Cystic Fibrosis Trust

UK Cystic Fibrosis Registry
2022 Annual Data Report

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Version 3

Uniting for a life unlimited

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UK Cystic Fibrosis Registry
2022 Annual Data Report

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

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Acknowledgements

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

Contact information

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Cystic Fibrosis Trust

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Foreword

I am pleased to share the 2022 UK CF Registry annual data report with you. In 2022, for the first time ever, we report over 11,000 people with CF registered on the UK CF Registry. I would like to thank all people with cystic fibrosis across the UK, and their families, who consent to sharing their data along with the clinical teams who enter it; without whom the ongoing work of the UK CF Registry would simply not be possible.

As well as our community growing in size, many are continuing to live longer and healthier lives. We have seen the median age of people with CF reach 22 years, 63% are now 16 and over, and, for the first time, we have expanded our reporting age categories for people over 60. Once again, we report a substantial increase in the number of women with CF (140) having babies. The median predicted survival age for people born today has increased to 56, with the gap between men and women decreasing. The results from the James Lind Alliance research priority refresh concluded how we manage an aging population with CF to be a key priority for our community. The data collected by the UK CF Registry will no doubt contribute to future research in this area.

More people than ever were reported to be in employment or education. In 2022 the Trust launched Work Forwards, a programme of free tailored careers support for people with CF and their loved ones, supported by the National Lottery Community Fund. Work Forwards offers information, advice, signposting, and practical support. To find out more, please visit **cysticfibrosis.org.uk/workforwards** or email the team at **workforwards@cysticfibrosis.org.uk**.

In early 2022, we saw the approval of Kaftrio for use in eligible children aged 6-11, and by the end of the year, 7,950 people across the UK were taking a modulator. The data in this year's report shows how modulator use has changed since 2020. 2022 also saw the final year of data collection to support the NICE appraisal of the CFTR modulator therapies. The team have analysed the UK CF Registry data and submitted the final data report to support the appraisal. I would like to thank the CF teams across the country who have entered additional encounters onto the system, making this important work possible. The Trust will continue to contribute to the evidence base as the appraisal process concludes in the coming months.

Whilst 2022 saw the end of most COVID-19 restrictions, we continued to see increasing numbers of infections, with over 2,000 cases reported in 2022. The number of people infected requiring hospitalisation has, however, continued to decrease since early in the pandemic, with a much lower rate of 3.8% hospitalised in 2022 compared to 24% in 2020. The team continue to play an important role in the global collaboration group, monitoring and reporting on the impact on people with CF internationally.

The data insights presented in the 2022 Registry annual data report reflect the outcomes of people with CF as a whole – it doesn't reflect the personal stories and individual experiences of our community. We know that not everyone can benefit from modulator therapies and that there is still much more work to do, which is why we won't stop uniting to make sure everyone with CF can live a life unlimited.

I hope you find the report useful and insightful. We would love to hear your feedback on this report and what you would like to see in future reports. You can contact us by email at **registry@cysticfibrosis.org.uk** or via social media if you have any comments or questions.

Finally, I would like to thank Professor Siobhan Carr who after eight years as Chair of the Registry Steering Committee has handed over the chair to Dr Jamie Duckers. Professor Carr will continue to work closely with the team and I wish her every success in her future projects. I warmly welcome Dr Duckers to his new role.

David Ramsden
Chief Executive of Cystic Fibrosis Trust

Executive summary

The 2022 Registry data remains a hugely valuable and reliable resource to help CF teams, researchers and those living with cystic fibrosis understand the current health of people with CF in the UK. It is a rich information source which the UK CF community as a whole should be proud of, celebrate and use to drive forward improvements in CF care as we emerge from COVID-19 and into an era of expanding novel therapies. I would like to highlight some aspects of this year's report.

- 11,148 people with CF are registered within the UK CF Registry of whom 92% had an annual review this year (Section 1.1)
- 62.9% of the population are over 16 years of age (Section 1.1)
- 5.4 % of the UK CF population report being non-white or of mixed ethnicity (Section 1.4)
- 66.1% of people with CF over 16years of age are in work or studying (Section 1.9)
- The median best FEV_1% continues to rise and is now 88.9% (section 1.14)
- Nutritional status is changing (section 1.8) shows BMI trends over the last four years, with a smaller proportion now being underweight but an increasing proportion of adults with a BMI >= 25.
- Depression reported in those ≥16 years old has fallen slightly from 8.1% in 2021 to 7.6% this year (section 1.22)
- 140 women had babies in 2022, which is another large annual increase
- 28.7% of people with CF over 10 years of age are on CF diabetes therapies (Section 1.24)
- 90.1% of people with CF had one respiratory culture sent this year (Section 1.19) but the sputum samples made up a smaller proportion of the sample type
- NTM prevalence has fallen again this year to 3.1% with reductions in *M. abscessus* prevalence to 0.9% (Section 1.20)
- The percentage of people receiving at least one course of IV antibiotics (22.3%) has dropped again this year (Section 1.25)
- 22.4% of people with CF remain on the combination of inhaled antibiotics, DNase and hypertonic saline or mannitol (Section 1.33) 19.5% of people with CF are on none of these inhaled therapies
- 7,950 people with CF were reported as being on a CFTR modulator by December 2022 (section 1.34) - reflected in a new graph breaking down each modulator use by month

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 and types of mucolytics used. Tables of outcome data for centres must be interpreted with caution; a lot of centres are not large enough to allow meaningful comparisons.

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

Jamie Duckers
Chair of the UK CF Registry Steering Committee

Introduction

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

You can find a Glossary of scientific and clinical terms on page 65.

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

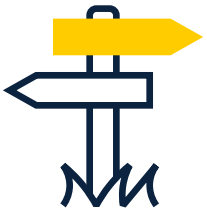
Cystic fibrosis

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

UK Cystic Fibrosis Registry

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

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



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

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

monitoring the safety and effectiveness of new treatments for cystic fibrosis

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

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

Governance

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

Please see Appendix 1: UK CF Registry Committee Structure.

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

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

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

Data collection

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

Where can I find more information?

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

Section 1: UK-wide analysis

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

1.1 Summary of the UK Cystic Fibrosis Registry

	2017	2018	2019	2020	2021	2022
CF patients Registered[1]	10469	10509	10655	10837	10908	11148
Excluding diagnoses that year	10255	10287	10462	10632	10720	10925
CF patients with an annual review; n(%)[2]	9887 (96)	9847 (96)	10070 (96)	9922 (93*)	10175 (95)	10251 (94)
Age in years; median[3]	20	20	21	21	21	22
All newly diagnosed patients (NBS and other)[4]	214	222	193	205	188	223
All newly diagnosed patients (amended)[5]	(304)	(303)	(282)	(262)	(246)	(TBD)
Number of patients born identified by NBS[4]	192	167	150	152	134	162
Number of patients born identified by NBS (amended)[5]	(208)	(178)	(171)	(176)	(155)	(TBD)
Age at diagnosis in months; median[3]	2	2	2	2	2	2
Adults aged 16 years and over; %[3]	60.6	60.4	60.6	60.6	61.9	62.9
Males; %[3]	53.3	53	53.2	53.1	53.2	53.1
Genotyped; %[3]	99.3**	99.1	99.2	99.2	99.1	99.5
Total deaths reported during annual review year (%)[6]	132 (1.3%)	137 (1.3%)	114 (1.1%)	97 (0.9%)	66 (0.6%)	64 (0.6%)
Total deaths reported amended(%)	143	143	119	102	68	(TBD)
Age at death in years; median (95% CI)[6]	31(29, 35)	32 (29, 35)	31 (29, 34)	36 (32, 38)	39 (36, 42)	33 (31, 39)

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

Notes:

* Corrected from 2020 report.

** Corrected from 2017 report.

1 Number of patients diagnosed with CF, seen in the last two years, and alive at 1 January in the given year.

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

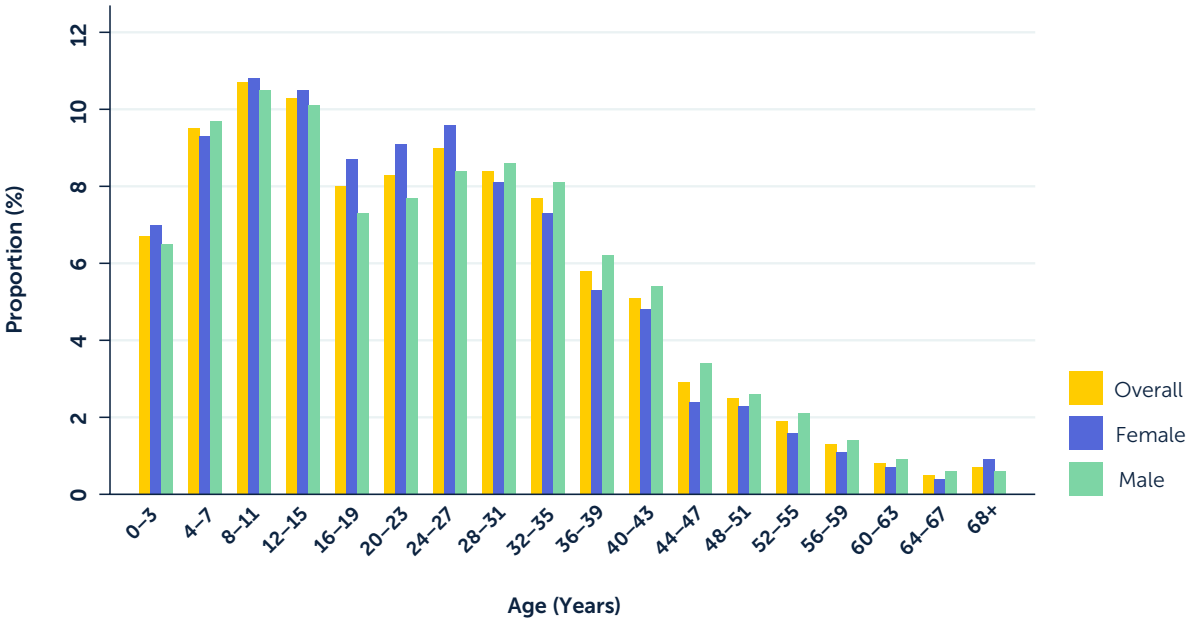
3 Calculated from patients with an annual review in the given year (see footnote 5 below).

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

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

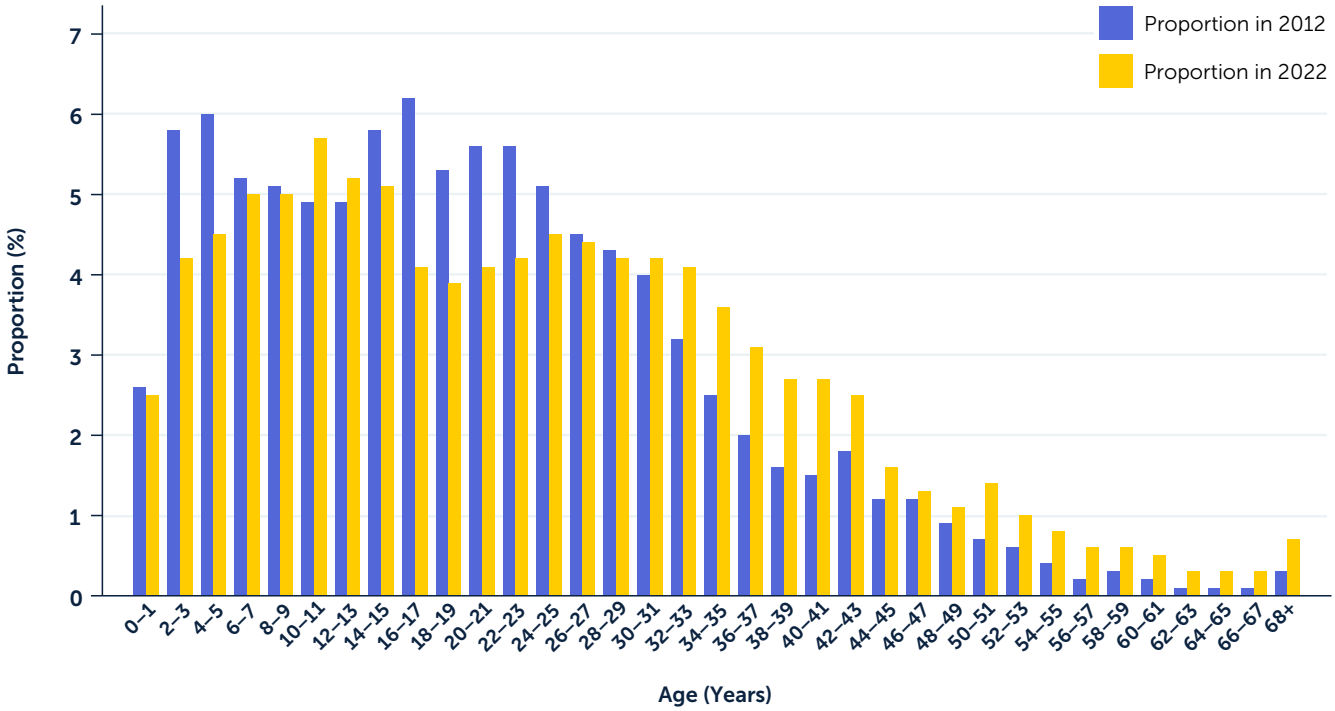
6 Calculated from all registered patients who died in the given year.

1.2 Age distribution by sex
N=10251



Age	All; n(%)	Females; n(%)	Males; n(%)
0-3	689 (6.7)	337 (7.0)	352 (6.5)
4-7	971 (9.5)	445 (9.3)	526 (9.7)
8-11	1093 (10.7)	521 (10.8)	572 (10.5)
12-15	1053 (10.3)	503 (10.5)	550 (10.1)
16-19	818 (8.0)	419 (8.7)	399 (7.3)
20-23	853 (8.3)	436 (9.1)	417 (7.7)
24-27	919 (9.0)	460 (9.6)	459 (8.4)
28-31	859 (8.4)	389 (8.1)	470 (8.6)
32-35	793 (7.7)	350 (7.3)	443 (8.1)
36-39	594 (5.8)	255 (5.3)	339 (6.2)
40-43	524 (5.1)	231 (4.8)	293 (5.4)
44-47	300 (2.9)	114 (2.4)	186 (3.4)
48-51	254 (2.5)	112 (2.3)	142 (2.6)
52-55	190 (1.9)	78 (1.6)	112 (2.1)
56-59	130 (1.3)	55 (1.1)	75 (1.4)
60-63	82 (0.8)	35 (0.7)	47 (0.9)
64-67	54 (0.5)	21 (0.4)	33 (0.6)
68+	75 (0.7)	44 (0.9)	31 (0.6)
<16	3806 (37.1)	1806 (37.6)	2000 (36.7)
≥16	6445 (62.9)	2999 (62.4)	3446 (63.3)
<18	4226 (41.2)	2015 (41.9)	2211 (40.6)
≥18	6025 (58.8)	2790 (58.1)	3235 (59.4)
Overall	**10251**	**4805**	**5446**

1.3 Age distribution of the UK CF population in 2012 vs 2022
N=10251 in 2022, N=8794 in 2012



1.4 Ethnicity

Ethnicity n(%)	2012	2017	2022
Total	8794	9887	10251
Total known[1]	8751	9750	9933
White	**8340 (95.3)**	**9326 (95.7)**	**9398 (94.6)**
Asian	**229 (2.6)**	**265 (2.7)**	**312 (3.1)**
Bangladeshi	32 (0.4)	33 (0.3)	42 (0.4)
Indian	29 (0.3)	41 (0.4)	50 (0.5)
Pakistani	145 (1.7)	163 (1.7)	189 (1.9)
Other (Asian)	23 (0.3)	28 (0.3)	31 (0.3)
Black	**24 (0.3)**	**33 (0.3)**	**28 (0.3)**
Black African	9 (0.1)	13 (0.1)	12 (0.1)
Black Caribbean	12 (0.1)	15 (0.2)	11 (0.1)
Other (Black)	<5*	5 (0.1)	5 (0.1)
Mixed	**69 (0.8)**	**40 (0.4)**	**85 (0.9)**
Mixed (white-Asian)	-	9 (0.1)	20 (0.2)
Mixed (white-Black African)	-	6 (0.1)	12 (0.1)
Mixed (white-Black Caribbean)	-	12 (0.1)	28 (0.3)
Other (mixed)	-	13 (0.1)	25 (0.3)
Other	**89 (1.0)**	**86 (0.9)**	**110 (1.1)**

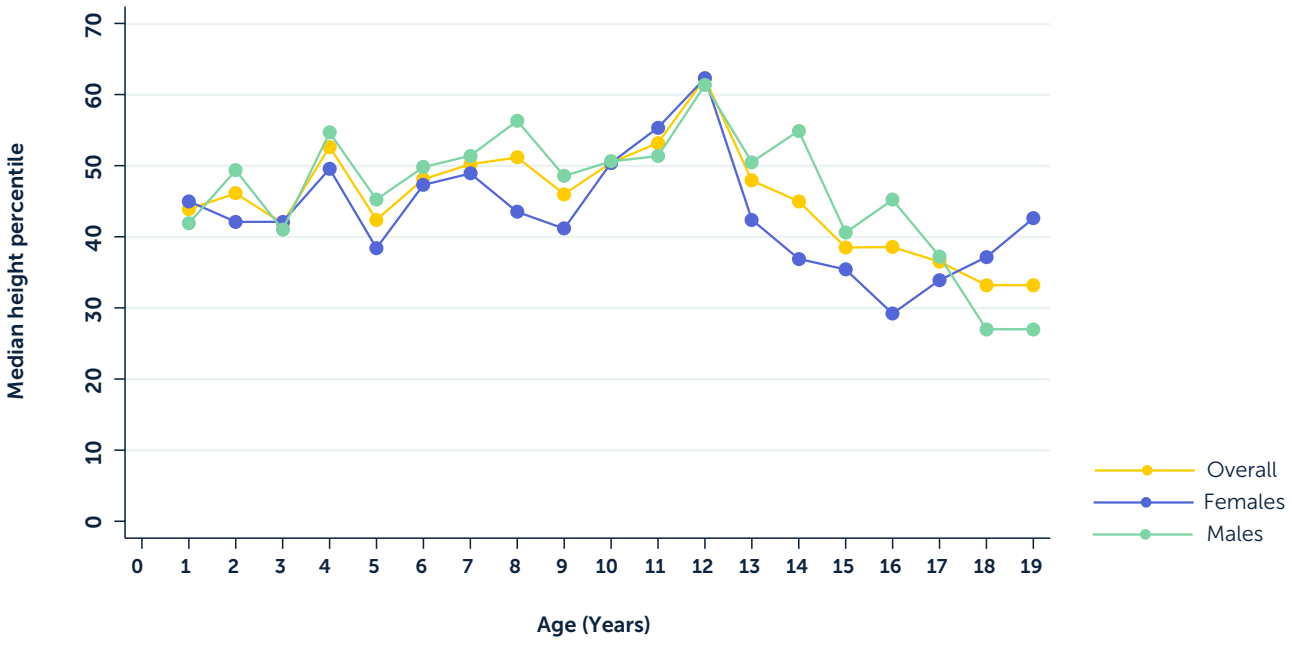
* Redacted to adhere to statistical disclosure guidelines.

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

[1] Proportions are calculated from total known ethnicities.

1.5 Height percentiles of children and young people (<20 years)[1] N=4624

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



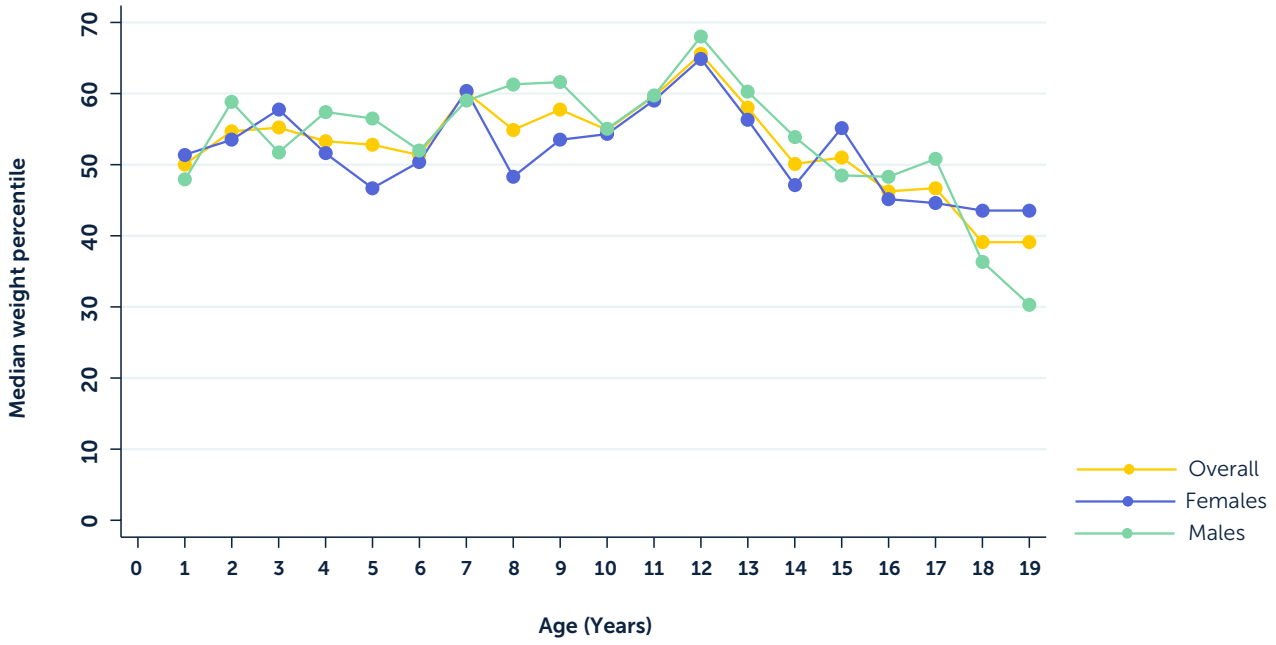
	Overall			Female			Male		
Age	n	Median	IQR	n	Median	IQR	n	Median	IQR
1	191	43.9	18.2-78.7	99	45.0	12.6-80.4	92	41.9	19.8-74.3
2	181	46.1	21.3-75.9	87	42.1	21.1-75.9	94	49.3	22.4-76.2
3	227	41.9	19.3-72.8	103	42.1	15.8-71.7	124	41.0	22.2-73.7
4	196	52.7	22.5-75.8	82	49.5	21.8-75.0	114	54.7	24.8-76.5
5	255	42.4	17.7-76.8	123	38.4	14.4-68.7	132	45.2	18.7-82.4
6	257	48.1	25.4-75.6	116	47.3	24.7-75.9	141	49.8	26.9-73.5
7	238	50.2	25.8-81.0	112	48.9	26.3-79.2	126	51.4	25.8-81.0
8	240	51.2	24.0-79.4	109	43.5	22.2-75.0	131	56.3	27.3-84.0
9	260	46.0	21.0-71.4	122	41.2	19.4-76.0	138	48.5	25.1-69.1
10	291	50.4	25.6-76.1	151	50.4	26.6-78.8	140	50.6	24.8-75.2
11	284	53.2	28.3-82.0	132	55.3	24.7-82.8	152	51.3	29.2-81.3
12	269	62.3	34.2-83.7	129	62.3	37.7-83.9	140	61.3	28.7-82.1
13	248	47.9	28.2-75.9	112	42.4	20.4-69.4	136	50.5	33.6-77.9
14	254	45.0	21.8-68.4	133	36.9	20.0-59.0	121	54.9	27.2-73.5
15	263	38.5	16.4-64.5	122	35.4	15.3-61.5	141	40.6	17.1-64.7
16	206	38.6	15.4-63.4	106	29.2	12.0-63.4	100	45.2	20.2-63.8
17	209	36.5	15.9-57.6	99	33.9	15.9-54.5	110	37.2	16.1-60.6
18	215	33.2	13.6-59.4	122	37.2	13.8-60.4	93	27.0	9.7-54.7
19	183	33.2	13.1-59.0	88	42.6	16.3-59.0	95	27.0	9.1-59.1
Overall	**4467***	**46.0**	**21.1-73.7**	**2147**	**43.2**	**19.8-72.0**	**2320**	**48.3**	**22.4-74.2**

* Number with non-missing data.

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

1.6 Weight percentiles of children and young people (<20 years)[1] N=4624

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.



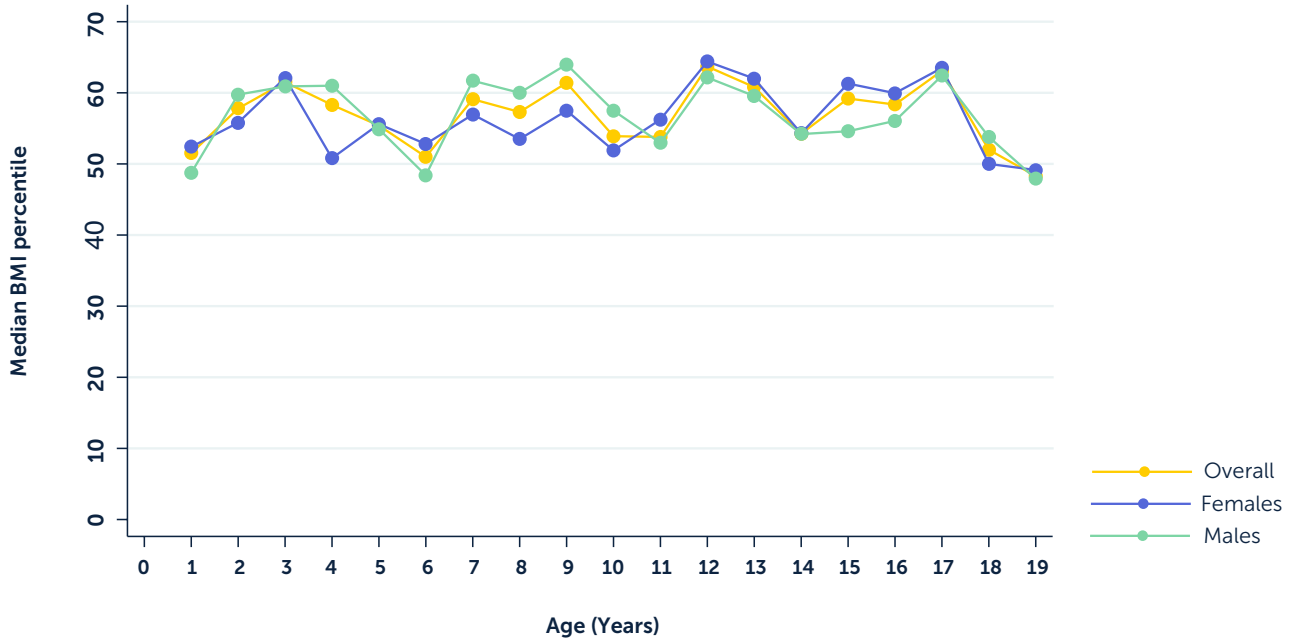
	Overall			Female			Male		
Age	n	Median	IQR	n	Median	IQR	n	Median	IQR
1	200	50.0	22.8-76.6	104	51.4	23.6-81.9	96	47.9	21.8-73.8
2	189	54.7	26.0-78.8	92	53.5	26.0-77.9	97	58.8	26.0-79.8
3	232	55.2	30.8-77.4	104	57.8	25.8-76.7	128	51.8	31.5-80.9
4	201	53.3	31.8-80.9	84	51.6	26.2-73.1	117	57.4	33.3-85.0
5	256	52.8	25.0-76.4	124	46.7	24.7-73.4	132	56.5	25.4-79.1
6	260	51.3	30.5-74.5	118	50.4	30.4-74.9	142	52.0	30.8-74.1
7	239	60.2	31.8-80.1	112	60.4	32.8-77.1	127	59.0	31.8-83.3
8	242	54.9	31.4-83.7	110	48.3	24.0-78.8	132	61.3	41.4-85.9
9	260	57.8	30.3-78.1	123	53.5	25.1-76.8	137	61.6	33.6-78.7
10	291	54.9	29.1-81.8	151	54.3	29.1-76.8	140	55.1	28.5-85.1
11	285	59.6	32.5-85.6	132	59.0	26.9-85.7	153	59.7	34.2-85.6
12	270	65.6	37.9-86.8	129	64.9	39.8-86.8	141	68.0	30.3-86.4
13	247	58.0	30.7-83.6	111	56.3	30.0-81.0	136	60.2	32.1-87.1
14	257	50.1	25.8-74.5	133	47.1	21.3-69.7	124	53.9	29.3-79.2
15	263	51.0	23.6-81.9	122	55.2	23.6-82.4	141	48.5	24.0-77.6
16	206	46.2	25.7-72.3	106	45.2	22.3-70.8	100	48.3	30.7-75.3
17	209	46.7	19.2-75.4	100	44.6	24.1-77.7	109	50.8	18.7-71.2
18	214	39.1	14.4-71.3	122	43.5	16.7-72.5	92	36.3	10.7-65.3
19	183	39.1	9.4-65.1	88	43.6	18.4-64.4	95	30.3	7.0-69.6
Overall	**4504***	**53.8**	**26.5-79.0**	**2165**	**52.4**	**25.6-77.1**	**2339**	**54.9**	**27.8-80.8**

* Number with non-missing data.

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

1.7a Body Mass Index (BMI) percentiles in children and young people (<20 years)[1]
N=4624

The following chart and table show the BMI percentiles 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, it means that only 40% of the population at the same age have the same BMI or lower; 60% have a higher BMI.



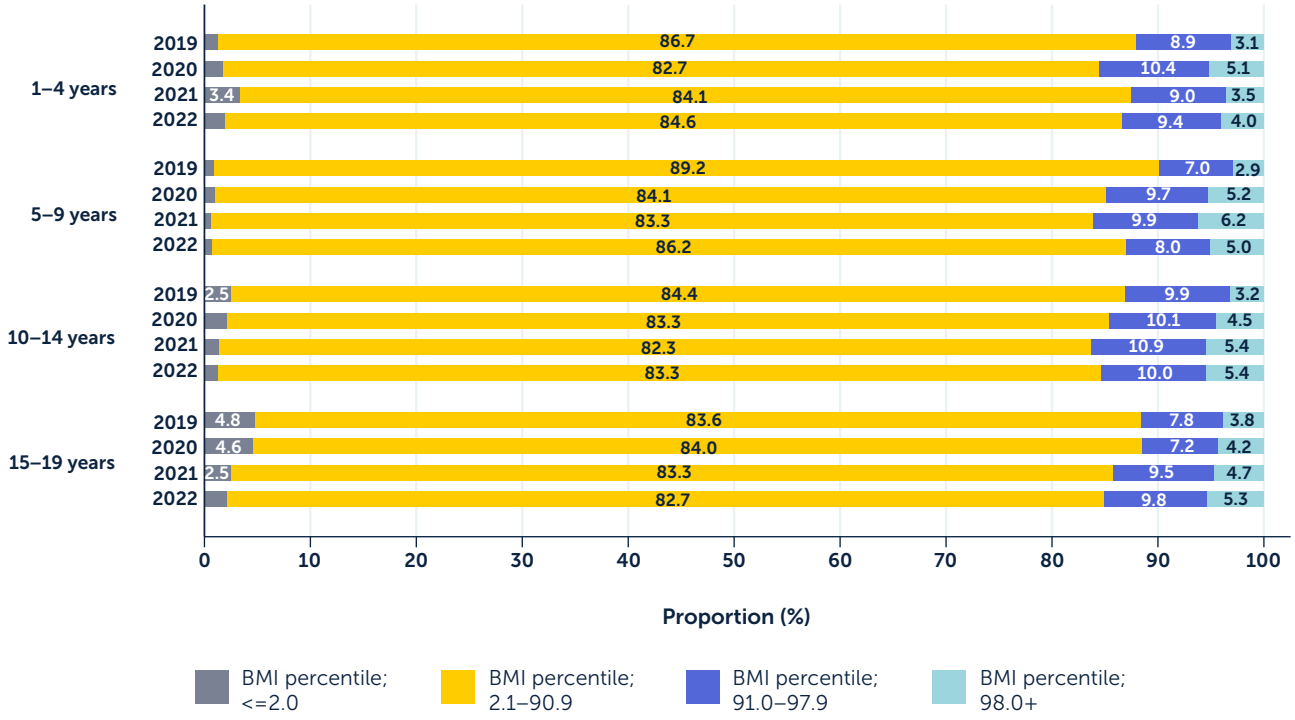
	Overall			Female			Male		
Age	n	Median	IQR	n	Median	IQR	n	Median	IQR
1	191	51.5	22.3-77.7	99	52.4	23.0-75.1	92	48.8	21.0-77.8
2	181	57.8	25.3-81.7	87	55.8	26.0-79.5	94	59.8	21.0-84.6
3	227	61.5	32.7-80.7	103	62.1	31.5-79.6	124	60.9	33.1-83.2
4	196	58.3	30.1-81.9	82	50.8	28.5-77.9	114	61.0	32.5-85.6
5	255	55.4	34.1-76.6	123	55.6	34.0-76.3	132	54.9	34.7-77.8
6	257	51.0	31.4-74.0	116	52.8	32.2-76.6	141	48.4	28.4-73.8
7	238	59.1	33.4-79.9	112	56.9	36.5-78.2	126	61.7	30.6-81.8
8	240	57.3	37.2-78.7	109	53.5	34.9-74.0	131	60.0	38.6-84.6
9	259	61.4	41.2-86.0	122	57.5	38.9-83.0	137	64.0	43.5-86.1
10	291	53.9	31.0-81.2	151	51.9	32.9-78.0	140	57.5	29.9-85.7
11	284	53.8	33.3-84.6	132	56.2	33.0-80.9	152	53.0	34.3-86.3
12	269	63.7	32.3-85.6	129	64.4	33.7-86.3	140	62.2	31.0-84.3
13	247	60.8	31.1-85.6	111	62.0	33.5-84.0	136	59.6	25.5-87.1
14	254	54.2	33.4-82.8	133	54.3	34.4-81.3	121	54.2	33.4-84.6
15	263	59.2	33.2-84.9	122	61.3	33.2-86.4	141	54.6	34.2-79.6
16	206	58.3	35.8-79.3	106	60.0	36.8-79.1	100	56.0	31.9-80.8
17	208	63.1	34.8-82.5	99	63.5	27.7-84.0	109	62.4	37.1-81.1
18	214	52.0	28.1-82.7	122	50.0	29.9-84.2	92	53.8	25.5-76.6
19	183	48.3	20.5-73.1	88	49.2	23.7-71.9	95	47.9	18.2-76.0
Overall	**4463***	**56.6**	**31.9-81.5**	**2146**	**56.1**	**32.1-80.0**	**2317**	**57.4**	**31.9-83.4**

* Number with non-missing data.

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

1.7b Body Mass Index (BMI) percentiles in children and young people (<20 years)[1] for 2019–2022

The following graph shows the change in BMI groups for children and young people with CF from 2019 to 2022.



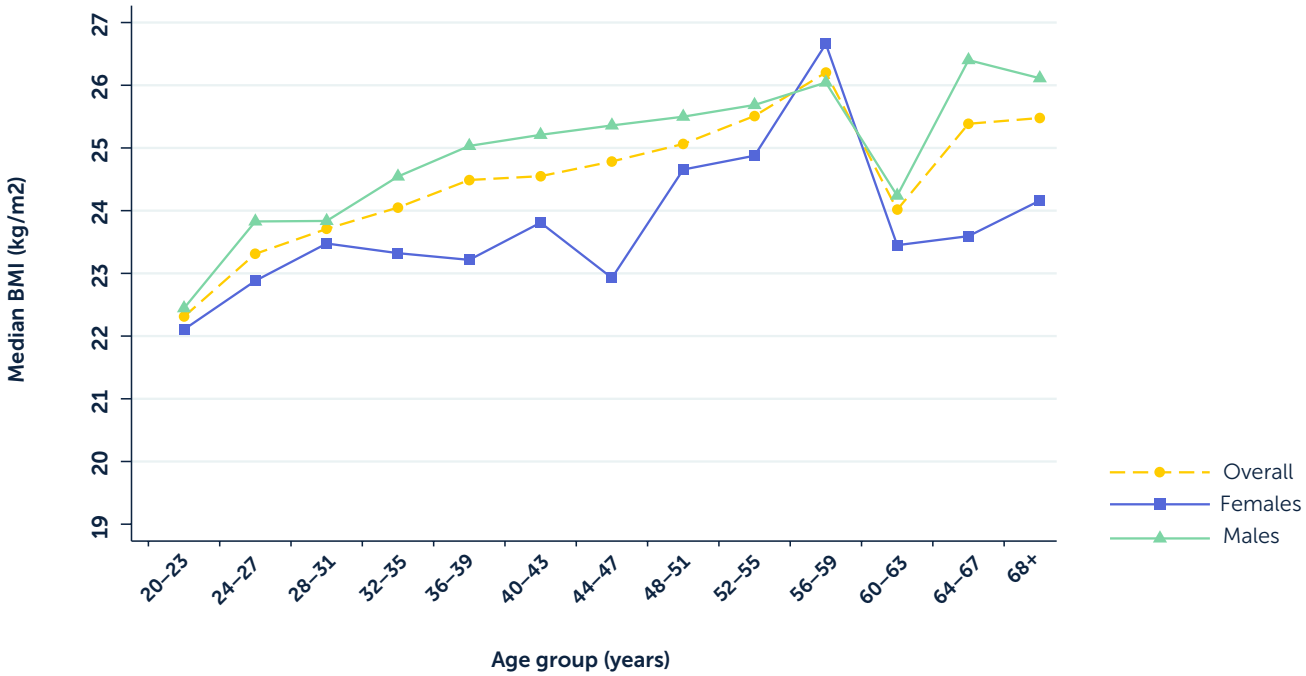
			BMI category by age and year : n*(%)			
Age group	Year	Total number of people in each age group	BMI percentile; <=2.0	BMI percentile; 2.1-90.9	BMI percentile; 91.0-97.9	BMI percentile; 98.0+
1-4 years	2019	1064	13 (1.3)	837 (86.6)	86 (8.9)	30 (3.1)
	2020	985	13 (1.8)	589 (82.7)	74 (10.4)	36 (5.1)
	2021	967	26 (3.4)	652 (84.1)	70 (9.0)	27 (3.5)
	2022	894	16 (2.0)	672 (84.5)	75 (9.4)	32 (4.0)
5-9 years						
	2019	1395	12 (0.9)	1242 (89.2)	98 (7.0)	40 (2.9)
	2020	1361	11 (1.0)	942 (84.2)	108 (9.7)	58 (5.2)
	2021	1321	7 (0.6)	1023 (83.3)	122 (9.9)	76 (6.2)
	2022	1279	10 (0.8)	1076 (86.1)	100 (8.0)	63 (5.0)
10-14 years						
	2019	1262	31 (2.5)	1061 (84.4)	125 (9.9)	40 (3.2)
	2020	1307	23 (2.1)	917 (83.4)	111 (10.1)	49 (4.5)
	2021	1360	18 (1.4)	1047 (82.2)	139 (10.9)	69 (5.4)
	2022	1368	17 (1.3)	1122 (83.4)	134 (10.0)	72 (5.4)
15-19 years						
	2019	1057	51 (4.8)	880 (83.6)	82 (7.8)	40 (3.8)
	2020	1064	41 (4.6)	757 (84.0)	65 (7.2)	38 (4.2)
	2021	1015	24 (2.5)	788 (83.3)	90 (9.5)	44 (4.7)
	2022	1083	24 (2.2)	888 (82.7)	105 (9.8)	57 (5.3)

* Number with non-missing data.

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

1.8a Body Mass Index (BMI) in adults (20 years and over) N=5627

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

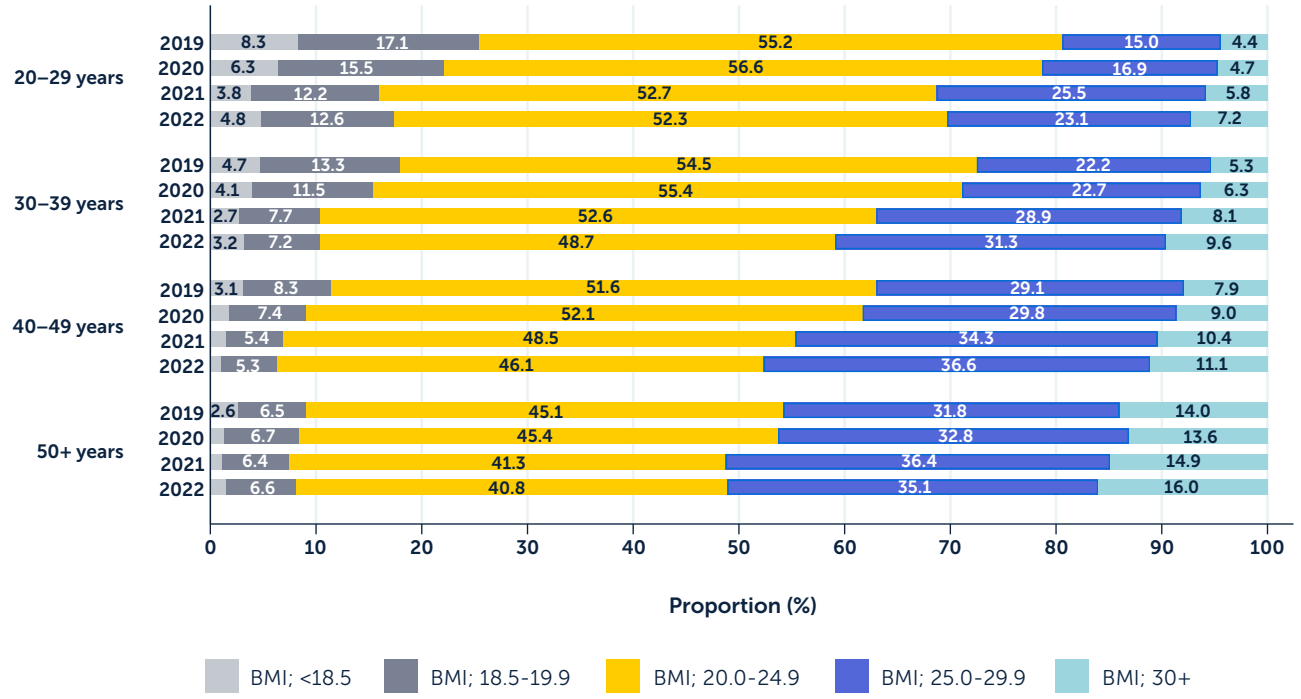


	Overall			Female			Male		
Age	n	Median	IQR	n	Median	IQR	n	Median	IQR
20-23	829	22.3	20.2-25.3	426	22.1	20.1-25.4	403	22.4	20.4-25.0
24-27	902	23.3	21.0-25.9	449	22.9	21.0-25.5	453	23.8	21.0-26.2
28-31	837	23.7	21.5-26.3	374	23.5	21.2-26.2	463	23.8	21.8-26.5
32-35	778	24.1	21.9-26.7	344	23.3	21.4-26.1	434	24.5	22.6-27.0
36-39	584	24.5	22.0-27.1	252	23.2	21.2-26.2	332	25.0	23.1-27.3
40-43	516	24.6	22.1-27.2	225	23.8	21.4-27.1	291	25.2	22.7-27.2
44-47	297	24.8	22.0-27.5	114	22.9	21.6-25.7	183	25.4	23.5-28.2
48-51	251	25.1	23.2-27.5	110	24.7	22.3-27.6	141	25.5	23.6-27.4
52-55	187	25.5	22.3-28.4	78	24.9	21.5-28.4	109	25.7	22.8-28.3
56-59	125	26.2	23.4-29.4	53	26.7	22.4-30.7	72	26.0	24.2-28.8
60-63	80	24.0	22.7-27.4	33	23.4	21.9-26.6	47	24.2	23.2-28.0
64-67	54	25.4	22.0-29.7	21	23.6	20.8-27.8	33	26.4	22.4-29.9
68+	72	25.5	22.1-28.9	43	24.2	20.5-30.1	29	26.1	24.0-27.8
Overall	**5512***	**23.9**	**21.6-26.8**	**2522**	**23.3**	**21.1-26.3**	**2990**	**24.3**	**22.0-26.9**

* Number with non-missing data.

1.8b Body Mass Index (BMI) in adults for 2019–2022

The following graph shows the change in the proportion of people in each BMI group from 2019 to 2022.



			BMI category by age and year : n*(%)				
Age group	Year	Total number of people in each age group	BMI; <18.5	BMI; 18.5-19.9	BMI; 20.0-24.9	BMI; 25.0-29.9	BMI; 30+
20-29 years	2019	2353	195 (8.3)	401 (17.1)	1293 (55.1)	352 (15.0)	104 (4.4)
	2020	2239	115 (6.3)	281 (15.5)	1027 (56.6)	306 (16.9)	86 (4.7)
	2021	2261	75 (3.8)	240 (12.2)	1033 (52.7)	499 (25.5)	113 (5.8)
	2022	2202	104 (4.8)	271 (12.6)	1126 (52.3)	498 (23.1)	154 (7.2)
30-39 years							
	2019	1656	78 (4.7)	219 (13.3)	900 (54.5)	366 (22.2)	87 (5.3)
	2020	1643	54 (4.1)	153 (11.5)	736 (55.4)	302 (22.7)	84 (6.3)
	2021	1776	42 (2.7)	118 (7.7)	809 (52.6)	444 (28.9)	124 (8.1)
	2022	1816	57 (3.2)	128 (7.2)	865 (48.7)	556 (31.3)	171 (9.6)
40-49 years							
	2019	774	24 (3.1)	64 (8.3)	398 (51.6)	224 (29.1)	61 (7.9)
	2020	807	11 (1.7)	48 (7.4)	336 (52.1)	192 (29.8)	58 (9.0)
	2021	856	11 (1.5)	40 (5.4)	360 (48.5)	255 (34.3)	77 (10.4)
	2022	939	9 (1.0)	49 (5.3)	427 (46.1)	339 (36.6)	103 (11.1)
50+ years							
	2019	509	13 (2.6)	33 (6.5)	228 (45.1)	161 (31.8)	71 (14.0)
	2020	516	6 (1.5)	27 (6.7)	183 (45.4)	132 (32.8)	55 (13.6)
	2021	619	6 (1.1)	35 (6.4)	227 (41.3)	200 (36.4)	82 (14.9)
	2022	670	10 (1.5)	43 (6.6)	267 (40.8)	230 (35.1)	105 (16.0)

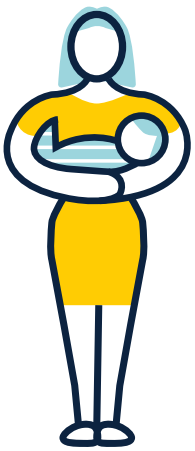
1.9 Education and employment in adults (16 years and over) N=6445

The following table shows how people with CF reported their education and employment status in 2022.

	2019	2020	2021	2022		
	Overall	Overall	Overall	Overall	Male	Female
Number of patients	6104	6012	6297	6445	3446	2999
Number who completed questionnaire; n(%)	6103 (100.0)	5968 (99.3)	6296 (100.0)	6442 (100.0)	3444 (99.9)	2998 (100.0)
Full-time employment; n(%)	2048 (33.6)	1975 (32.9)	2097 (33.3)	2228 (34.6)	1472 (42.7)	756 (25.2)
Part-time employment; n(%)	958 (15.7)	894 (14.9)	915 (14.5)	981 (15.2)	362 (10.5)	619 (20.6)
Student; n(%)	969 (15.9)	1015 (16.9)	1061 (16.8)	1046 (16.2)	498 (14.5)	548 (18.3)
Homemaker; n(%)	231 (3.8)	200 (3.3)	251 (4.0)	249 (3.9)	36 (1.0)	213 (7.1)
Unemployed; n(%)	825 (13.5)	847 (14.1)	791 (12.6)	767 (11.9)	446 (12.9)	321 (10.7)
Disabled; n(%)	327 (5.4)	274 (4.6)	255 (4.0)	228 (3.5)	124 (3.6)	104 (3.5)
Retired; n(%)	145 (2.4)	139 (2.3)	162 (2.6)	170 (2.6)	94 (2.7)	76 (2.5)
Volunteer; n(%)	8 (0.1)	11 (0.2)	12 (0.2)	14 (0.2)	7 (0.2)	7 (0.2)
Unknown entered; n(%)	592 (9.7)	613 (10.2)	752 (11.9)	759 (11.8)	405 (11.8)	354 (11.8)
No. in work or study; n(%)	3975 (65.1)	3884 (65.1)	4073 (64.7)	4255 (66.1)	2332 (67.7)	1923 (64.1)

1.10 Parenthood

	2019	2020	2021	2022
Women with CF who had babies; n	58	56	103	140
Men with CF who became fathers; n	45	44	30	33

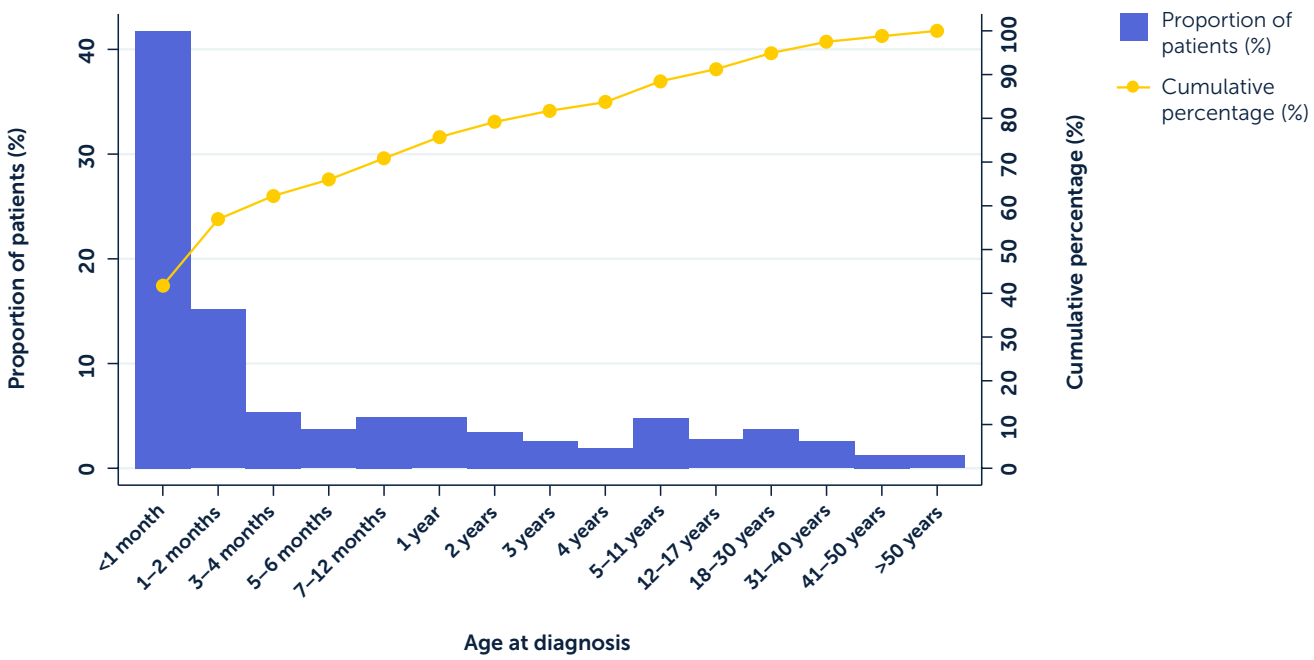


140 women with CF had babies in 2022

33 men with CF became fathers in 2022

Diagnosis of cystic fibrosis

1.11 Age at diagnosis
N=11148



The median age at diagnosis for patients aged under 16 in 2022 is **21 days**.

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

A total of **162 (73%)** out of 233 patients diagnosed in 2022 were identified by newborn screening (including those without complete data).

1,063 (9.5%) of adults with CF in the Registry in 2022 were diagnosed at age 16 or over.

In 2022, **33** people aged 16 or over were newly diagnosed with cystic fibrosis.

1.12 Mode of presentation

The following tables show the top five most frequent modes of presentation for those diagnosed between 2012–2022 and those born between 2012–2022, excluding those recorded as being diagnosed through newborn screening (NBS) or genotype. Patients may present with multiple symptoms so percentages may not add to 100.

	All patients diagnosed 2012-2022	Age <16 at diagnosis	Age ≥16 at diagnosis
Total patients	**2940**	**2510**	**430**
Number diagnosed by newborn screening	2072	2072	0
Total non-NBS	868	438	430

Presentation type	All patients diagnosed 2012-2022	Age<16 at diagnosis	Age >=16 at diagnosis
Persistent or acute respiratory infection	268 (30.9)	114 (26.0)	154 (35.8)
Meconium ileus	137 (15.8)	137 (31.3)	0 (0.0)
Family history	130 (15.0)	81 (18.5)	49 (11.4)
Bronchiectasis	110 (12.7)	10 (2.3)	100 (23.3)
Failure to thrive/ malnutrition	73 (8.4)	66 (15.1)	7 (1.6)

	All patients born 2012-2022
Total patients	**2344**
Number diagnosed by newborn screening or genotype	2017
Total non-NBS or genotype	327

Presentation type	All patients born 2012-2022
Meconium ileus	136 (41.6)
Family history	66 (20.2)
Persistent or acute respiratory infection	52 (15.9)
Failure to thrive/malnutrition	45 (13.8)
Prenatal	43 (13.1)

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

** Redacted to adhere to statistical disclosure guidelines.

Lung health

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

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

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

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

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

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

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

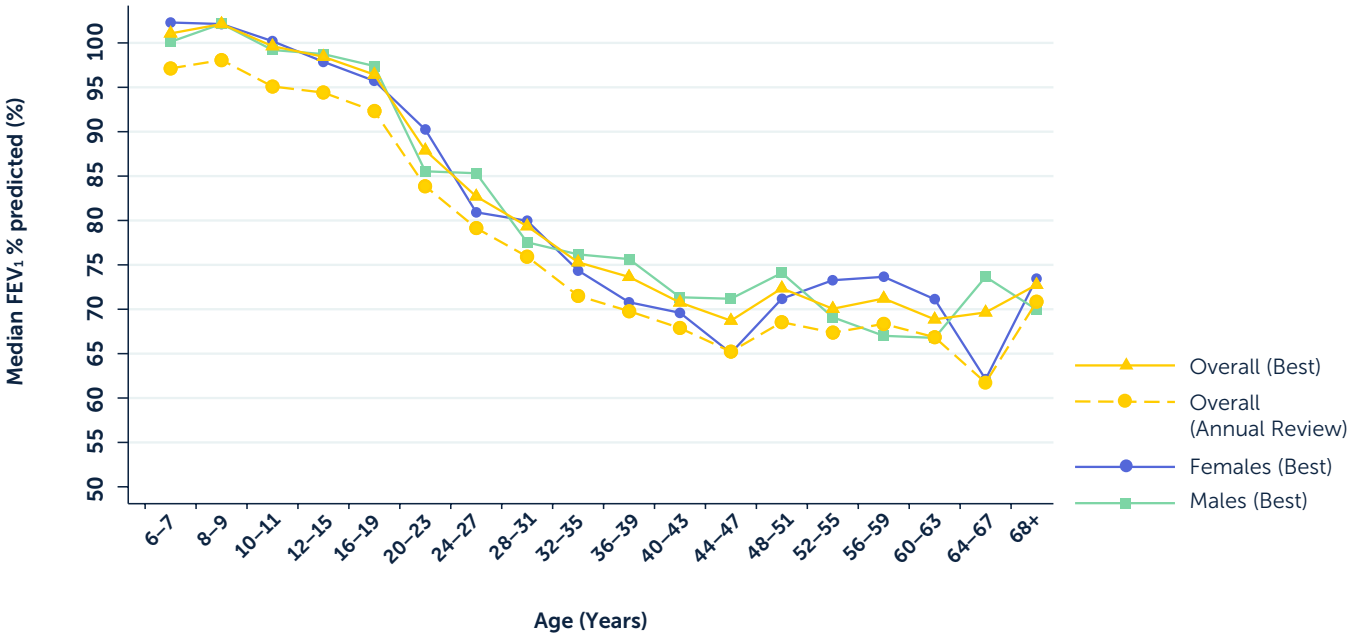
1.13 Annual review FEV_1% predicted (GLI equations) in patients age six years and older who have not had a lung transplant
N=8813

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 (yrs)	n	Median	IQR	n	Median	IQR	n	Median	IQR
6-7	425	97.1	88.0-106.9	198	97.8	89.5-107.8	227	96.4	87.4-106.5
8-9	449	98.1	89.4-106.7	209	98.0	89.9-105.8	240	98.3	89.2-106.9
10-11	528	95.1	86.8-104.2	258	95.6	86.8-105.1	270	94.6	86.9-103.2
12-15	960	94.4	83.9-104.1	457	94.0	83.4-103.8	503	94.8	84.0-104.4
16-19	737	92.3	81.5-100.9	386	91.3	79.5-99.9	351	93.5	82.0-102.8
20-23	757	83.9	67.4-96.6	393	85.0	70.2-96.6	364	82.2	64.7-96.6
24-27	820	79.1	62.1-93.9	409	76.1	58.3-92.3	411	82.1	65.0-94.3
28-31	736	75.9	56.8-90.8	342	76.6	56.5-90.8	394	75.7	57.2-90.7
32-35	670	71.5	54.7-86.6	299	69.9	54.7-85.2	371	72.2	54.6-87.1
36-39	496	69.8	50.1-85.6	209	66.9	49.1-83.0	287	72.2	52.3-88.2
40-43	441	67.9	49.5-85.9	192	65.7	49.2-85.8	249	69.8	50.9-87.3
44-47	265	65.2	48.1-83.1	100	59.0	46.1-80.5	165	68.4	50.5-83.4
48-51	214	68.5	49.0-86.0	92	67.1	46.3-81.3	122	70.6	51.7-87.6
52-55	156	67.4	47.0-86.8	65	70.1	51.2-91.1	91	66.2	43.0-82.9
56-59	109	68.3	50.3-83.6	47	71.3	56.2-83.1	62	65.6	46.3-83.7
60-63	73	66.8	47.8-82.4	32	68.0	51.6-80.6	41	62.1	44.0-85.9
64-67	53	61.7	43.2-85.3	20	59.6	39.7-70.1	33	70.9	48.2-90.4
68+	62	70.8	56.3-88.3	36	72.2	59.2-89.3	26	68.0	49.8-81.5
<16	2362	95.9	86.4-105.2	1122	96.0	86.7-105.2	1240	95.7	86.1-105.2
≥16	5589	77.3	57.6-92.3	2622	77.5	56.9-91.9	2967	77.1	58.0-92.9
<18	2735	95.5	86.0-104.8	1309	95.8	86.2-104.7	1426	95.4	85.9-104.9
≥18	5216	75.9	56.1-91.1	2435	75.7	55.5-90.5	2781	76.0	56.5-91.7
Overall	**7951***	**84.8**	**65.6-97.9**	**3744**	**84.8**	**65.6-97.6**	**4207**	**84.8**	**65.5-98.1**

* Number with non-missing data.

1.14 Best* FEV_1% predicted (GLI equations) in patients aged six years and older who have not had a lung transplant N=8831

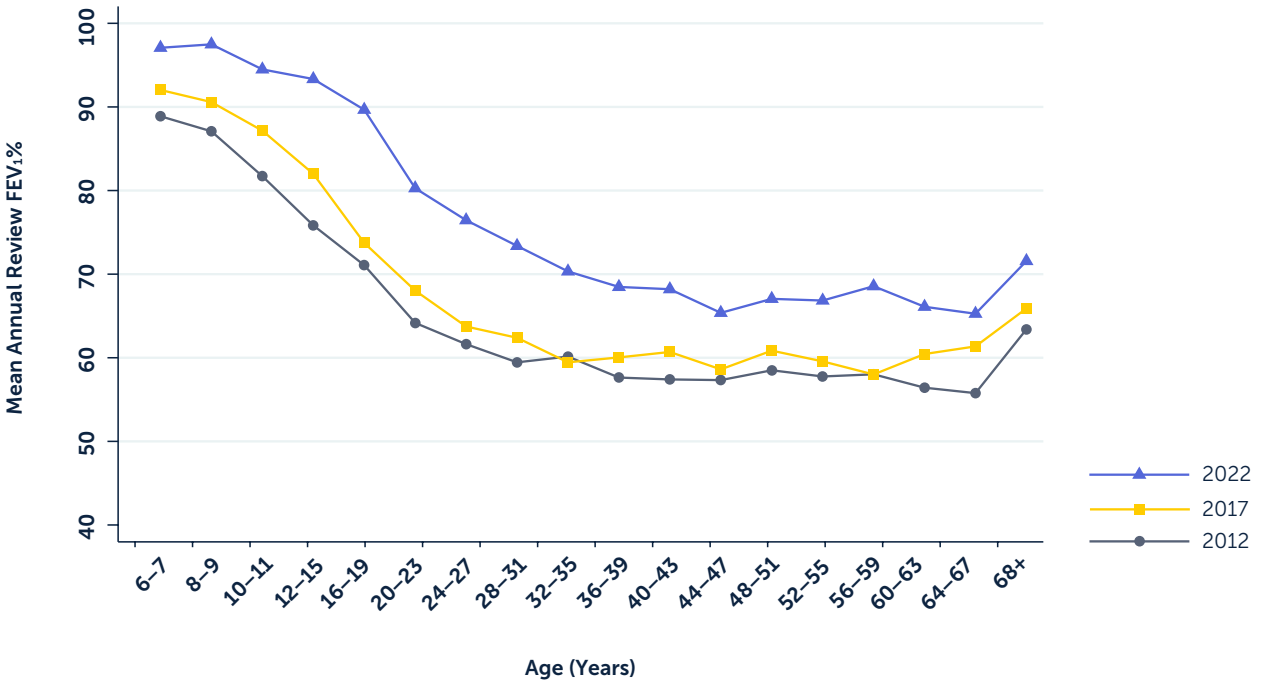


	Overall			Female			Male		
Age (yrs)	n	Median	IQR	n	Median	IQR	n	Median	IQR
6-7	464	101.1	92.5-110.4	219	102.3	93.4-111.8	245	100.1	90.7-109.4
8-9	496	102.1	93.9-110.0	230	102.1	93.8-109.7	266	102.1	93.9-110.5
10-11	562	99.6	90.9-107.1	276	100.2	91.2-107.4	286	99.2	90.6-106.1
12-15	1026	98.4	89.1-107.9	491	97.9	88.0-107.6	535	98.7	89.7-108.1
16-19	787	96.4	85.2-105.5	410	95.7	83.8-104.3	377	97.4	87.1-106.4
20-23	819	87.9	72.1-100.5	418	90.2	75.4-100.9	401	85.5	69.3-100.0
24-27	872	82.7	65.6-96.9	437	80.9	64.1-96.5	435	85.3	67.8-98.0
28-31	800	79.3	60.7-93.5	364	80.0	61.3-92.5	436	77.5	60.4-94.4
32-35	721	75.3	57.7-89.8	318	74.4	58.7-88.0	403	76.2	56.3-91.3
36-39	534	73.6	54.3-89.0	223	70.8	50.5-87.0	311	75.7	56.3-91.0
40-43	472	70.8	53.3-89.2	207	69.6	54.2-87.8	265	71.3	52.7-90.1
44-47	276	68.7	51.5-86.1	101	65.1	49.7-83.7	175	71.2	53.7-88.7
48-51	224	72.4	53.5-87.2	96	71.2	50.8-85.8	128	74.1	55.9-89.1
52-55	168	70.1	53.0-88.8	70	73.3	55.9-94.1	98	69.1	48.4-86.8
56-59	115	71.2	50.2-87.0	50	73.7	60.1-84.5	65	67.0	49.5-90.2
60-63	74	68.9	47.4-83.4	32	71.1	54.0-80.6	42	66.8	44.7-89.9
64-67	53	69.6	48.2-89.1	20	62.1	45.7-76.9	33	73.7	50.1-91.4
68+	70	72.8	55.5-88.7	41	73.5	59.0-92.5	29	70.0	50.3-83.8
<16	2548	100.0	91.1-108.6	1216	100.3	91.2-109.1	1332	99.8	91.1-108.4
≥16	5985	81.2	61.2-95.9	2787	81.5	61.2-95.8	3198	80.8	61.2-96.2
<18	2953	99.6	90.7-108.2	1420	99.9	90.4-108.3	1533	99.5	90.8-108.0
≥18	5580	79.3	59.7-94.7	2583	79.6	60.0-94.4	2997	78.7	59.6-95.1
Overall	**8533****	**88.9**	**69.7-101.4**	**4003**	**88.9**	**70.0-101.5**	**4530**	**88.9**	**69.4-101.4**

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

1.15 Annual review FEV_1% predicted (GLI equations) over time in patients aged six years and older who have not had a lung transplant N=8831 in 2022, N=8168 in 2017, N=7071 in 2012

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 2022 compares to Registry data from 2012 and 2017.



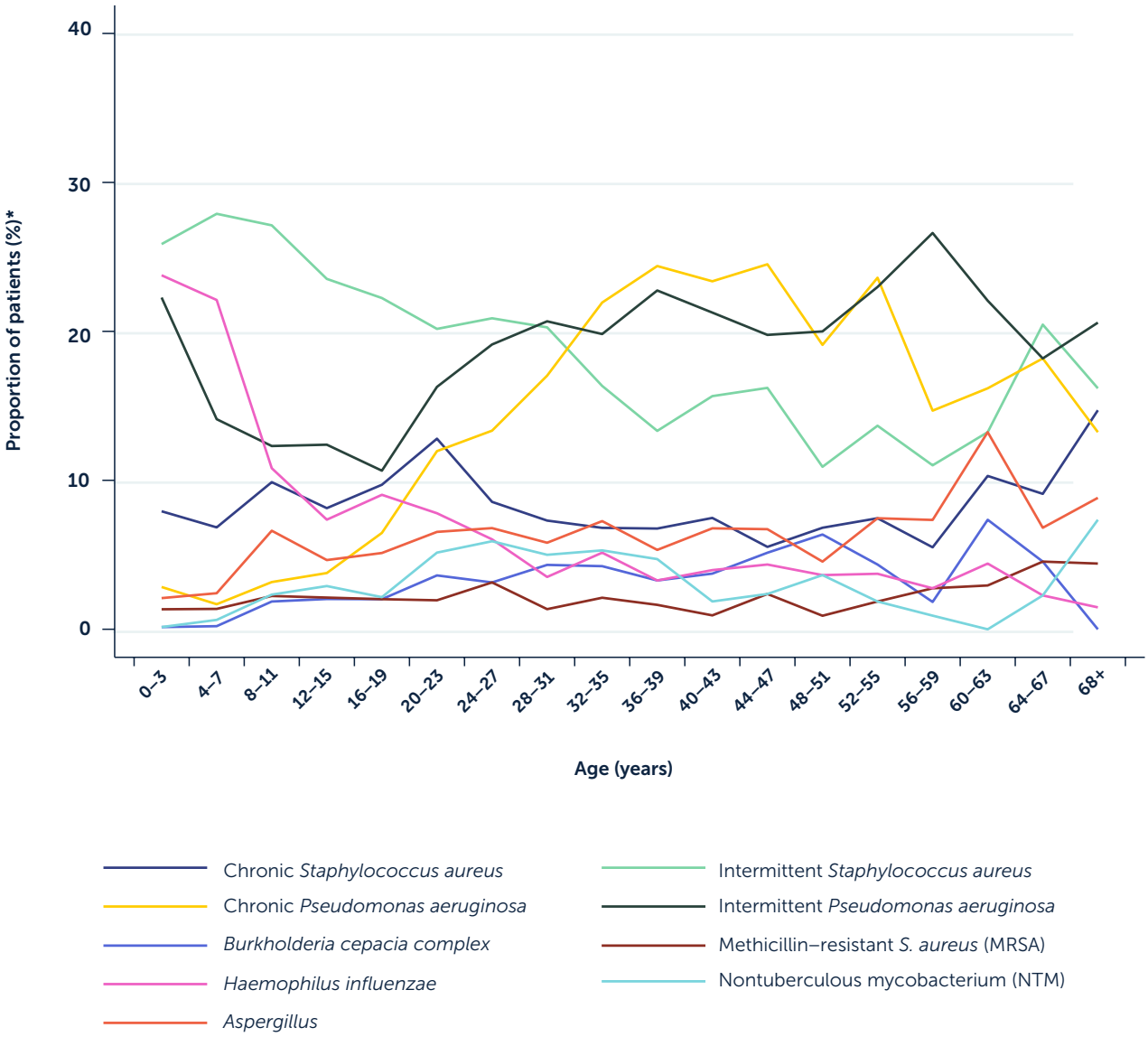
	2012		2017		2022		
Age (years)	n	FEV_1% :Mean (SD)	n	FEV_1% :Mean (SD)	n	FEV_1% :Mean (SD)	p-values (t-test*)
6-7	375	88.9 (15.9)	526	92.0 (16.0)	425	97.1 (15.8)	<0.001
8-9	399	87.1 (15.8)	511	90.6 (15.9)	449	97.5 (14.7)	<0.001
10-11	392	81.7 (16.5)	488	87.2 (15.7)	528	94.5 (15.5)	<0.001
12-15	914	75.8 (18.3)	862	82.0 (17.8)	960	93.3 (16.0)	<0.001
16-19	920	71.1 (21.8)	887	73.7 (21.5)	737	89.7 (17.9)	<0.001
20-23	915	64.2 (23.6)	945	68.0 (23.3)	757	80.3 (22.4)	<0.001
24-27	747	61.6 (23.8)	860	63.7 (23.0)	820	76.5 (22.6)	<0.001
28-31	600	59.5 (22.8)	725	62.4 (23.8)	736	73.4 (23.0)	<0.001
32-35	394	60.1 (23.3)	609	59.5 (23.7)	670	70.3 (22.5)	<0.001
36-39	256	57.6 (22.5)	436	60.0 (24.3)	496	68.5 (22.8)	<0.001
40-43	241	57.4 (22.7)	276	60.7 (24.0)	441	68.2 (23.8)	<0.001
44-47	167	57.3 (25.5)	237	58.6 (23.4)	265	65.4 (23.7)	0.001
48-51	111	58.5 (23.8)	172	60.9 (25.4)	214	67.0 (22.9)	0.012
52-55	66	57.8 (26.6)	119	59.6 (25.4)	156	66.8 (24.1)	0.016
56-59	37	58.0 (22.3)	68	58.0 (24.1)	109	68.6 (23.6)	0.005
60-63	18	56.4 (26.9)	45	60.5 (23.1)	73	66.1 (22.8)	0.196
64-67	17	55.8 (23.1)	28	61.4 (22.6)	53	65.3 (25.4)	0.497
68+	17	63.4 (25.6)	41	65.9 (28.3)	62	71.6 (24.5)	0.283
<16	2080	81.5 (18.0)	2387	87.1 (17.1)	2362	95.1 (15.7)	-
≥16	4506	62.8 (23.6)	5448	64.5 (23.9)	5589	74.6 (23.5)	-
<18	2561	79.8 (18.8)	2802	85.5 (17.9)	2735	94.6 (15.8)	-
≥18	4025	61.7 (23.7)	5033	63.5 (23.9)	5216	73.4 (23.5)	-

* T-test comparing 2022 with 2017.

Lung infections

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

1.16 Lung infections in 2022
N=9221*



* 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 2022 (contd.)
<16 years N=3806, ≥16 years N=6445

	Paediatric Age Range (Years)				Overall
	0-3	4-7	8-11	12-15	Paediatric (<16 years)
Number in age range	689	971	1093	1053	3806
Number who had culture taken*	669	950	1073	1033	3725
Chronic *S. aureus* n(%)	53 (7.9)	65 (6.8)	106 (9.9)	84 (8.1)	308 (8.3)
Intermittent *S. aureus* n(%)	173 (25.9)	265 (27.9)	291 (27.1)	243 (23.5)	972 (26.1)
Chronic *P. aeruginosa* n(%)	19 (2.8)	16 (1.7)	34 (3.2)	39 (3.8)	108 (2.9)
Intermittent *P. aeruginosa* n(%)	149 (22.3)	134 (14.1)	132 (12.3)	128 (12.4)	543 (14.6)
B. cepacia complex n(%)	<5	<5	20 (1.9)	21 (2.0)	44 (1.2)
B. cenocepacia n(%)	<5	<5	8 (0.7)	7 (0.7)	17 (0.5)
B. multivorans n(%)	<5	<5	5 (0.5)	6 (0.6)	12 (0.3)
B. other cepacia n(%)	<5	<5	<5	<5	7 (0.2)
MRSA n(%)	9 (1.3)	13 (1.4)	24 (2.2)	22 (2.1)	68 (1.8)
H. influenza n(%)	159 (23.8)	210 (22.1)	116 (10.8)	76 (7.4)	561 (15.1)
NTM n(%)	<5	6 (0.6)	25 (2.3)	30 (2.9)	-**
Aspergillus fumigatus n(%)	14 (2.1)	23 (2.4)	71 (6.6)	48 (4.6)	156 (4.2)

Infections in this table reflect those grown in the 12 months prior to the 2022 annual review.
The UK CF Registry definition of 'chronic' is three or more isolates in the last 12 months.

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

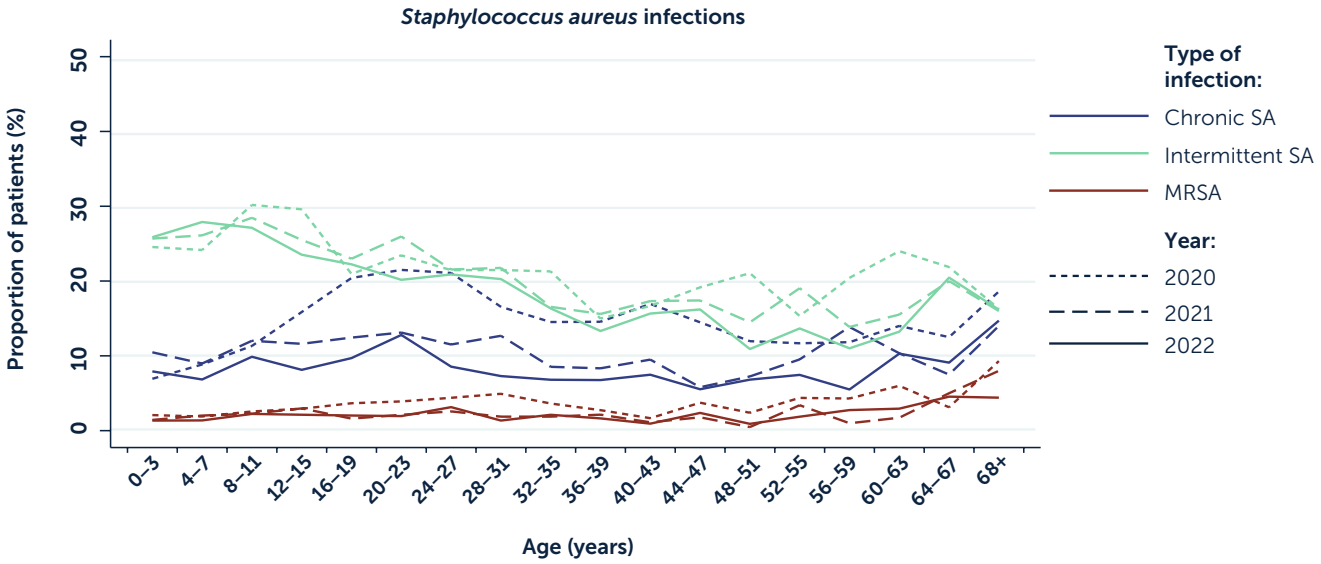
Lung infections in 2022 (contd.)
<16 years N=3806, ≥16 years N=6445

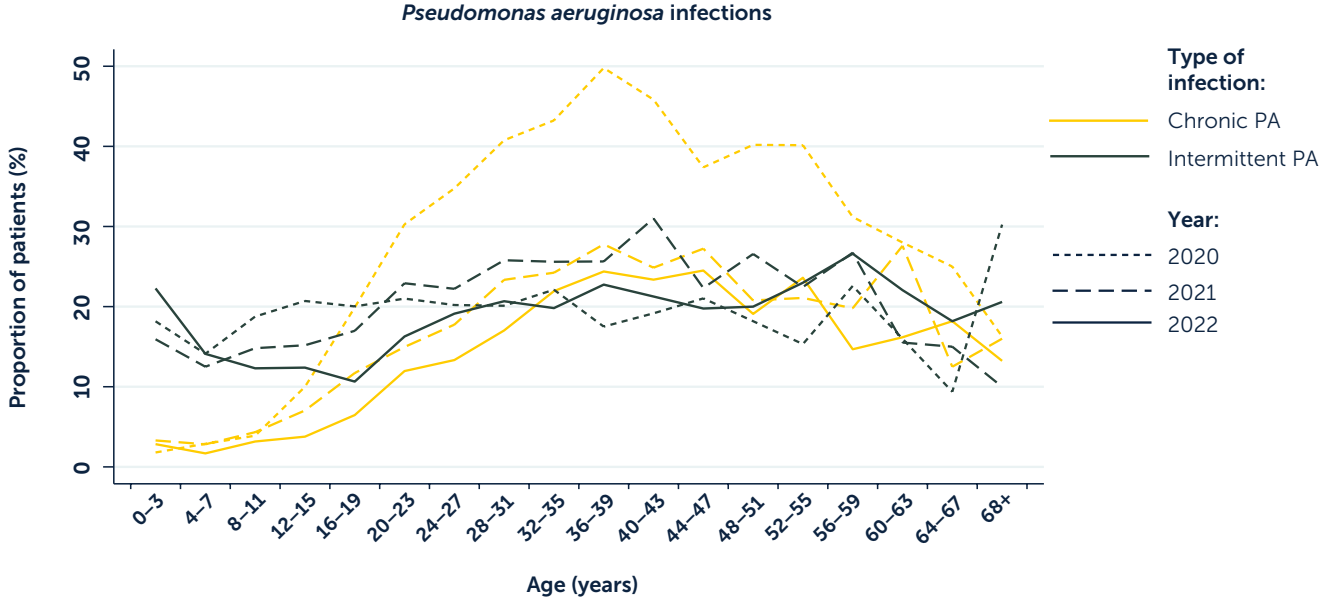
	Adult Age Range (Years)							Overall
	16-19	20-23	24-27	28-31	32-35	36-39	40-43	Adults (≥16 years)
Number in age range	818	853	919	859	793	594	524	6445
Number who had culture taken*	742	719	795	740	661	488	428	5496
Chronic *S. aureus* n(%)	72 (9.7)	92 (12.8)	68 (8.6)	54 (7.3)	45 (6.8)	33 (6.8)	32 (7.5)	464 (8.4)
Intermittent *S. aureus* n(%)	165 (22.2)	145 (20.2)	166 (20.9)	150 (20.3)	108 (16.3)	65 (13.3)	67 (15.7)	994 (18.1)
Chronic *P. aeruginosa* n(%)	48 (6.5)	86 (12.0)	106 (13.3)	126 (17.0)	145 (21.9)	119 (24.4)	100 (23.4)	916 (16.7)
Intermittent *P. aeruginosa* n(%)	79 (10.6)	117 (16.3)	152 (19.1)	153 (20.7)	131 (19.8)	111 (22.7)	91 (21.3)	1031 (18.8)
B. cepacia complex n(%)	15 (2.0)	26 (3.6)	25 (3.1)	32 (4.3)	28 (4.2)	16 (3.3)	16 (3.7)	201 (3.7)
B. cenocepacia n(%)	<5	8 (1.1)	8 (1.0)	10 (1.4)	7 (1.1)	6 (1.2)	5 (1.2)	61 (1.1)
B. multivorans n(%)	<5	10 (1.4)	9 (1.1)	15 (2.0)	18 (2.7)	9 (1.8)	8 (1.9)	88 (1.6)
B. other cepacia n(%)	7 (0.9)	<5	<5	6 (0.8)	<5	<5	<5	35 (0.6)
MRSA n(%)	15 (2.0)	14 (1.9)	25 (3.1)	10 (1.4)	14 (2.1)	8 (1.6)	<5	111 (2.0)
H. influenza n(%)	67 (9.0)	56 (7.8)	48 (6.0)	26 (3.5)	34 (5.1)	16 (3.3)	17 (4.0)	297 (5.4)
NTM n(%)	16 (2.2)	37 (5.1)	47 (5.9)	37 (5.0)	35 (5.3)	23 (4.7)	8 (1.9)	227 (4.1)
Aspergillus fumigatus n(%)	38 (5.1)	47 (6.5)	54 (6.8)	43 (5.8)	48 (7.3)	26 (5.3)	29 (6.8)	350 (6.4)

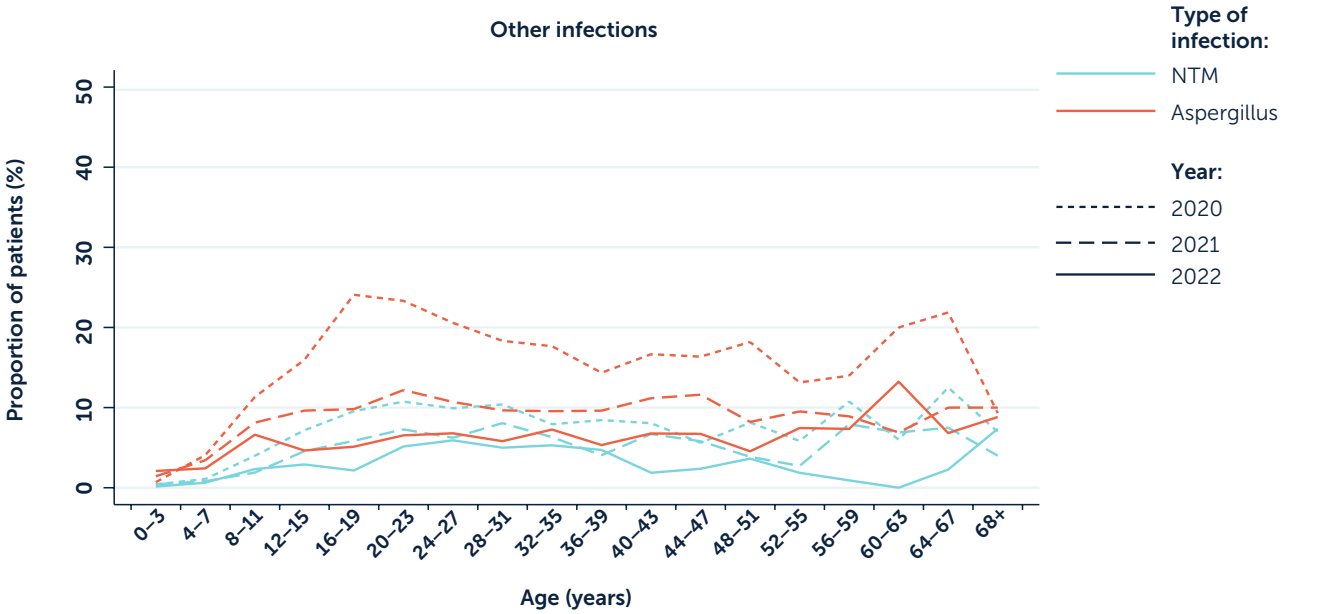
	Adult Age Range (Years)							Overall
	44-47	48-51	52-55	56-59	60-63	64-67	68+	Adults (≥16 years)
Number in age range	300	254	190	130	82	54	75	6445
Number who had culture taken*	253	220	161	109	68	44	68	5496
Chronic *S. aureus* n(%)	14 (5.5)	15 (6.8)	12 (7.5)	6 (5.5)	7 (10.3)	<5	10 (14.7)	-**
Intermittent *S. aureus* n(%)	41 (16.2)	24 (10.9)	22 (13.7)	12 (11.0)	9 (13.2)	9 (20.5)	11 (16.2)	994 (18.1)
Chronic *P. aeruginosa* n(%)	62 (24.5)	42 (19.1)	38 (23.6)	16 (14.7)	11 (16.2)	8 (18.2)	9 (13.2)	916 (16.7)
Intermittent *P. aeruginosa* n(%)	50 (19.8)	44 (20.0)	37 (23.0)	29 (26.6)	15 (22.1)	8 (18.2)	14 (20.6)	1031 (18.8)
B. cepacia complex n(%)	13 (5.1)	14 (6.4)	7 (4.3)	<5	5 (7.4)	<5	<5	201 (3.7)
B. cenocepacia n(%)	<5	5 (2.3)	<5	<5	<5	<5	<5	61 (1.1)
B. multivorans n(%)	6 (2.4)	5 (2.3)	<5	<5	<5	<5	<5	88 (1.6)
B. other cepacia n(%)	<5	<5	<5	<5	<5	<5	<5	35 (0.6)
MRSA n(%)	6 (2.4)	<5	<5	<5	<5	<5	<5	111 (2.0)
H. influenza n(%)	11 (4.3)	8 (3.6)	6 (3.7)	<5	<5	<5	<5	297 (5.4)
NTM n(%)	6 (2.4)	8 (3.6)	<5	<5	<5	<5	5 (7.4)	227 (4.1)
Aspergillus fumigatus n(%)	17 (6.7)	10 (4.5)	12 (7.5)	8 (7.3)	9 (13.2)	<5	6 (8.8)	-**

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

1.18 Lung infections 2020–2022







1.19 Respiratory culture sample type

Overall	2020	2021	2022
Number of people with an annual review (n)	9922	10175	10251
Number of people with at least 3 samples of any type taken n(%)*	7563 (76.2)	6082 (59.8)	6442 (62.8)
Number of people with at least 1 sample of any type taken n(%)*	9368 (94.4)	8921 (87.7)	9240 (90.1)
Sample type[1]**			
Sputum; n(%)	6250 (66.7)	5196 (58.2)	5319 (57.6)
Cough; n(%)	6021 (64.3)	6048 (67.8)	6434 (69.6)
Bronchoalveolar lavage; n(%)	528 (5.6)	224 (2.5)	264 (2.9)
Age <16 years	**2020**	**2021**	**2022**
Number of people with an annual review (n)	3910	3878	3806
Number of people with at least 3 samples of any type taken n(%)*	3626 (92.7)	3473 (89.6)	3510 (92.2)
Number of people with at least 1 sample of any type taken n(%)*	3851 (98.5)	3808 (98.2)	3730 (98.0)
Sample type[1]**			
Sputum; n(%)	1437 (37.3)	1125 (29.5)	1129 (30.3)
Cough; n(%)	3696 (96.0)	3704 (97.3)	3666 (98.3)
Bronchoalveolar lavage; n(%)	254 (6.6)	172 (4.5)	181 (4.9)
Age>=16 years	**2020**	**2021**	**2022**
Number of people with an annual review (n)	6012	6297	6445
Number of people with at least 3 samples of any type taken n(%)*	3937 (65.5)	2609 (41.4)	2932 (45.5)
Number of people with at least 1 sample of any type taken n(%)*	5517 (91.8)	5113 (81.2)	5510 (85.5)
Sample type**			
Sputum; n(%)	4813 (87.2)	4071 (79.6)	4190 (76.0)
Cough; n(%)	2325 (42.1)	2344 (45.8)	2768 (50.2)
Bronchoalveolar lavage; n(%)	274 (5.0)	52 (1.0)	83 (1.5)

* % 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.

	2020	2021	2022
Number with annual review	**(n=9922)**	**(n=10175)**	**(n=10251)**
NTM Prevalence; n(%)	620 (6.2*)	397 (3.9)	289 (3.1)
On NTM treatment in the given year; n (% of NTM prevalence in given year)	326 (52.6)	231 (58.1)	153 (52.9)
NTM Incidence[1]	226 (2.5)	154 (1.7)	147 (1.5)
M. abscessus prevalence	361 (3.6*)	216 (2.1)	90 (0.9)
M. abscessus incidence[2]	103 (1.1)	58 (0.6)	29 (0.3)

* correction for 2020 data

[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

1.21 COVID-19* infection in 2022

COVID-19 management and outcomes for people with CF infected with COVID-19 during the calendar year of 2022 are described below. Information is stratified by sex, ethnicity, age, organ transplant status and Best FEV_1% prior to catching COVID-19.

		COVID-19 Management			Outcomes
	Total	Symptomatic	IV antibiotics	Oral antibiotics	Hospitalised
Overall; n(%)					
All cases	2159 (100.0)	1685 (78.0)	69 (3.2)	564 (26.1)	83 (3.8)
Sex; n(%)					
Female	1114 (51.6)	875 (78.5)	37 (3.3)	301 (27.0)	39 (3.5)
Male	1045 (48.4)	810 (77.5)	32 (3.1)	263 (25.2)	44 (4.2)
Ethnicity; n(%)					
White	2022 (93.7)	1586 (78.4)	60 (3.0)	529 (26.2)	70 (3.5)
Non-White	73 (3.4)	53 (72.6)	8 (11.0)	19 (26.0)	10 (13.7)
Unknown	64 (3.0)	46 (71.9)	1 (1.6)	16 (25.0)	3 (4.7)
Age; n(%)					
Under 16	675 (31.3)	521 (77.2)	13 (1.9)	161 (23.9)	23 (3.4)
>= 16	1484 (68.7)	1164 (78.4)	56 (3.8)	403 (27.2)	60 (4.0)
Transplants; n(%)					
No	2100 (97.3)	1641 (78.1)	60 (2.9)	557 (26.5)	74 (3.5)
Yes	59 (2.7)	44 (74.6)	9 (15.3)	7 (11.9)	9 (15.3)
****BestFEV; n(%)**					
<40	92 (4.3)	75 (81.5)	14 (15.2)	21 (22.8)	12 (13.0)
40-70	441 (20.4)	354 (80.3)	19 (4.3)	129 (29.3)	21 (4.8)
>70	1626 (75.3)	1256 (77.2)	36 (2.2)	414 (25.5)	50 (3.1)

Of the 83 patients hospitalised after a positive test for COVID-19, 14 of them were also given oxygen.

* COVID-19 cases confirmed with positive PCR or lateral flow tests.

** Patients who had a lung transplant were excluded.

Complications

1.22 Complications in 2022

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

Complications	Overall	<16 years	≥16 years
Respiratory related			
Nasal polyps requiring surgery	414 (4.0)	114 (3.0)	300 (4.7)
Sinus disease	810 (7.9)	41 (1.1)	769 (11.9)
Asthma	663 (6.5)	137 (3.6)	526 (8.2)
ABPA	515 (5.0)	88 (2.3)	427 (6.6)
Any haemoptysis	-*	<5	144 (2.2)
Massive haemoptysis	10 (0.1)	0	10 (0.2)
Pneumothorax requiring chest tube	13 (0.1)	0	13 (0.2)
Cardiac complications			
Tachyarrhythmia	-*	<5	7 (0.1)
Bradycardia	-*	0	<5
Cardiac arrest	-*	0	<5
Cardiomyopathy	10 (0.1)	0	10 (0.2)
Congenital heart disease	18 (0.2)	8 (0.2)	10 (0.2)
Heart failure	9 (0.1)	0	9 (0.1)
Ischaemic heart disease	7 (0.1)	0	7 (0.1)
Valvular disease	-*	0	<5
Other	77 (0.8)	11 (0.3)	66 (1.0)
Pancreas and hepatobiliary disease			
Raised liver enzymes	1425 (13.9)	403 (10.6)	1022 (15.9)
Liver disease	1828 (17.8)	391 (10.3)	1437 (22.3)
Cirrhosis with no portal hypertension	85 (0.8)	9 (0.2)	76 (1.2)
Cirrhosis with portal hypertension	150 (1.5)	25 (0.7)	125 (1.9)
Gall bladder disease requiring surgery	246 (2.4)	42 (1.1)	204 (3.2)
Pancreatitis	66 (0.6)	8 (0.2)	58 (0.9)
Upper gastrointestinal (GI)			
Gastro-oesphageal reflux disease (GORD)	1665 (16.2)	215 (5.6)	1450 (22.5)
Peptic ulcer	-*	0	<5
GI bleed (varices as source)	-*	<5	13 (0.2)
GI bleed (non varices as source)	-*	<5	13 (0.2)
Lower gastrointestinal			
Intestinal obstruction	52 (0.5)	15 (0.4)	37 (0.6)
DIOS	453 (4.4)	83 (2.2)	370 (5.7)
Fibrosing colonopathy / colonic stricture	-*	0	<5
Rectal prolapse	13 (0.1)	5 (0.1)	8 (0.1)
Renal			
Kidney stones	165 (1.6)	5 (0.1)	160 (2.5)
Renal failure	92 (0.9)	0	92 (1.4)
Musculoskeletal			
Arthritis	-*	<5	108 (1.7)
Arthropathy	257 (2.5)	6 (0.2)	251 (3.9)
Bone fracture	56 (0.5)	19 (0.5)	37 (0.6)
Osteopenia	1065 (10.4)	10 (0.3)	1055 (16.4)
Osteoporosis	-*	<5	480 (7.4)
Other			
Cancer confirmed by histology	-*	<5	30 (0.5)
Port inserted or replaced	168 (1.6)	53 (1.4)	115 (1.8)
Depression	505 (4.9)	15 (0.4)	490 (7.6)
Hearing loss	367 (3.6)	28 (0.7)	339 (5.3)
Hypertension	-*	<5	206 (3.2)

* Redacted to adhere to statistical disclosure guidelines.

1.23 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.

	2021			2022		
	Overall (n=10175)	<16 years (n=3878)	≥16 years (n=6297)	Overall (n=10251)	<16 years (n=3806)	≥16 years (n=6445)
ABPA	149 (1.5)	71 (1.8)	78 (1.3)	112 (1.1)	47 (1.2)	65 (1.1)
Cirrhosis - no portal hypertension	57 (0.6)	19 (0.5)	38 (0.6)	41 (0.4)	6 (0.2)	35 (0.6)
Cirrhosis - with portal hypertension	39 (0.4)	11 (0.3)	28 (0.5)	41 (0.4)	9 (0.2)	32 (0.5)
Cancer confirmed by histology	*	<5	15 (0.2)	*	<5	16 (0.3)

1.24 CF diabetes**
N=8078

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=8078)	10-15 years (n=1633)	≥16 years (n=6445)
On CFD treatment; n(%)	2315 (28.7)	129 (7.9)	2186 (33.9)
Of those on treatment			
Insulin[1]; n(%)	1952 (84.3)	124 (96.1)	1828 (83.6)
CFD Screening ; n(%)			
Yes	3477 (43.0)	1051 (64.4)	2426 (37.6)
Screening Type			
Continuous glucose monitoring[2];n(%)	1015 (29.2)	271 (25.8)	744 (30.7)
Oral glucose tolerance test[2]; n(%)	1356 (39.0)	493 (46.9)	863 (35.6)
Not screened (other)	2330 (28.8)	96 (5.9)	2234 (34.7)
Not screened (known CFD)	2153 (26.7)	429 (26.3)	1724 (26.7)
Unknown	106 (1.3)	56 (3.4)	50 (0.8)

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

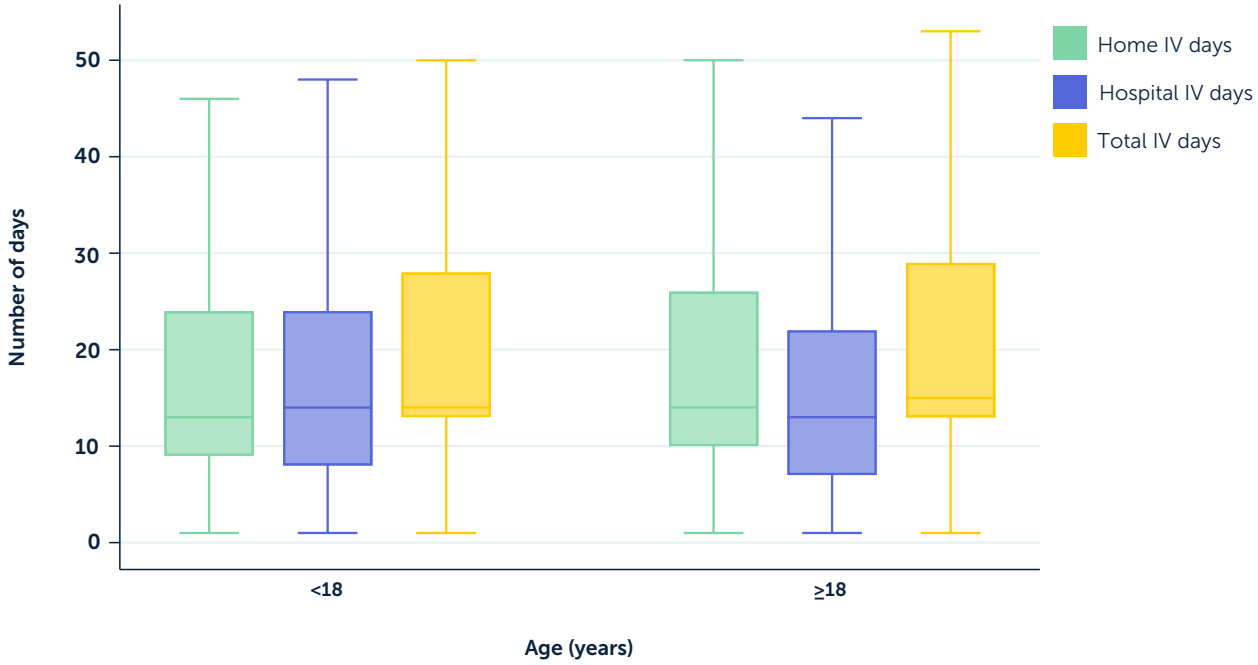
1.25 Intravenous (IV) antibiotics
N=10251

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

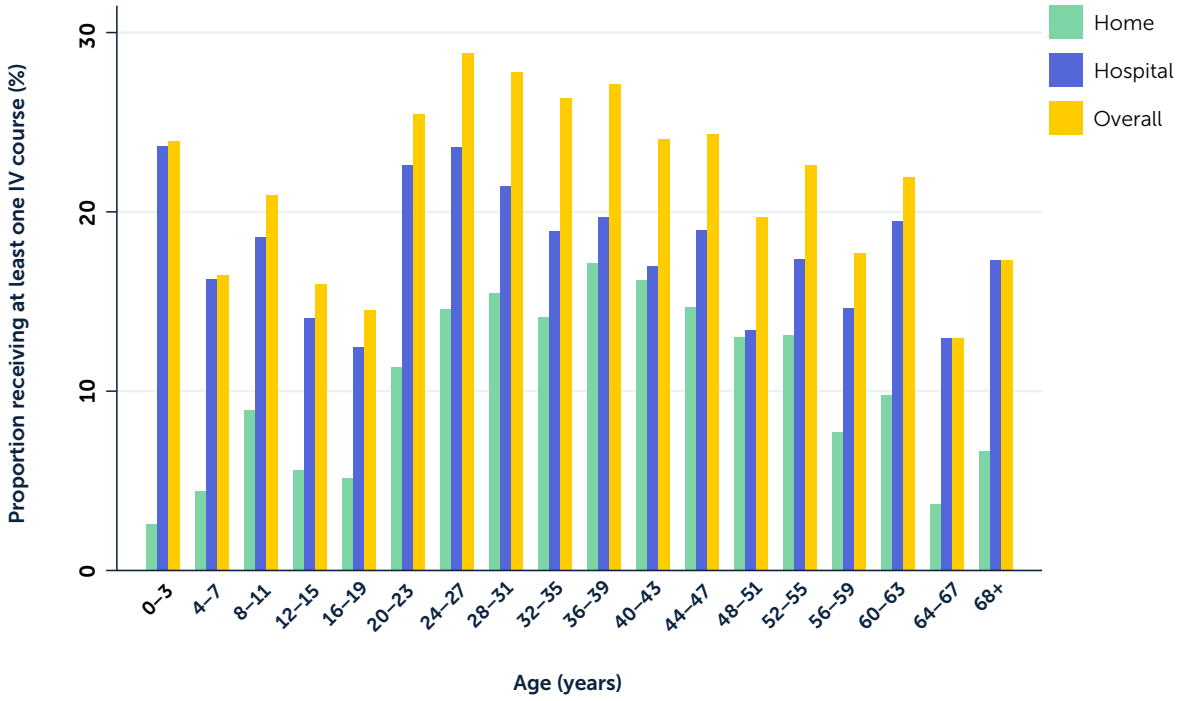
		Home		Hospital		Total	
Age	n	Patients n(%)	Median days (IQR)	Patients n(%)	Median days (IQR)	Patients n(%)	Median days (IQR)
0-3	689	18 (2.6)	7 (6-15)	163 (23.7)	13 (9-16)	165 (23.9)	14 (10-19)
4-7	971	43 (4.4)	12 (7-18)	158 (16.3)	13 (7-17)	160 (16.5)	14 (10-24)
8-11	1093	98 (9.0)	13 (9-21)	203 (18.6)	14 (7-28)	229 (21.0)	15 (14-32)
12-15	1053	59 (5.6)	14 (12-28)	148 (14.1)	14 (8-28)	168 (16.0)	15 (14-39)
16-19	818	42 (5.1)	14 (9-28)	102 (12.5)	14 (9-27)	119 (14.5)	16 (13-31)
20-23	853	97 (11.4)	14 (8-22)	193 (22.6)	13 (7-25)	217 (25.4)	14 (12-28)
24-27	919	134 (14.6)	14 (10-27)	217 (23.6)	13 (7-23)	265 (28.8)	15 (13-29)
28-31	859	133 (15.5)	14 (10-24)	184 (21.4)	13 (8-23)	239 (27.8)	14 (13-31)
32-35	793	112 (14.1)	14 (9-28)	150 (18.9)	11 (6-15)	209 (26.4)	14 (11-28)
36-39	594	102 (17.2)	14 (12-28)	117 (19.7)	13 (6-21)	161 (27.1)	19 (14-31)
40-43	524	85 (16.2)	16 (13-28)	89 (17.0)	14 (9-25)	126 (24.0)	22 (14-42)
44-47	300	44 (14.7)	14 (12-26)	57 (19.0)	13 (6-20)	73 (24.3)	16 (14-29)
48-51	254	33 (13.0)	14 (10-15)	34 (13.4)	12 (8-22)	50 (19.7)	14 (13-28)
52-55	190	25 (13.2)	14 (14-21)	33 (17.4)	9 (6-20)	43 (22.6)	15 (9-37)
56-59	130	10 (7.7)	14 (10-14)	19 (14.6)	14 (10-17)	23 (17.7)	14 (13-26)
60-63	82	8 (9.8)	11 (9-14)	16 (19.5)	13 (6-17)	18 (22.0)	14 (13-27)
64-67	54	<5	11 (2-20)	7 (13.0)	14 (8-43)	7 (13.0)	16 (14-43)
68+	75	5 (6.7)	18 (12-26)	13 (17.3)	14 (7-33)	13 (17.3)	15 (12-33)
<16	3806	218 (5.7)	13 (9-22)	672 (17.7)	14 (8-22)	722 (19.0)	14 (13-28)
≥16	6445	832 (12.9)	14 (10-26)	1231 (19.1)	13 (7-23)	1563 (24.3)	15 (13-29)
<18	4226	235 (5.6)	13 (9-24)	722 (17.1)	14 (8-24)	782 (18.5)	14 (13-28)
≥18	6025	815 (13.5)	14 (10-26)	1181 (19.6)	13 (7-22)	1503 (24.9)	15 (13-29)
Overall	**10251**	**1050 (10.2)**	**14 (10-26)**	**1903 (18.6)**	**13 (7-23)**	**2285 (22.3)**	**14 (13-28)**

* Redacted to adhere to statistical disclosure guidelines.

This box plot graph illustrates the spread of the number of days on IV antibiotics in the UK CF population, stratified by age. A guide on how to correctly interpret this box plot graph can be found on page 53.



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 10.2% and in hospital was 18.6%. The proportion receiving any IVs was 22.3%.



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

	2012			2017			2022		
	Overall	<16 years	≥16 years	Overall	<16 years	≥16 years	Overall	<16 years	≥16 years
Patients with chronic *P. aeruginosa*	3041	377	2664	2749	209	2540	1024	108	916
Tobramycin solution; n(%)	1018 (33.5)	120 (31.8)	898 (33.7)	626 (22.8)	72 (34.4)	554 (21.8)	294 (28.7)	42 (38.9)	252 (27.5)
Other aminoglycoside; n(%)	104 (3.4)	11 (2.9)	93 (3.5)	-*	<5	51 (2.0)	-*	<5	32 (3.5)
Colistin; n(%)	1326 (43.6)	214 (56.8)	1112 (41.7)	680 (24.7)	79 (37.8)	601 (23.7)	317 (31.0)	55 (50.9)	262 (28.6)
Promixin; n(%)	810 (26.6)	133 (35.3)	677 (25.4)	859 (31.2)	98 (46.9)	761 (30.0)	271 (26.5)	36 (33.3)	235 (25.7)
Aztreonam; n(%)	0	0	0	628 (22.8)	10 (4.8)	618 (24.3)	281 (27.4)	8 (7.4)	273 (29.8)
Colistimethate (DPI); n(%)	0	0	0	531 (19.3)	15 (7.2)	516 (20.3)	197 (19.2)	7 (6.5)	190 (20.7)
Tobramycin Inhalation Powder; n(%)	0	0	0	782 (28.4)	26 (12.4)	756 (29.8)	-*	<5	174 (19.0)
Levofloxacin; n(%)	0	0	0	0	0	0	27 (2.6)	0	27 (2.9)
At least one of the above; n(%)	2444 (80.4)	340 (90.2)	2104 (79.0)	2469 (89.8)	191 (91.4)	2278 (89.7)	888 (86.7)	96 (88.9)	792 (86.5)

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

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(%)
2012	**Overall**	3475	1974 (56.8)	1501 (43.2)
	0-3 years	-*	<5	35 (92.1)
	4-15 years	608	156 (25.7)	452 (74.3)
	≥ 16 years	2829	1815 (64.2)	1014 (35.8)
2017	**Overall**	4103	1922 (46.8)	2181 (53.2)
	0-3 years	-*	<5	34 (91.9)
	4-15 years	676	89 (13.2)	587 (86.8)
	≥ 16 years	3390	1830 (54.0)	1560 (46.0)
2022	**Overall**	4034	669 (16.6)	3365 (83.4)
	0-3 years	-*	<5	47 (94.0)
	4-15 years	523	41 (7.8)	482 (92.2)
	≥ 16 years	3461	625 (18.1)	2836 (81.9)

* Redacted to adhere to statistical disclosure guidelines.

1.28 Prophylactic flucloxacillin use

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

Age	Total patients	Patients on prophylactic flucloxacillin; n(%)
0-3	689	329 (47.8)
4-7	971	308 (31.7)
8-11	1093	280 (25.6)
12-15	1053	237 (22.5)
16-19	818	171 (20.9)
20-23	853	227 (26.6)
24-27	919	120 (13.1)
28-31	859	60 (7.0)
32-35	793	47 (5.9)
36-39	594	21 (3.5)
40-43	524	26 (5.0)
44-47	300	12 (4.0)
48-51	254	14 (5.5)
52-55	190	12 (6.3)
56-59	130	5 (3.8)
60-63	82	<5
64-67	54	<5
68+	75	<5
<16 years	3806	1154 (30.3)
≥16 years	6445	723 (11.2)
<18 years	4226	1240 (29.3)
≥18 years	6025	637 (10.6)
Overall	**10251**	**1877 (18.3)**

* Redacted to adhere to statistical disclosure guidelines.

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(%)
0-3	689	128 (18.6)	51 (7.4)	<5
4-7	971	308 (31.7)	130 (13.4)	28 (2.9)
8-11	1093	556 (50.9)	207 (18.9)	132 (12.1)
12-15	1053	600 (57.0)	177 (16.8)	203 (19.3)
16-19	818	536 (65.5)	136 (16.6)	240 (29.3)
20-23	853	631 (74.0)	203 (23.8)	301 (35.3)
24-27	919	649 (70.6)	168 (18.3)	264 (28.7)
28-31	859	632 (73.6)	164 (19.1)	312 (36.3)
32-35	793	580 (73.1)	159 (20.1)	293 (36.9)
36-39	594	423 (71.2)	112 (18.9)	219 (36.9)
40-43	524	372 (71.0)	103 (19.7)	223 (42.6)
44-47	300	214 (71.3)	63 (21.0)	116 (38.7)
48-51	254	171 (67.3)	47 (18.5)	114 (44.9)
52-55	190	117 (61.6)	46 (24.2)	70 (36.8)
56-59	130	97 (74.6)	22 (16.9)	49 (37.7)
60-63	82	59 (72.0)	22 (26.8)	26 (31.7)
64-67	54	38 (70.4)	11 (20.4)	22 (40.7)
68+	75	54 (72.0)	19 (25.3)	29 (38.7)
<16 years	3806	1592 (41.8)	565 (14.8)	-*
≥16 years	6445	4573 (71.0)	1275 (19.8)	2278 (35.3)
<18 years	4226	1853 (43.8)	636 (15.0)	479 (11.3)
≥18 years	6025	4312 (71.6)	1204 (20.0)	2166 (36.0)
Overall	**10251**	**6165 (60.1)**	**1840 (17.9)**	**2645 (25.8)**

* Redacted to adhere to statistical disclosure guidelines.

Muco-active therapies

1.30 Mannitol

	2017		2022	
Age	Total patients	Patients on Mannitol; n(%)	Total patients	Patients on Mannitol; n(%)
0-3	854	0	689	0
4-7	1124	0	971	0
8-11	1031	0	1093	0
12-15	889	<5	1053	<5
16-19	918	21 (2.3)	818	6 (0.7)
20-23	1010	60 (5.9)	853	27 (3.2)
24-27	942	63 (6.7)	919	40 (4.4)
28-31	810	58 (7.2)	859	41 (4.8)
32-35	698	47 (6.7)	793	53 (6.7)
36-39	483	33 (6.8)	594	30 (5.1)
40-43	311	22 (7.1)	524	29 (5.5)
44-47	280	14 (5.0)	300	23 (7.7)
48-51	201	5 (2.5)	254	11 (4.3)
52-55	138	<5	190	6 (3.2)
56-59	76	<5	130	5 (3.8)
60-63	48	0	82	<5
64-67	32	<5	54	0
68+	42	0	75	<5
<16 years	3898	<5	3806	<5
≥16 years	5989	330 (5.5)	6445	273 (4.2)
<18 years	4329	6 (0.1)	4226	6 (0.1)
≥18 years	5558	325 (5.8)	6025	270 (4.5)
Overall	**9887**	**331 (3.3)**	**10251**	**276 (2.7)**

1.31 DNase

	2012		2017		2022	
Age	Total patients	Patients on DNase; n(%)	Total patients	Patients on DNase; n(%)	Total patients	Patients on DNase; n(%)
0-3	929	97 (10.4)	854	128 (15.0)	689	137 (19.9)
4-7	962	306 (31.8)	1124	580 (51.6)	971	515 (53.0)
8-11	862	445 (51.6)	1031	790 (76.6)	1093	862 (78.9)
12-15	979	612 (62.5)	889	725 (81.6)	1053	864 (82.1)
16-19	993	587 (59.1)	918	742 (80.8)	818	690 (84.4)
20-23	1011	626 (61.9)	1010	728 (72.1)	853	755 (88.5)
24-27	816	475 (58.2)	942	641 (68.0)	919	747 (81.3)
28-31	679	395 (58.2)	810	536 (66.2)	859	603 (70.2)
32-35	459	237 (51.6)	698	431 (61.7)	793	540 (68.1)
36-39	303	141 (46.5)	483	292 (60.5)	594	363 (61.1)
40-43	291	139 (47.8)	311	179 (57.6)	524	320 (61.1)
44-47	192	84 (43.8)	280	153 (54.6)	300	179 (59.7)
48-51	132	66 (50.0)	201	105 (52.2)	254	138 (54.3)
52-55	79	33 (41.8)	138	68 (49.3)	190	101 (53.2)
56-59	43	14 (32.6)	76	41 (53.9)	130	74 (56.9)
60-63	27	11 (40.7)	48	21 (43.8)	82	47 (57.3)
64-67	18	7 (38.9)	32	19 (59.4)	54	31 (57.4)
68+	19	11 (57.9)	42	15 (35.7)	75	34 (45.3)
<16 years	3732	1460 (39.1)	3898	2223 (57.0)	3806	2378 (62.5)
≥16 years	5062	2826 (55.8)	5989	3971 (66.3)	6445	4622 (71.7)
<18 years	4251	1783 (41.9)	4329	2565 (59.3)	4226	2724 (64.5)
≥18 years	4543	2503 (55.1)	5558	3629 (65.3)	6025	4276 (71.0)
Overall	**8794**	**4286 (48.7)**	**9887**	**6194 (62.6)**	**10251**	**7000 (68.3)**

1.32 Hypertonic saline

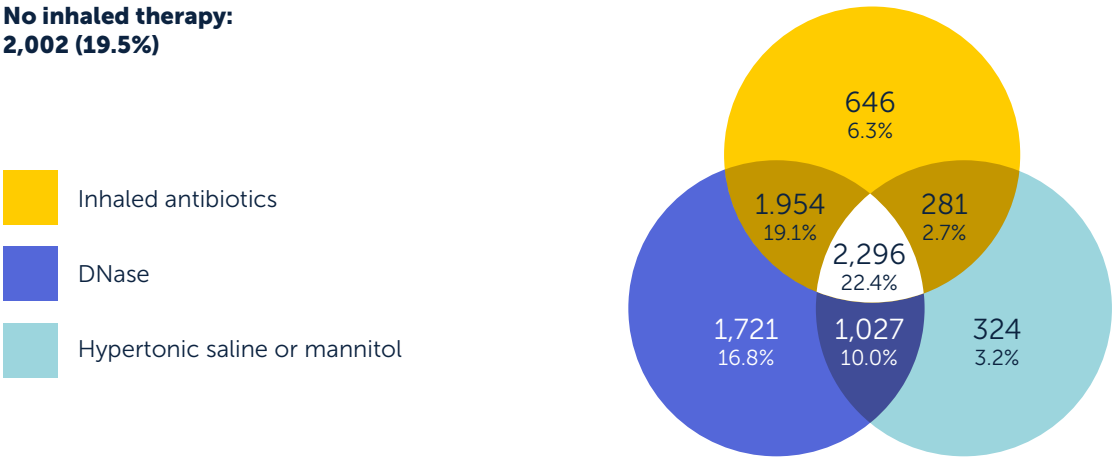
This treatment helps to thin mucus so that it is easier to cough out of the body.

	2012		2017		2022	
Age	Total patients	Patients on hypertonic saline; n(%)	Total patients	Patients on hypertonic saline; n(%)	Total patients	Patients on hypertonic saline; n(%)
0-3	929	51 (5.5)	854	70 (8.2)	689	146 (21.2)
4-7	962	127 (13.2)	1124	271 (24.1)	971	287 (29.6)
8-11	862	212 (24.6)	1031	352 (34.1)	1093	454 (41.5)
12-15	979	260 (26.6)	889	397 (44.7)	1053	468 (44.4)
16-19	993	231 (23.3)	918	415 (45.2)	818	393 (48.0)
20-23	1011	234 (23.1)	1010	331 (32.8)	853	492 (57.7)
24-27	816	205 (25.1)	942	265 (28.1)	919	400 (43.5)
28-31	679	167 (24.6)	810	256 (31.6)	859	261 (30.4)
32-35	459	101 (22.0)	698	251 (36.0)	793	209 (26.4)
36-39	303	59 (19.5)	483	160 (33.1)	594	181 (30.5)
40-43	291	56 (19.2)	311	103 (33.1)	524	153 (29.2)
44-47	192	43 (22.4)	280	78 (27.9)	300	89 (29.7)
48-51	132	30 (22.7)	201	50 (24.9)	254	76 (29.9)
52-55	79	21 (26.6)	138	38 (27.5)	190	50 (26.3)
56-59	43	6 (14.0)	76	23 (30.3)	130	29 (22.3)
60-63	27	7 (25.9)	48	7 (14.6)	82	31 (37.8)
64-67	18	<5	32	7 (21.9)	54	10 (18.5)
68+	19	<5	42	15 (35.7)	75	25 (33.3)
<16 years	3732	650 (17.4)	3898	1090 (28.0)	3806	1355 (35.6)
≥16 years	5062	1166 (23.0)	5989	1999 (33.4)	6445	2399 (37.2)
<18 years	4251	779 (18.3)	4329	1304 (30.1)	4226	1536 (36.3)
≥18 years	4543	1037 (22.8)	5558	1785 (32.1)	6025	2218 (36.8)
Overall	**8794**	**1816 (20.7)**	**9887**	**3089 (31.2)**	**10251**	**3754 (36.6)**

1.33 Burden of treatment

The Venn diagram shows how many people with CF are on one or more inhaled therapy and the combinations they take. 2,002 (19.5%) people are on no inhaled therapies.

No inhaled therapy: 2,002 (19.5%)



Other therapies

1.34 CFTR modulators

In 2022, the CFTR modulators were available to the following people across the UK with cystic fibrosis under a managed access agreement. The access arrangements prior to 2022 are described in previous annual reports.

Ivacaftor

In 2022, ivacaftor has approval for use for people aged four months and older with at least one copy of a CFTR 'gating' mutation, and for people aged four months and over with the R117H.

Lumacaftor/ivacaftor

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

Tezacaftor/ivacaftor

Tezacaftor/ivacaftor is licenced 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.

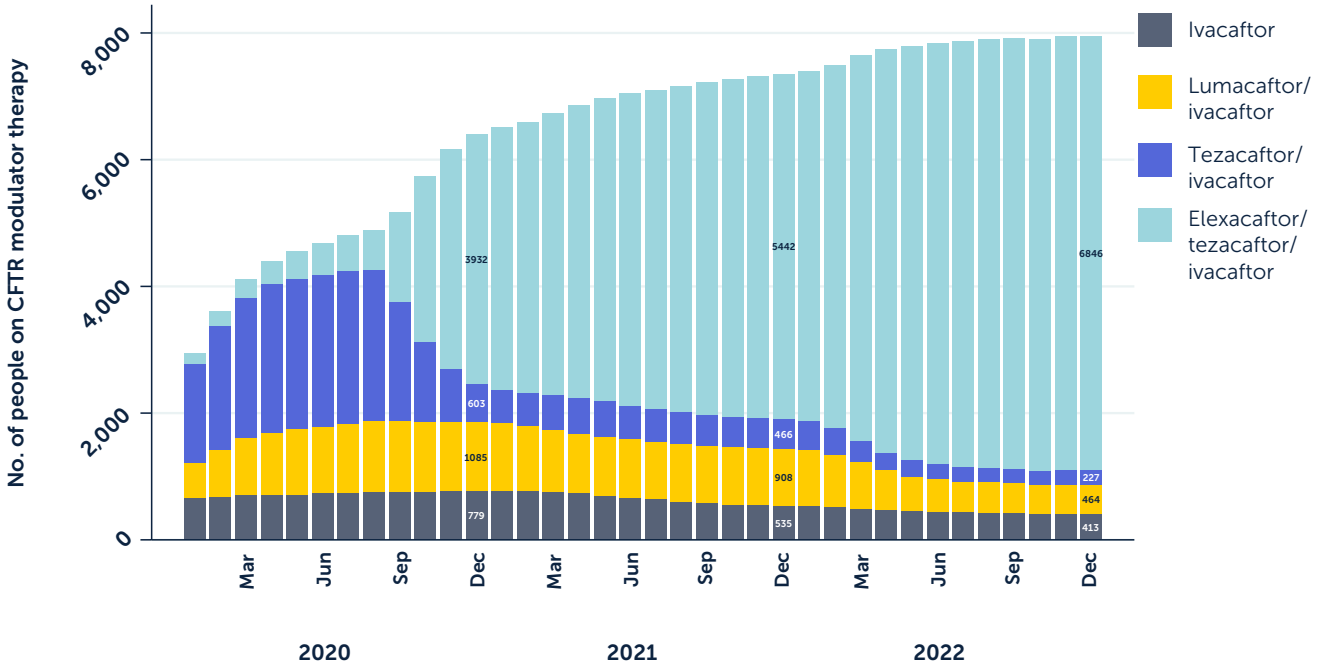
Elexacaftor/tezacaftor/ivacaftor

During 2022, elexacaftor/tezacaftor/ ivacaftor was available in the UK for patients with cystic fibrosis aged 6 and over who have two copies of the F508del mutation, or a single copy of F508del and one minimal function mutation.

Guidance has been issued throughout the year from NHS commissioners across the devolved nations to support the prescribing of CFTR modulators "off-label"; this varies slightly across the devolved nations but covers the 177 mutations that are on an approved "FDA list".

CFTR modulator use 2020–2022

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. By December 2022, 7,950 people were taking a CFTR modulator.



1.35 Oxygen and non-invasive ventilation

	Overall (n=10251)	<16 years (n=3806)	≥16 years (n=6445)	<18 years (n=4226)	≥18 years (n=6025)
Non Invasive Ventilation (NIV); n(%)	135 (1.3)	14 (0.4)	121 (1.9)	19 (0.4)	116 (1.9)
Any oxygen use; n(%)	333 (3.2)	48 (1.3)	285 (4.4)	49 (1.2)	284 (4.7)
Among those who had oxygen use:					
Continuously	46 (13.8)	<5	45 (15.8)	<5	45 (15.8)
Nocturnal or with exertion	149 (44.7)	11 (22.9)	138 (48.4)	12 (24.5)	137 (48.2)
As required (PRN)	32 (9.6)	<5	31 (10.9)	<5	31 (10.9)
With exacerbation	106 (31.8)	35 (72.9)	71 (24.9)	35 (71.4)	71 (25.0)

1.36 Physiotherapy

Physiotherapy helps people with CF clear sticky mucus from their lungs.

	Overall (n=10251)	<16 years (n=3806)	≥16 years (n=6445)	<18 years (n=4226)	≥18 years (n=6025)
Active cycle of breathing techniques; n(%)	1182 (11.5)	265 (7.0)	917 (14.2)	302 (7.1)	880 (14.6)
Autogenic drainage (including assisted autogenic drainage); n(%)	1752 (17.1)	156 (4.1)	1596 (24.8)	191 (4.5)	1561 (25.9)
Postural drainage; n(%)	571 (5.6)	448 (11.8)	123 (1.9)	466 (11.0)	105 (1.7)
Any form of PEP; n(%)	5997 (58.5)	2863 (75.2)	3134 (48.6)	3181 (75.3)	2816 (46.7)
VEST; n(%)	131 (1.3)	63 (1.7)	68 (1.1)	74 (1.8)	57 (0.9)
Exercise; n(%)	5963 (58.2)	2401 (63.1)	3562 (55.3)	2683 (63.5)	3280 (54.4)
Other; n(%)	1374 (13.4)	812 (21.3)	562 (8.7)	859 (20.3)	515 (8.5)

Note that these techniques are not mutually exclusive and represent primary and secondary forms of physiotherapy.

1.37 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.

	Overall (n=10251)	<16 years (n=3806)	≥16 years (n=6445)	<18 years (n=4226)	≥18 years (n=6025)
Any supplemental feeding; n(%)	3240 (31.6)	957 (25.1)	2283 (35.4)	1093 (25.9)	2147 (35.6)
Nasogastric tube; n(%)	54 (0.5)	12 (0.3)	42 (0.7)	15 (0.4)	39 (0.6)
Gastrostomy tube/button; n(%)	396 (3.9)	148 (3.9)	248 (3.8)	168 (4.0)	228 (3.8)
Jejunal; n(%)	7 (0.1)	0 (0.0)	7 (0.1)	0 (0.0)	7 (0.1)
Total Parenteral Nutrition (TPN); n(%)	5 (0.0)	<5	<5	<5	<5

* Redacted to adhere to statistical disclosure guidelines.

1.38 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.

	2017	2018	2019	2020	2021	2022
Number evaluated	235	247	241	175	78	41
Number accepted	121	104	96	66	23	22
Number receiving aged <16 years	5	<5	<5	0	0	0
Bilateral lung	<5	0	<5	0	0	0
Liver	0	<5	<5	0	0	0
Other	<5	0	0	0	0	0
Number receiving aged 16+ years	-*	63	54	15	5	6
Bilateral lung	51	58	49	12	<5	<5
Liver	0	<5	<5	<5	0	0
Other	<5	<5	<5	<5	<5	<5

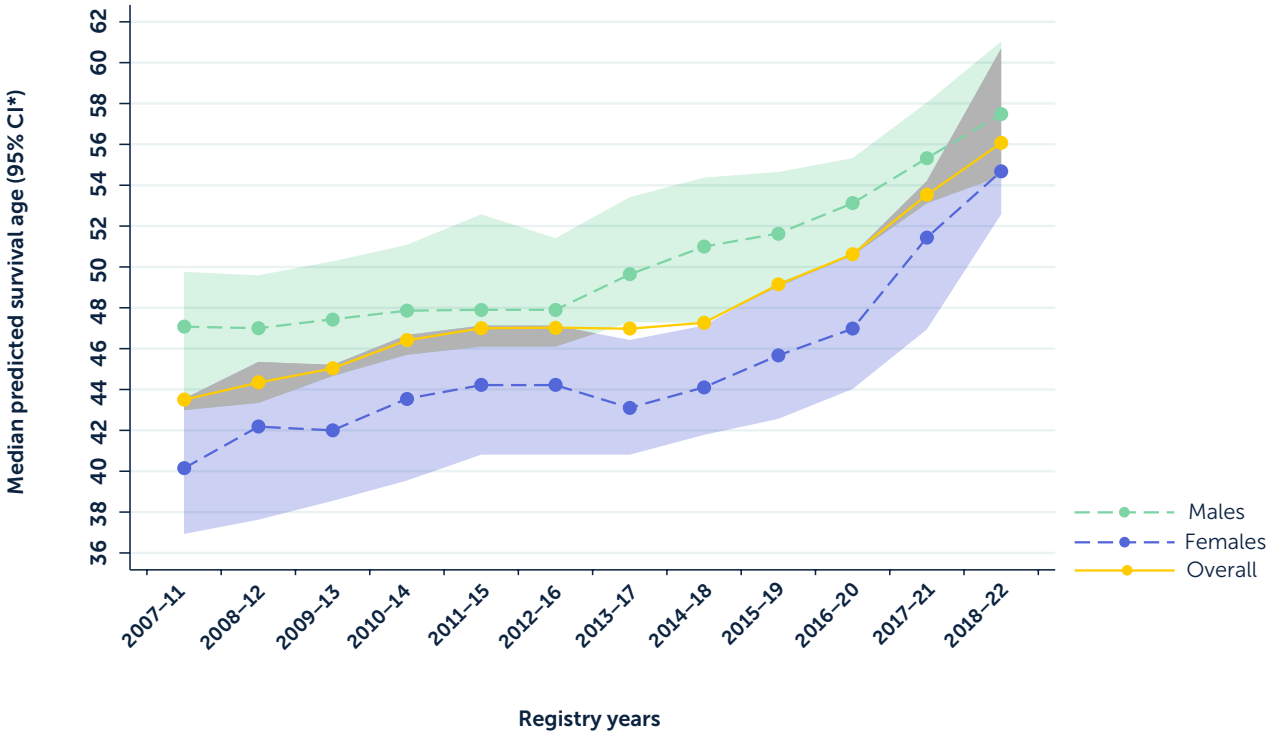
* Redacted to adhere to statistical disclosure guidelines.

Survival

1.39 Median predicted survival age

The calculation of median predicted survival age is based on people with CF who are recorded in the Registry as alive in the given year. A mathematical formula[1] predicts how long we expect half of people with CF born today will live. Half of people born today are predicted to live to at least **56.1** years. Half are therefore predicted to die before they reach that age.

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



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

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

* Confidence interval.

1.40 Age distribution of deaths in 2022

The table below shows the ages of the 64 people with CF who died in 2022.
In 2022 the median age of the 64 people who died was 33.

Age at death	Number of patients
0-19	10
20-25	11
26-30	7
31-35	8
36-40	9
41-45	8
46-50	<5
51-55	<5
56+	**5**
Total	**64**

1.41 Causes of death

This table shows all the recorded causes of death between 2020–2022.

Cause of death	Number of people (n=227)
Respiratory/cardiorespiratory	121 (53.3)
Other	26 (11.5)
Transplant-related	26 (11.5)
Not known	20 (8.8)
Cancer	18 (7.9)
Covid-19	9 (4.0)
Liver disease/liver failure	6 (2.6)
Trauma or suicide	<5
Total	-*

* 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 of the gene for CFTR; one on each allele. One is inherited from their mother, and one from their father. If both mutations (or genotypes) are the same, the person is said to be homozygous. Someone who has two different variants is heterozygous.

Data completeness	n(%)
Patients genotyped with at least one mutation recorded	11092 (99.5)
Patients genotyped with both mutations recorded	10877 (97.6)
F508del mutations	
Homozygous F508del	5324 (47.8)
Heterozygous F508del	4617 (41.4)

1.42 Mutation combinations in the UK population

This tabulation shows the proportion(%) of patients with the most common mutation combinations in their genotype. For example, 4.1% of the UK population have one copy of F508del and one copy of G551D.

Mutation	F508del	R117H	G551D	G542X	621+1G->T	Other	Unknown	Total
F508del	47.8							47.8
R117H	5.1	0.1						5.2
G551D	4.1	0.2	0.2					4.5
G542X	2.5	0.1	0.1	0.1				2.8
621+1G->T	1.7	0.1	0.1	0.1	0.1			2.1
Other	26.8	0.6	1.0	0.8	0.5	5.7		35.4
Unknown	1.3	0.1	0.0	0.1	0.0	0.4	0.5	2.4
Total	**89.2**	**1.3**	**1.4**	**1.0**	**0.6**	**6.1**	**0.5**	**100.0**

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

1.43 Mutations in the UK population

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

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

Nucleotide	Protein	Legacy name	n	%
c.1521_1523delCTT	p.Phe508del	F508del	9941	89.2
c.350G->A	p.Arg117His	R117H	704	6.3
c.1652G->A	p.Gly551Asp	G551D	636	5.7
c.1624G->T	p.Gly542X	G542X	407	3.7
c.489+1G->T		621+1G->T	291	2.6
c.3909C->G	p.Asn1303Lys	N1303K	177	1.6
c.1585-1G->A		1717-1G->A	176	1.6
c.1766+1G->A		1898+1G->A	159	1.4
c.3454G->C	p.Asp1152His	D1152H	151	1.4
c.200C->T	p.Pro67Leu	P67L	148	1.3
c.3140-26A->G		3272-26A->G	127	1.1
c.3528delC	p.Lys1177SerfsX15	3659delC	122	1.1
c.1679G->C	p.Arg560Thr	R560T	104	0.9
c.1519_1521delATC	p.Ile507del	I507del	94	0.8
c.1477C->T	p.Gln493X	Q493X	94	0.8
c.3717+12191C->T		3849+10kbC->T	92	0.8
c.1657C->T	p.Arg553X	R553X	87	0.8
c.254G->A	p.Gly85Glu	G85E	85	0.8
c.2657+5G->A		2789+5G->A	82	0.7
c.178G->T	p.Glu60X	E60X	80	0.7

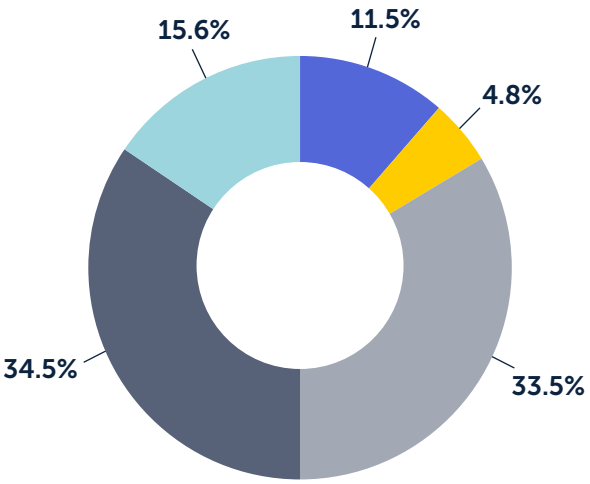
1.44 Mutation prevalence by devolved nation

This table shows the distribution of individual mutations 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 mutations appear twice in the table.

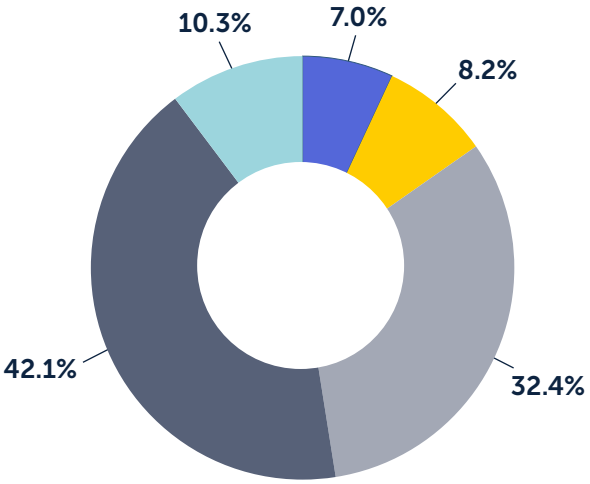
Legacy name	England n=9259		Scotland n=933		Wales n=465		Northern Ireland n=491	
	n	%	n	%	n	%	n	%
F508del	8264	89.3%	843	90.4%	418	89.9%	416	84.7%
R117H	535	5.8%	79	8.5%	17	3.7%	73	14.9%
G551D	481	5.2%	91	9.8%	16	3.4%	48	9.8%
G542X	294	3.2%	60	6.4%	23	4.9%	30	6.1%
621+1G->T	211	2.3%	10	1.1%	52	11.2%	18	3.7%
N1303K	149	1.6%	11	1.2%	7	1.5%	10	2.0%
1717-1G->A	156	1.7%	16	1.7%	<5	-	<5	-
1898+1G->A	123	1.3%	5	0.5%	30	6.5%	<5	-
D1152H	120	1.3%	18	1.9%	<5	-	10	2.0%
P67L	76	0.8%	50	5.4%	<5	-	20	4.1%

1.45 Genotype prevalence by devolved nation

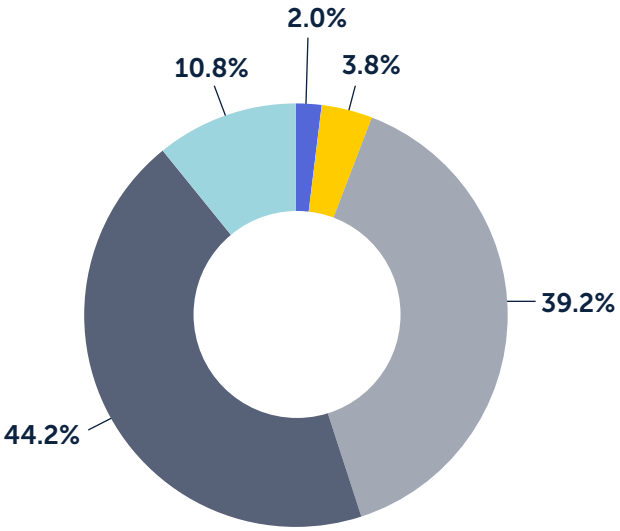
These charts show the distribution of mutation combinations across the devolved nations. The number of patients for each devolved nation is based on the location of the CF centre at which the patient receives care and does not account for patients who travel between devolved nations for care.



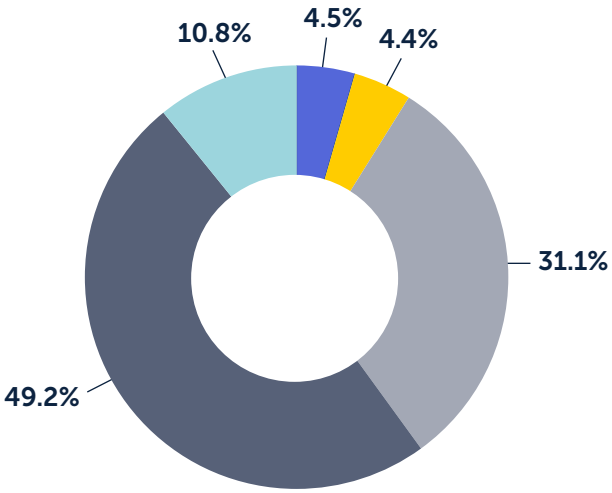
Northern Ireland
N=491



Scotland
N=933



Wales
N=465



England
N=9259

Homozygous F508del

Heterozygous F508del

F508del + gating (incl. G551D)

F508del + R117H

Other

Section 2 and 3: Centre-level analysis

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

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

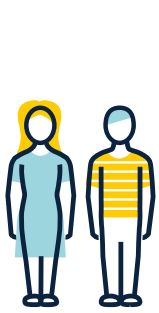
Section 2 shows analysis of data for individual CF centres. This allows people with CF, their families, and healthcare providers, to review a centre's use of some medications and outcome data alongside national averages. This transparency is intended to help improve standards of care overall.

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

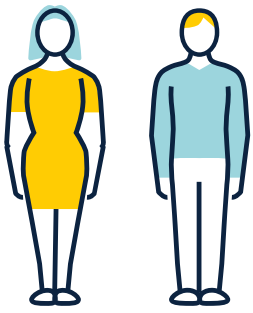
If a person with CF or a member of their family has questions about the results for their CF centre or clinic, they should discuss this with their CF team.

Full tables of the data are shown in appendix 2 on page 68.

Key



Paediatric centre

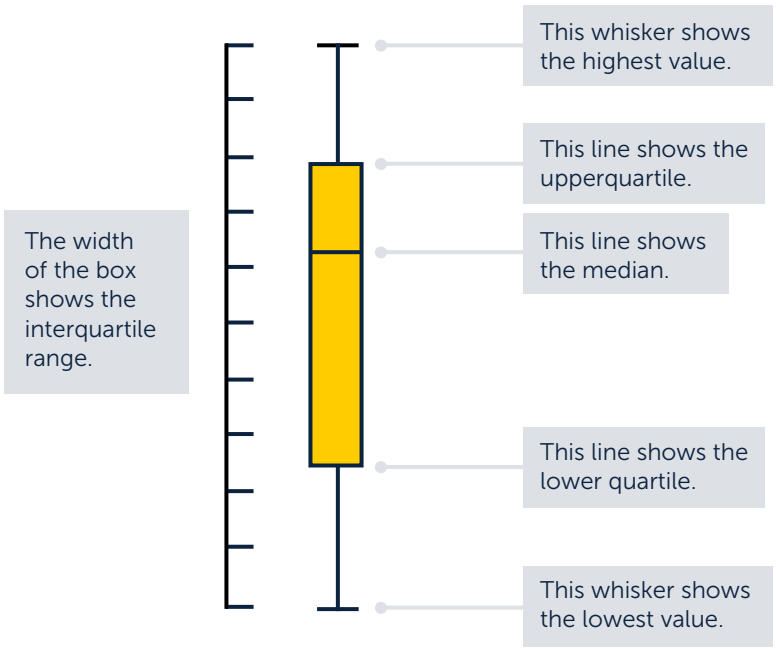


Adult centre

A guide to the charts

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

Box plots



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

Funnel plots

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

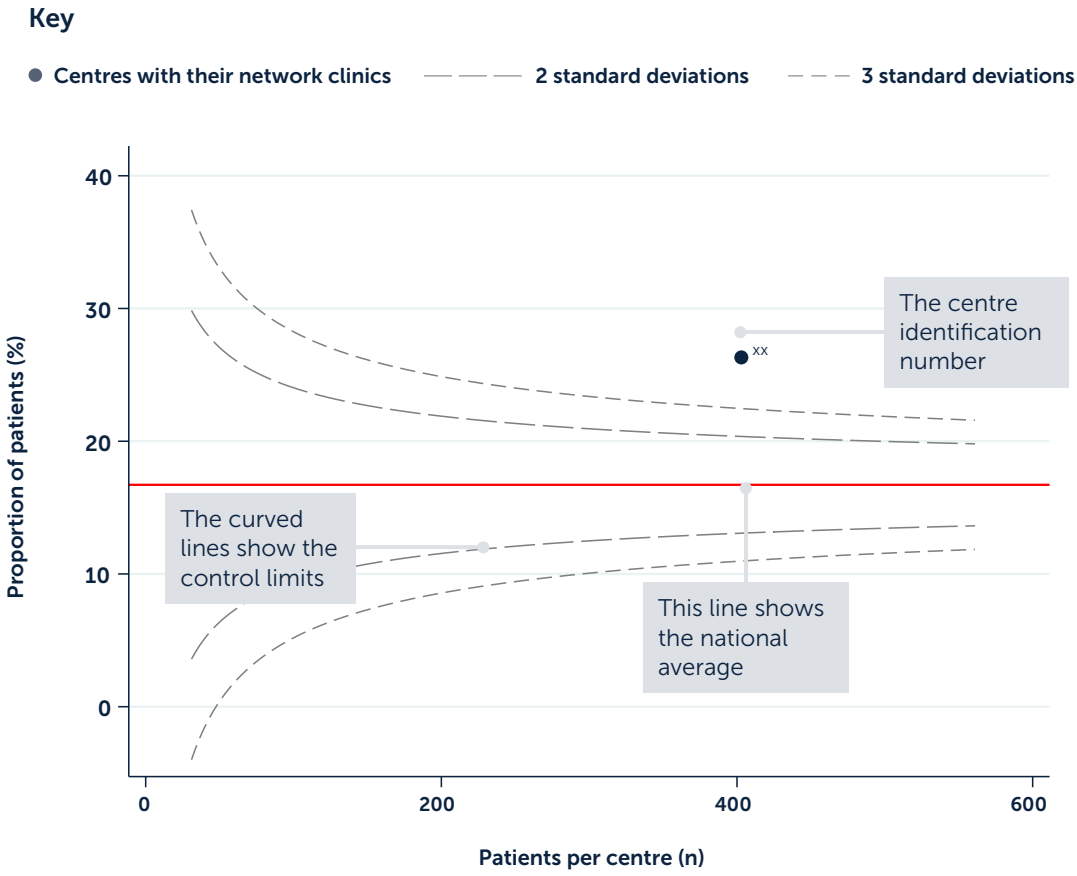
There will always be some natural variation between centres because of differences between the populations receiving care. Using only the national average as a standard can make it difficult to tell whether a survival rate that sits above the national average is higher than we would expect it to be,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

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

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

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

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

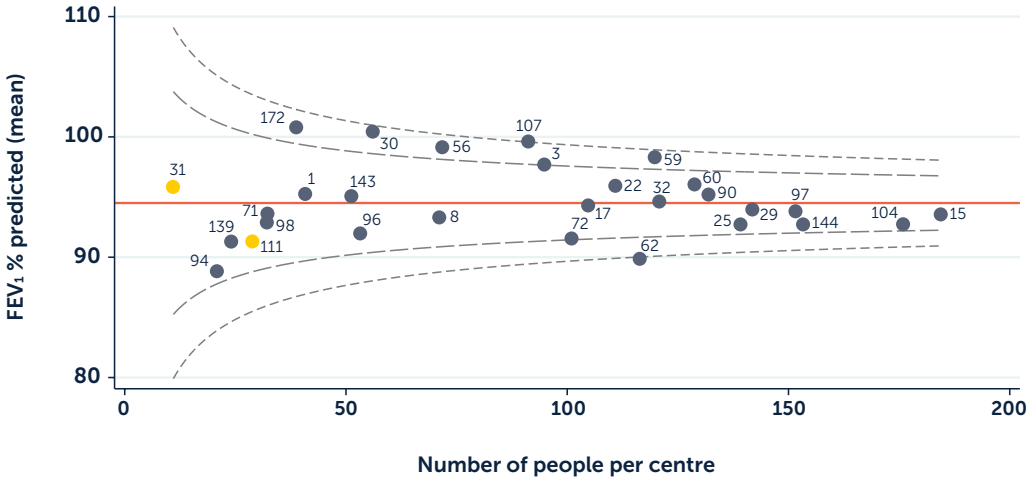


Section 2 Paediatric centre analysis

N=4099

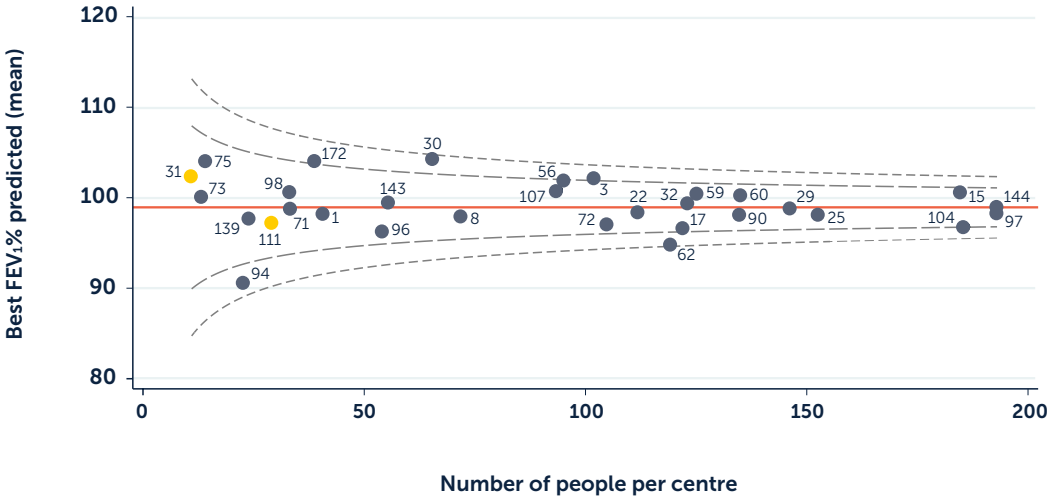
In the UK, paediatric CF care is led by 30 specialist CF centres and two standalone clinics (●). Some paediatric centres oversee care delivered by 68 smaller, networked clinics. Data from smaller networked clinics is included in the paediatric centre's data.

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



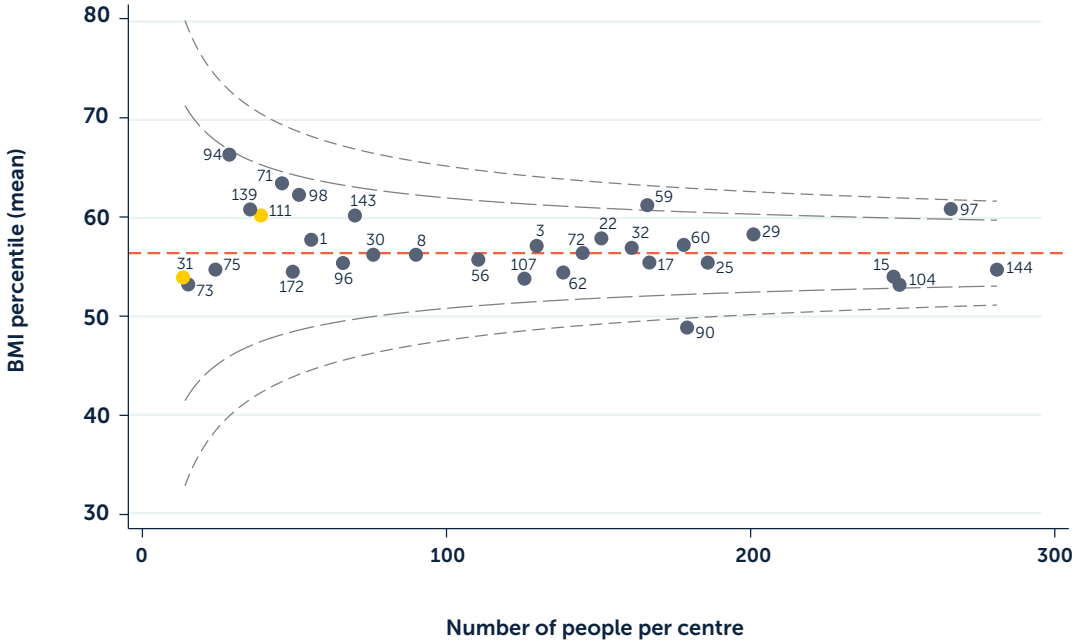
The mean FEV_1% predicted for patients attending paediatric centres/clinics is 94.5% predicted.

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



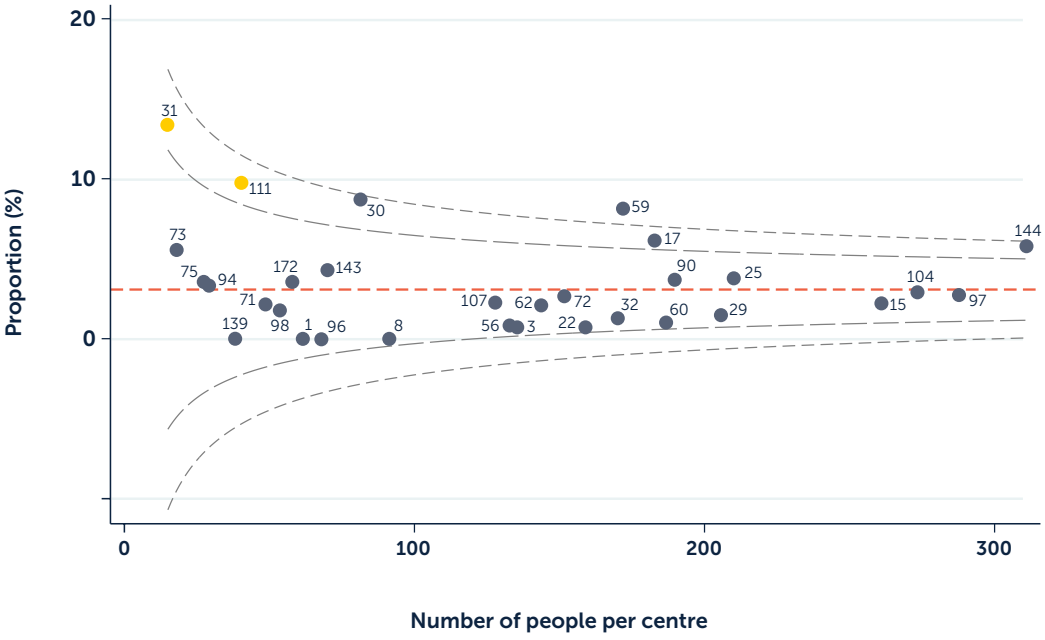
The mean Best FEV_1% predicted for patients attending paediatric centres/clinics is 98.9% predicted. Where Best FEV_1% predicted was missing, the FEV_1% predicted at annual review was used.

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



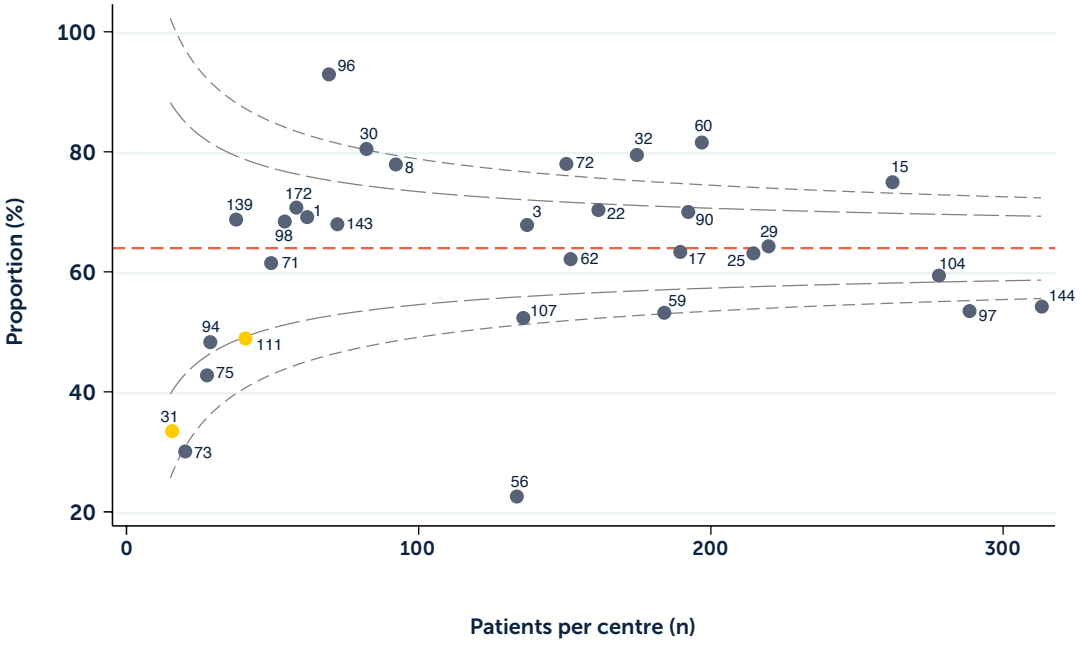
The mean BMI percentile for patients attending paediatric centres/clinics is 56.4%.

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



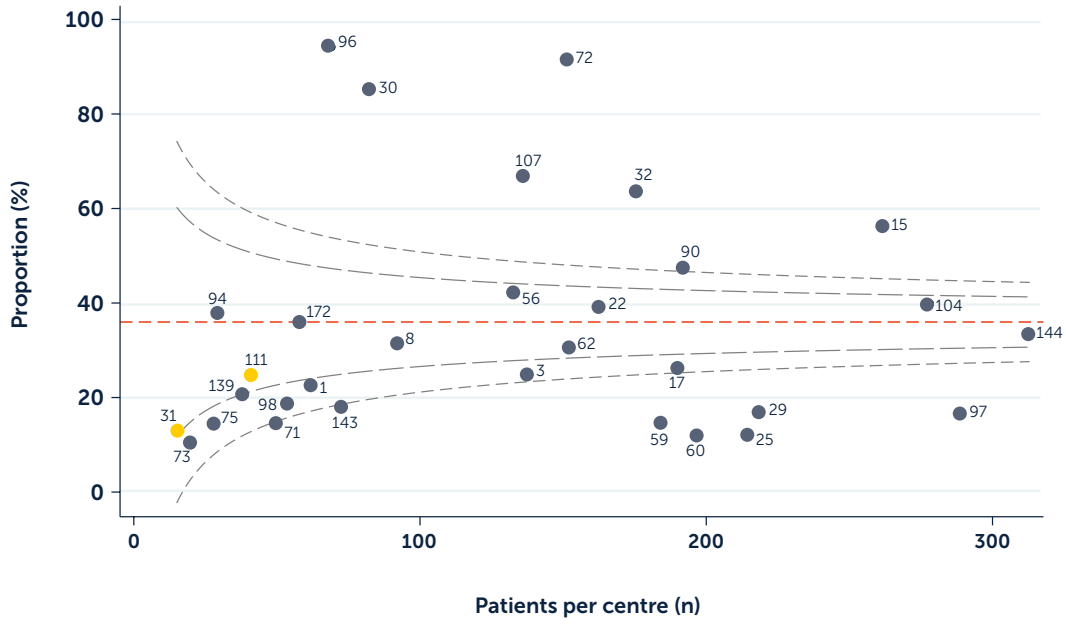
The proportion of patients with chronic *Pseudomonas aeruginosa* in paediatric centres/clinics is 3.1%.

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



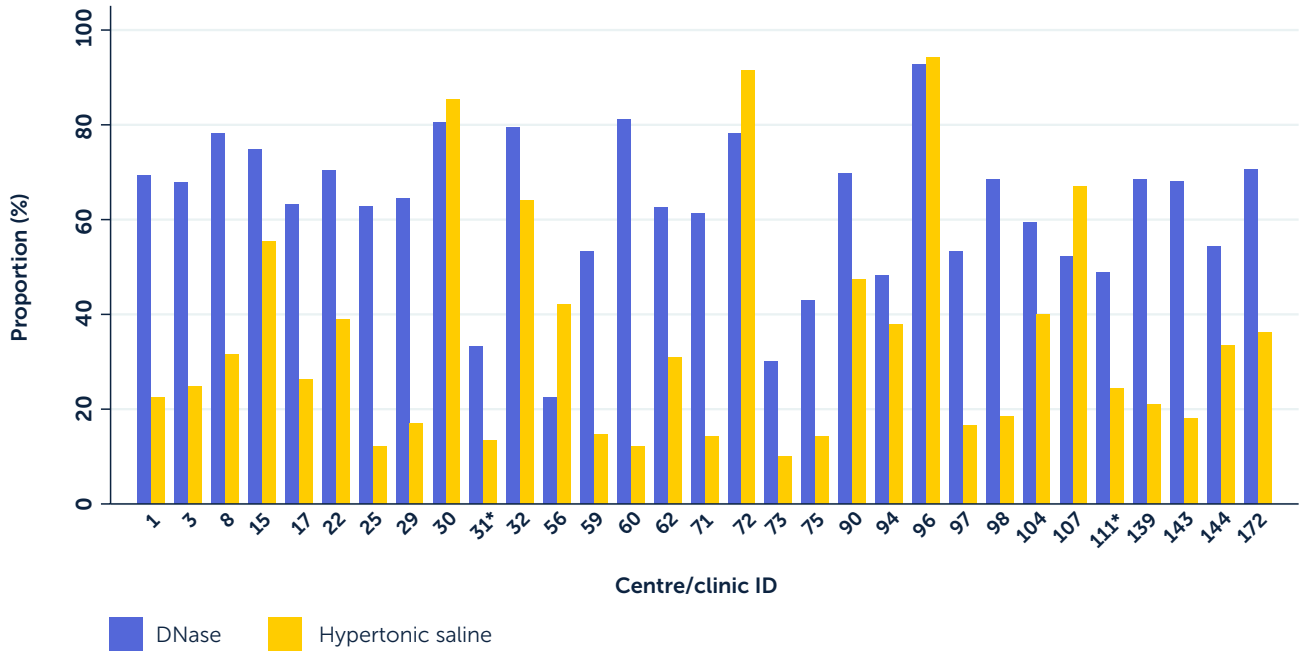
The proportion of patients receiving DNase treatment in paediatric centres/clinics is 63.9%.

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



The proportion of patients receiving hypertonic saline or mannitol treatment in paediatric centres/clinics is 36.0%.

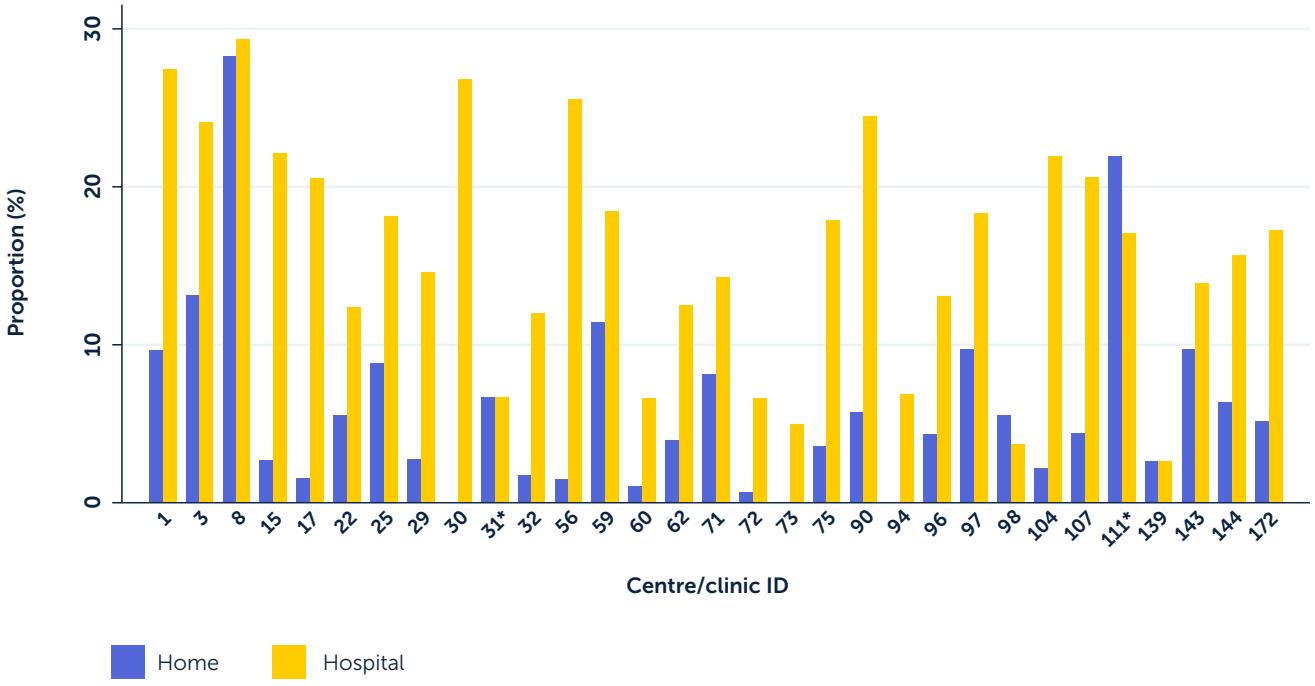
2.7 Proportion of patients receiving DNase/hypertonic saline/ mannitol treatment by paediatric centre/clinic



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

2.8 IV use by paediatric centre/clinic

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



The proportion of patients receiving IVs at home was 5.9% and in hospital was 15.5%. The proportion receiving any IVs was 17.2%.

* Standalone clinics.

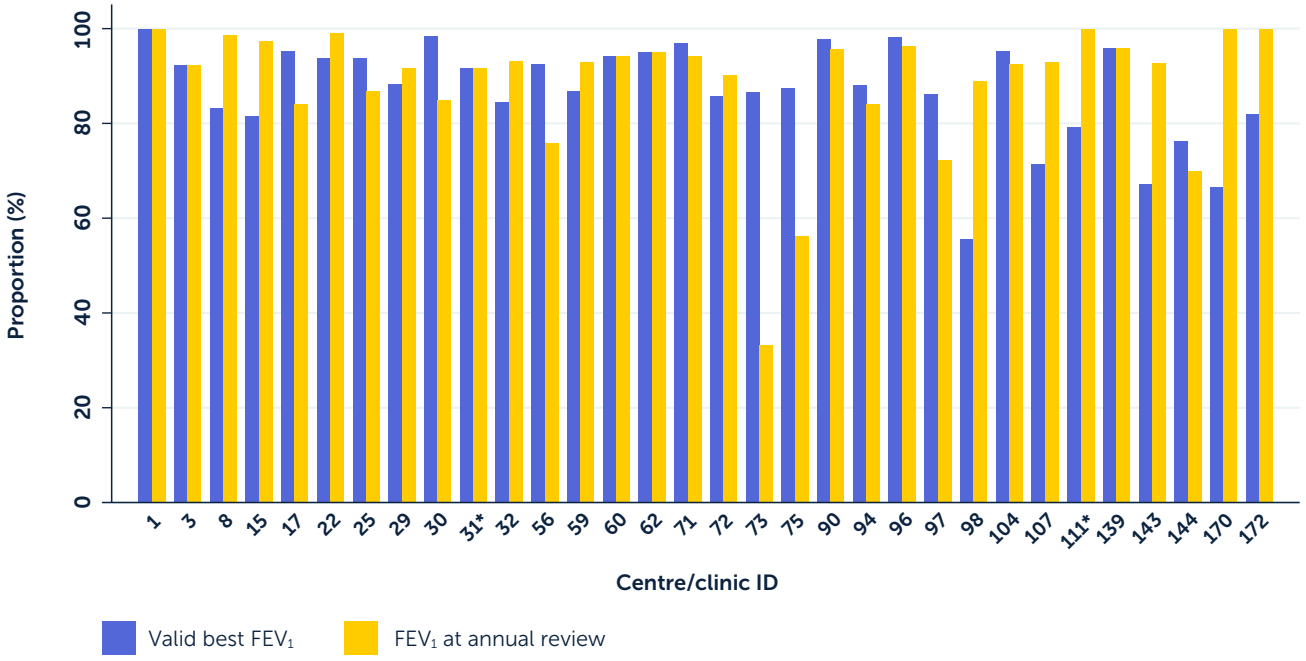
2.9 Inhaled antibiotic use for patients with chronic *Pseudomonas aeruginosa*, by paediatric centre/clinic

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

Centre/clinic ID	Proportion(%)
17	100.0
59	100.0
144	94.4

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

2.10 Data completeness by paediatric centre/clinic**



* Standalone clinics.

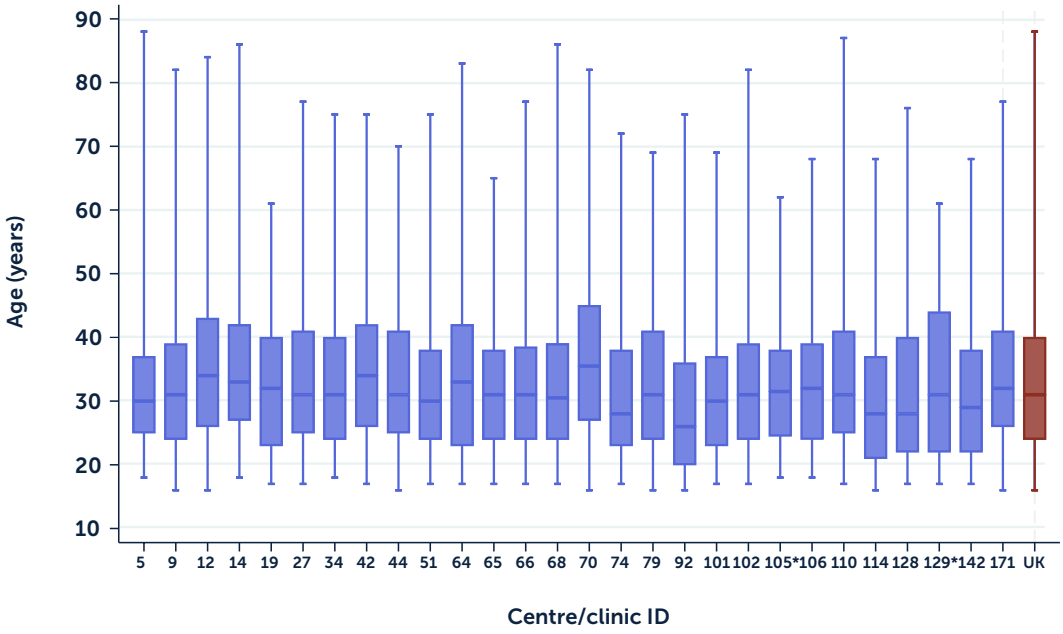
** The chart above shows the proportion of patients who had a valid best FEV_1% and an FEV_1% at annual review, excluding patients under six years of age. Best FEV_1% was considered valid if it was not missing, and the per cent predicted was not more than 0.5% lower than the annual review value. For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

Section 3: Adult centre analysis

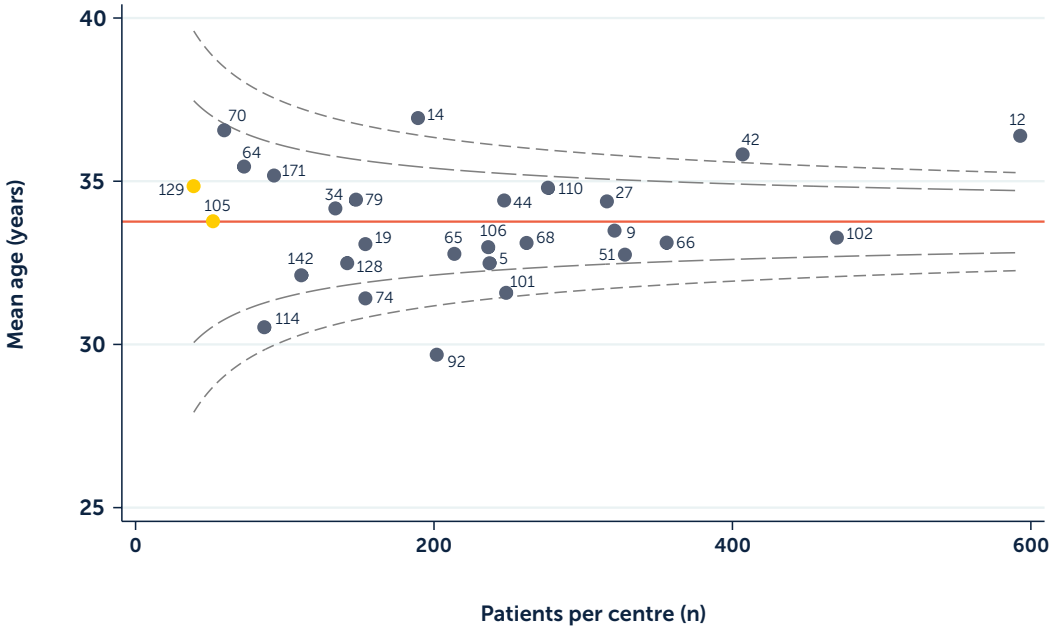
N=6152

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

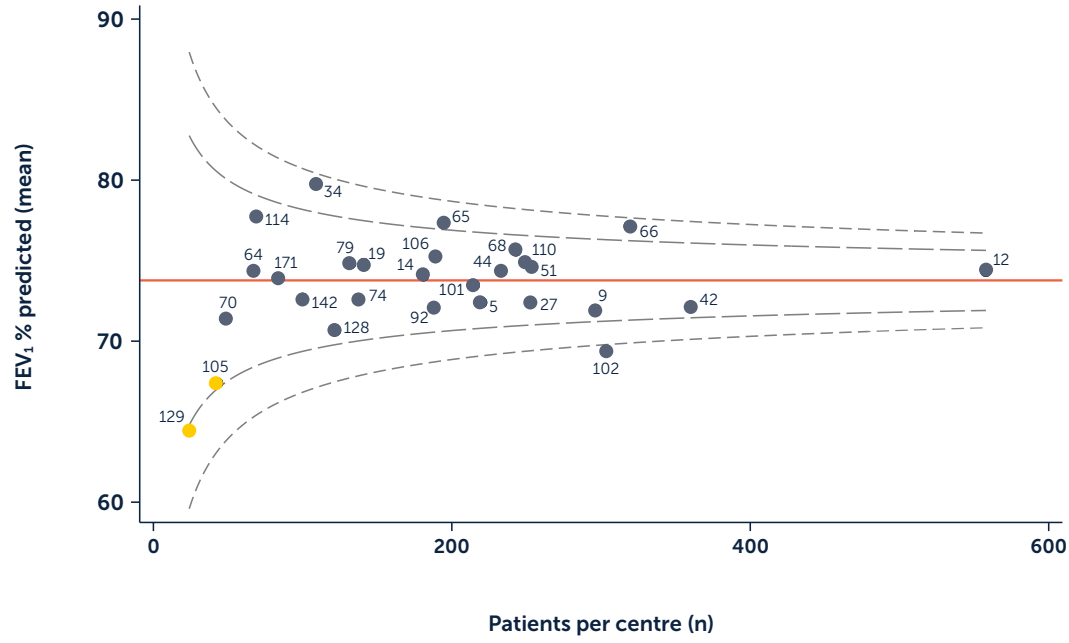
3.1 Age distribution by adults centre/clinic



The funnel plot below shows how the mean age in adult centres compares to the national mean. In 2022 the national mean age of patients at CF centres was 33.8 years.

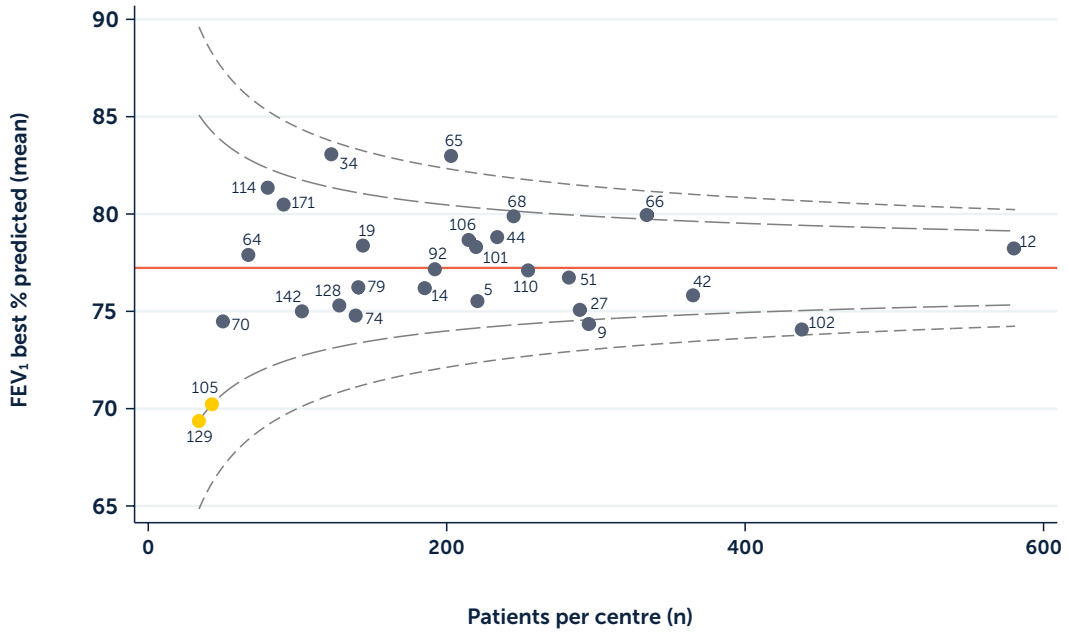


3.2 Age adjusted FEV_1 % predicted at annual review in patients without a history of lung transplant, by adult centre/clinic



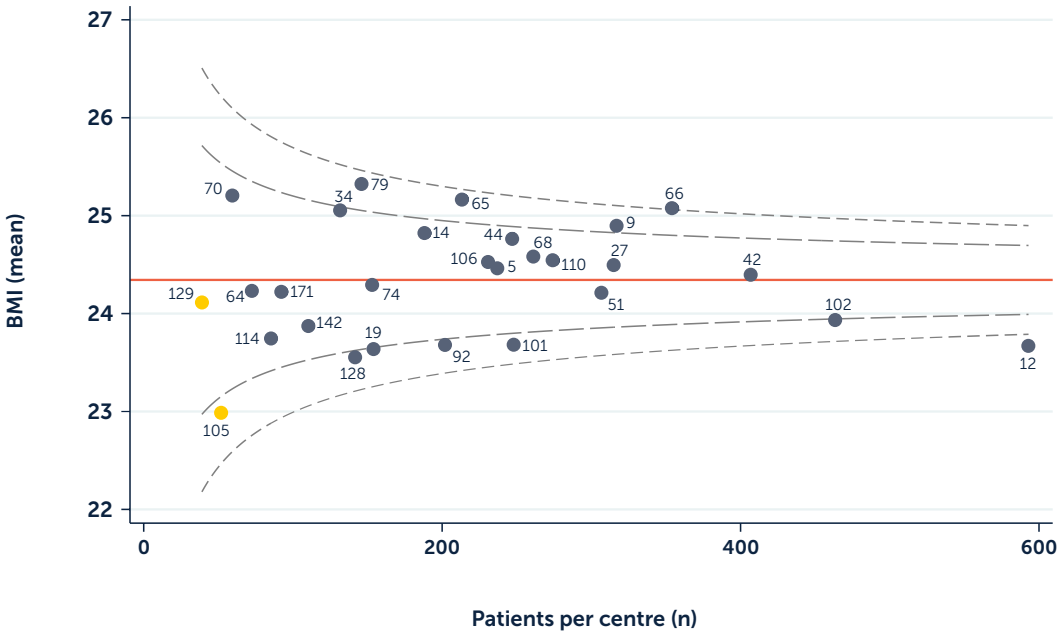
The mean FEV_1% predicted in adult centres/clinics is 73.8%.

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



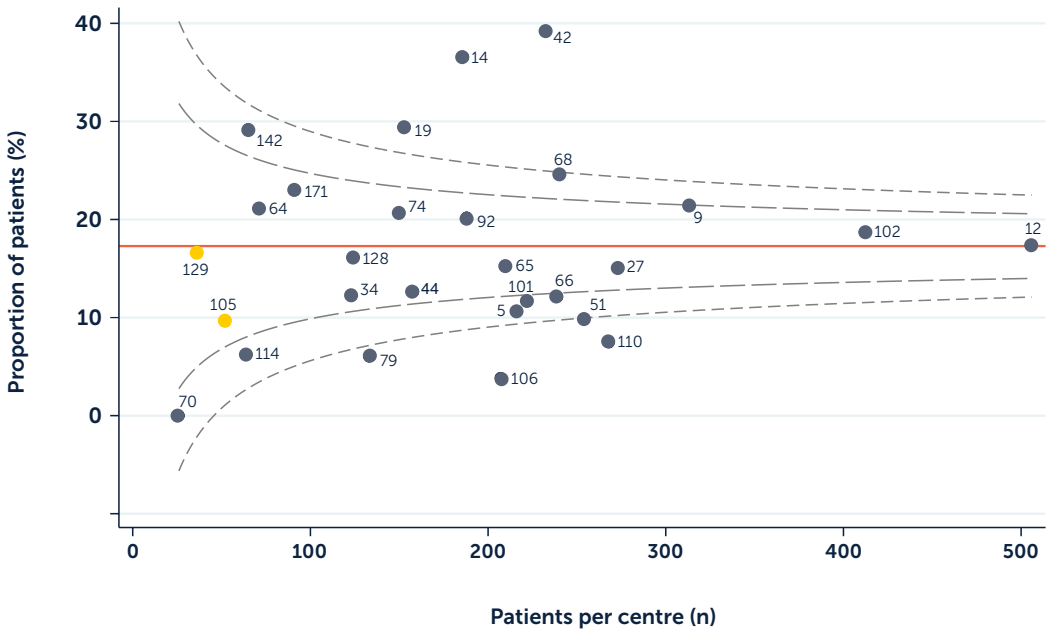
In 2022 the national mean was 77.2%. Where Best FEV_1% predicted was missing, or lower than the FEV_1 at annual review, the FEV_1% value at annual review was used.

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



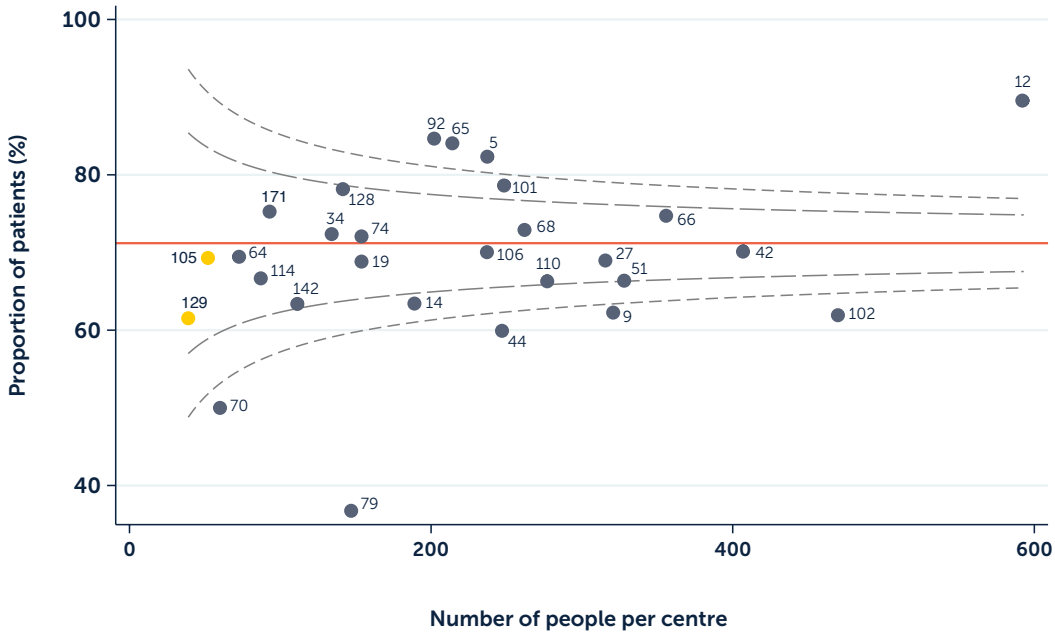
The mean BMI in adult centres/clinics is 24.3.

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



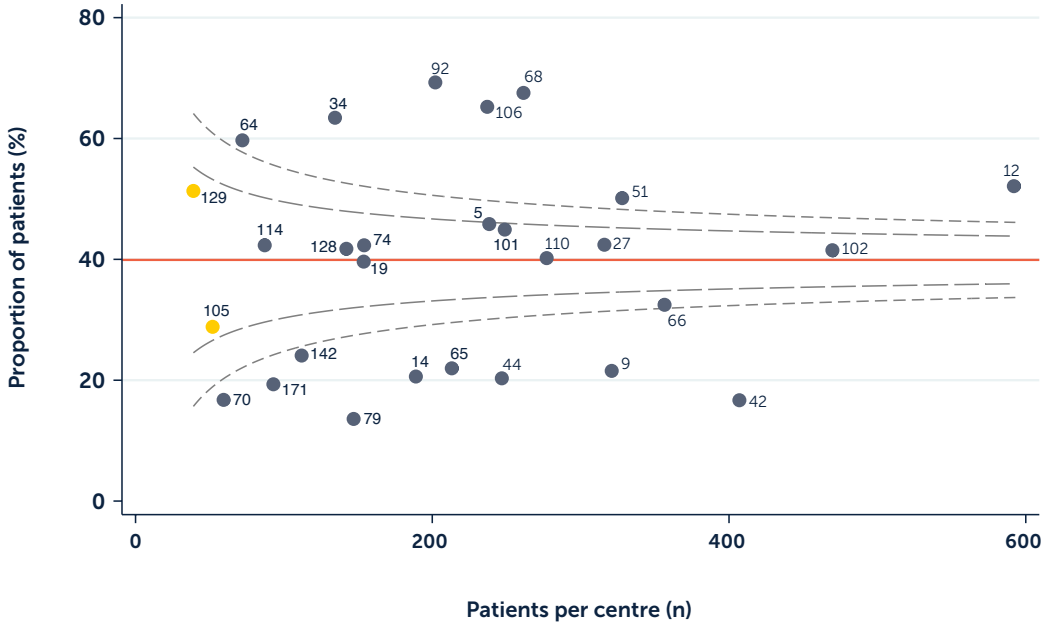
The proportion of patients with chronic *P. aeruginosa* in adult centres/clinics is 17.3%.

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



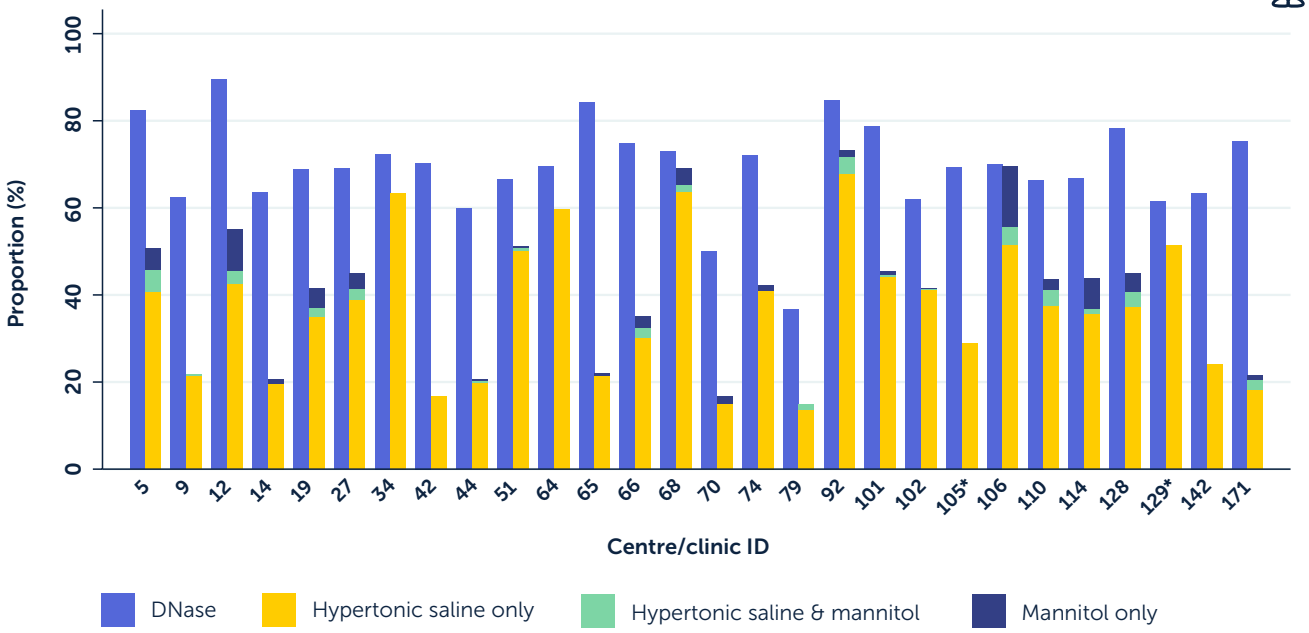
The proportion of patients receiving DNase treatment in adult centres/clinics is 71.2%.

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



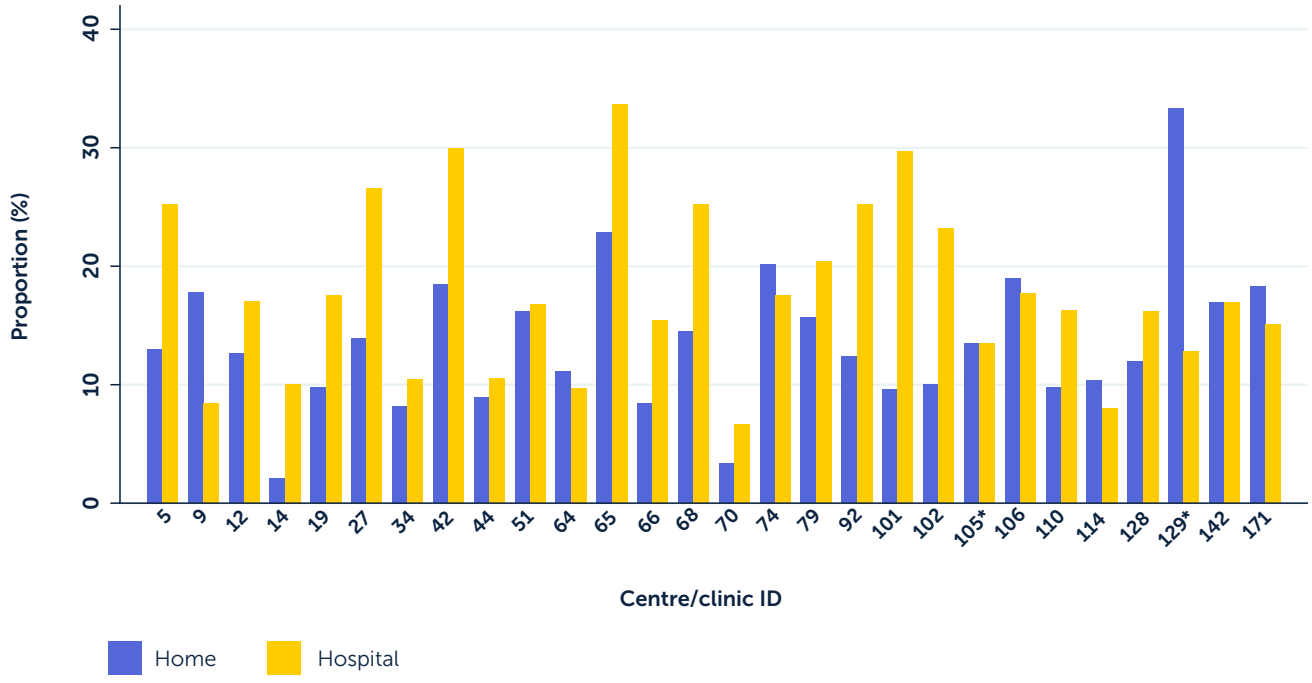
The proportion of patients receiving hypertonic saline or mannitol treatment in adult centres/clinics is 39.9%.

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



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

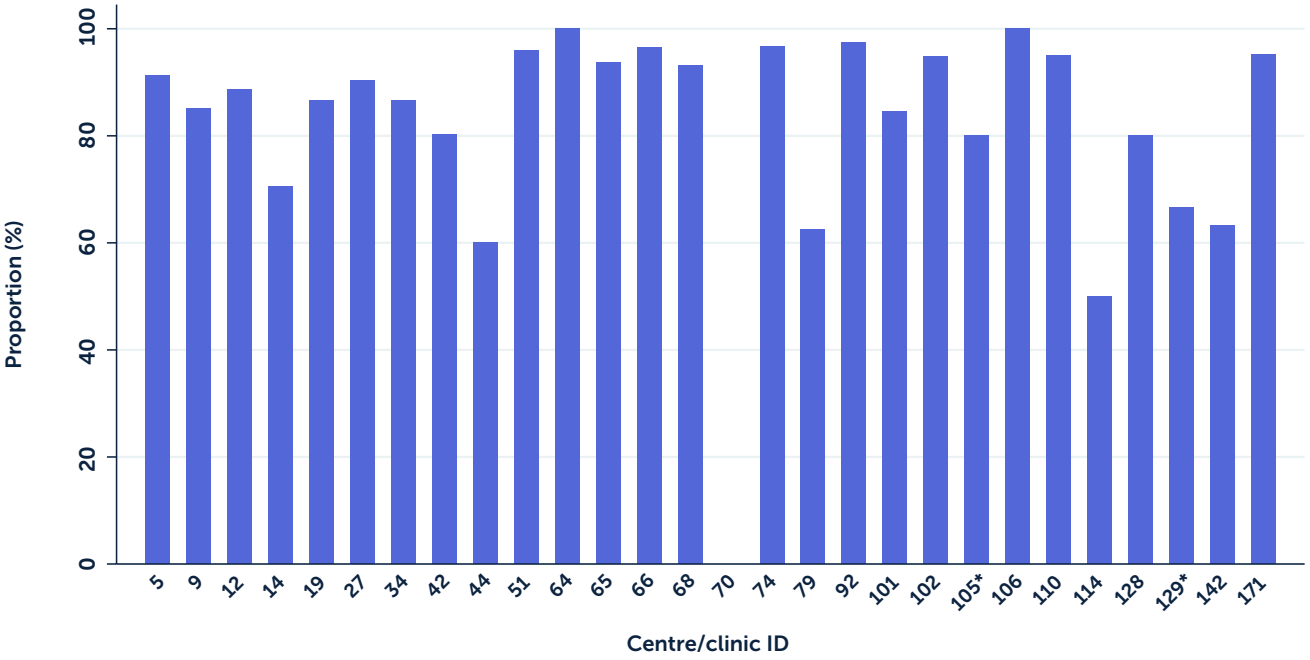
The chart below shows the proportion of patients with at least one IV day at home and/or in hospital. Patients may have a combination of home and hospital IV days.



The proportion of patients in adult centres receiving IV antibiotics at home was 13.6% and in hospital was 17.7%. The proportion receiving any IVs was 23.6%.

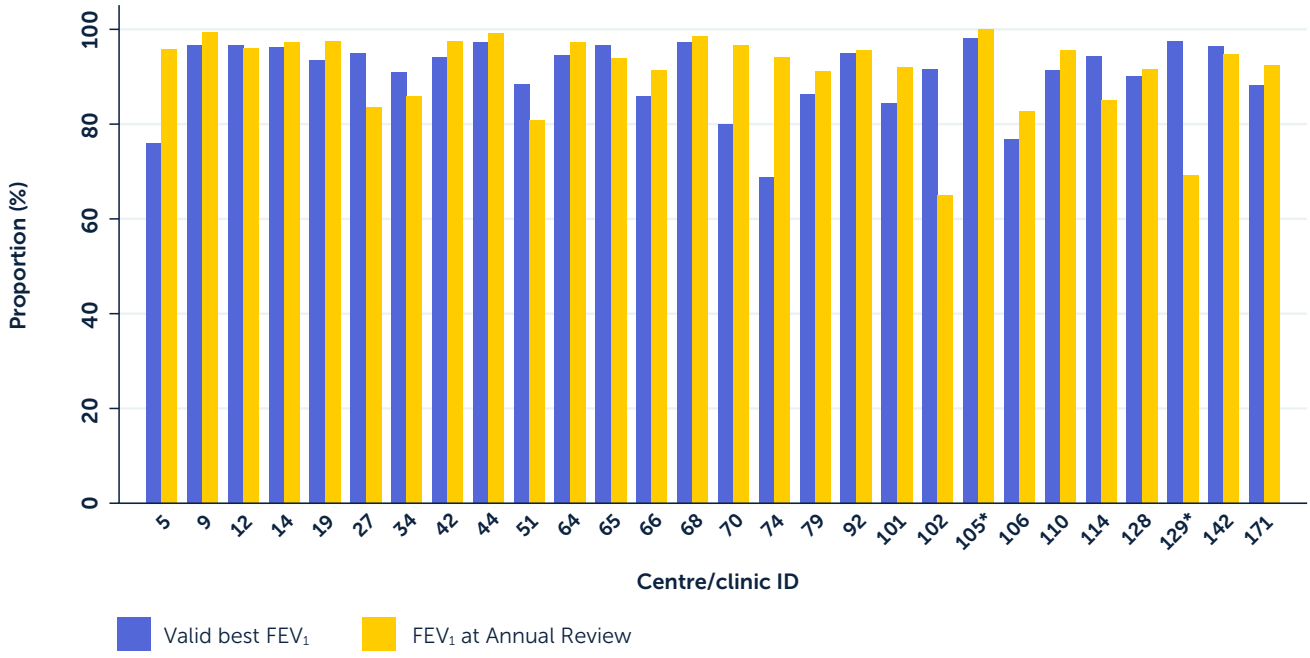
* Standalone clinics.

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



84.6% of patients in adult centres with chronic *P. aeruginosa* received inhaled antibiotics. Centres with fewer than 10 people with chronic *P. aeruginosa* were excluded.

3.11 Data completeness by adult centre/clinic*



* 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.

Glossary

Word/Phrase	Meaning
2022	1 January 2022–31 December 2021.
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).
GERD (gastroesophageal reflux disease)	A chronic symptom of damage caused by stomach acid coming up from the stomach into the oesophagus.
GI bleed	Bleeding in the gastrointestinal tract.
GLI equations	Global Lung Initiative, the equation used for calculating FEV_1% predicted from absolute FEV_1, which takes into account age, gender, height and ethnicity.
H. influenza	*Haemophilus influenza* is a bacterium that can cause serious illness.
Haemoptysis	The coughing up of blood.
Hepatobiliary disease	A liver or biliary disorder.
Heterozygous	Everyone living with cystic fibrosis has two mutations of the gene for CFTR, one inherited from their mother and one from their father. Someone who has two different mutations is heterozygous.

Word/Phrase	Meaning
Homozygous	Everyone living with cystic fibrosis has two mutations of the gene for CFTR, one inherited from their mother and one from their father. If both mutations (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.
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.

Appendix 1: UK CF Registry Committee structure

UK CF Registry Steering Committee

Role	Forename	Surname	Organisation
Director Research & Healthcare Data †	Lucy	Allen	Cystic Fibrosis Trust
Commissioner, England	Kathy	Blacker	NHS England
CF physician - Paediatrics	Malcolm	Brodlie	Newcastle Paediatrics CF Centre
CF Physician - Paediatrics (Outgoing Chair)*	Siobhán	Carr	Royal Brompton Hospital
Analytical team rep †	Susan	Charman	Cystic Fibrosis Trust
Associate Director of Data & QI #	Sarah	Clarke	Cystic Fibrosis Trust
Chair of the Research Committee	Steve	Cunningham	Royal Hospital for Sick Children, Edinburgh
CF Physician - Paediatrics	Gwyneth	Davies	UCL Great Ormond Street Institute of Child Health
CF Physician - Adults (Incoming Chair)*	Jamie	Duckers	All Wales Adult CF Centre, Cardiff
Parent of Child with CF	Catherine	Farrer	N/A
Registry data manager †	Elaine	Gunn	Cystic Fibrosis Trust
Allied Health Professional	Rebecca	Heise	Kings College Adult CF Centre
Cystic Centre Data Manager	Erin	Hodgetts	North West Midlands Adult & Paediatrics CF Centres
Commissioner, Scotland	Roseanne	McDonald	NHS Scotland
Welsh Commissioner	Richard	Palmer	NHS Wales
CF Physician - Adults	Simon	Range	Leicester Adult CF Centre

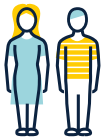
* Chair † Non-voting member # Caldicott guardian

UK CF Registry Research Committee

Role	Forename	Surname	Organisation
Pharmacovigilance PI, CF Physician - Paediatrics	Siobhán	Carr	Royal Brompton Hospital
Analytical team rep †	Susan	Charman	Cystic Fibrosis Trust
Associate Director of Data & QI #	Sarah	Clarke	Cystic Fibrosis Trust
Pharmacovigilance PI, CF physician – paediatrics*	Steve	Cunningham	Royal Hospital for Sick Children, Edinburgh
Registry data manager †	Elaine	Gunn	Cystic Fibrosis Trust
Pharmacovigilance PI, CF Physician - Adults	Dilip	Nazareth	Liverpool Heart and Chest Hospital, Liverpool
Pharmacovigilance PI , CF physician - Adults	Nick	Simmonds	Royal Brompton Hospital
Person with CF	James	Thompson	N/A

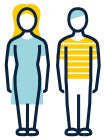
* Chair † Non-voting member # Caldicott guardian

Appendix 2: Centre-level data tables



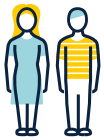
Paediatric centres/clinics providing data in 2022 – ordered alphabetically by country/city

Location	Name	Clinic ID	Total Active	Number with annual review
England				
Birmingham	Birmingham Children's Hospital	104	302	278
Brighton	Royal Alexandra Children's Hospital	172	60	58
Bristol	Bristol Royal Hospital for Children	32	189	175
Cambridge	Addenbrookes Hospital	107	141	136
Cornwall	Royal Cornwall Hospital	94	33	29
Exeter	Royal Devon & Exeter Hospital	96	71	69
Hull	Hull University Teaching Hospitals NHS Trust	111	43	41
Leeds	St James's University Hospital	25	226	215
Leicester	Leicester Royal Infirmary	1	74	62
Liverpool	Alder Hey Children's Hospital	97	308	289
London - Central	Great Ormond Street Hospital for Children	90	198	192
London - East	Royal London Hospital	30	92	82
London - South East	King's College Hospital	17	205	190
London - South West	Royal Brompton Hospital	15	276	262
Manchester	Royal Manchester Children's Hospital	144	333	313
Newcastle	Great North Children's Hospital	59	201	184
North West Midlands	University Hospital of North Midlands	8	97	92
Norwich	Norfolk & Norwich University Hospital	98	62	54
Nottingham	Nottingham University Hospitals	62	161	152
Oxford	John Radcliffe Hospital	22	171	162
Plymouth	Derriford Hospital	139	39	38
Sheffield	Sheffield Children's Hospital	3	150	137
Southampton	Southampton General Hospital	29	237	219
Teeside	James Cook University Hospital	71	55	49
Northern Ireland				
Belfast	Royal Belfast Hospital for Sick Children	60	212	197
Scotland				
Aberdeen	Royal Aberdeen Children's Hospital	75	30	28
Ayr	University Hospital Crosshouse	170	23	5
Dundee	Ninewells Hospital	73	21	20
Edinburgh	Royal Hospital for Sick Children	143	125	72
Glasgow	Royal Hospital for Sick Children	56	160	133
Inverness	Raigmore Hospital	31	16	15
Wales				
Cardiff	Children's Hospital for Wales	72	158	151



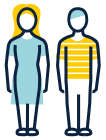
	Age		FEV$_1$% predicted at annual review				Best FEV$_1$% predicted			
Clinic ID	Mean	Median	Number	Mean - unadjusted	Mean - adjusted	Median	Number*	Mean - unadjusted	Mean - adjusted	Median
104	9.0	9.0	176	92.7	92.8	94.6	186	96.8	96.8	99.5
172	8.2	8.8	39	101.2	100.7	102.2	39	104.6	104.1	104.3
32	9.2	9.2	121	94.9	94.8	94.9	123	99.7	99.5	99.4
107	8.9	9.0	91	100.1	99.7	99.0	94	101.3	100.8	100.3
94	9.8	10.0	21	89.6	88.8	89.9	23	91.1	90.7	91.2
96	9.7	9.5	53	92.3	92.0	91.5	54	96.5	96.0	96.3
111	9.2	9.6	29	91.5	91.2	95.7	29	97.5	97.2	101.8
25	9.3	9.7	139	93.0	92.6	95.3	152	98.5	98.0	101.3
1	8.4	7.9	41	95.5	95.2	95.5	41	98.7	98.3	99.1
97	9.4	9.7	152	93.8	93.8	95.8	193	98.7	98.7	98.6
90	9.2	9.5	132	95.5	95.2	98.3	135	98.4	98.1	100.9
30	10.0	11.1	56	100.2	100.3	102.9	65	104.4	104.4	106.9
17	8.6	8.0	105	94.4	94.3	97.4	122	96.8	96.6	97.9
15	9.0	9.2	184	93.9	93.5	94.9	185	101.0	100.6	101.0
144	9.1	9.4	153	92.8	92.8	94.3	193	98.9	98.9	100.3
59	9.0	9.1	120	98.6	98.3	100.1	125	100.6	100.4	101.2
8	10.4	11.3	71	93.0	93.3	94.2	72	97.7	97.9	99.3
98	8.8	8.0	32	93.0	93.0	91.3	33	100.6	100.6	95.3
62	10.1	10.2	116	89.8	89.9	90.8	119	94.6	94.8	95.5
22	9.3	9.8	111	95.8	95.9	97.8	112	98.4	98.5	99.5
139	8.2	8.0	24	92.1	91.4	94.2	24	98.6	97.8	99.4
3	9.4	9.6	95	97.7	97.6	99.1	102	102.4	102.3	102.7
29	9.4	9.4	142	93.6	93.9	95.0	146	98.5	98.8	99.9
71	10.1	11.2	32	92.1	93.2	95.0	33	97.6	98.8	101.4
60	9.2	9.0	129	95.8	95.9	96.1	135	100.2	100.3	99.3
75	7.7	6.1	9	98.2	97.2	98.3	14	104.3	103.8	102.1
170	10.0	11.6	<5	91.0	91.6	78.8	<5	100.3	100.9	104.0
73	8.6	9.6	5	99.9	99.6	103.5	13	100.7	99.9	100.8
143	10.2	10.8	51	95.0	95.1	94.4	55	99.4	99.6	99.8
56	8.9	9.3	72	99.7	99.2	97.2	95	102.5	102.0	100.9
31	9.3	8.8	11	96.4	96.0	94.7	11	103.0	102.5	103.8
72	9.6	10.7	101	91.5	91.6	91.8	105	96.9	96.9	96.8

* 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 2022 – ordered alphabetically by country/city

			BMI			
Location	Name	Clinic ID	Number	Mean - unadjusted	Mean - adjusted	Median
England						
Birmingham	Birmingham Children's Hospital	104	249	53.1	53.1	52.2
Brighton	Royal Alexandra Children's Hospital	172	49	54.5	54.5	52.2
Bristol	Bristol Royal Hospital for Children	32	161	56.9	56.9	60.5
Cambridge	Addenbrookes Hospital	107	126	53.8	53.8	54.3
Cornwall	Royal Cornwall Hospital	94	28	66.3	66.3	69.3
Exeter	Royal Devon & Exeter Hospital	96	66	55.6	55.5	54.1
Hull	Hull University Teaching Hospitals NHS Trust	111	39	60.2	60.2	63.7
Leeds	St James's University Hospital	25	186	55.4	55.4	56.2
Leicester	Leicester Royal Infirmary	1	56	57.6	57.7	63.3
Liverpool	Alder Hey Children's Hospital	97	266	60.8	60.8	62.2
London - Central	Great Ormond Street Hospital for Children	90	179	49.0	49.0	46.6
London - East	Royal London Hospital	30	76	56.3	56.2	53.8
London - South East	King's College Hospital	17	167	55.3	55.4	57.1
London - South West	Royal Brompton Hospital	15	247	53.8	53.9	52.8
Manchester	Royal Manchester Children's Hospital	144	281	54.7	54.7	54.8
Newcastle	Great North Children's Hospital	59	166	61.2	61.2	66.8
North West Midlands	University Hospital of North Midlands	8	90	56.4	56.3	56.3
Norwich	Norfolk & Norwich University Hospital	98	52	62.1	62.2	65.7
Nottingham	Nottingham University Hospitals	62	139	54.5	54.4	53.9
Oxford	John Radcliffe Hospital	22	151	57.9	57.9	61.5
Plymouth	Derriford Hospital	139	36	60.7	60.8	60.6
Sheffield	Sheffield Children's Hospital	3	130	56.9	56.9	59.1
Southampton	Southampton General Hospital	29	201	58.4	58.3	63.2
Teeside	James Cook University Hospital	71	45	63.5	63.4	68.3
Northern Ireland						
Belfast	Royal Belfast Hospital for Sick Children	60	178	57.2	57.1	59.0
Scotland						
Aberdeen	Royal Aberdeen Children's Hospital	75	24	54.5	54.6	57.6
Ayr	University Hospital Crosshouse	170	5	65.0	64.9	69.7
Dundee	Ninewells Hospital	73	15	53.4	53.4	52.3
Edinburgh	Royal Hospital for Sick Children	143	70	60.4	60.3	63.9
Glasgow	Royal Hospital for Sick Children	56	111	55.7	55.7	56.7
Inverness	Raigmore Hospital	31	14	53.5	53.6	51.2
Wales						
Cardiff	Children's Hospital for Wales	72	145	56.4	56.4	57.4



	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	8	2.9	64	23.0	165	59.4	111	39.9	8	100.0
172	<5	3.4	11	19.0	41	70.7	21	36.2	<5	100.0
32	<5	1.2	21	12.0	139	79.4	112	64.0	<5	100.0
107	<5	2.3	28	20.6	71	52.2	91	66.9	<5	100.0
94	<5	3.4	<5	6.9	14	48.3	11	37.9	<5	100.0
96	0	0.0	9	13.0	64	92.8	65	94.2	0	0.0
111	<5	9.8	11	26.8	20	48.8	10	24.4	<5	25.0
25	8	3.8	41	19.1	135	62.8	26	12.1	8	100.0
1	0	0.0	17	27.4	43	69.4	14	22.6	0	0.0
97	8	2.8	63	21.8	154	53.3	48	16.6	6	75.0
90	7	3.7	47	24.5	134	69.8	91	47.4	7	100.0
30	7	8.6	22	26.8	66	80.5	70	85.4	7	100.0
17	11	6.0	39	20.5	120	63.2	50	26.3	11	100.0
15	6	2.3	59	22.5	196	74.8	147	56.1	6	100.0
144	18	5.8	53	16.9	170	54.3	105	33.5	17	94.4
59	14	8.1	40	21.7	98	53.3	27	14.7	14	100.0
8	0	0.0	39	42.4	72	78.3	29	31.5	0	0.0
98	<5	1.9	<5	7.4	37	68.5	10	18.5	<5	100.0
62	<5	2.1	22	14.5	95	62.5	47	30.9	<5	33.3
22	<5	0.6	24	14.8	114	70.4	63	38.9	<5	100.0
139	0	0.0	<5	5.3	26	68.4	8	21.1	0	0.0
3	<5	0.7	34	24.8	93	67.9	34	24.8	<5	100.0
29	<5	1.5	32	14.6	141	64.4	37	16.9	<5	66.7
71	<5	2.0	8	16.3	30	61.2	7	14.3	0	0.0
60	<5	1.1	13	6.6	160	81.2	24	12.2	<5	100.0
75	<5	3.6	5	17.9	12	42.9	<5	14.3	<5	100.0
170	0	0.0	0	0.0	0	0.0	0	0.0	0	0.0
73	<5	5.6	<5	5.0	6	30.0	<5	10.0	0	0.0
143	<5	4.3	13	18.1	49	68.1	13	18.1	<5	100.0
56	<5	0.8	34	25.6	30	22.6	56	42.1	<5	100.0
31	<5	13.3	<5	6.7	5	33.3	<5	13.3	<5	100.0
72	<5	2.6	11	7.3	118	78.1	138	91.4	<5	75.0

* Redacted to adhere to statistical disclosure guidelines.

Appendix 2: Centre-level data tables

Adult centres/clinics providing data in 2022 – ordered alphabetically by country/city

Location	Name	Clinic ID	Total Active	Number with annual review
England				
Birmingham	Birmingham Heartlands Hospital	27	339	316
Bristol	Bristol Royal Infirmary	106	250	237
Cambridge	Royal Papworth Hospital	51	370	328
Cornwall	Royal Cornwall Hospital	129	41	39
Exeter	Royal Devon & Exeter Hospital	34	142	134
Frimley	Frimley Park Hospital	19	162	154
Leeds	St James's University Hospital	42	416	407
Leicester	Glenfield Hospital	142	118	112
Liverpool	Liverpool Heart and Chest Hospital	66	378	356
London - East	St Bartholomew's Hospital	92	223	202
London - South East	King's College Hospital	5	260	238
London - South East	University Hospital Lewisham	105	57	52
London - South West	Royal Brompton Hospital	12	604	593
Manchester	Wythenshawe Hospital	102	481	470
Newcastle	Royal Victoria Infirmary	9	333	321
North West Midlands	University Hospital of North Midlands	74	160	154
Norwich	Norfolk & Norwich University Hospital	114	87	87
Nottingham	Nottingham University Hospitals	101	255	249
Oxford	Oxford University Hospitals	128	158	142
Plymouth	Derriford Hospital	64	73	72
Sheffield	Northern General Hospital	65	221	214
Southampton	Southampton General Hospital	110	311	277
York and Hull	York Hospital	171	97	93
Northern Ireland				
Belfast	Belfast City Hospital	14	279	189
Scotland				
Aberdeen	Aberdeen Royal Infirmary	70	74	60
Edinburgh	Western General Hospital	44	260	247
Glasgow	Queen Elizabeth University Hospital	79	224	147
Wales				
Llandough	Llandough Hospital	68	307	262

	Age		FEV$_1$% predicted at annual review				Best FEV$_1$% predicted			
Clinic ID	Mean	Median	Number	Mean - unadjusted	Mean - adjusted	Median	Number*	Mean - unadjusted	Mean - adjusted	Median
27	34.4	31.8	253	72.0	72.4	72.6	290	74.7	75.1	76.8
106	33.0	32.0	189	75.0	75.2	79.7	216	78.7	78.6	82.1
51	32.8	30.5	254	74.9	74.6	76.2	282	77.4	76.7	80.4
129	34.9	31.8	24	66.2	64.5	67.2	34	69.9	69.4	71.3
34	34.2	31.8	109	79.7	79.8	84.7	123	82.8	83.1	86.3
19	33.1	32.5	141	75.1	74.8	74.5	144	78.7	78.4	79.2
42	35.8	34.2	360	71.2	72.1	74.3	365	74.9	75.8	78.7
142	32.1	29.7	100	74.1	72.7	78.1	103	76.7	75.0	81.5
66	33.1	31.9	319	77.1	77.2	78.7	335	80.0	80.0	81.2
92	29.7	26.6	188	75.1	72.1	77.2	192	80.4	77.2	81.8
5	32.5	30.6	220	72.7	72.4	76.6	221	75.9	75.5	79.7
105	33.8	31.8	43	67.2	67.4	68.8	43	70.1	70.3	70.6
12	36.4	34.6	558	72.9	74.3	73.8	581	76.8	78.2	77.3
102	33.3	31.0	304	69.1	69.4	69.7	438	73.9	74.1	74.8
9	33.5	31.0	296	72.5	72.0	76.7	296	74.9	74.3	78.5
74	31.4	28.0	138	74.3	72.6	77.4	139	76.8	74.8	83.5
114	30.5	28.8	69	79.6	77.7	83.3	80	83.5	81.4	88.1
101	31.6	30.0	215	74.6	73.5	76.1	220	79.6	78.3	82.3
128	32.5	28.1	122	72.3	70.7	72.5	128	77.3	75.3	77.8
64	35.4	33.5	67	74.7	74.4	79.9	67	78.3	77.9	81.4
65	32.8	31.2	195	77.7	77.4	82.1	203	83.5	83.0	87.8
110	34.8	31.8	250	75.0	74.9	76.4	255	77.2	77.1	78.4
171	35.2	32.0	84	73.3	73.9	72.5	91	80.1	80.5	80.7
14	36.9	33.9	181	72.9	74.1	75.8	186	75.0	76.2	78.8
70	36.6	35.9	48	70.8	71.4	69.3	50	73.9	74.5	71.6
44	34.4	31.9	233	74.4	74.4	76.5	234	78.9	78.8	82.0
79	34.4	31.9	132	74.6	74.8	78.5	141	75.8	76.2	80.9
68	33.1	31.0	243	76.1	75.6	79.9	245	80.5	79.9	84.5

* 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 2022 – ordered alphabetically by country/city

			BMI			
Location	Name	Clinic ID	Number	Mean - unadjusted	Mean - adjusted	Median
England						
Birmingham	Birmingham Heartlands Hospital	27	315	24.6	24.5	24.2
Bristol	Bristol Royal Infirmary	106	231	24.5	24.5	24.1
Cambridge	Royal Papworth Hospital	51	307	24.1	24.2	23.5
Cornwall	Royal Cornwall Hospital	129	39	24.2	24.1	22.0
Exeter	Royal Devon & Exeter Hospital	34	132	25.1	25.1	24.5
Frimley	Frimley Park Hospital	19	154	23.6	23.6	23.2
Leeds	St James's University Hospital	42	407	24.6	24.4	24.2
Leicester	Glenfield Hospital	142	111	23.7	23.9	23.8
Liverpool	Liverpool Heart and Chest Hospital	66	355	25.0	25.1	24.2
London - East	St Bartholomew's Hospital	92	202	23.3	23.7	22.5
London - South East	King's College Hospital	5	237	24.4	24.5	23.4
London - South East	University Hospital Lewisham	105	52	23.0	23.0	22.2
London - South West	Royal Brompton Hospital	12	593	23.9	23.7	23.6
Manchester	Wythenshawe Hospital	102	464	23.9	23.9	23.2
Newcastle	Royal Victoria Infirmary	9	317	24.9	24.9	24.0
North West Midlands	University Hospital of North Midlands	74	153	24.1	24.3	23.7
Norwich	Norfolk & Norwich University Hospital	114	86	23.4	23.7	22.8
Nottingham	Nottingham University Hospitals	101	248	23.5	23.7	22.5
Oxford	Oxford University Hospitals	128	142	23.4	23.6	22.8
Plymouth	Derriford Hospital	64	72	24.2	24.2	24.2
Sheffield	Northern General Hospital	65	214	25.1	25.2	24.2
Southampton	Southampton General Hospital	110	275	24.6	24.5	23.7
York and Hull	York Hospital	171	93	24.3	24.2	23.8
Northern Ireland						
Belfast	Belfast City Hospital	14	189	25.1	24.8	24.4
Scotland						
Aberdeen	Aberdeen Royal Infirmary	70	60	25.5	25.2	24.8
Edinburgh	Western General Hospital	44	247	24.8	24.8	24.1
Glasgow	Queen Elizabeth University Hospital	79	146	25.4	25.3	24.3
Wales						
Llandough	Llandough Hospital	68	261	24.5	24.6	23.8

	Chronic *pseudomonas*		Having at least 1 IV days		Receiving DNase treatment		Receiving hypertonic saline or mannitol treatment		Inhaled antibiotic use among patients with chronic *pseudomonas*	
Clinic ID	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)
27	41	15.0	99	31.3	218	69.0	134	42.4	37	90.2
106	8	3.9	67	28.3	166	70.0	155	65.4	8	100.0
51	25	9.8	79	24.1	218	66.5	165	50.3	24	96.0
129	6	16.7	14	35.9	24	61.5	20	51.3	<5	66.7
34	15	12.2	16	11.9	97	72.4	85	63.4	13	86.7
19	45	29.4	34	22.1	106	68.8	61	39.6	39	86.7
42	91	39.2	136	33.4	286	70.3	68	16.7	73	80.2
142	19	29.2	31	27.7	71	63.4	27	24.1	12	63.2
66	29	12.1	72	20.2	266	74.7	116	32.6	28	96.6
92	38	20.2	55	27.2	171	84.7	140	69.3	37	97.4
5	23	10.6	67	28.2	196	82.4	109	45.8	21	91.3
105	5	9.6	11	21.2	36	69.2	15	28.8	<5	80.0
12	88	17.4	132	22.3	531	89.5	309	52.1	78	88.6
102	77	18.7	127	27.0	291	61.9	194	41.3	73	94.8
9	67	21.4	63	19.6	200	62.3	69	21.5	57	85.1
74	31	20.7	42	27.3	111	72.1	65	42.2	30	96.8
114	<5	6.2	12	13.8	58	66.7	37	42.5	<5	50.0
101	26	11.7	79	31.7	196	78.7	112	45.0	22	84.6
128	20	16.1	28	19.7	111	78.2	59	41.5	16	80.0
64	15	21.1	11	15.3	50	69.4	43	59.7	15	100.0
65	32	15.2	94	43.9	180	84.1	47	22.0	30	93.8
110	20	7.5	55	19.9	184	66.4	111	40.1	19	95.0
171	21	23.1	23	24.7	70	75.3	18	19.4	20	95.2
14	68	36.6	19	10.1	120	63.5	39	20.6	48	70.6
70	0	0.0	5	8.3	30	50.0	10	16.7	0	0.0
44	20	12.7	32	13.0	148	59.9	50	20.2	12	60.0
79	8	6.0	36	24.5	54	36.7	20	13.6	5	62.5
68	59	24.6	76	29.0	191	72.9	177	67.6	55	93.2

* Redacted to adhere to statistical disclosure guidelines.

Appendix 3: Full list of mutations in the UK CF population

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

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

Nucleotide	Protein	Legacy name	N	%
c.1521_1523delCTT	p.Phe508del	F508del	9941	89.2
c.350G->A	p.Arg117His	R117H	704	6.3
c.1652G->A	p.Gly551Asp	G551D	636	5.7
c.1624G->T	p.Gly542X	G542X	407	3.7
c.489+1G->T		621+1G->T	291	2.6
c.3909C->G	p.Asn1303Lys	N1303K	177	1.6
c.1585-1G->A		1717-1G->A	176	1.6
c.1766+1G->A		1898+1G->A	159	1.4
c.3454G->C	p.Asp1152His	D1152H	151	1.4
c.200C->T	p.Pro67Leu	P67L	148	1.3
c.3140-26A->G		3272-26A->G	127	1.1
c.3528delC	p.Lys1177SerfsX15	3659delC	122	1.1
c.1679G->C	p.Arg560Thr	R560T	104	0.9
c.1519_1521delATC	p.Ile507del	I507del	94	0.8
c.1477C->T	p.Gln493X	Q493X	94	0.8
c.3717+12191C->T		3849+10kbC->T	92	0.8
c.1657C->T	p.Arg553X	R553X	87	0.8
c.254G->A	p.Gly85Glu	G85E	85	0.8
c.2657+5G->A		2789+5G->A	82	0.7
c.178G->T	p.Glu60X	E60X	80	0.7
c.1022_1023insTC	p.Phe342HisfsX28	1154insTC	75	0.7
c.3846G->A	p.Trp1282X	W1282X	67	0.6
c.1364C->A	p.Ala455Glu	A455E	58	0.5
c.1646G->A	p.Ser549Asn	S549N	58	0.5
c.617T->G	p.Leu206Trp	L206W	53	0.5
c.948delT	p.Phe316LeufsX12	1078delT	52	0.5
c.2052delA	p.Lys684AsnfsX38	2184delA	49	0.4
c.1040G->C	p.Arg347Pro	R347P	44	0.4
c.2657+2_2657+3insA		2789+2insA	39	0.3
c.579+3A->G		711+3A->G	37	0.3
c.3718-2477C->T		3849+10kbC->T	34	0.3
c.1558G->T	p.Val520Phe	V520F	32	0.3
c.3484C->T	p.Arg1162X	R1162X	30	0.3
c.1040G->A	p.Arg347His	R347H	30	0.3
c.1000C->T	p.Arg334Trp	R334W	28	0.3
c.2988+1G->A		3120+1G->A	28	0.3
c.1367T->C	p.Val456Ala	V456A	27	0.2
c.1753G->T	p.Glu585X	E585X	27	0.2

Nucleotide	Protein	Legacy name	N	%
c.3472C->T	p.Arg1158X	R1158X	25	0.2
c.1055G->A	p.Arg352Gln	R352Q	25	0.2
c.1523T->G	p.Phe508Cys	F508C	24	0.2
c.1006_1007insG	p.Ile336SerfsX28	1138insG	23	0.2
c.2583delT	p.Phe861LeufsX3	2711delT	22	0.2
c.2490+1G->A		2622+1G->A	22	0.2
c.1705T->G	p.Tyr569Asp	Y569D	22	0.2
c.1393-1G->A		1525-1G->A	21	0.2
c.2125C->T	p.Arg709X	R709X	21	0.2
c.3873G->C	p.Gln1291His	Q1291H	21	0.2
c.3197G->A	p.Arg1066His	R1066H	21	0.2
c.2052_2053insA	p.Gln685ThrfsX4	2184insA	20	0.2
c.532G->A	p.Gly178Arg	G178R	19	0.2
c.349C->T	p.Arg117Cys	R117C	19	0.2
c.1210-12[5] (AJ574948.1:g.152T[5])		5T	18	0.2
c.2834C->T	p.Ser945Leu	S945L	18	0.2
c.3806T->A	p.Ile1269Asn	I1269N	18	0.2
c.658C->T	p.Gln220X	Q220X	16	0.1
c.3737C->T	p.Thr1246Ile	T1246I	15	0.1
c.579+1G->T		711+1G->T	15	0.1
c.2537G->A	p.Trp846X	W846X	13	0.1
c.2875delG	p.Ala959HisfsX9	3007delG	13	0.1
c.292C->T	p.Gln98X	Q98X	13	0.1
c.1029delC	p.Cys343X	1161delC	12	0.1
c.1329_1330insAGAT	p.Ile444ArgfsX3	1461ins4	12	0.1
c.2051_2052delAAinsG	p.Lys684SerfsX38	2183AA->G or 2183delAA->G	11	0.1
c.[1210–12[5];1210-34TG[12]]		5T;TG12	11	0.1
c.3196C->T	p.Arg1066Cys	R1066C	11	0.1
c.1466C->A	p.Ser489X	S489X	11	0.1
c.3705T->G	p.Ser1235Arg	S1235R	10	0.1
c.2988G->A		3120G->A	10	0.1
c.3761T->G	p.Leu1254X	L1254X	10	0.1
c.1679+1G->C		1811+1G->C	10	0.1
c.3468G->A		3600G->A	9	0.1
c.54-5940_273+10250del21kb	p.Ser18ArgfsX16	CFTRdele2,3	9	0.1
c.1675G->A	p.Ala559Thr	A559T	9	0.1

Nucleotide	Protein	Legacy name	N	%
c.3208C->T	p.Arg1070Trp	R1070W	9	0.1
c.709C->G	p.Gln237Glu	Q237E	8	0.1
c.1687T->A	p.Tyr563Asn	Y563N	8	0.1
c.1645A->C or c.1647T->G or c.1647T->A	p.Ser549Arg	S549R	8	0.1
c.494T->C	p.Leu165Ser	L165S	8	0.1
c.224G->A	p.Arg75Gln	R75Q	8	0.1
c.695T->A	p.Val232Asp	V232D	8	0.1
c.2353C->T	p.Arg785X	R785X	8	0.1
c.[1210–12[5];1210-34TG[13]]		5T;TG13	7	0.1
c.3353C->T	p.Ser1118Phe	S1118F	7	0.1
c.262_263delTT	p.Leu88IlefsX22	394delTT	7	0.1
c.2012delT	p.Leu671X	2143delT	7	0.1
c.1986_1989delAACT	p.Thr663ArgfsX8	2118del4	7	0.1
c.1721C->A	p.Pro574His	P574H	7	0.1
c.223C->T	p.Arg75X	R75X	6	0.1
c.2128A->T	p.Lys710X	K710X	6	0.1
c.1766+1G->T		1898+1G->T	6	0.1
c.4196_4197delTC	p.Cys1400X	4326delTC	6	0.1
c.2900T->C	p.Leu967Ser	L967S	6	0.1
c.[1210–12[5];1210-34TG[11]]		5T;TG11	5	0.0
c.3718-1G->A		3850-1G->A	5	0.0
c.349C->G	p.Arg117Gly	R117G	5	0.0
c.2290C->T	p.Arg764X	R764X	5	0.0
c.3848G->T	p.Arg1283Met	R1283M	5	0.0
c.3292T->C	p.Trp1098Arg	W1098R	5	0.0
c.3884_3885insT	p.Ser1297PhefsX5	4016insT	5	0.0
c.1116+1G->A		1248+1G->A	5	0.0
c.443T->C	p.Ile148Thr	I148T	5	0.0
c.2991G->C	p.Leu997Phe	L997F	5	0.0
c.1538A->G	p.Asp513Gly	D513G	5	0.0
c.2551C->T	p.Arg851X	R851X	5	0.0
c.(743+1_744-1)_(1584+1_1585-1)dup		CFTRdup6b-10	<5	-
c.429delT	p.Phe143LeufsX10	557delT	<5	-
c.3964-78_4242+577del		CFTRdele22,23	<5	-
c.3095A->G	p.Tyr1032Cys	Y1032C	<5	-
c.1585-8G->A		1717-8G->A	<5	-
c.2249C->T	p.Pro750Leu	P750L	<5	-
c.1680A->C	p.Arg560Ser	R560S	<5	-
c.2215delG	p.Val739TyrfsX16	2347delG	<5	-
c.1393-2A->G		1525-2A->G	<5	-
c.2896delA	p.Thr966ArgfsX2	3028delA	<5	-
c.2464G->T	p.Glu822X	E822X	<5	-
c.595C->T	p.His199Tyr	H199Y	<5	-

Nucleotide	Protein	Legacy name	N	%
c.850dupA	p.Met284AsnfsX3	977insA	<5	-
c.1046C->T	p.Ala349Val	A349V	<5	-
c.2600_2601insA	p.Val868SerfsX28	2732insA	<5	-
c.2491G->T	p.Glu831X	E831X	<5	-
c.4004T->C	p.Leu1335Pro	L1335P	<5	-
c.3988C->T	p.Gln1330X	Q1330X	<5	-
c.165-3C>T		297-3C->T	<5	-
c.4147_4148insA	p.Ile1383AsnfsX3	4279insA	<5	-
c.1584G->A	p.Glu528Glu	1716G/A	<5	-
c.1545_1546delTA	p.Tyr515X	1677delTA	<5	-
c.2909G->A	p.Gly970Asp	G970D	<5	-
c.1679G->A	p.Arg560Lys	R560K	<5	-
c.1572C->A	p.Cys524X	C524X	<5	-
c.350G->T	p.Arg117Leu	R117L	<5	-
c.274G->A	p.Glu92Lys	E92K	<5	-
c.577G->T	p.Glu193X	E193X	<5	-
c.1651G->A	p.Gly551Ser	G551S	<5	-
c.3080T->C	p.Ile1027Thr	I1027T	<5	-
c.1736A->G	p.Asp579Gly	D579G	<5	-
c.1505T>C	p.Ile502Thr	I502T	<5	-
c.4111G->T	p.Glu1371X	E1371X	<5	-
c.2374C->T	p.Arg792X	R792X	<5	-
c.91C->T	p.Arg31Cys	R31C	<5	-
c.3659delC	p.Thr1220LysfsX8	3791delC	<5	-
c.233dupT	p.Trp79LeufsX32	365-366insT	<5	-
c.3872A->G	p.Gln1291Arg	Q1291R	<5	-
c.3752G->A	p.Ser1251Asn	S1251N	<5	-
c.1327G->T	p.Asp443Tyr	D443Y	<5	-
c.328G->C	p.Asp110His	D110H	<5	-
c.1724T->A	p.Phe575Tyr	F575Y	<5	-
c.4046G->A	p.Gly1349Asp	G1349D	<5	-
c.442delA	p.Ile148LeufsX5	574delA	<5	-
c.3266G->A	p.Trp1089X	W1089X	<5	-
c.3297C->A	p.Phe1099Leu	F1099L	<5	-
c.3908delA	p.Asn1303ThrfsX25	4040delA	<5	-
c.2668C->T	p.Gln890X	Q890X	<5	-
c.296C->T	p.Pro99Leu	P99L	<5	-
c.79G->T	p.Gly27X	G27X	<5	-
c.1046C>T	p.Ala349Val	A349V	<5	-
c.1766+1G->C		1898+1G->C	<5	-
c.1117-1G>A		1249-1G->A	<5	-
c.4077_4080delTGTTinsAA	p.Val1360delfsX?	4209TGTT->AA	<5	-
c.1682C->A	p.Ala561Glu	A561E	<5	-
c.3476C->T	p.Ser1159Phe	S1159F	<5	-
c.3158C->T	p.Thr1053Ile	T1053I	<5	-
c.3882_3885delTATT	p.Ile1295PhefsX32	4010del4	<5	-

Nucleotide	Protein	Legacy name	N	%
c.1001G>A	p.Arg334Gln	R334Q	<5	-
c.1727G->C	p.Gly576Ala	G576A	<5	-
c.2260G->A	p.Val754Met	V754M	<5	-
c.3017C->A	p.Ala1006Glu	A1006E	<5	-
c.2780T->C	p.Leu927Pro	L927P	<5	-
c.164+2T>C		296+2T->C	<5	-
c.601G->A	p.Val201Met	V201M	<5	-
c.3475T->C	p.Ser1159Pro	S1159P	<5	-
c.509G->A	p.Arg170His	R170H	<5	-
c.1007T->A	p.Ile336Lys	I336K	<5	-
c.3763T->C	p.Ser1255Pro	S1255P	<5	-
c.1766+5G->T		1898+5G->T	<5	-
c.3205G->A	p.Gly1069Arg	G1069R	<5	-
c.1679+1.6kbA->G		1811+1.6kbA->G	<5	-
c.(53+1_54-1)_(489+1_490-1) del		CFTRdele2-4	<5	-
c.2855T->C	p.Met952Thr	M952T	<5	-
c.220C->T	p.Arg74Trp	R74W	<5	-
c.1135G->T	p.Glu379X	E379X	<5	-
c.(273+1_274-1)_ (1679+1_1680-1)del		CFTRdele4-11	<5	-
c.3310G->T	p.Glu1104X	E1104X	<5	-
c.2195T->G	p.Leu732X	L732X	<5	-
c.1477_1478delCA	p.Gln493ValfsX10	1609delCA	<5	-
c.273+1G->A		405+1G->A	<5	-
c.263T>A or c.263T>G	p.Leu88X	L88X	<5	-
c.933C>G	p.Phe311Leu	F311L	<5	-
c.1340delA	p.Lys447ArgfsX2	1471delA	<5	-
c.50delT	p.Phe17SerfsX8	182delT	<5	-
c.3302T->A	p.Met1101Lys	M1101K	<5	-
c.613C->T	p.Pro205Ser	P205S	<5	-
c.1037T->C	p.Leu346Pro	L346P	<5	-
c.933_935delCTT	p.Phe312del	F311del	<5	-
c.1882G->C or c.1882G->A	p.Gly628Arg	G628R	<5	-
c.3194T->C	p.Leu1065Pro	L1065P	<5	-
c.1209+1G->A		1341+1G->A	<5	-
c.[1523T->G;3752G->A]	p.[Phe508Cys;Ser1251Asn]	F508C;S1251N	<5	-
c.2989-1G->A		3121-1G->A	<5	-
c.3181G->C	p.Gly1061Arg	G1061R	<5	-
c.571T->G	p.Phe191Val	F191V	<5	-
c.2645G->A	p.Trp882X	W882X	<5	-
c.470_483del14	p.Phe157X	602del14	<5	-
c.2859_2890delACATT CTGTTCTTCAAGCA CCTATGTCAACCC	p.Leu953PhefsX11	2991del32	<5	-
c.11C>A	p.Ser4X	S4X	<5	-
c.3971T->C	p.Leu1324Pro	L1324P	<5	-

Nucleotide	Protein	Legacy name	N	%
c.137C->A	p.Ala46Asp	A46D	<5	-
c.2735C->A	p.Ser912X	S912X	<5	-
c.1687T->G	p.Tyr563Asp	Y563D	<5	-
c.1654C->T	p.Gln552X	Q552X	<5	-
c.3717G->A		3849G->A	<5	-
c.1573C->T	p.Gln525X	Q525X	<5	-
c.987delA	p.Gly330GlufsX39	1119delA	<5	-
c.2175_2176insA	p.Glu726ArgfsX4	2307insA	<5	-
c.3717+5G->A		3849+5G->A	<5	-
c.3011_3019delCTATAGCAG or c.3009_3017delAGCTATAGC	p.Ala1004_ Ala1006del	3143del9	<5	-
c.1A->G	p.Met1Val	M1V	<5	-
c.3700A->G	p.Ile1234Val	I1234V	<5	-
c.2930C->T	p.Ser977Phe	S977F	<5	-
c.859_863delAACTT	p.Asn287LysfsX19	991del5	<5	-
c.2421A->G	p.Ile807Met	I807M	<5	-
c.1801A->T	p.Ile601Phe	I601F	<5	-
c.3873+2T->C		4005+2T->C	<5	-
c.164+1G>A		296+1G->A	<5	-
c.3745G->A	p.Gly1249Arg	G1249R	<5	-
c.2739T->A	p.Tyr913X	Y913X	<5	-
c.53+1G->T		185+1G->T	<5	-
c.1013C->T	p.Thr338Ile	T338I	<5	-
c.3435G->A	p.Trp1145X	W1145X	<5	-
c.1420G->A	p.Glu474Lys	E474K	<5	-
c.3773_3774insT	p.Leu1258PhefsX7	3905insT	<5	-
c.3209G->A	p.Arg1070Gln	R1070Q	<5	-
c.2620-26A->G		2752-26A->G	<5	-
c.413_415dupTAC	p.Leu138dup	L138ins	<5	-
c.1081delT	p.Trp361GlyfsX8	1213delT	<5	-
c.3718-3T->G		3850-3T->G	<5	-
c.1703delT	p.Leu568CysfsX4	1833delT	<5	-
c.(53+1_54-1)_(164+1_165-1)del		CFTRdele2	<5	-
c.1670delC	p.Ser557PhefsX2	1802delC	<5	-
c.1766+3A->G		1898+3A->G	<5	-
c.274-2A->G		406-2A->G	<5	-
c.3230T->C	p.Leu1077Pro	L1077P	<5	-
c.717delG	p.Leu240X	849delG	<5	-
c.2002C->T	p.Arg668Cys	R668C	<5	-
c.3458T->A	p.Val1153Glu	V1153E	<5	-
c.1240C->T	p.Gln414X	Q414X	<5	-
c.1837G->A	p.Ala613Thr	A613T	<5	-
c.1418delG	p.Gly473GlufsX54	1548delG	<5	-
c.4231C->T	p.Gln1411X	Q1411X	<5	-
'Other' selected			**736**	**6.6**

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.

cysticfibrosis.org.uk

Uniting for a life unlimited