German Cystic Fibrosis Registry

Annual Report | 2023

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Status: 26.06.2024

MUKOVISZIDOSE e.V.
Helfen. Forschen. Heilen.

Impressum

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Publisher:
Mukoviszidose e.V. & Mukoviszidose Institut gGmbH
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Data Management &
Statistical Analyses:
Universitätsmedizin der
Johannes Gutenberg Universität
Interdisziplinäres Zentrum Klinische Studien (IZKS)
Langenbeckstraße 1, 55131 Mainz
www.izks-mainz.de

Typesetting & Layout:
hazel | GRAFIK + DESIGN
www.hazel-design.de

Foreword

Prof. Dr. med. Lutz Nährlich

Medical Director
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Registry

In addition to the current state of health of people with cystic fibrosis (CF) in Germany, the German Cystic Fibrosis Registry presents the development over the last two decades and the development for selected birth cohorts in this 2023 report.

The living population in the German Cystic Fibrosis Registry 2023 has reached a new high of 7,587 people with cystic fibrosis – 7,181 (95 % of the living population) with follow-up data. The year 2023 was marked by an expansion of the approval of CFTR modulator therapy (lumacaftor/ivacaftor) for children from 1 (instead of 2) year of age and of highly effective CFTR modulator therapy (elexacaftor/tezacaftor/ivacaftor) from 2 (instead of 6) years of age. In 2022, CFTR modulator therapy was available for 82 % of all people with cystic fibrosis in Germany and used in 85 % of this group. The report shows the medium-term effects on the health status of all people with cystic fibrosis. It should be noted that the presentation does not distinguish between people with CF with or without CFTR modulator therapy and therefore only partially reflects the individual effect. A particular highlight is the development of the median age of survival to 66.8 years for the current period (2019 – 2023); we have been able to optimize our follow-up and, in contrast to the previous year, are now reporting for the first time on the most recent period.

Stabilization of lung function continues for all birth cohorts. Among 18 – 19 year-olds, 81 % have an FEV1%pred of over 80 % compared to 32 % in 2000. The rate of chronic Pseudomonas infections has fallen to 7.2 % among children and adolescents and 48 % in adults, and due to the low detection rates, there is an increasing discrepancy between annual detection (24% across all MmM) and chronic infection according to the adapted Leeds criteria (31 % across all MmM). Another positive development is the high rate of CF patients without exacerbations treated with antibiotics (70%) and without hospitalizations (80 %). The decline in underweight, particularly in adults, is offset by an increase in overweight and obesity, which, however, is stable compared to the previous year. The basic and indication therapy shows a decline in the therapy load for the first time. All this underlines the continued need for close clinical care by the multidisciplinary cystic fibrosis team in order to jointly identify and discuss medium-term changes and challenges for the future. The German Cystic Fibrosis Registry can provide the (data) basis for this and has made it available to people with cystic fibrosis through the “MukoMe” project.

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. For this, I would like to express my sincere thanks to all outpatient teams and people with cystic fibrosis who, with their consent, enable us to document and evaluate their data. Many thanks to everyone involved in data entry and evaluation. I would also like to thank the Registry Working Group, the company Axaris (Ms. Jaumann, Mr. Müller, Mr. Volk) and the data management team of the Interdisciplinary Center for Clinical Studies (IZKS) at the University of Mainz (Ms. Wosniok, Ms. Wollscheid, Ms. Endres, Mr. Kronfeld, Mr. Ruckes). My special thanks go to Mr. Burkhart from the Cystic Fibrosis Institute for his tireless work in project management.

We hope you will continue to support the registry.

Collective description

The history data records of 7181 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 2023. In addition, patients without history data were also included in the evaluations of new CF diagnoses and mortality (16 newly diagnosed patients and 4 deceased patients without history data in 2023).

All 443 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 6837 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 2023

The map shows all 85 CF centers participating in the Registry in 2023.
A list of all CF facilities and the reported patient numbers can be found on page 66.
Write to us if your facility is not represented: mburkhart@muko.info

01

Brief overview

Brief overview	2000	2005	2010	2015	2020	2022	2023
Data status	26.06.24	26.06.24	26.06.24	26.06.24	26.06.24	26.06.24	26.06.24
Participating centers	78	83	81	89	88	85	85
Living Population [1, 2, 4]	4,249	5,209	5,897	6,658	7,442	7,480	7,587
Living patients with transplants[4]	66	172	291	452	564	468	443
Participating patients with annual data	3,614	4,691	5,085	5,732	6,629	6,962	7,181
of these, transplant patients	43	121	202	293	365	366	344
Age in years; Median[1]	14	16	18	20	21	22	23
Proportion of adults (≥ 18 years) in %	36.3	45.9	51.3	56.7	58.7	60.2	61.0
Male patients in %	52.4	52.1	51.9	52.0	52.0	51.6	51.9
New diagnoses in the reporting year[1]	227	209	220	228	236	190	146
Age for new diagnoses in years; Median[1]	1.4	0.9	1.0	0.9	0.2	0.1	0.1
of these, diagnosis via newborn screening	1.8	2.9	3.6	11.4	56.8	70.0	66.4
Maternities in the reporting year	8	9	16	23	37	57	63
Paternities in the reporting year	4	3	7	8	18	18	16
Deaths in the reporting year [1, 4]	43	72	75	100	61	34	22
Deaths: % of all patients[1]	1.2	1.5	1.5	1.7	0.9	0.5	0.3
Age at death in years; median	21	26	28	31,5	35	37	38,5
(25^{th} – 75^{th} pctl)	(15 – 28)	(19 – 35)	(24 – 37)	(25 – 37)	(28 – 46)	(23 – 53)	(24 – 50)
Transplant patients in the reporting year[1]	9	36	33	34	33	9	5
Lung transplants [3]	7	32	29	30	29	4	4
Liver transplants [3]	2	3	4	7	5	4	2
Renal transplantation[3]	0	1	1	0	1	1	0
Pancreas transplantation[3]	0	0	0	1	0	1	0

Table 1: Brief overview of pwCF with follow-up data, valid informed consent and cystic fibrosis diagnosis in the reporting years 2000 – 2023 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.
[3] Multiple answers possible
[4] Data status 05.9.2024

Patient numbers development

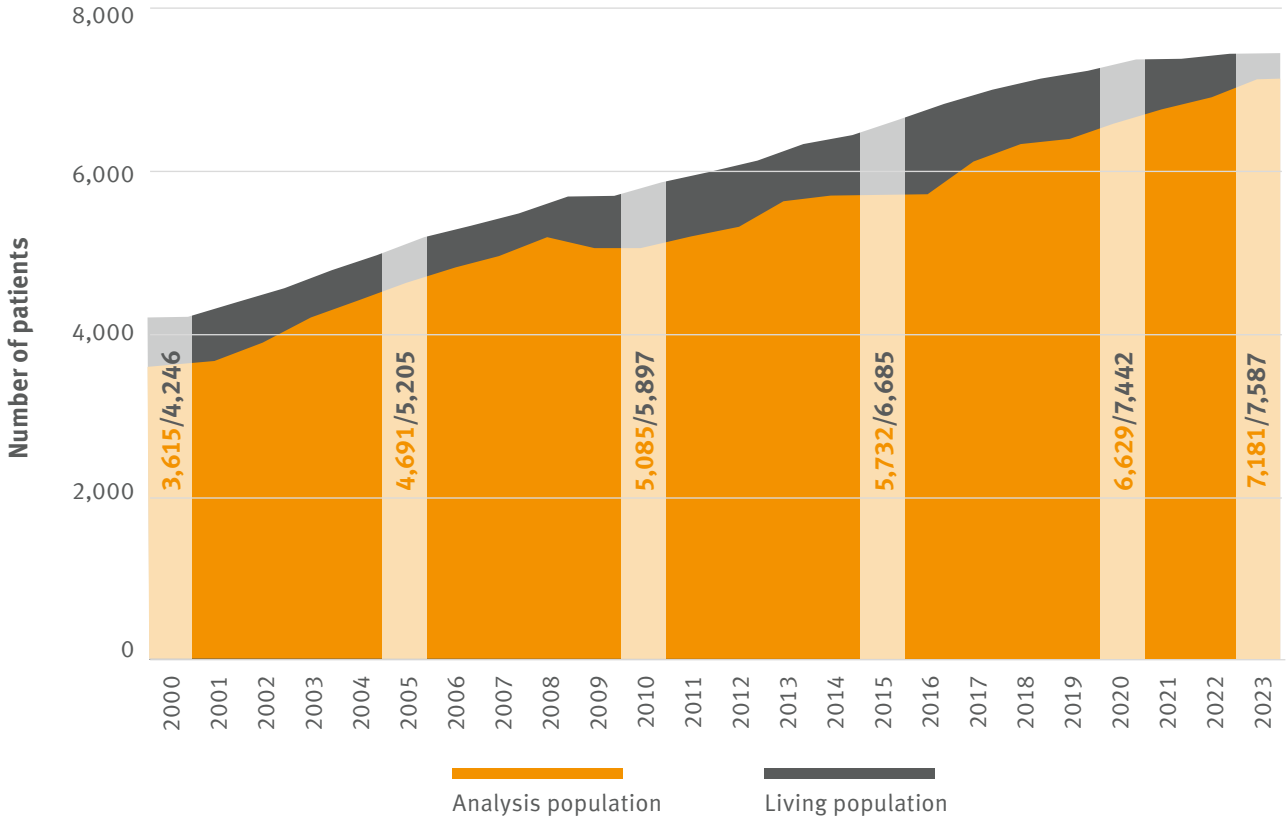

Figure 2: Number of pwCF documented in the registry 2000 – 2023

Reporting year	Analysis population	Living population
2000	3,614	4,249
2001	3,732	4,409
2002	3,940	4,578
2003	4,258	4,803
2004	4,437	4,985
2005	4,691	5,209
2006	4,846	5,370
2007	4,984	5,524
2008	5,222	5,712
2009	5,058	5,772
2010	5,085	5,897
2011	5,214	6,022

Reporting year	Analysis population	Living population
2012	5,352	6,172
2013	5,659	6,360
2014	5,728	6,505
2015	5,732	6,658
2016	5,779	6,848
2017	6,176	7,043
2018	6,370	7,154
2019	6,450	7,286
2020	6,629	7,442
2021	6,779	7,440
2022	6,962	7,480
2023	7,181	7,587

Table 2: Number of pwCF documented in the registry 2000 – 2023

03

Age structure

The age structure calculations include all 7181 pwCF with annual data for 2023. 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.

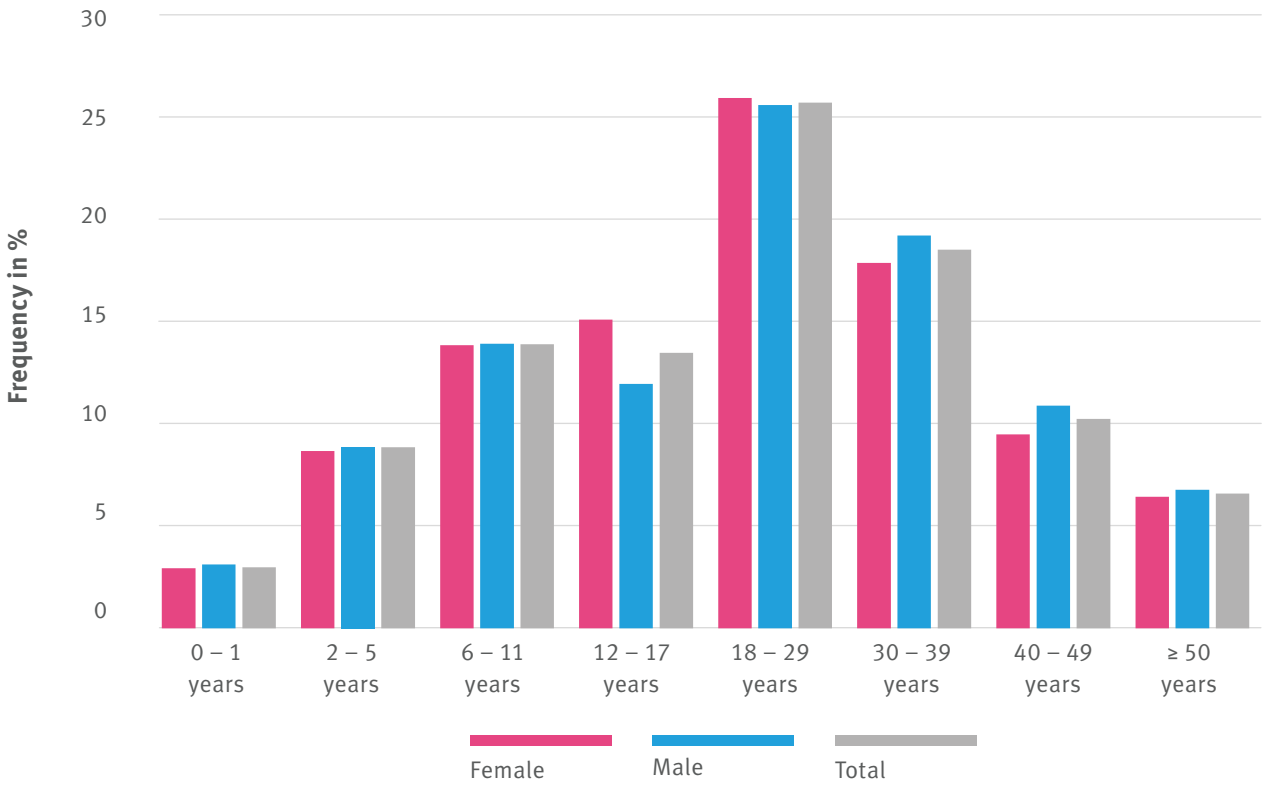

Figure 3: Age distribution of pwCF in 2023

	Male	Female	Total
Numbers	3,457	3,724	7,181
Mean value (years)	23,6	24,3	24,0
Median (years)	22	24	23
Minimum (years)	–	–	–
Maximum(years)	86	85	86
25th percentile (years)	11	11	11
75th percentile (years)	34	35	35
Number < 18 years	1,399	1,403	2,802
Number ≥ 18 years	2,058	2,321	4,379

Table 3: Age distribution of pwCF in 2023

Age structure

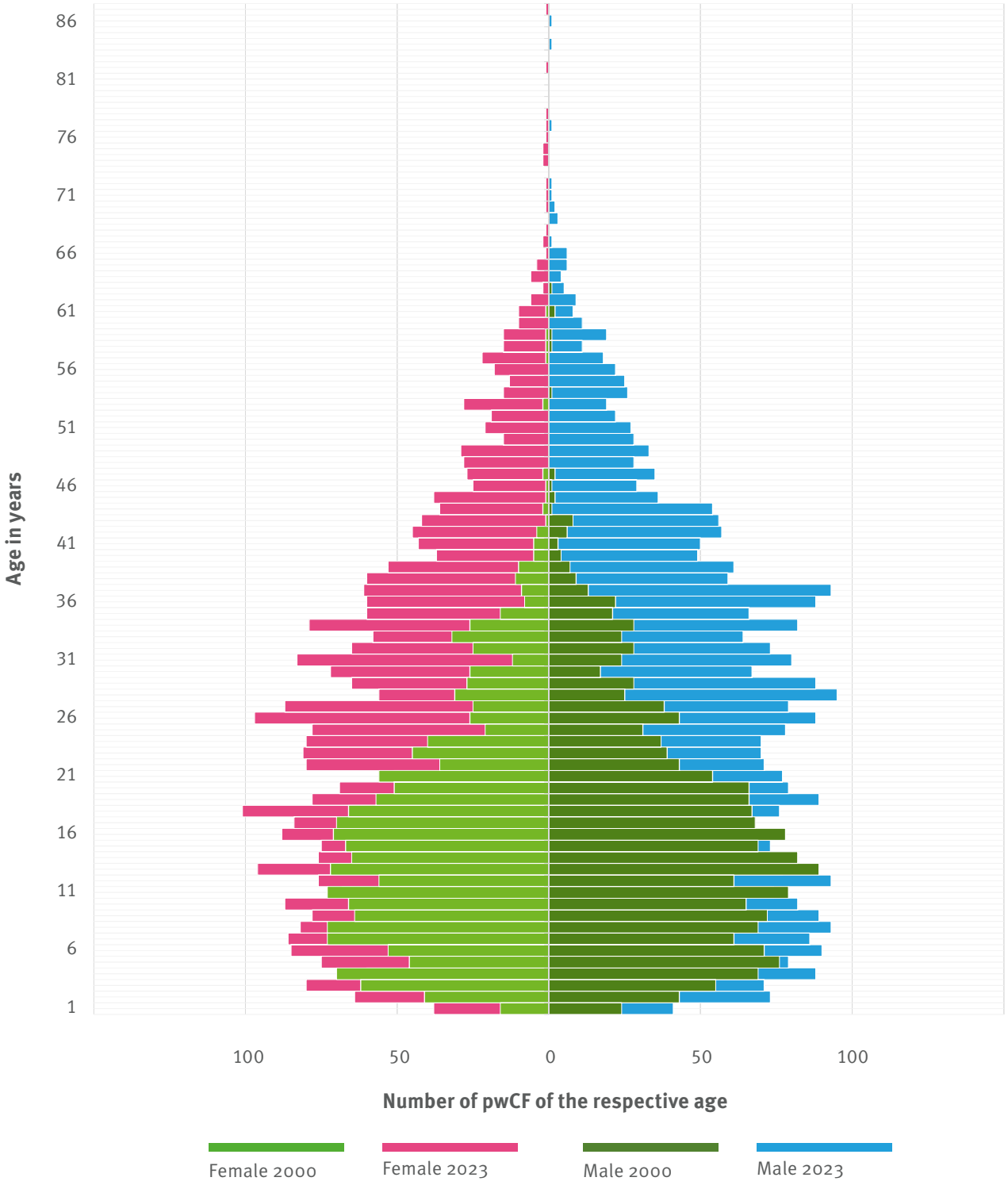

Figure 4: Age pyramid of pwCF in 2000 vs. 2023

03

Age structure

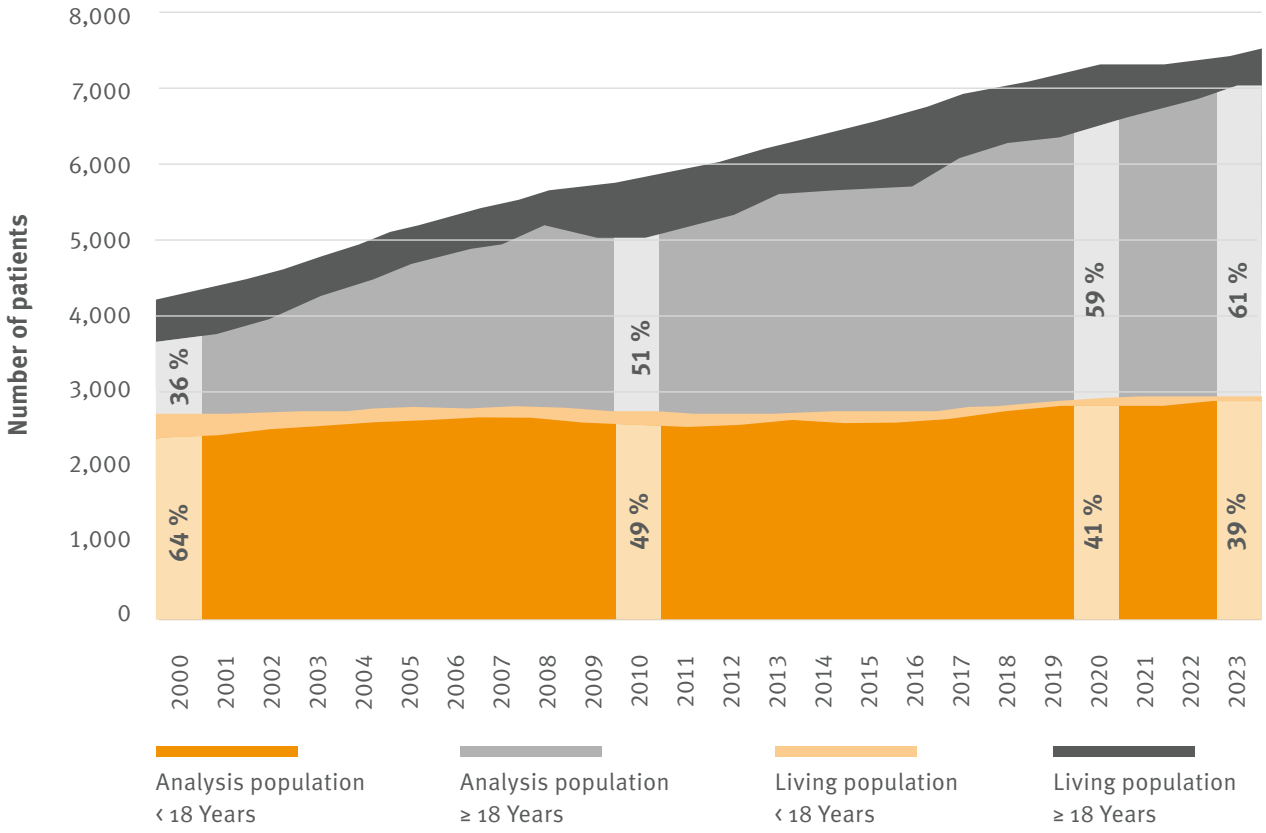

Figure 5: Development of the age distribution (‹ 18 vs. ≥ 18 years) for the years 2000 – 2023

Age structure

Reporting year	Analysis population Total	Analysis population Frequency (%)		Living population Total	Living population frequency (%)	
		‹ 18Years	≥ 18 Years		‹ 18 Years	≥ 18 Years
2000	3,614	63.7	36.3	4,249	61.0	39.0
2001	3,732	62.5	37.5	4,409	59.3	40.7
2002	3,940	61.4	38.6	4,578	57.6	42.4
2003	4,258	57.7	42.3	4,803	55.5	44.5
2004	4,437	56.5	43.5	4,985	53.6	46.4
2005	4,691	54.1	45.9	5,209	52.0	48.0
2006	4,846	52.1	47.9	5,370	49.9	50.1
2007	4,984	51.7	48.3	5,524	49.3	50.7
2008	5,222	49.1	50.9	5,712	47.2	52.8
2009	5,058	48.5	51.5	5,772	45.8	54.2
2010	5,085	48.7	51.3	5,897	45.1	54.9
2011	5,214	46.0	54.0	6,022	43.6	56.4
2012	5,352	45.6	54.4	6,172	42.5	57.5
2013	5,659	44.2	55.8	6,360	41.6	58.4
2014	5,728	43.8	56.2	6,505	40.8	59.2
2015	5,732	43.3	56.7	6,658	39.6	60.4
2016	5,779	43.1	56.9	6,848	38.8	61.2
2017	6,176	41.7	58.3	7,043	38.5	61.5
2018	6,370	41.3	58.7	7,154	38.3	61.7
2019	6,450	41.7	58.3	7,286	38.3	61.7
2020	6,629	41.3	58.7	7,442	38.1	61.9
2021	6,779	40.5	59.5	7,440	38.3	61.7
2022	6,962	39.8	60.2	7,480	38.3	61.7
2023	7,181	39.0	61.0	7,587	37.6	62.4

Table 4: Development of the age distribution (‹ 18 vs. ≥ 18 years) for the years 2000 – 2023

04

Cystic fibrosis diagnosis

4a. Diagnoses in 2023

146 patients were diagnosed in 2023; annual data is available for 130 of these patients (89.0 %). The age distribution of all patients newly diagnosed in 2023 is shown in the following tables.

	N	Mean value	Median	Min	Max	25th percentile	75th percentile	Missing
Age in years	143	5.2	0.1	0	63.1	0.1	3.5	3

Table 5: Age at diagnosis of all pwCF diagnosed in 2023

Newborn screening was performed in 97 (66.4 %) of the pwCF diagnosed in 2023. 12 patients (8.3 %) had a meconium ileus. The age at diagnosis of the patients newly diagnosed via newborn screening in 2023 is as follows:

	N	Mean value	Median	Min	Max	25th percentile	75th percentile	Missing
Age in days	96	77.8	28	0	2.334	18	44.5	1

Table 6: Age at diagnosis of all pwCF diagnosed via newborn screening in 2023

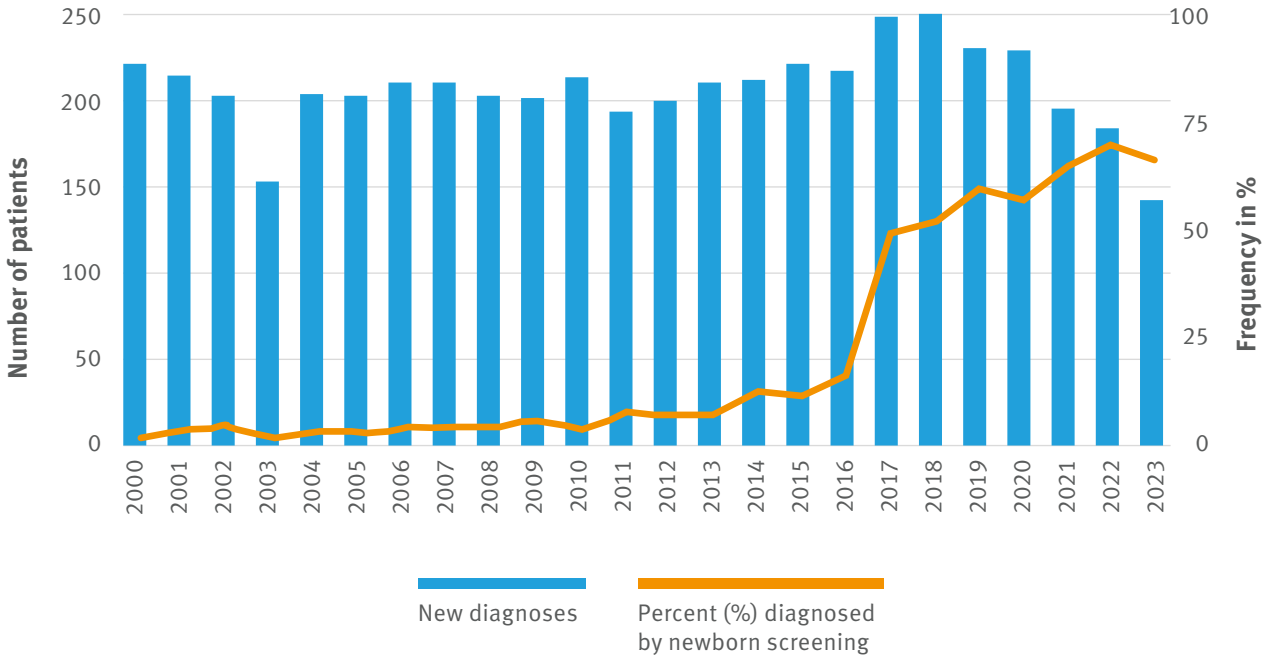

Figure 6: Number of new diagnoses and percentage frequency of pwCF diagnosed through newborn screening 2000 – 2023

Cystic fibrosis diagnosis

4b. Age at diagnosis (Status 2023)

The distribution of the age at diagnosis of the 7,181 patients with follow-up data in 2023 is shown in the following figures and tables below. No information on the date of diagnosis was available for 228 patients (3.1 %).

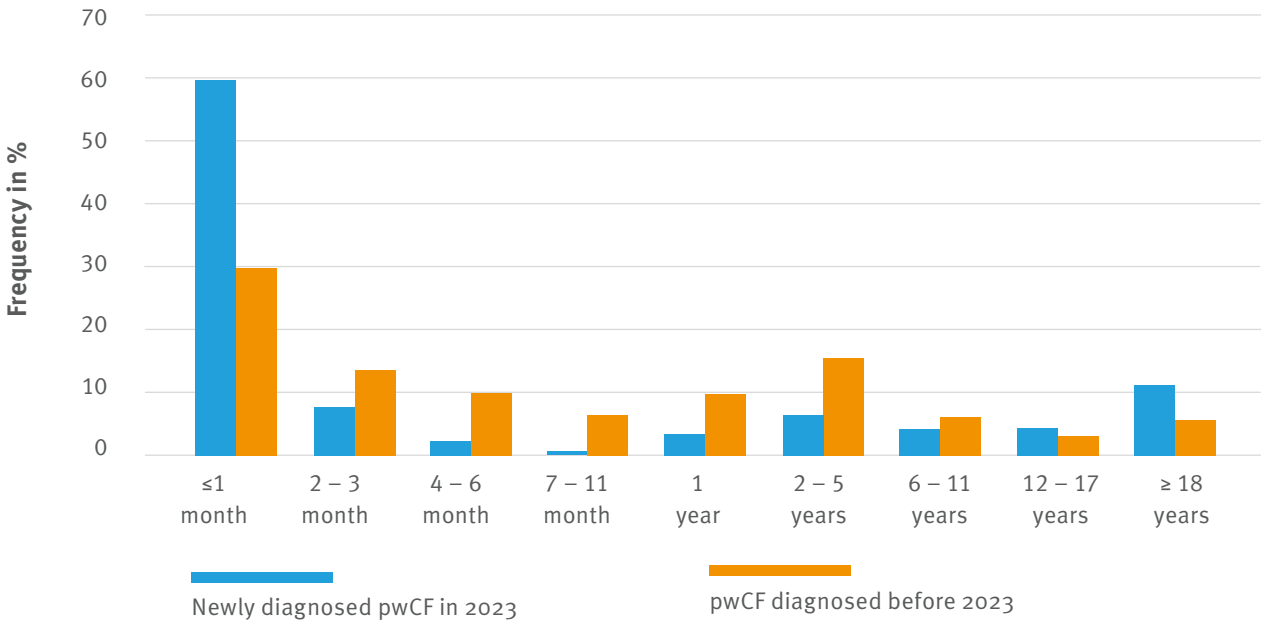

Figure 7: Age-related frequencies in diagnosed pwCF

Age at diagnoses	In 2023			Bevor 2023		
	Frequency	Percent	Accumulated percentages	Frequency	Percent	Accumulated percentages
≤ 1 month	86	60.1	60.1	2,086	30.0	30.0
2 – 3 months	11	7.7	67.8	947	13.6	43.6
4 – 6 months	3	2.1	69.9	668	9.6	53.2
7 – 11 months	1	0.7	70.6	443	6.4	59.6
1 year	5	3.5	74.1	665	9.6	69.2
2 – 5 years	9	6.3	80.4	1,084	15.6	84.8
6 – 11 years	6	4.2	84.6	432	6.2	91.0
12 – 17 years	6	4.2	88.8	227	3.3	94.2
≥ 18 years	16	11.2	100.0	401	5.8	100.0
Total	143	100.0	–	6,953	100.0	–
Missing	3	–	–	228	–	–

Table 7: Age at diagnosis in diagnosed pwCF

04

Cystic fibrosis diagnosis

4c. Genotyping

Genotyping was available for 7,165 patients (99.7 %). Missing information was treated as 'Mutation not identified' in the following presentation.

Mutationskombinationen	Häufigkeit	Prozent
F508del homozygot	3,349	46.6
F508del heterozygous: Second mutation identified	2,815	39.2
F508del heterozygous: Second mutation not identified	49	0.7
No verification of F508del: Both mutations identified	898	12.5
No verification of F508del: Only one mutation identified	24	0.3
No verification of F508del: No mutations identified	46	0.6
Total	7,181	100.0

Table 8: Mutation combinations in pwCF in 2023

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

Erste und zweite Mutation	Anzahl	Prozent
F508del (p.Phe508del / c.1521_1523delCTT)	9,562	66.8
G542X (p.Gly542X / c.1624G>T)	311	2.2
N1303K (p.Asn1303Lys / c.3909C>G)	290	2.0
R553X (p.Arg553X / c.1657C>T)	253	1.8
G551D (p.Gly551Asp / c.1652G>A)	222	1.6
CFTRdele2,3 (p.Ser18ArgfsX16 / c.54-5940_273+10250del21kb)	219	1.5
R347P (p.Arg347Pro / c.1040G>C)	193	1.4
3849+10kbC->T (c.3718-2477C>T)	165	1.2
1717-1G->A (c.1585-1G>A)	120	0.8
2789+5G->A (c.2657+5G>A)	111	0.8
2183AA->G (p.Lys684SerfsX38 / c.2051_2052delAAinsG)	92	0.6
W1282X (p.Trp1282X / c.3846G>A)	91	0.6
2184insA (p.Gln685ThrfsX4 or p.Gln685Thrfs*4 / c.2052dupA or c.2052dup)	75	0.5
3272-26A->G (c.3140-26A>G)	73	0.5
R117H (p.Arg117His / c.350G>A)	68	0.5
I336K (p.Ile336Lys / c.1007T>A)	63	0.4
M1101K (p.Met1101Lys / c.3302T>A)	61	0.4
R1162X (p.Arg1162X / c.3484C>T)	61	0.4
1677delTA (p.Tyr515X / c.1545_1546delTA)	56	0.4
2143delT (p.Leu671X / c.2012delT)	53	0.4
621+1G->T (c.489+1G>T)	53	0.4
Other Mutation	2,005	14.0
Unknown/Mutation not identified	123	0.9
Total	14,320	100

Table 9: CFTR genotyping of pwCF in 2023

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05

Nutritional status

5a. Children and adolescents under the age of 18

All patients aged 2 – 17 years without transplantation with follow-up data 2023 (n=2,607) were included.
No information on nutritional status was available for 3 patients (0.1 %). For the assessment of the nutritional status in children and adolescents, the BMI percentiles according to KiGGS were used. The age was calculated at the time of the physical examination.

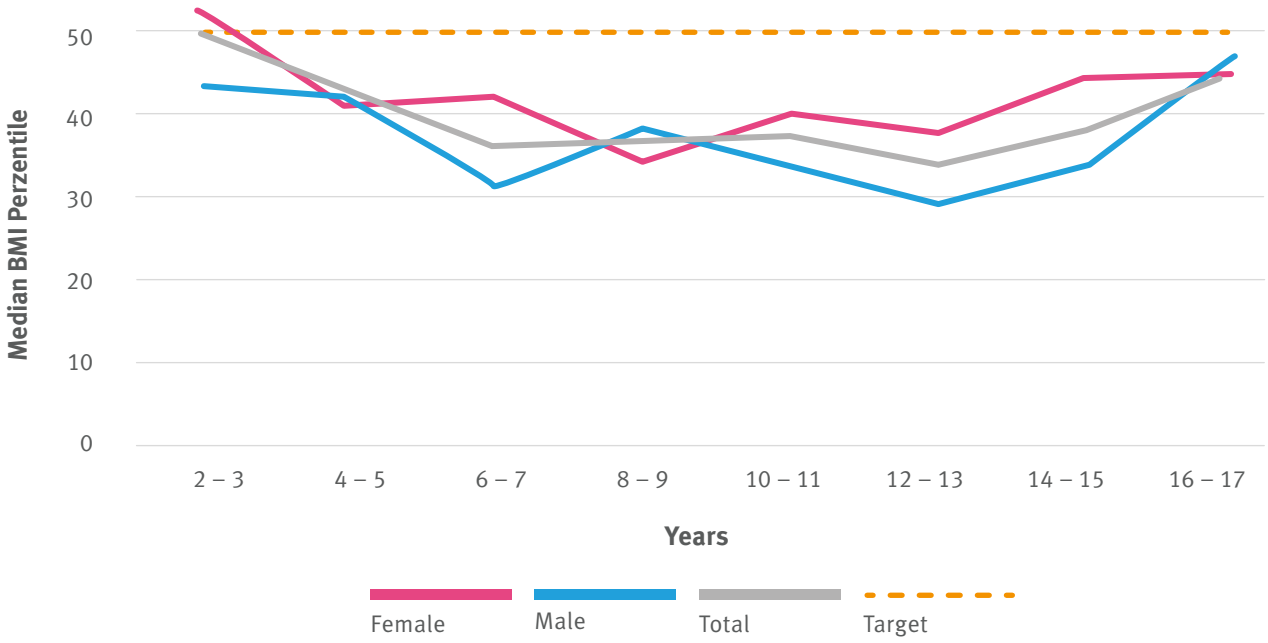

Figure 8: Median BMI percentiles of children and adolescents aged between 2 and 17 in 2023

Age (years)	Male			Female			Total		
	N	Median	25th - 75th pctl.	N	Median	25th - 75th pctl.	N	Median	25th - 75th pctl.
2 – 3	156	43.5	20.0 – 70.5	134	53.0	20.0 – 74.0	290	49.5	20.0 – 72.0
4 – 5	180	42.0	20.0 – 64.0	168	41.0	15.5 – 68.0	348	42.0	17.5 – 65.5
6 – 7	190	31.0	13.0 – 59.0	175	42.0	21.0 – 64.0	365	36.0	16.0 – 62.0
8 – 9	157	38.0	21.0 – 58.0	167	34.0	15.0 – 61.0	324	37.0	18.0 – 59.0
10 – 11	180	33.5	17.5 – 58.0	153	40.0	20.0 – 53.0	333	37.0	18.0 – 56.0
12 – 13	139	29.0	11.0 – 49.0	158	37.5	20.0 – 61.0	297	33.0	15.0 – 58.0
14 – 15	145	34.0	19.0 – 54.0	158	44.5	19.0 – 67.0	303	38.0	19.0 – 61.0
16 – 17	156	47.0	16.0 – 70.0	188	45.0	18.0 – 66.0	344	45.0	17.5 – 68.0
Total	1.303	37.0	17.0 – 61.0	1.301	42.0	18.0 – 65.0	2.604	39.0	18.0 – 63.0

Table 10: BMI percentiles of children and adolescents aged 2 – 17 in 2023

Nutritional status

5a. Children and adolescents under 18 years of age

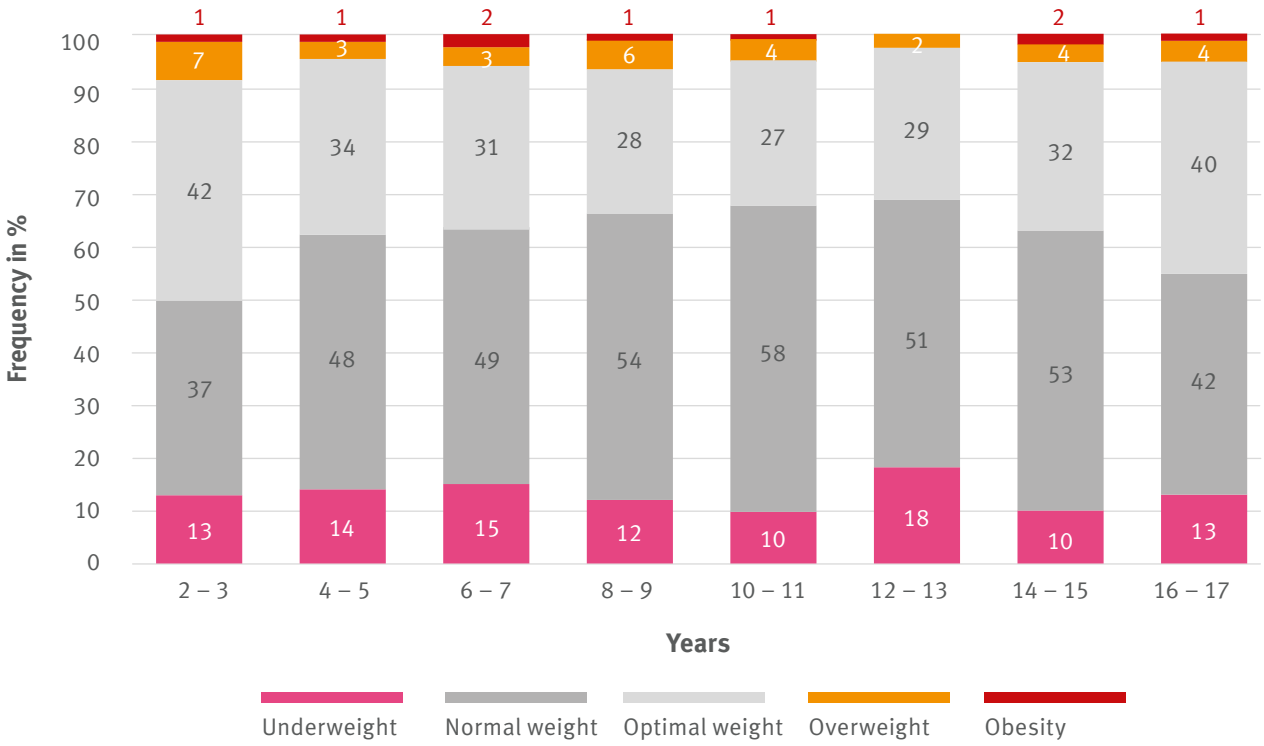

Figure 9: Weight categories of children and adolescents between 2 – 17 years 2023, underweight: BMI percentiles < 10, normal weight: BMI percentiles 10 – 49, optimal weight: BMI percentiles 50 – 89, overweight: BMI percentiles 90 – 96, obesity: BMI percentiles ≥ 97

BMI Perzentile KIGGS	Male	Female	Total
Underweight	13.8	12.3	13.1
Normal weight	51.0	46.7	48.9
Optimal weight	29.3	36.4	32.8
Overweight	4.2	3.9	4.1
Obesity	1.6	0.7	1.2

Table 11: Weight categories of children and adolescents between 2 – 17 years 2023, 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

weight-for-length	Male		Female		Total	
	0 – 12 months	13 – 24 months	0 – 12 months	13 – 24 months	0 – 12 months	13 – 24 months
Underweight	27.8	6.0	38.8	12.5	33.0	9.2
Normal weight	68.5	92.0	55.1	72.9	62.1	82.7
Overweight/Obesity	3.7	2.0	6.1	14.6	4.9	8.2

Table 12: Weight categories of children under 2 years of age (frequencies in %) according to weight-for-length (LSG) 2023
Underweight: LSG <90 %; normal weight: LSG 90 – 110 %; overweight/obesity: LSG >110 %

05

Nutritional status

5b. Development of nutritional status 2000 – 2023 Children and adolescents under 18 years of age

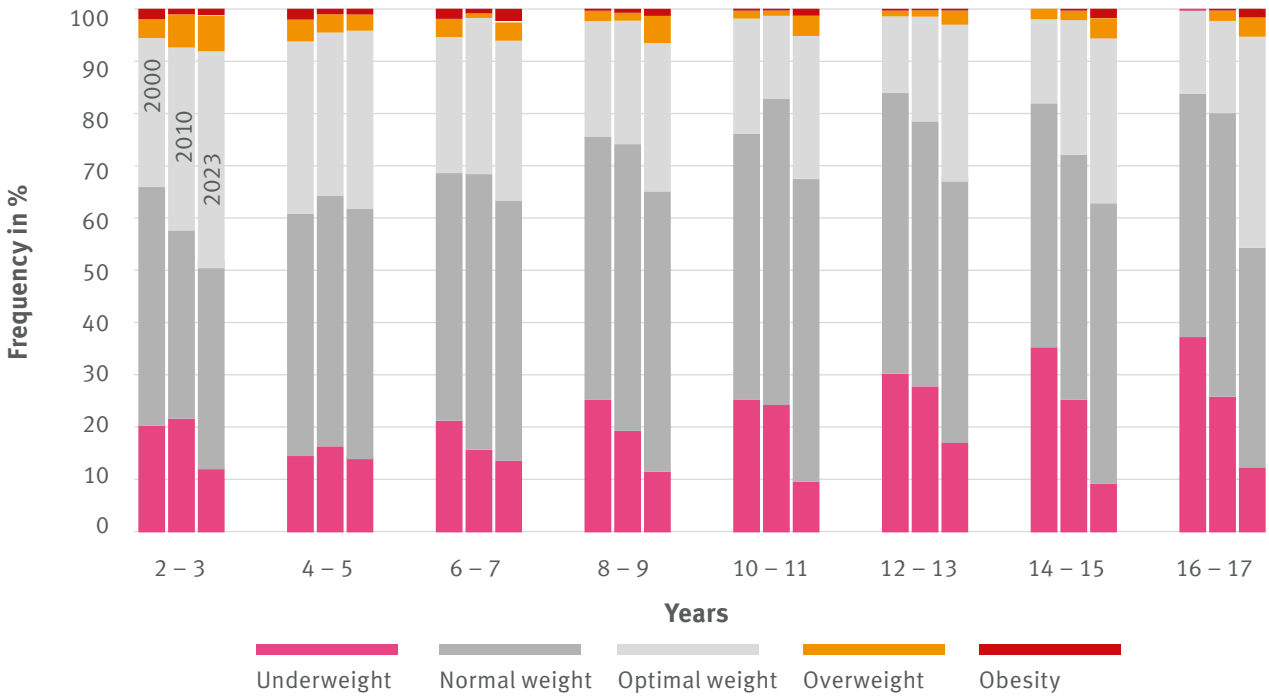

Figure 10: Development of the weight categories of children and adolescents up to 17 years 2000 – 2023, underweight: BMI percentiles < 10; Normal weight: BMI percentiles 10 – 49; optimal weight: BMI percentiles 50 – 89; overweight: BMI percentiles 90 – 96, obesity: BMI percentiles ≥ 97

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
	2023	12.8	14.4	14.5	12.0	10.2	17.5	10.2	12.8
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
	2023	37.2	47.7	49.0	53.7	57.7	50.5	52.8	41.9
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
	2023	42.1	34.2	31.0	27.8	27.0	29.3	31.7	40.1
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
	2023	6.9	3.2	3.0	5.9	3.9	2.4	3.6	4.1
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
	2023	1.0	0.6	2.5	0.6	1.2	0.3	1.7	1.2

Table 13: Development of the weight categories of children and adolescents up to the age of 17 (frequencies in %) 2000 – 2023, 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

5b.i Median BMI percentile by birth cohort

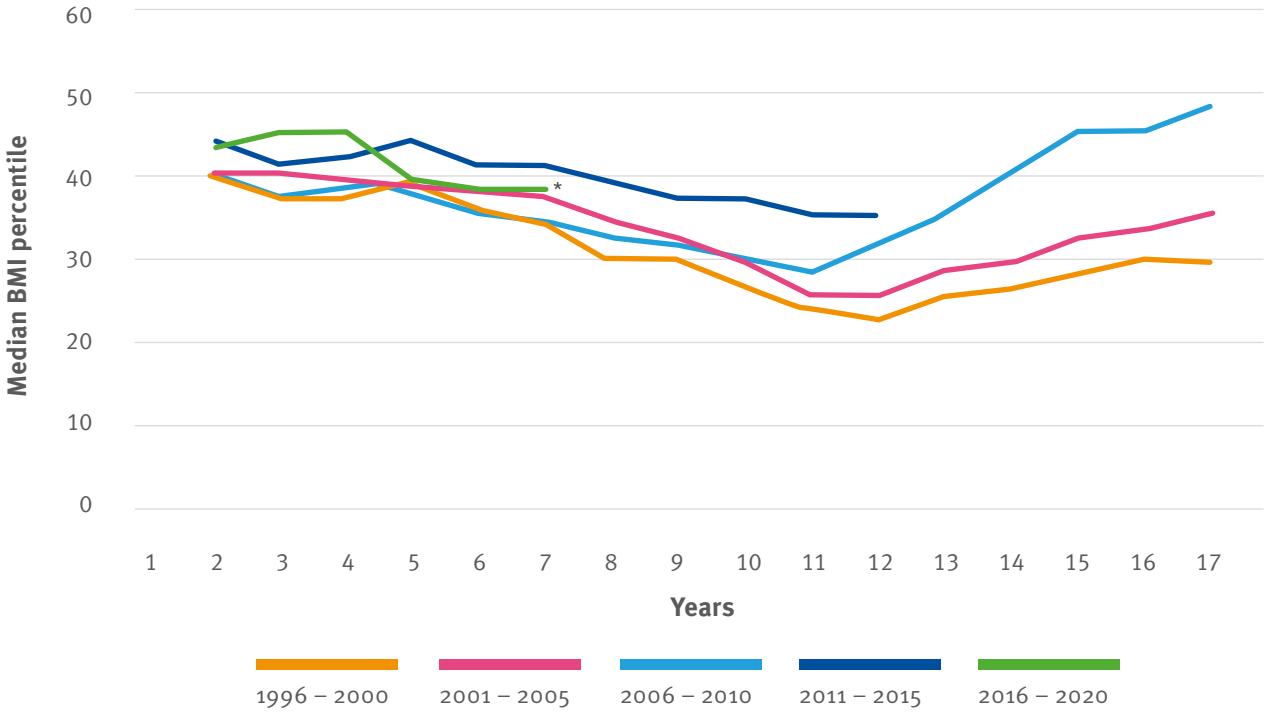

Figure 11: Development of median BMI percentiles of children and adolescents under 18 years of age by birth cohort 1996 – 2020 for the years 1996 – 2023. Until 2014, the BMI close to the date of birth was recorded, from 2014 onwards the BMI at the time of recording the best lung function of the calendar year.
* Limited significance due to the small cohort size.

Birth Cohort	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	29	25	25	28	29	32	33	35
2006 – 2010	40	37	38	39	35	34	32	31	29	28	31	35	40	45	45	48
2011 – 2015	44	41	42	44	41	41	39	37	37	35	35	–	–	–	–	–
2016 – 2020	43	45	45	39	38	38*	–	–	–	–	–	–	–	–	–	–

Table 14: Development of median BMI percentiles of children and adolescents under 18 years of age by birth cohort 1996 – 2020 for the years 1996 – 2023. 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 function of the calendar year. * Limited significance due to the small cohort size.

05

Nutritional status

5c. Adults

Adult patients without transplantation with follow-up data 2023 (n=3,952) were considered. For 22 patients (0.6 %) no information was available for the nutritional status. Age was calculated at the time of the physical examination.

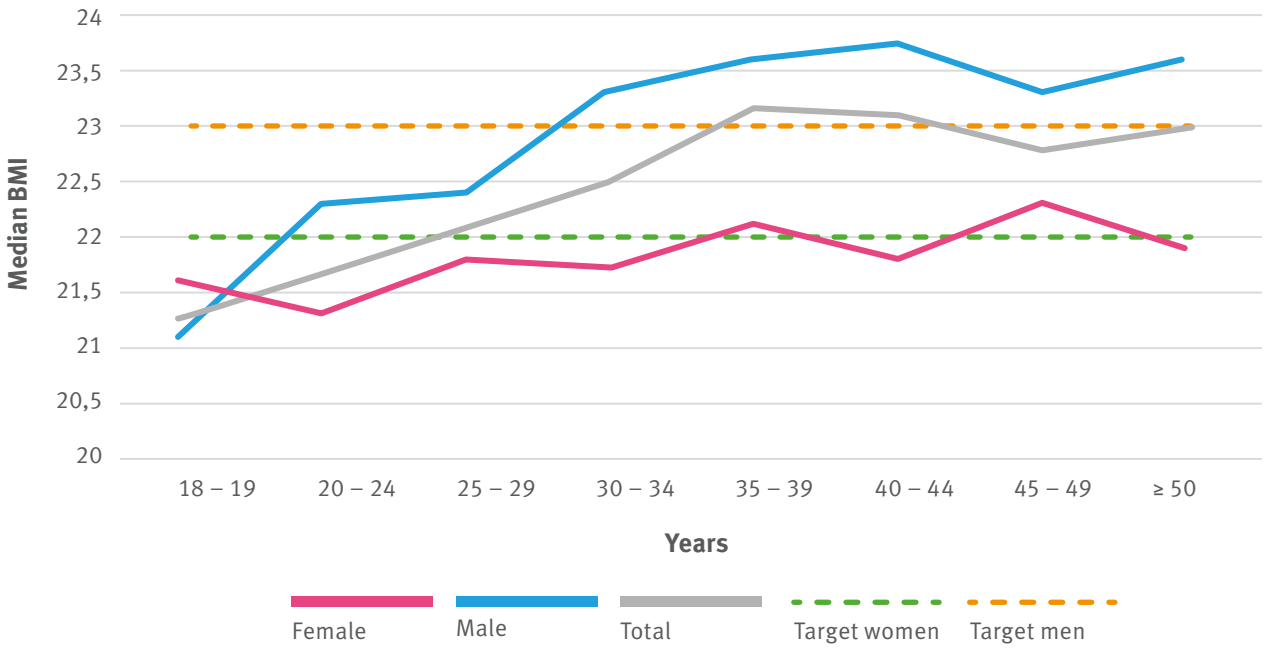

Figure 12: Median BMI of adults aged 18 and over in 2023

Age (years)	Male			Female			Total		
	N	Median	25th - 75th pctl.	N	Median	25th - 75th pctl.	N	Median	25th - 75th pctl.
18 – 19	149	21.1	19.6 – 23.3	131	21.6	19.9 – 23.4	280	21.3	19.8 – 23.4
20 – 24	359	22.3	20.0 – 24.6	364	21.3	19.6 – 23.2	723	21.7	19.7 – 23.9
25 – 29	387	22.4	20.4 – 25.2	360	21.8	19.9 – 23.7	747	22.1	20.2 – 24.5
30 – 34	352	23.3	21.4 – 25.6	289	21.7	20.0 – 24.1	641	22.5	20.6 – 25.0
35 – 39	291	23.6	21.8 – 25.6	247	22.1	20.6 – 24.8	538	23.2	21.2 – 25.4
40 – 44	220	23.8	21.8 – 26.1	155	21.8	20.2 – 24.2	375	23.1	20.8 – 25.3
45 – 49	132	23.3	21.3 – 25.6	97	22.3	20.0 – 24.2	229	22.8	20.6 – 25.1
≥ 50	211	23.6	21.9 – 25.9	190	21.9	20.6 – 24.6	401	23.0	21.1 – 25.6
Total	2.101	23.0	21.0 – 25.3	1.833	21.7	20.0 – 23.9	3.934	22.3	20.4 – 24.7

Table 15: BMI of adults aged 18 and over in 2023

Nutritional status

5c. Adults

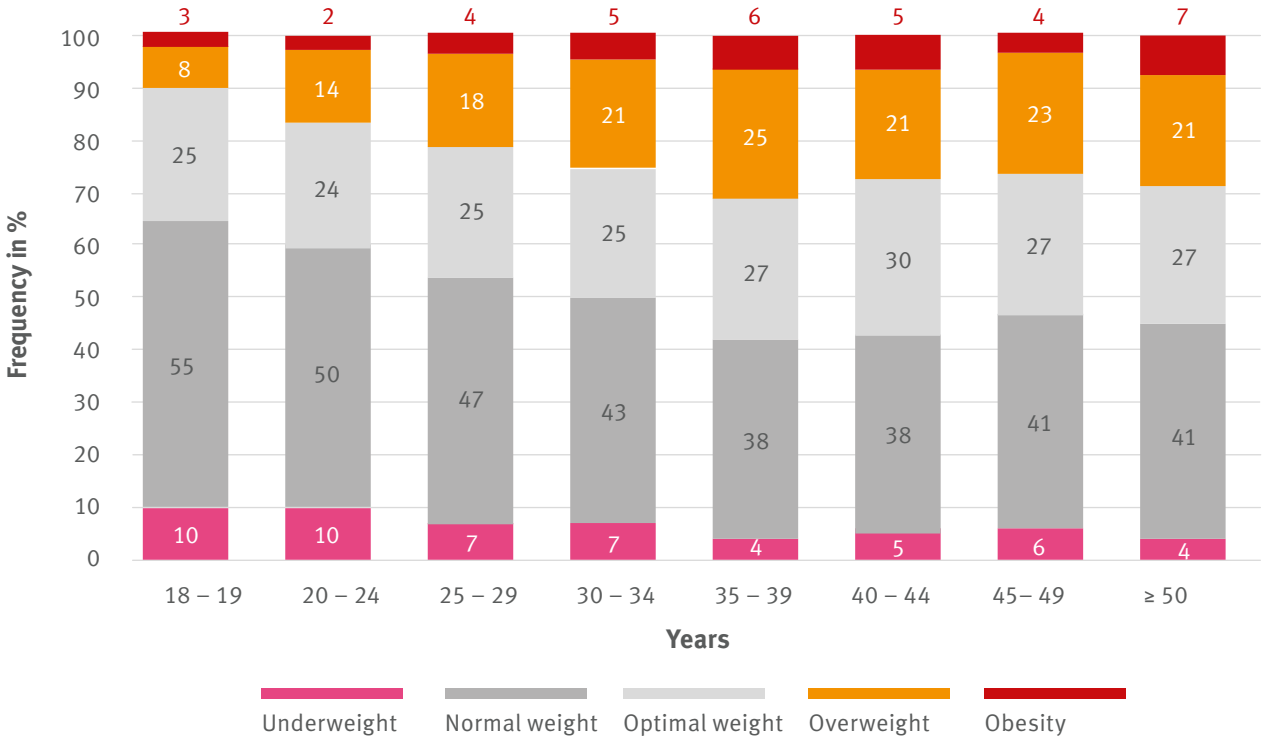

Figure 13: Weight categories for adults aged 18 and over in 2023,
Underweight: BMI ‹ 18.5 kg/sqm; normal weight: BMI men 18.5 – 22.9 kg/sqm; BMI women 18.5 – 21.9 kg/sqm;
Optimal weight: BMI men 23.0 – 24.9 kg/qm; BMI women 22.0 – 24.9 kg/qm; overweight: BMI 25.0 – 29.9 kg/qm; obesity: BMI ≥ 30 kg/qm

	Male	Female	Total
Underweight	5.1	8.7	6.8
Normal weight	44.0	44.5	44.3
Optimal weight	23.0	29.1	25.9
Overweight	23.2	13.9	18.8
Obesity	4.8	3.8	4.3

Table 16: Weight categories of adults aged 18 and over (frequencies in %) 2023,
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; obesity: BMI ≥ 30 kg/qm

05

Nutritional status

5d. Development of nutritional status 2000 – 2023 Adults

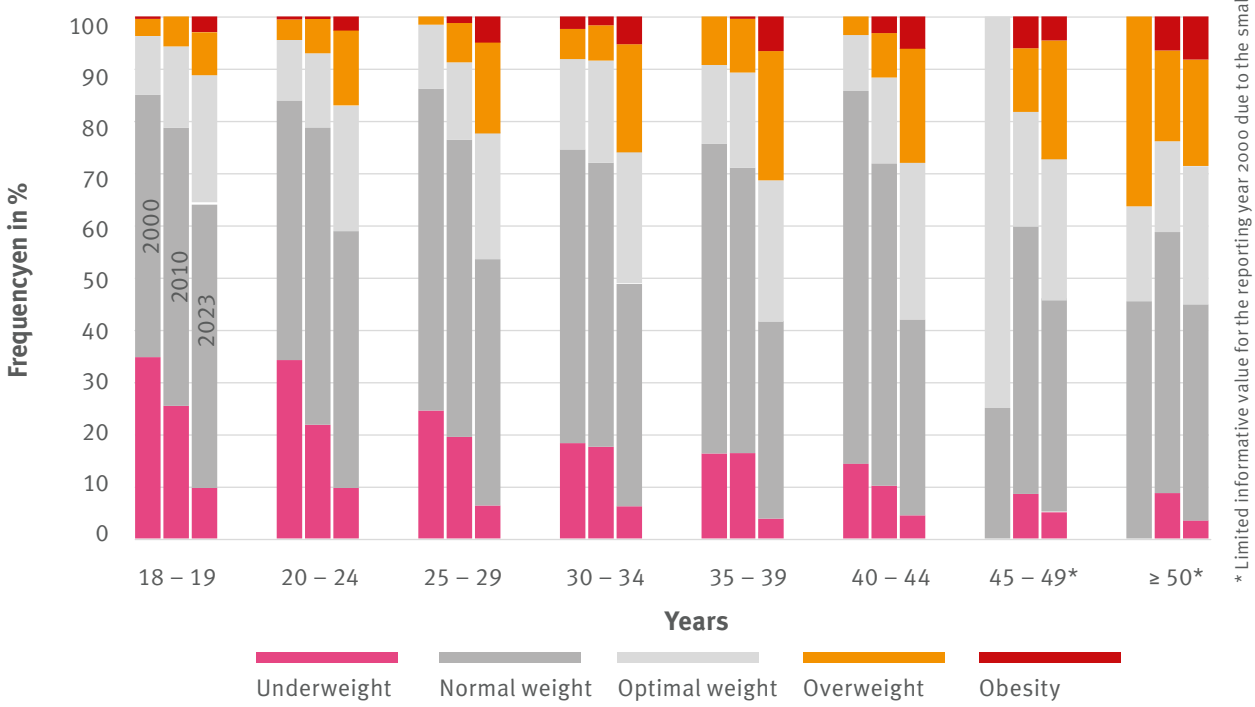

Figure 14: Development of the weight categories of adults aged 18 and over 2000 – 2023,
Underweight: BMI < 18.5 kg/sqm; normal weight: BMI men 18.5 – 22.9 kg/sqm; BMI women 18.5 – 21.9 kg/sqm;
Optimal weight: BMI men 23.0 – 24.9 kg/qm, BMI women 22.0 – 24.9 kg/qm; overweight: BMI 25.0 – 29.9 kg/qm; obesity: 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
Underweight	2000	34.7	34.2	24.5	18.3	16.5	14.3	0.0	0.0
	2010	25.4	21.8	19.5	17.5	16.4	10.1	8.5	8.7
	2023	10.0	10.0	7.0	6.9	4.3	4.8	5.7	4.0
Normal weight	2000	50.2	49.7	61.7	56.3	58.8	71.4	25.0	45.5
	2010	53.3	56.8	56.9	54.6	54.9	61.4	51.2	50.0
	2023	54.6	49.5	47.0	42.8	37.9	37.9	40.6	41.4
Optimal weight	2000	11.3	11.6	12.3	17.3	15.3	10.7	75.0	18.2
	2010	15.6	14.2	14.9	19.5	17.8	16.9	22.0	17.4
	2023	24.6	24.3	24.5	25.0	27.1	30.4	27.1	26.7
Overweight	2000	3.3	4.0	1.6	5.8	9.4	3.6	0.0	36.4
	2010	5.7	6.6	7.5	6.7	10.3	8.5	12.2	17.4
	2023	8.2	14.4	17.8	20.9	24.5	21.1	22.7	21.0
Obesity	2000	0.5	0.6	0.0	2.4	0.0	0.0	0.0	0.0
	2010	0.0	0.6	1.3	1.7	0.5	3.2	6.1	6.5
	2023	2.5	1.8	3.8	4.5	6.1	5.9	3.9	7.0

Table 17: Development of the weight categories of adults aged 18 and over (frequencies in %) 2000 – 2023
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/sqm, BMI women 22.0 – 24.9 kg/qm; overweight: BMI 25.0 – 29.9 kg/qm; obesity: BMI ≥ 30 kg/qm

Nutritional status

5d.i Median BMI by birth cohort

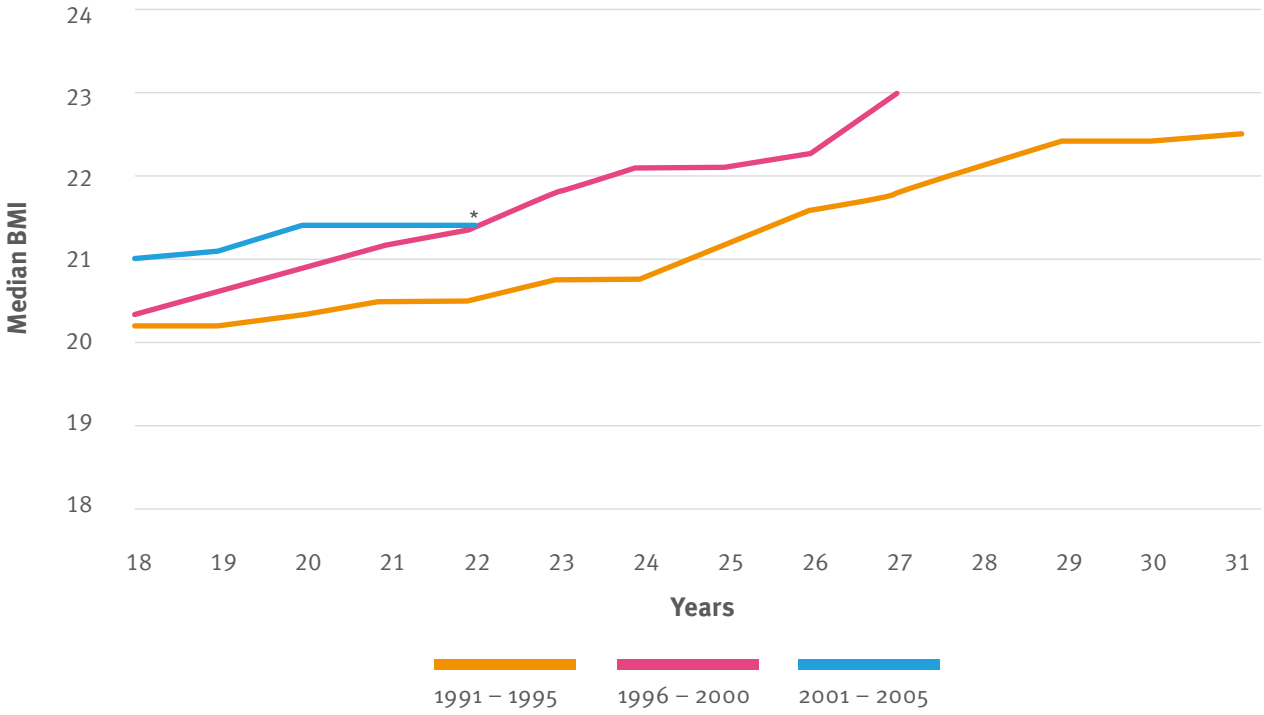

Figure 15: Development of median BMI of adults aged 18 and over by birth cohort 1991 – 2005 for the years 2009 – 2023.
Until 2014, the BMI close to the date of birth was recorded, from 2014 the BMI at the time of recording the best FEV1% of the calendar year.
* Limited significance due to the small cohort size

Birth Cohort	Age in years													
	18	19	20	21	22	23	24	25	26	27	28	29	30	31
1991 – 1995	20	20	20	21	21	21	21	21	22	22	22	22	22	23
1996 – 2000	20	21	21	21	21	22	22	22	22	23	–	–	–	–
2001 – 2005	21	21	21	21	21*	–	–	–	–	–	–	–	–	–

Table 18: Development of median BMI of adults aged 18 and over by birth cohort 1991 – 2005 for the years 2009 – 2023.
Until 2014, the BMI close to the date of birth was recorded, from 2014 the BMI at the time of recording the best FEV1% of the calendar year.
* Limited significance due to the small cohort size

06

Lung function

6a. Overview

For the evaluation of lung function, all patients aged 6 years and older without transplantation, with lung function measurement 2023 were taken into account. A total of 5,793 data records were available.

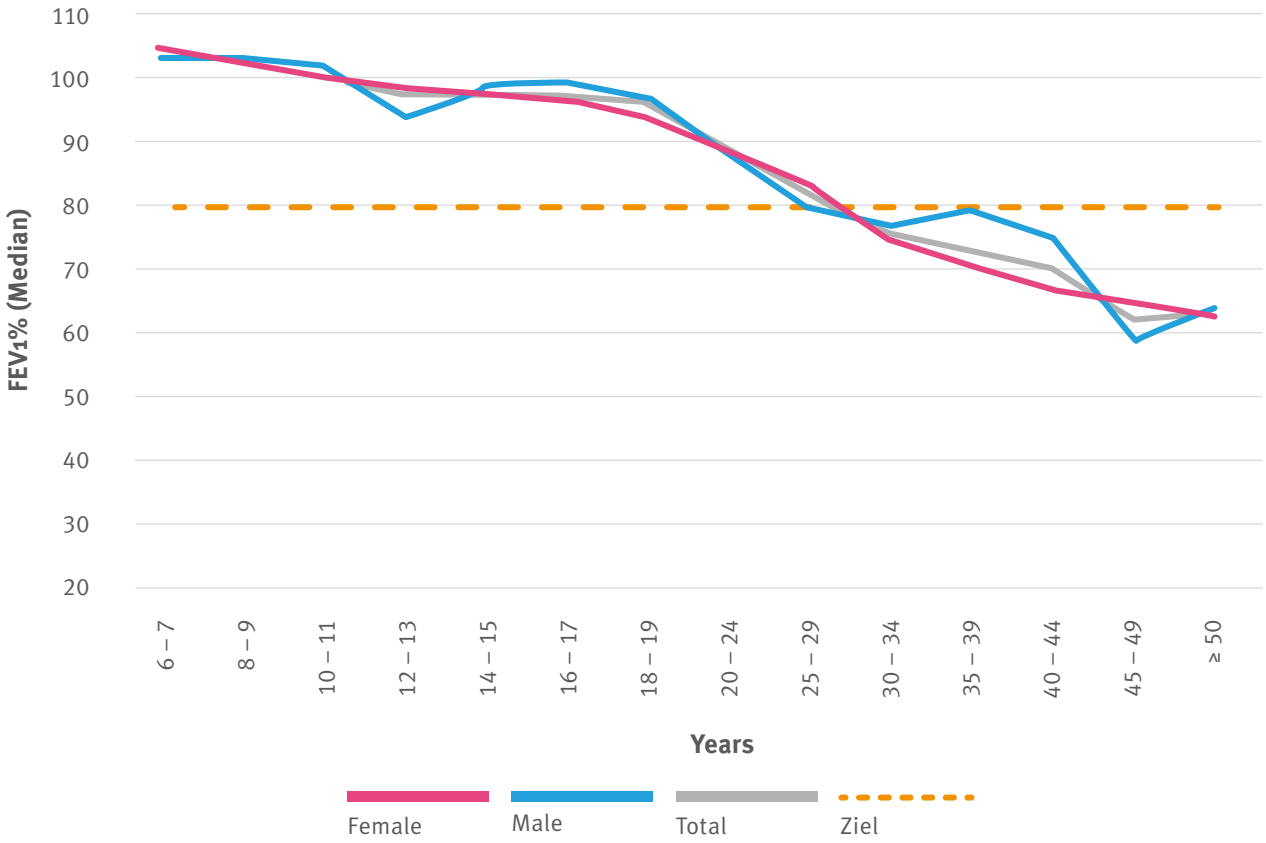

Figure 16: FEV1% value 2023 according to the Global Lung Function Initiative (GLI)

Lung function

Age (years)	Male			Female			Total		
	Anzahl	Median	25. – 75 P.	Anzahl	Median	25. – 75 P.	Anzahl	Median	25. – 75 P.
6 – 7	183	103.0	93 – 113	166	105.0	96 – 113	349	104.0	95 – 113
8 – 9	156	103.0	94 – 112	165	103.0	95 – 112	321	103.0	94 – 112
10 – 11	179	101.0	92 – 108	153	100.0	91 – 107	332	100.0	92 – 107
12 – 13	135	94.0	88 – 104	153	98.0	86 – 110	288	97.0	87 – 106
14 – 15	142	99.0	88 – 108	154	97.0	87 – 105	296	98.0	88 – 107
16 – 17	152	99.0	89 – 109	179	96.0	85 – 106	331	97.0	87 – 107
18 – 19	143	96.0	85 – 105	121	94.0	86 – 104	264	96.0	85 – 105
20 – 24	354	88.0	75 – 100	358	88.0	69 – 101	712	88.0	71 – 101
25 – 29	383	79.0	58 – 98	358	83.0	65 – 98	741	81.0	60 – 98
30 – 34	350	76.0	58 – 92	285	74.0	58 – 93	635	75.0	58 – 93
35 – 39	290	79.0	56 – 93	245	70.0	52 – 86	535	73.0	55 – 90
40 – 44	215	74.0	50 – 91	155	66.0	52 – 87	370	70.0	51 – 89
45 – 49	130	59.0	42 – 80	96	64.0	50 – 79	226	61.0	45 – 80
≥ 50	208	63.0	43 – 85	185	62.0	48 – 77	393	63.0	46 – 82
< 18	947	100.0	91 – 109	970	100.0	91 – 109	1,917	100,0	91 – 109
≥ 18	2,073	80,0	57 – 95	1,803	77,0	58 – 95	3,876	78,0	57 – 95
Total	3,020	88,0	67 – 102	2,773	88,0	67 – 102	5,793	88,0	67 – 102

Table 19: FEV1% value 2023 according to the Global Lung Function Initiative (GLI)

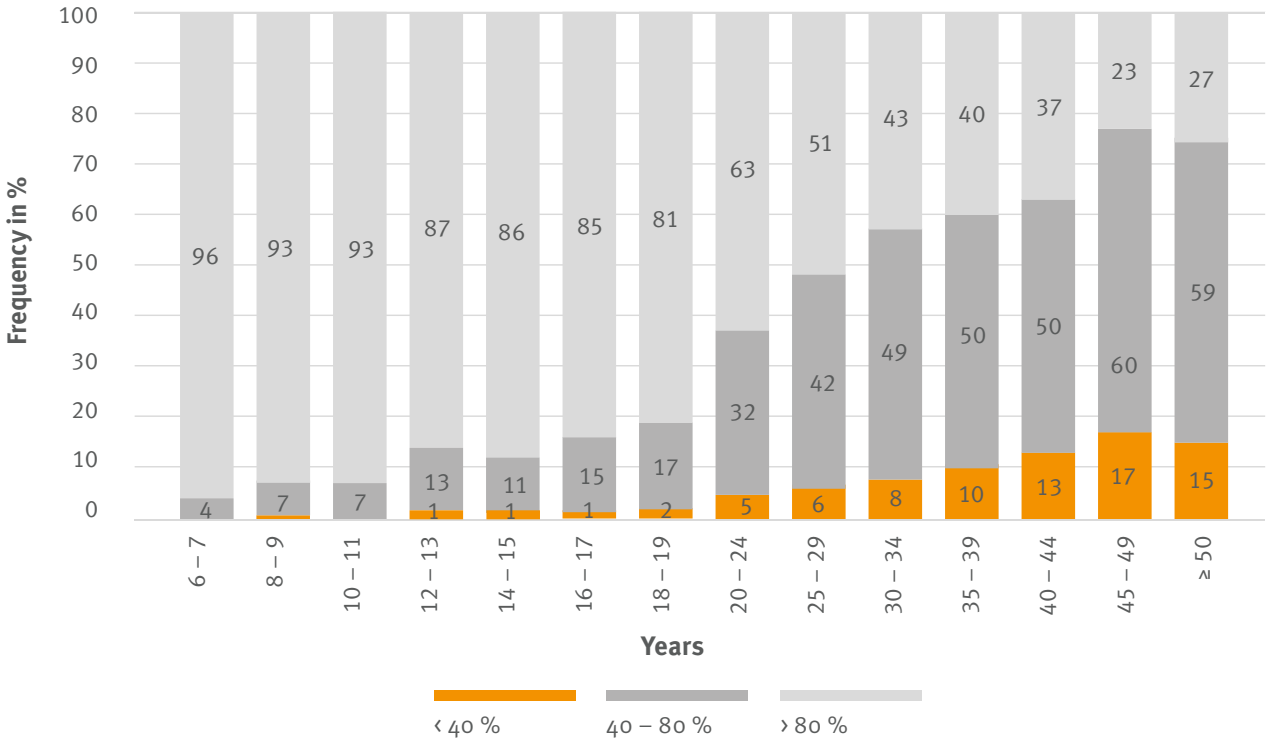

Figure 17: Severity of FEV1% (categories < 40 %, 40 – 80 %, > 80 %) in 2023 according to the Global Lung Function Initiative (GLI)

06

Lung function

6b. Development of lung function 2000 – 2023

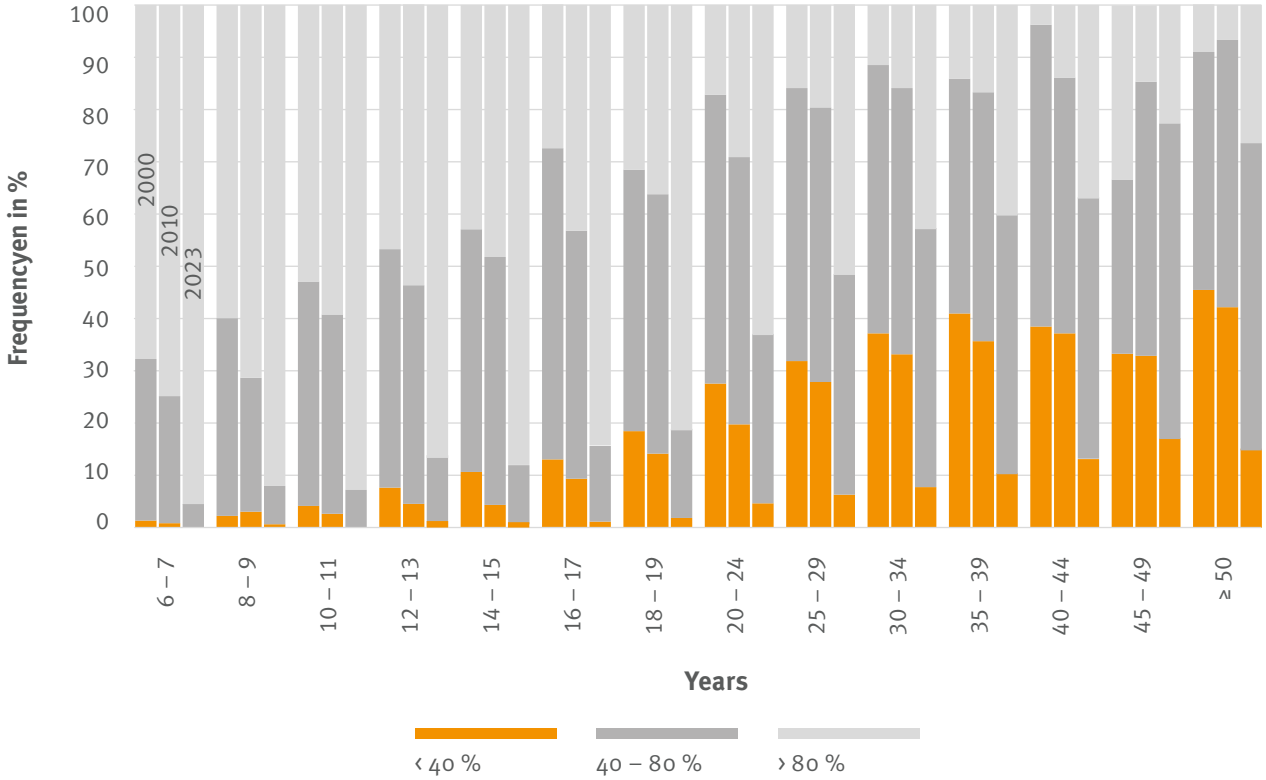

Figure 18: Development of age-related frequencies (in %) of the severity of FEV1% according to the Global Lung Function Initiative (GLI) 2000 – 2023

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	40.3	38.5	33.3	45.5
	2010	0.9	3.1	2.7	4.6	4.4	9.4	14.2	19.7	27.9	33.2	35.9	37.0	32.9	42.2
	2023	0.3	0.3	0.0	0.7	0.7	0.9	1.5	4.8	6.3	7.9	10.1	13.0	16.8	14.8
40 – 80 %	2000	30.9	37.8	42.9	45.6	46.4	59.5	50.0	55.2	52.2	51.3	45.5	57.7	33.3	45.5
	2010	24.3	25.6	38.1	41.8	47.4	47.4	49.6	51.2	52.5	50.9	47.9	48.6	52.4	51.1
	2023	4.0	7.2	6.9	12.5	11.2	14.5	17.1	32.2	42.2	49.3	49.7	50.0	60.2	58.5
› 80 %	2000	67.7	59.9	52.9	46.8	42.9	27.5	31.5	17.2	15.9	11.5	14.3	3.9	33.3	9.1
	2010	74.8	71.3	59.2	53.7	48.2	43.2	36.2	29.1	19.6	15.9	16.3	14.4	14.6	6.7
	2023	95.7	92.5	93.1	86.8	88.2	84.6	81.4	63.1	51.4	42.8	40.2	37.0	23.0	26.7

Table 20: Development of age-related frequencies (in %) of the severity of FEV1% according to the Global Lung Function Initiative (GLI) 2000 – 2023

Lung function

6c. Median FEV1% by birth cohort

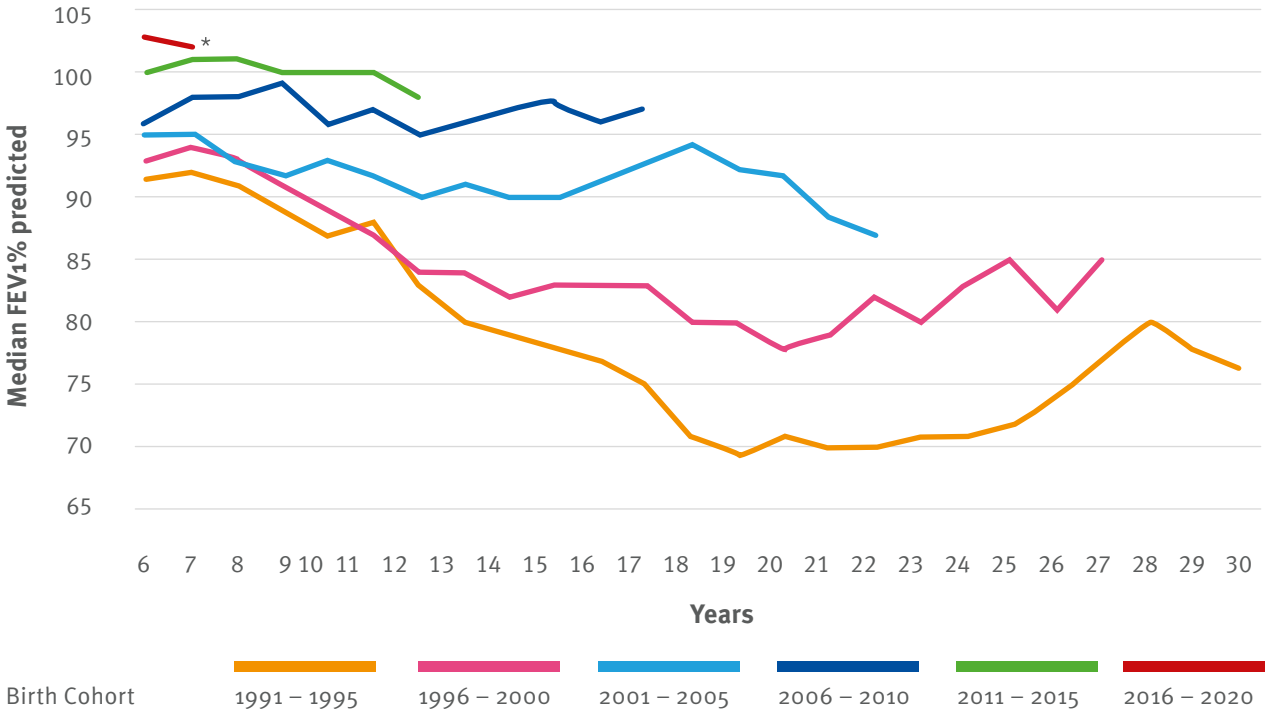

Figure 19: Development of median FEV1% of children and adults by birth cohort 1991 – 2020 for the years 1997 – 2023.
Until 2014, the FEV1% close to the date of birth was recorded, from 2014 the best FEV1% of the calendar year.
* Limited significance due to the small cohort size

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	30	31
1991 – 1995	92	92	91	89	87	88	83	80	79	78	77	75	71	70	71	70	70	71	71	72	74	77	80	78	77	76
1996 – 2000	93	94	93	91	89	87	84	84	82	83	83	83	80	80	78	79	82	80	83	85	81	85	–	–	–	–
2001 – 2005	95	95	93	92	93	92	90	91	90	90	91	93	94	93	92	89	87	–	–	–	–	–	–	–	–	–
2006 – 2010	96	98	98	99	96	97	95	96	97	98	96	97	–	–	–	–	–	–	–	–	–	–	–	–	–	–
2011 – 2015	100	101	101	100	100	100	98	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–
2016 – 2020	103	102*	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–	–

Table 21: Development of median FEV1% of children and adults by birth cohort 1991 – 2020 for the years 1997 – 2023.
Until 2014, the FEV1% close to the date of birth was recorded, from 2014 the best FEV1% of the calendar year.
* Limited significance due to the small cohort size

07

Lung infection

7a. Annual verification at leasrt once

All pwCF without transplantation who had at least one microbiological examination in the calendar year were included in the calendar year (n=6,719). No information on the microbiological test in the calendar year was available for 118 patients (1.7 %).

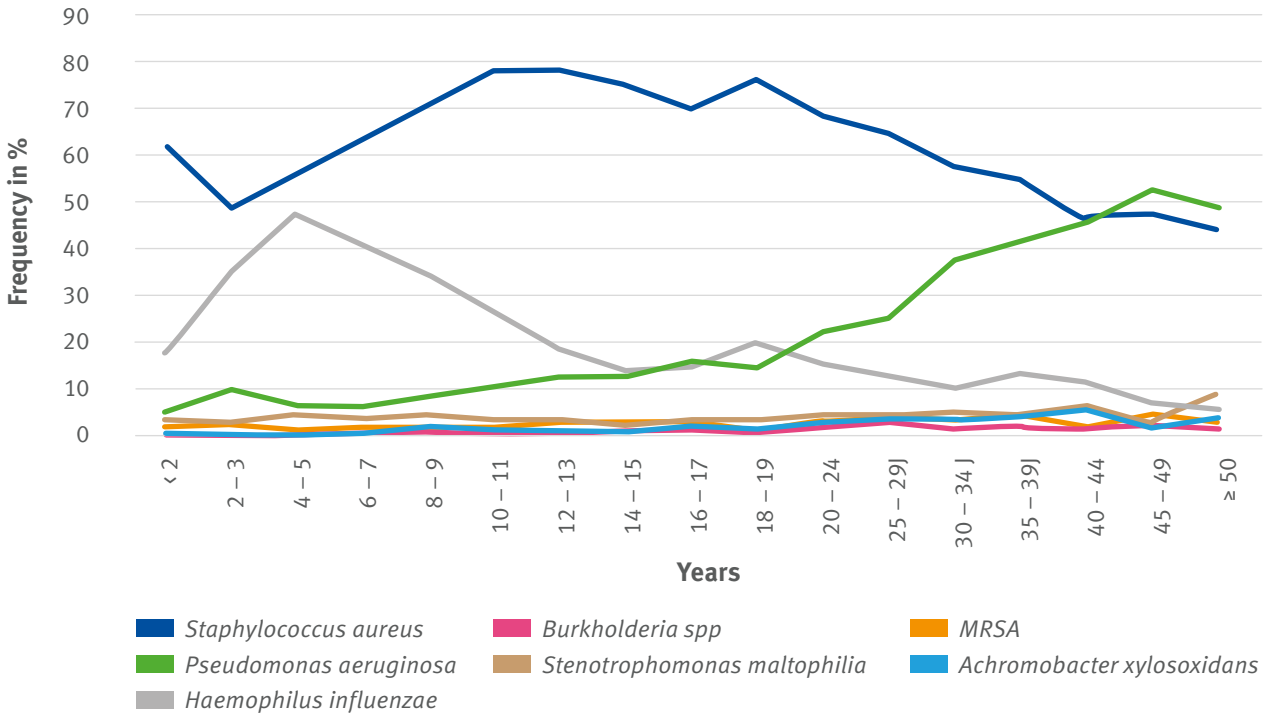

Figure 20: Detection of bacteria in pwCF with microbiological examination 2023

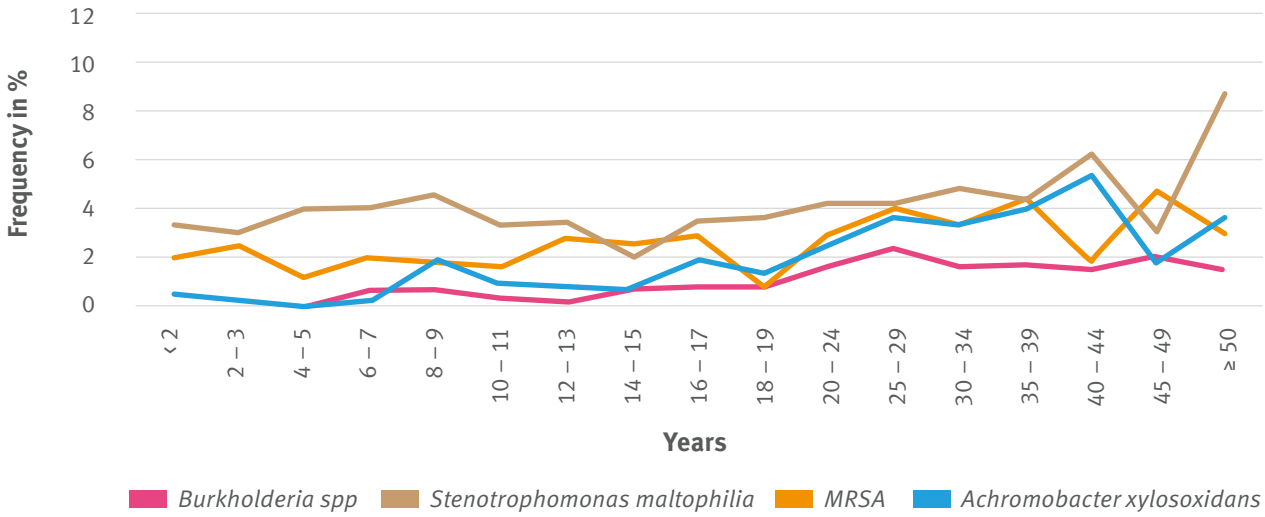

Figure 21: Bacterial detection in pwCF with microbiological examination (without the presentation of Pseudomonas aeruginosa, Staphylococcus aureus and Haemophilus influenzae) 2023

Lung infection

7a. Annual verification at leasrt once

Age (years)	*Staphylococcus aureus inklusive MRSA*	*MRSA*	*Pseudomonas aeruginosa (PSA)*	*davon MRGN*	*Burkholderia spp*	*Stenotrophomonas maltophilia*	*Achromobacter xylosoxidans*	*Haemophilus influenzae*
< 2	61.8	1.9	4.7	0.0	0.0	3.3	0.5	18.4
2 – 3	49.0	2.4	9.5	3.6	0.0	3.0	0.3	34.8
4 – 5	55.8	1.2	6.4	4.8	0.0	4.0	0.0	47.3
6 – 7	62.8	2.0	6.3	0.0	0.6	4.0	0.3	40.3
8 – 9	70.4	1.8	8.4	14.3	0.6	4.5	1.8	34.0
10 – 11	78.1	1.6	10.0	0.0	0.3	3.2	1.0	26.4
12 – 13	78.2	2.8	12.3	10.0	0.3	3.4	0.9	18.5
14 – 15	75.2	2.6	12.4	8.1	0.7	2.0	0.7	13.4
16 – 17	69.9	2.9	15.8	22.0	0.9	3.5	1.9	14.2
18 – 19	76.3	0.7	14.8	15.6	0.7	3.6	1.3	19.7
20 – 24	68.9	2.9	22.3	20.3	1.7	4.2	2.6	15.1
25 – 29	64.6	4.0	24.9	28.1	2.3	4.2	3.5	12.8
30 – 34	57.8	3.3	37.2	27.1	1.6	4.9	3.3	10.1
35 – 39	55.0	4.4	41.7	35.2	1.7	4.4	4.0	13.1
40 – 44	46.9	1.8	45.6	40.1	1.5	6.2	5.4	11.3
45 – 49	46.8	4.7	51.9	35.0	2.1	3.0	1.7	6.4
≥ 50	44.0	3.0	48.8	39.3	1.5	8.7	3.7	5.7
Total	62.3	2.8	24.0	27.9	1.2	4.3	2.3	18.7
< 18	67.1	2.1	9.7	9.0	0.4	3.5	0.8	28.1
≥ 18	58.9	3.2	34.0	31.7	1.7	4.9	3.3	12.1

Table 22: Detection of bacteria in pwCF with microbiological examination (frequencies in %) 2023

07

Lung infection

7a. Annual verification at least once

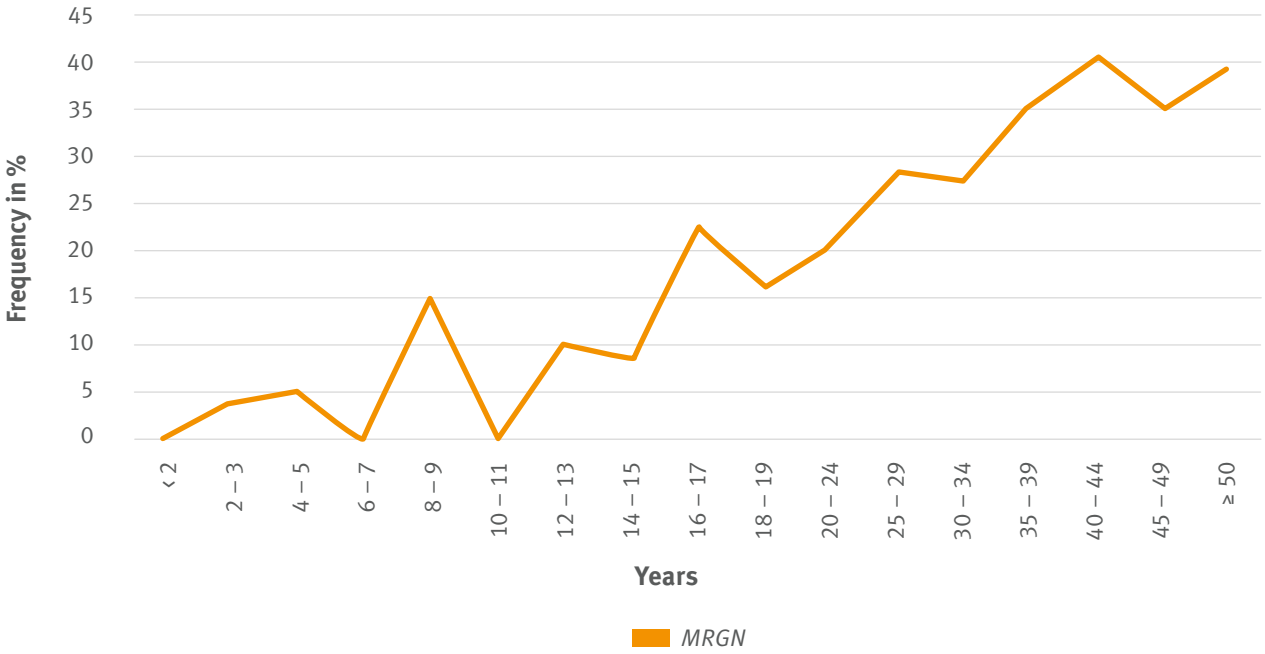

Figure 22: Bacterial detections for PSA multidrug-resistant (MRGN) in pwCF with PSA infection 2023

7b. Development of infections with Pseudomonas aeruginosa 2000 – 2023

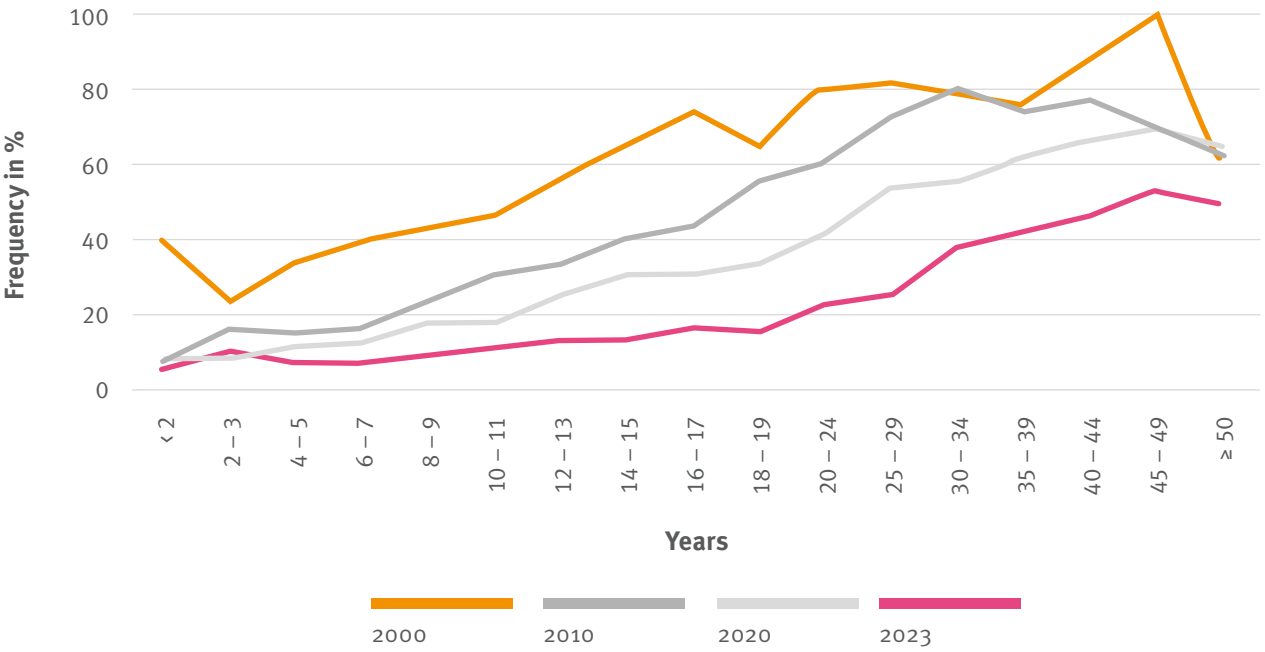

Figure 23: Development of Pseudomonas aeruginosa detection in pwCF with microbiological examination and at least annual detection 2000 – 2023

Lung infection

7b. Development of infections with Pseudomonas aeruginosa 2000 – 2023

Age (years)	2000	2010	2020	2023
<2	38.3	6.7	6.8	4.7
2 – 3	23.1	15.4	7.8	9.5
4 – 5	32.8	14.6	10.4	6.4
6 – 7	38.8	16.2	11.8	6.3
8 – 9	42.9	23.2	16.7	8.4
10 – 11	46.7	30.1	17.3	10.0
12 – 13	56.4	32.7	24.3	12.3
14 – 15	64.5	39.6	29.8	12.4
16 – 17	72.2	43.1	30.3	15.8
18 – 19	64.9	54.6	32.9	14.8
20 – 24	79.6	60.3	41.2	22.3
25 – 29	81.0	72.0	53.6	24.9
30 – 34	77.8	79.6	55.5	37.2
35 – 39	75.6	73.1	61.5	41.7
40 – 44	87.5	76.6	66.0	45.6
45 – 49	100.0	69.1	68.8	51.9
≥ 50	60.0	62.0	63.7	48.8
Total	57.1	46.9	37.4	23.9
< 18	47.2	26.7	17.3	9.6
≥ 18	76.7	67.9	53.1	34.0

Table 23: Development of Pseudomonas aeruginosa detection in pwCF with microbiological examination and at least once a year (frequencies in %) 2000 – 2023

07

Lung infection

7c. Chronic lung infections

All patients without transplantation were included in the analysis of chronic lung infections, who had at least one microbiological examination in the calendar year (n=6,719).No information on the microbiological examination in the calendar year was available for 118 patients (1.7 %).

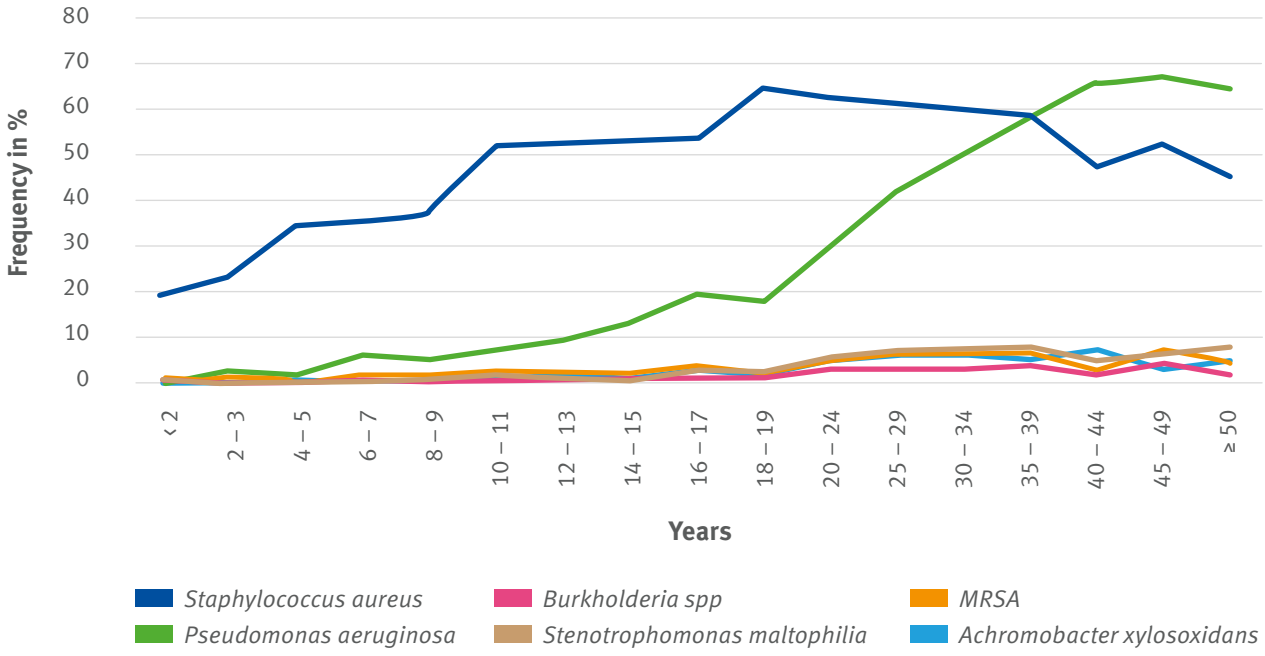

Figure 24: Chronic lung infections in pwCF with microbiological examination 2023

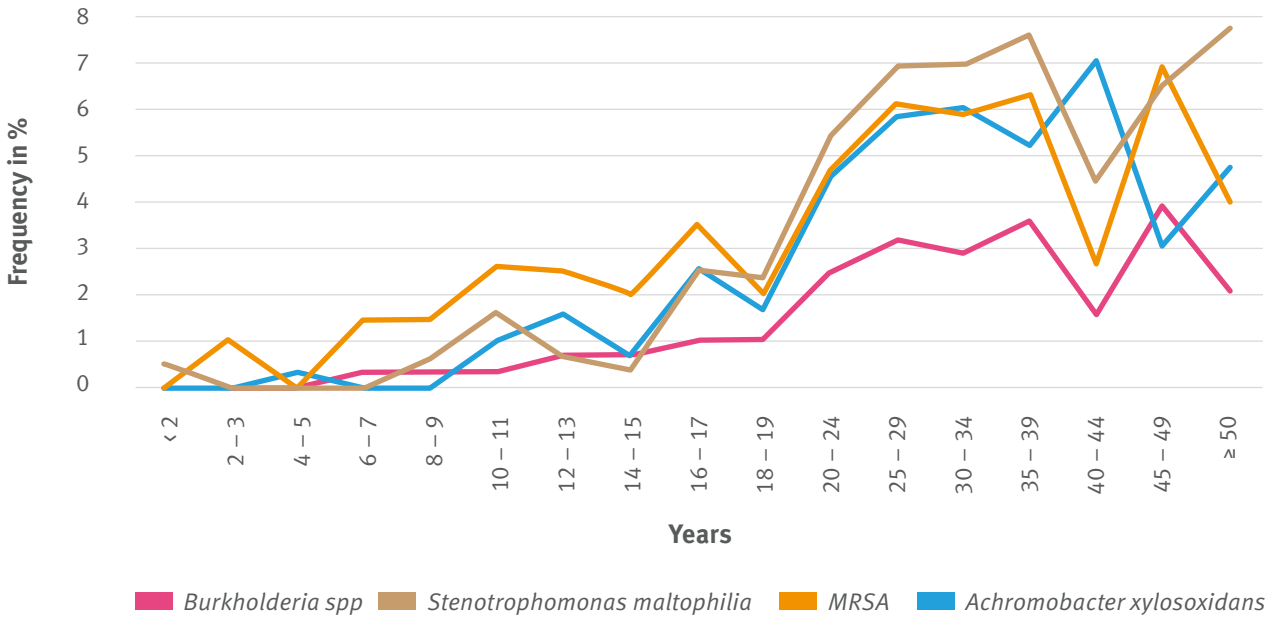

Figure 25: Chronic lung infections in pwCF with microbiological examination (without the presentation of Pseudomonas aeruginosa and Staphylococcus aureus) 2023

Lung infection

7c. Chronic lung infections

Age (years)	*Staphylococcus aureus inklusive MRSA chronisch*	*MRSA chronisch*	*Pseudomonas aeruginosa (PSA) chronisch*	*Burkholderia spp chronisch*	*Stenotrophomonas maltophilia chronisch*	*Achromobacter xylosoxidans chronisch*
<2	19.3	0.0	0.0	0.0	0.5	0.0
2 – 3	23.3	1.0	2.4	0.0	0.0	0.0
4 – 5	34.2	0.0	1.8	0.0	0.3	0.3
6 – 7	35.2	1.4	5.8	0.3	0.0	0.0
8 – 9	37.3	1.5	5.1	0.3	0.6	0.0
10 – 11	52.1	2.6	7.1	0.3	1.6	1.0
12 – 13	52.6	2.5	8.9	0.6	0.6	1.5
14 – 15	52.9	2.0	13.0	0.7	0.3	0.7
16 – 17	53.5	3.5	19.0	1.0	2.5	2.5
18 – 19	63.9	2.0	17.7	1.0	2.3	1.6
20 – 24	62.9	4.6	30.0	2.5	5.4	4.5
25 – 29	60.6	6.1	41.9	3.1	6.9	5.8
30 – 34	59.8	5.8	50.0	2.8	6.9	6.0
35 – 39	58.5	6.3	58.1	3.5	7.6	5.2
40 – 44	46.9	2.6	65.3	1.6	4.4	7.0
45 – 49	51.9	6.9	66.5	3.9	6.4	3.0
≥ 50	44.8	4.0	63.9	2.0	7.7	4.7
Total	50.5	3.6	30.8	1.7	3.9	3.2
<18	40.8	1.7	7.2	0.4	0.7	0.7
≥ 18	57.4	5.0	47.5	2.6	6.2	5.0

Table 24: Chronic lung infections in pwCF with microbiological examination (frequencies in %) 2023

07

Lung infection

7d. Atypical mycobacteria

The analysis included all patients without a transplant and at least one examination for mycobacteria in 2023 (n=1,511).

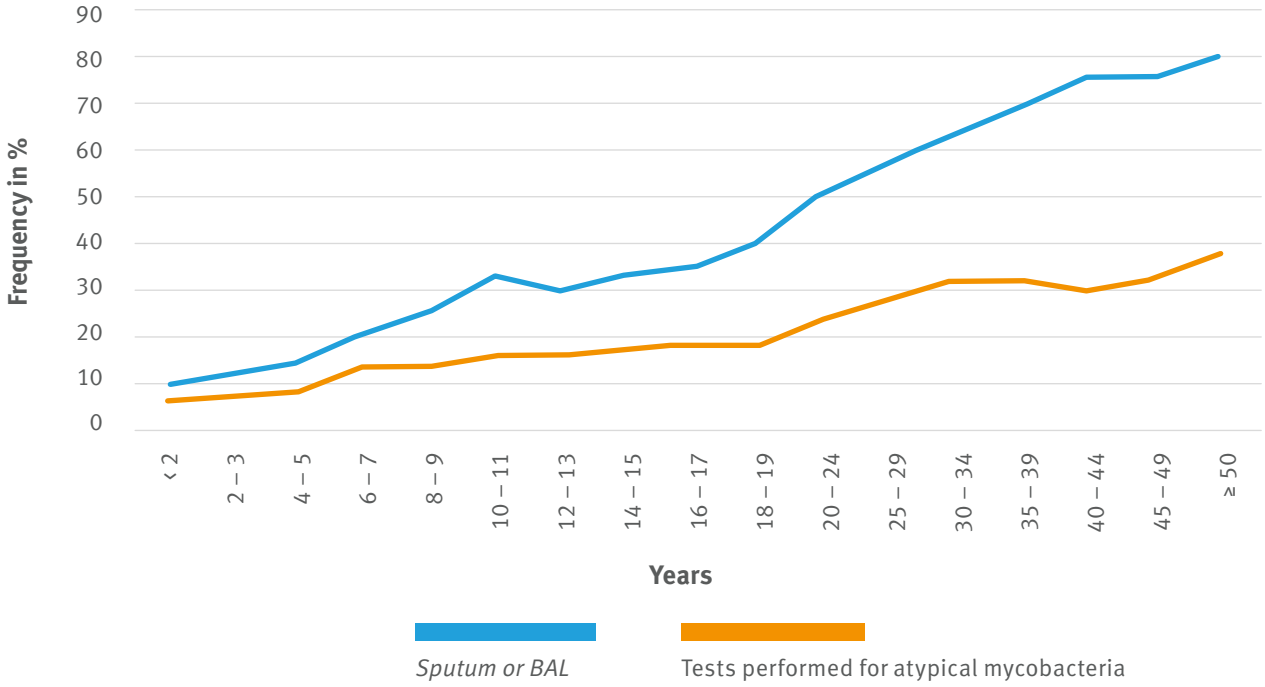

Figure 26: Frequency of pwCF with sputum or BAL and the tests performed for atypical mycobacteria 2023

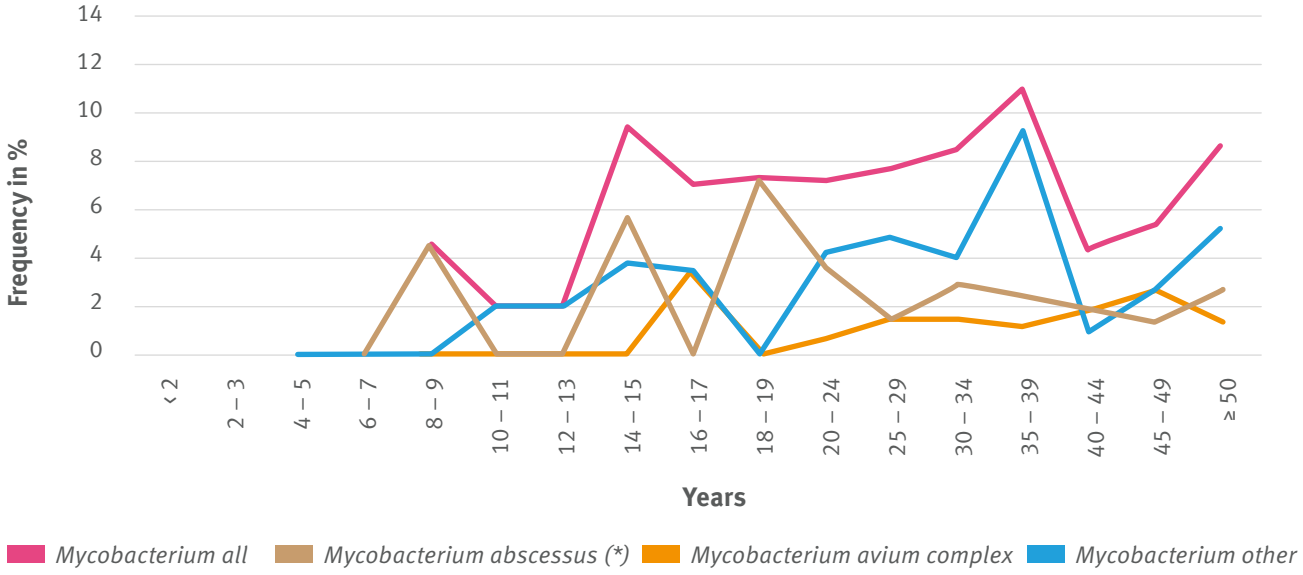

Figure 27: Age-dependent frequency of pwCF with tests for atypical mycobacteria 2023

(*) 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	20	9.4	14	70.0	0	0.0	0	0.0	0	0.0	0	0.0
2 – 3	34	11.5	22	64.7	1	4.6	0	0.0	0	0.0	1	5.3
4 – 5	47	14.3	27	57.5	0	0.0	0	0.0	0	0.0	0	0.0
6 – 7	72	20.8	48	66.7	0	0.0	0	0.0	0	0.0	0	2.4
8 – 9	86	25.7	44	51.2	2	4.6	0	0.0	0	0.0	2	2.2
10 – 11	103	33.1	50	48.5	0	0.0	0	0.0	1	2.0	1	0.0
12 – 13	95	29.2	51	53.7	0	0.0	0	0.0	1	2.0	1	0.0
14 – 15	105	34.1	53	50.5	3	5.7	0	0.0	2	3.8	5	6.1
16 – 17	111	35.1	57	51.4	0	0.0	2	3.5	2	3.5	4	4.4
18 – 19	122	40.0	55	45.1	4	7.3	0	0.0	0	0.0	4	9.1
20 – 24	351	50.8	167	47.6	6	3.6	1	0.6	7	4.2	12	8.2
25 – 29	424	57.1	208	49.1	3	1.4	3	1.4	10	4.8	16	8.7
30 – 34	402	63.3	201	50.0	6	3.0	3	1.5	8	4.0	17	6.6
35 – 39	376	69.1	173	46.0	4	2.3	2	1.2	16	9.3	19	7.1
40 – 44	293	75.5	115	39.3	2	1.7	2	1.7	1	0.9	5	3.1
45 – 49	176	75.5	75	42.6	1	1.3	2	2.7	2	2.7	4	2.5
≥ 50	321	79.9	151	47.0	4	2.7	2	1.3	8	5.3	13	12.8
Total	3,138	46.7	1,511	48.2	36	2.4	17	1.1	58	3.8	104	6.3
<18	673	24.2	366	54.4	6	1.6	2	0.6	6	1.6	14	2.2
≥ 18	2,465	62.6	1,145	46.5	30	2.6	15	1.3	52	4.5	90	7.6

Table 25: pwCF with a test for atypical mycobacteria (frequency in %) 2023

[1] Sputum or BAL: In relation 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

For the analysis of complications, all patients without transplantation who answered the question about complications were included. There were 6,829 data sets available. For a total of 8 patients (0.1 %) the question about complications was not answered.

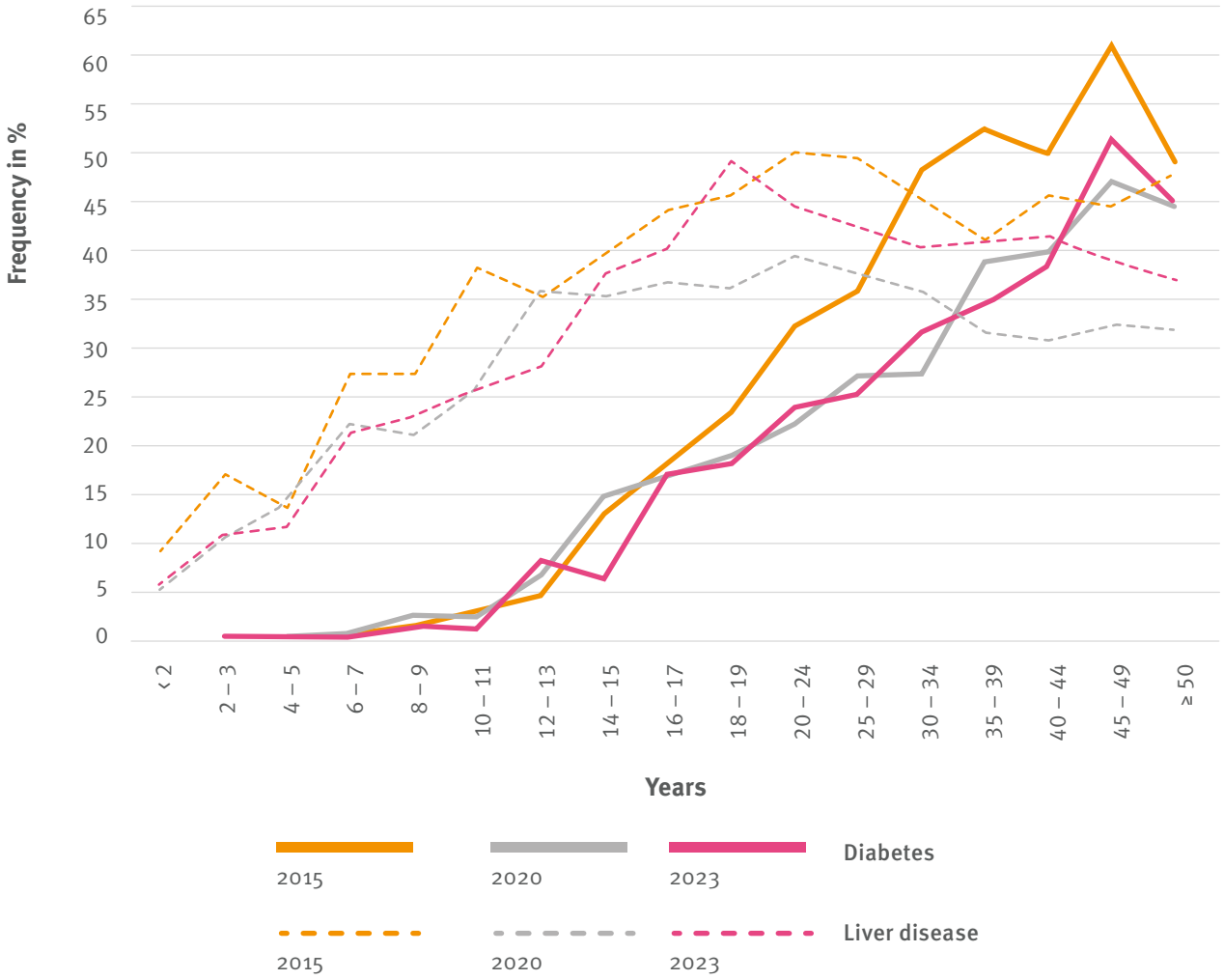

Figure 28: Development of diabetes detection and liver disease in pwCF 2015 – 2023

Complications

8a. 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	0.4	2.3	1.0
Haemoptysis	0.0	0.0	0.2	0.1
of these, at least one serious episode (› 240 ml in 24h)	–	–	0.0	0.0
Pneumothorax	0.0	0.1	0.0	0.0
of these, requiring drainage	–	0.0	–	0.0
Respiratory insufficiency	0.6	0.6	1.2	0.8
of these, partial insufficiency	100.0	100.0	81.8	90.9
of these, global insufficiency	0.0	0.0	9.1	4.6
Gastrointestinal complications				
Exocrine pancreatic insufficiency	89.3	91.9	90.8	90.7
Distal intestinal obstruction syndrome (DIOS)	1.8	2.1	2.6	2.2
Liver disease	9.8	23.2	35.1	23.2
of these, liver cirrhosis	0.0	5.2	16.8	10.5
of these, with portal hypertension	0.0	2.6	6.6	4.3
of these, without portal hypertension	0.0	2.6	8.1	5.1
Gastroesophageal reflux	1.2	1.2	3.2	1.9
Other complications/comorbidities				
Diabetes mellitus	0.2	1.0	10.5	4.0
of these, Type 3	50.0	60.0	95.0	91.1
of these, not Type 3	50.0	40.0	5.0	8.9
Bone disease				
Osteopenia	0.1	0.2	0.8	0.4
Osteoporosis	0.0	0.0	0.0	0.0
Arthritis/Arthropathy	0.0	0.3	1.0	0.4
Pansinusitis/Polyps	2.5	11.3	18.5	11.1
Impaired hearing	0.4	0.8	1.3	0.8
Renal insufficiency	0.1	0.1	0.1	0.1
Depression	0.0	0.0	2.0	0.7
Malignant disease	0.2	0.0	0.3	0.2
Salt-losing syndrome	1.3	0.4	0.0	0.5

Table 26: pwCF under the age of 18 with complications (frequencies in %) 2023

08

Complications

8b. Adults

Complication	18 – 29 years	30 – 39 years	≥ 40 years	Total
Pulmonary complications				
Allergic bronchopulmonary aspergillosis	3.8	3.5	2.5	3.4
Haemoptysis	2.7	4.1	5.4	3.8
of these, at least one serious episode (› 240 ml in 24h)	2.3	0.0	2.1	1.5
Pneumothorax	0.3	0.2	0.4	0.3
of these, requiring drainage	66.7	100.0	100.0	83.3
Respiratory insufficiency	4.6	7.2	12.1	7.3
of these, partial insufficiency	67.5	70.6	71.7	70.2
of these, global insufficiency	22.5	18.8	15.0	18.2
Gastrointestinal complications				
Exocrine pancreatic insufficiency	91.7	89.8	85.6	89.5
Distal intestinal obstruction syndrome (DIOS)	3.7	3.5	2.5	3.3
Liver disease	44.7	40.8	39.1	42.1
of these, liver cirrhosis	19.1	15.8	13.8	16.8
of these, with portal hypertension	8.9	6.7	7.7	8.0
of these, without portal hypertension	7.1	5.7	3.1	5.7
Gastroesophageal reflux	7.3	6.5	11.5	8.2
Other complications/comorbidities				
Diabetes mellitus	23.5	32.9	43.8	31.6
of these, Type 3	96.9	96.0	96.8	96.6
of these, not Type 3	3.1	4.0	3.2	3.5
Bone disease				
Osteopenia	10.9	19.0	25.8	17.2
Osteoporosis	4.2	8.8	17.3	9.0
Arthritis/Arthropathy	3.5	6.3	10.9	6.3
Pansinusitis/Polyps	28.8	33.9	34.8	31.9
Impaired hearing	1.6	2.5	5.5	2.9
Renal insufficiency	0.7	2.1	4.7	2.2
Depression	8.5	7.4	8.0	8.0
Malignant disease	0.2	0.6	4.2	1.4
Salzverlustsyndrom	0.2	0.3	0.1	0.2

Table 27: Cystic fibrosis patients aged 18 and over with complications (frequencies in %) 2023

Complications

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	66.0	72.7	77.9	74.1	70.2	67.7	71.7	72.5	71.1
1	16.1	15.4	13.4	16.4	18.0	18.5	16.5	14.9	17.5
2	10.0	5.5	5.4	5.4	7.1	7.9	6.7	6.8	6.6
3	4.1	3.2	1.8	2.1	2.9	3.2	2.8	3.0	2.6
4	1.9	1.1	0.6	1.2	1.0	1.1	1.1	1.2	1.1
5+	1.8	1.7	0.8	0.8	0.5	0.7	1.0	1.4	0.7
unbekannt	0.1	0.3	0.1	0.1	0.3	0.9	0.3	0.2	0.4

Table 28: Number of exacerbations treated with antibiotics per cystic fibrosis patient (frequencies in %) 2023

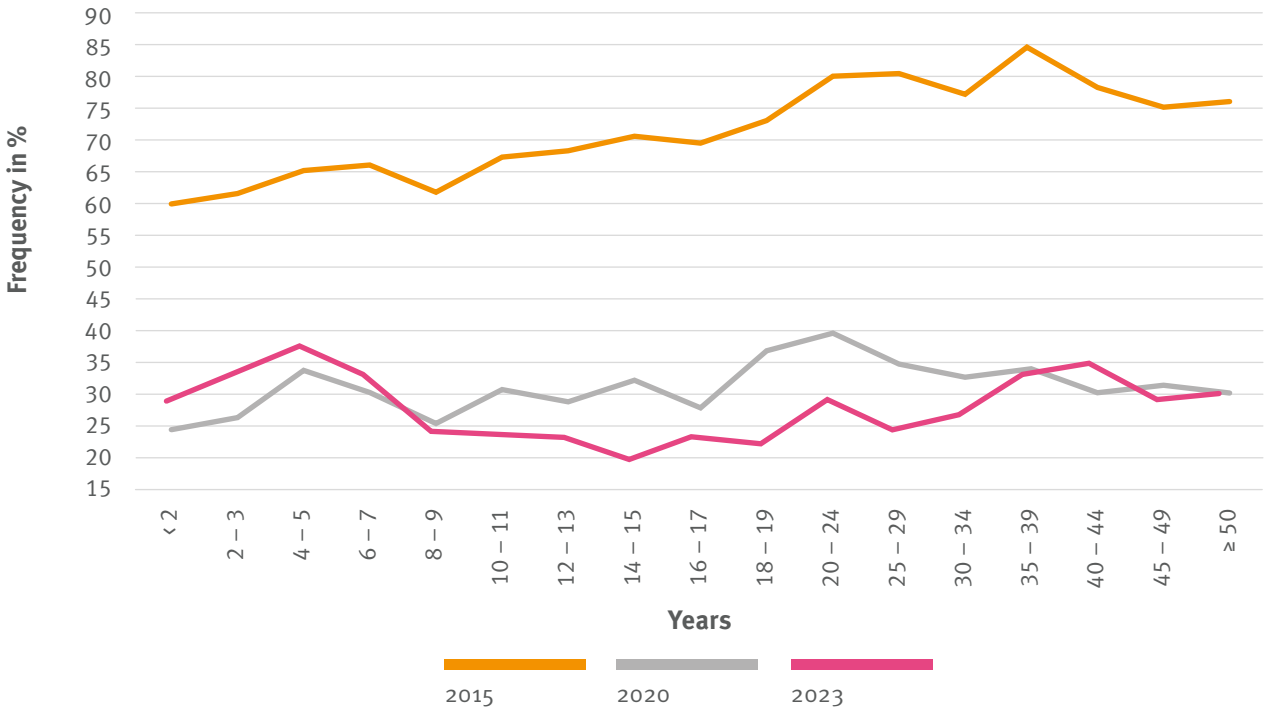

Figure 29: Development of age-related frequencies of pwCF with at least 1 antibiotic-treated exacerbation 2015 – 2023

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	59.7	61.5	65.5	66.1	62.0	67.2	68.3	70.5	69.5	73.1	80.1	80.5	77.3	84.6	78.3	75.1	76.3
2020	24.2	26.2	34.0	30.2	25.5	30.8	28.9	32.6	27.9	36.9	39.7	34.8	32.8	33.9	30.2	31.2	30.0
2023	28.7	33.4	37.7	32.6	24.1	23.8	22.9	19.7	23.3	22.5	28.9	24.3	26.8	32.7	34.4	29.2	29.9

Table 29: Development of age-related frequencies (in %) of pwCF with at least 1 antibiotic-treated exacerbation 2015 – 2023

09

Therapies

9a. Basic therapy

For the evaluation of the basic and indication therapy, all patients without transplantation were included who answered the question about long-term gastrointestinal or pulmonary therapy were included.The data sets of 2,784 patients under the age of 18 and 4,044 patients aged 18 and over were included in the analyses. For 10 patients (0.2 %), the question about long-term gastrointestinal or pulmonary therapy was not answered.

9a.i Children and adolescents under 18 years of age

Basic therapy	0 – 5 years	6 – 11 years	12 – 17 years	Total
DNase	15.7	39.5	50.5	36.1
Mannitol	0.0	0.0	0.4	0.1
Hypotonic saline solution (≥3 %)	90.6	96.8	91.5	93.1
of these 3 – 5,7 %	22.3	21.5	17.8	20.5
of these ≥ 5,8 %	77.7	78.5	82.2	79.5
At least one mucolytic therapy (Mannitol, DNAse, Hypertonic saline solution ≥3 %)	90.7	97.6	93.6	94.2
ß2-sympathomimetics				
Short-acting (SABA)	61.6	69.8	66.2	66.1
Long-acting (LABA)	4.9	11.3	20.4	12.5
Anticholinergics	5.3	9.4	13.2	9.4
Antistaphylococcal therapy	7.3	6.3	7.6	7.1
Steroids				
Nasal	10.8	20.2	21.7	17.9
Inhalative	8.9	13.6	21.6	14.9
Orale	0.7	0.7	1.5	1.0
Vitamins				
Vitamin A	79.5	82.9	80.7	81.1
Vitamin D	96.7	96.2	95.7	96.2
Vitamin E	68.0	73.2	76.7	72.8
Vitamin K	75.0	77.0	73.6	75.2
Hormonal contraception for women[1]	–	–	7.7	23.4

Table 30: pwCF under the age of 18 with basic therapy (frequencies in %) 2023
[1] Survey from the age of 14

Therapies

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

The graph on inhalation and combination therapies includes all minors who answered the question about long-term pulmonary therapy (2,784 patients). 148 patients (5.3 %) of these did not receive any inhalation therapy.

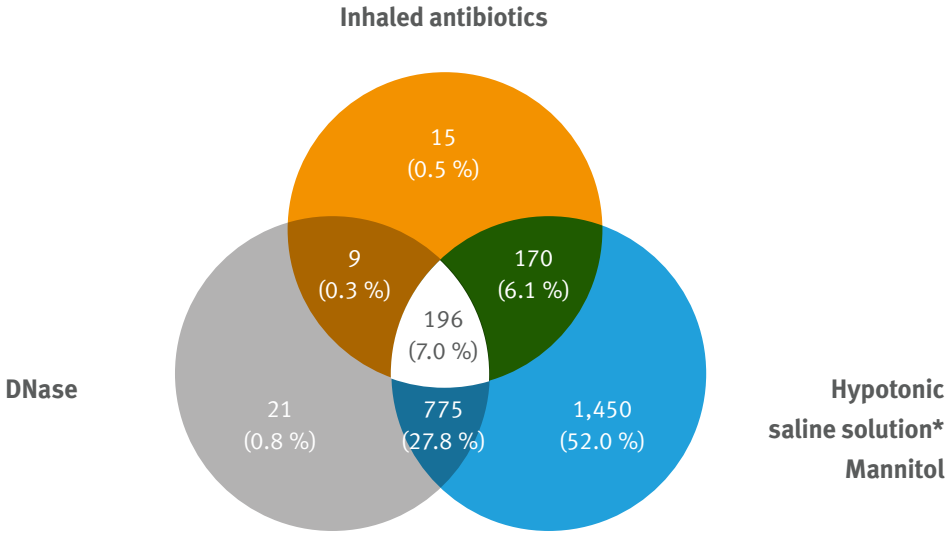

Figure 30: Inhalation and combination therapies for pwCF under 18 years of age 2023
*Hypertonic saline solution ≥ 3 %

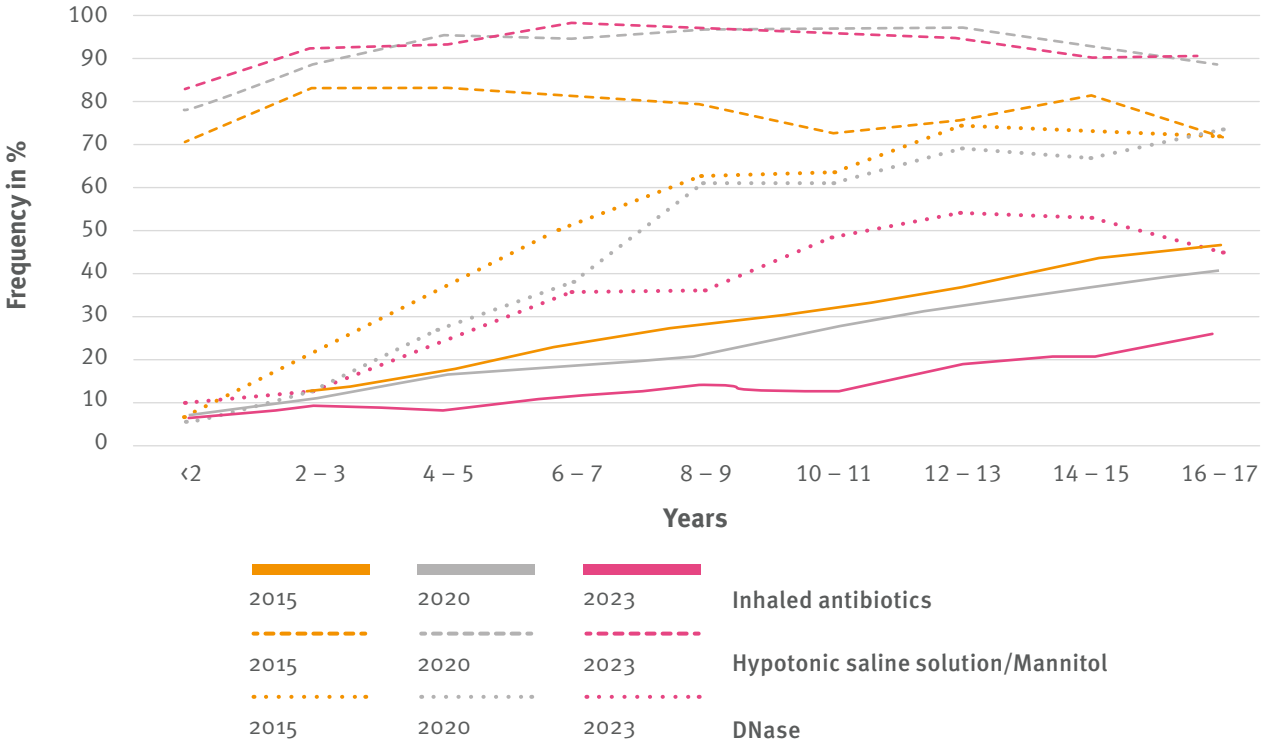

Figure 31: Development of basic therapy for pwCF 2015 – 2023

09

Therapies

9a. Basic therapy

9a.iii Adults

Basic therapy	18 – 29 years	30 – 39 years	≥ 40 years	Total
DNase	47.8	40.0	36.6	42.5
Mannitol	2.3	4.8	4.4	3.6
Hypotonic saline solution (≥3 %)	79.0	68.8	65.3	72.4
of these 3 – 5,7 %	20.9	23.0	26.1	22.7
of these ≥ 5,8 %	79.1	77.0	74.0	77.3
At least one mucolytic therapy (Mannitol, DNAse, Hypertonic saline solution ≥3 %)	86.3	79.4	77.1	81.8
ß2-sympathomimetics				
Short-acting (SABA)	61.1	55.3	61.8	59.5
Long-acting (LABA)	35.6	57.0	66.5	50.2
Anticholinergics	28.1	46.4	61.4	42.3
Antistaphylococcal therapy	6.4	4.5	2.6	4.8
Steroids				
Nasal	22.1	23.4	20.5	22.1
Inhalative	28.5	44.9	52.9	39.8
Orale	3.3	4.3	8.6	5.0
Vitamins				
Vitamin A	75.6	67.1	60.6	69.1
Vitamin D	94.6	93.2	92.7	93.7
Vitamin E	74.7	65.7	59.9	68.1
Vitamin K	71.7	64.0	59.4	66.1
Hormonal contraception for women	31.5	21.2	11.1	20.9

Table 31: pwCF aged 18 and over with basic therapy (frequencies in %) 2023

Therapies

9a.iiii Inhalation and combination therapies Adults

The graph on inhalation and combination therapies includes all adults who answered the question about long-term pulmonary therapy (4,044 patients).Of these, 478 patients (12.0 %) did not receive any pulmonary therapy.

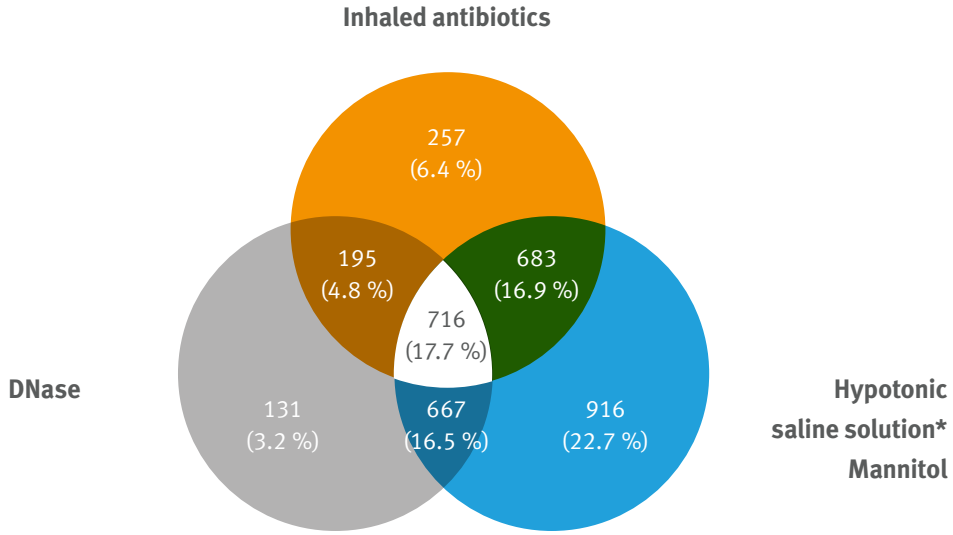

Figure 32: Inhalation and combination therapies for pwCF under 18 years of age 2023
*Hypertonic saline solution ≥ 3 %

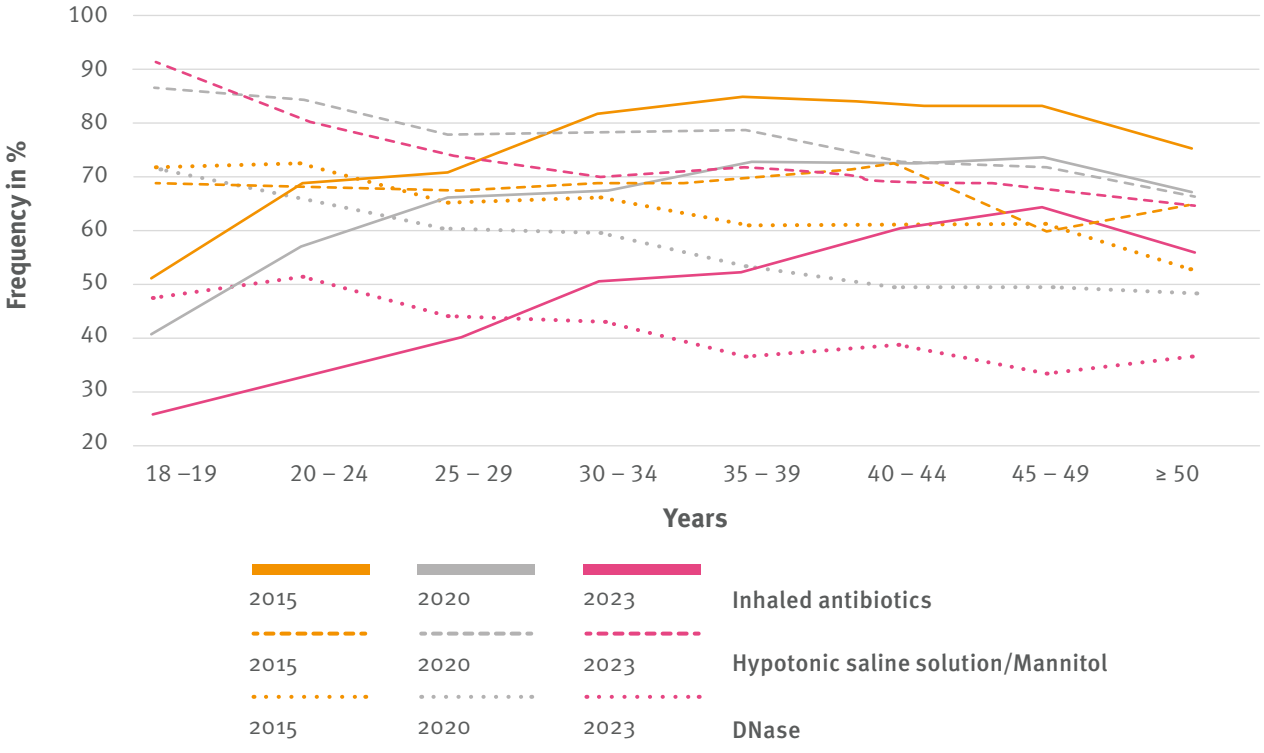

Figure 33: Development of basic therapy for pwCF 2015 – 2023

09

Therapies

9b. Indication therapy

9b.i Children and adolescents under 18 years of age

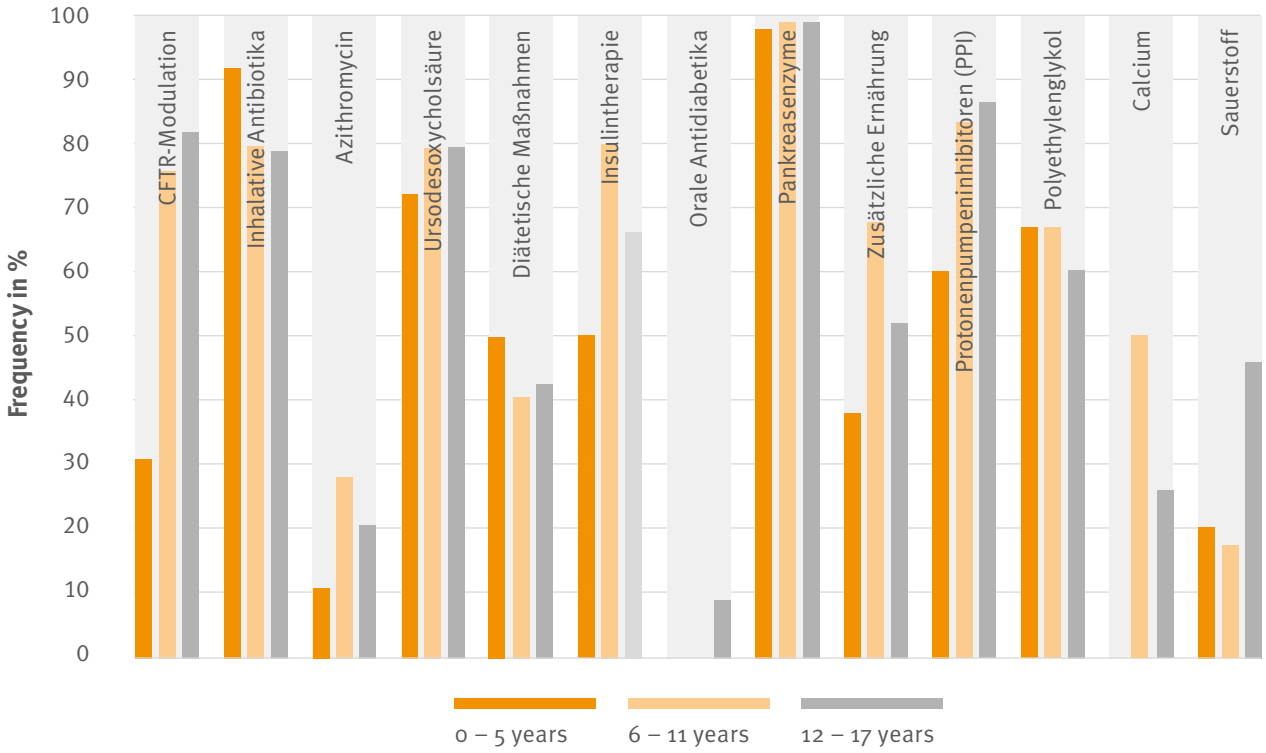

Figure 34: pwCF under the age of 18 with indication therapy 2023

Status of the approval of CFTR modulators 31/12/2023
Only the last modulator used in the reporting year is shown here.

[1] Ivacaftor has been authorised from the age of 1 year since 2018 and from 4 months since 2020 for patients with the mutations G551D, G1244E, G1349D, G178R,G551S, S1251N, S1255P, S549N or S549R.
Ivacaftor has been approved for patients with the R117H mutation from the age of 18 since 2018, also from 4 months of age since 2020.

[2] Lumacaftor/ivacaftor has been approved for F508del homozygous patients from 6 years of age since 2018, from 2 years of age since 2019 and from 1 year of age since July 2023.

[3] Tezacaftor/ivacaftor has been approved for F508del homozygous patients from 12 years of age since 2019 or for F508del heterozygous patients with one of the following mutations P67L, R117C, L206W, R352Q, A544E, 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 the age of 12 for F508del homozygous patients or F508del heterozygous patients with a minimal function (MF) mutation, since 2021 for all patients from the age of 12 years of age, since 2022 from 6 years of age and since November 2023 from 2 years of age with at least one F508del mutation. The use of elexacaftor/tezacaftor/ivacaftor before 2020 was possible in clinical trials.

Therapies

9b. Indication therapy

9b.i Children and adolescents under 18 years of age

Indication therapy	0 – 5 years	6 – 11 years	12 – 17 years	Total
CFTR-Modulation	30.4	75.6	81.6	64.0
of these, Ivacaftor[1]	2.5	2.3	3.1	2.6
of these, Lumacaftor/ Ivacaftor[2]	24.4	8.9	1.6	11.1
of these, Tezacaftor/ Ivacaftor[3]	0.0	0.5	0.8	0.5
of these, Elexacaftor/ Tezacaftor/ Ivacaftor[4]	6.0	73.8	80.3	59.0
Inhalative antibiotics *with chronic Pseudomonas infection*	92.3	79.7	79.1	80.1
of these, inhalative tobramycin	33.3	45.8	45.3	44.7
of these, inhalative colistin	75.0	55.9	54.3	56.0
of these, inhalative aztreonam	0.0	5.1	10.2	8.1
of these, DPI tobramycin	0.0	0.0	4.7	3.0
of these, DPI colistin	0.0	5.1	10.2	8.1
of these, levofloxacin	0.0	3.4	0.0	1.0
of these, inhalative gentamicin	0.0	0.0	0.0	0.0
of these, others	0.0	3.4	1.6	2.0
Azithromycin with *chronic Pseudomonas infection*	9.1	27.1	19.5	21.2
Ursodeoxycholic acid *with liver disease*	72.0	79.1	79.3	78.3
Dietary measures *with Diabetes mellitus*	50.0	40.0	42.0	42.0
Insulin therapy *with Diabetes mellitus*	50.0	80.0	66.0	67.0
Oral antidiabetics *with Diabetes mellitus*	0.0	0.0	8.0	7.1
Pancreatic enzymes *with exocrine pancreatic insufficiency*	98.1	99.1	99.1	98.8
Additional nutrition *with underweight*	37.4	67.7	51.6	52.5
Additional oral nutrition	36.3	64.9	46.8	49.6
PEG	0.8	2.3	4.8	2.6
Proton pump inhibitors (PPI) *with gastroesophageal reflux*	60.0	83.3	86.7	80.8
Polyethylene glycol *with DIOS*	66.7	66.7	60.0	63.9
Calcium *with osteoporosis/osteopenia*	0.0	50.0	25.0	27.3
Oxygen *with respiratory insufficiency*	20.0	16.7	45.5	31.8

Table 32: MpwCF under the age of 18 with indication therapy (frequencies in %) 2023

09

Therapies

9b. Indication therapy

9b.ii Adults

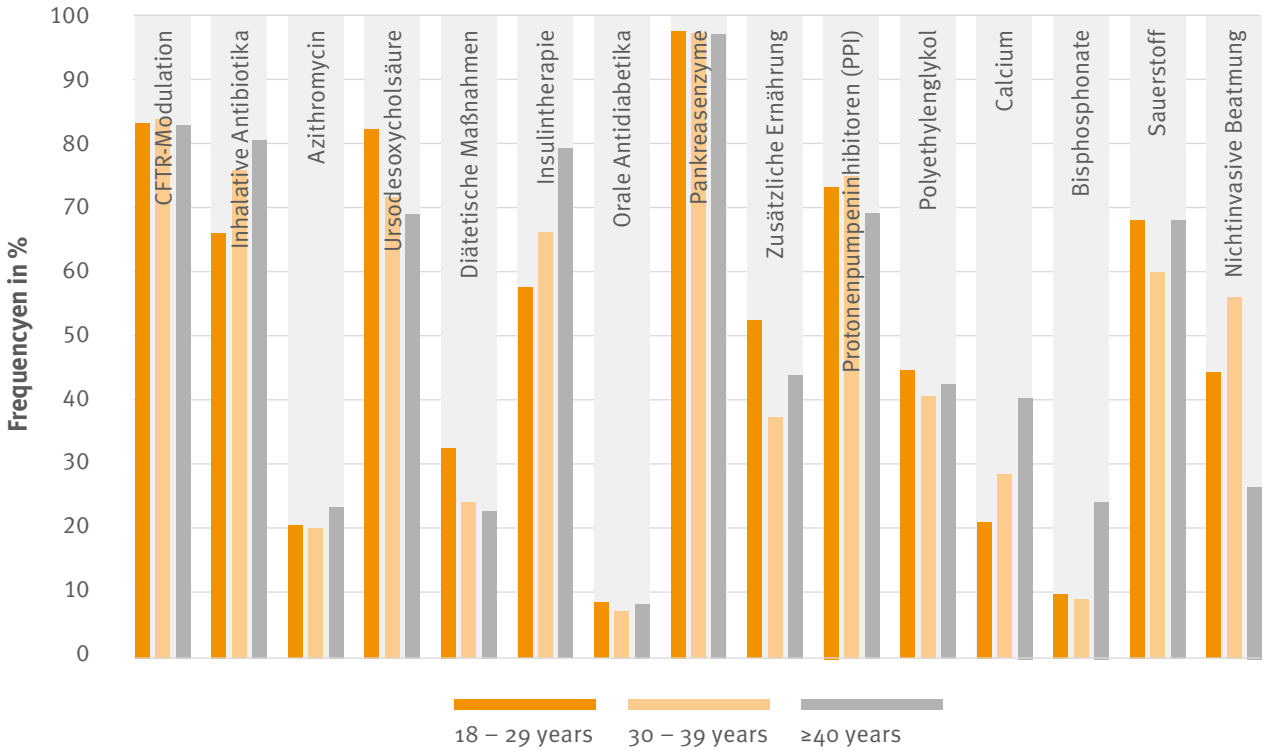

Figure 35: pwCF aged 18 and over with indication therapy 2023

Status of the approval of CFTR modulators 31/12/2023
Only the last modulator used in the reporting year is shown here.

[1] Ivacaftor has been authorised from the age of 1 year since 2018 and from 4 months since 2020 for patients with the mutations G551D, G1244E, G1349D, G178R,G551S, S1251N, S1255P, S549N or S549R.
Ivacaftor has been approved for patients with the R117H mutation from the age of 18 since 2018, also from 4 months of age since 2020.

[2] Lumacaftor/ivacaftor has been approved for F508del homozygous patients from 6 years of age since 2018, from 2 years of age since 2019 and from 1 year of age since July 2023.

[3] Tezacaftor/ivacaftor has been approved for F508del homozygous patients from 12 years of age since 2019 or for F508del heterozygous patients with one of the following mutations P67L, R117C, L206W, R352Q, A544E, 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 the age of 12 for F508del homozygous patients or F508del heterozygous patients with a minimal function (MF) mutation, since 2021 for all patients from the age of 12 years of age, since 2022 from 6 years of age and since November 2023 from 2 years of age with at least one F508del mutation. The use of elexacaftor/tezacaftor/ivacaftor before 2020 was possible in clinical trials.

Therapies

9b. Indication therapy

9b.ii Adults

Indication therapy	18 – 29 years	30 – 39 years	≥40 years	Total
CFTR-Modulation	83.2	83.6	82.8	83.2
of these, Ivacaftor[1]	3.1	1.6	3.0	2.6
of these, Lumacaftor/ Ivacaftor[2]	0.7	0.8	0.0	0.6
of these, Tezacaftor/ Ivacaftor[3]	1.6	1.2	2.9	1.8
of these, Elexacaftor/ Tezacaftor/ Ivacaftor[4]	81.9	82.3	80.8	81.7
Inhalative antibiotics *with chronic Pseudomonas infection*	66.0	75.9	80.4	74.5
of these, inhalative tobramycin	22.5	22.3	17.1	20.5
of these, inhalative colistin	42.0	41.8	48.9	44.4
of these, inhalative aztreonam	15.7	26.8	31.5	25.1
of these, DPI tobramycin	10.6	11.1	7.3	9.6
of these, DPI colistin	11.4	13.9	11.7	12.3
of these, levofloxacin	5.7	9.4	15.4	10.4
of these, inhalative gentamicin	0.0	0.2	0.0	0.1
of these, others	1.6	2.2	2.6	2.2
Azithromycin with *chronic Pseudomonas infection*	20.3	20.1	23.3	21.3
Ursodeoxycholic acid *with liver disease*	82.2	71.6	68.8	75.9
Dietary measures *with Diabetes mellitus*	32.5	24.0	22.5	26.2
Insulin therapy *with Diabetes mellitus*	57.6	66.3	79.5	68.3
Oral antidiabetics *with Diabetes mellitus*	8.3	7.1	8.1	7.8
Pancreatic enzymes *with exocrine pancreatic insufficiency*	97.5	97.2	97.0	97.3
Additional nutrition *with underweight*	52.2	37.3	43.8	47.1
Additional oral nutrition	49.1	35.8	40.8	44.4
PEG	3.1	1.5	4.1	2.9
Proton pump inhibitors (PPI) *with gastroesophageal reflux*	73.1	74.7	68.9	71.9
Polyethylene glycol *with DIOS*	44.6	40.5	42.3	42.9
Calcium *with osteoporosis/osteopenia*	20.4	28.2	40.1	31.4
Bisphosphonates *with osteoporosis*	9.3	8.6	24.0	16.5
Oxygen *with respiratory insufficiency*	67.9	59.8	68.0	65.5
Non-invasive ventilation (NIPPV) *with respiratory global insufficiency*	44.4	56.3	26.3	41.5

Table 33: pwCF aged 18 and over with indication therapy (frequencies in %) 2023

09

Therapies

see page 50

9c. Development of CFTR modulation therapy 2018 – 2023

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.

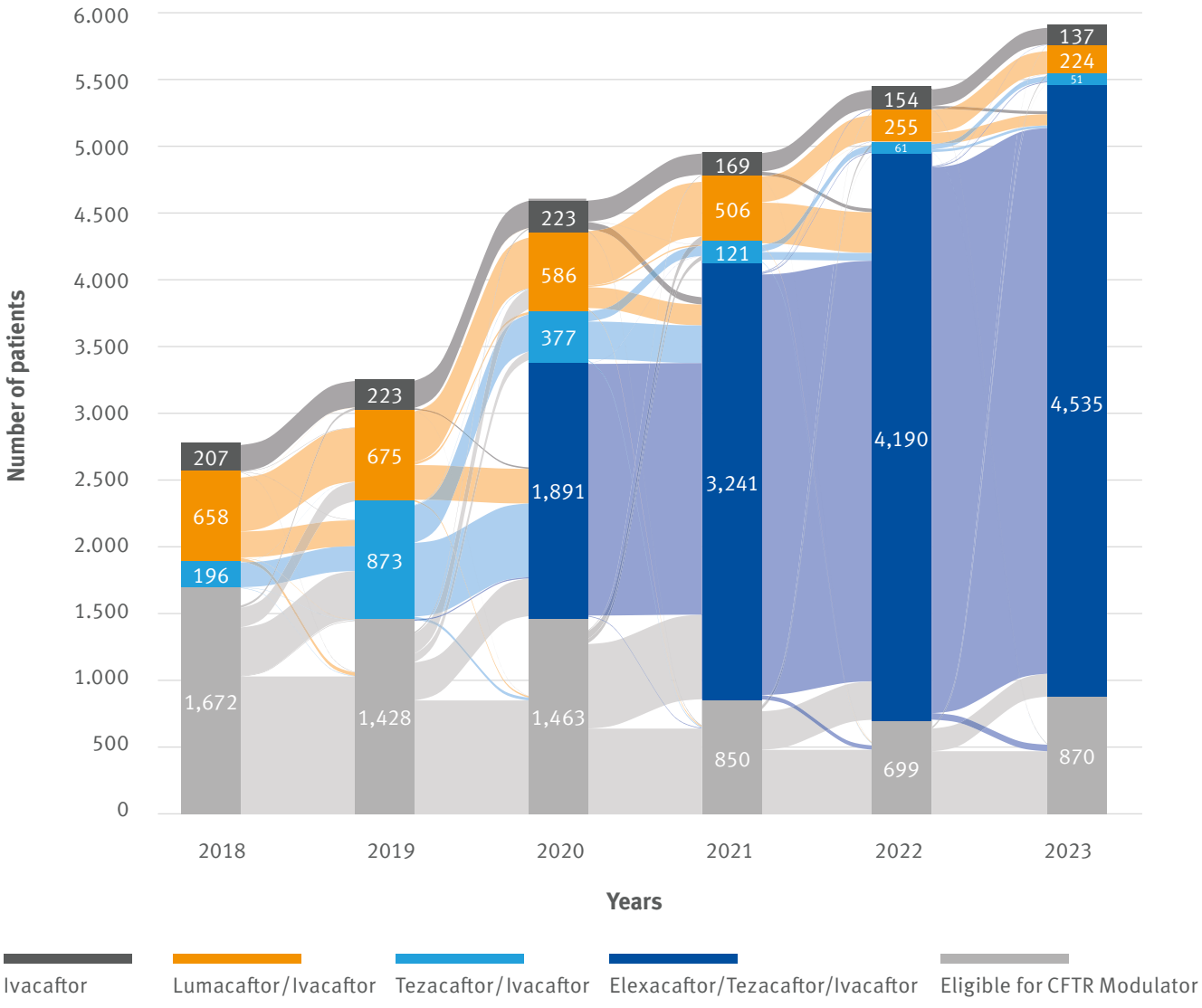

Figure 36: Number of pwCF with modulation therapy and number of patients for whom a suitable modulator is approved 2018 – 2023

Mortality

10.a Age at death

Lifespan is described by the average age at death, the average survival rate and the average age-specific life expectancy. life expectancy. The current mean age at death is reported for the year 2023, while the mean survival and life expectancy refer to a 5-year period (e.g. 2019 – 2023).

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 39 (37) years in the reporting year 2023 (2022).

In the reporting year 2023, 22 patients (11 girls/women and 11 boys/men) died. The main causes of death were cardiopulmonary diseases (36.4 %), malignant diseases (13.6 %), liver diseases/failure (9.1 %) and gastrointestinal diseases (9.1 %). Other or unknown causes were present in 31.8 % of cases. The age at death is distributed as follows:

	Number	Mean value	Median	Minimum	Maximum	25th percentile	75th percentile
Age at death in full years	22	36.9	38.5	3.0	61.0	24.0	50.0

Table 34: Age at death 2023

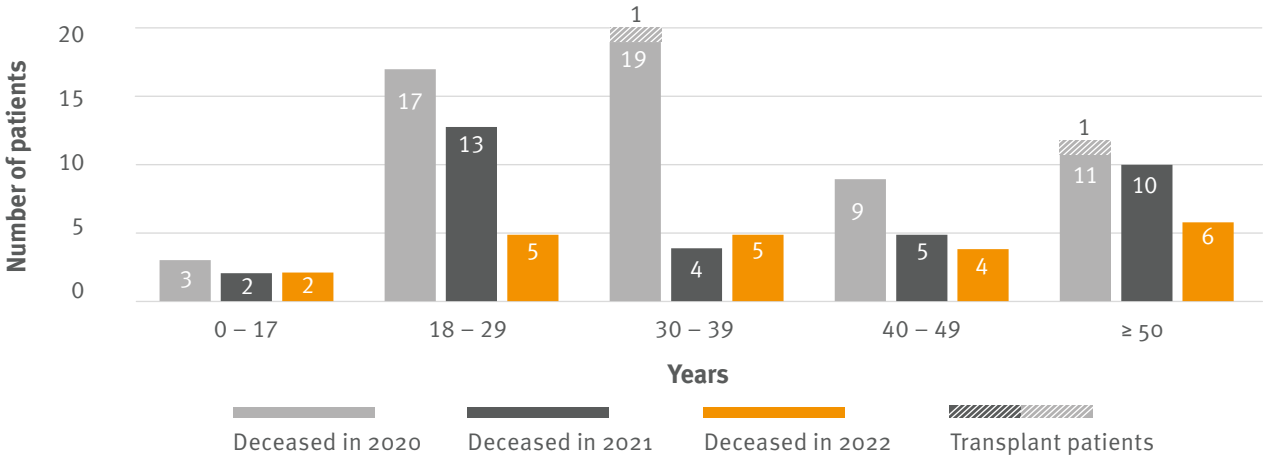

Figure 37: pwCF who died in the years 2020 – 2023

Age (years)	2020				2022				2023			
	No TX		TX		No TX		TX		No TX		TX	
	Number	%	Number	%	Number	%	Number	%	Number	%	Number	%
0 – 17	3	100.0	0	0.0	2	100.0	0	0.0	2	100.0	0	0.0
18 – 29	17	100.0	0	0.0	13	100.0	0	0.0	5	100.0	0	0.0
30 – 39	19	95.0	1	5.0	4	100.0	0	0.0	5	100.0	0	0.0
40 – 49	9	100.0	0	0.0	5	100.0	0	0.0	4	100.0	0	0.0
≥ 50	11	91.7	1	8.3	10	100.0	0	0.0	6	100.0	0	0.0
Total	59	–	2	–	34	–	0	–	22	–	0	–

Table 35: Deceased pwCF 2020 – 2023

10

Mortality

(Data status: 05.09.2024)

10.b. Median predicted survival age

The calculation of the median predicted survival age and also the median predicted life expectancy shown under point 10.c is based on the number of predicted life expectancy is based on the number of people with cystic fibrosis documented as living/deceased in the registry in the respective year and does not with cystic fibrosis in the respective year and does not take into account the possible effects of CFTR modulator therapies and other advances in clinical care. and other advances in clinical care. In recent years, the annual number of deaths has have declined sharply in recent years, resulting in less precision in predicting survival age and life expectancy. and life expectancy. This is also reflected in wider confidence intervals in the forecasts.

Median predicted survival age 2022

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. 8252 people with Cystic Fibrosis (including patients with transplants) and 297 deaths were recorded In the 5-year window between 2018 and 2022. 536 patients (6.5 %) were lost from the follow-up. The median survival age was 58.4 years (confidence interval: 56.4 to 63.8).

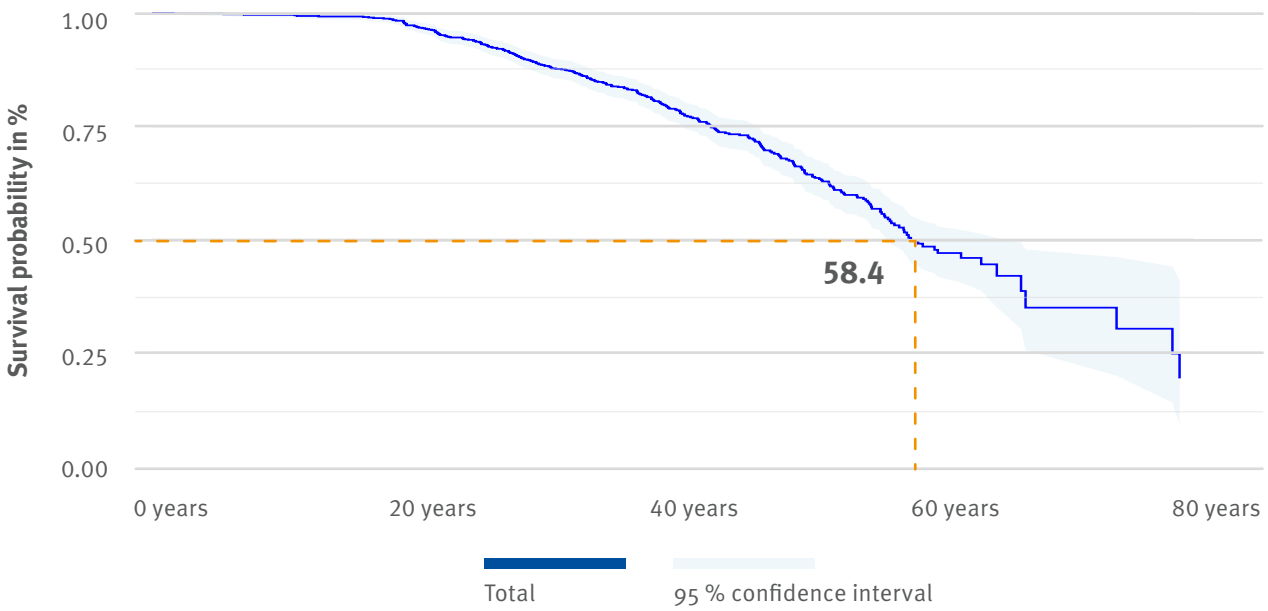

Figure 38: Median survival age for pwCF for the period 2018 – 2022

Mortality

Median predicted survival age 2023

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. 8369 people with Cystic Fibrosis (including patients with transplants) and 244 deaths were recorded In the 5-year window between 2019 and 2023. 570 patients (6.8%) were lost from the follow-up. The median survival age was 66.8 years (confidence interval: 59.3 to 74.1).

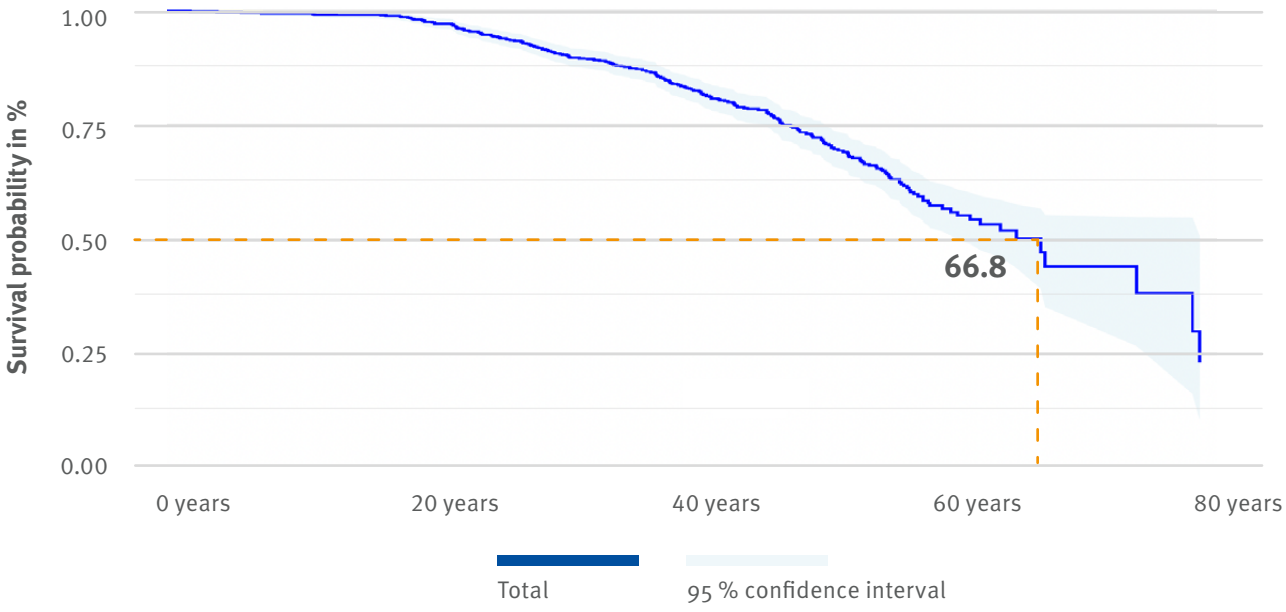

Figure 39: Median survival age for pwCF for the period 2019 – 2023

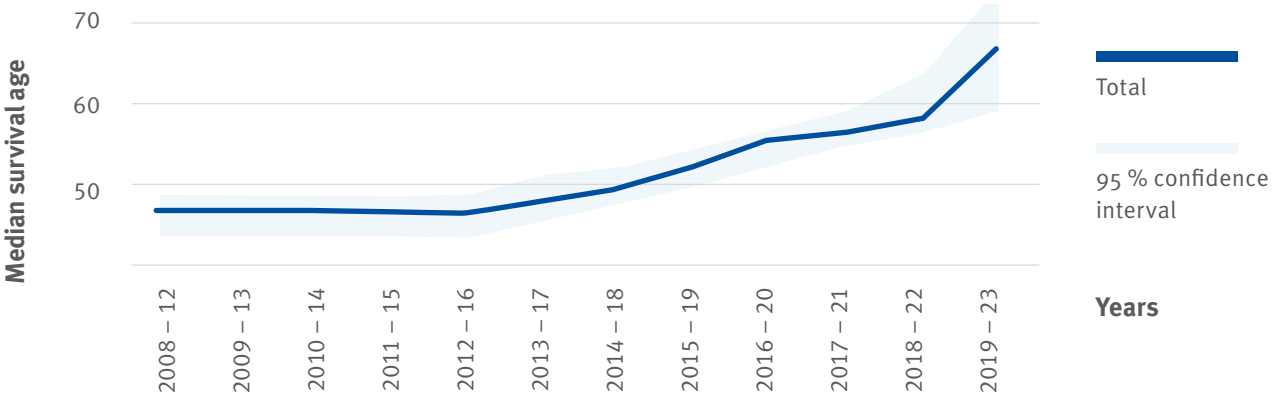

Figure 40: Median predicted survival age in the years 2012 – 2023

10

Mortality

10.c Life expectancy

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 78.2 years and that of a female newborn 83.0 years (www.statista.de). The life expectancy is different for each age and does not correspond to the median survival age.

Life expectancy 2022

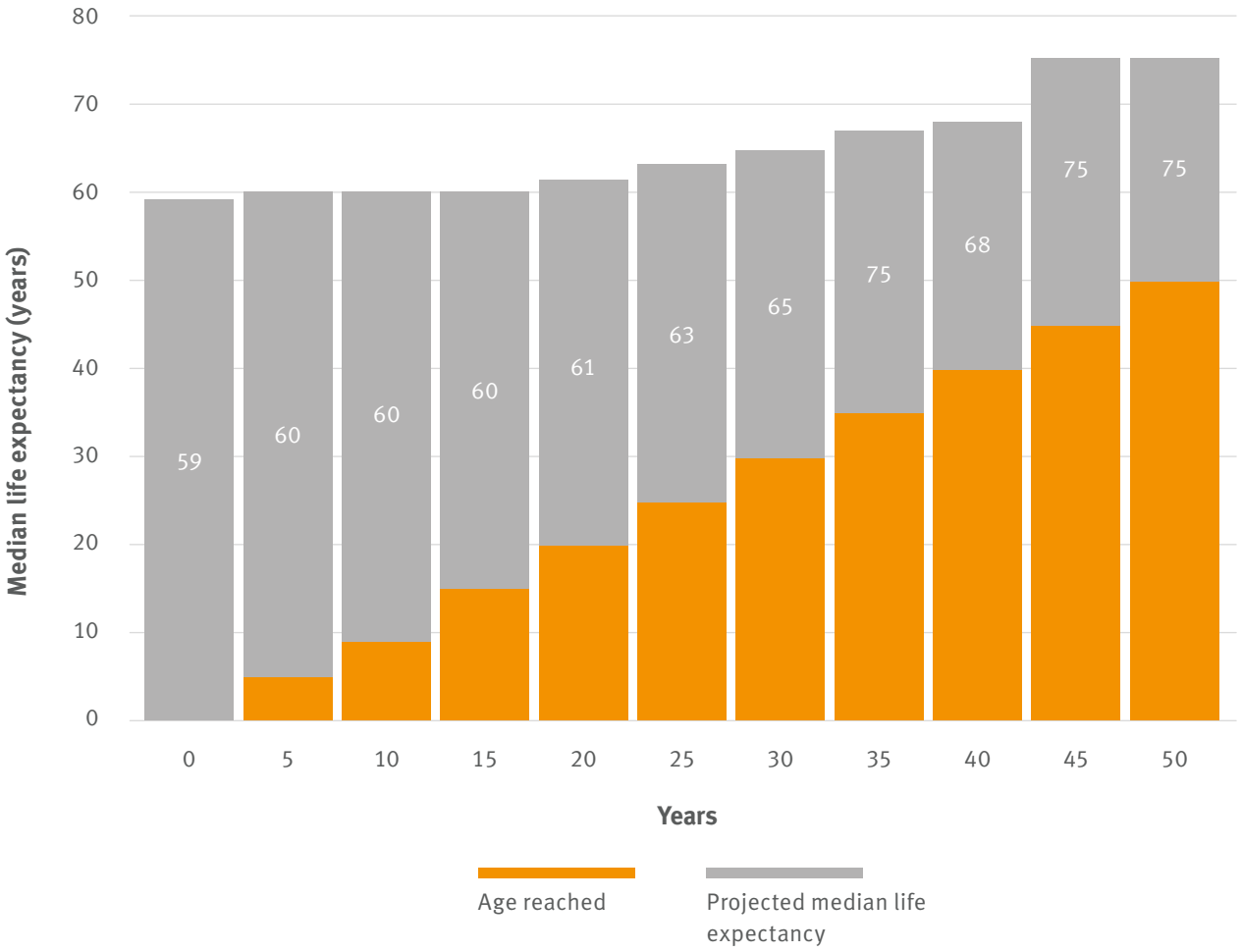

Figure 41: Predicted median life expectancy for pwCF 2018 – 2022

Mortality

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.

Life expectancy 2023

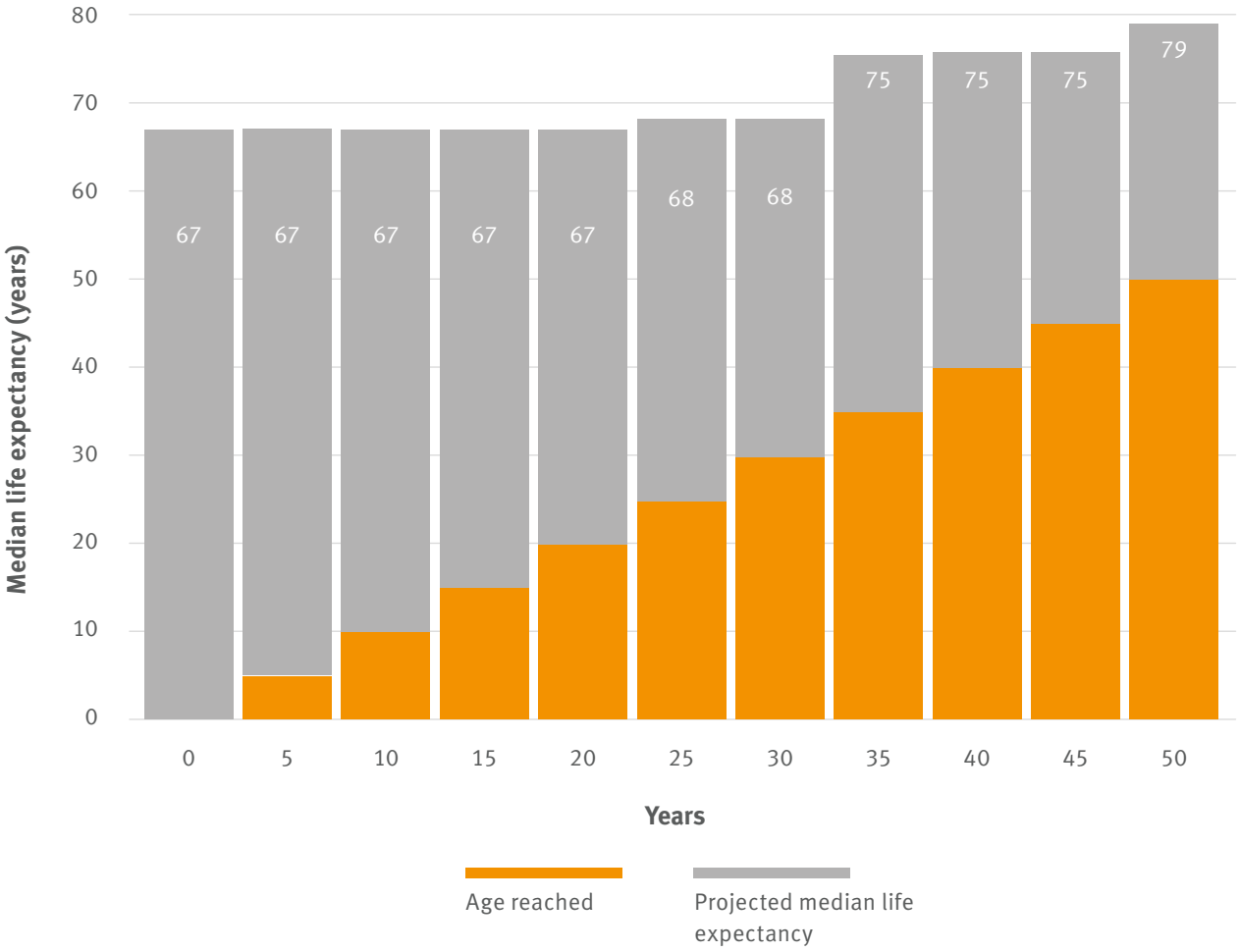

Figure 42: Predicted median life expectancy for pwCF 2019 – 2023

11

Structure of care

11a. Size of the participating CF-centres

In the 2023 reporting year, 85 centres participated in the cystic fibrosis register. 40 centres cared for fewer than 50 patients and 45 centres more than 82 % of the patients documented in the registry.

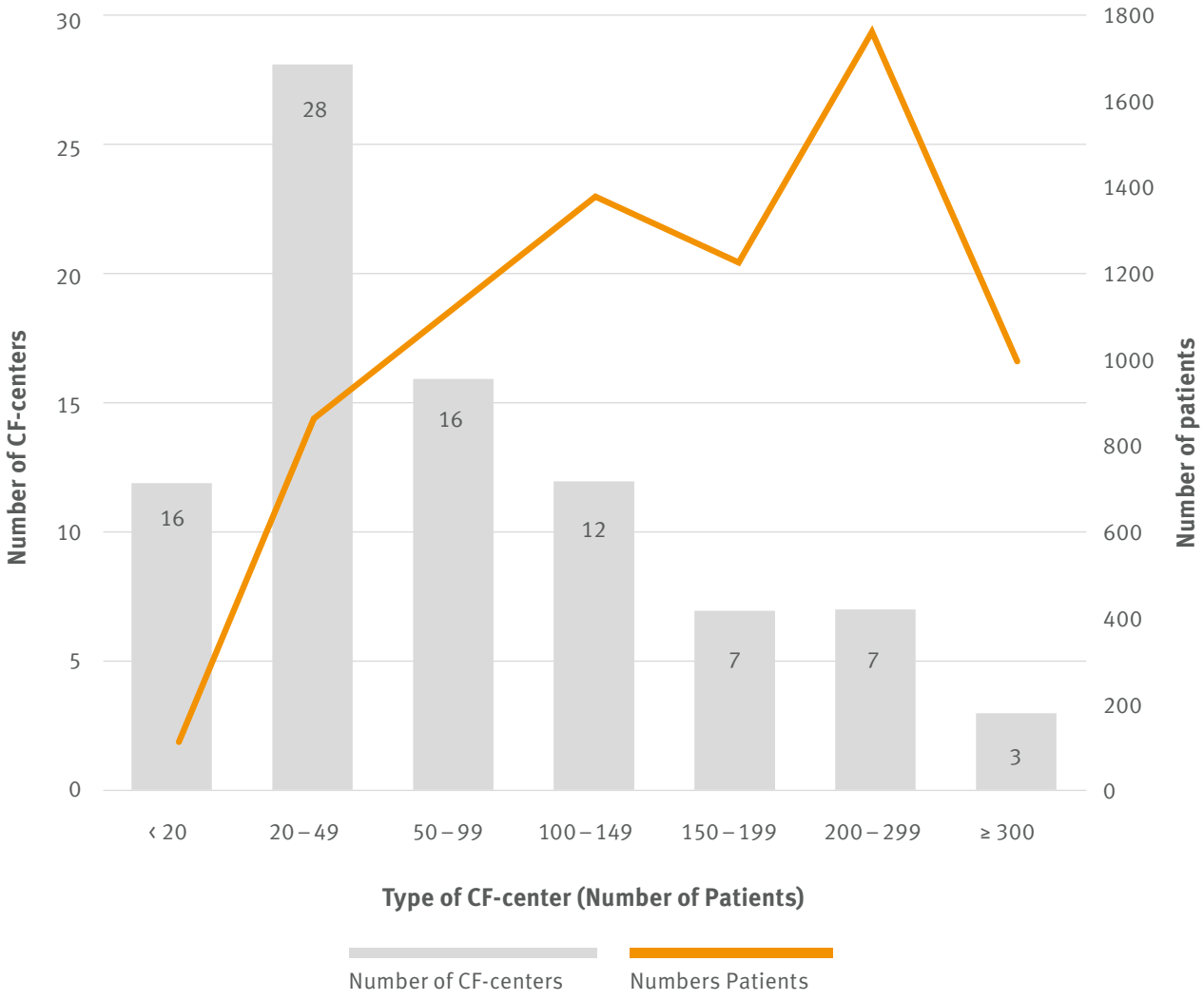

Figure 43: Number of documented pwCF and number of facilities in 2023

Structure of care

11b. Outpatient care

All patients with follow-up data in 2023 (n=7,181) 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*	74.5	72.3	68.8	50.8	51.1	54.2	59.6
Nutritional therapy *in the outpatient clinic*	54.4	47.6	40.8	18.7	12.5	14.0	27.9
Psychosocial support *in the outpatient clinic*	44.2	43.2	41.3	32.4	23.9	27.0	34.0
Anxiety and depression screening[1]	–	–	33.6	47.7	49.5	48.4	45.8
Imaging Thorax	70.5	80.0	68.2	74.5	75.4	72.7	73.6
Bildgebung Abdomen	95.1	80.0	92.4	66.7	64.4	67.8	73.2
Bildgebung Bone density measurement	0.0	0.0	7.6	8.5	16.9	16.1	10.4
Laboratory	90.9	95.2	95.2	96.9	96.5	95.9	95.5
Rehabilitation stay	5.1	6.8	5.9	1.9	3.1	5.7	4.3
oGT test[2] *in patients without diabetes mellitus in the previous year*	0.0	13.6	48.7	35.2	28.6	25.6	30.7

Table 36: pwCF with outpatient care (frequencies in %) 2023
[1] related to cystic fibrosis patients aged 12 and over; 2 related to cystic fibrosis patients aged 6 and over

11

Structure of care

11c. CF-related hospitalizations

Age (years)	Number of CF-relevant hospitalisations per patient						
	0	1	2	3	4	5+	unknown
0 – 5	73.3	20.6	4.6	1.1	0.1	0.2	0.0
6 – 11	81.2	15.5	2.4	0.4	0.2	0.3	0.0
12 – 17	79.5	15.8	2.8	1.1	0.5	0.2	0.1
18 – 29	81.1	12.7	3.7	1.3	0.8	0.2	0.2
30 – 39	83.2	12.0	3.2	1.1	0.3	0.0	0.2
≥ 40	80.6	13.8	3.3	1.0	0.4	0.3	0.5
Total	80.3	14.5	3.3	1.0	0.4	0.2	0.2
< 18	78.2	17.1	3.2	0.9	0.3	0.2	0.0
≥ 18	81.6	12.8	3.4	1.2	0.5	0.2	0.3

Table 37: Number of cystic fibrosis-related hospitalisations per patient (frequencies in %) 2023

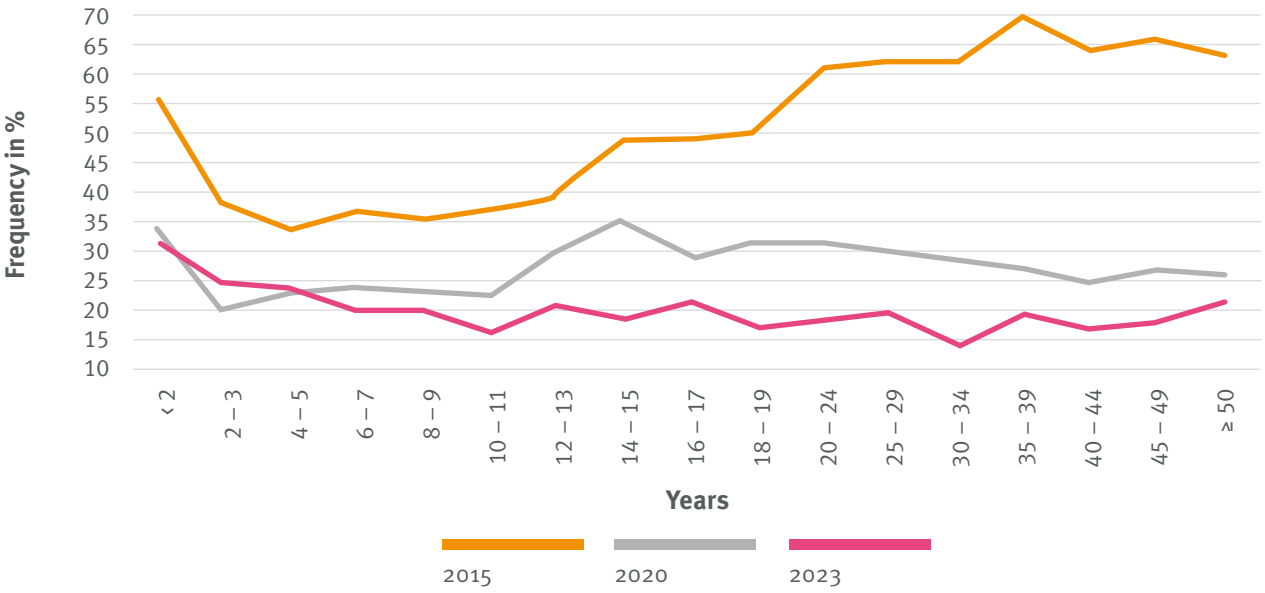

Figure 44: Development of the proportion of pwCF with at least 1 hospitalisation 2015 – 2023

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	55.8	38.0	33.7	36.9	34.9	37.4	40.2	48.7	49.0	50.0	60.9	62.5	62.7	70.0	64.3	65.7	63.6
2020	34.0	20.3	23.0	23.7	23.0	22.5	30.2	34.9	29.3	31.9	31.6	29.8	28.2	26.9	24.8	26.6	26.1
2023	33.3	24.7	24.0	19.6	20.2	16.4	20.7	18.8	21.7	17.1	18.5	19.5	14.1	19.5	16.8	17.7	21.5

Table 38: Development of age-related frequencies (in %) of pwCF with at least 1 hospitalisation 2015 – 2023

Structure of care

11d. Outpatient visits

Years	Outpatient visits in the calendar year			
	1	2	3	≥ 4
2018	7.1	10.7	20.7	61.5
2019	6.1	11.0	23.6	59.4
2020	7.7	14.8	25.1	52.4
2021	6.0	11.6	24.9	57.5
2022	5.9	13.2	30.7	50.1
2023	11.3	14.2	29.8	44.7

Table 39: Development of the number of documented outpatient visits (frequencies in %) 2018 – 2023

Figure 45: Development of the number of documented outpatient visits 2018 – 2023

11

Structure of care

11e. Transplants

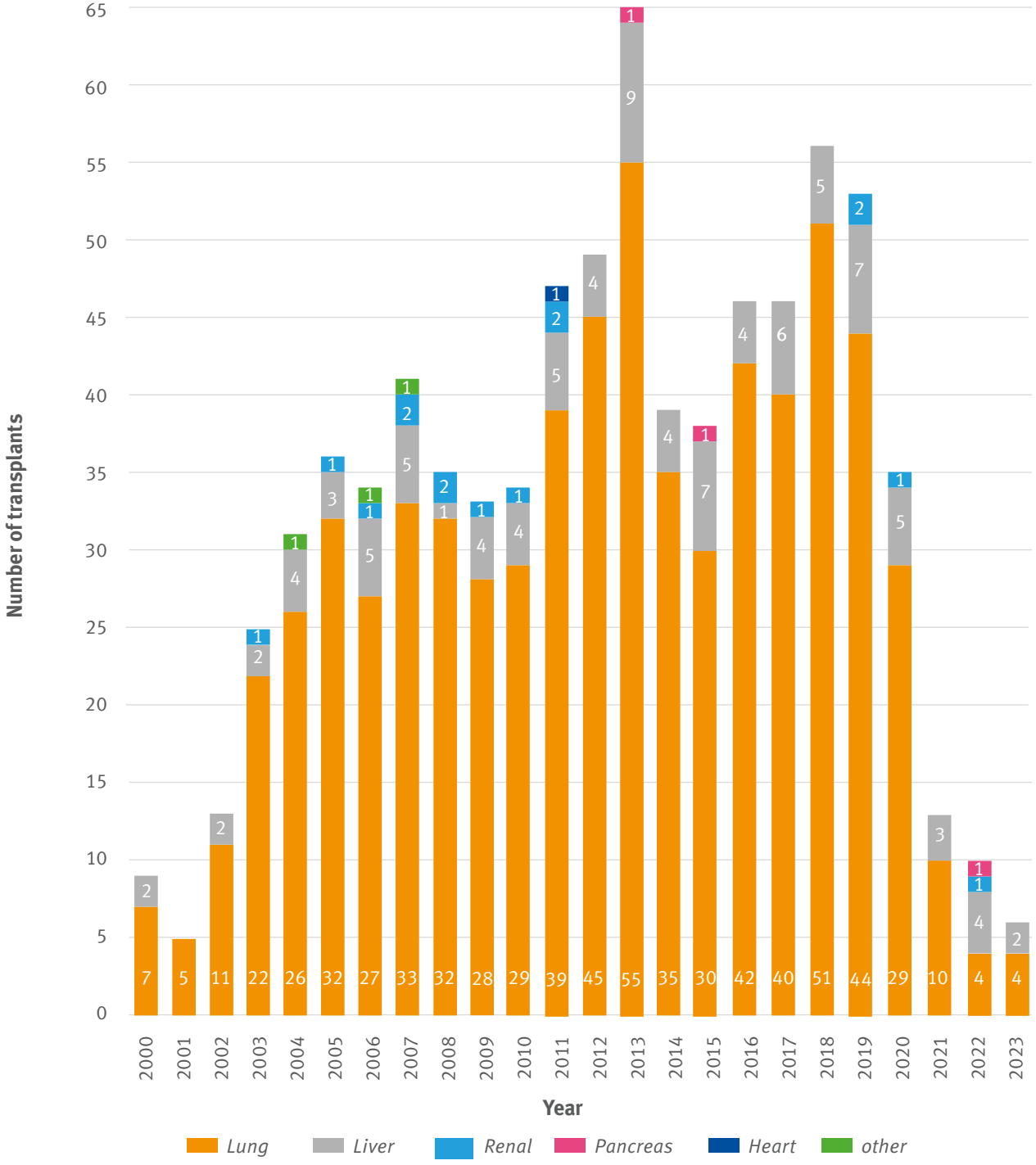

Figure 46: Number of transplants for the years 2000 – 2023

Structure of care

11e. Transplants

Reporting year	Lung	Liver	Renal	Pancreas	Heart	other	unknown
2000	7	2	0	0	0	0	0
2001	5	0	0	0	0	0	0
2002	11	2	0	0	0	0	0
2003	22	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	28	4	1	0	0	0	0
2010	29	4	1	0	0	0	0
2011	39	5	2	0	1	0	0
2012	45	4	0	0	0	0	0
2013	55	9	0	1	0	0	0
2014	35	4	0	0	0	0	0
2015	30	7	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	44	7	2	0	0	0	0
2020	29	5	1	0	0	0	0
2021	10	3	0	0	0	0	0
2022	4	4	1	1	0	0	0
2023	4	2	0	0	0	0	0

Table 40: Number of transplants for the years 2000 – 2023

12

Overview of Registry requests

Receipt	Applicant	Institution	Subject / Title	Status
2017	Dittrich	Universität Heidelberg	Survival-Adjusted FEV1 and BMI Percentiles for Patients with Cystic Fibrosis before the Era of Triple CFTR Modulator Therapy in Germany	Completed – Published
2017	Schwarz	Charité Universitätsmedizin Berlin	Risk factors for respiratory Aspergillus fumigatus in German Cystic Fibrosis patients and impact on lung function	Completed – Published
2017	Prinz	Universität Ulm	Mukoviszidose und Glukosetoleranz	Completed – Published
2017	Grehn	Charité Universitätsmedizin Berlin	Risk factors for cystic fibrosis arthropathy: Data from the German cystic fibrosis registry	Completed – Published
2017	S.p.A Chiesi Farmaceutici	Chiesi	Quinsair PASS	Study in progress
2018	Ballmann	Kinder-und Jugendklinik Universitätsmedizin Rostock	Diabetes Sonderauswertung	Completed
2018	Vertex Pharmaceuticals	Vertex	TEZ/IVA PASS	Completed
2018	Hogardt	Universitätsklinikum Frankfurt	Prävalenz des B. Cepacia-Komplex bei CF-Patienten	Completed – Published (MIQ)
2019	Steindor/ Ringshausen	Universitätsklinik Essen/ Medizinische Hochschule Hannover	Epidemiological trends in nontuberculous mycobacterial infection among people with cystic fibrosis in Germany	Completed – Published
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	Chronic inhaled antibiotic therapy in people with cystic fibrosis with Pseudomonas aeruginosa infection in Germany	Completed – Published
2019	Stanke	Medizinische Hochschule Hannover	Genetische Prädiktoren für schwere CF bei europäischen Zwillingen und Geschwistern	Under evaluation
2019	Hogardt	Universitätsklinikum Frankfurt	Molecular Epidemiology of Mycobacterium abscessus Isolates Recovered from German Cystic Fibrosis Patients	Completed – Published
2020	Vertex Pharmaceuticals (Germany) GmbH	Vertex	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 – Übergewicht bei CF (DMT 2020 Vortrag)	Completed
2020	Vertex Pharmaceuticals (Germany) GmbH	Vertex	Dossier Nutzenbewertung Triple-Therapie – Indikationserweiterung	Completed
2020	Vertex Pharmaceuticals (Germany) GmbH	Vertex	ETI PASS	Study in progress
2021	Vertex Pharmaceuticals (Germany) GmbH	Vertex	Dossier Nutzenbewertung Triple-Therapie	Completed
2022	Dittrich, Tümmler	Medizinische Hochschule Hannover	Use of elexacaftor/tezacaftor/ivacaftor leads to changes in detection frequencies of Staphylococcus aureus and Pseudomonas aeruginosa dependent on age and lung function in people with CF	Completed – Published
2022	Splisense, Israel		Number of CF patients carrying the 3849 +10kb C->T mutation in Germany	Completed

Receipt	Applicant	Institution	Subject / Title	Status
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	Completed
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	Dillenhöfer/ Mukoviszidose Institut	Universitätsklinikum der Ruhr-Universität Bochum/ ETI Publikationsinitiative	Long-Term Impact of Elexacaftor/Tezacaftor/ Ivacaftor on small and large Airways in pwCF aged over 6 years: 24-month real-world evidence from the German CF Registry	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 evaluation
2023	Nährlich/ Brockow	Justus-Liebig-Universität Gießen/DGNS	Vergleich von Diagnosedaten nach der Einführung des Mukoviszidosescreenings bei Neugeborenen in Deutschland	Completed – Published
2024	Sutharsan/ Mukoviszidose Institut gGmbH	Universitätsmedizin Essen/ ETI Publikationsinitiative	Working Title: Impact ETI on BMI & HbA1c	Under evaluation
2024	Vertex Pharmaceuticals (Germany) GmbH	Vertex	AMNOG Nutzenbewertung Vanzacaftor/Tezacaftor/Deutivacaftor	Completed
2024	Nährlich/Burkhart/ Kurch-Beck	Justus-Liebig-Universität Gießen/ Mukoviszidose Institut/KBV	The prevalence of cystic fibrosis – a comparison of patient registry data and billing data within the German statutory health insurance system, 2023	Completed – Published

You can also find all published registry requests at:
www.muko.info/was-wir-tun/register/publikationen/publizierte-registeranfragen

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Participating CF centers 2023

City	CF center	Department	Number of patients[1]
Aachen	Kinderarztpraxis Laurensberg	Aachener Mukoviszidose Ambulanz für Kinder und Jugendliche	45
Aachen	Luisenhospital Aachen	Mukoviszidose-Zentrum für Erwachsene, Innere Medizin	91
Aue	HELIOS Klinikum Aue - CF-Ambulanz	Klinik für Kinder- und Jugendmedizin	10
Augsburg	Universitätsklinikum Augsburg AöR	Pneumologie	1
Augsburg	KJF Klinik Josefinum	Klinik für Kinder- und Jugendmedizin Sozialpädagogisches Zentrum (SPZ) Mukoviszidose-Ambulanz	20
Augsburg	Universitätsklinikum Augsburg	I. Klinik für Kinder und Jugendliche, Kinderpneumologie - Allergologie, Mukoviszidose Ambulanz	23
Baden Baden	Klinikum Mittelbaden gGmbH	Baden-Baden Balg, Lungenzentrum, Mukoviszidose Ambulanz	6
Berlin	Sana Klinikum Lichtenberg	Oskar-Ziethen-Krankenhaus, Klinik für Kinder- und Jugendmedizin, Pneumologie, Mukoviszidose-Zentrum, Allergologie	63
Berlin	Charité	Christiane Herzog-Zentrum Berlin, Klinik für Pädiatrie m. S. Pädiatrische Pneumologie und Immunologie	296
Bielefeld	Evangelisches Klinikum Bethel gGmbH	Lehrkrankenhaus der Universität Münster, Klinik für Kinder- und Jugendmedizin, Tagesklinik für Allergologie und Pneumologie	25
Bochum	Universitätsklinikum der Ruhr-Universität Bochum	St. Josef-Hospital am Katholischen Klinikum Bochum, Klinik für Kinder- und Jugendmedizin, Christiane Herzog Zentrum Ruhr (CHCR)	79
Brandenburg	Medizinische Hochschule Brandenburg (MHB) Klinikum West-Brandenburg	Kinder- und Jugendklinik, CF-Ambulanz	34
Bremen	Klinikum Bremen Mitte	Eltern-Kind-Zentrum Prof. Hess, Christiane Herzog Ambulanz für MukoviszidoseEltern-Kind-Zentrum Prof. Hess, Christiane Herzog	88
Chemnitz	Poliklinik Chemnitz gGmbH	Praxis für Kinder- und Jugendmedizin	40
Cottbus	Carl-Thiem-Klinikum Cottbus gGmbH	Interdisziplinäre Studienzentrale	25
Dresden	Medizinische Fakultät der TU Dresden	Klinik und Poliklinik für Kinder- und Jugendmedizin, Mukoviszidose-Centrum "Christiane Herzog"	208
Düsseldorf	UKD Universitätsklinikum Düsseldorf	Klink für Allgemeine Pädiatrie, Neonatologie und Kinderkardiologie, Ambulanz für Kinderpneumologie und Allergologie	32
Erfurt	HELIOS Klinikum Erfurt	Kinderklinik und Jugendmedizin, CF-Ambulanz	21
Erlangen	Universitätsklinikum Erlangen	Kinder- und Jugendklinik, Sozialpädiatrisches Zentrum	175
Essen	Universitätsklinikum Essen	Zentrum für Kinder- und Jugendmedizin, Pädiatrische Pneumologie und Schlafmedizin, Christiane Herzog Centrum Ruhr	101
Essen	Universitätsmedizin Essen	Ruhrlandklinik – Pneumologie	330
Frankfurt	St. Elisabethen Krankenhaus	Katharina-Kaspar Kliniken, Innere Medizin – Pneumologie	22
Frankfurt	Universitätsklinikum Frankfurt	Goethe Universität, Christiane Herzog CF-Zentrum für Kinder, Jugendliche und Erwachsene	263
Freiburg	Universitätsklinikum Freiburg	Abteilung Pneumologie, Erwachsenenambulanz	45
Freiburg	Universitätsklinikum Freiburg	Zentrum für Kinder- und Jugendmedizin – Mukoviszidose-Ambulanz	79
Gießen	Universitätsklinik Gießen und Marburg GmbH	Zentrum für Kinder- und Jugendmedizin, Mukoviszidose-Zentrum	100
Gießen	Universitätsklinik Gießen und Marburg GmbH	Zentrum für Kinder- und Jugendmedizin, Mukoviszidose-Zentrum	100
Greifswald	Universitätsmedizin Greifswald	Klinik und Poliklinik für Kinder- und Jugendmedizin	23
Halle	Universitätsklinikum Halle (Saale) (UKH)	Medizinische Fakultät der Martin-Luther-Universität, Mukoviszidose-Zentrum	80
Hamburg	Universitätsklinikum Eppendorf	II. Medizinische Klinik - Sektion Pneumologie	98

[1] patients may have been documented in several outpatient clinics

Participating CF centers 2023

City	CF center	Department	Number of patients[1]
Hamburg	Kinder- und Jugendärztliche Gemeinschaftspraxis	Kinderärzte im Friesenweg, CF Centrum Altona	123
Hamm	Evangelisches Krankenhaus Hamm (EVK) gGmbH	Klinik für Kinder- und Jugendmedizin, Pulmologie/Allergologie	12
Hannover	Medizinische Hochschule Hannover	Klinik für Pädiatrische Pneumologie, Allergologie und Neonatologie	178
Hannover	Medizinische Hochschule Hannover	Klinik für Innere Medizin, Pneumologische Ambulanz (Erwachsene)	259
Heidelberg	Universitätsklinikum Heidelberg	Sektion Pädiatrische Pneumologie, Allergologie und Mukoviszidose-Zentrum	132
Heidelberg	Thoraxklinik am Universitätsklinikum Heidelberg	Abteilung für Pneumologie und Beatmungstherapie, CF Ambulanz für Erwachsene	253
Heilbronn	SLK-Kliniken Heilbronn GmbH	Klinik für Kinder- und Jugendmedizin, Klinikum am Gesundbrunnen, Perinatalzentrum	14
Homburg	Universitätsklinikum des Saarlandes	Klinik für Allgemeine Pädiatrie und Neonatologie – Mukoviszidose-Ambulanz	58
Homburg	Universitätsklinikum des Saarlandes	Innere Medizin 5, CF-Ambulanz für Erwachsene	58
Jena	Universitätsklinikum Jena	Klinik für Kinder- und Jugendmedizin, Ambulanz für Pädiatrische Pneumologie, Allergologie, Mukoviszidosezentrum	173
Karlsruhe	Städtisches Klinikum Karlsruhe gGmbH	Klinik für Kinder- und Jugendmedizin	25
Kassel	Klinikum Kassel	Pädiatrische Hämatologie und Onkologie, Psychosomatik und Systemerkrankungen Mukoviszidose-Ambulanz	44
Kiel	Städtisches Krankenhaus Kiel GmbH	Klinik für Kinder- und Jugendmedizin, Mukoviszidose-Zentrum	51
Kiel	Städtisches Krankenhaus Kiel GmbH	Mukoviszidose-Zentrum für Erwachsene, 4. Medizinische Klinik	114
Koblenz	Gemeinschaftsklinikum Mittelrhein gGmbH	Klinik für Kinder- und Jugendmedizin, Pädiatrische Pneumologie und Allergologie, Mukoviszidose Ambulanz	43
Köln	Kliniken der Stadt Köln	Lungenklinik Merheim	84
Köln	Universitätsklinikum Köln	Klinik und Poliklinik für Kinder- und Jugendmedizin, Mukoviszidose-Zentrum	244
Krefeld	Helios Klinikum Krefeld	Zentrum für Kinder- und Jugendmedizin, Mukoviszidose-Ambulanz	37
Leipzig	Universitätsklinikum Leipzig	Klinik und Poliklinik für Kinder- und Jugendmedizin, CF-Ambulanz	71
Löwenstein	Fachklinik Löwenstein	Klinik für Pneumologie, Intensiv- und Beatmungsmedizin	32
Lübeck	Universitätsklinkum Schleswig Holstein (UKSH)	Campus Lübeck, Klinik für Kinder- und Jugendmedizin, Pädiatrische Pneumologie	33
Magdeburg	Otto-von-Guericke Universität Magdeburg	Universitätsklinik für Pneumologie	15
Magdeburg	Otto-von-Guericke Universität Magdeburg	Klinik für Allgemeinpädiatrie und Neonatologie, CF-Ambulanz	19
Mainz	Universitätsmedizin Mainz	Zentrum für Kinder- und Jugendmedizin Klinik und Poliklinik für Kinder- und Jugendmedizin	109
Mannheim	Universitätsmedizin Mannheim	Klinik für Kinder- und Jugendmedizin, Pulmologie, Infektiologie und Allergologie	6
Marburg	Zentrum für Kinderheilkunde	Mukoviszidose-Ambulanz	14
Memmingen	Klinikum Memmingen	Klinik für Kinder- und Jugendmedizin, Sozialpädiatrisches Zentrum	11
München	Kinderpoliklinik Schwabing	CF-Ambulanz	31
München	Lungenheilkunde München Pasing	Mukoviszidose-Zentrum München West	182
München	LMU Klinikum der Universität München	Campus Innenstadt, Medizinische Klinik – Pneumologie	230

[1] patients may have been documented in several outpatient clinics

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Participating CF centers 2023

City	CF center	Department	Number of patients[1]
München	LMU Klinikum der Universität München	Kinderklinik und Kinderpoliklinik im Dr. von Haunerschen Kinderspital, Christiane Herzog-Ambulanz	314
Münster	Universitätsklinikum Münster UKM	Klinik für Kinder- und Jugendmedizin, Allgemeine Pädiatrie Mukoviszidose-Ambulanz	63
Münster	Clemenshospital	Mukoviszidose-Ambulanz	125
Neubranden-burg	Dietrich Bonhoeffer Klinikum	Klinik für Kinder- u. Jugendmedizin	22
Oldenburg	Klinikum Oldenburg AöR	Klinik für Pädiatrische Pneumologie und Allergologie, Neonatologie und Intensivmedizin	107
Osnabrück	Christliches Kinderhospital Osnabrück	Zentrum für Kinder- und Jugendmedizin, Mukoviszidose Ambulanz	67
Passau	Ordenskliniken München-Passau gGmbH	Standort Kinderklinik Dritter Orden Passau	26
Potsdam	Klinikum Westbrandenburg gGmbH	Kinder- und Jugendklinik, Mukoviszidose Ambulanz	245
Ravensburg	Oberschwabenklinik (OSK) gGmbH Ravensburg	Krankenhaus St. Elisabeth, Klinik für Kinder und Jugendliche	2
Regensburg	Klinik Donaustauf	Pneumologische Ambulanz	52
Regensburg	KUNO Klinik St. Hedwig	Kinder- und Jugendmedizin	75
Rostock	Universitätsmedizin Rostock	Kinder- und Jugendmedizin	26
Rüdersdorf bei Berlin	Immanuel Klinik Rüdersdorf	Kinder- und Jugendmedizin	5
Schwerin	HELIOS Kliniken Schwerin	Kinder- und Jugendmedizin, Mukoviszidose-Ambulanz	25
Stuttgart	Klinikum Stuttgart	Christiane Herzog Transitionszentrum	151
Stuttgart	Robert Bosch Krankenhaus RBK	Pneumologie und Beatmungsmedizin, Mukoviszidose-Ambulanz	167
Trier	Klinikum Mutterhaus der Borromäerinnen gGmbH	Kinder-u. Jugendmedizin	18
Trier	Klinikum Mutterhaus der Borromäerinnen gGmbH	Klinikum Mutterhaus Mitte, Innere Medizin 1	23
Tübingen	Universitätsklinik Tübingen	Klinik für Kinder- und Jugendmedizin, Mukoviszidose-Ambulanz	93
Ulm	Universitätsklinikum Ulm	Klinik für Kinder- und Jugendmedizin, Mukoviszidose-Ambulanz	107
Wangen	Fachkliniken Wangen gGmbH	Rehabilitationsklinik für Kinder und Jugendliche, CF-Ambulanz	8
Wangen	Fachkliniken Wangen	Waldburg Zeil Kliniken, Klinik für Pneumologie	27
Wesel	Marienhospital Wesel	Akademisches Lehrkrankenhaus der Westfälischen Wilhelms-Universität Münster, Klinik für Kinder- und Jugendmedizin	35
Worms	Klinikum Worms gGmbH	Klinik für Kinder- und Jugendmedizin	51
Würzburg	Universitätsklinikum Würzburg	Kinderpoliklinik Christiane Herzog-Zentrum Unterfranken Mukoviszidose-Ambulanz	161

[1] patients may have been documented in several outpatient clinics

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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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