2018

ECFS Patient Registry
Annual Data Report

European Cystic Fibrosis Society
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ECFS Patient Registry

Annual Data Report

2018 data

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Preface

We are pleased to share with you the 2018 Annual Report from the European Cystic Fibrosis Society Patient Registry (ECFSPR). This 14th report contains demographic and clinical data of 49,886 consenting CF patients from 38 countries. The epidemiological data is provided by national cystic fibrosis (CF) registries and individual CF centres throughout Europe and neighbouring countries. Due to problems with data collection as a result of the COVID-19 pandemic, the data-set for France is missing in this report, and demographic data from 2017 has been included instead.

It is the ECFSPR's mission to provide a reliable and comprehensive picture of clinical outcomes in CF across Europe that will help enhance quality of CF care in Europe and worldwide. In recent years, the number of participating countries and centres has increased steadily and there are few regions in Europe that are not yet involved; we are making every effort to include them in future data collections and analyses. Our strong collaboration with CF Europe and national patient organisations guarantees that the ECFSPR data is helpful for the community and we are grateful to all people with CF, and their families, throughout Europe, for their willingness to participate in the Registry.

The 2018 data has been collected with a technologically updated software, ECFSTracker. We increased the number of data variables collected to include useful information on CFTR modulator therapy, steroids, DIOS (distal intestinal obstruction syndrome), MRSA (multi-resistant Staphylococcus aureus) and days on intravenous antibiotics at home and in hospital (for more detailed information on the new variables, please refer to the List of Variables and Definitions in Appendix 2, page 168). In addition, we collected specific information from each country regarding the status of licencing and reimbursement of CF-specific pharmacotherapies. This information should help the reader to interpret correctly the country-specific differences in the use of therapies, with particular regard to the availability of rhDNAse and CFTR modulators.

The analyses presented in this report have been carried out by the ECFSPR statisticians using the 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 variations 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 168).

To ensure the highest quality data is collected the ECFSPR has implemented an ongoing data quality project: on-site data validation visits to assess data accuracy and completeness and identify areas for improvement are carried out in participating CF centres and at National Registries. Outcomes from the visits help us to work together with the centres and countries to optimise how source data is documented and to enhance quality in a sustainable manner. This is of paramount importance not only with regard to the validity and acceptance of the ECFSPR as a data source for scientific publications, but it is also mandatory in this era of registry-based pharmacovigilance studies which will continue to evolve in the upcoming years. In 2018 the European Medicines Agency (EMA) qualified the ECFSPR and its data collection software, ECFSTracker, as a platform and data source for post-authorisation safety (PASS) and efficacy (PAESS) studies. This is an important milestone in the field of registry-based clinical research; it opens new horizons for the ECFSPR and brings opportunities for collaboration with the EMA and the pharmaceutical industry that will employ high quality post-marketing data from the largest CF data-set in the world to monitor new drugs that come onto the market.

The countries and national registries that participate in the ECFSPR, represented by their country coordinators, patients, and their families, have dedicated their valuable time and resources to the Registry. They are supported by the patient organisations, the ECFSPR staff, the Executive and Scientific Committees and a vast number of voluntary working group members of the many projects undertaken by the ECFSPR. I would like to

thank all of these stakeholders for their huge efforts in making the Registry an important and invaluable database with regard to CF care. Finally, we are thankful for all our sponsors and supporters who provide the financial background to make the ECFSPR possible.

Sincerely,

Andreas Jung, ECFSPR Director

To the people with cystic fibrosis

This report is about you and how cystic fibrosis (CF) affects you and other people all over Europe. The report is based on information collected by individual CF centres and the national CF registries that participate in the European Cystic Fibrosis Society Patient Registry (ECFSPR). We have tried to make the presentation of this data as clear as possible and hope that you will find the report interesting and easy to understand.

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

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

The Registry's presence on social media is increasing, since we launched a Facebook account www.facebook.com/EuropeanCysticFibrosisPatientRegistry/ in June 2019 and a Twitter account @ECFSRegistry in 2018. News, updates and other interesting information is posted regularly on both media.

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 Registry please 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 the data

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

In larger print: the name of the country representative in the ECFSPR Steering Group;
Underlined: the name of the database manager for the national registry;
In Italics: new participants with 2018 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	14 individual centres:	Andreas Pfleger
	Medizinische Universität Graz, Universitätsklinik für Kinder- und Jugendheilkunde, Klinische Abteilung für Pädiatrische Pulmonologie und Allergologie und CF Zentrum für Kinder, Jugendliche und Erwachsene, Graz	Ernst Eber Andreas Pfleger Maria Gaber
	Medizinische Universität Innsbruck, Zertifiziertes CF Zentrum für Kinder, Jugendliche und Erwachsene, Innsbruck	Helmut Ellemunter Johannes Eder
	Klinikum Klagenfurt am Wörthersee, Abteilung für Kinder- und Jugendheilkunde, Pädiatrische Pulmologie/ Allergologie, Klagenfurt	Franz Hubert Wadlegger
	Univ. Klinik für Kinder- und Jugendheilkunde, Kepler Universitätsklinikum, Linz	Adrienne Molnar Julia Pichler Christina Thir
	Kepler Universitats Klinikum, Klinik für Lungenheilkunde/ Pneumologie, Linz	Martin Stadlinger Viktoria Reinelt Katrin Scheich
	Kardinal Schwarzenberg Klinikum, Abteilung für Kinder- und Jugendmedizin, Schwarzach im Pongau	Josef Riedler Christoph Seelbach
	Salzburger Landeskliniken, Universitätsklinik für Pneumologie, Salzburg	Michael Studnicka Natalie Firlei-Fleischmann
	Landeskrankenhaus Steyr, Abteilung für Kinder- und Jugendheilkunde und Abteilung für Lungenheilkunde, Steyr	Alexander Ebner Margit Kalinger Monika Pell
	Medizinische Universität, Allgemeines Krankenhaus Wien, Universitätsklinik für Chirurgie, Klinische Abteilung für Thoraxchirurgie, Vienna	Peter Jaksch Dagmar Liebhart
	Medizinische Universität Wien, 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

Country	Centre/National Registry name	Contact
	Klinikum Wels-Grieskirchen, Abteilung für Kinder- und Jugendheilkunde, Wels	Franz Eitelberger Beatrix Wintersteiger Vera Karin Bauer
	Klinikum Wels-Grieskirchen, Abteilung für Lungenkrankheiten, Wels	Alexander Leitner Andrea Ringl
Belarus	*Belarusian Republic Children's Center of Pulmonology and Cystic Fibrosis, Pulmonary Department, 3rd City Children's Clinical Hospital, Minsk*	*Vladimir Bobrovnichiy* *Svetlana Keegan*
Belgium	Belgian Cystic Fibrosis Registry	Géraldine Daneau Simeon Wanyama
Bulgaria	2 individual centres:	Guergana Petrova
	Alexandrovska University Hospital, Pediatric Clinic, Sofia	Guergana Petrova
	University Hospital St. Marina, 2nd Paediatric Clinic, Varna	Miglena Georgieva Nataliya Dobrudzhanska Margarita Nikolova Ruzha Pancheva
Croatia	1 individual centre:	Duska Tjesic-Drinkovic Andrea Vukic Dugac
	University Hospital Centre Zagreb, Cystic Fibrosis Centre – Paediatrics and Adults, Zagreb	Duska Tjesić-Drinković Dorian Tjesić-Drinković Andrea Vukic Dugac
	On behalf of the Croatian CF Patient Database	Ivan Bambir Ivona Markelic
Cyprus	*Medical School, University of Cyprus (children and adults)*	*Panayiotis Yiallouros* *Andreas Matthaiou* *Panayiotis Kouis* *Pinelopi Anagnostopoulou*
Czech Republic	Cystic Fibrosis Registry of the Czech Republic	Pavel Drevinek Alena Bilkova Milan Macek Marek Turnovec
Denmark	Cystic Fibrosis Registry Denmark	Hanne Vebert Olesen Tania Pressler
France	Registre Français de la Mucoviscidose	Lydie Lemonnier Clémence Dehillotte
Germany	German Cystic Fibrosis Registry	Lutz Nährlich Sylvia Hafkemeyer Julia Wosniok
Greece	3 individual centres:	Elpis Hatziagorou
	Aghia Sophia Children's Hospital, CF Centre, Athens	Athanasios Kaditis Ioanna Loukou Argyri Petrocheilou
	Sismanoglio General Hospital of Attica, Adult Cystic Fibrosis Unit, Athens	Filia Diamantea Irene Bourgani
	Aristotle University of Thessaloniki, Hippokration General Hospital, Cystic Fibrosis Centre, Thessaloniki	John Tsanakas Elpis Hatziagorou Maria Fotoulaki John Kioumis Aikaterini Manika

Country	Centre/National Registry name	Contact
Georgia	*I. Tsitsishvili Children's Clinic, CF Centre, Tblisi*	*Ia Khurtsilava* *Tsitsino Parulava*
Hungary	Cystic Fibrosis Registry of Hungary	Andrea Párniczky Géza Marsal Attila Hornyák-Kovács
Ireland	Cystic Fibrosis Registry of Ireland	Godfrey Fletcher Laura Kirwan
Israel	Cystic Fibrosis Registry of Israel	Meir Mei-Zahav
Italy	Italian Cystic Fibrosis Registry	Rita Padoan Marco Salvatore Annalisa Amato Gianluca Ferrari
Latvia	1 individual centre: Rīga Stradinš University, Children's Clinical University Hospital, Department of Pneumology, Riga	Elina Aleksejeva Elina Aleksejeva Liga Berke
Lithuania	2 individual centres: Hospital of theLithuanian University of Health Sciences Kauno Klinikos, Adult Cystic Fibrosis Centre, Kaunas *Hospital of Lithuanian University of Health Sciences Kauno Klinikos, Centre of Pediatric Chronic Respiratory Diseases, Kaunas*	Kęstutis Malakauskas Kęstutis Malakauskas Virginija Kalinauskaitė - Žukauskė *Valdone Misevičiene*
Luxembourg	1 individual centre: Centre Hospitalier de Luxembourg	Marc Schlesser Marc Schlesser Elisabeth Da Silva
Rep. of North Macedonia	2 individual centres: University Children's Hospital, Centre for Cystic Fibrosis, Skopje *Institute for respiratory diseases in children, Kozle, Center for cystic fibrosis, Children and adults*	Stojka Fustik Stojka Fustik Ana Stomatova *Tatjana Jakovska- Maretti* *Ivana Arnaudova*
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	Lukasz Wozniacki Radoslawa Staszak – Kowalska Mikolaj Kowalski Maria Trawinska-Bartnicka Ewa Sapiejka Grzegorz Gaszczyk Monika Rams Stanislaw Stepniewski Daria Dziecichowicz-Latala

Country	Centre/National Registry name	Contact
Poland (cont.)	University Hospital of Lords Transfiguration, Dept. of Pulmonology, Allergology and Pulmonary Oncology, Poznan	Szczepan Cofta Agata Nowicka
	Karol Jonscher University Hospital of Poznan University of Medical Sciences, Poznan	Irena Wojsyk-Banaszak
	Institute of Tuberculosis and Lung Diseases, Rabka-Zdrój Branch, Dept. of Pneumology and Cystic Fibrosis, Rabka Zdroj	Henryk Mazurek Lidia Pawlik
	Provincial Clinical Hospital no. 2, Dept of Allergology and Cystic Fibrosis, St Jadwigi Krolowej in Rzeszow	Krzysztof Balata Marta Rachel
	Dziekanow Paediatric Hospital, Cystic Fibrosis Centre, Institute of Mother and Child, Warsaw	Dorota Sands Lukasz Wozniacki
Portugal	Cystic Fibrosis Registry of Portugal	Luísa Pereira
Romania	6 individual centres:	Liviu Pop
	Regional Cystic Fibrosis Centre Brasov, Brasov	*Laura Larisa Dracea*
	Clinical Children's Hospital Grigore Alexandrescu, Bukarest	Simona Mosescu
	Mother & Child Health Institute, Bukarest	Suciu Nicolae Iustina Stan
	Regional Cystic Fibrosis Centre Cluj, Cluj-Napoca	Radu Sorin Şerban Szabo Csilla-Enikő
	"S.F. Maria" Children's Emergency Hospital, Iasi	*Dana Anton-Paduraru*
	National Cystic Fibrosis Centre, 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, Detská fakultná nemocnica Košice, Košice	Anna Fetekeova Zuzana Hribíková
Slovenia	3 individual centres:	Uroš Krivec
	University Clinic of Pulmonary and Allergic Diseases, Golnik	Matjaž Fležar Tjaša Brus Pičman Julij Šelb

Country	Centre/National Registry name	Contact
Slovenia (cont.)	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	24 individual centres:	M Dolores Pastor-Vivero
	Parc Taulí Hospital Universitario, Hospital de Sabadell, Unitat de Pneumologia Pediátrica i Unitat de Fibrosi Quística, Sabadell, Barcelona	Oscar Asensio de la Cruz Miguel Garcia Gonzàlez Xavier Pomares Amigó Concepción Montón Soler
	Hospital Sant Joan de Déu, Unitat de Pneumologia Pediátrica i Fibrosi Quística, Barcelona	María Cols Roig Jordi Costa i Colomer
	Hospital Universitari Vall d'Hebron, Adult Cystic Fibrosis unit, Barcelona	*Antonio Alvarez Fernández*
	Hospital Vall d'Hebron, Unidad Fibrosis Quística y Neumología 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
	Hospital Universitario La Paz, Unidad de Fibrosis Quìstica Adultos, Servicio de Neumología, Madrid	Concha Prados
	Hospital Infantil La Paz, Sección de Neumología Pediátrica, Unidad de Fibrosis Quística Pediátrica, Madrid	María Isabel Barrio Gomez de Agüero Marta Ruiz de Valbuena Maiz
	Hospital Universitario La Princesa, Neumología Adultos, Madrid	Rosa María Girón
	Hospital Niño Jesús, Sección de Neumología Pediátrica, Unidad de Fibrosis Quística, Madrid	José R. Villa Asensi 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 Fibrosis Quística, Oviedo	José Ramón Gutiérrez Martínez Marta Garcia Clemente
	Hospital Universitari Son Espases, Servicio de Neumología y Servicio de Pediatría, Unidad de Neumología y Alergia Pediátrica, Palma de Mallorca	*Alexandre Palou-Rotger* *Catalina Bover-Bauza* *Joan Figuerola Mulet* *Margalida Barceló Bobillo*

Country	Centre/National Registry name	Contact
Spain (cont.)	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 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 M Dolores Pastor-Vivero
	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
	Kantonsspital Aarau AG, Klinik für Pneumologie und Schlafmedizin, Aarau	Sarosh Irani G. Mauro Tini Lydia Eisenmann
	Universitätsspital Basel, Klinik für Pneumologie, Adulte Cystische Fibrose, Basel	Michael Tamm Kathleen Jahn
	UKBB Universitäts-Kinderspital beider Basel, Abteilung Intensivmedizin & Pneumologie, Basel	Jürg Hammer Daniel Trachsel Anja Jochmann
	Inselspital, Universitätsklinik für Pneumologie, Abteiling 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 Nadège Gabent
	Hôpitaux Universitaires de Genève, Département de Médecine, Service de Pneumologie, Consultation de Mucoviscidose Adulte, Genève	Paola Gasche Jérôme Plojoux

Country	Centre/National Registry name	Contact
Switzerland (cont.)	Centre Hospitalier Universitaire Vaudois (CHUV), Département femme-mère-enfant, Service de pédiatrie, Unité de pneumologie et mucoviscidose pédiatrique, Lausanne	Isabelle Rochat Laurence Mioranza
	Consultation Adulte de Mucoviscidose Service de Pneumologie, Département de Médecine, Centre Hospitalier Universitaire Vaudois (CHUV), Lausanne	Isabelle Huart Bellavere Caroline Dutoit
	Luzerner Kantonsspital, Zentrum für Zystische Fibrose für Kinder und Jugendliche, Luzern	Nicolas Regamey Michael Hitzler Marco Lurà
	Luzerner Kantonsspital, Abteilung für Pneumologie, Luzern	Christian Murer Natascha Sidler
	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, Zentrum für Cystische Fibrose für Erwachsene, St. Gallen	Martin Brutsche Otto Schoch Anna-Lena Walter
	Kantonsspital Winterthur, Klinik für Pneumologie und Klinik für Innere Medizin, Adulte Cystische Fibrose, Winterthur	Markus Hofer Sieghart Filippi
	Universitäts-Kinderspital Zürich, Abteilung für Pneumologie, Zürich	Andreas Jung Alexander Möller Demet Inci
	Universitätsspital Zürich, Klinik für Pneumologie, Adultes CF Zentrum, Zürich	Christian Benden Thomas Kurowski
Turkey	Cystic Fibrosis Registry of Turkey	Deniz Dogru
	Marmara University Faculty of Medicine, Division of Pulmonology, Istanbul	Bülent Karadağ Yasemin Gökdemir Ela Eralp
	Medipol University Faculty of Medicine, Division of Pediatric Pulmonology, Istanbul	*Sedat Öktem* *Füsun Ünal*
Ukraine	1 individual centre: Cystic Fibrosis Centre of Western Ukrainian Specialized Children's Medical Centre, Lviv	Halyna Makukh Lyudmyla Bober Halyna Makukh
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:

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Contributing country managers and national representatives (the names are listed on page 5-11);
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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 pseudonymised, and only year/ month of birth and randomised centre and patient 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) give or deny their approval for the use of the data from their country; this decision is final. Requests originating from Industry are also reviewed by the ECFS Clinical Trials Network. All applications must meet the European and individual country data protection legislation regarding patient anonymity.

For more information, please visit our website www.ecfs.eu/ecfspr.

General Considerations

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, because either some types of information are not collected, or they are collected by the national registry using a different method. When the national registries upload their data they are also asked to state whether their variables definitions meet those of the ECFSPR or not. Where major discrepancies between the definitions occur, those variables have been omitted from the annual report for that country; in the case of minor discrepancies an explanatory footnote has been added to the graphs and tables. For example, the ECFSPR collects information on the presence of chronic *Pseudomonas aeruginosa* infection according to the modified Leeds criteria, and/or the presence of elevated *Pseudomonas* antibodies (see Appendix 2 on page 168). If a national registry collects such information as “at least one positive *Pseudomonas aeruginosa* culture this year”, this information would be too different from the ECFSPR definition of chronic *Pseudomonas aeruginosa*, and we would set this variable to “missing” for that particular country. If, instead, a country defines chronic *Pseudomonas aeruginosa* as “the presence of more than four positive cultures in 6 months”, the data of this variable would be included in the annual report since the definition is much closer to the ECFSPR definition and a footnote would be added to the relevant tables and graphs.

If a country does not collect a certain variable (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; all of the data, however, is presented in the report. The same applies for countries where the information for a variable is missing for more than 10% of the patients. The countries with less than 5 patients in an age group (e.g.

less than 5 adults) are excluded both from the graphs and from 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 will be anything between 7 and 27%.

You will find some differences between the findings of the national registries' own reports and the ECFSPR report. This is because some variable values are recoded or computed in different ways. For example, some national registries compute the age of the patient at the date of the annual visit and consider 16 years as the cut-off for adult age. The ECFSPR computes the age at FEV1/height/weight measurement and the age at follow-up (the end of the year) and considers 18 years as the cut-off for adult age. Since clinical outcomes do not change very much over a 12-month period we do not consider this to be a serious obstacle to interpretation. Another example: for lung function values such as FEV1 the raw data values, reported in litres, are not informative unless they are expressed in relation to the age, sex and height of the patient. We therefore needed to transform the raw values into new variables in order to compare lung function between 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 coverage, i.e. the percentage of people with CF included in the national registry or national data presented by the country, varies; see table 1.1, page 19. These differences can influence how the data is interpreted, and we therefore advise comparisons to be made only between countries with similar coverage.

Glossary and Abbreviations

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

Explanation of terms:

ABPA: allergic bronchopulmonary aspergillosis is an allergic lung disease characterised by an excessive response to the mould *Aspergillus fumigatus.*

BMI: body mass index: weight (kg) / [height (m)]^2.

Bronchodilator: medication that relaxes the muscles of the airways, used also for asthma.

CFRD: CF related diabetes.

CFTR: CF transmembrane conductance regulator, is a protein at the cell surface that controls the salt and water balance across a cell. The gene that causes CF is the blueprint for the CFTR protein. Everyone has two copies of the gene for CFTR, but to be born with CF; both CFTR genes must be affected by a CF-causing mutation.

CFTR modulator therapy: are therapies designed to correct the mal-functioning CFTR protein. Since different mutations cause different defects in the protein structure or functionality CFTR modulators either correct or potentiate CFTR assembly or function, and can be combined to become more efficient. As the CFTR modulator therapies are specific to certain mutation classes the currently available therapies are effective only in people with those mutations.

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

FEV$_1$%: the FEV$_1$ as a percentage of the average value for healthy people of the same age, height and sex.

Haemoptysis: coughing up blood. This happens frequently in small amounts in CF, so the complication we asked for here is major bleeding (more than 250 ml).

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

Heterozygous: CF is caused by mutations of the CFTR gene, one on each allele. One is inherited from the mother and one from the father. If these are two different mutations, the person is considered to be heterozygous.

ICM: Intestinal current measurement is a method to diagnose or exclude CF difficult situations (e.g. unclear relevance of CFTR mutations). CF is caused by abnormalities in the mechanism that carries salt into and out of cells. With ICM, the rate of salt transport is measured in tissue samples (rectal biopsy) taken from the person and measured against reference values of a healthy population. ICM can be carried out at any age.
LCI: Lung clearance index, measured by multiple breath washout (MBW); this is a test that measures inhomogeneity of lung ventilation. A tracer gas is inhaled, and the time to exhale a defined proportion of the gas is determined (LCI). MBW is sensitive and particularly useful to measure lung function in children and people with milder forms of CF.
Macrolides: a type of antibiotic with anti-inflammatory properties. Azithromycin is a macrolide often used in people with CF who have chronic Pseudomonas aeruginosa lung infection.
Max: maximum. It is the highest value.
Mean: it is the average value of a set of measurements. For example, if the mean age at diagnosis is 3 years, it means that, on average, the patients are diagnosed when they are 3 years old.
Meconium ileus: 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.
NIPPV: Non-invasive positive pressure ventilation; this refers to mechanical ventilation that helps patients with breathing difficulties. It is done with the help of a face mask and does not require the insertion of an artificial airway (tube). It can be one of 2 types: BiPaP (Bi-level positive air pressure) or CPaP (continuous positive air pressure).
NPD: Nasal Potential Difference; this is a method to diagnose or exclude CF in unclear cases and involves placing an electrode on the surface of the inside of the nose to measure the electrical potential difference across the nasal epithelium. The NPD is a result of the transport of ions such as sodium and chloride in and out of the cells, a mechanism that is affected by defects in the CFTR protein.
Pancreatic insufficiency: the absence of pancreatic enzymes in the gut leading to malnutrition if not treated (in the ECFSPR pancreatic insufficiency is therefore defined as the use of pancreatic enzyme supplementation).
25th Pctl: 25th percentile, also called first quartile. It is the value that separates the set of measurements in two parts, so that one quarter (25%) of the measurements is below it and the other three quarters are above it. For example, if the 25th percentile for age at diagnosis is 1 month, it means that a quarter of the patients were diagnosed before they were a month old, and the other three quarters were diagnosed after they were a month old.
50th Pctl: 50th percentile, also called second quartile or median (please refer to definition of Median).
75th Pctl: 75th percentile, also called third quartile. It is the value that separates the set of measurements in two parts, so that three quarters (75%) are below it and the other quarter is above it. For example, if the 75th percentile for age at diagnosis is 3 years, it means that three quarters of the patients are diagnosed before they were 3 years old, and the remaining quarter was diagnosed after they reached 3 years of age.
Pneumothorax: collapsed lung. In CF usually because of severe lung damage.
PPI: Proton Pump Inhibitors, is medication that reduces, or at least ameliorates, stomach acid levels.
Quartiles: The 25th Percentile, the median (the 50th Percentile) and the 75th percentile are collectively called quartiles, because they divide the set of measurements into quarters.
rhDNase: recombinant human DNase (marketed as Pulmozyme®).
Z-score (or standardised scores): 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 year old boy is -2, it means that the BMI for that boy is 2 standard deviations below the mean BMI of 10 year old boys of the reference population.

Summary of data report

Outcome		Females	Males	Total
Patients registered in the ECFSPR	n (%)	23688 (47.5)	26198 (52.5)	49886
Age at follow-up (in years; patients alive on 31/12/2018)	mean median	20.5 18.0	21.2 19.0	20.9 18.5
Patients ≥ 18 years (patients alive on 31/12/2018)	%	49.8	52.4	51.2
Age at diagnosis*	mean (years) median (months)	4.1 4.3	3.9 4.1	4.0 4.1
Patients with at least one F508del allele recorded*	%	80.6	80.6	80.6
Patients living with lung transplant*	n (%)	922 (4.7)	894 (4.2)	1816 (4.4)
Patients living with liver transplant*	n (%)	72 (0.4)	153 (0.7)	225 (0.6)
Patients deceased in 2018**	n (%)	217 (1.1)	229 (1.0)	446 (1.1)
Age at death (years)**	mean median	29.6 27.0	32.8 31.0	31.2 29.0

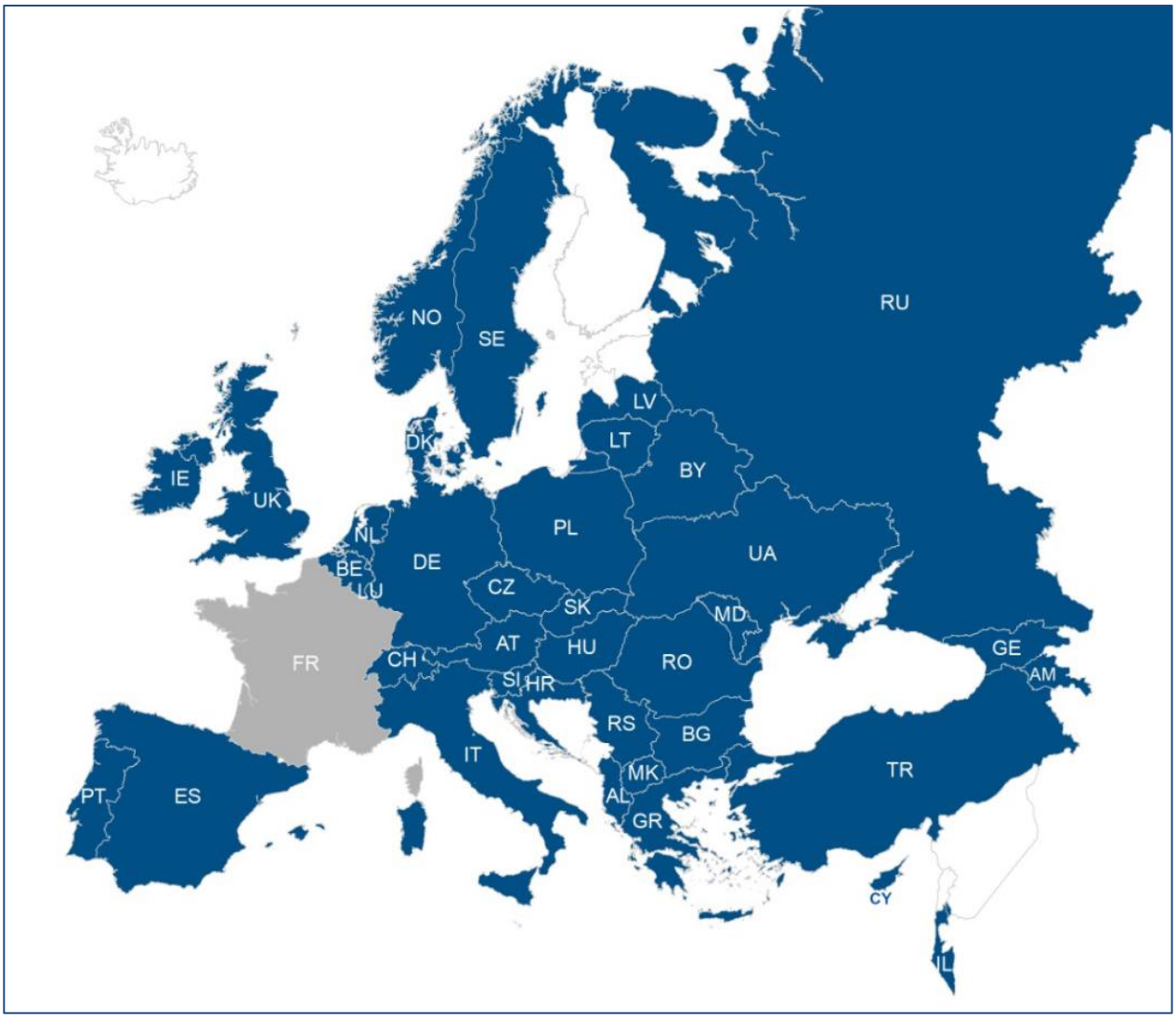
* Only patients seen during the year are presented. The total number of patients presented is 48,211.

** Only patients seen during the year are presented. For the United Kingdom, all patients with confirmed diagnosis of Cystic Fibrosis (seen and not seen) were included (N=10,509). The total number of patients presented is 41,933.

Data report

1. Demographics

Figure 1.1 Map of countries that contributed to the ECFSPR in year 2018.



Countries that contributed 2018 data are marked in blue.

France is in grey, since no 2018 data have been reported due to the COVID-19 pandemic in 2020. In this chapter, 2017 data are used for France.

Table 1.1 Number of patients in year 2018, by country.

Country	Patients registered, not lost to follow-up	Patients seen	Estimated coverage 2018
Albania	128	112	80%
Armenia	33	28	>70%
Austria	816	793	>90%
Belarus[1]	111	111	>60%
Belgium*	1314	1298	>90%
Bulgaria	193	192	>70%
Croatia**[1]	132	123	>95%
Cyprus	26	15	>80%
Czech Republic*	636	615	99%
Denmark*	528	512	99%
France*[2]	6940	6940	>90%
Georgia[3]	72	71	>80%
Germany*	6361	6361	80%
Greece**	611	588	>95%
Hungary*	498	496	90%
Ireland*[4]	1288	1224	90%
Israel**	589	537	>95%
Italy*	5531	5501	95%
Latvia	42	37	>90%
Lithuania	28	24	52%
Luxembourg	39	38	>95%
Rep of Moldova	64	56	>90%
The Netherlands*	1473	1394	>90%
North Macedonia	127	119	>90%
Norway*	294	290	>80%
Poland	917	882	>60%
Portugal**	307	296	>95%
Romania	224	215	35%
Russian Federation*	3425	3137	>95%
Serbia	198	180	>90%
Slovak Republic**	308	278	>90%
Slovenia	113	109	>95%
Spain	2320	2192	75%
Sweden*	689	689	>95%
Switzerland**	983	950	>95%
Turkey	1811	1807	>50%
Ukraine	208	154	>20%
United Kingdom*	10509	9847	99%
Total	49886	48211	

* Countries with an established national CF registry.

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

[1] Croatia reports on behalf of the Croatian CF Patient Database.

[2] For France 2017 demographic data are reported.

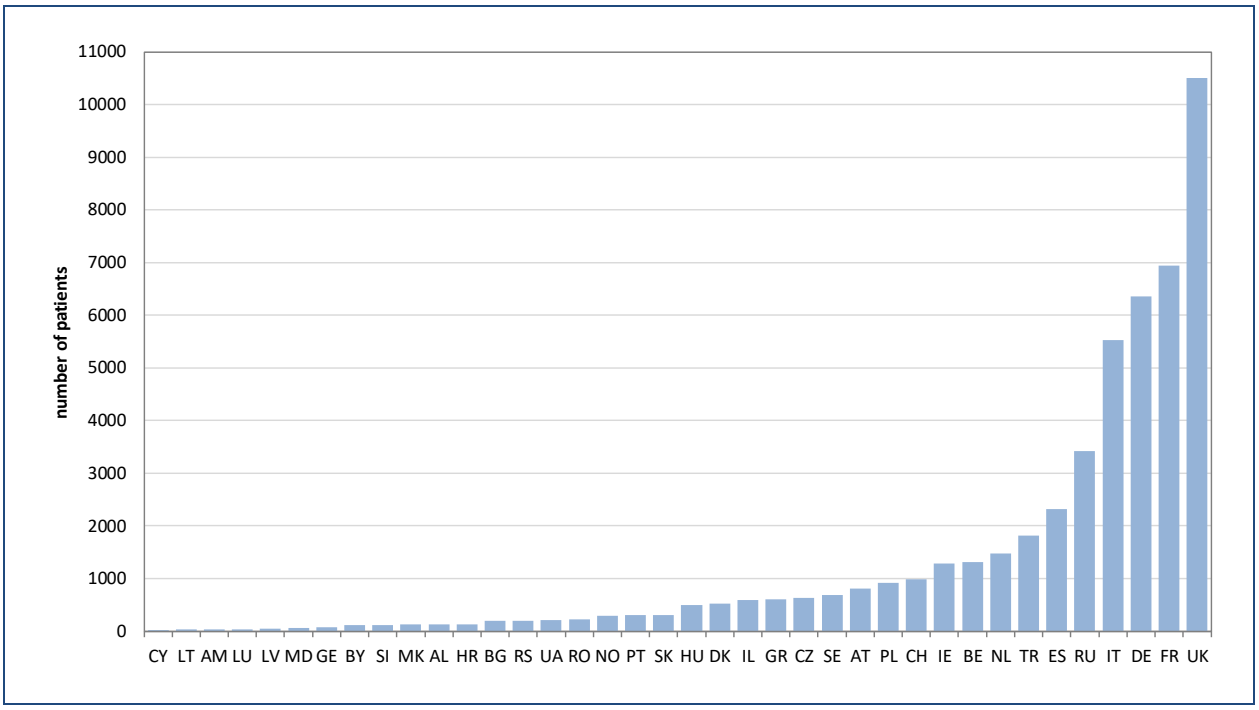
[3] For Georgia no adult patients are reported.

[4] The number of patients in this report differs from that in the Irish 2018 annual report, because for 2 patients the diagnosis was later reversed.

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

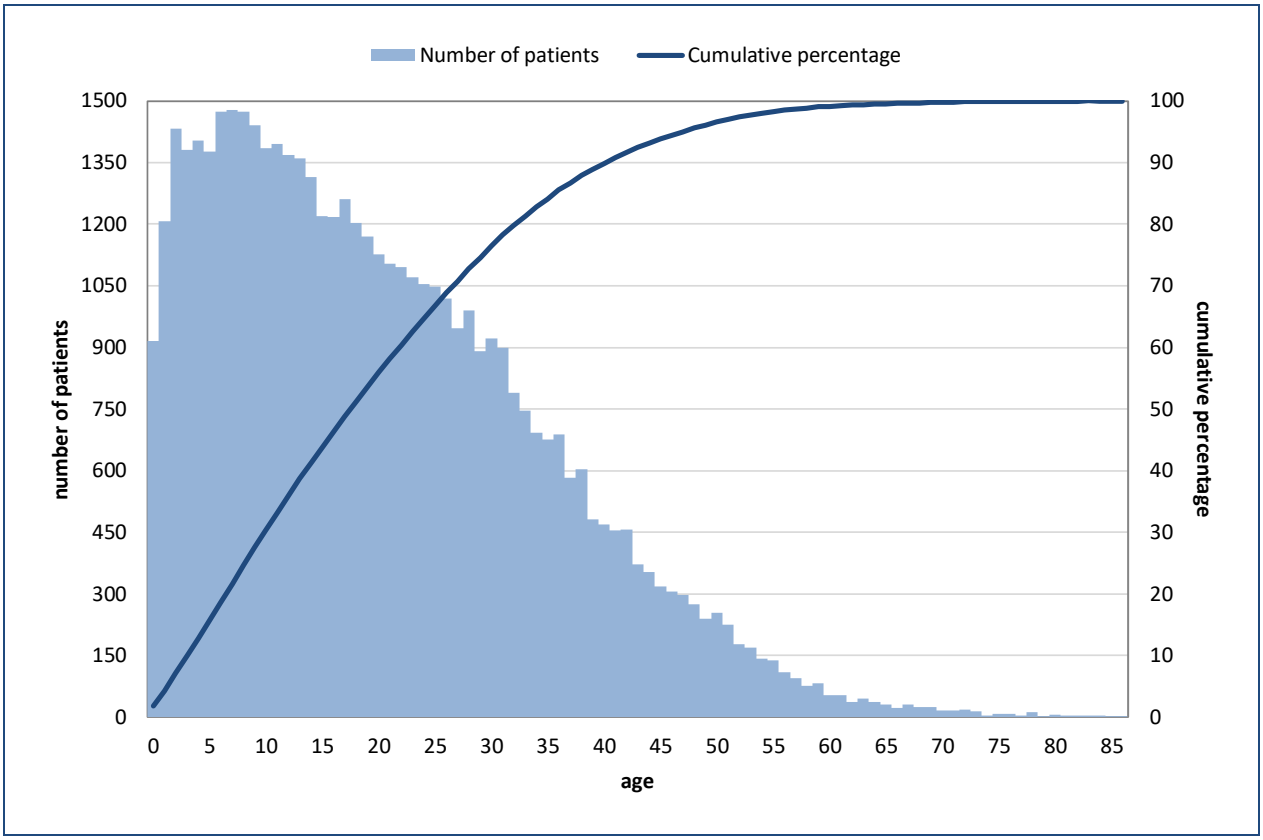
The column “patients seen” presents only the patients who have attended the clinic during the year. The column “Estimated coverage 2018” shows the estimated percentage of people with CF living in that country who are included in the national registry/national data collection as reported by the country. For some countries, one individual centre may include almost all patients, e.g. Latvia and Luxembourg.

Figure 1.2 Number of patients registered in the ECFSPR in year 2018, by country.



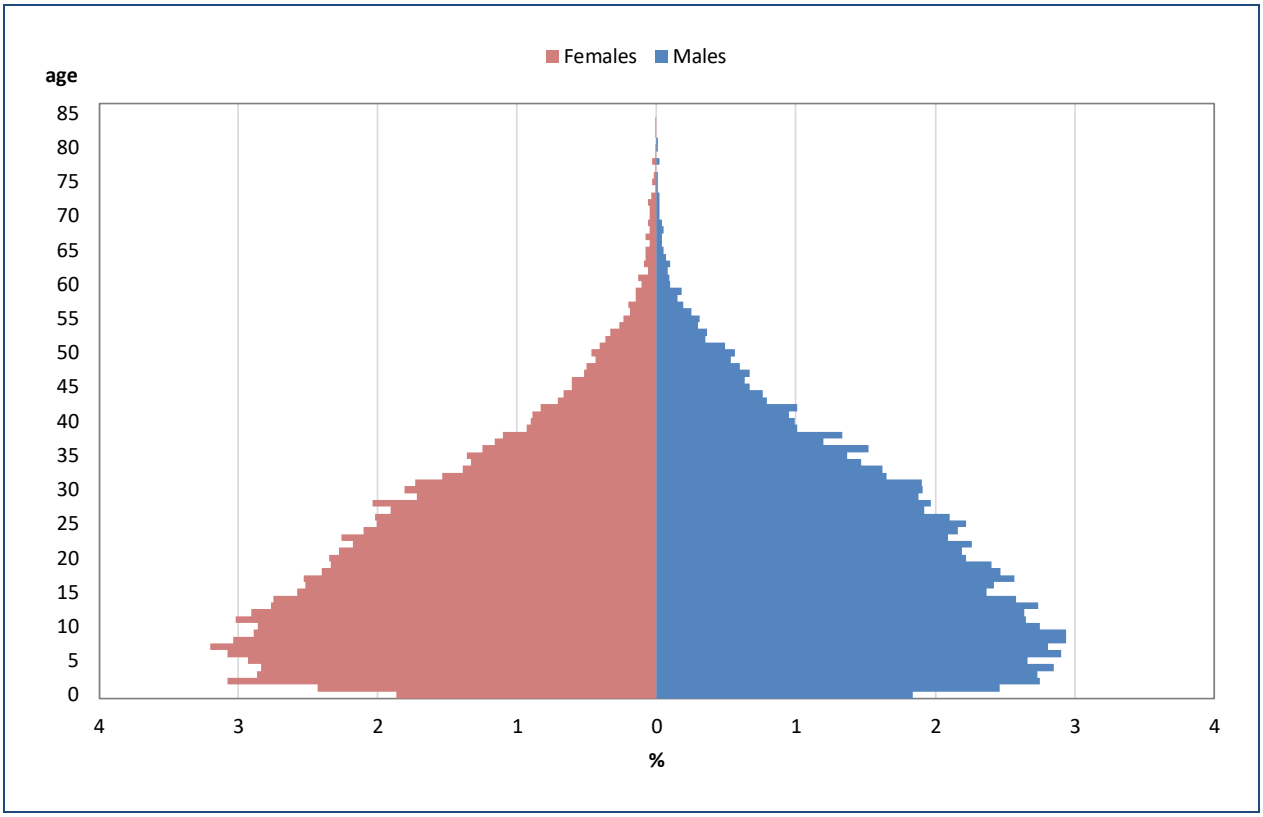
Each vertical bar shows the number of patients living in that country in 2018. Please refer to table 1.1 for the coverage in each country.

Figure 1.3 Age at follow-up distribution. Patients alive on 31/12/2018.



Each blue vertical bar represents the number of patients of that age alive in 2018. 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 31/12/2018.



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 4.0 years, see table 2.1).

Table 1.2 Proportion of children (<18 years) and adults (≥18 years), by country. Patients alive on 31/12/2018.

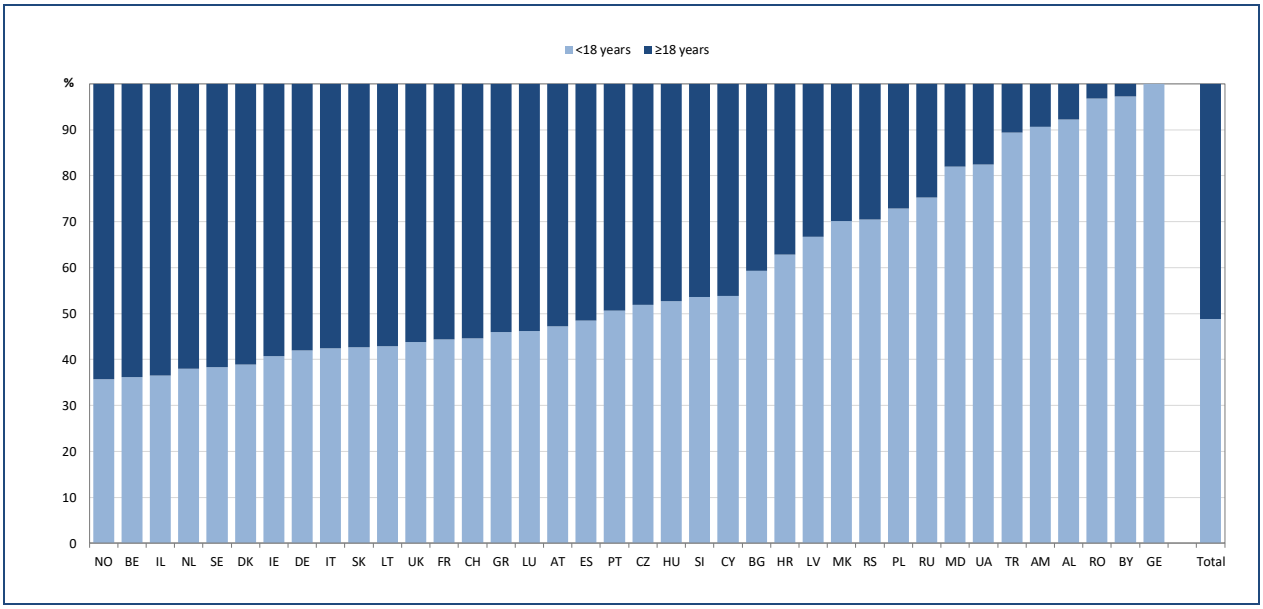
Country	Children (<18 years) number (%)	Adults (≥18 years) number (%)
Albania	118 (92.19)	10 (7.81)
Armenia	29 (90.63)	3 (9.38)
Austria	383 (47.23)	428 (52.77)
Belarus	108 (97.30)	3 (2.70)
Belgium	470 (36.21)	828 (63.79)
Bulgaria	111 (59.36)	76 (40.64)
Croatia	83 (62.88)	49 (37.12)
Cyprus	14 (53.85)	12 (46.15)
Czech Republic	324 (51.84)	301 (48.16)
Denmark	202 (38.92)	317 (61.08)
France	3051 (44.32)	3833 (55.68)
Georgia	72 (100)	0 -
Germany	2640 (41.94)	3654 (58.06)
Greece	280 (45.98)	329 (54.02)
Hungary	258 (52.65)	232 (47.35)
Ireland	519 (40.74)	755 (59.26)
Israel	214 (36.52)	372 (63.48)
Italy	2334 (42.49)	3159 (57.51)
Latvia	28 (66.67)	14 (33.33)
Lithuania	12 (42.86)	16 (57.14)
Luxembourg	18 (46.15)	21 (53.85)
Rep of Moldova	50 (81.97)	11 (18.03)
The Netherlands	556 (37.98)	908 (62.02)
North Macedonia	89 (70.08)	38 (29.92)

Note: Belarus and Georgia have 0% coverage for adult patients.

[table 1.2 continued]

Country	Children (<18 years) number (%)	Adults (≥18 years) number (%)
Norway	104 (35.74)	187 (64.26)
Poland	668 (72.85)	249 (27.15)
Portugal	152 (50.67)	148 (49.33)
Romania	216 (96.86)	7 (3.14)
Russian Federation	2541 (75.29)	834 (24.71)
Serbia	136 (70.47)	57 (29.53)
Slovak Republic	129 (42.72)	173 (57.28)
Slovenia	60 (53.57)	52 (46.43)
Spain	1117 (48.44)	1189 (51.56)
Sweden	263 (38.34)	423 (61.66)
Switzerland	434 (44.60)	539 (55.40)
Turkey	1607 (89.43)	190 (10.57)
Ukraine	169 (82.44)	36 (17.56)
United Kingdom	4544 (43.81)	5828 (56.19)
Total	24103 (48.81)	25281 (51.19)

Figure 1.5 Proportion of children (<18 years) and adults (≥18 years), by country. Patients alive on 31/12/2018.



Note: Belarus and Georgia have 0% coverage for adult patients.

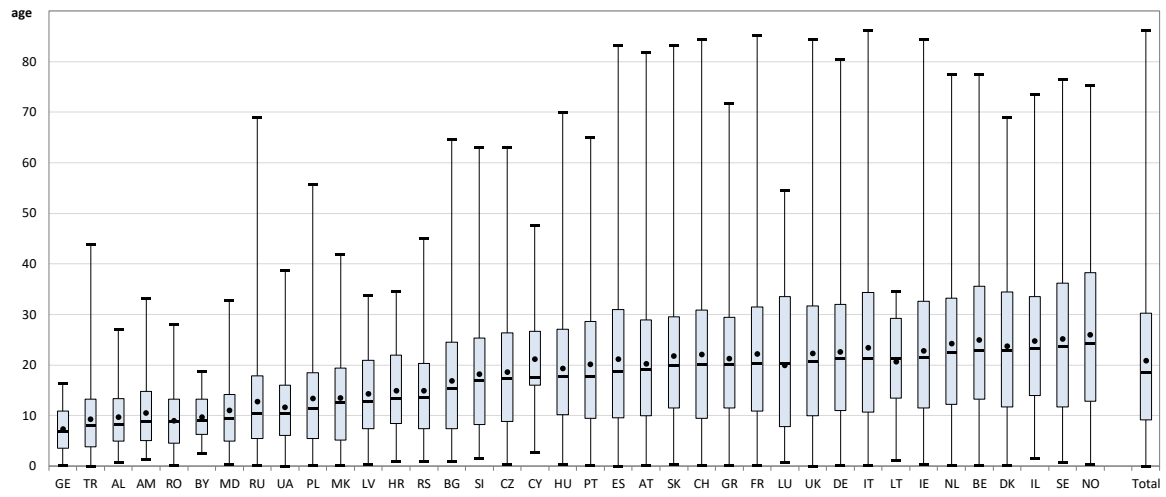
This graph shows the percentage of patients in each country who are adults (dark blue) or children (light blue). The percentage of adult patients varies considerably between the different countries, but this is partly an effect of the way the patients are included: for some countries only a few individual centres send data to the ECFSPR, and the proportion of children and adults may reflect only the proportion of paediatric and adult centres in that country who participate in the ECFSPR and not the real proportion in the country. Please refer to table 1.1, page 20, for national coverage.

Table 1.3 Age at follow-up: descriptive statistics, by country and overall. Patients alive on 31/12/2018.

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	128	9.7	0.6	5.0	8.3	13.4	27.0
Armenia	32	10.6	1.2	5.1	8.8	14.8	33.2
Austria	811	20.3	0.2	10.0	19.2	28.9	81.7
Belarus	111	9.7	2.5	6.3	9.0	13.2	18.7
Belgium	1298	25.0	0.2	13.2	23.0	35.6	77.5
Bulgaria	187	16.9	0.9	7.4	15.4	24.5	64.5
Croatia	132	15.0	0.8	8.4	13.3	22.0	34.5
Cyprus	26	21.2	2.8	16.0	17.6	26.7	47.5
Czech Republic	625	18.6	0.3	8.8	17.4	26.4	63.0
Denmark	519	23.8	0.1	11.7	23.0	34.5	69.0
France	6884	22.2	0.1	10.9	20.3	31.5	85.1
Georgia	72	7.4	0.1	3.5	6.9	10.9	16.4
Germany	6294	22.6	0.1	11.0	21.2	32.0	80.5
Greece	609	21.3	0.2	11.5	20.2	29.4	71.8
Hungary	490	19.4	0.4	10.2	17.7	27.1	69.9
Ireland	1274	22.8	0.3	11.5	21.5	32.6	84.4
Israel	586	24.8	1.5	14.0	23.4	33.5	73.5
Italy	5493	23.5	0.1	10.7	21.2	34.3	86.1
Latvia	42	14.4	0.4	7.4	12.8	20.9	33.7
Lithuania	28	20.7	1.0	13.5	21.3	29.2	34.5
Luxembourg	39	20.0	0.6	7.8	20.3	33.5	54.5
Rep of Moldova	61	11.1	0.3	5.0	9.3	14.2	32.7
The Netherlands	1464	24.3	0.1	12.2	22.5	33.2	77.5
North Macedonia	127	13.5	0.2	5.2	12.5	19.4	41.8
Norway	291	26.0	0.4	12.8	24.2	38.3	75.3
Poland	917	13.4	0.1	5.5	11.5	18.5	55.7
Portugal	300	20.2	0.2	9.5	17.7	28.6	65.0
Romania	223	9.0	0.2	4.5	8.9	13.2	28.0
Russian Federation	3375	12.8	0.1	5.5	10.4	17.9	69.0
Serbia	193	15.0	0.9	7.4	13.5	20.3	45.1
Slovak Republic	302	21.8	0.3	11.5	20.0	29.5	83.2
Slovenia	112	18.2	1.4	8.2	17.0	25.3	63.1
Spain	2306	21.2	0.0	9.6	18.8	31.0	83.2
Sweden	686	25.2	0.7	11.7	23.6	36.2	76.5
Switzerland	973	22.1	0.1	9.5	20.1	30.9	84.3
Turkey	1797	9.3	0.0	3.8	8.0	13.2	43.8
Ukraine	205	11.7	0.0	6.1	10.5	16.0	38.7
United Kingdom	10372	22.3	0.0	10.0	20.7	31.7	84.4
Total	49384	20.9	0.0	9.2	18.5	30.3	86.1

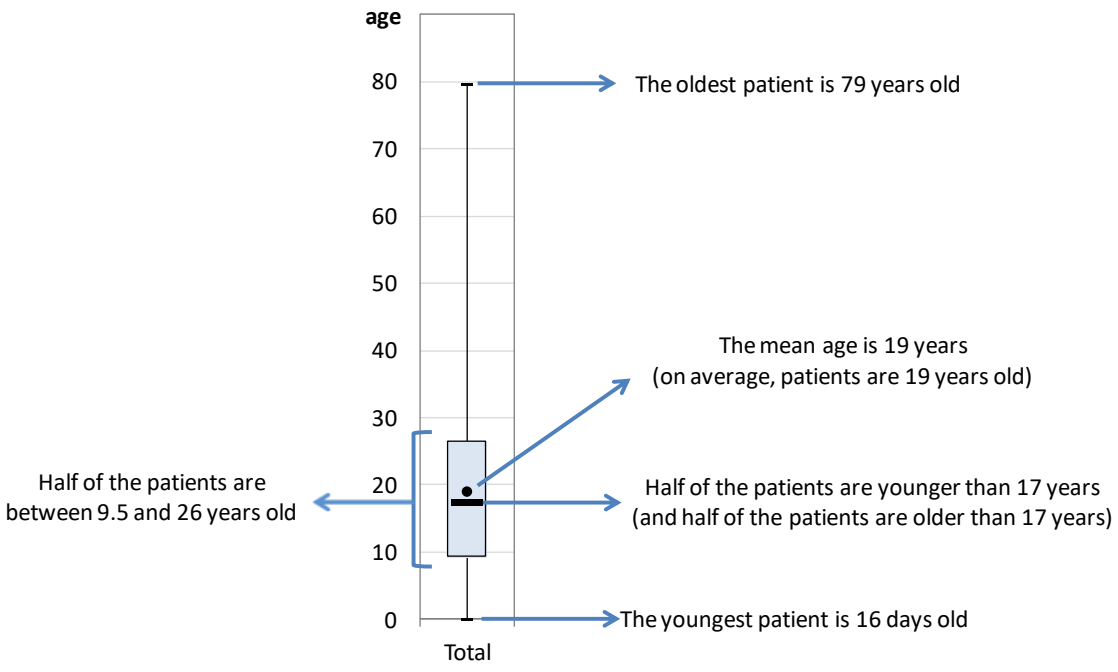
This table shows the descriptive statistics for age at follow-up of the patients by country and overall. Only patients who were alive on 31/12/2018 are included.

Figure 1.6 Age at follow-up: box-plot, by country and overall. Patients alive on 31/12/2018.



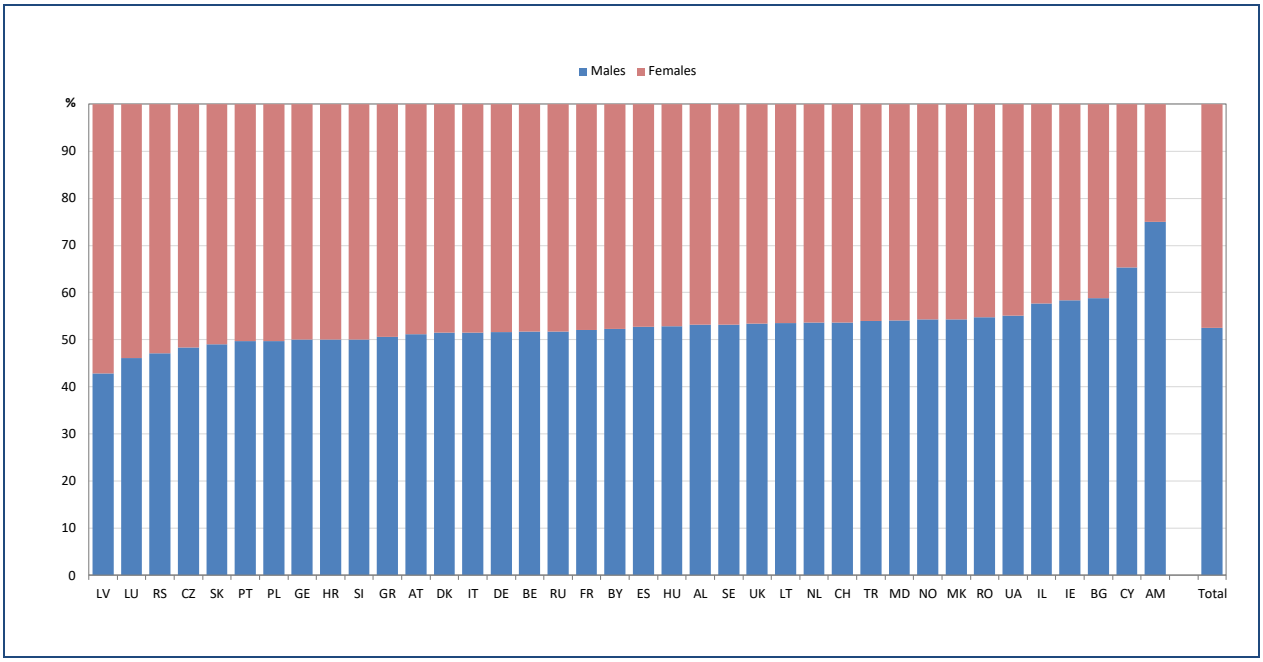
This box-plot is a graphic representation of the age detailed in table 1.3. For each country the dash (black line crossing the blue box) is the median, the black dot is the mean and the whiskers (vertical lines with a T-shaped end) are the minimum and the maximum.

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



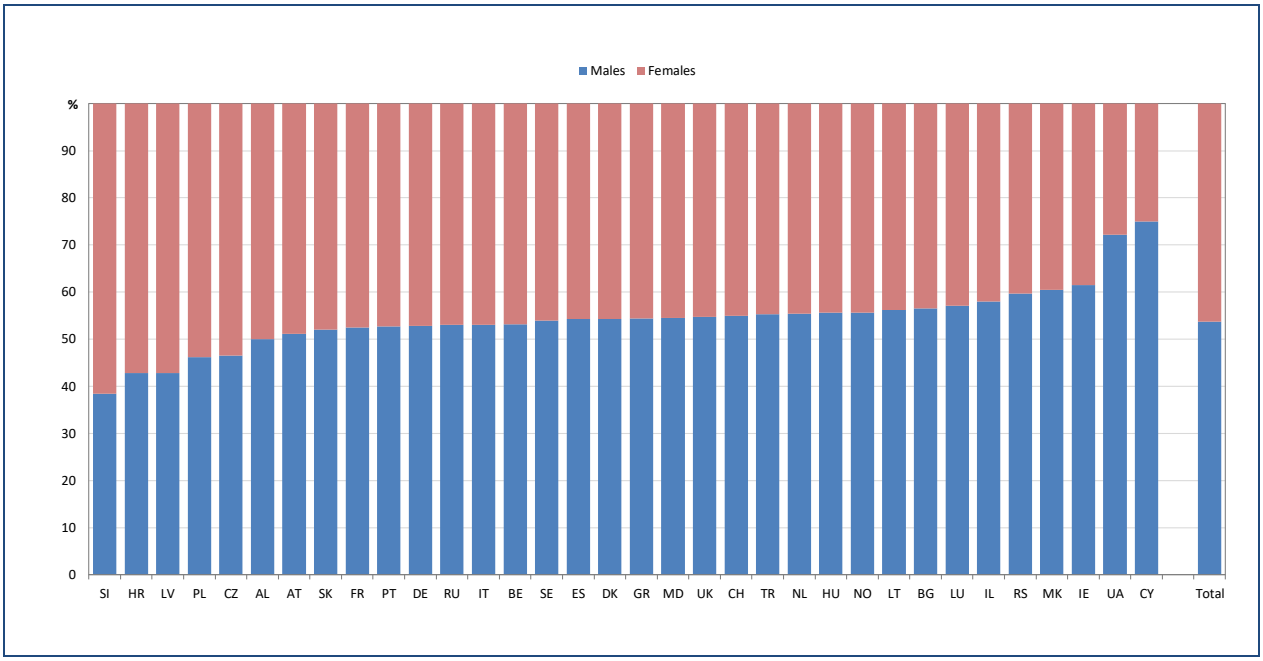
Note: This is an example of how to read a box-plot. The numbers used in this figure are not real and do not refer to figure 1.6.

Figure 1.7 Sex distribution, by country and overall. Patients alive on 31/12/2018.



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 31/12/2018 and aged 18 years or more.



Note: Belarus and Georgia have 0% coverage for adults and are excluded from the table.
Armenia and Romania have <5 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. The 2017 data from France (N=6,940) are not included.

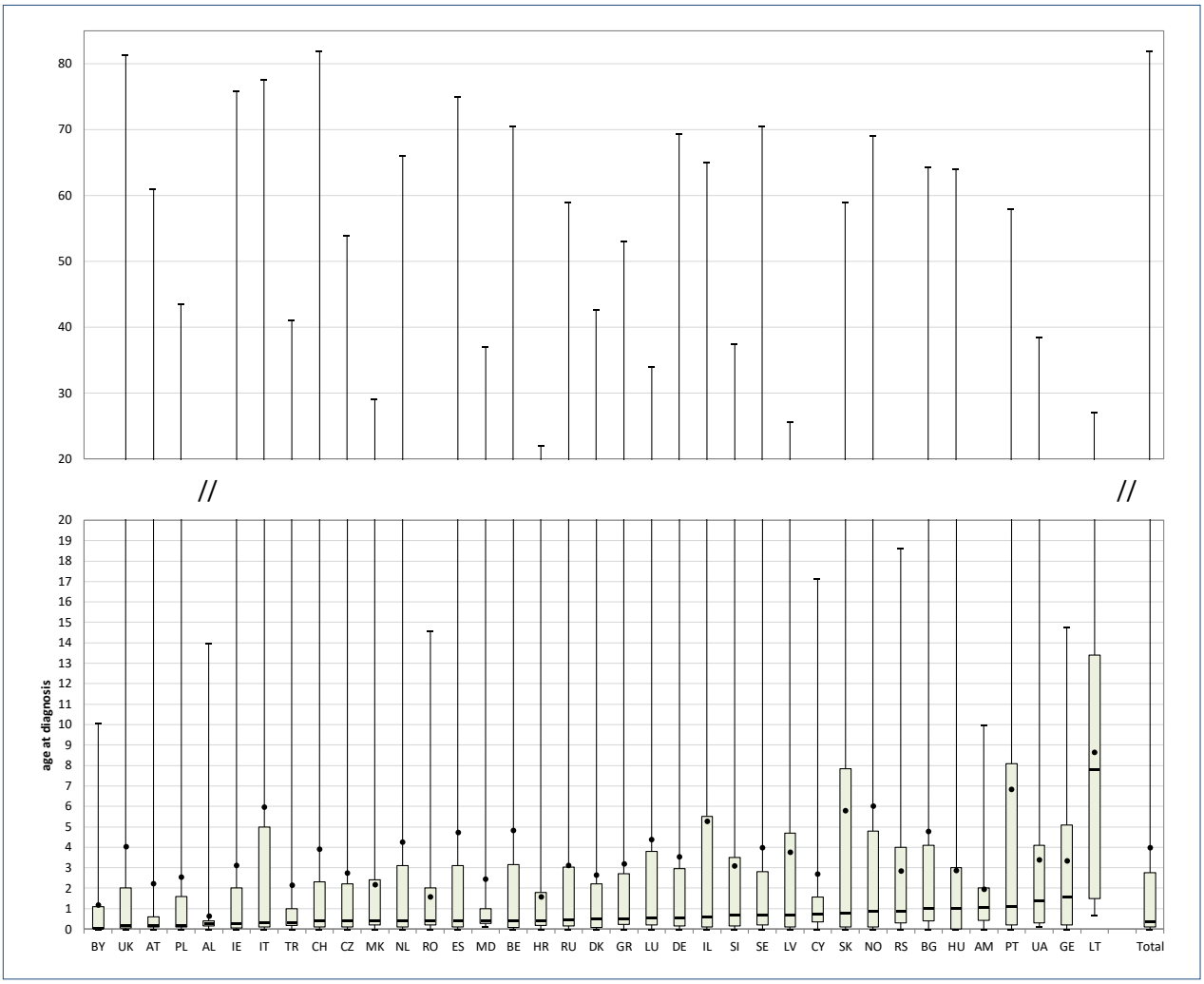
Table 2.1 Age at diagnosis (in years): descriptive statistics, by country and overall. All patients seen in 2018.

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	112	0	0.66	0	0.16	0.25	0.41	14.00
Armenia	28	0	1.97	0	0.42	1.04	2.00	10.00
Austria	731	62	2.23	0	0.10	0.20	0.60	61.00
Belarus	103	8	1.19	0.01	0.02	0.05	1.10	10.08
Belgium	1295	3	4.83	0	0.08	0.42	3.16	70.55
Bulgaria	187	5	4.79	0	0.40	1.00	4.10	64.30
Croatia	118	5	1.59	0	0.17	0.42	1.80	22.00
Cyprus	15	0	2.71	0.01	0.35	0.76	1.56	17.11
Czech Republic	609	6	2.77	0	0.10	0.40	2.20	53.90
Denmark	512	0	2.65	0	0.08	0.50	2.21	42.67
Georgia	69	2	3.36	0	0.20	1.60	5.10	14.80
Germany	6115	246	3.55	0	0.16	0.58	2.96	69.41
Greece	580	8	3.21	0	0.22	0.53	2.70	53.00
Hungary	427	69	2.89	0	0.00	1.00	3.00	64.00
Ireland	1219	5	3.13	0	0.06	0.29	2.00	75.83
Israel	532	5	5.28	0	0.11	0.59	5.50	65.00
Italy	5411	90	5.99	0	0.10	0.31	4.98	77.62
Latvia	37	0	3.78	0	0.10	0.70	4.70	25.60
Lithuania	23	1	8.65	0.7	1.50	7.80	13.4	27.10
Luxembourg	38	0	4.39	0	0.20	0.55	3.80	34.00
Rep of Moldova	56	0	2.47	0.1	0.28	0.41	1.00	37.00
The Netherlands	1351	43	4.26	0	0.10	0.40	3.10	66.00
North Macedonia	119	0	2.19	0	0.20	0.40	2.40	29.00
Norway	279	11	6.04	0	0.10	0.90	4.80	69.00
Poland	879	3	2.56	0	0.10	0.20	1.60	43.50
Portugal	286	10	6.86	0	0.20	1.10	8.10	58.00
Romania	208	7	1.59	0	0.20	0.40	2.00	14.60
Russian Federation	3109	28	3.14	0	0.15	0.45	3.03	58.91
Serbia	177	3	2.85	0	0.30	0.90	4.00	18.60
Slovak Republic	248	30	5.80	0	0.10	0.80	7.85	59.00
Slovenia	107	2	3.10	0	0.16	0.67	3.50	37.50
Spain	2166	26	4.74	0	0.10	0.41	3.10	75.00
Sweden	677	12	4.00	0	0.19	0.68	2.81	70.57
Switzerland	838	112	3.93	0	0.10	0.40	2.30	81.90
Turkey	1794	13	2.17	0	0.17	0.33	1.00	41.00
Ukraine	154	0	3.40	0.1	0.30	1.40	4.10	38.50
United Kingdom	9759	88	4.04	0	0.05	0.17	2.00	81.35
Total	40368	903	4.00	0	0.10	0.34	2.76	81.90

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

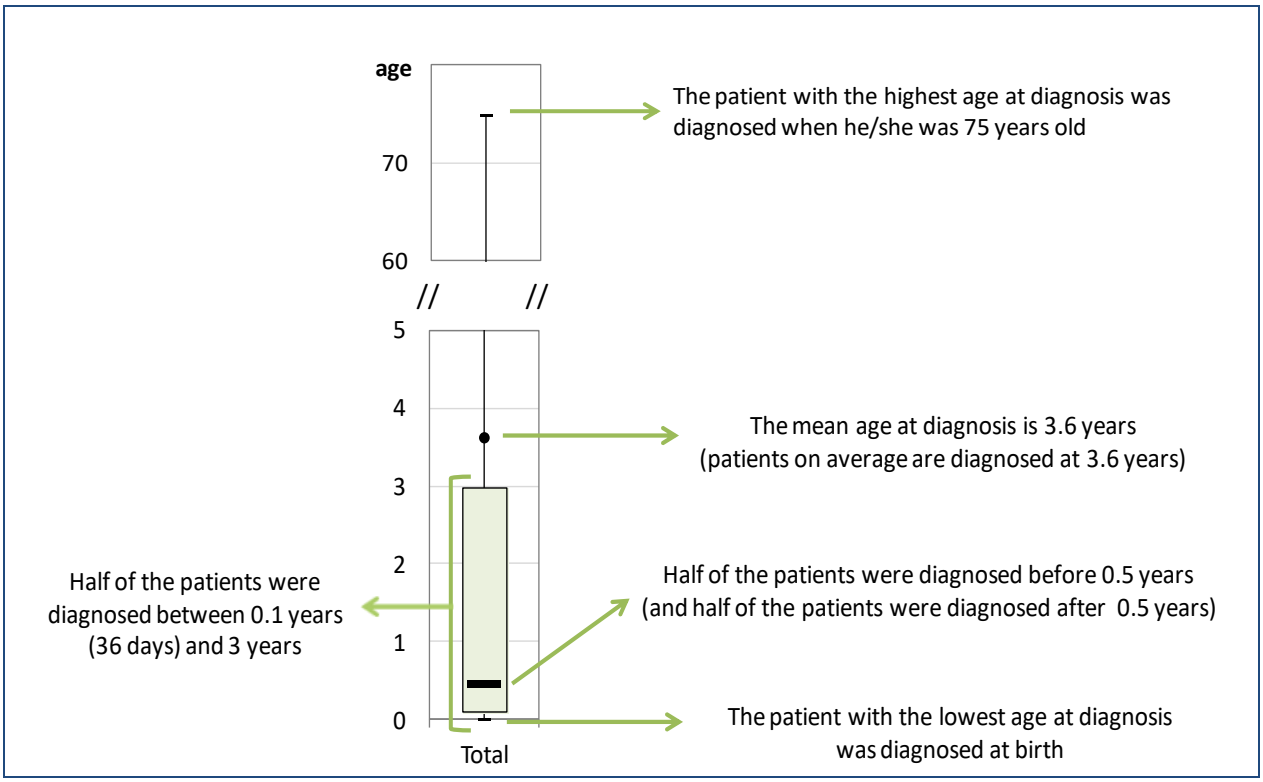
Table 2.1 shows the descriptive statistics for age at diagnosis by country and overall. For prenatal diagnoses (children diagnosed before birth), the age at diagnosis has been set to 0.

Figure 2.1 Age at diagnosis (in years): box-plot, by country and overall. All patients seen in 2018.



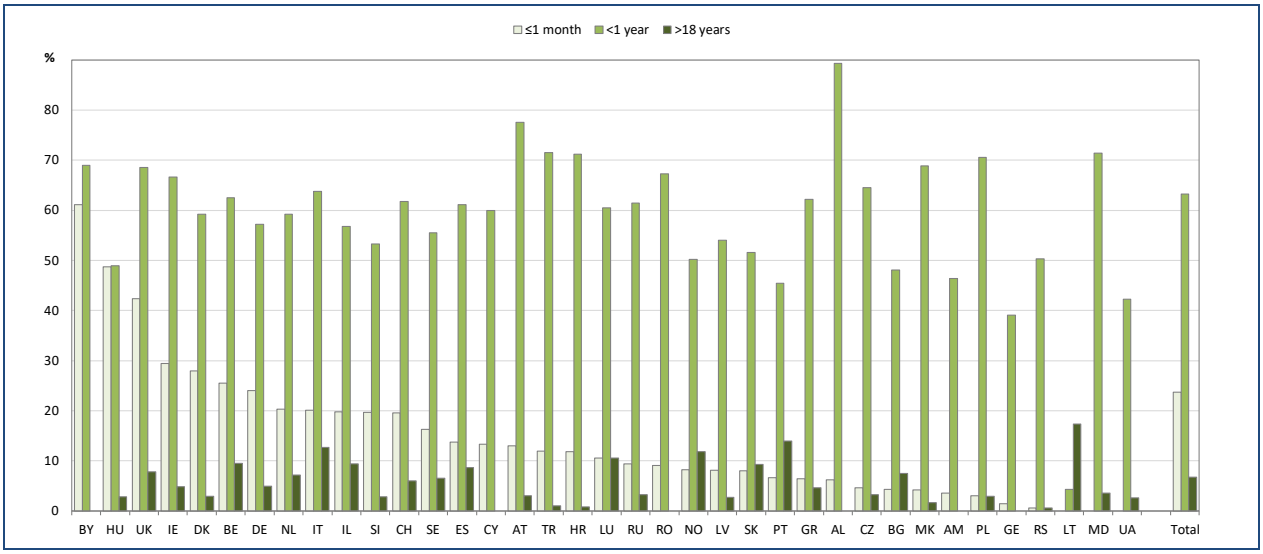
Note: For Hungary, Slovak Republic and Switzerland the information on age at diagnosis is missing for more than 10% of the patients.

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



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

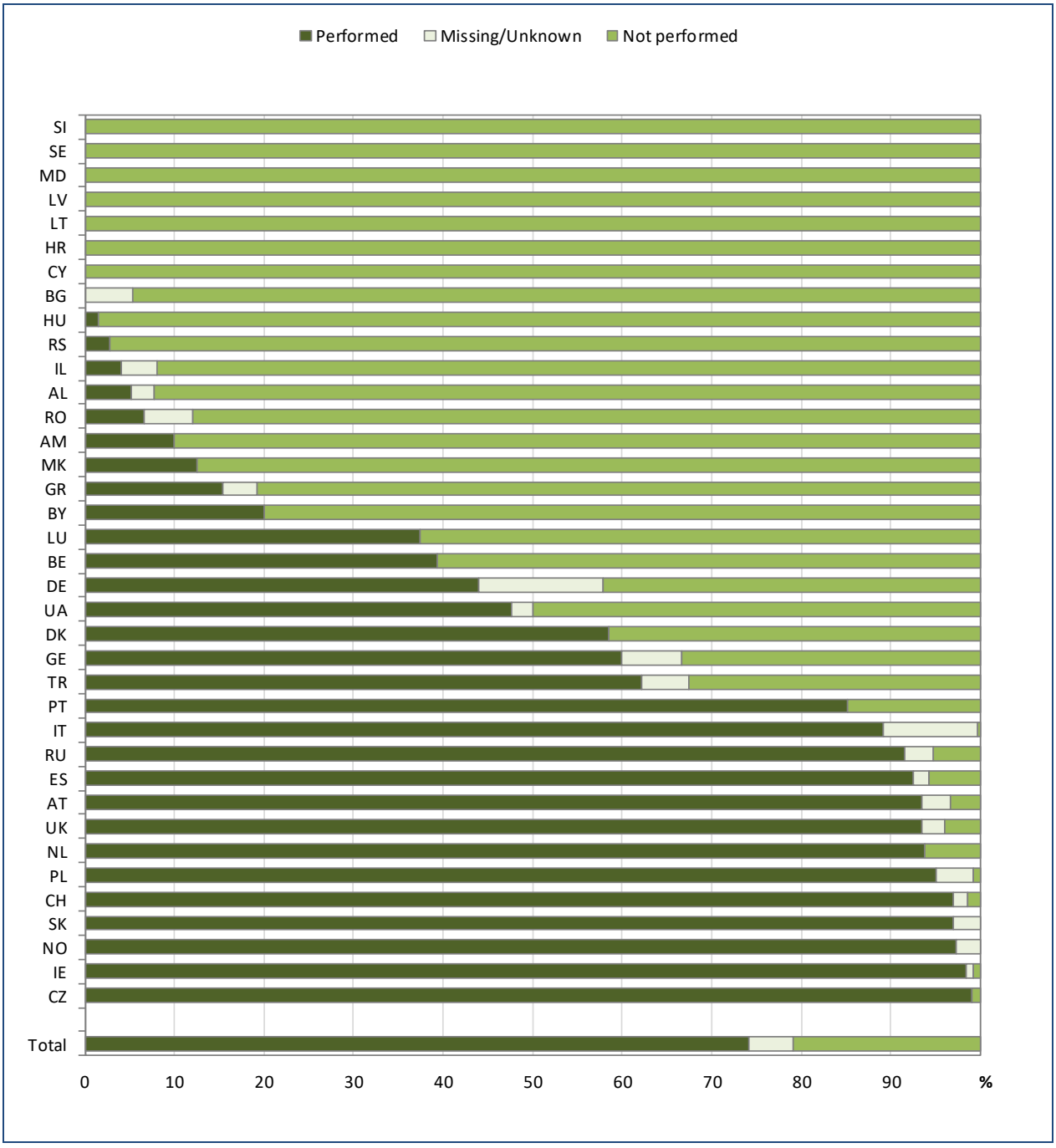
Figure 2.2 Proportion of patients diagnosed at age 1 month or younger, younger than 1 year and older than 18 years, by country and overall. All patients seen in 2018.



Note: For Hungary, Slovak Republic and Switzerland the information on age at diagnosis is missing for more than 10% of the patients.
The high proportion of patients diagnosed at the age of 1 month or younger for Belarus and Hungary might be caused by an approximation of the age at diagnosis by the clinician.
The high proportion of patients diagnosed at the age of 18 years or older for Lithuania is due to the limited number of paediatric patients reported.

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

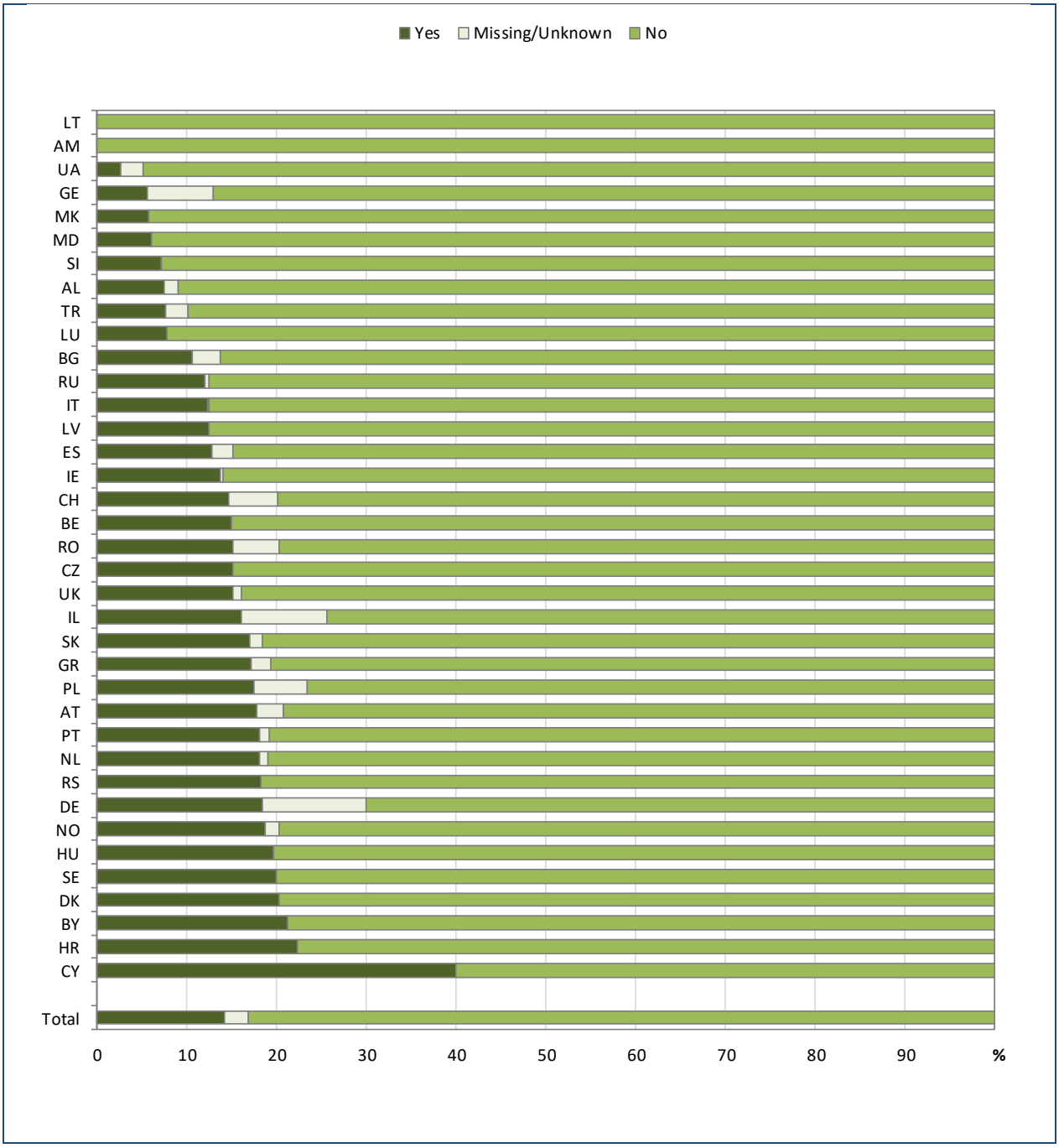
Figure 2.3 Proportion of patients who underwent neonatal screening, by country and overall. Patients 5 years old or younger seen in 2018.



Note: For Germany and Italy the information on neonatal screening is missing for more than 10% of the patients.
For Belgium and UK positive answers ("neonatal screening performed") are reported only when neonatal screening is one of the factors that led to CF diagnosis.

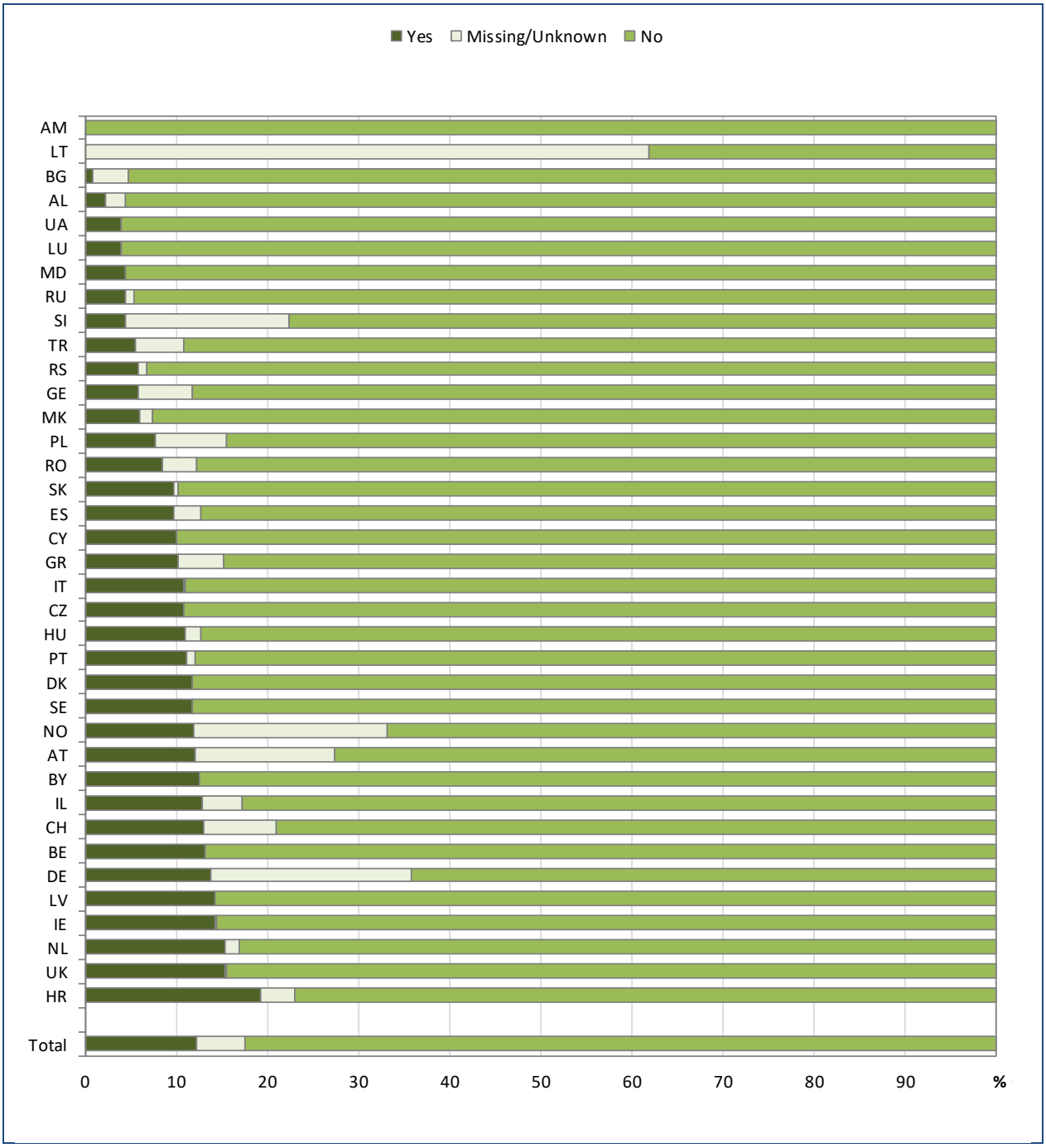
This graph shows the percentage of patients at the age of 5 years or younger in 2018 who were screened at birth, (see country-specific notes above). Dark green horizontal bars represent neonatal screening "performed", light green ones "not performed". This graph shows that, in the five years previous to 2018, in many countries the CF patients underwent neonatal, i.e. newborn screening, and that in some countries there is no neonatal screening programme. In total, 74% of all children of 5 years old or younger registered in the ECFSPR in 2018 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 Germany, the information on meconium ileus is missing for more than 10% of the patients aged 10 years or younger.

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



Note: For Austria, Germany, Lithuania, Norway and Slovenia the information on meconium ileus is missing for more than 10% of the patients aged 11 years 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 (>11 years). This difference is not because of an increase in the prevalence of meconium ileus in the younger generations but could be due to the fact that some older patients with meconium ileus have died and are therefore not present in the current data collection (which refers to patients seen in 2018). The graphs also show that the frequency of reported meconium ileus varies between countries.

3. Genetics

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

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

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

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

Table 3.1 Proportion of patients with DNA analysis and the result of this, by country and overall. All patients seen in 2018.

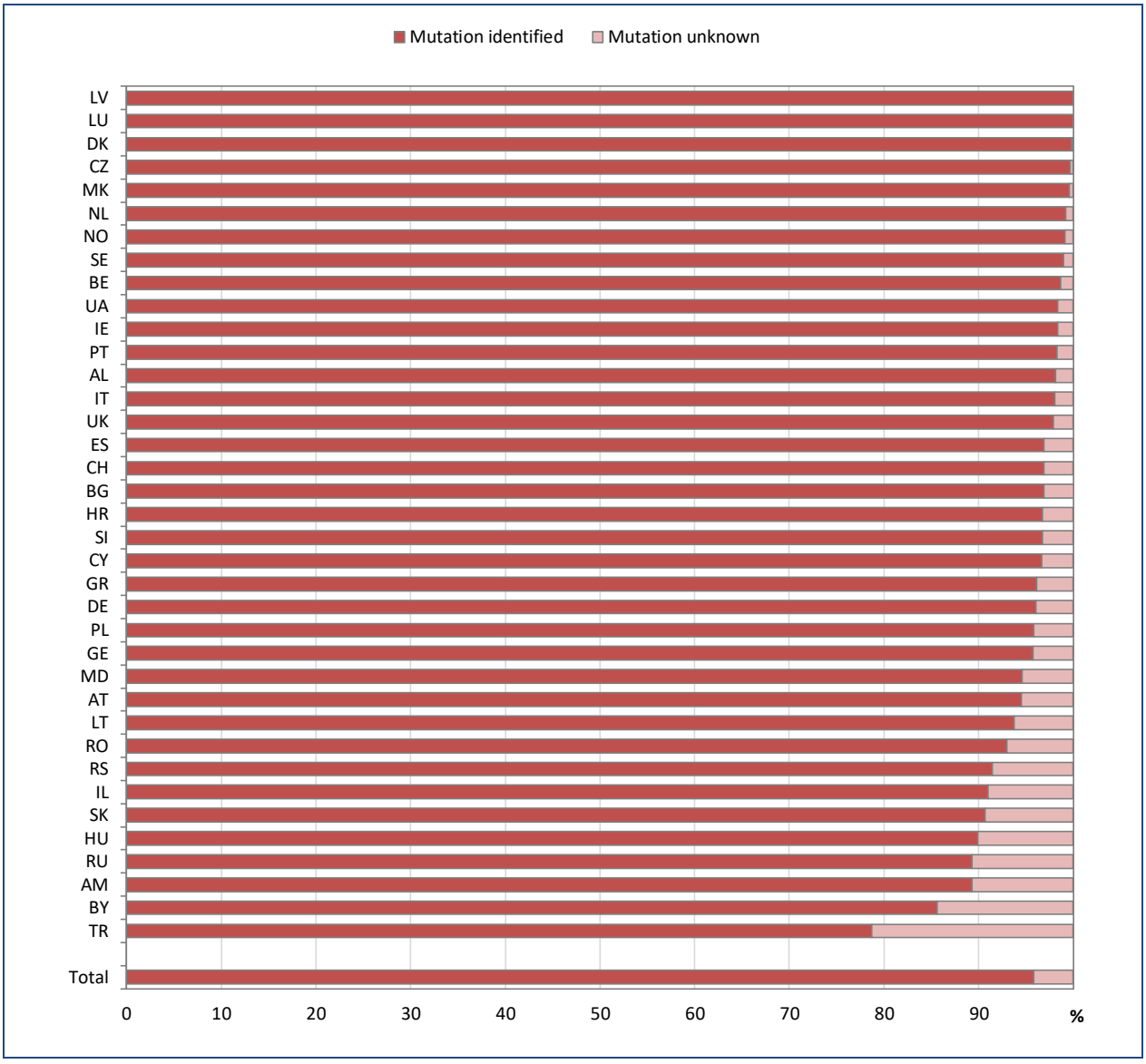
Country	N	Genotyping		Among genotyping done	
		not done	done	two mutations identified	at least one mutation unknown
		number (%)	number (%)	number (%)	number (%)
Albania	112	7 (6.25)	105 (93.75)	101 (96.19)	4 (3.81)
Armenia	28	0 -	28 (100)	22 (78.57)	6 (21.43)
Austria	793	1 (0.13)	792 (99.87)	730 (92.17)	62 (7.83)
Belarus	111	0 -	111 (100)	85 (76.58)	26 (23.42)
Belgium	1298	3 (0.23)	1295 (99.77)	1266 (97.76)	29 (2.24)
Bulgaria	192	0 -	192 (100)	180 (93.75)	12 (6.25)
Croatia	123	0 -	123 (100)	116 (94.31)	7 (5.69)
Cyprus	15	0 -	15 (100)	14 (93.33)	1 (6.67)
Czech Republic	615	1 (0.16)	614 (99.84)	611 (99.51)	3 (0.49)
Denmark	512	0 -	512 (100)	511 (99.80)	1 (0.20)
Georgia	71	0 -	71 (100)	65 (91.55)	6 (8.45)
Germany	6361	17 (0.27)	6344 (99.73)	5949 (93.77)	395 (6.23)
Greece	588	0 -	588 (100)	554 (94.22)	34 (5.78)
Hungary	496	4 (0.81)	492 (99.19)	414 (84.15)	78 (15.85)
Ireland	1224	0 -	1224 (100)	1191 (97.30)	33 (2.70)
Israel	537	0 -	537 (100)	474 (88.27)	63 (11.73)
Italy	5501	12 (0.22)	5489 (99.78)	5305 (96.65)	184 (3.35)
Latvia	37	0 -	37 (100)	37 (100)	0 -
Lithuania	24	0 -	24 (100)	22 (91.67)	2 (8.33)
Luxembourg	38	0 -	38 (100)	38 (100)	0 -
Rep of Moldova	56	0 -	56 (100)	51 (91.07)	5 (8.93)
The Netherlands	1394	3 (0.22)	1391 (99.78)	1373 (98.71)	18 (1.29)
North Macedonia	119	1 (0.84)	118 (99.16)	117 (99.15)	1 (0.85)
Norway	290	0 -	290 (100)	287 (98.97)	3 (1.03)

[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	882	2 (0.23)	880 (99.77)	827 (93.98)	53 (6.02)
Portugal	296	0 -	296 (100)	288 (97.30)	8 (2.70)
Romania	215	0 -	215 (100)	191 (88.84)	24 (11.16)
Russian Federation	3137	177 (5.64)	2960 (94.36)	2437 (82.33)	523 (17.67)
Serbia	180	4 (2.22)	176 (97.78)	151 (85.80)	25 (14.20)
Slovak Republic	278	0 -	278 (100)	233 (83.81)	45 (16.19)
Slovenia	109	2 (1.83)	107 (98.17)	101 (94.39)	6 (5.61)
Spain	2192	1 (0.05)	2191 (99.95)	2067 (94.34)	124 (5.66)
Sweden	689	0 -	689 (100)	679 (98.55)	10 (1.45)
Switzerland	950	5 (0.53)	945 (99.47)	904 (95.66)	41 (4.34)
Turkey	1807	141 (7.80)	1666 (92.20)	1210 (72.63)	456 (27.37)
Ukraine	154	0 -	154 (100)	149 (96.75)	5 (3.25)
United Kingdom	9847	35 (0.36)	9812 (99.64)	9462 (96.43)	350 (3.57)
Total	41271	416 (1.01)	40855 (98.99)	38212 (93.53)	2643 (6.47)

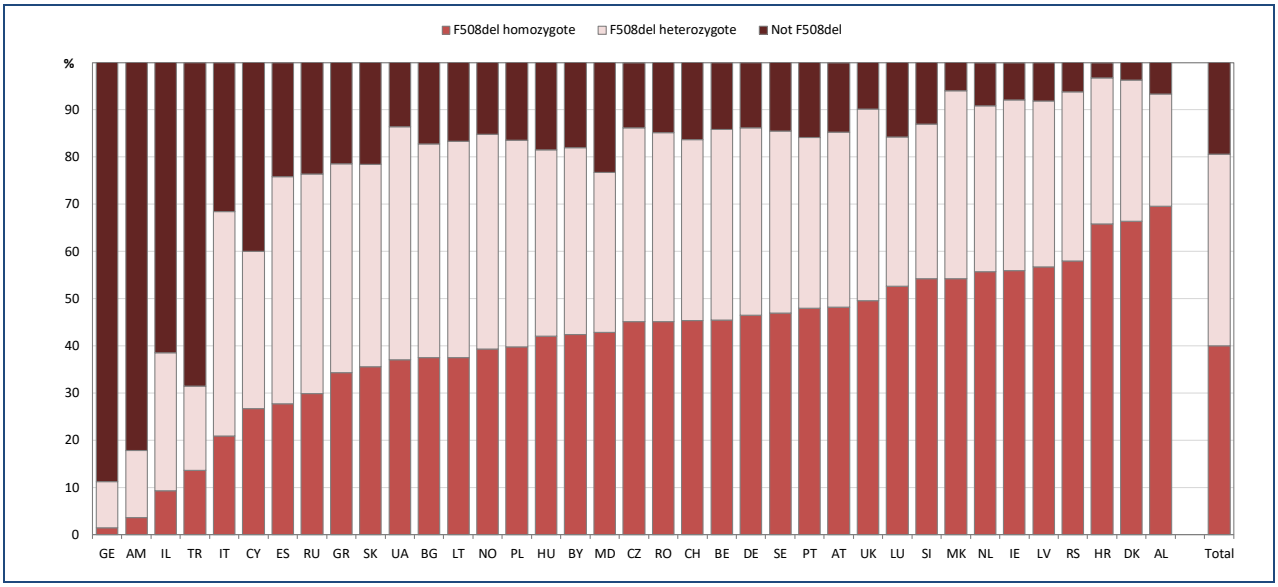
The table shows how many patients underwent DNA analysis to identify the CFTR mutations (column “genotyping done”) and, for those patients, how many patients had both mutations identified (column “two mutations identified”) and for how many one or both mutations remained unidentified (column “at least 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 6.47% 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 2018.



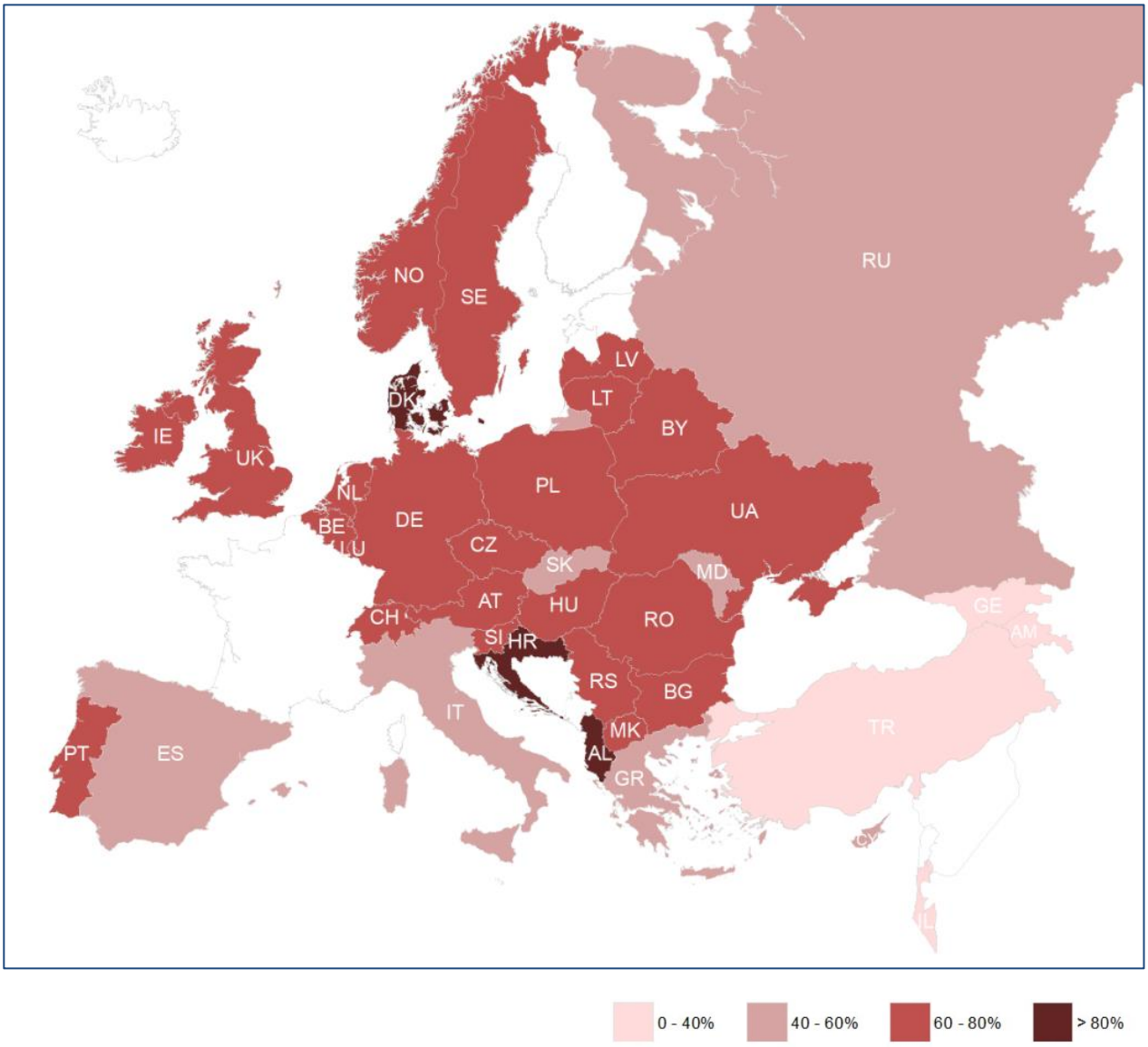
F508del is the name of the most commonly occurring CFTR mutation in the world. Patients who carry two F508del mutations are often described as having “classic CF”, but other combinations of mutations may cause the same degree of disease. We have grouped the patients in F508del homozygous (patients who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

Table 3.2 Allelic frequencies of the 18 most common mutations in the ECFSPR database.

Mutation name	Number of alleles	Percentage among tested	Country with highest allele frequency
F508del	49246	60.27	Albania (81.4%)
G542X	2219	2.72	Armenia (8.9%)
N1303K	1734	2.12	Italy (5.6%)
G551D	1120	1.37	Ireland (8.5%)
W1282X	937	1.15	Israel (23.0%)
CFTRdele2,3	899	1.10	Belarus (6.8%)
3849+10kbC->T	846	1.04	Lithuania (14.6%)
R117H	846	1.04	Ireland (3.0%)
2789+5G->A	823	1.01	Italy (2.9%)
1717-1G->A	680	0.83	Switzerland (3.1%)
R553X	666	0.82	Switzerland (2.2%)
621+1G->T	597	0.73	Greece (6.4%)
2183AA->G	565	0.69	Armenia (8.9%)
D1152H	504	0.62	Israel (5.5%)
G85E	451	0.55	Israel (2.6%)
R347P	449	0.55	Luxembourg (2.6%)
1677delTA	423	0.52	Georgia (44.4%)
R1162X	410	0.50	Slovenia (5.1%)

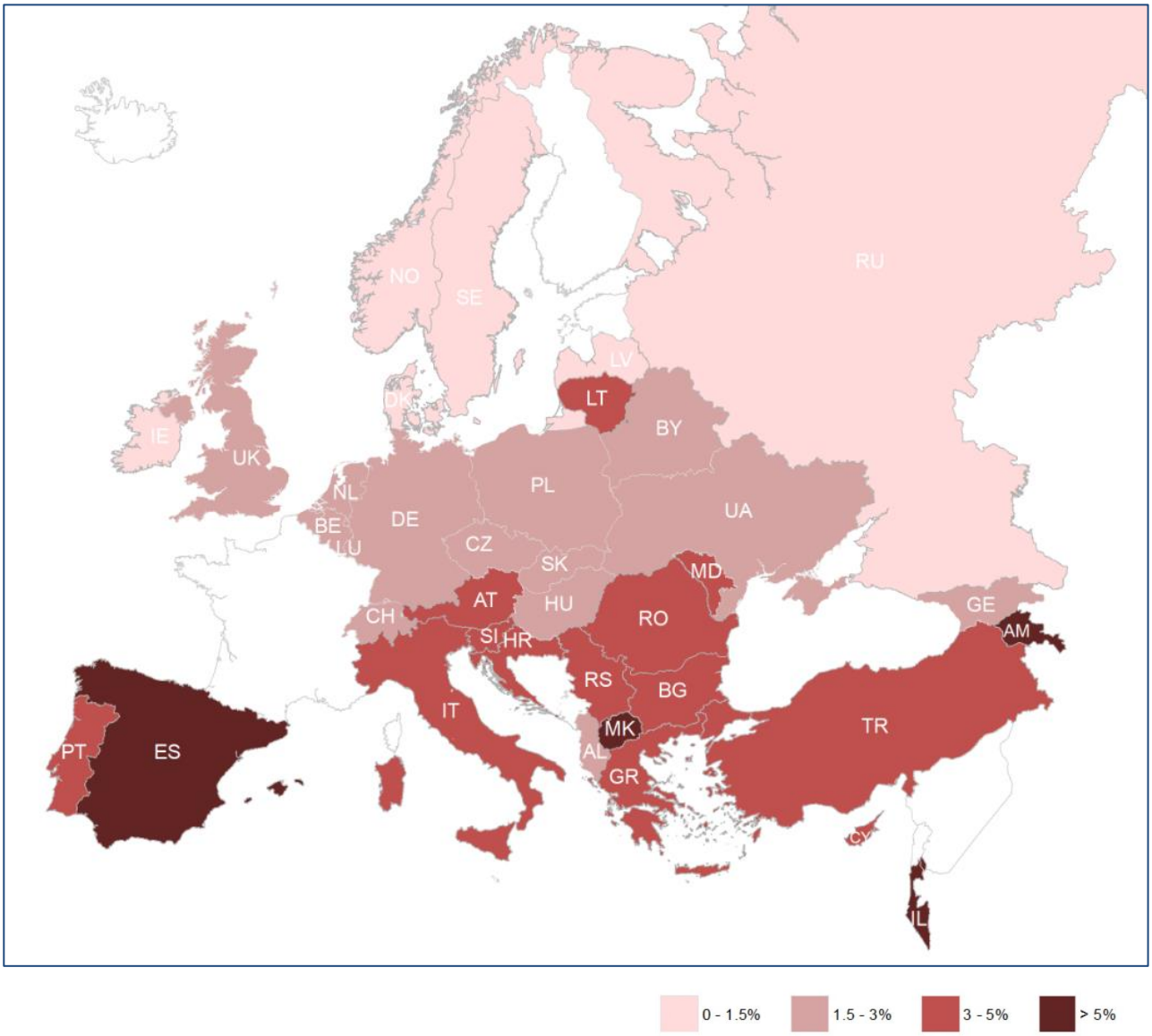
This table presents the allele frequency of the 18 most commonly occurring mutations found in the ECFSPR database. The last column reports in which country this particular mutation is most frequent. F508del is, by far, the most frequent mutation.

Figure 3.3 Geographical distribution of mutation F508del.



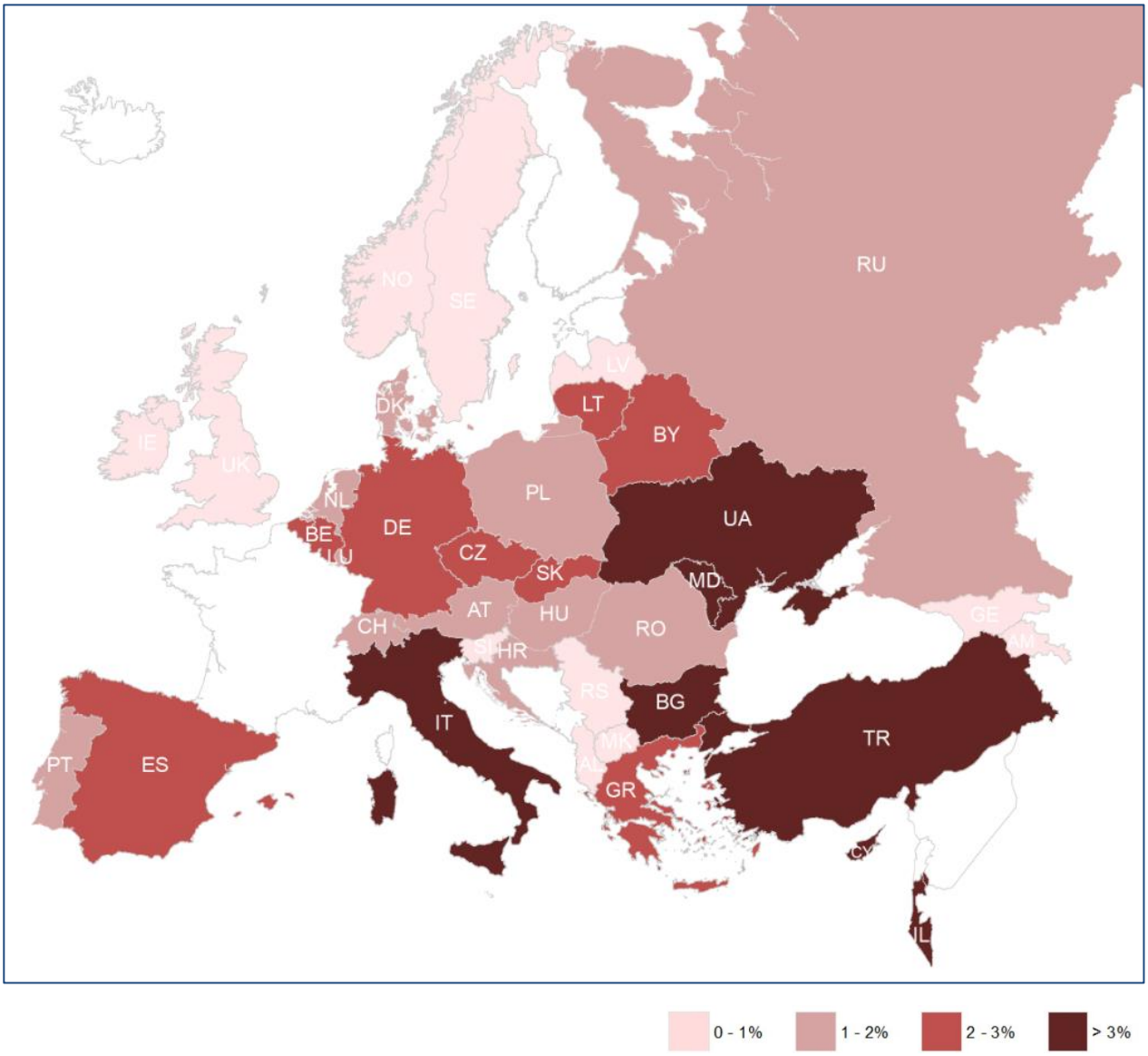
Although this mutation is the most common in all countries, it is most frequent in the South East of Europe. The highest frequency occurs in Albania (81.4%) and Croatia (81.3%), and in the north of Europe, in Denmark (81.4%).

Figure 3.4 Geographical distribution of mutation G542X.



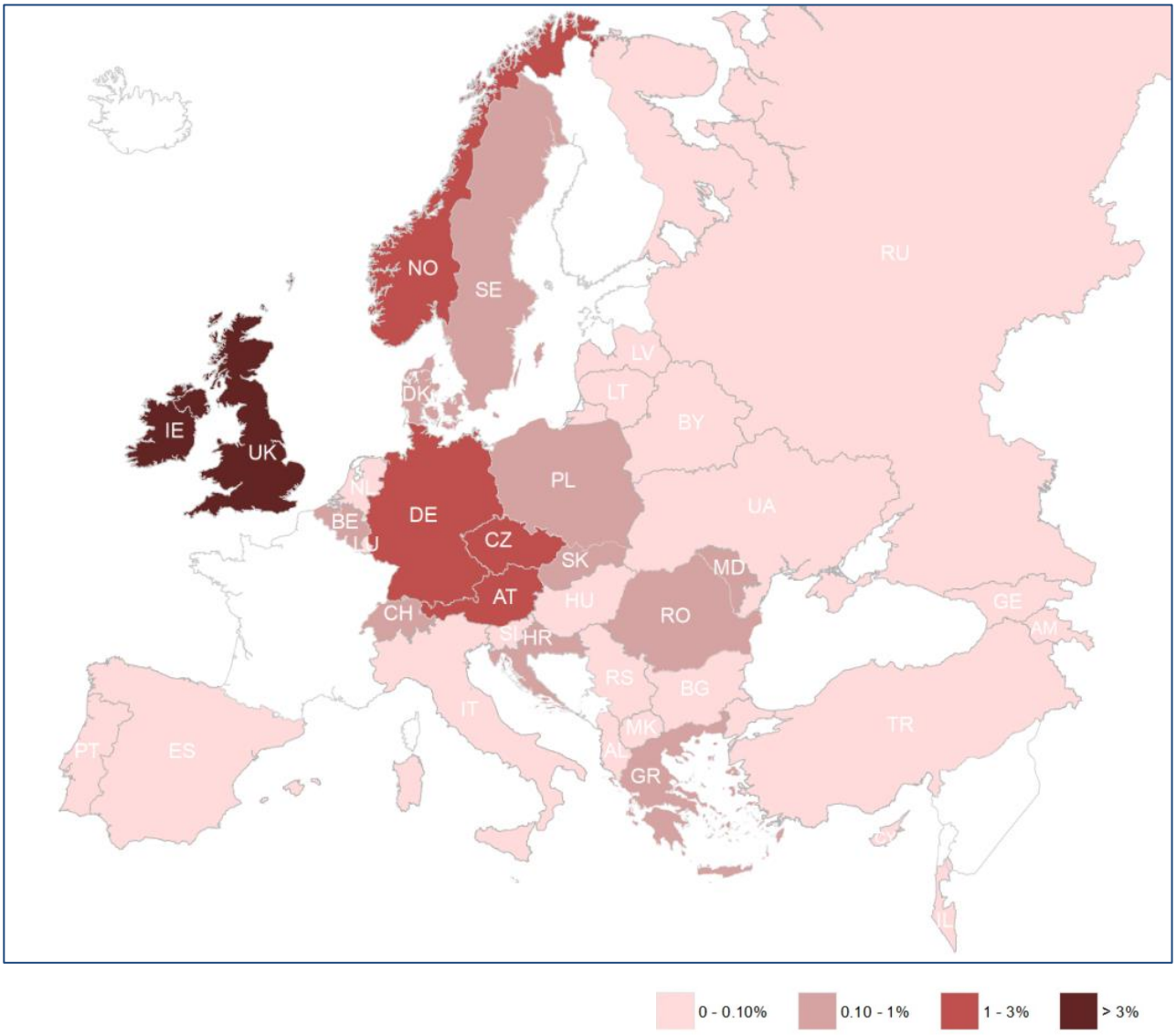
This mutation is most frequent in Southern Europe, with the highest allele frequency in Armenia (8.9%), whereas it is very rarely found in Ireland, the Scandinavian and Baltic countries and the Russian Federation.

Figure 3.5 Geographical distribution of mutation N1303K.



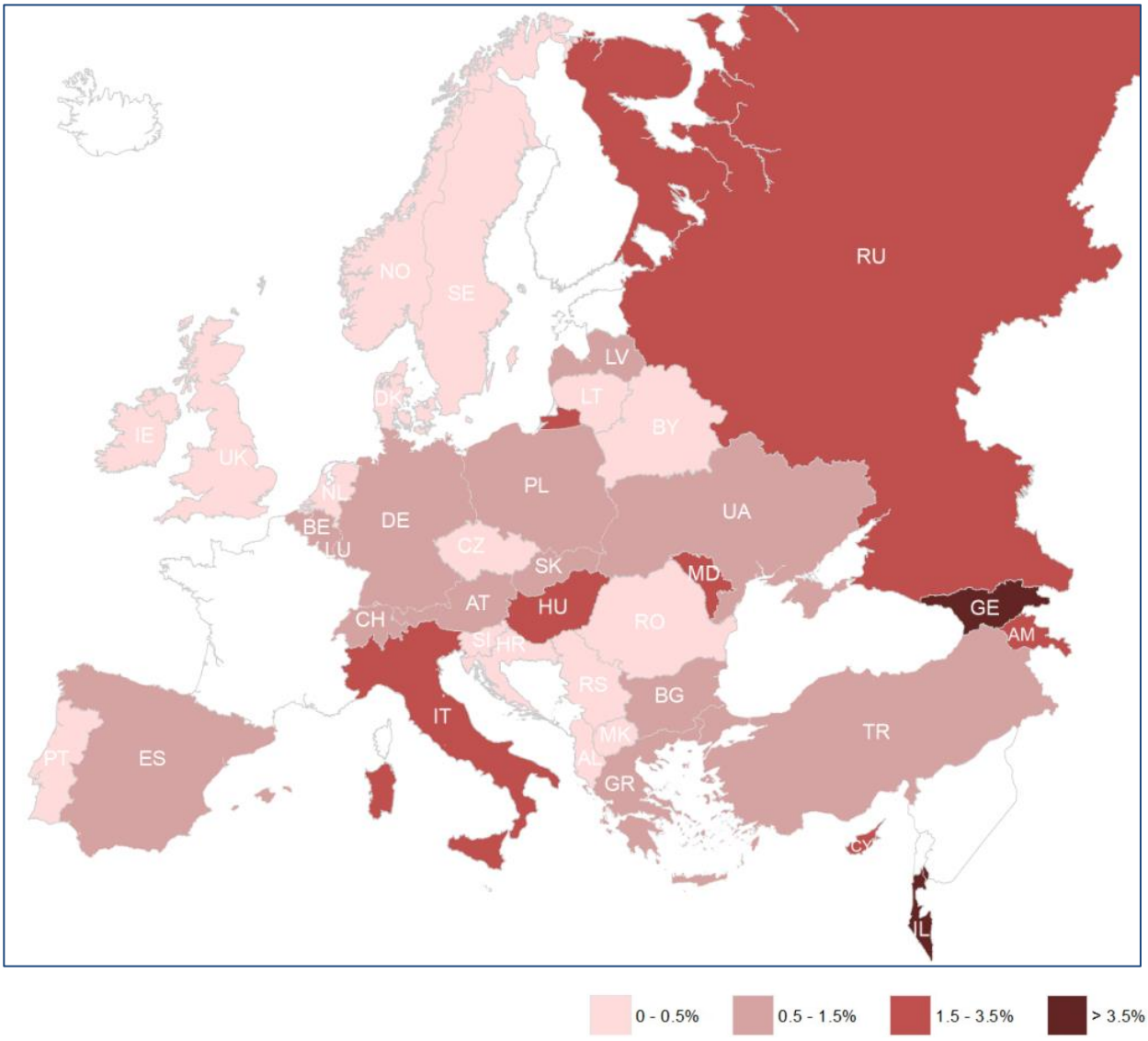
This mutation is most frequent in Italy (5.6%) and other countries in Southern and Eastern Europe, but rare in Northern Europe.

Figure 3.6 Geographical distribution of mutation G551D.



This mutation is most frequent in Ireland (8.5%) and United Kingdom (3.0%), whereas it is rare in Southern and Eastern Europe.

Figure 3.7 Geographical distribution of mutation W1282X.



This mutation, of Middle-Eastern origin, is by far most frequent in Israel (23.0%) with a very high allele frequency in Ashkenazi Jews; it is also frequent in Georgia (9.2%).

4. Lung function

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

We used the Global Lung Function Initiative equations described by Quanjer PH et al. for this report (for the full reference we refer you to Appendix 1, page 167). This is the global reference for spirometry and it has been agreed, as part of the CF global harmonisation project, that this is the best way to present lung function.

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

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

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

We excluded patients from the analyses on FEV_1 who have had one or more lung transplants, since their lung function does not reflect the severity of their CF lung disease. Moreover, we also excluded patients who had one or more transplant of other organs since the follow-up data of those patients is sometimes missing.

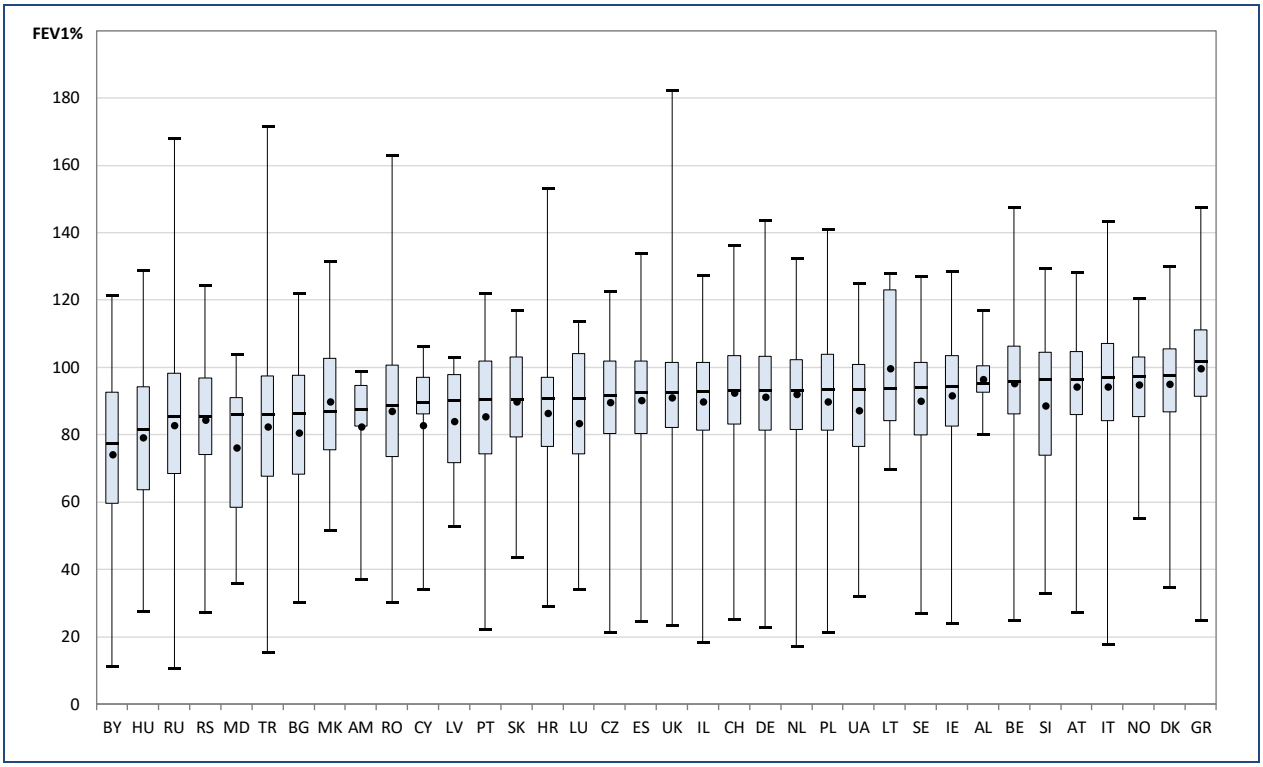
Table 4.1 FEV_1% of predicted: descriptive statistics, by country. Patients aged 6-17 years who have never had a transplant.

Country	N	N Miss	Mean (average FEV_1%)	Min	25th pctl (25% of patients have FEV_1% below this value)	Median (50% of patients have FEV_1% below this value)	75th pctl (75% of patients have FEV_1% below this value)	Max
Albania	57	8	96.3	80.0	92.7	95.2	100.4	116.9
Armenia	17	0	82.3	37.0	82.5	87.6	94.6	98.7
Austria	236	2	94.1	26.9	86.0	96.4	104.7	128.2
Belarus	53	28	74.0	11.2	59.6	77.2	92.6	121.4
Belgium	345	2	95.1	24.7	86.2	95.8	106.3	147.3
Bulgaria	66	6	80.5	30.1	68.2	86.4	97.6	122.0
Croatia	51	4	86.3	28.8	76.6	90.7	97.0	152.8
Cyprus	<10	<10	82.6	34.1	86.2	89.3	97.1	106.1
Czech Republic	212	1	89.6	21.2	80.4	91.6	101.9	122.3
Denmark	132	3	95.0	34.7	86.8	97.3	105.5	129.7
Germany	1733	32	91.1	22.7	81.4	93.1	103.3	143.4
Greece	214	6	99.6	24.7	91.5	101.6	111.2	147.4
Hungary	144	38	79.1	27.4	63.6	81.5	94.2	128.8
Ireland	381	10	91.6	23.9	82.5	94.1	103.5	128.3
Israel	172	1	89.8	18.4	81.4	92.8	101.4	127.2
Italy	1503	84	94.2	17.5	84.2	96.8	107.1	143.2
Latvia	21	0	83.9	52.7	71.7	90.1	97.8	102.9
Lithuania	<10	<10	99.6	69.7	84.2	93.7	123.0	127.7
Luxembourg	<10	<10	83.3	34.1	74.3	90.8	104.2	113.5
Rep of Moldova	23	3	76.1	35.9	58.4	85.9	91.1	103.6
The Netherlands	399	6	92.0	16.9	81.6	93.2	102.3	132.4
North Macedonia	48	3	89.7	51.4	75.5	86.8	102.6	131.4
Norway	67	2	94.8	55.0	85.3	97.2	103.1	120.4
Poland	381	24	89.8	21.2	81.3	93.3	103.9	140.8
Portugal	99	7	85.4	21.9	74.4	90.3	101.9	121.9
Romania	84	30	87.0	30.0	73.6	88.8	100.7	162.7
Russian Federation	1040	388	82.6	10.6	68.5	85.3	98.3	167.7
Serbia	85	3	84.3	27.0	74.1	85.4	96.8	124.2
Slovak Republic	87	1	89.8	43.4	79.3	90.4	103.1	116.7
Slovenia	42	0	88.5	32.8	74.0	96.2	104.6	129.2
Spain	692	15	90.1	24.6	80.3	92.3	101.9	133.8
Sweden	191	7	89.9	26.8	79.9	93.9	101.5	127.0
Switzerland	285	4	92.4	25.1	83.2	92.9	103.5	136.2
Turkey	652	227	82.2	15.2	67.6	85.9	97.5	171.4
Ukraine	73	2	87.1	32.0	76.5	93.4	100.9	124.9
United Kingdom	2867	9	90.9	23.1	82.2	92.3	101.5	182.2
Total	12477	994	90	10.6	80.3	92.6	102.6	182.2

Note: Georgia has <5 patients aged 6-17 years at FEV_1 measurement and is excluded from this table.
Note: 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 from the previous calendar year.

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

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



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

This box-plot is a graphic representation of the FEV_1 in children, expressed as % of predicted, detailed in table 4.1. For each country, the dash (black line crossing the blue box) is the median, the black dot is the mean and the whiskers (vertical lines with a T-shaped end) are the minimum and the maximum

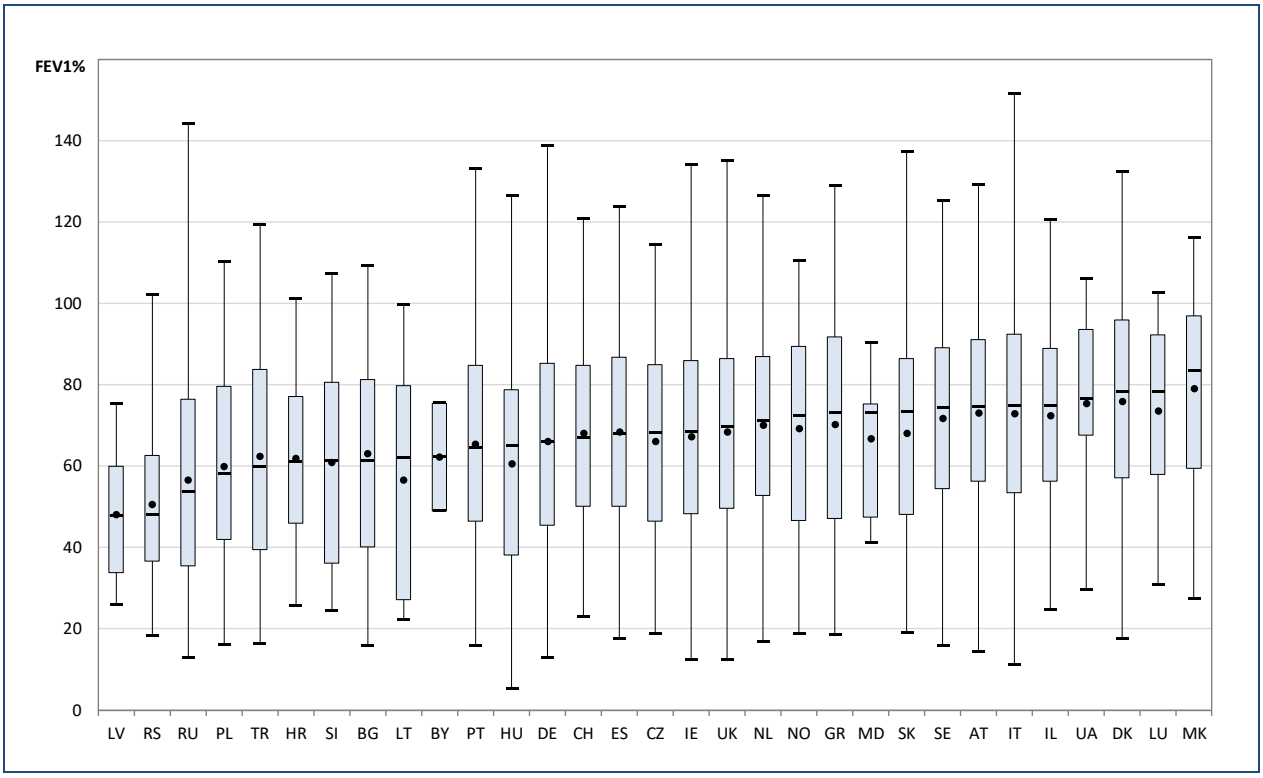
Table 4.2 FEV_1% of predicted: descriptive statistics, by country. Patients aged 18 years or older who have never had a transplant.

Country	N	N Miss	Mean (average FEV_1%)	Min	25th pctl (25% of patients have FEV_1% below this value)	Median (50% of patients have FEV_1% below this value)	75th pctl (75% of patients have FEV_1% below this value)	Max
Austria	316	3	73	14.5	56.3	74.6	91.1	129.1
Belgium	609	9	73.8	18.3	56.0	75.7	92.0	134.9
Bulgaria	74	0	63	15.9	40.1	61.4	81.3	109.3
Croatia	36	0	61.9	25.6	45.9	61.1	77.1	101.1
Czech Republic	237	5	66	18.7	46.4	68.2	84.9	114.5
Denmark	254	0	75.9	17.4	57.1	78.3	96.0	132.3
Germany	3119	63	66	13.0	45.5	65.9	85.3	138.6
Greece	261	2	70.2	18.4	47.1	73.0	91.8	128.9
Hungary	145	31	60.5	5.3	38.2	65.0	78.7	126.4
Ireland	575	8	67.2	12.2	48.3	68.3	85.9	134.1
Israel	305	0	72.4	24.6	56.3	74.9	88.9	120.5
Italy	2614	113	72.9	11.3	53.5	74.7	92.4	151.6
Latvia	<10	<10	48.1	26.0	33.9	47.9	60.0	75.4
Lithuania	15	0	56.6	22.3	27.2	62.0	79.8	99.7
Luxembourg	18	0	73.5	30.9	57.9	78.3	92.2	102.6
Rep of Moldova	<10	<10	66.7	41.2	47.5	73.0	75.2	90.2
The Netherlands	769	10	70	16.7	52.8	71.0	86.9	126.5
North Macedonia	32	1	79.1	27.4	59.5	83.5	96.9	116.0
Norway	150	0	69.2	18.8	46.7	72.4	89.4	110.5
Poland	182	8	59.9	16.1	41.9	58.0	79.6	110.4
Portugal	115	0	65.4	15.9	46.5	64.4	84.7	133.0
Russian Federation	474	200	56.5	12.7	35.4	53.6	76.4	144.2
Serbia	45	1	50.6	18.2	36.7	48.1	62.7	102.0
Slovak Republic	136	3	68	19.1	48.2	73.4	86.4	137.2
Slovenia	31	0	60.9	24.5	36.1	61.3	80.6	107.3
Spain	863	13	68.3	17.6	50.2	67.9	86.7	123.7
Sweden	323	12	71.7	15.8	54.5	74.3	89.1	125.2
Switzerland	444	1	68.1	22.9	50.1	67.0	84.7	120.8
Turkey	152	22	62.4	16.5	39.5	59.8	83.8	119.3
Ukraine	22	5	75.4	29.6	67.6	76.6	93.6	106.0
United Kingdom	5127	4	68.3	12.2	49.6	69.5	86.4	134.9
Total	17467	516	68.5	5.3	49.1	69.4	87.4	151.6

Note: Belarus and Georgia have 0% coverage for adults and are excluded from the table.
Note: Albania, Armenia, Cyprus and Romania have <5 patients aged 18 years or more at FEV_1 measurement and are excluded from this table.
Note: 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 from the previous calendar year.

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

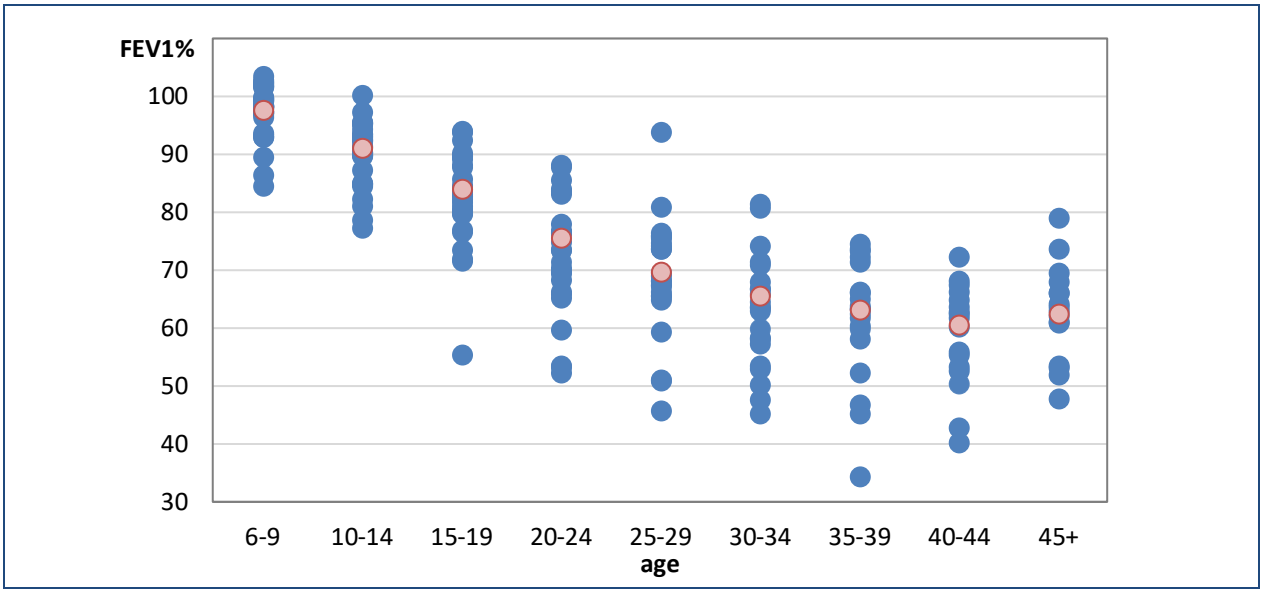
Figure 4.2 FEV_1% of predicted: box-plot, by country and overall. Patients aged 18 years or older who have never had a transplant.



Note: Belarus and Georgia have 0% coverage for adults and are excluded from the table.
Note: Albania, Armenia, Cyprus and Romania have <5 patients aged 18 years or more at FEV_1 measurement and are excluded from this graph.
Note: 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 from the previous calendar year.

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

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



Note: We excluded from the analyses those age groups 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 35-39, and then levels out. The patients in the oldest age groups are patients that survived, and may therefore represent the patients with less disease severity. There is considerable variability between countries.

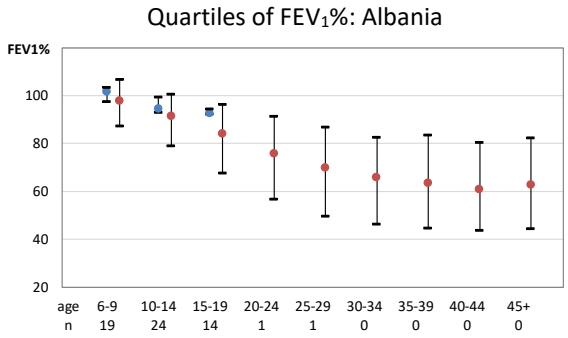
Table 4.3 FEV_1% of predicted: descriptive statistics by age group (patients aged 6 years or older) who have never had a transplant.

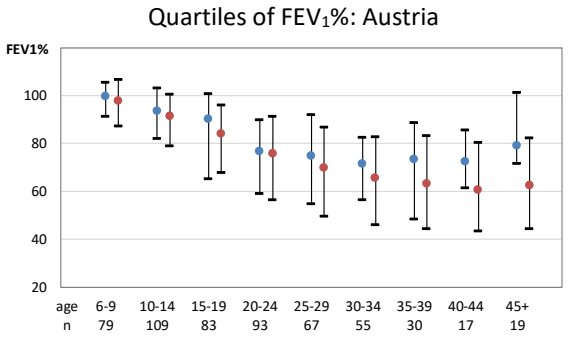
Age at FEV_1 measurement	N	N Miss	Mean	Min	25th pctl	Median	75th pctl	Max
6-9	4353	572	96.6	21.3	87.5	97.7	106.9	182.2
10-14	5246	292	88.5	10.6	79.3	91.3	100.8	171.4
15-19	4645	188	80.6	5.3	68.0	84.1	96.4	181.1
20-24	4040	120	73.5	11.7	56.8	75.8	91.4	144.2
25-29	3431	110	68.4	13.2	49.7	69.9	87.2	138.6
30-34	2654	93	65.2	11.3	46.4	65.7	82.8	137.2
35-39	1956	56	64.2	12.7	44.7	63.3	83.6	151.6
40-44	1384	29	62.6	13.0	43.9	60.7	80.6	131.0
45+	2234	51	64.3	14.2	44.6	62.6	82.6	147.1

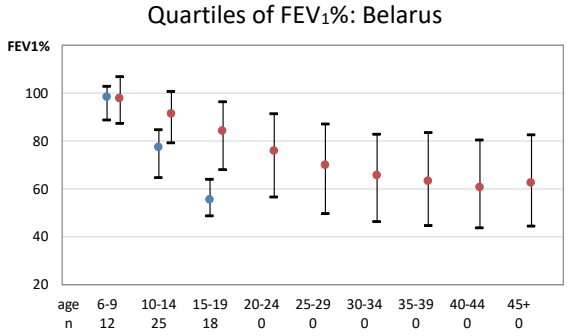
This table shows FEV_1% by age group for the total data-set. The median values reported in this table are shown as red dots in fig 4.3.

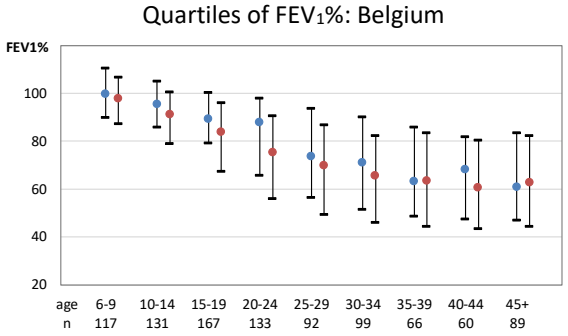
Figure 4.4 Quartiles of FEV$_1$% of predicted by age group and by country. Patients aged 6 years or older and who have never had a transplant.

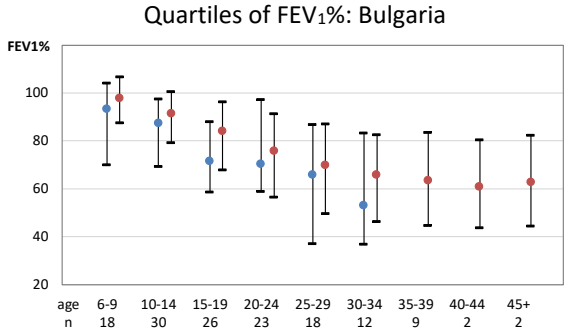
The figures below show the FEV$_1$% in different age groups, separately for each country. The dot shows the median, and the whiskers show the 25th and 75th percentiles (the median, the 25th percentile and the 75th percentile are collectively named "quartiles"). In blue are the quartiles for the country,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

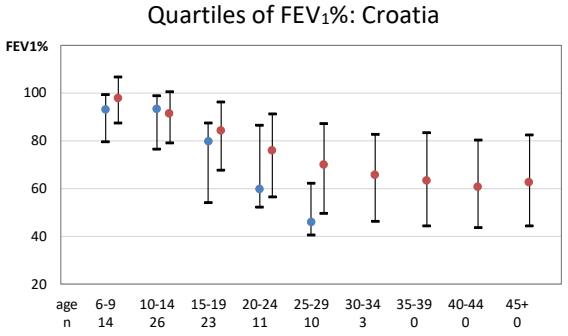


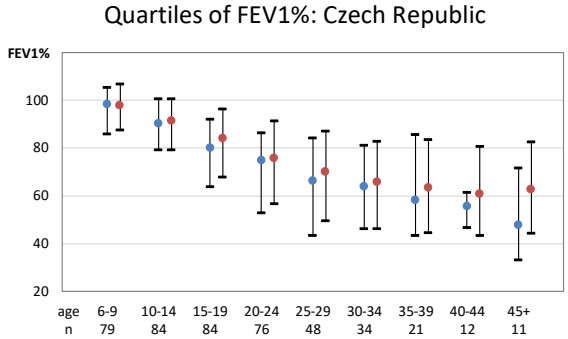


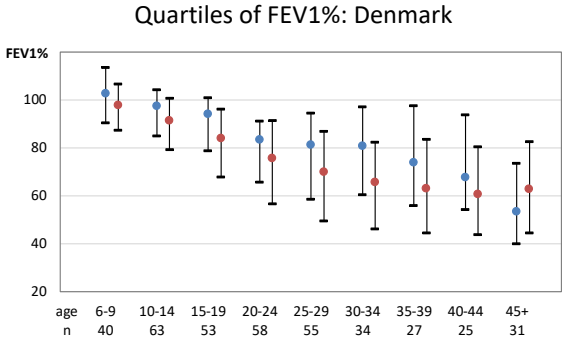




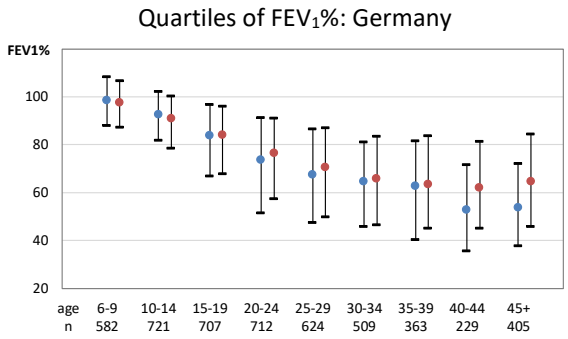


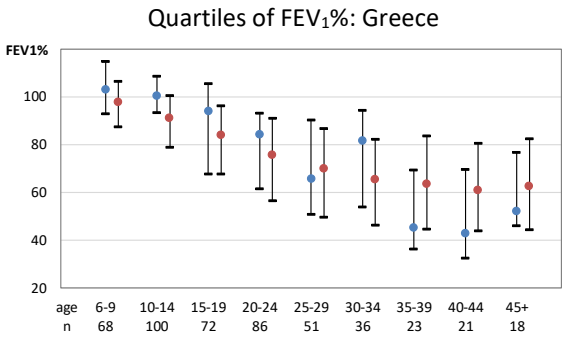


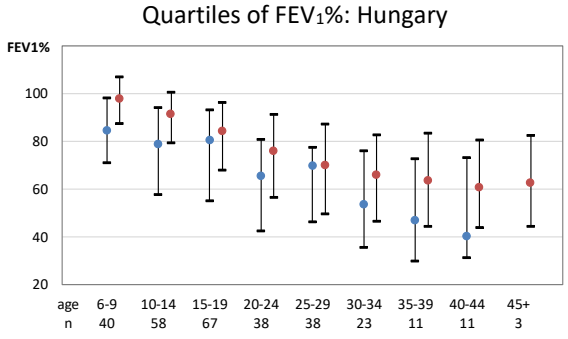


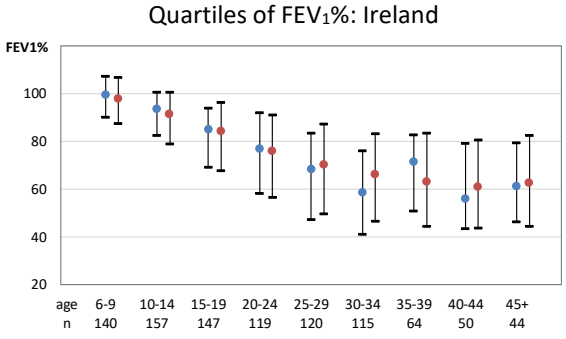


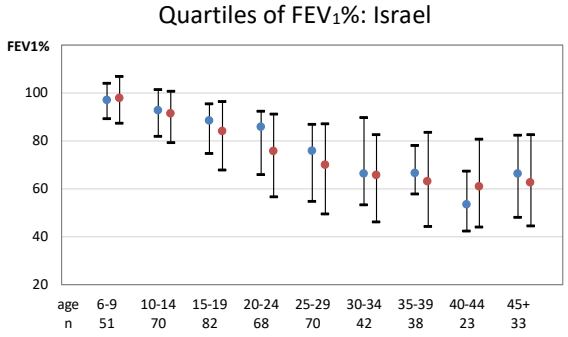
[figure 4.4 continued]

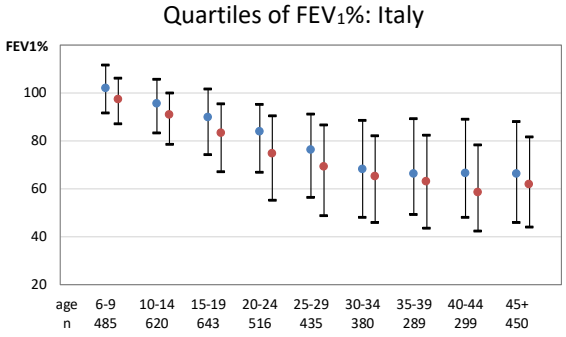


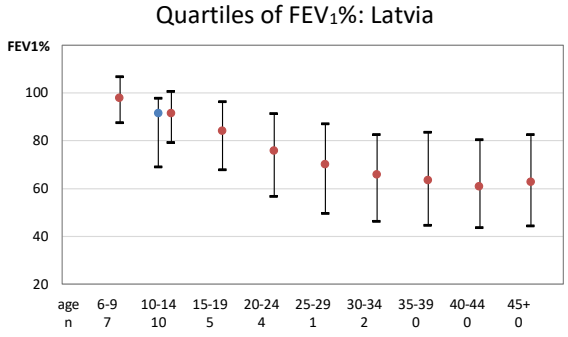


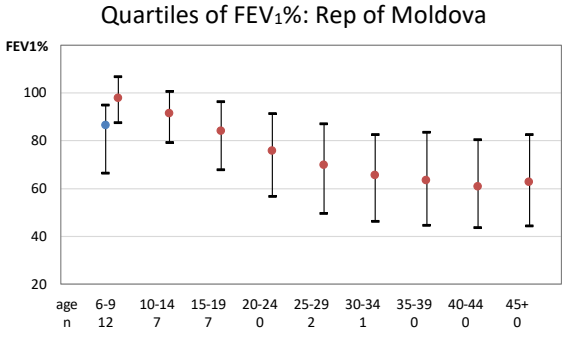


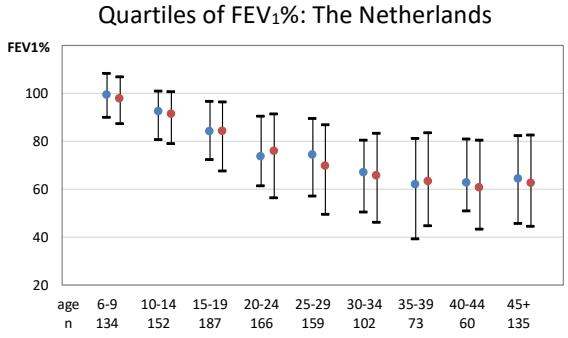


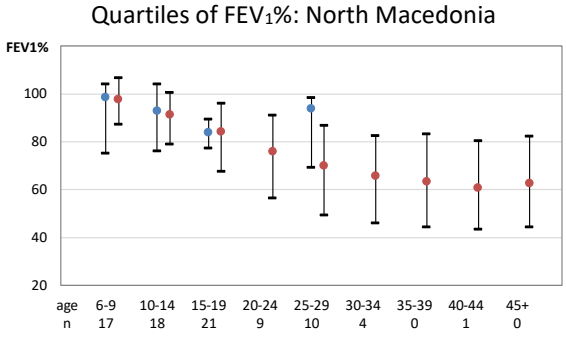






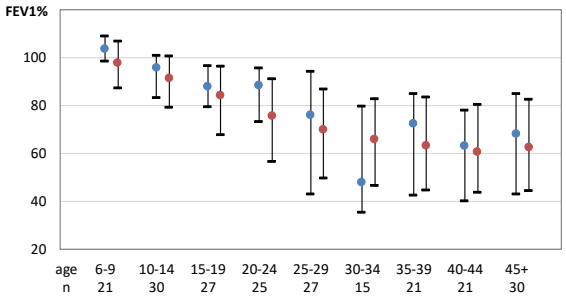




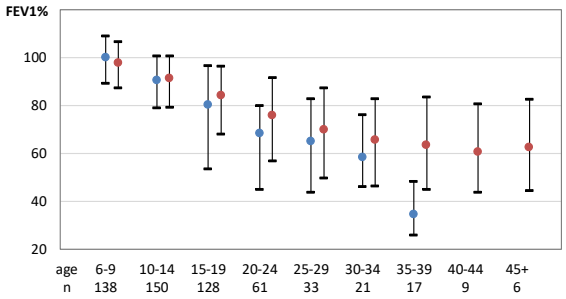


[figure 4.4 continued]

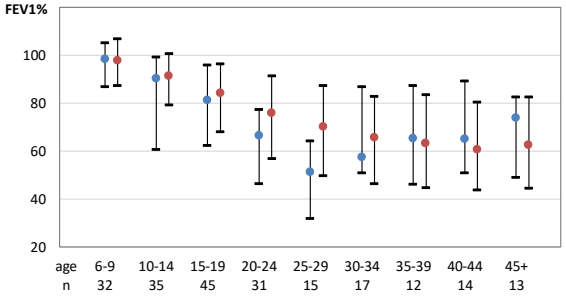
Quartiles of FEV$_1$%: Norway



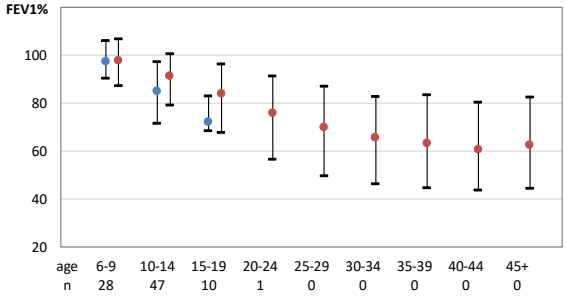
Quartiles of FEV$_1$%: Poland



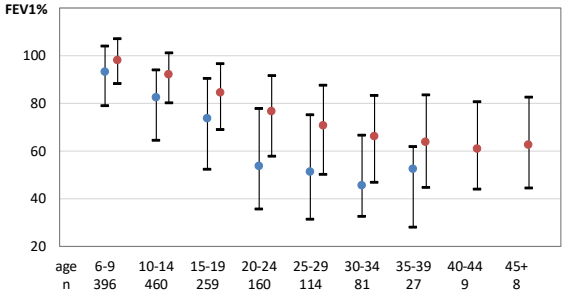
Quartiles of FEV$_1$%: Portugal



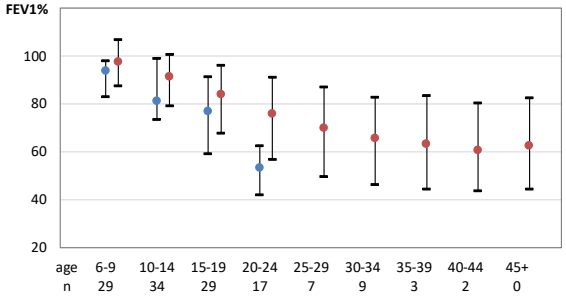
Quartiles of FEV$_1$%: Romania



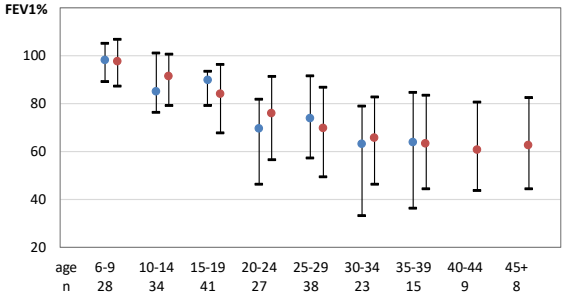
Quartiles of FEV$_1$%: Russian Federation



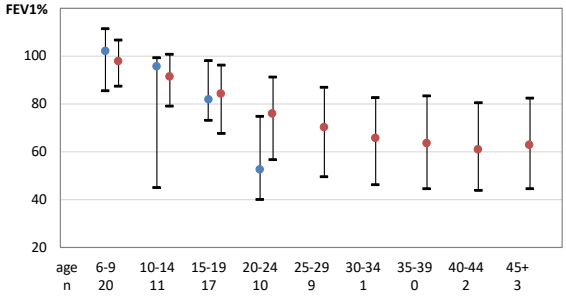
Quartiles of FEV$_1$%: Serbia



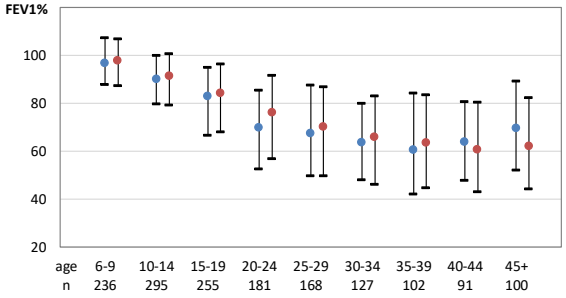
Quartiles of FEV$_1$%: Slovak Republic



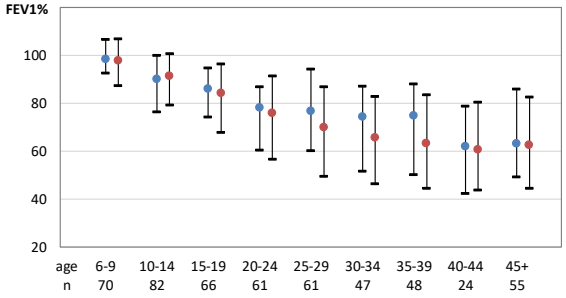
Quartiles of FEV$_1$%: Slovenia



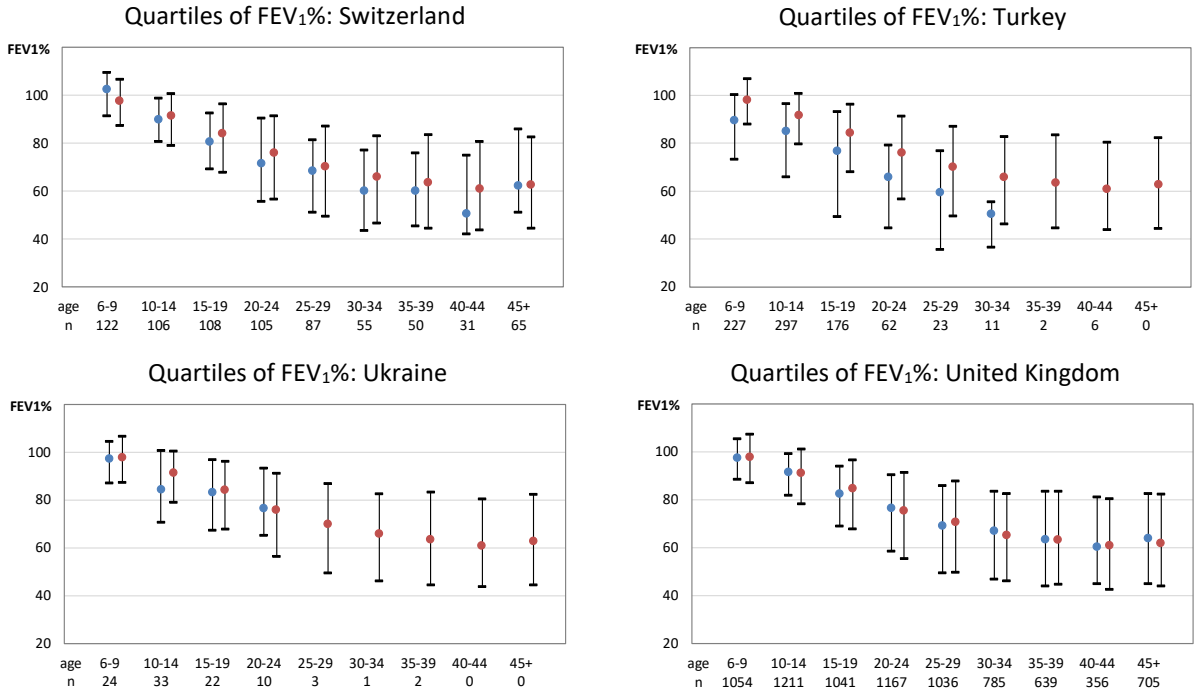
Quartiles of FEV$_1$%: Spain



Quartiles of FEV$_1$%: Sweden

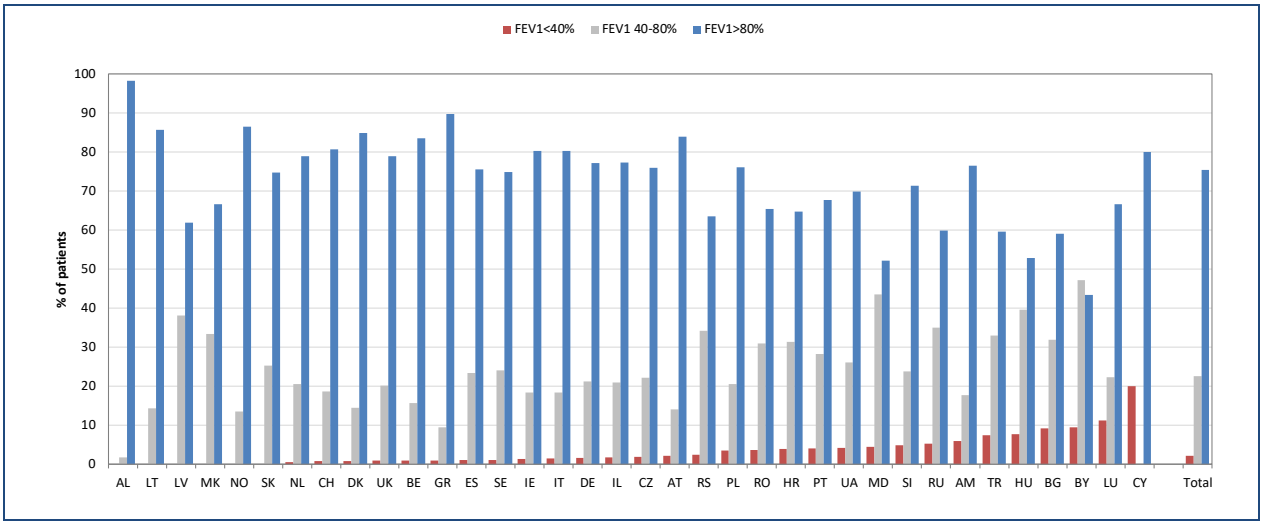


[figure 4.4 continued]



Note: The 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.

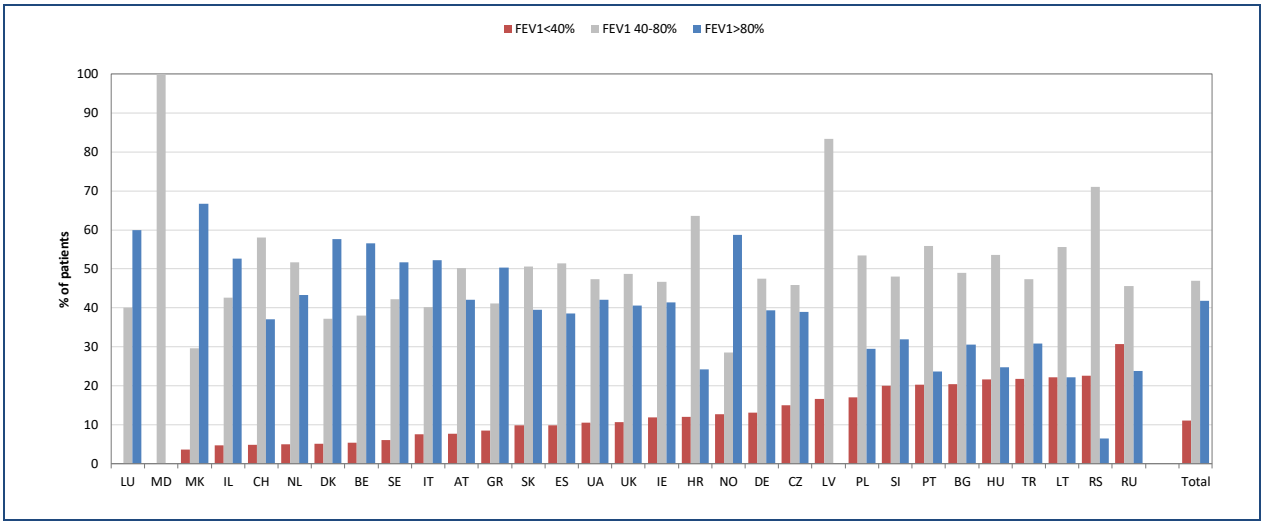
Figure 4.5 FEV_1% of predicted according to severity group and age group, by country and overall. Patients aged 6-17 years who have never had a transplant.



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

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

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

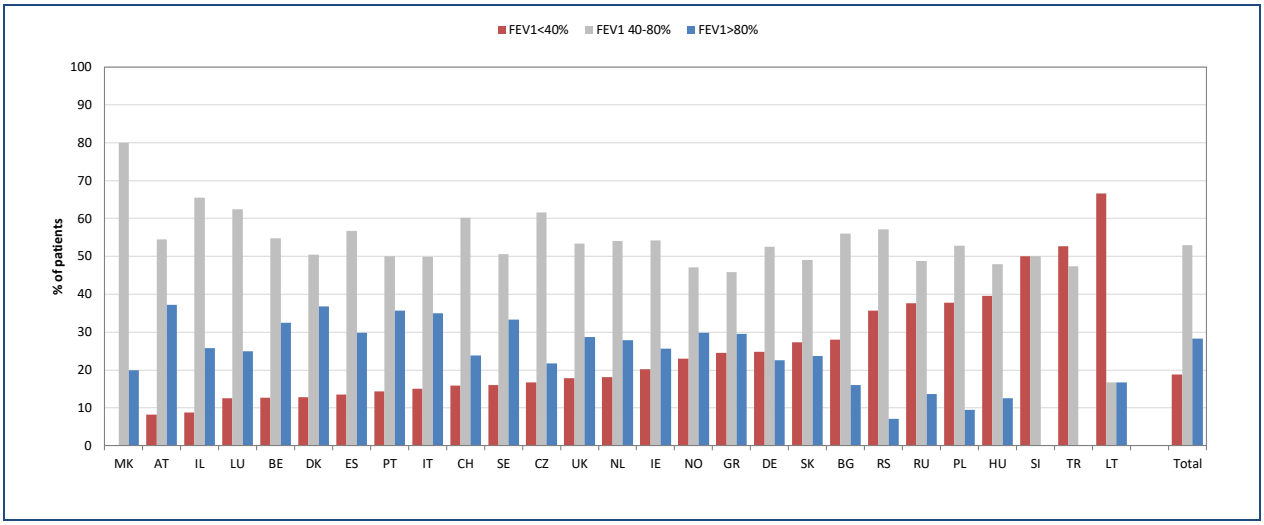


Note: Belarus and Georgia have 0% coverage for adults and are excluded from the table.

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

Note: 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 from the previous calendar year.

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



Note: Belarus and Georgia have 0% coverage for adults and are excluded from the table.

Note: Albania, Armenia, Croatia, Cyprus, Latvia, Rep of Moldova, Romania and Ukraine have <5 patients aged 30 years or more at FEV_1 measurement and are excluded from this graph.

Note: 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 from the previous calendar year.

5. Microbiology

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

In the microbiology category, discrepancies exist between the ECFSPR definition of chronicity and those of the national registries. The ECFSPR definition of chronic infection (see Appendix 2, page 168) is:

Patient should be defined as chronically infected if he/she fulfils the criteria now or has done in recent years and the physician has no reason to think the status has changed, when:

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

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

This year, for the first time, information on intravenous (IV) antibiotic therapy – days on IV antibiotics at home and in hospital – and days in hospital (for any reason, excluding regular check-up) has been collected. The results are not presented in this report, however, since the information is not reliable because of missing data.

Table 5.1 Prevalence of bacterial infection in all patients seen in 2018 who have never had a transplant, by country.

Country	Chronic *Pseudomonas aeruginosa* number (%)			Chronic *Burkholderia cepacia complex species* number (%)			*Haemophilus influenzae* number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	4 (3.60)	77 (69.37)	30 (27.03)	7 (6.31)	104 (93.69)	0 (0.00)	4 (3.60)	102 (91.89)	5 (4.50)
Armenia	0 (0.00)	18 (64.29)	10 (35.71)	19 (67.86)	9 (32.14)	0 (0.00)	3 (10.71)	24 (85.71)	1 (3.57)
Austria	10 (1.42)	514 (73.22)	178 (25.36)	9 (1.28)	673 (95.87)	20 (2.85)	6 (0.85)	474 (67.52)	222 (31.62)
Belarus	0 (0.00)	84 (75.68)	27 (24.32)	0 (0.00)	108 (97.30)	3 (2.70)	0 (0.00)	101 (90.99)	10 (9.01)
Belgium	31 (2.79)	815 (73.23)	267 (23.99)	27 (2.43)	1062 (95.42)	24 (2.16)	0 (0.00)	841 (75.56)	272 (24.44)
Bulgaria	0 (0.00)	89 (46.84)	101 (53.16)	1 (0.53)	188 (98.95)	1 (0.53)	0 (0.00)	176 (92.63)	14 (7.37)
Croatia	7 (6.03)	73 (62.93)	36 (31.03)	6 (5.17)	110 (94.83)	0 (0.00)	4 (3.45)	104 (89.66)	8 (6.90)
Cyprus	8 (53.33)	2 (13.33)	5 (33.33)	8 (53.33)	7 (46.67)	0 (0.00)	2 (13.33)	4 (26.67)	9 (60.00)
Czech Republic	4 (0.70)	451 (79.26)	114 (20.04)	8 (1.41)	519 (91.21)	42 (7.38)	3 (0.53)	482 (84.71)	84 (14.76)
Denmark	0 (0.00)	344 (74.30)	119 (25.70)	0 (0.00)	439 (94.82)	24 (5.18)	0 (0.00)	304 (65.66)	159 (34.34)
Georgia	3 (4.23)	41 (57.75)	27 (38.03)	5 (7.04)	66 (92.96)	0 (0.00)	3 (4.23)	68 (95.77)	0 (0.00)
Germany	176 (3.02)	3724 (63.82)	1935 (33.16)	165 (2.83)	5546 (95.05)	124 (2.13)	146 (2.50)	4637 (79.47)	1052 (18.03)
Greece	31 (5.62)	270 (48.91)	251 (45.47)	9 (1.63)	543 (98.37)	0 (0.00)	12 (2.17)	498 (90.22)	42 (7.61)
Hungary	5 (1.14)	280 (63.93)	153 (34.93)	6 (1.37)	422 (96.35)	10 (2.28)	438 (100)	0 -	0 -
Ireland[1]	7 (0.63)	836 (74.84)	274 (24.53)	8 (0.72	1087 (97.31	22 (1.97	6 (0.54)	851 (76.19)	260 (23.28)
Israel	17 (3.28)	266 (51.25)	236 (45.47)	22 (4.24)	493 (94.99)	4 (0.77)	20 (3.85)	431 (83.04)	68 (13.10)
Italy[2]	5 (0.10)	3642 (69.93)	1561 (29.97)	28 (0.54)	5081 (97.56)	99 (1.90)	4117 (79.05)	1062 (20.39)	29 (0.56)
Latvia	6 (16.22)	24 (64.86)	7 (18.92)	7 (18.92)	29 (78.38)	1 (2.70)	6 (16.22)	22 (59.46)	9 (24.32)
Lithuania	2 (8.33)	16 (66.67)	6 (25.00)	1 (4.17)	21 (87.50)	2 (8.33)	2 (8.33)	18 (75.00)	4 (16.67)
Luxembourg	0 (0.00)	29 (80.56)	7 (19.44)	0 (0.00)	33 (91.67)	3 (8.33)	0 (0.00)	27 (75.00)	9 (25.00)
Rep of Moldova	2 (3.57)	28 (50.00)	26 (46.43)	0 (0)	56 (100)	0 (0)	3 (5.36)	49 (87.50)	4 (7.14)
The Netherlands	228 (16.91)	777 (57.64)	343 (25.45)	44 (3.26)	1280 (94.96)	24 (1.78)	44 (3.26)	1083 (80.34)	221 (16.39)
North Macedonia	6 (5.08)	74 (62.71)	38 (32.20)	7 (5.93)	111 (94.07)	0 (0.00)	6 (5.08)	109 (92.37)	3 (2.54)

[1] Ireland: chronicity for *Pseudomonas aeruginosa* and *Burkholderia cepacia complex species* is defined as: at least 3 or more positive isolates during the last 12 months preceding the last reported culture in 2018.

[2] Italy: chronicity for *Pseudomonas aeruginosa* and *Burkholderia cepacia complex species* is defined as: at least 3 or more positive cultures during 2018.

[table 5.1 continued]

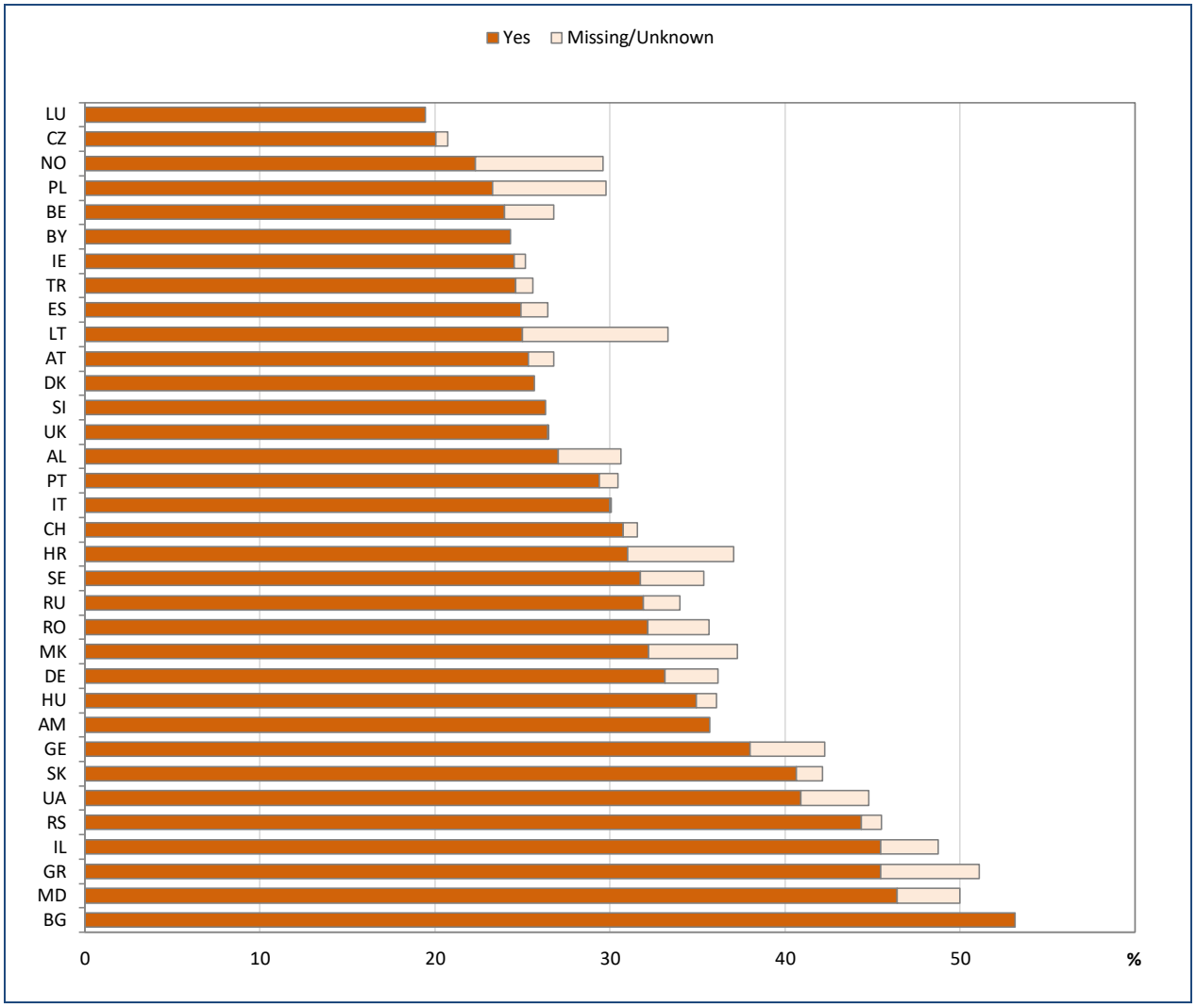
Country	Chronic *Pseudomonas aeruginosa* number (%)			Chronic *Burkholderia cepacia complex species* number (%)			*Haemophilus influenzae* number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Norway	19 (7.31)	183 (70.38)	58 (22.31)	28 (10.77)	226 (86.92)	6 (2.31)	15 (5.77)	163 (62.69)	82 (31.54)
Poland	56 (6.49)	606 (70.22)	201 (23.29)	59 (6.84)	790 (91.54)	14 (1.62)	28 (3.24)	693 (80.30)	142 (16.45)
Portugal	3 (1.08)	194 (69.53)	82 (29.39)	4 (1.43)	260 (93.19)	15 (5.38)	3 (1.08)	205 (73.48)	71 (25.45)
Romania	7 (3.52)	128 (64.32)	64 (32.16)	27 (13.57)	172 (86.43)	0 (0.00)	6 (3.02)	186 (93.47)	7 (3.52)
Russian Federation	65 (2.10)	2042 (65.98)	988 (31.92)	61 (1.97)	2859 (92.37)	175 (5.65)	157 (5.07)	2799 (90.44)	139 (4.49)
Serbia	2 (1.12)	97 (54.49)	79 (44.38)	1 (0.56)	161 (90.45)	16 (8.99)	4 (2.25)	88 (49.44)	86 (48.31)
Slovak Republic	4 (1.47)	158 (57.88)	111 (40.66)	4 (1.47)	253 (92.67)	16 (5.86)	5 (1.83)	239 (87.55)	29 (10.62)
Slovenia	0 (0.00)	70 (73.68)	25 (26.32)	1 (1.05)	94 (98.95)	0 (0.00)	1 (1.05)	53 (55.79)	41 (43.16)
Spain	31 (1.54)	1478 (73.53)	501 (24.93)	42 (2.09)	1878 (93.43)	90 (4.48)	40 (1.99)	1607 (79.95)	363 (18.06)
Sweden	22 (3.62)	393 (64.64)	193 (31.74)	0 (0.00)	597 (98.19)	11 (1.81)	0 (0.00)	487 (80.10)	121 (19.90)
Switzerland	7 (0.79)	603 (68.44)	271 (30.76)	10 (1.14)	851 (96.59)	20 (2.27)	14 (1.59)	646 (73.33)	221 (25.09)
Turkey	18 (1.00)	1339 (74.39)	443 (24.61)	22 (1.22)	1767 (98.17)	11 (0.61)	688 (38.22)	1002 (55.67)	110 (6.11)
Ukraine	6 (3.90)	85 (55.19)	63 (40.91)	24 (15.58)	127 (82.47)	3 (1.95)	5 (3.25)	141 (91.56)	8 (5.19)
United Kingdom[3]	4 (0.04)	6973 (73.49)	2511 (26.47)	4 (0.04)	9153 (96.47)	331 (3.49)	4 (0.04)	8045 (84.79)	1439 (15.17)
Total	796 (2.04)	26823 (68.85)	11341 (29.11)	674 (1.73)	37175 (95.42)	1111 (2.85)			

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

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

Table 5.1 shows, separately by country, the frequency of chronic *Pseudomonas aeruginosa*, chronic *Burkholderia cepacia complex species* and *Haemophilus influenza*, which is not chronic. 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 the culture techniques employed.

Figure 5.1 Prevalence of chronic Pseudomonas aeruginosa infection in all patients seen in 2018 who have never had a transplant, by country.

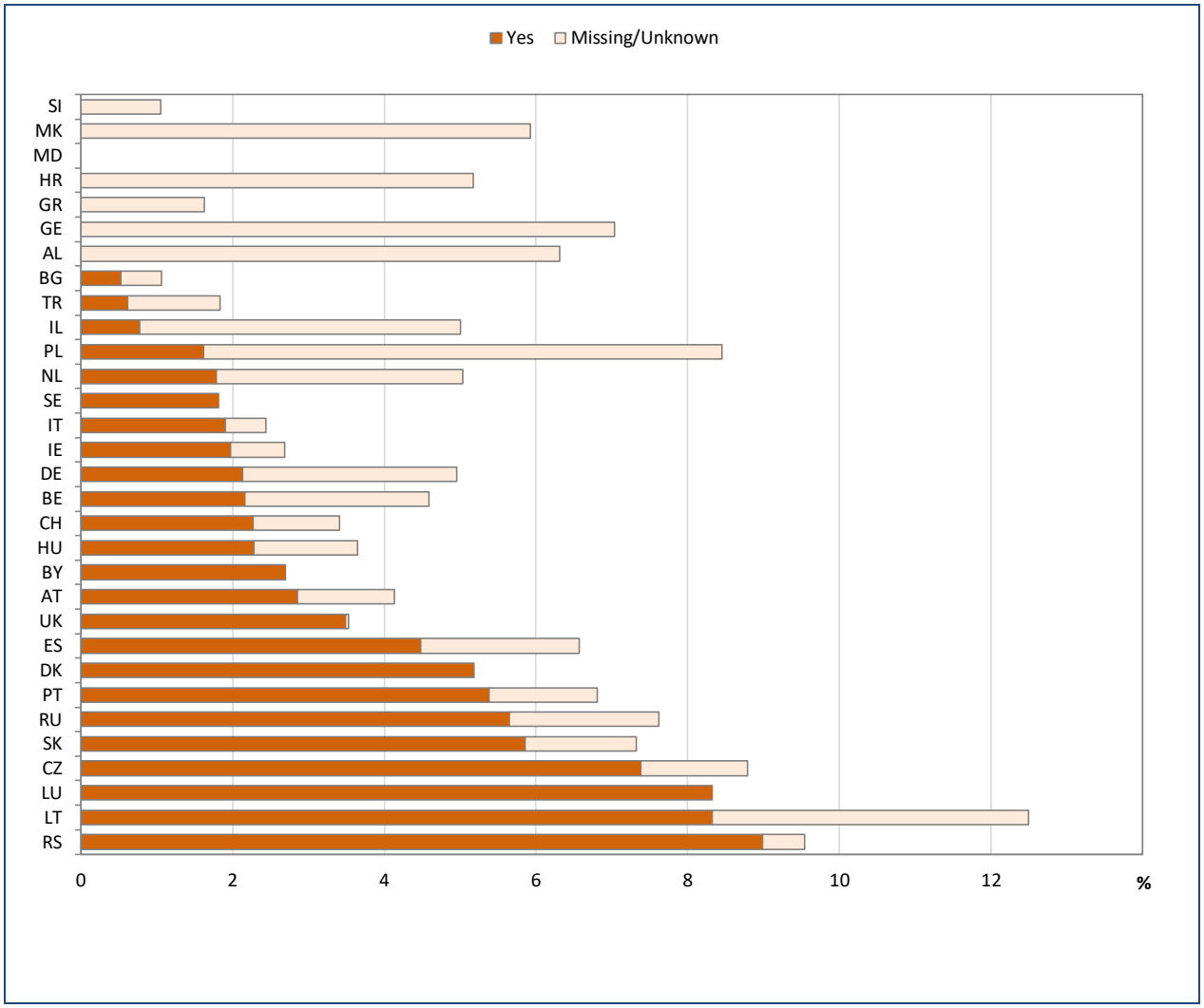


Note: We excluded from the graph the countries for which the information on *Pseudomonas aeruginosa* was missing for more than 10% of the patients.

Note: Ireland: chronicity for *Pseudomonas aeruginosa* is defined as: at least 3 or more positive isolates during the last 12 months preceding the last reported culture in 2018.
Italy: chronicity for *Pseudomonas aeruginosa* is defined as: at least 3 or more positive cultures during 2018.
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 2018 who have never had a transplant, by country.

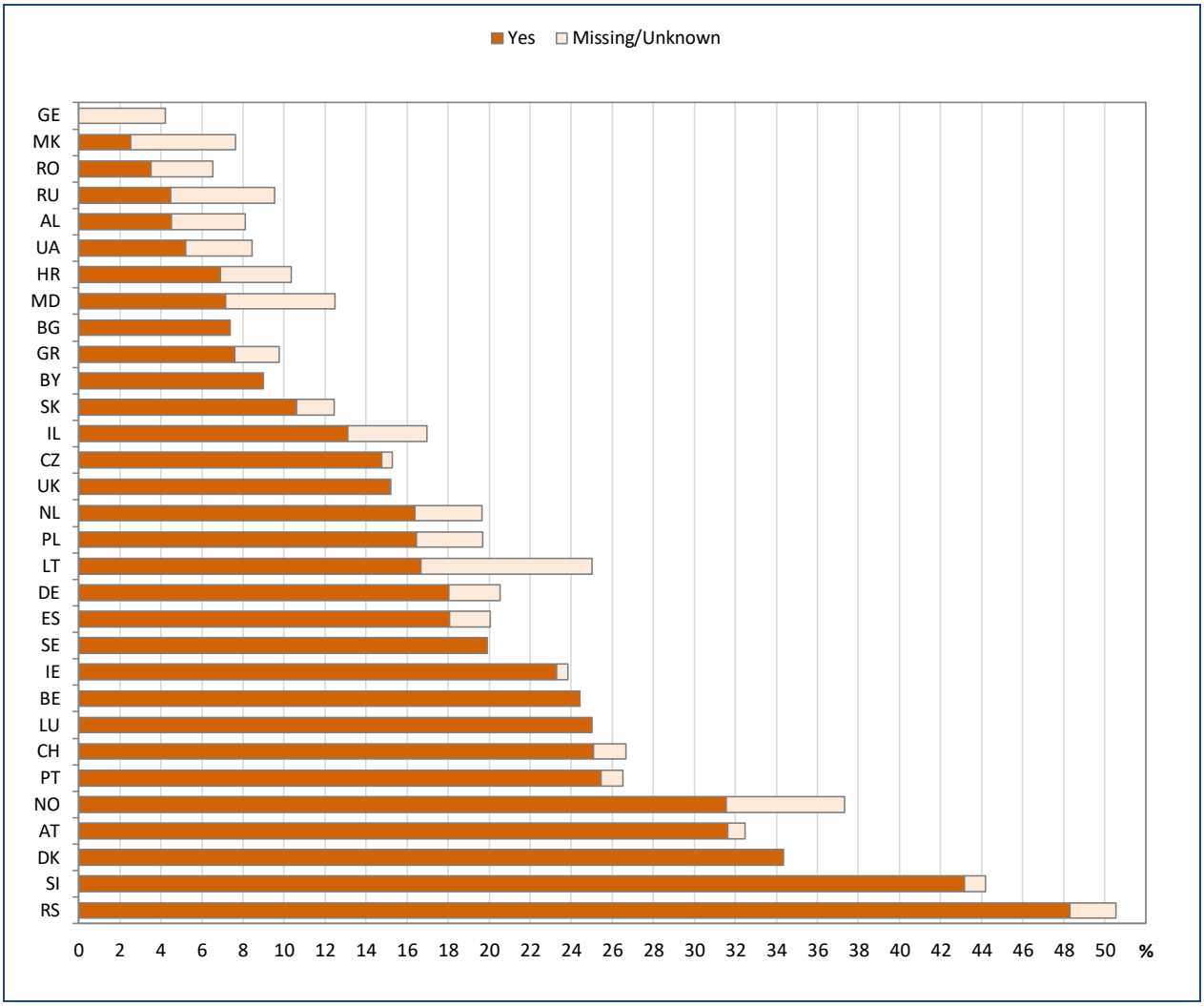


Note: We excluded from the graph the countries for which the information on *Burkholderia cepacia complex species* was missing for more than 10% of the patients.

Note: Ireland: chronicity for *Burkholderia cepacia complex species* is defined as: at least 3 or more positive isolates during the last 12 months preceding the last reported culture in 2018.
Italy: chronicity for *Burkholderia cepacia complex species* is defined as: at least 3 or more positive cultures during 2018.
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 Haemophilus influenzae infection in all patients seen in 2018 who have never had a transplant, by country.



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

The horizontal bars represent the percentage of patients with *Haemophilus influenzae* infection (in dark orange) and the percentage of patients where information on *Haemophilus influenzae* 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 bacterial infection in children seen in 2018 who have never had a transplant, by country.

Country	Chronic *Pseudomonas aeruginosa* number (%)			Chronic *Burkholderia cepacia complex species* number (%)			*Haemophilus influenzae* number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	3 (2.83)	74 (69.81)	29 (27.36)	6 (5.66)	100 (94.34)	0 (0.00)	3 (2.83)	98 (92.45)	5 (4.72)
Armenia	0 (0.00)	18 (64.29)	10 (35.71)	19 (67.86)	9 (32.14)	0 (0.00)	3 (10.71)	24 (85.71)	1 (3.57)
Austria	1 (0.27)	340 (91.15)	32 (8.58)	1 (0.27)	369 (98.93)	3 (0.80)	1 (0.27)	222 (59.52)	150 (40.21)
Belarus	0 (0.00)	82 (75.93)	26 (24.07)	0 (0.00)	105 (97.22)	3 (2.78)	0 (0.00)	99 (91.67)	9 (8.33)
Belgium	13 (2.80)	417 (89.68)	35 (7.53)	11 (2.37)	450 (96.77)	4 (0.86)	0 (0.00)	288 (61.94)	177 (38.06)
Bulgaria	0 (0.00)	69 (61.61)	43 (38.39)	1 (0.89)	110 (98.21)	1 (0.89)	0 (0.00)	103 (91.96)	9 (8.04)
Croatia	5 (6.41)	61 (78.21)	12 (15.38)	5 (6.41)	73 (93.59)	0 (0.00)	3 (3.85)	67 (85.90)	8 (10.26)
Cyprus	7 (58.33	1 (8.33))	4 (33.33)	7 (58.33)	5 (41.67)	0 (0.00)	1 (8.33)	3 (25.00)	8 (66.67)
Czech Republic	1 (0.31)	291 (91.51)	26 (8.18)	5 (1.57)	309 (97.17)	4 (1.26)	0 (0.00)	258 (81.13)	60 (18.87)
Denmark	0 (0.00)	188 (93.53)	13 (6.47)	0 (0.00)	199 (99.00)	2 (1.00)	0 (0.00)	103 (51.24)	98 (48.76)
Georgia	3 (4.23)	41 (57.75)	27 (38.03)	5 (7.04)	66 (92.96)	0 (0.00)	3 (4.23)	68 (95.77)	0 (0.00)
Germany	44 (1.73)	2258 (88.62)	246 (9.65)	38 (1.49)	2491 (97.76)	19 (0.75)	34 (1.33)	1745 (68.49)	769 (30.18)
Greece	24 (8.73)	170 (61.82)	81 (29.45)	2 (0.73)	273 (99.27)	0 (0.00)	4 (1.45)	255 (92.73)	16 (5.82)
Hungary	1 (0.39	186 (73.23)	67 (26.38)	1 (0.39)	249 (98.03)	4 (1.57)	254 (100)	0 -	0 -
Ireland[1]	2 (0.39)	470 (92.70)	35 (6.90)	1 (0.20)	501 (98.82)	5 (0.99)	1 (0.20)	318 (62.72)	188 (37.08)
Israel	10 (4.85)	144 (69.90)	52 (25.24)	13 (6.31)	193 (93.69)	0 (0.00)	11 (5.34)	155 (75.24)	40 (19.42)
Italy[2]	2 (0.09)	2073 (89.90)	231 (10.02)	11 (0.48)	2292 (99.39)	3 (0.13)	1886 (81.79)	401 (17.39)	19 (0.82)
Latvia	6 (21.43)	21 (75.00)	1 (3.57)	6 (21.43)	21 (75.00)	1 (3.57)	6 (21.43)	15 (53.57)	7 (25.00)
Lithuania	1 (11.11)	6 (66.67)	2 (22.22)	0 (0.00)	8 (88.89)	1 (11.11)	0 (0.00)	7 (77.78)	2 (22.22)
Luxembourg	0 (0.00)	16 (88.89)	2 (11.11)	0 (0)	18 (100)	0 (0)	0 (0.00)	11 (61.11)	7 (38.89)
Rep of Moldova	0 (0.00)	27 (61.36)	17 (38.64)	0 (0)	44 (100)	0 (0)	0 (0.00)	40 (90.91)	4 (9.09)
The Netherlands	5 (0.93)	469 (87.34)	63 (11.73)	5 (0.93)	527 (98.14)	5 (0.93)	5 (0.93)	381 (70.95)	151 (28.12)
North Macedonia	3 (3.61)	61 (73.49)	19 (22.89)	4 (4.82)	79 (95.18)	0 (0.00)	3 (3.61)	77 (92.77)	3 (3.61)

[1] Ireland: chronicity for *Pseudomonas aeruginosa* and *Burkholderia cepacia complex species* is defined as: at least 3 or more positive isolates during the last 12 months preceding the last reported culture in 2018.

[2] Italy: chronicity for *Pseudomonas aeruginosa* and *Burkholderia cepacia complex species* is defined as: at least 3 or more positive cultures during 2018.

[table 5.2 continued]

Country	Chronic *Pseudomonas aeruginosa* number (%)			Chronic *Burkholderia cepacia complex species* number (%)			*Haemophilus influenzae* number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Norway	1 (0.96)	97 (93.27)	6 (5.77)	1 (0.96)	102 (98.08)	1 (0.96)	1 (0.96)	53 (50.96)	50 (48.08)
Poland	37 (5.75)	538 (83.54)	69 (10.71)	40 (6.21)	599 (93.01)	5 (0.78)	13 (2.02)	503 (78.11)	128 (19.88)
Portugal	1 (0.66)	116 (76.32)	35 (23.03)	2 (1.32)	144 (94.74)	6 (3.95)	1 (0.66)	100 (65.79)	51 (33.55)
Romania	7 (3.59)	127 (65.13)	61 (31.28)	27 (13.85)	168 (86.15)	0 (0.00)	6 (3.08)	182 (93.33)	7 (3.59)
Russian Federation	49 (2.08)	1701 (72.23)	605 (25.69)	44 (1.87)	2225 (94.48)	86 (3.65)	110 (4.67)	2119 (89.98)	126 (5.35)
Serbia	2 (1.55)	84 (65.12)	43 (33.33)	1 (0.78)	119 (92.25)	9 (6.98)	3 (2.33)	53 (41.09)	73 (56.59)
Slovak Republic	1 (0.80)	81 (64.80)	43 (34.40)	1 (0.80)	121 (96.80)	3 (2.40)	1 (0.80)	112 (89.60)	12 (9.60)
Slovenia	0 (0.00)	52 (88.14)	7 (11.86)	0 (0)	59 (100)	0 (0)	1 (1.69)	28 (47.46)	30 (50.85)
Spain	11 (1.02)	949 (88.03)	118 (10.95)	17 (1.58)	1041 (96.57)	20 (1.86)	14 (1.30)	789 (73.19)	275 (25.51)
Sweden	20 (7.66)	212 (81.23)	29 (11.11)	0 (0.00)	259 (99.23)	2 (0.77)	0 (0.00)	182 (69.73)	79 (30.27)
Switzerland	3 (0.71)	378 (89.15)	43 (10.14)	4 (0.94	417 (98.35	3 (0.71	3 (0.71)	270 (63.68)	151 (35.61)
Turkey	16 (0.99)	1257 (77.98)	339 (21.03)	20 (1.24)	1589 (98.57)	3 (0.19)	609 (37.78)	904 (56.08)	99 (6.14)
Ukraine	3 (2.44)	77 (62.60)	43 (34.96)	19 (15.45)	103 (83.74)	1 (0.81)	3 (2.44)	113 (91.87)	7 (5.69)
United Kingdom[3]	1 (0.02)	3917 (93.00)	294 (6.98)	1 (0.02)	4151 (98.55)	60 (1.42)	1 (0.02)	3187 (75.66)	1024 (24.31)
Total	283 (1.40)	17069 (84.67)	2808 (13.93)	318 (1.58)	19588 (97.16)	254 (1.26)			

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

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

Table 5.2 shows, separately by country, the frequency of chronic *Pseudomonas aeruginosa*, chronic *Burkholderia cepacia complex species* and *Haemophilus inflenzae* 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 bacterial infection in adults seen in 2018 who have never had a transplant, by country.

Country	Chronic *Pseudomonas aeruginosa* number (%)			Chronic *Burkholderia cepacia complex species* number (%)			*Haemophilus influenzae* number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	1 (20.00)	3 (60.00)	1 (20.00)	1 (20.00)	4 (80.00)	0 (0.00)	1 (20.00)	4 (80.00)	0 (0.00)
Austria	9 (2.74)	174 (52.89)	146 (44.38)	8 (2.43)	304 (92.40)	17 (5.17)	5 (1.52)	252 (76.60)	72 (21.88)
Belgium	18 (2.78)	398 (61.42)	232 (35.80)	16 (2.47)	612 (94.44)	20 (3.09)	0 (0.00)	553 (85.34)	95 (14.66)
Bulgaria	0 (0.00)	20 (25.64)	58 (74.36)	0 (0)	78 (100)	0 (0)	0 (0.00)	73 (93.59)	5 (6.41)
Croatia	2 (5.26)	12 (31.58)	24 (63.16)	1 (2.63)	37 (97.37)	0 (0.00)	1 (2.63)	37 (97.37)	0 (0.00)
Czech Republic	3 (1.20)	160 (63.75)	88 (35.06)	3 (1.20)	210 (83.67)	38 (15.14)	3 (1.20)	224 (89.24)	24 (9.56)
Denmark	0 (0.00)	156 (59.54)	106 (40.46)	0 (0.00)	240 (91.60)	22 (8.40)	0 (0.00)	201 (76.72)	61 (23.28)
Germany	132 (4.02)	1466 (44.60)	1689 (51.38)	127 (3.86)	3055 (92.94)	105 (3.19)	112 (3.41)	2892 (87.98)	283 (8.61)
Greece	7 (2.53)	100 (36.10)	170 (61.37)	7 (2.53)	270 (97.47)	0 (0.00)	8 (2.89)	243 (87.73)	26 (9.39)
Hungary	4 (2.17)	94 (51.09)	86 (46.74)	5 (2.72)	173 (94.02)	6 (3.26)	184 (100)	0 -	0 -
Ireland[1]	5 (0.82)	366 (60.00)	239 (39.18)	7 (1.15)	586 (96.07)	17 (2.79)	5 (0.82)	533 (87.38)	72 (11.80)
Israel	7 (2.24)	122 (38.98)	184 (58.79)	9 (2.88)	300 (95.85)	4 (1.28)	9 (2.88)	276 (88.18)	28 (8.95)
Italy[2]	3 (0.10)	1569 (54.07)	1330 (45.83)	17 (0.59)	2789 (96.11)	96 (3.31)	2231 (76.88)	661 (22.78)	10 (0.34)
Latvia	0 (0.00)	3 (33.33)	6 (66.67)	1 (11.11)	8 (88.89)	0 (0.00)	0 (0.00)	7 (77.78)	2 (22.22)
Lithuania	1 (6.67)	10 (66.67)	4 (26.67)	1 (6.67)	13 (86.67)	1 (6.67)	2 (13.33)	11 (73.33)	2 (13.33)
Luxembourg	0 (0.00)	13 (72.22)	5 (27.78)	0 (0.00)	15 (83.33)	3 (16.67)	0 (0.00)	16 (88.89)	2 (11.11)
Rep of Moldova	2 (16.67	1 (8.33))	9 (75.00)	0 (0)	12 (100)	0 (0)	3 (25.00)	9 (75.00)	0 (0.00)
The Netherlands	223 (27.50)	308 (37.98)	280 (34.53)	39 (4.81)	753 (92.85)	19 (2.34)	39 (4.81)	702 (86.56)	70 (8.63)
North Macedonia	3 (8.57)	13 (37.14)	19 (54.29)	3 (8.57)	32 (91.43)	0 (0.00)	3 (8.57)	32 (91.43)	0 (0.00)
Norway	18 (11.54	86 (55.13	52 (33.33	27 (17.31)	124 (79.49)	5 (3.21)	14 (8.97)	110 (70.51)	32 (20.51)
Poland	19 (8.68)	68 (31.05)	132 (60.27)	19 (8.68)	191 (87.21)	9 (4.11)	15 (6.85)	190 (86.76)	14 (6.39)

[1] Ireland: chronicity for *Pseudomonas aeruginosa* and *Burkholderia cepacia complex species* is defined as: at least 3 or more positive isolates during the last 12 months preceding the last reported culture in 2018.

[2] Italy: chronicity for *Pseudomonas aeruginosa* and *Burkholderia cepacia complex species* is defined as: at least 3 or more positive cultures during 2018.

[table 5.3 continued]

Country	Chronic *Pseudomonas aeruginosa* number (%)			Chronic *Burkholderia cepacia complex species* number (%)			*Haemophilus influenzae* number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Portugal	2 (1.57)	78 (61.42)	47 (37.01)	2 (1.57)	116 (91.34)	9 (7.09)	2 (1.57)	105 (82.68)	20 (15.75)
Russian Federation	16 (2.16)	341 (46.08)	383 (51.76)	17 (2.30)	634 (85.68)	89 (12.03)	47 (6.35)	680 (91.89)	13 (1.76)
Serbia	0 (0.00)	13 (26.53)	36 (73.47)	0 (0.00)	42 (85.71)	7 (14.29)	1 (2.04)	35 (71.43)	13 (26.53)
Slovak Republic	3 (2.03)	77 (52.03)	68 (45.95)	3 (2.03)	132 (89.19)	13 (8.78)	4 (2.70)	127 (85.81)	17 (11.49)
Slovenia	0 (0.00)	18 (50.00)	18 (50.00)	1 (2.78)	35 (97.22)	0 (0.00)	0 (0.00)	25 (69.44)	11 (30.56)
Spain	20 (2.15)	529 (56.76)	383 (41.09)	25 (2.68)	837 (89.81)	70 (7.51)	26 (2.79)	818 (87.77)	88 (9.44)
Sweden	2 (0.58)	181 (52.16)	164 (47.26)	0 (0.00)	338 (97.41)	9 (2.59)	0 (0.00)	305 (87.90)	42 (12.10)
Switzerland	4 (0.88)	225 (49.23)	228 (49.89)	6 (1.31)	434 (94.97)	17 (3.72)	11 (2.41)	376 (82.28)	70 (15.32)
Turkey	2 (1.06)	82 (43.62)	104 (55.32)	2 (1.06)	178 (94.68)	8 (4.26)	79 (42.02)	98 (52.13)	11 (5.85)
Ukraine	3 (9.68)	8 (25.81)	20 (64.52)	5 (16.13)	24 (77.42)	2 (6.45)	2 (6.45)	28 (90.32)	1 (3.23)
United Kingdom[3]	3 (0.06)	3056 (57.92)	2217 (42.02)	3 (0.06)	5002 (94.81)	271 (5.14)	3 (0.06)	4858 (92.08)	415 (7.87)
Total	513 (2.73)	9754 (51.88)	8533 (45.39)	356 (1.89)	17587 (93.55)	857 (4.56)			

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

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

Note: Belarus and Georgia have 0% coverage for adults and are excluded from the table.

Note: Armenia, Belarus, Cyprus and Romania have <5 patients aged 18 years or more at 31/12/2018 and are not shown in this table, but are considered in the total.

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

Table 5.4 Prevalence of chronic Staphylococcus aureus and methicillin-resistant Staphylococcus aureus (MRSA) in all patients seen in 2018 who have never had a transplant, by country.

Country	Chronic *Staphylococcus aureus* number (%)			MRSA number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	3 (2.70)	72 (64.86)	36 (32.43)	3 (2.70)	106 (95.50)	2 (1.80)
Armenia	0 (0.00)	3 (10.71)	25 (89.29)	22 (78.57)	4 (14.29)	2 (7.14)
Austria	9 (1.28)	250 (35.61)	443 (63.11)	12 (1.71)	676 (96.30)	14 (1.99)
Belarus	0 (0.00)	50 (45.05)	61 (54.95)	111 (100)	0 -	0 -
Belgium	1113 (100)	0 -	0 -	0 (0.00)	1045 (93.89)	68 (6.11)
Bulgaria	0 (0.00)	136 (71.58)	54 (28.42)	2 (1.05)	184 (96.84)	4 (2.11)
Croatia	6 (5.17)	68 (58.62)	42 (36.21)	4 (3.45)	106 (91.38)	6 (5.17)
Cyprus	8 (53.33)	3 (20.00)	4 (26.67)	2 (13.33)	13 (86.67)	0 (0.00)
Czech Republic	5 (0.88)	293 (51.49)	271 (47.63)	5 (0.88)	548 (96.31)	16 (2.81)
Denmark	0 (0.00)	333 (71.92)	130 (28.08)	0 (0.00)	460 (99.35)	3 (0.65)
Georgia	4 (5.63)	35 (49.30)	32 (45.07)	3 (4.23)	68 (95.77)	0 (0.00)
Germany	170 (2.91)	3232 (55.39)	2433 (41.70)	147 (2.52)	5384 (92.27)	304 (5.21)
Greece	14 (2.54)	373 (67.57)	165 (29.89)	10 (1.81)	471 (85.33)	71 (12.86)
Hungary	4 (0.91)	203 (46.35)	231 (52.74)	4 (0.91)	399 (91.10)	35 (7.99)
Ireland[1]	6 (0.54)	706 (63.21)	405 (36.26)	6 (0.54)	1030 (92.21)	81 (7.25)
Israel	18 (3.47)	295 (56.84)	206 (39.69)	20 (3.85)	469 (90.37)	30 (5.78)
Italy[2]	140 (2.69)	2941 (56.47)	2127 (40.84)	4117 (79.05)	978 (18.78)	113 (2.17)
Latvia	6 (16.22)	7 (18.92)	24 (64.86)	6 (16.22)	31 (83.78)	0 (0.00)
Lithuania	1 (4.17)	10 (41.67)	13 (54.17)	2 (8.33)	22 (91.67)	0 (0.00)
Luxembourg	0 (0.00)	16 (44.44)	20 (55.56)	0 (0.00)	33 (91.67)	3 (8.33)
Rep of Moldova	2 (3.57)	10 (17.86)	44 (78.57)	5 (8.93)	50 (89.29)	1 (1.79)
The Netherlands	44 (3.26)	682 (50.59)	622 (46.14)	228 (16.91)	1103 (81.82)	17 (1.26)
North Macedonia	6 (5.08)	75 (63.56)	37 (31.36)	6 (5.08)	85 (72.03)	27 (22.88)

[1] Ireland: chronicity for *Staphylococcus aureus* is defined as: at least 3 or more positive isolates during the last 12 months preceding the last reported culture in 2018.

[2] Italy: chronicity for *Staphylococcus aureus* is defined as: at least 3 or more positive cultures during 2018.

[table 5.4 continued]

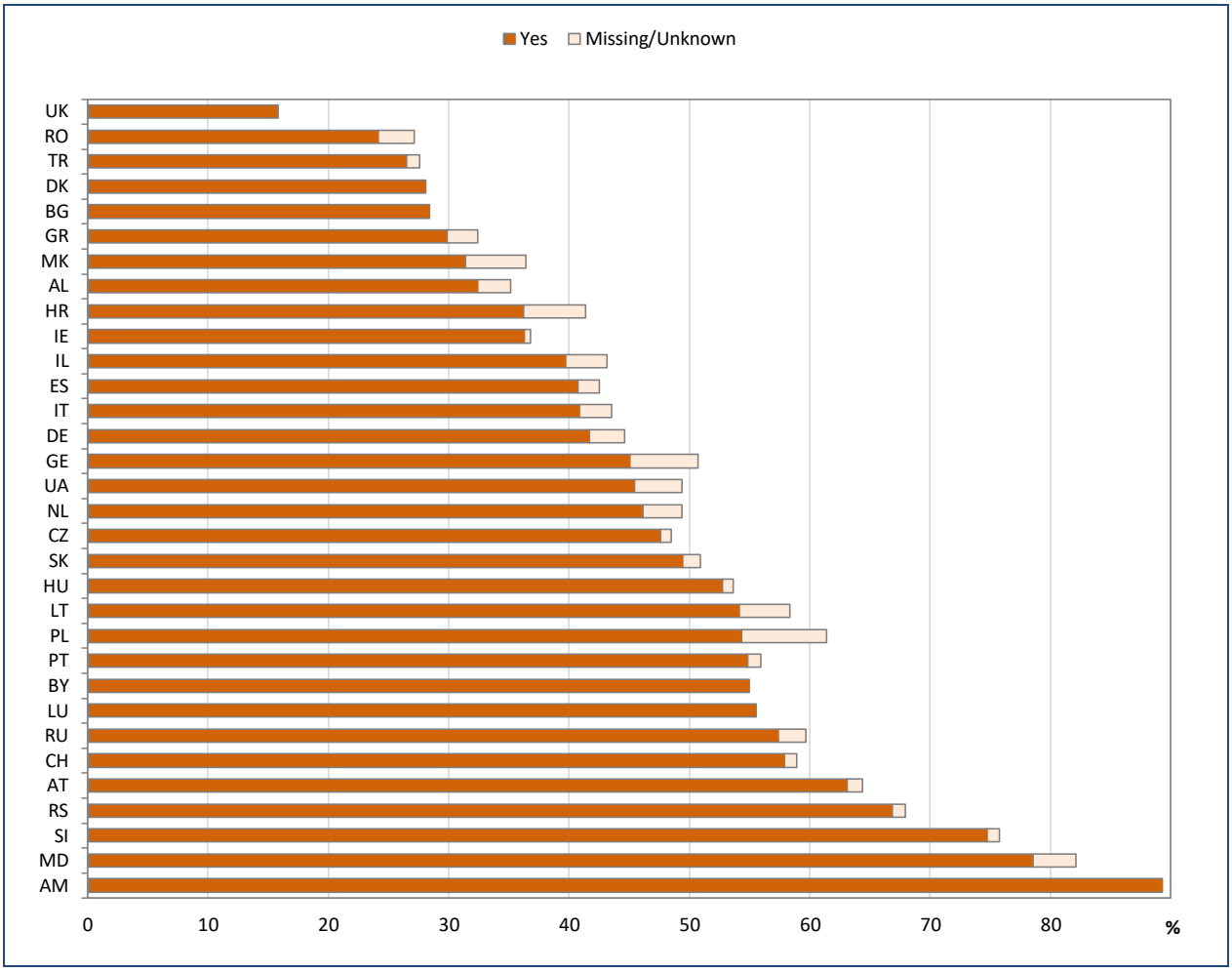
Country	Chronic *Staphylococcus aureus* number (%)			MRSA number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Norway	26 (10.00)	97 (37.31)	137 (52.69)	16 (6.15)	242 (93.08)	2 (0.77)
Poland	61 (7.07)	333 (38.59)	469 (54.35)	24 (2.78	807 (93.51	32 (3.71
Portugal	3 (1.08)	123 (44.09)	153 (54.84)	4 (1.43)	237 (84.95)	38 (13.62)
Romania	6 (3.02)	145 (72.86)	48 (24.12)	5 (2.51)	155 (77.89)	39 (19.60)
Russian Federation	70 (2.26)	1248 (40.32)	1777 (57.42)	102 (3.30)	2874 (92.86)	119 (3.84)
Serbia	2 (1.12)	57 (32.02)	119 (66.85)	1 (0.56)	143 (80.34)	34 (19.10)
Slovak Republic	4 (1.47)	134 (49.08)	135 (49.45)	8 (2.93)	248 (90.84)	17 (6.23)
Slovenia	1 (1.05)	23 (24.21)	71 (74.74)	3 (3.16)	89 (93.68)	3 (3.16)
Spain	36 (1.79)	1155 (57.46)	819 (40.75)	42 (2.09)	1844 (91.74)	124 (6.17)
Sweden	61 (10.03)	350 (57.57)	197 (32.40)	0 (0.00)	602 (99.01)	6 (0.99)
Switzerland	9 (1.02)	362 (41.09)	510 (57.89)	15 (1.70)	848 (96.25)	18 (2.04)
Turkey	19 (1.06)	1304 (72.44)	477 (26.50)	693 (38.50)	949 (52.72)	158 (8.78)
Ukraine	6 (3.90)	78 (50.65)	70 (45.45)	5 (3.25)	135 (87.66)	14 (9.09)
United Kingdom[3]	4 (0.04)	7991 (84.22)	1493 (15.74)	4 (0.04)	9208 (97.05)	276 (2.91)
Total	1867 (4.79)	23193 (59.53)	13900 (35.68)			

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

Note: For methicillin-resistant *Staphylococcus aureus* (MRSA) the total percentage of missing information is higher than 10%, therefore the totals are excluded from the table.

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

Figure 5.4 Prevalence of chronic Staphylococcus aureus infection in all patients seen in 2018 who have never had a transplant, by country.

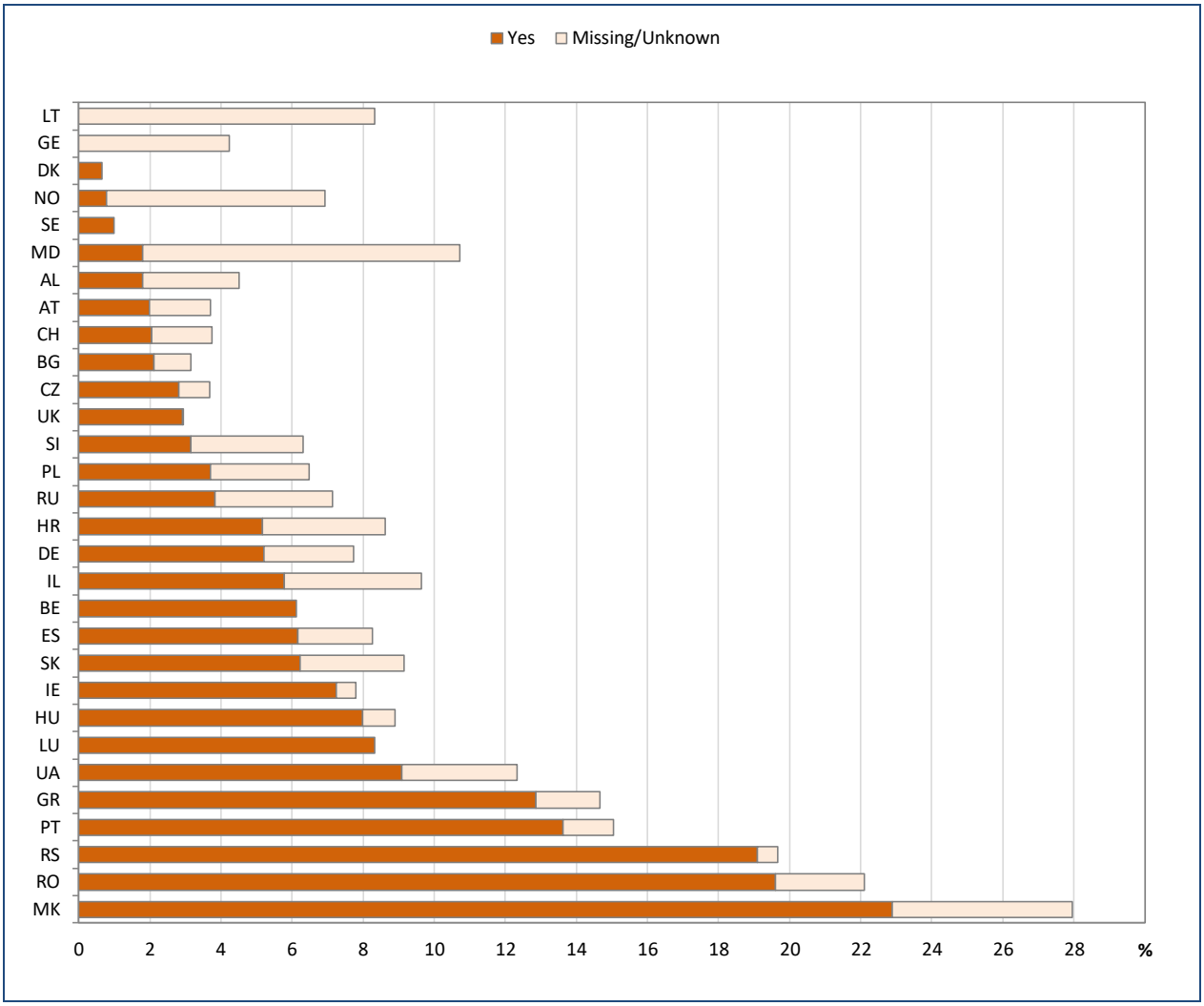


Note: We excluded from the graph the countries for which the information on *Staphylococcus aureus* was missing for more than 10% of the patients.

Note: Ireland: chronicity for *Staphylococcus aureus* is defined as: at least 3 or more positive isolates during the last 12 months preceding the last reported culture in 2018.
Italy: chronicity for *Staphylococcus aureus* is defined as: at least 3 or more positive cultures during 2018.
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.

Figure 5.5 Prevalence of methicillin-resistant Staphylococcus aureus (MRSA) infection in all patients seen in 2018 who have never had a transplant, by country.



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

The horizontal bars represent the percentage of patients with methicillin-resistant *Staphylococcus aureus* infection (in dark orange) and the percentage of patients where information on methicillin-resistant *Staphylococcus aureus* was missing (in light orange).

Table 5.5 Prevalence of chronic Staphylococcus aureus and methicillin-resistant Staphylococcus aureus (MRSA) in children seen in 2018 who have never had a transplant, by country.

Country	Chronic *Staphylococcus aureus* number (%)			MRSA number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	2 (1.89)	71 (66.98)	33 (31.13)	2 (1.89)	102 (96.23)	2 (1.89)
Armenia	0 (0.00)	3 (10.71)	25 (89.29)	22 (78.57)	4 (14.29)	2 (7.14)
Austria	1 (0.27)	140 (37.53)	232 (62.20)	3 (0.80)	363 (97.32)	7 (1.88)
Belarus	0 (0.00)	50 (46.30)	58 (53.70)	108 (100)	0 -	0 -
Belgium	465 (100)	0 -	0 -	0 (0.00)	440 (94.62)	25 (5.38)
Bulgaria	0 (0.00)	80 (71.43)	32 (28.57)	2 (1.79)	107 (95.54)	3 (2.68)
Croatia	5 (6.41)	44 (56.41)	29 (37.18)	3 (3.85)	71 (91.03)	4 (5.13)
Cyprus	7 (58.33)	2 (16.67)	3 (25.00)	1 (8.33)	11 (91.67)	0 (0.00)
Czech Republic	2 (0.63)	175 (55.03)	141 (44.34)	2 (0.63)	312 (98.11)	4 (1.26)
Denmark	0 (0.00)	147 (73.13)	54 (26.87)	0 (0.00)	200 99.50)	1 (0.50)
Georgia	4 (5.63)	35 (49.30)	32 (45.07)	3 (4.23)	68 (95.77)	0 (0.00)
Germany	39 (1.53)	1576 (61.85)	933 (36.62)	34 (1.33)	2418 (94.90)	96 (3.77)
Greece	7 (2.55)	202 (73.45)	66 (24.00)	3 (1.09)	235 (85.45)	37 (13.45)
Hungary	0 (0.00)	129 (50.79)	125 (49.21)	1 (0.39)	236 (92.91)	17 (6.69)
Ireland[1]	1 (0.20)	312 (61.54)	194 (38.26)	1 (0.20)	472 (93.10)	34 (6.71)
Israel	11 (5.34)	95 (46.12)	100 (48.54)	11 (5.34)	187 (90.78)	8 (3.88)
Italy[2]	87 (3.77)	1353 (58.67)	866 (37.55)	1886 (81.79)	373 (16.18)	47 (2.04)
Latvia	6 (21.43)	4 (14.29)	18 (64.29)	6 (21.43)	22 (78.57)	0 (0.00)
Lithuania	0 (0.00)	3 (33.33)	6 (66.67)	0 (0)	9 (100)	0 (0)
Luxembourg	0 (0.00)	9 (50.00)	9 (50.00)	0 (0)	18 (100)	0 (0)
Rep of Moldova	0 (0.00)	8 (18.18)	36 (81.82)	1 (2.27)	42 (95.45)	1 (2.27)
The Netherlands	5 (0.93)	295 (54.93)	237 (44.13)	5 (0.93)	523 (97.39)	9 (1.68)
North Macedonia	3 (3.61)	55 (66.27)	25 (30.12)	3 (3.61)	61 (73.49)	19 (22.89)

[1] Ireland: chronicity for *Staphylococcus aureus* is defined as: at least 3 or more positive isolates during the last 12 months preceding the last reported culture in 2018.

[2] Italy: chronicity for *Staphylococcus aureus* is defined as: at least 3 or more positive cultures during 2018.

[table 5.5 continued]

Country	Chronic *Staphylococcus aureus* number (%)			MRSA number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Norway	2 (1.92)	46 (44.23)	56 (53.85)	2 (1.92)	102 (98.08)	0 (0.00)
Poland	43 (6.68)	244 (37.89)	357 (55.43)	11 (1.71)	616 (95.65)	17 (2.64)
Portugal	1 (0.66)	72 (47.37)	79 (51.97)	1 (0.66)	132 (86.84)	19 (12.50)
Romania	6 (3.08)	143 (73.33)	46 (23.59)	5 (2.56)	153 (78.46)	37 (18.97)
Russian Federation	48 (2.04)	902 (38.30)	1405 (59.66)	64 (2.72)	2212 (93.93)	79 (3.35)
Serbia	2 (1.55)	36 (27.91)	91 (70.54)	1 (0.78)	102 (79.07)	26 (20.16)
Slovak Republic	1 (0.80)	54 (43.20)	70 (56.00)	4 (3.20)	116 (92.80)	5 (4.00)
Slovenia	0 (0.00)	14 (23.73)	45 (76.27)	1 (1.69)	57 (96.61)	1 (1.69)
Spain	12 (1.11)	671 (62.24)	395 (36.64)	14 (1.30)	1014 (94.06)	50 (4.64)
Sweden	29 (11.11)	182 (69.73)	50 (19.16)	0 (0.00)	257 (98.47)	4 (1.53)
Switzerland	3 (0.71)	184 (43.40)	237 (55.90)	5 (1.18)	412 (97.17)	7 (1.65)
Turkey	17 (1.05)	1194 (74.07)	401 (24.88)	614 (38.09)	853 (52.92)	145 (9.00)
Ukraine	4 (3.25)	67 (54.47)	52 (42.28)	3 (2.44)	110 (89.43)	10 (8.13)
United Kingdom[3]	1 (0.02)	3827 (90.86)	384 (9.12)	1 (0.02)	4109 (97.55)	102 (2.42)
Total	814 (4.04)	12424 (61.63)	6922 (34.34)			

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

Note: For methicillin-resistant *Staphylococcus aureus* (MRSA) the total percentage of missing information is higher than 10%, therefore the totals are excluded from the table.

Table 5.5 shows, separately by country, the frequency of chronic *Staphylococcus aureus* and methicillin-resistant *Staphylococcus aureus* (MRSA) in children. The number of missing values is also included.

Table 5.6 Prevalence of chronic Staphylococcus aureus, methicillin-resistant Staphylococcus aureus (MRSA) in adults seen in 2018 who have never had a transplant, by country.

Country	Chronic *Staphylococcus aureus* number (%)			MRSA number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	1 (20.00)	1 (20.00)	3 (60.00)	1 (20.00)	4 (80.00)	0 (0.00)
Austria	8 (2.43)	110 (33.43)	211 (64.13)	9 (2.74)	313 (95.14)	7 (2.13)
Belgium	648 (100)	0 -	0 -	0 (0.00)	605 (93.36)	43 (6.64)
Bulgaria	0 (0.00)	56 (71.79)	22 (28.21)	0 (0.00)	77 (98.72)	1 (1.28)
Croatia	1 (2.63)	24 (63.16)	13 (34.21)	1 (2.63)	35 (92.11)	2 (5.26)
Czech Republic	3 (1.20)	118 (47.01)	130 (51.79)	3 (1.20)	236 (94.02)	12 (4.78)
Denmark	0 (0.00)	186 (70.99)	76 (29.01)	0 (0.00)	260 (99.24)	2 (0.76)
Germany	131 (3.99)	1656 (50.38)	1500 (45.63)	113 (3.44)	2966 (90.23)	208 (6.33)
Greece	7 (2.53)	171 (61.73)	99 (35.74)	7 (2.53)	236 (85.20)	34 (12.27)
Hungary	4 (2.17)	74 (40.22)	106 (57.61)	3 (1.63)	163 (88.59)	18 (9.78)
Ireland[1]	5 (0.82)	394 (64.59)	211 (34.59)	5 (0.82)	558 (91.48)	47 (7.70)
Israel	7 (2.24)	200 (63.9)	106 (33.87)	9 (2.88)	282 (90.10)	22 (7.03)
Italy[2]	53 (1.83)	1588 (54.72)	1261 (43.45)	2231 (76.88)	605 (20.85)	66 (2.27)
Latvia	0 (0.00)	3 (33.33)	6 (66.67)	0 (0)	9 (100)	0 (0)
Lithuania	1 (6.67)	7 (46.67)	7 (46.67)	2 (13.33)	13 (86.67)	0 (0.00)
Luxembourg	0 (0.00)	7 (38.89)	11 (61.11)	0 (0.00)	15 (83.33)	3 (16.67)
Rep of Moldova	2 (16.67)	2 (16.67)	8 (66.67)	4 (33.33)	8 (66.67)	0 (0.00)
The Netherlands	39 (4.81)	387 (47.72)	385 (47.47)	223 (27.50)	580 (71.52)	8 (0.99)
North Macedonia	3 (8.57)	20 (57.14)	12 (34.29)	3 (8.57)	24 (68.57)	8 (22.86)
Norway	24 (15.38)	51 (32.69)	81 (51.92)	14 (8.97)	140 (89.74)	2 (1.28)
Poland	18 (8.22)	89 (40.64)	112 (51.14)	13 (5.94)	191 (87.21)	15 (6.85)

[1] Ireland: chronicity for *Staphylococcus aureus* is defined as: at least 3 or more positive isolates during the last 12 months preceding the last reported culture in 2018.

[2] Italy: chronicity for *Staphylococcus aureus* is defined as: at least 3 or more positive cultures during 2018.

[table 5.6 continued]

Country	Chronic *Staphylococcus aureus* number (%)			MRSA number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Portugal	2 (1.57)	51 (40.16)	74 (58.27)	3 (2.36)	105 (82.68)	19 (14.96)
Russian Federation	22 (2.97)	346 (46.76)	372 (50.27)	38 (5.14)	662 (89.46)	40 (5.41)
Serbia	0 (0.00)	21 (42.86)	28 (57.14)	0 (0.00)	41 (83.67)	8 (16.33)
Slovak Republic	3 (2.03)	80 (54.05)	65 (43.92)	4 (2.70)	132 (89.19)	12 (8.11)
Slovenia	1 (2.78	9 (25.00	26 (72.22)	2 (5.56)	32 (88.89)	2 (5.56)
Spain	24 (2.58)	484 (51.93)	424 (45.49)	28 (3.00)	830 (89.06)	74 (7.94)
Sweden	32 (9.22)	168 (48.41)	147 (42.36)	0 (0.00)	345 (99.42)	2 (0.58)
Switzerland	6 (1.31)	178 (38.95)	273 (59.74)	10 (2.19)	436 (95.40)	11 (2.41)
Turkey	2 (1.06)	110 (58.51)	76 (40.43)	79 (42.02)	96 (51.06)	13 (6.91)
Ukraine	2 (6.45)	11 (35.48)	18 (58.06)	2 (6.45)	25 (80.65)	4 (12.90)
United Kingdom[3]	3 (0.06)	4164 (78.92)	1109 (21.02)	3 (0.06)	5099 (96.65)	174 (3.30)
Total	1053 (5.60)	10769 (57.28)	6978 (37.12)			

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

Note: For methicillin-resistant *Staphylococcus aureus* (MRSA) the total percentage of missing information is higher than 10%, therefore the totals are excluded from the table.

Note: Belarus and Georgia have 0% coverage for adults and are excluded from the table.

Note: Armenia, Belarus, Cyprus and Romania have <5 patients aged 18 years or more at 31/12/2018 and are not shown in this table, but are considered in the total.

Table 5.5 shows, separately by country, the frequency of chronic *Staphylococcus aureus* and methicillin-resistant *Staphylococcus aureus* (MRSA) in adults. The number of missing values is also included.

Table 5.7 Prevalence of non-tuberculous mycobacteria, Stenotrophomonas maltophilia and Achromobacter species infection in all patients seen in 2018 who have never had a transplant, by country.

Country	Non-tuberculous mycobacteria (NTM) infection this year number (%)			*Stenotrophomonas maltophilia* infection this year number (%)			*Achromobacter species* infection this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	3 (2.70)	108 (97.30)	0 (0.00)	5 (4.50)	105 (94.59)	1 (0.90)	4 (3.60)	106 (95.50)	1 (0.90)
Armenia	22 (78.57)	6 (21.43)	0 (0.00)	25 (89.29)	3 (10.71)	0 (0.00)	25 (89.29)	3 (10.71)	0 (0.00)
Austria	88 (12.54)	582 (82.91)	32 (4.56)	10 (1.42)	600 (85.47)	92 (13.11)	10 (1.42)	651 (92.74)	41 (5.84)
Belarus	102 (91.89)	9 (8.11)	0 (0.00)	0 (0.00)	110 (99.10)	1 (0.90)	111 (100)	0 -	0 -
Belgium	0 (0.00)	1096 (98.47)	17 (1.53)	0 (0.00)	972 (87.33)	141 (12.67)	0 (0.00)	1003 (90.12)	110 (9.88)
Bulgaria	177 (93.16)	12 (6.32)	1 (0.53)	0 (0.00)	186 (97.89)	4 (2.11)	0 (0.00)	181 (95.26)	9 (4.74)
Croatia	32 (27.59)	83 (71.55)	1 (0.86)	4 (3.45)	106 (91.38)	6 (5.17)	4 (3.45)	110 (94.83)	2 (1.72)
Cyprus	2 (13.33)	13 (86.67)	0 (0.00)	2 (13.33)	12 (80.00)	1 (6.67)	2 (13.33)	11 (73.33)	2 (13.33)
Czech Republic	271 (47.63)	279 (49.03)	19 (3.34)	4 (0.70)	514 (90.33)	51 (8.96)	10 (1.76)	550 (96.66)	9 (1.58)
Denmark	0 (0.00)	443 (95.68)	20 (4.32)	0 (0.00)	347 (74.95)	116 (25.05)	0 (0.00)	417 (90.06)	46 (9.94)
Georgia	2 (2.82)	69 (97.18)	0 (0.00)	5 (7.04)	66 (92.96)	0 (0.00)	3 (4.23)	68 (95.77)	0 (0.00)
Germany[1]	3766 (64.54)	1896 (32.49)	173 (2.96)	146 (2.50)	5063 (86.77)	626 (10.73)	146 (2.50)	5382 (92.24)	307 (5.26)
Greece[2]	296 (53.62)	249 (45.11)	7 (1.27)	8 (1.45)	482 (87.32)	62 (11.23)	13 (2.36)	490 (88.77)	49 (8.88)
Hungary	14 (3.20)	415 (94.75)	9 (2.05)	3 (0.68)	414 (94.52)	21 (4.79)	438 (100)	0 -	0 -
Ireland	6 (0.54)	1075 (96.24)	36 (3.22)	6 (0.54)	1010 (90.42)	101 (9.04)	6 (0.54)	1076 (96.33)	35 (3.13)
Israel	26 (5.01)	456 (87.86)	37 (7.13)	24 (4.62)	457 (88.05)	38 (7.32)	21 (4.05)	477 (91.91)	21 (4.05)
Italy	468 (8.99)	4725 (90.73)	15 (0.29)	468 (8.99)	4594 (88.21)	146 (2.80)	4134 (79.38)	971 (18.64)	103 (1.98)
Latvia	8 (21.62)	29 (78.38)	0 (0.00)	6 (16.22)	29 (78.38)	2 (5.41)	6 (16.22)	29 (78.38)	2 (5.41)
Lithuania	1 (4.17)	23 (95.83)	0 (0.00)	1 (4.17)	21 (87.50)	2 (8.33)	2 (8.33)	21 (87.50)	1 (4.17)
Luxembourg	0 (0.00)	34 (94.44)	2 (5.56)	0 (0.00)	32 (88.89)	4 (11.11)	0 (0)	36 (100)	0 (0)
Rep of Moldova	48 (85.71)	8 (14.29)	0 (0.00)	0 (0)	56 (100)	0 (0)	0 (0)	56 (100)	0 (0)
The Netherlands	95 (7.05)	1212 (89.91)	41 (3.04)	228 (16.91)	982 (72.85)	138 (10.24)	228 (16.91)	1073 (79.60)	47 (3.49)
North Macedonia	6 (5.08)	111 (94.07)	1 (0.85)	6 (5.08)	111 (94.07)	1 (0.85)	6 (5.08)	112 (94.92)	0 (0.00)

[1] Germany: If patients could not produce sputum or were not investigated for NTM, “missing/unknown” was used.
[2] Greece: one centre reported “missing” when children could not provide sputum for cultures for NTM.

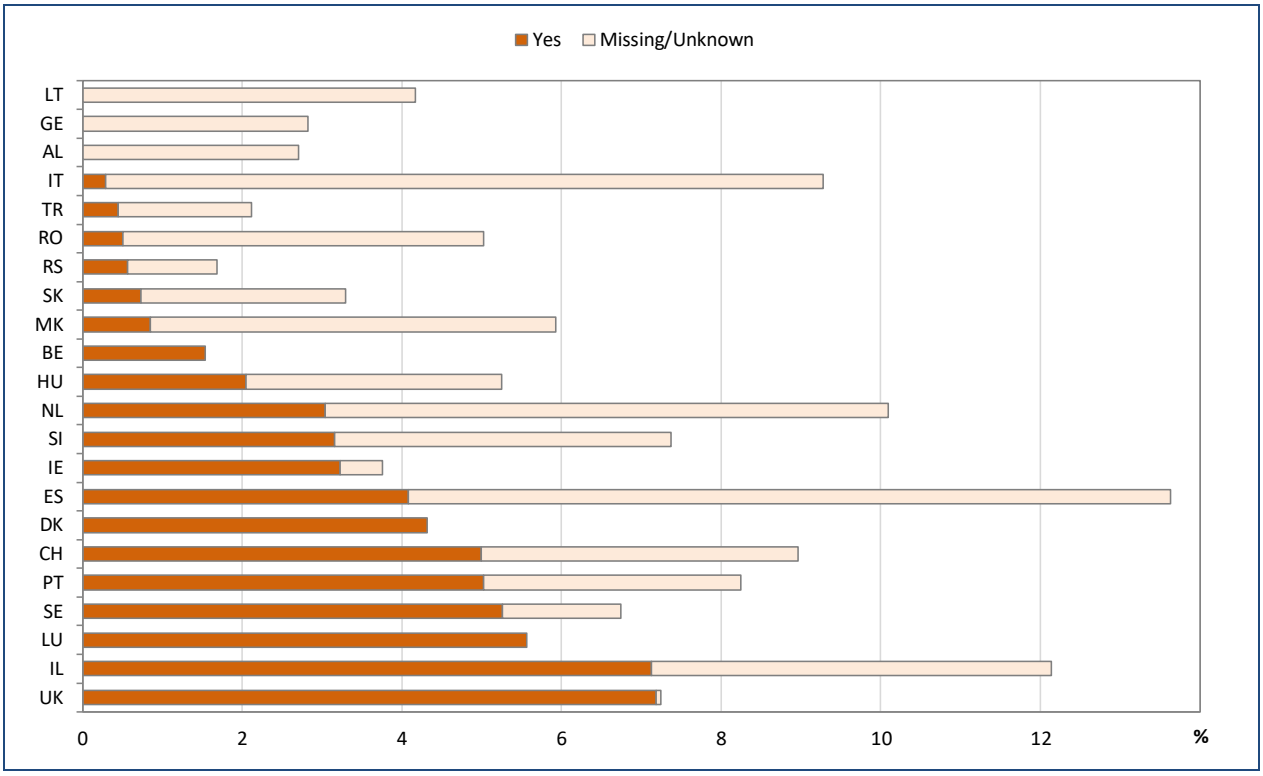
[table 5.7 continued]

Country	Non-tuberculous mycobacteria (NTM) infection this year number (%)			*Stenotrophomonas maltophilia* infection this year number (%)			*Achromobacter species* infection this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Norway	54 (20.77)	190 (73.08)	16 (6.15)	13 (5.00)	201 (77.31)	46 (17.69)	15 (5.77)	233 (89.62)	12 (4.62)
Poland	286 (33.14)	572 (66.28)	5 (0.58)	31 (3.59)	800 (92.70)	32 (3.71)	29 (3.36)	792 (91.77)	42 (4.87)
Portugal	9 (3.23)	256 (91.76)	14 (5.02)	5 (1.79)	247 (88.53)	27 (9.68)	4 (1.43)	257 (92.11)	18 (6.45)
Romania	9 (4.52)	189 (94.97)	1 (0.50)	6 (3.02)	190 (95.48)	3 (1.51)	4 (2.01)	193 (96.98)	2 (1.01)
Russian Federation	460 (14.86)	2611 (84.36)	24 (0.78)	89 (2.88)	2877 (92.96)	129 (4.17)	78 (2.52)	2844 (91.89)	173 (5.59)
Serbia	2 (1.12)	175 (98.31)	1 (0.56)	1 (0.56)	160 (89.89)	17 (9.55)	2 (1.12)	167 (93.82)	9 (5.06)
Slovak Republic	7 (2.56)	264 (96.70)	2 (0.73)	4 (1.47)	253 (92.67)	16 (5.86)	8 (2.93)	254 (93.04)	11 (4.03)
Slovenia	4 (4.21)	88 (92.63)	3 (3.16)	7 (7.37)	75 (78.95)	13 (13.68)	2 (2.11)	91 (95.79)	2 (2.11)
Spain	192 (9.55)	1737 (86.42)	81 (4.03)	32 (1.59)	1819 (90.50)	159 (7.91)	42 (2.09)	1825 (90.80)	143 (7.11)
Sweden	9 (1.48)	567 (93.26)	32 (5.26)	0 (0.00)	543 (89.31)	65 (10.69)	0 (0.00)	593 (97.53)	15 (2.47)
Switzerland	35 (3.97)	802 (91.03)	44 (4.99)	13 (1.48)	753 (85.47)	115 (13.05)	15 (1.70)	826 (93.76)	40 (4.54)
Turkey	30 (1.67)	1762 (97.89)	8 (0.44)	20 (1.11)	1702 (94.56)	78 (4.33)	691 (38.39)	1067 (59.28)	42 (2.33)
Ukraine	144 (93.51)	10 (6.49)	0 (0.00)	5 (3.25)	139 (90.26)	10 (6.49)	5 (3.25)	148 (96.10)	1 (0.65)
United Kingdom	6 (0.06)	8801 (92.76)	681 (7.18)	4 (0.04)	8756 (92.28)	728 (7.67)	4 (0.04)	9140 (96.33)	344 (3.63)
Total	6680	30957	1323	1181 (3.03)	34787 (89.29)	2992 (7.68)			

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

Table 5.7 shows the frequency of three other infections, *non-tuberculous mycobacteria* (NTM), *Stenotrophomonas maltophilia* and *Achromobacter species.* All 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 the culture techniques employed. For example, the detection of NTM is dependent on sputum production, which is not always possible for all patients, especially younger patients.

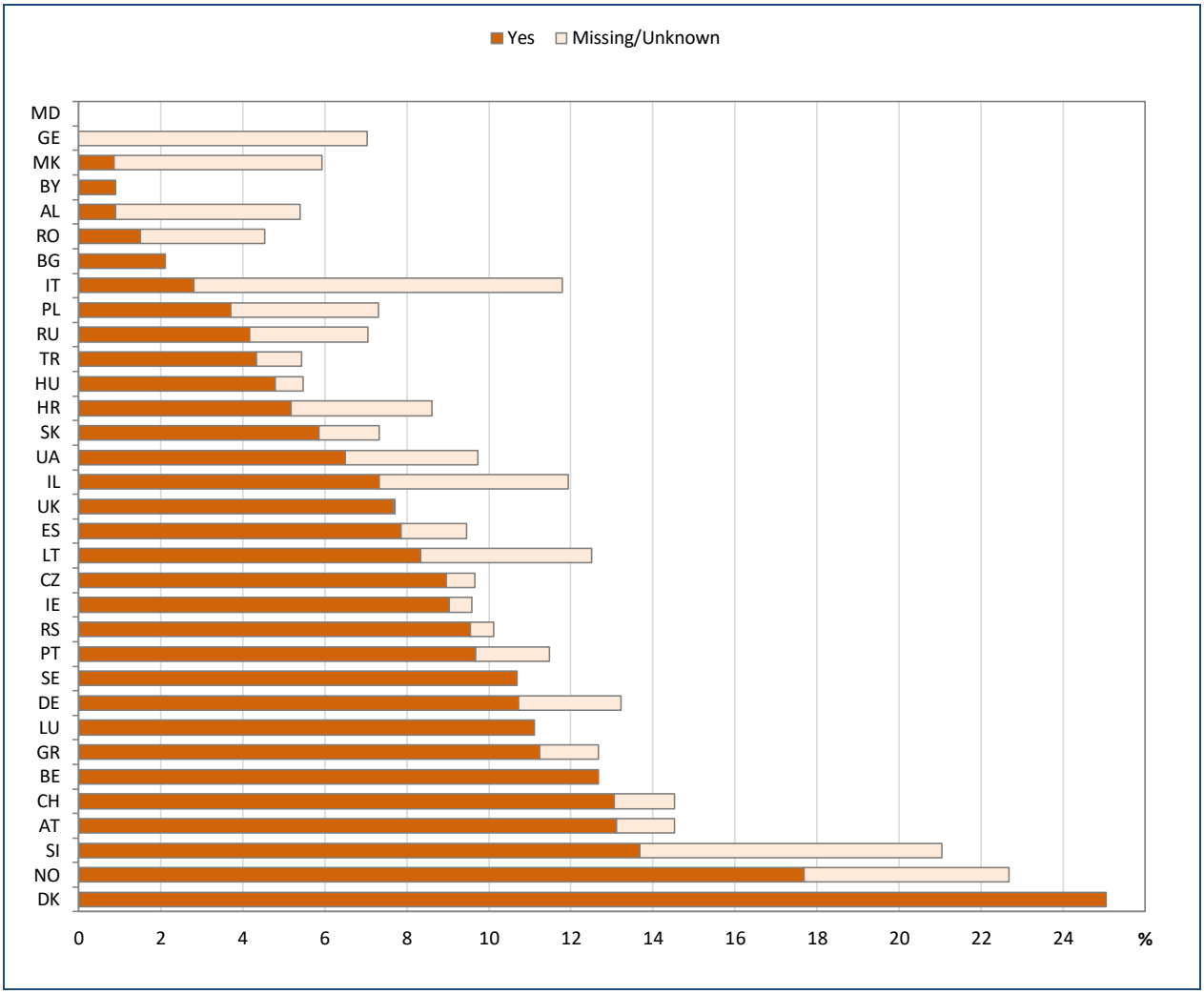
Figure 5.6 Prevalence of non-tuberculous mycobacteria in all patients seen in 2018 who have never had a transplant, by country.



Note: We excluded from the graph the countries for which the information on non-tuberculous mycobacteria was missing for more than 10% of the 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). Detection of non-tuberculous mycobacteria infection depends on sputum production, which is not always possible for all patients, especially younger patients. Generally, infections from these bacteria are not very frequent in any country.

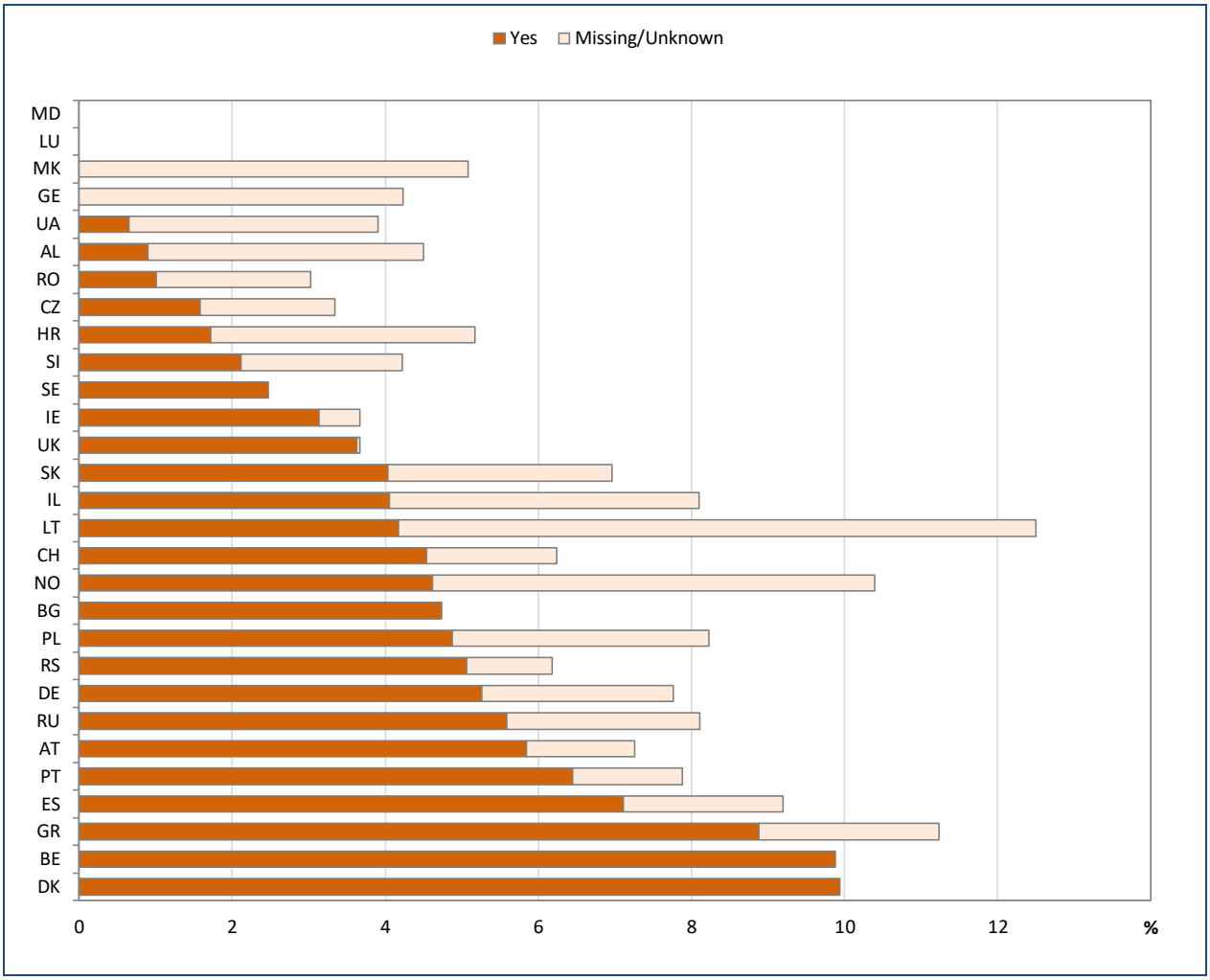
Figure 5.7 Prevalence of Stenotrophomonas maltophilia infection in all patients seen in 2018 who have never had a transplant, by country.



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

Figure 5.8 Prevalence of Achromobacter species in all patients seen in 2018 who have never had a transplant, by country.



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

The horizontal bars represent the percentage of patients with *Achromobacter species* infection (in dark orange) and the percentage of patients where information on *Achromobacter species* infection was missing (in light orange).

Table 5.8 Prevalence of non-tuberculous mycobacteria, Stenotrophomonas maltophilia and Achromobacter species infection in children seen in 2018 who have never had a transplant, by country.

Country	Non-tuberculous mycobacteria (NTM) infection this year number (%)			*Stenotrophomonas maltophilia* infection this year number (%)			*Achromobacter species* infection this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	2 (1.89)	104 (98.11)	0 (0.00)	4 (3.77)	101 (95.28)	1 (0.94)	3 (2.83)	102 (96.23)	1 (0.94)
Armenia	22 (78.57)	6 (21.43)	0 (0.00)	25 (89.29)	3 (10.71)	0 (0.00)	25 (89.29)	3 (10.71)	0 (0.00)
Austria	9 (2.41)	359 (96.25)	5 (1.34)	0 (0.00)	342 (91.69)	31 (8.31)	1 (0.27)	359 (96.25)	13 (3.49)
Belarus	100 (92.59)	8 (7.41)	0 (0.00)	0 (0.00)	107 (99.07)	1 (0.93)	108 (100)	0 -	0 -
Belgium	0 (0.00)	461 (99.14)	4 (0.86)	0 (0.00)	394 (84.73)	71 (15.27)	0 (0.00)	440 (94.62)	25 (5.38)
Bulgaria	103 (91.96)	9 (8.04)	0 (0.00)	0 (0.00)	110 (98.21)	2 (1.79)	0 (0.00)	105 (93.75)	7 (6.25)
Croatia	27 (34.62)	51 (65.38)	0 (0.00)	3 (3.85)	72 (92.31)	3 (3.85)	3 (3.85)	74 (94.87)	1 (1.28)
Cyprus	1 (8.33)	11 (91.67)	0 (0.00)	1 (8.33)	10 (83.33)	1 (8.33)	1 (8.33)	10 (83.33)	1 (8.33)
Czech Republic	251 (78.93)	62 (19.50)	5 (1.57)	1 (0.31)	289 (90.88)	28 (8.81)	2 (0.63)	310 (97.48)	6 (1.89)
Denmark	0 (0.00)	198 (98.51)	3 (1.49)	0 (0.00)	156 (77.61)	45 (22.39)	0 (0.00)	189 (94.03)	12 (5.97)
Georgia	2 (2.82)	69 (97.18)	0 (0.00)	5 (7.04)	66 (92.96)	0 (0.00)	3 (4.23)	68 (95.77)	0 (0.00)
Germany	2020 (79.28)	501 (19.66)	27 (1.06)	34 (1.33)	2269 (89.05)	245 (9.62)	34 (1.33)	2441 (95.80)	73 (2.86)
Greece	160 (58.18)	115 (41.82)	0 (0.00)	1 (0.36)	243 (88.36)	31 (11.27)	6 (2.18)	250 (90.91)	19 (6.91)
Hungary	8 (3.15)	243 (95.67)	3 (1.18)	0 (0.00)	246 (96.85)	8 (3.15)	254 (100)	0 -	0 -
Ireland	1 (0.20)	493 (97.24)	13 (2.56)	1 (0.20)	463 (91.32)	43 (8.48)	1 (0.20)	492 (97.04)	14 (2.76)
Israel	13 (6.31)	181 (87.86)	12 (5.83)	14 (6.80)	173 (83.98)	19 (9.22)	13 (6.31)	189 (91.75)	4 (1.94)
Italy	227 (9.84)	2074 (89.94)	5 (0.22)	227 (9.84)	2021 (87.64)	58 (2.52)	1896 (82.22)	380 (16.48)	30 (1.30)
Latvia	7 (25.00)	21 (75.00)	0 (0.00)	6 (21.43)	20 (71.43)	2 (7.14)	6 (21.43)	22 (78.57)	0 (0.00)
Lithuania	0 (0)	9 (100)	0 (0)	0 (0.00)	7 (77.78)	2 (22.22)	0 (0.00)	8 (88.89)	1 (11.11)
Luxembourg	0 (0)	18 (100)	0 (0)	0 (0.00)	15 (83.33)	3 (16.67)	0 (0)	18 (100))	0 (0
Rep of Moldova	36 (81.82)	8 (18.18)	0 (0.00)	0 (0)	44 (100)	0 (0)	0 (0)	44 (100)	0 (0)
The Netherlands	6 (1.12)	518 (96.46)	13 (2.42)	5 (0.93)	469 (87.34)	63 (11.73)	5 (0.93)	523 (97.39)	9 (1.68)
North Macedonia	3 (3.61)	80 (96.39)	0 (0.00)	3 (3.61)	79 (95.18)	1 (1.20)	3 (3.61)	80 (96.39)	0 (0.00)

[table 5.8 continued]

Country	Non-tuberculous mycobacteria (NTM) infection this year number (%)			*Stenotrophomonas maltophilia* infection this year number (%)			*Achromobacter species* infection this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Norway	17 (16.35)	84 (80.77)	3 (2.88)	1 (0.96)	91 (87.5)	12 (11.54)	1 (0.96)	101 (97.12)	2 (1.92)
Poland	236 (36.65)	404 (62.73)	4 (0.62)	18 (2.80)	603 (93.63)	23 (3.57)	13 (2.02)	618 (95.96)	13 (2.02)
Portugal	6 (3.95)	145 (95.39)	1 (0.66)	2 (1.32)	129 (84.87)	21 (13.82)	1 (0.66)	144 (94.74)	7 (4.61)
Romania	9 (4.62)	185 (94.87)	1 (0.51)	6 (3.08)	186 (95.38)	3 (1.54)	4 (2.05)	190 (97.44)	1 (0.51)
Russian Federation	374 (15.88)	1973 (83.78)	8 (0.34)	55 (2.34)	2201 (93.46)	99 (4.20)	51 (2.17)	2196 (93.25)	108 (4.59)
Serbia	2 (1.55)	127 (98.45)	0 (0.00)	1 (0.78)	115 (89.15)	13 (10.08)	2 (1.55)	122 (94.57)	5 (3.88)
Slovak Republic	3 (2.40)	121 (96.80)	1 (0.80)	1 (0.80)	118 (94.40)	6 (4.80)	5 (4.00)	117 (93.60)	3 (2.40)
Slovenia	0 (0.00)	57 (96.61)	2 (3.39)	0 (0.00)	53 (89.83)	6 (10.17)	0 (0.00)	58 (98.31)	1 (1.69)
Spain	155 (14.38)	898 (83.30)	25 (2.32)	10 (0.93)	988 (91.65)	80 (7.42)	16 (1.48)	1010 (93.69)	52 (4.82)
Sweden	7 (2.68)	246 (94.25)	8 (3.07)	0 (0.00)	235 (90.04)	26 (9.96)	0 (0.00)	257 (98.47)	4 (1.53)
Switzerland	20 (4.72)	397 (93.63)	7 (1.65)	5 (1.18)	382 (90.09)	37 (8.73)	3 (0.71)	410 (96.70)	11 (2.59)
Turkey	28 (1.74)	1579 (97.95)	5 (0.31)	18 (1.12)	1526 (94.67)	68 (4.22)	612 (37.97)	962 (59.68)	38 (2.36)
Ukraine	117 (95.12)	6 (4.88)	0 (0.00)	3 (2.44)	110 (89.43)	10 (8.13)	3 (2.44)	119 (96.75)	1 (0.81)
United Kingdom	3 (0.07)	4048 (96.11)	161 (3.82)	1 (0.02)	3932 (93.35)	279 (6.62)	1 (0.02)	4130 (98.05)	81 (1.92)
Total				451 (2.24)	18368 (91.11)	1341 (6.65)			

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

Table 5.8 shows the frequency of three other infections, non-tuberculous mycobacteria (NTM), *Stenotrophomonas maltophilia* and *Achromobacter species* in children.

Table 5.9 Prevalence of non-tuberculous mycobacteria, Stenotrophomonas maltophilia and Achromobacter species infection in adults seen in 2018 who have never had a transplant, by country.

Country	Non-tuberculous mycobacteria (NTM) infection this year number (%)			*Stenotrophomonas maltophilia* infection this year number (%)			*Achromobacter species* infection this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	1 (20.00)	4 (80.00)	0 (0.00)	1 (20.00)	4 (80.00)	0 (0.00)	1 (20.00)	4 (80.00)	0 (0.00)
Austria	79 (24.01)	223 (67.78)	27 (8.21)	10 (3.04)	258 (78.42)	61 (18.54)	9 (2.74)	292 (88.75)	28 (8.51)
Belgium	0 (0.00)	635 (97.99)	13 (2.01)	0 (0.00)	578 (89.20)	70 (10.80)	0 (0.00)	563 (86.88)	85 (13.12)
Bulgaria	74 (94.87)	3 (3.85)	1 (1.28)	0 (0.00)	76 (97.44)	2 (2.56)	0 (0.00)	76 (97.44)	2 (2.56)
Croatia	5 (13.16)	32 (84.21)	1 (2.63)	1 (2.63)	34 (89.47)	3 (7.89)	1 (2.63)	36 (94.74)	1 (2.63)
Czech Republic	20 (7.97)	217 (86.45)	14 (5.58)	3 (1.20)	225 (89.64)	23 (9.16)	8 (3.19)	240 (95.62)	3 (1.20)
Denmark	0 (0.00)	245 (93.51)	17 (6.49)	0 (0.00)	191 (72.90)	71 (27.10)	0 (0.00)	228 (87.02)	34 (12.98)
Germany	1746 (53.12)	1395 (42.44)	146 (4.44)	112 (3.41)	2794 (85.00)	381 (11.59)	112 (3.41)	2941 (89.47)	234 (7.12)
Greece	136 (49.10)	134 (48.38)	7 (2.53)	7 (2.53)	239 (86.28)	31 (11.19)	7 (2.53)	240 (86.64)	30 (10.83)
Hungary	6 (3.26)	172 (93.48)	6 (3.26)	3 (1.63)	168 (91.30)	13 (7.07)	184 (100)	0 -	0 -
Ireland	5 (0.82)	582 (95.41)	23 (3.77)	5 (0.82)	547 (89.67)	58 (9.51)	5 (0.82)	584 (95.74)	21 (3.44)
Israel	13 (4.15)	275 (87.86)	25 (7.99)	10 (3.19)	284 (90.73)	19 (6.07)	8 (2.56)	288 (92.01)	17 (5.43)
Italy	241 (8.30)	2651 (91.35)	10 (0.34)	241 (8.30)	2573 (88.66)	88 (3.03)	2238 (77.12)	591 (20.37)	73 (2.52)
Latvia	1 (11.11)	8 (88.89)	0 (0.00)	0 (0)	9 (100)	0 (0)	0 (0.00)	7 (77.78)	2 (22.22)
Lithuania	1 (6.67)	14 (93.33)	0 (0.00)	1 (6.67)	14 (93.33)	0 (0.00)	2 (13.33)	13 (86.67)	0 (0.00)
Luxembourg	0 (0.00)	16 (88.89)	2 (11.11)	0 (0.00)	17 (94.44)	1 (5.56)	0 (0)	18 (100)	0 (0)
Rep of Moldova	12 (100)	0 -	0 -	0 (0)	12 (100)	0 (0)	0 (0)	12 (100)	0 (0)
The Netherlands	89 (10.97)	694 (85.57)	28 (3.45)	223 (27.50)	513 (63.26)	75 (9.25)	223 (27.50)	550 (67.82)	38 (4.69)
North Macedonia	3 (8.57)	31 (88.57)	1 (2.86)	3 (8.57)	32 (91.43)	0 (0.00)	3 (8.57)	32 (91.43)	0 (0.00)

[table 5.9 continued]

Country	Non-tuberculous mycobacteria (NTM) infection this year number (%)			*Stenotrophomonas maltophilia* infection this year number (%)			*Achromobacter species* infection this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Norway	37 (23.72)	106 (67.95)	13 (8.33)	12 (7.69)	110 (70.51)	34 (21.79)	14 (8.97)	132 (84.62)	10 (6.41)
Poland	50 (22.83)	168 (76.71)	1 (0.46)	13 (5.94)	197 (89.95)	9 (4.11)	16 (7.31)	174 (79.45)	29 (13.24)
Portugal	3 (2.36)	111 (87.40)	13 (10.24)	3 (2.36)	118 (92.91)	6 (4.72)	3 (2.36)	113 (88.98)	11 (8.66)
Russian Federation	86 (11.62)	638 (86.22)	16 (2.16)	34 (4.59)	676 (91.35)	30 (4.05)	27 (3.65)	648 (87.57)	65 (8.78)
Serbia	0 (0.00)	48 (97.96)	1 (2.04)	0 (0.00)	45 (91.84)	4 (8.16)	0 (0.00)	45 (91.84)	4 (8.16)
Slovak Republic	4 (2.70)	143 (96.62)	1 (0.68)	3 (2.03)	135 (91.22)	10 (6.76)	3 (2.03)	137 (92.57)	8 (5.41)
Slovenia	4 (11.11)	31 (86.11)	1 (2.78)	7 (19.44)	22 (61.11)	7 (19.44)	2 (5.56)	33 (91.67)	1 (2.78)
Spain	37 (3.97)	839 (90.02)	56 (6.01)	22 (2.36)	831 (89.16)	79 (8.48)	26 (2.79)	815 (87.45)	91 (9.76)
Sweden	2 (0.58)	321 (92.51)	24 (6.92)	0 (0.00)	308 (88.76)	39 (11.24)	0 (0.00)	336 (96.83)	11 (3.17)
Switzerland	15 (3.28)	405 (88.62)	37 (8.10)	8 (1.75)	371 (81.18)	78 (17.07)	12 (2.63)	416 (91.03)	29 (6.35)
Turkey	2 (1.06)	183 (97.34)	3 (1.60)	2 (1.06)	176 (93.62)	10 (5.32)	79 (42.02)	105 (55.85)	4 (2.13)
Ukraine	27 (87.10)	4 (12.90)	0 (0.00)	2 (6.45)	29 (93.55)	0 (0.00)	2 (6.45)	29 (93.55)	0 (0.00)
United Kingdom	3 (0.06)	4753 (90.09)	520 (9.86)	3 (0.06)	4824 (91.43)	449 (8.51)	3 (0.06)	5010 (94.96)	263 (4.98)
Total				730 (3.88)	16419 (87.34)	1651 (8.78)			

Note: For non-tuberculous mycobacteria (NTM) and *Achromobacter species* the total percentage of missing information is higher than 10%, therefore the totals are excluded from the table.
Note: Belarus and Georgia have 0% coverage for adults and are excluded from the table.
Note: Armenia, Belarus, Cyprus and Romania have <5 patients aged 18 years or more at 31/12/2018 and are not shown in this table, but considered in the total.

Table 5.9 shows the frequency of three other infections, non-tuberculous mycobacteria (NTM), *Stenotrophomonas maltophilia* and *Achromobacter species* in adults.

6. Nutrition

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

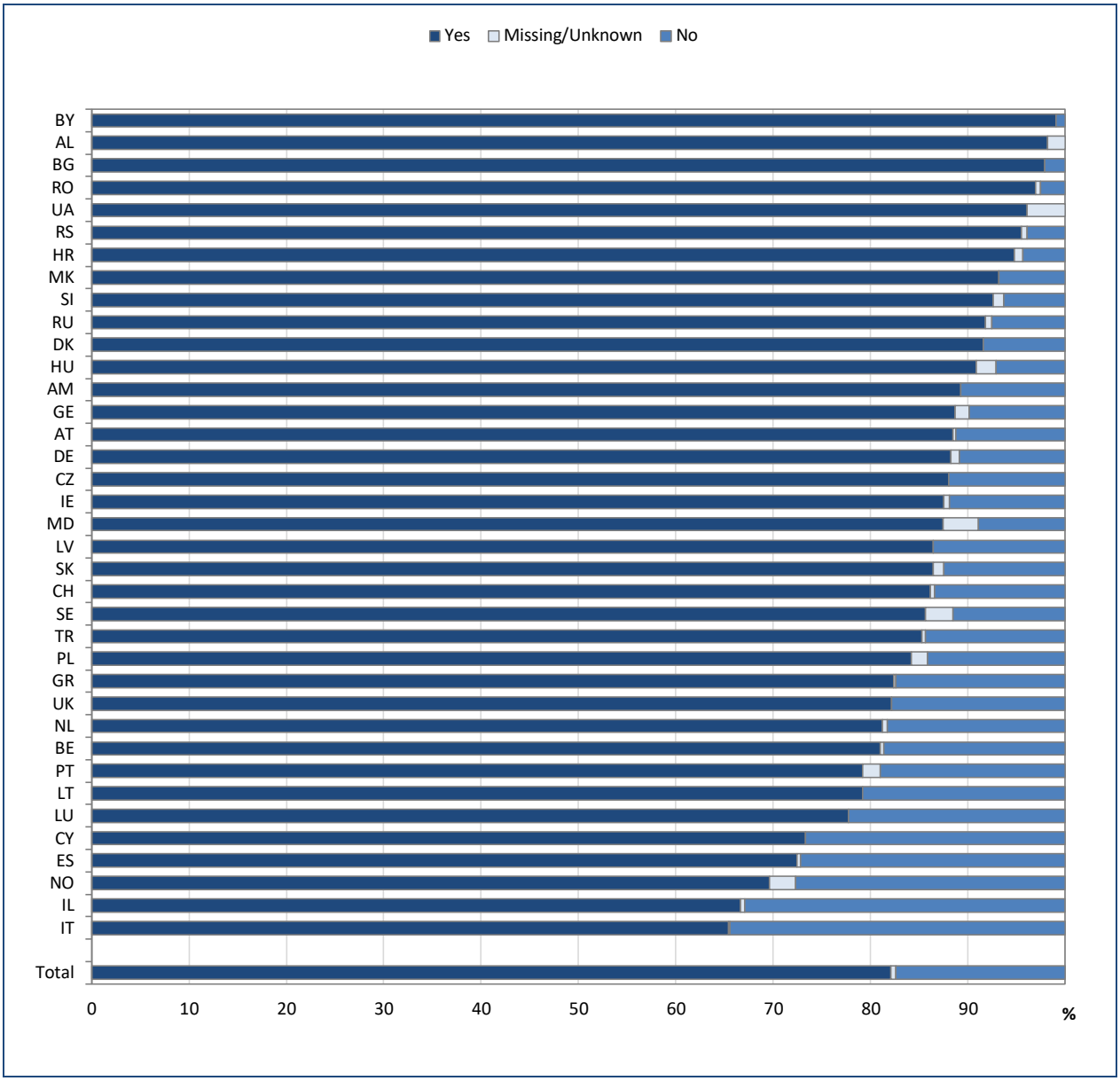
We collected weight and height measured on the date the best FEV1 value (of the highest FEV1% predicted of the year) was recorded. For patients that did not perform spirometry, the last height and weight measurements of the year were considered. From these raw values we calculated body mass index (BMI). A patient with a low weight is not necessarily underweight if the height is also low, therefore BMI may better illustrate the nutritional status because it describes the weight/height relationship. The ECFS Standards of Care guidelines recommend: for adults, a BMI of above 20 kg/m^2; for older children and adolescents, the 50th percentile for BMI; for infants and children up to 2 years of age, weight and height percentiles similar to those for the non-CF population.[1]

Weight, height and BMI were then expressed in terms of z-scores by using a reference population of healthy individuals (in this case the US population with reference values issued by the Centre for Disease Control, USA, see Appendix 1, page 170, for details).

A z-score of 0 means that the height/weight/BMI is equal to the mean height/weight/BMI of people of the same age and sex of the reference population. A z-score of -2 means that the height/weight/BMI value is 2 standard deviations below the mean height/weight/BMI of people of the same age and sex of the reference population; a z-score of +2 means that the value is 2 standard deviations above that mean. In the reference population, 95% of all individuals have a z-score for weight between -2 and +2 (the same for height) and it is expected that the same happens for approximately 95% of individuals of a population without conditions that affect weight (or height). The average z-score for a largely healthy population should be very close to zero.

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

Figure 6.1 Use of pancreatic enzymes in 2018 for all patients who have never had a transplant, by country and overall.



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

Table 6.1 Number of patients for whom height and weight measurements were available. All patients seen in 2018 who never had a transplant.

Country	Number of patients	Height				Weight			
		N	(%)	N miss	(%)	N	(%)	N miss	(%)
Albania	111	96	(86.49)	15	(13.51)	106	(95.50)	5	(4.50)
Armenia	28	28	(100)	0	(0.00)	28	(100)	0	(0.00)
Austria	702	687	(97.86)	15	(2.14)	688	(98.01)	14	(1.99)
Belarus	111	110	(99.10)	1	(0.90)	110	(99.10)	1	(0.90)
Belgium	1113	1107	(99.46)	6	(0.54)	1107	(99.46)	6	(0.54)
Bulgaria	190	183	(96.32)	7	(3.68)	185	(97.37)	5	(2.63)
Croatia	116	112	(96.55)	4	(3.45)	115	(99.14)	1	(0.86)
Cyprus	15	13	(86.67)	2	(13.33)	14	(93.33)	1	(6.67)
Czech Republic	569	565	(99.30)	4	(0.70)	565	(99.30)	4	(0.70)
Denmark	463	459	(99.14)	4	(0.86)	459	(99.14)	4	(0.86)
Georgia	71	65	(91.55)	6	(8.45)	65	(91.55)	6	(8.45)
Germany	5835	5794	(99.30)	41	(0.70)	5794	(99.30)	41	(0.70)
Greece	552	534	(96.74)	18	(3.26)	537	(97.28)	15	(2.72)
Hungary	438	365	(83.33)	73	(16.67)	366	(83.56)	72	(16.44)
Ireland	1117	1088	(97.40)	29	(2.60)	997	(89.26)	120	(10.74)
Israel	519	506	(97.50)	13	(2.50)	505	(97.30)	14	(2.70)
Italy	5208	4980	(95.62)	228	(4.38)	4996	(95.93)	212	(4.07)
Latvia	37	37	(100)	0	(0.00)	37	(100)	0	(0.00)
Lithuania	24	24	(100)	0	(0.00)	24	(100)	0	(0.00)
Luxembourg	36	36	(100)	0	(0.00)	36	(100)	0	(0.00)
Rep of Moldova	56	52	(92.86)	4	(7.14)	52	(92.86)	4	(7.14)
The Netherlands	1348	1338	(99.26)	10	(0.74)	1337	(99.18)	11	(0.82)
North Macedonia	118	115	(97.46)	3	(2.54)	116	(98.31)	2	(1.69)
Norway	260	257	(98.85)	3	(1.15)	255	(98.08)	5	(1.92)
Poland	863	836	(96.87)	27	(3.13)	843	(97.68)	20	(2.32)
Portugal	279	267	(95.70)	12	(4.30)	269	(96.42)	10	(3.58)
Romania	199	184	(92.46)	15	(7.54)	187	(93.97)	12	(6.03)
Russian Federation	3095	3001	(96.96)	94	(3.04)	3008	(97.19)	87	(2.81)
Serbia	178	170	(95.51)	8	(4.49)	171	(96.07)	7	(3.93)
Slovak Republic	273	247	(90.48)	26	(9.52)	251	(91.94)	22	(8.06)
Slovenia	95	94	(98.95)	1	(1.05)	94	(98.95)	1	(1.05)
Spain	2010	1933	(96.17)	77	(3.83)	1937	(96.37)	73	(3.63)
Sweden[1]	608	606	(99.67)	2	(0.33)	604	(99.34)	4	(0.66)
Switzerland	881	862	(97.84)	19	(2.16)	866	(98.30)	15	(1.70)
Turkey	1800	1786	(99.22)	14	(0.78)	1787	(99.28)	13	(0.72)
Ukraine	154	148	(96.10)	6	(3.90)	148	(96.10)	6	(3.90)
United Kingdom[2]	9488	8310	(87.58)	1178	(12.42)	8264	(87.10)	1224	(12.90)

[1] Sweden: if there is no lung function measurement, the height and weight reported are those measured at annual review.

[2] UK: height and weight at date of annual data is used instead of the date of best FEV1. If no lung function measurement is reported, the date of the last visit is used.

Table 6.2 Z-scores for height: descriptive statistics by country. Patients aged 17 years or younger who have never had a transplant.

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	94	-0.6	-3.9	-1.5	-0.6	0.2	1.9
Armenia	28	-0.6	-3.0	-1.3	-0.5	-0.1	2.4
Austria	369	0.1	-4.6	-0.7	0.0	0.7	4.1
Belarus	108	-1.6	-8.5	-2.9	-1.4	0.0	3.4
Belgium	489	-0.4	-3.5	-1.0	-0.4	0.4	2.9
Bulgaria	109	-0.6	-5.3	-1.5	-0.6	0.4	3.2
Croatia	76	0.2	-2.6	-0.7	0.2	1.1	4.7
Cyprus	10	-0.1	-1.5	-0.8	-0.2	0.6	2.3
Czech Republic	323	-0.1	-4.1	-0.8	-0.1	0.6	3.0
Denmark	205	0.0	-2.5	-0.5	0.1	0.6	2.6
Georgia	65	-0.5	-3.4	-1.3	-0.4	0.5	1.9
Germany	2615	-0.2	-7.0	-0.9	-0.1	0.5	3.9
Greece	271	-0.2	-4.2	-0.9	-0.2	0.5	3.2
Hungary	217	0.3	-3.5	-0.6	0.3	1.2	7.5
Ireland	505	-0.2	-3.6	-0.8	-0.2	0.4	2.4
Israel	201	-0.6	-4.3	-1.3	-0.5	0.2	2.1
Italy	2285	-0.2	-5.3	-0.9	-0.2	0.5	4.0
Latvia	29	0.4	-2.6	-0.3	0.4	1.0	4.4
Lithuania	<10	0.0	-1.4	-0.6	-0.4	0.6	2.3
Luxembourg	18	0.5	-2.2	-0.5	0.7	1.5	2.8
Rep of Moldova	44	-0.9	-2.9	-1.4	-1.0	-0.2	0.8
The Netherlands	559	0.3	-2.8	-0.4	0.3	0.9	3.4
North Macedonia	82	-0.4	-3.8	-1.4	-0.4	0.7	2.3
Norway[1]	107	0.1	-2.1	-0.4	0.2	0.8	2.3
Poland	646	0.0	-5.1	-0.7	0.0	0.7	4.0
Portugal	152	-0.6	-3.1	-1.2	-0.6	0.1	1.4
Romania	181	-0.5	-6.2	-1.4	-0.5	0.4	5.0
Russian Federation	2330	-0.4	-9.9	-1.2	-0.4	0.5	6.9
Serbia	125	-0.2	-3.9	-1.0	-0.2	0.6	3.4
Slovak Republic	108	0.3	-2.2	-0.6	0.1	1.1	3.9
Slovenia	63	0.2	-2.4	-0.5	0.1	0.9	2.5
Spain	1062	-0.2	-4.6	-0.9	-0.2	0.5	3.4
Sweden	271	-0.1	-5.1	-0.7	-0.1	0.6	3.5
Switzerland	417	-0.2	-5.1	-0.9	-0.2	0.5	3.7
Turkey	1613	-0.5	-8.1	-1.4	-0.5	0.3	6.4
Ukraine	121	-0.4	-3.0	-1.0	-0.3	0.3	2.9
United Kingdom	3183	-0.4	-5.4	-1.1	-0.4	0.3	8.0

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

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 who have never had a transplant.

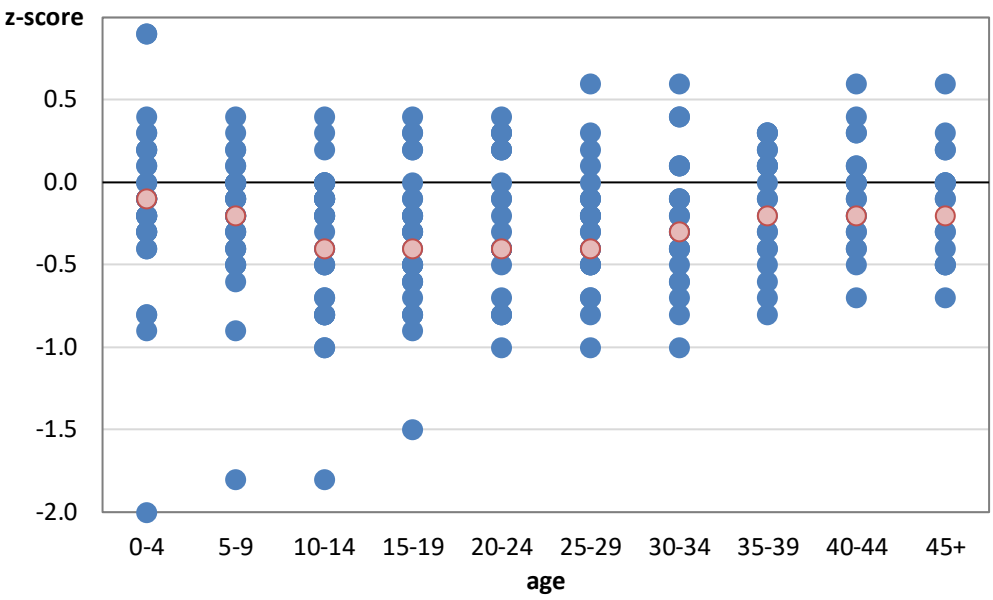
Country	N	Mean	Min	25th pctl (25% of the patients are below this z-score for height)	Median (50% of the patients are below this z-score for height)	75th pctl (75% of the patients are below this z-score for height)	Max
Austria	318	-0.2	-3.4	-0.8	-0.3	0.3	2.7
Belgium	618	-0.3	-3.9	-1.0	-0.2	0.4	2.7
Bulgaria	74	-0.1	-2.5	-0.8	-0.3	0.7	2.1
Croatia	36	0.0	-3.3	-0.5	0.1	0.8	1.6
Czech Republic	242	-0.1	-3.4	-0.7	-0.1	0.4	2.7
Denmark	254	0.2	-2.2	-0.5	0.2	0.9	3.2
Germany	3179	-0.1	-4.0	-0.8	-0.1	0.7	4.0
Greece	263	-0.5	-4.0	-1.3	-0.5	0.2	2.0
Hungary	148	-0.2	-3.2	-1.0	-0.2	0.5	2.7
Ireland	583	-0.4	-5.1	-1.0	-0.4	0.3	2.1
Israel	305	-0.6	-4.3	-1.3	-0.5	0.2	2.4
Italy	2695	-0.5	-4.4	-1.2	-0.5	0.2	4.0
Latvia	<10	0.4	-1.1	-0.3	0.5	1.1	1.5
Lithuania	15	0.9	-0.8	0.3	0.9	1.7	2.4
Luxembourg	18	-0.2	-2.6	-0.8	0.0	0.3	2.4
Rep of Moldova	<10	-0.1	-1.9	-1.3	0.0	1.1	1.3
The Netherlands	779	0.3	-2.8	-0.4	0.3	1.0	3.9
North Macedonia	33	-0.5	-2.5	-1.2	-0.7	-0.2	2.4
Norway[1]	150	0.4	-2.5	-0.3	0.4	1.0	2.8
Poland	190	-0.2	-2.8	-1.0	-0.2	0.4	2.8
Portugal	115	-0.8	-3.0	-1.5	-0.8	-0.2	1.4
Russian Federation	671	-0.3	-5.7	-1.0	-0.3	0.4	3.4
Serbia	45	0.1	-1.2	-0.5	0.0	0.7	2.3
Slovak Republic	139	0.1	-3.0	-0.5	0.1	0.9	2.4
Slovenia	31	0.2	-1.5	-0.8	0.2	0.7	2.9
Spain	871	-0.6	-4.7	-1.3	-0.7	0.0	2.3
Sweden	335	0.1	-2.8	-0.5	0.2	0.7	3.3
Switzerland	445	-0.2	-3.7	-0.8	-0.2	0.4	2.8
Turkey	173	-0.8	-6.3	-1.4	-0.7	-0.1	1.3
Ukraine	27	-0.7	-3.0	-1.0	-0.8	-0.2	0.9
United Kingdom	5127	-0.4	-6.0	-1.0	-0.4	0.3	3.7

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

Note: Belarus and Georgia have 0% coverage for adults and are excluded from the table.
Note: Albania, Armenia, Cyprus 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 2018 who never had a transplant.



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

This graph shows the median z-scores for height by age group. Each country is represented by a dot (in blue) and the overall median estimate is in red. The overall median z-scores for height tend to slowly decrease up to the teenage years and then rise again 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 2018 who never had a transplant.

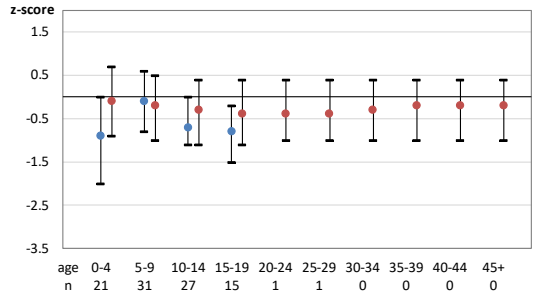
Age at height measurement	N	Mean	Min	25th pctl	Median	75th pctl	Max
0-4	4519	-0.1	-9.9	-0.9	-0.1	0.7	7.5
5-9	6075	-0.3	-7.0	-1.0	-0.2	0.5	8.0
10-14	5511	-0.4	-8.1	-1.1	-0.4	0.4	4.1
15-19	4801	-0.4	-6.3	-1.1	-0.4	0.4	3.4
20-24	4146	-0.3	-6.0	-1.0	-0.4	0.4	3.7
25-29	3522	-0.3	-3.9	-1.0	-0.4	0.4	4.0
30-34	2734	-0.3	-4.3	-1.0	-0.3	0.4	3.6
35-39	2005	-0.2	-4.7	-1.0	-0.2	0.4	3.3
40-44	1410	-0.2	-3.4	-1.0	-0.2	0.4	3.3
45+	2272	-0.2	-5.1	-1.0	-0.2	0.4	4.0

This table reports the median z-score for height and other descriptive statistics by age group for all the patients seen in 2018 who never had a transplant. The median values reported in this table are shown as red dots in fig 6.2.

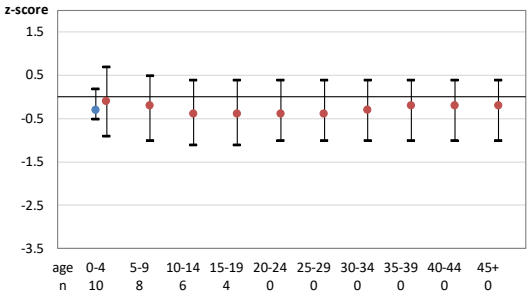
Figure 6.3 Quartiles of z-scores for height by age group and by country. All patients seen in 2018 who never had a transplant.

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

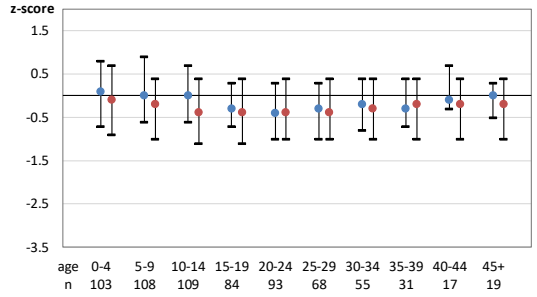
Quartiles of z-scores for height: Albania



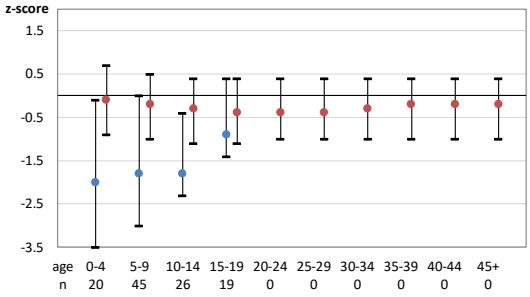
Quartiles of z-scores for height: Armenia



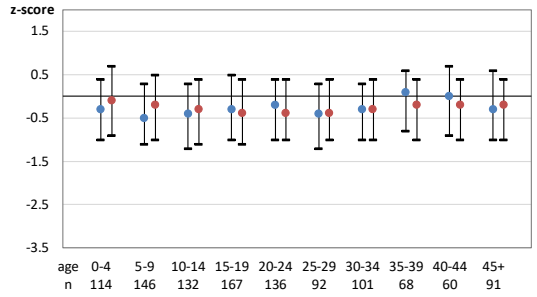
Quartiles of z-scores for height: Austria



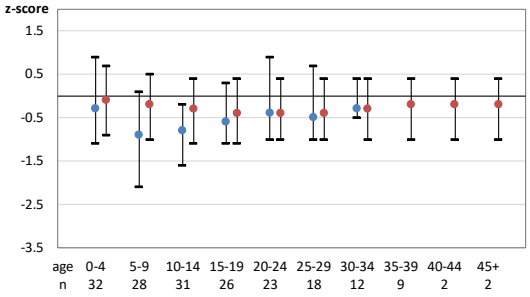
Quartiles of z-scores for height: Belarus



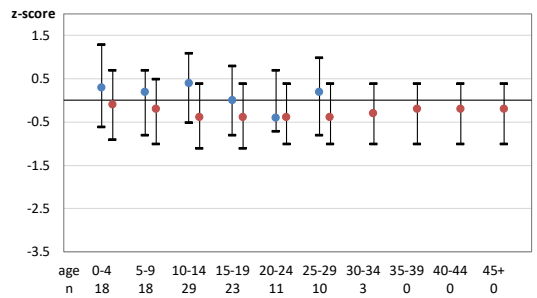
Quartiles of z-scores for height: Belgium



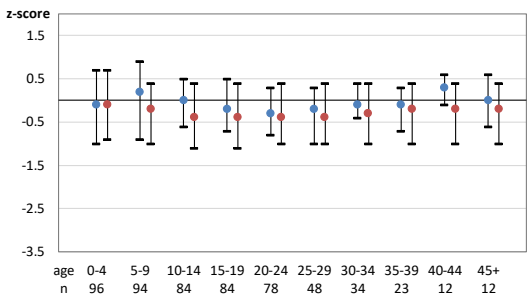
Quartiles of z-scores for height: Bulgaria



Quartiles of z-scores for height: Croatia

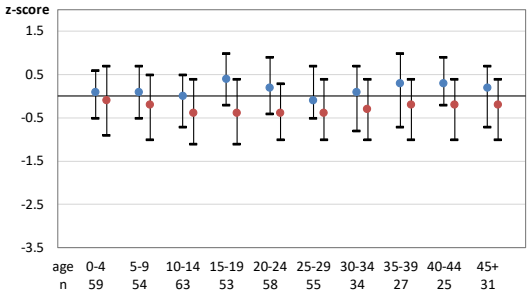


Quartiles of z-scores for height: Czech Republic

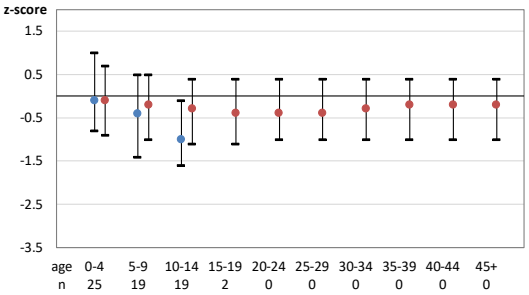


[figure 6.3 continued]

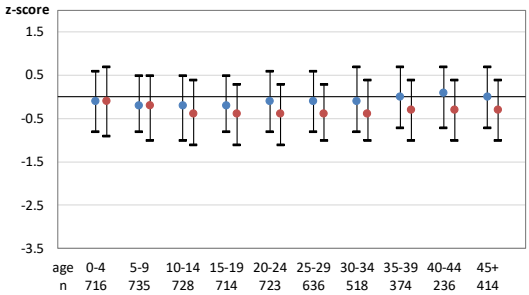
Quartiles of z-scores for height: Denmark



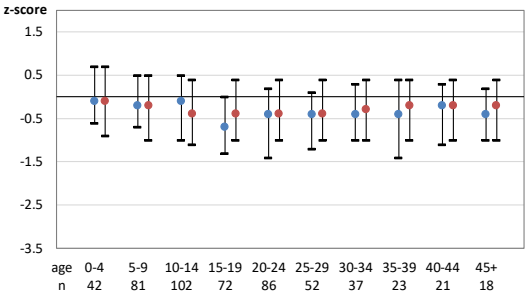
Quartiles of z-scores for height: Georgia



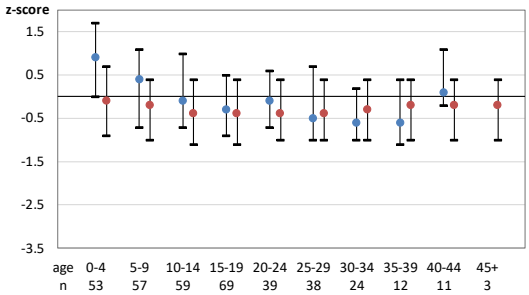
Quartiles of z-scores for height: Germany



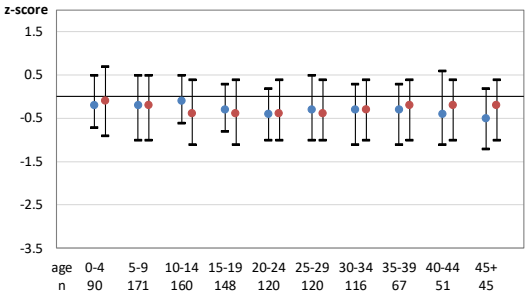
Quartiles of z-scores for height: Greece



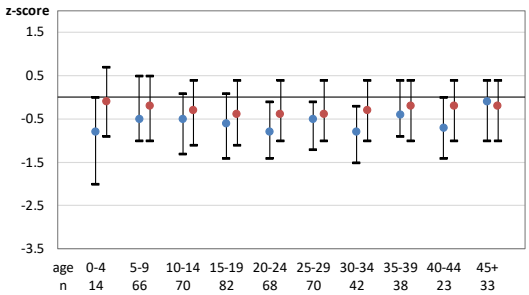
Quartiles of z-scores for height: Hungary



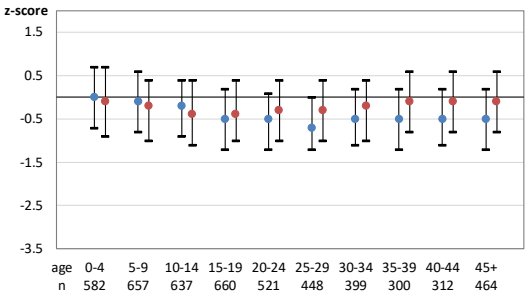
Quartiles of z-scores for height: Ireland



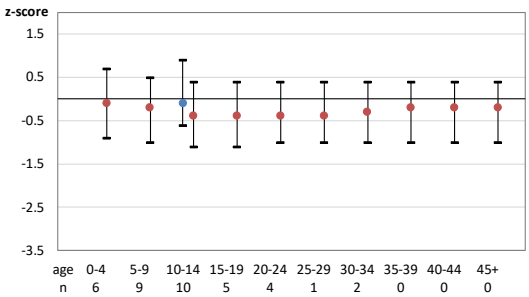
Quartiles of z-scores for height: Israel



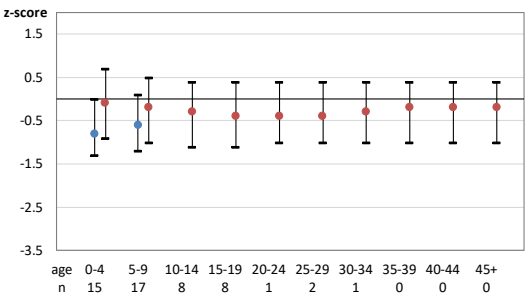
Quartiles of z-scores for height: Italy



Quartiles of z-scores for height: Latvia

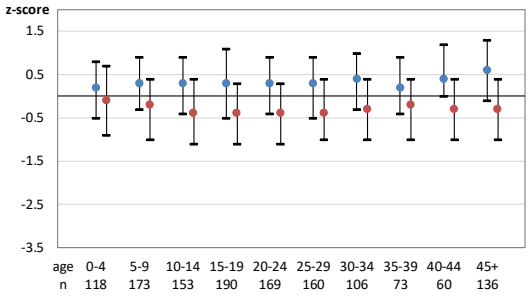


Quartiles of z-scores for height: Rep of Moldova

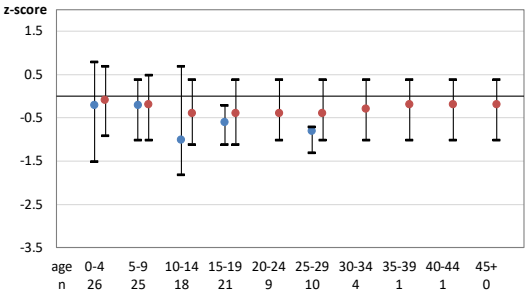


[figure 6.3 continued]

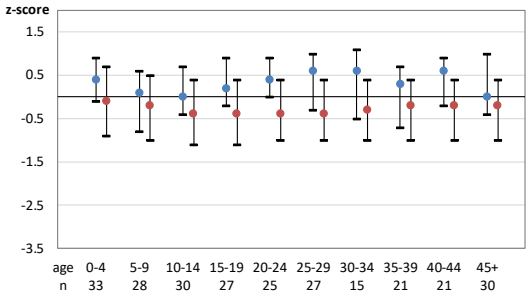
Quartiles of z-scores for height: The Netherlands



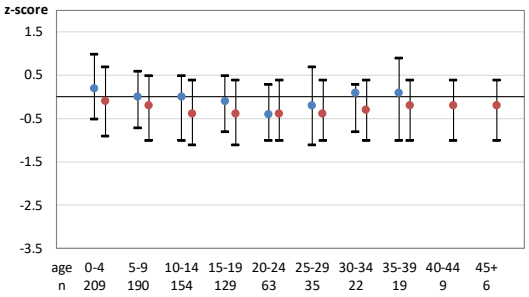
Quartiles of z-scores for height: North Macedonia



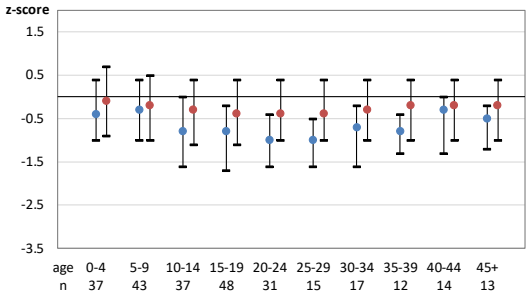
Quartiles of z-scores for height: Norway



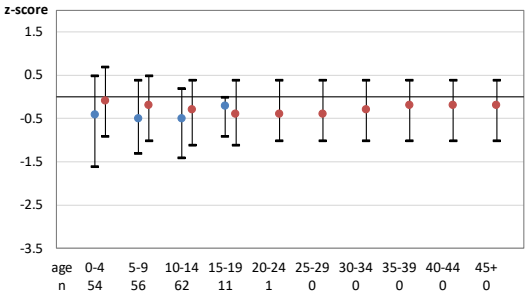
Quartiles of z-scores for height: Poland



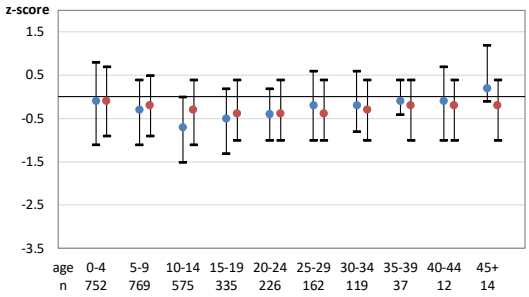
Quartiles of z-scores for height: Portugal



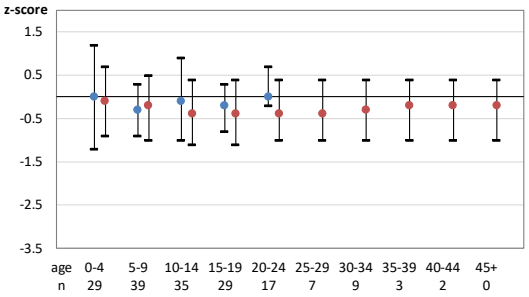
Quartiles of z-scores for height: Romania



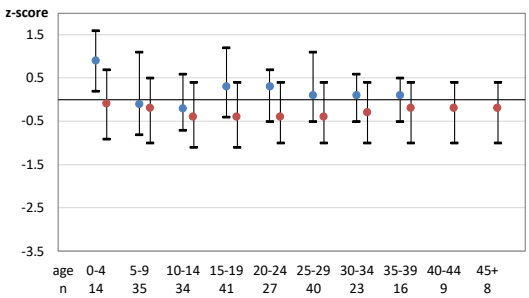
Quartiles of z-scores for height: Russian Federation



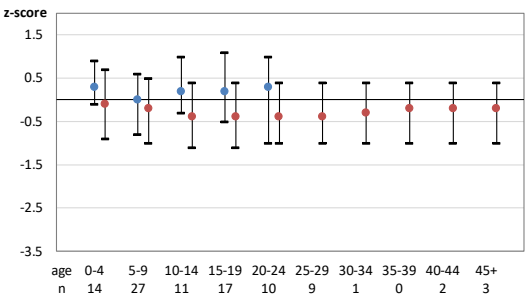
Quartiles of z-scores for height: Serbia



Quartiles of z-scores for height: Slovak Republic

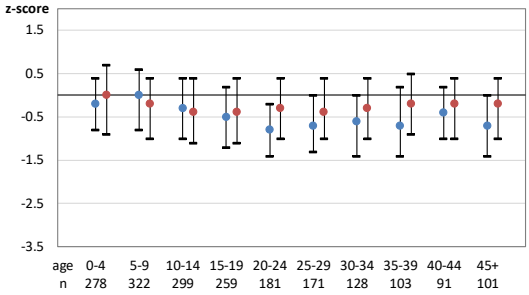


Quartiles of z-scores for height: Slovenia

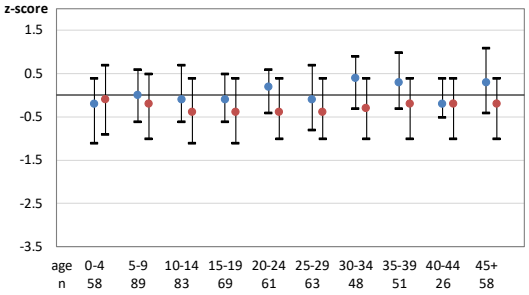


[figure 6.3 continued]

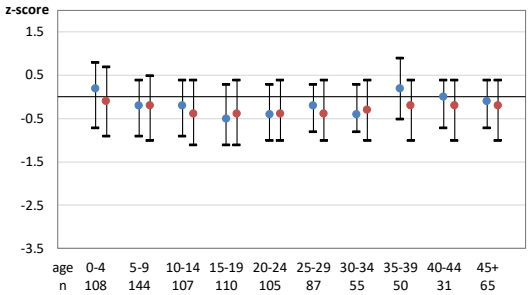
Quartiles of z-scores for height: Spain



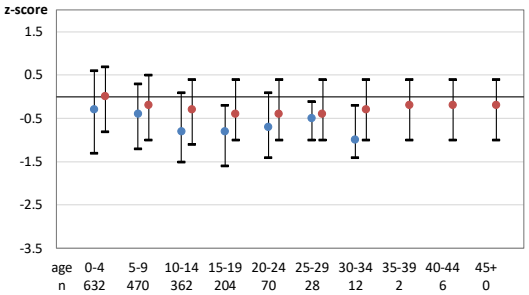
Quartiles of z-scores for height: Sweden



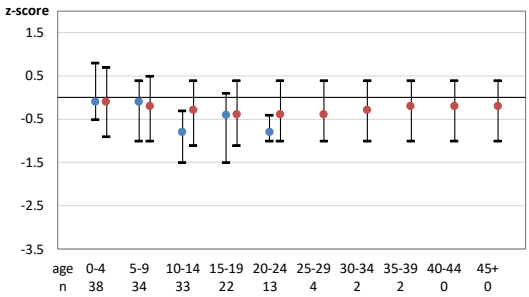
Quartiles of z-scores for height: Switzerland



Quartiles of z-scores for height: Turkey



Quartiles of z-scores for height: Ukraine



Quartiles of z-scores for height: United Kingdom

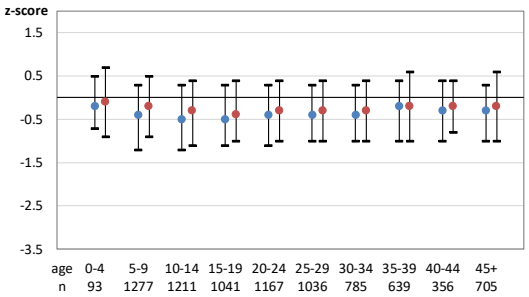


Table 6.5 Z-scores for weight: descriptive statistics by country. Patients aged 17 years or younger who never had a transplant.

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	104	-0.8	-6.5	-1.5	-0.7	0.0	2.9
Armenia	28	-0.5	-3.5	-1.2	-0.1	0.4	1.3
Austria	370	-0.2	-3.9	-0.8	-0.1	0.4	2.5
Belarus	108	-1.8	-7.9	-2.9	-1.6	-0.5	2.3
Belgium	489	-0.5	-4.2	-1.1	-0.4	0.1	2.7
Bulgaria	111	-1.1	-8.9	-1.7	-1.0	0.0	2.6
Croatia	79	-0.3	-4.0	-1.0	-0.4	0.5	4.7
Cyprus	11	-0.2	-2.8	-1.1	-0.3	0.4	2.5
Czech Republic	323	-0.3	-4.8	-1.0	-0.2	0.5	2.6
Denmark	205	-0.3	-3.6	-1.0	-0.3	0.4	2.0
Georgia	65	-0.5	-3.5	-1.4	-0.6	0.3	1.7
Germany	2621	-0.4	-9.1	-1.0	-0.3	0.3	2.6
Greece	274	0.1	-3.7	-0.7	0.1	0.8	2.9
Hungary	216	-0.4	-4.7	-1.1	-0.2	0.5	4.9
Ireland	516	0.0	-9.0	-0.6	0.0	0.6	2.7
Israel	201	-0.5	-6.1	-1.2	-0.4	0.3	2.8
Italy	2291	-0.3	-6.1	-0.9	-0.2	0.5	5.1
Latvia	29	-0.3	-2.5	-0.9	-0.2	0.3	3.6
Lithuania	<10	-0.7	-1.6	-1.3	-1.1	-0.1	0.9
Luxembourg	18	0.0	-2.6	-0.6	-0.1	0.5	3.2
Rep of Moldova	44	-1.1	-4.0	-2.1	-0.9	-0.2	1.0
The Netherlands	559	-0.1	-3.0	-0.6	-0.1	0.5	2.2
North Macedonia	83	-0.4	-2.9	-1.2	-0.6	0.6	2.2
Norway[1]	108	-0.2	-2.3	-0.7	-0.1	0.4	1.9
Poland	653	-0.3	-6.7	-0.9	-0.2	0.4	2.6
Portugal	154	-0.6	-3.7	-1.3	-0.6	0.2	2.0
Romania	184	-1.0	-5.7	-1.7	-0.9	-0.1	2.4
Russian Federation	2337	-0.8	-9.8	-1.6	-0.7	0.1	9.6
Serbia	125	-0.5	-6.3	-1.1	-0.5	0.3	2.6
Slovak Republic	112	-0.1	-2.4	-0.8	-0.1	0.7	2.6
Slovenia	63	-0.2	-3.5	-0.7	-0.2	0.3	1.6
Spain	1064	-0.3	-5.8	-1.0	-0.3	0.4	2.8
Sweden	271	-0.3	-4.3	-1.0	-0.2	0.4	2.9
Switzerland	421	-0.3	-3.7	-0.9	-0.3	0.3	2.9
Turkey	1616	-0.9	-9.4	-1.7	-0.7	0.1	5.4
Ukraine	121	-1.0	-6.1	-1.9	-1.0	-0.2	3.3
United Kingdom	3151	-0.3	-5.3	-0.9	-0.2	0.4	4.4

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

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 who never had a transplant.

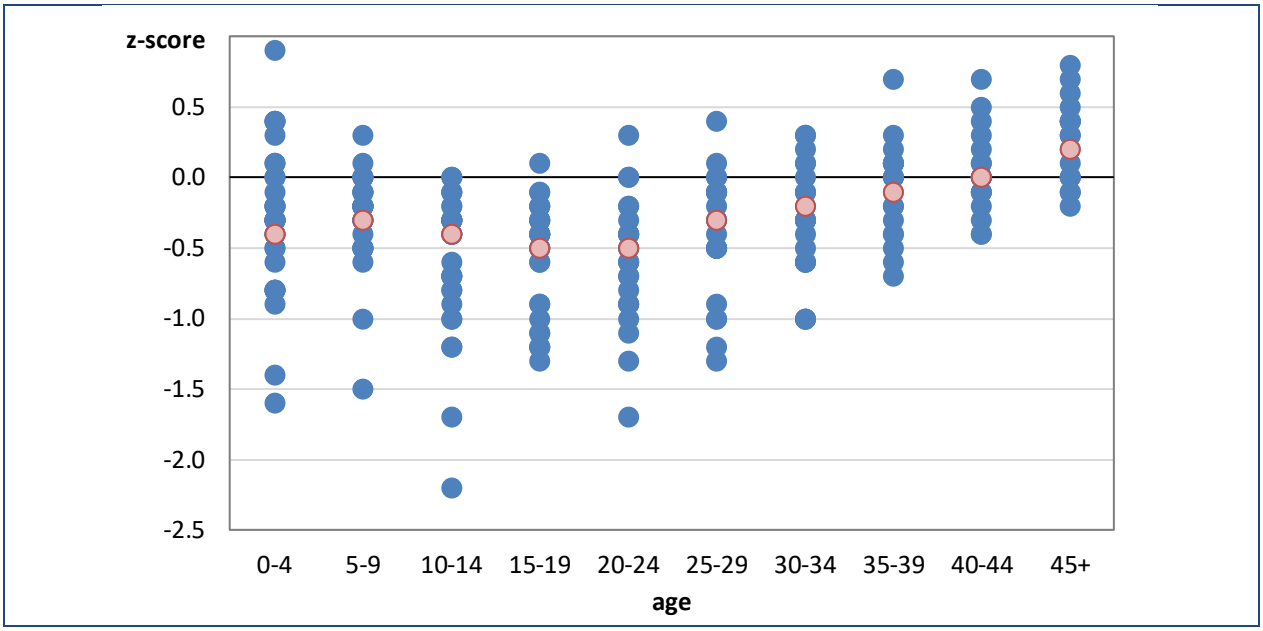
Country	N	Mean	Min	25th pctl (25% of the patients are below this z-score for weight)	Median (50% of the patients are below this z-score for weight)	75th pctl (75% of the patients are below this z-score for weight)	Max
Austria	318	-0.4	-4.0	-1.2	-0.3	0.4	2.3
Belgium	618	-0.3	-4.1	-1.0	-0.2	0.4	2.4
Bulgaria	74	-0.9	-4.7	-1.7	-0.7	-0.1	2.5
Croatia	36	-0.3	-2.3	-0.8	-0.1	0.4	1.4
Czech Republic	242	-0.5	-5.7	-1.2	-0.4	0.3	2.0
Denmark	254	0.0	-3.1	-0.7	0.1	0.8	2.5
Germany	3173	-0.4	-6.2	-1.0	-0.3	0.4	2.8
Greece	263	-0.4	-6.8	-1.0	-0.1	0.4	2.0
Hungary	150	-0.9	-4.6	-1.7	-0.9	0.2	2.3
Ireland	481	-0.2	-6.3	-0.8	-0.1	0.5	2.8
Israel	304	-0.4	-4.6	-1.1	-0.2	0.5	2.3
Italy	2705	-0.5	-6.8	-1.1	-0.4	0.3	3.4
Latvia	<10	-1.1	-2.5	-1.8	-1.3	-0.6	1.0
Lithuania	15	-0.4	-3.3	-0.7	-0.2	0.0	1.3
Luxembourg	18	-0.1	-3.7	-1.0	0.1	0.5	2.0
Rep of Moldova	<10	-1.1	-3.9	-2.5	-0.5	0.3	1.0
The Netherlands	778	0.1	-3.6	-0.5	0.1	0.7	2.6
North Macedonia	33	-0.4	-2.5	-0.9	-0.4	0.4	1.5
Norway[1]	147	0.1	-4.0	-0.4	0.2	0.8	2.8
Poland	190	-0.7	-3.4	-1.3	-0.7	0.1	1.9
Portugal	115	-0.7	-6.8	-1.2	-0.5	0.1	2.5
Russian Federation	671	-1.2	-7.6	-2.0	-1.1	-0.3	2.3
Serbia	46	-0.9	-3.9	-1.5	-0.9	0.2	1.1
Slovak Republic	139	-0.4	-4.2	-1.1	-0.3	0.4	2.4
Slovenia	31	-0.5	-2.5	-1.2	-0.3	0.3	1.1
Spain	873	-0.4	-5.6	-1.1	-0.4	0.3	2.7
Sweden	333	0.0	-3.7	-0.6	0.1	0.6	3.0
Switzerland	445	-0.4	-5.4	-0.9	-0.3	0.3	2.1
Turkey	171	-1.1	-6.2	-1.8	-1.0	-0.2	1.8
Ukraine	27	-1.5	-4.6	-2.5	-1.2	-0.4	0.8
United Kingdom	5113	-0.2	-6.9	-0.9	-0.1	0.6	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: Belarus and Georgia have 0% coverage for adults and are excluded from the table.
Note: Albania, Armenia, Cyprus 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 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 2018 who never had a transplant.



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

This graph shows the median z-scores for weight by age group. Each country is represented by a dot (in blue) and the overall estimate is in red. Overall, the median z-scores for weight decrease from the third youngest age group to the 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 2018 who never had a transplant.

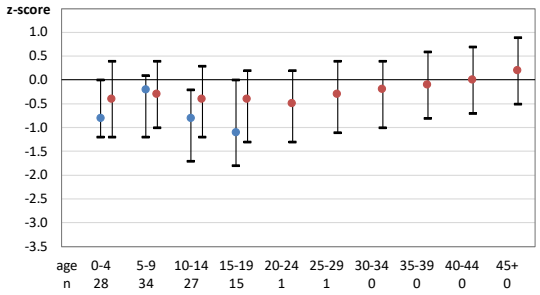
Age at weight measurement	N	Mean	Min	25th pctl	Median	75th pctl	Max
0-4	4584	-0.5	-9.8	-1.2	-0.4	0.4	9.6
5-9	6074	-0.3	-6.0	-1.0	-0.3	0.4	4.4
10-14	5500	-0.5	-9.4	-1.2	-0.4	0.3	2.9
15-19	4777	-0.6	-9.1	-1.3	-0.5	0.2	2.8
20-24	4122	-0.6	-7.6	-1.3	-0.5	0.2	2.9
25-29	3503	-0.4	-6.2	-1.1	-0.3	0.4	2.8
30-34	2714	-0.3	-5.6	-1.0	-0.2	0.4	3.4
35-39	1990	-0.1	-4.6	-0.8	-0.1	0.6	3.0
40-44	1397	-0.1	-4.2	-0.7	0.0	0.7	2.8
45+	2262	0.1	-6.9	-0.5	0.2	0.9	3.0

This table reports the median z-score for weight and other descriptive statistics by age group for all the patients seen in 2018 who never had a transplant. The median values reported in this table are shown as red dots in fig 6.4.

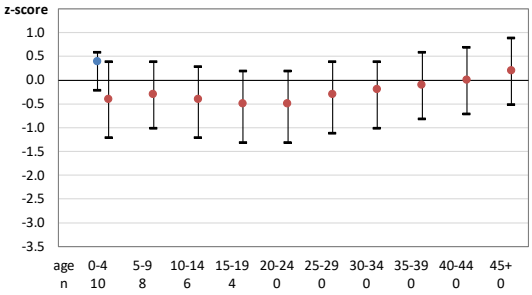
Figure 6.5 Quartiles of z-scores for weight by age group and by country. All patients seen in 2018 who never had a transplant.

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

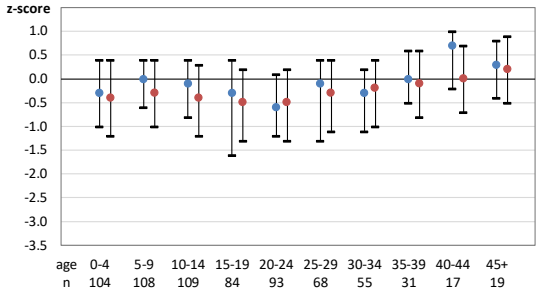
Quartiles of z-scores for weight: Albania



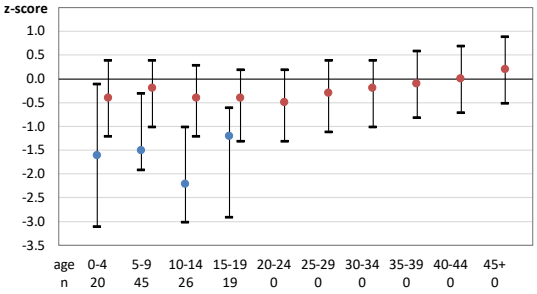
Quartiles of z-scores for weight: Armenia



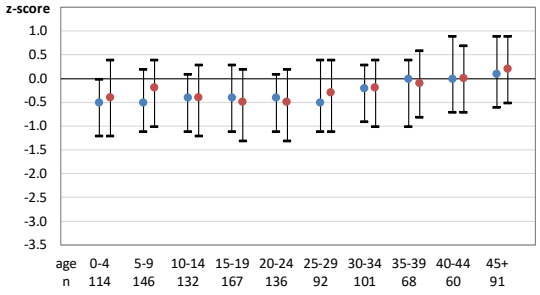
Quartiles of z-scores for weight: Austria



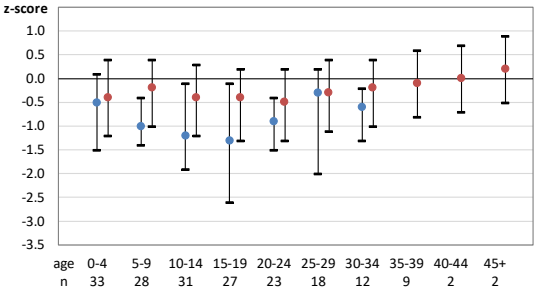
Quartiles of z-scores for weight: Belarus



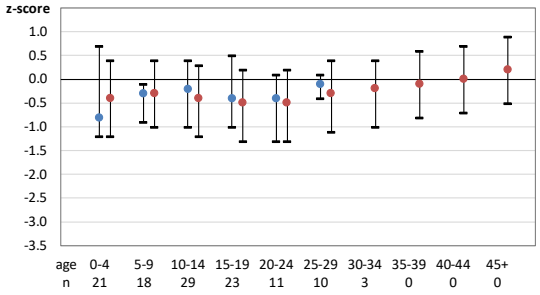
Quartiles of z-scores for weight: Belgium



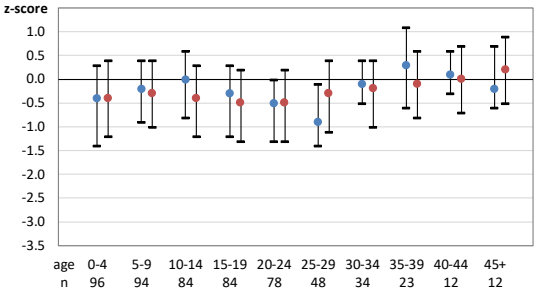
Quartiles of z-scores for weight: Bulgaria



Quartiles of z-scores for weight: Croatia

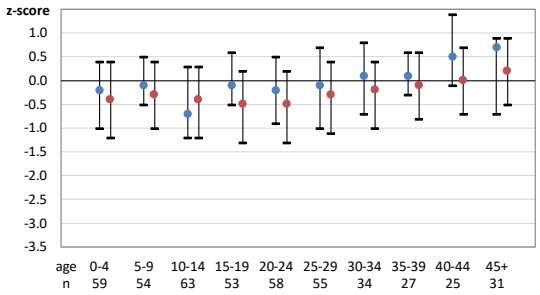


Quartiles of z-scores for weight: Czech Republic

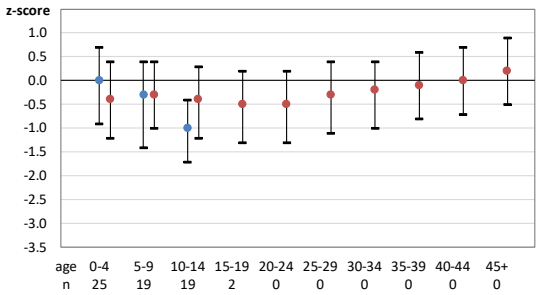


[figure 6.5 continued]

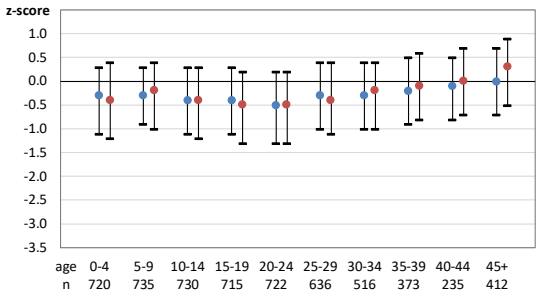
Quartiles of z-scores for weight: Denmark



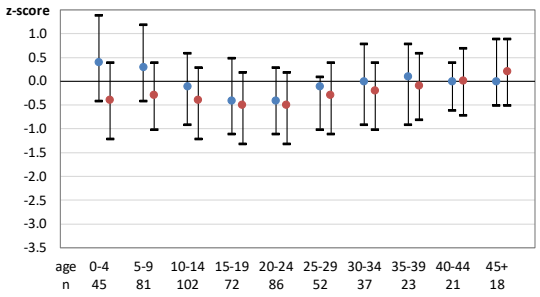
Quartiles of z-scores for weight: Georgia



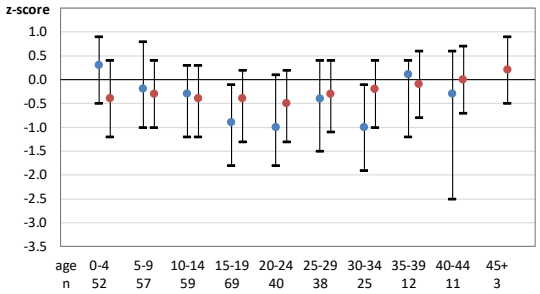
Quartiles of z-scores for weight: Germany



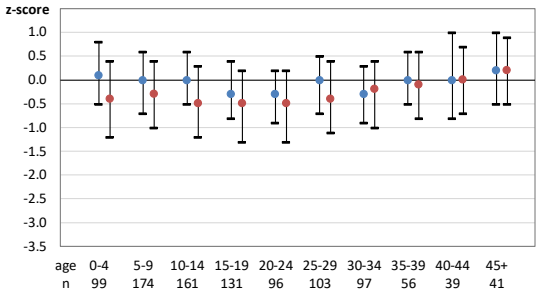
Quartiles of z-scores for weight: Greece



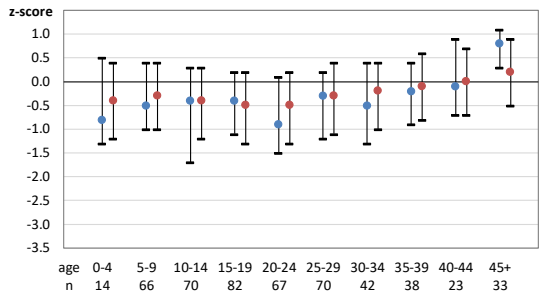
Quartiles of z-scores for weight: Hungary



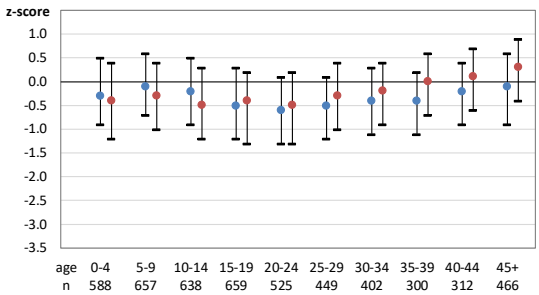
Quartiles of z-scores for weight: Ireland



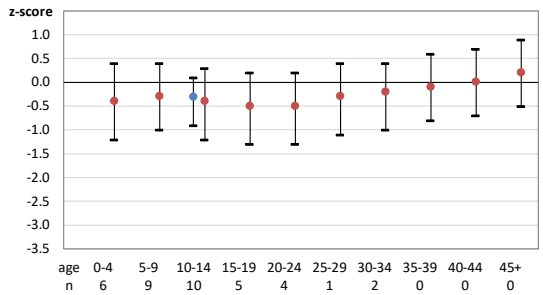
Quartiles of z-scores for weight: Israel



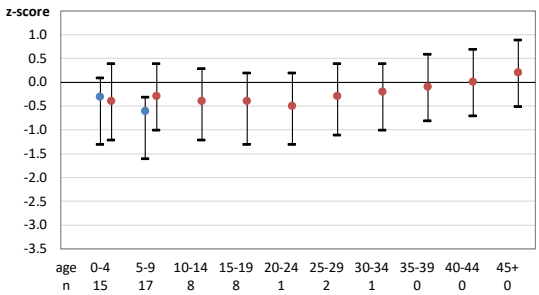
Quartiles of z-scores for weight: Italy



Quartiles of z-scores for weight: Latvia

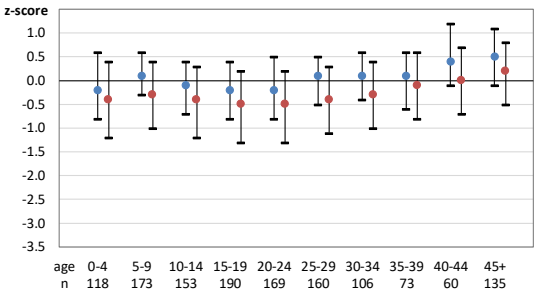


Quartiles of z-scores for weight: Rep of Moldova

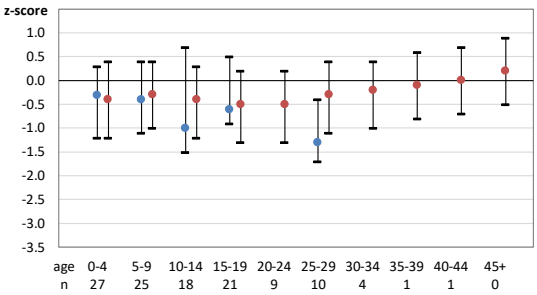


[figure 6.5 continued]

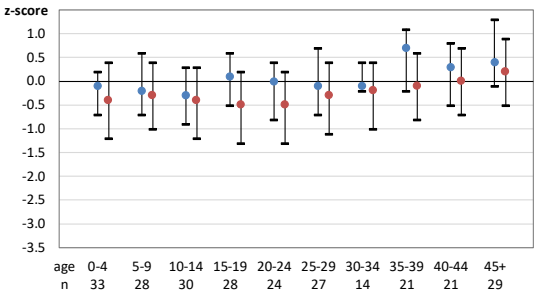
Quartiles of z-scores for weight: The Netherlands



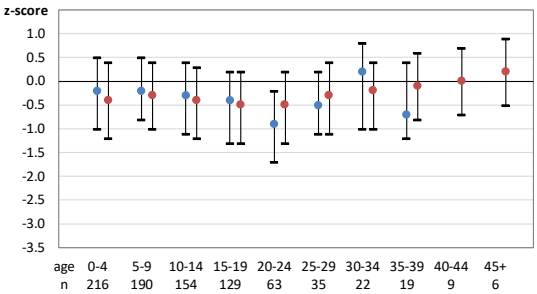
Quartiles of z-scores for weight: North Macedonia



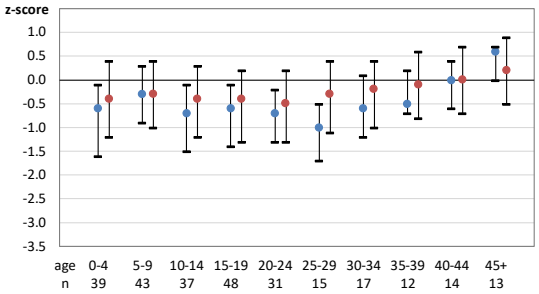
Quartiles of z-scores for weight: Norway



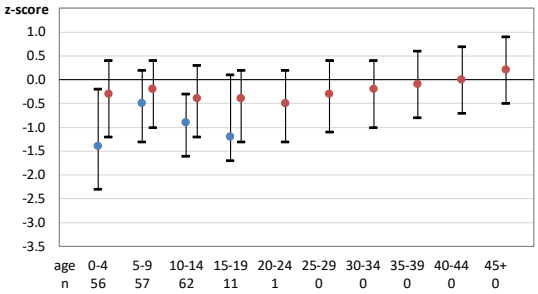
Quartiles of z-scores for weight: Poland



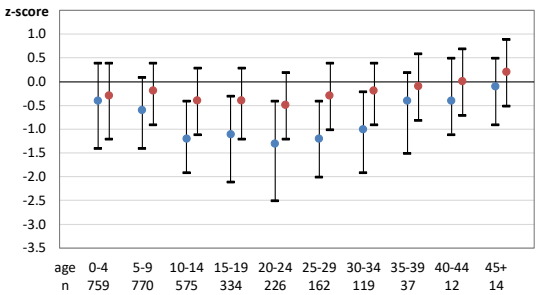
Quartiles of z-scores for weight: Portugal



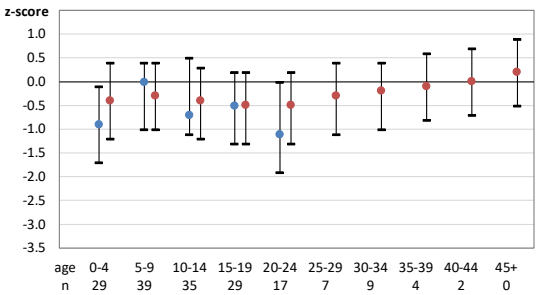
Quartiles of z-scores for weight: Romania



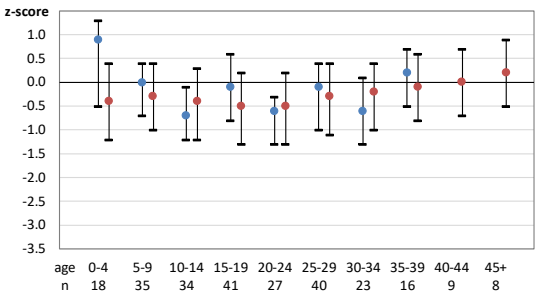
Quartiles of z-scores for weight: Russian Federation



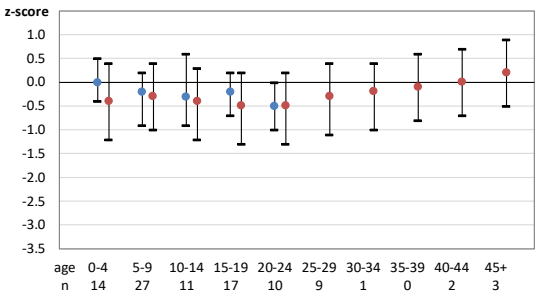
Quartiles of z-scores for weight: Serbia



Quartiles of z-scores for weight: Slovak Republic

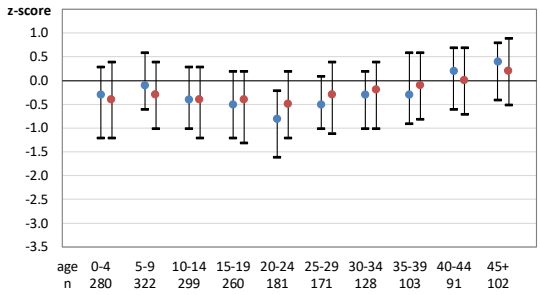


Quartiles of z-scores for weight: Slovenia

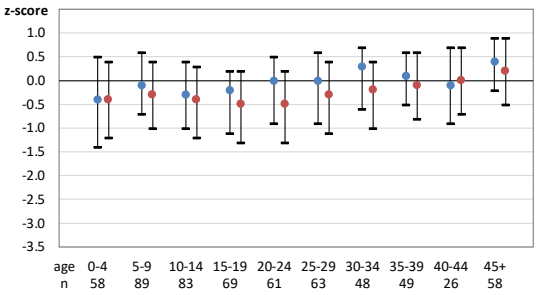


[figure 6.5 continued]

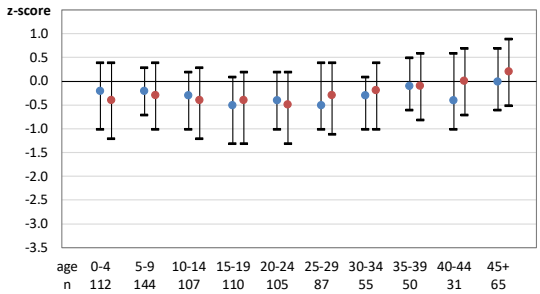
Quartiles of z-scores for weight: Spain



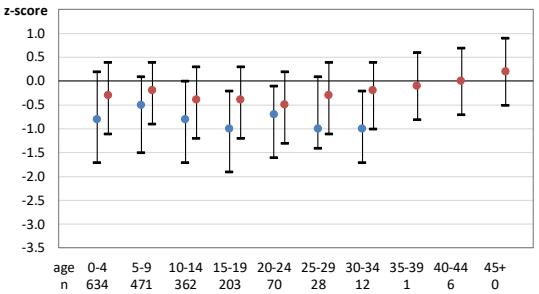
Quartiles of z-scores for weight: Sweden



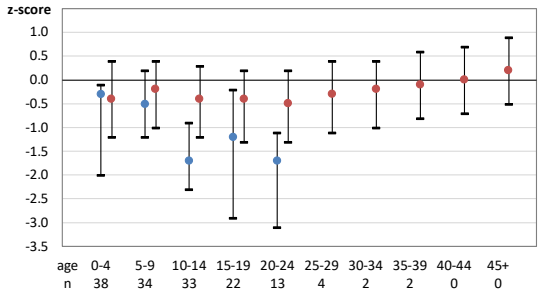
Quartiles of z-scores for weight: Switzerland



Quartiles of z-scores for weight: Turkey



Quartiles of z-scores for weight: Ukraine



Quartiles of z-scores for weight: United Kingdom

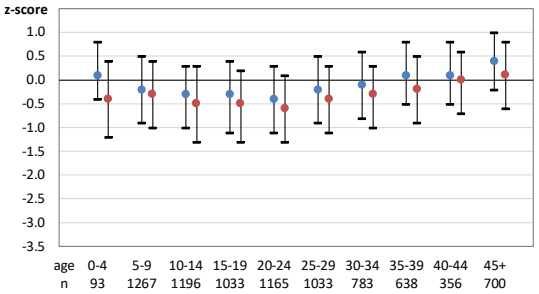


Table 6.8 Z-scores for BMI: descriptive statistics by country. All patients seen in 2018 aged 2-17 years who never had a transplant.

Country	N	N Miss	Mean	Min	25th pctl (25% of the patients are below this z-score for BMI)	Median (50% of the patients are below this z-score for BMI)	75th pctl (75% of the patients are below this z-score for BMI)	Max
Albania	91	9	-0.5	-4.8	-1.4	-0.3	0.3	3.4
Armenia	24	0	-0.1	-5.4	-0.9	0.3	1.0	2.1
Austria	329	0	-0.3	-5.0	-0.8	-0.2	0.5	2.2
Belarus	108	1	-0.9	-4.5	-1.7	-0.7	0.1	2.3
Belgium	456	0	-0.4	-3.8	-1.0	-0.4	0.3	2.6
Bulgaria	96	2	-0.8	-4.0	-1.7	-0.8	-0.2	2.4
Croatia	68	3	-0.7	-4.2	-1.2	-0.6	0.2	1.9
Cyprus	10	2	-0.3	-2.7	-1.5	-0.4	0.4	2.0
Czech Republic	281	0	-0.3	-4.2	-0.9	-0.2	0.4	2.6
Denmark	169	0	-0.4	-3.5	-1.0	-0.3	0.3	2.0
Georgia	57	0	-0.3	-1.9	-1.1	-0.3	0.3	2.5
Germany	2339	3	-0.4	-6.8	-0.9	-0.3	0.3	2.8
Greece	261	1	0.2	-3.2	-0.5	0.3	1.0	2.9
Hungary	201	42	-0.7	-5.2	-1.3	-0.6	0.0	2.6
Ireland	480	20	0.2	-3.0	-0.4	0.2	0.7	3.8
Israel	197	0	-0.2	-3.4	-0.9	-0.2	0.5	2.6
Italy	2063	11	-0.1	-4.5	-0.8	-0.1	0.6	3.3
Latvia	25	0	-0.8	-2.2	-1.5	-0.8	-0.2	0.9
Lithuania	<10	0	-0.9	-2.1	-1.6	-1.0	0.0	0.0
Luxembourg	12	0	-0.5	-1.9	-1.4	-0.4	0.3	1.0
Rep of Moldova	40	0	-0.8	-3.7	-1.5	-0.8	0.1	2.7
The Netherlands	519	0	-0.2	-3.5	-0.8	-0.2	0.4	2.1
North Macedonia	70	1	-0.2	-3.9	-1.0	-0.2	0.6	2.2
Norway[1]	90	1	-0.3	-2.3	-1.0	-0.3	0.4	1.5
Poland	562	3	-0.4	-6.1	-1.0	-0.3	0.4	2.2
Portugal	143	1	-0.4	-5.1	-1.0	-0.3	0.3	2.4
Romania	159	4	-0.9	-6.5	-1.7	-0.9	0.2	3.1
Russian Federation	2066	9	-0.8	-9.3	-1.6	-0.7	0.1	3.9
Serbia	113	0	-0.3	-3.6	-1.1	-0.2	0.4	2.2
Slovak Republic	108	4	-0.4	-3.3	-1.0	-0.4	0.2	2.8
Slovenia	62	0	-0.4	-4.0	-0.9	-0.3	0.3	1.6
Spain	973	1	-0.2	-5.2	-0.9	-0.2	0.5	2.5
Sweden	249	0	-0.3	-4.6	-0.8	-0.3	0.3	3.1
Switzerland	379	2	-0.2	-3.1	-0.8	-0.2	0.4	4.9
Turkey	1380	5	-0.7	-8.6	-1.5	-0.6	0.3	3.8
Ukraine	114	0	-1.1	-5.6	-1.8	-1.1	-0.2	3.2
United Kingdom	3151	41	0.0	-7.0	-0.6	0.0	0.6	3.3

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

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 2018 aged 18 years or older who never had a transplant.

Country	N	N Miss	Mean	Min	25th pctl (25% of the patients are below this BMI)	Median (50% of the patients are below this BMI)	75th pctl (75% of the patients are below this BMI)	Max
Austria	318	1	21.8	14.5	19.5	21.2	23.6	35.5
Belgium	618	0	22.2	15.8	19.9	21.9	23.7	34.3
Bulgaria	74	0	20.5	13.8	17.9	19.8	22.5	40.1
Croatia	36	0	21.5	15.4	19.8	21.5	22.9	28.1
Czech Republic	242	0	21.5	14.3	19.0	21.1	23.3	33.3
Denmark	254	0	22.7	16.0	19.9	22.0	24.6	38.2
Germany	3171	11	21.7	11.8	19.4	21.2	23.4	40.6
Greece	263	0	22.5	15.0	20.5	22.2	24.1	32.0
Hungary	148	28	20.6	14.3	18.3	20.0	22.3	39.8
Ireland	481	102	22.8	15.5	20.4	22.4	24.6	43.4
Israel	304	1	22.8	14.4	20.3	22.4	25.0	37.9
Italy	2695	32	22.2	12.9	19.9	21.8	24.0	44.8
Latvia	<10	0	18.6	16.3	16.5	17.9	19.6	24.5
Lithuania	15	0	19.9	15.3	18.0	19.6	21.2	24.6
Luxembourg	18	0	22.9	16.8	21.4	22.4	23.8	35.2
Rep of Moldova	<10	0	19.9	15.8	17.8	20.1	22.1	23.7
The Netherlands	778	1	22.5	15.5	20.3	21.9	23.9	45.9
North Macedonia	33	0	22.1	17.0	20.6	21.8	23.4	26.5
Norway[1]	147	3	22.9	15.8	20.0	22.5	24.7	37.7
Poland	190	0	20.9	14.9	18.3	20.4	22.7	36.0
Portugal	115	0	22.2	14.5	20.1	21.6	24.1	38.7
Russian Federation	671	3	19.6	11.8	17.4	19.2	21.3	35.8
Serbia	45	1	19.7	14.4	17.9	19.5	21.8	25.9
Slovak Republic	139	0	21.3	14.2	18.8	21.1	23.3	33.7
Slovenia	31	0	20.6	15.1	18.9	21.1	23.0	24.3
Spain	871	5	22.5	14.9	20.2	22.0	24.2	41.4
Sweden	333	2	22.7	15.6	20.2	22.1	24.2	41.7
Switzerland	445	0	21.7	14.0	19.6	21.5	23.4	37.0
Turkey	171	3	20.7	13.3	18.9	20.1	22.5	31.7
Ukraine	27	0	19.4	15.2	17.3	19.2	21.3	26.0
United Kingdom	5113	18	23.1	13.2	20.4	22.5	25.1	49.4

[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: Belarus and Georgia have 0% coverage for adults and are excluded from the table.
Note: Albania, Armenia, Cyprus and Romania have <5 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 patients aged 18 years or older, by country.

Table 6.10 BMI: descriptive statistics by country. All male patients seen in 2018 aged 18 years or older who never had a transplant.

Country	N	N Miss	Mean	Min	25th pctl (25% of the patients are below this BMI)	Median (50% of the patients are below this BMI)	75th pctl (75% of the patients are below this BMI)	Max
Austria	165	1	22.5	16.7	19.9	22.1	24.8	32.9
Belgium	335	0	22.6	15.8	20.3	22.3	24.4	34.3
Bulgaria	40	0	21.5	13.8	17.7	21.1	23.2	40.1
Croatia	16	0	21.7	15.4	20.0	21.7	22.9	28.1
Czech Republic	115	0	22.0	15.9	19.2	21.6	24.3	33.3
Denmark	141	0	22.9	16.0	20.6	22.6	25.5	36.6
Germany	1692	6	22.1	12.9	19.8	21.7	24.1	39.1
Greece	140	0	23.1	16.3	21.1	22.9	25.1	32.0
Hungary	74	18	21.5	15.2	18.9	21.2	24.0	34.7
Ireland	287	57	23.2	15.5	21.0	23.0	25.1	34.1
Israel	174	0	23.1	14.4	20.8	22.8	25.1	37.9
Italy	1440	16	22.8	14.5	20.7	22.4	24.6	44.8
Lithuania	<10	0	20.7	17.6	18.7	20.2	22.7	24.6
Luxembourg	10	0	22.1	16.8	21.7	22.4	23.6	23.9
The Netherlands	423	0	22.6	15.6	20.5	22.2	24.2	35.7
North Macedonia	21	0	22.5	18.9	20.6	22.3	24.5	26.5
Norway[1]	81	1	23.5	15.9	20.6	23.0	25.9	37.7
Poland	81	0	21.3	14.9	18.2	20.9	23.3	36.0
Portugal	60	0	22.0	16.3	20.0	21.4	24.5	30.9
Russian Federation	345	1	19.8	11.8	17.6	19.4	21.8	33.4
Serbia	26	0	19.8	15.1	17.9	19.3	21.8	25.9
Slovak Republic	66	0	22.2	14.2	19.8	22.0	24.2	33.7
Slovenia	13	0	21.8	17.3	19.5	23.4	23.8	24.3
Spain	480	3	22.9	14.9	20.7	22.6	24.5	40.2
Sweden	180	1	23.2	16.3	20.8	22.4	24.8	38.2
Switzerland	244	0	22.2	14.0	20.3	22.1	23.8	32.4
Turkey	93	1	20.9	14.5	18.9	20.6	22.9	31.7
Ukraine	18	0	19.7	15.2	17.4	19.6	22.0	26.0
United Kingdom	2787	7	23.3	13.2	20.9	23.0	25.4	40.1

[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: Belarus and Georgia have 0% coverage for adults and are excluded from the table.
Note: Albania, Armenia, Cyprus, 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 2018 aged 18 years or older who never had a transplant.

Country	N	N Miss	Mean	Min	25th pctl (25% of the patients are below this BMI)	Median (50% of the patients are below this BMI)	75th pctl (75% of the patients are below this BMI)	Max
Austria	153	0	21.1	14.5	19.4	20.7	22.5	35.5
Belgium	283	0	21.8	15.8	19.5	21.5	23.2	34.2
Bulgaria	34	0	19.3	15.2	17.9	19.0	20.4	27.7
Croatia	20	0	21.4	17.7	19.8	21.3	22.9	24.8
Czech Republic	127	0	21.0	14.3	18.9	20.6	23.0	33.0
Denmark	113	0	22.4	16.4	19.6	21.5	23.7	38.2
Germany	1479	5	21.1	11.8	19.0	20.7	22.6	40.6
Greece	123	0	21.7	15.0	19.7	21.7	23.1	31.7
Hungary	74	10	19.8	14.3	17.9	19.2	21.6	39.8
Ireland	194	45	22.1	15.9	19.7	21.6	23.4	43.4
Israel	130	1	22.4	16.4	19.6	21.4	24.4	37.4
Italy	1255	16	21.5	12.9	19.3	21.0	23.1	42.4
Latvia	<10	0	17.0	16.3	16.4	16.5	17.3	18.8
Lithuania	<10	0	19.0	15.3	15.6	19.0	21.0	24.2
Luxembourg	<10	0	23.8	18.2	20.6	22.4	25.4	35.2
The Netherlands	355	1	22.3	15.5	20.1	21.6	23.6	45.9
North Macedonia	12	0	21.5	17.0	20.5	21.6	22.4	24.9
Norway[1]	66	2	22.1	15.8	19.7	21.2	23.7	33.1
Poland	109	0	20.6	14.9	18.4	20.2	21.7	35.7
Portugal	55	0	22.4	14.5	20.3	21.9	23.3	38.7
Russian Federation	326	2	19.4	13.0	17.3	18.8	21.1	35.8
Serbia	19	1	19.7	14.4	17.7	19.6	22.1	25.2
Slovak Republic	73	0	20.5	14.3	18.2	19.6	22.5	26.0
Slovenia	18	0	19.8	15.1	17.5	20.2	21.3	23.0
Spain	391	2	21.9	15.1	19.4	21.5	23.4	41.4
Sweden	153	1	22.1	15.6	19.9	21.7	23.5	41.7
Switzerland	201	0	21.2	14.3	19.0	20.7	22.3	37.0
Turkey	78	2	20.3	13.3	18.8	19.9	21.9	29.9
Ukraine	<10	0	18.8	16.6	17.3	18.5	20.8	21.4
United Kingdom	2326	11	22.9	13.5	20.1	21.9	24.5	49.4

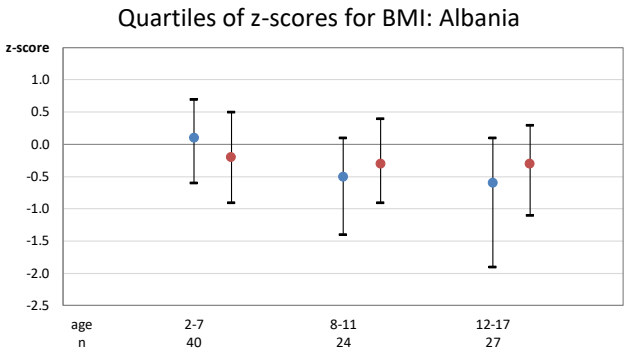
[1] Norway: sometimes any value (instead of last of the year) for height and weight is used when no lung function test was available.

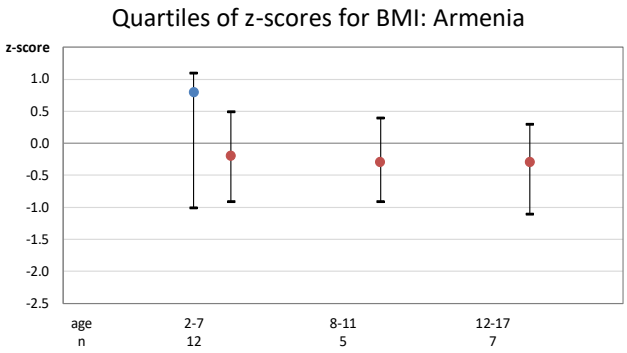
Note: Belarus and Georgia have 0% coverage for adults and are excluded from the table.
Note: Albania, Armenia, Cyprus, Rep of Moldova and Romania have <5 female patients aged 18 years or more at BMI measurement and are excluded from this table.

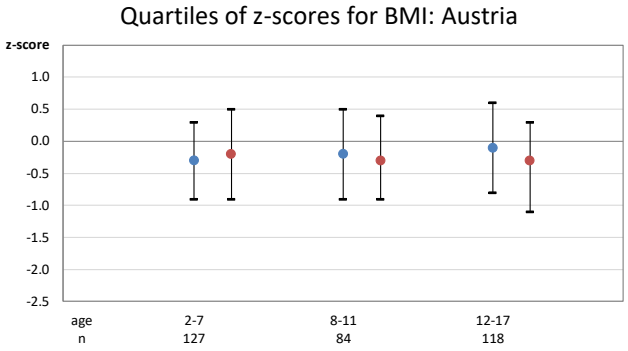
This table reports the median BMI (expressed as absolute values, not as z-scores), the mean BMI and other descriptive statistics for female patients aged 18 years or older, by country.

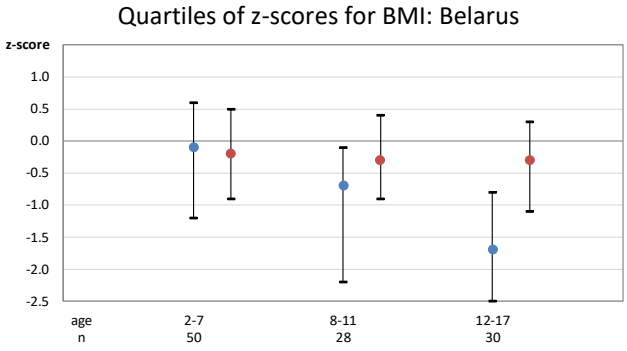
Figure 6.6 Quartiles of z-scores for BMI by age group and country. Patients aged 2-17 years in 2018 who never had a transplant.

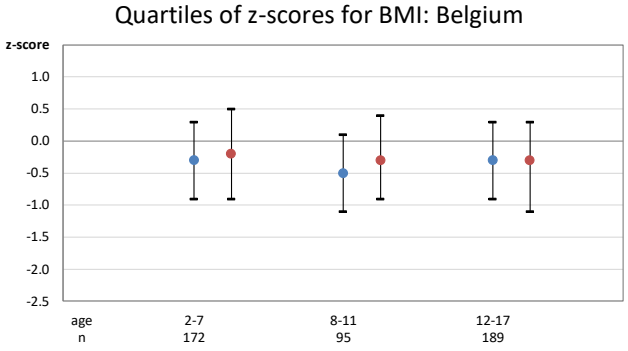
The figures below show the z-scores for BMI by country. The dot is the median, and the whiskers show the 25th and 75th percentiles. In blue are the quartiles for the country, in red are the pooled quartiles computed on all other countries (i.e. excluding that country). We did not compute quartiles where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

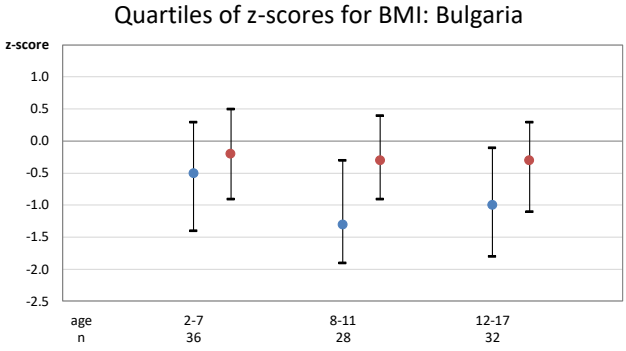


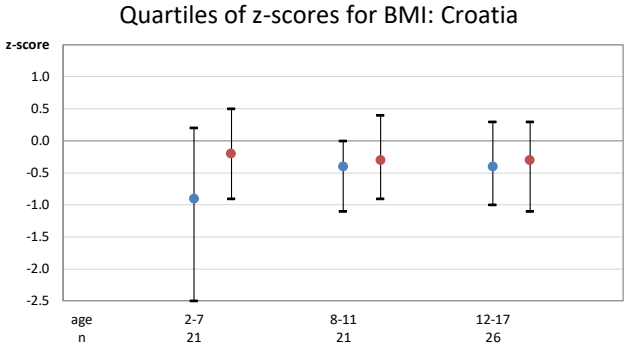


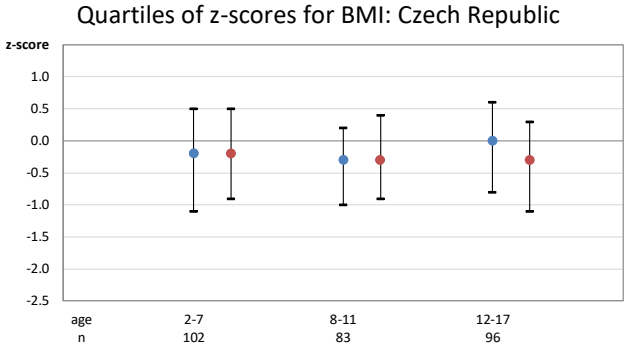






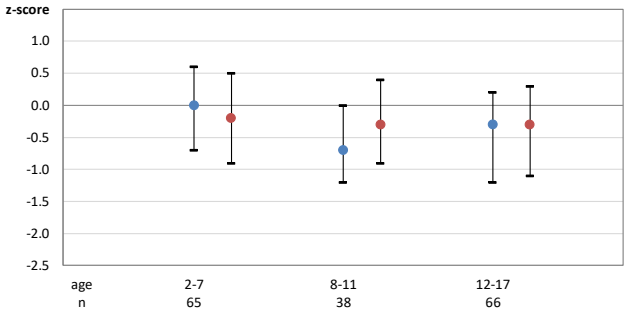




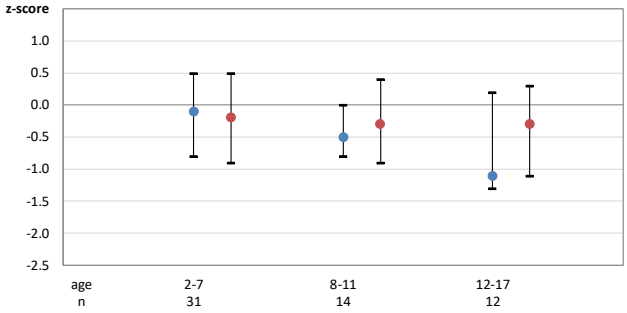


[figure 6.6 continued]

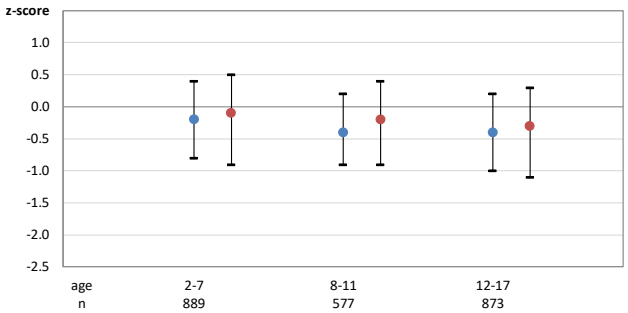
Quartiles of z-scores for BMI: Denmark



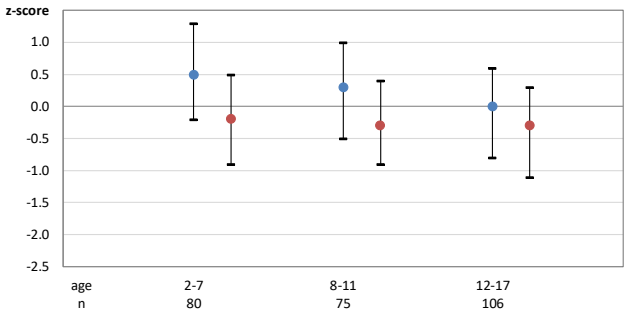
Quartiles of z-scores for BMI: Georgia



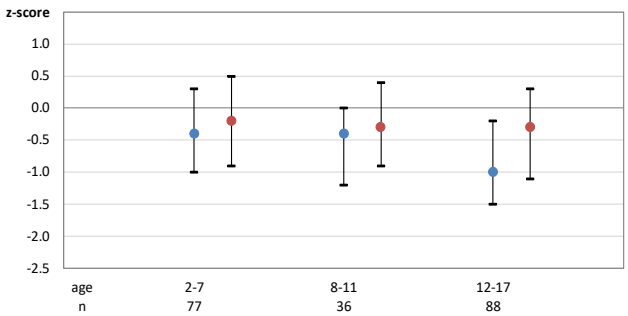
Quartiles of z-scores for BMI: Germany



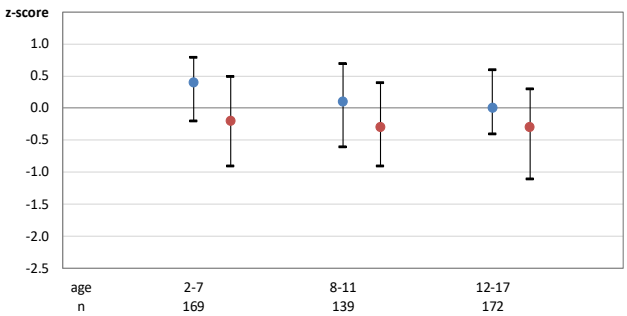
Quartiles of z-scores for BMI: Greece



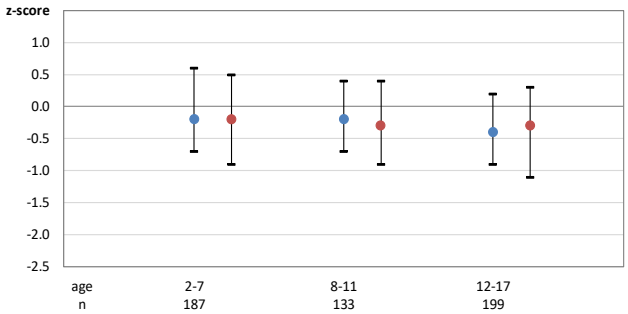
Quartiles of z-scores for BMI: Hungary



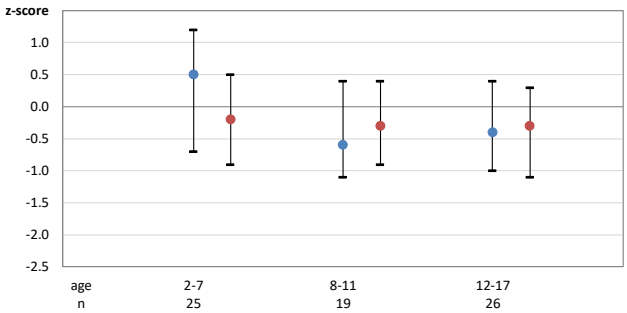
Quartiles of z-scores for BMI: Ireland



Quartiles of z-scores for BMI: The Netherlands

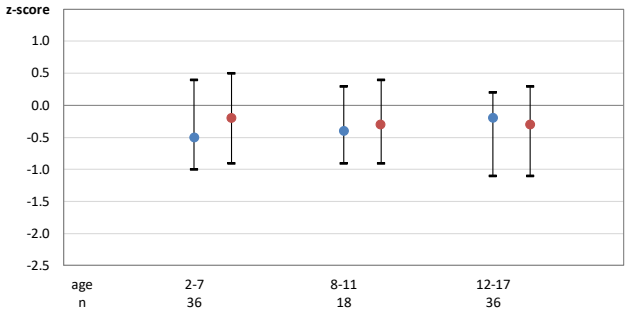


Quartiles of z-scores for BMI: North Macedonia

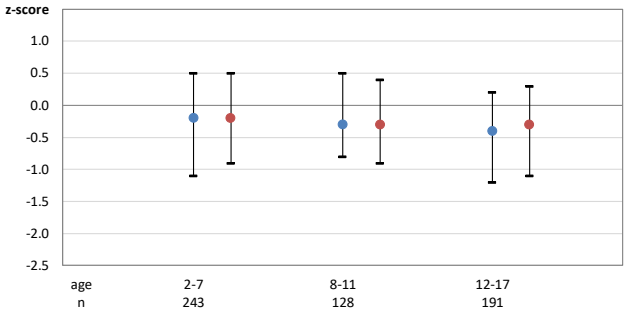


[figure 6.6 continued]

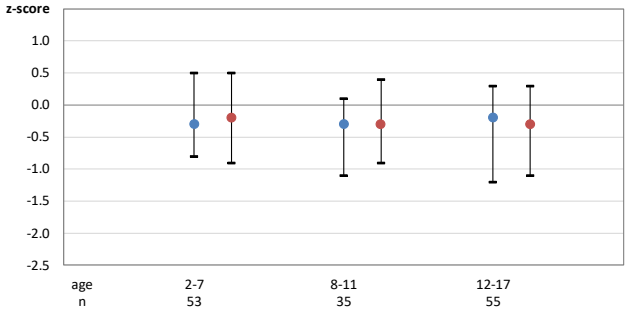
Quartiles of z-scores for BMI: Norway



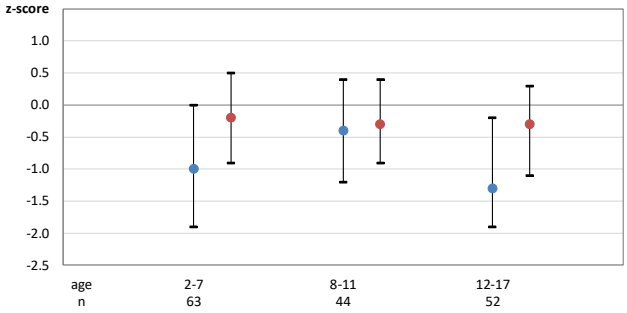
Quartiles of z-scores for BMI: Poland



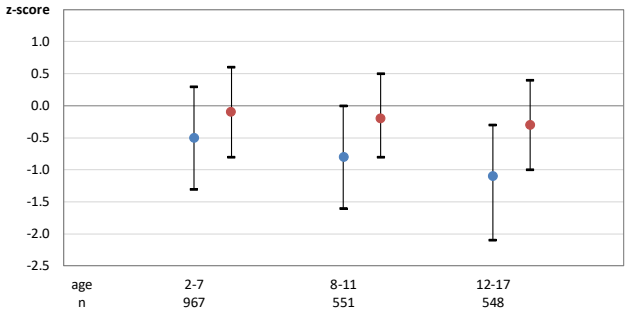
Quartiles of z-scores for BMI: Portugal



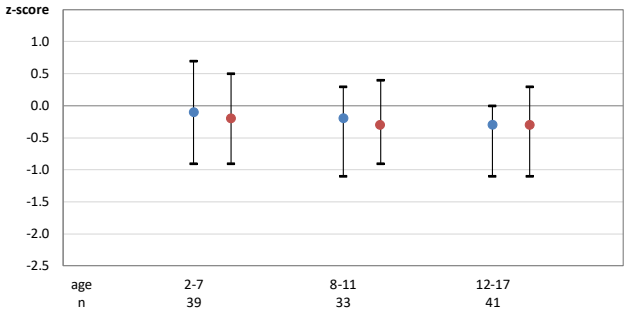
Quartiles of z-scores for BMI: Romania



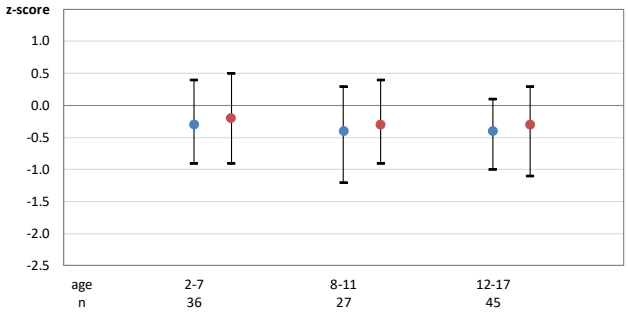
Quartiles of z-scores for BMI: Russian Federation



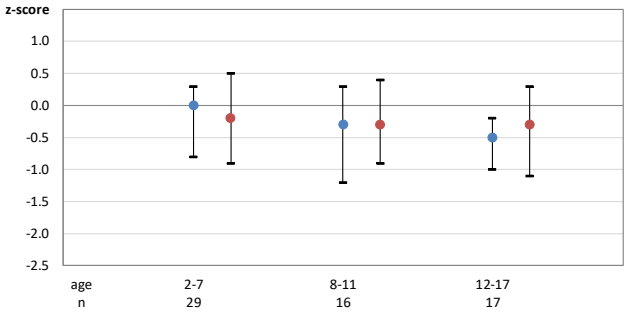
Quartiles of z-scores for BMI: Serbia



Quartiles of z-scores for BMI: Slovak Republic

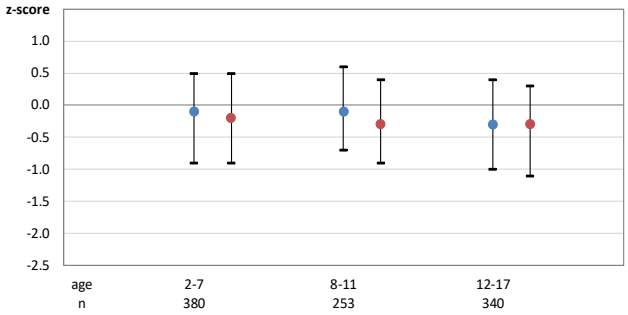


Quartiles of z-scores for BMI: Slovenia

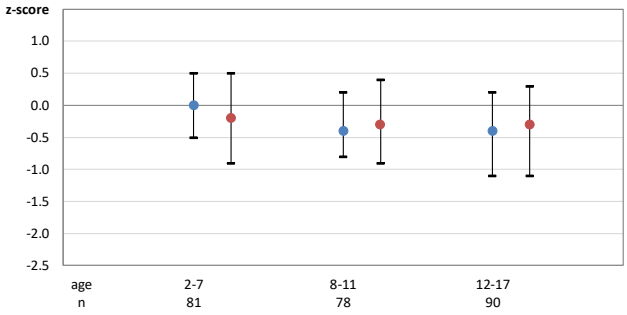


[figure 6.6 continued]

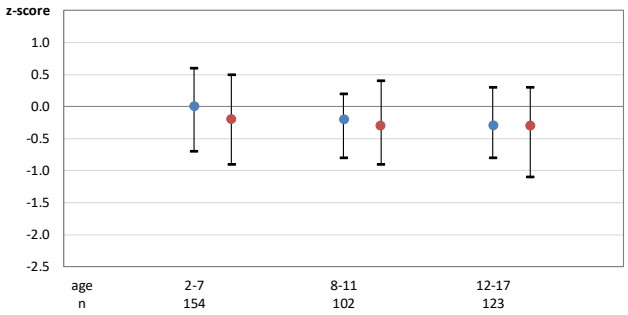
Quartiles of z-scores for BMI: Spain



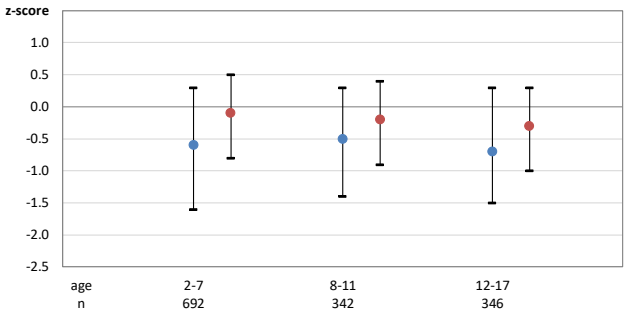
Quartiles of z-scores for BMI: Sweden



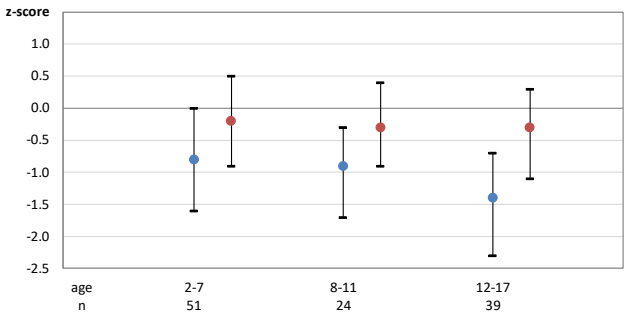
Quartiles of z-scores for BMI: Switzerland



Quartiles of z-scores for BMI: Turkey



Quartiles of z-scores for BMI: Ukraine



Quartiles of z-scores for BMI: United Kingdom

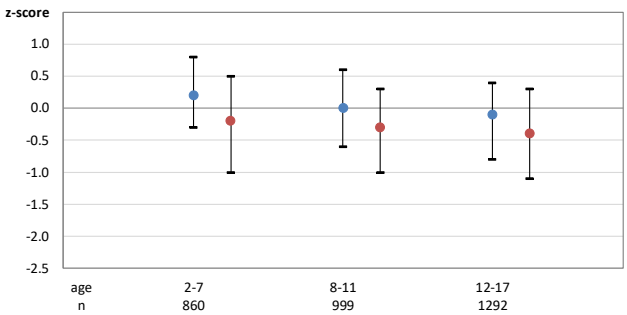
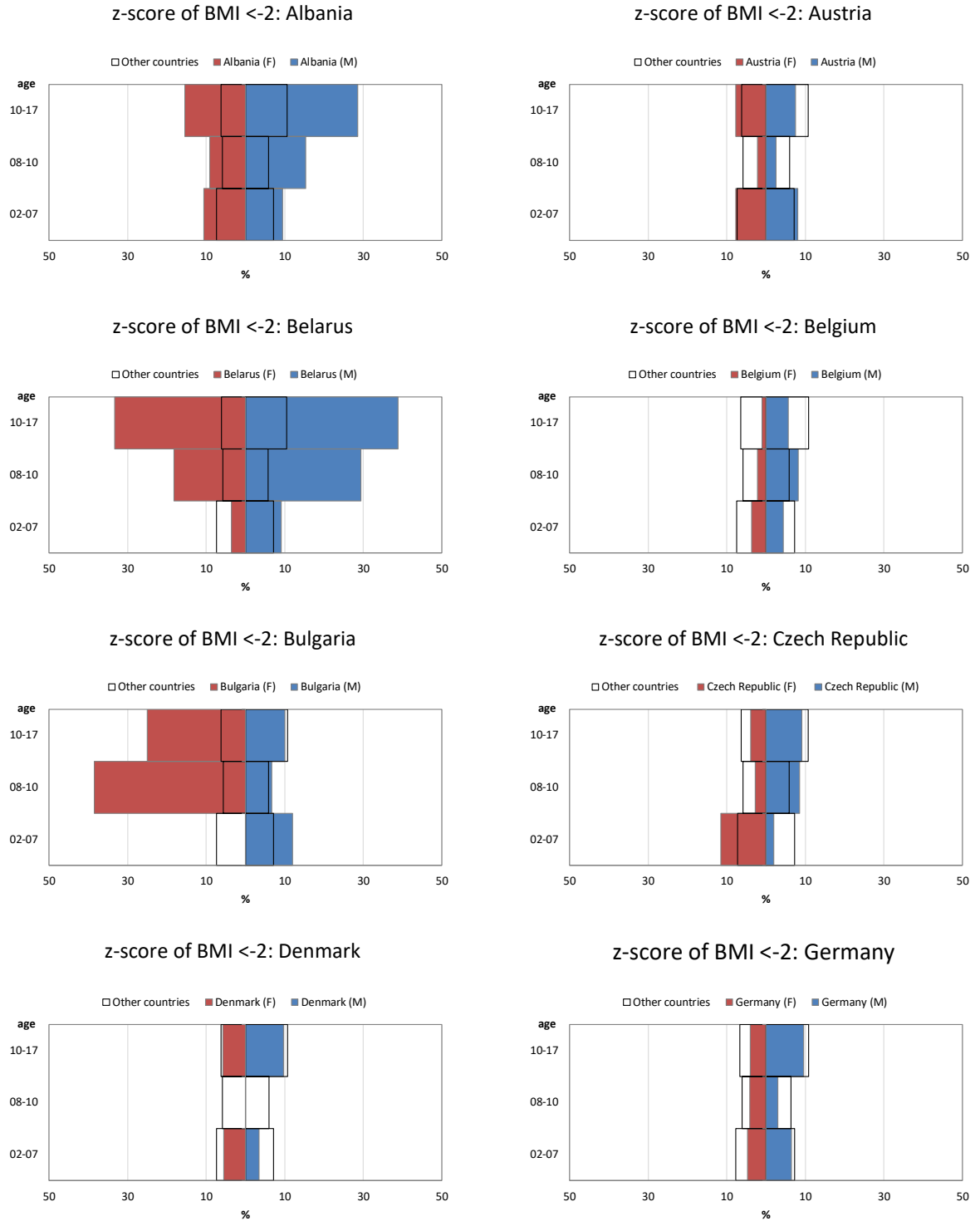


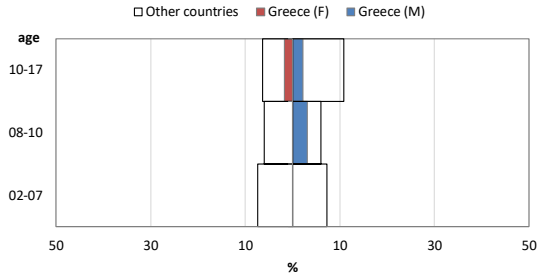
Figure 6.7 Proportion of children underweight (z-score of BMI<-2): age and sex pyramids, by country and overall. Patients aged 2-17 years in 2018 who never had a transplant.

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

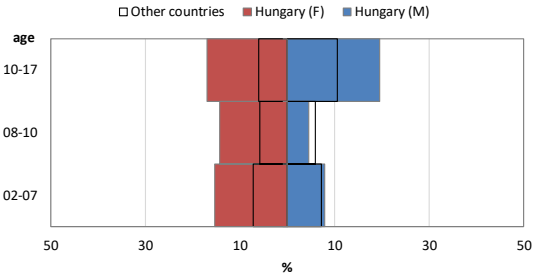


[figure 6.7 continued]

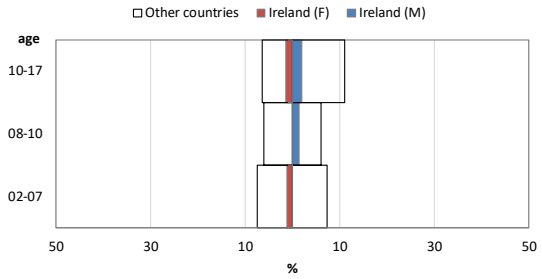
z-score of BMI <-2: Greece



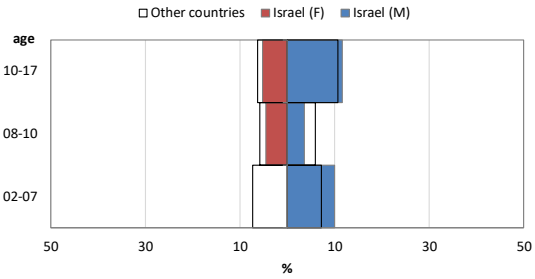
z-score of BMI <-2: Hungary



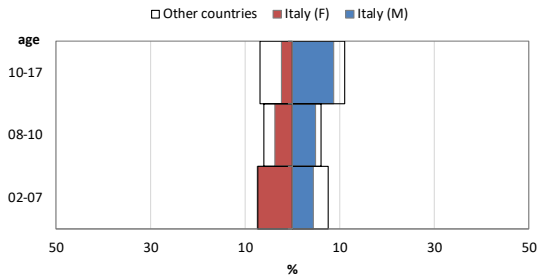
z-score of BMI <-2: Ireland



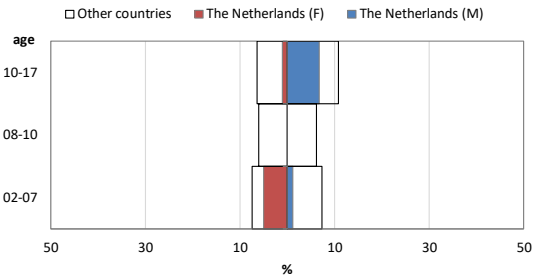
z-score of BMI <-2: Israel



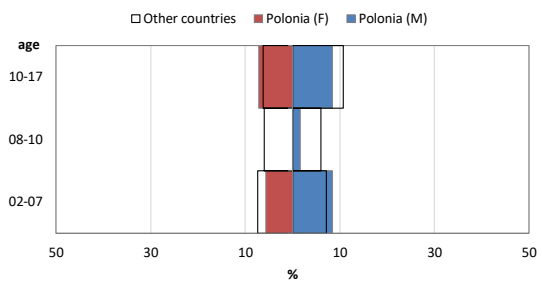
z-score of BMI <-2: Italy



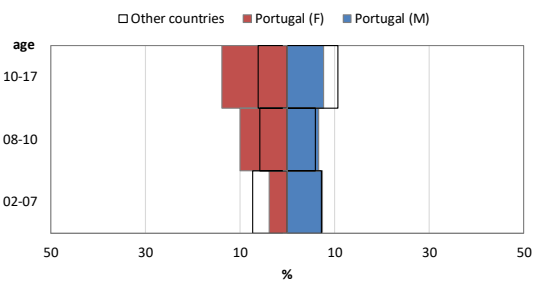
z-score of BMI <-2: The Netherlands



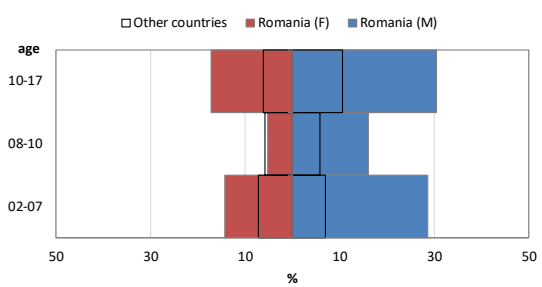
z-score of BMI <-2: Poland



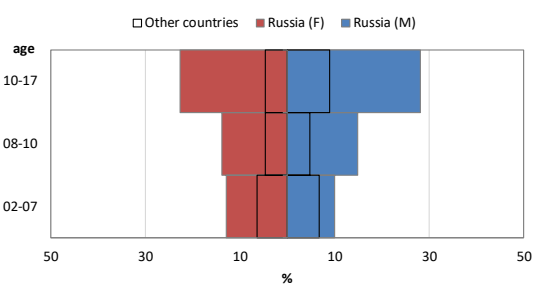
z-score of BMI <-2: Portugal



z-score of BMI <-2: Romania

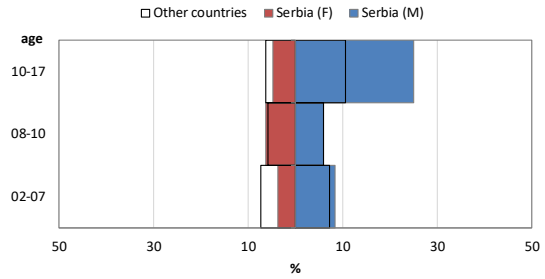


z-score of BMI <-2: Russian Federation

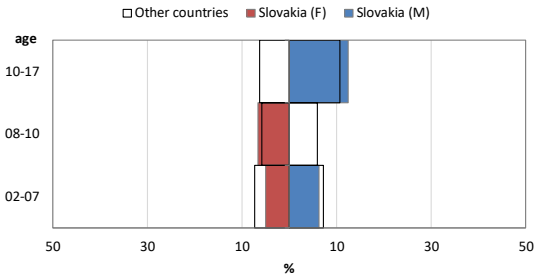


[figure 6.7 continued]

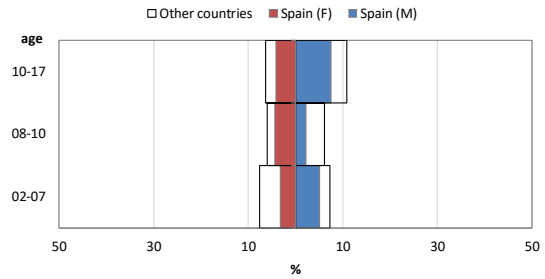
z-score of BMI <-2: Serbia



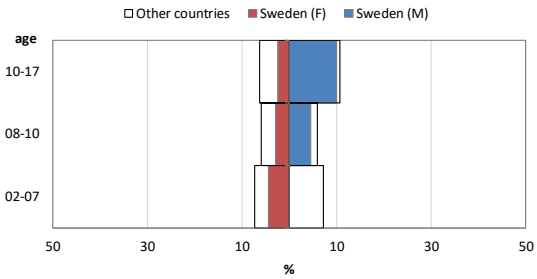
z-score of BMI <-2: Slovak Republic



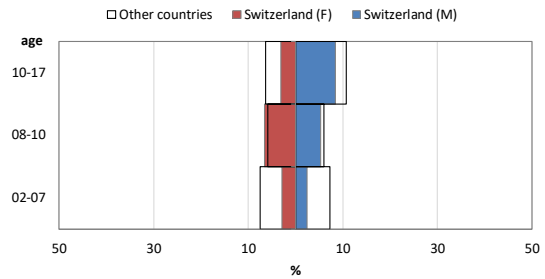
z-score of BMI <-2: Spain



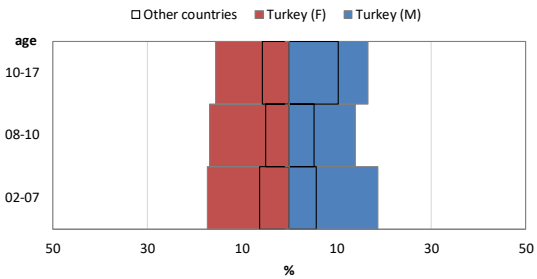
z-score of BMI <-2: Sweden



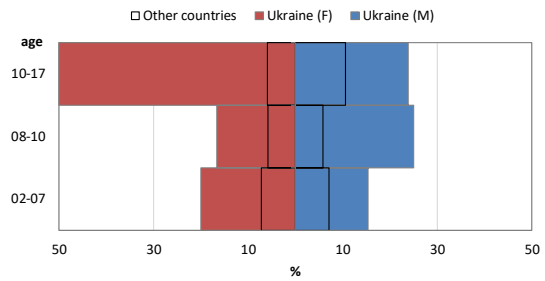
z-score of BMI <-2: Switzerland



z-score of BMI <-2: Turkey



z-score of BMI <-2: Ukraine



z-score of BMI <-2: United Kingdom

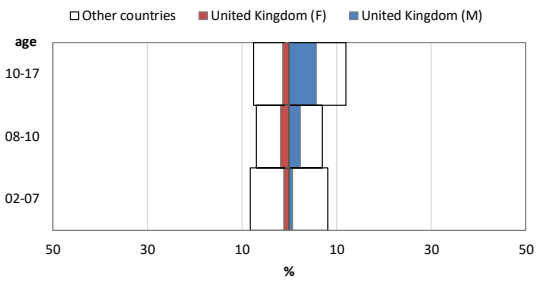
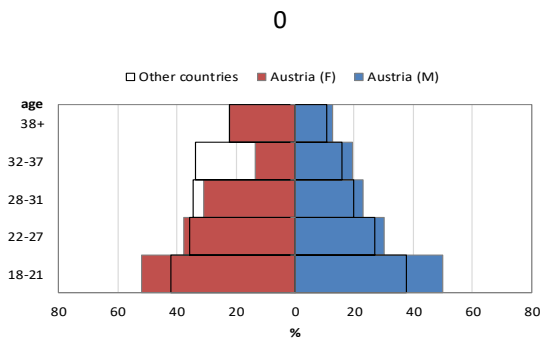
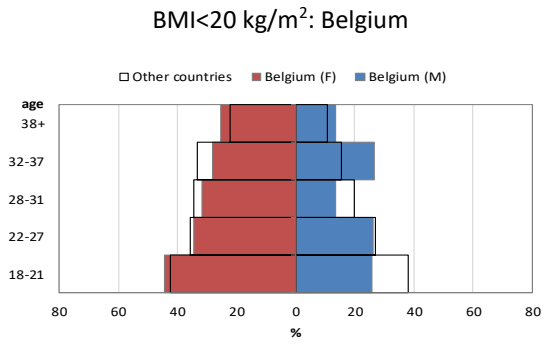
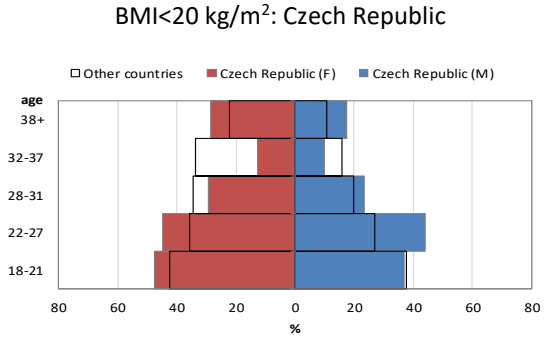


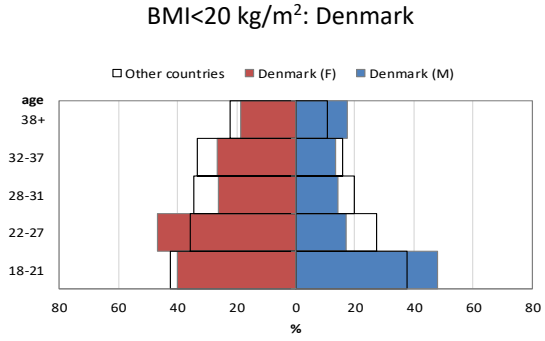
Figure 6.8 Proportion of adult patients with BMI<20: age and sex pyramids, by country and overall. Patients aged 18 years or older in 2018 who never had a transplant.

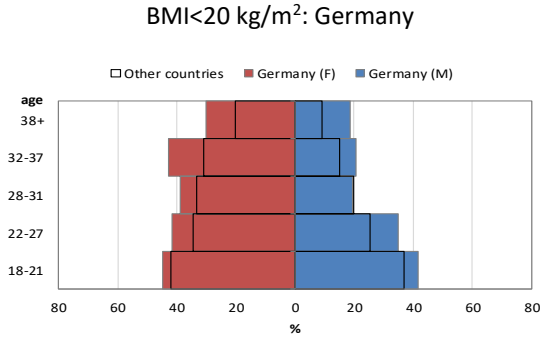
The coloured bars (red for females, blue for males) represent the percentage of underweight patients in the selected country, whereas the non-coloured bars represent the percentage of underweight patients in all the remaining countries (i.e. excluding that country). We therefore excluded from the graphs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

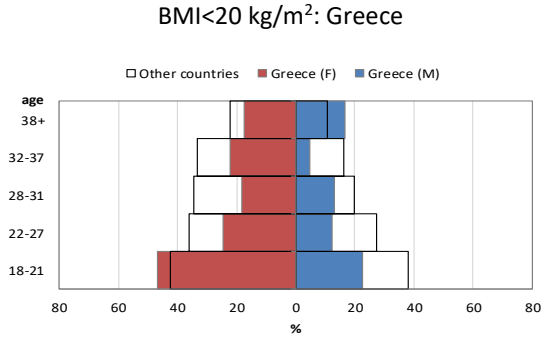


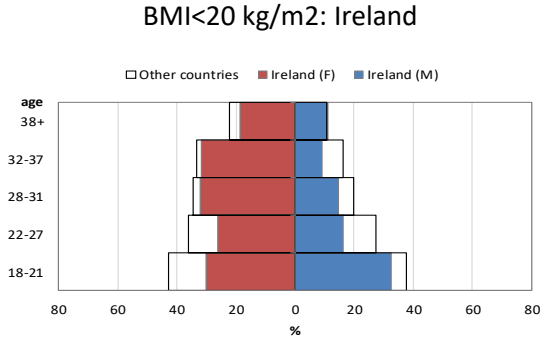


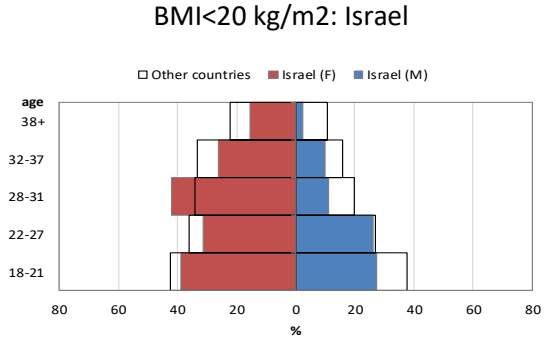






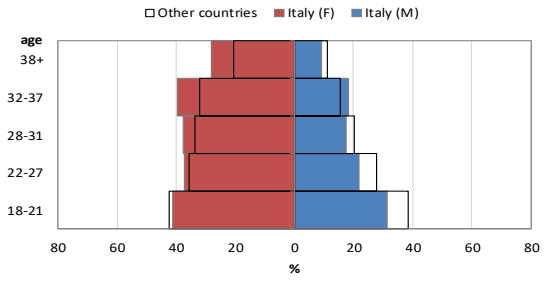




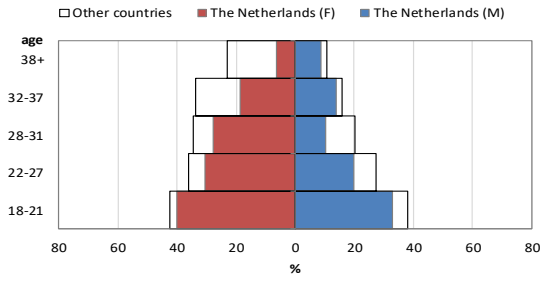


[figure 6.8 continued]

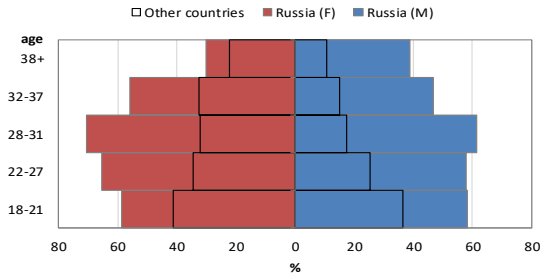
BMI<20 kg/m^2: Italy



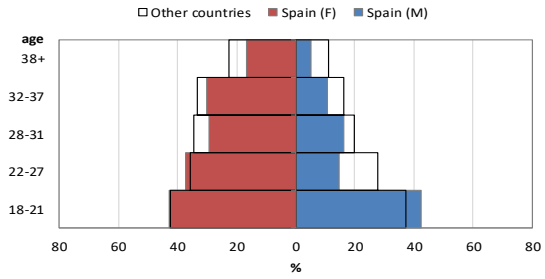
BMI<20 kg/m^2: The Netherlands



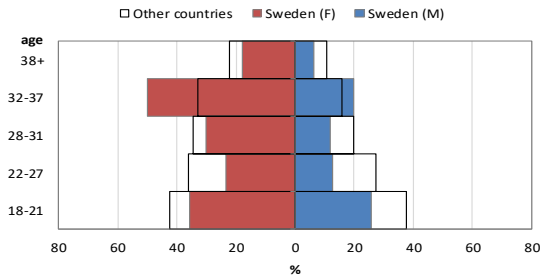
BMI<20 kg/m^2: Russian Federation



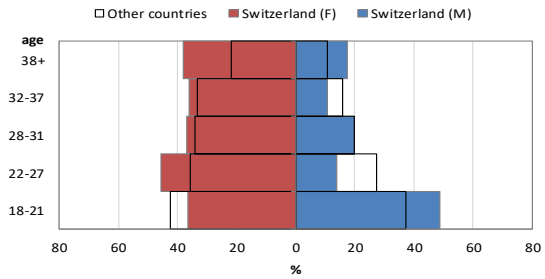
BMI<20 kg/m2: Spain



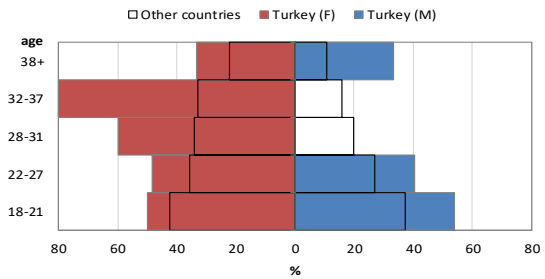
BMI<20 kg/m2: Sweden



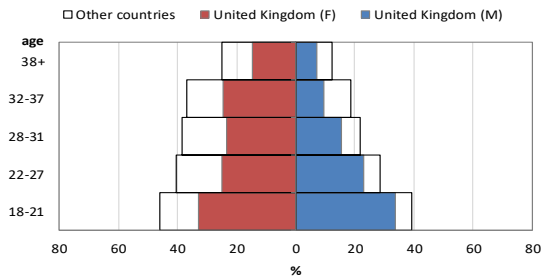
BMI<20 kg/m2: Switzerland



BMI<20 kg/m2: Turkey



BMI<20 kg/m2: United Kingdom



7. Complications and Therapy

The information in this section should not be considered complete, for several reasons: national registries may use a different definition or different parameters for a complication; data about one or more of the complications is not collected; the status of the complication is truly unknown (e.g. liver disease, where the definition requires ultrasound examination, is not always done for a patient). In the tables, therefore, we show the number of missing values for the various complications, but in the graphs we have included only countries where less than 10% of the data was missing. For a full list of complications and definitions please see Appendix 2 on page 168.

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”.

New information available from 2018

For the first time, information on a new class of drugs, the CFTR modulators is presented. These therapies target the function of the cystic fibrosis transmembrane conductance regulator (CFTR) protein.

Also, for the first time we present information about the status and availability of CF-specific pharmacotherapies for each country. The information should help to interpret country-specific differences in the use of therapies correctly.

Table 7.1 Prevalence of allergic bronchopulmonary aspergillosis (ABPA) (all patients seen in 2018 who never had a transplant) and CF-related diabetes (CFRD) in 2018 (patients aged 18 years or older seen in 2018 who never had a transplant), by country.

Country	ABPA this year number (%)			CFRD this year number (%)					
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes, treated with daily insulin	Yes, treated with oral hypogly-caemic agents	Yes, dietary advice only	Yes, therapy unknown
Albania	9 (8.11)	100 (90.09)	2 (1.80)	1 (20.00)	4 (80.00)	0 (0.00)	0 (0.00)	0 (0.00)	0 (0.00)
Armenia	0 (0)	28 (100)	0 (0)	-	-	-	-	-	-
Austria	8 (1.14)	673 (95.87)	21 (2.99)	2 (0.61)	243 (73.86)	71 (21.58)	1 (0.30)	12 (3.65)	0 (0.00)
Belarus	0 (0.00)	109 (98.20)	2 (1.80)	-	-	-	-	-	-
Belgium	6 (0.54)	1010 (90.75)	97 (8.72)	0 (0.00)	460 (80.00)	135 (20.83)	15 (2.31)	0 (0.00)	38 (5.86)
Bulgaria	0 (0.00)	185 (97.37)	5 (2.63)	0 (0.00)	69 (88.46)	8 (10.26)	0 (0.00)	1 (1.28)	0 (0.00)
Croatia	0 (0.00)	114 (98.28)	2 (1.72)	0 (0.00)	30 (78.95)	8 (21.05)	0 (0.00)	0 (0.00)	0 (0.00)
Cyprus	0 (0)	15 (100)	0 (0)	-	-	-	-	-	-
Czech Republic	1 (0.18)	554 (97.36)	14 (2.46)	0 (0.00)	179 (71.31)	72 (28.69)	0 (0.00)	0 (0.00)	0 (0.00)
Denmark	0 (0.00)	447 (96.54)	16 (3.46)	0 (0.00)	186 (70.99)	76 (29.01)	0 (0.00)	0 (0.00)	0 (0.00)
Georgia	3 (4.23)	68 (95.77)	0 (0.00)	-	-	-	-	-	-
Germany	57 (0.98)	5454 (93.47)	324 (5.55)	62 (1.89)	2251 (68.48)	710 (21.60)	51 (1.55)	31 (0.94)	182 (5.54)
Greece	1 (0.18)	536 (97.10)	15 (2.72)	1 (0.36)	212 (76.54)	63 (22.74)	0 (0.00)	0 (0.00)	1 (0.36)
Hungary	4 (0.91)	430 (98.17)	4 (0.91)	4 (2.17)	148 (80.44)	32 (17.39)	0 (0.00)	0 (0.00)	0 (0.00)
Ireland	6 (0.54)	1046 (93.64)	65 (5.82)	5 (0.82)	450 (73.77)	123 (20.16)	0 (0.00)	32 (5.25)	0 (0.00)
Israel	11 (2.12)	478 (92.10)	30 (5.78)	2 (0.64)	214 (68.36)	87 (27.80)	3 (0.96)	5 (1.60)	2 (0.64)
Italy	92 (1.77)	4958 (95.20)	158 (3.03)	42 (1.45)	2276 (78.43)	584 (20.12)	0 (0.00)	0 (0.00)	0 (0.00)
Latvia	0 (0)	37 (100)	0 (0)	0 (0.00)	8 (88.89)	1 (11.11)	0 (0.00)	0 (0.00)	0 (0.00)
Lithuania	0 (0)	24 (100)	0 (0)	0 (0)	15 (100)	0 (0)	0 (0)	0 (0)	0 (0)
Luxembourg	0 (0.00)	32 (88.89)	4 (11.11)	0 (0.00)	11 (61.11)	7 (38.89)	0 (0.00)	0 (0.00)	0 (0.00)

[table 7.1 continued]

Country	ABPA this year number (%)			CFRD this year number (%)					
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes, treated with daily insulin	Yes, treated with oral hypo-glycaemic agents	Yes, dietary advice only	Yes, therapy unknown
Rep of Moldova	0 (0)	56 (100)	0 (0)	0 (0.00)	11 (91.67)	1 (8.33)	0 (0.00)	0 (0.00)	0 (0.00)
The Netherlands[1]	13 (0.96)	1247 (92.51)	88 (6.53)	19 (2.34)	541 (66.71)	251 (30.95)	0 (0.00)	0 (0.00)	0 (0.00)
North Macedonia	4 (3.39)	113 (95.76)	1 (0.85)	2 (5.71)	22 (62.86)	11 (31.43)	0 (0.00)	0 (0.00)	0 (0.00)
Norway	6 (2.31)	249 (95.77)	5 (1.92)	0 (0.00)	130 (83.34)	23 (14.74)	0 (0.00)	3 (1.92)	0 (0.00)
Poland	14 (1.62)	833 (96.52)	16 (1.85)	18 (8.22)	159 (72.60)	32 (14.61)	0 (0.00)	10 (4.57)	0 (0.00)
Portugal	2 (0.72)	273 (97.85)	4 (1.43)	1 (0.79)	110 (86.62)	14 (11.02)	0 (0.00)	2 (1.57)	0 (0.00)
Romania	4 (2.01)	195 (97.99)	0 (0.00)	-	-	-	-	-	-
Russian Federation	58 (1.87)	2980 (96.28)	57 (1.84)	11 (1.49)	654 (88.37)	61 (8.24)	5 (0.68)	1 (0.14)	8 (1.08)
Serbia	1 (0.56)	176 (98.88)	1 (0.56)	0 (0.00)	31 (63.27)	18 (36.73)	0 (0.00)	0 (0.00)	0 (0.00)
Slovak Republic	3 (1.10)	249 (91.21)	21 (7.69)	3 (2.03)	131 (88.51)	13 (8.78)	0 (0.00)	1 (0.68)	0 (0.00)
Slovenia	0 (0.00)	94 (98.95)	1 (1.05)	0 (0.00)	25 (69.44)	9 (25.00)	0 (0.00)	2 (5.56)	0 (0.00)
Spain	24 (1.19)	1893 (94.18)	93 (4.63)	15 (1.61)	732 (78.54)	165 (17.70)	5 (0.54)	15 (1.61)	0 (0.00)
Sweden	0 (0.00)	593 (97.53)	15 (2.47)	0 (0.00)	264 (76.08)	60 (17.29)	8 (2.31)	15 (4.32)	0 (0.00)
Switzerland	6 (0.68)	834 (94.67)	41 (4.65)	2 (0.44)	319 (69.80)	106 (23.19)	7 (1.53)	19 (4.16)	4 (0.88)
Turkey	6 (0.33)	1754 (97.44)	40 (2.22)	11 (5.85)	162 (86.17)	8 (4.26)	0 (0.00)	7 (3.72)	0 (0.00)
Ukraine	5 (3.25)	149 (96.75)	0 (0.00)	3 (9.68)	27 (87.10)	1 (3.23)	0 (0.00)	0 (0.00)	0 (0.00)
United Kingdom[2]	1 (0.01)	8778 (92.52)	709 (7.47)	0 (0.00)	3607 (68.37)	1463 (27.73)	115 (2.18)	86 (1.63)	5 (0.09)
Total	345 (0.89)	36764 (94.36)	1851 (4.75)	204 (1.09)	13691 (72.82)	4213 (22.41)	210 (1.12)	242 (1.29)	240 (1.28)

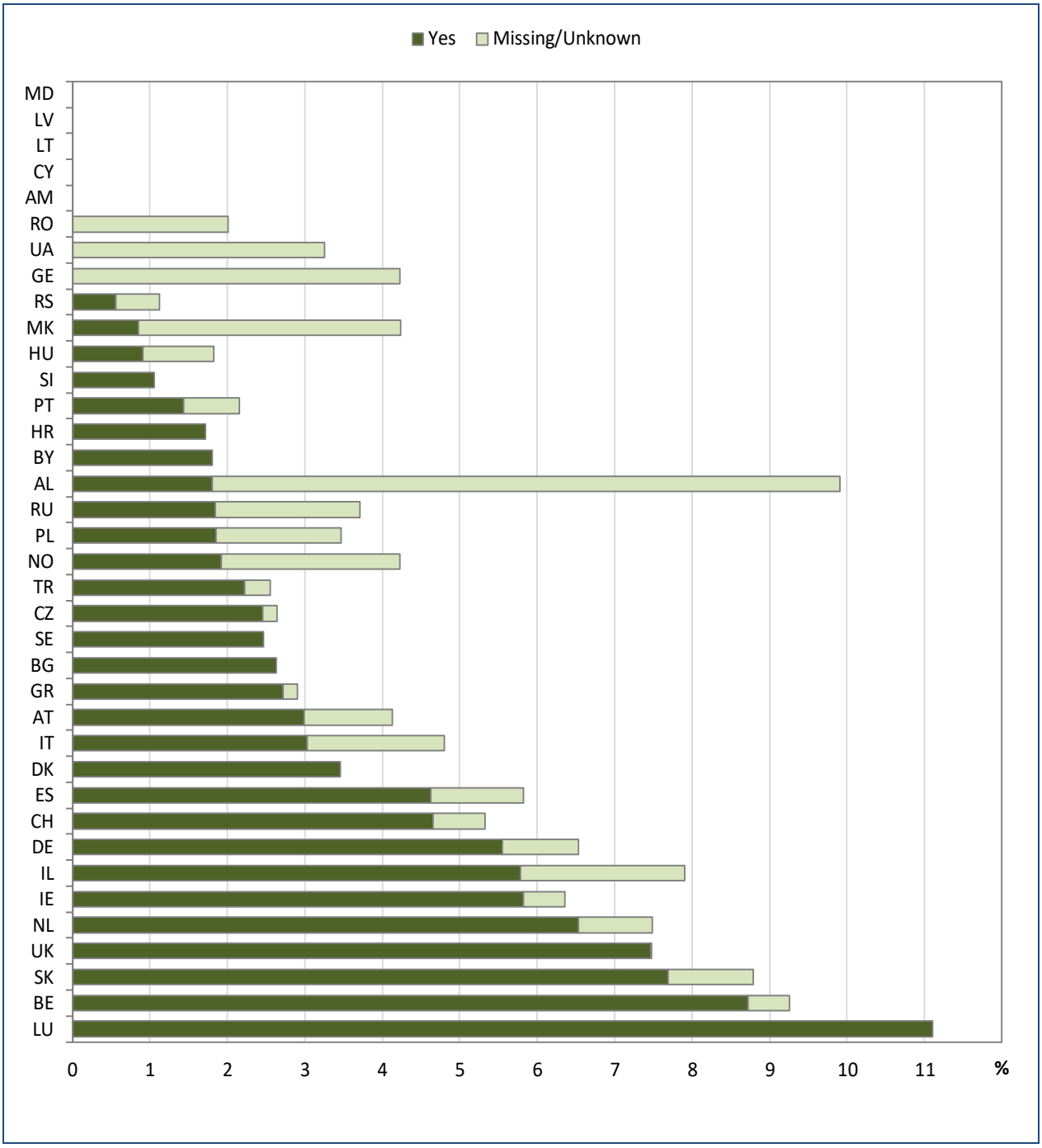
[1] The Netherlands: only diabetes treated with daily insulin is recorded.

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

Note: Belarus and Georgia have 0% coverage for adult patients and are excluded from the table.
Note: Armenia, Cyprus and Romania have <5 patients aged 18 years or more on 31/12/2018 and are not shown in the CFRD table, but the patients are included in the total.

Table 7.1 shows the frequency of allergic bronchopulmonary aspergillosis (see Appendix 2, page 168, for ABPA definitions) and CF-related diabetes (CFRD) by country. For CFRD only patients 18 years and older are included.

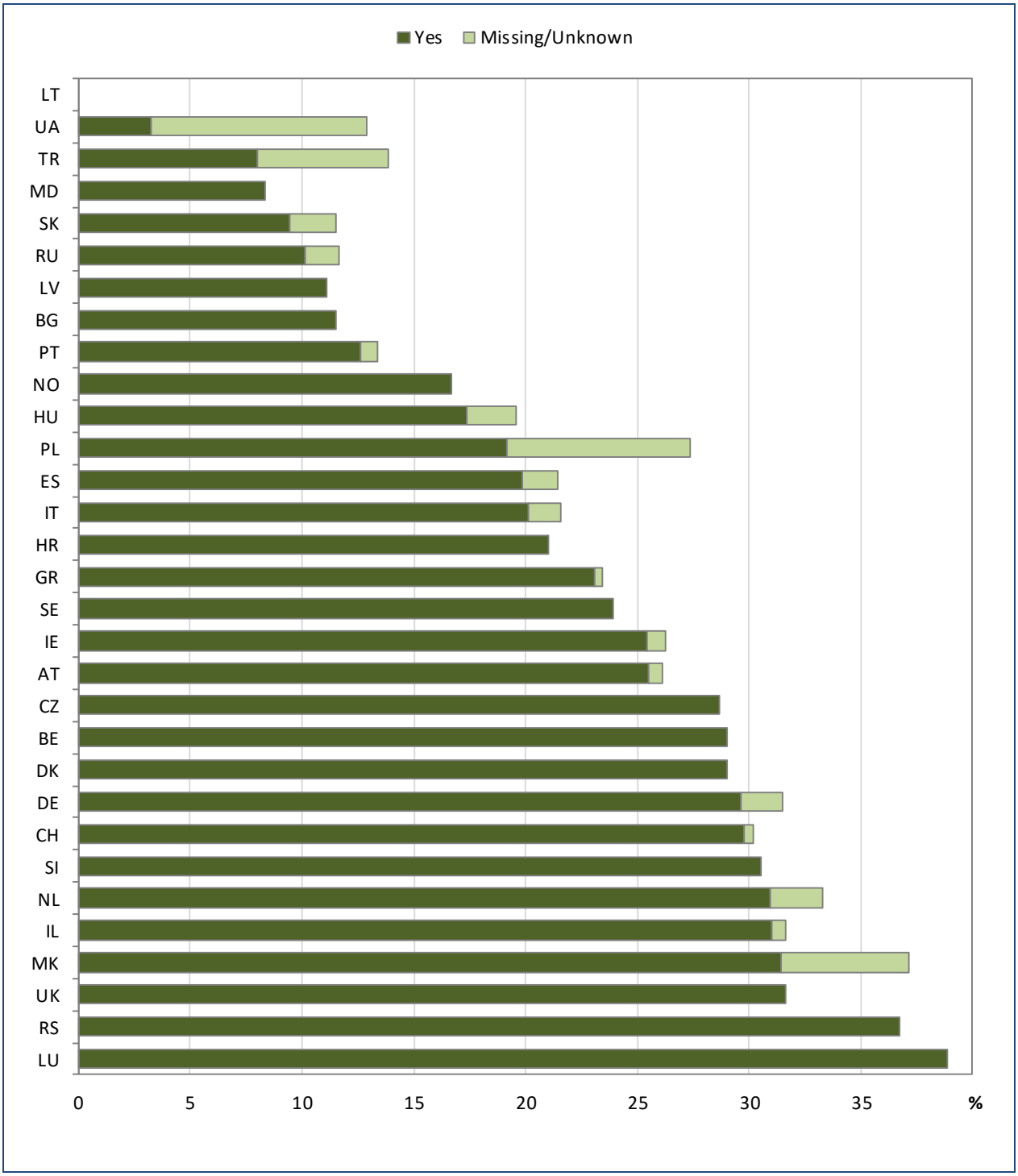
Figure 7.1 Prevalence of allergic bronchopulmonary aspergillosis in all patients seen in 2018 who have never had a transplant, by country.



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

This graph shows the frequency of allergic bronchopulmonary aspergillosis (ABPA) by country. For the definition of ABPA see Appendix 2 (page 168) the dark green part of the bar shows the percentage of patients with ABPA, the light green part shows the percentage of patients for which this information was missing.

Figure 7.2 Prevalence of CFRD, by country. All patients seen in 2018 aged 18 years or older who have never had a transplant.



Note: We excluded from the graph the countries for which the information on CFRD was missing for more than 10% of the adults patients.
Note: Belarus and Georgia have 0% coverage for adult patients and are excluded from the graph.
Note: Armenia, Cyprus and Romania have <5 patients aged 18 years or more on 31/12/2018, therefore no information is included in the graph.

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

Table 7.2 Prevalence of pneumothorax and haemoptysis major in all patients seen in 2018 who have never had a transplant, by country.

Country	Pneumothorax this year number (%)					Haemoptysis major over 250 ml this year number (%)		
	Missing/ unknown	No	Yes, chest drain	Yes, observation only	Yes, therapy unknown	Missing/ unknown	No	Yes
Albania	7 (6.31)	104 (93.69)	0 (0.00)	0 (0.00)	0 (0.00)	6 (5.41)	104 (93.69)	1 (0.90)
Armenia	0 (0.00)	26 (92.86)	0 (0.00)	2 (7.14)	0 (0.00)	0 (0)	28 (100)	0 (0)
Austria	3 (0.43)	697 (99.29)	1 (0.14)	1 (0.14)	0 (0.00)	8 (1.14)	682 (97.15)	12 (1.71)
Belarus	0 (0)	111 (100)	0 (0)	0 (0)	0 (0)	0 (0.00)	108 (97.30)	3 (2.70)
Belgium	219 (19.68)	890 (79.96)	0 (0.00)	0 (0.00)	4 (0.36)	219 (19.68)	884 (79.42)	10 (0.90)
Bulgaria	0 (0.00)	189 (99.47)	1 (0.53)	0 (0.00)	0 (0.00)	0 (0.00)	172 (90.53)	18 (9.47)
Croatia	0 (0.00)	115 (99.14)	1 (0.86)	0 (0.00)	0 (0.00)	0 (0.00)	112 (96.55)	4 (3.45)
Cyprus	0 (0)	15 (100)	0 (0)	0 (0)	0 (0)	0 (0)	15 (100)	0 (0)
Czech Republic	3 (0.53)	564 (99.12)	2 (0.35)	0 (0.00)	0 (0.00)	7 (1.23)	554 (97.36)	8 (1.41)
Denmark	0 (0.00)	462 (99.78)	1 (0.22)	0 (0.00)	0 (0.00)	0 (0.00)	461 (99.57)	2 (0.43)
Georgia	3 (4.23)	68 (95.77)	0 (0.00)	0 (0.00)	0 (0.00)	4 (5.63)	65 (91.55)	2 (2.82)
Germany[1]	47 (0.81)	5743 (98.42)	22 (0.38)	14 (0.24)	9 (0.15)	322 (5.52)	5509 (94.41)	4 (0.07)
Greece	1 (0.18)	546 (98.92)	3 (0.54)	1 (0.18)	1 (0.18)	1 (0.18)	543 (98.37)	8 (1.45)
Hungary	5 (1.14)	431 (98.40)	2 (0.46)	0 (0.00)	0 (0.00)	15 (3.42)	411 (93.84)	12 (2.74)
Ireland	6 (0.54)	1110 (99.37)	1 (0.09)	0 (0.00)	0 (0.00)	7 (0.63)	1110 (99.37)	0 (0.00)
Israel	9 (1.73)	510 (98.27)	0 (0.00)	0 (0.00)	0 (0.00)	9 (1.73)	505 (97.30)	5 (0.96)
Italy[2]	91 (1.75)	5108 (98.08)	9 (0.17)	0 (0.00)	0 (0.00)	716 (13.75)	4262 (81.84)	230 (4.42)
Latvia	0 (0)	37 (100)	0 (0)	0 (0)	0 (0)	0 (0)	37 (100)	0 (0)
Lithuania	1 (4.17)	23 (95.83)	0 (0.00)	0 (0.00)	0 (0.00)	0 (0.00)	23 (95.83)	1 (4.17)
Luxembourg	0 (0)	36 (100)	0 (0)	0 (0)	0 (0)	0 (0)	36 (100)	0 (0)
Rep of Moldova	0 (0)	56 (100)	0 (0)	0 (0)	0 (0)	0 (0.00)	55 (98.21)	1 (1.79)
The Netherlands	106 (7.86)	1240 (91.99)	2 (0.15)	0 (0.00)	0 (0.00)	307 (22.77)	1001 (74.26)	40 (2.97)

[1] Germany: defines haemoptysis major over 240 ml.
[2] Italy: one centre did not provide data on haemoptysis.

[table 7.2 continued]

Country	Pneumothorax this year number (%)					Haemoptysis major over 250 ml this year number (%)		
	Missing/ unknown	No	Yes, chest drain	Yes, observation only	Yes, therapy unknown	Missing/ unknown	No	Yes
North Macedonia	2 (1.69)	116 (98.31)	0 (0.00)	0 (0.00)	0 (0.00)	2 (1.69)	115 (97.46)	1 (0.85)
Norway	2 (0.77)	254 (97.70)	3 (1.15)	1 (0.38)	0 (0.00)	4 (1.54)	254 (97.69)	2 (0.77)
Poland	17 (1.97)	843 (97.68)	1 (0.12)	2 (0.23)	0 (0.00)	21 (2.43)	803 (93.05)	39 (4.52)
Portugal	6 (2.15)	270 (96.77)	3 (1.08)	0 (0.00)	0 (0.00)	3 (1.08)	262 (93.91)	14 (5.02)
Romania	3 (1.51)	196 (98.49)	0 (0.00)	0 (0.00)	0 (0.00)	3 (1.51)	194 (97.49)	2 (1.01)
Russian Federation	16 (0.52)	3062 (98.93)	16 (0.52)	1 (0.03)	0 (0.00)	53 (1.71)	3016 (97.45)	26 (0.84)
Serbia	1 (0.56)	177 (99.44)	0 (0.00)	0 (0.00)	0 (0.00)	1 (0.56)	170 (95.51)	7 (3.93)
Slovak Republic	4 (1.47)	266 (97.43)	2 (0.73)	1 (0.37)	0 (0.00)	4 (1.47)	247 (90.48)	22 (8.06)
Slovenia	0 (0)	95 (100)	0 (0)	0 (0)	0 (0)	0 (0.00)	93 (97.89)	2 (2.11)
Spain	35 (1.74)	1969 (97.96)	5 (0.25)	1 (0.05)	0 (0.00)	31 (1.54)	1893 (94.18)	86 (4.28)
Sweden	0 (0)	608 (100)	0 (0)	0 (0)	0 (0)	0 (0.00)	605 (99.51)	3 (0.49)
Switzerland	8 (0.91)	869 (98.64)	4 (0.45)	0 (0.00)	0 (0.00)	6 (0.68)	856 (97.16)	19 (2.16)
Turkey	6 (0.33)	1793 (99.61)	1 (0.06)	0 (0.00)	0 (0.00)	7 (0.39)	1781 (98.94)	12 (0.67)
Ukraine[3]	6 (3.90)	145 (94.15)	1 (0.65)	2 (1.30)	0 (0.00)	7 (4.55)	127 (82.47)	20 (12.99)
United Kingdom	1 (0.01)	9451 (99.61)	36 (0.38)	0 (0.00)	0 (0.00)	0 (0.00)	9468 (99.79)	20 (0.21)
Total	608 (1.56)	38195 (98.04)	117 (0.30)	26 (0.07)	14 (0.04)	1763 (4.53)	36561 (93.84)	636 (1.63)

[3] Ukraine: considered also patients with haemoptysis <250ml.

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

Table 7.3 Prevalence of liver disease in all patients seen in 2018 who have never had a transplant, by country.

Country	Liver disease this year number (%)						
	Missing/ unknown	No liver disease	Cirrhosis			Liver disease without cirrhosis	Variceal bleeding
			Cirrhosis with portal hypertension/ hypersplenism	Cirrhosis no portal hypertension/ hypersplenism	Cirrhosis. portal hypertension unknown		
Albania	6 (5.41)	59 (53.15)	2 (1.80)	0 (0.00)	0 (0.00)	44 (39.64)	0 (0.00)
Armenia	0 (0.00)	16 (57.14)	0 (0.00)	1 (3.57)	0 (0.00)	11 (39.29)	0 (0.00)
Austria	3 (0.43)	384 (54.70)	18 (2.56)	11 (1.57)	3 (0.43)	283 (40.31)	0 (0.00)
Belarus	0 (0.00)	74 (66.67)	4 (3.60)	2 (1.80)	0 (0.00)	31 (27.93)	0 (0.00)
Belgium[1]	4 (0.36)	1063 (95.51)	46 (4.13)	0 (0.00)	0 (0.00)	0 (0.00)	0 (0.00)
Bulgaria	0 (0.00)	140 (73.68)	10 (5.26)	1 (0.53)	0 (0.00)	39 (20.53)	0 (0.00)
Croatia	1 (0.86)	100 (86.21)	4 (3.45)	1 (0.86)	0 (0.00)	10 (8.62)	0 (0.00)
Cyprus	0 (0)	15 (100)	0 (0)	0 (0)	0 (0)	0 (0)	0 (0)
Czech Republic	16 (2.81)	456 (80.14)	5 (0.88)	6 (1.05)	1 (0.18)	85 (14.94)	0 (0.00)
Denmark	0 (0.00)	385 (83.15)	19 (4.10)	7 (1.51)	3 (0.65)	49 (10.58)	0 (0.00)
Georgia	3 (4.23)	64 (90.14)	0 (0.00)	2 (2.82)	0 (0.00)	2 (2.82)	0 (0.00)
Germany[2]	405 (6.94)	4068 (69.72)	123 (2.11)	83 (1.42)	62 (1.06)	1094 (18.75)	0 (0.00)
Greece	4 (0.72)	396 (71.74)	8 (1.45)	6 (1.09)	2 (0.36)	136 (24.64)	0 (0.00)
Hungary	7 (1.60)	328 (74.89)	68 (15.53)	16 (3.65)	15 (3.42)	4 (0.91)	0 (0.00)
Ireland[3]	6 (0.54)	968 (86.66)	30 (2.69)	8 (0.72)	10 (0.90)	95 (8.50)	0 (0.00)
Israel	12 (2.31)	417 (80.35)	13 (2.50)	3 (0.58)	0 (0.00)	74 (14.26)	0 (0.00)
Italy	62 (1.19)	3745 (71.91)	68 (1.31)	44 (0.84)	1 (0.02)	1288 (24.73)	0 (0.00)
Latvia	0 (0.00)	23 (62.16)	2 (5.41)	0 (0.00)	0 (0.00)	12 (32.43)	0 (0.00)
Lithuania	1 (4.17)	23 (95.83)	0 (0.00)	0 (0.00)	0 (0.00)	0 (0.00)	0 (0.00)
Luxembourg	0 (0.00)	29 (80.56)	2 (5.56)	0 (0.00)	0 (0.00)	4 (11.11)	1 (2.78)
Rep of Moldova	0 (0.00)	51 (91.07)	0 (0.00)	0 (0.00)	0 (0.00)	5 (8.93)	0 (0.00)
The Netherlands[2]	150 (11.13)	943 (69.96)	65 (4.82)	28 (2.08)	4 (0.30)	158 (11.72)	0 (0.00)
North Macedonia	2 (1.69)	58 (49.15)	5 (4.24)	11 (9.32)	2 (1.69)	40 (33.90)	0 (0.00)

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

[2] Germany and The Netherlands: variceal bleeding information was not reported.

[table 7.3 continued]

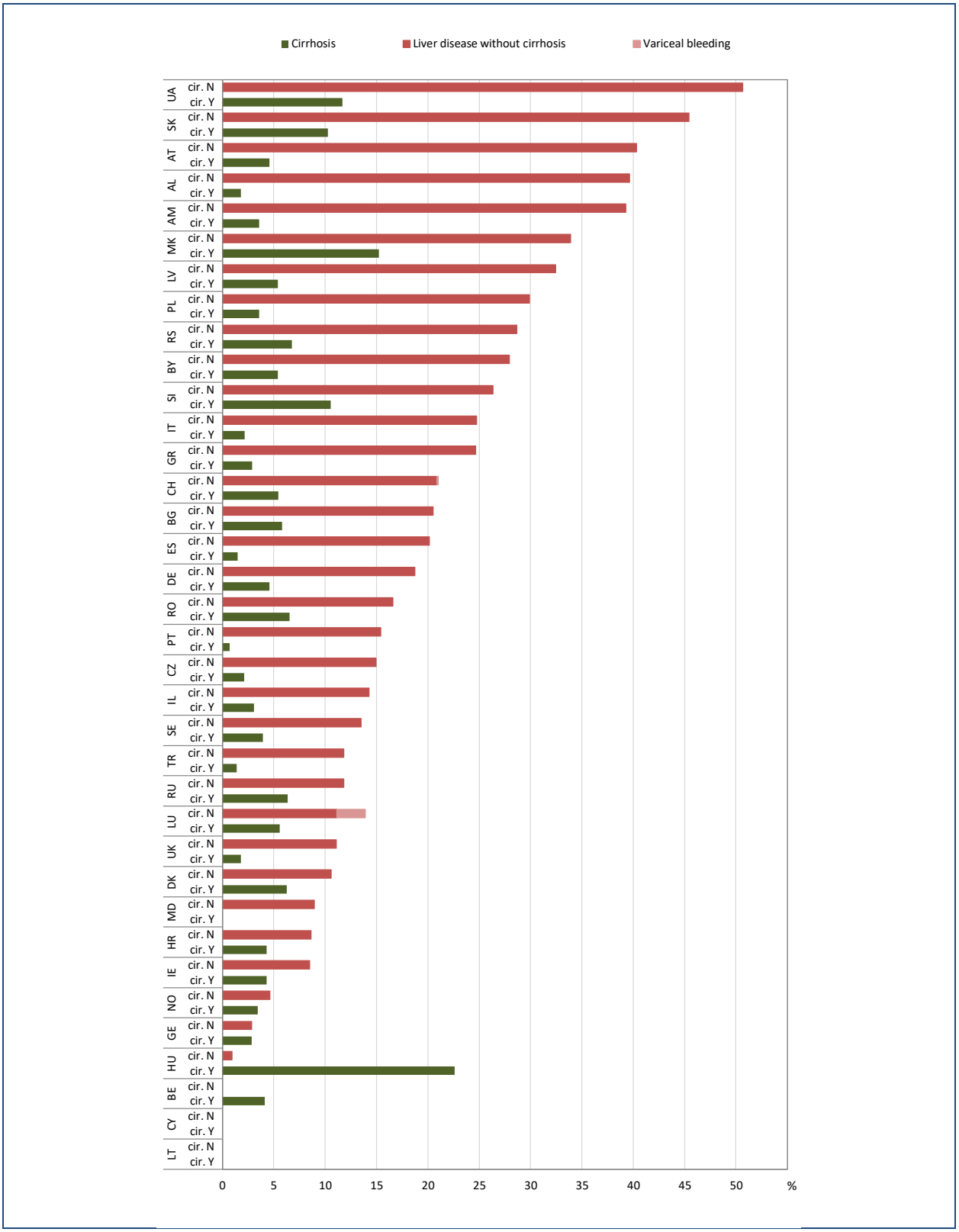
Country	Liver disease this year number (%)						
	Missing/ unknown	No liver disease	Cirrhosis			Liver disease without cirrhosis	Variceal bleeding
			Cirrhosis with portal hypertension/ hypersplenism	Cirrhosis no portal hypertension/ hypersplenism	Cirrhosis. portal hypertension unknown		
Norway	1 (0.38)	238 (91.54)	5 (1.92)	3 (1.15)	1 (0.38)	12 (4.62)	0 (0.00)
Poland	14 (1.62)	560 (64.89)	22 (2.55)	4 (0.46)	5 (0.58)	258 (29.90)	0 (0.00)
Portugal	4 (1.43)	230 (82.44)	1 (0.36)	0 (0.00)	1 (0.36)	43 (15.41)	0 (0.00)
Romania	3 (1.51)	150 (75.38)	6 (3.02)	4 (2.01)	3 (1.51)	33 (16.58)	0 (0.00)
Russian Federation	31 (1.00)	2503 (80.87)	116 (3.75)	68 (2.20)	12 (0.39)	365 (11.79)	0 (0.00)
Serbia[3]	1 (0.56)	114 (64.04)	7 (3.93)	4 (2.25)	1 (0.56)	51 (28.65)	0 (0.00)
Slovak Republic	2 (0.73)	119 (43.59)	13 (4.76)	15 (5.49)	0 (0.00)	124 (45.42)	0 (0.00)
Slovenia	0 (0.00)	60 (63.16)	4 (4.21)	4 (4.21)	2 (2.11)	25 (26.32)	0 (0.00)
Spain	27 (1.34)	1548 (77.01)	22 (1.09)	6 (0.30)	2 (0.10)	405 (20.15)	0 (0.00)
Sweden[3]	0 (0.00)	502 (82.57)	12 (1.97)	7 (1.15)	5 (0.82)	82 (13.49)	0 (0.00)
Switzerland	13 (1.48)	635 (72.08)	31 (3.52)	16 (1.82)	1 (0.11)	184 (20.89)	1 (0.11)
Turkey	6 (0.33)	1556 (86.44)	10 (0.56)	12 (0.67)	3 (0.17)	213 (11.83)	0 (0.00)
Ukraine	5 (3.25)	53 (34.42)	10 (6.49)	7 (4.55)	1 (0.65)	78 (50.65)	0 (0.00)
United Kingdom	0 (0.00)	8265 (87.11)	100 (1.05)	70 (0.74)	0 (0.00)	1053 (11.10)	0 (0.00)
Total	789 (2.03)	30338 (77.87)	851 (2.18)	450 (1.16)	140 (0.36)	6390 (16.40)	2 (0.01)

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

This table shows the frequency and severity of liver disease according to the ECFSPR definitions (see Appendix 2, page 168). The frequency and severity of liver disease differs greatly throughout the ECFSPR data.

The following graph (figure 7.3) shows the frequency of liver disease by country. Liver disease is defined according to severity of portal hypertension (increased blood pressure in the liver veins, often resulting in blood shunting past the cirrhotic liver) divided into five categories, including no liver disease (see Appendix 2, page 168). 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 7.3 Prevalence and severity of liver disease in all patients seen in 2018 who have never had a transplant, 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.

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

Table 7.4 Prevalence of the use of ursodeoxycholic acid in all patients seen in 2018 who have never had a transplant, by country.

Country	Ursodeoxycholic acid this year number (%)		
	Missing/ unknown	No	Yes
Albania	5 (4.50)	59 (53.15)	47 (42.34)
Armenia	1 (3.57)	16 (57.14)	11 (39.29)
Austria	3 (0.43)	367 (52.28)	332 (47.29)
Belarus	0 (0.00)	7 (6.31)	104 (93.69)
Belgium	4 (0.36)	891 (80.05)	218 (19.59)
Bulgaria	0 (0.00)	138 (72.63)	52 (27.37)
Croatia	0 (0.00)	74 (63.79)	42 (36.21)
Cyprus	0 (0.00)	14 (93.33)	1 (6.67)
Czech Republic	0 (0.00)	390 (68.54)	179 (31.46)
Denmark	0 (0.00)	325 (70.19)	138 (29.81)
Georgia	3 (4.23)	57 (80.28)	11 (15.49)
Germany	57 (0.98)	2947 (50.51)	2831 (48.52)
Greece	4 (0.72)	386 (69.93)	162 (29.35)
Hungary	18 (4.11)	244 (55.71)	176 (40.18)
Ireland[3]	6 (0.54)	1016 (90.96)	95 (8.50)
Israel	6 (1.16)	431 (83.04)	82 (15.80)
Italy	15 (0.29)	3467 (66.57)	1726 (33.14)
Latvia	0 (0.00)	25 (67.57)	12 (32.43)
Lithuania	0 (0.00)	18 (75.00)	6 (25.00)
Luxembourg	0 (0.00)	20 (55.56)	16 (44.44)
Rep of Moldova	3 (5.36)	45 (80.36)	8 (14.29)
The Netherlands	153 (11.35)	883 (65.50)	312 (23.15)
North Macedonia	1 (0.85)	62 (52.54)	55 (46.61)
Norway	23 (8.85)	223 (85.77)	14 (5.38)

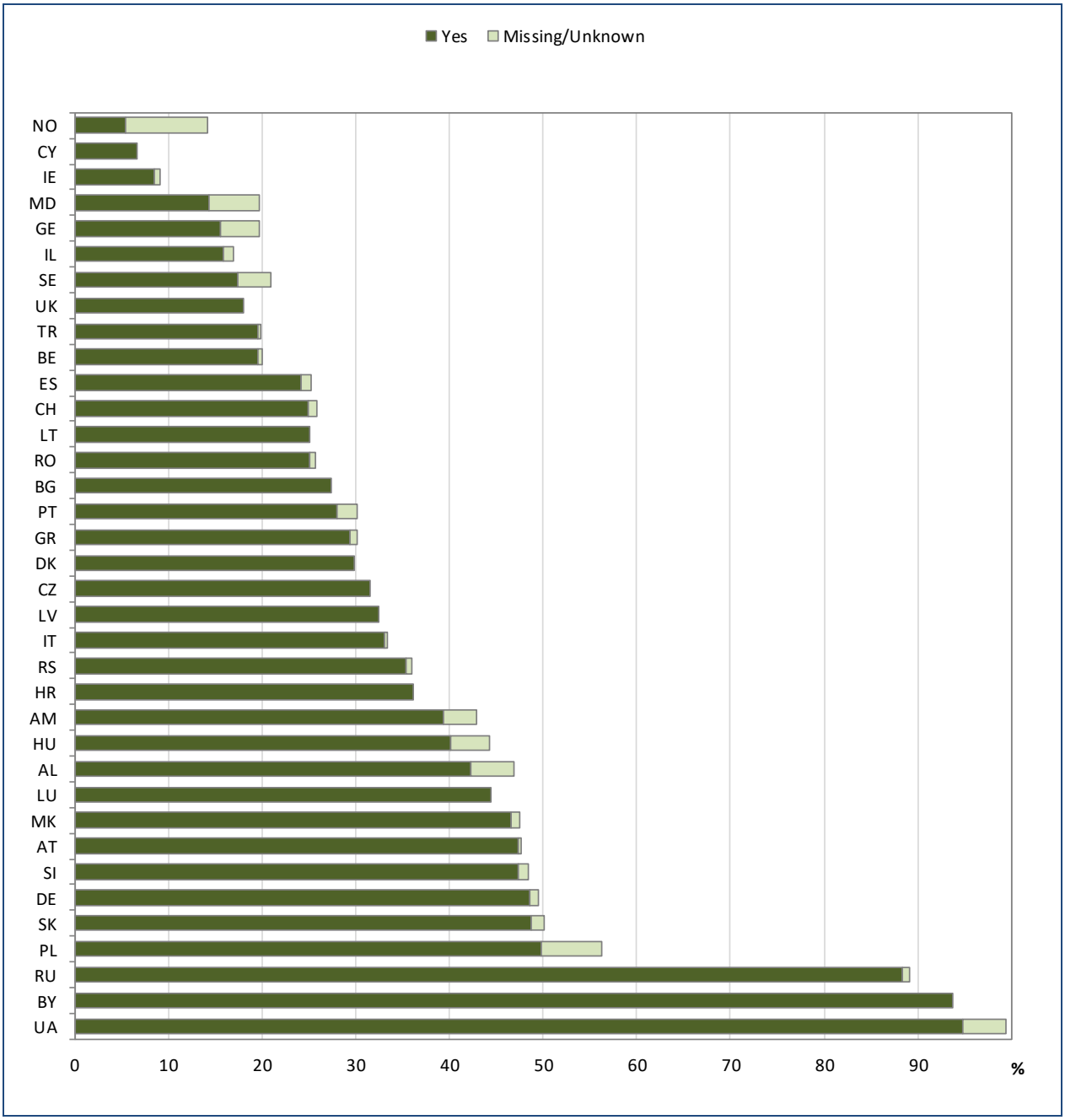
[table 7.4 continued]

Country	Ursodeoxycholic acid this year number (%)		
	Missing/ unknown	No	Yes
Poland	55 (6.37)	378 (43.80)	430 (49.83)
Portugal	6 (2.15)	195 (69.89)	78 (27.96)
Romania	1 (0.50)	148 (74.37)	50 (25.13)
Russian Federation	24 (0.78)	338 (10.92)	2733 (88.30)
Serbia	1 (0.56)	114 (64.04)	63 (35.39)
Slovak Republic	4 (1.47)	136 (49.82)	133 (48.72)
Slovenia	1 (1.05)	49 (51.58)	45 (47.37)
Spain	23 (1.14)	1502 (74.73)	485 (24.13)
Sweden	21 (3.45)	481 (79.11)	106 (17.43)
Switzerland	9 (1.02)	653 (74.12)	219 (24.86)
Turkey	6 (0.33)	1443 (80.17)	351 (19.50)
Ukraine	7 (4.55)	1 (0.65)	146 (94.81)
United Kingdom	0 (0.00)	7780 (82.00)	1708 (18.00)
Total	460 (1.18)	25323 (65.00)	13177 (33.82)

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

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

Figure 7.4 Use of ursodeoxycholic acid in all patients seen in 2018 who have never had a transplant, 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.

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

This graph shows how many patients used ursodeoxycholic acid during 2018. Ursodeoxycholic acid is used as a treatment for CF liver disease. The dark green part of the bar indicates the percentage of patients taking this drug, the light green part shows the percentage of patients for whom this information is missing.

Table 7.5 Occurrence of malignancy this year and prevalence of distal intestinal obstruction syndrome (DIOS) in all patients seen in 2018 who have never had a transplant, by country.

Country	Malignancy occurred this year number(%)			DIOS this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	6 (5.41)	105 (94.59)	0 (0.00)	5 (4.50)	103 (92.79)	3 (2.70)
Armenia	0 (0)	28 (100)	0 (0)	0 (0.00)	24 (85.71)	4 (14.29)
Austria	12 (1.71)	689 (98.15)	1 (0.14)	2 (0.28)	677 (96.44)	23 (3.28)
Belarus	0 (0)	111 (100)	0 (0)	111 (100)	0 -	0 -
Belgium	0 (0.00)	1110 (99.73)	3 (0.27)	216 (19.41)	865 (77.72)	32 (2.88)
Bulgaria	0 (0.00)	189 (99.47)	1 (0.53)	0 (0.00)	189 (99.47)	1 (0.53)
Croatia	1 (0.86)	115 (99.14)	0 (0.00)	0 (0.00)	110 (94.83)	6 (5.17)
Cyprus	0 (0)	15 (100)	0 (0)	0 (0)	15 (100)	0 (0)
Czech Republic	0 (0)	569 (100)	0 (0)	0 (0.00)	565 (99.30)	4 (0.70)
Denmark	0 (0.00)	462 (99.78)	1 (0.22)	463 (100)	0 -	0 -
Georgia	7 (9.86)	64 (90.14)	0 (0.00)	4 (5.63)	67 (94.37)	0 (0.00)
Germany	81 (1.39)	5707 (97.81)	47 (0.81)	85 (1.46)	5469 (93.73)	281 (4.82)
Greece	2 (0.36)	548 (99.28)	2 (0.36)	2 (0.36)	538 (97.46)	12 (2.17)
Hungary	6 (1.37)	431 (98.40)	1 (0.23)	438 (100)	0 -	0 -
Ireland	6 (0.54)	1109 (99.28)	2 (0.18)	12 (1.07)	1097 (98.21)	8 (0.72)
Israel	9 (1.73)	508 (97.88)	2 (0.39)	10 (1.93)	496 (95.57)	13 (2.50)
Italy	745 (14.30)	4441 (85.27)	22 (0.42)	630 (12.10)	4509 (86.58)	69 (1.32)
Latvia	1 (2.70)	36 (97.30)	0 (0.00)	0 (0)	37 (100)	0 (0)
Lithuania	0 (0)	24 (100)	0 (0)	0 (0)	24 (100)	0 (0)
Luxembourg	0 (0.00)	35 (97.22)	1 (2.78)	0 (0)	36 (100)	0 (0)
Rep of Moldova	0 (0)	56 (100)	0 (0)	0 (0)	56 (100)	0 (0)
The Netherlands	29 (2.15)	1315 (97.55)	4 (0.30)	144 (10.68)	1148 (85.16)	56 (4.15)

[table 7.5 continued]

Country	Malignancy occurred this year number(%)			DIOS this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
North Macedonia	2 (1.69)	116 (98.31)	0 (0.00)	2 (1.69)	113 (95.76)	3 (2.54)
Norway	4 (1.54)	256 (98.46)	0 (0.00)	16 (6.15)	235 (90.38)	9 (3.46)
Poland	12 (1.39)	851 (98.61)	0 (0.00)	12 (1.39)	846 (98.03)	5 (0.58)
Portugal	3 (1.08)	276 (98.92)	0 (0.00)	2 (0.72)	274 (98.21)	3 (1.08)
Romania	3 (1.51)	195 (97.99)	1 (0.50)	4 (2.01)	187 (93.97)	8 (4.02)
Russian Federation	16 (0.52)	3073 (99.29)	6 (0.19)	66 (2.13)	2976 (96.16)	53 (1.71)
Serbia	1 (0.56)	177 (99.44)	0 (0)	1 (0.56)	173 (97.19)	4 (2.25)
Slovak Republic	3 (1.10)	270 (98.90)	0 (0)	7 (2.56)	265 (97.07)	1 (0.37)
Slovenia	0 (0)	95 (100)	0 (0)	0 (0.00)	93 (97.89)	2 (2.11)
Spain	23 (1.14)	1979 (98.46)	8 (0.40)	21 (1.04)	1955 (97.26)	34 (1.69)
Sweden	0 (0.00)	605 (99.51)	3 (0.49)	0 (0.00)	589 (96.88)	19 (3.13)
Switzerland	7 (0.79)	871 (98.86)	3 (0.33)	9 (1.02)	843 (95.69)	29 (3.29)
Turkey	6 (0.33)	1794 (99.67)	0 (0.00)	1464 (81.33)	336 (18.67)	0 (0.00)
Ukraine	5 (3.25)	149 (96.75)	0 (0.00)	5 (3.25)	148 (96.10)	1 (0.65)
United Kingdom	7 (0.07)	9461 (99.72)	20 (0.21)	1 (0.01)	9010 (94.96)	477 (5.03)
Total	997 (2.56)	37835 (97.11)	128 (0.33)	3732 (9.58)	34068 (87.44)	1160 (2.98)

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

Table 7.6 Use of hypertonic saline, rhDNase and inhaled mannitol in all patients seen in 2018 who have never had a transplant, by country.

Country	Hypertonic saline (NaCl) inhaled > 3 months this year number (%)			rhDNase inhaled > 3 months this year number (%)			Mannitol inhaled > 3 months this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	2 (1.80)	5 (4.50)	104 (93.69)	2 (1.80)	103 (92.79)	6 (5.41)	6 (5.41)	104 (93.69)	1 (0.90)
Armenia	0 (0.00)	1 (3.57)	27 (96.43)	1 (3.57)	22 (78.57)	5 (17.86)	0 (0)	28 (100)	0 (0)
Austria	4 (0.57)	188 (26.78)	510 (72.65)	5 (0.71)	336 (47.86)	361 (51.42)	4 (0.57)	690 (98.29)	8 (1.14)
Belarus	0 (0.00)	56 (50.45)	55 (49.55)	0 (0.00)	81 (72.97)	30 (27.03)	0 (0)	111 (100)	0 (0)
Belgium	42 (3.77)	326 (29.29)	745 (66.94)	42 (3.77)	130 (11.68)	941 (84.55)	1113 (100)	0 -	0 -
Bulgaria	1 (0.53)	76 (40.00)	113 (59.47)	0 (0.00)	39 (20.53)	151 (79.47)	0 (0.00)	189 (99.47)	1 (0.53)
Croatia	1 (0.86)	11 (9.48)	104 (89.66)	1 (0.86)	26 (22.41)	89 (76.72)	1 (0.86)	114 (98.28)	1 (0.86)
Cyprus	0 (0.00)	12 (80.00)	3 (20.00)	0 (0.00)	6 (40.00)	9 (60.00)	0 (0)	15 (100)	0 (0)
Czech Republic	0 (0.00)	109 (19.16)	460 (80.84)	0 (0.00)	187 (32.86)	382 (67.14)	0 (0.00)	564 (99.12)	5 (0.88)
Denmark	463 (100)	0 -	0 -	0 (0.00)	51 (11.02)	412 (88.98)	463 (100)	0 -	0 -
Georgia	1 (1.41)	51 (71.83)	19 (26.76)	2 (2.82)	69 (97.18)	0 (0.00)	1 (1.41)	70 (98.59)	0 (0.00)
Germany	65 (1.11)	1109 (19.01)	4661 (79.88)	59 (1.01)	2852 (48.88)	2924 (50.11)	63 (1.08)	5504 (94.33)	268 (4.59)
Greece	1 (0.18)	354 (64.13)	197 (35.69)	2 (0.36)	136 (24.64)	414 (75.00)	2 (0.36)	536 (97.10)	14 (2.54)
Hungary	10 (2.28)	75 (17.12)	353 (80.59)	18 (4.11)	125 (28.54)	295 (67.35)	438 (100)	0 -	0 -
Ireland	6 (0.54)	464 (41.54)	647 (57.92)	6 (0.54)	474 (42.44)	637 (57.03)	1117 (100)	0 -	0 -
Israel	3 (0.58)	124 (23.89)	392 (75.53)	4 (0.77)	131 (25.24)	384 (73.99)	3 (0.58)	514 (99.04)	2 (0.39)
Italy	17 (0.33)	2953 (56.70)	2238 (42.97)	17 (0.33)	3338 (64.09)	1853 (35.58)	4560 (87.56)	636 (12.21)	12 (0.23)
Latvia	0 (0.00)	3 (8.11)	34 (91.89)	0 (0.00)	19 (51.35)	18 (48.65)	0 (0)	37 (100)	0 (0)
Lithuania	0 (0.00)	19 (79.17)	5 (20.83)	0 (0.00)	6 (25.00)	18 (75.00)	1 (4.17)	23 (95.83)	0 (0.00)
Luxembourg	1 (2.78)	8 (22.22)	27 (75.00)	1 (2.78)	12 (33.33)	23 (63.89)	0 (0.00)	35 (97.22)	1 (2.78)
Rep of Moldova	3 (5.36)	7 (12.50)	46 (82.14)	0 (0)	56 (100)	0 (0)	3 (5.36)	53 (94.64)	0 (0.00)
The Netherlands	156 (11.57)	748 (55.49)	444 (32.94)	150 (11.13)	391 (29.01)	807 (59.87)	1348 (100)	0 -	0 -

[table 7.6 continued]

Country	Hypertonic saline (NaCl) inhaled > 3 months this year number (%)			rhDNase inhaled > 3 months this year number (%)			Mannitol inhaled > 3 months this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
North Macedonia	1 (0.85)	40 (33.90)	77 (65.25)	1 (0.85)	31 (26.27)	86 (72.88)	1 (0.85)	116 (98.31)	1 (0.85)
Norway	4 (1.54)	73 (28.08)	183 (70.38)	4 (1.54)	109 (41.92)	147 (56.54)	10 (3.85)	250 (96.15)	0 (0.00)
Poland	21 (2.43)	162 (18.77)	680 (78.79)	16 (1.85)	164 (19.00)	683 (79.14)	53 (6.14)	810 (93.86)	0 (0.00)
Portugal	5 (1.79)	189 (67.74)	85 (30.47)	5 (1.79)	51 (18.28)	223 (79.93)	5 (1.79)	274 (98.21)	0 (0.00)
Romania	3 (1.51)	84 (42.21)	112 (56.28)	2 (1.01)	40 (20.10)	157 (78.89)	2 (1.01)	197 (98.99)	0 (0.00)
Russian Federation	39 (1.26)	979 (31.63)	2077 (67.11)	19 (0.61)	116 (3.75)	2960 (95.64)	49 (1.58)	3006 (97.12)	40 (1.29)
Serbia	1 (0.56)	8 (4.49)	169 (94.94)	1 (0.56)	52 (29.21)	125 (70.22)	1 (0.56)	176 (98.88)	1 (0.56)
Slovak Republic	2 (0.73)	189 (69.23)	82 (30.04)	2 (0.73)	94 (34.43)	177 (64.84)	3 (1.10)	270 (98.90)	0 (0.00)
Slovenia	0 (0.00)	2 (2.11)	93 (97.89)	0 (0.00)	63 (66.32)	32 (33.68)	1 (1.05)	93 (97.89	1 (1.05))
Spain	17 (0.85)	721 (35.87)	1272 (63.28)	18 (0.90)	1221 (60.75)	771 (38.36)	21 (1.04)	1981 (98.56)	8 (0.40)
Sweden	19 (3.13)	107 (17.60)	482 (79.28)	22 (3.62)	408 (67.11)	178 (29.28)	608 (100)	0 -	0 -
Switzerland	4 (0.45)	220 (24.97)	657 (74.57)	5 (0.57)	458 (51.99)	418 (47.45)	5 (0.57)	875 (99.32)	1 (0.11)
Turkey	7 (0.39)	1584 (88.00)	209 (11.61)	6 (0.33)	194 (10.78)	1600 (88.89)	7 (0.39)	1707 (94.83)	86 (4.78)
Ukraine	7 (4.55)	3 (1.95)	144 (93.51)	6 (3.90)	37 (24.03)	111 (72.08)	5 (3.25)	149 (96.75)	0 (0.00)
United Kingdom[1]	0 (0.00)	6311 (66.52)	3177 (33.48)	0 (0.00)	3180 (33.52)	6308 (66.48)	0 (0.00)	9155 (96.49)	333 (3.51)
Total	906 (2.33)	17371 (44.59)	20683 (53.09)	417 (1.07)	14808 (38.01)	23735 (60.92)	9894	28282	784

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

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

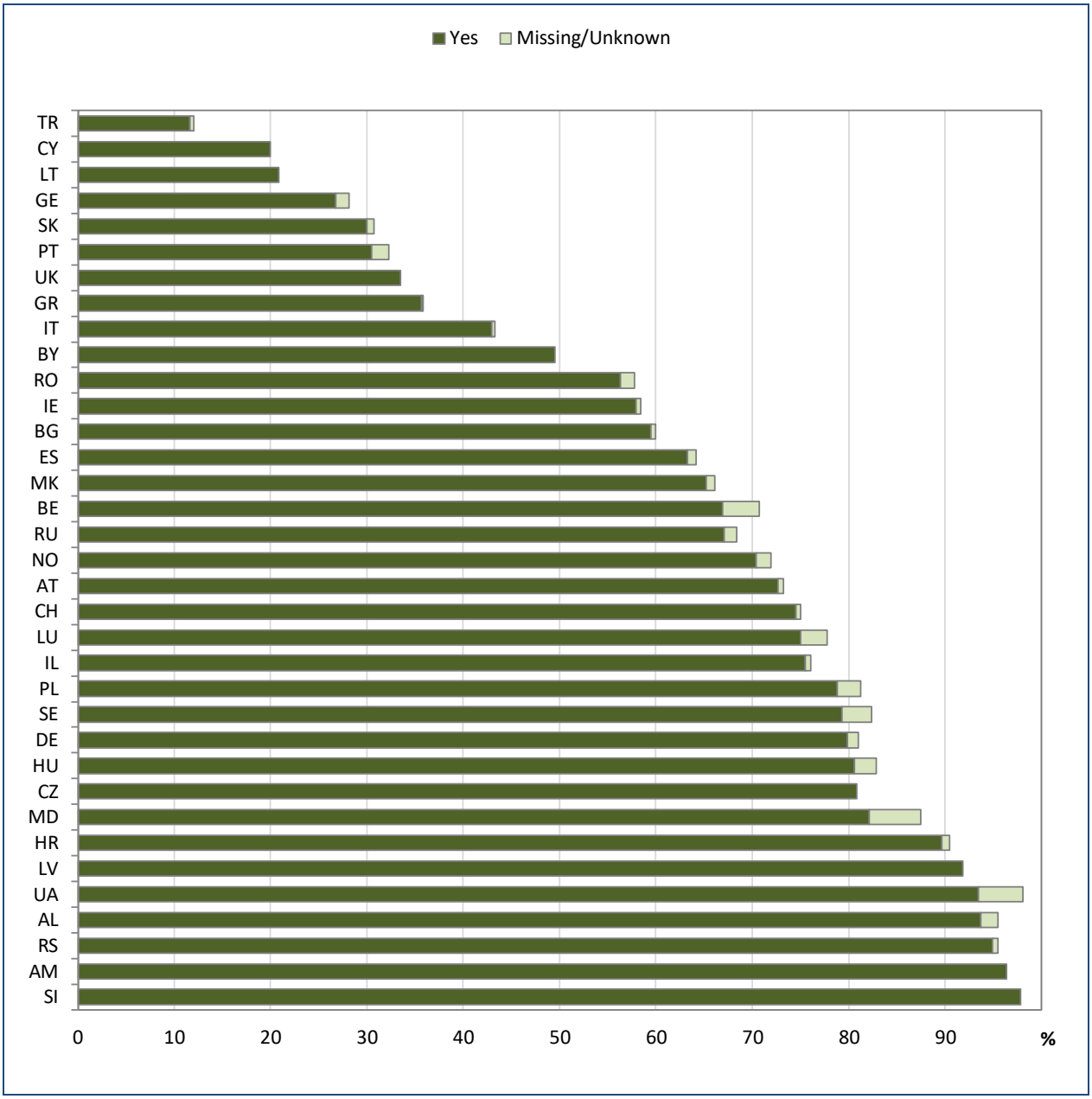
Note: Inhaled hypertonic saline is reimbursed in most countries except in Albania, Bulgaria, Lithuania, Moldova, Poland, Romania, Serbia, Sweden and Turkey, and for children in the Ukraine.

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

Note: Inhaled Mannitol is reimbursed in Austria, Czech Republic, Denmark, Germany (≥18 years), Italy (≥18 years), Russia (> 5 years), Slovenia, Turkey (> 6 years), United Kingdom (≥18 years), but not in the other countries.

Table 7.6 shows the use of three different inhaled medications: hypertonic saline, rhDNase (Pulmozyme®) and mannitol (see page 15 for abbreviations). Hypertonic saline can be any saline of a concentration >0.9% NaCl, but most commonly between 3% and 11%.

Figure 7.5 Use of inhaled hypertonic saline in all patients seen in 2018 who have never had a transplant, by country.



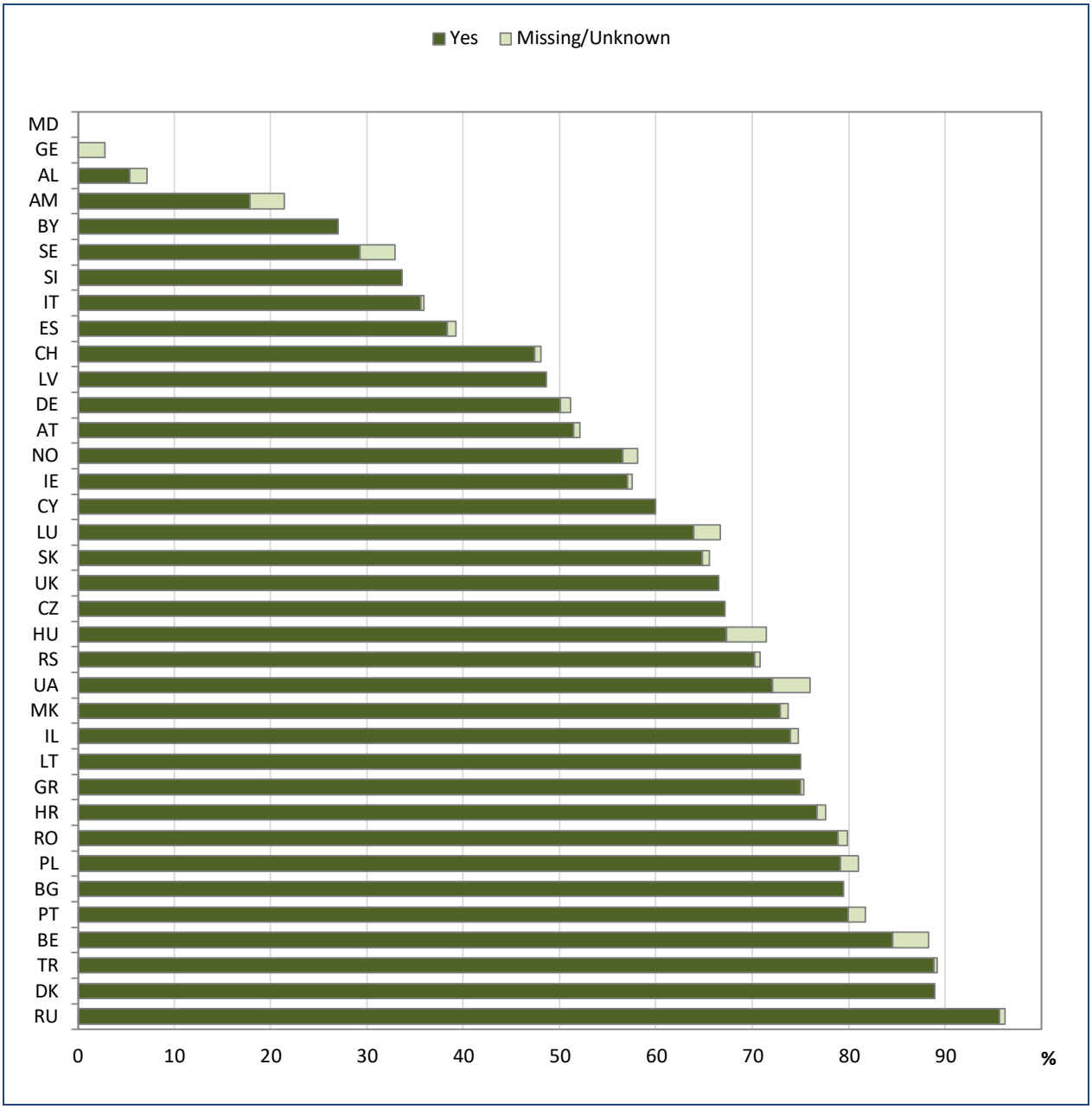
Note: We excluded from the graph the countries for which the information on inhaled hypertonic saline was missing for more than 10% of the patients.

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

Note: Inhaled hypertonic saline is reimbursed everywhere except in Albania, Bulgaria, Lithuania, Moldova, Poland, Romania, Serbia, Sweden and Turkey; in the Ukraine it is reimbursed only for children.

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

Figure 7.6 Use of rhDNase in all patients seen in 2018 who have never had a transplant, 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.

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

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

Table 7.7 Use of inhaled antibiotics, bronchodilators and macrolides in all patients seen in 2018 who have never had a transplant, by country.

Country	Antibiotics inhaled > 3 months this year number (%)			Bronchodilators inhaled > 3 months this year number (%)			Macrolides > 3 months this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	3 (2.70)	71 (63.96)	37 (33.33)	4 (3.60)	31 (27.93)	76 (68.47)	7 (6.31)	89 (80.18)	15 (13.51)
Armenia	0 (0.00)	18 (64.29)	10 (35.71)	0 (0.00)	1 (3.57)	27 (96.43)	0 (0.00)	14 (50.00)	14 (50.00)
Austria	2 (0.28)	449 (63.96)	251 (35.75)	2 (0.28)	42 (5.98)	658 (93.73)	5 (0.71)	664 (94.59)	33 (4.70)
Belarus	0 (0.00)	75 (67.57)	36 (32.43)	0 (0.00)	47 (42.34)	64 (57.66)	0 (0.00)	72 (64.86)	39 (35.14)
Belgium	6 (0.54)	490 (44.03)	617 (55.44)	42 (3.77)	209 (18.78)	862 (77.45)	4 (0.36)	502 (45.10)	607 (54.54)
Bulgaria	0 (0.00)	94 (49.47)	96 (50.53)	0 (0.00)	143 (75.26)	47 (24.74)	0 (0.00)	181 (95.26)	9 (4.74)
Croatia	1 (0.86)	61 (52.59)	54 (46.55)	1 (0.86)	96 (82.76)	19 (16.38)	1 (0.86)	65 (56.03)	50 (43.10)
Cyprus	0 (0)	10 (66.67)	5 (33.33)	0 (0.00)	7 (46.67)	8 (53.33)	0 (0.00)	8 (53.33)	7 (46.67)
Czech Republic	0 (0.00)	428 (75.22)	141 (24.78)	0 (0.00)	230 (40.42)	339 (59.58)	0 (0.00)	531 (93.32)	38 (6.68)
Denmark[1]	180 (38.88)	150 (32.40)	133 (28.73)	463 (100)	0 -	0 -	0 (0.00)	346 (74.73)	117 (25.27)
Georgia	2 (2.82)	68 (95.77)	1 (1.41)	3 (4.23)	65 (91.55)	3 (4.23)	4 (5.63)	67 (94.37)	0 (0.00)
Germany	81 (1.39)	3045 (52.19)	2709 (46.43)	61 (1.05)	901 (15.44)	4873 (83.51)	63 (1.08)	4914 (84.22)	858 (14.70)
Greece	1 (0.18)	204 (36.96)	347 (62.86)	1 (0.18)	223 (40.40)	328 (59.42)	2 (0.36)	311 (56.34)	239 (43.30)
Hungary	9 (2.05)	213 (48.63)	216 (49.32)	11 (2.51)	164 (37.44)	263 (60.05)	19 (4.34)	299 (68.26)	120 (27.40)
Ireland	6 (0.54)	627 (56.13)	484 (43.33)	6 (0.54)	311 (27.84)	800 (71.62)	6 (0.54)	570 (51.03)	541 (48.43)
Israel	3 (0.58)	228 (43.93)	288 (55.49)	2 (0.39)	177 (34.10)	340 (65.51)	3 (0.58)	278 (53.56)	238 (45.86)
Italy	14 (0.27)	3299 (63.34)	1895 (36.39)	17 (0.33)	1529 (29.36)	3662 (70.31)	14 (0.27)	3890 (74.69)	1304 (25.04)
Latvia	0 (0.00)	31 (83.78)	6 (16.22)	0 (0.00)	2 (5.41)	35 (94.59)	0 (0.00)	33 (89.19)	4 (10.81)
Lithuania	0 (0.00)	19 (79.17)	5 (20.83)	0 (0.00)	10 (41.67)	14 (58.33)	0 (0.00)	23 (95.83)	1 (4.17)
Luxembourg	0 (0.00)	21 (58.33)	15 (41.67)	1 (2.78)	7 (19.44)	28 (77.78)	0 (0.00)	22 (61.11)	14)(38.89)
Rep of Moldova	3 (5.36)	28 (50.00)	25 (44.64)	0 (0.00)	50 (89.29)	6 (10.71)	3 (5.36)	50 (89.29)	3 (5.36)
The Netherlands	7 (0.52)	794 (58.90)	547 (40.58)	151 (11.20)	526 (39.02)	671 (49.78)	6 (0.45)	835 (61.94)	507 (37.61)
North Macedonia	3 (2.54)	60 (50.85)	55 (46.61)	2 (1.69)	5 (4.24)	111 (94.07)	2 (1.69)	99 (83.9)	17 (14.41)

[1] Denmark: the high rate of missing information is due to the fact that only one of the two centres reported these data.

[table 7.7 continued]

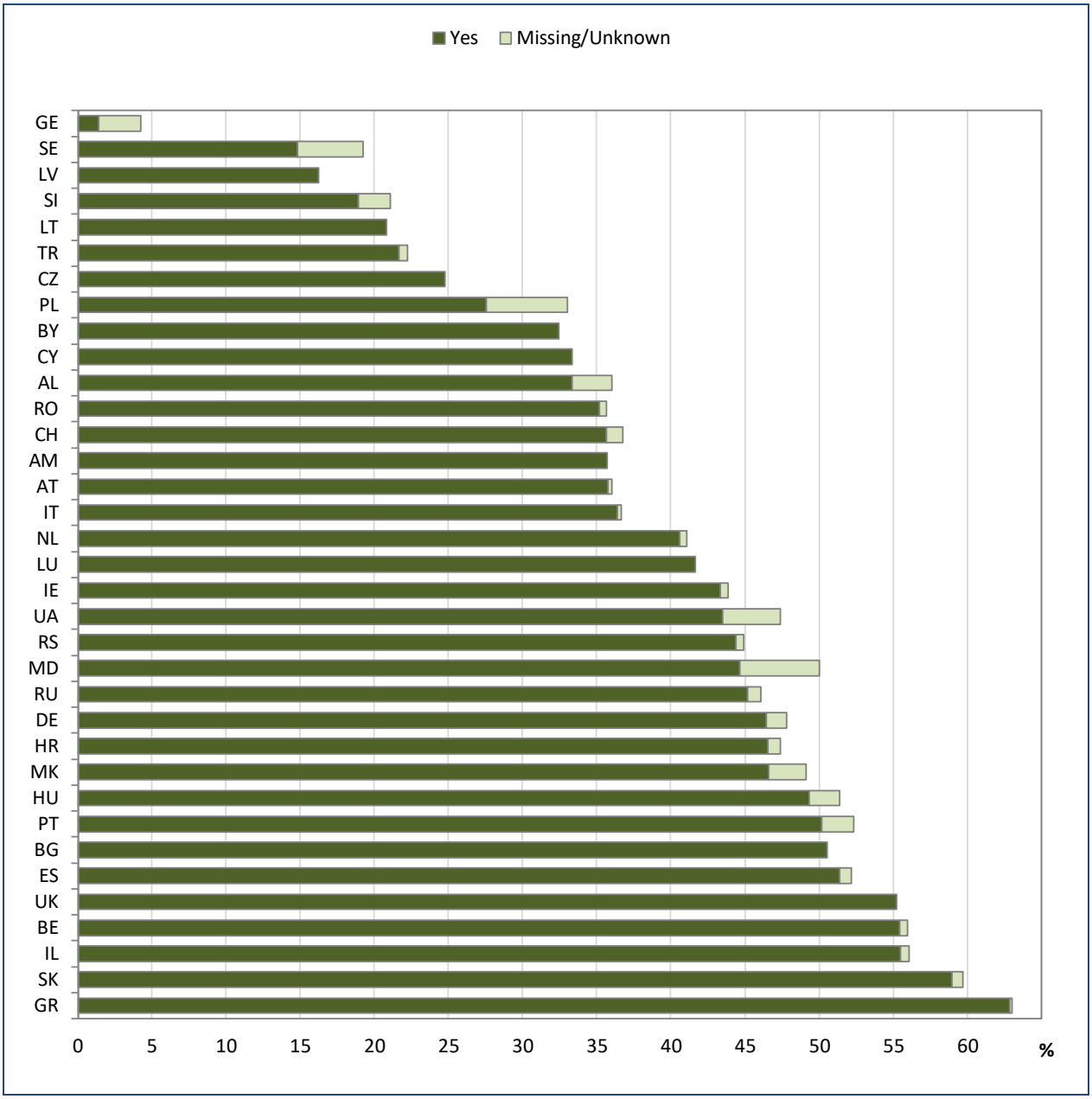
Country	Antibiotics inhaled > 3 months this year number (%)			Bronchodilators inhaled > 3 months this year number (%)			Macrolides > 3 months this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Norway	27 (10.38)	186 (71.54)	47 (18.08)	4 (1.54)	54 (20.77)	202 (77.69)	11 (4.23)	217 (83.46)	32 (12.31)
Poland	47 (5.45)	578 (66.98)	238 (27.58)	35 (4.06)	104 (12.05)	724 (83.89)	51 (5.91)	703 (81.46)	109 (12.63)
Portugal	6 (2.15)	133 (47.67)	140 (50.18)	5 (1.79)	112 (40.14)	162 (58.06)	7 (2.51)	181 (64.87)	91 (32.62)
Romania	1 (0.50)	128 (64.32)	70 (35.18)	1 (0.50)	114 (57.29)	84 (42.21)	2 (1.01)	178 (89.45)	19 (9.55)
Russian Federation	28 (0.90)	1669 (53.93)	1398 (45.17)	25 (0.81)	1543 (49.85)	1527 (49.34)	49 (1.58)	2079 (67.17)	967 (31.24)
Serbia	1 (0.56)	98 (55.06)	79 (44.38)	1 (0.56)	3 (1.69)	174 (97.75)	1 (0.56)	156 (87.64)	21 (11.80)
Slovak Republic	2 (0.73)	110 (40.29)	161 (58.97)	2 (0.73)	108 (39.56)	163 (59.71)	3 (1.10)	161 (58.97)	109 (39.93)
Slovenia	2 (2.11)	75 (78.95)	18 (18.95)	1 (1.05)	77 (81.05)	17 (17.89)	2 (2.11)	85 (89.47)	8 (8.42)
Spain	16 (0.80)	961 (47.81)	1033 (51.39)	15 (0.75)	579 (28.81)	1416 (70.45)	19 (0.95)	1221 (60.75)	770 (38.31)
Sweden	27 (4.44)	491 (80.76)	90 (14.8)	19 (3.13)	38 (6.25)	551 (90.63)	24 (3.95)	438 (72.04)	146 (24.01)
Switzerland	10 (1.14)	557 (63.22)	314 (35.64)	5 (0.57)	151 (17.14)	725 (82.29)	10 (1.14)	654 (74.23)	217 (24.63)
Turkey	10 (0.56)	1400 (77.78)	390 (21.67)	6 (0.33)	1230 (68.33)	564 (31.33)	6 (0.33)	1651 (91.72)	143 (7.94)
Ukraine	6 (3.90)	81 (52.60)	67 (43.51)	7 (4.55)	17 (11.04)	130 (84.42)	7 (4.55)	15 (9.74)	132 (85.71)
United Kingdom[2]	0 (0.00)	4249 (44.78)	5239 (55.22)	0 (0.00)	3877 (40.86)	5611 (59.14)	0 (0.00)	6085 (64.13)	3403 (35.87)
Total	504 (1.29)	21199 (54.41)	17257 (44.29)	893 (2.29)	12783 (32.81)	25284 (64.90)	331 (0.85)	27687 (71.07)	10942 (28.09)

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

Note: Inhaled antibiotics are reimbursed in all countries. In Bulgaria the therapy is available to patients >7 years old only.
Note: Inhaled bronchodilators are reimbursed in most countries except in Bulgaria and Serbia. In the Ukraine the therapy is reimbursed for children, but not for adults.
Note: Oral macrolides are reimbursed in most countries except in Bulgaria, Moldova and Serbia.

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

Figure 7.7 Use of inhaled antibiotics in all patients seen in 2018 who have never had a transplant, by country.

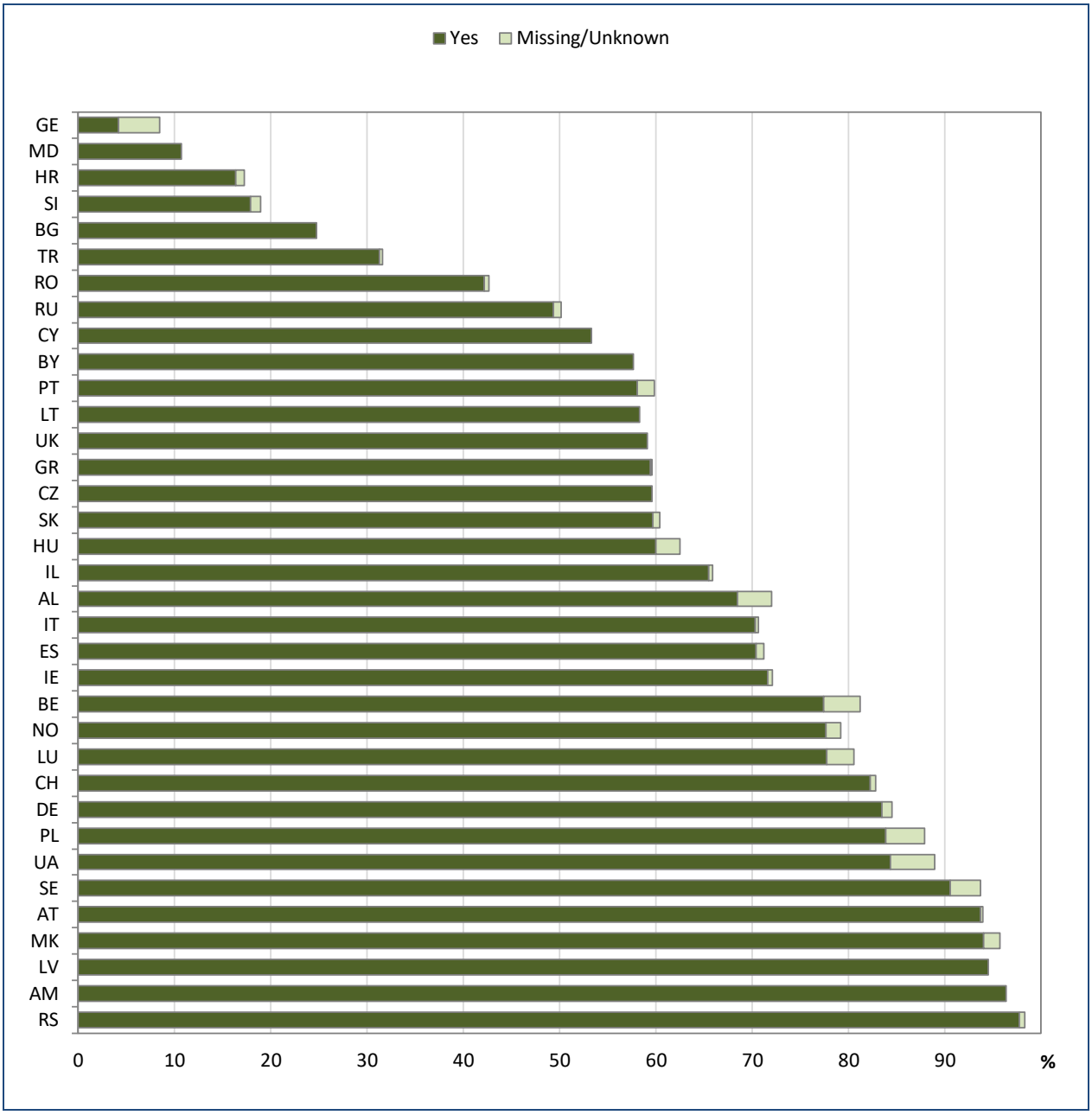


Note: We excluded from the graph the countries for which the information on inhaled antibiotics was missing for more than 10% of the patients.

Note: Inhaled antibiotics are reimbursed in all countries. In Bulgaria the therapy is available to patients >7 years only.

This graph shows the use of inhaled antibiotics (of any kind) for more than three months during the survey year. The frequency varies considerably, from 1.5 to 63%. The dark green part of the bar shows the percentage of patients taking this drug, the light green part shows the percentage of patients for whom this information is missing.

Figure 7.8 Use of bronchodilators in all patients seen in 2018 who have never had a transplant, by country.



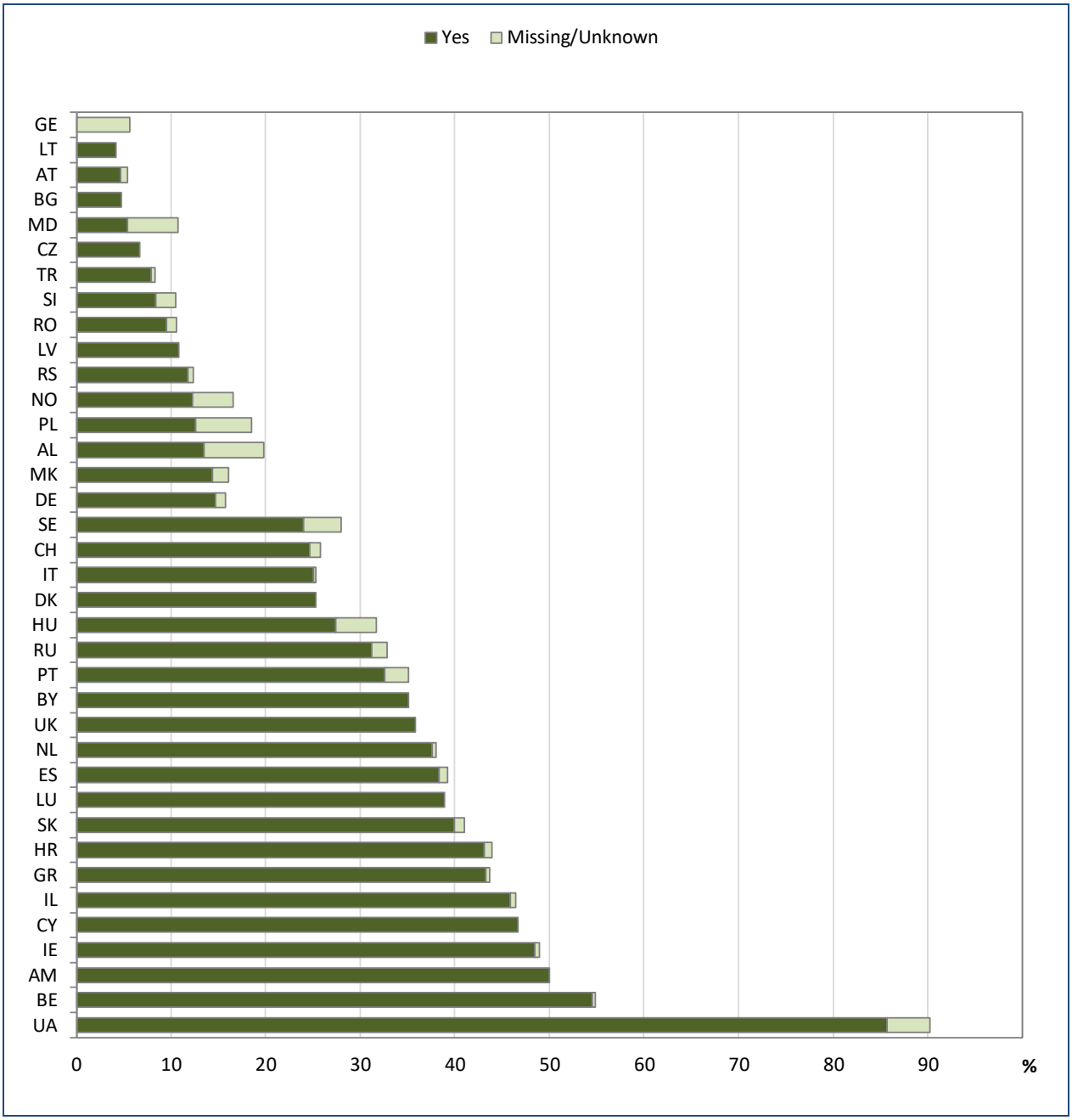
Note: We excluded from the graph the countries for which the information on the use of bronchodilators was missing for more than 10% of the patients.

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

Note: Inhaled bronchodilators are reimbursed in most countries except in Bulgaria and Serbia. In the Ukraine the therapy is reimbursed for children, but not for adults.

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

Figure 7.9 Use of macrolides in all patients seen in 2018 who have never had a transplant, 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: United Kingdom: the duration of use of macrolides is not specified.
Note: Oral macrolides are reimbursed in most countries except in Bulgaria, Moldova and Serbia.

This graph shows the use of macrolides (e.g. azithromycin) for more than 3 months during 2018. Macrolides are antibiotics, but taken continuously they can also modulate the immune system, probably by their anti-inflammatory properties. Clinical studies have shown that patients with chronic Pseudomonas aeruginosa infection benefit from continuous azithromycin treatment with regard to lung function and pulmonary exacerbation rates.

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

Table 7.8 Use of oxygen and non-invasive positive pressure ventilation (NIPPV) in all patients seen in 2018 who have never had a transplant, by country.

Country	Oxygen therapy this year number (%)			NIPPV > 3 months this year number (%)			
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes, BiPAP (Bilevel Positive Airways Pressure)	Yes, CPAP (Continuous Positive Airways Pressure)
Albania	2 (1.80)	105 (94.59)	4 (3.60)	4 (3.60)	107 (96.40)	0 (0.00)	0 (0.00)
Armenia	0 (0.00)	26 (92.86)	2 (7.14)	0 (0.00)	27 (96.43)	0 (0.00)	1 (3.57)
Austria	3 (0.43)	669 (95.30)	30 (4.27)	4 (0.57)	696 (99.15)	1 (0.14)	1 (0.14)
Belarus	0 (0)	111 (100)	0 (0)	0 (0)	111 (100)	0 (0)	0 (0)
Belgium	41 (3.68)	1040 (93.44)	32 (2.88)	1113 (100)	0 -	0 -	0 -
Bulgaria	1 (0.53)	183 (96.32)	6 (3.16)	0 (0.00)	189 (99.47)	0 (0.00)	1 (0.53)
Croatia	2 (1.72)	108 (93.10)	6 (5.17)	1 (0.86)	115 (99.14)	0 (0.00)	0 (0.00)
Cyprus	0 (0)	15 (100)	0 (0)	0 (0)	15 (100)	0 (0)	0 (0)
Czech Republic	0 (0.00)	558 (98.07)	11 (1.93)	0 (0.00)	568 (99.82)	0 (0.00)	1 (0.18)
Denmark	283 (61.12)	176 (38.01)	4 (0.86)	463 (100)	0 -	0 -	0 -
Georgia	3 (4.23)	68 (95.77)	0 (0.00)	1 (1.41)	70 (98.59)	0 (0.00)	0 (0.00)
Germany[1]	59 (1.01)	5340 (91.52)	436 (7.47)	68 (1.17)	5695 (97.60)	0 (0.00)	72 (1.23)
Greece	1 (0.18)	542 (98.19)	9 (1.63)	1 (0.18)	551 (99.82)	0 (0.00)	0 (0.00)
Hungary	19 (4.34)	382 (87.21)	37 (8.45)	438 (100)	0 -	0 -	0 -
Ireland	6 (0.54))	1071 (95.88	40 (3.58)	6 (0.54)	1064 (95.26)	47 (4.21)	0 (0.00)
Israel	3 (0.58)	505 (97.30)	11 (2.12)	2 (0.39)	503 (96.92)	13 (2.50)	1 (0.19)
Italy	662 (12.71)	4290 (82.37)	256 (4.92)	4560 (87.56)	295 (5.66)	0 (0.00)	353 (6.78)
Latvia	0 (0.00)	36 (97.30)	1 (2.70)	0 (0)	37 (100)	0 (0)	0 (0)
Lithuania	0 (0.00)	23 (95.83)	1 (4.17)	0 (0.00)	23 (95.83)	0 (0.00)	1 (4.17)
Luxembourg	0 (0.00)	35 (97.22)	1 (2.78)	0 (0)	36 (100)	0 (0)	0 (0)
Rep of Moldova	3 (5.36)	51 (91.07)	2 (3.57)	3 (5.36)	53 (94.64)	0 (0.00)	0 (0.00)
The Netherlands[1]	149 (11.05)	1159 (85.98)	40 (2.97)	146 (10.83)	1190 (88.28)	0 (0.00)	12 (0.89)

[1] Germany and The Netherlands: don't use the categories BiPAP or CPAP and reported all patients with NIPPV as Continuous Positive Airways Pressure (CPAP).

[table 7.8 continued]

Country	Oxygen therapy this year number (%)			NIPPV > 3 months this year number (%)			
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes, BiPAP (Bilevel Positive Airways Pressure)	Yes, CPAP (Continuous Positive Airways Pressure)
North Macedonia	1 (0.85)	113 (95.76)	4 (3.39)	1 (0.85)	117 (99.15)	0 (0.00)	0 (0.00)
Norway[2]	1 (0.38)	254 (97.69)	5 (1.92)	2 (0.77)	256 (98.46)	1 (0.38)	1 (0.38)
Poland	53 (6.14)	780 (90.38)	30 (3.48)	54 (6.26)	806 (93.40)	3 (0.35)	0 (0.00)
Portugal	5 (1.79)	255 (91.40)	19 (6.81)	6 (2.15)	264 (94.62)	9 (3.23)	0 (0.00)
Romania	2 (1.01)	190 (95.48)	7 (3.52)	1 (0.50)	198 (99.50)	0 (0.00)	0 (0.00)
Russian Federation	21 (0.68)	2946 (95.19)	128 (4.14)	21 (0.68)	3040 (98.22)	23 (0.74)	11 (0.36)
Serbia	1 (0.56)	171 (96.07)	6 (3.37)	1 (0.56)	175 (98.31)	2 (1.12)	0 (0.00)
Slovak Republic	2 (0.73)	257 (94.14)	14 (5.13)	4 (1.47)	266 (97.44)	0 (0.00)	3 (1.10)
Slovenia	0 (0.00)	92 (96.84)	3 (3.16)	0 (0.00)	92 (96.84)	3 (3.16)	0 (0.00)
Spain	16 (0.80)	1931 (96.07)	63 (3.13)	17 (0.85)	1986 (98.81)	6 (0.30)	1 (0.05)
Sweden	19 (3.13)	575 (94.57)	14 (2.30)	607 (99.84)	1 (0.16)	0 (0.00)	0 (0.00)
Switzerland	9 (1.02)	840 (95.35)	32 (3.63)	8 (0.91)	866 (98.30)	3 (0.34)	4 (0.45)
Turkey	6 (0.33)	1750 (97.22)	44 (2.44)	6 (0.33)	1749 (97.17)	42 (2.33)	3 (0.17)
Ukraine	6 (3.90)	123 (79.87)	25 (16.23)	6 (3.90)	148 (96.10)	0 (0.00)	0 (0.00)
United Kingdom	0 (0.00)	8913 (93.94)	575 (6.06)	0 (0.00)	9310 (98.12)	0 (0.00)	178 (1.88)
Total	1379 (3.54)	35683 (91.59)	1898 (4.87)	7544	30619	153	644

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

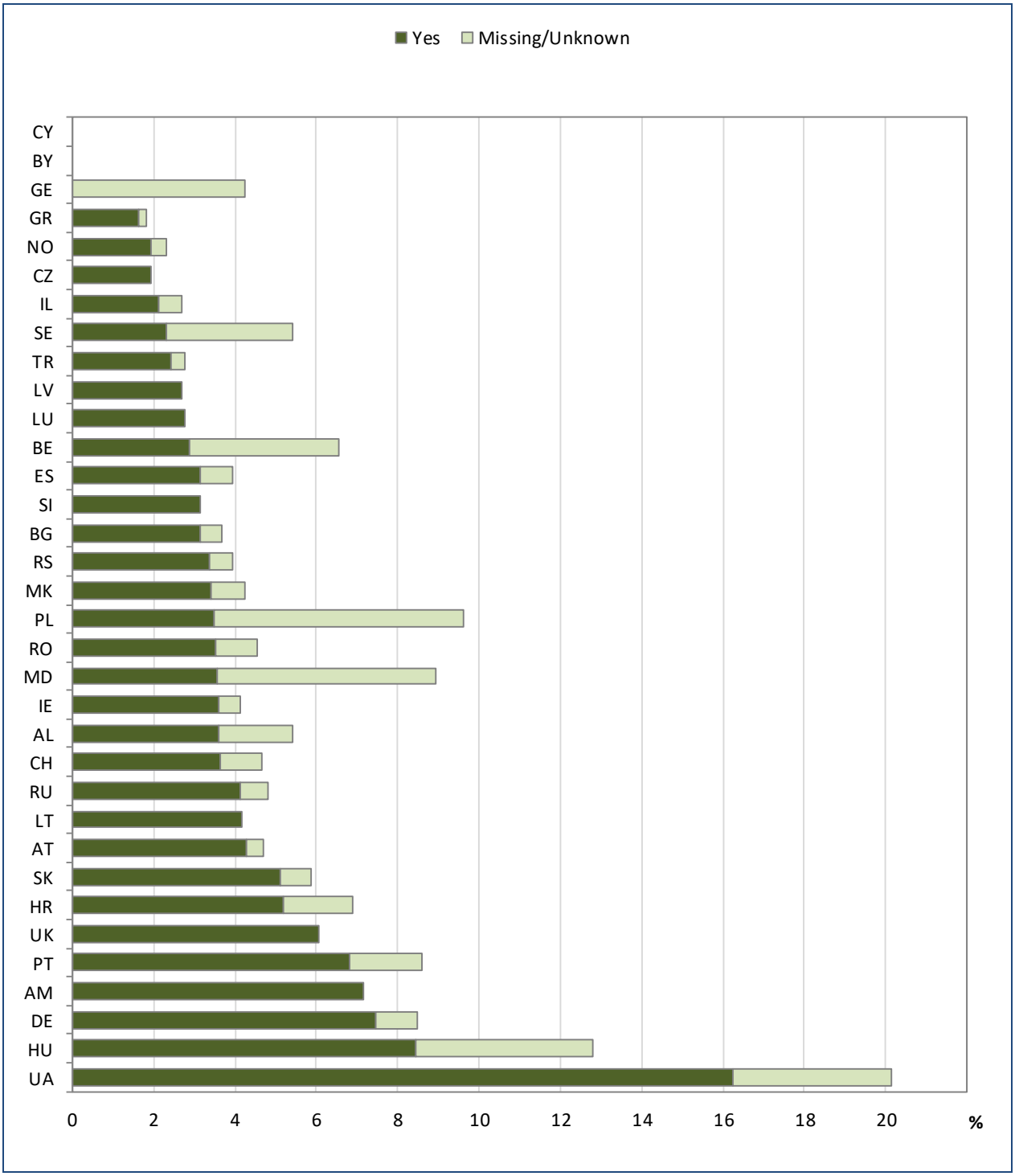
Note: For non-invasive positive pressure ventilation (NIPPV) the percentage of missing information is higher than 10%, therefore the percentages are excluded from the table.

Note: Oxygen therapy is reimbursed in most countries except in Bulgaria, Moldova, Serbia, Russia and Ukraine. In Latvia the therapy is reimbursed for adults only.

Note: Noninvasive positive pressure ventilation (NIPPV) is reimbursed in most countries, except in Albania, Belarus, Belgium, Bulgaria, Hungary, Latvia, Macedonia, Moldova, Serbia, Russia and Ukraine.

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

Figure 7.10 Use of oxygen in all patients seen in 2018 who have never had a transplant, by country.



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

Note: Oxygen therapy is reimbursed in most countries except in Bulgaria, Moldova, Serbia, Russia and Ukraine. In Latvia the therapy is reimbursed for adults only.

This graph shows the use of oxygen during 2018. Oxygen is used for severe lung disease. The dark green part of the bar indicates the percentage of patients using oxygen supplementation, the light green part shows the percentage of patients for whom this information is missing.

Table 7.9 Use of inhaled steroids, oral steroids and proton pump inhibitors (PPI) in all patients seen in 2018 who have never had a transplant, by country.

Country	Inhaled steroids > 3 months this year number (%)			Oral steroids > 3 months this year number (%)			PPI > 3 months this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Albania	3 (2.70)	88 (79.28)	20 (18.02)	5 (4.50)	104 (93.69)	2 (1.80)	4 (3.60)	100 (90.09)	7 (6.31)
Armenia	0 (0.00)	20 (71.43)	8 (28.57)	0 (0)	28 (100)	0 (0)	0 (0.00)	6 (21.43)	22 (78.57)
Austria	3 (0.43)	624 (88.89)	75 (10.68)	5 (0.71)	678 (96.58)	19 (2.71)	4 (0.57)	650 (92.59)	48 (6.84)
Belarus	0 (0.00)	63 (56.76)	48 (43.24)	0 (0.00)	98 (88.29)	13 (11.71)	0 (0)	111 (100)	0 (0)
Belgium	0 (0.00)	545 (48.97)	568 (51.03)	0 (0.00)	1059 (95.15)	54 (4.85)	0 (0.00)	570 (51.21)	543 (48.79)
Bulgaria	0 (0.00)	175 (92.11)	15 (7.89)	0 (0.00)	188 (98.95)	2 (1.05)	1 (0.53)	162 (85.26)	27 (14.21)
Croatia	1 (0.86)	106 (91.38)	9 (7.76)	1 (0.86)	115 (99.14)	0 (0.00)	1 (0.86)	87 (75.00)	28 (24.14)
Cyprus	0 (0.00)	8 (53.33)	7 (46.67)	0 (0)	15 (100)	0 (0)	0 (0)	15 (100)	0 (0)
Czech Republic	0 (0.00)	366 (64.32)	203 (35.68)	0 (0.00)	557 (97.89)	12 (2.11)	0 (0.00)	451 (79.26)	118 (20.74)
Denmark	463 (100)	0 -	0 -	463 (100)	0 -	0 -	463 (100)	0 -	0 -
Georgia	2 (2.82)	69 (97.18)	0 (0.00)	1 (1.41)	70 (98.59)	0 (0.00)	1 (1.41)	68 (95.77)	2 (2.82)
Germany	62 (1.06)	3702 (63.44)	2071 (35.49)	68 (1.17)	5455 (93.49)	312 (5.35)	63 (1.08)	4519 (77.45)	1253 (21.47)
Greece	1 (0.18)	461 (83.51)	90 (16.30)	1 (0.18)	543 (98.37)	8 (1.45)	2 (0.36)	487 (88.22)	63 (11.41)
Hungary	438 (100)	0 -	0 -	438 (100)	0 -	0 -	438 (100)	0 -	0 -
Ireland	13 (1.16)	769 (68.85)	335 (29.99)	1117 (100)	0 -	0 -	23 (2.06)	593 (53.09)	501 (44.85)
Israel	6 (1.16)	276 (53.18)	237 (45.66)	3 (0.58)	482 (92.87)	34 (6.55)	2 (0.39)	350 (67.44)	167 (32.18)
Italy	4560 (87.56)	566 (10.87)	82 (1.57)	4560 (87.56)	573 (11.00)	75 (1.44)	4560 (87.56)	295 (5.66)	353 (6.78)
Latvia	0 (0.00)	33 (89.19)	4 (10.81)	0 (0)	37 (100)	0 (0)	0 (0.00)	32 (86.49)	5 (13.51)
Lithuania	0 (0.00)	22 (91.67)	2 (8.33)	0 (0)	24 (100)	0 (0)	0 (0.00)	20 (83.33)	4 (16.67)
Luxembourg	1 (2.78)	10 (27.78)	25 (69.44)	0 (0.00)	33 (91.67)	3 (8.33)	0 (0.00)	17 (47.22)	19 (52.78)
Rep of Moldova	3 (5.36)	49 (87.50)	4 (7.14)	3 (5.36)	51 (91.07)	2 (3.57)	3 (5.36)	49 (87.50)	4 (7.14)
The Netherlands	153 (11.35)	684 (50.74)	511 (37.91)	152 (11.28)	1068 (79.23)	128 (9.50)	206 (15.28)	613 (45.47)	529 (39.24)
North Macedonia	3 (2.54)	107 (90.68)	8 (6.78)	1 (0.85)	117 (99.15)	0 (0.00)	2 (1.69)	63 (53.39)	53 (44.92)
Norway	3 (1.15)	216 (83.08)	41 (15.77)	5 (1.92)	252 (96.92)	3 (1.15)	13 (5.00)	198 (76.15)	49 (18.85)

[table 7.9 continued]

Country	Inhaled steroids > 3 months this year number (%)			Oral steroids > 3 months this year number (%)			PPI > 3 months this year number (%)		
	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes	Missing/ unknown	No	Yes
Poland	55 (6.37)	637 (73.81)	171 (19.81)	55 (6.37)	799 (92.58)	9 (1.04)	52 (6.03)	667 (77.29)	144 (16.69)
Portugal	6 (2.15)	199 (71.33)	74 (26.52)	6 (2.15)	270 (96.77)	3 (1.08)	5 (1.79)	204 (73.12)	70 (25.09)
Romania	2 (1.01)	189 (94.97)	8 (4.02)	1 (0.50)	197 (98.99)	1 (0.50)	1 (0.50)	176 (88.44)	22 (11.06)
Russian Federation	24 (0.78)	2619 (84.62)	452 (14.6)	24 (0.78)	2980 (96.28)	91 (2.94)	42 (1.36)	2488 (80.39)	565 (18.26)
Serbia	1 (0.56)	123 (69.10)	54 (30.34)	1 (0.56)	175 (98.31)	2 (1.12)	1 (0.56)	122 (68.54)	55 (30.90)
Slovak Republic	2 (0.73)	121 (44.32)	150 (54.95)	1 (0.37)	239 (87.55)	33 (12.09)	2 (0.73)	212 (77.66)	59 (21.61)
Slovenia	1 (1.05)	88 (92.63)	6 (6.32)	2 (2.11)	92 (96.84)	1 (1.05)	5 (5.26)	70 (73.68)	20 (21.05)
Spain	19 (0.95)	1210 (60.20)	781 (38.86)	19 (0.95)	1917 (95.37)	74 (3.68)	15 (0.75)	1418 (70.55)	577 (28.71)
Sweden	32 (5.26)	412 (67.76)	164 (26.97)	23 (3.78)	579 (95.23)	6 (0.99)	32 (5.26)	458 (75.33)	118 (19.41)
Switzerland	6 (0.68)	603 (68.44)	272 (30.87)	5 (0.57)	836 (94.89)	40 (4.54)	8 (0.91)	695 (78.89)	178 (20.20)
Turkey	5 (0.28)	1448 (80.44)	347 (19.28)	6 (0.33)	1771 (98.39)	23 (1.28)	5 (0.28)	1573 (87.39)	222 (12.33)
Ukraine	6 (3.90)	79 (51.30)	69 (44.81)	7 (4.55)	145 (94.16)	2 (1.30)	6 (3.90)	65 (42.21)	83 (53.90)
United Kingdom	0 (0.00)	7862 (82.86)	1626 (17.14)	0 (0.00)	8790 (92.64)	698 (7.36)	0 (0.00)	5142 (54.19)	4346 (45.81)
Total	5874	24549	8537	6973	30337	1650	5960	22746	10254

Note: For inhaled and oral steroids and proton pump inhibitors (PPI) the percentage of missing information is higher than 10%, therefore the percentages are excluded from the table.

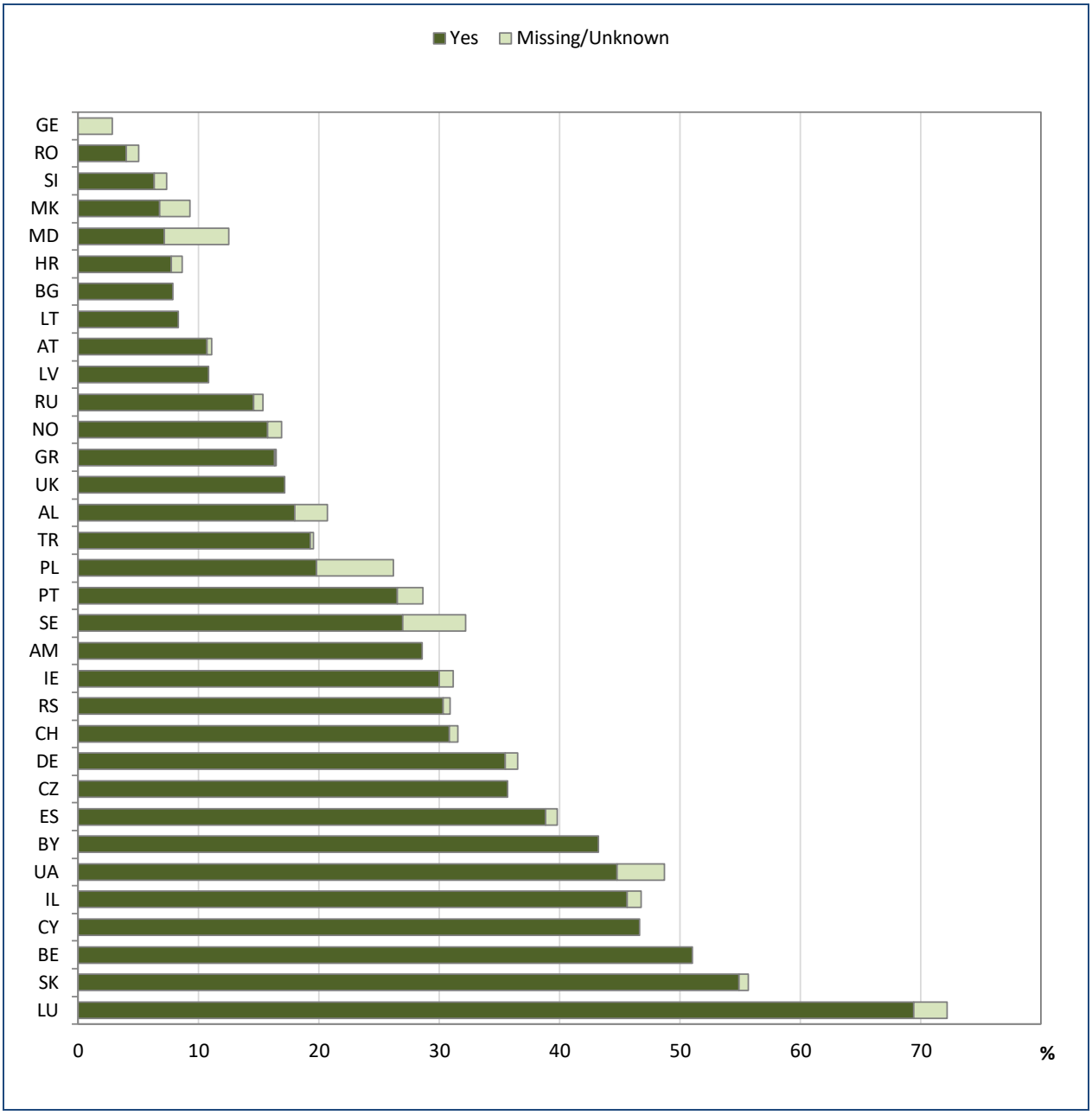
Note: Inhaled steroids are reimbursed in most countries except in Georgia, Lithuania, Poland and Serbia. In Moldova they are reimbursed for children only.

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

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

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

Figure 7.11 Use of inhaled steroids in all patients seen in 2018 who have never had a transplant, by country.

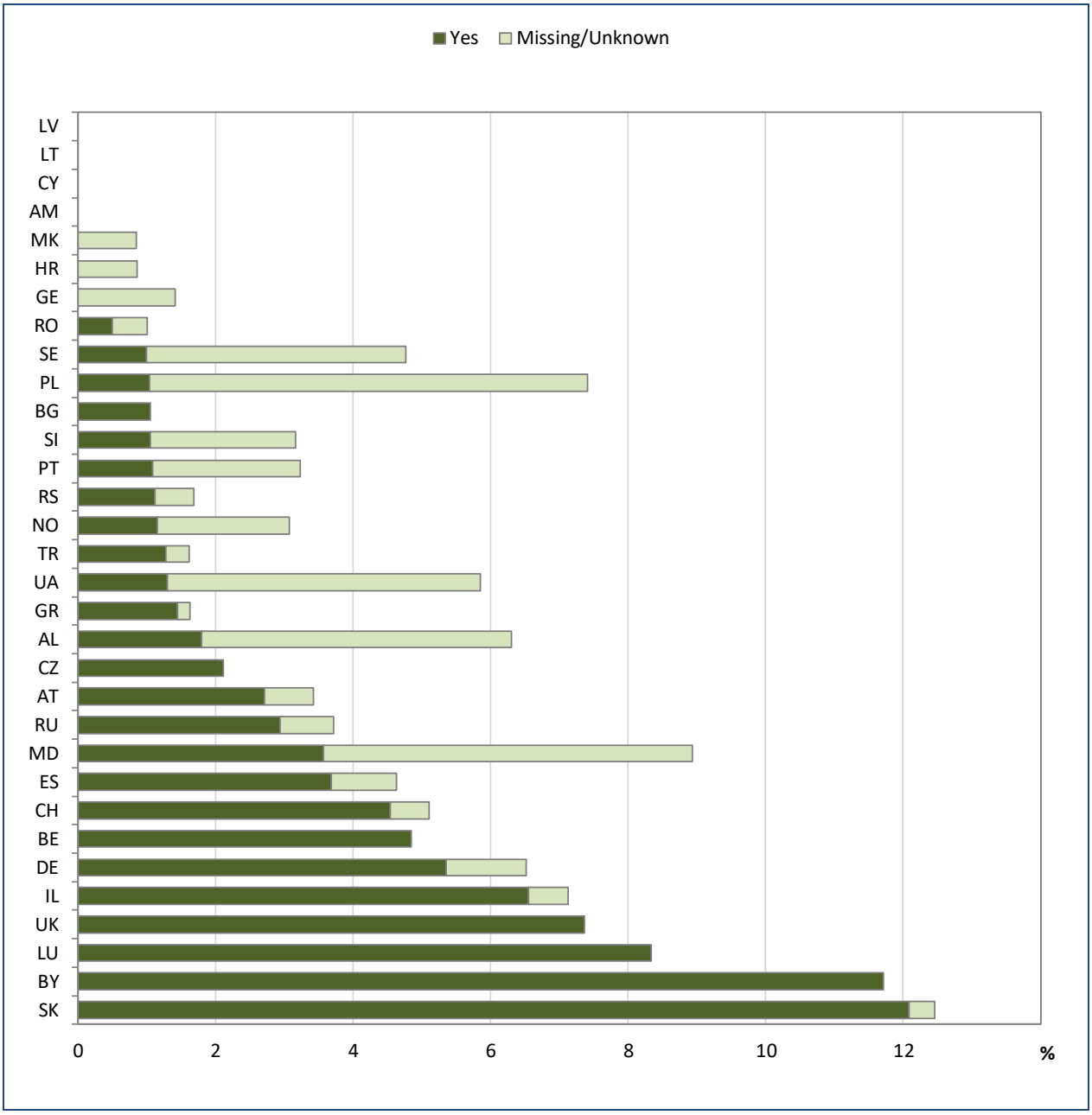


Note: We excluded from the graph the countries for which the information on use of inhaled steroids was missing for more than 10% of the patients.

Note: Inhaled steroids are reimbursed in most countries except in Georgia, Lithuania, Poland and Serbia. In Moldova they are reimbursed for children only.

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

Figure 7.12 Use of oral steroids in all patients seen in 2018 who have never had a transplant, by country.

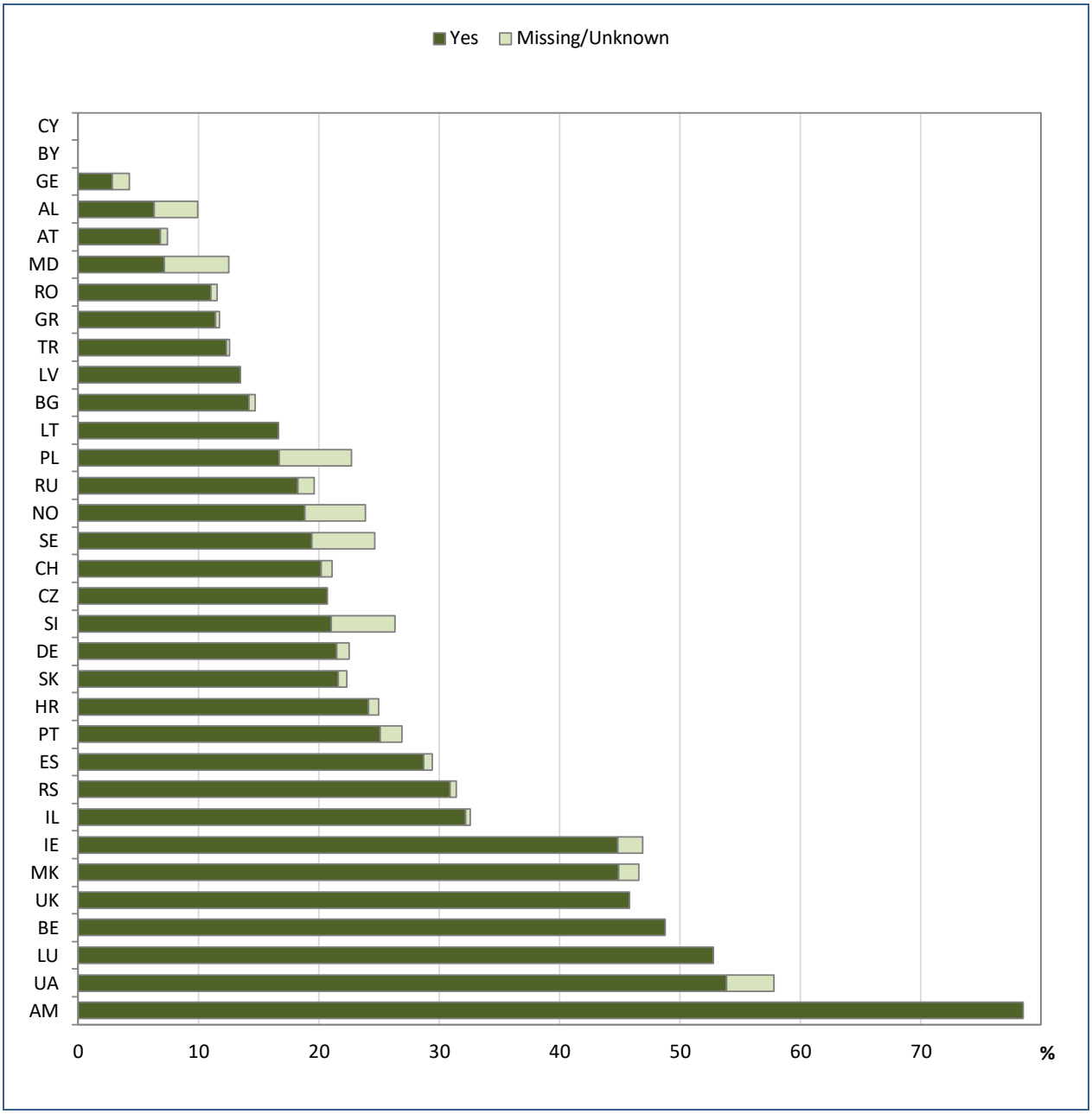


Note: We excluded from the graph the countries for which the information on use of oral steroids was missing for more than 10% of the patients.

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

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

Figure 7.13 Use of proton pump inhibitors (PPI) in all patients seen in 2018 who have never had a transplant, by country.

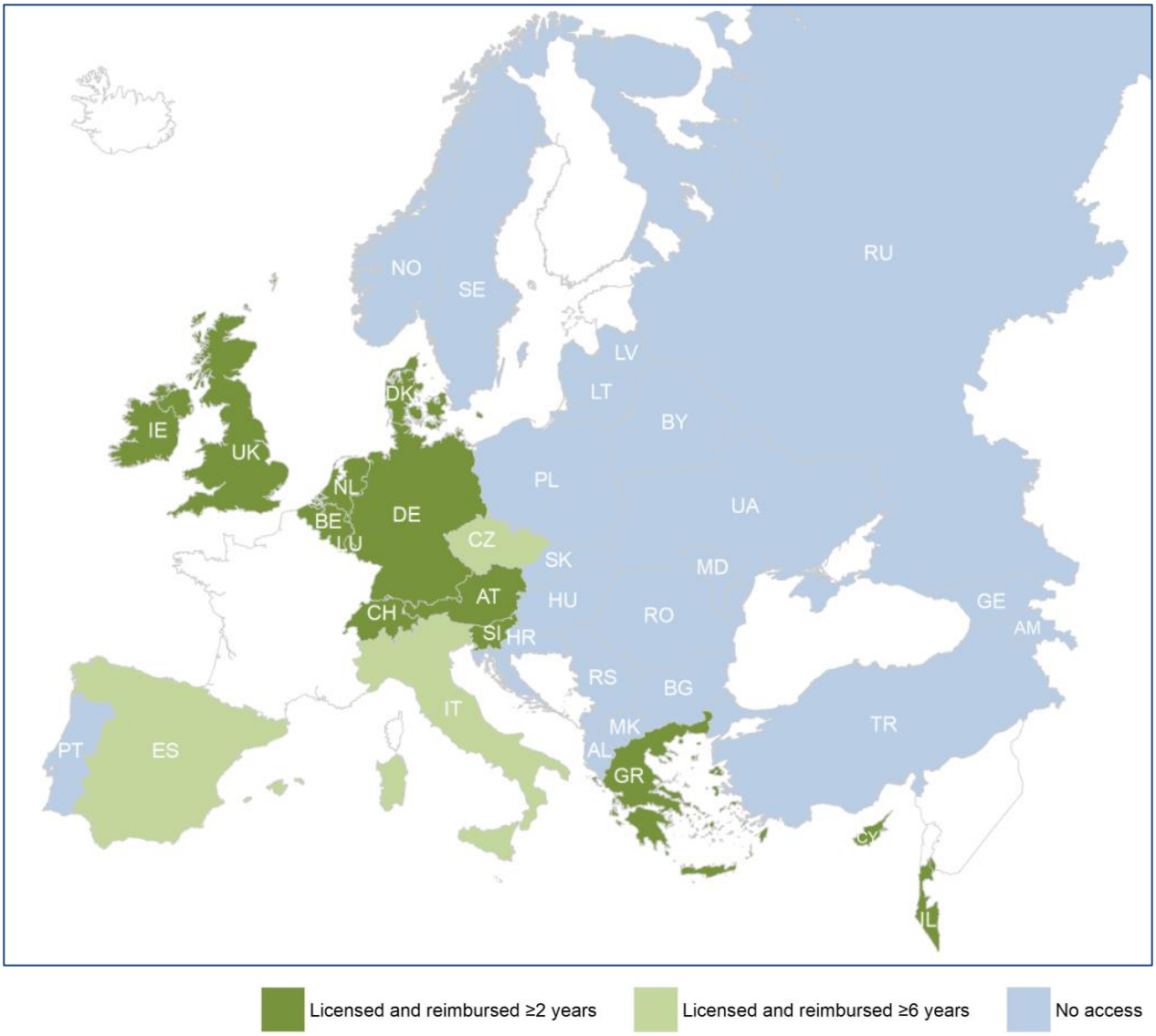


Note: We excluded from the graph the countries for which the information on the use of PPI was missing for more than 10% of the patients.

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

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

Figure 7.14 Use of Ivacaftor in all eligible patients seen in 2018 who had never had a transplant, by country and age group.



In this graph we highlighted the countries where Ivacaftor is licensed and reimbursed; for 2 years and older the country is shaded in green; for 6 years and older light green is used.

Table 7.10 Use of Ivacaftor in all eligible patients seen in 2018 who had never had a transplant, by country and age group.

Country	Age at follow-up (years)	Use of Ivacaftor this year number (%)					
		Missing/unknown		No		Yes	
Albania	**1-17**	0	(0)	2	(100)	0	(0)
Austria	**1-17**	0	(0.00)	7	(58.33)	5	(41.67)
(L &R ≥ 2 yrs)	**≥18**	0	(0.00)	6	(35.29)	11	(64.71)
Belgium	**1-17**	0	(0)	2	(11.76)	15	(88.24)
(L&R ≥ 2 yrs, no R117H)	**≥18**	0	(0.00)	19	(37.25)	32	(62.75)
Bulgaria	**1-17**	0	(0)	5	(100)	0	(0)
(L)	**≥18**	0	(0)	1	(100)	0	(0)
Croatia (L)	**1-17**	0	(0)	1	(100)	0	(0)
Czech Republic	**1-17**	0	(0)	2	(18.18)	9	(81.82)
(L&R ≥ 6yrs)	**≥18**	0	(0.00)	7	(46.67)	8	(53.33)
Denmark (L&R ≥ 2yrs)	**≥18**	0	(0.00)	13	(86.67)	2	(13.33)
Germany	**1-17**	2	(2.59)	17	(22.08)	58	(75.32)
(L&R ≥ 2 yrs)	**≥18**	2	(1.13)	51	(28.81)	124	(70.06)
Greece	**1-17**	0	(0)	0	(0)	5	(100)
(L&R ≥ 2 yrs)	**≥18**	0	(0)	0	(0)	1	(100)
Hungary L	**≥18**	2	(100)	-		-	
Ireland	**1-17**	0	(0.00)	2	(2.38)	82	(97.62)
(L&R ≥ 2 yrs)	**≥18**	7	(4.52)	25	(16.13)	123	(79.35)
Israel	**1-17**	0	(0)	0	(0)	8	(100)
(L&R ≥2 yrs)	**≥18**	0	(0.00)	2	(28.57)	5	(71.43)
Italy	**1-17**	75	(96.15)	2	(2.56)	1	(1.28)
(L&R > 6 yrs, no R117H)	**≥18**	107	(94.69)	2	(1.77)	4	(3.54)
Rep of Moldova	**6-11**	0	(0)	1	(100)	0	(0)
The Netherlands	**1-17**	0	(0)	0	(0)	10	(100)
(L&R ≥ 2 yrs)	**≥18**	0	(0.00)	13	(35.14)	24	(64.86)
North Macedonia	**1-17**	0	(0)	1	(100)	0	(0)
	≥18	0	(0)	2	(100)	0	(0)
Norway	**1-17**	0	(0)	0	(0)	3	(100)
(L ≥ 6 yrs)	**≥18**	0	(0.00)	5	(38.46)	8	(61.54)
Poland	**1-17**	0	(0)	1	(100)	0	(0)
(L)	**≥18**	0	(0)	2	(100)	0	(0)
Portugal (L)	**≥18**	0	(0)	2	(100)	0	(0)
Romania (L)	**1-17**	0	(0)	2	(100)	0	(0)
Russian Federation	**1-17**	0	(0)	4	(100)	0	(0)
	≥18	0	(0)	1	(100)	0	(0)
Slovak Republic	**6-11**	0	(0)	1	(100)	0	(0)
(L)	**≥18**	0	(0)	5	(100)	0	(0)
Spain	**1-17**	0	(0)	1	(11.11)	8	(88.89)
(L&R ≥ 6 yrs)	**≥18**	0	(0.00)	9	(37.50)	15	(62.50)
Sweden	**1-17**	7	(100)	-		-	
(L≥ 6 yrs, no R117H)	**≥18**	13	(100)	-		-	
Switzerland	**1-17**	0	(0)	3	(33.33)	6	(66.67)
(L&R ≥ 2 yrs)	**≥18**	0	(0.00)	7	(77.78)	2	(22.22)
Turkey	**1-17**	0	(0)	12	(100)	0	(0)
	≥18	0	(0)	1	(100)	0	(0)
United Kingdom	**1-17**	0	(0.00)	43	(16.73)	214	(83.27)
(L&R ≥ 2 yrs)	**≥18**	0	(0.00)	275	(42.64)	370	(57.36)
Total	**1-17**	84	(13.61)	109	(17.67)	424	(68.72)
	≥18	131	(10.02)	448	(34.25)	729	(55.73)

Note: L = licensed, R = reimbursed.

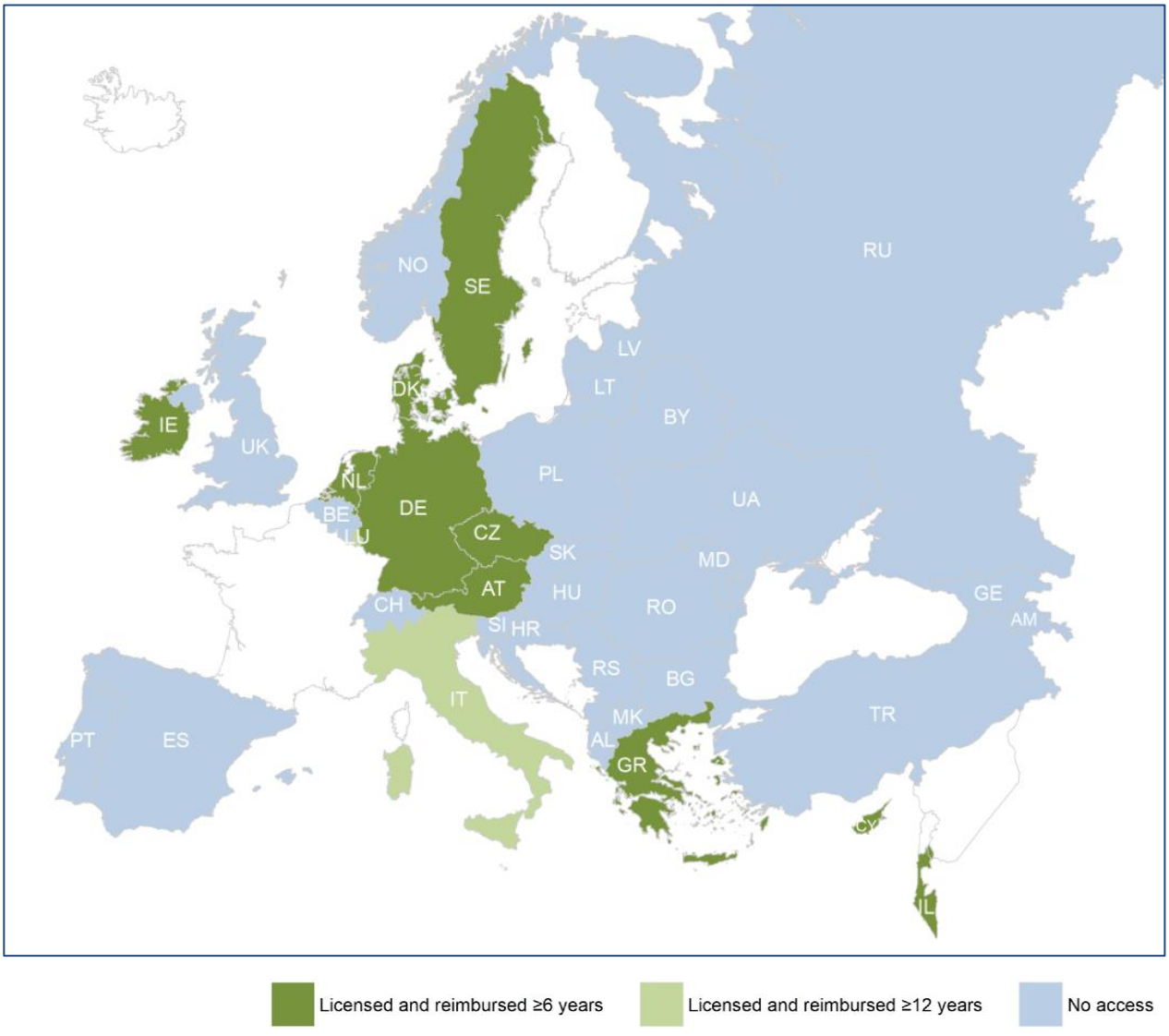
Note: Countries that do not have patients that are eligible for Ivacaftor are not included in the table.

In table 7.10 we indicated in which countries Ivacaftor is licensed (i.e. authorised), or licensed and reimbursed.

The eligibility criteria for Ivacaftor in 2018 are:

- The patient must be 1 year and older with at least one of the mutations G551D, G1244E, G1349D, G178R, G551S, S1251N, S1255P, S549N, S549R;
- The patient must be 18 years and older with at least one of the mutations G551D, G1244E, G1349D, G178R, G551S, S1251N, S1255P, S549R, S549N, R117H.

Figure 7.15 Use of Lumacaftor/Ivacaftor in all eligible patients seen in 2018 who had never had a transplant, by country and age group.



In this graph we highlighted the countries where Lumacaftor/Ivacaftor is licensed and reimbursed; for 6 years and older the country is shaded in green; for 12 years and older light green is used.

Table 7.11 Use of Lumacaftor/Ivacaftor in all eligible patients seen in 2018 who had never had a transplant, by country and age group.

Country	Age at follow-up (years)	Use of Lumacaftor/Ivacaftor this year number (%)					
		Missing/ unknown		No		Yes	
Albania	**6-17**	0	(0)	49	(100)	0	(0)
	≥18	0	(0)	1	(100)	0	(0)
Austria	**6-17**	0	(0.00)	93	(81.58)	21	(18.42)
(L&R ≥ 6 yrs)	**≥18**	1	(0.62)	101	(62.73)	59	(36.65)
Belarus	**6-17**	0	(0)	35	(100)	0	(0)
	≥18	0	(0)	1	(100)	0	(0)
Belgium	**6-17**	0	(0.00)	116	(86.57)	18	(13.43)
(L)	**≥18**	2	(0.73)	224	(81.45)	49	(17.82)
Bulgaria	**6-17**	0	(0)	36	(100)	0	(0)
(L)	**≥18**	0	(0.00)	23	(95.83)	1	(4.17)
Croatia	**6-17**	0	(0)	32	(100)	0	(0)
(L)	**≥18**	0	(0)	28	(100)	0	(0)
Cyprus	**6-17**	0	(0)	1	(50.00)	1	(50.00)
(L&R ≥ 6 yrs)	**≥18**	0	(0)	1	(100)	0	(0)
Czech Republic	**6-17**	0	(0.00)	95	(96.94)	3	(3.06)
(L ≥ 6 yrs)	**≥18**	0	(0.00)	85	(84.16)	16	(15.84)
Denmark	**6-17**	0	(0.00)	33	(36.67)	57	(63.33)
(L&R ≥ 6 yrs)	**≥18**	0	(0.00)	101	(59.76)	68	(40.24)
Germany	**6-17**	2	(0.25)	606	(74.54)	205	(25.22)
(L&R ≥ 6 yrs)	**≥18**	20	(1.30)	1050	(68.05)	473	(30.65)
Greece	**6-17**	0	(0.00)	13	(15.48)	71	(84.52)
(L&R ≥ 6 yrs)	**≥18**	0	(0.00)	21	(25.00)	63	(75.00)
Hungary	**6-17**	74	(100)	-		-	
(L)	**≥18**	63	(100)	-		-	
Ireland	**6-17**	1	(0.45)	10	(4.55)	209	(95.00)
(L&R ≥ 6 yrs)	**≥18**	3	(0.93)	68	(21.05)	252	(78.02)
Israel	**6-17**	0	(0)	0	(0)	15	(100)
(L&R ≥ 6 yrs)	**≥18**	0	(0.00)	8	(23.53)	26	(76.47)
Italy	**6-17**	310	(89.86)	23	(6.67)	12	(3.48)
(L&R > 12 yrs)	**≥18**	481	(83.22)	37	(6.40)	60	(10.38)
Latvia	**6-17**	0	(0)	10	(100)	0	(0)
(L)	**≥18**	0	(0)	6	(100)	0	(0)
Lithuania	**6-17**	0	(0)	2	(100)	0	(0)
(L ≥ 6 yrs)	**≥18**	0	(0)	6	(100)	0	(0)
Luxembourg	**6-17**	0	(0.00)	5	(83.33)	1	(16.67)
(L&R ≥ 12 yrs)	**≥18**	0	(0.00)	5	(45.45)	6	(54.55)
Rep of Moldova	**6-17**	0	(0)	10	(100)	0	(0)
	≥18	0	(0)	2	(100)	0	(0)
The Netherlands	**6-17**	0	(0.00)	26	(10.70)	217	(89.30)
(L&R ≥ 6 yrs)	**≥18**	0	(0.00)	89	(21.04)	334	(78.96)
North Macedonia	**6-17**	0	(0)	33	(100)	0	(0)
	≥18	0	(0)	14	(100)	0	(0)
Norway	**6-17**	0	(0.00)	20	(71.43)	8	(28.57)
(L ≥ 12 yrs)	**≥18**	1	(1.96)	33	(64.71)	17	(33.33)
Poland	**6-17**	12	(7.45)	148	(91.93)	1	(0.62)
(L)	**≥18**	6	(6.82)	78	(88.63)	4	(4.55)
Portugal	**6-17**	2	(3.33)	53	(88.33)	5	(8.33)
(L)	**≥18**	1	(2.33)	35	(81.39)	7	(16.28)
Romania	**6-17**	0	(0)	55	(100)	0	(0)
(L)	**≥18**	0	(0)	4	(100)	0	(0)
Russian Federation	**6-17**	4	(0.90)	439	(98.87)	1	(0.23)
	≥18	2	(1.23)	159	(97.55)	2	(1.23)
Serbia	**6-17**	0	(0)	51	(100)	0	(0)
	≥18	0	(0)	27	(100)	0	(0)

[table 7.11 continued]

Country	Age at follow-up (years)	Use of Lumacaftor/Ivacaftor this year number (%)					
		Missing/ unknown		No		Yes	
Slovak Republic	**6-17**	0	(0)	33	(100)	0	(0)
(L)	**≥18**	0	(0)	46	(100)	0	(0)
Slovenia	**6-17**	0	(0)	26	(100)	0	(0)
(L)	**≥18**	0	(0)	15	(100)	0	(0)
Spain	**6-17**	1	(0.44)	208	(91.23)	19	(8.33)
(L)	**≥18**	3	(1.24)	220	(91.29)	18	(7.47)
Sweden	**6-17**	92	(100)	-		-	
(L&R ≥ 6 yrs)	**≥18**	152	(100)	-		-	
Switzerland	**6-17**	1	(0.80)	122	(97.60)	2	(1.60)
(L ≥ 6 yrs)	**≥18**	1	(0.48)	148	(70.81)	60	(28.71)
Turkey	**6-17**	0	(0)	117	(100)	0	(0)
	≥18	0	(0)	25	(100)	0	(0)
Ukraine	**6-17**	1	(3.45)	28	(96.55)	0	(0)
	≥18	0	(0)	9	(100)	0	(0)
United Kingdom	**6-17**	0	(0.00)	1494	(96.51)	54	(3.49)
	≥18	0	(0.00)	2146	(86.32)	340	(13.68)
Total	**6-17**	500	(9.19)	4022	(73.91)	920	(16.91)
	≥18	736	(9.94)	4816	(65.02)	1855	(25.04)

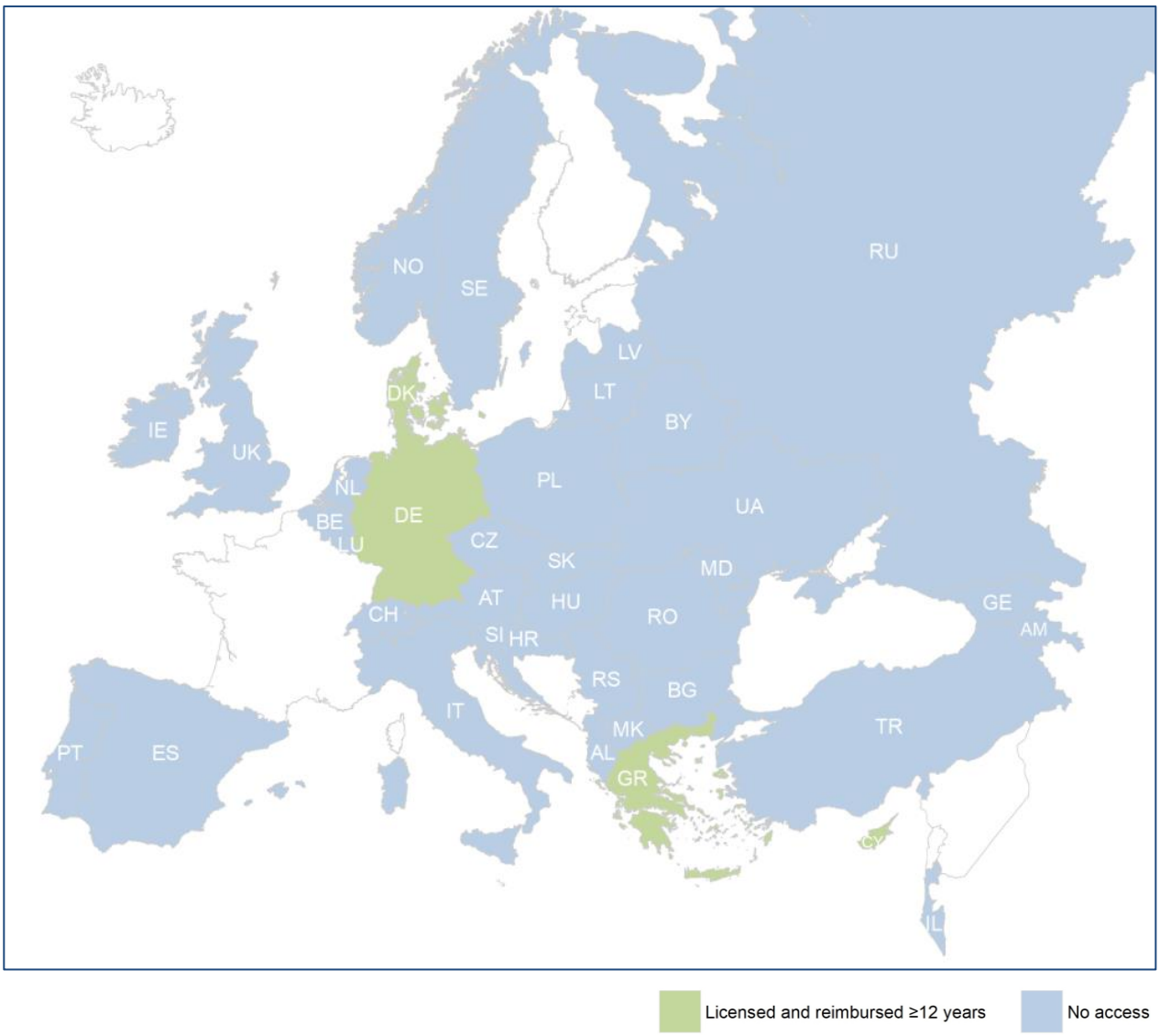
Note: L = licensed, R = reimbursed.

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

In table 7.11 we indicated in which countries Lumacaftor/Ivacaftor is licensed, or licensed and reimbursed.

The eligibility criteria for Lumacaftor/Ivacaftor in 2018 are:
The patient must be 6 years or older and have the mutation F508del/F508del.

Figure 7.16 Use of Tezacaftor/Ivacaftor in all eligible patients seen in 2018 who had never had a transplant, by country and age group.



In this graph we highlighted in light green the countries where Tezacaftor/Ivacaftor is licensed and reimbursed from 12 years.

Table 7.12 Use of Tezacaftor/Ivacaftor in all eligible patients seen in 2018 who had never had a transplant, by country and age group.

Country	Age at follow-up (years)	Use of Tezacaftor/Ivacaftor this year number (%)					
		Missing/ unknown		No		Yes	
Albania	**12-17**	0	(0)	22	(100)	0	(0)
	≥18	0	(0)	1	(100)	0	(0)
Austria	**12-17**	0	(0)	56	(100)	0	(0)
(L≥ 12 yrs)	**≥18**	1	(0.58)	171	(98.84)	1	(0.58)
Belarus	**12-17**	0	(0)	13	(100)	0	(0)
	≥18	0	(0)	1	(100)	0	(0)
Belgium	**12-17**	0	(0.00)	81	(97.59)	2	(2.41)
(L≥ 12 yrs)	**≥18**	2	(0.58)	338	(98.26)	4	(1.16)
Bulgaria	**12-17**	0	(0)	16	(100)	0	(0)
(L≥ 12 yrs)	**≥18**	0	(0)	30	(100)	0	(0)
Croatia	**12-17**	0	(0)	15	(100)	0	(0)
(L≥ 12 yrs)	**≥18**	0	(0)	28	(100)	0	(0)
Cyprus	**12-17**	0	(0)	2	(100)	0	(0)
(L&R ≥ 12 yrs)	**≥18**	0	(0)	2	(100)	0	(0)
Czech Republic	**12-17**	0	(0)	56	(100)	0	(0)
(L ≥ 12 yrs)	**≥18**	0	(0)	124	(100)	0	(0)
Denmark	**12-17**	0	(0.00)	52	(98.11)	1	(1.89)
(L&R ≥ 12 yrs)	**≥18**	0	(0.00)	154	(87.50)	22	(12.50)
Germany	**12-17**	2	(0.45)	436	(97.76)	8	(1.79)
(L&R ≥ 12 yrs)	**≥18**	23	(1.37)	1582	(94.05)	77	(4.58)
Greece	**12-17**	0	(0)	44	(100)	0	(0)
(L&R ≥ 12 yrs)	**≥18**	0	(0)	107	(100)	0	(0)
Hungary	**12-17**	41	(100)	-		-	
(L≥ 12 yrs)	**≥18**	66	(100)	-		-	
Ireland	**12-17**	1	(0.98)	101	(99.02)	0	(0.00)
(L≥ 12 yrs)	**≥18**	3	(0.89)	328	(97.33)	6	(1.78)
Israel	**12-17**	0	(0)	10	(100)	0	(0)
	≥18	0	(0.00)	46	(85.19)	8	(14.81)
Italy	**12-17**	180	(91.37)	17	(8.63)	0	(0.00)
(L≥ 12 yrs)	**≥18**	717	(85.46)	121	(14.42)	1	(0.12)
Latvia	**12-17**	0	(0)	6	(100)	0	(0)
(L≥ 12 yrs)	**≥18**	0	(0)	6	(100)	0	(0)
Lithuania	**12-17**	0	(0)	1	(100)	0	(0)
(L ≥ 12 yrs)	**≥18**	0	(0)	11	(100)	0	(0)
Luxembourg	**12-17**	0	(0)	1	(100)	0	(0)
(L≥ 12 yrs)	**≥18**	0	(0)	12	(100)	0	(0)
Rep of Moldova	**12-17**	0	(0)	3	(100)	0	(0)
	≥18	0	(0)	2	(100)	0	(0)
The Netherlands	**12-17**	0	(0.00)	127	(94.07)	8	(5.93)
(L≥ 12 yrs)	**≥18**	0	(0.00)	526	(97.05)	16	(2.95)
North Macedonia	**12-17**	0	(0)	17	(100)	0	(0)
	≥18	0	(0)	14	(100)	0	(0)
Norway	**12-17**	0	(0)	13	(100)	0	(0)
	≥18	1	(1.37)	72	(98.63)	0	(0.00)
Poland	**12-17**	5	(5.56)	85	(94.44)	0	(0.00)
(L≥ 12 yrs)	**≥18**	7	(6.36)	103	(93.64)	0	(0.00)
Portugal	**12-17**	2	(6.25)	30	(93.75)	0	(0.00)
(L≥ 12 yrs)	**≥18**	1	(1.85)	53	(98.15)	0	(0.00)
Romania	**12-17**	0	(0)	24	(100)	0	(0)
(L≥ 12 yrs)	**≥18**	0	(0)	4	(100)	0	(0)
Russian Federation	**12-17**	2	(1.11)	178	(98.89)	0	(0.00)
	≥18	2	(0.88)	223	(98.68)	1	(0.44)

[table 7.12 continued]

Country	Age at follow-up (years)	Use of Tezacaftor/Ivacaftor this year number (%)					
		Missing/ unknown		No		Yes	
Serbia	**12-17**	0	(0)	24	(100)	0	(0)
	≥18	0	(0)	28	(100)	0	(0)
Slovak Republic	**12-17**	0	(0)	19	(100)	0	(0)
(L≥ 12 yrs)	**≥18**	1	(1.56)	62	(96.88)	1	(1.56)
Slovenia	**12-17**	0	(0)	7	(100)	0	(0)
(L≥ 12 yrs)	**≥18**	0	(0)	15	(100)	0	(0)
Spain	**12-17**	1	(0.70)	134	(94.37)	7	(4.93)
(L≥ 12 yrs)	**≥18**	3	(0.95)	271	(85.49)	43	(13.56)
Sweden	**12-17**	39	(100)	-		-	
(L≥ 12 yrs)	**≥18**	177	(100)	-		-	
Switzerland	**12-17**	0	(0.00)	54	(93.10)	4	(6.90)
	≥18	1	(0.45)	200	(90.91)	19	(8.64)
Turkey	**12-17**	0	(0)	53	(100)	0	(0)
	≥18	0	(0)	33	(100)	0	(0)
Ukraine	**12-17**	1	(6.25)	15	(93.75)	0	(0.00)
	≥18	0	(0)	11	(100)	0	(0)
United Kingdom	**12-17**	0	(0.00)	763	(99.74)	2	(0.26)
	≥18	0	(0.00)	2809	(99.22)	22	(0.78)
Total	**12-17**	274	(9.85)	2475	(89.00)	32	(1.15)
	≥18	1005	(11.53)	7488	(85.93)	221	(2.54)

Note: L = licensed, R = reimbursed.

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

In table 7.11 we indicated in which countries Tezacaftor/Ivacaftor is licensed, or licensed and reimbursed.

The eligibility criteria for Tezacaftor/Ivacaftor in 2018 are:
The patients must be aged 12 years or more and be F508del homozygote or F508del heterozygote plus one of the following mutations: P67L, R117C, L206W, R352Q, A455E, D579G, 711+3A→G, S945L, S977F, R1070W, D1152H, 2789+5G→A, 3272-26A→G, 3849+10kbC→T.

8. Transplantation

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

Table 8.1 Number of patients living in 2018 with transplanted lungs, by age and sex.

Age	Males	Females	Total	Transplants carried out in 2018
5-9	0	2	2	1
10-14	6	13	19	5
15-19	21	36	57	10
20-24	69	110	179	40
25-29	127	145	272	37
30-34	157	167	324	39
35-39	183	175	358	49
40-44	119	126	245	14
45+	212	148	360	15
Total	894	922	1816	210

This table shows the number of patients alive in 2018 who have had a lung transplant at some time in their life, by age group, as well as the number of patients transplanted during 2018.

Table 8.2 Number of patients living in 2018 with transplanted liver, by age and sex.

Age	Males	Females	Total	Transplants carried out In 2018
5-9	3	0	3	0
10-14	11	9	20	6
15-19	28	15	43	7
20-24	27	10	37	2
25-29	24	15	39	1
30-34	26	8	34	2
35-39	16	6	22	0
40-44	8	4	12	0
45+	10	5	15	0
Total	153	72	225	18

This table shows the number of patients alive in 2018 who have had a liver transplant at some time in their life, by age group, as well as the number of patients transplanted during 2018.

Table 8.3 Number of patients living in 2018 with transplanted kidneys, by age and sex.

Age	Males	Females	Total	Transplants carried out In 2018
20-24	2	0	2	1
25-29	0	3	3	1
30-34	2	7	9	3
35-39	6	5	11	1
40-44	8	9	17	2
45+	10	4	14	0
Total	28	28	56	8

Note: Denmark does not collect information on kidney transplant.

This table shows the number of patients alive in 2018 who have had a kidney transplant at some time in their life, by age group, as well as the number of patients transplanted during 2018.

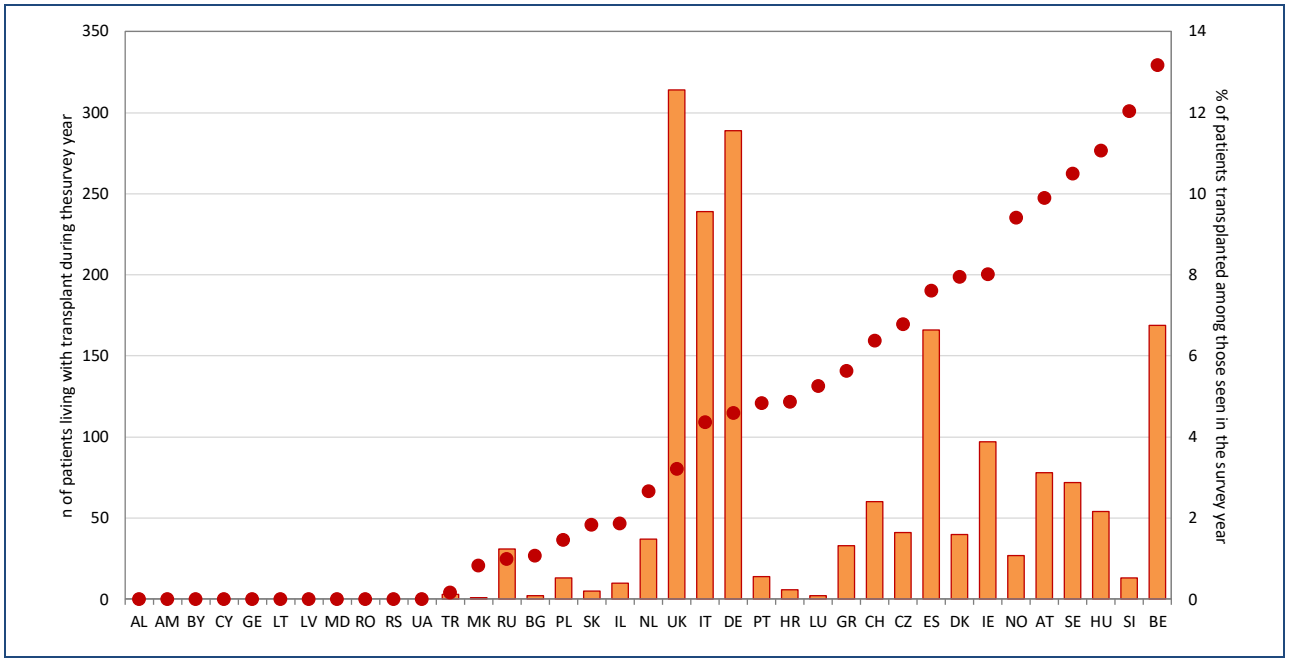
Table 8.4 Number of patients living in 2018 with other transplanted organs (not lung, liver, kidney), by age and sex.

Age	Males	Females	Total	Other Transplants carried out in 2018
5-9	1	0	1	0
10-14	0	0	0	0
15-19	2	1	3	1
20-24	2	4	6	0
25-29	1	1	2	0
30-34	2	1	3	0
35-39	1	0	1	0
40-44	2	2	4	0
45+	6	0	6	0
Total	17	9	26	1

Note: Denmark does not collect information on other organ transplant.

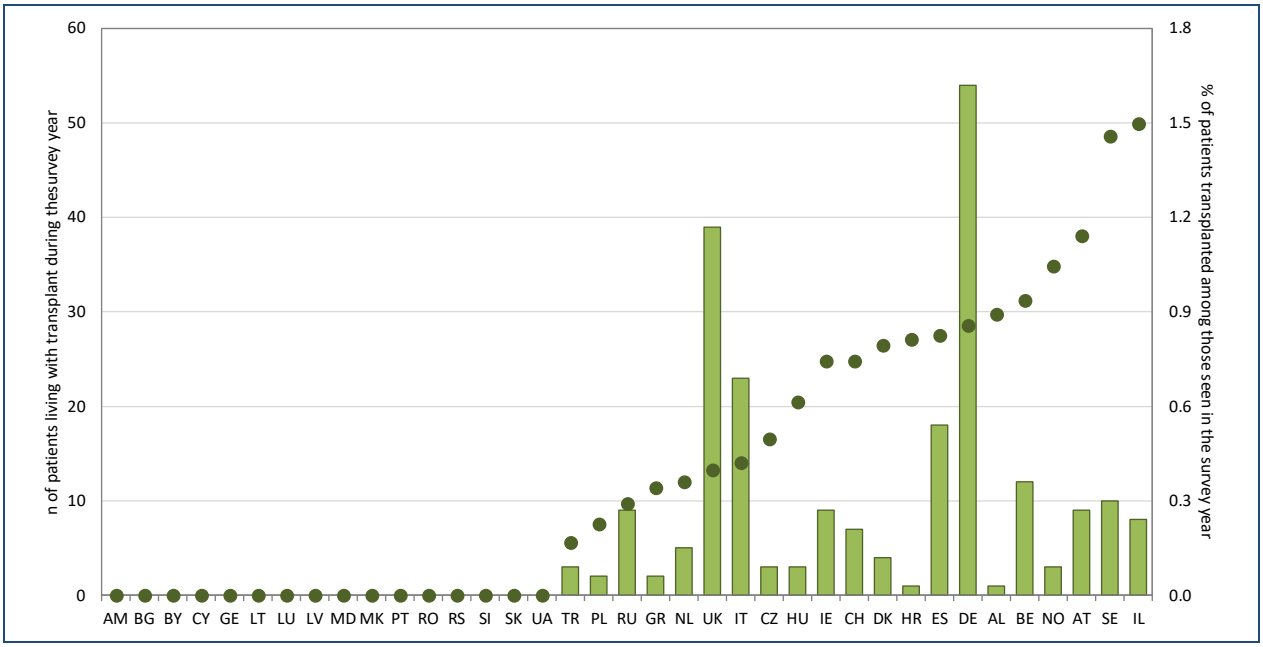
This table shows the number of patients alive in 2018 who have had an organ transplant different from lung, liver or kidney at some time in their life, by age group, as well as the number of patients transplanted during 2018.

Figure 8.1 Number of patients living in 2018 with transplanted lungs, by country.



This graph shows the number of patients alive at 31/12/2018 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 transplant lungs in 2018 among the patients that were seen in 2018.

Figure 8.2 Number of patients living in 2018 with transplanted liver, by country.



This graph shows the number of patients alive at 31/12/2018 who have had a liver transplant (green bars) at some point in their life. The dark green dots (right axis) show the percentage of patients that are living with transplanted liver in 2018 among the patients that were seen in 2018. Note that on the vertical axis the number of patients who had a liver transplant is much lower than the number who had a lung transplant. The main reason for this is that liver disease is only found in a subset of CF patients, whereas lung disease affects almost all patients.

9. Mortality

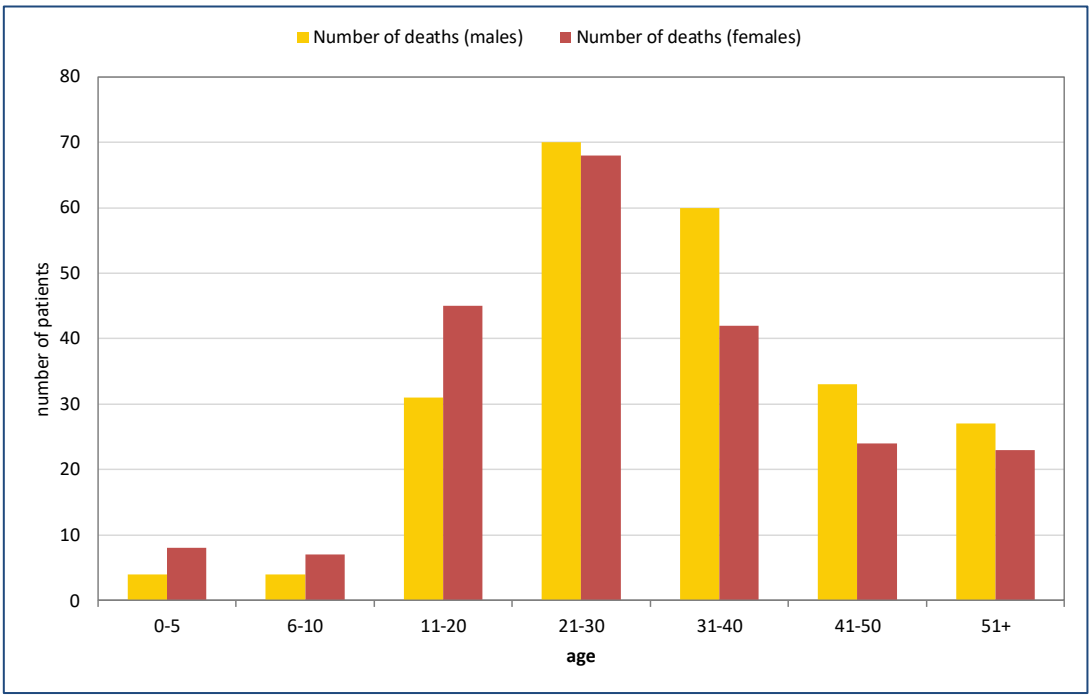
Table 9.1 Number of deaths in 2018, by age and sex.

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	4	1.75	8	3.69	12	2.69
6-10	4	1.75	7	3.23	11	2.47
11-20	31	13.54	45	20.7	76	17.04
21-30	70	30.57	68	31.3	138	30.94
31-40	60	26.2	42	19.4	102	22.87
41-50	33	14.41	24	11.1	57	12.78
51+	27	11.79	23	10.6	50	11.21
Total	229	100	217	100	446	100

Note: For the United Kingdom, all patients with a confirmed diagnosis of CF were included (N=10,509). The total number of patients presented is 41,933.

This table shows the number of deaths in 2018 by age group and sex. Death in small children is very rare, and the most frequent range of age at death for both sexes is 21-30 years. It is possible that the number of deceased patients is under reported because some of the patients were not seen at the centre during the year, and therefore the information may not have been recorded.

Figure 9.1 Age at death distribution of patients deceased in 2018, by sex.



This graph shows the distribution of age at death of patients who died in 2018, separately by males (yellow) and females (red).

Table 9.2 Cause of death distribution of deaths in 2018.

Cause of death	Number of deaths	Percentage of all deaths
Respiratory disease	295	66.14
Transplantation related	36	8.07
Non-CF related	24	5.38
Other-CF related	19	4.26
Cancer	18	4.04
Liver-GI related	15	3.36
Suicide	6	1.35
Trauma	1	0.22
Missing/Unknown	32	7.17
Total	446	

Note: For the United Kingdom, all patients with a confirmed diagnosis of CF were included (N=10,509). The total number of patients presented is 41,933.
Note: Germany and the United Kingdom record Cause of death as “cardio/respiratory”.

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

Publications

The ECFSPR database is a useful source for research and the data is actively used. Applications for data are conscientiously handled in accordance with the ECFSPR guidelines, which you will find here.

In the period 2011 – 2020 we received 85 applications to use Registry data. The majority of these requests originated from researchers (82%) from the European Cystic Fibrosis Society and other institutes, and 18% of the applications derived from Industry.

Several of these research projects have resulted in publications and other publications are in the pipeline. Articles published in the period 2019 – 2020 using Registry data are:

- Dornase alfa and rate of lung function decline in European patients with cystic fibrosis: a retrospective registry cohort study.
  McKone EF, Jackson AD, Fletcher F, Kirwan L. J Cyst Fibros. 2020 August 21. DOI: https://doi.org/10.1016/j.jcf.2020.08.004.

- The c.3140-26A>G Variant of the CFTR Gene in homozygous state causes mild cystic fibrosis. Overview of longitudinal clinical data of the patient managed in our centre and review of the literature. Kotnik Pirš A, Krivec U, Trebušak Podkrajšek K. Acta Chim. Slov. 2020; 67: 666–673. DOI: https://dx.doi.org/10.17344/acsi.2019.5677.

- Cystic Fibrosis Related Diabetes in Europe: Prevalence, Risk Factors and Outcome.
  Olesen HV, et al on behalf of the ECFSPR Steering Group. J Cyst Fibros. 2019 Oct. 31.
  DOI: https://doi.org/10.1016/j.jcf.2019.10.009

- Changing Epidemiology of the respiratory bacteriology of patients with cystic fibrosis from the European Cystic Fibrosis Society Patient Registry.
  Hatziagorou E, Orenti A, Drevinek P, Kashirskaya N, Mei-Zahav M, de Boeck K, on behalf of the ECFSPR. J Cyst Fibros. 2019 Sept. 3. DOI: https://doi.org/10.1016/j.jcf.2019.08.006.

- Characteristics of Cystic Fibrosis-related diabetes: Data from two different sources, the European Cystic Fibrosis Society Patient registry and German/Austrian diabetes prospective follow-up registry. Prinz N et al on behalf of the ECFS Patient Registry and the DPV Registry. Pediatr Diabetes. 2019;20(3):255-262. DOI: https://doi.org/10.1111/pedi.12831.

A complete overview of publications using ECFSPR data is available here.

The following abstracts were accepted in 2019-2020:

- Distribution of cystic fibrosis patients not eligible to studied CFTR modulators in Europe.
  Zolin A, Orenti A, Barbier A, van Rens J, Naehrlich L on behalf of the ECFSPR.
  ECFSPR. J Cyst Fibros. 2020; 19 (S22). DOI: https://doi.org/10.1016/S1569-1993(20)30236-8.

- Epidemiology of European adults with cystic fibrosis.
  Zolin A, Naehrlich L, Fox A, Krasnyk M, Orenti A, van Rens J, on behalf of the ECFSPR.
  ECFSPR. J Cyst Fibros. 2020; 19 (S35). DOI: https://doi.org/10.1016/S1569-1993(20)30276-9.
  1st Adult CF International Workshop, 5-6 September 2019, Milan. European Respiratory & Pulmonary Diseases 2019; 5 (1;S2).

- The European Cystic Fibrosis Society Patient Registry's Data Quality programme.
  Van Rens J, Fox A, Krasnyk M, Orenti A, Zolin A, Naehrlich L on behalf of the ECFSPR.
  10th European Conference on Rare Diseases & Orphan Products, 14-15 May 2020 (online).
  Theme 6, p194.

An overview of the approved applications for data, not associated with a publication, can be found here.

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.
For pre-natal diagnoses, we set age at diagnosis equal to 0.
We checked for outliers and, whenever possible, we corrected the values according to the instructions of the national registries'/individual centres. If, after the data quality controls, aberrant values were still present in the database, we set them to missing.

Reference populations used for computing z-scores

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

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

Reference populations used for computing FEV_1 predicted values

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

Software used for data management and statistical analyses

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

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

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

Variables

Demographics

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

Diagnosis

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

Genotype

First mutation
Second mutation

Therapy

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

Complications

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

Microbiology

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

Follow-up

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

Transplant

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

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

■ Marked in blue are the variables that have been collected, for the first time, in the 2018 data and are presented in this report.

Inclusion criteria

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

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

Definitions used by the EFCSPR

SWEAT TEST

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

2. a. The sweat sample should be processed immediately after sweat test collection;
   b. Chloride concentration measurement is the preferred analysis;
   c. Chloride value: the Chloride value should be measured in millimols per litre (mmol/L);
   if duplicate tests were completed on the same day, **the highest positive value** should be considered;
   d. A sweat chloride value > 59 mmol/L is consistent with a diagnosis of CF;
   e. A sweat chloride value < 30 mmol/L makes the diagnosis of CF unlikely (However, specific CF causing mutations can be associated with a sweat test value of below 30 mmol/L).

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

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

References:

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

NUTRITION

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

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

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

References:

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

SPIROMETRY

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

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

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

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

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

References:

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

CHRONIC INFECTION IN THE LOWER AIRWAYS

1. Chronic Pseudomonas aeruginosa infection:

    A patient should be considered chronically infected if the modified Leeds criteria are met (a) below, and/or anti-pseudomonas antibodies are detected (b) below.

A patient should be defined as chronically infected if he/she fulfils the criteria now, or has done so in recent years, and the physician has no reason to think that the status has changed:

a. Modified Leeds criteria, chronic infection: >50% of the samples (sputum/other) collected during the last 12 months should be positive; at least 4 samples collected.
b. Significantly raised anti-pseudomonas antibodies according to local laboratories.

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

References:

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

ALLERGIC BRONCHOPULMONARY ASPERGILLOSIS (ABPA)

Diagnostic criteria:

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

References:

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

LIVER DISEASE

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

These definitions discriminate patients with severe liver disease (with portal hypertension) from milder cases (cirrhosis without portal hypertension).

Cirrhosis with Hypertension: scarring of the liver related to underlying CF, typically in a biliary pattern. Severe liver disease may include portal hypertension and/or hypersplenism;

Cirrhosis without Hypertension: scarring of the liver related to underlying CF;

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

PANCREATIC STATUS

To define pancreatic insufficiency two determinations are mandatory:

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

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

For the ECFSPR, pancreatic status is assessed as follows:
Pancreatic insufficiency: Faecal elastase <200 µg/g (twice), and faecal fat high* (twice);
Pancreatic sufficiency: Faecal elastase ≥200 µg/g (twice) and Faecal fat normal* (twice).

*see definition above.

References:

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

SALT LOSS SYNDROME

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

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

TRANSPLANTATION

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