 (98.02)	1 (0.66)	0 (0.00)	0 (0.00)	2 (1.32)	147 (97.36)	2 (1.32)
United Kingdom[42]	0 (0.00)	9676 (99.76)	23 (0.24)	0 (0.00)	0 (0.00)	0 (0.00)	9663 (99.63)	36 (0.37)
Total	355 (0.77)	45441 (98.93)	105 (0.23)	22 (0.05)	10 (0.02)	1462 (3.18)	43696 (95.13)	775 (1.69)

[4] United Kingdom: defines haemoptysis major over 240 ml.

Table 7.2 shows the frequency of two rare complications: Pneumothorax (collapsed lung) and haemoptysis (coughing up of blood) major of more than 250 ml. Both of these complications are extremely rare.

Table 7.3 Prevalence of liver disease in all patients seen in 2019 who have never had a transplant, by country and overall.

Country	Liver disease this year number (%)						
	Missing/ unknown	No liver disease	Cirrhosis			Liver disease without cirrhosis	Variceal bleeding
			Cirrhosis with portal hypertension/ hypersplenism	Cirrhosis no portal hypertension/ hypersplenism	Cirrhosis. portal hypertension unknown		
Albania	1 (0.94)	52 (49.06)	2 (1.89)	0 (0.00)	0 (0.00)	51 (48.11)	0 (0.00)
Armenia	0 (0.00)	13 (52.00)	0 (0.00)	1 (4.00)	0 (0.00)	11 (44.00)	0 (0.00)
Austria	4 (0.56)	363 (50.55)	13 (1.81)	20 (2.79)	2 (0.28)	316 (44.01)	0 (0.00)
Belarus	0 (0.00)	96 (61.94)	4 (2.58)	2 (1.29)	0 (0.00)	53 (34.19)	0 (0.00)
Belgium[1]	2 (0.18)	1069 (95.96)	43 (3.86)	0 (0.00)	0 (0.00)	0 (0.00)	0 (0.00)
Bulgaria	1 (0.54)	129 (69.35)	4 (2.15)	6 (3.23)	0 (0.00)	46 (24.73)	0 (0.00)
Croatia	1 (0.76)	114 (87.03)	4 (3.05)	3 (2.29)	1 (0.76)	8 (6.11)	0 (0.00)
Cyprus	0 (0.00)	25 (92.60)	1 (3.70)	0 (0.00)	0 (0.00)	1 (3.70)	0 (0.00)
Czech Republic	0 (0.00)	483 (81.45)	10 (1.69)	7 (1.18)	1 (0.17)	92 (15.51)	0 (0.00)
Denmark	2 (0.43)	386 (82.30)	19 (4.05)	9 (1.92)	2 (0.43)	51 (10.87)	0 (0.00)
France	0 (0.00)	4914 (84.48)	110 (1.89)	121 (2.08)	0 (0.00)	672 (11.55)	0 (0.00)
Georgia	2 (2.78)	65 (90.27)	0 (0.00)	3 (4.17)	0 (0.00)	2 (2.78)	0 (0.00)
Germany[2]	360 (6.08)	4065 (68.60)	128 (2.16)	84 (1.42)	83 (1.40)	1205 (20.34)	0 (0.00)
Greece	17 (4.26)	249 (62.41)	16 (4.01)	3 (0.75)	2 (0.50)	112 (28.07)	0 (0.00)
Hungary	7 (1.57)	318 (71.46)	87 (19.55)	14 (3.15)	16 (3.60)	3 (0.67)	0 (0.00)
Ireland	2 (0.18)	1000 (87.86)	29 (2.55)	7 (0.62)	9 (0.79)	91 (8.00)	0 (0.00)
Israel	15 (2.92)	400 (77.98)	9 (1.75)	7 (1.36)	0 (0.00)	81 (15.80)	1 (0.19)
Italy	27 (0.51)	3652 (69.24)	72 (1.36)	37 (0.70)	8 (0.15)	1479 (28.04)	0 (0.00)
Latvia	2 (4.44)	27 (60.01)	2 (4.44)	0 (0.00)	0 (0.00)	14 (31.11)	0 (0.00)
Lithuania	0 (0.00)	26 (96.30)	1 (3.70)	0 (0.00)	0 (0.00)	0 (0.00)	0 (0.00)
Luxembourg	0 (0.00)	28 (77.78)	3 (8.33)	0 (0.00)	0 (0.00)	5 (13.89)	0 (0.00)

[1] Belgium: collects only cirrhosis with portal hypertension “yes” or “no”, therefore no liver disease means no cirrhosis with portal hypertension.

[2] Germany: variceal bleeding information is not reported.

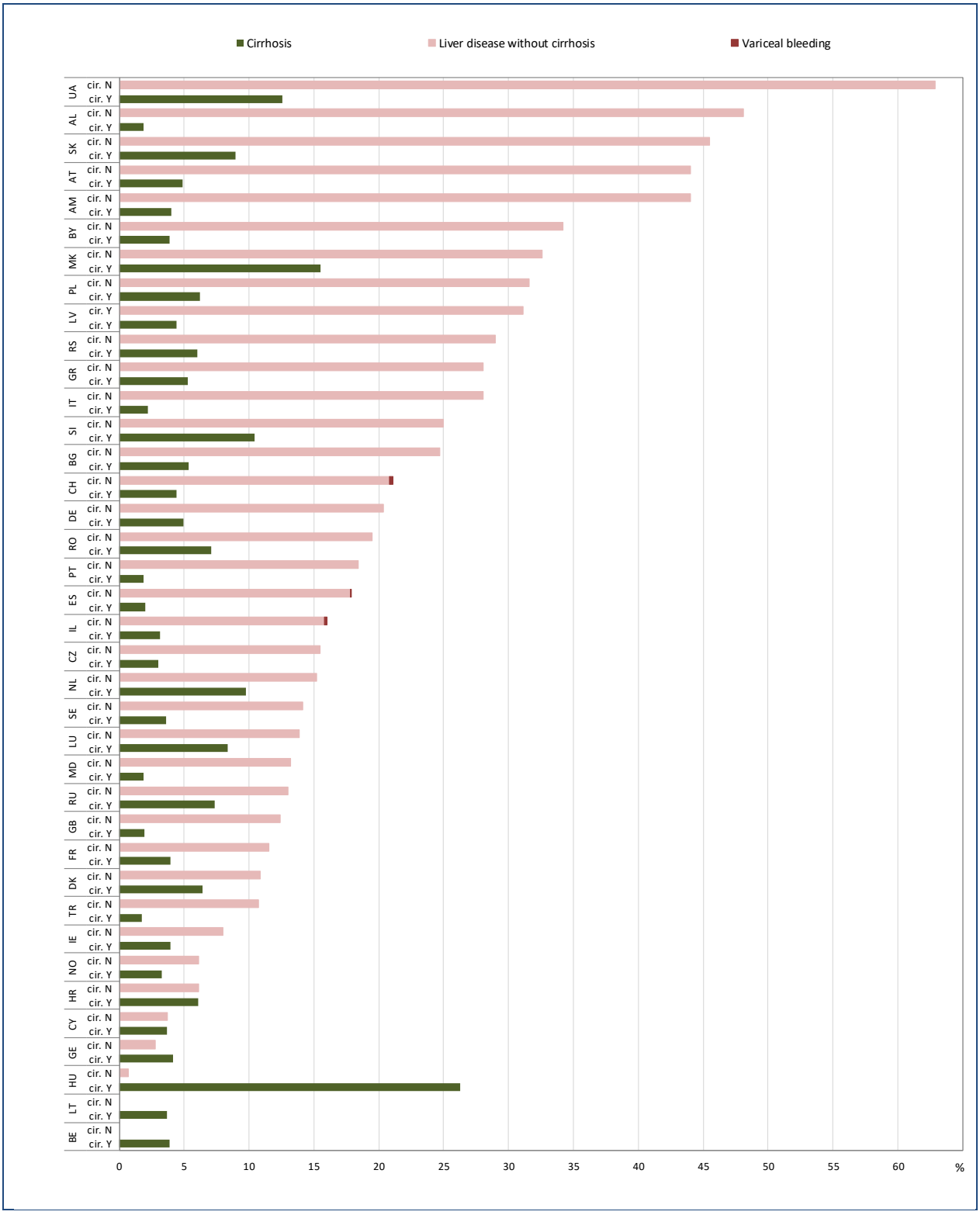
[table 7.3 continued]

Country	Liver disease this year number (%)						
	Missing/ unknown	No liver disease	Cirrhosis			Liver disease without cirrhosis	Variceal bleeding
			Cirrhosis with portal hypertension/ hypersplenism	Cirrhosis no portal hypertension/ hypersplenism	Cirrhosis. portal hypertension unknown		
Rep of Moldova	0 (0.00)	45 (84.90)	0 (0.00)	1 (1.89)	0 (0.00)	7 (13.21)	0 (0.00)
The Netherlands[3]	2 (0.14)	1044 (74.89)	97 (6.96)	35 (2.51)	4 (0.29)	212 (15.21)	0 (0.00)
North Macedonia	0 (0.00)	67 (51.94)	6 (4.65)	14 (10.85)	0 (0.00)	42 (32.56)	0 (0.00)
Norway	5 (1.81)	246 (88.80)	5 (1.81)	0 (0.00)	4 (1.44)	17 (6.14)	0 (0.00)
Poland	29 (2.47)	701 (59.71)	58 (4.94)	10 (0.85)	5 (0.43)	371 (31.60)	0 (0.00)
Portugal	6 (1.84)	254 (77.92)	6 (1.84)	0 (0.00)	0 (0.00)	60 (18.40)	0 (0.00)
Romania	1 (0.44)	165 (73.01)	9 (3.98)	4 (1.77)	3 (1.33)	44 (19.47)	0 (0.00)
Russian Federation	27 (0.87)	2456 (78.81)	149 (4.78)	72 (2.31)	8 (0.26)	404 (12.97)	0 (0.00)
Serbia[4]	2 (1.09)	117 (63.94)	7 (3.83)	2 (1.09)	2 (1.09)	53 (28.96)	0 (0.00)
Slovak Republic	4 (1.43)	123 (44.09)	16 (5.73)	7 (2.51)	2 (0.72)	127 (45.52)	0 (0.00)
Slovenia	1 (1.04)	61 (63.54)	7 (7.29)	3 (3.13)	0 (0.00)	24 (25.00)	0 (0.00)
Spain	31 (1.47)	1661 (78.69)	30 (1.42)	10 (0.47)	2 (0.09)	376 (17.81)	1 (0.05)
Sweden	12 (1.95)	493 (80.30)	12 (1.95)	10 (1.63)	0 (0.00)	87 (14.17)	0 (0.00)
Switzerland	12 (1.32)	664 (73.21)	29 (3.20)	9 (0.99)	2 (0.22)	189 (20.84)	2 (0.22)
Turkey	7 (0.35)	1729 (87.24)	23 (1.16)	7 (0.35)	4 (0.20)	212 (10.70)	0 (0.00)
Ukraine[5]	2 (1.32)	35 (23.19)	11 (7.28)	6 (3.97)	2 (1.32)	95 (62.92)	0 (0.00)
United Kingdom	0 (0.00)	8310 (85.68)	125 (1.29)	63 (0.65)	0 (0.00)	1201 (12.38)	0 (0.00)
Total	584 (1.27)	35645 (77.6)	1147 (2.5)	577 (1.26)	162 (0.35)	7814 (17.01)	4 (0.01)

[3] The Netherlands: variceal bleeding information is not reported.
[4] Serbia: cirrhosis without portal hypertension/hypersplenism means the presence of CF-related liver disease with normal liver function.
[5] Ukraine: Liver disease without cirrhosis also includes ultrasound signs of changes in the liver.

This table shows the frequency and severity of liver disease according to the ECFSPR definitions (see Appendix 3, page 156). The frequency and severity of liver disease differs greatly throughout the ECFSPR data.

Figure 7.3 Prevalence and severity of liver disease in all patients seen in 2019 who have never had a transplant, by country.



Note: Belgium: collects only cirrhosis with portal hypertension “yes” or “no”, therefore no liver disease means no cirrhosis with portal hypertension.
Note: Germany and The Netherlands: variceal bleeding information not reported.
Note: Serbia: cirrhosis without portal hypertension/hypersplenism means the presence of CF-related disease with normal liver function.

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 3, page 156). This graph emphasises better than the table the vast differences in frequency and severity, which may be due to problems in definitions and diagnostic tools.

Table 7.4 Prevalence of the use of ursodeoxycholic acid in all patients seen in 2019 who have never had a transplant, by country and overall.

Country	Ursodeoxycholic acid this year number (%)		
	Missing/ unknown	No	Yes
Albania	2 (1.89)	48 (45.28)	56 (52.83)
Armenia	0 (0.00)	17 (68.00)	8 (32.00)
Austria	4 (0.56)	368 (51.25)	346 (48.19)
Belarus	0 (0.00)	31 (20.00)	124 (80.00)
Belgium	1 (0.09)	894 (80.25)	219 (19.66)
Bulgaria	0 (0.00)	131 (70.43)	55 (29.57)
Croatia	4 (3.05)	92 (70.23)	35 (26.72)
Cyprus	1 (3.70)	24 (88.89)	2 (7.41)
Czech Republic	0 (0.00)	410 (69.14)	183 (30.86)
Denmark	0 (0.00)	335 (71.43)	134 (28.57)
France	0 (0.00)	4462 (76.71)	1355 (23.29)
Georgia	1 (1.39)	53 (73.61)	18 (25.00)
Germany	43 (0.73)	2977 (50.24)	2905 (49.03)
Greece	12 (3.01)	262 (65.66)	125 (31.33)
Hungary	7 (1.57)	246 (55.28)	192 (43.15)
Ireland	2 (0.18)	1049 (92.18)	87 (7.64)
Israel	6 (1.17)	427 (83.24)	80 (15.59)
Italy	15 (0.28)	3466 (65.71)	1794 (34.01)
Latvia	0 (0.00)	29 (64.44)	16 (35.56)
Lithuania	0 (0.00)	20 (74.07)	7 (25.93)
Luxembourg	0 (0.00)	21 (58.33)	15 (41.67)
Rep of Moldova	0 (0.00)	40 (75.47)	13 (24.53)

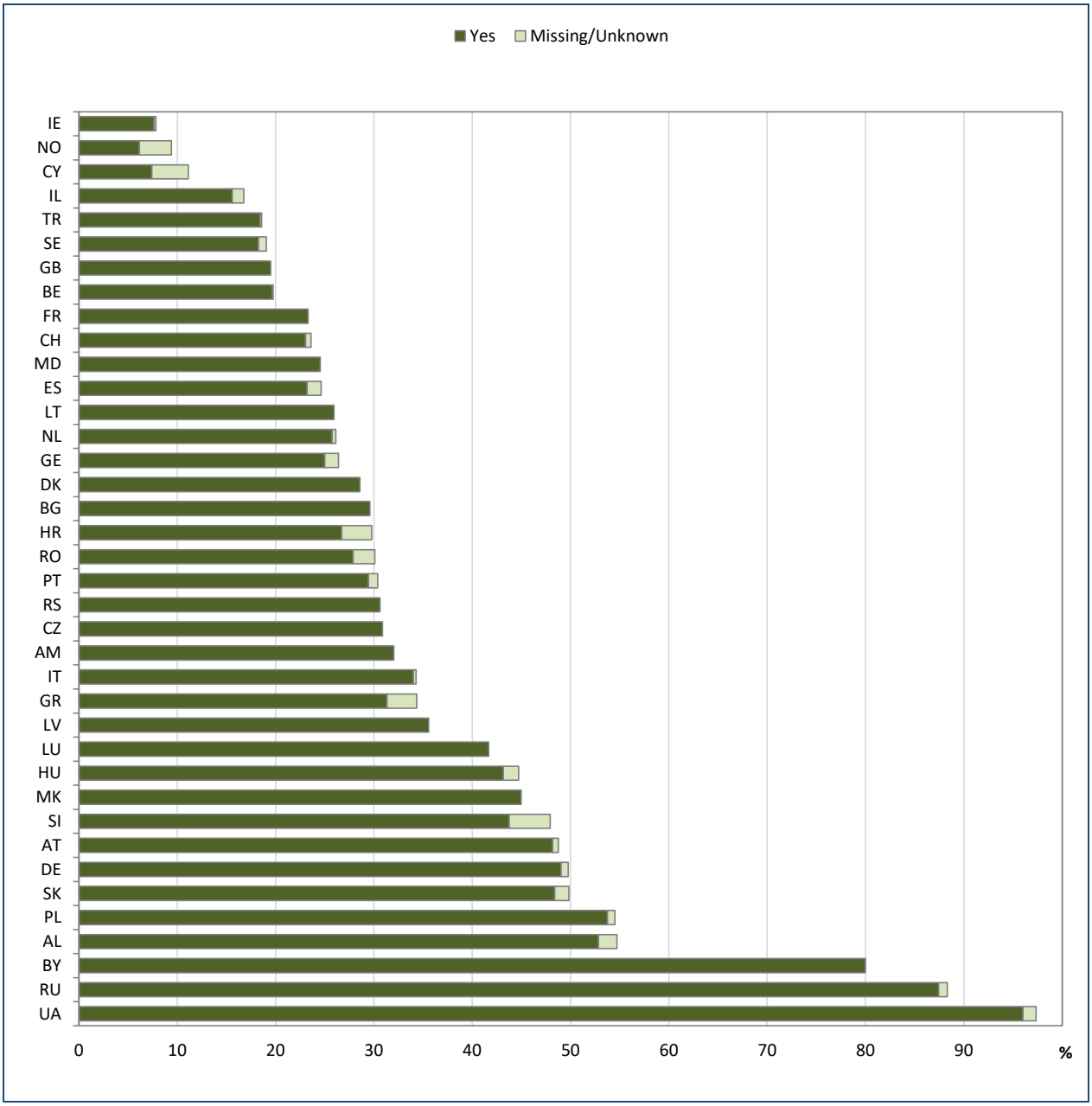
[table 7.4 continued]

Country	Ursodeoxycholic acid this year number (%)		
	Missing/ unknown	No	Yes
The Netherlands	5 (0.36)	1030 (73.89)	359 (25.75)
North Macedonia	0 (0.00)	71 (55.04)	58 (44.96)
Norway	9 (3.25)	251 (90.61)	17 (6.14)
Poland	9 (0.77)	534 (45.49)	631 (53.75)
Portugal	3 (0.92)	227 (69.63)	96 (29.45)
Romania	5 (2.21)	158 (69.91)	63 (27.88)
Russian Federation	27 (0.87)	364 (11.68)	2725 (87.45)
Serbia	0 (0.00)	127 (69.40)	56 (30.60)
Slovak Republic	4 (1.43)	140 (50.18)	135 (48.39)
Slovenia	4 (4.17)	50 (52.08)	42 (43.75)
Spain	30 (1.42)	1591 (75.37)	490 (23.21)
Sweden	5 (0.81)	497 (80.95)	112 (18.24)
Switzerland	5 (0.55)	693 (76.41)	209 (23.04)
Turkey	3 (0.15)	1614 (81.43)	365 (18.42)
Ukraine	2 (1.32)	4 (2.65)	145 (96.03)
United Kingdom	0 (0.00)	7810 (80.52)	1889 (19.48)
Total	209 (0.46)	30563 (66.54)	15161 (33.01)

Note: Oral ursodesoxycholic acid is reimbursed in most countries in Europe, except in Armenia, Bulgaria, Lithuania, Serbia and Ukraine. In Republic of Moldova it is reimbursed for children.

This table shows the frequency of the use of ursodeoxycholic acid, a commonly used treatment for CF liver disease. The frequency and severity of liver disease differs greatly throughout the ECFSPR data and does not correspond to the number of patients on ursodeoxycholic acid.

Figure 7.4 Use of ursodeoxycholic acid in all patients seen in 2019 who have never had a transplant, by country.



Note: Oral ursodesoxycholic acid is reimbursed in most countries in Europe, except in Armenia, Bulgaria, Lithuania, Serbia and Ukraine. In Republic of Moldova it is only reimbursed for children.

This graph shows how many patients used ursodeoxycholic acid during 2019. Ursodeoxycholic acid is used as a treatment for CF liver disease. The dark green part of the bar indicates the percentage of patients taking this drug, the light green part shows the percentage of patients for whom this information is missing.

Table 7.5 Occurrence of malignancy this year and prevalence of distal intestinal obstruction syndrome (DIOS) in all patients seen in 2019 who have never had a transplant, by country and overall.

Country	Malignancy occurred this year number(%)			DIOS this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	3 (2.83)	103 (97.17)	0 (0.00)	3 (2.83)	100 (94.34)	3 (2.83)
Armenia	0 (0)	25 (100)	0 (0)	0 (0.00)	23 (92.00)	2 (8.00)
Austria	5 (0.70)	713 (99.30)	0 (0.00)	1 (0.14)	693 (96.52)	24 (3.34)
Belarus	0 (0)	155 (100)	0 (0)	0 (0.00)	152 (98.06)	3 (1.94)
Belgium	0 (0.00)	1111 (99.73)	3 (0.27)	12 (1.08)	1062 (95.33)	40 (3.59)
Bulgaria	2 (1.08)	183 (98.38)	1 (0.54)	1 (0.54)	185 (99.46)	0 (0.00)
Croatia	1 (0.76)	130 (99.24)	0 (0.00)	1 (0.76)	126 (96.19)	4 (3.05)
Cyprus	0 (0)	27 (100)	0 (0)	0 (0)	27 (100)	0 (0)
Czech Republic	7 (1.18)	585 (98.65)	1 (0.17)	2 (0.34)	588 (99.15)	3 (0.51)
Denmark[1]	2 (0.43)	464 (98.93)	3 (0.64)	280 (59.70)	189 (40.30)	0 (0.00)
France	5 (0.09)	5795 (99.62)	17 (0.29)	0 (0.00)	5623 (96.66)	194 (3.34)
Georgia	1 (1.39)	71 (98.61)	0 (0.00)	2 (2.78)	68 (94.44)	2 (2.78)
Germany	77 (1.30)	5805 (97.97)	43 (0.73)	86 (1.45)	5605 (94.60)	234 (3.95)
Greece	15 (3.76)	377 (94.49)	7 (1.75)	10 (2.51)	382 (95.74)	7 (1.75)
Hungary	7 (1.57)	435 (97.76)	3 (0.67)	445 (100)	0 (0)	0 (0)
Ireland	2 (0.18)	1135 (99.73)	1 (0.09)	2 (0.18)	1099 (96.57)	37 (3.25)
Israel	15 (2.92)	494 (96.30)	4 (0.78)	15 (2.92)	487 (94.94)	11 (2.14)
Italy	63 (1.19)	5195 (98.49)	17 (0.32)	685 (12.99)	4509 (85.47)	81 (1.54)
Latvia	2 (4.44)	43 (95.56)	0 (0.00)	2 (4.44)	43 (95.56)	0 (0.00)
Lithuania	0 (0)	27 (100)	0 (0)	0 (0)	27 (100)	0 (0)
Luxembourg	0 (0.00)	35 (97.22)	1 (2.78)	2 (5.56)	34 (94.44)	0 (0.00)
Rep of Moldova	0 (0)	53 (100)	0 (0)	0 (0)	53 (100)	0 (0)

[1] Denmark has only reported DIOS requiring hospitalisation.

[table 7.5 continued]

Country	Malignancy occurred this year number(%)			DIOS this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
The Netherlands	0 (0.00)	1392 (99.86)	2 (0.14)	3 (0.22)	1320 (94.69)	71 (5.09)
North Macedonia	0 (0)	129 (100)	0 (0)	0 (0.00)	125 (96.90)	4 (3.10)
Norway	13 (4.69)	264 (95.31)	0 (0.00)	17 (6.14)	252 (90.97)	8 (2.89)
Poland	33 (2.81)	1141 (97.19)	0 (0.00)	24 (2.04)	1140 (97.11)	10 (0.85)
Portugal	4 (1.23)	321 (98.46)	1 (0.31)	3 (0.92)	319 (97.85)	4 (1.23)
Romania	2 (0.88)	223 (98.68)	1 (0.44)	2 (0.88)	220 (97.35)	4 (1.77)
Russian Federation	30 (0.96)	3081 (98.88)	5 (0.16)	68 (2.18)	2994 (96.09)	54 (1.73)
Serbia	1 (0.55)	182 (99.45)	0 (0.00)	1 (0.55)	180 (98.36)	2 (1.09)
Slovak Republic	7 (2.51)	272 (97.49)	0 (0.00)	4 (1.43)	270 (96.78)	5 (1.79)
Slovenia	1 (1.04)	95 (98.96)	0 (0.00)	0 (0.00)	94 (97.92)	2 (2.08)
Spain	35 (1.66)	2071 (98.10)	5 (0.24)	30 (1.42)	2048 (97.02)	33 (1.56)
Sweden	11 (1.79)	603 (98.21)	0 (0.00)	11 (1.79)	583 (94.95)	20 (3.26)
Switzerland	5 (0.55)	899 (99.12)	3 (0.33)	5 (0.55)	879 (96.91)	23 (2.54)
Turkey	6 (0.30)	1974 (99.60)	2 (0.10)	5 (0.25)	1963 (99.04)	14 (0.71)
Ukraine	2 (1.32)	149 (98.68)	0 (0.00)	3 (1.99)	147 (97.35)	1 (0.66)
United Kingdom	11 (0.11)	9665 (99.65)	23 (0.24)	0 (0.00)	9171 (94.56)	528 (5.44)
Total	368 (0.8)	45422 (98.89)	143 (0.31)	1725 (3.76)	42780 (93.14)	1428 (3.11)

Table 7.5 shows the frequency of two rare complications: occurrence of malignancy (cancer) and distal intestinal obstruction syndrome (DIOS). Both these complications are rare.

Table 7.6 Use of hypertonic saline, rhDNase and inhaled mannitol in all patients seen in 2019 who have never had a transplant, by country and overall.

Country	Hypertonic saline (NaCl) inhaled > 3 months this year number (%)			rhDNase inhaled > 3 months this year number (%)			Mannitol inhaled > 3 months this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	8 (7.55)	0 (0.00)	98 (92.45)	3 (2.83)	96 (90.57)	7 (6.60)	2 (1.89)	104 (98.11)	0 (0.00)
Armenia	0 (0)	0 (0)	25 (100)	0 (0.00)	19 (76.00)	6 (24.00)	0 (0)	25 (100)	0 (0)
Austria	1 (0.14)	142 (19.78)	575 (80.08)	1 (0.14)	327 (45.54)	390 (54.32)	1 (0.14)	708 (98.61)	9 (1.25)
Belarus	0 (0.00)	54 (34.84)	101 (65.16)	0 (0.00)	150 (96.77)	5 (3.23)	0 (0)	155 (100)	0 (0)
Belgium	37 (3.32)	333 (29.89)	744 (66.79)	37 (3.32)	130 (11.67)	947 (85.01)	1114 (100)	0 (0)	0 (0)
Bulgaria	0 (0.00)	96 (51.61)	90 (48.39)	0 (0.00)	28 (15.05)	158 (84.95)	0 (0)	186 (100)	0 (0)
Croatia	2 (1.53)	16 (12.21)	113 (86.26)	5 (3.82)	37 (28.24)	89 (67.94)	5 (3.82)	125 (95.42)	1 (0.76)
Cyprus	1 (3.70)	22 (81.49)	4 (14.81)	1 (3.70)	7 (25.93)	19 (70.37)	1 (3.70)	26 (96.30)	0 (0.00)
Czech Republic	0 (0.00)	119 (20.07)	474 (79.93)	0 (0.00)	196 (33.05)	397 (66.95)	0 (0.00)	586 (98.82)	7 (1.18)
Denmark	469 (100)	0 (0)	0 (0)	0 (0.00)	40 (8.53)	429 (91.47)	469 (100)	0 (0)	0 (0)
France	0 (0.00)	5011 (86.14)	806 (13.86)	0 (0.00)	3116 (53.57)	2701 (46.43)	5817 (100)	0 (0)	0 (0)
Georgia	0 (0.00)	48 (66.67)	24 (33.33)	0 (0)	72 (100)	0 (0)	0 (0)	72 (100)	0 (0)
Germany	46 (0.78)	1180 (19.92)	4699 (79.30)	47 (0.79)	2749 (46.40)	3129 (52.81)	58 (0.98)	5636 (95.12)	231 (3.90)
Greece	6 (1.50)	291 (72.94)	102 (25.56)	4 (1.00)	117 (29.33)	278 (69.67)	8 (2.01)	381 (95.48)	10 (2.51)
Hungary	9 (2.02)	80 (17.98)	356 (80.00)	9 (2.02)	133 (29.89)	303 (68.09)	445 (100)	0 (0)	0 (0)
Ireland	2 (0.18)	466 (40.95)	670 (58.87)	2 (0.18)	496 (43.58)	640 (56.24)	1138 (100)	0 (0)	0 (0)
Israel	6 (1.17)	118 (23.00)	389 (75.83)	3 (0.58)	123 (23.98)	387 (75.44)	2 (0.39)	510 (99.42)	1 (0.19)
Italy	15 (0.28)	2886 (54.72)	2374 (45.00)	15 (0.28)	3020 (57.26)	2240 (42.46)	1493 (28.30)	3707 (70.28)	75 (1.42)
Latvia	0 (0.00)	4 (8.89)	41 (91.11)	0 (0.00)	25 (55.56)	20 (44.44)	0 (0)	45 (100)	0 (0)
Lithuania	0 (0.00)	22 (81.48)	5 (18.52)	0 (0.00)	4 (14.81)	23 (85.19)	0 (0)	27 (100)	0 (0)
Luxembourg	0 (0.00)	3 (8.33)	33 (91.67)	0 (0.00)	10 (27.78)	26 (72.22)	0 (0.00)	35 (97.22)	1 (2.78)
Rep of Moldova	0 (0.00)	3 (5.66)	50 (94.34)	0 (0.00)	52 (98.11)	1 (1.89)	0 (0)	53 (100)	0 (0)

[table 7.6 continued]

Country	Hypertonic saline (NaCl) inhaled > 3 months this year number (%)			rhDNase inhaled > 3 months this year number (%)			Mannitol inhaled > 3 months this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
The Netherlands	2 (0.14)	902 (64.71)	490 (35.15)	3 (0.22)	482 (34.57)	909 (65.21)	1394 (100)	0 (0)	0 (0)
North Macedonia	0 (0.00)	40 (31.01)	89 (68.99)	0 (0.00)	41 (31.78)	88 (68.22)	3 (2.33)	126 (97.67)	0 (0.00)
Norway	3 (1.08)	57 (20.58)	217 (78.34)	0 (0.00)	94 (33.94)	183 (66.06)	2 (0.72)	275 (99.28)	0 (0.00)
Poland	5 (0.43)	143 (12.18)	1026 (87.39)	4 (0.34)	170 (14.48)	1000 (85.18)	8 (0.68)	1164 (99.15)	2 (0.17)
Portugal	2 (0.61)	197 (60.43)	127 (38.96)	3 (0.92)	50 (15.34)	273 (83.74)	3 (0.92)	322 (98.77)	1 (0.31)
Romania	1 (0.44)	59 (26.11)	166 (73.45)	1 (0.44)	45 (19.91)	180 (79.65)	1 (0.44)	225 (99.56)	0 (0.00)
Russian Federation	56 (1.80)	856 (27.47)	2204 (70.73)	22 (0.71)	117 (3.75)	2977 (95.54)	27 (0.87)	2991 (95.98)	98 (3.15)
Serbia	0 (0.00)	7 (3.83)	176 (96.17)	0 (0.00)	49 (26.78)	134 (73.22)	0 (0)	183 (100)	0 (0)
Slovak Republic	0 (0.00)	151 (54.12)	128 (45.88)	3 (1.08)	87 (31.18)	189 (67.74)	4 (1.43)	275 (98.57)	0 (0.00)
Slovenia	0 (0.00)	4 (4.17)	92 (95.83)	1 (1.04)	65 (67.71)	30 (31.25)	3 (3.13)	92 (95.83)	1 (1.04)
Spain	8 (0.38)	728 (34.48)	1375 (65.14)	8 (0.38)	1293 (61.25)	810 (38.37)	10 (0.47)	2093 (99.15)	8 (0.38)
Sweden	6 (0.98)	103 (16.77)	505 (82.25)	7 (1.14)	419 (68.24)	188 (30.62)	0 (0.00)	608 (99.02)	6 (0.98)
Switzerland	5 (0.55)	200 (22.05)	702 (77.40)	3 (0.33)	466 (51.38)	438 (48.29)	6 (0.66)	900 (99.23)	1 (0.11)
Turkey	4 (0.20)	1630 (82.24)	348 (17.56)	4 (0.20)	172 (8.68)	1806 (91.12)	4 (0.20)	1882 (94.96)	96 (4.84)
Ukraine	3 (1.99)	1 (0.66)	147 (97.35)	3 (1.99)	19 (12.58)	129 (85.43)	2 (1.32)	149 (98.68)	0 (0.00)
United Kingdom[1]	0 (0.00)	6309 (65.05)	3390 (34.95)	0 (0.00)	3002 (30.95)	6697 (69.05)	0 (0.00)	9359 (96.49)	340 (3.51)
Total	697 (1.52)	22281 (48.51)	22955 (49.97)	189 (0.41)	17518 (38.14)	28226 (61.45)			

[1] United Kingdom: the duration of use of inhaled hypertonic saline is not specified.

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

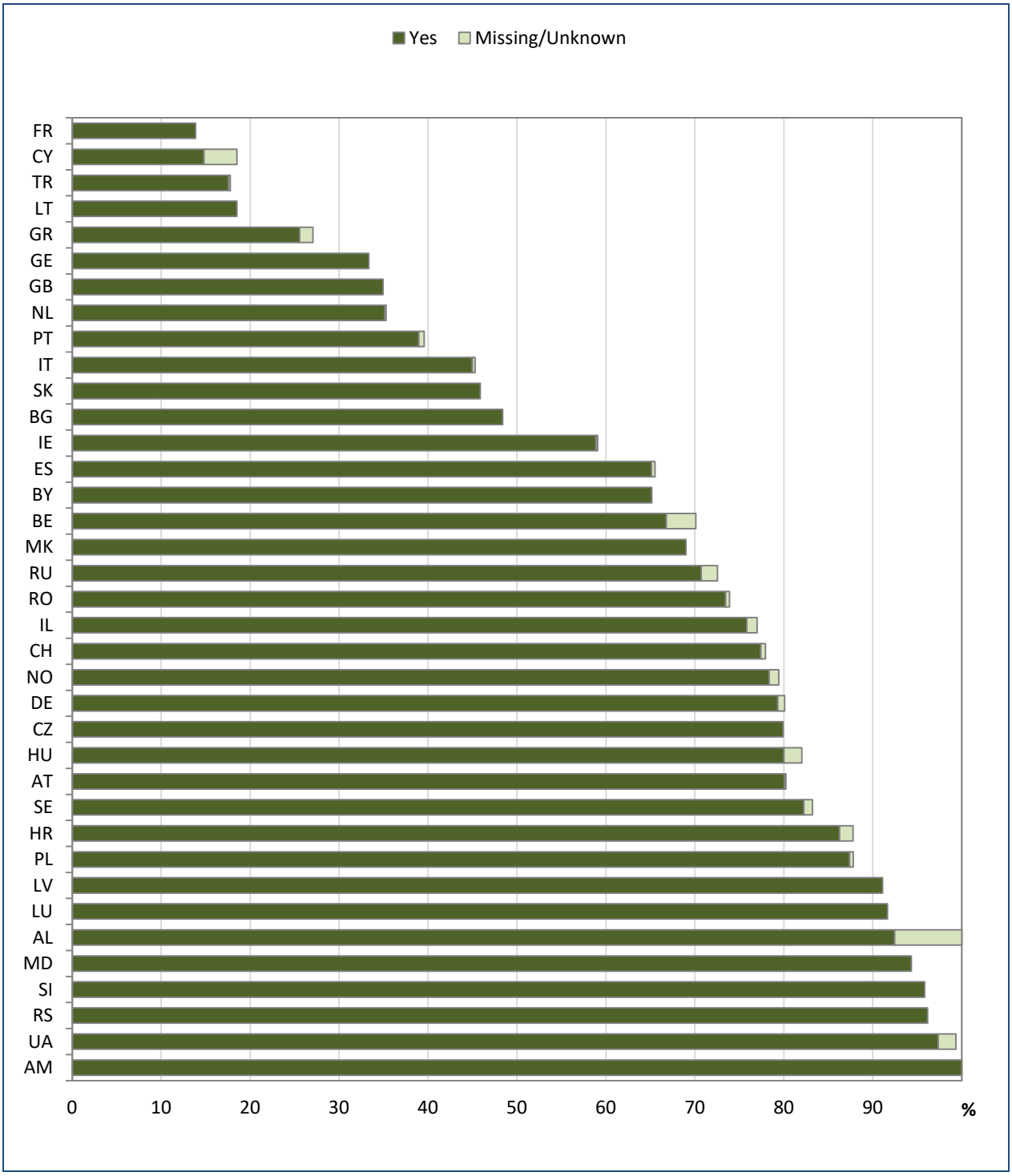
Note: Inhaled hypertonic saline is reimbursed in most countries except in Albania, Armenia, Bulgaria, Lithuania, Republic of Moldova, Poland, Romania, Russian Federation, Serbia, Sweden. In Ukraine it is y reimbursed for children.

Note: Inhaled rhDNase is reimbursed in most countries except in Albania, Armenia and Republic of Moldova. In Bulgaria, Croatia, Georgia, Germany, Israel, Luxembourg, North Macedonia, Norway, Romania, Spain, Ukraine, United Kingdom it is reimbursed for patients ≥ 5 years; in Latvia it is reimbursed for patients ≥ 6 years. In Belgium it is reimbursed for patients with FVC% >=40 only.

Note: Inhaled Mannitol is reimbursed in Austria, Czech Republic, Denmark, France, Germany (≥ 18 years), Greece (≥ 18 years), Italy (≥ 18 years), Russian Federation (depending on the region of residence), Slovenia, Turkey (≥ 6 years), United Kingdom (≥ 18 years), but not in the other countries.

Table 7.6 shows the use of three different inhaled medications: hypertonic saline, rhDNase (Pulmozyme®) and mannitol (see page 8 for abbreviations). Hypertonic saline can be any saline of a concentration >0.9% NaCl, but most commonly between 3% and 11%.

Figure 7.5 Use of inhaled hypertonic saline in all patients seen in 2019 who have never had a transplant, by country.



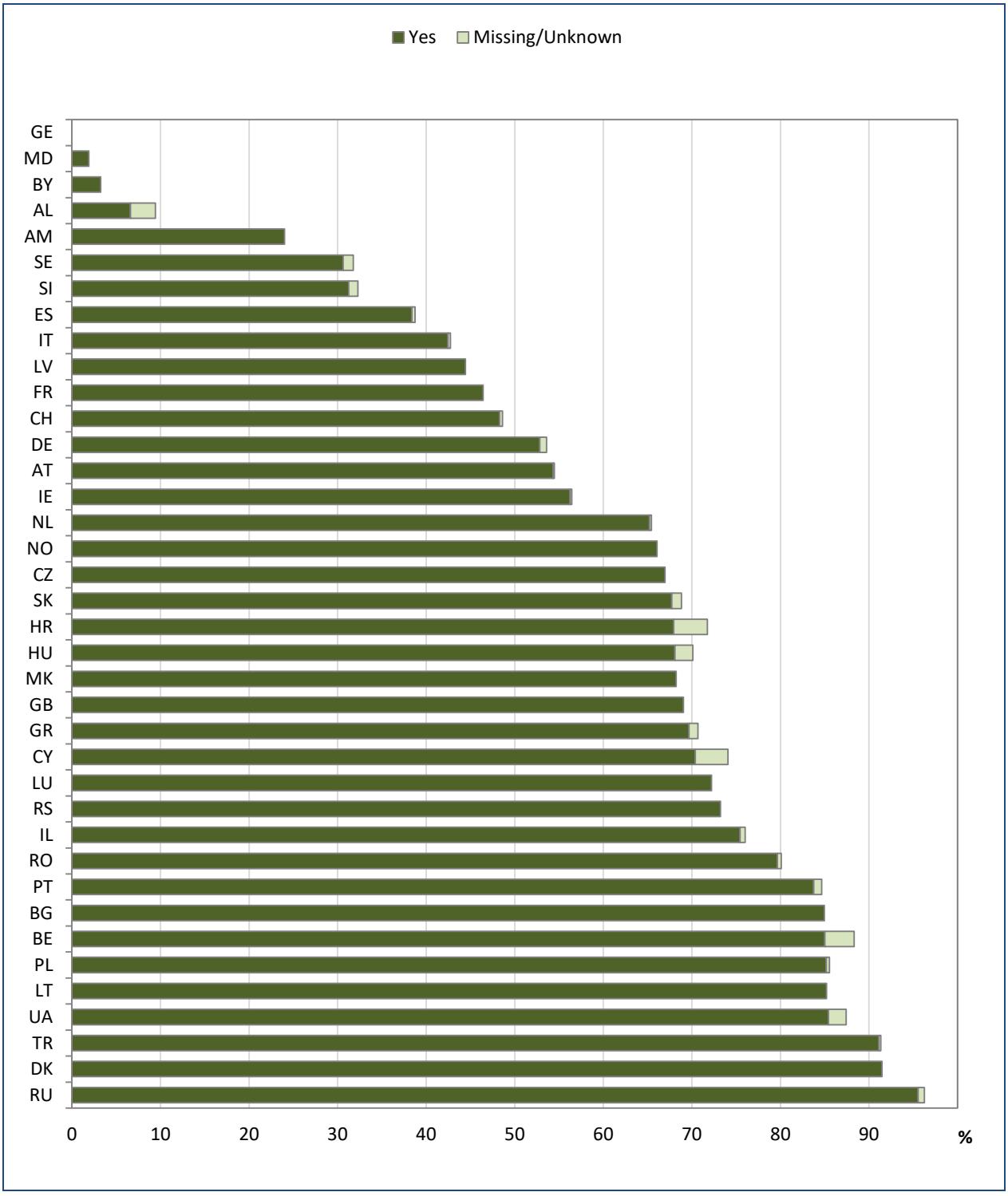
Note: We excluded from the graph the countries for which the information on inhaled hypertonic saline was missing for more than 10% of the patients.

Note: United Kingdom: the duration of use of inhaled hypertonic saline is not specified.

Note: Inhaled hypertonic saline is reimbursed in most countries except in Albania, Armenia, Bulgaria, Lithuania, Republic of Moldova, Poland, Romania, Russian Federation, Serbia, Sweden. In Ukraine it is only reimbursed for children.

This graph shows the use of inhaled hypertonic saline for more than three months during the survey year. The dark green part of the bar indicates the percentage of patients taking the medication, the light green part shows the percentage of patients for whom this information is missing.

Figure 7.6 Use of rhDNase in all patients seen in 2019 who have never had a transplant, by country.



Note: Inhaled rhDNase is reimbursed in most countries except in Albania, Armenia and Republic of Moldova. In Bulgaria, Croatia, Georgia, Germany, Israel, Luxembourg, Macedonia, Republic of Moldova, Norway, Romania, Slovenia, Spain, Ukraine, United Kingdom it is reimbursed for patients > 5 years; in Latvia it is reimbursed for patients > 6 years.

This graph shows the use of rhDNase as inhalations for more than 3 months during the survey year. The dark green part of the bar indicates the percentage of patients taking this drug, the light green part shows the percentage of patients for whom this information is missing.

Table 7.7 Use of inhaled antibiotics, bronchodilators and macrolides in all patients seen in 2019 who have never had a transplant, by country and overall.

Country	Antibiotics inhaled > 3 months this year number (%)			Bronchodilators inhaled > 3 months this year number (%)			Macrolides > 3 months this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	2 (1.89)	73 (68.86)	31 (29.25)	1 (0.94)	16 (15.09)	89 (83.97)	3 (2.83)	85 (80.19)	18 (16.98)
Armenia	0 (0.00)	15 (60.00)	10 (40.00)	0 (0.00)	1 (4.00)	24 (96.00)	0 (0.00)	14 (56.00)	11 (44.00)
Austria	1 (0.14)	444 (61.84)	273 (38.02)	0 (0.00)	24 (3.34)	694 (96.66)	1 (0.14)	679 (94.57)	38 (5.29)
Belarus	0 (0.00)	102 (65.81)	53 (34.19)	0 (0.00)	100 (64.52)	55 (35.48)	0 (0.00)	112 (72.26)	43 (27.74)
Belgium	2 (0.18)	524 (47.04)	588 (52.78)	37 (3.32)	210 (18.85)	867 (77.83)	1 (0.09)	515 (46.23)	598 (53.68)
Bulgaria	0 (0.00)	97 (52.15)	89 (47.85)	0 (0.00)	138 (74.19)	48 (25.81)	0 (0.00)	175 (94.09)	11 (5.91)
Croatia	6 (4.58)	62 (47.33)	63 (48.09)	5 (3.82)	96 (73.28)	30 (22.90)	5 (3.82)	81 (61.83)	45 (34.35)
Cyprus	1 (3.70)	17 (62.97)	9 (33.33)	1 (3.70)	15 (55.56)	11 (40.74)	1 (3.70)	14 (51.86)	12 (44.44)
Czech Republic	0 (0.00)	458 (77.23)	135 (22.77)	0 (0.00)	247 (41.65)	346 (58.35)	0 (0.00)	560 (94.44)	33 (5.56)
Denmark	2 (0.43)	423 (90.19)	44 (9.38)	469 (100)	0 (0)	0 (0)	1 (0.21)	343 (73.14)	125 (26.65)
France	0 (0.00)	3375 (58.02)	2442 (41.98)	0 (0.00)	1970 (33.87)	3847 (66.13)	0 (0.00)	3825 (65.76)	1992 (34.24)
Georgia	0 (0.00)	71 (98.61)	1 (1.39)	0 (0)	72 (100)	0 (0)	0 (0.00)	58 (80.56)	14 (19.44)
Germany	86 (1.45)	3364 (56.78)	2475 (41.77)	48 (0.81)	1113 (18.78)	4764 (80.41)	62 (1.05)	4959 (83.69)	904 (15.26)
Greece	5 (1.25)	173 (43.36)	221 (55.39)	3 (0.75)	151 (37.84)	245 (61.41)	7 (1.75)	274 (68.68)	118 (29.57)
Hungary	7 (1.57)	209 (46.97)	229 (51.46)	10 (2.25)	169 (37.98)	266 (59.77)	14 (3.15)	299 (67.19)	132 (29.66)
Ireland	3 (0.26)	653 (57.38)	482 (42.36)	2 (0.18)	300 (26.36)	836 (73.46)	2 (0.18)	600 (52.72)	536 (47.10)
Israel	3 (0.58)	237 (46.20)	273 (53.22)	7 (1.36)	199 (38.79)	307 (59.85)	5 (0.97)	282 (54.98)	226 (44.05)
Italy	14 (0.27)	3301 (62.57)	1960 (37.16)	14 (0.27)	1356 (25.71)	3905 (74.02)	18 (0.34)	3807 (72.17)	1450 (27.49)
Latvia	0 (0.00)	33 (73.33)	12 (26.67)	0 (0.00)	2 (4.44)	43 (95.56)	0 (0.00)	41 (91.11)	4 (8.89)
Lithuania	1 (3.70)	20 (74.07)	6 (22.22)	1 (3.70)	14 (51.86)	12 (44.44)	0 (0.00)	26 (96.30)	1 (3.70)
Luxembourg	0 (0.00)	19 (52.78)	17 (47.22)	0 (0.00)	5 (13.89)	31 (86.11)	1 (2.78)	26 (72.22)	9 (25.00)
Rep of Moldova	0 (0.00)	20 (37.74)	33 (62.26)	0 (0.00)	44 (83.02)	9 (16.98)	0 (0.00)	46 (86.79)	7 (13.21)

[table 7.7 continued]

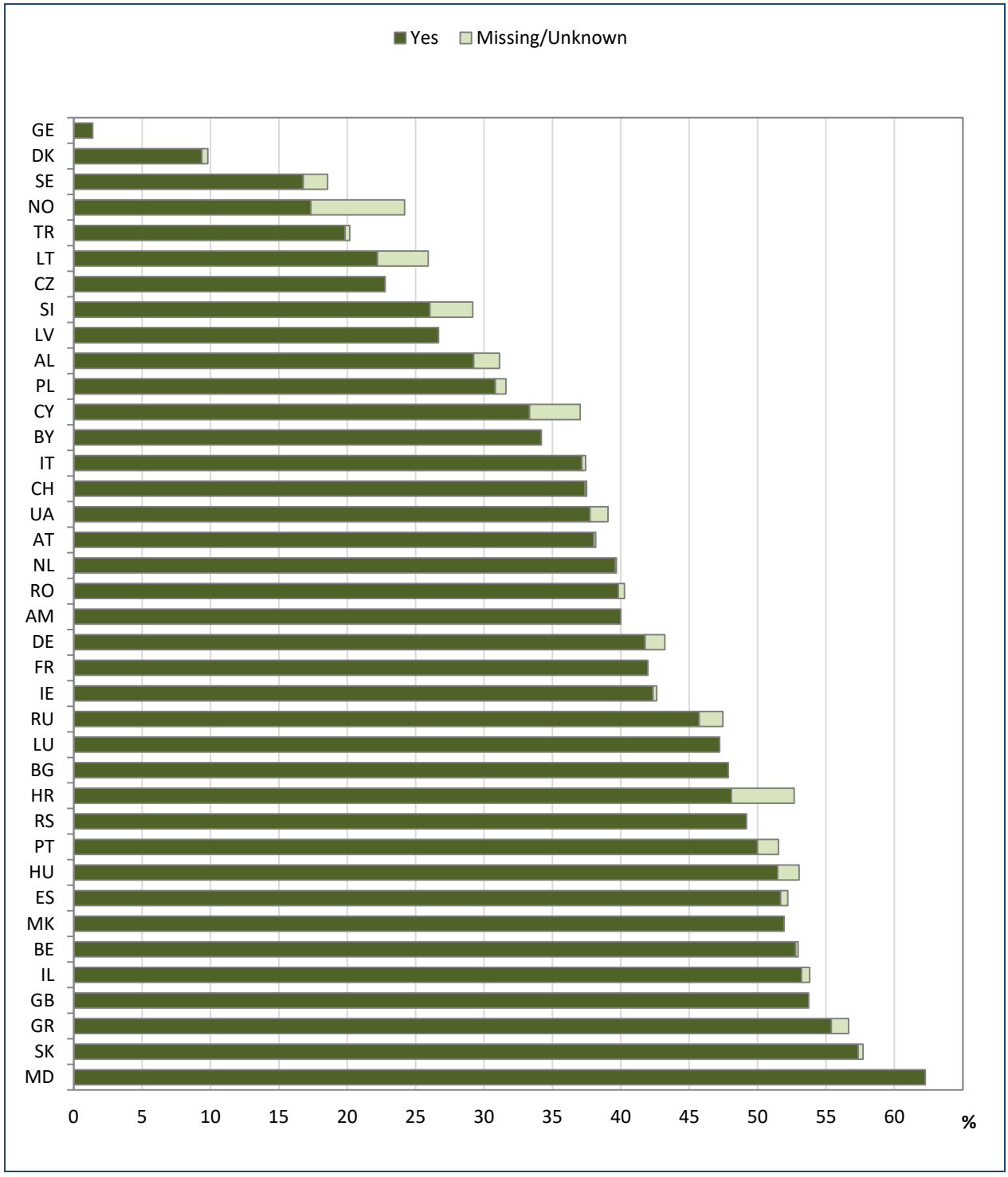
Country	Antibiotics inhaled > 3 months this year number (%)			Bronchodilators inhaled > 3 months this year number (%)			Macrolides > 3 months this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
The Netherlands	1 (0.07)	841 (60.33)	552 (39.60)	0 (0.00)	598 (42.90)	796 (57.10)	1 (0.07)	885 (63.49)	508 (36.44)
North Macedonia	0 (0.00)	62 (48.06)	67 (51.94)	0 (0.00)	9 (6.98)	120 (93.02)	0 (0.00)	102 (79.07)	27 (20.93)
Norway	19 (6.86)	210 (75.81)	48 (17.33)	3 (1.08)	55 (19.86)	219 (79.06)	7 (2.53)	231 (83.39)	39 (14.08)
Poland	9 (0.77)	803 (68.40)	362 (30.83)	9 (0.77)	257 (21.89)	908 (77.34)	15 (1.28)	955 (81.34)	204 (17.38)
Portugal	5 (1.53)	158 (48.47)	163 (50.00)	1 (0.31)	134 (41.10)	191 (58.59)	3 (0.92)	215 (65.95)	108 (33.13)
Romania	1 (0.44)	135 (59.74)	90 (39.82)	2 (0.88)	136 (60.18)	88 (38.94)	1 (0.44)	197 (87.17)	28 (12.39)
Russian Federation	54 (1.73)	1637 (52.54)	1425 (45.73)	56 (1.80)	1523 (48.88)	1537 (49.32)	72 (2.31)	2120 (68.04)	924 (29.65)
Serbia	0 (0.00)	93 (50.82)	90 (49.18)	0 (0.00)	1 (0.55)	182 (99.45)	0 (0.00)	161 (87.98)	22 (12.02)
Slovak Republic	1 (0.36)	118 (42.29)	160 (57.35)	0 (0.00)	131 (46.95)	148 (53.05)	4 (1.43)	148 (53.05)	127 (45.52)
Slovenia	3 (3.13)	68 (70.83)	25 (26.04)	3 (3.13)	70 (72.91)	23 (23.96)	6 (6.25)	79 (82.29)	11 (11.46)
Spain	11 (0.52)	1009 (47.80)	1091 (51.68)	13 (0.62)	595 (28.19)	1503 (71.19)	18 (0.85)	1291 (61.16)	802 (37.99)
Sweden	11 (1.79)	500 (81.43)	103 (16.78)	6 (0.98)	38 (6.19)	570 (92.83)	6 (0.98)	474 (77.20)	134 (21.82)
Switzerland	1 (0.11)	567 (62.51)	339 (37.38)	2 (0.22)	174 (19.18)	731 (80.60)	3 (0.33)	671 (73.98)	233 (25.69)
Turkey	7 (0.35)	1582 (79.82)	393 (19.83)	3 (0.15)	1298 (65.49)	681 (34.36)	3 (0.15)	1800 (90.82)	179 (9.03)
Ukraine	2 (1.32)	92 (60.93)	57 (37.75)	3 (1.99)	15 (9.93)	133 (88.08)	3 (1.99)	18 (11.92)	130 (86.09)
United Kingdom[1]	0 (0.00)	4488 (46.27)	5211 (53.73)	0 (0.00)	3931 (40.53)	5768 (59.47)	0 (0.00)	6474 (66.75)	3225 (33.25)
Total	258 (0.56)	26053 (56.72)	19622 (42.72)	699 (1.52)	15207 (33.11)	30027 (65.37)	263 (0.57)	32642 (71.06)	13028 (28.36)

[1] United Kingdom: the duration of use of bronchodilators and macrolides is not specified.

Note: Inhaled antibiotics are reimbursed in all countries with the exception of Armenia.
Note: Inhaled bronchodilators are reimbursed in most countries except in Bulgaria and Serbia. In Ukraine they are reimbursed for children.
Note: Oral macrolides are reimbursed in most countries except in Bulgaria, Republic of Moldova and Serbia.

This table shows the use of three treatments: inhaled antibiotics for more than 3 months during the survey year (any kind); inhaled bronchodilators for more than 3 months during the survey year (any kind); macrolides (e.g. azithromycin) for more than three months.

Figure 7.7 Use of inhaled antibiotics in all patients seen in 2019 who have never had a transplant, by country.

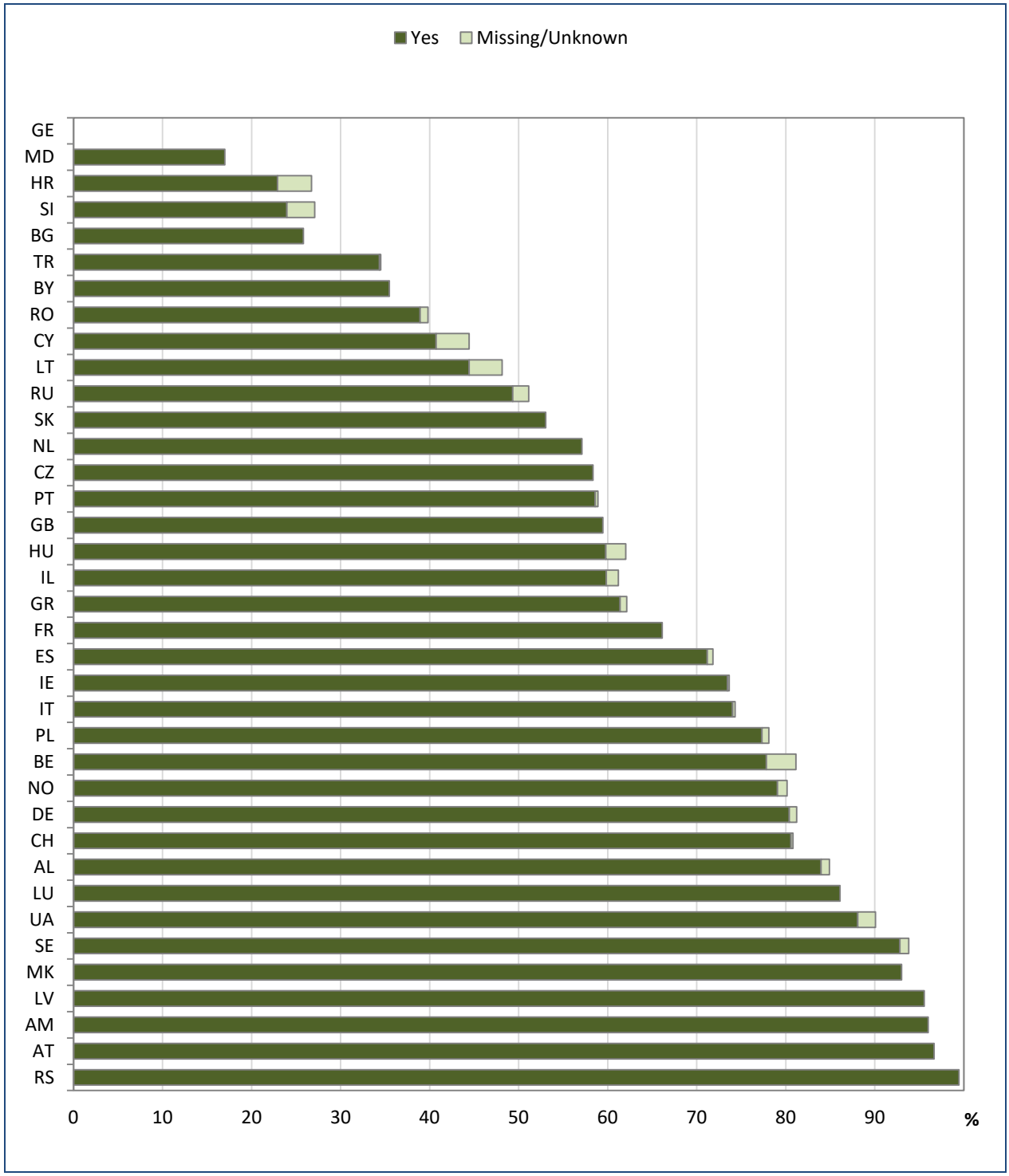


Note: We excluded from the graph the countries for which the information on inhaled antibiotics was missing for more than 10% of the patients.

Note: Inhaled antibiotics are reimbursed in all countries with the exception of Armenia.

This graph shows the use of inhaled antibiotics (of any kind) for more than three months during the survey year. The frequency varies considerably, from 1.4 to 62.3%. The dark green part of the bar shows the percentage of patients taking this drug, the light green part shows the percentage of patients for whom this information is missing.

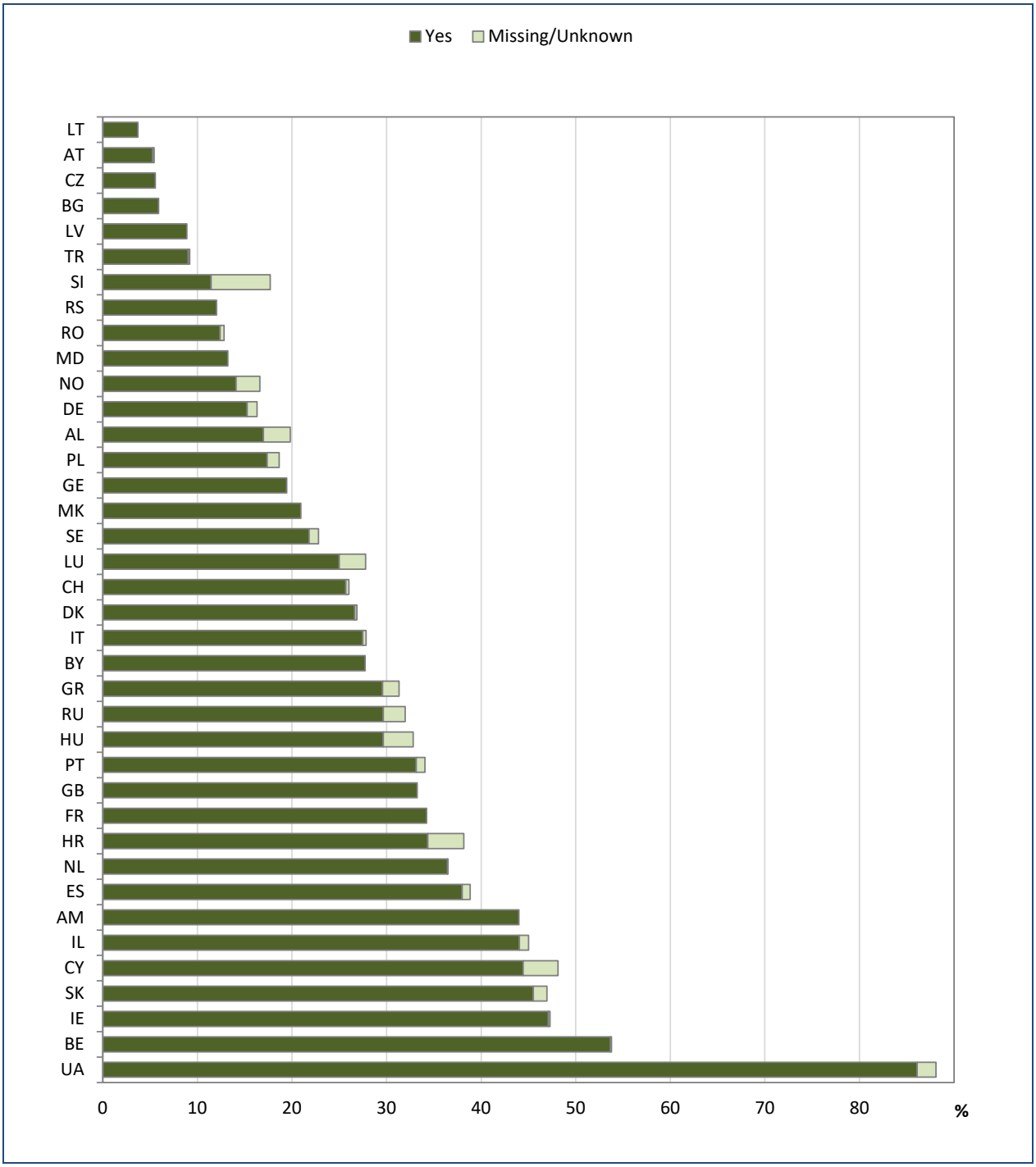
Figure 7.8 Use of bronchodilators in all patients seen in 2019 who have never had a transplant, by country.



Note: We excluded from the graph the countries for which the information on the use of bronchodilators was missing for more than 10% of the patients.
Note: United Kingdom: the duration of use of bronchodilators is not specified.
Note: Inhaled bronchodilators are reimbursed in most countries except in Bulgaria and Serbia. In Ukraine they are reimbursed for children.

This graph shows the use of bronchodilators for more than three months during the survey year. This is the most widely used inhaled medication, but still there are significant differences in frequency of use between countries. The dark green part of the bar indicates the percentage of patients taking this drug, the light green part shows the percentage of patients for whom this information is missing.

Figure 7.9 Use of macrolides in all patients seen in 2019 who have never had a transplant, by country.



Note: France: collects only use of azithromycin for macrolides.
United Kingdom: the duration of use of macrolides is not specified.
Note: Oral macrolides are reimbursed in most countries except in Bulgaria, Republic of Moldova and Serbia.

This graph shows the use of macrolides (e.g. azithromycin) for more than 3 months during 2019. Macrolides are antibiotics, but taken continuously they can also modulate the immune system, probably by their anti-inflammatory properties. Clinical studies have shown that patients with chronic Pseudomonas aeruginosa infection benefit from continuous azithromycin treatment with regard to lung function and pulmonary exacerbation rates.

The dark green part of the bar indicates the percentage of patients taking this drug, the light green part shows the percentage of patients for whom this information is missing.

Table 7.8 Use of oxygen and non-invasive positive pressure ventilation (NIPPV) in all patients seen in 2019 who have never had a transplant, by country and overall.

Country	Oxygen therapy this year number (%)			NIPPV > 3 months this year number (%)			
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes, BiPAP (Bilevel Positive Airways Pressure)	Yes, CPAP (Continuous Positive Airways Pressure)
Albania	4 (3.77)	99 (93.40)	3 (2.83)	4 (3.77)	102 (96.23)	0 (0.00)	0 (0.00)
Armenia	0 (0.00)	23 (92.00)	2 (8.00)	0 (0)	25 (100)	0 (0)	0 (0)
Austria	1 (0.14)	686 (95.54)	31 (4.32)	1 (0.14)	715 (99.58)	0 (0.00)	2 (0.28)
Belarus	0 (0.00)	150 (96.77)	5 (3.23)	0 (0.00)	154 (99.35)	0 (0.00)	1 (0.65)
Belgium	184 (16.52)	904 (81.15)	26 (2.33)	1114 (100)	0 (0)	0 (0)	0 (0)
Bulgaria	0 (0.00)	180 (96.77)	6 (3.23)	0 (0.00)	185 (99.46)	1 (0.54)	0 (0.00)
Croatia	4 (3.05)	117 (89.32)	10 (7.63)	4 (3.05)	126 (96.19)	1 (0.76)	0 (0.00)
Cyprus	1 (3.70)	26 (96.30)	0 (0.00)	1 (3.70)	26 (96.30)	0 (0.00)	0 (0.00)
Czech Republic	0 (0.00)	581 (97.98)	12 (2.02)	0 (0.00)	589 (99.33)	4 (0.67)	0 (0.00)
Denmark	279 (59.48)	188 (40.09)	2 (0.43)	278 (59.27)	189 (40.30)	0 (0.00)	2 (0.43)
France[1]	0 (0.00)	5579 (95.91)	238 (4.09)	5817 (100)	0 (0)	0 (0)	0 (0)
Georgia	0 (0.00)	71 (98.61)	1 (1.39)	1 (1.39)	70 (97.22)	0 (0.00)	1 (1.39)
Germany[2]	46 (0.78)	5443 (91.86)	436 (7.36)	49 (0.83)	5804 (97.95)	0 (0.00)	72 (1.22)
Greece	3 (0.75)	386 (96.74)	10 (2.51)	6 (1.50)	391 (98.00)	2 (0.50)	0 (0.00)
Hungary	13 (2.92)	394 (88.54)	38 (8.54)	445 (100)	0 (0)	0 (0)	0 (0)
Ireland	2 (0.18)	1059 (93.05)	77 (6.77)	2 (0.18)	1078 (94.72)	0 (0.00)	58 (5.10)
Israel	5 (0.97)	499 (97.28)	9 (1.75)	4 (0.78)	493 (96.11)	15 (2.92)	1 (0.19)
Italy	14 (0.27)	5011 (94.99)	250 (4.74)	4808 (91.15)	458 (8.68)	1 (0.02)	8 (0.15)
Latvia	0 (0.00)	44 (97.78)	1 (2.22)	0 (0)	45 (100)	0 (0)	0 (0)
Lithuania	0 (0.00)	25 (92.59)	2 (7.41)	0 (0)	27 (100)	0 (0)	0 (0)
Luxembourg	0 (0.00)	35 (97.22)	1 (2.78)	0 (0)	36 (100)	0 (0)	0 (0)

[1] France: Continuous (>3 months) NIPPV during the year of follow-up is collected but the definition differs.

[2] Germany reported all patients with NIPPV as Continuous Positive Airways Pressure (CPAP); they don't use the categories BiPAP or CPAP.

[table 7.8 continued]

Country	Oxygen therapy this year number (%)			NIPPV > 3 months this year number (%)			
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes, BiPAP (Bilevel Positive Airways Pressure)	Yes, CPAP (Continuous Positive Airways Pressure)
Rep of Moldova	0 (0.00)	51 (96.23)	2 (3.77)	0 (0)	53 (100)	0 (0)	0 (0)
The Netherlands	5 (0.36)	1354 (97.13)	35 (2.51)	5 (0.36)	1355 (97.20)	0 (0.00)	34 (2.44)
North Macedonia	0 (0.00)	119 (92.25)	10 (7.75)	1 (0.78)	128 (99.22)	0 (0.00)	0 (0.00)
Norway[3]	0 (0.00)	274 (98.92)	3 (1.08)	1 (0.36)	275 (99.28)	1 (0.36)	0 (0.00)
Poland	9 (0.77)	1105 (94.12)	60 (5.11)	13 (1.11)	1152 (98.12)	8 (0.68)	1 (0.09)
Portugal	3 (0.92)	303 (92.95)	20 (6.13)	2 (0.61)	310 (95.09)	13 (3.99)	1 (0.31)
Romania	2 (0.88)	217 (96.02)	7 (3.10)	4 (1.77)	219 (96.91)	2 (0.88)	1 (0.44)
Russian Federation	9 (0.29)	2981 (95.67)	126 (4.04)	19 (0.61)	3066 (98.39)	13 (0.42)	18 (0.58)
Serbia	0 (0.00)	178 (97.27)	5 (2.73)	0 (0.00)	181 (98.91)	2 (1.09)	0 (0.00)
Slovak Republic	4 (1.43)	264 (94.63)	11 (3.94)	3 (1.08)	274 (98.20)	0 (0.00)	2 (0.72)
Slovenia	1 (1.04)	91 (94.79)	4 (4.17)	3 (3.13)	90 (93.75)	1 (1.04)	2 (2.08)
Spain	12 (0.57)	2036 (96.45)	63 (2.98)	14 (0.66)	2082 (98.63)	11 (0.52)	4 (0.19)
Sweden	6 (0.98)	593 (96.58)	15 (2.44)	4 (0.65)	602 (98.05)	8 (1.30)	0 (0.00)
Switzerland	5 (0.55)	864 (95.26)	38 (4.19)	3 (0.33)	898 (99.01)	3 (0.33)	3 (0.33)
Turkey	3 (0.15)	1916 (96.67)	63 (3.18)	4 (0.20)	1908 (96.27)	66 (3.33)	4 (0.20)
Ukraine	3 (1.99)	120 (79.47)	28 (18.54)	2 (1.32)	149 (98.68)	0 (0.00)	0 (0.00)
United Kingdom	0 (0.00)	9142 (94.26)	557 (5.74)	0 (0.00)	9541 (98.37)	0 (0.00)	158 (1.63)
Total	618 (1.35)	43108 (93.85)	2207 (4.80)				

[3] Norway: for NIPPV collected for ventilator support (not short term use for exacerbations or drainage) the duration is not specified.

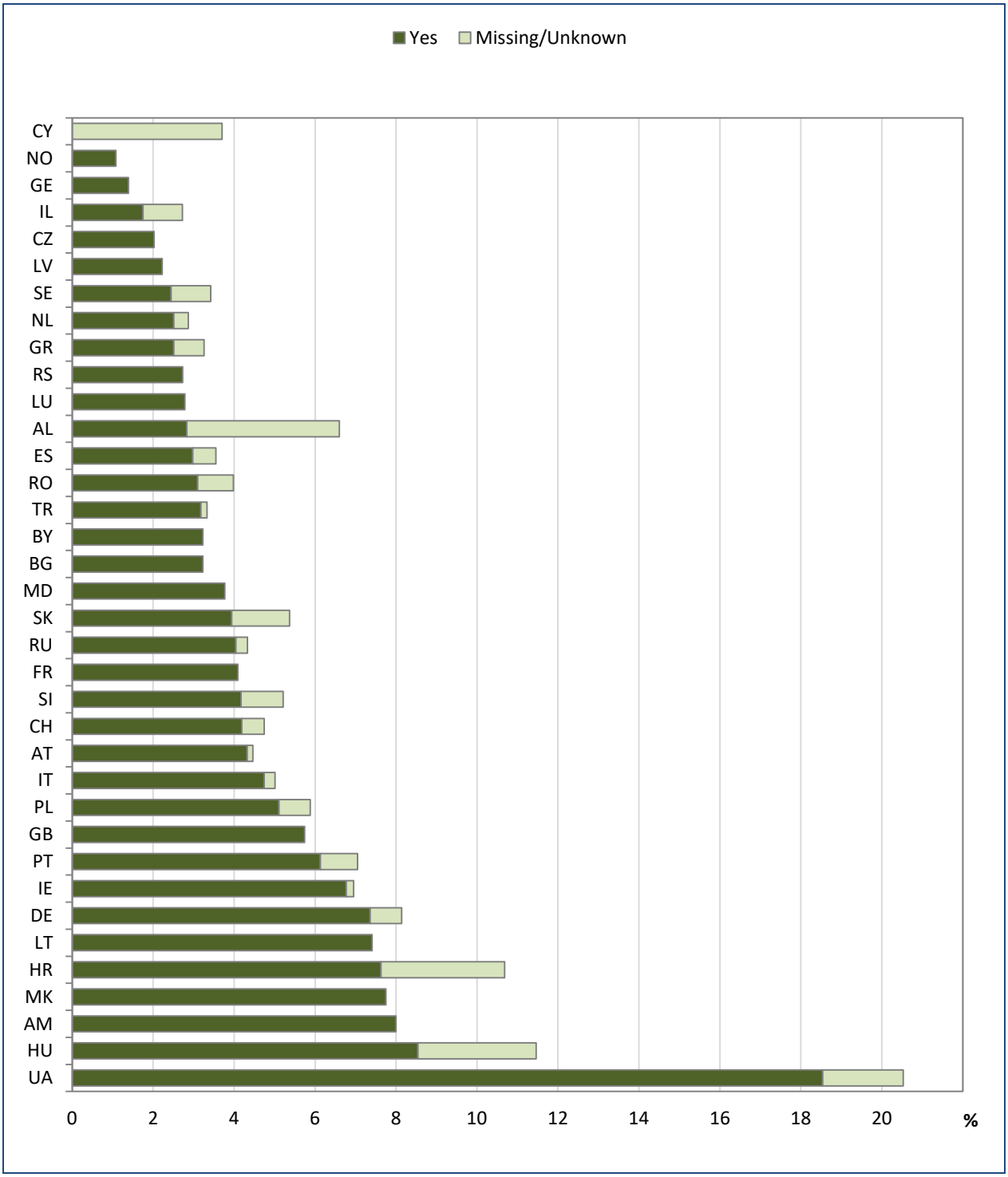
Note: For non-invasive positive pressure ventilation (NIPPV) the total percentage of missing information is higher than 10%, therefore the totals are excluded from the table.

Note: Oxygen therapy is reimbursed in most countries except in Armenia, Bulgaria, Republic of Moldova, Russian Federation, Serbia and Ukraine.

Note: Noninvasive positive pressure ventilation (NIPPV) is reimbursed in most countries except in Albania, Belarus, Belgium, Bulgaria, Hungary, North Macedonia, Republic of Moldova, Russian Federation, Serbia and Ukraine.

This table shows the use of two treatments: use of oxygen and non-invasive positive pressure ventilation (NIPPV) for more than three months.

Figure 7.10 Use of oxygen in all patients seen in 2019 who have never had a transplant, by country.



Note: We excluded from the graph the countries for which the information on the use of oxygen was missing for more than 10% of the patients.

Note: Oxygen therapy is reimbursed in most countries except in Armenia, Bulgaria, Republic of Moldova, Russian Federation, Serbia and Ukraine.

This graph shows the use of oxygen during 2019. Oxygen is used for severe lung disease. The dark green part of the bar indicates the percentage of patients using oxygen supplementation, the light green part shows the percentage of patients for whom this information is missing.

Table 7.9 Use of inhaled steroids, oral steroids and proton pump inhibitors (PPI) in all patients seen in 2019 who have never had a transplant, by country and overall.

Country	Inhaled steroids > 3 months this year number (%)			Oral steroids > 3 months this year number (%)			PPI > 3 months this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	2 (1.89)	85 (80.19)	19 (17.92)	4 (3.77)	102 (96.23)	0 (0.00)	0 (0.00)	94 (88.68)	12 (11.32)
Armenia	0 (0.00)	20 (80.00)	5 (20.00)	0 (0)	25 (100)	0 (0)	0 (0.00)	7 (28.00)	18 (72.00)
Austria	2 (0.28)	623 (86.77)	93 (12.95)	3 (0.42)	699 (97.35)	16 (2.23)	3 (0.42)	650 (90.53)	65 (9.05)
Belarus	0 (0.00)	108 (69.68)	47 (30.32)	0 (0.00)	145 (93.55)	10 (6.45)	0 (0.00)	127 (81.94)	28 (18.06)
Belgium	1 (0.09)	528 (47.40)	585 (52.51)	1 (0.09)	1080 (96.95)	33 (2.96)	0 (0.00)	560 (50.27)	554 (49.73)
Bulgaria	0 (0.00)	169 (90.86)	17 (9.14)	0 (0)	186 (100)	0 (0)	0 (0.00)	163 (87.63)	23 (12.37)
Croatia	4 (3.05)	103 (78.63)	24 (18.32)	6 (4.58)	123 (93.89)	2 (1.53)	4 (3.05)	89 (67.94)	38 (29.01)
Cyprus	1 (3.70)	16 (59.26)	10 (37.04)	1 (3.70)	26 (96.30)	0 (0.00)	1 (3.70)	23 (85.19)	3 (11.11)
Czech Republic	0 (0.00)	389 (65.60)	204 (34.40)	0 (0.00)	582 (98.15)	11 (1.85)	0 (0.00)	472 (79.60)	121 (20.40)
Denmark	469 (100)	0 (0)	0 (0)	0 (0.00)	451 (96.16)	18 (3.84)	1 (0.21)	299 (63.76)	169 (36.03)
France	0 (0.00)	2475 (42.55)	3342 (57.45)	0 (0.00)	5561 (95.60)	256 (4.40)	0 (0.00)	3515 (60.43)	2302 (39.57)
Georgia	1 (1.39)	71 (98.61)	0 (0.00)	2 (2.78)	70 (97.22)	0 (0.00)	2 (2.78)	69 (95.83)	1 (1.39)
Germany	57 (0.96)	3721 (62.80)	2147 (36.24)	59 (1.00)	5559 (93.82)	307 (5.18)	48 (0.81)	4607 (77.76)	1270 (21.43)
Greece	2 (0.50)	263 (65.92)	134 (33.58)	2 (0.50)	384 (96.24)	13 (3.26)	30 (7.52)	308 (77.19)	61 (15.29)
Hungary	445 (100)	0 (0)	0 (0)	445 (100)	0 (0)	0 (0)	445 (100)	0 (0)	0 (0)
Ireland	2 (0.18)	759 (66.69)	377 (33.13)	1138 (100)	0 (0)	0 (0)	2 (0.18)	602 (52.90)	534 (46.92)
Israel	3 (0.58)	271 (52.83)	239 (46.59)	3 (0.58)	486 (94.74)	24 (4.68)	3 (0.58)	332 (64.72)	178 (34.70)
Italy	1461 (27.70)	2762 (52.36)	1052 (19.94)	1475 (27.96)	3013 (57.12)	787 (14.92)	1417 (26.86)	2843 (53.90)	1015 (19.24)
Latvia	0 (0.00)	38 (84.44)	7 (15.56)	0 (0.00)	44 (97.78)	1 (2.22)	0 (0.00)	31 (68.89)	14 (31.11)
Lithuania	0 (0.00)	26 (96.30)	1 (3.70)	1 (3.70)	26 (96.30)	0 (0.00)	0 (0.00)	20 (74.07)	7 (25.93)
Luxembourg	0 (0.00)	18 (50.00)	18 (50.00)	1 (2.78)	33 (91.66)	2 (5.56)	0 (0.00)	16 (44.44)	20 (55.56)
Rep of Moldova	0 (0.00)	47 (88.68)	6 (11.32)	0 (0.00)	50 (94.34)	3 (5.66)	0 (0)	53 (100)	0 (0)

[table 7.9 continued]

Country	Inhaled steroids > 3 months this year number (%)			Oral steroids > 3 months this year number (%)			PPI > 3 months this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
The Netherlands	0 (0.00)	769 (55.16)	625 (44.84)	0 (0.00)	1224 (87.80)	170 (12.20)	0 (0.00)	695 (49.86)	699 (50.14)
North Macedonia	0 (0.00)	119 (92.25)	10 (7.75)	1 (0.78)	128 (99.22)	0 (0.00)	0 (0.00)	67 (51.94)	62 (48.06)
Norway	2 (0.72)	245 (88.45)	30 (10.83)	3 (1.08)	269 (97.11)	5 (1.81)	11 (3.97)	216 (77.98)	50 (18.05)
Poland	12 (1.02)	949 (80.84)	213 (18.14)	16 (1.36)	1137 (96.85)	21 (1.79)	21 (1.79)	945 (80.49)	208 (17.72)
Portugal	1 (0.31)	235 (72.08)	90 (27.61)	5 (1.53)	311 (95.40)	10 (3.07)	2 (0.61)	239 (73.32)	85 (26.07)
Romania	4 (1.77)	214 (94.69)	8 (3.54)	2 (0.88)	224 (99.12)	0 (0.00)	1 (0.44)	210 (92.92)	15 (6.64)
Russian Federation	55 (1.77)	2621 (84.11)	440 (14.12)	18 (0.58)	3011 (96.63)	87 (2.79)	72 (2.31)	2437 (78.21)	607 (19.48)
Serbia	0 (0.00)	132 (72.13)	51 (27.87)	0 (0.00)	178 (97.27)	5 (2.73)	0 (0.00)	129 (70.49)	54 (29.51)
Slovak Republic	3 (1.08)	145 (51.97)	131 (46.95)	1 (0.36)	255 (91.40)	23 (8.24)	5 (1.79)	225 (80.65)	49 (17.56)
Slovenia	6 (6.25)	81 (84.37)	9 (9.38)	7 (7.29)	88 (91.67)	1 (1.04)	8 (8.33)	65 (67.71)	23 (23.96)
Spain	14 (0.66)	1264 (59.88)	833 (39.46)	15 (0.71)	2030 (96.16)	66 (3.13)	19 (0.90)	1467 (69.49)	625 (29.61)
Sweden	8 (1.30)	414 (67.43)	192 (31.27)	8 (1.30)	589 (95.93)	17 (2.77)	12 (1.95)	465 (75.74)	137 (22.31)
Switzerland	3 (0.33)	605 (66.70)	299 (32.97)	2 (0.22)	873 (96.25)	32 (3.53)	8 (0.88)	713 (78.61)	186 (20.51)
Turkey	4 (0.20)	1580 (79.72)	398 (20.08)	3 (0.15)	1953 (98.54)	26 (1.31)	3 (0.15)	1759 (88.75)	220 (11.10)
Ukraine	4 (2.65)	76 (50.33)	71 (47.02)	2 (1.32)	142 (94.04)	7 (4.64)	5 (3.31)	30 (19.87)	116 (76.82)
United Kingdom	0 (0.00)	7930 (81.76)	1769 (18.24)	0 (0.00)	9023 (93.03)	676 (6.97)	0 (0.00)	5233 (53.95)	4466 (46.05)
Total	2566 (5.59)	29871 (65.03)	13496 (29.38)	3224 (7.02)	40080 (87.26)	2629 (5.72)	2123 (4.62)	29775 (64.82)	14035 (30.56)

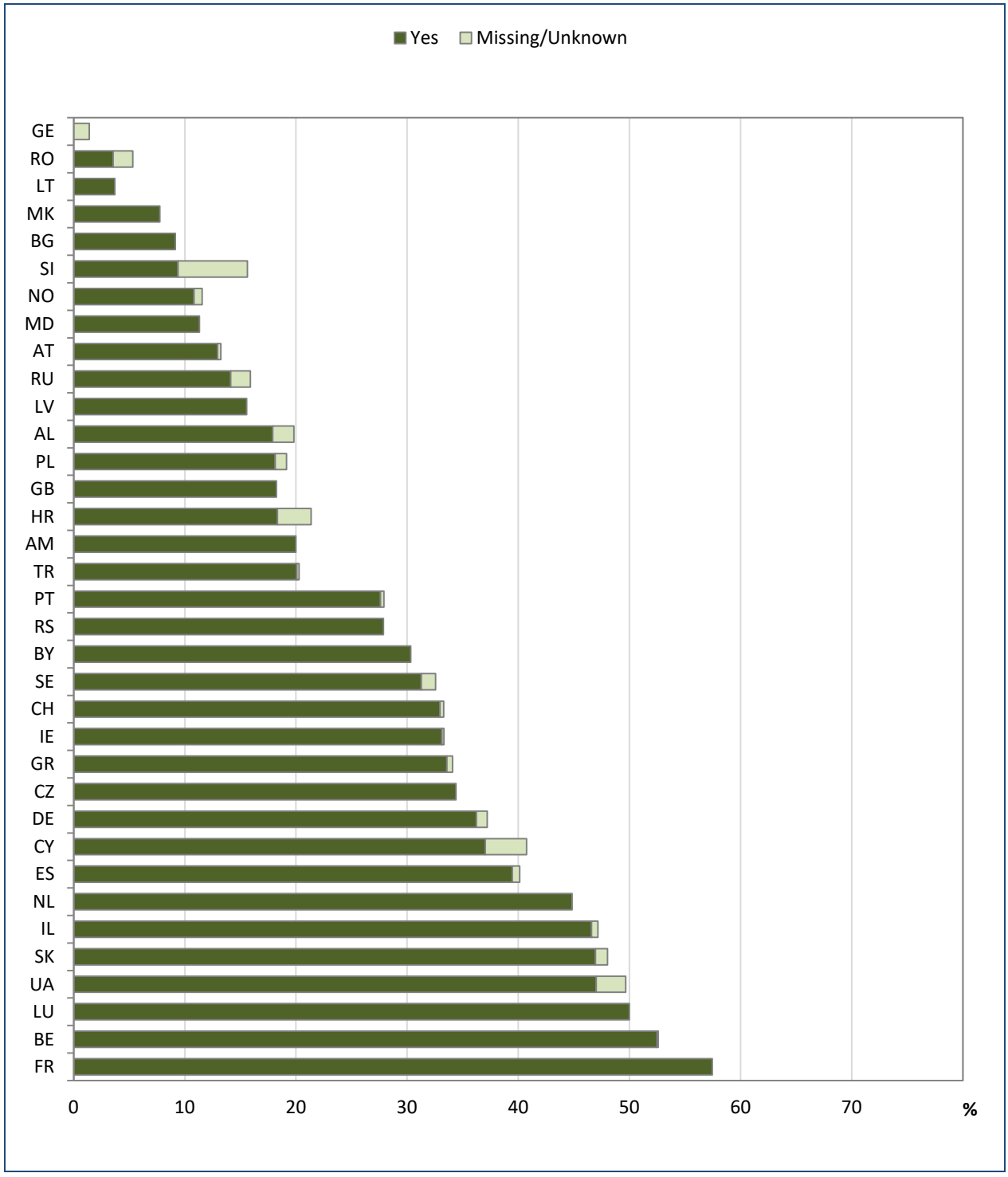
Note: Inhaled steroids are reimbursed in most countries except in Georgia, Lithuania and Serbia. In Republic of Moldova they are reimbursed for children. In Bulgaria it is reimbursed if patients also have an Asthma or chronic obstructive pulmonary disease (COPD) diagnosis.

Note: Oral steroids are reimbursed in most countries except in Bulgaria, Lithuania, Republic of Moldova, Poland and the Ukraine. In Latvia they are reimbursed for children.

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

This table shows the use of three treatments: inhaled steroids for more than 3 months; oral steroids for more than three months; proton pump inhibitors (PPI) for more than 3 months during the survey year.

Figure 7.11 Use of inhaled steroids in all patients seen in 2019 who have never had a transplant, by country.

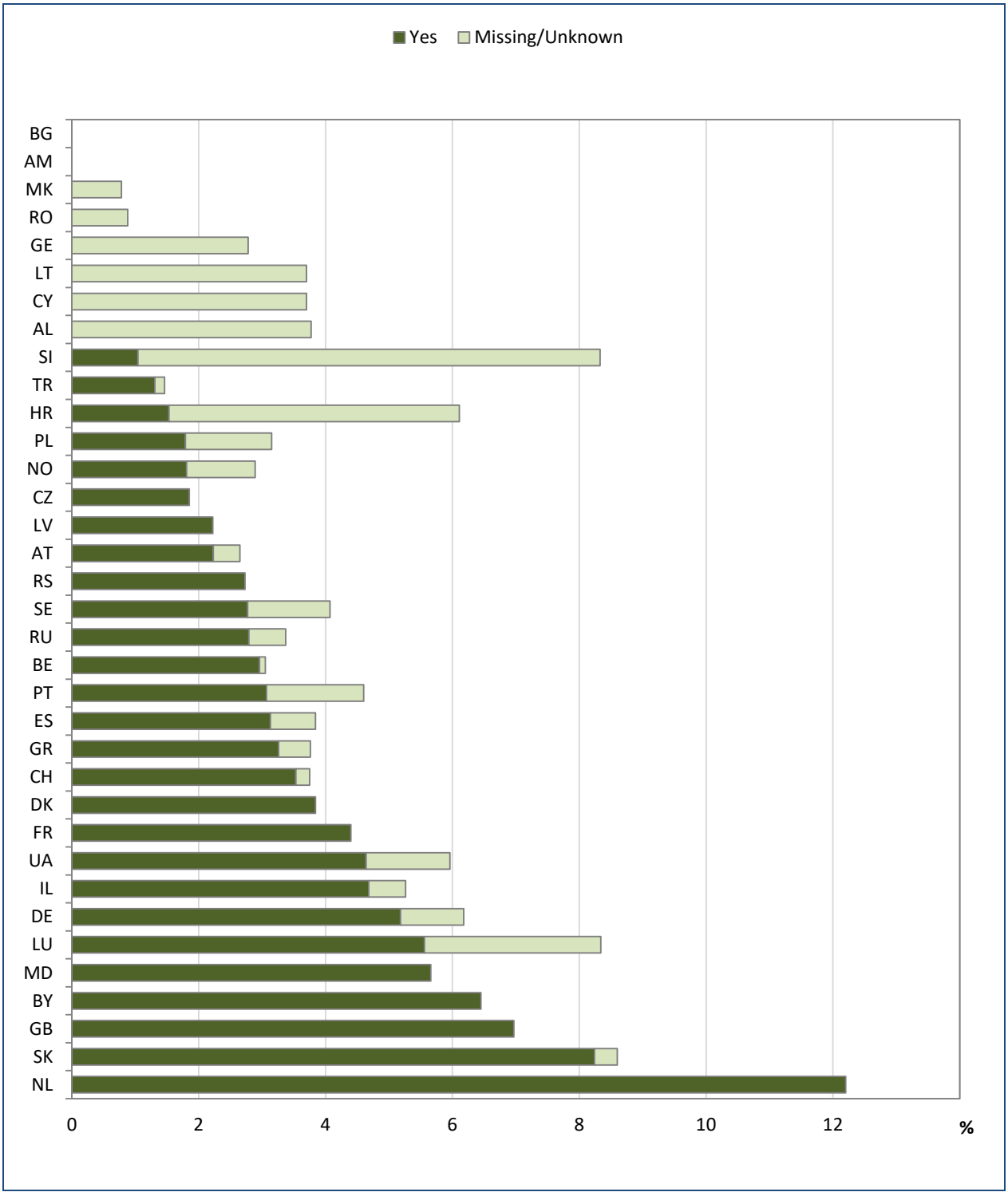


Note: We excluded from the graph the countries for which the information on use of inhaled steroids was missing for more than 10% of the patients.

Note: Inhaled steroids are reimbursed in most countries except in Georgia, Lithuania and Serbia. In Republic of Moldova they are reimbursed for children. In Bulgaria it is reimbursed if patients also have an Asthma or chronic obstructive pulmonary disease (COPD) diagnosis.

This graph shows the use of inhaled steroids for more than 3 months during the survey year. The dark green part of the bar indicates the percentage of patients taking these drugs, the light green part shows the percentage of patients for whom this information is missing.

Figure 7.12 Use of oral steroids in all patients seen in 2019 who have never had a transplant, by country.

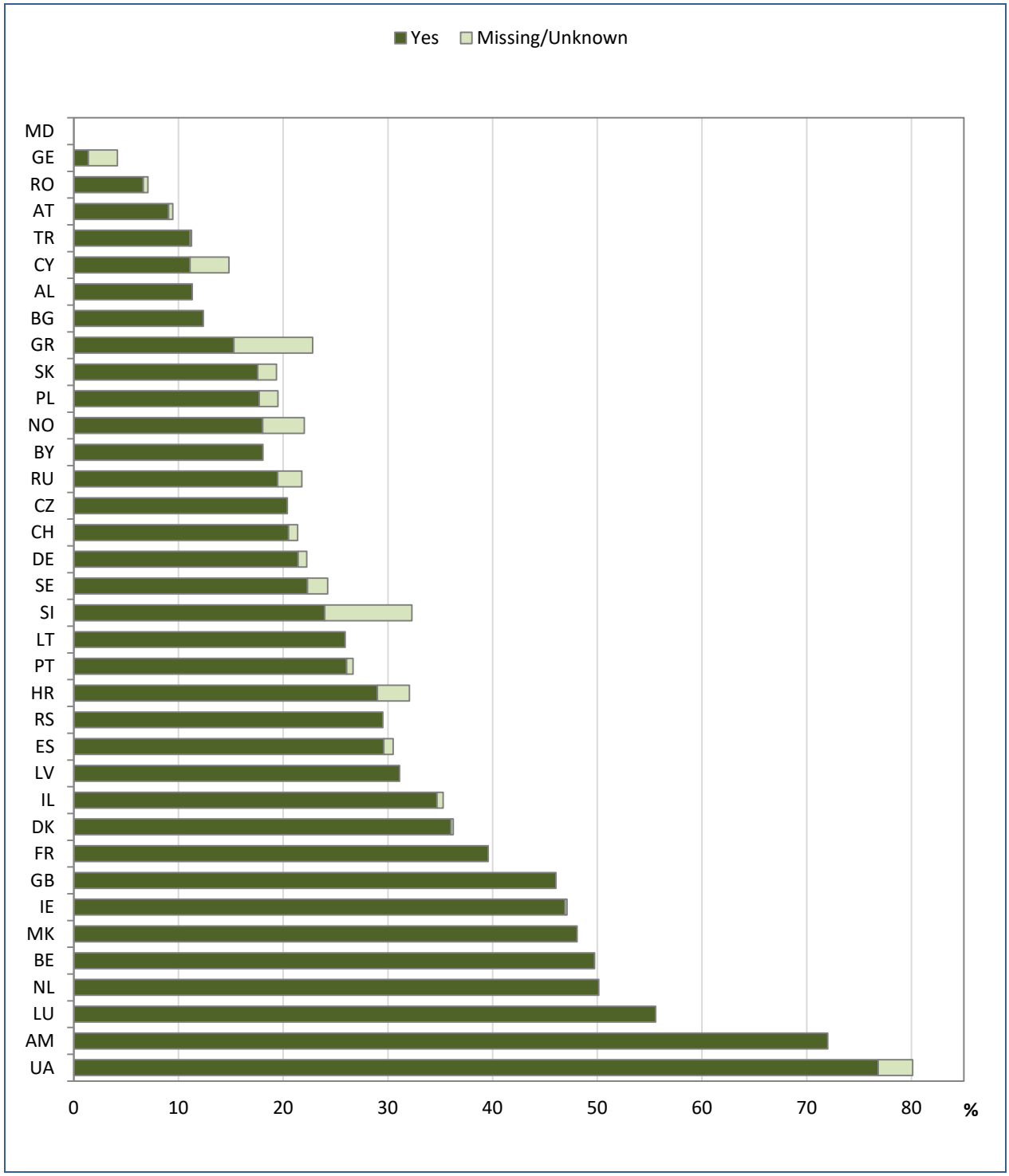


Note: We excluded from the graph the countries for which the information on use of oral steroids was missing for more than 10% of the patients.

Note: Oral steroids are reimbursed in most countries except in Bulgaria, Lithuania, Republic of Moldova, Poland and the Ukraine. In Latvia they are reimbursed for children.

This graph shows the use of oral steroids for more than 3 months during the survey year. The dark green part of the bar indicates the percentage of patients taking these drugs, the light green part shows the percentage of patients for whom this information is missing.

Figure 7.13 Use of proton pump inhibitors (PPI) in all patients seen in 2019 who have never had a transplant, by country.



Note: We excluded from the graph the countries for which the information on the use of PPI was missing for more than 10% of the patients.

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

This graph shows the use of proton pump inhibitors (PPI) for more than 3 months during the survey year. The dark green part of the bar indicates the percentage of patients using PPI, the light green part shows the percentage of patients for whom this information is missing.

Table 7.10 Use of Ivacaftor in all eligible patients seen in 2019 who had never had a transplant, by country and age group.

Country	Age at follow-up (years)	Use of Ivacaftor this year, number (%)					
		Missing/unknown		No		Yes	
Albania	**1-17**	0	(0)	2	(100)	0	(0)
Austria	**1-17**	0	(0.00)	3	(25.00)	9	(75.00)
	≥18	0	(0.00)	6	(35.29)	11	(64.71)
Belgium	**1-17**	0	(0)	0	(0)	15	(100)
	≥18	0	(0.00)	21	(38.89)	33	(61.11)
Bulgaria	**1-17**	0	(0)	5	(100)	0	(0)
	≥18	0	(0)	1	(100)	0	(0)
Croatia	**1-17**	0	(0)	1	(100)	0	(0)
	≥18	0	(0)	1	(100)	0	(0)
Czech Republic	**1-17**	0	(0.00)	3	(27.27)	8	(72.73)
	≥18	0	(0.00)	5	(29.41)	12	(70.59)
Denmark	**≥18**	0	(0.00)	11	(84.62)	2	(15.38)
France	**1-17**	0	(0.00)	10	(13.70)	63	(86.30)
	≥18	0	(0.00)	26	(23.85)	83	(76.15)
Germany	**1-17**	0	(0.00)	15	(18.99)	64	(81.01)
	≥18	3	(1.69)	40	(22.47)	135	(75.84)
Greece	**≥18**	0	(0)	0	(0)	1	(100)
Hungary	**1-17**	0	(0)	1	(100)	0	(0)
	≥18	0	(0)	2	(100)	0	(0)
Ireland	**1-17**	0	(0.00)	1	(1.30)	76	(98.70)
	≥18	0	(0.00)	22	(13.50)	141	(86.50)
Israel	**1-17**	0	(0)	0	(0)	7	(100)
	≥18	0	(0)	0	(0)	8	(100)
Italy	**1-17**	28	(34.15)	5	(6.10)	49	(59.75)
	≥18	32	(28.57)	9	(8.04)	71	(63.39)
Rep of Moldova	**1-17**	0	(0)	1	(100)	0	(0)
The Netherlands	**1-17**	0	(0)	0	(0)	10	(100)
	≥18	0	(0.00)	11	(28.21)	28	(71.79)
North Macedonia	**1-17**	0	(0)	2	(100)	0	(0)
	≥18	0	(0)	2	(100)	0	(0)
Norway	**1-17**	0	(0)	0	(0)	4	(100)
	≥18	0	(0.00)	4	(30.77)	9	(69.23)
Poland	**1-17**	0	(0)	1	(100)	0	(0)
	≥18	0	(0)	2	(100)	0	(0)
Portugal	**≥18**	0	(0.00)	3	(75.00)	1	(25.00)
Romania	**1-17**	0	(0)	2	(100)	0	(0)
Russian Federation	**1-17**	0	(0)	5	(100)	0	(0)
	≥18	0	(0)	1	(100)	0	(0)
Slovak Republic	**1-17**	0	(0)	1	(100)	0	(0)
	≥18	0	(0)	5	(100)	0	(0)
Spain	**1-17**	0	(0)	0	(0)	9	(100)
	≥18	0	(0.00)	12	(44.44)	15	(55.56)
Sweden	**1-17**	0	(0)	0	(0)	7	(100)
	≥18	0	(0.00)	12	(80.00)	3	(20.00)
Switzerland	**1-17**	0	(0)	0	(0)	8	(100)
	≥18	0	(0.00)	6	(60.00)	4	(40.00)
Turkey	**1-17**	0	(0)	14	(100)	0	(0)
	≥18	0	(0)	2	(100)	0	(0)
United Kingdom	**1-17**	0	(0.00)	37	(14.02)	227	(85.98)
	≥18	0	(0.00)	289	(42.44)	392	(57.56)
Total	**1-17**	28	(4.04)	109	(15.73)	556	(80.23)
	≥18	35	(2.37)	493	(33.38)	949	(64.25)

Note: Countries that do not have patients who are eligible for Ivacaftor are not included in the table.

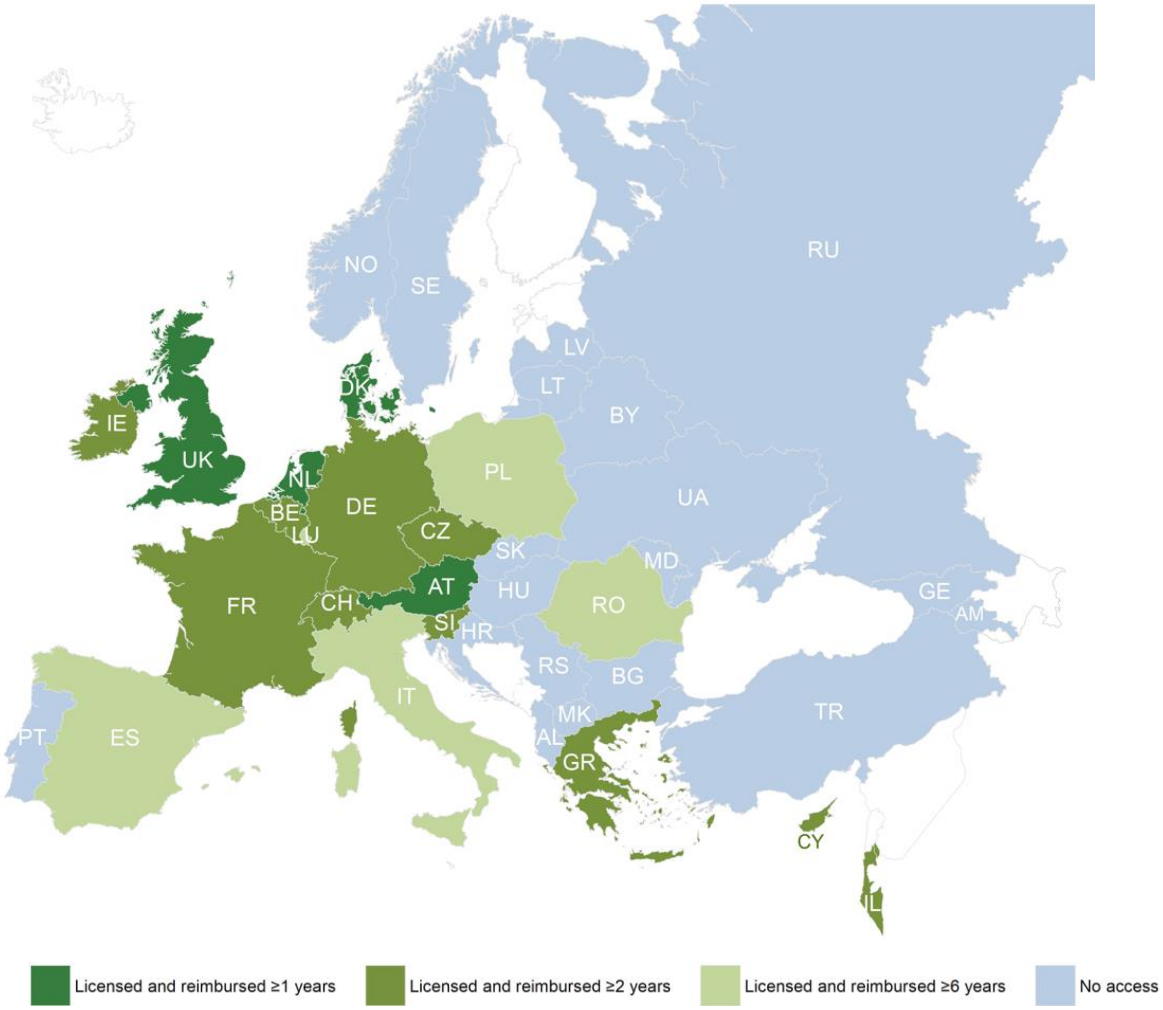
We adopted the eligibility criteria of the European Medicine Agency (EMA) for the countries in and outside Europe. Exceptions are Israel and Switzerland where specific eligibility criteria have been applied.

The eligibility criteria for Ivacaftor in 2019 are:

- The patient is 1 year (2 years in Israel) and older with at least one of the following mutations: G551D, G1244E, G1349D, G178R, G551S, S1251N, S1255P, S549N, S549R;
- The patient is 18 years and older with at least one of the following mutations: G551D, G1244E, G1349D, G178R, G551S, S1251N, S1255P, S549R, S549N, R117H (Czech Republic, France and Israel have no approval for this mutation).

In countries where the therapy is licensed but not reimbursed, or not licensed nor reimbursed, eligible patients may sometimes have access to the therapy because of a clinical trial or a compassionate use programme.

Figure 7.14 Countries where Ivacaftor is reimbursed in year 2019.



Note: Belgium: Reimbursement only for patients with two CF-causing mutations or sweat chloride > 60mmol/L and with pulmonary or GI symptoms and/or growth deviation. Excluded are patients with the R117H mutation and patients with a lung transplantation.

In this graph we highlighted the countries where Ivacaftor was licensed and reimbursed in 2019. If the therapy is reimbursed for patients of 1 year and older the country is shaded in dark green, for patients of 2 years and older the country is marked in green, and for patients of 6 years and older in light green.

Table 7.11 Use of Lumacaftor/Ivacaftor in all eligible patients seen in 2019 who had never had a transplant, by country and age group.

Country	Age at follow-up (years)	Use of Lumacaftor/Ivacaftor this year number (%)					
		Missing/unknown		No		Yes	
Albania	**2-17**	0	(0)	63	(100)	0	(0)
	≥18	0	(0.00)	5	(83.33)	1	(16.67)
Armenia	**2-17**	0	(0)	1	(100)	0	(0)
Austria	**2-17**	0	(0.00)	101	(61.96)	62	(38.04)
	≥18	0	(0.00)	108	(62.07)	66	(37.93)
Belarus	**2-17**	0	(0)	45	(100)	0	(0)
	≥18	0	(0)	4	(100)	0	(0)
Belgium	**2-17**	0	(0.00)	172	(92.47)	14	(7.53)
	≥18	0	(0.00)	225	(83.64)	44	(16.36)
Bulgaria	**2-17**	0	(0)	43	(100)	0	(0)
	≥18	0	(0)	21	(100)	0	(0)
Croatia	**2-17**	0	(0)	48	(100)	0	(0)
	≥18	0	(0)	30	(100)	0	(0)
Cyprus	**2-17**	0	(0.00)	2	(50.00)	2	(50.00)
	≥18	0	(0)	0	(0)	1	(100)
Czech Republic	**2-17**	0	(0.00)	119	(96.75)	4	(3.25)
	≥18	0	(0.00)	92	(82.88)	19	(17.12)
Denmark	**2-17**	0	(0.00)	14	(11.67)	106	(88.33)
	≥18	0	(0.00)	106	(59.55)	72	(40.45)
France	**2-17**	0	(0.00)	721	(66.39)	365	(33.61)
	≥18	0	(0.00)	459	(35.80)	823	(64.20)
Germany	**2-17**	7	(0.66)	694	(65.47)	359	(33.87)
	≥18	22	(1.42)	1124	(72.42)	406	(26.16)
Georgia	**2-17**	0	(0)	1	(100)	0	(0)
Greece	**2-17**	0	(0.00)	14	(37.84)	23	(62.16)
	≥18	1	(1.14)	16	(18.18)	71	(80.68)
Hungary	**2-17**	0	(0)	100	(100)	0	(0)
	≥18	0	(0)	66	(100)	0	(0)
Ireland	**2-17**	0	(0.00)	15	(5.49)	258	(94.51)
	≥18	2	(0.61)	78	(23.71)	249	(75.68)
Israel	**6-17**	0	(0.00)	3	(17.65)	14	(82.35)
	≥18	0	(0.00)	9	(26.47)	25	(73.53)
Italy	**2-17**	240	(53.57)	60	(13.39)	148	(33.04)
	≥18	129	(21.68)	69	(11.60)	397	(66.72)
Latvia	**2-17**	0	(0)	14	(100)	0	(0)
	≥18	0	(0)	8	(100)	0	(0)
Lithuania	**2-17**	0	(0)	4	(100)	0	(0)
	≥18	0	(0)	4	(100)	0	(0)
Luxembourg	**2-17**	0	(0.00)	4	(66.67)	2	(33.33)
	≥18	0	(0.00)	6	(46.15)	7	(53.85)
Rep of Moldova	**2-17**	0	(0)	19	(100)	0	(0)
	≥18	0	(0)	1	(100)	0	(0)
The Netherlands	**2-17**	0	(0.00)	28	(9.30)	273	(90.70)
	≥18	0	(0.00)	131	(29.11)	319	(70.89)
North Macedonia	**2-17**	0	(0)	46	(100)	0	(0)
	≥18	0	(0)	17	(100)	0	(0)
Norway	**2-17**	0	(0.00)	32	(82.05)	7	(17.95)
	≥18	0	(0.00)	29	(54.72)	24	(45.28)
Poland	**2-17**	3	(0.93)	316	(97.83)	4	(1.24)
	≥18	0	(0.00)	141	(95.92)	6	(4.08)
Portugal	**2-17**	0	(0.00)	88	(87.13)	13	(12.87)
	≥18	0	(0.00)	36	(75.00)	12	(25.00)

[table 7.11 continued]

Country	Age at follow-up (years)	Use of Lumacaftor/Ivacaftor this year number (%)					
		Missing/unknown		No		Yes	
Romania	**2-17**	0	(0)	84	(100)	0	(0)
	≥18	0	(0)	5	(100)	0	(0)
Russian Federation	**2-17**	0	(0)	654	(100)	0	(0)
	≥18	0	(0.00)	169	(98.26)	3	(1.74)
Serbia	**2-17**	0	(0)	64	(100)	0	(0)
	≥18	0	(0)	33	(100)	0	(0)
Slovak Republic	**2-17**	0	(0.00)	48	(97.96)	1	(2.04)
	≥18	0	(0)	48	(100)	0	(0)
Slovenia	**2-17**	0	(0)	34	(100)	0	(0)
	≥18	0	(0)	20	(100)	0	(0)
Spain	**2-17**	0	(0.00)	259	(88.1)	35	(11.9)
	≥18	0	(0.00)	235	(92.16)	20	(7.84)
Sweden	**2-17**	0	(0.00)	10	(8.33)	110	(91.67)
	≥18	0	(0.00)	31	(20.67)	119	(79.33)
Switzerland	**6-17**	0	(0.00)	118	(97.52)	3	(2.48)
	≥18	0	(0.00)	155	(69.51)	68	(30.49)
Turkey	**2-17**	0	(0)	192	(100)	0	(0)
	≥18	0	(0)	33	(100)	0	(0)
Ukraine	**2-17**	1	(2.22)	42	(93.33)	2	(4.44)
	≥18	0	(0)	6	(100)	0	(0)
United Kingdom	**2-17**	0	(0.00)	1979	(96.91)	63	(3.09)
	≥18	0	(0.00)	2164	(85.29)	376	(14.89)
Total	**2-17**	251	(3.00)	6251	(74.68)	1868	(22.32)
	≥18	154	(1.72)	5684	(63.39)	3128	(34.89)

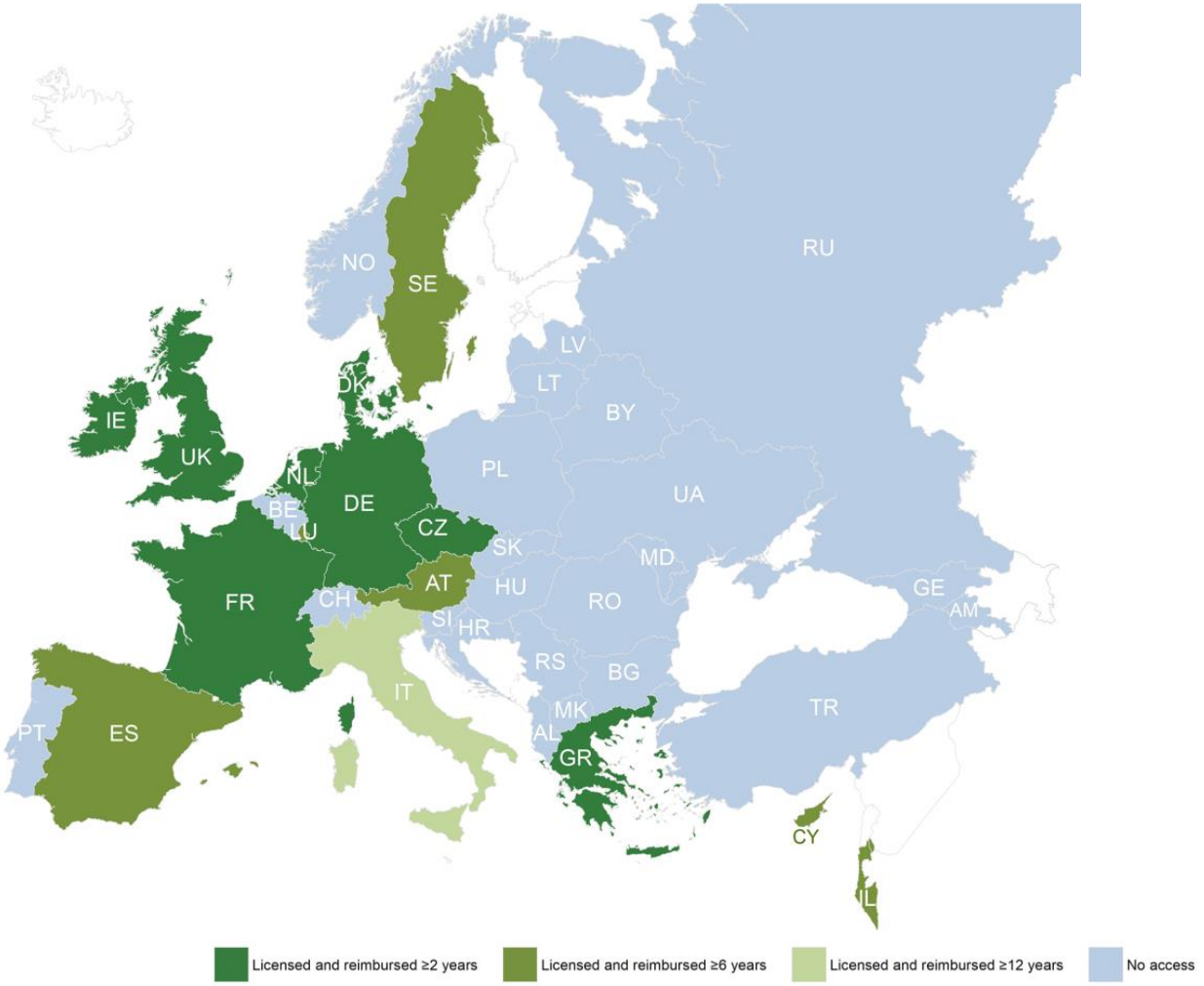
Note: Countries that do not have patients who are eligible for Lumacaftor/Ivacaftor are not included in the table.

We adopted the eligibility criteria of the European Medicine Agency (EMA) criteria for the countries in and outside Europe. Exceptions are Israel and Switzerland where specific eligibility criteria have been applied.

The eligibility criteria for Lumacaftor/Ivacaftor in 2019 are:
The patient must be 2 years (6 years for Israel and Switzerland) or older and have the mutation F508del/ F508del.

In countries where the therapy is licensed but not reimbursed, or not licensed nor reimbursed, eligible patients may sometimes have access to the therapy because of a clinical trial or a compassionate use programme.

Figure 7.15 Countries where Lumacaftor/Ivacaftor is reimbursed in year 2019.



In this graph we highlighted the countries where Lumacaftor/Ivacaftor was licensed and reimbursed in 2019. If the therapy is reimbursed for patients who are 2 years and older the country is shaded in dark green, if for patients who are 6 years and older the country is marked in green, and for patients of 12 years and older in light green.

Table 7.12 Use of Tezacaftor/Ivacaftor in all eligible patients seen in 2019 who had never had a transplant, by country and age group.

Country	Age at follow-up (years)	Use of Tezacaftor/Ivacaftor this year number (%)					
		Missing/unknown		No		Yes	
Albania	**12-17**	0	(0)	22	(100)	0	(0)
	≥18	0	(0)	6	(100)	0	(0)
Austria	**12-17**	0	(0)	48	(87.27)	7	(12.73)
	≥18	1	(0.53)	138	(73.8)	48	(25.67)
Belarus	**12-17**	0	(0)	12	(100)	0	(0)
	≥18	0	(0)	4	(100)	0	(0)
Belgium	**12-17**	0	(0)	70	(93.33)	5	(6.67)
	≥18	0	(0)	332	(97.08)	10	(2.92)
Bulgaria	**12-17**	0	(0)	16	(100)	0	(0)
	≥18	0	(0)	27	(100)	0	(0)
Croatia	**12-17**	0	(0)	19	(100)	0	(0)
	≥18	0	(0)	30	(100)	0	(0)
Cyprus	**12-17**	0	(0)	2	(100)	0	(0)
	≥18	0	(0)	2	(100)	0	(0)
Czech Republic	**12-17**	0	(0)	50	(100)	0	(0)
	≥18	0	(0)	136	(100)	0	(0)
Denmark	**12-17**	0	(0)	13	(24.53)	40	(75.47)
	≥18	0	(0)	61	(33.15)	123	(66.85)
France	**12-17**	501	(99.8)	0	(0)	1	(0.2)
	≥18	1566	(99.49)	0	(0)	8	(0.51)
Germany	**12-17**	3	(0.7)	335	(78.45)	89	(20.84)
	≥18	22	(1.29)	968	(56.84)	713	(41.87)
Greece	**12-17**	0	(0)	15	(100)	0	(0)
	≥18	0	(0)	88	(81.48)	20	(18.52)
Hungary	**12-17**	0	(0)	39	(100)	0	(0)
	≥18	0	(0)	72	(100)	0	(0)
Ireland	**12-17**	0	(0)	100	(93.46)	7	(6.54)
	≥18	2	(0.58)	203	(58.84)	140	(40.58)
Israel	**12-17**	0	(0)	4	(28.57)	10	(71.43)
	≥18	0	(0)	19	(34.55)	36	(65.45)
Italy	**12-17**	157	(77.34)	38	(18.72)	8	(3.94)
	≥18	641	(73.76)	192	(22.09)	36	(4.14)
Latvia	**12-17**	0	(0)	7	(100)	0	(0)
	≥18	0	(0)	8	(100)	0	(0)
Lithuania	**12-17**	0	(0)	1	(100)	0	(0)
	≥18	0	(0)	9	(100)	0	(0)
Luxembourg	**12-17**	0	(0)	1	(100)	0	(0)
	≥18	0	(0)	14	(100)	0	(0)
Rep of Moldova	**12-17**	0	(0)	4	(100)	0	(0)
	≥18	0	(0)	1	(100)	0	(0)
The Netherlands	**12-17**	0	(0)	27	(20.61)	104	(79.39)
	≥18	2	(0.35)	340	(59.44)	230	(40.21)
North Macedonia	**12-17**	0	(0)	19	(100)	0	(0)
	≥18	0	(0)	17	(100)	0	(0)
Norway	**12-17**	0	(0)	15	(100)	0	(0)
	≥18	0	(0)	75	(100)	0	(0)
Poland	**12-17**	1	(0.78)	127	(99.22)	0	(0)
	≥18	0	(0)	174	(100)	0	(0)
Portugal	**12-17**	0	(0)	48	(100)	0	(0)
	≥18	0	(0)	59	(100)	0	(0)

[table 7.12 continued]

Country	Age at follow-up (years)	Use of Tezacaftor/Ivacaftor this year number (%)					
		Missing/unknown		No		Yes	
Romania	**12-17**	0	(0)	35	(100)	0	(0)
	≥18	0	(0)	5	(100)	0	(0)
Russian Federation	**12-17**	0	(0)	197	(99.49)	1	(0.51)
	≥18	0	(0)	230	(98.71)	3	(1.29)
Serbia	**12-17**	0	(0)	27	(100)	0	(0)
	≥18	0	(0)	34	(100)	0	(0)
Slovak Republic	**12-17**	0	(0)	22	(100)	0	(0)
	≥18	0	(0)	65	(98.48)	1	(1.52)
Slovenia	**12-17**	0	(0)	4	(100)	0	(0)
	≥18	0	(0)	20	(100)	0	(0)
Spain	**12-17**	0	(0)	138	(92)	12	(8)
	≥18	0	(0)	299	(85.43)	51	(14.57)
Sweden	**12-17**	0	(0)	45	(100)	0	(0)
	≥18	0	(0)	162	(93.1)	12	(6.9)
Switzerland	**12-17**	0	(0)	52	(92.86)	4	(7.14)
	≥18	0	(0)	202	(85.23)	35	(14.77)
Turkey	**12-17**	0	(0)	66	(100)	0	(0)
	≥18	0	(0)	43	(100)	0	(0)
Ukraine	**12-17**	1	(5.56)	17	(94.44)	0	(0)
	≥18	0	(0)	8	(100)	0	(0)
United Kingdom	**12-17**	0	(0)	776	(98.73)	10	(1.27)
	≥18	0	(0)	2723	(93.57)	187	(6.43)
Total	**12-17**	663	(19.66)	2411	(71.50)	298	(8.84)
	≥18	2234	(20.97)	6766	(63.51)	1653	(15.52)

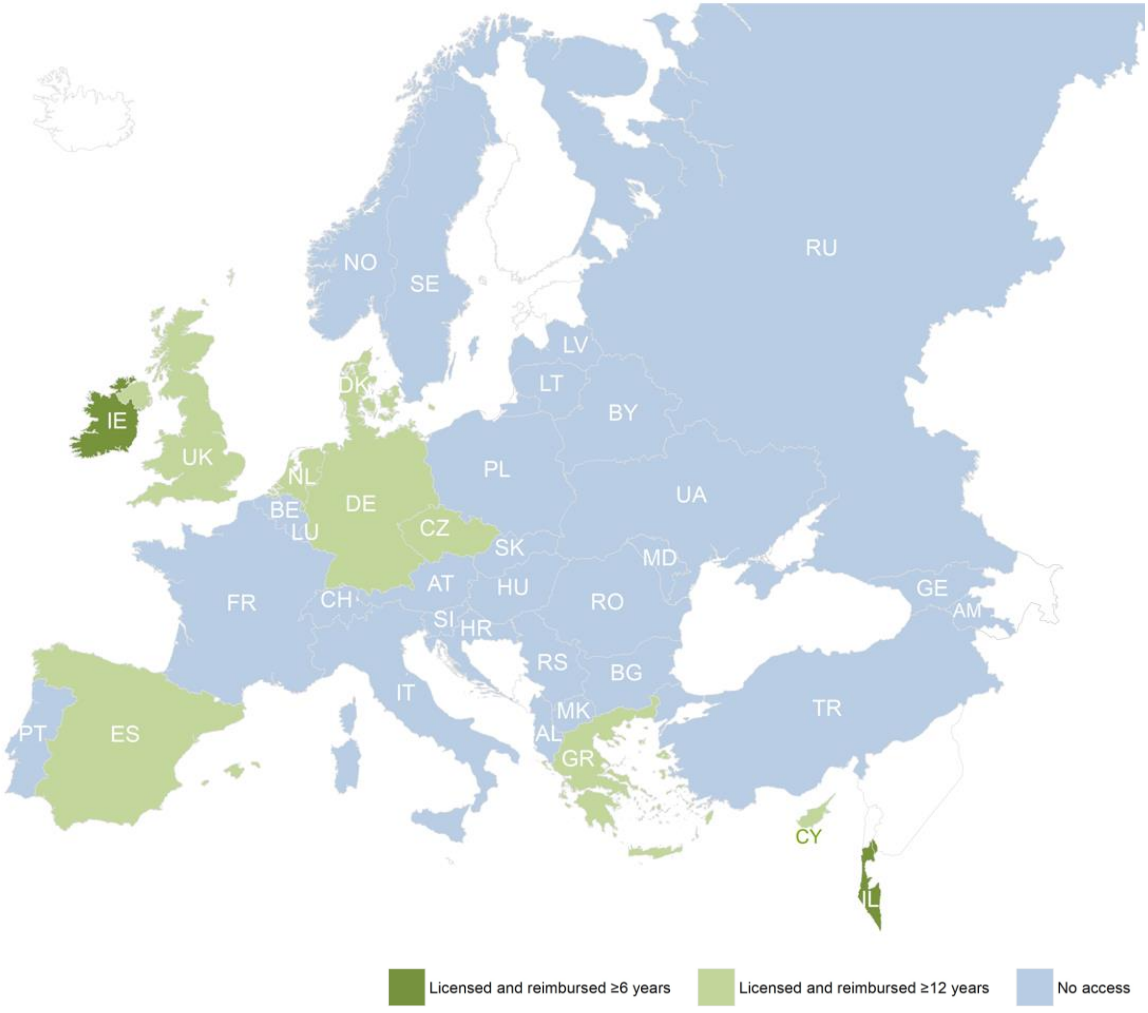
Note: Countries that do not have patients who are eligible for Tezacaftor/Ivacaftor are not included in the table.

We adopted the eligibility criteria of the European Medicine Agency (EMA) criteria for the countries, in and outside Europe. Exceptions are Israel and Switzerland where specific eligibility criteria have been applied.

The eligibility criteria for Tezacaftor/Ivacaftor in 2019 are:
The patients is 12 years or older and is F508del homozygote, or F508del heterozygote with one of the following mutations: P67L, R117C, L206W, R352Q, A455E, D579G, 711+3A→G, S945L, S977F, R1070W, D1152H, 2789+5G→A, 3272-26A→G, 3849+10kbC→T.

In countries where the therapy is licensed but not reimbursed, or not licensed nor reimbursed, eligible patients may sometimes have access to the therapy because of a clinical trial or a compassionate use programme.

Figure 7.16 Countries where Tezacaftor/Ivacaftor is reimbursed in year 2019.



Note: The Netherlands: only for patients with the mutation F508del homozygote.

In this graph we highlighted the countries where Tezacaftor/Ivacaftor was licensed and reimbursed in 2019. If the therapy is reimbursed for patients who are 6 years and older the country is shaded in green and if reimbursed for patients of 12 years and older light green is used.

8. Transplantation

We ask the countries whether their patients are transplanted or not (lung, liver, other transplant), and if they are, in which year they had their (latest) transplant. In some countries transplanted patients are no longer registered in the database of the CF centres or the national CF registry because the patients have been transferred to a transplant centre. For this reason, the figures below may report a lower number of transplanted patients than the true number, but it has not been possible to acquire more accurate data.

Table 8.1 Number of patients living in 2019 with transplanted lung(s), by age and sex.

Age	Males	Females	Total	Transplants carried out in 2019
5-9	0	2	2	1
10-14	5	11	16	3
15-19	34	59	93	29
20-24	91	143	234	47
25-29	192	241	433	56
30-34	259	247	506	39
35-39	299	280	579	54
40-44	202	203	405	26
45+	323	254	577	40
Total	1405	1440	2845	295

This table shows the number of patients alive in 2019 who have had one or more lung transplant(s) at some time in their life, by age group, as well as the number of patients transplanted during 2019.

Table 8.2 Number of patients living in 2019 with transplanted liver, by age and sex.

Age	Males	Females	Total	Transplants carried out In 2019
0-4	1	0	1	1
5-9	4	0	4	2
10-14	9	8	17	3
15-19	32	16	48	3
20-24	33	17	50	3
25-29	35	17	52	5
30-34	32	22	54	2
35-39	23	7	30	1
40-44	14	6	20	0
45+	13	8	21	1
Total	196	101	297	21

This table shows the number of patients alive in 2019 who have had a liver transplant at some time in their life, by age group, as well as the number of patients transplanted during 2019.

Table 8.3 Number of patients living in 2019 with transplanted kidney, by age and sex.

Age	Males	Females	Total	Transplants carried out In 2019
5-9	1	0	1	0
15-19	1	1	2	2
20-24	3	1	4	1
25-29	2	8	10	3
30-34	6	9	15	4
35-39	15	17	32	8
40-44	20	14	34	0
45+	22	21	43	2
Total	70	71	141	20

Note: Hungary does not collect information on kidney transplant and the Netherlands have more than 90% missing.

This table shows the number of patients alive in 2019 who have had a kidney transplant at some time in their life, by age group, as well as the number of patients transplanted during 2019.

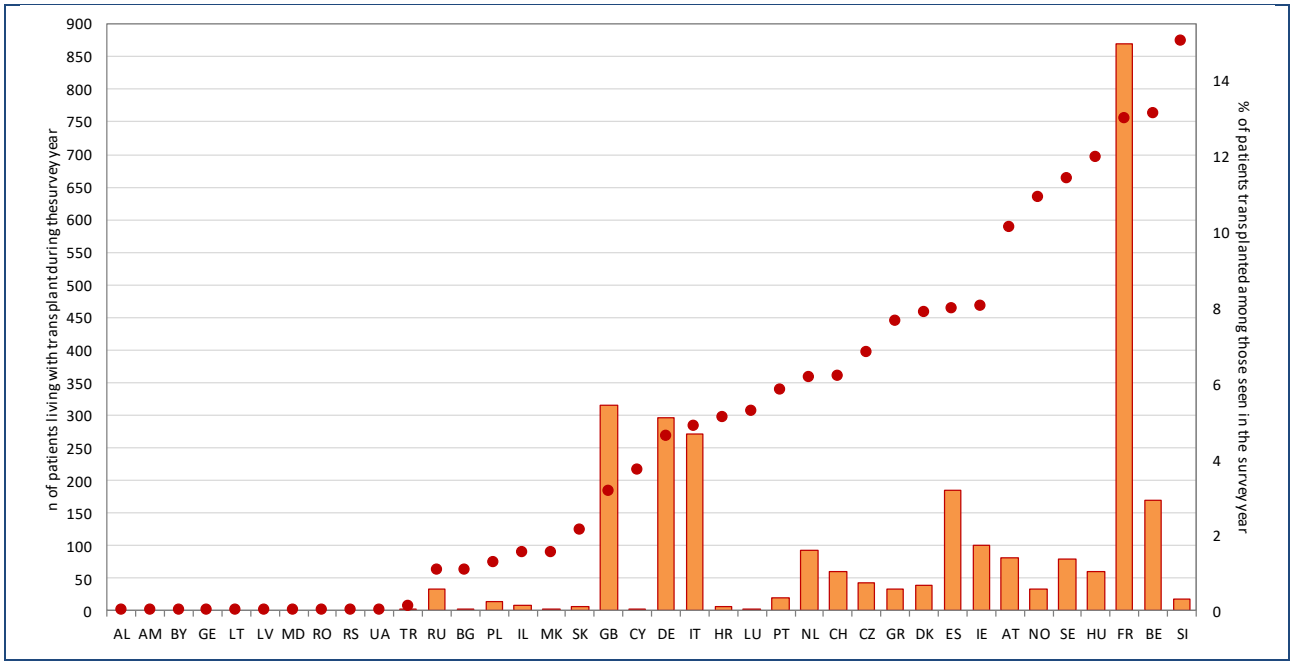
Table 8.4 Number of patients living in 2019 with other transplanted organs (not lung, liver, kidney), by age and sex.

Age	Males	Females	Total	Other Transplants carried out in 2019
0-4	0	1	1	1
10-14	0	2	2	0
15-19	0	1	1	0
20-24	4	4	8	0
25-29	3	6	9	0
30-34	2	5	7	0
35-39	6	3	9	2
40-44	7	6	13	0
45+	12	8	20	2
Total	34	36	70	5

Note: Hungary does not collect information on other organ transplant and the Netherlands have more than 90% missing.

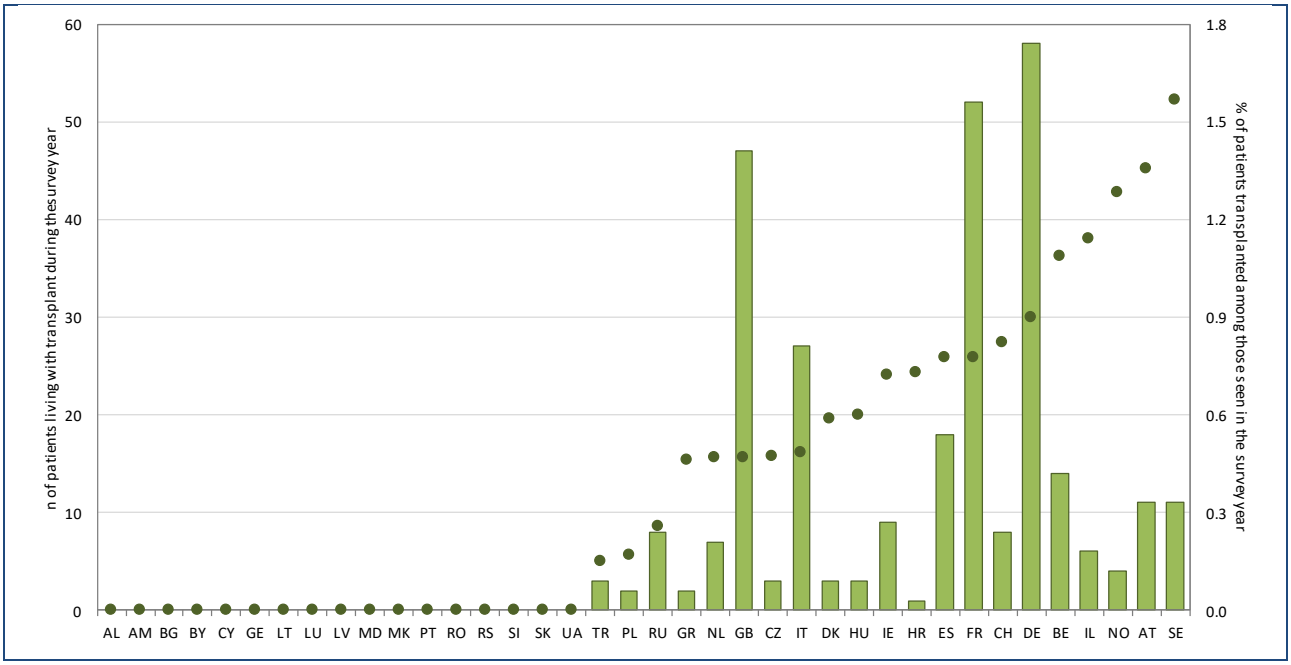
This table shows the number of patients alive in 2019 who have had an organ transplant different from lung, liver or kidney at some time in their life, by age group, as well as the number of patients transplanted during 2019.

Figure 8.1 Number of patients living in 2019 with transplanted lungs, by country.



This graph shows the number of patients alive at 31/12/2019 who have had one or more lung(s) transplant (orange bars) at some point in their life. The red dots (right axis) show the percentage of patients that are living with transplant lungs in 2019 among the patients that were seen in 2019.

Figure 8.2 Number of patients living in 2019 with transplanted liver, by country.



This graph shows the number of patients alive at 31/12/2019 who have had a liver transplant (green bars) at some point in their life. The dark green dots (right axis) show the percentage of patients that are living with transplanted liver in 2019 among the patients that were seen in 2019. Note that on the vertical axis the number of patients who had a liver transplant is much lower than the number who had a lung transplant. The main reason for this is that liver disease is only found in a subset of CF patients, whereas lung disease affects almost all patients.

9. Mortality

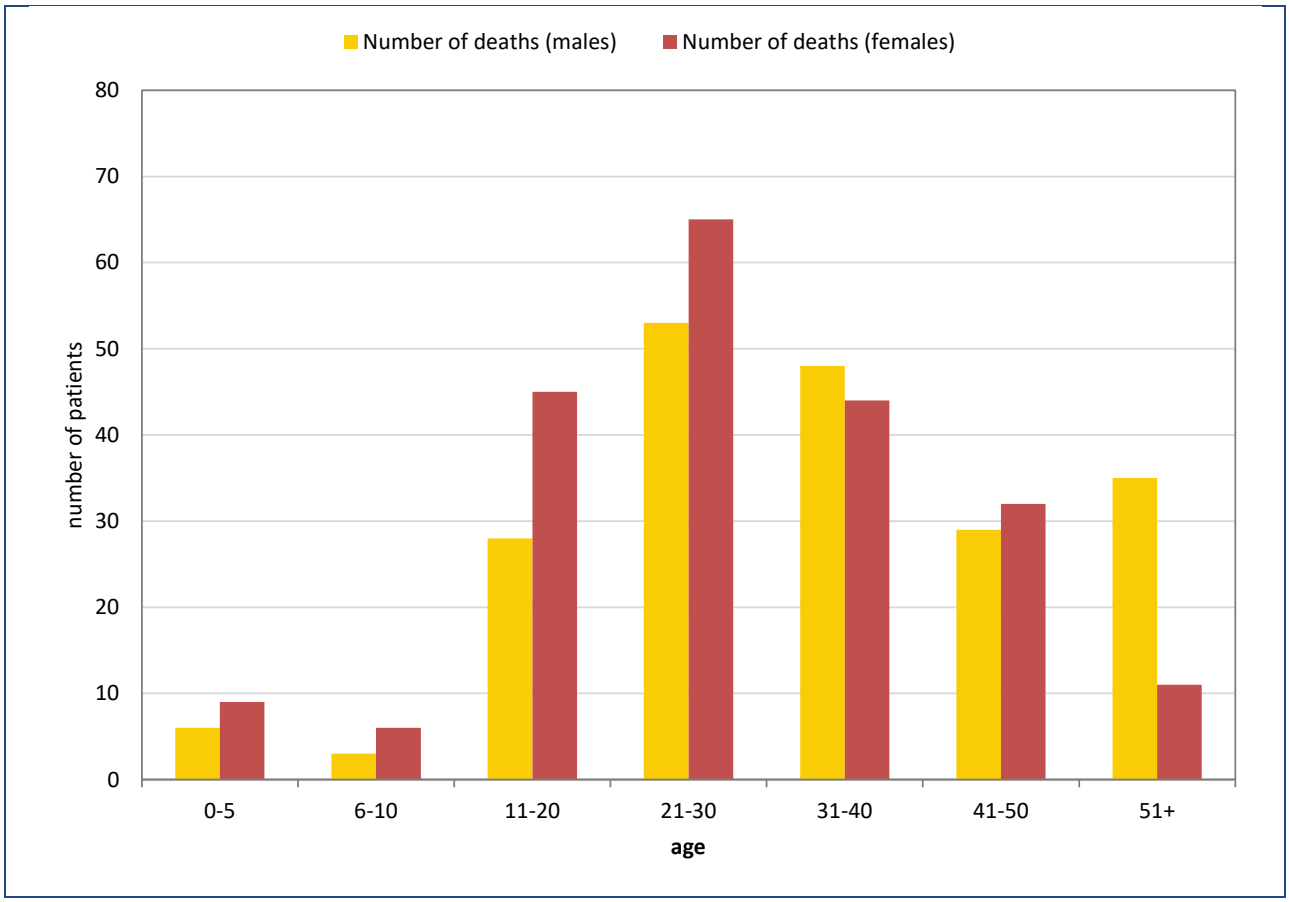
Table 9.1 Number of deaths in 2019, by age and sex.

Age at death	Number of male patients	% of deaths in this age group (of all male deaths)	Number of female patients	% of deaths in this age group (of all female deaths)	Total	% Total
0-5	6	2.97	9	4.25	15	3.62
6-10	3	1.49	6	2.83	9	2.17
11-20	28	13.86	45	21.23	73	17.63
21-30	53	26.23	65	30.66	118	28.51
31-40	48	23.76	44	20.75	92	22.23
41-50	29	14.36	32	15.09	61	14.73
51+	35	17.33	11	5.19	46	11.11
Total	202	100.00	212	100.00	414	100.00

Note: For the United Kingdom, all patients with a confirmed diagnosis of CF were included (N=10,665). The total number of patients presented is 49,925.

This table shows the number of deaths in 2019 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 21-30 years. It is possible that the number of deceased patients is under reported because some of the patients were not seen at the centre during the year, and therefore the information may not have been recorded.

Figure 9.1 Age at death distribution of patients deceased in 2019, by sex.



This graph shows the distribution of age at death of patients who died in 2019, separately by males (yellow) and females (red).

Table 9.2 Cause of death distribution of deaths in 2019.

Cause of death	Number of deaths	Percentage of all deaths
Respiratory	259	62.56
Transplantation	63	15.22
Non-CF related	23	5.56
Unknown	22	5.31
Other CF related	18	4.35
Liver-GI	15	3.62
Cancer	9	2.17
Suicide	4	0.97
Trauma	1	0.24
Total	414	100.00

Note: For the United Kingdom, all patients with a confirmed diagnosis of CF were included (N=10,665). The total number of patients presented is 49,925.
Note: Germany and the United Kingdom record Cause of death as “cardio/respiratory”.
Note: The Netherlands does not record “Cancer” and “Other-CF related” as cause of death.

The table shows cause of death for the deceased patients. The most frequent cause of death is respiratory disease.

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. More information on the data application process you will find on the website www.ecfs.eu/projects/ecfs-patient-registry/data-request-application.

In the period 2011 – 2021 we received 98 applications to use Registry data. The majority of these requests, 84%, originated from researchers from the European Cystic Fibrosis Society and other institutes, and 16% of the applications derived from Industry.

Several of these research projects have resulted in publications and other publications are in the pipeline. Articles using Registry data published in the period November 2020 to October 2021 are:

- Factors for severe outcomes following SARS-CoV-2 infection in people with cystic fibrosis in Europe. Jung A, Orenti A, Dunlevy F, et al. ERJ Open Research 2021; DOI: 10.1183/23120541.00411-2021. https://doi.org/10.1183/23120541.00411-2021

- Incidence of SARS-CoV-2 in people with cystic fibrosis in Europe between February and June 2020. Naehrlich L, Orenti A, Dunlevy F et al. J Cyst Fibros. 2021; April 17. www.cysticfibrosisjournal.com/article/S1569-1993(21)00099-0/fulltext#seccesectitle0022

- Survival estimates in European cystic fibrosis patients and the impact of socioeconomic factors: a retrospective cohort study. McKone EF, Ariti C, Jackson A, Zolin A, Carr SB, Orenti A, van Rens JG, Lemonnier L, Macek M Jr, Keogh RH, Naehrlich L, on behalf of the ECFSPR. European Respiratory Journal 2021 58 (3) 2002288; DOI 10.1183/13993003.02288-2020. https://erj.ersjournals.com/content/58/3/2002288

A complete overview of publications using ECFSPR data is available on www.ecfs.eu/projects/ecfs-patient-registry/articles.

The following abstracts were accepted in the period November 2020 to October 2021:

- Incidence of SARS-CoV-2 and risk factors for severe outcomes in people with cystic fibrosis in Europe. Orenti A, Dunlevy F, Zolin A, van Rens J, Jung A, Naehrlich L. Società Italiana Statistica Medica ed Epidemiologia Clinica, 15-17 September 2021.

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National Patient Organisations

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Appendix 1: List of contributing centres and national registries

List of individual centres and national registries that contributed to the ECFSPR. In larger print: 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 2019 data.

Country	Centre/National Registry name	Contact
Albania	1 individual centre:	Irena Kasmi
	"Mother Thereza" Hospital Center, Department of Paediatrics, Tirana	Irena Kasmi Evda Vevecka
Armenia	1 individual centre:	Satenik Harutyunyan
	Yerevan State Medical University, Muratsan University Hospital, Cystic Fibrosis Center, Yerevan	
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
	Medizinische Universität Innsbruck, Zertifiziertes CF Zentrum für Kinder, Jugendliche und Erwachsene, Innsbruck	Helmut Ellemunter Johannes Eder
	Klinikum Klagenfurt am Wörthersee, Abteilung für Kinder- und Jugendheilkunde, Pädiatrische Pulmologie/ Allergologie, Klagenfurt	Franz Hubert Wadlegger
	Kepler Universitätsklinikum, Universitätsklinik für Kinder- und Jugendheilkunde, Linz	Adrienne Molnar Julia Pichler Christina Thir
	Kepler Universitätsklinikum, Klinik für Lungenheilkunde/ Pneumologie, Linz	Martin Stadlinger Viktoria Reinelt Katrin Scheich
	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
	Landeskrankenhaus Steyr, Abteilung für Kinder- und Jugendheilkunde und Abteilung für Lungenheilkunde, Steyr	Alexander Ebner Margit Kalinger 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	Thomas Frischer Kerstin Tiringer Katharina Kainz
	Krankenhaus Hietzing, Abteilung für Atmungs- und Lungenerkrankungen, Vienna	Andrea Lakatos–Krepcik Dr. Sabine Burghart

Country	Centre/National Registry name	Contact
	Klinikum Wels-Grieskirchen, Abteilung für Kinder- und Jugendheilkunde, Wels	Franz Eitelberger Beatrix Wintersteiger Vera Karin Bauer
	Klinikum Wels-Grieskirchen, Abteilung für Lungenkrankheiten, Wels	Alexander Leitner Matthäus Ploder Thomas Tempelmayer Andrea Ringl
Belarus	Belarusian Republic Children's Center of Pulmonology and Cystic Fibrosis, Pulmonary Department, 3rd City Children's Clinical Hospital, Minsk	Vladimir Bobrovnichiy Svetlana Keegan
Belgium	Belgian Cystic Fibrosis Registry	Géraldine Daneau Simeon Wanyama
Bulgaria	2 individual centres:	Guergana Petrova
	Alexandrovska University Hospital, Pediatric Clinic, Sofia	Guergana Petrova
	University Hospital St. Marina, 2nd Paediatric Clinic, Varna	Miglena Georgieva Nataliya Dobrudzhanska Margarita Nikolova Ruzha Pancheva
Croatia	1 individual centre:	Duska Tjesic-Drinkovic Andrea Vukic Dugac
	University Hospital Centre Zagreb, Cystic Fibrosis Centre – Paediatrics and Adults, Zagreb	Duska Tjesić-Drinković Dorian Tjesić-Drinković Andrea Vukic Dugac
	On behalf of the Croatian CF Patient Database	Ivan Bambir Ivona Markelic
Cyprus	Medical School, University of Cyprus (children and adults)	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 Tania Pressler
France	Registre Français de la Mucoviscidose	Lydie Lemonnier Clémence Dehillotte
Germany	German Cystic Fibrosis Registry	Lutz Naehrlich Julia Wosniok
Greece	2 individual centres:	Elpis Hatziagorou
	Sismanoglio General Hospital of Attica, Adult Cystic Fibrosis Unit, Athens	Filia Diamantea Margarita Gkotsina
	Aristotle University of Thessaloniki, Cystic Fibrosis Centre, Thessaloniki	John Tsanakas Elpis Hatziagorou Aikaterini Manika Maria Sionidou Maria Fotoulaki
Georgia	I. Tsitsishvili Children's Clinic, CF Centre, Tblisi	Ia Khurtsilava Tsitsino Parulava

Country	Centre/National Registry name	Contact
Hungary	LaylaCystic Fibrosis Registry of Hungary	Andrea Párniczky Géza Marsal
Ireland	Cystic Fibrosis Registry of Ireland	Godfrey Fletcher Laura Kirwan
Israel	6 individual centres:	Meir Mei-Zahav
	Soroka Medical Centre, Ben Gurion University, Beer Sheva	Micha Aviram
	Carmel Medical Centre, Haifa	Galit Livnat
	Meyer Children's Hospital of Haifa, Rambam Medical Center, Haifa	Michal Gur
	Hadassah Medical Centre, Mount Scopus, Jerusalem	Malen Cohen-Cymberknoh
	Schneider Children's Medical Centre of Israel, Petach Tikvah	Meir Mei-Zahav
	Safra Children's Hospital, Sheba Medical Center, Ramat Gan	Ori Efrati
Italy	Italian Cystic Fibrosis Registry	Rita Padoan Marco Salvatore Annalisa Amato Gianluca Ferrari
Latvia	1 individual centre:	Elina Aleksejeva
	Rīga Stradinš 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
	Hospitalof Lithuanian University of Health Sciences Kauno Klinikos, Centre of Pediatric Chronic Respiratory Diseases, Kaunas	Valdone Misevičiene
Luxembourg	1 individual centre:	Anna-Maria Charatsi
	Centre Hospitalier de Luxembourg	Romain Nati Anna-Maria Charatsi Meriem Mastouri Flore Nzuangue
Rep. of North Macedonia	2 individual centres:	Stojka Fustik
	University Children's Hospital, Centre for Cystic Fibrosis, Skopje	Stojka Fustik Ana Stamatova
	Institute for respiratory diseases in children Kozle, Center for cystic fibrosis, Children and adults, Kozle	Tatjana Jakovska-Maretti Ivana Arnaudova Danevska
Rep. of Moldova	Outpatient Center for Cystic Fibrosis and Other Rare Diseases	Oxana Turcu
Netherlands	Dutch Cystic Fibrosis Registry	Vincent Gulmans Domenique Zomer
Norway	Norwegian Cystic Fibrosis Patient Registry	Egil Bakkeheim Anita Senstad Wathne

Country	Centre/National Registry name	Contact
Poland	12 individual centres:	Lukasz Wozniacki
	Voivodeship Children's Hospital, Dept. of Paediatric Pneumology and Allergology, Bydgoszcz	Radoslawa Staszak – Kowalska Mikolaj Kowalski
	Cystic Fibrosis Centre, Polanki Paediatric Hospital, Gdansk	Maria Trawinska-Bartnicka Ewa Sapiejka
	Centrum Medyczne Karpacz, Children/Adults' Hospital, Karpacz	Grzegorz Gaszczyk Monika Rams
	Dept of Pediatric Clinic John Paul II ,Upper Silesian Child Health Center, The independent Public Clinical Hospital no 6 of the Medical University of Silesian in Katowice	*Urszula Grzybowska-Chlebowczyk* *Bozena Kordys-Darmolinska*
	St. Louis Regional Specialised Children's Hospital, Krakow	Stanislaw Stepniewski Daria Dziecichowicz-Latala
	Wojewodzkie Wielospecjalistyczne centrum onkologii i traumatologii im. m. Kopernika w Lodzi. Ośrodek Pediatryczny im. Dr J. Korczak	*Iwona Stelmach* *Agnieszka Koniarek-Maniecka*
	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, Dept of Allergology and Cystic Fibrosis, St Jadwigi Krolowej in Rzeszow	Marta Rachel
	Szczecin Hospital "Zdroje" Dep. of Pediatrics, Allergology and Pulmonology	Pawel Gonerko Pawel Fabisiak
	Dziekanow Paediatric Hospital, Cystic Fibrosis Centre, Institute of Mother and Child, Warsaw	Dorota Sands Lukasz Wozniacki
Portugal	Cystic Fibrosis Registry of Portugal	Luísa Pereira
Romania	7 individual centres:	Liviu Pop
	Regional Cystic Fibrosis Centre Brasov, Brasov	Laura Larisa Dracea
	Children Emergency Hospital "Maria S. Curie", Bucharest	*Maria Brustan*
	Clinical Children's Hospital Grigore Alexandrescu, Bucharest	Simona Mosescu
	Mother & Child Health Institute, Bucharest	Iustina Stan
	Regional Cystic Fibrosis Centre Cluj, Cluj-Napoca	Radu Sorin Şerban Szabo Csilla-Enikő
	"S.F. Maria" Children's Emergency Hospital, Iasi	Dana Anton-Paduraru
	National Cystic Fibrosis Centre, Timişoara	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:	Milan Rodic
	National Centre for Cystic Fibrosis, Mother and Child Health Institute of Serbia "Dr Vukan Cupic", Belgrade	Predrag Minić Milan Rodić Aleksandar Sovtić
Slovakia	6 individual centres:	Hana Kayserova
	Childrens CF Centre, DFN Banská Bystrica, Banská Bystrica	Branko Takáč
	Centrum cystickej fibrozy pre dospelych FNSP FDR, Banská Bystrica	Eva Bérešova
	Centrum cystickej fibrozy pre dospelych, Klinika pneumologie l.SZU a Univerzitna nemocnica, Bratislava	Marta Hajkova
	Klinika detskej pneumologie SZU UN Bratislava, pracovisko Podunajské Biskupice, Bratislava	Hana Kayserova Nina Bližňáková
	CF Adult centre, University Hospital L Pasteura, Košice	Lenka Kopčová
	Centrum cystickej fibrozy detí, Detská fakultná nemocnica Košice, Košice	Anna Fetekeova Zuzana Hribíková
Slovenia	3 individual centres:	Uroš Krivec
	University Clinic of Pulmonary and Allergic Diseases, Golnik	Matjaž Fležar Tjaša Brus Pičman Julij Šelb
	University Medical Centre Ljubljana, University Children`s Hospital, Unit for pulmonary diseases	Uroš Krivec Jasna Rodman Berlot Majda Oštir
	University Medical Centre Ljubljana, Department of Pulmonology and Allergy	Izidor Kos Barbara Salobir
Spain	26 individual centres:	Mª Dolores Pastor Vivero
	Parc Taulí Hospital Universitario, Hospital de Sabadell, Unitat de Pneumologia Pediátrica i Unitat de Fibrosi Quística, Sabadell, Barcelona	Oscar Asensio de la Cruz Miguel Garcia Gonzàlez Xavier Pomares Amigó Concepción Montón Soler
	Hospital Sant Joan de Déu, Unitat de Pneumologia Pediàtrica i Fibrosi Quística, Barcelona	Maria Cols Roig Jordi Costa i Colomer
	Hospital Universitari Vall d'Hebron, Unidad de Fibrosis Quística del Adulto, Barcelona	Antonio Alvarez Fernández
	Hospital Vall d'Hebron, Unidad Fibrosis Quística y Neumología Pediátrica, Barcelona	Silvia Gartner
	Hospital Universitario de Cruces, Unidad de Fibrosis Quística, Bizkaia	Mª Dolores Pastor Vivero Félix Baranda García Ainhoa Gómez Bonilla Beatriz Gómez Crespo
	Hospital Universitario Reina Sofia, Dpto. Especialidades Médico-quirúrgicas, Área de Pediatría, Unidad de Alergia y Neumología Pediátricas, Unidad de Gestión Clínica de Pediatría y sus Especialidades, Cordoba	Javier Torres Borrego Noelia Sancho Montero
	Complejo Hospitalario Universitario Insular Materno Infantil, Las Palmas de Gran Canaria	Antonio José Aguilar Fernández
	Hospital Universitario La Paz, Unidad de Fibrosis Quìstica Adultos, Servicio de Neumología, Madrid	Concha Prados

Country	Centre/National Registry name	Contact
	Hospital Infantil La Paz, Sección de Neumología Pediátrica, Unidad de Fibrosis Quística Pediátrica, Madrid	María Isabel Barrio Gomez de Agüero Marta Ruiz de Valbuena Maiz Cristina de Manuel Gómez
	Hospital Universitario La Princesa, Neumología Adultos, Madrid	Rosa María Girón
	Hospital Niño Jesús, Sección de Neumología Pediátrica, Unidad de Fibrosis Quística, Madrid	José R. Villa Asensi Patricia Fernandez Garcia Alejandro López Neyra Verónica Sanz Santiago Rosa Ana Muñoz Codoceo
	Hospital Universitario de Ramón y Cajal, Unidad de Fibrosis Quística, Madrid	Luis Maiz Carro Rosa Maria Nieto Royo Adelaida Lamas Ferreiro Saioa Vicente Santamaria
	Hospital Universitario 12 de Octubre, Unidad de Fibrosis Quística Adultos, Madrid	Layla Diab Cáceres
	Hospital Universitario 12 de Octubre, Unidad de Fibrosis Quística Pediátrica, Madrid	Carmen Luna Paredes Enrique Salcedo Lobato
	Hospital Regional Universitario de Málaga, Unidad Fibrosis Quística Adultos, Málaga	Casilda Olveira Fuster Gabriel María Olveira Fuster Nuria Porras Pèrez
	Hospital Regional Universitario de Málaga, Unidad de Fibrosis Quística Pediátrica, Málaga	Francisco Javier Pèrez Frias Estela Pèrez-Ruiz Pilar Caro-Aguilera
	Hospital Clínico Universitario Virgen de la Arrixaca, Unidad de Fibrosis Quística, Murcia	Pedro Mondéjar-López
	Hospital Universitario Central de Asturias, Unidad de Fibrosis Quística, Oviedo	José Ramón Gutiérrez Martínez Marta Garcia Clemente
	Hospital Universitari 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 Margalida Barceló Bobillo
	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 Callejon Orlando Mesa
	Hospital Clínico Universitario de Valencia, Unidad de Fibrosis Quística Pediátrica, Valencia	Amparo Escribano 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, Vigo	*Cristina Ramos Hernández* *María Jesús Rodriguez*
	Hospital Universitario Miguel Servet, Unidad de Neumología Pediátrica y Fibrosis Quística, Zaragoza	Carlos Martín de Vicente
	Hospital Universitario Miguel Servet, Unidad de Neumología y Fibrosis Quística (Adultos), Zaragoza	Maria Inés Herrero Labarga
Sweden	Cystic Fibrosis Registry of Sweden	Isabelle de Monestrol Anders Lindblad

Country	Centre/National Registry name	Contact
Switzerland	20 individual centres:	Andreas Jung
	Kantonsspital Aarau AG, Klinik für Kinder und Jugendliche, Abteilung pädiatrische Pneumologie, Allergologie und Immunologie, Aarau	Dominik Müller-Suter Peter Eng Rachel Kusche
	Kantonsspital Aarau AG, Klinik für Pneumologie und Schlafmedizin, Aarau	Sarosh Irani G. Mauro Tini Lydia Eisenmann
	Universitätsspital Basel, Klinik für Pneumologie, Adulte Cystische Fibrose, Basel	Michael Tamm Kathleen Jahn
	UKBB Universitäts-Kinderspital beider Basel, Abteilung Intensivmedizin & Pneumologie, Basel	Jürg Hammer Daniel Trachsel Anja Jochmann Diana Reppucci
	Inselspital, Universitätsklinik für Pneumologie, Abteiling Cystische Fibrose, Bern	Thomas Geiser Dagmar Lin Michaela Semmler
	Lindenhofspital Quartier Bleu, Bern	Reta Fischer Iris Schmid Bernhard Schwizer Patrizia Bevilacqua
	Universitätsklinik für Kinderheilkunde, Zentrum für Cystische Fibrose und Pulmonologie, Inselspital, Bern	Philipp Latzin Romy Rodriguez Florian Singer
	Hôpital Cantonal Fribourg, Pädiatrie, Fribourg	Denise Herzog Johannes Wildhaber
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	Hôpitaux Universitaires de Genève, Département de Médecine, Service de Pneumologie, Consultation de Mucoviscidose Adulte, Genève	Paola Gasche Jérôme Plojoux Valerie Durand
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	Children's Hospital of Eastern Switzerland, Division of Paediatric Pulmonology & CF Centre, St Gallen	Jürg Barben Christine Baumgartner

Country	Centre/National Registry name	Contact
	Kantonsspital St. Gallen, Lungenzentrum, Zentrum für Cystische Fibrose für Erwachsene, St. Gallen	Martin Brutsche Otto Schoch Anna-Lena Walter
	Kantonsspital Winterthur, Klinik für Pneumologie und Klinik für Innere Medizin, Adulte Cystische Fibrose, Winterthur	Markus Hofer Sieghart Filippi
	Universitäts-Kinderspital Zürich, Abteilung für Pneumologie, Zürich	Andreas Jung Alexander Möller Demet Inci
	Universitätsspital Zürich, Klinik für Pneumologie, Adultes CF Zentrum, Zürich	Christian Benden Thomas Kurowski
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	Medeniyet University, Faculty of Medicine, Division of Pediatric Pulmonology, Istanbul	Saniye Girit Yetkin Ayhan
Ukraine	1 individual centre:	Halyna Makukh
	Cystic Fibrosis Centre of Western Ukrainian Specialized Children's Medical Centre, Lviv	Lyudmyla Bober Halyna Makukh
United Kingdom	UK Cystic Fibrosis Registry	Rebecca Cosgriff Susan Charman Elaine Gunn Siobhán Carr Sarah Clarke

Appendix 2: Technical notes

Patient inclusion criteria

The ECFSPR registers patients diagnosed with CF in accordance with agreed definitions (see Appendix 2). Data of patients with a diagnosis that does not meet the agreed definitions are accepted in the database but not included in the analyses.

Data manipulation

To ensure that data was anonymous, we collected only year and month of birth and the day of birth was set to the 15th of the month.
For pre-natal 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.

Reference populations used for computing z-scores

The value of a z-score depends on the anthropometric reference chart: if different reference values are used, the same value of height (or weight or BMI) will result in different values of z-scores, and these differences might be of clinical importance. In order to compare the nutritional status of CF patients with that of healthy individuals an appropriate reference population must be used: ideally, a fair comparison requires that CF patients and healthy individuals belong to the same population. This implies the availability of a national reference.

The lack of a national reference for most countries participating in the ECFSPR obliged us to use an international reference to compute z-scores for height, weight and BMI. We decided to use the CDC 2000 reference charts (Kuczmarski RJ, Ogden CL, Guo SS et al. 2000 CDC Growth Charts for the United States: Methods and Development. National Centre for Health Statistics. Vital Health Stat 2002; 11(246):1-190.), which were derived from samples of healthy individuals from the USA[1]. The choice of CDC charts as a reference, although not necessarily the most suitable to assess the nutritional status of European CF patients, is justified by the widespread use of these charts at international level.

Reference populations used for computing FEV_1 predicted values

We computed the percent of predicted values for FEV_1 and FVC using:
The 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.

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.

[1] For details on the target population, please see www.cdc.gov/growthcharts/2000growthchart-us.pdf.

Appendix 3: List of variables, inclusion criteria and definitions used by the ECFSPR

Variables

Demographics

CF centre code
Patient code
Year of follow-up
Date of birth (year and month)
Gender
Status of patient
Cause of death
Date of death

Diagnosis

Diagnosis confirmed
Age at diagnosis
Sweat test type and value
Electrolytes
Chloride value
Meconium Ileus
Nasal Potential Difference (NPD)
CF-typical NPD
Date of NPD
Intestinal current measurement (ICM)
CF-typical ICM
Date of ICM
Neonatal screening

Genotype

First mutation
Second mutation

Therapy

Inhaled continuous hypertonic NaCl this year
Inhaled continuous Mannitol
Inhaled continuous antibiotic this year
Inhaled continuous bronchodilators this year
In Oxygen therapy this year
Use of Non-invasive positive pressure ventilation (NIPPV)
Use of rhDNase this year
Use of continuous Inhaled steroids
Use of continuous Oral steroids
Use of continuous azithromycin (or other macrolide) this year
Use of ursodeoxycholic acid this year
Use of pancreatic enzymes this year
Use of proton pump inhibitors (PPI)
Use of CFTR Modifier Therapy

Complications

Allergic broncho-pulmonary aspergillosis this year
Diabetes treated this year
Pneumothorax this year
Distal intestinal obstruction syndrome (DIOS)
Salt depletion this year
Liver disease this year
Haemoptysis major over 250 ml this year
Pancreatic status: faecal elastase
Pancreatic status: faecal fat
Occurrence of malignancy this year

Microbiology

Chronic *Pseudomonas aeruginosa*
Chronic *Staphylococcus aureus*
Chronic *Burkholderia cepacia complex*
Stenotrophomonas maltophilia this year
Nontuberculous mycobacteria this year
Achromobacter spp this year
Haemophilus influenza this year
MRSA this year
Total days on iv antibiotics at home and in hospital this year
Total days on iv antibiotics in hospital this year
Total days in hospital this year

Follow-up

Date of best FEV_{1*} recorded this year
Value of best FEV_{1**} recorded this year
Value of best FVC recorded this year
Date of lowest LCI 2.5% this year
Value of lowest LCI 2.5% this year
Type of device
Height measured at date of best FEV_1 (or in case of no FEV_1 last height of the year)
Weight measured at date of best FEV_1 (or in case of no FEV_1 last height of the year)

Transplant

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

*FEV1 of highest FEV1% predicted
**FVC at time of best FEV1

Inclusion criteria

Only patients who fulfil the diagnostic criteria below should be included in the Registry:

1. **Two sweat tests value > 59 mmol/L chloride**: CF diagnosis accepted.
2. **One sweat test value > 59 mmol/L chloride and DNA Analysis/Genotyping – two identified disease causing CF mutations**: CF diagnosis accepted.
3. **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:
   a. DNA Analysis/Genotyping: two identified disease causing CF mutations;
   b. Transepithelial (Nasal) Potential Difference or Intestinal current measurement: result consistent with a diagnosis of CF;
   c. Clinical Presentation: typical features of CF.
4. **Diagnosis reversal**:
   If the patient's CF diagnosis was reversed during the year, must be due to one of the options listed:
   a. DNA Analysis: unable to identify two disease causing CF mutations;
   b. Transepithelial (Nasal) Potential Difference and/or Intestinal current measurement: result not consistent with a diagnosis of CF;
   c. Repeated normal values from sweat tests and confirmed by the clinical team.

Definitions used by the EFCSPR

SWEAT TEST

1. Diagnostic standards: the quantity of sweat should indicate an adequate rate of sweat production.

2. a. The sweat sample should be processed immediately after sweat test collection;
   b. Chloride concentration measurement is the preferred analysis;
   c. Chloride value: the Chloride value should be measured in millimols per litre (mmol/L);
   if duplicate tests were completed on the same day, **the highest positive value** should be considered;
   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 value of below 30 mmol/L).

   Note: The acceptable range for Chloride values is 1-160 mmol/L. Anyone who has a Chloride value above 160 mmol/L must be re-tested.

3. The ECFSPR considers only Titration/Chloride values in the analyses.

References:

- ECFS Standards of Care Best Practices Guidelines, Smyth AR et al. J Cyst. Fibros. 13 (2014) S23-S42.
- Standards of Care for Cystic Fibrosis ten years later. Castellani C, Conway S, Smyth AR, Stern M, Elborn JS. J Cyst. Fibros. 13 (2014) S1-S2.

NUTRITION

Measurements: Weight and height should be measured in accordance with the EuroCareCF guidelines:

a. weight: removal of outer clothing, shoes and socks;
b. height: without shoes and socks; stadiometer: top of head in contact with head board, slight pressure;
c. date: the recorded height and weight should be the measurements taken the same day as the best FEV1 (FEV1 of the highest FEV1% predicted). If spirometry was not done the last weight and height measurements of the year, and the date they were measured, should be recorded.

Note: z-scores for height, weight and BMI are calculated using the CDC reference values (Kuczmarski et al, 2002).

References:

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

SPIROMETRY

The ECFS Patient Registry collects data on spirometry values in order to obtain standardised data for comparison with other centres/countries and for use in epidemiological studies. Some of the conditions for this (see below) may not be met at every clinical visit for all patients, and, for the ECFSPR, only spirometry tests fulfilling the criteria must be recorded by centres, or extracted by the National Registries.

All spirometry tests must be carried out in accordance with the ATS/ERS guidelines: www.thoracic.org/statements/resources/pfet/PFT2.pdf.

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

1. Pre-test:
    a. date of birth, gender and height must be recorded for calculation of predicted values;
    b. all recorded spirometry tests must be pre-bronchodilator* values
        i. short-acting bronchodilators: at least 4 hours pre-test;
        ii. long-acting bronchodilators: at least 12 hours pre-test.
2. Values to report:
    a. FEV1 value to report: value of FEV1, in litres (up to 2 decimals), of the highest FEV1% predicted of the year, in accordance with local reference values;
    b. The FEV1 and FVC measurements must be reported in litres (L), to max 2 decimal points;
    c. The FVC measurement is the FVC from the same test as the recorded FEV1 and it must be greater than or equal to the FEV1 measurement;
    d. For the reported spirometry value, the date of the test and the patient's height and weight at that date must also be recorded so that the percentage of predicted values can be calculated;
    e. Only tests deemed valid according to ATS/ERS guidelines to be reported.
3. Calculation of percent of predicted values:
    A common set of reference values is used: Global Lung Function Initiative equations described by Quanjer PH et al.

    Note: The ECFSPR Definitions Group considered the issue of race-specific reference values. The decision was to not record race for European patients and therefore not to calculate race-specific values.

References:

- 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.
- Standardisation of spirometry. Miller et al. Eur Respir J 2005; 26: 319–338.
- General considerations for lung function testing. Miller et al. Eur Respir J 2005; 26: 153–161.
- Cystic Fibrosis Foundation Patient Registry User's Guide, Version 4.0. 2006.
- Task Force to Evaluate Choice of Spirometric Reference Equations for the National Patient Registry: Summary and Recommendations. Rosenfeld et al. Cystic Fibrosis Foundation Registry Committee, 2005.

CHRONIC INFECTION IN THE LOWER AIRWAYS

1. 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 anti-pseudomonas antibodies according to local laboratories.

2. Chronic infection with other gram-negative bacteria should meet the same criteria as described above.

References:

- Evaluation of a new definition for chronic Pseudomonas aeruginosa in cystic fibrosis patients. Lee TWR, Brownlee KG, Conway SP, Denton M, Littlewood JM. J Cyst Fibros. 2003 Mar;2(1):29-34.
- Evaluating the "Leeds criteria" for Pseudomonas aeruginosa infection in a cystic fibrosis centre. Proesmans M, Balinska-Miskiewiscz, Dupont L et al. Eur Resp J 2006;27:937-943.
- Antibiotic therapy against Pseudomonas aeruginosa in cystic fibrosis: a European consensus. Döring G, Conway SP, Heijerman HG, et al. Eur Respir J 2000;16:749-767.

ALLERGIC BRONCHOPULMONARY ASPERGILLOSIS (ABPA)

Diagnostic criteria:

1. Acute or subacute clinical deterioration (cough, wheeze, exercise intolerance, exercise-induced asthma, change in pulmonary function, or increased sputum production) not attributable to another etiology.
2. Total IgE > 500 IU/ml.
3. Positive skin prick test for Aspergillus antigen (> 3 mm) or positive specific IgE for *A. fumigatus*.
4. Either:
   a. precipitins to *A. fumigatus* or in vitro demonstration of IgG antibody to *A. fumigatus*;
   b. or new or recent abnormalities on chest radiography (infiltrates or mucus plugging) or chest CT (characteristic changes) that have not cleared with antibiotics and standard physiotherapy.

References:

Allergic bronchopulmonary aspergillosis in cystic fibrosis--state of the art: Cystic Fibrosis Foundation Consensus Conference. Stevens DA, Moss RB, Kurup VP, Knutsen AP, Greenberger P, Judson MA, Denning DW, Crameri R, Brody AS, Light M, Skov M, Maish W, Mastella G; Participants in the Cystic Fibrosis Foundation Consensus Conference. Clin Infect Dis. 2003 Oct 1;37 Suppl 3:S225-64.

LIVER DISEASE

The ECFSPR has adopted the definitions for Liver Disease used by the Cystic Fibrosis Registry in the United Kingdom.

These definitions discriminate patients 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.

PANCREATIC STATUS

To define pancreatic insufficiency two determinations are mandatory:

- Young children: Stool fat (van de Kamer) > 4-5 g/d;
- Children older than 10 years and adults: Stool fat (van de Kamer) >7g/d and/or faecal pancreatic elastase-1 < 200 ug/g.

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

For the ECFSPR, pancreatic status is 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).

*see definition above.

References:

- Nutrition in patients with cystic fibrosis. A European consensus. Sinaasappel M, Stern M, Littlewood J, Wolfe S, Steinkamp G, Heijerman HGM, Robberecht E, Döring G. Nutrition J Cystic Fibros.K 2002; 1:51-75.
- Invited review: Indirect pancreatic function tests in children. Walkowiak J, Nousia-Arvanitakis S, Henker J, Stern M, Sinaasappel M, Dodge JA. J Pediatr Gastroenterol Nutr 2005; 40:107-114.
- Metabolic alkalosis with hypoelectrolytemia in infants with cystic fibrosis. Fustik S, Pop-Jordanova N, Slaveska N, Koceva S, Efremov G. Pediatr int 2002; 44: 289-92.

SALT LOSS SYNDROME

Primary metabolic alkalosis with blood pH > 7.45, serum sodium < 130 mmol/l and serum chloride < 90 mmol/l.

References:

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.

TRANSPLANTATION

1. For patients who had a transplant during the year of follow up:
   The best FEV1 (of the highest FEV1% predicted) before transplantation must be used;
   Therapy, complications and microbiology from before transplantation must be recorded.
2. For patients who had a transplant before the current follow-up year, record all information available.