Cystic Fibrosis Trust

UK Cystic Fibrosis Registry
2021 Annual Data Report

September 2022

Version 2

Uniting for a life unlimited

Version history

Version	Date	Action
1	September 2022	Initial publication
2	December 2022	Correction to Appendix 3

UK Cystic Fibrosis Registry
2021 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

This report covers a year of continued challenge and change, with the ongoing impact of COVID-19, but also real progress for people with cystic fibrosis, as access to Kaftrio increases across the community.

As this new data reflects, during the second year of the pandemic, the way in which people with CF used health services continued to show differences from previous years. The proportion of people having annual reviews remains stable when compared to 2020 at 93%, but remains below the figures shown pre-2019. It is also interesting to note that 30% of annual reviews were recorded as taking place virtually, which indicates the persistence of some of the new ways of delivering care adopted during the first wave of COVID-19.

The scale and scope of the data collected by the UK CF Registry continued to grow, as more frequent data collection to support the reporting of the effectiveness of new medicines continued. CF teams entered over 32,000 encounters onto the system, including over 10,000 annual review datasets. I would like to thank everyone involved in making this possible. It is an amazing achievement – and we couldn't do it without the dedication of CF teams across the country.

As the CF landscape continues to change, the fact that 99% of people with CF and their families consent to their data being captured on the UK CF Registry means we can understand the increasing diversity of our community and use this to ensure that research is targeted so that nobody in the community is left behind.

It also means that throughout the year, the UK CF Registry was able to support domestic and international efforts to monitor the impact of COVID-19, receiving reports of infections and recording vaccinations. During 2021 we recorded 814 cases of infection, with a much lower rate of hospitalisation of 7.6%, compared to the 2020 figure of 24%.

The team continues to focus on ways to use the Registry to make a difference for people with cystic fibrosis. Over the last 12 months we have continued the technical development of the Registry platform to support CF STORM, a Registry-based clinical trial. The trial is focused on the safety and impact of stopping some medicines that people may not need when they are taking elexacaftor/tezacaftor/ivacaftor. We are continuing to support the forthcoming NICE (National Institute of Health and Care Excellence) appraisal of CFTR modulator therapies, through additional data collection and analysis.

In a time of profound change, this report brings so many useful insights that really can shape our work moving forwards. There are positives we can take, with an increase in the number of people with CF having families, and a much-improved picture for overall health. The predicted median survival age of people born today has increased to 53 years. But this is still far too young – and shows there is still much more work we need to do. We won't stop until everyone with CF can truly live a life unlimited: mentally well, physically well, and personally fulfilled.

I hope you enjoy reading the report and would love to hear your feedback. Please contact us on social media or by emailing **registry@cysticfibrosis.org.uk** to let us know your comments and questions.

Finally, and most importantly, I want to extend my thanks to people with cystic fibrosis for continuing to support the UK CF Registry, as well as their families and their clinical teams for coming together to make this report possible.

David Ramsden
Chief Executive of Cystic Fibrosis Trust

Executive summary

The 2021 Registry data continues to be a rich resource to help CF teams, researchers and people with cystic fibrosis understand the current health of people with CF in the UK. The report will still be impacted by the effects of the COVID-19 pandemic, with some CF teams again re-deployed to other areas of the hospital and most hospital clinics still running some or all of their outpatient appointments remotely. This again is reflected in the slightly lower percentage of people having annual reviews (93%) compared to pre-pandemic.

Nevertheless, it is still a rich resource and I will try to bring to your attention some of the highlights of this year's report.

- 10,175 people had an annual review, and they form the basis of this report
- 61.9% of the population are over 16 years of age (57.9% are ≥ 18 years)
- 5.8% of the UK CF population report being non-white or of mixed ethnicity
- The slight downward trend in numbers of people newly diagnosed with CF continues, with the amended number for 2020 being 239. We have made a further update to the 2019 figure, which is now 276
- Nutritional status is improving, and a new graph (section 1.9) shows BMI trends over the last four years, with a smaller proportion now being underweight
- The median best FEV_1 continues to rise and is now 86.9% (section 1.15). The comparison since 2011 is shown in the next graph, with a jump apparent in the ≥ 12-year mark
- Depression in those ≥16 years old remains constant at 8.1% (section 1.22)
- 103 women had babies in 2021, nearly double the number of the previous year
- The figures for *Pseudomonas aeruginosa* infections continue to change, the intermittent figure for adults increasing (23.4%), with the chronic again decreasing (20.2%)
- Growths of NTM (6.2%) and Aspergillus (10.3%) (section 1.17, p29) have both dropped significantly, which may be related to improved health or the CFTR modulator effect of less sputum production
- The proportion of people receiving at least one course of IV antibiotics has dropped again, with only 24.3% reported compared to 39.2% in 2020. This represents 1,418 less people needing IV's in 2021 (section 1.25)
- The proportion of adults requiring oxygen has dropped in a year from 8.3% to 6.1% (section 1.35)
- The overall numbers reported as having any supplemental feeds have dropped as well, from 43.2% to 34.6% in 2021 (section 1.37)
- 7,384 people were reported as being on a CFTR modulator by December 2021 (section 1.34)
- There are quite different genotype distributions across the devolved nations, shown in section 1.45 (p49).

Sections 2 and 3 are the centre level reports which continue to show interesting differences between centres in the use of home compared to hospital IV antibiotics. They also highlight the differences in 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.

The trends of not just the COVID-19 pandemic but the more widespread introduction of CFTR modulators appear to be showing through in the data. This is important information that we hope you find useful. Again, I would like to thank the people with CF for consenting to have their anonymised clinical data recorded and the clinical teams for entering it into the Registry.

Dr Siobhán B Carr
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 64.

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

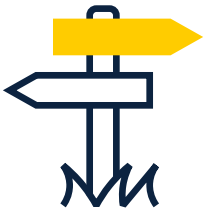
Cystic fibrosis

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-related diabetes (CFRD). Around 85% 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

	2016	2017	2018	2019	2020	2021
CF patients Registered[1]	10461	10469	10509	10655	10837	10908
Excluding diagnoses that year	10214	10255	10287	10462	10632	10720
CF patients with an annual review; n(%)[2]	9695 (95)	9887 (96)	9847 (96)	10070 (96)	9922 (93*)	10175 (93)
Age in years; median[3]	20	20	20	21	21	21
All newly diagnosed patients (NBS and other)[4]	247	214	222	193	205	188
All newly diagnosed patients (amended)[5]	(322)	(304)	(301)	(276)	(239)	(TBD)
Number of patients born identified by NBS[4]	216	192	167	150	152	134
Age at diagnosis in months; median[3]	2	2	2	2	2	2
Adults aged 16 years and over; %[3]	60.4	60.6	60.4	60.6	60.6	61.9
Males; %[3]	53.2	53.3	53	53.2	53.1	53.2
Genotyped; %[3]	98.4	99.3**	99.1	99.2	99.2	99.1
Total deaths reported during annual review year (%)[6]	148 (1.5%)	132 (1.3%)	137 (1.3%)	114 (1.1%)	97 (0.9%)	66 (0.6%)
Total deaths reported amended (%)[5]	159 (1.5%)	143 (1.4%)	143 (1.4%)	118 (1.1%)	101 (1.0%)	(TBD)
Age at death in years; median (95% CI)[6]	31 (29, 33)	31(29, 35)	32 (29, 35)	31 (29, 34)	36 (32, 38)	38 (36, 42)

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.

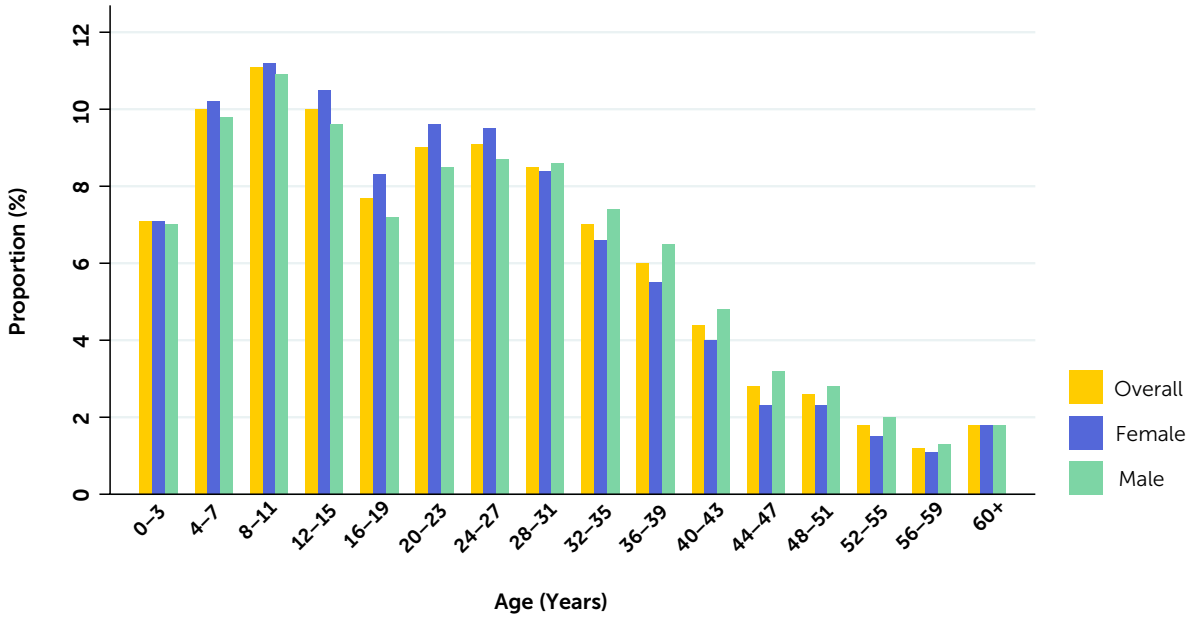
3 Calculated from patients with an annual review in the given year (see footnote 5 below).

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 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.

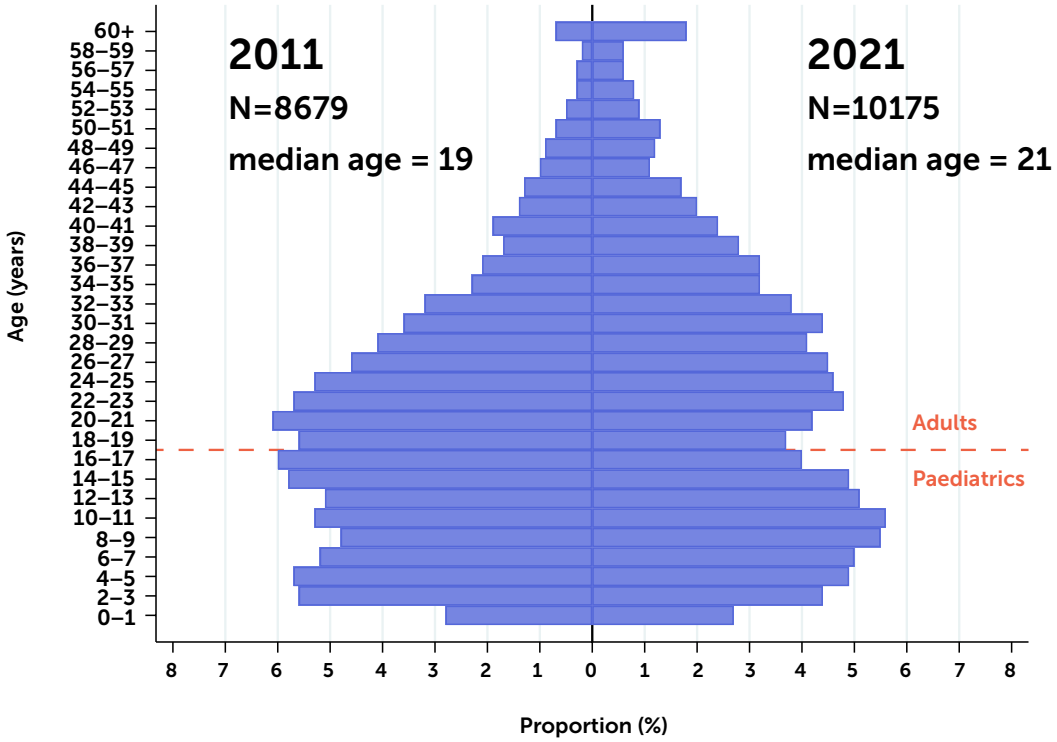
6 Calculated from all registered patients who died in the given year.

1.2 Age distribution by sex
N=10175



Age	All; n (%)	Females; n (%)	Males; n (%)
0-3	718 (7.1)	340 (7.1)	378 (7.0)
4-7	1013 (10.0)	485 (10.2)	528 (9.8)
8-11	1127 (11.1)	535 (11.2)	592 (10.9)
12-15	1020 (10.0)	498 (10.5)	522 (9.6)
16-19	785 (7.7)	397 (8.3)	388 (7.2)
20-23	919 (9.0)	458 (9.6)	461 (8.5)
24-27	926 (9.1)	454 (9.5)	472 (8.7)
28-31	865 (8.5)	399 (8.4)	466 (8.6)
32-35	713 (7.0)	315 (6.6)	398 (7.4)
36-39	614 (6.0)	262 (5.5)	352 (6.5)
40-43	451 (4.4)	190 (4.0)	261 (4.8)
44-47	280 (2.8)	108 (2.3)	172 (3.2)
48-51	261 (2.6)	111 (2.3)	150 (2.8)
52-55	179 (1.8)	72 (1.5)	107 (2.0)
56-59	123 (1.2)	52 (1.1)	71 (1.3)
60+	181 (1.8)	85 (1.8)	96 (1.8)
<16	3878 (38.1)	1858 (39.0)	2020 (37.3)
≥16	6297 (61.9)	2903 (61.0)	3394 (62.7)
<18	4283 (42.1)	2064 (43.4)	2219 (41.0)
≥18	5892 (57.9)	2697 (56.6)	3195 (59.0)
Overall	10175	4761	5414

1.3 Age distribution of the UK CF population in 2011 vs 2021



1.4 Ethnicity

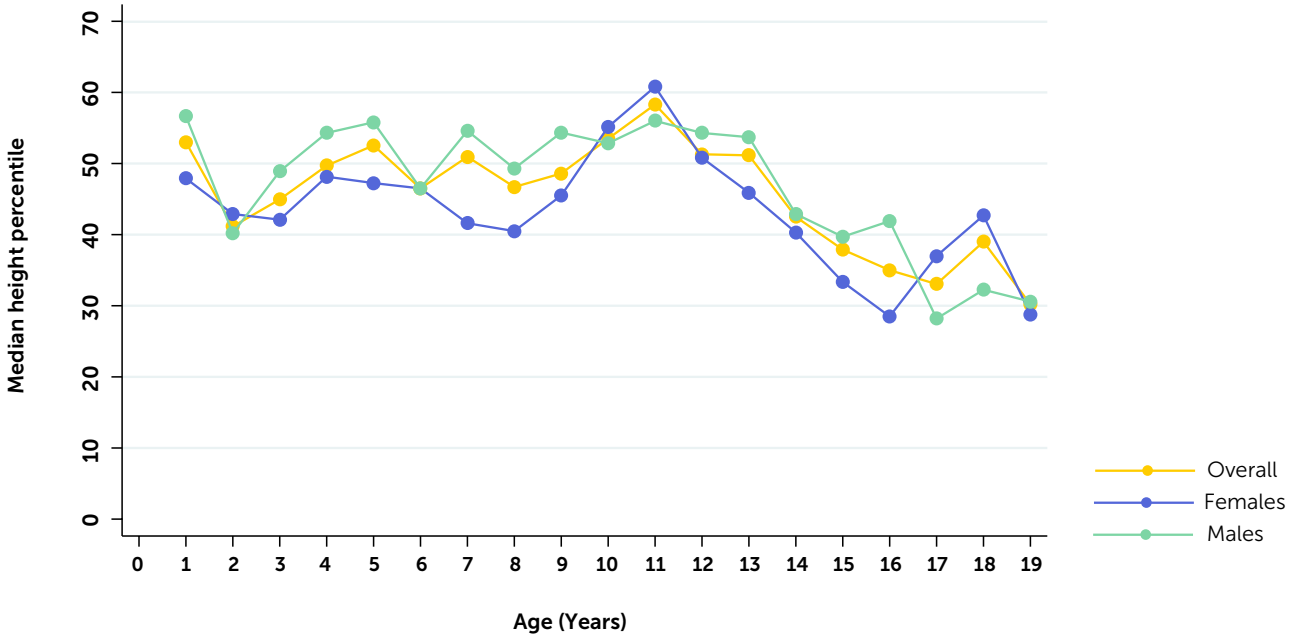
	2011	2016	2021
Total	8679	9695	10175
White			
White	8251 (95.1)	9186 (94.7)	9375 (92.1)
Asian			
Bangladeshi	32 (0.4)	33 (0.3)	46 (0.5)
Indian	25 (0.3)	36 (0.4)	48 (0.5)
Pakistani	139 (1.6)	155 (1.6)	183 (1.8)
Other (Asian)	22 (0.3)	24 (0.2)	32 (0.3)
Black			
Black African	-*	12 (0.1)	13 (0.1)
Black Caribbean	14 (0.2)	16 (0.2)	10 (0.1)
Other (Black)	<5	<5	5 (0.0)
Mixed**			
Mixed	51 (0.6)	24 (0.2)	71 (0.7)
Mixed (white-Asian)	-	6 (0.1)	17 (0.2)
Mixed (white-Black African)	-	<5	12 (0.1)
Mixed (white-Black Caribbean)	-	9 (0.1)	24 (0.2)
Other (mixed)	-	6 (0.1)	18 (0.2)
Other/Unknown			
Other	81 (0.9)	92 (0.9)	105 (1.0)
Unknown	49 (0.6)	113 (1.2)	287 (2.8)

* redacted to adhere to statistical disclosure guidelines.

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

1.5 Height percentiles of children and young people (<20 years)[1] N=4663

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.



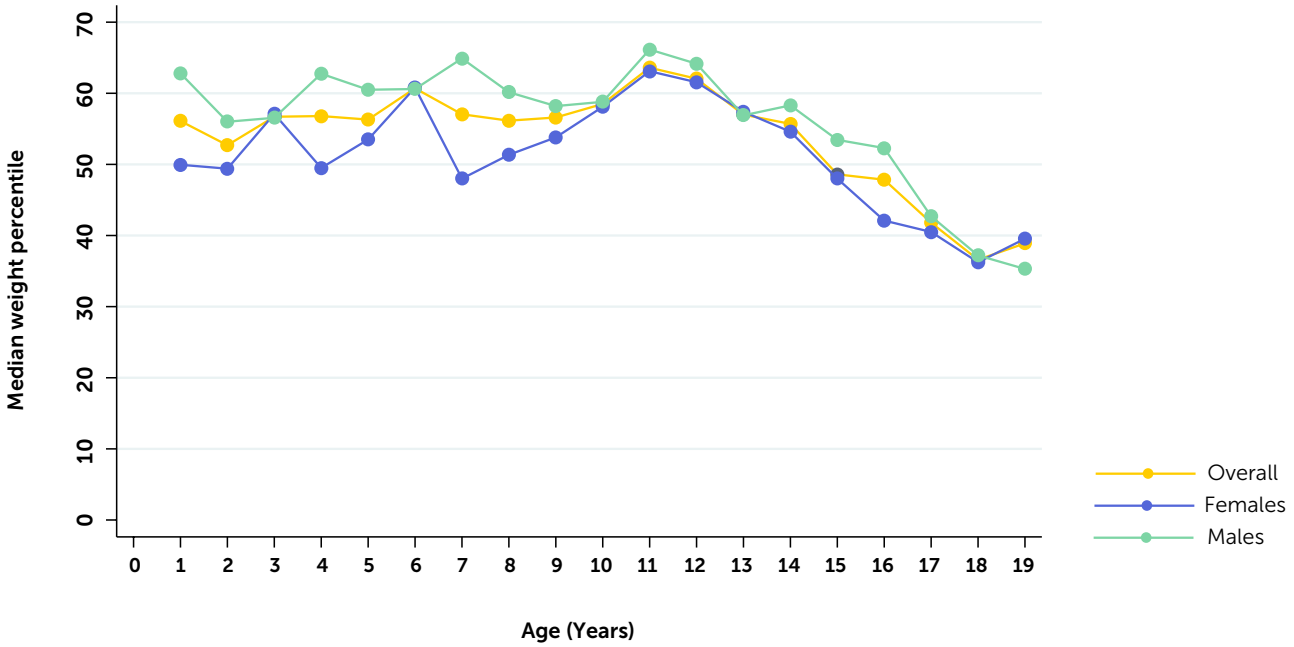
	Overall			Female			Male		
Age	n	Median	IQR	n	Median	IQR	n	Median	IQR
1	165	53.0	26.6-83.7	83	47.9	21.6-84.0	82	56.7	28.5-80.9
2	196	41.2	19.5-73.4	88	42.9	17.6-72.4	108	40.2	19.8-73.4
3	190	44.9	24.1-75.2	79	42.1	22.9-75.2	111	48.9	25.4-75.3
4	225	49.7	21.1-81.0	112	48.2	17.0-75.9	113	54.3	25.0-84.0
5	230	52.5	24.9-75.1	116	47.3	23.8-73.2	114	55.8	27.2-77.0
6	235	46.5	22.1-76.2	110	46.5	22.1-75.6	125	46.5	24.3-76.2
7	244	50.9	23.8-72.4	117	41.6	16.4-69.1	127	54.6	28.7-80.8
8	239	46.7	22.1-77.2	107	40.5	20.2-78.7	132	49.3	24.5-76.3
9	281	48.6	24.5-75.2	141	45.5	24.1-76.3	140	54.3	25.8-74.8
10	266	53.5	27.6-78.2	136	55.2	23.3-78.2	130	52.8	30.2-77.6
11	259	58.3	32.8-80.0	120	60.8	36.6-80.4	139	56.0	27.5-77.1
12	245	51.3	25.9-76.7	123	50.8	25.2-75.0	122	54.3	26.4-78.0
13	248	51.2	25.6-76.6	117	45.9	21.7-69.4	131	53.7	28.6-81.1
14	255	42.5	19.7-71.4	118	40.3	19.7-70.8	137	42.9	21.4-71.4
15	223	37.9	16.7-62.0	114	33.3	12.3-63.1	109	39.7	21.1-61.5
16	214	35.0	16.3-60.6	102	28.5	11.1-45.9	112	41.9	20.0-66.9
17	172	33.1	10.9-59.7	93	37.0	13.9-59.7	79	28.2	8.9-55.2
18	178	39.0	10.4-65.3	88	42.7	15.7-62.3	90	32.3	7.2-65.6
19	193	30.2	11.9-59.0	98	28.8	13.6-65.3	95	30.6	9.0-53.9
Overall	**4258***	**45.9**	**21.4-73.7**	**2062**	**43.8**	**19.9-73.4**	**2196**	**48.0**	**22.7-74.4**

* 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=4663

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.



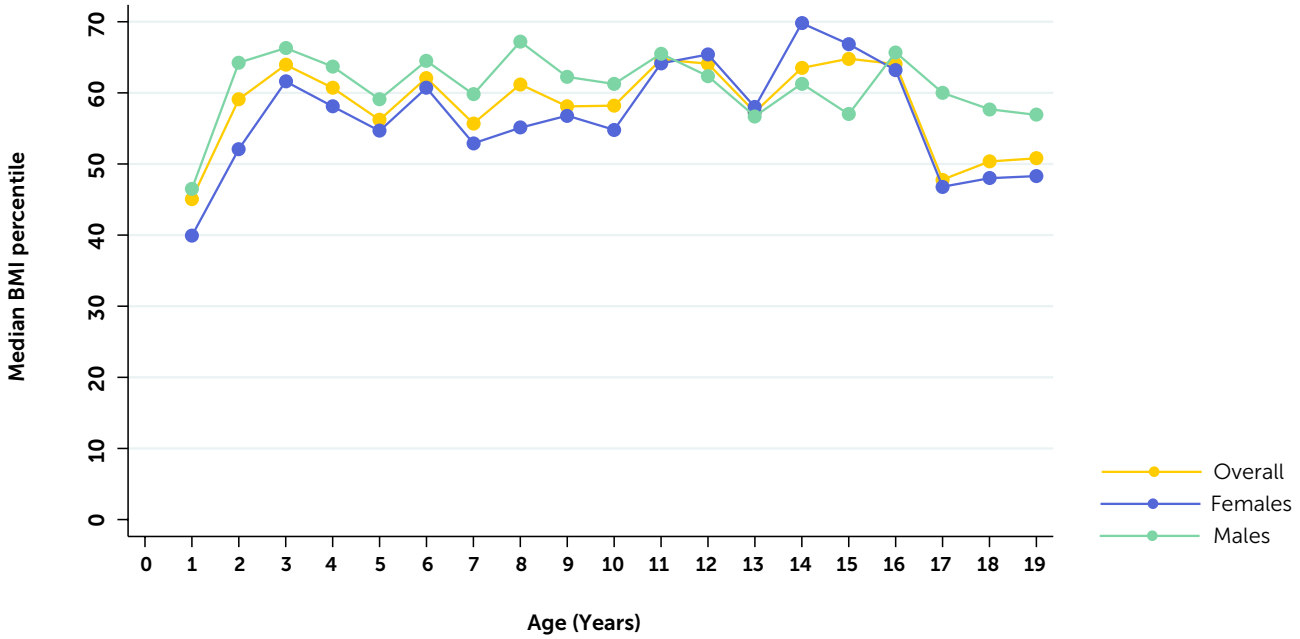
	Overall			Female			Male		
Age	n	Median	IQR	n	Median	IQR	n	Median	IQR
1	183	56.1	24.2-81.9	92	50.0	19.5-80.8	91	62.8	26.1-83.8
2	217	52.7	28.4-77.6	99	49.4	21.6-73.9	118	56.0	32.6-79.7
3	203	56.7	31.5-80.1	83	57.1	29.1-84.1	120	56.6	33.3-78.8
4	236	56.8	30.8-81.9	114	49.5	28.6-75.9	122	62.8	37.3-85.8
5	236	56.3	31.8-78.9	119	53.5	30.3-78.3	117	60.5	35.0-80.4
6	242	60.7	32.1-80.6	111	60.8	32.8-79.2	131	60.6	29.7-83.4
7	248	57.1	29.0-82.7	118	48.0	25.5-73.7	130	64.9	34.8-85.4
8	246	56.2	31.1-82.3	112	51.3	22.5-82.3	134	60.2	38.3-82.7
9	285	56.6	31.3-80.8	142	53.8	28.5-80.1	143	58.2	35.4-82.3
10	276	58.5	29.0-83.7	140	58.1	22.8-83.9	136	58.8	32.8-83.1
11	265	63.6	39.2-84.6	121	63.1	40.8-83.1	144	66.2	36.0-85.1
12	248	62.1	35.7-83.5	124	61.6	30.3-81.9	124	64.2	39.5-85.3
13	254	57.0	30.2-83.6	121	57.4	29.3-78.1	133	56.9	30.8-85.4
14	260	55.7	25.8-85.4	121	54.6	27.2-85.6	139	58.3	25.7-84.7
15	225	48.6	28.1-76.1	115	48.0	27.1-77.0	110	53.4	28.1-74.0
16	214	47.8	26.6-74.5	103	42.1	26.1-69.7	111	52.3	27.2-77.6
17	173	41.8	16.8-69.1	94	40.5	22.6-71.9	79	42.7	11.6-66.0
18	169	36.6	12.5-68.2	82	36.3	12.6-65.3	87	37.2	11.5-72.6
19	175	38.9	14.5-71.0	90	39.6	21.0-63.6	85	35.3	8.2-75.3
Overall	**4355***	**54.9**	**28.3-80.1**	**2101**	**51.9**	**26.9-78.3**	**2254**	**57.3**	**29.6-81.6**

* number with non-missing data.

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

1.7 Body Mass Index (BMI) percentiles in children and young people (<20 years)[1]
N=4663

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.



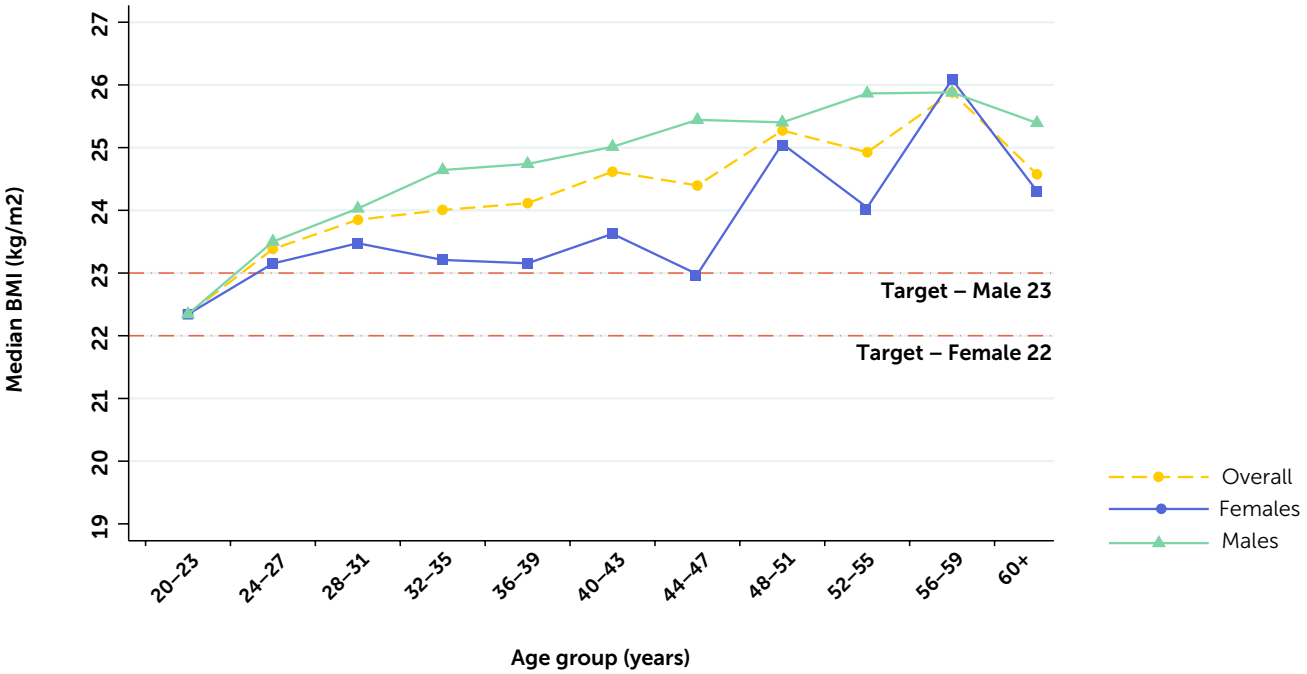
	Overall			Female			Male		
Age	n	Median	IQR	n	Median	IQR	n	Median	IQR
1	165	45.1	17.5-77.5	83	39.9	14.5-72.1	82	46.5	20.5-81.0
2	195	59.1	33.7-81.0	87	52.1	28.1-76.0	108	64.3	39.1-83.1
3	190	63.9	38.4-84.2	79	61.6	33.8-84.2	111	66.3	39.3-84.2
4	225	60.7	33.0-81.2	112	58.1	31.0-77.6	113	63.7	33.9-86.0
5	229	56.2	35.2-80.7	116	54.7	38.4-80.4	113	59.1	33.8-80.7
6	235	62.1	39.0-84.0	110	60.7	39.4-79.6	125	64.5	37.5-86.2
7	244	55.7	33.8-83.2	117	52.9	31.0-74.0	127	59.8	38.5-86.3
8	239	61.2	38.1-84.9	107	55.1	36.9-83.0	132	67.2	41.8-85.1
9	281	58.1	32.0-81.4	141	56.8	29.3-79.4	140	62.3	33.9-82.6
10	266	58.2	30.9-84.0	136	54.8	29.7-80.6	130	61.3	31.4-86.2
11	259	64.7	36.7-86.7	120	64.1	34.8-82.9	139	65.5	36.7-89.4
12	245	64.1	34.4-85.5	123	65.4	34.4-80.2	122	62.3	33.3-87.7
13	248	57.3	30.8-83.4	117	58.0	30.1-82.6	131	56.7	31.5-85.0
14	255	63.5	36.1-85.1	118	69.8	37.7-85.6	137	61.3	33.5-84.9
15	223	64.8	35.1-85.6	114	66.8	39.5-87.0	109	57.0	32.5-84.4
16	210	64.1	38.3-83.0	101	63.2	41.0-83.0	109	65.7	35.2-84.0
17	171	47.8	29.3-78.6	93	46.8	29.5-79.9	78	60.0	29.3-76.9
18	168	50.3	26.0-74.8	81	48.0	18.5-70.4	87	57.7	30.3-77.5
19	174	50.8	25.2-78.7	89	48.3	26.2-71.6	85	56.9	25.2-85.4
Overall	**4222***	**59.7**	**32.7-82.9**	**2044**	**57.2**	**31.3-80.3**	**2178**	**61.2**	**33.8-84.7**

* number with non-missing data.

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

1.8 Body Mass Index (BMI) in adults (20 years and over) N=5512

The following chart and table show the BMI of people with CF aged 20 and over in relation to the target BMI for adults; 22 for women and 23 for men[1].



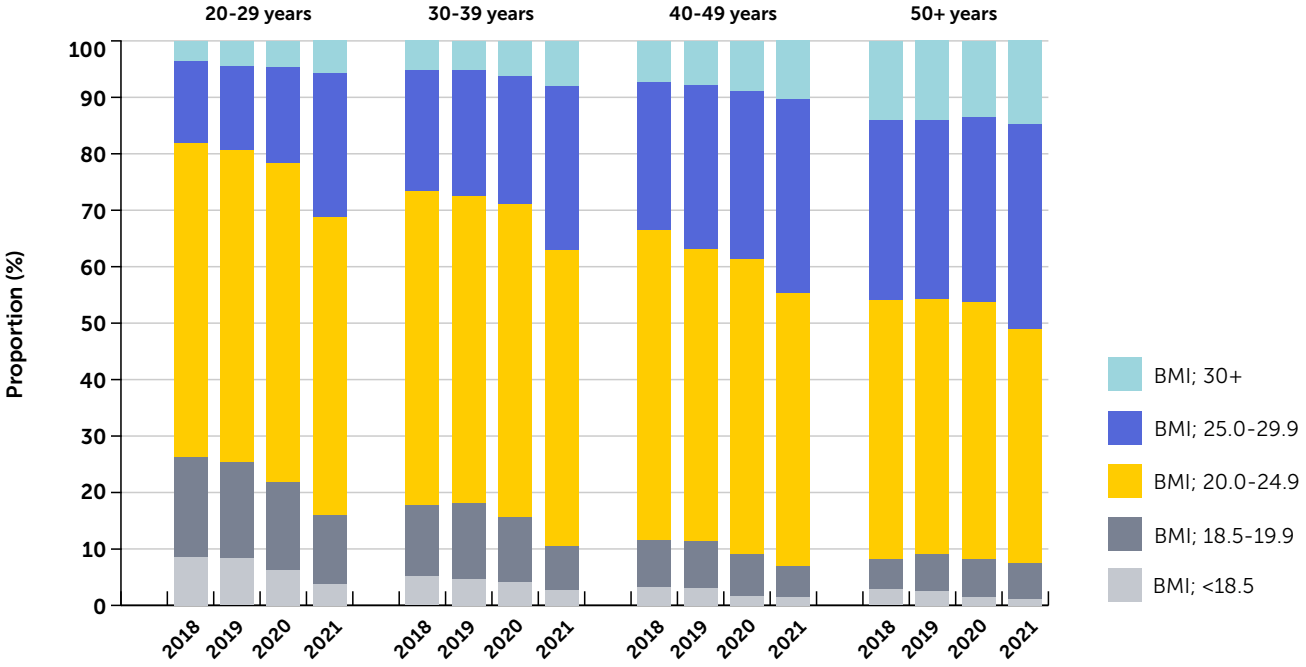
	Overall			Female			Male		
Age	n	Median	IQR	n	Median	IQR	n	Median	IQR
20-23	802	22.3	20.3-25.1	403	22.3	20.3-24.9	399	22.3	20.3-25.1
24-27	799	23.4	21.1-26.1	399	23.2	20.9-25.8	400	23.5	21.3-26.2
28-31	750	23.8	21.9-26.2	348	23.5	21.5-26.0	402	24.0	22.1-26.4
32-35	621	24.0	21.9-26.6	280	23.2	21.5-26.1	341	24.6	22.6-27.0
36-39	525	24.1	22.0-26.4	223	23.2	21.4-25.7	302	24.7	22.5-26.8
40-43	390	24.6	22.5-27.2	166	23.6	21.6-26.6	224	25.0	23.1-27.5
44-47	247	24.4	22.2-26.9	96	23.0	21.5-25.0	151	25.4	23.2-27.8
48-51	217	25.3	23.3-28.1	94	25.1	22.6-28.3	123	25.4	23.8-27.9
52-55	160	24.9	22.7-28.1	66	24.0	21.3-26.8	94	25.9	23.1-28.7
56-59	111	25.9	23.7-29.0	46	26.1	22.6-30.8	65	25.9	24.1-28.4
60+	168	24.6	22.3-28.1	79	24.3	21.2-28.0	89	25.4	22.8-28.2
Overall	**4790***	**23.9**	**21.6-26.5**	**2200**	**23.3**	**21.2-26.1**	**2590**	**24.3**	**22.1-26.9**

* number with non-missing data.

[1] Stallings et al, J Am Diet Assoc. 2008;108:832-839.

1.9 Body Mass Index (BMI) in adults for 2018 – 2021

The following graph shows the change in the proportion of people in each BMI group from 2018 to 2021.



		Proportion (%) of age group in BMI category, by year				
Age group		<18.5	18.5-19.9	20.0-24.9	25.0-29.9	30+
20-29 years	2018	8.6	17.7	55.6	14.5	3.6
	2019	8.3	17.1	55.1	15.0	4.4
	2020	6.3	15.5	56.6	16.9	4.7
	2021	3.8	12.2	52.8	25.5	5.8
30-39 years						
	2018	5.1	12.7	55.6	21.3	5.4
	2019	4.7	13.3	54.5	22.2	5.3
	2020	4.1	11.5	55.4	22.7	6.3
	2021	2.7	7.7	52.6	28.9	8.1
40-49 years						
	2018	3.3	8.2	55.0	26.1	7.3
	2019	3.1	8.3	51.6	29.1	7.9
	2020	1.7	7.4	52.1	29.8	9.0
	2021	1.5	5.4	48.5	34.3	10.4
50+ years						
	2018	2.8	5.4	45.9	31.8	14.1
	2019	2.6	6.5	45.1	31.8	14.0
	2020	1.5	6.7	45.4	32.8	13.6
	2021	1.1	6.4	41.3	36.4	14.9

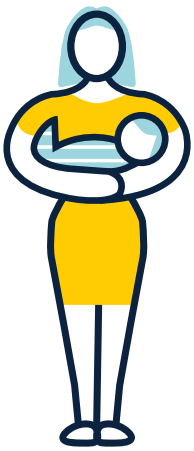
1.10 Education and employment in adults (16 years and over) N=6297

The following table shows how people with CF reported their education and employment status in 2021.

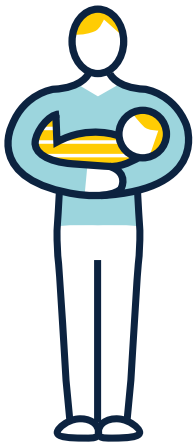
	2018	2019	2020	2021		
	Overall	Overall	Overall	Overall	Male	Female
Number of patients	5952	6104	6012	6297	3394	2903
Number who completed questionnaire; n (%)	5950 (100.0)	6103 (100.0)	5968 (99.3)	6296 (100.0)	3394 (100.0)	2902 (100.0)
Full-time employment; n (%)	1956 (32.9)	2048 (33.6)	1975 (32.9)	2097 (33.3)	1388 (40.9)	709 (24.4)
Part-time employment; n (%)	926 (15.6)	958 (15.7)	894 (14.9)	915 (14.5)	354 (10.4)	561 (19.3)
Student; n (%)	937 (15.7)	969 (15.9)	1015 (16.9)	1061 (16.8)	515 (15.2)	546 (18.8)
Homemaker; n (%)	237 (4.0)	231 (3.8)	200 (3.3)	251 (4.0)	38 (1.1)	213 (7.3)
Unemployed; n (%)	814 (13.7)	825 (13.5)	847 (14.1)	791 (12.6)	458 (13.5)	333 (11.5)
Disabled; n (%)	359 (6.0)	327 (5.4)	274 (4.6)	255 (4.0)	144 (4.2)	111 (3.8)
Retired; n (%)	133 (2.2)	145 (2.4)	139 (2.3)	162 (2.6)	88 (2.6)	74 (2.5)
Volunteer; n (%)	-*	8 (0.1)	11 (0.2)	12 (0.2)	5 (0.1)	7 (0.2)
Unknown entered; n (%)	588 (9.9)	592 (9.7)	613 (10.2)	752 (11.9)	404 (11.9)	348 (12.0)
No. in work or study; n (%)	3819 (64.2)	3975 (65.1)	3884 (65.1)	4073 (64.7)	2257 (66.5)	1816 (62.6)

1.11 Pregnancy

	2018	2019	2020	2021
Women with CF who had babies; n	65	58	56	103
Men with CF who became fathers; n	45	45	44	30



103 women with CF had babies in 2021



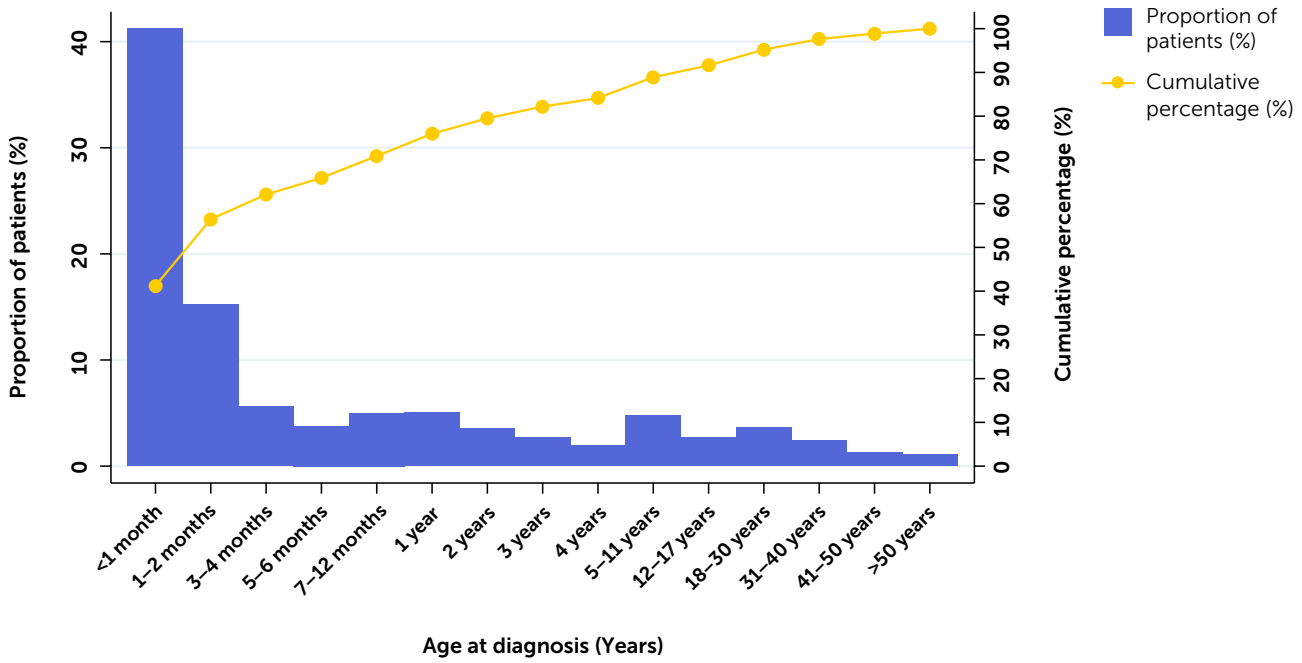
30 men with CF became fathers in 2021

*Newly added in 2019.

** Redacted to adhere to statistical disclosure guidelines.

Diagnosis of cystic fibrosis

1.12 Age at diagnosis
N=10174



The median age at diagnosis for patients aged under 16 in 2021 is **22 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 **134 (71%)** out of 188 patients born in 2021 were identified by newborn screening (including those without complete data). As there is a delay between newborn screening tests being performed and the results entering the Registry, these statistics are updated retrospectively each year to take updated data into account. The number of patients identified in 2020 is slightly lower (148) in this report than was recorded in the previous annual report.

926 (14.7%) of adults with CF in the Registry in 2021 were diagnosed at age 16 or over.

In 2021, **20** people aged 16 or over were newly diagnosed with cystic fibrosis.

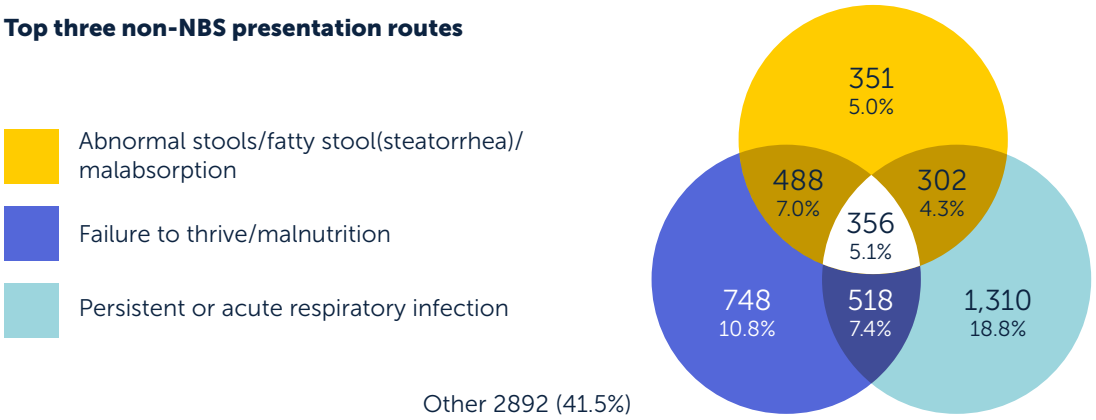
1.13 Mode of presentation

The following table shows the number of patients diagnosed through each mode of presentation. Patients may present with multiple symptoms. The Venn diagram below shows the three most common modes of presentation excluding newborn screening (NBS), and the combinations of them.

	All patients	Age <16 at diagnosis*	Age ≥16 at diagnosis*
Total patients	10175	9248	926
Number diagnosed by newborn screening	3209	3209	0
Total non-NBS	**6966**	**6039**	**926**

Presentation type	All patients	Age <16 at diagnosis*	Age ≥16 at diagnosis*
Total	6965 (100.0)	6039 (100.0)	926 (100.0)
Persistent or acute respiratory infection	2486 (35.7)	1987 (32.9)	499 (53.9)
Failure to thrive/malnutrition	2110 (30.3)	2083 (34.5)	27 (2.9)
Abnormal stools/fatty stool(steatorrhea)/malabsorption	1497 (21.5)	1445 (23.9)	52 (5.6)
Meconium ileus	1296 (18.6)	1290 (21.4)	6 (0.6)
Family history	913 (13.1)	783 (13.0)	130 (14.0)
Genotype	699 (10.0)	479 (7.9)	220 (23.8)
Unknown	328 (4.7)	271 (4.5)	57 (6.2)
Rectal prolapse	-**	236 (3.9)	<5
Nasal polyps	144 (2.1)	78 (1.3)	66 (7.1)
Bronchiectasis	103 (1.5)	10 (0.2)	93 (10.0)
Prenatal	-**	95 (1.6)	<5
Electrolyte imbalance	60 (0.9)	55 (0.9)	5 (0.5)
Fertility	-**	<5	47 (5.1)
Liver disease	-**	43 (0.7)	<5
Pancreatitis	21 (0.3)	6 (0.1)	15 (1.6)
Oedema	9 (0.1)	9 (0.1)	0 (0.0)

Top three non-NBS presentation routes



Other 2892 (41.5%)

*Age-stratified figures are presented only for those with non-missing diagnosis date.
** 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

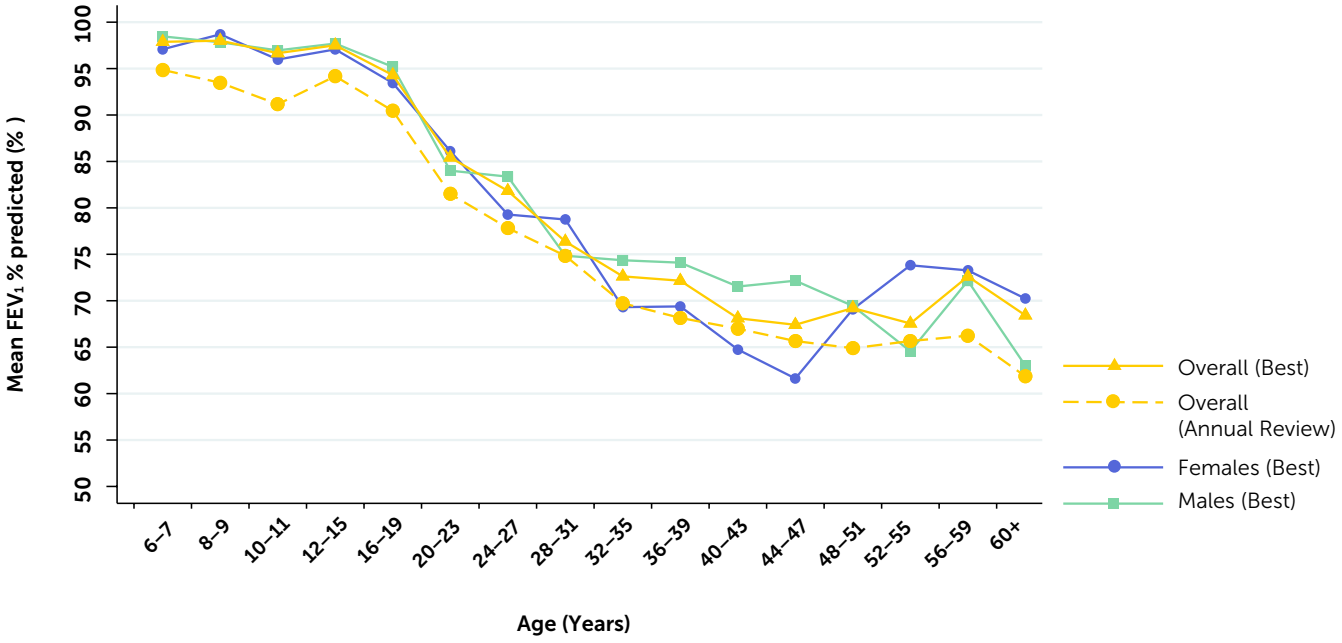
1.14 Annual review FEV_1% predicted (GLI equations) in patients age six years and older who have not had a lung transplant
N= 8659

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	388	94.9	84.7-103.3	188	95.4	87.7-103.1	200	94.0	82.9-104.0
8-9	478	93.5	84.6-101.9	229	94.0	85.0-101.7	249	93.3	84.5-102.0
10-11	490	91.2	83.0-100.0	234	91.0	82.8-98.8	256	91.3	83.0-100.6
12-15	902	94.2	83.1-102.5	438	94.1	81.8-102.1	464	94.2	84.1-102.6
16-19	676	90.5	78.7-100.6	342	90.0	77.3-99.9	334	90.7	80.8-102.2
20-23	759	81.5	65.6-95.0	386	81.8	66.2-97.7	373	81.3	65.4-93.5
24-27	741	77.8	62.0-93.4	369	75.1	57.6-92.5	372	81.6	64.2-93.9
28-31	695	74.8	55.0-89.2	321	77.0	57.9-88.6	374	72.1	51.0-89.6
32-35	553	69.7	52.1-86.6	253	66.5	52.0-83.3	300	72.5	52.6-88.7
36-39	453	68.2	47.8-86.2	201	67.0	44.7-82.0	252	69.3	50.9-88.5
40-43	358	67.0	49.0-85.1	149	64.2	50.1-85.1	209	69.0	48.6-84.7
44-47	222	65.7	48.2-83.0	87	59.2	47.5-81.5	134	68.9	49.6-83.1
48-51	188	64.9	48.0-81.0	78	65.8	51.2-77.2	110	64.3	48.0-84.8
52-55	141	65.7	45.7-84.3	56	66.7	52.0-87.4	85	61.8	42.4-80.3
56-59	100	66.2	47.8-80.9	43	65.5	52.2-76.9	57	66.9	44.8-81.6
60+	155	61.9	44.4-79.1	68	62.7	47.9-76.1	87	59.2	40.2-79.3
<16	2258	93.4	83.5-102.0	1089	93.6	83.4-101.5	1169	93.3	83.5-102.5
≥16	5040	76.4	56.2-91.3	2353	75.7	56.2-91.1	2687	76.8	56.1-91.3
<18	2609	93.4	83.3-102.0	1268	93.6	83.0-101.6	1341	93.3	83.4-102.4
≥18	4689	74.7	54.6-89.9	2174	73.6	55.0-89.5	2515	75.4	54.3-90.3
Overall	**7298***	**83.4**	**64.2-96.4**	**3442**	**83.3**	**63.7-96.5**	**3856**	**83.5**	**64.6-96.2**

*number with non-missing data.

1.15 Best* FEV_1% predicted (GLI equations) in patients aged six years and older who have not had a lung transplant
N= 8659

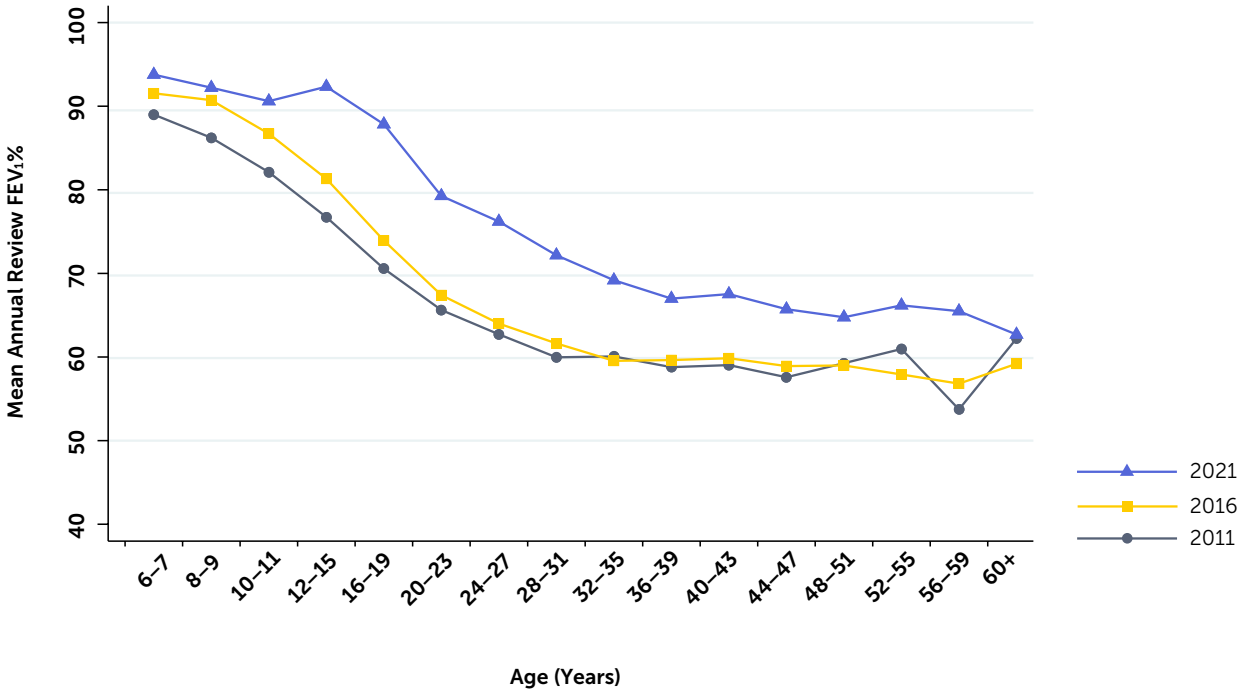


	Overall			Female			Male		
Age (yrs)	N	Median	IQR	N	Median	IQR	N	Median	IQR
6-7	439	97.9	89.8-107.3	209	97.1	90.9-107.7	230	98.5	88.1-107.1
8-9	524	98.0	90.3-106.8	251	98.7	89.7-107.4	273	97.8	90.4-106.5
10-11	538	96.7	87.8-104.9	256	96.0	87.6-104.5	282	97.0	88.1-105.4
12-15	984	97.5	87.6-106.2	480	97.1	87.3-106.3	504	97.7	88.2-106.2
16-19	743	94.3	83.8-104.4	377	93.4	83.4-103.8	366	95.2	84.1-105.6
20-23	862	85.4	69.5-97.7	429	86.1	70.3-99.8	433	84.0	68.8-95.7
24-27	856	81.8	64.1-96.5	420	79.3	61.1-95.9	436	83.4	66.6-96.8
28-31	809	76.4	57.8-92.1	377	78.8	61.4-92.5	432	74.8	53.3-91.9
32-35	642	72.6	56.5-89.0	283	69.3	55.5-86.6	359	74.4	56.6-90.4
36-39	518	72.2	51.9-88.4	227	69.4	49.5-86.2	291	74.1	52.6-89.6
40-43	408	68.1	52.1-87.1	167	64.8	52.1-87.4	241	71.5	52.1-87.0
44-47	245	67.4	49.6-85.4	95	61.6	47.5-83.5	150	72.2	51.2-86.0
48-51	218	69.2	53.2-85.3	89	69.1	53.4-84.5	129	69.4	52.7-86.7
52-55	159	67.6	47.8-86.4	65	73.8	51.3-91.9	94	64.6	44.2-81.9
56-59	111	72.6	51.2-87.6	48	73.3	57.7-85.4	63	72.1	48.3-88.7
60+	164	68.4	47.5-84.6	75	70.2	51.2-84.8	89	63.0	43.2-84.3
<16	2485	97.4	88.5-106.3	1196	97.1	88.2-106.4	1289	97.7	88.8-106.2
≥16	5735	79.5	59.2-94.3	2652	79.6	59.4-94.6	3083	79.3	58.9-94.1
<18	2866	97.4	88.3-106.3	1390	97.1	88.0-106.4	1476	97.7	88.7-106.3
≥18	5354	77.7	57.9-92.8	2458	77.8	58.4-92.8	2896	77.6	57.6-92.8
Overall	**8220****	**86.9**	**67.7-99.7**	**3848**	**87.3**	**68.0-99.9**	**4372**	**86.7**	**67.5-99.6**

* Where Best FEV_1% was missing or less than the FEV1% at annual review, annual review FEV1% was used instead.
** number with non-missing data.

1.16 Annual review FEV_1% predicted (GLI equations) over time in patients aged six years and older who have not had a lung transplant N= 8659 in 2021, N= 7977 in 2016, N= 6986 in 2011

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 2021 compares to Registry data from 2011 and 2016.



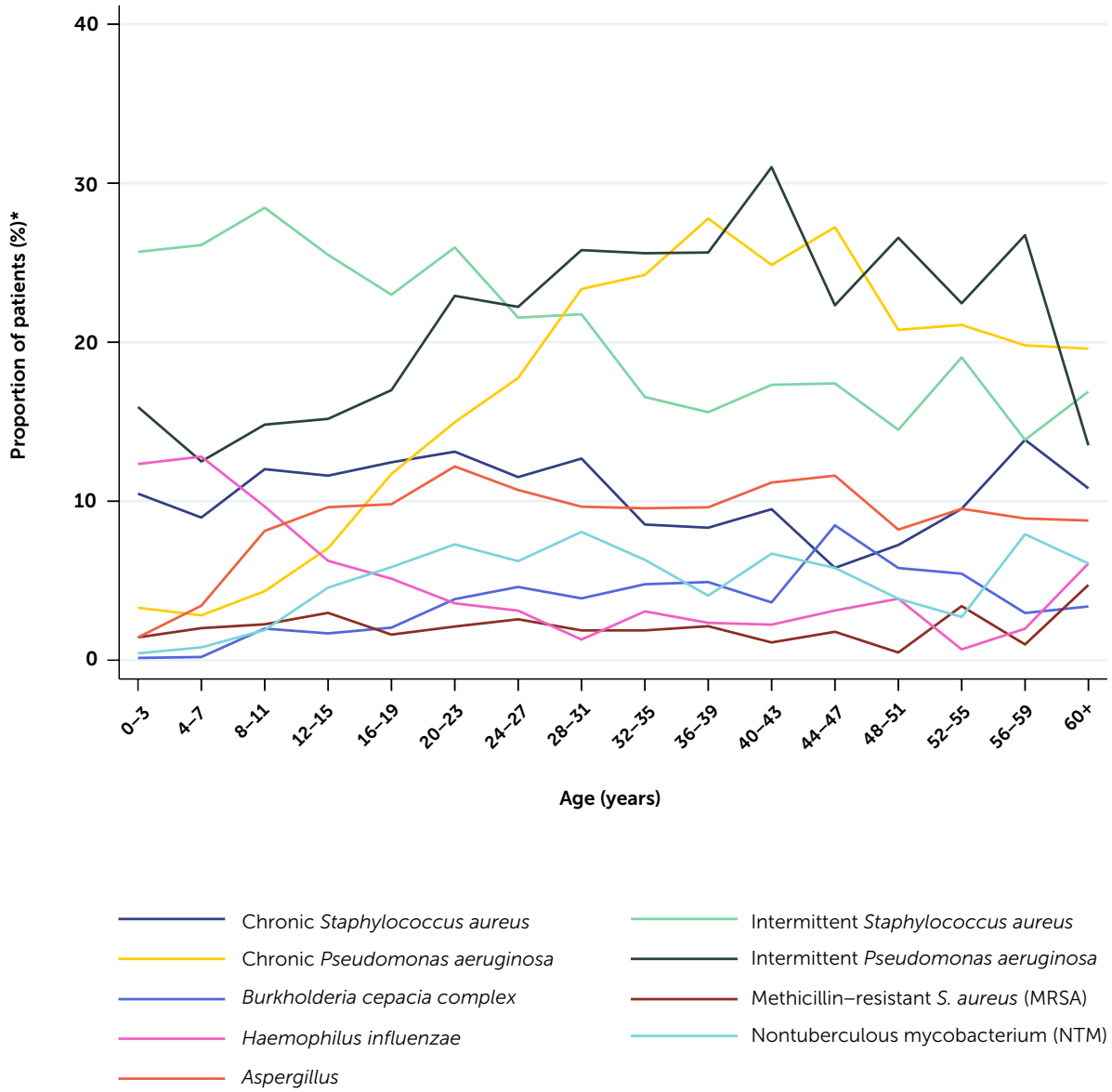
	2011		2016		2021		
Age (years)	n	FEV_1% : Mean (SD)	n	FEV_1% : Mean (SD)	n	FEV_1% : Mean (SD)	p-values (t-test)
6-7	389	89.0 (15.2)	500	91.5 (15.9)	388	93.8 (15.3)	0.035
8-9	372	86.2 (16.1)	488	90.7 (15.7)	478	92.2 (14.8)	0.131
10-11	384	82.1 (16.1)	440	86.7 (15.9)	490	90.6 (15.0)	<0.001
12-15	920	76.7 (18.7)	872	81.3 (17.2)	902	92.3 (16.0)	<0.001
16-19	949	70.6 (21.8)	918	73.9 (21.3)	676	87.8 (18.9)	<0.001
20-23	948	65.6 (23.8)	933	67.4 (23.0)	759	79.3 (21.7)	<0.001
24-27	734	62.7 (23.6)	881	64.0 (23.2)	741	76.2 (22.4)	<0.001
28-31	589	60.0 (22.6)	713	61.6 (23.8)	695	72.2 (23.5)	<0.001
32-35	388	60.1 (22.7)	564	59.5 (23.3)	553	69.2 (23.3)	<0.001
36-39	249	58.8 (22.6)	392	59.6 (23.4)	453	67.0 (23.6)	<0.001
40-43	230	59.0 (22.8)	256	59.9 (22.9)	358	67.5 (23.7)	<0.001
44-47	165	57.6 (24.9)	225	58.9 (22.5)	222	65.7 (24.2)	0.002
48-51	106	59.3 (22.9)	155	59.0 (25.9)	188	64.8 (22.7)	0.029
52-55	53	61.0 (27.3)	108	57.9 (24.2)	141	66.2 (24.6)	0.009
56-59	33	53.7 (21.8)	59	56.8 (26.5)	100	65.5 (22.1)	0.028
60+	48	62.2 (24.4)	100	59.2 (23.7)	155	62.7 (23.3)	0.245
<16	2065	81.8 (17.9)	2300	86.6 (16.9)	2258	92.2 (15.4)	-
≥16	4492	63.6 (23.4)	5304	64.3 (23.7)	5041	73.7 (23.6)	-
<18	2533	80.1 (18.7)	2739	84.8 (17.9)	2609	92.0 (15.7)	-
≥18	4024	62.6 (23.5)	4865	63.3 (23.7)	4690	72.4 (23.6)	-

** t-test comparing 2021 with 2016.

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 data set.

1.17 Lung infections in 2021
N=8913



* 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 2021 (contd.)
<16 years N=3878, ≥16 years N=6297

	Paediatric Age Range (Years)				Overall
	0-3	4-7	8-11	12-15	Paediatric (<16 years)
Number in age range	718	1013	1127	1020	3878
Number who had culture taken*	697	992	1107	1008	3804
Chronic *S. aureus* n (%)	73 (10.5)	89 (9.0)	133 (12.0)	117 (11.6)	412 (10.8)
Intermittent *S. aureus* n (%)	179 (25.7)	259 (26.1)	315 (28.5)	257 (25.5)	1010 (26.6)
Chronic *P. aeruginosa* n (%)	23 (3.3)	28 (2.8)	48 (4.3)	71 (7.0)	170 (4.5)
Intermittent *P. aeruginosa* n (%)	111 (15.9)	124 (12.5)	164 (14.8)	153 (15.2)	552 (14.5)
B. cepacia complex n (%)	<5	<5	22 (2.0)	17 (1.7)	42 (1.1)
B. cenocepacia n (%)	<5	<5	9 (0.8)	<5	13 (0.3)
B. multivorans n (%)	<5	<5	7 (0.6)	6 (0.6)	14 (0.4)
B. other cepacia n (%)	<5	<5	<5	<5	8 (0.2)
MRSA n (%)	10 (1.4)	20 (2.0)	25 (2.3)	30 (3.0)	85 (2.2)
H. influenza n (%)	86 (12.3)	127 (12.8)	107 (9.7)	63 (6.3)	383 (10.1)
NTM n (%)	<5	-**	21 (1.9)	46 (4.6)	78 (2.1)
Aspergillus fumigatus n (%)	10 (1.4)	34 (3.4)	90 (8.1)	97 (9.6)	231 (6.1)

Infections in this table reflect those grown in the 12 months prior to the 2021 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 2021 (contd.)
<16 years N=3878, ≥16 years N=6297

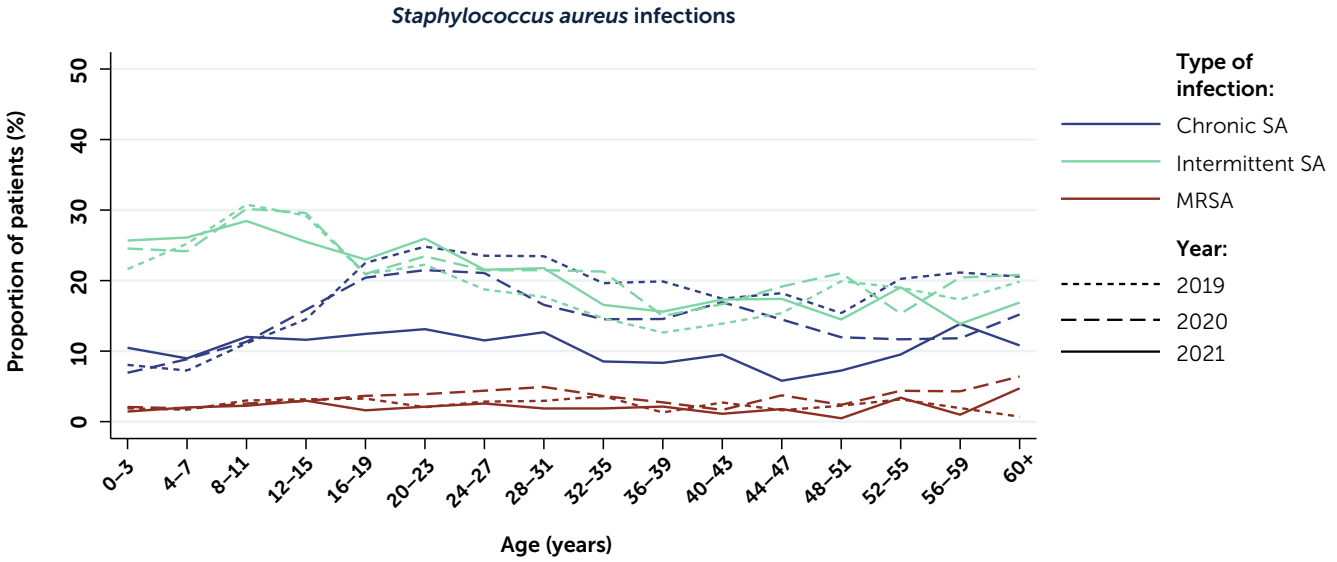
	Adult Age Range (Years)						Overall
	16-19	20-23	24-27	28-31	32-35	36-39	Adults (≥16 years)
Number in age range	785	919	926	865	713	614	6297
Number who had culture taken*	683	755	738	694	586	468	5109
Chronic *S. aureus* n (%)	85 (12.4)	99 (13.1)	85 (11.5)	88 (12.7)	50 (8.5)	39 (8.3)	552 (10.8)
Intermittent *S. aureus* n (%)	157 (23.0)	196 (26.0)	159 (21.5)	151 (21.8)	97 (16.6)	73 (15.6)	1031 (20.2)
Chronic *P. aeruginosa* n (%)	80 (11.7)	113 (15.0)	131 (17.8)	162 (23.3)	142 (24.2)	130 (27.8)	1031 (20.2)
Intermittent *P. aeruginosa* n (%)	116 (17.0)	173 (22.9)	164 (22.2)	179 (25.8)	150 (25.6)	120 (25.6)	1198 (23.4)
B. cepacia complex n (%)	14 (2.0)	29 (3.8)	34 (4.6)	27 (3.9)	28 (4.8)	23 (4.9)	215 (4.2)
B. cenocepacia n (%)	<5	-**	9 (1.2)	7 (1.0)	9 (1.5)	5 (1.1)	58 (1.1)
B. multivorans n (%)	6 (0.9)	9 (1.2)	15 (2.0)	17 (2.4)	17 (2.9)	13 (2.8)	105 (2.1)
B. other cepacia n (%)	<5	7 (0.9)	8 (1.1)	<5	<5	<5	30 (0.6)
MRSA n (%)	11 (1.6)	16 (2.1)	19 (2.6)	13 (1.9)	11 (1.9)	10 (2.1)	102 (2.0)
H. influenza n (%)	35 (5.1)	27 (3.6)	23 (3.1)	9 (1.3)	18 (3.1)	11 (2.4)	158 (3.1)
NTM n (%)	40 (5.9)	55 (7.3)	46 (6.2)	56 (8.1)	37 (6.3)	19 (4.1)	319 (6.2)
Aspergillus fumigatus n (%)	67 (9.8)	92 (12.2)	79 (10.7)	67 (9.7)	56 (9.6)	45 (9.6)	525 (10.3)

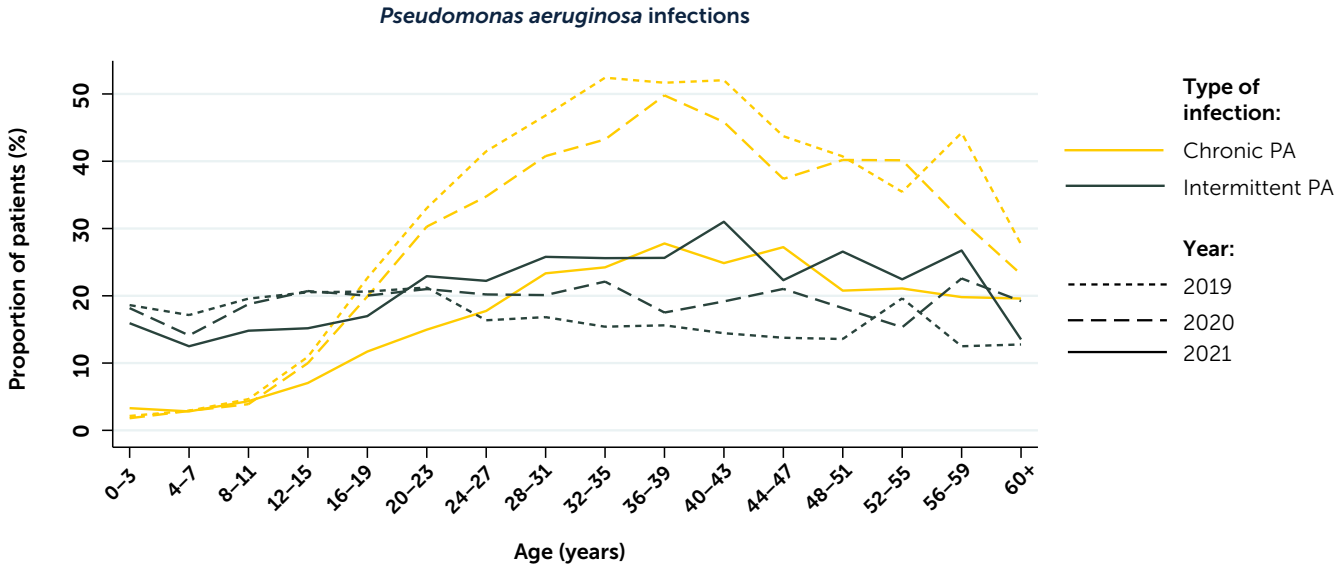
	Adult Age Range (Years)						Overall
	40-43	44-47	48-51	52-55	56-59	60+	Adults (≥16 years)
Number in age range	451	280	261	179	123	181	6297
Number who had culture taken*	358	224	207	147	101	148	5109
Chronic *S. aureus* n (%)	34 (9.5)	13 (5.8)	15 (7.2)	14 (9.5)	14 (13.9)	16 (10.8)	552 (10.8)
Intermittent *S. aureus* n (%)	62 (17.3)	39 (17.4)	30 (14.5)	28 (19.0)	14 (13.9)	25 (16.9)	1031 (20.2)
Chronic *P. aeruginosa* n (%)	89 (24.9)	61 (27.2)	43 (20.8)	31 (21.1)	20 (19.8)	29 (19.6)	1031 (20.2)
Intermittent *P. aeruginosa* n (%)	111 (31.0)	50 (22.3)	55 (26.6)	33 (22.4)	27 (26.7)	20 (13.5)	1198 (23.4)
B. cepacia complex n (%)	13 (3.6)	19 (8.5)	12 (5.8)	8 (5.4)	<5	5 (3.4)	215 (4.2)
B. cenocepacia n (%)	6 (1.7)	<5	<5	<5	<5	<5	58 (1.1)
B. multivorans n (%)	7 (2.0)	9 (4.0)	7 (3.4)	<5	<5	<5	105 (2.1)
B. other cepacia n (%)	<5	<5	<5	<5	<5	<5	30 (0.6)
MRSA n (%)	<5	<5	<5	5 (3.4)	<5	7 (4.7)	102 (2.0)
H. influenza n (%)	8 (2.2)	7 (3.1)	8 (3.9)	<5	<5	9 (6.1)	158 (3.1)
NTM n (%)	24 (6.7)	13 (5.8)	8 (3.9)	<5	-**	9 (6.1)	319 (6.2)
Aspergillus fumigatus n (%)	40 (11.2)	26 (11.6)	17 (8.2)	14 (9.5)	9 (8.9)	13 (8.8)	525 (10.3)

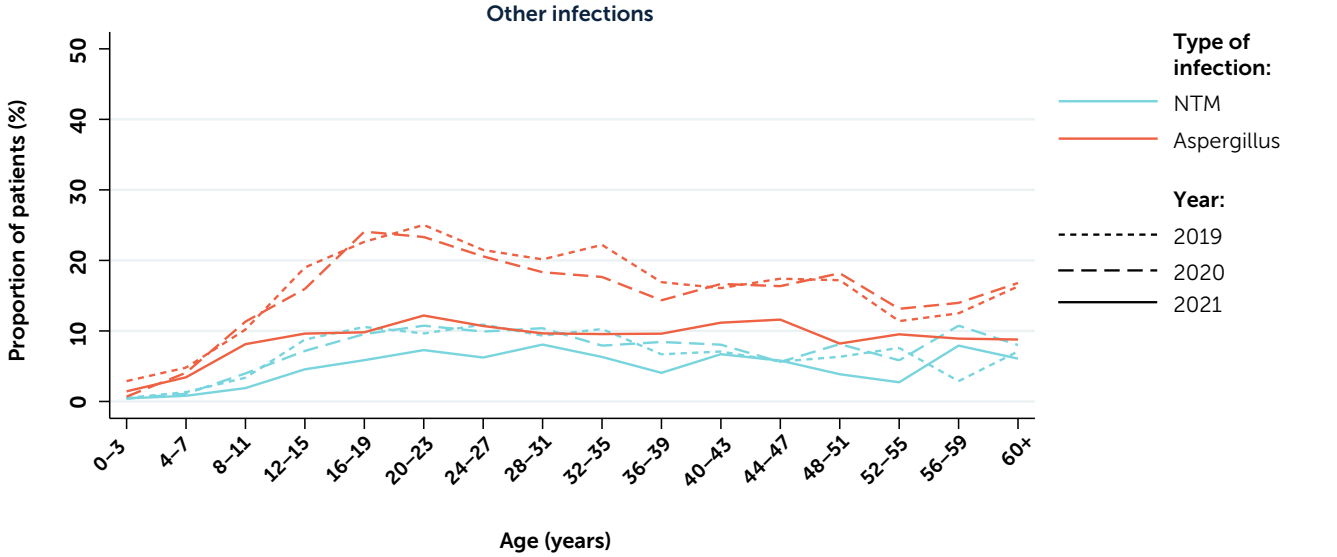
* Proportions are calculated from the number of people who were recorded as having at least one respiratory culture sample taken.

** Redacted to adhere to statistical disclosure guidelines

1.18 Lung infections 2019-2021







1.19 Respiratory culture sample type

Overall	2019	2020	2021
Number of people with an annual review (N)	10070	9922	10175
Number of people with at least 1 sample of any type taken N(%)*	9847 (97.8)	9368 (94.4)	8921 (87.7)
Sample type**			
Sputum; N(%)	6865 (69.7)	6250 (66.7)	5196 (58.2)
Cough; N(%)	6198 (62.9)	6021 (64.3)	6048 (67.8)
Bronchoalveolar lavage; N(%)	485 (4.9)	528 (5.6)	224 (2.5)
Age <16 years	**2019**	**2020**	**2021**
Number of people with an annual review (N)	3966	3910	3878
Number of people with at least 1 sample of any type taken N(%)*	3937 (99.3)	3851 (98.5)	3808 (98.1)
Sample type**			
Sputum; N(%)	1668 (42.4)	1437 (37.3)	1125 (29.5)
Cough; N(%)	3767 (95.7)	3696 (96.0)	3704 (97.3)
Bronchoalveolar lavage; N(%)	343 (8.7)	254 (6.6)	172 (4.5)
Age>=16 years	**2019**	**2020**	**2021**
Number of people with an annual review (N)	6104	6012	6297
Number of people with at least 1 sample of any type taken N(%)*	5910 (96.8)	5517 (98.1)	5113 (81.2)
Sample type**			
Sputum; N(%)	5197 (87.9)	4813 (87.2)	4071 (79.6)
Cough; N(%)	2431 (41.1)	2325 (42.1)	2344 (45.8)
Bronchoalveolar lavage; N(%)	142 (2.4)	274 (5.0)	52 (1.0)

* % is of those people with an Annual Review.

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

1.20 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.

	2019	2020	2021
Number with annual review	**(n=10070)**	**(n=9922)**	**(n=10175)**
NTM Prevalence; n (%)	674 (6.7)	620 (6.2*)	397 (3.9)
On NTM treatment in the given year; n (% of NTM prevalence in given year)	362 (53.7)	326 (52.6)	231 (58.1)
NTM Incidence[1]	279 (3.0)	226 (2.5)	154 (1.7)
M. abscessus prevalence	382 (3.8)	361 (3.6*)	216 (2.1)
M. abscessus incidence[2]	126 (1.3)	103 (1.1)	58 (0.6)

* 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 2021

COVID-19 management and outcomes for people with CF infected with COVID-19 during the calendar year of 2021 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	814	570 (70.0)	41 (5.0)	249 (30.6)	62 (7.6)
Sex; n(%)					
Female	422 (51.8)	305 (72.3)	26 (6.2)	134 (31.8)	35 (8.3)
Male	392 (48.2)	265 (67.6)	15 (3.8)	115 (29.3)	27 (6.9)
Ethnicity; n(%)					
White	765 (94.0)	534 (69.8)	36 (4.7)	232 (30.3)	51 (6.7)
Non-White	29 (3.6)	20 (69.0)	<5	9 (31.0)	9 (31.0)
Unknown	20 (2.5)	16 (80.0)	<5	8 (40.0)	<5
Age; n(%)					
Under 16	255 (31.3)	161 (63.1)	<5	56 (22.0)	10 (3.9)
>= 16	559 (68.7)	409 (73.2)	37 (6.6)	193 (34.5)	52 (9.3)
Transplants; n(%)					
No	774 (95.1)	540 (69.8)	25 (3.2)	239 (30.9)	42 (5.4)
Yes	40 (4.9)	30 (75.0)	16 (40.0)	10 (25.0)	20 (50.0)
****BestFEV; n(%)**					
<40	38 (4.7)	29 (76.3)	8 (21.1)	18 (47.4)	10 (26.3)
40-70	180 (22.1)	123 (68.3)	18 (10.0)	53 (29.4)	23 (12.8)
>70	596 (73.2)	418 (70.1)	15 (2.5)	178 (29.9)	29 (4.9)

Of the 62 patients hospitalised after a positive test for COVID-19, 17 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 2021

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

	Overall (N=10175) n (%)	<16 years (N=3878) n(%)	≥16 years (N=6297) n(%)
Respiratory related			
Nasal polyps requiring surgery	445 (4.4)	129 (3.3)	316 (5.0)
Sinus disease	714 (7.0)	53 (1.4)	661 (10.5)
Asthma	738 (7.3)	183 (4.7)	555 (8.8)
ABPA	605 (5.9)	122 (3.1)	483 (7.7)
Any haemoptysis	-*	<5	188 (3.0)
Massive haemoptysis	12 (0.1)	0	12 (0.2)
Pneumothorax requiring chest tube	12 (0.1)	0	12 (0.2)
Pancreas and hepatobiliary disease			
Raised liver enzymes	1243 (12.2)	373 (9.6)	870 (13.8)
Liver disease	1553 (15.3)	361 (9.3)	1192 (18.9)
Cirrhosis with no portal hypertension	96 (0.9)	19 (0.5)	77 (1.2)
Cirrhosis with portal hypertension	139 (1.4)	29 (0.7)	110 (1.7)
Gall bladder disease requiring surgery	147 (1.4)	29 (0.7)	118 (1.9)
Pancreatitis	73 (0.7)	9 (0.2)	64 (1.0)
Upper gastrointestinal (GI)			
Gastro-oesophageal reflux disease (GORD)	1764 (17.3)	252 (6.5)	1512 (24.0)
Peptic ulcer	0 (0.0)	0	0
GI bleed (varices as source)	16 (0.2)	7 (0.2)	9 (0.1)
GI bleed (non varices as source)	17 (0.2)	8 (0.2)	9 (0.1)
Lower gastrointestinal			
Intestinal obstruction	34 (0.3)	17 (0.4)	17 (0.3)
DIOS	493 (4.8)	102 (2.6)	391 (6.2)
Fibrosing colonopathy / colonic stricture	-*	0	<5
Rectal prolapse	12 (0.1)	8 (0.2)	<5
Renal			
Kidney stones	144 (1.4)	10 (0.3)	134 (2.1)
Renal failure	-*	<5	81 (1.3)
Musculoskeletal			
Arthritis	-*	<5	111 (1.8)
Arthropathy	230 (2.3)	7 (0.2)	223 (3.5)
Bone fracture	37 (0.4)	16 (0.4)	21 (0.3)
Osteopenia	925 (9.1)	7 (0.2)	918 (14.6)
Osteoporosis	408 (4.0)	7 (0.2)	401 (6.4)
Other			
Cancer confirmed by histology	-*	<5	28 (0.4)
Port inserted or replaced	210 (2.1)	64 (1.7)	146 (2.3)
Depression	522 (5.1)	15 (0.4)	507 (8.1)
Hearing loss	358 (3.5)	34 (0.9)	324 (5.1)
Hypertension	-*	<5	197 (3.1)

* 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.

	2020			2021		
	Overall (n=9922)	<16 years (n=3910)	≥16 years (n=6012)	Overall (n=10175)	<16 years (n=3878)	≥16 years (n=6297)
ABPA	216 (2.2)	87 (2.2)	129 (2.2)	153 (1.5)	71 (1.8)	82 (1.3)
Cirrhosis - no portal hypertension	40 (0.4)	11 (0.3)	29 (0.5)	57 (0.6)	19 (0.5)	38 (0.6)
Cirrhosis - with portal hypertension	46 (0.5)	11 (0.3)	35 (0.6)	39 (0.4)	11 (0.3)	28 (0.5)
Cancer confirmed by histology	*	<5	17 (0.3)	*	<5	15 (0.2)

1.24 CF-related diabetes
N= 7887

Cystic fibrosis-related diabetes (CFRD) 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 CFRD. CFRD is different from type 1 and type 2 diabetes, but has features of both.

	All ≥10 years (N=7887)	10-15 years (N=1590)	≥16 years (N=6297)
On CFRD treatment; n (%)	2349 (29.8)	132 (8.3)	2217 (35.2)
Of those on treatment			
Insulin[1]; n (%)	1999 (85.1)	126 (95.5)	1873 (84.5)
CFRD Screening ; n(%)			
Yes	2993 (37.9)	1041 (65.5)	1952 (31.0)
Screening Type			
Continuous glucose monitoring[2]; n (%)	1069 (35.7)	265 (25.5)	804 (41.2)
Oral glucose tolerance test[2]; n (%)	1048 (35.0)	465 (44.7)	583 (29.9)
Not screened (other)	2258 (28.6)	122 (7.7)	2136 (33.9)
Not screened (known CFRD)	2470 (31.3)	349 (21.9)	2121 (33.7)
Unknown	162 (2.1)	77 (4.8)	85 (1.3)

1 Proportion of patients on treatment
2 Proportion of patients screened
* redacted to adhere to statistical disclosure guidelines

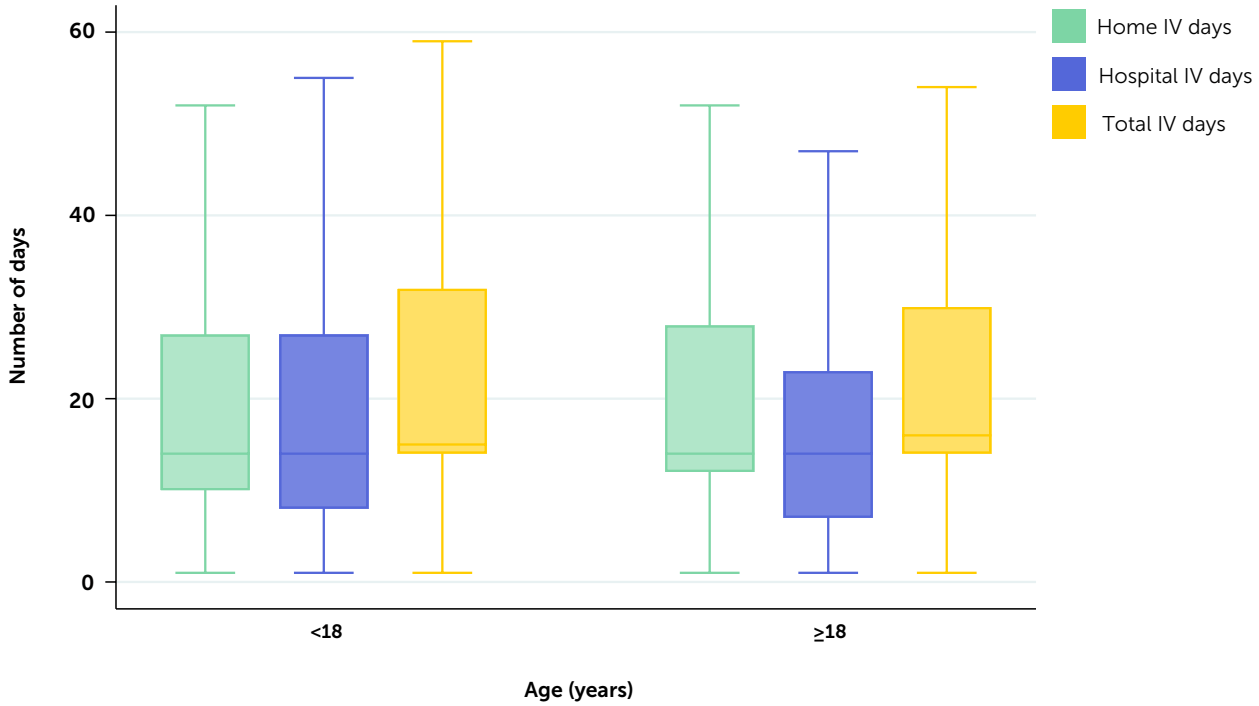
Antibiotics

1.25 Intravenous (IV) antibiotics
N=10175

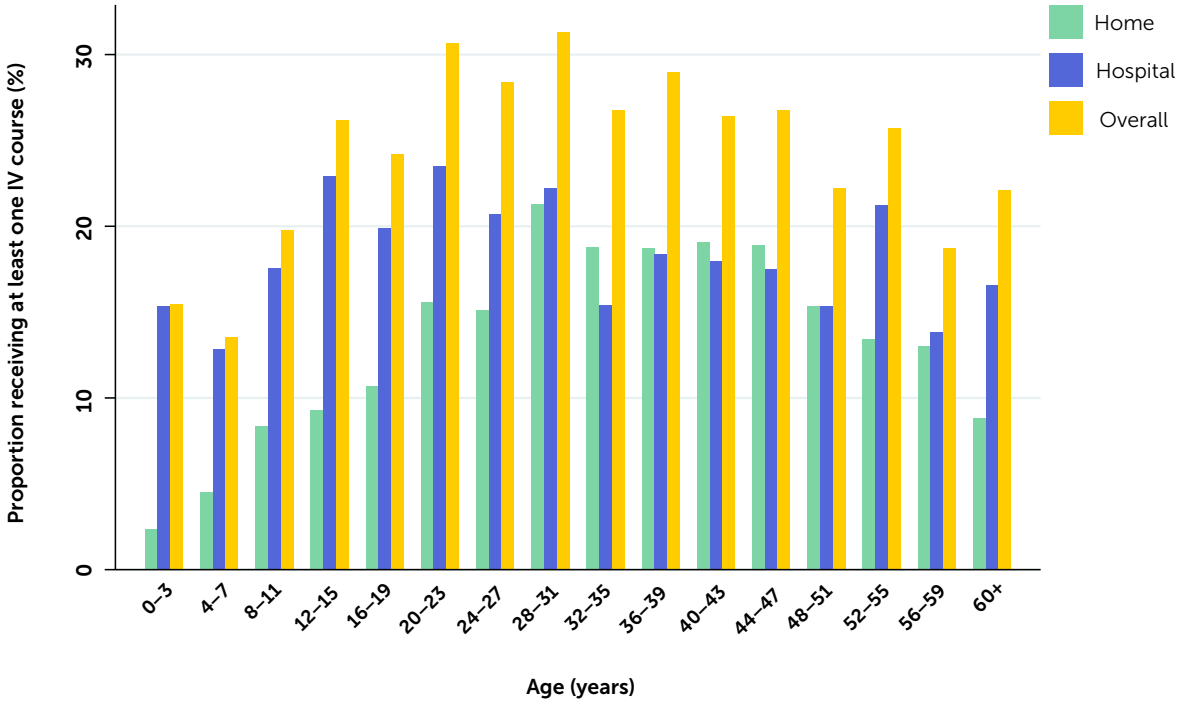
When someone with CF becomes unwell with an infection, they might be prescribed intravenous (IV) antibiotics. IV antibiotics are given to the patient through their veins. This treatment can take a number of days and may take place as a hospital inpatient, or at home.

		Home		Hospital		Total	
Age	N	Patients n (%)	Median days (IQR)	Patients n (%)	Median days (IQR)	Patients n (%)	Median days (IQR)
0-3	718	17 (2.4)	7 (5-9)	110 (15.3)	14 (7-19)	111 (15.5)	14 (8-22)
4-7	1013	46 (4.5)	13 (9-18)	130 (12.8)	14 (7-27)	137 (13.5)	14 (13-30)
8-11	1127	94 (8.3)	14 (10-27)	198 (17.6)	14 (8-28)	223 (19.8)	19 (14-38)
12-15	1020	95 (9.3)	14 (12-29)	234 (22.9)	14 (9-28)	267 (26.2)	21 (14-40)
16-19	785	84 (10.7)	14 (11-26)	156 (19.9)	13 (8-23)	190 (24.2)	14 (12-28)
20-23	919	143 (15.6)	14 (10-28)	216 (23.5)	14 (7-25)	282 (30.7)	15 (13-30)
24-27	926	140 (15.1)	14 (11-26)	192 (20.7)	14 (7-22)	263 (28.4)	16 (14-28)
28-31	865	184 (21.3)	14 (12-25)	192 (22.2)	13 (7-23)	271 (31.3)	17 (14-35)
32-35	713	134 (18.8)	14 (13-28)	110 (15.4)	13 (7-23)	191 (26.8)	16 (14-32)
36-39	614	115 (18.7)	15 (14-28)	113 (18.4)	14 (8-25)	178 (29.0)	20 (14-32)
40-43	451	86 (19.1)	14 (12-26)	81 (18.0)	12 (5-19)	119 (26.4)	17 (14-31)
44-47	280	53 (18.9)	14 (14-28)	49 (17.5)	14 (6-24)	75 (26.8)	16 (14-39)
48-51	261	40 (15.3)	16 (11-28)	40 (15.3)	10 (5-19)	58 (22.2)	18 (14-28)
52-55	179	24 (13.4)	14 (13-25)	38 (21.2)	14 (7-23)	46 (25.7)	21 (14-30)
56-59	123	16 (13.0)	14 (12-15)	17 (13.8)	10 (8-20)	23 (18.7)	14 (14-28)
60+	181	16 (8.8)	14 (10-14)	30 (16.6)	14 (8-20)	40 (22.1)	14 (11-22)
<16	3878	252 (6.5)	14 (10-27)	672 (17.3)	14 (8-28)	738 (19.0)	15 (14-32)
≥16	6297	1035 (16.4)	14 (12-28)	1234 (19.6)	14 (7-23)	1736 (27.6)	16 (14-30)
<18	4283	298 (7.0)	14 (10-27)	754 (17.6)	14 (8-27)	841 (19.6)	15 (14-32)
≥18	5892	989 (16.8)	14 (12-28)	1152 (19.6)	14 (7-23)	1633 (27.7)	16 (14-30)
Overall	**10175**	**1287 (12.6)**	**14 (11-28)**	**1906 (18.7)**	**14 (7-24)**	**2474 (24.3)**	**15 (14-31)**

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 51.



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 12.6% and in hospital was 18.7%. The proportion receiving any IVs was 24.3%.



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

	2011			2016			2021		
	Overall	<16 years	≥16 years	Overall	<16 years	≥16 years	Overall	<16 years	≥16 years
Patients with chronic *P. aeruginosa*	3138	407	2731	2831	246	2585	1201	170	1031
Tobramycin solution; n (%)	922 (29.4)	103 (25.3)	819 (30.0)	655 (23.1)	79 (32.1)	576 (22.3)	360 (30.0)	73 (42.9)	287 (27.8)
Other aminoglycoside; n (%)	63 (2.0)	14 (3.4)	49 (1.8)	74 (2.6)	12 (4.9)	62 (2.4)	39 (3.2)	<5	-*
Colistin; n (%)	1378 (43.9)	231 (56.8)	1147 (42.0)	791 (27.9)	117 (47.6)	674 (26.1)	406 (33.8)	74 (43.5)	332 (32.2)
Promixin; n (%)	858 (27.3)	112 (27.5)	746 (27.3)	888 (31.4)	104 (42.3)	784 (30.3)	329 (27.4)	70 (41.2)	259 (25.1)
Aztreonam; n (%)	-	-	-	551 (19.5)	13 (5.3)	538 (20.8)	282 (23.5)	7 (4.1)	275 (26.7)
Colistimethate (DPI); n (%)	-	-	-	513 (18.1)	29 (11.8)	484 (18.7)	216 (18.0)	14 (8.2)	202 (19.6)
Tobramycin Inhalation Powder; n (%)	-	-	-	865 (30.6)	34 (13.8)	831 (32.1)	217 (18.1)	8 (4.7)	209 (20.3)
Levofloxacin ;n(%)	-	-	-	0	0	0	27 (2.2)	0	27 (2.6)
At least one of the above; n (%)	2589 (82.5)	366 (89.9)	2223 (81.4)	2519 (89.0)	230 (93.5)	2289 (88.5)	1044 (86.9)	152 (89.4)	892 (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 (%)
2011	**Overall**	3646	2109 (57.8)	1537 (42.2)
	0-3 years	38	5 (13.2)	33(86.8)
	4-15 years	717	190 (26.5)	527 (73.5)
	≥ 16 years	2891	1914 (66.2)	977 (33.8)
2016	**Overall**	3833	1859 (48.5)	1974(51.5)
	0-3 years	32	5 (15.6)	27 (84.4)
	4-15 years	642	974(14.6)	548(85.4)
	≥ 16 years	3159	1760(55.7)	1399 (44.3)
2021	**Overall**	4160	759 (18.2)	3401 (81.8)
	0-3 years	39	<5	-*
	4-15 years	599	63 (10.5)	536 (89.5)
	≥ 16 years	3522	692 (19.6)	2830 (80.4)

* 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	718	360 (50.1)
4-7	1013	346 (34.2)
8-11	1127	291 (25.8)
12-15	1020	242 (23.7)
16-19	785	178 (22.7)
20-23	919	238 (25.9)
24-27	926	94 (10.2)
28-31	865	65 (7.5)
32-35	713	37 (5.2)
36-39	614	32 (5.2)
40-43	451	26 (5.8)
44-47	280	14 (5.0)
48-51	261	14 (5.4)
52-55	179	10 (5.6)
56-59	123	<5
60+	181	-*
<16 years	3878	1239 (31.9)
≥16 years	6297	720 (11.4)
<18 years	4283	1313 (30.7)
≥18 years	5892	646 (11.0)
Overall	**10175**	**1959 (19.3)**

* Redacted to adhere to statistical disclosure guidelines.

Bronchodilators & Corticosteroids

1.29 Inhaled bronchodilators & corticosteroids

Age	Total patients	Patients on inhaled bronchodilators; n(%)	Patients on inhaled corticosteroids; n(%)	Patients on inhaled combination corticosteroids/ bronchodilators; n(%)
0-3	718	120 (16.7)	43 (6.0)	<5
4-7	1013	341 (33.7)	151 (14.9)	-*
8-11	1127	563 (50.0)	222 (19.7)	155 (13.8)
12-15	1020	596 (58.4)	183 (17.9)	246 (24.1)
16-19	785	533 (67.9)	150 (19.1)	271 (34.5)
20-23	919	690 (75.1)	205 (22.3)	333 (36.2)
24-27	926	647 (69.9)	188 (20.3)	310 (33.5)
28-31	865	631 (72.9)	173 (20.0)	379 (43.8)
32-35	713	529 (74.2)	148 (20.8)	315 (44.2)
36-39	614	443 (72.1)	123 (20.0)	275 (44.8)
40-43	451	322 (71.4)	98 (21.7)	204 (45.2)
44-47	280	197 (70.4)	53 (18.9)	133 (47.5)
48-51	261	177 (67.8)	57 (21.8)	115 (44.1)
52-55	179	121 (67.6)	40 (22.3)	77 (43.0)
56-59	123	87 (70.7)	22 (17.9)	47 (38.2)
60+	181	130 (71.8)	41 (22.7)	76 (42.0)
<16 years	3878	1620 (41.8)	599 (15.4)	428 (11.0)
≥16 years	6297	4507 (71.6)	1298 (20.6)	2535 (40.3)
<18 years	4283	1890 (44.1)	657 (15.3)	555 (13.0)
≥18 years	5892	4237 (71.9)	1240 (21.0)	2408 (40.9)
Overall	**10175**	**6127 (60.2)**	**1897 (18.6)**	**2963 (29.1)**

* Redacted to adhere to statistical disclosure guidelines.

Muco-active therapies

1.30 Mannitol

	2016		2021	
Age	Total patients	Patients on Mannitol; n (%)	Total patients	Patients on Mannitol; n (%)
0-3	844	0	718	0
4-7	1119	0	1013	0
8-11	975	0	1127	0
12-15	906	<5	1020	<5
16-19	943	23 (2.4)	785	9 (1.1)
20-23	998	59 (5.9)	919	33 (3.6)
24-27	965	70 (7.3)	926	44 (4.8)
28-31	802	57 (7.1)	865	54 (6.2)
32-35	648	42 (6.5)	713	55 (7.7)
36-39	443	28 (6.3)	614	32 (5.2)
40-43	303	17 (5.6)	451	27 (6.0)
44-47	267	16 (6.0)	280	23 (8.2)
48-51	181	<5	261	10 (3.8)
52-55	125	5 (4.0)	179	5 (2.8)
56-59	67	<5	123	<5
60+	109	<5	181	<5
<16 years	3844	<5	3878	<5
≥16 years	5851	326 (5.6)	6297	298 (4.7)
<18 years	4292	7 (0.2)	4283	5 (0.1)
≥18 years	5403	322 (6.0)	5892	296 (5.0)
Overall	**9695**	**329 (3.4)**	**10175**	**301 (3.0)**

1.31 DNase

	2011		2016		2021	
Age	Total patients	Patients on DNase; n (%)	Total patients	Patients on DNase; n (%)	Total patients	Patients on DNase; n (%)
0-3	964	97 (10.1)	844	126 (14.9)	718	124 (17.3)
4-7	932	280 (30.0)	1119	539 (48.2)	1013	538 (53.1)
8-11	835	395 (47.3)	975	717 (73.5)	1127	897 (79.6)
12-15	1015	587 (57.8)	906	719 (79.4)	1020	863 (84.6)
16-19	1016	575 (56.6)	943	728 (77.2)	785	683 (87.0)
20-23	1003	576 (57.4)	998	696 (69.7)	919	823 (89.6)
24-27	803	437 (54.4)	965	623 (64.6)	926	745 (80.5)
28-31	642	358 (55.8)	802	508 (63.3)	865	620 (71.7)
32-35	433	211 (48.7)	648	402 (62.0)	713	492 (69.0)
36-39	293	120 (41.0)	443	261 (58.9)	614	395 (64.3)
40-43	278	114 (41.0)	303	162 (53.5)	451	274 (60.8)
44-47	192	86 (44.8)	267	144 (53.9)	280	175 (62.5)
48-51	122	53 (43.4)	181	94 (51.9)	261	139 (53.3)
52-55	61	24 (39.3)	125	73 (58.4)	179	101 (56.4)
56-59	38	11 (28.9)	67	34 (50.7)	123	70 (56.9)
60+	52	19 (36.5)	109	47 (43.1)	181	102 (56.4)
<16 years	3746	1359 (36.3)	3844	2101 (54.7)	3878	2422 (62.5)
≥16 years	4933	2584 (52.4)	5851	3772 (64.5)	6297	4619 (73.4)
<18 years	4254	1658 (39.0)	4292	2452 (57.1)	4283	2770 (64.7)
≥18 years	4425	2285 (51.6)	5403	3421 (63.3)	5892	4271 (72.5)
Overall	**8679**	**3943 (45.4)**	**9695**	**5873 (60.6)**	**10175**	**7041 (69.2)**

1.32 Hypertonic saline

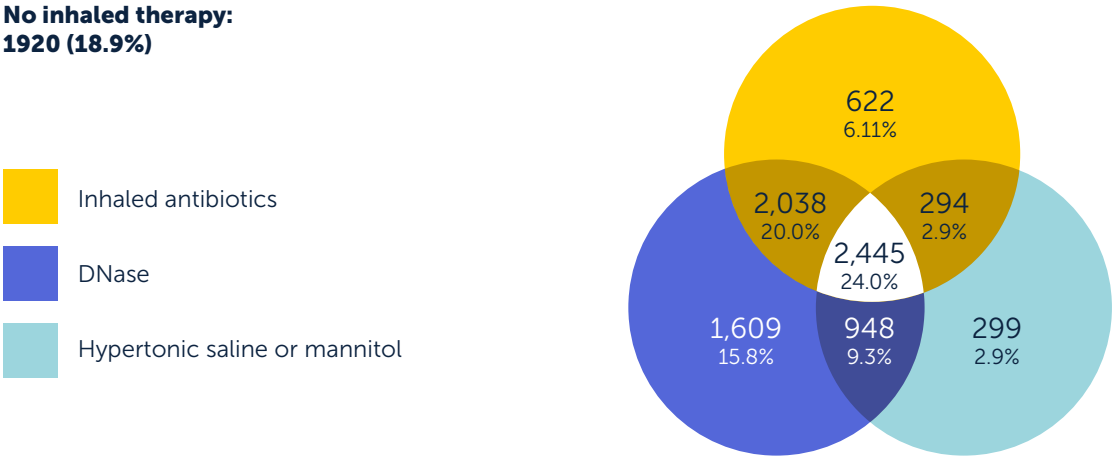
This treatment helps to thin mucus so that it is easier to cough out of the body.

	2011		2016		2021	
Age	Total patients	Patients on hypertonic saline; n (%)	Total patients	Patients on hypertonic saline; n (%)	Total patients	Patients on hypertonic saline; n (%)
0-3	964	32 (3.3)	844	71 (8.4)	718	111 (15.5)
4-7	932	77 (8.3)	1119	245 (21.9)	1013	300 (29.6)
8-11	835	163 (19.5)	975	320 (32.8)	1127	464 (41.2)
12-15	1015	196 (19.3)	906	425 (46.9)	1020	494 (48.4)
16-19	1016	184 (18.1)	943	356 (37.8)	785	404 (51.5)
20-23	1003	170 (16.9)	998	306 (30.7)	919	524 (57.0)
24-27	803	162 (20.2)	965	267 (27.7)	926	390 (42.1)
28-31	642	138 (21.5)	802	264 (32.9)	865	259 (29.9)
32-35	433	88 (20.3)	648	221 (34.1)	713	211 (29.6)
36-39	293	49 (16.7)	443	144 (32.5)	614	207 (33.7)
40-43	278	52 (18.7)	303	87 (28.7)	451	139 (30.8)
44-47	192	36 (18.8)	267	69 (25.8)	280	87 (31.1)
48-51	122	21 (17.2)	181	49 (27.1)	261	75 (28.7)
52-55	61	9 (14.8)	125	41 (32.8)	179	45 (25.1)
56-59	38	5 (13.2)	67	18 (26.9)	123	29 (23.6)
60+	52	9 (17.3)	109	27 (24.8)	181	58 (32.0)
<16 years	3746	468 (12.5)	3844	1061 (27.6)	3878	1369 (35.3)
≥16 years	4933	923 (18.7)	5851	1849 (31.6)	6297	2428 (38.6)
<18 years	4254	565 (13.3)	4292	1260 (29.4)	4283	1562 (36.5)
≥18 years	4425	826 (18.7)	5403	1650 (30.5)	5892	2235 (37.9)
Overall	**8679**	**1391 (16.0)**	**9695**	**2910 (30.0)**	**10175**	**3797 (37.3)**

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. 1920 (18.9%) people are on no inhaled therapies.

No inhaled therapy: 1920 (18.9%)



Other therapies

1.34 CFTR modulators

In 2021, the CFTR modulators were available to the following people across the UK with cystic fibrosis under a managed access agreement:

Ivacaftor

In 2021, 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 six months and over with the R117H mutation.

Lumacaftor/ivacaftor

Lumacaftor/ivacaftor is licensed for use in the UK for patients aged two 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

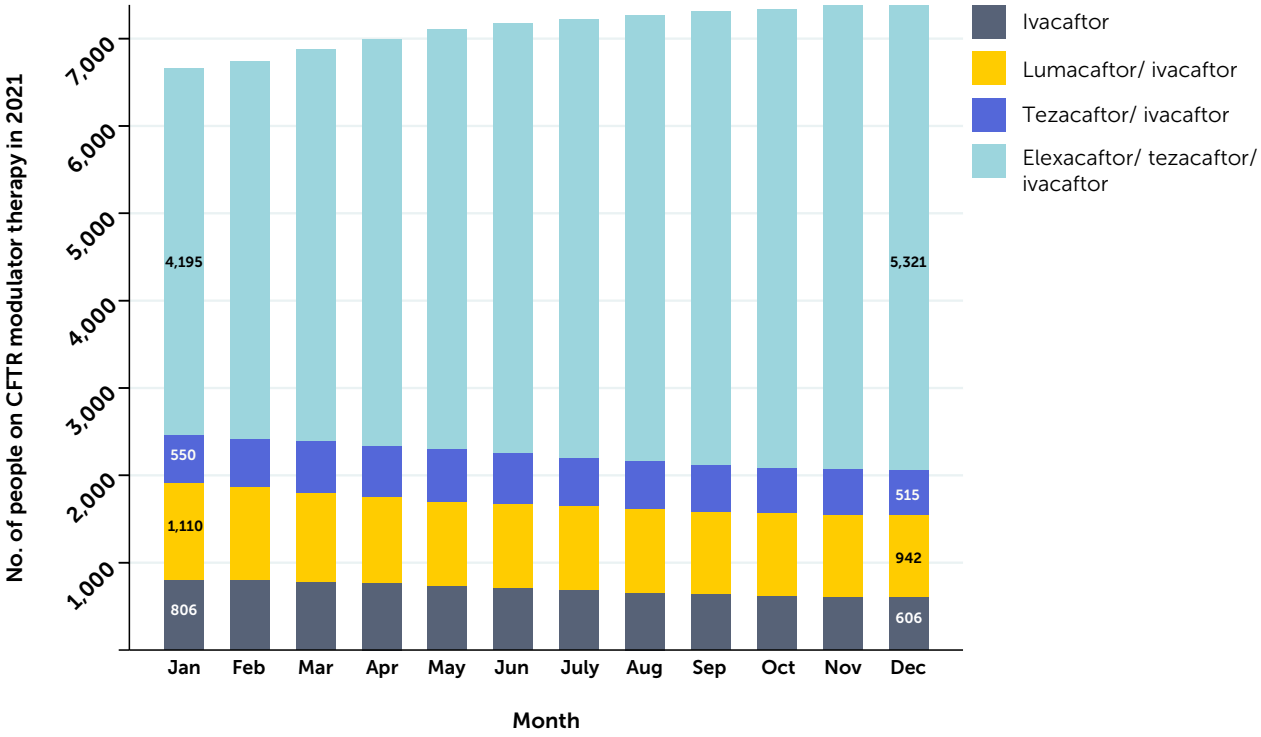
During 2021, Elexacaftor/tezacaftor/ ivacaftor was available in the UK for patients with cystic fibrosis aged 12 and over who have two copies of the F508del mutation, or a single copy of F508del and one minimal function mutation.

In January 2021 guidance was issued to clinicians supporting prescribing of a CFTR modulator to people with one copy of a F508del mutation but for whom the drug was not currently licenced.

The access arrangements prior to 2021 are described in previous annual reports.

CFTR modulator use in 2021

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, 7384 people were taking a CFTR modulator.



1.35 Oxygen and non-invasive ventilation

	Overall (n=10175)	<16 years (n=3878)	≥16 years (n=6297)	<18 years (n=4283)	≥18 years (n=5892)
Non Invasive Ventilation (NIV); n (%)	146 (1.4)	17 (0.4)	129 (2.0)	20 (0.5)	126 (2.1)
Any oxygen use; n (%)	415 (4.1)	48 (1.2)	367 (5.8)	53 (1.2)	362 (6.1)
Among those who had oxygen use:					
Continuously	-*	<5	57 (15.5)	<5	57 (15.7)
Nocturnal or with exertion	200 (48.2)	10 (20.8)	190 (51.8)	12 (22.6)	188 (51.9)
As required (PRN)	50 (12.0)	5 (10.4)	45 (12.3)	5 (9.4)	45 (12.4)
With exacerbation	106 (25.5)	31 (64.6)	75 (20.4)	34 (64.2)	72 (19.9)

1.36 Physiotherapy

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

	Overall (n=10175)	<16 years (n=3878)	≥16 years (n=6297)	<18 years (n=4283)	≥18 years (n=5892)
Active cycle of breathing techniques; n (%)	1273 (12.5)	320 (8.3)	953 (15.1)	367 (8.6)	906 (15.4)
Autogenic drainage (including assisted autogenic drainage); n (%)	1810 (17.8)	157 (4.0)	1653 (26.3)	205 (4.8)	1605 (27.2)
Postural drainage; n (%)	633 (6.2)	475 (12.2)	158 (2.5)	505 (11.8)	128 (2.2)
Any form of PEP; n (%)	6074 (59.7)	2930 (75.6)	3144 (49.9)	3241 (75.7)	2833 (48.1)
VEST; n (%)	160 (1.6)	81 (2.1)	79 (1.3)	93 (2.2)	67 (1.1)
Exercise; n (%)	6095 (59.9)	2523 (65.1)	3572 (56.7)	2797 (65.3)	3298 (56.0)
Other; n (%)	1794 (17.6)	1022 (26.4)	772 (12.3)	1083 (25.3)	711 (12.1)

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=10175)	<16 years (n=3878)	≥16 years (n=6297)	<18 years (n=4283)	≥18 years (n=5892)
Any supplemental feeding; n(%)	3523 (34.6)	1016 (26.2)	2507 (39.8)	1149 (26.8)	2374 (40.3)
Nasogastric tube; n(%)	55 (0.5)	14 (0.4)	41 (0.7)	15 (0.4)	40 (0.7)
Gastrostomy tube/button; n(%)	457 (4.5)	177 (4.6)	280 (4.4)	205 (4.8)	252 (4.3)
Jejunal; n(%)	7 (0.1)	0 (0.0)	7 (0.1)	0 (0.0)	7 (0.1)
Total Parenteral Nutrition (TPN); n(%)	<5	<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.

	2016	2017	2018	2019	2020	2021
Number evaluated	221	235	247	241	175	78
Number accepted	96	121	104	96	66	23
Number receiving aged <16 years	<5	5	<5	<5	0	0
Bilateral lung	<5	<5	0	<5	0	0
Liver	0	0	<5	<5	0	0
Other	0	<5	0	0	0	0
Number receiving aged 16+ years	51	53	63	54	15	5
Bilateral lung	46	51	58	49	12	<5
Liver	<5	0	<5	<5	<5	0
Other	<5	<5	<5	<5	<5	<5

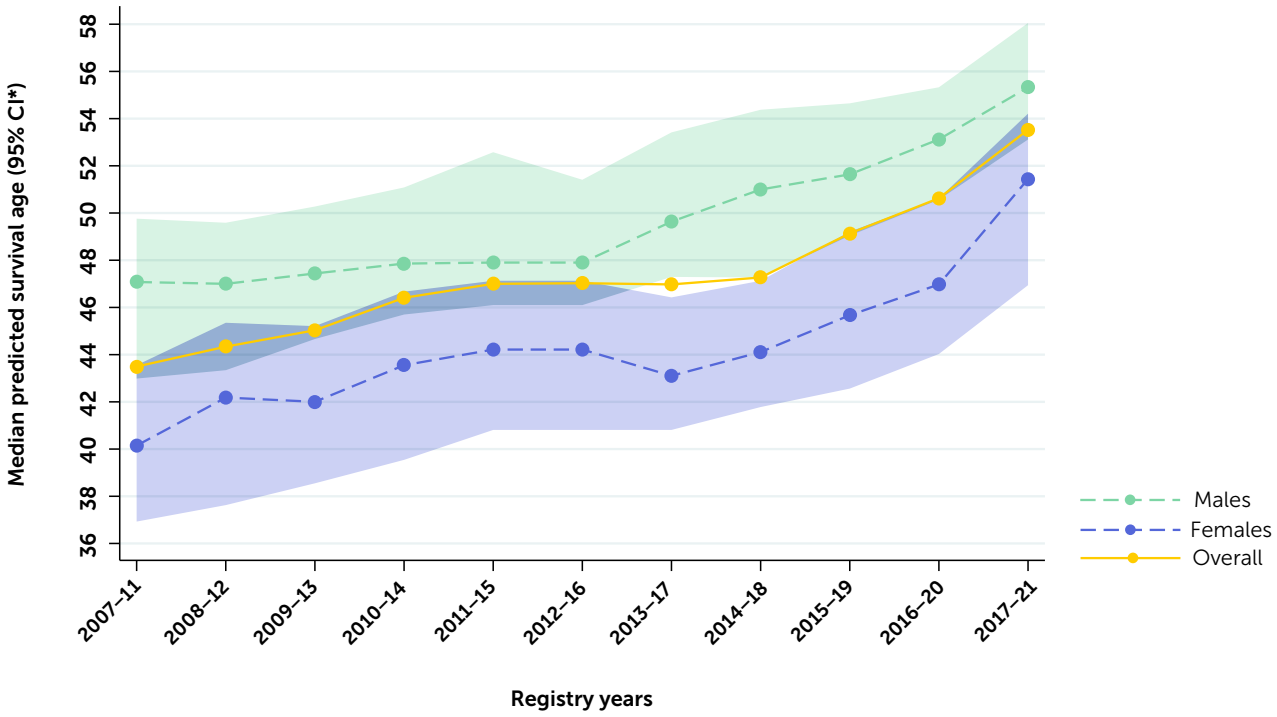
* Redacted to adhere to statistical disclosure guidelines.

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 **53.3** 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.

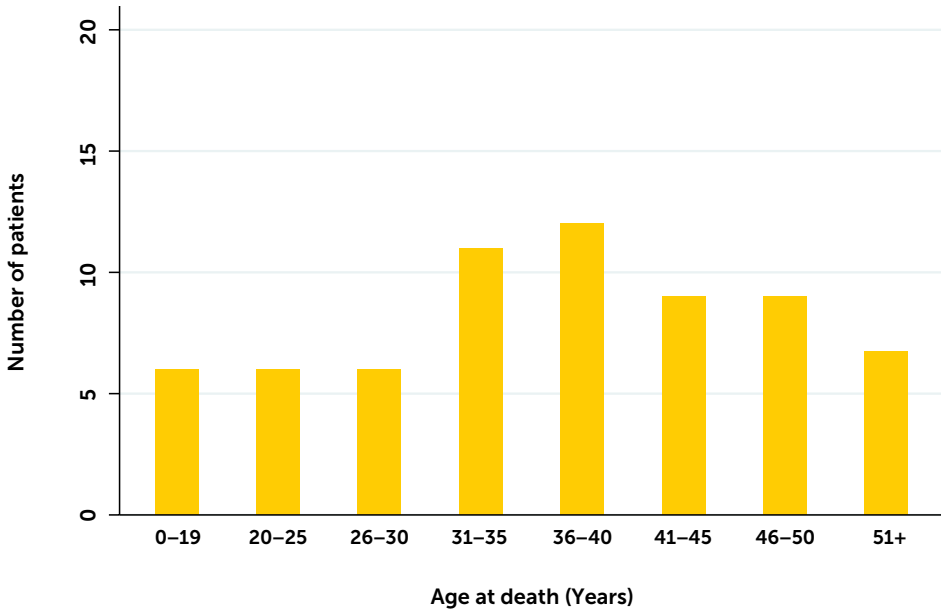


Mean predicted survival age; years (95% CI*)				
Years	Overall	Female	Male	p-value (males vs females)
2007-11	43.5(41.9-45.9)	40.1(36.9-43.6)	47.1(43.0-49.8)	<0.001
2008-12	44.3(42.4-46.5)	42.2(37.6-45.3)	47.0(43.3-49.6)	<0.001
2009-13	45.0(42.8-47.0)	42.0(38.5-45.2)	47.4(44.7-50.3)	<0.001
2010-14	46.4(43.7-47.9)	43.6(39.5-46.7)	47.9(45.7-51.1)	<0.001
2011-15	47.0(44.3-48.2)	44.2(40.8-47.1)	47.9(46.1-52.6)	0.004
2012-16	47.0(44.7-48.2)	44.2(40.8-47.1)	47.9(46.1-51.4)	0.003
2013-17	47.0(44.8-48.2)	43.1(40.8-46.4)	49.6(47.3-53.4)	<0.001
2014-18	47.3(45.7-49.6)	44.1(41.8-47.1)	51.0(47.3-54.4)	<0.001
2015-19	49.1(47.0-51.4)	45.7(42.6-49.2)	51.6(49.0-54.6)	<0.001
2016-20	50.6(48.2-53.1)	47.0(44.0-50.6)	53.1(50.6-55.3)	0.004
2017-21	53.5(51.5-55.2)	51.4(46.9-54.2)	55.3(53.1-58.1)	0.002

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

1.40 Age distribution of deaths in 2021

The table below shows the ages of the 66 people with CF who died in 2021. In 2021 the median age of the 66 people who died was 38.



Age at death	Number of patients
0-19	6
20-25	6
26-30	6
31-35	11
36-40	12
41-45	9
46-50	9
51+	7
Total	**66**

1.41 Causes of death

This table shows all the recorded causes of death between 2019 – 2021.

Cause of death	Number (%)
Respiratory/cardiorespiratory	171 (61.7)
Transplant-related	36 (13.0)
Not known	20 (7.2)
Cancer	16 (5.8)
Other	15 (5.4)
Liver disease/liver failure	9 (3.2)
COVID-19	-*
Trauma or Suicide	<5
Total	277

* 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	10801 (99.0)
Patients genotyped with both mutations recorded	10503 (96.3)
F508del mutations	
Homozygous F508del	5206 (47.7)
Heterozygous F508del	4508 (41.3)

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, 4.2% 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	47.7							47.7
R117H	5.0	0.1						5.1
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			2.0
Other	26.1	0.6	0.9	0.7	0.5	5.3		34.1
Unknown	1.9	0.1	0.1	0.1	0.0	0.5	1.0	3.7
Total	**89.1**	**1.2**	**1.4**	**1.0**	**0.6**	**5.8**	**1.0**	**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	9714	89.1
c.350G->A	p.Arg117His	R117H	680	6.2
c.1652G->A	p.Gly551Asp	G551D	625	5.7
c.1624G->T	p.Gly542X	G542X	397	3.6
c.489+1G->T		621+1G->T	280	2.6
c.3909C->G	p.Asn1303Lys	N1303K	172	1.6
c.1585-1G->A		1717-1G->A	167	1.5
c.1766+1G->A		1898+1G->A	150	1.4
c.3454G->C	p.Asp1152His	D1152H	144	1.3
c.200C->T	p.Pro67Leu	P67L	144	1.3
c.3140-26A->G		3272-26A->G	118	1.1
c.3528delC	p.Lys1177SerfsX15	3659delC	115	1.1
c.1679G->C	p.Arg560Thr	R560T	102	0.9
c.1519_1521delATC	p.Ile507del	I507del	93	0.9
c.1477C->T	p.Gln493X	Q493X	92	0.8
c.3717+12191C->T		3849+10kbC->T	85	0.8
c.1657C->T	p.Arg553X	R553X	84	0.8
c.254G->A	p.Gly85Glu	G85E	82	0.8
c.178G->T	p.Glu60X	E60X	77	0.7
c.2657+5G->A		2789+5G->A	74	0.7

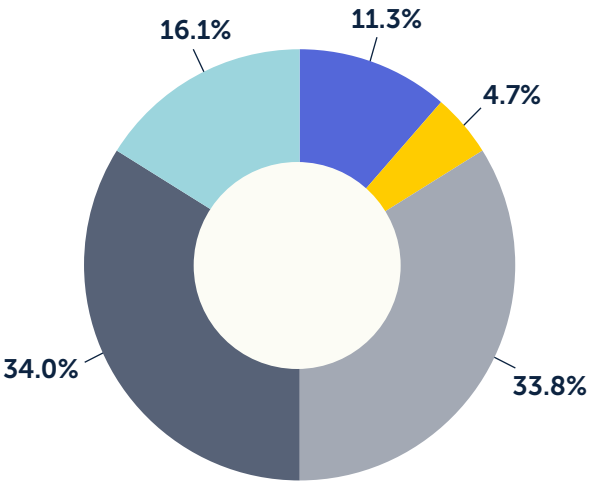
1.44 Mutation prevalence by devolved nation

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.

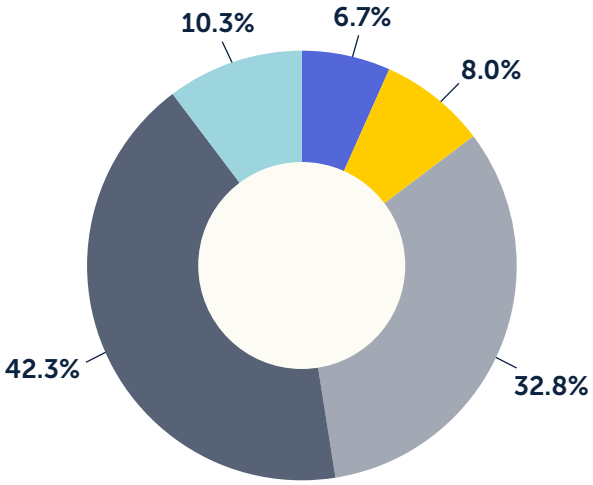
Legacy name	England N=9044		Scotland N=925		Wales N=454		Northern Ireland N=485	
	n	%	n	%	n	%	n	%
F508del	8072	89.3%	830	89.7%	405	89.2%	407	83.9%
R117H	510	5.6%	78	8.4%	17	3.7%	75	15.5%
G551D	471	5.2%	92	9.9%	15	3.3%	47	9.7%
G542X	284	3.1%	63	6.8%	22	4.8%	28	5.8%
621+1G->T	204	2.3%	10	1.1%	49	10.8%	17	3.5%
N1303K	144	1.6%	12	1.3%	7	1.5%	9	1.9%
1717-1G->A	149	1.6%	15	1.6%	<5	-	<5	-
1898+1G->A	119	1.3%	<5	-	27	5.9%	0	0.0%
P67L	75	0.8%	49	5.3%	<5	-	18	3.7%
D1152H	112	1.2%	18	1.9%	<5	-	11	2.3%

1.45 Genotype prevalence by devolved nation

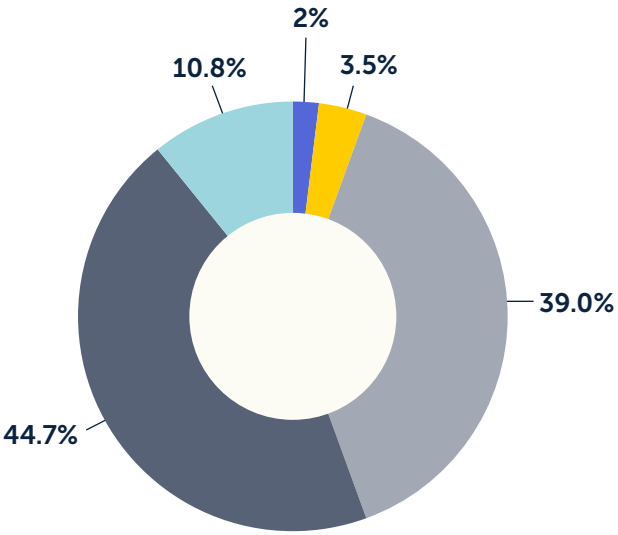
These charts show the distribution of mutation combinations across the devolved nations. The number of patients for each devolved nation is based on the location of the CF centre at which the patient receives care and does not account for patients who travel between devolved nations for care.



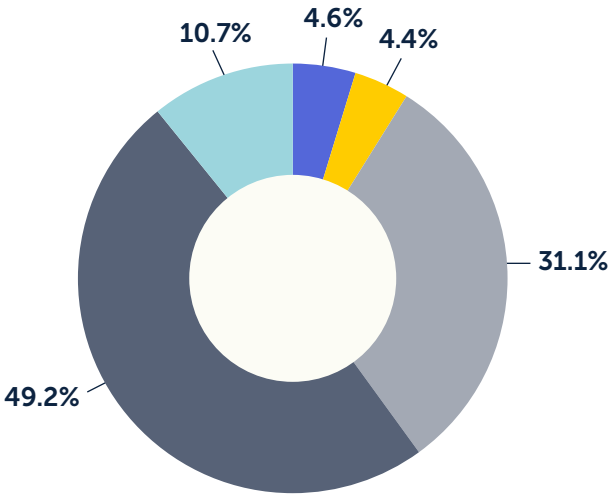
Northern Ireland
N=485



Scotland
N=925



Wales
N=454



England
N=9044

Homozygous F508del

F508del + other

F508del + gating (incl. G551D)

F508del + R117H

Non-F508del combinations

Section 2 and 3: Centre-level analysis

Cystic fibrosis care in the UK is led by 56 regional centres, 4 stand-alone clinics and 76 networked clinics. The breakdown between centres and clinics delivering paediatric and adult care is shown below:

	Paediatric	Adult	Total
Centres	30	26	56
Stand-alone clinics	2	2	4
Networked clinics	69	7	76

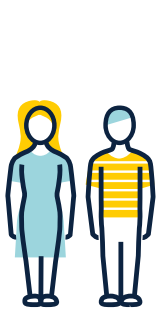
Section 2 shows analysis of data for individual CF centres. This allows people with CF, their families, and healthcare providers, to review a centre's use of some medications and outcome data alongside national averages. This transparency is intended to help improve standards of care overall.

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.

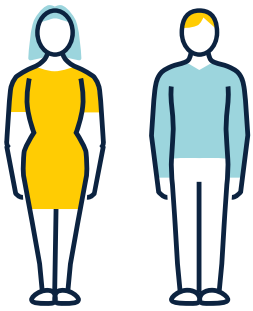
If a person with CF or a member of their family has questions about the results for their CF centre or clinic, they should discuss this with their CF team.

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

Key



Paediatric centre

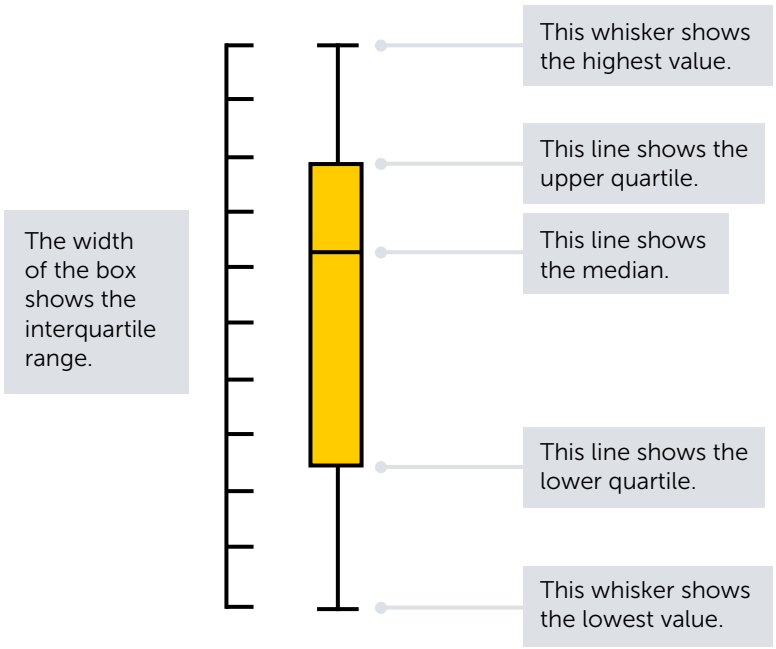


Adult centre

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 of people with CF are treated at a site, even a single outcome that is unusual affects the overall result for that site much more.

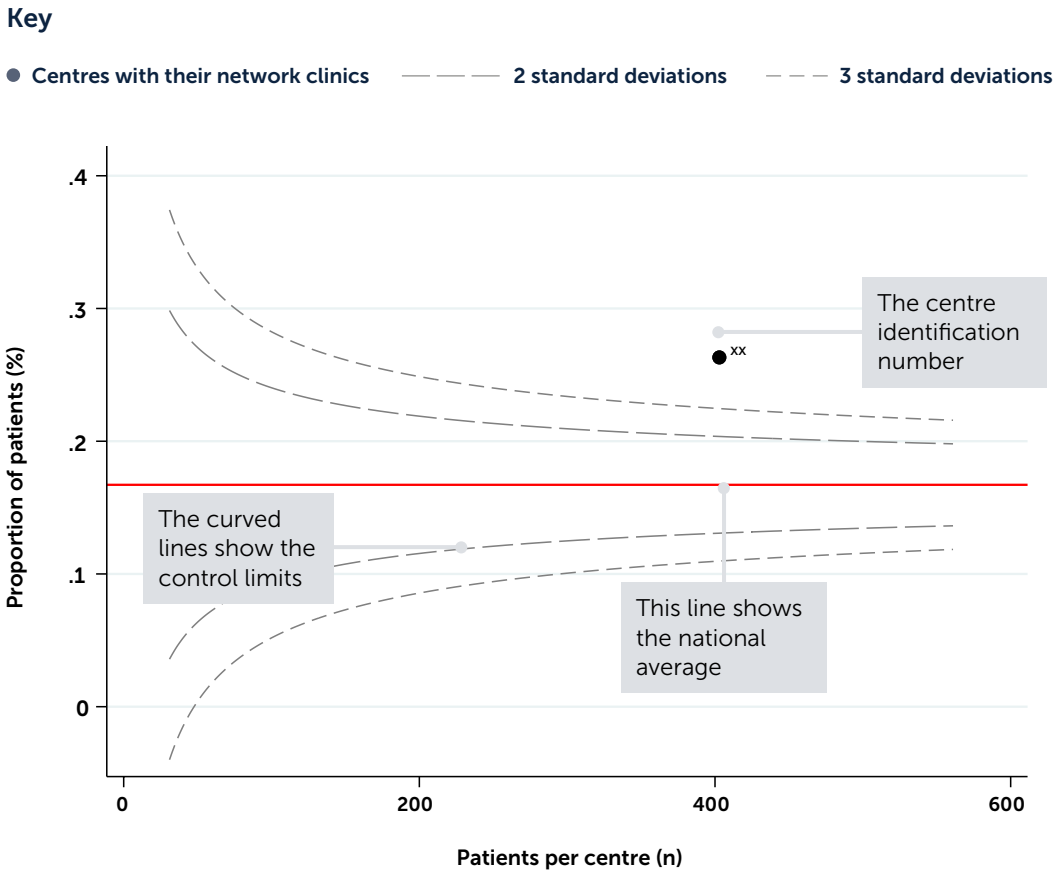
There will always be some natural variation between centres because of differences between the populations receiving care. Using only the national average as a standard can make it difficult to tell whether a survival rate that sits above the national average is higher than we would expect it to be,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

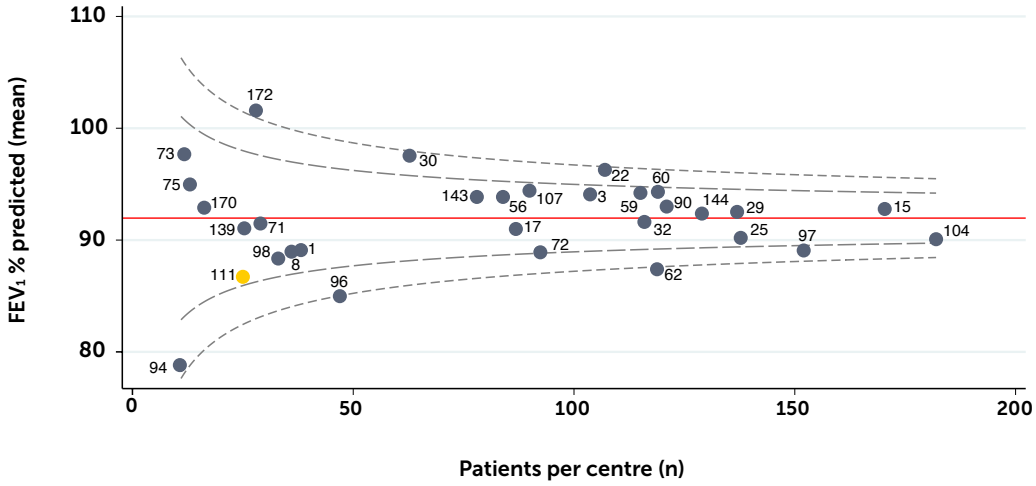


Section 2 Paediatric centre analysis

N=4,187

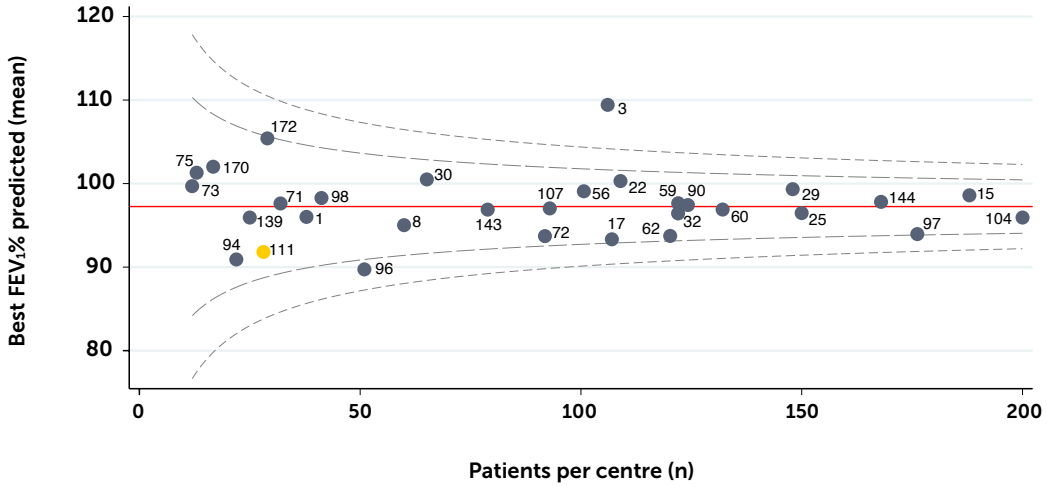
In the UK, paediatric CF care is led by 30 specialist CF centres and two stand-alone clinics (●). Some paediatric centres oversee care delivered by 69 smaller, networked clinics. Data from smaller networked clinics is included in the paediatric centre's data.

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



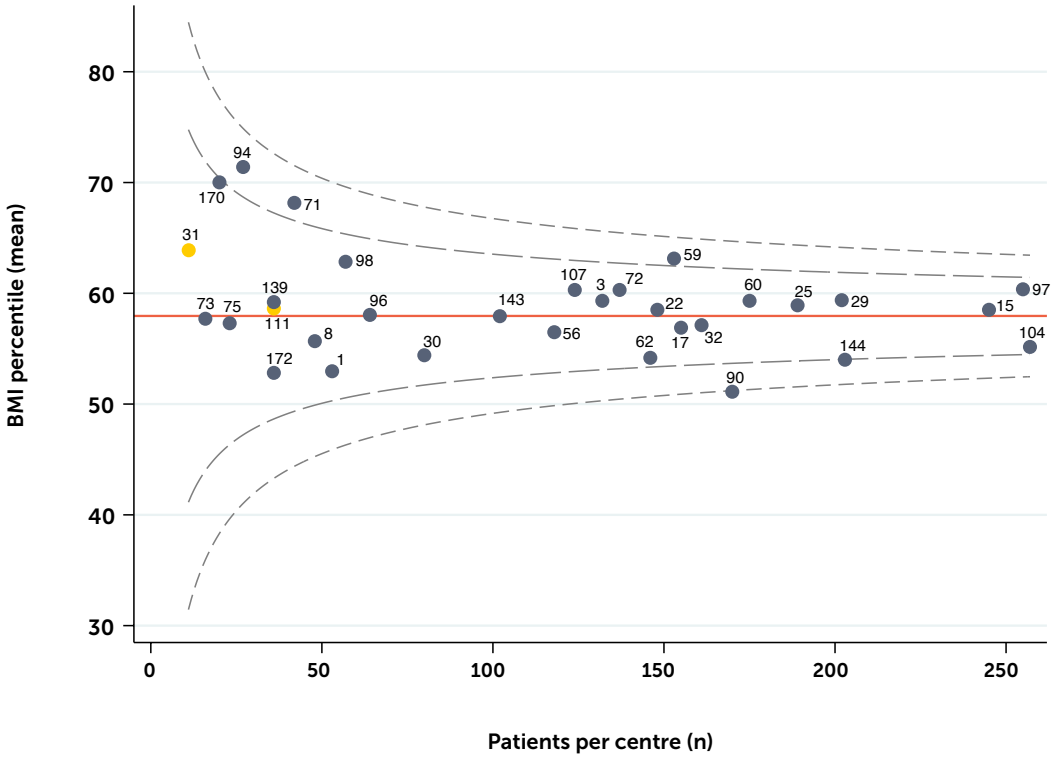
The mean FEV_1% predicted for patients attending paediatric centres/clinics is 92.0% predicted.

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



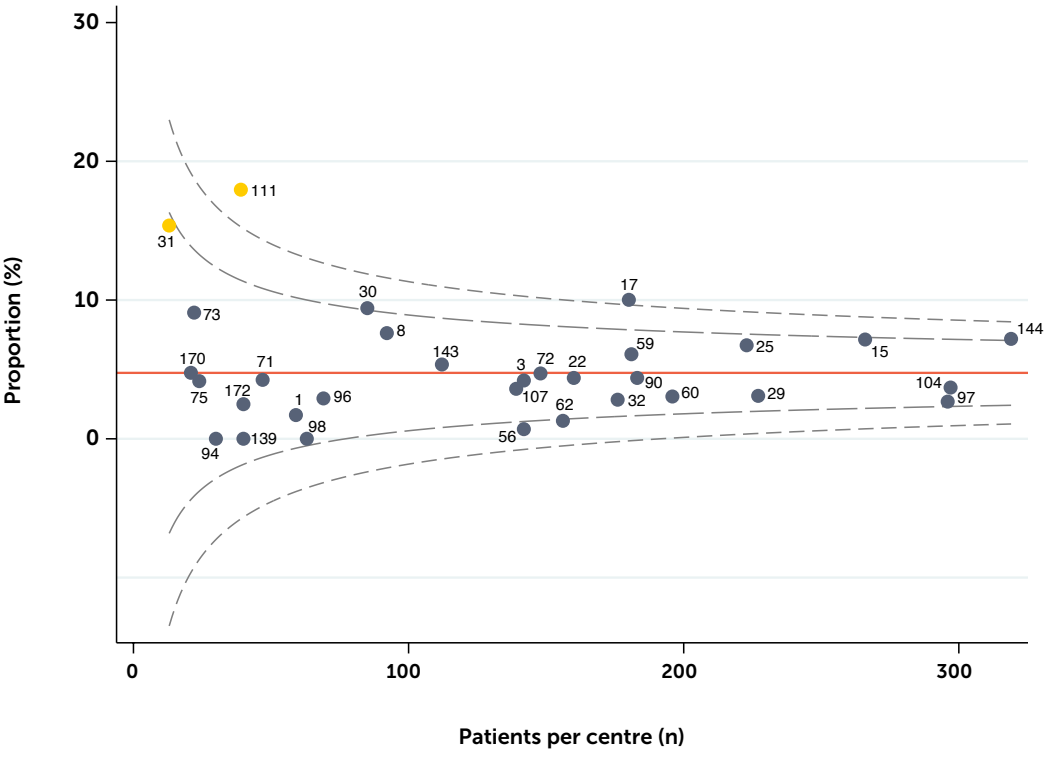
The mean Best FEV_1% predicted for patients attending paediatric centres/clinics is 97.3% predicted. Where Best FEV_1% predicted was missing, the FEV_1% predicted at annual review was used.

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



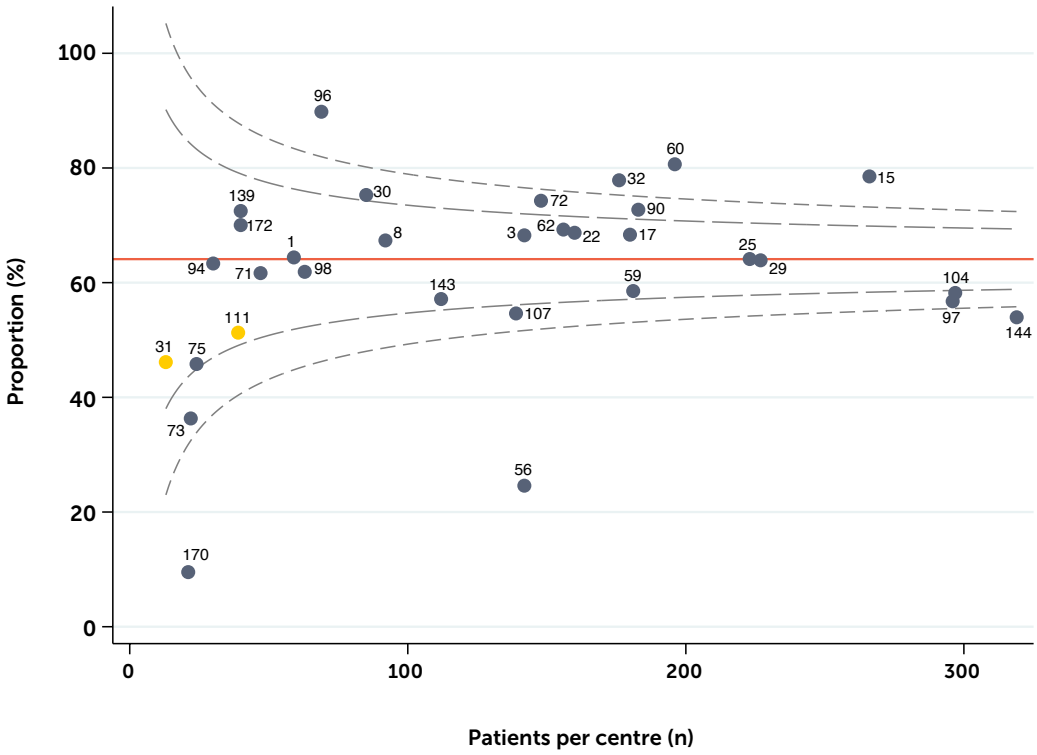
The mean BMI percentile for patients attending paediatric centres/clinics is 58.0%.

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



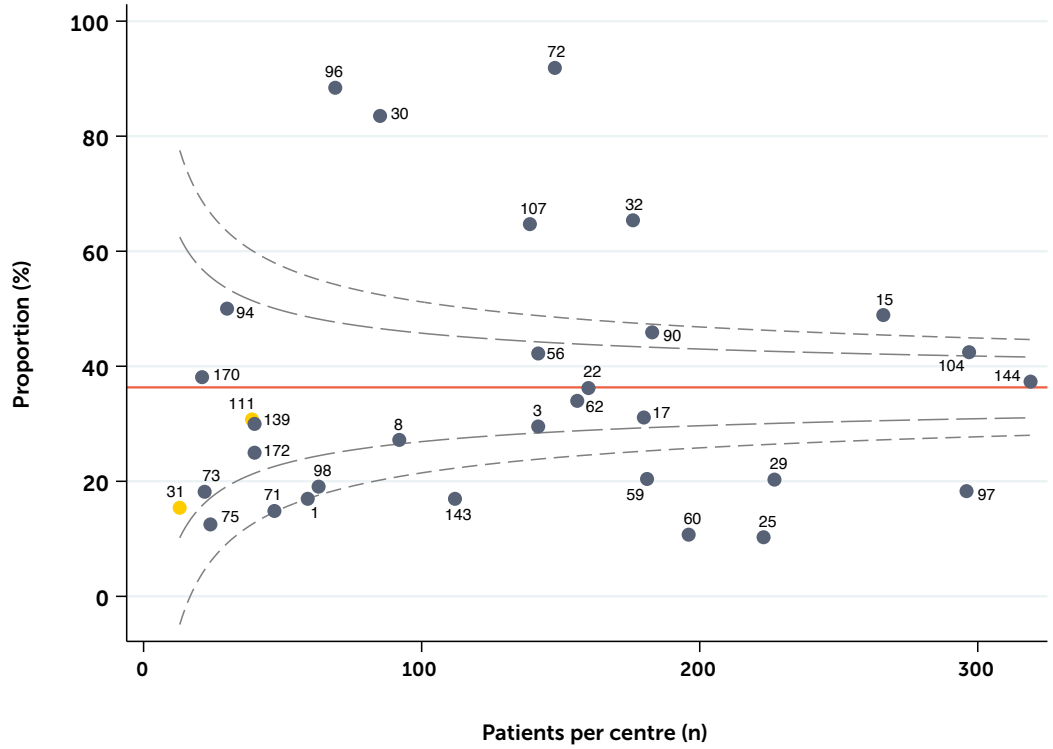
The proportion of patients with chronic *Pseudomonas aeruginosa* in paediatric centres/clinics is 4.8%.

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



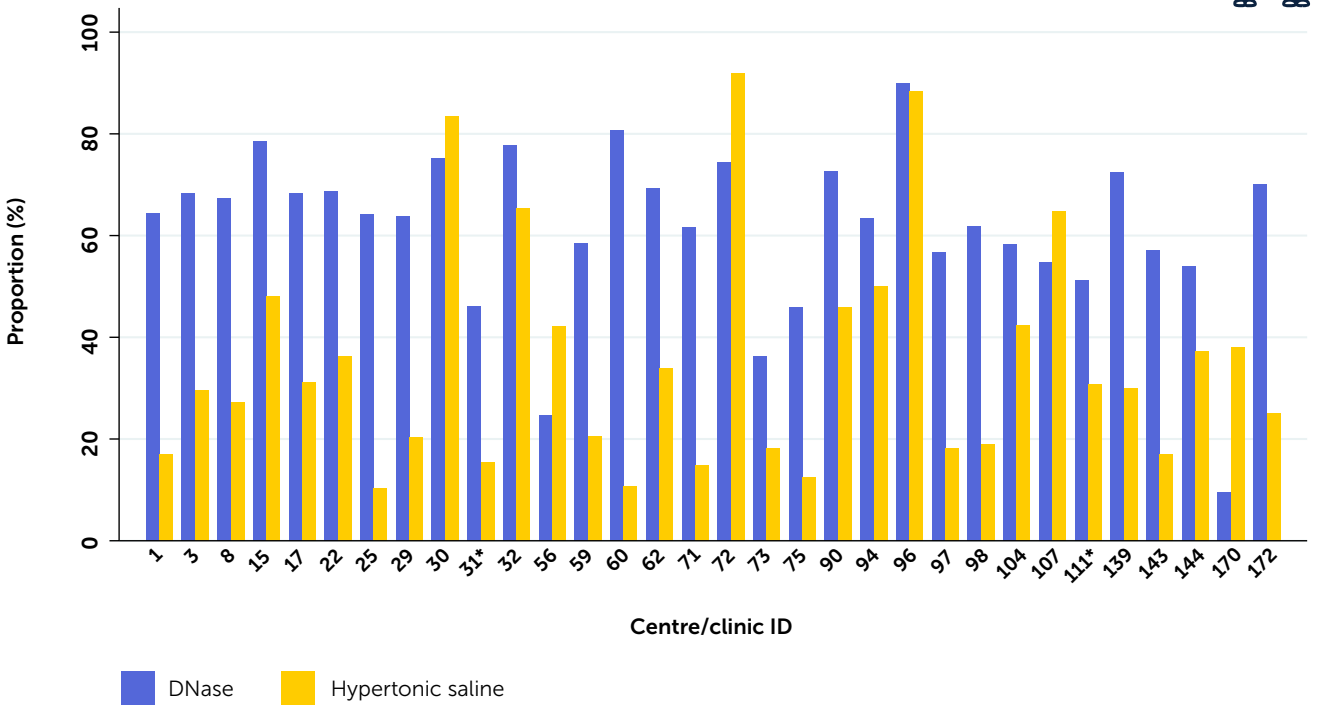
The proportion of patients receiving DNase treatment in paediatric centres/clinics is 64.1%.

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 36.3%.

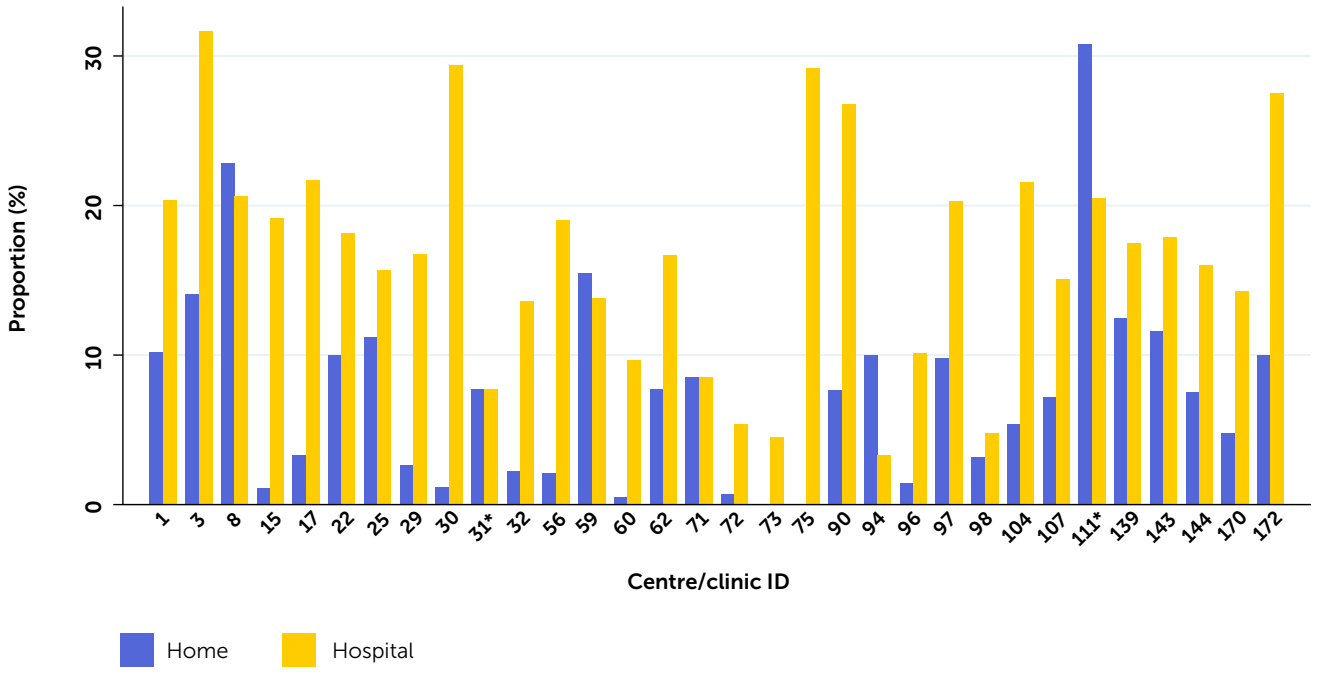
2.7 Proportion of patients receiving DNase/hypertonic saline/ mannitol treatment by paediatric centre/clinic



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

2.8 IV use by paediatric centre/clinic

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

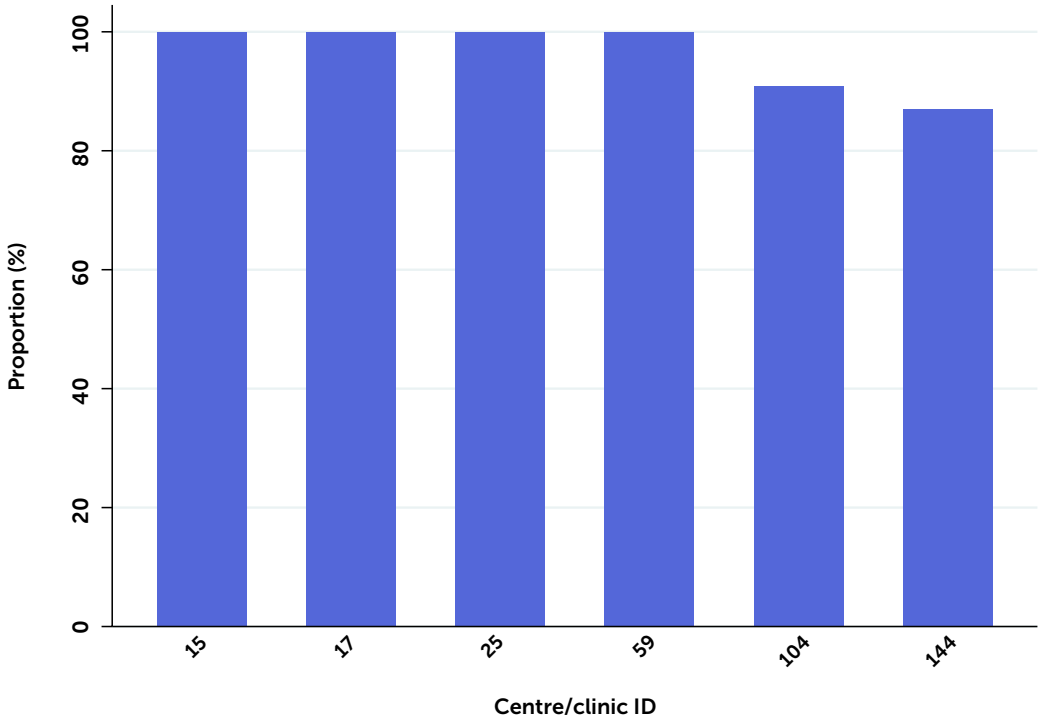


The proportion of patients receiving IVs at home was 7.0% and in hospital was 17.7%. The proportion receiving any IVs was 19.7%.

*Stand-alone clinics

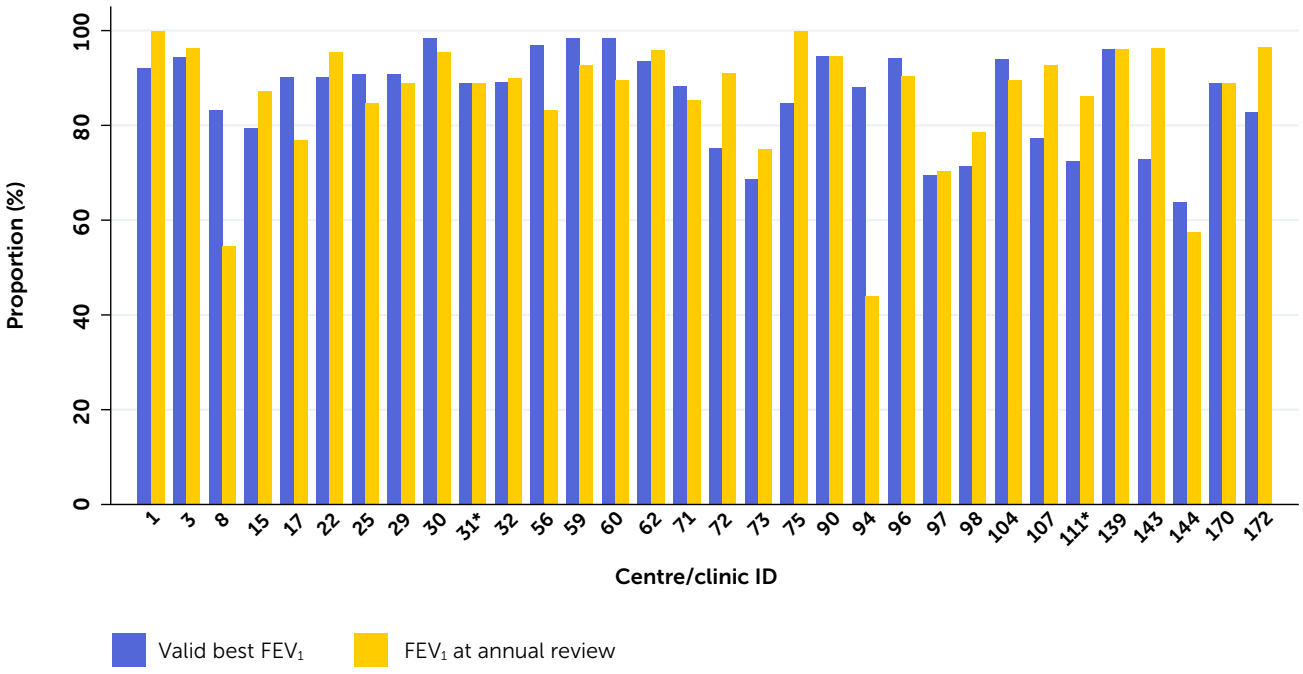
2.9 Inhaled antibiotic use for patients with chronic *Pseudomonas aeruginosa*, by paediatric centre/clinic

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



91.5% of patients with chronic *P. aeruginosa* received inhaled antibiotics.

2.10 Data completeness by paediatric centre/clinic**



*Stand-alone clinics

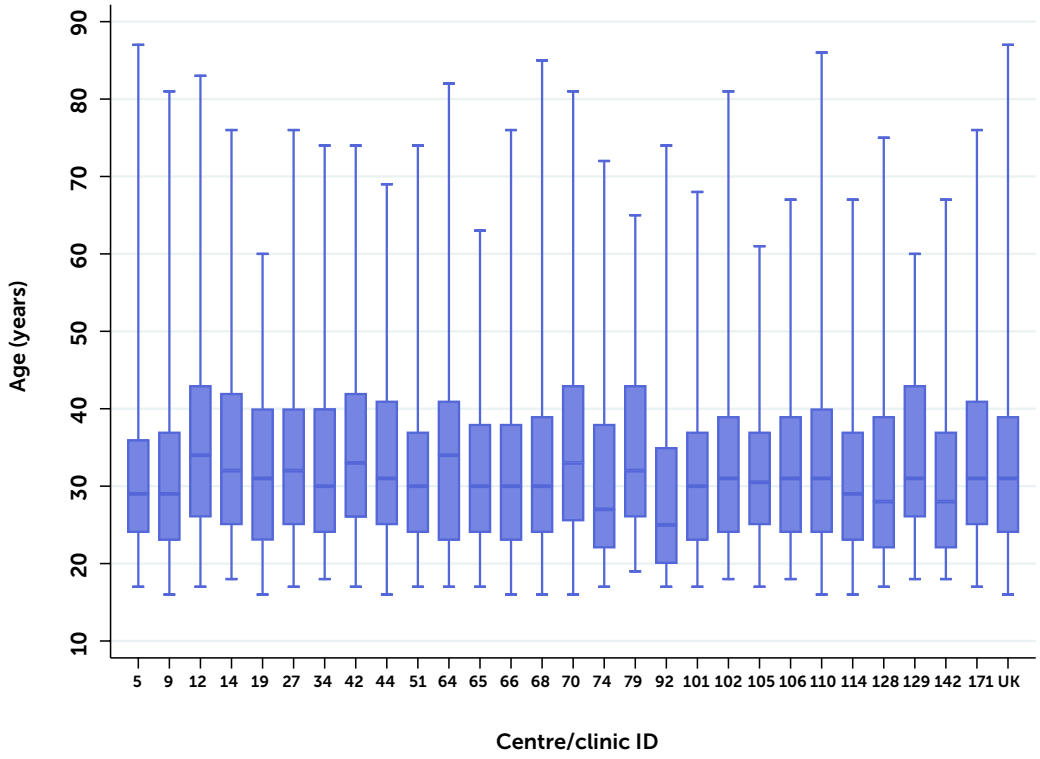
**The chart above shows the proportion of patients who had a valid best FEV_1% and an FEV_1% at annual review, excluding patients under six years of age. Best FEV_1% was considered valid if it was not missing, and the per cent predicted was not more than 0.5% lower than the annual review value. For some 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

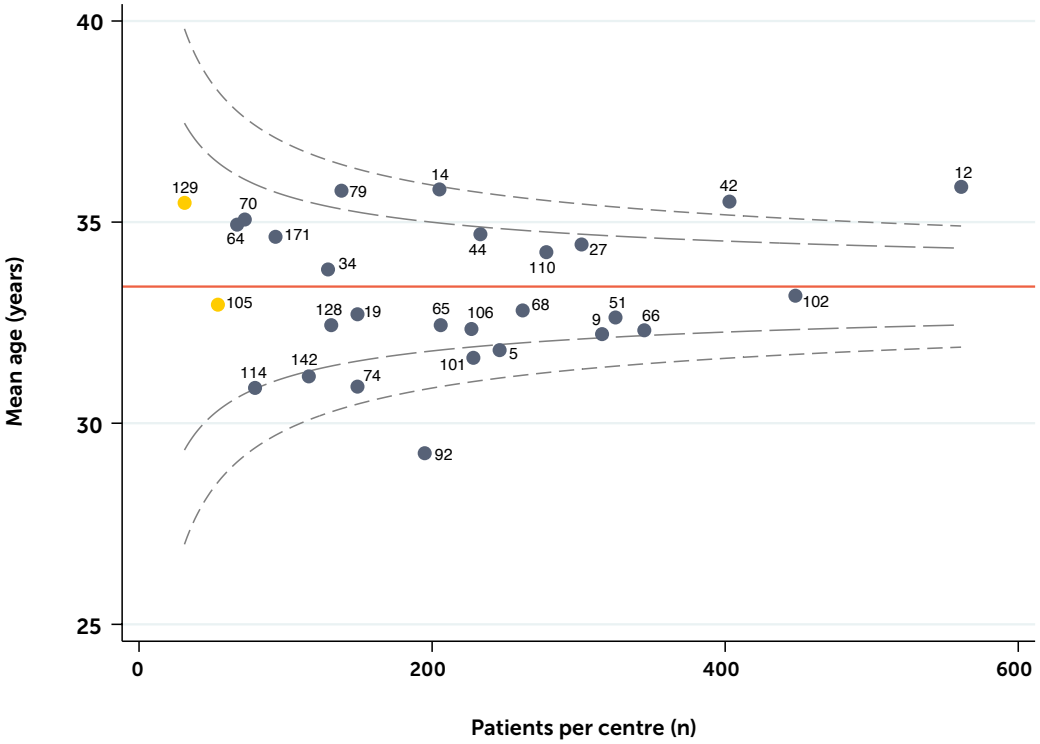
N=5,988

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

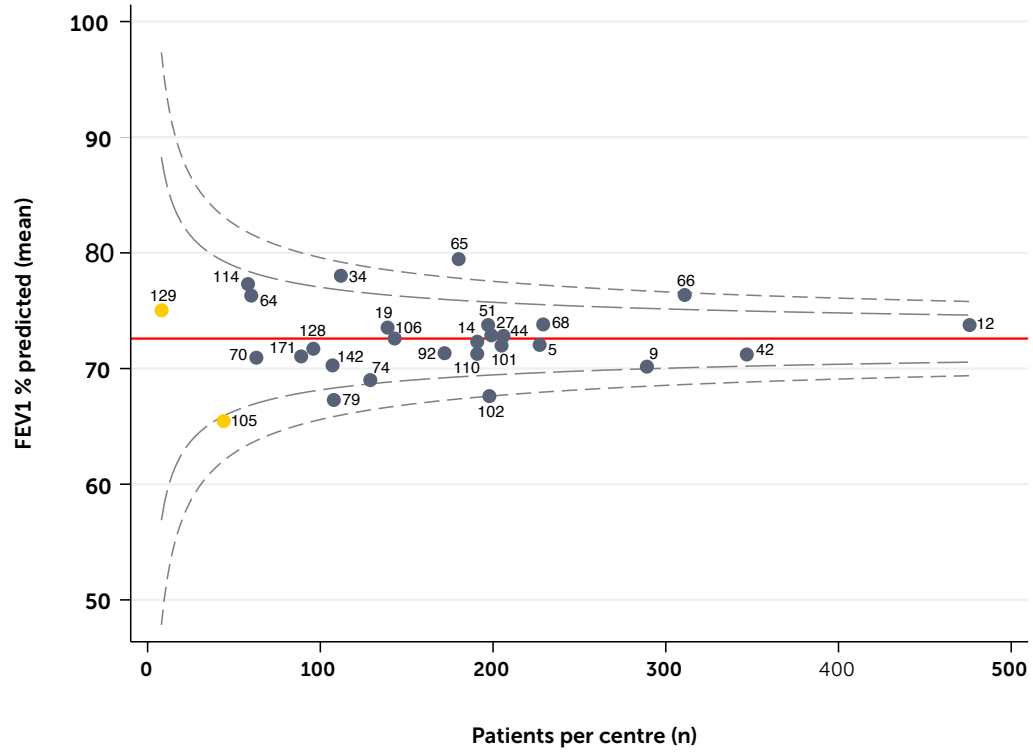
3.1 Age distribution by adults centre / clinic



The funnel plot below shows how the mean age in adult centres compares to the national mean. In 2021 the national mean age of patients at CF centres was 32.7 years.

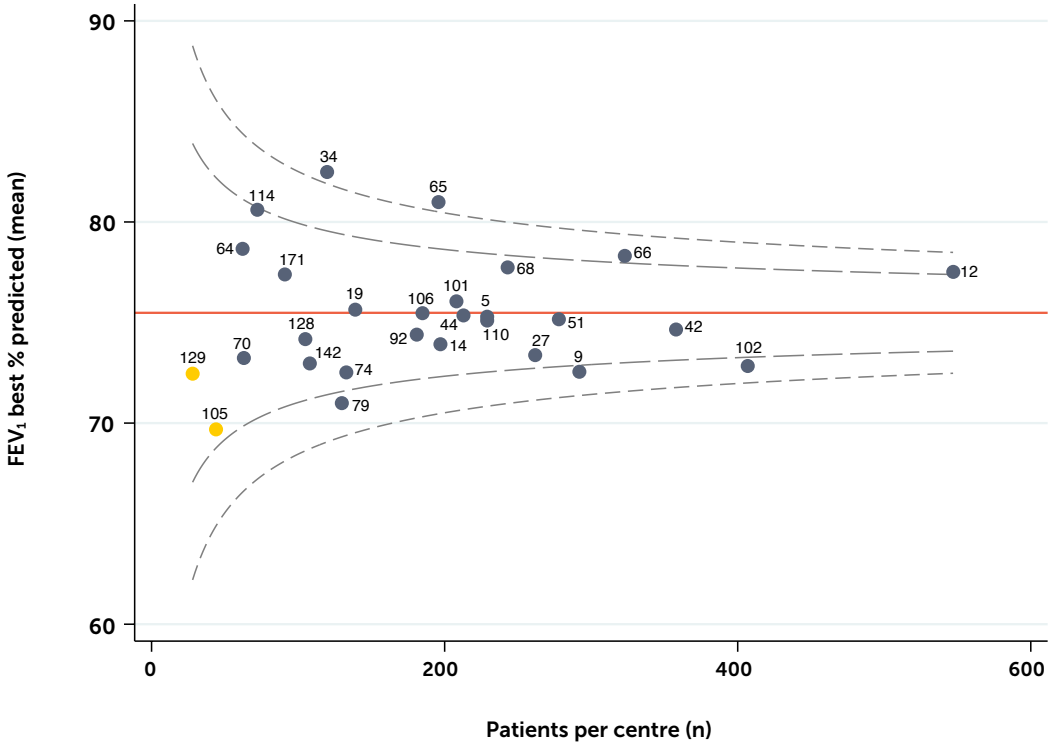


3.2 Age adjusted FEV_1 % predicted at annual review in patients without a history of lung transplant, by adult centre / clinic



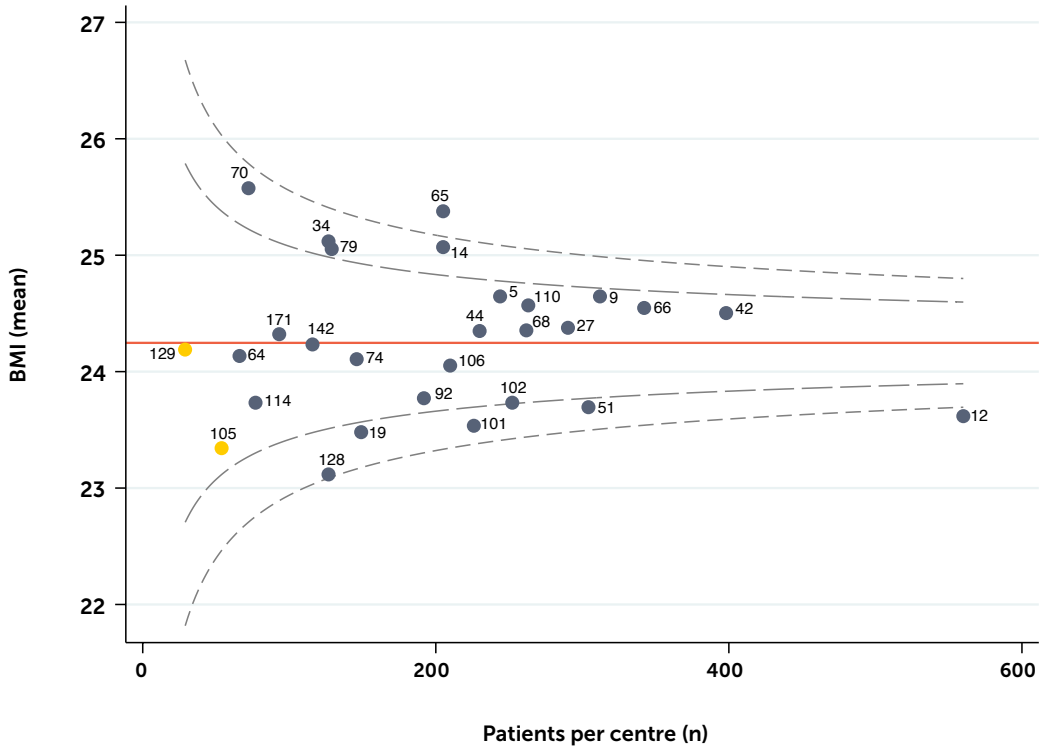
The mean FEV_1% predicted in adult centres/clinics is 72.6%.

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



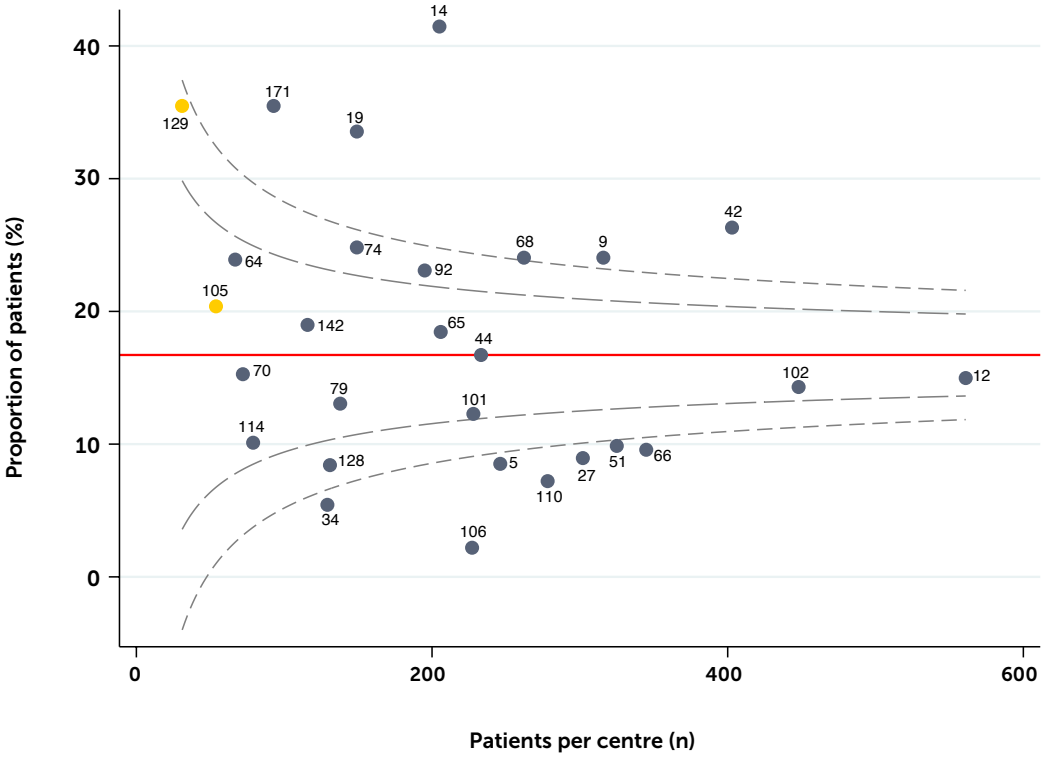
In 2021 the national mean was 75.5%. Where Best FEV_1% predicted was missing, or lower than the FEV_1 at annual review, the FEV_1% value at annual review was used.

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



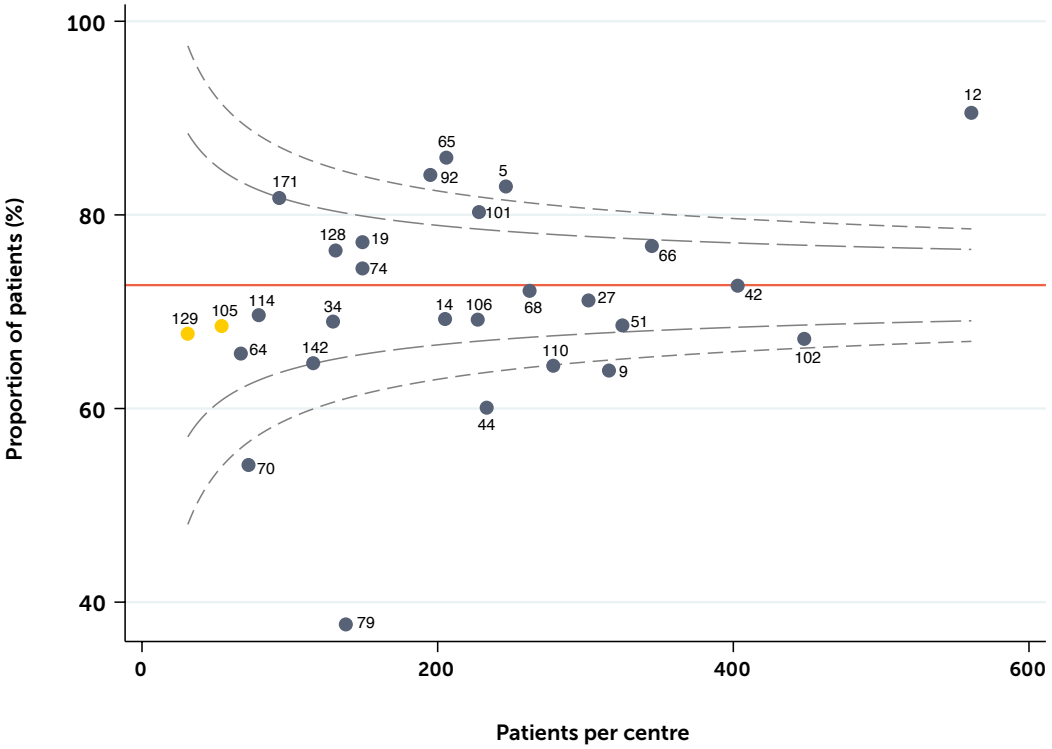
The mean BMI in adult centres/clinics is 24.2.

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



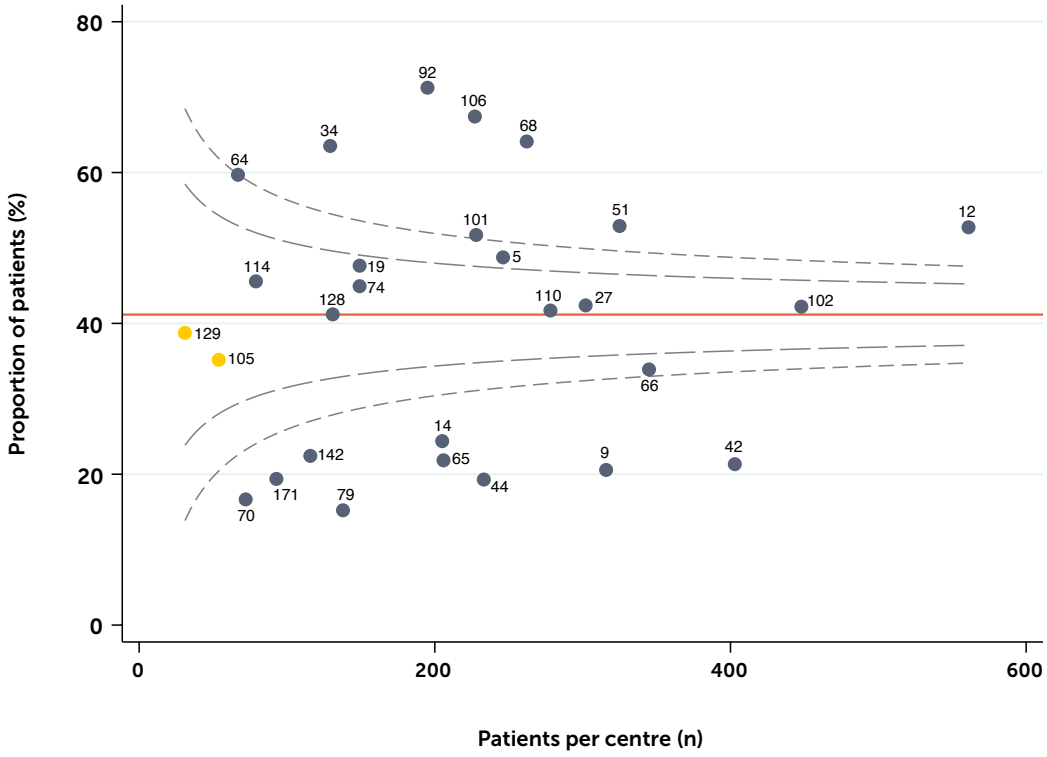
The proportion of patients with chronic *P. aeruginosa* in adult centres/clinics is 32.6%.

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



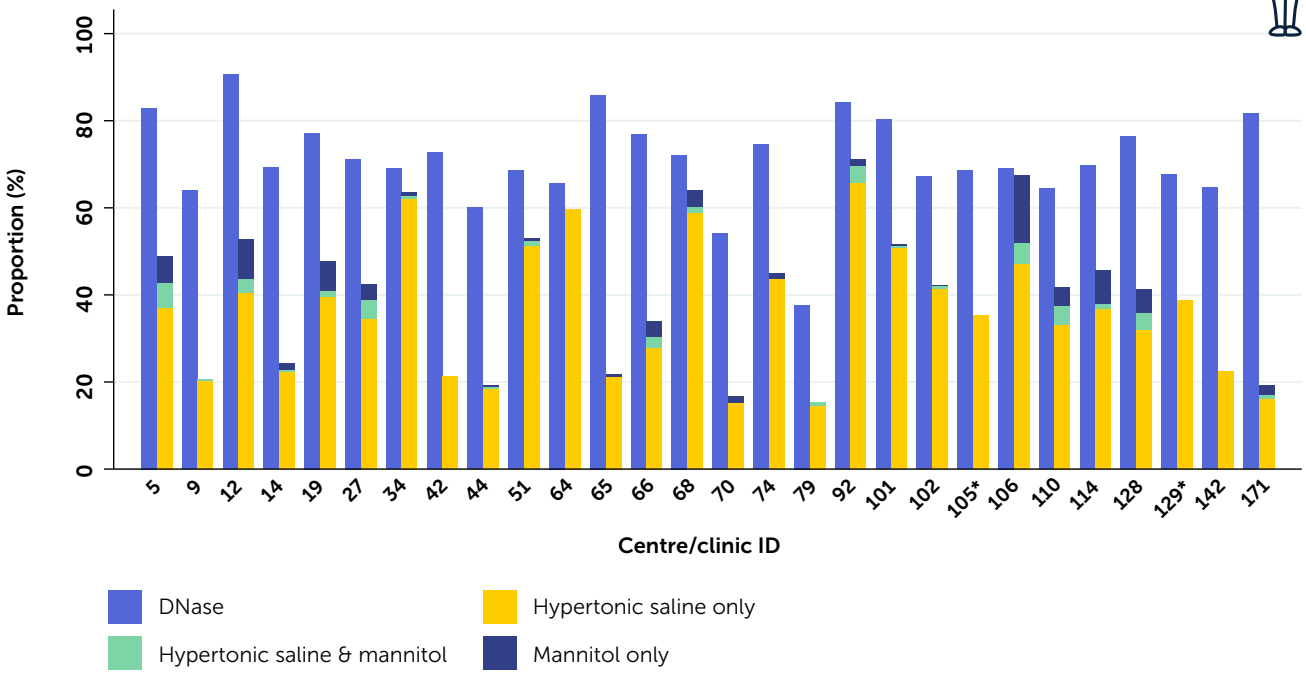
The proportion of patients receiving DNase treatment in adult centres/clinics is 72.7%.

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



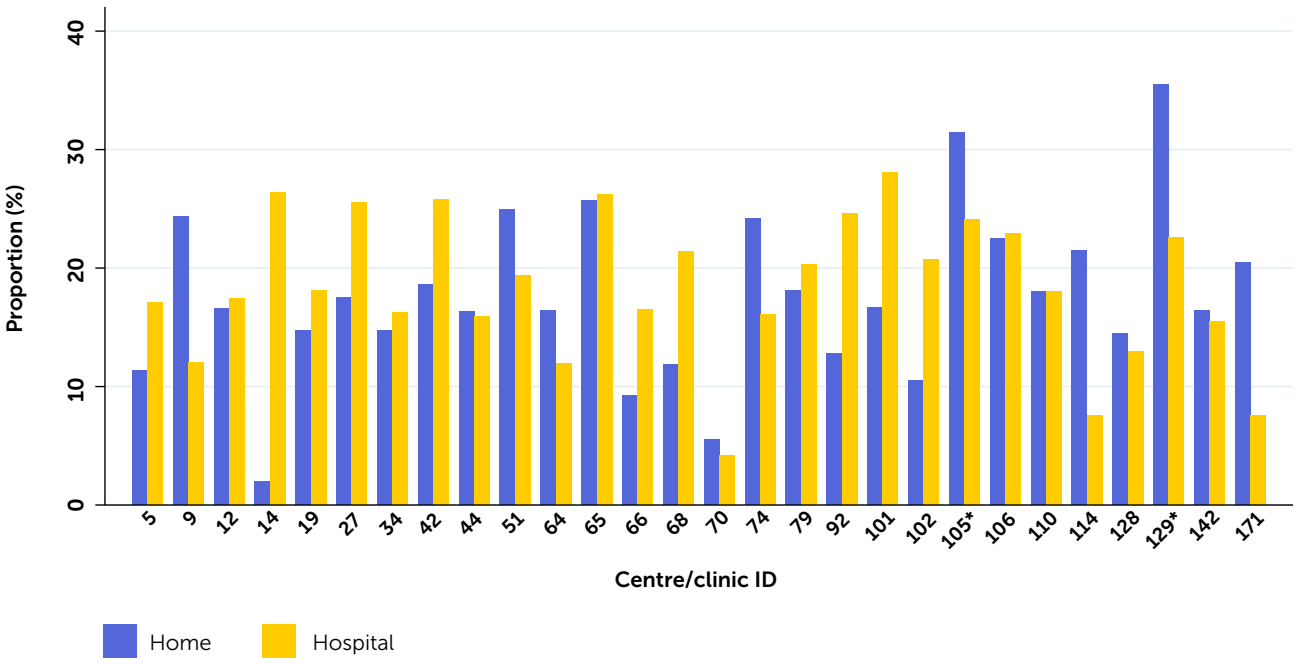
The proportion of patients receiving hypertonic saline or mannitol treatment in adult centres/ clinics is 41.2%.

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

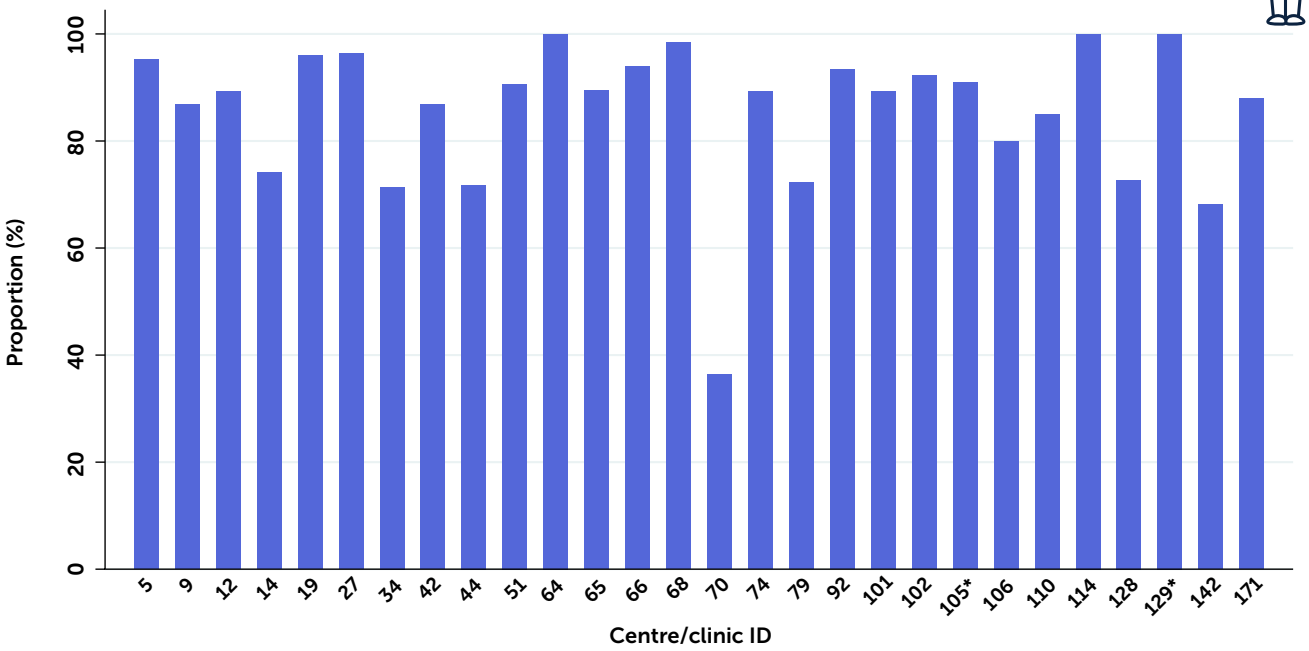
The chart below shows the proportion of patients with at least one IV day at home and/or in hospital. Patients may have a combination of home and hospital IV days.



The proportion of patients in adult centres receiving IV antibiotics at home was 17.6% and in hospital was 18.4%. The proportion receiving any IVs was 27.5%.

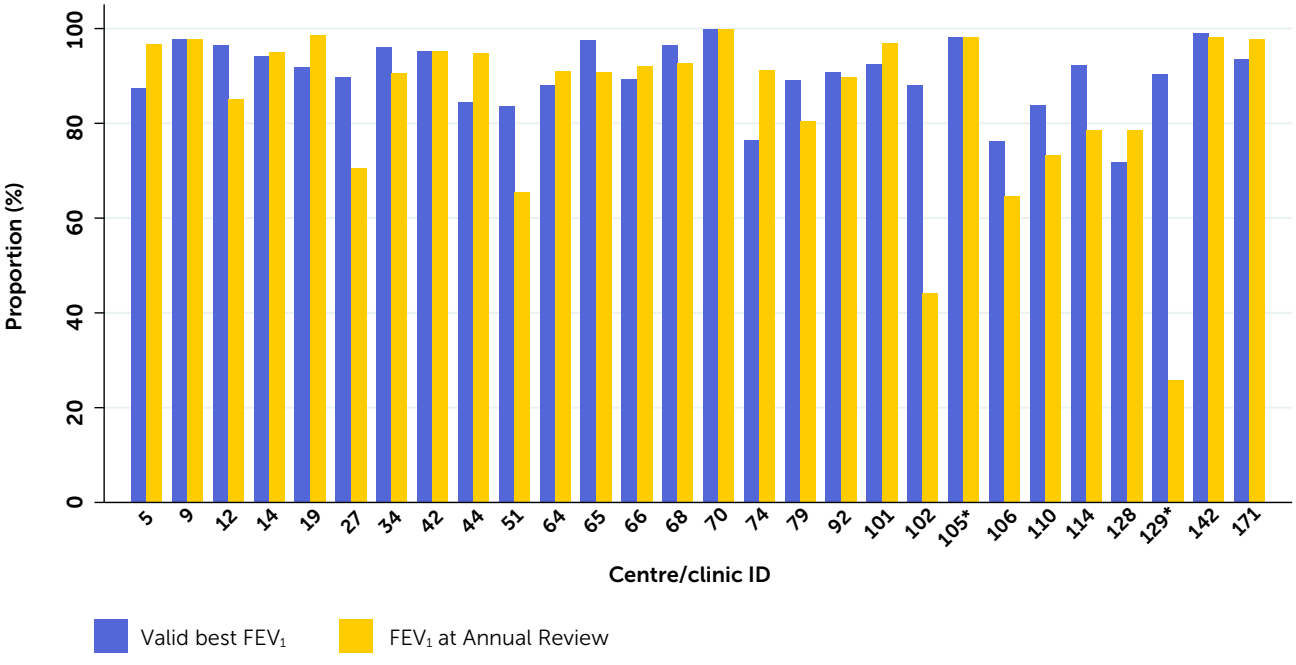
*Stand-alone clinics

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



85.6% 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
2021	1 January 2021 – 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 (eg BMI). It gives a range of results that is likely to include the 'true' value for the population. A narrow confidence interval indicates a more precise estimate. A wide confidence interval indicates more uncertainty about the true value of the clinical measure, often because a small group of patients has been studied. The confidence interval is usually stated as '95% CI', which means that the range of values has a 95 in 100 chance of including the 'true' value.
Enzymes	Biological molecules that help complex reactions, such as the digestion of food, occur in the body.
FEV_1 (forced expiratory volume in one second)	This is the amount of air that a person can blow out of the lungs in the first second of a forced exhaled breath. People with healthy lungs can blow out most of the air held in this time.
FEV_1% predicted	The FEV_1 can be converted from absolute litres of air blown out into a predicted percentage (%). A healthy range for % predicted is calculated from a very large population sample, and is normally considered to be between 80-120% predicted.
Fibrosing colonopathy	A condition causing narrowing of part of the colon.
Gall bladder	The small sac-shaped organ under the liver that stores bile after it is secreted by the liver, before it is released into the intestine.
Gastrointestinal (GI) tract	The GI tract is an organ system responsible for digesting food, absorbing nutrients and expelling waste.
Genotype	Part of the genetic makeup of a cell, organism or individual that usually controls a particular characteristic (known as a phenotype).
GORD (gastro-esophageal 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
Commissioner, England	Kathy	Blacker	NHS England
CF physician - Paediatrics	Malcolm	Brodlie	Newcastle Paediatric CF Centre
CF physician – Paediatrics*	Siobhán	Carr	Royal Brompton Hospital
Analytical team rep †	Susan	Charman	Cystic Fibrosis Trust
Head of Healthcare Data and Pharmacovigilance #	Sarah	Clarke	Cystic Fibrosis Trust
Director of Data & Quality Improvement	Rebecca	Cosgriff	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 Centre Data Manager	Lance	Dennard	Lewisham Hospital, London
CF physician – Adults	Jamie	Duckers	All Wales Adult CF Centre, Cardiff
Registry data manager †	Elaine	Gunn	Cystic Fibrosis Trust
Allied Health Professional	Rebecca	Heise	Kings College Adult CF Centre
CF Centre Data Manager	Erin	Hodgetts	North West Midlands Adults & Paediatric CF Centres
Person with CF	Flora	Kennedy-McConnell	N/A
CF physician - Adults	Simon	Range	Leicester Adult CF Centre
Commissioner, Wales	Andrea	Richards	Welsh Commissioning Board
Commissioner, Scotland	David	Steele	NHS Scotland
Parent representative	Vacant		
Registry development manager †	Mary	Yip	Cystic Fibrosis Trust

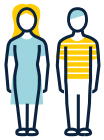
*Chair † Non-voting member # Caldicott guardian

UK CF Registry Research Committee

Role	Forename	Surname	Organisation
Pharmacovigilance PI	Diana	Bilton	Royal Brompton Hospital, London
Pharmacovigilance PI, CF physician – paediatrics	Siobhán	Carr	Royal Brompton Hospital, London
Analytical team rep †	Susan	Charman	Cystic Fibrosis Trust
Head of Healthcare Data & Pharmacovigilance #	Sarah	Clarke	Cystic Fibrosis Trust
Director of Data & QI	Rebecca	Cosgriff	Cystic Fibrosis Trust
Pharmacovigilance PI, CF physician – paediatrics *	Steve	Cunningham	Royal Hospital for Sick Children, Edinburgh
Parent Representative	Marian	Dmochowska	N/A
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, London
Person with CF	James	Thompson	N/A
Registry development manager †	Mary	Yip	Cystic Fibrosis Trust

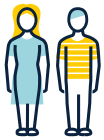
*Chair † Non-voting member # Caldicott guardian

Appendix 2: Centre-level data tables



Paediatric centres/clinics providing data in 2021 – ordered alphabetically by country/city

Location	Name	Clinic ID	Total Active	Number with annual review
England				
Birmingham	Birmingham Children's Hospital	104	314	297
Brighton	Royal Alexandra Children's Hospital	172	55	40
Bristol	Bristol Royal Hospital for Children	32	187	176
Cambridge	Addenbrooke's Hospital	107	147	139
Cornwall	Royal Cornwall Hospital	94	37	30
Exeter	Royal Devon & Exeter Hospital	96	74	69
Hull	Hull University Teaching Hospitals	111	43	39
Leeds	St James's University Hospital	25	230	223
Leicester	Leicester Royal Infirmary	1	64	59
Liverpool	Alder Hey Children's Hospital	97	308	296
London - Central	Great Ormond Street Hospital for Children	90	197	183
London - East	Royal London Hospital	30	92	85
London - South East	King's College Hospital	17	194	180
London - South West	Royal Brompton Hospital	15	293	266
Manchester	Royal Manchester Children's Hospital	144	339	319
Newcastle	Great North Children's Hospital	59	194	181
North West Midlands	University Hospital of North Midlands	8	95	92
Norwich	Norfolk & Norwich University Hospital	98	66	63
Nottingham	Nottingham University Hospitals	62	165	156
Oxford	John Radcliffe Hospital	22	165	160
Plymouth	Derriford Hospital	139	41	40
Sheffield	Sheffield Children's Hospital	3	147	142
Southampton	Southampton General Hospital	29	235	227
Teesside	James Cook University Hospital	71	52	47
Northern Ireland				
Belfast	Royal Belfast Hospital for Sick Children	60	212	196
Scotland				
Aberdeen	Royal Aberdeen Children's Hospital	75	26	24
Ayr	University Hospital Crosshouse	170	23	21
Dundee	Ninewells Hospital	73	23	22
Edinburgh	Royal Hospital for Sick Children	143	137	112
Glasgow	Royal Hospital for Sick Children	56	164	142
Inverness	Raigmore Hospital	31	16	13
Wales				
Cardiff	Children's Hospital for Wales	72	162	148



	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	9.2	8.9	182	90.1	90.1	92.6	200	95.9	95.9	97.3
172	8.6	8.5	28	101.9	101.6	103.2	29	105.6	105.3	105.5
32	9.2	9.1	116	91.7	91.6	92.9	122	96.7	96.6	96.3
107	8.6	9.0	90	94.6	94.4	95.8	93	97.3	97.0	97.2
94	10.3	9.5	11	78.5	78.6	78.6	22	91.0	91.0	90.9
96	9.5	9.0	47	85.2	85.0	88.1	51	89.8	89.6	93.6
111	9.1	9.4	25	86.9	86.7	88.1	28	92.0	91.9	93.6
25	9.1	9.3	138	90.4	90.3	93.5	150	96.7	96.5	96.5
1	8.0	8.3	38	89.4	89.1	90.5	38	96.3	96.0	96.8
97	9.4	9.3	152	89.2	89.2	90.8	176	94.0	94.0	93.8
90	8.8	9.1	121	93.1	93.0	93.5	124	97.7	97.5	97.6
30	9.9	10.7	63	97.6	97.6	98.1	65	100.7	100.6	101.3
17	8.3	7.9	87	91.1	91.0	93.4	107	93.5	93.3	95.8
15	9.1	9.1	170	92.9	92.8	94.5	188	98.9	98.7	99.2
144	9.1	9.3	129	92.4	92.4	94.7	168	97.8	97.8	98.0
59	8.5	8.8	115	94.4	94.3	97.4	122	97.7	97.5	97.7
8	9.8	10.7	36	88.9	89.0	88.8	60	95.0	95.0	96.5
98	8.9	9.0	33	88.4	88.4	85.0	41	98.3	98.3	95.6
62	10.1	10.0	119	87.4	87.4	87.7	120	93.8	93.8	94.7
22	9.1	9.5	107	96.3	96.3	95.5	109	100.4	100.3	100.2
139	8.2	8.5	25	91.4	91.2	95.9	25	96.1	95.9	99.4
3	9.3	9.3	104	94.2	94.1	95.3	106	109.5	109.4	101.0
29	9.1	9.1	137	92.6	92.6	94.9	148	99.2	99.2	100.0
71	9.7	10.8	29	91.5	91.5	91.4	32	97.5	97.6	95.6
60	9.1	9.3	119	94.3	94.3	94.8	132	96.9	96.9	96.7
75	7.8	7.9	13	95.0	95.0	97.6	13	101.5	101.3	100.3
170	10.6	10.1	16	92.9	92.9	98.7	17	102.1	102.0	103.3
73	8.7	9.7	12	97.8	97.6	95.4	12	99.9	99.7	99.7
143	9.5	10.0	78	93.9	93.9	92.5	79	96.7	96.7	96.9
56	9.1	9.8	84	93.9	93.8	92.7	101	99.2	99.1	98.4
31	8.7	7.9	8	95.7	95.7	92.7	8	98.7	98.6	97.1
72	9.1	10.1	92	89.0	89.0	89.2	92	93.7	93.7	93.9

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

			BMI			
Location	Name	Clinic ID	Number	Mean - unadjusted	Mean - adjusted	Median
England						
Birmingham	Birmingham Children's Hospital	104	257	55.2	55.2	56.0
Brighton	Royal Alexandra Children's Hospital	172	36	52.8	52.8	52.3
Bristol	Bristol Royal Hospital for Children	32	161	57.1	57.1	61.4
Cambridge	Addenbrooke's Hospital	107	124	60.3	60.3	60.2
Cornwall	Royal Cornwall Hospital	94	27	71.4	71.4	75.8
Exeter	Royal Devon & Exeter Hospital	96	64	58.0	58.0	63.7
Hull	Hull University Teaching Hospitals	111	36	58.6	58.6	57.8
Leeds	St James's University Hospital	25	189	58.9	58.9	63.8
Leicester	Leicester Royal Infirmary	1	53	53.0	53.0	56.3
Liverpool	Alder Hey Children's Hospital	97	255	60.4	60.3	65.1
London - Central	Great Ormond Street Hospital for Children	90	170	51.1	51.1	50.7
London - East	Royal London Hospital	30	80	54.4	54.4	53.1
London - South East	King's College Hospital	17	155	56.9	56.9	57.2
London - South West	Royal Brompton Hospital	15	245	58.5	58.5	58.3
Manchester	Royal Manchester Children's Hospital	144	203	54.0	54.0	53.9
Newcastle	Great North Children's Hospital	59	153	63.2	63.2	70.4
North West Midlands	University Hospital of North Midlands	8	48	55.7	55.7	59.2
Norwich	Norfolk & Norwich University Hospital	98	57	62.8	62.8	66.2
Nottingham	Nottingham University Hospitals	62	146	54.2	54.2	56.1
Oxford	John Radcliffe Hospital	22	148	58.5	58.5	57.9
Plymouth	Derriford Hospital	139	36	59.2	59.2	64.9
Sheffield	Sheffield Children's Hospital	3	132	59.3	59.3	61.0
Southampton	Southampton General Hospital	29	202	59.4	59.4	64.3
Teesside	James Cook University Hospital	71	42	68.2	68.2	76.7
Northern Ireland						
Belfast	Royal Belfast Hospital for Sick Children	60	175	59.3	59.3	65.4
Scotland						
Aberdeen	Royal Aberdeen Children's Hospital	75	23	57.2	57.3	61.4
Ayr	University Hospital Crosshouse	170	20	70.0	70.0	74.5
Dundee	Ninewells Hospital	73	16	57.7	57.7	62.2
Edinburgh	Royal Hospital for Sick Children	143	102	57.9	57.9	60.3
Glasgow	Royal Hospital for Sick Children	56	118	56.5	56.5	63.1
Inverness	Raigmore Hospital	31	11	63.9	63.9	65.0
Wales						
Cardiff	Children's Hospital for Wales	72	137	60.3	60.3	65.8

	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 (%)
104	11	3.7	64	21.5	173	58.2	126	42.4	10	90.9
172	<5	-*	11	27.5	28	70.0	10	25.0	<5	100.0
32	5	2.8	25	14.2	137	77.8	115	65.3	5	100.0
107	5	3.6	23	16.5	76	54.7	90	64.7	<5	80.0
94	0	0.0	<5	-*	19	63.3	15	50.0	0	0.0
96	<5	-*	7	10.1	62	89.9	61	88.4	<5	100.0
111	7	17.9	13	33.3	20	51.3	12	30.8	<5	42.9
25	15	6.7	45	20.2	143	64.1	23	10.3	15	100.0
1	<5	-*	12	20.3	38	64.4	10	16.9	<5	100.0
97	8	2.7	67	22.6	168	56.8	54	18.2	8	100.0
90	8	4.4	49	26.8	133	72.7	84	45.9	8	100.0
30	8	9.4	25	29.4	64	75.3	71	83.5	8	100.0
17	18	10.0	39	21.7	123	68.3	56	31.1	18	100.0
15	19	7.1	54	20.3	209	78.6	130	48.9	19	100.0
144	23	7.2	56	17.6	172	53.9	119	37.3	20	87.0
59	11	6.1	41	22.7	106	58.6	37	20.4	11	100.0
8	7	7.6	28	30.4	62	67.4	25	27.2	6	85.7
98	0	0.0	<5	-*	39	61.9	12	19.0	0	0.0
62	<5	-*	30	19.2	108	69.2	53	34.0	<5	100.0
22	7	4.4	35	21.9	110	68.8	58	36.3	5	71.4
139	0	0.0	8	20.0	29	72.5	12	30.0	0	0.0
3	6	4.2	47	33.1	97	68.3	42	29.6	6	100.0
29	7	3.1	38	16.7	145	63.9	46	20.3	5	71.4
71	<5	-*	5	10.6	29	61.7	7	14.9	<5	100.0
60	6	3.1	20	10.2	158	80.6	21	10.7	6	100.0
75	<5	-*	7	29.2	11	45.8	<5	-*	<5	100.0
170	<5	-*	<5	-*	<5	-*	8	38.1	<5	100.0
73	<5	-*	<5	-*	8	36.4	<5	-*	<5	50.0
143	6	5.4	26	23.2	64	57.1	19	17.0	6	100.0
56	<5	-*	27	19.0	35	24.6	60	42.3	0	0.0
31	<5	-*	<5	7.7	6	46.2	<5	-*	<5	100.0
72	7	4.7	9	6.1	110	74.3	136	91.9	7	100.0

* Redacted to adhere to statistical disclosure guidelines.

Appendix 2: Centre-level data tables

Adult centres/clinics providing data in 2021 – ordered alphabetically by country/city

Location	Name	Clinic ID	Total active	Number with annual review
England				
Birmingham	Birmingham Heartlands Hospital	27	323	302
Bristol	Bristol Royal Infirmary	106	239	227
Cambridge	The Royal Papworth Hospital	51	350	325
Cornwall	Royal Cornwall Hospital	129	38	31
Exeter	Royal Devon & Exeter Hospital	34	133	129
Frimley	Frimley Park Hospital	19	151	149
Leeds	St James's University Hospital	42	410	403
Leicester	Glenfield Hospital	142	117	116
Liverpool	Liverpool Heart and Chest Hospital	66	361	345
London - East	St. Bartholomew's Hospital	92	215	195
London - South East	University Hospital Lewisham	105	55	54
London - South East	King's College Hospital	5	258	246
London - South West	Royal Brompton Hospital	12	578	561
Manchester	Wythenshawe Hospital	102	472	448
Newcastle	Royal Victoria Infirmary	9	326	316
North West Midlands	University Hospital of North Midlands	74	150	149
Norwich	Norfolk & Norwich University Hospital	114	82	79
Nottingham	Nottingham University Hospitals	101	239	228
Oxford	Oxford University Hospitals	128	149	131
Plymouth	Derriford Hospital	64	67	67
Sheffield	Northern General Hospital	65	214	206
Southampton	Southampton General Hospital	110	291	278
York	York Hospital	171	93	93
Northern Ireland				
Belfast	Belfast City Hospital	14	273	205
Scotland				
Aberdeen	Aberdeen Royal Infirmary	70	74	72
Edinburgh	Western General Hospital	44	243	233
Glasgow	Queen Elizabeth University Hospital	79	219	138
Wales				
Llandough	Llandough Hospital	68	292	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	32.0	199	72.1	72.9	74.5	262	72.9	73.4	74.5
106	32.3	31.0	143	72.9	72.6	76.6	185	75.8	75.5	78.6
51	32.6	30.4	197	74.4	73.8	75.1	278	75.7	75.2	74.7
129	35.5	31.8	8	71.4	75.0	70.1	28	72.0	72.5	71.4
34	33.8	30.9	112	77.4	78.0	79.6	120	82.2	82.5	87.6
19	32.7	31.9	139	73.9	73.5	76.8	139	76.0	75.6	77.9
42	35.5	33.6	347	70.3	71.2	73.8	358	73.7	74.7	77.9
142	31.2	28.8	107	72.1	70.3	78.4	108	74.9	73.0	79.6
66	32.3	30.8	311	76.8	76.4	79.1	323	78.7	78.3	81.1
92	29.2	25.9	172	74.0	71.3	76.2	181	77.3	74.4	80.0
105	32.9	31.0	44	65.7	65.5	64.9	44	69.9	69.7	71.2
5	31.8	29.8	227	72.8	72.1	76.0	229	76.1	75.3	82.6
12	35.9	34.2	476	72.2	73.8	72.2	547	76.2	77.5	76.2
102	33.2	31.0	198	67.5	67.6	67.2	407	72.5	72.9	72.9
9	32.2	29.9	289	71.1	70.1	73.6	292	73.6	72.6	77.3
74	30.9	27.3	129	71.4	69.0	77.3	133	74.7	72.5	81.1
114	30.9	29.1	58	78.0	77.3	82.7	72	81.9	80.6	85.4
101	31.6	30.4	205	73.0	72.0	75.3	208	77.1	76.1	79.4
128	32.4	28.3	96	73.5	71.7	73.4	105	76.0	74.2	76.4
64	34.9	34.1	60	75.9	76.3	78.4	62	78.7	78.7	81.0
65	32.4	30.7	180	79.7	79.5	83.3	196	81.4	81.0	85.9
110	34.3	31.3	191	71.3	71.3	72.4	229	75.3	75.1	76.5
171	34.6	31.3	89	70.6	71.1	70.7	91	77.1	77.4	79.4
14	35.8	32.3	191	71.5	72.3	76.0	197	73.1	73.9	77.1
70	35.1	33.3	63	70.7	71.0	68.7	63	73.0	73.2	73.0
44	34.7	31.9	206	72.7	72.8	73.6	213	75.1	75.3	78.0
79	35.8	32.5	108	65.4	67.3	66.7	130	69.3	71.0	71.0
68	32.8	30.4	229	74.4	73.8	77.0	243	78.4	77.7	82.4

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

			BMI			
Location	Name	Clinic ID	Number	Mean - unadjusted	Mean - adjusted	Median
England						
Birmingham	Birmingham Heartlands Hospital	27	290	24.5	24.4	24.2
Bristol	Bristol Royal Infirmary	106	210	24.0	24.1	23.3
Cambridge	The Royal Papworth Hospital	51	304	23.7	23.7	23.2
Cornwall	Royal Cornwall Hospital	129	29	24.4	24.2	21.9
Exeter	Royal Devon & Exeter Hospital	34	127	25.2	25.1	24.4
Frimley	Frimley Park Hospital	19	149	23.5	23.5	23.4
Leeds	St James's University Hospital	42	398	24.7	24.5	24.3
Leicester	Glenfield Hospital	142	116	24.0	24.2	23.8
Liverpool	Liverpool Heart and Chest Hospital	66	342	24.5	24.5	23.9
London - East	St. Bartholomew's Hospital	92	192	23.4	23.8	22.6
London - South East	University Hospital Lewisham	105	54	23.3	23.3	22.5
London - South East	King's College Hospital	5	244	24.5	24.6	24.0
London - South West	Royal Brompton Hospital	12	560	23.8	23.6	23.5
Manchester	Wythenshawe Hospital	102	252	23.6	23.7	23.2
Newcastle	Royal Victoria Infirmary	9	312	24.5	24.6	23.6
North West Midlands	University Hospital of North Midlands	74	146	23.9	24.1	23.6
Norwich	Norfolk & Norwich University Hospital	114	77	23.6	23.7	23.1
Nottingham	Nottingham University Hospitals	101	226	23.4	23.5	23.0
Oxford	Oxford University Hospitals	128	127	23.0	23.1	22.7
Plymouth	Derriford Hospital	64	66	24.2	24.1	24.3
Sheffield	Northern General Hospital	65	205	25.3	25.4	24.2
Southampton	Southampton General Hospital	110	263	24.6	24.6	24.0
York	York Hospital	171	93	24.4	24.3	23.7
Northern Ireland						
Belfast	Belfast City Hospital	14	205	25.2	25.1	24.1
Scotland						
Aberdeen	Aberdeen Royal Infirmary	70	72	25.7	25.6	24.6
Edinburgh	Western General Hospital	44	230	24.4	24.4	23.9
Glasgow	Queen Elizabeth University Hospital	79	129	25.2	25.1	24.9
Wales						
Llandough	Llandough Hospital	68	262	24.3	24.4	23.7

	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	27	8.9	96	31.8	215	71.2	128	42.4	26	96.3
106	5	2.2	85	37.4	157	69.2	153	67.4	<5	-*
51	32	9.8	108	33.2	223	68.6	172	52.9	29	90.6
129	11	35.5	15	48.4	21	67.7	12	38.7	11	100.0
34	7	5.4	24	18.6	89	69.0	82	63.6	5	71.4
19	50	33.6	39	26.2	115	77.2	71	47.7	48	96.0
42	106	26.3	134	33.3	293	72.7	86	21.3	92	86.8
142	22	19.0	33	28.4	75	64.7	26	22.4	15	68.2
66	33	9.6	76	22.0	265	76.8	117	33.9	31	93.9
92	45	23.1	55	28.2	164	84.1	139	71.3	42	93.3
105	11	20.4	19	35.2	37	68.5	19	35.2	10	90.9
5	21	8.5	59	24.0	204	82.9	120	48.8	20	95.2
12	84	15.0	142	25.3	508	90.6	296	52.8	75	89.3
102	64	14.3	110	24.6	301	67.2	189	42.2	59	92.2
9	76	24.1	88	27.8	202	63.9	65	20.6	66	86.8
74	37	24.8	48	32.2	111	74.5	67	45.0	33	89.2
114	8	10.1	19	24.1	55	69.6	36	45.6	8	100.0
101	28	12.3	73	32.0	183	80.3	118	51.8	25	89.3
128	11	8.4	27	20.6	100	76.3	54	41.2	8	72.7
64	16	23.9	15	22.4	44	65.7	40	59.7	16	100.0
65	38	18.4	85	41.3	177	85.9	45	21.8	34	89.5
110	20	7.2	72	25.9	179	64.4	116	41.7	17	85.0
171	33	35.5	20	21.5	76	81.7	18	19.4	29	87.9
14	85	41.5	55	26.8	142	69.3	50	24.4	63	74.1
70	11	15.3	5	6.9	39	54.2	12	16.7	<5	-*
44	39	16.7	49	21.0	140	60.1	45	19.3	28	71.8
79	18	13.0	35	25.4	52	37.7	21	15.2	13	72.2
68	63	24.0	64	24.4	189	72.1	168	64.1	62	98.4

* 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	9714	89.1
c.350G->A	p.Arg117His	R117H	680	6.2
c.1652G->A	p.Gly551Asp	G551D	625	5.7
c.1624G->T	p.Gly542X	G542X	397	3.6
c.489+1G->T		621+1G->T	280	2.6
c.3909C->G	p.Asn1303Lys	N1303K	172	1.6
c.1585-1G->A		1717-1G->A	167	1.5
c.1766+1G->A		1898+1G->A	150	1.4
c.3454G->C	p.Asp1152His	D1152H	144	1.3
c.200C->T	p.Pro67Leu	P67L	144	1.3
c.3140-26A->G		3272-26A->G	118	1.1
c.3528delC	p.Lys1177SerfsX15	3659delC	115	1.1
c.1679G->C	p.Arg560Thr	R560T	102	0.9
c.1519_1521delATC	p.Ile507del	I507del	93	0.9
c.1477C->T	p.Gln493X	Q493X	92	0.8
c.3717+12191C->T		3849+10kbC->T	85	0.8
c.1657C->T	p.Arg553X	R553X	84	0.8
c.254G->A	p.Gly85Glu	G85E	82	0.8
c.178G->T	p.Glu60X	E60X	77	0.7
c.2657+5G->A		2789+5G->A	74	0.7
c.1022_1023insTC	p.Phe342HisfsX28	1154insTC	73	0.7
c.3846G->A	p.Trp1282X	W1282X	61	0.6
c.948delT	p.Phe316LeufsX12	1078delT	56	0.5
c.1646G->A	p.Ser549Asn	S549N	56	0.5
c.1364C->A	p.Ala455Glu	A455E	51	0.5
c.2052delA	p.Lys684AsnfsX38	2184delA	50	0.5
c.617T->G	p.Leu206Trp	L206W	45	0.4
c.1040G->C	p.Arg347Pro	R347P	42	0.4
c.2657+2_2657+3insA		2789+2insA	38	0.3
c.579+3A->G		711+3A->G	35	0.3
c.1558G->T	p.Val520Phe	V520F	33	0.3
c.3484C->T	p.Arg1162X	R1162X	31	0.3
c.1000C->T	p.Arg334Trp	R334W	29	0.3
c.1040G->A	p.Arg347His	R347H	29	0.3
c.1753G->T	p.Glu585X	E585X	28	0.3
c.2988+1G->A		3120+1G->A	27	0.2
c.3718-2477C->T		3849+10kbC->T	26	0.2
c.3472C->T	p.Arg1158X	R1158X	25	0.2

Nucleotide	Protein	Legacy name	N	%
c.1055G->A	p.Arg352Gln	R352Q	24	0.2
c.2583delT	p.Phe861LeufsX3	2711delT	22	0.2
c.1523T->G	p.Phe508Cys	F508C	22	0.2
c.1006_1007insG	p.Ile336SerfsX28	1138insG	22	0.2
c.2490+1G->A		2622+1G->A	21	0.2
c.1367T->C	p.Val456Ala	V456A	21	0.2
c.3873G->C	p.Gln1291His	Q1291H	20	0.2
c.1210-12[5] (AJ574948.1:g.152T[5])		5T	20	0.2
c.1393-1G->A		1525-1G->A	20	0.2
c.1705T->G	p.Tyr569Asp	Y569D	20	0.2
c.532G->A	p.Gly178Arg	G178R	19	0.2
c.2125C->T	p.Arg709X	R709X	19	0.2
c.3197G->A	p.Arg1066His	R1066H	19	0.2
c.2834C->T	p.Ser945Leu	S945L	17	0.2
c.3806T->A	p.Ile1269Asn	I1269N	17	0.2
c.2052_2053insA	p.Gln685ThrfsX4	2184insA	17	0.2
c.349C->T	p.Arg117Cys	R117C	15	0.1
c.658C->T	p.Gln220X	Q220X	14	0.1
c.579+1G->T		711+1G->T	13	0.1
c.2537G->A	p.Trp846X	W846X	13	0.1
c.292C->T	p.Gln98X	Q98X	13	0.1
c.2875delG	p.Ala959HisfsX9	3007delG	13	0.1
c.3737C->T	p.Thr1246Ile	T1246I	12	0.1
c.1029delC	p.Cys343X	1161delC	12	0.1
c.2988G->A		3120G->A	11	0.1
c.1466C->A	p.Ser489X	S489X	11	0.1
c.1645A->C or c.1647T->G or c.1647T->A	p.Ser549Arg	S549R	9	0.1
c.3705T->G	p.Ser1235Arg	S1235R	9	0.1
c.224G->A	p.Arg75Gln	R75Q	9	0.1
c.3196C->T	p.Arg1066Cys	R1066C	9	0.1
c.3468G->A		3600G->A	8	0.1
c.1679+1G->C		1811+1G->C	8	0.1
c.3208C->T	p.Arg1070Trp	R1070W	8	0.1
c.1675G->A	p.Ala559Thr	A559T	8	0.1
c.494T->C	p.Leu165Ser	L165S	8	0.1
c.1687T->A	p.Tyr563Asn	Y563N	7	0.1
c.[1210–12[5];1210-34TG[12]]		5T;TG12	7	0.1

Nucleotide	Protein	Legacy name	N	%
c.2051_2052delAAinsG	p.Lys684SerfsX38	2183AA->G or 2183delAA->G	7	0.1
c.695T->A	p.Val232Asp	V232D	7	0.1
c.3761T->G	p.Leu1254X	L1254X	7	0.1
c.2012delT	p.Leu671X	2143delT	7	0.1
c.54-5940_273+10250del21kb	p.Ser18ArgfsX16	CFTRdele2,3	7	0.1
c.1721C->A	p.Pro574His	P574H	6	0.1
c.1116+1G->A		1248+1G->A	6	0.1
c.4196_4197delTC	p.Cys1400X	4326delTC	6	0.1
c.3884_3885insT	p.Ser1297PhefsX5	4016insT	6	0.1
c.2353C->T	p.Arg785X	R785X	6	0.1
c.709C->G	p.Gln237Glu	Q237E	6	0.1
c.1329_1330insAGAT	p.Ile444ArgfsX3	1461ins4	6	0.1
c.1986_1989delAACT	p.Thr663ArgfsX8	2118del4	6	0.1
c.2128A->T	p.Lys710X	K710X	6	0.1
c.1766+1G->T		1898+1G->T	6	0.1
c.2900T->C	p.Leu967Ser	L967S	6	0.1
c.[1521_1523delCTT; 3080T->C]	p.[Phe508del; Ile1027Thr]	F508del;I1027T	6	0.1
c.[1210–12[5];1210-34TG[13]]		5T;TG13	6	0.1
c.3848G->T	p.Arg1283Met	R1283M	5	0.0
c.223C->T	p.Arg75X	R75X	5	0.0
c.2551C->T	p.Arg851X	R851X	5	0.0
c.262_263delTT	p.Leu88IlefsX22	394delTT	5	0.0
c.2290C->T	p.Arg764X	R764X	5	0.0
c.3718-1G->A		3850-1G->A	5	0.0
c.349C->G	p.Arg117Gly	R117G	5	0.0
c.1679G->A	p.Arg560Lys	R560K	<5	-
c.2464G->T	p.Glu822X	E822X	<5	-
c.1393-2A->G		1525-2A->G	<5	-
c.443T->C	p.Ile148Thr	I148T	<5	-
c.2215delG	p.Val739TyrfsX16	2347delG	<5	-
c.3964-78_4242+577del		CFTRdele22,23	<5	-
c.3988C->T	p.Gln1330X	Q1330X	<5	-
c.1538A->G	p.Asp513Gly	D513G	<5	-
c.(743+1_744-1)_(1584+1_1585-1)dup		CFTRdup6b-10	<5	-
c.2249C->T	p.Pro750Leu	P750L	<5	-
c.1680A->C	p.Arg560Ser	R560S	<5	-
c.850dupA	p.Met284AsnfsX3	977insA	<5	-
c.3353C->T	p.Ser1118Phe	S1118F	<5	-
c.3292T->C	p.Trp1098Arg	W1098R	<5	-
c.2909G->A	p.Gly970Asp	G970D	<5	-
c.165-3C>T		297-3C->T	<5	-
c.3095A->G	p.Tyr1032Cys	Y1032C	<5	-
c.429delT	p.Phe143LeufsX10	557delT	<5	-
c.595C->T	p.His199Tyr	H199Y	<5	-

Nucleotide	Protein	Legacy name	N	%
c.3080T->C	p.Ile1027Thr	I1027T	<5	-
c.274G->A	p.Glu92Lys	E92K	<5	-
c.1651G->A	p.Gly551Ser	G551S	<5	-
c.2600_2601insA	p.Val868SerfsX28	2732insA	<5	-
c.1046C->T	p.Ala349Val	A349V	<5	-
c.91C->T	p.Arg31Cys	R31C	<5	-
c.2491G->T	p.Glu831X	E831X	<5	-
c.1585-8G->A		1717-8G->A	<5	-
c.1736A->G	p.Asp579Gly	D579G	<5	-
c.577G->T	p.Glu193X	E193X	<5	-
c.2991G->C	p.Leu997Phe	L997F	<5	-
c.509G->A	p.Arg170His	R170H	<5	-
c.4147_4148insA	p.Ile1383AsnfsX3	4279insA	<5	-
c.1572C->A	p.Cys524X	C524X	<5	-
c.1724T->A	p.Phe575Tyr	F575Y	<5	-
c.328G->C	p.Asp110His	D110H	<5	-
c.1340delA	p.Lys447ArgfsX2	1471delA	<5	-
c.4046G->A	p.Gly1349Asp	G1349D	<5	-
c.4004T->C	p.Leu1335Pro	L1335P	<5	-
c.1505T>C	p.Ile502Thr	I502T	<5	-
c.350G->T	p.Arg117Leu	R117L	<5	-
c.3659delC	p.Thr1220LysfsX8	3791delC	<5	-
c.2896delA	p.Thr966ArgfsX2	3028delA	<5	-
c.2260G->A	p.Val754Met	V754M	<5	-
c.3205G->A	p.Gly1069Arg	G1069R	<5	-
c.1001G>A	p.Arg334Gln	R334Q	<5	-
c.164+2T>C		296+2T->C	<5	-
c.296C->T	p.Pro99Leu	P99L	<5	-
c.1545_1546delTA	p.Tyr515X	1677delTA	<5	-
c.3763T->C	p.Ser1255Pro	S1255P	<5	-
c.2780T->C	p.Leu927Pro	L927P	<5	-
c.3297C->A	p.Phe1099Leu	F1099L	<5	-
c.1007T->A	p.Ile336Lys	I336K	<5	-
c.2374C->T	p.Arg792X	R792X	<5	-
c.1766+5G->T		1898+5G->T	<5	-
c.1727G->C	p.Gly576Ala	G576A	<5	-
c.3882_3885delTATT	p.Ile1295PhefsX32	4010del4	<5	-
c.3908delA	p.Asn1303ThrfsX25	4040delA	<5	-
c.4077_4080delT GTTinsAA	p.Val1360delfsX?	4209TGTT->AA	<5	-
c.3017C->A	p.Ala1006Glu	A1006E	<5	-
c.3266G->A	p.Trp1089X	W1089X	<5	-
c.1679+1.6kbA->G		1811+1.6kbA->G	<5	-
c.4111G->T	p.Glu1371X	E1371X	<5	-
c.3872A->G	p.Gln1291Arg	Q1291R	<5	-
c.1477_1478delCA	p.Gln493ValfsX10	1609delCA	<5	-
c.(273+1_274-1)_(1679+1_1680-1)del		CFTRdele4-11	<5	-

Nucleotide	Protein	Legacy name	N	%
c.442delA	p.Ile148LeufsX5	574delA	<5	-
c.2195T->G	p.Leu732X	L732X	<5	-
c.[1210–12[5];1210-34TG[11]]		5T;TG11	<5	-
c.220C->T	p.Arg74Trp	R74W	<5	-
c.1327G->T	p.Asp443Tyr	D443Y	<5	-
c.3310G->T	p.Glu1104X	E1104X	<5	-
c.1766+1G->C		1898+1G->C	<5	-
c.3752G->A	p.Ser1251Asn	S1251N	<5	-
c.2855T->C	p.Met952Thr	M952T	<5	-
c.2668C->T	p.Gln890X	Q890X	<5	-
c.79G->T	p.Gly27X	G27X	<5	-
c.933_935delCTT	p.Phe312del	F311del	<5	-
c.3230T->C	p.Leu1077Pro	L1077P	<5	-
c.1A->G	p.Met1Val	M1V	<5	-
c.3158C->T	p.Thr1053Ile	T1053I	<5	-
c.1209+1G->A		1341+1G->A	<5	-
c.(53+1_54-1)_(164+1_165-1)del		CFTRdele2	<5	-
c.3011_3019del CTATAGCAG or c.3009_3017del AGCTATAGC	p.Ala1004_Ala1006del	3143del9	<5	-
c.413_415dupTAC	p.Leu138dup	L138ins	<5	-
c.3476C->T	p.Ser1159Phe	S1159F	<5	-
c.[1523T->G;3752G->A]	p.[Phe508Cys;Ser1251Asn]	F508C;S1251N	<5	-
c.274-2A->G		406-2A->G	<5	-
c.1573C->T	p.Gln525X	Q525X	<5	-
c.1240C->T	p.Gln414X	Q414X	<5	-
c.3717+5G->A		3849+5G->A	<5	-
c.2645G->A	p.Trp882X	W882X	<5	-
c.1418delG	p.Gly473GlufsX54	1548delG	<5	-
c.1117-1G>A		1249-1G->A	<5	-
c.1081delT	p.Trp361GlyfsX8	1213delT	<5	-
c.3475T->C	p.Ser1159Pro	S1159P	<5	-
c.571T->G	p.Phe191Val	F191V	<5	-
c.164+1G>A		296+1G->A	<5	-
c.1801A->T	p.Ile601Phe	I601F	<5	-
c.4231C->T	p.Gln1411X	Q1411X	<5	-
c.(53+1_54-1)_(489+1_490-1)del		CFTRdele2-4	<5	-
c.3209G->A	p.Arg1070Gln	R1070Q	<5	-
c.3458T->A	p.Val1153Glu	V1153E	<5	-
c.3435G->A	p.Trp1145X	W1145X	<5	-
c.2930C->T	p.Ser977Phe	S977F	<5	-
c.3717G->A		3849G->A	<5	-
c.1210–12[9]		9T	<5	-
c.3181G->C	p.Gly1061Arg	G1061R	<5	-
c.470_483del14	p.Phe157X	602del14	<5	-

Nucleotide	Protein	Legacy name	N	%
c.2739T->A	p.Tyr913X	Y913X	<5	-
c.137C->A	p.Ala46Asp	A46D	<5	-
c.717delG	p.Leu240X	849delG	<5	-
c.263T>A or c.263T>G	p.Leu88X	L88X	<5	-
c.1703delT	p.Leu568CysfsX4	1833delT	<5	-
c.613C->T	p.Pro205Ser	P205S	<5	-
c.1021T->C	p.Ser341Pro	S341P	<5	-
c.2735C->A	p.Ser912X	S912X	<5	-
c.1687T->G	p.Tyr563Asp	Y563D	<5	-
c.3773_3774insT	p.Leu1258PhefsX7	3905insT	<5	-
c.2859_2890del ACATTCTGTTCTT CAAGCACCTATGTCAACCC	p.Leu953PhefsX11	2991del32	<5	-
c.3302T->G	p.Met1101Arg	M1101R	<5	-
c.53+1G->T		185+1G->T	<5	-
c.1654C->T	p.Gln552X	Q552X	<5	-
c.1135G->T	p.Glu379X	E379X	<5	-
c.233dupT	p.Trp79LeufsX32	365-366insT	<5	-
c.3745G->A	p.Gly1249Arg	G1249R	<5	-
c.987delA	p.Gly330GlufsX39	1119delA	<5	-
c.859_863delAACTT	p.Asn287LysfsX19	991del5	<5	-
c.1037T->C	p.Leu346Pro	L346P	<5	-
c.273+1G->A		405+1G->A	<5	-
c.2989-1G->A		3121-1G->A	<5	-
c.3718-3T->G		3850-3T->G	<5	-
c.2421A->G	p.Ile807Met	I807M	<5	-
c.1837G->A	p.Ala613Thr	A613T	<5	-
c.1882G->C or c.1882G->A	p.Gly628Arg	G628R	<5	-
c.11C>A	p.Ser4X	S4X	<5	-
c.3194T->C	p.Leu1065Pro	L1065P	<5	-
c.1682C->A	p.Ala561Glu	A561E	<5	-
c.3700A->G	p.Ile1234Val	I1234V	<5	-
c.3873+2T->C		4005+2T->C	<5	-
c.50delT	p.Phe17SerfsX8	182delT	<5	-
c.2620-26A->G		2752-26A->G	<5	-
c.1013C->T	p.Thr338Ile	T338I	<5	-
'Other' selected			734	6.7

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