German Cystic Fibrosis Registry

Annual Report | 2022

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Foreword

Prof. Dr. med. Lutz Nährlich

Medical Director
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Cystic Fibrosis
Registry

In addition to the current state of health of people with cystic fibrosis in Germany, the German Cystic Fibrosis Registry presents the development of the last two decades in the 27th Annual Report for the year 2022 and also describes the development for selected birth cohorts.

The end of the coronavirus pandemic and an extension of approval for the highly effective CFTR modulator therapy (elexacaftor/tezacaftor/ivacaftor) for children between the age of 6 and 12 years characterized the year 2022. A CFTR modulator therapy was available for approx. 78 % of all people with cystic fibrosis in Germany in 2022 and was used in 86 % of this group. The Annual Report shows the medium-term effects on the health status of all people with cystic fibrosis. However, it should be borne in mind that the report does not make a distinction between people with cystic fibrosis (CF) with and without CFTR modulator therapy and therefore only partly reflects the individual effect. In addition to the number of CF cases with annual follow-up data, we also show for the first time the number of CF cases without annual follow-up data but with a confirmed living status. Together they form the so-called live population of the Registry.

The stabilisation of the pulmonary function is continuing for all cohorts. 78 % of the 18- to 19-year-olds have a FEV1%pred of more than 80 % compared with 32 % in 2000. The rate of chronic Pseudomonas infection has fallen to 8.3 % in children and adolescents and to 49 % in adults. Resistance against various antibiotics (MRGN) continues to be a challenge with up to 18 % of all CF cases and chronic Pseudomonas infection in patients aged from 35 – 39 years. The relatively low rate of exacerbations treated with antibiotics (30 %) and hospitalisations (20 %) is another positive development. The decline in underweight particularly in adults, contrasts with an increase in overweight and adiposity prevalence.

The basic and indication therapy remained largely unchanged for the time being. All this underlines the continuing necessity for the close clinical care provided by the multidisciplinary cystic fibrosis team, in order to recognise and discuss the medium-term changes and challenges ahead on a collective basis. The German Cystic Fibrosis Registry can provide the (data) basis for this purpose.

This detailed overview of the health status of people with cystic fibrosis would not be possible without your trust in the German Cystic Fibrosis Registry. I would like to express my thanks to all the outpatient clinic teams and the people with cystic fibrosis who allow us to document and evaluate their data. Many thanks to all those involved in the data entry and analysis. My thanks also go to the Registry Work Group, the Axaris company (Ms Jaumann, Mr Müller, Mr Volk) and the data management team of the Interdisciplinary Center for Clinical Studies (IZKS) of the University of Mainz (Ms Wosniok, Ms Wollscheid, Ms Regenfuß, Mr Kronfeld, Mr Ruckes).

My special thanks go to Mr Burkhart of the Mukoviszidose Institut for his tireless efforts in project management.

Please keep supporting the Registry.

Collective description

The history data records of 6973 people with cystic fibrosis (pwCF) are included in the analyses of the demography, Cystic Fibrosis diagnosis, mortality and structure of care for the reporting year 2022. In addition, patients without history data were also included in the evaluations of new CF diagnoses and mortality (10 newly diagnosed patients and 7 deceased patients without history data in 2022).

All 364 transplant patients were excluded from the evaluations of nutritional status, pulmonary function, lung infections, complications and therapies, regardless of the type of transplant. This results in a number of 6609 patients for the analysis of the history data.

Further definitions apply to the various evaluation groups in some cases. These are described in more detail in the respective chapters.

A current declaration of consent is available for all evaluated patients, or they died before consent could be renewed. Patients who withdrew their consent before death were excluded from the mortality analyses.

The age of the patients was calculated in completed years at the end of the respective reporting year for patients not documented as deceased. The age at the time of death was calculated in completed years for patients who died during the reporting year. The age was calculated in completed years at the end of the reporting year for deceased patients for whom no date of death was documented. The age of newly diagnosed patients was calculated at the time of diagnosis.

The pulmonary function was calculated and reported using the reference values of the Global Lung Function Initiative (Quanjer et al; Eur Respir J 2012; 40: 1324).

The reference values according to the KiGGS study were used for calculation of the BMI percentiles for 2 – 18-year-old patients (Robert Koch Institute: Reference percentiles for anthropometric measures and blood pressure based on the German Health Interview and Examination Survey for Children and Adolescents (KiGGS); Berlin: RKI-Hausdruckerei; 2013).

Missing values were not taken into account for the calculation of the percentages.

Missing values were not taken into account for the calculation of the percentages.

The history data records are documented once a year in the so-called Level 1 documentation as the status for the entire calendar year or are aggregated from the visit- related data records of the so-called Level 2 documentation. The examination date with the best FEV1%pred and the relevant body measurements are selected as the examination time point in the reporting year for patients older than 6 years with a pulmonary function measurement.

The last body measurements available in the reporting year are used in the absence of an FEV1 value and for children younger than 6 years. A complication occurring at least once a year or a longterm therapy, microbiological indication or a chronic infection determine the intensity for the entire reporting year. If history data sets from several outpatient clinics are available for a patient, they are aggregated in a single data set for the annual data report in accordance with the above rules.

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Map of participating CF centers

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Figure 1: CF centers participating in 2022

The map shows all 85 CF centers participating in the Registry in 2022.
A list of all CF facilities and the reported patient numbers can be found on page 64.
Write to us if your facility is not represented: mburkhart@muko.info

01

Brief overview

	2000	2005	2010	2015	2020	2021	2022
Data status	30.05.23	30.05.23	30.05.23	30.05.23	30.05.23	30.05.23	30.05.23
Participating centers	78	83	81	89	88	87	85
Living Population [1, 2, 4]	4246	5205	5887	6656	7416	7364	–
Living patients with transplants[4]	67	174	291	454	559	507	413
Participating patients with annual data	3615	4693	5088	5751	6643	6809	6973
of these, transplant patients	43	122	201	293	363	375	364
Age in years; Median[1]	14	16	18	20	21	22	22
Proportion of adults (≥ 18 years) in %	36.3	45.9	51.3	56.7	58.6	59.4	60.1
Male patients in %	52.4	52	51.9	52.0	52.0	51.7	51.6
New diagnoses in the reporting year[1]	225	206	219	227	227	188	157
Age for new diagnoses in years; Median[1]	1.38	0.83	1.00	0.92	0.17	0.08	0.13
of these, diagnosis via newborn screening	1.8	3.4	3.7	10.6	58.1	65.4	72.0
Maternities in the reporting year	4	8	13	20	35	43	52
Paternities in the reporting year	1		7	6	15	11	15
Deaths in the reporting year [1, 4]	43	72	75	100	61	44	32
Deaths: % of all patients[1]	1.2	1.5	1.5	1.7	0.9	0.6	0.5
Age at death in years; median	21	26	28	31.5	35	42.5	37
(25^{th} – 75^{th} pctl)	(15 – 28)	(19 – 35)	(24 – 37)	(25 – 37)	(28 – 46)	(26 – 50)	(23 – 54)
Transplant patients in the reporting year[1]	9	36	33	33	33	12	8
Lung transplants [3]	7	32	29	30	29	10	4
Liver transplants [3]	2	3	4	6	5	2	3
Renal transplantation[3]	0	1	1	0	1	0	1
Pancreas transplantation[3]	0	0	0	1	0	0	1

Table 1: Brief overview of cystic fibrosis patients with follow-up data, valid informed consent and cystic fibrosis diagnosis in the reporting years 2000 – 2022 in Germany

[1] The information on living patients, new diagnoses, deaths and transplants also includes patients without follow-up data

[2] As a result of the anonymisation of patient data at the end of 2021, the number of living patients mentioned here will be reduced in subsequent years, as it is no longer possible to determine the living status of anonymised patients. The living population for 2022 will be published in the 2023.

[3] Multiple answers possible

[4] Data status 25.9.2023

Patient numbers development

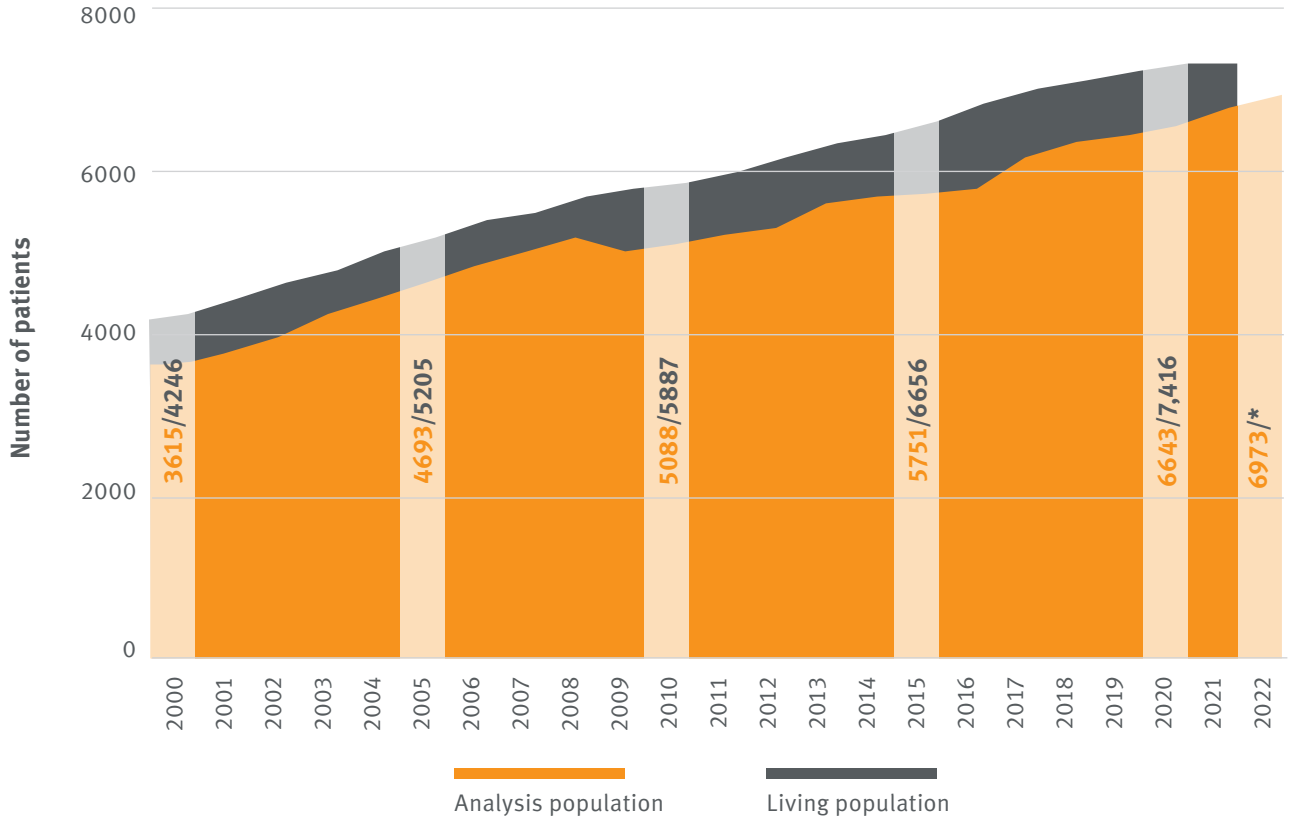

Figure 2: Number of pwCF documented in the registry 2000 – 2022, * the living population for 2022 will be published in the 2023 reporting year

Reporting year	Analysis population	Living population
2000	3615	4246
2001	3732	4406
2002	3939	4574
2003	4257	4798
2004	4436	4980
2005	4693	5205
2006	4848	5367
2007	4986	5521
2008	5225	5709
2009	5059	5765
2010	5088	5887
2011	5218	6014

Reporting year	Analysis population	Living population
2012	5359	6167
2013	5666	6355
2014	5736	6501
2015	5751	6656
2016	5804	6847
2017	6206	7041
2018	6405	7148
2019	6478	7273
2020	6643	7416
2021	6809	7364
2022	6973	–*

Table 2: Number of pwCF documented in the registry 2000 – 2022, * the living population for 2022 will be published in the 2023 reporting year

03

Age structure

The age structure calculations include all 6973 pwCF with annual data for 2022. The age of the patients was calculated in completed years at the end of the respective reporting year for patients not documented as deceased as well as for those without a date of death. The age at the time of death was calculated in completed years if the date of death was available.

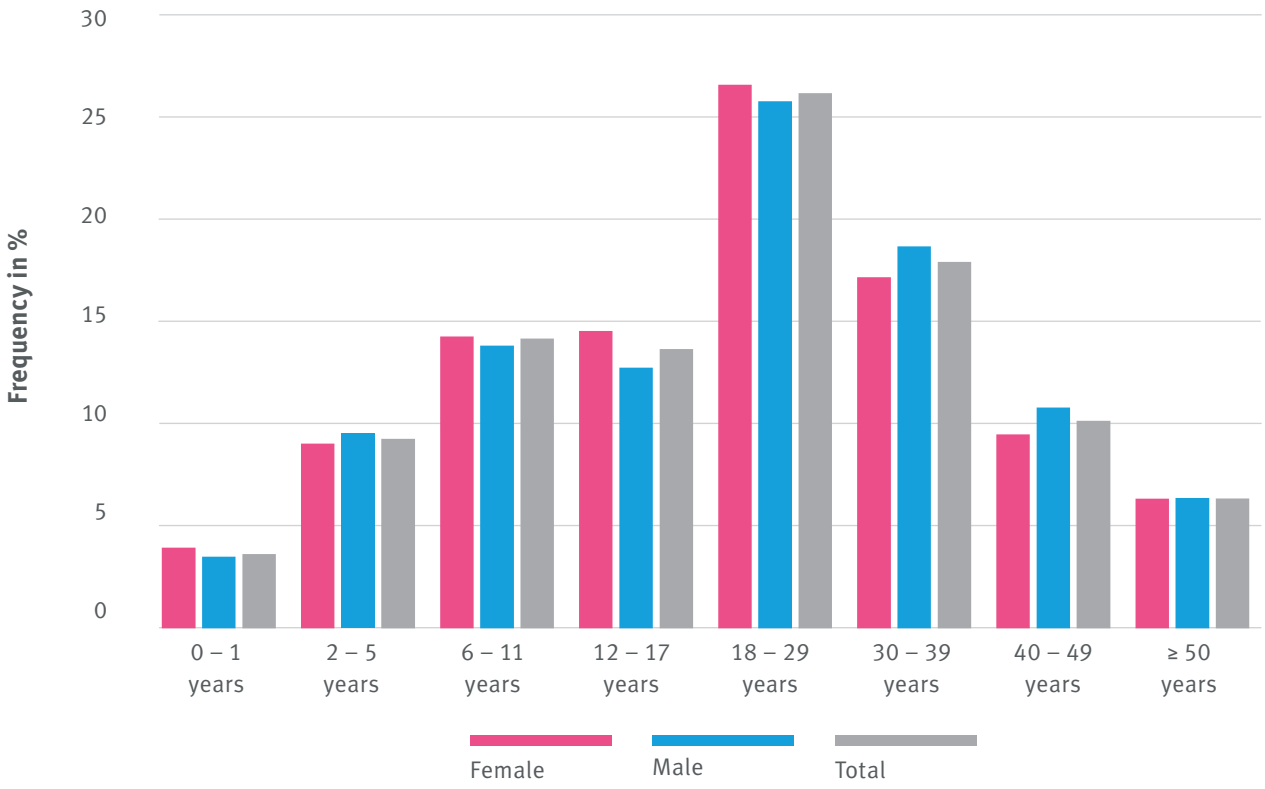

Figure 3: Age distribution of pwCF in 2022

	Male	Female	Total
Numbers	3598	3375	6973
Mean value (years)	23.8	23.2	23.5
Median (years)	23.0	22.0	22.0
Minimum (years)	0.0	0.0	0.0
Maximum(years)	82.0	85.0	85.0
25th percentile (years)	11.0	11.0	11.0
75th percentile (years)	34.0	33.0	34.0
Number < 18 years	1398	1384	2782
Number ≥ 18 years	2200	1991	4191

Table 3: Age distribution of pwCF in 2022

Age structure

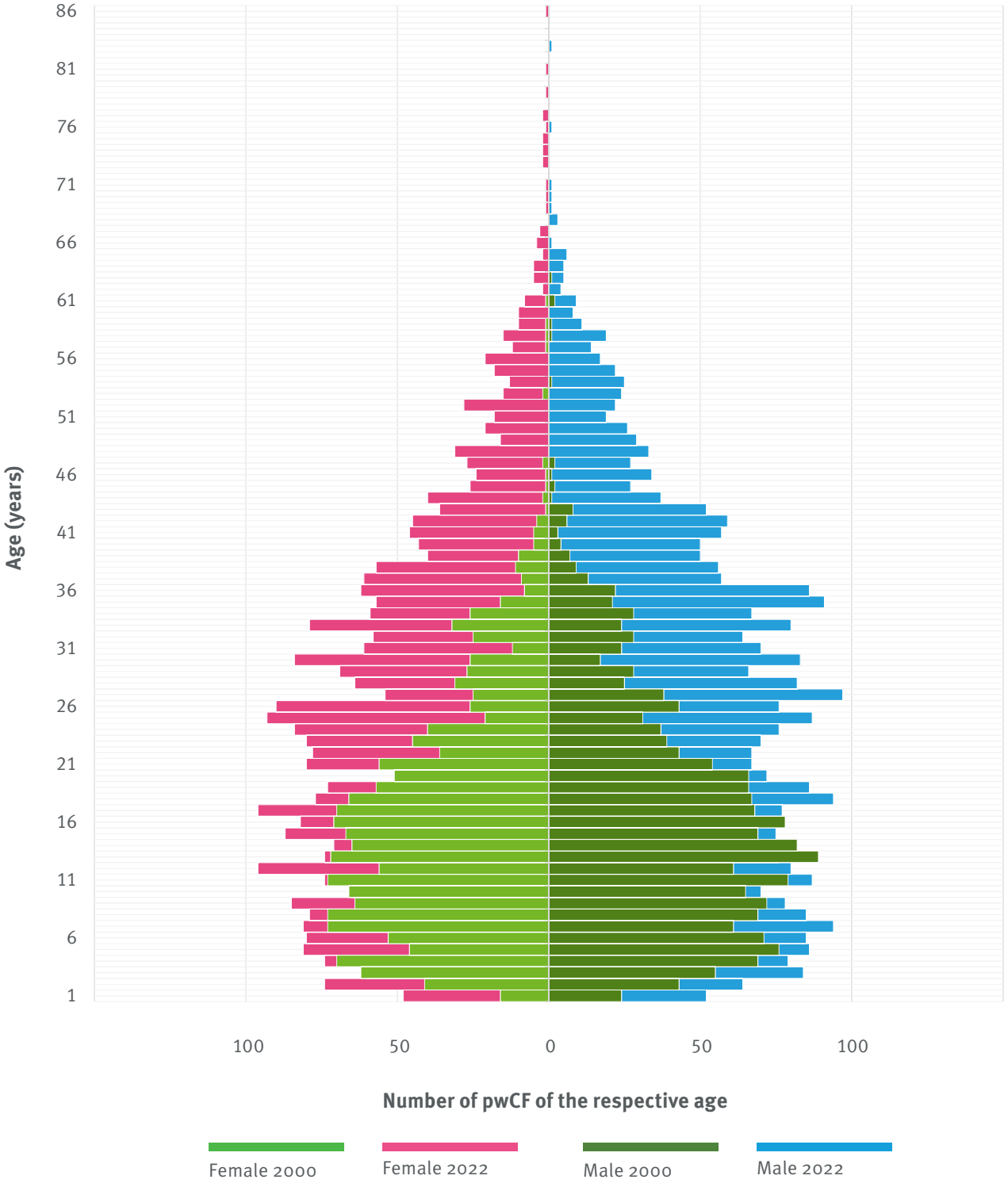

Figure 4: Age pyramid pwCF 2000 vs. 2022

03

Age structure

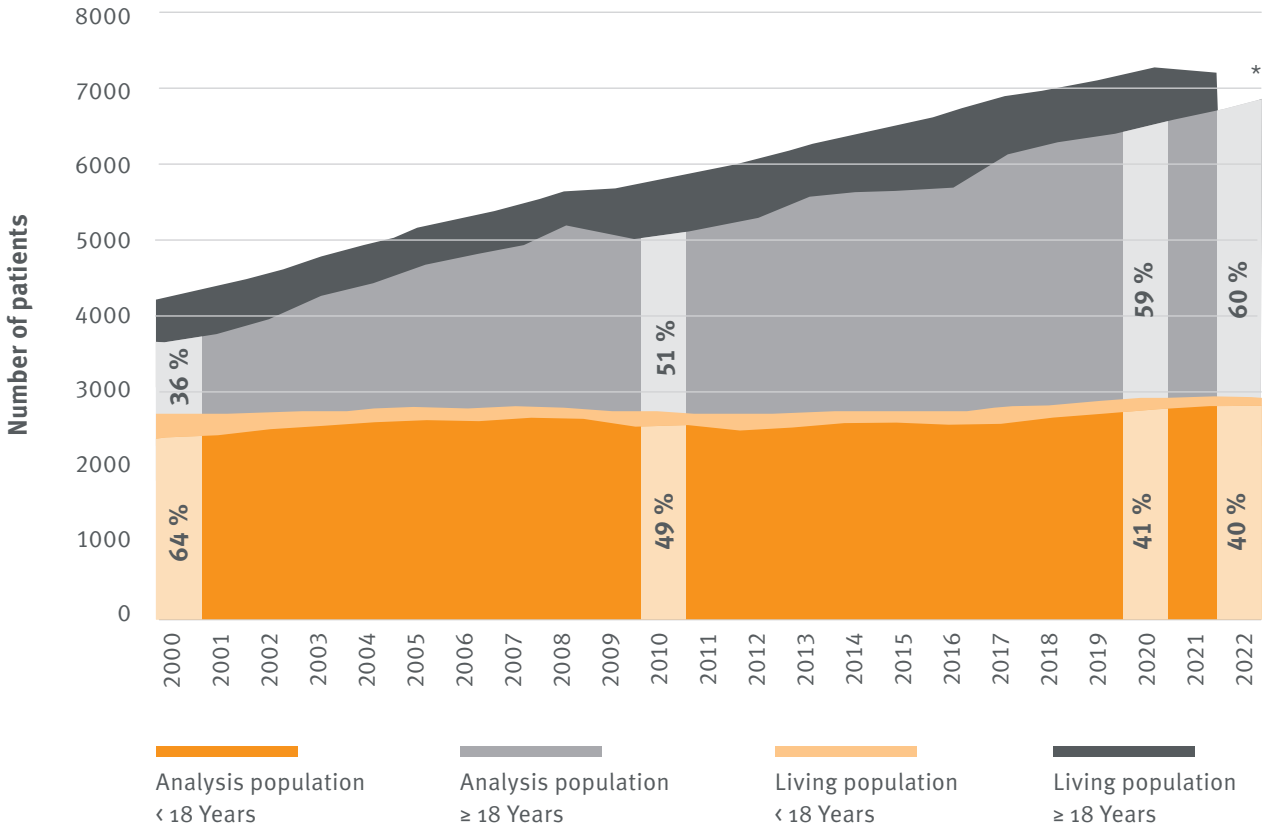

Figure 5: Development of the age distribution (‹ 18 vs. ≥ 18 years) for the years 2000 – 2022
* the live population for 2022 will be published in the 2023 reporting year

Age structure

Reporting year	Analysis population Total	Analysis population Frequency (%)		Living population Total	Living population frequency (%)	
		< 18Years	≥ 18 Years		< 18 Years	≥ 18 Years
2000	3615	63.7	36.3	4246	61.0	39.0
2001	3732	62.5	37.5	4406	59.3	40.7
2002	3939	61.4	38.6	4574	57.6	42.4
2003	4257	57.7	42.3	4798	55.5	44.5
2004	4436	56.5	43.5	4980	53.7	46.3
2005	4693	54.1	45.9	5205	52.0	48.0
2006	4848	52.1	47.9	5367	49.9	50.1
2007	4986	51.7	48.3	5521	49.3	50.7
2008	5225	49.1	50.9	5709	47.3	52.7
2009	5059	48.5	51.5	5765	45.8	54.2
2010	5088	48.7	51.3	5887	45.1	54.9
2011	5218	46.0	54.0	6014	43.6	56.4
2012	5359	45.6	54.4	6167	42.6	57.4
2013	5666	44.2	55.8	6355	41.6	58.4
2014	5736	43.8	56.2	6501	40.9	59.1
2015	5751	43.3	56.7	6656	39.7	60.3
2016	5804	43.1	56.9	6847	38.8	61.2
2017	6206	41.7	58.3	7041	38.5	61.5
2018	6405	41.3	58.7	7148	38.4	61.6
2019	6478	41.7	58.3	7273	38.4	61.6
2020	6643	41.4	58.6	7416	38.3	61.7
2021	6809	40.6	59.4	7364	38.7	61.3
2022	6973	39.9	60.1	–*	–*	–*

Table 4: Development of the age distribution (<18 vs ≥ 18 years) for the years 2000 – 2022
* the living population for 2022 will be published in the 2023 reporting year

04

Cystic fibrosis diagnosis

4a. Diagnoses in 2022

157 patients were diagnosed in 2022; annual data is available for 147 of these patients (89.3 %). The age distribution of all patients newly diagnosed in 2022 is shown in the following tables.

	N	Mean value	Median	Min	Max	25th percentile	75th percentile	Missing
Age in years	155	5.0	0.1	0.0	72.7	0.1	0.4	2

Table 5: Age at diagnosis of all pwCF diagnosed in 2022

Newborn screening was performed in 113 (72 %) of the pwCF diagnosed in 2022. 16 patients (8.1 %) had a meconium ileus. The age at diagnosis of the patients newly diagnosed via newborn screening in 2022 is as follows:

	N	Mean value	Median	Mini	Max	25th percentile	75th percentile	Missing
Age in days	112	37.5	29.5	0.0	266	19.5	45.5	1

Table 6: Age at diagnosis of all pwCF diagnosed via newborn screening in 2022

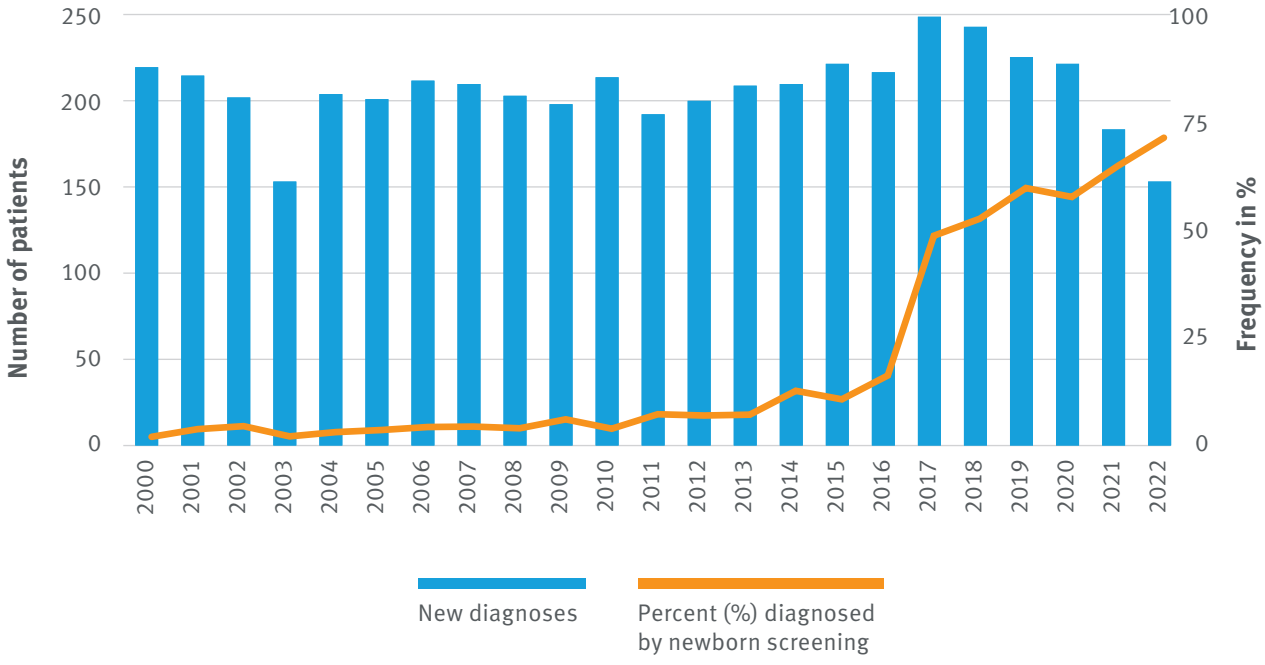

Figure 6: Number of new diagnoses and percentage frequency of pwCF diagnosed by newborn screening 2000 – 2022

Cystic fibrosis diagnosis

4b. Age at diagnosis (Status 2022)

The age distribution at diagnosis of the 6973 patients with follow-up data in 2022 is shown in the following figure and table below. No information on the date of diagnosis was available for 221 patients (3.1 %).

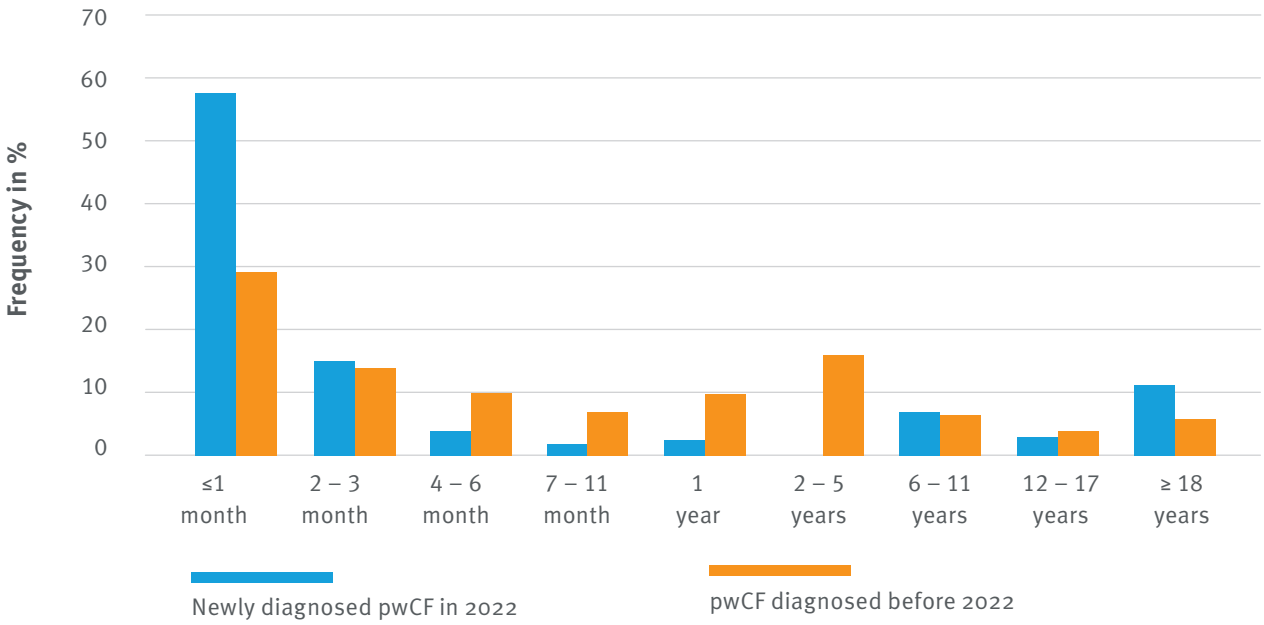

Figure 7: Age-related frequencies in diagnosed pwCF

Age at diagnoses	In 2022			Before 2022		
	Frequency	Percent	Accumulated percentages	Frequency	Percent	Accumulated percentages
≤ 1 month	91	58.7	58.7	1980	29.3	29.3
2 – 3 months	23	14.8	73.6	927	13.7	43.1
4 – 6 months	5	3.2	76.8	660	9.8	52.8
7 – 11 months	2	1.3	78.1	440	6.5	59.3
1 year	3	1.9	80.0	647	9.6	68.9
2 – 5 years	0	0.0	0.0	1086	16.1	85.0
6 – 11 years	10	6.5	86.5	415	6.2	91.2
12 – 17 years	4	2.6	89.0	221	3.3	94.4
≥ 18 years	17	11.0	100.0	376	5.6	100.0
Total	155	100.00	–	6752	100.00	–
Missing	2	–	–	221	–	–

Table 7: Age at diagnosis in diagnosed pwCF

04

Cystic fibrosis diagnosis

4c. Genotyping

Genotyping was available for 6,926 patients (99.3 %). Missing information was treated in the following presentation as "Mutation not identified" in the following presentation.

Mutation combinations	Frequency	Percent
F508del homozygot	3230	46.3
F508del heterozygous: Second mutation identified	2707	38.8
F508del heterozygous: Second mutation not identified	74	1.1
No verification of F508del: Both mutations identified	842	12.1
No verification of F508del: Only one mutation identified	33	0.5
No verification of F508del: No mutations identified	87	1.3
Total	6973	100.0

Table 8: Mutation combinations in pwCF in 2022

The frequencies for the individual alleles are shown below, whereby only those with an absolute frequency of at least 50 are shown individually:

First and second mutation	Number	Percent
F508del (p.Phe508del / c.1521_1523delCTT)	9241	66.8
G542X (p.Gly542X / c.1624G>T)	294	2.1
N1303K (p.Asn1303Lys / c.3909C>G)	276	2.0
R553X (p.Arg553X / c.1657C>T)	248	1.8
G551D (p.Gly551Asp / c.1652G>A)	221	1.6
CFTRdele2,3 (p.Ser18ArgfsX16 / c.54-5940_273+10250del21kb)	213	1.5
R347P (p.Arg347Pro / c.1040G>C)	192	1.4
3849+10kbC->T (c.3718-2477C>T)	146	1.1
1717-1G->A (c.1585-1G>A)	116	0.8
2789+5G->A (c.2657+5G>A)	111	0.8
W1282X (p.Trp1282X / c.3846G>A)	94	0.7
2183AA->G (p.Lys684SerfsX38 / c.2051_2052delAAinsG)	93	0.7
3272-26A->G (c.3140-26A>G)	69	0.5
R117H (p.Arg117His / c.350G>A)	69	0.5
2184insA (p.Gln685ThrfsX4 or p.Gln685Thrfs*4 / c.2052dupA or c.2052dup)	63	0.5
M1101K (p.Met1101Lys / c.3302T>A)	60	0.4
621+1G->T (c.489+1G>T)	57	0.4
R1162X (p.Arg1162X / c.3484C>T)	57	0.4
I336K (p.Ile336Lys / c.1007T>A)	56	0.4
1677delTA (p.Tyr515X / c.1545_1546delTA)	51	0.4
Other Mutation	1938	14.0
Unknown/Mutation not identified	180	1.3
Total	13845	100

Table 9: CFTR genotyping of pwCF 2022

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05

Nutritional status

5a. Children and adolescents under 18 years

All patients aged 2 – 17 years without transplantation with follow-up data 2022 (n=2550) were included. For 3 patients (0.1%), no information on nutritional status was available. For the assessment of nutritional status in children and adolescents, the BMI percentiles according to KiGGS were used. The age was calculated at the time of the physical calculated at the time of the physical examination.

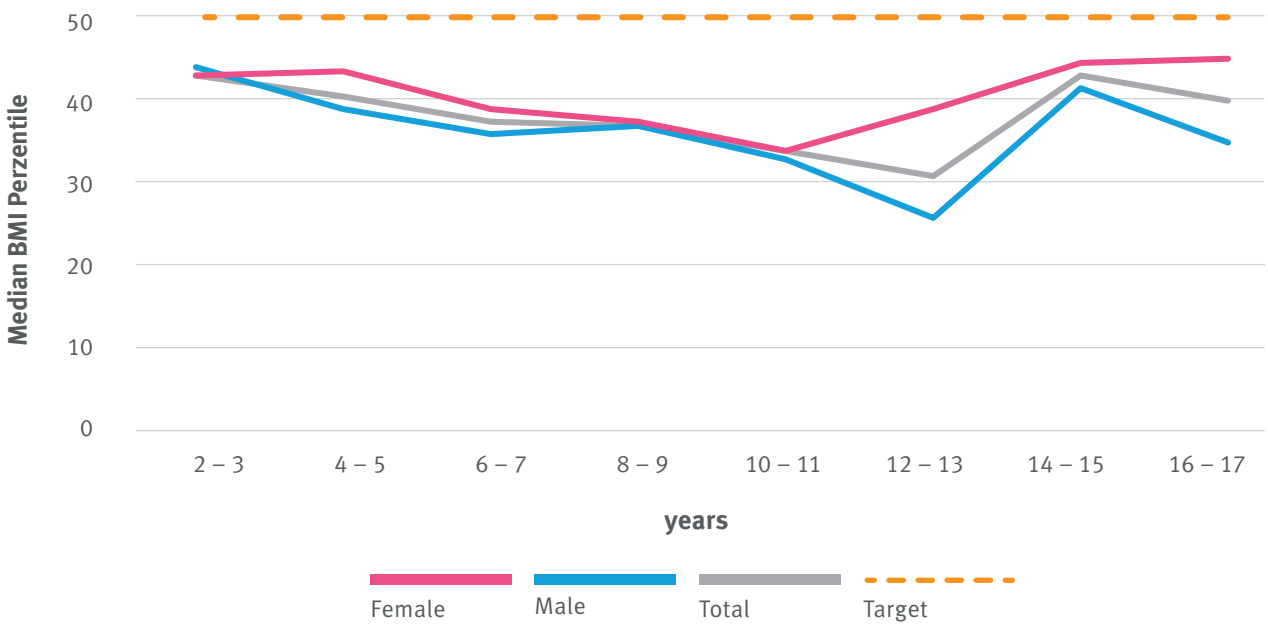

Figure 8: Median BMI percentiles of children and adolescents between 2 – 17 years 2022

Age (years)	Male			Female			Total		
	N	Median	25th - 75th pctl.	N	Median	25th - 75th pctl.	N	Median	25th - 75th pctl.
2 – 3	155	44.0	19.0 – 69.0	137	43.0	19.0 – 67.0	292	43.0	19.0 – 67.0
4 – 5	184	39.0	19.0 – 64.0	172	43.5	23.0 – 70.5	356	40.5	20.0 – 69.0
6 – 7	185	36.0	17.0 – 63.0	159	39.0	18.0 – 60.0	344	37.5	18.0 – 62.5
8 – 9	143	37.0	21.0 – 58.0	158	37.5	18.0 – 57.0	301	37.0	20.0 – 57.0
10 – 11	157	33.0	14.0 – 53.0	170	34.0	18.0 – 54.0	327	34.0	15.0 – 54.0
12 – 13	148	26.0	13.0 – 52.0	149	39.0	18.0 – 66.0	297	31.0	16.0 – 61.0
14 – 15	132	41.5	22.0 – 61.5	170	44.5	23.0 – 67.0	302	43.0	23.0 – 65.0
16 – 17	174	35.0	16.0 – 66.0	154	45.0	21.0 – 68.0	328	40.0	19.0 – 66.0
Total	1278	36.0	17.0 – 61.0	1269	40.0	20.0 – 64.0	2547	38.0	19.0 – 63.0

Table 10: BMI percentiles of children and adolescents aged 2 – 17 years 2022

Nutritional status

5a. Children and adolescents under 18 years

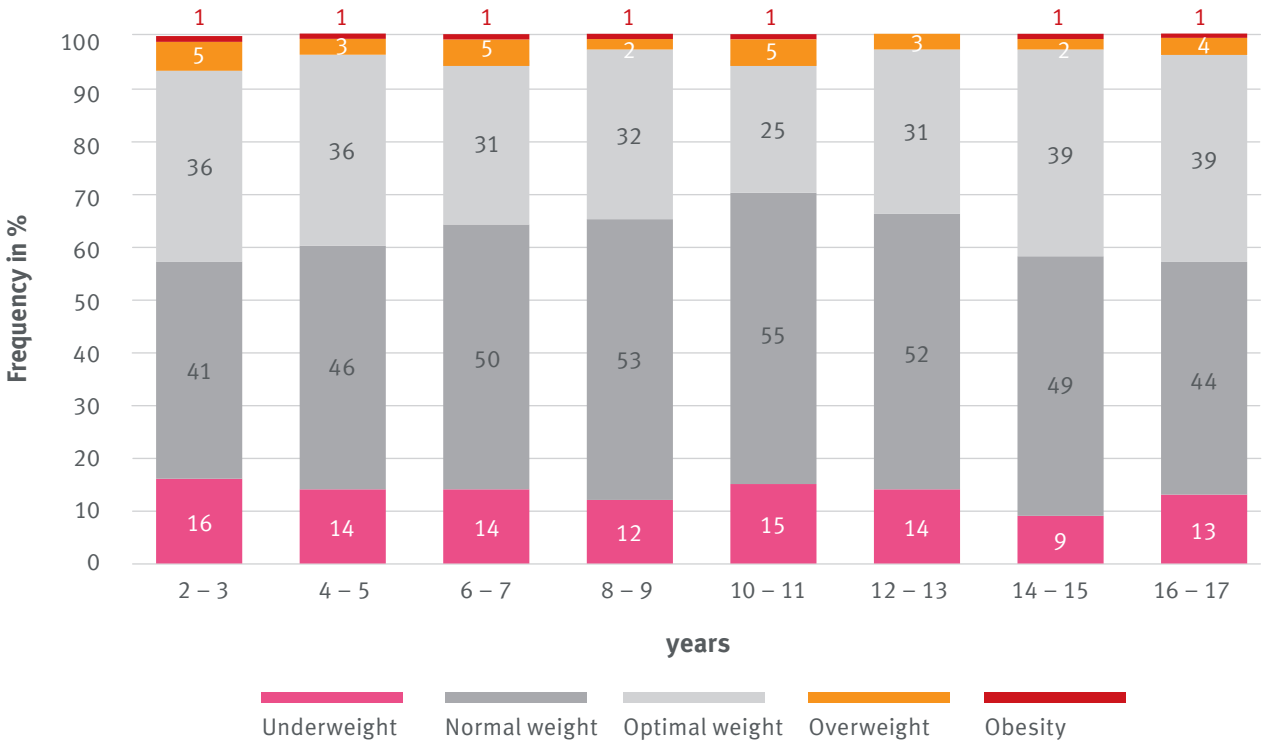

Figure 9: Weight categories of children and adolescents between 2 – 17 years 2022, underweight: BMI percentiles ‹ 10, Normal weight: BMI percentiles 10 – 49, Optimal weight: BMI percentiles 50 – 89, Overweight: BMI percentiles 90 – 96, Obesity: BMI percentiles ≥ 97

BMI Perzentile KIGGS	Male	Female	Total
Underweight	14.4	12.5	13.4
Normal weight	50.3	46.9	48.6
Optimal weight	31.1	35.9	33.5
Overweight	3.0	4.2	3.6
Obesity	1.3	0.6	0.9

Table 11: Weight categories of children and adolescents between 2– 17 years 2022, Underweight: BMI percentiles ‹ 10; Normal weight: BMI percentiles 10 – 49; optimal weight: BMI percentiles 50 – 89; overweight: BMI percentiles 90– 96; obese: BMI percentiles ≥ 97

Längensollgewicht	Male		Female		Total	
	0 – 12 months	13 – 24 months	0 – 12 months	13 – 24 months	0 – 12 months	13 – 24 months
Underweight	19.1	16.7	49.2	10.4	33.1	13.3
Normal weight	72.1	78.6	44.1	87.5	59.1	83.3
Overweight/Obesity	8.8	4.8	6.8	2.1	7.9	3.3

Table 12: Weight categories of children under 2 years of age (frequencies in %) according to weight-for-length (LSG) 2022, Underweight: LSG ‹ 90 %; normal weight: LSG 90 – 110 %; overweight/obese: LSG › 110 %

05

Nutritional status

5b. Adults 18 years and older

Adult patients without transplantation with follow-up data 2022 (n=3764) were included. For 23 patients (0.6 %) no information was available for the nutritional status. Age was calculated at the time of the physical examination was calculated.

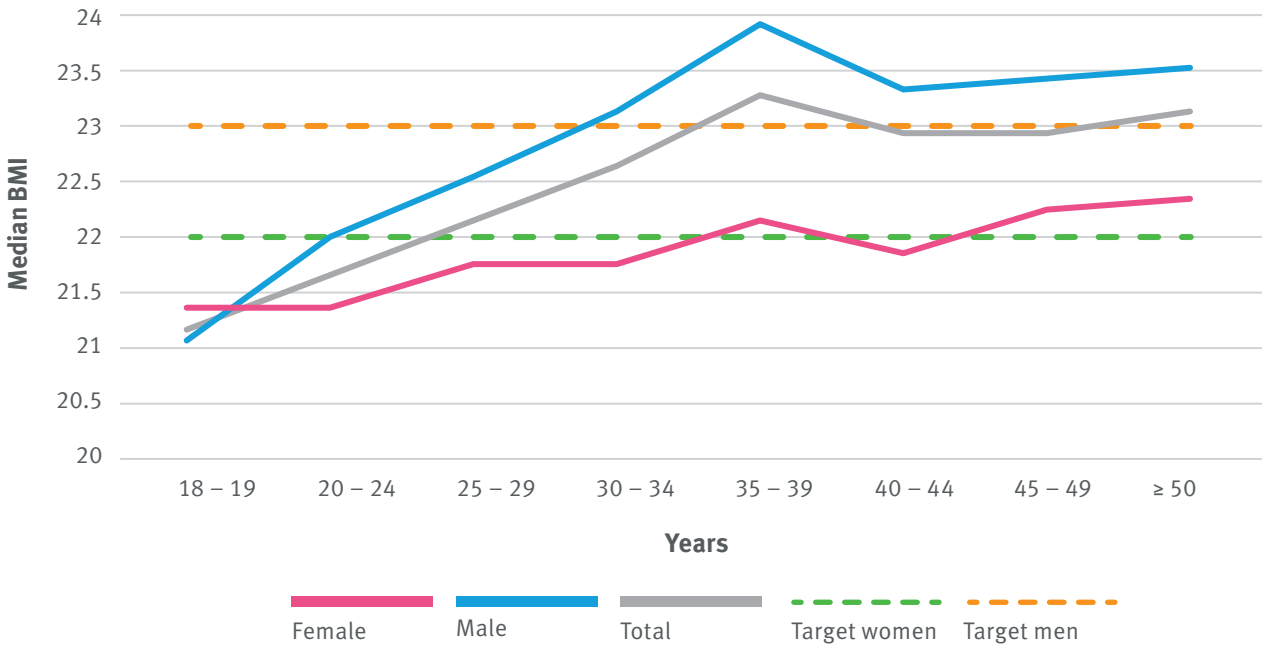

Figure 10: Median BMI of adults aged 18 and over 2022

Age (years)	Male			Female			Total		
	N	Median	25th - 75th pctl.	N	Median	25th - 75th pctl.	N	Median	25th - 75th pctl.
18 – 19	144	21.0	19.2 – 23.1	117	21.3	19.8 – 22.9	261	21.1	19.7 – 23.0
20 – 24	352	22.0	19.8 – 23.8	406	21.3	19.6 – 23.0	758	21.6	19.7 – 23.4
25 – 29	374	22.5	20.4 – 24.7	325	21.7	19.9 – 23.5	699	22.1	20.2 – 24.3
30 – 34	357	23.1	21.3 – 25.5	279	21.7	19.9 – 24.2	636	22.6	20.5 – 25.0
35 – 39	250	23.9	21.8 – 25.9	222	22.1	20.2 – 24.2	472	23.3	21.2 – 25.2
40 – 44	196	23.3	21.4 – 25.9	139	21.8	19.9 – 24.6	335	22.9	20.7 – 25.4
45 – 49	129	23.4	21.6 – 25.8	101	22.2	20.6 – 25.2	230	22.9	21.1 – 25.7
≥ 50	181	23.5	21.7 – 26.0	170	22.3	20.7 – 25.7	351	23.1	21.2 – 25.8
Total	1983	22.9	20.9 – 25.0	1759	21.7	19.9 – 23.9	3742	22.3	20.3 – 24.6

Table 13: BMI of adults aged 18 and over 2022

Nutritional status

5b. Adults 18 years and older

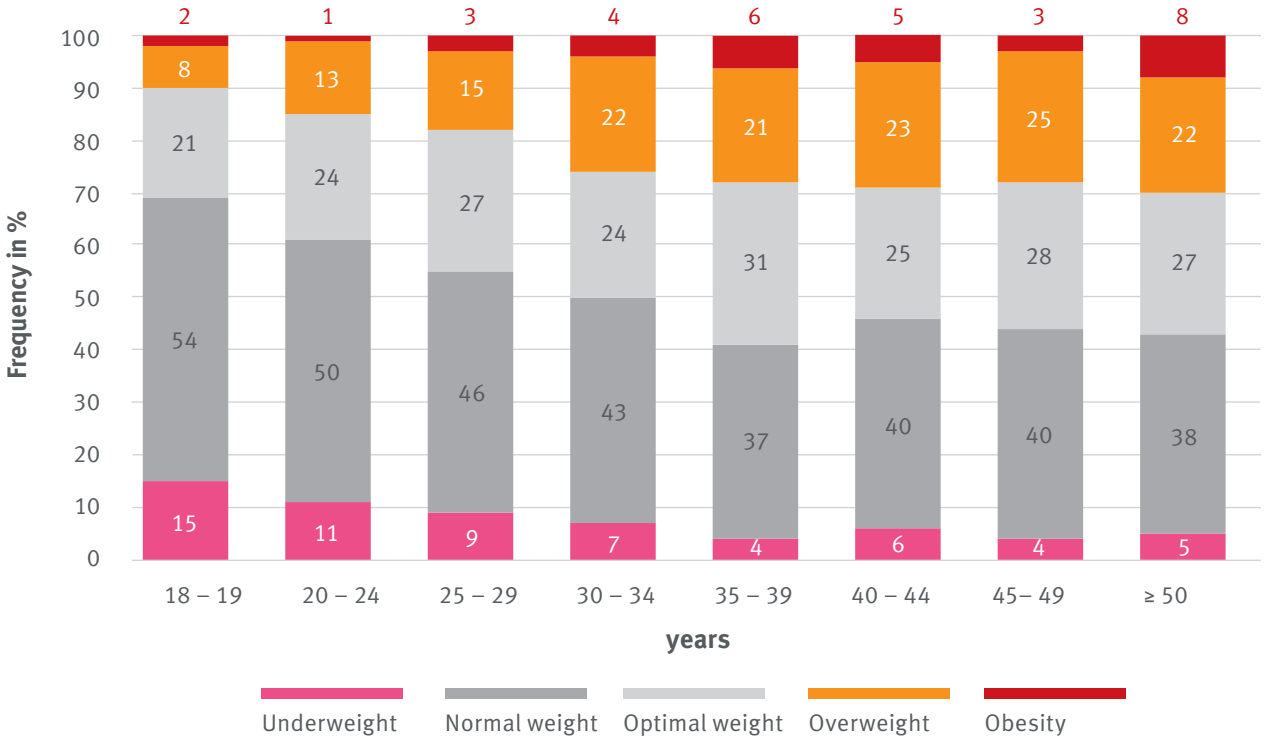

Figure 11: Weight categories adults aged 18 and over 2022,
Underweight: BMI ‹ 18.5 kg/sqm; normal weight: BMI men 18.5 – 22.9 kg/sqm; BMI women 18.5 – 21.9 kg/sqm;
Optimal weight: BMI men 23.0 – 24.9 kg/qm; BMI women 22.0 – 24.9 kg/qm; overweight: BMI 25.0 – 29.9 kg/qm; obese: BMI ≥ 30 kg/qm

	Male	Female	Total
Underweight	6.6	9.5	8.0
Normal weight	44.1	43.9	44.0
Optimal weight	23.1	29.4	26.0
Overweight	21.9	13.7	18.1
Obesity	4.3	3.5	3.9

Table 14: Weight categories of adults aged 18 and over (frequencies in %) 2022,
Underweight: BMI ‹ 18.5 kg/sqm; normal weight: BMI men 18.5 – 22.9 kg/sqm; BMI women 18.5 – 21.9 kg/sqm;
Optimal weight: BMI men 23.0 – 24.9 kg/qm; BMI women 22.0 – 24.9 kg/qm; overweight: BMI 25.0 – 29.9 kg/qm; obese: BMI ≥ 30 kg/qm

05

Nutritional status

5c. Development of nutritional status 2000 – 2022 Children and adolescents under 18 years of age

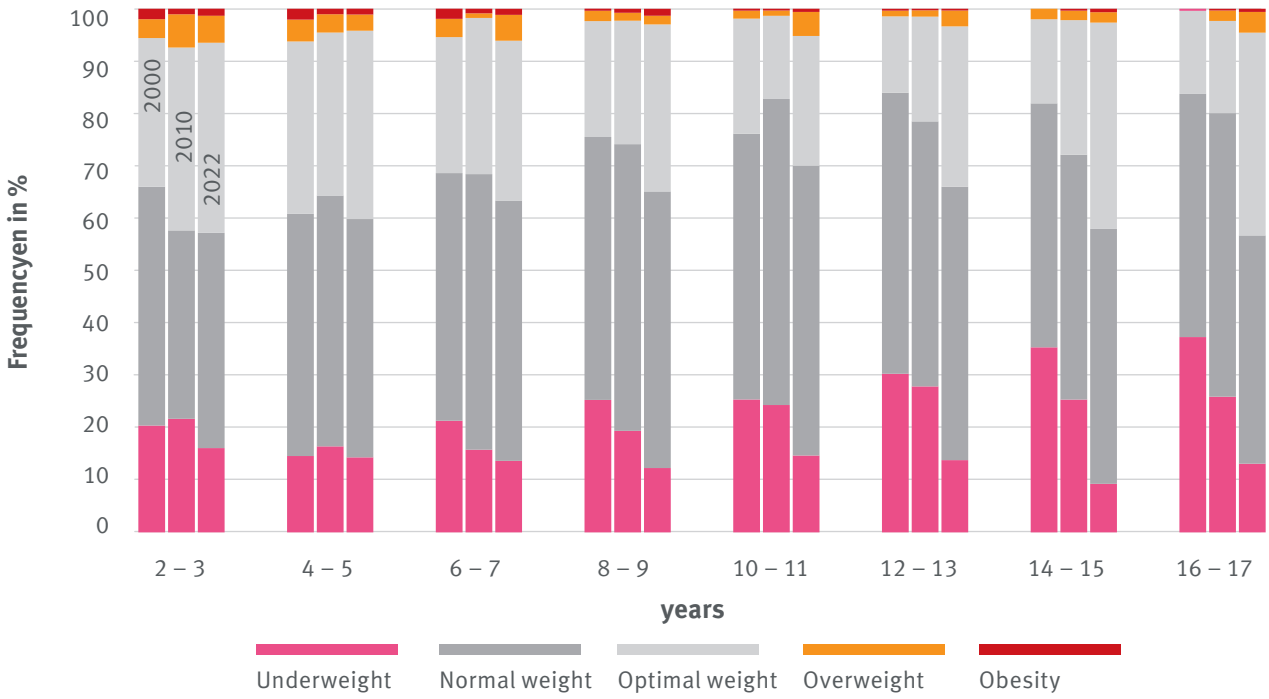

Figure 12: Development of the weight categories of children and adolescents up to 17 years 2000 – 2022, underweight: BMI percentiles < 10; Normal weight: BMI percentiles 10 – 49; optimal weight: BMI percentiles 50 – 89; overweight: BMI percentiles 90 – 96, obese: BMI percentiles ≥ 97

Weight categories	Reporting year	Age in years							
		2 – 3	4 – 5	6 – 7	8 – 9	10 – 11	12 – 13	14 – 15	16 – 17
Underweight	2000	20.4	14.6	21.3	25.3	25.4	30.3	35.3	37.3
	2010	21.7	16.4	15.8	19.1	24.3	27.9	25.4	25.9
	2022	16.1	14.3	13.7	12.3	14.7	13.8	9.3	13.1
Normal weight	2000	45.6	46.3	47.3	50.2	50.8	53.7	46.6	46.4
	2010	36.0	47.9	52.6	55.0	58.5	50.6	46.7	54.2
	2022	41.1	45.5	49.7	52.8	55.4	52.2	48.7	43.6
Optimal weight	2000	28.4	32.9	26.0	22.1	22.0	14.6	16.1	15.9
	2010	34.9	31.1	29.8	23.7	15.9	20.0	25.7	17.6
	2022	36.3	36.0	30.5	31.9	24.8	30.6	39.4	38.7
Overweight	2000	3.6	4.2	3.5	2.0	1.5	1.1	2.0	0.4
	2010	6.4	3.5	0.9	1.5	1.0	1.2	1.8	2.0
	2022	5.1	3.1	4.9	1.7	4.6	3.0	2.0	4.0
Obesity	2000	2.0	2.1	1.9	0.4	0.4	0.4	0.0	0.0
	2010	1.1	1.1	0.9	0.8	0.3	0.3	0.4	0.3
	2022	1.4	1.1	1.2	1.3	0.6	0.3	0.7	0.6

Table 15: Development of the weight categories of children and adolescents up to the age of 17 (frequencies in %) 2000 – 2022, Underweight: BMI percentiles < 10; normal weight: BMI percentiles 10 – 49; Optimal weight: BMI percentiles 50 – 89; Overweight: BMI percentiles 90 – 96, obesity: BMI percentiles ≥ 97

Nutritional status

5c.i Median BMI percentiles by birth cohort

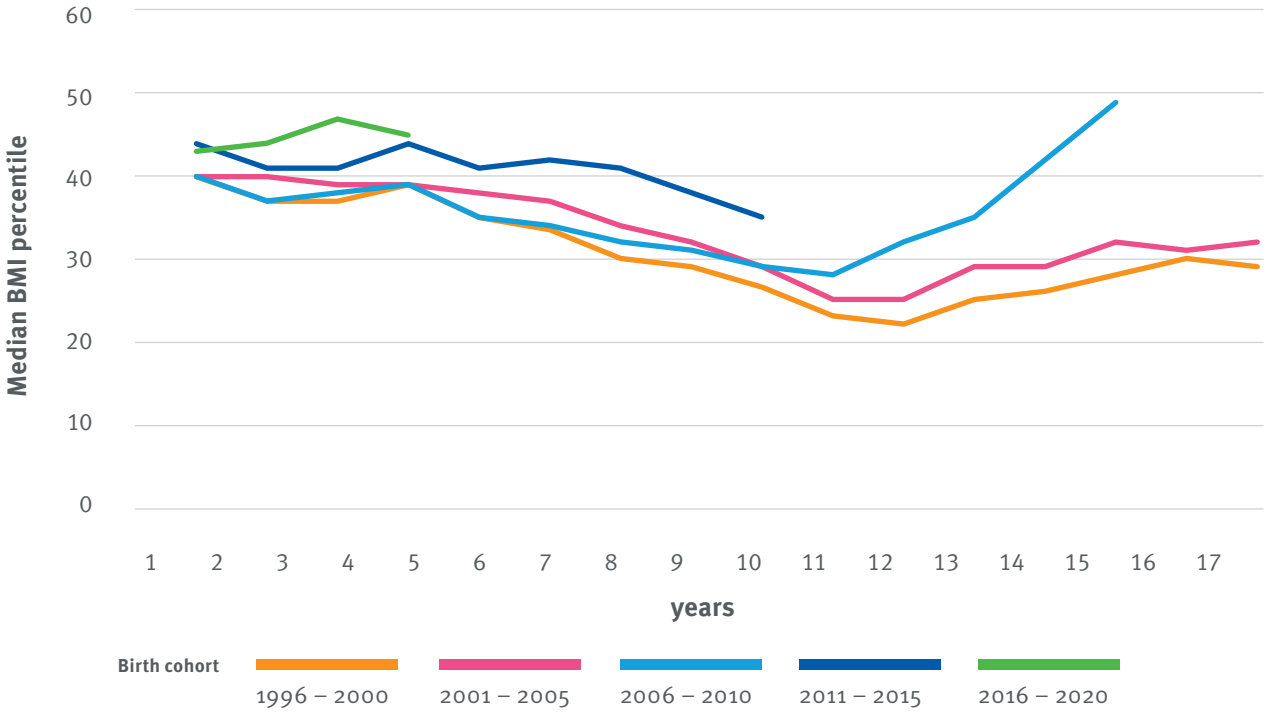

Figure 13: Development of median BMI percentiles of children and adolescents under 18 years of age by birth cohort 1996 – 2020 for the data from 1996 – 2022. Until 2014, the BMI close to the date of birth was recorded, from 2014 onwards the BMI at the time of recording the best lung functionof the calendar year.

Birth Cohorat	Age in years															
	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17
1996 – 2000	40	37	37	39	35	34	30	29	27	23	22	25	26	28	30	29
2001 – 2005	40	40	39	39	38	37	34	32	30	25	25	29	29	32	31	32
2006 – 2010	40	37	38	39	35	34	32	31	29	28	32	35	42	49	–	–
2011 – 2015	44	41	41	44	41	42	41	38	35	–	–	–	–	–	–	–
2016 – 2020	43	44	47	45	–	–	–	–	–	–	–	–	–	–	–	–

Table 16: Development of median BMI percentiles of children and adolescents under 18 years of age by birth cohort 1996 – 2020 for the years 1996 – 2022. Until 2014, the FEV1% close to the date of birth was recorded, from 2014 the best FEV1% of the calendar year.

05

Nutritional status

5d. Development of nutritional status 2000 – 2022 Adults aged 18 and over

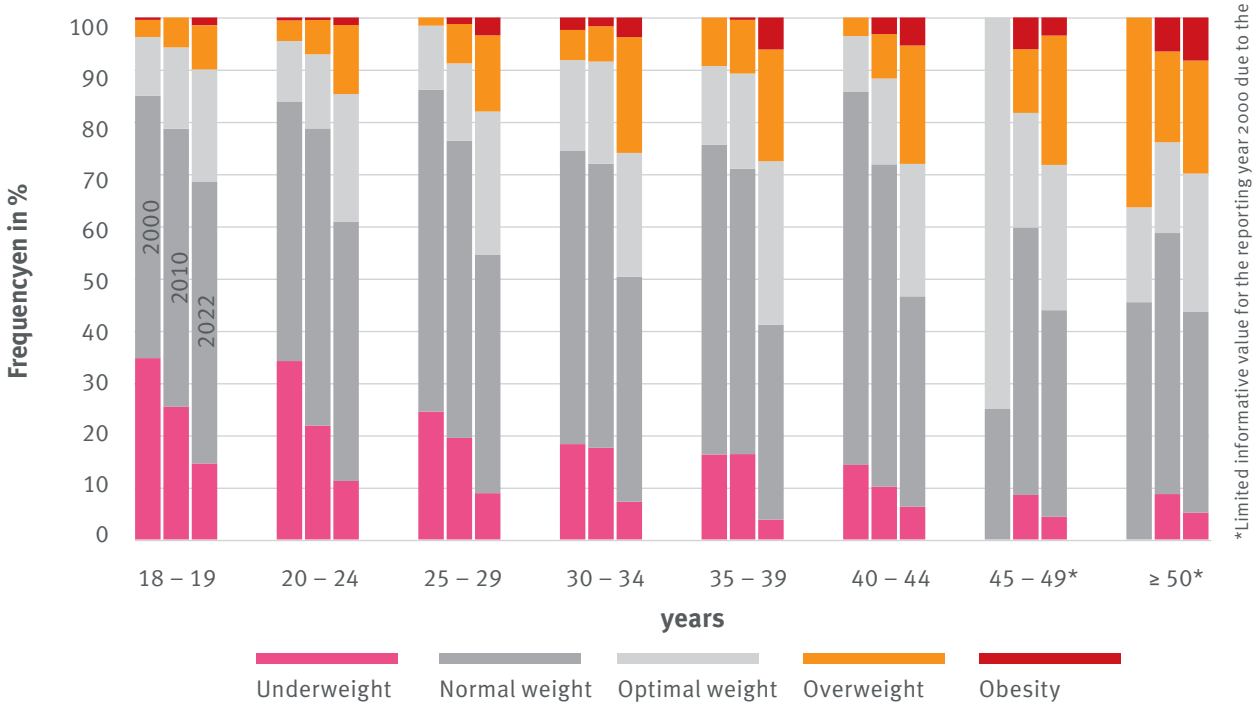

Figure 14: Development of weight categories of adults aged 18 and over 2000 – 2022,
Underweight: BMI < 18.5 kg/sqm; normal weight: BMI men 18.5 – 22.9 kg/sqm; BMI women 18.5 – 21.9 kg/sqm;
Optimal weight: BMI men 23.0 – 24.9 kg/qm, BMI women 22.0 – 24.9 kg/qm; overweight: BMI 25.0 – 29.9 kg/qm; obese: BMI ≥ 30 kg/qm

Weight categories	Reporting year	Age in years							
		18 – 19	20 – 24	25 – 29	30 – 34	35 – 39	40 – 44	45 – 49	≥ 50
Untergewicht	2000	34.7	34.2	24.5	18.3	16.3	14.3	0.0	0.0
	2010	25.4	21.8	19.5	17.6	16.4	10.1	8.5	8.7
	2022	14.6	11.2	8.9	7.2	3.8	6.3	4.4	5.1
Normalgewicht	2000	50.2	49.7	61.7	56.3	59.3	71.4	25.0	45.5
	2010	53.3	56.9	56.9	54.4	54.7	61.7	51.2	50.0
	2022	54.0	49.6	45.6	43.1	37.3	40.3	39.6	38.5
Optimalgewicht	2000	11.3	11.6	12.3	17.3	15.1	10.7	75.0	18.2
	2010	15.6	14.3	14.9	19.6	18.2	16.5	22.0	17.4
	2022	21.5	24.4	27.5	23.7	31.4	25.4	27.8	26.5
Overweight	2000	3.3	4.0	1.6	5.8	9.3	3.6	0.0	36.4
	2010	5.7	6.6	7.5	6.8	10.3	8.5	12.2	17.4
	2022	8.4	13.3	14.6	22.2	21.4	22.7	24.8	21.7
Obesity	2000	0.5	0.6	0.0	2.4	0.0	0.0	0.0	0.0
	2010	0.0	0.5	1.3	1.7	0.5	3.2	6.1	6.5
	2022	1.5	1.5	3.4	3.8	6.1	5.4	3.5	8.3

Table 17: Development of the weight categories of adults aged 18 and over (frequencies in %) 2000 – 2022
Underweight: BMI < 18.5 kg/sqm; normal weight: BMI men 18. – 22.9 kg/sqm; BMI women 18.5 – 21.9 kg/sqm; optimal weight: BMI men 23.0 – 24.9 kg/sqm, BMI women 22.0 – 24.9 kg/qm; overweight: BMI 25.0 – 29.9 kg/qm; obese: BMI ≥ 30 kg/qm

Nutritional status

5d.i Median BMI by birth cohort

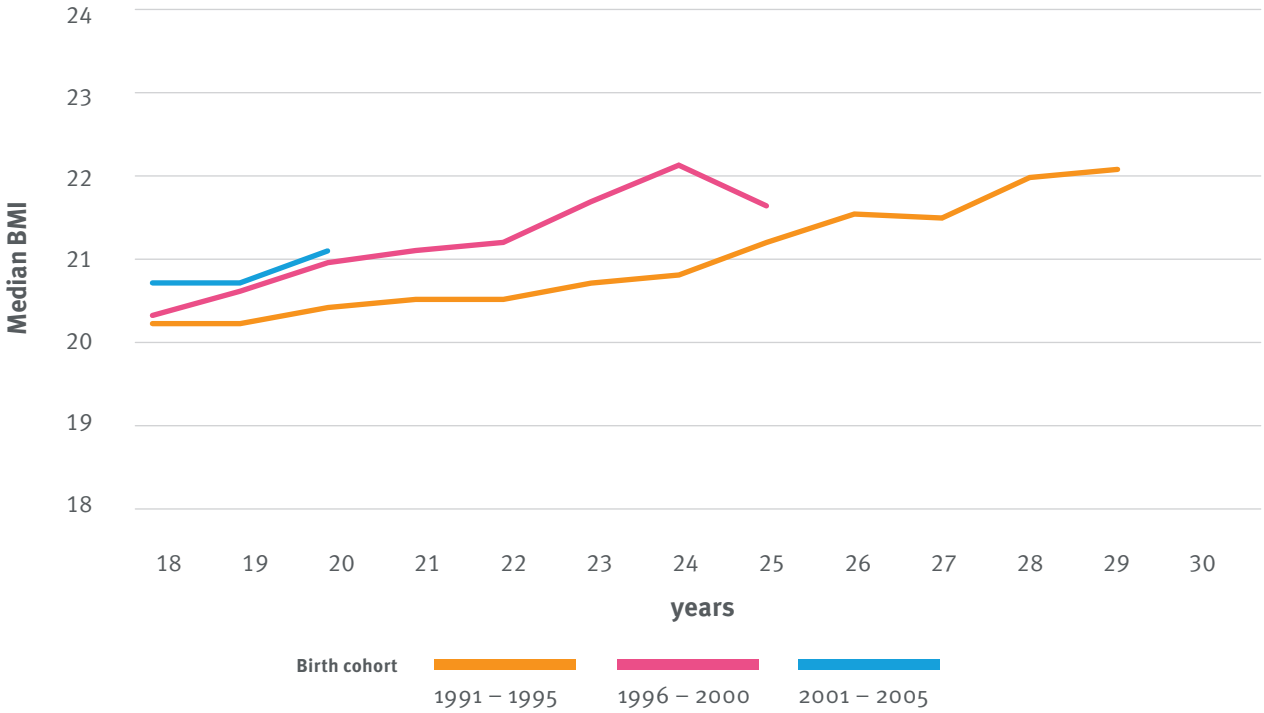

Figure 15: Development of median BMI of adults aged 18 and over by birth cohort 1991 – 2005 for the years 2009 – 2022.
Until 2014, the BMI close to the date of birth was recorded, from 2014 onwards the BMI at the time of recording the best FEV1% of the calendar year.

Birth Cohorat	Age in years											
	18	19	20	21	22	23	24	25	26	27	28	29
1991 – 1995	20.2	20.2	20.4	20.5	20.5	20.7	20.8	21.2	21.6	21.5	22.0	22.1
1996 – 2000	20.3	20.6	21.0	21.1	21.2	21.7	22.2	21.7	–	–	–	–
2001 – 2005	20.7	20.7	21.1*	–	–	–	–	–	–	–	–	–

Table 18: Development of median BMI of adults aged 18 and over by birth cohort 1991 – 2005 for the years 2009 – 2022.
Up to 2014, the FEV1% close to the date of birth was recorded; from 2014, the best FEV1% of the calendar year was recorded.

* Limited informative value due to the small cohort size

06

Lung function

6a. Overview of lung function

For the evaluations of lung function, all patients aged 6 years and older without transplantation, with lung function measurement 2022 were taken into account. A total of 5565 data sets were available.

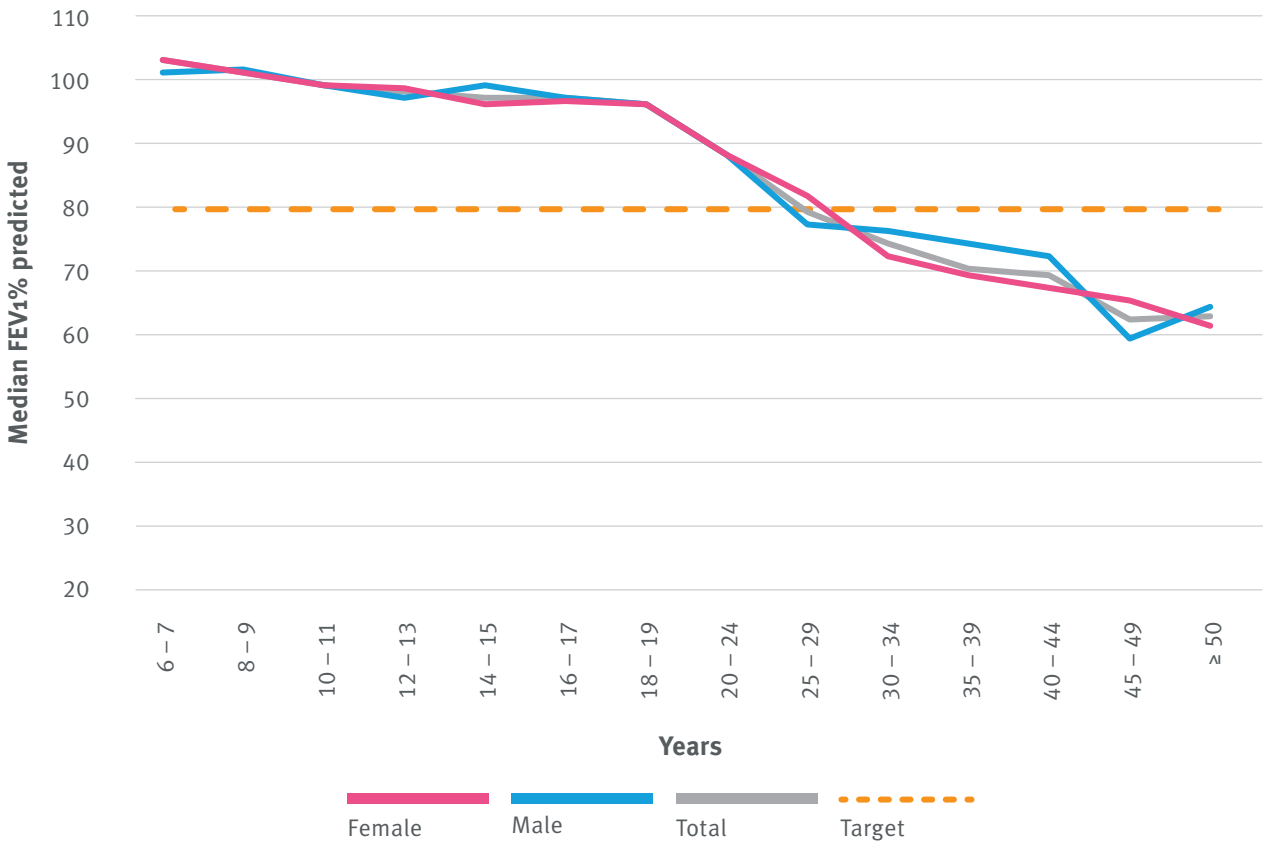

Figure 16: FEV1% value 2022 according to Global Lung Function Initiative (GLI)

Lung function

Age in years	Male			Female			Total		
	N	Median	25th - 75th pctl.	N	Median	25th - 75th pctl.	N	Median	25th - 75th pctl.
6 – 7	179	101.0	91 – 109	156	103.0	92 – 112	335	103.0	92 – 111
8 – 9	142	101.5	91 – 111	155	101.0	93 – 109	297	101.0	92 – 109
10 – 11	155	99.0	91 – 108	167	99.0	91 – 108	322	99.0	91 – 108
12 – 13	147	97.0	88 – 106	148	98.5	89 – 107	295	98.0	88 – 107
14 – 15	131	99.0	89 – 108	169	96.0	87 – 106	300	97.0	88 – 107
16 – 17	173	97.0	87 – 107	154	96.5	87 – 106	327	97.0	87 – 107
18 – 19	142	96.0	83 – 103	116	96.0	79 – 104	258	96.0	82 – 103
20 – 24	347	88.0	69 – 100	397	88.0	68 – 102	744	88.0	69 – 101
25 – 29	368	77.0	58 – 96	322	81.5	62 – 96	690	79.0	59 – 96
30 – 34	355	76.0	56 – 92	273	72.0	56 – 89	628	74.0	56 – 92
35 – 39	246	74.0	52 – 91	214	69.0	56 – 85	460	70.0	53 – 90
40 – 44	195	72.0	46 – 89	136	67.0	53 – 85	331	69.0	50 – 88
45 – 49	129	59.0	40 – 86	99	65.0	51 – 80	228	62.0	44 – 84
≥ 50	181	64.0	43 – 84	169	61.0	48 – 76	350	62.5	46 – 81

Table 19: FEV1% value 2022 according to Global Lung Function Initiative (GLI)

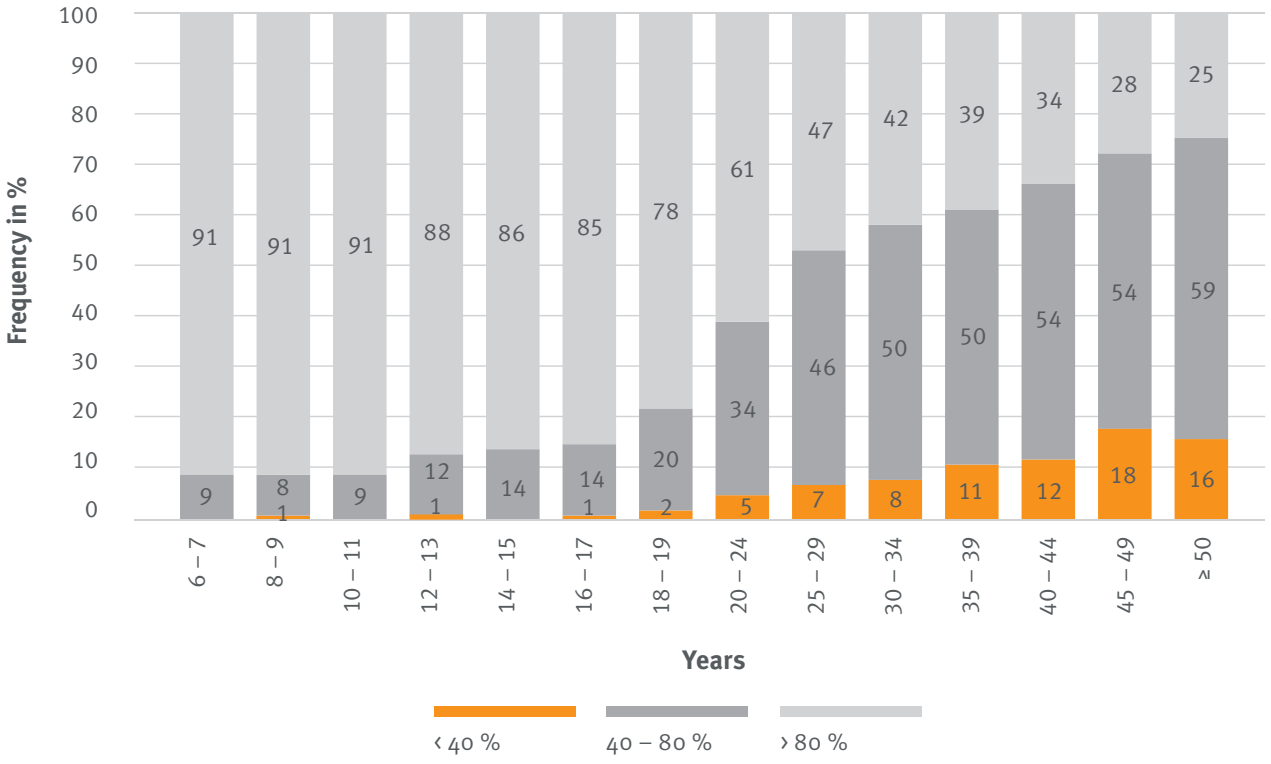

Figure 17: Severity of FEV1% (categories ‹ 40 %, 40 – 80 %, › 80 %) 2022 according to Global Lung Function Initiative (GLI)

06

Lung function

6b. Development of lung function 2000 – 2022

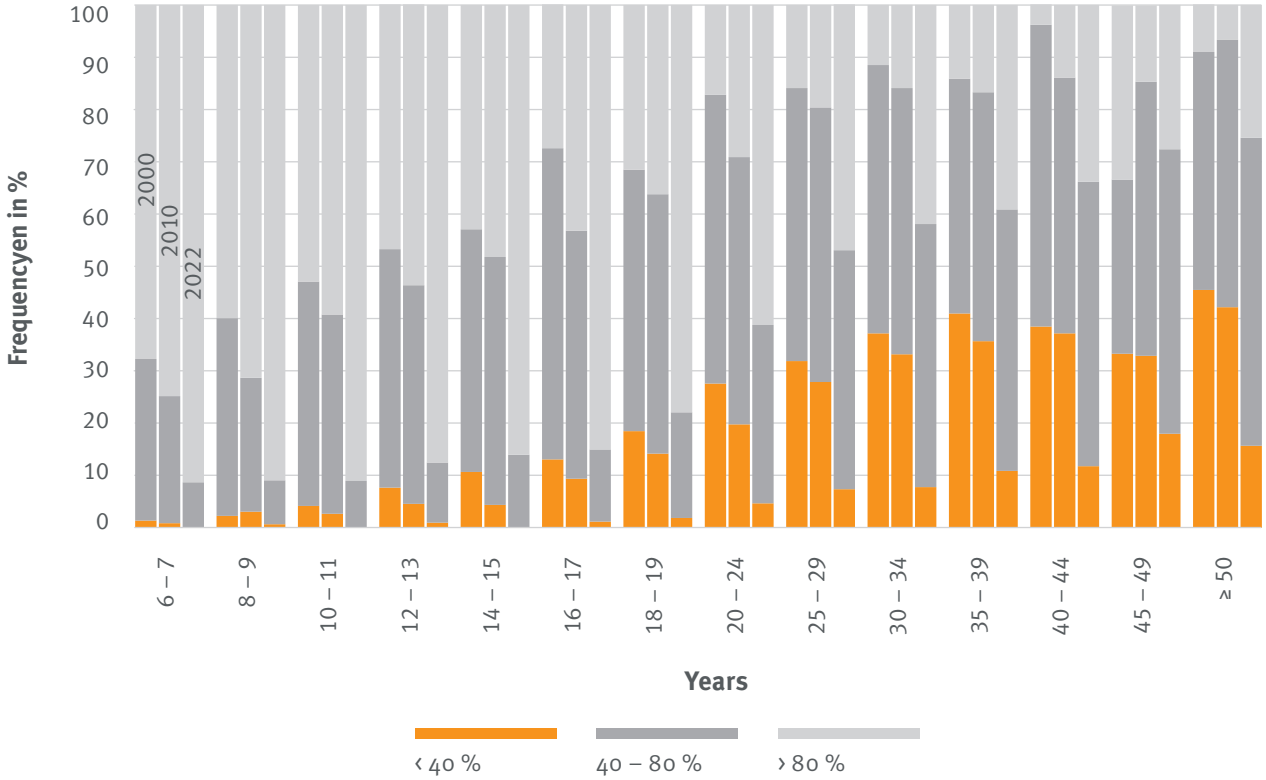

Figure 18: Development of age-related frequencies (in %) of the severity of FEV1% according to the Global Lung Initiative (GLI) 2000 – 2022

Severity levels of FEV1%	Reporting year	Age in years													
		6 – 7	8 – 9	10 – 11	12 – 13	14 – 15	16 – 17	18 – 19	20 – 24	25 – 29	30 – 34	35 – 39	40 – 44	45 – 49	≥ 50
‹ 40 %	2000	1.4	2.3	4.2	7.7	10.7	13.1	18.5	27.6	31.9	37.2	41.0	38.5	33.3	45.5
	2010	0.9	3.1	2.7	4.6	4.4	9.4	14.2	19.8	27.9	33.2	35.7	37.2	32.9	42.2
	2022	0.0	0.7	0.0	1.0	0.0	1.2	1.9	4.7	7.4	7.8	10.9	11.8	18.0	15.7
40 – 80 %	2000	30.9	37.8	42.9	45.6	46.4	59.5	50.0	55.2	52.2	51.3	44.9	57.7	33.3	45.5
	2010	24.3	25.3	38.1	41.8	47.4	47.4	49.6	51.1	52.5	50.9	47.6	48.9	52.4	51.1
	2022	8.7	8.4	9.0	11.5	14.0	13.8	20.2	34.1	45.7	50.3	50.0	54.4	54.4	58.9
› 80 %	2000	67.7	59.9	52.9	46.8	42.9	27.5	31.5	17.2	15.9	11.5	14.1	3.9	33.3	9.1
	2010	74.8	71.6	59.2	53.7	48.2	43.2	36.2	29.1	19.6	15.9	16.7	13.9	14.6	6.7
	2022	91.3	90.9	91.0	87.5	86.0	85.0	77.9	61.2	47.0	41.9	39.1	33.8	27.6	25.4

Table 20: Development of age-related frequencies (in %) of the severity of FEV1% according to the Global Lung Initiative (GLI) 2000 – 2022

Lung function

6c. Median FEV1% by birth cohort

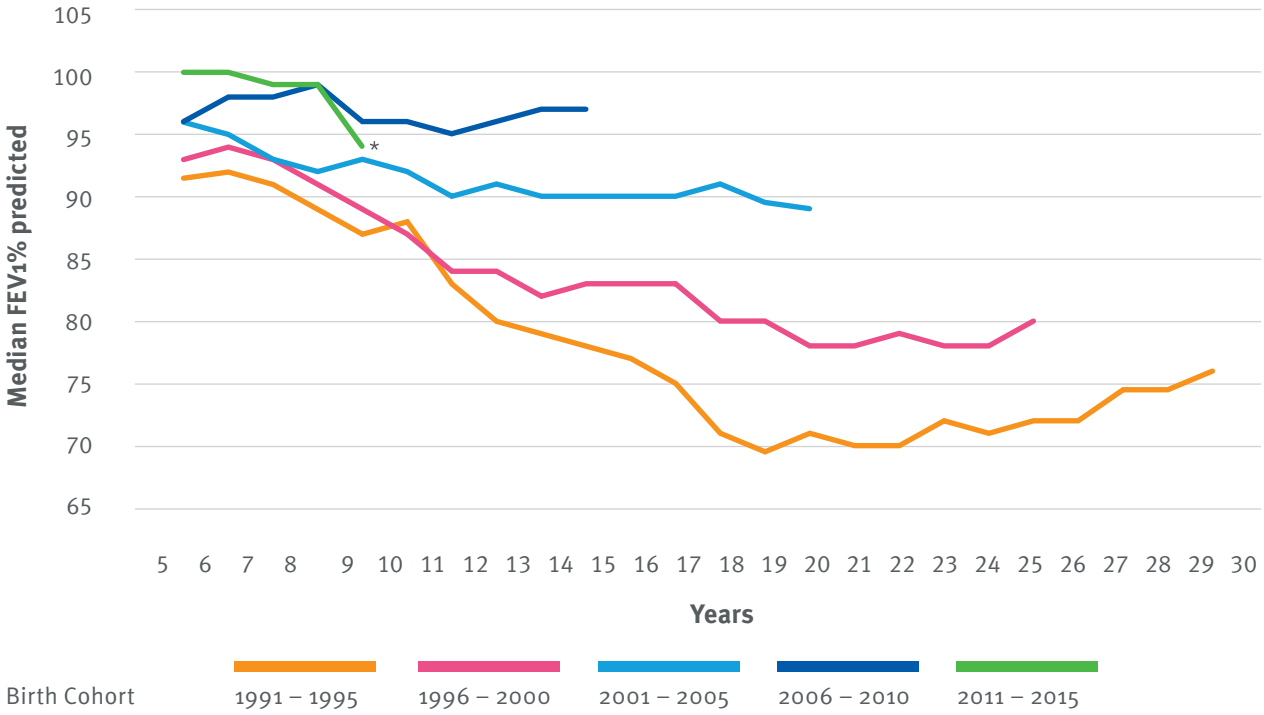

Figure 19: Development of median FEV1% of children and adults by birth cohort 1991 – 2015 for the years 1997 – 2022.
Until 2014, the FEV1% close to the date of birth was recorded; from 2014 onwards, the best FEV1% of the calendar year was recorded.

Birth Cohort	Age in years																							
	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20	21	22	23	24	25	26	27	28	29
1991 – 1995	92	92	91	89	87	88	83	80	79	78	77	75	71	70	71	70	70	72	71	72	72	75	75	76
1996 – 2000	93	94	93	91	89	87	84	84	82	83	83	83	80	80	78	78	79	78	78	80	–	–	–	–
2001 – 2005	96	95	93	92	93	92	90	91	90	90	91	90	91	90	89	–	–	–	–	–	–	–	–	–
2006 – 2010	96	98	98	99	96	96	95	96	97	97	–	–	–	–	–	–	–	–	–	–	–	–	–	–
2011 – 2015	100	100	99	99	94*	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	-	-

Table 21: Development of median FEV1% of children and adults by birth cohort 1991 – 2015 for the years 1997 – 2022.
Until 2014, the FEV1% close to the date of birth was recorded; from 2014 onwards, the best FEV1% of the calendar year was recorded.
* Limited informative value due to the small cohort size

07

Lung infection

7a. Annual verification at least once

All patients without transplantation who had at least one microbiological examination in the calendar year were included in the calendar year (n=6487). No information on the microbiological test in the calendar year was available for 122 patients (1.9 %).

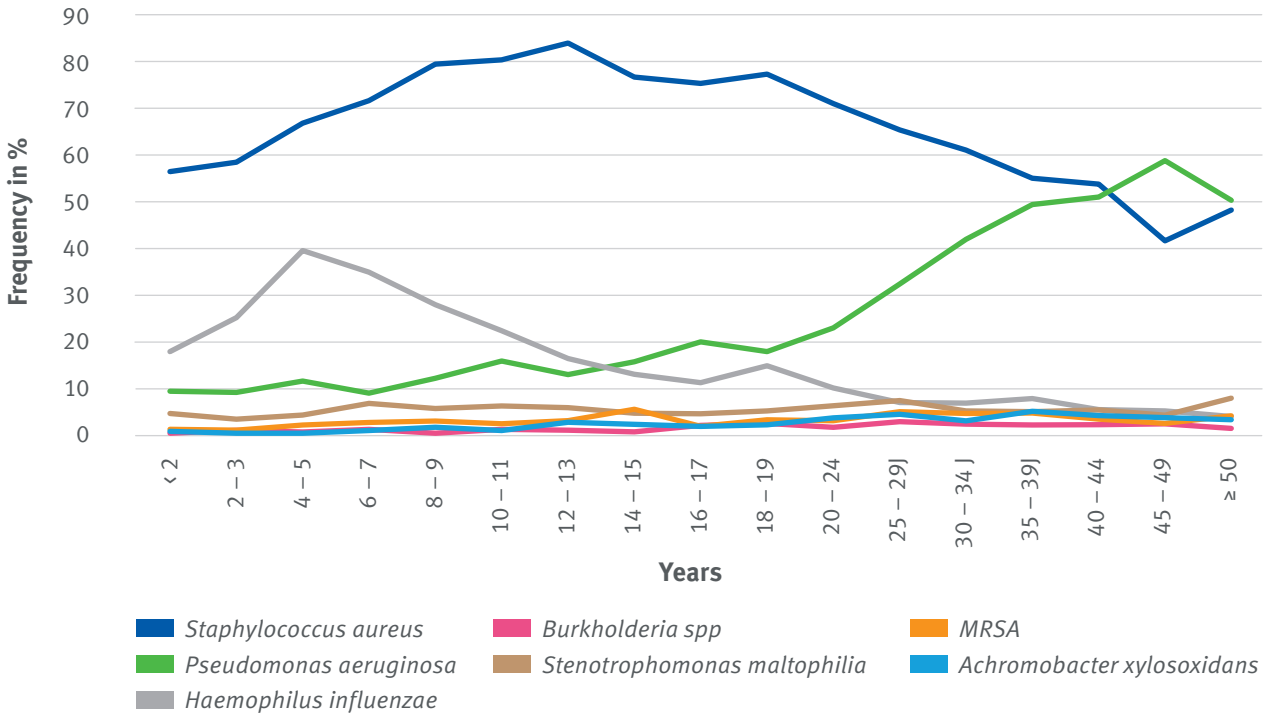

Figure 20: Bacterial detection in pwCF with microbiological testing 2022

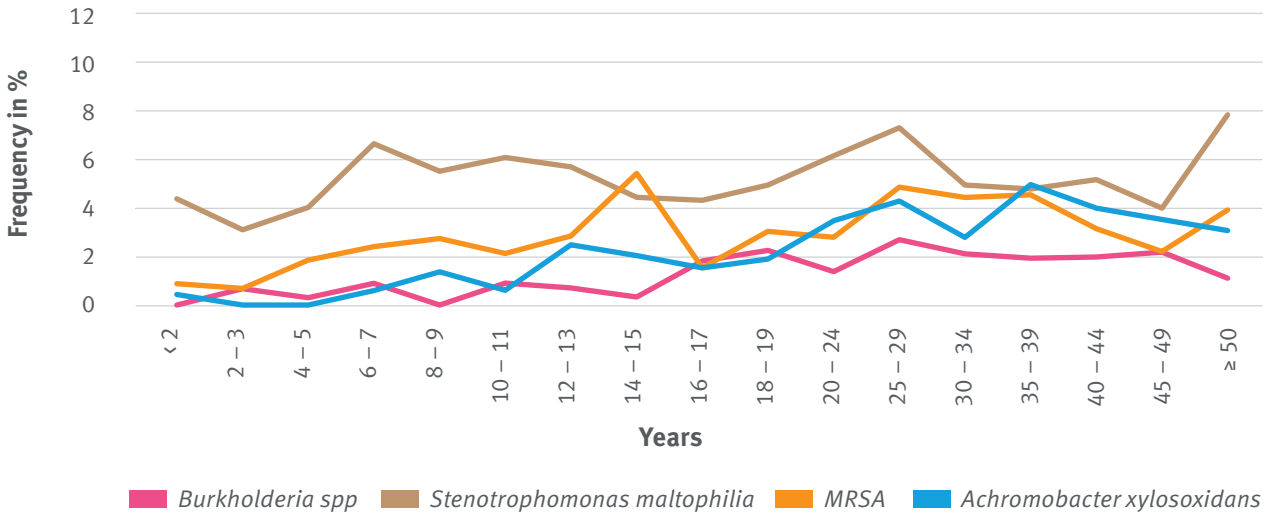

Figure 21: Bacterial detection in pwCF with microbiological examination (without the presentation of Pseudomonas aeruginosa, Staphylococcus aureus and Haemophilus influenzae) 2022

Lung infection

7a. Annual verification at least once

Age (years)	*Staphylococcus aureus inklusive MRSA*	*MRSA*	*Pseudomonas aeruginosa (PSA)*	*of these MRGN*	*Burkholderia spp*	*Stenotrophomonas maltophilia*	*Achromobacter xylosoxidans*	*Haemophilus influenzae*
< 2	56.2	0.9	9.0	5.0	0.0	4.3	0.4	17.6
2 – 3	58.3	0.7	8.8	11.5	0.7	3.0	0.0	24.9
4 – 5	66.7	1.8	11.2	2.7	0.3	3.9	0.0	39.4
6 – 7	71.5	2.4	8.6	6.9	0.9	6.5	0.6	34.7
8 – 9	79.4	2.7	11.8	5.7	0.0	5.4	1.4	27.7
10 – 11	80.3	2.1	15.5	5.8	0.9	6.0	0.6	22.1
12 – 13	83.9	2.8	12.6	11.4	0.7	5.6	2.5	16.1
14 – 15	76.6	5.4	15.4	15.2	0.3	4.4	2.0	12.7
16 – 17	75.2	1.5	19.6	13.9	1.8	4.2	1.5	10.9
18 – 19	77.2	3.0	17.5	23.4	2.2	4.9	1.9	14.6
20 – 24	70.9	2.8	22.7	25.5	1.4	6.0	3.4	9.8
25 – 29	65.2	4.8	32.1	24.3	2.7	7.2	4.2	6.6
30 – 34	60.8	4.4	41.8	35.4	2.1	4.9	2.8	6.5
35 – 39	54.8	4.5	49.3	42.2	1.9	4.7	4.9	7.5
40 – 44	53.5	3.1	50.9	38.6	2.0	5.1	3.9	5.1
45 – 49	41.3	2.2	58.7	37.6	2.2	3.9	3.5	4.8
≥ 50	47.9	3.9	50.1	41.8	1.1	7.7	3.0	3.6
Total	65.8	3.1	27.3	29.7	1.4	5.4	2.5	14.1
< 18	72.4	2.3	12.7	9.3	0.7	4.9	1.0	23.3
≥ 18	60.9	3.7	38.1	34.6	2.0	5.8	3.6	7.3

Table 22: Detection of bacteria in pwCF with microbiological examination (frequencies in %) 2022

07

Lung infection

7a. Annual verification at least once

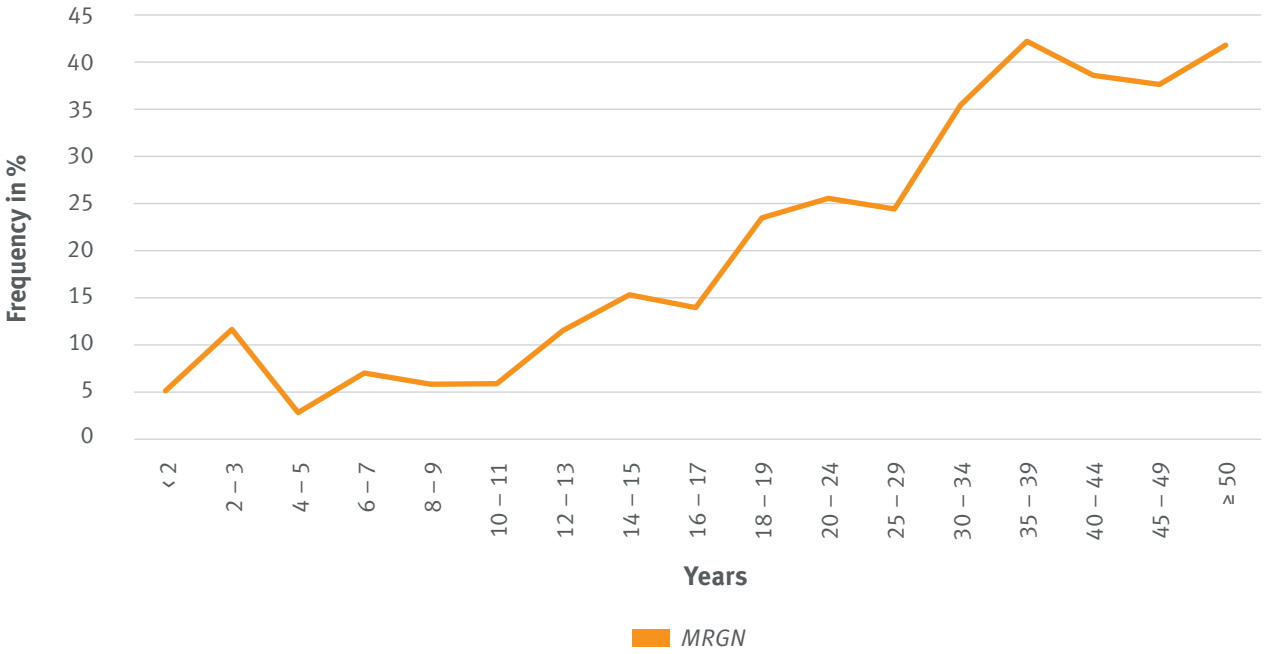

Figure 22: Bacterial detection for PSA multidrug-resistant (MRGN) in pwCF with PSA infection 2022

7b. Development of infections with Pseudomonas aeruginosa 2000 – 2022

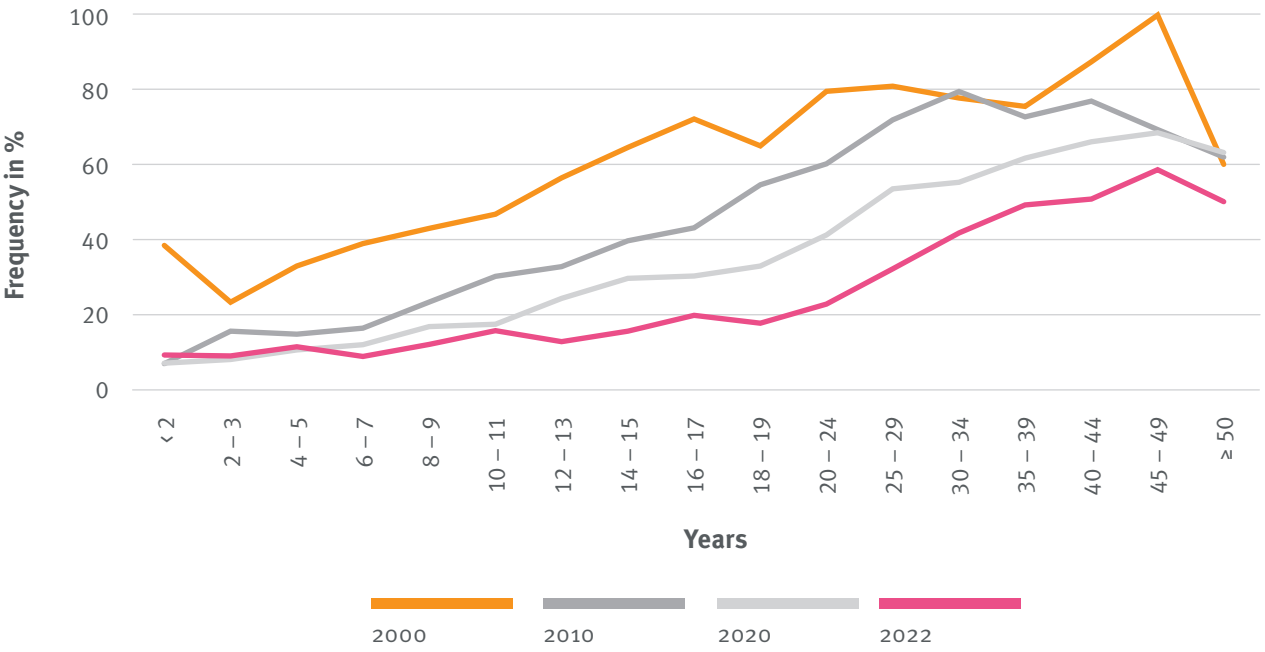

Figure 23: Development of Pseudomonas aeruginosa detection in pwCF with microbiological testing 2000 – 2022

Lung infection

7b. Development of infections with Pseudomonas aeruginosa 2000 – 2022

Age (years)	2000	2010	2020	2022
< 2	38.3	6.7	6.8	9.0
2 – 3	23.1	15.4	7.8	8.8
4 – 5	32.8	14.6	10.4	11.2
6 – 7	38.8	16.2	11.8	8.6
8 – 9	42.9	23.3	16.6	11.8
10 – 11	46.7	30.1	17.3	15.5
12 – 13	56.4	32.7	24.2	12.6
14 – 15	64.5	39.6	29.6	15.4
16 – 17	72.2	43.1	30.2	19.6
18 – 19	64.9	54.6	32.9	17.5
20 – 24	79.6	60.2	41.2	22.7
25 – 29	81.0	72.0	53.5	32.1
30 – 34	77.8	79.6	55.3	41.7
35 – 39	75.6	72.8	61.8	49.3
40 – 44	87.5	77.0	66.2	50.8
45 – 49	100.0	69.4	68.6	58.7
≥ 50	60.0	62.0	63.3	50.1

Table 23: Development of Pseudomonas aeruginosa detection in pwCF with microbiological testing (frequencies in %) 2000 – 2022

07

Lung infection

7c. Chronic lung infections

All patients without transplantation were included in the analyses of chronic lung infections, who had at least one microbiological examination in the calendar year (n=6487). Of 122 patients (1.9 %) were no information on the microbiological examination in the calendar year was available.

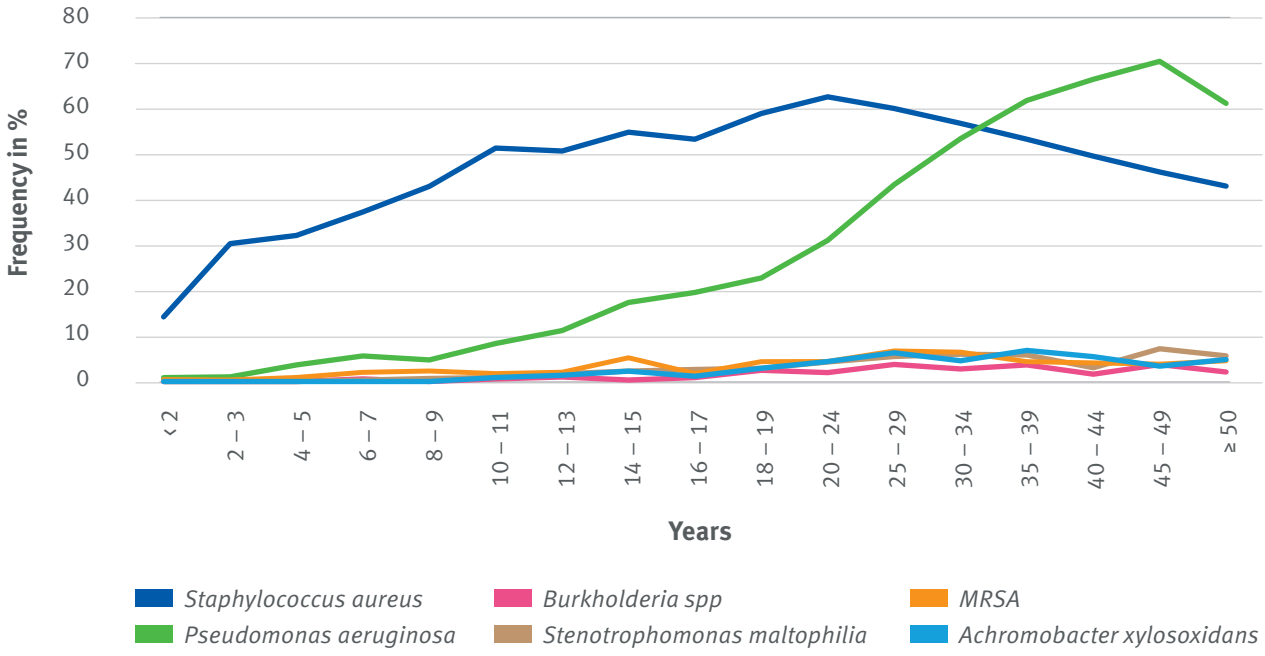

Figure 24: Chronic lung infections in pwCF with microbiological examination 2022

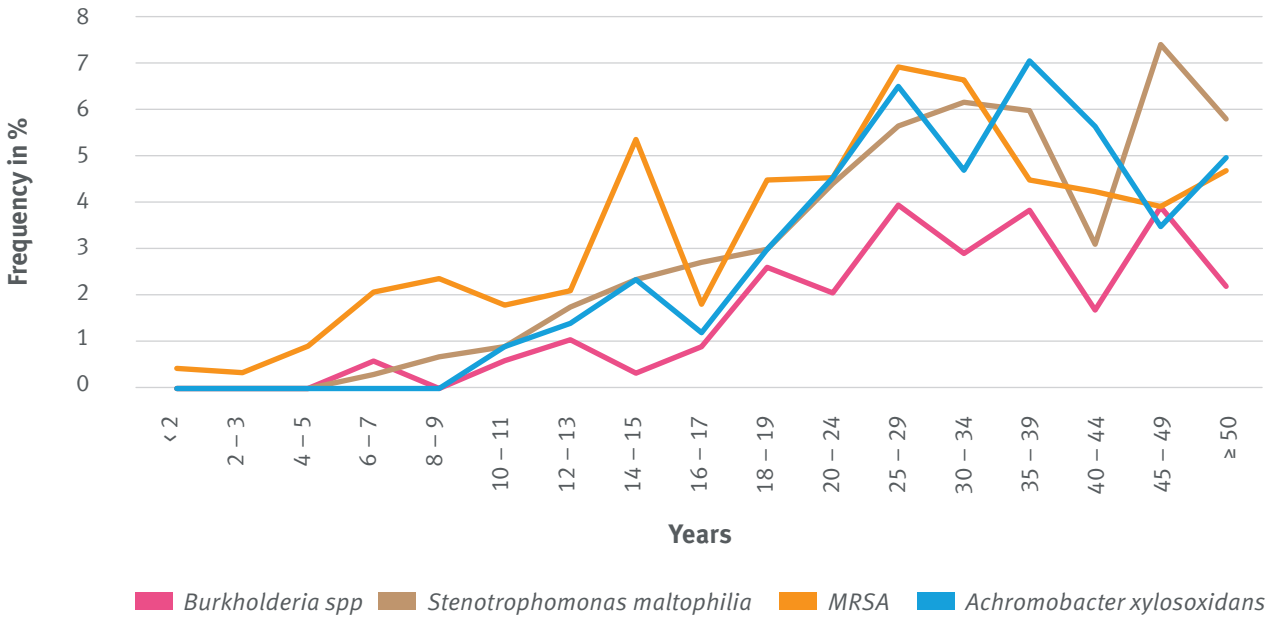

Figure 25: Chronic lung infections in pwCF with microbiological examination (without the presentation of Pseudomonas aeruginosa and Staphylococcus aureus) 2022

Lung infection

7c. Chronic lung infections

Alter (Jahre)	*Staphylococcus aureus inklusive MRSA chronic*	*MRSA chronic*	*Pseudomonas aeruginosa (PSA) chronisch*	*Burkholderia spp chronic*	*Stenotrophomonas maltophilia chronic*	*Achromobacter xylosoxidans chronic*
< 2	14.2	0.4	0.9	0.0	0.0	0.0
2 – 3	30.3	0.3	1.0	0.0	0.0	0.0
4 – 5	32.1	0.9	3.6	0.0	0.0	0.0
6 – 7	37.3	2.1	5.6	0.6	0.3	0.0
8 – 9	42.9	2.4	4.7	0.0	0.7	0.0
10 – 11	51.3	1.8	8.4	0.6	0.9	0.9
12 – 13	50.7	2.1	11.2	1.1	1.8	1.4
14 – 15	54.9	5.4	17.4	0.3	2.3	2.3
16 – 17	53.3	1.8	19.6	0.9	2.7	1.2
18 – 19	59.0	4.5	22.8	2.6	3.0	3.0
20 – 24	62.6	4.5	31.0	2.1	4.4	4.5
25 – 29	60.1	6.9	43.3	4.0	5.6	6.5
30 – 34	56.8	6.6	53.4	2.9	6.2	4.7
35 – 39	53.3	4.5	61.8	3.8	6.0	7.0
40 – 44	49.6	4.2	66.5	1.7	3.1	5.6
45 – 49	46.1	3.9	70.4	3.9	7.4	3.5
≥ 50	43.0	4.7	61.2	2.2	5.8	5.0
Total	49.6	3.9	31.8	1.9	3.4	3.3
< 18	41.5	1.9	8.3	0.4	1.0	0.7
≥ 18	55.6	5.3	49.0	2.9	5.2	5.2

Table 24: Chronic lung infections in pwCF with microbiological examination (frequencies in %) 2022

07

Lung infection

7d. Atypical mycobacteria

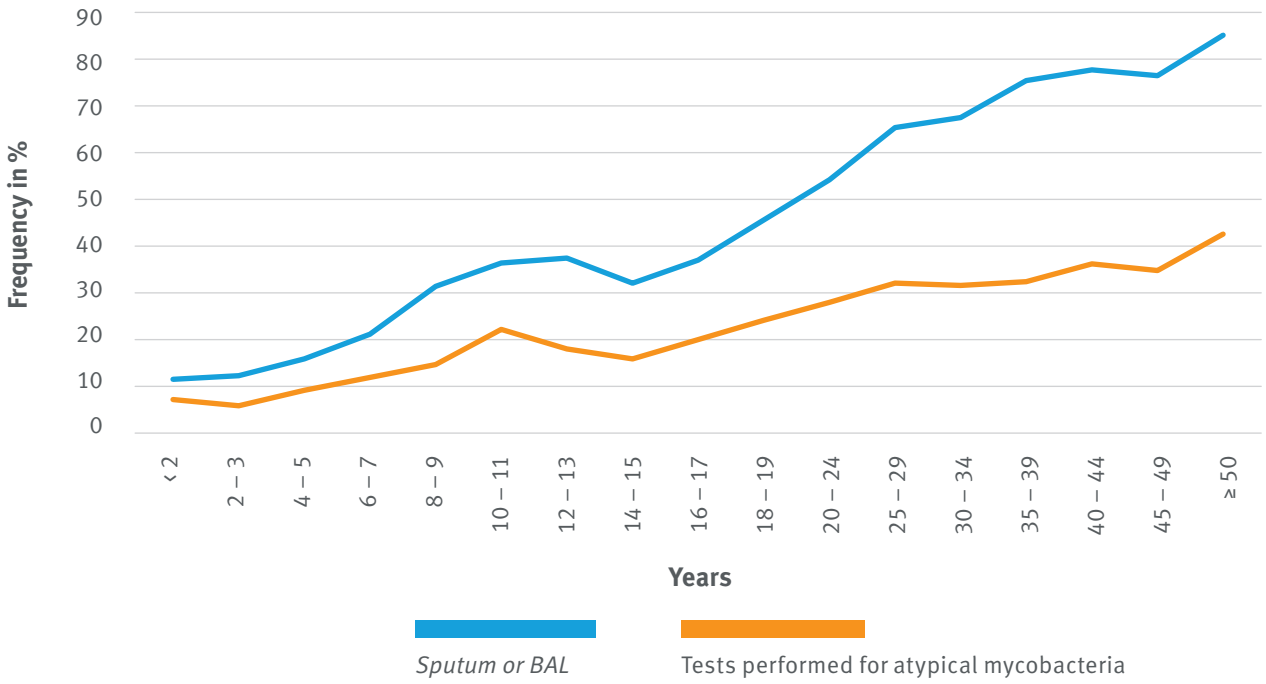

Figure 26: Frequencies of patients with sputum or BAL and tests performed for atypical mycobacteria 2022

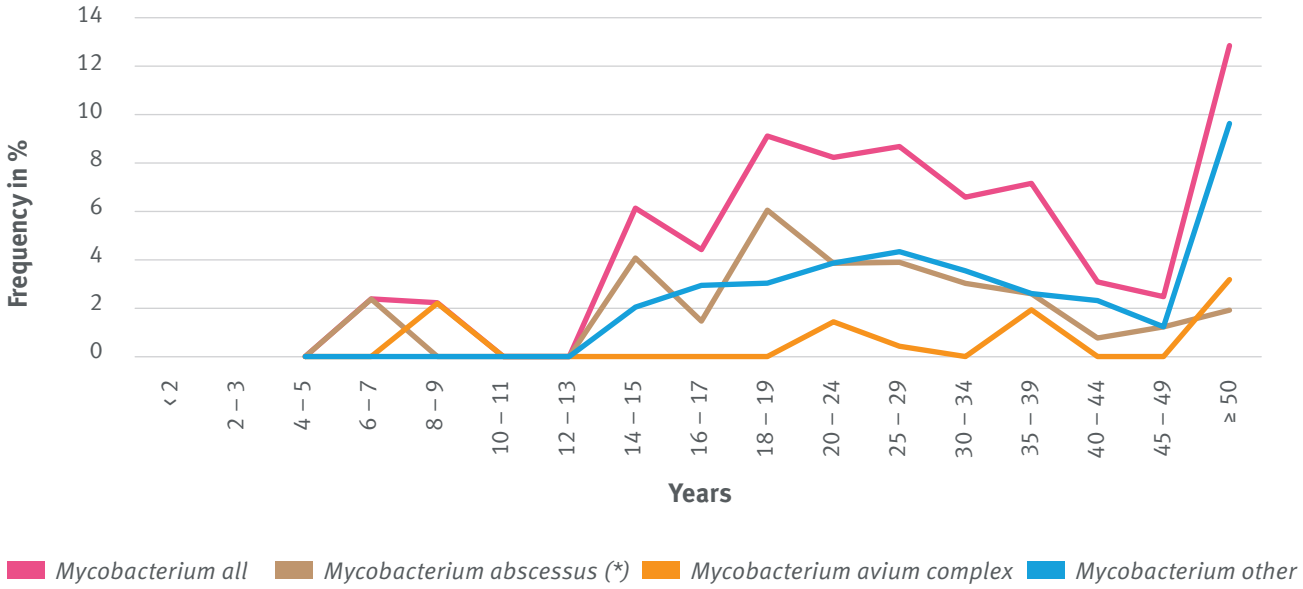

Figure 27: Age-dependent frequency of patients with tests for atypical mycobacteria 2022

(*) Mycobacterium abscessus includes Mycobacterium abscessus bolleti, Mycobacterium abscessus massiliense, Mycobacterium abscessus abscessus and Mycobacterium abscessus, multiple answers possible

Lung infection

7d. Atypical mycobacteria

Age (years)	Sputum or BAL performed	Sputum or BAL Frequency in %[1]	Examination for atypical mycobacteria performed Number of patients[2]	Untersuchung auf atypischeExamination for atypical mycobacteria performed Frequency in % [2]	Mycobacterium abscessus (*) Number of patients[3]	Mycobacterium abscessus (*) Frequencyen in %[3]	Mycobacterium avium complex Anzahl Patienten[3]	Mycobacterium avium complex Frequencyen in %[3]	Mycobacteriumothers Anzahl Patienten[3]	Mycobacteriumothers Frequencyen in %[3]	Mycobacterium all Anzahl Patienten[3]	Mycobacterium all Frequencyen in %[3]
< 2	28	12.0	18	64.3	0	0.0	0	0.0	0	0.0	0	0.0
2 – 3	38	12.8	19	50.0	0	0.0	0	0.0	1	5.3	1	5.3
4 – 5	54	16.4	32	59.3	0	0.0	0	0.0	0	0.0	0	0.0
6 – 7	73	21.6	42	57.5	1	2.4	0	0.0	0	0.0	1	2.4
8 – 9	94	31.8	45	47.9	0	0.0	1	2.2	0	0.0	1	2.2
10 – 11	123	36.7	76	61.8	0	0.0	0	0.0	0	0.0	0	0.0
12 – 13	108	37.8	53	49.1	0	0.0	0	0.0	0	0.0	0	0.0
14 – 15	97	32.4	49	50.5	2	4.1	0	0.0	1	2.0	3	6.1
16 – 17	124	37.4	68	54.8	1	1.5	0	0.0	2	2.9	3	4.4
18 – 19	123	45.9	66	53.7	4	6.1	0	0.0	2	3.0	6	9.1
20 – 24	396	54.4	207	52.3	8	3.9	3	1.5	8	3.9	17	8.2
25 – 29	465	65.5	231	49.7	9	3.9	1	0.4	10	4.3	20	8.7
30 – 34	418	67.6	198	47.4	6	3.0	0	0.0	7	3.5	13	6.6
35 – 39	354	75.5	154	43.5	4	2.6	3	2.0	4	2.6	11	7.1
40 – 44	276	77.8	130	47.1	1	0.8	0	0.0	3	2.3	4	3.1
45 – 49	176	76.5	81	46.0	1	1.2	0	0.0	1	1.2	2	2.5
≥ 50	309	85.1	156	50.5	3	1.9	5	3.2	15	9.6	20	12.8
Total	3256	50.2	1625	49.9	40	2.5	13	0.8	54	3.3	102	6.3
< 18	739	26.9	402	54.4	4	1.0	1	0.3	4	1.0	9	2.2
≥ 18	2517	67.3	1223	48.6	36	2.9	12	1.0	50	4.1	93	7.6

Table 25: pwCF with a test for atypical mycobacteria (frequency in %) 2022

[1] Sputum or BAL: Related to all patients with microbiological examination.
[2] Examination for atypical mycobacteria: Related to patients with sputum/BAL
[3] For the individual mycobacteria: In relation to patients with testing for atypical mycobacteria

(*) Mycobacterium abscessus includes Mycobacterium abscessus bolleti, Mycobacterium abscessus massiliense, Mycobacterium abscessus abscessus and Mycobacterium abscessus

08

Complications extended

All patients without a transplant who answered the question about complications were included in the analysis of complications. There were 6599 data sets available. For a total of 10 patients (0.2 %) the question about complications was not answered.

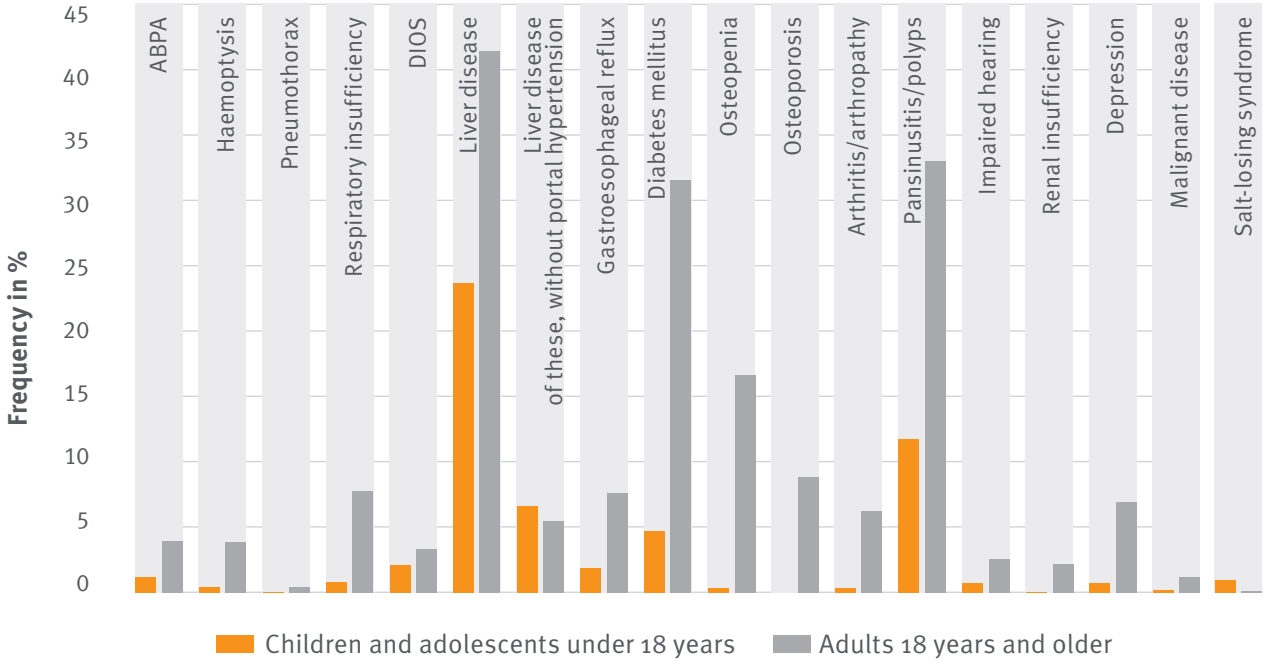

Figure 28: pwCF with complications (without the presentation of pancreatic insufficiency) 2022

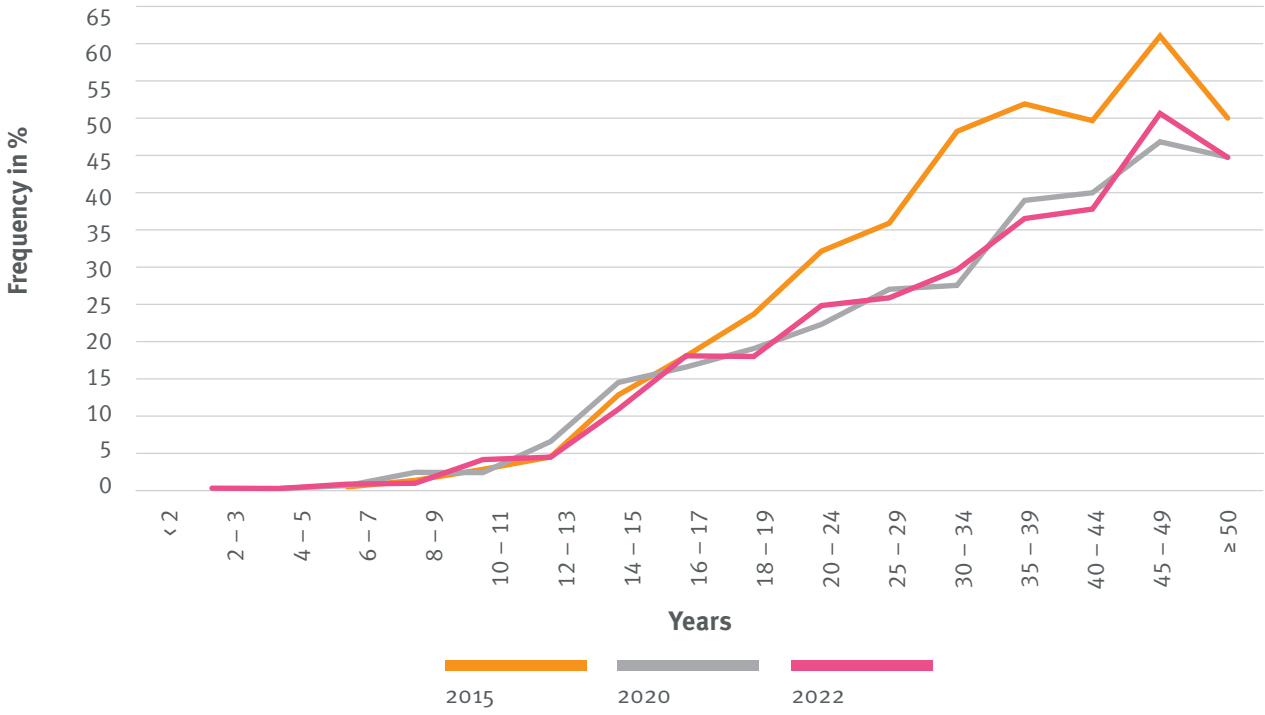

Figure 29: Development of diabetes detection in pwCF 2015 – 2022

Complications extended

Children and adolescents under the age of 18

Complication	0 – 5 years	6 – 11 years	12 – 17 years	Total
Pulmonary complications				
Allergic bronchopulmonary aspergillosis	0.1	0.8	2.6	1.2
Haemoptysis	0.1	0.3	0.8	0.4
of these, at least one serious episode (› 240 ml in 24h)	0.0	0.0	0.0	0.0
Pneumothorax	0.0	0.1	0.0	0.0
of these, requiring drainage	–	0.0	–	0.0
Respiratory insufficiency	0.9	0.7	0.8	0.8
of these, partial insufficiency	75.0	100.0	85.7	86.4
of these, global insufficiency	12.5	0.0	14.3	9.1
Gastrointestinal complications				
Exocrine pancreatic insufficiency	90.4	91.8	89.7	90.6
Distal intestinal obstruction syndrome (DIOS)	2.0	2.3	2.2	2.1
Liver disease	10.5	21.0	38.8	23.7
of these, liver cirrhosis	1.1	6.4	19.7	13.0
of these, with portal hypertension	0.0	3.4	6.7	4.7
of these, without portal hypertension	1.1	2.9	10.0	6.6
Gastroesophageal reflux	0.7	1.8	3.0	1.8
Other complications/comorbidities				
Diabetes mellitus	0.2	2.1	11.5	4.7
of these, Type 3	50.0	80.0	93.5	90.7
of these, not Type 3	50.0	20.0	6.5	9.3
Bone disease				
Osteopenia	0.1	0.2	0.7	0.3
Osteoporosis	0.0	0.0	0.0	0.0
Arthritis/Arthropathy	0.0	0.1	0.9	0.3
Pansinusitis/Polyps	2.8	13.5	18.3	11.8
Impaired hearing	0.5	0.6	1.1	0.7
Renal insufficiency	0.1	0.0	0.1	0.1
Depression	0.0	0.0	2.1	0.7
Malignant disease	0.4	0.0	0.2	0.2
Salt-losing syndrome	2.1	0.5	0.3	0.9

Table 26: pwCF under the age of 18 with complications (frequencies in %) 2022

08

Complications extended

8b. Adults 18 years and older

Complication	18 – 29 years	30 – 39 years	≥ 40 years	Total
Pulmonary complications				
Allergic bronchopulmonary aspergillosis	4.1	4.2	3.4	4.0
Haemoptysis	3.5	3.8	4.5	3.8
of these, at least one serious episode (› 240 ml in 24h)	7.8	2.6	0.0	3.9
Pneumothorax	0.5	0.4	0.5	0.4
of these, requiring drainage	62.5	75.0	80.0	70.6
Respiratory insufficiency	5.0	7.6	12.8	7.7
of these, partial insufficiency	65.5	71.8	73.4	70.6
of these, global insufficiency	24.1	17.7	12.1	17.2
Gastrointestinal complications				
Exocrine pancreatic insufficiency	92.8	90.8	85.7	90.4
Distal intestinal obstruction syndrome (DIOS)	3.6	3.6	2.6	3.3
Liver disease	43.4	41.4	37.8	41.4
of these, liver cirrhosis	20.5	14.2	14.2	17.2
of these, with portal hypertension	8.9	7.1	8.2	8.2
of these, without portal hypertension	7.6	3.5	3.6	5.4
Gastroesophageal reflux	6.4	6.3	11.4	7.6
Other complications/comorbidities				
Diabetes mellitus	24.2	32.7	43.6	31.6
of these, Type 3	96.2	95.1	96.9	96.1
of these, not Type 3	3.8	4.9	3.1	3.9
Bone disease				
Osteopenia	10.4	17.8	26.5	16.6
Osteoporosis	4.6	8.6	16.9	8.9
Arthritis/Arthropathy	3.8	7.5	9.3	6.3
Pansinusitis/Polyps	31.1	34.6	34.4	33.0
Impaired hearing	1.5	2.2	4.9	2.6
Renal insufficiency	1.1	2.1	4.2	2.2
Depression	7.6	6.7	5.9	6.9
Malignant disease	0.2	0.4	3.8	1.2
Salt-losing syndrome	0.1	0.1	0.2	0.1

Table 27: pwCF aged 18 and over with complications (frequencies in %) 2022

Complications extended

8c. Exacerbations treated with antibiotics

Number of antibiotic-treated Exacerbations per patient	Age (years)								
	0 – 5	6 – 11	12 – 17	18 – 29	30 – 39	≥ 40	Total	< 18	≥ 18
0	62.1	69.8	77.8	72.7	71.9	70.4	71.1	70.1	71.8
1	18.9	16.9	14.3	16.5	17.5	17.5	16.9	16.7	17.0
2	10.3	6.4	4.5	5.2	5.9	7.1	6.3	6.9	5.9
3	4.4	3.9	1.9	2.7	1.9	2.3	2.8	3.4	2.4
4	1.8	1.3	0.4	1.7	1.1	1.2	1.3	1.2	1.4
5+	2.1	1.3	1.0	0.9	0.7	0.4	1.0	1.4	0.7
unknown	0.5	0.3	0.1	0.4	1.0	1.1	0.6	0.3	0.7

Table 28: Number of exacerbations treated with antibiotics per cystic fibrosis patient (frequencies in %) 2022

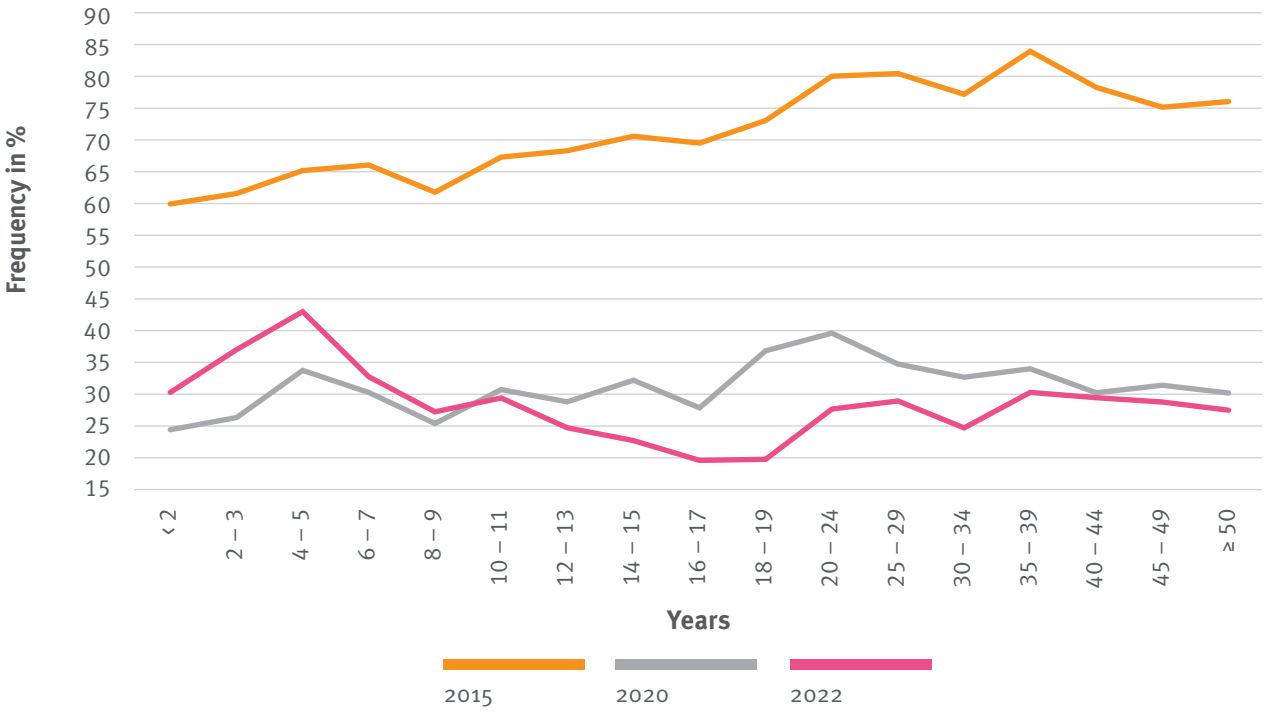

Figure 30: Development of age-related frequencies of cystic fibrosis patients with at least 1 antibiotic-treated exacerbation 2015 – 2022

Reporting year	Age (years)															
	< 2	2 – 3	4 – 5	6 – 7	8 – 9	10 – 11	12 – 13	14 – 15	16 – 17	18 – 19	20 – 24	25 – 29	30 – 34	35 – 39	40 – 44	45 – 49	≥ 50
2015	59.9	61.5	65.2	66.1	61.7	67.3	68.3	70.6	69.5	73.1	80.1	80.6	77.3	84.1	78.4	75.2	76.1
2020	24.3	26.2	33.8	30.2	25.3	30.7	28.8	32.2	27.8	36.9	39.7	34.8	32.7	34.0	30.2	31.4	30.1
2022	30.3	37.0	43.1	32.7	27.2	29.4	24.7	22.6	19.5	19.6	27.6	28.9	24.6	30.2	29.4	28.7	27.4

Table 29: Development of age-related frequencies (in %) of pwCF with at least 1 antibiotic-treated exacerbation 2015 –2022

09

Therapies

9a. Basic therapy

All patients without transplantation who answered the question about long-term gastrointestinal or pulmonary therapy were included. The data sets of 2767 patients under the age of 18 and 3839 patients aged 18 and over were included in the analyses. For 2 patients (0.03 %) the question about gastrointestinal or pulmonary long-term therapy was not answered.

9a.i Children and adolescents under 18 years

Basic therapy	0 – 5 years	6 – 11 years	12 – 17 years	Total
DNase	13.1	47.8	58.5	40.5
Mannitol	0.0	0.0	0.3	0.1
Hypotonic saline solution (≥3 %)	88.0	95.9	93.3	92.6
of these 3 – 5,7 %	27.1	24.0	22.5	24.4
of these ≥ 5,8 %	72.9	76.1	77.6	75.6
At least one mucolytic therapy (Mannitol, DNAse, Hypertonic saline solution ≥3 %)	88.4	97.0	95.1	93.7
ß2-sympathomimetics				
Short-acting (SABA)	60.7	71.4	70.7	67.8
Long-acting (LABA)	5.4	13.7	24.0	14.5
Anticholinergics	5.6	11.8	14.6	10.8
Antistaphylococcal therapy	7.9	7.9	8.6	8.1
Steroids				
Nasal	10.3	21.6	26.3	19.6
Inhalative	9.7	16.9	25.3	17.4
Orale	0.9	1.6	2.4	1.6
Vitamins				
Vitamin A	80.1	86.2	83.7	83.5
Vitamin D	96.0	96.9	96.4	96.5
Vitamin E	71.6	78.4	81.5	77.3
Vitamin K	73.8	79.7	76.3	76.7
Hormonal contraception for women[1]	–	–	9.3	24.0

Table 30: pwCF under the age of 18 with basic therapy (frequencies in %) 2022
[1] Survey from 14 years

Therapies

9a.ii Inhalation and combination therapies Children and adolescents under 18 years of age

The graph on inhalation and combination therapies takes into account all minors who answered the question who answered the question about long-term pulmonary therapy (2767 patients).Of these, 153 patients (5.5 %) did not receive any inhalation therapy.

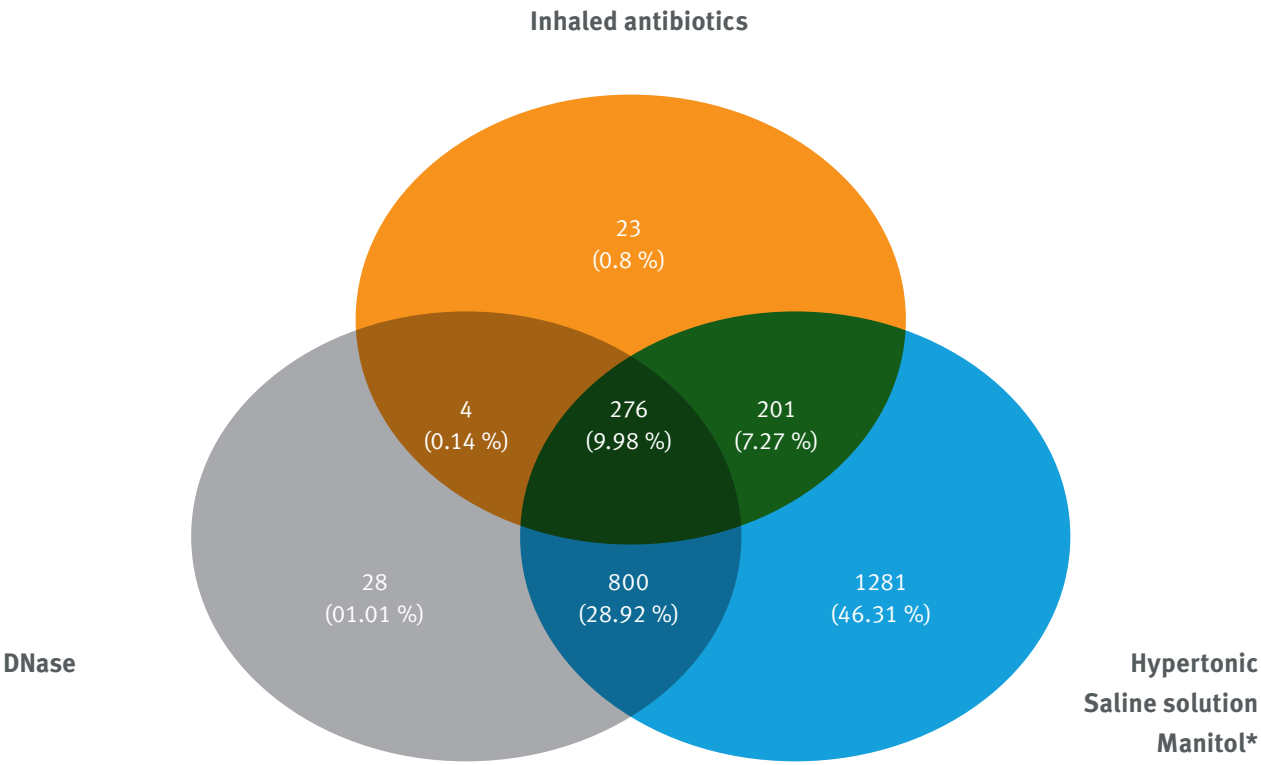

Figure 31: Inhalation and combination therapies in pwCF under 18 years of age 2022
*Hypertonic saline solution ≥ 3 %

09

Therapies

9a. Basic therapy

9a.iii Adults 18 years and older

Basic therapy	18 – 29 years	30 – 39 years	≥ 40 years	Total
DNase	53.9	47.0	41.1	48.6
Mannitol	2.7	6.1	4.6	4.2
Hypotonic saline solution (≥3 %)	80.2	71.5	66.1	74.1
of these 3 – 5,7 %	22.0	23.1	25.6	23.1
of these ≥ 5,8 %	78.0	76.9	74.4	76.9
At least one mucolytic therapy (Mannitol, DNAse, Hypertonic saline solution ≥3 %)	88.7	83.2	78.1	84.4
ß2-sympathomimetics				
Short-acting (SABA)	65.0	60.4	64.8	63.6
Long-acting (LABA)	43.0	60.6	72.7	55.8
Anticholinergics	34.2	50.0	63.9	46.5
Antistaphylococcal therapy	7.2	5.5	3.6	5.8
Steroids				
Nasal	24.3	23.9	19.2	22.9
Inhalative	35.1	50.0	57.7	45.3
Orale	5.0	6.6	9.2	6.6
Vitamins				
Vitamin A	79.5	70.3	62.5	72.5
Vitamin D	95.2	94.5	92.3	94.3
Vitamin E	79.3	68.3	62.2	71.8
Vitamin K	74.7	65.8	60.8	68.5
Hormonal contraception for women	32.7	20.2	11.6	21.7

Table 31: pwCF aged 18 and over with basic therapy (frequencies in %) 2022

Therapies

9a.iii Inhalation and combination therapies Adults aged 18 and over

The graph on inhalation and combination therapies includes all adults who answered the question about long-term pulmonary therapy (3839 patients). Of these, 354 patients (9.2 %) did not receive any inhalation therapy.

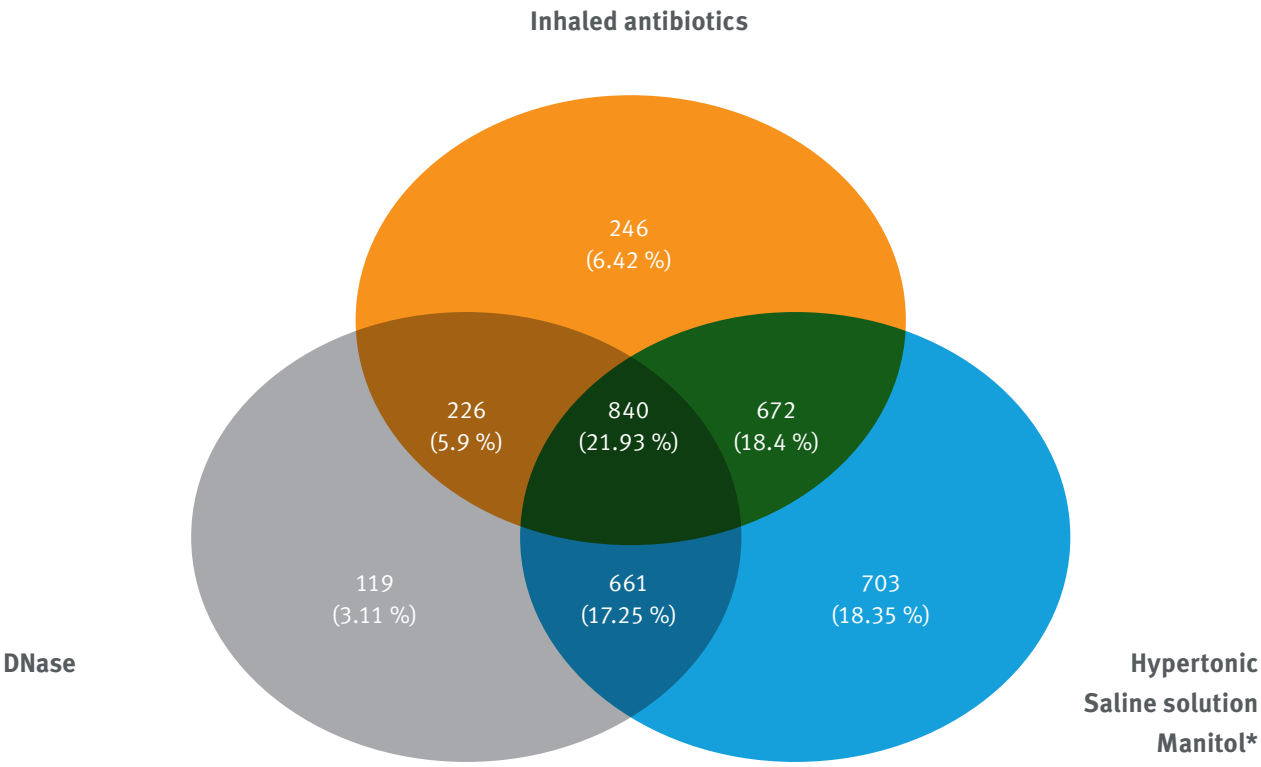

Figure 32: Inhalation and combination therapies in pwCF over 18 years of age 2022
*Hypertonic saline solution ≥ 3 %

09

Therapies

9b. Indication therapy

9b.i Children and adolescents under 18 years

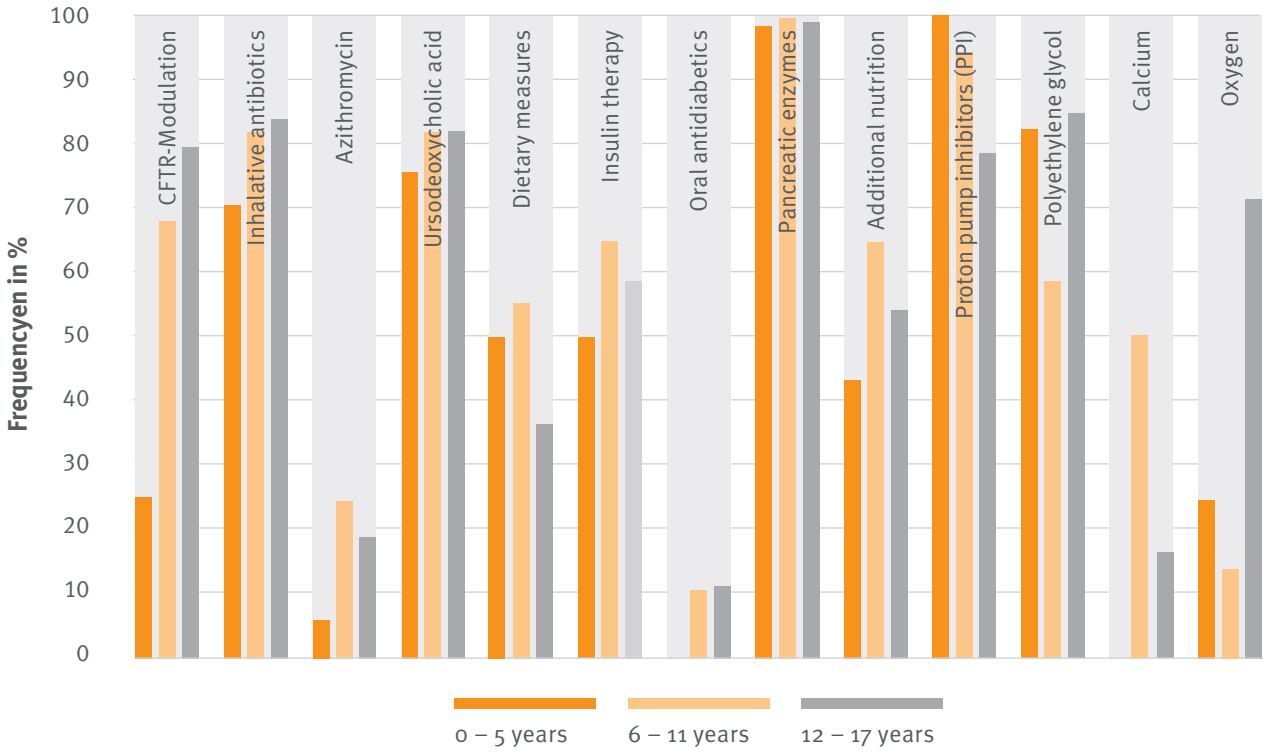

Figure 33: Status of approval of CFTR modulators

Status of the approval of CFTR modulators 31.12.2022
Only the last modulator used in the reporting year is shown here.

[1] Ivacaftor has been approved since 2018 from the 1st year of life and since 2020 from the 4th month of life for patients with the G551D, G1244E, G1349D, G178R, G551S, S1251N, S1255P, S549N or S549R mutations. For patients with the R117H mutation, ivacaftor has been approved in over 18s since 2018, since 2020, also from 4 months of age

[2] Lumacaftor/ivacaftor has been approved for F508del homozygous patients since 2018 from 6 years of age. and since 2019 from 2 years of age.

[3] Tezacaftor/ivacaftor has been approved for F508del homozygous patients from 12 years of age since 2019. or for F508del heterozygous patients with any of the following mutations: P67L, R117C, L206W, R352Q, A455E, D579G, 711+3A->G, S945L, S977F, R1070W, D1152H, 2789+5G->A, 3272-26A->G and 3849+10kbC->T.

[4] Elexacaftor/tezacaftor/ivacaftor has been available since 2020 from age 12 for F508del homozygous patients or F508del heterozygous patients with a minimal function (MF) mutation and since 2021 for all patients aged 12 and older and since 2022 from the age of 6 with at least one F508del mutation. It was possible to take elexacaftor/tezacaftor/ivacaftor before 2020 as part of clinical trials.

Therapies

9b. Indication therapy

9b.i Children and adolescents under 18 years

Indication therapy	0 – 5 years	6 – 11 years	12 – 17 years	Total
CFTR-Modulation	25.1	68.2	79.7	58.6
of these, Ivacaftor[1]	2.2	3.5	3.8	3.2
of these, Lumacaftor/ Ivacaftor[2]	21.7	28.6	5.8	18.8
of these, Tezacaftor/ Ivacaftor[3]	0.0	2.6	2.2	1.6
of these, Elexacaftor/ Tezacaftor/ Ivacaftor[4]	1.3	60.5	73.6	46.4
Inhalative antibiotics *with chronic Pseudomonas infection*	70.6	82.0	83.9	82.4
of these, inhalative tobramycin	41.2	50.8	50.7	50.0
of these, inhalative colistin	58.8	55.7	56.1	56.2
of these, inhalative aztreonam	0.0	9.8	13.0	11.2
of these, DPI tobramycin	0.0	1.6	6.9	4.9
of these, DPI colistin	0.0	4.9	12.2	9.3
of these, levofloxacin	0.0	3.3	1.4	1.8
of these, inhalative gentamicin	0.0	1.6	0.0	0.5
of these, others	0.0	1.6	0.7	0.9
Azithromycin with *chronic Pseudomonas infection*	5.9	24.6	19.2	19.6
Ursodeoxycholic acid *with liver disease*	75.8	81.9	81.9	81.1
Dietary measures *with Diabetes mellitus*	50.0	55.0	36.8	39.8
Insulin therapy *with Diabetes mellitus*	50.0	65.0	58.9	59.7
Oral antidiabetics *with Diabetes mellitus*	0.0	10.0	11.2	10.9
Pancreatic enzymes *with exocrine pancreatic insufficiency*	98.7	99.3	99.2	99.1
Additional nutrition *with underweight*	43.5	64.8	54.9	53.8
Additional oral nutrition	38.8	61.6	47.8	48.8
PEG	2.0	3.2	4.4	3.1
Proton pump inhibitors (PPI) *with gastroesophageal reflux*	100.0	94.1	78.6	86.3
Polyethylene glycol *with DIOS*	82.4	59.1	85.0	74.6
Calcium *with osteoporosis/osteopenia*	0.0	50.0	16.7	22.2
Oxygen *with respiratory insufficiency*	25.0	14.3	71.4	36.4

Table 32: pwCF under the age of 18 with indication therapy (frequencies in %) 2022

09

Therapies

9b.Indication therapy

9b.ii Adults 18 years and older

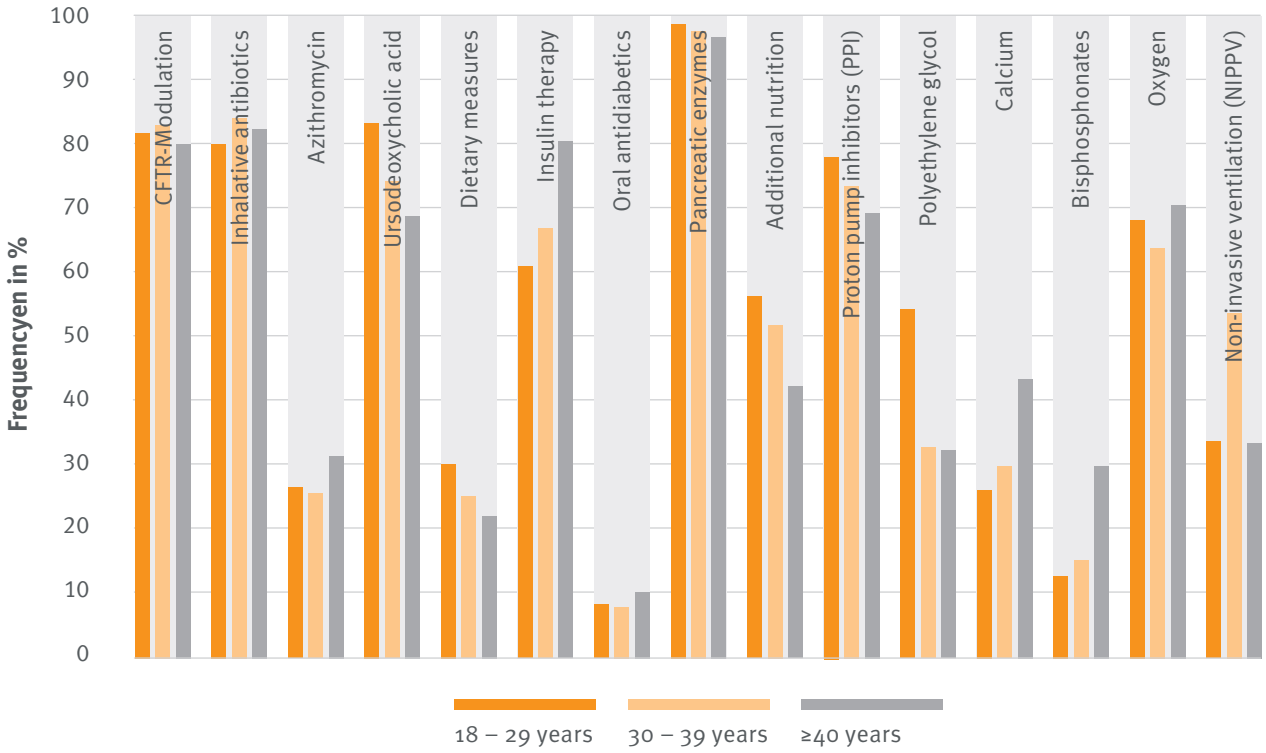

Figure 34: pwCF aged 18 and over with indication therapy 2022

Status of the approval of CFTR modulators 31.12.2022
Only the last modulator used in the reporting year is shown here.

[1] Ivacaftor has been approved since 2018 from the 1st year of life and since 2020 from the 4th month of life for patients with the G551D, G1244E, G1349D, G178R, G551S, S1251N, S1255P, S549N or S549R mutations. For patients with the R117H mutation, ivacaftor has been approved in over 18s since 2018, since 2020, also from 4 months of age

[2] Lumacaftor/ivacaftor has been approved for F508del homozygous patients since 2018 from 6 years of age. and since 2019 from 2 years of age.

[3] Tezacaftor/ivacaftor has been approved for F508del homozygous patients from 12 years of age since 2019. or for F508del heterozygous patients with any of the following mutations: P67L, R117C, L206W, R352Q, A455E, D579G, 711+3A->G, S945L, S977F, R1070W, D1152H, 2789+5G->A, 3272-26A->G and 3849+10kbC->T.

[4] Elexacaftor/tezacaftor/ivacaftor has been available since 2020 from age 12 for F508del homozygous patients or F508del heterozygous patients with a minimal function (MF) mutation and since 2021 for all patients aged 12 and older and since 2022 from the age of 6 with at least one F508del mutation. It was possible to take elexacaftor/tezacaftor/ivacaftor before 2020 as part of clinical trials.

Therapies

9b. Indication therapy

9b.ii Adults 18 years and older

Indication therapy	18 – 29 years	30 – 39 years	≥40 years	Total
CFTR-Modulation	81.3	82.7	80.1	81.4
of these, Ivacaftor[1]	3.1	2.1	3.5	2.9
of these, Lumacaftor/ Ivacaftor[2]	1.4	1.0	0.2	1.0
of these, Tezacaftor/ Ivacaftor[3]	2.2	2.2	3.5	2.5
of these, Elexacaftor/ Tezacaftor/ Ivacaftor[4]	76.2	78.7	74.5	76.5
Inhalative antibiotics *with chronic Pseudomonas infection*	80.0	83.5	81.9	81.8
of these, inhalative tobramycin	27.9	23.9	18.0	23.2
of these, inhalative colistin	49.1	49.6	50.1	49.6
of these, inhalative aztreonam	20.6	31.3	33.4	28.6
of these, DPI tobramycin	13.5	13.9	6.9	11.4
of these, DPI colistin	15.1	15.5	14.6	15.1
of these, levofloxacin	8.0	11.2	17.4	12.3
of these, inhalative gentamicin	0.2	0.3	0.0	0.2
of these, others	2.8	2.8	3.1	2.9
Azithromycin with *chronic Pseudomonas infection*	26.1	25.4	30.9	27.5
Ursodeoxycholic acid *with liver disease*	82.9	73.9	68.6	77.0
Dietary measures *with Diabetes mellitus*	29.8	25.1	21.6	25.5
Insulin therapy *with Diabetes mellitus*	60.8	66.5	80.3	69.3
Oral antidiabetics *with Diabetes mellitus*	7.9	7.7	10.0	8.6
Pancreatic enzymes *with exocrine pancreatic insufficiency*	98.1	97.3	96.3	97.4
Additional nutrition *with underweight*	56.0	51.5	41.7	52.8
Additional oral nutrition	48.7	48.5	40.8	47.4
PEG	7.9	4.6	2.0	6.2
Proton pump inhibitors (PPI) *with gastroesophageal reflux*	77.7	73.2	68.8	73.3
Polyethylene glycol *with DIOS*	54.0	32.5	32.0	43.0
Calcium *with osteoporosis/osteopenia*	25.5	29.6	42.8	34.2
Bisphosphonates *with osteoporosis*	12.5	14.6	29.5	21.2
Oxygen *with respiratory insufficiency*	67.8	63.5	70.2	67.6
Non-invasive ventilation (NIPPV) *with respiratory global insufficiency*	33.3	53.3	33.3	39.2

Table 33: pwCF aged 18 and over with indication therapy (frequencies in %) 2022

09

Therapies

9c. Development of CFTR modulation therapy 2018 – 2022

For the presentation of CFTR modulation therapy, all patients without transplantation and with modulator use were taken into account or all patients for whom a suitable modulator is approved in the respective reporting year.

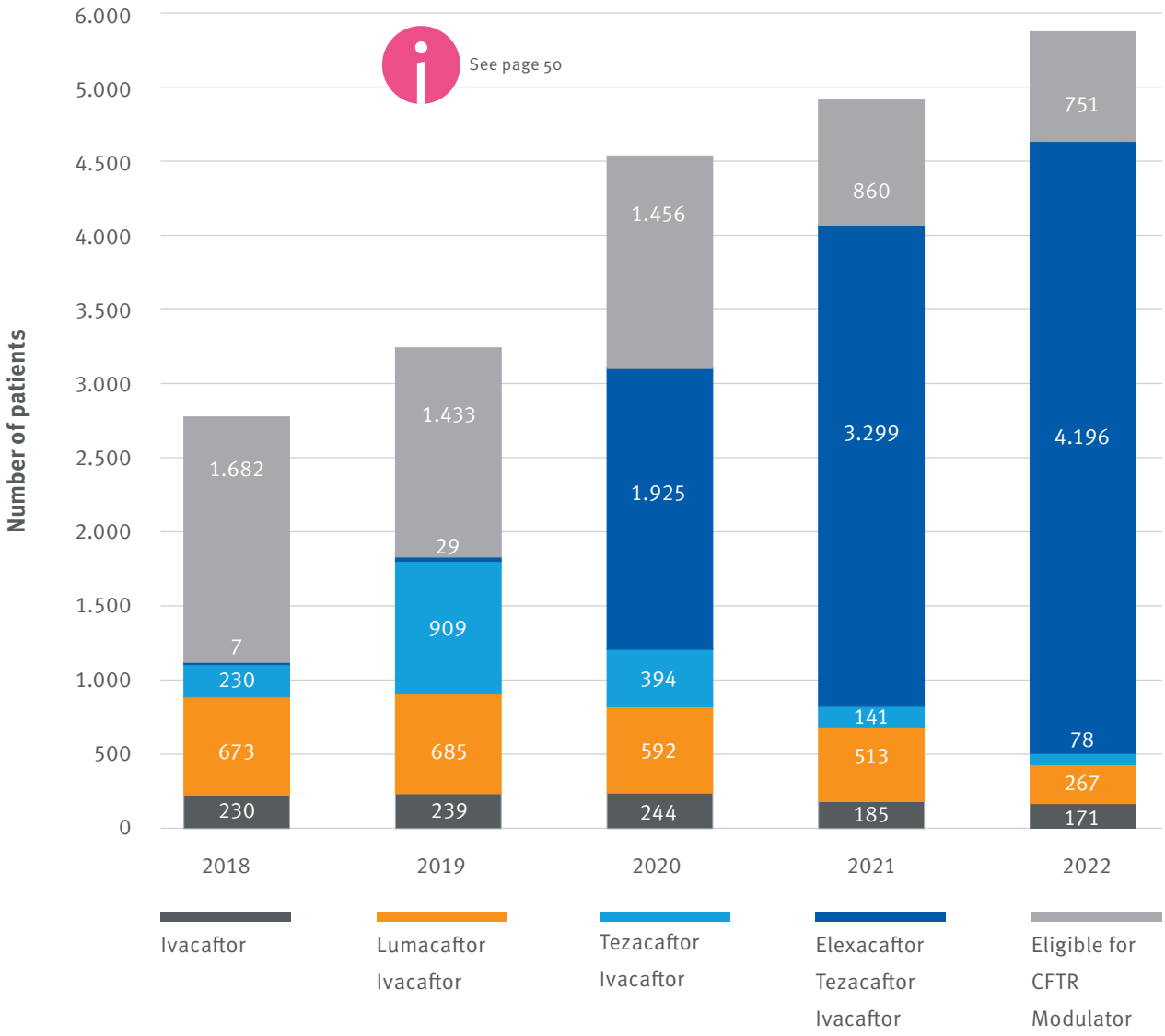

Figure 35: Number of pwCF with modulation therapy and number of patients for whom a suitable modulator is approved 2018 – 2022

Mortality

32 patients (19 girls/women and 13 boys/men) died in the reporting year 2022. The main causes of death were cardiopulmonary diseases (40.6 %), transplants (3.1 %), liver diseases/failure (6.3 %) and malignant diseases (9.4 %). Malignant diseases (9.4 %). Other or unknown causes were present in 37.4% of cases. The age at death is distributed as follows:

	Mean value	Median	Minimum	Maximum	25th percentile	75th percentile
Age at death in full years	39.0	37.0	7.0	79.0	22.5	54.0

Table 34: Age at death 2022

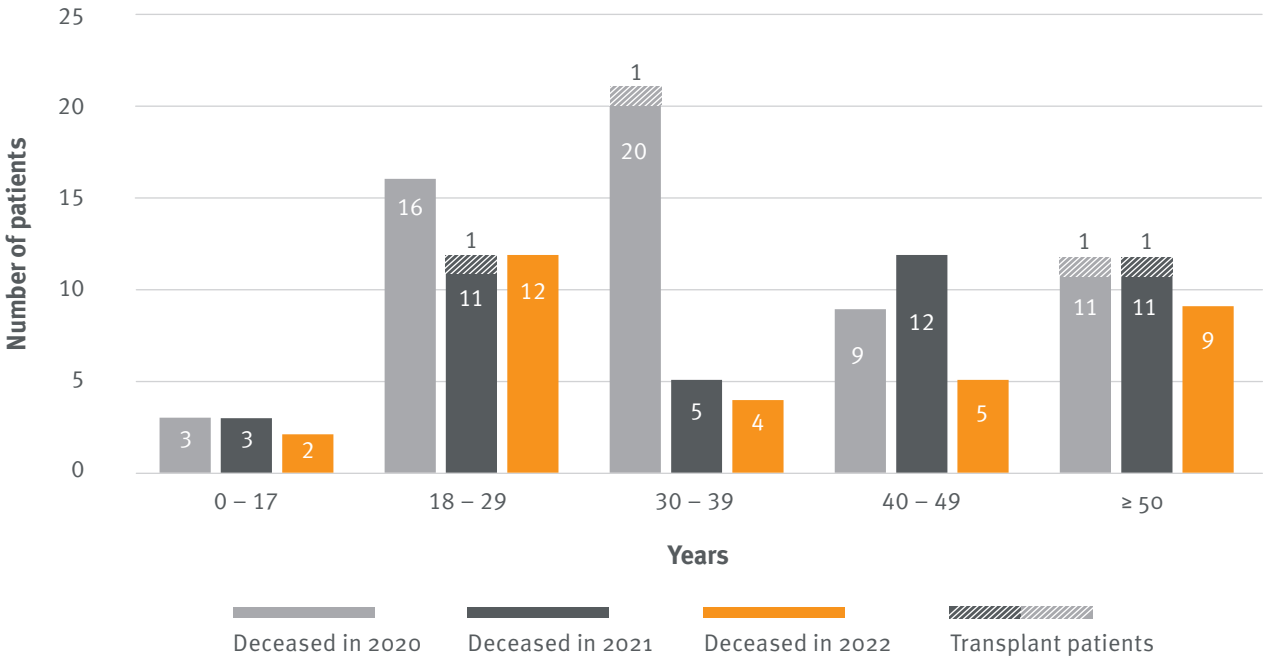

Figure 36: PwCF who died in the years 2020 – 2022

Age (years)	2020				2021				2022			
	No TX		TX		No TX		TX		No TX		TX	
	Number	%	Number	%	Number	%	Number	%	Number	%	Number	%
0 – 17	3	100.0	0	0.0	3	100.0	0	0.0	2	100.0	0	0.0
18 – 29	16	100.0	0	0.0	11	91.7	1	8.3	12	100.0	0	0.0
30 – 39	20	95.2	1	4.8	5	100.0	0	0.0	4	100.0	0	0.0
40 – 49	9	100.0	0	0.0	12	100.0	0	0.0	5	100.0	0	0.0
≥ 50	11	91.7	1	8.3	11	91.7	1	8.3	9	100.0	0	0.0
Total	59	–	2	–	42	–	2	–	32	–	0	–

Table 35: Deceased pwCF 2020 – 2022

10

Mortality

(Status: 28.09.2023)

The lifespan is described by the average age at death, the median survival age and the average age-specific life expectancy. We will present these statistical values in this annual data report on the basis of internationally accepted and comparable analytical methods. Owing to the higher number of patients lost from the follow-up for the reporting year 2022, we decided to report the current median age at death for the year 2022 as well as the average survival age and the life expectancy with respect to the period 2017 – 2021.

Average age at death

The average age at death for a given year describes the age at which half of the patients died. The average age at death was 37 (43) years in the reporting year 2022 (2021).

Median survival age 2021

The median survival age describes the expected age at which only 50 % of the patients are still alive. A COX PH regression analysis according to Sykes (Journal of Clinical Epidemiology 2016; 70: is conducted over a 5-year period to compensate for variations in the annual number of deaths. 8107 people with Cystic Fibrosis (including patients with transplants) and 316 deaths were recorded In the 5-year window between 2017 and 2021. 2017 patients (6.3%) were lost from the follow-up. The median survival age was 56.9 years (confidence interval: 55.1 to 58.9).

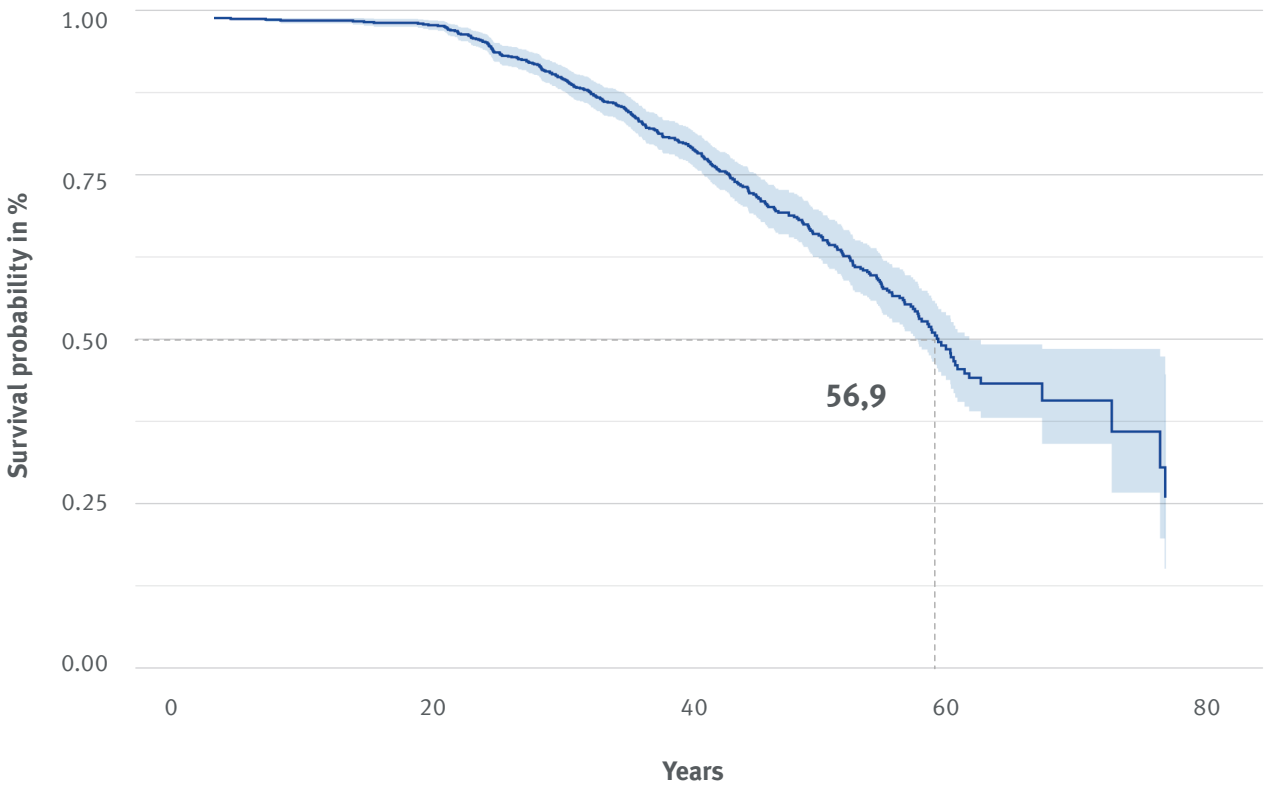

Figure 37: Median survival age for pwCF for the period 2017 – 2021

Mortality

Life expectancy 2022

Life expectancy is the average time a person can be expected to live from a specified age until death. It is calculated for a fixed period of time and is based on current and age-specific death rates. Currently the life expectancy of a healthy male newborn in Germany is 78 years and that of a female newborn 83 years (www.statista.de). The life expectancy is different for each age and does not correspond to the median survival age.

All statistical values refer to the population of Cystic Fibrosis patients in Germany, who vary greatly from individual to individual. As a result, only allow limited conclusions can be drawn about the individual. According to the literature, important influencing factors include gender, the existing gene mutation and the exocrine pancreatic function. All calculations are based on the current death rate, which has fortunately been steadily decreasing over the past years.

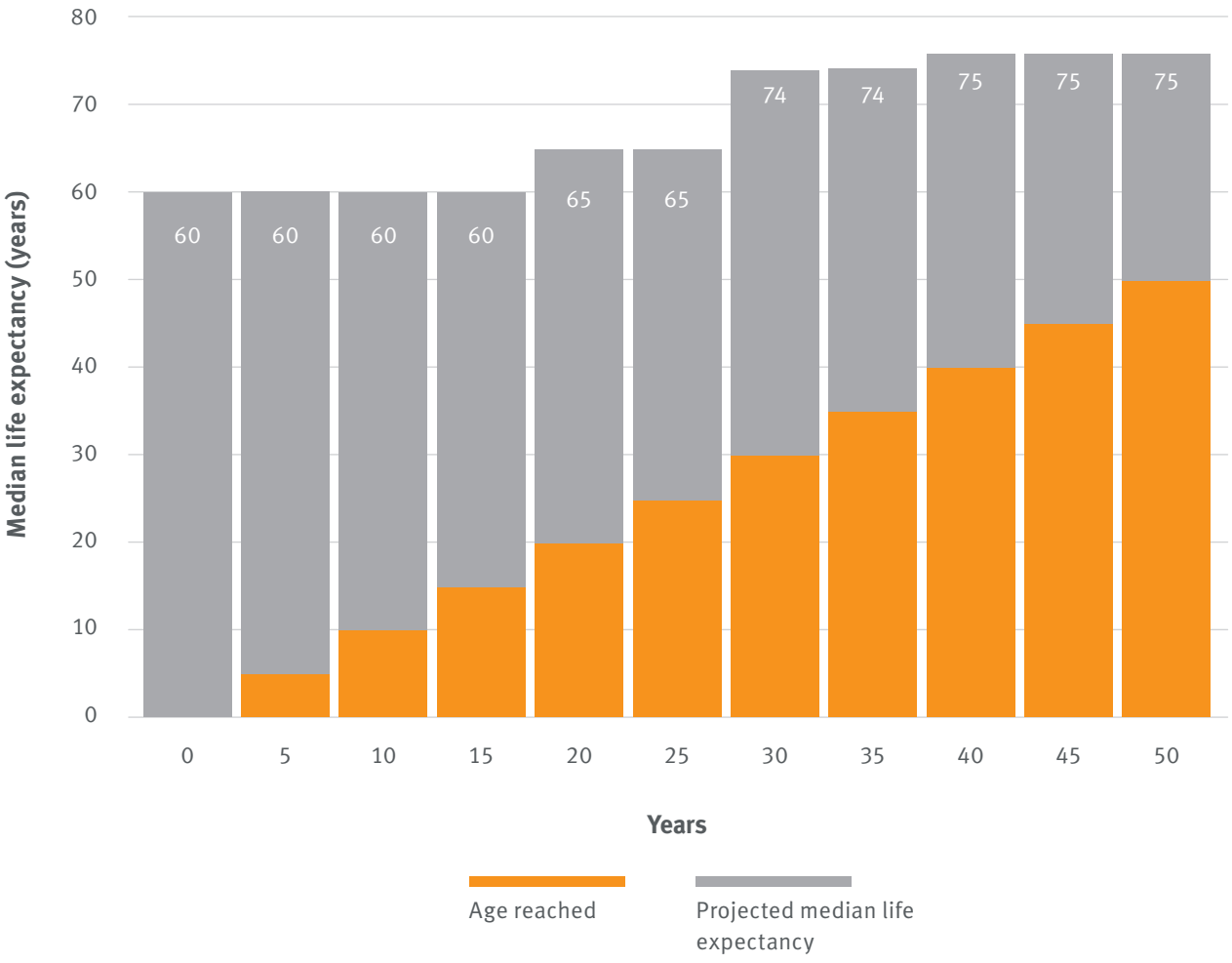

Figure 38: Projected median life expectancy for pwCF 2017 – 2021

11

Structure of care

11a. Size of the participating CF-centers

85 facilities participated in the cystic fibrosis registry in the reporting year 2022. 42 facilities cared for fewer than 50 patients and 43 facilities cared for more than 50 patients. More than 85% of the patients documented in the patients documented in the registry.

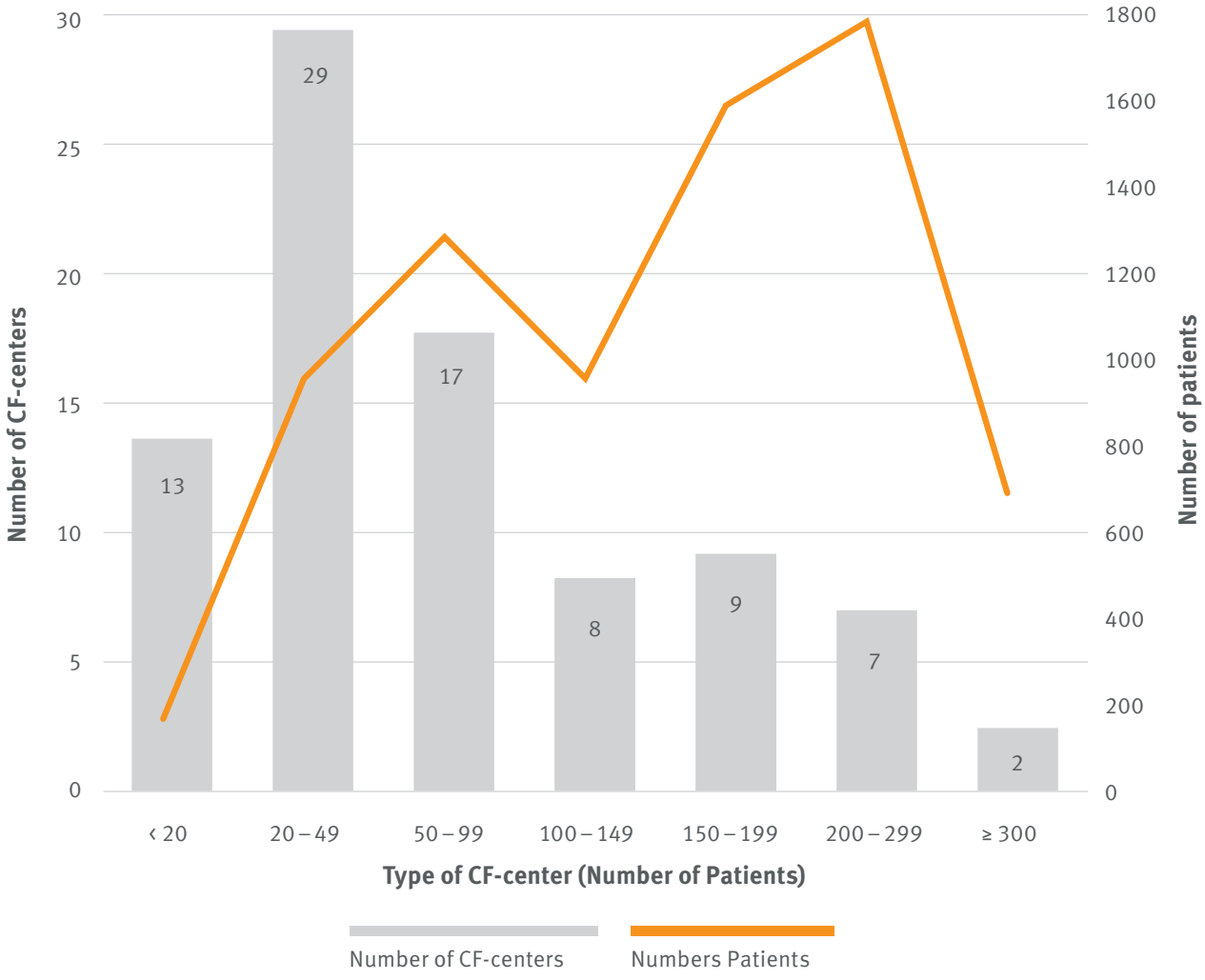

Figure 39: Number of documented pwCF and number of CF-centers in 2022

Structure of care

11b. Outpatient care

All patients with annual data in 2022 (n=6,973) were included in the following analyses.

	Age (years)						
	0 – 5	6 – 11	12 – 17	18 – 29	30 – 39	≥ 40	Total
Physiotherapy *in the outpatient clinic*	67.9	65.6	59.9	50.5	41.5	45.4	53.6
Nutritional therapy *in the outpatient clinic*	57.2	49.5	42.9	20.0	16.5	16.2	30.6
Psychosocial support *in the outpatient clinic*	47.6	42.7	45.7	32.5	21.3	25.4	34.4
Anxiety and depression screening[1]	–	–	28.2	32.8	33.6	36.0	32.8
Imaging Thorax	52.1	54.8	53.3	48.9	41.9	47.4	49.2
Bildgebung Abdomen	67.3	65.1	65.2	53.0	51.6	53.6	58.0
Bildgebung Bone density measurement	0.0	0.4	3.7	8.8	11.0	16.5	7.5
Laboratory	90.8	95.4	95.4	96.7	96.6	96.9	95.6
Rehabilitation stay	4.5	7.3	6.1	3.2	4.2	6.9	5.1
oGT test[2] *in patients without diabetes mellitus in the previous year*	0.0	13.0	44.4	31.5	31.8	22.8	24.4

Table 36: PwCF with outpatient care (frequencies in %) 2022,
[1] related to pwCF aged 12 and over; [2] related to pwCF aged 6 and over

11c. Outpatient visits

Outpatient visits	Outpatient visits in the reporting year			
	1	2	3	≥ 4
2018	7.2	10.7	20.7	61.4
2019	6.2	11.1	23.5	59.2
2020	7.8	14.9	25.0	52.3
2021	6.2	11.8	24.8	57.2
2022	6.0	13.3	30.7	50.0

Table 37: Development of the number of documented outpatient visits (frequencies in %) 2018 – 2022

11

Structure of care

11d. CF-related hospitalizations

Age (years)	Number of CF-relevant hospitalisations per patient						
	0	1	2	3	4	5+	unknown
0 – 5	75	18.2	4.2	1.2	0.3	0.2	0.9
6 – 11	79.2	16.2	2.3	0.8	0.5	0.2	0.8
12 – 17	78.1	14.2	4.3	1.4	0.4	0.4	1.2
18 – 29	78.9	13.7	4.3	1.7	0.3	0.4	0.7
30 – 39	82.6	12.2	.0	1.0	0.6	0.2	0.4
≥ 40	80.3	12.9	4.1	1.3	0.6	0.2	0.6
Total	79.2	14.3	3.7	1.3	0.5	0.3	0.7
< 18	77.5	16.2	3.5	1.1	0.4	0.3	1.0
≥ 18	80.4	13.1	3.8	1.4	0.5	0.3	0.6

Table 38: Number of cystic fibrosis-related hospitalizations per patient (frequencies in %) 2022

Structure of care

11d. CF-related hospitalizations

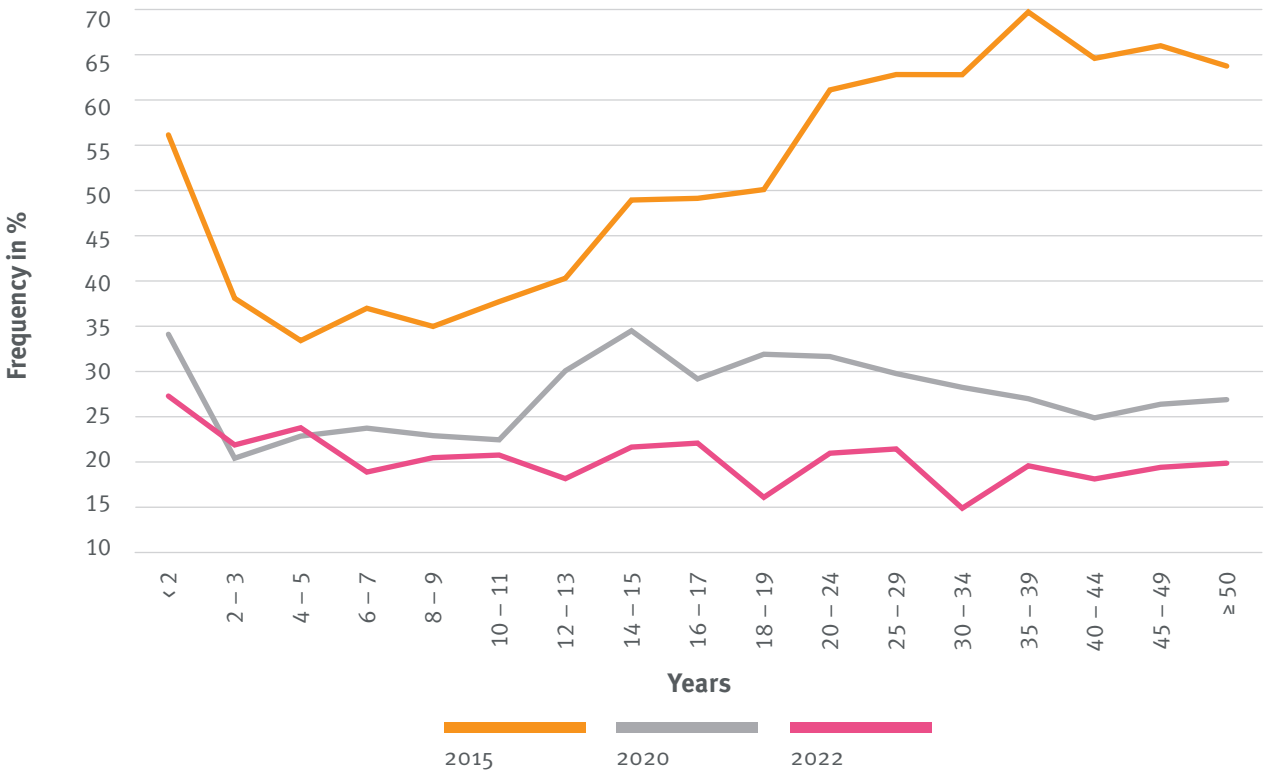

Figure 40: Development of the frequency (in %) of pwCF with at least 1 hospitalization 2015 – 2022

Years	Age (years)																
	< 2	2 – 3	4 – 5	6 – 7	8 – 9	10 – 11	12 – 13	14 – 15	16 – 17	18 – 19	20 – 24	25 – 29	30 – 34	35 – 39	40 – 44	45 – 49	≥ 50
2015	56.0	38.0	33.3	36.9	34.9	37.6	40.2	48.8	49.0	50.0	61.0	62.7	62.7	69.6	64.5	65.9	63.6
2020	34.1	20.4	22.8	23.7	22.9	22.4	30.1	34.5	29.2	31.9	31.6	29.8	28.2	27.0	24.8	26.4	26.9
2022	27.3	21.9	23.8	18.9	20.5	20.8	18.2	21.6	22.1	16.1	21.0	21.4	14.9	19.6	18.1	19.4	19.9

Table 39: Development of age-related frequencies (in %) of pwCF with at least 1 hospitalization 2015 – 2022

11

Structure of care

11e. Transplants

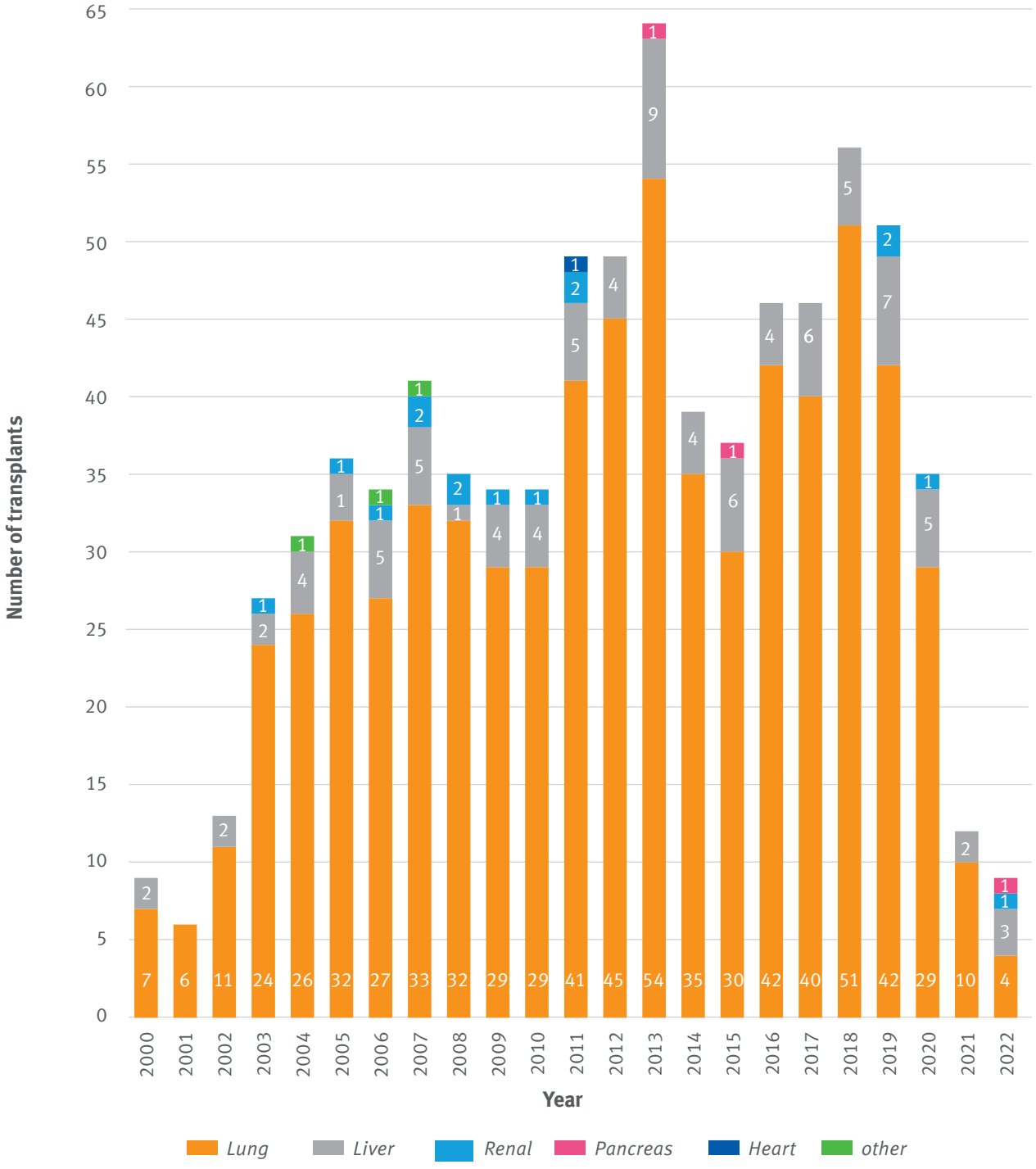

Figure 41: Number of transplants over the period from 2000 for the years 2000 – 2022

Structure of care

11e. Transplants

Reporting year	Lung	Liver	Renal	Pancreas	Heart	other	unknown
2000	7	2	0	0	0	0	0
2001	6	0	0	0	0	0	0
2002	11	2	0	0	0	0	0
2003	24	2	1	0	0	0	0
2004	26	4	0	0	0	1	0
2005	32	3	1	0	0	0	0
2006	27	5	1	0	0	1	0
2007	33	5	2	0	0	1	0
2008	32	1	2	0	0	0	0
2009	29	4	1	0	0	0	0
2010	29	4	1	0	0	0	0
2011	41	5	2	0	1	0	0
2012	45	4	0	0	0	0	0
2013	54	9	0	1	0	0	0
2014	35	4	0	0	0	0	0
2015	30	6	0	1	0	0	0
2016	42	4	0	0	0	0	0
2017	40	6	0	0	0	0	0
2018	51	5	0	0	0	0	0
2019	42	7	2	0	0	0	0
2020	28	5	1	0	0	0	0
2021	10	2	0	0	0	0	0
2022	4	3	1	1	0	0	0

Table 40: Number of transplants over time from the year 2000 for the years 2000 – 2022

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Overview of Registry requests

Receipt	Applicant	Institution	Subject / Title	Status
2017	Dittrich	Universität Heidelberg	Referenzperzentilen für FEV1 und BMI bei Mukoviszidose	Under evaluation
2017	Schwarz	Charité - Universitätsmedizin Berlin	Art4Fun/Schimmelpilz-assoziierte Erkrankungen	Completed – Published
2017	Prinz	Universität Ulm	Mukoviszidose und Glukosetoleranz	Completed – Published
2017	Grehn	Charité - Universitätsmedizin Berlin	Arthropathie bei Patienten mit Mukoviszidose	Completed
2017	Chiesi Farmaceutici S.p.A Chiesi Farmaceutici	---	Quinsair PASS	Completed
2018	Ballmann	Kinder-und Jugendklinik Universitätsmedizin Rostock	Diabetes Sonderauswertung	Completed
2018	Vertex Pharmaceuticals (Germany) GmbH	---	TEZ/IVA PASS	canceled
2018	Waldmann	Universität Erlangen/Nürnberg	Advanced Statistical Inference in Joint Models for Longitudinal and Time to Event Data	Under evaluation
2018	Hogardt	Universitätsklinikum Frankfurt	Prävalenz des B. Cepacia-Komplex bei CF-Patienten	Completed – Published
2019	Steindor/ Ringshausen	Universitätsklinik Essen/ Medizinische Hochschule Hannover	NTM bei CF-Patienten in Deutschland	Completed
2019	Moos-Thiele/ Muko.fit	Mukoviszidose e.V.	Kontrollgruppe aus Register zur Überprüfung der Repräsentanz der Muko.fit Gruppe	Under preparation
2019	Hebestreit	Universitätsklinikum Würzburg	Kontrollgruppe aus Register zur Überprüfung der Repräsentanz der VEMSE-Population	Completed
2019	Nährig/ Schulte-Hubbert	Klinikum der Universität München/ Uniklinikum Dresden	Datenanalyse zur antibiotischen Inhalationstherapie bei CF-Patienten mit chron. Pseudomonas-Infektion	Completed
2019	Stanke	Medizinische Hochschule Hannover	Genetische Prädiktoren für schwere CF bei europäischen Zwillingen und Geschwistern	Under evaluation
2019	Hogardt	Universitätsklinikum Frankfurt	Molekulare Epidemiologie von Mycobacterium abscessus bei CF-Patienten aus Deutschland	Completed
2020	Vertex Pharmaceuticals (Germany) GmbH		Dossier Nutzenbewertung Triple-Therapie	Completed
2020	Eickmeier/Gardecki	Universitätsklinikum Frankfurt	Patient Science zur Erforschung Seltener Erkrankungen - eine bürgerwissenschaftliche Studie am Beispiel der Mukoviszidose	Completed
2020	Müller	Universität Siegen	Einfluß hormoneller Kontrazeptiva auf Pneumonien bei CF Patientinnen	Under evaluation
2020	Van Dullemen	Universitätsklinikum Frankfurt	Mutationsspezifische Therapie - Overweight bei CF (DMT 2020 Vortrag)	Completed
2020	Vertex Pharmaceuticals (Germany) GmbH	Vertex	Dossier Nutzenbewertung Triple-Therapie – Indikationserweiterung	Completed
2020	Vertex Pharmaceuticals (Germany) GmbH	---	ETI PASS	Under evaluation
2021	Vertex Pharmaceuticals (Germany) GmbH	Vertex	Dossier Nutzenbewertung Triple-Therapie	Completed
2022	Dittrich, Tümmler	Medizinische Hochschule Hannover	MicroChange: Changes in Culture-dependent Microbiology after initiation of highly-effective CFTR modulator therapy	Completed – Published

Receipt	Applicant	Institution	Subject / Title	Status
2022	Splisense, Israel		Number of CF patients carrying the 3849 +10kb C->T mutation in Germany	Completed
2022	Nährig, Smaczny	Klinikum der Universität München/ Universitätsklinikum Frankfurt	Verlauf von Schwangerschaften	Under evaluation
2022	Sutharsan/ Mukoviszidose Institut gGmbH	Universitätsmedizin Essen/ ETI Publikationsinitiative	Impact of elexacaftor/tezacaftor/ivacaftor on lung function, nutritional status, pulmonary exacerbation frequency and sweat chloride in people with cystic fibrosis: real-world evidence from the German CF Registry	Completed – Published
2022	Steindor/ Ringshausen	Universitätsmedizin Essen/ Medizinische Hochschule Hannover	Epidemiological trends in nontuberculous mycobacterial infection among people with cystic fibrosis in Germany, 2023	Completed – Published
2023	Staab	Charité Universitätsmedizin Berlin	Ergänzende Daten zum Coach Projekt	Completed
2023	Vertex Pharmaceuticals (Germany) GmbH	Vertex	Antrag Registerauswertung G-BA Nutzendossier Indikationserweiterung Ivacaftor/Tezacaftor/Elexacaftor	Completed
2023	Stahl	Charité Universitätsmedizin Berlin	ETI off label use	Under evaluation
2023	Stahl	Charité Universitätsmedizin Berlin	Azithromycin	Under evaluation
2023	Vertex Pharmaceuticals (Germany) GmbH	Vertex	Antrag Registerauswertung G-BA Nutzendossier Indikationserweiterung Ivacaftor/Tezacaftor/Elexacaftor	Under evaluation
2023	Athing	Universitätsklinikum Leipzig	Every CFTR variant counts – Target-capture based next-generation-sequencing for molecular diagnosis in the German CF Registry	Completed – Published
2023	Welsner/ Mukoviszidose Institut gGmbH	Universitätsmedizin Essen/ ETI Publikationsinitiative	ETI Non-Responder	Under evaluation
2023	Prenzel/ Mukoviszidose Institut gGmbH	Universitätsklinikum Leipzig/ ETI Publikationsinitiative	Efficacy of CF triple modulators is associated with therapy reduction in a real-world analysis	Under preparation

13

Participating CF centers 2022

City	CF center	Department	Documentation level[1]	Number of patients[2]
Aachen	Luisenhospital Aachen	Mukoviszidose-Zentrum für Erwachsene, Innere Medizin	Level 1	98
Aachen	Kinderarztpraxis Laurensberg	Aachener Mukoviszidose Ambulanz für Kinder und Jugendliche	Level 1	46
Aue	HELIOS Klinikum Aue - CF-Ambulanz	Klinik für Kinder- und Jugendmedizin	Level 1	11
Augsburg	KJF Klinik Josefinum	Klinik für Kinder- und Jugendmedizin Sozialpädagogisches Zentrum (SPZ) Mukoviszidose-Ambulanz	Level 2	21
Augsburg	Universitätsklinikum Augsburg	II. Klinik für Kinder und Jugendliche, Kinderpneumologie - Allergologie, Mukoviszidose Ambulanz	Level 1	26
BadenBaden	Klinikum Mittelbaden GmbH	Baden-Baden Balg, Lungenzentrum, Mukoviszidose Ambulanz	Level 1	8
Berlin	Charité	Christiane Herzog-Zentrum Berlin, Klinik für Pädiatrie m. S. Pädiatrische Pneumologie und Immunologie	Level 2	286
Berlin	Sana Klinikum Lichtenberg	Oskar-Ziethen-Krankenhaus, Klinik für Kinder- und Jugendmedizin, Pneumologie, Mukoviszidose-Zentrum, Allergologie	Level 2	62
Berlin	Kinderarztpraxis Karow	Kinder- und Jugendmedizin, Kinderpneumologie	Level 1	16
Bielefeld	Evangelisches Klinikum Bethel gGmbH	Lehrkrankenhaus der Universität Münster, Klinik für Kinder- und Jugendmedizin, Tagesklinik für Allergologie und Pneumologie	Level 2	26
Bochum	Universitätsklinikum der Ruhr-Universität Bochum	St. Josef-Hospital am Katholischen Klinikum Bochum, Klinik für Kinder- und Jugendmedizin, Christiane Herzog Zentrum Ruhr (CHCR), Sekretariat Sylvia Meier	Level 2	72
Brandenburg	Medizinische Hochschule Brandenburg (MHB) Klinikum West-Brandenburg	Kinder- und Jugendklinik, CF-Ambulanz	Level 2	36
Bremen	Klinikum Bremen Mitte	Eltern-Kind-Zentrum Prof. Hess, Christiane Herzog Ambulanz für Mukoviszidose	Level 2	95
Chemnitz	Poliklinik Chemnitz gGmbH	Praxis für Kinder- und Jugendmedizin	Level 1	42
Cottbus	Carl-Thiem-Klinikum Cottbus gGmbH	Interdisziplinäre Studienzentrale	Level 2	27
Dresden	Medizinische Fakultät der TU Dresden	Klinik und Poliklink für Kinder- und Jugendmedizin, Mukoviszidose-Centrum "Christiane Herzog"	Level 2	184
Düsseldorf	UKD Universitätsklinikum Düsseldorf	Klink für Allgemeine Pädiatrie, Neonatologie und Kinderkardiologie, Ambulanz für Kinderpneumologie und Allergologie	Level 1	36
Erfurt	HELIOS Klinikum Erfurt	Kinderklinik und Jugendmedizin, CF-Ambulanz	Level 1	22
Erlangen	Universitätsklinikum Erlangen	Kinder- und Jugendklinik, Sozialpädiatrisches Zentrum	Level 2	184
Essen	Universitätsmedizin Essen	Ruhrlandklinik - Pneumologie	Level 2	342
Essen	Universitätsklinikum Essen	Zentrum für Kinder- und Jugendmedizin, Pädiatrische Pneumologie und Schlafmedizin, Christiane Herzog Centrum Ruhr	Level 2	103
Frankfurt	Universitätsklinikum Frankfurt	Goethe Universität, Christiane Herzog CF-Zentrum für Kinder, Jugendliche und Erwachsene	Level 2	261
Frankfurt	St. Elisabethen Krankenhaus	Katharina-Kaspar Kliniken, Innere Medizin - Pneumologie	Level 2	25
Freiburg	Universitätsklinikum Freiburg	Klinik für Allgemeine Kinder- und Jugendmedizin, Ambulanz und Arbeitsgruppe Pneumologie, Allergologie und Mukoviszidose	Level 2	78
Freiburg	Universitätsklinikum Freiburg	Abteilung Pneumologie, Erwachsenenambulanz	Level 2	49
Gießen	Universitätsklinik Gießen und Marburg GmbH	Zentrum für Kinder- und Jugendmedizin, Mukoviszidose-Zentrum	Level 2	104
Gießen	Universitätsklinik Gießen und Marburg GmbH	Zentrum für Kinder- und Jugendmedizin, Mukoviszidose-Zentrum	Level 2	99

[1] See the collective description for the definition of the documentation level, [2] patients may have been documented in several outpatient clinics

City	CF center	Department	Documentation level[1]	Number of patients[2]
Greifswald	Ernst-Moritz-Arndt Universität	Universitätsmedizin Greifswald, Klinik und Poliklinik für Kinder- und Jugendmedizin	Level 2	24
Halle	Universitätsklinikum Halle (Saale) (UKH)	Medizinische Fakultät der Martin-Luther-Universität, Mukoviszidose-Zentrum	Level 2	83
Hamburg	Kinder- und Jugendärztliche Gemeinschaftspraxis	Kinderärzte im Friesenweg, CF Centrum Altona	Level 2	141
Hamburg	Universitätsklinikum Eppendorf	II. Medizinische Klinik - Sektion Pneumologie	Level 2	49
Hamm	Evangelisches Krankenhaus Hamm (EVK) gGmbH	Klinik für Kinder- und Jugendmedizin, Pulmologie/Allergologie	Level 1	13
Hannover	Medizinische Hochschule Hannover	Klinik für Pädiatrische Pneumologie, Allergologie und Neonatologie	Level 2	193
Hannover	Medizinische Hochschule Hannover	Klinik für Innere Medizin, Pneumologische Ambulanz (Erwachsene)	Level 2	246
Heidelberg	Universitätsklinikum Heidelberg	Sektion Pädiatrische Pneumologie, Allergologie und Mukoviszidose-Zentrum	Level 2	124
Heidelberg	Thoraxklinik am Universitätsklinikum Heidelberg	Abteilung für Pneumologie und Beatmungstherapie, CF Ambulanz für Erwachsene	Level 2	234
Heilbronn	SLK-Kliniken Heilbronn GmbH	Klinik für Kinder- und Jugendmedizin, Klinikum am Gesundbrunnen, Perinatalzentrum	Level 2	19
Homburg	Universitätsklinikum des Saarlandes	Klinik für Allgemeine Pädiatrie und Neonatologie	Level 2	66
Homburg	Universitätsklinikum des Saarlandes	Innere Medizin 5, CF-Ambulanz für Erwachsene	Level 1	57
Jena	Universitätsklinikum Jena	Klinik für Kinder- und Jugendmedizin, Ambulanz für Pädiatrische Pneumologie, Allergologie, Mukoviszidosezentrum	Level 2	172
Karlsruhe	Städtisches Klinikum Karlsruhe gGmbH	Klinik für Kinder- und Jugendmedizin	Level 2	26
Kassel	Klinikum Kassel	Klinik für Kinder- und Jugendmedizin, Pneumologie, CF-Ambulanz	Level 2	48
Kiel	Städtisches Krankenhaus Kiel GmbH	Mukoviszidose-Zentrum für Erwachsene, 4. Medizinische Klinik	Level 2	117
Kiel	Städtisches Krankenhaus Kiel GmbH	Klinik für Kinder- und Jugendmedizin, Mukoviszidose-Zentrum	Level 2	50
Koblenz	Gemeinschaftsklinikum Mittelrhein gGmbH	Klinik für Kinder- und Jugendmedizin, Pädiatrische Pneumologie und Allergologie, Mukoviszidose Ambulanz	Level 2	45
Köln	Universitätsklinikum Köln	Klinik und Poliklinik für Kinder- und Jugendmedizin, Mukoviszidose-Zentrum	Level 2	247
Köln	Kliniken der Stadt Köln	Lungenklinik Merheim	Level 2	85
Krefeld	Helios Klinikum Krefeld	Zentrum für Kinder- und Jugendmedizin, Mukoviszidose-Ambulanz	Level 2	39
Leipzig	Universitätsklinikum Leipzig	Klinik und Poliklinik für Kinder- und Jugendmedizin, CF-Ambulanz	Level 2	69
Löwenstein	Fachklinik Löwenstein	Klinik für Pneumologie, Intensiv- und Beatmungsmedizin	Level 2	31
Lübeck	Universitätsklinkum Schleswig Holstein (UKSH)	Campus Lübeck, Klinik für Kinder- und Jugendmedizin, Pädiatrische Pneumologie	Level 1	31
Magdeburg	Otto-von-Guericke Universität Magdeburg	Universitätsklinik für Pneumologie	Level 2	18
Magdeburg	Otto-von-Guericke Universität Magdeburg	Klinik für Allgemeinpädiatrie und Neonatologie, CF-Ambulanz	Level 2	21
Mainz	Universitätsmedizin Mainz	Zentrum für Kinder- und Jugendmedizin - Pädiatrische Pneumologie, Allergologie, Mukoviszidose	Level 2	111

[1] See the collective description for the definition of the documentation level, [2] patients may have been documented in several outpatient clinics

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Participating CF centers 2022

City	CF center	Department	Documentation level[1]	Number of patients[2]
Mannheim	Universitätsmedizin Mannheim	Klinik für Kinder- und Jugendmedizin, Pulmologie, Infektiologie und Allergologie	Level 2	17
Marburg	Zentrum für Kinderheilkunde	Mukoviszidose-Ambulanz	Level 2	14
München	Kinderpoliklinik Schwabing	CF-Ambulanz	Level 2	38
München	LMU Klinikum der Universität München	Kinderklinik und Kinderpoliklinik im Dr. von Haunerschen Kinderspital, Christiane Herzog-Ambulanz	Level 2	339
München	LMU Klinikum der Universität München	Campus Innenstadt, Medizinische Klinik - Pneumologie	Level 2	233
München	Lungenheilkunde München Pasing	Mukoviszidose-Zentrum München West	Level 2	168
Münster	Clemenshospital	Mukoviszidose-Ambulanz	Level 2	129
Münster	Universitätsklinikum Münster UKM	Klinik für Kinder- und Jugendmedizin, Allgemeine Pädiatrie Mukoviszidose-Ambulanz	Level 1	61
Neubrandenburg	Dietrich Bonhoeffer Klinikum	Klinik für Kinder- u. Jugendmedizin	Level 2	22
Oldenburg	Klinikum Oldenburg AöR	Klinik für Pädiatrische Pneumologie und Allergologie, Neonatologie und Intensivmedizin	Level 1	99
Osnabrück	Christliches Kinderhospital Osnabrück	Zentrum für Kinder- und Jugendmedizin, Mukoviszidose Ambulanz	Level 2	62
Passau	Kinderklinik Dritter Orden	Zentrum für Kinder- und Jugendgesundheit, Sozialpädiatrisches Zentrum	Level 2	25
Potsdam	Klinikum Westbrandenburg gGmbH	Kinder- und Jugendklinik, Mukoviszidose Ambulanz	Level 2	255
Ravensburg	Oberschwabenklinik (OSK) gGmbH Ravensburg	Krankenhaus St. Elisabeth, Klinik für Kinder und Jugendliche	Level 1	4
Regensburg	Klinik Donaustauf	Pneumologische Ambulanz	Level 2	48
Regensburg	KUNO Klinik St. Hedwig	Kinder- und Jugendmedizin	Level 2	81
Rostock	Universitätsmedizin Rostock	Kinder- und Jugendmedizin	Level 2	32
Rüdersdorf bei Berlin	Immanuel Klinik Rüdersdorf	Kinder- und Jugendmedizin	Level 2	6
Schwerin	HELIOS Kliniken Schwerin	Kinder- und Jugendmedizin, Mukoviszidose-Ambulanz	Level 2	24
Stuttgart	Klinikum Stuttgart	Christiane Herzog Transitionszentrum	Level 2	176
Stuttgart-Gerlingen	Robert Bosch Krankenhaus RBK	Klinik Schillerhöhe, Mukoviszidose Ambulanz	Level 2	171
Trier	Klinikum Mutterhaus der Borromäerinnen gGmbH	Klinikum Mutterhaus Mitte, Innere Medizin 1	Level 1	26
Trier	Klinikum Mutterhaus der Borromäerinnen gGmbH	Kinder- und Jugendmedizin	Level 2	17
Tübingen	Universitätsklinik Tübingen	Klinik für Kinder- und Jugendmedizin, Mukoviszidose-Ambulanz	Level 2	157
Ulm	Universitätsklinikum Ulm	Klinik für Kinder- und Jugendmedizin, Mukoviszidose-Ambulanz	Level 2	115
Wangen	Fachkliniken Wangen gGmbH	Waldburg Zeil Kliniken, Klinik für Pneumologie	Level 2	26
Wangen	Fachkliniken Wangen gGmbH	Rehabilitationsklinik für Kinder und Jugendliche, CF-Ambulanz	Level 2	7
Wesel	Marienhospital Wesel	Akademisches Lehrkrankenhaus der Westfälischen Wilhelms-Universität Münster, Klinik für Kinder- und Jugendmedizin	Level 2	34
Worms	Klinikum Worms gGmbH	Klinik für Kinder- und Jugendmedizin	Level 2	51
Würzburg	Universitätsklinikum Würzburg	Kinderpoliklinik Christiane Herzog-Zentrum Unterfranken Mukoviszidose-Ambulanz	Level 2	164
Zwickau	Heinrich Braun Klinikum gGmbH	Standort Zwickau, Kinderzentrum	Level 1	11

[1] See the collective description for the definition of the documentation level, [2] patients may have been documented in several outpatient clinics

Glossary

Term	Definition
ABPA Allergic bronchopulmonary aspergillosis	Development of an allergic reaction to *Aspergillus fumigatus*.
Anticholinergics	An anticholinergic has a relaxing effect on the smooth musculature and inhibits secretion.
Arthritis	A condition which causes pain and inflammation in the joints.
Arthropathy	A condition which causes pain in the joints.
Pancreas	An organ in the digestive system which produces insulin and digestive enzymes.
ß2-sympathomimetics	Betasympathomimetics are pharmaceutical substances which stimulate the beta receptors of the sympathetic nervous system.
BMI (Body Mass Index)	A measure for evaluating a person's body weight in relation to their height.
Burkholderia cepacia	Burkholderia cepacia is a species of bacterium in the Burkholderia genus. Several of these bacteria are a potential threat to the health of people with Cystic Fibrosis.
CF (Cystic fibrosis)	Mucoviscidosis; Cystic Fibrosis
CFTR Regulator of the transmembrane conductance in Cystic Fibrosis	A protein on the cell surface which controls the sodium and water balance of a cell. The gene which causes Cystic Fibrosis is the blueprint for the CFTR protein. Every person has two copies of the gene for CFTR. Both CFTR genes must be affected by a mutation which causes CF, in order for someone to be born with Cystic Fibrosis.
Enzymes	Biological molecules present in the body (i.e. molecules occurring as metabolic products in the living cell) which support complex reactions such as the digestion of food.
FEV1 one-second capacity	The one-second capacity is the largest-possible quantity of air which can be forced out of the lungs within 1 second. The FEV1 value is part of the pulmonary function and can be measured in a pulmonary function test.
FEV1% predicted	The FEV1% is the percentage value of the average FEV1 which healthy people of the same age, gender and height can achieve. It is normally between 80 – 120%.
Gastroesophageal reflux disease	A chronic symptom of damage caused by gastric acid rising from the gastric mucosa.
Genotype	A characteristic part of the genetic structure of a cell, an organism or an individual.
Haemophilus influenza	Haemophilus influenza is a bacterium which can cause severe illness.
Haemoptysis	Coughing up blood.
Hepatobiliary disease	A liver or biliary disease.

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Glossary

Term	Definition
Heterozygous	Everyone living with Cystic Fibrosis has two mutations of the gene for CFTR. One mutation is inherited from the mother and one from the father. If both mutations (or genotypes) are different, the person is heterozygous.
Homozygous	Everyone living with Cystic Fibrosis has two mutations of the gene for CFTR. One mutation is inherited from the mother and one from the father. If both mutations (or genotypes) are the same, the person is homozygous.
Interquartile range	The interquartile range is a measure of dispersion in descriptive statistics. If the sample is sorted by size, it indicates the width of the interval in which the mean 50% of the sample elements lie. It shows the difference between the upper and lower quartile: IQR = Q3 - Q1.
Confidence interval	An expectancy range to express how confident we are about our statistical estimates of a clinical measure. It shows a series of results which are likely to include the correct values for the population under study. A narrow confidence interval indicates a more accurate estimate. A wide confidence interval indicates greater uncertainty about the exact value of the measurement, often because only a small group of patients was studied.
Digestive tract / Gastrointestinal tract (GI)	The gastrointestinal tract (GI) is the main part of the digestive system which extends from the oesophagus to the anus. The GI is an organ system responsible for digesting food, absorbing nutrients and excreting faeces.
Median	The middle number when all numbers are arranged from the smallest to the largest number.
Median survival prognosis	A mathematical formula which can be used to predict the age which half the people born with CF today will reach. For example: 50 % of the people born today will reach the age of at least 47. The other 50% of these people will probably die before they reach this age.
Mean value	An average value calculated by adding up all the values and dividing by the number of values.
Average age at death	The average age at death is based on the people with CF who died in one year.
MRSA	Methicillin-resistant Staphylococcus aureus is a bacterial species which is resistant to a series of widely-used antibiotics.
Mutation	A mutation is an alteration to a gene. If both parents of a child are carriers of a mutation which causes Cystic Fibrosis , there is a 25% chance that the child will have CF. There are over 1,400 different mutations of the CFTR gene.
Hepatobiliary disease	Small sacciform growths caused by chronic inflammation of the nasal mucosa.

Glossary

Term	Definition
Newborn screening	Newborn screening is an examination of newborns which aims to detect congenital diseases at an early stage, e.g. Cystic Fibrosis.
Non-tuberculous mycobacteria (NTM)	A mycobacterium which does not cause tuberculosis but can still be the cause of respiratory tract infections. Several types are known.
Osteopenia	A disease which is less severe than osteoporosis and in which the mineral content of bones is reduced.
Osteoporosis	A condition in which the bones become brittle due to the loss of tissue.
Percentile	A percentile indicates where a value is relative to the rest of the data. If a value is higher than 90% of the rest of the data, it is referred to as the 90th percentile.
Pneumothorax	An accumulation of air in the cavity between the lung and the chest wall which can cause a pulmonary collapse on the affected side.
Prevalence	The total number of people with this disease in the last 12 months.
Pseudomonas aeruginosa	A strain of bacteria which rarely affects healthy people but can lead to a variety of infections in a weakened immune system. These infections often become chronic.
Liver cirrhosis	A chronic liver disease.

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Notes

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