Cystic Fibrosis Trust

UK Cystic Fibrosis Registry
2024 Annual Data Report

October 2025

Uniting for a life unlimited

UK Cystic Fibrosis Registry
2024 Annual Data Report

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

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Cystic Fibrosis Trust

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Foreword

I'm pleased to share with you the UK Cystic Fibrosis Registry 2024 Annual Data Report.

The data presented in this report provides valuable insights into the health of people living with cystic fibrosis (CF) today, helping us to shape our understanding and work to ensure all people can live a life without limits imposed by CF.

Over recent years, we have witnessed significant progress in CF: improvements in diagnosis, management of infections, and inhaled and CFTR modulator therapies have all contributed to our CF population increasing in size and growing older. In 2024, 65% of all people on the UK CF Registry were aged 16 or older, with more than 260 people aged 60 or over. The median predicted survival continues to increase – in 2024, we predict half of people born today will live to at least 66 years. However, this still means, that sadly, half may die before this age, possibly decades earlier. Across the population as a whole, the positive trends in improving lung health observed in the last few years were sustained in 2024. Improvements in lung function (median FEV_1 percent predicted) are seen across all age groups and the proportion of people receiving IV antibiotics both at home and in hospital, typically received after a pulmonary exacerbation, has decreased since last year, with 20% of people receiving IV antibiotics in 2024.

Yet challenges remain for many living with CF, including managing complications like CF diabetes and liver disease. In 2024, more than 27% of people living with CF were also dealing with treatments for CF diabetes; over 23% of adults were living with liver disease, a 5% increase since 2019. Other complications of the gut, including gastro-oesophageal reflux disease (GORD) and Distal Intestinal Obstruction Syndrome (DIOS), continue to impact some people with CF. Almost 80% of people were prescribed pancreatic enzyme supplementation. While nutritional status is improving, there is an increasing proportion of adults with higher BMIs, indicating changing health needs for some.

After more than a decade of campaigning, in 2024 the CFTR modulator therapies were permanently approved for use on the NHS across the UK. Data collected by the Registry over several years played a vital role in that decision. We report over 85% of people with CF had used a CFTR modulator therapy during 2024. Yet we know that they are not a cure and some people cannot benefit from these treatments. In section 1.36, we present more information on those who have no record of CFTR modulator use, including new information this year on their lung and nutritional health.

The Registry's comprehensive data collection enables us to monitor trends and changes in the health of people with CF closely. This report emphasises the importance of continued data collection through the Registry to monitor and understand the long-term effects of new treatments, provide insights to support those not eligible for modulators, and generate the evidence needed to adapt care and research strategies as the CF population evolves.

As we look ahead, this report serves not only as a snapshot of the current health of people with CF but also as a foundation for future innovation and hope.

Finally, I extend my deepest gratitude to everyone who supports the Registry – from those living with CF and their families who generously consent to sharing their data, to the NHS care teams and administrators who collect and enter the data. Your participation is essential to shaping a life unlimited by cystic fibrosis.

David Ramsden
Chief Executive of Cystic Fibrosis Trust

Executive summary

The 2024 Registry data continues to be a hugely valuable asset in driving forward improvements in CF care which the UK CF community should be proud of. I would like to highlight some aspects of this year's report.

- 11,381 people with CF are registered within the UK CF Registry of whom 93% had an annual review this year (Section 1.1).
- 65% of the UK population living with CF are over 16 years of age (Section 1.1) and 17.2% are over 40 years of age (Section 1.5). The changing age distribution of the UK CF population over the past 10 years is illustrated in Section 1.6.
- 68.2% of people with CF over 16 years of age are in work or studying – a figure that continues to increase annually (Section 1.12).
- Half of people born today with CF in the UK are predicted to live at least 66.2 years (Section 1.44), which has increased again from previous years.
- 5.4% of the UK CF population report being non-white or of mixed ethnicity (Section 1.7).
- The median Best FEV_1% continues to rise and is now 89.4% (Section 1.14).
- 17.2% 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 >= 25. The use of oral supplements has fallen again to 14.9% of the UK CF Population (Section 1.42).
- 27.8% of people with CF over 10 years of age are on CF diabetes therapies (Section 1.23).
- Depression is reported in 7.3% of people with CF over 16 years of age (Section 1.21).
- 111 women with CF had babies in 2024 and 35 men with CF became fathers (Section 1.13).
- The number of people with CF who had at least one respiratory culture remained high (93.4%) (Section 1.19), but sputum samples made up a smaller proportion of the sample type (58.8%).
- The percentage of people receiving at least one course of IV antibiotics (20%) has dropped again this year (Section 1.24). There has been a large drop in the number of ports inserted or replaced (0.8%) (Section 1.21).
- The proportion of people with CF that remain on the combination of inhaled antibiotics, DNase and hypertonic saline or mannitol has dropped from 14.9% last year to 12.3% this year (Section 1.33). 27.8% of people with CF are on none of these inhaled therapies as compared to 24.6% last year.
- 8,830 people with CF with a 2024 annual review have at least one record of using a CFTR modulator in 2024 (Section 1.35a). Tables in Sections 1.35b and 1.36 illustrate modulator use by genotype group and demographics of those eligible and ineligible for current modulator therapies respectively. 12.4% of people with CF with a 2024 annual review have never used a CFTR modulator (Section 1.35a).

Sections 2 and 3 are the centre-level reports, which centres may find helpful when analysing their pattern of home compared to hospital IV antibiotics use 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 would like to express our gratitude to those living with CF for consenting to have their clinical data recorded and the clinical teams for collecting and entering it into the Registry. This collective effort has ensured that the CF community continues to benefit from comprehensive, accurate and timely data to continue to further knowledge of CF and to drive ongoing improvements in care of people living with CF.

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 96.

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

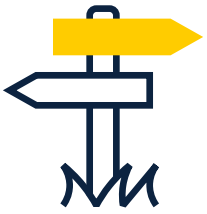
Cystic fibrosis

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. Many people with CF have difficulty digesting food. People with CF may also develop other problems, such as liver disease or CF diabetes (CFD).

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 2: 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

*	2019	2020	2021	2022	2023	2024
CF patients registered; n[1]	10655	10837	10908	11148	11318	11381
Excluding diagnoses that year; n[2]	10449	10632	10715	10928	11144	11217
CF patients with an annual review; n(%)[3]	10070 (96)	9922 (93)**	10175 (95)	10251 (94)	10344 (93)	10424 (93)
New diagnoses						
All newly diagnosed patients; n[2]	206	205	193	220***	174	164
Patients identified by NBS; n[2]	149	154	136	159	125	123
Patients diagnosed age 16 years or older; n[4]	29	14	21	33	25	17
Demographics						
Age in years; median (IQR)[5]	20 (9, 32)	21 (10, 32)	21 (10, 33)	22 (11, 33)	22 (11, 34)	23 (12, 35)
Adults aged 16 years and over; %[6]	60.4	61.0	62.1	62.9	64.2	65.2
Males; %[6]	53.2	53.2	53.2	53.4	53.2	53.2
Genotyped; %[6]	98.9	98.9	99.0	99.3	99.3	99.3
Total deaths during annual review year; n(%)[7]	118 (1.1)	100 (0.9)	67 (0.6)	68 (0.6)	51 (0.5)	51 (0.4)
Age at death in years; median (IQR)[7]	36 (28, 47)	37 (29, 48)	38 (31, 49)	38 (31, 51)	42 (33, 54)	42 (31, 51)

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:

* Unless otherwise stated in a footnote, all proportions are calculated from all CF patients registered (row 1).

** Corrected from 2020 report.

*** Amended from previously published reports and is a lower figure because individual demographic records were updated and the date of diagnosis was amended to an earlier year.

1 Number of patients alive on 1 January in the given year diagnosed with CF, with at least 1 annual review in either the given year or the previous 2 annual review years or at least 1 recorded encounter in the given year, plus any newly diagnosed and/or newly consented individuals. We use this definition as a proxy for the sub-population of people with CF who are actively engaged with NHS services.

2 This figure may be amended from previous publications if the diagnosis date of an individual registered in the given year has since been updated. This happens in cases where it was not recorded at the time of publishing or was since corrected. These figures only include new diagnoses among registered patients in the given year. The subsequent diagnosis section details all new diagnoses by calendar year, irrespective of when the individual was registered on the UK CF registry.

3 Newly diagnosed patients in the 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.

4 This figure is the total number of individuals newly diagnosed in the given year who were 16 years or older at the time of diagnosis.

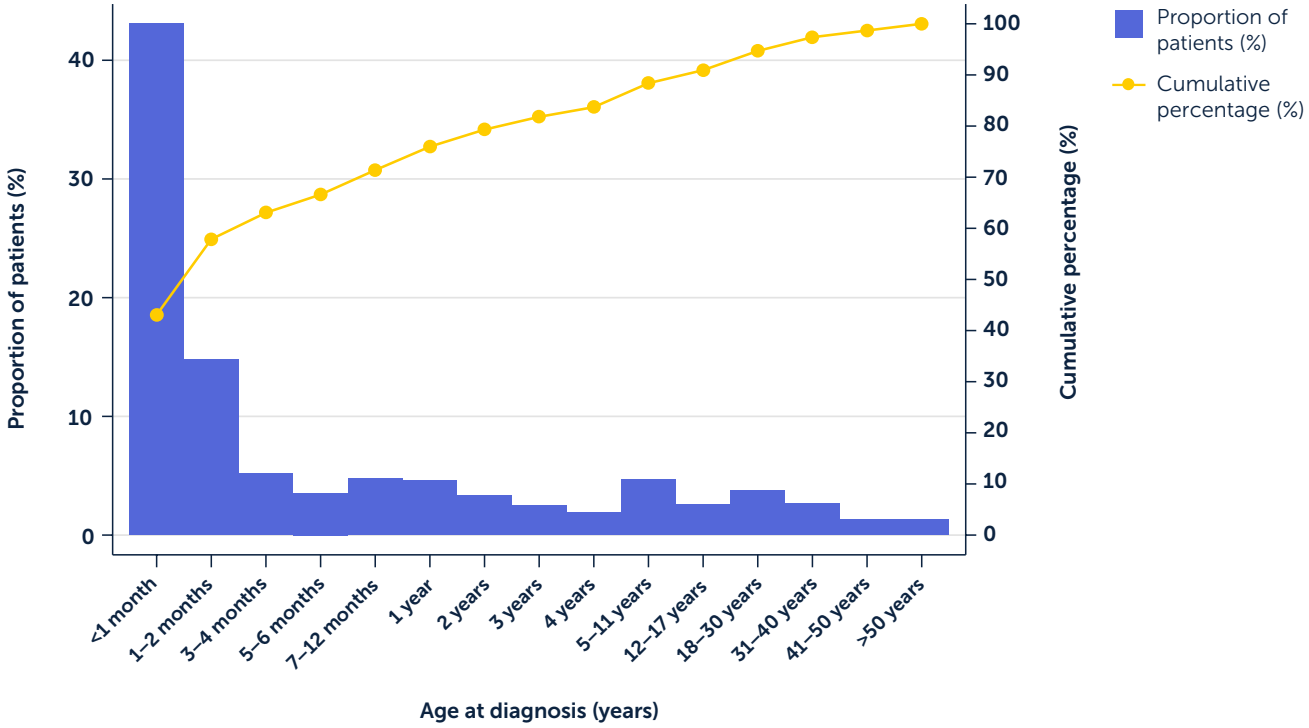
5 Age is taken at the given year's annual review; if the individual did not have an annual review during the year in question, their age was taken on 1 January of the given year.

6 Calculated from all registered patients in the given year. Figures may be different from previous reports as they were previously calculated from patients with an annual review in the given year.

7 This figure may be amended from previous publications if the date of death for an individual in the given year was unknown at the time of analysis and subsequently entered on to the Registry following the given year's annual report publication.

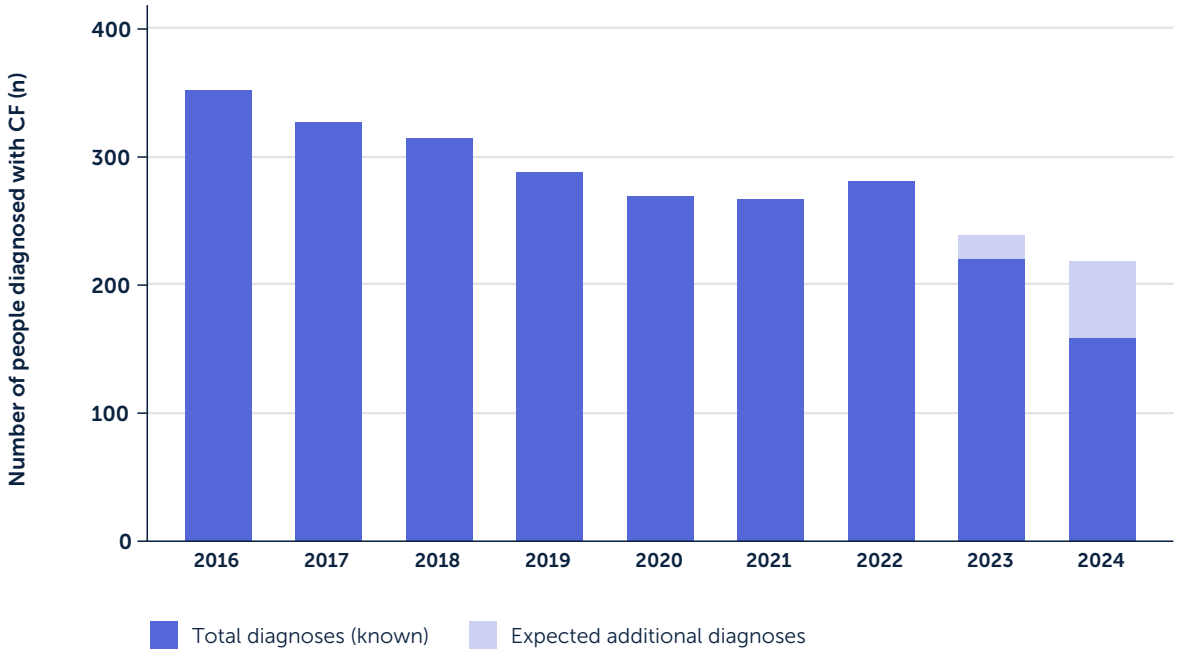
Diagnosis of cystic fibrosis

1.2 Age at diagnosis
N=11381



1.3 New diagnoses of cystic fibrosis by year, 2016–2024 N=2479

This chart shows all new diagnoses of cystic fibrosis by year in the UK for all people registered between 2016–2024 (see table 1.1 for definition of patients registered). Due to timelines in diagnosing CF, obtaining consent to join the Registry, and other factors, individuals are not always registered in the same year in which they were diagnosed; there is typically up to a 2-year lag in registering people with a new diagnosis. For 2023 and 2024 we have also included an estimate[1] of the number of additional diagnoses we expect to see when we have collected and analysed Registry data in future years.



1 Expected additional diagnoses were estimated by averaging the ratio of "lagged" registrations (patients added to the UK CF Registry 1 year after diagnosis and 2+ years after diagnosis) to patients registered in the same year they were diagnosed. We used this ratio to estimate the expected additional diagnoses for 2023 and 2024 based on the number of new diagnoses added to the Registry in the year they were diagnosed.

1.4 Mode of presentation
N=1202

The following table shows the most frequent modes of presentation for people diagnosed between 2020–2024. Individuals included in this table were registered and diagnosed between 2020–2024. Patients not diagnosed through newborn screening (NBS) may present with multiple symptoms so the categories are not mutually exclusive, and percentages may not add up to 100.

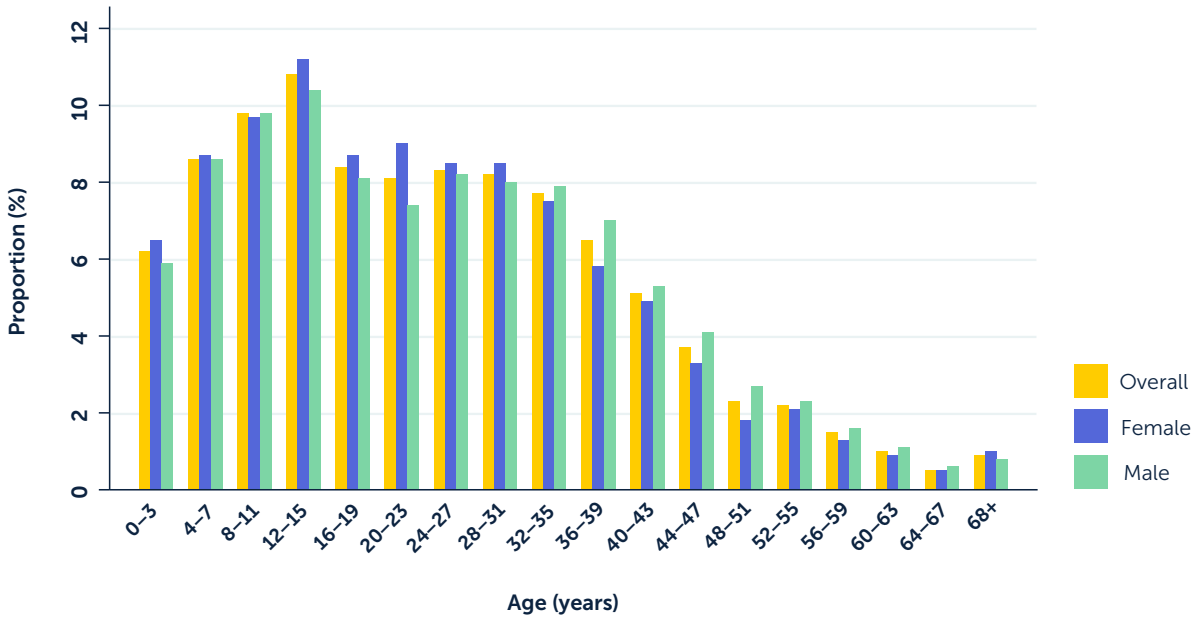
	All patients diagnosed 2020–2024	Age <1 year at diagnosis	Age ≥1 year & < 16 years at diagnosis	Age ≥16 years at diagnosis
Total patients	**1202**	**928**	**81**	**193**
Number diagnosed by newborn screening (NBS)[1]	839	828	11	0
Total non-NBS	363	100	70	193
Individuals not diagnosed by NBS	**363 (100.0)**	**100 (100.0)**	**70 (100.0)**	**193 (100.0)**
Genotype	95 (26.2)	0 (0.0)	19 (27.1)	76 (39.4)
Persistant or acute respiratory infection	84 (23.1)	9 (9.0)	30 (42.9)	45 (23.3)
Bronchiectasis	64 (17.6)	0 (0.0)	7 (10.0)	57 (29.5)
Family history	59 (16.3)	21 (21.0)	17 (24.3)	21 (10.9)
Meconium ileus	49 (13.5)	49 (49.0)	0 (0.0)	0 (0.0)
Unknown	26 (7.2)	5 (5.0)	0 (0.0)	21 (10.9)
Fertility	26 (7.2)	0 (0.0)	0 (0.0)	26 (13.5)
Prenatal	24 (6.6)	22 (22.0)	0 (0.0)	<5
Failure to thrive/malnutrition	20 (5.5)	11 (11.0)	8 (11.4)	<5
Pancreatitis	18 (5.0)	0 (0.0)	5 (7.1)	13 (6.7)

1 If the method of diagnosis was genotyping and the age of diagnosis was <1 year, this was counted as a diagnosis by newborn screening (n=34).

Demographics

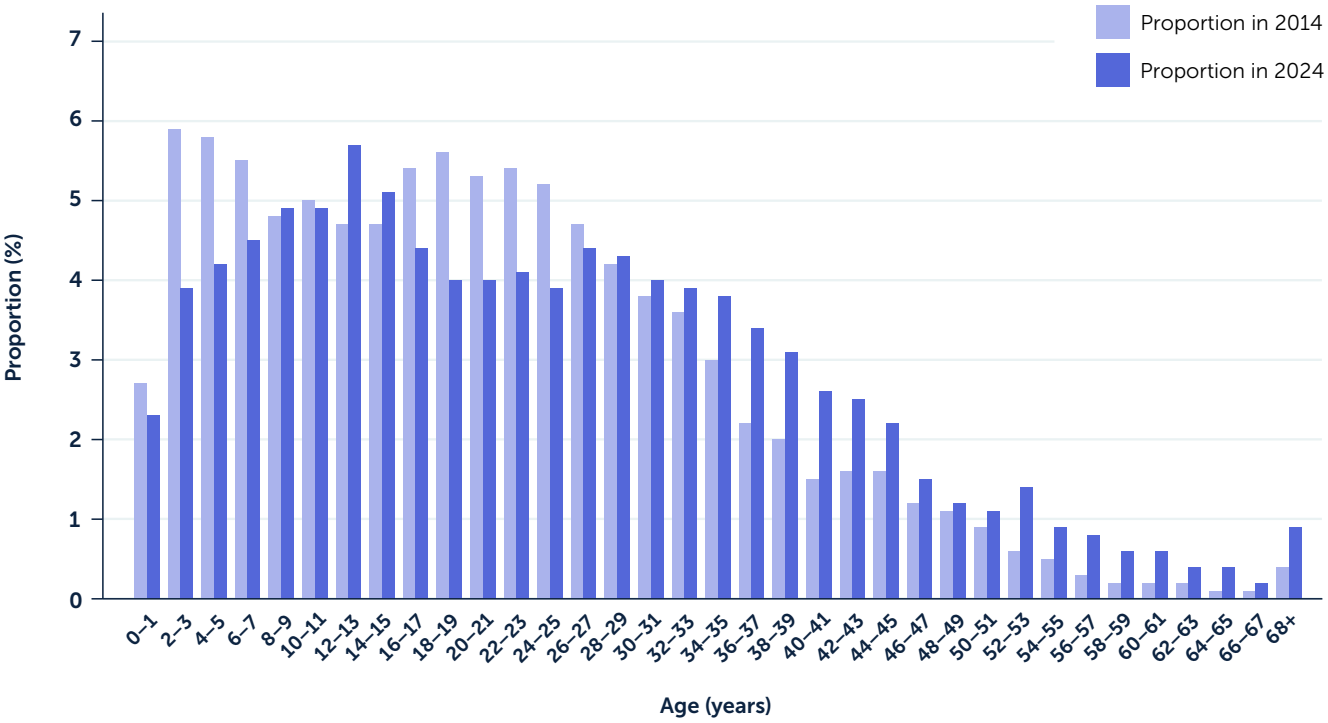
1.5 Age distribution by sex
N=10424

This graph shows the age distribution by sex for people with an annual review in 2024.



Age	All; n (%)	Females; n (%)	Males; n (%)
0–3	645 (6.2)	318 (6.5)	327 (5.9)
4–7	899 (8.6)	424 (8.7)	475 (8.6)
8–11	1020 (9.8)	473 (9.7)	547 (9.8)
12–15	1124 (10.8)	544 (11.2)	580 (10.4)
16–19	874 (8.4)	422 (8.7)	452 (8.1)
20–23	845 (8.1)	436 (9.0)	409 (7.4)
24–27	870 (8.3)	414 (8.5)	456 (8.2)
28–31	859 (8.2)	416 (8.5)	443 (8.0)
32–35	806 (7.7)	365 (7.5)	441 (7.9)
36–39	673 (6.5)	282 (5.8)	391 (7.0)
40–43	533 (5.1)	237 (4.9)	296 (5.3)
44–47	387 (3.7)	160 (3.3)	227 (4.1)
48–51	242 (2.3)	90 (1.8)	152 (2.7)
52–55	234 (2.2)	104 (2.1)	130 (2.3)
56–59	152 (1.5)	64 (1.3)	88 (1.6)
60–63	107 (1.0)	46 (0.9)	61 (1.1)
64–67	57 (0.5)	25 (0.5)	32 (0.6)
68+	97 (0.9)	50 (1.0)	47 (0.8)
<16	3688 (35.4)	1759 (36.1)	1929 (34.7)
≥16	6736 (64.6)	3111 (63.9)	3625 (65.3)
<18	4143 (39.7)	1985 (40.8)	2158 (38.9)
≥18	6281 (60.3)	2885 (59.2)	3396 (61.1)
Overall	**10424**	**4870**	**5554**

1.6 Age distribution of the UK CF population in 2014 and 2024
N=10424 in 2024, N=9432 in 2014



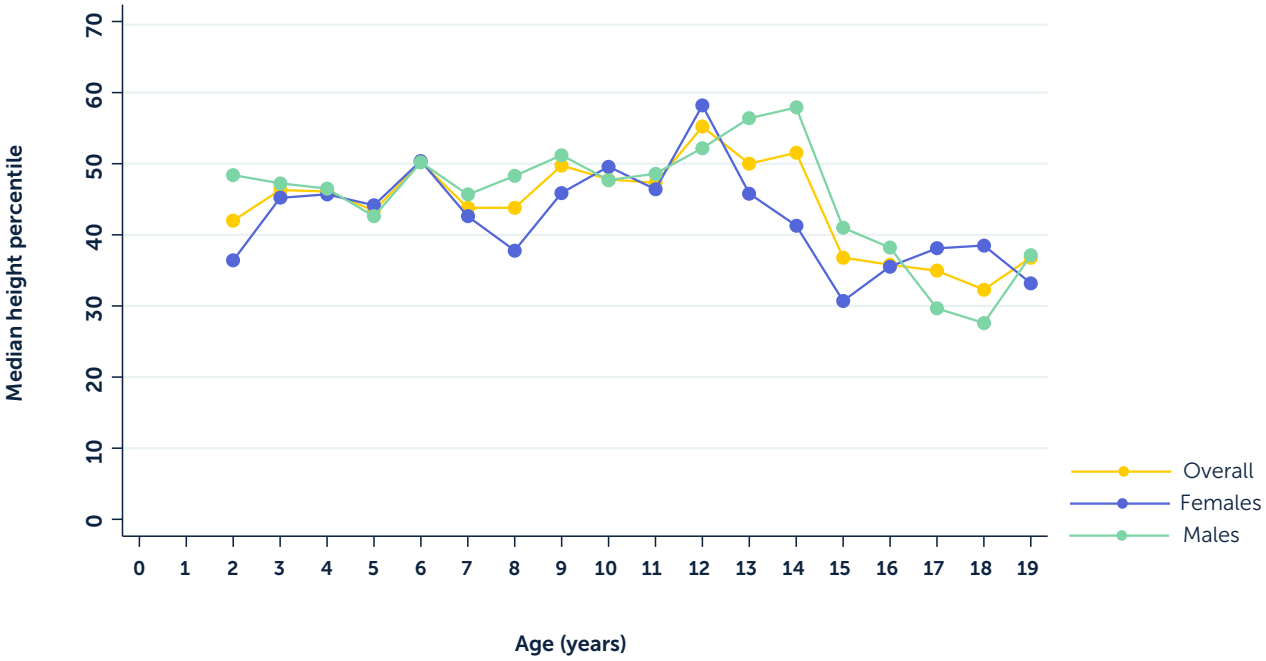
1.7 Ethnicity

Ethnicity n (%)	2014	2019	2024
Total	9432	10070	10424
Total known[1]	9336	9833	10227
White	**8939 (95.7)**	**9396 (95.6)**	**9673 (94.6)**
Asian	**240 (2.6)**	**286 (2.9)**	**344 (3.4)**
Bangladeshi	32 (0.3)	38 (0.4)	46 (0.4)
Indian	33 (0.4)	45 (0.5)	59 (0.6)
Pakistani	150 (1.6)	173 (1.8)	194 (1.9)
Other (Asian)	25 (0.3)	30 (0.3)	45 (0.4)
Black	**28 (0.3)**	**28 (0.3)**	**31 (0.3)**
Black African	–*	–*	–*
Black Caribbean	14 (0.1)	12 (0.1)	14 (0.1)
Other (Black)	<5	<5	<5
Mixed[2]	**81 (0.9)**	**65 (0.7)**	**119 (1.2)**
Mixed (white-Asian)	–	15 (0.2)	30 (0.3)
Mixed (white-Black African)	–	9 (0.1)	15 (0.1)
Mixed (white-Black Caribbean)	–	19 (0.2)	33 (0.3)
Other (mixed)	-	22 (0.2)	41 (0.4)
Other	**48 (0.5)**	**58 (0.6)**	**60 (0.6)**

* Redacted to adhere to statistical disclosure guidelines
1 Proportions are calculated from total known ethnicities
2 Further detail on mixed ethnicity categories were collected from 2016 onwards

1.8 Height percentiles of children and young people (age 2–19)[1] N=4562

The following chart and table show the height percentiles of people with CF, ages 2 to 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.



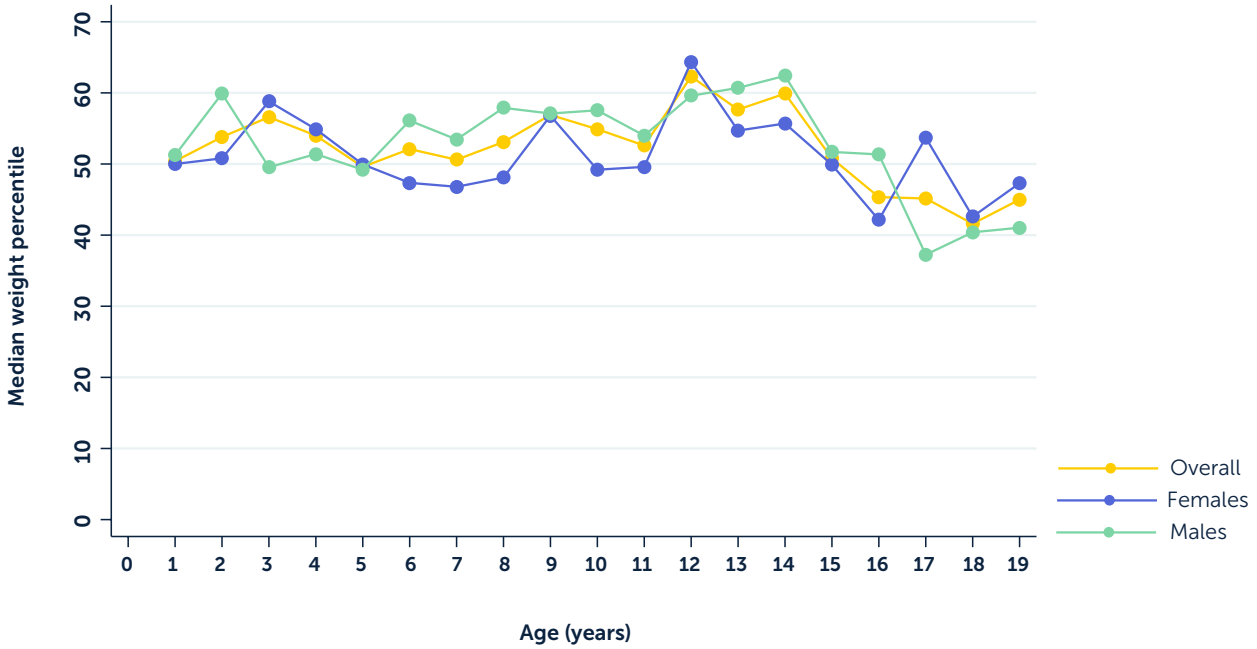
	Overall			Female			Male		
Age	n	Median	IQR	n	Median	IQR	n	Median	IQR
2	182	42.0	20.3–68.0	89	36.4	14.1–62.4	93	48.4	23.3–74.1
3	207	46.3	20.4–68.7	109	45.2	23.5–69.0	98	47.2	18.2–68.2
4	196	46.1	19.0–72.1	97	45.7	19.0–67.2	99	46.5	19.8–76.1
5	235	43.4	18.3–71.6	110	44.2	15.9–71.6	125	42.6	20.8–69.7
6	210	50.2	21.5–70.9	88	50.4	16.7–70.9	122	50.2	24.7–70.0
7	247	43.8	21.2–77.1	123	42.6	19.5–71.9	124	45.7	21.9–86.4
8	245	43.8	22.1–73.4	114	37.8	21.4–71.8	131	48.3	25.8–74.3
9	260	49.8	24.2–73.9	122	45.9	24.6–75.5	138	51.2	23.9–73.7
10	238	47.8	23.3–75.4	111	49.6	20.4–75.6	127	47.7	27.5–75.4
11	266	47.3	26.1–72.2	123	46.4	22.5–79.5	143	48.6	27.5–69.5
12	285	55.2	30.2–80.6	142	58.2	36.2–80.6	143	52.2	27.2–81.4
13	297	50.0	26.7–77.6	145	45.8	25.9–72.0	152	56.4	31.3–81.6
14	268	51.5	24.7–73.8	135	41.3	20.4–70.2	133	57.9	32.1–79.2
15	256	36.8	16.7–59.0	115	30.7	13.3–58.6	141	41.0	20.4–59.4
16	231	35.8	16.4–59.0	118	35.5	15.1–55.9	113	38.2	16.8–59.9
17	220	34.9	11.1–56.8	106	38.1	13.3–59.7	114	29.6	10.0–53.7
18	208	32.3	10.0–59.7	100	38.5	13.6–71.2	108	27.6	9.2–56.1
19	210	36.8	13.6–57.5	95	33.2	13.6–52.4	115	37.1	11.7–59.5
Overall	**4261***	**44.1**	**20.6–70.9**	**2042**	**42.3**	**19.3–70.6**	**2219**	**46.5**	**21.6–71.3**

* Number with non-missing data.

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

1.9 Weight percentiles of children and young people (<20 years)[1] N=4511

The following chart and table show the weight of people with CF, aged 19 and under, 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.



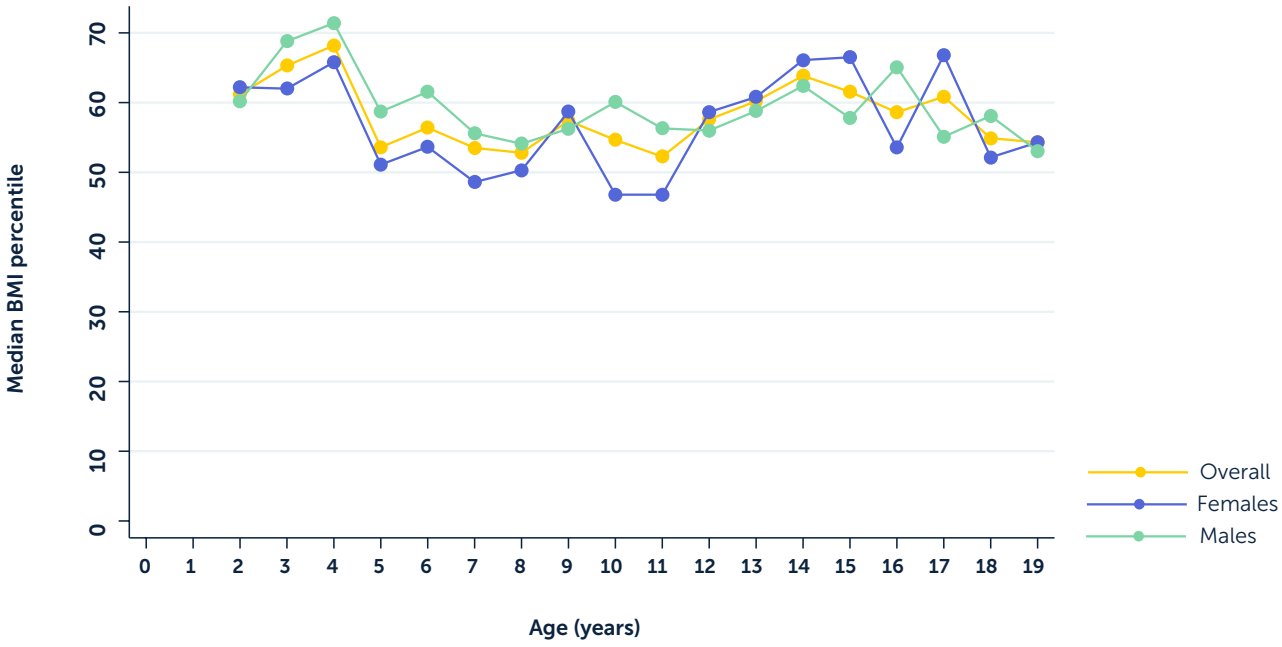
	Overall			Female			Male		
Age	n	Median	IQR	n	Median	IQR	n	Median	IQR
1	181	50.4	23.3–71.7	87	50.0	22.7–72.9	94	51.2	24.3–68.5
2	189	53.8	30.6–77.9	93	50.8	22.1–70.5	96	59.9	37.1–82.3
3	209	56.6	30.3–80.7	111	58.8	30.3–79.4	98	49.5	28.0–82.2
4	197	54.0	31.1–81.5	97	54.9	26.4–79.6	100	51.4	32.9–86.0
5	235	49.6	26.1–76.3	110	50.0	25.3–72.8	125	49.2	27.7–80.5
6	210	52.1	28.1–77.8	88	47.3	23.8–74.4	122	56.1	32.9–82.0
7	247	50.6	26.0–80.1	123	46.8	24.5–70.5	124	53.4	27.1–86.3
8	245	53.1	29.5–79.1	114	48.1	26.4–78.8	131	57.9	32.6–80.0
9	261	56.9	30.4–83.2	122	56.8	29.5–80.3	139	57.1	30.4–86.4
10	239	54.9	30.9–81.4	111	49.2	27.6–73.9	128	57.6	35.0–82.2
11	266	52.6	30.9–79.0	123	49.6	28.2–82.7	143	54.0	34.5–77.8
12	285	62.3	34.4–85.0	142	64.4	39.6–85.0	143	59.6	28.0–86.7
13	298	57.7	31.6–83.7	145	54.7	31.4–81.3	153	60.7	33.4–85.4
14	269	59.9	34.5–85.3	135	55.7	32.3–85.2	134	62.4	37.6–85.4
15	256	50.8	28.0–73.8	115	49.9	25.3–75.3	141	51.7	29.4–73.3
16	232	45.3	21.0–75.1	118	42.2	16.9–71.3	114	51.3	25.5–78.7
17	220	45.2	17.3–79.0	106	53.7	21.5–85.4	114	37.2	14.1–68.4
18	207	41.6	13.3–73.3	100	42.6	15.2–73.0	107	40.4	11.3–75.0
19	205	45.0	16.9–71.3	93	47.3	20.8–73.8	112	41.0	13.5–67.5
Overall	**4451***	**52.6**	**26.9–79.6**	**2133**	**51.6**	**25.9–78.5**	**2318**	**54.2**	**28.0–80.5**

* Number with non-missing data.

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

1.10a Body Mass Index (BMI) percentiles in children and young people (<20 years)[1]
N=4324

The following chart and table show the BMI percentiles of people with CF, ages 2 to 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.



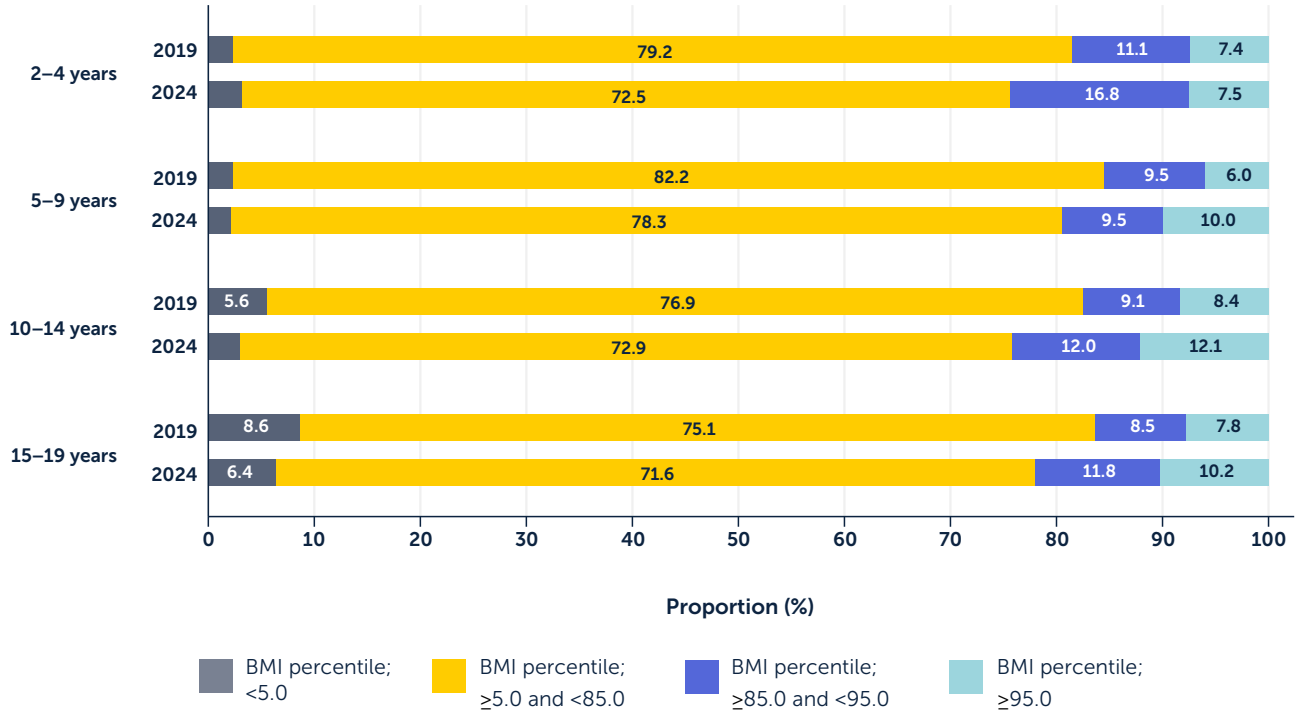
	Overall			Female			Male		
Age	n	Median	IQR	n	Median	IQR	n	Median	IQR
2	182	61.2	37.9–78.7	89	62.2	36.7–76.5	93	60.2	38.0–83.5
3	207	65.3	41.4–85.9	109	62.0	35.4–85.9	98	68.8	47.9–85.8
4	196	68.2	44.3–84.0	97	65.8	43.5–83.2	99	71.4	44.4–84.3
5	235	53.6	35.0–78.1	110	51.1	33.7–74.4	125	58.7	38.5–80.4
6	210	56.4	34.8–79.1	88	53.7	34.5–78.1	122	61.5	37.1–81.2
7	247	53.5	31.2–76.5	123	48.6	31.3–73.7	124	55.6	31.0–78.8
8	245	52.8	34.9–81.2	114	50.3	31.9–81.7	131	54.1	35.4–81.2
9	260	57.4	31.3–86.2	122	58.7	29.5–83.9	138	56.2	32.4–90.9
10	238	54.7	29.9–81.2	111	46.8	26.2–77.0	127	60.1	33.6–85.1
11	266	52.2	32.0–78.7	123	46.8	28.5–77.3	143	56.3	34.6–82.9
12	285	57.6	32.0–83.1	142	58.7	37.1–83.1	143	56.0	25.4–84.6
13	297	60.2	35.5–83.2	145	60.8	36.8–82.8	152	58.8	33.5–83.4
14	268	63.8	39.5–89.1	135	66.1	41.9–90.5	133	62.4	31.5–87.4
15	256	61.5	34.8–82.8	115	66.5	39.8–82.6	141	57.8	30.7–83.0
16	231	58.6	28.9–83.3	118	53.6	27.4–83.2	113	65.1	31.6–83.4
17	218	60.8	29.8–85.3	105	66.8	30.7–89.3	113	55.1	29.7–76.0
18	207	54.9	27.1–83.0	100	52.2	25.2–79.0	107	58.1	29.4–84.1
19	205	54.3	24.2–79.1	93	54.3	25.9–81.0	112	53.0	23.2–75.7
Overall	**4253***	**58.2**	**32.6–82.6**	**2039**	**57.6**	**32.4–82.0**	**2214**	**58.8**	**32.8–83.2**

* Number with non-missing data.

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

1.10b Body Mass Index (BMI) percentiles in children and young people (<20 years)[1] for 2019 and 2024

The following graph shows the change in BMI groups for children and young people with CF from 2019 and 2024.

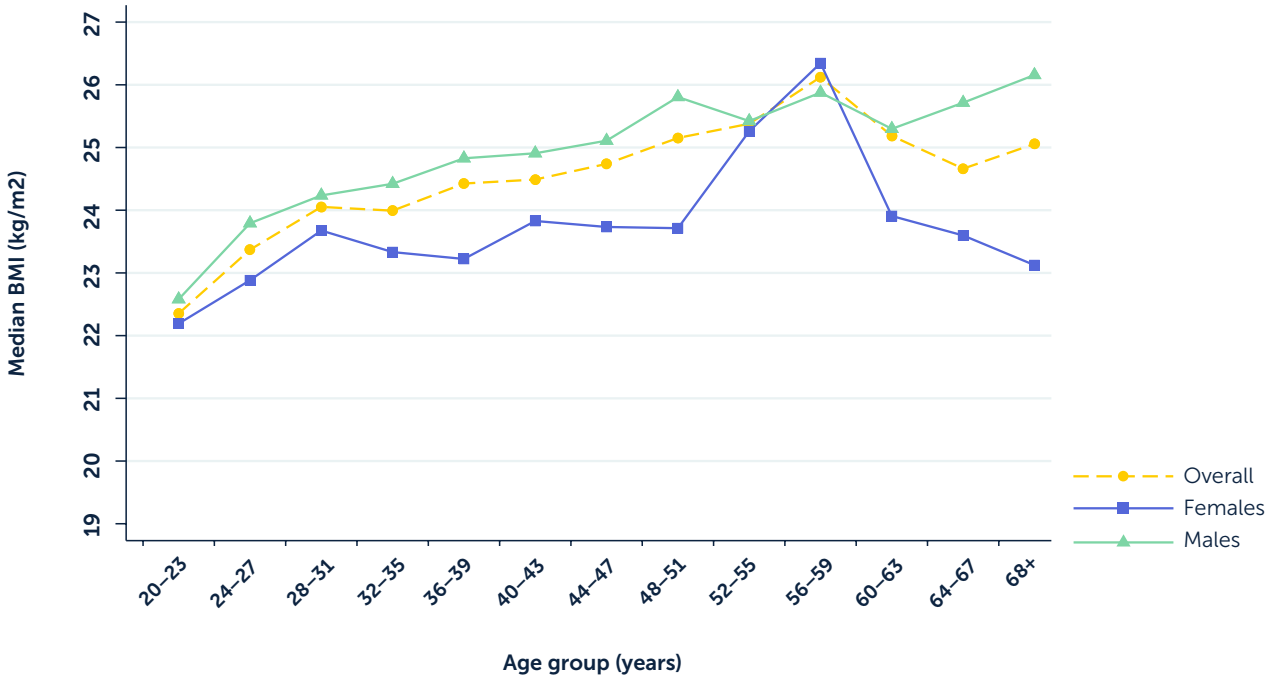


			BMI category by age and year : n(%)			
Age group	Year	Total number of people in each age group*	BMI percentile; <5.0	BMI percentile; ≥5.0 and <85.0	BMI percentile; ≥85.0 and <95.0	BMI percentile; ≥95.0
2–4 years	2019	746	17 (2.3)	591 (79.2)	83 (11.1)	55 (7.4)
	2024	585	19 (3.2)	424 (72.5)	98 (16.8)	44 (7.5)
5–9 years	2019	1392	32 (2.3)	1145 (82.3)	132 (9.5)	83 (6.0)
	2024	1197	26 (2.2)	937 (78.3)	114 (9.5)	120 (10.0)
10–14 years	2019	1257	70 (5.6)	968 (77.0)	114 (9.1)	105 (8.4)
	2024	1354	41 (3.0)	986 (72.8)	163 (12.0)	164 (12.1)
15–19 years	2019	1053	91 (8.6)	790 (75.0)	90 (8.5)	82 (7.8)
	2024	1117	71 (6.4)	800 (71.6)	132 (11.8)	114 (10.2)

* With non-missing BMI data

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

1.11a Body Mass Index (BMI) in adults ages 20 and older
N=5862

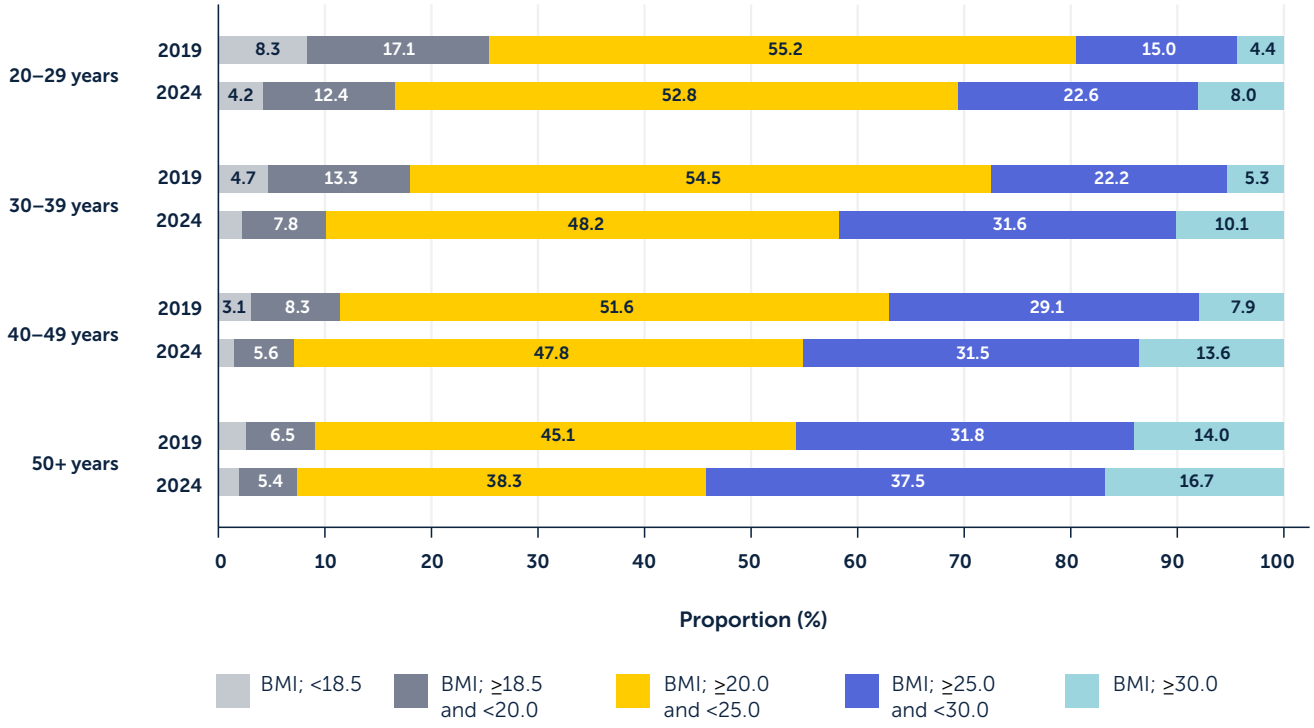


	Overall			Female			Male		
Age	n	Median	IQR	n	Median	IQR	n	Median	IQR
20–23	825	22.4	20.4–24.7	421	22.2	20.3–24.7	404	22.6	20.6–24.7
24–27	846	23.4	21.2–26.2	404	22.9	21.1–26.3	442	23.8	21.3–26.2
28–31	829	24.1	21.5–26.4	405	23.7	21.2–26.2	424	24.2	21.9–26.6
32–35	782	24.0	22.0–26.9	355	23.3	21.6–26.2	427	24.4	22.4–27.2
36–39	649	24.4	21.9–27.4	270	23.2	21.1–26.7	379	24.8	22.8–27.6
40–43	520	24.5	22.2–27.1	229	23.8	21.7–26.5	291	24.9	22.8–27.6
44–47	373	24.7	22.2–27.8	153	23.7	21.3–28.3	220	25.1	23.2–27.7
48–51	237	25.2	22.7–27.7	89	23.7	22.0–26.8	148	25.8	23.8–28.1
52–55	226	25.4	22.9–28.4	100	25.3	21.9–29.2	126	25.4	23.3–27.9
56–59	148	26.1	22.4–28.7	62	26.3	22.1–29.5	86	25.9	23.1–28.6
60–63	104	25.2	22.9–29.6	45	23.9	22.7–29.8	59	25.3	23.9–29.3
64–67	56	24.7	23.1–27.1	24	23.6	21.7–25.5	32	25.7	23.7–27.3
68+	94	25.1	21.7–28.9	48	23.1	20.3–28.6	46	26.2	23.9–28.9
Overall	**5689***	**24.0**	**21.6–26.9**	**2605**	**23.3**	**21.1–26.5**	**3084**	**24.4**	**22.1–27.0**

* Number with non-missing data.

1.11b Body Mass Index (BMI) in adults for 2019 and 2024

The following graph shows the change in the proportion of people in each BMI group for 2019 and 2024.



			BMI category by age and year : n(%)				
Age group	Year	Total number of people in each age group*	BMI; <18.5	BMI; ≥18.5 and <20.0	BMI; ≥20.0 and <25.0	BMI; ≥25.0 and <30.0	BMI; ≥30
20–29 years	2019	2345	195 (8.3)	401 (17.1)	1293 (55.1)	352 (15.0)	104 (4.4)
	2024	2104	88 (4.2)	260 (12.4)	1112 (52.9)	475 (22.6)	169 (8.0)
30–39 years	2019	1650	78 (4.7)	219 (13.3)	900 (54.5)	366 (22.2)	87 (5.3)
	2024	1826	42 (2.3)	142 (7.8)	881 (48.2)	577 (31.6)	184 (10.1)
40–49 years	2019	771	24 (3.1)	64 (8.3)	398 (51.6)	224 (29.1)	61 (7.9)
	2024	1020	15 (1.5)	57 (5.6)	488 (47.8)	321 (31.5)	139 (13.6)
50+ years	2019	506	13 (2.6)	33 (6.5)	228 (45.1)	161 (31.8)	71 (14.0)
	2024	738	15 (2.0)	40 (5.4)	283 (38.3)	277 (37.5)	123 (16.7)

* With non–missing BMI data

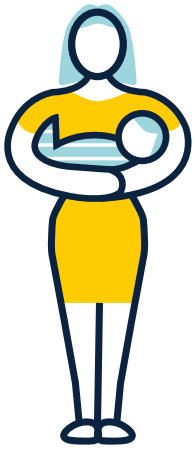
1.12 Education and employment in adults (16 years and over) N=6736

The following table shows how people with CF aged 16 and older reported their education and employment status in 2024.

	2021	2022	2023	2024		
	Overall	Overall	Overall	Overall	Male	Female
Number of patients	6297	6445	6588	6736	3625	3111
Number who completed questionnaire; n(%)[1]	6296 (100.0)	6442 (100.0)	6587 (100.0)	6726 (99.9)	3617 (99.8)	3109 (99.9)
Full-time employment; n(%)	2097 (33.3)	2228 (34.6)	2379 (36.1)	2538 (37.7)	1673 (46.2)	865 (27.8)
Part-time employment; n(%)	915 (14.5)	981 (15.2)	1031 (15.6)	997 (14.8)	355 (9.8)	642 (20.6)
Student; n(%)	1061 (16.8)	1046 (16.2)	1049 (15.9)	1053 (15.6)	501 (13.8)	552 (17.7)
Homemaker; n(%)	251 (4.0)	249 (3.9)	257 (3.9)	255 (3.8)	27 (0.7)	228 (7.3)
Unemployed; n(%)	791 (12.6)	767 (11.9)	744 (11.3)	775 (11.5)	459 (12.7)	316 (10.2)
Disabled; n(%)	255 (4.0)	228 (3.5)	237 (3.6)	235 (3.5)	121 (3.3)	114 (3.7)
Retired; n(%)	162 (2.6)	170 (2.6)	184 (2.8)	174 (2.6)	99 (2.7)	75 (2.4)
Volunteer; n(%)	12 (0.2)	14 (0.2)	21 (0.3)	16 (0.2)	8 (0.2)	8 (0.3)
Unknown entered; n(%)	752 (11.9)	759 (11.8)	685 (10.4)	683 (10.1)	374 (10.3)	309 (9.9)
No. in work or study; n(%)	**4073 (64.7)**	**4255 (66.1)**	**4459 (67.7)**	**4588 (68.2)**	**2529 (69.9)**	**2059 (66.2)**

1.13 Parenthood

	2021	2022	2023	2024
Women with CF who had babies; n	103	140	116	111
Men with CF who became fathers; n	30	33	31	35



111 women with CF had babies in 2024

35 men with CF became fathers in 2024

1 Proportions below are calculated from the total who completed the questionnaire

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

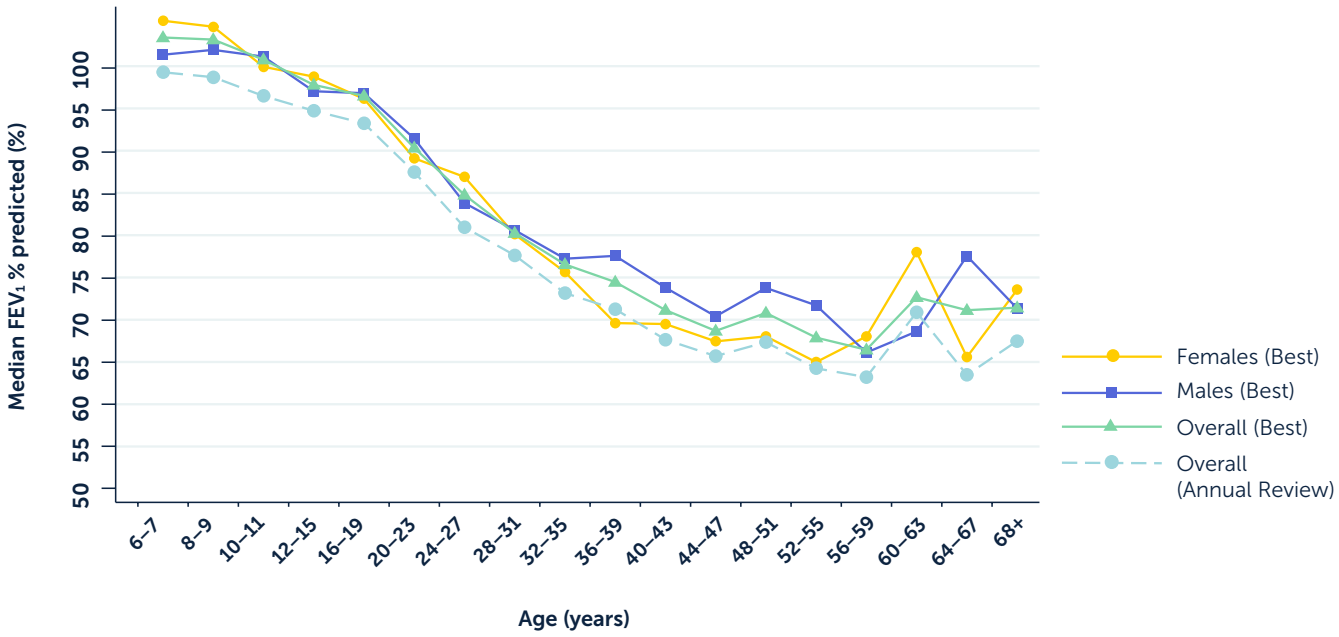
1.14 Annual review FEV_1% predicted (GLI equations) in patients aged 6 years and older who have not had a lung transplant N=9084

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	424	99.5	89.4–109.0	198	100.4	92.7–109.0	226	98.7	88.2–109.0
8–9	484	98.9	90.3–106.4	228	100.7	92.2–108.2	256	96.8	89.1–104.5
10–11	484	96.7	89.4–104.6	224	95.8	87.6–104.5	260	97.9	90.6–105.2
12–15	1066	94.9	84.8–103.2	516	96.1	86.6–103.7	550	93.7	83.6–102.1
16–19	833	93.4	83.3–102.2	406	93.4	82.2–102.1	427	93.8	84.3–102.4
20–23	785	87.6	72.9–98.8	409	87.4	71.5–97.4	376	88.3	73.8–99.9
24–27	787	81.1	65.3–95.5	374	82.7	64.7–97.3	413	80.0	65.4–94.2
28–31	774	77.7	58.2–91.6	380	76.9	56.4–91.5	394	78.3	59.6–92.8
32–35	714	73.2	56.5–88.5	317	72.7	57.3–86.3	397	74.3	55.5–89.3
36–39	594	71.3	52.2–87.7	250	67.3	50.8–84.6	344	75.2	54.2–89.5
40–43	441	67.7	47.1–85.3	198	67.1	44.4–83.0	243	70.4	49.9–87.3
44–47	341	65.7	51.5–84.5	135	62.7	50.5–81.3	206	67.4	52.5–85.7
48–51	197	67.4	48.3–81.8	72	61.8	46.3–79.7	125	70.0	53.9–85.4
52–55	190	64.3	49.2–85.1	85	63.1	51.5–83.5	105	68.1	46.2–85.1
56–59	134	63.2	47.3–80.8	59	63.2	48.0–81.7	75	63.3	46.6–80.8
60–63	93	70.9	51.8–82.6	42	74.6	60.9–83.0	51	64.7	44.7–82.3
64–67	51	63.5	40.1–86.2	24	58.6	42.1–77.5	27	76.6	37.3–89.9
68+	91	67.5	46.6–84.6	46	67.0	52.2–83.0	45	68.6	43.9–85.1
<16	2458	96.8	87.6–105.2	1166	97.4	88.5–106.0	1292	96.1	86.7–104.5
≥16	6025	79.4	59.0–93.9	2797	79.2	58.7–93.5	3228	79.7	59.6–94.2
<18	2891	96.4	87.3–104.8	1382	96.9	87.6–105.3	1509	96.0	86.7–104.3
≥18	5592	77.6	57.8–92.8	2581	77.2	57.4–92.4	3011	78.0	58.0–93.0
Overall	**8483***	**86.1**	**67.3–98.7**	**3963**	**86.3**	**66.5–98.9**	**4520**	**86.0**	**67.9–98.6**

* Number with non-missing data

1.15 Best* FEV_1% predicted (GLI equations) in patients aged 6 years and older who have not had a lung transplant N=9084



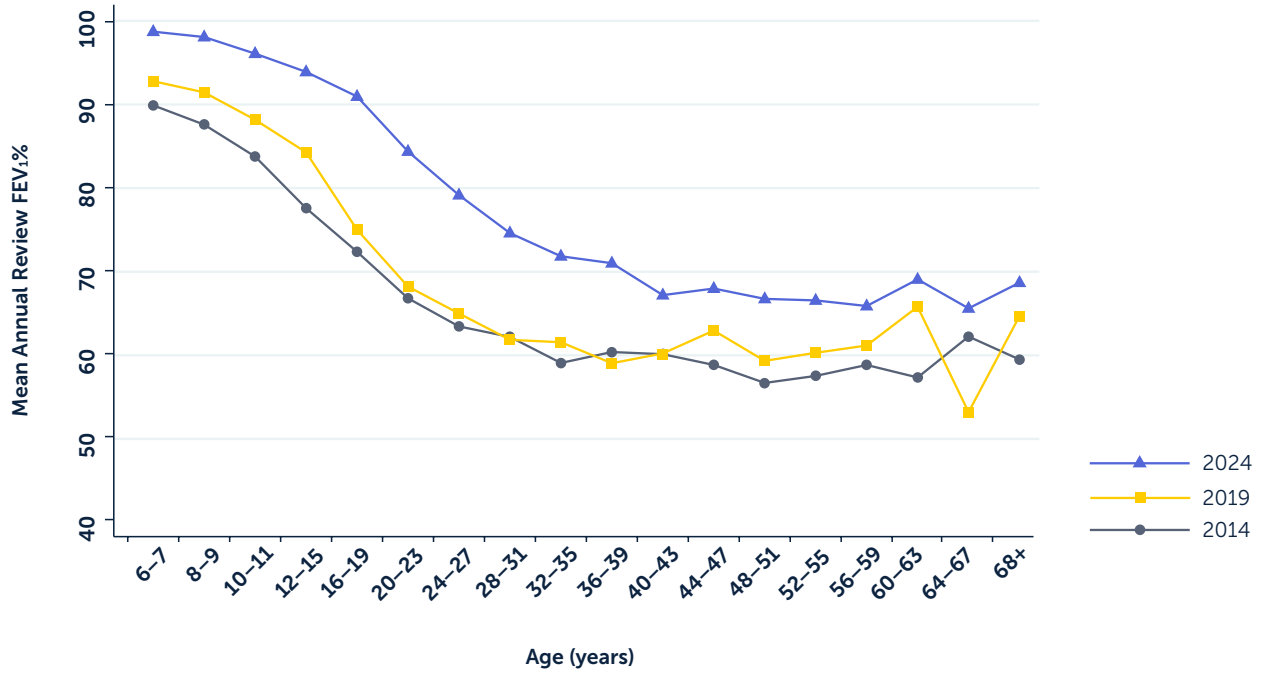
	Overall			Female			Male		
Age (years)	n	Median	IQR	n	Median	IQR	n	Median	IQR
6–7	442	103.6	94.4–112.9	205	105.6	97.2–113.8	237	101.6	93.4–112.3
8–9	493	103.4	95.9–110.7	231	104.9	97.3–111.6	262	102.2	94.5–109.6
10–11	498	101.0	92.8–108.5	229	100.2	92.2–108.3	269	101.4	93.7–109.2
12–15	1099	98.0	89.5–106.9	532	99.0	90.3–107.5	567	97.2	88.2–105.8
16–19	857	96.7	86.8–105.0	416	96.3	86.7–104.4	441	97.0	86.8–105.5
20–23	821	90.5	75.8–101.4	427	89.3	74.6–100.8	394	91.6	77.5–102.5
24–27	840	84.9	69.0–98.0	398	87.1	67.8–100.1	442	83.9	69.3–96.5
28–31	814	80.4	61.0–94.4	399	80.2	60.4–94.2	415	80.7	62.5–94.9
32–35	757	76.6	59.3–90.6	340	75.7	60.0–89.3	417	77.3	58.3–91.8
36–39	625	74.5	55.3–90.4	261	69.7	53.5–87.8	364	77.7	57.0–91.2
40–43	461	71.2	50.3–88.3	206	69.5	48.6–84.2	255	73.9	53.3–91.0
44–47	354	68.7	53.5–85.9	137	67.5	52.2–84.5	217	70.5	55.8–88.3
48–51	211	70.9	52.5–85.0	79	68.1	49.2–81.5	132	73.8	58.2–87.3
52–55	201	67.9	51.7–89.0	91	65.0	52.9–91.1	110	71.8	50.5–86.6
56–59	140	66.5	50.5–85.3	61	68.1	53.2–84.1	79	66.2	48.7–87.7
60–63	97	72.7	54.4–87.2	43	78.1	64.6–84.0	54	68.7	47.0–89.3
64–67	53	71.2	53.5–92.0	24	65.6	54.1–90.0	29	77.6	43.4–93.5
68+	96	71.5	53.2–89.4	49	73.6	60.2–89.1	47	71.4	49.0–90.4
<16	2532	100.7	92.2–109.2	1197	101.6	93.0–109.6	1335	100.1	91.8–108.5
≥16	6327	82.2	62.5–96.6	2931	81.8	61.5–96.1	3396	82.6	63.2–97.0
<18	2974	100.3	91.7–108.8	1417	100.8	92.3–109.2	1557	99.9	91.3–108.4
≥18	5885	80.7	60.9–95.1	2711	80.4	60.0–94.9	3174	81.0	61.6–95.3
Overall	**8859****	**89.4**	**70.6–101.8**	**4128**	**89.5**	**69.9–102.0**	**4731**	**89.3**	**71.2–101.6**

* 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.

1.16 Annual review FEV_1% predicted (GLI equations) over time in patients aged 6 years and older who have not had a lung transplant N=9084 in 2024, N=8419 in 2019, N=7626 in 2014

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 2024 compares to Registry data from 2014 and 2019.



	2014		2019		2024		
Age (years)	n	FEV_1% : Mean (SD)	n	FEV_1% : Mean (SD)	n	FEV_1% : Mean (SD)	p-values (t-test*)
6–7	455	89.9 (15.9)	529	92.8 (16.1)	424	98.8 (16.0)	<0.001
8–9	420	87.6 (15.1)	555	91.5 (14.9)	484	98.2 (13.8)	<0.001
10–11	428	83.8 (16.1)	509	88.2 (16.2)	484	96.2 (14.5)	<0.001
12–15	866	77.5 (18.3)	962	84.3 (17.7)	1065	94.0 (14.6)	<0.001
16–19	950	72.3 (21.4)	791	74.9 (20.5)	832	91.0 (17.4)	<0.001
20–23	920	66.7 (23.4)	954	68.1 (23.4)	780	84.4 (20.3)	<0.001
24–27	835	63.3 (24.0)	846	64.8 (23.4)	785	79.1 (22.2)	<0.001
28–31	652	62.0 (23.4)	776	61.7 (23.1)	773	74.5 (23.1)	<0.001
32–35	525	58.9 (23.4)	620	61.4 (23.5)	714	71.7 (23.0)	<0.001
36–39	317	60.2 (24.0)	487	58.8 (24.2)	593	70.9 (23.4)	<0.001
40–43	253	59.9 (23.1)	339	60.0 (24.2)	440	67.1 (23.7)	<0.001
44–47	186	58.6 (24.7)	220	62.8 (22.7)	341	67.8 (23.1)	0.011
48–51	153	56.5 (23.6)	200	59.1 (22.9)	197	66.6 (22.0)	<0.001
52–55	79	57.3 (24.9)	146	60.1 (24.5)	189	66.4 (23.3)	0.017
56–59	47	58.6 (22.7)	97	61.0 (24.6)	134	65.7 (23.4)	0.137
60–63	27	57.1 (22.3)	54	65.6 (25.9)	93	69.0 (20.8)	0.397
64–67	21	62.1 (28.8)	31	53.0 (18.5)	50	65.5 (27.4)	0.028
68+	30	59.3 (26.0)	48	64.5 (26.6)	91	68.6 (23.6)	0.362
<16	2169	83.3 (17.5)	2555	88.4 (16.9)	2457	96.1 (14.8)	–
≥16	4995	64.0 (23.7)	5609	64.6 (23.7)	6012	76.1 (23.4)	–
<18	2623	81.7 (18.4)	2939	87.0 (17.5)	2889	95.5 (15.2)	–
≥18	4541	63.0 (23.8)	5225	63.7 (23.7)	5580	74.9 (23.4)	–
Overall	**7164****	**69.9 (23.8)**	**8164**	**72.1 (24.4)**	**8469**	**81.9 (23.1)**	.

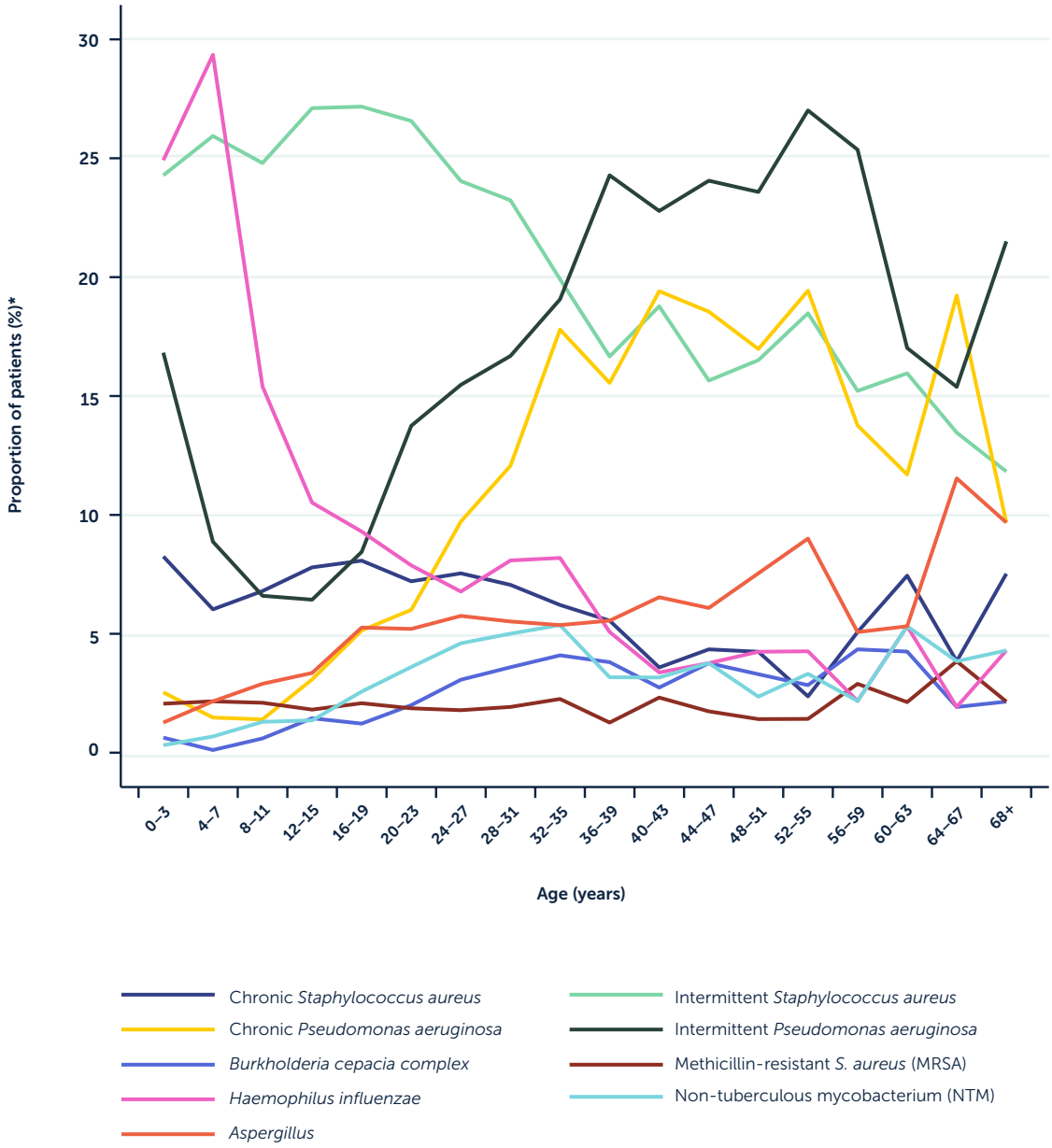
* T-test comparing 2024 with 2019

** Number with non-missing data

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.

1.17 Lung infections in 2024
N=9696*



* 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 2024 (cont.)
<16 years N=3688, ≥16 years N=6736

Infections in this table reflect those grown in the 12 months prior to the 2024 annual review.
The UK CF Registry definition of 'chronic' is three or more isolates in the last 12 months.

	Paediatric age range (years)				Overall
	0–3	4–7	8–11	12–15	Paediatric (<16 years)
Number in age range	645	899	1020	1124	3688
Number who had culture taken*	630	879	1000	1103	3612
Chronic *S. aureus* n(%)	52 (8.3)	53 (6.0)	68 (6.8)	86 (7.8)	259 (7.2)
Intermittent *S. aureus* n(%)	153 (24.3)	228 (25.9)	248 (24.8)	299 (27.1)	928 (25.7)
Chronic *P. aeruginosa* n(%)	16 (2.5)	13 (1.5)	14 (1.4)	34 (3.1)	77 (2.1)
Intermittent *P. aeruginosa* n(%)	106 (16.8)	78 (8.9)	66 (6.6)	71 (6.4)	321 (8.9)
B. cepacia complex n(%)	<5	<5	6 (0.6)	16 (1.5)	27 (0.7)
B. cenocepacia n(%)	<5	<5	<5	10 (0.9)	14 (0.4)
B. multivorans n(%)	<5	<5	<5	<5	<5
B. other cepacia n(%)	<5	<5	<5	5 (0.5)	9 (0.2)
MRSA n(%)	13 (2.1)	19 (2.2)	21 (2.1)	20 (1.8)	73 (2.0)
H. influenza n(%)	157 (24.9)	258 (29.4)	154 (15.4)	116 (10.5)	685 (19.0)
NTM n(%)	<5	6 (0.7)	13 (1.3)	15 (1.4)	36 (1.0)
Aspergillus fumigatus n(%)	8 (1.3)	19 (2.2)	29 (2.9)	37 (3.4)	93 (2.6)

* Proportions are calculated from the number of people who were recorded as having at least one respiratory culture sample taken.

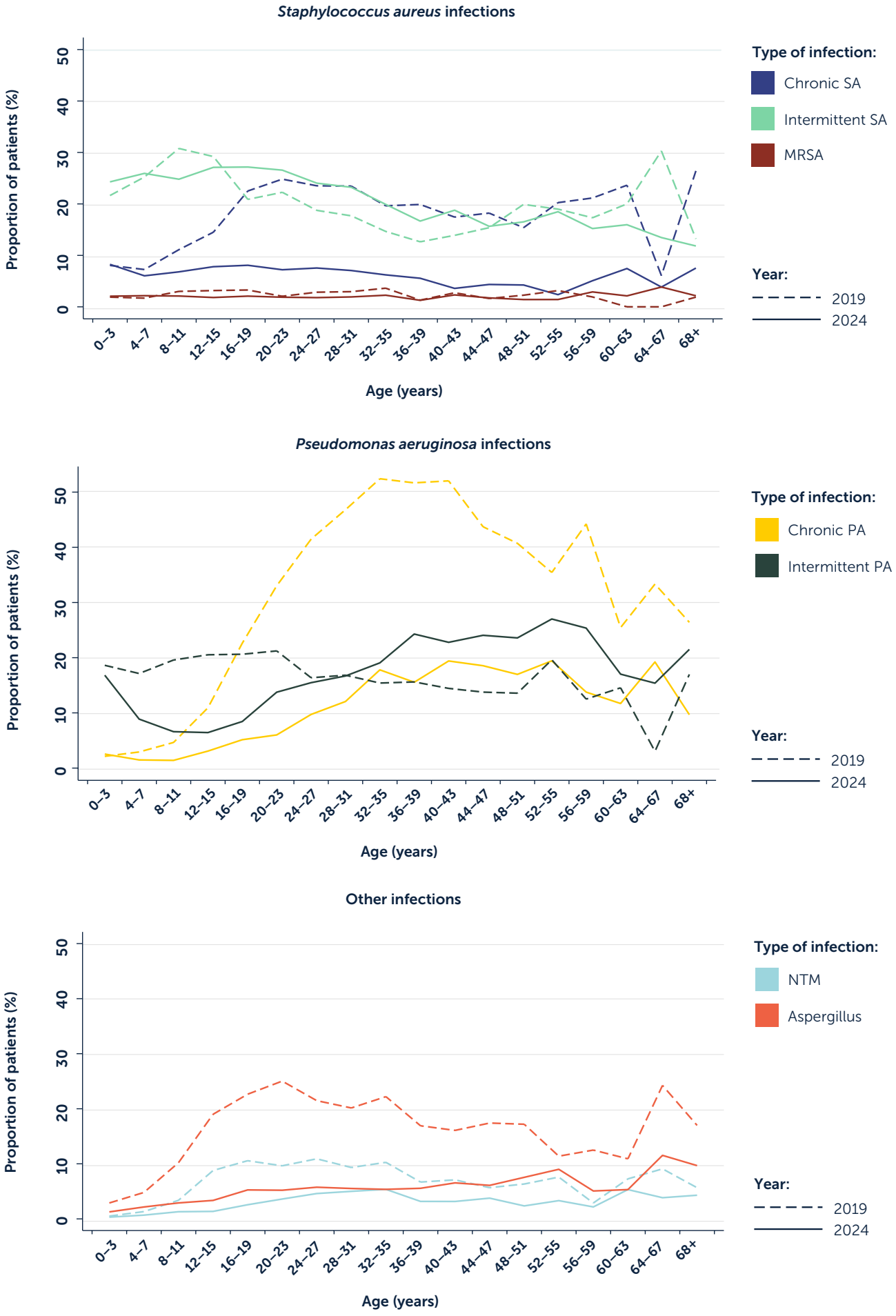
1.17 Lung infections in 2024 (cont.)
<16 years N=3688, ≥16 years N=6736

	Adult 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	874	845	870	859	806	673	533	6736
Number who had culture taken*	817	749	782	779	708	630	474	6084
Chronic *S. aureus* n(%)	66 (8.1)	54 (7.2)	59 (7.5)	55 (7.1)	44 (6.2)	35 (5.6)	17 (3.6)	382 (6.3)
Intermittent *S. aureus* n(%)	222 (27.2)	199 (26.6)	188 (24.0)	181 (23.2)	141 (19.9)	105 (16.7)	89 (18.8)	1307 (21.5)
Chronic *P. aeruginosa* n(%)	42 (5.1)	45 (6.0)	76 (9.7)	94 (12.1)	126 (17.8)	98 (15.6)	92 (19.4)	763 (12.5)
Intermittent *P. aeruginosa* n(%)	69 (8.4)	103 (13.8)	121 (15.5)	130 (16.7)	135 (19.1)	153 (24.3)	108 (22.8)	1088 (17.9)
B. cepacia complex n(%)	10 (1.2)	15 (2.0)	24 (3.1)	28 (3.6)	29 (4.1)	24 (3.8)	13 (2.7)	182 (3.0)
B. cenocepacia n(%)	<5	5 (0.7)	6 (0.8)	8 (1.0)	12 (1.7)	6 (1.0)	<5	56 (0.9)
B. multivorans n(%)	5 (0.6)	7 (0.9)	11 (1.4)	16 (2.1)	11 (1.6)	14 (2.2)	9 (1.9)	85 (1.4)
B. other cepacia n(%)	<5	<5	<5	<5	<5	<5	<5	24 (0.4)
MRSA n(%)	17 (2.1)	14 (1.9)	14 (1.8)	15 (1.9)	16 (2.3)	8 (1.3)	11 (2.3)	117 (1.9)
H. influenza n(%)	76 (9.3)	59 (7.9)	53 (6.8)	63 (8.1)	58 (8.2)	32 (5.1)	16 (3.4)	401 (6.6)
NTM n(%)	21 (2.6)	27 (3.6)	36 (4.6)	39 (5.0)	38 (5.4)	20 (3.2)	15 (3.2)	235 (3.9)
Aspergillus fumigatus n(%)	43 (5.3)	39 (5.2)	45 (5.8)	43 (5.5)	38 (5.4)	35 (5.6)	31 (6.5)	357 (5.9)

	Adult age range (years)							Overall
	44–47	48–51	52–55	56–59	60–63	64–67	68+	Adults (≥16 years)
Number in age range	387	242	234	152	107	57	97	6736
Number who had culture taken*	345	212	211	138	94	52	93	6084
Chronic *S. aureus* n(%)	15 (4.3)	9 (4.2)	5 (2.4)	7 (5.1)	7 (7.4)	<5	7 (7.5)	382 (6.3)
Intermittent *S. aureus* n(%)	54 (15.7)	35 (16.5)	39 (18.5)	21 (15.2)	15 (16.0)	7 (13.5)	11 (11.8)	1307 (21.5)
Chronic *P. aeruginosa* n(%)	64 (18.6)	36 (17.0)	41 (19.4)	19 (13.8)	11 (11.7)	10 (19.2)	9 (9.7)	763 (12.5)
Intermittent *P. aeruginosa* n(%)	83 (24.1)	50 (23.6)	57 (27.0)	35 (25.4)	16 (17.0)	8 (15.4)	20 (21.5)	1088 (17.9)
B. cepacia complex n(%)	13 (3.8)	7 (3.3)	6 (2.8)	6 (4.3)	<5	<5	<5	182 (3.0)
B. cenocepacia n(%)	<5	<5	<5	<5	<5	<5	<5	56 (0.9)
B. multivorans n(%)	7 (2.0)	<5	<5	<5	<5	<5	<5	85 (1.4)
B. other cepacia n(%)	<5	<5	<5	<5	<5	<5	<5	24 (0.4)
MRSA n(%)	6 (1.7)	<5	<5	<5	<5	<5	<5	117 (1.9)
H. influenza n(%)	13 (3.8)	9 (4.2)	9 (4.3)	<5	5 (5.3)	<5	<5	401 (6.6)
NTM n(%)	13 (3.8)	5 (2.4)	7 (3.3)	<5	5 (5.3)	<5	<5	235 (3.9)
Aspergillus fumigatus n(%)	21 (6.1)	16 (7.5)	19 (9.0)	7 (5.1)	5 (5.3)	6 (11.5)	9 (9.7)	357 (5.9)

* Proportions are calculated from the number of people who were recorded as having at least one respiratory culture sample taken.

1.18 Lung infections 2019–2024



1.19 Respiratory culture sample type

Overall	2019	2024
Number of people with an annual review (n)	10070	10424
Number of people with at least 3 samples of any type taken n(%)*	8656 (86.0)	6719 (64.5)
Number of people with at least 1 sample of any type taken n(%)*	9847 (97.8)	9734 (93.4)
Sample type[1]**		
Sputum; n(%)	6865 (69.7)	5726 (58.8)
Cough; n(%)	6198 (62.9)	6802 (69.9)
Bronchoalveolar lavage; n(%)	485 (4.9)	279 (2.9)
Age <16 years	**2019**	**2024**
Number of people with an annual review (n)	3966	3688
Number of people with at least 3 samples of any type taken n(%)*	3817 (96.2)	3398 (92.1)
Number of people with at least 1 sample of any type taken n(%)*	3937 (99.3)	3612 (97.9)
Sample type[1]**		
Sputum; n(%)	1668 (42.4)	1116 (30.9)
Cough; n(%)	3767 (95.7)	3530 (97.7)
Bronchoalveolar lavage; n(%)	343 (8.7)	176 (4.9)
Age ≥16 years	**2019**	**2024**
Number of people with an annual review (n)	6104	6736
Number of people with at least 3 samples of any type taken n(%)*	4839 (79.3)	3321 (49.3)
Number of people with at least 1 sample of any type taken n(%)*	5910 (96.8)	6122 (90.9)
Sample type[1]**		
Sputum; n(%)	5197 (87.9)	4610 (75.3)
Cough; n(%)	2431 (41.1)	3272 (53.4)
Bronchoalveolar lavage; n(%)	142 (2.4)	103 (1.7)

* % 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.

	2022	2023	2024
Number with annual review	**(n=10251)**	**(n=10344)**	**(n=10424)**
NTM Prevalence; n(%)	289 (3.1)	273 (2.6)	271 (2.6)
On NTM treatment in the given year; n (% of NTM prevalence in given year)	153 (52.9)	111 (40.7)	101 (37.3)
NTM Incidence[1]	147 (1.5)	144 (1.5)	151 (1.5)
M. abscessus prevalence	90 (0.9)	81 (0.8)	74 (0.7)
M. abscessus incidence[2]	29 (0.3)	32 (0.3)	34 (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

1.21a Complications 2019 and 2024 in young people (<16 years) N=3688 in 2024; N=3966 in 2019

The number shown is for a complication that has been present in the 12 months prior to the 2019 and 2024 annual reviews respectively. Proportions are from all people younger than 16 years at annual review.[1]

Complications	2019 n(%)	2024 n(%)
Respiratory related		
Nasal polyps*	120 (3.0)	66 (1.8)
Sinus disease	52 (1.3)	25 (0.7)
Asthma	272 (6.9)	89 (2.4)
ABPA	138 (3.5)	45 (1.2)
Any haemoptysis	16 (0.4)	<5
Massive haemoptysis	<5	0
Pneumothorax requiring chest tube	0	0
Cardiac complications		
Tachyarrhythmia	0	<5
Bradycardia	0	0
Cardiac arrest	0	0
Cardiomyopathy	0	0
Congenital heart disease	9 (0.2)	11 (0.3)
Heart failure	0	0
Ischaemic heart disease	0	0
Valvular disease	<5	<5
Other	9 (0.2)	15 (0.4)
Pancreas and hepatobiliary disease		
Raised liver enzymes	302 (7.6)	355 (9.6)
Liver disease	357 (9.0)	341 (9.2)
Cirrhosis with no portal hypertension	15 (0.4)	5 (0.1)
Cirrhosis with portal hypertension	28 (0.7)	11 (0.3)
Gall bladder disease requiring surgery	24 (0.6)	25 (0.7)
Pancreatitis	9 (0.2)	<5
Upper gastrointestinal (GI)		
Gastro-oesphageal reflux disease (GORD)	252 (6.4)	165 (4.5)
Peptic ulcer	0	0
GI bleed (varices as source)	<5	<5
GI bleed (non varices as source)	<5	<5

* Previous reports listed this complication as "Nasal polyps requiring surgery". The UK CF Registry does not specify a difference between nasal polyps and nasal polyps requiring surgery therefore we have changed the wording.

1 See Appendix 6 (page 94) for overall figures.

1.21a Complications 2019 and 2024 in young people (<16 years) (cont.)
N=3688 in 2024; N=3966 in 2019

Complications	2019 n(%)	2024 n(%)
Lower gastrointestinal		
Intestinal obstruction	12 (0.3)	20 (0.5)
DIOS	100 (2.5)	54 (1.5)
Fibrosing colonopathy / colonic stricture	0	0
Rectal prolapse	7 (0.2)	7 (0.2)
Renal		
Kidney stones	11 (0.3)	7 (0.2)
Renal failure	5 (0.1)	0
Musculoskeletal		
Arthritis	9 (0.2)	<5
Arthropathy	14 (0.4)	<5
Bone fracture	13 (0.3)	12 (0.3)
Osteopenia	25 (0.6)	23 (0.6)
Osteoporosis	5 (0.1)	<5
Other		
Cancer confirmed by histology	0	<5
Port inserted or replaced	85 (2.1)	29 (0.8)
Depression	14 (0.4)	<5
Hearing loss	39 (1.0)	27 (0.7)
Hypertension	5 (0.1)	<5
Urinary incontinence	81 (2.0)	54 (1.5)
Faecal incontinence	25 (0.6)	22 (0.6)
Postural anomaly	81 (2.0)	30 (0.8)

1.21b Complications 2019 and 2024 in adults (ages 16 years and older) N=6736 in 2024; N=6104 in 2019

The number shown is for a complication that has been present in the 12 months prior to the 2019 and 2024 annual reviews respectively. Proportions are from all people ages 16 years and older at annual review.[1]

Complications	2019 n(%)	2024 n(%)
Respiratory related		
Nasal polyps*	306 (5.0)	310 (4.6)
Sinus disease	745 (12.2)	757 (11.2)
Asthma	653 (10.7)	600 (8.9)
ABPA	616 (10.1)	446 (6.6)
Any haemoptysis	404 (6.6)	159 (2.4)
Massive haemoptysis	36 (0.6)	6 (0.1)
Pneumothorax requiring chest tube	27 (0.4)	5 (0.1)
Cardiac complications		
Tachyarrhythmia	25 (0.4)	26 (0.4)
Bradycardia	5 (0.1)	<5
Cardiac arrest	<5	<5
Cardiomyopathy	8 (0.1)	18 (0.3)
Congenital heart disease	9 (0.1)	12 (0.2)
Heart failure	<5	10 (0.1)
Ischaemic heart disease	6 (0.1)	9 (0.1)
Valvular disease	5 (0.1)	5 (0.1)
Other	58 (1.0)	64 (1.0)
Pancreas and hepatobiliary disease		
Raised liver enzymes	708 (11.6)	915 (13.6)
Liver disease	1110 (18.2)	1577 (23.4)
Cirrhosis with no portal hypertension	48 (0.8)	87 (1.3)
Cirrhosis with portal hypertension	107 (1.8)	114 (1.7)
Gall bladder disease requiring surgery	119 (1.9)	237 (3.5)
Pancreatitis	56 (0.9)	48 (0.7)
Upper gastrointestinal (GI)		
Gastro-oesphageal reflux disease (GORD)	1444 (23.7)	1469 (21.8)
Peptic ulcer	<5	<5
GI bleed (varices as source)	10 (0.2)	10 (0.1)
GI bleed (non varices as source)	11 (0.2)	14 (0.2)

* Previous reports listed this complication as "Nasal polyps requiring surgery". The UK CF Registry does not specify a difference between nasal polyps and nasal polyps requiring surgery therefore we have changed the wording.

1 See Appendix 6 (page 94) for overall figures.

1.21b Complications 2019 and 2024 in adults (ages 16 years and older) (cont.)
N=6736 in 2024; N=6104 in 2019

Complications	2019 n(%)	2024 n(%)
Lower gastrointestinal		
Intestinal obstruction	22 (0.4)	29 (0.4)
DIOS	470 (7.7)	310 (4.6)
Fibrosing colonopathy / colonic stricture	<5	<5
Rectal prolapse	5 (0.1)	6 (0.1)
Renal		
Kidney stones	100 (1.6)	170 (2.5)
Renal failure	92 (1.5)	92 (1.4)
Musculoskeletal		
Arthritis	91 (1.5)	118 (1.8)
Arthropathy	236 (3.9)	201 (3.0)
Bone fracture	34 (0.6)	24 (0.4)
Osteopenia	950 (15.6)	1123 (16.7)
Osteoporosis	409 (6.7)	536 (8.0)
Other		
Cancer confirmed by histology	28 (0.5)	29 (0.4)
Port inserted or replaced	188 (3.1)	56 (0.8)
Depression	432 (7.1)	492 (7.3)
Hearing loss	300 (4.9)	381 (5.7)
Hypertension	121 (2.0)	225 (3.3)
Urinary incontinence	858 (14.1)	644 (9.6)
Faecal incontinence	49 (0.8)	59 (0.9)
Postural anomaly	883 (14.5)	508 (7.5)

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.

	2023			2024		
	Overall (n=10344)	<16 years (n=3755)	≥16 years (n=6588)	Overall (n=10424)	<16 years (n=3688)	≥16 years (n=6736)
ABPA	118 (1.1)	23 (0.6)	95 (1.4)	120 (1.2)	17 (0.5)	103 (1.5)
Cirrhosis – no portal hypertension	–*	<5	36 (0.5)	–*	<5	41 (0.6)
Cirrhosis – with portal hypertension	43 (0.4)	7 (0.2)	36 (0.5)	–*	<5	28 (0.4)
Cancer confirmed by histology	20 (0.2)	0	20 (0.3)	–*	<5	21 (0.3)

1.23 CF diabetes**
N=8369

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=8369)	10-15 years (n=1633)	≥16 years (n=6736)
On CFD treatment			
People on CFD treatment; n (%)	2327 (27.8)	151 (9.2)	2176 (32.3)
Insulin; n(%)[1]	1911 (82.1)	144 (95.4)	1767 (81.2)
CFD Screening			
People screened; n(%)	3841 (45.9)	1077 (66.0)	2764 (41.0)
Screening Type			
Continous glucose monitoring; n(%)[2]	1468 (38.2)	375 (34.8)	1093 (39.5)
Oral glucose tolerance test; n(%)[2]	1471 (38.3)	538 (50.0)	933 (33.8)
Not screened (other)	2363 (28.2)	88 (5.4)	2275 (33.8)
Not screened (known CFD)	1933 (23.1)	375 (23.0)	1558 (23.1)
Unknown	232 (2.8)	93 (5.7)	139 (2.1)

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

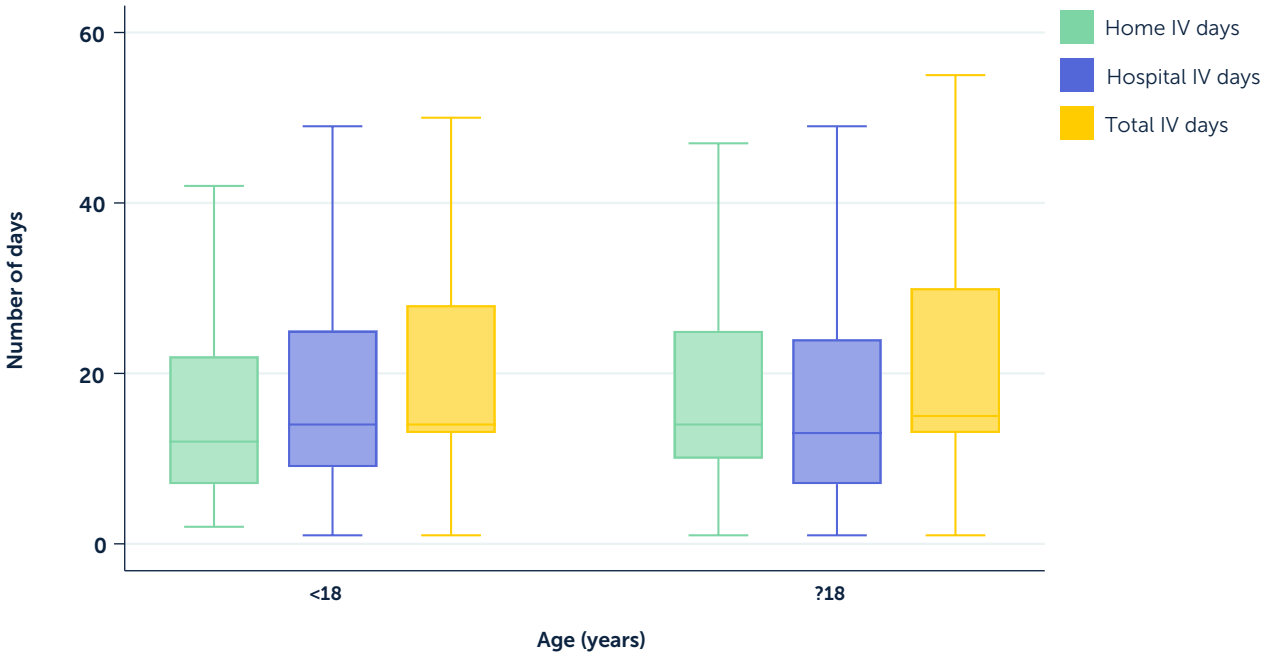
Antibiotics

1.24 Intravenous (IV) antibiotics
N=10424

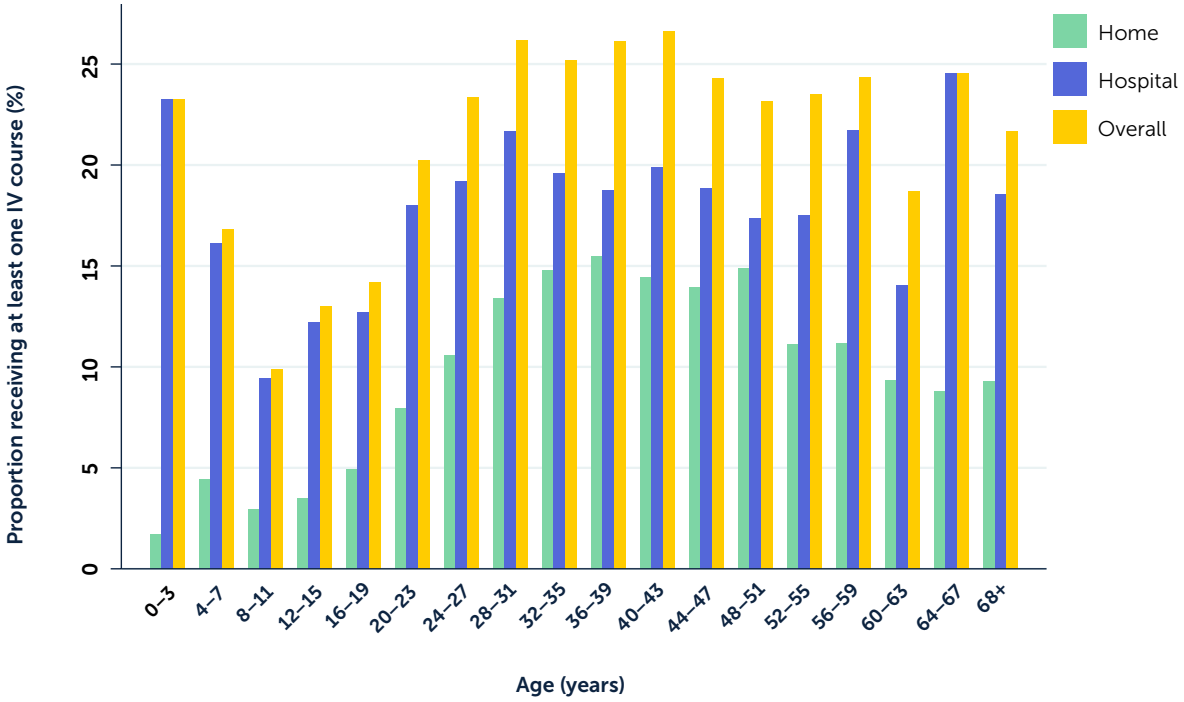
When someone with CF becomes unwell with an infection, they might be prescribed intravenous (IV) antibiotics. This treatment can take a number of days and may take place as a hospital inpatient, or at home.

		Home		Hospital		Total	
Age	n	Patients n(%)	Median days (IQR)	Patients n(%)	Median days (IQR)	Patients n(%)	Median days (IQR)
0–3	645	11 (1.7)	10 (7–13)	150 (23.3)	14 (9–19)	150 (23.3)	14 (10–24)
4–7	899	40 (4.4)	10 (7–14)	145 (16.1)	14 (7–18)	151 (16.8)	14 (12–24)
8–11	1020	30 (2.9)	12 (8–25)	96 (9.4)	14 (8–21)	101 (9.9)	14 (13–28)
12–15	1124	39 (3.5)	13 (7–22)	137 (12.2)	14 (11–31)	146 (13.0)	14 (14–36)
16–19	874	43 (4.9)	14 (11–27)	111 (12.7)	14 (9–28)	124 (14.2)	16 (14–34)
20–23	845	67 (7.9)	13 (7–21)	152 (18.0)	14 (9–24)	171 (20.2)	14 (12–30)
24–27	870	92 (10.6)	14 (10–24)	167 (19.2)	13 (7–24)	203 (23.3)	14 (13–30)
28–31	859	115 (13.4)	14 (9–28)	186 (21.7)	13 (7–24)	225 (26.2)	14 (13–31)
32–35	806	119 (14.8)	14 (11–27)	158 (19.6)	14 (7–26)	203 (25.2)	17 (14–39)
36–39	673	104 (15.5)	14 (12–22)	126 (18.7)	13 (7–23)	176 (26.2)	14 (13–28)
40–43	533	77 (14.4)	14 (13–28)	106 (19.9)	14 (7–22)	142 (26.6)	18 (13–31)
44–47	387	54 (14.0)	14 (8–21)	73 (18.9)	13 (6–21)	94 (24.3)	14 (13–28)
48–51	242	36 (14.9)	14 (10–20)	42 (17.4)	10 (6–21)	56 (23.1)	14 (14–30)
52–55	234	26 (11.1)	14 (13–27)	41 (17.5)	14 (8–17)	55 (23.5)	14 (14–28)
56–59	152	17 (11.2)	14 (11–22)	33 (21.7)	14 (9–28)	37 (24.3)	20 (14–30)
60–63	107	10 (9.3)	14 (10–14)	15 (14.0)	14 (10–46)	20 (18.7)	14 (14–30)
64–67	57	5 (8.8)	11 (5–16)	14 (24.6)	10 (7–13)	14 (24.6)	14 (11–16)
68+	97	9 (9.3)	11 (6–14)	18 (18.6)	14 (9–24)	21 (21.6)	14 (14–42)
<16	3688	120 (3.3)	12 (7–22)	528 (14.3)	14 (9–24)	548 (14.9)	14 (13–28)
≥16	6736	774 (11.5)	14 (10–25)	1242 (18.4)	14 (7–24)	1541 (22.9)	15 (13–30)
<18	4143	135 (3.3)	12 (7–22)	580 (14.0)	14 (9–25)	606 (14.6)	14 (13–28)
≥18	6281	759 (12.1)	14 (10–25)	1190 (18.9)	13 (7–24)	1483 (23.6)	15 (13–30)
Overall	**10424**	**894 (8.6)**	**14 (10–24)**	**1770 (17.0)**	**14 (7–24)**	**2089 (20.0)**	**14 (13–29)**

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 72.



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 8.6% and in hospital was 17.0%. The proportion receiving any IVs was 20.0%.



1.25 Inhaled antibiotic use
N=10424

	2024		
	Overall	<16 years	≥16 years
Number of patients	10424	3688	6736
Tobramycin solution; n(%)	859 (8.2)	190 (5.2)	669 (9.9)
Other aminoglycoside; n(%)	42 (0.4)	5 (0.1)	37 (0.5)
Colistin; n(%)	1026 (9.8)	338 (9.2)	688 (10.2)
Promixin; n(%)	1145 (11.0)	219 (5.9)	926 (13.7)
Aztreonam; n(%)	672 (6.4)	35 (0.9)	637 (9.5)
Colistimethate (DPI); n(%)	833 (8.0)	45 (1.2)	788 (11.7)
Tobramycin Inhalation Powder; n(%)	562 (5.4)	15 (0.4)	547 (8.1)
Levofloxacin; n(%)	71 (0.7)	0 (0.0)	71 (1.1)
At least one of the above; n(%)	3762 (36.1)	649 (17.6)	3113 (46.2)

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 *Pseudomonas aeruginosa* should be prescribed at least one of the below inhaled antibiotics.

	2014			2019			2024		
	Overall	<16 years	≥16 years	Overall	<16 years	≥16 years	Overall	<16 years	≥16 years
Patients with chronic *P. aeruginosa*	2963	305	2658	2609	206	2403	840	77	763
Tobramycin solution; n(%)	841 (28.4)	96 (31.5)	745 (28.0)	623 (23.9)	87 (42.2)	536 (22.3)	176 (21.0)	29 (37.7)	147 (19.3)
Other aminoglycoside; n(%)	139 (4.7)	22 (7.2)	117 (4.4)	29 (1.1)	0	29 (1.2)	–*	<5	10 (1.3)
Colistin; n(%)	1101 (37.2)	156 (51.1)	945 (35.6)	615 (23.6)	81 (39.3)	534 (22.2)	185 (22.0)	26 (33.8)	159 (20.8)
Promixin; n(%)	919 (31.0)	134 (43.9)	785 (18.2)	771 (29.6)	97 (47.1)	674 (28.0)	177 (21.1)	32 (41.6)	145 (19.0)
Aztreonam; n(%)	395 (13.3)	10 (3.3)	385 (14.5)	690 (26.4)	15 (7.3)	675 (28.1)	212 (25.2)	6 (7.8)	206 (27.0)
Colistimethate (DPI); n(%)	433 (14.6)	21 (6.9)	412 (15.5)	457 (17.5)	11 (5.3)	446 (18.6)	163 (19.4)	6 (7.8)	157 (20.6)
Tobramycin Inhalation Powder; n(%)	802 (27.1)	24 (7.9)	778 (29.3)	573 (22.0)	9 (4.4)	564 (23.5)	–*	<5	96 (12.6)
Levofloxacin; n(%)	–**	–**	–**	<5	0	<5	37 (4.4)	0	37 (4.8)
At least one of the above; n(%)	**2625 (88.6)**	**288 (94.4)**	**2337 (87.9)**	**2315 (88.7)**	**189 (91.7)**	**2126 (88.5)**	**702 (83.6)**	**72 (93.5)**	**630 (82.6)**

* Redacted to adhere to statistical disclosure guidelines

** No data in the given year as the drug in question was not licensed for use on the NHS

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(%)
2014	Overall	3705	1945 (52.5)	1760 (47.5)
	0–3 years	39	5 (12.8)	34 (87.2)
	4–15 years	594	101 (17.0)	493 (83.0)
	≥ 16 years	3072	1839 (59.9)	1233 (40.1)
2019	Overall	4130	1772 (42.9)	2358 (57.1)
	0–3 years	–*	<5	44 (95.7)
	4–15 years	672	99 (14.7)	573 (85.3)
	≥ 16 years	3412	1671 (49.0)	1741 (51.0)
2024	Overall	3232	513 (15.9)	2719 (84.1)
	0–3 years	–*	<5	62 (95.4)
	4–15 years	350	27 (7.7)	323 (92.3)
	≥ 16 years	2817	483 (17.1)	2334 (82.9)

1.28 Prophylactic flucloxacillin use**

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

	2019		2024	
Age	Total patients	Patients on prophylactic flucloxacillin; n(%)	Total patients	Patients on prophylactic flucloxacillin; n(%)
0–3	803	454 (56.5)	645	318 (49.3)
4–7	1075	297 (27.6)	899	193 (21.5)
8–11	1105	294 (26.6)	1020	168 (16.5)
12–15	983	229 (23.3)	1124	186 (16.5)
16–19	812	172 (21.2)	874	107 (12.2)
20–23	996	113 (11.3)	845	82 (9.7)
24–27	902	69 (7.6)	870	77 (8.9)
28–31	852	66 (7.7)	859	37 (4.3)
32–35	695	35 (5.0)	806	37 (4.6)
36–39	564	37 (6.6)	673	24 (3.6)
40–43	379	32 (8.4)	533	20 (3.8)
44–47	256	16 (6.2)	387	15 (3.9)
48–51	229	14 (6.1)	242	7 (2.9)
52–55	164	9 (5.5)	234	12 (5.1)
56–59	110	5 (4.5)	152	8 (5.3)
60–63	57	<5	107	<5
64–67	33	<5	57	<5
68+	55	<5	97	<5
<16 years	3966	1274 (32.1)	3688	865 (23.5)
≥16 years	6104	576 (9.4)	6736	432 (6.4)
<18 years	4362	1374 (31.5)	4143	920 (22.2)
≥18 years	5708	476 (8.3)	6281	377 (6.0)
Overall	**10070**	**1850 (18.4)**	**10424**	**1297 (12.4)**

* Redacted to adhere to statistical disclosure guidelines.

** Data incudes patients that have been recruited and randomised for the CF START trial from 2018–2023. The CF START trial is investigating the use of prophylactic flucloxacillin use in infants. You can learn more about this trial here: **cfstart.org.uk**

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	1079	255 (23.6)	89 (8.2)	8 (0.7)
6–≤16 years	2842	1264 (44.5)	390 (13.7)	290 (10.2)
6–≤18 years	3273	1500 (45.8)	447 (13.7)	368 (11.2)
<16 years	3688	1398 (37.9)	459 (12.4)	269 (7.3)
≥16 years	6736	4479 (66.5)	1093 (16.2)	1723 (25.6)
<18 years	4143	1643 (39.7)	516 (12.5)	333 (8.0)
≥18 years	6281	4234 (67.4)	1036 (16.5)	1659 (26.4)
Overall	**10424**	**5877 (56.4)**	**1552 (14.9)**	**1992 (19.1)**

Mucoactive therapies

1.30 Mannitol**

	2019		2024	
Age	Total patients	Patients on Mannitol; n (%)	Total patients	Patients on Mannitol; n (%)
<16 years	3966	<5	3688	<5
≥16 years	6104	342 (5.6)	6736	194 (2.9)
<18 years	4362	–*	4143	–*
≥18 years	5708	337 (5.9)	6281	190 (3.0)
Overall	**10070**	**–***	**10424**	**–***

1.31 DNase**

	2014		2019		2024	
Age	Total patients	Patients on DNase; n(%)	Total patients	Patients on DNase; n(%)	Total patients	Patients on DNase; n(%)
0–3	963	114 (11.8)	803	155 (19.3)	645	139 (21.6)
4–7	1044	415 (39.8)	1075	584 (54.3)	899	458 (50.9)
8–11	906	558 (61.6)	1105	858 (77.6)	1020	722 (70.8)
12–15	927	648 (69.9)	983	841 (85.6)	1124	808 (71.9)
16–19	1020	701 (68.7)	812	682 (84.0)	874	600 (68.6)
20–23	1002	632 (63.1)	996	795 (79.8)	845	523 (61.9)
24–27	932	609 (65.3)	902	684 (75.8)	870	528 (60.7)
28–31	728	447 (61.4)	852	615 (72.2)	859	491 (57.2)
32–35	589	355 (60.3)	695	477 (68.6)	806	447 (55.5)
36–39	361	186 (51.5)	564	376 (66.7)	673	377 (56.0)
40–43	309	150 (48.5)	379	235 (62.0)	533	282 (52.9)
44–47	231	114 (49.4)	256	145 (56.6)	387	210 (54.3)
48–51	179	94 (52.5)	229	133 (58.1)	242	121 (50.0)
52–55	96	47 (49.0)	164	84 (51.2)	234	115 (49.1)
56–59	55	27 (49.1)	110	62 (56.4)	152	72 (47.4)
60–63	33	17 (51.5)	57	31 (54.4)	107	58 (54.2)
64–67	27	14 (51.9)	33	18 (54.5)	57	35 (61.4)
68+	30	15 (50.0)	55	26 (47.3)	97	48 (49.5)
<16 years	3840	1735 (45.2)	3966	2438 (61.5)	3688	2127 (57.7)
≥16 years	5592	3408 (60.9)	6104	4363 (71.5)	6736	3907 (58.0)
<18 years	4328	2078 (48.0)	4362	2770 (63.5)	4143	2463 (59.4)
≥18 years	5104	3065 (60.1)	5708	4031 (70.6)	6281	3571 (56.9)
Overall	**9432**	**5143 (54.5)**	**10070**	**6801 (67.5)**	**10424**	**6034 (57.9)**

* Redacted to adhere to statistical disclosure guidelines.

** CF STORM, a clinical trial investigating the withdrawal of mucoactive therapies, was enrolling patients during the 2024 data collection year. See **www.cfstorm.org.uk** for more information.

1.32 Hypertonic saline*

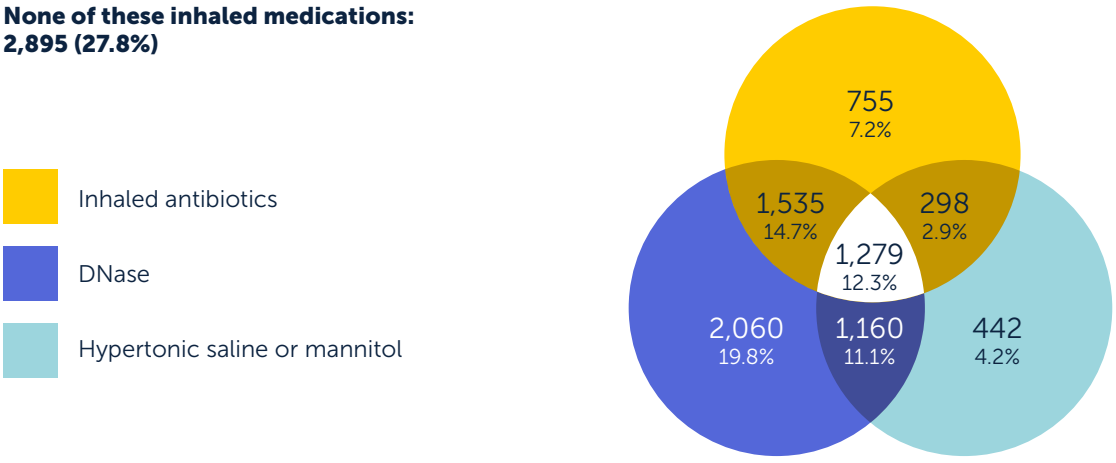
This treatment helps to thin mucus so that it is easier to cough out of the body.

	2014		2019		2024	
Age	Total patients	Patients on hypertonic saline; n(%)	Total patients	Patients on hypertonic saline; n(%)	Total patients	Patients on hypertonic saline; n(%)
0–3	963	64 (6.6)	803	91 (11.3)	645	125 (19.4)
4–7	1044	173 (16.6)	1075	304 (28.3)	899	323 (35.9)
8–11	906	234 (25.8)	1105	416 (37.6)	1020	360 (35.3)
12–15	927	340 (36.7)	983	458 (46.6)	1124	445 (39.6)
16–19	1020	328 (32.2)	812	409 (50.4)	874	304 (34.8)
20–23	1002	285 (28.4)	996	389 (39.1)	845	261 (30.9)
24–27	932	269 (28.9)	902	307 (34.0)	870	246 (28.3)
28–31	728	251 (34.5)	852	269 (31.6)	859	211 (24.6)
32–35	589	186 (31.6)	695	233 (33.5)	806	162 (20.1)
36–39	361	96 (26.6)	564	202 (35.8)	673	145 (21.5)
40–43	309	65 (21.0)	379	123 (32.5)	533	147 (27.6)
44–47	231	63 (27.3)	256	80 (31.2)	387	80 (20.7)
48–51	179	45 (25.1)	229	65 (28.4)	242	59 (24.4)
52–55	96	27 (28.1)	164	46 (28.0)	234	59 (25.2)
56–59	55	14 (25.5)	110	28 (25.5)	152	36 (23.7)
60–63	33	8 (24.2)	57	12 (21.1)	107	25 (23.4)
64–67	27	8 (29.6)	33	8 (24.2)	57	16 (28.1)
68+	30	10 (33.3)	55	20 (36.4)	97	23 (23.7)
<16 years	3840	811 (21.1)	3966	1269 (32.0)	3688	1253 (34.0)
≥16 years	5592	1655 (29.6)	6104	2191 (35.9)	6736	1774 (26.3)
<18 years	4328	970 (22.4)	4362	1473 (33.8)	4143	1432 (34.6)
≥18 years	5104	1496 (29.3)	5708	1987 (34.8)	6281	1595 (25.4)
Overall	**9432**	**2466 (26.1)**	**10070**	**3460 (34.4)**	**10424**	**3027 (29.0)**

1.33 Inhaled therapies*

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: 2,895 (27.8%)



* CF STORM, a clinical trial investigating the withdrawal of mucoactive therapies, was enrolling patients during the 2024 data collection year. See **www.cfstorm.org.uk** for more information

CFTR modulators

Here we describe the eligibility for and access to CFTR modulators throughout[1] 2024, including the NHS agreement in mid-2024 for long-term access to CFTR modulators. The access arrangements prior to 2024 are described in previous annual reports.

Elexacaftor/tezacaftor/ivacaftor

During 2024, elexacaftor/tezacaftor/ivacaftor (ETI) was available in the UK for patients with cystic fibrosis aged 2 years and over who have at least one copy of the F508del variant. Guidance was issued throughout the year from NHS commissioners across the devolved nations to support the prescribing of ETI "off-label". This varies slightly across the devolved nations but covers the 178 variants that are on an approved FDA list and the 7 named variants in the French Compassionate Use Programme. Individuals with at least one copy of one of the variants on the approved list were potentially eligible for ETI.

Lumacaftor/ivacaftor

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

Ivacaftor

Ivacaftor is licensed for use for people aged four months and older with at least one copy of a CFTR gating mutation, one copy of the R117H gene, or one copy of one of the 97 named variants on the approved FDA list[2], which are mutations considered responsive to ivacaftor monotherapy based on in-vitro and clinical data.

Tezacaftor/ivacaftor

Tezacaftor/ivacaftor is licensed for use in patients aged six and over who have two copies of the F508del mutation, or a single copy of F508del and one of 14 residual function mutations[3]. Tezacaftor/ivacaftor is also available for use in patients aged six and over who have at least one copy of one of the 154 named variants on the approved FDA list[4], which are mutations considered responsive to tezacaftor/ivacaftor combination therapy based on in-vitro and clinical data.

1 **www.cysticfibrosis.org.uk/the-work-we-do/campaigning-hard/life-saving-drugs/nice-modulator-appraisal**. Note: In December 2024, access expanded to 272 named variants. The additional 94 variants named in this expansion are not considered in this report as "eligible" to access Kaftrio as this change was made at the very end of the 2024 annual review year.

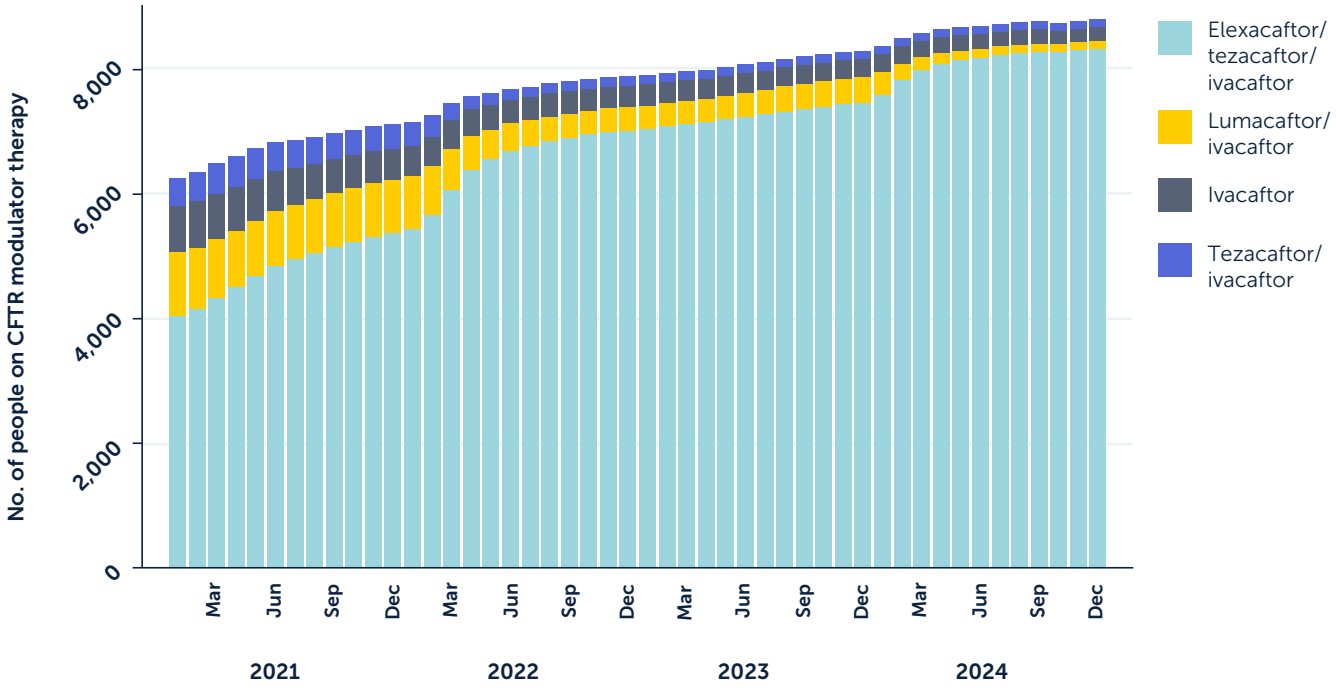
2 **pi.vrtx.com/files/uspi_ivacaftor.pdf**

3 **www.cysticfibrosis.org.uk/what-is-cystic-fibrosis/cystic-fibrosis-care/treatments-and-medication/symkevi**

4 **pi.vrtx.com/files/uspi_tezacaftor_ivacaftor.pdf**

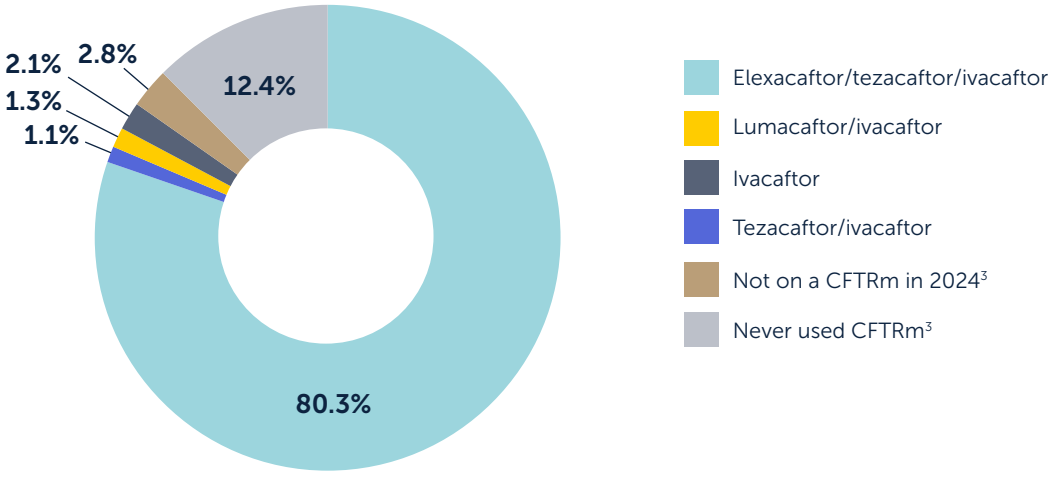
1.34 CFTR modulator uptake 2021–2024 among people with a 2024 annual review[1]
N=10,424

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 counted.



1.35a CFTR modulator use in 2024
N=10,414[2]

This chart shows the distribution of CFTR modulators taken during 2024. Where an individual had multiple prescriptions, only the most recent one was counted. 8,830 people had a record of using a CFTRm in 2024.



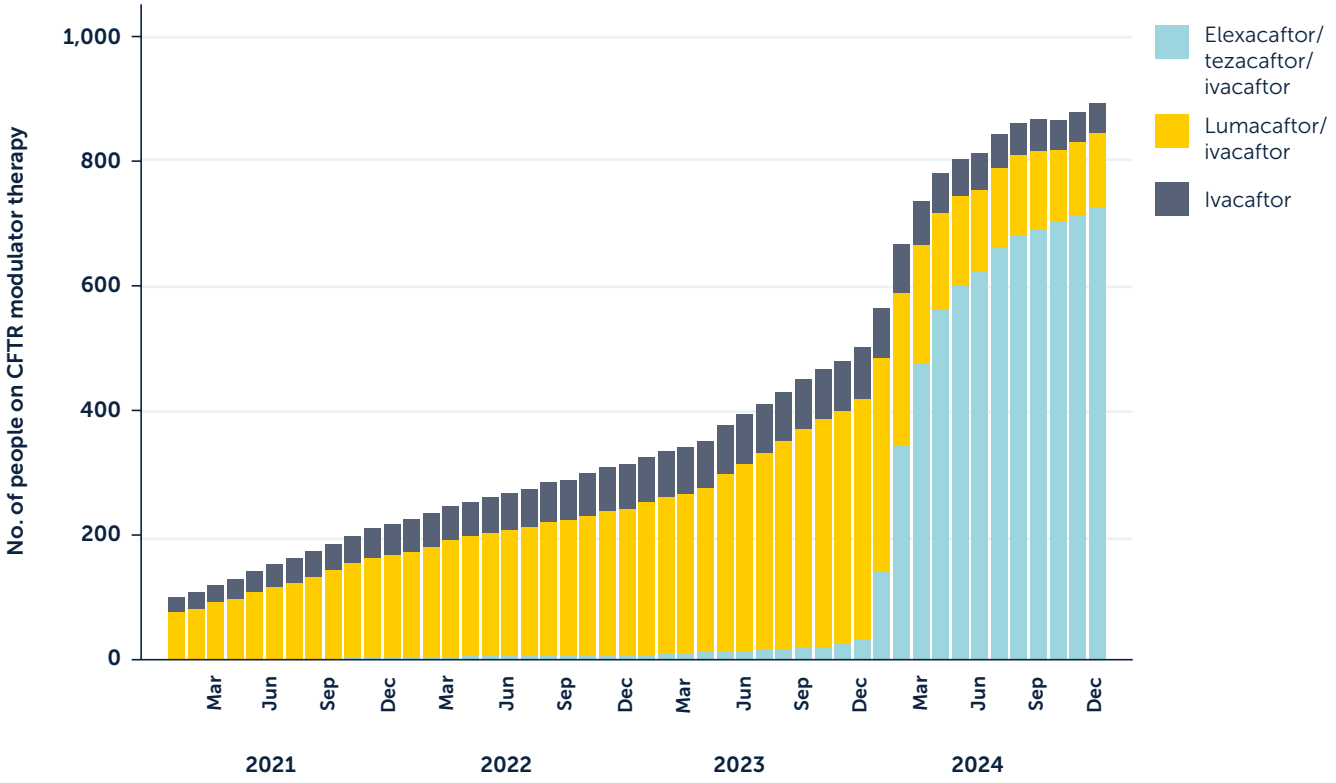
1 All people of all ages with a 2024 annual review are included.

2 10 people were excluded because their last prescribed CFTRm treatment was through a clinical trial and specific treatment was unknown.

3 An individual was categorized as 'Not on a CFTRm in 2024' if they had a previous record of CFTRm use but no record of CFTRm use in 2024. Individuals categorized as 'Never used CFTRm' had no records of CFTRms prior to or in 2024. These figures include eligible and non-eligible people.

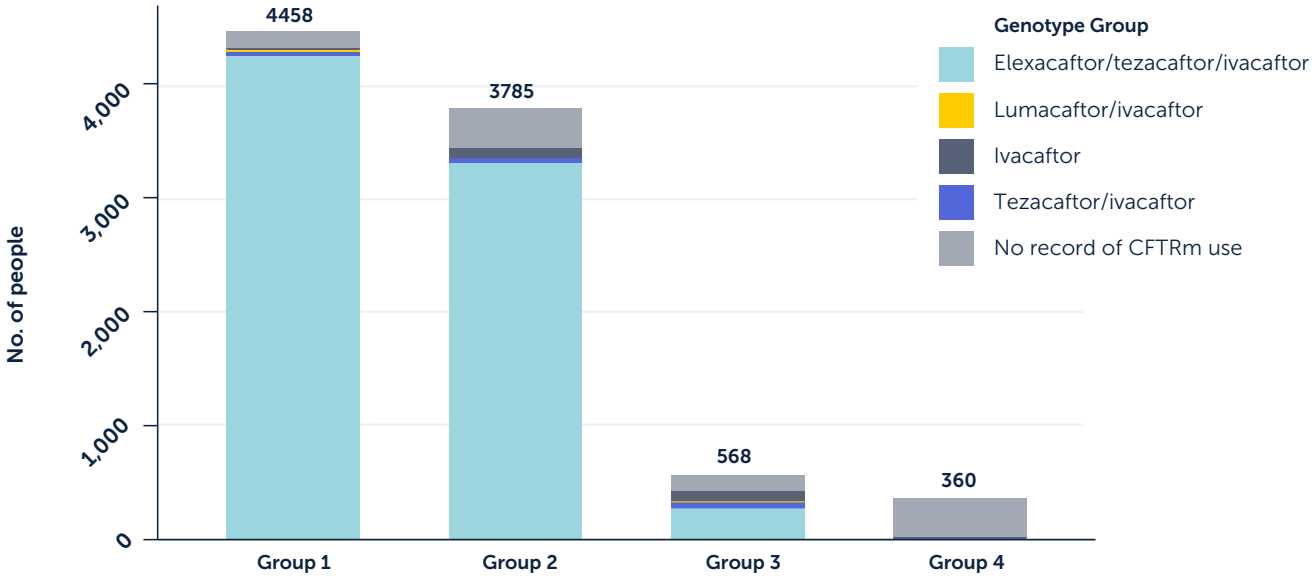
1.35b CFTR modulator uptake in 2021–2024 in people younger than 6 years of age[1]
N=1,196

The landscape of eligibility for CFTR modulator treatment has continued to expand. In 2024, elexacaftor/tezacaftor/ivacaftor, lumacaftor/ivacaftor, and ivacaftor monotherapy were available to people younger than 6 years old, with ivacaftor available for those as young as 4 months old. 76.3% of people younger than 6 years of age in 2024 have had a CFTRm record either prior to or in 2024. This graph shows the uptake of CFTR modulator use from 2021–2024 among those who were younger than 6 years on 1 January 2024.



1 All individuals included in this visual are also included in figure 1.34

1.36a CFTR modulator use in all people aged 6 years and older by genotype group[1,2]
N=9,316



Genotype group definitions	
Group 1	F508del Homozygous
Group 2	F508del Heterozygous
Group 3	No F508del, but at least one variant responsive to one of the available CFTR modulators as defined by FDA lists or French Compassionate Use Programme list (see Appendix 5 for full lists).
Group 4	No F508del and no variants from FDA and French Compassionate Use Programme lists (see Appendix 5).

		Groups			
		Group 1	Group 2	Group 3	Group 4
Most recent CFTRm used	Elexacaftor/ tezacaftor/ ivacaftor	4246 (95.2)	3309 (87.4)	276 (48.6)	17 (4.7)
	Tezacaftor/ ivacaftor	36 (0.8)	41 (1.1)	-*	<5
	Lumacaftor/ ivacaftor	-*	0 (0.0)	<5	0 (0.0)
	Ivacaftor	<5	92 (2.4)	95 (16.7)	<5
	No record of CFTRm use	149 (3.3)	343 (9.1)	134 (23.6)	338 (93.9)
	Total	**4458**	**3785**	**568**	**360**

1 All individuals included in this visual are also included in figure 1.34
2 46 people were excluded because their genotype data is missing or unknown.
* Redacted to adhere to statistical disclosure guidelines

1.36b Demographic characteristics in all people aged 6 years and older by genotype group and CFTRm use[1,2]
N=9,316

	Groups 1, 2, and 3[3]		Group 4[4]
	CFTRm use recorded	no record of CFTRm use	no record of CFTRm use
Number of individuals (n)	8195	626	338
Age			
6–11 years; n(%)	1374 (16.8)	99 (15.8)	59 (17.5)
12–17 years; n(%)	1367 (16.7)	77 (12.3)	65 (19.2)
18+ years; n(%)	5454 (66.6)	450 (71.9)	214 (63.3)
Median years (IQR)	25 (14, 35)	33 (15, 43)	22 (14, 33)
Demographics			
Male; n (%)	4369 (53.3)	336 (53.7)	191 (56.5)
In full or part time employment or school; n(% of people age 18+ years)	3691 (67.7)	256 (56.9)	141 (65.9)
Ethnicity; n (%)			
White	7842 (97.2)	578 (94.9)	193 (59.8)
Asian	144 (1.8)	22 (3.6)	112 (34.7)
Black	16 (0.2)	<5	-*
Mixed	28 (0.3)	<5	<5
Other	36 (0.4)	<5	9 (2.8)
Lung Health			
FEV_1 predicted; median(IQR)	86 (67, 99)	88 (71, 100)	74 (54, 87)
Best FEV_1 pred; median(IQR)	89 (70, 102)	89 (74, 101)	79 (60, 95)
At least 1 pulmonary exacerbation since last annual review; n(%)	1292 (15.8)	78 (12.5)	155 (45.9)
Nutritional Health			
Individuals ages 20 years and older; n	5054	434	193
BMI for individuals ages 20+ years; median (IQR)	22 (19, 26)	23 (20, 26)	20 (18, 24)

Genotype group definitions	
Group 1[3]	F508del Homozygous
Group 2[3]	F508del Heterozygous
Group 3[3]	No F508del, but at least one variant responsive to one of the available CFTR modulators as defined by FDA lists or French Compassionate Use Programme list (see Appendix 5 for full lists).
Group 4[4]	No F508del and no variants from FDA and French Compassionate Use Programme lists (see Appendix 5).

1 10 people were excluded because their last prescribed CFTRm treatment was through a clinical trial and specific treatment was unknown.
2 46 people were excluded because their genotype data is missing or unknown
3 People in groups 1, 2, and 3 for visual 1,36 are considered potentially eligible for CFTRms
4 People in group 4 for visual 1.36 are considered likely not to be eligible for CFTRms.
* 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=10424)	<16 years (n=3688)	≥16 years (n=6736)	<18 years (n=4143)	≥18 years (n=6281)
Active cycle of breathing techniques	515 (4.9)	33 (0.9)	482 (7.2)	43 (1.0)	472 (7.5)
Assisted autogenic drainage	114 (1.1)	17 (0.5)	97 (1.4)	25 (0.6)	89 (1.4)
Autogenic drainage	900 (8.6)	29 (0.8)	871 (12.9)	46 (1.1)	854 (13.6)
Exercise	1795 (17.2)	454 (12.3)	1341 (19.9)	539 (13.0)	1256 (20.0)
Forced expiration	50 (0.5)	11 (0.3)	39 (0.6)	11 (0.3)	39 (0.6)
High Pressure PEP	29 (0.3)	17 (0.5)	12 (0.2)	18 (0.4)	11 (0.2)
Incentive spirometer	–*	<5	11 (0.2)	<5	10 (0.2)
Manual in/ex-sufflation (cough assist)	–*	<5	13 (0.2)	<5	12 (0.2)
Manual techniques (percussion over pressures vibrations)	273 (2.6)	223 (6.0)	50 (0.7)	224 (5.4)	49 (0.8)
NIV (non-invasive ventillation)	–*	<5	35 (0.5)	<5	35 (0.6)
Oscillating PEP	2775 (26.6)	1301 (35.3)	1474 (21.9)	1490 (36.0)	1285 (20.5)
PEP	1733 (16.6)	1052 (28.5)	681 (10.1)	1130 (27.3)	603 (9.6)
Postural drainage	138 (1.3)	118 (3.2)	20 (0.3)	119 (2.9)	19 (0.3)
VEST	39 (0.4)	15 (0.4)	24 (0.4)	17 (0.4)	22 (0.4)
Other	541 (5.2)	241 (6.5)	300 (4.5)	259 (6.3)	282 (4.5)
None	1451 (13.9)	170 (4.6)	1281 (19.0)	213 (5.1)	1238 (19.7)

* Redacted to adhere to statistical disclosure guidelines

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=10424)	<16 years (n=3688)	≥16 years (n=6736)	<18 years (n=4143)	≥18 years (n=6281)
Active cycle of breathing techniques	1055 (10.1)	184 (5.0)	871 (12.9)	217 (5.2)	838 (13.3)
Assisted autogenic drainage	187 (1.8)	60 (1.6)	127 (1.9)	71 (1.7)	116 (1.8)
Autogenic drainage	1536 (14.7)	73 (2.0)	1463 (21.7)	105 (2.5)	1431 (22.8)
Exercise; of which:	6112 (58.6)	2447 (66.4)	3665 (54.4)	2765 (66.7)	3347 (53.3)
Exercise listed as only airway clearance technique	1019 (9.8)	133 (3.6)	886 (13.2)	172 (4.2)	847 (13.5)
Forced expiration	539 (5.2)	306 (8.3)	233 (3.5)	348 (8.4)	191 (3.0)
High Pressure PEP	38 (0.4)	24 (0.7)	14 (0.2)	25 (0.6)	13 (0.2)
Incentive spirometer	35 (0.3)	9 (0.2)	26 (0.4)	10 (0.2)	25 (0.4)
Manual in/ex-sufflation (cough assist)	32 (0.3)	6 (0.2)	26 (0.4)	8 (0.2)	24 (0.4)
Manual techniques (percussion over pressures vibrations)	547 (5.2)	414 (11.2)	133 (2.0)	422 (10.2)	125 (2.0)
NIV (non-invasive ventillation)	-*	<5	80 (1.2)	<5	80 (1.3)
Oscillating PEP	3821 (36.7)	1635 (44.3)	2186 (32.5)	1891 (45.6)	1930 (30.7)
PEP	2334 (22.4)	1350 (36.6)	984 (14.6)	1441 (34.8)	893 (14.2)
Postural drainage	421 (4.0)	341 (9.2)	80 (1.2)	353 (8.5)	68 (1.1)
VEST	91 (0.9)	34 (0.9)	57 (0.8)	38 (0.9)	53 (0.8)
Other	1282 (12.3)	632 (17.1)	650 (9.6)	683 (16.5)	599 (9.5)

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=10424)	<16 years (n=3688)	≥16 years (n=6736)	<18 years (n=4143)	≥18 years (n=6281)
Yes[1]	1280 (12.3)	374 (10.1)	906 (13.5)	450 (10.9)	830 (13.2)
No	7404 (71.0)	2536 (68.8)	4868 (72.3)	2844 (68.6)	4560 (72.6)
Not known or missing	1740 (16.7)	778 (21.1)	962 (14.3)	849 (20.5)	891 (14.2)
Type of exercise test[2,3]					
CPET	224 (17.5)	66 (17.6)	158 (17.4)	76 (16.9)	148 (17.8)
Shuttle test	202 (15.8)	156 (41.7)	46 (5.1)	170 (37.8)	32 (3.9)
Step test	297 (23.2)	69 (18.4)	228 (25.2)	88 (19.6)	209 (25.2)
6 minute walk test	-*	<5	88 (9.7)	-*	85 (10.2)
Other	278 (21.7)	86 (23.0)	192 (21.2)	103 (22.9)	175 (21.1)
Missing	387 (30.2)	110 (29.4)	277 (30.6)	135 (30.0)	252 (30.4)

* Redacted to adhere to statistical disclosure guidelines

1 Exercise test represents all types of testing listed including submaximal, shuttle test, 6 minute walk test, step test and other test.

2 Proportion of patients who answered 'Yes' above

3 More than one type of test can be recorded so percentages may not sum to 100%

Other therapies

1.40 Oxygen and non-invasive ventilation

	Overall (n=10424)	<16 years (n=3688)	≥16 years (n=6736)	<18 years (n=4143)	≥18 years (n=6281)
Non invasive ventilation (NIV); n (%)	128 (1.2)	18 (0.5)	110 (1.6)	25 (0.6)	103 (1.6)
Any oxygen use; n (%)	298 (2.9)	52 (1.4)	246 (3.7)	60 (1.4)	238 (3.8)
Among those who had oxygen use:					
Continuously	-*	<5	48 (19.5)	<5	47 (19.7)
Nocturnal or with exertion	108 (36.2)	5 (9.6)	103 (41.9)	8 (13.3)	100 (42.0)
As required (PRN)	-*	<5	20 (8.1)	<5	20 (8.4)
With exacerbation	119 (39.9)	44 (84.6)	75 (30.5)	48 (80.0)	71 (29.8)

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. Only individuals with an annual review in the given year are considered. N=10,424 in 2024.

	2019	2020	2021	2022	2023	2024
Number evaluated	241	175	78	41	37	28
Number accepted	96	66	23	22	21	14
Number receiving aged <16 years	<5	0	0	0	0	0
Bilateral lung	<5	0	0	0	0	0
Liver	<5	0	0	0	0	0
Other	0	0	0	0	0	0
Number receiving aged 16+ years	54	15	5	6	<5	6
Bilateral lung	49	12	<5	<5	<5	<5
Liver	<5	<5	0	0	<5	<5
Other	<5	<5	<5	<5	0	<5

* 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. The category "Any supplemental feeding" includes the subsequent categories: oral, nasogastric tube, gastronomy tube/button, jejunal, and total parenteral nutrition, and unknown or other supplemental feeding. The subsequent categories are not mutually exclusive.

Year		Overall	<16 years	≥16 years	<18 years	≥18 years
2014	**Total; n**	**9432**	**3840**	**5592**	**4328**	**5104**
	Any supplemental feeding; n(%)	3001 (31.8)	1032 (26.9)	1969 (35.2)	1192 (27.5)	1809 (35.4)
	Oral; n(%)	2520 (26.7)	844 (22.0)	1676 (30.0)	971 (22.4)	1549 (30.3)
	Nasogastric tube; n(%)	114 (1.2)	12 (0.3)	102 (1.8)	16 (0.4)	98 (1.9)
	Gastrostomy tube/Button; n(%)	572 (6.1)	221 (5.8)	351 (6.3)	258 (6.0)	314 (6.2)
	Jejunal; n(%)	6 (0.1)	0 (0.0)	6 (0.1)	0 (0.0)	6 (0.1)
	Total Parenteral Nutrition (TPN); n(%)	<5	<5	<5	<5	<5
2019	**Total; n**	**9847**	**3894**	**5953**	**4313**	**5534**
	Any supplemental feeding; n(%)	3504 (35.6)	1224 (31.4)	2280 (38.3)	1393 (32.3)	2111 (38.1)
	Oral; n(%)	2749 (27.9)	950 (24.4)	1799 (30.2)	1086 (25.2)	1663 (30.1)
	Nasogastric tube; n(%)	105 (1.1)	17 (0.4)	88 (1.5)	21 (0.5)	84 (1.5)
	Gastrostomy tube/Button; n(%)	552 (5.6)	211 (5.4)	341 (5.7)	242 (5.6)	310 (5.6)
	Jejunal; n(%)	-*	<5	5 (0.1)	<5	<5
	Total Parenteral Nutrition (TPN); n(%)	6 (0.1)	<5	<5	<5	<5
2024	**Total; n**	**10424**	**3688**	**6736**	**4143**	**6281**
	Any supplemental feeding; n(%)	2732 (26.2)	841 (22.8)	1891 (28.1)	934 (22.5)	1798 (28.6)
	Oral; n(%)	1550 (14.9)	476 (12.9)	1074 (15.9)	528 (12.7)	1022 (16.3)
	Nasogastric tube; n(%)	39 (0.4)	7 (0.2)	32 (0.5)	9 (0.2)	30 (0.5)
	Gastrostomy tube/Button; n(%)	315 (3.0)	115 (3.1)	200 (3.0)	132 (3.2)	183 (2.9)
	Jejunal; n(%)	-*	<5	9 (0.1)	<5	9 (0.1)
	Total Parenteral Nutrition (TPN); n(%)	5 (0.0)	<5	<5	<5	<5

1.43 Pancreatic enzyme supplementation

Year		Overall	<16 years	≥16 years	<18 years	≥18 years
2014	**Total; n**	**9432**	**3840**	**5592**	**4328**	**5104**
	Pancreatic enzyme supplements; n(%)	8027 (85.1)	3296 (85.8)	4731 (84.6)	3733 (86.3)	4294 (84.1)
2019	**Total; n**	**9847**	**3894**	**5953**	**4313**	**5534**
	Pancreatic enzyme supplements; n(%)	8140 (82.7)	3212 (82.5)	4928 (82.8)	3578 (83.0)	4562 (82.4)
2024	**Total; n**	**10424**	**3688**	**6736**	**4143**	**6281**
	Pancreatic enzyme supplements; n(%)	8329 (79.9)	2923 (79.3)	5406 (80.3)	3300 (79.7)	5029 (80.1)

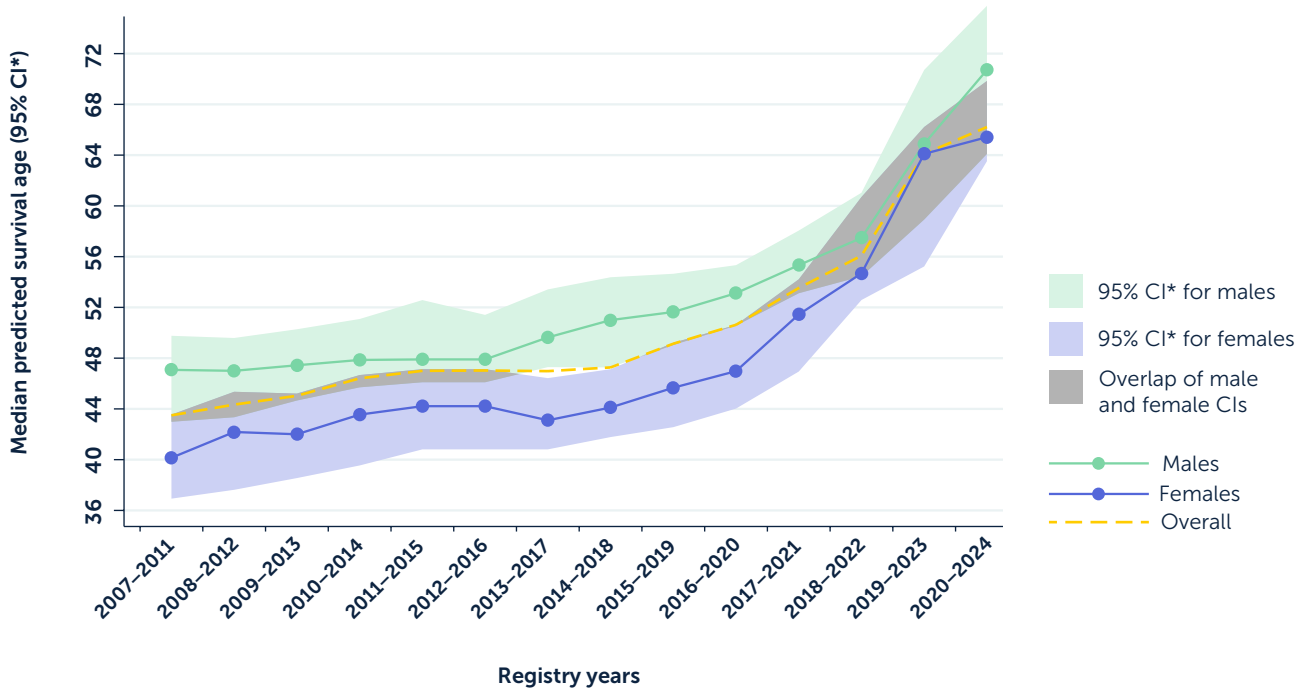
* Redacted to adhere to statistical disclosure guidelines.

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 to live. Half of people born today are predicted to live to at least **66.2** years. Half are therefore predicted to die before they reach that age. However, this does not consider individual variations in survival; for example, predicted survival is likely to be lower for those who are not eligible for CFTR modulators.

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[2] (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
2020–2024	66.2(64.1-71.2)	65.4(63.5-69.8)	70.7(64.1-75.8)	0.271

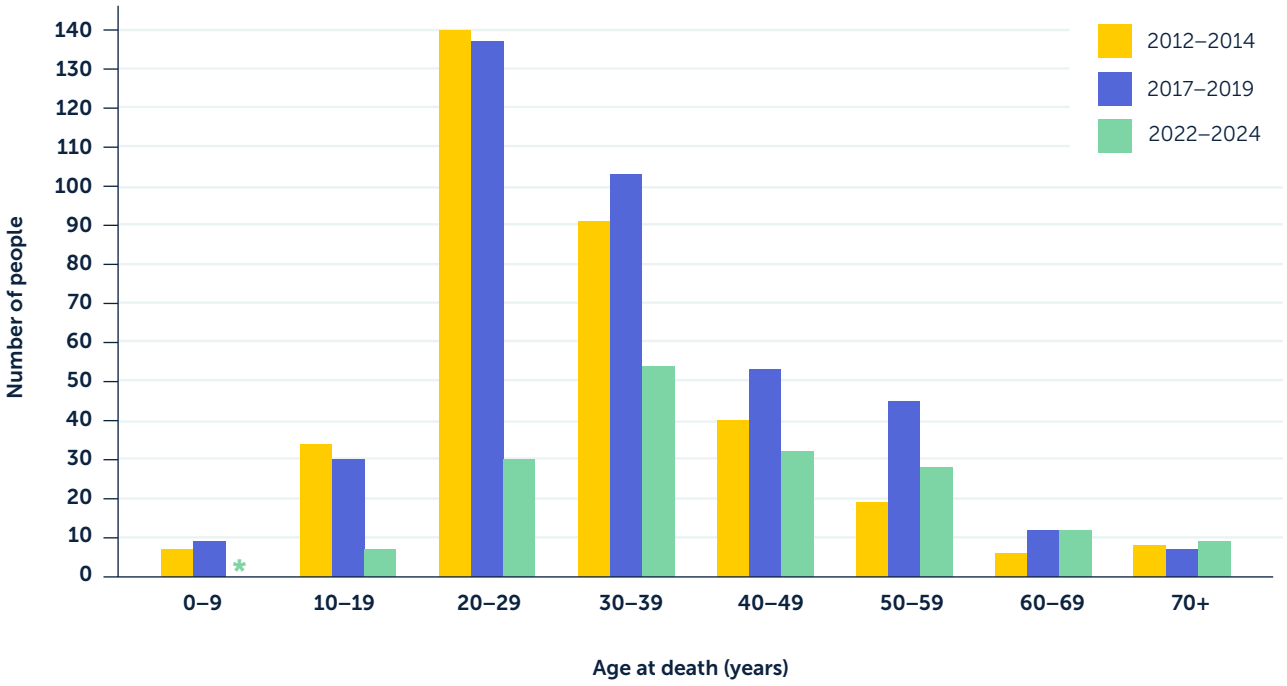
1 Sykes, Jenna et al. J Clin Epidemiol. 2016;70:206-213.

2 p-values represents the likelihood of a difference between two estimates. See glossary for more information.

* Confidence interval.

1.45 Age distribution of deaths in 2022–2024, 2017–2019, and 2012–2014

The table below shows the age distribution of people with CF who died in the last three years (2022–2024) compared to a three-year period five years ago (2017–2019) and 10 years ago (2012–2014).



Age group	2012–2014	2017–2019	2022–2024
0–9	7	9	<5
10–19	34	30	7
20–29	140	137	30
30–39	91	103	54
40–49	40	53	32
50–59	19	45	28
60–69	6	12	12
70+	8	7	9

1.46 Causes of death

This table shows all the recorded causes of death between 2022–2024.

Cause of death	2022 n(%)	2023 n(%)	2024 n(%)
Respiratory/cardiorespiratory	25 (36.8)	28 (54.9)	27 (52.9)
Other	12 (17.6)	6 (11.8)	5 (9.8)
Cancer	7 (10.3)	5 (9.8)	5 (9.8)
Liver disease/liver failure	<5	<5	<5
Transplant-related	9 (13.2)	<5	<5
Not known	8 (11.8)	<5	7 (13.7)
Trauma/suicide	<5	<5	<5
Total	68	51	51

* 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 mutations of the CF gene cause their cystic fibrosis. Everyone living with CF has two mutations (variants) of the gene for CFTR, one on each allele. One is inherited from their mother, and one from their father. If both variants 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	11302 (99.3)
Patients genotyped with both CFTR variants recorded	11035 (97.0)
F508del variants	
Homozygous F508del	5404 (47.5)
Heterozygous F508del	4732 (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	47.6							47.6
R117H	5.0	0.1						5.1
G551D	4.0	0.2	0.2					4.4
G542X	2.5	0.1	0.1	0.1				2.8
621+1G->T	1.8	0.1	0.1	0.1	0.1			2.1
Other	26.8	0.6	0.9	0.8	0.5	5.4		35.0
Unknown	1.5	0.1	0.1	0.1	0.0	0.6	0.7	3.0
Total	**89.2**	**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) with known genotypes.

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 may appear twice in the table.

These are the 20 most common 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	10136	89.1
c.350G->A	p.Arg117His	R117H	703	6.2
c.1652G->A	p.Gly551Asp	G551D	639	5.6
c.1624G->T	p.Gly542X	G542X	420	3.7
c.489+1G->T		621+1G->T	301	2.6
c.1585-1G->A		1717-1G->A	191	1.7
c.3909C->G	p.Asn1303Lys	N1303K	182	1.6
c.3454G->C	p.Asp1152His	D1152H	160	1.4
c.1766+1G->A		1898+1G->A	160	1.4
c.200C->T	p.Pro67Leu	P67L	157	1.4
c.3140-26A->G		3272-26A->G	139	1.2
c.3528delC	p.Lys1177SerfsX15	3659delC	131	1.2
c.3718-2477C->T		3849+10kbC->T	127	1.1
c.1679G->C	p.Arg560Thr	R560T	106	0.9
c.1477C->T	p.Gln493X	Q493X	98	0.9
c.1022_1023insTC	p.Phe342HisfsX28	1154insTC	96	0.8
c.1519_1521delATC	p.Ile507del	I507del	95	0.8
c.1657C->T	p.Arg553X	R553X	91	0.8
c.254G->A	p.Gly85Glu	G85E	89	0.8
c.2657+5G->A		2789+5G->A	88	0.8

1.49 CFTR variant prevalence by devolved nation

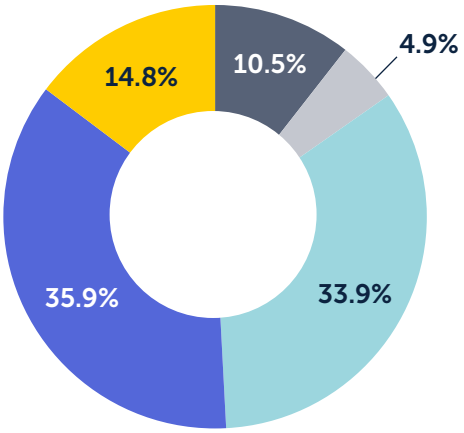
This table shows the distribution of individual variants across the devolved nations. The number of patients for each devolved nation is based on the location of the CF centre at which the patient receives care and does not account for patients who travel between devolved nations for care. The groups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

Legacy name	England n=9436		Scotland n=969		Wales n=483		Northern Ireland n=493	
	n	%	n	%	n	%	n	%
F508del	8406	89.1	880	90.8	431	89.2	419	85
R117H	541	5.7	75	7.7	19	3.9	68	13.8
G551D	485	5.1	92	9.5	18	3.7	44	8.9
G542X	308	3.3	58	6	25	5.2	29	5.9
621+1G->T	216	2.3	12	1.2	54	11.2	19	3.9
1717-1G->A	165	1.7	20	2.1	<5	-*	<5	-*
N1303K	153	1.6	13	1.3	7	1.4	9	1.8
1898+1G->A	125	1.3	5	0.5	30	6.2	0	0
D1152H	122	1.3	24	2.5	<5	-*	11	2.2
P67L	79	0.8	55	5.7	<5	-*	21	4.3

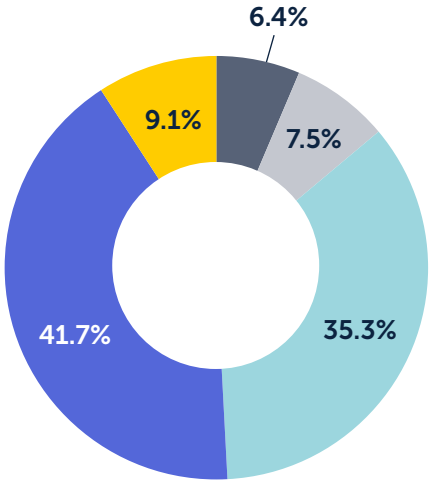
* Redacted to adhere to statistical disclosure guidelines.

1.50 Genotype prevalence by devolved nation

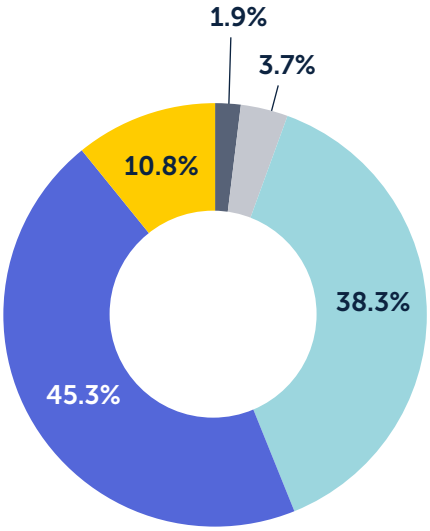
These charts show the distribution of CFTR variant combinations across the devolved nations. The number of patients for each devolved nation is based on the location of the CF centre at which the patient receives care and does not account for patients who travel between devolved nations for care.



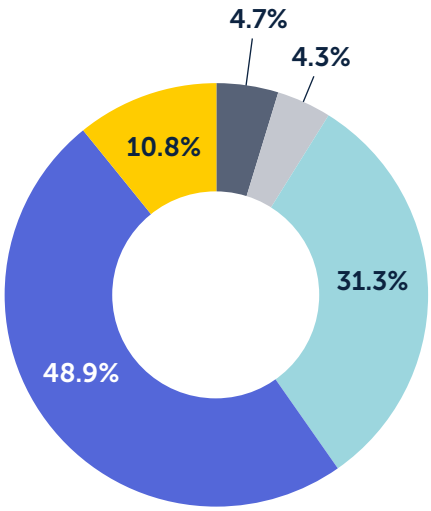
Northern Ireland
n=493



Scotland
n=969



Wales
n=483



England
n=9436

Homozygous F508del

F508del + gating (incl. G551D)

F508del + R117H

Heterozygous F508del + Other

Other

Sections 2 and 3: Centre-level analysis

Cystic fibrosis care in the UK is led by 56 regional centres, 2 standalone clinics, and 75 networked clinics. The breakdown between centres and clinics delivering paediatric and adult care is shown below:

	Paediatric	Adult	Total
Centres	29	27	56
Standalone clinics	1	1	2
Networked clinics	68	7	75

Sections 2 and 3 show 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. Data from smaller networked clinics is included in the centre data.

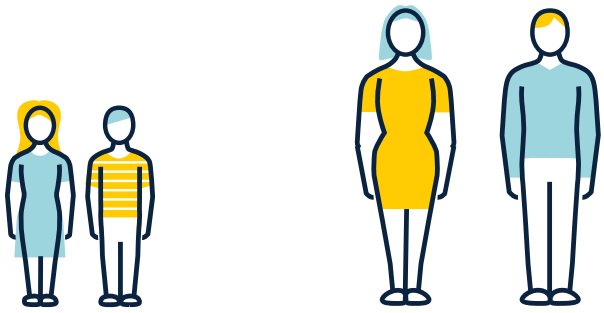
Lots of different factors can affect the outcomes of people with CF in centres, not all of which are within a centre's control. This might include the economic profile of the area, the age at which the person with CF was diagnosed and referred to the centre, and certain patient characteristics such as 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 3.

A guide on how to interpret the different graphs (funnel plots and box plots) can be found in Appendix 1.

Key to sections 2 and 3



Paediatric centre

Adult centre

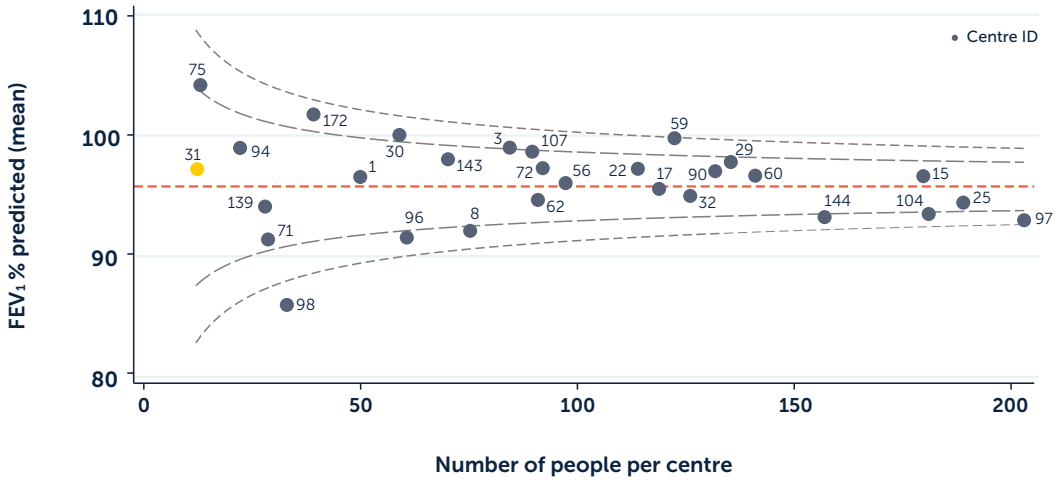
Funnel plot

- Standalone clinic
- Centre ID

Section 2: Paediatric centre analysis

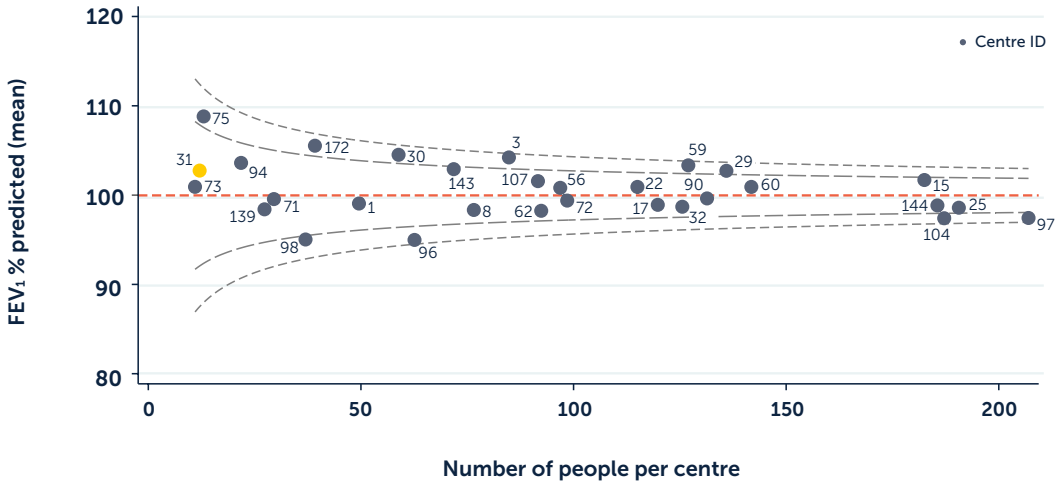
N=3995

2.1 Age-adjusted FEV_1 % predicted at annual review, in patients aged 6 and over without a history of lung transplant



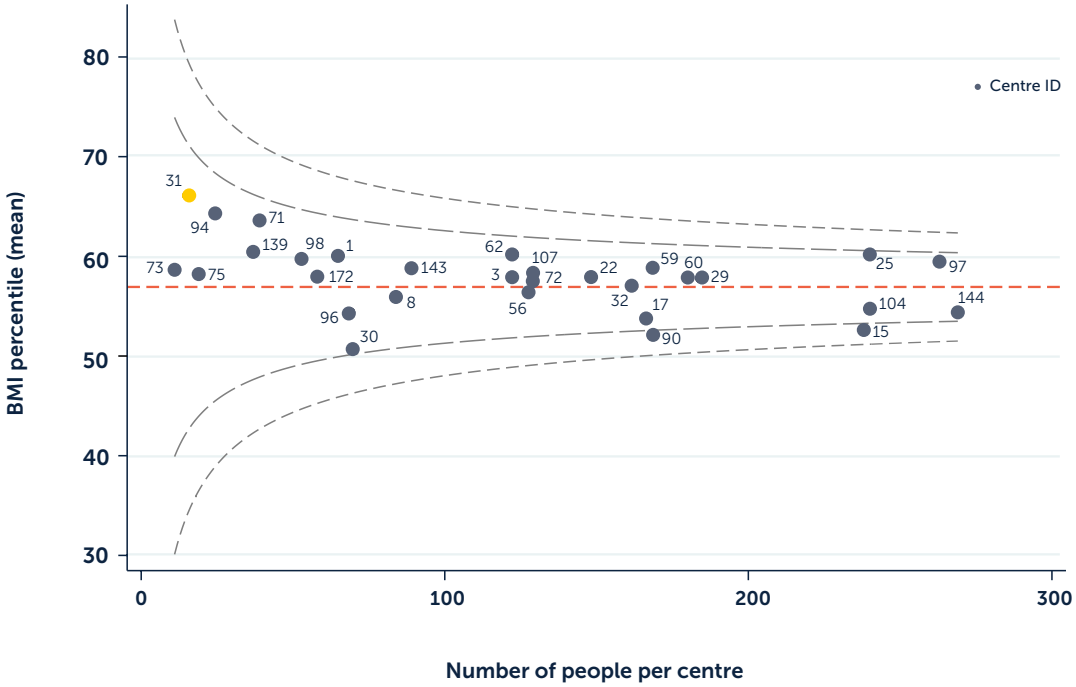
The mean FEV_1% predicted for patients attending paediatric centres/clinics is 95.7% predicted.

2.2 Age-adjusted Best FEV_1 % predicted at annual review, in patients aged 6 and over without a history of lung transplant



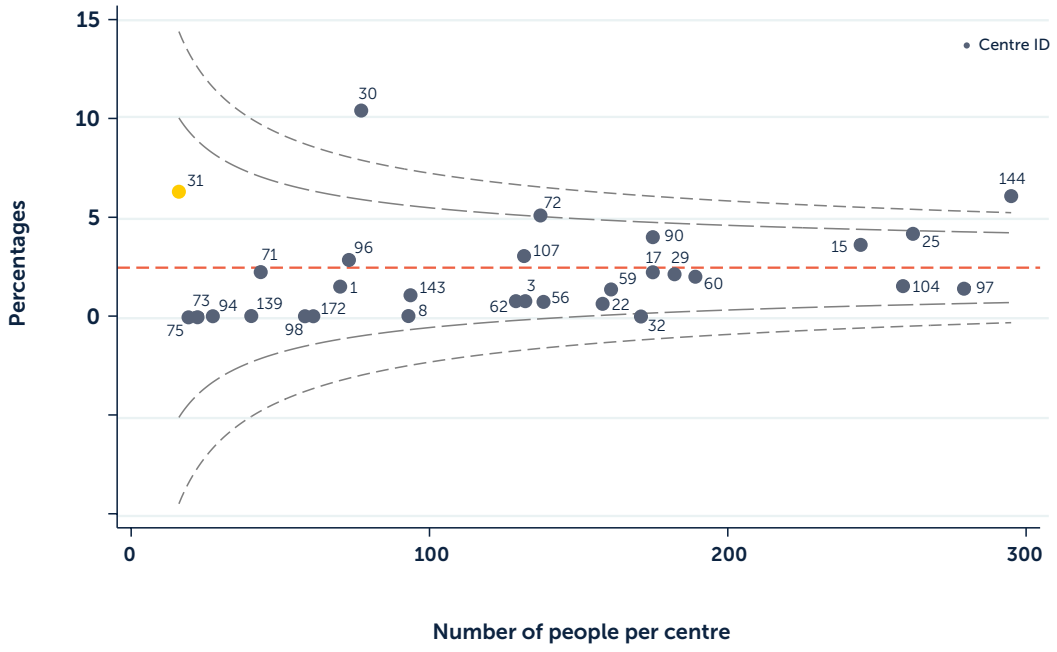
The mean Best FEV_1% predicted for patients attending paediatric centres/clinics is 100.0% predicted. Where Best FEV_1% predicted was missing, the FEV_1% predicted at annual review was used.

2.3 Age-adjusted Body Mass Index (BMI) percentile in patients aged 1–15 years



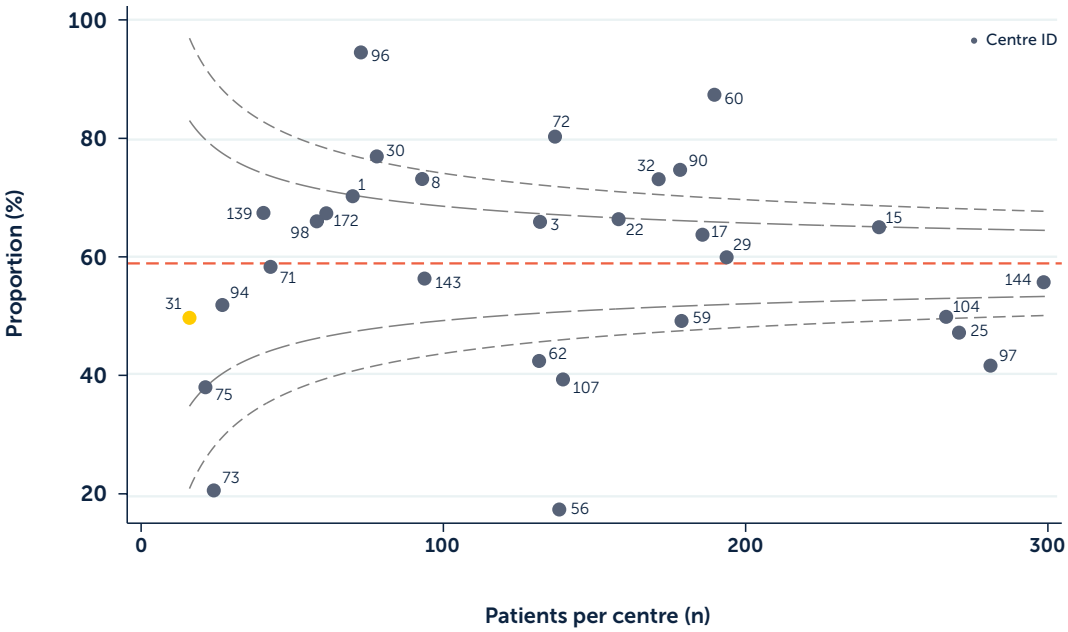
The mean BMI percentile for patients attending paediatric centres/clinics is 57.0%.

2.4 Proportion of patients with chronic *Pseudomonas aeruginosa*



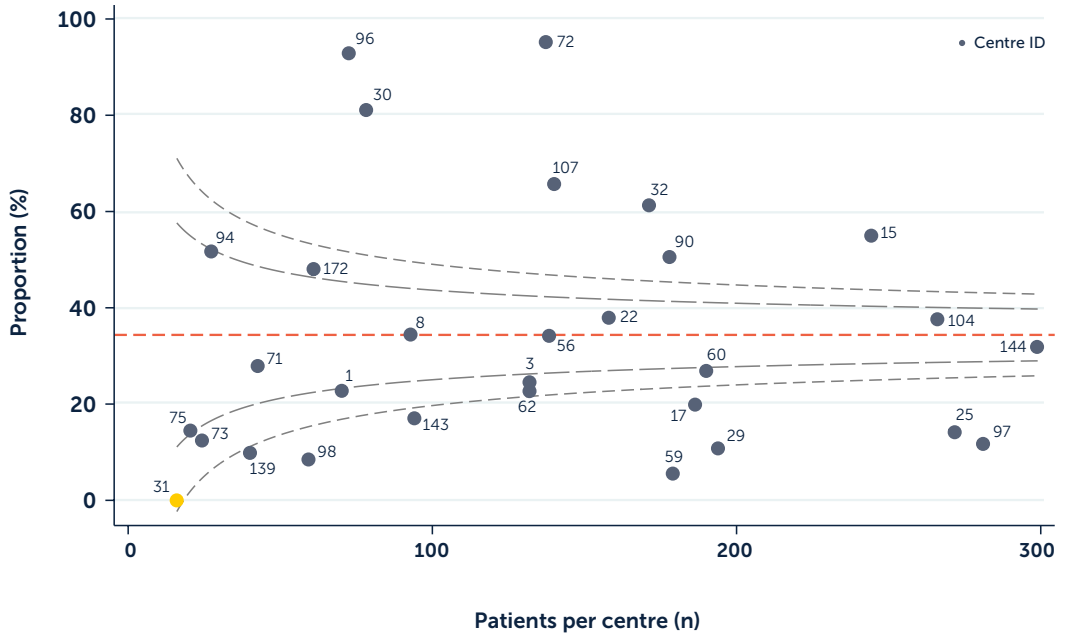
The proportion of patients with chronic *Pseudomonas aeruginosa* in paediatric centres/clinics is 2.5%.

2.5 Proportion of patients receiving DNase treatment



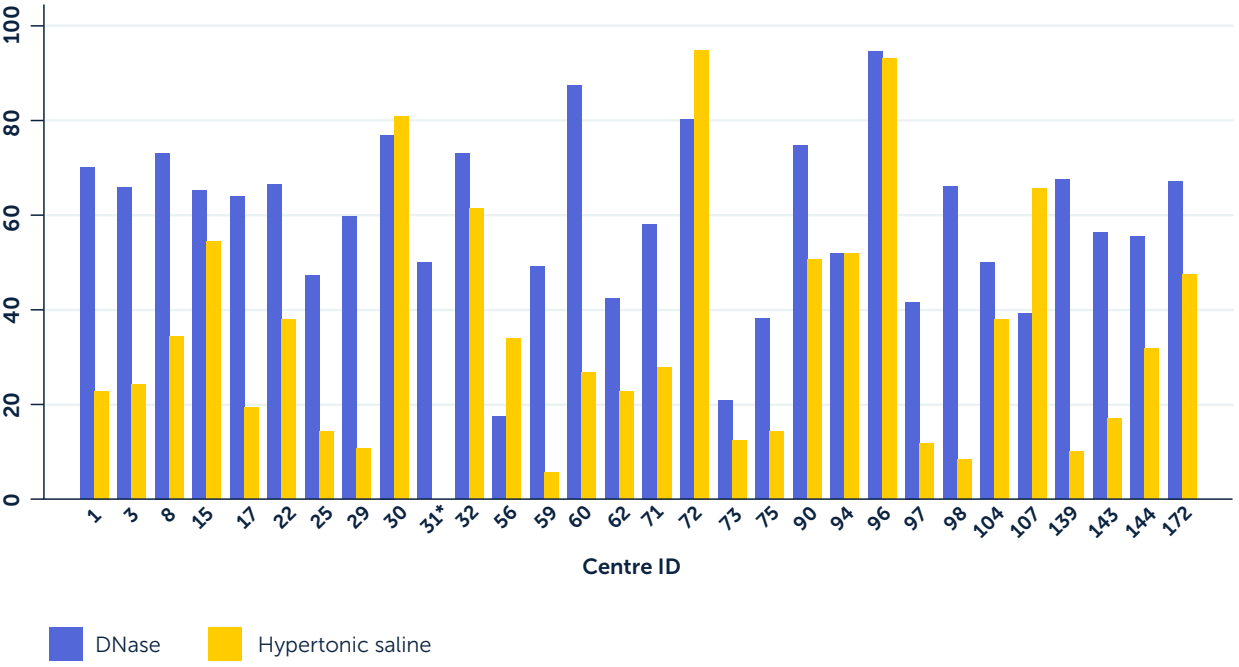
The proportion of patients receiving DNase treatment in paediatric centres/clinics is 58.9%.

2.6 Proportion of patients on hypertonic saline or mannitol treatment



The proportion of patients receiving hypertonic saline or mannitol treatment in paediatric centres/clinics is 34.4%.

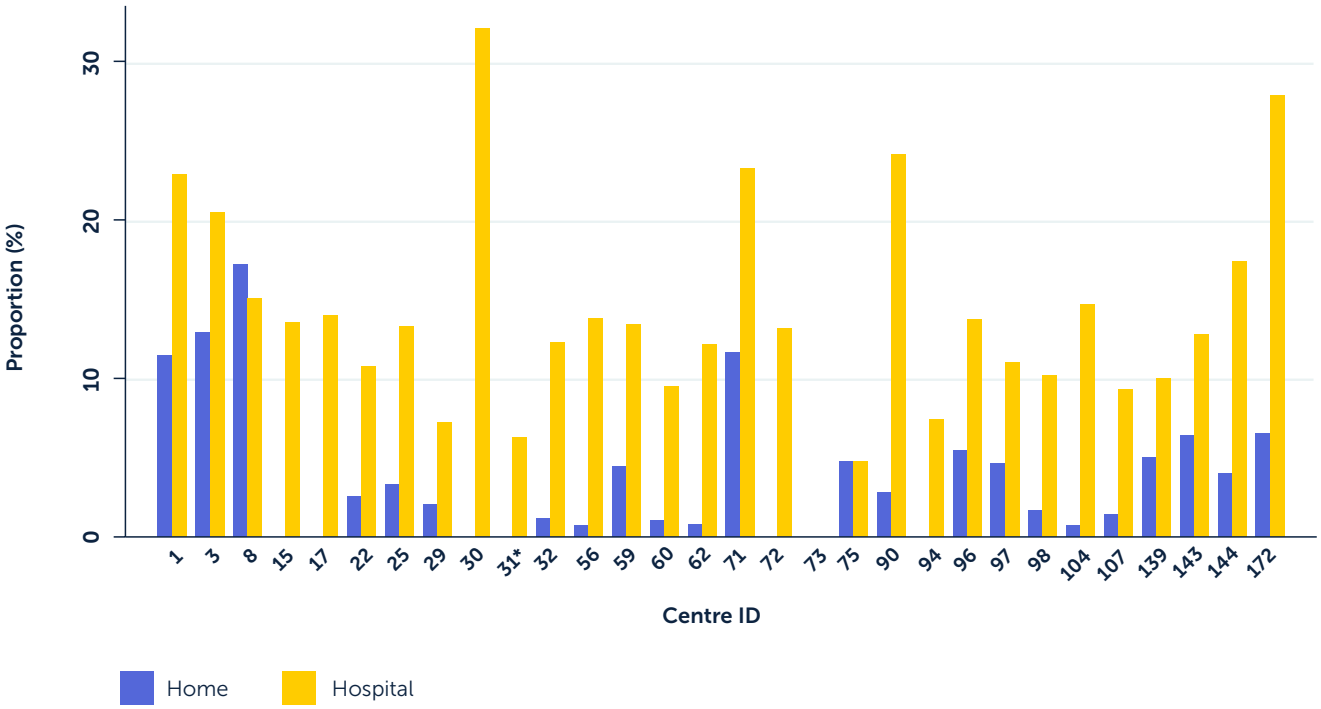
2.7 Proportion of patients receiving DNase/hypertonic saline/ mannitol treatment



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 3.2% and in hospital was 14.1%. The proportion receiving any IVs was 14.8%.

* Standalone clinics.

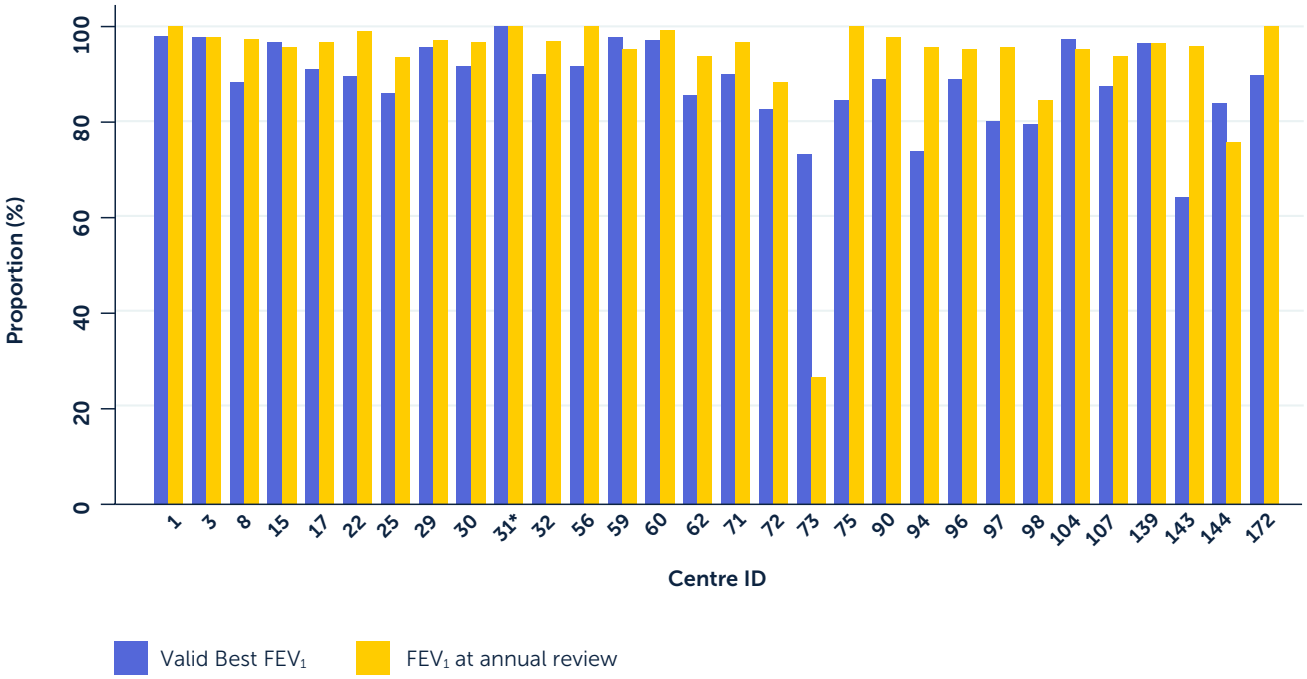
2.9 Inhaled antibiotic use for patients with chronic *Pseudomonas aeruginosa*

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

Centre ID	Proportion (%)
25	100.0
144	83.3

92.71% of patients with chronic *P. aeruginosa* received inhaled antibiotics.

2.10 Data completeness by paediatric centre/clinic[1]

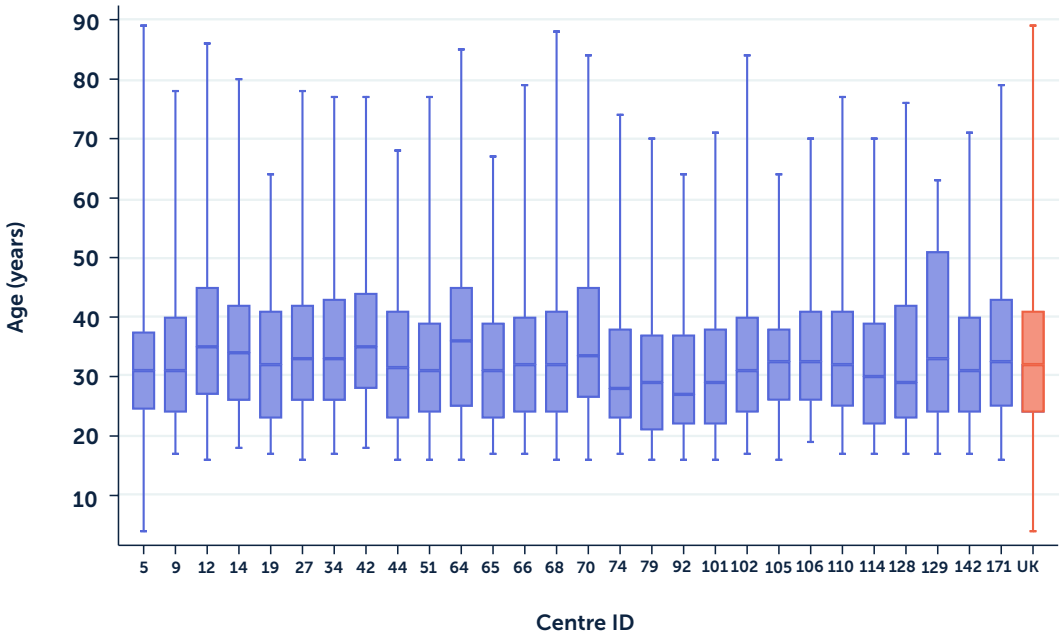


1 The chart above shows the proportion of patients who had a valid Best FEV_1% and an FEV_1% at annual review, excluding patients under six years of age. Best FEV_1% was considered valid if it was not missing, and the percent predicted was not more than 0.5% lower than the annual review value. For some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

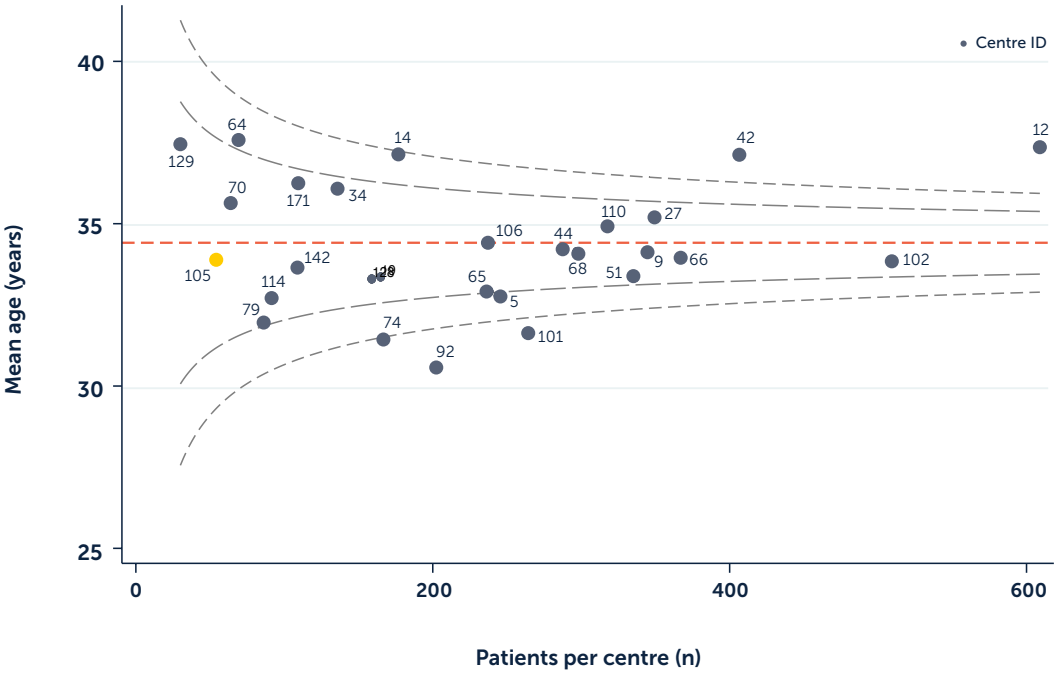
Section 3: Adult centre analysis

N=6429

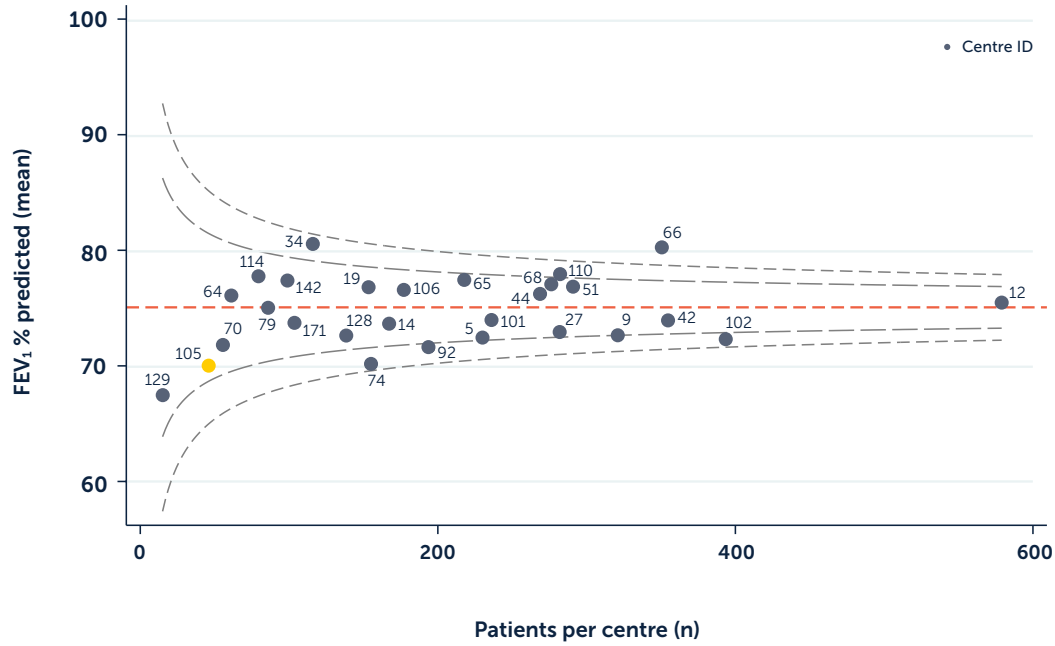
3.1 Age distribution



The funnel plot below shows how the mean age in adult centres compares to the national mean. In 2024 the national mean age of patients at CF centres was 34.4 years.

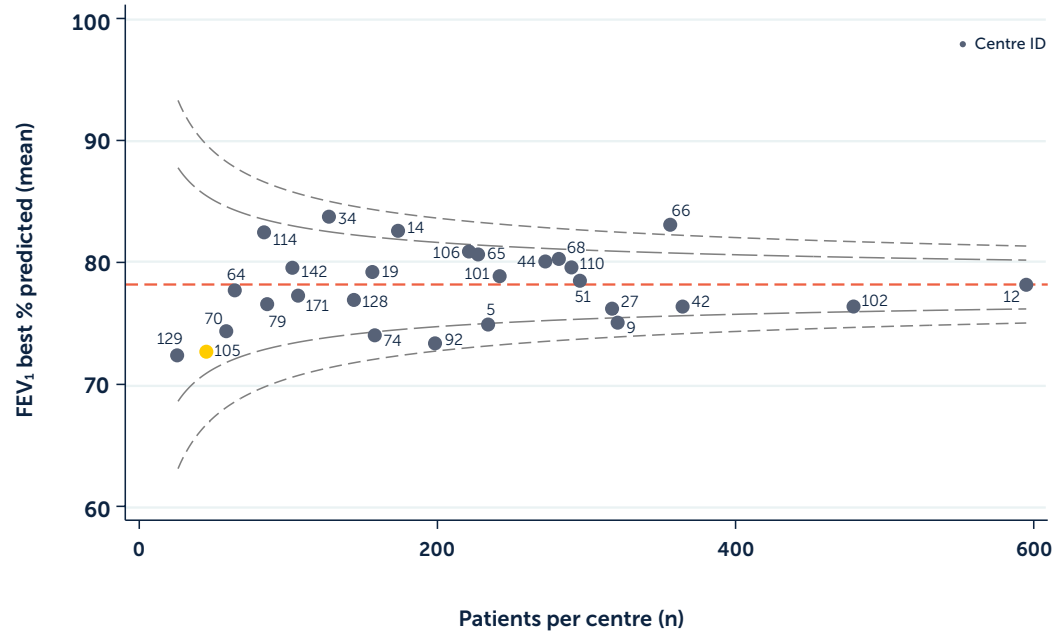


3.2 Age adjusted FEV_1 % predicted at annual review in patients without a history of lung transplant



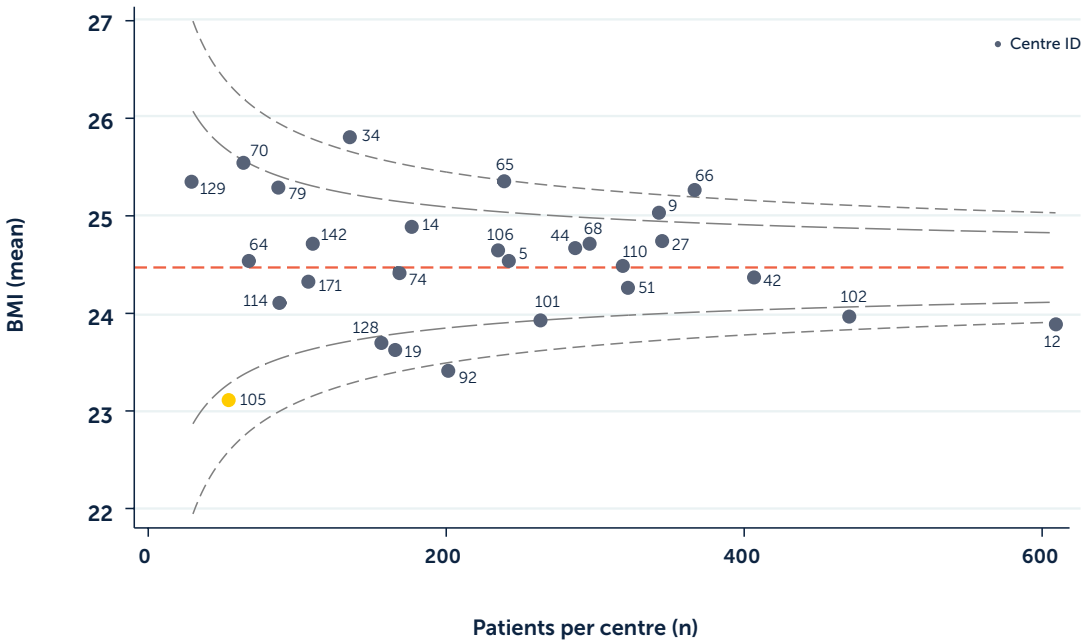
The mean FEV_1% predicted in adult centres/clinics is 75.1%.

3.3 Age adjusted Best FEV_1 % predicted at annual review in patients without a history of lung transplant



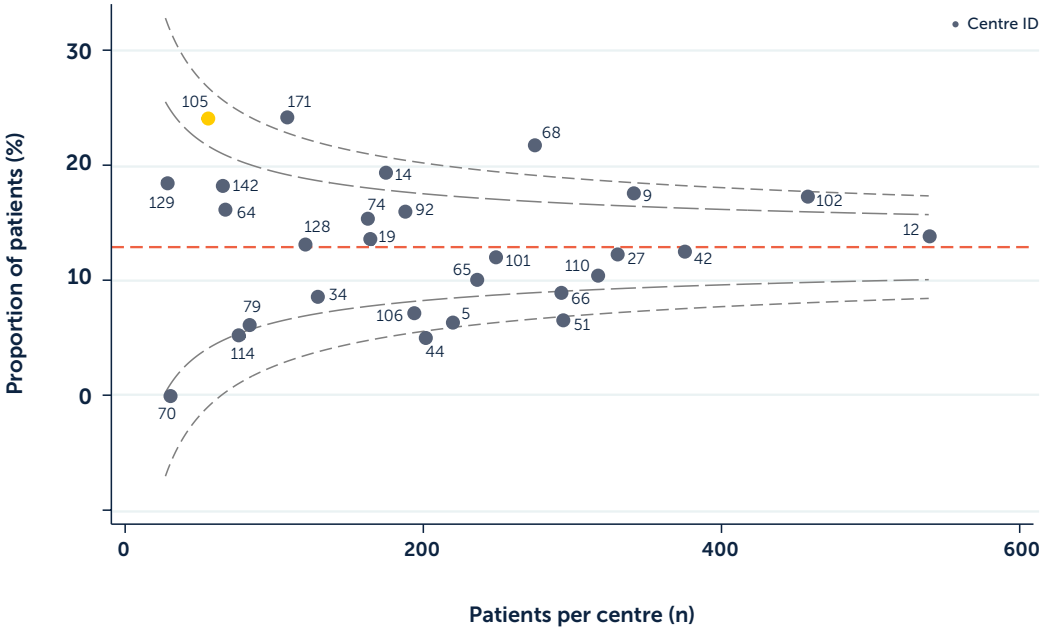
In 2024 the national mean was 78.2%. Where Best FEV_1% predicted was missing, or lower than the FEV_1 at annual review, the FEV_1% value at annual review was used.

3.4 Age-adjusted Body Mass Index (BMI) among patients aged 16 years and older



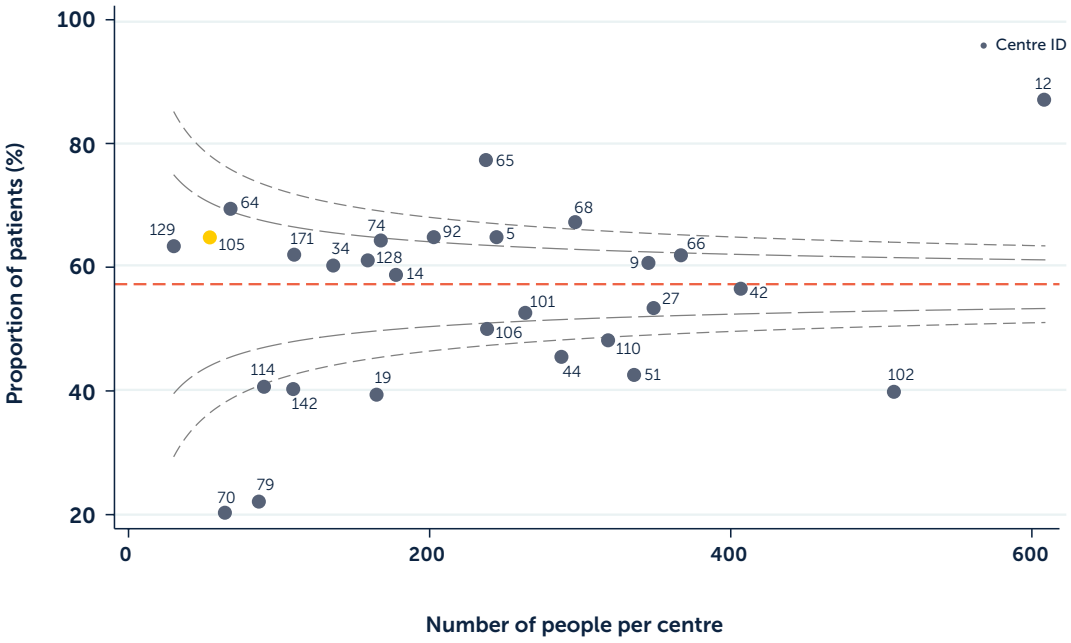
The mean BMI in adult centres/clinics is 24.5.

3.5 Proportion of patients with chronic *Pseudomonas aeruginosa*



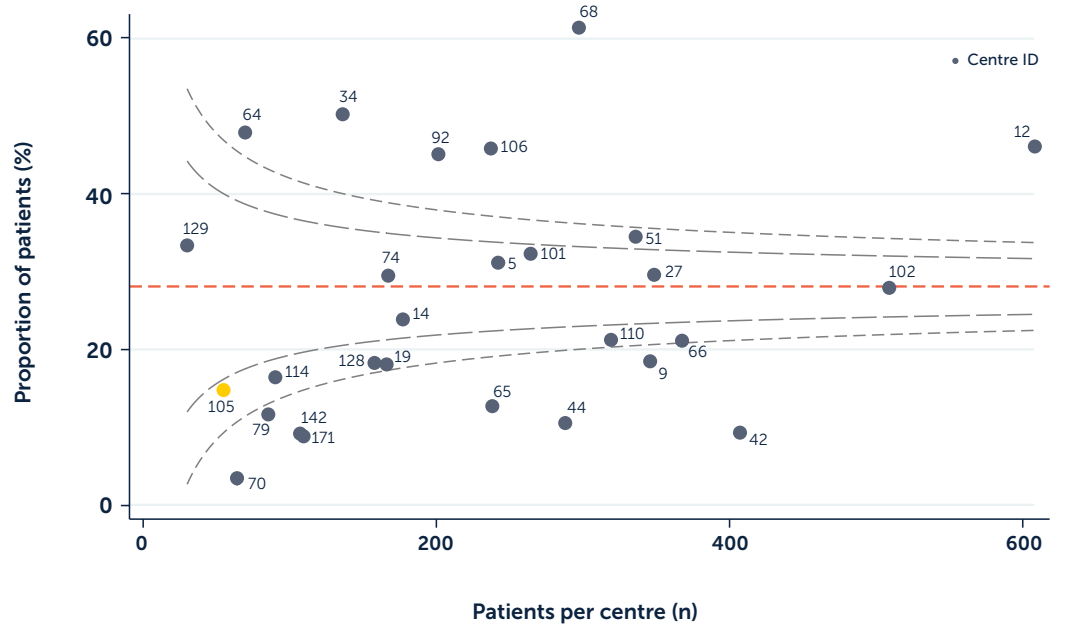
The proportion of patients with chronic *P. aeruginosa* in adult centres/clinics is 12.9%.

3.6 Proportion of patients receiving DNase treatment



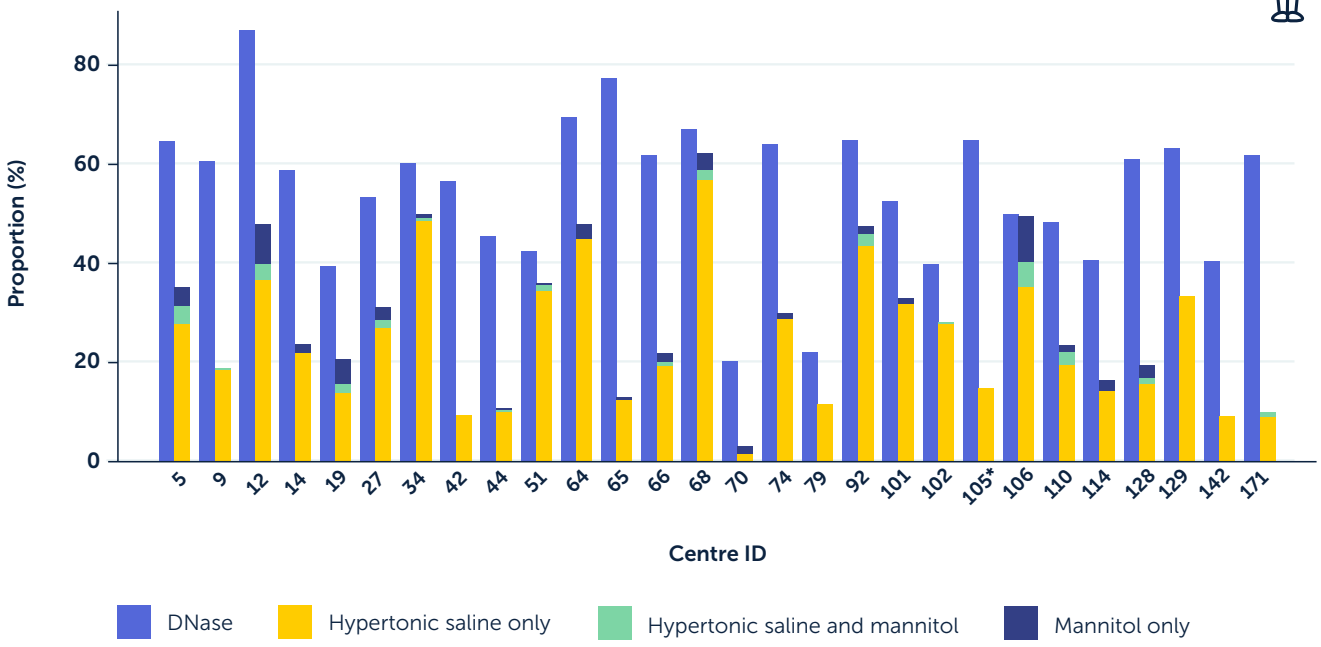
The proportion of patients receiving DNase treatment in adult centres/clinics is 57.3%.

3.7 Proportion of patients receiving hypertonic saline or mannitol



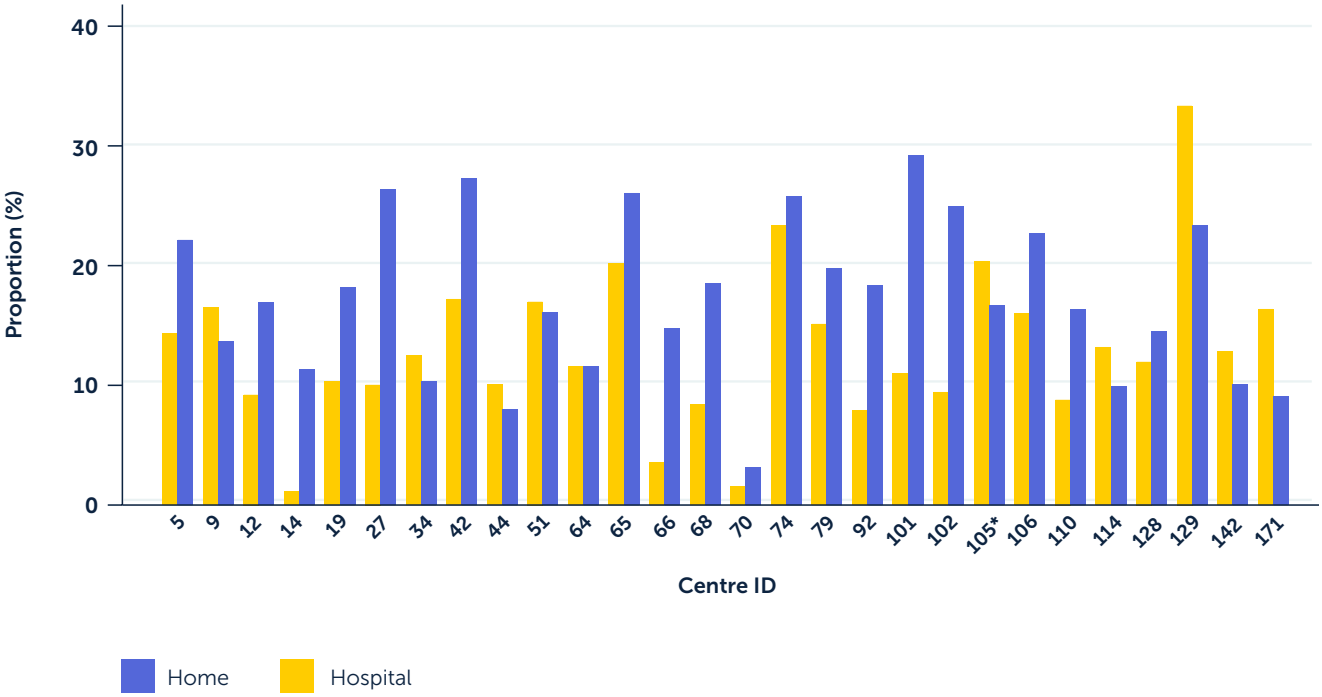
The proportion of patients receiving hypertonic saline or mannitol treatment in adult centres/clinics is 28.1%.

3.8 Proportion of patients receiving DNase/hypertonic saline/ mannitol treatment



3.9 Intravenous (IV) antibiotic use

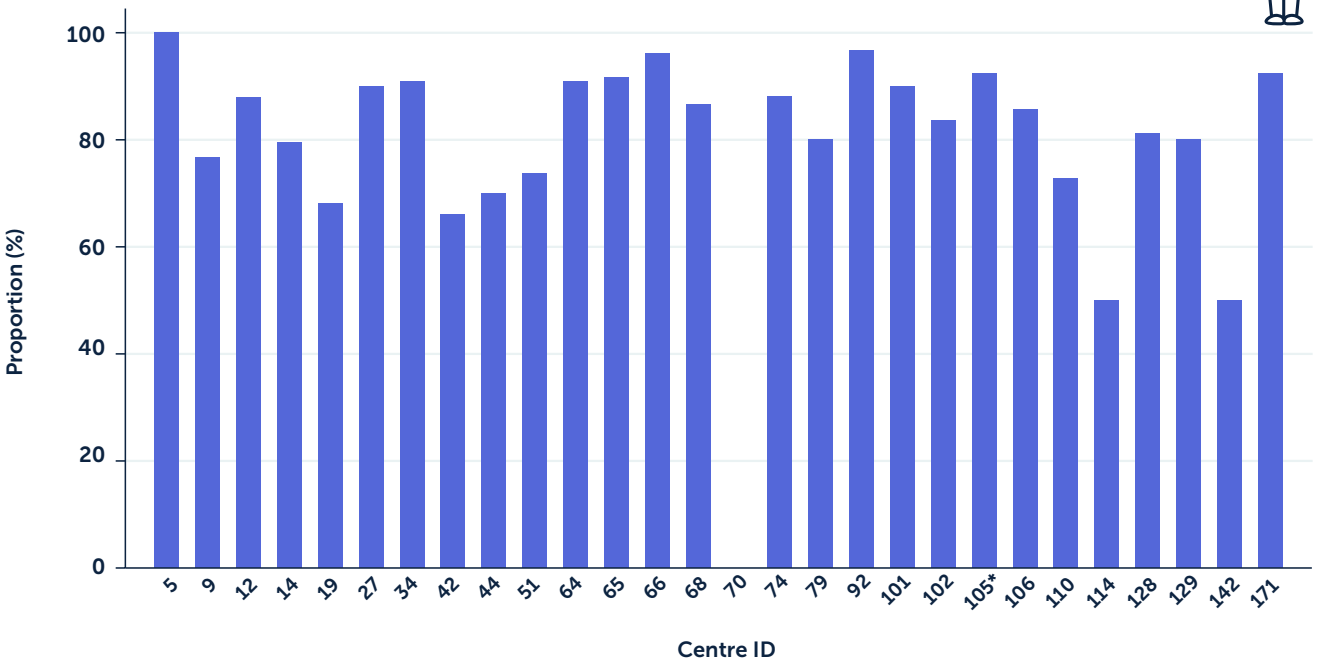
The chart below shows the proportion of patients with at least one IV day at home and/or in hospital. Patients may have a combination of home and hospital IV days.



The proportion of patients in adult centres receiving IV antibiotics at home was 11.9% and in hospital was 18.7%. The proportion receiving any IVs was 23.3%.

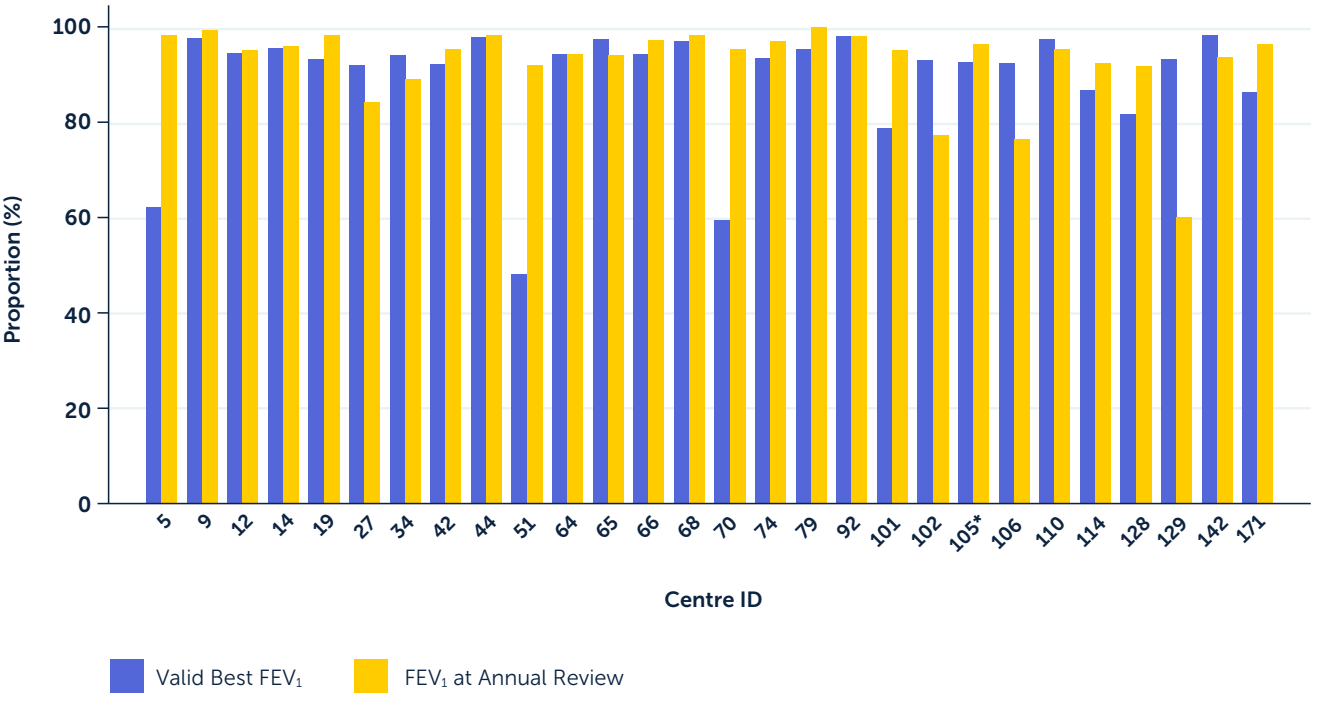
* Standalone clinics.

3.10 Inhaled antibiotic use for patients with chronic *Pseudomonas aeruginosa*



83.2% 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[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.

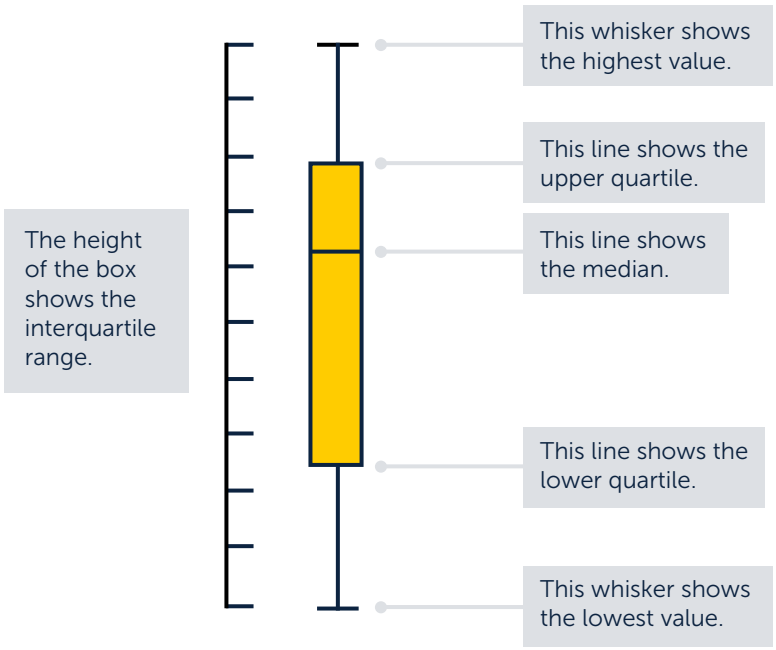
* Standalone clinics.

Appendices and Glossary

Appendix 1: 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.

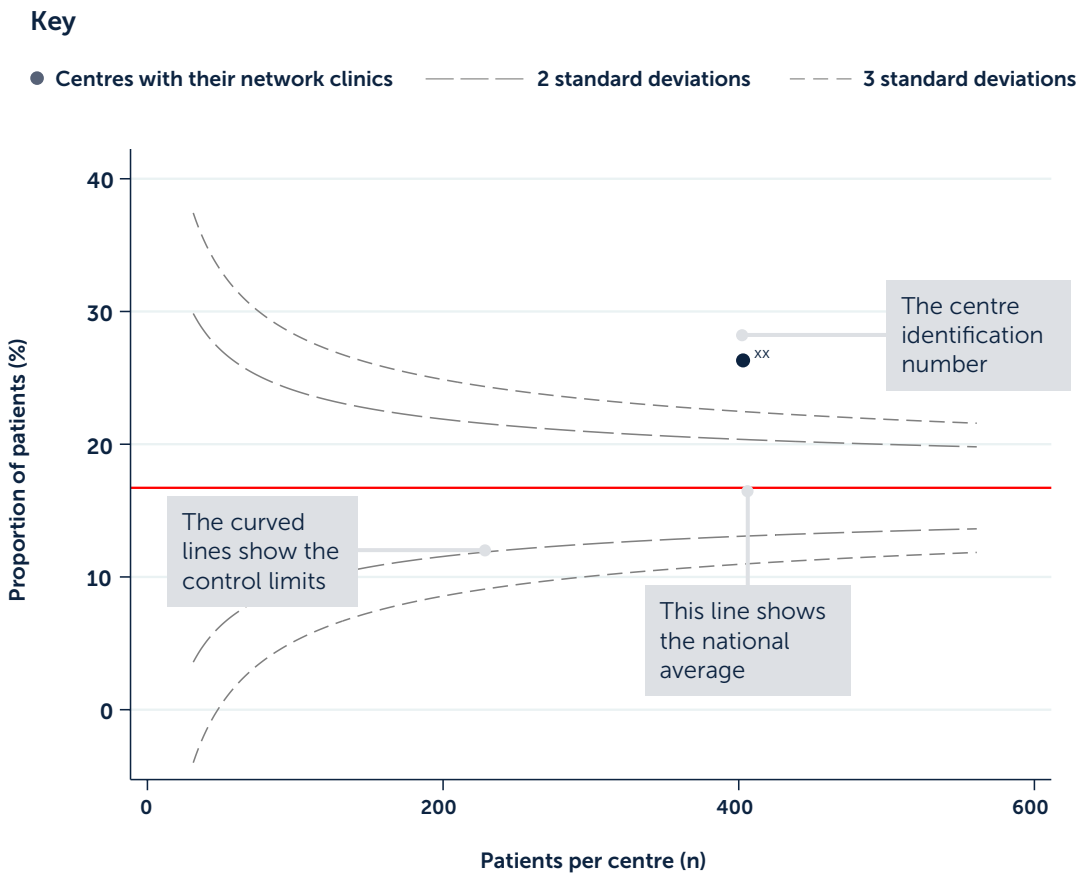
There will always be some natural variation between centres because of differences between the populations receiving care. Using only the national average as a standard can make it difficult to tell whether a survival rate that sits above the national average is higher than we would expect it to be, 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.



Appendix 2: UK CF Registry Committee structure

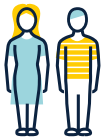
UK CF Registry Steering Committee

Role	Forename	Surname	Organisation
Director Research and Healthcare Data	Lucy	Allen	Cystic Fibrosis Trust
Commissioner, England	Kathy	Blacker	NHS England
CF Physician – Paediatrics	Malcolm	Brodlie	Great North Children's Hospital, Newcastle
CF Physician – Paediatrics	Siobhán	Carr	Royal Brompton and Harefield hospitals, London
Analytical team rep (*non-voting member*)	Susan	Charman	Cystic Fibrosis Trust
Associate Director of Data and QI, and Caldicott Guardian	Sarah	Clarke	Cystic Fibrosis Trust
CF Physician – Paediatrics	Gwyneth	Davies	UCL and Great Ormond Street Institute of Child Health, London
Chair, UK CF Registry Steering Committee and CF Physician – Adults	Jamie	Duckers	University Hospital of Wales, Cardiff
Parent of child with CF	Catherine	Farrer	N/A
Registry data manager (*non-voting member*)	Elaine	Gunn	Cystic Fibrosis Trust
Allied Health Professional	Rebecca	Heise	King's College Hospital, London
Cystic Fibrosis Centre Data Manager	Erin	Hodgetts	Royal Stoke University Hospital, Stoke
Registry Systems Development Manager	Kerry	Laidlaw	Cystic Fibrosis Trust
Welsh Commissioner	Richard	Palmer	NHS Wales
CF Physician – Adults	Simon	Range	Leicester's Hospitals, Leicester
Scotland Representative	Helen	Rodgers	NHS Scotland
Chair, UK CF Registry Research Committee, and CF Physician – Adults	Nick	Simmonds	Royal Brompton and Harefield hospitals, London
Person with CF	Hannah	Gales	N/A
Head of Registry Operations (*non-voting member*)	Mary	Kisanga	Cystic Fibrosis Trust
Head of Registry Operations (*non-voting member*)	Joanne	Osmond	Cystic Fibrosis Trust
Allied Health Professional	Jacqui	Cowlard	The Royal London Hospital, London
Allied Health Professional	Joanna	Snowball	John Radcliffe Hospital, Oxford

UK CF Registry Research Committee

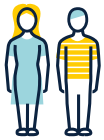
Role	Forename	Surname	Organisation
Pharmacovigilance PI, CF Physician – Paediatrics	Siobhán	Carr	Royal Brompton and Harefield hospitals, London
Analytical team rep (*non-voting member*)	Susan	Charman	Cystic Fibrosis Trust
Associate Director of Data and QI, and Caldicott Guardian	Sarah	Clarke	Cystic Fibrosis Trust
Pharmacovigilance PI, CF physician – Paediatrics	Steve	Cunningham	Royal Hospital for Children and Young People, Edinburgh
Registry data manager (*non-voting member*)	Elaine	Gunn	Cystic Fibrosis Trust
Pharmacovigilance PI, CF Physician – Adults	Dilip	Nazareth	Liverpool Heart and Chest Hospital, Liverpool
Pharmacovigilance PI , Chair of RSC, CF physician – Adults	Nick	Simmonds	Royal Brompton and Harefield hospitals, London
Person with CF	Dawn	Bostock	N/A
Pharmacovigilance PI (retired CF Physician – Adults)	Diana	Bilton	Imperial College London
Head of Registry Operations (*non-voting member*)	Mary	Kisanga	Cystic Fibrosis Trust
Head of Registry Operations (*non-voting member*)	Joanne	Osmond	Cystic Fibrosis Trust
Registry Systems Development Manager	Kerry	Laidlaw	Cystic Fibrosis Trust
Director Research and Healthcare Data (*non-voting member*)	Lucy	Allen	Cystic Fibrosis Trust
CF Physician – Paediatrics	Francis	Gilchrist	Royal Stoke University Hospital, Stoke

Appendix 3: Centre-level data tables



Paediatric centres/clinics providing data in 2024 – ordered alphabetically by country/city

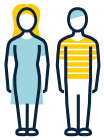
Location	Name	Clinic ID	Total Active	Number with annual review
England				
Birmingham	Birmingham Children's Hospital	104	281	266
Brighton	Royal Alexandra Children's Hospital	172	65	61
Bristol	Bristol Royal Hospital for Children	32	184	171
Cambridge	Addenbrookes Hospital	107	144	140
Cornwall	Royal Cornwall Hospital	94	35	27
Exeter	Royal Devon and Exeter Hospital	96	75	73
Leeds	St James's University Hospital	25	284	271
Leicester	Leicester Royal Infirmary	1	76	70
Liverpool	Alder Hey Children's Hospital	97	294	281
London – Central	Great Ormond Street Hospital for Children	90	188	178
London – East	The Royal London Hospital	30	80	78
London – South East	King's College Hospital	17	200	186
London – South West	Royal Brompton Hospital	15	261	244
Manchester	Royal Manchester Children's Hospital	144	320	299
Newcastle	Great North Children's Hospital	59	201	179
North West Midlands	University Hospital of North Midlands	8	96	93
Norwich	Norfolk and Norwich University Hospital	98	64	59
Nottingham	Nottingham University Hospitals	62	142	132
Oxford	John Radcliffe Hospital	22	168	158
Plymouth	Derriford Hospital	139	41	40
Sheffield	Sheffield Children's Hospital	3	139	132
Southampton	Southampton General Hospital	29	221	194
Teesside	The James Cook University Hospital	71	52	43
Northern Ireland				
Belfast	Royal Belfast Hospital for Sick Children	60	210	190
Scotland				
Aberdeen	Royal Aberdeen Children's Hospital	75	30	21
Dundee	Ninewells Hospital	73	25	24
Edinburgh	Royal Hospital for Children and Young People	143	114	94
Glasgow	Royal Hospital for Children and Young People	56	165	138
Inverness	Raigmore Hospital	31	16	16
Wales				
Cardiff	Noah's Ark Children's Hospital for Wales	72	146	137



	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	9.0	8.8	181	93.7	93.4	95.2	188	97.9	97.5	98.6
172	8.8	9.0	39	101.6	101.7	100.8	39	105.5	105.6	104.3
32	10.1	10.2	126	94.8	95.1	95.9	126	98.5	98.8	99.2
107	8.9	9.1	90	98.6	98.5	100.4	92	101.7	101.4	102.5
94	10.6	11.2	22	98.8	98.8	95.9	22	103.4	103.4	99.9
96	10.4	10.5	61	91.7	91.5	90.5	63	95.1	94.9	95.8
25	9.5	10.0	189	94.1	94.1	97.3	191	98.3	98.3	100.1
1	9.4	9.2	50	96.1	96.2	96.0	50	99.4	99.4	99.1
97	9.7	9.7	203	92.9	92.9	94.6	207	97.8	97.7	99.2
90	9.7	10.0	132	97.1	97.0	99.0	132	99.7	99.6	100.9
30	10.1	10.4	59	99.5	99.7	102.2	59	104.0	104.3	105.7
17	8.6	8.9	119	95.8	95.5	97.3	120	99.5	99.1	100.6
15	9.6	10.1	180	96.5	96.3	96.5	183	102.1	101.8	101.6
144	9.3	9.9	157	93.0	93.0	94.7	186	98.4	98.5	99.8
59	9.3	9.9	123	100.0	99.9	100.2	127	103.4	103.2	102.7
8	10.8	12.0	75	91.5	92.0	94.3	77	97.8	98.3	98.4
98	8.6	8.0	33	85.8	85.7	88.6	37	95.4	95.2	93.5
62	9.6	10.1	91	94.3	94.6	94.4	93	97.8	98.1	98.9
22	9.7	10.0	114	97.0	97.2	96.8	115	100.4	100.5	99.7
139	9.3	9.5	28	94.0	93.9	97.0	28	98.9	98.6	100.6
3	8.2	9.0	84	99.5	99.0	100.1	85	104.8	104.1	103.4
29	9.3	9.2	135	97.7	97.7	98.1	136	102.6	102.6	104.1
71	9.4	10.0	29	90.6	91.0	93.2	29	98.9	99.3	98.3
60	9.9	9.9	141	96.4	96.7	96.5	142	100.4	100.7	101.3
75	7.8	7.0	13	105.1	104.1	103.7	13	110.0	108.7	106.9
73	8.4	9.1	<5	97.3	98.3	96.5	11	100.8	100.9	99.7
143	9.7	10.3	70	98.0	97.9	97.0	72	103.2	102.9	101.8
56	9.3	9.5	97	95.9	95.9	95.6	97	100.5	100.4	100.3
31	9.2	9.0	12	97.5	97.1	98.4	12	103.0	102.5	104.7
72	9.9	9.8	92	97.1	97.2	97.0	98	99.8	99.8	99.8

* Number with non-missing data.

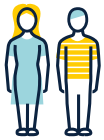
** Where 'Best' values were missing, or lower than FEV_1% predicted taken at annual review, the annual review value was used.



Paediatric centres/clinics providing data in 2024 – ordered alphabetically by country/city

			BMI			
Location	Name	Clinic ID	Number*	Mean – unadjusted	Mean – adjusted	Median
England						
Birmingham	Birmingham Children's Hospital	104	240	54.6	54.6	53.7
Brighton	Royal Alexandra Children's Hospital	172	58	58.0	58.0	54.5
Bristol	Bristol Royal Hospital for Children	32	162	57.0	57.1	61.3
Cambridge	Addenbrookes Hospital	107	129	58.0	58.0	60.2
Cornwall	Royal Cornwall Hospital	94	25	64.2	64.3	66.6
Exeter	Royal Devon & Exeter Hospital	96	69	54.3	54.4	54.6
Leeds	St James's University Hospital	25	240	60.2	60.2	65.4
Leicester	Leicester Royal Infirmary	1	65	60.1	60.1	69.9
Liverpool	Alder Hey Children's Hospital	97	263	59.4	59.5	60.7
London – Central	Great Ormond Street Hospital for Children	90	169	52.2	52.2	53.1
London – East	Royal London Hospital	30	70	50.5	50.6	49.8
London – South East	King's College Hospital	17	167	53.8	53.8	55.8
London – South West	Royal Brompton Hospital	15	238	52.7	52.7	52.2
Manchester	Royal Manchester Children's Hospital	144	269	54.4	54.4	52.3
Newcastle	Great North Children's Hospital	59	169	58.9	58.9	61.4
North West Midlands	University Hospital of North Midlands	8	84	55.8	55.9	56.9
Norwich	Norfolk & Norwich University Hospital	98	53	59.9	59.9	63.0
Nottingham	Nottingham University Hospitals	62	123	60.2	60.2	63.7
Oxford	John Radcliffe Hospital	22	148	57.8	57.9	61.7
Plymouth	Derriford Hospital	139	37	60.5	60.4	65.5
Sheffield	Sheffield Children's Hospital	3	122	57.9	57.8	58.0
Southampton	Southampton General Hospital	29	184	57.8	57.8	58.6
Teesside	James Cook University Hospital	71	39	63.6	63.6	65.0
Northern Ireland						
Belfast	Royal Belfast Hospital for Sick Children	60	180	57.7	57.7	59.3
Scotland						
Aberdeen	Royal Aberdeen Children's Hospital	75	19	58.4	58.2	61.7
Dundee	Ninewells Hospital	73	11	58.6	58.6	67.4
Edinburgh	Royal Hospital for Sick Children	143	89	58.9	58.9	56.3
Glasgow	Royal Hospital for Sick Children	56	128	56.6	56.6	59.8
Inverness	Raigmore Hospital	31	15	66.1	66.1	75.2
Wales						
Cardiff	Children's Hospital for Wales	72	129	57.6	57.6	59.8

* Number with non-missing data.



	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	<5	1.5	39	14.7	133	50.0	101	38.0	<5	100.0
172	0	0.0	17	27.9	41	67.2	29	47.5	0	0.0
32	0	0.0	21	12.3	125	73.1	105	61.4	0	0.0
107	<5	3.0	14	10.0	55	39.3	92	65.7	<5	75.0
94	0	0.0	<5	7.4	14	51.9	14	51.9	0	0.0
96	<5	2.7	10	13.7	69	94.5	68	93.2	<5	100.0
25	11	4.2	38	14.0	128	47.2	39	14.4	11	100.0
1	<5	1.4	16	22.9	49	70.0	16	22.9	<5	100.0
97	<5	1.4	36	12.8	117	41.6	33	11.7	<5	100.0
90	7	4.0	43	24.2	133	74.7	90	50.6	7	100.0
30	8	10.4	25	32.1	60	76.9	63	80.8	7	87.5
17	<5	2.3	26	14.0	119	64.0	37	19.9	<5	100.0
15	9	3.7	33	13.5	159	65.2	135	55.3	9	100.0
144	18	6.1	54	18.1	166	55.5	95	31.8	15	83.3
59	<5	1.2	28	15.6	88	49.2	10	5.6	<5	100.0
8	0	0.0	22	23.7	68	73.1	32	34.4	0	0.0
98	0	0.0	6	10.2	39	66.1	5	8.5	0	0.0
62	<5	0.8	17	12.9	56	42.4	30	22.7	<5	100.0
22	<5	0.6	18	11.4	105	66.5	60	38.0	<5	100.0
139	0	0.0	<5	10.0	27	67.5	<5	10.0	0	0.0
3	<5	0.8	27	20.5	87	65.9	32	24.2	<5	100.0
29	<5	2.2	14	7.2	116	59.8	21	10.8	<5	75.0
71	<5	2.3	11	25.6	25	58.1	12	27.9	0	0.0
60	<5	2.1	18	9.5	166	87.4	51	26.8	<5	100.0
75	0	0.0	<5	4.8	8	38.1	<5	14.3	0	0.0
73	0	0.0	0	0.0	5	20.8	<5	12.5	0	0.0
143	<5	1.1	12	12.8	53	56.4	16	17.0	<5	100.0
56	<5	0.7	19	13.8	24	17.4	47	34.1	<5	100.0
31	<5	6.2	<5	6.2	8	50.0	0	0.0	<5	100.0
72	7	5.1	18	13.1	110	80.3	130	94.9	7	100.0

* Redacted to adhere to statistical disclosure guidelines.

Adult centres/clinics providing data in 2024 – ordered alphabetically by country/city

Location	Name	Clinic ID	Total Active	Number with annual review
England				
Birmingham	Heartlands Hospital	27	356	349
Bristol	Bristol Royal Infirmary	106	258	238
Cambridge	Royal Papworth Hospital	51	387	336
Cornwall	Royal Cornwall Hospital	129	44	30
Exeter	Royal Devon and Exeter Hospital	34	142	136
Frimley	Frimley Park Hospital	19	172	165
Leeds	St James's University Hospital	42	422	407
Leicester	Glenfield Hospital	142	116	109
Liverpool	Liverpool Heart and Chest Hospital	66	402	367
London – East	St Bartholomew's Hospital	92	240	202
London – South East	University Hospital Lewisham	105	59	54
London – South East	King's College Hospital	5	276	244
London – South West	Royal Brompton Hospital	12	616	609
Manchester	Wythenshawe Hospital	102	520	509
Newcastle	Royal Victoria Infirmary	9	356	345
North West Midlands	University Hospital of North Midlands	74	171	167
Norwich	Norfolk and Norwich University Hospital	114	92	91
Nottingham	Nottingham University Hospitals	101	269	264
Oxford	Oxford University Hospitals	128	168	159
Plymouth	Derriford Hospital	64	71	69
Sheffield	Northern General Hospital	65	245	238
Southampton	Southampton General Hospital	110	331	319
York and Hull	York Hospital	171	113	110
Northern Ireland				
Belfast	Belfast City Hospital	14	283	177
Scotland				
Aberdeen	Aberdeen Royal Infirmary	70	78	64
Edinburgh	Western General Hospital	44	298	288
Glasgow	Queen Elizabeth University Hospital	79	243	86
Wales				
Llandough	University Hospital Llandough	68	337	297

	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	35.2	33.1	282	72.8	72.9	76.3	318	75.8	76.0	78.8
106	34.4	32.5	177	76.0	76.6	79.6	223	80.3	80.9	84.4
51	33.3	31.1	291	77.7	76.8	78.1	295	79.5	78.5	81.2
129	37.5	33.3	15	66.1	67.5	65.3	26	71.6	72.4	72.1
34	36.1	33.9	116	79.5	80.6	81.9	128	82.8	83.8	87.9
19	33.4	32.4	154	77.3	76.8	79.7	157	79.8	79.3	82.1
42	37.1	35.4	354	72.3	73.9	75.7	364	74.6	76.2	78.9
142	33.6	31.6	98	78.2	77.3	83.2	103	80.3	79.6	87.4
66	34.0	32.8	350	80.8	80.3	84.0	357	83.6	83.1	86.6
92	30.6	27.3	195	73.9	71.5	75.5	198	75.7	73.2	77.8
105	33.9	33.1	45	69.9	70.0	67.1	45	72.6	72.7	76.2
5	32.8	31.5	231	73.1	72.4	78.2	233	75.5	74.9	80.7
12	37.4	35.1	579	73.7	75.5	75.6	595	76.5	78.2	77.6
102	33.8	31.9	393	72.3	72.3	74.3	480	76.4	76.3	77.3
9	34.1	31.9	321	73.3	72.7	77.0	321	75.7	75.0	80.3
74	31.5	28.5	156	72.5	70.2	77.8	158	76.5	74.0	81.8
114	32.7	30.9	80	78.7	77.8	83.6	85	83.5	82.5	86.2
101	31.6	29.8	237	75.8	74.0	79.2	241	80.8	78.8	82.8
128	33.3	29.8	138	74.3	72.6	75.4	144	78.7	77.0	78.2
64	37.6	36.6	62	75.8	76.1	79.9	65	77.4	77.8	82.0
65	32.9	31.5	218	78.3	77.4	82.6	226	81.7	80.7	85.5
110	34.9	32.1	282	77.9	77.8	79.8	290	79.8	79.4	82.1
171	36.3	33.0	104	73.2	73.7	74.5	106	76.9	77.3	79.6
14	37.2	34.2	167	72.4	73.7	76.9	174	81.4	82.5	80.8
70	35.6	33.5	56	71.8	71.8	72.9	58	74.3	74.4	75.1
44	34.2	32.0	270	77.1	76.3	80.8	273	80.9	80.1	84.2
79	32.0	29.9	86	76.9	75.0	77.6	86	78.6	76.6	80.2
68	34.1	32.3	277	77.6	77.1	81.4	280	80.8	80.3	83.9

* Number with non-missing data.

** 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 2024 – ordered alphabetically by country/city

			BMI			
Location	Name	Clinic ID	Number*	Mean - unadjusted	Mean - adjusted	Median
England						
Birmingham	Birmingham Heartlands Hospital	27	345	24.8	24.7	24.3
Bristol	Bristol Royal Infirmary	106	236	24.7	24.6	24.0
Cambridge	Royal Papworth Hospital	51	322	24.2	24.3	23.5
Cornwall	Royal Cornwall Hospital	129	30	25.5	25.3	23.5
Exeter	Royal Devon and Exeter Hospital	34	136	26.0	25.8	24.7
Frimley	Frimley Park Hospital	19	165	23.6	23.6	23.3
Leeds	St James's University Hospital	42	407	24.7	24.4	24.0
Leicester	Glenfield Hospital	142	109	24.7	24.7	24.2
Liverpool	Liverpool Heart and Chest Hospital	66	367	25.2	25.2	24.4
London – East	St Bartholomew's Hospital	92	202	23.1	23.4	22.5
London – South East	University Hospital Lewisham	105	54	23.2	23.1	22.5
London – South East	King's College Hospital	5	242	24.4	24.5	23.2
London – South West	Royal Brompton Hospital	12	609	24.2	23.9	23.6
Manchester	Wythenshawe Hospital	102	471	23.9	24.0	23.3
Newcastle	Royal Victoria Infirmary	9	342	25.0	25.0	24.0
North West Midlands	University Hospital of North Midlands	74	167	24.2	24.4	23.7
Norwich	Norfolk and Norwich University Hospital	114	90	24.0	24.1	23.2
Nottingham	Nottingham University Hospitals	101	263	23.7	23.9	22.8
Oxford	Oxford University Hospitals	128	157	23.5	23.7	22.5
Plymouth	Derriford Hospital	64	69	24.7	24.5	24.0
Sheffield	Northern General Hospital	65	237	25.2	25.4	24.5
Southampton	Southampton General Hospital	110	319	24.5	24.5	23.4
York and Hull	York Hospital	171	109	24.4	24.3	24.0
Northern Ireland						
Belfast	Belfast City Hospital	14	177	25.1	24.9	24.5
Scotland						
Aberdeen	Aberdeen Royal Infirmary	70	64	25.6	25.5	24.7
Edinburgh	Western General Hospital	44	287	24.6	24.7	23.8
Glasgow	Queen Elizabeth University Hospital	79	86	25.0	25.3	24.1
Wales						
Llandough	University Hospital Llandough	68	296	24.7	24.7	23.9

* Number with non-missing data.

	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	40	12.1	101	28.9	186	53.3	103	29.5	36	90.0
106	14	7.2	71	29.8	119	50.0	109	45.8	12	85.7
51	19	6.5	79	23.5	143	42.6	116	34.5	14	73.7
129	5	18.5	12	40.0	19	63.3	10	33.3	<5	80.0
34	11	8.5	19	14.0	82	60.3	68	50.0	10	90.9
19	22	13.3	36	21.8	65	39.4	30	18.2	15	68.2
42	47	12.5	133	32.7	230	56.5	38	9.3	31	66.0
142	12	18.2	21	19.3	44	40.4	10	9.2	6	50.0
66	26	8.9	62	16.9	227	61.9	77	21.0	25	96.2
92	30	16.0	41	20.3	131	64.9	91	45.0	29	96.7
105	13	24.1	18	33.3	35	64.8	8	14.8	12	92.3
5	14	6.4	67	27.5	158	64.8	76	31.1	14	100.0
12	74	13.7	124	20.4	530	87.0	279	45.8	65	87.8
102	79	17.2	145	28.5	203	39.9	142	27.9	66	83.5
9	60	17.5	77	22.3	209	60.6	64	18.6	46	76.7
74	25	15.4	55	32.9	107	64.1	49	29.3	22	88.0
114	<5	--**	17	18.7	37	40.7	15	16.5	<5	50.0
101	30	12.0	85	32.2	139	52.7	85	32.2	27	90.0
128	16	13.2	30	18.9	97	61.0	29	18.2	13	81.2
64	11	16.2	13	18.8	48	69.6	33	47.8	10	90.9
65	24	10.2	81	34.0	184	77.3	30	12.6	22	91.7
110	33	10.4	58	18.2	154	48.3	68	21.3	24	72.7
171	26	24.1	22	20.0	68	61.8	10	9.1	24	92.3
14	34	19.4	20	11.3	104	58.8	42	23.7	27	79.4
70	0	0.0	<5	--**	13	20.3	<5	--**	0	0.0
44	10	5.0	32	11.1	131	45.5	30	10.4	7	70.0
79	5	6.0	18	20.9	19	22.1	10	11.6	<5	80.0
68	60	21.8	59	19.9	199	67.0	182	61.3	52	86.7

* Redacted to adhere to statistical disclosure guidelines.

Appendix 4: 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	10136	89.1
c.350G->A	p.Arg117His	R117H	703	6.2
c.1652G->A	p.Gly551Asp	G551D	639	5.6
c.1624G->T	p.Gly542X	G542X	420	3.7
c.489+1G->T		621+1G->T	301	2.6
c.1585-1G->A		1717-1G->A	191	1.7
c.3909C->G	p.Asn1303Lys	N1303K	182	1.6
c.3454G->C	p.Asp1152His	D1152H	160	1.4
c.1766+1G->A		1898+1G->A	160	1.4
c.200C->T	p.Pro67Leu	P67L	157	1.4
c.3140-26A->G		3272-26A->G	139	1.2
c.3528delC	p.Lys1177SerfsX15	3659delC	131	1.2
c.3718-2477C->T		3849+10kbC->T	127	1.1
c.1679G->C	p.Arg560Thr	R560T	106	0.9
c.1477C->T	p.Gln493X	Q493X	98	0.9
c.1022_1023insTC	p.Phe342HisfsX28	1154insTC	96	0.8
c.1519_1521delATC	p.Ile507del	I507del	95	0.8
c.1657C->T	p.Arg553X	R553X	91	0.8
c.254G->A	p.Gly85Glu	G85E	89	0.8
c.2657+5G->A		2789+5G->A	88	0.8
c.178G->T	p.Glu60X	E60X	79	0.7
c.3846G->A	p.Trp1282X	W1282X	67	0.6
c.617T->G	p.Leu206Trp	L206W	61	0.5
c.1210-12T[5]		5T	59	0.5
c.2052delA	p.Lys684AsnfsX38	2184delA	57	0.5
c.1364C->A	p.Ala455Glu	A455E	57	0.5
c.1646G->A	p.Ser549Asn	S549N	55	0.5
c.948delT	p.Phe316LeufsX12	1078delT	53	0.5
c.1040G->C	p.Arg347Pro	R347P	49	0.4
c.2657+2_2657+3insA		2789+2insA	45	0.4
c.579+3A->G		711+3A->G	39	0.3
c.3484C->T	p.Arg1162X	R1162X	35	0.3
c.1367T->C	p.Val456Ala	V456A	34	0.3
c.1558G->T	p.Val520Phe	V520F	33	0.3
c.1040G->A	p.Arg347His	R347H	32	0.3
c.[1210-12[5];1210-34TG[12]]		5T;TG12	32	0.3
c.1753G->T	p.Glu585X	E585X	30	0.3
c.1000C->T	p.Arg334Trp	R334W	30	0.3
c.1523T->G	p.Phe508Cys	F508C	30	0.3
c.3472C->T	p.Arg1158X	R1158X	30	0.3
c.2988+1G->A		3120+1G->A	29	0.3

Nucleotide	Protein	Legacy name	N	%
c.1329_1330insAGAT	p.Ile444ArgfsX3	1461ins4	27	0.2
c.1705T->G	p.Tyr569Asp	Y569D	26	0.2
c.1055G->A	p.Arg352Gln	R352Q	24	0.2
c.1006_1007insG	p.Ile336SerfsX28	1138insG	24	0.2
c.3197G->A	p.Arg1066His	R1066H	23	0.2
c.349C->T	p.Arg117Cys	R117C	22	0.2
c.2125C->T	p.Arg709X	R709X	22	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	20	0.2
c.653T>A	p.Leu218X	L218X	20	0.2
c.2834C->T	p.Ser945Leu	S945L	20	0.2
c.2583delT	p.Phe861LeufsX3	2711delT	20	0.2
c.3806T->A	p.Ile1269Asn	I1269N	20	0.2
c.2052_2053insA	p.Gln685ThrfsX4	2184insA	20	0.2
c.54-5940_273+10250del21kb	p.Ser18ArgfsX16	CFTRdele2,3	19	0.2
c.3874-2A>G	p.?	4006-2A->G	19	0.2
c.532G->A	p.Gly178Arg	G178R	19	0.2
c.3737C->T	p.Thr1246Ile	T1246I	18	0.2
c.[1521_1523delCTT;3080T->C]	p.[Phe508del;Ile1027Thr]	F508del;I1027T	17	0.1
c.658C->T	p.Gln220X	Q220X	16	0.1
c.579+1G->T		711+1G->T	15	0.1
c.2537G->A	p.Trp846X	W846X	14	0.1
c.2875delG	p.Ala959HisfsX9	3007delG	14	0.1
c.1029delC	p.Cys343X	1161delC	13	0.1
c.2T>C	p.?	M1T	12	0.1
c.1679+1G->C		1811+1G->C	12	0.1
c.292C->T	p.Gln98X	Q98X	12	0.1
c.1645A->C or c.1647T->G or c.1647T->A	p.Ser549Arg	S549R	11	0.1
c.1466C->A	p.Ser489X	S489X	11	0.1
c.3761T->G	p.Leu1254X	L1254X	11	0.1
c.3196C->T	p.Arg1066Cys	R1066C	11	0.1
c.494T->C	p.Leu165Ser	L165S	10	0.1
c.2353C->T	p.Arg785X	R785X	10	0.1
c.1675G->A	p.Ala559Thr	A559T	10	0.1
c.224G->A	p.Arg75Gln	R75Q	10	0.1
c.2988G->A		3120G->A	10	0.1
c.2051_2052delAAinsG	p.Lys684SerfsX38	2183AA->G or 2183delAA->G	9	0.1
c.3468G->A		3600G->A	9	0.1
c.1687T->A	p.Tyr563Asn	Y563N	9	0.1

Nucleotide	Protein	Legacy name	N	%
c.(53+1_54-1)_(489+1_490-1)del		CFTRdele2-4	9	0.1
c.3276C->A or c.3276C->G	p.Tyr1092X	Y1092X	9	0.1
c.3353C->T	p.Ser1118Phe	S1118F	9	0.1
c.3705T->G	p.Ser1235Arg	S1235R	8	0.1
c.709C->G	p.Gln237Glu	Q237E	8	0.1
c.[1210-12[5];1210-34TG[13]]		5T;TG13	8	0.1
c.4196_4197delTC	p.Cys1400X	4326delTC	8	0.1
c.1721C->A	p.Pro574His	P574H	8	0.1
c.695T->A	p.Val232Asp	V232D	8	0.1
c.1766+1G->T		1898+1G->T	7	0.1
c.2012delT	p.Leu671X	2143delT	7	0.1
c.3208C->T	p.Arg1070Trp	R1070W	7	0.1
c.[1210-12[5];1210-34TG[11]] or c.1210-33_1210-6GT[11]T[4] or c.1210-7_1210-6del		5T;TG11	7	0.1
c.(743+1_744-1)_(1584+1_1585-1) dup		CFTRdup6b-10	7	0.1
c.262_263delTT	p.Leu88IlefsX22	394delTT	7	0.1
c.(273+1_274-1)_(1679+1_1680-1) del		CFTRdele4-11	7	0.1
c.349C->G	p.Arg117Gly	R117G	7	0.1
c.1538A->G	p.Asp513Gly	D513G	7	0.1
c.(53+1_54-1)_(164+1_165-1)del		CFTRdele2	6	0.1
c.2991G->C	p.Leu997Phe	L997F	6	0.1
c.3964-78_4242+577del		CFTRdele22,23	6	0.1
c.2900T->C	p.Leu967Ser	L967S	6	0.1
c.1986_1989delAACT	p.Thr663ArgfsX8	2118del4	6	0.1
c.2128A->T	p.Lys710X	K710X	6	0.1
c.2600_2601insA	p.Val868SerfsX28	2732insA	6	0.1
c.2290C->T	p.Arg764X	R764X	6	0.1
c.4147_4148insA	p.Ile1383AsnfsX3	4279insA	6	0.1
c.2551C->T	p.Arg851X	R851X	5	0.0
c.165-3C->T		297-3C->T	5	0.0
c.(273+1_274-1)_(1584+1_1585-1) del		CFTRdele4-10	5	0.0
c.1116+1G->A		1248+1G->A	5	0.0
c.443T->C	p.Ile148Thr	I148T	5	0.0
c.1393-2A->G		1525-2A->G	5	0.0
c.3292T->C	p.Trp1098Arg	W1098R	5	0.0
c.4077_4080delTGTTinsAA	p.Val1360ThrfsX3	4209TGTT->AA	5	0.0
c.3718-1G->A		3850-1G->A	5	0.0
c.3848G->T	p.Arg1283Met	R1283M	5	0.0
c.2491G->T	p.Glu831X	E831X	5	0.0
c.2464G->T	p.Glu822X	E822X	5	0.0
c.4065_4066del	p.Leu1356GlyfsX2	4197_4198delCT	5	0.0
c.1679G->A	p.Arg560Lys	R560K	5	0.0
c.3822G>A	p.Trp1274X	W1274X	5	0.0
c.1545_1546delTA	p.Tyr515X	1677delTA	5	0.0
c.1397C->A or c.1397C->G	p.Ser466X	S466X	5	0.0
c.3884_3885insT	p.Ser1297PhefsX5	4016insT	5	0.0
c.2249C->T	p.Pro750Leu	P750L	5	0.0
c.223C->T	p.Arg75X	R75X	5	0.0

Nucleotide	Protein	Legacy name	N	%
c.168delA	p.Glu56AspfsX35	300delA	5	0.0
c.1210-12T[7]		7T	<5	-
c.429delT	p.Phe143LeufsX10	557delT	<5	-
c.2896delA	p.Thr966ArgfsX2	3028delA	<5	-
c.850dupA	p.Met284AsnfsX3	977insA	<5	-
c.1736A->G	p.Asp579Gly	D579G	<5	-
c.1572C->A	p.Cys524X	C524X	<5	-
c.1219G>T	p.Glu407X	E407X	<5	-
c.575A->G	p.Asp192Gly	D192G	<5	-
c.3254A>C	p.His1085Pro	H1085P	<5	-
c.489+2T>C	p.?	621+2T->C	<5	-
c.2909G->A	p.Gly970Asp	G970D	<5	-
c.933C>G or c.933C>A	p.Phe311Leu	F311L	<5	-
c.3988C->T	p.Gln1330X	Q1330X	<5	-
c.2215delG	p.Val739TyrfsX16	2347delG	<5	-
c.1680A->C	p.Arg560Ser	R560S	<5	-
c.4028dup	p.Cys1344LeufsX15	4160insG	<5	-
c.3095A->G	p.Tyr1032Cys	Y1032C	<5	-
c.595C->T	p.His199Tyr	H199Y	<5	-
c.79G->T	p.Gly27X	G27X	<5	-
c.328G->C	p.Asp110His	D110H	<5	-
c.4139del	p.Thr1380AsnfsX4	4271delC	<5	-
c.2051_2052del	p.Lys684ThrfsX4	2183delAA	<5	-
c.3743C>A|c.3743C>G	p.Ser1248X	S1248X	<5	-
c.274G->A	p.Glu92Lys	E92K	<5	-
c.4046G->A	p.Gly1349Asp	G1349D	<5	-
c.91C->T	p.Arg31Cys	R31C	<5	-
c.577G->T	p.Glu193X	E193X	<5	-
c.4004T->C	p.Leu1335Pro	L1335P	<5	-
c.4111G->T	p.Glu1371X	E1371X	<5	-
c.1651G->A	p.Gly551Ser	G551S	<5	-
c.1327G->T	p.Asp443Tyr	D443Y	<5	-
c.2855T->C	p.Met952Thr	M952T	<5	-
c.3495del	p.Lys1165AsnfsX27	c.3495delG	<5	-
c.2374C->T	p.Arg792X	R792X	<5	-
c.1243_1247del	p.Asn415X	1367del5	<5	-
c.3659delC	p.Thr1220LysfsX8	3791delC	<5	-
c.476T>A	p.Leu159X	L159X	<5	-
c.2924_2925del	p.Arg975IlefsX10	3056delGA	<5	-
c.233dupT	p.Trp79LeufsX32	365-366insT	<5	-
c.442delA	p.Ile148LeufsX5	574delA	<5	-
c.461dup	p.Ala155SerfsX4	593insT	<5	-
c.3872A->G	p.Gln1291Arg	Q1291R	<5	-
c.3752G->A	p.Ser1251Asn	S1251N	<5	-
c.1505T>C	p.Ile502Thr	I502T	<5	-
c.3758dup	p.Leu1253PhefsX12	3889dupT	<5	-
c.1727G->C	p.Gly576Ala	G576A	<5	-
c.743+1G>C	p.?	875+1G->C	<5	-
c.509G->A	p.Arg170His	R170H	<5	-
c.1585-8G->A		1717-8G->A	<5	-
c.1584G->A	p.Glu528Glu	1716G/A	<5	-

Nucleotide	Protein	Legacy name	N	%
c.350G->T	p.Arg117Leu	R117L	<5	-
c.1679+1.6kbA->G		1811+1.6kbA->G	<5	-
c.54-1G>A	p.?	186-1G->A	<5	-
c.3067_3072delATAGTG	p.Ile1023_Val1024del	3199del6	<5	-
c.1340delA	p.Lys447ArgfsX2	1471delA	<5	-
c.1135G->T	p.Glu379X	E379X	<5	-
c.1724T->A	p.Phe575Tyr	F575Y	<5	-
c.601G->A	p.Val201Met	V201M	<5	-
c.3618del	p.Gly1208AlafsX3	3750delA	<5	-
c.(3963+1_3964-1)_(4136+1_4137-1)dup	p.?	CFTRdup22	<5	-
c.3882_3885delTATT	p.Ile1295PhefsX32	4010del4	<5	-
c.1477_1478delCA	p.Gln493ValfsX10	1609delCA	<5	-
c.1766+1G->C		1898+1G->C	<5	-
c.3266G->A	p.Trp1089X	W1089X	<5	-
c.1001G->A	p.Arg334Gln	R334Q	<5	-
c.3017C->A	p.Ala1006Glu	A1006E	<5	-
c.3929G>A	p.Trp1310X	W1310X	<5	-
c.1302del	p.Leu435PhefsX7	1434delA	<5	-
c.2668C->T	p.Gln890X	Q890X	<5	-
c.3476C->T	p.Ser1159Phe	S1159F	<5	-
c.3310G->T	p.Glu1104X	E1104X	<5	-
c.2002C->T	p.Arg668Cys	R668C	<5	-
c.2805_2810del	p.Pro936_Leu937del	2937_2942delinsTCAGA	<5	-
c.413_415dupTAC	p.Leu138dup	L138ins	<5	-
c.1579G>T	p.Glu527X	E527X	<5	-
c.273+1G->A		405+1G->A	<5	-
c.1679+2T>C	p.?	1811+2T->C	<5	-
c.3080T->C	p.Ile1027Thr	I1027T	<5	-
c.3475T->C	p.Ser1159Pro	S1159P	<5	-
c.2523_2548del	p.Ala842SerfsX45	2655del26	<5	-
c.1046C->T	p.Ala349Val	A349V	<5	-
c.1670delC	p.Ser557PhefsX2	1802delC	<5	-
c.3546C>G	p.Tyr1182X	Y1182X	<5	-
c.220C->T	p.Arg74Trp	R74W	<5	-
c.2051_2052delinsG	p.Lys684SerfsX38	2183AA->G	<5	-
c.2739T->A	p.Tyr913X	Y913X	<5	-
c.3365del	p.Thr1122LysfsX12	3497delC	<5	-
c.2260G->A	p.Val754Met	V754M	<5	-
c.3835_3836del	p.Leu1279AlafsX22	3967delTT	<5	-
c.1007T->A	p.Ile336Lys	I336K	<5	-
c.2195T->G	p.Leu732X	L732X	<5	-
c.3763T->C	p.Ser1255Pro	S1255P	<5	-
c.3397del	p.Leu1133X	3528delC	<5	-
c.307G>T	p.Gly103X	G103X	<5	-
c.3717+5G->A		3849+5G->A	<5	-
c.2930C->T	p.Ser977Phe	S977F	<5	-
c.2036G>A|c.2037G>A	p.Trp679X	W679X	<5	-
c.3158C->T	p.Thr1053Ile	T1053I	<5	-
c.3908delA	p.Asn1303ThrfsX25	4040delA	<5	-

Nucleotide	Protein	Legacy name	N	%
c.2274_2275delinsT	p.Thr760ArgfsX11	2406_2407delinsT	<5	-
c.1202G->A or c.1203G->A	p.Trp401X	W401X	<5	-
c.1682C->A	p.Ala561Glu	A561E	<5	-
c.1766+5G->T		1898+5G->T	<5	-
c.1766+3A->G		1898+3A->G	<5	-
c.3874-1G>A	p.?	4006-1G->A	<5	-
c.470_483del14	p.Phe157X	602del14	<5	-
c.3205G->A	p.Gly1069Arg	G1069R	<5	-
c.164+2T->C		296+2T->C	<5	-
c.2810_2811insT	p.Val938GlyfsX37	2942insT	<5	-
c.2780T->C	p.Leu927Pro	L927P	<5	-
c.263T->A or c.263T->G	p.Leu88X	L88X	<5	-
c.2148del	p.Thr717LeufsX5	c.2148delG	<5	-
c.(2988+1_2989-1)_(3367+1_3368-1)del		CFTRdele17a,17b	<5	-
c.3700A->G	p.Ile1234Val	I1234V	<5	-
c.1209+1G->A		1341+1G->A	<5	-
c.2836A>T	p.Lys946X	K946X	<5	-
c.165-2A>G	p.?	297-2A->G	<5	-
c.(2490+1_2491-1)_(2908+1_2909-1)del	p.?	CFTRdele14a-15	<5	-
c.(2908+1_2909-1)_(3873+1_3874-1)dup	p.?	CFTRdup16-20	<5	-
c.3678del	p.Leu1227X	c.3678delA	<5	-
c.3368-2A->G		3500-2A->G	<5	-
c.3181G->C	p.Gly1061Arg	G1061R	<5	-
c.531dup	p.Gly178TrpfsX5	663insT	<5	-
c.3745G->A	p.Gly1249Arg	G1249R	<5	-
c.3176T>G	p.Leu1059X	L1059X	<5	-
c.3717+1G>A	p.?	3849+1G->A	<5	-
c.490-1G>C	p.?	622-1G->C	<5	-
c.4090del	p.Ala1364ArgfsX16	4222delG	<5	-
c.4170del	p.Ala1391HisfsX7	4301delA	<5	-
c.828C->A	p.Cys276X	C276X	<5	-
c.11C->A	p.Ser4X	S4X	<5	-
c.-8G->C		125G/C	<5	-
c.1703delT	p.Leu568CysfsX4	1833delT	<5	-
c.1546A>G	p.Arg516Gly	R516G	<5	-
c.1324A>T	p.Lys442X	K442X	<5	-
c.2620-26A->G		2752-26A->G	<5	-
c.1A>C	p.?	M1L	<5	-
c.571T->G	p.Phe191Val	F191V	<5	-
c.4200_4201del	p.Cys1400X	4332delTG	<5	-
c.1116+2T>A	p.?	1248+2T->A	<5	-
c.(1392+1_1393-1)_(1584+1_1585-1)del	p.?	CFTRdele10	<5	-
c.1954C>T	p.Gln652X	Q652X	<5	-
c.1393-1G>C	p.?	1525-1G->C	<5	-
c.(3468+1_3469-1)_(3717+1_3718-1)dup	p.?	CFTRdup19	<5	-
c.3458T->A	p.Val1153Glu	V1153E	<5	-
c.558del	p.Asn186LysfsX3	690delC	<5	-

Nucleotide	Protein	Legacy name	N	%
c.50delT	p.Phe17SerfsX8	182delT	<5	-
c.908_911del	p.Arg303ThrfsX24	1040del4	<5	-
c.164+1G->A		296+1G->A	<5	-
c.3302T->A	p.Met1101Lys	M1101K	<5	-
c.613C->T	p.Pro205Ser	P205S	<5	-
c.1013C->T	p.Thr338Ile	T338I	<5	-
c.3611G->A or c.3612G->A	p.Trp1204X	W1204X	<5	-
c.743+1G>A	p.?	875+1G->A	<5	-
c.1654C->T	p.Gln552X	Q552X	<5	-
c.2908+1G>A	p.?	3040+1G->A	<5	-
c.(3963+1_3964-1)_(4242+1_4243-1)dup	p.?	CFTRdup22,23	<5	-
c.2735C->A	p.Ser912X	S912X	<5	-
c.2645G->A	p.Trp882X	W882X	<5	-
c.1177del	p.Val393X	1309delG	<5	-
c.1A->G	p.Met1Val	M1V	<5	-
c.137C->A	p.Ala46Asp	A46D	<5	-
c.(2619+1_2620-1)_(2657+1_2658-1)del	p.?	CFTRdele14b	<5	-
c.296C->T	p.Pro99Leu	P99L	<5	-
c.1882G->C or c.1882G->A	p.Gly628Arg	G628R	<5	-
c.4231C->T	p.Gln1411X	Q1411X	<5	-
c.1037T->C	p.Leu346Pro	L346P	<5	-
c.(3873+1_3874-1)_(3963+1_3964-1)del		CFTRdele21	<5	-
c.1911delG	p.Gln637HisfsX26	2043delG	<5	-
c.1420G->A	p.Glu474Lys	E474K	<5	-
c.3773_3774insT	p.Leu1258PhefsX7	3905insT	<5	-
c.234del	p.Trp79GlyfsX12	c.234delC	<5	-
c.2175_2176insA	p.Glu726ArgfsX4	2307insA	<5	-
c.3587C->G	p.Ser1196X	S1196X	<5	-
c.3718-3T->G		3850-3T->G	<5	-
c.2421A->G	p.Ile807Met	I807M	<5	-
c.274-1G->A		406-1G->A	<5	-
c.859_863delAACTT	p.Asn287LysfsX19	991del5	<5	-
c.(2908+1_2909-1)_(3139+1_3140-1)del	p.?	CFTRdele16,17a	<5	-
c.1801A->T	p.Ile601Phe	I601F	<5	-
c.717delG	p.Leu240X	849delG	<5	-
c.2989-1G->A		3121-1G->A	<5	-
c.2835del	p.Ile947PhefsX21	2967delG	<5	-
c.2859_2890delACATTCTGTTCTTCAAGCACCTATGTCAACCC	p.Leu953PhefsX11	2991del32	<5	-
c.1684G->A	p.Val562Ile	V562I	<5	-
c.(1584+1_1585-1)_(1679+1_1680-1)del		CFTRdele11	<5	-
c.4035_4038dup	p.Ser1347ProfsX13	p.S1347PfsX13	<5	-
c.2380del	p.Val794CysfsX9	2512delG	<5	-
c.1081delT	p.Trp361GlyfsX8	1213delT	<5	-
c.1208del	p.Glu403GlyfsX39	1340delA	<5	-
c.4364C->G	p.Ser1455X	S1455X	<5	-
c.2433_2437delinsATA	p.Leu812TyrfsX10	c.2433_2437delinsATA	<5	-

Nucleotide	Protein	Legacy name	N	%
c.50dup	p.Ser18GlnfsX27	175insT	<5	-
c.2277del	p.Thr760ArgfsX11	2409delC	<5	-
c.3011_3019delCTATAGCAG or c.3009_3017delAGCTATAGC	p.Ala1004_Ala1006del	3143del9	<5	-
c.1837G->A	p.Ala613Thr	A613T	<5	-
c.53+1G->T		185+1G->T	<5	-
c.2083del	p.Glu695LysfsX27	2215delG	<5	-
c.273+2T>G	p.?	405+2T->G	<5	-
c.3971T->C	p.Leu1324Pro	L1324P	<5	-
c.285dup	p.Ala96SerfsX15	415insA	<5	-
c.3194T->C	p.Leu1065Pro	L1065P	<5	-
c.310delA	p.Arg104GlufsX3	442delA	<5	-
c.(2988+1_2989-1)_(3468+1_3469-1)del		CFTRdele17a-18	<5	-
c.472dup	p.Ser158LysfsX5	604insA	<5	-
c.88_89del	p.Gln30AlafsX14	c.88_89delCA	<5	-
c.927del	p.Phe310SerfsX18	1058delC	<5	-
c.1573C->T	p.Gln525X	Q525X	<5	-
c.1418delG	p.Gly473GlufsX54	1548delG	<5	-
c.3230T->C	p.Leu1077Pro	L1077P	<5	-
c.1820_1903del84	p.Met607_Gln634del	1949del84	<5	-
c.445G>A	p.Gly149Arg	G149R	<5	-
c.3808G->A	p.Asp1270Asn	D1270N	<5	-
c.1585-1G>T	p.?	1717-1G->T	<5	-
c.3717G->A		3849G->A	<5	-
c.274-2A->G		406-2A->G	<5	-
c.2619+2T>C	p.?	2751+2T->C	<5	-
c.53+2T>G	p.?	185+2T->G	<5	-
c.1687T->G	p.Tyr563Asp	Y563D	<5	-
c.2089dup	p.Arg697LysfsX33	2221insA	<5	-
c.1969A>T	p.Arg657X	R657X	<5	-
c.3530del	p.Lys1177SerfsX15	3662delA	<5	-
c.1301C->A or c.1301C->G	p.Ser434X	S434X	<5	-
c.869+1G>C	p.?	1001+1G->C	<5	-
c.3297C->A	p.Phe1099Leu	F1099L	<5	-
c.(3367+1_3368-1)_(3873+1_3874-1)del	p.?	CFTRdele18-20	<5	-
c.3932_3933delinsAATATG	p.Ser1311LysfsX12	4064-4065delinsAATATG	<5	-
c.3209G->A	p.Arg1070Gln	R1070Q	<5	-
c.1117-1G->A		1249-1G->A	<5	-
c.2909del	p.Gly970ValfsX11	3041delG	<5	-
c.1469del	p.Phe490SerfsX37	1601delT	<5	-
'Other' selected			269	2.4
Missing/not known			346	3.0

* Redacted to adhere to statistical disclosure guidelines.

Appendix 5: Genotype lists for CFTRm eligibility

FDA list of CFTR variants potentially responsive to elexacaftor/tezacaftor/ivacaftor[1]

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

1 **pi.vrtx.com/files/uspi_elexacaftor_tezacaftor_ivacaftor.pdf** (accessed prior to December 2024, has since been updated to include 94 additional mutations as of December 2024, current prescribing information contains the additional 94 mutations listed here: **www.cff.org/news/2024-12/fda-approves-trikafta-additional-rare-cftr-mutations**. These were not considered in this report as they were not named eligible variants during the 2024 annual review year.

FDA list of additional[2] CFTR variants potentially responsive to either tezacaftor/ivacaftor or ivacaftor

	Named variant
Tezacaftor/ivacaftor and Ivacaftor extension list eligible	2789+5G->A
	3272-26A->G
	3849+10kbC->T
	711+3A->G
	D443Y;G576A;R668C
	E831X
	F508C;S1251N
Tezacaftor/ivacaftor ONLY extension list eligible	G576A;R668C
	R74W;D1270N
	R74W;V201M
	R74W;V201M;D1270N

Potentially responsive CFTR variants according to French Compassionate Use Programme[3]

	Named variant
French compassionate use programme	2789+5G->A
	3041-15T->G
	3272-26A->G
	3849+10kbC->T
	N1303K
	R1066C
	R334W

2 **www.cff.org/managing-cf/cftr-modulator-therapies**; there are a total of 154 named variants potentially responsive to tezacaftor/ivacaftor and 97 variants potentially responsive to ivacaftor. The table above is the list of variants responsive to either tezacaftor/ivacaftor or ivacaftor which are not named as potentially responsive to elexacaftor/tezacaftor/ivacaftor.

3 Burgel PR et al. Eur Respir J. Feb 2023; variants may overlap with named variants in list 4.2.

Appendix 6: Additional tables

Complications (all ages) 2019 and 2024

Complications	2019	2024
Respiratory related		
Nasal polyps	426 (4.2)	376 (3.6)
Sinus disease	797 (7.9)	782 (7.5)
Asthma	925 (9.1)	689 (6.6)
ABPA	754 (7.4)	491 (4.7)
Any haemoptysis	420 (4.1)	-*
Massive haemoptysis	-*	6 (0.1)
Pneumothorax requiring chest tube	27 (0.3)	5 (0.0)
Cardiac complications		
Tachyarrhythmia	25 (0.2)	-*
Bradycardia	5 (0.0)	<5
Cardiac arrest	<5	<5
Cardiomyopathy	8 (0.1)	18 (0.2)
Congenital heart disease	18 (0.2)	23 (0.2)
Heart failure	<5	10 (0.1)
Ischaemic heart disease	6 (0.1)	9 (0.1)
Valvular disease	-*	-*
Other	67 (0.7)	79 (0.8)
Pancreas and hepatobiliary disease		
Raised liver enzymes	1010 (10.0)	1270 (12.1)
Liver disease	1467 (14.5)	1918 (18.3)
Cirrhosis with no portal hypertension	63 (0.6)	92 (0.9)
Cirrhosis with portal hypertension	135 (1.3)	125 (1.2)
Gall bladder disease requiring surgery	143 (1.4)	262 (2.5)
Pancreatitis	65 (0.6)	-*
Upper gastrointestinal (GI)		
Gastro-oesphageal reflux disease (GORD)	1696 (16.7)	1634 (15.6)
Peptic ulcer	<5	<5
GI bleed (varices as source)	-*	11 (0.1)
GI bleed (non varices as source)	-*	16 (0.2)

Complications (all ages) 2019 and 2024 (cont.)

Complications	2019	2024
Lower gastrointestinal		
Intestinal obstruction	34 (0.3)	49 (0.5)
DIOS	570 (5.6)	364 (3.5)
Fibrosing colonopathy / colonic stricture	<5	<5
Rectal prolapse	12 (0.1)	13 (0.1)
Renal		
Kidney stones	111 (1.1)	177 (1.7)
Renal failure	97 (1.0)	92 (0.9)
Musculoskeletal		
Arthritis	100 (1.0)	-*
Arthropathy	250 (2.5)	-*
Bone fracture	47 (0.5)	36 (0.3)
Osteopenia	975 (9.6)	1146 (10.9)
Osteoporosis	414 (4.1)	-*
Other		
Cancer confirmed by histology	28 (0.3)	-*
Port inserted or replaced	273 (2.7)	85 (0.8)
Depression	446 (4.4)	-*
Hearing loss	339 (3.3)	408 (3.9)
Hypertension	126 (1.2)	-*
Urinary incontinence	939 (9.3)	698 (6.7)
Faecal incontinence	74 (0.7)	81 (0.8)
Postural Anomaly	964 (9.5)	538 (5.1)

CF population by devolved nation

Country	Number registered	Number with annual review
England	9436	8892
Northern Ireland	493	367
Scotland	969	731
Wales	483	434

Glossary

Word/phrase	Meaning
2024	1 January 2024–31 December 2024.
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 mutation.
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).
GORD (gastro-oesophageal 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.
Mutation	A mutation is a change in a gene. When both of a child's parents are carriers of a CF-causing mutation there is a 25% chance that the child will have cystic fibrosis. There are over 1,400 different mutations of the CFTR gene that can cause cystic fibrosis.
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.
p-value	A number that describes the likelihood of obtaining the observed data under the null hypothesis in a statistical test. It indicates how likely it is that the observed results occurred by random chance. A smaller p-value suggests stronger evidence against the null hypothesis.
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.

Cystic Fibrosis Trust

Cystic Fibrosis Trust is the charity uniting people to stop cystic fibrosis. Our community will improve care, speak out, support each other and fund vital research as we race towards effective treatments for all.

We won't stop until everyone can live without the limits of cystic fibrosis.

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