Cystic Fibrosis Trust

UK Cystic Fibrosis Registry
2023 Annual Data Report

October 2024

Uniting for a life unlimited

UK Cystic Fibrosis Registry
2023 Annual Data Report

An at-a-glance version of this report can be found at cysticfibrosis.org.uk/registry

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The UK CF Registry Steering Committee

Acknowledgements

First and foremost, the UK Cystic Fibrosis Registry team would like to thank people with cystic fibrosis and their families for their support, as well as anyone who has generously donated to Cystic Fibrosis Trust. We would also like to express our gratitude to the UK cystic fibrosis centres and clinics for their continued dedication to obtaining consent and submitting data to the Registry.

Contact information

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Cystic Fibrosis Trust

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Foreword

In the year we're marking 60 years of Cystic Fibrosis Trust, I am really pleased to share with you the 2023 UK CF Registry Annual Report. First established in 1995, the data collected within the UK CF Registry has provided valuable insights into the health of people with CF and how this has changed over time. Twenty years ago in 2003, 6,861 people with CF were registered on the then UK CF Database. In 2023, our UK CF Registry Report is based on the experiences of 11,318 people. I would like to thank everyone with CF and their families who consent to sharing their data, along with the clinical care teams who collect and submit it. This ongoing support ensures the Registry can continue to document, inform and make change for people with CF across the UK.

Cystic Fibrosis Trust took over the sponsorship and management of the database in 2007 and I wanted to take this opportunity to reflect on some key UK CF Registry milestones. In 2012, the Registry started the production of reports used by the NHS England to make payment by results (PbR) tariff payments to CF centres. The Registry's pharmacovigilance programme was established in the same year, and in 2023, three studies reviewing the safety of medicines were concluded.

In 2016, the first randomised Registry-based trial, CF START began, with CF STORM following in 2021, and earlier this year, the UK CF Registry received its 500th data request - demonstrating the significance of the Registry to researchers, clinicians and many others. More recently, Registry data has contributed to the NICE appraisal of the CFTR modulator therapies, leading to the life changing decision to recommend long-term access to these drugs. But not everyone can benefit from these drugs, and they are not a cure. We are presenting new data on modulator use, including demographic information data on those not taking modulator therapies, in this report. We continue to support research into new therapies for those unable to benefit from modulators and our Clinical Trials Accelerator Platform is supporting several trials. You can find out more about opportunities to participate in research here: **cysticfibrosis.org.uk/research/clinical-trials**

The number of people living with CF has increased over the last 20 years and we are seeing people live longer and healthier lives. In 2003, the median age of those with CF was 16.1 years old and 50.8% were adults aged 16 or over. In 2023, we report a median age of people with CF of 22 years and 63.7% aged 16 or over. FEV_1% predicted lung function measures are improving across all age groups (p.27) and chronic lung infections are decreasing (p.31). In 2023, our 60th year, we report for the first time, the median predicted survival for babies born today increasing to over 60 years of age to 64.1 years, with the gap between men and women almost closed (p.50).

In noting this progress, it's important to highlight that people with CF still face many challenges. We report increases since 2018 in some CF complications, such as raised liver enzymes and liver disease. Over 33% of adults with CF are also living with CF diabetes, a number that hasn't changed over the last five years. And over 80% of people with CF are taking pancreatic enzyme supplements like Creon, with some people with CF and their families experiencing medium-term interruptions in supply since late 2023, causing worry and stress for those affected. The data within the 2023 Registry Annual Report reflects the overall CF population health; it doesn't tell us the individual stories of each person living with CF.

As we reflect on the last 60 years of Cystic Fibrosis Trust, we also look towards the next 60 years. For those in our community living longer, healthier lives, every journey experienced will be different, and the UK CF Registry will need to adapt to monitor new and emerging health outcomes alongside the existing measures reported within this report.

I hope you enjoy reading the report and find the insights presented useful. We would love to hear your feedback or thoughts on future reports. You can contact us by emailing **registry@cysticfibrosis.org.uk** or via social media if you have any comments or questions.

David Ramsden
Chief Executive of Cystic Fibrosis Trust

Executive summary

The 2023 Registry data is a hugely rich information source which the UK CF community should be proud of, celebrate and use to drive forward improvements in CF care in the ever changing landscape of cystic fibrosis. I would like to highlight some aspects of this year's report.

- 11,318 people with CF are registered within the UK CF Registry of whom 93% had an annual review this year (Section 1.1)
- 63.7% of the UK population living with CF are over 16 years of age (Section 1.1) and 16.7% are over 40 years of age (Section 1.2). The changing age distribution of the UK CF population over the past ten years is illustrated in Section 1.3
- Half of people born today with CF in the UK are predicted to live at least 64.1 years (Section 1.44) which has increased from previous years and the gap between predicted survival of men and women with CF has narrowed
- 94.5% of the UK CF population describe their ethnicity as white (Section 1.4)
- 67.7% of people with CF over 16 years of age are in work or studying (Section 1.9)
- The median best FEV_1% continues to rise and is now 89.1% (section 1.14)
- 17.5% of people with CF are using exercise as their primary airway clearance technique (Section 1.37)
- There were fewer than five lung transplants for people with CF in the UK last year (Section 1.41)
- Nutritional status of the CF population is changing (section 1.8) with a smaller proportion now being underweight but an increasing proportion of adults with a BMI $\geq$ 25. The use of oral supplements has fallen to 18.1% of the UK CF population (Section 1.42)
- 28.6% of people with CF over 10 years of age are on CF diabetes therapies (Section 1.23)
- Depression is reported in 6.9% of people with CF over 16 years of age (Section 1.21)
- 116 women with CF had babies in 2023 and 31 men with CF became fathers (Section 1.10)
- 92.7% of people with CF had at least one respiratory culture sent this year (Section 1.19) but sputum samples made up a smaller proportion of the sample type
- The percentage of people receiving at least one course of IV antibiotics (22%) has dropped again this year (Section 1.24)
- The proportion of people with CF that remain on the combination of inhaled antibiotics, DNase and hypertonic saline or mannitol has dropped from 22% to 14.9% (Section 1.33). 24.6% of people with CF are on none of these inhaled therapies as compared to 19.9% last year
- 8,212 people with CF were reported as being on a CFTR modulator by December 2023 (section 1.34) with new tables in Sections 1.35b and 1.36 illustrating modulator use by genotype group and demographics of those eligible and ineligible for current modulator therapies respectively.

Sections 2 is the centre level reports which centres may find helpful when analysing their pattern of home compared to hospital IV antibiotics and types of mucolytics used. Tables of outcome data for centres must be interpreted with caution as some centres are not large enough to allow meaningful comparisons.

We hope the Registry data continues to be valuable to the whole CF community and would like to express our gratitude to people with CF for consenting to have their clinical data recorded and the clinical teams for collecting and entering it into the Registry.

Jamie Duckers
Chair of the UK CF Registry Steering Committee

Introduction

This report is aimed at anyone who is interested in the health, care, and outcomes of people with cystic fibrosis (CF) in the UK. This includes people with CF, their families and clinical teams, healthcare managers, commissioners, and policy makers.

You can find a Glossary of scientific and clinical terms on page 70.

An at-a-glance version of this report can be found at **cysticfibrosis.org.uk/registry**.

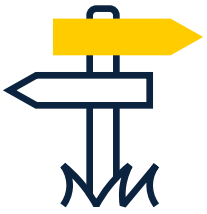
Cystic fibrosis

Cystic fibrosis is an inherited disease caused by a faulty version of a gene known as 'CFTR'. The gene and the protein it makes help control the movement of salt and water in and out of cells. When the gene, and the protein it makes, is faulty, it can cause thicker mucus. One of the main areas affected is the lungs; over time this thick mucus blocks and damages airways, leading to infections and making it hard to breathe. People with CF may also develop other problems, such as liver disease or CF diabetes (CFD). Around 80% of people with CF also have difficulty digesting food.

UK Cystic Fibrosis Registry

The UK CF Registry has been sponsored and hosted by Cystic Fibrosis Trust since 2007. It is a database of consenting people with CF in the UK. The Registry collects demographic, treatment and health outcomes data. You can find a full list of the data items we collect at **cysticfibrosis.org.uk/registry**.

The purpose of the UK CF Registry is to improve the health of people with cystic fibrosis. This is done in a number of ways:



helping people with CF and their families understand CF, and make informed decisions

giving clinical teams the evidence they need to improve the quality of care

monitoring the safety and effectiveness of new treatments for cystic fibrosis

providing data for research to find out the best ways to treat cystic fibrosis

helping commissioners provide funding to NHS CF centres that is proportionate to the severity of their patients' condition

supporting clinical trials through feasibility studies and pragmatic data collection

Governance

The Registry Steering Committee (RSC) is responsible for making sure that the UK CF Registry is compliant with data protection legislation, and its Research Ethics Committee-approved Study Protocol. It also makes recommendations about the future development of the Registry. A sub-committee of the RSC, the Registry Research Committee, assesses applications for data and guides the Registry research strategy.

Please see Appendix 1: UK CF Registry Committee Structure.

Data are only recorded on the UK CF Registry if explicit consent is given by the person with CF, or, if they're a child, their parent or guardian.

When data are provided to third parties, such as the NHS or university researchers, they are either anonymised (all identifiable data removed completely) or pseudonymised (all identifiable data replaced with a unique identification number). Pseudonymisation is used so that data can be traced back to what is in the 'live' database by the Registry team for the purposes of updating the data or answering queries. This means that the Registry data used for research, and the results that come from it, cannot identify the people whose data are stored on the UK CF Registry.

If requests from pharmaceutical companies are granted, for research, or submissions to regulators or the NHS, the data are analysed and aggregated by Registry statisticians and only summary data are provided.

Data collection

Data are entered onto the UK CF Registry by NHS employees at CF centres in the UK using a secure web portal.

Where can I find more information?

You can find out more about CF, and the UK CF Registry, at **cysticfibrosis.org.uk/registry**.

Section 1: UK-wide analysis

This section provides an overview of the cystic fibrosis (CF) population, health outcomes, and care in the United Kingdom, including CF centres in England, Northern Ireland, Scotland, and Wales.

1.1 Summary of the UK Cystic Fibrosis Registry

	2018	2019	2020	2021	2022	2023
CF patients registered[1]	10509	10655	10837	10908	11148	11318
Excluding diagnoses that year	10287	10462	10632	10720	10925	11146
CF patients with an annual review; n(%)[2]	9847 (96)	10070 (96)	9922 (93*)	10175 (95)	10251 (94)	10344 (93)
Age in years; median[3]	20	21	21	21	22	22
All newly diagnosed patients (NBS and other)[4]	222	193	205	188	223	173
All newly diagnosed patients (amended)[5]	(305)	(283)	(264)	(258)	(268)	(TBD)
Number of patients born identified by NBS[4]	167	150	152	134	162	124
Number of patients born identified by NBS (amended)[5]	(179)	(172)	(177)	(167)	(180)	(TBD)
Age at diagnosis in months; median[3]	2.0	2.0	1.8	1.6	1.5	1.3
Adults aged 16 years and over; %[3]	60.4	60.6	60.6	61.9	62.9	63.7
Males; %[3]	53.0	53.2	53.1	53.2	53.1	53.3
Genotyped; %[3]	99.1	99.2	99.2	99.1	99.5	99.4
Total deaths reported during annual review year (%)[6]	137 (1.3%)	114 (1.1%)	97 (0.9%)	66 (0.6%)	64 (0.6%)	49 (0.4%)
Total deaths reported amended (%)	143	119	102	69	72	(TBD)
Age at death in years; median (95% CI)[6]	32 (29, 35)	31 (29, 34)	36 (32, 38)	39 (36, 42)	33 (31, 39)	46 (37, 55)

Annual review: A Registry annual review form records a combination of data relating to a person with CF's once-yearly annual review appointment at their CF centre and their clinical care and health over the past 12 months.

Notes:

* Corrected from 2020 report.

[1] Number of patients diagnosed with CF, seen in the last two years, and alive at 1 January in the given year. Figure has been amended since the publication of the Highlights report 2023.

[2] Newly diagnosed patients in a given year may not have their first annual review in the same year, so the proportion with an annual review is calculated from the total registered excluding those diagnosed in the given year.

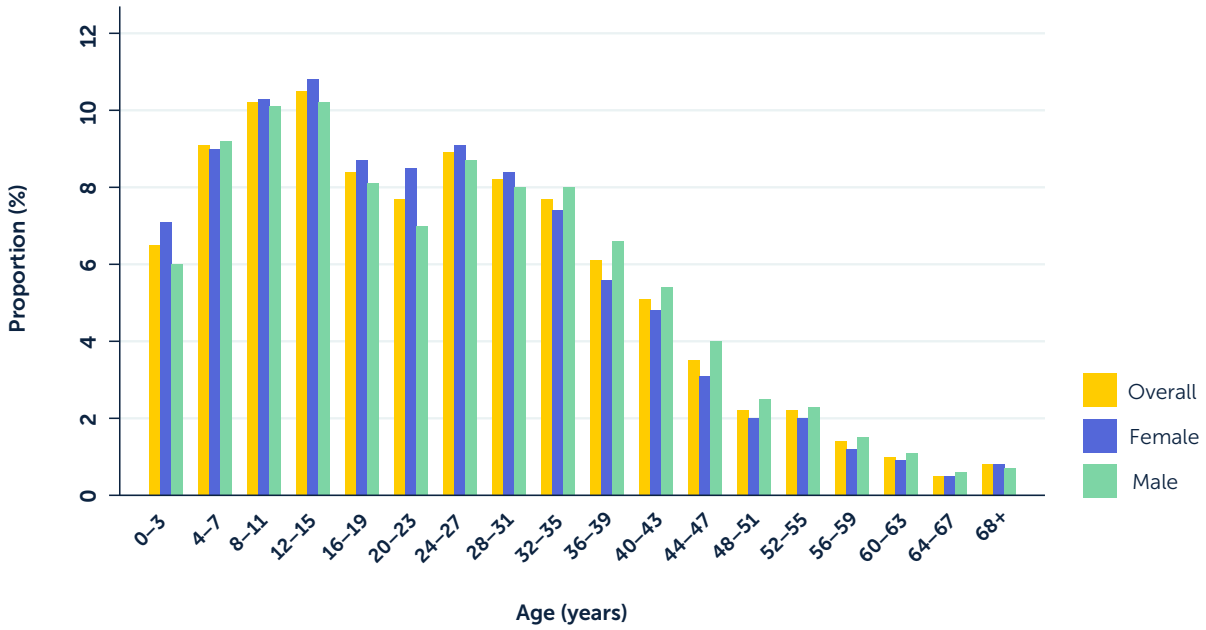
[3] Calculated from patients with an annual review in the given year (see footnote 5 below).

[4] Calculated from all patients registered on the database. Some diagnosis data are added after the data entry closure each year, so figures are updated the following year (see below).

[5] Amended values refer to new diagnoses, identification by NBS or deaths that occurred within the given year but were not recorded on the Registry until after data collection closure. We first presented the amended figures in the 2019 data report. In this report we have completed an additional data cleaning exercise and so some earlier figures have also been updated. We have also added in amended figures for those born identified by NBS.

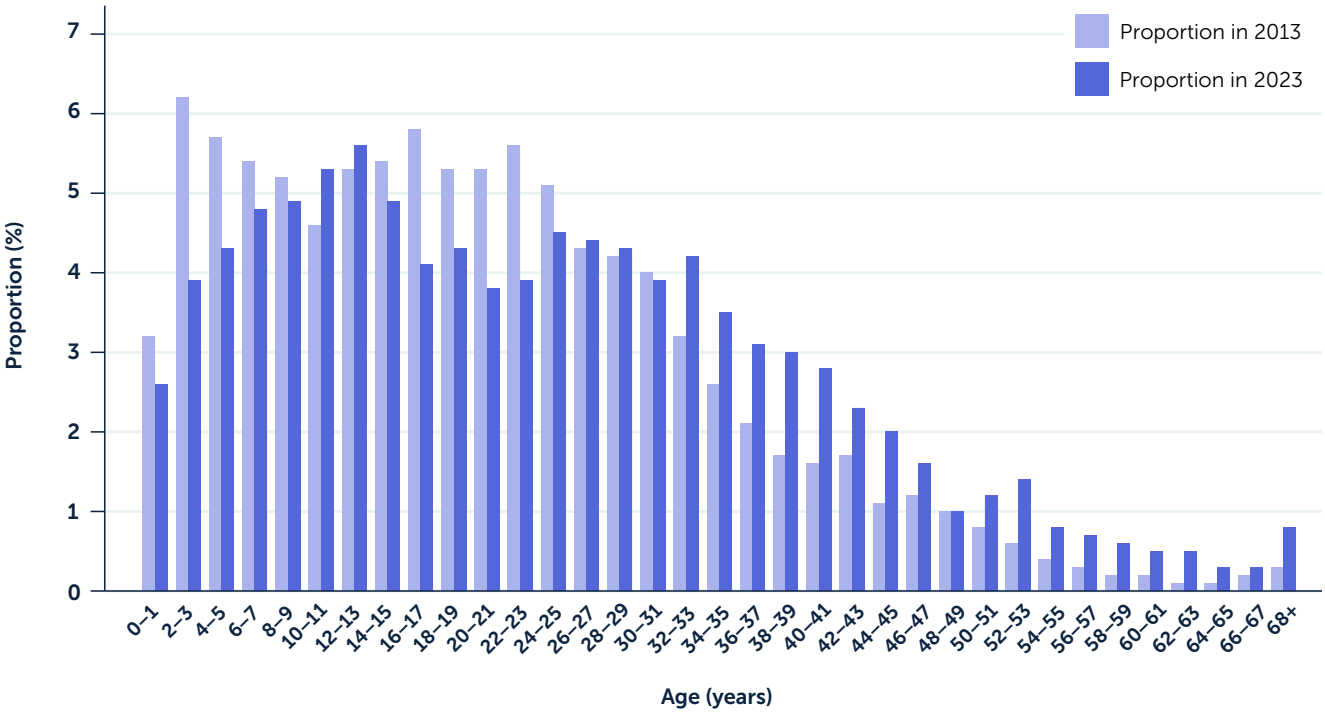
[6] Calculated from all registered patients who died in the given year.

1.2 Age distribution by sex
N=10344



Age	All; n (%)	Females; n (%)	Males; n (%)
0-3	676 (6.5)	343 (7.1)	333 (6.0)
4-7	942 (9.1)	434 (9.0)	508 (9.2)
8-11	1053 (10.2)	498 (10.3)	555 (10.1)
12-15	1085 (10.5)	521 (10.8)	564 (10.2)
16-19	866 (8.4)	419 (8.7)	447 (8.1)
20-23	796 (7.7)	412 (8.5)	384 (7.0)
24-27	918 (8.9)	439 (9.1)	479 (8.7)
28-31	848 (8.2)	405 (8.4)	443 (8.0)
32-35	800 (7.7)	360 (7.4)	440 (8.0)
36-39	636 (6.1)	273 (5.6)	363 (6.6)
40-43	527 (5.1)	230 (4.8)	297 (5.4)
44-47	367 (3.5)	148 (3.1)	219 (4.0)
48-51	230 (2.2)	95 (2.0)	135 (2.5)
52-55	225 (2.2)	97 (2.0)	128 (2.3)
56-59	140 (1.4)	56 (1.2)	84 (1.5)
60-63	103 (1.0)	45 (0.9)	58 (1.1)
64-67	54 (0.5)	22 (0.5)	32 (0.6)
68+	78 (0.8)	39 (0.8)	39 (0.7)
<16	3756 (36.3)	1796 (37.1)	1960 (35.6)
≥16	6588 (63.7)	3040 (62.9)	3548 (64.4)
<18	4181 (40.4)	2003 (41.4)	2178 (39.5)
≥18	6163 (59.6)	2833 (58.6)	3330 (60.5)
Overall	**10344**	**4836**	**5508**

1.3 Age distribution of the UK CF population in 2013 vs 2023
N=10344 in 2023, N=9052 in 2013



1.4 Ethnicity

Ethnicity n (%)	2013	2018	2023
Total	9052	9847	10344
Total known[1]	9003	9691	10192
White	**8575 (95.2)**	**9244 (95.4)**	**9632 (94.5)**
Asian	**231 (2.6)**	**277 (2.9)**	**327 (3.2)**
Bangladeshi	31 (0.3)	36 (0.4)	44 (0.4)
Indian	29 (0.3)	42 (0.4)	57 (0.6)
Pakistani	146 (1.6)	169 (1.7)	189 (1.9)
Other (Asian)	25 (0.3)	30 (0.3)	37 (0.4)
Black	**-***	**26 (0.3)**	**-***
Black African	10 (0.1)	11 (0.1)	15 (0.1)
Black Caribbean	13 (0.1)	10 (0.1)	13 (0.1)
Other (Black)	<5	5 (0.1)	<5
Mixed**	**77 (0.9)**	**57 (0.6)**	**113 (1.1)**
Mixed (White-Asian)	-	14 (0.1)	31 (0.3)
Mixed (White-Black African)	-	9 (0.1)	13 (0.1)
Mixed (White-Black Caribbean)	-	16 (0.2)	34 (0.3)
Other (Mixed)	-	18 (0.2)	35 (0.3)
Other	**94 (1.0)**	**87 (0.9)**	**89 (0.9)**

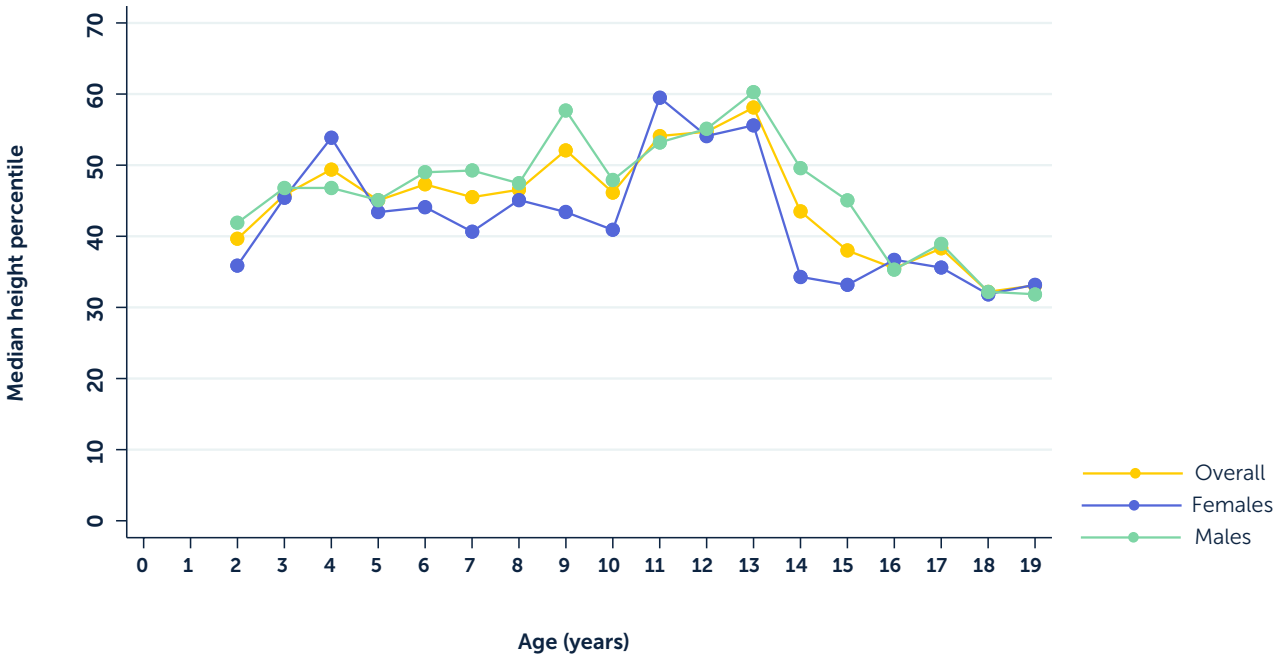
* Redacted to adhere to statistical disclosure guidelines.

** Further detail on mixed ethnicity categories were collected from 2016 onwards.

[1] Proportions are calculated from total known ethnicities.

1.5 Height percentiles of children and young people (<20 years)[1] N=4622

The following chart and table show the height percentiles of people with CF, aged between 2 and 19, in relation to UK growth data for the general population. If a person with CF is on the 40th percentile, only 40% of people the same age are their height or shorter; 60% are taller.



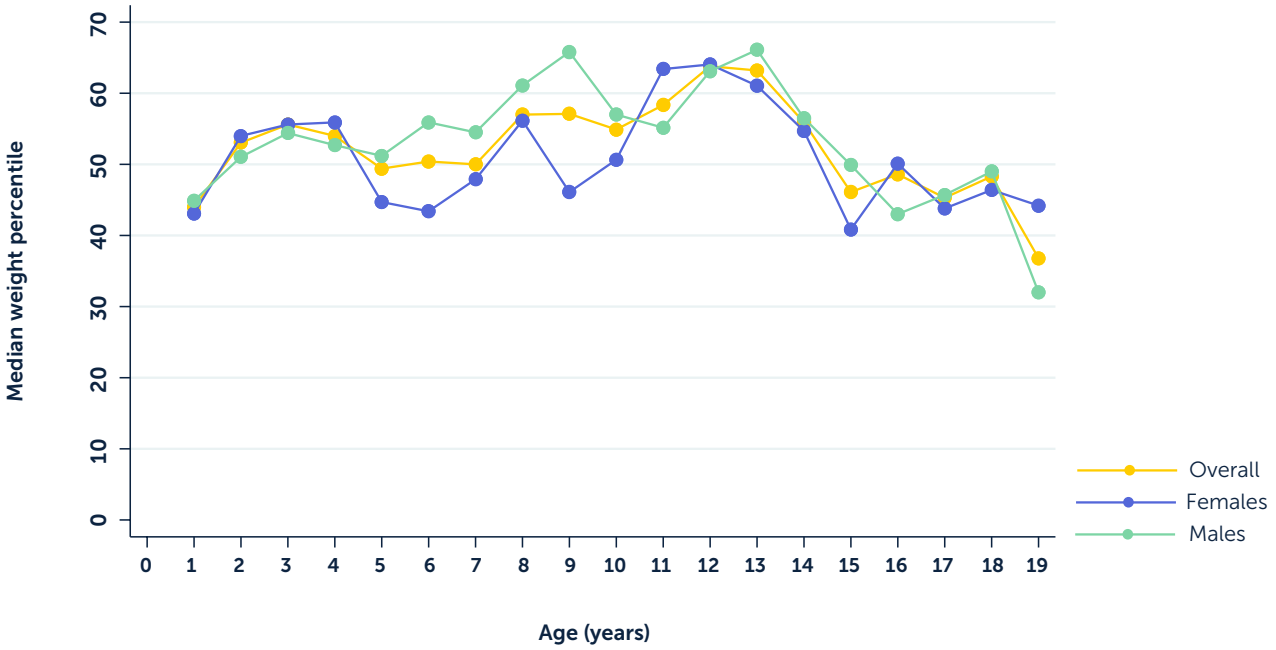
Age	Overall	Median	IQR	Female	Median	IQR	Male	Median	IQR
2	196	39.7	12.9-71.5	103	35.9	12.5-70.3	93	41.9	15.3-73.5
3	196	45.8	20.3-74.6	99	45.4	19.3-72.0	97	46.8	20.4-77.0
4	223	49.4	20.4-77.5	104	53.8	18.9-76.4	119	46.8	21.5-78.9
5	216	45.0	18.4-65.1	87	43.4	16.6-68.9	129	45.1	21.9-62.1
6	237	47.3	19.2-80.3	120	44.1	16.7-75.2	117	49.0	22.2-86.4
7	252	45.5	23.5-72.8	116	40.7	23.1-73.2	136	49.2	24.9-72.8
8	260	46.6	22.6-72.1	120	45.1	22.5-71.2	140	47.5	23.7-73.2
9	237	52.1	23.0-78.2	110	43.4	20.1-71.0	127	57.7	26.7-80.2
10	262	46.2	25.4-73.0	128	40.9	21.0-75.9	134	47.9	27.5-71.8
11	282	54.1	31.3-77.3	133	59.5	31.9-81.3	149	53.2	30.7-73.1
12	291	54.7	28.5-80.0	145	54.1	28.5-78.2	146	55.1	29.4-81.3
13	276	58.1	30.5-81.4	132	55.6	28.7-79.7	144	60.2	36.7-82.7
14	246	43.5	23.1-67.3	115	34.3	15.1-64.8	131	49.6	25.3-73.0
15	258	38.0	16.2-62.4	124	33.2	14.8-52.2	134	45.0	20.0-67.1
16	243	35.5	14.0-60.9	116	36.7	13.5-64.2	127	35.3	14.4-57.8
17	175	38.3	11.8-62.6	89	35.6	13.9-69.8	86	39.0	11.8-59.3
18	215	32.2	12.8-58.1	96	31.9	13.8-52.7	119	32.2	10.4-62.5
19	223	33.1	13.6-65.1	115	33.2	13.6-65.3	108	31.8	14.8-61.0
Overall	**4288**	**45.3**	**21.2-72.0**	**2052**	**42.5**	**19.7-71.9**	**2236**	**47.5**	**22.4-72.2**

* Number with non-missing data.

[1] Based on UK-WHO growth charts, 1990 (updated 1996).

1.6 Weight percentiles of children and young people (<20 years)[1] N=4622

The following chart and table show the weight of people with CF, aged between 1 and 19, in relation to the UK growth data for the general population. If a person with CF is on the 40th percentile, only 40% of people the same age are their weight or lower; 60% weigh more.



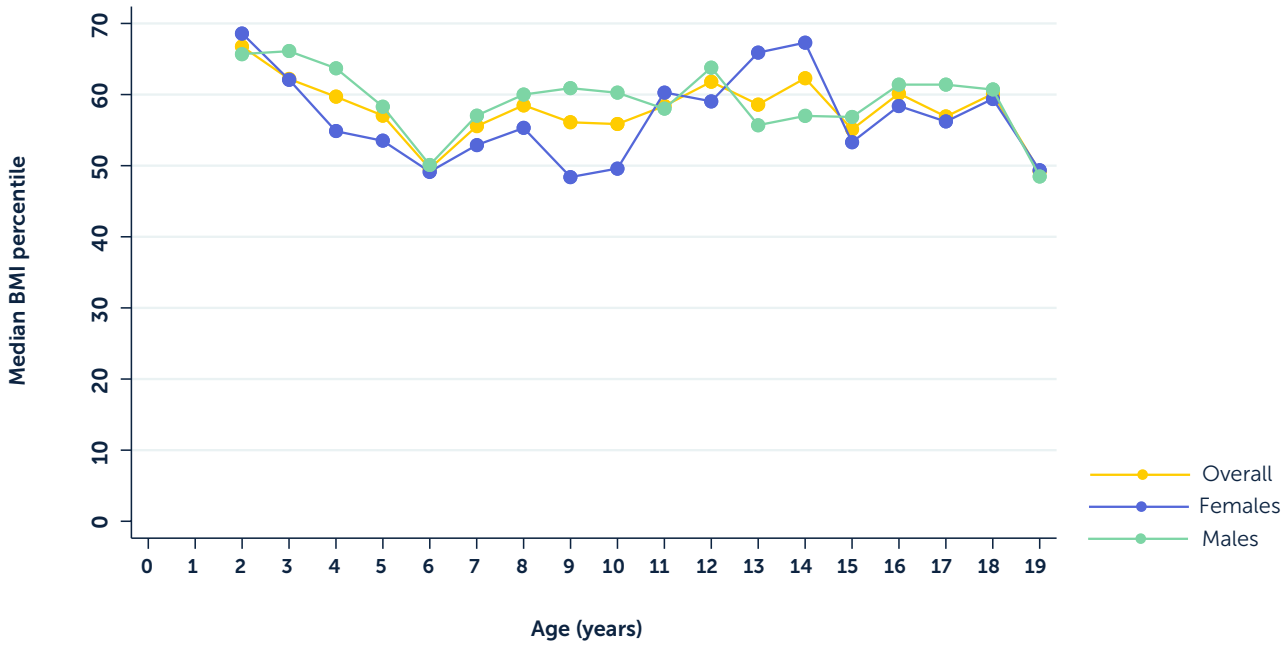
	Overall			Female			Male		
Age	n	Median	IQR	n	Median	IQR	n	Median	IQR
1	204	44.0	20.5-73.2	105	43.1	15.8-74.4	99	44.9	22.7-72.0
2	204	53.1	26.0-82.2	108	54.0	26.5-84.7	96	51.0	25.8-80.2
3	197	55.6	30.3-80.6	99	55.6	24.5-80.6	98	54.4	33.8-81.5
4	224	54.0	28.5-79.4	105	55.9	28.2-74.4	119	52.7	28.5-82.6
5	218	49.4	25.7-74.5	87	44.7	22.5-75.1	131	51.2	26.6-74.3
6	237	50.4	26.3-78.0	120	43.4	25.8-75.2	117	55.9	26.3-81.6
7	254	50.0	31.4-77.7	117	47.9	31.3-77.7	137	54.5	32.6-77.1
8	263	57.0	28.9-78.2	122	56.2	30.5-74.5	141	61.1	26.7-80.6
9	238	57.1	27.5-83.4	111	46.1	24.7-74.3	127	65.8	32.6-86.0
10	263	54.9	31.2-79.2	128	50.7	26.9-77.1	135	57.0	36.4-81.5
11	282	58.3	32.9-84.8	132	63.4	33.5-84.9	150	55.2	28.9-83.1
12	291	63.8	30.9-85.6	144	64.1	27.5-85.1	147	63.1	31.9-86.7
13	275	63.2	38.1-87.4	132	61.0	39.7-86.2	143	66.1	36.2-87.8
14	246	55.9	34.0-78.2	115	54.7	31.4-78.3	131	56.5	35.3-78.2
15	259	46.1	20.4-74.3	124	40.8	18.0-72.7	135	49.9	24.3-77.1
16	243	48.6	20.8-75.8	117	50.1	26.1-77.3	126	43.0	14.2-72.5
17	175	45.3	20.4-71.9	89	43.8	14.5-69.5	86	45.7	24.0-77.6
18	211	48.3	13.9-76.9	94	46.4	20.0-81.8	117	49.0	10.6-71.6
19	219	36.8	13.9-69.0	112	44.2	15.6-73.5	107	32.0	13.5-67.4
Overall	**4503**	**53.0**	**26.7-79.2**	**2161**	**51.7**	**26.0-78.5**	**2342**	**54.0**	**27.1-79.7**

* Number with non-missing data.

[1] Based on UK-WHO growth charts, 1990 (updated 1996).

1.7a Body Mass Index (BMI) percentiles in children and young people (<20 years)[1]
N=4622

The following chart and table show the BMI percentiles of people with CF, aged between 2 and 19, in relation to the UK growth data for the general population. If a person with CF is on the 40th percentile, it means that only 40% of the population at the same age have the same BMI or lower; 60% have a higher BMI.



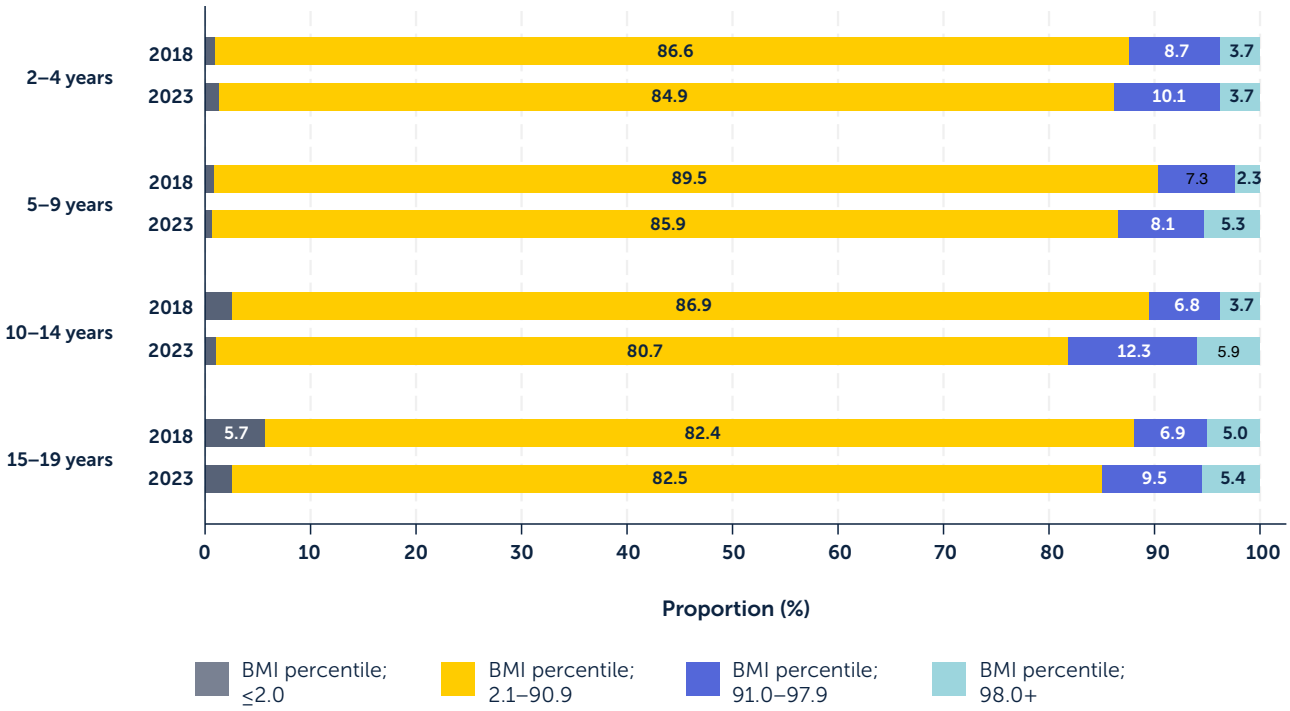
	Overall			Female			Male		
Age	n	Median	IQR	n	Median	IQR	n	Median	IQR
2	196	66.8	36.4-84.9	103	68.6	39.7-86.3	93	65.7	35.1-83.0
3	196	62.2	37.4-83.2	99	62.1	38.4-80.7	97	66.1	37.4-85.6
4	223	59.7	33.5-79.7	104	54.8	31.5-75.9	119	63.7	35.0-80.9
5	216	57.0	31.2-78.8	87	53.5	32.0-72.5	129	58.3	30.8-80.6
6	237	49.6	28.4-73.9	120	49.2	27.2-74.1	117	50.1	31.3-73.4
7	252	55.6	35.0-76.3	116	52.9	33.8-75.2	136	57.1	36.8-76.3
8	260	58.5	35.7-81.0	120	55.3	36.0-77.9	140	60.0	35.3-85.4
9	237	56.1	31.7-83.2	110	48.4	24.6-75.3	127	60.9	37.2-85.4
10	262	55.8	35.6-82.2	128	49.6	32.0-74.5	134	60.2	40.2-85.5
11	281	58.4	28.5-84.0	132	60.3	32.5-83.4	149	58.0	26.9-84.0
12	290	61.8	32.2-85.1	144	59.1	31.5-82.5	146	63.8	36.0-86.6
13	274	58.6	31.6-88.4	131	65.9	36.9-89.5	143	55.7	27.8-88.4
14	246	62.3	32.4-85.6	115	67.3	43.6-85.5	131	57.0	27.0-85.8
15	258	55.1	30.8-82.7	124	53.3	28.5-83.3	134	56.8	34.2-82.0
16	242	60.2	34.1-82.7	116	58.4	39.1-81.9	126	61.4	27.8-85.0
17	175	56.9	33.4-80.7	89	56.2	24.8-76.8	86	61.4	39.5-82.0
18	211	60.1	27.0-81.9	94	59.4	27.0-82.9	117	60.7	27.1-80.0
19	219	49.2	22.3-75.6	112	49.3	22.8-78.2	107	48.5	22.3-73.7
Overall	**4275**	**58.0**	**32.1-82.1**	**2044**	**56.8**	**32.1-81.1**	**2231**	**58.6**	**32.1-83.0**

* Number with non-missing data.

[1] Based on UK-WHO growth charts, 1990 (updated 1996).

1.7b Body Mass Index (BMI) percentiles in children and young people (<20 years)[1] for 2018 and 2023

The following graph shows the change in BMI groups for children and young people with CF from 2018 and 2023.



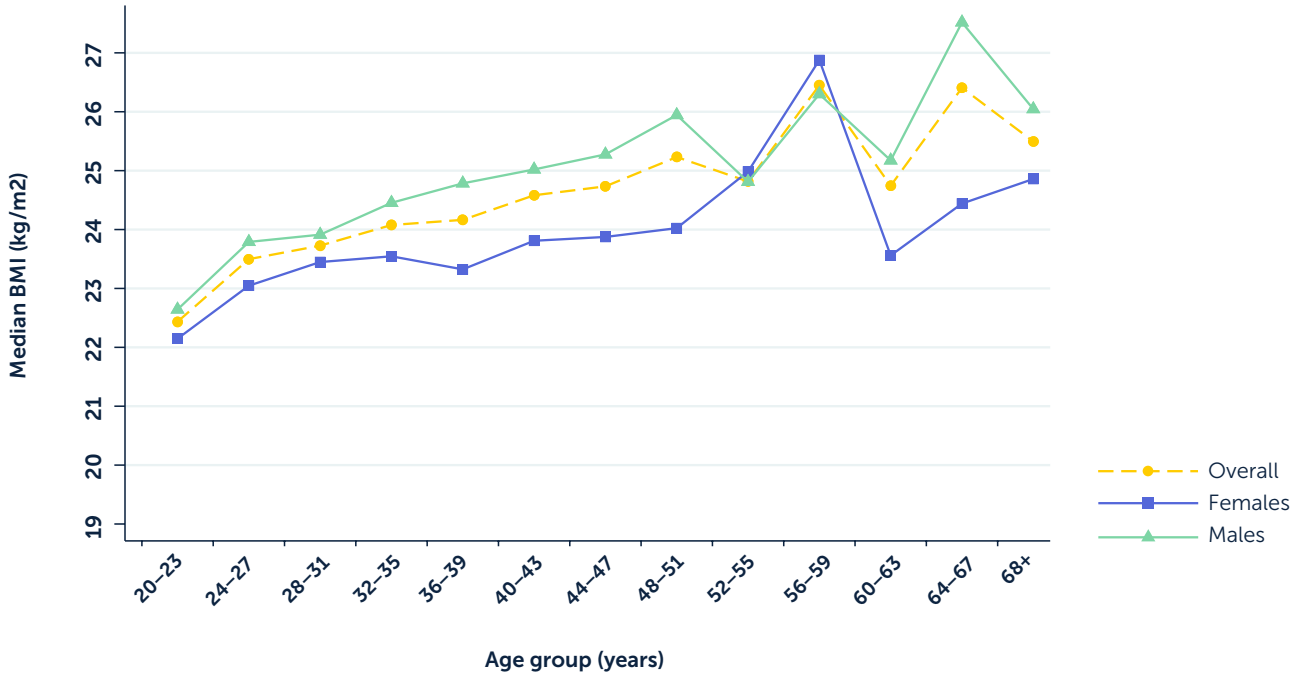
			BMI category by age and year : n*(%)			
Age group	Year	Total number of people in each age group	BMI percentile; ≤2.0	BMI percentile; 2.1-90.9	BMI percentile; 91.0-97.9	BMI percentile; 98.0+
2-4 years	2018	790	7 (1.0)	580 (86.6)	58 (8.7)	25 (3.7)
	2023	632	8 (1.3)	522 (84.9)	62 (10.1)	23 (3.7)
5-9 years	2018	1381	11 (0.9)	1114 (89.5)	91 (7.3)	29 (2.3)
	2023	1222	9 (0.7)	1032 (85.9)	97 (8.1)	64 (5.3)
10-14 years	2018	1222	29 (2.6)	971 (86.9)	76 (6.8)	41 (3.7)
	2023	1371	15 (1.1)	1091 (80.6)	167 (12.3)	80 (5.9)
15-19 years	2018	1063	55 (5.7)	798 (82.4)	67 (6.9)	48 (5.0)
	2023	1128	29 (2.6)	911 (82.4)	105 (9.5)	60 (5.4)

* Number with non-missing data.

[1] Based on UK-WHO growth charts, 1990 (updated 1996).

1.8a Body Mass Index (BMI) in adults (20 years and over) N=5722

The following chart and table show the BMI of people with CF aged 20 and over.

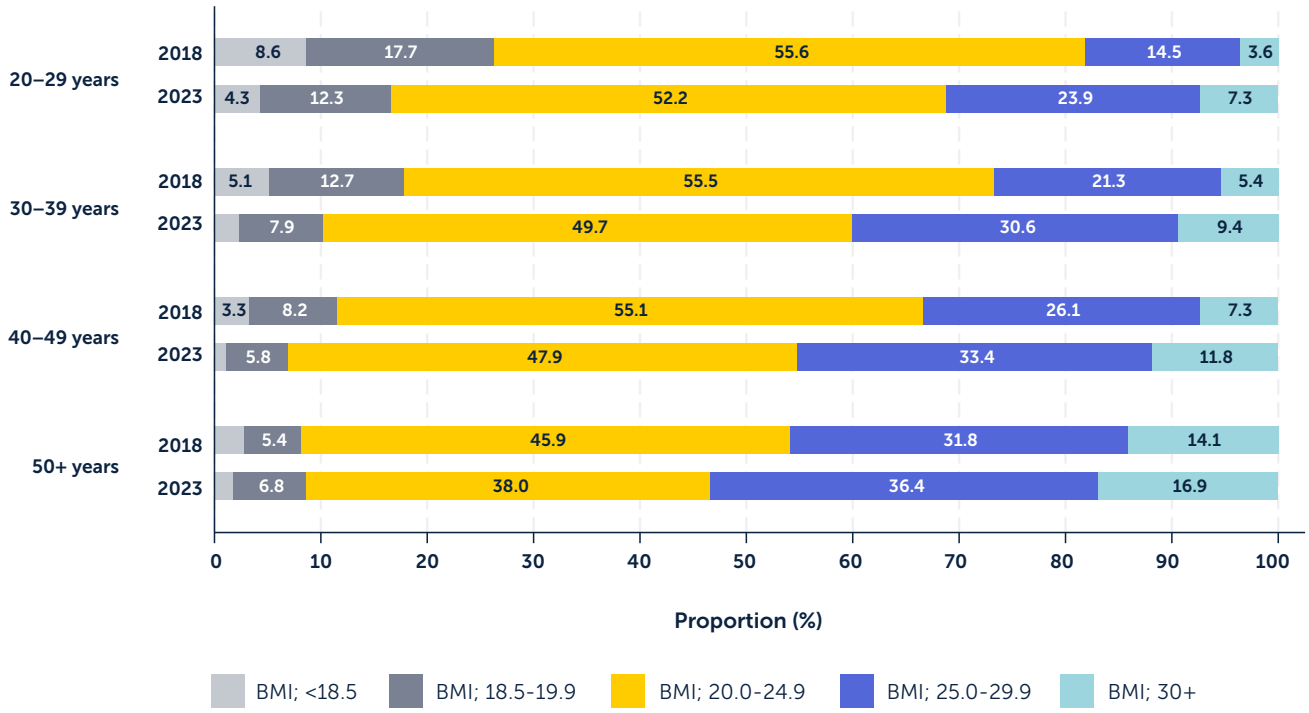


	Overall			Female			Male		
Age	n	Median	IQR	n	Median	IQR	n	Median	IQR
20-23	785	22.4	20.4-25.0	408	22.1	20.2-25.1	377	22.6	20.6-25.0
24-27	898	23.5	21.1-26.2	431	23.0	21.0-26.2	467	23.8	21.1-26.1
28-31	832	23.7	21.8-26.4	399	23.4	21.2-26.2	433	23.9	22.1-26.6
32-35	782	24.1	21.9-26.6	349	23.5	21.4-26.4	433	24.5	22.3-26.8
36-39	622	24.2	21.8-27.0	267	23.3	21.1-26.4	355	24.8	22.6-27.4
40-43	514	24.6	22.1-27.1	221	23.8	21.4-26.7	293	25.0	22.8-27.3
44-47	362	24.7	22.3-27.5	147	23.9	21.6-27.0	215	25.3	23.1-27.6
48-51	224	25.2	22.3-27.6	95	24.0	21.4-27.1	129	25.9	23.4-28.2
52-55	217	24.8	22.4-28.4	93	25.0	21.0-29.4	124	24.8	23.0-27.7
56-59	135	26.4	23.5-29.4	53	26.9	22.6-29.7	82	26.3	24.1-28.8
60-63	102	24.7	22.7-27.7	44	23.6	22.1-27.1	58	25.2	23.2-28.1
64-67	54	26.4	22.7-29.1	22	24.4	20.7-28.1	32	27.5	24.5-29.7
68+	77	25.5	22.2-28.3	38	24.9	19.7-28.8	39	26.0	23.5-28.2
Overall	**5604***	**23.9**	**21.6-26.8**	**2567**	**23.3**	**21.0-26.5**	**3037**	**24.4**	**22.1-26.9**

* Number with non-missing data.

1.8b Body Mass Index (BMI) in adults for 2018 and 2023

The following graph shows the change in the proportion of people in each BMI group for 2018 and 2023.



			BMI category by age and year : n*(%)				
Age group	Year	Total number of people in each age group	BMI; <18.5	BMI; 18.5-19.9	BMI; 20.0-24.9	BMI; 25.0-29.9	BMI; 30+
20-29 years	2018	2341	201 (8.6)	413 (17.7)	1297 (55.6)	337 (14.5)	84 (3.6)
	2023	2154	90 (4.3)	260 (12.3)	1104 (52.2)	506 (23.9)	154 (7.3)
30-39 years	2018	1574	79 (5.1)	198 (12.7)	868 (55.6)	333 (21.3)	84 (5.4)
	2023	1844	42 (2.3)	143 (7.9)	897 (49.7)	553 (30.6)	170 (9.4)
40-49 years	2018	704	23 (3.3)	57 (8.2)	383 (55.0)	182 (26.1)	51 (7.3)
	2023	998	11 (1.1)	57 (5.8)	468 (47.9)	326 (33.4)	115 (11.8)
50+ years	2018	469	13 (2.8)	25 (5.4)	212 (45.9)	147 (31.8)	65 (14.1)
	2023	726	13 (1.8)	48 (6.8)	269 (38.0)	258 (36.4)	120 (16.9)

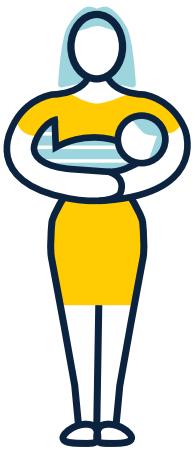
1.9 Education and employment in adults (16 years and over) N=6588

The following table shows how people with CF reported their education and employment status in 2023.

	2020	2021	2022	2023		
	Overall	Overall	Overall	Overall	Male	Female
Number of patients	6012	6297	6445	6588	3548	3040
Number who completed questionnaire; n(%)	5968 (99.3)	6296 (100.0)	6442 (100.0)	6587 (100.0)	3547 (100.0)	3040 (100.0)
Full-time employment; n(%)	1975 (32.9)	2097 (33.3)	2228 (34.6)	2379 (36.1)	1573 (44.3)	806 (26.5)
Part-time employment; n(%)	894 (14.9)	915 (14.5)	981 (15.2)	1031 (15.6)	376 (10.6)	655 (21.5)
Student; n(%)	1015 (16.9)	1061 (16.8)	1046 (16.2)	1049 (15.9)	520 (14.7)	529 (17.4)
Homemaker; n(%)	200 (3.3)	251 (4.0)	249 (3.9)	257 (3.9)	29 (0.8)	228 (7.5)
Unemployed; n(%)	847 (14.1)	791 (12.6)	767 (11.9)	744 (11.3)	432 (12.2)	312 (10.3)
Disabled; n(%)	274 (4.6)	255 (4.0)	228 (3.5)	237 (3.6)	120 (3.4)	117 (3.8)
Retired; n(%)	139 (2.3)	162 (2.6)	170 (2.6)	184 (2.8)	108 (3.0)	76 (2.5)
Volunteer; n(%)	11 (0.2)	12 (0.2)	14 (0.2)	21 (0.3)	10 (0.3)	11 (0.4)
Unknown entered; n(%)	613 (10.2)	752 (11.9)	759 (11.8)	685 (10.4)	379 (10.7)	306 (10.1)
No. in work or study; n(%)	**3884 (65.1)**	**4073 (64.7)**	**4255 (66.1)**	**4459 (67.7)**	**2469 (69.6)**	**1990 (65.5)**

1.10 Parenthood

	2020	2021	2022	2023
Women with CF who had babies; n	56	103	140	116
Men with CF who became fathers; n	44	30	33	31

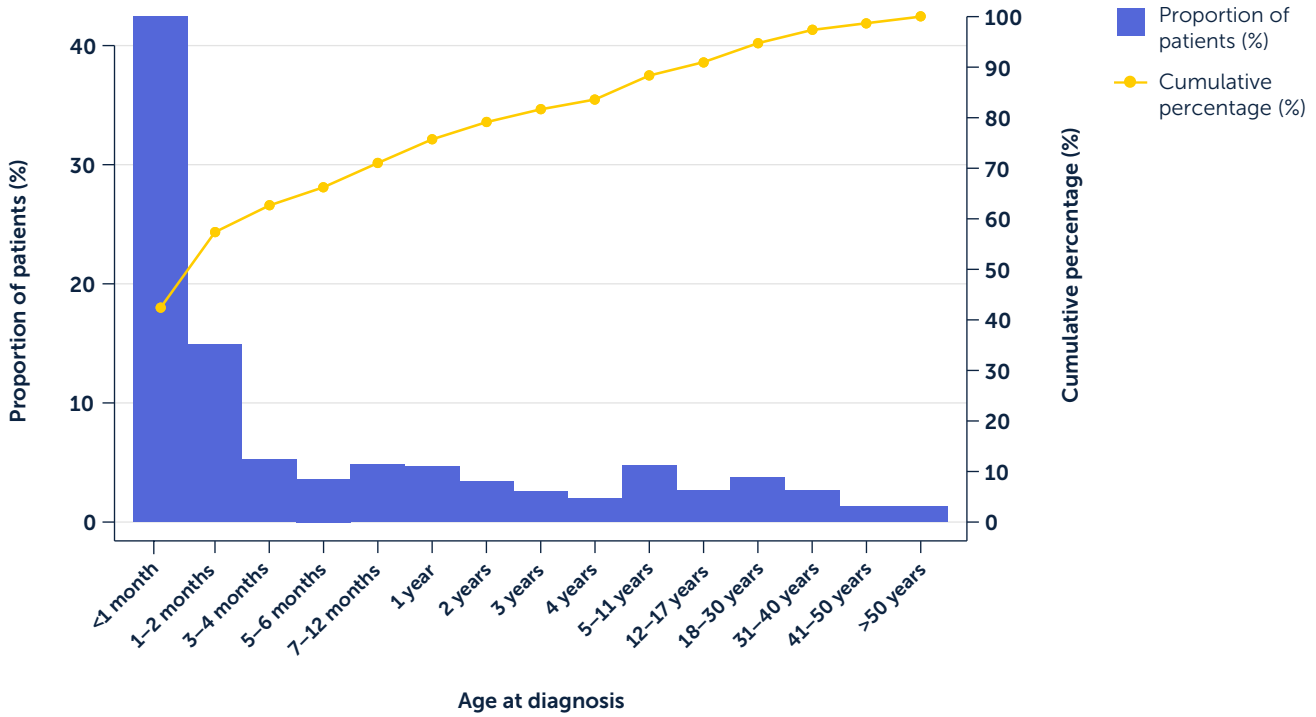


116 women with CF had babies in 2023

31 men with CF became fathers in 2023

Diagnosis of cystic fibrosis

1.11 Age at diagnosis
N=11318



The median age at diagnosis for patients aged under 16 in 2023 is **21 days**.

Newborn screening for CF has been done routinely in the whole of the UK since mid-2007. It is part of the heel prick blood spot testing done at 5–7 days of age. The blood sample is tested for a number of conditions, including cystic fibrosis. This means that more babies born after 2007 receive an early diagnosis than those born prior to 2007.

A total of **124 (72%)** out of 173 patients diagnosed in 2023 were identified by newborn screening (including those without an annual review).

1,102 (14.9%) of adults with CF in the Registry in 2023 were diagnosed at age 16 or over.

In 2023, **25** people aged 16 or over were newly diagnosed with cystic fibrosis.

1.12 Mode of presentation

The following tables show the top five most frequent modes of presentation for those diagnosed between 2013–2023 and those born between 2013–2023, excluding those recorded as being diagnosed through newborn screening (NBS) or genotype. Patients may present with multiple symptoms so percentages may not add to 100.

	All patients diagnosed 2013-2023	Age <16 at diagnosis	Age ≥16 at diagnosis
Total patients	**2864**	**2428**	**-***
Number diagnosed by newborn screening	2027	2026	<5
Total non-NBS or Genotype	837	402	431
Presentation type			
Persistent or acute respiratory infection	254 (30.5)	101 (25.1)	153 (35.5)
Meconium ileus	140 (16.8)	140 (34.8)	0 (0.0)
Family history	135 (16.2)	77 (19.2)	58 (13.5)
Bronchiectasis	116 (13.9)	9 (2.2)	107 (24.8)
Failure to thrive/malnutrition	60 (7.2)	55 (13.7)	5 (1.2)

	All patients born 2013-2023
Total patients	**2274**
Number diagnosed by newborn screening or genotype	1970
Total non-NBS or Genotype	304
Presentation Type	
Meconium ileus	138 (45.5)
Family history	66 (21.8)
Persistent or acute respiratory infection	45 (14.9)
Prenatal	43 (14.2)
Failure to thrive/malnutrition	35 (11.6)

* Multiple presentation types can be indicated so percentage may not add up to 100.

** Redacted to adhere to statistical disclosure guidelines.

Lung health

For people with CF, mucus in the lungs is linked to repeat or chronic infections. This can cause permanent damage, making it harder to breathe.

In CF, the condition of the lungs is often measured using FEV_1; the Forced Expiratory Volume of air in the first second of a forced exhaled breath. In this report, an FEV_1% predicted is based on the FEV_1 we would expect for a person without CF of the same age, sex, height, and ethnicity.

A person with CF who has FEV_1% predicted of 100% can breathe out the same amount of air in the first second of an exhaled breath as we would expect from a comparable person without cystic fibrosis. A person with CF who has an FEV_1% predicted of 50% breathes out half the volume of air as a comparable person without cystic fibrosis.

For people with CF, an FEV_1% predicted of 85% or higher is the target, as this indicates normal or near-normal lung health. Each individual with CF will have their own FEV_1 target, based on their own lung function results and trends.

An aim of CF care is to prevent FEV_1% predicted from falling as much as possible, for as long as possible. This is often a team effort between people with CF, their family, and their medical team, which can include doctors, nurses, physiotherapists, dietitians, and psychologists.

The FEV_1% predicted values shown in this report are calculated using an equation called Global Lungs Initiative, or GLI.[1]

[1] Quanjer et al. Eur respir J. 2012 40(6):1324-1343.

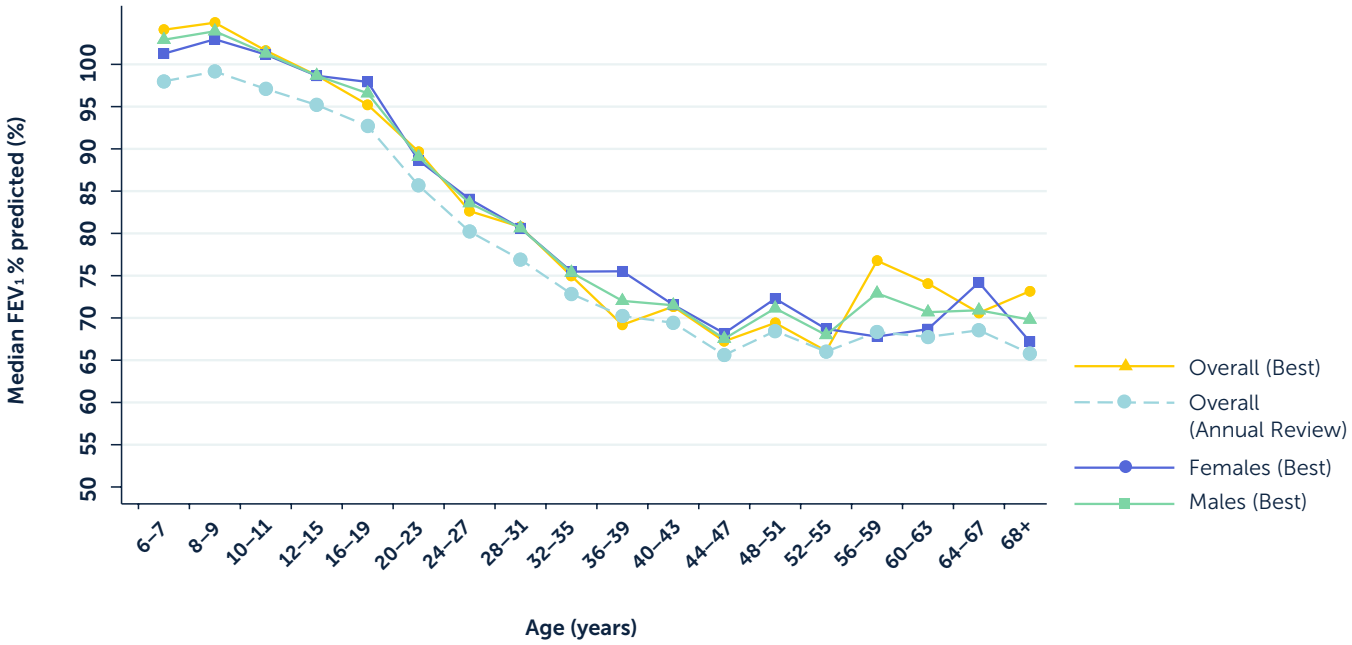
1.13 Annual review FEV_1% predicted (GLI equations) in patients aged six years and older who have not had a lung transplant N=9073

People with CF who have had lung transplants are excluded, as their new 'non-CF' lungs may have lung health similar to a person without cystic fibrosis.

	Overall			Female			Male		
Age (years)	n	Median	IQR	n	Median	IQR	n	Median	IQR
6-7	438	98.0	88.7-107.7	215	98.4	89.9-109.2	223	97.6	87.2-106.2
8-9	469	99.2	91.3-107.4	219	100.3	93.2-109.1	250	97.6	88.9-106.5
10-11	508	97.1	86.8-104.8	240	97.3	86.9-105.2	268	96.4	86.5-104.3
12-15	1014	95.2	85.8-104.3	492	94.9	86.1-103.9	522	95.2	85.4-104.8
16-19	815	92.7	81.8-101.8	399	91.8	80.9-100.7	416	93.9	83.3-102.3
20-23	738	85.7	72.6-98.2	386	86.3	72.7-98.3	352	85.2	72.5-98.1
24-27	829	80.2	63.5-94.2	401	80.2	62.6-94.2	428	80.5	65.0-94.2
28-31	769	76.9	58.9-90.9	378	75.5	58.0-89.5	391	77.7	59.3-91.8
32-35	710	72.8	55.8-88.4	316	72.3	56.5-86.9	394	73.2	55.4-89.7
36-39	572	70.2	49.9-85.7	246	67.7	48.4-81.7	326	73.5	51.1-88.0
40-43	470	69.4	48.9-85.8	207	65.5	47.4-83.9	263	70.4	50.3-87.6
44-47	330	65.6	49.6-82.9	134	64.9	49.6-80.8	196	67.5	49.5-84.0
48-51	207	68.4	50.0-83.9	87	65.1	44.8-82.1	120	70.0	54.4-85.1
52-55	199	66.0	49.6-83.4	86	61.6	49.6-84.1	113	67.4	50.5-83.1
56-59	128	68.3	49.3-86.3	52	75.5	53.5-89.8	76	65.0	48.1-85.3
60-63	97	67.8	52.0-88.1	43	67.8	52.6-83.5	54	67.1	43.2-88.9
64-67	49	68.5	53.2-83.3	19	67.4	53.2-78.3	30	70.3	53.1-85.8
68+	73	65.8	48.4-83.9	37	70.8	50.0-85.6	36	65.4	42.2-82.0
<16	2429	97.0	87.3-105.9	1166	97.5	88.0-106.5	1263	96.4	86.7-105.2
≥16	5986	78.3	58.7-92.8	2791	77.8	58.3-92.2	3195	78.8	59.1-93.4
<18	2831	96.6	86.6-105.3	1364	97.0	87.0-105.9	1467	96.3	86.1-105.1
≥18	5584	76.8	57.3-91.7	2593	76.1	56.7-90.9	2991	77.6	57.8-92.3
Overall	**8415***	**85.3**	**66.4-98.6**	**3957**	**85.3**	**66.4-98.5**	**4458**	**85.2**	**66.4-98.6**

* Number with non-missing data.

1.14 Best* FEV_1% predicted (GLI equations) in patients aged six years and older who have not had a lung transplant N=9073

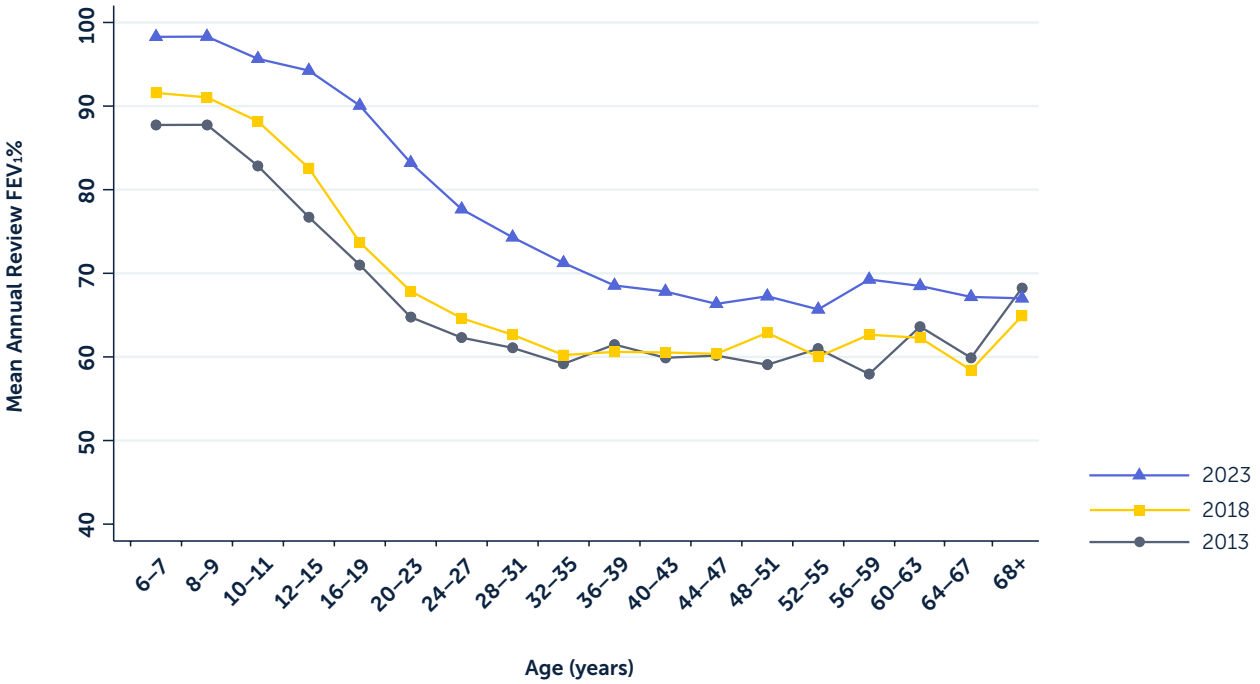


	Overall			Female			Male		
Age (years)	n	Median	IQR	n	Median	IQR	n	Median	IQR
6-7	471	102.9	94.4-112.3	230	104.1	95.3-114.5	241	101.3	94.2-110.6
8-9	490	103.9	96.6-111.2	228	104.9	97.4-112.5	262	103.0	95.6-110.3
10-11	533	101.3	93.5-108.6	254	101.6	93.8-110.3	279	101.2	93.4-107.7
12-15	1060	98.7	89.7-108.1	511	98.7	89.8-108.0	549	98.7	89.7-108.3
16-19	848	96.6	85.4-104.9	414	95.2	84.8-103.5	434	97.9	86.1-106.2
20-23	779	89.1	75.9-100.6	403	89.7	76.5-100.9	376	88.6	75.2-100.3
24-27	882	83.6	66.9-97.3	424	82.6	66.1-97.6	458	84.1	67.6-96.7
28-31	816	80.6	60.7-94.1	395	80.8	60.1-93.6	421	80.6	62.2-94.5
32-35	750	75.4	58.8-90.4	338	75.0	61.1-89.9	412	75.5	57.8-90.6
36-39	599	72.0	54.1-89.2	254	69.2	52.3-87.0	345	75.5	55.3-89.9
40-43	487	71.5	51.6-89.1	214	71.3	50.2-87.6	273	71.6	52.2-89.9
44-47	348	67.5	53.3-86.6	138	67.2	54.0-84.4	210	68.2	52.3-87.1
48-51	216	71.1	55.1-86.6	89	69.4	49.5-86.1	127	72.3	58.8-89.0
52-55	208	68.0	51.1-86.7	89	66.1	51.5-89.3	119	68.7	51.0-86.3
56-59	133	72.9	53.1-88.0	55	76.8	57.3-90.3	78	67.8	49.9-88.0
60-63	101	70.7	53.1-88.1	44	74.1	54.0-87.9	57	68.7	43.9-92.2
64-67	54	70.9	58.3-89.3	22	70.6	60.5-86.0	32	74.2	55.3-92.0
68+	77	69.8	48.8-90.3	39	73.2	59.6-92.8	38	67.2	42.9-83.9
<16	2554	101.1	92.3-109.6	1223	101.3	92.8-110.5	1331	101.1	92.0-109.2
≥16	6298	81.4	62.0-96.0	2918	81.5	62.3-95.8	3380	81.3	61.9-96.1
<18	2969	100.7	91.7-109.2	1427	100.6	91.9-109.5	1542	100.7	91.4-108.6
≥18	5883	79.9	60.5-94.5	2714	79.7	60.6-94.3	3169	80.1	60.5-94.7
Overall	**8852****	**89.1**	**70.0-101.8**	**4141**	**89.6**	**70.3-101.9**	**4711**	**88.8**	**69.7-101.7**

* Where Best FEV_1% was missing or less than the FEV_1% at annual review, annual review FEV_1% was used instead.
** Number with non-missing data.

1.15 Annual review FEV_1% predicted (GLI equations) over time in patients aged six years and older who have not had a lung transplant N=9073 in 2023, N=8320 in 2018, N=7504 in 2013

As we learn more about CF and how to treat it, we hope to improve the outcomes of people with the condition. The chart below shows how FEV_1 in 2023 compares to Registry data from 2013 and 2018.



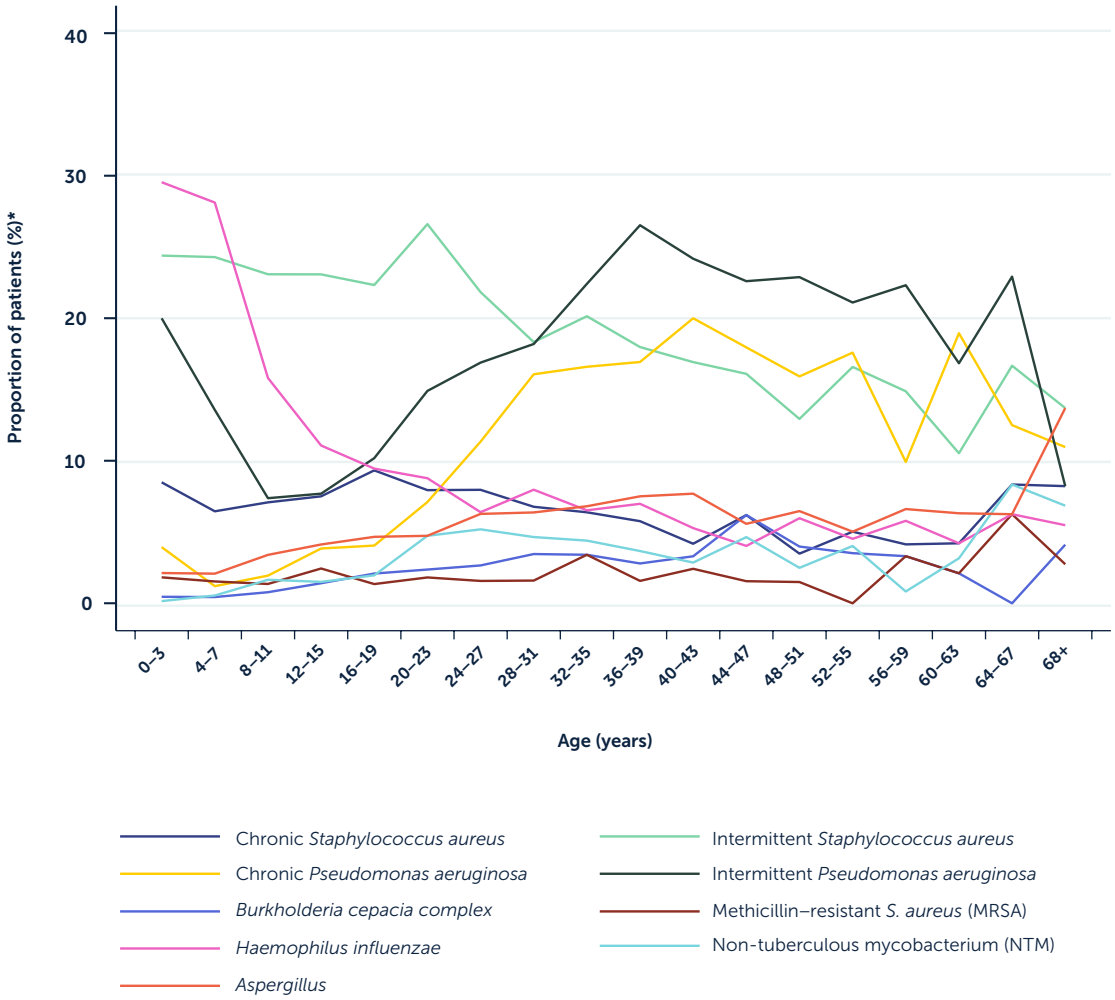
	2013		2018		2023		
Age (years)	n	FEV_1% : Mean (SD)	n	FEV_1% : Mean (SD)	n	FEV_1% : Mean (SD)	p-values (t-test*)
6-7	399	87.7 (16.3)	539	91.6 (16.9)	438	98.3 (17.1)	<0.001
8-9	434	87.8 (15.7)	526	91.0 (15.6)	469	98.3 (13.9)	<0.001
10-11	401	82.8 (15.9)	502	88.2 (16.2)	508	95.7 (14.0)	<0.001
12-15	908	76.7 (18.1)	890	82.6 (17.4)	1014	94.2 (15.6)	<0.001
16-19	944	71.0 (22.1)	835	73.7 (20.9)	815	90.1 (17.6)	<0.001
20-23	934	64.8 (23.5)	954	67.9 (23.0)	738	83.2 (20.4)	<0.001
24-27	782	62.3 (24.2)	857	64.6 (23.4)	829	77.7 (22.7)	<0.001
28-31	669	61.1 (23.4)	779	62.7 (23.7)	769	74.3 (22.6)	<0.001
32-35	479	59.2 (22.9)	601	60.2 (23.4)	710	71.2 (23.4)	<0.001
36-39	299	61.5 (23.3)	493	60.6 (23.4)	572	68.5 (23.3)	<0.001
40-43	273	59.9 (23.5)	310	60.5 (24.3)	470	67.8 (23.7)	<0.001
44-47	198	60.2 (26.6)	244	60.4 (22.8)	330	66.3 (23.0)	0.002
48-51	138	59.1 (24.4)	199	62.9 (24.3)	207	67.3 (22.6)	0.061
52-55	72	61.0 (26.2)	143	60.0 (25.8)	199	65.7 (22.4)	0.031
56-59	46	57.9 (23.3)	84	62.7 (25.9)	128	69.3 (23.5)	0.056
60-63	28	63.6 (24.7)	54	62.3 (22.3)	97	68.5 (24.4)	0.125
64-67	20	59.9 (27.6)	26	58.4 (25.1)	49	67.2 (21.5)	0.118
68+	21	68.2 (25.8)	46	64.9 (23.7)	73	67.0 (24.9)	0.648
<16	2142	82.2 (17.6)	2457	87.5 (17.1)	2429	96.1 (15.3)	-
≥16	4903	63.6 (23.8)	5625	64.8 (23.6)	5986	75.3 (23.4)	-
<18	2616	80.4 (18.7)	2857	86.0 (17.7)	2831	95.2 (15.8)	-
≥18	4429	62.6 (23.9)	5225	63.8 (23.7)	5584	74.2 (23.4)	-

* T-test comparing 2023 with 2018.

Lung infections

Lung infections can permanently reduce lung function in people with cystic fibrosis. Some lung infections can become 'chronic', meaning that they can't ever be removed completely using medicines. All other infections are reported if they have occurred at least once as a positive growth in the 12 months prior to the patient's annual review.

1.16 Lung infections in 2023
N=9579*



* Proportions are calculated from the number of patients with at least one sample taken in the relevant age group, This is a change from the 2020 data report where they were calculated from the number of people with annual reviews in the age group.

1.17 Lung infections in 2023 (contd.)
<16 years N=3756, ≥16 years N=6588

	Paediatric age range (years)				Overall
	0-3	4-7	8-11	12-15	Paediatric (<16 years)
Number in age range	676	942	1053	1085	3756
Number who had culture taken*	660	914	1031	1066	3671
Chronic *S. aureus* n(%)	56 (8.5)	59 (6.5)	73 (7.1)	80 (7.5)	268 (7.3)
Intermittent *S. aureus* n(%)	161 (24.4)	222 (24.3)	238 (23.1)	246 (23.1)	867 (23.6)
Chronic *P. aeruginosa* n(%)	26 (3.9)	11 (1.2)	20 (1.9)	41 (3.8)	98 (2.7)
Intermittent *P. aeruginosa* n(%)	132 (20.0)	124 (13.6)	76 (7.4)	82 (7.7)	414 (11.3)
B. cepacia complex n(%)	<5	<5	8 (0.8)	15 (1.4)	30 (0.8)
B. cenocepacia n(%)	<5	<5	<5	9 (0.8)	14 (0.4)
B. multivorans n(%)	<5	<5	<5	<5	<5
B. other cepacia n(%)	<5	<5	<5	<5	7 (0.2)
MRSA n(%)	12 (1.8)	14 (1.5)	14 (1.4)	26 (2.4)	66 (1.8)
H. influenza n(%)	195 (29.5)	257 (28.1)	163 (15.8)	118 (11.1)	733 (20.0)
NTM n(%)	<5	5 (0.5)	17 (1.6)	16 (1.5)	39 (1.1)
Aspergillus fumigatus n(%)	14 (2.1)	19 (2.1)	35 (3.4)	44 (4.1)	112 (3.1)

Infections in this table reflect those grown in the 12 months prior to the 2023 annual review.
The UK CF Registry definition of 'chronic' is three or more isolates in the last 12 months.

* Proportions are calculated from the number of people who were recorded as having at least one respiratory culture sample taken.
** Redacted to adhere to statistical disclosure guidelines.

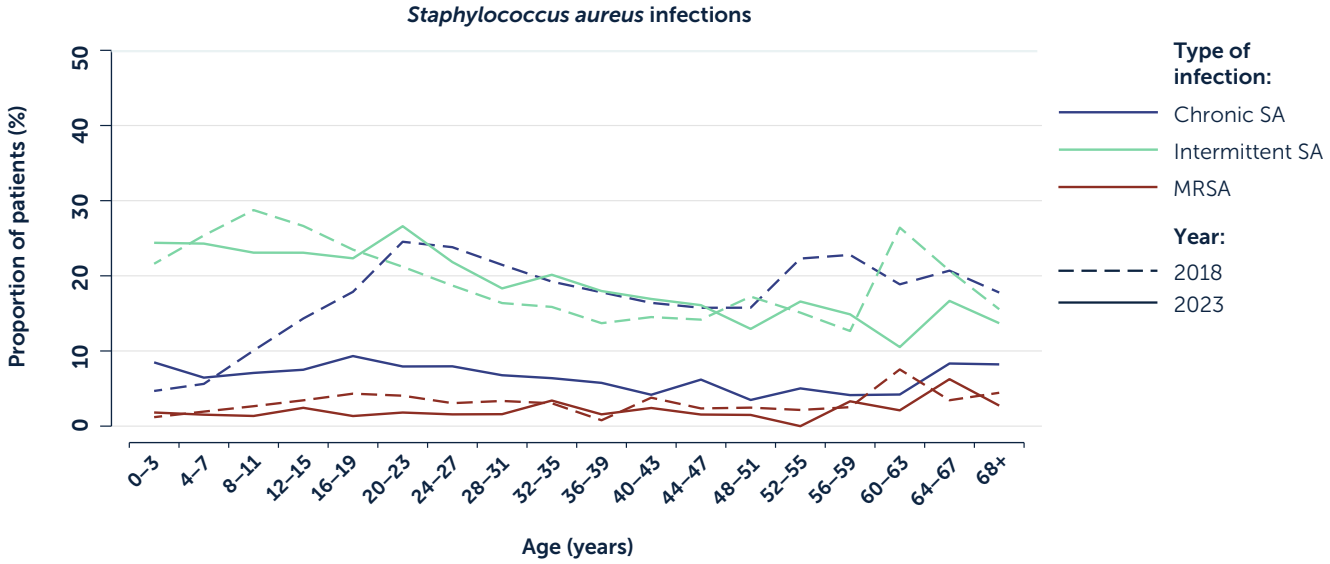
Lung infections in 2023 (contd.)
<16 years N=3756, ≥16 years N=6588

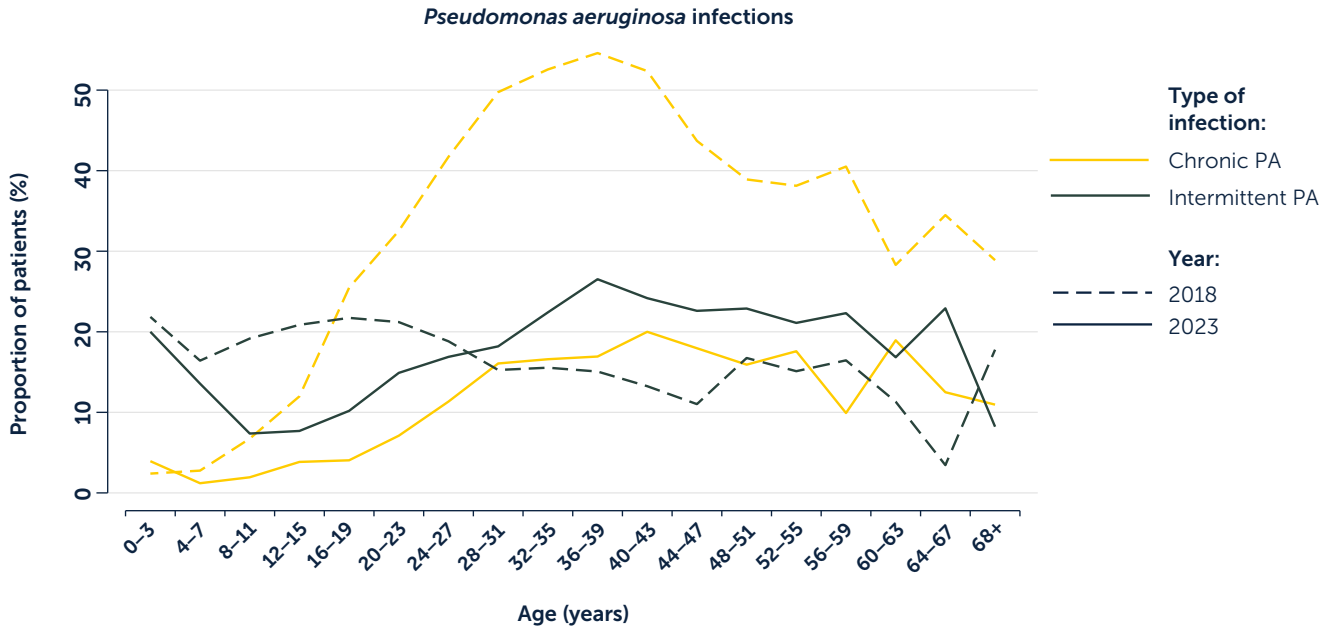
	Paediatric age range (years)							Overall
	16-19	20-23	24-27	28-31	32-35	36-39	40-43	Adults (≥16 years)
Number in age range	866	796	918	848	800	636	527	6588
Number who had culture taken*	815	718	829	753	705	573	455	5908
Chronic *S. aureus* n(%)	76 (9.3)	57 (7.9)	66 (8.0)	51 (6.8)	45 (6.4)	33 (5.8)	19 (4.2)	403 (6.8)
Intermittent *S. aureus* n(%)	182 (22.3)	191 (26.6)	181 (21.8)	138 (18.3)	142 (20.1)	103 (18.0)	77 (16.9)	1171 (19.8)
Chronic *P. aeruginosa* n(%)	33 (4.0)	51 (7.1)	94 (11.3)	121 (16.1)	117 (16.6)	97 (16.9)	91 (20.0)	773 (13.1)
Intermittent *P. aeruginosa* n(%)	83 (10.2)	107 (14.9)	140 (16.9)	137 (18.2)	158 (22.4)	152 (26.5)	110 (24.2)	1108 (18.8)
B. cepacia complex n(%)	17 (2.1)	17 (2.4)	22 (2.7)	26 (3.5)	24 (3.4)	16 (2.8)	15 (3.3)	181 (3.1)
B. cenocepacia n(%)	<5	7 (1.0)	<5	8 (1.1)	9 (1.3)	<5	6 (1.3)	53 (0.9)
B. multivorans n(%)	7 (0.9)	7 (1.0)	13 (1.6)	11 (1.5)	14 (2.0)	10 (1.7)	6 (1.3)	89 (1.5)
B. other cepacia n(%)	5 (0.6)	<5	<5	5 (0.7)	<5	<5	<5	22 (0.4)
MRSA n(%)	11 (1.3)	13 (1.8)	13 (1.6)	12 (1.6)	24 (3.4)	9 (1.6)	11 (2.4)	112 (1.9)
H. influenza n(%)	77 (9.4)	63 (8.8)	53 (6.4)	60 (8.0)	46 (6.5)	40 (7.0)	24 (5.3)	415 (7.0)
NTM n(%)	16 (2.0)	34 (4.7)	43 (5.2)	35 (4.6)	31 (4.4)	21 (3.7)	13 (2.9)	234 (4.0)
Aspergillus fumigatus n(%)	38 (4.7)	34 (4.7)	52 (6.3)	48 (6.4)	48 (6.8)	43 (7.5)	35 (7.7)	-**

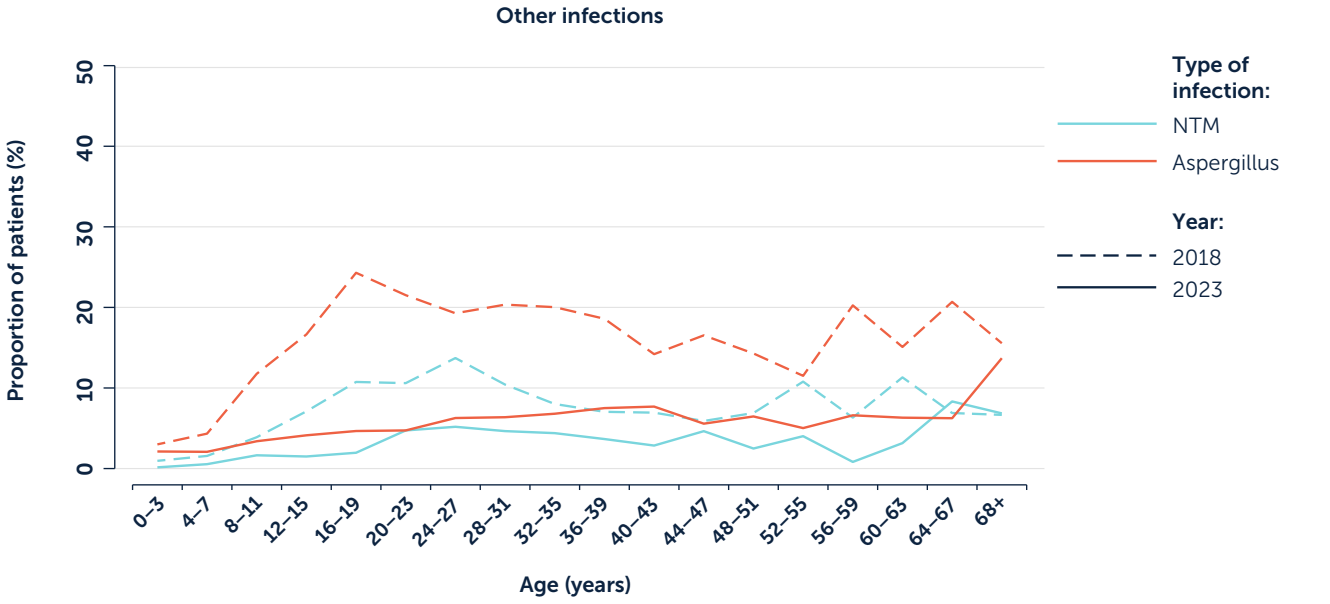
	Paediatric age range (years)							Overall
	44-47	48-51	52-55	56-59	60-63	64-67	68+	Adults (≥16 years)
Number in age range	367	230	225	140	103	54	78	6588
Number who had culture taken*	323	201	199	121	95	48	73	5908
Chronic *S. aureus* n(%)	20 (6.2)	7 (3.5)	10 (5.0)	5 (4.1)	<5	<5	6 (8.2)	403 (6.8)
Intermittent *S. aureus* n(%)	52 (16.1)	26 (12.9)	33 (16.6)	18 (14.9)	10 (10.5)	8 (16.7)	10 (13.7)	1171 (19.8)
Chronic *P. aeruginosa* n(%)	58 (18.0)	32 (15.9)	35 (17.6)	12 (9.9)	18 (18.9)	6 (12.5)	8 (11.0)	773 (13.1)
Intermittent *P. aeruginosa* n(%)	73 (22.6)	46 (22.9)	42 (21.1)	27 (22.3)	16 (16.8)	11 (22.9)	6 (8.2)	1108 (18.8)
B. cepacia complex n(%)	20 (6.2)	8 (4.0)	7 (3.5)	<5	<5	<5	<5	181 (3.1)
B. cenocepacia n(%)	5 (1.5)	<5	<5	<5	<5	<5	<5	53 (0.9)
B. multivorans n(%)	12 (3.7)	<5	<5	<5	<5	<5	<5	89 (1.5)
B. other cepacia n(%)	<5	<5	<5	<5	<5	<5	<5	22 (0.4)
MRSA n(%)	5 (1.5)	<5	<5	<5	<5	<5	<5	112 (1.9)
H. influenza n(%)	13 (4.0)	12 (6.0)	9 (4.5)	7 (5.8)	<5	<5	<5	415 (7.0)
NTM n(%)	15 (4.6)	5 (2.5)	8 (4.0)	<5	<5	<5	5 (6.8)	234 (4.0)
Aspergillus fumigatus n(%)	18 (5.6)	13 (6.5)	10 (5.0)	8 (6.6)	6 (6.3)	<5	10 (13.7)	-**

* Proportions are calculated from the number of people who were recorded as having at least one respiratory culture sample taken.
** Redacted to adhere to statistical disclosure guidelines.

1.18 Lung infections 2018–2023







1.19 Respiratory culture sample type

Overall	2018	2023
Number of people with an annual review (n)	9847	10344
Number of people with at least 3 samples of any type taken n(%)*	8540 (86.7)	6664 (64.4)
Number of people with at least 1 sample of any type taken n(%)*	9637 (97.9)	9590 (92.7)
Sample type[1]**		
Sputum; n(%)	6623 (68.7)	5429 (56.6)
Cough; n(%)	6112 (63.4)	6774 (70.6)
Bronchoalveolar lavage; n(%)	419 (4.3)	248 (2.6)
Age <16 years	**2018**	**2023**
Number of people with an annual review (n)	3894	3756
Number of people with at least 3 samples of any type taken n(%)*	3774 (96.9)	3483 (92.7)
Number of people with at least 1 sample of any type taken n(%)*	3872 (99.4)	3676 (97.9)
Sample type[1]**		
Sputum; n(%)	1563 (40.4)	1087 (29.6)
Cough; n(%)	3708 (95.8)	3613 (98.3)
Bronchoalveolar lavage; n(%)	295 (7.6)	191 (5.2)
Age ≥16 years	**2018**	**2023**
Number of people with an annual review (n)	5953	6588
Number of people with at least 3 samples of any type taken n(%)*	4766 (80.1)	3181 (48.3)
Number of people with at least 1 sample of any type taken n(%)*	5765 (96.8)	5914 (89.8)
Sample type[1]**		
Sputum; n(%)	5060 (87.8)	4342 (73.4)
Cough; n(%)	2404 (41.7)	3161 (53.4)
Bronchoalveolar lavage; n(%)	124 (2.2)	57 (1.0)

* % is of those people with an annual review.

** Patients can have more than one sample taken so the % total may not add up to 100%.

[1] Proportions are calculated from the number of people with at least 1 sample of any type taken.

1.20 Non-tuberculous mycobacteria (NTM) or atypical mycobacteria

Non-tuberculous mycobacterium is slow to grow and takes time to treat. It may be present for several years before eradication, or may never be cleared. In the table below, 'prevalence' represents all people reported in that year as having a positive culture. 'Incidence' represents all positive cultures in individuals that have not reported having any in the previous two years of data.

	2021	2022	2023
Number with annual review	**(n=10175)**	**(n=10251)**	**(n=10344)**
NTM Prevalence; n(%)	397 (3.9)	289 (3.1)	273 (2.6)
On NTM treatment in the given year; n (% of NTM prevalence in given year)	231 (58.1)	153 (52.9)	111 (40.7)
NTM Incidence[1]	154 (1.7)	147 (1.5)	144 (1.5)
M. abscessus prevalence	216 (2.1)	90 (0.9)	81 (0.8)
M. abscessus incidence[2]	58 (0.6)	29 (0.3)	32 (0.3)

[1] Proportion based on the number of patients with non-positive NTM tests in the previous two data years

[2] Proportion based on the number of patients with non-positive *M.abscessus* tests in the previous two data years

Complications

1.21 Complications in 2023

The number shown is for a complication that has been present in the preceding 12 months.

Complications	Overall	<16 years	≥16 years
Respiratory related			
Nasal polyps requiring surgery	394 (3.8)	90 (2.4)	304 (4.6)
Sinus disease	805 (7.8)	34 (0.9)	771 (11.7)
Asthma	596 (5.8)	85 (2.3)	511 (7.8)
ABPA	487 (4.7)	61 (1.6)	426 (6.5)
Any haemoptysis	-*	<5	136 (2.1)
Massive haemoptysis	11 (0.1)	0	11 (0.2)
Pneumothorax requiring chest tube	7 (0.1)	0	7 (0.1)
Cardiac complications			
Tachyarrhythmia	-*	<5	26 (0.4)
Bradycardia	-*	0	<5
Cardiac arrest	-*	0	<5
Cardiomyopathy	13 (0.1)	0	13 (0.2)
Congenital heart disease	23 (0.2)	12 (0.3)	11 (0.2)
Heart failure	16 (0.2)	0	16 (0.2)
Ischaemic heart disease	13 (0.1)	0	13 (0.2)
Valvular disease	14 (0.1)	5 (0.1)	9 (0.1)
Other	-*	<5	26 (0.4)
Pancreas and hepatobiliary disease			
Raised liver enzymes	1375 (13.3)	397 (10.6)	978 (14.8)
Liver disease	1888 (18.3)	350 (9.3)	1538 (23.3)
Cirrhosis with no portal hypertension	-*	<5	82 (1.2)
Cirrhosis with portal hypertension	140 (1.4)	17 (0.5)	123 (1.9)
Gall bladder disease requiring surgery	237 (2.3)	32 (0.9)	205 (3.1)
Pancreatitis	64 (0.6)	5 (0.1)	59 (0.9)
Upper gastrointestinal (GI)			
Gastro-oesophageal reflux disease (GORD)	1717 (16.6)	207 (5.5)	1510 (22.9)
Peptic ulcer	-*	0	<5
GI bleed (varices as source)	-*	<5	6 (0.1)
GI bleed (non varices as source)	-*	<5	9 (0.1)
Lower gastrointestinal			
Intestinal obstruction	38 (0.4)	14 (0.4)	24 (0.4)
DIOS	372 (3.6)	74 (2.0)	298 (4.5)
Fibrosing colonopathy / colonic stricture	-*	0	<5
Rectal prolapse	14 (0.1)	6 (0.2)	8 (0.1)
Renal			
Kidney stones	-*	<5	160 (2.4)
Renal failure	100 (1.0)	0	100 (1.5)
Musculoskeletal			
Arthritis	125 (1.2)	6 (0.2)	119 (1.8)
Arthropathy	241 (2.3)	6 (0.2)	235 (3.6)
Bone fracture	50 (0.5)	12 (0.3)	38 (0.6)
Osteopenia	1100 (10.6)	16 (0.4)	1084 (16.5)
Osteoporosis	-*	<5	539 (8.2)
Other			
Cancer confirmed by histology	-*	<5	33 (0.5)
Port inserted or replaced	143 (1.4)	41 (1.1)	102 (1.5)
Depression	463 (4.5)	8 (0.2)	455 (6.9)
Hearing loss	381 (3.7)	25 (0.7)	356 (5.4)
Hypertension	-*	<5	211 (3.2)
Urinary incontinence	700 (6.8)	54 (1.4)	646 (9.8)
Faecal incontinence	79 (0.8)	17 (0.5)	62 (0.9)
Postural anomaly	604 (5.8)	42 (1.1)	562 (8.5)

* Redacted to adhere to statistical disclosure guidelines.

1.22 Incidence of complications

The table below describes new cases of a complication that have not been reported for an individual in at least the previous two years.

	2022†			2023		
	Overall (n=10251)	<16 years (n=3806)	≥16 years (n=6445)	Overall (n=10344)	<16 years (n=3755)	≥16 years (n=6588)
ABPA	125 (1.2)	40 (1.1)	85 (1.3)	118 (1.1)	23 (0.6)	95 (1.4)
Cirrhosis - no portal hypertension	44 (0.4)	6 (0.2)	38 (0.6)	-*	<5	36 (0.5)
Cirrhosis - with portal hypertension	44 (0.4)	9 (0.2)	35 (0.5)	43 (0.4)	7 (0.2)	36 (0.5)
Cancer confirmed by histology	-*	<5	19 (0.3)	20 (0.2)	0	20 (0.3)

† 2022 figures have been updated since their publication in the 2022 Registry Report as a result of data cleaning.

1.23 CF diabetes**
N=8221

Cystic fibrosis diabetes (CFD) is common in adults and adolescents with cystic fibrosis. This is because, for many people with CF, the pancreas does not work properly. This can mean that not enough insulin is produced, or it may not work properly, causing CFD. CFD is different from type 1 and type 2 diabetes, but has features of both.

	All ≥10 years (n=8221)	10-15 years (n=1633)	≥16 years (n=6588)
On CFD treatment; n(%)			
Of those on treatment	2353 (28.6)	133 (8.1)	2220 (33.7)
Insulin[1]; n(%)	1940 (82.4)	128 (96.2)	1812 (81.6)
CFD Screening ; n(%)			
Yes	3779 (46.0)	1092 (66.9)	2687 (40.8)
Screening Type			
Continuous glucose monitoring[2]; n(%)	1204 (31.9)	364 (33.3)	840 (31.3)
Oral glucose tolerance test[2]; n(%)	1652 (43.7)	521 (47.7)	1131 (42.1)
Not screened (other)	2324 (28.3)	74 (4.5)	2250 (34.2)
Not screened (known CFD)	2001 (24.3)	406 (24.9)	1595 (24.2)
Unknown	117 (1.4)	61 (3.7)	56 (0.9)

[1] Proportion of patients on treatment.
[2] Proportion of patients screened.
* Redacted to adhere to statistical disclosure guidelines.
** Alternatively known as CF related diabetes.

Antibiotics

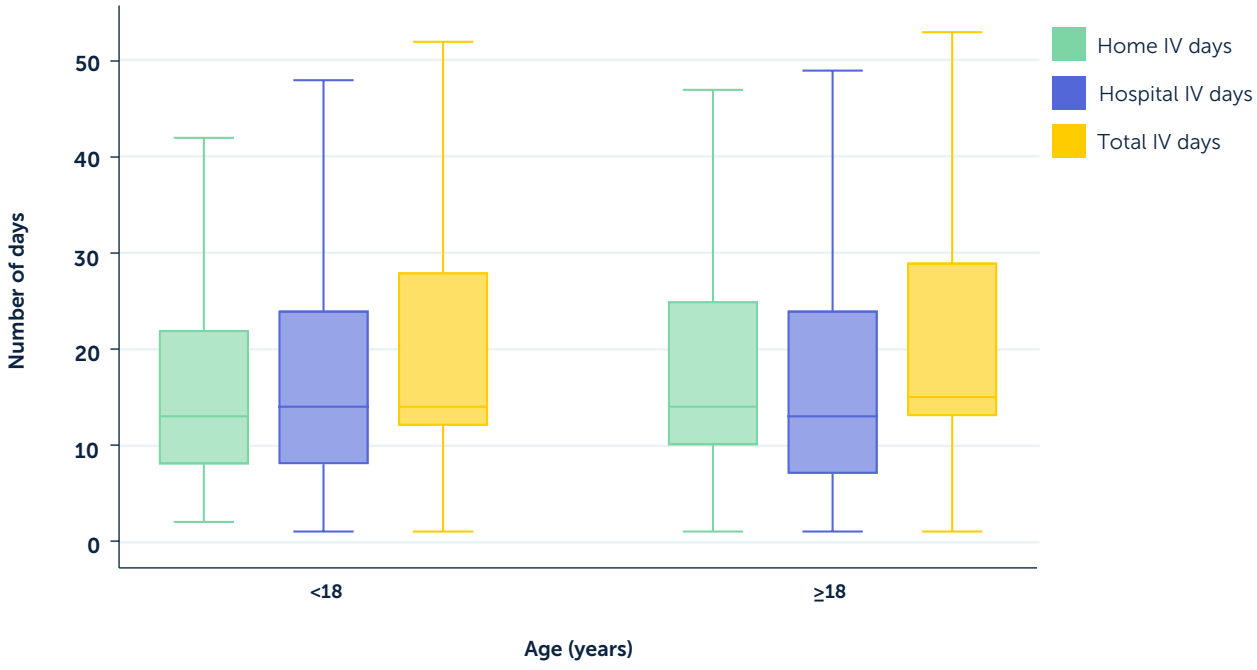
1.24 Intravenous (IV) antibiotics
N=10344

When someone with CF becomes unwell with an infection, they might be prescribed intravenous (IV) antibiotics. IV antibiotics are given to the patient through their veins. This treatment can take a number of days and may take place as a hospital inpatient, or at home.

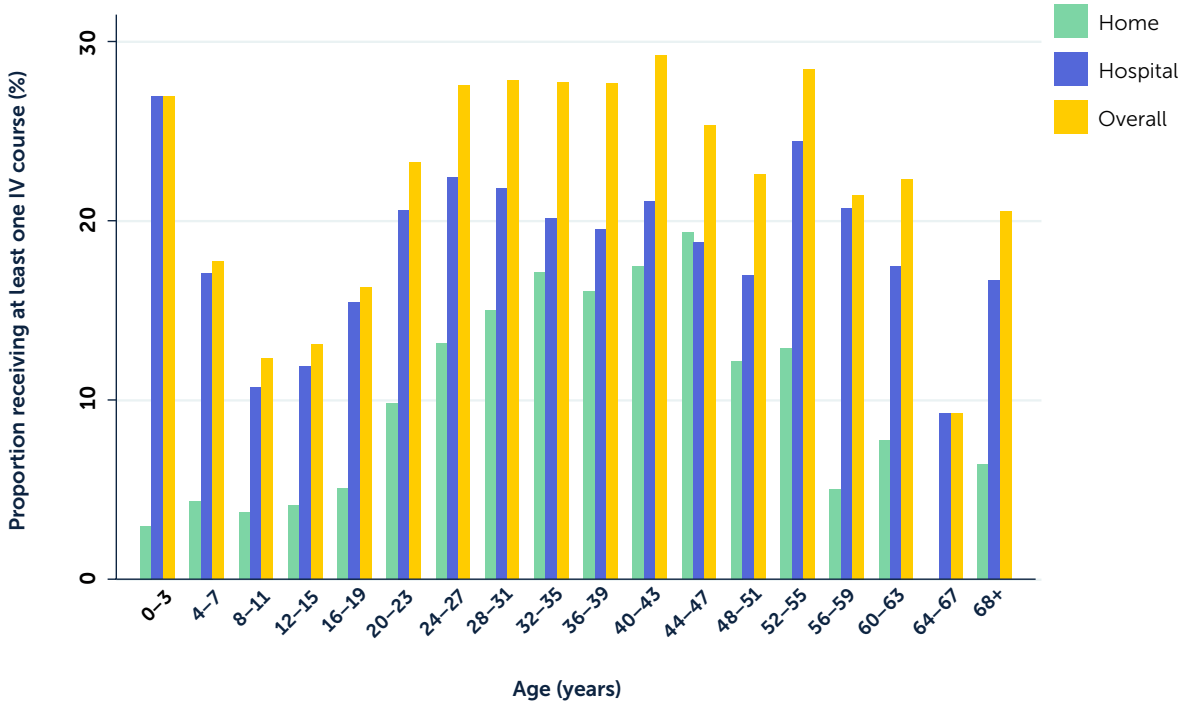
		Home		Hospital		Total	
Age	n	Patients n(%)	Median days (IQR)	Patients n(%)	Median days (IQR)	Patients n(%)	Median days (IQR)
0-3	676	20 (3.0)	8 (6-13)	182 (26.9)	13 (7-17)	182 (26.9)	14 (9-17)
4-7	942	41 (4.4)	13 (7-22)	161 (17.1)	14 (10-19)	167 (17.7)	14 (13-27)
8-11	1053	39 (3.7)	14 (10-20)	113 (10.7)	14 (8-22)	130 (12.3)	14 (13-28)
12-15	1085	45 (4.1)	14 (8-26)	129 (11.9)	14 (10-36)	142 (13.1)	21 (14-40)
16-19	866	44 (5.1)	11 (7-20)	134 (15.5)	13 (7-25)	141 (16.3)	14 (11-28)
20-23	796	78 (9.8)	14 (9-21)	164 (20.6)	12 (8-26)	185 (23.2)	14 (12-28)
24-27	918	121 (13.2)	14 (11-25)	206 (22.4)	13 (7-22)	253 (27.6)	15 (14-29)
28-31	848	127 (15.0)	14 (11-22)	185 (21.8)	14 (7-23)	236 (27.8)	15 (14-28)
32-35	800	137 (17.1)	14 (11-23)	161 (20.1)	13 (7-24)	222 (27.8)	14 (14-30)
36-39	636	102 (16.0)	14 (11-28)	124 (19.5)	14 (7-28)	176 (27.7)	18 (14-32)
40-43	527	92 (17.5)	14 (14-32)	111 (21.1)	12 (7-20)	154 (29.2)	16 (14-35)
44-47	367	71 (19.3)	14 (10-23)	69 (18.8)	12 (6-24)	93 (25.3)	15 (14-41)
48-51	230	28 (12.2)	13 (7-16)	39 (17.0)	13 (7-21)	52 (22.6)	14 (13-28)
52-55	225	29 (12.9)	14 (10-27)	55 (24.4)	11 (7-23)	64 (28.4)	14 (13-28)
56-59	140	7 (5.0)	8 (5-21)	29 (20.7)	15 (10-26)	30 (21.4)	16 (14-29)
60-63	103	8 (7.8)	14 (9-27)	18 (17.5)	16 (14-26)	23 (22.3)	20 (14-27)
64-67	54	0 (0.0)	-	5 (9.3)	14 (11-19)	5 (9.3)	14 (11-19)
68+	78	5 (6.4)	14 (14-14)	13 (16.7)	20 (12-24)	16 (20.5)	16 (12-28)
<16	3756	145 (3.9)	14 (8-21)	585 (15.6)	14 (8-23)	621 (16.5)	14 (12-28)
≥16	6588	849 (12.9)	14 (10-25)	1313 (19.9)	13 (7-24)	1650 (25.0)	15 (13-29)
<18	4181	165 (3.9)	13 (8-22)	653 (15.6)	14 (8-24)	691 (16.5)	14 (12-28)
≥18	6163	829 (13.5)	14 (10-25)	1245 (20.2)	13 (7-24)	1580 (25.6)	15 (13-29)
Overall	**10344**	**994 (9.6)**	**14 (10-24)**	**1898 (18.3)**	**14 (8-24)**	**2271 (22.0)**	**14 (13-29)**

* Redacted to adhere to statistical disclosure guidelines.

This box plot graph illustrates the spread of the number of days on IV antibiotics in the UK CF population, stratified by age. A guide on how to correctly interpret this box plot graph can be found on page 56.



The bar graph below summarises the proportion of people receiving at least one course of IV antibiotics across different age groups within the UK CF population. Overall, the proportion of patients receiving at least one IV course at home was 9.6% and in hospital was 18.3%. The proportion receiving any IVs was 22.0%.



1.25 Inhaled antibiotic use
N=10344

	2023		
	Overall	<16 years	≥16 years
Number of patients	10344	3756	6588
Tobramycin solution; n(%)	965 (9.3)	219 (5.8)	746 (11.3)
Other aminoglycoside; n(%)	44 (0.4)	5 (0.1)	39 (0.6)
Colistin; n(%)	1196 (11.6)	422 (11.2)	774 (11.7)
Promixin; n(%)	1363 (13.2)	315 (8.4)	1048 (15.9)
Aztreonam; n(%)	759 (7.3)	50 (1.3)	709 (10.8)
Colistimethate (DPI); n(%)	892 (8.6)	55 (1.5)	837 (12.7)
Tobramycin Inhalation Powder; n(%)	656 (6.3)	18 (0.5)	638 (9.7)
Levofloxacin; n(%)	-*	<5	70 (1.1)
At least one of the above; n(%)	4189 (40.5)	823 (21.9)	3366 (51.1)

1.26 Inhaled antibiotic use among people with chronic *Pseudomonas aeruginosa*

The consensus view in the UK is that 90% of people chronically infected with *P. aeruginosa* should be prescribed at least one of the above inhaled antibiotics.

	2013			2018			2023		
	Overall	<16 years	≥16 years	Overall	<16 years	≥16 years	Overall	<16 years	≥16 years
Patients with chronic *P. aeruginosa*	2960	329	2631	2611	229	2382	871	98	773
Tobramycin solution; n(%)	929 (31.4)	103 (31.3)	826 (31.4)	638 (24.4)	81 (35.4)	557 (23.4)	184 (21.1)	29 (29.6)	155 (20.1)
Other aminoglycoside; n(%)	108 (3.6)	13 (4.0)	95 (3.6)	-*	<5	40 (1.7)	-*	<5	5 (0.6)
Colistin; n(%)	1173 (39.6)	176 (53.5)	997 (37.9)	647 (24.8)	91 (39.7)	556 (23.3)	208 (23.9)	44 (44.9)	164 (21.2)
Promixin; n(%)	881 (29.8)	140 (42.6)	741 (28.2)	797 (30.5)	103 (45.0)	694 (29.1)	167 (19.2)	33 (33.7)	134 (17.3)
Aztreonam; n(%)	201 (6.8)	<5	199 (7.6)	645 (24.7)	15 (6.6)	630 (26.4)	211 (24.2)	9 (9.2)	202 (26.1)
Colistimethate (DPI); n(%)	-	-	-	448 (17.2)	13 (5.7)	435 (18.3)	164 (18.8)	6 (6.1)	158 (20.4)
Tobramycin Inhalation Powder; n(%)	-	-	-	635 (24.3)	17 (7.4)	618 (25.9)	-*	<5	100 (12.9)
Levofloxacin; n(%)	-	-	-	-	-	-	30 (3.4)	0	30 (3.9)
At least one of the above; n(%)	**2368 (80.0)**	**302 (91.8)**	**2066 (78.5)**	**2322 (88.9)**	**206 (90.0)**	**2116 (88.8)**	**716 (82.2)**	**87 (88.8)**	**629 (81.4)**

* Redacted to adhere to statistical disclosure guidelines.

1.27 Long-term azithromycin use

Azithromycin is an antibiotic with some anti-inflammatory properties. It is recommended for long-term use as a prophylactic antibiotic in people with chronic *Pseudomonas aeruginosa*.

		Number of patients on azithromycin; n	Patients with chronic *P. aeruginosa*; n(%)	Patients without chronic *P. aeruginosa*; n(%)
2013	Overall	3619	2022 (55.9)	1597 (44.1)
	0-3 years	-*	<5	25 (92.6)
	4-15 years	620	141 (22.7)	479 (77.3)
	≥ 16 years	2972	1879 (63.2)	1093 (36.8)
2018	Overall	4111	1794 (43.6)	2317 (56.4)
	0-3 years	-*	<5	48 (94.1)
	4-15 years	657	104 (15.8)	553 (84.2)
	≥ 16 years	3403	1687 (49.6)	1716 (50.4)
2023	Overall	3459	532 (15.4)	2927 (84.6)
	0-3 years	50	7 (14.0)	43 (86.0)
	4-15 years	415	29 (7.0)	386 (93.0)
	≥ 16 years	2994	496 (16.6)	2498 (83.4)

1.28 Prophylactic flucloxacillin use**

Flucloxacillin is an antibiotic that is used prophylactically to prevent infection with bacteria.

	2018		2023	
Age	Total patients	Patients on prophylactic flucloxacillin; n(%)	Total patients	Patients on prophylactic flucloxacillin; n(%)
0-3	839	516 (61.5)	676	318 (47.0)
4-7	1093	292 (26.7)	942	237 (25.2)
8-11	1059	267 (25.2)	1053	208 (19.8)
12-15	903	222 (24.6)	1085	211 (19.4)
16-19	865	174 (20.1)	866	115 (13.3)
20-23	986	103 (10.4)	796	100 (12.6)
24-27	906	65 (7.2)	918	71 (7.7)
28-31	833	59 (7.1)	848	48 (5.7)
32-35	653	44 (6.7)	800	41 (5.1)
36-39	537	38 (7.1)	636	17 (2.7)
40-43	330	25 (7.6)	527	20 (3.8)
44-47	267	16 (6.0)	367	14 (3.8)
48-51	210	15 (7.1)	230	10 (4.3)
52-55	147	6 (4.1)	225	10 (4.4)
56-59	88	5 (5.7)	140	11 (7.9)
60-63	55	<5	103	<5
64-67	30	<5	54	<5
68+	46	<5	78	<5
<16 years	3894	1297 (33.3)	3756	974 (25.9)
≥16 years	5953	556 (9.3)	6588	463 (7.0)
<18 years	4313	1391 (32.3)	4181	1029 (24.6)
≥18 years	5534	462 (8.3)	6163	408 (6.6)
Overall	**9847**	**1853 (18.8)**	**10344**	**1437 (13.9)**

* Redacted to adhere to statistical disclosure guidelines.

** Data incudes patients that have been recruited and randomised for the CF START trial in 2018 and 2023

Bronchodilators and corticosteroids

1.29 Inhaled bronchodilators and corticosteroids

Age	Total patients	Patients on inhaled bronchodilators; n(%)	Patients on inhaled corticosteroids; n(%)	Patients on inhaled combination corticosteroids/ bronchodilators; n(%)
<6 years	1122	273 (24.3)	101 (9.0)	12 (1.1)
6 - ≤16 years	2883	1354 (47.0)	404 (14.0)	333 (11.6)
6 - ≤18 years	3276	1595 (48.7)	457 (13.9)	409 (12.5)
<16 years	3756	1478 (39.4)	471 (12.5)	299 (8.0)
≥16 years	6588	4510 (68.5)	1125 (17.1)	1887 (28.6)
<18 years	4181	1726 (41.3)	526 (12.6)	386 (9.2)
≥18 years	6163	4262 (69.2)	1070 (17.4)	1800 (29.2)
Overall	**10344**	**5988 (57.9)**	**1596 (15.4)**	**2186 (21.1)**

* Redacted to adhere to statistical disclosure guidelines.

Muco-active therapies

1.30 Mannitol

	2018		2023	
Age	Total patients	Patients on Mannitol; n (%)	Total patients	Patients on Mannitol; n (%)
<16 years	3894	<5	3756	<5
≥16 years	5953	336 (5.6)	6588	223 (3.4)
<18 years	4313	<5	4181	<5
≥18 years	5534	334 (6.0)	6163	222 (3.6)
Overall	**9847**	**-***	**10344**	**_***

1.31 DNase**

	2013		2018		2023	
Age	Total patients	Patients on DNase; n(%)	Total patients	Patients on DNase; n(%)	Total patients	Patients on DNase; n(%)
0-3	981	100 (10.2)	840	153 (18.2)	676	157 (23.2)
4-7	1004	332 (33.1)	1093	576 (52.7)	942	479 (50.8)
8-11	899	496 (55.2)	1059	825 (77.9)	1053	803 (76.3)
12-15	955	627 (65.7)	903	757 (83.8)	1085	828 (76.3)
16-19	1005	635 (63.2)	865	701 (81.0)	866	644 (74.4)
20-23	994	625 (62.9)	986	759 (77.0)	796	536 (67.3)
24-27	836	537 (64.2)	906	642 (70.9)	918	591 (64.4)
28-31	703	413 (58.7)	833	585 (70.2)	848	523 (61.7)
32-35	503	283 (56.3)	653	418 (64.0)	800	480 (60.0)
36-39	315	157 (49.8)	537	343 (63.9)	636	384 (60.4)
40-43	294	141 (48.0)	330	202 (61.2)	527	287 (54.5)
44-47	213	102 (47.9)	267	150 (56.2)	367	203 (55.3)
48-51	152	79 (52.0)	209	107 (51.2)	230	118 (51.3)
52-55	76	32 (42.1)	147	77 (52.4)	225	118 (52.4)
56-59	48	24 (50.0)	88	48 (54.5)	140	74 (52.9)
60-63	29	12 (41.4)	55	27 (49.1)	103	57 (55.3)
64-67	22	10 (45.5)	30	19 (63.3)	54	31 (57.4)
68+	23	10 (43.5)	46	19 (41.3)	78	39 (50.0)
<16 years	3839	1555 (40.5)	3895	2311 (59.3)	3756	2267 (60.4)
≥16 years	5213	3060 (58.7)	5952	4097 (68.8)	6588	4085 (62.0)
<18 years	4354	1891 (43.4)	4314	2650 (61.4)	4181	2601 (62.2)
≥18 years	4698	2724 (58.0)	5533	3758 (67.9)	6163	3751 (60.9)
Overall	**9052**	**4615 (51.0)**	**9847**	**6408 (65.1)**	**10344**	**6352 (61.4)**

** CF STORM was enrolling patients throughout the 2023 data collection year.

1.32 Hypertonic saline

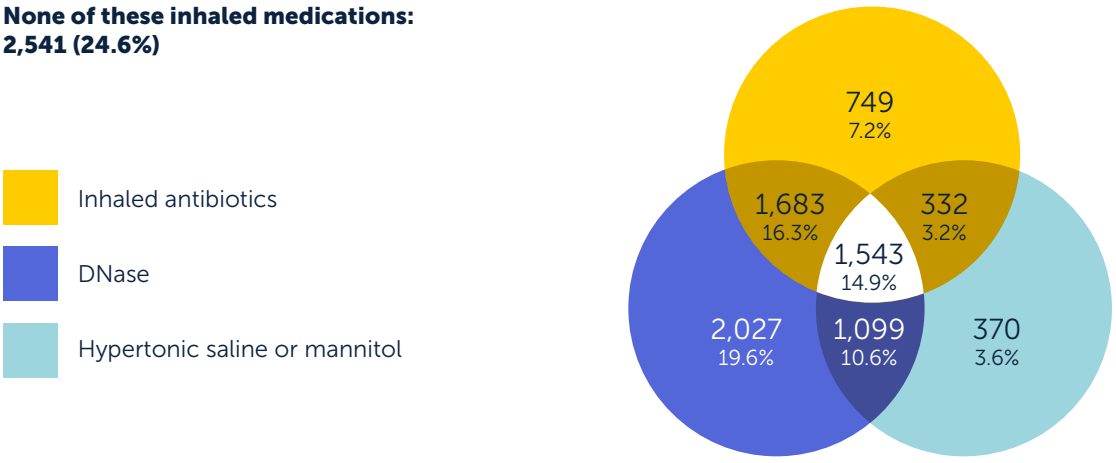
This treatment helps to thin mucus so that it is easier to cough out of the body.

	2013		2018		2023	
Age	Total patients	Patients on hypertonic saline; n(%)	Total patients	Patients on hypertonic saline; n(%)	Total patients	Patients on hypertonic saline; n(%)
0-3	981	49 (5.0)	839	79 (9.4)	676	135 (20.0)
4-7	1004	157 (15.6)	1093	288 (26.3)	942	310 (32.9)
8-11	899	225 (25.0)	1059	386 (36.4)	1053	381 (36.2)
12-15	955	303 (31.7)	903	418 (46.3)	1085	441 (40.6)
16-19	1005	287 (28.6)	865	408 (47.2)	866	315 (36.4)
20-23	994	263 (26.5)	986	350 (35.5)	796	307 (38.6)
24-27	836	220 (26.3)	906	275 (30.4)	918	268 (29.2)
28-31	703	206 (29.3)	833	263 (31.6)	848	226 (26.7)
32-35	503	131 (26.0)	653	238 (36.4)	800	182 (22.8)
36-39	315	76 (24.1)	537	188 (35.0)	636	170 (26.7)
40-43	294	64 (21.8)	330	110 (33.3)	527	139 (26.4)
44-47	213	50 (23.5)	267	81 (30.3)	367	88 (24.0)
48-51	152	35 (23.0)	210	52 (24.8)	230	57 (24.8)
52-55	76	23 (30.3)	147	41 (27.9)	225	60 (26.7)
56-59	48	9 (18.8)	88	27 (30.7)	140	29 (20.7)
60-63	29	6 (20.7)	55	10 (18.2)	103	28 (27.2)
64-67	22	8 (36.4)	30	7 (23.3)	54	12 (22.2)
68+	23	5 (21.7)	46	17 (37.0)	78	23 (29.5)
<16 years	3839	734 (19.1)	3894	1171 (30.1)	3756	1267 (33.7)
≥16 years	5213	1383 (26.5)	5953	2067 (34.7)	6588	1904 (28.9)
<18 years	4354	879 (20.2)	4313	1386 (32.1)	4181	1437 (34.4)
≥18 years	4698	1238 (26.4)	5534	1852 (33.5)	6163	1734 (28.1)
Overall	**9052**	**2117 (23.4)**	**9847**	**3238 (32.9)**	**10344**	**3171 (30.7)**

1.33 Burden of treatment*

The Venn diagram shows how many people with CF are on one or more inhaled therapy and the combinations they take.

None of these inhaled medications: 2,541 (24.6%)



* As a result of data cleaning these figures differ slightly from the Registry Highlights report published in June 2023

CFTR modulators

During 2023 the CFTR modulators (CFTRm) were available to people with cystic fibrosis under a managed access agreement.

Ivacaftor

In 2023 ivacaftor had approval for use in people aged 4 months and older with at least one copy of a CFTR "gating" variant and for people with the R117H variant.

Lumacaftor/ivacaftor

Lumacaftor / ivacaftor is licensed for use in the UK for people with CF aged one and over with two copies of the F508del variant.

Tezacaftor/ivacaftor

Tezacaftor / ivacaftor is licensed for use in people with CF aged six years and over who have two copies of the F508del variant and, or a single copy of F508del and one of 14 "residual" function variants.

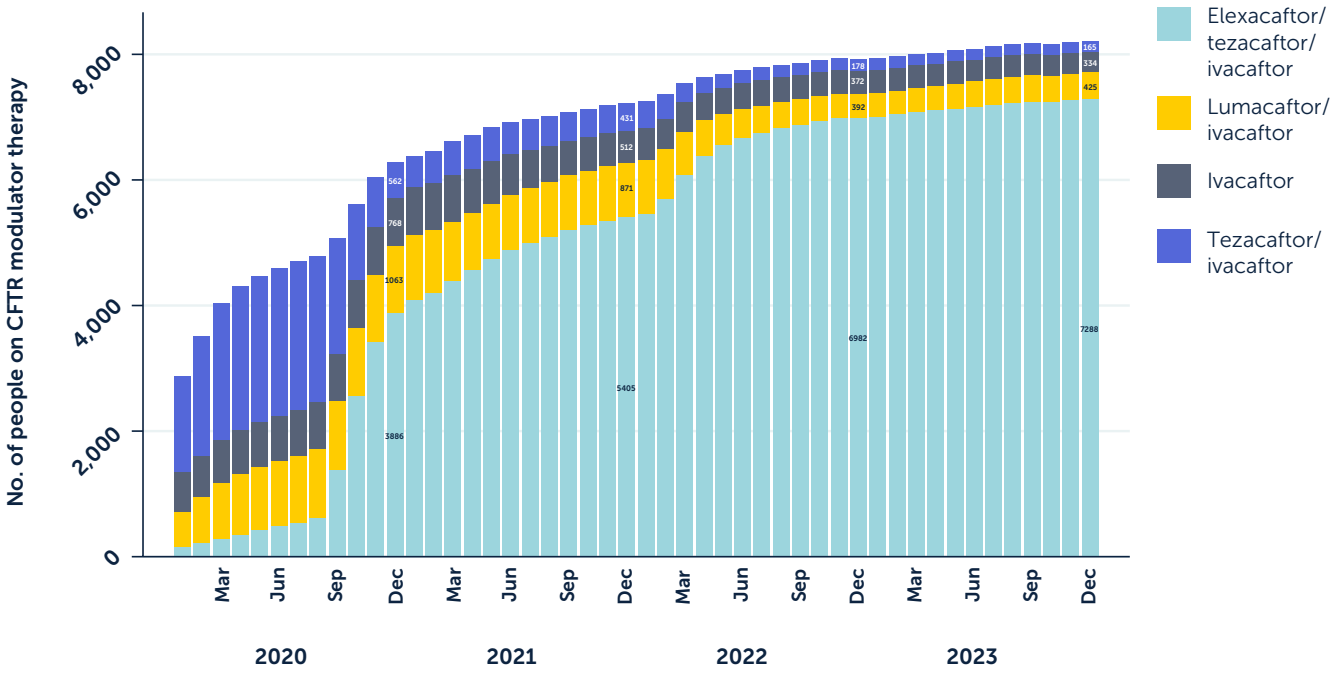
Elexacaftor/tezacaftor/ivacaftor

Elexacaftor/ tezacaftor/ ivacaftor was available for people with CF aged 6 years and over who have two copies of the F508del variant, or a single copy of F508del and one minimal function variant. In November 2023, this was extended to include children aged 2 and over. NHS commissioning statements adopted across the UK support the prescribing of CFTR modulators "off label"; the arrangement varies slightly across devolved nations but covers the 177 CFTR variants on an approved "FDA list", and in some devolved nations, responsive genotypes identified through the "French Compassionate Use Programme".

Access arrangements for the CFTR modulators prior to 2023 can be found in previous annual reports and on our website here: **cysticfibrosis.org.uk/treatmentsandmedication**

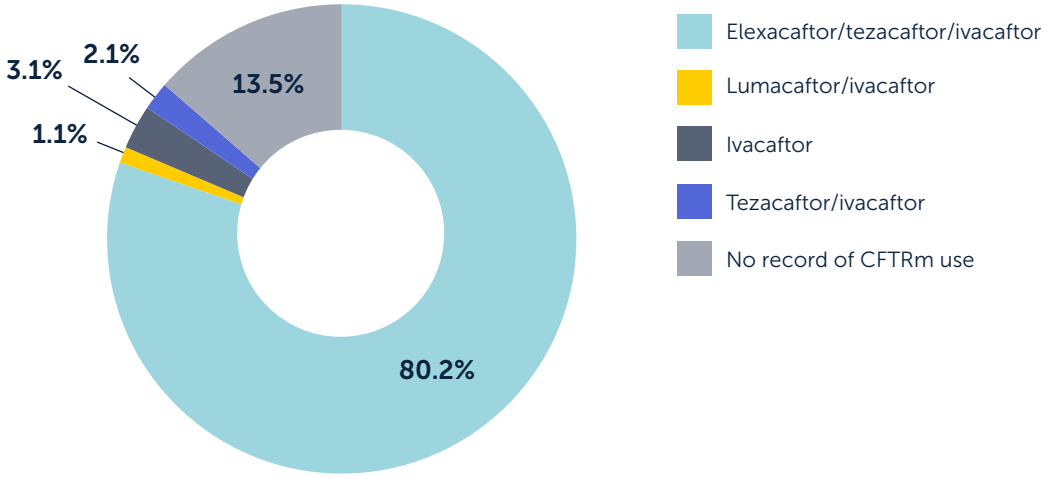
1.34 CFTR modulator use 2020–2023

The graph below shows the number of people taking each drug by month. Where people switched modulators, the most recent prescription is counted. Only patients who had an annual review are included. By December 2023, 8,212 people were taking a CFTR modulator.



1.35a CFTR modulator use in all people aged six years and older[1] N=9316

The chart shows the distribution of CFTR modulators taken in those aged 6 or over as of 31/12/2023. The last used CFTR modulator as of 31/12/2023 is shown in the chart. 13.5% of people aged 6 and over had no record of any CFTRm use on the Registry.

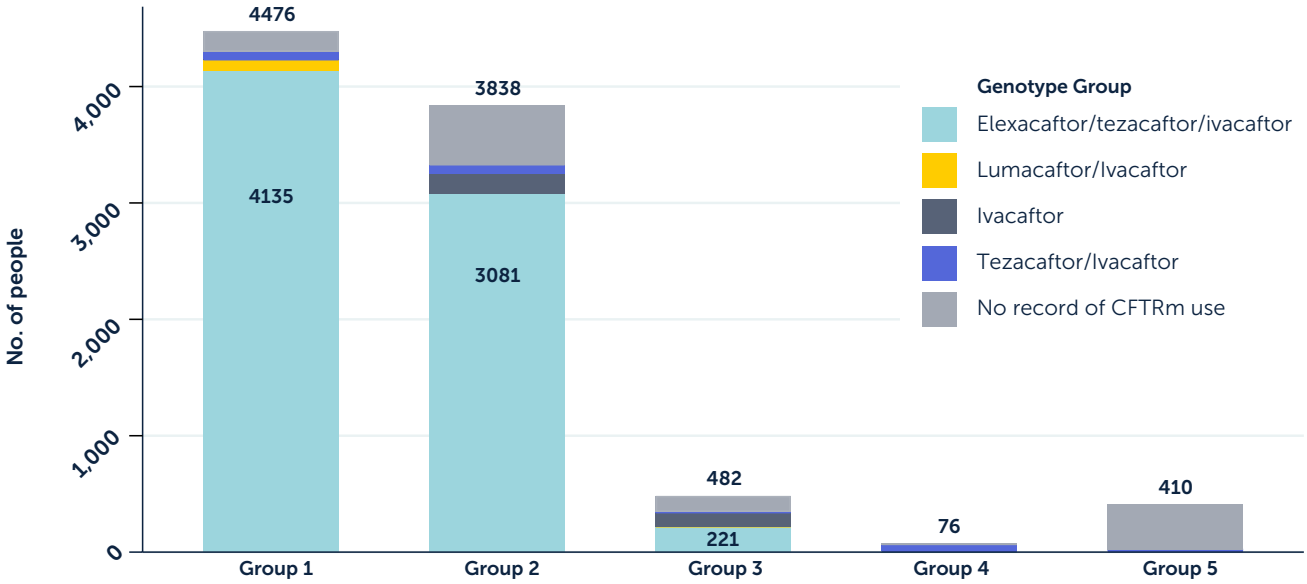


[1] 7 patients were excluded because their last recorded CFTRm treatment was as part of drug trial and specific drug was unknown

1.35b CFTR modulator use in all people aged six years and older by genotype group [1,2,3]
N=9316

The chart below shows the most recent CFTR modulator recorded for people aged six and older and according to their genotype group as defined in the table below. The full list of CFTR variants included within these groups can be found in appendix 4.

Some people with a potentially responsive variant (Groups 1 to 4) have no recorded use of any CFTR modulator on the Registry. There could be several reasons for this: for example, a person may have had a transplant or there could be a data entry error. There are a small number of people[3] thought to have non-responsive genotypes (group 5) who are currently taking or have previously taken a modulator. It is not possible to tell from the Registry record why they received a CFTR modulator.



Genotype Group Definitions	
Group 1	F508del Homozygous
Group 2	F508del Heterozygous
Group 3	no F508del, but ETI* responsive variant as defined by FDA list (4a**) or French Compassionate Use Programme list (4b**)
Group 4	no F508del, no ETI* responsive variant as defined by FDA list (4a**) or French Compassionate Use Programme list (4b**), but at least 1 variant from lists 4c or 4d**
Group 5	no F508del, no ETI* responsive variant as defined by FDA list (4a**) or French Compassionate Use Programme list (4b**), no variant from lists 4c** or 4d**

[1] 7 patients were excluded because their last recorded CFTRm treatment was as part of a drug trial and specific drug was unknown

[2] 27 people were excluded because their genotype is missing or unknown

[3] for numbers and % details see Appendix 4 table 4e

* ETI is Elexacaftor/Tezacaftor/ivacaftor

** see Appendix 4 for lists of relevant variants

1.36 Demographic characteristics for people aged six years and older, by genotype group and CFTR modulator use[1,2]

	All potential responders*		Likely non-responders**[6]
	CFTRm use recorded[3]	no record of CFTRm use[3]	no record of CFTRm use[3]
Number of Individuals (n)	8031	841	390
Male n (%)	4312 (53.69)	420 (49.94)	214 (54.87)
Ethnicity[4]			
White n (%)	7711 (96.6)	782 (94.8)	233 (60.8)
Asian n (%)	131 (1.6)	22 (2.7)	120 (31.3)
Black n (%)	14 (0.2)	<5	9 (2.4)
Mixed n (%)	65 (0.8)	10 (1.2)	9 (2.4)
Other n (%)	59 (0.8)	-***	12 (3.1)
Age (years)[5]			
Mean (sd)	27 (14)	31 (17)	27 (16)
Median (IQR)	25 (15,36)	31 (14, 43)	24 (15,35)

[1] 7 patients were excluded because their last recorded CFTRm treatment was as part of a drug trial and specific drug was unknown

[2] 27 people were excluded because their genotype is missing or unknown

[3] Record / no record of CFTRm use defined as if record / no record as of 31/12/2023

[4] Reported ethnicty percentages are out of those with known ethnicity. 74 people had missing or unknown ethnicity

[5] Age at 31/12/2023

[6] Details for 20 people considered non-responders who had a record of CFTRm use are not included.

* Defined at least one F508del or ETI responsive variant as defined by FDA list (4a) or French Compassionate Use list (4b) or variant from lists 4c or 4d

** Defined as no F508del, no ETI responsive variant as defined by FDA list (4a) or French Compassionate Use list (4b) , no variant from lists 4c or 4d.

*** Redacted to adhere to statistical disclosure guidelines

Physiotherapy

1.37 Primary airway clearance technique

Physiotherapy helps people with CF clear sticky mucus from their lungs. The listed techniques represent the primary form of physiotherapy recorded for an individual on the Registry. One primary airway clearance technique can be recorded for an individual.

	Overall (n=10344)	<16 years (n=3756)	≥16 years (n=6588)	<18 years (n=4181)	≥18 years (n=6163)
Active cycle of breathing techniques	527 (5.1)	43 (1.1)	484 (7.3)	48 (1.1)	479 (7.8)
Assisted autogenic drainage	120 (1.2)	20 (0.5)	100 (1.5)	25 (0.6)	95 (1.5)
Autogenic drainage	901 (8.7)	33 (0.9)	868 (13.2)	49 (1.2)	852 (13.8)
Exercise	1812 (17.5)	500 (13.3)	1312 (19.9)	569 (13.6)	1243 (20.2)
Forced expiration	66 (0.6)	13 (0.3)	53 (0.8)	13 (0.3)	53 (0.9)
High Pressure PEP	35 (0.3)	22 (0.6)	13 (0.2)	23 (0.6)	12 (0.2)
Incentive spirometer	-*	<5	9 (0.1)	<5	9 (0.1)
Manual in/ex-sufflation (cough assist)	-*	<5	13 (0.2)	<5	12 (0.2)
Manual techniques (percussion over pressures vibrations)	221 (2.1)	160 (4.3)	61 (0.9)	161 (3.9)	60 (1.0)
NIV (non-invasive ventilation)	-*	<5	33 (0.5)	<5	32 (0.5)
Oscillating PEP	2804 (27.1)	1315 (35.0)	1489 (22.6)	1499 (35.9)	1305 (21.2)
PEP	1755 (17.0)	1043 (27.8)	712 (10.8)	1115 (26.7)	640 (10.4)
Postural drainage	175 (1.7)	149 (4.0)	26 (0.4)	150 (3.6)	25 (0.4)
VEST	45 (0.4)	17 (0.5)	28 (0.4)	19 (0.5)	26 (0.4)
Other	468 (4.5)	269 (7.2)	199 (3.0)	293 (7.0)	175 (2.8)
None	1356 (13.1)	168 (4.5)	1188 (18.0)	211 (5.0)	1145 (18.6)

1.38 Primary or secondary airway clearance technique

People with CF may receive more than one airway clearance technique. These techniques are not mutually exclusive and represent both primary and secondary forms of physiotherapy received by people with CF.

	Overall (n=10344)	<16 years (n=3756)	≥16 years (n=6588)	<18 years (n=4181)	≥18 years (n=6163)
Active cycle of breathing techniques	1115 (10.8)	222 (5.9)	893 (13.6)	252 (6.0)	863 (14.0)
Assisted autogenic drainage	174 (1.7)	44 (1.2)	130 (2.0)	50 (1.2)	124 (2.0)
Autogenic drainage	1535 (14.8)	86 (2.3)	1449 (22.0)	116 (2.8)	1419 (23.0)
Exercise; of which	6091 (58.9)	2472 (65.8)	3619 (54.9)	2754 (65.9)	3337 (54.1)
Exercise listed as only airway clearance technique	1001 (9.7)	157 (4.2)	844 (12.8)	187 (4.5)	814 (13.2)
Forced expiration	581 (5.6)	334 (8.9)	247 (3.7)	363 (8.7)	218 (3.5)
High Pressure PEP	44 (0.4)	28 (0.7)	16 (0.2)	29 (0.7)	15 (0.2)
Incentive spirometer	30 (0.3)	6 (0.2)	24 (0.4)	6 (0.1)	24 (0.4)
Manual in/ex-sufflation (cough assist)	-*	<5	26 (0.4)	6 (0.1)	24 (0.4)
Manual techniques (percussion over pressures vibrations)	480 (4.6)	336 (8.9)	144 (2.2)	342 (8.2)	138 (2.2)
NIV (non-invasive ventilation)	-*	<5	80 (1.2)	5 (0.1)	76 (1.2)
Oscillating PEP	3830 (37.0)	1663 (44.3)	2167 (32.9)	1903 (45.5)	1927 (31.3)
PEP	2371 (22.9)	1345 (35.8)	1026 (15.6)	1431 (34.2)	940 (15.3)
Postural drainage	465 (4.5)	373 (9.9)	92 (1.4)	382 (9.1)	83 (1.3)
VEST	99 (1.0)	43 (1.1)	56 (0.9)	49 (1.2)	50 (0.8)
Other	1213 (11.7)	688 (18.3)	525 (8.0)	733 (17.5)	480 (7.8)

1.39 Exercise testing

Exercise testing provides valuable information on an individual's physical abilities which gives insights into prognosis or oxygen requirements. Physiotherapists and exercise specialists use test results to individualise exercise programmes and target specific needs. Results of exercise tests can also be motivating to the individual and can be used to set future exercise goals.

Exercise test	Overall (n=10344)	<16 years (n=3756)	≥16 years (n=6588)	<18 years (n=4181)	≥18 years (n=6163)
Yes*	1066 (10.3)	318 (8.5)	748 (11.4)	372 (8.9)	694 (11.3)
No	7186 (69.5)	2447 (65.1)	4739 (71.9)	2699 (64.6)	4487 (72.8)
Not known or missing	2092 (20.2)	991 (26.4)	1101 (16.7)	1110 (26.5)	982 (15.9)
Type of exercise test[1,2]					
CPET	183 (17.2)	80 (25.2)	103 (13.8)	88 (23.7)	95 (13.7)
Shuttle test	135 (12.7)	89 (28.0)	46 (6.1)	97 (26.1)	38 (5.5)
Step test	213 (20.0)	53 (16.7)	160 (21.4)	58 (15.6)	155 (22.3)
6 minute walk test	93 (8.7)	8 (2.5)	85 (11.4)	10 (2.7)	83 (12.0)
Other	271 (25.4)	75 (23.6)	196 (26.2)	95 (25.5)	176 (25.4)
Missing	332 (31.1)	94 (29.6)	238 (31.8)	110 (29.6)	222 (32.0)

* Exercise test represents all types of testing listed including Cardiopulmonary Exercise Test (CPET), shuttle test, 6 minute walk test, step test and other test.

[1] Proportion of patients who answered Yes above.

[2] More than one type of test can be recorded so % total may not sum to 100%.

Other therapies

1.40 Oxygen and non-invasive ventilation

	Overall (n=10344)	<16 years (n=3756)	≥16 years (n=6588)	<18 years (n=4181)	≥18 years (n=6163)
Non invasive ventilation (NIV); n (%)	130 (1.3)	19 (0.5)	111 (1.7)	21 (0.5)	109 (1.8)
Any oxygen use; n (%)	319 (3.1)	47 (1.3)	272 (4.1)	49 (1.2)	270 (4.4)
Among those who had oxygen use:					
Continuously	-*	<5	41 (15.1)	<5	41 (15.2)
Nocturnal or with exertion	-*	<5	111 (40.8)	<5	110 (40.7)
As required (PRN)	-*	<5	34 (12.5)	<5	34 (12.6)
With exacerbation	125 (39.2)	39 (83.0)	86 (31.6)	40 (81.6)	85 (31.5)

1.41 Transplants

Lung transplantation has been available to people with CF for almost 30 years. Today the most common operation carried out is a double lung transplant, or bilateral sequential lung transplant. The following table gives information about transplant activity over time.

	2018	2019	2020	2021	2022	2023
Number evaluated	247	241	175	78	41	37
Number accepted	104	96	66	23	22	21
Number receiving aged <16 years	<5	<5	0	0	0	0
Bilateral lung	0	<5	0	0	0	0
Liver	<5	<5	0	0	0	0
Other	0	0	0	0	0	0
Number receiving aged 16+ years	63	54	15	5	6	<5
Bilateral lung	58	49	12	<5	<5	<5
Liver	<5	<5	<5	0	0	<5
Other	<5	<5	<5	<5	<5	0

* Redacted to adhere to statistical disclosure guidelines.

1.42 Feeding

Supplementary feeding, often using a nasogastric (via the nose) or gastrostomy (via the abdomen) tube directly to the stomach, is considered when a person with CF has poor weight gain, or progressive weight loss, despite efforts to increase oral intake.

Year		Overall	<16 years	≥16 years	<18 years	≥18 years
2013	**Total; n**	**9052**	**3839**	**5213**	**4354**	**4698**
2013	Any supplemental feeding; n(%)	2826 (31.2)	1020 (26.6)	1806 (34.6)	1182 (27.1)	1644 (35.0)
2013	Oral; n(%)	2329 (25.7)	843 (22.0)	1486 (28.5)	966 (22.2)	1363 (29.0)
2013	Nasogastric tube; n(%)	110 (1.2)	13 (0.3)	97 (1.9)	20 (0.5)	90 (1.9)
2013	Gastrostomy tube/Button; n(%)	548 (6.1)	204 (5.3)	344 (6.6)	243 (5.6)	305 (6.5)
2013	Jejunal; n(%)	-*	<5	7 (0.1)	<5	7 (0.1)
2013	Total Parenteral Nutrition (TPN); n(%)	-*	<5	5 (0.1)	6 (0.1)	<5
2018	**Total; n**	**9847**	**3895**	**5952**	**4314**	**5533**
2018	Any supplemental feeding; n(%)	3504 (35.6)	1225 (31.5)	2279 (38.3)	1394 (32.3)	2110 (38.1)
2018	Oral; n(%)	2749 (27.9)	951 (24.4)	1798 (30.2)	1087 (25.2)	1662 (30.0)
2018	Nasogastric tube; n(%)	105 (1.1)	17 (0.4)	88 (1.5)	21 (0.5)	84 (1.5)
2018	Gastrostomy tube/Button; n(%)	552 (5.6)	211 (5.4)	341 (5.7)	242 (5.6)	310 (5.6)
2018	Jejunal; n(%)	-*	<5	5 (0.1)	<5	<5
2018	Total Parenteral Nutrition (TPN); n(%)	6 (0.1)	<5	<5	<5	<5
2023	**Total; n**	**10344**	**3756**	**6588**	**4181**	**6163**
2023	Any supplemental feeding; n(%)	2996 (29.0)	940 (25.0)	2056 (31.2)	1048 (25.1)	1948 (31.6)
2023	Oral; n(%)	1871 (18.1)	580 (15.4)	1291 (19.6)	638 (15.3)	1233 (20.0)
2023	Nasogastric tube; n(%)	40 (0.4)	11 (0.3)	29 (0.4)	15 (0.4)	25 (0.4)
2023	Gastrostomy tube/Button; n(%)	353 (3.4)	129 (3.4)	224 (3.4)	148 (3.5)	205 (3.3)
2023	Jejunal; n(%)	-*	<5	9 (0.1)	<5	7 (0.1)
2023	Total Parenteral Nutrition (TPN); n(%)	<5	<5	0 (0.0)	<5	0 (0.0)

1.43 Pancreatic enzyme supplementation

Year		Overall	<16 years	≥16 years	<18 years	≥18 years
2013	**Total; n**	**9052**	**3839**	**5213**	**4354**	**4698**
2013	Pancreatic enzyme supplements; n(%)	7768 (85.8)	3326 (86.6)	4442 (85.2)	3793 (87.1)	3975 (84.6)
2018	**Total; n**	**9847**	**3895**	**5952**	**4314**	**5533**
2018	Pancreatic enzyme supplements; n(%)	8140 (82.7)	3212 (82.5)	4928 (82.8)	3578 (82.9)	4562 (82.5)
2023	**Total; n**	**10344**	**3756**	**6588**	**4181**	**6163**
2023	Pancreatic enzyme supplements; n(%)	8297 (80.2)	2984 (79.4)	5313 (80.6)	3342 (79.9)	4955 (80.4)

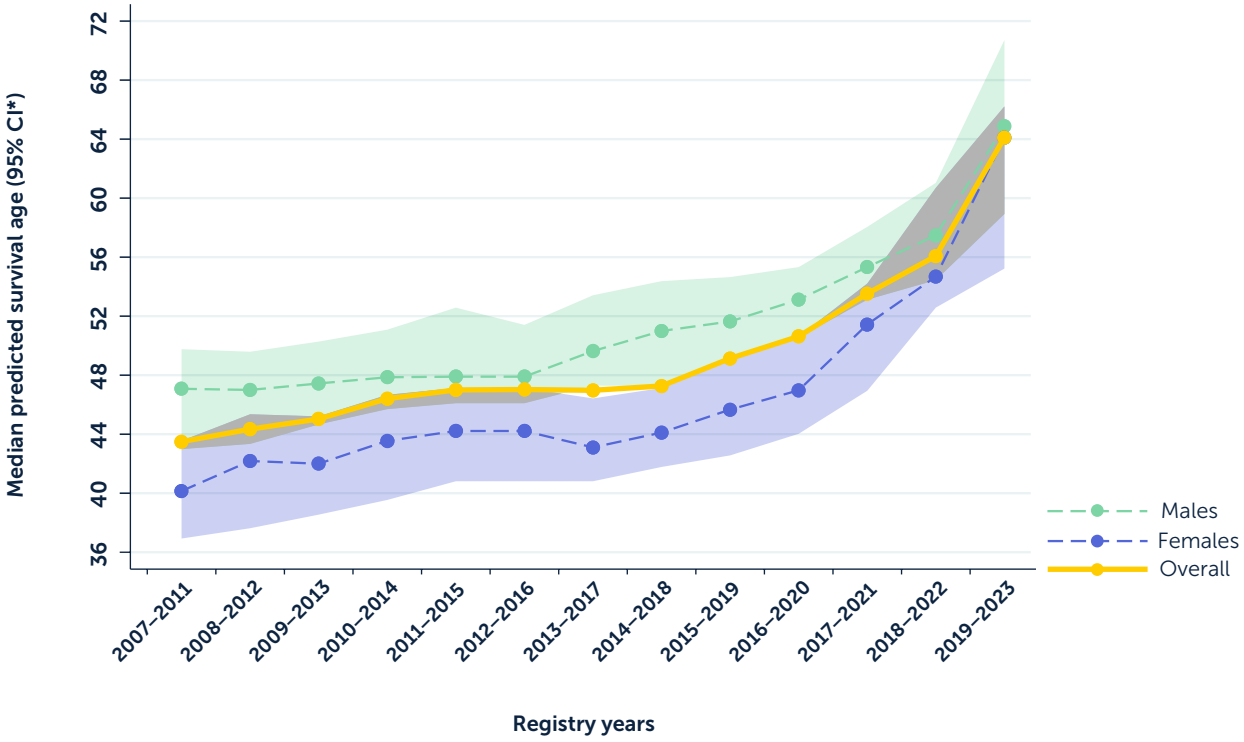
* Redacted to adhere to statistical disclosure guidelines.

Survival

1.44 Median predicted survival age

The calculation of median predicted survival age is based on people with CF who are recorded in the Registry as alive in the given year. A mathematical formula[1] predicts how long we expect half of people with CF born today will live. Half of people born today are predicted to live to at least **64.1** years. Half are therefore predicted to die before they reach that age.

Grouping together several years of data gives a better estimate of predicted survival. One-year data can show big variations in median predicted survival age from year to year, which may be due to chance alone and does not necessarily reflect a change in real-world outcomes. A rolling five-year predicted survival is therefore shown to try to smooth out these fluctuations.



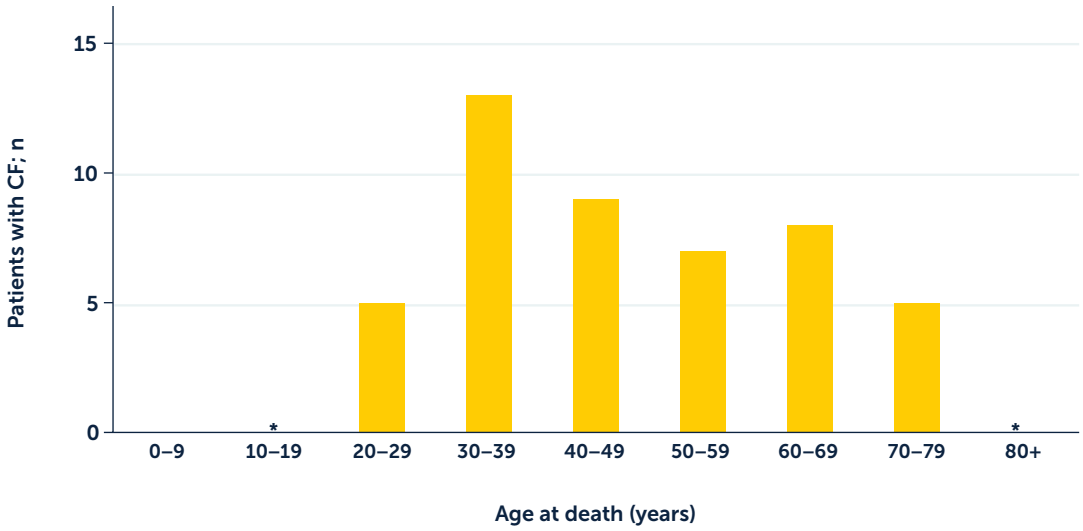
Median predicted survival age; years (95% CI*)				
Years	Overall	Female	Male	p-value (males vs females)
2007-2011	43.5(41.9-45.9)	40.1(36.9-43.6)	47.1(43.0-49.8)	<0.001
2008-2012	44.3(42.4-46.5)	42.2(37.6-45.3)	47.0(43.3-49.6)	<0.001
2009-2013	45.0(42.8-47.0)	42.0(38.5-45.2)	47.4(44.7-50.3)	<0.001
2010-2014	46.4(43.7-47.9)	43.6(39.5-46.7)	47.9(45.7-51.1)	<0.001
2011-2015	47.0(44.3-48.2)	44.2(40.8-47.1)	47.9(46.1-52.6)	0.004
2012-2016	47.0(44.7-48.2)	44.2(40.8-47.1)	47.9(46.1-51.4)	0.003
2013-2017	47.0(44.8-48.2)	43.1(40.8-46.4)	49.6(47.3-53.4)	<0.001
2014-2018	47.3(45.7-49.6)	44.1(41.8-47.1)	51.0(47.3-54.4)	<0.001
2015-2019	49.1(47.0-51.4)	45.7(42.6-49.2)	51.6(49.0-54.6)	<0.001
2016-2020	50.6(48.2-53.1)	47.0(44.0-50.6)	53.1(50.6-55.3)	0.004
2017-2021	53.5(51.5-55.2)	51.4(46.9-54.2)	55.3(53.1-58.1)	0.002
2018-2022	56.1(54.4-59.0)	54.7(52.6-60.7)	57.5(54.4-61.0)	0.057
2019-2023	64.1(58.9-67.0)	64.1(55.2-66.2)	64.9(58.9-70.7)	0.068

[1] Sykes, Jenna et al. J Clin Epidemiol. 2016;70:206-213.

* Confidence interval.

1.45 Age distribution of deaths 2021–2023

The table below shows the ages of the 49 people with CF who died in 2023.
In 2023 the median age of the 49 people who died was 46.



Age at death	2021 (n)	2022 (n)	2023 (n)
0-9	<5	0	0
10-19	<5	<5	<5
20-29	13	19	5
30-39	24	24	13
40-49	22	12	9
50-59	<5	12	7
60-69	<5	<5	8
70-79	<5	<5	5
80+	**0**	**0**	**<5**
Total	**69**	**72**	**49**

1.46 Causes of death

This table shows all the recorded causes of death between 2021–2023.

Cause of death	2021 n(%)	2022 n(%)	2023 n(%)
Respiratory/cardiorespiratory	33 (47.8)	29 (40.3)	27 (55.1)
Other	6 (8.7)	15 (20.8)	6 (12.2)
Cancer	5 (7.2)	6 (8.3)	5 (10.2)
Transplant-related	9 (13.0)	9 (12.5)	<5
Liver disease/liver failure	<5	<5	<5
Trauma or suicide	<5	0 (0.0)	<5
Not known	7 (10.1)	9 (12.5)	<5
COVID-19	7 (10.1)	<5	0 (0.0)
Total	69	72	49

* Redacted to adhere to statistical disclosure guidelines.

Genotypes*

Genotypes are part of the genetic makeup of an individual that usually control a particular characteristic, known as a phenotype. For people with CF, their genotype reveals which variants of the CF gene cause their cystic fibrosis. Everyone living with CF has two variants of the gene for CFTR; one on each allele. One is inherited from their mother, and one from their father. If both variants (or genotypes) are the same, the person is said to be homozygous. Someone who has two different variants is heterozygous.

Data completeness	n(%)
Patients genotyped with at least one CFTR variant recorded	11240 (99.3)
Patients genotyped with both CFTR variants recorded	10987 (97.1)
F508del variants	
Homozygous F508del	5365 (47.4)
Heterozygous F508del	4708 (41.6)

1.47 CFTR variant combinations in the UK population

This tabulation shows the proportion(%) of patients with the most common CFTR variant combinations in their genotype. For example, 4.0% of the UK population have one copy of F508del and one copy of G551D.

CFTR variant	F508del	R117H	G551D	G542X	621+1G->T	Other	Unknown	Total
F508del	47.5							47.5
R117H	5.1	0.1						5.2
G551D	4.0	0.2	0.2					4.4
G542X	2.5	0.1	0.1	0.1				2.8
621+1G->T	1.7	0.1	0.1	0.1	0.1			2.0
Other	26.8	0.6	1.0	0.8	0.5	5.5		35.2
Unknown	1.5	0.1	0.1	0.1	0.0	0.5	0.7	2.9
Total	**89.1**	**1.2**	**1.4**	**1.0**	**0.6**	**6.0**	**0.7**	**100.0**

* In this section, we include everyone who is registered (see table 1.1) and where CFTR variants are available.

1.48 CFTR variants in the UK population

The table below shows the number of people with CF who carry at least one of each CFTR variant. The groups are not mutually exclusive because people with heterozygous variants appear twice in the table.

These are the 20 most common CFTR variants in the UK population. The full list of recorded variants can be found in Appendix 3.

Nucleotide	Protein	Legacy name	n	%
c.1521_1523delCTT	p.Phe508del	F508del	10073	89.0
c.350G->A	p.Arg117His	R117H	714	6.3
c.1652G->A	p.Gly551Asp	G551D	641	5.7
c.1624G->T	p.Gly542X	G542X	418	3.7
c.489+1G->T		621+1G->T	292	2.6
c.3909C->G	p.Asn1303Lys	N1303K	182	1.6
c.1585-1G->A		1717-1G->A	181	1.6
c.3454G->C	p.Asp1152His	D1152H	163	1.4
c.1766+1G->A		1898+1G->A	160	1.4
c.200C->T	p.Pro67Leu	P67L	155	1.4
c.3140-26A->G		3272-26A->G	129	1.1
c.3528delC	p.Lys1177SerfsX15	3659delC	127	1.1
c.1679G->C	p.Arg560Thr	R560T	106	0.9
c.1477C->T	p.Gln493X	Q493X	95	0.8
c.1519_1521delATC	p.Ile507del	I507del	94	0.8
c.1657C->T	p.Arg553X	R553X	88	0.8
c.254G->A	p.Gly85Glu	G85E	87	0.8
c.2657+5G->A		2789+5G->A	87	0.8
c.3717+12191C->T		3849+10kbC->T	86	0.8
c.178G->T	p.Glu60X	E60X	80	0.7

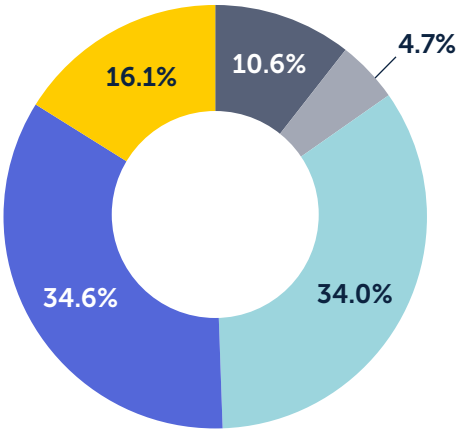
1.49 CFTR variant prevalence by devolved nation

This table shows the distribution of individual CFTR variants across the devolved nations. The number of patients for each devolved nation is based on the location of the CF centre at which the patient receives care and does not account for patients who travel between devolved nations for care. The groups are not mutually exclusive because people with heterozygous variants appear twice in the table.

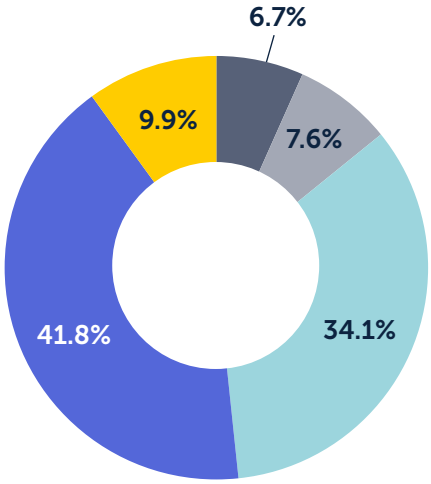
Legacy name	England n=9364		Scotland n=972		Wales n=473		Northern Ireland n=509	
	n	%	n	%	n	%	n	%
F508del	8347	89.1%	876	90.1%	424	89.6%	426	83.7%
R117H	545	5.8%	79	8.1%	18	3.8%	72	14.1%
G551D	481	5.1%	95	9.8%	18	3.8%	47	9.2%
G542X	301	3.2%	62	6.4%	24	5.1%	31	6.1%
621+1G->T	211	2.3%	10	1.0%	53	11.2%	18	3.5%
N1303K	153	1.6%	12	1.2%	7	1.5%	10	2.0%
1717-1G->A	158	1.7%	18	1.9%	<5	-	<5	-
D1152H	126	1.3%	23	2.4%	<5	-	11	2.2%
1898+1G->A	125	1.3%	5	0.5%	30	6.3%	0	0.0%
P67L	78	0.8%	54	5.6%	<5	-	21	4.1%

1.50 Genotype prevalence by devolved nation

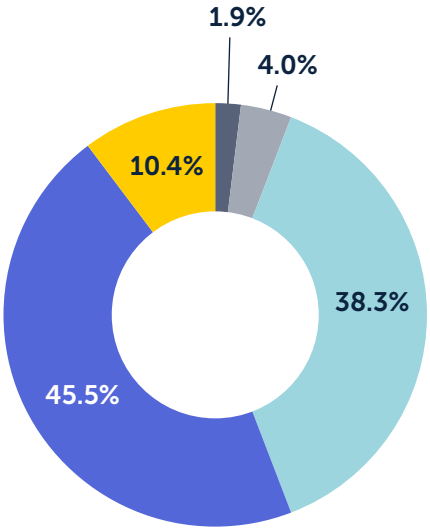
These charts show the distribution of CFTR variant combinations across the devolved nations. The number of patients for each devolved nation is based on the location of the CF centre at which the patient receives care and does not account for patients who travel between devolved nations for care.



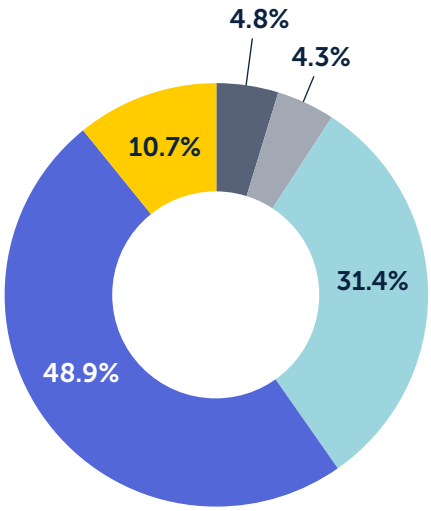
Northern Ireland
N=509



Scotland
N=972



Wales
N=473



England
N=9364

F508del + R117H

F508del + gating (incl. G551D)

Heterozygous F508del

Homozygous F508del

Other

Section 2 and 3: Centre-level analysis

Cystic fibrosis care in the UK is led by 56 regional centres, 4 standalone clinics, and 75 networked clinics. The breakdown between centres and clinics delivering paediatric and adult care is shown below:

	Paediatric	Adult	Total
Centres	30	26	56
Standalone clinics	2	2	4
Networked clinics	68	7	75

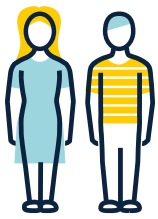
Section 2 shows analysis of data for individual CF centres. This allows people with CF, their families, and healthcare providers, to review a centre's use of some medications and outcome data alongside national averages. This transparency is intended to help improve standards of care overall.

Lots of different factors can affect the outcomes of people with CF in centres, not all of which are within a centre's control. This might include the economic profile of the area, the age at which the person with CF was diagnosed and referred to the centre, certain patient characteristics such as their gender, as well as facilities, care pathways, and the medical team providing care.

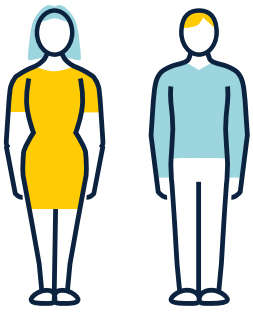
If a person with CF or a member of their family has questions about the results for their CF centre or clinic, they should discuss this with their CF team

Full tables of the data are shown in appendix 2 on page 74.

Key



Paediatric centre

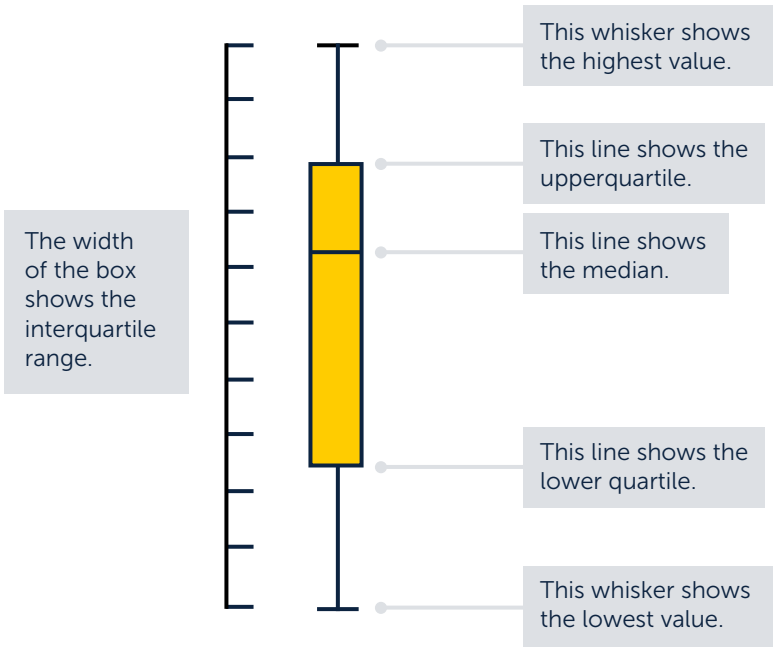


Adult centre

A guide to the charts

Some of the data in this section are shown as 'box plots'. We also show the data in 'funnel plots'.

Box plots



- The 'box' shows the middle half of the data for that centre, going from the first quartile to the third quartile. The longer the box, the more varied the data for that centre.
- The horizontal line within the box shows the median result for that centre.
- The 'whiskers' above and below the box show the highest and lowest values for that centre, excluding any outliers.
- The position of the box between the whiskers shows any skew in the data. If a box is towards the top of the whisker, more of the people for this centre were recorded at the high end of the scale.

Funnel plots

The more people with CF at a care site, the closer to the national average you would expect the results to be. This is because high numbers in one centre affect the overall average across the country, 'pulling' the average towards them. When a small number people with CF are treated at a site, even a single outcome that is unusual affects the overall result for that site much more.

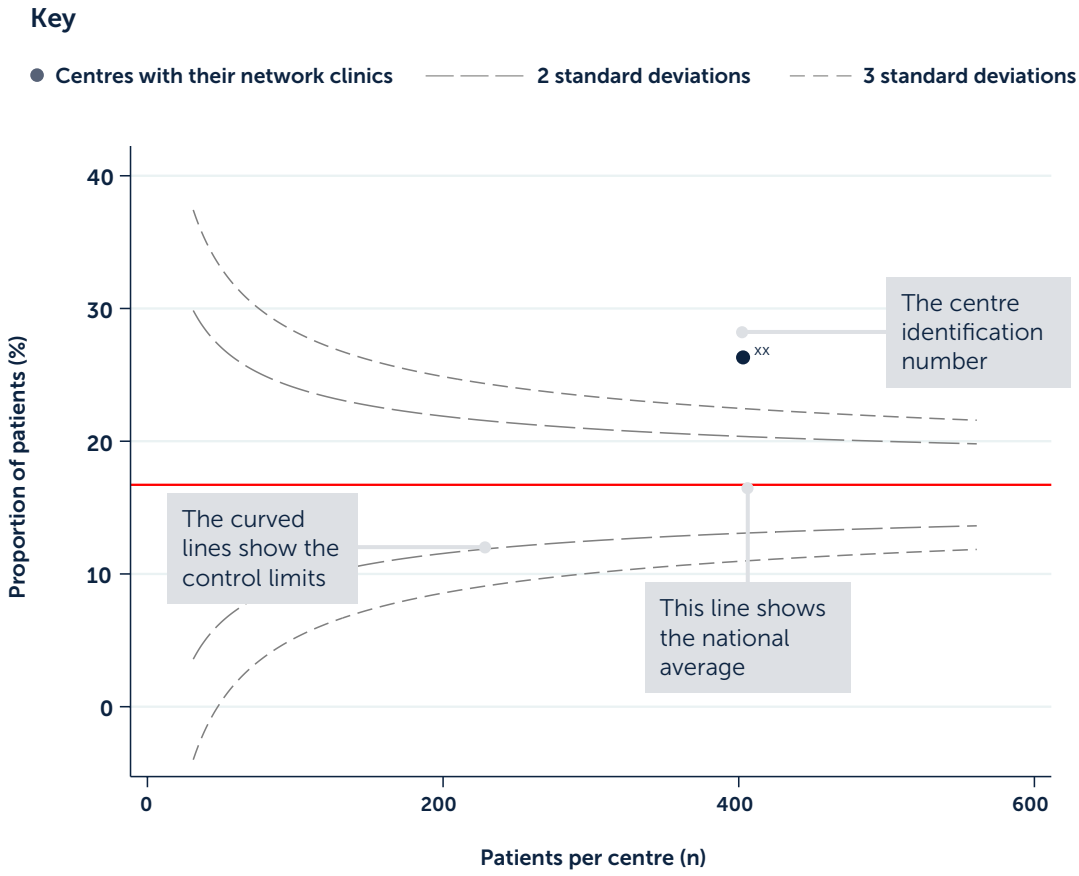
There will always be some natural variation between centres because of differences between the populations receiving care. Using only the national average as a standard can make it difficult to tell whether a survival rate that sits above the national average is higher than we would expect it to be, or not.

For this reason, the funnel plots also show 'control limits'; the curved lines on the charts that give them the 'funnel' shape. The horizontal line in the middle of the funnel shows the national average. Control limits show the rate we would expect, based on the number of people with CF at that site.

If the result for a CF centre is between the two 'control limits', it is 'as expected' and any variation above or below the national average may be due to chance alone. If a result is below the bottom control it is lower than expected, if it is above the upper control, it is higher than expected. Being outside the control limits can be a good thing, for example if a site's lung function results are exceptionally high.

A centre's data can sit outside of the control limits for a number of reasons, including patient characteristics (for example, an adult centre with younger patients might have a higher average lung function than one with older patients), problems with data submitted to the Registry, specialist practice, chance, or the care being delivered.

Where charts have been adjusted for age, this means that the data have been fine-tuned to take account of the different spread of ages across centres and clinics. The adjusted values are intended to show what the average lung function or BMI percentile would be for that centre/clinic if the age spread is the same as the spread of age in the whole population. Because it is difficult for adjustment to fully account for all factors that might affect clinical outcomes, we should be very careful about drawing conclusions based on adjusted outcomes alone.

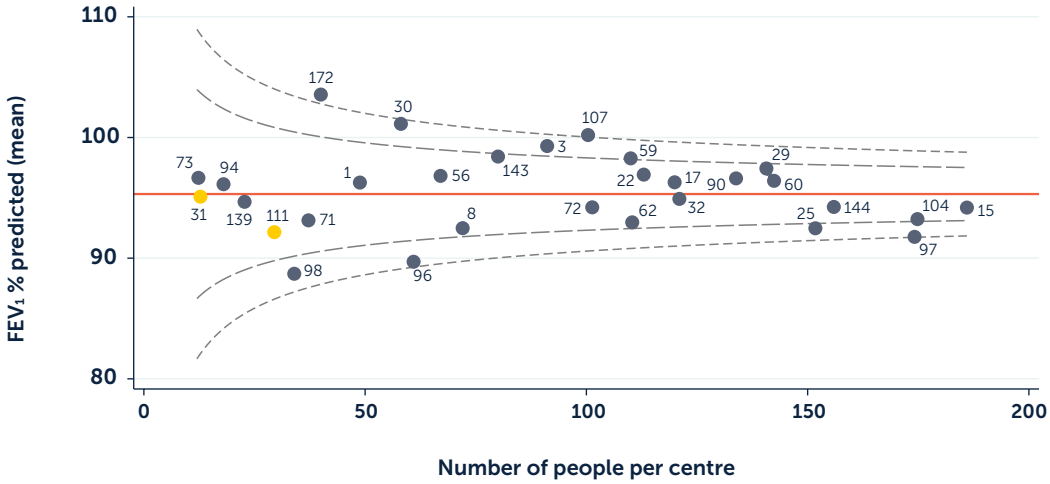


Section 2 Paediatric centre analysis

N=4082

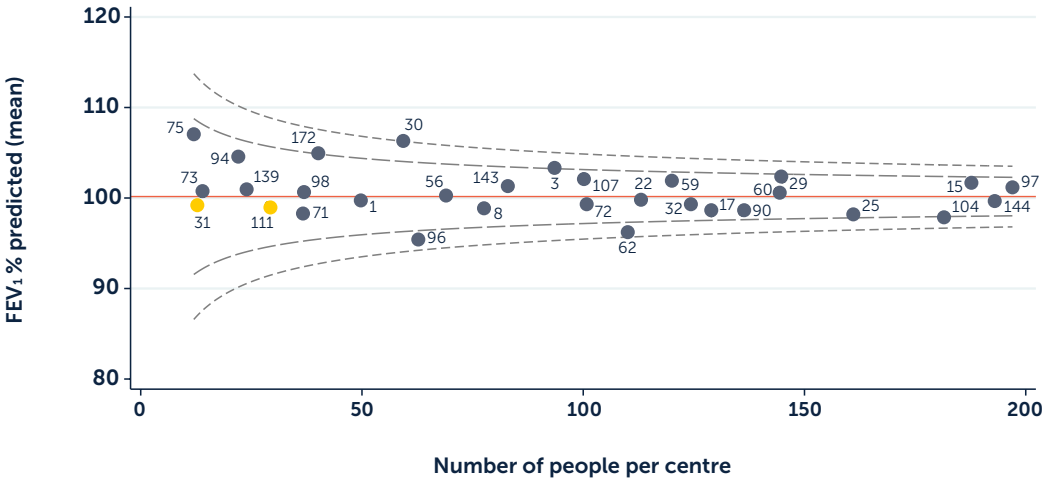
In the UK, paediatric CF care is led by 30 specialist CF centres and two standalone clinics (●). Some paediatric centres oversee care delivered by 68 smaller, networked clinics. Data from smaller networked clinics is included in the paediatric centre's data.

2.1 Age-adjusted FEV_1 % predicted at annual review, in patients aged six and over without a history of lung transplant, by paediatric centre/clinic



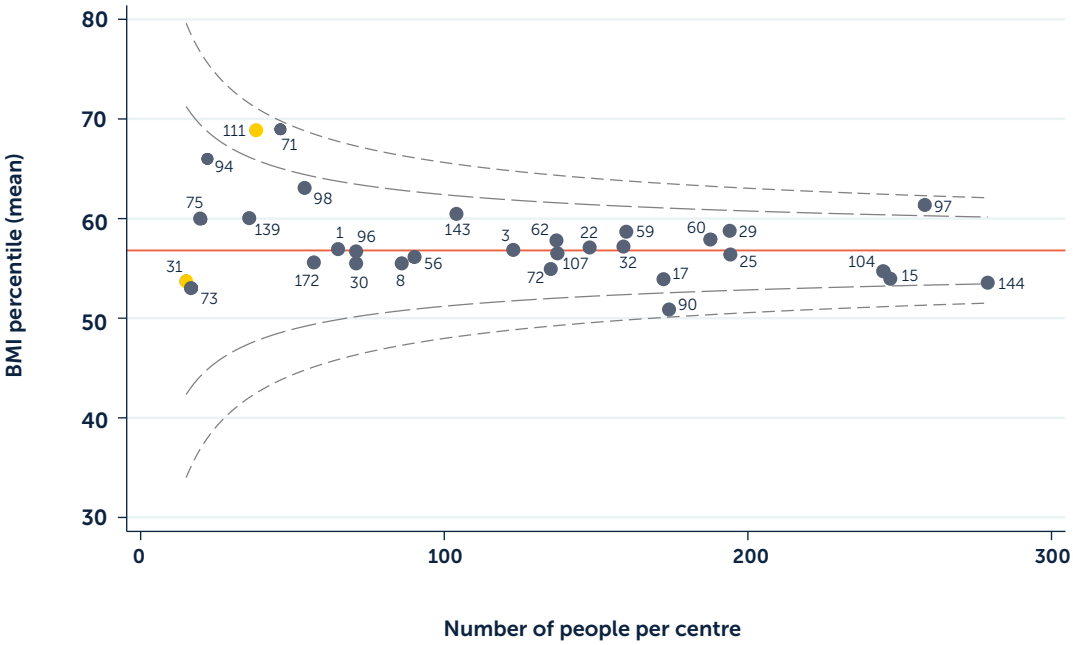
The mean FEV_1% predicted for patients attending paediatric centres/clinics is 95.3% predicted.

2.2 Age-adjusted Best FEV_1 % predicted at annual review, in patients aged six and over without a history of lung transplant, by paediatric centre/clinic



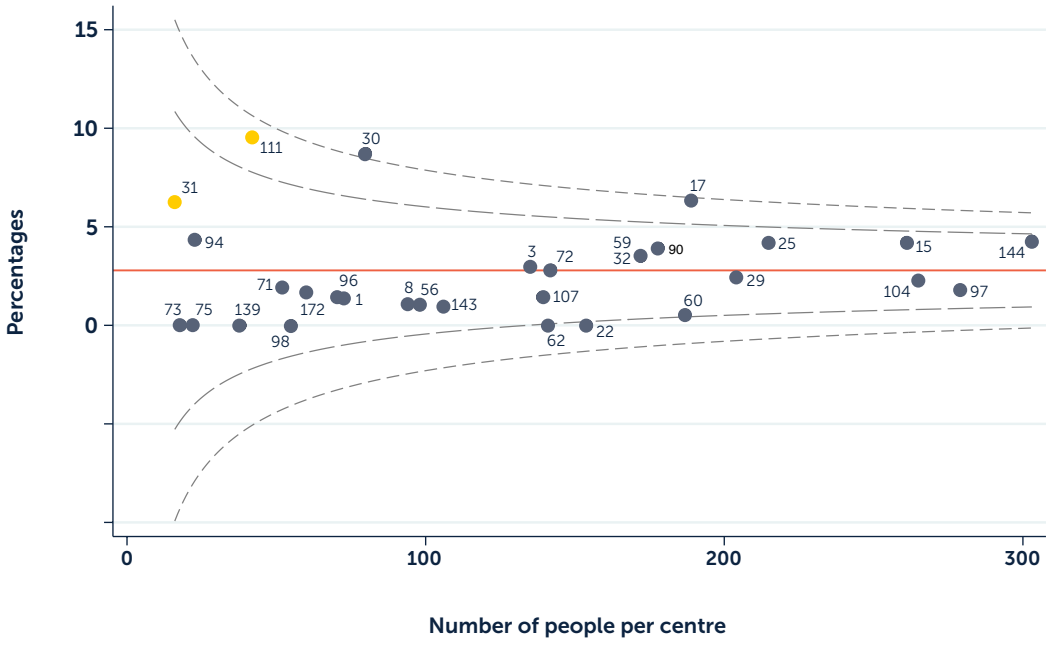
The mean Best FEV_1% predicted for patients attending paediatric centres/clinics is 100.2% predicted. Where Best FEV_1% predicted was missing, the FEV_1% predicted at annual review was used.

2.3 Age-adjusted Body Mass Index (BMI) percentile in patients aged 1-15 years by paediatric centre/clinic



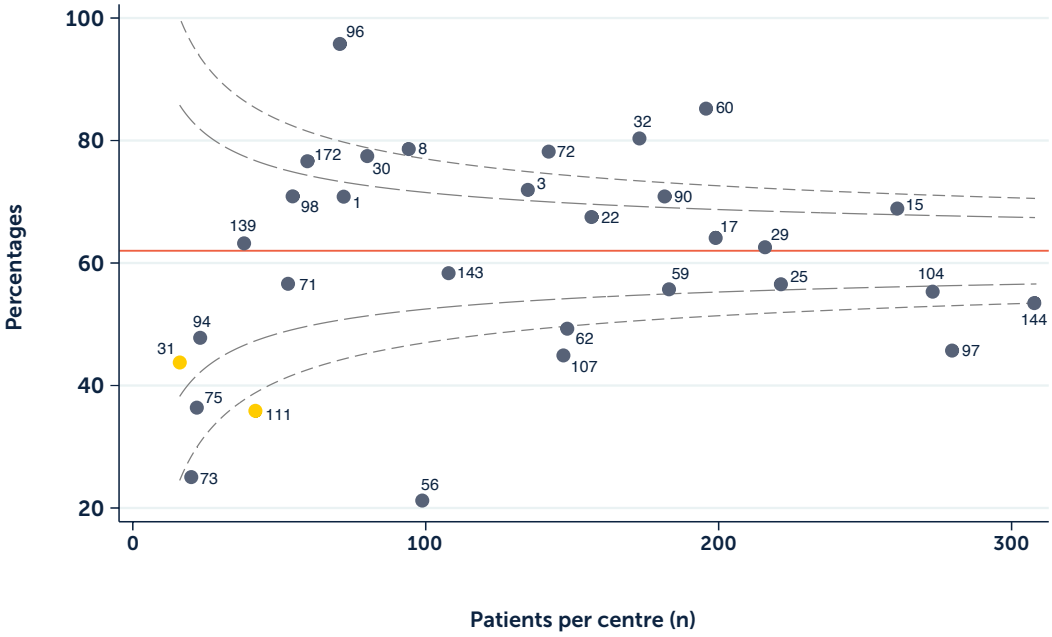
The mean BMI percentile for patients attending paediatric centres/clinics is 56.8%.

2.4 Proportion of patients with chronic *Pseudomonas aeruginosa* by paediatric centre/clinic



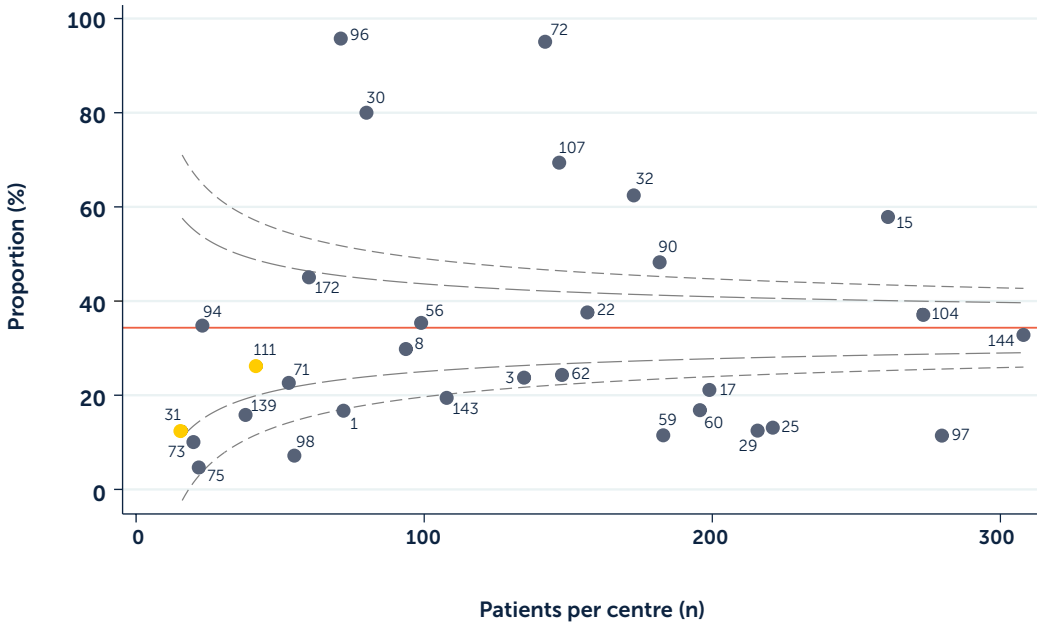
The proportion of patients with chronic *Pseudomonas aeruginosa* in paediatric centres/clinics is 2.8%.

2.5 Proportion of patients receiving DNase treatment by paediatric centre/clinic



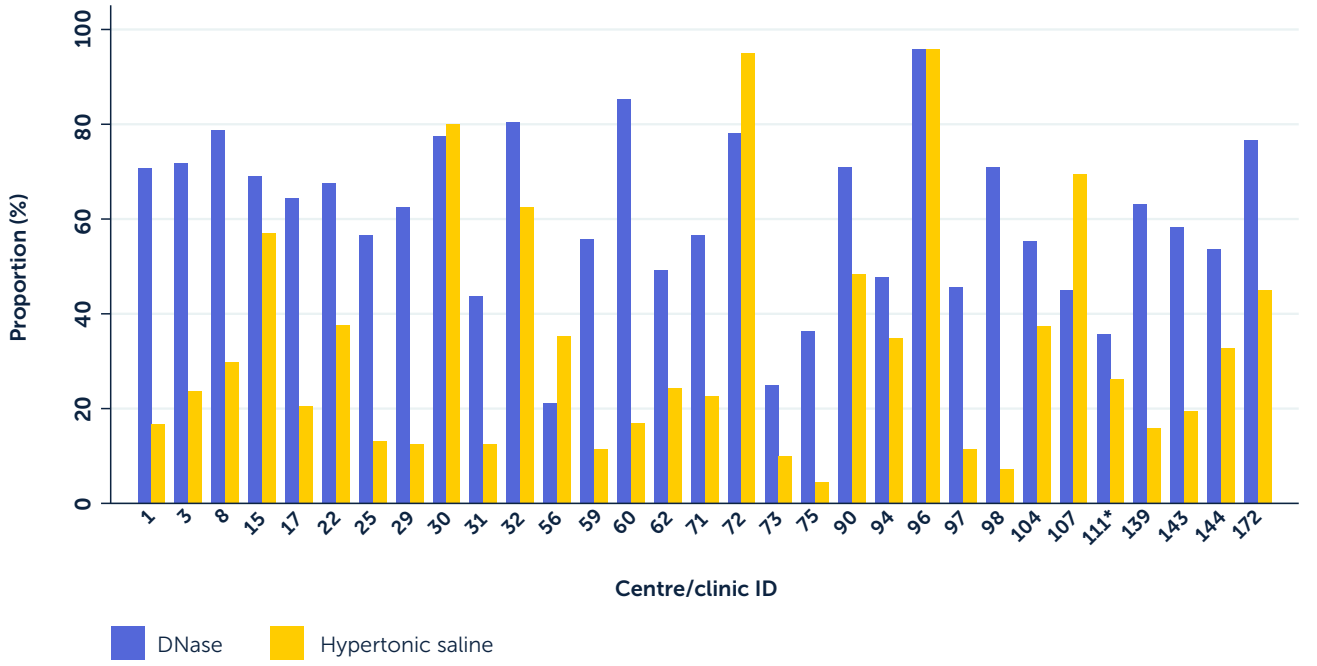
The proportion of patients receiving DNase treatment in paediatric centres/clinics is 62.0%.

2.6 Proportion of patients on hypertonic saline or mannitol treatment by paediatric centre/clinic



The proportion of patients receiving hypertonic saline or mannitol treatment in paediatric centres/clinics is 34.3%.

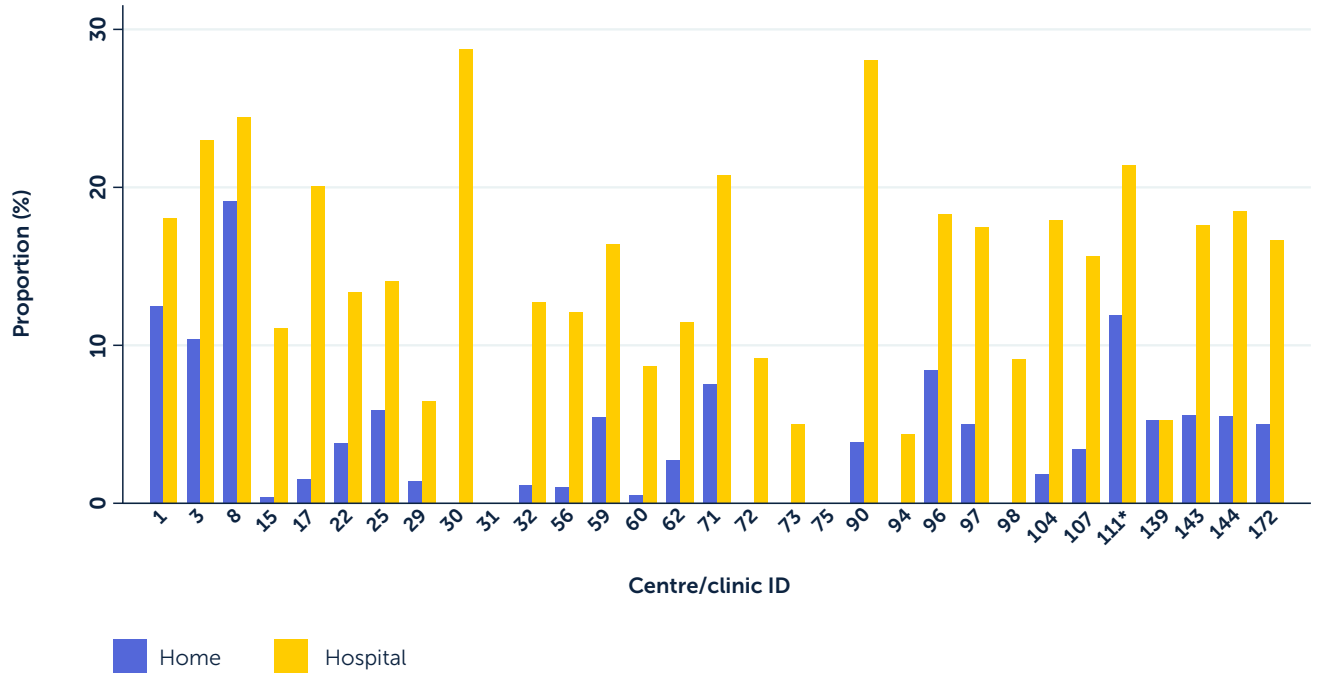
2.7 Proportion of patients receiving DNase/hypertonic saline/ mannitol treatment by paediatric centre/clinic



Due to the small number of paediatric patients that received mannitol (<5 across all clinics/centres), receipt of mannitol is omitted from the above graph.

2.8 IV use by paediatric centre/clinic

The chart below shows the proportion of patients with at least one IV day at home and/or in hospital. Patients may have a combination of home and hospital IV days.



The proportion of patients receiving IVs at home was 3.9% and in hospital was 15.6%. The proportion receiving any IVs was 16.5%.

* Standalone clinics.

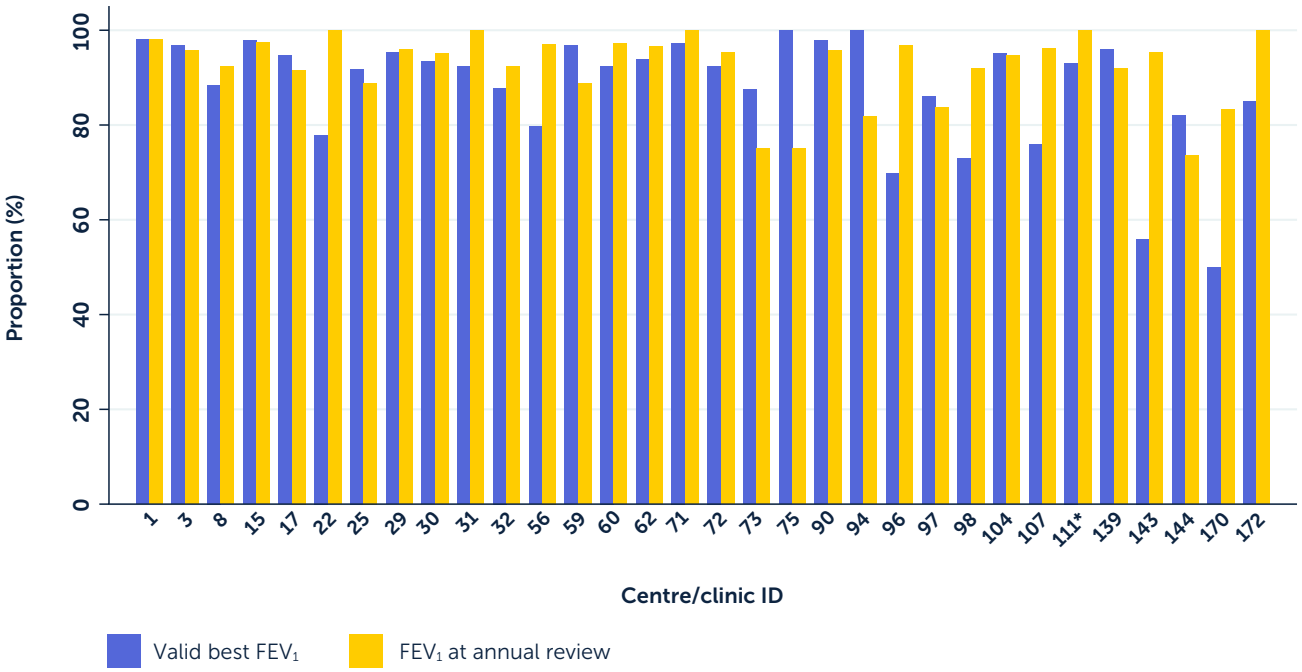
2.9 Inhaled antibiotic use for patients with chronic *Pseudomonas aeruginosa*, by paediatric centre/clinic

This table excludes centres where fewer than 10 patients had chronic *P. aeruginosa*.

Centre/clinic ID	Proportion(%)
15	90.9
17	100.0
144	84.6

84.1% of patients with chronic *P. aeruginosa* received inhaled antibiotics.

2.10 Data completeness by paediatric centre/clinic**



* Standalone clinics.

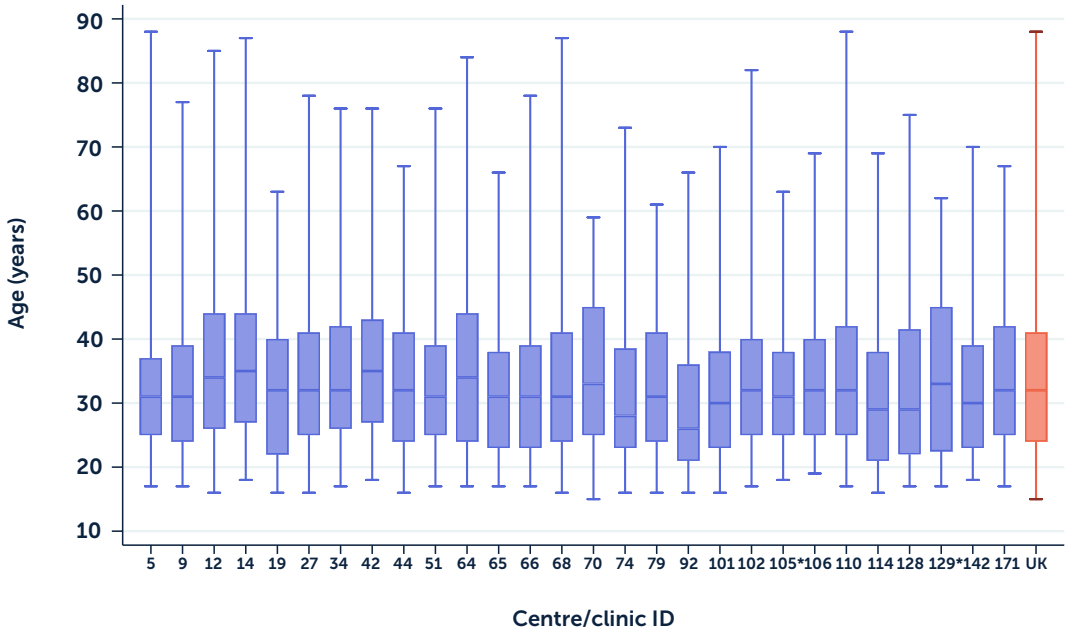
** The chart above shows the proportion of patients who had a valid best FEV_1% and an FEV_1% at annual review, excluding patients under six years of age. Best FEV_1% was considered valid if it was not missing, and the per cent predicted was not more than 0.5% lower than the annual review value. For some patients there may be medical reasons why FEV_1 could not be taken, so centres may not be able to get 100% completeness.

Section 3: Adult centre analysis

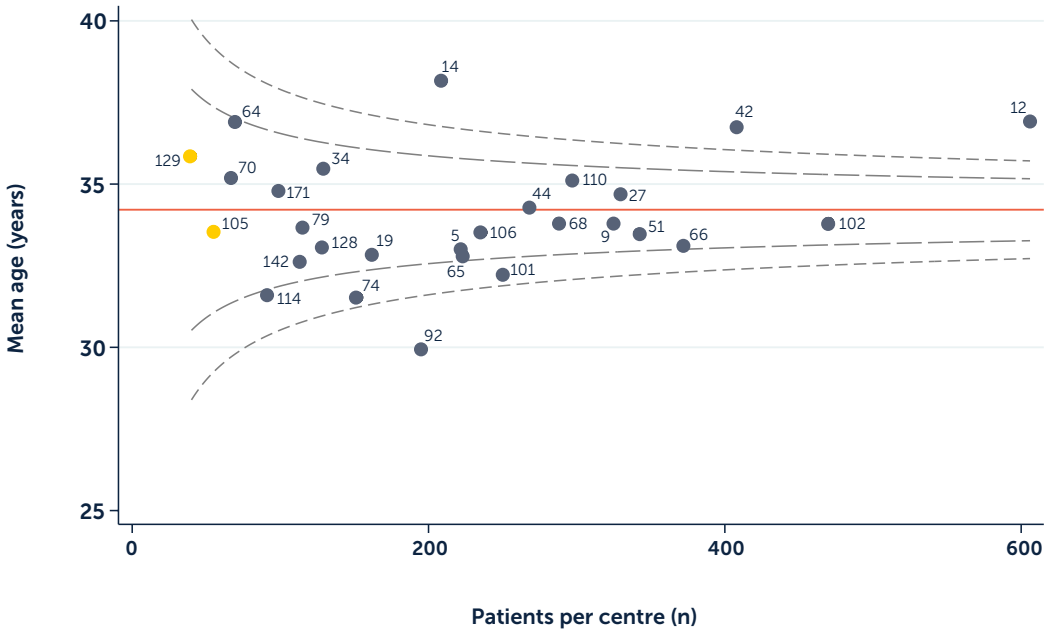
N=6262

In the UK, CF care is led by 26 adult specialist CF centres and two standalone clinics (🟡). People with CF transfer to adult care centres between the ages of 16 and 18 years.

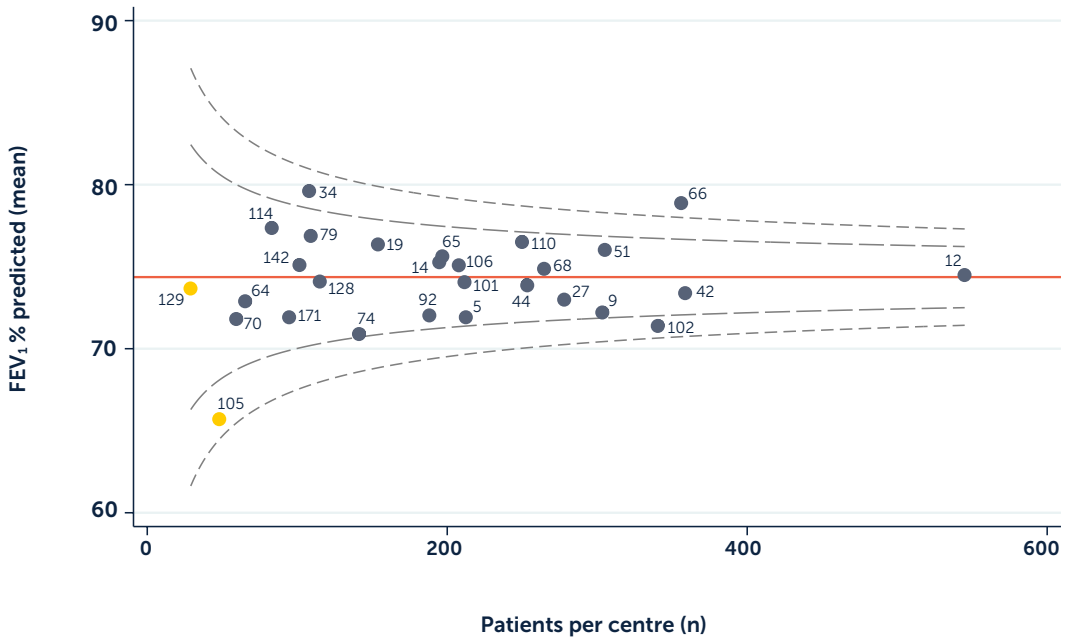
3.1 Age distribution by adults centre/clinic



The funnel plot below shows how the mean age in adult centres compares to the national mean. In 2023 the national mean age of patients at CF centres was 34.2 years.

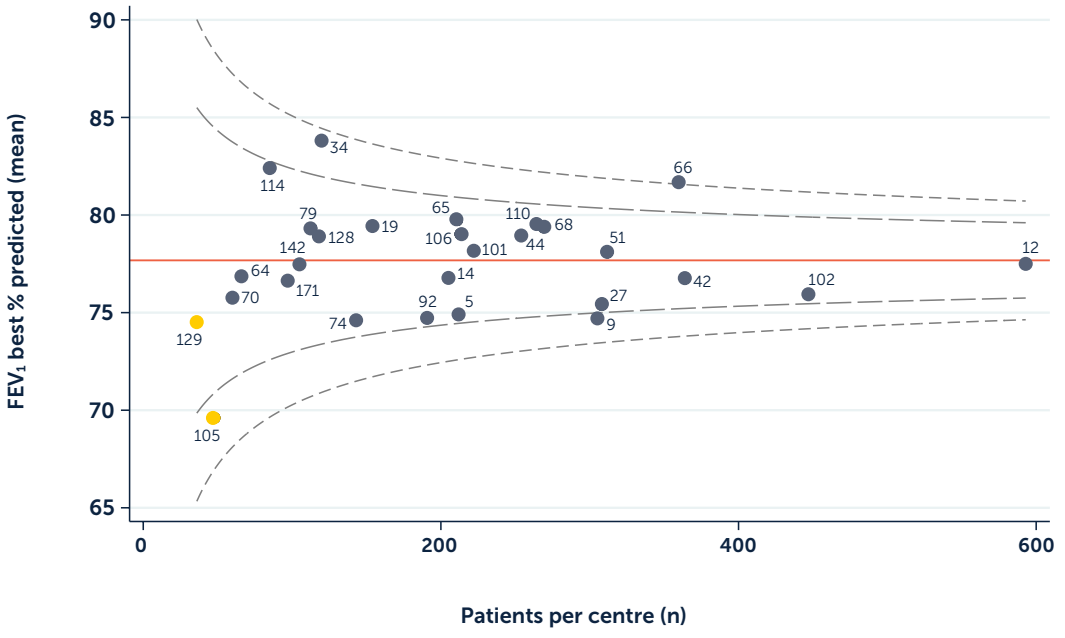


3.2 Age adjusted FEV_1 % predicted at annual review in patients without a history of lung transplant, by adult centre/clinic



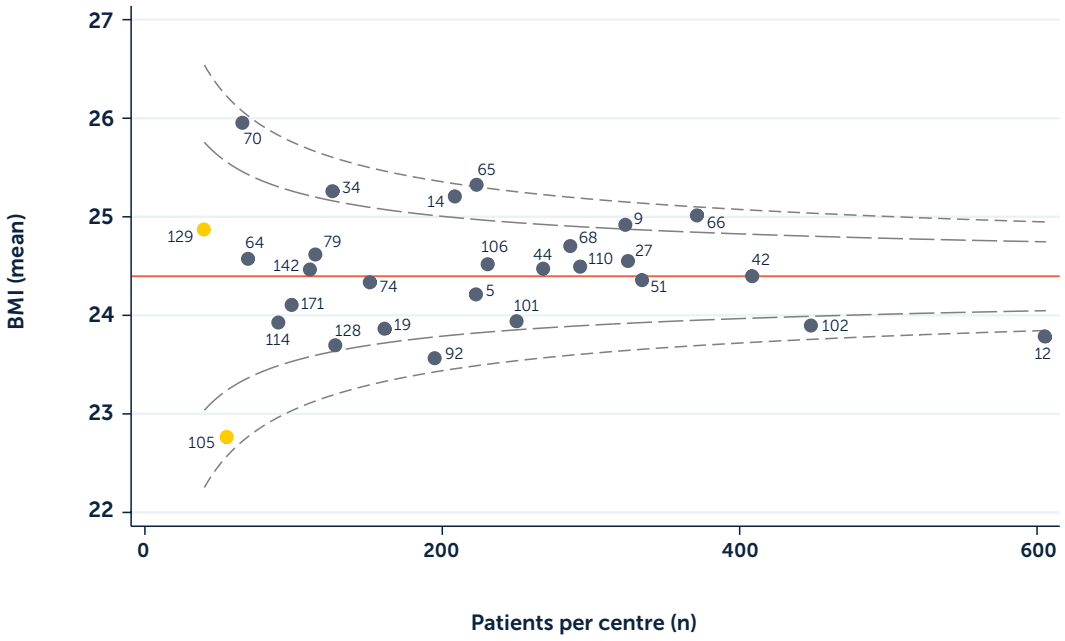
The mean FEV_1% predicted in adult centres/clinics is 74.4%.

3.3 Age adjusted Best FEV_1 % predicted at annual review in patients without a history of lung transplant, by adult centre/clinic



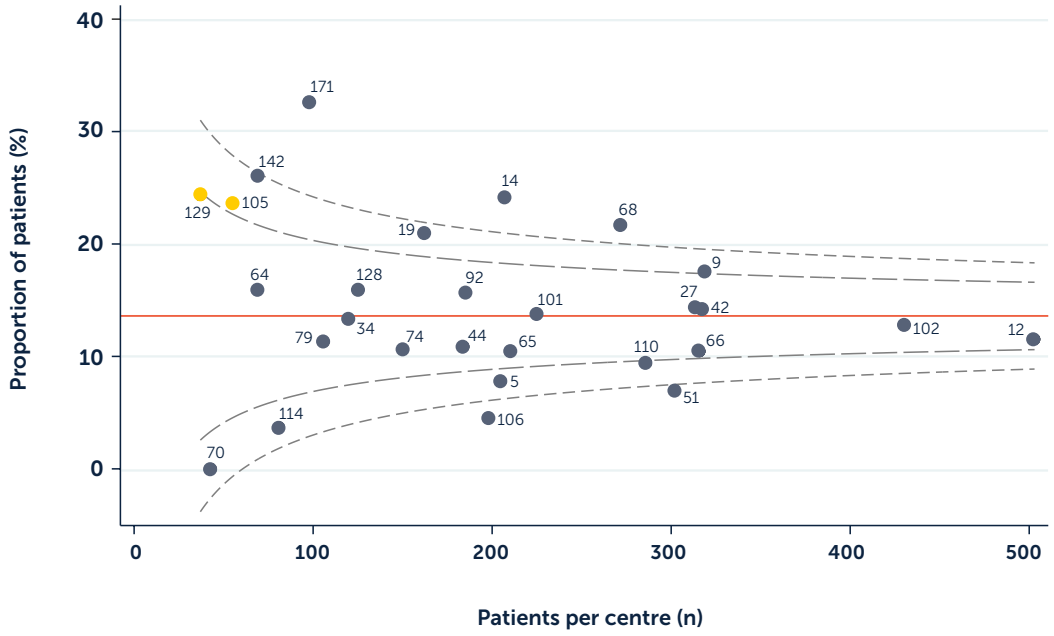
In 2023 the national mean was 77.7%. Where Best FEV_1% predicted was missing, or lower than the FEV_1 at annual review, the FEV_1% value at annual review was used.

3.4 Age-adjusted Body Mass Index (BMI) among patients aged 16 years and older by adult centre/clinic



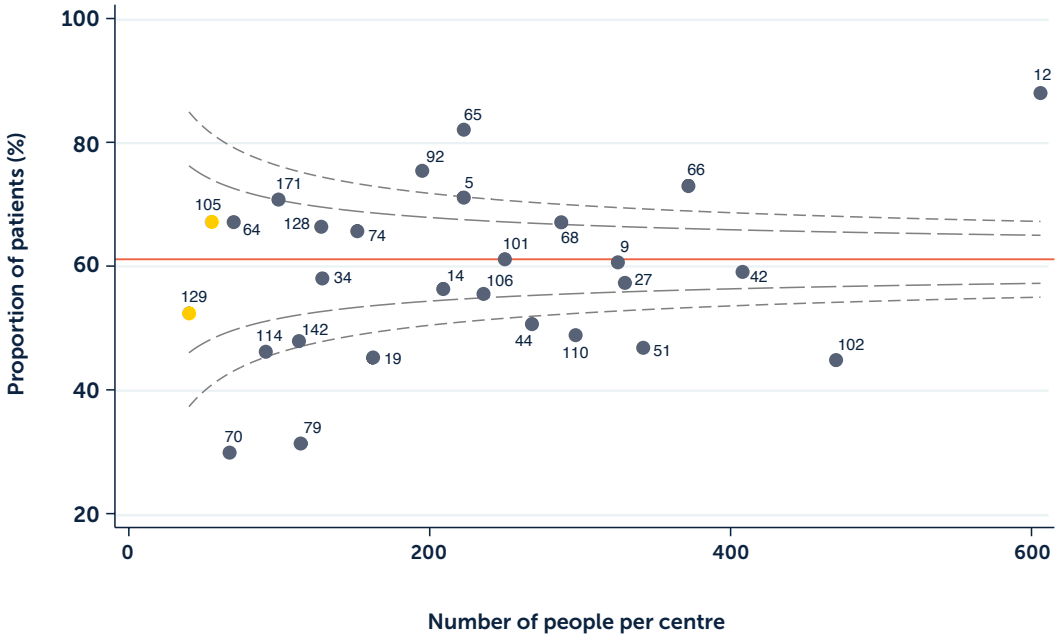
The mean BMI in adult centres/clinics is 24.4.

3.5 Proportion of patients with chronic *Pseudomonas aeruginosa* by adult centre/clinic



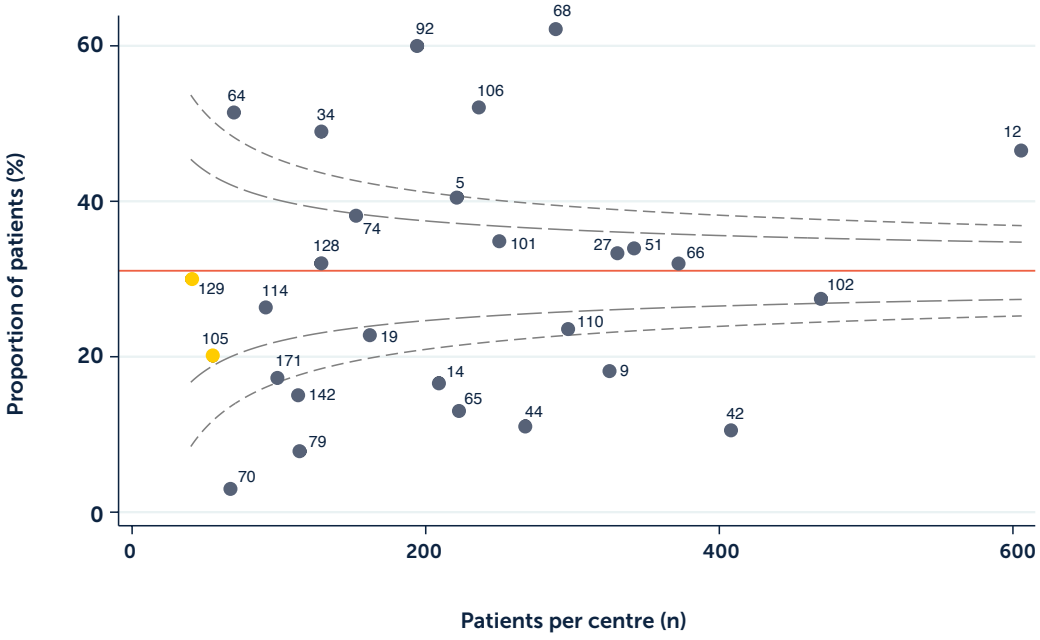
The proportion of patients with chronic *P. aeruginosa* in adult centres/clinics is 13.6%.

3.6 Proportion of patients receiving DNase treatment by adult centre/clinic



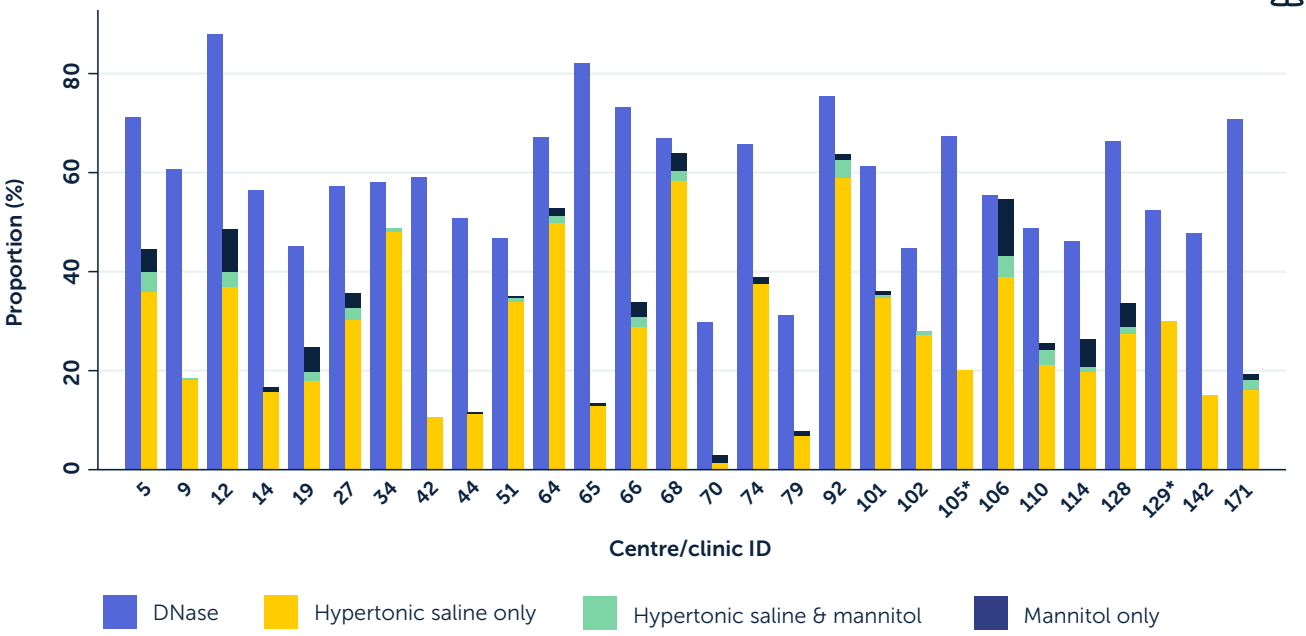
The proportion of patients receiving DNase treatment in adult centres/clinics is 61.1%.

3.7 Proportion of patients receiving hypertonic saline or mannitol by adult centre/clinic



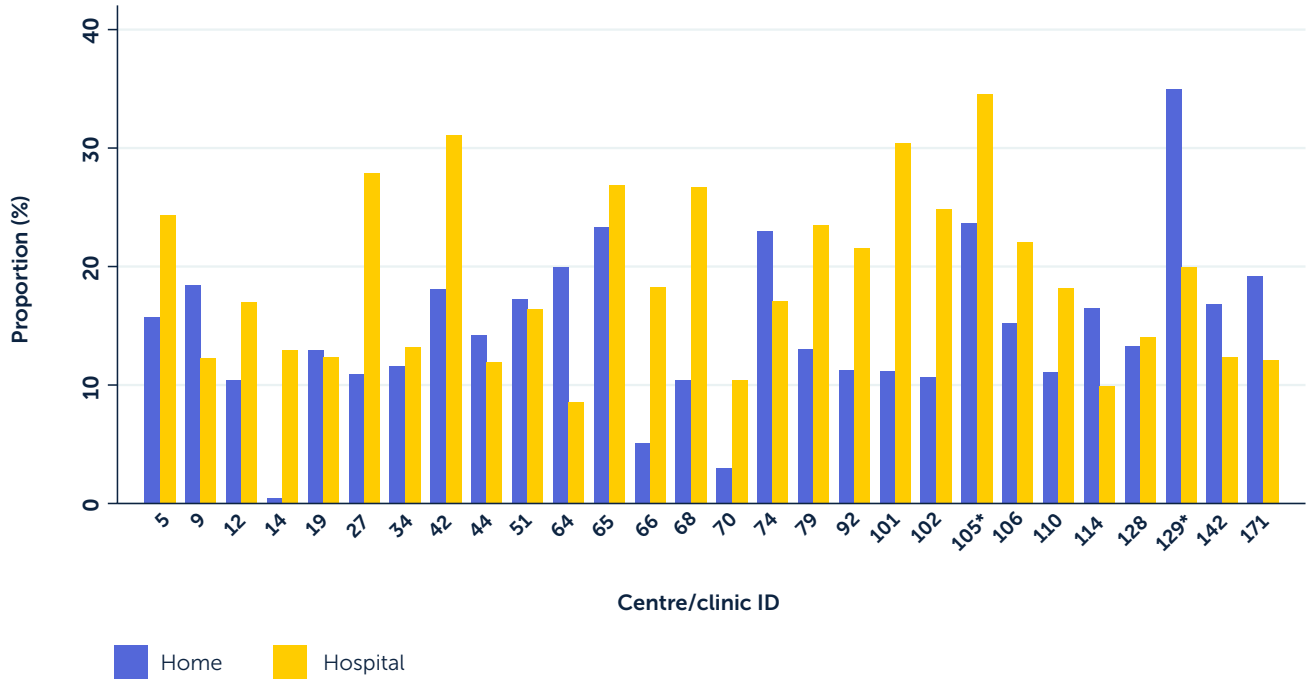
The proportion of patients receiving hypertonic saline or mannitol treatment in adult centres/clinics is 31.1%.

3.8 Proportion of patients receiving DNase/hypertonic saline/ mannitol treatment by adult centre/clinic



3.9 Intravenous (IV) antibiotic use by adult centre/clinic

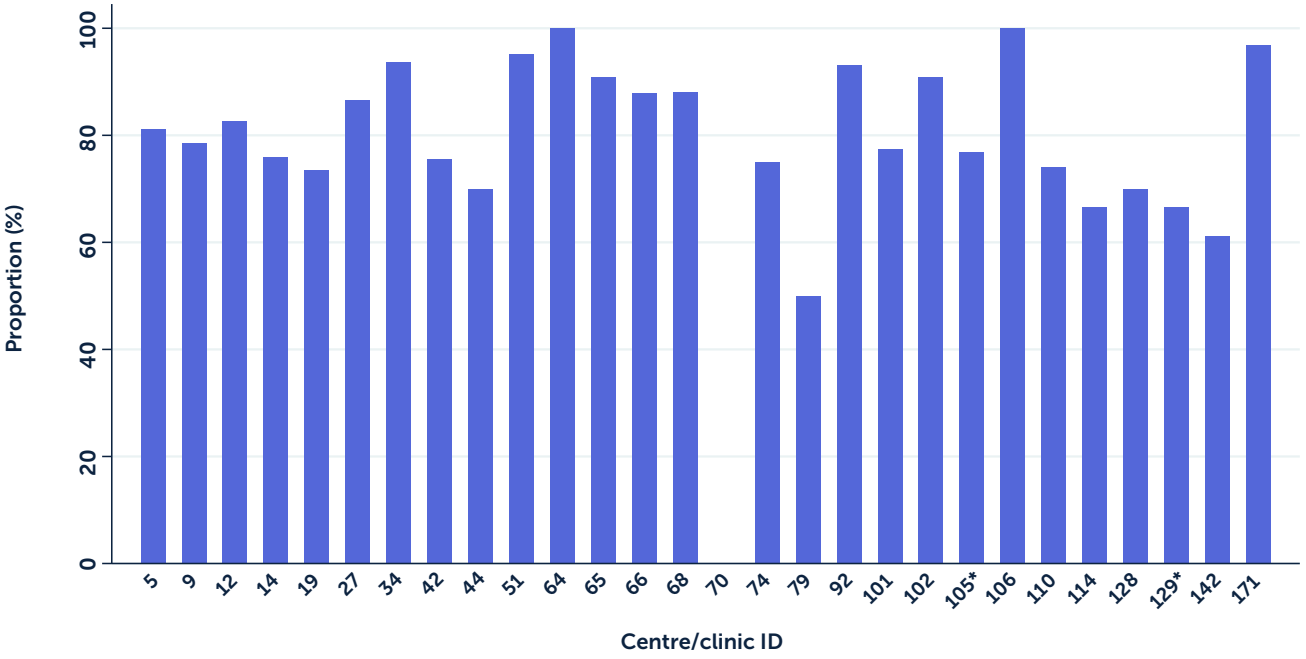
The chart below shows the proportion of patients with at least one IV day at home and/or in hospital. Patients may have a combination of home and hospital IV days.



The proportion of patients in adult centres receiving IV antibiotics at home was 13.3% and in hospital was 20.1%. The proportion receiving any IVs was 25.5%.

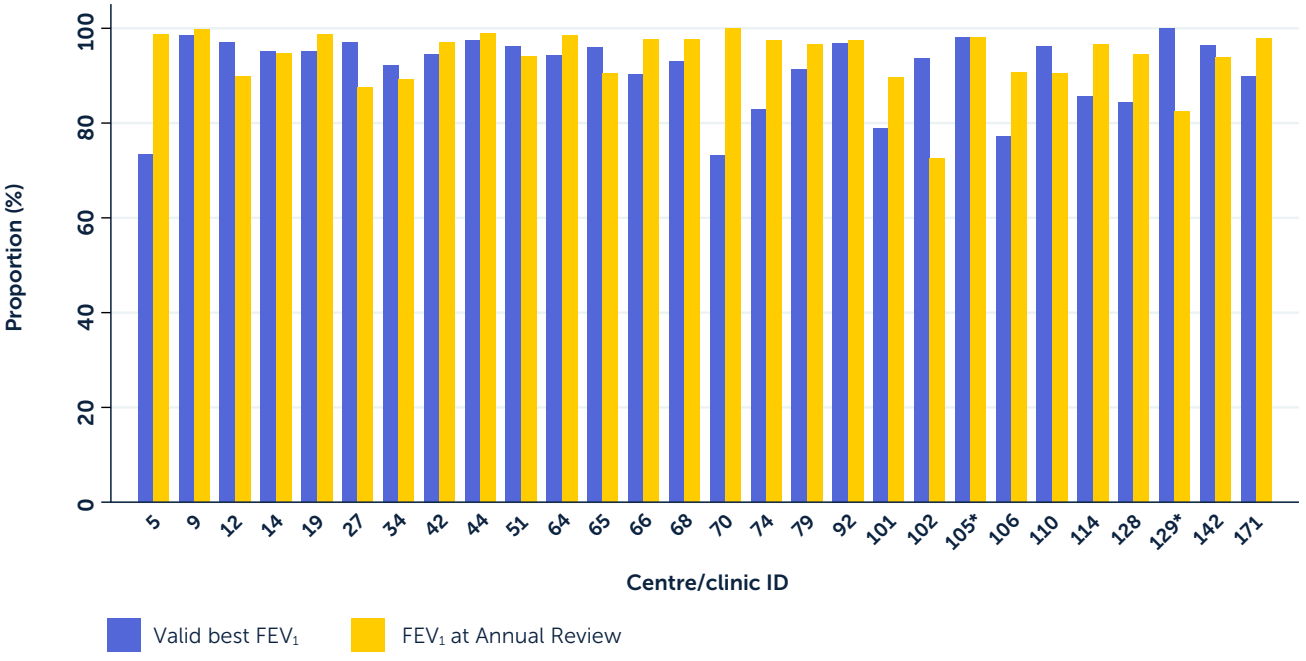
* Standalone clinics.

3.10 Inhaled antibiotic use for patients with chronic *Pseudomonas aeruginosa* by adult centre/clinic



82.2% of patients in adult centres with chronic *P. aeruginosa* received inhaled antibiotics. Centres with fewer than 10 people with chronic *P. aeruginosa* were excluded.

3.11 Data completeness by adult centre/clinic[1]



[1] FEV_1 was considered valid if it was not missing, and the percent predicted was not more than 0.5% lower than the annual review value. For some patients there may be medical reasons why FEV_1 could not be taken, so centres may not be able to get 100% completeness.

* Standalone clinics.

Glossary

Word/Phrase	Meaning
2023	1 January 2023–31 December 2023.
ABPA (allergic bronchopulmonary aspergillosis)	When a person develops a respiratory allergic reaction to *Aspergillus fumigatus*.
Arthritis	A condition causing pain and inflammation in the joints.
Arthropathy	A condition causing pain in the joints.
Asthma	A respiratory condition causing reversible episodes of difficulty breathing, often associated with wheezing.
***B. cepacia* complex**	The *Burkholderia cepacia* complex is a group of bacteria, some of which threaten the health of people with cystic fibrosis.
BMI (Body Mass Index)	A measure designed to show whether a person is a healthy weight for their height.
CF	Cystic fibrosis.
CFTR (cystic fibrosis transmembrane conductance regulator)	A protein at the cell surface that controls the salt and water balance across a cell. The gene that causes cystic fibrosis is the blueprint for the CFTR protein. Everyone has two copies of the gene for CFTR. To be born with cystic fibrosis, both CFTR genes must be affected by a CF-causing variant.
Chronic	Persistent, or long-lasting.
Cirrhosis	A chronic liver disease.
CI (confidence interval)	A way of expressing how certain we are about our statistical estimates of a clinical measure (for example BMI). It gives a range of results that is likely to include the 'true' value for the population. A narrow confidence interval indicates a more precise estimate. A wide confidence interval indicates more uncertainty about the true value of the clinical measure, often because a small group of patients has been studied. The confidence interval is usually stated as '95% CI', which means that the range of values has a 95 in 100 chance of including the 'true' value.
Enzymes	Biological molecules that help complex reactions, such as the digestion of food, occur in the body.
FEV_1 (forced expiratory volume in one second)	This is the amount of air that a person can blow out of the lungs in the first second of a forced exhaled breath. People with healthy lungs can blow out most of the air held in this time.
FEV_1% predicted	The FEV_1 can be converted from absolute litres of air blown out into a predicted percentage (%). A healthy range for % predicted is calculated from a very large population sample, and is normally considered to be between 80-120% predicted.
Fibrosing colonopathy	A condition causing narrowing of part of the colon.
Gall bladder	The small sac-shaped organ under the liver that stores bile after it is secreted by the liver, before it is released into the intestine.
Gastrointestinal (GI) tract	The GI tract is an organ system responsible for digesting food, absorbing nutrients and expelling waste.
Genotype	Part of the genetic makeup of a cell, organism or individual that usually controls a particular characteristic (known as a phenotype).
GERD (gastroesophageal reflux disease)	A chronic symptom of damage caused by stomach acid coming up from the stomach into the oesophagus.
GI bleed	Bleeding in the gastrointestinal tract.
GLI equations	Global Lung Initiative, the equation used for calculating FEV_1% predicted from absolute FEV_1, which takes into account age, gender, height and ethnicity.
H. influenza	*Haemophilus influenza* is a bacterium that can cause serious illness.
Haemoptysis	The coughing up of blood.
Hepatobiliary disease	A liver or biliary disorder.
Heterozygous	Everyone living with cystic fibrosis has two variants of the gene for CFTR, one inherited from their mother and one from their father. Someone who has two different CFTR variants is heterozygous.

Word/Phrase	Meaning
Homozygous	Everyone living with cystic fibrosis has two variants of the gene for CFTR, one inherited from their mother and one from their father. If both CFTR variants (or genotypes) are the same, the person is said to be homozygous.
Hypertension	High blood pressure.
Incidence	The number of people newly diagnosed with a condition in the given year.
IQR (interquartile range)	Also called the mid-spread, or middle fifty, IQR is a measure of the spread of data. It shows the difference between the upper and lower quartiles. IQR = Q3 – Q1.
Mean	A type of average, calculated by adding up all the values and dividing by the number of values.
Median	The middle number, when all numbers are arranged from smallest to largest.
Median age of death	Median age of death is based on the people with CF who died in any given year.
Median predicted survival age	A prediction of how long we expect half of the people with CF born today live for.
MRSA	Methicillin-resistant *Staphylococcus aureus* is a type of bacteria that is resistant to a number of widely used antibiotics.
Nasal polyps	Small, sac-like growths of inflamed mucus membrane caused by chronic inflammation of the nasal lining.
NBS (newborn screening)	Newborn screening is part of the heel prick blood spot testing carried out on all babies at 5–7 days of age. The blood sample is tested for a number of conditions, including cystic fibrosis.
NTM (non-tuberculous mycobacteria)	A mycobacterium that does not cause tuberculosis, but which can cause respiratory infection. There are several types known.
Osteopenia	A medical condition less severe than osteoporosis, where the mineral content of bone is reduced.
Osteoporosis	A condition where the bones become brittle from loss of tissue.
Pancreas	An organ in the digestive system that produces insulin and digestive enzymes.
Pancreatitis	Inflammation of the pancreas.
Peptic ulcer	Or stomach ulcer; an open sore that develops in the lining of the stomach.
Percentile	A percentile shows where a value stands, relative to the rest of the data. If a value is higher than 90% of the rest of the data, it is on the 90th percentile.
Pneumothorax	A collection of air in the cavity between the lungs and the chest wall causing collapse of the lung on the affected side.
Portal hypertension	High blood pressure in the portal vein system, which is the blood system of the liver.
Prenatal	Before birth, whilst the baby is still in the womb.
Prevalence	The overall number of people with the condition in the last 12 months.
Pseudomonas aeruginosa	A tough bacterial strain. Rarely affecting healthy people, it can cause a wide range of infections, particularly in those with a weakened immune system.
Rectal prolapse	When the rectal wall slides through the anus.
Renal	Relating to the kidneys.
Staphylococcus aureus	*Staphylococcus aureus* is a type of bacteria that can cause disease if it enters the body.
Sinus disease	When the sinuses, which are usually filled with air, are typically full of thick sticky mucus.
Statistically significant	This phrase means there is statistical evidence that the results we observe (such as a difference in median predicted survival age between males and females) are unlikely to have occurred due to chance.
Variant	A variant is a change in a gene. When both of a child's parents are carriers of a CF-causing variant there is a 25% chance that the child will have cystic fibrosis. There are over 1,400 different variants of the CFTR gene that can cause cystic fibrosis.

Appendix 1: UK CF Registry Committee structure

UK CF Registry Steering Committee

Role	Forename	Surname	Organisation
Director Research & Healthcare Data†	Lucy	Allen	Cystic Fibrosis Trust
NHS England Commissioner	Kathy	Blacker	NHS England
CF Physician - Paediatrics	Malcolm	Brodlie	Newcastle Paediatrics CF Centre
CF Physician - Paediatrics	Siobhán	Carr	Royal Brompton Hospital
Analytical team rep†	Susan	Charman	Cystic Fibrosis Trust
Associate Director of Data & QI#	Sarah	Clarke	Cystic Fibrosis Trust
CF Physician - Paediatrics	Gwyneth	Davies	UCL Great Ormond Street Institute of Child Health
CF Physician - Adults*	Jamie	Duckers	All Wales Adult CF Centre, Cardiff
Parent of Child with CF	Catherine	Farrer	N/A
Registry Clinical Data Manager†	Elaine	Gunn	Cystic Fibrosis Trust
Allied Health Professional	Rebecca	Heise	Kings College Adult CF Centre
Cystic Fibrosis Centre Data Manager	Erin	Hodgetts	North West Midlands Adult & Paediatrics CF Centres
Registry Systems Development Manager†	Kerry	Laidlaw	Cystic Fibrosis Trust
Welsh Commissioner	Richard	Palmer	NHS Wales
CF Physician - Adults	Simon	Range	Leicester Adult CF Centre
Scotland Representative	Helen	Rodgers	Western General Hospital
Chair of the UK CF Registry Research Committee*	Nick	Simmonds	Royal Brompton Hospital
Person with CF	Hannah	Gales	N/A
Head of Registry Operations (on Mat leave)†	Mary	Kisanga	Cystic Fibrosis Trust
Head of Registry Operations (Mat cover)†	Joanne	Osmond	Cystic Fibrosis Trust
Allied Health Professional	Jacqui	Cowlard	Royal London Hospital
Allied Health Professional	Joanna	Snowball	John Radcliffe Hospital, Oxford

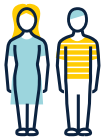
* Chair † Non-voting member # Caldicott guardian

UK CF Registry Research Committee

Role	Forename	Surname	Organisation
Director Research & Healthcare Data†	Lucy	Allen	Cystic Fibrosis Trust
Pharmacovigilance PI, Retired Professor in Respiratory Medicine	Diana	Bilton	N/A
Person with CF	Dawn	Bostock	N/A
Pharmacovigilance PI, CF Physician - Paediatrics	Siobhán	Carr	Royal Brompton Hospital
Analytical team rep†	Susan	Charman	Cystic Fibrosis Trust
Associate Director of Data & QI	Sarah	Clarke	Cystic Fibrosis Trust
Pharmacovigilance PI, CF Physician - Paediatrics	Steve	Cunningham	Royal Hospital for Sick Children, Edinburgh
CF Physician - Paediatrics	Francis	Gilchrist	University Hospitals of North Midlands NHS Trust
Registry Clinical Data Manager†	Elaine	Gunn	Cystic Fibrosis Trust
Head of Registry Operations (on Mat leave)†	Mary	Kisanga	Cystic Fibrosis Trust
Registry Systems Development Manager†	Kerry	Laidlaw	Cystic Fibrosis Trust
Pharmacovigilance PI, CF Physician - Adults	Dilip	Nazareth	Liverpool Heart and Chest Hospital, Liverpool
Head of Registry Operations (Mat cover)†	Joanne	Osmond	Cystic Fibrosis Trust
Pharmacovigilance PI, CF Physician - Adults*	Nick	Simmonds	Royal Brompton Hospital

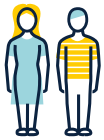
* Chair † Non-voting member # Caldicott guardian

Appendix 2: Centre-level data tables



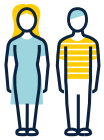
Paediatric centres/clinics providing data in 2023 – ordered alphabetically by country/city

Location	Name	Clinic ID	Total Active	Number with annual review
England				
Birmingham	Birmingham Children's Hospital	104	294	273
Brighton	Royal Alexandra Children's Hospital	172	62	60
Bristol	Bristol Royal Hospital for Children	32	189	173
Cambridge	Addenbrookes Hospital	107	151	147
Cornwall	Royal Cornwall Hospital	94	33	23
Exeter	Royal Devon & Exeter Hospital	96	75	71
Hull	Hull University Teaching Hospitals NHS Trust	111	43	42
Leeds	St James's University Hospital	25	237	221
Leicester	Leicester Royal Infirmary	1	74	72
Liverpool	Alder Hey Children's Hospital	97	303	280
London - Central	Great Ormond Street Hospital for Children	90	190	182
London - East	Royal London Hospital	30	85	80
London - South East	King's College Hospital	17	209	199
London - South West	Royal Brompton Hospital	15	272	261
Manchester	Royal Manchester Children's Hospital	144	327	308
Newcastle	Great North Children's Hospital	59	207	183
North West Midlands	University Hospital of North Midlands	8	97	94
Norwich	Norfolk & Norwich University Hospital	98	64	55
Nottingham	Nottingham University Hospitals	62	159	148
Oxford	John Radcliffe Hospital	22	168	157
Plymouth	Derriford Hospital	139	39	38
Sheffield	Sheffield Children's Hospital	3	149	135
Southampton	Southampton General Hospital	29	228	216
Teeside	James Cook University Hospital	71	55	53
Northern Ireland				
Belfast	Royal Belfast Hospital for Sick Children	60	211	196
Scotland				
Aberdeen	Royal Aberdeen Children's Hospital	75	29	22
Ayr	University Hospital Crosshouse	170	19	8
Dundee	Ninewells Hospital	73	21	20
Edinburgh	Royal Hospital for Sick Children	143	124	108
Glasgow	Royal Hospital for Sick Children	56	160	99
Inverness	Raigmore Hospital	31	17	16
Wales				
Cardiff	Children's Hospital for Wales	72	154	142



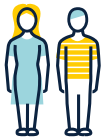
	Age		FEV$_1$% predicted at annual review				Best FEV$_1$% predicted			
Clinic ID	Mean	Median	Number	Mean - unadjusted	Mean - adjusted	Median	Number*	Mean - unadjusted	Mean - adjusted	Median
104	8.8	8.8	175	93.5	93.2	94.4	182	98.4	97.9	98.5
172	8.5	8.8	40	104.0	103.6	103.1	40	105.6	105.0	104.9
32	9.7	10.0	121	94.9	95.0	96.5	124	99.4	99.4	100.2
107	9.0	9.1	100	100.6	100.2	100.0	100	102.9	102.3	102.5
94	10.9	11.4	18	96.8	96.0	96.0	22	105.2	104.5	102.9
96	10.5	10.2	61	89.8	89.6	91.4	63	96.0	95.5	95.7
111	8.8	9.4	29	92.7	92.2	96.9	29	99.8	99.2	105.0
25	9.6	10.0	152	92.9	92.6	94.8	161	98.5	98.1	100.4
1	8.6	8.4	49	96.6	96.1	94.8	50	100.6	99.8	100.4
97	9.3	9.8	174	92.0	91.8	94.4	197	101.4	101.0	100.6
90	9.4	9.8	134	97.0	96.5	98.8	136	99.1	98.4	100.8
30	9.6	10.0	58	101.4	101.2	102.9	59	106.5	106.2	107.7
17	8.5	8.2	120	96.5	96.3	98.1	129	98.9	98.4	100.0
15	9.3	9.9	186	94.5	94.2	94.9	188	102.1	101.7	101.5
144	9.3	9.9	156	94.1	94.2	96.6	193	99.7	99.8	101.1
59	8.9	9.3	110	98.5	98.3	100.4	120	102.4	102.1	101.9
8	10.6	11.8	72	92.3	92.4	94.2	78	98.8	98.9	98.7
98	8.8	8.0	34	89.1	88.7	89.2	37	101.1	100.5	98.2
62	10.0	10.9	110	92.6	92.8	93.2	110	95.9	96.1	96.2
22	9.6	9.8	113	96.9	96.9	98.6	113	100.0	99.9	100.0
139	8.8	8.9	23	95.3	94.7	98.3	24	101.4	100.8	100.7
3	8.9	9.2	91	99.6	99.3	101.8	94	103.9	103.4	105.0
29	9.2	8.9	141	97.1	97.2	97.7	145	102.1	102.1	101.9
71	9.4	9.4	37	92.7	93.0	94.5	37	98.1	98.3	97.7
60	9.7	9.9	142	96.4	96.6	97.2	144	100.8	100.8	99.6
75	7.4	6.5	9	109.1	108.1	108.8	12	108.2	107.1	107.5
170	9.9	12.0	5	98.6	99.3	98.9	5	101.1	102.0	98.9
73	9.6	10.8	12	97.1	96.6	95.1	14	101.1	100.5	101.4
143	9.8	10.1	80	98.7	98.4	99.0	83	101.6	101.2	101.3
56	8.9	8.8	67	97.3	96.8	95.5	69	101.0	100.3	100.4
31	9.7	9.4	13	95.5	95.1	96.3	13	100.0	99.4	99.0
72	9.9	10.0	101	94.2	94.2	94.7	101	99.5	99.5	98.0

* Where 'Best' values were missing, or lower than FEV$_1$% predicted taken at annual review, the annual review value was used.



Paediatric centres/clinics providing data in 2023 – ordered alphabetically by country/city

			BMI			
Location	Name	Clinic ID	Number	Mean - unadjusted	Mean - adjusted	Median
England						
Birmingham	Birmingham Children's Hospital	104	245	54.7	54.7	52.8
Brighton	Royal Alexandra Children's Hospital	172	57	55.5	55.6	50.0
Bristol	Bristol Royal Hospital for Children	32	159	57.3	57.2	59.2
Cambridge	Addenbrookes Hospital	107	137	56.5	56.5	58.3
Cornwall	Royal Cornwall Hospital	94	22	66.1	66.0	70.2
Exeter	Royal Devon & Exeter Hospital	96	71	56.7	56.6	54.5
Hull	Hull University Teaching Hospitals NHS Trust	111	38	68.9	68.9	71.2
Leeds	St James's University Hospital	25	194	56.4	56.4	57.8
Leicester	Leicester Royal Infirmary	1	65	56.9	56.9	63.5
Liverpool	Alder Hey Children's Hospital	97	258	61.4	61.4	64.1
London - Central	Great Ormond Street Hospital for Children	90	174	50.9	50.9	49.9
London - East	Royal London Hospital	30	71	55.6	55.6	61.0
London - South East	King's College Hospital	17	172	53.9	53.9	57.5
London - South West	Royal Brompton Hospital	15	247	54.0	54.0	54.4
Manchester	Royal Manchester Children's Hospital	144	279	53.6	53.6	54.4
Newcastle	Great North Children's Hospital	59	160	58.7	58.7	58.2
North West Midlands	University Hospital of North Midlands	8	86	55.6	55.6	58.1
Norwich	Norfolk & Norwich University Hospital	98	54	63.0	63.1	67.3
Nottingham	Nottingham University Hospitals	62	137	57.8	57.8	60.0
Oxford	John Radcliffe Hospital	22	148	57.1	57.1	59.5
Plymouth	Derriford Hospital	139	36	60.0	60.0	61.8
Sheffield	Sheffield Children's Hospital	3	123	56.8	56.8	55.2
Southampton	Southampton General Hospital	29	194	58.8	58.8	63.3
Teeside	James Cook University Hospital	71	46	69.0	69.0	76.6
Northern Ireland						
Belfast	Royal Belfast Hospital for Sick Children	60	188	58.0	57.9	60.7
Scotland						
Aberdeen	Royal Aberdeen Children's Hospital	75	20	59.8	59.9	58.3
Ayr	University Hospital Crosshouse	170	5	54.9	54.7	53.5
Dundee	Ninewells Hospital	73	17	53.0	53.0	54.6
Edinburgh	Royal Hospital for Sick Children	143	104	60.5	60.5	61.4
Glasgow	Royal Hospital for Sick Children	56	90	56.1	56.1	58.8
Inverness	Raigmore Hospital	31	15	53.8	53.8	56.8
Wales						
Cardiff	Children's Hospital for Wales	72	135	55.0	54.9	56.1



	Chronic *pseudomonas*		Having at least 1 IV days		Receiving DNase treatment		Receiving hypertonic saline or mannitol treatment		Inhaled antibiotic use among patients with chronic *pseudomonas*	
Clinic ID	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)
104	6	2.3	49	17.9	151	55.3	102	37.4	6	100.0
172	<5	1.7	10	16.7	46	76.7	27	45.0	<5	100.0
32	6	3.5	23	13.3	139	80.3	108	62.4	6	100.0
107	<5	1.4	24	16.3	66	44.9	102	69.4	<5	100.0
94	<5	4.3	<5	4.3	11	47.8	8	34.8	<5	100.0
96	<5	1.4	13	18.3	68	95.8	68	95.8	0	0.0
111	<5	9.5	11	26.2	15	35.7	11	26.2	<5	25.0
25	9	4.2	33	14.9	125	56.6	29	13.1	9	100.0
1	<5	1.4	14	19.4	51	70.8	12	16.7	<5	100.0
97	5	1.8	54	19.3	128	45.7	32	11.4	<5	80.0
90	7	3.9	51	28.0	129	70.9	88	48.4	7	100.0
30	7	8.8	23	28.8	62	77.5	64	80.0	7	100.0
17	12	6.3	41	20.6	128	64.3	42	21.1	12	100.0
15	11	4.2	29	11.1	180	69.0	151	57.9	10	90.9
144	13	4.3	59	19.2	165	53.6	101	32.8	11	84.6
59	6	3.5	36	19.7	102	55.7	21	11.5	6	100.0
8	<5	1.1	30	31.9	74	78.7	28	29.8	<5	100.0
98	0	0.0	5	9.1	39	70.9	<5	7.3	0	0.0
62	0	0.0	18	12.2	73	49.3	36	24.3	0	0.0
22	0	0.0	24	15.3	106	67.5	59	37.6	0	0.0
139	0	0.0	<5	7.9	24	63.2	6	15.8	0	0.0
3	<5	3.0	32	23.7	97	71.9	32	23.7	<5	100.0
29	5	2.5	14	6.5	135	62.5	27	12.5	<5	80.0
71	<5	1.9	13	24.5	30	56.6	12	22.6	0	0.0
60	<5	0.5	17	8.7	167	85.2	33	16.8	<5	100.0
75	0	0.0	0	0.0	8	36.4	<5	4.5	0	0.0
170	0	0.0	<5	25.0	0	0.0	0	0.0	0	0.0
73	0	0.0	<5	5.0	5	25.0	<5	10.0	0	0.0
143	<5	0.9	19	17.6	63	58.3	21	19.4	<5	100.0
56	<5	1.0	12	12.1	21	21.2	35	35.4	<5	100.0
31	<5	6.2	0	0.0	7	43.8	<5	12.5	<5	100.0
72	<5	2.8	13	9.2	111	78.2	135	95.1	<5	100.0

* Redacted to adhere to statistical disclosure guidelines.

Appendix 2: Centre-level data tables

Adult centres/clinics providing data in 2023 – ordered alphabetically by country/city

Location	Name	Clinic ID	Total Active	Number with annual review
England				
Birmingham	Birmingham Heartlands Hospital	27	338	330
Bristol	Bristol Royal Infirmary	106	252	236
Cambridge	Royal Papworth Hospital	51	374	342
Cornwall	Royal Cornwall Hospital	129	44	40
Exeter	Royal Devon & Exeter Hospital	34	142	129
Frimley	Frimley Park Hospital	19	172	162
Leeds	St James's University Hospital	42	414	408
Leicester	Glenfield Hospital	142	117	113
Liverpool	Liverpool Heart and Chest Hospital	66	392	372
London - East	St Bartholomew's Hospital	92	224	195
London - South East	University Hospital Lewisham	105	57	55
London - South East	King's College Hospital	5	267	222
London - South West	Royal Brompton Hospital	12	620	606
Manchester	Wythenshawe Hospital	102	492	470
Newcastle	Royal Victoria Infirmary	9	348	325
North West Midlands	University Hospital of North Midlands	74	160	152
Norwich	Norfolk & Norwich University Hospital	114	93	91
Nottingham	Nottingham University Hospitals	101	258	250
Oxford	Oxford University Hospitals	128	167	128
Plymouth	Derriford Hospital	64	74	70
Sheffield	Northern General Hospital	65	226	223
Southampton	Southampton General Hospital	110	321	297
York and Hull	York Hospital	171	104	99
Northern Ireland				
Belfast	Belfast City Hospital	14	298	209
Scotland				
Aberdeen	Aberdeen Royal Infirmary	70	78	67
Edinburgh	Western General Hospital	44	282	268
Glasgow	Queen Elizabeth University Hospital	79	242	115
Wales				
Llandough	Llandough Hospital	68	319	288

	Age		FEV$_1$% predicted at annual review				Best FEV$_1$% predicted			
Clinic ID	Mean	Median	Number	Mean - unadjusted	Mean - adjusted	Median	Number*	Mean - unadjusted	Mean - adjusted	Median
27	34.7	32.0	278	73.1	73.0	74.3	308	75.2	75.4	78.1
106	33.5	32.0	208	75.3	75.1	79.6	213	79.0	79.0	83.7
51	33.5	31.3	305	76.4	76.1	79.1	312	78.3	78.1	80.4
129	35.9	33.4	29	73.8	73.7	71.3	36	74.8	74.5	75.4
34	35.5	32.9	108	78.6	79.7	79.2	120	83.0	83.8	87.7
19	32.8	32.0	154	77.0	76.3	80.3	154	80.0	79.4	83.1
42	36.7	35.1	359	72.1	73.4	75.4	364	75.3	76.7	79.4
142	32.6	30.7	101	76.3	75.1	81.7	105	78.4	77.5	83.2
66	33.1	31.8	356	79.5	78.9	81.3	360	82.2	81.7	84.6
92	29.9	26.8	188	74.9	72.1	76.7	191	77.4	74.7	80.6
105	33.6	31.8	48	65.7	65.7	64.7	48	69.5	69.6	68.4
5	33.0	31.4	212	72.1	71.9	76.3	212	74.9	74.9	78.3
12	36.9	34.7	545	72.9	74.5	74.2	593	75.9	77.5	77.0
102	33.8	32.0	341	70.8	71.4	72.9	447	75.5	75.9	74.7
9	33.8	31.2	304	72.8	72.2	77.9	305	75.2	74.7	79.7
74	31.5	28.6	142	72.9	70.9	78.1	143	76.4	74.6	79.6
114	31.6	29.9	83	79.1	77.4	82.7	85	84.0	82.4	85.6
101	32.2	30.6	212	75.4	74.1	76.7	222	79.3	78.2	81.3
128	33.1	29.1	115	75.9	74.1	77.5	118	80.6	78.9	80.0
64	36.9	34.8	66	72.7	72.9	77.7	66	76.6	76.9	81.3
65	32.8	31.5	197	76.3	75.6	81.0	211	80.3	79.7	83.5
110	35.1	32.2	250	76.5	76.6	77.8	265	79.4	79.5	80.2
171	34.8	32.1	95	71.8	71.9	68.9	97	76.4	76.6	75.1
14	38.2	35.8	195	73.6	75.3	78.2	205	75.0	76.8	79.2
70	35.2	33.4	60	72.0	71.8	74.3	60	75.8	75.8	77.6
44	34.3	32.0	253	74.5	73.9	77.7	254	79.3	79.0	83.1
79	33.7	31.0	109	77.3	76.9	80.1	113	79.6	79.3	83.4
68	33.8	31.6	265	75.3	74.9	79.6	269	79.7	79.4	83.3

* Where 'Best' values were missing, or lower than FEV$_1$% predicted taken at annual review, the annual review value was used.

Adult centres/clinics providing data in 2023 – ordered alphabetically by country/city

			BMI			
Location	Name	Clinic ID	Number	Mean - unadjusted	Mean - adjusted	Median
England						
Birmingham	Birmingham Heartlands Hospital	27	325	24.6	24.6	24.0
Bristol	Bristol Royal Infirmary	106	231	24.5	24.5	24.0
Cambridge	Royal Papworth Hospital	51	334	24.3	24.4	23.7
Cornwall	Royal Cornwall Hospital	129	40	24.9	24.9	22.9
Exeter	Royal Devon & Exeter Hospital	34	127	25.4	25.3	24.5
Frimley	Frimley Park Hospital	19	162	23.8	23.9	23.4
Leeds	St James's University Hospital	42	408	24.6	24.4	24.2
Leicester	Glenfield Hospital	142	111	24.3	24.5	23.6
Liverpool	Liverpool Heart and Chest Hospital	66	372	24.9	25.0	24.2
London - East	St Bartholomew's Hospital	92	195	23.2	23.6	22.6
London - South East	University Hospital Lewisham	105	55	22.7	22.8	22.3
London - South East	King's College Hospital	5	222	24.2	24.2	23.0
London - South West	Royal Brompton Hospital	12	606	24.0	23.8	23.5
Manchester	Wythenshawe Hospital	102	448	23.9	23.9	23.3
Newcastle	Royal Victoria Infirmary	9	323	24.9	24.9	24.1
North West Midlands	University Hospital of North Midlands	74	152	24.1	24.3	23.7
Norwich	Norfolk & Norwich University Hospital	114	90	23.7	23.9	22.6
Nottingham	Nottingham University Hospitals	101	250	23.8	23.9	22.9
Oxford	Oxford University Hospitals	128	128	23.5	23.7	22.7
Plymouth	Derriford Hospital	64	70	24.7	24.6	23.8
Sheffield	Northern General Hospital	65	223	25.2	25.3	24.4
Southampton	Southampton General Hospital	110	293	24.6	24.5	23.8
York and Hull	York Hospital	171	99	24.1	24.1	23.3
Northern Ireland						
Belfast	Belfast City Hospital	14	208	25.5	25.2	25.0
Scotland						
Aberdeen	Aberdeen Royal Infirmary	70	66	26.1	26.0	25.0
Edinburgh	Western General Hospital	44	268	24.4	24.5	23.7
Glasgow	Queen Elizabeth University Hospital	79	115	24.6	24.6	24.0
Wales						
Llandough	Llandough Hospital	68	286	24.7	24.7	23.8

	Chronic *pseudomonas*		Having at least 1 IV days		Receiving DNase treatment		Receiving hypertonic saline or mannitol treatment		Inhaled antibiotic use among patients with chronic *pseudomonas*	
Clinic ID	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)
27	45	14.3	100	30.3	189	57.3	110	33.3	39	86.7
106	9	4.5	67	28.4	131	55.5	123	52.1	9	100.0
51	21	7.0	90	26.3	160	46.8	116	33.9	20	95.2
129	9	24.3	14	35.0	21	52.5	12	30.0	6	66.7
34	16	13.3	20	15.5	75	58.1	63	48.8	15	93.8
19	34	21.0	34	21.0	73	45.1	37	22.8	25	73.5
42	45	14.2	147	36.0	241	59.1	43	10.5	34	75.6
142	18	26.1	26	23.0	54	47.8	17	15.0	11	61.1
66	33	10.4	78	21.0	272	73.1	119	32.0	29	87.9
92	29	15.7	49	25.1	147	75.4	117	60.0	27	93.1
105	13	23.6	24	43.6	37	67.3	11	20.0	10	76.9
5	16	7.8	65	29.3	158	71.2	90	40.5	13	81.2
12	58	11.5	129	21.3	533	88.0	282	46.5	48	82.8
102	55	12.8	139	29.6	210	44.7	129	27.4	50	90.9
9	56	17.6	74	22.8	197	60.6	59	18.2	44	78.6
74	16	10.7	43	28.3	100	65.8	58	38.2	12	75.0
114	<5	3.7	19	20.9	42	46.2	24	26.4	<5	66.7
101	31	13.8	80	32.0	153	61.2	87	34.8	24	77.4
128	20	16.0	23	18.0	85	66.4	41	32.0	14	70.0
64	11	15.9	19	27.1	47	67.1	36	51.4	11	100.0
65	22	10.5	80	35.9	183	82.1	29	13.0	20	90.9
110	27	9.4	58	19.5	145	48.8	70	23.6	20	74.1
171	32	32.7	22	22.2	70	70.7	17	17.2	31	96.9
14	50	24.2	27	12.9	118	56.5	35	16.7	38	76.0
70	0	0.0	9	13.4	20	29.9	<5	3.0	0	0.0
44	20	10.9	46	17.2	136	50.7	30	11.2	14	70.0
79	12	11.3	31	27.0	36	31.3	9	7.8	6	50.0
68	59	21.7	84	29.2	193	67.0	179	62.2	52	88.1

* Redacted to adhere to statistical disclosure guidelines.

Appendix 3: Full list of CFTR variants in the UK CF population

The table below shows the number of people with CF who carry at least one of each variant.

The groups are not mutually exclusive, as people with heterozygous variants appear twice in the table.

Nucleotide	Protein	Legacy name	N	%
c.1521_1523delCTT	p.Phe508del	F508del	10073	89.0
c.350G->A	p.Arg117His	R117H	714	6.3
c.1652G->A	p.Gly551Asp	G551D	641	5.7
c.1624G->T	p.Gly542X	G542X	418	3.7
c.489+1G->T		621+1G->T	292	2.6
c.3909C->G	p.Asn1303Lys	N1303K	182	1.6
c.1585-1G->A		1717-1G->A	181	1.6
c.3454G->C	p.Asp1152His	D1152H	163	1.4
c.1766+1G->A		1898+1G->A	160	1.4
c.200C->T	p.Pro67Leu	P67L	155	1.4
c.3140-26A->G		3272-26A->G	129	1.1
c.3528delC	p.Lys1177SerfsX15	3659delC	127	1.1
c.1679G->C	p.Arg560Thr	R560T	106	0.9
c.1477C->T	p.Gln493X	Q493X	95	0.8
c.1519_1521delATC	p.Ile507del	I507del	94	0.8
c.1657C->T	p.Arg553X	R553X	88	0.8
c.254G->A	p.Gly85Glu	G85E	87	0.8
c.2657+5G->A		2789+5G->A	87	0.8
c.3717+12191C->T		3849+10kbC->T	86	0.8
c.178G->T	p.Glu60X	E60X	80	0.7
c.1022_1023insTC	p.Phe342HisfsX28	1154insTC	74	0.7
c.3846G->A	p.Trp1282X	W1282X	67	0.6
c.617T->G	p.Leu206Trp	L206W	59	0.5
c.1364C->A	p.Ala455Glu	A455E	58	0.5
c.1646G->A	p.Ser549Asn	S549N	57	0.5
c.2052delA	p.Lys684AsnfsX38	2184delA	53	0.5
c.948delT	p.Phe316LeufsX12	1078delT	52	0.5
c.1040G->C	p.Arg347Pro	R347P	47	0.4
c.2657+2_2657+3insA		2789+2insA	43	0.4
c.3718-2477C->T		3849+10kbC->T	40	0.4
c.579+3A->G		711+3A->G	40	0.4
c.3484C->T	p.Arg1162X	R1162X	32	0.3
c.1558G->T	p.Val520Phe	V520F	32	0.3
c.1040G->A	p.Arg347His	R347H	29	0.3
c.1753G->T	p.Glu585X	E585X	29	0.3
c.1000C->T	p.Arg334Trp	R334W	28	0.2
c.2988+1G->A		3120+1G->A	28	0.2
c.1367T->C	p.Val456Ala	V456A	28	0.2

Nucleotide	Protein	Legacy name	N	%
c.3472C->T	p.Arg1158X	R1158X	26	0.2
c.1523T->G	p.Phe508Cys	F508C	26	0.2
c.1055G->A	p.Arg352Gln	R352Q	25	0.2
c.1705T->G	p.Tyr569Asp	Y569D	23	0.2
c.3197G->A	p.Arg1066His	R1066H	23	0.2
c.1006_1007insG	p.Ile336SerfsX28	1138insG	23	0.2
c.1393-1G->A		1525-1G->A	22	0.2
c.3873G->C	p.Gln1291His	Q1291H	21	0.2
c.2490+1G->A		2622+1G->A	21	0.2
c.2125C->T	p.Arg709X	R709X	21	0.2
c.349C->T	p.Arg117Cys	R117C	20	0.2
c.2583delT	p.Phe861LeufsX3	2711delT	20	0.2
c.2052_2053insA	p.Gln685ThrfsX4	2184insA	19	0.2
c.532G->A	p.Gly178Arg	G178R	19	0.2
c.1210-12[5](AJ574948.1: g.152T[5])		5T	19	0.2
c.2834C->T	p.Ser945Leu	S945L	19	0.2
c.3806T->A	p.Ile1269Asn	I1269N	18	0.2
c.[1521_1523delCTT; 3080T->C]	p.[Phe508del; Ile1027Thr]	F508del;I1027T	16	0.1
c.658C->T	p.Gln220X	Q220X	16	0.1
c.3737C->T	p.Thr1246Ile	T1246I	16	0.1
c.579+1G->T		711+1G->T	15	0.1
c.292C->T	p.Gln98X	Q98X	13	0.1
c.2537G->A	p.Trp846X	W846X	13	0.1
c.2875delG	p.Ala959HisfsX9	3007delG	13	0.1
c.[1210–12[5];1210-34TG[12]]		5T;TG12	12	0.1
c.54-5940_273+10250del21kb	p.Ser18ArgfsX16	CFTRdele2,3	12	0.1
c.2051_2052delAAinsG	p.Lys684SerfsX38	2183AA->G or 2183delAA->G	12	0.1
c.1029delC	p.Cys343X	1161delC	12	0.1
c.1329_1330insAGAT	p.Ile444ArgfsX3	1461ins4	12	0.1
c.1466C->A	p.Ser489X	S489X	11	0.1
c.1645A->C or c.1647T->G or c.1647T->A	p.Ser549Arg	S549R	11	0.1
c.1679+1G->C		1811+1G->C	11	0.1
c.3196C->T	p.Arg1066Cys	R1066C	11	0.1
c.3468G->A		3600G->A	10	0.1
c.3761T->G	p.Leu1254X	L1254X	10	0.1

Nucleotide	Protein	Legacy name	N	%
c.2988G->A		3120G->A	10	0.1
c.1687T->A	p.Tyr563Asn	Y563N	9	0.1
c.224G->A	p.Arg75Gln	R75Q	9	0.1
c.1675G->A	p.Ala559Thr	A559T	9	0.1
c.3705T->G	p.Ser1235Arg	S1235R	9	0.1
c.3208C->T	p.Arg1070Trp	R1070W	9	0.1
c.709C->G	p.Gln237Glu	Q237E	8	0.1
c.695T->A	p.Val232Asp	V232D	8	0.1
c.1721C->A	p.Pro574His	P574H	8	0.1
c.2353C->T	p.Arg785X	R785X	8	0.1
c.494T->C	p.Leu165Ser	L165S	8	0.1
c.3353C->T	p.Ser1118Phe	S1118F	8	0.1
c.1986_1989delAACT	p.Thr663ArgfsX8	2118del4	7	0.1
c.262_263delTT	p.Leu88IlefsX22	394delTT	7	0.1
c.[1210–12[5];1210-34TG[13]]		5T;TG13	7	0.1
c.2900T->C	p.Leu967Ser	L967S	7	0.1
c.4196_4197delTC	p.Cys1400X	4326delTC	7	0.1
c.2012delT	p.Leu671X	2143delT	7	0.1
c.2991G->C	p.Leu997Phe	L997F	6	0.1
c.1766+1G->T		1898+1G->T	6	0.1
c.[1210–12[5];1210-34TG[11]]		5T;TG11	6	0.1
c.2128A->T	p.Lys710X	K710X	6	0.1
c.1538A->G	p.Asp513Gly	D513G	6	0.1
c.223C->T	p.Arg75X	R75X	6	0.1
c.2290C->T	p.Arg764X	R764X	6	0.1
c.349C->G	p.Arg117Gly	R117G	6	0.1
c.3718-1G->A		3850-1G->A	5	0.0
c.3884_3885insT	p.Ser1297PhefsX5	4016insT	5	0.0
c.1046C->T	p.Ala349Val	A349V	5	0.0
c.2491G->T	p.Glu831X	E831X	5	0.0
c.3292T->C	p.Trp1098Arg	W1098R	5	0.0
c.3848G->T	p.Arg1283Met	R1283M	5	0.0
c.1393-2A->G		1525-2A->G	5	0.0
c.1679G->A	p.Arg560Lys	R560K	5	0.0
c.1116+1G->A		1248+1G->A	5	0.0
c.443T->C	p.Ile148Thr	I148T	5	0.0
c.2551C->T	p.Arg851X	R851X	5	0.0
c.4147_4148insA	p.Ile1383AsnfsX3	4279insA	<5	-
c.3988C->T	p.Gln1330X	Q1330X	<5	-
c.850dupA	p.Met284AsnfsX3	977insA	<5	-
c.2249C->T	p.Pro750Leu	P750L	<5	-
c.(743+1_744-1)_(1584+1_1585-1)dup		CFTRdup6b-10	<5	-
c.1585-8G->A		1717-8G->A	<5	-
c.3964-78_4242+577del		CFTRdele22,23	<5	-
c.2600_2601insA	p.Val868SerfsX28	2732insA	<5	-
c.2896delA	p.Thr966ArgfsX2	3028delA	<5	-
c.1736A->G	p.Asp579Gly	D579G	<5	-

Nucleotide	Protein	Legacy name	N	%
c.1584G->A	p.Glu528Glu	1716G/A	<5	-
c.1545_1546delTA	p.Tyr515X	1677delTA	<5	-
c.2215delG	p.Val739TyrfsX16	2347delG	<5	-
c.165-3C>T		297-3C->T	<5	-
c.2909G->A	p.Gly970Asp	G970D	<5	-
c.2464G->T	p.Glu822X	E822X	<5	-
c.429delT	p.Phe143LeufsX10	557delT	<5	-
c.3095A->G	p.Tyr1032Cys	Y1032C	<5	-
c.595C->T	p.His199Tyr	H199Y	<5	-
c.509G->A	p.Arg170His	R170H	<5	-
c.1680A->C	p.Arg560Ser	R560S	<5	-
c.233dupT	p.Trp79LeufsX32	365-366insT	<5	-
c.1572C->A	p.Cys524X	C524X	<5	-
c.3080T->C	p.Ile1027Thr	I1027T	<5	-
c.3872A->G	p.Gln1291Arg	Q1291R	<5	-
c.1327G->T	p.Asp443Tyr	D443Y	<5	-
c.91C->T	p.Arg31Cys	R31C	<5	-
c.1210–12[7]		7T	<5	-
c.350G->T	p.Arg117Leu	R117L	<5	-
c.2374C->T	p.Arg792X	R792X	<5	-
c.4004T->C	p.Leu1335Pro	L1335P	<5	-
c.1727G->C	p.Gly576Ala	G576A	<5	-
c.3752G->A	p.Ser1251Asn	S1251N	<5	-
c.577G->T	p.Glu193X	E193X	<5	-
c.3659delC	p.Thr1220LysfsX8	3791delC	<5	-
c.79G->T	p.Gly27X	G27X	<5	-
c.274G->A	p.Glu92Lys	E92K	<5	-
c.328G->C	p.Asp110His	D110H	<5	-
c.1505T>C	p.Ile502Thr	I502T	<5	-
c.1724T->A	p.Phe575Tyr	F575Y	<5	-
c.4046G->A	p.Gly1349Asp	G1349D	<5	-
c.4111G->T	p.Glu1371X	E1371X	<5	-
c.933C>G	p.Phe311Leu	F311L	<5	-
c.1651G->A	p.Gly551Ser	G551S	<5	-
c.(53+1_54-1)_ (489+1_490-1)del		CFTRdele2-4	<5	-
c.1340delA	p.Lys447ArgfsX2	1471delA	<5	-
c.3476C->T	p.Ser1159Phe	S1159F	<5	-
c.(273+1_274-1)_ (1679+1_1680-1)del		CFTRdele4-11	<5	-
c.3017C->A	p.Ala1006Glu	A1006E	<5	-
c.1046C>T	p.Ala349Val	A349V	<5	-
c.164+2T>C		296+2T->C	<5	-
c.2195T->G	p.Leu732X	L732X	<5	-
c.263T>A or c.263T>G	p.Leu88X	L88X	<5	-
c.1766+5G->T		1898+5G->T	<5	-
c.3205G->A	p.Gly1069Arg	G1069R	<5	-
c.3266G->A	p.Trp1089X	W1089X	<5	-
c.1766+3A->G		1898+3A->G	<5	-

Nucleotide	Protein	Legacy name	N	%
c.1001G>A	p.Arg334Gln	R334Q	<5	-
c.1679+1.6kbA->G		1811+1.6kbA->G	<5	-
c.2260G->A	p.Val754Met	V754M	<5	-
c.2780T->C	p.Leu927Pro	L927P	<5	-
c.442delA	p.Ile148LeufsX5	574delA	<5	-
c.1766+1G->C		1898+1G->C	<5	-
c.413_415dupTAC	p.Leu138dup	L138ins	<5	-
c.2855T->C	p.Met952Thr	M952T	<5	-
c.273+1G->A		405+1G->A	<5	-
c.3475T->C	p.Ser1159Pro	S1159P	<5	-
c.1477_1478delCA	p.Gln493ValfsX10	1609delCA	<5	-
c.1007T->A	p.Ile336Lys	I336K	<5	-
c.3763T->C	p.Ser1255Pro	S1255P	<5	-
c.296C->T	p.Pro99Leu	P99L	<5	-
c.3158C->T	p.Thr1053Ile	T1053I	<5	-
c.4077_4080delTGTTinsAA	p.Val1360delfsX?	4209TGTT->AA	<5	-
c.601G->A	p.Val201Met	V201M	<5	-
c.1135G->T	p.Glu379X	E379X	<5	-
c.3310G->T	p.Glu1104X	E1104X	<5	-
c.2668C->T	p.Gln890X	Q890X	<5	-
c.3882_3885delTATT	p.Ile1295PhefsX32	4010del4	<5	-
c.2930C->T	p.Ser977Phe	S977F	<5	-
c.220C->T	p.Arg74Trp	R74W	<5	-
c.1682C->A	p.Ala561Glu	A561E	<5	-
c.3297C->A	p.Phe1099Leu	F1099L	<5	-
c.1670delC	p.Ser557PhefsX2	1802delC	<5	-
c.3908delA	p.Asn1303ThrfsX25	4040delA	<5	-
c.3368-2A->G		3500-2A->G	<5	-
c.2175_2176insA	p.Glu726ArgfsX4	2307insA	<5	-
c.(53+1_54-1)_(164+1_165-1)del		CFTRdele2	<5	-
c.137C->A	p.Ala46Asp	A46D	<5	-
c.3764C->A	p.Ser1255X	S1255X	<5	-
c.1210-12[5](AJ574948.1:g.152T[7])		7T	<5	-
c.1013C->T	p.Thr338Ile	T338I	<5	-
c.613C->T	p.Pro205Ser	P205S	<5	-
c.3808G->A	p.Asp1270Asn	D1270N	<5	-
c.1A->G	p.Met1Val	M1V	<5	-
c.50delT	p.Phe17SerfsX8	182delT	<5	-
c.11C>A	p.Ser4X	S4X	<5	-
c.3718-3T->G		3850-3T->G	<5	-
c.274-2A->G		406-2A->G	<5	-
c.3209G->A	p.Arg1070Gln	R1070Q	<5	-
c.3717+5G->A		3849+5G->A	<5	-
c.1420G->A	p.Glu474Lys	E474K	<5	-
c.1703delT	p.Leu568CysfsX4	1833delT	<5	-
c.1882G->C or c.1882G->A	p.Gly628Arg	G628R	<5	-
c.3700A->G	p.Ile1234Val	I1234V	<5	-

Nucleotide	Protein	Legacy name	N	%
c.2421A->G	p.Ile807Met	I807M	<5	-
c.2002C->T	p.Arg668Cys	R668C	<5	-
c.571T->G	p.Phe191Val	F191V	<5	-
c.3458T->A	p.Val1153Glu	V1153E	<5	-
c.1209+1G->A		1341+1G->A	<5	-
c.1117-1G>A		1249-1G->A	<5	-
c.4364C->G	p.Ser1455X	S1455X	<5	-
c.1837G->A	p.Ala613Thr	A613T	<5	-
c.859_863delAACTT	p.Asn287LysfsX19	991del5	<5	-
c.1654C->T	p.Gln552X	Q552X	<5	-
c.2989-1G->A		3121-1G->A	<5	-
c.2645G->A	p.Trp882X	W882X	<5	-
c.3717G->A		3849G->A	<5	-
c.-8G->C		125G/C	<5	-
c.1801A->T	p.Ile601Phe	I601F	<5	-
c.2735C->A	p.Ser912X	S912X	<5	-
c.(3873+1_3874-1)_ (3963+1_3964-1)del		CFTRdele21	<5	-
c.1573C->T	p.Gln525X	Q525X	<5	-
c.3011_3019delCTATAGCAG or c.3009_3017delAGCTATAGC	p.Ala1004_Ala1006del	3143del9	<5	-
c.164+1G>A		296+1G->A	<5	-
c.3745G->A	p.Gly1249Arg	G1249R	<5	-
c.53+1G->T		185+1G->T	<5	-
c.470_483del14	p.Phe157X	602del14	<5	-
c.3971T->C	p.Leu1324Pro	L1324P	<5	-
c.3587C->G	p.Ser1196X	S1196X	<5	-
c.2739T->A	p.Tyr913X	Y913X	<5	-
c.717delG	p.Leu240X	849delG	<5	-
c.(2988+1_2989-1)_ (3468+1_3469-1)del		CFTRdele17a-18	<5	-
c.933_935delCTT	p.Phe312del	F311del	<5	-
c.1418delG	p.Gly473GlufsX54	1548delG	<5	-
c.3773_3774insT	p.Leu1258PhefsX7	3905insT	<5	-
c.3181G->C	p.Gly1061Arg	G1061R	<5	-
c.3230T->C	p.Leu1077Pro	L1077P	<5	-
c.1687T->G	p.Tyr563Asp	Y563D	<5	-
c.3302T->A	p.Met1101Lys	M1101K	<5	-
c.3873+2T->C		4005+2T->C	<5	-
c.3435G->A	p.Trp1145X	W1145X	<5	-
c.1081delT	p.Trp361GlyfsX8	1213delT	<5	-
c.2859_2890del ACATTCTGTTCTTC AAGCACCTATGTCAACCC	p.Leu953PhefsX11	2991del32	<5	-
c.1037T->C	p.Leu346Pro	L346P	<5	-
c.274-1G->A		406-1G->A	<5	-
c.2620-26A->G		2752-26A->G	<5	-
c.4231C->T	p.Gln1411X	Q1411X	<5	-
c.3194T->C	p.Leu1065Pro	L1065P	<5	-
'Other' selected			675	6.0

Appendix 4: Legacy names lists for modulator eligibility*

List 4a: FDA list of CFTR variants potentially responsive to elexacaftor/ tezacaftor/ ivacaftor[1]

3141del9	E822K	G1244E	L997F	R117P	S945L
546insCTA	F191V	G1249R	L1077P	R170H	S977F
A46D	F311del	G1349D	L1324P	R258G	S1159F
A120T	F311L	H139R	L1335P	R334L	S1159P
A234D	F508C	H199Y	L1480P	R334Q	S1251N
A349V	F508C;S1251N	H939R	M152V	R347H	S1255P
A455E	F575Y	H1054D	M265R	R347L	T338I
A554E	F1016S	H1085P	M952I	R347P	T1036N
A1006E	F1052V	H1085R	M952T	R352Q	T1053I
A1067T	F1074L	H1375P	M1101K	R352W	V201M
D110E	F1099L	I148T	P5L	R553Q	V232D
D110H	G27R	I175V	P67L	R668C	V456A
D192G	G85E	I336K	P205S	R751L	V456F
D443Y	G126D	I502T	P574H	R792G	V562I
D443Y;G576A;R668C	G178E	I601F	Q98R	R933G	V754M
D579G	G178R	I618T	Q237E	R1066H	V1153E
D614G	G194R	I807M	Q237H	R1070Q	V1240G
D836Y	G194V	I980K	Q359R	R1070W	V1293G
D924N	G314E	I1027T	Q1291R	R1162L	W361R
D979V	G463V	I1139V	R31L	R1283M	W1098C
D1152H	G480C	I1269N	R74Q	R1283S	W1282R
D1270N	G551D	I1366N	R74W	S13F	Y109N
E56K	G551S	K1060T	R74W;D1270N	S341P	Y161D
E60K	G576A	L15P	R74W;V201M	S364P	Y161S
E92K	G576A;R668C	L165S	R74W;V201M;D1270N	S492F	Y563N
E116K	G622D	L206W	R75Q	S549N	Y1014C
E193K	G628R	L320V	R117C	S549R	Y1032C
E403D	G970D	L346P	R117G	S589N	
E474K	G1061R	L453S	R117H	S737F	
E588V	G1069R	L967S	R117L	S912L	

[1] https://pi.vrtx.com/files/uspi_elexacaftor_tezacaftor_ivacaftor.pdf

List 4b: potentially responsive CFTR variants according to French Compassionate Use Programme[2]

R334W	3849+10kbC->T
R1066C	3272-26A->G
2789+5G->A	3041-15T->G
	N1303K

[2] Burgel PR et al, Eur Respir J. Feb. 2023

* Please see graphs 1.35b and 1.36 in the report which reference these lists

List 4c: CFTR variants considered suitable for tezacaftor / ivacaftor use[3]

Named variants	E56K	P67L	D110H
	R117C	E193K	R347H
	L206W	R352Q	A455E
	711+3A->G	E831X	S945L
	K1060T	A1067T	2789+5G->A
	3272-26A->G	3849+10kbC->T	
Variants with "varying clinical consequence" (VCC)	R74W	D110E	D579G
	S977F	F1052V	R1070W
	F1074L	D1152H	D1270N

[3] https://www.england.nhs.uk/wp-content/uploads/2020/08/Urgent-policy-statement-CFTR-off-label-rarer-mutations.pdf

List 4d: CFTR variants considered suitable for ivacaftor use[3]

Named variants	E56K	P67L	D110H
	R117C	E193K	R347H
	L206W	R352Q	A455E
	711+3A->G	E831X	S945L
	K1060T	A1067T	2789+5G->A
	3272-26A->G	3849+10kbC->T	
Variants with "varying clinical consequence" (VCC)	R74W	D110E	D579G
	S977F	F1052V	G1069R
	R1070Q	R1070W	F1074L
	D1152H	D1270N	

[3] https://www.england.nhs.uk/wp-content/uploads/2020/08/Urgent-policy-statement-CFTR-off-label-rarer-mutations.pdf

Table 4e : CFTR modulator use in people aged six years and older by genotype group[4,5]

		Genotype Group				
		Group 1	**Group 2**	**Group 3**	**Group 4**	**Group 5**
Most recent CFTRm Used	ETI*	4135 (92.4)	3081 (80.3)	221 (45.9)	-**	16 (3.9)
	Tezacaftor/ivacaftor	-**	69 (1.8)	-**	48 (63.2)	<5
	Lumacaftor/ivacaftor	98 (2.2)	0 (0)	<5	0 (0)	<5
	Ivacaftor	<5	173 (4.5)	112 (23.2)	<5	<5
	Never Used	**176 (3.9)**	**515 (13.4)**	**135 (28)**	**15 (19.7)**	**390 (95.1)**
	Total	**4476 (100)**	**3838 (100)**	**482 (100)**	**76 (100)**	**410 (100)**

[4] 7 patients were excluded because their last recorded CFTRm treatment was as part of a drug trial and specific drug was unknown
[5] 27 people were excluded because their genotype is missing or unknown
* ETI is Elexacaftor/Tezacaftor/ivacaftor
** Redacted to adhere to statistical guidelines

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