2017

ECFS Patient Registry
Annual Data Report

European Cystic Fibrosis Society
Kastanieparken 7
7470 Karup
Denmark

ECFS Patient Registry

Annual Data Report

2017 data

ecfs-pr@uzleuven.be
www.ecfs.eu/ecfspr
@ECFSRegistry
www.facebook.com/EuropeanCysticFibrosisPatientRegistry/

Table of contents

Published: July 2019

Preface

We are pleased to share with you the 2017 Annual Report from the European Cystic Fibrosis Society Patient Registry (ECFSPR). This 13th report contains demographic and clinical data of 48,204 consenting CF patients from 35 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 clear and comprehensive picture of CF clinical outcomes across Europe. The analyses presented in this report have been carried out by the ECFSPR statisticians using the anonymised, raw data submitted by the participating countries. The results of analyses for some countries as presented in the ECSFPR report may differ from the data published in their national annual registry report; differences can originate from variation in patient inclusion criteria, the definitions used for disease complications and the employment of different reference values. Further details on how this occurs and is dealt with can be found in the report and in the List of ECFSPR Variables and Definitions in Appendix 2 (page 144).

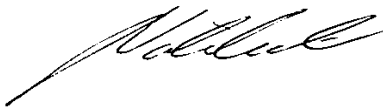
During the past years the Registry has grown considerably and has become the largest CF database in the world, thanks to the essential support from the contributing centres and national registries in Europe and neighbouring countries. In the following years our focus will remain on improving and monitoring data quality, and on increasing the use of the data for scientific publications and long-term safety and efficacy studies of new therapies. Essential elements to achieve those objectives are complete longitudinal data-sets of high quality data and a coverage in each participating country of 80% or more.

We will also continue the invaluable collaboration with CF Europe and the ECFS Pharmacovigilance Group, which has led to the implementation of several important projects.

The management of the ECFSPR and the development of this report take a considerable amount of work. I would like to thank the national registries and individual centres, as well as the country representatives, for submitting data to the ECFSPR, and the ECFSPR staff for their hard work in making this report possible. Managing the Registry comes with a cost and we are indebted to our sponsors whose unrestricted grants help to support the running and expansion of the Registry.

Finally, I would like to thank all the people with CF throughout Europe for their willingness to participate in the Registry, because without them it would not exist. We hope that the information in this report is useful for people with CF, their families and caregivers and that it will lead to improved CF care throughout Europe.

Sincerely,



Lutz Naehrlich, MD
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.

We will continue to publish a separate At-a-Glance report containing key information, from the ECFSPR Report, relevant for people with CF and their families: www.ecfs.eu/projects/ecfs-patient-registry/annual-reports. Interactive maps with country-relevant information are available on our website: www.ecfs.eu/ecfspr.

Recently we developed country posters with information and basic statistics from the Registry for display in CF-clinics. Every 3 to 4 years the data in the posters will be updated. The posters are also published online: www.ecfs.eu/ecfspr/posters.

We also increased the Registry's presence on social media: in June 2019 we launched a Facebook account www.facebook.com/EuropeanCysticFibrosisPatientRegistry/ where we will post news, updates and other interesting information. You can also follow us on Twitter @ECFSRegistry.

In the next few years we will carry on, together with the patient organisations, with our projects aimed at increasing awareness of the Registry among people with CF and their families.

If you have any suggestions on how we can improve the information, or if something is unclear, you are welcome to contact us by sending an email to: ecfs-pr@uzleuven.be.

For discussions about the results in your country we encourage you to contact your CF centre.

For more detailed information about the ECFSPR visit the patient-dedicated page on our website: www.ecfs.eu/projects/ecfs-patient-registry/information-about-ecfspr-cf-patients.

List of centres and national registries that provided data

List of individual centres and national registries that contributed to the ECFSPR.

In large 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 since the report with 2017 data.

Country	Centre/National Registry name	Contact
Albania	1 individual centre:	Irena Kasmi
	"Mother Thereza" Hospital Center, Department of Paediatrics, Tirana	*Irena Kasmi* *Evda Vevecka*
Armenia	1 individual centre:	Satenik Harutyunyan
	Yerevan University CF Centre, Muratsan Hospital, Yerevan	*Satenik Harutyunyan*
Austria	13 individual centres:	Andreas Pfleger
	Medizinische Universität Graz, Universitätsklinik für Kinder- und Jugendheilkunde, Klinische Abteilung für Pädiatrische Pulmonologie und Allergologie und CF Zentrum für Kinder, Jugendliche und Erwachsene, Graz	Ernst Eber Andreas Pfleger Maria Wagenhofer
	Medizinische Universität Innsbruck, Zertifiziertes CF Zentrum für Kinder, Jugendliche und Erwachsene, Innsbruck	Helmut Ellemunter Johannes Eder
	Klinikum Klagenfurt am Wörthersee, Abteilung für Kinder- und Jugendheilkunde, Pädiatrische Pulmologie/ Allergologie, Klagenfurt	Franz Hubert Wadlegger
	Univ. Klinik für Kinder- und Jugendheilkunde, Kepler Universitätsklinikum, Linz	Wolfgang Högler Julia Pichler
	Kardinal Schwarzenberg Klinikum, Abteilung für Kinder- und Jugendmedizin, Schwarzach im Pongau	Josef Riedler Christoph Seelbach
	Landeskrankenhaus Steyr, Abteilung für Kinder- und Jugendheilkunde und Abteilung für Lungenheilkunde, Steyr	Josef Emhofer Alexander Ebner
	Medizinische Universität, Allgemeines Krankenhaus Wien, Universitätsklinik für Chirurgie, Klinische Abteilung für Thoraxchirurgie, Vienna	Peter Jaksch Dagmar Liebhart
	Medizinische Universität, Klinik für Kinder- und Jugendheilkunde, Cystische Fibrose Ambulanz, Vienna	Sabine Renner Brigitte Mersi
	Wilhelminenspital, Abteilung für Kinder- und Jugendheilkunde mit Ambulanz, Vienna	Thomas Frischer Kerstin Tiringer Katharina Kainz
	Krankenhaus Hietzing, Abteilung für Atmungs- und Lungenerkrankungen, Vienna	Andrea Lakatos–Krepcik Ingrid Kaluza
	Klinikum Wels-Grieskirchen, Abteilung für Kinder- und Jugendheilkunde, Wels	Franz Eitelberger Beatrix Wintersteiger Vera Karin Bauer
	Klinikum Wels-Grieskirchen, Abteilung für Lungenkrankheiten, Wels	Carolin Großruck Helmut Feizelmeier
	Salzburger Landeskliniken, Universitätsklinik für Pneumologie, Salzburg	*Michael Studnicka* *Natalie Firlei-Fleischmann*

Country	Centre/National Registry name	Contact
Belgium	Belgian Cystic Fibrosis Registry	Géraldine Daneau Simeon Wanyama Muriel Thomas
Bulgaria	2 individual centres: Alexandrovska University Hospital, Pediatric Clinic, Sofia *University Hospital St. Marina, 2nd Paediatric Clinic, Varna*	Guergana Petrova Guergana Petrova *Miglena Georgieva* *Margarita Nikolova* *Ruzha Pancheva*
Croatia	1 individual centre: *University Hospital Centre Zagreb, Cystic Fibrosis Centre – Paediatrics and Adults, Zagreb*	Duska Tjesic-Drinkovic Andrea Vukic Dugac *Andrea Vukic Dugac* *Ivona Markelic* *Duska Tjesić-Drinković* *Dorian Tjesić-Drinković* *Ivan Bambir*
Czech Republic	Cystic Fibrosis Registry of the Czech Republic	Pavel Drevinek Alena Bilkova Milan Macek Marek Turnovec
Denmark	Cystic Fibrosis Registry of Denmark	Hanne Vebert Olesen Tania Pressler
France	Registre Français de la Mucoviscidose	Anne Farge Lydie Lemonnier Clémence Dehillotte
Germany	Qualitätssicherung Mukoviszidose	Lutz Naehrlich Julia Wosniok
Greece	3 individual centres: Aghia Sophia Children's Hospital, CF Centre, Athens Sismanoglio General Hospital of Attica, Adult Cystic Fibrosis Unit, Athens Aristotle University of Thessaloniki, Hippokration General Hospital, Cystic Fibrosis Centre, Thessaloniki	Elpis Hatziagorou Athanasios Kaditis Ioanna Loukou Argyri Petrocheilou Filia Diamantea Dimitrios Papadopoulos John Tsanakas Elpis Hatziagorou Maria Fotoulaki John Kioumis
Hungary	Cystic Fibrosis Registry of Hungary	Rita Ujhelyi Géza Marsal Attila Hornyák
Ireland	Cystic Fibrosis Registry of Ireland	Godfrey Fletcher Laura Kirwan Abaigeal Jackson Shijun Zhou
Israel	Cystic Fibrosis Registry of Israel	Meir Mei-Zahav
Italy	Italian Cystic Fibrosis Registry	Rita Padoan Marco Salvatore Annalisa Amato Gianluca Ferrari

Country	Centre/National Registry name	Contact
Latvia	1 individual centre: Rīga Stradiņš University, Children's Clinical University Hospital, Department of Pneumology, Riga	Zane Timpare Vija Švabe Zane Timpare Liga Berke
Lithuania	1 individual centre: Hospital of Lithuanian University of Health Sciences, Kaunas Clinics, Adult Cystic Fibrosis Centre, Kaunas	Kęstutis Malakauskas Kęstutis Malakauskas Virginija Kalinauskaitė-Žukauskė
Luxembourg	1 individual centre: Centre Hospitalier de Luxembourg	Marc Schlesser Marc Schlesser Elisabeth Da Silva Inesse Denine
Rep. of North Macedonia	1 individual centre: University Children's Hospital, Centre for Cystic Fibrosis, Skopje	Stojka Fustik Stojka Fustik
Rep. of Moldova	Cystic Fibrosis Registry of Moldova	Oxana Turcu
Netherlands	Dutch Cystic Fibrosis Registry	Vincent Gulmans Domenique Zomer
Norway	Norwegian Cystic Fibrosis Patient Registry	Egil Bakkeheim Anita Senstad Wathne
Poland	9 individual centres: *Voivodeship Children's Hospital, Dept. of Paediatric Pneumology and Allergology, Bydgoszcz* *Cystic Fibrosis Centre, Polanki Paediatric Hospital, Gdansk* *Centrum Medyczne Karpacz, Children/Adults' Hospital, Karpacz* *St. Louis Regional Specialised Children's Hospital, Krakow* *University Hospital of Lords Transfiguration, Dept. of Pulmonology, Allergology and Pulmonary Oncology, Poznan* *Karol Jonscher University Hospital of Poznan University of Medical Sciences, Poznan* *Institue of Tuberculosis and Lung Disorders, Dept. of Pneumology and Cystic Fibrosis (Children & Adults), Rabka Zdroj* *Provincial Clinical Hospital no. 2, Dept of Allergology and Cystic Fibrosis, St Jadwigi Krolowej in Rzeszow* *Dziekanow Paediatric Hospital, Cystic Fibrosis Centre, Institute of Mother and Child, Warsaw*	Lukasz Wozniacki *Radoslawa Staszak – Kowalska* *Mikolaj Kowalski* *Maria Trawinska-Bartnicka* *Grzegorz Gaszczyk* *Monika Rams* *Stanislaw Stepniewski* *Daria Dziecichowicz-Latala* *Szczepan Cofta* *Agata Nowicka* *Irena Wojsyk-Banaszak* *Henryk Mazurek* *Patrycja Gburzynska-Czudzinowicz* *Krzysztof Balata* *Marta Rachel* *Dorota Sands* *Lukasz Wozniacki*
Portugal	Cystic Fibrosis Registry of Portugal	Luísa Pereira
Romania	4 individual centres: Clinical Children's Hospital Grigore Alexandrescu, Bukarest *Mother & Child Health Institute, Bukarest*	Liviu Pop Simona Mosescu *Suciu Nicolae* *Iustina Stan*

Country	Centre/National Registry name	Contact
Romania (cont.)	*Regional Cystic Fibrosis Centre Cluj, Cluj-Napoca*	*Şerban Radua* *Szabo Csilla-Enikö*
	National Cystic Fibrosis Centre, Timişoare	*Liviu Pop* *Ioana Ciuca*
Russian Federation	Cystic Fibrosis Registry of the Russian Federation	Nataliya Kashirskaya Elena Amelina Marina Starinova Elena Kondratyeva Stanislav Krasovskiy Anna Voronkova Nataliya Kashirskaya
Serbia	1 individual centre:	Milan Rodic
	National Centre for Cystic Fibrosis, Mother and Child Health Institute of Serbia "Dr Vukan Cupic", Belgrade	Predrag Minić Milan Rodić
Slovakia	6 individual centres:	Hana Kayserova
	Childrens CF Centre, DFN Banská Bystrica, Banská Bystrica	Branko Takáč
	Centrum cystickej fibrozy pre dospelych FNSP FDR, Banská Bystrica	Eva Bérešova
	Centrum cystickej fibrozy pre dospelych, Klinika pneumologie l.SZU a Univerzitna nemocnica, Bratislava	Marta Hajkova
	Klinika detskej pneumologie SZU UN Bratislava, pracovisko Podunajské Biskupice, Bratislava	Hana Kayserova Nina Bližňáková
	CF Adult centre, University Hospital L Pasteura, Košice	Lenka Kopčová
	Centrum cystickej fibrozy detí, Klinika detí a dorastu, Kosice	Anna Fetekeova Zuzana Hribíková
Slovenia	3 individual centres:	Uroš Krivec
	University Clinic of Pulmonary and Allergic Diseases, Golnik	Matjaž Fležar Tjaša Brus Pičman
	University Medical Centre Ljubljana, University Children`s Hospital, Unit for pulmonary diseases	Uroš Krivec Ana Kotnik Pirš
	University Medical Center Ljubljana, Department of Pulmonology and Allergy	*Izidor Kos* *Barbara Salobir*
Spain	22 individual centres:	Carlos Vazquez-Cordero
	Parc Taulí Hospital Universitario, Hospital de Sabadell, Unitat de Pneumologia Pediátrica i Unitat de Fibrosi Quística, Sabadell, Barcelona	Oscar Asensio de la Cruz Miguel Garcia Gonzàlez
	Hospital Sant Joan de Déu, Unitat de Pneumologia Pediátrica i Fibrosi Quística, Barcelona	María Cols Roig Jordi Costa i Colomer
	Hospital Vall d'Hebron, Unidad Fibrosis Quística y Neumologia Pediátrica, Barcelona	Silvia Gartner
	Hospital Universitario Reina Sofia, Dpto. Especialidades Médico-quirúrgicas, Área de Pediatría, Unidad de Alergia y Neumología Pediátricas, Unidad de Gestión Clínica de Pediatría y sus Especialidades, Cordoba	Javier Torres Borrego Noelia Sancho Montero
	Complejo Hospitalario Universitario Insular Materno Infantil, Las Palmas de Gran Canaria	Antonio José Aguilar Fernández

Country	Centre/National Registry name	Contact
Spain (cont.)	Hospital Universitario La Paz, Unidad de Fibrosis Quìstica Adultos, Servicio de Neumología, Madrid	Concha Prados
	Hospital Infantil La Paz, Unidad de Neumologìa Pediàtrica, Madrid	María Isabel Barrio Gomez de Agüero Marta Ruiz de Valbuena
	Hospital Universitario La Princesa, Neumologia Adultos, Madrid	Rosa María Girón
	Hospital Niño Jesús, Sección de Neumologia Pediátrica, Unidad de Fibrosis Quística, Madrid	José R. Villa Asensi Maribel Gonzàlez Álvarez
	Hospital Universitario de Ramón y Cajal, Unidad de Fibrosis Quística, Madrid	Adelaida Lamas Ferreiro Alejandro López Neyra Saioa Vicente Santamaria
	Hospital 12 de Octubre, Unidad de Fibrosis Quística, Madrid	Carmen Luna Paredes
	Hospital Regional Universitario de Málaga, Unidad Fibrosis Quística Adultos, Málaga	Casilda Olveira Fuster Gabriel María Olveira Fuster Nuria Porras Pèrez
	Hospital Regional Universitario de Málaga, Unidad de Fibrosis Quística Pediátrica, Málaga	Francisco Javier Pèrez -Frias Estela Pèrez -Ruiz
	Hospital Clínico Universitario Virgen de la Arrixaca, Unidad de Fibrosis Quística, Murcia	Pedro Mondéjar-López
	Hospital Universitario Central de Asturias, Unidad de Pediatría, Oviedo	Carlos Bousoño-García Ramon Gutierrez
	Hospital Universitario Virgen del Rocío, Unidad de Fibrosis Quística, Sevilla	Isabel Delgado Pecellín Esther Quintana Gallego
	Hospital Universitario Nuestra Señora de Candelaria, Santa Cruz de Tenerife, Tenerife	Alicia Callejon Orlando Mesa
	Hospital Clínico Universitario de Valencia, Unidad de Fibrosis Quística Pediátrica, Valencia	Amparo Escribano Montaner Silvia Castillo Corullón
	Hospital Universitario y Politécnico La Fe, Unidad de Trasplante Pulmonar y Fibrosis Quística, Valencia	Amparo Solé Jover Carmen Inés Perez Munoz
	Hospital Universitario de Cruces, Unidad de Fibrosis Quística, Vizkaya	Carlos Vazquez Cordero Maria Dolores Pastor
	Hospital Universitario Miguel Servet, Unidad de Neumología Pediátrica y Fibrosis Quística, Zaragoza	Carlos Martín de Vicente
	Hospital Universitario Miguel Servet, Unidad de Neumología y Fibrosis Quística (Adultos), Zaragoza	Maria Inés Herrero Labarga
Sweden	Cystic Fibrosis Registry of Sweden	Isabelle de Monestrol Anders Lindblad
Switzerland	19 individual centres:	Andreas Jung
	Kantonsspital Aarau AG, Klinik für Kinder und Jugendliche, Abteilung pädiatrische Pneumologie, Allergologie und Immunologie, Aarau	Dominik Müller-Suter Rachel Kusche
	Kantonsspital Aarau AG, Klinik für Pneumologie und Schlafmedizin, Aarau	Sarosh Irani
	Universitätsspital Basel, Klinik für Pneumologie, Adulte Cystische Fibrose, Basel	Michael Tamm Kathleen Jahn

Country	Centre/National Registry name	Contact
Switzerland *(cont.)*	UKBB Universitäts-Kinderspital beider Basel, Abteilung Intensivmedizin & Pneumologie, Basel	Jürg Hammer Daniel Trachsel
	Inselspital, Universitätsklinik für Pneumologie, Abteilung Cystische Fibrose, Bern	Thomas Geiser Dagmar Lin
	Lindenhofspital Quartier Bleu, Bern	Reta Fischer Iris Schmid Bernhard Schwizer Patrizia Bevilacqua
	Universitätsklinik für Kinderheilkunde, Zentrum für Cystische Fibrose und Pulmonologie, Inselspital, Bern	Philipp Latzin Romy Rodriguez Florian Singer
	Hôpital Cantonal Fribourg, Pädiatrie, Fribourg	Denise Herzog Johannes Wildhaber
	Hôpitaux Universitaires de Genève, Département de l'Enfant et de l'Adolescent, Unité de Pneumologie Pédiatrique, Genève	Constance Barazzone Anne Mornand
	Hôpitaux Universitaires de Genève, Département des Spécialités de Médecine, Service de Pneumologie, Consultation de Mucoviscidose Adulte, Genève	Paola Gasche Jérôme Plojoux
	Centre Hospitalier Universitaire Vaudois (CHUV), Départment Médico-Chirurgical de Pédiatrie, Pneumologie Pédiatrique et Mucoviscidose, Lausanne	Isabelle Rochat Laurence Mioranza
	Consultation Adulte de Mucoviscidose Service de Pneumologie, Département de Médecine, Centre Hospitalier Universitaire Vaudois (CHUV), Lausanne	Isabelle Huart Bellavere Isabelle Petigenet
	Luzerner Kantonsspital, Zentrum für Zystische Fibrose für Kinder und Erwachsene, Abteilungen für Pneumologie, Pädiatrische Pneumologie und Pädiatrische Gastroenterologie, Luzern	Nicolas Regamey Johannes Spalinger Christian Murer
	Hôpital Neuchâtelois – Pourtales, Consultation de Mucoviscidose Adulte, Neuchâtel	Alain Sauty Jean Marc Fellrath Marie Hofer
	Children's Hospital of Eastern Switzerland, Division of Paediatric Pulmonology & CF Centre, St Gallen	Jürg Barben Christine Gasser
	Kantonsspital St. Gallen, Lungenzentrum, Spezialsprechstunde für Adulte Cystische Fibrose, St. Gallen	Martin Brutsche Anna-Lena Walter Rebekka Kleiner
	Kantonsspital Winterthur, Klinik für Pneumologie und Klinik für Innere Medizin, Adulte Cystische Fibrose, Winterthur	Markus Hofer Thomas Hess
	Universitäts-Kinderspital Zürich, Abteilung für Pneumologie, Zürich	Andreas Jung Alexander Möller Demet Inci
	Universitätsspital Zürich, Klinik für Pneumologie, Adultes CF Zentrum, Zürich	Christian Benden Thomas Kurowski
Turkey	Cystic Fibrosis Registry of Turkey	Deniz Dogru
	Marmara University Faculty of Medicine, Division of Pulmonology, Istanbul	Bülent Karadağ Yasemin Gökdemir Ela Eralp

Country	Centre/National Registry name	Contact
Ukraine	1 individual centre: Cystic Fibrosis Centre of Western Ukrainian Specialized Children's Medical Centre, Lviv	Halyna Makukh Lyudmyla Bober Natalia Rohovyk
United Kingdom	UK Cystic Fibrosis Registry	Rebecca Cosgriff Susan Charman Elaine Gunn Siobhán Carr

Authors

For this report, the tables and graphs were written, commented and/or revised by:

Anna Zolin and **Annalisa Orenti**, Italy, ECFSPR Statisticians, Department of Clinical Sciences and Community Health, University of Milan;
Jacqui van Rens, Belgium, ECFSPR Executive Coordinator;
Alice Fox, Italy, **Marko Krasnyk**, Ukraine, ECFSPR Service Desk;
Andreas Jung, Switzerland, **Meir Mei-Zahav,** Israel, **Rebecca Cosgriff**, United Kingdom;
Valérie Storms, Belgium: CF Europe representative;
Contributing country managers and national representatives (the names are listed on page 5-11);
Lutz Naehrlich, Germany, ECFSPR Director.

Suggested citation for this report:

ECFSPR Annual Report 2017, Zolin A, Orenti A, Naehrlich L, van Rens J et al, 2019.

Introduction

The European Cystic Fibrosis Society Patient Registry (ECFSPR)

The ECFSPR collects demographic and clinical data of consenting cystic fibrosis (CF) patients 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 patient data from their own database and import the data into the ECFSPR software;
- Individual centres enter patient 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 anonymous, and only year/ month of birth and randomised centre numbers 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 request is approved or not; this decision is final. Requests originating from the 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

For the national registries it is possible that some of their definitions and data coding do not fully correspond to those employed by the ECFSPR, either because some types of information are not collected, or are collected by the national registry using a different method. When the national registries upload their data they are also asked to state in a document whether their variables definitions meet those of the ECFSPR. Where major discrepancies between the definitions occur, those variables have been omitted from the annual report, and 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 2 on page 144). 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. Where this is the case, a footnote has been added to the relevant tables and graphs.

If a country does not collect a certain variable (or if it is completely different from the ECFSPR definitions as described above), we have omitted that country from the relevant graphs in the report. The same applies for countries where the information for a variable is missing for more than 10% of the patients. All data, however, is presented in the tables. The number of missing values is important for the interpretation of the results, since it is impossible to know if a patient with a missing value for a given complication has

this complication or not, which makes the given frequencies less accurate. For example, in a country where 7% of the patients have liver disease but 20% of patients have unknown/missing information on liver disease, the true frequency of liver disease can 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 at 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 patients 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 percentage of people with CF, per country, included in the national registry or national data presented by the country, varies; see table 1.1, page 18. These differences can influence how the data is interpreted, and we therefore advise comparisons to be made only between countries with a similar percentage coverage.

Glossary and Abbreviations

Country codes:

AL:	Albania
AM:	Armenia
AT:	Austria
BE:	Belgium
BG:	Bulgaria
CH:	Switzerland
CZ:	Czech Republic
DE:	Germany
DK:	Denmark
ES:	Spain
FR:	France
GR:	Greece
HR:	Croatia
HU:	Hungary
IE:	Ireland
IL:	Israel
IT:	Italy
LT:	Lithuania
LU:	Luxembourg
LV:	Latvia
MD:	Republic of Moldova
MK:	Republic of North Macedonia
NL:	The Netherlands
NO:	Norway
PL:	Poland
PT:	Portugal
RO:	Romania
RS:	Serbia
RU:	Russian Federation
SE:	Sweden
SI:	Slovenia
SK:	Slovak Republic
TR:	Turkey
UA:	Ukraine
UK:	United Kingdom

Explanation of terms:

ABPA: allergic bronchopulmonary aspergillosis, an allergic reaction 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.
FEV_1: forced expiratory volume in one second (lung function parameter).
FEV_1%: the FEV_1 as a percentage of the average value for healthy people of the same age, height and sex.
Haemoptysis: coughing up blood. This happens frequently in small amounts in CF, so the complication we asked for here is major bleeding (more than 250 ml).
Homozygous: CF is caused by mutations of the CFTR gene, one on each allele. One is inherited from the mother and one from the father. If both mutations are the same, the person is said to be homozygous for this mutation.
Heterozygous: CF is caused by mutations of the CFTR gene, one on each allele. One is inherited from the mother and one from the father. If these are two different mutations, the person is considered to be heterozygous.
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: congenital obstruction of the gut with thick, sticky faeces.
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 was missing.
NaCl: sodium chloride. Here: inhaled hypertonic saline.
Pancreatic insufficiency: the absence of pancreatic enzymes in the gut leading to malnutrition if not treated (pancreatic insufficiency is therefore defined as the use of pancreatic enzyme supplementation).
25th Pctl: 25th percentile, also called first quartile. It is the value that separates the set of measurements in two parts, so that one quarter (25%) of the measurements is below it and the other three quarters are above it. For example, if the 25th percentile for age at diagnosis is 1 month, it means that a quarter of the patients were diagnosed before they were a month old, and the other three quarters were diagnosed after they were a month old.
50th Pctl: 50th percentile, also called second quartile or median (please refer to definition of Median).
75th Pctl: 75th percentile, also called third quartile. It is the value that separates the set of measurements in two parts, so that three quarters (75%) are below it and the other quarter is above it. For example, if the 75th percentile for age at diagnosis is 3 years, it means that three quarters of the patients are diagnosed before they were 3 years old, and the remaining quarter was diagnosed after they reached 3 years of age.
Pneumothorax: collapsed lung, in CF usually because of severe lung damage.
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): they are a way to compare results from a test to a "normal" population, to give scores (or data-values) a common standard: a mean of 0 and a standard deviation of 1 to indicate how far a value is from the mean value of a reference population (see Appendix 1 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. 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 years old boy is -2, it means that the BMI for that boy is 2 standard deviations below the mean BMI of 10 years old boys of the reference population.

Summary of data report

Outcome		Females	Males	Total
Patients registered in the ECFSPR	n (%)	22847 (47.4)	25357 (52.6)	48204
Age at follow-up (in years; patients alive on 31/12/2017)	mean median	20.4 17.9	21.2 19.0	20.8 18.5
Patients ≥ 18 years (patients alive on 31/12/2017)	%	49.8	52.6	51.30
Age at diagnosis*	mean (years) median (months)	4.2 4.0	4.0 4.0	4.1 4.0
Patients with at least one F508del allele recorded*	%	81.2	81.0	81.1
Patients living with lung transplant*	n (%)	1296 (5.9)	1293 (5.3)	2589 (5.6)
Patients living with liver transplant*	n (%)	97 (0.44)	172 (0.71)	269 (0.58)
Patients deceased in 2017**	n (%)	237 (1.05)	225 (0.90)	462 (0.97)
Age at death (years)**	mean median	30.5 29.0	31.9 31.0	31.2 29.0

* Only patients seen during the year are presented. The total number of patients presented is 46,832.

** Only patients seen during the year are presented. For the United Kingdom, all patients with confirmed diagnosis of CF were included (N=10,468). The total number of patients presented is 47,413.

Data report

1. Demographics

Figure 1.1 Map of countries that contributed to the ECFSPR in year 2017.

Countries that contributed 2017 data are marked in blue.

Table 1.1 Number of patients in year 2017, by country.

Country	Patients registered, not lost to follow-up	Patients seen	Estimated coverage 2017
Albania	123	122	80%
Armenia	33	32	>70%
Austria	800	757	>90%
Belgium*	1319	1287	>90%
Bulgaria	155	148	>70%
Croatia	93	87	70%
Czech Republic*	619	605	>95%
Denmark*	510	496	99%
France*	6940	6940	>90%
Germany*	6119	6119	>80%
Greece**	621	599	>95%
Hungary*	507	504	>95%
Ireland*	1284	1219	>90%
Israel**	597	547	>95%
Italy*	5565	5561	>95%
Latvia	41	39	>90%
Lithuania[1]	14	14	20%[1]
Luxembourg	36	36	>95%
Rep of Moldova	62	50	>90%
The Netherlands*	1473	1470	>95%
Rep of North Macedonia	119	115	>90%
Norway*	254	251	>70%
Poland	721	656	>35%
Portugal**	341	327	>95%
Romania	167	159	35%
Russian Federation*	3269	3080	95%
Serbia	196	172	>90%
Slovak Republic**	294	266	>90%
Slovenia	112	109	>95%
Spain	2075	2002	75%
Sweden*	686	686	>95%
Switzerland**	963	914	>95%
Turkey	1447	1411	>40%
Ukraine	181	165	>20%
United Kingdom*[2]	10468[2]	9887	>95%
Total	48204	46832	

* Countries with an established national CF registry.

** These countries are defined as a national registry, because all centres participate in the ECFSPR and use the direct data-entry function of ECFSTracker.

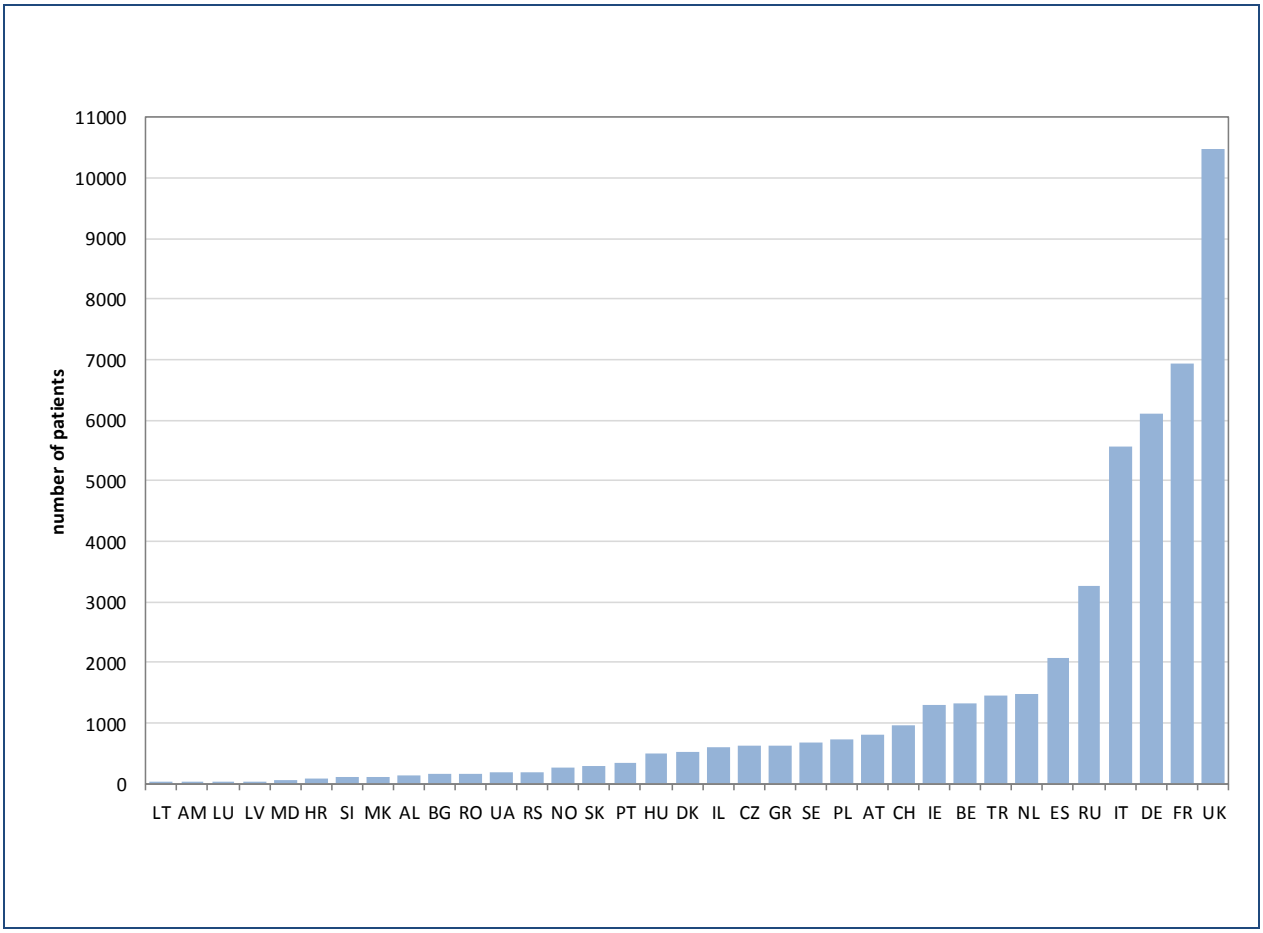
1 Coverage is 100% for adults and 0% for children.

2 The number of registered patients in this report differs from the number 10,469 reported in the UK 2017 annual data report, because additional data cleaning was done.

[Table 1.1 continued]

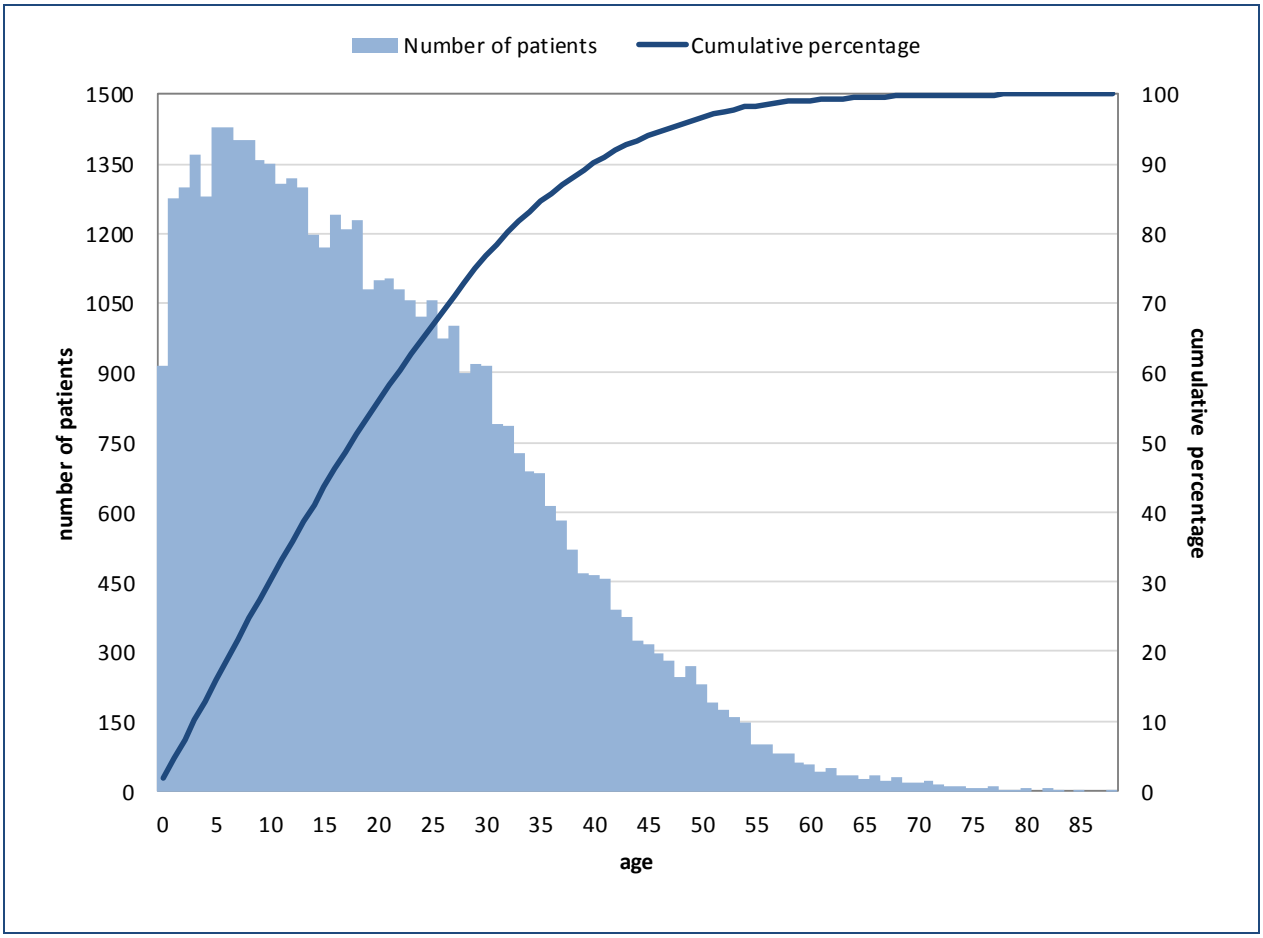
The column “Patients registered, not lost to follow-up” shows the patients that attend centres and includes patients that have not been seen during the year but are known to be alive that year. The column “patients seen” presents only the patients who have attended the clinic during the year. The column “Estimated coverage 2017” shows the estimated percentage of CF patients 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 Luxembourg.

Figure 1.2 Number of patients registered in the ECFSPR in year 2017, by country.



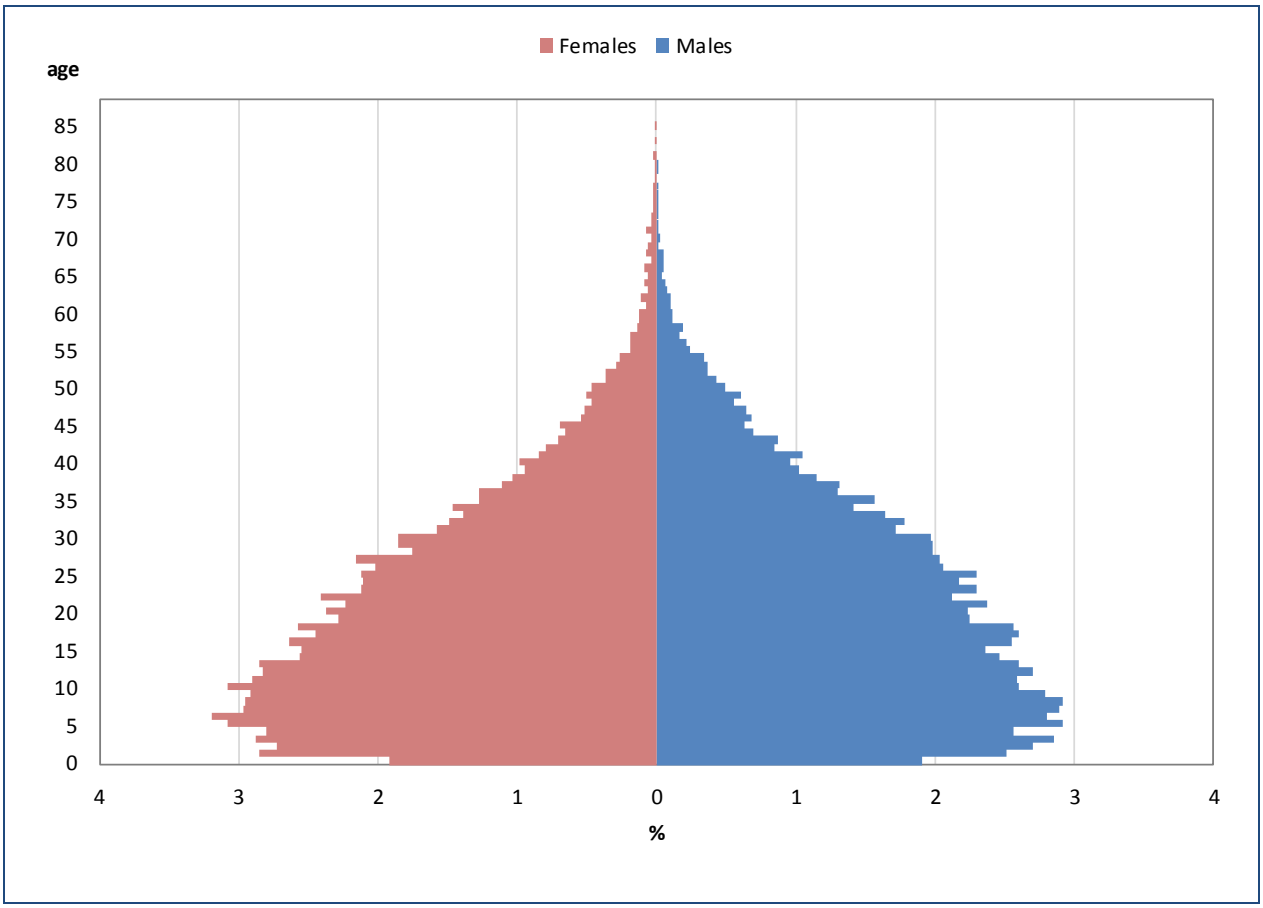
Each vertical bar shows the number of patients living in that country in 2017. Please refer to table 1.1 for the coverage in each country.

Figure 1.3 Age at follow-up distribution. Patients alive on 31/12/2017.



Each blue vertical bar represents the number of patients of that age alive in 2017. 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 18.5 years of age).

Figure 1.4 Age at follow-up distribution by sex. Patients alive on 31/12/2017.



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 4.12 years, see table 2.1).

Table 1.2 Proportion of adults (≥18 years) and children (<18 years), by country. Patients registered, alive on 31/12/2017.

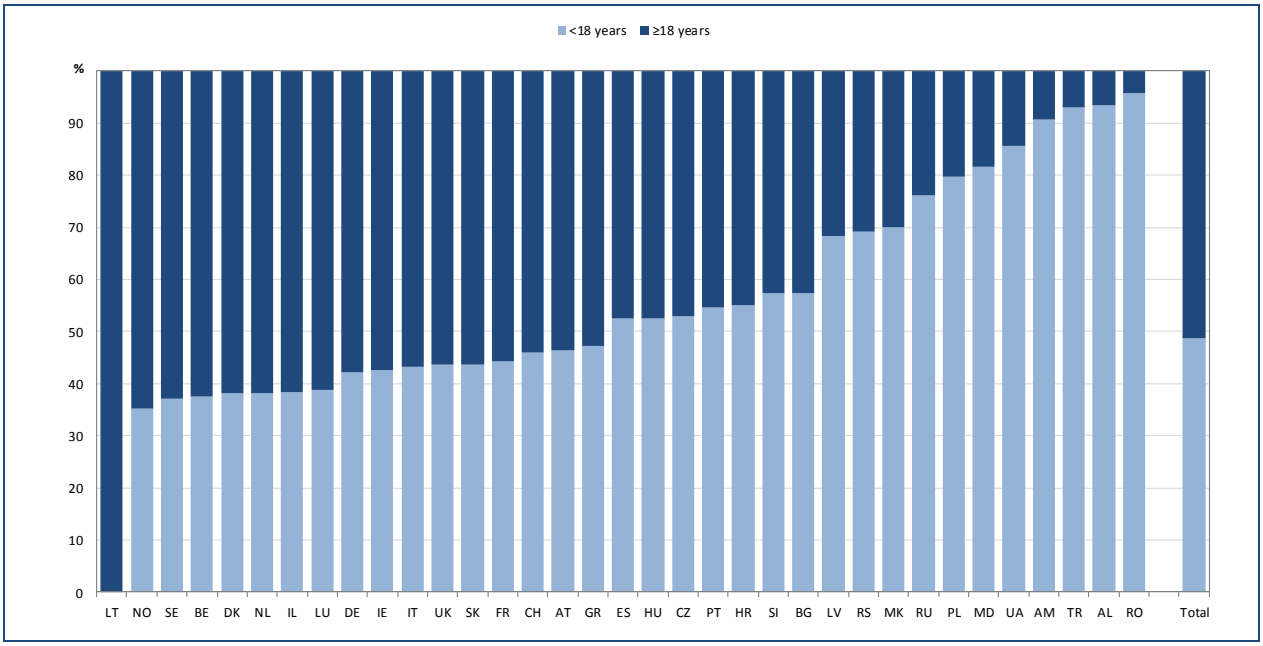
Country	Children (<18 years) number (%)	Adults (≥18 years) number (%)
Albania	115 (93.50)	8 (6.50)
Armenia	29 (90.63)	3 (9.38)
Austria	370 (46.42)	427 (53.58)
Belgium	492 (37.47)	821 (62.53)
Bulgaria	86 (57.33)	64 (42.67)
Croatia	50 (54.95)	41 (45.05)
Czech Republic	324 (52.94)	288 (47.06)
Denmark	191 (38.05)	311 (61.95)
France	3051 (44.32)	3833 (55.68)
Germany	2559 (42.15)	3512 (57.85)
Greece	290 (47.15)	325 (52.85)
Hungary	264 (52.59)	238 (47.41)
Ireland	539 (42.54)	728 (57.46)
Israel	227 (38.34)	365 (61.66)
Italy	2379 (43.14)	3136 (56.86)
Latvia	28 (68.29)	13 (31.71)
Lithuania	0 (0.00)	14 (100.00)
Luxembourg	14 (38.89)	22 (61.11)
Rep of Moldova	49 (81.67)	11 (18.33)
The Netherlands	559 (38.18)	905 (61.82)
North Macedonia	82 (70.09)	35 (29.91)
Norway	89 (35.18)	164 (64.82)

Note: Lithuania has 100% coverage for adults and 0% coverage for children.

[table 1.2 continued]

Country	Children (<18 years) number (%)	Adults (≥18 years) number (%)
Poland	575 (79.75)	146 (20.25)
Portugal	185 (54.57)	154 (45.43)
Romania	158 (95.76)	7 (4.24)
Russian Federation	2452 (76.13)	769 (23.87)
Serbia	134 (69.07)	60 (30.93)
Slovak Republic	128 (43.69)	165 (56.31)
Slovenia	63 (57.27)	47 (42.73)
Spain	1082 (52.45)	981 (47.55)
Sweden	254 (37.19)	429 (62.81)
Switzerland	439 (45.92)	517 (54.08)
Turkey	1329 (93.00)	100 (7.00)
Ukraine	155 (85.64)	26 (14.36)
United Kingdom	4510 (43.63)	5826 (56.37)
Total	23251 (48.70)	24491 (51.30)

Figure 1.5 Proportion of adults (≥18 years) and children (<18 years). Patients alive on 31/12/2017.



Note: Lithuania has 100% coverage for adults and 0% coverage for children.

This graph shows the percentage of patients in each country who are adults (dark blue) or children (light blue). The percentage of adult patients varies considerably between the different countries, but this is partly an effect of the way the patients are included: for some countries only a few individual centres send data to the ECFSPR, and the proportion of children and adults may reflect the proportion of paediatric and adult centres in that country who participate in the ECFSPR. Please refer to table 1.1, page 18, for national coverage.

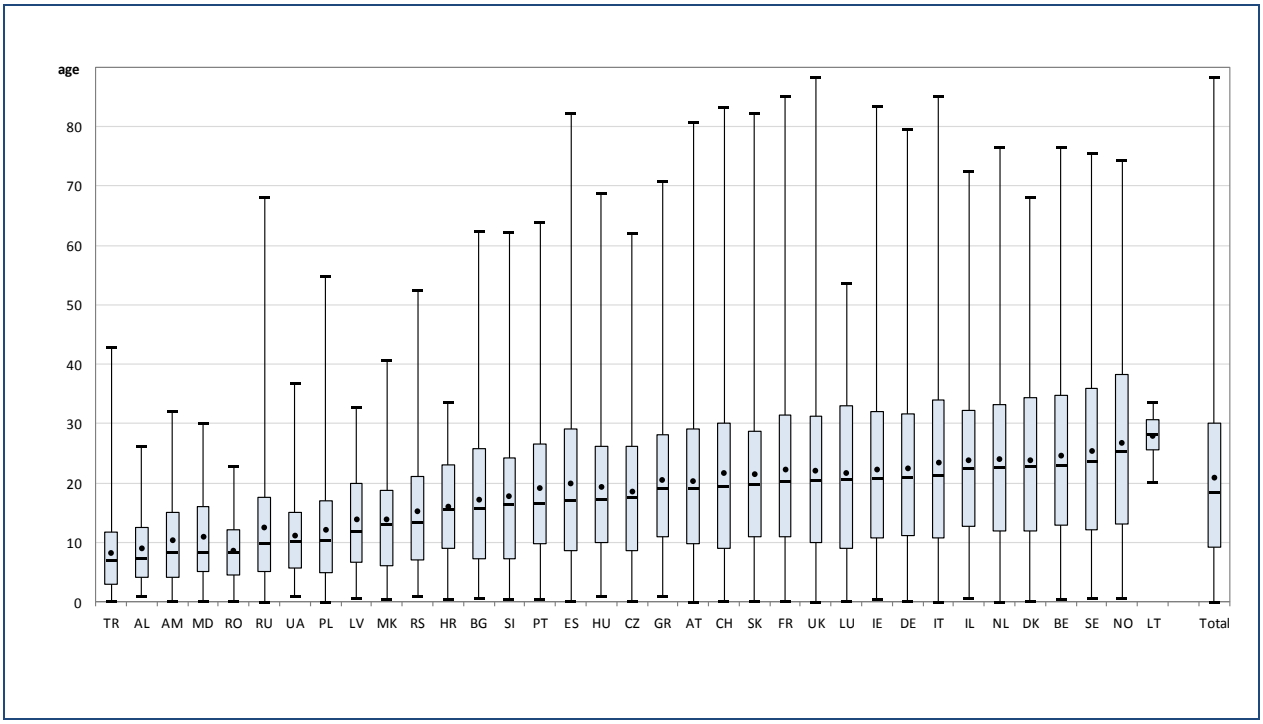
Table 1.3 Age at follow-up: descriptive statistics, by country and overall. Patients registered, alive on 31/12/2017.

Country	N	Mean (average age)	Min (age of the youngest patient)	25th pctl (25% of the patients are younger than this age)	Median (half the patients are younger than this age)	75th pctl (75% of the patients are younger than this age)	Max (age of the oldest patient)
Albania	123	8.8	0.9	4.2	7.4	12.5	26.0
Armenia	32	10.2	0.2	4.1	8.3	15.0	32.2
Austria	797	20.2	0.0	9.8	19.1	29.0	80.7
Belgium	1313	24.6	0.3	12.9	22.9	34.7	76.5
Bulgaria	150	17.1	0.5	7.3	15.8	25.8	62.3
Croatia	91	16.0	0.3	9.0	15.5	23.1	33.5
Czech Republic	612	18.4	0.1	8.6	17.5	26.1	62.0
Denmark	502	23.8	0.1	12.0	22.8	34.4	68.0
France	6884	22.2	0.1	10.9	20.3	31.5	85.1
Germany	6071	22.4	0.1	11.2	20.9	31.6	79.5
Greece	615	20.5	0.8	10.9	19.0	28.2	70.8
Hungary	502	19.2	0.9	10.0	17.3	26.2	68.9
Ireland	1267	22.2	0.3	10.8	20.8	32.0	83.4
Israel	592	23.8	0.5	12.7	22.4	32.3	72.5
Italy	5515	23.3	0.0	10.7	21.2	34.0	85.1
Latvia	41	13.8	0.5	6.6	11.9	19.9	32.7
Lithuania	14	27.8	20.1	25.5	28.2	30.7	33.5
Luxembourg	36	21.5	0.1	9.0	20.6	32.9	53.5
Rep of Moldova	60	10.9	0.2	5.0	8.4	16.0	30.0
The Netherlands	1464	24.0	0.0	12.0	22.5	33.1	76.5
North Macedonia	117	13.8	0.3	6.1	12.9	18.8	40.8
Norway	253	26.7	0.6	13.0	25.3	38.2	74.3
Poland	721	12.1	0.0	4.9	10.3	17.0	54.7
Portugal	339	19.1	0.4	9.8	16.5	26.6	64.0
Romania	165	8.6	0.2	4.6	8.4	12.2	22.8
Russian Federation	3221	12.4	0.0	5.1	9.9	17.5	68.0
Serbia	194	15.2	0.9	7.0	13.4	21.0	52.4
Slovak Republic	293	21.3	0.2	11.0	19.9	28.7	82.2
Slovenia	110	17.6	0.4	7.3	16.4	24.3	62.1
Spain	2063	19.8	0.1	8.6	17.0	29.0	82.2
Sweden	683	25.2	0.5	12.2	23.6	36.0	75.5
Switzerland	956	21.5	0.1	9.0	19.5	30.0	83.3
Turkey	1429	8.2	0.1	3.0	7.0	11.8	42.8
Ukraine	181	11.1	0.9	5.7	10.0	15.0	36.8
United Kingdom	10336	22.0	0.0	9.9	20.5	31.3	88.4
Total	47742	20.8	0.0	9.1	18.5	30.0	88.4

Note: Lithuania has 100% coverage for adults and 0% coverage for children.

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 December 2017 are included.

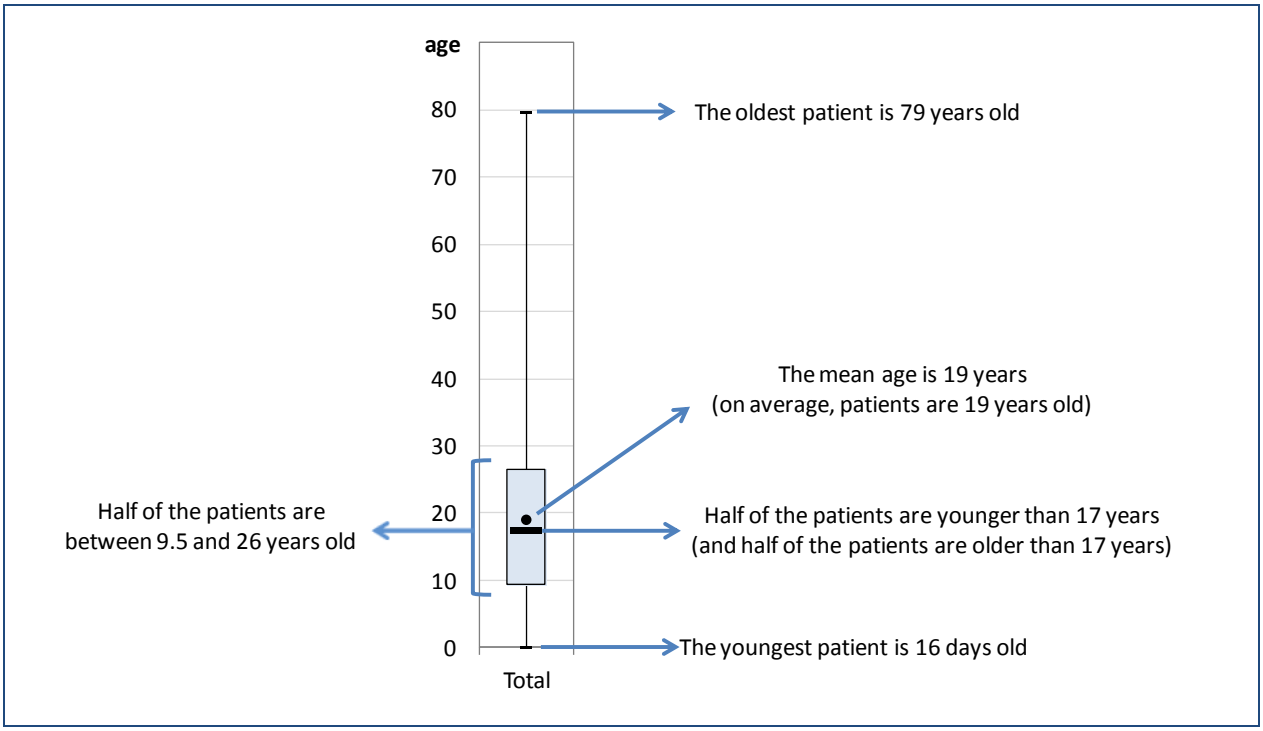
Figure 1.6 Age at follow-up: box-plot, by country and overall. Patients alive on 31/12/2017.



Note: Lithuania has 100% coverage for adults and 0% coverage for children.

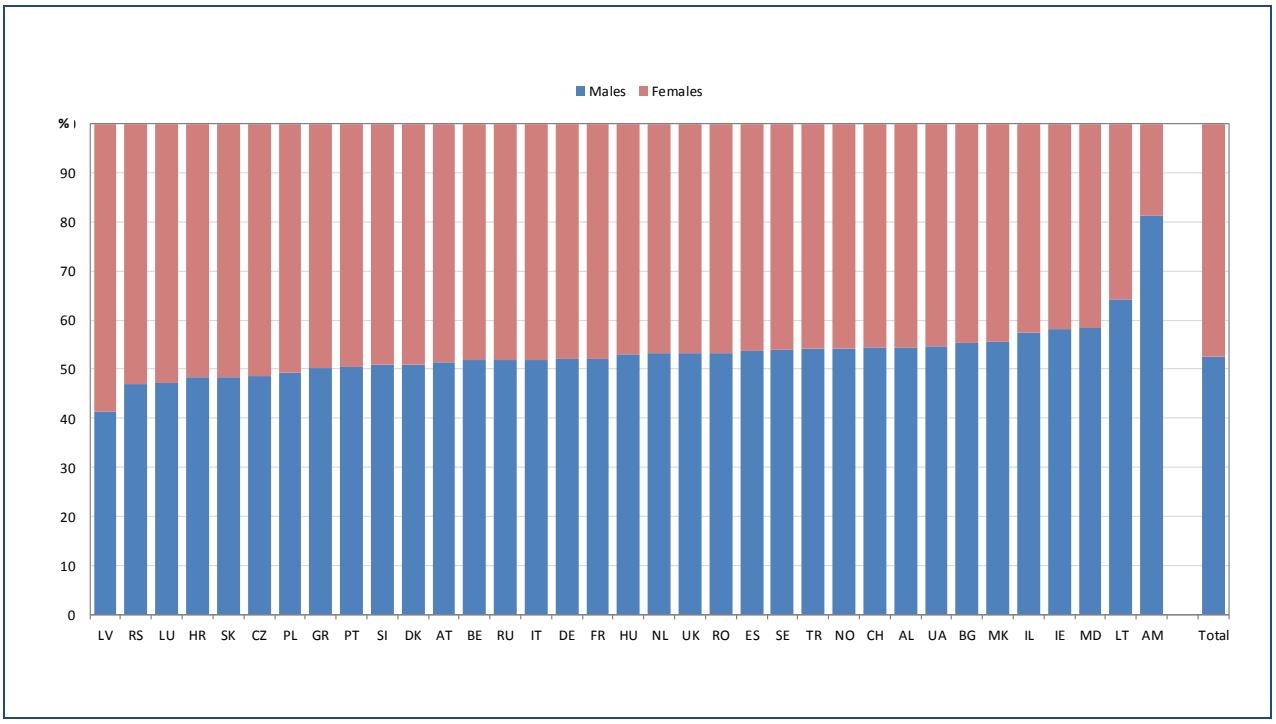
This box-plot is a graphic representation of the age detailed in table 1.3. For each country the dash (black line crossing the blue box) is the median, the black dot is the mean and the whiskers (vertical lines with a T-shaped end) are the minimum and the maximum.

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



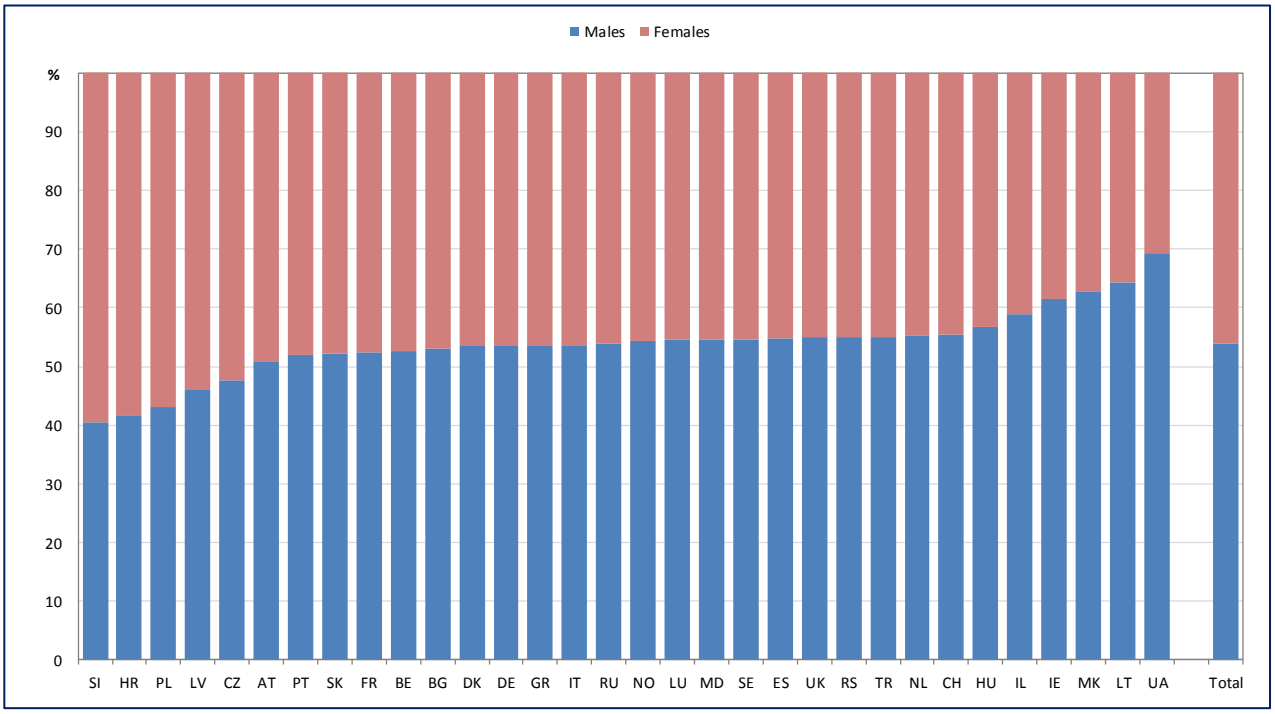
Note: This is an example of how to read a box-plot. The numbers used in this figure are not real and do not refer to figure 1.6.

Figure 1.7 Sex distribution, by country and overall. Patients alive on 31/12/2017.



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 31/12/2017 and aged 18 years or more.



Note: Albania, Armenia and Romania have only few patients aged 18 years or more and are excluded from this graph.

Sex distribution for adult patients. 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.

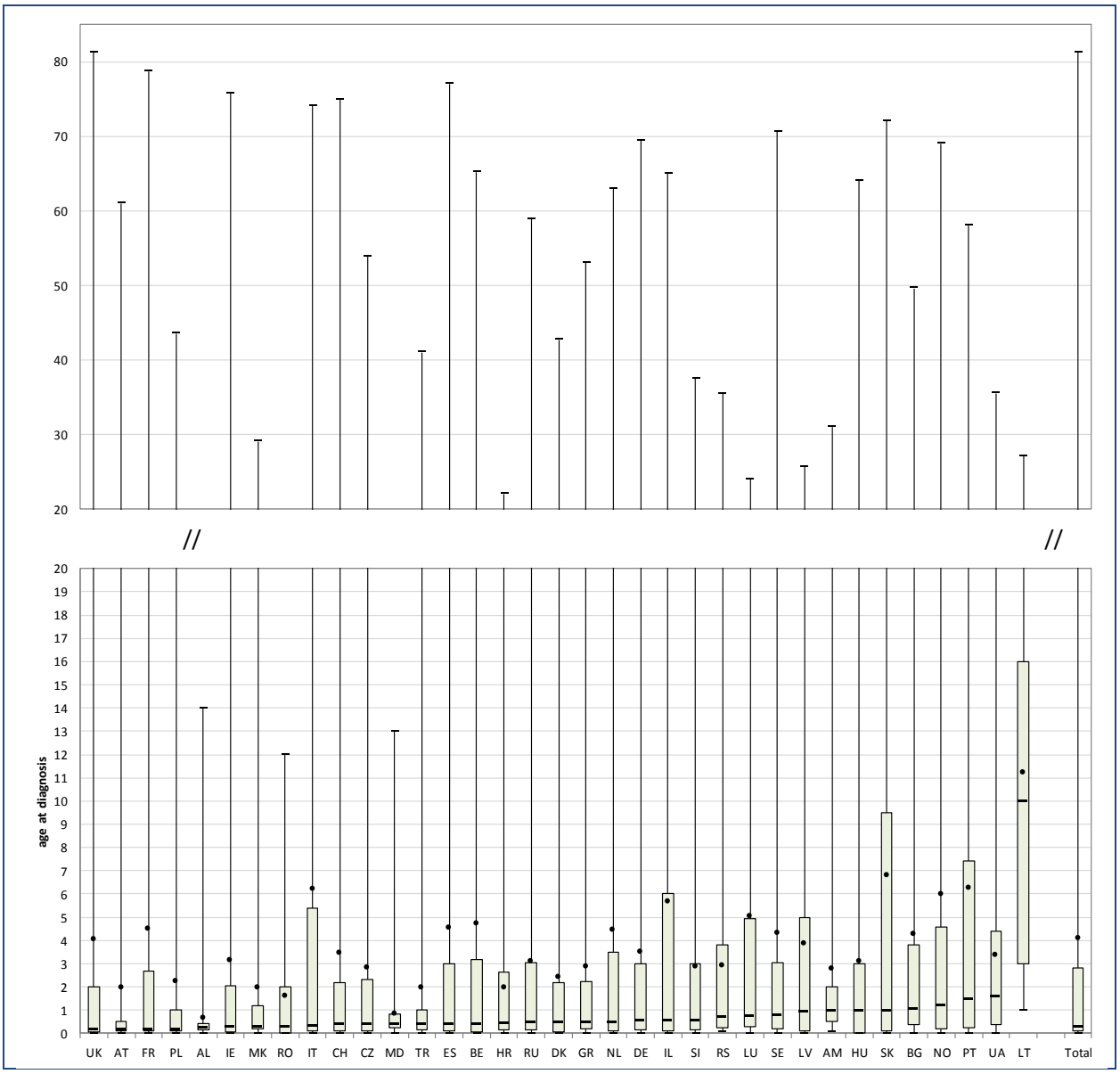
Table 2.1 Age at diagnosis (in years): descriptive statistics, by country and overall. All patients seen in 2017.

Country	N	N miss	Mean (average age at diagnosis)	Min (lowest age at diagnosis)	25th pctl (25 % of the patients were diagnosed before this age)	Median (half the patients were diagnosed before this age)	75th pctl (75% of the patients were diagnosed before this age)	Max (highest age at diagnosis)
Albania	122	0	0.66	0.0	0.16	0.25	0.41	14.00
Armenia	31	1	2.81	0.1	0.50	1.00	2.00	31.00
Austria	685	72	1.98	0.0	0.10	0.20	0.50	61.00
Belgium	1284	3	4.75	0.0	0.08	0.42	3.18	65.24
Bulgaria	145	3	4.27	0.0	0.40	1.10	3.80	49.60
Croatia	84	3	1.98	0.0	0.17	0.46	2.63	22.00
Czech Republic	597	8	2.86	0.0	0.10	0.40	2.30	53.90
Denmark	496	0	2.42	0.0	0.08	0.50	2.17	42.67
France	6870	70	4.53	0.0	0.10	0.20	2.70	78.70
Germany	5857	262	3.53	0.0	0.17	0.59	3.00	69.41
Greece	589	10	2.90	0.0	0.22	0.50	2.25	53.00
Hungary	439	65	3.09	0.0	0.00	1.00	3.00	64.00
Ireland	1215	4	3.17	0.0	0.06	0.30	2.05	75.83
Israel	541	6	5.70	0.0	0.10	0.60	6.00	65.00
Italy	5510	51	6.22	0.0	0.11	0.35	5.39	74.15
Latvia	39	0	3.90	0.0	0.10	0.95	5.00	25.70
Lithuania	13	1	11.24	1.0	3.00	10.00	16.00	27.10
Luxembourg	36	0	5.05	0.0	0.30	0.77	4.95	24.00
Rep of Moldova	50	0	0.86	0.0	0.25	0.40	0.83	13.00
The Netherlands	1362	108	4.45	0.0	0.10	0.50	3.50	63.00
North Macedonia	115	0	1.97	0.0	0.20	0.30	1.20	29.00
Norway	237	14	5.99	0.0	0.20	1.20	4.60	69.00
Poland	655	1	2.24	0.0	0.10	0.20	1.00	43.50
Portugal	316	11	6.25	0.0	0.25	1.50	7.40	58.00
Romania	154	5	1.60	0.0	0.00	0.30	2.00	12.00
Russian Federation	3061	19	3.11	0.0	0.15	0.49	3.05	58.91
Serbia	168	4	2.91	0.1	0.25	0.70	3.80	35.40
Slovak Republic	227	39	6.82	0.0	0.11	1.00	9.50	72.00
Slovenia	107	2	2.89	0.0	0.16	0.60	3.00	37.50
Spain	1963	39	4.56	0.0	0.10	0.41	3.00	77.00
Sweden	670	16	4.32	0.0	0.19	0.79	3.04	70.61
Switzerland	790	124	3.48	0.0	0.10	0.40	2.20	75.00
Turkey	1395	16	1.99	0.0	0.17	0.40	1.00	41.00
Ukraine	165	0	3.39	0.0	0.40	1.60	4.40	35.50
United Kingdom	9791	96	4.06	0.0	0.05	0.17	2.00	81.35
Total	45779	1053	4.12	0.0	0.10	0.33	2.83	81.35

Note: For Austria, Hungary, Slovak Republic and Switzerland the information on age at diagnosis is missing for more than 10% of the patients.
Lithuania has 100% coverage for adults and 0% coverage for children.

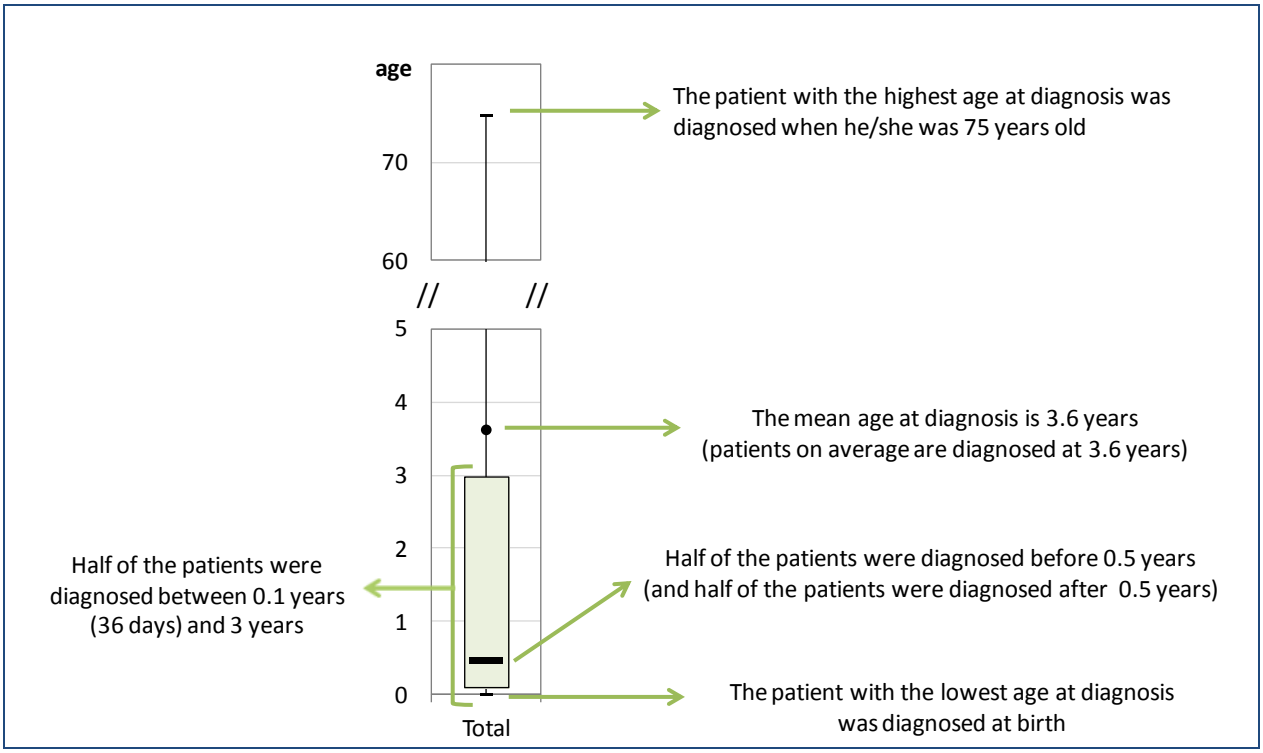
Table 2.1 shows the descriptive statistics for age at diagnosis by country and overall. For prenatal diagnoses (children diagnosed before birth), the age at diagnosis has been set to 0.

Figure 2.1 Age at diagnosis (in years): box-plot, by country and overall. All patients seen in 2017.



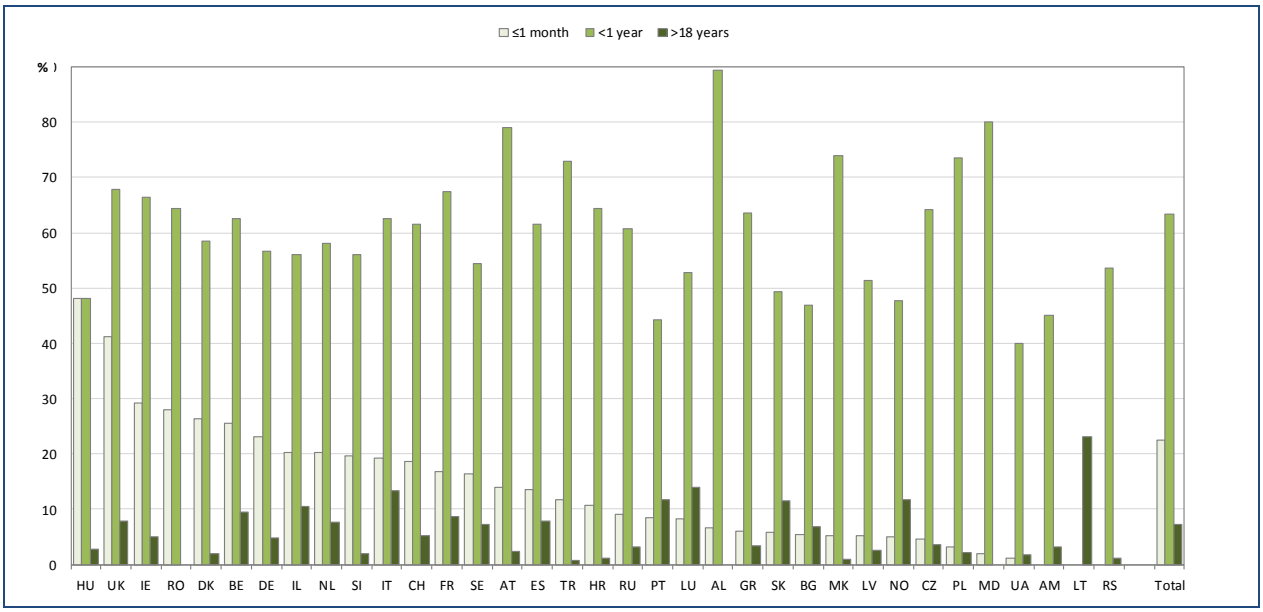
Note: For Austria, Hungary, Slovak Republic and Switzerland the information on age at diagnosis is missing for more than 10% of the patients.
Lithuania has 100% coverage for adults and 0% coverage for children.

This box-plot is a graphic representation of age at diagnosis as detailed in table 2.1. For each country the dash (black line crossing the blue box) is the median, the black dot is the mean and the whiskers (vertical lines with a T-shaped end) are the minimum and the maximum. Please note that the vertical axis is interrupted to emphasise the change of scale in the upper part of the graph. The figure on the next page explains how to read the box-plot.



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

Figure 2.2 Proportion of patients diagnosed at age 1 month or younger, younger than 1 year and older than 18 years, by country and overall. All patients seen in 2017.

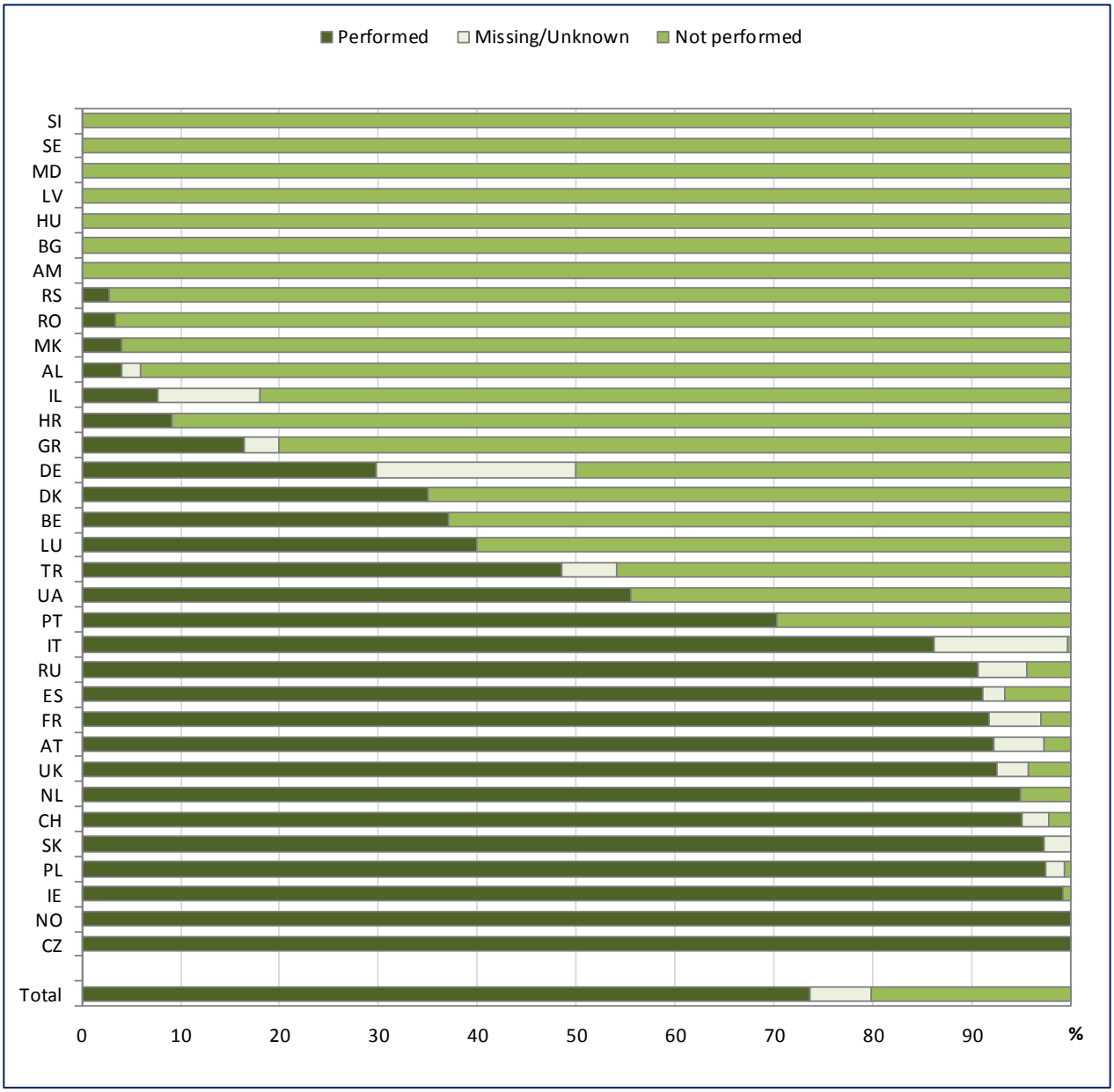


Note: For Austria, Hungary, Slovak Republic and Switzerland the information on age at diagnosis is missing for more than 10% of the patients.
Lithuania has 100% coverage for adults and 0% coverage for children.

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 diagnoses included in the sub-group for within 1 month are also part of the diagnoses in the sub-group for 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 up to 100%.

Figure 2.3 Proportion of patients who underwent neonatal screening, by country and overall. Patients 5 years old or younger seen in 2017.

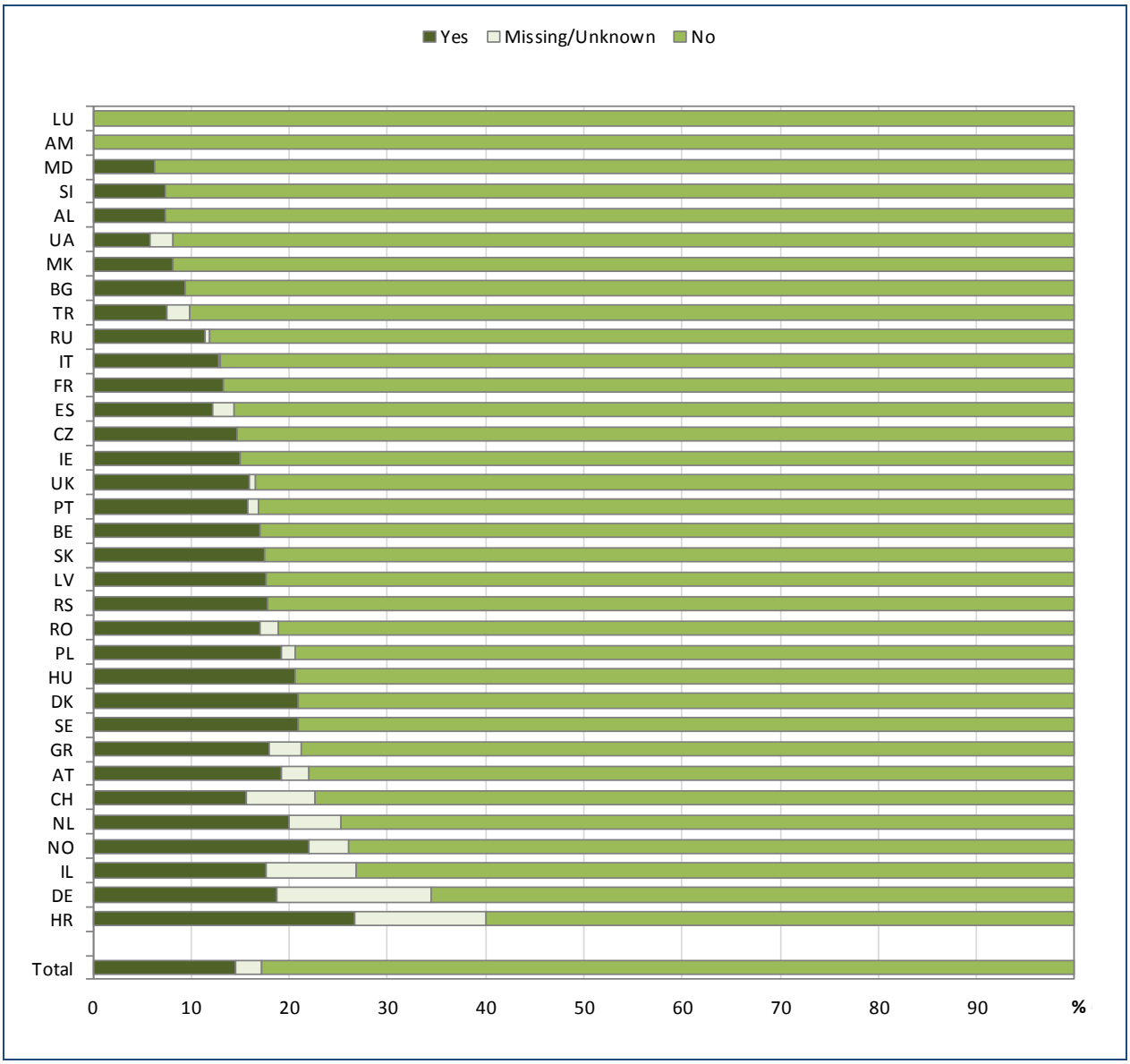


Note: For Germany, Israel and Italy the information on neonatal screening is missing for more than 10% of the patients.
Lithuania: 0% coverage for children, therefore the country was excluded from this graph.
For Belgium, Czech Republic, France and UK positive answers ("neonatal screening performed") are reported 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 2017 who were screened at birth, (see country specific notes above). Dark green horizontal bars represent neonatal screening "performed", light green ones "not performed".

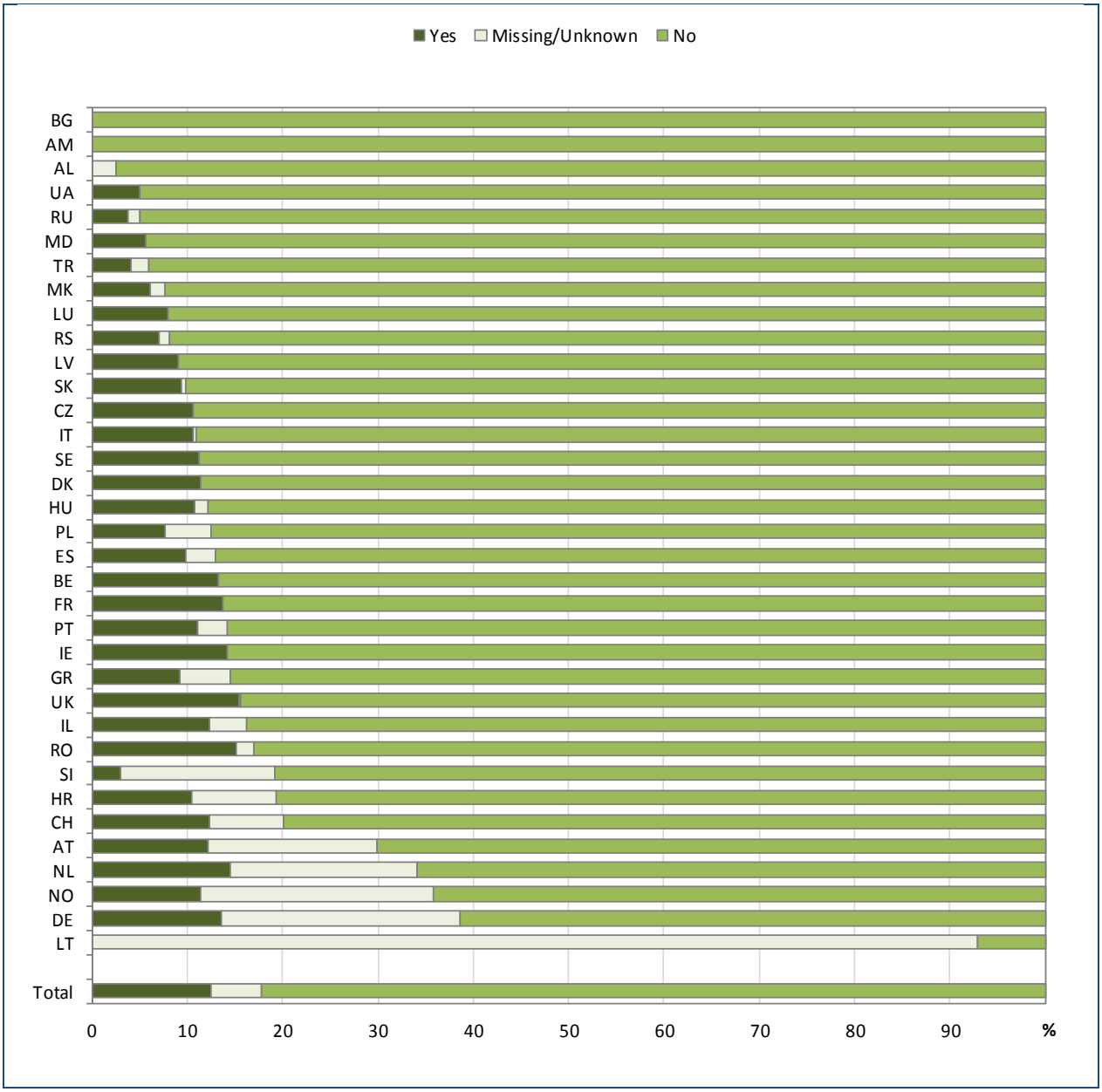
This graph shows that, in the five years previous to 2017, in many countries the CF patients underwent newborn screening and that in some countries there is no neonatal screening programme. In total, 74% of all children of 5 years old or younger registered in the ECFSPR in 2017 were screened at birth. 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 Croatia and Germany the information on meconium ileus is missing for more than 10% of the patients aged 10 years or younger.
Lithuania: 0% coverage for children, therefore this country was excluded from this graph.

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



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

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 (>10 years). This difference is not due to 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 2017). 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 said to be 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.

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 2017.

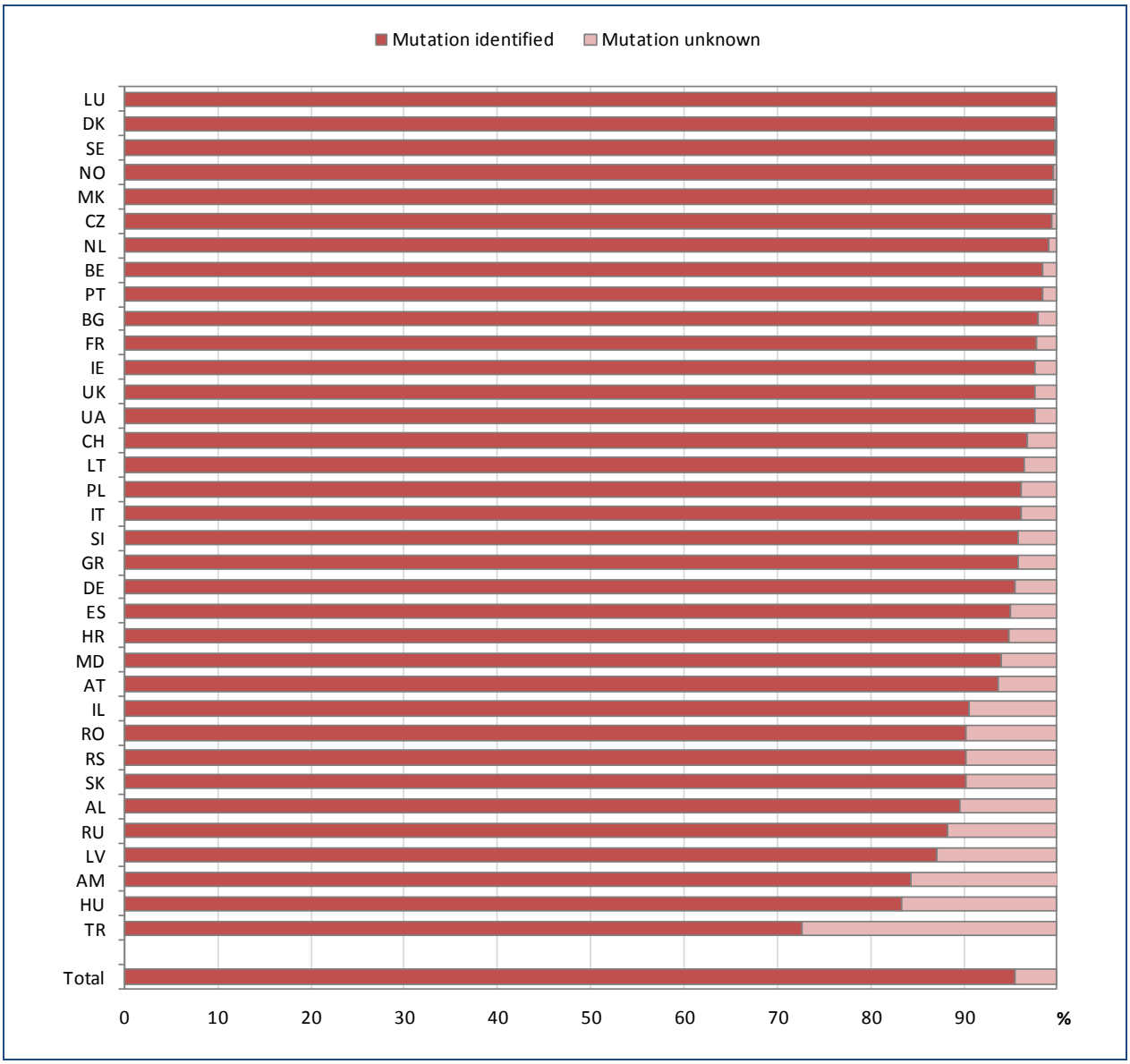
Country	N	Genotyping		Among genotyping done	
		not done	done	two mutations identified	at least one mutation unknown
		number (%)	number (%)	number (%)	number (%)
Albania	122	12 (9.84)	110 (90.16)	91 (82.73)	19 (17.27)
Armenia	32	0 (0)	32 (100)	23 (71.88)	9 (28.13)
Austria	757	2 (0.26)	755 (99.74)	688 (91.13)	67 (8.87)
Belgium	1287	0 (0)	1287 (100)	1255 (97.51)	32 (2.49)
Bulgaria	148	0 (0)	148 (100)	142 (95.95)	6 (4.05)
Croatia	87	0 (0)	87 (100)	80 (91.95)	7 (8.05)
Czech Republic	605	1 (0.17)	604 (99.83)	598 (99.01)	6 (0.99)
Denmark	496	0 (0)	496 (100)	495 (99.80)	1 (0.20)
France	6940	0 (0)	6940 (100)	6722 (96.86)	218 (3.14)
Germany	6119	11 (0.18)	6108 (99.82)	5658 (92.63)	450 (7.37)
Greece	599	0 (0)	599 (100)	561 (93.66)	38 (6.34)
Hungary	504	4 (0.79)	500 (99.21)	380 (76.00)	120 (24.00)
Ireland	1219	0 (0)	1219 (100)	1175 (96.39)	44 (3.61)
Israel	547	0 (0)	547 (100)	479 (87.57)	68 (12.43)
Italy	5561	27 (0.49)	5534 (99.51)	5160 (93.24)	374 (6.76)
Latvia	39	0 (0)	39 (100)	29 (74.36)	10 (25.64)
Lithuania	14	0 (0)	14 (100)	13 (92.86)	1 (7.14)
Luxembourg	36	0 (0)	36 (100)	36 (100)	0 (0)
Rep of Moldova	50	0 (0)	50 (100)	44 (88.00)	6 (12.00)
The Netherlands	1470	15 (1.02)	1455 (98.98)	1434 (98.56)	21 (1.44)
North Macedonia	115	1 (0.87)	114 (99.13)	113 (99.12)	1 (0.88)
Norway	251	0 (0)	251 (100)	250 (99.60)	1 (0.40)

[table 3.1 continued]

Country	N	Genotyping		Among genotyping done	
		not done	done	two mutations identified	at least one mutation unknown
		number (%)	number (%)	number (%)	number (%)
Poland	656	0 (0)	656 (100)	622 (94.82)	34 (5.18)
Portugal	327	0 (0)	327 (100)	320 (97.86)	7 (2.14)
Romania	159	0 (0)	159 (100)	137 (86.16)	22 (13.84)
Russian Federation	3080	222 (7.21)	2858 (92.79)	2297 (80.37)	561 (19.63)
Serbia	172	3 (1.74)	169 (98.26)	141 (83.43)	28 (16.57)
Slovak Republic	266	0 (0)	266 (100)	222 (83.46)	44 (16.54)
Slovenia	109	1 (0.92)	108 (99.08)	101 (93.52)	7 (6.48)
Spain	2002	2 (0.10)	2000 (99.90)	1820 (91.00)	180 (9.00)
Sweden	686	0 (0)	686 (100)	683 (99.56)	3 (0.44)
Switzerland	914	5 (0.55)	909 (99.45)	869 (95.60)	40 (4.40)
Turkey	1411	158 (11.20)	1253 (88.80)	815 (65.04)	438 (34.96)
Ukraine	165	0 (0)	165 (100)	158 (95.76)	7 (4.24)
United Kingdom	9887	43 (0.43)	9844 (99.57)	9444 (95.94)	400 (4.06)
Total	46832	507 (1.08)	46325 (98.92)	43055 (92.94)	3270 (7.06)

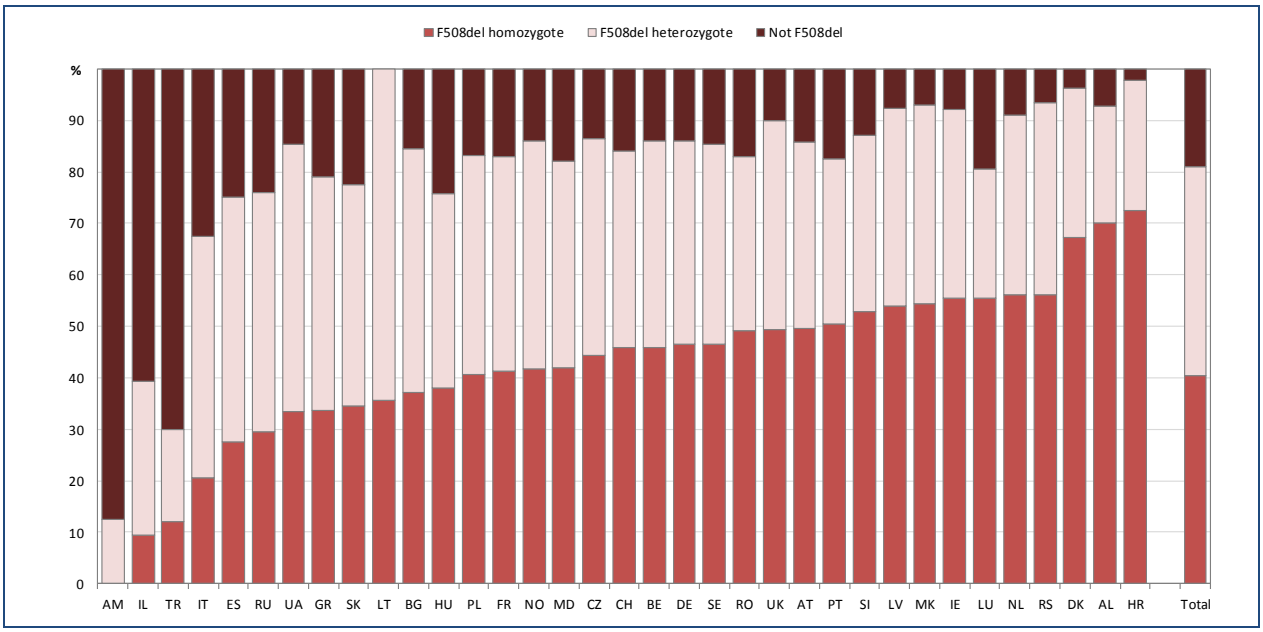
The table shows how many patients underwent DNA analysis to identify the CFTR mutations (column “genotyping done”) and, for those patients, how many patients had both mutations identified (column “two mutations identified”) and for how many one or both mutations remained unidentified (column “at least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

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



This graph shows the percentage of mutations that are not identified (unknown 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 4% of mutations remain unidentified after DNA analysis, leaving 7.06% 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 2017.



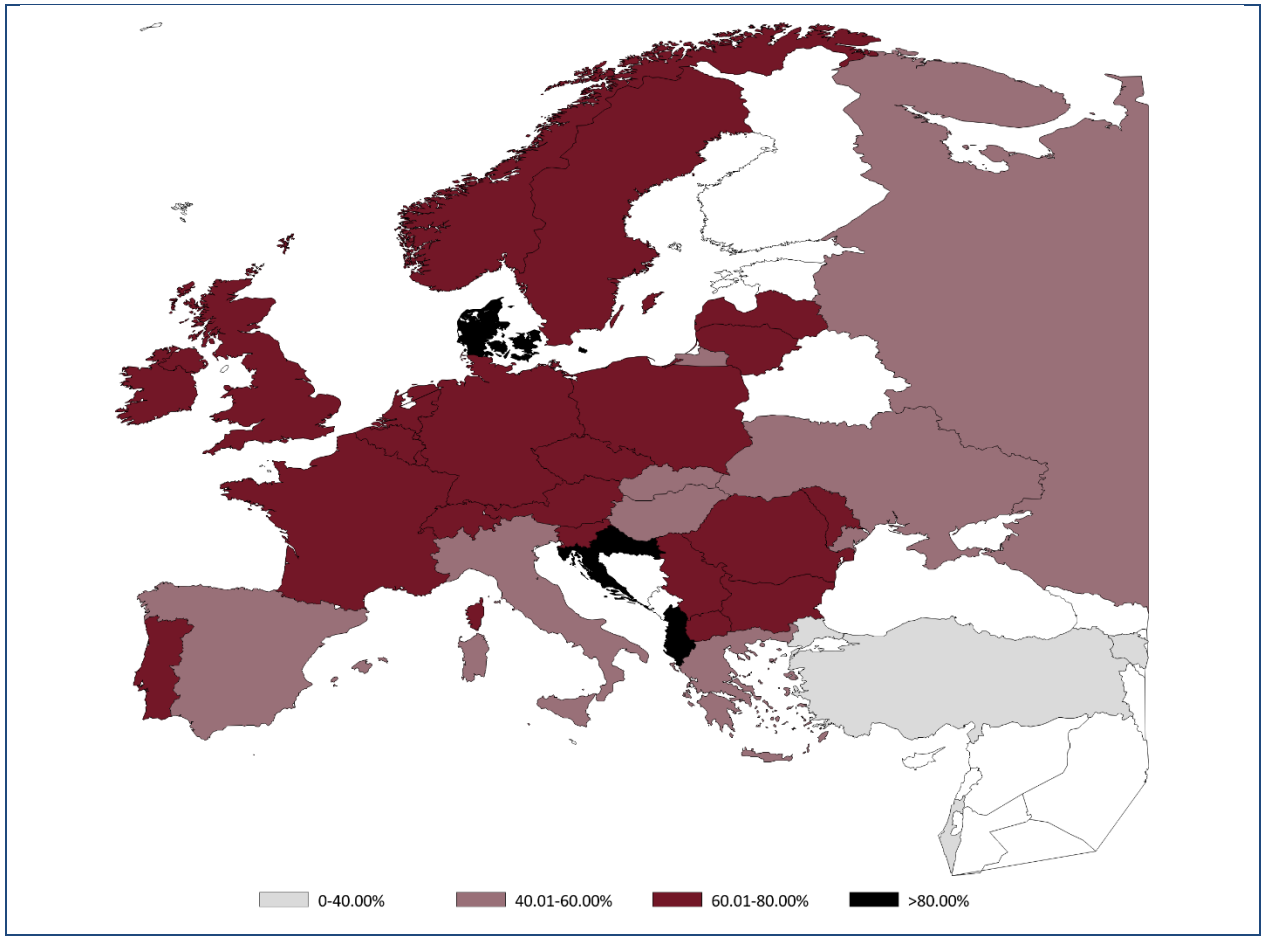
F508del is the name of the most commonly occurring CFTR mutation in the world. Patients who carry two F508del mutations are often described as having “classic CF”, but other combinations of mutations may cause the same degree of disease. We have grouped the patients in F508del homozygous (patients who have two F508del mutations), F508del heterozygous (patients who have one F508del mutation and another mutation, different from F508del), and patients without F508del mutations. Only patients for whom the genotype is known have been included in this graph. “Unknown” mutations have been classified as “other”, since F508del is included in all genotyping kits and would have been identified. Please note that the genotype grouping in this graph does not reflect the severity of the disease in the countries.

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

Mutation name	Number of alleles	Percentage among tested	Country with highest allele frequency
F508del	56285	60.75	Croatia (85.1%)
G542X	2495	2.69	Armenia (7.8%)
N1303K	2027	2.19	Italy (5.5%)
G551D	1226	1.32	Ireland (8.1%)
W1282X	1009	1.09	Israel (23.0%)
R117H	969	1.05	Ireland (3.0%)
2789+5G->A	936	1.01	Italy (2.9%)
3849+10kbC->T	863	0.93	Lithuania (14.3%)
CFTRdele2,3	838	0.9	Russia (6.2%)
1717-1G->A	816	0.88	Switzerland (3.1%)
R553X	778	0.84	Lithuania (3.6%)
621+1G->T	606	0.65	Greece (6.3%)
2183AA->G	583	0.63	Armenia (9.4%)
D1152H	562	0.61	Israel (5.6%)
R347P	496	0.54	Luxembourg (2.8%)
G85E	483	0.52	Israel (2.6%)
R1162X	483	0.52	Slovenia (5.1%)

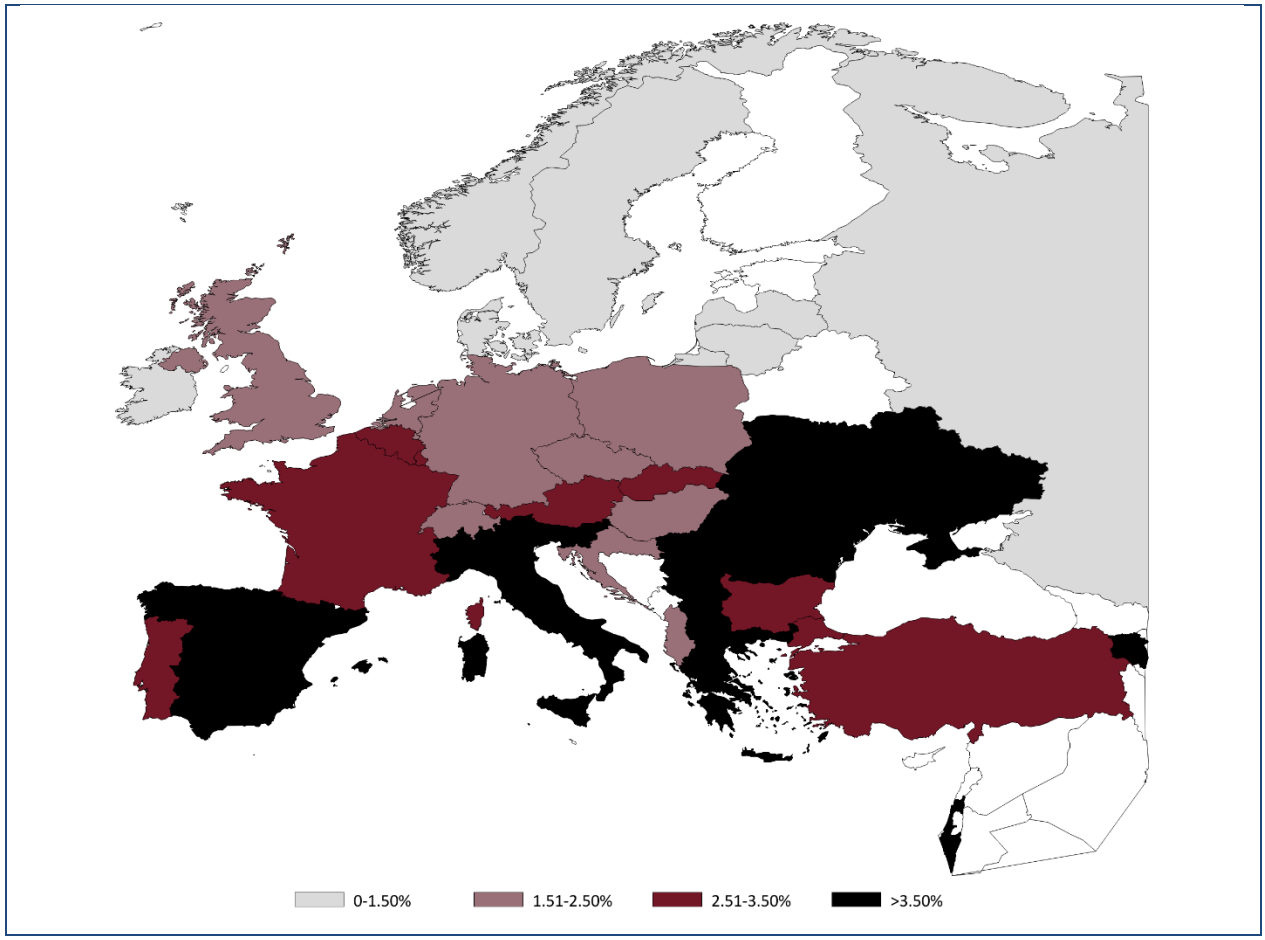
This table presents the allele frequency of the 17 most commonly occurring mutations found in the ECFSPR database. The last column reports in which country this particular mutation is most frequent. F508del is by far the most frequent mutation. Additionally, since F508del is included in all genetic screening tests, this is also the mutation with the highest detection rate.

Figure 3.3 Geographical distribution of mutation F508del.



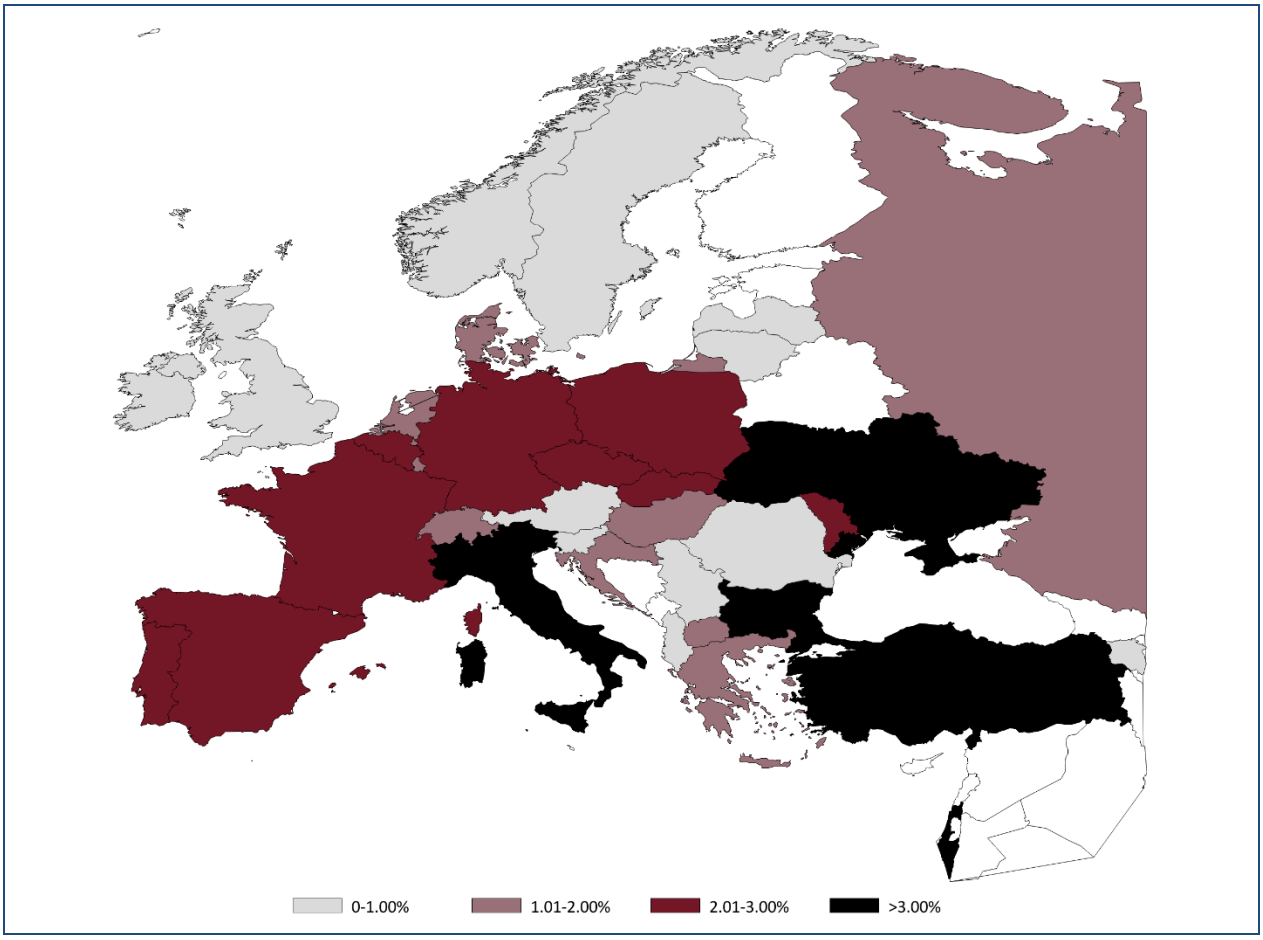
Although this mutation is the most common in all countries, it is most frequent in the South East of Europe, the highest frequency occurs in Croatia (85.1%) and Albania (81.4%), and in the north of Europe, in Denmark (81.7%).

Figure 3.4 Geographical distribution of mutation G542X.



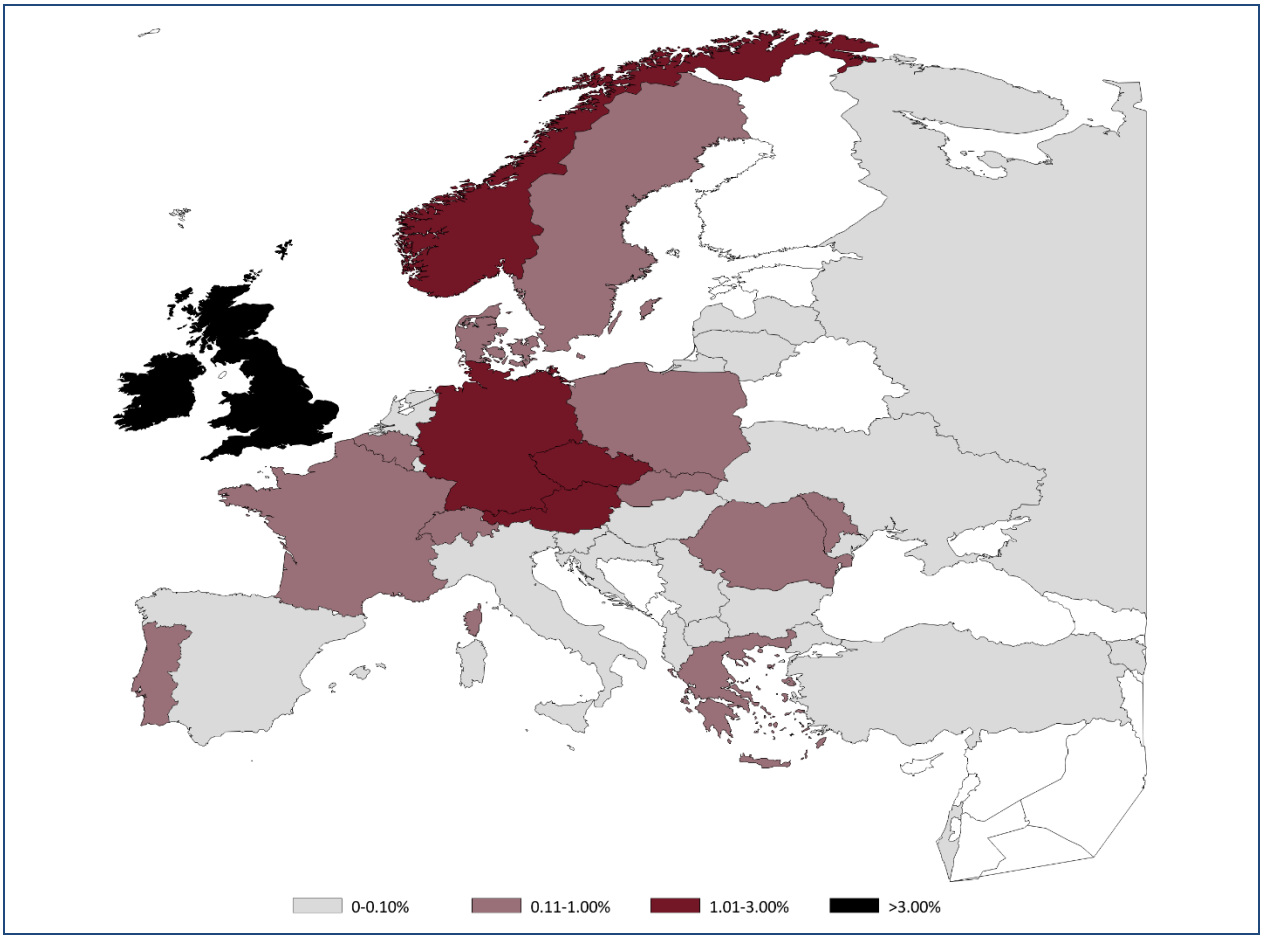
This mutation is most frequent in Southern Europe, with the highest allele frequency in Armenia (7.8%), whereas it is very rarely found in Ireland, the Scandinavian and Baltic countries or the Russian Federation.

Figure 3.5 Geographical distribution of mutation N1303K.



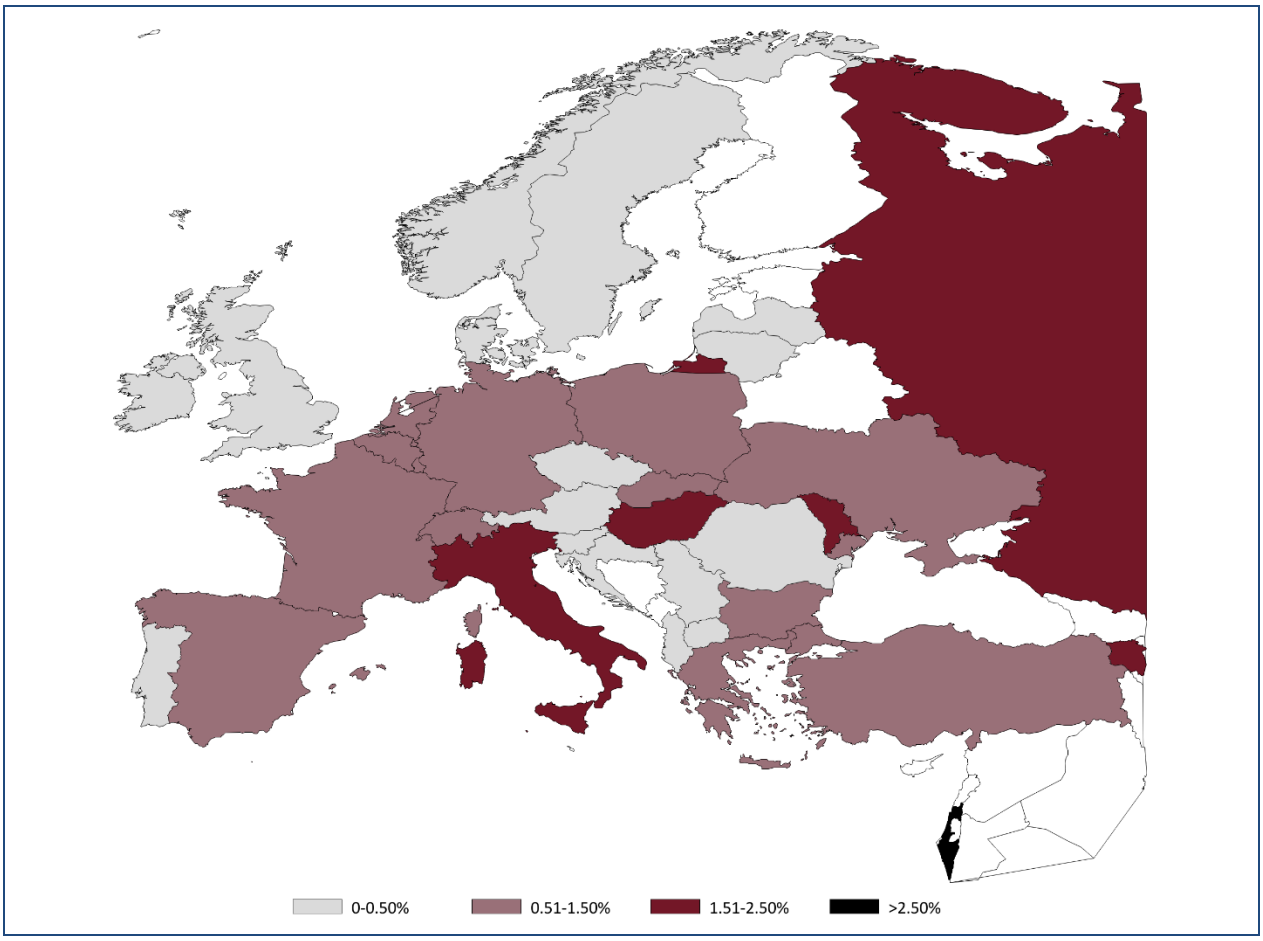
This mutation is most frequent in Italy (5.5%) and other countries in Southern Europe and in Eastern Europe, but rare in Northern Europe.

Figure 3.6 Geographical distribution of mutation G551D.



This mutation is most frequent in Ireland (8.1%) and United Kingdom (3.0%), whereas it is rare in Southern Europe.

Figure 3.7 Geographical distribution of mutation W1282X.



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

4. Lung function

FEV_1 is measured in litres but it is normally expressed as a percentage of the expected value (FEV_1%). The expected 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 full reference we refer you to Appendix 1, page 144). This is the global reference for spirometry and it has been agreed, as part of the CF global harmonisation project, that this is the best way to present lung function.

A FEV_1% of 100 means that the lung function measurement is equal to the mean lung function measurement of people of the same age, sex and height of the healthy reference population.

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

We asked the countries to report the best FEV_1 recorded throughout the year (according to the FEV_1% computed at the CF centres) to the ECFSPR. Whereas in the past some national registries recorded a different value, we are pleased to announce that in this report all countries report the best FEV1.

We excluded patients from the analyses on FEV_1 who have had one or more lung transplants, since their lung function does not reflect the severity of their CF lung disease.

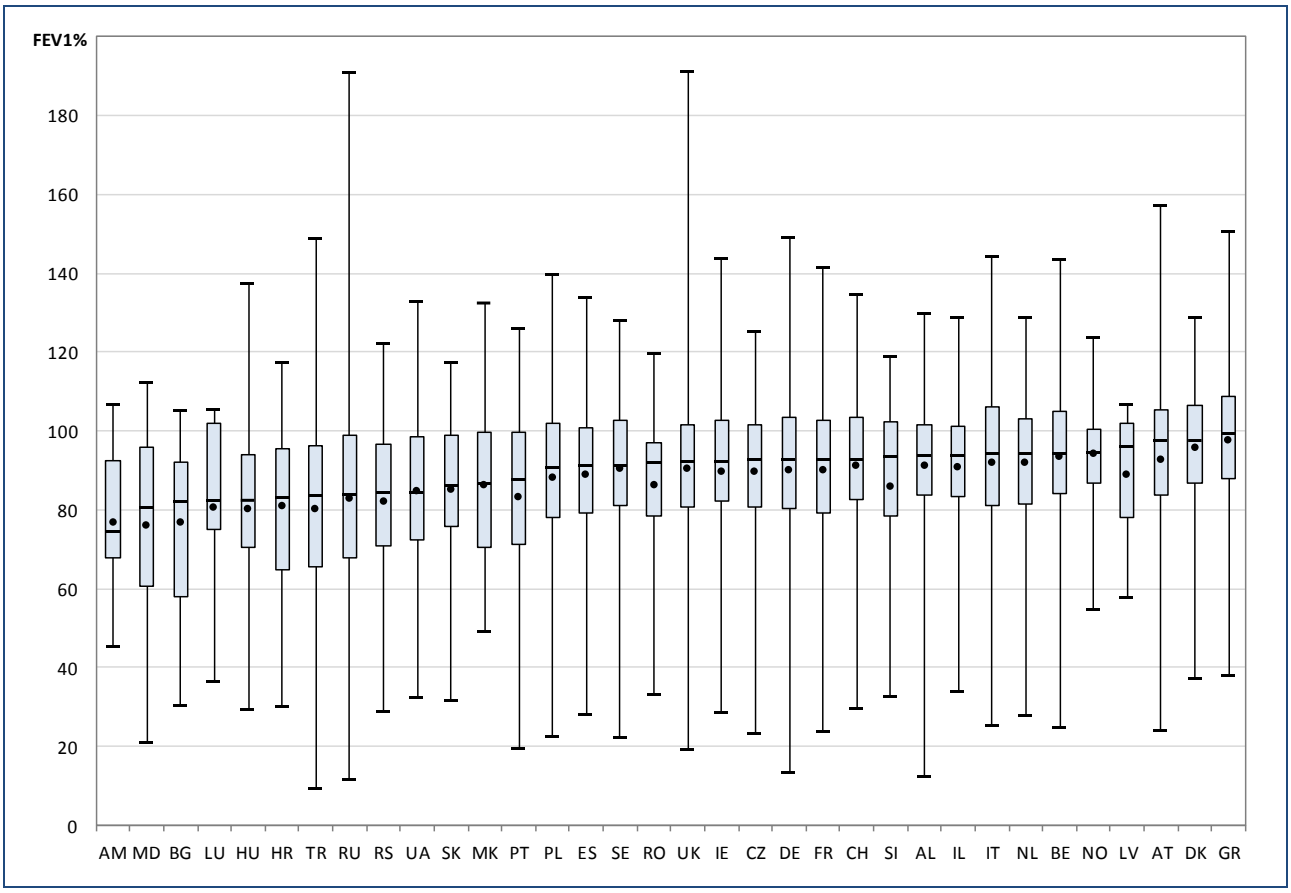
Table 4.1 FEV_1% of predicted: descriptive statistics, by country. Patients aged 6-17 years who have never had a lung transplant.

Country	N	N Miss	Mean (average FEV_1%)	Min	25th pctl (25% of patients have FEV_1% below this value)	Median (50% of patients have FEV_1% below this value)	75th pctl (75% of patients have FEV_1% below this value)	Max
Albania	55	0	91.0	12.3	83.6	93.6	101.6	129.7
Armenia	16	0	76.9	45.2	67.8	74.3	92.6	106.6
Austria	243	3	92.8	24.0	83.9	97.4	105.5	157.1
Belgium	346	10	93.5	24.8	84.1	94.3	104.9	143.4
Bulgaria	41	1	76.7	30.7	57.9	81.9	92.0	105.3
Croatia	34	2	80.8	30.3	64.7	83.0	95.5	117.4
Czech Republic	211	12	89.5	23.4	80.6	92.5	101.5	125.1
Denmark	129	0	95.5	37.2	86.8	97.7	106.6	128.6
France	2088	135	90.0	23.7	79.3	92.5	102.6	141.7
Germany	1746	32	90.1	13.3	80.5	92.5	103.3	149.0
Greece	219	9	97.5	38.0	88.1	99.5	108.6	150.5
Hungary	164	24	80.1	29.3	70.5	82.6	94.1	137.5
Ireland	394	7	89.8	28.7	82.1	92.4	102.8	143.9
Israel	178	0	90.7	33.9	83.4	93.6	101.3	128.6
Italy	1298	341	92.0	25.1	81.0	94.1	106.1	144.1
Latvia	19	1	88.7	57.8	78.0	96.0	101.8	106.8
Luxembourg	<10	0	80.7	36.6	75.0	82.5	101.8	105.6
Rep of Moldova	28	2	76.1	20.9	60.6	80.5	95.9	112.5
The Netherlands	396	16	91.9	27.9	81.6	94.2	103.1	128.5
North Macedonia	50	2	86.3	49.0	70.5	86.4	99.6	132.4
Norway	58	0	94.0	54.8	86.7	94.6	100.4	123.9
Poland	303	19	88.1	22.5	78.1	90.6	102.1	139.7
Portugal	121	7	83.0	19.5	71.3	87.9	99.7	125.8
Romania	70	24	86.2	33.1	78.6	91.8	96.9	119.7
Russian Federation	956	440	82.9	11.5	67.7	83.9	98.8	190.6
Serbia	76	5	82.1	29.0	71.0	84.4	96.7	122.3
Slovak Republic	85	3	84.9	31.5	75.6	86.1	98.9	117.3
Slovenia	44	0	86.0	32.7	78.5	93.5	102.4	118.9
Spain	699	15	88.9	28.4	79.3	91.2	100.7	133.8
Sweden	187	13	90.5	22.1	81.1	91.3	102.6	127.9
Switzerland	275	10	91.1	29.9	82.7	92.6	103.5	134.7
Turkey	478	159	80.2	9.2	65.6	83.4	96.1	148.6
Ukraine	83	8	84.7	32.5	72.4	84.4	98.7	132.9
United Kingdom	2618	277	90.2	19.1	80.8	92.3	101.4	191.0

Note: Lithuania has 0% coverage for children.
The UK reports best FEV1 from the annual review which is the time period between data sets and is not a calendar year. Therefore, in some cases month and day of FEV1 could be dated in 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 lung 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 lung transplant.



Note: Lithuania has 0% coverage for children.

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.

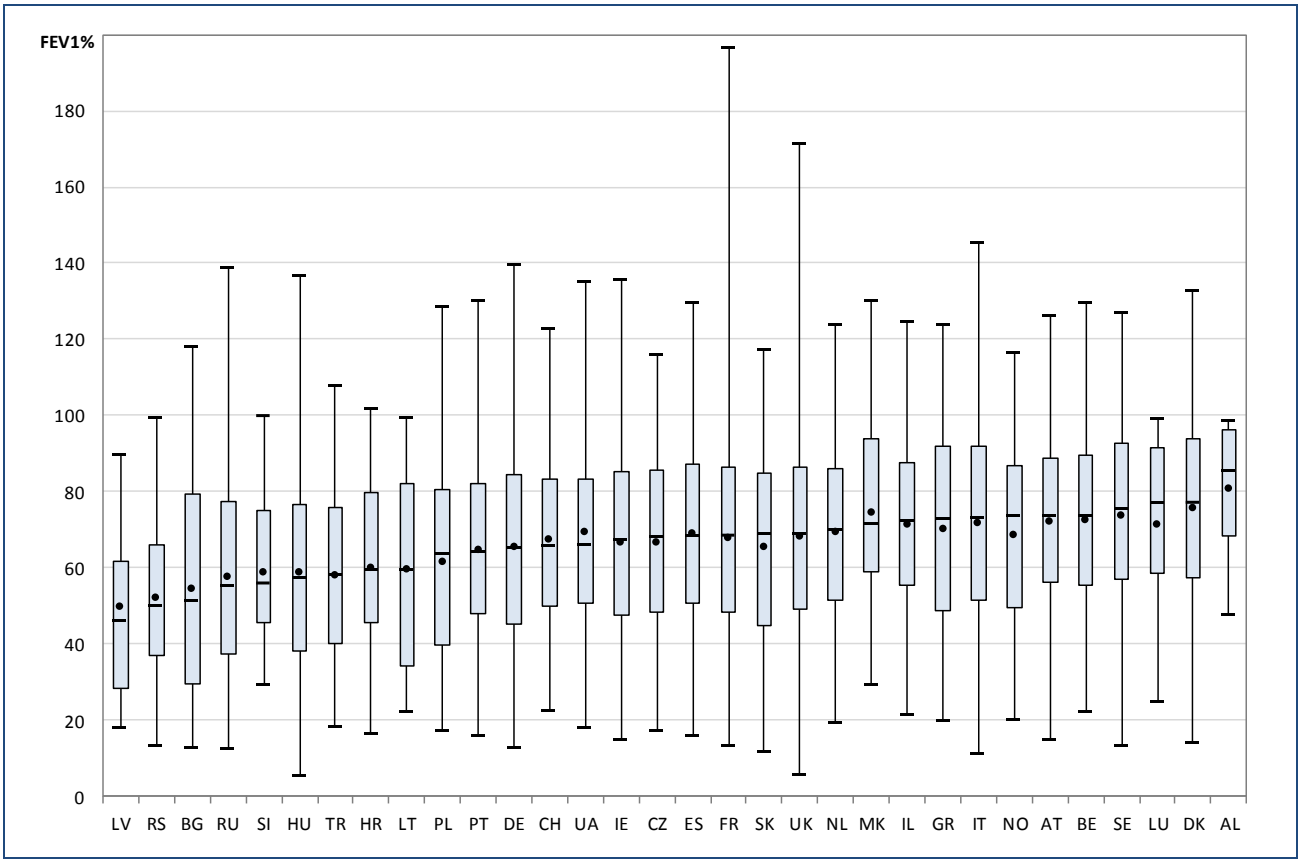
Table 4.2 FEV_1% of predicted: descriptive statistics, by country. Patients aged 18 years or older who have never had a lung transplant.

Country	N	N Miss	Mean (average FEV_1%)	Min	25th pctl (25% of patients have FEV_1% below this value)	Median (50% of patients have FEV_1% below this value)	75th pctl (75% of patients have FEV_1% below this value)	Max
Albania	8	0	80.9	48.0	68.3	85.6	96.3	98.8
Austria	309	4	72.1	14.7	56.4	73.8	88.8	126.2
Belgium	602	9	72.6	22.5	55.3	73.9	89.6	129.7
Bulgaria	59	2	54.6	13.1	29.4	51.2	79.3	117.9
Croatia	25	6	60.1	16.4	45.5	59.7	79.8	102.0
Czech Republic	223	12	66.7	17.3	48.6	68.2	85.6	116.2
Denmark	242	2	75.7	13.9	57.4	77.3	93.8	133
France	2874	67	67.8	13.3	48.6	68.6	86.5	196.5
Germany	2861	76	65.4	12.8	45.4	65.6	84.4	139.5
Greece	262	2	70.3	19.8	48.9	73.1	91.9	124.0
Hungary	157	28	58.7	5.3	38.4	57.7	76.6	137.0
Ireland	554	14	66.6	14.7	47.5	67.6	85.2	135.7
Israel	307	1	71.4	21.5	55.4	72.4	87.8	124.8
Italy	2461	371	71.8	11.3	51.4	73.5	91.9	145.6
Latvia	10	0	49.9	17.9	28.5	46.2	61.7	89.9
Lithuania	14	0	59.5	22.5	34.2	59.7	82.1	99.4
Luxembourg	21	0	71.4	25.2	58.7	77.1	91.7	99.3
The Netherlands	763	21	69.4	19.4	51.6	70.0	86.0	124.1
North Macedonia	28	2	74.4	29.5	59.0	71.6	94.1	130.1
Norway	134	1	68.8	20.5	49.4	73.7	86.9	116.3
Poland	104	2	61.4	17.1	39.6	63.9	80.4	128.8
Portugal	115	6	64.5	16.0	47.9	64.1	82.2	130.0
Russian Federation	451	163	57.5	12.5	37.4	55.3	77.4	138.7
Serbia	46	1	52.2	13.2	37.2	50.0	66.1	99.5
Slovak Republic	127	4	65.4	11.9	44.7	69.1	84.7	117.4
Slovenia	29	1	58.8	29.4	45.5	56.2	75.0	99.9
Spain	727	17	69.0	15.9	50.9	68.5	87.3	129.8
Sweden	326	22	73.8	13.3	56.9	75.8	92.9	127.0
Switzerland	410	1	67.6	22.9	49.9	65.9	83.5	122.6
Turkey	70	24	58.0	18.4	40.3	58.5	75.8	107.9
Ukraine	17	5	69.5	18.1	50.6	66.1	83.2	135.3
United Kingdom	5027	204	68.1	6.0	49.2	69.1	86.4	171.3

Note: Armenia, Rep of Moldova and Romania have <5 patients aged 18 years or more at FEV_1 measurement and are excluded from this table.

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

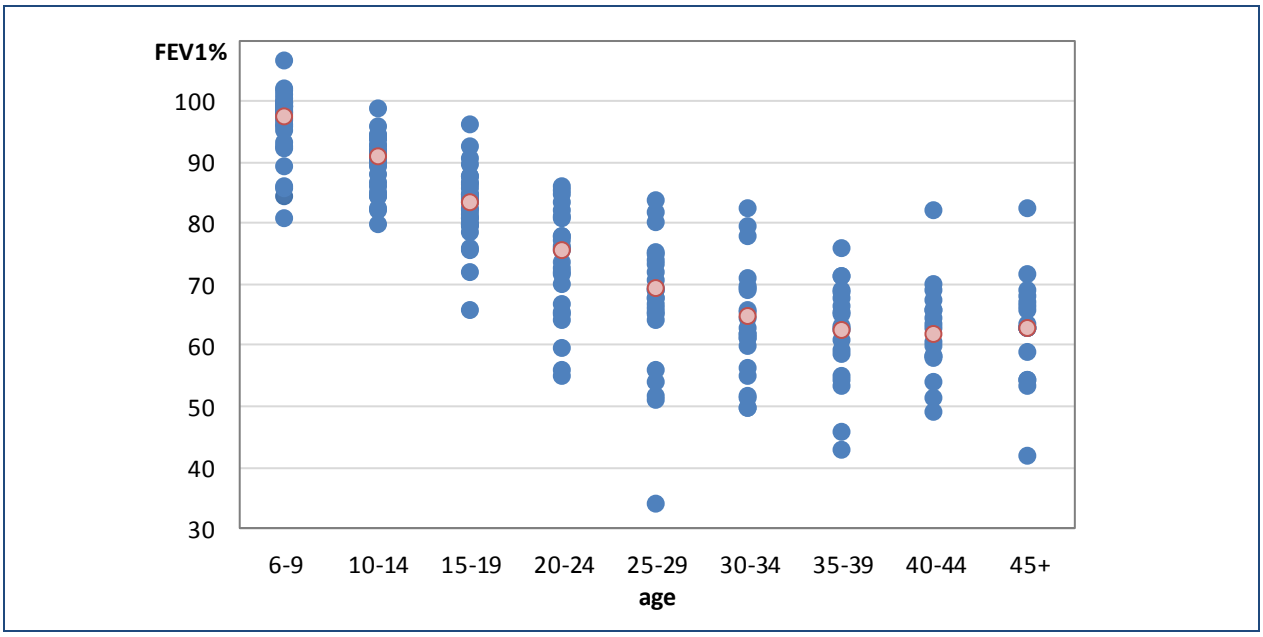
Figure 4.2 FEV_1% of predicted: box-plot, by country and overall. Patients aged 18 years or older who have never had a lung transplant.



Note: Armenia, Rep of Moldova and Romania have <5 patients aged 18 years or more at FEV_1 measurement and are excluded from this graph.

This box-plot is a graphic representation of the FEV_1 in adults, expressed as % of predicted detailed in table 4.2. For each country the dash (black line crossing the blue box) is the median, the black dot is the mean and the whiskers (vertical lines with a T-shaped end) are the minimum and the maximum.

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



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

This graph shows the median FEV_1% (the value that separates the highest and lowest half of the patients) by age group. Each country is represented by a dot (in blue) and the overall estimate is in red. The general pattern shows that the FEV_1% slowly decreases until the age of 30-34, 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.

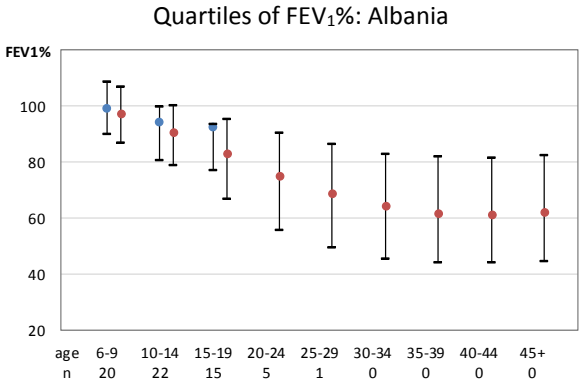
Table 4.3 FEV_1% of predicted: descriptive statistics by age group (patients aged 6 years or older) who have never had a lung transplant.

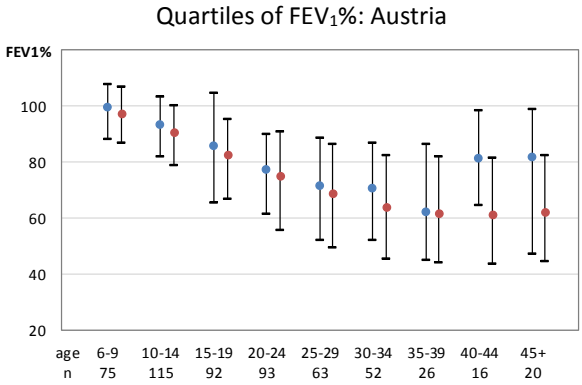
Age at FEV_1 measurement	N	N Miss	Mean	Min	25th pctl	Median	75th pctl	Max
6-9	4687	758	96.0	11.5	86.6	97.3	106.9	191.0
10-14	5686	549	88.2	9.2	78.7	90.7	100.3	157.1
15-19	5252	395	80.0	5.3	66.6	83.0	95.1	186.4
20-24	4570	256	72.7	11.7	55.6	75.1	90.5	138.7
25-29	3964	241	67.9	11.3	49.5	68.9	86.2	139.5
30-34	2966	167	64.5	6.0	45.3	64.3	82.7	196.5
35-39	2151	93	63.4	12.5	43.9	62.0	82.0	147.8
40-44	1436	62	63.2	15.5	43.8	61.5	81.3	138.6
45+	2378	124	64.0	14.1	44.3	62.4	82.2	171.3

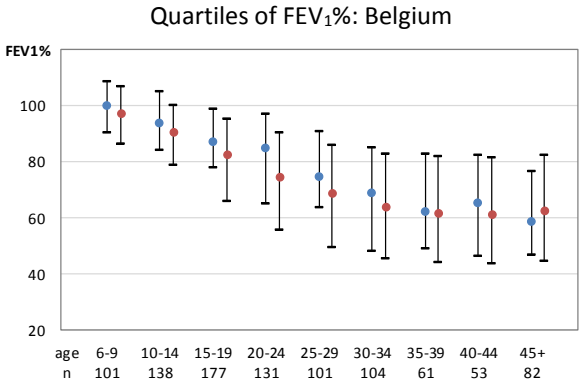
This table shows FEV_1% by age group for the total data set. The median values reported in this table are shown as red dots in fig 4.3.

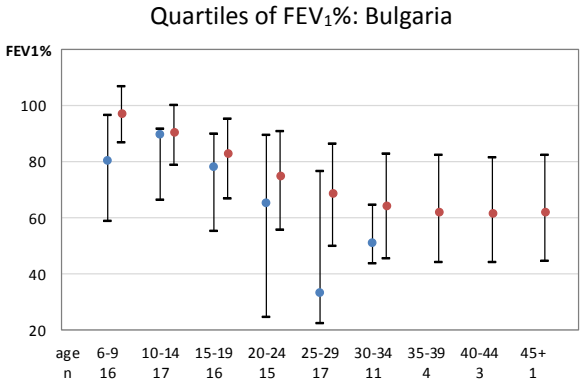
Figure 4.4 Quartiles of FEV$_1$% of predicted by age group and by country. Patients aged 6 years or older and who have never had a lung transplant.

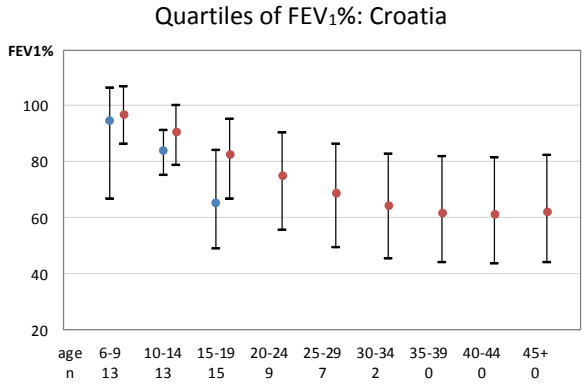
The figures below show the FEV$_1$% in different age groups, separately for each country. The dot shows the median, and the whiskers show the 25th and 75th percentiles (the median, the 25th percentile and the 75th percentile are collectively named "quartiles"). In blue are the quartiles for the country,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

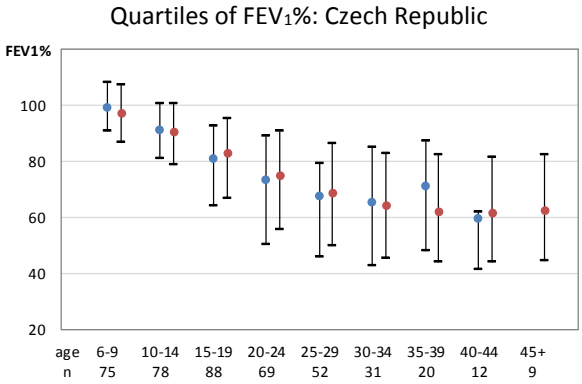




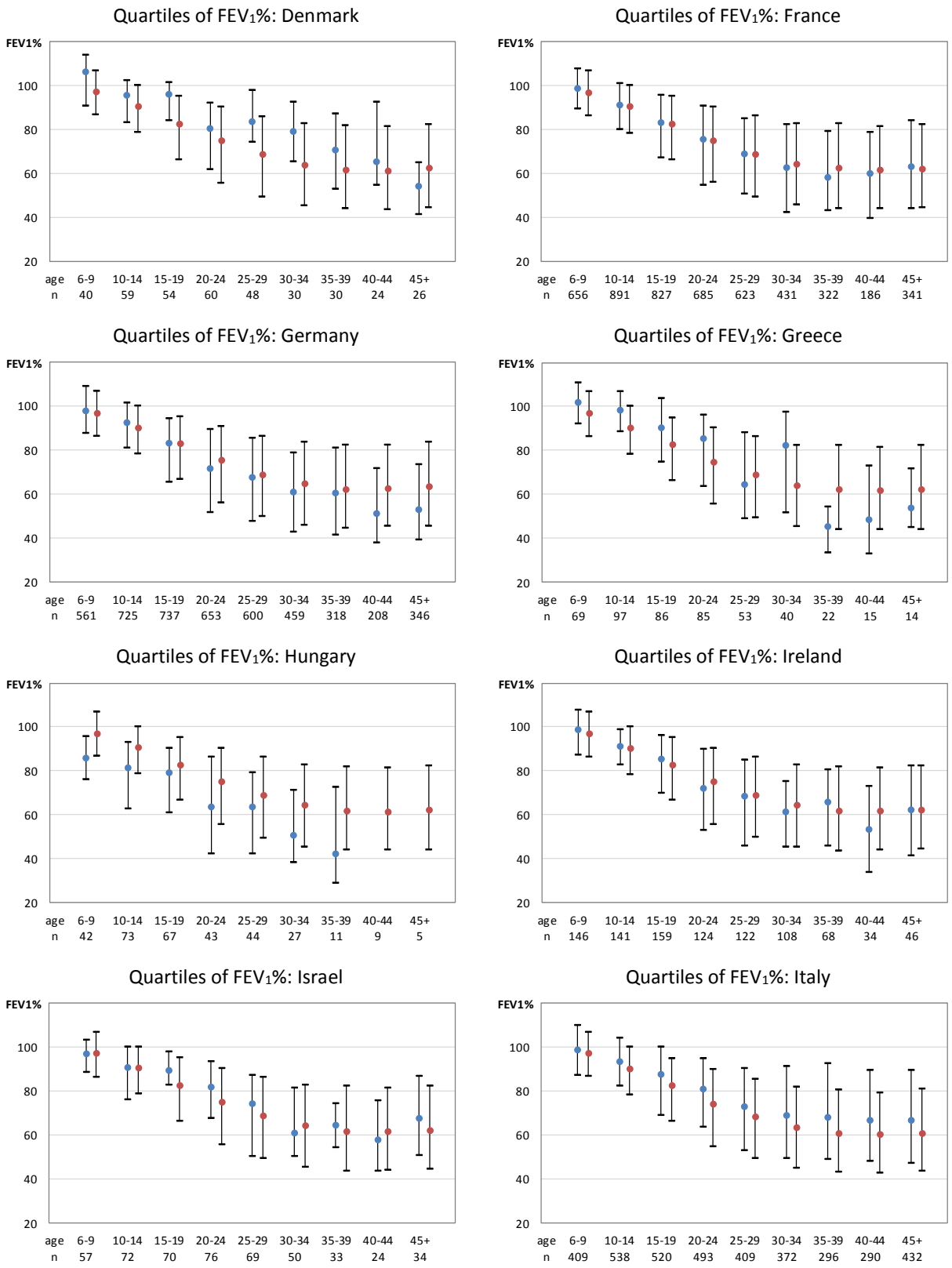






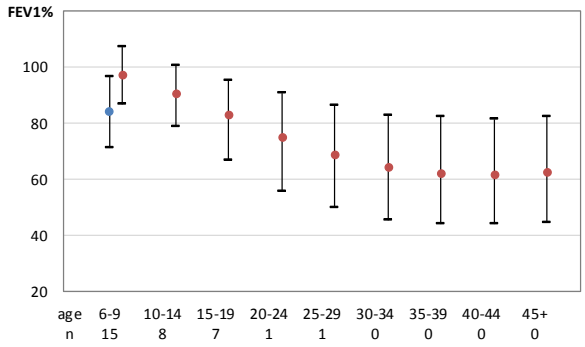


[figure 4.4 continued]

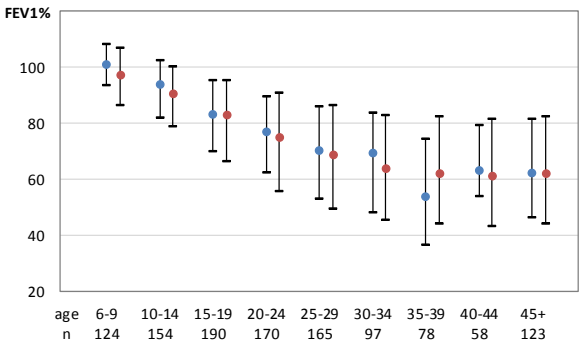


[figure 4.4 continued]

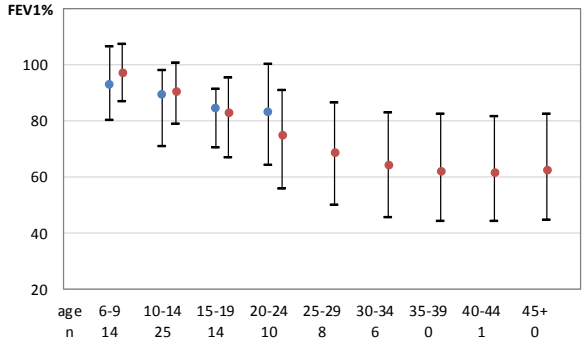
Quartiles of FEV$_1$%: Rep of Moldova



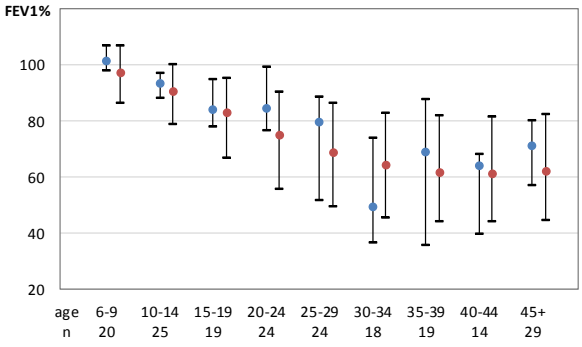
Quartiles of FEV$_1$%: The Netherlands



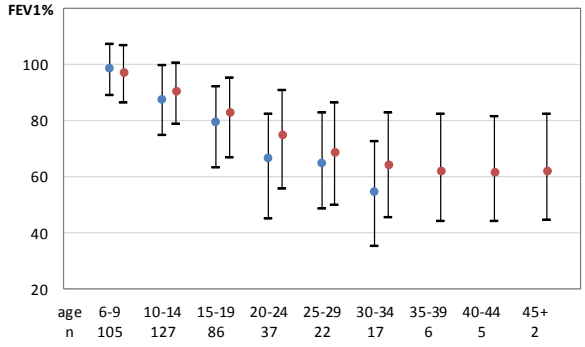
Quartiles of FEV$_1$%: North Macedonia



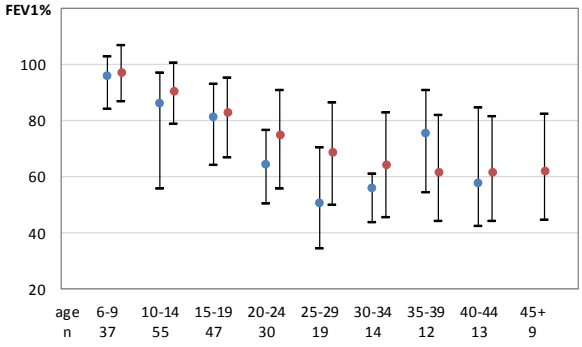
Quartiles of FEV$_1$%: Norway



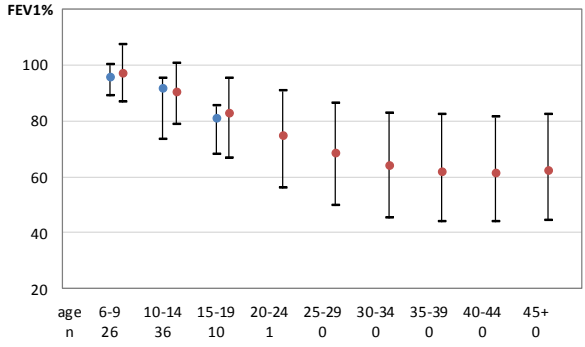
Quartiles of FEV$_1$%: Poland



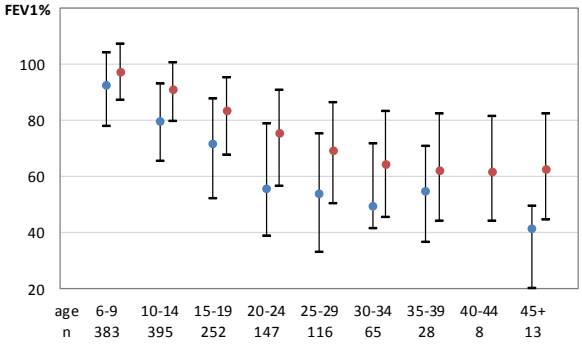
Quartiles of FEV$_1$%: Portugal



Quartiles of FEV$_1$%: Romania

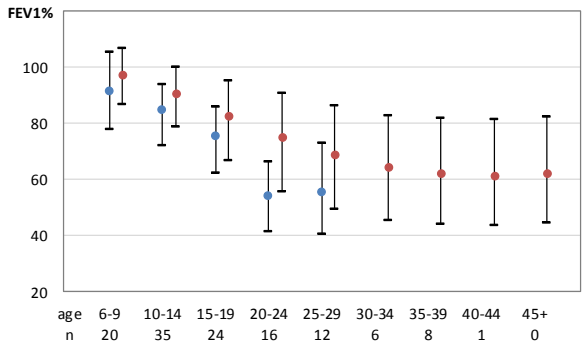


Quartiles of FEV$_1$%: Russian Federation

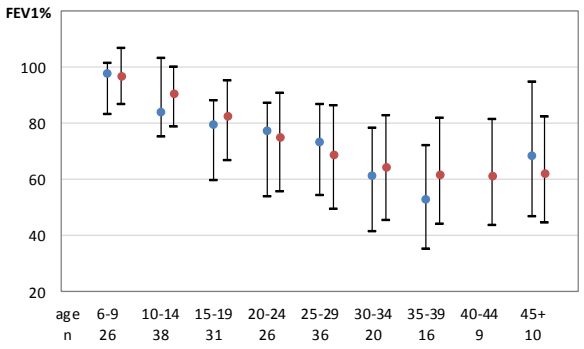


[figure 4.4 continued]

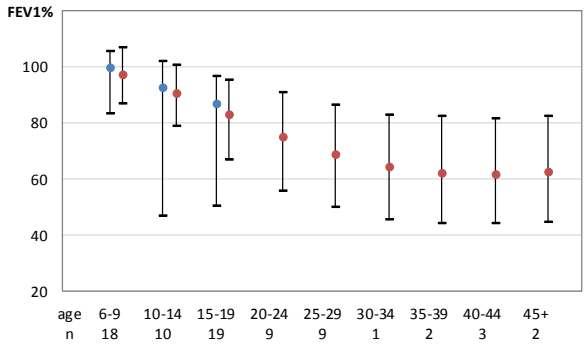
Quartiles of FEV$_1$%: Serbia



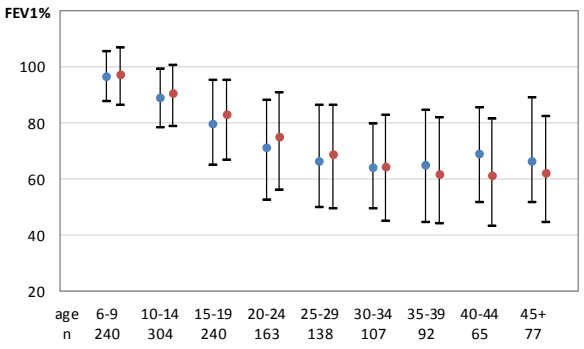
Quartiles of FEV$_1$%: Slovak Republic



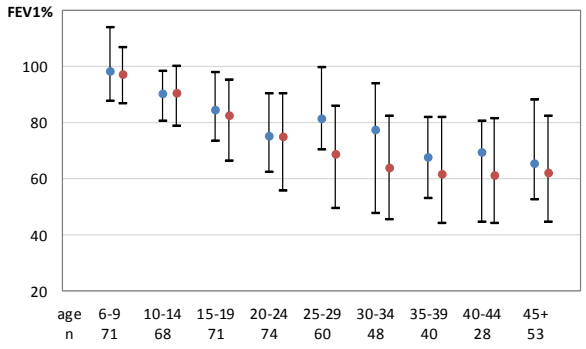
Quartiles of FEV$_1$%: Slovenia



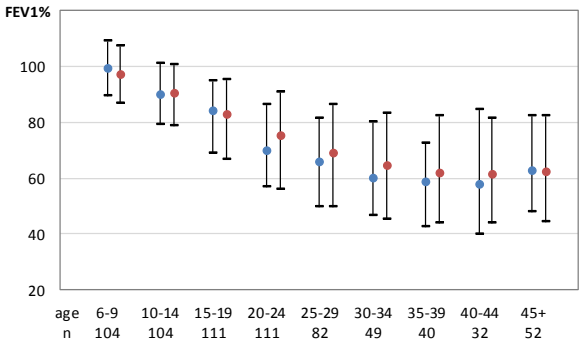
Quartiles of FEV$_1$%: Spain



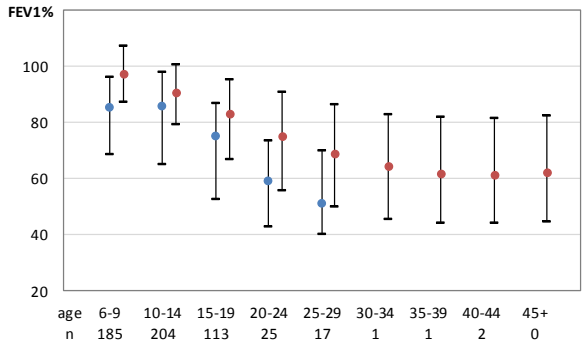
Quartiles of FEV$_1$%: Sweden



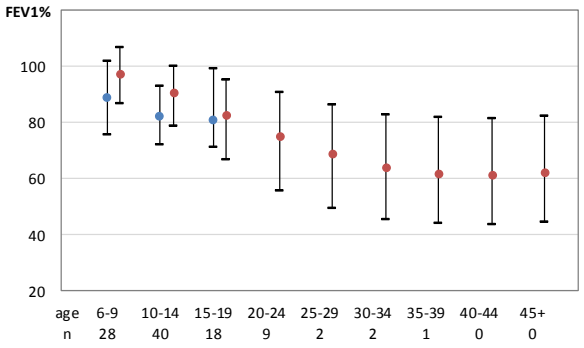
Quartiles of FEV$_1$%: Switzerland



Quartiles of FEV$_1$%: Turkey



Quartiles of FEV$_1$%: Ukraine



[figure 4.4 continued]

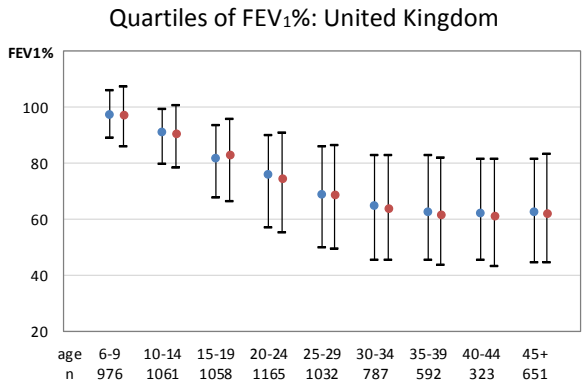
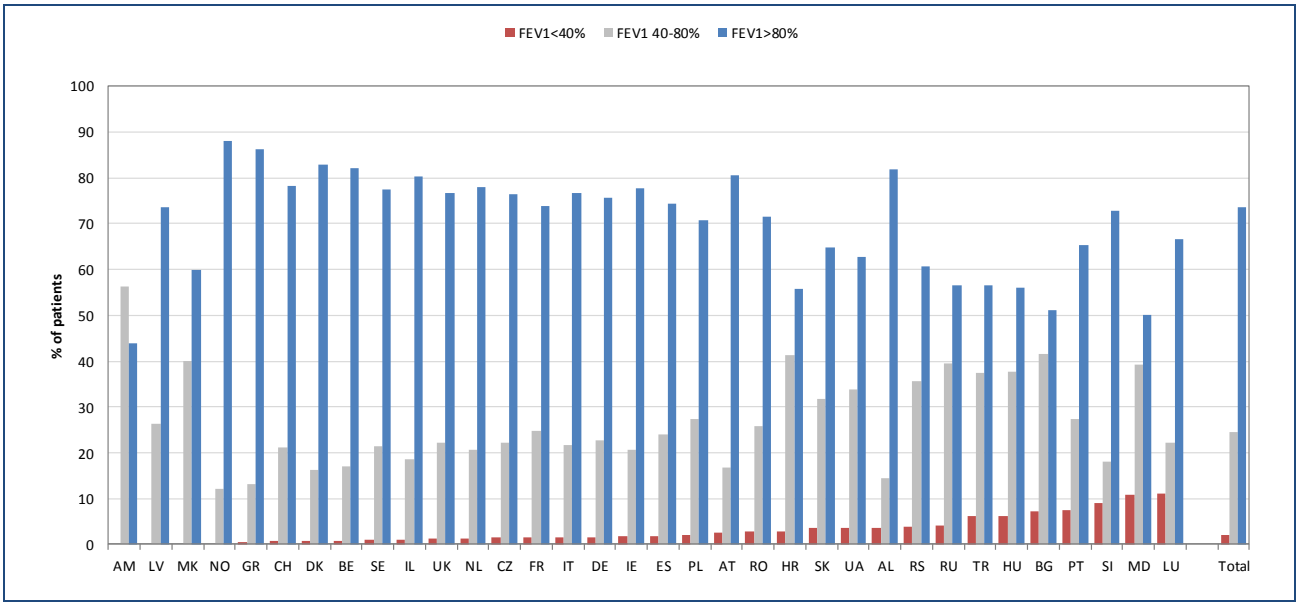


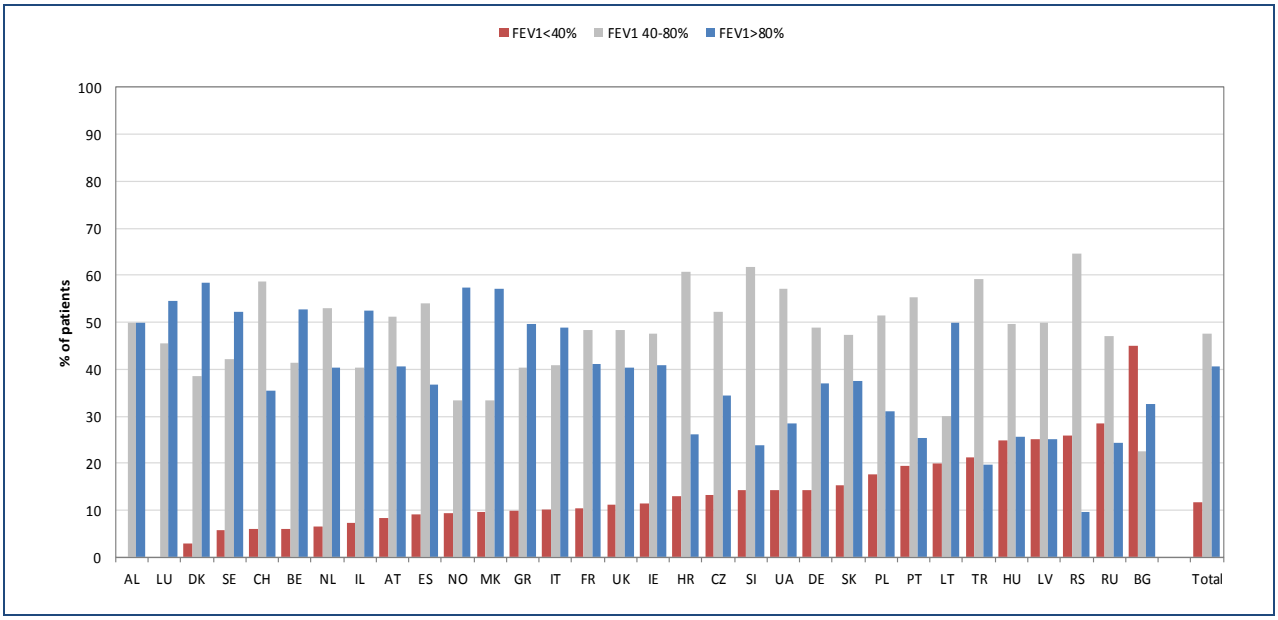
Figure 4.5 FEV_1% of predicted according to severity group and age group, by country and overall. Patients aged 6-17 years who have never had a lung transplant.



Note: Lithuania has 0% coverage for children.

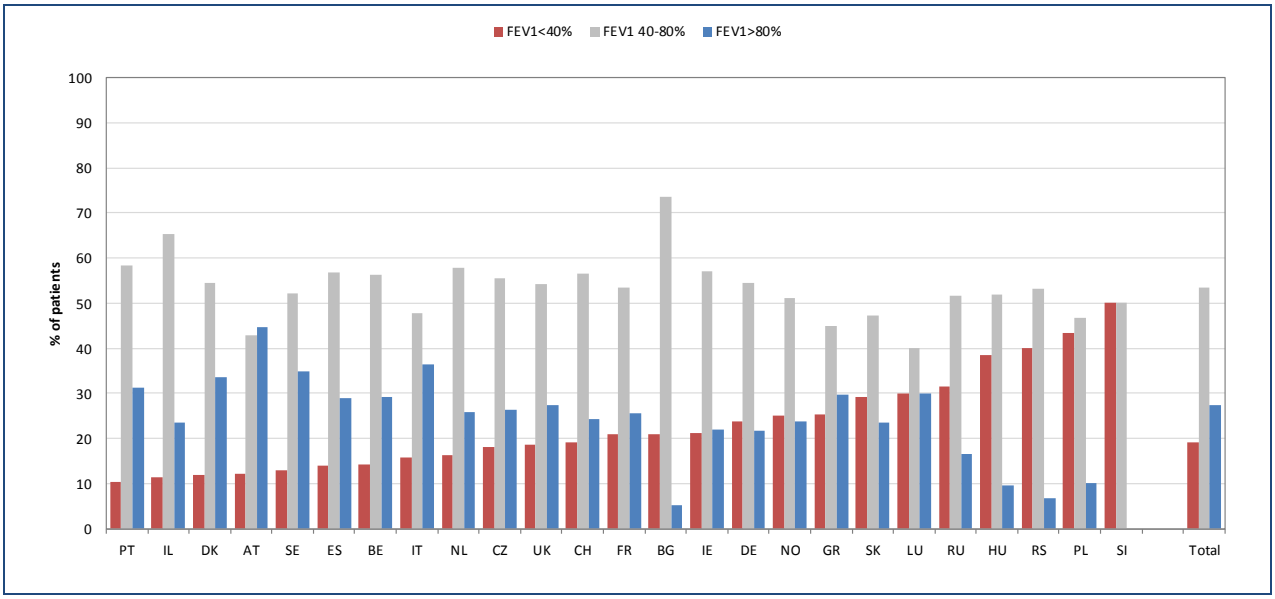
Figures 4.5, 4.6 and 4.7 show the FEV_1% by severity group, by country and overall. Patients with an FEV_1% higher than 80% are generally considered to have mild lung disease, patients with FEV_1% between 80% and 40% moderate lung disease, and patients with FEV_1 <40% severe lung disease. However, since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

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



Note: Armenia, Rep of Moldova and Romania have <5 patients aged 18-29 years at FEV_1 measurement and are excluded from this graph.

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



Note: Albania, Armenia, Croatia, Latvia, Lithuania, North Macedonia, Rep of Moldova, Romania, Turkey and Ukraine have <5 patients aged 30 years or older and are excluded from this graph.

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) and *Stenotrophomonas maltophilia*.

In the microbiology category discrepancies exist between the ECFSPR definitions and those of the national registries. The ECFSPR definition of chronic infection (see Appendix 2, page 144) 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:

a. modified Leeds criteria, chronic infection: >50% of respiratory samples collected during the last 12 months are positive. At least 4 samples during that period;
b. and/or significantly raised bacteria-specific antibodies according to local laboratories.

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 chronic bacterial infection in all patients seen in 2017, by country.

Country	Chronic *Pseudomonas aeruginosa* number (%)			Chronic *Burkholderia cepacia complex species* number (%)			Chronic *Staphylococcus aureus* number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	4 (3.28)	95 (77.87)	23 (18.85)	3 (2.46)	119 (97.54)	0 (0)	3 (2.46)	84 (68.85)	35 (28.69)
Armenia	4 (12.50)	16 (50.00)	12 (37.50)	30 (93.75)	2 (6.25)	0 (0)	3 (9.38)	2 (6.25)	27 (84.38)
Austria	5 (0.66)	536 (70.81)	216 (28.53)	2 (0.26)	729 (96.30)	26 (3.43)	4 (0.53)	336 (44.39)	417 (55.09)
Belgium[1]	216 (16.78)	788 (61.23)	283 (21.99)	216 (16.78)	1046 (81.27)	25 (1.94)	1287 (100)	-	-
Bulgaria	4 (2.70)	52 (35.14)	92 (62.16)	4 (2.70)	142 (95.95)	2 (1.35)	4 (2.70)	112 (75.68)	32 (21.62)
Croatia	4 (4.60)	43 (49.43)	40 (45.98)	5 (5.75)	81 (93.10)	1 (1.15)	4 (4.60)	37 (42.53)	46 (52.87)
Czech Republic	31 (5.12)	454 (75.04)	120 (19.83)	31 (5.12)	526 (86.94)	48 (7.93)	29 (4.79)	313 (51.74)	263 (43.47)
Denmark	0 (0)	339 (68.35)	157 (31.65)	0 (0)	463 (93.35)	33 (6.65)	496 (100)	-	-
France	0 (0)	5517 (79.50)	1423 (20.50)	0 (0)	6853 (98.75)	87 (1.25)	0 (0)	4418 (63.66)	2522 (36.34)
Germany	278 (4.54)	3736 (61.06)	2105 (34.40)	273 (4.46)	5719 (93.46)	127 (2.08)	277 (4.53)	3457 (56.50)	2385 (38.98)
Greece	43 (7.18)	285 (47.58)	271 (45.24)	22 (3.67)	575 (95.99)	2 (0.33)	25 (4.17)	408 (68.11)	166 (27.71)
Hungary	6 (1.19)	300 (59.52)	198 (39.29)	6 (1.19)	486 (96.43)	12 (2.38)	6 (1.19)	265 (52.58)	233 (46.23)
Ireland[2]	5 (0.41)	892 (73.17)	322 (26.42)	5 (0.41)	1185 (97.21)	29 (2.38)	5 (0.41)	761 (62.43)	453 (37.16)
Israel	24 (4.39)	286 (52.29)	237 (43.33)	26 (4.75)	513 (93.78)	8 (1.46)	25 (4.57)	316 (57.77)	206 (37.66)
Italy[3]	209 (3.76)	3349 (60.22)	2003 (36.02)	209 (3.76)	5227 (93.99)	125 (2.25)	210 (3.78)	2378 (42.76)	2973 (53.46)
Latvia	2 (5.13)	29 (74.36)	8 (20.51)	1 (2.56)	37 (94.87)	1 (2.56)	1 (2.56)	19 (48.72)	19 (48.72)
Lithuania	0 (0)	12 (85.71)	2 (14.29)	0 (0)	13 (92.86)	1 (7.14)	0 (0)	5 (35.71)	9 (64.29)
Luxembourg	0 (0)	27 (75.00)	9 (25.00)	0 (0)	34 (94.44)	2 (5.56)	0 (0)	15 (41.67)	21 (58.33)
Rep of Moldova	0 (0)	23 (46.00)	27 (54.00)	50 (100)	-	-	0 (0)	9 (18.00)	41 (82.00)
The Netherlands	132 (8.98)	871 (59.25)	467 (31.77)	132 (8.98)	1311 (89.18)	27 (1.84)	132 (8.98)	736 (50.07)	602 (40.95)
North Macedonia	2 (1.74)	71 (61.74)	42 (36.52)	2 (1.74)	113 (98.26)	0 (0)	2 (1.74)	82 (71.30)	31 (26.96)
Norway	30 (11.95)	160 (63.75)	61 (24.30)	31 (12.35)	212 (84.46)	8 (3.19)	35 (13.94)	92 (36.65)	124 (49.40)

[1] Belgium: *Chronic Pseudomonas aeruginosa* and *Chronic Burkholderia cepacia complex species* are not collected for transplanted patients and most of the missing data refers to this sub-population. *Chronic Staphylococcus* aureus is not collected.

[2] Ireland: chronicity for *Pseudomonas aeruginosa*, *Burkholderia cepacia complex species* and *Staphylococcus aureus* is defined as: at least 3 or more positive isolates during the last 12 months window preceding the last reported culture in 2017.

[3] Italy: chronicity for *Pseudomonas aeruginosa*, *Burkholderia cepacia complex species* and *Staphylococcus aureus* is defined as: at least 3 or more positive cultures during 2017.

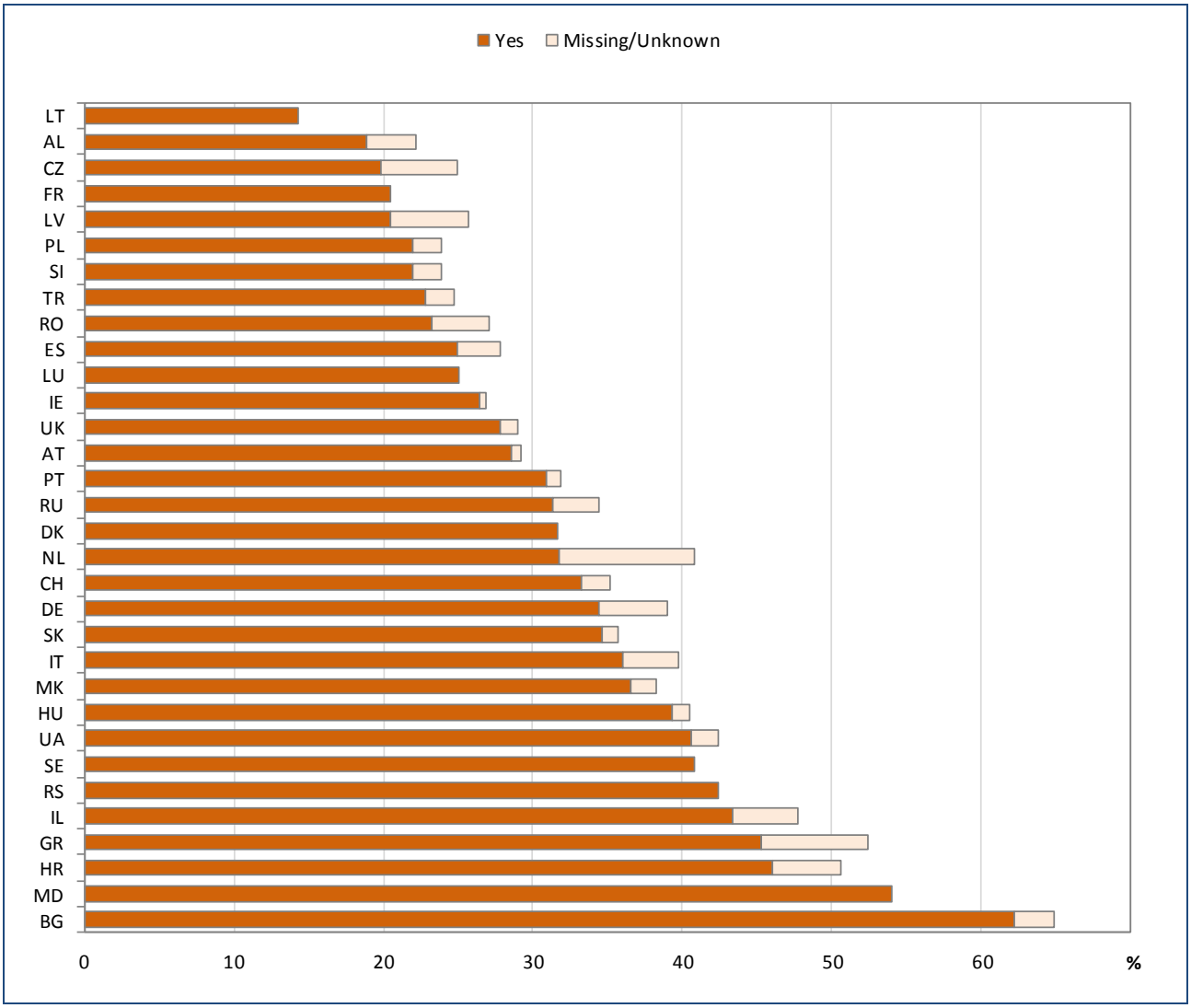
[table 5.1 continued]

Country	Chronic *Pseudomonas aeruginosa* number (%)			Chronic *Burkholderia cepacia complex species* number (%)			Chronic *Staphylococcus aureus* number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Poland	13 (1.98)	499 (76.07)	144 (21.95)	13 (1.98)	633 (96.49)	10 (1.52)	13 (1.98)	268 (40.85)	375 (57.16)
Portugal	3 (0.92)	223 (68.20)	101 (30.89)	4 (1.22)	303 (92.66)	20 (6.12)	4 (1.22)	165 (50.46)	158 (48.32)
Romania	6 (3.77)	116 (72.96)	37 (23.27)	7 (4.40)	152 (95.60)	0 (0)	6 (3.77)	128 (80.50)	25 (15.72)
Russian Federation	96 (3.12)	2020 (65.58)	964 (31.30)	72 (2.34)	2821 (91.59)	187 (6.07)	92 (2.99)	1284 (41.69)	1704 (55.32)
Serbia	0 (0)	99 (57.56)	73 (42.44)	0 (0)	151 (87.79)	21 (12.21)	0 (0)	47 (27.33)	125 (72.67)
Slovak Republic	3 (1.13)	171 (64.29)	92 (34.59)	3 (1.13)	250 (93.98)	13 (4.89)	4 (1.50)	126 (47.37)	136 (51.13)
Slovenia	2 (1.83)	83 (76.15)	24 (22.02)	4 (3.67)	104 (95.41)	1 (0.92)	3 (2.75)	45 (41.28)	61 (55.96)
Spain	57 (2.85)	1446 (72.23)	499 (24.93)	64 (3.20)	1850 (92.41)	88 (4.40)	59 (2.95)	1136 (56.74)	807 (40.31)
Sweden	0 (0)	406 (59.18)	280 (40.82)	0 (0)	669 (97.52)	17 (2.48)	79 (11.52)	423 (61.66)	184 (26.82)
Switzerland	17 (1.86)	593 (64.88)	304 (33.26)	18 (1.97)	871 (95.30)	25 (2.74)	20 (2.19)	399 (43.65)	495 (54.16)
Turkey	27 (1.91)	1062 (75.27)	322 (22.82)	36 (2.55)	1361 (96.46)	14 (0.99)	30 (2.13)	1022 (72.43)	359 (25.44)
Ukraine	3 (1.82)	95 (57.58)	67 (40.61)	2 (1.21)	156 (94.55)	7 (4.24)	2 (1.21)	83 (50.30)	80 (48.48)
United Kingdom[4]	115 (1.16)	7023 (71.03)	2749 (27.80)	115 (1.16)	9427 (95.35)	345 (3.49)	115 (1.16)	8247 (83.41)	1525 (15.42)

[4] United Kingdom: chronicity for *Pseudomonas aeruginosa* and *Staphylococcus aureus* 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.

Table 5.1 shows, separately by country, the frequency of chronic *Pseudomonas aeruginosa*, chronic *Burkholderia cepacia complex species* and chronic *Staphylococcus aureus*. 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.

Figure 5.1 Prevalence of chronic Pseudomonas aeruginosa infection in all patients seen in 2017, by country.

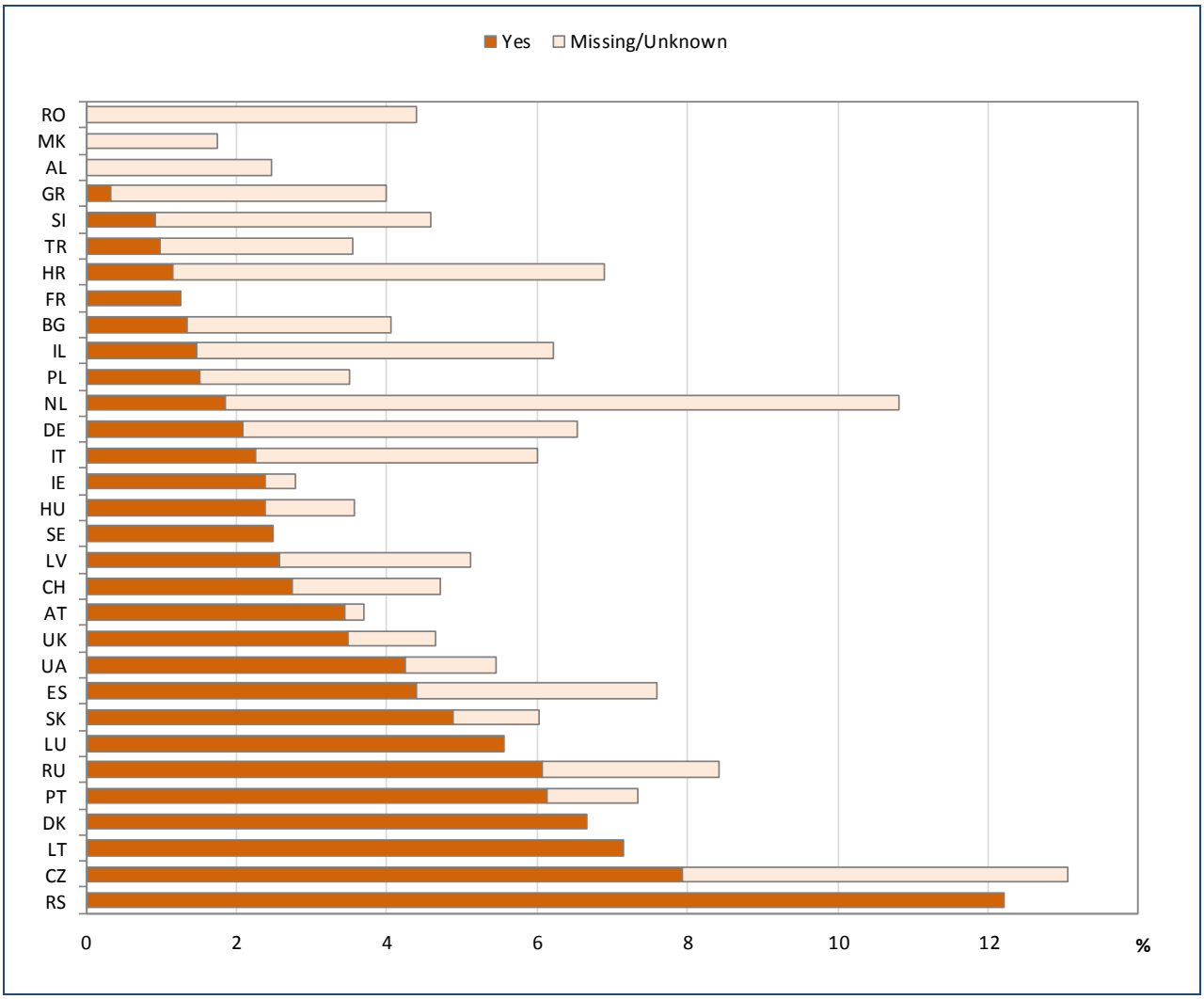


Note: We excluded from the graph the countries for which the information on *Pseudomonas aeruginosa* was missing for more than 10% of the patients.

Note: Ireland: chronicity for *Pseudomonas aeruginosa* is defined as: at least 3 or more positive isolates during the last 12 months window preceding the last reported culture in 2017.
Italy: chronicity for *Pseudomonas aeruginosa* is defined as: at least 3 or more positive cultures during 2017.
United Kingdom: for chronic *Pseudomonas aeruginosa* the definition is: 3 or more positive isolates during the last 12 months.

The horizontal bars represent the percentage of patients with chronic *Pseudomonas aeruginosa* infection (in dark orange) and the percentage of patients where information on *Pseudomonas aeruginosa* infection was missing (in light orange). This is a frequent infection, but prevalence varies considerably between countries.

Figure 5.2 Prevalence of chronic Burkholderia cepacia complex species infection in all patients seen in 2017, by country.

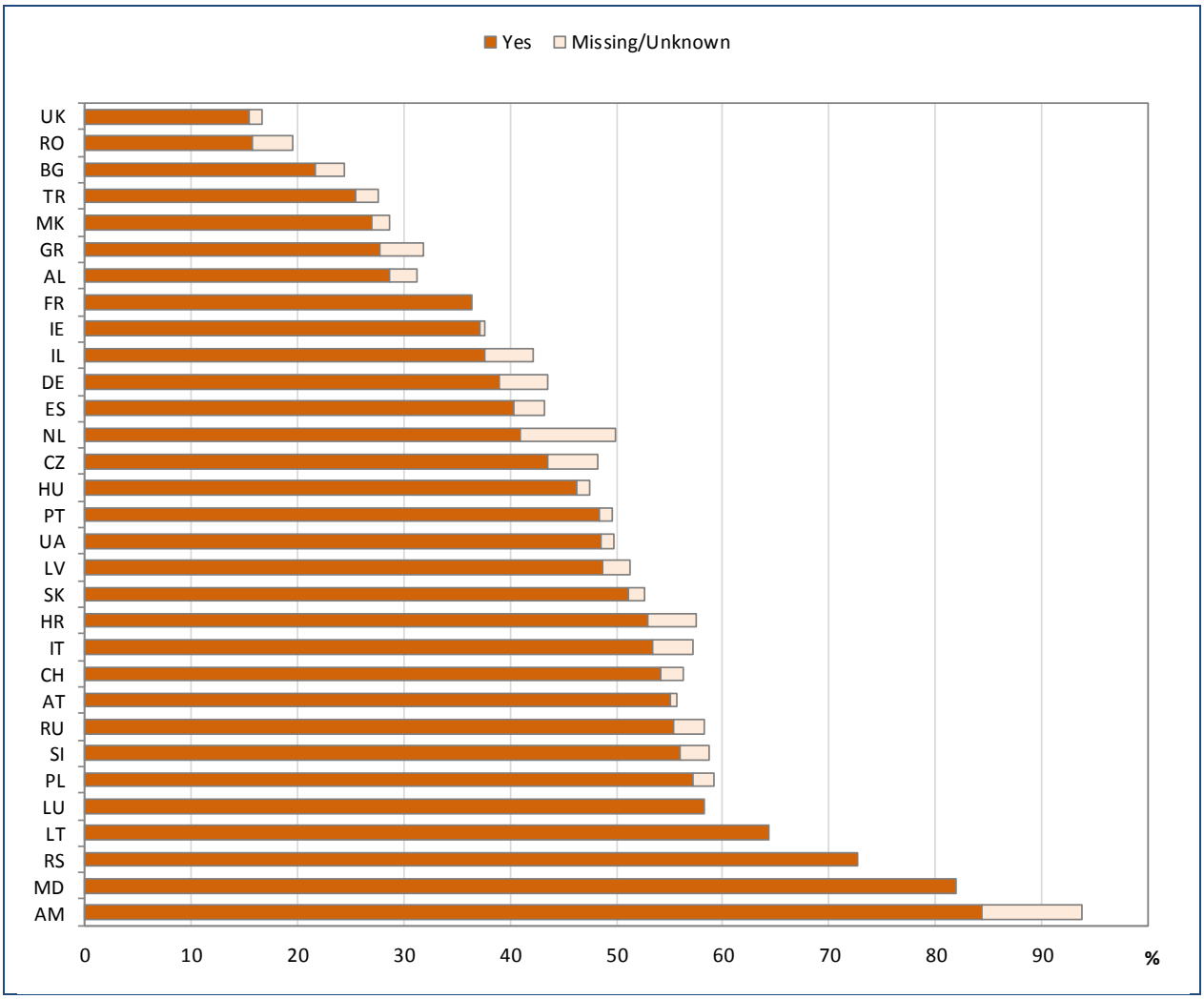


Note: We excluded from the graph the countries for which the information on *Burkholderia cepacia complex species* was missing for more than 10% of the patients.

Note: Ireland: chronicity for *Burkholderia cepacia complex species* is defined as: at least 3 or more positive isolates during the last 12 months window preceding the last reported culture in 2017.
Italy: chronicity for *Burkholderia cepacia complex species* is defined as: at least 3 or more positive cultures during 2017.
United Kingdom: information on *Burkholderia cepacia complex species* is collected as: *Burkholderia cepacia complex species* grown since last annual review, not necessarily chronic.

The horizontal bars represent the percentage of patients with chronic *Burkholderia* infection (in dark orange) and the percentage of patients where information on *Burkholderia* infection was missing (in light orange). This infection is much less frequent than *Pseudomonas aeruginosa* (note the different scale on the horizontal axis), and there is also some variation.

Figure 5.3 Prevalence of chronic Staphylococcus aureus infection in all patients seen in 2017, 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 patients.

Note: Ireland: chronicity for *Staphylococcus aureus* is defined as: at least 3 or more positive isolates during the last 12 months window preceding the last reported culture in 2017.
Italy: chronicity for *Staphylococcus aureus* is defined as: at least 3 or more positive cultures during 2017.
United Kingdom: for chronic Staphylococcus aureus the definition is: 3 or more positive isolates during the last 12 months.

The horizontal bars represent the percentage of patients with chronic *Staphylococcus aureus* infection (in dark orange) and the percentage of patients where information on *Staphylococcus aureus* was missing (in light orange). This infection is as frequent as chronic *Pseudomonas aeruginosa* infection and a similar degree of variation between the countries can be observed.

Table 5.2 Prevalence of chronic bacterial infection in children seen in 2017, by country.

Country	Chronic *Pseudomonas aeruginosa* number (%)			Chronic *Burkholderia cepacia complex species* number (%)			Chronic *Staphylococcus aureus* number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	3 (2.63)	90 (78.95)	21 (18.42)	3 (2.63)	111 (97.37)	0 (0)	3 (2.63)	83 (72.81)	28 (24.56)
Armenia	3 (10.34)	16 (55.17)	10 (34.48)	27 (93.10)	2 (6.90)	0 (0)	2 (6.90)	2 (6.90)	25 (86.21)
Austria	2 (0.55)	326 (89.81)	35 (9.64)	2 (0.55)	355 (97.80)	6 (1.65)	2 (0.55)	158 (43.53)	203 (55.92)
Belgium[1]	11 (2.24)	433 (88.19)	47 (9.57)	11 (2.24)	476 (96.95)	4 (0.81)	491 (100)	-	-
Bulgaria	1 (1.23)	44 (54.32)	36 (44.44)	1 (1.23)	79 (97.53)	1 (1.23)	1 (1.23)	61 (75.31)	19 (23.46)
Croatia	1 (2.22)	34 (75.56)	10 (22.22)	2 (4.44)	43 (95.56)	0 (0)	1 (2.22)	19 (42.22)	25 (55.56)
Czech Republic	15 (4.66)	286 (88.82)	21 (6.52)	14 (4.35)	300 (93.17)	8 (2.48)	14 (4.35)	167 (51.86)	141 (43.79)
Denmark	0 (0)	175 (92.59)	14 (7.41)	0 (0)	187 (98.94)	2 (1.06)	189 (100)	-	-
France	0 (0)	2837 (92.86)	218 (7.14)	0 (0)	3042 (99.57)	13 (0.43)	0 (0)	1875 (61.37)	1180 (38.63)
Germany	70 (2.73)	2231 (87.11)	260 (10.15)	65 (2.54)	2473 (96.56)	23 (0.90)	65 (2.54)	1553 (60.64)	943 (36.82)
Greece	21 (7.32)	190 (66.20)	76 (26.48)	1 (0.35)	286 (99.65)	0 (0)	3 (1.05)	220 (76.66)	64 (22.30)
Hungary	1 (0.38)	192 (73.00)	70 (26.62)	1 (0.38)	258 (98.10)	4 (1.52)	0 (0)	140 (53.23)	123 (46.77)
Ireland[2]	1 (0.19)	478 (90.36)	50 (9.45)	1 (0.19)	521 (98.49)	7 (1.32)	1 (0.19)	290 (54.82)	238 (44.99)
Israel	8 (3.76)	159 (74.65)	46 (21.60)	9 (4.23)	203 (95.31)	1 (0.47)	8 (3.76)	109 (51.17)	96 (45.07)
Italy[3]	98 (4.11)	1914 (80.29)	372 (15.60)	98 (4.11)	2280 (95.64)	6 (0.25)	98 (4.11)	1010 (42.37)	1276 (53.52)
Latvia	2 (7.14)	25 (89.29)	1 (3.57)	1 (3.57)	26 (92.86)	1 (3.57)	1 (3.57)	15 (53.57)	12 (42.86)
Luxembourg	0 (0)	12 (85.71)	2 (14.29)	0 (0)	14 (100)	0 (0)	0 (0)	7 (50.00)	7 (50.00)
Rep of Moldova	0 (0)	23 (51.11)	22 (48.89)	45 (100)	-	-	0 (0)	8 (17.78)	37 (82.22)
The Netherlands	14 (2.50)	476 (85.00)	70 (12.50)	14 (2.50)	537 (95.89)	9 (1.61)	14 (2.50)	334 (59.64)	212 (37.86)
North Macedonia	1 (1.25)	59 (73.75)	20 (25.00)	1 (1.25)	79 (98.75)	0 (0)	1 (1.25)	59 (73.75)	20 (25.00)
Norway	4 (4.49)	79 (88.76)	6 (6.74)	6 (6.74)	82 (92.13)	1 (1.12)	7 (7.87)	41 (46.07)	41 (46.07)

[1] Belgium: *Chronic Pseudomonas aeruginosa, Chronic Burkholderia cepacia complex species, and Chronic Staphylococcus aureus* are not collected for transplanted patients and most of the missing data refers to this sub-population.

[2] Ireland: chronicity for *Pseudomonas aeruginosa*, *Burkholderia* and *Staphylococcus aureus* is defined as: at least 3 or more positive isolates during the last 12 months window preceding the last reported culture in 2017.

[3] Italy: chronicity for *Pseudomonas aeruginosa*, *Burkholderia* and *Staphylococcus aureus* is defined as: at least 3 or more positive cultures during 2017.

Note: Lithuania has 0% coverage for children.

[table 5.2 continued]

Country	Chronic *Pseudomonas aeruginosa* number (%)			Chronic *Burkholderia cepacia complex species* number (%)			Chronic *Staphylococcus aureus* number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Poland	10 (1.88)	451 (84.62)	72 (13.51)	9 (1.69)	521 (97.75)	3 (0.56)	9 (1.69)	211 (39.59)	313 (58.72)
Portugal	1 (0.54)	143 (77.72)	40 (21.74)	1 (0.54)	172 (93.48)	11 (5.98)	1 (0.54)	98 (53.26)	85 (46.20)
Romania	5 (3.25)	115 (74.68)	34 (22.08)	6 (3.90)	148 (96.10)	0 (0)	5 (3.25)	125 (81.17)	24 (15.58)
Russian Federation	49 (2.06)	1715 (72.18)	612 (25.76)	27 (1.14)	2247 (94.57)	102 (4.29)	39 (1.64)	973 (40.95)	1364 (57.41)
Serbia	0 (0)	86 (71.07)	35 (28.93)	0 (0)	111 (91.74)	10 (8.26)	0 (0)	28 (23.14)	93 (76.86)
Slovak Republic	0 (0)	96 (78.05)	27 (21.95)	0 (0)	122 (99.19)	1 (0.81)	0 (0)	62 (50.41)	61 (49.59)
Slovenia	0 (0)	56 (88.89)	7 (11.11)	0 (0)	63 (100)	0 (0)	0 (0)	26 (41.27)	37 (58.73)
Spain	3 (0.28)	930 (87.00)	136 (12.72)	9 (0.84)	1032 (96.54)	28 (2.62)	4 (0.37)	657 (61.46)	408 (38.17)
Sweden	0 (0)	213 (83.86)	41 (16.14)	0 (0)	252 (99.21)	2 (0.79)	20 (7.87)	190 (74.80)	44 (17.32)
Switzerland	5 (1.18)	376 (88.47)	44 (10.35)	3 (0.71)	419 (98.59)	3 (0.71)	5 (1.18)	194 (45.65)	226 (53.18)
Turkey	24 (1.83)	1017 (77.52)	271 (20.66)	32 (2.44)	1272 (96.95)	8 (0.61)	26 (1.98)	963 (73.40)	323 (24.62)
Ukraine	0 (0)	92 (65.71)	48 (34.29)	0 (0)	134 (95.71)	6 (4.29)	0 (0)	72 (51.43)	68 (48.57)
United Kingdom[4]	6 (0.14)	3948 (93.11)	286 (6.75)	6 (0.14)	4171 (98.37)	63 (1.49)	6 (0.14)	3882 (91.56)	352 (8.30)

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

Table 5.2 shows, separately by country, the frequency of chronic *Pseudomonas aeruginosa*, chronic *Burkholderia cepacia complex species* and chronic *Staphylococcus aureus* in children. The number of missing values is also included. The identification rate of *Burkholderia cepacia complex species* in particular may also be influenced by differences in culture techniques employed.

Table 5.3 Prevalence of chronic bacterial infection in adults seen in 2017, by country.

Country	Chronic *Pseudomonas aeruginosa* number (%)			Chronic *Burkholderia cepacia complex species* number (%)			Chronic *Staphylococcus aureus* number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	1 (12.50)	5 (62.50)	2 (25.00)	0 (0)	8 (100)	0 (0)	0 (0)	1 (12.50)	7 (87.50)
Austria	3 (0.76)	210 (53.30)	181 (45.94)	0 (0)	374 (94.92)	20 (5.08)	2 (0.51)	178 (45.18)	214 (54.31)
Belgium[1]	205 (25.75)	355 (44.60)	236 (29.65)	205 (25.75)	570 (71.61)	21 (2.64)	796 (100)	-	-
Bulgaria	3 (4.48)	8 (11.94)	56 (83.58)	3 (4.48)	63 (94.03)	1 (1.49)	3 (4.48)	51 (76.12)	13 (19.40)
Croatia	3 (7.14)	9 (21.43)	30 (71.43)	3 (7.14)	38 (90.48)	1 (2.38)	3 (7.14)	18 (42.86)	21 (50.00)
Czech Republic	16 (5.65)	168 (59.36)	99 (34.98)	17 (6.01)	226 (79.86)	40 (14.13)	15 (5.3)	146 (51.59)	122 (43.11)
Denmark	0 (0)	164 (53.42)	143 (46.58)	0 (0)	276 (89.90)	31 (10.10)	307 (100)	-	-
France	0 (0)	2680 (68.98)	1205 (31.02)	0 (0)	3811 (98.10)	74 (1.90)	0 (0)	2543 (65.46)	1342 (34.54)
Germany	208 (5.85)	1505 (42.30)	1845 (51.85)	208 (5.85)	3246 (91.23)	104 (2.92)	212 (5.96)	1904 (53.51)	1442 (40.53)
Greece	22 (7.05)	95 (30.45)	195 (62.50)	21 (6.73)	289 (92.63)	2 (0.64)	22 (7.05)	188 (60.26)	102 (32.69)
Hungary	5 (2.12)	107 (45.34)	124 (52.54)	5 (2.12)	224 (94.92)	7 (2.97)	6 (2.54)	120 (50.85)	110 (46.61)
Ireland[2]	4 (0.58)	414 (60.00)	272 (39.42)	4 (0.58)	664 (96.23)	22 (3.19)	4 (0.58)	471 (68.26)	215 (31.16)
Israel	16 (4.79)	127 (38.02)	191 (57.19)	17 (5.09)	310 (92.81)	7 (2.10)	17 (5.09)	207 (61.98)	110 (32.93)
Italy[3]	111 (3.49)	1435 (45.17)	1631 (51.34)	111 (3.49)	2947 (92.76)	119 (3.75)	112 (3.53)	1368 (43.06)	1697 (53.42)
Latvia	0 (0)	4 (36.36)	7 (63.64)	0 (0)	11 (100)	0 (0)	0 (0)	4 (36.36)	7 (63.64)
Lithuania	0 (0)	12 (85.71)	2 (14.29)	0 (0)	13 (92.86)	1 (7.14)	0 (0)	5 (35.71)	9 (64.29)
Luxembourg	0 (0)	15 (68.18)	7 (31.82)	0 (0)	20 (90.91)	2 (9.09)	0 (0)	8 (36.36)	14 (63.64)
The Netherlands	118 (12.97)	395 (43.41)	397 (43.63)	118 (12.97)	774 (85.05)	18 (1.98)	118 (12.97)	402 (44.18)	390 (42.86)
North Macedonia	1 (2.86)	12 (34.29)	22 (62.86)	1 (2.86)	34 (97.14)	0 (0)	1 (2.86)	23 (65.71)	11 (31.43)
Norway	26 (16.05)	81 (50.00)	55 (33.95)	25 (15.43)	130 (80.25)	7 (4.32)	28 (17.28)	51 (31.48)	83 (51.23)
Poland	3 (2.44)	48 (39.02)	72 (58.54)	4 (3.25)	112 (91.06)	7 (5.69)	4 (3.25)	57 (46.34)	62 (50.41)

[1] Belgium: *Chronic Pseudomonas aeruginosa* and *Chronic Burkholderia cepacia complex species* are not collected for transplanted patients and most of the missing data refers to this sub-population. *Chronic Staphylococcus* aureus is not collected.

[2] Ireland: chronicity for *Pseudomonas aeruginosa*, *Burkholderia* and *Staphylococcus aureus* is defined as: at least 3 or more positive isolates during the last 12 months window preceding the last reported culture in 2017.

[3] Italy: chronicity for *Pseudomonas aeruginosa*, *Burkholderia* and *Staphylococcus aureus* is defined as: at least 3 or more positive cultures during 2017.

[table 5.3 continued]

Country	Chronic *Pseudomonas aeruginosa* number (%)			Chronic *Burkholderia cepacia complex species* number (%)			Chronic *Staphylococcus aureus* number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Portugal	2 (1.40)	80 (55.94)	61 (42.66)	3 (2.10)	131 (91.61)	9 (6.29)	3 (2.10)	67 (46.85)	73 (51.05)
Russian Federation	47 (6.68)	305 (43.32)	352 (50.00)	45 (6.39)	574 (81.53)	85 (12.07)	53 (7.53)	311 (44.18)	340 (48.30)
Serbia	0 (0)	13 (25.49)	38 (74.51)	0 (0)	40 (78.43)	11 (21.57)	0 (0)	19 (37.25)	32 (62.75)
Slovak Republic	3 (2.10)	75 (52.45)	65 (45.45)	3 (2.10)	128 (89.51)	12 (8.39)	4 (2.80)	64 (44.76)	75 (52.45)
Slovenia	2 (4.35)	27 (58.70)	17 (36.96)	4 (8.70)	41 (89.13)	1 (2.17)	3 (6.52)	19 (41.30)	24 (52.17)
Spain	53 (5.69)	516 (55.36)	363 (38.95)	54 (5.79)	818 (87.77)	60 (6.44)	54 (5.79)	479 (51.39)	399 (42.81)
Sweden	0 (0)	193 (44.68)	239 (55.32)	0 (0)	417 (96.53)	15 (3.47)	59 (13.66)	233 (53.94)	140 (32.41)
Switzerland	12 (2.45)	217 (44.38)	260 (53.17)	15 (3.07)	452 (92.43)	22 (4.50)	15 (3.07)	205 (41.92)	269 (55.01)
Turkey	3 (3.03)	45 (45.45)	51 (51.52)	4 (4.04)	89 (89.90)	6 (6.06)	4 (4.04)	59 (59.60)	36 (36.36)
Ukraine	3 (12.00)	3 (12.00)	19 (76.00)	2 (8.00)	22 (88.00)	1 (4.00)	2 (8.00)	11 (44.00)	12 (48.00)
United Kingdom[4]	109 (1.93)	3075 (54.45)	2463 (43.62)	109 (1.93)	5256 (93.08)	282 (4.99)	109 (1.93)	4365 (77.30)	1173 (20.77)

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

Note: Armenia, Rep of Moldova and Romania have <5 patients aged 18 years or more at 31/12/2017 and are excluded from this table.

This table shows, separately by country, the frequency of chronic *Pseudomonas aeruginosa*, chronic *Burkholderia cepacia complex species* and chronic *Staphylococcus aureus* in adults. The number of missing values is also included. The identification rate of *Burkholderia cepacia complex species* in particular may also be influenced by differences in culture techniques employed.

Table 5.4 Prevalence of non-tuberculous mycobacteria and Stenotrophomonas maltophilia infection in all patients seen in 2017, by country.

Country	Non-tuberculous mycobacteria (NTM) infection this year number (%)			*Stenotrophomonas maltophilia* infection this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	1 (0.82)	121 (99.18)	0 (0)	1 (0.82)	121 (99.18)	0 (0)
Armenia	14 (43.75)	17 (53.13)	1 (3.13)	30 (93.75)	2 (6.25)	0 (0)
Austria	22 (2.91)	700 (92.47)	35 (4.62)	2 (0.26)	671 (88.64)	84 (11.10)
Belgium[1]	174 (13.52)	1094 (85.00)	19 (1.48)	174 (13.52)	980 (76.15)	133 (10.33)
Bulgaria	147 (99.32)	1 (0.68)	0 (0)	5 (3.38)	141 (95.27)	2 (1.35)
Croatia	22 (25.29)	62 (71.26)	3 (3.45)	6 (6.90)	74 (85.06)	7 (8.05)
Czech Republic	292 (48.26)	301 (49.75)	12 (1.98)	26 (4.30)	532 (87.93)	47 (7.77)
Denmark	0 (0)	480 (96.77)	16 (3.23)	191 (38.51)	282 (56.85)	23 (4.64)
France	0 (0)	6757 (97.36)	183 (2.64)	0 (0)	6206 (89.42)	734 (10.58)
Germany	4052 (66.22)	1890 (30.89)	177 (2.89)	257 (4.20)	5275 (86.21)	587 (9.59)
Greece	356 (59.43)	228 (38.06)	15 (2.50)	34 (5.68)	514 (85.81)	51 (8.51)
Hungary	13 (2.58)	484 (96.03)	7 (1.39)	5 (0.99)	480 (95.24)	19 (3.77)
Ireland	5 (0.41)	1179 (96.72)	35 (2.87)	5 (0.41)	1108 (90.89)	106 (8.70)
Israel	28 (5.12)	477 (87.20)	42 (7.68)	35 (6.40)	464 (84.83)	48 (8.78)
Italy	48 (0.86)	5452 (98.04)	61 (1.10)	48 (0.86)	5193 (93.38)	320 (5.75)
Latvia	28 (71.79)	11 (28.21)	0 (0)	1 (2.56)	33 (84.62)	5 (12.82)
Lithuania	0 (0)	13 (92.86)	1 (7.14)	0 (0)	14 (100)	0 (0)
Luxembourg	0 (0)	33 (91.67)	3 (8.33)	0 (0)	30 (83.33)	6 (16.67)
Rep of Moldova	50 (100)	-	-	50 (100)	-	-
The Netherlands	180 (12.24)	1251 (85.10)	39 (2.65)	133 (9.05)	1159 (78.84)	178 (12.11)
North Macedonia	2 (1.74)	112 (97.39)	1 (0.87)	2 (1.74)	113 (98.26)	0 (0)
Norway	27 (10.76)	211 (84.06)	13 (5.18)	5 (1.99)	200 (79.68)	46 (18.33)

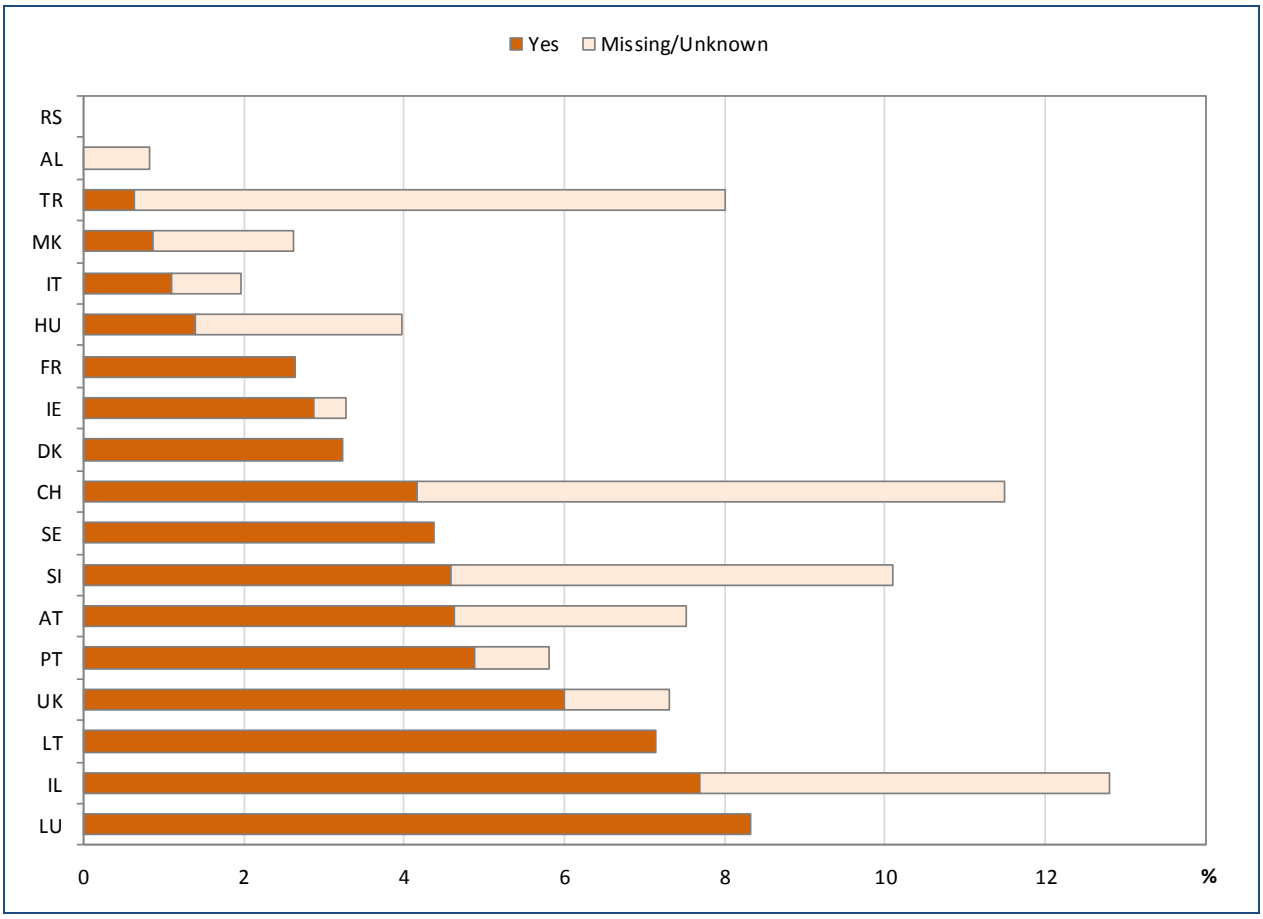
[1] Belgium: *Non-tuberculous mycobacteria* and *Stenotrophomonas maltophilia* infections are not collected for transplanted patients and most of the missing data refers to this sub-population.

[table 5.4 continued]

Country	Non-tuberculous mycobacteria (NTM) infection this year number (%)			*Stenotrophomonas maltophilia* infection this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Poland	155 (23.63)	497 (75.76)	4 (0.61)	16 (2.44)	614 (93.60)	26 (3.96)
Portugal	3 (0.92)	308 (94.19)	16 (4.89)	3 (0.92)	291 (88.99)	33 (10.09)
Romania	27 (16.98)	132 (83.02)	0 (0)	11 (6.92)	148 (93.08)	0 (0)
Russian Federation	756 (24.55)	2307 (74.90)	17 (0.55)	93 (3.02)	2882 (93.57)	105 (3.41)
Serbia	0 (0)	172 (100)	0 (0)	0 (0)	158 (91.86)	14 (8.14)
Slovak Republic	31 (11.65)	234 (87.97)	1 (0.38)	4 (1.50)	244 (91.73)	18 (6.77)
Slovenia	6 (5.50)	98 (89.91)	5 (4.59)	10 (9.17)	93 (85.32)	6 (5.50)
Spain	261 (13.04)	1679 (83.87)	62 (3.10)	44 (2.20)	1807 (90.26)	151 (7.54)
Sweden	0 (0)	656 (95.63)	30 (4.37)	0 (0)	626 (91.25)	60 (8.75)
Switzerland	67 (7.33)	809 (88.51)	38 (4.16)	20 (2.19)	795 (86.98)	99 (10.83)
Turkey	104 (7.37)	1298 (91.99)	9 (0.64)	25 (1.77)	1346 (95.39)	40 (2.83)
Ukraine	164 (99.39)	1 (0.61)	0 (0)	5 (3.03)	152 (92.12)	8 (4.85)
United Kingdom	130 (1.31)	9165 (92.70)	592 (5.99)	115 (1.16)	9030 (91.33)	742 (7.50)

Table 5.4 shows the frequency of two other infections, non-tuberculous mycobacteria (NTM) and *Stenotrophomonas maltophilia.* Both these infections seem to be relatively rare, in line with the frequencies of *Burkholderia* infection. The identification rate of these bacteria may also be influenced by differences in culture techniques employed.

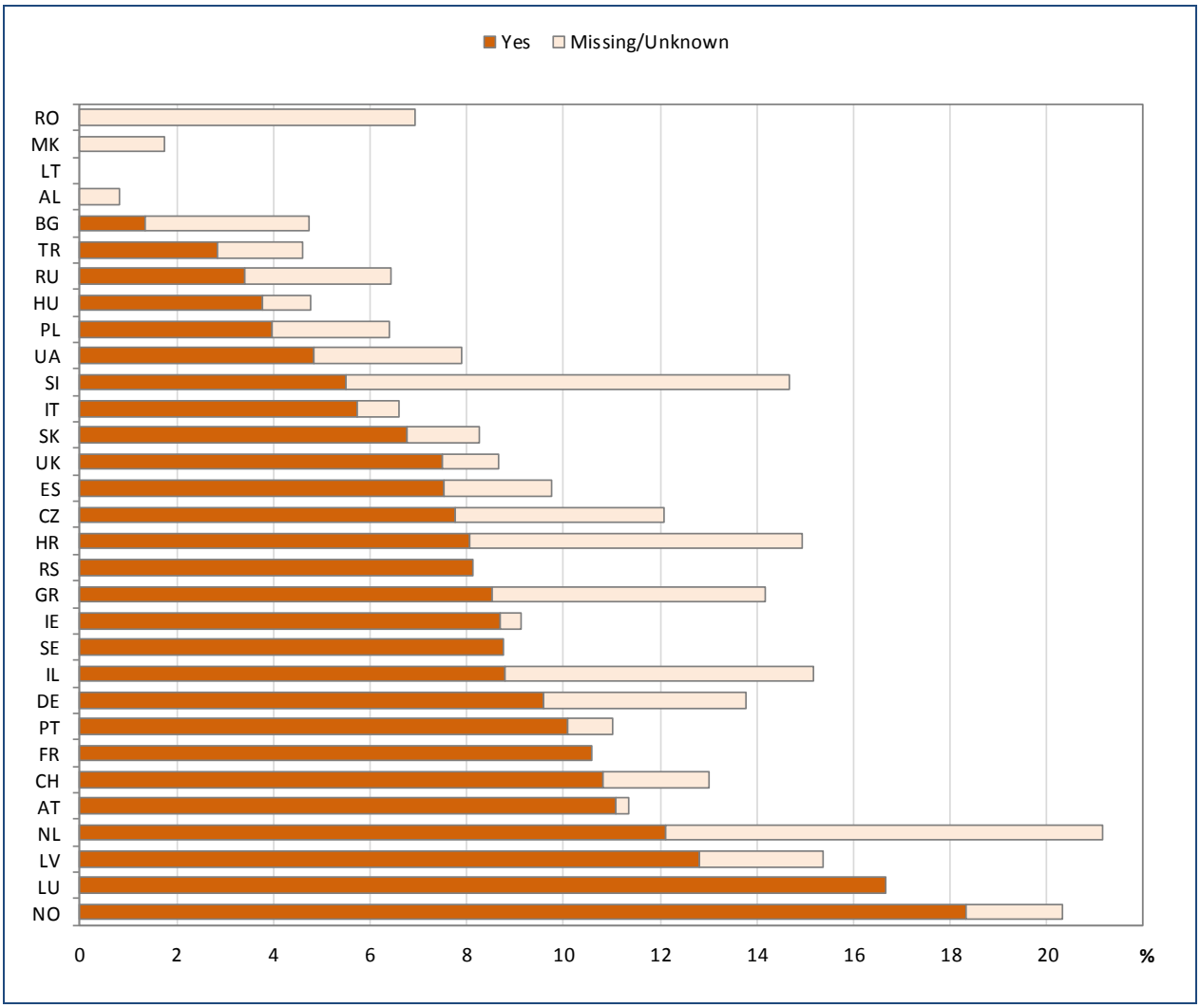
Figure 5.4 Prevalence of non-tuberculous mycobacteria in all patients seen in 2017, by country.



Note: We excluded from the graph the countries for which the information on non-tuberculous mycobacteria was missing for more than 10% of the patients.

The horizontal bars represent the percentage of patients with non-tuberculous mycobacteria infection (in dark orange) and the percentage of patients where information on non-tuberculous mycobacteria infection was missing (in light orange). Generally, infections from these bacteria are not very frequent in any country.

Figure 5.5 Prevalence of Stenotrophomonas maltophilia infection in all patients seen in 2017, 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 patients.

The horizontal bars represent the percentage of patients with *Stenotrophomonas maltophilia* infection (in dark orange) and the percentage of patients where information on *Stenotrophomonas maltophilia* was missing (light orange). The frequency varies considerably between countries.

Table 5.5 Prevalence of non-tuberculous mycobacteria and Stenotrophomonas maltophilia infection in children seen in 2017, by country.

Country	Non-tuberculous mycobacteria (NTM) infection this year number (%)			*Stenotrophomonas maltophilia* infection this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	1 (0.88)	113 (99.12)	0 (0)	1 (0.88)	113 (99.12)	0 (0)
Armenia	12 (41.38)	16 (55.17)	1 (3.45)	27 (93.1)	2 (6.9)	0 (0)
Austria	19 (5.23)	340 (93.66)	4 (1.1)	1 (0.28)	329 (90.63)	33 (9.09)
Belgium[1]	3 (0.61)	485 (98.78)	3 (0.61)	3 (0.61)	420 (85.54)	68 (13.85)
Bulgaria	80 (98.77)	1 (1.23)	0 (0)	2 (2.47)	79 (97.53)	0 (0)
Croatia	17 (37.78)	27 (60.00)	1 (2.22)	3 (6.67)	39 (86.67)	3 (6.67)
Czech Republic	249 (77.33)	72 (22.36)	1 (0.31)	14 (4.35)	282 (87.58)	26 (8.07)
Denmark[2]	0 (0)	186 (98.41)	3 (1.59)	97 (51.32)	86 (45.50)	6 (3.17)
France	0 (0)	3010 (98.53)	45 (1.47)	0 (0)	2703 (88.48)	352 (11.52)
Germany	2027 (79.15)	492 (19.21)	42 (1.64)	61 (2.38)	2252 (87.93)	248 (9.68)
Greece	179 (62.37)	106 (36.93)	2 (0.70)	1 (0.35)	268 (93.38)	18 (6.27)
Hungary	6 (2.28)	256 (97.34)	1 (0.38)	0 (0)	256 (97.34)	7 (2.66)
Ireland	1 (0.19)	519 (98.11)	9 (1.70)	1 (0.19)	479 (90.55)	49 (9.26)
Israel	10 (4.69)	190 (89.20)	13 (6.10)	13 (6.10)	178 (83.57)	22 (10.33)
Italy	18 (0.76)	2352 (98.66)	14 (0.59)	18 (0.76)	2253 (94.51)	113 (4.74)
Latvia	19 (67.86)	9 (32.14)	0 (0)	1 (3.57)	23 (82.14)	4 (14.29)
Luxembourg	0 (0)	14 (100)	0 (0)	0 (0)	10 (71.43)	4 (28.57)
Rep of Moldova	45 (100)	-	-	45 (100)	-	-
The Netherlands	14 (2.50)	533 (95.18)	13 (2.32)	15 (2.68)	475 (84.82)	70 (12.50)
North Macedonia	1 (1.25)	79 (98.75)	0 (0)	1 (1.25)	79 (98.75)	0 (0)
Norway	6 (6.74)	79 (88.76)	4 (4.49)	3 (3.37)	71 (79.78)	15 (16.85)

[1] Belgium: *Non-tuberculous mycobacteria* and *Stenotrophomonas maltophilia* infections are not collected for transplanted patients and most of the missing data refers to this sub-population.

[2] Denmark: the high number of missing information is due to only one of two centres reporting these data.

Note: Lithuania has 0% coverage for children.

[table 5.5 continued]

Country	Non-tuberculous mycobacteria (NTM) infection this year number (%)			*Stenotrophomonas maltophilia* infection this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Poland	104 (19.51)	425 (79.74)	4 (0.75)	11 (2.06)	497 (93.25)	25 (4.69)
Portugal	1 (0.54)	181 (98.37)	2 (1.09)	1 (0.54)	153 (83.15)	30 (16.30)
Romania	26 (16.88)	128 (83.12)	0 (0)	10 (6.49)	144 (93.51)	0 (0)
Russian Federation	552 (23.23)	1814 (76.35)	10 (0.42)	39 (1.64)	2255 (94.91)	82 (3.45)
Serbia	0 (0)	121 (100)	0 (0)	0 (0)	110 (90.91)	11 (9.09)
Slovak Republic	24 (19.51)	99 (80.49)	0 (0)	0 (0)	120 (97.56)	3 (2.44)
Slovenia	0 (0)	60 (95.24)	3 (4.76)	0 (0)	61 (96.83)	2 (3.17)
Spain	186 (17.40)	862 (80.64)	21 (1.96)	7 (0.65)	976 (91.30)	86 (8.04)
Sweden	0 (0)	249 (98.03)	5 (1.97)	0 (0)	238 (93.70)	16 (6.30)
Switzerland	18 (4.24)	405 (95.29)	2 (0.47)	5 (1.18)	384 (90.35)	36 (8.47)
Turkey	97 (7.39)	1209 (92.15)	6 (0.46)	22 (1.68)	1261 (96.11)	29 (2.21)
Ukraine	139 (99.29)	1 (0.71)	0 (0)	2 (1.43)	131 (93.57)	7 (5.00)
United Kingdom	10 (0.24)	4094 (96.56)	136 (3.21)	6 (0.14)	3952 (93.21)	282 (6.65)

Table 5.6 Prevalence of non-tuberculous mycobacteria and Stenotrophomonas maltophilia infection in adults seen in 2017, by country.

Country	Non-tuberculous mycobacteria (NTM) infection this year number (%)			*Stenotrophomonas maltophilia* infection this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	0 (0)	8 (100)	0 (0)	0 (0	8 (100)	0 (0)
Austria	3 (0.76)	360 (91.37)	31 (7.87)	1 (0.25)	342 (86.80)	51 (12.94)
Belgium[1]	171 (21.48)	609 (76.51)	16 (2.01)	171 (21.48)	560 (70.35)	65 (8.17)
Bulgaria	67 (100)	-	-	3 (4.48)	62 (92.54)	2 (2.99)
Croatia	5 (11.90)	35 (83.33)	2 (4.76)	3 (7.14)	35 (83.33)	4 (9.52)
Czech Republic	43 (15.19)	229 (80.92)	11 (3.89)	12 (4.24)	250 (88.34)	21 (7.42)
Denmark[2]	0 (0)	294 (95.77)	13 (4.23)	94 (30.62)	196 (63.84)	17 (5.54)
France	0 (0)	3747 (96.45)	138 (3.55)	0 (0)	3503 (90.17)	382 (9.83)
Germany	2025 (56.91)	1398 (39.29)	135 (3.79)	196 (5.51)	3023 (84.96)	339 (9.53)
Greece	177 (56.73)	122 (39.10)	13 (4.17)	33 (10.58)	246 (78.85)	33 (10.58)
Hungary	7 (2.97)	223 (94.49)	6 (2.54)	5 (2.12)	219 (92.80)	12 (5.08)
Ireland	4 (0.58)	660 (95.65)	26 (3.77)	4 (0.58)	629 (91.16)	57 (8.26)
Israel	18 (5.39)	287 (85.93)	29 (8.68)	22 (6.59)	286 (85.63)	26 (7.78)
Italy	30 (0.94)	3100 (97.58)	47 (1.48)	30 (0.94)	2940 (92.54)	207 (6.52)
Latvia	9 (81.82)	2 (18.18)	0 (0)	0 (0)	10 (90.91)	1 (9.09)
Lithuania	0 (0)	13 (92.86)	1 (7.14)	0 (0)	14 (100)	0 (0)
Luxembourg	0 (0)	19 (86.36)	3 (13.64)	0 (0)	20 (90.91)	2 (9.09)
The Netherlands	166 (18.24)	718 (78.90)	26 (2.86)	118 (12.97)	684 (75.16)	108 (11.87)
North Macedonia	1 (2.86)	33 (94.29)	1 (2.86)	1 (2.86)	34 (97.14)	0 (0)
Norway	21 (12,96)	132 (81,48)	9 (5,56)	2 (1,23)	129 (79,63)	31 (19,14)

[1] Belgium: *Non-tuberculous mycobacteria* and *Stenotrophomonas maltophilia* infections are not collected for transplanted patients and most of the missing data refers to this sub-population.

[2] Denmark: the high number of missing information is due to only one of two centres reporting these data.

[table 5.6 continued]

Country	Non-tuberculous mycobacteria (NTM) infection this year number (%)			*Stenotrophomonas maltophilia* infection this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Poland	51 (41.46)	72 (58.54)	0 (0)	5 (4.07)	117 (95.12)	1 (0.81)
Portugal	2 (1.40)	127 (88.81)	14 (9.79)	2 (1.40)	138 (96.50)	3 (2.10)
Russian Federation	204 (28.98)	493 (70.03)	7 (0.99)	54 (7.67)	627 (89.06)	23 (3.27)
Serbia	0 (0)	51 (100)	0 (0)	0 (0)	48 (94.12)	3 (5.88)
Slovak Republic	7 (4.90)	135 (94.41)	1 (0.70)	4 (2.80)	124 (86.71)	15 (10.49)
Slovenia	6 (13.04)	38 (82.61)	2 (4.35)	10 (21.74)	32 (69.57)	4 (8.70)
Spain	74 (7.94)	817 (87.66)	41 (4.40)	36 (3.86)	831 (89.16)	65 (6.97)
Sweden	0 (0)	407 (94.21)	25 (5.79)	0 (0)	388 (89.81)	44 (10.19)
Switzerland	49 (10.02)	404 (82.62)	36 (7.36)	15 (3.07)	411 (84.05)	63 (12.88)
Turkey	7 (7.07)	89 (89.90)	3 (3.03)	3 (3.03)	85 (85.86)	11 (11.11)
Ukraine	25 (100)	-	-	3 (12.00)	21 (84.00)	1 (4.00)
United Kingdom	120 (2.13)	5071 (89.80)	456 (8.08)	109 (1.93)	5078 (89.92)	460 (8.15)

Note: Armenia, Rep of Moldova and Romania have <5 patients aged 18 years or more at 31/12/2017 and are excluded from this table.

6. Nutrition

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

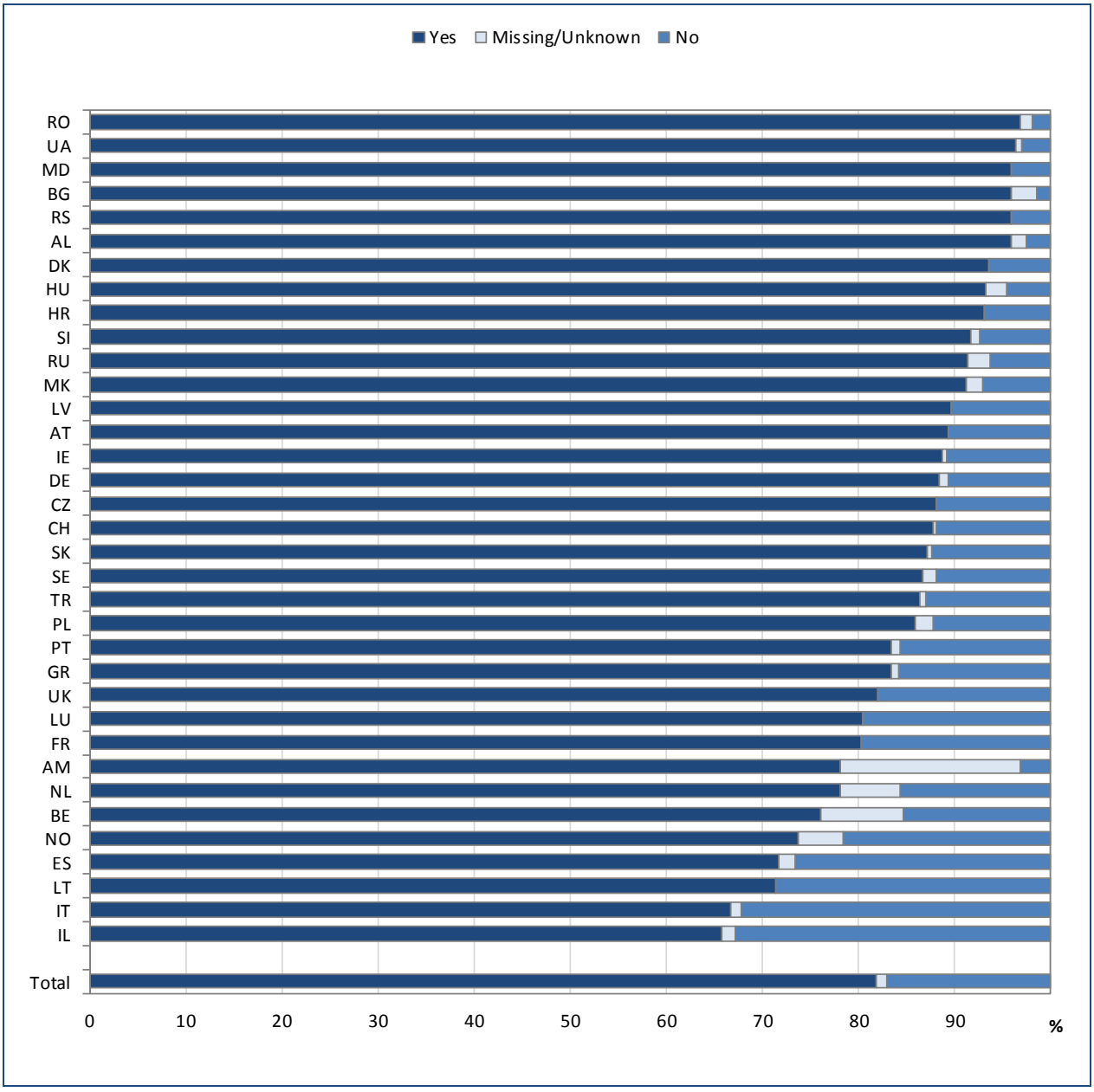
We collected weight and height measured on the date the best FEV1 value was recorded and, for patients that did not perform spirometry, the last measurements of the year were considered. From these raw values we calculated body mass index (BMI). A patient with a low weight is not necessarily underweight if the height is also low, and BMI may better illustrate the nutritional status; BMI describes the weight/height relationship and is considered a good measure of nutritional status. The ECFS Standards of Care guidelines recommend: for adults, a BMI of above 20 kg/m^2; for older children and adolescents, the 50th percentile for BMI; for infants and children up to 2 years of age, weight and height percentiles similar to those for the non-CF population.[2]

Weight, height and BMI were then expressed in terms of so-called 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 1, page 143, 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.

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

Figure 6.1 Use of pancreatic enzymes in 2017 for all patients, by country and overall.



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

Table 6.1 Number of patients for whom height and weight measurements were available. All patients seen in 2017.

Country	Number of patients	Height		Weight	
		N (%)	N miss (%)	N (%)	N miss (%)
Albania[1]	122	66 (54.10)	56 (45.90)	66 (54.10)	56 (45.90)
Armenia	32	32 (100)	0 (0)	32 (100)	0 (0)
Austria	757	755 (99.74)	2 (0.26)	755 (99.74)	2 (0.26)
Belgium	1287	1246 (96.81)	41 (3.19)	1246 (96.81)	41 (3.19)
Bulgaria	148	132 (89.19)	16 (10.81)	131 (88.51)	17 (11.49)
Croatia	87	87 (100)	0 (0)	87 (100)	0 (0)
Czech Republic	605	577 (95.37)	28 (4.63)	577 (95.37)	28 (4.63)
Denmark	496	491 (98.99)	5 (1.01)	481 (96.98)	15 (3.02)
France	6940	6833 (98.46)	107 (1.54)	6836 (98.50)	104 (1.50)
Germany	6119	6064 (99.10)	55 (0.90)	6058 (99.00)	61 (1.00)
Greece	599	573 (95.66)	26 (4.34)	574 (95.83)	25 (4.17)
Hungary	504	469 (93.06)	35 (6.94)	470 (93.25)	34 (6.75)
Ireland	1219	1178 (96.64)	41 (3.36)	1070 (87.78)	149 (12.22)
Israel	547	538 (98.35)	9 (1.65)	538 (98.35)	9 (1.65)
Italy	5561	5319 (95.65)	242 (4.35)	5326 (95.77)	235 (4.23)
Latvia	39	39 (100)	0 (0)	39 (100)	0 (0)
Lithuania	14	14 (100)	0 (0)	14 (100)	0 (0)
Luxembourg	36	34 (94.44)	2 (5.56)	36 (100)	0 (0)
Rep of Moldova	50	49 (98.00)	1 (2.00)	49 (98.00)	1 (2.00)
The Netherlands	1470	1434 (97.55)	36 (2.45)	1432 (97.41)	38 (2.59)
North Macedonia	115	113 (98.26)	2 (1.74)	113 (98.26)	2 (1.74)
Norway	251	243 (96.81)	8 (3.19)	243 (96.81)	8 (3.19)
Poland	656	639 (97.41)	17 (2.59)	645 (98.32)	11 (1.68)
Portugal	327	319 (97.55)	8 (2.45)	319 (97.55)	8 (2.45)
Romania	159	155 (97.48)	4 (2.52)	155 (97.48)	4 (2.52)
Russian Federation	3080	2960 (96.10)	120 (3.90)	2979 (96.72)	101 (3.28)
Serbia	172	168 (97.67)	4 (2.33)	169 (98.26)	3 (1.74)
Slovak Republic	266	257 (96.62)	9 (3.38)	259 (97.37)	7 (2.63)
Slovenia	109	108 (99.08)	1 (0.92)	109 (100)	0 (0)
Spain	2002	1894 (94.61)	108 (5.39)	1899 (94.86)	103 (5.14)
Sweden	686	680 (99.13)	6 (0.87)	677 (98.69)	9 (1.31)
Switzerland	914	909 (99.45)	5 (0.55)	909 (99.45)	5 (0.55)
Turkey	1411	1381 (97.87)	30 (2.13)	1394 (98.80)	17 (1.20)
Ukraine	165	162 (98.18)	3 (1.82)	162 (98.18)	3 (1.82)
United Kingdom[2]	9887	8223 (83.17)	1664 (16.83)	7956 (80.47)	1931 (19.53)

[1] Albania: height and weight for patients of 6 years and older are included.
[2] UK: height and weight at date of annual data is used instead of date of best FEV1. If no lung function measurement, the date of the last visit is used.

Table 6.2 Z-scores for height: descriptive statistics by country. Patients aged 17 years or younger.

Country	N	Mean	Min	25th pctl (25% of the patients are below this z-score for height)	Median (50% of the patients are below this z-score for height)	75th pctl (75% of the patients are below this z-score for height)	Max
Albania	58	-0.5	-4.1	-1.3	-0.6	0.1	1.7
Armenia	29	-0.2	-2.1	-1.1	-0.1	0.6	2.0
Austria	376	0.0	-3.1	-0.6	0.0	0.7	3.7
Belgium	492	-0.4	-3.5	-1.1	-0.4	0.3	2.9
Bulgaria	71	-0.7	-3.3	-1.7	-0.8	0.0	3.1
Croatia	50	0.1	-2.3	-0.5	0.3	0.8	2.4
Czech Republic	314	0.0	-4.6	-0.7	0.0	0.7	2.7
Denmark	195	0.0	-2.1	-0.5	0.0	0.6	2.6
France	3123	-0.4	-5.7	-1.1	-0.4	0.3	5.2
Germany	2649	-0.2	-7.4	-0.9	-0.2	0.5	4.7
Greece	292	-0.2	-4.2	-0.9	-0.2	0.5	2.9
Hungary	260	0.1	-4.1	-0.8	0.1	0.9	8.2
Ireland	520	-0.2	-4.4	-0.9	-0.2	0.4	2.5
Israel	222	-0.5	-4.0	-1.2	-0.5	0.3	3.9
Italy	2384	-0.1	-5.6	-0.9	-0.1	0.6	6.2
Latvia	29	0.3	-2.3	-0.1	0.4	1.1	2.2
Luxembourg	12	0.2	-1.7	-0.7	0.4	0.9	2.5
Rep of Moldova	45	-1.0	-4.9	-1.5	-1.0	-0.3	0.8
The Netherlands	573	0.3	-4.0	-0.4	0.3	1.0	5.3
North Macedonia	82	-0.5	-4.3	-1.3	-0.6	0.3	3.3
Norway[1]	86	0.1	-1.6	-0.5	0.1	0.6	2.2
Poland	528	0.0	-5.7	-0.7	0.1	0.8	5.0
Portugal	182	-0.6	-3.3	-1.3	-0.6	0.2	2.0
Romania	152	-0.5	-4.9	-1.4	-0.5	0.5	4.7
Russian Federation	2362	-0.4	-7.3	-1.2	-0.4	0.4	8.9
Serbia	119	-0.2	-2.7	-1.0	-0.2	0.5	2.9
Slovak Republic	125	0.3	-2.0	-0.6	0.1	1.1	4.0
Slovenia	68	0.2	-1.8	-0.5	0.2	0.7	2.9
Spain	1041	-0.2	-4.1	-0.9	-0.2	0.5	3.9
Sweden	266	-0.1	-4.2	-0.7	-0.1	0.7	2.9
Switzerland	437	-0.2	-3.1	-0.8	-0.2	0.4	3.1
Turkey	1290	-0.4	-9.0	-1.4	-0.5	0.6	9.3
Ukraine	140	-0.5	-2.9	-1.2	-0.4	0.2	4.5
United Kingdom	2914	-0.5	-9.2	-1.2	-0.5	0.3	4.8

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

Note: Lithuania has 0% coverage for children.

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 6.3 Z-scores for height: descriptive statistics by country. Patients aged 18 years or older.

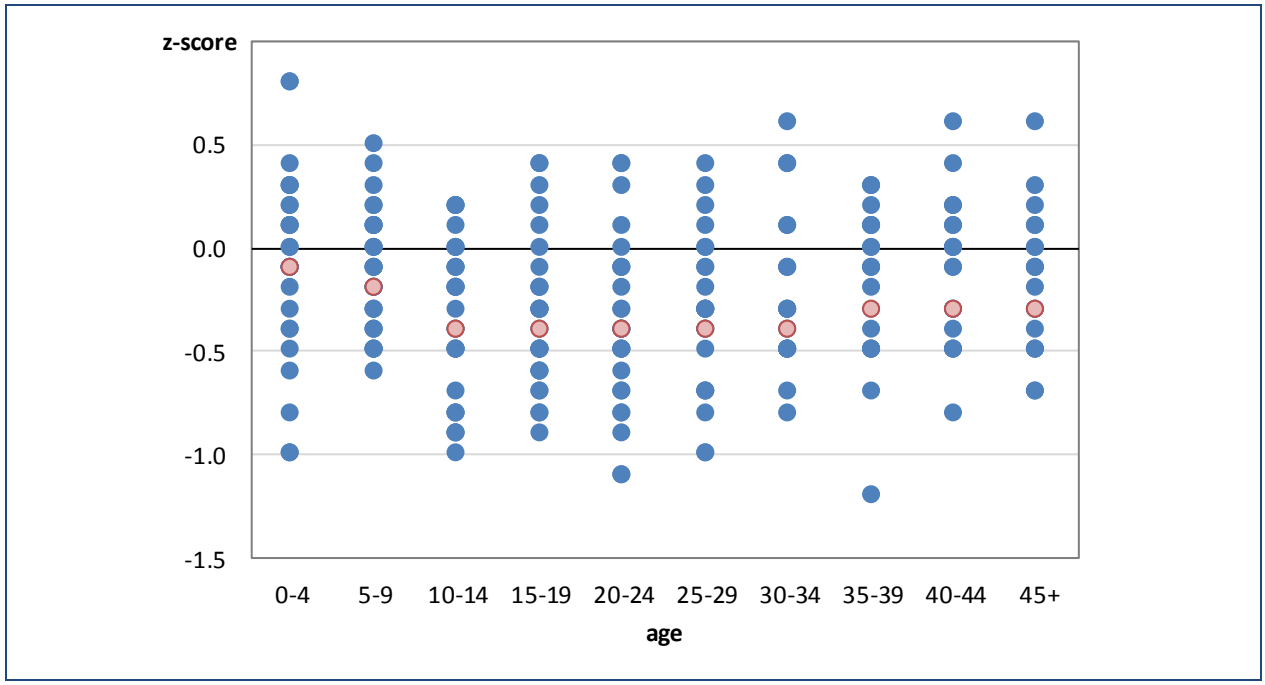
Country	N	Mean	Min	25th pctl (25% of the patients are below this z-score for height)	Median (50% of the patients are below this z-score for height)	75th pctl (75% of the patients are below this z-score for height)	Max
Albania	8	-0.9	-1.7	-1.4	-1.0	-0.7	0.8
Austria	379	-0.3	-3.4	-1.0	-0.3	0.3	2.7
Belgium	754	-0.3	-3.9	-1.0	-0.4	0.4	3.1
Bulgaria	61	-0.3	-2.5	-1.0	-0.5	0.6	1.9
Croatia	37	0.0	-2.1	-0.8	-0.3	0.5	2.5
Czech Republic	263	-0.1	-3.4	-0.7	-0.1	0.4	3.1
Denmark	296	0.2	-3.4	-0.5	0.1	0.9	3.2
France	3710	-0.5	-5.7	-1.2	-0.5	0.1	3.4
Germany	3415	-0.1	-5.6	-0.8	-0.1	0.6	4.0
Greece	281	-0.5	-3.6	-1.2	-0.5	0.2	2.0
Hungary	209	-0.2	-9.4	-1.0	-0.2	0.6	3.6
Ireland	658	-0.4	-5.2	-1.0	-0.4	0.3	2.1
Israel	316	-0.6	-4.3	-1.4	-0.7	0.1	2.4
Italy	2935	-0.6	-4.4	-1.2	-0.5	0.1	3.8
Latvia	10	0.4	-1.1	-0.4	0.6	1.1	1.5
Lithuania	14	1.0	-0.7	0.4	0.9	1.7	2.6
Luxembourg	22	-0.2	-2.6	-1.1	0.0	0.3	2.4
The Netherlands	861	0.3	-3.0	-0.4	0.3	1.0	4.1
North Macedonia	31	-0.5	-2.6	-1.2	-0.5	0.1	2.4
Norway[1]	157	0.2	-2.9	-0.4	0.2	0.9	2.8
Poland	111	-0.3	-2.2	-1.0	-0.3	0.3	2.8
Portugal	137	-0.8	-3.0	-1.5	-0.8	-0.3	1.5
Russian Federation	598	-0.3	-5.7	-1.0	-0.3	0.4	3.4
Serbia	49	0.1	-1.3	-0.4	0.0	0.7	2.3
Slovak Republic	132	0.1	-3.7	-0.5	0.1	0.8	2.4
Slovenia	40	-0.1	-1.5	-0.8	0.0	0.5	2.3
Spain	853	-0.7	-3.8	-1.4	-0.7	-0.1	2.3
Sweden	414	0.1	-2.8	-0.7	0.2	0.7	3.3
Switzerland	472	-0.3	-3.7	-0.8	-0.3	0.4	2.7
Turkey	91	-1.0	-3.6	-1.7	-1.0	-0.5	1.7
Ukraine	22	-0.6	-1.7	-1.3	-0.7	-0.2	0.7
United Kingdom	5309	-0.4	-6.0	-1.0	-0.4	0.3	4.0

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

Note: Armenia, Rep of Moldova and Romania have <5 patients aged 18 years or more at height measurement and are excluded from this 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 2017.



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

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

Table 6.4 Z-scores for height: descriptive statistics by age group. All patients seen in 2017.

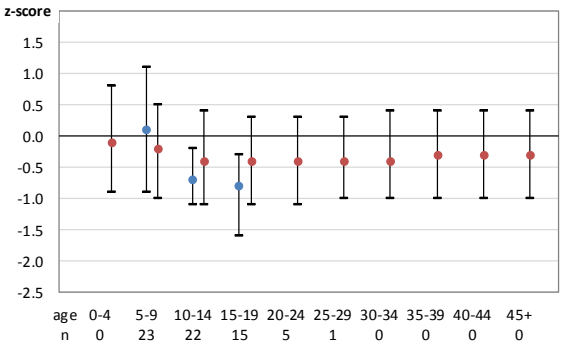
Age at height measurement	N	Mean	Min	25th pctl	Median	75th pctl	Max
0-4	5088	1212	0.0	-9.0	-0.9	-0.1	0.8
5-9	6645	274	-0.2	-7.4	-1.0	-0.2	0.5
10-14	6140	157	-0.4	-5.9	-1.1	-0.4	0.4
15-19	5670	140	-0.4	-9.2	-1.1	-0.4	0.3
20-24	5064	106	-0.4	-6.0	-1.1	-0.4	0.3
25-29	4574	134	-0.4	-4.6	-1.0	-0.4	0.3
30-34	3579	117	-0.3	-4.9	-1.0	-0.4	0.4
35-39	2663	79	-0.3	-9.4	-1.0	-0.3	0.4
40-44	1807	47	-0.3	-3.7	-1.0	-0.3	0.4
45+	2911	86	-0.3	-5.2	-1.0	-0.3	0.4

This table reports the median z-score for height and other descriptive statistics by age group for all the patients seen in 2017. The median values reported in this table are shown as red dots in fig 6.2.

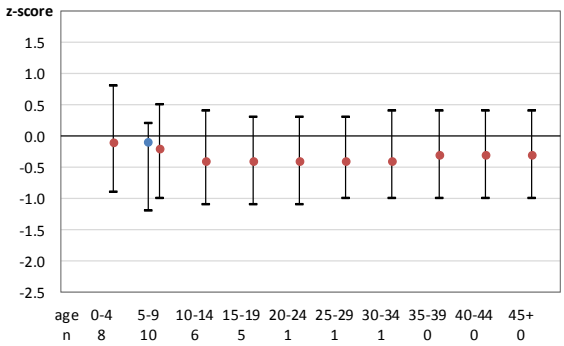
Figure 6.3 Quartiles of z-scores for height by age group and by country. All patients seen in 2017.

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 Latvia, Lithuania and Luxembourg from the graphs because none of the age groups in these countries had more than 10 patients.

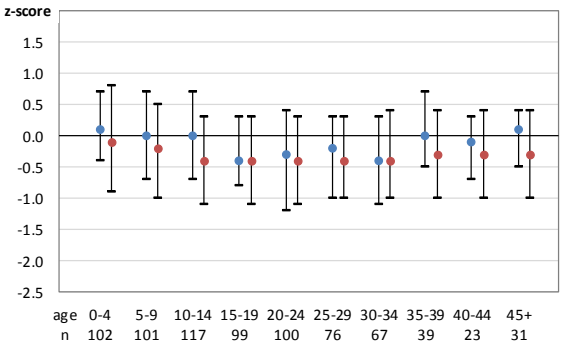
Quartiles of z-scores for height: Albania



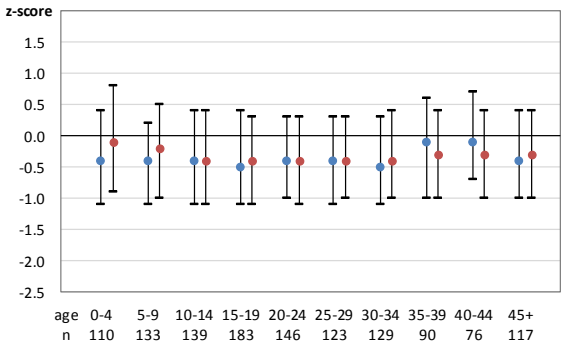
Quartiles of z-scores for height: Armenia



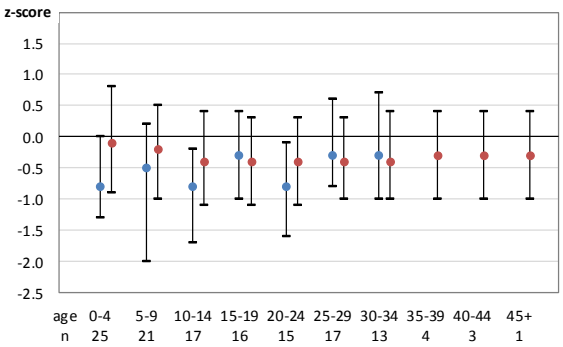
Quartiles of z-scores for height: Austria



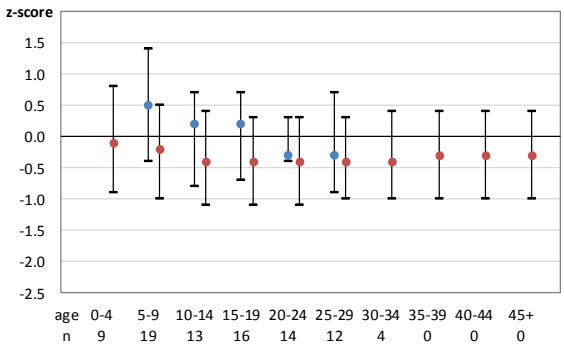
Quartiles of z-scores for height: Belgium



Quartiles of z-scores for height: Bulgaria

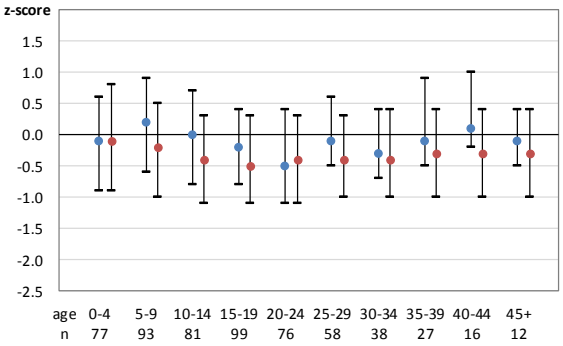


Quartiles of z-scores for height: Croatia

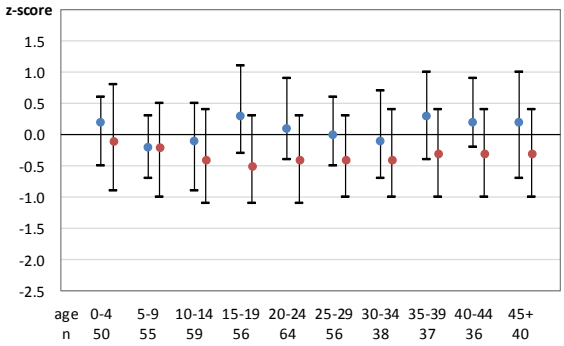


[figure 6.3 continued]

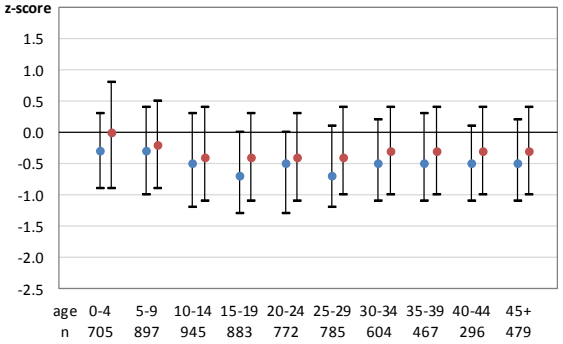
Quartiles of z-scores for height: Czech Republic



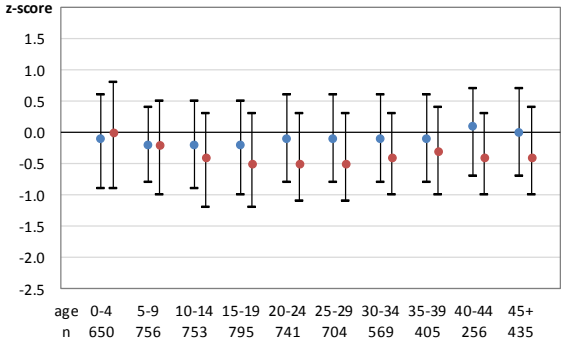
Quartiles of z-scores for height: Denmark



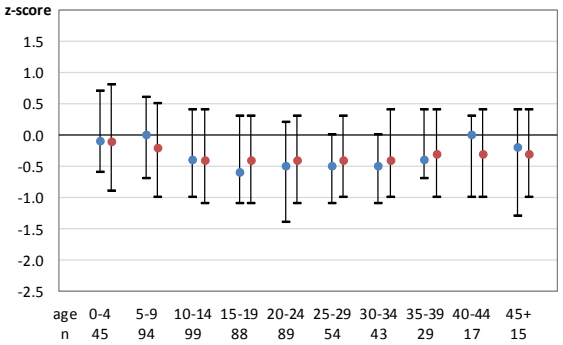
Quartiles of z-scores for height: France



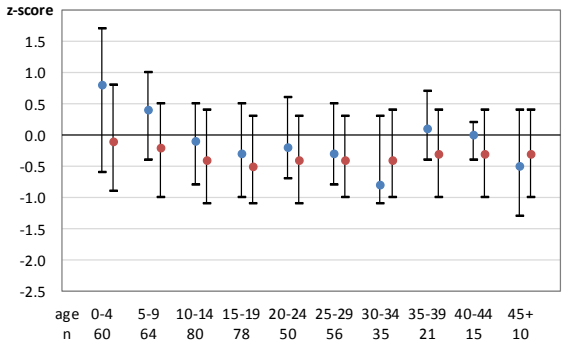
Quartiles of z-scores for height: Germany



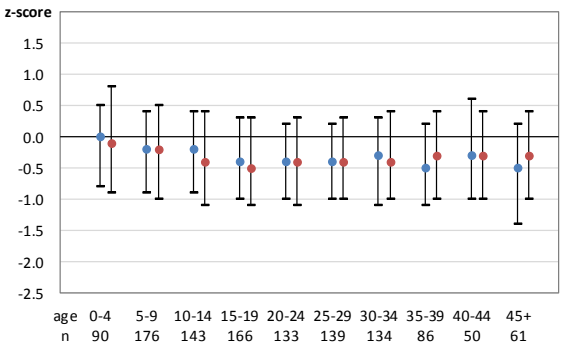
Quartiles of z-scores for height: Greece



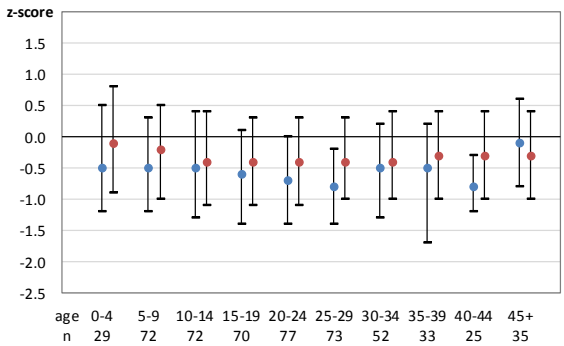
Quartiles of z-scores for height: Hungary



Quartiles of z-scores for height: Ireland

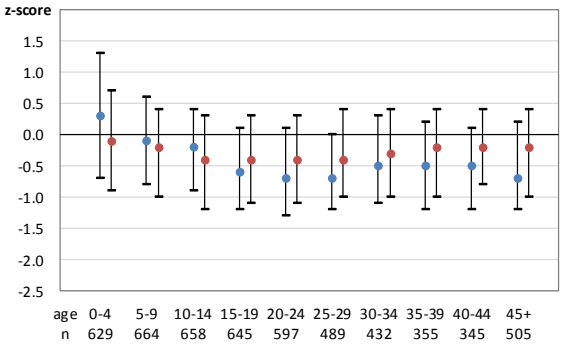


Quartiles of z-scores for height: Israel

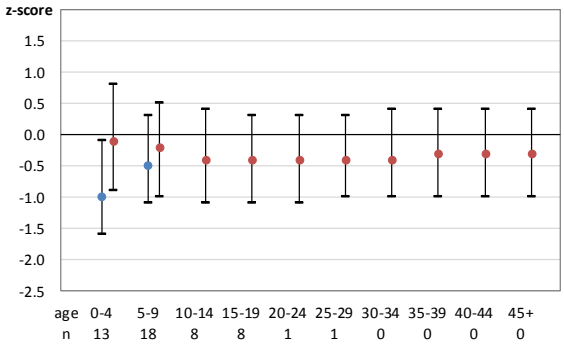


[figure 6.3 continued]

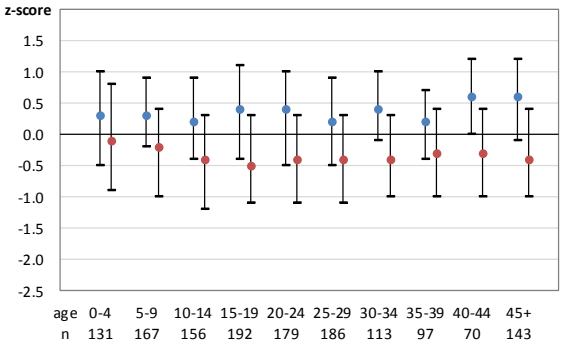
Quartiles of z-scores for height: Italy



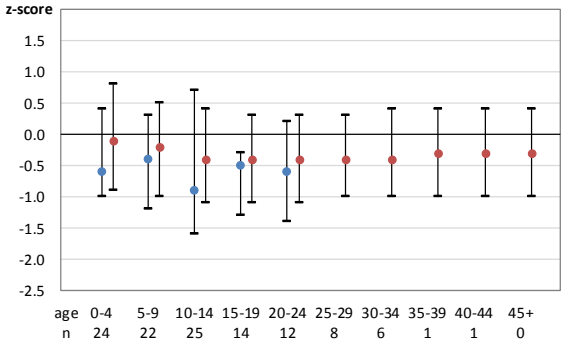
Quartiles of z-scores for height: Rep of Moldova



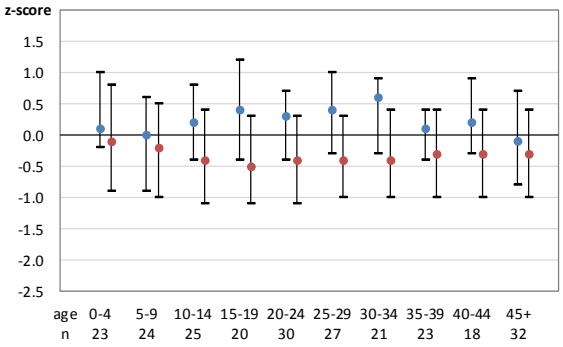
Quartiles of z-scores for height: The Netherlands



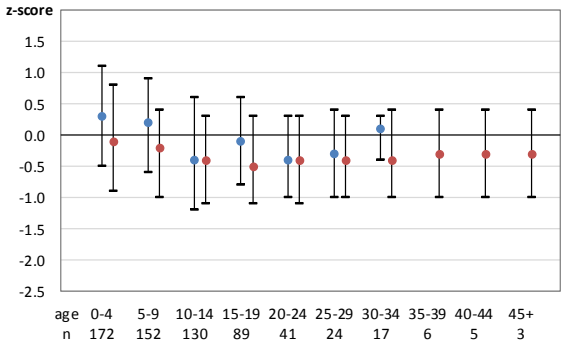
Quartiles of z-scores for height: North Macedonia



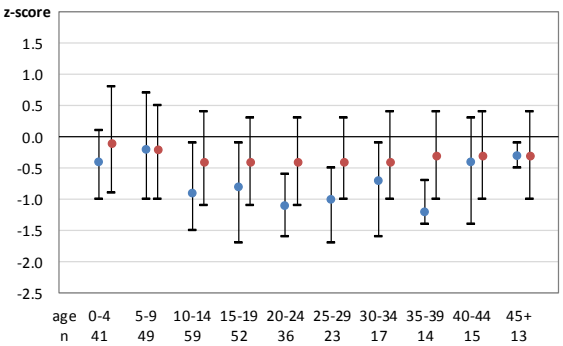
Quartiles of z-scores for height: Norway



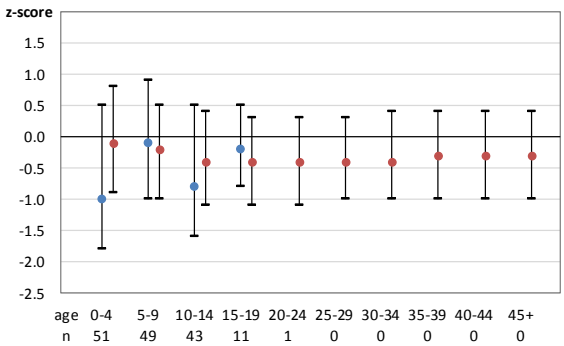
Quartiles of z-scores for height: Poland



Quartiles of z-scores for height: Portugal

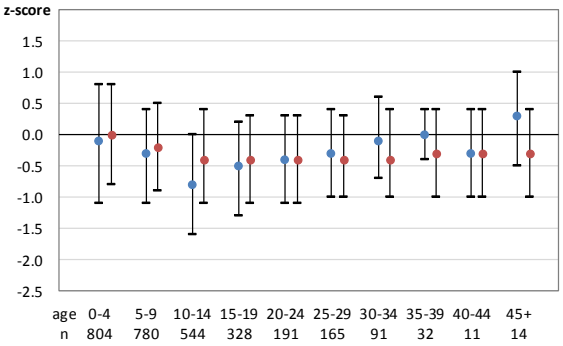


Quartiles of z-scores for height: Romania

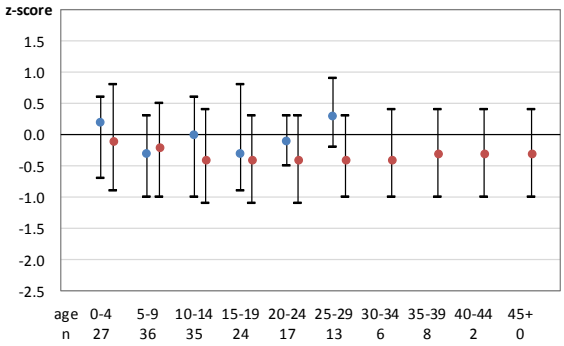


[figure 6.3 continued]

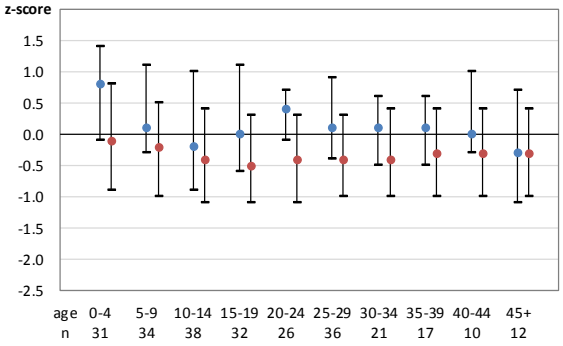
Quartiles of z-scores for height: Russian Federation



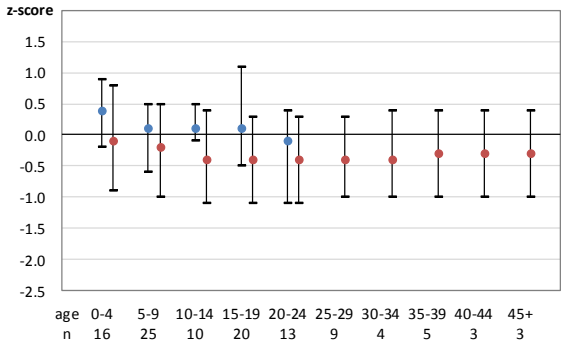
Quartiles of z-scores for height: Serbia



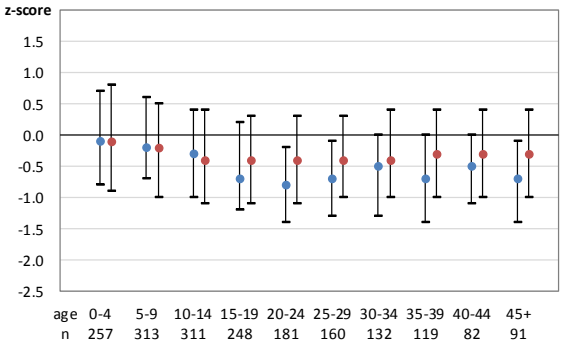
Quartiles of z-scores for height: Slovak Republic



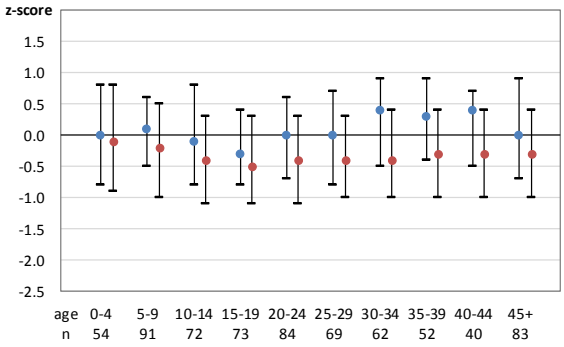
Quartiles of z-scores for height: Slovenia



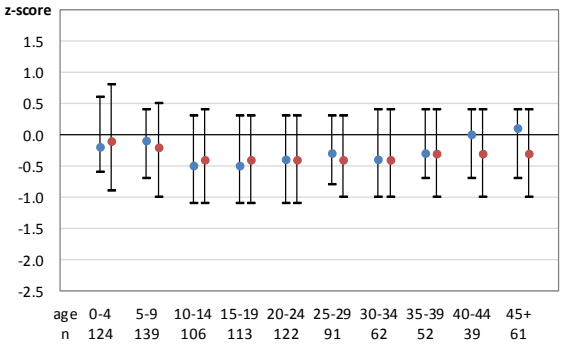
Quartiles of z-scores for height: Spain



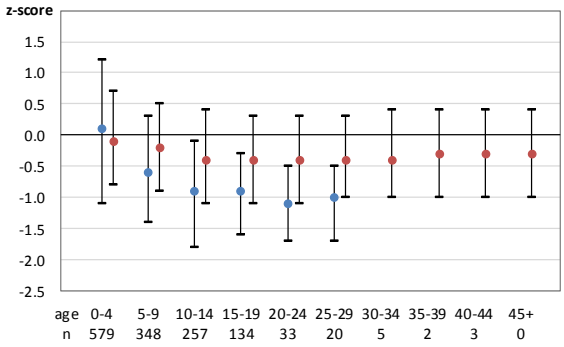
Quartiles of z-scores for height: Sweden



Quartiles of z-scores for height: Switzerland

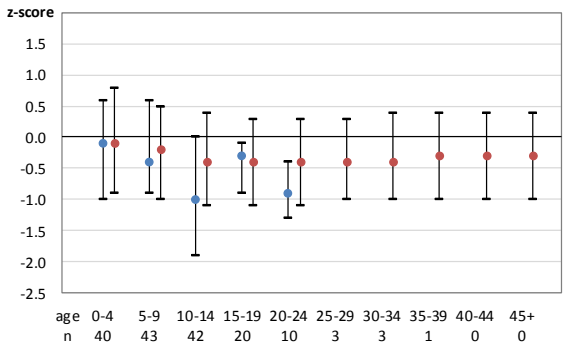


Quartiles of z-scores for height: Turkey



[figure 6.3 continued]

Quartiles of z-scores for height: Ukraine



Quartiles of z-scores for height: United Kingdom

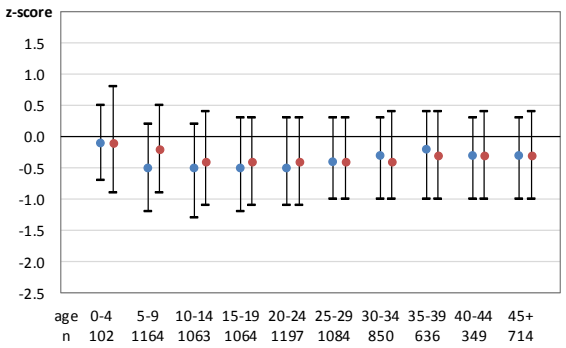


Table 6.5 Z-scores for weight: descriptive statistics by country. Patients aged 17 years or younger.

Country	N	Mean	Min	25th pctl (25% of the patients are below this z-score for weight)	Median (50% of the patients are below this z-score for weight)	75th pctl (75% of the patients are below this z-score for weight)	Max
Albania	58	-0.7	-5.3	-1.4	-0.6	0.2	1.9
Armenia	29	-0.4	-2.8	-1.1	-0.1	0.4	1.5
Austria	376	-0.3	-4.5	-0.9	-0.2	0.4	2.3
Belgium	492	-0.5	-3.7	-1.2	-0.4	0.2	2.6
Bulgaria	70	-1.2	-5.1	-2.0	-0.9	-0.2	1.0
Croatia	50	-0.4	-4.2	-0.8	-0.2	0.3	2.4
Czech Republic	314	-0.3	-4.2	-0.9	-0.2	0.5	3.2
Denmark	195	-0.3	-3.0	-1.0	-0.3	0.4	2.1
France	3129	-0.6	-6.7	-1.2	-0.5	0.1	3.2
Germany	2655	-0.4	-5.8	-1.1	-0.3	0.3	2.7
Greece	293	0.0	-4.2	-0.7	0.1	0.8	2.9
Hungary	261	-0.5	-4.4	-1.2	-0.3	0.4	2.8
Ireland	530	-0.1	-7.2	-0.7	-0.1	0.5	3.0
Israel	222	-0.4	-5.0	-1.2	-0.3	0.3	3.2
Italy	2393	-0.2	-9.4	-0.9	-0.1	0.6	7.6
Latvia	29	-0.4	-2.4	-0.8	-0.4	0.2	1.1
Luxembourg	14	-0.3	-1.4	-1.1	-0.5	0.1	2.0
Rep of Moldova	45	-1.5	-8.4	-2.2	-1.3	-0.3	0.5
The Netherlands	571	0.0	-3.9	-0.6	0.0	0.5	4.3
North Macedonia	82	-0.5	-4.5	-1.4	-0.6	0.6	2.5
Norway[1]	85	-0.1	-2.2	-0.8	-0.1	0.5	3.8
Poland	534	-0.3	-7.1	-0.9	-0.2	0.5	2.6
Portugal	182	-0.6	-5.5	-1.3	-0.5	0.1	2.1
Romania	152	-1.0	-4.7	-1.8	-0.9	-0.2	1.9
Russian Federation	2375	-0.8	-8.5	-1.6	-0.8	0.1	9.6
Serbia	120	-0.5	-3.7	-1.3	-0.5	0.2	3.0
Slovak Republic	127	-0.1	-3.1	-0.9	-0.3	0.7	3.0
Slovenia	68	-0.2	-3.3	-0.6	-0.1	0.4	1.7
Spain	1042	-0.3	-5.9	-0.9	-0.2	0.5	2.8
Sweden	265	-0.3	-3.6	-0.8	-0.1	0.4	2.7
Switzerland	437	-0.3	-4.0	-0.9	-0.3	0.3	2.9
Turkey	1302	-0.8	-9.3	-1.6	-0.6	0.2	7.9
Ukraine	140	-1.0	-4.9	-1.9	-0.9	-0.1	4.0
United Kingdom	2802	-0.3	-5.2	-1.0	-0.3	0.4	2.9

[1] Norway: sometimes any value (instead of last of the year) for weight is used when no lung function test was available.

Note: Lithuania has 0% coverage for children.µ

This table 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 6.6 Z-scores for weight: descriptive statistics by country. Patients aged 18 years or older.

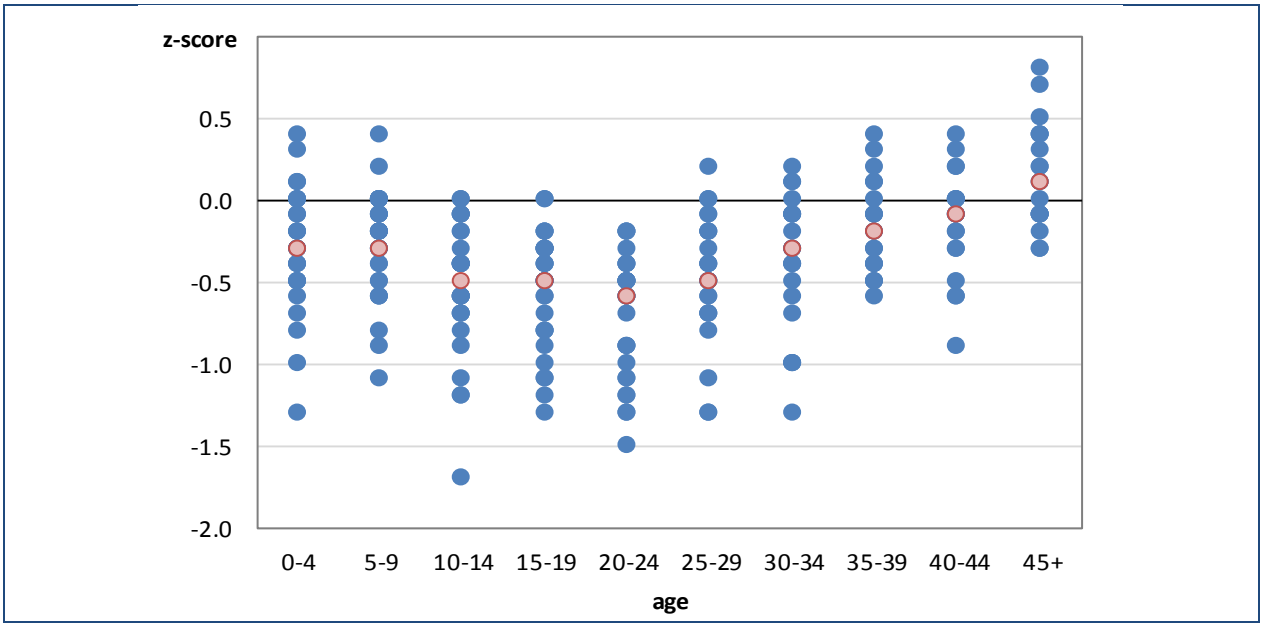
Country	N	Mean	Min	25th pctl (25% of the patients are below this z-score for weight)	Median (50% of the patients are below this z-score for weight)	75th pctl (75% of the patients are below this z-score for weight)	Max
Albania	8	-1.2	-2.4	-1.9	-1.1	-0.6	-0.2
Austria	379	-0.6	-4.3	-1.3	-0.5	0.2	2.3
Belgium	754	-0.5	-4.8	-1.2	-0.4	0.3	2.3
Bulgaria	61	-1.2	-4.6	-1.9	-1.2	-0.4	2.5
Croatia	37	-0.3	-2.3	-0.7	-0.2	0.3	1.6
Czech Republic	263	-0.5	-5.0	-1.3	-0.4	0.3	1.9
Denmark	286	-0.1	-3.0	-0.9	0.0	0.8	2.4
France	3707	-0.8	-7.2	-1.5	-0.7	0.0	3.0
Germany	3403	-0.5	-6.8	-1.2	-0.4	0.4	3.0
Greece	281	-0.4	-5.0	-1.1	-0.4	0.2	2.0
Hungary	209	-0.8	-5.0	-1.7	-0.7	0.0	2.0
Ireland	540	-0.3	-5.5	-0.8	-0.2	0.5	2.8
Israel	316	-0.4	-4.6	-1.2	-0.3	0.5	2.3
Italy	2933	-0.5	-6.4	-1.2	-0.5	0.2	3.1
Latvia	10	-0.5	-1.9	-1.5	-0.5	-0.1	0.9
Lithuania	14	-0.5	-3.0	-0.7	-0.3	0.0	1.2
Luxembourg	22	-0.1	-3.5	-0.9	0.1	0.8	2.1
Rep of Moldova	861	0.0	-3.6	-0.6	0.0	0.6	2.5
The Netherlands	158	0.0	-3.6	-0.6	0.0	0.6	2.8
North Macedonia	31	-0.7	-3.1	-1.5	-0.7	0.1	1.4
Norway[1]	111	-0.7	-3.9	-1.5	-0.6	0.0	2.0
Poland	137	-0.8	-8.6	-1.5	-0.7	0.1	2.3
Portugal	604	-1.3	-5.8	-2.0	-1.1	-0.3	2.3
Russian Federation	49	-0.9	-3.6	-1.5	-0.9	-0.3	1.2
Serbia	132	-0.4	-5.4	-1.0	-0.3	0.4	2.5
Slovak Republic	41	-0.8	-3.7	-1.7	-0.7	0.1	1.6
Slovenia	857	-0.5	-5.8	-1.2	-0.5	0.3	2.7
Spain	412	-0.1	-4.6	-0.7	0.0	0.6	2.9
Sweden	472	-0.6	-5.0	-1.2	-0.5	0.1	2.1
Switzerland	92	-1.5	-5.8	-1.9	-1.3	-0.3	1.3
Turkey	22	-1.5	-3.3	-2.3	-1.3	-0.6	0.2
Ukraine	5154	-0.2	-8.7	-0.9	-0.1	0.6	3.7
United Kingdom	8	-1.2	-2.4	-1.9	-1.1	-0.6	-0.2

[1] Norway: sometimes any value (instead of last of the year) for weight is used when no lung function test was available.

Note: Armenia, Rep of Moldova and Romania have <5 patients aged 18 years or more at weight measurement and are excluded from this table.

This table 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 2017.



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

This graph shows the median z-scores for weight by age group. Each country is represented by a dot (in blue) and the overall estimate is in red. Overall, the median z-scores for weight decrease from the third youngest age group to the 20-24 years age group 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 6.7 Z-scores for weight: descriptive statistics by age group. All patients seen in 2017.

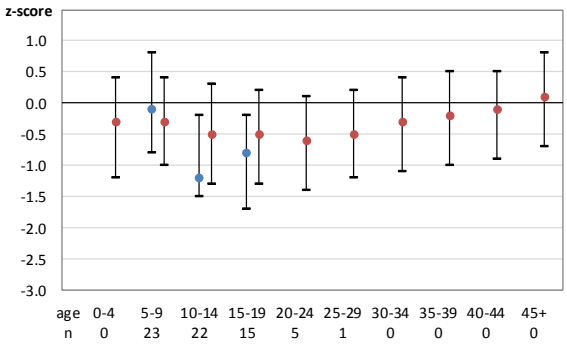
Age at weight measurement	N	Mean	Min	25th pctl	Median	75th pctl	Max
0-4	5144	1156	-0.4	-9.3	-1.2	-0.3	0.4
5-9	6613	306	-0.3	-6.7	-1.0	-0.3	0.4
10-14	6103	194	-0.5	-9.4	-1.3	-0.5	0.3
15-19	5615	195	-0.7	-8.6	-1.3	-0.5	0.2
20-24	5014	156	-0.7	-6.3	-1.4	-0.6	0.1
25-29	4524	184	-0.5	-6.1	-1.2	-0.5	0.2
30-34	3529	167	-0.4	-8.7	-1.1	-0.3	0.4
35-39	2622	120	-0.3	-5.5	-1.0	-0.2	0.5
40-44	1785	69	-0.2	-5.4	-0.9	-0.1	0.5
45+	2856	141	0.0	-6.9	-0.7	0.1	0.8

This table reports the median z-score for weight and other descriptive statistics by age group for all the patients seen in 2017. The median values reported in this table are shown as red dots in fig 6.4.

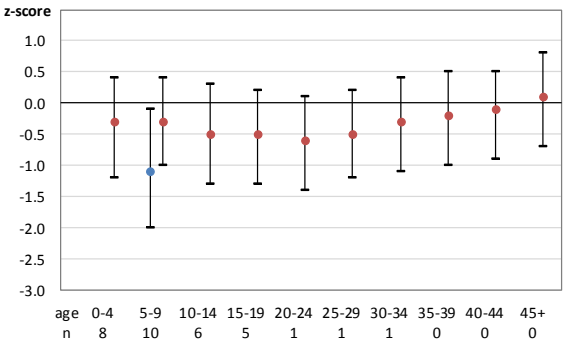
Figure 6.5 Quartiles of z-scores for weight by age group and by country. All patients seen in 2017.

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 Latvia, Lithuania and Luxembourg from the graphs because none of the age groups in these countries had more than 10 patients.

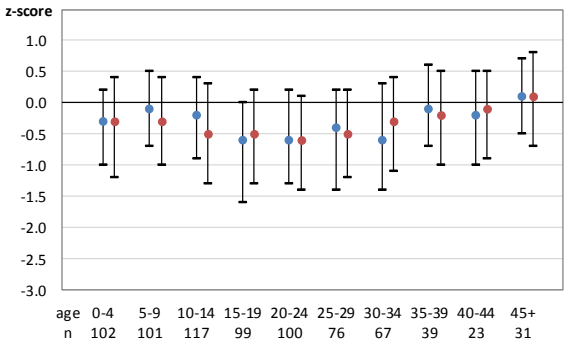
Quartiles of z-scores for weight: Albania



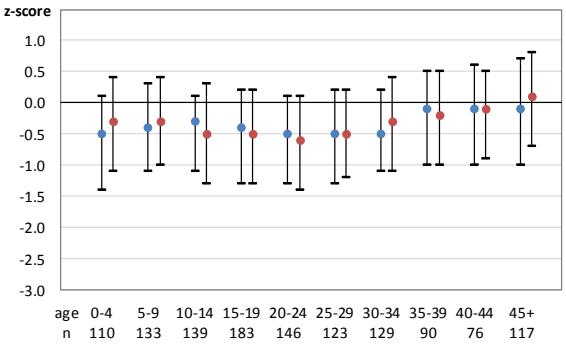
Quartiles of z-scores for weight: Armenia



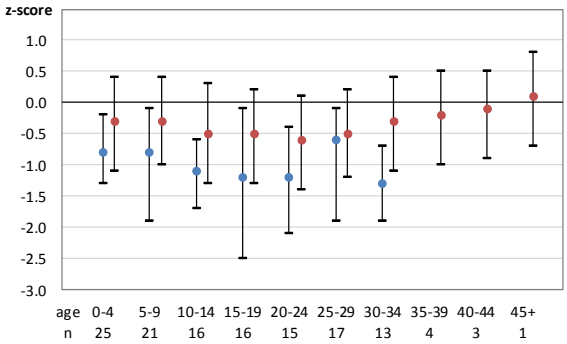
Quartiles of z-scores for weight: Austria



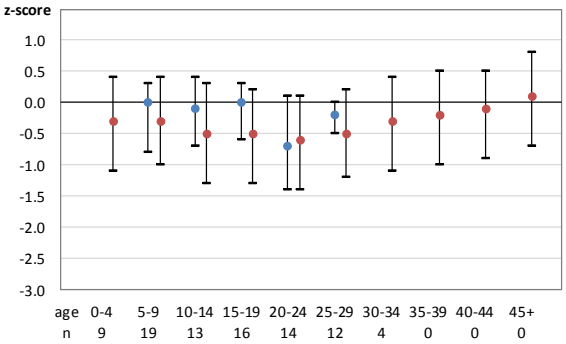
Quartiles of z-scores for weight: Belgium



Quartiles of z-scores for weight: Bulgaria

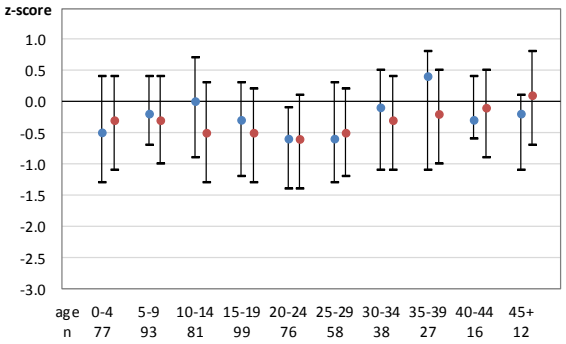


Quartiles of z-scores for weight: Croatia

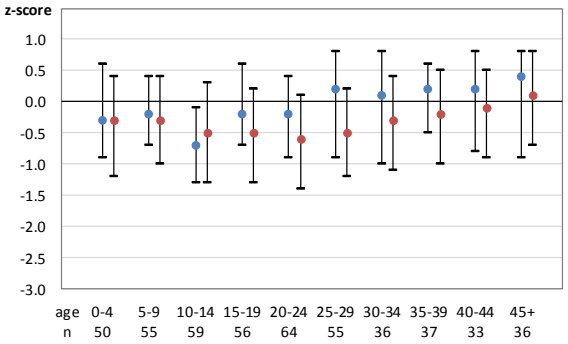


[figure 6.5 continued]

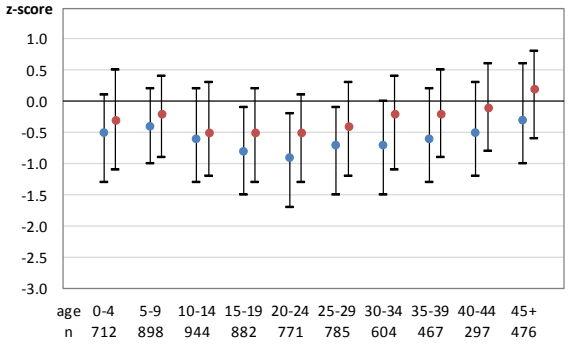
Quartiles of z-scores for weight: Czech Republic



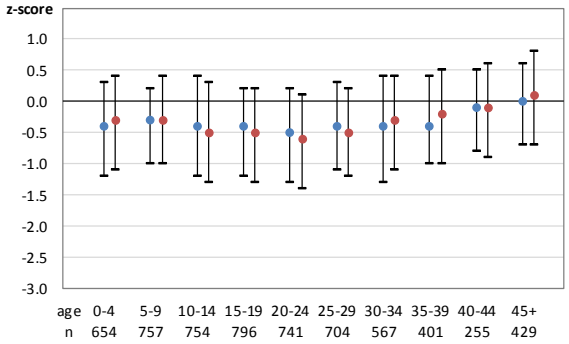
Quartiles of z-scores for weight: Denmark



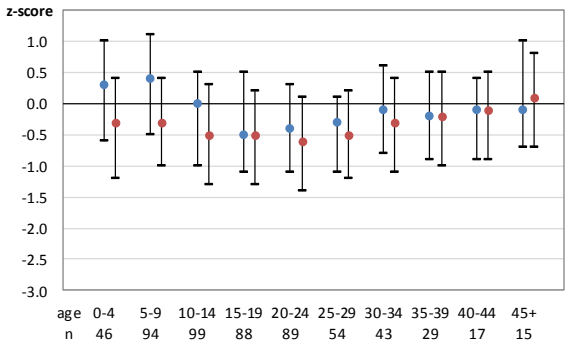
Quartiles of z-scores for weight: France



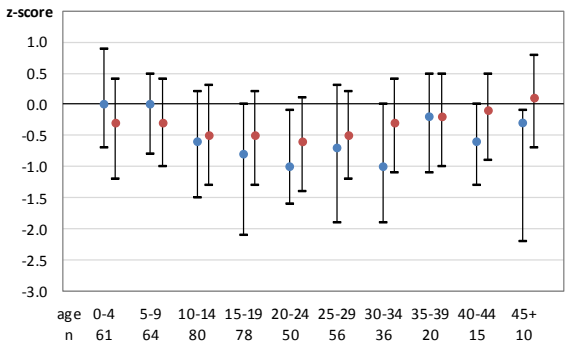
Quartiles of z-scores for weight: Germany



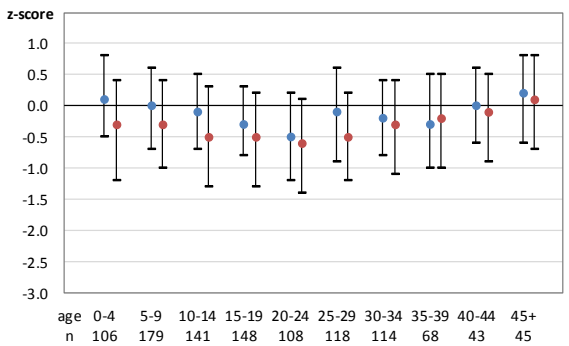
Quartiles of z-scores for weight: Greece



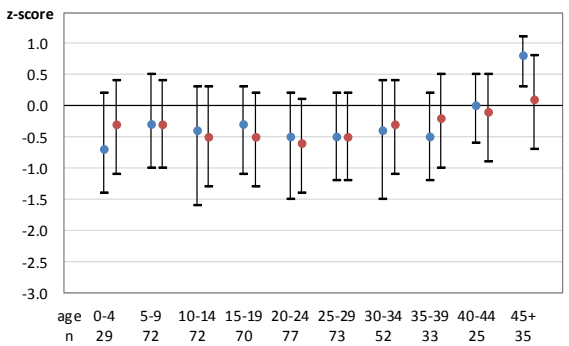
Quartiles of z-scores for weight: Hungary



Quartiles of z-scores for weight: Ireland

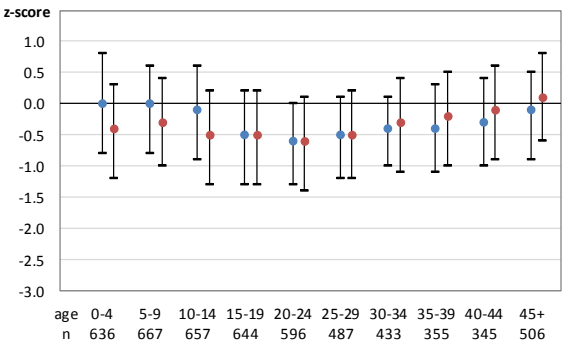


Quartiles of z-scores for weight: Israel

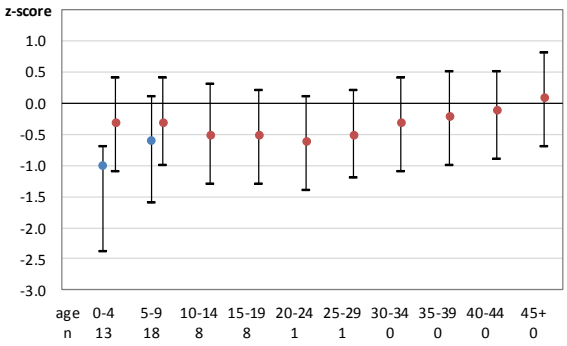


[figure 6.5 continued]

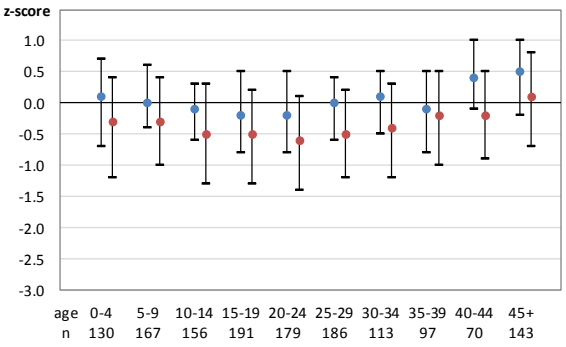
Quartiles of z-scores for weight: Italy



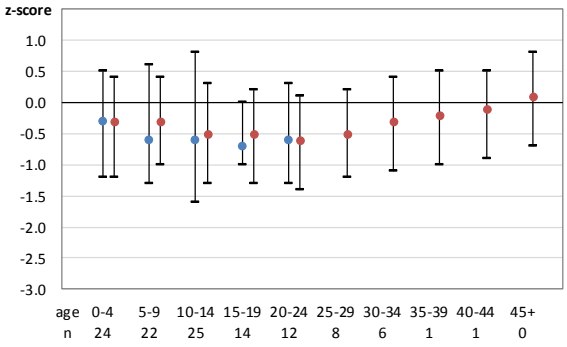
Quartiles of z-scores for weight: Rep of Moldova



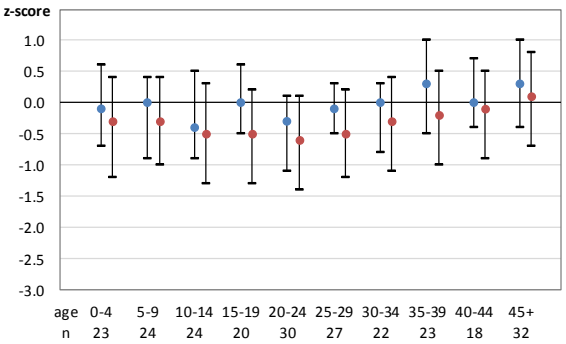
Quartiles of z-scores for weight: The Netherlands



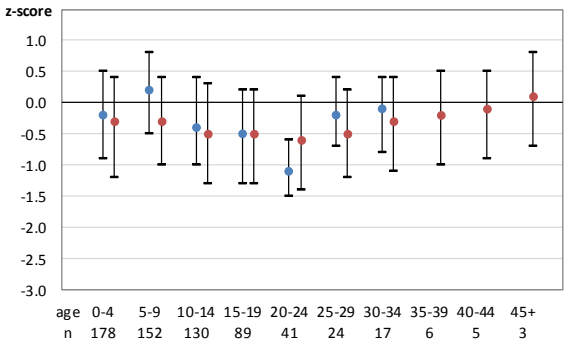
Quartiles of z-scores for weight: North Macedonia



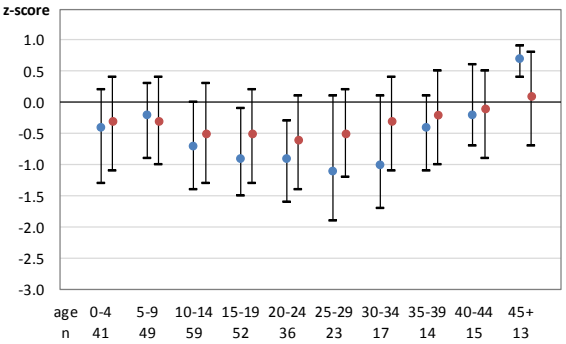
Quartiles of z-scores for weight: Norway



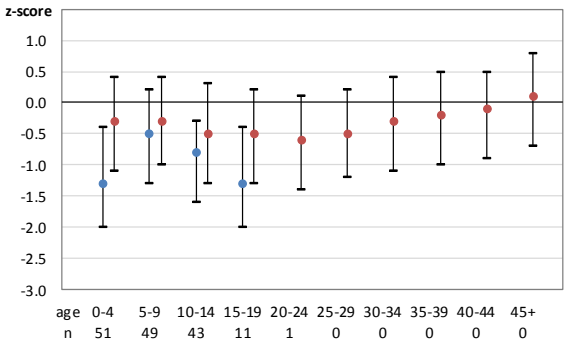
Quartiles of z-scores for weight: Poland



Quartiles of z-scores for weight: Portugal

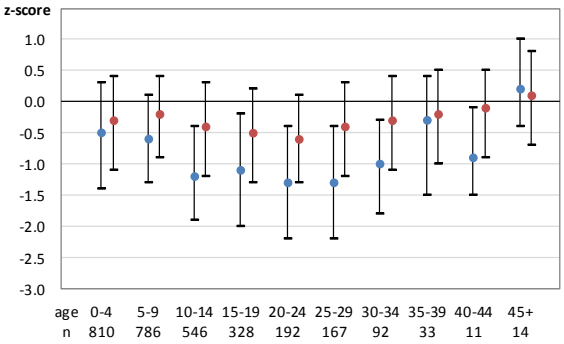


Quartiles of z-scores for weight: Romania

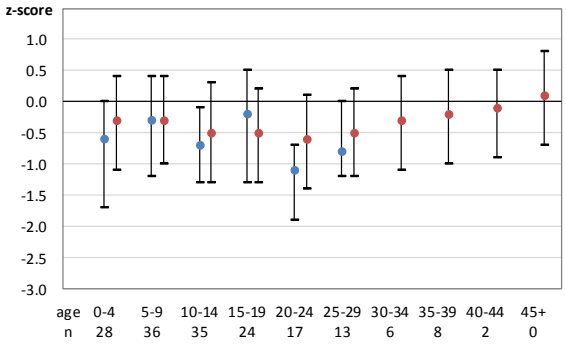


[figure 6.5 continued]

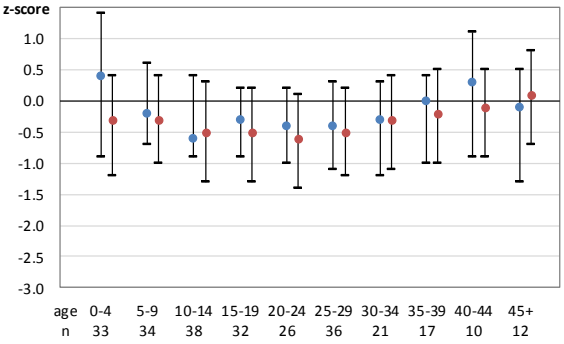
Quartiles of z-scores for weight: Russian Federation



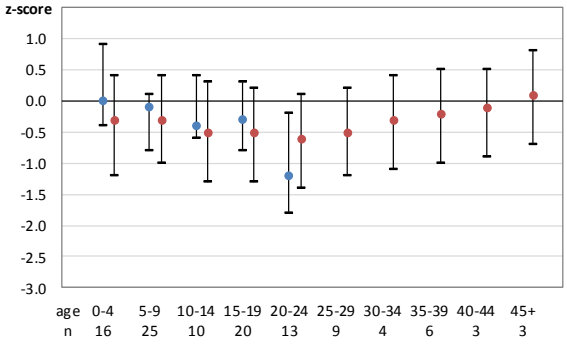
Quartiles of z-scores for weight: Serbia



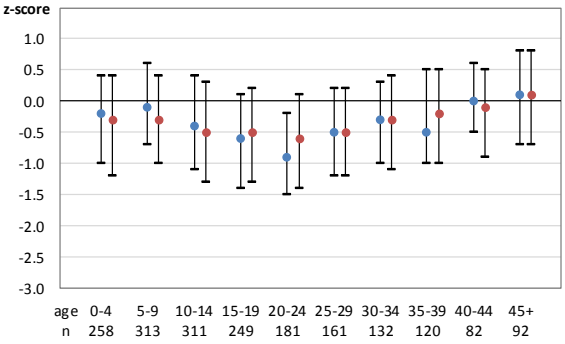
Quartiles of z-scores for weight: Slovak Republic



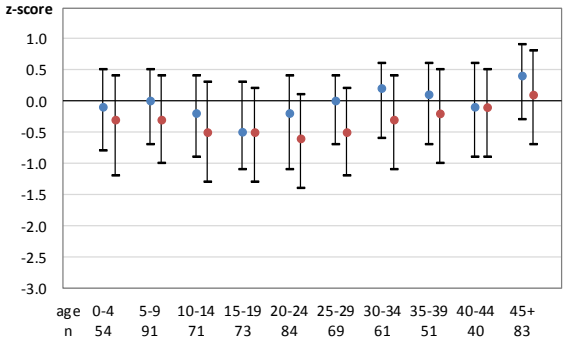
Quartiles of z-scores for weight: Slovenia



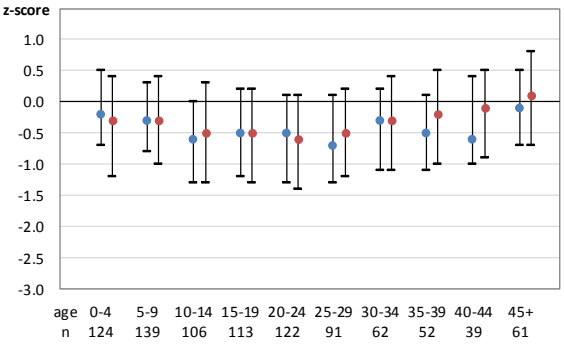
Quartiles of z-scores for weight: Spain



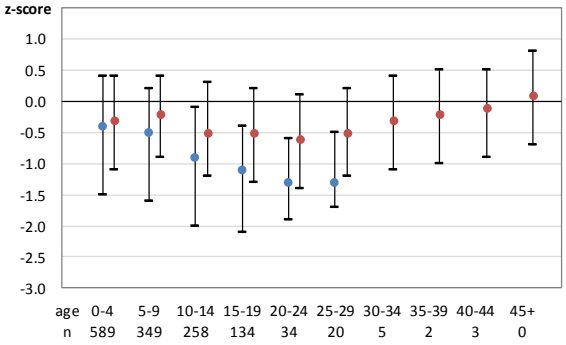
Quartiles of z-scores for weight: Sweden



Quartiles of z-scores for weight: Switzerland

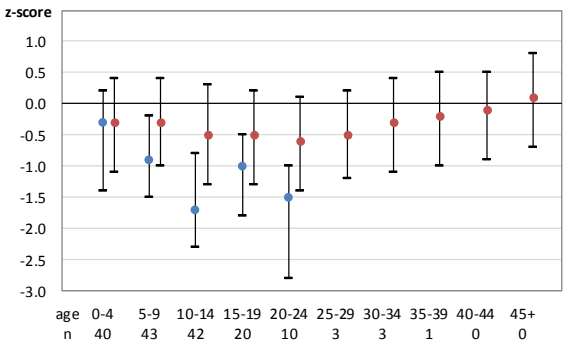


Quartiles of z-scores for weight: Turkey



[figure 6.5 continued]

Quartiles of z-scores for weight: Ukraine



Quartiles of z-scores for weight: United Kingdom

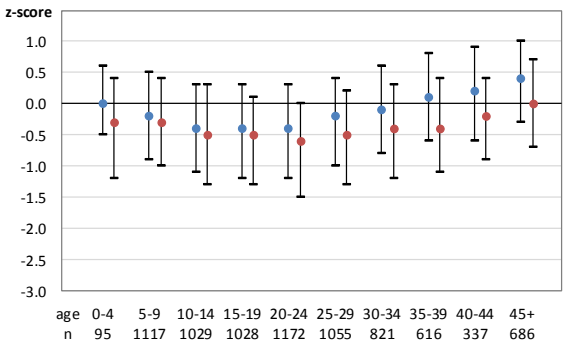


Table 6.8 Z-scores for BMI: descriptive statistics by country. All patients seen in 2017 aged 2-17 years.

Country	N	N Miss	Mean	Min	25th pctl (25% of the patients are below this z-score for BMI)	Median (50% of the patients are below this z-score for BMI)	75th pctl (75% of the patients are below this z-score for BMI)	Max
Albania	58	0	-0.7	-7.3	-1.7	-0.2	0.5	1.9
Armenia	25	0	-0.6	-5.1	-0.9	-0.3	0.4	1.5
Austria	338	0	-0.4	-5.0	-1.0	-0.2	0.3	2.4
Belgium	453	12	-0.3	-4.1	-0.9	-0.2	0.4	2.5
Bulgaria	60	1	-1.0	-5.3	-1.9	-0.5	0.0	2.0
Croatia	47	0	-0.6	-5.0	-1.0	-0.3	0.2	2.0
Czech Republic	278	11	-0.3	-3.8	-1.1	-0.2	0.5	2.7
Denmark	172	0	-0.4	-2.8	-1.2	-0.3	0.3	2.1
France	2855	38	-0.4	-4.4	-1.0	-0.4	0.2	3.8
Germany	2396	10	-0.4	-5.1	-1.0	-0.3	0.3	2.4
Greece	280	1	0.1	-3.4	-0.6	0.3	0.9	2.6
Hungary	243	9	-0.7	-5.0	-1.4	-0.6	0.1	2.2
Ireland	497	21	0.1	-5.4	-0.5	0.1	0.6	2.9
Israel	214	0	-0.1	-3.3	-0.7	-0.1	0.5	2.4
Italy	2127	53	-0.1	-5.8	-0.8	-0.1	0.6	3.0
Latvia	26	0	-0.7	-2.6	-1.2	-0.7	-0.2	0.9
Luxembourg	11	1	-0.4	-1.4	-0.8	-0.5	0.0	0.5
Rep of Moldova	37	0	-1.1	-3.9	-1.7	-0.9	-0.3	0.6
The Netherlands	520	14	-0.2	-3.5	-0.8	-0.2	0.4	2.4
North Macedonia	73	0	-0.2	-3.2	-0.9	-0.5	0.6	2.4
Norway[1]	76	1	-0.3	-2.4	-0.9	-0.2	0.4	2.2
Poland	461	0	-0.3	-4.6	-1.0	-0.3	0.5	3.7
Portugal	167	0	-0.4	-4.2	-1.1	-0.3	0.4	2.2
Romania	134	1	-0.9	-5.6	-1.8	-0.9	0.2	3.2
Russian Federation	2062	35	-0.8	-8.8	-1.5	-0.7	0.1	2.8
Serbia	109	0	-0.4	-3.3	-1.0	-0.3	0.4	2.5
Slovak Republic	115	2	-0.4	-3.4	-1.1	-0.4	0.4	2.9
Slovenia	65	0	-0.5	-4.8	-0.9	-0.2	0.3	1.7
Spain	962	2	-0.2	-3.8	-0.8	-0.1	0.5	2.6
Sweden	245	2	-0.3	-4.2	-0.8	-0.3	0.3	2.3
Switzerland	394	0	-0.3	-3.0	-0.9	-0.2	0.4	2.5
Turkey	1025	15	-0.7	-8.7	-1.5	-0.5	0.3	3.5
Ukraine	130	0	-1.0	-5.1	-1.9	-1.0	-0.2	4.5
United Kingdom	2799	1158	0.0	-8.9	-0.6	0.0	0.6	2.8

[1] Norway: sometimes any value (instead of last of the year) for height and weight is used when no lung function test was available.

Note: Lithuania has 0% coverage for children.

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 6.9 BMI: descriptive statistics by country. All patients seen in 2017 aged 18 years or older.

Country	N	N Miss	Mean	Min	25th pctl (25% of the patients are below this BMI)	Median (50% of the patients are below this BMI)	75th pctl (75% of the patients are below this BMI)	Max
Albania	8	0	20.1	17.3	19.1	20.0	20.8	23.5
Austria	379	0	21.4	13.6	19.3	20.9	23.1	34.4
Belgium	754	20	21.8	14.4	19.4	21.4	23.4	35.2
Bulgaria	61	2	19.9	13.7	17.3	19.4	21.6	40.1
Croatia	37	0	21.6	17.4	19.8	21.1	22.9	30.5
Czech Republic	263	8	21.3	12.9	19.0	20.8	23.3	32.4
Denmark	286	10	22.4	13.6	19.8	21.8	24.5	37.0
France	3704	45	21.3	12.9	19.1	20.8	23.0	47.3
Germany	3402	41	21.5	12.7	19.2	21.1	23.2	46.1
Greece	281	9	22.1	15.8	20.1	22.0	23.9	35.5
Hungary	207	25	20.5	13.8	18.1	20.1	22.4	30.0
Ireland	539	120	22.7	14.7	20.3	22.3	24.6	42.6
Israel	316	0	22.8	15.3	20.1	22.3	25.1	37.4
Italy	2925	172	22.1	13.9	19.8	21.7	23.9	45.7
Latvia	10	0	20.0	15.9	18.5	19.3	21.8	24.3
Lithuania	14	0	19.6	15.3	18.0	19.6	20.7	24.4
Luxembourg	22	0	23.2	17.3	21.4	22.7	25.2	36.1
The Netherlands	861	20	22.2	15.5	20.2	21.7	23.7	43.5
North Macedonia	31	0	21.3	15.8	19.4	20.9	23.5	27.2
Norway[1]	157	1	22.5	14.8	20.2	21.7	24.2	36.6
Poland	111	0	20.8	15.5	18.3	20.1	22.3	35.4
Portugal	137	1	22.0	12.4	19.6	21.3	23.7	37.3
Russian Federation	598	59	19.6	13.3	17.3	19.1	21.5	34.0
Serbia	49	0	19.7	14.7	18.2	19.6	21.0	26.3
Slovak Republic	132	4	21.4	14.2	19.1	21.2	23.4	34.9
Slovenia	40	1	20.4	16.4	18.2	20.1	21.7	27.5
Spain	852	8	22.3	15.0	20.0	21.9	24.2	40.0
Sweden	412	4	22.4	14.4	20.1	22.0	24.2	41.4
Switzerland	472	0	21.3	14.5	19.3	21.0	23.1	35.3
Turkey	91	3	20.2	12.8	18.2	20.0	22.2	30.8
Ukraine	22	0	19.2	15.8	17.7	19.5	20.1	22.8
United Kingdom	5153	387	23.0	12.3	20.4	22.4	25.0	49.7

[1] Norway: sometimes any value (instead of last of the year) for height and weight is used when no lung function test was available.

Note: Armenia, Rep of Moldova and Romania have <5 patients aged 18 years or more at height measurement and are excluded from this table.

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

Table 6.10 BMI: descriptive statistics by country. All male patients seen in 2017 aged 18 years or older.

Country	N	N Miss	Mean	Min	25th pctl (25% of the patients are below this BMI)	Median (50% of the patients are below this BMI)	75th pctl (75% of the patients are below this BMI)	Max
Austria	191	0	21.9	14.5	19.4	21.5	23.9	31.8
Belgium	397	8	22.0	15.1	19.7	22.0	23.7	34.3
Bulgaria	33	1	20.8	14.9	18.2	20.0	22.6	40.1
Croatia	14	0	21.8	17.7	20.4	21.3	22.9	27.8
Czech Republic	126	5	21.6	14.5	19.0	21.3	24.2	32.4
Denmark	154	6	22.8	14.3	20.5	22.1	24.9	36.6
France	1947	30	21.5	13.3	19.3	21.1	23.3	47.3
Germany	1815	22	21.9	13.3	19.6	21.6	23.9	46.1
Greece	150	4	22.8	16.6	20.5	22.5	24.6	32.0
Hungary	119	16	21.1	13.8	18.7	20.9	23.1	29.4
Ireland	326	77	23.2	15.6	21.1	22.8	25.3	35.5
Israel	180	0	23.0	15.3	20.5	22.7	25.3	35.0
Italy	1557	92	22.6	14.3	20.4	22.4	24.4	39.6
Lithuania	9	0	20.5	17.9	19.4	20.2	20.7	24.4
Luxembourg	12	0	22.6	17.3	22.0	22.7	23.3	25.6
The Netherlands	474	16	22.3	15.5	20.4	22.0	23.9	32.7
North Macedonia	19	0	22.0	17.4	20.4	22.0	24.2	27.2
Norway[1]	86	0	23.3	17.1	20.3	22.6	24.8	36.6
Poland	46	0	21.2	16.3	17.7	20.9	23.1	35.4
Portugal	72	1	21.7	12.8	19.2	20.9	23.6	30.9
Russian Federation	312	34	20.0	14.2	17.6	19.4	22.0	34.0
Serbia	30	0	19.8	14.7	18.3	19.8	21.0	26.3
Slovak Republic	67	0	22.1	14.2	19.8	22.1	24.2	34.9
Slovenia	17	1	21.5	17.9	19.3	20.5	22.9	27.5
Spain	461	4	22.9	15.0	20.8	22.6	24.6	40.0
Sweden	227	1	23.1	14.4	20.8	22.6	24.9	37.2
Switzerland	266	0	21.8	14.5	20.0	21.8	23.5	33.4
Turkey	51	0	20.1	12.8	18.0	19.9	23.1	28.4
Ukraine	15	0	19.4	15.8	18.3	19.7	21.0	22.8
United Kingdom	2795	233	23.3	12.6	20.8	23.0	25.4	49.7

[1] Norway: sometimes any value (instead of last of the year) for height and weight is used when no lung function test was available.

Note: Albania, Armenia, Latvia, Rep of Moldova and Romania have <5 male patients aged 18 years or more at BMI measurement and are excluded from this table.

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

Table 6.11 BMI: descriptive statistics by country. All female patients seen in 2017 aged 18 years or older.

Country	N	N Miss	Mean	Min	25th pctl (25% of the patients are below this BMI)	Median (50% of the patients are below this BMI)	75th pctl (75% of the patients are below this BMI)	Max
Austria	188	0	20.8	13.6	19.1	20.4	22.3	34.4
Belgium	357	12	21.4	14.4	19.2	20.8	22.9	35.2
Bulgaria	28	1	18.8	13.7	17.2	18.2	20.4	27.7
Croatia	23	0	21.5	17.4	19.6	21.1	22.9	30.5
Czech Republic	137	3	21.0	12.9	19.1	20.7	22.7	32.3
Denmark	132	4	21.9	13.6	19.4	20.9	24.0	37.0
France	1757	15	21.1	12.9	18.8	20.5	22.5	45.1
Germany	1587	19	21.0	12.7	18.9	20.5	22.6	40.6
Greece	131	5	21.4	15.8	19.6	21.3	22.7	35.5
Hungary	88	9	19.7	14.4	17.8	19.4	21.3	30.0
Ireland	213	43	21.9	14.7	19.8	21.4	23.3	42.6
Israel	136	0	22.4	15.6	19.5	21.9	24.7	37.4
Italy	1368	80	21.6	13.9	19.3	21.0	23.1	45.7
Latvia	6	0	18.5	15.9	17.4	18.6	19.5	21.2
Lithuania	5	0	18.0	15.3	15.8	18.1	18.9	21.8
Luxembourg	10	0	23.9	17.6	20.4	22.5	25.8	36.1
The Netherlands	387	4	22.0	16.2	19.9	21.4	23.3	43.5
North Macedonia	12	0	20.2	15.8	19.1	19.9	22.1	23.8
Norway[1]	71	1	21.7	14.8	19.4	21.0	23.0	34.2
Poland	65	0	20.5	15.5	18.6	20.0	21.6	31.2
Portugal	65	0	22.3	12.4	20.1	21.8	23.8	37.3
Russian Federation	286	25	19.1	13.3	17.1	18.7	21.0	33.9
Serbia	19	0	19.5	15.3	17.3	19.4	21.2	24.7
Slovak Republic	65	4	20.7	14.4	18.8	20.2	22.6	30.1
Slovenia	23	0	19.6	16.4	17.6	19.0	21.4	27.4
Spain	391	4	21.7	15.0	19.4	21.1	23.5	39.0
Sweden	185	3	21.6	14.5	19.3	21.0	23.3	41.4
Switzerland	206	0	20.7	14.7	18.8	20.2	22.1	35.3
Turkey	40	3	20.4	14.0	19.1	20.2	22.1	30.8
Ukraine	7	0	18.8	16.4	17.7	19.0	20.1	21.2
United Kingdom	2358	154	22.6	12.3	19.9	21.8	24.2	48.2

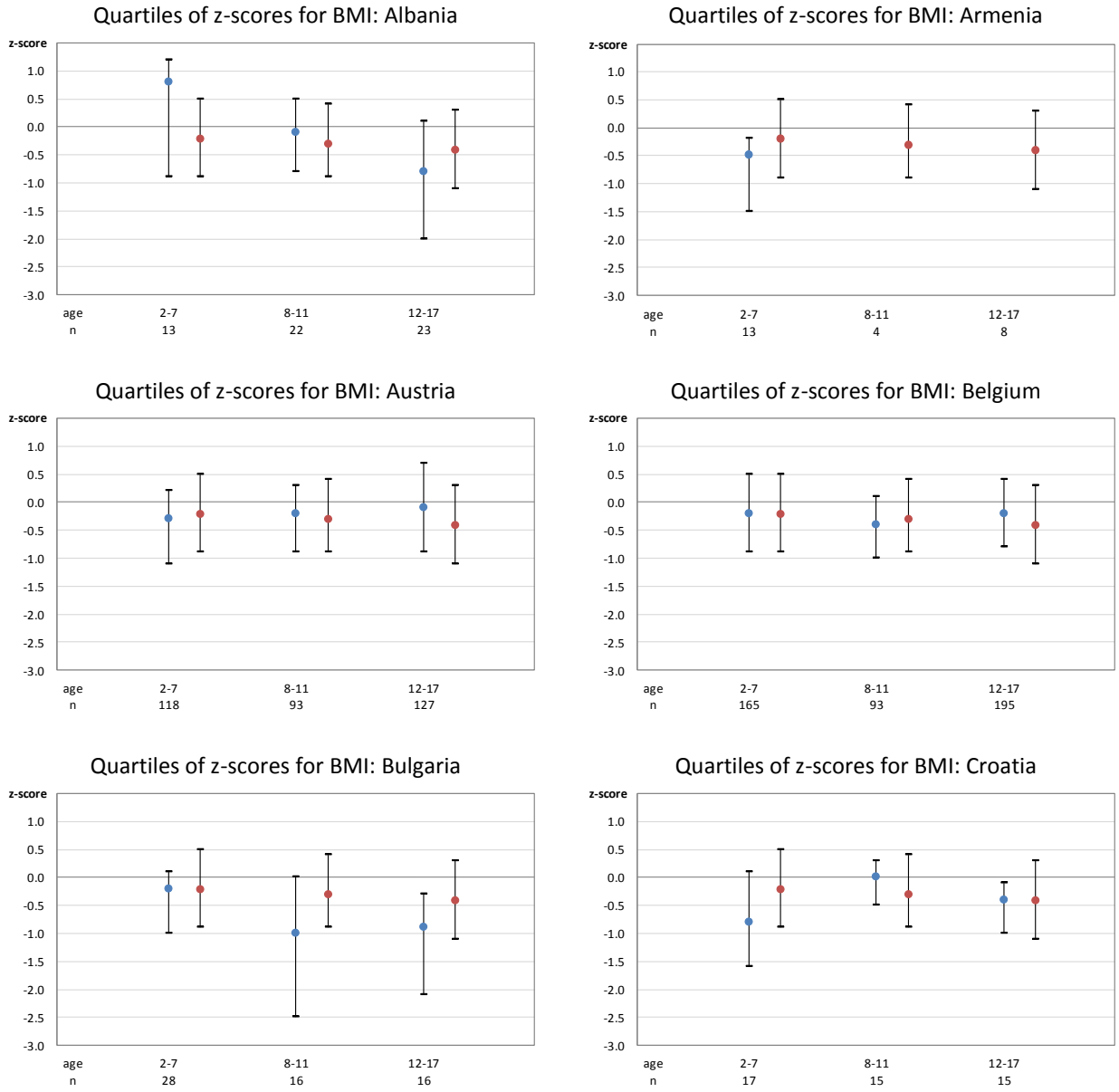
[1] Norway: sometimes any value (instead of last of the year) for height and weight is used when no lung function test was available.

Note: Albania, Armenia, Rep of Moldova and Romania have <5 male patients aged 18 years or more at BMI measurement and are excluded from this table.

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

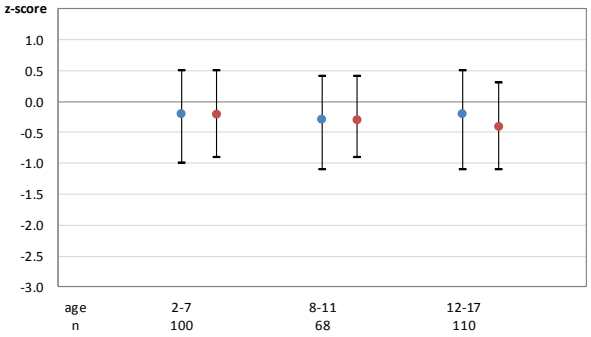
Figure 6.6 Quartiles of z-scores for BMI by age group and country. Patients aged 2-17 years in 2017.

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.

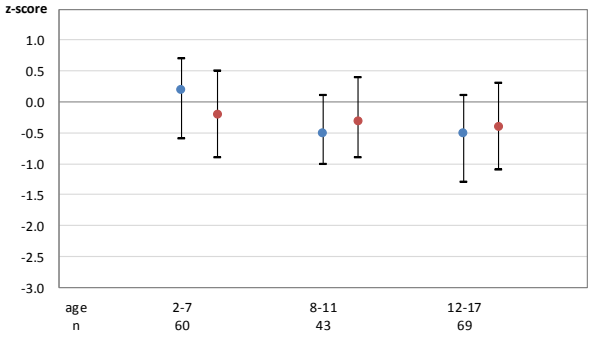


[figure 6.6 continued]

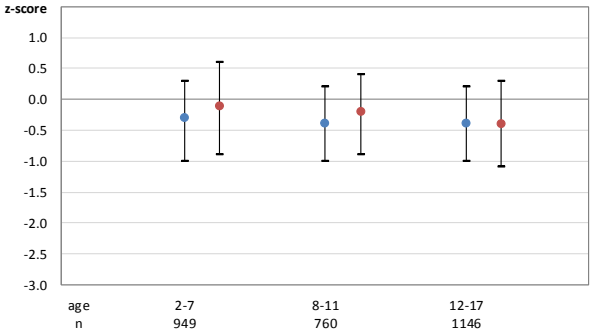
Quartiles of z-scores for BMI: Czech Republic



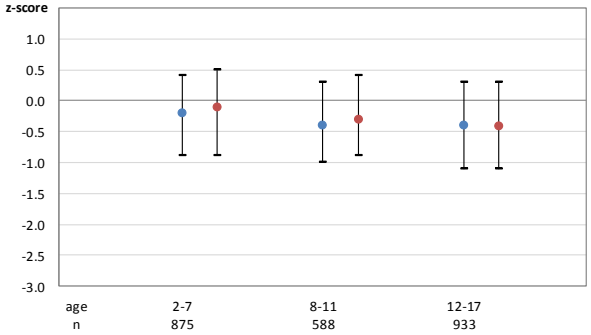
Quartiles of z-scores for BMI: Denmark



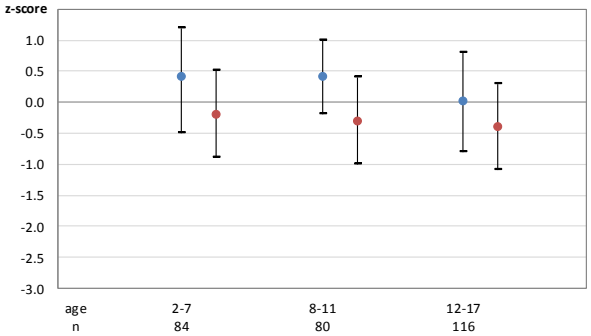
Quartiles of z-scores for BMI: France



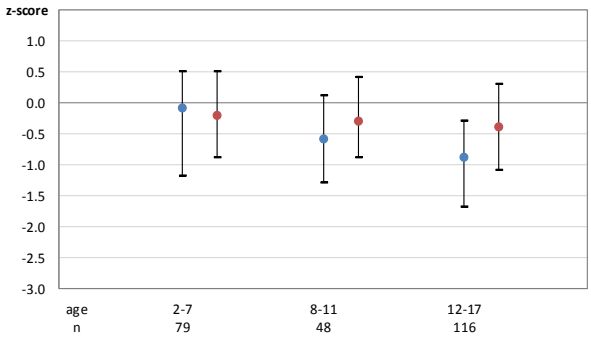
Quartiles of z-scores for BMI: Germany



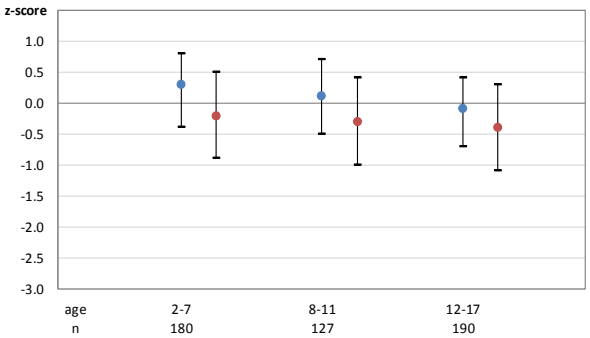
Quartiles of z-scores for BMI: Greece



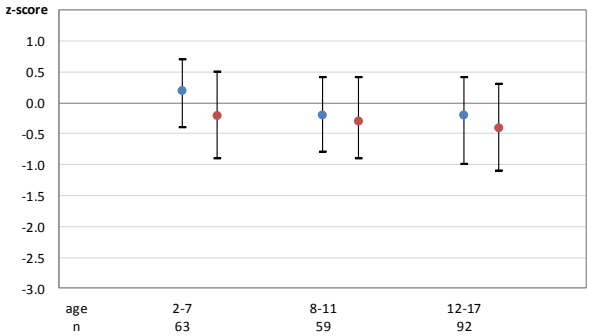
Quartiles of z-scores for BMI: Hungary



Quartiles of z-scores for BMI: Ireland

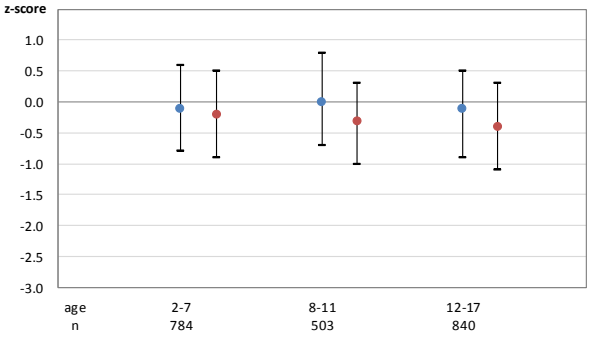


Quartiles of z-scores for BMI: Israel

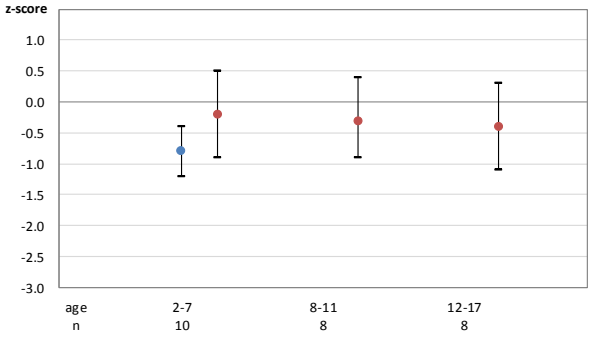


[figure 6.6 continued]

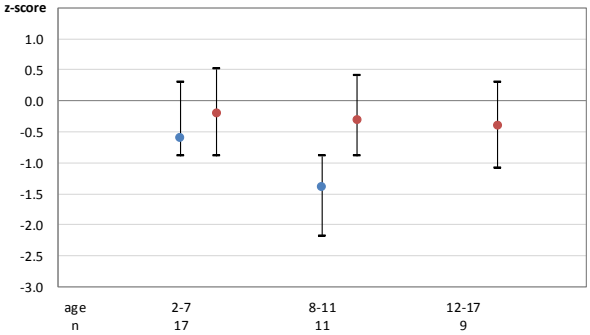
Quartiles of z-scores for BMI: Italy



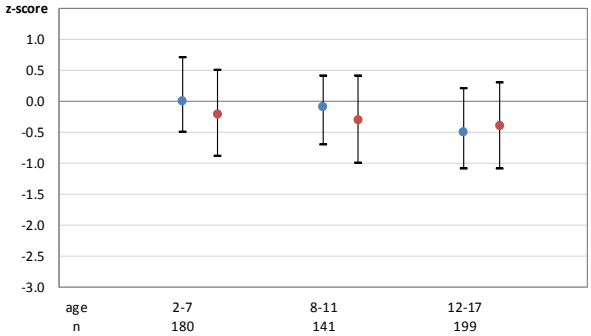
Quartiles of z-scores for BMI: Latvia



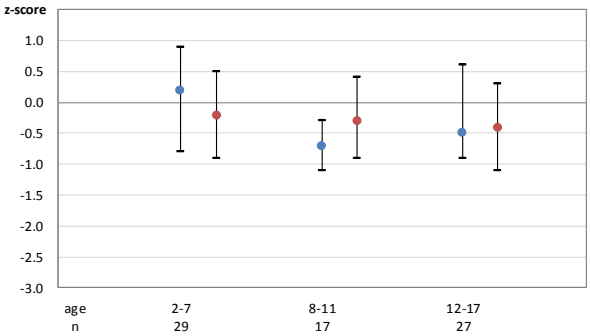
Quartiles of z-scores for BMI: Rep of Moldova



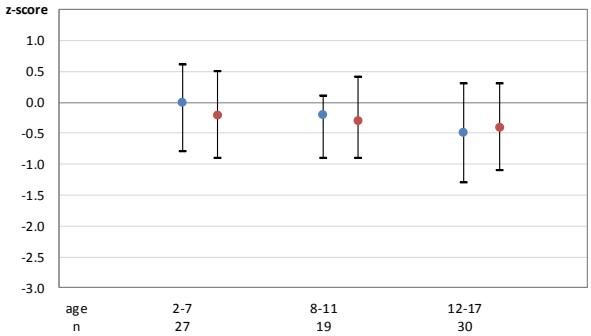
Quartiles of z-scores for BMI: The Netherlands



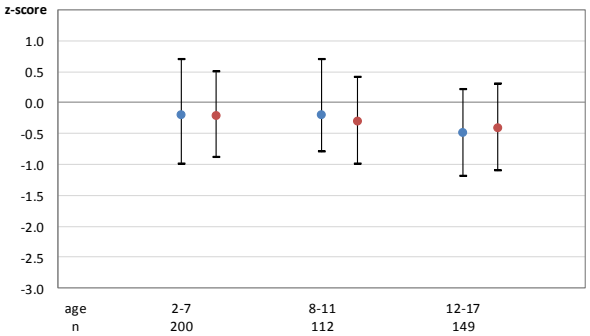
Quartiles of z-scores for BMI: North Macedonia



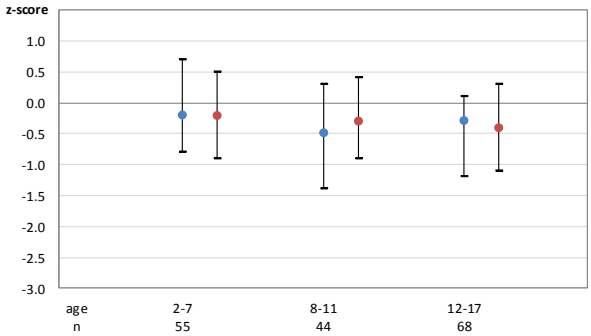
Quartiles of z-scores for BMI: Norway



Quartiles of z-scores for BMI: Poland

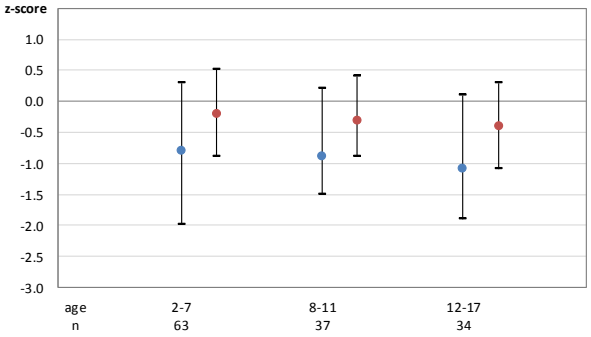


Quartiles of z-scores for BMI: Portugal

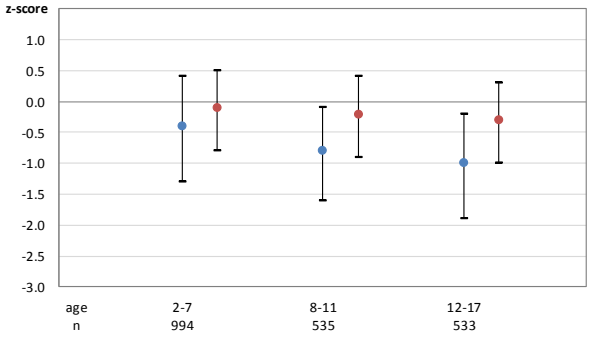


[figure 6.6 continued]

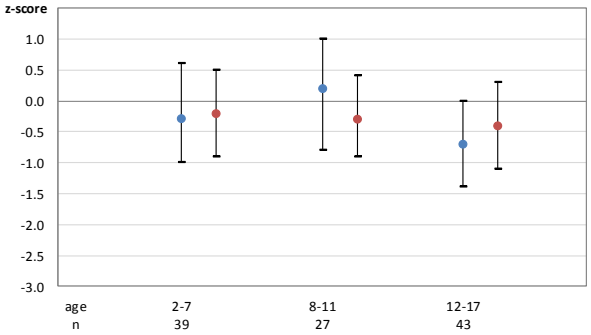
Quartiles of z-scores for BMI: Romania



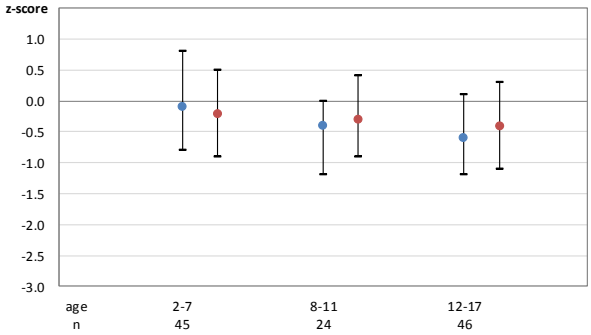
Quartiles of z-scores for BMI: Russian Federation



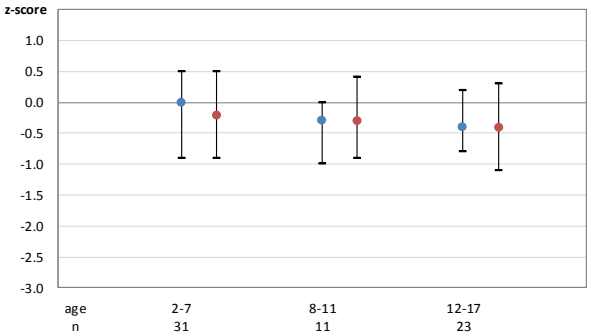
Quartiles of z-scores for BMI: Serbia



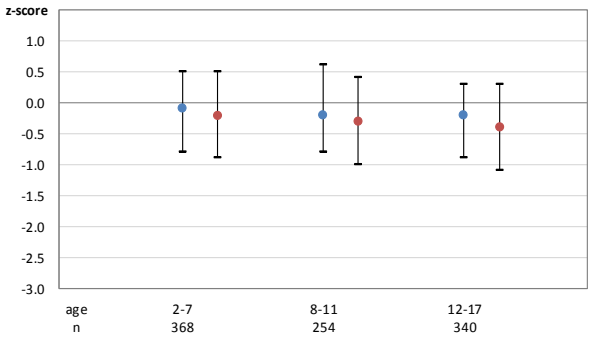
Quartiles of z-scores for BMI: Slovak Republic



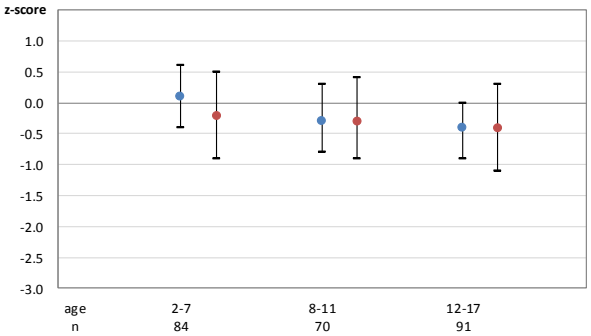
Quartiles of z-scores for BMI: Slovenia



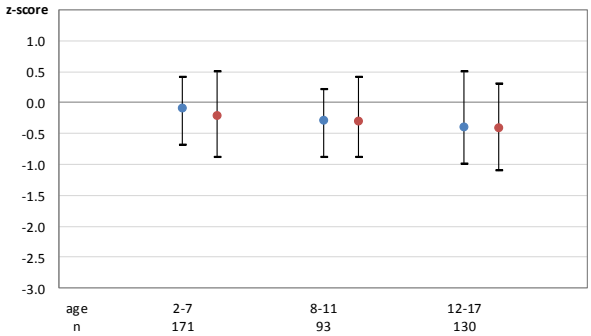
Quartiles of z-scores for BMI: Spain



Quartiles of z-scores for BMI: Sweden

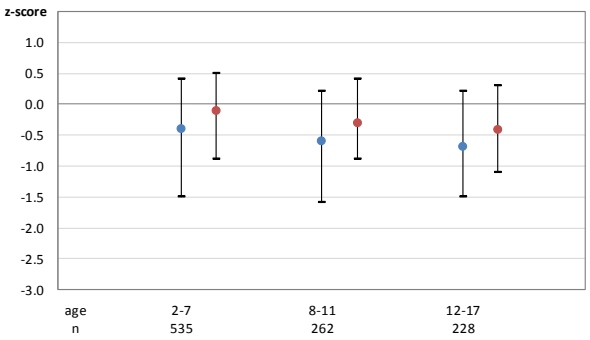


Quartiles of z-scores for BMI: Switzerland

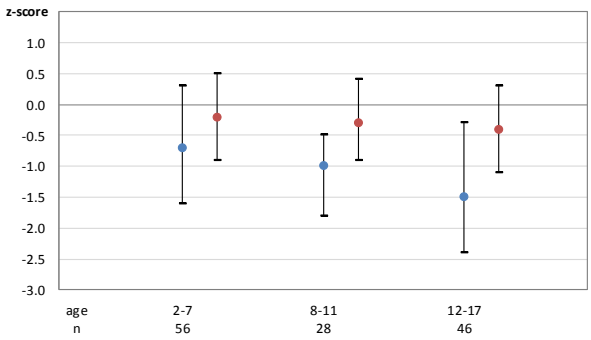


[figure 6.6 continued]

Quartiles of z-scores for BMI: Turkey



Quartiles of z-scores for BMI: Ukraine



Quartiles of z-scores for BMI: United Kingdom

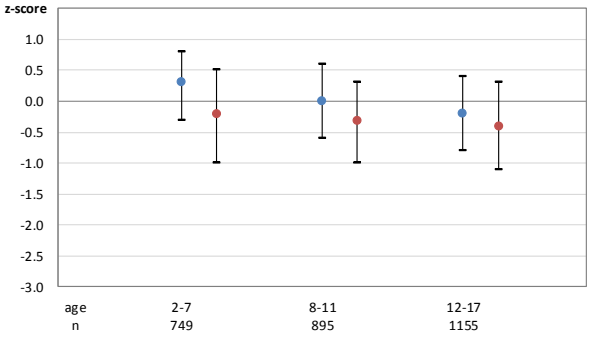
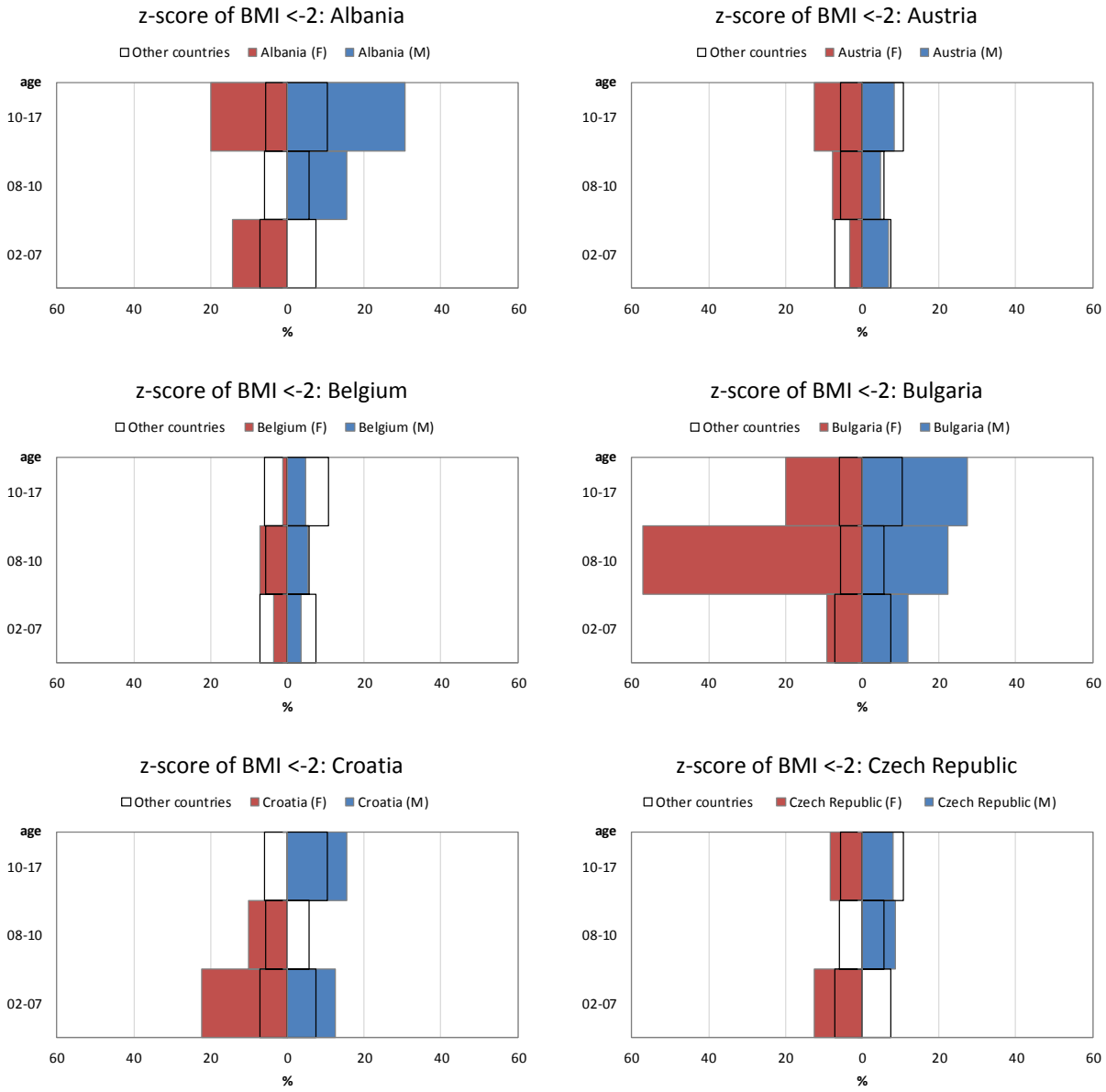
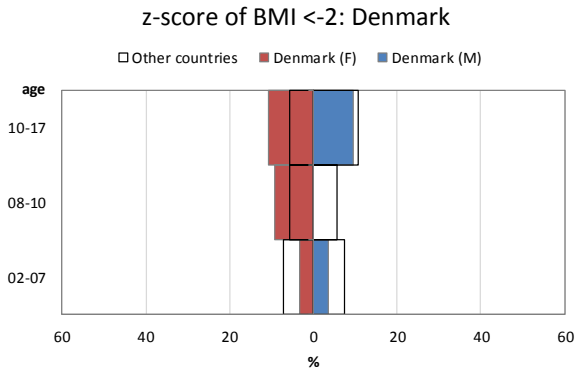


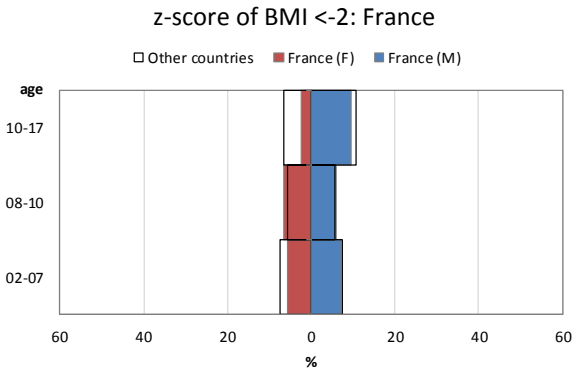
Figure 6.7 Proportion of child patients underweight (z-score of BMI<-2): age and sex pyramids, by country and overall. Patients aged 2-17 years in 2017.

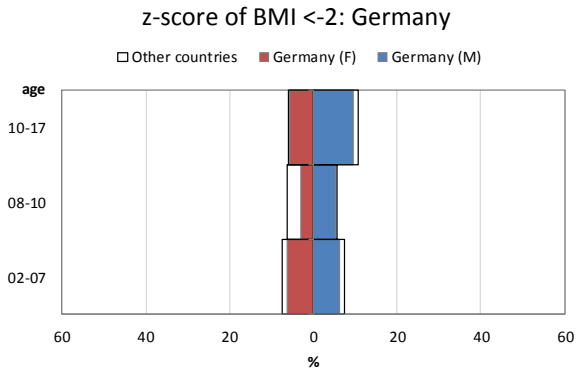
The coloured bars (red for females, blue for males) represent the percentage of underweight patients in the selected country, whereas the non-coloured bars represent the percentage of underweight patients in all the remaining countries (i.e. excluding that country). We excluded from the analyses those age groups where the number of patients was <10. We therefore excluded from the graphs Armenia, Latvia, Lithuania, Luxembourg and Republic of Moldova because some of the age groups in these countries had fewer than 10 patients.

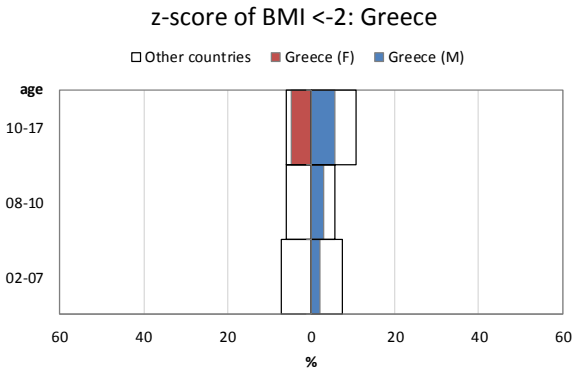


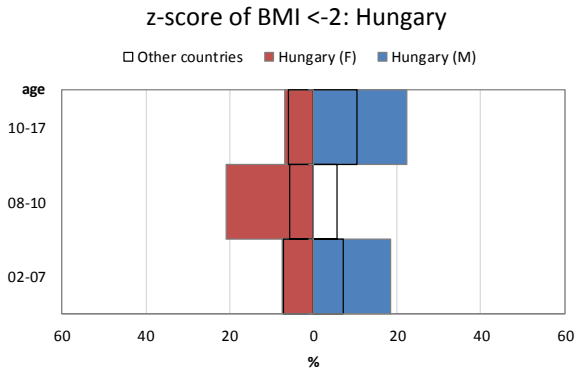
[figure 6.7 continued]

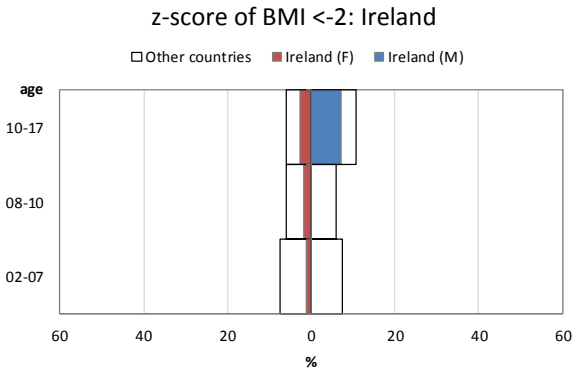


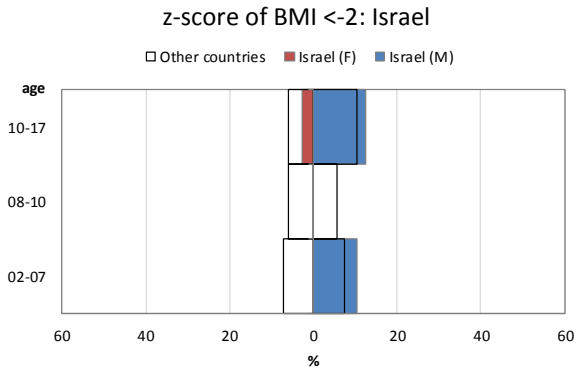


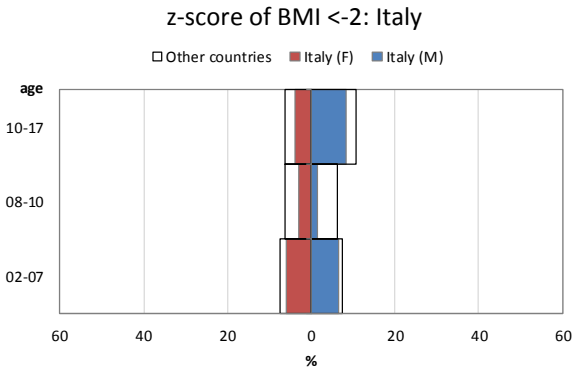




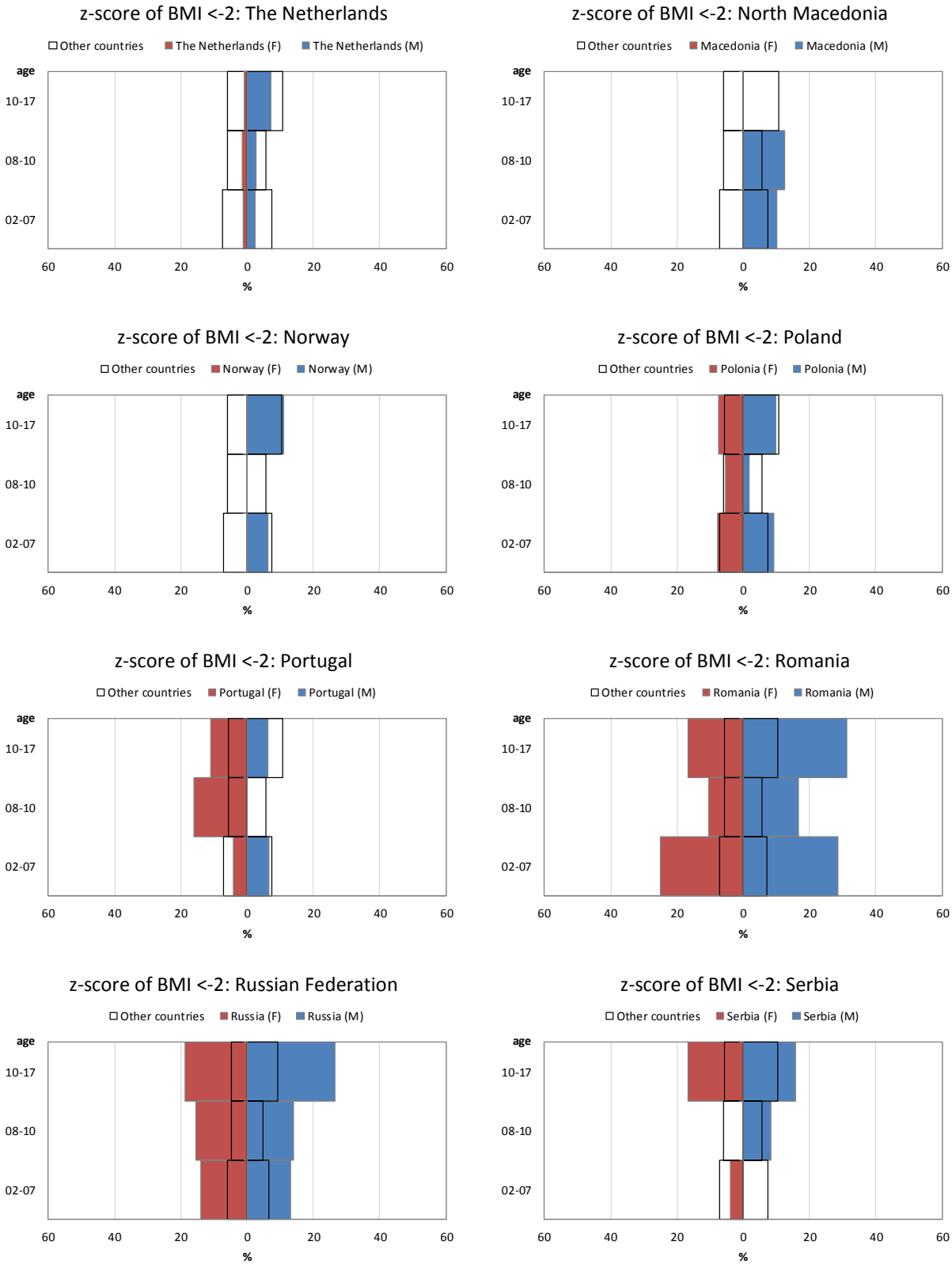








[figure 6.7 continued]



[figure 6.7 continued]

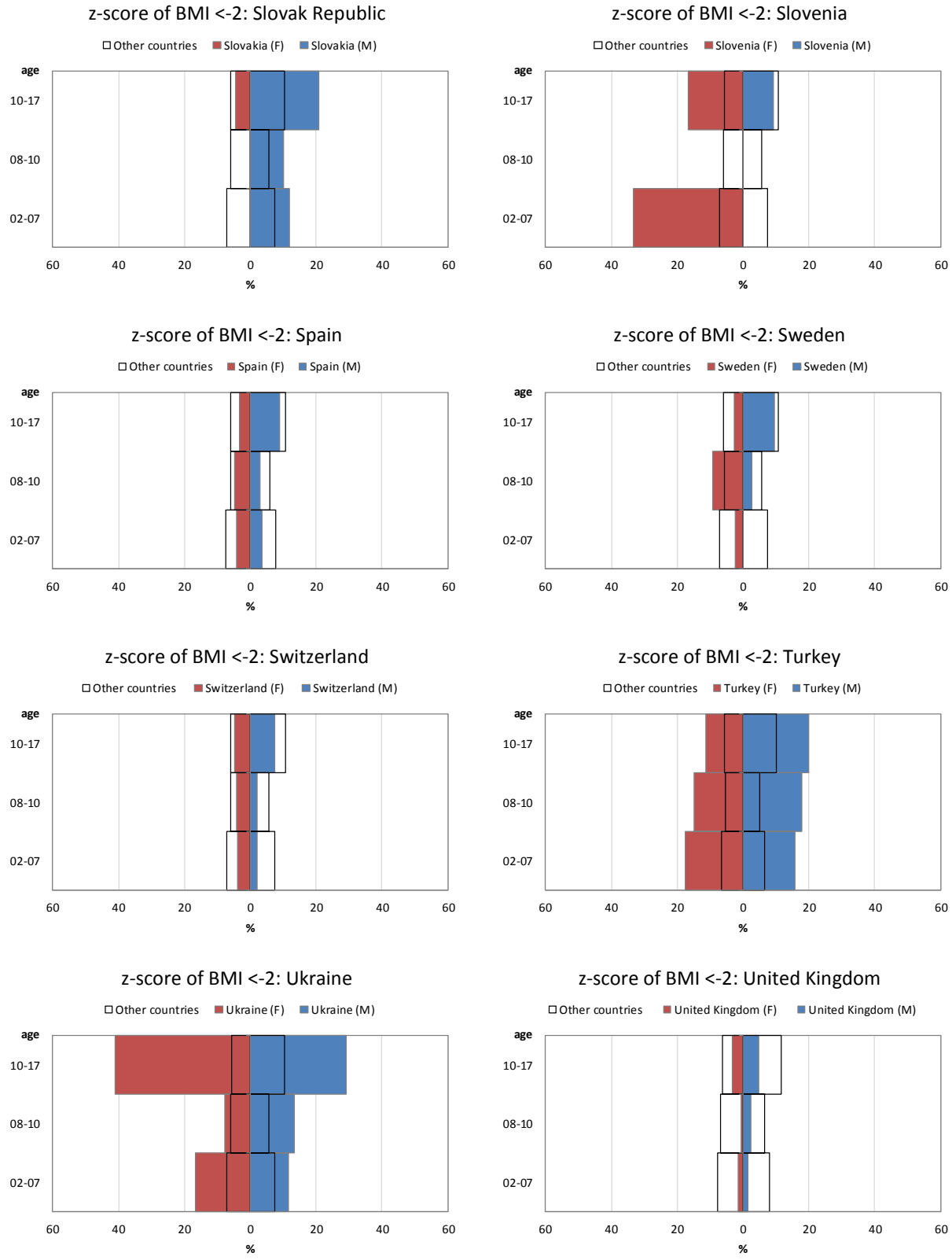
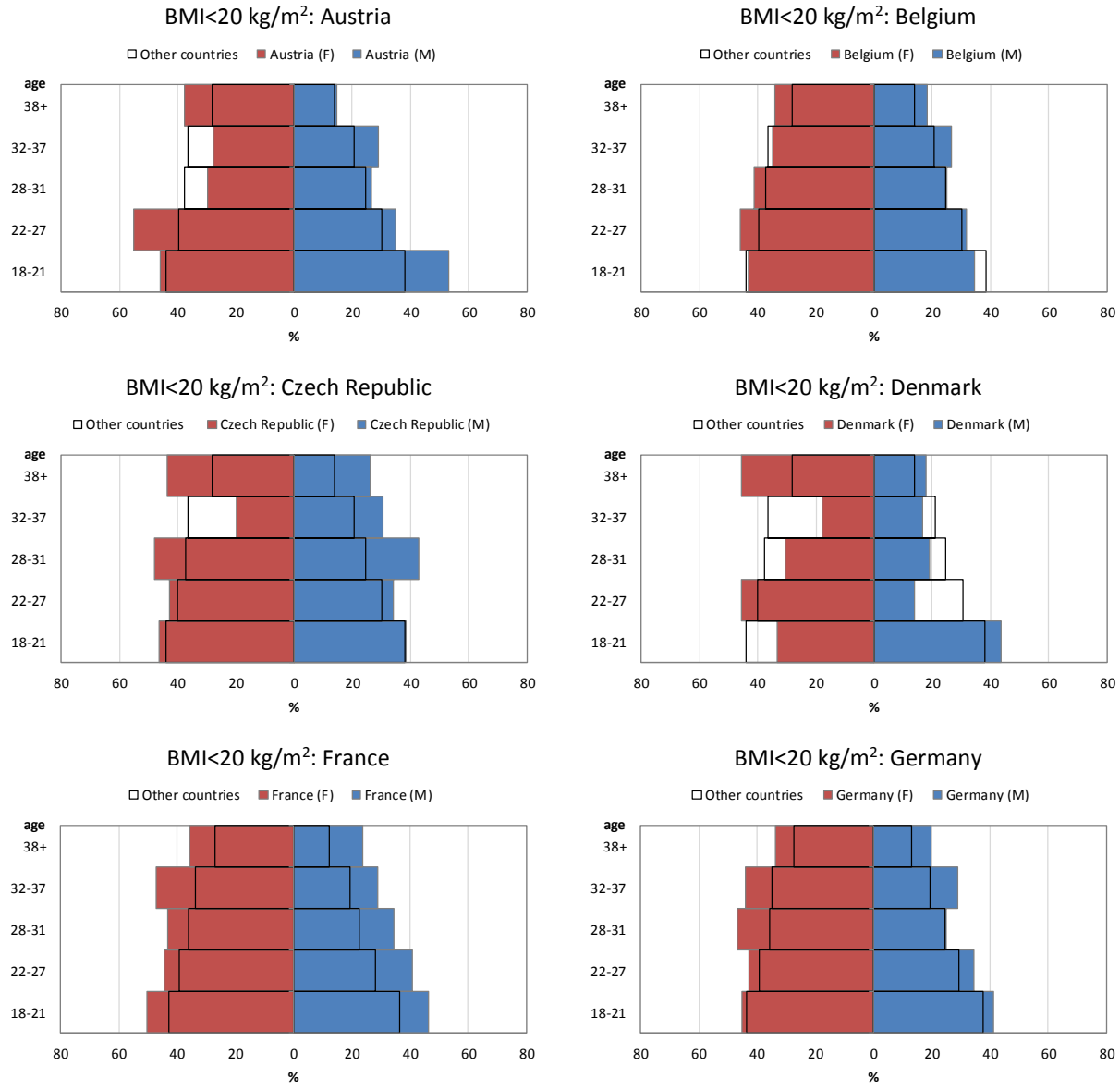
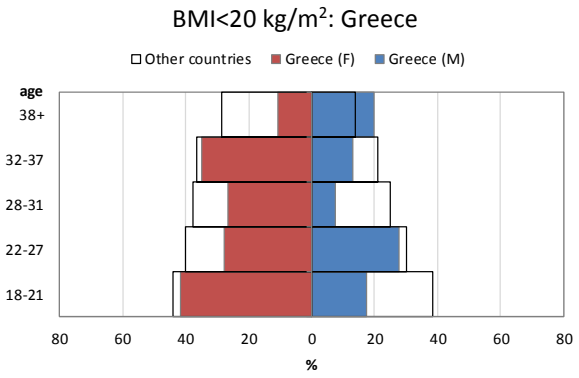


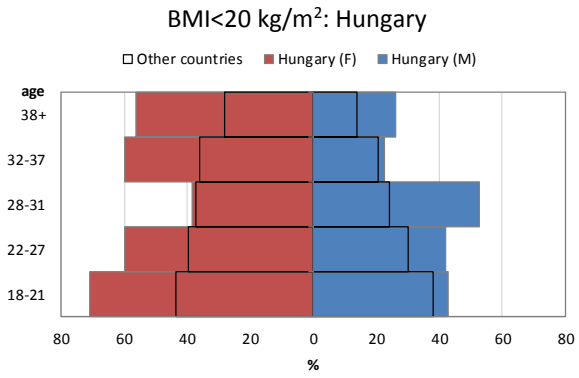
Figure 6.8 Proportion of adult patients with BMI<20: age and sex pyramids, by country and overall. Patients aged 18 years or older in 2017.

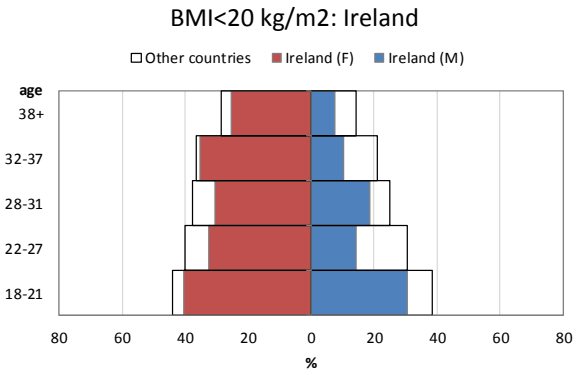
The coloured bars (red for females, blue for males) represent the percentage of underweight patients in the selected country, whereas the non-coloured bars represent the percentage of underweight patients in all the remaining countries (i.e. excluding that country). We excluded from the analyses those age groups where the number of patients was <10. We therefore excluded from the graphs Albania, Armenia, Bulgaria, Croatia, Latvia, Lithuania, Luxembourg, Republic of Moldova, Republic of Macedonia, Romania, Serbia, Slovenia, Turkey and Ukraine because some of the age groups in these countries had fewer than 10 patients.

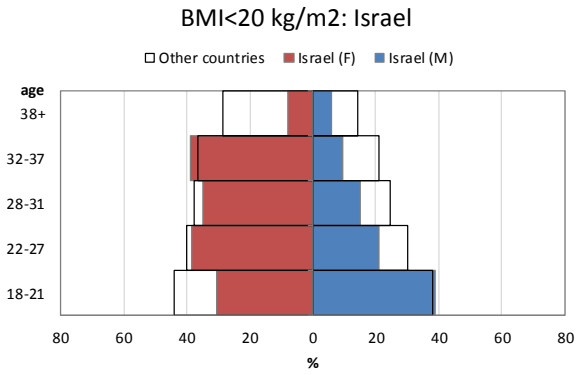


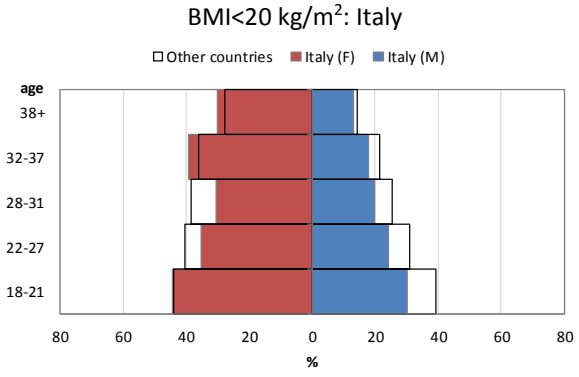
[figure 6.8 continued]

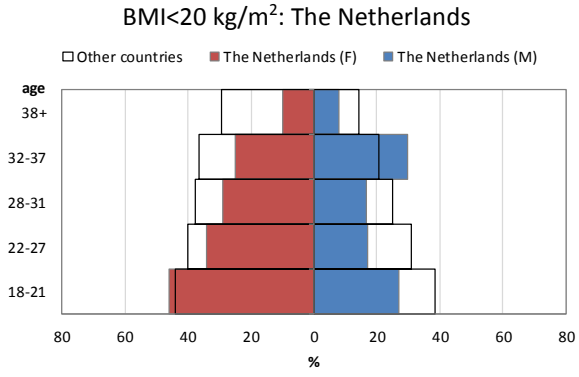


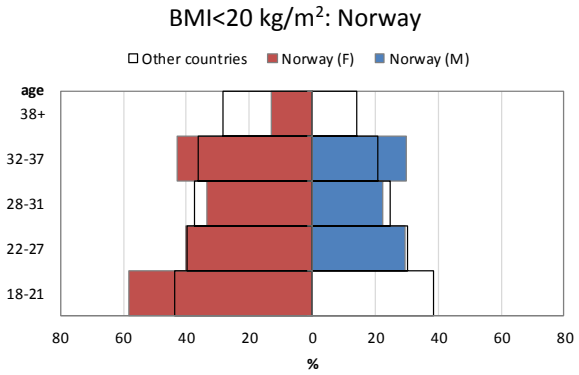


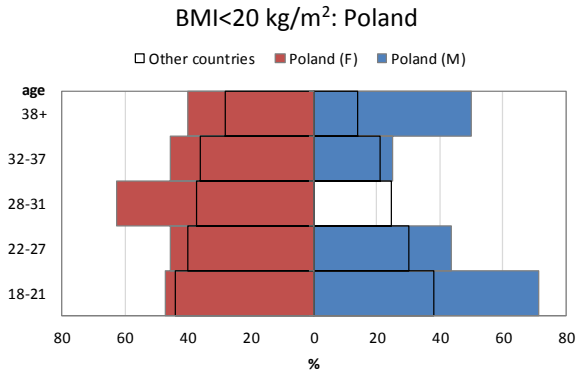






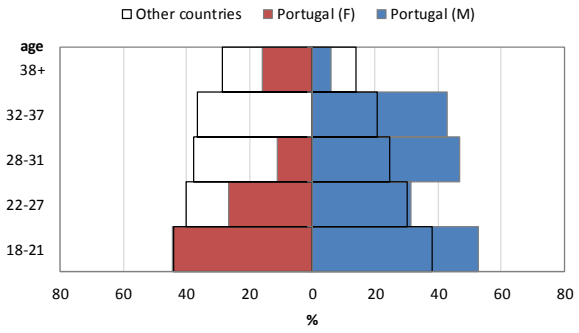




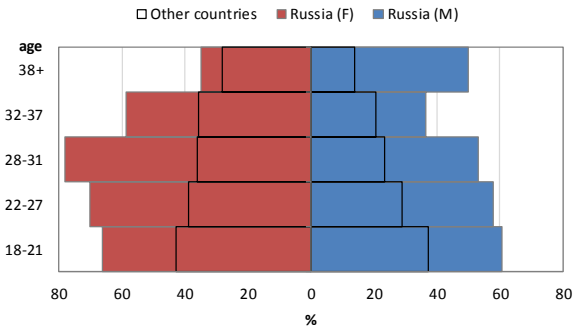


[figure 6.8 continued]

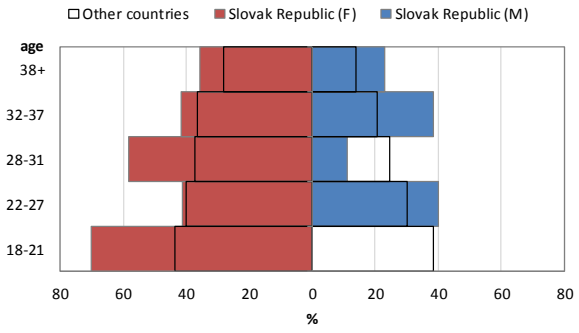
BMI<20 kg/m^2: Portugal



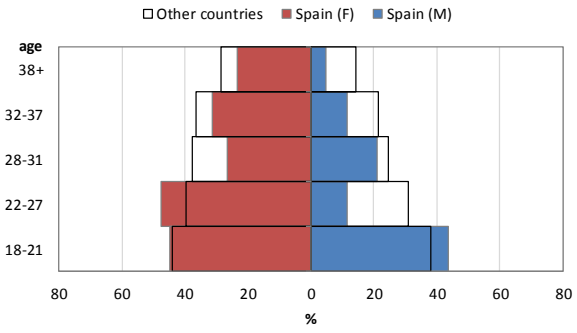
BMI<20 kg/m^2: Russian Federation



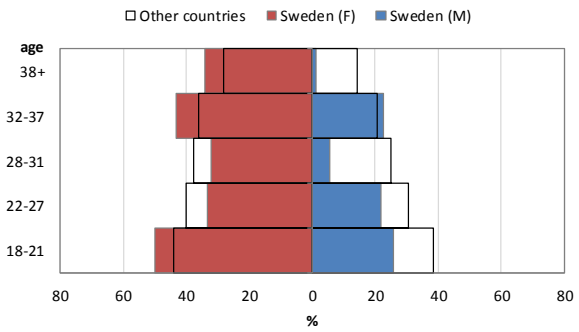
BMI<20 kg/m^2: Slovak Republic



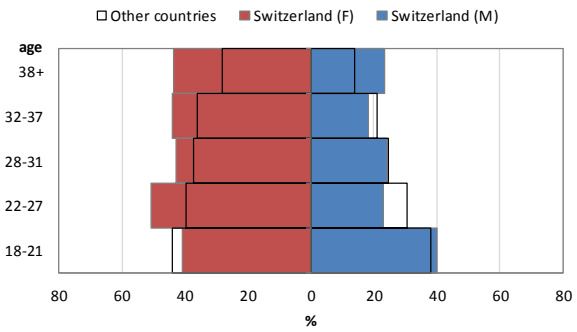
BMI<20 kg/m2: Spain



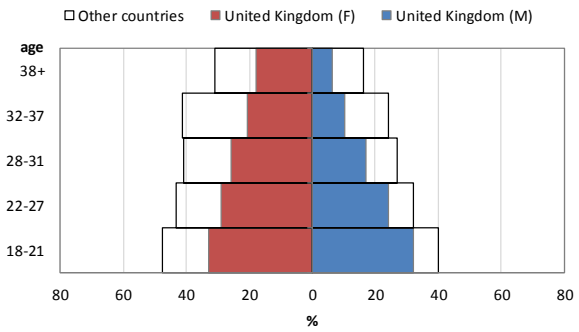
BMI<20 kg/m2: Sweden



BMI<20 kg/m2: Switzerland



BMI<20 kg/m2: United Kingdom



7. Complications and therapy

The information in this section should not be considered complete, for several reasons: national registries may use a different definition data about one or more complications is not collected, or the status of the complication is truly unknown (e.g. liver disease, where the definition requires ultrasound examination). 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 2 on page 144.

In this section we also present data on selected therapies. We collected information on therapies using the generic name of the drug, and 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”.

Table 7.1 Prevalence of allergic bronchopulmonary aspergillosis (ABPA) (all patients seen in 2017) and CF-related diabetes (CFRD) treated with insulin in 2017 (patients aged 18 years or older), by country.

Country	ABPA this year number (%)			CFRD with daily use of insulin this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	1 (0.82)	119 (97.54)	2 (1.64)	0 (0)	7 (87.50)	1 (12.50)
Armenia	10 (31.25)	20 (62.50)	2 (6.25)	-	-	-
Austria	1 (0.13)	727 (96.04)	29 (3.83)	1 (0.25)	271 (68.78)	122 (30.96)
Belgium[1]	315 (24.48)	892 (69.31)	80 (6.22)	0 (0)	563 (70.73)	233 (29.27)
Bulgaria	4 (2.70)	142 (95.95)	2 (1.35)	3 (4.48)	54 (80.60)	10 (14.93)
Croatia	2 (2.30)	83 (95.40)	2 (2.30)	1 (2.38)	34 (80.95)	7 (16.67)
Czech Republic	2 (0.33)	593 (98.02)	10 (1.65)	0 (0)	177 (62.54)	106 (37.46)
Denmark	496 (100)	-	-	0 (0)	197 (64.17)	110 (35.83)
France	0 (0)	6410 (92.36)	530 (7.64)	0 (0)	2848 (73.31)	1037 (26.69)
Germany	82 (1.34)	5608 (91.65)	429 (7.01)	197 (5.54)	2489 (69.96)	872 (24.51)
Greece	5 (0.83)	581 (96.99)	13 (2.17)	4 (1.28)	230 (73.72)	78 (25.00)
Hungary	4 (0.79)	497 (98.61)	3 (0.60)	4 (1.69)	165 (69.92)	67 (28.39)
Ireland	5 (0.41)	1139 (93.44)	75 (6.15)	4 (0.58)	506 (73.33)	180 (26.09)
Israel	15 (2.74)	504 (92.14)	28 (5.12)	10 (2.99)	220 (65.87)	104 (31.14)
Italy	111 (2)	5330 (95.85)	120 (2.16)	60 (1.89)	2382 (74.98)	735 (23.14)
Latvia	0 (0)	39 (100)	0 (0)	0 (0)	10 (90.91)	1 (9.09)
Lithuania	0 (0)	14 (100)	0 (0)	0 (0)	14 (100)	0 (0)
Luxembourg	0 (0)	31 (86.11)	5 (13.89)	0 (0)	13 (59.09)	9 (40.91)
Rep of Moldova	0 (0)	50 (100)	0 (0)	-	-	-
The Netherlands	243 (16.53)	1120 (76.19)	107 (7.28)	88 (9.67)	529 (58.13)	293 (32.20)
North Macedonia	2 (1.74)	111 (96.52)	2 (1.74)	1 (2.86)	23 (65.71)	11 (31.43)
Norway	9 (3.59)	238 (94.82)	4 (1.59)	8 (4.94)	123 (75.93)	31 (19.14)

[1] Belgium: ABPA is not collected for transplanted patients and most of the missing data refers to this sub-population.

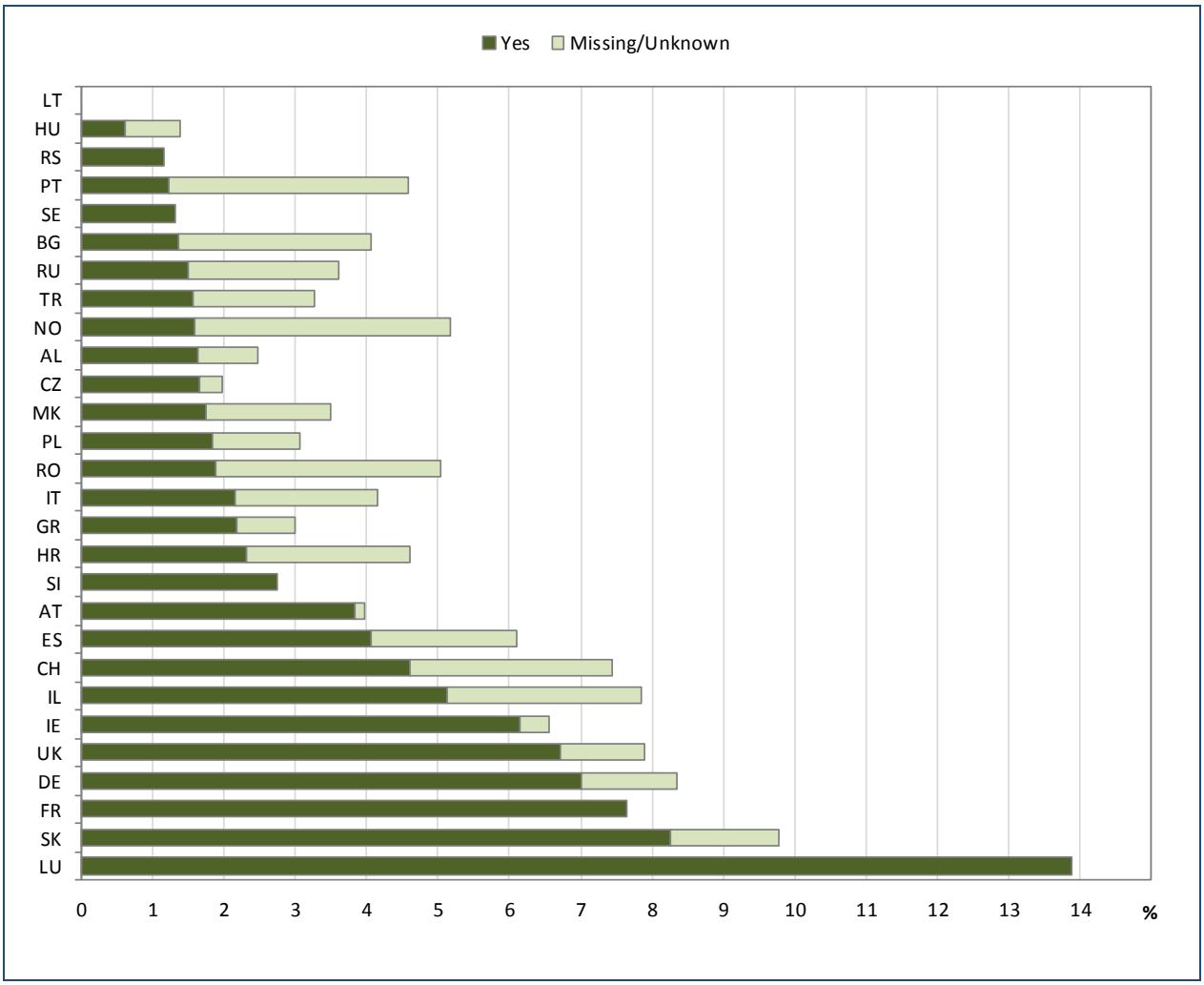
Note: Armenia, Rep of Moldova and Romania have <5 patients aged 18 years or more on 31/12/2017, therefore no information is included in the table for CFRD.

[table 7.1 continued]

Country	ABPA this year number (%)			CFRD with daily use of insulin this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Poland	8 (1.22)	636 (96.95)	12 (1.83)	1 (0.81)	105 (85.37)	17 (13.82)
Portugal	11 (3.36)	312 (95.41)	4 (1.22)	2 (1.40)	118 (82.52)	23 (16.08)
Romania	5 (3.14)	151 (94.97)	3 (1.89)	-	-	-
Russian Federation	65 (2.11)	2969 (96.40)	46 (1.49)	43 (6.11)	599 (85.09)	62 (8.81)
Serbia	0 (0)	170 (98.84)	2 (1.16)	0 (0)	34 (66.67)	17 (33.33)
Slovak Republic	4 (1.50)	240 (90.23)	22 (8.27)	2 (1.40)	127 (88.81)	14 (9.79)
Slovenia	0 (0)	106 (97.25)	3 (2.75)	1 (2.17)	34 (73.91)	11 (23.91)
Spain	41 (2.05)	1880 (93.91)	81 (4.05)	24 (2.58)	669 (71.78)	239 (25.64)
Sweden	0 (0)	677 (98.69)	9 (1.31)	0 (0)	318 (73.61)	114 (26.39)
Switzerland	26 (2.84)	846 (92.56)	42 (4.60)	3 (0.61)	338 (69.12)	148 (30.27)
Turkey	24 (1.70)	1365 (96.74)	22 (1.56)	2 (2.02)	79 (79.80)	18 (18.18)
Ukraine	2 (1.21)	163 (98.79)	0 (0)	2 (8.00)	22 (88.00)	1 (4.00)
United Kingdom	116 (1.17)	9107 (92.11)	664 (6.72)	0 (0)	3919 (69.40)	1728 (30.60)

Table 7.1 shows the frequency of allergic bronchopulmonary aspergillosis (see Appendix 2, page 144, for ABPA definitions) and CF-related diabetes (CFRD) with daily use of insulin this year, 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 2017, by country.

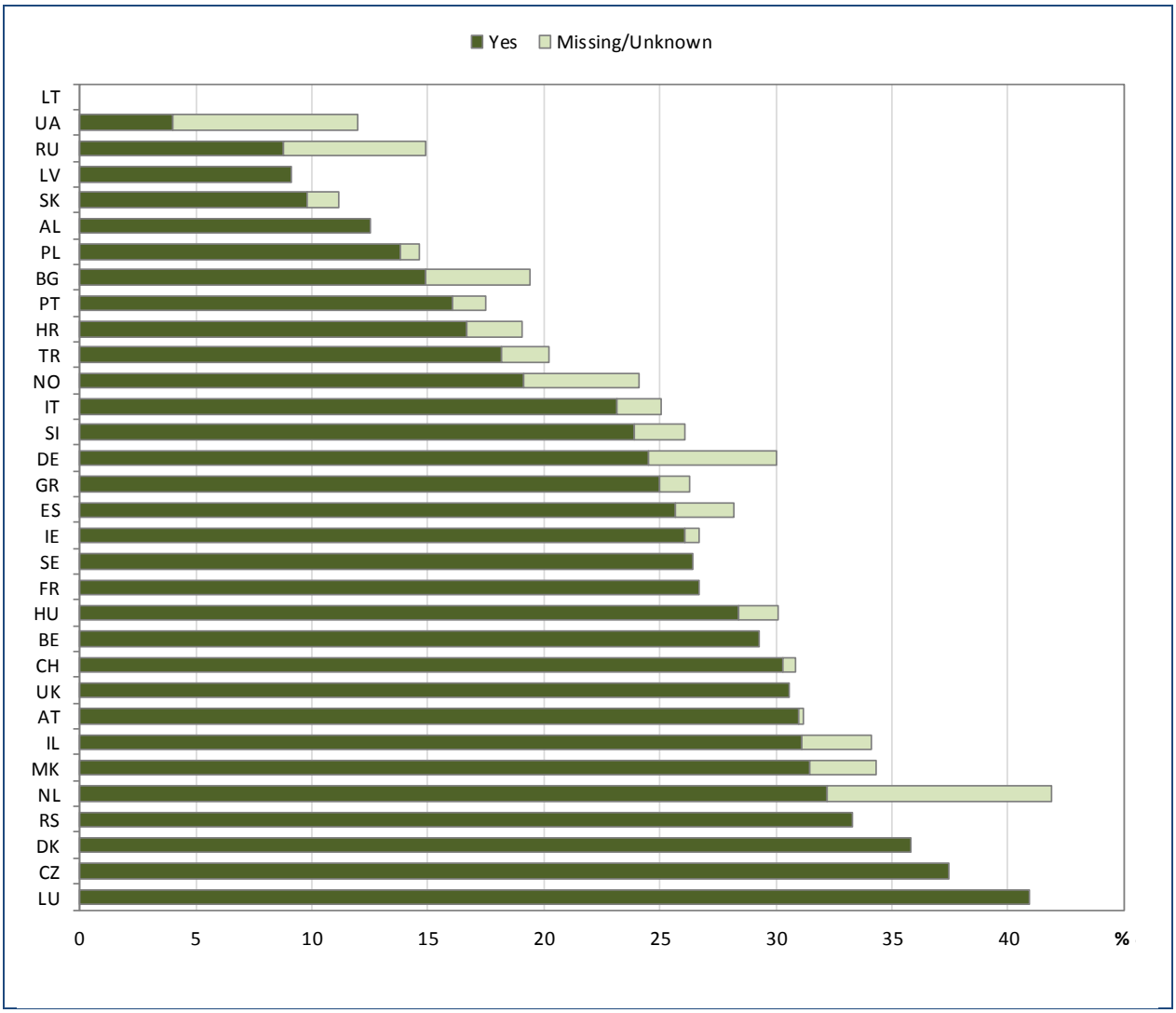


Note: We excluded from the graph the countries for which the information on allergic bronchopulmonary aspergillosis (ABPA) was missing for more than 10% of the patients.

Note: Belgium: ABPA is not collected for transplanted patients and most of the missing data refers to this sub-population. France collected ABPA as Aspergillosis (ABPA and other) if treated.

This graph shows the frequency of allergic bronchopulmonary aspergillosis by country (for the definition of ABPA see Appendix 2, page 144). 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 requiring daily insulin treatment, by country. All patients seen in 2017 aged 18 years or older.



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

This graph shows the prevalence of CF-related diabetes (CFRD) with daily use of insulin this year, by country. CFRD is recorded differently among the national registries. As a substitute marker of diabetes, we have collected data on the use of insulin on a daily basis. The dark green part of the bar shows the percentage of patients who use insulin daily, the light green part shows the percentage of patients for whom this information was missing. Only patients aged 18 years or older were included in this graph.

Table 7.2 Prevalence of pneumothorax, haemoptysis and malignancy in all patients seen in 2017, by country.

Country	Pneumothorax requiring chest tube this year number (%)			Haemoptysis major over 250 ml this year number (%)			Malignancy occurred this year number(%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	2 (1.64)	120 (98.36)	0 (0)	5 (4.10)	117 (95.90)	0 (0)	2 (1.64)	120 (98.36)	0 (0)
Armenia	2 (6.25)	30 (93.75)	0 (0)	2 (6.25)	19 (59.38)	11 (34.38)	2 (6.25)	30 (93.75)	0 (0)
Austria	1 (0.13)	753 (99.47)	3 (0.40)	8 (1.06)	739 (97.62)	10 (1.32)	3 (0.40)	752 (99.34)	2 (0.26)
Belgium[1]	314 (24.40)	971 (75.45)	2 (0.16)	314 (24.40)	968 (75.21)	5 (0.39)	0 (0)	1282 (99.61)	5 (0.39)
Bulgaria	4 (2.70)	143 (96.62)	1 (0.68)	5 (3.38)	130 (87.84)	13 (8.78)	4 (2.70)	143 (96.62)	1 (0.68)
Croatia	1 (1.15)	86 (98.85)	0 (0)	1 (1.15)	82 (94.25)	4 (4.60)	1 (1.15)	86 (98.85)	0 (0)
Czech Republic	15 (2.48)	585 (96.69)	5 (0.83)	12 (1.98)	589 (97.36)	4 (0.66)	0 (0)	603 (99.67)	2 (0.33)
Denmark	0 (0)	495 (99.80)	1 (0.20)	496 (100)	-	-	0 (0)	494 (99.60)	2 (0.40)
France	0 (0)	6908 (99.54)	32 (0.46)	0 (0)	6889 (99.27)	51 (0.73)	0 (0)	6871 (99.01)	69 (0.99)
Germany	95 (1.55)	5990 (97.89)	34 (0.56)	137 (2.24)	5964 (97.47)	18 (0.29)	89 (1.45)	5961 (97.42)	69 (1.13)
Greece	6 (1.00)	591 (98.66)	2 (0.33)	7 (1.17)	586 (97.83)	6 (1.00)	6 (1.00)	592 (98.83)	1 (0.17)
Hungary	5 (0.99)	496 (98.41)	3 (0.60)	13 (2.58)	479 (95.04)	12 (2.38)	5 (0.99)	493 (97.82)	6 (1.19)
Ireland	5 (0.41)	1213 (99.51)	<5 (0.08)	5 (0.41)	1211 (99.34)	<5 (0.25)	5 (0.41)	1212 (99.43)	<5 (0.16)
Israel	14 (2.56)	531 (97.07)	2 (0.37)	17 (3.11)	487 (89.03)	43 (7.86)	12 (2.19)	534 (97.62)	1 (0.18)
Italy	95 (1.71)	5446 (97.93)	20 (0.36)	97 (1.74)	5406 (97.21)	58 (1.04)	97 (1.74)	5435 97.73)	29 (0.52)
Latvia	0 (0)	39 (100)	0 (0)	0 (0)	39 (100)	0 (0)	0 (0)	39 (100)	0 (0)
Lithuania	0 (0)	14 (100)	0 (0)	0 (0)	14 (100)	0 (0)	0 (0)	14 (100)	0 (0)
Luxembourg	0 (0)	36 (100)	0 (0)	0 (0)	36 (100)	0 (0)	0 (0)	35 (97.22)	1 (2.78)
North Macedonia	2 (1.74)	112 (97.39)	1 (0.87)	2 (1.74)	113 (98.26)	0 (0)	2 (1.74)	113 (98.26)	0 (0)
Rep of Moldova	0 (0)	50 (100)	0 (0)	0 (0)	45 (90.00)	5 (10.00)	0 (0)	50 (100)	0 (0)
The Netherlands	257 (17.48)	1208 (82.18)	5 (0.34)	269 (18.30)	1093 (74.35)	108 (7.35)	99 (6.73)	1369 (93.13)	2 (0.14)
Norway	8 (3.19)	242 (96.41)	1 (0.40)	7 (2.79)	242 (96.41)	2 (0.80)	6 (2.39)	243 (96.81)	2 (0.80)

[1] Belgium: pneumothorax requiring chest tube and haemoptysis major over 250 ml are not collected for transplanted patients and most of the missing data refers to this sub-population.

Note: Ireland: when the number of patients is less than 5 the information is suppressed.

[table 7.2 continued]

Country	Pneumothorax requiring chest tube this year number (%)			Haemoptysis major over 250 ml this year number (%)			Malignancy occurred this year number(%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Poland	10 (1.52)	643 (98.02)	3 (0.46)	12 (1.83)	607 (92.53)	37 (5.64)	10 (1.52)	645 (98.32)	1 (0.15)
Portugal	10 (3.06)	315 (96.33)	2 (0.61)	10 (3.06)	289 (88.38)	28 (8.56)	10 (3.06)	317 (96.94)	0 (0)
Romania	4 (2.52)	155 (97.48)	0 (0)	6 (3.77)	150 (94.34)	3 (1.89)	26 (16.35)	133 (83.65)	0 (0)
Russian Federation	62 (2.01)	3000 (97.40)	18 (0.58)	77 (2.50)	2959 (96.07)	44 (1.43)	76 (2.47)	2997 (97.31)	7 (0.23)
Serbia	0 (0)	172 (100)	0 (0)	0 (0)	164 (95.35)	8 (4.65)	0 (0)	171 (99.42)	1 (0.58)
Slovak Republic	2 (0.75)	264 (99.25)	0 (0)	2 (0.75)	250 (93.98)	14 (5.26)	4 (1.5)	259 (97.37)	3 (1.13)
Slovenia	0 (0)	108 (99.08)	1 (0.92)	3 (2.75)	106 (97.25)	0 (0)	1 (0.92)	108 (99.08)	0 (0)
Spain	32 (1.60)	1959 (97.85)	11 (0.55)	33 (1.65)	1897 (94.76)	72 (3.60)	37 (1.85)	1955 (97.65)	10 (0.50)
Sweden	0 (0)	684 (99.71)	2 (0.29)	0 (0)	686 (100)	0 (0)	0 (0)	684 (99.71)	2 (0.29)
Switzerland	24 (2.63)	886 (96.94)	4 (0.44)	28 (3.06)	856 (93.65)	30 (3.28)	25 (2.74)	884 (96.72)	5 (0.55)
Turkey	16 (1.13)	1393 (98.72)	2 (0.14)	19 (1.35)	1382 (97.94)	10 (0.71)	20 (1.42)	1390 (98.51)	1 (0.07)
Ukraine	2 (1.21)	161 (97.58)	2 (1.21)	1 (0.61)	152 (92.12)	12 (7.27)	0 (0)	165 (100)	0 (0)
United Kingdom	116 (1.17)	9738 (98.49)	33 (0.33)	0 (0)	9856 (99.69)	31 (0.31)	0 (0)	9853 (99.66)	34 (0.34)

Table 7.2 shows the frequency of three rare complications: Pneumothorax (collapsed lung) requiring chest tube, haemoptysis (coughing up of blood) of more than 250 ml and occurrence of malignancy (cancer). All these complications are extremely rare.

Table 7.3 Prevalence of liver disease and use of ursodeoxycholic acid in all patients seen in 2017, by country.

Country	Liver disease this year number (%)						Ursodeoxycholic acid this year number (%)		
	Missing/ unknown	No liver disease	Cirrhosis			Liver disease without cirrhosis	Missing/ unknown	No	Yes
			Cirrhosis with portal hypertension/ hypersplenism	Cirrhosis no portal hypertension/ hypersplenism	Cirrhosis. portal hypertension unknown				
Albania	1 (0.82)	80 (65.57)	0 (0)	1 (0.82)	0 (0)	40 (32.79)	3 (2.46)	81 (66.39)	38 (31.15)
Armenia	2 (6.25)	12 (37.50)	2 (6.25)	5 (15.63)	0 (0)	11 (34.38)	6 (18.75)	15 (46.88)	11 (34.38)
Austria	7 (0.92)	439 (57.99)	23 (3.04)	7 (0.92)	5 (0.66)	276 (36.46)	0 (0)	409 (54.03)	348 (45.97)
Belgium[1]	3 (0.23)	1216 (94.48)	68 (5.28)	0 (0)	0 (0)	0 (0)	111 (8.62)	940 (73.04)	236 (18.34)
Bulgaria	4 (2.70)	110 (74.32)	8 (5.41)	0 (0)	0 (0)	26 (17.57)	5 (3.38)	101 (68.24)	42 (28.38)
Croatia	1 (1.15)	69 (79.31)	7 (8.05)	0 (0)	0 (0)	10 (11.49)	0 (0)	51 (58.62)	36 (41.38)
Czech Republic	16 (2.64)	445 (73.55)	4 (0.66)	7 (1.16)	0 (0)	133 (21.98)	0 (0)	405 (66.94)	200 (33.06)
Denmark	0 (0)	403 (81.25)	21 (4.23)	8 (1.61)	0 (0)	64 (12.90)	0 (0)	351 (70.77)	145 (29.23)
France	0 (0)	5961 (85.89)	126 (1.82)	135 (1.95)	0 (0)	718 (10.35)	0 (0)	5381 (77.54)	1559 (22.46)
Germany	757 (12.37)	3926 (64.16)	133 (2.17)	85 (1.39)	101 (1.65)	1117 (18.25)	63 (1.03)	3053 (49.89)	3003 (49.08)
Greece	5 (0.83)	439 (73.29)	13 (2.17)	8 (1.34)	6 (1.00)	128 (21.37)	5 (0.83)	432 (72.12)	162 (27.05)
Hungary	6 (1.19)	392 (77.78)	67 (13.29)	17 (3.37)	16 (3.17)	6 (1.19)	19 (3.77)	277 (54.96)	208 (41.27)
Ireland[3]	5 (0.41)	1048 (85.97)	43 (3.53)	7 (0.57)	8 (0.66)	108 (8.86)	5 (0.41)	1084 (88.93)	130 (10.66)
Israel	15 (2.74)	433 (79.16)	12 (2.19)	4 (0.73)	1 (0.18)	82 (14.99)	8 (1.46)	454 (83.00)	85 (15.54)
Italy	107 (1.92)	3976 (71.50)	57 (1.02)	49 (0.88)	8 (0.14)	1364 (24.53)	62 (1.11)	3570 (64.20)	1929 (34.69)
Latvia	0 (0)	20 (51.28)	2 (5.13)	0 (0)	1 (2.56)	16 (41.03)	0 (0)	24 (61.54)	15 (38.46)
Lithuania	0 (0)	14 (100)	0 (0)	0 (0)	0 (0)	0 (0)	0 (0)	14 (100)	0 (0)
Luxembourg	0 (0)	25 (69.44)	2 (5.56)	0 (0)	0 (0)	9 (25.00)	0 (0)	19 (52.78)	17 (47.22)
North Macedonia	2 (1.74)	59 (51.30)	3 (2.61)	15 (13.04)	0 (0)	36 (31.30)	2 (1.74)	59 (51.30)	54 (46.96)
Rep of Moldova	0 (0)	44 (88.00)	0 (0)	0 (0)	0 (0)	6 (12.00)	0 (0)	45 (90.00)	5 (10.00)
The Netherlands	289 (19.66)	929 (63.20)	67 (4.56)	27 (1.84)	4 (0.27)	154 (10.48)	93 (6.33)	1017 (69.18)	360 (24.49)
Norway	5 (1.99)	215 (85.66)	8 (3.19)	5 (1.99)	0 (0)	18 (7.17)	12 (4.78)	219 (87.25)	20 (7.97)

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

[table 7.3 continued]

Country	Liver disease this year number (%)						Ursodeoxycholic acid this year number (%)		
	Missing/ unknown	No liver disease	Cirrhosis			Liver disease without cirrhosis	Missing/ unknown	No	Yes
			Cirrhosis with portal hypertension/ hypersplenism	Cirrhosis no portal hypertension/ hypersplenism	Cirrhosis. portal hypertension unknown				
Poland	17 (2.59)	415 (63.26)	26 (3.96)	8 (1.22)	3 (0.46)	187 (28.51)	11 (1.68)	281 (42.84)	364 (55.49)
Portugal	14 (4.28)	246 (75.23)	4 (1.22)	0 (0)	0 (0)	63 (19.27)	3 (0.92)	234 (71.56)	90 (27.52)
Romania	5 (3.14)	118 (74.21)	6 (3.77)	2 (1.26)	3 (1.89)	25 (15.72)	2 (1.26)	122 (76.73)	35 (22.01)
Russian Federation	91 (2.95)	2291 (74.38)	136 (4.42)	68 (2.21)	20 (0.65)	474 (15.39)	70 (2.27)	309 (10.03)	2701 (87.69)
Serbia[2]	0 (0)	112 (65.12)	7 (4.07)	3 (1.74)	1 (0.58)	49 (28.49)	0 (0)	113 (65.70)	59 (34.30)
Slovak Republic	2 (0.75)	120 (45.11)	9 (3.38)	18 (6.77)	1 (0.38)	116 (43.61)	1 (0.38)	123 (46.24)	142 (53.38)
Slovenia	0 (0)	73 (66.97)	3 (2.75)	8 (7.34)	0 (0)	25 (22.94)	0 (0)	54 (49.54)	55 (50.46)
Spain	40 (2.00)	1501 (74.98)	39 (1.95)	7 (0.35)	3 (0.15)	412 (20.58)	40 (2.00)	1501 (74.98)	461 (23.03)
Sweden[3]	0 (0)	541 (78.86)	15 (2.19)	11 (1.60)	0 (0)	119 (17.35)	11 (1.60)	533 (77.70)	142 (20.70)
Switzerland	41 (4.49)	637 (69.69)	28 (3.06)	13 (1.42)	3 (0.33)	192 (21.01)	3 (0.33)	659 (72.10)	252 (27.57)
Turkey	16 (1.13)	1233 (87.38)	7 (0.50)	3 (0.21)	3 (0.21)	149 (10.56)	23 (1.63)	1156 (81.93)	232 (16.44)
Ukraine	1 (0.61)	41 (24.85)	11 (6.67)	9 (5.45)	5 (3.03)	98 (59.39)	1 (0.61)	5 (3.03)	159 (96.36)
United Kingdom[4]	0 (0)	8344 (84.39)	149 (1.51)	125 (1.26)	0 (0)	1269 (12.84)	0 (0)	8423 (85.19)	1464 (14.81)

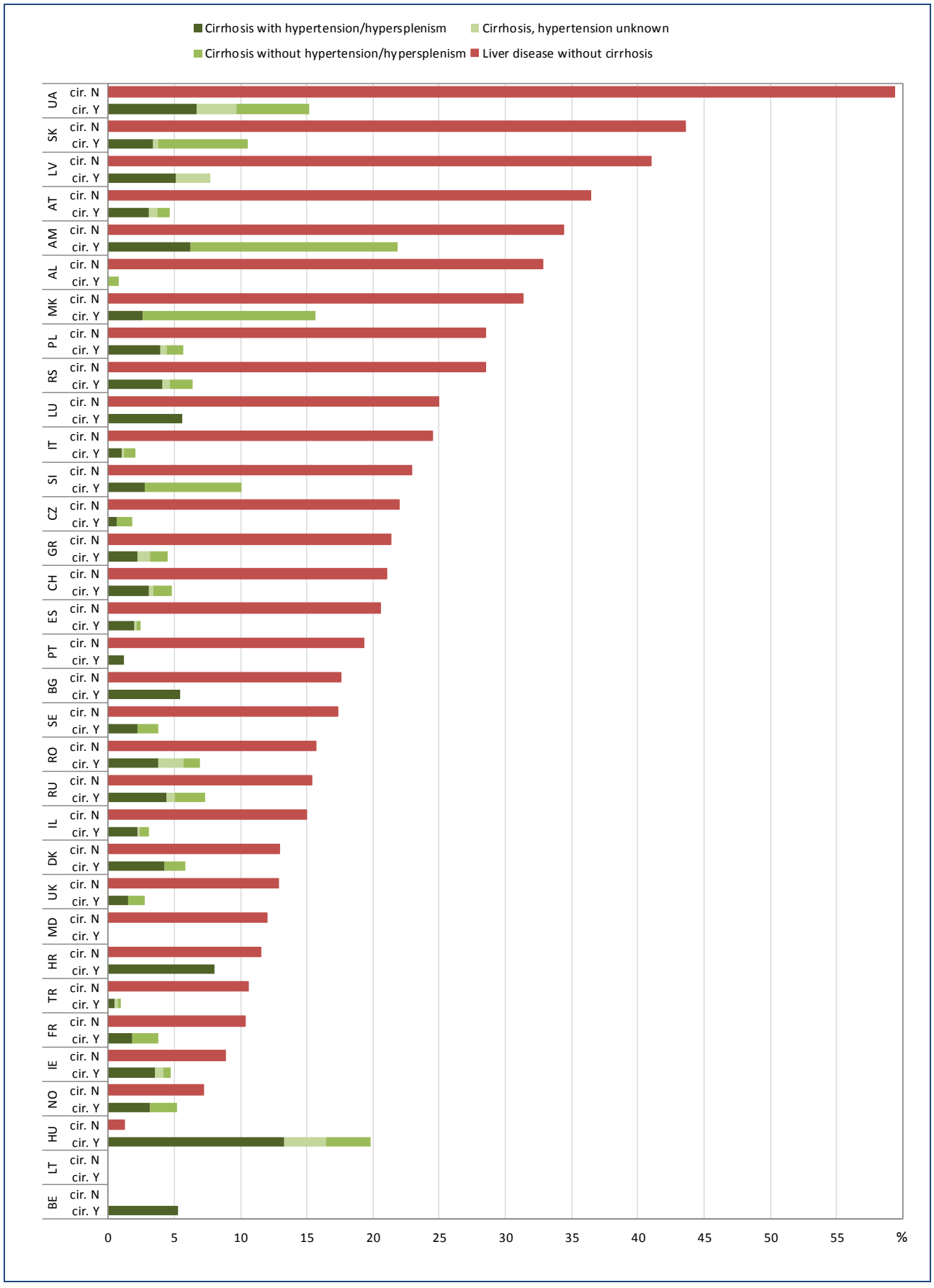
[2] Serbia: cirrhosis without portal hypertension/hypersplenism means the presence of CF-related liver disease with normal liver function.

[3] Sweden: has only collected cirrhosis with portal hypertension yes or no this year. The rest have been set to No liver disease due to software issues. The prevalence of use of ursodeoxycholic acid could be used as an indicator of the total prevalence of liver disease of all categories.

[4] UK: after additional data cleaning, these figures do not match those in the UK 2017 data annual report.

This table shows the frequency and severity of liver disease according to the ECFSPR definitions (see Appendix 2, page 144) and use of ursodeoxycholic acid, a commonly used treatment for CF liver disease. The frequency and severity of liver disease differs greatly throughout the Registry data and does not correspond to the number of patients on ursodeoxycholic acid.

Figure 7.3 Prevalence and severity of liver disease in all patients seen in 2017, by country.



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

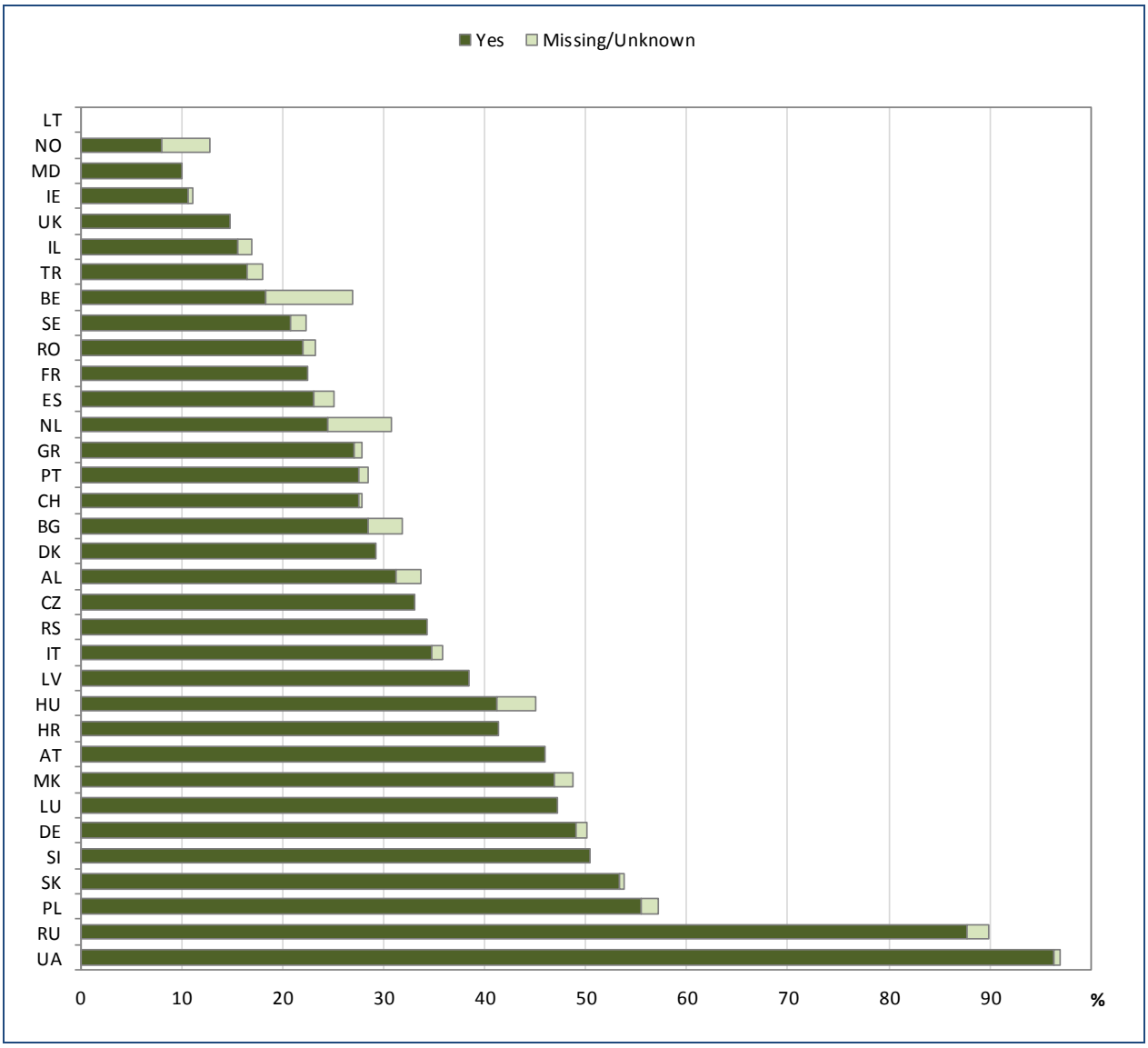
[Figure 7.4 continued]

Note: Belgium: collects only cirrhosis with portal hypertension yes or no. No liver disease therefore means no cirrhosis with portal hypertension.
Serbia: cirrhosis without portal hypertension/hypersplenism means the presence of CF-related disease with normal liver function.
Sweden: has only collected cirrhosis with portal hypertension yes or no this year. The rest have been set to "No liver disease" due to software issues. The prevalence of use of ursodeoxycholic acid could be used as an indicator of the total amount of liver disease of all categories.
UK: after additional data cleaning, these figures do not match those in the UK 2017 data annual report.

This graph 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 2, page 144).

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.

Figure 7.5 Use of ursodeoxycholic acid in all patients seen in 2017, by country.



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

This graph shows how many patients used ursodeoxycholic acid during the survey year. 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 7.4 Use of hypertonic saline, rhDNase and bronchodilators in all patients seen in 2017, by country.

Country	Hypertonic saline (NaCl) inhaled > 3 months this year number (%)			rhDNase inhaled > 3 months this year number (%)			Bronchodilators inhaled > 3 months this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	2 (1.64)	13 (10.66)	107 (87.70)	2 (1.64)	115 (94.26)	5 (4.10)	1 (0.82)	46 (37.70)	75 (61.48)
Armenia	6 (18.75)	0 (0	26 (81.25)	6 (18.75)	20 (62.5)	6 (18.75)	6 (18.75)	4 (12.5)	22 (68.75)
Austria	1 (0.13)	290 (38.31)	466 (61.56)	5 (0.66)	392 (51.78)	360 (47.56)	0 (0)	95 (12.55)	662 (87.45)
Belgium[1]	236 (18.34)	347 (26.96)	704 (54.70)	236 (18.34)	127 (9.87)	924 (71.79)	236 (18.34)	219 (17.02)	832 (64.65)
Bulgaria	4 (2.70)	46 (31.08)	98 (66.22)	4 (2.70)	28 (18.92)	116 (78.38)	4 (2.70)	102 (68.92)	42 (28.38)
Croatia	0 (0)	14 (16.09)	73 (83.91)	0 (0)	15 (17.24)	72 (82.76)	0 (0)	67 (77.01)	20 (22.99)
Czech Republic[2]	0 (0)	167 (27.60)	438 (72.40)	0 (0)	217 (35.87)	388 (64.13)	0 (0)	230 (38.02)	375 (61.98)
Denmark	496 (100)	-	-	0 (0)	90 (18.15)	406 (81.85)	496 (100)	-	-
France	0 (0)	6129 (88.31)	811 (11.69)	0 (0)	3919 (56.47)	3021 (43.53)	0 (0)	2852 (41.10)	4088 (58.90)
Germany	64 (1.05)	1281 (20.93)	4774 (78.02)	65 (1.06)	2942 (48.08)	3112 (50.86)	59 (0.96)	1072 (17.52)	4988 (81.52)
Greece	5 (0.83)	375 (62.60)	219 (36.56)	5 (0.83)	180 (30.05)	414 (69.12)	10 (1.67)	261 (43.57)	328 (54.76)
Hungary	13 (2.58)	148 (29.37)	343 (68.06)	17 (3.37)	185 (36.71)	302 (59.92)	13 (2.58)	194 (38.49)	297 (58.93)
Ireland	5 (0.41)	565 (46.35)	649 (53.24)	5 (0.41)	586 (48.07)	628 (51.52)	5 (0.41)	380 (31.17)	834 (68.42)
Israel	6 (1.10)	146 (26.69)	395 (72.21)	8 (1.46)	159 (29.07)	380 (69.47)	8 (1.46)	198 (36.20)	341 (62.34)
Italy	63 (1.13)	3033 (54.54)	2465 (44.33)	64 (1.15)	3660 (65.82)	1837 (33.03)	63 (1.13)	1548 (27.84)	3950 (71.03)
Latvia	0 (0)	2 (5.13)	37 (94.87)	1 (2.56)	18 (46.15)	20 (51.28)	0 (0)	2 (5.13)	37 (94.87)
Lithuania	0 (0)	14 (100)	0 (0)	0 (0)	3 (21.43)	11 (78.57)	0 (0)	8 (57.14)	6 (42.86)
Luxembourg	0 (0)	8 (22.22)	28 (77.78)	0 (0)	13 (36.11)	23 (63.89)	0 (0)	12 (33.33)	24 (66.67)
North Macedonia	2 (1.74)	53 (46.09)	60 (52.17)	2 (1.74)	26 (22.61)	87 (75.65)	2 (1.74)	8 (6.96)	105 (91.3)
Rep of Moldova	0 (0)	11 (22)	39 (78)	0 (0)	50 (100)	0 (0)	0 (0)	47 (94)	3 (6)
The Netherlands	101 (6.87)	909 (61.84)	460 (31.29)	92 (6.26)	479 (32.59)	899 (61.16)	92 (6.26)	669 (45.51)	709 (48.23)
Norway	7 (2.79)	87 (34.66)	157 (62.55)	5 (1.99)	106 (42.23)	140 (55.78)	5 (1.99)	55 (21.91)	191 (76.1)

[1] Belgium: Use of hypertonic saline, rhDNase and bronchodilators infections are not collected for transplanted patients and most of the missing data refers to this sub-population.

[2] Czech Republic: Since 2017 NaCl is prescribed more frequently; in previous years inhaled Amiloride Chloride was used more often.

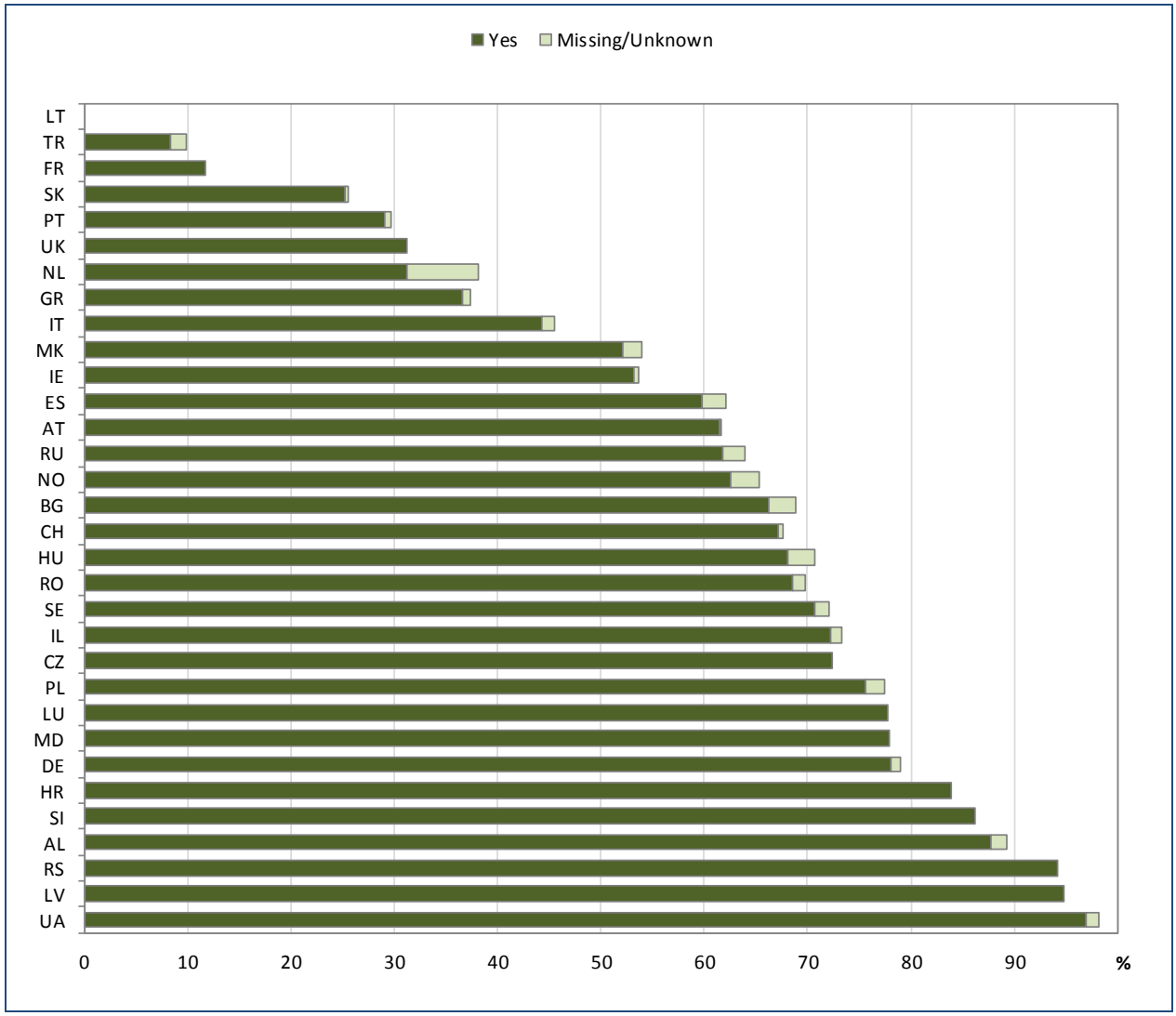
[table 7.4 continued]

Country	Hypertonic saline (NaCl) inhaled > 3 months this year number (%)			rhDNase inhaled > 3 months this year number (%)			Bronchodilators inhaled > 3 months this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Poland	12 (1.83)	148 (22.56)	496 (75.61)	12 (1.83)	135 (20.58)	509 (77.59)	14 (2.13)	103 (15.70)	539 (82.16)
Portugal	2 (0.61)	230 (70.34)	95 (29.05)	2 (0.61)	71 (21.71)	254 (77.68)	4 (1.22)	144 (44.04)	179 (54.74)
Romania	2 (1.26)	48 (30.19)	109 (68.55)	2 (1.26)	24 (15.09)	133 (83.65)	2 (1.26)	82 (51.57)	75 (47.17)
Russian Federation	63 (2.05)	1113 (36.14)	1904 (61.82)	56 (1.82)	155 (5.03)	2869 (93.15)	56 (1.82)	1408 (45.71)	1616 (52.47)
Serbia	0 (0)	10 (5.81)	162 (94.19)	0 (0)	74 (43.02)	98 (56.98)	0 (0)	1 (0.58)	171 (99.42)
Slovak Republic	1 (0.38)	198 (74.44)	67 (25.19)	1 (0.38)	101 (37.97)	164 (61.65)	1 (0.38)	108 (40.60)	157 (59.02)
Slovenia	0 (0)	15 (13.76)	94 (86.24)	0 (0)	76 (69.72)	33 (30.28)	2 (1.83)	91 (83.49)	16 (14.68)
Spain	46 (2.30)	759 (37.91)	1197 (59.79)	48 (2.40)	1295 (64.69)	659 (32.92)	44 (2.20)	652 (32.57)	1306 (65.23)
Sweden	10 (1.46)	191 (27.84)	485 (70.70)	9 (1.31)	484 (70.55)	193 (28.13)	9 (1.31)	86 (12.54)	591 (86.15)
Switzerland	4 (0.44)	296 (32.39)	614 (67.18)	3 (0.33)	511 (55.91)	400 (43.76)	3 (0.33)	147 (16.08)	764 (83.59)
Turkey	22 (1.56)	1271 (90.08)	118 (8.36)	9 (0.64)	147 (10.42)	1255 (88.94)	8 (0.57)	911 (64.56)	492 (34.87)
Ukraine	2 (1.21)	3 (1.82)	160 (96.97)	4 (2.42)	61 (36.97)	100 (60.61)	2 (1.21)	15 (9.09)	148 (89.70)
United Kingdom[1]	0 (0)	6798 (68.76)	3089 (31.24)	0 (0)	3694 (37.36)	6193 (62.64)	0 (0)	4226 (42.74)	5661 (57.26)

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

Table 7.4 shows the use of three different inhaled medications: hypertonic saline, rhDNase (Pulmozyme®) and bronchodilators (see page 15 for abbreviations).

Figure 7.6 Use of inhaled hypertonic saline in all patients seen in 2017, by country.

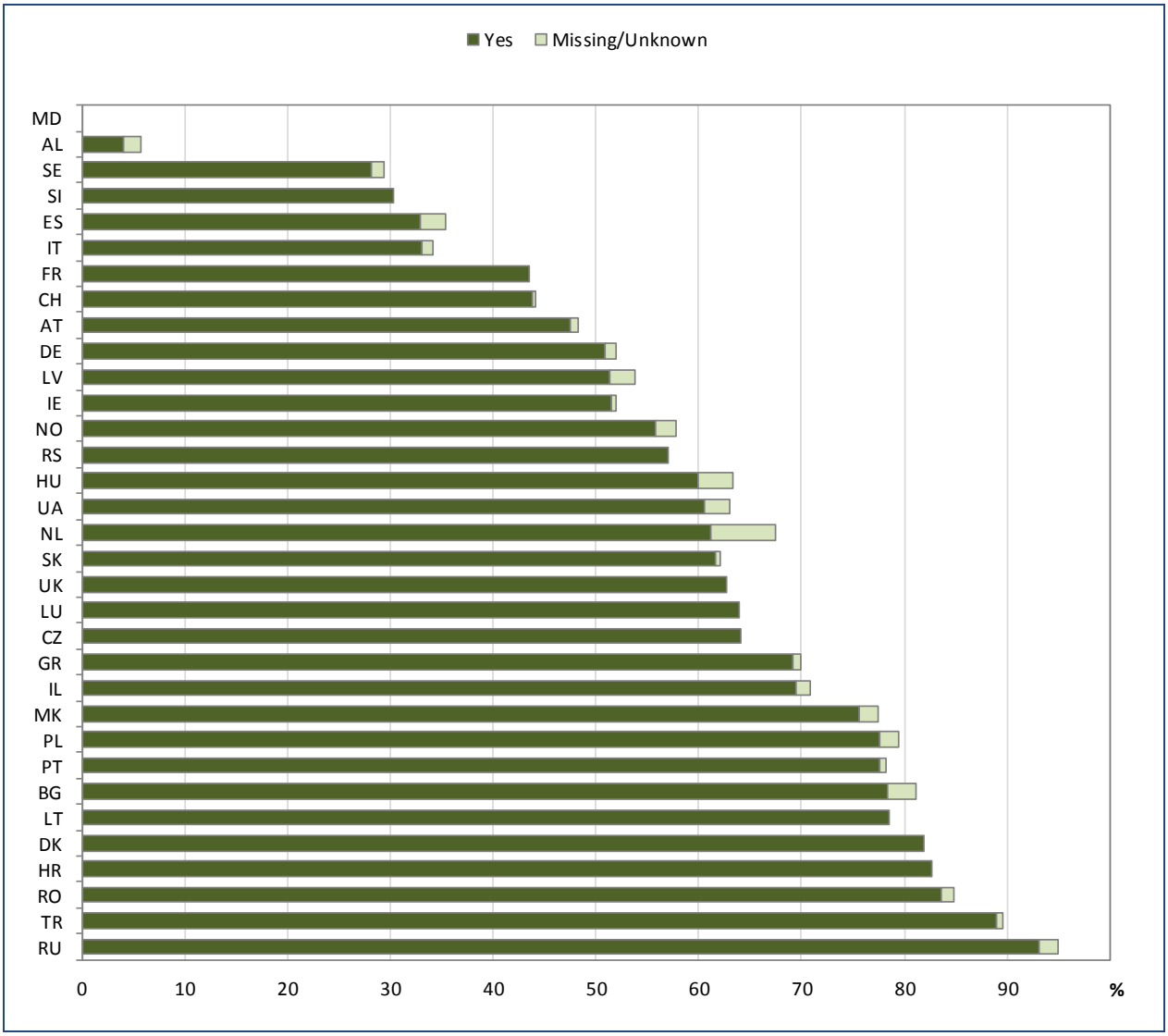


Note: We excluded from the graph the countries for which the information on inhaled hypertonic saline was missing for more than 10% of the patients.

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

This table 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.

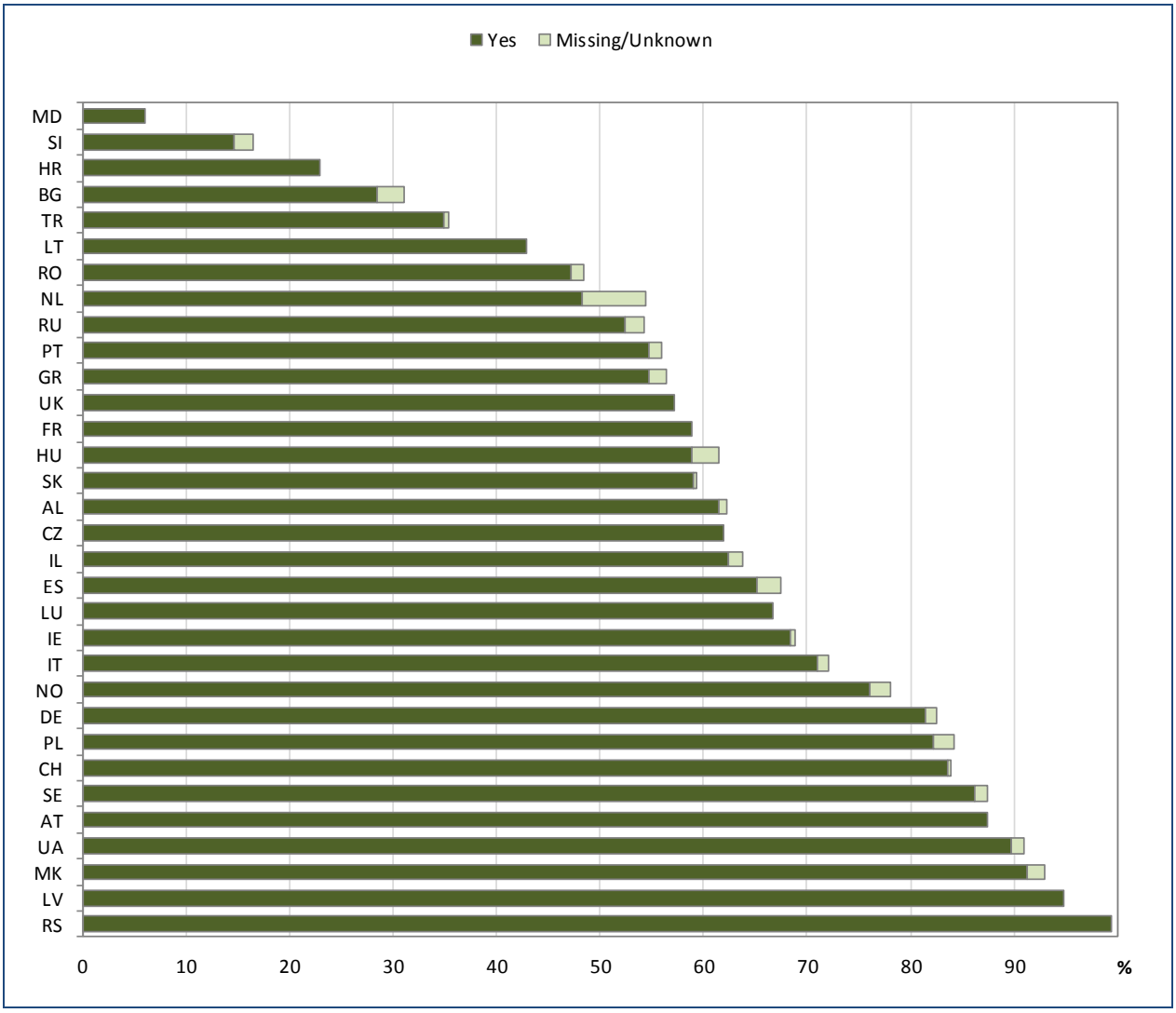
Figure 7.7 Use of rhDNase in all patients seen in 2017, by country.



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

This graph shows the use of rhDNase (marketed as Pulmozyme®) 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.

Figure 7.8 Use of bronchodilators in all patients seen in 2017, by country.



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

Note: United Kingdom: the duration of use of bronchodilators is not specified.

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.

Table 7.5 Use of inhaled antibiotics, oxygen and macrolides in all patients seen in 2017, by country.

Country	Inhaled antibiotics inhaled > 3 months this year number (%)			Oxygen therapy this year number (%)			Macrolides > 3 months this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	2 (1.64)	87 (71.31)	33 (27.05)	2 (1.64)	115 (94.26)	5 (4.1)	1 (0.82)	102 (83.61)	19 (15.57)
Armenia	7 (21.88)	18 (56.25)	7 (21.88)	6 (18.75)	24 (75.00)	2 (6.25)	6 (18.75)	14 (43.75)	12 (37.50)
Austria	1 (0.13)	494 (65.26)	262 (34.61)	0 (0)	733 (96.83)	24 (3.17)	1 (0.13)	702 (92.73)	54 (7.13)
Belgium[1]	179 (13.91)	500 (38.85)	608 (47.24)	129 (10.02)	1131 (87.88)	27 (2.10)	110 (8.55)	541 (42.04)	636 (49.42)
Bulgaria	4 (2.7)	54 (36.49)	90 (60.81)	5 (3.38)	131 (88.51)	12 (8.11)	15 (10.14)	128 (86.49)	5 (3.38)
Croatia	1 (1.15)	38 (43.68)	48 (55.17)	0 (0)	76 (87.36)	11 (12.64)	0 (0)	46 (52.87)	41 (47.13)
Czech Republic	0 (0)	454 (75.04)	151 (24.96)	0 (0)	590 (97.52)	15 (2.48)	0 (0)	542 (89.59)	63 (10.41)
Denmark[2]	496 (100)	-	-	302 (60.89)	187 (37.70)	7 (1.41)	496 (100)	0	-
France[3]	0 (0)	4170 (60.09)	2770 (39.91)	0 (0)	6626 (95.48)	314 (4.52)	0 (0)	4585 (66.07)	2355 (33.93)
Germany	59 (0.96)	3134 (51.22)	2926 (47.82)	60 (0.98)	5590 (91.35)	469 (7.66)	60 (0.98)	4956 (80.99)	1103 (18.03)
Greece	5 (0.83)	237 (39.57)	357 (59.60)	5 (0.83)	568 (94.82)	26 (4.34)	5 (0.83)	333 (55.59)	261 (43.57)
Hungary	10 (1.98)	264 (52.38)	230 (45.63)	18 (3.57)	440 (87.30)	46 (9.13)	19 (3.77)	338 (67.06)	147 (29.17)
Ireland	5 (0.41)	632 (51.85)	582 (47.74)	5 (0.41)	1100 (90.24)	114 (9.35)	5 (0.41)	573 (47.01)	641 (52.58)
Israel	8 (1.46)	222 (40.59)	317 (57.95)	7 (1.28)	527 (96.34)	13 (2.38)	11 (2.01)	279 (51.01)	257 (46.98)
Italy	62 (1.11)	3328 (59.85)	2171 (39.04)	64 (1.15)	5191 (93.35)	306 (5.50)	62 (1.11)	3799 (68.32)	1700 (30.57)
Latvia	0 (0)	26 (66.67)	13 (33.33)	0 (0)	38 (97.44)	1 (2.56)	0 (0)	35 (89.74)	4 (10.26)
Lithuania	0 (0)	13 (92.86)	1 (7.14)	0 (0)	13 (92.86)	1 (7.14)	0 (0)	13 (92.86)	1 (7.14)
Luxembourg	0 (0)	21 (58.33)	15 (41.67)	0 (0)	34 (94.44)	2 (5.56)	0 (0)	21 (58.33)	15 (41.67)
North Macedonia	2 (1.74)	68 (59.13)	45 (39.13)	2 (1.74)	112 (97.39)	1 (0.87)	2 (1.74)	92 (80.00)	21 (18.26)
Rep of Moldova	0 (0)	29 (58.00)	21 (42.00)	0 (0)	47 (94.00)	3 (6.00)	0 (0)	48 (96.00)	2 (4.00)
The Netherlands	91 (6.19)	782 (53.20)	597 (40.61)	255 (17.35)	1165 (79.25)	50 (3.40)	93 (6.33)	803 (54.63)	574 (39.05)
Norway	12 (4.78)	184 (73.31)	55 (21.91)	5 (1.99)	240 (95.62)	6 (2.39)	8 (3.19)	208 (82.87)	35 (13.94)

[1] Belgium: inhaled antibiotics is not collected for transplanted patients and most of the missing data refers to this sub-population.
[2] Denmark: the high number of missing information is due to only one of two centres reporting these data.
[3] France: collects only use of azithromycin for macrolides.

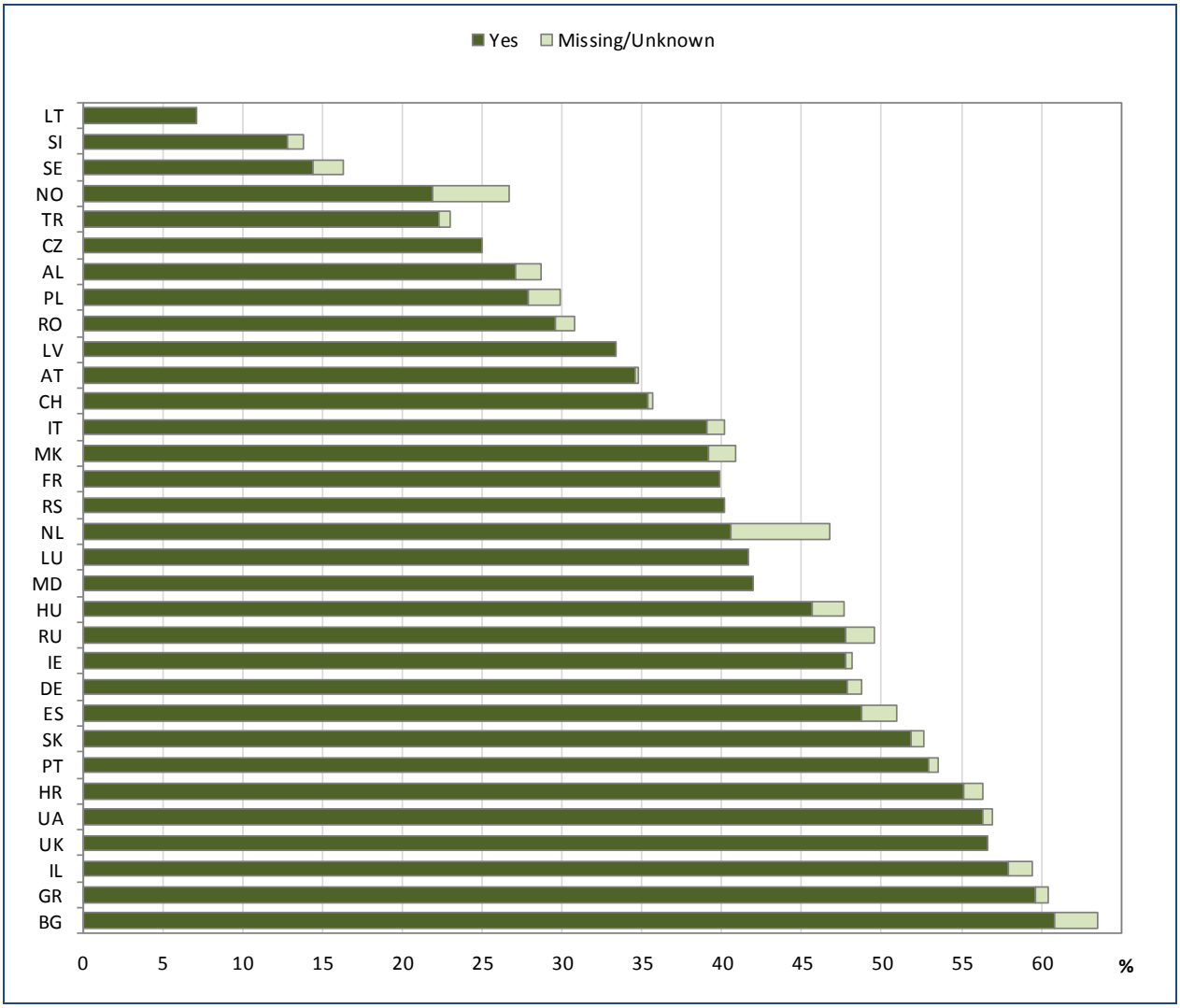
[table 7.5 continued]

Country	Inhaled antibiotics inhaled > 3 months this year number (%)			Oxygen therapy this year number (%)			Macrolides > 3 months this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Poland	13 (1.98)	460 (70.12)	183 (27.90)	12 (1.83)	624 (95.12)	20 (3.05)	15 (2.29)	538 (82.01)	103 (15.70)
Portugal	2 (0.61)	152 (46.48)	173 (52.91)	4 (1.22)	299 (91.44)	24 (7.34)	3 (0.92)	232 (70.95)	92 (28.13)
Romania	2 (1.26)	110 (69.18)	47 (29.56)	3 (1.89)	150 (94.34)	6 (3.77)	2 (1.26)	138 (86.79)	19 (11.95)
Russian Federation	55 (1.79)	1555 (50.49)	1470 (47.73)	54 (1.75)	2901 (94.19)	125 (4.06)	61 (1.98)	2110 (68.51)	909 (29.51)
Serbia	0 (0)	103 (59.88)	69 (40.12)	0 (0)	171 (99.42)	1 (0.58)	0 (0)	155 (90.12)	17 (9.88)
Slovak Republic	2 (0.75)	126 (47.37)	138 (51.88)	2 (0.75)	253 (95.11)	11 (4.14)	1 (0.38)	165 (62.03)	100 (37.59)
Slovenia	1 (0.92)	94 (86.24)	14 (12.84)	0 (0)	105 (96.33)	4 (3.67)	1 (0.92)	96 (88.07)	12 (11.01)
Spain	45 (2.25)	981 (49)	976 (48.75)	39 (1.95)	1897 (94.76)	66 (3.30)	57 (2.85)	1174 (58.64)	771 (38.51)
Sweden	13 (1.90)	574 (83.67)	99 (14.43)	10 (1.46)	659 (96.06)	17 (2.48)	16 (2.33)	479 (69.83)	191 (27.84)
Switzerland	3 (0.33)	588 (64.33)	323 (35.34)	4 (0.44)	884 (96.72)	26 (2.84)	4 (0.44)	661 (72.32)	249 (27.24)
Turkey	10 (0.71)	1087 (77.04)	314 (22.25)	11 (0.78)	1357 (96.17)	43 (3.05)	8 (0.57)	1360 (96.39)	43 (3.05)
Ukraine	1 (0.61)	71 (43.03)	93 (56.36)	1 (0.61)	147 (89.09)	17 (10.30)	5 (3.03)	17 (10.30)	143 (86.67)
United Kingdom[4]	0 (0)	4291 (43.40)	5596 (56.60)	0 (0)	9266 (93.72)	621 (6.28)	0 (0)	6070 (61.39)	3817 (38.61)

[4] United Kingdom: the duration of use of macrolides is not specified.

This table shows the use of three treatments: inhaled antibiotics for more than 3 months during the survey year (any kind); macrolides (e.g. azithromycin) for more than three months; oxygen for home treatment. Both inhaled antibiotics and macrolides are frequently used but with marked differences between countries. Oxygen is used less frequently (severe lung disease).

Figure 7.9 Use of inhaled antibiotics in all patients seen in 2017, by country.

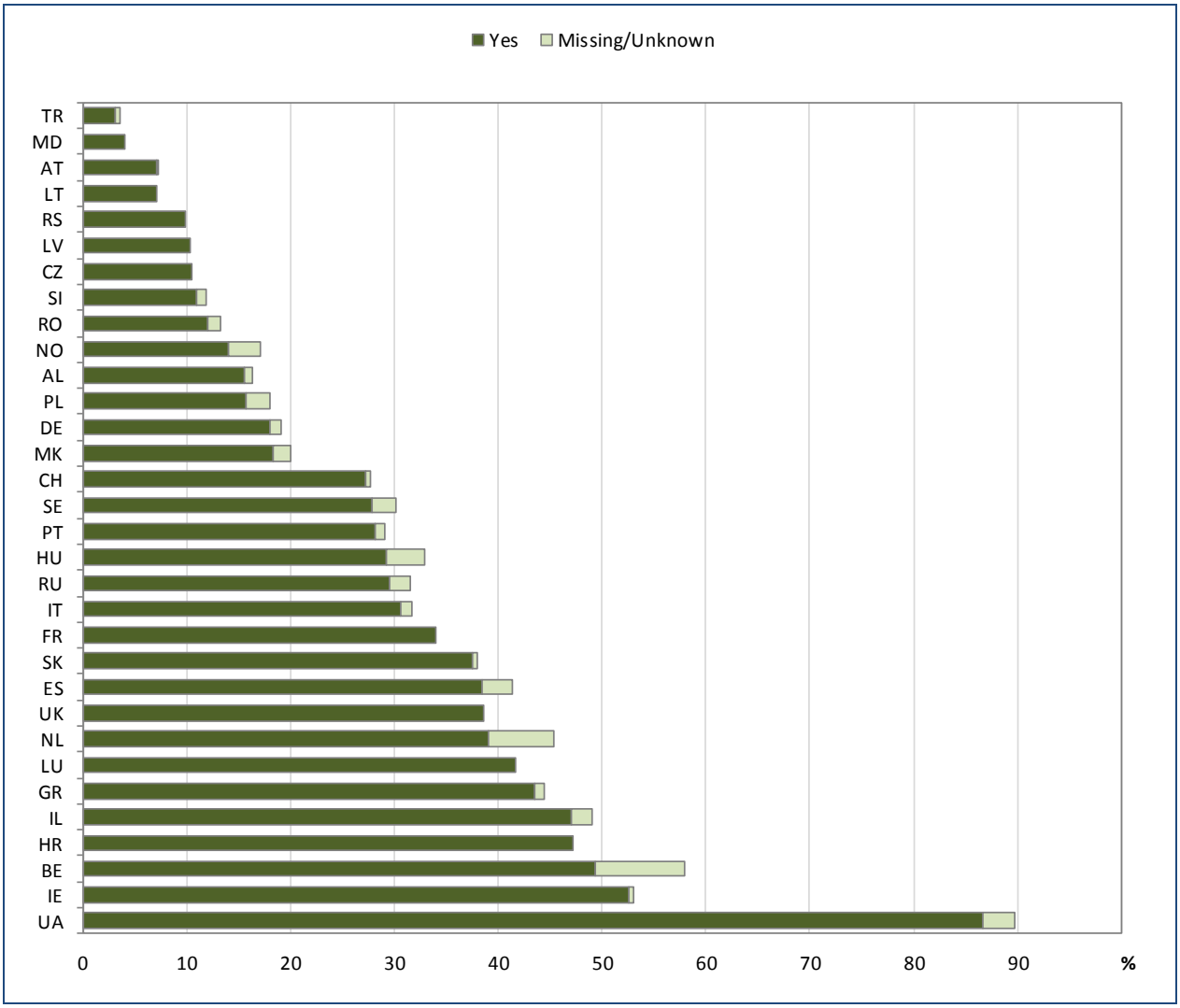


Note: We excluded from the graph the countries for which the information on inhaled antibiotics was missing for more than 10% of the patients.

Note: Belgium: inhaled antibiotics is not collected for transplanted patients and most of the missing data refers to this sub-population.

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 8 to 61%. 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 7.10 Use of macrolides in all patients seen in 2017, by country.

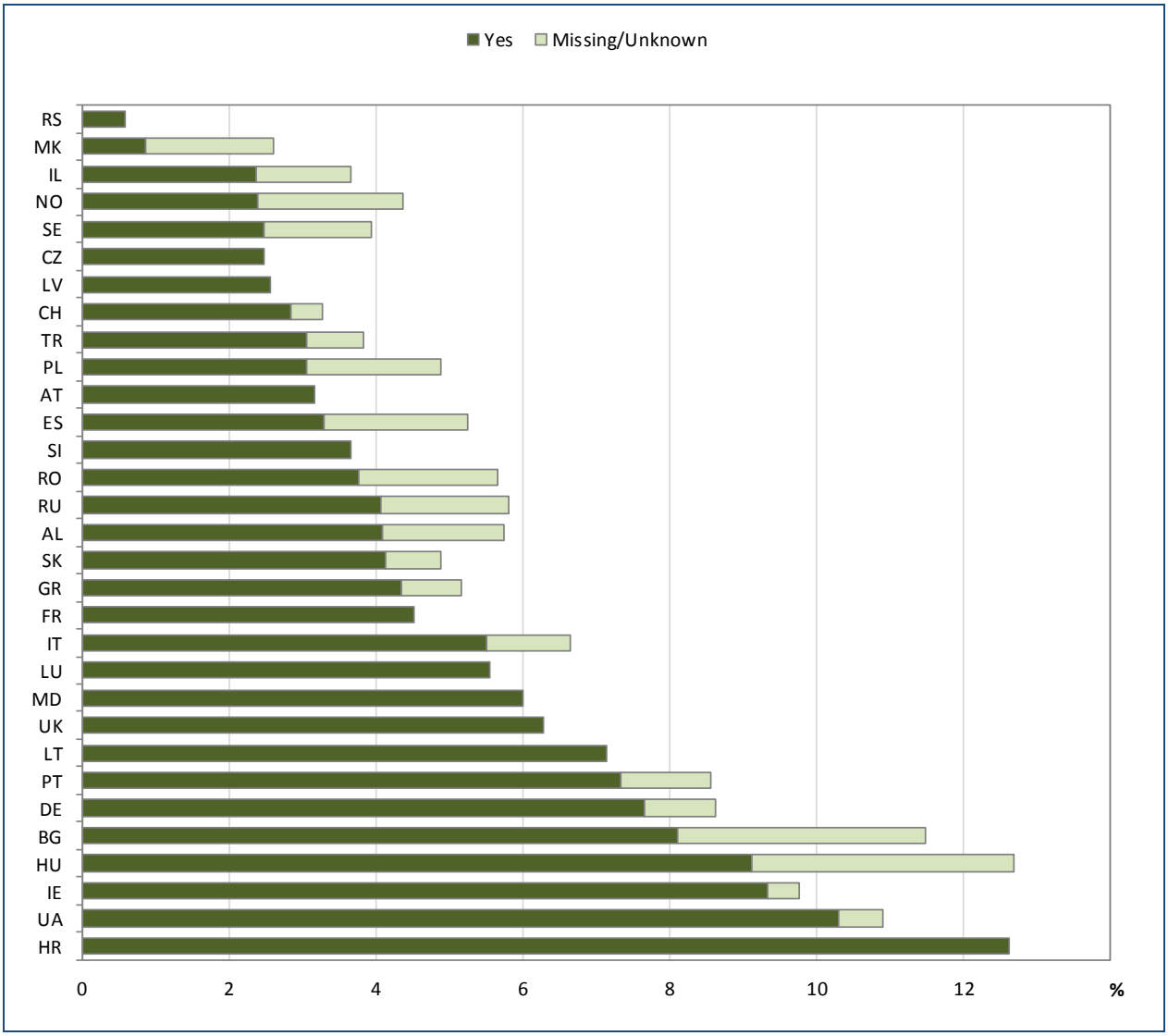


Note: We excluded from the graph the countries for which the information on use of macrolides was missing for more than 10% of the patients.

Note: France: collects only use of azithromycin for macrolides.
United Kingdom: the duration of use of macrolides is not specified.

This graph shows the use of macrolides (e.g. azithromycin) for more than 3 months during the survey year. Macrolides are antibiotics, but taken continuously they also modulate the immune system. 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 7.11 Use of oxygen in all patients seen in 2017, 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.

This graph shows the use of oxygen at home during the survey year. 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.

8. Transplantation

We ask the countries whether their patients are transplanted or not, and if they are, in which year they had their (latest) transplant.

In some countries transplanted patients are no longer registered in the CF centres' database and the CF national 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 2017 with transplanted lungs, by age and sex.

Age	Males	Females	Total	Transplants performed during the survey year
5-9	1	1	2	1
10-14	12	18	30	9
15-19	47	54	101	22
20-24	98	154	252	60
25-29	198	224	422	73
30-34	250	246	496	43
35-39	233	231	464	34
40-44	181	167	348	26
45+	273	201	474	31
Total	1293	1296	2589	299

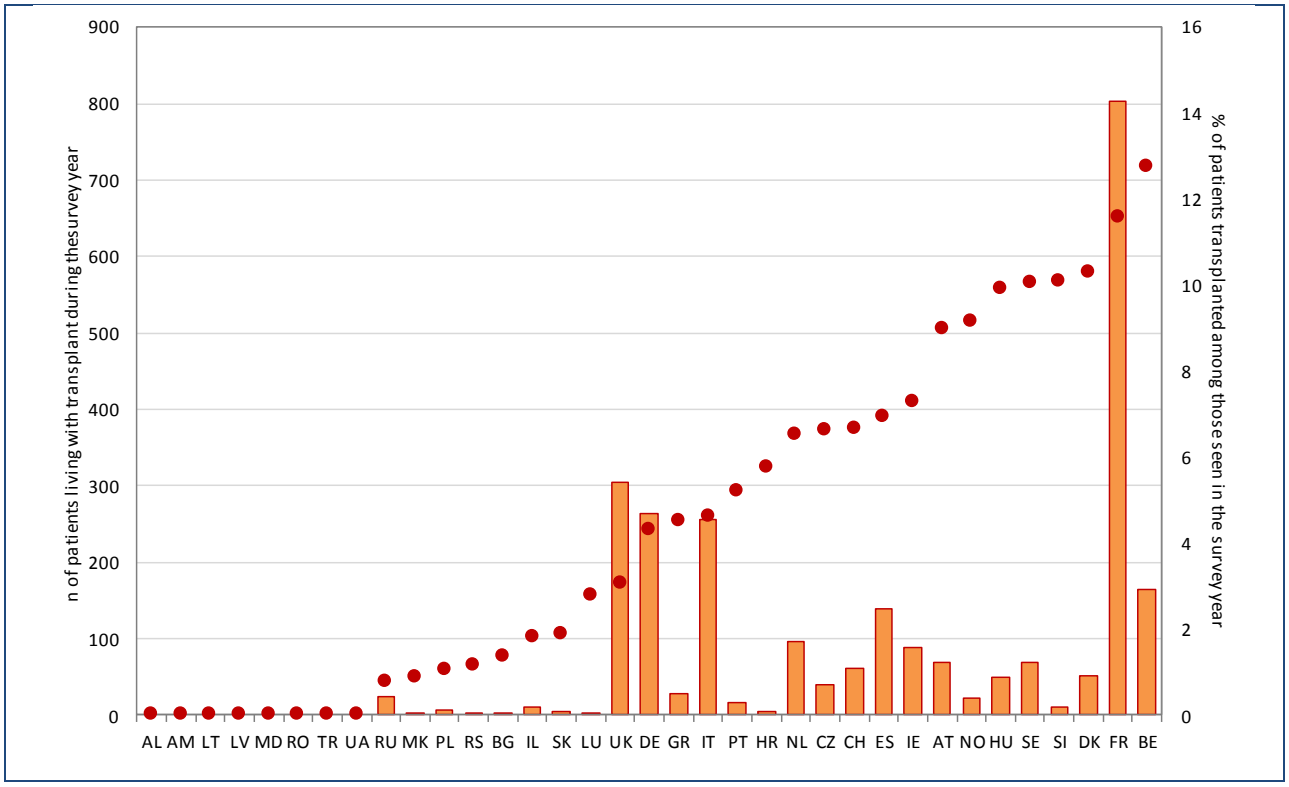
This table shows the number of patients alive in 2017 who have had a lung transplant at some time in their life, by age group, as well as the number of patients transplanted during 2017.

Table 8.2 Number of patients living in 2017 with transplanted liver, by age and sex.

Age	Males	Females	Total	Transplants performed during the survey year
0-4	1	1	2	0
5-9	4	1	5	1
10-14	8	12	20	6
15-19	29	12	41	2
20-24	28	19	47	4
25-29	29	20	49	1
30-34	35	16	51	4
35-39	20	8	28	0
40-44	9	5	14	1
45+	9	3	12	1
Total	172	97	269	20

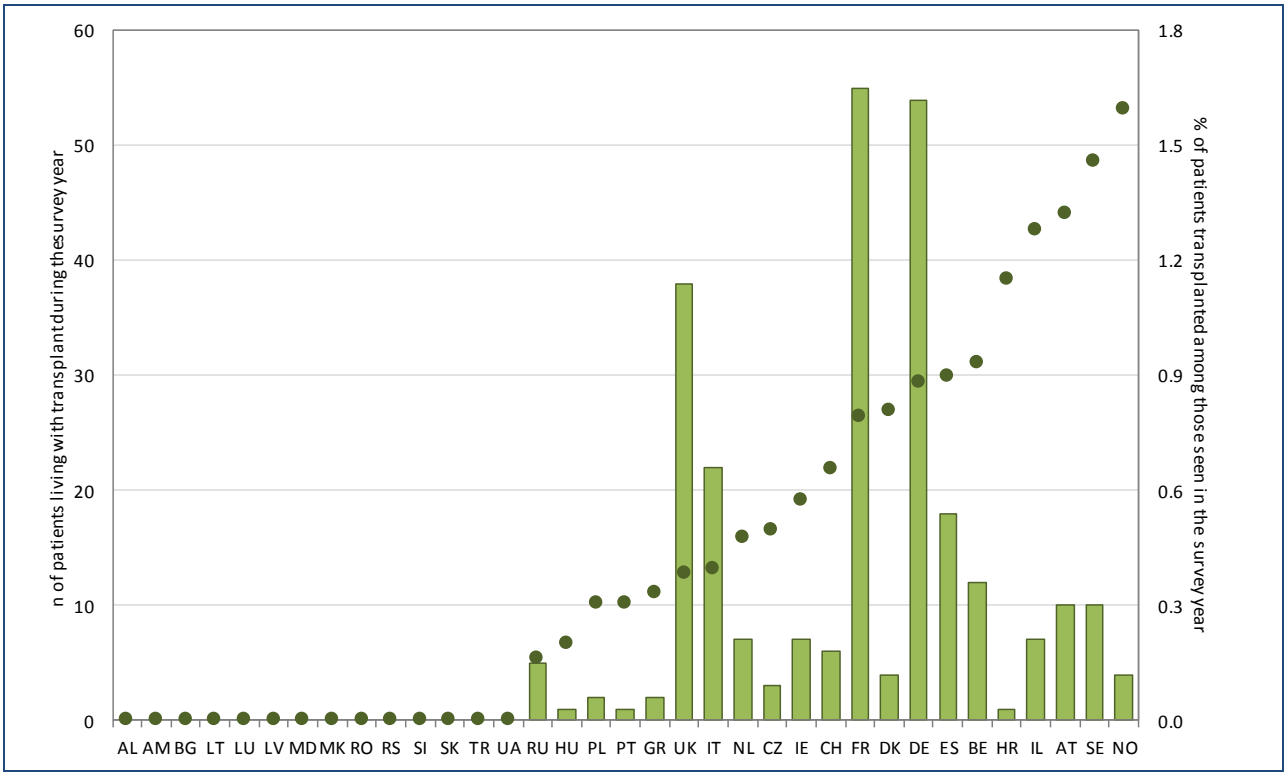
This table shows the number of patients alive in 2017 who have had a liver transplant at some time in their life, by age group, as well as the number of patients transplanted during 2017.

Figure 8.1 Number of patients living in 2017 with transplanted lungs, by country.



This graph shows the number of patients alive at 31/12/2017 who have had a lung transplant (orange bars) at some point in their life. The red dots (right axis) show the percentage of patients that are living with lung transplant in 2017 among the patients that were seen in 2017.

Figure 8.2 Number of patients living in 2017 with transplanted liver, by country.



This graph shows the number of patients alive at 31/12/2017 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 liver transplant in 2017 among the patients that were seen in 2017.

Note that on the vertical axis the number of patients with liver transplant is much lower than the number with 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 2017, by age and sex.

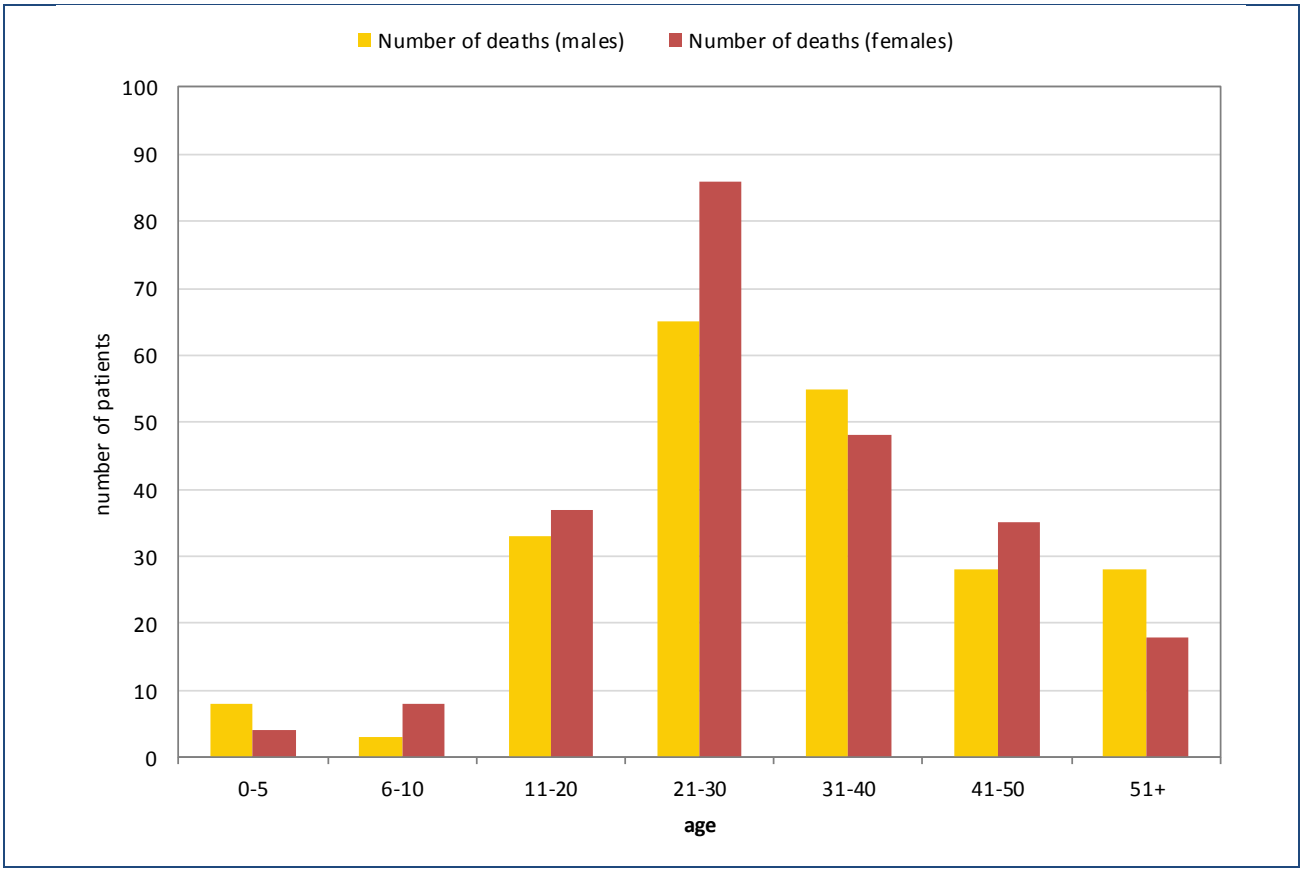
Age at death	Number of male patients	% of deaths in this age group of all male deaths	Number of female patients	% of deaths in this age group of all female deaths	Total	% Total
0-5	8	3.64	4	1.69	12	2.63
6-10	3	1.36	8	3.39	11	2.41
11-20	33	15.00	37	15.68	70	15.35
21-30	65	29.55	86	36.44	151	33.11
31-40	55	25.00	48	20.34	103	22.59
41-50	28	12.73	35	14.83	63	13.82
51+	28	12.73	18	7.63	46	10.09
Total	220	0.90	236	1.05	456	0.96

Note: For the United Kingdom, all patients with confirmed diagnosis of CF were included (N=10,468). The total number of patients presented is 47,413.
Note: For 5 male patients and 1 female patient date at death, and thereby age at death, is unknown.

This table shows the number of deaths in 2017 by age group and sex. Death in small children is very rare, and the most frequent range of age of death for both sexes is 21-30 years.

Please note: 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 2017, by sex.



Note: For the United Kingdom, all patients with confirmed diagnosis of CF were included (N=10,468). The total number of patients presented is 47,413.
Note: For 5 male patients and 1 female patient date at death, and thereby age at death, is unknown.

This graph shows the distribution of age at death of patients who died in 2017, separately by males (yellow) and females (red).

Table 9.2 Cause of death distribution of deaths in 2017.

Cause of death	Number of deaths	Percentage of all deaths
Respiratory disease	307	66.45
Transplantation related	55	11.90
Non-CF related	49	10.61
Liver-GI related	12	2.60
Trauma	3	0.65
Suicide	1	0.22
Unknown	35	7.60
Total	462	100.00

Note: Germany and the United Kingdom collect Cause of death “respiratory disease” as “cardio/respiratory”.

The table shows cause of death for the deceased patients. The most frequent cause of death is respiratory disease. Please note that only a limited number of causes of death are collected, therefore if some deaths are due to rare complications of CF, they may have been classified as “Unknown”.

Publications

The ECFSPR data has been actively used for research. Data applications are handled in accordance with the ECFSPR guidelines, for more information we refer you to the webpage www.ecfs.eu/projects/ecfs-patient-registry/data-request-application.

In the period 2011-2018 we received 69 applications for data. The majority of these requests originated from researchers (83%) from within and outside the European Cystic Fibrosis Society, and 17% of the applications derived from the Industry.

Several of these research projects have resulted in publications and others are in the pipeline. Published articles in the period 2016 – 2018 are:

- International prospective study of distal intestinal obstruction syndrome in cystic fibrosis: Associated factors and outcome.
  Munck A, Alberti C, Colombo C, Kashirskaya N, Ellemunter H, Fotoulaki M, Houwen R, Robberecht E, Boizeau E, Wilschanski M et al, on behalf of the CF/Pancreas ESPGHAN Working Group and DIOS Study Group. J Cyst Fibros 2016; 15 (4): 531-539.
  www.cysticfibrosisjournal.com/article/S1569-1993(16)00014-X/fulltext

- Epidemiology of nontuberculous mycobacteria (NTM) amongst individuals with cystic fibrosis (CF). Viviani L, Harrison MJ, Zolin A, Haworth CS, Floto RA. J Cyst Fibros. 2016; 15 (5): 619–623.
  www.cysticfibrosisjournal.com/article/S1569-1993(16)30008-X/fulltext

- Year to year change in FEV1 in patients with cystic fibrosis and different mutation classes.
  De Boeck K, Zolin A. J Cyst Fibros. 2017; 16 (2): 239-245.
  www.cysticfibrosisjournal.com/article/S1569-1993(16)30611-7/fulltext

- Effect of allergic bronchopulmonary aspergillosis on FEV1 in children and adolescents with cystic fibrosis: a European Cystic Fibrosis Society Patient Registry analysis.
  Kaditis AG, Miligkos M, Bossi A, Colombo C, Hatziagorou E, Kashirskaya N, de Monestrol I, Thomas M, Mei-Zahav M, Chrousos G, Zolin A. Arch Dis Child 2017; 102 (8): 742-747.
  https://adc.bmj.com/content/archdischild/102/8/742.full.pdf

- Creating longitudinal datasets and cleaning existing data identifiers in a cystic fibrosis registry using a novel Bayesian probabilistic approach from astronomy.
  Hurley PD, Oliver S, Mehta A. PLoS ONE; 13 (7): e0199815.
  https://doi.org/10.1371/journal.pone.0199815

- Cystic fibrosis mortality in childhood. Data from European Cystic Fibrosis Society Patient Registry. Zolin A , Bossi A, Cirilli N, Kashirskaya N, Padoan R. Int. J. Environ. Res. Public Health 2018; 15; 2020.
  www.mdpi.com/1660-4601/15/9/2020/pdf

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

The following abstracts were accepted in 2016 – 2018:

- Impact of Dornase Alfa on rate of decline in lung function in European CF patients.
  McKone EF, Kirwan L, Zolin A, Jackson A. 32nd Annual North American Cystic Fibrosis Conference, 18-20 October 2018, Denver. *Pediatric Pulmonology* 2018; 53 (S2): 354-354.
  https://onlinelibrary.wiley.com/doi/epdf/10.1002/ppul.24152

- Cystic Fibrosis Survival and Socioeconomic status across Europe.
  McKone EF, Ariti C, Jackson A, et al. *J Cyst Fibros*. 2017; 16 (S20).
  www.cysticfibrosisjournal.com/article/S1569-1993(17)30221-7/pdf

- Genetic Epidemiology of Nonsense CFTR mutations in Europe.
  McKone EF, Jackson A, Zolin A, et al. *J Cyst Fibros.* 2017; 16 (S47).
  www.cysticfibrosisjournal.com/article/S1569-1993(17)30313-2/pdf

- Guidelines to use Cystic Fibrosis registries as a tool for pharmacovigilance.
  Rens J van, McKone EF, on behalf of the ECFSPR.
  9th European Conference on Rare Diseases & Orphan Products, 10-12 May 2018, Vienna.

- The European CF Patient Registry, a useful tool for people with cystic fibrosis.
  Rens J van, McKone EF, on behalf of the ECFSPR.
  8th European Conference on Rare Diseases & Orphan Products, 26-28 May 2016, Edinburgh.

An overview of the approved applications for data, not yet associated with a publication, can be found on the website www.ecfs.eu/projects/ecfs-patient-registry/overview-data-applications.

Partners

Contributors

Appendix 1: 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.
Unknown dates of lung function tests and of height/weight measurements were set to July 1st of the survey year.
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 national registries'/individual centres' instructions. If, after the data quality controls, aberrant values were still present in the database, we set them to missing for the purposes of this report.

Reference populations used for computing z-scores

The value of a z-score depends on the reference anthropometric 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. 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 U.S. healthy individuals[1]. The choice of CDC charts as a reference, although not the most suitable to assess the nutritional status of European CF patients, is justified by the widespread use of these charts at international level.

Reference populations used for computing FEV_1 predicted values

We computed the percent of predicted values for FEV_1 and FVC using:
The multi-ethnic reference values for spirometry for the 3-95-yr age range: the global lung function 2012 equations. Eur Respir J 2012; 40: 1324–1343.

Software used for data management and statistical analyses

SAS software, Version 9.4. Copyright, SAS Institute Inc. SAS and all other SAS Institute Inc. product or service names are registered trademarks or trademarks of SAS Institute Inc., Cary, NC, USA.

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

Appendix 2: List of variables, inclusion criteria and definitions used by the ECFSPR

List of variables

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

Therapy
Inhaled continuous hypertonic NaCl this year
Inhaled continuous antibiotic this year
Inhaled continuous bronchodilators this year
In Oxygen therapy this year
Use of rhDNase this year
Use of continuous azithromycin (or other macrolide) this year
Use of ursodeoxycholic acid this year
Use of pancreatic enzymes this year

Diagnosis
Diagnosis confirmed
Age at diagnosis
Type of sweat test
Electrolytes
Chloride value
Meconium Ileus
Neonatal screening

Complications
Allergic bronchopulmonary aspergillosis this year
Diabetes: daily insulin treated this year
Pneumothorax requiring chest drain 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

Genotype
First mutation
Second mutation

Microbiology
Chronic *Burkholderia cepacia complex*
Nontuberculous mycobacteria this year
Chronic *Pseudomonas aeruginosa*
Chronic *Staphylococcus aureus*
Stenotrophomonas maltophilia this year

Follow-up
Date of best FEV_1 recorded this year
Value of best FEV_1 recorded this year
Value of best FVC recorded this year
Height measured at date of best FEV_1 (or in case of no FEV_1 last height of the year)
Weight measured at date of best FEV_1 (or in case of no FEV_1 last height of the year)

Transplant
Liver transplant
Year of latest liver transplant (if occurred before or during this year)
Lung transplant
Year of latest lung transplant (if occurred before or during this year)

Inclusion criteria

Only patients who fulfil the diagnostic criteria below should be included in the registry.

a. **Two sweat tests value > 60 mmol/L chloride**: CF diagnosis accepted
b. **One sweat test value > 60 mmol/L chloride and DNA Analysis/Genotyping – two identified disease causing CF mutations**: CF diagnosis accepted
c. **Sweat value less than or equal to 60 mmol/L chloride**: if the sweat value is less than or equal to 60 mmol/L chloride, then at least 2 of these should be met:
    i. DNA Analysis/Genotyping – two identified disease causing CF mutations.
    ii. Transepithelial (Nasal) Potential Difference – study consistent with a diagnosis of CF.
    iii. Clinical Presentation – typical features of CF.
d. **Diagnosis reversal**: if the patient's CF diagnosis reversed during the year, identify the reason from the following options:
    i. DNA Analysis – unable to identify two disease causing CF mutations.
    ii. Transepithelial (Nasal) Potential Difference – study not consistent with a diagnosis of CF.
    iii. Repeat normal sweat testing – confirm with clinical team.

Definitions for EFCSPR

SWEAT TEST

If a sweat test was not performed on a patient, record "not done". If a sweat test is "not done" then two known genotype mutations must be reported.

i. Sweat Test: record the patient's sweat test.
ii. Electrolytes: Chloride concentration measurement is the preferred analysis.
iii. Chloride value: report the Chloride value in millimols per litre (mmol/L). If duplicate tests were completed on the same day, report the highest positive value.

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.

SPIROMETRY

The purpose of recording data on spirometry values for the ECFS Patient Registry is to obtain standardised comparable data for comparison with other centres/countries and for use in specific epidemiological studies. Some of the conditions for this (see below) may not be met at every clinic visit for all patients. Therefore, for the purpose of the registry, only the spirometry tests fulfilling the criteria should be recorded/extracted for the ECFS Patient Registry. For all tests the spirometry should be performed according to the common ATS/ERS guidelines: (www.thoracic.org/statements/resources/pfet/PFT2.pdf).

Furthermore for the values reported to the registry the following criteria should be met

1. Pre-test:
    a. date of birth, gender and height should be recorded for calculation of predicted values
    b. all recorded spirometry tests should be pre-bronchodilator* values
        i. short-acting bronchodilators: at least 4 hours pre-test
        ii. long-acting bronchodilators: at least 12 hours pre-test

    *This was decided according to the PortCF official definitions.
2. Reported values:
    a. for values reported to national registries or to centres and extracted to the ECFS Patient Registry, the value in litres of the highest available value of FEV_1% of predicted (according to local references) of the year should be extracted
    b. each patient's FVC and FEV_1 measurement must be reported in litres (L), with up to two places to the right of the decimal
    c. the FVC measurement must be greater than or equal to the FEV_1 measurement
    d. for each reported spirometry value, the date of the test and the patient's height at that date should be reported in order to perform the calculation of percent of predicted values
    e. only tests deemed valid according to ATS/ERS guidelines should 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. (Multi-ethnic reference values for spirometry for the 3-95-yr age range: the global lung function 2012 equations. Eur Respir J 2012; 40: 1324–1343).

The ECFSPR Definition Group considered the issue of race-specific reference values and decided not to do this calculation and not to record race for European patients.

References:
a) Miller et al. Standardisation of spirometry. Eur Respir J 2005; 26: 319–338
b) Miller et al. General considerations for lung function testing. Eur Respir J 2005; 26: 153–161
c) Cystic Fibrosis Foundation Patient Registry User's Guide, Version 4.0. 2006
d) Rosenfeld et al. Task Force to Evaluate Choice of Spirometric Reference Equations for the National Patient Registry: Summary and Recommendations. Cystic Fibrosis Foundation Registry Committee; 2005
e) Hankinson JL, Odencrantz RJ, Fedan KB. Spirometric reference values from a sample of the general U.S. population. Am J Respr Crit Care Med 1999;159:179-87
f) Wang X, Dockery DW, Wypij D, Fay ME, Ferris BG. Pulmonary function between 6 and 18 years of age. Pediatr Pulmonol 1993;15:75-88.

NUTRITION

Measurements: weight and height are measured according to 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. it should be the value at the day of the recorded FEV_1

z-scores for height, weight and BMI will be calculated using the CDC reference values [Kuczmarski et al (2002)]

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

DEFINITION OF CHRONIC INFECTION IN THE LOWER AIRWAYS

1. Chronic PA infection should be defined by local physician according to modified Leeds criteria[a] and/or anti-pseudomonas antibodies[b]. 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 the status has changed:
2. modified Leeds criteria, chronic infection: >50% of respiratory samples collected during the last 12 months are positive. At least 4 samples during that period;
3. and/or significantly raised anti-pseudomonas antibodies according to local laboratories.
4. Chronic infection with other gram-negative bacteria should be recorded by the same criteria as above.

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

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

LIVER DISEASE

We adopt the definitions for Liver Disease used by the UK Registry. These definitions discriminate patients with severe liver disease (with portal hypertension) from milder cases (cirrhosis without portal hypertension).

Cirrhosis with Hypertension: scaring 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: scaring of the liver relating to underlying CF.

Liver disease without cirrhosis: this includes fatty liver or viral hepatitis but not biliary cirrhosis.

PANCREATIC STATUS

Definition:

Stool fat (van de Kamer) > 4-5 g/d in young children, > 7g/d in children above 10 yrs and adults and/or faecal pancreatic elastase-1 < 200 ug/g.

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

Pancreatic status will be assessed at the registry level, according to the following:

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)*

*according to definition above

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