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Published: December 2021
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Suggested citation for this report:

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 ECFSPR 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 organisations on 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 value for a given complication has this complication or not, meaning given frequencies are less accurate. For example, in a country where 7% of the people with CF have liver disease but 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
SL: 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 CF both 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<sub>1</sub>: the Forced Expiratory Volume of air in the first second of a forced exhaled breath.

FEV<sub>1</sub>%: the FEV<sub>1</sub> 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 were diagnosed after 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

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

* 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.

<table>
<thead>
<tr>
<th>Country</th>
<th>Patients registered, not lost to follow-up</th>
<th>Patients seen</th>
<th>Estimated coverage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Albania</td>
<td>133</td>
<td>106</td>
<td>80%</td>
</tr>
<tr>
<td>Armenia</td>
<td>30</td>
<td>25</td>
<td>&gt;70%</td>
</tr>
<tr>
<td>Austria</td>
<td>833</td>
<td>816</td>
<td>&gt;90%</td>
</tr>
<tr>
<td>Belarus</td>
<td>155</td>
<td>155</td>
<td>90%</td>
</tr>
<tr>
<td>Belgium*</td>
<td>1360</td>
<td>1294</td>
<td>&gt;90%</td>
</tr>
<tr>
<td>Bulgaria</td>
<td>197</td>
<td>188</td>
<td>&gt;70%</td>
</tr>
<tr>
<td>Croatia**</td>
<td>146</td>
<td>139</td>
<td>&gt;95%</td>
</tr>
<tr>
<td>Cyprus</td>
<td>31</td>
<td>28</td>
<td>&gt;80%</td>
</tr>
<tr>
<td>Czech Republic*</td>
<td>654</td>
<td>643</td>
<td>99%</td>
</tr>
<tr>
<td>Denmark*</td>
<td>530</td>
<td>514</td>
<td>99%</td>
</tr>
<tr>
<td>France*</td>
<td>6729</td>
<td>6729</td>
<td>&gt;90%</td>
</tr>
<tr>
<td>Georgia1</td>
<td>80</td>
<td>72</td>
<td>&gt;80%</td>
</tr>
<tr>
<td>Germany*</td>
<td>6481</td>
<td>6481</td>
<td>80%</td>
</tr>
<tr>
<td>Greece2</td>
<td>451</td>
<td>436</td>
<td>&gt;70%</td>
</tr>
<tr>
<td>Hungary*</td>
<td>528</td>
<td>509</td>
<td>90%</td>
</tr>
<tr>
<td>Ireland*</td>
<td>1268</td>
<td>1251</td>
<td>90%</td>
</tr>
<tr>
<td>Israel**</td>
<td>588</td>
<td>529</td>
<td>&gt;95%</td>
</tr>
<tr>
<td>Italy*</td>
<td>5618</td>
<td>5585</td>
<td>95%</td>
</tr>
<tr>
<td>Latvia</td>
<td>47</td>
<td>45</td>
<td>&gt;90%</td>
</tr>
<tr>
<td>Lithuania</td>
<td>32</td>
<td>28</td>
<td>52%</td>
</tr>
<tr>
<td>Luxembourg</td>
<td>40</td>
<td>38</td>
<td>&gt;95%</td>
</tr>
<tr>
<td>Rep of Moldova</td>
<td>59</td>
<td>53</td>
<td>&gt;90%</td>
</tr>
<tr>
<td>The Netherlands*</td>
<td>1533</td>
<td>1501</td>
<td>95%</td>
</tr>
<tr>
<td>North Macedonia</td>
<td>141</td>
<td>131</td>
<td>&gt;90%</td>
</tr>
<tr>
<td>Norway*</td>
<td>319</td>
<td>312</td>
<td>90%</td>
</tr>
<tr>
<td>Poland</td>
<td>1242</td>
<td>1194</td>
<td>&gt;60%</td>
</tr>
<tr>
<td>Portugal**</td>
<td>368</td>
<td>347</td>
<td>&gt;95%</td>
</tr>
<tr>
<td>Romania</td>
<td>246</td>
<td>230</td>
<td>50%</td>
</tr>
<tr>
<td>Russian Federation*</td>
<td>3433</td>
<td>3162</td>
<td>&gt;95%</td>
</tr>
<tr>
<td>Serbia</td>
<td>202</td>
<td>183</td>
<td>&gt;90%</td>
</tr>
<tr>
<td>Slovak Republic**</td>
<td>326</td>
<td>287</td>
<td>&gt;90%</td>
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<tr>
<td>Slovenia</td>
<td>113</td>
<td>113</td>
<td>&gt;95%</td>
</tr>
<tr>
<td>Spain</td>
<td>2417</td>
<td>2329</td>
<td>80%</td>
</tr>
<tr>
<td>Sweden*</td>
<td>704</td>
<td>704</td>
<td>95%</td>
</tr>
<tr>
<td>Switzerland**</td>
<td>999</td>
<td>974</td>
<td>&gt;95%</td>
</tr>
<tr>
<td>Turkey</td>
<td>2002</td>
<td>1988</td>
<td>&gt;60%</td>
</tr>
<tr>
<td>Ukraine</td>
<td>212</td>
<td>151</td>
<td>23%</td>
</tr>
<tr>
<td>United Kingdom**</td>
<td>10655</td>
<td>10070</td>
<td>99%</td>
</tr>
<tr>
<td>Total</td>
<td>50902</td>
<td>49340</td>
<td></td>
</tr>
</tbody>
</table>

* 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](http://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.

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

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

<table>
<thead>
<tr>
<th>Country</th>
<th>Children (&lt;18 years)</th>
<th>Adults (≥18 years)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>number (%)</td>
<td>number (%)</td>
</tr>
<tr>
<td>Norway</td>
<td>114 (35.85)</td>
<td>204 (64.15)</td>
</tr>
<tr>
<td>Poland</td>
<td>863 (70.11)</td>
<td>368 (29.89)</td>
</tr>
<tr>
<td>Portugal</td>
<td>191 (52.47)</td>
<td>173 (47.53)</td>
</tr>
<tr>
<td>Romania</td>
<td>229 (94.63)</td>
<td>13 (5.37)</td>
</tr>
<tr>
<td>Russian Federation</td>
<td>2526 (74.71)</td>
<td>855 (25.29)</td>
</tr>
<tr>
<td>Serbia</td>
<td>134 (66.67)</td>
<td>67 (33.33)</td>
</tr>
<tr>
<td>Slovak Republic</td>
<td>134 (41.74)</td>
<td>187 (58.26)</td>
</tr>
<tr>
<td>Slovenia</td>
<td>57 (50.44)</td>
<td>56 (49.56)</td>
</tr>
<tr>
<td>Spain</td>
<td>1112 (46.26)</td>
<td>1292 (53.74)</td>
</tr>
<tr>
<td>Sweden</td>
<td>264 (37.61)</td>
<td>438 (62.39)</td>
</tr>
<tr>
<td>Switzerland</td>
<td>427 (42.91)</td>
<td>568 (57.09)</td>
</tr>
<tr>
<td>Turkey</td>
<td>1752 (87.95)</td>
<td>240 (12.05)</td>
</tr>
<tr>
<td>Ukraine</td>
<td>169 (80.48)</td>
<td>41 (19.52)</td>
</tr>
<tr>
<td>United Kingdom</td>
<td>4515 (42.83)</td>
<td>6026 (57.17)</td>
</tr>
<tr>
<td>Total</td>
<td>24041 (47.62)</td>
<td>26447 (52.38)</td>
</tr>
</tbody>
</table>
**Figure 1.5 Proportion of children (<18 years) and adults (≥18 years), by country and overall. Patients alive on 31/12/2019.**

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.

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

<table>
<thead>
<tr>
<th>Country</th>
<th>N</th>
<th>Mean (average age)</th>
<th>Min (age of the youngest patient)</th>
<th>25th pctl (25% of the patients are younger than this age)</th>
<th>Median (half the patients are younger than this age)</th>
<th>75th pctl (75% of the patients are younger than this age)</th>
<th>Max (age of the oldest patient)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Albania</td>
<td>133</td>
<td>10.5</td>
<td>0.5</td>
<td>6.0</td>
<td>9.2</td>
<td>14.4</td>
<td>28.0</td>
</tr>
<tr>
<td>Armenia</td>
<td>30</td>
<td>11.1</td>
<td>2.2</td>
<td>5.9</td>
<td>9.2</td>
<td>14.8</td>
<td>34.2</td>
</tr>
<tr>
<td>Austria</td>
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<td>20.8</td>
<td>0.3</td>
<td>10.5</td>
<td>20.0</td>
<td>29.6</td>
<td>82.7</td>
</tr>
<tr>
<td>Belarus</td>
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<td>0.3</td>
<td>5.0</td>
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<td>12.8</td>
<td>19.7</td>
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<tr>
<td>Belgium</td>
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<td>13.6</td>
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<td>36.4</td>
<td>78.5</td>
</tr>
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<td>24.0</td>
<td>65.5</td>
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<td>7.8</td>
<td>14.0</td>
<td>22.0</td>
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<td>Cyprus</td>
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<td>27.7</td>
<td>49.5</td>
</tr>
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<td>0.1</td>
<td>8.9</td>
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<td>11.9</td>
<td>21.5</td>
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<td>84.6</td>
</tr>
<tr>
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<td>3.7</td>
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<td>9.3</td>
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<td>28.0</td>
<td>70.9</td>
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<td>85.4</td>
</tr>
<tr>
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<td>82.5</td>
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<td>5.8</td>
<td>12.4</td>
<td>20.5</td>
<td>34.7</td>
</tr>
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<td>13.3</td>
<td>20.9</td>
<td>28.8</td>
<td>35.5</td>
</tr>
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<td>55.5</td>
</tr>
<tr>
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<td>1.3</td>
<td>5.5</td>
<td>9.8</td>
<td>14.5</td>
<td>33.7</td>
</tr>
<tr>
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<td>12.7</td>
<td>23.3</td>
<td>34.1</td>
<td>78.5</td>
</tr>
<tr>
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<td>5.0</td>
<td>12.0</td>
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<td>19.5</td>
<td>56.7</td>
</tr>
<tr>
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</tr>
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<td>9.3</td>
<td>13.8</td>
<td>29.0</td>
</tr>
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<td>0.0</td>
<td>5.7</td>
<td>10.8</td>
<td>18.1</td>
<td>62.0</td>
</tr>
<tr>
<td>Serbia</td>
<td>201</td>
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<td>0.1</td>
<td>7.6</td>
<td>13.5</td>
<td>20.5</td>
<td>46.1</td>
</tr>
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<td>11.0</td>
<td>20.9</td>
<td>29.5</td>
<td>80.0</td>
</tr>
<tr>
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<td>1.0</td>
<td>9.0</td>
<td>18.0</td>
<td>26.3</td>
<td>64.1</td>
</tr>
<tr>
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<td>2404</td>
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<td>0.1</td>
<td>10.1</td>
<td>19.8</td>
<td>32.5</td>
<td>84.2</td>
</tr>
<tr>
<td>Sweden</td>
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<td>0.2</td>
<td>12.3</td>
<td>24.5</td>
<td>37.2</td>
<td>77.5</td>
</tr>
<tr>
<td>Switzerland</td>
<td>995</td>
<td>22.8</td>
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<td>10.0</td>
<td>20.9</td>
<td>31.9</td>
<td>85.3</td>
</tr>
<tr>
<td>Turkey</td>
<td>1992</td>
<td>9.7</td>
<td>0.1</td>
<td>4.1</td>
<td>8.0</td>
<td>13.9</td>
<td>44.8</td>
</tr>
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<td>Ukraine</td>
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<td>1.0</td>
<td>6.4</td>
<td>10.9</td>
<td>16.9</td>
<td>39.7</td>
</tr>
<tr>
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<td>0.0</td>
<td>10.3</td>
<td>21.2</td>
<td>32.5</td>
<td>85.4</td>
</tr>
<tr>
<td>Total</td>
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<td>0.0</td>
<td>9.4</td>
<td>19.0</td>
<td>30.9</td>
<td>85.4</td>
</tr>
</tbody>
</table>

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.**

<table>
<thead>
<tr>
<th>Country</th>
<th>N</th>
<th>N miss</th>
<th>Mean (average age at diagnosis)</th>
<th>Min (lowest age at diagnosis)</th>
<th>25th pctl (25 % of the patients were diagnosed before this age)</th>
<th>Median (half the patients were diagnosed before this age)</th>
<th>75th pctl (75 % of the patients were diagnosed before this age)</th>
<th>Max (highest age at diagnosis)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Albania</td>
<td>106</td>
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<td>0.75</td>
<td>0.00</td>
<td>0.16</td>
<td>0.25</td>
<td>0.41</td>
<td>16.00</td>
</tr>
<tr>
<td>Armenia</td>
<td>25</td>
<td>0</td>
<td>1.72</td>
<td>0.10</td>
<td>0.50</td>
<td>1.00</td>
<td>2.00</td>
<td>6.17</td>
</tr>
<tr>
<td>Austria</td>
<td>751</td>
<td>65</td>
<td>2.20</td>
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<td>0.10</td>
<td>0.20</td>
<td>0.60</td>
<td>61.00</td>
</tr>
<tr>
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<td>155</td>
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<td>0.01</td>
<td>0.05</td>
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<td>5.01</td>
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<td>0.40</td>
<td>3.16</td>
<td>75.53</td>
</tr>
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<td>0.10</td>
<td>0.31</td>
<td>2.70</td>
<td>81.90</td>
</tr>
</tbody>
</table>

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.**

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.
The patient with the highest age at diagnosis was diagnosed when he/she was 75 years old.

The mean age at diagnosis is 3.6 years (patients on average are diagnosed at 3.6 years).

Half of the patients were diagnosed before 0.5 years (and half of the patients were diagnosed after 0.5 years).

The patient with the lowest age at diagnosis was diagnosed at birth.

Half of the patients were diagnosed between 0.1 years (36 days) and 3 years.

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.

<table>
<thead>
<tr>
<th>Country</th>
<th>Genotyping not done</th>
<th>Genotyping done</th>
<th>Among genotyping done at least one mutation unknown number (%)</th>
<th>Among genotyping done two mutations identified number (%)</th>
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<tbody>
<tr>
<td></td>
<td>number (%)</td>
<td>number (%)</td>
<td></td>
<td></td>
</tr>
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<td>10 (7.19)</td>
<td>129 (92.81)</td>
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<td>2 (7.14)</td>
<td>26 (92.86)</td>
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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”).

<table>
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<th>Genotyping done</th>
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<th>Two mutations identified</th>
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<td>number (%)</td>
<td>at least one mutation unknown number (%)</td>
<td>number (%)</td>
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<td>46293 (94.52)</td>
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</table>
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.**

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

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

FEV₁ is measured in litres but it is normally expressed as a percentage of the expected (i.e. predicted) value (FEV₁%). 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 FEV₁% 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 FEV₁, 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 FEV₁% values only for patients aged 6 years or older.

We asked the countries to report the best FEV₁ recorded throughout the year (relative to the best FEV₁% computed at the CF centres) to the ECFSPR.

We excluded patients from the analyses on FEV₁ 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.

<table>
<thead>
<tr>
<th>Country</th>
<th>N</th>
<th>N Miss</th>
<th>Mean (average FEV(_1),%)</th>
<th>Min</th>
<th>25(^{th}) pctl (25% of patients have FEV(_1),% below this value)</th>
<th>Median (50% of patients have FEV(_1),% below this value)</th>
<th>75(^{th}) pctl (75% of patients have FEV(_1),% below this value)</th>
<th>Max</th>
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<td>93.8</td>
<td>97.8</td>
<td>101.3</td>
<td>116.2</td>
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</tr>
</tbody>
</table>

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 FEV\(_1\) 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 FEV\(_1\) could be from the previous calendar year.

This table shows some descriptive statistics for FEV\(_1\) 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.

<table>
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<tr>
<th>Country</th>
<th>N</th>
<th>N Miss</th>
<th>Mean (average FEV$_1$%)</th>
<th>Min</th>
<th>25$^{th}$ pctl (25% of patients have FEV$_1$% below this value)</th>
<th>Median (50% of patients have FEV$_1$% below this value)</th>
<th>75$^{th}$ pctl (75% of patients have FEV$_1$% below this value)</th>
<th>Max</th>
</tr>
</thead>
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<td>27.9</td>
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<td>90.0</td>
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<td>70.2</td>
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<td>16.5</td>
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<td>85.4</td>
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<td>3</td>
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<td>40.3</td>
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<td>93.1</td>
<td>110.2</td>
</tr>
<tr>
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<td>47</td>
<td>68.0</td>
<td>10.5</td>
<td>49.5</td>
<td>69.2</td>
<td>86.2</td>
<td>158.5</td>
</tr>
<tr>
<td>Total</td>
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<td>594</td>
<td>68.8</td>
<td>10.5</td>
<td>49.3</td>
<td>69.7</td>
<td>87.8</td>
<td>158.5</td>
</tr>
</tbody>
</table>

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 FEV₁ % 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 FEV₁ 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 FEV₁ 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.

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.

<table>
<thead>
<tr>
<th>Age at FEV$_1$ measurement</th>
<th>N</th>
<th>N Miss</th>
<th>Mean</th>
<th>Min</th>
<th>25$^{th}$ pctl</th>
<th>Median</th>
<th>75$^{th}$ pctl</th>
<th>Max</th>
</tr>
</thead>
<tbody>
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<td>6-9</td>
<td>5051</td>
<td>624</td>
<td>96.9</td>
<td>18.1</td>
<td>88.0</td>
<td>98.2</td>
<td>107.6</td>
<td>185.1</td>
</tr>
<tr>
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<td>6235</td>
<td>371</td>
<td>89.0</td>
<td>13.6</td>
<td>80.1</td>
<td>91.5</td>
<td>101.1</td>
<td>157.5</td>
</tr>
<tr>
<td>15-19</td>
<td>5537</td>
<td>225</td>
<td>81.3</td>
<td>13.1</td>
<td>68.3</td>
<td>84.9</td>
<td>96.7</td>
<td>158.5</td>
</tr>
<tr>
<td>20-24</td>
<td>4825</td>
<td>149</td>
<td>73.2</td>
<td>10.5</td>
<td>56.1</td>
<td>75.8</td>
<td>91.1</td>
<td>142.4</td>
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<tr>
<td>25-29</td>
<td>4147</td>
<td>122</td>
<td>69.2</td>
<td>11.4</td>
<td>50.6</td>
<td>70.1</td>
<td>87.5</td>
<td>144.8</td>
</tr>
<tr>
<td>30-34</td>
<td>3298</td>
<td>92</td>
<td>65.3</td>
<td>13.8</td>
<td>46.3</td>
<td>64.8</td>
<td>83.2</td>
<td>136.3</td>
</tr>
<tr>
<td>35-39</td>
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<td>62</td>
<td>63.7</td>
<td>11.3</td>
<td>44.2</td>
<td>62.6</td>
<td>82.6</td>
<td>146.7</td>
</tr>
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<td>40-44</td>
<td>1620</td>
<td>39</td>
<td>63.8</td>
<td>12.2</td>
<td>44.0</td>
<td>62.5</td>
<td>81.6</td>
<td>147.1</td>
</tr>
<tr>
<td>45+</td>
<td>2813</td>
<td>59</td>
<td>64.5</td>
<td>11.8</td>
<td>45.2</td>
<td>62.8</td>
<td>83.1</td>
<td>141.2</td>
</tr>
</tbody>
</table>

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.
The figures below show the FEV₁% in different age groups, separately for each country. The dot shows the median, and the whiskers show the 25<sup>th</sup> and 75<sup>th</sup> percentiles (the median, the 25<sup>th</sup> percentile and the 75<sup>th</sup> 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.
Quartiles of FEV₁%: France

Quartiles of FEV₁%: Germany

Quartiles of FEV₁%: Greece

Quartiles of FEV₁%: Hungary

Quartiles of FEV₁%: Ireland

Quartiles of FEV₁%: Israel

Quartiles of FEV₁%: Italy

Quartiles of FEV₁%: Rep of Moldova

Quartiles of FEV₁%: The Netherlands

Quartiles of FEV₁%: North Macedonia
Figure 4.4 continued

Quartiles of FEV₁%: Norway

Quartiles of FEV₁%: Poland

Quartiles of FEV₁%: Portugal

Quartiles of FEV₁%: Romania

Quartiles of FEV₁%: Russian Federation

Quartiles of FEV₁%: Serbia

Quartiles of FEV₁%: Slovak Republic

Quartiles of FEV₁%: Slovenia

Quartiles of FEV₁%: Spain

Quartiles of FEV₁%: 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  FEV₁% 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 FEV₁ 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 FEV₁% by severity group, by country and overall. Patients with an FEV₁% higher than 80% are generally considered to have mild lung disease, patients with FEV₁% between 80% and 40% moderate lung disease, and patients with FEV₁ <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 FEV₁%, 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\textsubscript{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\textsubscript{1} measurement and are excluded from the graph.
Note: The United Kingdom reports best FEV\textsubscript{1} 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 FEV\textsubscript{1} could be from the previous calendar year.

**Figure 4.7** FEV\textsubscript{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\textsubscript{1} measurement and are excluded from the graph.
Note: The United Kingdom reports best FEV\textsubscript{1} 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 FEV\textsubscript{1} 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.

<table>
<thead>
<tr>
<th>Country</th>
<th>Chronic <em>Pseudomonas aeruginosa</em> number (%)</th>
<th>Chronic <em>Burkholderia cepacia complex species</em> number (%)</th>
<th><em>Haemophilus influenzae</em> number (%)</th>
</tr>
</thead>
<tbody>
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<td>Missing/unknown</td>
<td>No</td>
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</tr>
<tr>
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<td>(30.93)</td>
</tr>
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<td>(41.67)</td>
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<td>(0.27)</td>
<td>(92.28)</td>
<td>(7.45)</td>
</tr>
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<td>Belarus</td>
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<td>(74.15)</td>
<td>(25.85)</td>
</tr>
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<td>(0.91)</td>
<td>(87.64)</td>
<td>(11.45)</td>
</tr>
</tbody>
</table>

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 shows, separately by country, the frequency of chronic *Pseudomonas aeruginosa*, chronic *Burkholderia cepacia complex species* and *Haemophilus influenzae* number 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>
<thead>
<tr>
<th>Country</th>
<th>Chronic <em>Pseudomonas aeruginosa</em></th>
<th>Chronic <em>Burkholderia cepacia complex species</em></th>
<th>Haemophilus influenzae</th>
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</tr>
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</tr>
<tr>
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<td>(26.97)</td>
</tr>
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<td>3</td>
</tr>
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<td>(94.64)</td>
<td>(2.68)</td>
</tr>
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<tr>
<td></td>
<td>(1.67)</td>
<td>(87.11)</td>
<td>(11.22)</td>
</tr>
<tr>
<td>Turkey</td>
<td>14</td>
<td>1421</td>
<td>307</td>
</tr>
<tr>
<td></td>
<td>(0.80)</td>
<td>(81.58)</td>
<td>(17.62)</td>
</tr>
<tr>
<td>Ukraine</td>
<td>2</td>
<td>71</td>
<td>48</td>
</tr>
<tr>
<td></td>
<td>(1.65)</td>
<td>(58.68)</td>
<td>(39.67)</td>
</tr>
<tr>
<td>United Kingdom$^3$</td>
<td>5</td>
<td>3978</td>
<td>257</td>
</tr>
<tr>
<td></td>
<td>(0.12)</td>
<td>(93.82)</td>
<td>(6.06)</td>
</tr>
<tr>
<td>Total</td>
<td>362</td>
<td>19847</td>
<td>2968</td>
</tr>
<tr>
<td></td>
<td>(1.56)</td>
<td>(85.63)</td>
<td>(12.81)</td>
</tr>
</tbody>
</table>

$^3$ 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>
<thead>
<tr>
<th>Country</th>
<th>Chronic <em>Pseudomonas aeruginosa</em> number (%)</th>
<th>Chronic <em>Burkholderia cepacia</em> complex species number (%)</th>
<th>Haemophilus influenzae number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Missing/unknown</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Albania</td>
<td>(0.00)</td>
<td>(66.67)</td>
<td>(33.33)</td>
</tr>
<tr>
<td>Austria</td>
<td>(0.29)</td>
<td>(54.10)</td>
<td>(45.61)</td>
</tr>
<tr>
<td>Belarus</td>
<td>(0.00)</td>
<td>(62.50)</td>
<td>(37.50)</td>
</tr>
<tr>
<td>Belgium</td>
<td>(0.15)</td>
<td>(64.83)</td>
<td>(35.02)</td>
</tr>
<tr>
<td>Bulgaria</td>
<td>(0.00)</td>
<td>(30.67)</td>
<td>(69.33)</td>
</tr>
<tr>
<td>Croatia</td>
<td>(4.88)</td>
<td>(19.51)</td>
<td>(75.61)</td>
</tr>
<tr>
<td>Cyprus</td>
<td>(0.00)</td>
<td>(64.29)</td>
<td>(35.71)</td>
</tr>
<tr>
<td>Czech Republic</td>
<td>7 (2.61)</td>
<td>(67.54)</td>
<td>(29.85)</td>
</tr>
<tr>
<td>Denmark</td>
<td>(0.74)</td>
<td>(59.26)</td>
<td>(40.00)</td>
</tr>
<tr>
<td>France</td>
<td>(0.00)</td>
<td>(64.73)</td>
<td>(35.27)</td>
</tr>
<tr>
<td>Germany</td>
<td>(3.17)</td>
<td>(45.69)</td>
<td>(51.14)</td>
</tr>
<tr>
<td>Greece</td>
<td>(3.69)</td>
<td>(24.16)</td>
<td>(72.15)</td>
</tr>
<tr>
<td>Hungary</td>
<td>(2.05)</td>
<td>(46.67)</td>
<td>(51.28)</td>
</tr>
<tr>
<td>Ireland¹</td>
<td>(0.31)</td>
<td>(58.25)</td>
<td>(41.44)</td>
</tr>
<tr>
<td>Israel</td>
<td>(3.46)</td>
<td>(40.25)</td>
<td>(56.29)</td>
</tr>
<tr>
<td>Italy²</td>
<td>(6.95)</td>
<td>(51.67)</td>
<td>(41.38)</td>
</tr>
<tr>
<td>Latvia</td>
<td>(3.07)</td>
<td>(23.08)</td>
<td>(46.15)</td>
</tr>
<tr>
<td>Lithuania</td>
<td>(0.00)</td>
<td>(84.62)</td>
<td>(15.38)</td>
</tr>
<tr>
<td>Luxembourg</td>
<td>(4.76)</td>
<td>(61.91)</td>
<td>(33.33)</td>
</tr>
<tr>
<td>Moldova</td>
<td>(11.11)</td>
<td>(11.11)</td>
<td>(77.78)</td>
</tr>
<tr>
<td>The Netherlands</td>
<td>43 (5.09)</td>
<td>(47.28)</td>
<td>(47.63)</td>
</tr>
</tbody>
</table>

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**

<table>
<thead>
<tr>
<th>Country</th>
<th>Chronic <em>Pseudomonas aeruginosa</em> number (%)</th>
<th>Chronic <em>Burkholderia cepacia complex</em> species number (%)</th>
<th>Haemophilus influenzae number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Missing/unknown</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>North Macedonia</td>
<td>1 (2.50)</td>
<td>12</td>
<td>27</td>
</tr>
<tr>
<td>Norway</td>
<td>4 (2.42)</td>
<td>106</td>
<td>55</td>
</tr>
<tr>
<td>Poland</td>
<td>5 (1.48)</td>
<td>136</td>
<td>196</td>
</tr>
<tr>
<td>Portugal</td>
<td>6 (4.35)</td>
<td>83</td>
<td>49</td>
</tr>
<tr>
<td>Romania</td>
<td>1 (9.09)</td>
<td>3</td>
<td>7</td>
</tr>
<tr>
<td>Russian Federation</td>
<td>7 (0.92)</td>
<td>353</td>
<td>405</td>
</tr>
<tr>
<td>Serbia</td>
<td>1 (1.67)</td>
<td>18</td>
<td>41</td>
</tr>
<tr>
<td>Slovak Republic</td>
<td>4 (2.68)</td>
<td>77</td>
<td>68</td>
</tr>
<tr>
<td>Slovenia</td>
<td>1 (2.44)</td>
<td>17</td>
<td>23</td>
</tr>
<tr>
<td>Spain</td>
<td>13 (1.26)</td>
<td>605</td>
<td>414</td>
</tr>
<tr>
<td>Sweden</td>
<td>4 (1.14)</td>
<td>175</td>
<td>173</td>
</tr>
<tr>
<td>Switzerland</td>
<td>7 (1.43)</td>
<td>241</td>
<td>240</td>
</tr>
<tr>
<td>Turkey</td>
<td>4 (1.67)</td>
<td>125</td>
<td>111</td>
</tr>
<tr>
<td>Ukraine</td>
<td>0 (0.00)</td>
<td>9</td>
<td>21</td>
</tr>
<tr>
<td>United Kingdom$^3$</td>
<td>3 (0.05)</td>
<td>3194</td>
<td>2262</td>
</tr>
<tr>
<td>Total</td>
<td>464 (2.04)</td>
<td>12316</td>
<td>9976</td>
</tr>
</tbody>
</table>

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 influenzae* 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.**

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.

**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.
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.

<table>
<thead>
<tr>
<th>Country</th>
<th>Chronic <em>Staphylococcus aureus</em> number (%)</th>
<th>MRSA number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Missing/unknown</td>
<td>No</td>
</tr>
<tr>
<td>Albania</td>
<td>5 (5.15)</td>
<td>59 (60.83)</td>
</tr>
<tr>
<td>Armenia</td>
<td>0 (0.00)</td>
<td>5 (20.83)</td>
</tr>
<tr>
<td>Austria</td>
<td>1 (0.27)</td>
<td>139 (36.97)</td>
</tr>
<tr>
<td>Belarus</td>
<td>0 (0.00)</td>
<td>72 (48.98)</td>
</tr>
<tr>
<td>Belgium</td>
<td>460 (100)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Bulgaria</td>
<td>0 (0.00)</td>
<td>86 (77.48)</td>
</tr>
<tr>
<td>Croatia</td>
<td>11 (12.22)</td>
<td>43 (47.78)</td>
</tr>
<tr>
<td>Cyprus</td>
<td>0 (0.00)</td>
<td>11 (84.62)</td>
</tr>
<tr>
<td>Czech Republic</td>
<td>3 (0.92)</td>
<td>169 (52.00)</td>
</tr>
<tr>
<td>Denmark</td>
<td>0 (0.00)</td>
<td>141 (70.85)</td>
</tr>
<tr>
<td>France</td>
<td>0 (0.00)</td>
<td>1554 (57.24)</td>
</tr>
<tr>
<td>Georgia</td>
<td>0 (0.00)</td>
<td>50 (69.44)</td>
</tr>
<tr>
<td>Germany</td>
<td>29 (1.12)</td>
<td>1591 (61.55)</td>
</tr>
<tr>
<td>Greece</td>
<td>1 (0.99)</td>
<td>61 (60.40)</td>
</tr>
<tr>
<td>Hungary</td>
<td>0 (0.00)</td>
<td>108 (43.20)</td>
</tr>
<tr>
<td>Ireland(^1)</td>
<td>0 (0.00)</td>
<td>303 (60.48)</td>
</tr>
<tr>
<td>Israel</td>
<td>7 (3.59)</td>
<td>99 (50.77)</td>
</tr>
<tr>
<td>Italy(^2)</td>
<td>141 (6.14)</td>
<td>1385 (60.27)</td>
</tr>
<tr>
<td>Latvia</td>
<td>16 (50.00)</td>
<td>1 (3.13)</td>
</tr>
<tr>
<td>Lithuania</td>
<td>1 (7.14)</td>
<td>3 (21.43)</td>
</tr>
<tr>
<td>Luxembourg</td>
<td>0 (0.00)</td>
<td>7 (46.67)</td>
</tr>
<tr>
<td>Moldova</td>
<td>0 (0.00)</td>
<td>4 (9.09)</td>
</tr>
<tr>
<td>The Netherlands</td>
<td>5 (0.91)</td>
<td>355 (64.54)</td>
</tr>
</tbody>
</table>

\(^1\) 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 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.

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

<table>
<thead>
<tr>
<th>Country</th>
<th>Chronic <em>Staphylococcus aureus</em> number (%)</th>
<th>MRSA number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Missing/unknown</td>
<td>No</td>
</tr>
<tr>
<td>Albania</td>
<td>1 (11.1)</td>
<td>2</td>
</tr>
<tr>
<td>Austria</td>
<td>1 (0.29)</td>
<td>122</td>
</tr>
<tr>
<td>Belarus</td>
<td>0 (0.00)</td>
<td>3</td>
</tr>
<tr>
<td>Belgium</td>
<td>654 (100)</td>
<td>0</td>
</tr>
<tr>
<td>Bulgaria</td>
<td>0 (0.00)</td>
<td>57</td>
</tr>
<tr>
<td>Croatia</td>
<td>2 (4.88)</td>
<td>17</td>
</tr>
<tr>
<td>Cyprus</td>
<td>2 (14.29)</td>
<td>8</td>
</tr>
<tr>
<td>Czech Republic</td>
<td>10 (3.73)</td>
<td>130</td>
</tr>
<tr>
<td>Denmark</td>
<td>2 (0.74)</td>
<td>165</td>
</tr>
<tr>
<td>France</td>
<td>0 (0.00)</td>
<td>1748</td>
</tr>
<tr>
<td>Germany</td>
<td>103 (3.08)</td>
<td>1652</td>
</tr>
<tr>
<td>Greece</td>
<td>13 (3.73)</td>
<td>158</td>
</tr>
<tr>
<td>Hungary</td>
<td>4 (2.05)</td>
<td>80</td>
</tr>
<tr>
<td>Ireland¹</td>
<td>2 (0.31)</td>
<td>411</td>
</tr>
<tr>
<td>Israel</td>
<td>11 (3.46)</td>
<td>201</td>
</tr>
<tr>
<td>Italy²</td>
<td>206 (6.92)</td>
<td>1704</td>
</tr>
<tr>
<td>Latvia</td>
<td>7 (53.85)</td>
<td>0</td>
</tr>
<tr>
<td>Lithuania</td>
<td>0 (0.00)</td>
<td>3</td>
</tr>
<tr>
<td>Luxembourg</td>
<td>1 (4.76)</td>
<td>8</td>
</tr>
<tr>
<td>Moldova</td>
<td>1 (11.11)</td>
<td>3</td>
</tr>
<tr>
<td>The Netherlands</td>
<td>45 (5.33)</td>
<td>394</td>
</tr>
</tbody>
</table>

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

<table>
<thead>
<tr>
<th>Country</th>
<th>Chronic Staphylococcus aureus number (%)</th>
<th>MRSA number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Missing/unknown</td>
<td>No</td>
</tr>
<tr>
<td>North Macedonia</td>
<td>0</td>
<td>27</td>
</tr>
<tr>
<td></td>
<td>(0.00)</td>
<td>(67.50)</td>
</tr>
<tr>
<td>Norway</td>
<td>8</td>
<td>64</td>
</tr>
<tr>
<td></td>
<td>(4.85)</td>
<td>(38.79)</td>
</tr>
<tr>
<td>Poland</td>
<td>6</td>
<td>130</td>
</tr>
<tr>
<td></td>
<td>(1.78)</td>
<td>(38.58)</td>
</tr>
<tr>
<td>Portugal</td>
<td>5</td>
<td>60</td>
</tr>
<tr>
<td></td>
<td>(3.62)</td>
<td>(43.48)</td>
</tr>
<tr>
<td>Romania</td>
<td>0</td>
<td>6</td>
</tr>
<tr>
<td></td>
<td>(0.00)</td>
<td>(54.55)</td>
</tr>
<tr>
<td>Russian Federation</td>
<td>11</td>
<td>345</td>
</tr>
<tr>
<td></td>
<td>(1.44)</td>
<td>(45.10)</td>
</tr>
<tr>
<td>Serbia</td>
<td>1</td>
<td>28</td>
</tr>
<tr>
<td></td>
<td>(1.67)</td>
<td>(46.67)</td>
</tr>
<tr>
<td>Slovak Republic</td>
<td>5</td>
<td>67</td>
</tr>
<tr>
<td></td>
<td>(3.36)</td>
<td>(44.97)</td>
</tr>
<tr>
<td>Slovenia</td>
<td>1</td>
<td>11</td>
</tr>
<tr>
<td></td>
<td>(2.44)</td>
<td>(26.83)</td>
</tr>
<tr>
<td>Spain</td>
<td>15</td>
<td>558</td>
</tr>
<tr>
<td></td>
<td>(1.45)</td>
<td>(54.07)</td>
</tr>
<tr>
<td>Sweden</td>
<td>36</td>
<td>166</td>
</tr>
<tr>
<td></td>
<td>(10.23)</td>
<td>(47.16)</td>
</tr>
<tr>
<td>Switzerland</td>
<td>10</td>
<td>180</td>
</tr>
<tr>
<td></td>
<td>(2.05)</td>
<td>(36.88)</td>
</tr>
<tr>
<td>Turkey</td>
<td>3</td>
<td>137</td>
</tr>
<tr>
<td></td>
<td>(1.25)</td>
<td>(57.08)</td>
</tr>
<tr>
<td>Ukraine</td>
<td>0</td>
<td>11</td>
</tr>
<tr>
<td></td>
<td>(0.00)</td>
<td>(36.67)</td>
</tr>
<tr>
<td>United Kingdom4</td>
<td>3</td>
<td>4262</td>
</tr>
<tr>
<td></td>
<td>(0.05)</td>
<td>(78.08)</td>
</tr>
<tr>
<td>Total</td>
<td>1169</td>
<td>12918</td>
</tr>
<tr>
<td></td>
<td>(5.14)</td>
<td>(56.76)</td>
</tr>
</tbody>
</table>

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.

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

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.

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.
### 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.

<table>
<thead>
<tr>
<th>Country</th>
<th>Non-tuberculous mycobacteria (NTM) infection this year</th>
<th><em>Stenotrophomonas maltophilia</em> infection this year</th>
<th><em>Achromobacter</em> species infection this year</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Missing/unknown</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Albania</td>
<td>(5.15)</td>
<td>92</td>
<td>0</td>
</tr>
<tr>
<td>Armenia</td>
<td>(50.00)</td>
<td>12</td>
<td>0</td>
</tr>
<tr>
<td>Austria</td>
<td>(1.86)</td>
<td>364</td>
<td>5</td>
</tr>
<tr>
<td>Belarus</td>
<td>(100)</td>
<td>147</td>
<td>0</td>
</tr>
<tr>
<td>Belgium</td>
<td>(0.00)</td>
<td>453</td>
<td>7</td>
</tr>
<tr>
<td>Bulgaria</td>
<td>(89.19)</td>
<td>12</td>
<td>0</td>
</tr>
<tr>
<td>Croatia</td>
<td>(61.11)</td>
<td>35</td>
<td>0</td>
</tr>
<tr>
<td>Cyprus</td>
<td>(0.00)</td>
<td>12</td>
<td>1</td>
</tr>
<tr>
<td>Czech Republic</td>
<td>(72.61)</td>
<td>84</td>
<td>5</td>
</tr>
<tr>
<td>Denmark</td>
<td>(0.00)</td>
<td>197</td>
<td>2</td>
</tr>
<tr>
<td>France</td>
<td>(0.00)</td>
<td>2647</td>
<td>68</td>
</tr>
<tr>
<td>Georgia</td>
<td>(100)</td>
<td>72</td>
<td>0</td>
</tr>
<tr>
<td>Germany</td>
<td>(76.67)</td>
<td>574</td>
<td>29</td>
</tr>
<tr>
<td>Greece</td>
<td>(0.00)</td>
<td>101</td>
<td>0</td>
</tr>
<tr>
<td>Hungary</td>
<td>(3.20)</td>
<td>240</td>
<td>2</td>
</tr>
<tr>
<td>Ireland</td>
<td>(0.00)</td>
<td>486</td>
<td>15</td>
</tr>
<tr>
<td>Israel</td>
<td>(5.64)</td>
<td>169</td>
<td>15</td>
</tr>
<tr>
<td>Italy</td>
<td>(0.26)</td>
<td>2268</td>
<td>24</td>
</tr>
<tr>
<td>Latvia</td>
<td>(46.88)</td>
<td>17</td>
<td>0</td>
</tr>
<tr>
<td>Lithuania</td>
<td>(0.00)</td>
<td>14</td>
<td>0</td>
</tr>
<tr>
<td>Luxembourg</td>
<td>(0.00)</td>
<td>15</td>
<td>0</td>
</tr>
<tr>
<td>Moldova</td>
<td>(0.00)</td>
<td>44</td>
<td>0</td>
</tr>
<tr>
<td>The Netherlands</td>
<td>(1.82)</td>
<td>522</td>
<td>18</td>
</tr>
</tbody>
</table>
Table 5.5 shows the frequency of three other infections, non-tuberculous mycobacteria (NTM), Stenotrophomonas maltophilia and Achromobacter species in children.

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 5.6 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.

<table>
<thead>
<tr>
<th>Country</th>
<th>Non-tuberculous mycobacteria (NTM) infection this year number (%)</th>
<th><em>Stenotrophomonas maltophilia</em> infection this year number (%)</th>
<th><em>Achromobacter</em> species infection this year number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Missing/unknown</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td><strong>Albania</strong></td>
<td>0 (0)</td>
<td>9</td>
<td>0</td>
</tr>
<tr>
<td><strong>Austria</strong></td>
<td>34 (9.94)</td>
<td>279</td>
<td>29</td>
</tr>
<tr>
<td><strong>Belarus</strong></td>
<td>8 (100)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td><strong>Belgium</strong></td>
<td>1 (0.15)</td>
<td>631</td>
<td>22</td>
</tr>
<tr>
<td><strong>Bulgaria</strong></td>
<td>68 (90.67)</td>
<td>6</td>
<td>0</td>
</tr>
<tr>
<td><strong>Croatia</strong></td>
<td>4 (9.76)</td>
<td>35</td>
<td>2</td>
</tr>
<tr>
<td><strong>Cyprus</strong></td>
<td>2 (14.29)</td>
<td>12</td>
<td>0</td>
</tr>
<tr>
<td><strong>Czech Republic</strong></td>
<td>47 (17.54)</td>
<td>204</td>
<td>17</td>
</tr>
<tr>
<td><strong>Denmark</strong></td>
<td>2 (0.74)</td>
<td>250</td>
<td>18</td>
</tr>
<tr>
<td><strong>France</strong></td>
<td>0 (0.00)</td>
<td>2973</td>
<td>179</td>
</tr>
<tr>
<td><strong>Germany</strong></td>
<td>1729 (51.77)</td>
<td>1468</td>
<td>143</td>
</tr>
<tr>
<td><strong>Greece</strong></td>
<td>0 (0.00)</td>
<td>284</td>
<td>14</td>
</tr>
<tr>
<td><strong>Hungary</strong></td>
<td>5 (12.56)</td>
<td>179</td>
<td>11</td>
</tr>
<tr>
<td><strong>Ireland</strong></td>
<td>2 (0.31)</td>
<td>616</td>
<td>19</td>
</tr>
<tr>
<td><strong>Israel</strong></td>
<td>16 (5.03)</td>
<td>269</td>
<td>33</td>
</tr>
<tr>
<td><strong>Italy</strong></td>
<td>17 (0.57)</td>
<td>2927</td>
<td>33</td>
</tr>
<tr>
<td><strong>Latvia</strong></td>
<td>4 (30.77)</td>
<td>9</td>
<td>0</td>
</tr>
<tr>
<td><strong>Lithuania</strong></td>
<td>0 (0)</td>
<td>13</td>
<td>0</td>
</tr>
<tr>
<td><strong>Luxembourg</strong></td>
<td>2 (9.52)</td>
<td>18</td>
<td>1</td>
</tr>
<tr>
<td><strong>Moldova</strong></td>
<td>9 (100)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td><strong>The Netherlands</strong></td>
<td>101 (11.97)</td>
<td>711</td>
<td>32</td>
</tr>
</tbody>
</table>
### Table 5.6 continued

<table>
<thead>
<tr>
<th>Country</th>
<th>Non-tuberculous mycobacteria (NTM) infection this year number (%)</th>
<th>Stenotrophomonas maltophilia infection this year number (%)</th>
<th>Achromobacter species infection this year number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Missing/ unknown  No  Yes</td>
<td>Missing/ unknown  No  Yes</td>
<td>Missing/ unknown  No  Yes</td>
</tr>
<tr>
<td>North Macedonia</td>
<td>0  39 (2.50)</td>
<td>0  40 (100)</td>
<td>0  40 (100)</td>
</tr>
<tr>
<td>Norway</td>
<td>15  141 (9.09)</td>
<td>5  131 (3.03)</td>
<td>5  151 (3.03)</td>
</tr>
<tr>
<td>Poland</td>
<td>27  307 (8.01)</td>
<td>7  307 (2.50)</td>
<td>8  298 (3.62)</td>
</tr>
<tr>
<td>Portugal</td>
<td>5  124 (3.62)</td>
<td>5  120 (3.62)</td>
<td>5  121 (3.62)</td>
</tr>
<tr>
<td>Romania</td>
<td>1  10 (9.09)</td>
<td>0  11 (0.00)</td>
<td>0  11 (0.00)</td>
</tr>
<tr>
<td>Russian Federation</td>
<td>38  708 (4.97)</td>
<td>21  711 (2.75)</td>
<td>15  684 (1.96)</td>
</tr>
<tr>
<td>Serbia</td>
<td>1  59 (1.67)</td>
<td>1  56 (1.67)</td>
<td>2  53 (3.33)</td>
</tr>
<tr>
<td>Slovak Republic</td>
<td>5  141 (3.36)</td>
<td>6  130 (4.03)</td>
<td>7  132 (4.70)</td>
</tr>
<tr>
<td>Slovenia</td>
<td>4  37 (9.76)</td>
<td>6  29 (14.63)</td>
<td>3  37 (7.32)</td>
</tr>
<tr>
<td>Spain</td>
<td>22  933 (2.13)</td>
<td>13  928 (1.26)</td>
<td>23  905 (2.23)</td>
</tr>
<tr>
<td>Sweden</td>
<td>2  326 (0.57)</td>
<td>0  306 (0.00)</td>
<td>0  339 (0.00)</td>
</tr>
<tr>
<td>Switzerland</td>
<td>24  423 (4.92)</td>
<td>9  408 (1.84)</td>
<td>11  445 (2.25)</td>
</tr>
<tr>
<td>Turkey</td>
<td>6  230 (2.50)</td>
<td>4  221 (1.67)</td>
<td>1  222 (0.42)</td>
</tr>
<tr>
<td>Ukraine</td>
<td>26  326 (86.67)</td>
<td>6  23 (20.00)</td>
<td>1  29 (3.33)</td>
</tr>
<tr>
<td>United Kingdom</td>
<td>3  4958 (0.05)</td>
<td>3  4690 (0.05)</td>
<td>3  5162 (0.05)</td>
</tr>
<tr>
<td>Total</td>
<td>2230  19283 (9.80)</td>
<td>308  20179 (5.46)</td>
<td>499  20589 (1.35)</td>
</tr>
</tbody>
</table>

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.**

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/m²; 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.

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

\textsuperscript{1} Norway: sometimes any value (instead of last of the year) for height is used when no lung function test was available.

\textsuperscript{2} Sweden: if there is no lung function measurement, the height and weight reported are those measured at annual review.

\textsuperscript{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.

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

<sup>1</sup> Norway: sometimes any value (instead of last of the year) for height is used when no lung function test was available.

<sup>2</sup> Sweden: if there is no lung function measurement, the height and weight reported are those measured at annual review.

<sup>3</sup> 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.

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.

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

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: Latvia

Quartiles of z-scores for height: Italy
[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

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

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

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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.

<table>
<thead>
<tr>
<th>Country</th>
<th>N</th>
<th>N miss</th>
<th>Mean</th>
<th>Min</th>
<th>25th pctl (25% of the patients are below this z-score for weight)</th>
<th>Median (50% of the patients are below this z-score for weight)</th>
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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.**

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<th>N miss</th>
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<th>Min</th>
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<th>Median (50% of the patients are below this z-score for weight)</th>
<th>75th pctl (75% of the patients are below this z-score for weight)</th>
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</tbody>
</table>

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.**

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.**

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

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.
[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

Quartiles of z-scores for weight: Israel

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

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

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

---

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

<table>
<thead>
<tr>
<th>Country</th>
<th>N</th>
<th>N Miss</th>
<th>Mean</th>
<th>Min</th>
<th>25th pctl (25% of the patients are below this z-score for BMI)</th>
<th>Median</th>
<th>75th pctl (75% of the patients are below this z-score for BMI)</th>
<th>Max</th>
</tr>
</thead>
<tbody>
<tr>
<td>Albania</td>
<td>81</td>
<td>13</td>
<td>-0.4</td>
<td>-5.0</td>
<td>-1.3</td>
<td>-0.2</td>
<td>0.4</td>
<td>2.8</td>
</tr>
<tr>
<td>Armenia</td>
<td>25</td>
<td>0</td>
<td>-0.7</td>
<td>-6.8</td>
<td>-1.1</td>
<td>-0.6</td>
<td>0.2</td>
<td>2.3</td>
</tr>
<tr>
<td>Austria</td>
<td>346</td>
<td>0</td>
<td>-0.2</td>
<td>-3.9</td>
<td>-0.8</td>
<td>-0.2</td>
<td>0.4</td>
<td>2.5</td>
</tr>
<tr>
<td>Belarus</td>
<td>84</td>
<td>3</td>
<td>-0.9</td>
<td>-6.8</td>
<td>-1.7</td>
<td>-0.7</td>
<td>0.0</td>
<td>1.7</td>
</tr>
<tr>
<td>Belgium</td>
<td>440</td>
<td>0</td>
<td>-0.3</td>
<td>-2.9</td>
<td>-0.9</td>
<td>-0.3</td>
<td>0.3</td>
<td>2.6</td>
</tr>
<tr>
<td>Bulgaria</td>
<td>97</td>
<td>0</td>
<td>-0.8</td>
<td>-6.7</td>
<td>-1.6</td>
<td>-0.8</td>
<td>0.1</td>
<td>2.3</td>
</tr>
<tr>
<td>Croatia</td>
<td>75</td>
<td>2</td>
<td>-0.5</td>
<td>-5.5</td>
<td>-1.2</td>
<td>-0.4</td>
<td>0.1</td>
<td>3.7</td>
</tr>
<tr>
<td>Cyprus</td>
<td>12</td>
<td>0</td>
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<td>-3.2</td>
<td>-1.1</td>
<td>-0.5</td>
<td>-0.3</td>
<td>1.7</td>
</tr>
<tr>
<td>Czech Republic</td>
<td>292</td>
<td>0</td>
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<td>-0.9</td>
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<tr>
<td>Denmark</td>
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<td>0.3</td>
<td>1.9</td>
</tr>
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<td>-0.5</td>
<td>-9.9</td>
<td>-1.1</td>
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<td>0.2</td>
<td>3.5</td>
</tr>
<tr>
<td>Georgia</td>
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<td>-2.9</td>
<td>-0.6</td>
<td>0.1</td>
<td>0.7</td>
<td>2.9</td>
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<tr>
<td>Germany</td>
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<td>-6.5</td>
<td>-0.9</td>
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<td>0.3</td>
<td>2.8</td>
</tr>
<tr>
<td>Greece</td>
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<td>-3.0</td>
<td>-0.5</td>
<td>0.4</td>
<td>1.0</td>
<td>3.1</td>
</tr>
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<td>-5.3</td>
<td>-1.5</td>
<td>-0.6</td>
<td>0.1</td>
<td>2.6</td>
</tr>
<tr>
<td>Ireland</td>
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<td>-0.3</td>
<td>0.2</td>
<td>0.8</td>
<td>2.8</td>
</tr>
<tr>
<td>Israel</td>
<td>197</td>
<td>0</td>
<td>-0.3</td>
<td>-5.2</td>
<td>-0.9</td>
<td>-0.2</td>
<td>0.6</td>
<td>2.2</td>
</tr>
<tr>
<td>Italy</td>
<td>2086</td>
<td>25</td>
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<td>-4.3</td>
<td>-0.8</td>
<td>-0.1</td>
<td>0.6</td>
<td>3.9</td>
</tr>
<tr>
<td>Latvia</td>
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<td>-0.9</td>
<td>-2.3</td>
<td>-1.6</td>
<td>-1.1</td>
<td>-0.2</td>
<td>0.8</td>
</tr>
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<td>-1.5</td>
<td>-1.0</td>
<td>-0.4</td>
<td>0.4</td>
</tr>
<tr>
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<td>-0.3</td>
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<td>-0.9</td>
<td>-0.3</td>
<td>0.2</td>
<td>0.9</td>
</tr>
<tr>
<td>Rep of Moldova</td>
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<td>-4.2</td>
<td>-1.9</td>
<td>-1.1</td>
<td>0.1</td>
<td>2.9</td>
</tr>
<tr>
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<td>-0.2</td>
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<td>-0.8</td>
<td>-0.1</td>
<td>0.4</td>
<td>1.8</td>
</tr>
<tr>
<td>North Macedonia</td>
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<td>-1.2</td>
<td>-0.2</td>
<td>0.7</td>
<td>2.1</td>
</tr>
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<td>-1.0</td>
<td>-0.3</td>
<td>0.4</td>
<td>1.7</td>
</tr>
<tr>
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<td>0.4</td>
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</tr>
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<td>-0.9</td>
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<td>0.3</td>
<td>1.7</td>
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<td>Romania</td>
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<td>-1.0</td>
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<td>-0.7</td>
<td>0.0</td>
<td>1.9</td>
</tr>
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<td>-1.5</td>
<td>-0.6</td>
<td>0.2</td>
<td>4.0</td>
</tr>
<tr>
<td>Serbia</td>
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<td>-0.6</td>
<td>-5.0</td>
<td>-1.2</td>
<td>-0.6</td>
<td>0.2</td>
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</tr>
<tr>
<td>Slovak Republic</td>
<td>118</td>
<td>4</td>
<td>-0.5</td>
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<td>-1.3</td>
<td>-0.5</td>
<td>0.2</td>
<td>2.3</td>
</tr>
<tr>
<td>Slovenia</td>
<td>56</td>
<td>0</td>
<td>-0.4</td>
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<td>-1.0</td>
<td>-0.3</td>
<td>0.2</td>
<td>1.9</td>
</tr>
<tr>
<td>Spain</td>
<td>995</td>
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<td>-0.2</td>
<td>-4.6</td>
<td>-0.8</td>
<td>-0.1</td>
<td>0.5</td>
<td>2.6</td>
</tr>
<tr>
<td>Sweden2</td>
<td>244</td>
<td>0</td>
<td>-0.2</td>
<td>-3.0</td>
<td>-0.7</td>
<td>-0.1</td>
<td>0.5</td>
<td>2.0</td>
</tr>
<tr>
<td>Switzerland</td>
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<td>-0.2</td>
<td>-3.1</td>
<td>-0.7</td>
<td>-0.2</td>
<td>0.4</td>
<td>3.9</td>
</tr>
<tr>
<td>Turkey</td>
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<td>-0.7</td>
<td>-9.3</td>
<td>-1.6</td>
<td>-0.6</td>
<td>0.3</td>
<td>4.4</td>
</tr>
<tr>
<td>Ukraine</td>
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<td>-6.5</td>
<td>-1.6</td>
<td>-1.0</td>
<td>-0.2</td>
<td>2.7</td>
</tr>
<tr>
<td>United Kingdom2</td>
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<td>-7.0</td>
<td>-0.5</td>
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</tr>
<tr>
<td>Total</td>
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<td>-9.9</td>
<td>-0.9</td>
<td>-0.2</td>
<td>0.4</td>
<td>4.4</td>
</tr>
</tbody>
</table>

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

<table>
<thead>
<tr>
<th>Country</th>
<th>N</th>
<th>N Miss</th>
<th>Mean</th>
<th>Min</th>
<th>25th pctl (25% of the patients are below this BMI)</th>
<th>Median (50% of the patients are below this BMI)</th>
<th>75th pctl (75% of the patients are below this BMI)</th>
<th>Max</th>
</tr>
</thead>
<tbody>
<tr>
<td>Austria</td>
<td>335</td>
<td>0</td>
<td>21.8</td>
<td>14.9</td>
<td>19.5</td>
<td>21.3</td>
<td>23.6</td>
<td>38.5</td>
</tr>
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<td>Belgium</td>
<td>637</td>
<td>2</td>
<td>22.5</td>
<td>15.1</td>
<td>20.2</td>
<td>22.1</td>
<td>24.2</td>
<td>36.5</td>
</tr>
<tr>
<td>Bulgaria</td>
<td>67</td>
<td>0</td>
<td>20.2</td>
<td>12.3</td>
<td>17.9</td>
<td>19.7</td>
<td>21.8</td>
<td>40.1</td>
</tr>
<tr>
<td>Croatia</td>
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<td>0</td>
<td>21.5</td>
<td>15.1</td>
<td>20.2</td>
<td>21.8</td>
<td>23.1</td>
<td>27.8</td>
</tr>
<tr>
<td>Cyprus</td>
<td>13</td>
<td>0</td>
<td>23.5</td>
<td>19.1</td>
<td>21.6</td>
<td>22.6</td>
<td>24.1</td>
<td>34.7</td>
</tr>
<tr>
<td>Czech Republic</td>
<td>253</td>
<td>0</td>
<td>21.9</td>
<td>13.7</td>
<td>19.7</td>
<td>21.3</td>
<td>23.9</td>
<td>34.4</td>
</tr>
<tr>
<td>Denmark</td>
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<td>22.6</td>
<td>13.8</td>
<td>20.0</td>
<td>22.3</td>
<td>24.1</td>
<td>39.3</td>
</tr>
<tr>
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<td>2921</td>
<td>50</td>
<td>21.6</td>
<td>12.1</td>
<td>19.4</td>
<td>21.1</td>
<td>23.1</td>
<td>46.7</td>
</tr>
<tr>
<td>Germany</td>
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<td>13</td>
<td>21.8</td>
<td>13.1</td>
<td>19.4</td>
<td>21.3</td>
<td>23.5</td>
<td>42.1</td>
</tr>
<tr>
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<td>4</td>
<td>22.6</td>
<td>15.0</td>
<td>20.5</td>
<td>22.1</td>
<td>24.2</td>
<td>37.0</td>
</tr>
<tr>
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<td>20.5</td>
<td>14.4</td>
<td>18.6</td>
<td>20.1</td>
<td>22.3</td>
<td>30.8</td>
</tr>
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<td>15.2</td>
<td>20.8</td>
<td>22.6</td>
<td>24.8</td>
<td>44.7</td>
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<td>23.1</td>
<td>16.2</td>
<td>20.4</td>
<td>22.7</td>
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<tr>
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<td>19.6</td>
<td>21.0</td>
<td>24.7</td>
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<td>40.8</td>
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<td>20.6</td>
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<td>22.2</td>
<td>12.3</td>
<td>19.8</td>
<td>21.3</td>
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<td>18.2</td>
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<td>16.3</td>
<td>17.7</td>
<td>19.5</td>
<td>23.9</td>
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<td>19.7</td>
<td>12.0</td>
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<td>19.2</td>
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<td>17.9</td>
<td>19.6</td>
<td>21.5</td>
<td>25.1</td>
</tr>
<tr>
<td>Slovak Republic</td>
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<td>1</td>
<td>21.3</td>
<td>14.2</td>
<td>19.0</td>
<td>20.9</td>
<td>23.6</td>
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<td>21.0</td>
<td>15.1</td>
<td>19.7</td>
<td>21.3</td>
<td>22.0</td>
<td>25.2</td>
</tr>
<tr>
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<td>5</td>
<td>22.7</td>
<td>12.2</td>
<td>20.2</td>
<td>22.2</td>
<td>24.3</td>
<td>43.4</td>
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<tr>
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<td>10</td>
<td>22.8</td>
<td>13.9</td>
<td>20.4</td>
<td>22.3</td>
<td>24.4</td>
<td>39.0</td>
</tr>
<tr>
<td>Switzerland</td>
<td>467</td>
<td>1</td>
<td>21.7</td>
<td>13.8</td>
<td>19.4</td>
<td>21.3</td>
<td>23.4</td>
<td>36.6</td>
</tr>
<tr>
<td>Turkey</td>
<td>220</td>
<td>0</td>
<td>20.5</td>
<td>12.9</td>
<td>18.0</td>
<td>19.9</td>
<td>23.0</td>
<td>31.8</td>
</tr>
<tr>
<td>Ukraine</td>
<td>27</td>
<td>0</td>
<td>19.8</td>
<td>16.4</td>
<td>17.5</td>
<td>19.9</td>
<td>21.3</td>
<td>25.9</td>
</tr>
<tr>
<td>United Kingdom3</td>
<td>5213</td>
<td>49</td>
<td>23.2</td>
<td>12.1</td>
<td>20.6</td>
<td>22.6</td>
<td>25.2</td>
<td>49.3</td>
</tr>
<tr>
<td>Total</td>
<td>21487</td>
<td>308</td>
<td>22.3</td>
<td>12.0</td>
<td>19.8</td>
<td>21.8</td>
<td>24.1</td>
<td>49.5</td>
</tr>
</tbody>
</table>

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.
[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
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**

![Greece BMI chart](image)

**z-score of BMI < -2: Hungary**

![Hungary BMI chart](image)

**z-score of BMI < -2: Ireland**

![Ireland BMI chart](image)

**z-score of BMI < -2: Israel**

![Israel BMI chart](image)

**z-score of BMI < -2: Italy**

![Italy BMI chart](image)

**z-score of BMI < -2: The Netherlands**

![The Netherlands BMI chart](image)

**z-score of BMI < -2: Norway**

![Norway BMI chart](image)

**z-score of BMI < -2: Poland**

![Poland BMI chart](image)

**z-score of BMI < -2: Portugal**

![Portugal BMI chart](image)

**z-score of BMI < -2: Romania**

![Romania BMI chart](image)
[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.
[figure 6.8 continued]
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.

<table>
<thead>
<tr>
<th>Country</th>
<th>ABPA this year number (%)</th>
<th>CF RD this year number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Missing/unknown</td>
<td>No</td>
</tr>
<tr>
<td>Albania</td>
<td>1 (0.94)</td>
<td>105</td>
</tr>
<tr>
<td>Armenia</td>
<td>0 (0)</td>
<td>25</td>
</tr>
<tr>
<td>Austria</td>
<td>2 (0.28)</td>
<td>696</td>
</tr>
<tr>
<td>Belarus</td>
<td>0 (0.00)</td>
<td>154</td>
</tr>
<tr>
<td>Belgium</td>
<td>6 (0.54)</td>
<td>1004</td>
</tr>
<tr>
<td>Bulgaria</td>
<td>1 (0.54)</td>
<td>181</td>
</tr>
<tr>
<td>Croatia</td>
<td>1 (0.76)</td>
<td>129</td>
</tr>
<tr>
<td>Cyprus</td>
<td>0 (0)</td>
<td>27</td>
</tr>
<tr>
<td>Czech Republic</td>
<td>0 (0.00)</td>
<td>576</td>
</tr>
<tr>
<td>Denmark</td>
<td>279 (59.49)</td>
<td>186</td>
</tr>
<tr>
<td>France</td>
<td>0 (0.00)</td>
<td>5325</td>
</tr>
<tr>
<td>Georgia</td>
<td>1 (1.39)</td>
<td>71</td>
</tr>
<tr>
<td>Germany</td>
<td>63 (1.06)</td>
<td>5585</td>
</tr>
<tr>
<td>Greece</td>
<td>11 (2.76)</td>
<td>377</td>
</tr>
<tr>
<td>Hungary</td>
<td>6 (1.35)</td>
<td>426</td>
</tr>
<tr>
<td>Ireland</td>
<td>2 (0.18)</td>
<td>1070</td>
</tr>
<tr>
<td>Israel</td>
<td>17 (3.31)</td>
<td>462</td>
</tr>
<tr>
<td>Italy</td>
<td>34 (0.64)</td>
<td>5090</td>
</tr>
<tr>
<td>Latvia</td>
<td>2 (4.44)</td>
<td>43</td>
</tr>
<tr>
<td>Lithuania</td>
<td>0 (0)</td>
<td>27</td>
</tr>
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</table>
### Table 7.1 continued

<table>
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<tr>
<th>Country</th>
<th>ABPA this year number (%)</th>
<th>CFRD this year number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
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</tr>
<tr>
<td>Luxembourg</td>
<td>0 (0.00)</td>
<td>32 (88.89)</td>
</tr>
<tr>
<td>Rep of Moldova</td>
<td>0 (0.00)</td>
<td>52 (98.11)</td>
</tr>
<tr>
<td>The Netherlands¹</td>
<td>7 (0.50)</td>
<td>1266 (90.82)</td>
</tr>
<tr>
<td>North Macedonia</td>
<td>0 (0.00)</td>
<td>128 (99.22)</td>
</tr>
<tr>
<td>Norway</td>
<td>2 (0.72)</td>
<td>271 (97.84)</td>
</tr>
<tr>
<td>Poland</td>
<td>29 (2.47)</td>
<td>1122 (95.57)</td>
</tr>
<tr>
<td>Portugal</td>
<td>5 (1.53)</td>
<td>313 (96.02)</td>
</tr>
<tr>
<td>Romania</td>
<td>1 (0.44)</td>
<td>224 (99.12)</td>
</tr>
<tr>
<td>Russian Federation</td>
<td>62 (1.99)</td>
<td>2987 (95.86)</td>
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<tr>
<td>Serbia</td>
<td>1 (0.55)</td>
<td>177 (96.72)</td>
</tr>
<tr>
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<td>3 (1.08)</td>
<td>262 (93.90)</td>
</tr>
<tr>
<td>Slovenia</td>
<td>2 (2.08)</td>
<td>92 (95.84)</td>
</tr>
<tr>
<td>Spain</td>
<td>24 (1.14)</td>
<td>2019 (95.64)</td>
</tr>
<tr>
<td>Sweden</td>
<td>11 (1.79)</td>
<td>585 (95.28)</td>
</tr>
<tr>
<td>Switzerland</td>
<td>6 (0.66)</td>
<td>868 (95.70)</td>
</tr>
<tr>
<td>Turkey</td>
<td>26 (1.31)</td>
<td>1913 (96.52)</td>
</tr>
<tr>
<td>Ukraine</td>
<td>2 (1.32)</td>
<td>147 (97.36)</td>
</tr>
<tr>
<td>United Kingdom²</td>
<td>0 (0.00)</td>
<td>8957 (92.35)</td>
</tr>
<tr>
<td>Total</td>
<td>607 (1.32)</td>
<td>42974 (93.56)</td>
</tr>
</tbody>
</table>

¹ The Netherlands: only diabetes treated with daily insulin is recorded.
² United Kingdom: for ABPA, clinician reported aspergillosis.

**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.

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.

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.
### 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.

<table>
<thead>
<tr>
<th>Country</th>
<th>Pneumothorax this year number (%)</th>
<th>Haemoptysis major over 250 ml this year number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Missing/unknown</td>
<td>No</td>
</tr>
<tr>
<td>Albania</td>
<td>1 (0.94)</td>
<td>105</td>
</tr>
<tr>
<td>Armenia¹</td>
<td>0 (0.00)</td>
<td>22</td>
</tr>
<tr>
<td>Austria</td>
<td>6 (0.84)</td>
<td>711</td>
</tr>
<tr>
<td>Belarus</td>
<td>0 (0.00)</td>
<td>155</td>
</tr>
<tr>
<td>Belgium</td>
<td>23 (2.06)</td>
<td>1087</td>
</tr>
<tr>
<td>Bulgaria</td>
<td>2 (1.08)</td>
<td>184</td>
</tr>
<tr>
<td>Croatia</td>
<td>1 (0.76)</td>
<td>129</td>
</tr>
<tr>
<td>Cyprus</td>
<td>1 (3.70)</td>
<td>26</td>
</tr>
<tr>
<td>Czech Republic</td>
<td>11 (1.85)</td>
<td>580</td>
</tr>
<tr>
<td>Denmark</td>
<td>1 (0.21)</td>
<td>467</td>
</tr>
<tr>
<td>France</td>
<td>0 (0.00)</td>
<td>5807</td>
</tr>
<tr>
<td>Georgia</td>
<td>1 (1.39)</td>
<td>70</td>
</tr>
<tr>
<td>Germany²</td>
<td>60 (1.01)</td>
<td>5832</td>
</tr>
<tr>
<td>Greece</td>
<td>12 (3.01)</td>
<td>385</td>
</tr>
<tr>
<td>Hungary</td>
<td>6 (1.35)</td>
<td>435</td>
</tr>
<tr>
<td>Ireland¹</td>
<td>2 (0.18)</td>
<td>1136</td>
</tr>
<tr>
<td>Israel</td>
<td>16 (3.12)</td>
<td>497</td>
</tr>
<tr>
<td>Italy</td>
<td>77 (1.46)</td>
<td>5192</td>
</tr>
<tr>
<td>Latvia</td>
<td>2 (4.44)</td>
<td>43</td>
</tr>
<tr>
<td>Lithuania</td>
<td>0 (0.00)</td>
<td>27</td>
</tr>
<tr>
<td>Luxembourg</td>
<td>0 (0.00)</td>
<td>36</td>
</tr>
<tr>
<td>Rep of Moldova</td>
<td>0 (0.00)</td>
<td>53</td>
</tr>
</tbody>
</table>

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]

<table>
<thead>
<tr>
<th>Country</th>
<th>Missing/unknown</th>
<th>No</th>
<th>Pneumothorax this year number (%)</th>
<th>Haemoptysis major over 250 ml this year number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td>Yes, chest drain</td>
<td>Yes, observation only</td>
</tr>
<tr>
<td>The Netherlands</td>
<td>5 (0.36)</td>
<td>1385</td>
<td>4 (0.29)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>North Macedonia</td>
<td>0 (0)</td>
<td>129</td>
<td>0 (0.00)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Norway</td>
<td>1 (0.36)</td>
<td>274</td>
<td>1 (0.36)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Poland</td>
<td>31 (2.64)</td>
<td>1141</td>
<td>2 (0.00)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Portugal</td>
<td>7 (2.15)</td>
<td>316</td>
<td>2 (0.00)</td>
<td>1 (0.00)</td>
</tr>
<tr>
<td>Romania</td>
<td>3 (1.33)</td>
<td>221</td>
<td>1 (0.44)</td>
<td>1 (0.44)</td>
</tr>
<tr>
<td>Russian Federation</td>
<td>17 (0.55)</td>
<td>3077</td>
<td>22 (0.71)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Serbia</td>
<td>1 (0.55)</td>
<td>182</td>
<td>0 (0.00)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Slovak Republic</td>
<td>6 (2.15)</td>
<td>272</td>
<td>1 (0.36)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Slovenia</td>
<td>0 (0)</td>
<td>96</td>
<td>0 (0.00)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Spain</td>
<td>36 (1.71)</td>
<td>2074</td>
<td>1 (0.05)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Sweden</td>
<td>11 (1.79)</td>
<td>603</td>
<td>0 (0.00)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Switzerland</td>
<td>6 (0.66)</td>
<td>899</td>
<td>2 (0.22)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Turkey</td>
<td>7 (0.35)</td>
<td>1969</td>
<td>6 (0.30)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Ukraine</td>
<td>2 (1.32)</td>
<td>148</td>
<td>1 (0.00)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>United Kingdom(^4)</td>
<td>0 (0.00)</td>
<td>9676</td>
<td>23 (0.24)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Total</td>
<td>355 (0.77)</td>
<td>45441</td>
<td>105 (0.23)</td>
<td>22 (0.05)</td>
</tr>
</tbody>
</table>

\(^4\) United Kingdom: defines haemoptysis major over 250 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.

<table>
<thead>
<tr>
<th>Country</th>
<th>Missing/unknown</th>
<th>No liver disease</th>
<th>Cirrhosis with portal hypertension/ hypersplenism</th>
<th>Cirrhosis no portal hypertension/ hypersplenism</th>
<th>Cirrhosis. portal hypertension unknown</th>
<th>Liver disease without cirrhosis</th>
<th>Variceal bleeding</th>
</tr>
</thead>
<tbody>
<tr>
<td>Albania</td>
<td>1 (0.94)</td>
<td>52 (49.06)</td>
<td>2 (1.89)</td>
<td>0 (0.00)</td>
<td>0 (0.00)</td>
<td>51 (48.11)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Armenia</td>
<td>0 (0.00)</td>
<td>13 (52.00)</td>
<td>0 (0.00)</td>
<td>1 (4.00)</td>
<td>0 (0.00)</td>
<td>11 (44.00)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Austria</td>
<td>4 (0.56)</td>
<td>363 (50.55)</td>
<td>13 (1.81)</td>
<td>20 (2.79)</td>
<td>2 (0.28)</td>
<td>316 (44.01)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Belarus</td>
<td>0 (0.00)</td>
<td>96 (61.94)</td>
<td>4 (2.58)</td>
<td>2 (1.29)</td>
<td>0 (0.00)</td>
<td>53 (34.19)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Belgium¹</td>
<td>2 (0.18)</td>
<td>1069 (95.96)</td>
<td>43 (3.86)</td>
<td>0 (0.00)</td>
<td>0 (0.00)</td>
<td>46 (0.00)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Bulgaria</td>
<td>1 (0.54)</td>
<td>129 (69.35)</td>
<td>4 (2.15)</td>
<td>6 (3.23)</td>
<td>0 (0.00)</td>
<td>46 (24.73)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Croatia</td>
<td>1 (0.76)</td>
<td>114 (87.03)</td>
<td>4 (3.05)</td>
<td>3 (2.29)</td>
<td>1 (0.76)</td>
<td>8 (6.11)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Cyprus</td>
<td>0 (0.00)</td>
<td>25 (92.60)</td>
<td>1 (3.70)</td>
<td>0 (0.00)</td>
<td>0 (0.00)</td>
<td>1 (3.70)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Czech Republic</td>
<td>0 (0.00)</td>
<td>483 (81.45)</td>
<td>10 (1.69)</td>
<td>7 (1.18)</td>
<td>1 (0.17)</td>
<td>92 (15.51)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Denmark</td>
<td>2 (0.43)</td>
<td>386 (82.30)</td>
<td>19 (4.05)</td>
<td>9 (1.92)</td>
<td>2 (0.43)</td>
<td>51 (10.87)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>France</td>
<td>0 (0.00)</td>
<td>4914 (84.48)</td>
<td>110 (1.89)</td>
<td>121 (2.08)</td>
<td>0 (0.00)</td>
<td>672 (11.55)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Georgia</td>
<td>2 (0.28)</td>
<td>65 (90.27)</td>
<td>0 (0.00)</td>
<td>3 (4.17)</td>
<td>0 (0.00)</td>
<td>2 (2.78)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Germany²</td>
<td>360 (6.08)</td>
<td>4065 (68.60)</td>
<td>128 (2.16)</td>
<td>84 (1.42)</td>
<td>83 (1.40)</td>
<td>1205 (20.34)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Greece</td>
<td>17 (4.26)</td>
<td>249 (62.41)</td>
<td>16 (4.01)</td>
<td>3 (0.75)</td>
<td>2 (0.50)</td>
<td>112 (28.07)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Hungary</td>
<td>7 (1.57)</td>
<td>318 (71.46)</td>
<td>87 (19.55)</td>
<td>14 (3.15)</td>
<td>16 (3.60)</td>
<td>3 (0.67)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Ireland</td>
<td>2 (0.18)</td>
<td>1000 (87.86)</td>
<td>29 (2.55)</td>
<td>7 (0.62)</td>
<td>9 (0.79)</td>
<td>91 (8.00)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Israel</td>
<td>15 (2.92)</td>
<td>400 (77.98)</td>
<td>9 (1.75)</td>
<td>7 (1.36)</td>
<td>0 (0.00)</td>
<td>81 (15.80)</td>
<td>1 (0.19)</td>
</tr>
<tr>
<td>Italy</td>
<td>27 (0.51)</td>
<td>3652 (69.24)</td>
<td>72 (1.36)</td>
<td>37 (0.70)</td>
<td>8 (0.15)</td>
<td>1479 (28.04)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Latvia</td>
<td>2 (0.44)</td>
<td>27 (60.01)</td>
<td>2 (4.44)</td>
<td>0 (0.00)</td>
<td>0 (0.00)</td>
<td>14 (31.11)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Lithuania</td>
<td>0 (0.00)</td>
<td>26 (96.30)</td>
<td>1 (3.70)</td>
<td>0 (0.00)</td>
<td>0 (0.00)</td>
<td>0 (0.00)</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Luxembourg</td>
<td>0 (0.00)</td>
<td>28 (77.78)</td>
<td>3 (8.33)</td>
<td>0 (0.00)</td>
<td>0 (0.00)</td>
<td>5 (13.89)</td>
<td>0 (0.00)</td>
</tr>
</tbody>
</table>

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.
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.

<table>
<thead>
<tr>
<th>Country</th>
<th>Missing/unknown</th>
<th>No liver disease</th>
<th>Liver disease this year number (%)</th>
<th>Liver disease this year number (%)</th>
<th>Liver disease this year number (%)</th>
<th>Liver disease this year number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Cirrhosis</td>
<td>Cirrhosis</td>
<td>Cirrhosis</td>
<td>Cirrhosis</td>
<td>Variceal bleeding</td>
</tr>
<tr>
<td></td>
<td></td>
<td>with portal</td>
<td>hypertension/hypersplenism</td>
<td>no portal</td>
<td>hypertension/</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>hypertension</td>
<td></td>
<td></td>
<td>hypersplenism unknown</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Rep of Moldova</td>
<td>0.00</td>
<td>45 (84.90)</td>
<td>0</td>
<td>1</td>
<td>0</td>
<td>7</td>
</tr>
<tr>
<td>The Netherlands</td>
<td>0.14</td>
<td>1044 (74.89)</td>
<td>97</td>
<td>35</td>
<td>4</td>
<td>212</td>
</tr>
<tr>
<td>North Macedonia</td>
<td>0.00</td>
<td>67 (51.94)</td>
<td>6</td>
<td>14</td>
<td>0</td>
<td>42</td>
</tr>
<tr>
<td>Norway</td>
<td>0.00</td>
<td>246 (188.80)</td>
<td>5</td>
<td>0</td>
<td>4</td>
<td>17</td>
</tr>
<tr>
<td>Poland</td>
<td>0.00</td>
<td>701 (59.71)</td>
<td>58</td>
<td>10</td>
<td>5</td>
<td>371</td>
</tr>
<tr>
<td>Portugal</td>
<td>0.00</td>
<td>254 (77.92)</td>
<td>6</td>
<td>0</td>
<td>0</td>
<td>60</td>
</tr>
<tr>
<td>Romania</td>
<td>0.00</td>
<td>165 (73.01)</td>
<td>9</td>
<td>4</td>
<td>3</td>
<td>44</td>
</tr>
<tr>
<td>Russian Federation</td>
<td>0.00</td>
<td>2456 (78.81)</td>
<td>149</td>
<td>72</td>
<td>8</td>
<td>404</td>
</tr>
<tr>
<td>Serbia</td>
<td>0.00</td>
<td>117 (63.94)</td>
<td>7</td>
<td>2</td>
<td>2</td>
<td>53</td>
</tr>
<tr>
<td>Slovak Republic</td>
<td>0.00</td>
<td>123 (44.09)</td>
<td>16</td>
<td>7</td>
<td>2</td>
<td>127</td>
</tr>
<tr>
<td>Slovenia</td>
<td>0.00</td>
<td>61 (63.54)</td>
<td>7</td>
<td>3</td>
<td>0</td>
<td>24</td>
</tr>
<tr>
<td>Spain</td>
<td>1.47</td>
<td>1661 (78.69)</td>
<td>30</td>
<td>10</td>
<td>2</td>
<td>376</td>
</tr>
<tr>
<td>Sweden</td>
<td>0.00</td>
<td>493 (80.30)</td>
<td>12</td>
<td>10</td>
<td>0</td>
<td>87</td>
</tr>
<tr>
<td>Switzerland</td>
<td>0.00</td>
<td>664 (73.21)</td>
<td>29</td>
<td>9</td>
<td>2</td>
<td>189</td>
</tr>
<tr>
<td>Turkey</td>
<td>0.00</td>
<td>1729 (87.24)</td>
<td>23</td>
<td>7</td>
<td>4</td>
<td>212</td>
</tr>
<tr>
<td>Ukraine</td>
<td>0.00</td>
<td>35 (23.19)</td>
<td>11</td>
<td>6</td>
<td>2</td>
<td>95</td>
</tr>
<tr>
<td>United Kingdom</td>
<td>0.00</td>
<td>8310 (85.68)</td>
<td>125</td>
<td>63</td>
<td>0</td>
<td>1201</td>
</tr>
<tr>
<td>Total</td>
<td>0.00</td>
<td>35645 (77.6)</td>
<td>1147</td>
<td>577</td>
<td>162</td>
<td>7814</td>
</tr>
</tbody>
</table>

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.
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.**

<table>
<thead>
<tr>
<th>Country</th>
<th>Ursodeoxycholic acid this year number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Missing/unknown</td>
</tr>
<tr>
<td>Albania</td>
<td></td>
</tr>
<tr>
<td></td>
<td>(1.89)</td>
</tr>
<tr>
<td>Armenia</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>(0.00)</td>
</tr>
<tr>
<td>Austria</td>
<td>4</td>
</tr>
<tr>
<td></td>
<td>(0.56)</td>
</tr>
<tr>
<td>Belarus</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>(0.00)</td>
</tr>
<tr>
<td>Belgium</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>(0.09)</td>
</tr>
<tr>
<td>Bulgaria</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>(0.00)</td>
</tr>
<tr>
<td>Croatia</td>
<td>4</td>
</tr>
<tr>
<td></td>
<td>(3.05)</td>
</tr>
<tr>
<td>Cyprus</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>(3.70)</td>
</tr>
<tr>
<td>Czech Republic</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>(0.00)</td>
</tr>
<tr>
<td>Denmark</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>(0.00)</td>
</tr>
<tr>
<td>France</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>(0.00)</td>
</tr>
<tr>
<td>Georgia</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>(1.39)</td>
</tr>
<tr>
<td>Germany</td>
<td>43</td>
</tr>
<tr>
<td></td>
<td>(0.73)</td>
</tr>
<tr>
<td>Greece</td>
<td>12</td>
</tr>
<tr>
<td></td>
<td>(3.01)</td>
</tr>
<tr>
<td>Hungary</td>
<td>7</td>
</tr>
<tr>
<td></td>
<td>(1.57)</td>
</tr>
<tr>
<td>Ireland</td>
<td>2</td>
</tr>
<tr>
<td></td>
<td>(0.18)</td>
</tr>
<tr>
<td>Israel</td>
<td>6</td>
</tr>
<tr>
<td></td>
<td>(1.17)</td>
</tr>
<tr>
<td>Italy</td>
<td>15</td>
</tr>
<tr>
<td></td>
<td>(0.28)</td>
</tr>
<tr>
<td>Latvia</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>(0.00)</td>
</tr>
<tr>
<td>Lithuania</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>(0.00)</td>
</tr>
<tr>
<td>Luxembourg</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>(0.00)</td>
</tr>
<tr>
<td>Rep of Moldova</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td>(0.00)</td>
</tr>
<tr>
<td>Country</td>
<td>Ursodeoxycholic acid this year number (%)</td>
</tr>
<tr>
<td>-------------------------</td>
<td>------------------------------------------</td>
</tr>
<tr>
<td></td>
<td>Missing/unknown</td>
</tr>
<tr>
<td>The Netherlands</td>
<td>5 (0.36)</td>
</tr>
<tr>
<td>North Macedonia</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Norway</td>
<td>9 (3.25)</td>
</tr>
<tr>
<td>Poland</td>
<td>9 (0.77)</td>
</tr>
<tr>
<td>Portugal</td>
<td>3 (0.92)</td>
</tr>
<tr>
<td>Romania</td>
<td>5 (2.21)</td>
</tr>
<tr>
<td>Russian Federation</td>
<td>27 (0.87)</td>
</tr>
<tr>
<td>Serbia</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Slovak Republic</td>
<td>4 (1.43)</td>
</tr>
<tr>
<td>Slovenia</td>
<td>4 (4.17)</td>
</tr>
<tr>
<td>Spain</td>
<td>30 (1.42)</td>
</tr>
<tr>
<td>Sweden</td>
<td>5 (0.81)</td>
</tr>
<tr>
<td>Switzerland</td>
<td>5 (0.55)</td>
</tr>
<tr>
<td>Turkey</td>
<td>3 (0.15)</td>
</tr>
<tr>
<td>Ukraine</td>
<td>2 (1.32)</td>
</tr>
<tr>
<td>United Kingdom</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Total</td>
<td>209 (0.46)</td>
</tr>
</tbody>
</table>

Note: Oral ursodeoxycholic 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.

<table>
<thead>
<tr>
<th>Country</th>
<th>Malignancy occurred this year number (%)</th>
<th>DIOS this year number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Missing/unknown</td>
<td>No</td>
</tr>
<tr>
<td>Albania</td>
<td>3 (2.83)</td>
<td>103</td>
</tr>
<tr>
<td>Armenia</td>
<td>0 (0)</td>
<td>25</td>
</tr>
<tr>
<td>Austria</td>
<td>5 (0.70)</td>
<td>713</td>
</tr>
<tr>
<td>Belarus</td>
<td>0 (0)</td>
<td>155</td>
</tr>
<tr>
<td>Belgium</td>
<td>0 (0.00)</td>
<td>1111</td>
</tr>
<tr>
<td>Bulgaria</td>
<td>2 (1.08)</td>
<td>183</td>
</tr>
<tr>
<td>Croatia</td>
<td>1 (0.76)</td>
<td>130</td>
</tr>
<tr>
<td>Cyprus</td>
<td>0 (0)</td>
<td>27</td>
</tr>
<tr>
<td>Czech Republic</td>
<td>7 (1.18)</td>
<td>585</td>
</tr>
<tr>
<td>Denmark¹</td>
<td>2 (0.43)</td>
<td>464</td>
</tr>
<tr>
<td>France</td>
<td>5 (0.09)</td>
<td>5795</td>
</tr>
<tr>
<td>Georgia</td>
<td>1 (1.39)</td>
<td>71</td>
</tr>
<tr>
<td>Germany</td>
<td>77 (1.30)</td>
<td>5805</td>
</tr>
<tr>
<td>Greece</td>
<td>15 (3.76)</td>
<td>377</td>
</tr>
<tr>
<td>Hungary</td>
<td>7 (1.57)</td>
<td>435</td>
</tr>
<tr>
<td>Ireland</td>
<td>2 (0.18)</td>
<td>1135</td>
</tr>
<tr>
<td>Israel</td>
<td>15 (2.92)</td>
<td>494</td>
</tr>
<tr>
<td>Italy</td>
<td>63 (1.19)</td>
<td>5195</td>
</tr>
<tr>
<td>Latvia</td>
<td>2 (4.44)</td>
<td>43</td>
</tr>
<tr>
<td>Lithuania</td>
<td>0 (0)</td>
<td>27</td>
</tr>
<tr>
<td>Luxembourg</td>
<td>0 (0.00)</td>
<td>35</td>
</tr>
<tr>
<td>Rep of Moldova</td>
<td>0 (0)</td>
<td>53</td>
</tr>
</tbody>
</table>

¹ Denmark has only reported DIOS requiring hospitalisation.
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.

<table>
<thead>
<tr>
<th>Country</th>
<th>Hypertonic saline (NaCl) inhaled &gt; 3 months this year number (%)</th>
<th>rhDNase inhaled &gt; 3 months this year number (%)</th>
<th>Mannitol inhaled &gt; 3 months this year number (%)</th>
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<td>Missing/unknown No Yes</td>
<td>Missing/unknown No Yes</td>
</tr>
<tr>
<td>Albania</td>
<td>8 (7.55) 0 (0.00) 98 (92.45)</td>
<td>3 (2.83) 96 (90.57) 7 (6.60)</td>
<td>2 (1.89) 104 (98.11) 0 (0.00)</td>
</tr>
<tr>
<td>Armenia</td>
<td>0 (0.00) 0 (0.00) 25 (100)</td>
<td>0 (0.00) 19 (76.00) 6 (24.00)</td>
<td>0 (0.00) 25 (100) 0 (0)</td>
</tr>
<tr>
<td>Austria</td>
<td>1 (0.14) 142 (19.78) 575 (80.08)</td>
<td>1 (0.14) 327 (45.54) 390 (54.32)</td>
<td>1 (0.14) 708 (98.61) 9 (1.25)</td>
</tr>
<tr>
<td>Belarus</td>
<td>0 (0.00) 54 (34.84) 101 (65.16)</td>
<td>0 (0.00) 150 (96.77) 5 (3.23)</td>
<td>0 (0.00) 155 (100) 0 (0)</td>
</tr>
<tr>
<td>Belgium</td>
<td>37 (3.32) 333 (29.89) 744 (66.79)</td>
<td>37 (3.32) 130 (11.67) 947 (85.01)</td>
<td>1114 (0.00) 0 (100) 0 (0)</td>
</tr>
<tr>
<td>Bulgaria</td>
<td>0 (0.00) 96 (51.61) 90 (48.39)</td>
<td>0 (0.00) 28 (15.05) 158 (84.95)</td>
<td>0 (0.00) 186 (100) 0 (0)</td>
</tr>
<tr>
<td>Croatia</td>
<td>2 (1.53) 16 (12.21) 113 (86.26)</td>
<td>5 (3.82) 37 (28.24) 89 (67.94)</td>
<td>5 (3.82) 125 (95.42) 1 (0.76)</td>
</tr>
<tr>
<td>Cyprus</td>
<td>1 (3.70) 22 (81.49) 4 (14.81)</td>
<td>1 (3.70) 7 (25.93) 19 (70.37)</td>
<td>1 (3.70) 26 (96.30) 0 (0.00)</td>
</tr>
<tr>
<td>Czech Republic</td>
<td>0 (0.00) 119 (20.07) 474 (79.93)</td>
<td>0 (0.00) 196 (33.05) 397 (66.95)</td>
<td>0 (0.00) 586 (100) 7 (1.18)</td>
</tr>
<tr>
<td>Denmark</td>
<td>469 (100) 0 (0) 0 (0)</td>
<td>0 (0.00) 40 (8.53) 429 (91.47)</td>
<td>469 (100) 0 (0) 0 (0)</td>
</tr>
<tr>
<td>France</td>
<td>0 (0.00) 5011 (86.14) 806 (13.86)</td>
<td>0 (0.00) 3116 (53.57) 2701 (46.43)</td>
<td>5817 (100) 0 (0) 0 (0)</td>
</tr>
<tr>
<td>Georgia</td>
<td>0 (0.00) 48 (66.67) 24 (33.33)</td>
<td>0 (0.00) 72 (100) 0 (0)</td>
<td>0 (0.00) 72 (100) 0 (0)</td>
</tr>
<tr>
<td>Germany</td>
<td>46 (0.78) 1180 (19.92) 4699 (79.30)</td>
<td>47 (0.79) 2749 (46.40) 3129 (52.81)</td>
<td>58 (0.98) 5636 (95.12) 231 (3.90)</td>
</tr>
<tr>
<td>Greece</td>
<td>6 (1.50) 291 (72.94) 102 (25.56)</td>
<td>4 (1.00) 117 (29.33) 278 (69.67)</td>
<td>8 (2.01) 381 (95.48) 10 (2.51)</td>
</tr>
<tr>
<td>Hungary</td>
<td>9 (2.02) 80 (17.98) 356 (80.00)</td>
<td>9 (2.02) 133 (29.89) 303 (68.99)</td>
<td>445 (100) 0 (0) 0 (0)</td>
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<tr>
<td>Ireland</td>
<td>2 (0.18) 466 (40.95) 670 (58.87)</td>
<td>2 (0.18) 496 (43.58) 640 (56.24)</td>
<td>1138 (100) 0 (0) 0 (0)</td>
</tr>
<tr>
<td>Israel</td>
<td>6 (1.17) 118 (23.00) 389 (75.83)</td>
<td>3 (0.58) 123 (23.98) 387 (75.44)</td>
<td>2 (0.39) 510 (99.42) 1 (0.19)</td>
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<td>Italy</td>
<td>15 (0.28) 2886 (54.72) 2374 (45.00)</td>
<td>15 (0.28) 3020 (57.26) 2240 (42.46)</td>
<td>1493 (28.30) 3707 (70.28) 75 (1.42)</td>
</tr>
<tr>
<td>Latvia</td>
<td>0 (0.00) 4 (8.89) 41 (91.11)</td>
<td>0 (0.00) 25 (55.66) 20 (44.44)</td>
<td>0 (0.00) 45 (100) 0 (0)</td>
</tr>
<tr>
<td>Lithuania</td>
<td>0 (0.00) 22 (81.48) 5 (18.52)</td>
<td>0 (0.00) 4 (14.81) 23 (85.19)</td>
<td>0 (0.00) 27 (100) 0 (0)</td>
</tr>
<tr>
<td>Luxembourg</td>
<td>0 (0.00) 3 (8.33) 33 (91.67)</td>
<td>0 (0.00) 10 (27.78) 26 (72.22)</td>
<td>0 (0.00) 35 (100) 1 (0)</td>
</tr>
<tr>
<td>Rep of Moldova</td>
<td>0 (0.00) 3 (5.66) 50 (94.34)</td>
<td>0 (0.00) 52 (98.11) 1 (1.89)</td>
<td>0 (0.00) 53 (100) 0 (0)</td>
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</table>
**[table 7.6 continued]**

<table>
<thead>
<tr>
<th>Country</th>
<th>Hypertonic saline (NaCl) inhaled &gt; 3 months this year number (%)</th>
<th>rhDNase inhaled &gt; 3 months this year number (%)</th>
<th>Mannitol inhaled &gt; 3 months this year number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Missing/unknown No Yes</td>
<td>Missing/unknown No Yes</td>
<td>Missing/unknown No Yes</td>
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<tr>
<td>The Netherlands</td>
<td>2 (0.14) 902 (35.15) 490</td>
<td>3 (0.22) 482 (65.21) 909</td>
<td>1394 0 0</td>
</tr>
<tr>
<td>North Macedonia</td>
<td>0 (0.00) 40 89</td>
<td>0 (0.00) 41 88</td>
<td>3 126 0</td>
</tr>
<tr>
<td>Norway</td>
<td>3 (1.08) 57 217</td>
<td>0 (0.00) 94 183</td>
<td>2 275 0</td>
</tr>
<tr>
<td>Poland</td>
<td>5 (0.43) 143 1026</td>
<td>4 (0.34) 170 1000</td>
<td>8 1164 2</td>
</tr>
<tr>
<td>Portugal</td>
<td>2 (0.61) 197 127</td>
<td>3 (0.92) 50 273</td>
<td>3 322 1</td>
</tr>
<tr>
<td>Romania</td>
<td>1 (0.44) 59 166</td>
<td>1 (0.92) 45 180</td>
<td>1 225 0</td>
</tr>
<tr>
<td>Russian Federation</td>
<td>56 (1.80) 856 2204</td>
<td>22 (0.71) 117 2977</td>
<td>27 2991 98</td>
</tr>
<tr>
<td>Serbia</td>
<td>0 (0.00) 7 176</td>
<td>0 (0.00) 49 134</td>
<td>0 183 0</td>
</tr>
<tr>
<td>Slovak Republic</td>
<td>0 (0.00) 151 128</td>
<td>3 (0.71) 87 189</td>
<td>4 275 0</td>
</tr>
<tr>
<td>Slovenia</td>
<td>0 (0.00) 4 92</td>
<td>1 (0.92) 65 30</td>
<td>3 92 1</td>
</tr>
<tr>
<td>Spain</td>
<td>8 (0.38) 728 1375</td>
<td>8 (0.38) 1293 810</td>
<td>10 2093 8</td>
</tr>
<tr>
<td>Sweden</td>
<td>6 (0.98) 103 505</td>
<td>7 (1.14) 419 188</td>
<td>0 608 6</td>
</tr>
<tr>
<td>Switzerland</td>
<td>5 (0.55) 200 702</td>
<td>3 (0.33) 466 438</td>
<td>6 900 1</td>
</tr>
<tr>
<td>Turkey</td>
<td>4 (0.20) 1630 348</td>
<td>4 (0.20) 172 1806</td>
<td>4 1882 96</td>
</tr>
<tr>
<td>Ukraine</td>
<td>3 (1.99) 1 147</td>
<td>3 (1.99) 19 129</td>
<td>2 149 0</td>
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<tr>
<td>United Kingdom¹</td>
<td>0 (0.00) 6309 3390</td>
<td>0 (0.00) 3002 6697</td>
<td>0 9359 340</td>
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<tr>
<td>Total</td>
<td>697 (1.52) 22281 22955</td>
<td>189 (0.41) 17518 28226</td>
<td>192 (3.51) 0</td>
</tr>
</tbody>
</table>

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.
- 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.
- 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.
- 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.

<table>
<thead>
<tr>
<th>Country</th>
<th>Antibiotics inhaled &gt; 3 months this year number (%)</th>
<th>Bronchodilators inhaled &gt; 3 months this year number (%)</th>
<th>Macrolides &gt; 3 months this year number (%)</th>
</tr>
</thead>
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<td>Missing/unknown No Yes Missing/unknown No Yes Missing/unknown No Yes</td>
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<td></td>
</tr>
<tr>
<td>Albania</td>
<td>(1.89) 68.86 (29.25) 1 (0.94) (15.09) (83.97) (2.83) (80.19) (16.98)</td>
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<td></td>
</tr>
<tr>
<td>Armenia</td>
<td>(0.00) 60.00 (40.00) 0 (0.00) (4.00) (96.00) (0.00) (56.00) (44.00)</td>
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<td></td>
</tr>
<tr>
<td>Austria</td>
<td>(0.14) 61.84 (38.02) 0 (0.00) (3.34) (96.66) (0.14) (94.57) (5.29)</td>
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<td></td>
</tr>
<tr>
<td>Belarus</td>
<td>(0.00) 65.81 (34.19) 0 (0.00) (64.52) (35.48) (0.00) (72.26) (27.74)</td>
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<td></td>
</tr>
<tr>
<td>Belgium</td>
<td>(0.18) 47.04 (52.78) 37 (0.00) (18.85) (77.83) (0.09) (46.23) (53.68)</td>
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<td></td>
</tr>
<tr>
<td>Bulgaria</td>
<td>(0.00) 52.15 (47.85) 0 (0.00) (74.19) (25.81) (0.00) (94.09) (5.91)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Croatia</td>
<td>(4.58) 47.33 (48.09) 5 (3.82) (73.28) (22.90) (3.82) (61.83) (34.35)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cyprus</td>
<td>(3.70) 62.97 (33.33) 1 (3.70) (55.56) (40.74) (3.70) (51.86) (44.44)</td>
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<td></td>
</tr>
<tr>
<td>Czech Republic</td>
<td>(0.00) 77.23 (22.77) 0 (0.00) (41.65) (58.35) (0.00) (94.44) (5.56)</td>
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<td></td>
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<td>Denmark</td>
<td>(0.43) 90.19 (9.38) (100) (0) (0.21) (73.14) (26.65)</td>
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<tr>
<td>France</td>
<td>(0.00) 58.02 (41.98) 0 (0.00) (33.87) (66.13) (0.00) (65.76) (34.24)</td>
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</tr>
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<td>Georgia</td>
<td>(0.00) 98.61 (1.39) 0 (0.00) (100) (0) (0.00) (80.56) (19.44)</td>
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<td></td>
</tr>
<tr>
<td>Germany</td>
<td>(1.45) 56.78 (41.77) 0.81 (18.78) (80.41) (1.05) (83.69) (15.26)</td>
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<td></td>
</tr>
<tr>
<td>Greece</td>
<td>(1.25) 43.36 (55.39) 0.75 (37.84) (61.41) (1.75) (68.68) (29.57)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hungary</td>
<td>(1.57) 46.97 (51.46) 0.25 (37.98) (59.77) (3.15) (67.19) (29.66)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ireland</td>
<td>(0.26) 57.38 (42.36) 0.18 (26.36) (73.46) (0.18) (52.72) (47.10)</td>
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</tr>
<tr>
<td>Israel</td>
<td>(0.58) 46.20 (53.22) 1.36 (38.79) (53.85) (0.97) (54.98) (44.05)</td>
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</tr>
<tr>
<td>Italy</td>
<td>(0.27) 62.57 (37.16) 0.27 (25.71) (74.02) (0.34) (72.17) (27.49)</td>
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<tr>
<td>Latvia</td>
<td>(0.00) 73.33 (26.67) 0 (0.00) (4.44) (95.56) (0.00) (91.11) (8.89)</td>
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<td></td>
</tr>
<tr>
<td>Lithuania</td>
<td>(3.70) 74.07 (22.22) 3.70 (51.86) (44.44) (0.00) (96.30) (3.70)</td>
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</tr>
<tr>
<td>Luxembourg</td>
<td>(0.00) 52.78 (47.22) 0 (0.00) (13.89) (86.11) (2.78) (72.22) (25.00)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Rep of Moldova</td>
<td>(0.00) 37.74 (62.26) 0 (0.00) (83.02) (16.98) (0.00) (86.79) (13.21)</td>
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</table>
### Table 7.7 continued

<table>
<thead>
<tr>
<th>Country</th>
<th>Antibiotics inhaled &gt; 3 months this year number (%)</th>
<th>Bronchodilators inhaled &gt; 3 months this year number (%)</th>
<th>Macrolides &gt; 3 months this year number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Missing/unknown No Yes</td>
<td>Missing/unknown No Yes</td>
<td>Missing/unknown No Yes</td>
</tr>
<tr>
<td>The Netherlands</td>
<td>1 841 552 (0.07) (60.33) (36.90)</td>
<td>0 598 766 (0.07) (42.90) (57.10)</td>
<td>1 885 508 (0.07) (63.49) (36.44)</td>
</tr>
<tr>
<td>North Macedonia</td>
<td>0 62 67 (0.00) (48.06) (51.94)</td>
<td>0 9 120 (0.00) (6.98) (93.02)</td>
<td>0 102 27 (0.00) (79.07) (20.93)</td>
</tr>
<tr>
<td>Norway</td>
<td>19 210 48 (6.86) (75.81) (17.33)</td>
<td>3 55 219 (1.08) (19.86) (79.06)</td>
<td>7 231 39 (2.53) (83.39) (14.08)</td>
</tr>
<tr>
<td>Poland</td>
<td>9 803 362 (0.77) (68.40) (30.83)</td>
<td>9 257 908 (0.77) (21.89) (77.34)</td>
<td>15 955 204 (1.28) (81.34) (17.38)</td>
</tr>
<tr>
<td>Portugal</td>
<td>5 158 163 (1.53) (48.47) (50.00)</td>
<td>1 134 191 (0.31) (41.10) (58.59)</td>
<td>3 215 108 (0.92) (65.95) (33.13)</td>
</tr>
<tr>
<td>Romania</td>
<td>1 135 90 (0.44) (59.74) (39.82)</td>
<td>2 136 88 (0.88) (60.18) (38.94)</td>
<td>1 197 28 (0.44) (87.17) (12.39)</td>
</tr>
<tr>
<td>Russian Federation</td>
<td>54 1637 1425 (1.73) (52.54) (45.73)</td>
<td>56 1523 1537 (1.80) (48.88) (49.32)</td>
<td>72 2120 924 (2.31) (68.04) (29.65)</td>
</tr>
<tr>
<td>Serbia</td>
<td>0 93 90 (0.00) (50.82) (49.18)</td>
<td>0 1 182 (0.00) (0.55) (99.45)</td>
<td>0 161 22 (0.00) (87.98) (12.02)</td>
</tr>
<tr>
<td>Slovak Republic</td>
<td>1 118 160 (0.36) (42.29) (57.35)</td>
<td>0 131 148 (0.00) (46.95) (53.05)</td>
<td>4 148 127 (1.43) (53.05) (45.52)</td>
</tr>
<tr>
<td>Slovenia</td>
<td>3 68 25 (3.13) (70.83) (26.04)</td>
<td>3 70 23 (3.13) (72.91) (23.96)</td>
<td>6 79 11 (3.65) (82.29) (11.46)</td>
</tr>
<tr>
<td>Spain</td>
<td>11 1009 1091 (0.52) (47.80) (51.68)</td>
<td>13 595 1503 (0.62) (28.19) (71.19)</td>
<td>18 1291 802 (0.85) (61.16) (37.99)</td>
</tr>
<tr>
<td>Sweden</td>
<td>11 500 103 (1.79) (81.43) (16.78)</td>
<td>6 38 570 (0.98) (6.19) (92.83)</td>
<td>6 474 134 (0.98) (77.20) (21.82)</td>
</tr>
<tr>
<td>Switzerland</td>
<td>1 567 339 (0.11) (62.51) (37.38)</td>
<td>2 174 731 (0.22) (19.18) (80.60)</td>
<td>3 671 233 (0.33) (73.98) (25.69)</td>
</tr>
<tr>
<td>Turkey</td>
<td>7 1582 393 (0.35) (79.82) (19.83)</td>
<td>3 1298 681 (0.15) (65.49) (34.36)</td>
<td>3 1800 179 (0.15) (90.82) (9.03)</td>
</tr>
<tr>
<td>Ukraine</td>
<td>2 92 57 (1.32) (60.93) (37.75)</td>
<td>3 15 133 (1.99) (9.93) (88.08)</td>
<td>3 18 130 (1.99) (81.19) (18.80)</td>
</tr>
<tr>
<td>United Kingdom1</td>
<td>0 4488 5211 (0.00) (46.27) (53.73)</td>
<td>0 3931 5768 (0.00) (40.53) (59.47)</td>
<td>0 6474 3225 (0.00) (66.75) (33.25)</td>
</tr>
<tr>
<td>Total</td>
<td>258 26053 19622 (0.56) (56.72) (42.72)</td>
<td>699 15207 30027 (1.52) (33.11) (65.37)</td>
<td>263 32642 13028 (0.57) (71.06) (28.36)</td>
</tr>
</tbody>
</table>

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.

<table>
<thead>
<tr>
<th>Country</th>
<th>Oxygen therapy this year number (%)</th>
<th>NIPPV &gt; 3 months this year number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Missing/unknown</td>
<td>No</td>
</tr>
<tr>
<td>Albania</td>
<td>4 (3.77)</td>
<td>99 (93.40)</td>
</tr>
<tr>
<td>Armenia</td>
<td>0 (0.00)</td>
<td>23 (92.00)</td>
</tr>
<tr>
<td>Austria</td>
<td>1 (0.14)</td>
<td>686 (95.54)</td>
</tr>
<tr>
<td>Belarus</td>
<td>0 (0.00)</td>
<td>150 (96.77)</td>
</tr>
<tr>
<td>Belgium</td>
<td>184 (16.52)</td>
<td>904 (81.15)</td>
</tr>
<tr>
<td>Bulgaria</td>
<td>0 (0.00)</td>
<td>180 (96.77)</td>
</tr>
<tr>
<td>Croatia</td>
<td>4 (3.05)</td>
<td>117 (89.32)</td>
</tr>
<tr>
<td>Cyprus</td>
<td>1 (3.70)</td>
<td>26 (96.30)</td>
</tr>
<tr>
<td>Czech Republic</td>
<td>0 (0.00)</td>
<td>581 (97.98)</td>
</tr>
<tr>
<td>Denmark</td>
<td>279 (59.48)</td>
<td>188 (40.90)</td>
</tr>
<tr>
<td>France¹</td>
<td>0 (0.00)</td>
<td>5579 (95.91)</td>
</tr>
<tr>
<td>Georgia</td>
<td>0 (0.00)</td>
<td>71 (98.61)</td>
</tr>
<tr>
<td>Germany²</td>
<td>46 (0.78)</td>
<td>5443 (91.86)</td>
</tr>
<tr>
<td>Greece</td>
<td>3 (0.75)</td>
<td>386 (96.74)</td>
</tr>
<tr>
<td>Hungary</td>
<td>13 (2.92)</td>
<td>394 (88.54)</td>
</tr>
<tr>
<td>Ireland</td>
<td>2 (0.18)</td>
<td>1059 (93.05)</td>
</tr>
<tr>
<td>Israel</td>
<td>5 (0.97)</td>
<td>499 (97.28)</td>
</tr>
<tr>
<td>Italy</td>
<td>14 (0.27)</td>
<td>5011 (94.99)</td>
</tr>
<tr>
<td>Latvia</td>
<td>0 (0.00)</td>
<td>44 (97.78)</td>
</tr>
<tr>
<td>Lithuania</td>
<td>0 (0.00)</td>
<td>25 (92.59)</td>
</tr>
<tr>
<td>Luxembourg</td>
<td>0 (0.00)</td>
<td>35 (97.22)</td>
</tr>
</tbody>
</table>

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

<table>
<thead>
<tr>
<th>Country</th>
<th>Oxygen therapy this year number (%)</th>
<th>NIPPV &gt; 3 months this year number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Missing/unknown No Yes Missing/unknown No Yes, BIPAP (Bilevel Positive Airways Pressure) Yes, CPAP (Continuous Positive Airways Pressure)</td>
<td></td>
</tr>
<tr>
<td>Rep of Moldova</td>
<td>0 (0.00) 51 (96.23) 2 (3.77) 0 (0) 53 (100) 0 (0) 0 (0)</td>
<td></td>
</tr>
<tr>
<td>The Netherlands</td>
<td>5 (0.36) 1354 (97.13) 35 (2.51) 5 (0.36) 1355 (97.20) 0 (0.00) 34 (2.44)</td>
<td></td>
</tr>
<tr>
<td>North Macedonia</td>
<td>0 (0.00) 119 (92.25) 10 (7.75) 1 (0.78) 128 (99.22) 0 (0.00) 0 (0.00)</td>
<td></td>
</tr>
<tr>
<td>Norway³</td>
<td>0 (0.00) 274 (98.92) 3 (1.08) 1 (0.36) 275 (99.28) 1 (0.36) 0 (0.00)</td>
<td></td>
</tr>
<tr>
<td>Poland</td>
<td>9 (0.77) 1105 (94.12) 60 (5.11) 13 (1.11) 1152 (98.12) 8 (0.68) 1 (0.09)</td>
<td></td>
</tr>
<tr>
<td>Portugal</td>
<td>3 (0.92) 303 (92.95) 20 (6.13) 2 (0.61) 310 (95.09) 13 (3.99) 1 (0.31)</td>
<td></td>
</tr>
<tr>
<td>Romania</td>
<td>2 (0.88) 217 (96.02) 7 (3.10) 4 (1.77) 219 (96.91) 2 (0.88) 1 (0.44)</td>
<td></td>
</tr>
<tr>
<td>Russian Federation</td>
<td>9 (0.29) 2981 (95.67) 126 (4.04) 19 (0.61) 3066 (98.39) 13 (0.42) 18 (0.58)</td>
<td></td>
</tr>
<tr>
<td>Serbia</td>
<td>0 (0.00) 178 (97.27) 5 (2.73) 0 (0.00) 181 (98.91) 2 (1.09) 0 (0.00)</td>
<td></td>
</tr>
<tr>
<td>Slovak Republic</td>
<td>4 (1.43) 264 (94.63) 11 (3.94) 3 (1.08) 274 (98.20) 0 (0.00) 2 (0.72)</td>
<td></td>
</tr>
<tr>
<td>Slovenia</td>
<td>1 (1.04) 91 (94.79) 4 (4.17) 3 (3.13) 90 (93.75) 1 (1.04) 2 (0.08)</td>
<td></td>
</tr>
<tr>
<td>Spain</td>
<td>12 (0.57) 2036 (96.45) 63 (2.98) 14 (0.66) 2082 (98.63) 11 (0.52) 4 (0.19)</td>
<td></td>
</tr>
<tr>
<td>Sweden</td>
<td>6 (0.98) 593 (96.58) 15 (3.42) 4 (0.65) 602 (98.05) 8 (1.30) 0 (0.00)</td>
<td></td>
</tr>
<tr>
<td>Switzerland</td>
<td>5 (0.55) 864 (95.26) 38 (4.19) 3 (0.33) 898 (99.01) 3 (0.33) 3 (0.33)</td>
<td></td>
</tr>
<tr>
<td>Turkey</td>
<td>3 (0.15) 1916 (96.67) 63 (3.18) 4 (0.20) 1908 (96.27) 66 (3.33) 4 (0.20)</td>
<td></td>
</tr>
<tr>
<td>Ukraine</td>
<td>3 (1.99) 120 (79.47) 28 (18.54) 2 (1.32) 149 (98.68) 0 (0.00) 0 (0.00)</td>
<td></td>
</tr>
<tr>
<td>United Kingdom</td>
<td>0 (0.00) 9142 (94.26) 557 (5.74) 0 (0.00) 9541 (98.37) 0 (0.00) 158 (1.63)</td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>618 (1.35) 43108 (93.85) 2207 (4.80)</td>
<td></td>
</tr>
</tbody>
</table>

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.
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.

<table>
<thead>
<tr>
<th>Country</th>
<th>Inhaled steroids &gt; 3 months this year number (%)</th>
<th>Oral steroids &gt; 3 months this year number (%)</th>
<th>PPI &gt; 3 months this year number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Missing/unknown</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Albania</td>
<td>2 (1.89)</td>
<td>85</td>
<td>19</td>
</tr>
<tr>
<td>Armenia</td>
<td>0 (0.00)</td>
<td>20</td>
<td>5</td>
</tr>
<tr>
<td>Austria</td>
<td>2 (0.28)</td>
<td>623</td>
<td>93</td>
</tr>
<tr>
<td>Belarus</td>
<td>0 (0.00)</td>
<td>108</td>
<td>47</td>
</tr>
<tr>
<td>Belgium</td>
<td>1 (0.09)</td>
<td>528</td>
<td>585</td>
</tr>
<tr>
<td>Bulgaria</td>
<td>0 (0.00)</td>
<td>169</td>
<td>17</td>
</tr>
<tr>
<td>Croatia</td>
<td>4 (3.05)</td>
<td>103</td>
<td>24</td>
</tr>
<tr>
<td>Cyprus</td>
<td>1 (3.70)</td>
<td>16</td>
<td>10</td>
</tr>
<tr>
<td>Czech Republic</td>
<td>0 (0.00)</td>
<td>389</td>
<td>204</td>
</tr>
<tr>
<td>Denmark</td>
<td>469 (0.10)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>France</td>
<td>0 (0.00)</td>
<td>2475</td>
<td>3342</td>
</tr>
<tr>
<td>Georgia</td>
<td>1 (1.39)</td>
<td>71</td>
<td>0</td>
</tr>
<tr>
<td>Germany</td>
<td>57 (0.96)</td>
<td>3721</td>
<td>2147</td>
</tr>
<tr>
<td>Greece</td>
<td>2 (0.50)</td>
<td>263</td>
<td>134</td>
</tr>
<tr>
<td>Hungary</td>
<td>445 (0.10)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Ireland</td>
<td>2 (0.18)</td>
<td>759</td>
<td>377</td>
</tr>
<tr>
<td>Israel</td>
<td>3 (0.58)</td>
<td>271</td>
<td>239</td>
</tr>
<tr>
<td>Italy</td>
<td>1461 (27.70)</td>
<td>2762</td>
<td>1052</td>
</tr>
<tr>
<td>Latvia</td>
<td>0 (0.00)</td>
<td>38</td>
<td>7</td>
</tr>
<tr>
<td>Lithuania</td>
<td>0 (0.00)</td>
<td>26</td>
<td>1</td>
</tr>
<tr>
<td>Luxembourg</td>
<td>0 (0.00)</td>
<td>18</td>
<td>18</td>
</tr>
<tr>
<td>Rep of Moldova</td>
<td>0 (0.00)</td>
<td>47</td>
<td>6</td>
</tr>
</tbody>
</table>
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.

<table>
<thead>
<tr>
<th>Country</th>
<th>Age at follow-up (years)</th>
<th>Use of Ivacaftor this year, number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Missing/unknown</td>
</tr>
<tr>
<td>Albania</td>
<td>1-17</td>
<td>0 (0)</td>
</tr>
<tr>
<td></td>
<td>≥18</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Austria</td>
<td>1-17</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td></td>
<td>≥18</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Belgium</td>
<td>1-17</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td></td>
<td>≥18</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Bulgaria</td>
<td>1-17</td>
<td>0 (0)</td>
</tr>
<tr>
<td></td>
<td>≥18</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Croatia</td>
<td>1-17</td>
<td>0 (0)</td>
</tr>
<tr>
<td></td>
<td>≥18</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Czech Republic</td>
<td>1-17</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td></td>
<td>≥18</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Denmark</td>
<td>≥18</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>France</td>
<td>1-17</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td></td>
<td>≥18</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Germany</td>
<td>1-17</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td></td>
<td>≥18</td>
<td>3 (1.69)</td>
</tr>
<tr>
<td>Greece</td>
<td>≥18</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Hungary</td>
<td>1-17</td>
<td>0 (0)</td>
</tr>
<tr>
<td></td>
<td>≥18</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Ireland</td>
<td>1-17</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td></td>
<td>≥18</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Israel</td>
<td>1-17</td>
<td>0 (0)</td>
</tr>
<tr>
<td></td>
<td>≥18</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Italy</td>
<td>1-17</td>
<td>28 (34.15)</td>
</tr>
<tr>
<td></td>
<td>≥18</td>
<td>32 (28.57)</td>
</tr>
<tr>
<td>Rep of Moldova</td>
<td>1-17</td>
<td>0 (0)</td>
</tr>
<tr>
<td>The Netherlands</td>
<td>1-17</td>
<td>0 (0)</td>
</tr>
<tr>
<td></td>
<td>≥18</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>North Macedonia</td>
<td>1-17</td>
<td>0 (0)</td>
</tr>
<tr>
<td></td>
<td>≥18</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Norway</td>
<td>1-17</td>
<td>0 (0)</td>
</tr>
<tr>
<td></td>
<td>≥18</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Poland</td>
<td>1-17</td>
<td>0 (0)</td>
</tr>
<tr>
<td></td>
<td>≥18</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Portugal</td>
<td>≥18</td>
<td>0 (0.00)</td>
</tr>
<tr>
<td>Romania</td>
<td>1-17</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Russian Federation</td>
<td>1-17</td>
<td>0 (0)</td>
</tr>
<tr>
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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.

<table>
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<tr>
<th>Country</th>
<th>Age at follow-up (years)</th>
<th>Use of Lumacaftor/Ivacaftor this year number (%)</th>
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### Table 7.11 continued

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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.
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.

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<td>22 (1.29)</td>
</tr>
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</tr>
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</tr>
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</tr>
<tr>
<td></td>
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</tr>
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<td>Ireland</td>
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</tr>
<tr>
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</tr>
<tr>
<td>Israel</td>
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</tr>
<tr>
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<td>≥18</td>
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</tr>
<tr>
<td>Italy</td>
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<td>≥18</td>
<td>641 (73.76)</td>
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<tr>
<td>Lithuania</td>
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</tr>
<tr>
<td></td>
<td>≥18</td>
<td>0 (0)</td>
</tr>
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</tr>
<tr>
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</tr>
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</tr>
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<td></td>
<td>≥18</td>
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</tr>
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<td>≥18</td>
<td>2 (0.35)</td>
</tr>
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</tr>
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<td>0 (0)</td>
</tr>
<tr>
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<td>≥18</td>
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[table 7.12 continued]

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

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.**

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

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.**

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

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.

<table>
<thead>
<tr>
<th>Age</th>
<th>Males</th>
<th>Females</th>
<th>Total</th>
<th>Transplants carried out in 2019</th>
</tr>
</thead>
<tbody>
<tr>
<td>5-9</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>15-19</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>20-24</td>
<td>3</td>
<td>1</td>
<td>4</td>
<td>1</td>
</tr>
<tr>
<td>25-29</td>
<td>2</td>
<td>8</td>
<td>10</td>
<td>3</td>
</tr>
<tr>
<td>30-34</td>
<td>6</td>
<td>9</td>
<td>15</td>
<td>4</td>
</tr>
<tr>
<td>35-39</td>
<td>15</td>
<td>17</td>
<td>32</td>
<td>8</td>
</tr>
<tr>
<td>40-44</td>
<td>20</td>
<td>14</td>
<td>34</td>
<td>0</td>
</tr>
<tr>
<td>45+</td>
<td>22</td>
<td>21</td>
<td>43</td>
<td>2</td>
</tr>
<tr>
<td>Total</td>
<td>70</td>
<td>71</td>
<td>141</td>
<td>20</td>
</tr>
</tbody>
</table>

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.

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

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.

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

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.

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

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:


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:

The ECFSPR is supported by:

National Patient Organisations

Industry with a Donation or Sponsorship
## 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.

<table>
<thead>
<tr>
<th>Country</th>
<th>Centre/National Registry name</th>
<th>Contact</th>
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<tbody>
<tr>
<td>Albania</td>
<td>1 individual centre: “Mother Thereza” Hospital Center, Department of Paediatrics, Tirana</td>
<td>Irena Kasmi</td>
</tr>
<tr>
<td>Armenia</td>
<td>1 individual centre: Yerevan State Medical University, Muratsan University Hospital, Cystic Fibrosis Center, Yerevan</td>
<td>Satenik Harutyunyan</td>
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</tbody>
</table>
| Austria | 14 individual centres:  
Medizinische Universität Graz, Universitätsklinik für Kinder- und Jugendheilkunde, Klinische Abteilung für Pädiatrische Pulmonologie und Allergologie und CF Zentrum für Kinder, Jugendliche und Erwachsene, Graz  
Medizinische Universität Innsbruck, Zertifiziertes CF Zentrum für Kinder, Jugendliche und Erwachsene, Innsbruck  
Klinikum Klagenfurt am Wörthersee, Abteilung für Kinder- und Jugendheilkunde, Pädiatrische Pulmologie/Allergologie, Klagenfurt  
Kepler Universitätsklinikum, Universitätsklinik für Kinder- und Jugendheilkunde, Linz  
Kepler Universitätsklinikum, Klinik für Lungenheilkunde/Pneumologie, Linz  
Kardinal Schwarzenberg Klinikum, Abteilung für Kinder- und Jugendmedizin, Schwarzach im Pongau  
Salzburger Landeskliniken, Universitätsklinik für Pneumologie, Salzburg  
Landeskrankenhaus Steyr, Abteilung für Kinder- und Jugendheilkunde und Abteilung für Lungenheilkunde, Steyr  
Medizinische Universität Wien, Allgemeines Krankenhaus Wien für Thoraxchirurgie, Vienna  
Medizinische Universität, Allgemeines Krankenhaus Wien, Universitätsklinik für Kinder-und Jugendheilkunde, Klinische Abteilung für Pädiatrische Pneumologie, Allergologie und Endokrinologie, Zentrum für Cystische Fibrose, Vienna  
Klinik Ottakring, Abteilung für Kinder- und Jugendheilkunde mit Ambulanz, Vienna  
Krankenhaus Hietzing, Abteilung für Atemungs- und Lungenerkrankungen, Vienna | Andreas Pfleger  
Ernst Eber  
Maria Gaber  
Helmut Ellemunter  
Johannes Eder  
Franz Hubert Wadlegger  
Adrienne Molnar  
Julia Pichler  
Christina Thir  
Martin Stadlinger  
Viktoria Reinelt  
Katrin Scheich  
Josef Riedler  
Christoph Seelbach  
Michael Studnicka  
Natalie Firlei-Fleischmann  
Alexander Ebner  
Margit Kalinger  
Monika Pell  
Peter Jaksch  
Dagmar Liebhart  
Sabine Renner  
Saskia Gruber  
Brigitte Mersi  
Thomas Frischer  
Kerstin Tiringer  
Katharina Kainz  
Andrea Lakatos–Krepcik  
Dr. Sabine Burghart |
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<tr>
<th>Country</th>
<th>Centre/National Registry name</th>
<th>Contact</th>
</tr>
</thead>
</table>
| Klinikum Wels-Grieskirchen, Abteilung für Kinder- und Jugendheilkunde, Wels | Franz Eitelberger  
Beatrix Wintersteiger  
Vera Karin Bauer  
Alexander Leitner  
Matthäus Ploder  
Thomas Tempelmayer  
Andrea Ringl |
| Klinikum Wels-Grieskirchen, Abteilung für Lungenkrankheiten, Wels          |                                                              |
| Belarus      | Belarusian Republic Children's Center of Pulmonology and Cystic Fibrosis, Pulmonary Department, 3rd City Children's Clinical Hospital, Minsk | Vladimir Bobrovniichiy  
Svetlana Keegan |
| Belgium      | Belgian Cystic Fibrosis Registry                                                               | Géraldine Daneau  
Simeon Wanyama |
| Bulgaria     | 2 individual centres:  
Alexandrovsk University Hospital, Pediatric Clinic, Sofia                               | Guergana Petrova  
Guergana Petrova  
Miglena Georgieva  
Nataliya Dobrudzhanska  
Margarita Nikolova  
Ruzha Pancheva |
|              | University Hospital St. Marina, 2nd Paediatric Clinic, Varna                               |                                                              |
| Croatia      | 1 individual centre:  
University Hospital Centre Zagreb, Cystic Fibrosis Centre – Paediatrics and Adults, Zagreb | Duska Tjesic-Drinkovic  
Andrea Vukic Dugac  
Duska Tjesić-Drinković  
Dorian Tjesić-Drinković  
Andrea Vukic Dugac  
Ivan Bambir  
Ivona Markelkic |
|              | On behalf of the Croatian CF Patient Database                                                  |                                                              |
| 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 Naehrlrich  
Julia Wosniok |
| Greece       | 2 individual centres:  
Sismanoglio General Hospital of Attica, Adult Cystic Fibrosis Unit, Athens                | Elpis Hatzigaporou  
Filla Diamantea  
Margarita Gkotsina  
John Tsanakas  
Elpis Hatzigaporou  
Aikaterini Manika  
Maria Sionidou  
Maria Fotoulaki |
|              | Aristotle University of Thessaloniki, Cystic Fibrosis Centre, Thessaloniki                   | Ia Khurtsilava  
Tsisino Parulava |
| Georgia      | I. Tsitsishvili Children’s Clinic, CF Centre, Tblisi                                           | Ia Khurtsilava  
Tsisino Parulava |
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<td>Meir Mei-Zahav</td>
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<td>Carmel Medical Centre, Haifa</td>
<td>Micha Aviram</td>
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<td>Meyer Children’s Hospital of Haifa, Rambam Medical Center, Haifa</td>
<td>Galit Livnat</td>
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<td></td>
<td>Hadassah Medical Centre, Mount Scopus, Jerusalem</td>
<td>Michal Gur</td>
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<tr>
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<td>Schneider Children’s Medical Centre of Israel, Petach Tikvah</td>
<td>Meir Mei-Zahav</td>
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<td>Safra Children’s Hospital, Sheba Medical Center, Ramat Gan</td>
<td>Ori Efrati</td>
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<td>Italian Cystic Fibrosis Registry</td>
<td>Rita Padoan Marco Salvatore Annalisa Amato Gianluca Ferrari</td>
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<td>Elina Aleksejeva Elina Aleksejeva Dita Gaïdule-Logina</td>
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<td>Institute for respiratory diseases in children Kozle, Center for cystic fibrosis, Children and adults, Kozle</td>
<td>Tatjana Jakovska-Maretti Ivana Arnaudova Danevska</td>
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<td>Rep. of Moldova</td>
<td>Outpatient Center for Cystic Fibrosis and Other Rare Diseases</td>
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<td>Egil Bakkeheim Anita Senstad Wathne</td>
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<td>Radoslawa Staszak – Kowalska</td>
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<td></td>
<td>Cystic Fibrosis Centre, Polanki Paediatric Hospital, Gdansk</td>
<td>Mikolaj Kowalski</td>
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<td></td>
<td>Centrum Medyczne Karpacz, Children/Adults’ Hospital, Karpacz</td>
<td>Maria Trawinska-Bartnicka Ewa Sapiejk</td>
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<td>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</td>
<td>Grzegorz Gaszczyn Monika Rams</td>
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<td>Urszula Grzybowska-Chlebowczyk Bozena Koryds-Dormolinska</td>
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<td>Wojewódzkie Wielospecjalistyczne centrum onkologii i traumatologii im. m. Kopernika w Łodzi. Ośrodek Pediatriczny im. Dr J. Korczak</td>
<td>Iwona Stelmach Agnieszka Koniarek-Maniecka</td>
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<td>University Hospital of Lords Transfiguration, Dept. of Pulmonology, Allergology and Pulmonary Oncology, Poznan</td>
<td>Szczepan Cofta Agata Nowicka</td>
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<td>Karol Jonscher University Hospital of Poznan University of Medical Sciences, Poznan</td>
<td>Irena Wojsyk-Banaszak</td>
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<td>Institute of Tuberculosis and Lung Diseases, Rabka-Zdró Branch, Dept. of Pneumology and Cystic Fibrosis, Rabka Zdroj</td>
<td>Henryk Mazurek Lidia Pawlik</td>
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<td>Provincial Clinical Hospital no. 2, Dept of Allergology and Cystic Fibrosis, St Jadwigi Krolowej in Rzeszow</td>
<td>Marta Rachel</td>
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<td>Dorota Sands Lukasz Wozniacki</td>
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<td>Liviu Pop</td>
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<td>Laura Larisa Dracea</td>
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<td>Children Emergency Hospital &quot;Maria S. Curie&quot;, Bucharest</td>
<td>Maria Brustan</td>
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<td>Clinical Children’s Hospital Grigore Alexandrescu, Bucharest</td>
<td>Simona Mosescu</td>
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<td>Mother &amp; Child Health Institute, Bucharest</td>
<td>Iustina Stan</td>
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<tr>
<td></td>
<td>Regional Cystic Fibrosis Centre Cluj, Cluj-Napoca</td>
<td>Radu Sorin Şerban</td>
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<td>“S.F. Maria” Children’s Emergency Hospital, Iasi</td>
<td>Szabo Csilla-Enikő</td>
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<td>National Cystic Fibrosis Centre, Timișoara</td>
<td>Dana Anton-Paduraru</td>
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<td>Serbia</td>
<td>1 individual centre: National Centre for Cystic Fibrosis, Mother and Child Health Institute of Serbia &quot;Dr Vukan Cupic&quot;, Belgrade</td>
<td>Milan Rodic, Predrag Minić, Milan Rodić, Aleksandar Sovtić</td>
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<td>Slovakia</td>
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<td>Hana Kayserova</td>
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<td>Centrum cystickej fibrozy pre dospelých FNSP FDR, Banská Bystrica</td>
<td>Branko Takáč</td>
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<td>Centrum cystickej fibrozy pre dospelých, Klinika pneumologie I.SZU a Univerzitna nemocnica, Bratislava</td>
<td>Eva Bérešova</td>
</tr>
<tr>
<td></td>
<td>Klinika detskej pneumologie SZU UN Bratislava, pracovisko Podunajské Biskupice, Bratislava</td>
<td>Marta Hajkova</td>
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<tr>
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<td>CF Adult centre, University Hospital L Pasteura, Košice</td>
<td>Lenka Kopčová</td>
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<td></td>
<td>Centrum cystickej fibrozy detí, Detská fakultná nemocnica Košice, Košice</td>
<td>Anna Fetekeoeva, Zuzana Hribiková</td>
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<tr>
<td>Slovenia</td>
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<td></td>
<td>University Clinic of Pulmonary and Allergic Diseases, Golnik</td>
<td>Uroš Krivec</td>
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<td></td>
<td>University Medical Centre Ljubljana, University Children`s Hospital, Unit for pulmonary diseases</td>
<td>Matjaž Fležar, Tjaša Brus Pičman, Julij Šelb</td>
</tr>
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<td></td>
<td>University Medical Centre Ljubljana, Department of Pulmonology and Allergy</td>
<td>Uroš Krivec</td>
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<td>Jasna Rodman Berlot, Majda Oštir, Izidor Kos, Barbara Salobir</td>
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<td>Spain</td>
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<td>Parc Taulí Hospital Universitario, Hospital de Sabadell, Unitat de Pneumologia Pediàtrica i Unitat de Fibrosi Quística, Sabadell, Barcelona</td>
<td>Mª Dolores Pastor Vivero, Oscar Asensio de la Cruz, Miguel García González, Xavier Pomares Amigó, Concepción Montón Soler, MariaColsRoig, Jordi Costa i Colomer</td>
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<tr>
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<td>Hospital Sant Joan de Déu, Unitat de Pneumologia Pediàtrica i Fibrosi Quística, Barcelona</td>
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<td>Hospital Universitari Vall d'Hebron, Unidad de Fibrosis Quística del Adulto, Barcelona</td>
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<td>Hospital Vall d’Hebron, Unidad Fibrosis Quística y Neumología Pediàtrica, Barcelona</td>
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<td>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</td>
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<td>Complejo Hospitalario Universitario Insular Materno Infantil, Las Palmas de Gran Canaria</td>
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<td>Hospital Universitario La Paz, Unidad de Fibrosis Quística Adultos, Servicio de Neumología, Madrid</td>
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<td>María Isabel Barrio Gomez de Agüero Marta Ruiz de Valbuena Maiz Cristina de Manuel Gómez Rosa María Girón José R. Villa Asensi Patricia Fernandez García Alejandro López Neyra Verónica Sanz Santiago Rosa Ana Muñoz Codoceo Luis Maiz Carro Rosa María Nieto Royo Adelaida Lamas Ferreiro Saioa Vicente Santamaría</td>
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<td>Layla Diab Cáceres Carmen Luna Paredes Enrique Salcedo Lobato Casilda Oliveira Fuster Gabriel María Oliveira Fuster Nuria Porras Pérez Francisco Javier Pérez Frias Estela Pérez Ruiz Pilar Caro-Aguilera</td>
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<td>Alicia Callejon Orlando Mesa Amparo Escribano Silvia Castillo Corullón Amparo Solé Jover Carmen Inés Perez Munoz Cristina Ramos Hernández Maria Jesús Rodríguez</td>
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<td>Constance Barazzone, Anne Mornand, Nadège Gabent</td>
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<td>Consultation de Mucoviscidose Adulte et de CFTR-related Disorders, Service de Pneumologie, Département de Médecine, Centre Hospitalier Universitaire Vaudois (CHUV), Lausanne</td>
<td>Angela Koutsokera, Zisis Balmouzis, Isabelle Huart Bellavere, Caroline Dutoit, Nicolas Regamey, Michael Hitzler, Marco Lurà, Lucia Eichhorn, Sonja Ettlin</td>
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<td>Children’s Hospital of Eastern Switzerland, Division of Paediatric Pulmonology &amp; CF Centre, St Gallen</td>
<td>Jürg Barben, Christine Baumgartner</td>
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<td>Turkey</td>
<td>Cystic Fibrosis Registry of Turkey</td>
<td>Deniz Dogru, Bülent Karadağ, Yasemin Gökdemir, Ela Eralp</td>
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<td>Marmara University Faculty of Medicine, Division of Pediatric Pulmonology, Istanbul</td>
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<td>Saniye Girit, Yetkin Ayhan</td>
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<td>1 individual centre: Cystic Fibrosis Centre of Western Ukrainian Specialized Children's Medical Centre, Lviv</td>
<td>Halyna Makukh, Lyudmyla Bober, Halyna Makukh</td>
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<tr>
<td>United Kingdom</td>
<td>UK Cystic Fibrosis Registry</td>
<td>Rebecca Cosgriff, Susan Charman, Elaine Gunn, Siobhán Carr, Sarah Clarke</td>
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</tbody>
</table>
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 USA1. 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 FEV1 predicted values
We computed the percent of predicted values for FEV1 and FVC using:

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

<table>
<thead>
<tr>
<th>Demographics</th>
<th>Diagnosis</th>
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<tr>
<td>CF centre code</td>
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<td>Patient code</td>
<td>Age at diagnosis</td>
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<tr>
<td>Year of follow-up</td>
<td>Sweat test type and value</td>
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<td>Date of birth (year and month)</td>
<td>Electrolytes</td>
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<td>Gender</td>
<td>Chloride value</td>
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<td>Status of patient</td>
<td>Meconium Ileus</td>
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<td>Cause of death</td>
<td>Nasal Potential Difference (NPD)</td>
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<td>Date of death</td>
<td>CF-typical NPD</td>
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<td>Date of NPD</td>
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<td>Intestinal current measurement (ICM)</td>
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<td>CF-typical ICM</td>
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<th>Genotype</th>
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<td>First mutation</td>
<td>Inhaled continuous hypertonic NaCl this year</td>
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<tr>
<td>Second mutation</td>
<td>Inhaled continuous Mannitol</td>
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<tr>
<td></td>
<td>Inhaled continuous antibiotic this year</td>
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<tr>
<td></td>
<td>Inhaled continuous bronchodilators this year</td>
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<td></td>
<td>In Oxygen therapy this year</td>
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<tr>
<td></td>
<td>Use of Non-invasive positive pressure ventilation (NIPPV)</td>
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<td></td>
<td>Use of rhDNase this year</td>
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<td></td>
<td>Use of continuous Inhaled steroids</td>
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<td></td>
<td>Use of continuous Oral steroids</td>
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<td></td>
<td>Use of continuous azithromycin (or other macrolide) this year</td>
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<td></td>
<td>Use of ursodeoxycholic acid this year</td>
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<td>Use of pancreatic enzymes this year</td>
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<td>Use of proton pump inhibitors (PPI)</td>
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<td></td>
<td>Use of CFTR Modifier Therapy</td>
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</tbody>
</table>
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

Follow-up

- Date of best FEV1 recorded this year
- Value of best FEV1 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 FEV1 (or in case of no FEV1 last height of the year)
- Weight measured at date of best FEV1 (or in case of no FEV1 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 ECFSPR

**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:

**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:

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:

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:

**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:

**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) > 7 g/d and/or faecal pancreatic elastase-1 < 200 μg/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:

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

References:

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.