Cystic Fibrosis strength in numbers

UK Cystic Fibrosis Registry
2020 Annual Data Report

Published December 2021

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

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The UK CF Registry Steering Committee

Acknowledgements

First and foremost, the UK Cystic Fibrosis Registry team would like to thank people with cystic fibrosis and their families for their support, as well as anyone who has generously donated to the 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.

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Foreword

This report covers a year of challenge and change for people with cystic fibrosis, dominated by the impact of the COVID-19 pandemic and the initial stages of the roll-out of Kaftrio.

In this different year, the way in which people with CF used health services changed, with fewer in-person clinic attendances and traditional annual reviews recorded than normal (2020 91.6%, 2019 96%). As members of clinical teams were redeployed to the front line of the fight against COVID-19 we saw access to inpatient NHS services fall by around 40% across the year and the proportion of virtual appointments recorded jump significantly (2020 42%, 2019 4%).

It was also a year of change for the UK CF Registry, with more frequent data collection to support the reporting of the effectiveness of new medicines. CF teams entered a mammoth 25,000 encounters onto the system, including over 8,000 Annual Review datasets. This is an incredible achievement, and I would like to publicly thank them.

There has though been some inevitable disruption to the collection and entry of data, which means some things look different to normal and need to be interpreted carefully. We have added guidance on how to do this.

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 to ensure the best health outcomes for everyone.

It also means it has been able to play a central role in monitoring the impact of COVID-19, receiving reports of infections and recording vaccinations. As I write, we have recorded over 500 cases of people in the UK with CF having COVID-19. Fortunately, many have had mild or no symptoms: however, around 15% of people have been admitted to hospital and we have sadly had reports of four people who have died because of COVID-19.

The power of the Registry has also been shown in the progress of the milestone clinical trial, CF STORM. The focus is on the safety and impact of stopping some medicines that may not be needed by people when they are taking highly effective modulators.

I hope you enjoy reading about the insights that this report brings. 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 during a truly challenging year to make this report possible.

We also want your feedback on the work we do, including this report. Please get in touch with us on social media or by emailing registry@cysticfibrosis.org.uk to let us know your comments and questions.

David Ramsden
Chief Executive

Executive summary

As everyone knows 2020 was a strange year for all and that includes people with cystic fibrosis and the centres looking after them. We are incredibly grateful to the CF centres teams and the people with CF who consent to having their data captured in the registry, which continues to form such a rich source of information.

This foreword normally highlights changes in clinical outcomes over the years and new variables that I feel the reader might find of interest. However, as discussed in the other summaries, some of the data collection has been hard this year and we have needed to make some changes to analysis methods.

Areas to draw some attention to are:

- A slight drop in the number designated as completing an annual review to 91.6%, but this still represents 9922 people contributing to the outcome data in this report.
- The Best FEV1 percent predicted for the population remains stable at 81% (section 1.14).
- The amended 2019 figure for those newly diagnosed with CF seems to be lower at 244 than previous years, which have ranged between 270 and 300.
- There are now 145 people over the age of 60 living with CF in the UK.
- Intermittent growth of Pseudomonas aeruginosa in 17.8% of adults is consistent with previous years but because the Registry definition for chronic growth is 3 positives in a year and many centres may not have been able to capture that many samples during early phases of the pandemic, we think the 31.9% described in the adult group is probably falsely low.
- The drop in reported lung transplants this year with only 12 people reported as receiving one. It may be some of this tail off in transplants is due to the availability of the Elexacaftor/tezacaftor/ivacaftor (Kaftrio) modulator, initially becoming available for people with severe disease on a managed access agreement followed by a UK wide roll out in late 2020.
- 3892 people (39.2%) received at least once course of iv antibiotics. There has been a trend in reduction in this over the last 10 years, but the larger drop of 5.3% this year is likely to be influenced by lockdowns with less respiratory infections circulating to precipitate some of the exacerbations.
- Predicted survival at 50.6 years (section 1.39) for those born today, has passed the 50-year mark for the first time and appears to now show a progressive improvement again.

We hope that you find this years report informative and we look forward to the results for 2021.

Dr Siobhán B Carr
Chair of the UK CF Registry
Steering Committee

How the pandemic affected data entry in 2020

We think it's important to show as much data as possible from 2020 about clinical care and health outcomes over time, as well as a snapshot of how things were in that year. However, data entry into the Registry was affected by the COVID-19 pandemic in five key ways:

1. Fewer face-to-face appointments
2. Less testing of lung function and for lung infection
3. CF team members were diverted to COVID-19 care
4. More virtual appointments
5. More home spirometry equipment available to people with CF

This means it's hard for us to tell whether any changes we see in the 2020 data are:

- 'Real' (associated with the health and treatment of people with CF)
- Caused by one or more of the five 'Pandemic factors'

…or a mixture of the two. Because of this, it is very important that results from 2020 are interpreted with caution.In the next year or two it will become clearer whether 2020 represents an unusual single year, or a new normal forpeople with cystic fibrosis. We have marked results that are at particular risk of being distorted because of thepandemic with this symbol:

Results for outcomes like lung function, complication rates, and infection prevalence may be different to normal because:

- There was less data available, and clinical teams had less time to enter it onto the system. This is a trend we have seen reflected in CF Registries around the world.
- For extended periods people with CF were advised to shield, and not leave their homes. Clinical teams who care for people with cystic fibrosis were also diverted to frontline COVID-19 care. This meant that in-person appointments, and the clinical measurements that would happen during then, took place less often than normal.
- Not everyone with CF had access to spirometry equipment at home. Although funding was provided to support this, those that did get access to this equipment may not have done until after their 2020 annual review.~We aren't sure if measurements taken at home are equivalent to those taken in clinic.
- Most CF centres didn't measure lung function in hospital during the pandemic because it's an aerosol generating procedure and might have spread COVID-19.

We helped CF centres as much as we could to represent a true picture of the health of their patients during 2020. This included reassigning some outpatient clinic appointments as an annual review, so that they could be included in this report. Where key data was not available at Annual review we used information provided at an encounter where it could sensibly be used as a substitute.

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 **www.cysticfibrosis.org.uk/registry**.

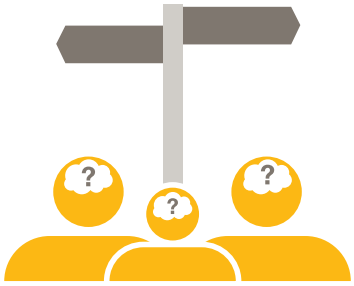
Cystic fibrosis

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 **www.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:

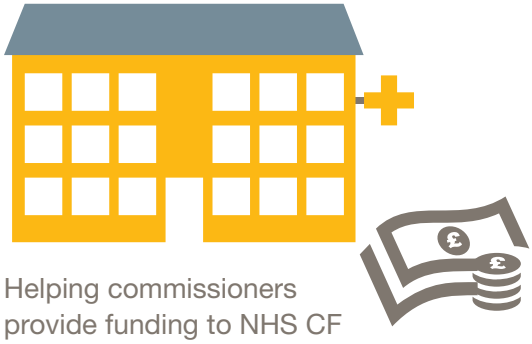


Helping people with CF and their families understand CF, and make informed decisions.

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 **www.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

	2015	2016	2017	2018	2019	2020
CF patients registered[1] Excluding diagnoses that year	10810 10586	10461 10214	10469 10255	10509 10287	10655 10462	10837 10632
CF patients with an annual review; n (%) [2]	9587 (91)	9695 (95)	9887 (96)	9847 (96)	10070 (96)	9922 (92)
Age in years; median[3]	19	20	20	20	21	21
All newly diagnosed patients (newborn screening [NBS] and other)[4]	224	247	214	222	193	205
All newly diagnosed patients (amended)[5]	(300)	(303)	(270)	(278)	(244)	(TBD)
Number of patients born identified by NBS[4]	168	216	192	167	150	152
Age at diagnosis in months; median[3]	2	2	2	2	2	2
Adults aged 16 years and over; %[3]	59.9	60.4	60.6	60.4	60.6	60.6
Males; %[3]	53	53.2	53.3	53	53.2	53.1
Genotyped; %[3]	98.1	98.4	99.3*	99.1	99.2	99.2
Total deaths reported during annual review year; n (%)[6]	125 (1.2)	148 (1.5)	132 (1.3)	137 (1.3)	114 (1.1)	97 (0.9)
Total deaths reported amended; n (%)[5]	135 (1.2)	159 (1.5)	143 (1.4)	143 (1.4)	118 (1.1)	(TBD)
Age at death in years; median (95% CI)[6]	28 (27, 33)	31 (29, 33)	31(29, 35)	32 (29, 35)	31 (29, 34)	36 (32, 38)

Annual Review: A Registry annual review form contains 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 2017 report.

[1] Number of patients diagnosed with CF, seen in the last two years, and alive at 1 January in the given year. This number reduced in 2016 as a result of a data cleaning exercise. We followed up on patients who were registered but did not have data submitted in 2016. If they were no longer being cared for within the NHS (*eg* they had moved abroad), they were marked as 'inactive' and excluded from this number.

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

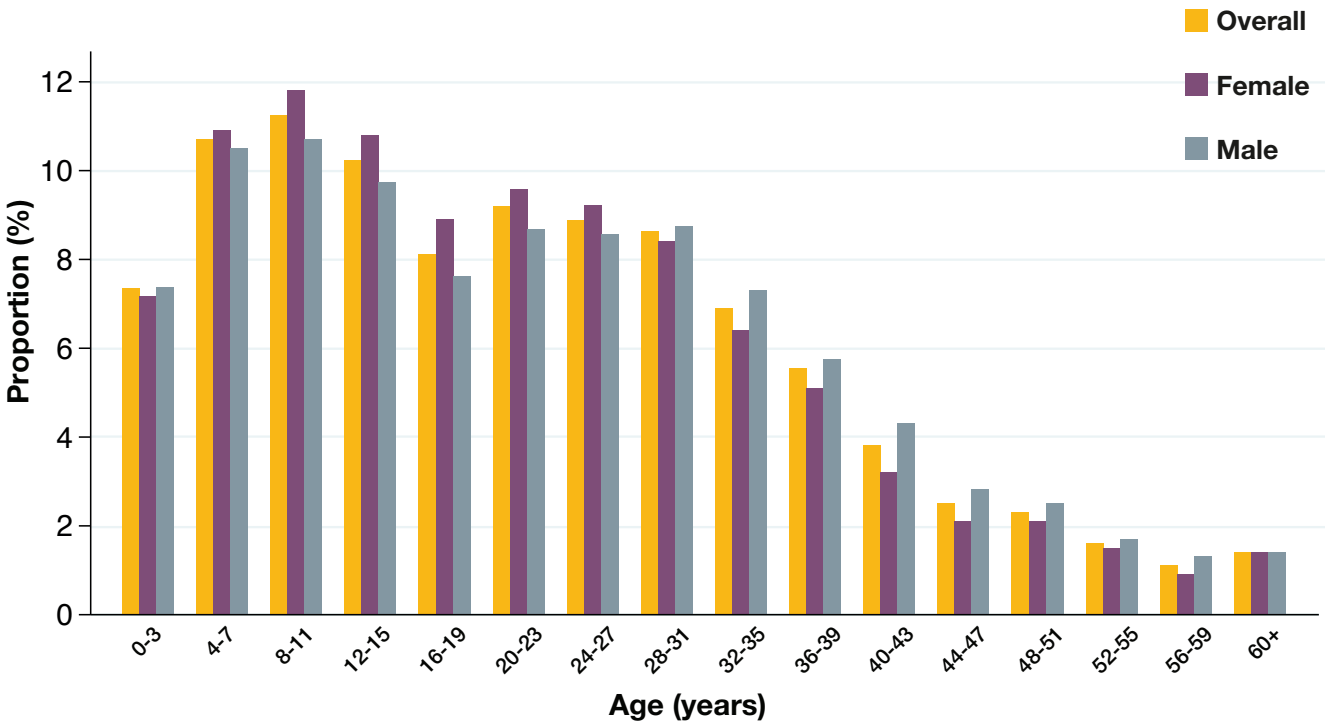
[3] Calculated from patients with an annual review in the given year (see footnote 5 below).

[4] Calculated from all patients registered on the database. Some diagnosis data are added after the data entry closure each year, so figures from previous years have been updated for this report.

[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.

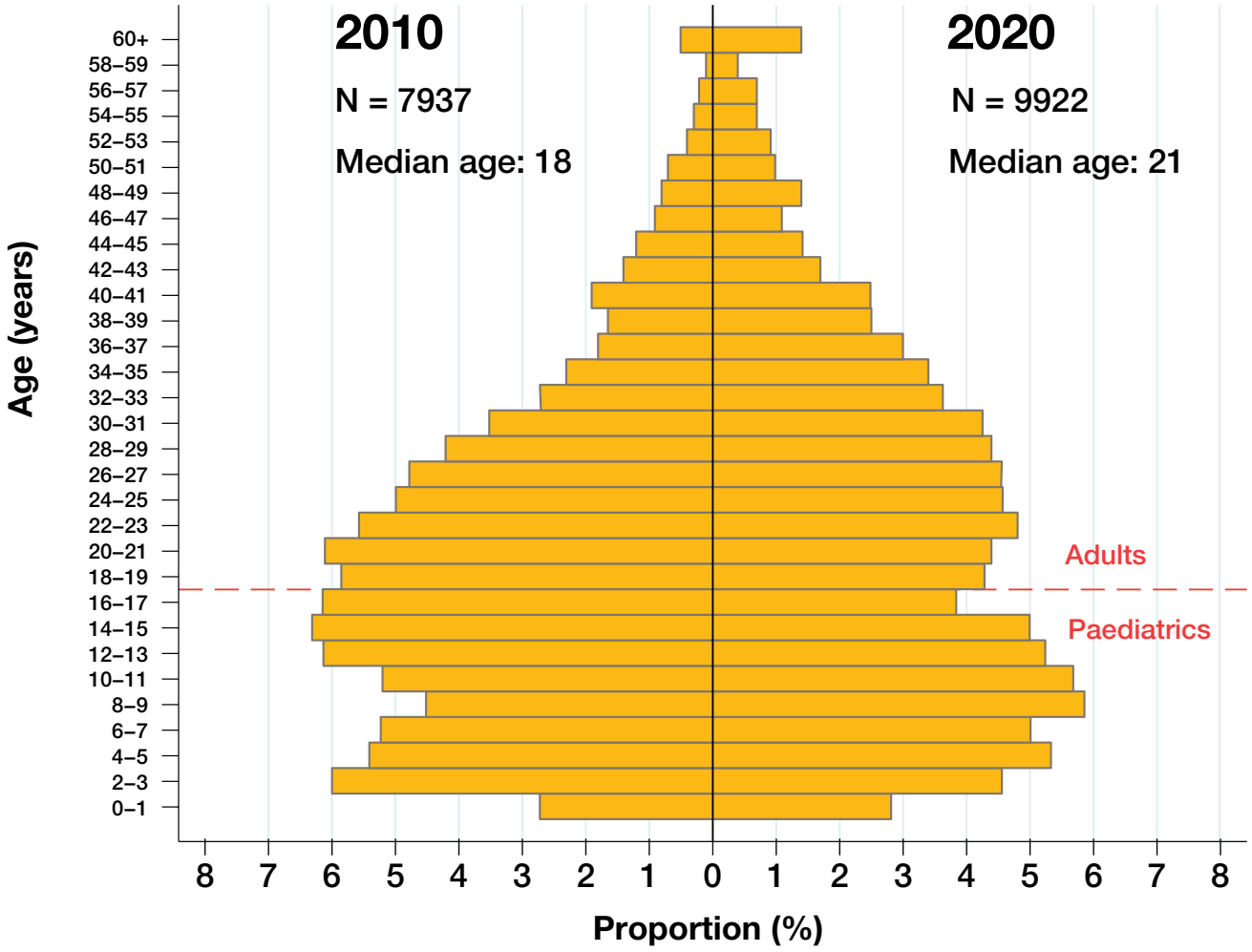
[6] Calculated from all registered patients who died in the given year.

1.2 Age distribution by sex
N=9922



Age	All; n (%)	Female; n (%)	Male; n (%)
0-3	730 (7.4)	339 (7.3)	391 (7.4)
4-7	1027 (10.4)	489 (10.5)	538 (10.2)
8-11	1147 (11.6)	554 (11.9)	593 (11.3)
12-15	1006 (10.1)	490 (10.5)	516 (9.8)
16-19	808 (8.1)	410 (8.8)	398 (7.6)
20-23	910 (9.2)	441 (9.5)	469 (8.9)
24-27	901 (9.1)	439 (9.4)	462 (8.8)
28-31	844 (8.5)	388 (8.3)	456 (8.7)
32-35	683 (6.9)	303 (6.5)	380 (7.2)
36-39	544 (5.5)	244 (5.2)	300 (5.7)
40-43	410 (4.1)	167 (3.6)	243 (4.6)
44-47	248 (2.5)	96 (2.1)	152 (2.9)
48-51	244 (2.5)	108 (2.3)	136 (2.6)
52-55	159 (1.6)	71 (1.5)	88 (1.7)
56-59	116 (1.2)	45 (1.0)	71 (1.3)
60+	145 (1.5)	68 (1.5)	77 (1.5)
<16	3910 (39.4)	1872 (40.2)	2038 (38.7)
≥16	6012 (60.6)	2780 (59.8)	3232 (61.3)
<18	4288 (43.2)	2070 (44.5)	2218 (42.1)
≥18	5634 (56.8)	2582 (55.5)	3052 (57.9)
Overall	**9922**	**4652**	**5270**

1.3 Age distribution of the UK CF population in 2010 vs 2020



Note the different demographic distribution across the years, namely higher proportion in older age groups in 2020.)

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

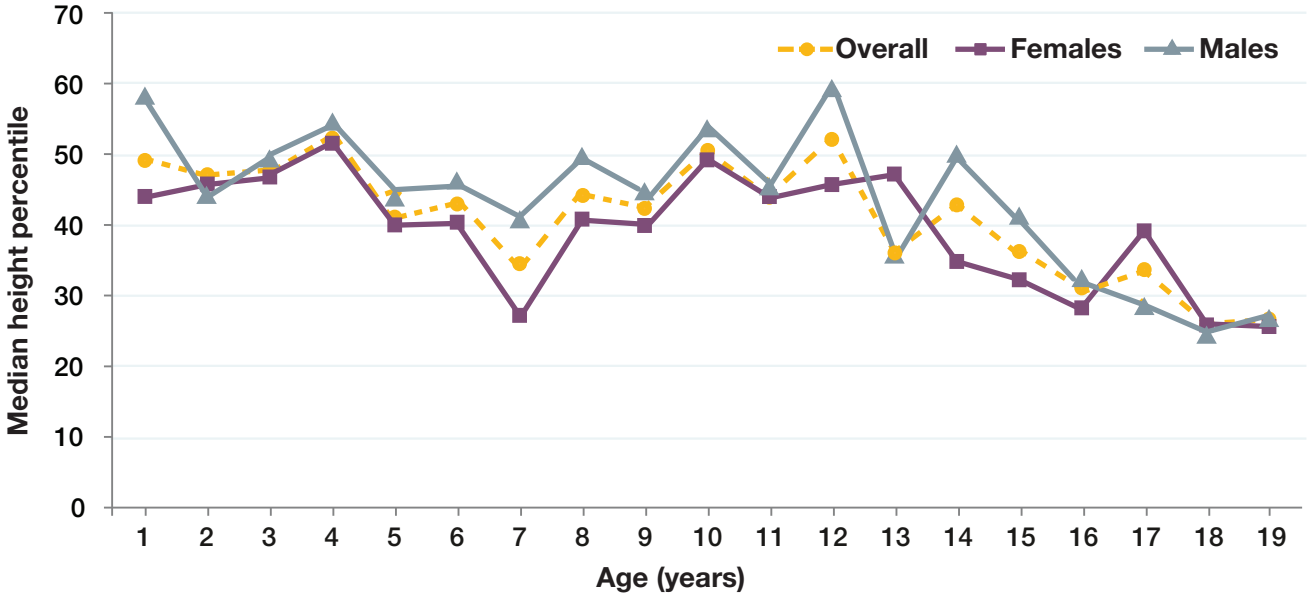
1.4 Ethnicity

	2010	2015	2020
Total	7937	9587	9922
White			
White (all); n (%)	7560 (95.3)	9081 (94.7)	9189 (92.6)
Asian			
Bangladeshi; n (%)	26 (0.3)	32 (0.3)	38 (0.4)
Indian; n (%)	23 (0.3)	35 (0.4)	48 (0.5)
Pakistani; n (%)	125 (1.6)	151 (1.6)	182 (1.8)
Other (Asian); n (%)	22 (0.3)	22 (0.2)	33 (0.3)
Black			
Black African; n (%)	11 (0.1)	13 (0.1)	12 (0.1)
Black Caribbean; n (%)	14 (0.2)	13 (0.1)	12 (0.1)
Other (Black); n (%)	<5	<5	<5
Mixed*			
Mixed (all); n (%)	40 (0.5)	81 (0.8)	63 (0.6)
Mixed (White-Asian); n (%)	-	-	13 (0.1)
Mixed (White-Black African); n (%)	-	-	9 (0.1)
Mixed (White-Black Caribbean); n (%)	-	-	22 (0.2)
Other (mixed); n (%)	-	-	19 (0.2)
Other/Unknown			
Other; n (%)	71 (0.9)	105 (1.1)	98 (1.0)
Unknown; n (%)	41 (0.5)	52 (0.5)	243 (2.4)

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

1.5 Height percentiles of children and young people (<20 years)[1] N=4718

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.

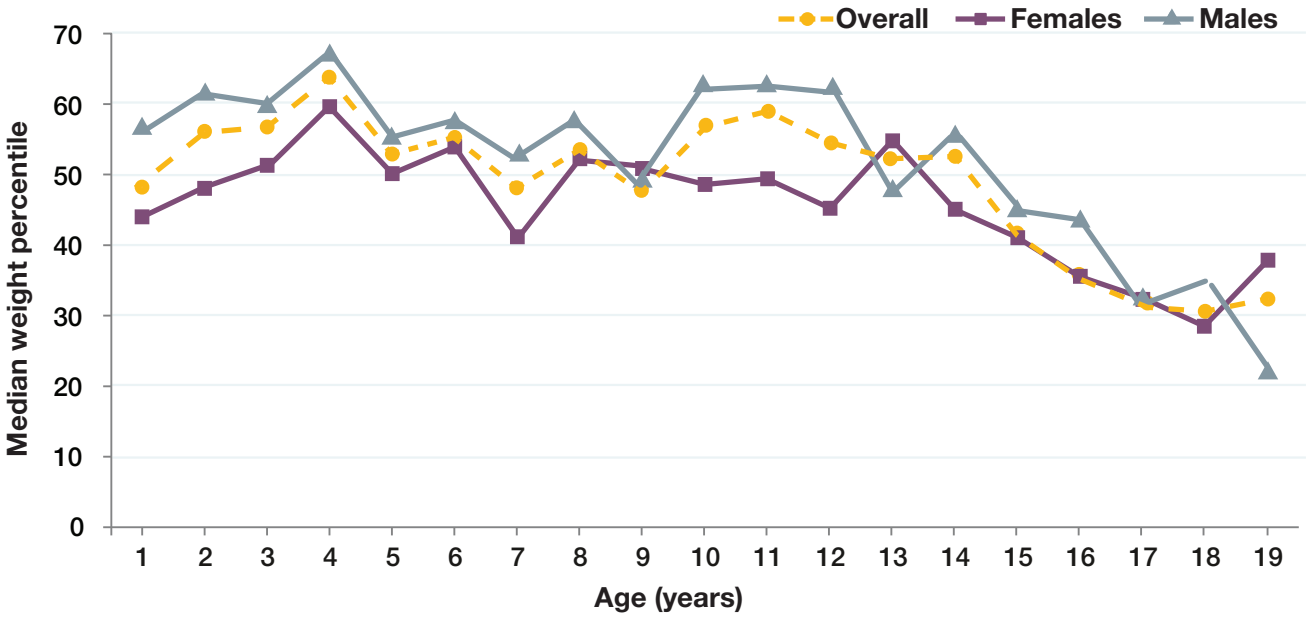


	Overall			Female			Male		
Age	n	Median	IQR	n	Median	IQR	n	Median	IQR
1	178	49.2	20.0-78.9	87	45.2	17.0-75.3	91	57.2	23.9-84.0
2	168	46.9	27.1-76.2	78	48.2	27.5-70.5	90	45.5	26.9-81.8
3	172	48.3	21.4-75.2	84	46.7	19.6-71.2	88	48.5	23.2-77.8
4	194	53.7	23.1-71.8	96	52.2	23.5-71.3	98	55.1	22.4-71.8
5	216	40.7	18.0-67.9	104	39.5	15.5-69.2	112	43.6	22.2-67.4
6	196	44.4	19.0-67.8	94	40.2	14.2-70.7	102	45.7	22.1-67.6
7	207	35.6	16.0-67.9	106	28.0	10.5-63.1	101	39.5	18.9-68.6
8	254	45.5	20.4-69.9	124	41.0	21.1-68.3	130	48.9	18.3-70.8
9	251	43.0	18.1-71.2	131	40.5	15.4-72.9	120	44.0	21.3-69.0
10	244	52.1	24.9-72.3	113	50.6	22.2-70.7	131	53.8	25.1-74.6
11	227	46.0	23.1-74.4	116	44.7	24.4-74.7	111	46.0	22.1-74.4
12	209	51.9	23.5-74.9	101	46.3	21.2-69.6	108	58.8	28.4-78.9
13	224	42.7	17.0-69.4	107	47.5	22.4-74.8	117	36.2	15.2-66.4
14	204	43.7	22.0-71.0	105	36.3	15.1-62.7	99	50.6	28.0-77.8
15	211	35.4	18.3-61.7	99	32.8	17.9-61.7	112	41.2	19.9-65.2
16	178	31.4	10.6-56.2	93	28.8	9.9-56.7	85	32.4	10.9-55.8
17	151	33.5	14.1-59.6	81	39.8	20.8-59.6	70	28.0	10.8-57.4
18	184	27.4	10.4-54.5	93	27.7	11.9-62.5	91	25.1	7.5-53.0
19	200	27.4	10.3-54.9	99	27.4	13.6-58.9	101	26.9	9.1-54.0
Overall	**3868***	**41.6**	**18.3-69.5**	**1911**	**40.0**	**17.6-68.2**	**1957**	**43.8**	**19.0-71.1**

* 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=4718

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.

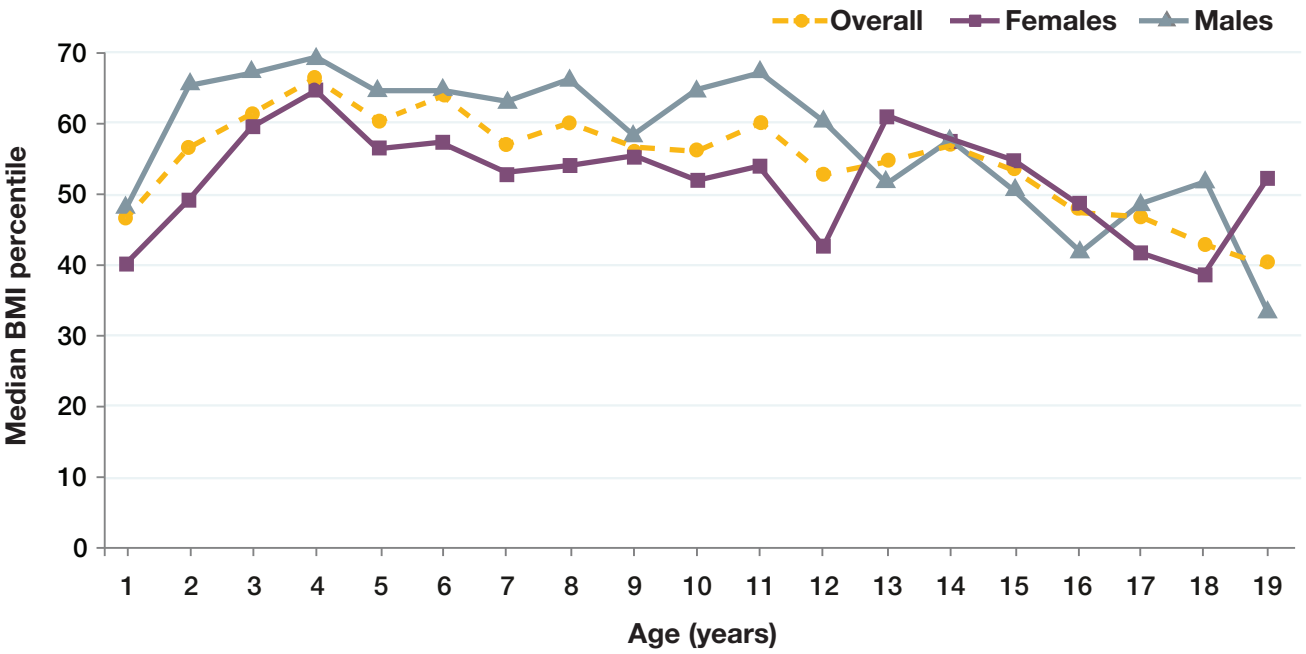


	Overall			Female			Male		
Age	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**
1	198	48.9	24.0-75.6	91	44.0	22.1-73.5	107	56.4	24.0-77.5
2	191	55.9	32.1-79.8	87	47.8	25.9-77.1	104	61.5	34.9-80.8
3	193	56.7	27.4-79.9	93	50.8	27.4-77.6	100	59.9	28.1-85.4
4	208	63.2	38.2-84.5	104	59.6	39.2-83.4	104	67.1	35.3-85.1
5	236	54.8	29.7-76.2	113	50.9	30.0-72.7	123	56.2	28.2-79.2
6	211	57.2	29.7-79.1	101	55.8	28.5-79.1	110	58.0	30.7-78.6
7	220	47.9	22.5-76.5	108	40.8	19.8-74.2	112	54.1	29.8-77.3
8	265	54.9	27.7-77.0	129	52.5	26.2-68.9	136	59.0	27.7-82.8
9	263	50.0	26.7-76.0	135	51.6	24.7-76.9	128	49.2	27.4-74.3
10	257	57.7	32.2-81.8	117	49.3	29.6-81.1	140	62.5	34.0-83.2
11	238	59.0	31.1-81.0	118	50.4	25.3-80.4	120	62.8	35.1-81.9
12	217	54.6	29.7-80.3	106	47.2	20.6-72.1	111	62.6	31.8-84.8
13	234	51.8	22.0-79.0	110	55.0	24.2-82.5	124	49.2	19.8-76.8
14	209	51.7	25.0-74.8	106	45.5	24.2-72.7	103	56.1	29.3-75.0
15	224	43.5	28.8-67.8	105	42.9	29.2-65.1	119	45.7	27.4-69.6
16	183	37.1	13.1-72.2	96	35.8	12.6-73.8	87	42.7	13.1-69.2
17	151	32.7	13.7-58.6	80	32.0	13.4-59.4	71	32.7	14.0-58.6
18	179	30.9	6.1-61.9	90	28.6	9.1-59.0	89	34.9	4.3-66.1
19	192	32.3	6.6-65.4	96	38.7	15.6-66.7	96	21.5	3.9-63.6
Overall	**4069***	**50.5**	**24.2-77.0**	**1985**	**47.4**	**23.1-74.6**	**2084**	**53.9**	**25.5-78.8**

* 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=4718

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.

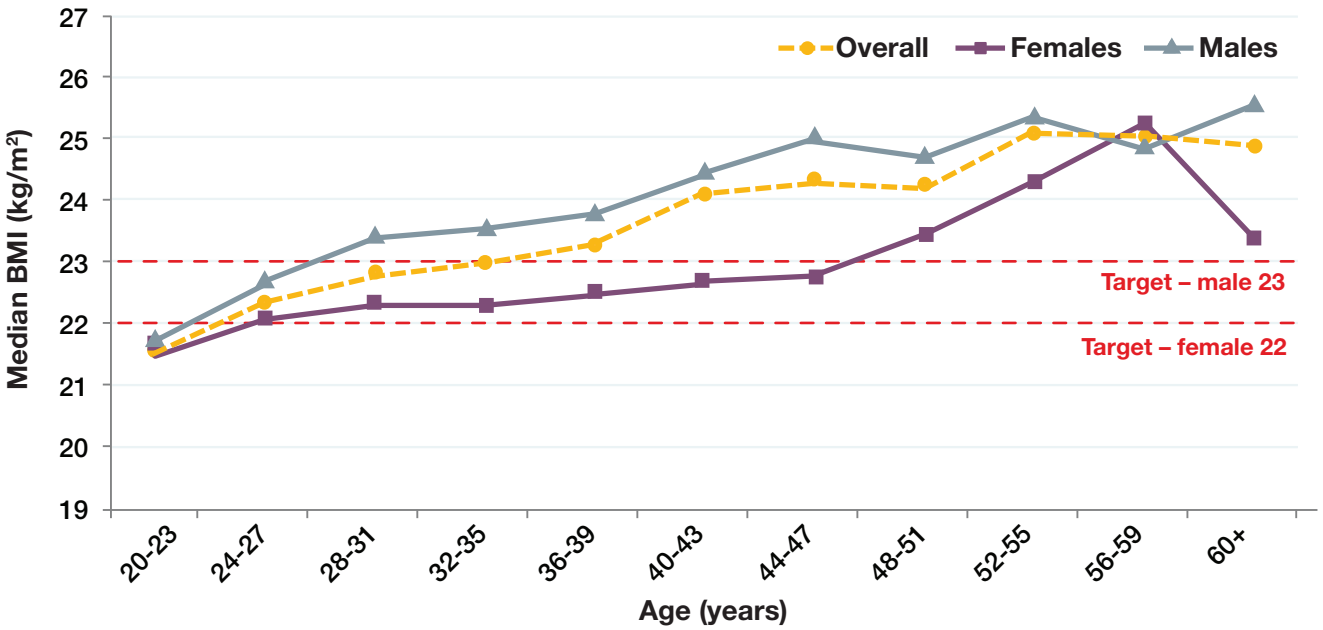


	Overall			Female			Male		
Age	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**
1	178	47.3	18.1-79.4	87	39.9	14.5-79.0	48.7	45.2	19.8-79.9
2	168	57.0	32.0-85.1	78	49.1	29.4-81.7	65.7	59.7	36.5-86.9
3	172	61.8	38.7-85.4	84	60.0	37.3-82.8	66.9	60.4	41.0-88.6
4	194	67.3	44.9-85.4	96	66.3	42.1-83.2	69.4	66.4	48.3-88.3
5	216	61.0	34.7-82.6	104	58.4	35.0-79.7	64.4	59.2	32.2-86.1
6	196	64.4	39.4-83.9	94	57.0	37.4-84.8	64.8	54.7	40.7-83.2
7	207	57.4	36.2-79.8	106	52.9	28.3-73.1	63.1	53.8	41.2-80.3
8	250	60.2	33.5-81.3	123	54.2	27.7-76.8	65.9	45.9	38.6-86.9
9	250	56.3	34.2-81.5	131	55.9	32.7-79.2	57.7	58.4	36.2-83.2
10	243	57.0	32.9-83.1	112	53.3	30.4-78.4	64.3	62.5	36.3-87.4
11	225	60.6	31.9-83.7	115	53.4	25.3-82.5	67.3	56.7	43.7-86.1
12	208	53.2	27.7-81.2	100	43.1	26.1-71.6	60.8	48.9	31.0-82.9
13	222	56.7	28.8-78.5	105	61.4	31.8-79.1	54.2	51.5	27.8-77.0
14	202	60.0	29.2-78.4	104	59.3	33.0-79.8	60.0	52.0	26.9-78.0
15	210	56.0	31.5-76.6	99	57.7	34.4-82.5	53.6	47.8	28.1-76.1
16	176	49.8	24.5-81.4	93	50.7	24.1-81.3	45.4	51.0	24.7-81.5
17	147	48.2	24.9-75.0	78	45.4	25.2-69.5	49.8	48.7	24.7-76.9
18	178	45.3	14.0-76.3	90	40.5	12.7-68.2	54.3	43.1	16.5-80.3
19	191	42.3	14.5-75.3	96	53.6	22.1-77.6	36.2	28.8	10.7-64.2
Overall	**3833***	**56.4**	**30.2-81.3**	**1895**	**54.1**	**29.2-79.0**	**58.9**	**53.2**	**31.4-83.1**

*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=5204

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	890	21.8	19.9-24.0	431	21.8	20.1-24.1	459	21.8	19.8-23.9
24-27	876	22.4	20.3-24.8	424	22.1	20.2-24.3	452	22.7	20.5-25.1
28-31	818	22.9	20.9-25.1	374	22.5	20.6-24.4	444	23.3	21.3-25.5
32-35	668	23.1	20.8-25.5	296	22.3	20.2-25.2	372	23.5	21.5-25.7
36-39	527	23.3	21.1-25.7	237	22.5	20.6-25.0	290	23.8	21.7-26.2
40-43	398	24.0	21.7-25.8	163	22.7	20.8-25.0	235	24.5	22.5-26.3
44-47	241	24.2	21.8-26.7	93	22.8	21.1-25.2	148	25.1	23.1-27.2
48-51	236	24.4	22.2-26.8	103	23.4	21.3-26.7	133	24.7	22.7-27.1
52-55	154	25.1	22.8-27.9	69	24.4	22.4-27.6	85	25.4	23.3-27.9
56-59	112	25.0	22.5-28.1	43	25.2	20.8-30.0	69	24.9	23.0-27.6
60+	141	24.8	22.1-27.8	67	24.3	20.6-27.8	74	25.5	23.0-28.1
Overall	**5061**	**23.0**	**20.9-25.4**	**2300**	**22.5**	**20.4-24.9**	**2761**	**23.5**	**21.3-25.8**

*number with non-missing data.

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

1.9 Education and employment in adults (16 years and over) N=6012

The following table shows how people with CF reported their education and employment status in 2020.

	2017	2018	2019	2020		
				Overall	Male	Female
Number of patients	5989	5952	6104	6012	3232	2780
Number who completed questionnaire; n (%)	5937 (99.1)	5950 (100.0)	6103 (100.0)	5968 (99.3)	3205 (99.2)	2763 (99.4)
Full-time employment; n (%)	1949 (32.5)	1956 (32.9)	2048 (33.6)	1975 (32.9)	1324 (41.0)	651 (23.4)
Part-time employment; n (%)	887 (14.8)	926 (15.6)	958 (15.7)	894 (14.9)	347 (10.7)	547 (19.7)
Student; n (%)	973 (16.2)	937 (15.7)	969 (15.9)	1015 (16.9)	492 (15.2)	523 (18.8)
Homemaker; n (%)	246 (4.1)	237 (4.0)	231 (3.8)	200 (3.3)	18 (0.6)	182 (6.5)
Unemployed; n (%)	837 (14.0)	814 (13.7)	825 (13.5)	847 (14.1)	486 (15.0)	361 (13.0)
Disabled; n (%)	352 (5.9)	359 (6.0)	327 (5.4)	274 (4.6)	139 (4.3)	135 (4.9)
Retired; n (%)	120 (2.0)	133 (2.2)	145 (2.4)	139 (2.3)	76 (2.4)	63 (2.3)
Volunteer; n(%)	-*	-*	8 (0.1)	-**	<5	7 (0.3)
Unknown entered; n (%)	573 (9.6)	588 (9.9)	592 (9.7)	613 (10.2)	319 (9.9)	294 (10.6)
In work or study; n (%)	**3809 (64.2)**	**3819 (64.2)**	**3975 (65.1)**	**3884 (65.1)**	**2163 (67.5)**	**1721 (62.3)**

1.10 Pregnancy

	2017	2018	2019	2020
Women with CF that had babies; n	58	65	58	56
Men with CF who became fathers; n	44	45	45	44

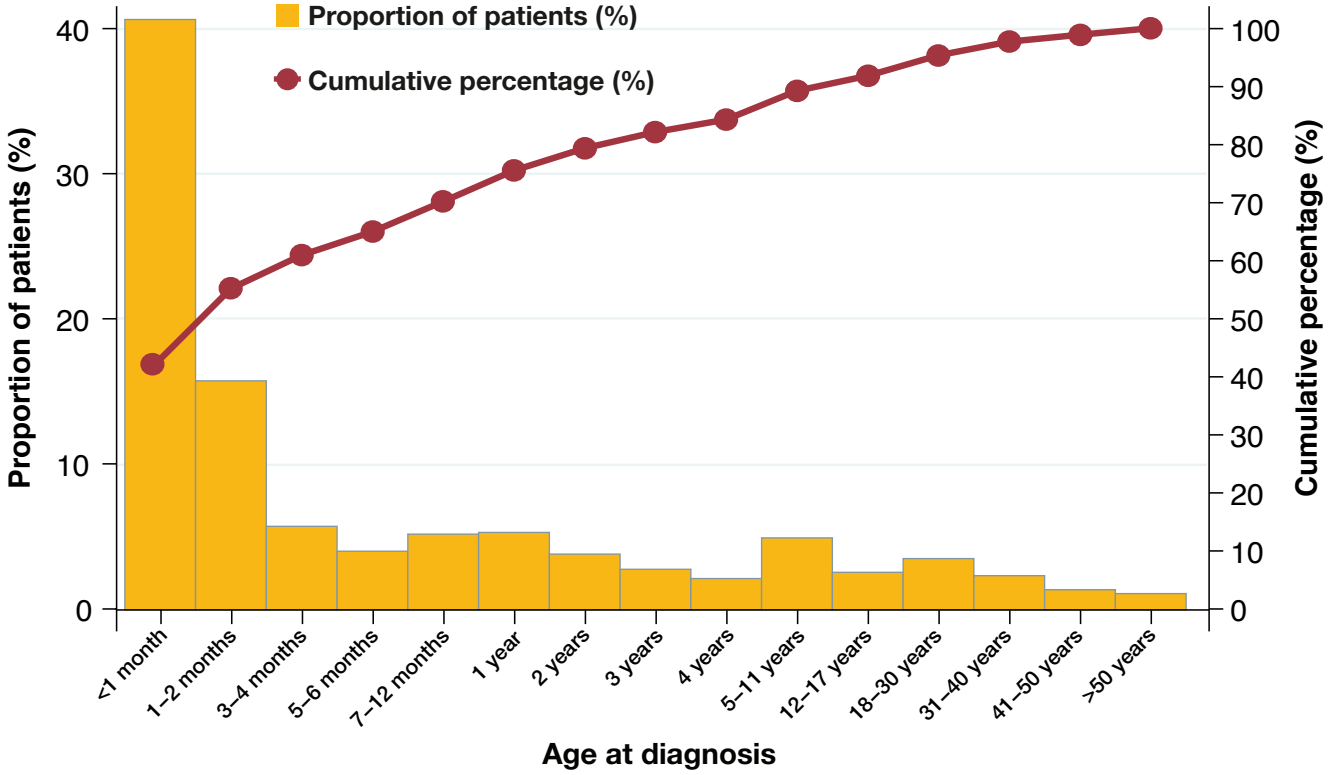
56 women with CF had babies in 2020

44 men with CF became fathers in 2020

*Newly added in 2019.
** Redacted to adhere to statistical disclosure guidelines.

Diagnosis of cystic fibrosis

1.11 Age at diagnosis in 2020
N=9922



The median age at diagnosis for patients aged under 16 in 2020 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 **152 (74.1%)** out of 205 patients born in 2020 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. Therefore the number of patients identified in 2020 is higher (150) in this report than was recorded in the previous.

857 (14.7%) of adults with CF in the Registry in 2020 were diagnosed at age 16 or over.

In 2020, **12** people aged 16 or over were newly diagnosed with cystic fibrosis.

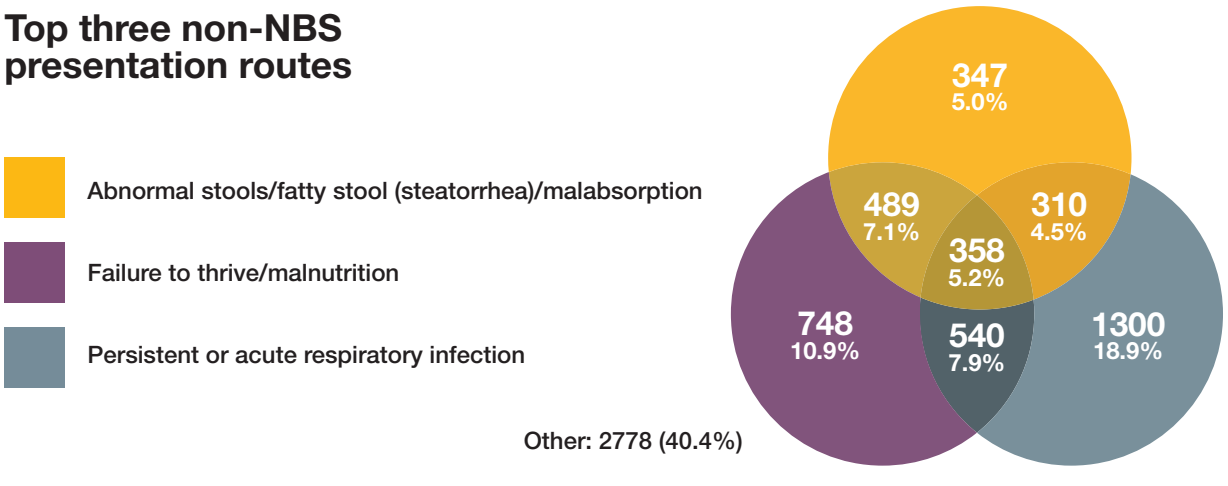
1.12 Mode of presentation

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	9922	9024	857
Number diagnosed by newborn screening	3011	3011	0
Total non-NBS	**6911**	**6013**	**857**

Mode of presentation (excluding NBS)	All patients ** (N=6870)	Age <16 at diagnosis (N=6013	Age >= 16 at diagnosis (N=857)
	n (%)	n(%)	n (%)
Persistent or acute respiratory infection	2508 (36.5)	2028 (33.7)	480 (56.0)
Failure to thrive/malnutrition	2135 (31.1)	2109 (35.1)	26 (3.0)
Abnormal stools/fatty stool(steatorrhea)/malabsorption	1504 (21.9)	1452 (24.1)	52 (6.1)
Meconium ileus	-***	1281 (21.3)	<5
Family history	902 (13.1)	787 (13.1)	115 (13.4)
Genotype	662 (9.6)	468 (7.8)	194 (22.6)
Unknown	310 (4.5)	263 (4.4)	47 (5.5)
Rectal prolapse	-***	238 (4.0)	<5
Nasal polyps	142 (2.1)	76 (1.3)	66 (7.7)
Bronchiectasis	82 (1.2)	8 (0.1)	74 (8.6)
Prenatal	77 (1.1)	77 (1.3)	0 (0.0)
Electrolyte imbalance	-***	59 (1.0)	<5
Liver disease	-***	41 (0.7)	<5
Fertility	-***	<5	40 (4.7)
Pancreatitis	20 (0.3)	6 (0.1)	14 (1.6)
Oedema	9 (0.1)	9 (0.1)	0 (0.0)

Top three non-NBS presentation routes



Other: 2778 (40.4%)

*Age-stratified figures are presented only for those with non-missing diagnosis date.
** Mode of presentation is described for people with a 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, gender, 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 FEV_1% predicted of 50% breathes out half the volume of air as a comparable person without cystic fibrosis.

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

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

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

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

1.13 Annual Review FEV_1% predicted (GLI equations) in patients aged six years and older who have not had a lung transplant N=8363

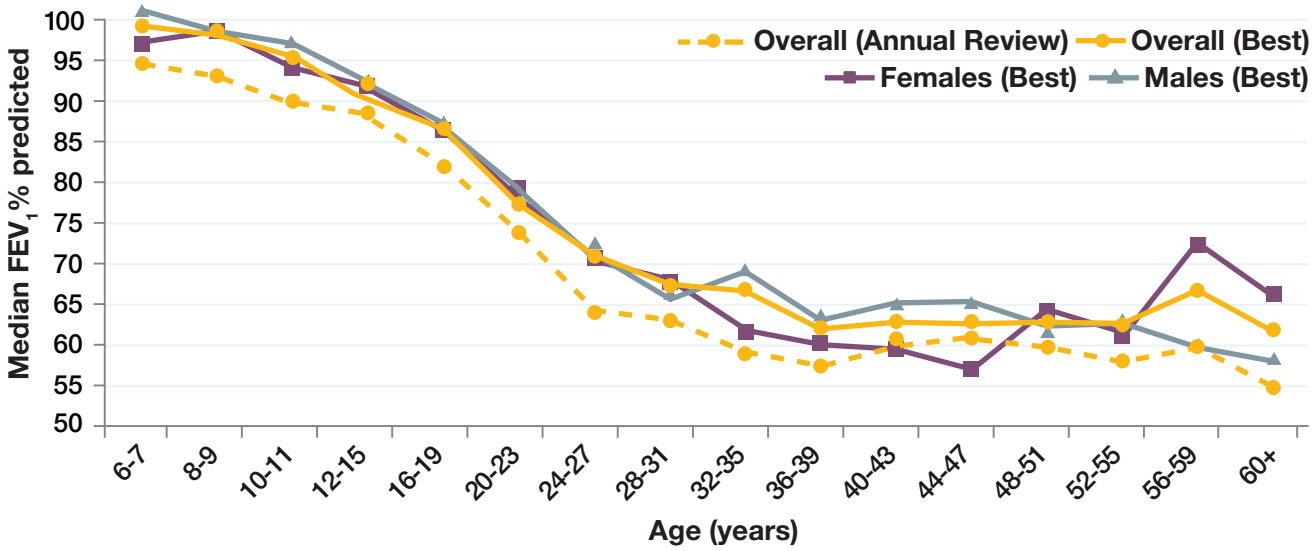
People with CF who have had lung transplants are excluded, as their new 'non-CF' lungs may have lung health similar to a person without cystic fibrosis.

For the best FEV_1 calculation, where best FEV_1% was missing or less than FEV_1% at annual review, the annual review FEV_1% was used.

	Overall			Female			Male		
Age (yrs)	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**
6-7	255	95.1	84.0-105.1	127	92.8	83.4-103.0	128	96.4	86.0-106.4
8-9	368	93.4	82.2-102.1	179	94.1	81.9-102.7	189	92.9	82.3-101.6
10-11	385	90.0	80.1-99.3	189	89.9	79.5-99.1	196	90.0	80.8-100.3
12-15	690	88.6	76.4-98.2	337	87.0	75.6-98.0	353	89.2	77.2-98.2
16-19	592	81.7	65.9-93.0	304	80.9	64.3-92.4	288	82.0	67.3-94.3
20-23	631	74.4	56.0-88.5	311	76.3	55.8-88.6	320	73.5	56.6-88.1
24-27	610	69.0	49.8-85.0	311	67.8	50.0-84.5	299	70.4	48.9-85.9
28-31	550	63.7	44.5-80.4	268	65.9	48.9-80.8	282	60.3	42.4-79.7
32-35	459	63.9	46.1-81.0	204	62.8	42.7-79.2	255	64.5	49.3-81.1
36-39	336	57.8	40.5-79.3	151	56.2	41.8-74.3	185	59.5	38.0-82.0
40-43	264	61.5	45.0-80.3	113	59.3	44.6-76.7	151	63.0	45.4-82.9
44-47	155	61.0	42.1-76.0	63	60.0	42.1-74.8	92	62.4	42.1-78.1
48-51	141	59.9	40.5-77.6	61	62.2	44.9-75.8	80	57.4	39.7-78.3
52-55	100	58.7	42.5-78.1	41	57.3	41.9-72.8	59	59.6	44.5-79.4
56-59	71	59.6	45.8-76.2	27	66.6	56.3-78.1	44	52.4	38.9-71.7
60+	91	55.1	38.4-76.4	44	56.5	41.7-76.9	47	52.8	35.4-76.4
<16	1698	90.8	79.7-100.2	832	90.3	78.6-99.7	866	91.3	80.5-100.4
≥16	4000	68.0	48.3-84.8	1898	68.3	49.3-84.1	2102	67.8	47.2-85.1
<18	1968	90.1	78.7-99.6	975	89.5	77.0-99.2	993	90.8	80.2-100.3
≥18	3730	66.9	47.1-83.3	1755	67.3	48.3-83.1	1975	66.4	46.0-83.7
Overall	**5698***	**76.9**	**55.8-92.0**	**2730**	**76.5**	**56.4-91.6**	**2968**	**77.2**	**55.0-92.3**

*number with non-missing data.

1.14 Best FEV_1% predicted (GLI equations) in patients aged six years and older who have not had a lung transplant
N=8363



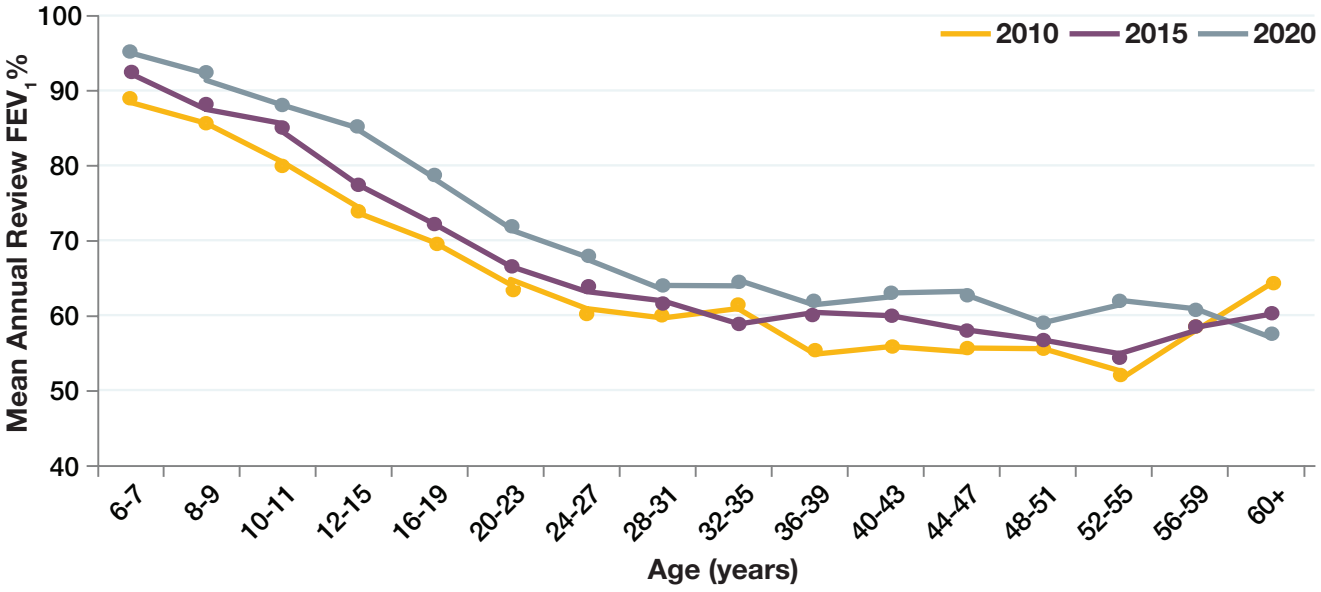
Where Best FEV_1% was missing or less than the FEV_1% at annual review, annual review FEV_1% was used instead.

	Overall			Female			Male		
Age (yrs)	n	Median	IQR	n	Median	IQR	n	Median	IQR
6-7	436	99.1	89.6-107.8	207	97.6	87.5-107.4	229	100.6	91.1-108.4
8-9	539	97.8	88.6-106.0	269	97.9	88.5-107.4	270	97.7	89.2-105.6
10-11	527	94.6	85.1-103.2	250	93.6	84.6-103.2	277	95.9	86.3-102.9
12-15	953	92.4	81.5-100.6	467	91.8	81.3-100.6	486	92.9	81.6-100.7
16-19	768	85.8	71.0-96.4	388	85.5	70.1-96.3	380	85.9	71.9-96.4
20-23	869	77.6	60.1-91.3	420	78.7	59.1-91.9	449	77.4	60.8-90.8
24-27	839	72.0	53.3-89.1	410	71.4	53.4-88.6	429	72.6	53.3-89.1
28-31	772	67.4	48.8-83.3	358	68.4	52.4-84.7	414	66.5	45.1-82.4
32-35	619	67.2	49.4-82.2	269	65.6	48.8-83.0	350	68.8	50.1-81.7
36-39	465	62.3	43.2-81.0	205	60.1	43.7-78.3	260	63.5	42.8-82.3
40-43	365	63.0	47.0-82.7	145	60.2	49.2-80.7	220	64.4	46.0-83.4
44-47	212	61.9	45.4-78.3	82	57.0	44.7-72.2	130	65.4	46.5-81.8
48-51	210	62.8	46.6-82.5	91	64.1	50.2-82.5	119	61.7	42.9-82.6
52-55	141	62.5	46.3-79.4	66	61.3	45.3-79.1	75	62.5	46.3-81.4
56-59	99	66.7	49.5-82.2	38	72.6	63.2-82.7	61	59.1	43.5-81.7
60+	130	62.1	43.4-82.9	62	66.1	47.5-85.5	68	58.7	38.4-79.9
<16	2455	95.5	84.6-103.6	1193	94.6	83.9-103.3	1262	96.0	85.5-103.9
≥16	5489	71.4	52.1-87.8	2534	71.3	52.7-88.4	2955	71.4	51.3-87.4
<18	2813	94.6	83.8-103.1	1381	93.6	82.8-102.9	1432	95.4	84.7-103.3
≥18	5131	70.0	50.9-86.4	2346	70.0	51.7-87.1	2785	70.0	50.1-86.0
Overall	**7944***	**81.0**	**60.3-95.8**	**3727**	**81.2**	**60.8-95.6**	**4217**	**80.8**	**59.8-95.8**

*number with non-missing data.

1.15 Annual Review FEV_1% predicted (GLI equations) over time in patients six years and older who have not had a lung transplant
N=8637 in 2020, N=7764 in 2015, N=6426 in 2010*

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 2020 compares to Registry data from 2010 and 2015.

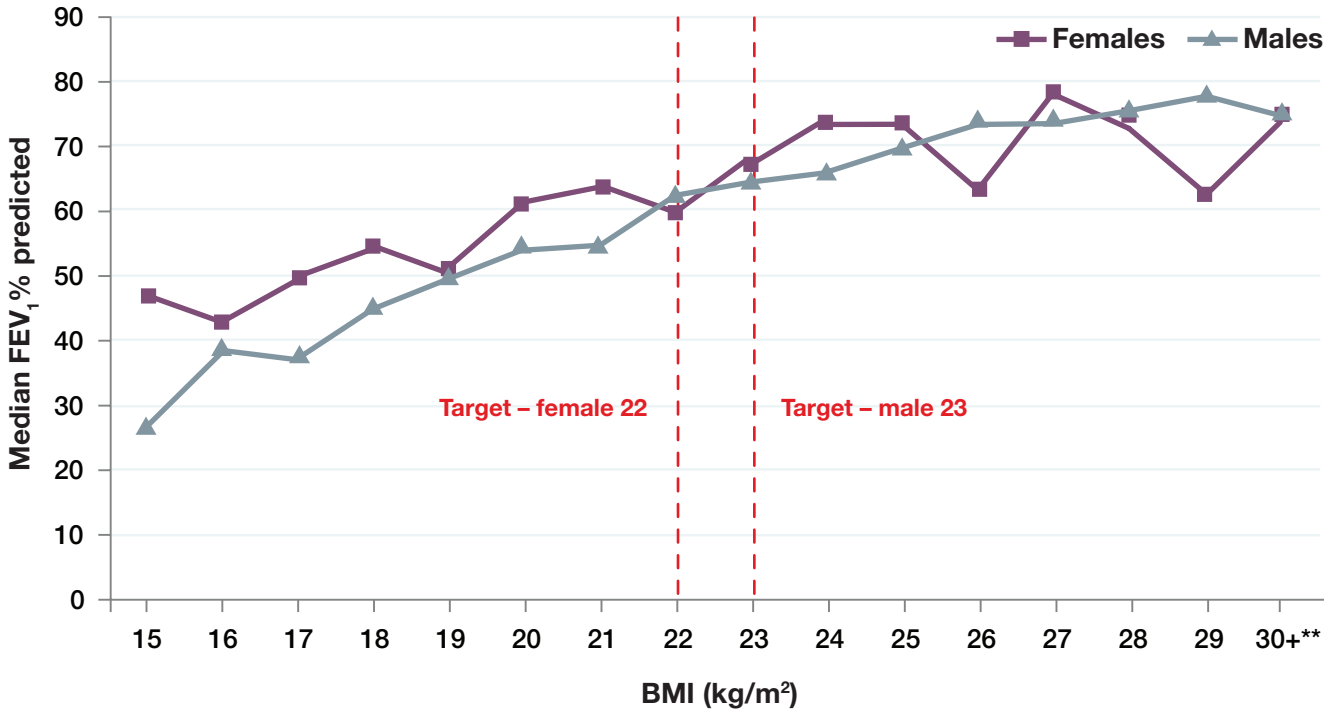


Age (years)	2010 mean FEV1%	2015 mean FEV1%	2020 mean FEV1%	p-values (t-test)**
6-7	89.5	91.7	94.4	0.033
8-9	85.2	88.3	92.1	0.001
10-11	80.0	84.9	88.6	<0.001
12-15	75.4	78.9	85.9	<0.001
16-19	70.0	72.6	78.9	<0.001
20-23	64.5	67.4	71.6	<0.001
24-27	60.5	63.7	68.0	<0.001
28-31	60.6	61.6	63.3	0.191
32-35	61.2	59.5	63.6	0.005
36-39	57.0	60.0	61.5	0.534
40-43	56.8	60.1	63.0	0.170
44-47	56.4	58.3	61.3	0.208
48-51	57.8	56.6	60.0	0.236
52-55	53.3	55.8	61.2	0.121
56-59	59.1	59.1	60.9	0.642
60+	64.5	60.1	58.9	0.742
<16	80.4	84.7	89.1	-
≥16	62.9	64.2	67.1	-
<18	79.2	82.8	88.2	-
≥18	61.6	63.2	66.0	-

*Due to missing data, means are calculated from a population of 8367 in 2020, 7764 in 2015 and 6426 in 2010.
** t-test comparing 2020 with 2015.

1.16 Annual Review FEV_1% predicted (GLI equations) and Body Mass Index (BMI) in people aged 20 years and over who have not had a lung transplant N=4970*

The goal BMI for adults is 22 for women, and 23 for men. The chart below shows the relationship between BMI and FEV_1% predicted. A healthy BMI can protect people with CF against lung infection, and help to preserve lung health. This chart excludes people who have had a lung transplant.

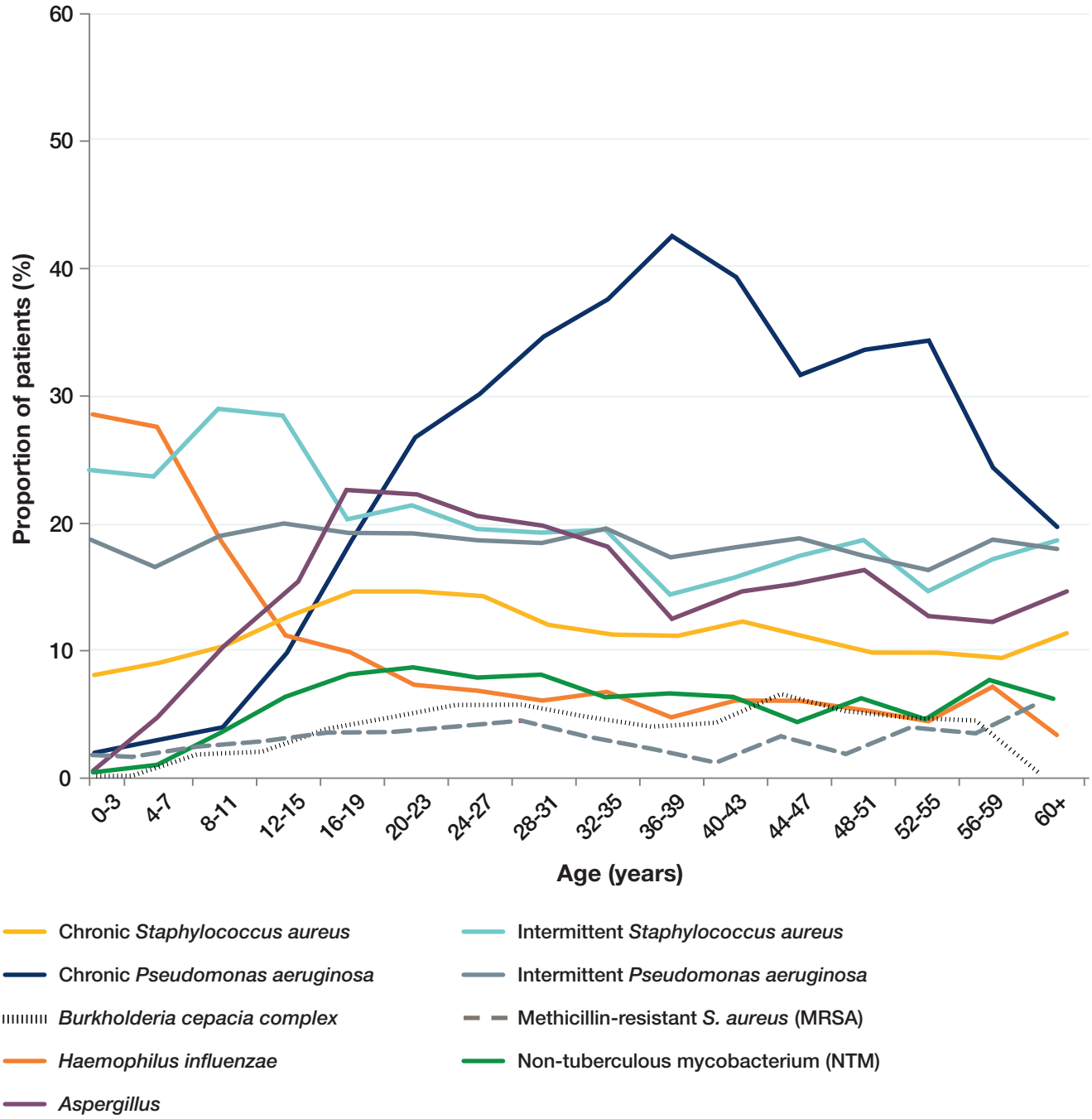


* Due to missing data, medians are calculated from a population of 4808.
** Each point represents the median FEV_1% predicted of patients for each given BMI value. Due to the wide range of BMIs in this population with a value of 30 of more, these are grouped into one.

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 2020
N=9922*



*Proportions are calculated from the number of patients with annual reviews, whereas previous reports based proportions on the number of patients who had a culture taken.

1.18 Lung infections in 2020
<16 years N=3910, ≥16 years N=6012

	Paediatric age range (years)				Overall
	0-3	4-7	8-11	12-15	Paediatric <16 years
Number in age range	730	1027	1147	1006	3910
Number who had culture taken*	721	1006	1130	990	3847
Chronic *Staphylococcus aureus*; n (%)	50 (6.8)	89 (8.7)	128 (11.2)	157 (15.6)	424 (10.8)
Intermittent *Staphylococcus aureus*; n (%)	177 (24.2)	243 (23.7)	341 (29.7)	293 (29.1)	1054 (27.0)
Chronic *Pseudomonas aeruginosa*; n (%)	13 (1.8)	29 (2.8)	44 (3.8)	99 (9.8)	185 (4.7)
Intermittent *Pseudomonas aeruginosa*; n (%)	131 (17.9)	142 (13.8)	212 (18.5)	205 (20.4)	690 (17.6)
Burkholderia cepacia complex; n (%)	<5	6 (0.6)	23 (2.0)	22 (2.2)	-**
B. cenocepacia; n (%)	<5	<5	5 (0.4)	<5	10 (0.3)
B. multivorans; n (%)	<5	<5	8 (0.7)	8 (0.8)	17 (0.4)
B. other cepacia; n (%)	<5	<5	<5	7 (0.7)	12 (0.3)
MRSA; n (%)	15 (2.1)	19 (1.9)	29 (2.5)	29 (2.9)	92 (2.4)
Haemophilus influenzae; n (%)	205 (28.1)	278 (27.1)	204 (17.8)	102 (10.1)	789 (20.2)
Non-tuberculous mycobacterium ; n (%)	<5	11 (1.1)	45 (3.9)	71 (7.1)	-**
Aspergillus; n (%)	5 (0.7)	41 (4.0)	128 (11.2)	158 (15.7)	332 (8.5)

Infections in this table reflect bugs grown in the 12 months prior to the 2020 annual review.
The UK CF Registry definition of 'chronic' is three or more isolates in the last 12 months.

* Proportions are calculated from number in age range, whereas prior to 2019 they were calculated from the number of people with CF who were recorded as having had a culture taken.
** Redacted to adhere to statistical disclosure guidelines.

	Adult age range (years)						Overall
	16-19	20-23	24-27	28-31	32-35	36-39	≥16 years
Number in age range	807	911	902	843	683	543	6012
Number who had culture taken*	764	819	797	731	606	474	5329
Chronic *S. aureus*; n (%)	156 (19.3)	176 (19.3)	168 (18.6)	121 (14.4)	88 (12.9)	69 (12.7)	941 (15.7)
Intermittent *S. aureus*; n (%)	160 (19.8)	192 (21.1)	171 (19.0)	157 (18.6)	129 (18.9)	71 (13.1)	1091 (18.1)
Chronic *P. aeruginosa*; n (%)	152 (18.8)	248 (27.2)	277 (30.7)	298 (35.3)	262 (38.4)	236 (43.5)	1915 (31.9)
Intermittent *P. aeruginosa*; n (%)	153 (19.0)	172 (18.9)	161 (17.8)	147 (17.4)	134 (19.6)	83 (15.3)	1068 (17.8)
B. cepacia complex; n (%)	30 (3.7)	41 (4.5)	48 (5.3)	45 (5.3)	31 (4.5)	21 (3.9)	273 (4.5)
B. cenocepacia; n (%)	5 (0.6)	8 (0.9)	12 (1.3)	14 (1.7)	6 (0.9)	<5	71 (1.2)
B. multivorans; n (%)	15 (1.9)	19 (2.1)	23 (2.5)	23 (2.7)	22 (3.2)	12 (2.2)	137 (2.3)
B. other cepacia; n (%)	5 (0.6)	12 (1.3)	7 (0.8)	5 (0.6)	<5	<5	43 (0.7)
Methicillin-resistant S. aureus; n (%)	28 (3.5)	32 (3.5)	35 (3.9)	36 (4.3)	22 (3.2)	13 (2.4)	203 (3.4)
H. influenzae; n (%)	71 (8.8)	56 (6.1)	51 (5.7)	41 (4.9)	38 (5.6)	19 (3.5)	333 (5.5)
NTM; n (%)	73 (9.0)	88 (9.7)	79 (8.8)	76 (9.0)	48 (7.0)	40 (7.4)	490 (8.2)
Aspergillus; n (%)	184 (22.8)	191 (21.0)	164 (18.2)	134 (15.9)	107 (15.7)	68 (12.5)	1033 (17.2)

	Adult age range (years)						Overall
	40-43	44-47	48-51	52-55	56-59	60+	≥16 years
Number in age range	411	248	245	157	117	145	6012
Number who had culture taken*	360	214	209	137	93	125	5329
Chronic *S. aureus*; n (%)	61 (14.8)	31 (12.5)	25 (10.2)	16 (10.2)	11 (9.4)	19 (13.1)	941 (15.7)
Intermittent *S. aureus*; n (%)	60 (14.6)	41 (16.5)	44 (18.0)	21 (13.4)	19 (16.2)	26 (17.9)	1091 (18.1)
Chronic *P. aeruginosa*; n (%)	165 (40.1)	80 (32.3)	84 (34.3)	55 (35.0)	29 (24.8)	29 (20.0)	1915 (31.9)
Intermittent *P. aeruginosa*; n (%)	69 (16.8)	45 (18.1)	38 (15.5)	21 (13.4)	21 (17.9)	24 (16.6)	1068 (17.8)
B. cepacia complex; n (%)	17 (4.1)	15 (6.0)	12 (4.9)	7 (4.5)	5 (4.3)	<5	273 (4.5)
B. cenocepacia; n (%)	<5	7 (2.8)	<5	6 (3.8)	<5	<5	71 (1.2)
B. multivorans; n (%)	9 (2.2)	5 (2.0)	6 (2.4)	<5	<5	<5	137 (2.3)
B. other cepacia; n (%)	<5	<5	<5	<5	<5	<5	43 (0.7)
MRSA; n (%)	6 (1.5)	8 (3.2)	5 (2.0)	6 (3.8)	<5	8 (5.5)	203 (3.4)
H. influenzae; n (%)	20 (4.9)	12 (4.8)	10 (4.1)	5 (3.2)	7 (6.0)	<5	333 (5.5)
NTM; n (%)	29 (7.1)	12 (4.8)	17 (6.9)	8 (5.1)	10 (8.5)	10 (6.9)	490 (8.2)
Aspergillus; n (%)	60 (14.6)	35 (14.1)	38 (15.5)	18 (11.5)	13 (11.1)	21 (14.5)	1033 (17.2)

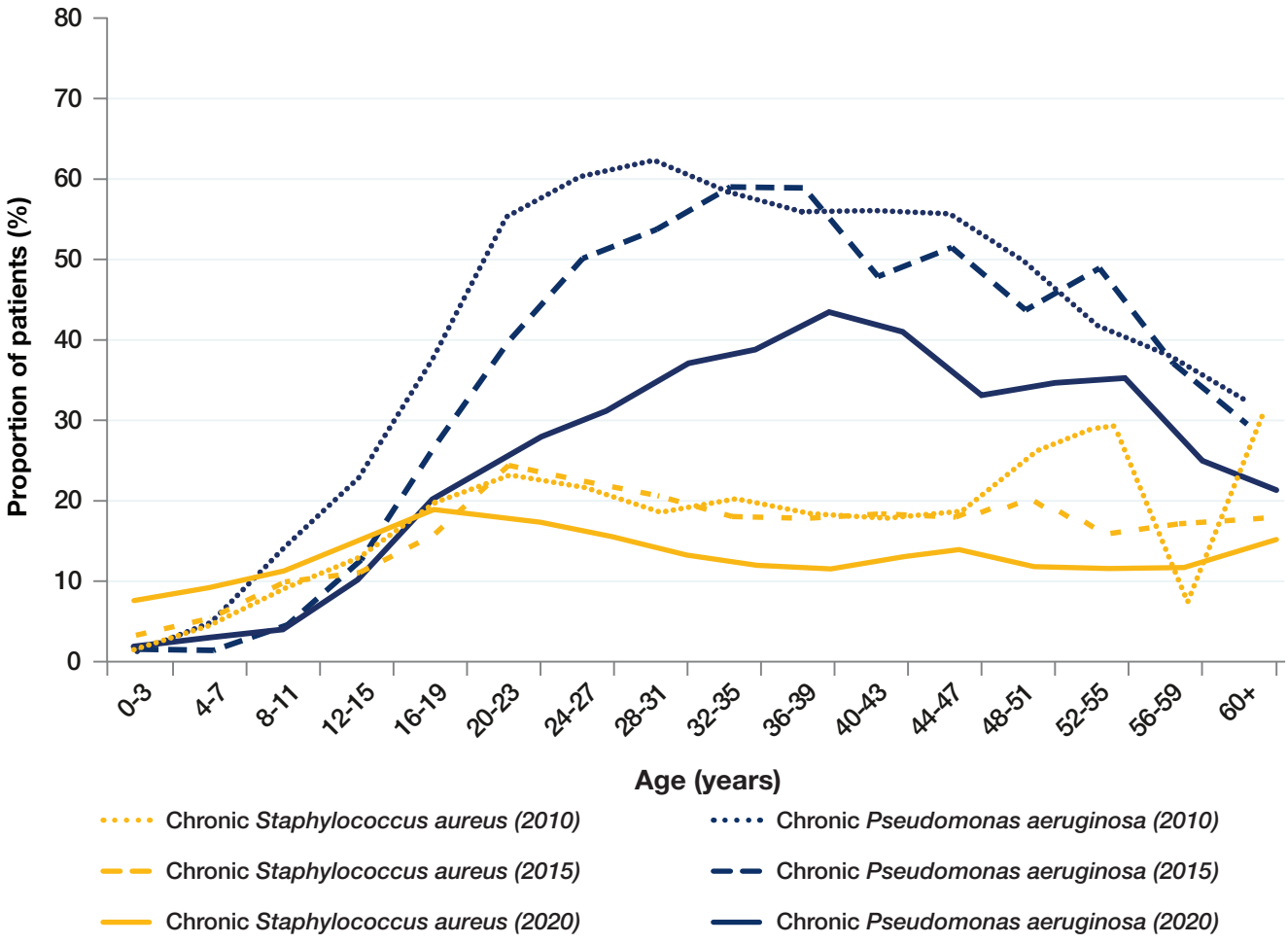
* Proportions are calculated from number in age range, whereas prior to 2019 they were calculated from the number of people with CF who were recorded as having had a culture taken.

1.19 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.

	2018 (n=9847)	2019 (n=10070)	2020 (n=9922)
NTM prevalence; n (%)	697 (7.1)	674 (6.7)	620 (6.9)
On NTM treatment in the given year; n (% of NTM prevalence in given year)	343 (49.2)	362 (53.7)	326 (52.6)
NTM incidence [1]; n (%)	293 (3.2)	279 (3.0)	226 (2.5)
M. abscessus prevalence	419 (4.3)	382 (3.8)	361 (3.9)
M. abscessus incidence [2]; n (%)	157 (1.6)	126 (1.3)	103 (1.1)

1.20 Lung infections over time
N=7937 in 2010, N=9587 in 2015, N=9922 in 2020



Due to the COVID-19 pandemic, a reduced number of patients received the necessary 3 tests needed to designate an infection as chronic. Reductions in chronic infections reported in 2020 should not be considered as strict reductions in prevalence. The median age of people with chronic *Pseudomonas aeruginosa* infection increased from 25 years in 2010 to 31 years in 2020.

[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.

Chronic *Staphylococcus aureus*			
Age (years)	**2010 (%)**	**2015 (%)**	**2020 (%)**
0-3	1.0	2.9	6.8
4-7	4.1	5.2	8.7
8-11	8.9	9.9	11.2
12-15	12.8	11.0	15.6
16-19	19.7	15.8	19.3
20-23	23.4	24.3	19.3
24-27	21.5	22.5	18.6
28-31	18.5	20.3	14.4
32-35	2.2	17.9	12.9
36-39	17.4	17.4	12.7
40-43	17.5	17.8	14.8
44-47	18.9	17.8	12.5
48-51	26.0	20.1	10.2
52-55	30.0	15.3	10.2
56-59	6.7	16.7	9.4
60+	30.0	17.3	13.1
<16 years	6.8	7.3	10.8
≥16 years	20.6	19.6	15.7
<18 years	8.1	8.1	11.6
≥18 years	20.9	20.1	15.4

Chronic *Pseudomonas aeruginosa*			
Age (years)	**2010 (%)**	**2015 (%)**	**2020 (%)**
0-3	2.6	2.3	1.8
4-7	6.2	2.2	2.8
8-11	15.6	5.3	3.8
12-15	24.0	13.4	9.8
16-19	38.6	27.3	18.8
20-23	56.6	39.7	27.2
24-27	60.7	50.0	30.7
28-31	63.3	52.8	35.3
32-35	59.2	58.5	38.4
36-39	53.8	57.4	43.5
40-43	56.1	46.4	40.1
44-47	57.3	51.0	32.3
48-51	50.0	40.7	34.3
52-55	42.5	47.6	35.0
56-59	36.7	36.4	24.8
60+	32.5	29.1	20.0
<16 years	12.5	5.7	4.7
≥16 years	53.7	45.3	31.9
<18 years	14.5	7.9	5.9
≥18 years	56.4	47.0	32.8

1.21 COVID-19* infection in 2020

COVID-19 management and outcomes for people with CF infected with COVID-19 during the calendar year of 2020 (01 of January 2020—31 of December 2020) are described below. Information is stratified by sex, ethnicity, age, organ transplant status and Best FEV1% prior to catching COVID-19

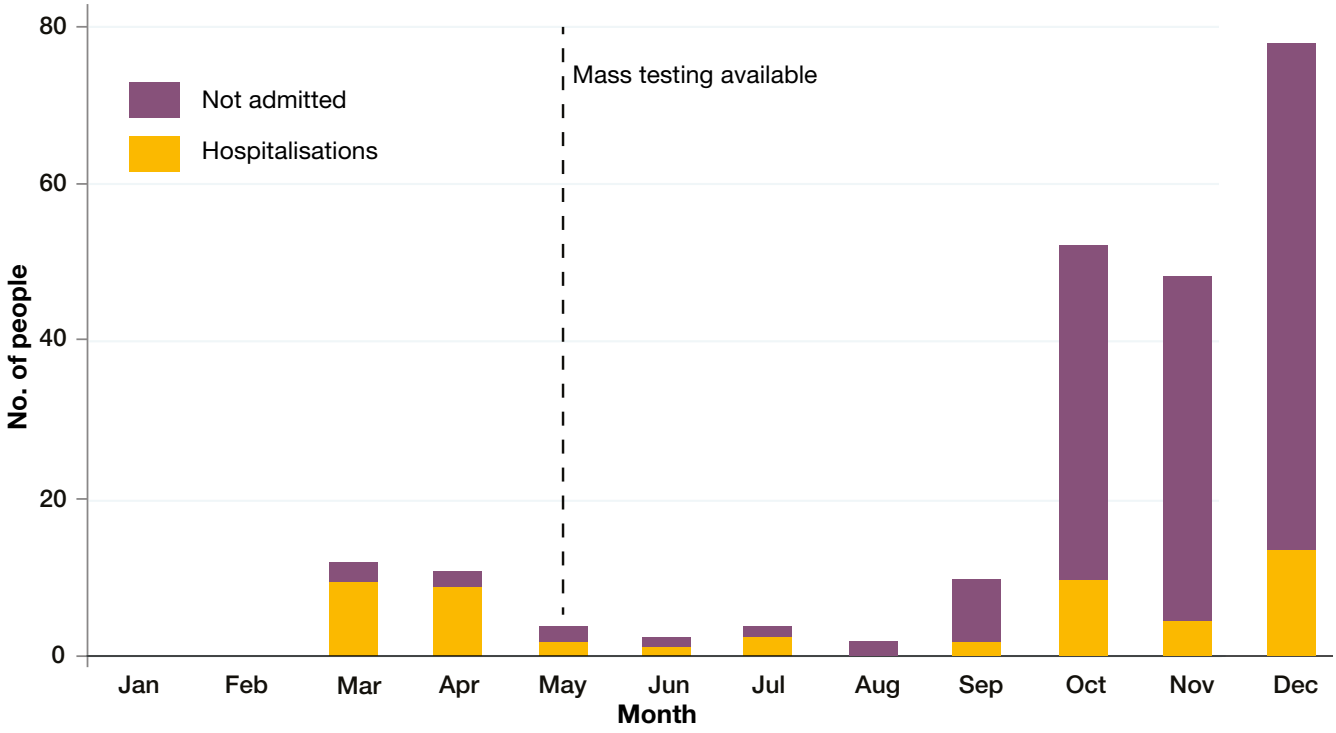
	Total	Symptomatic	COVID-19 Management		COVID-19 Outcomes		
			IV antibiotics	Oral antibiotics	Hospitalised	Additional oxygen	Hospitalised with additional oxygen
Overall; n (%)							
All cases	217 (100)	144 (66)	41 (19)	79 (36)	51 (24)	22 (10)	20 (9)
Sex; n (%)							
Male	113 (52)	35 (69)	19 (17)	42 (37)	24 (21)	13 (12)	12 (11)
Female	104 (48)	66 (63)	22 (21)	37 (36)	27 (26)	9 (9)	8 (8)
Ethnicity; n (%)							
White	194 (89)	129 (66)	33 (17)	70 (36)	43 (22)	21 (11)	19 (10)
Non-White	17 (8)	12 (71)	7 (41)	7 (41)	7 (41)	<5	<5
Unknown	6 (3)	<5	<5	<5	<5	0 (0)	0 (0)
Age; n (%)							
≥16	171 (79)	121 (71)	37 (22)	65 (38)	45 (26)	21 (12)	19 (11)
Under 16	46 (21)	23 (50)	4 (9)	14 (30)	6 (13)	<5	<5
Transplants; n (%)							
Yes	12 (6)	7 (58)	<5	5 (42)	7 (58)	<5	<5
No	205 (94)	137 (67)	37 (18)	74 (36)	44 (21)	20 (10)	18 (9)
**Best FEV; n (%)							
<40	21 (10)	15 (71)	12 (57)	11 (52)	15 (71)	8 (38)	8 (38)
40-70	60 (28)	38 (63)	14 (23)	22 (37)	17 (28)	9 (15)	7 (12)
>70	112 (52)	81 (72)	9 (8)	38 (34)	10 (9)	<5	<5

In 2020, five people were admitted to Intensive care with Covid-19 and less than 5 were thought to have died from Covid-19.

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

**Patients who had a lung transplant were excluded from the “Best FEV%” analysis*

The number of COVID-19 cases and hospitalisations from March 2020 to December 2020



Not admitted patients may have reported their Covid status because of symptoms, contact tracing or other screening. Some admitted patients may have been found to be positive due to routine hospital screening measures rather than new symptoms.

Complications

1.22 Complications in 2020

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

	Overall (N=9922)	<16 years (n=3910)	≥16 years (n=6012)
	n (%)		
Respiratory related			
Nasal polyps requiring surgery	435 (4.4)	127 (3.2)	308 (5.1)
Sinus disease	803 (8.1)	56 (1.4)	747 (12.4)
Asthma	816 (8.2)	189 (4.8)	627 (10.4)
ABPA	704 (7.1)	146 (3.7)	558 (9.3)
Any haemoptysis	340 (3.4)	12 (0.3)	328 (5.5)
Massive haemoptysis	-*	<5	16 (0.3)
Pneumothorax requiring chest tube	-*	<5	21 (0.3)
Pancreas and hepatobiliary disease			
Raised liver enzymes	1056 (10.6)	374 (9.6)	682 (11.3)
Liver disease	1561 (15.7)	358 (9.2)	1203 (20.0)
Cirrhosis with no portal hypertension	71 (0.7)	14 (0.4)	57 (0.9)
Cirrhosis with portal hypertension	137 (1.4)	30 (0.8)	107 (1.8)
Gall bladder disease requiring surgery	150 (1.5)	28 (0.7)	122 (2.0)
Pancreatitis	57 (0.6)	14 (0.4)	43 (0.7)
Upper gastrointestinal (GI)			
GERD	2112 (21.3)	264 (6.8)	1848 (30.7)
Peptic ulcer	-*	0	<5
GI bleed (varices as source)	11 (0.1)	5 (0.1)	6 (0.1)
GI bleed (non-varices as source)	-*	<5	14 (0.2)
Lower gastrointestinal			
Intestinal obstruction	34 (0.3)	14 (0.4)	20 (0.3)
DIOS	482 (4.9)	91 (2.3)	391 (6.5)
Fibrosing colonopathy / colonic stricture	-*	0	<5
Rectal prolapse	11 (0.1)	6 (0.2)	5 (0.1)
Renal			
Kidney stones	150 (1.5)	15 (0.4)	135 (2.2)
Renal failure	87 (0.9)	<5	85 (1.4)
Musculoskeletal			
Arthritis	107 (1.1)	7 (0.2)	100 (1.7)
Arthropathy	263 (2.7)	10 (0.3)	253 (4.2)
Bone fracture	48 (0.5)	14 (0.4)	34 (0.6)
Osteopenia	1114 (11.2)	15 (0.4)	1099 (18.3)
Osteoporosis	-*	<5	431 (7.2)
Other			
Cancer confirmed by histology	-*	<5	31 (0.5)
Port inserted or replaced	237 (2.4)	76 (1.9)	161 (2.7)
Depression	497 (5.0)	13 (0.3)	484 (8.1)
Hearing loss	355 (3.6)	37 (0.9)	318 (5.3)
Hypertension	-*	<5	152 (2.5)

* 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.

	2019			2020		
	Overall (n=10070)	<16 years (n=3966)	≥16 years (n=6104)	Overall (n=9922)	<16 years (n=3910)	≥16 years (n=6012)
ABPA; n (%)	225 (2.2)	62 (1.6)	163 (2.7)	216 (2.2)	87 (2.2)	129 (2.2)
Cirrhosis with no portal hypertension; n (%)	36 (0.4)	9 (0.2)	27 (0.4)	40 (0.4)	11 (0.3)	29 (0.5)
Cirrhosis with portal hypertension; n (%)	54 (0.5)	12 (0.3)	42 (0.7)	46 (0.5)	11 (0.3)	35 (0.6)
Cancer confirmed by histology; n (%)	20 (0.2)	0	20 (0.3)	-*	<5	17 (0.3)

1.24 CF-related diabetes
N=7576

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=7576)	10-15 years (n=1564)	≥16 years (n=6012)
On CFRD treatment; n (%)	2284 (30.1)	162 (10.4)	2122 (35.3)
Of those on treatment			
Insulin[1]; n (%)	2015 (88.2)	156 (96.3)	1859 (87.6)
CFRD Screening; n (%)			
Yes	2915 (38.5)	1001 (64.0)	1914 (31.8)
Screening type			
Continuous glucose monitoring[2]; n (%)	860 (29.5)	237 (23.7)	623 (32.5)
Oral glucose tolerance test[2]; n (%)	929 (31.9)	340 (34.0)	589 (30.8)
Not screened (known CFRD)	2015 (26.6)	112 (7.2)	1903 (31.7)
Not screened (other)	2413 (31.9)	413 (26.4)	2000 (33.3)
Unknown	87 (1.1)	36 (2.3)	51 (0.8)

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

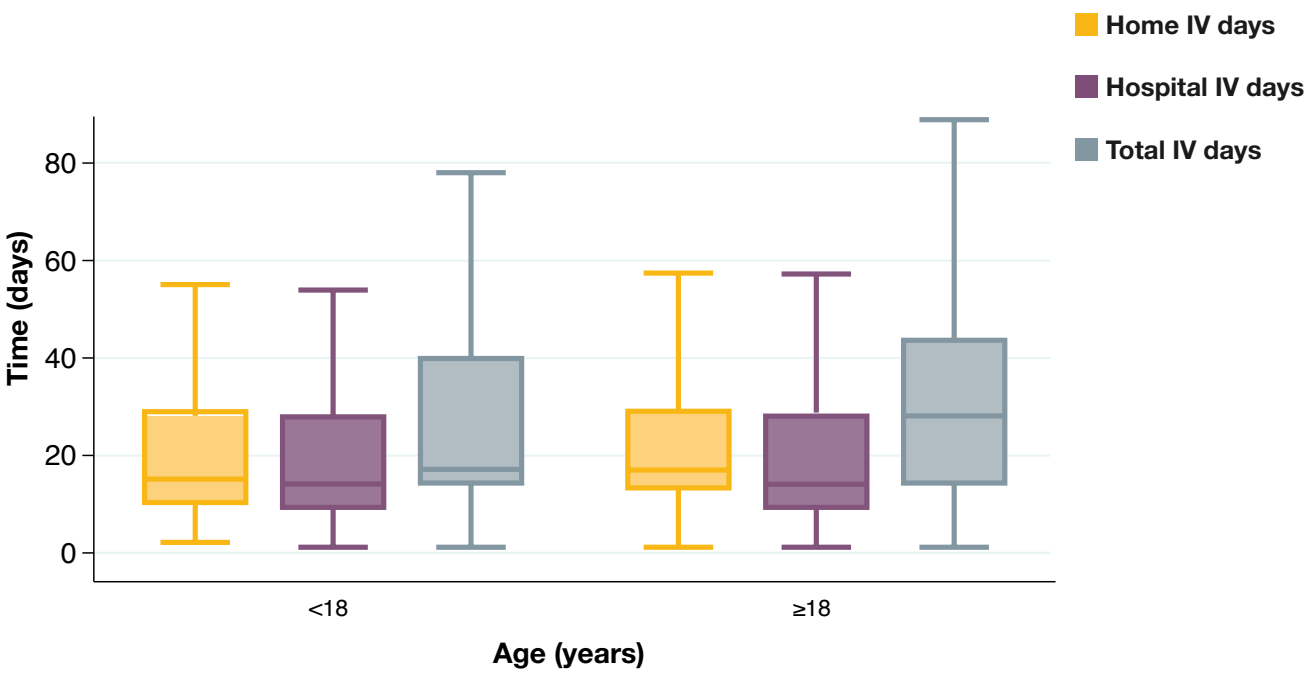
Antibiotics

1.25 Intravenous (IV) antibiotics
N=9922

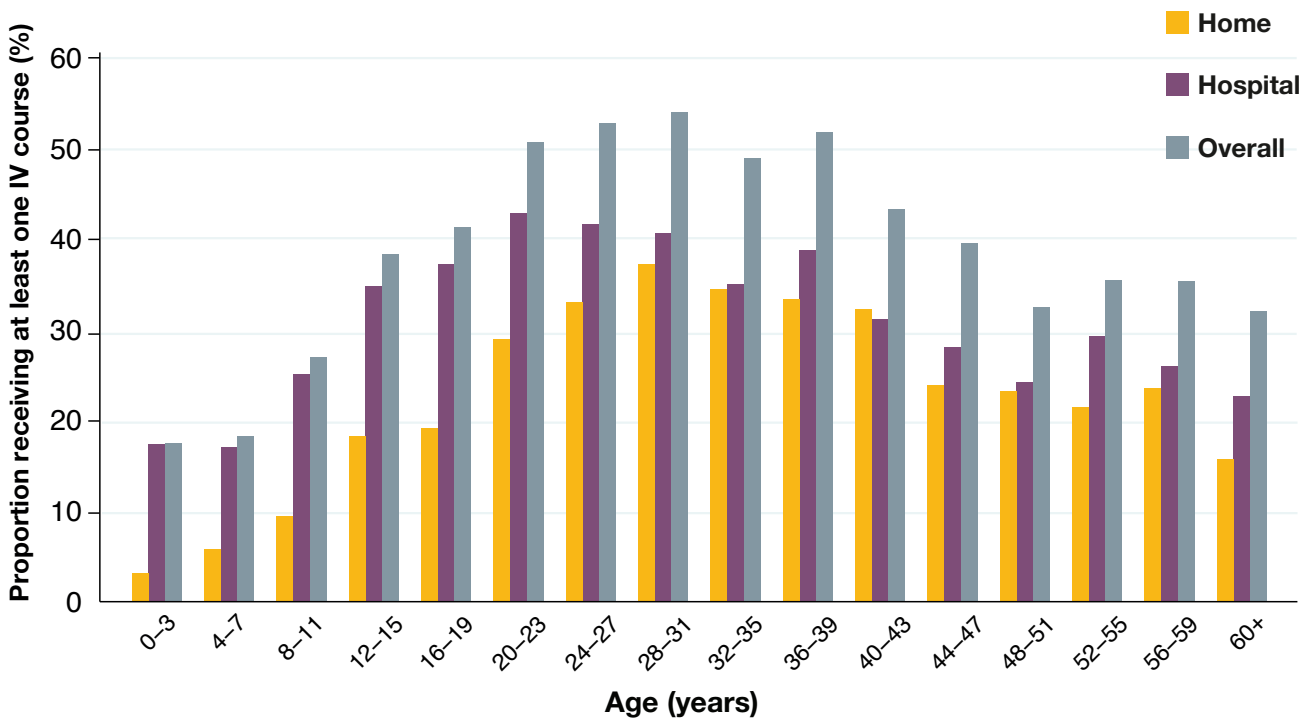
When someone with CF becomes unwell with an infection, they might be prescribed intravenous (IV) antibiotics. IV antibiotics are given to the patient through their veins. This treatment can take a number of days and might take place as a hospital inpatient, or at home.

		Home		Hospital		Total	
Age	N	Patients n (%)	Median days (IQR)	Patients n (%)	Median days (IQR)	Patients n (%)	Median days (IQR)
0-3	730	17 (2.3)	7 (6-12)	127 (17.4)	14 (6-12)	128 (17.5)	14 (10-16)
4-7	1027	57 (5.6)	14 (8-28)	182 (17.7)	14 (8-28)	193 (18.8)	14 (13-29)
8-11	1147	113 (9.9)	14 (10-29)	293 (25.5)	14 (10-29)	316 (27.6)	16 (14-37)
12-15	1006	171 (17.0)	16 (12-33)	343 (34.1)	17 (12-33)	389 (38.7)	28 (14-43)
16-19	807	153 (19.0)	14 (10-28)	297 (36.8)	15 (10-28)	335 (41.5)	22 (14-42)
20-23	911	271 (29.7)	15 (10-28)	388 (42.6)	14 (10-28)	473 (51.9)	25 (14-42)
24-27	902	301 (33.4)	17 (12-35)	375 (41.6)	14 (12-35)	478 (53.0)	26 (14-44)
28-31	843	313 (37.1)	20 (13-32)	345 (40.9)	14 (13-32)	452 (53.6)	27 (14-43)
32-35	683	240 (35.1)	18 (14-35)	242 (35.4)	14 (14-35)	340 (49.8)	27 (14-44)
36-39	543	185 (34.1)	21 (14-35)	213 (39.2)	14 (14-35)	282 (51.9)	27 (14-48)
40-43	411	134 (32.6)	14 (12-36)	132 (32.1)	13 (12-36)	180 (43.8)	19 (14-43)
44-47	248	61 (24.6)	16 (13-29)	72 (29.0)	17 (13-29)	100 (40.3)	24 (14-42)
48-51	245	57 (23.3)	25 (14-38)	60 (24.5)	15 (14-38)	81 (33.1)	28 (14-53)
52-55	157	34 (21.7)	20 (14-28)	46 (29.3)	17 (14-28)	56 (35.7)	28 (15-43)
56-59	117	28 (23.9)	14 (13-23)	31 (26.5)	14 (13-23)	42 (35.9)	15 (14-31)
60+	145	22 (15.2)	14 (11-28)	35 (24.1)	14 (11-28)	47 (32.4)	15 (14-37)
<16	3910	358 (9.2)	14 (10-28)	945 (24.2)	14 (10-28)	1026 (26.2)	16 (14-38)
≥16	6012	1799 (29.9)	17 (13-31)	2236 (37.2)	14 (13-31)	2866 (47.7)	25 (14-43)
<18	4289	428 (10.0)	14 (10-29)	1084 (25.3)	14 (10-29)	1183 (27.6)	18 (14-40)
≥18	5633	1729 (30.7)	17 (13-31)	2097 (37.2)	14 (13-31)	2709 (48.1)	25 (14-43)
Overall	**9922**	**2157 (21.7)**	**16 (12-31)**	**3181 (32.1)**	**14 (12-31)**	**3892 (39.2)**	**23 (14-42)**

This box plot graph illustrates the spread of the number of days on IV antibiotics in the UK 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 21.7% and in hospital was 32.1%. The proportion receiving any IVs was 39.2%.



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

	2010			2015			2020		
	Overall	<16 years	≥16 years	Overall	<16 years	≥16 years	Overall	<16 years	≥16 years
Patients with chronic *P. aeruginosa*; n	2861	433	2428	2859	239	2625	2100	185	1915
Tobramycin solution; n (%)	707 (24.7)	86 (19.9)	621 (25.6)	653 (22.8)	86 (36.8)	567 (21.6)	623 (29.7)	76 (41.1)	547 (28.6)
Other aminoglycoside; n (%)	74 (2.6)	17 (3.9)	57 (2.3)	95 (3.3)	15 (6.4)	80 (3.1)	-*	<5	68 (3.6)
Colistin; n (%)	1280 (44.7)	241 (55.7)	1039 (42.8)	840 (29.4)	108 (46.2)	732 (28.0)	629 (30.0)	68 (36.8)	561 (29.3)
Promixin; n (%)	741 (25.9)	107 (24.7)	634 (26.1)	896 (31.3)	111 (47.4)	785 (29.9)	704 (33.5)	83 (44.9)	621 (32.4)
Aztreonam; n (%)	-	-	-	485 (17.0)	8 (3.4)	477 (18.2)	600 (28.6)	16 (8.6)	584 (30.5)
Colistimethate (inh) inhalation powder; n (%)	-	-	-	537 (18.8)	22 (9.4)	515 (19.6)	389 (18.5)	15 (8.1)	374 (19.5)
Tobramycin inhalation powder; n (%)	-	-	-	888 (31.1)	25 (10.7)	863 (32.9)	460 (21.9)	7 (3.8)	453 (23.7)
Levofloxacin ;n(%)	-	-	-	-	-	-	55 (2.6)	0	55 (2.9)
At least one of the above; n (%)	2287 (79.9)	376 (86.8)	1911 (78.7)	2547 (89.1)	226 (96.6)	2321 (96.6)	1898 (90.4)	165 (89.2)	1733 (90.5)

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 (%)
2010	**Overall**	3188	1327 (41.6)	1861 (58.4)
	0-3 years	17	17 (100.0)	0
	4-15 years	593	440 (74.2)	153 (25.8)
	≥16 years	2578	870 (33.7)	1708 (66.3)
2015	**Overall**	3790	1916 (50.6)	1874 (49.4)
	0-3 years	-*	18 (85.7)	<5
	4-15 years	574	491 (85.5)	83 (14.5)
	≥16 years	3195	1407 (44.0)	1788 (56.0)
2020	**Overall**	4221	1435 (34.0)	2786 (66.0)
	0-3 years	-*	<5	40 (97.6)
	4-15 years	658	76 (11.6)	582 (88.4)
	≥16 years	3522	1358 (38.6)	2164 (61.4)

* 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	730	381 (52.2)
4-7	1027	325 (31.6)
8-11	1147	307 (26.8)
12-15	1006	241 (24.0)
16-19	807	204 (25.3)
20-23	911	224 (24.6)
24-27	902	72 (8.0)
28-31	843	72 (8.5)
32-35	683	32 (4.7)
36-39	543	35 (6.4)
40-43	411	24 (5.8)
44-47	248	14 (5.6)
48-51	245	13 (5.3)
52-55	157	10 (6.4)
56-59	117	5 (4.3)
60+	145	7 (4.8)
<16 years	3910	1254 (32.1)
≥16 years	6012	712 (11.8)
<18 years	4289	1339 (31.2)
≥18 years	5633	627 (11.1)
Overall	**9922**	**1966 (19.8)**

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	730	124 (17.0)	41 (5.6)	1 (0.1)
4-7	1027	383 (37.3)	165 (16.1)	40 (3.9)
8-11	1147	574 (50.0)	208 (18.1)	168 (14.6)
12-15	1006	612 (60.8)	186 (18.5)	284 (28.2)
16-19	807	565 (70.0)	176 (21.8)	287 (35.6)
20-23	911	655 (71.9)	202 (22.2)	356 (39.1)
24-27	902	639 (70.8)	173 (19.2)	353 (39.1)
28-31	843	627 (74.4)	186 (22.1)	415 (49.2)
32-35	683	507 (74.2)	143 (20.9)	319 (46.7)
36-39	543	402 (74.0)	126 (23.2)	270 (49.7)
40-43	411	289 (70.3)	86 (20.9)	205 (49.9)
44-47	248	169 (68.1)	46 (18.5)	130 (52.4)
48-51	245	161 (65.7)	58 (23.7)	119 (48.6)
52-55	157	107 (68.2)	42 (26.8)	71 (45.2)
56-59	117	80 (68.4)	25 (21.4)	40 (34.2)
60+	145	106 (73.1)	35 (24.1)	76 (52.4)
<16 years	3910	1693 (43.3)	600 (15.3)	493 (12.6)
≥16 years	6012	4307 (71.6)	1298 (21.6)	2641 (43.9)
<18 years	4289	1942 (45.3)	666 (15.5)	622 (14.5)
≥18 years	5633	4058 (72.0)	1232 (21.9)	2512 (44.6)
Overall	**9922**	**6000 (60.5)**	**1898 (19.1)**	**3134 (31.6)**

Muco-active therapies

1.30 Mannitol

Age	Total patients	Patients on Mannitol; n (%)
0-3	730	0
4-7	1027	0
8-11	1147	<5
12-15	1006	<5
16-19	807	13 (1.6)
20-23	911	40 (4.4)
24-27	902	60 (6.7)
28-31	843	63 (7.5)
32-35	683	56 (8.2)
36-39	543	32 (5.9)
40-43	411	30 (7.3)
44-47	248	20 (8.1)
48-51	245	10 (4.1)
52-55	157	8 (5.1)
56-59	117	5 (4.3)
60+	145	<5
<16 years	3910	<5
≥16 years	6012	338 (5.6)
<18 years	4289	5 (0.1)
≥18 years	5633	337 (6.0)
Overall	**9922**	**342 (3.4)**

1.31 DNase

	2010		2015		2020	
Age; years	Total patients	Patients on DNase; n (%)	Total patients	Patients on DNase; n (%)	Total patients	Patients on DNase; n (%)
0-3	919	87 (9.5)	921	121 (13.1)	730	143 (19.6)
4-7	785	172 (21.9)	1098	478 (43.5)	1027	524 (51.0)
8-11	858	367 (42.8)	946	665 (70.3)	1147	905 (78.9)
12-15	969	528 (54.5)	880	654 (74.3)	1006	856 (85.1)
16-19	942	509 (54.0)	999	733 (73.4)	807	712 (88.2)
20-23	917	491 (53.5)	1000	654 (65.4)	911	805 (88.4)
24-27	735	389 (52.9)	954	609 (63.8)	902	711 (78.8)
28-31	577	304 (52.7)	749	470 (62.8)	843	631 (74.9)
32-35	346	155 (44.8)	634	376 (59.3)	683	477 (69.8)
36-39	288	127 (44.1)	407	225 (55.3)	543	362 (66.7)
40-43	223	94 (42.2)	286	135 (47.2)	411	248 (60.3)
44-47	164	75 (45.7)	260	140 (53.8)	248	151 (60.9)
48-51	104	47 (45.2)	170	89 (52.4)	245	141 (57.6)
52-55	40	18 (45.0)	121	64 (52.9)	157	88 (56.1)
56-59	30	12 (40.0)	63	32 (50.8)	117	66 (56.4)
60+	40	16 (40.0)	99	50 (50.5)	145	82 (56.6)
<16 years	3531	1154 (32.7)	3845	1918 (49.9)	3910	2428 (62.1)
≥16 years	4406	2237 (50.8)	5742	3577 (62.3)	6012	4474 (74.4)
<18 years	3987	1394 (35.0)	4336	2300 (53.0)	4289	2766 (64.5)
≥18 years	3950	1997 (50.6)	5251	3195 (60.8)	5633	4136 (73.4)
Overall	**7937**	**3391 (42.7)**	**9587**	**5495 (57.3)**	**9922**	**6902 (69.6)**

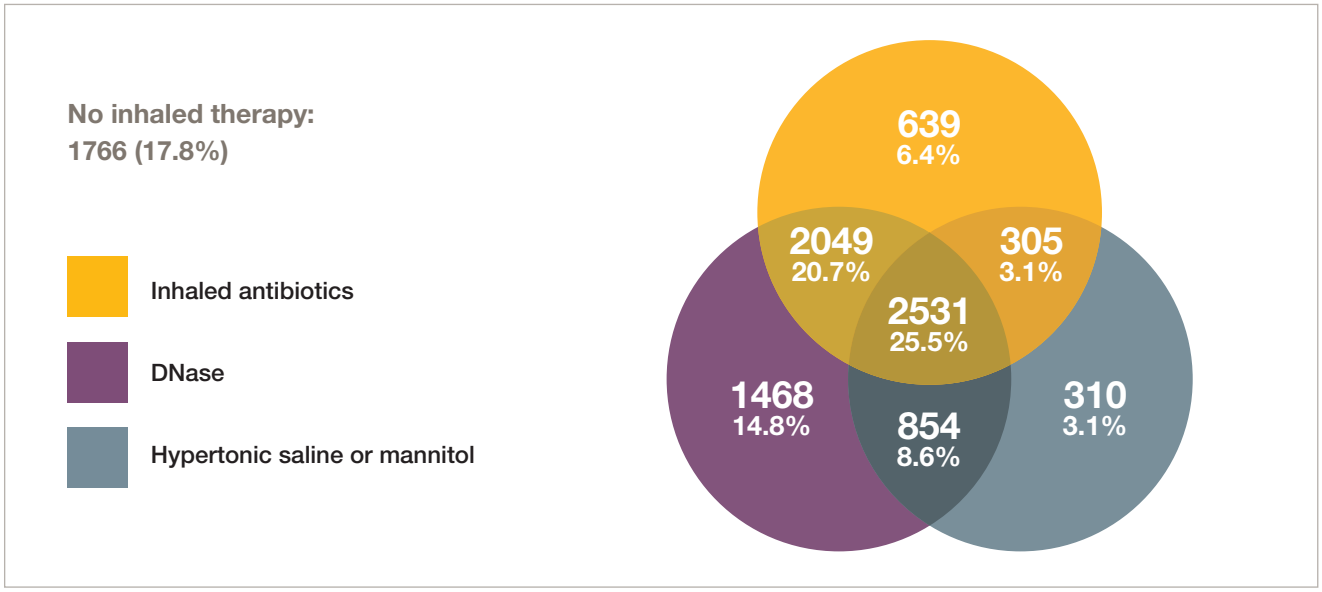
1.32 Hypertonic saline

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

	2010		2015		2020	
Age; years	**Total patients; N**	**Patients on hypertonic saline; n (%)**	**Total patients; N**	**Patients on hypertonic saline; n (%)**	**Total patients**	**Patients on hypertonic saline; n (%)**
0-3	919	14 (1.5)	921	55 (6.0)	730	109 (14.9)
4-7	785	37 (4.7)	1098	198 (18.0)	1027	317 (30.9)
8-11	858	85 (9.9)	946	282 (29.8)	1147	452 (39.4)
12-15	969	132 (13.6)	880	359 (40.8)	1006	504 (50.1)
16-19	942	129 (13.7)	999	354 (35.4)	807	461 (57.1)
20-23	917	127 (13.8)	1000	290 (29.0)	911	493 (54.1)
24-27	735	117 (15.9)	954	277 (29.0)	902	361 (40.0)
28-31	577	83 (14.4)	749	246 (32.8)	843	274 (32.5)
32-35	346	60 (17.3)	634	197 (31.1)	683	212 (31.0)
36-39	288	35 (12.2)	407	122 (30.0)	543	199 (36.6)
40-43	223	26 (11.7)	286	60 (21.0)	411	124 (30.2)
44-47	164	24 (14.6)	260	63 (24.2)	248	80 (32.3)
48-51	104	17 (16.3)	170	50 (29.4)	245	80 (32.7)
52-55	40	6 (15.0)	121	34 (28.1)	157	44 (28.0)
56-59	30	<5	63	16 (25.4)	117	32 (27.4)
60+	40	<5	99	25 (25.3)	145	43 (29.7)
<16 years	3531	268 (7.6)	3845	894 (23.3)	3910	1382 (35.3)
≥16 years	4406	630 (14.3)	5742	1734 (30.2)	6012	2403 (40.0)
<18 years	3987	329 (8.3)	4336	1084 (25.0)	4289	1588 (37.0)
≥18 years	3950	569 (14.4)	5251	1544 (29.4)	5633	2197 (39.0)
Overall	**7937**	**898 (11.3)**	**9587**	**2628 (27.4)**	**9922**	**3785 (38.1)**

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. 1766 (17.8%) people are on no inhaled therapies.



Other therapies

1.34 CFTR modifiers

Ivacaftor

As of November 2020, ivacaftor was approved for use on the NHS across the UK for people aged four months and older with at least one copy of a CFTR 'gating' mutation, and for people aged 6 months and over with the R117H. Additionally patients in Scotland and NI can access from age 4 months with specific rare mutations.

Lumacaftor/ivacaftor

Lumacaftor/ivacaftor is licensed for use in patients aged 2 and over with two copies of the F508del mutation. Managed access was agreed to make lumacaftor/ivacaftor available on the NHS from Autumn/Winter 2019.

Tezacaftor/ivacaftor

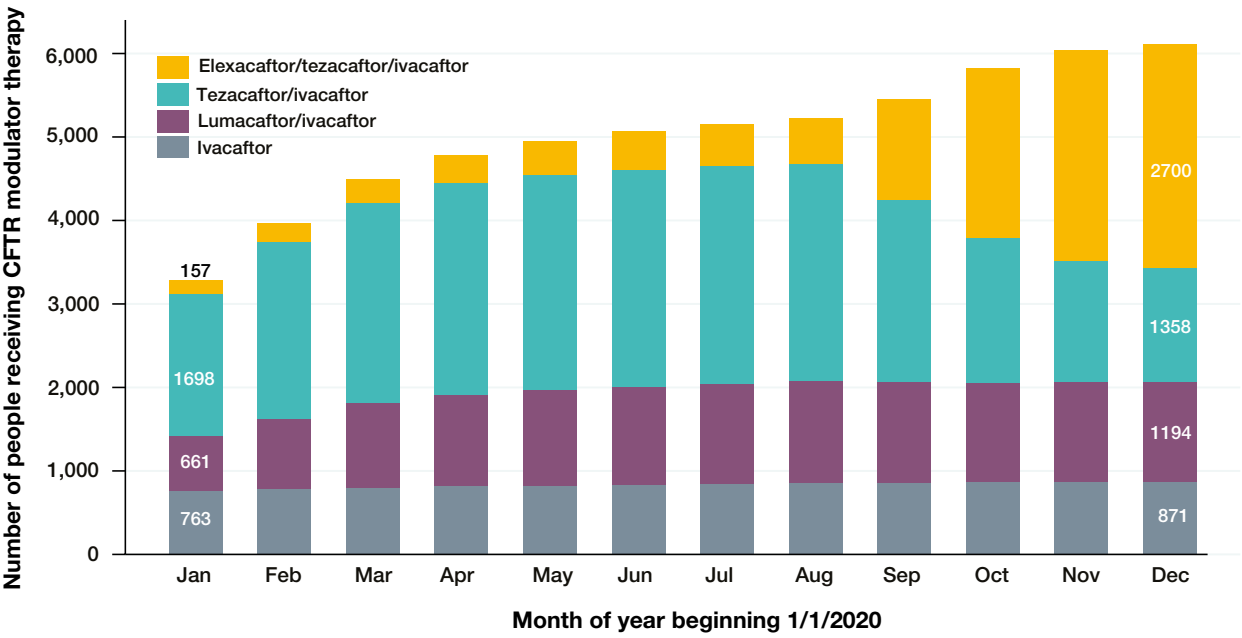
Tezacaftor/ivacaftor is licenced for use in patients aged 6 and over who have two copies of the F508del mutation, or a single copy of F508del and one of 14 residual function mutations. Managed access was agreed to make tezacaftor/ivacaftor available on the NHS from Autumn/Winter 2019. Patients aged 6+ with rare mutations gained access in Scotland and NI from December 2020.I.

Elexacaftor/tezacaftor/ivacaftor

As an extension of the managed access agreement, from August 2020 Elexacaftor/tezacaftor/ivacaftor was made available in the UK for those ages 12 and over who have two copies of the F508del mutation, or a single copy of F508del and one minimal function mutation.

CFTR modulator use in 2020

A total of 6208 people received CFTR modulator treatment in 2020. The graph below shows the number of people on each drug by month. Where people switched modulators, the most recent prescription is counted.



1.35 Oxygen and non-invasive ventilation

	Overall (N=9922)	<16 years (n=3910)	≥16 years (n=6012)	<18 years (n=4289)	≥18 years (n=5633)
Non-invasive ventilation (NIV); n (%)	176 (1.8)	16 (0.4)	160 (2.7)	21 (0.5)	155 (2.8)
Long-term oxygen; n (%)	537 (5.4)	56 (1.4)	481 (8.0)	70 (1.6)	467 (8.3)
Among those who have long-term oxygen:					
Continuously	-*	<5	65 (13.5)	<5	65 (13.9)
Nocturnal or with exertion	227 (42.3)	15 (26.8)	212 (44.1)	23 (32.9)	204 (43.7)
As required (PRN)	-*	<5	43 (8.9)	<5	41 (8.8)
With exacerbation	197 (36.7)	36 (64.3)	161 (33.5)	40 (57.1)	157 (33.6)

1.36 Physiotherapy

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

	Overall (N=9922)	<16 years (n=3910)	≥16 years (n=6012)	<18 years (n=4289)	≥18 years (n=5633)
Active cycle of breathing techniques; n (%)	1307 (13.2)	347 (8.9)	960 (16.0)	389 (9.1)	918 (16.3)
Autogenic drainage (including assisted autogenic drainage); n (%)	1865 (18.8)	166 (4.2)	1699 (28.3)	221 (5.2)	1644 (29.2)
Postural drainage; n (%)	682 (6.9)	537 (13.7)	145 (2.4)	560 (13.1)	122 (2.2)
Any form of positive expiratory pressure (PEP); n (%)	6036 (60.8)	2968 (75.9)	3068 (51.0)	3275 (76.4)	2761 (49.0)
VEST; n (%)	174 (1.8)	93 (2.4)	81 (1.3)	97 (2.3)	77 (1.4)
Exercise; n (%)	5867 (59.1)	2529 (64.7)	3338 (55.5)	2775 (64.7)	3092 (54.9)
Other; n (%)	1875 (18.9)	1011 (25.9)	864 (14.4)	1057 (24.6)	818 (14.5)

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

1.37 Feeding

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

	Overall (n=9922)	<16 years (n=3910)	≥16 years (n=6012)	<18 years (n=4289)	≥18 years (n=5633)
Any supplemental feeding; n(%)	4282 (43.2)	1328 (34.0)	2954 (49.1)	1487 (34.7)	2795 (49.6)
Nasogastric tube; n(%)	73 (0.7)	12 (0.3)	61 (1.0)	13 (0.3)	60 (1.1)
Gastrostomy tube/button; n(%)	499 (5.0)	196 (5.0)	303 (5.0)	227 (5.3)	272 (4.8)
Jejunal; n(%)	-*	<5	5 (0.1)	<5	5 (0.1)
Total parenteral nutrition (TPN); n(%)	-*	<5	6 (0.1)	<5	6 (0.1)

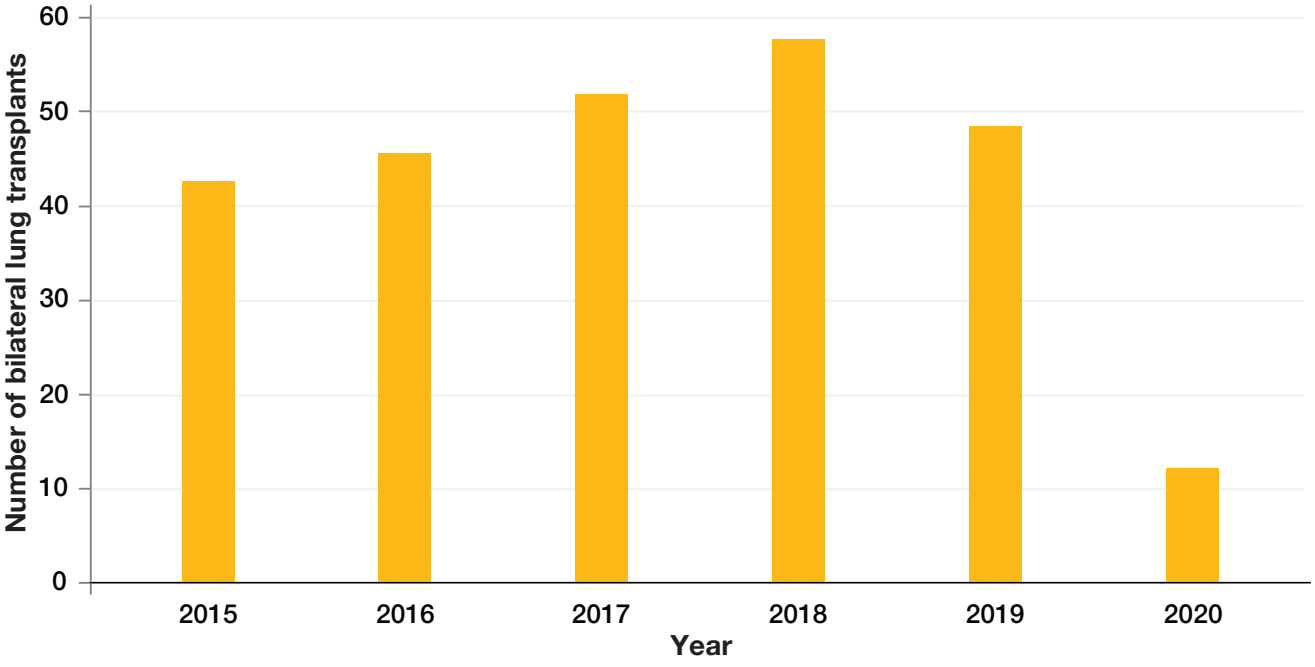
* Redacted to adhere to statistical disclosure guidelines.

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.

	2015	2016	2017	2018	2019	2020
Number evaluated	229	221	235	247	241	175
Number accepted	125	96	121	104	96	66
Number receiving aged <16 years	<5	<5	5	<5	<5	0
Bilateral lung	<5	<5	<5	0	<5	0
Liver	<5	0	0	<5	<5	0
Other	<5	0	<5	0	0	0
Number receiving aged ≥16 years	46	51	53	63	54	15
Bilateral lung	42	46	51	58	49	12
Liver	<5	<5	0	<5	<5	<5
Other	<5	<5	<5	<5	<5	<5

The graph below shows the total number of double lung transplants over time in patients aged 16 and over



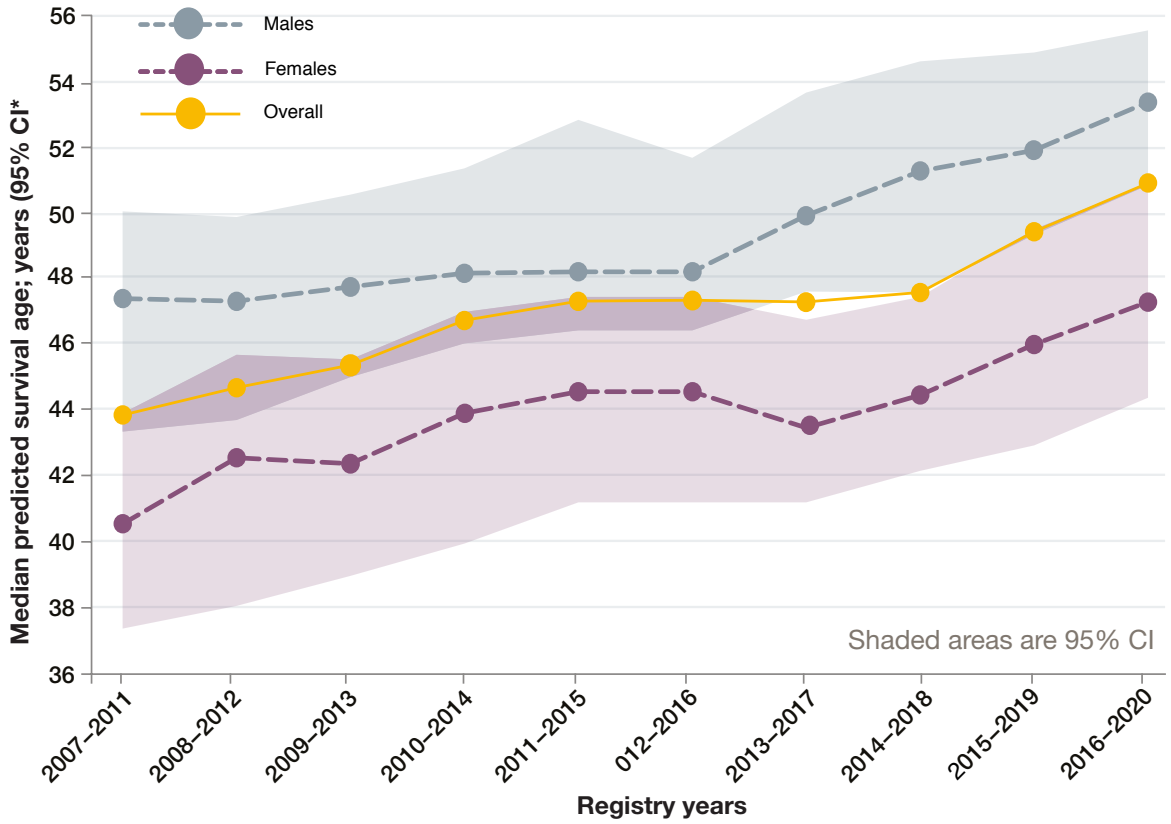
* Redacted to adhere to statistical disclosure guidelines.

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 **50.6** years. Half are therefore predicted to die before they reach that age.

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

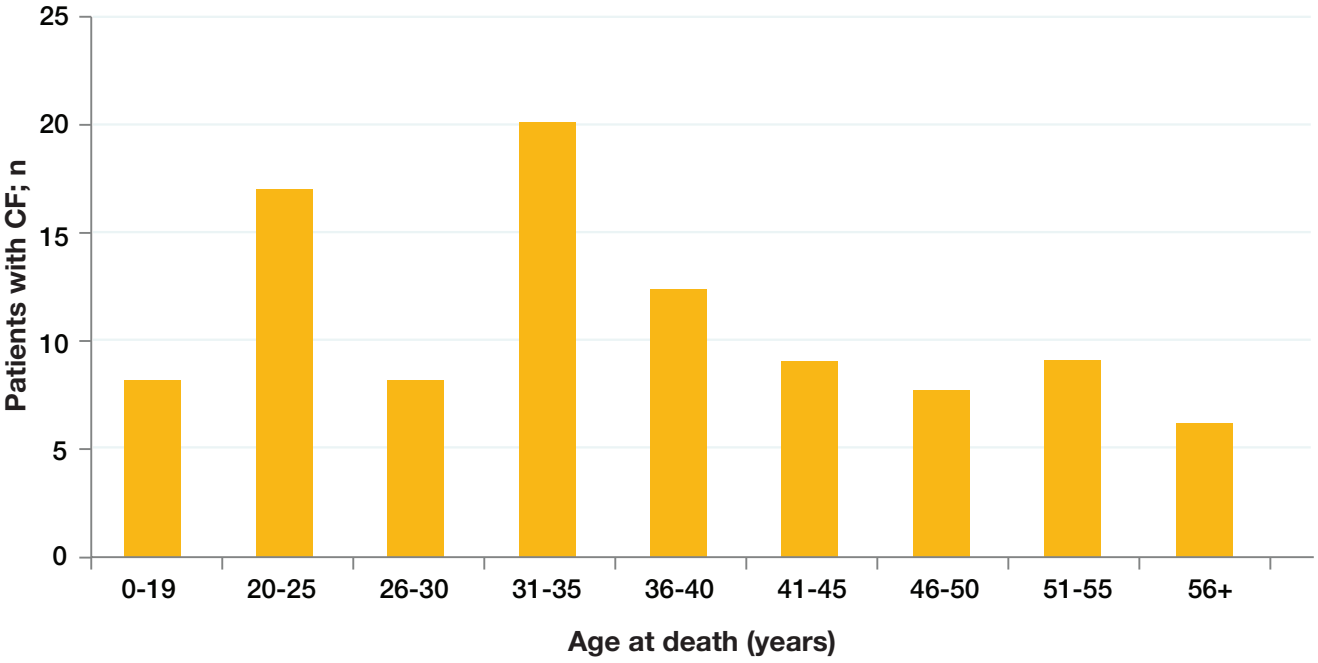


Median predicted survival age; years (95% CI*)				
Years	**Overall**	**Female**	**Male**	**p-value (males vs females)**
2007-2011	43.5 (41.9-45.9)	40.1 (36.9-43.6)	47.1 (43-49.8)	<0.001
2008-2012	44.3 (42.4-46.5)	42.2 (37.6-45.3)	47.0 (43.3-49.6)	<0.001
2009-2013	45.0 (42.8-47.0)	42.0 (38.5-45.2)	47.4 (44.7-50.3)	<0.001
2010-2014	46.4 (43.7-47.9)	43.6 (39.5-46.7)	47.9 (45.7-51.1)	<0.001
2011-2015	47.0 (44.3-48.2)	44.2 (40.8-47.1)	47.9 (46.1-52.6)	0.004
2012-2016	47.0 (44.7-48.2)	44.2 (40.8-47.1)	47.9 (46.1-51.4)	0.003
2013-2017	47.0 (44.8-48.2)	43.1 (40.8-46.4)	49.6 (47.3-53.4)	<0.001
2014-2018	47.3 (45.7-49.6)	44.1 (41.8-47.1)	51.0 (47.3-54.4)	<0.001
2015-2019	49.1 (47.0-51.4)	45.7 (42.6-49.2)	51.6 (49.0-54.6)	<0.001
2016-2020	50.6 (48.2-53.1)	47.0 (44.0-50.6)	53.1 (50.6-55.3)	p=0.004

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

1.40 Age distribution of deaths in 2020

The table below shows the ages of the 97 people with CF who died in 2020. In 2020 the median age of the 97 people who died was 36. Median age of death is based on the people with CF who died in any given year.



Age at death; years	Patients with CF; n
0-19	8
20-25	17
26-30	8
31-35	20
36-40	12
41-45	9
46-50	8
51-55	9
56+	6
Total	**97**

1.41 Causes of death

This table shows all the recorded causes of death between 2018 – 2020.

Cause of death	Patients with CF; n(%)
Respiratory/cardiorespiratory	236 (61.6)
Transplant-related	32 (8.4)
Other	29 (7.6)
Cancer	19 (5.0)
Not known	17 (4.4)
Liver disease/liver failure	12 (3.1)
Trauma or suicide	3 (0.8)
Total	**348**

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	10721 (98.9)
Patients genotyped with both mutations recorded	10417 (96.1)
F508del mutations	
Homozygous F508del	5169 (47.7)
Heterozygous F508del	4476 (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 1							
Mutation 2	**F508del**	**R117H**	**G551D**	**G542X**	**621+1G->T**	**Other**	**Unknown**	**Total**
	(%)							
F508del	47.7							**47.7**
R117H	4.9	0.1						**5.1**
G551D	4.2	0.2	0.2					**4.6**
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**
Other	26	0.6	0.9	0.7	0.5	5.3		**34**
Unknown	2	0.1	0.1	0.1	0	0.5	1.1	**3.9**
Total	**89.0**	**1.2**	**1.4**	**1.0**	**0.6**	**5.8**	**1.1**	**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, as 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	9645	89.0
c.350G->A	p.Arg117His	R117H	670	6.2
c.1652G->A	p.Gly551Asp	G551D	629	5.8
c.1624G->T	p.Gly542X	G542X	398	3.7
c.489+1G->T		621+1G->T	270	2.5
c.1585-1G->A		1717-1G->A	169	1.6
c.3909C->G	p.Asn1303Lys	N1303K	169	1.6
c.1766+1G->A		1898+1G->A	153	1.4
c.3454G->C	p.Asp1152His	D1152H	141	1.3
c.200C->T	p.Pro67Leu	P67L	141	1.3
c.3528delC	p.Lys1177SerfsX15	3659delC	115	1.1
c.3140-26A->G		3272-26A->G	109	1.0
c.1679G->C	p.Arg560Thr	R560T	102	0.9
c.1519_1521delATC	p.Ile507del	I507del	91	0.8
c.1477C->T	p.Gln493X	Q493X	89	0.8
c.1657C->T	p.Arg553X	R553X	88	0.8
c.3717+12191C->T		3849+10kbC->T	87	0.8
c.254G->A	p.Gly85Glu	G85E	80	0.7
c.178G->T	p.Glu60X	E60X	74	0.7
c.1022_1023insTC	p.Phe342HisfsX28	1154insTC	73	0.7

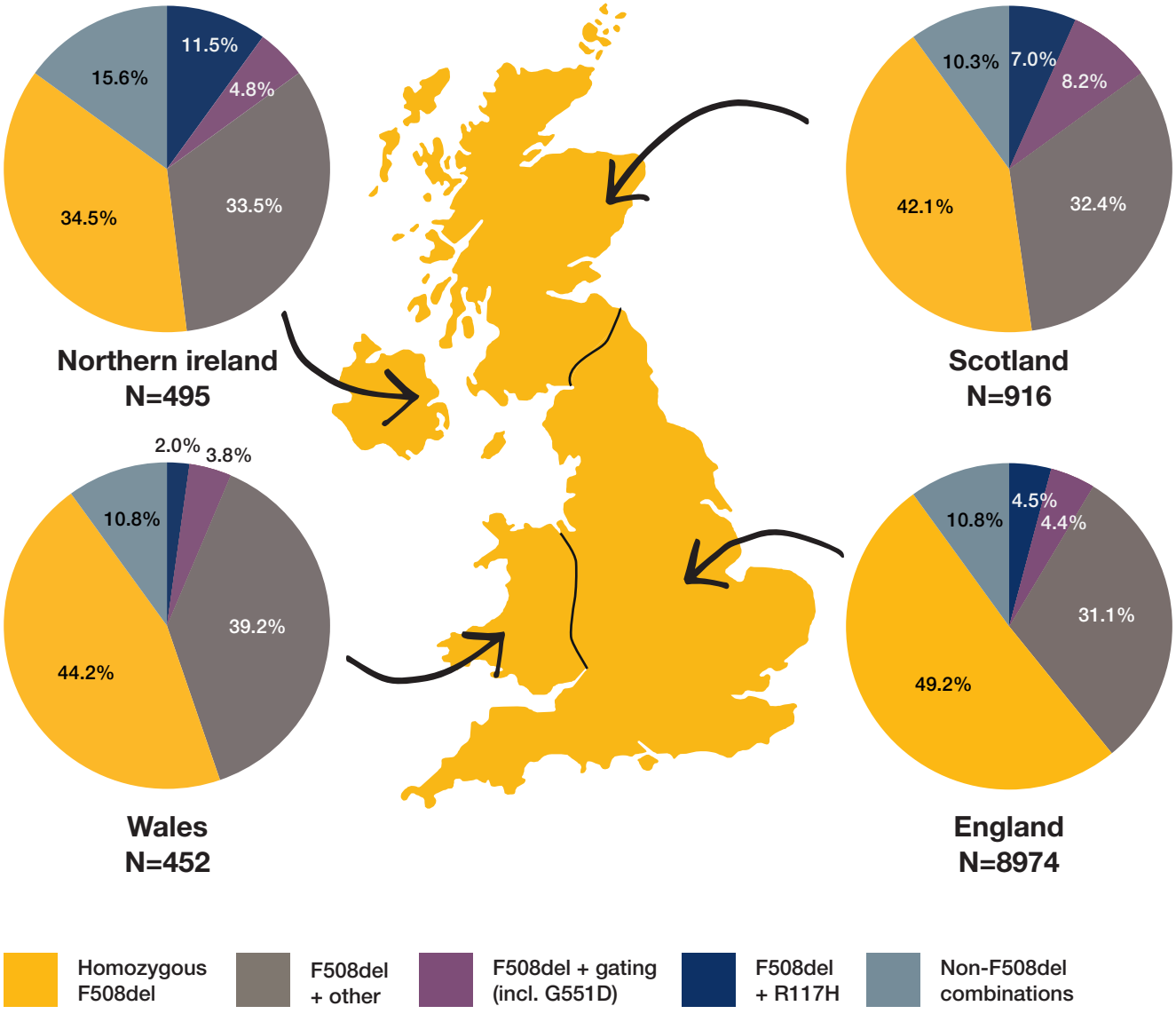
1.44 Mutation prevalence by devolved nation

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, as people with heterozygous mutations appear twice in the table.

Legacy name	England N=8974		Scotland N=916		Wales N=452		Northern Ireland N=495	
	n	%	n	%	n	%	n	%
F508del	8002	89.2	822	89.7	403	89.2	418	84.4
R117H	499	5.6	80	8.7	15	3.3	76	15.4
G551D	470	5.2	95	10.4	19	4.2	45	9.1
G542X	285	3.2	61	6.7	23	5.1	29	5.9
621+1G->T	196	2.2	11	1.2	48	10.6	15	3
1717-1G->A	150	1.7	16	1.7	<5	-	<5	-
N1303K	143	1.6	12	1.3	7	1.5	7	1.4
1898+1G->A	119	1.3	5	0.5	28	6.2	<5	-
P67L	71	0.8	49	5.3	<5	-	19	3.8
D1152H	114	1.3	15	1.6	<5	-	9	1.8

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.



Section 2 and 3: Centre-level analysis

Cystic fibrosis care in the UK is led by 57 regional centres, 4 stand-alone clinics and 72 networked clinics. The breakdown between centres and clinics delivering paediatric and adult care is shown below:

	Paediatric	Adult	Total
Centres	31	26	57
Stand-alone clinics	2	2	4
Networked clinics	65	7	72

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

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

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

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

Key

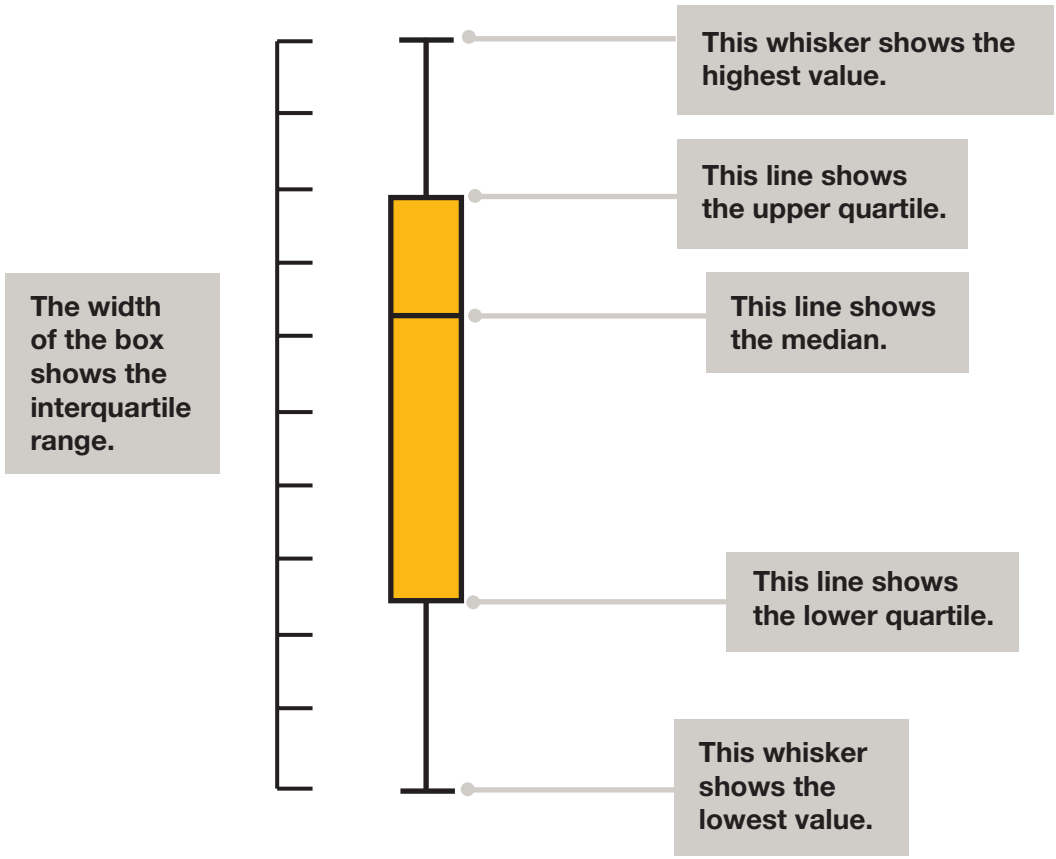
Paediatric centre

Adult centre

A guide to the charts

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

Box plots



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

Funnel plots

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

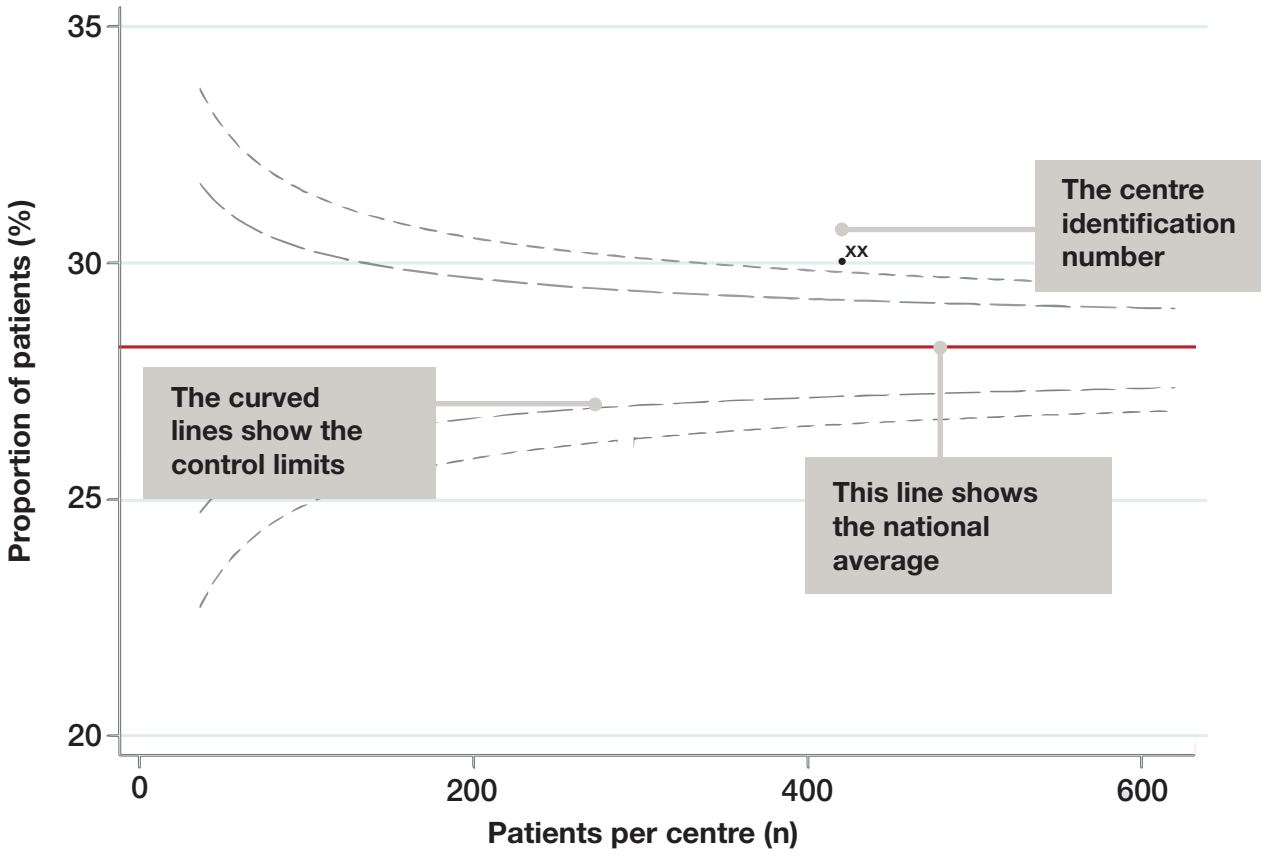
There will always be some natural variation between centres because of differences between the populations receiving care. Using only the national average as a standard can make it difficult to tell whether a survival rate that sits above the national average is higher than we would expect it to be,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

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

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

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

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



Section 2 Paediatric centre analysis

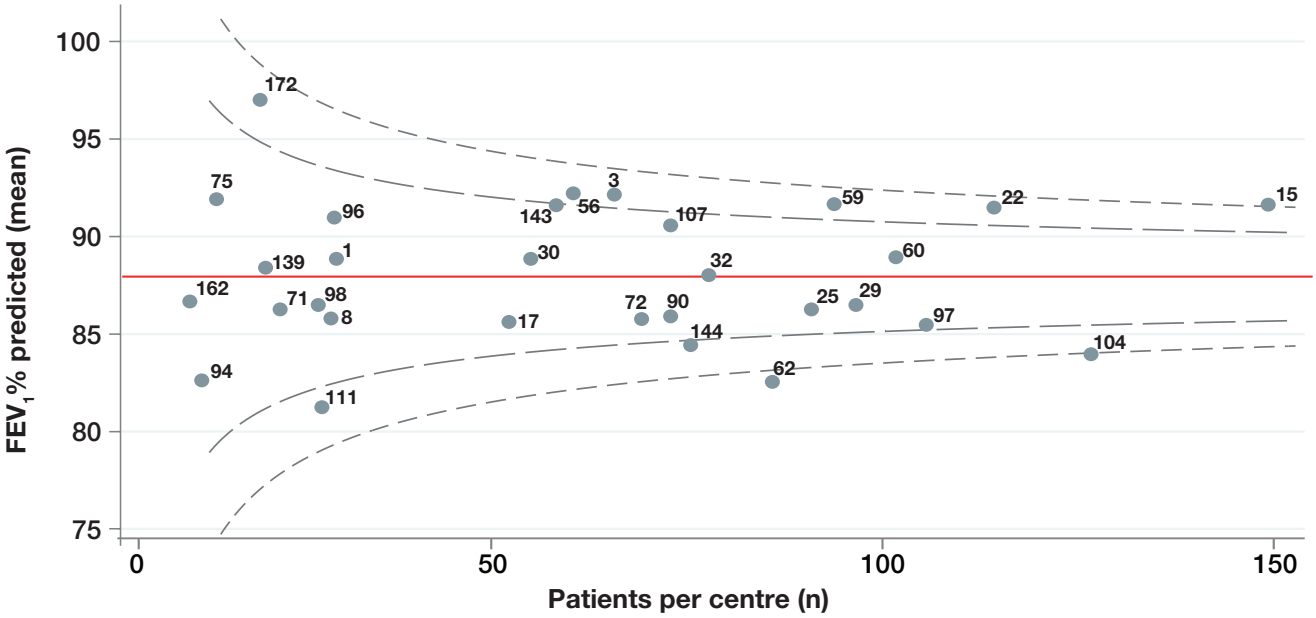
N=4231

In the UK, paediatric CF care is led by 31 specialist CF centres and two stand-alone clinics. Some paediatric centres oversee care delivered by 65 smaller, networked clinics. Data from smaller networked clinics is included in the paediatric centre's data.

Key

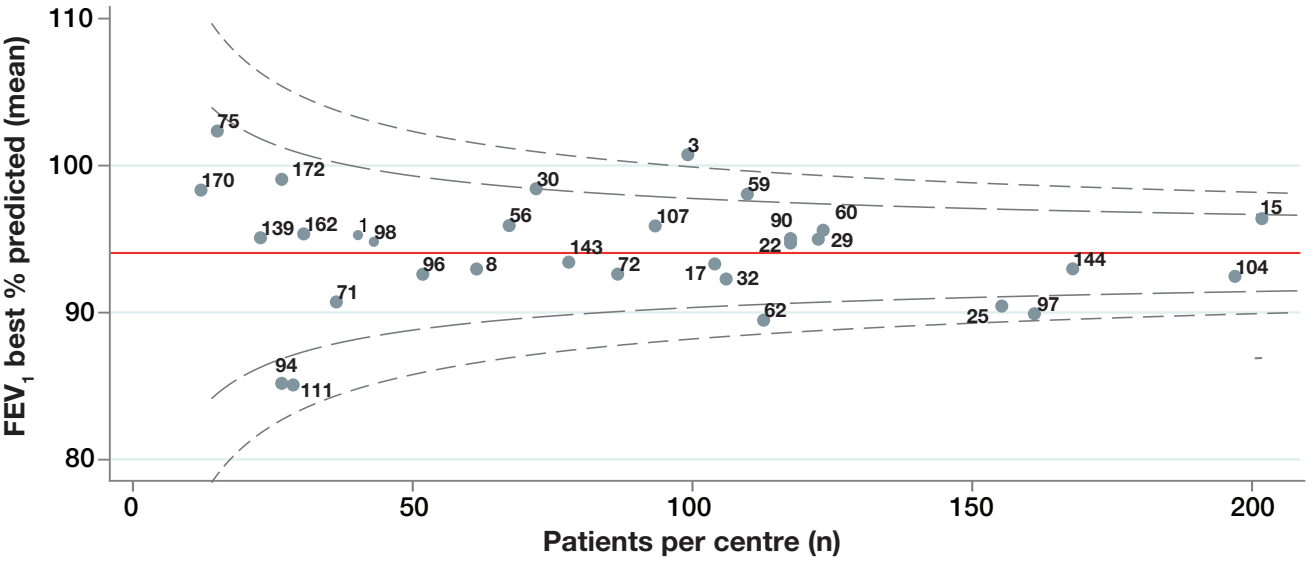
Centres with their network clinics · Stand-alone clinics · 2 standard deviations · 3 standard deviations

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



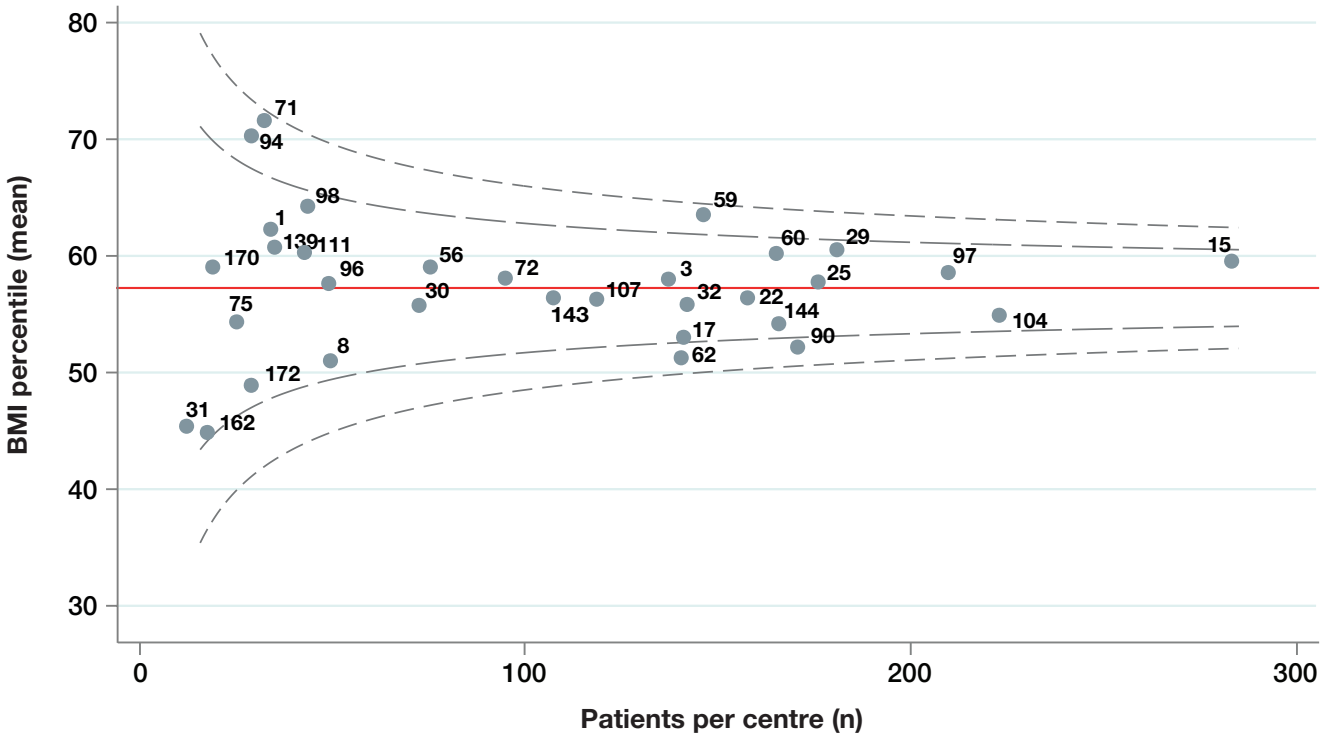
The mean FEV_1% predicted for patients attending paediatric centres/clinics is 88.6% predicted.

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



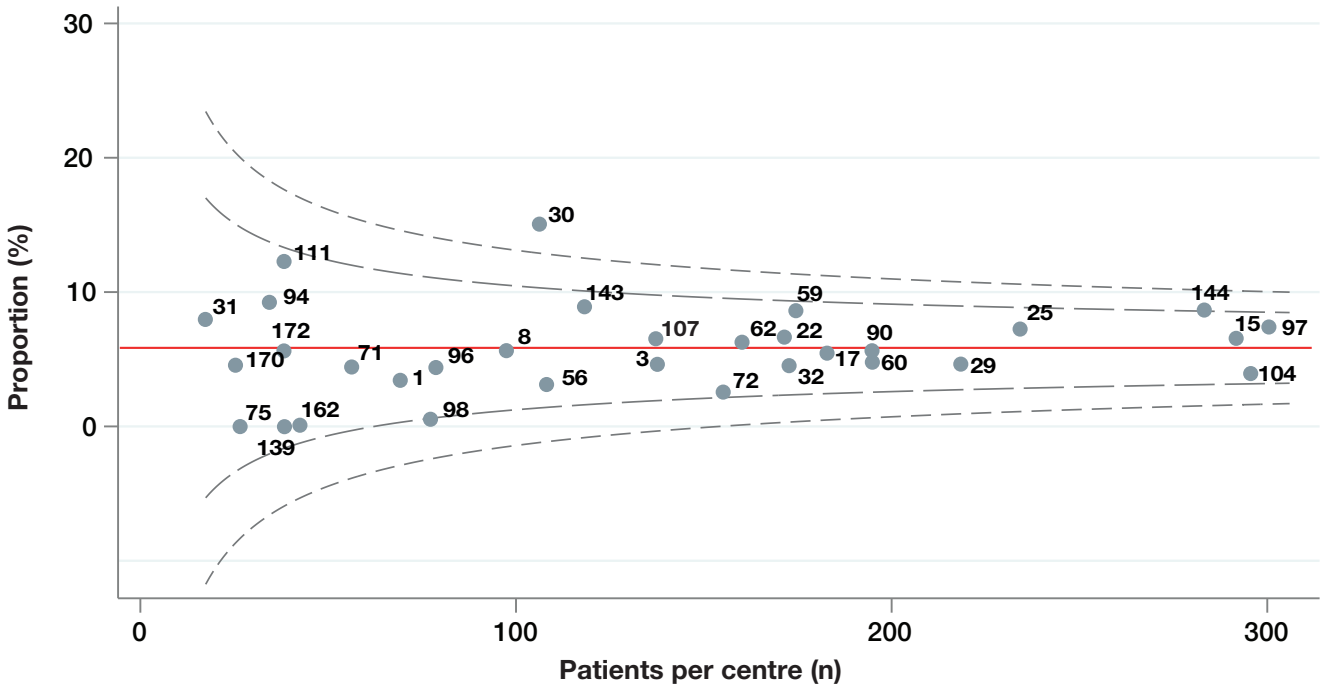
The mean Best FEV_1% predicted for patients attending paediatric centres/clinics is 93.4% predicted. Where Best FEV_1% predicted was missing, the FEV_1% predicted at annual review was used.

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



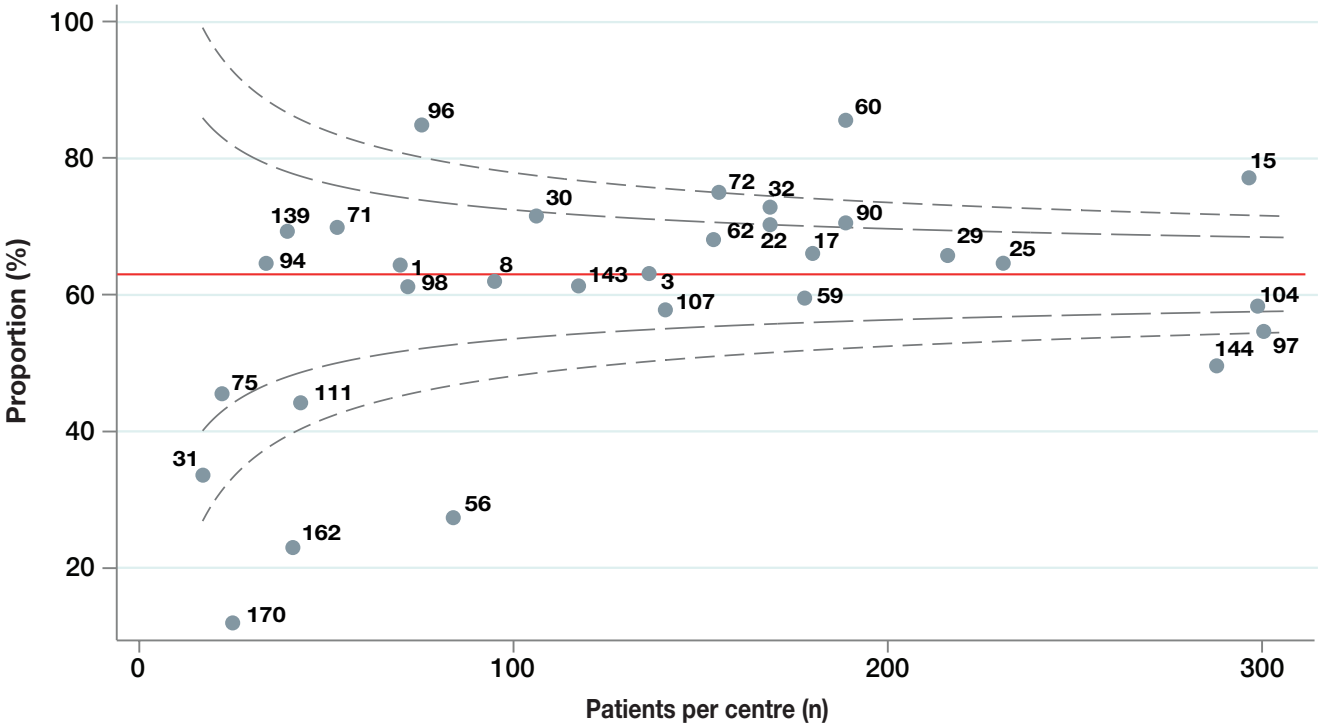
The mean BMI percentile for patients attending paediatric centres/clinics is 56.5.

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



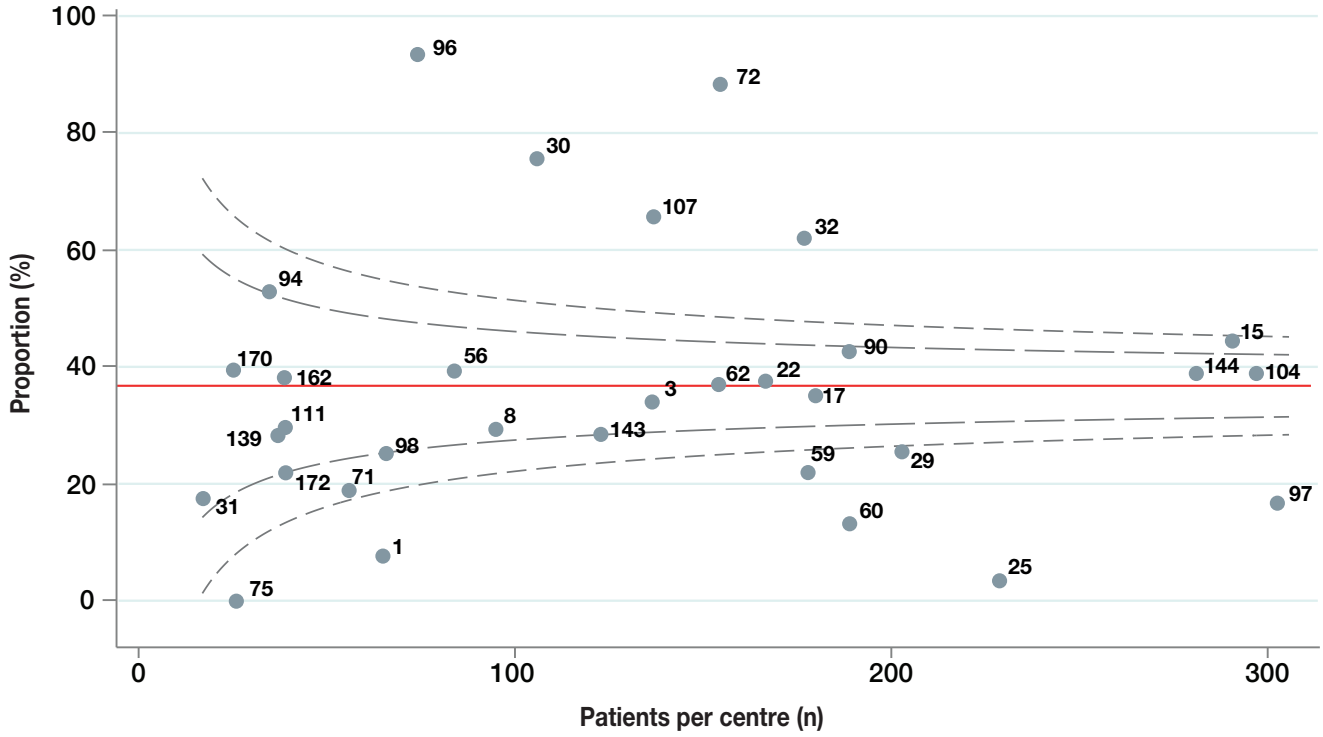
The proportion of patients with chronic *Pseudomonas aeruginosa* in paediatric centres/clinics is 5.9%.

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



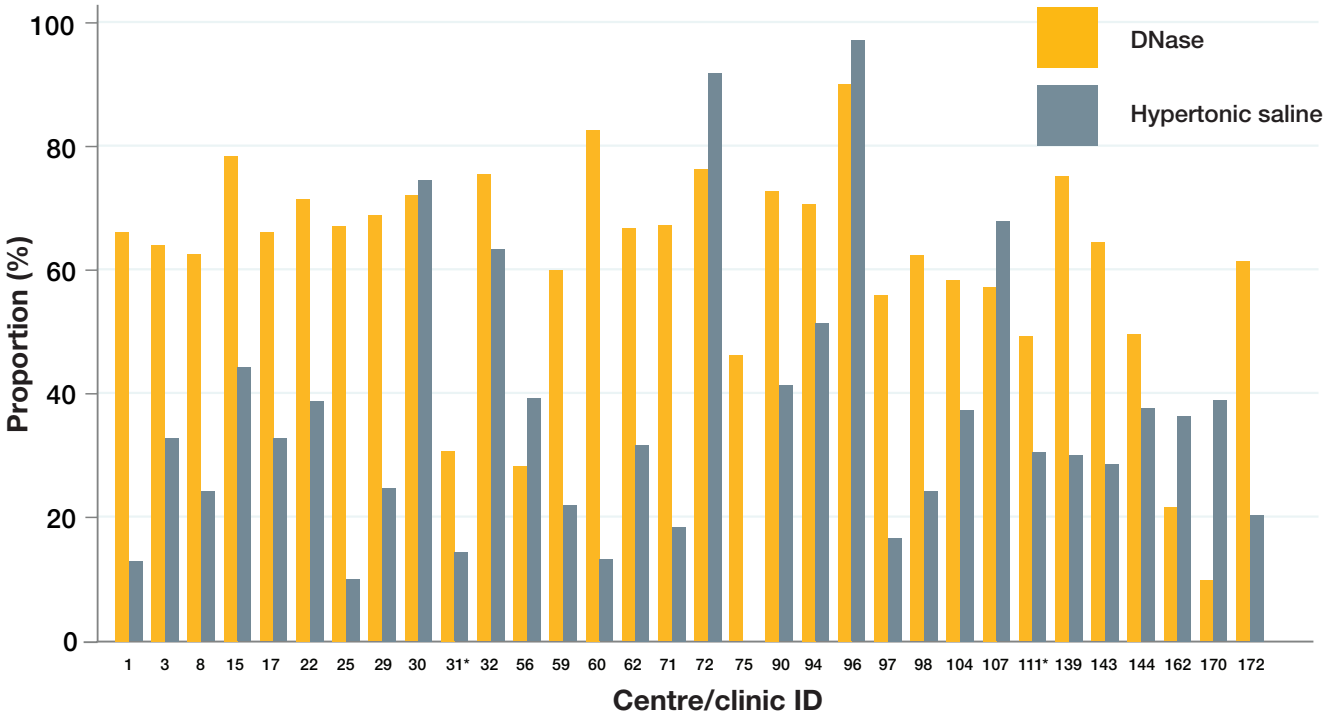
The proportion of patients receiving DNase treatment in paediatric centres/clinics is 64.2%.

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 36.8%.

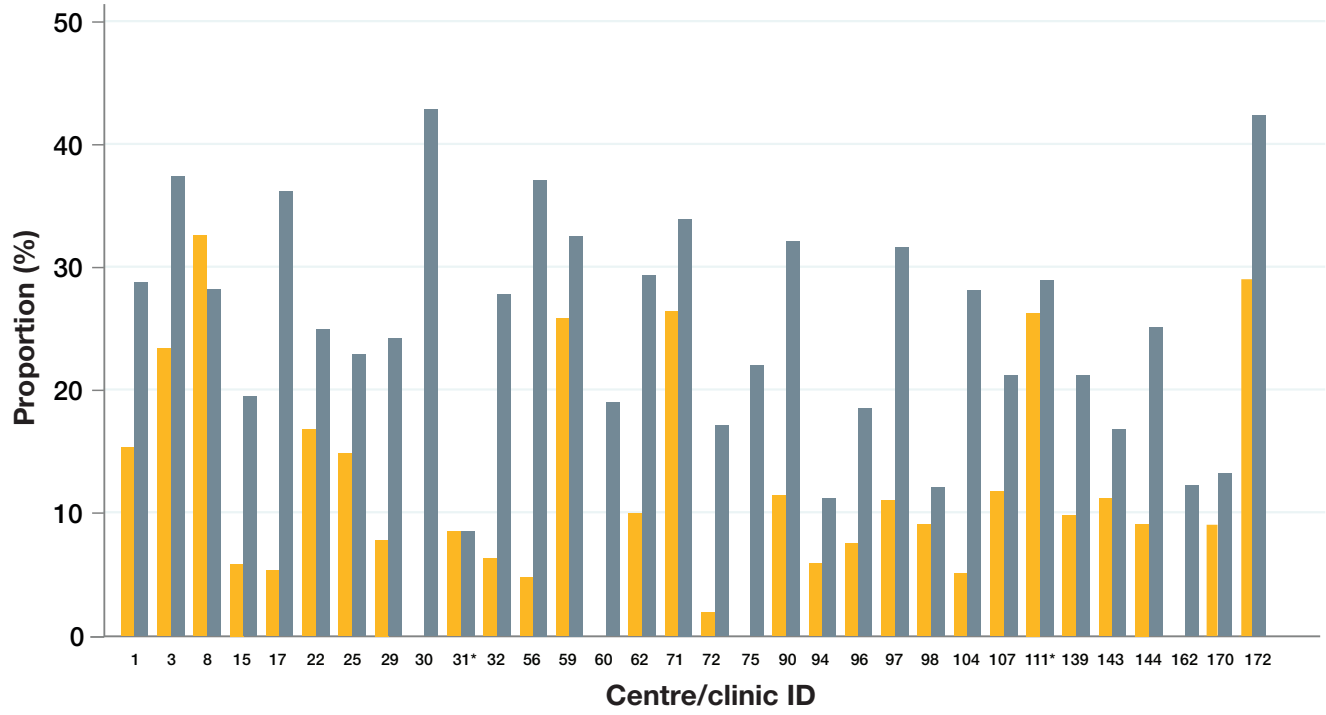
2.7 Proportion of patients receiving DNase/hypertonic saline/ mannitol treatment by paediatric centre/clinic



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.

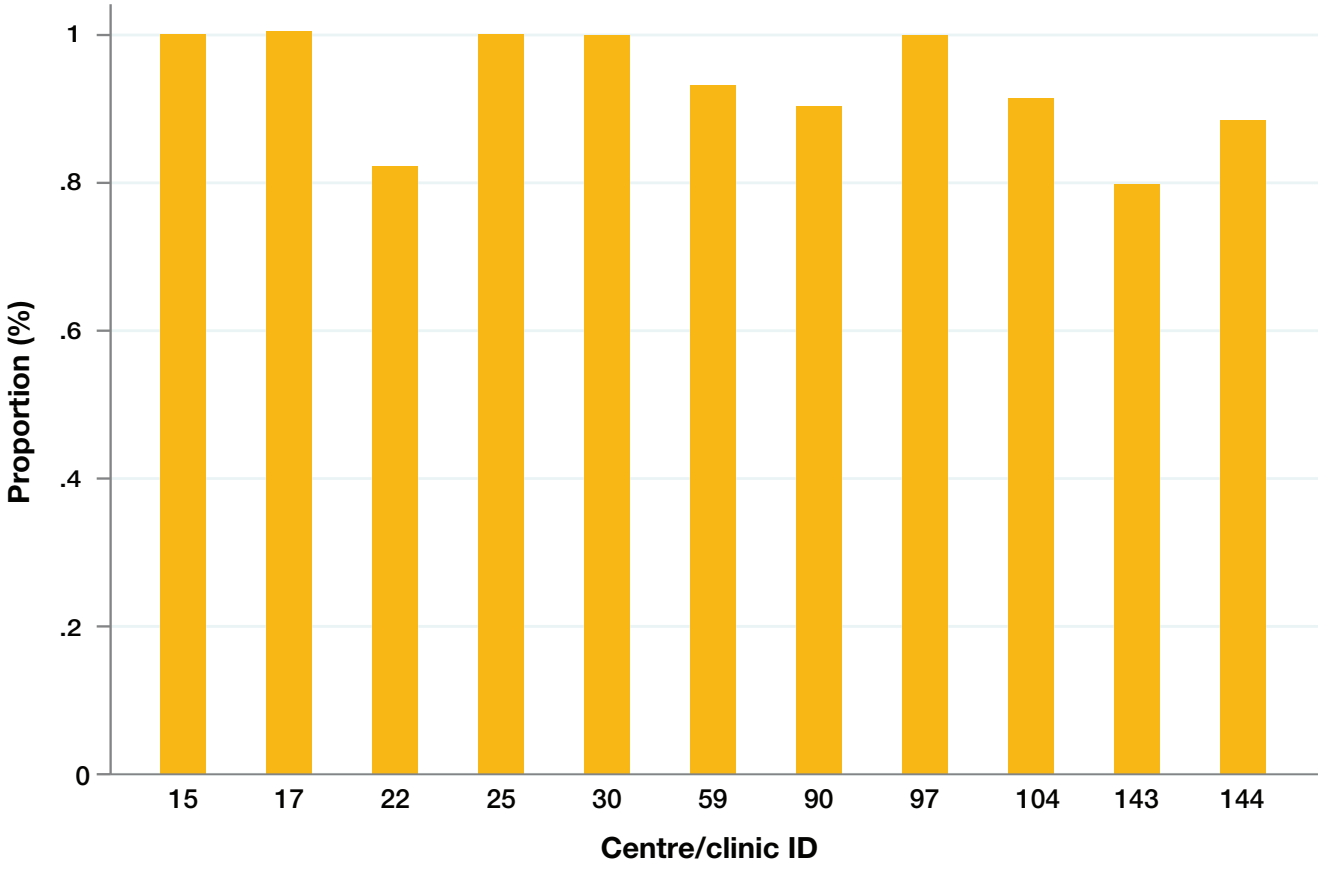


The proportion of patients receiving IVs at home was 11.2% and in hospital was 30.2%. The proportion receiving any IVs was 32.1%.

*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*.



94.4% of patients with chronic *P. aeruginosa* received inhaled antibiotics.

2.10 Data completeness

Due to the COVID-19 pandemic and prioritisation of frontline services, some sites were unable to complete data cleaning. As a result, the data completeness section has been omitted this year.

Section 3: Adult centre analysis

N=5836

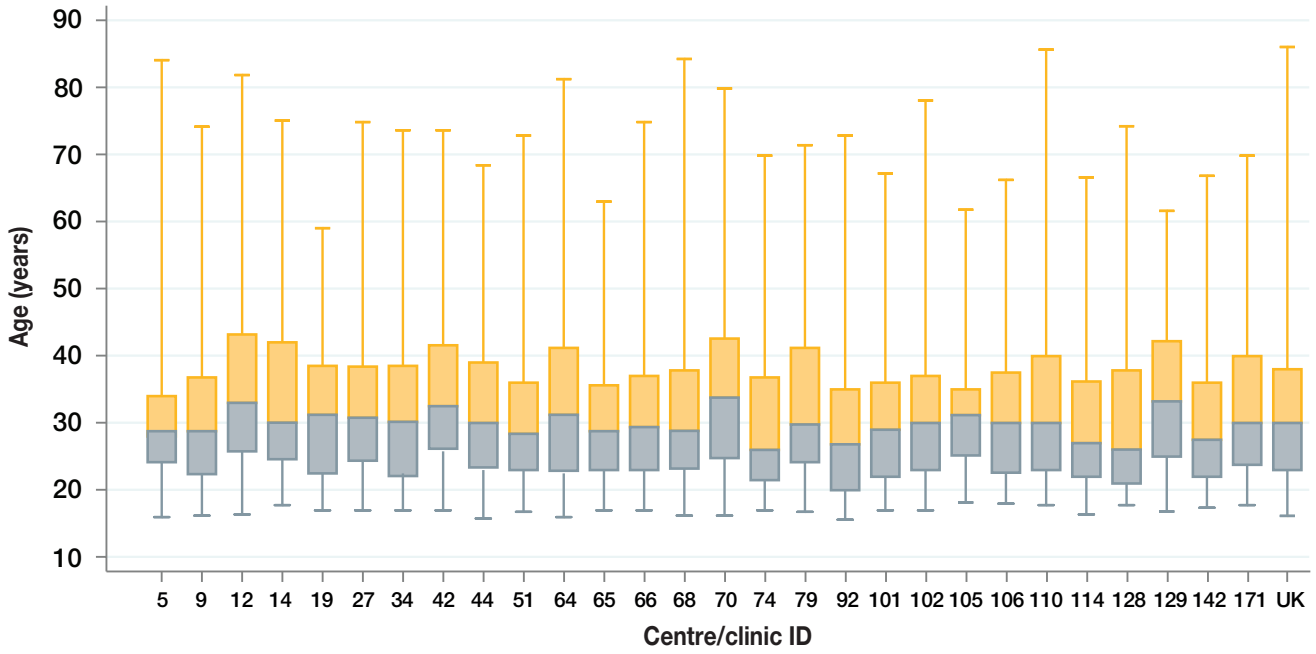
In the UK, CF care is led by 26 adult specialist CF centres. People with CF transfer to adult care centres between the ages of 16 and 18 years.

Key

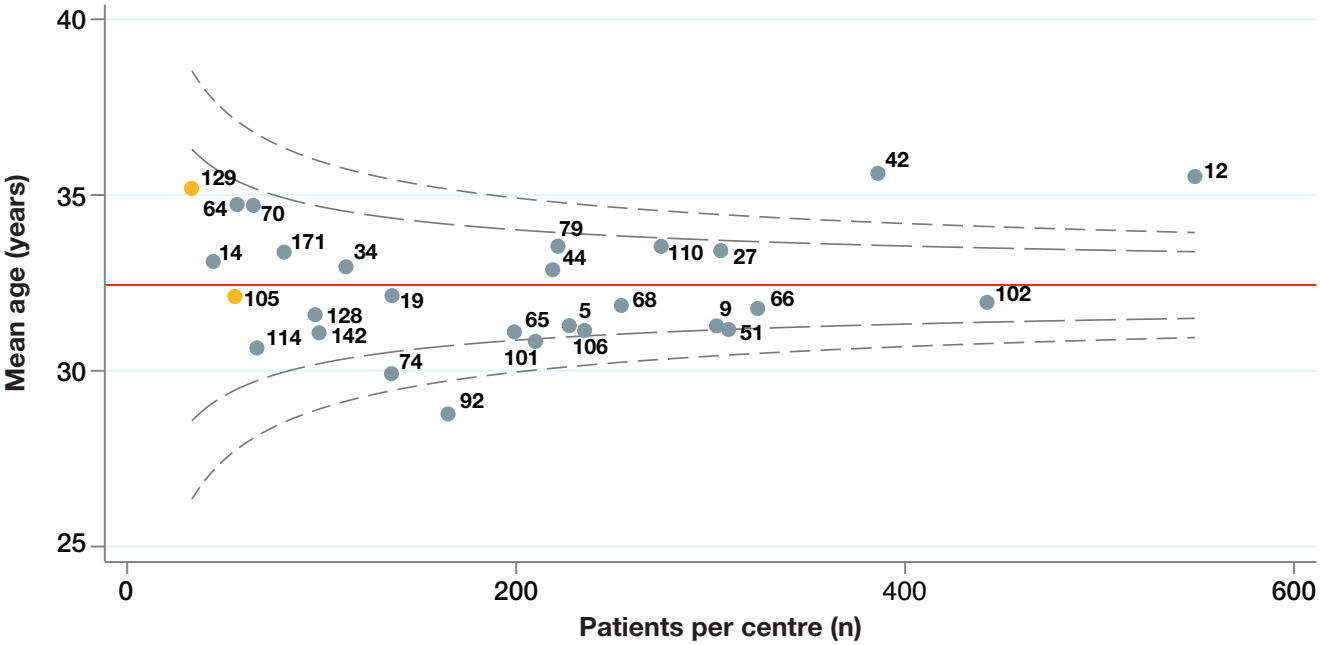
Centres with their network clinics · Stand-alone clinics · — — — 2 standard deviations · – – – 3 standard deviations

3.1 Age distribution by adults service

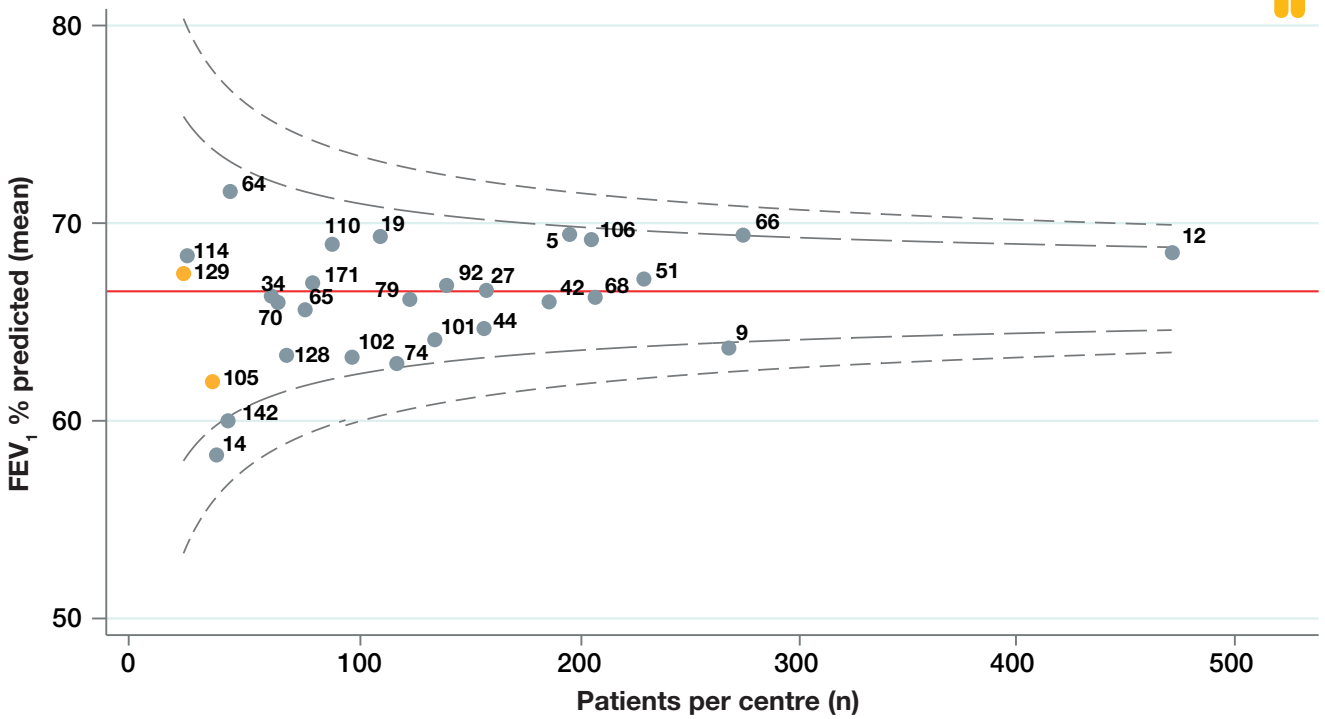
The box plot shows the age distribution of patients within each centre/clinic. In 2020 the median age in adults services was 30 years (IQR:23-38)



The funnel plot below shows how the mean age in adult centres compares to the national mean. In 2020 the national mean age of patients at CF centres was 32.5 years.

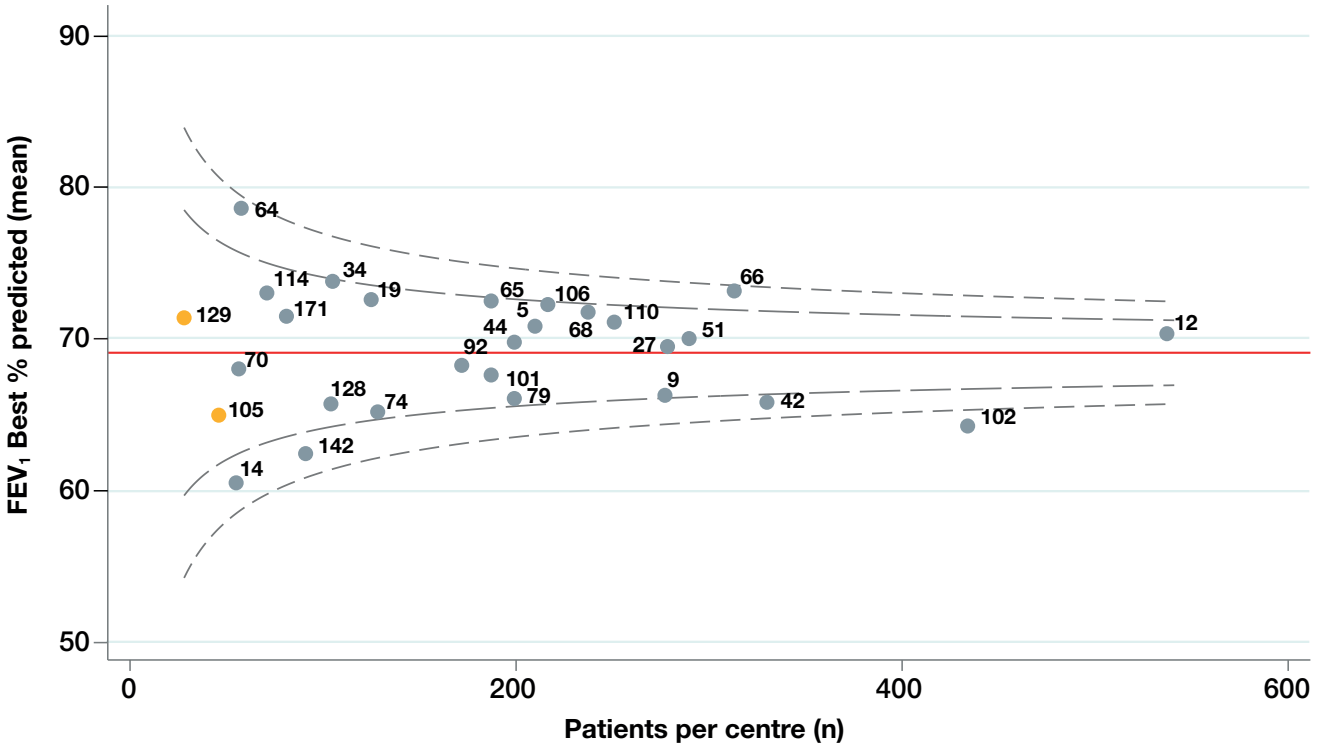


3.2 Age-adjusted FEV_1% predicted at annual review in patients without a history of lung transplant, by adult service



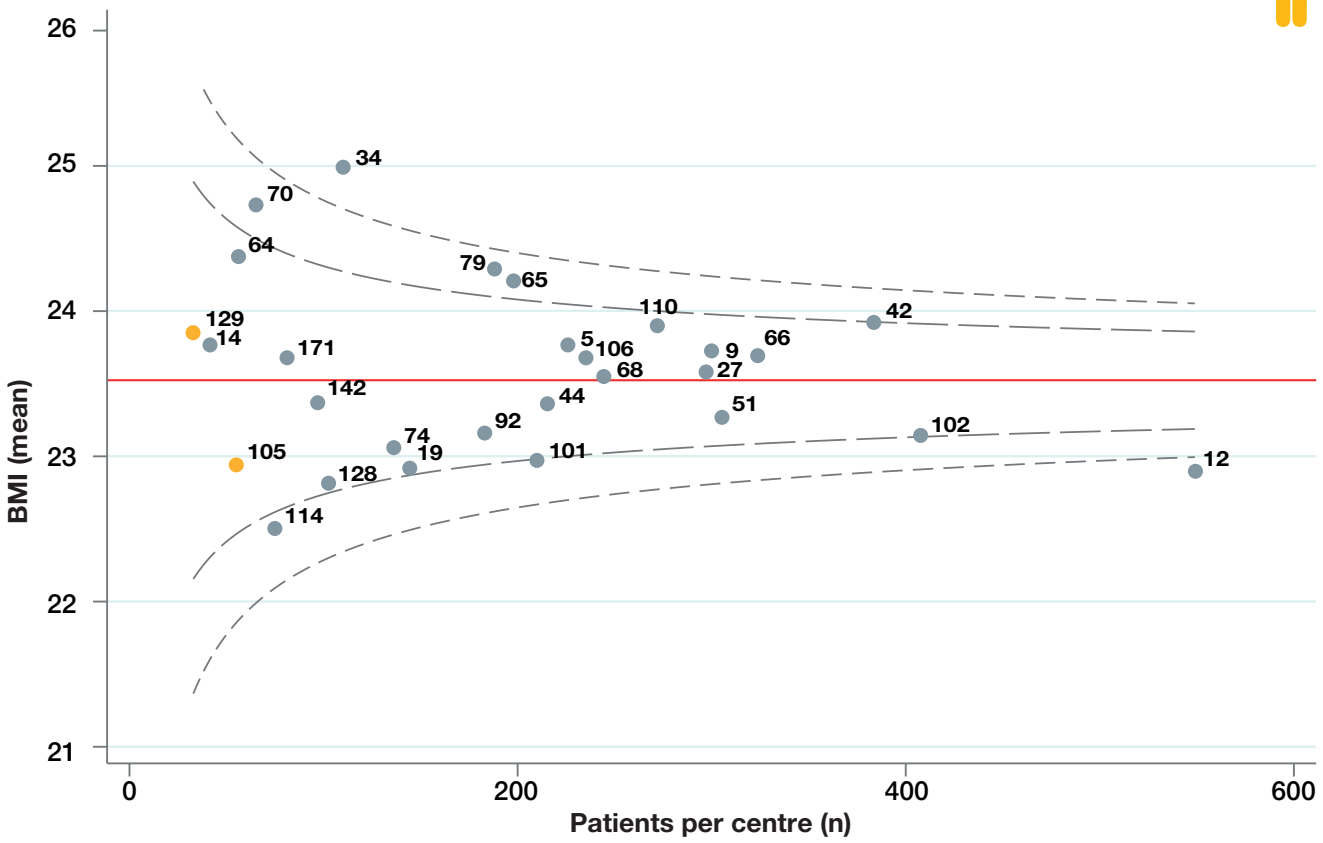
The mean FEV_1% predicted in adult services is 66.5%.

3.3 Age-adjusted Best FEV_1% predicted in patients without a history of lung transplant, by adult service



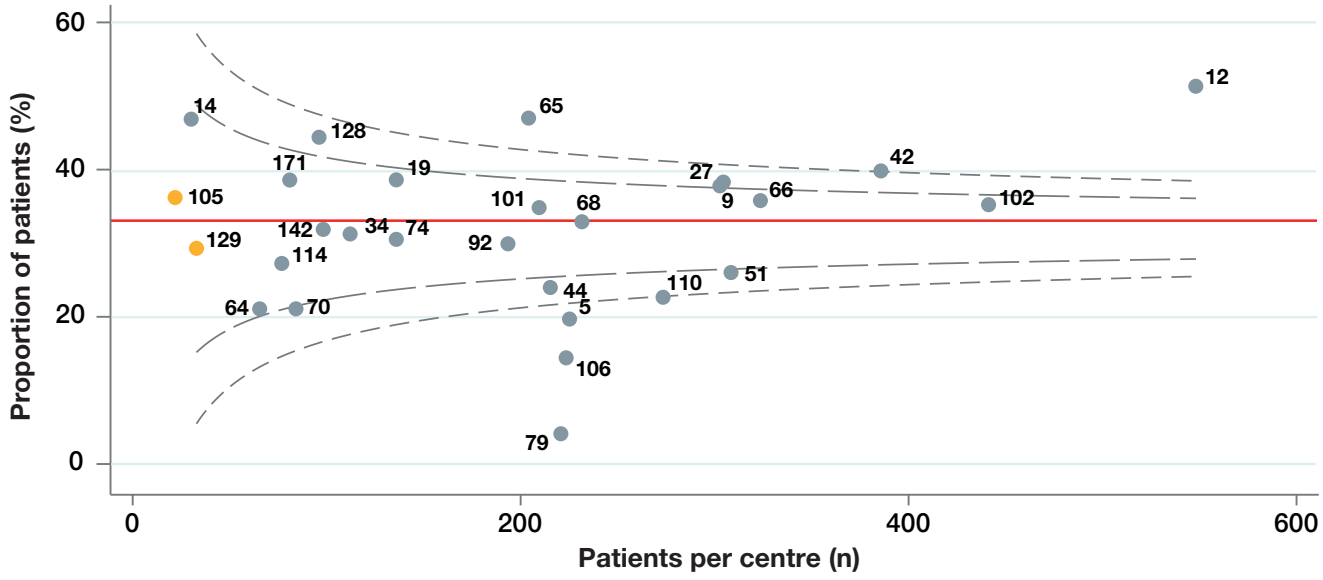
In 2020 the national mean was 69.4%. Where Best FEV_1% predicted was missing, or lower than the FEV_1 at annual review, the FEV_1% value at annual review was used.

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



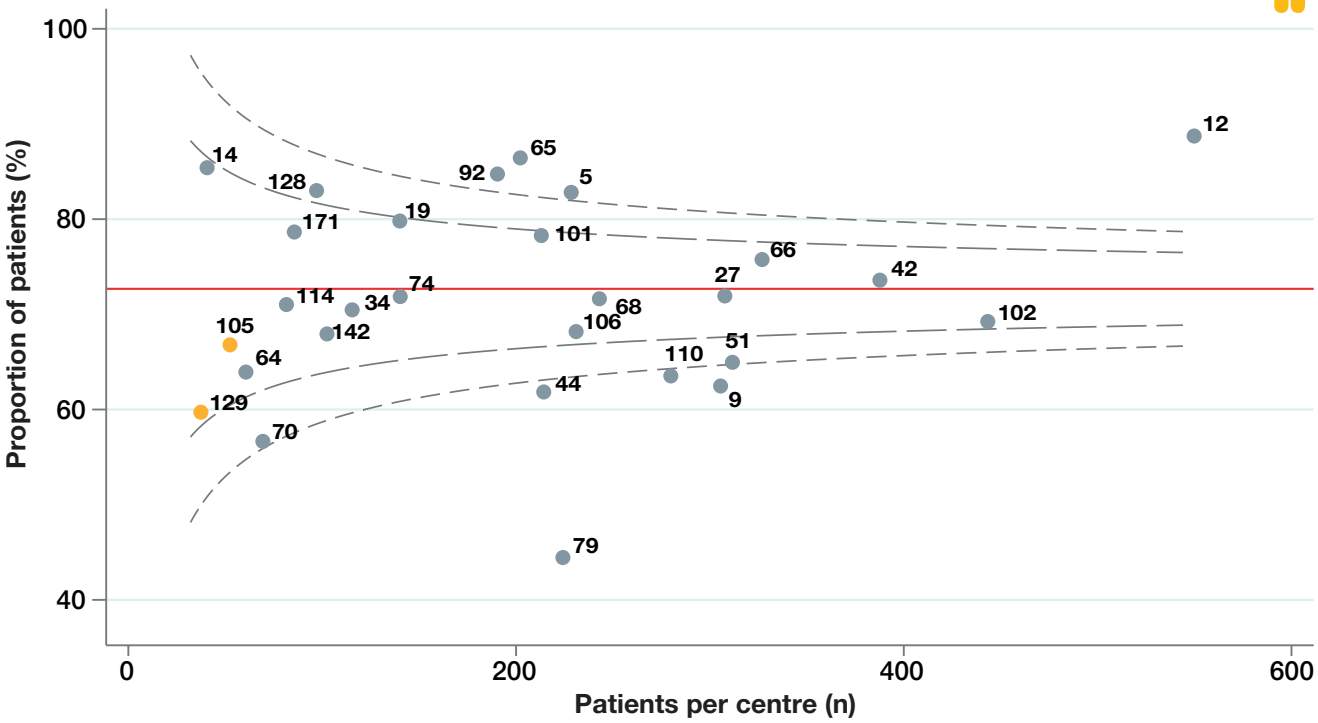
The mean BMI in adult services is 23.5.

3.5 Proportion of patients with chronic *Pseudomonas aeruginosa* by adult service



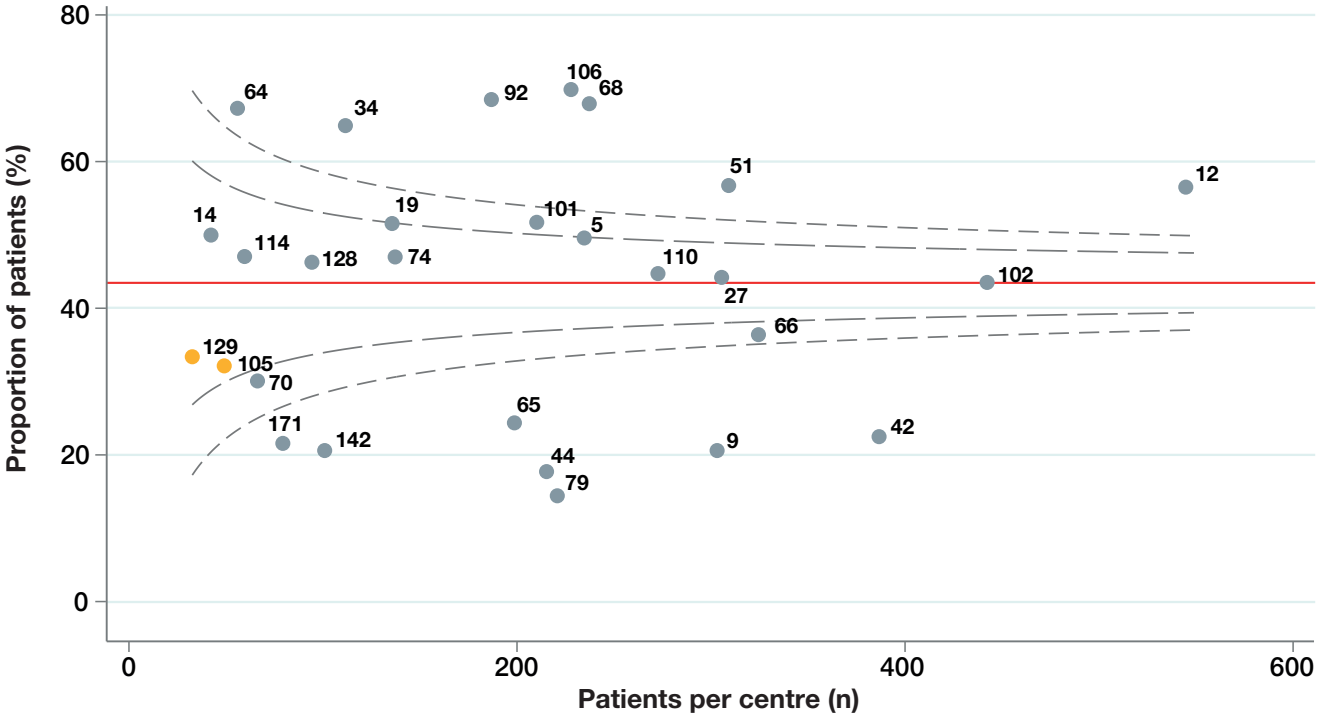
The proportion of patients with chronic *P. aeruginosa* in adult centres/clinics is 32.6%.

3.6 Proportion of patients receiving DNase treatment by adult service



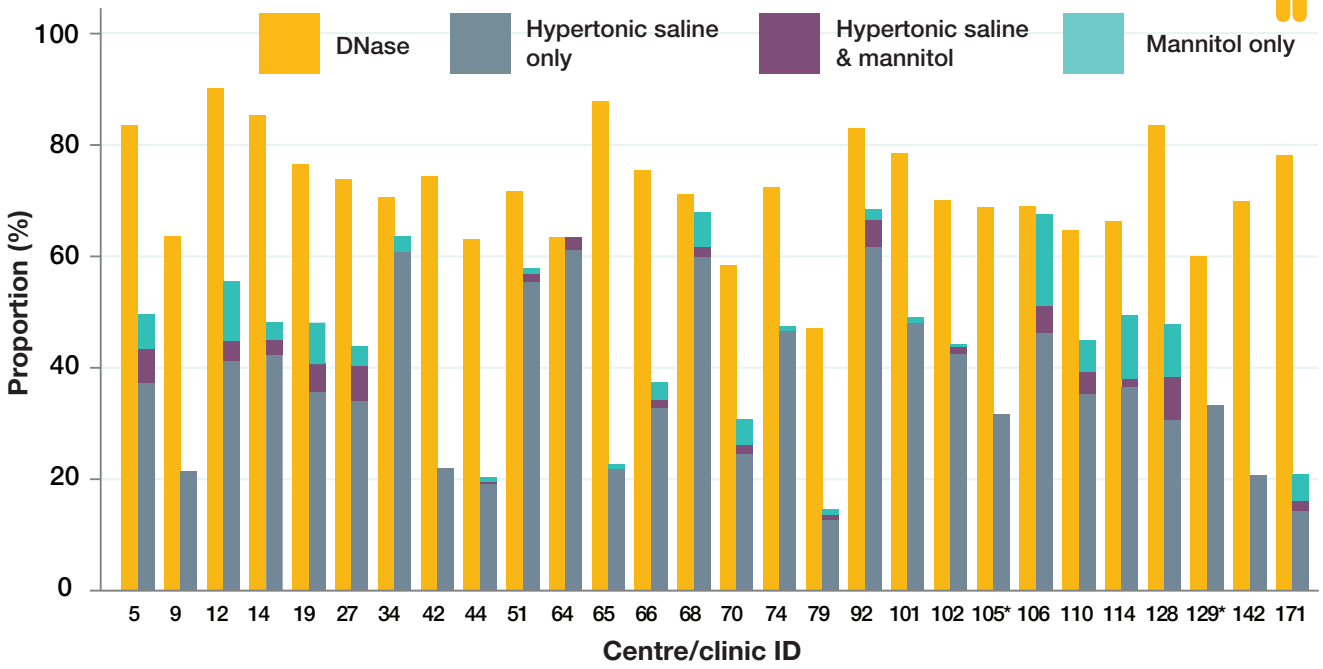
The proportion of patients receiving DNase treatment in adult centres/clinics is 73.6%.

3.7 Proportion of patients receiving hypertonic saline or mannitol by adult service



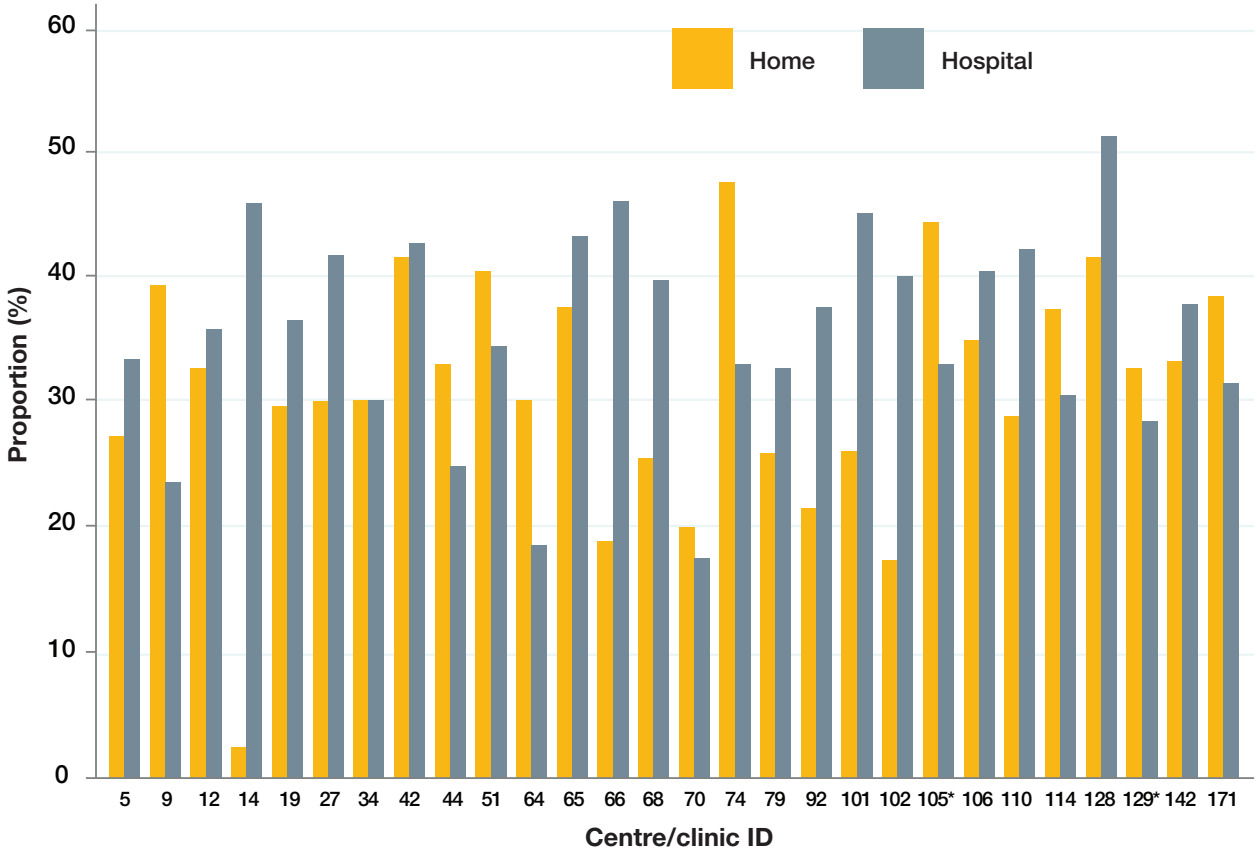
The proportion of patients receiving hypertonic saline or mannitol treatment in adult centres/clinics is 43.0%.

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



3.9 Intravenous (IV) antibiotic use by adult service

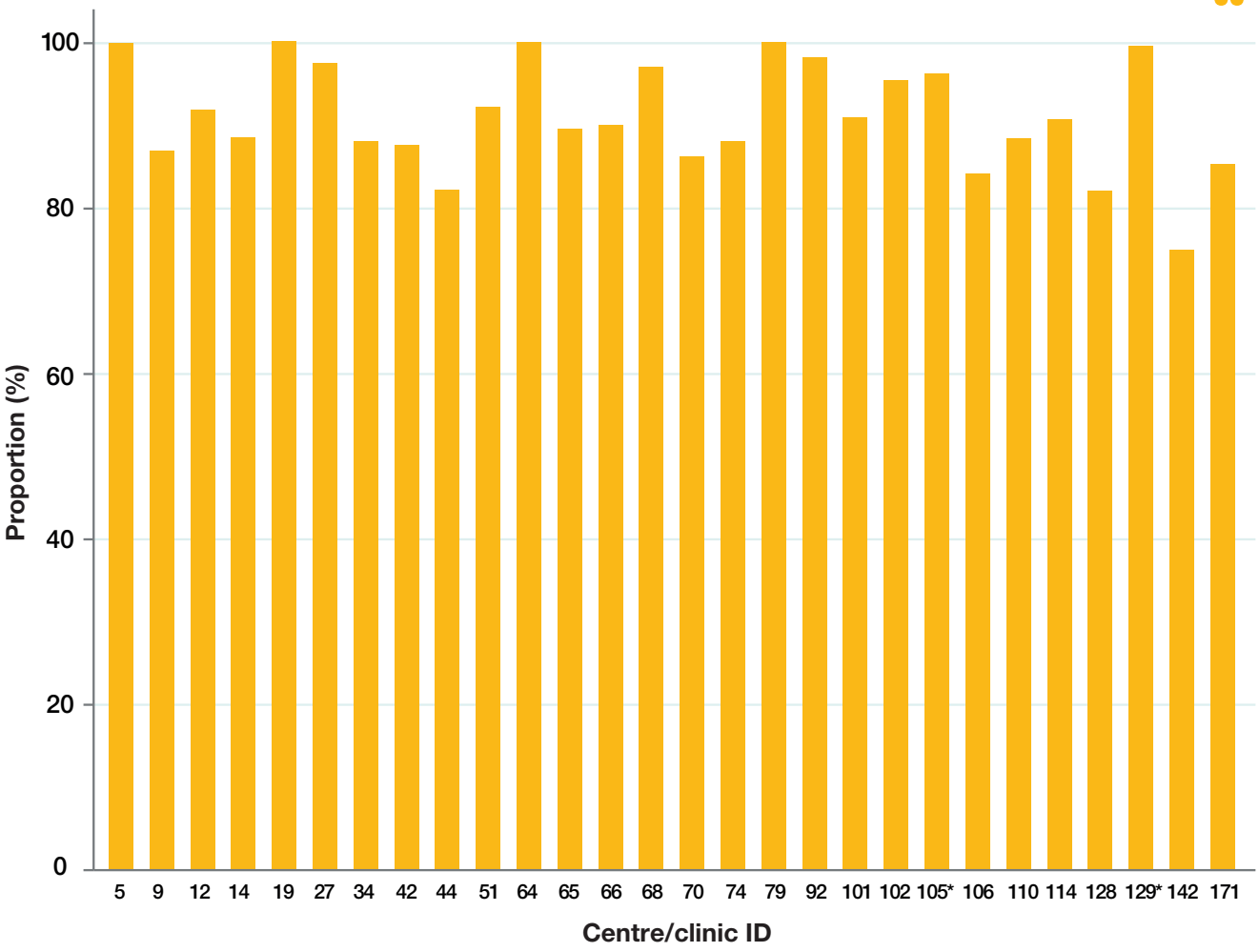
The chart below shows the proportion of patients with at least one IV day at home and/or in hospital. Patients may have a combination of home and hospital IV days.



The proportion of patients in adult centres receiving IV antibiotics at home was 30.7% and in hospital was 35.5%. The proportion receiving any IVs was 47.7%.

*Stand-alone clinics

3.10 Inhaled antibiotic use for patients with chronic *Pseudomonas aeruginosa* by adult service



91.4% 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

Due to the COVID-19 pandemic and prioritisation of frontline services, some sites were unable to complete data cleaning. As a result, the data completeness section has been omitted this year.

*Stand-alone clinics

Glossary

Word/Phrase	Meaning
2020	1 January 2020 – 31 December 2020.
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).
GERD (gastroesophageal reflux disease)	A chronic symptom of damage caused by stomach acid coming up from the stomach into the oesophagus.
GI bleed	Bleeding in the gastrointestinal tract.
GLI equations	Global Lung Initiative, the equation used for calculating FEV_1% predicted from absolute FEV_1, which takes into account age, gender, height and ethnicity.
H. influenza	*Haemophilus influenza* is a bacterium that can cause serious illness.
Haemoptysis	The coughing up of blood.
Hepatobiliary disease	A liver or biliary disorder.
Heterozygous	Everyone living with cystic fibrosis has two mutations of the gene for CFTR, one inherited from their mother and one from their father. Someone who has two different mutations is heterozygous.

Word/Phrase	Meaning
Homozygous	Everyone living with cystic fibrosis has two mutations of the gene for CFTR, one inherited from their mother and one from their father. If both mutations (or genotypes) are the same, the person is said to be homozygous.
Hypertension	High blood pressure.
Incidence	The number of people newly diagnosed with a condition in the given year.
IQR (interquartile range)	Also called the mid-spread, or middle fifty, IQR is a measure of the spread of data. It shows the difference between the upper and lower quartiles. IQR = Q3 – Q1.
Mean	A type of average, calculated by adding up all the values and dividing by the number of values.
Median	The middle number, when all numbers are arranged from smallest to largest.
Median age of death	Median age of death is based on the people with CF who died in any given year. So in 2020 the median age of the 97 people who died was 36..
Median predicted survival	A mathematical formula 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 49.1 years. Half of people are therefore predicted to die before they reach that age.
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 Steering Committee structure

UK CF Registry Steering Committee

Role	Forename	Surname	Organisation
Commissioner, England	Kathy	Blacker	NHS England
CF Physician - Paediatrics	Malcolm	Brodlie	Great North Children's Hospital, Newcastle
CF Physician – Paediatrics* #	Siobhán	Carr	Royal Brompton Hospital, London
Senior Statistician †	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
Non-consultant Grade - Paediatrics	Gwyneth	Davies	UCL & GOS Institute of Child Health, London
CF Physician - Paediatrics	Iolo	Doull	Children's Hospital for Wales, Cardiff
CF Physician - Adults	Jamie	Duckers	University Hospital, Llandough
CF Physician - Adults	Caroline	Elston	King's College Hospital, London
Registry Clinical Data Manager †	Elaine	Gunn	Cystic Fibrosis Trust
Cystic Fibrosis Centre Data Manager	Rebecca	Heise	King's College Hospital, London
Cystic Fibrosis Centre Administrator	Erin	Hodgetts	Royal Stoke University Hospital, Stoke
Person with CF	Flora	Kennedy McConnell	N/A
Medical Statistician †	Andrew	Lee	Cystic Fibrosis Trust
Medical Statistician †	Elliot	McClenaghan	Cystic Fibrosis Trust
CF Physician - Adults	Simon	Range	Glenfield Hospital, Leicester
Commissioner, Wales †	Andrea	Richards	NHS Wales
Commissioner, Scotland	David	Steele	NHS Scotland
Parent Representative	Grant	Valentine	N/A
Registry System Development Manager †	Mary	Yip	Cystic Fibrosis Trust

*Chair † Non-voting member # Caldicott guardian

UK CF Registry Research Committee

Role	Forename	Surname	Organisation
Senior Statistician †	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
Registry Clinical Data Manager †	Elaine	Gunn	Cystic Fibrosis Trust
Medical Statistician †	Andrew	Lee	Cystic Fibrosis Trust
Medical Statistician †	Elliot	McClenaghan	Cystic Fibrosis Trust
Registry Systems Development Manager †	Mary	Yip	Cystic Fibrosis Trust
CF Physician – Adults * #	Martin	Wildman	Northern General Hospital, Sheffield
Pharmacovigilance PI, CF Physician - Adults (retired)	Diana	Bilton	Royal Brompton Hospital, London
Pharmacovigilance PI, CF Physician - Paediatrics	Siobhán	Carr	Royal Brompton Hospital, London
Pharmacovigilance PI, CF Physician - Adults	Nick	Simmonds	Royal Brompton Hospital, London
Pharmacovigilance PI, CF Physician - Paediatrics	Steve	Cunningham	Royal Hospital for Sick Children, Edinburgh
Pharmacovigilance PI, CF Physician _ Adults	Dilip	Nazareth	Liverpool heart and Chest Hospital
Parent Representative	Marian	Dmochowska	N/A
Person with CF	James	Thomson	N/A

*Chair † Non-voting member # Caldicott guardian

Appendix 2: Centre-level data tables

Paediatric centres/clinics providing data in 2020 – ordered by clinic ID

Location	Name	Clinic ID	Total Active	Number with annual review *
Leicester	Leicester Royal Infirmary	1	75	65
Sheffield	Sheffield Children's Hospital	3	148	140
North West Midlands	University Hospital of North Midlands	8	97	91
London - South West	Royal Brompton Hospital	15	311	294
London - South East	King's College Hospital	17	189	181
Oxford	John Radcliffe Hospital	22	180	171
Leeds	St James's University Hospital	25	234	225
Southampton	Southampton General Hospital	29	228	211
London - East	Royal London Hospital	30	108	101
Inverness	Raigmore Hospital	31	16	13
Bristol	Bristol Royal Hospital for Children	32	179	171
Glasgow	Royal Hospital for Children	56	118	104
Newcastle	Great North Children's Hospital	59	190	173
Belfast	Royal Belfast Hospital for Sick Children	60	205	188
Nottingham	Nottingham University Hospitals	62	173	161
Teeside	James Cook University Hospital	71	52	49
Cardiff	Children's Hospital for Wales	72	176	157
Dundee	Ninewells Hospital	73	20	8
Aberdeen	Royal Aberdeen Children's Hospital	75	29	24
London - Central	Great Ormond Street Hospital for Children	90	196	188
Cornwall	Royal Cornwall Hospital	94	37	36
Exeter	Royal Devon & Exeter Hospital	96	75	74
Liverpool	Alder Hey Children's Hospital	97	320	303
Norwich	Norfolk & Norwich University Hospital	98	70	68
Birmingham	Birmingham Children's Hospital	104	311	298
Cambridge	Addenbrookes Hospital	107	153	141
Hull	Hull University Teaching Hospitals	111	43	42
Plymouth	Derriford Hospital	139	139	39
Edinburgh	Royal Hospital for Sick Children	143	142	117
Manchester	Royal Manchester Children's Hospital	144	342	287
Lanarkshire	Wishaw General Hospital	162	46	40
Ayr	University Hospital Crosshouse	170	25	23
Brighton	Royal Alexandra Children's Hospital	172	47	38

Age (years)		FEV$_1$% predicted at annual review				Best FEV$_1$% predicted			
Mean	Median	Number	Mean - unadjusted	Mean - adjusted	Median	Number*	Mean - unadjusted	Mean - adjusted	Median
8.5	8.5	30	88.4	89.1	93.2	40	94.2	94.6	94.7
9.1	9.0	65	92.8	92.9	93.7	101	98.8	98.8	99.3
9.2	9.9	29	86.9	86.4	86.8	62	91.9	92.3	94.4
9.1	9.0	149	92.8	92.8	93.1	207	95.6	95.6	97.6
8.1	7.1	53	85.9	85.6	89.4	106	92.9	92.9	94.8
9.5	9.7	117	91.5	92.1	94.8	120	93.6	94.4	96.6
9.1	9.2	93	87.1	86.7	91.6	159	90.8	90.6	93.5
9.0	9.0	98	86.9	87.3	89.5	125	93.6	94.5	95.1
10.1	10.7	56	88.3	89.2	89.2	73	96.1	97.1	97.8
10.0	11.0	8	85.3	84.8	85.3	9	87.2	87.0	86.6
8.7	8.8	79	88.4	88.5	90.6	108	92.0	92.0	94.1
9.2	9.8	61	92.1	92.5	94.8	68	94.6	95.1	95.8
8.3	8.1	96	92.9	92.1	93.7	112	97.1	96.6	97.6
9.3	9.4	104	88.8	89.3	90.0	126	94.3	94.8	94.5
10.0	10.0	87	82.3	83.1	84.4	115	88.7	89.4	89.4
10.3	10.2	23	84.8	86.8	82.1	36	89.0	90.4	87.5
9.2	9.8	71	85.8	86.5	83.8	88	91.3	92.2	92.2
9.2	10.0	<5	79.2	81.4	79.2	<5	88.0	88.3	88.2
7.1	7.0	13	94.2	92.9	90.0	14	102.2	100.8	101.2
8.4	8.3	73	86.4	86.7	88.4	120	95.1	94.6	96.1
9.7	9.3	13	80.9	83.0	82.3	26	85.7	86.0	89.5
9.5	9.0	30	90.9	91.3	89.1	52	91.4	91.9	90.5
9.0	8.7	108	85.2	85.9	86.6	165	89.7	90.1	89.3
9.1	10.1	28	85.4	86.8	87.4	43	92.5	94.0	89.1
9.2	8.8	129	83.5	84.6	85.6	202	91.3	92.0	94.0
8.7	9.0	74	91.8	91.2	95.4	95	95.3	94.9	95.7
8.6	8.6	28	82.3	81.7	90.3	28	86.3	85.9	91.3
7.5	8.0	21	89.7	88.7	88.0	22	95.3	94.4	97.6
9.8	9.9	58	91.8	92.1	93.4	79	92.8	93.2	94.2
9.3	9.3	76	84.4	85.1	86.8	172	92.1	92.7	95.9
9.2	9.1	11	89.4	87.3	91.1	30	95.0	94.7	95.4
9.2	8.8	9	89.0	88.1	86.9	11	99.7	98.5	95.7
9.2	9.4	20	97.5	98.0	97.2	26	97.9	98.7	98.4

* Annual review data may represent a construct derived from a recorded clinic encounter
** Where 'Best' values were missing, or lower than FEV$_1$% predicted taken at annual review, the annual review value was used.

			BMI percentile			
Location	Name	Clinic ID	Number	Mean - unadjusted	Mean - adjusted	Median
Leicester	Leicester Royal Infirmary	1	39	60.7	61.5	64.0
Sheffield	Sheffield Children's Hospital	3	124	57.1	57.0	57.6
North West Midlands	University Hospital of North Midlands	8	50	50.5	50.6	47.8
London - South West	Royal Brompton Hospital	15	272	58.9	58.7	60.0
London - South East	King's College Hospital	17	127	52.9	52.5	50.4
Oxford	John Radcliffe Hospital	22	158	55.6	55.7	56.8
Leeds	St James's University Hospital	25	173	57.3	57.0	55.5
Southampton	Southampton General Hospital	29	177	59.3	59.2	62.2
London - East	Royal London Hospital	30	71	53.9	54.4	59.6
Inverness	Raigmore Hospital	31	12	46.1	46.1	39.8
Bristol	Bristol Royal Hospital for Children	32	147	55.5	55.2	56.9
Glasgow	Royal Hospital for Children	56	75	57.7	58.1	56.8
Newcastle	Great North Children's Hospital	59	144	62.9	62.5	70.5
Belfast	Royal Belfast Hospital for Sick Children	60	164	59.0	59.0	62.8
Nottingham	Nottingham University Hospitals	62	127	50.3	50.7	52.4
Teeside	James Cook University Hospital	71	36	69.5	70.8	71.9
Cardiff	Children's Hospital for Wales	72	89	56.9	57.1	56.5
Dundee	Ninewells Hospital	73	6	51.8	51.6	59.0
Aberdeen	Royal Aberdeen Children's Hospital	75	22	55.6	54.2	60.2
London - Central	Great Ormond Street Hospital for Children	90	167	51.7	51.4	52.9
Cornwall	Royal Cornwall Hospital	94	32	70.2	70.4	75.1
Exeter	Royal Devon & Exeter Hospital	96	48	57.0	56.9	56.2
Liverpool	Alder Hey Children's Hospital	97	200	57.7	57.9	58.5
Norwich	Norfolk & Norwich University Hospital	98	43	62.3	62.6	65.9
Birmingham	Birmingham Children's Hospital	104	208	53.6	53.8	51.1
Cambridge	Addenbrookes Hospital	107	108	56.1	55.9	57.1
Hull	Hull University Teaching Hospitals	111	38	61.1	60.9	62.6
Plymouth	Derriford Hospital	139	34	60.9	60.1	63.5
Edinburgh	Royal Hospital for Sick Children	143	100	55.8	56.0	57.3
Manchester	Royal Manchester Children's Hospital	144	165	52.8	53.2	55.4
Lanarkshire	Wishaw General Hospital	162	24	46.3	45.7	42.3
Ayr	University Hospital Crosshouse	170	15	59.0	58.4	70.3
Brighton	Royal Alexandra Children's Hospital	172	30	48.3	48.3	43.8

Chronic *P. aeruginosa*		Having at least 1 IV day		Receiving DNase treatment		Receiving hypertonic saline/mannitol treatment		Inhaled antibiotic use among patients with chronic *P. aeruginosa*	
Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)
39	60.7	61.5	64.0	43	66.2	5	7.7	<5	100.0
124	57.1	57.0	57.6	86	63.2	40	29.4	5	100.0
50	50.5	50.6	47.8	64	67.4	23	24.2	<5	80.0
272	58.9	58.7	60.0	223	72.9	111	36.3	26	100
127	52.9	52.5	50.4	119	66.1	59	32.8	15	88.2
158	55.6	55.7	56.8	118	71.5	62	37.6	9	100.0
173	57.3	57.0	55.5	142	62.0	8	3.5	7	87.5
177	59.3	59.2	62.2	139	68.8	44	21.8	5	100.0
71	53.9	54.4	59.6	76	71.7	80	75.5	15	93.8
12	46.1	46.1	39.8	<5	11.8	<5	11.8	0	0.0
147	55.5	55.2	56.9	126	71.2	87	49.2	11	100.0
75	57.7	58.1	56.8	23	27.4	33	39.3	<5	100.0
144	62.9	62.5	70.5	99	55.6	39	21.9	13	86.7
164	59.0	59.0	62.8	162	85.7	25	13.2	11	100.0
127	50.3	50.7	52.4	104	65.0	46	28.8	10	100.0
36	69.5	70.8	71.9	40	75.5	13	24.5	<5	100.0
89	56.9	57.1	56.5	123	79.4	119	76.8	11	91.7
6	51.8	51.6	59.0	8	40.0	6	30.0	<5	66.7
22	55.6	54.2	60.2	10	38.5	0	0.0	0	0.0
167	51.7	51.4	52.9	123	65.1	83	43.9	<5	100.0
32	70.2	70.4	75.1	22	64.7	16	47.1	<5	100.0
48	57.0	56.9	56.2	60	81.1	69	93.2	6	100.0
200	57.7	57.9	58.5	164	56.0	49	16.7	21	100.0
43	62.3	62.6	65.9	44	66.7	16	24.2	<5	100.0
208	53.6	53.8	51.1	173	58.4	94	31.8	15	100.0
108	56.1	55.9	57.1	88	64.2	83	60.6	9	100.0
38	61.1	60.9	62.6	22	57.9	9	23.7	<5	100.0
34	60.9	60.1	63.5	24	63.2	8	21.1	0	0.0
100	55.8	56.0	57.3	67	51.5	30	23.1	7	87.5
165	52.8	53.2	55.4	149	49.7	131	43.7	16	100.0
24	46.3	45.7	42.3	8	19.5	13	31.7	<5	100.0
15	59.0	58.4	70.3	<5	12.0	8	32.0	<5	100.0
30	48.3	48.3	43.8	17	56.7	10	33.3	<5	100.0

Appendix 2: Centre-level data tables

Adult centres/clinics providing data in 2020 – ordered by clinic ID

Location	Name	Clinic ID	Total Active	Number with annual review *
London - South East	King's College Hospital	5	246	235
Newcastle	Royal Victoria Infirmary	9	322	308
London - South West	Royal Brompton Hospital	12	567	550
Belfast	Belfast City Hospital	14	290	48
Frimley	Frimley Park Hospital	19	143	140
Birmingham	Birmingham Heartlands Hospital	27	322	308
Exeter	Royal Devon & Exeter Hospital	34	123	122
Leeds	St James's University Hospital	42	401	390
Edinburgh	Western General Hospital	44	239	215
Cambridge	Royal Papworth Hospital	51	335	309
Plymouth	Derriford Hospital	64	66	64
Sheffield	Northern General Hospital	65	212	202
Liverpool	Liverpool Heart and Chest Hospital	66	355	335
Llandough	Llandough Hospital	68	276	240
Aberdeen	Aberdeen Royal Infirmary	70	74	72
North West Midlands	University Hospital of North Midlands	74	143	141
Glasgow	Queen Elizabeth University Hospital	79	226	218
London - East	St Bartholomew's Hospital	92	196	185
Nottingham	Nottingham University Hospitals	101	222	216
Manchester	Wythenshawe Hospital	102	466	441
London - South East	University Hospital Lewisham	105	56	53
Bristol	Bristol Royal Infirmary	106	242	230
Southampton	Southampton General Hospital	110	292	281
Norwich	Norfolk & Norwich University Hospital	114	79	73
Oxford	John Radcliffe Hospital	128	135	101
Cornwall	Royal Cornwall Hospital	129	36	35
Leicester	Glenfield Hospital	142	108	102
York & Hull	York Hospital	171	89	87

Age (years)		FEV$_1$% predicted at annual review				Best FEV$_1$% predicted			
Mean	Median	Number	Mean - unadjusted	Mean - adjusted	Median	Number**	Mean - unadjusted	Mean - adjusted	Median
31.4	29.3	204	69.5	69.3	70.3	208	71.4	70.9	71.9
31.7	29.0	272	63.7	63.2	65.4	282	67.5	66.9	70.1
35.4	33.6	472	67.0	68.4	68.0	536	68.9	70.2	70.6
33.6	30.7	43	57.5	58.1	56.8	48	60.6	61.0	56.4
32.6	32.0	122	68.9	69.4	69.8	129	71.8	72.2	71.7
34.0	31.5	158	65.7	66.5	66.0	270	69.0	69.6	68.9
33.1	30.1	62	65.3	66.2	64.4	109	73.4	73.7	74.0
35.3	33.6	183	65.0	65.9	67.2	330	65.3	66.6	66.9
33.4	30.8	157	64.0	64.2	63.8	196	69.8	70.0	69.8
31.6	29.3	215	67.0	67.0	66.8	287	70.4	70.2	70.3
34.7	33.5	50	71.1	71.7	70.9	58	78.1	78.3	79.5
31.5	29.8	77	65.8	65.5	66.9	191	73.0	72.8	77.9
32.2	30.0	287	69.1	69.3	69.5	321	73.2	73.2	74.2
32.3	29.9	197	66.2	66.1	68.4	225	72.4	72.1	73.9
34.7	33.6	65	65.4	66.1	65.7	65	68.0	68.5	65.7
30.4	26.9	112	63.3	62.1	64.2	124	67.0	65.7	69.1
34.0	30.4	123	65.5	66.0	64.9	196	66.3	66.9	66.0
29.3	27.1	147	68.5	66.8	69.1	171	70.4	68.6	72.5
31.2	29.7	135	64.0	63.5	65.1	187	68.8	68.0	69.3
32.5	30.9	95	63.0	62.6	62.8	419	64.4	64.6	62.6
32.7	31.3	39	61.6	61.8	57.8	44	65.6	66.1	63.6
31.5	30.0	212	69.3	69.2	71.1	212	72.9	72.7	74.9
34.0	30.6	99	67.9	69.0	68.5	240	71.0	71.0	71.1
30.8	28.3	27	67.7	68.1	69.1	66	72.6	72.3	72.8
31.9	27.5	66	63.7	62.5	61.7	90	67.3	66.1	66.4
35.7	33.2	24	66.5	67.8	68.6	27	70.4	71.4	70.1
31.5	28.8	55	60.8	59.8	66.9	87	63.5	62.8	67.5
33.9	30.8	81	66.0	66.7	64.5	83	70.6	71.0	71.7

* Annual review data may represent a construct derived from a recorded clinic encounter
** 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**
London - South East	King's College Hospital	223	25.3	25.5	22.8	22.7
Newcastle	Royal Victoria Infirmary	299	23.5	23.7	22.8	22.5
London - South West	Royal Brompton Hospital	550	23.0	22.7	22.5	22.4
Belfast	Belfast City Hospital	48	23.8	23.8	23.0	23.5
Frimley	Frimley Park Hospital	140	22.9	22.9	22.5	22.1
Birmingham	Birmingham Heartlands Hospital	295	23.6	23.5	23.4	23.0
Exeter	Royal Devon & Exeter Hospital	119	25.0	25.0	24.6	24.8
Leeds	St James's University Hospital	377	24.1	23.8	23.3	22.8
Edinburgh	Western General Hospital	215	24.0	23.9	22.7	22.5
Cambridge	Royal Papworth Hospital	300	23.1	23.2	22.8	22.3
Plymouth	Derriford Hospital	62	24.3	24.2	24.0	23.5
Sheffield	Northern General Hospital	200	24.1	24.2	23.3	22.9
Liverpool	Liverpool Heart and Chest Hospital	335	23.7	23.8	23.0	22.8
Llandough	Llandough Hospital	240	23.4	23.5	22.8	22.4
Aberdeen	Aberdeen Royal Infirmary	72	24.8	24.6	23.3	22.6
North West Midlands	University Hospital of North Midlands	130	22.8	23.1	22.8	22.2
Glasgow	Queen Elizabeth University Hospital	189	24.3	24.2	23.7	23.2
London - East	St Bartholomew's Hospital	182	22.7	23.2	22.3	21.8
Nottingham	Nottingham University Hospitals	213	22.8	22.9	22.0	21.8
Manchester	Wythenshawe Hospital	407	23.0	23.1	22.5	22.1
London - South East	University Hospital Lewisham	53	23.0	22.9	22.6	21.1
Bristol	Bristol Royal Infirmary	230	23.5	23.6	22.8	22.7
Southampton	Southampton General Hospital	277	23.8	23.8	23.1	22.5
Norwich	Norfolk & Norwich University Hospital	72	22.2	22.4	21.8	21.1
Oxford	John Radcliffe Hospital	99	22.6	22.7	22.1	21.9
Cornwall	Royal Cornwall Hospital	35	24.1	23.8	22.5	21.3
Leicester	Glenfield Hospital	98	23.2	23.3	22.2	21.9
York & Hull	York Hospital	86	23.7	23.6	23.3	22.0

Chronic *P. aeruginosa*		Having at least 1 IV day		Receiving DNase treatment		Receiving hypertonic saline/mannitol treatment		Inhaled antibiotic use among patients with chronic *P. aeruginosa*	
Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)
52	22.1	102	43.4	195	83.0	116	49.4	52	100.0
117	38.0	130	42.2	194	63.0	64	20.8	103	88.0
244	44.4	252	45.8	500	90.9	308	56.0	229	93.9
21	43.8	22	45.8	41	85.4	24	50.0	19	90.5
55	39.3	66	47.1	113	80.7	71	50.7	55	100.0
118	38.3	156	50.6	224	72.7	134	43.5	115	97.5
38	31.1	47	38.5	86	70.5	79	64.8	34	89.5
155	39.7	215	55.1	289	74.1	89	22.8	137	88.4
52	24.2	77	35.8	136	63.3	42	19.5	43	82.7
84	27.2	170	55.0	222	71.8	174	56.3	78	92.9
14	21.9	25	39.1	42	65.6	42	65.6	14	100.0
93	46.0	113	55.9	173	85.6	48	23.8	84	90.3
122	36.4	171	51.0	254	75.8	118	35.2	111	91.0
78	32.5	111	46.3	174	72.5	159	66.3	76	97.4
15	20.8	18	25.0	42	58.3	22	30.6	13	86.7
43	30.5	80	56.7	102	72.3	66	46.8	38	88.4
9	4.1	81	37.2	97	44.5	31	14.2	9	100.0
54	29.2	81	43.8	157	84.9	125	67.6	53	98.1
73	33.8	111	51.4	171	79.2	112	51.9	67	91.8
157	35.6	203	46.0	309	70.1	190	43.1	148	94.3
19	35.8	26	49.1	36	67.9	17	32.1	18	94.7
38	16.5	128	55.7	159	69.1	158	68.7	32	84.2
67	23.8	134	47.7	182	64.8	127	45.2	59	88.1
20	27.4	36	49.3	53	72.6	35	47.9	18	90.0
44	43.6	60	59.4	84	83.2	47	46.5	36	81.8
10	28.6	14	40.0	21	60.0	12	34.3	10	100.0
32	31.4	52	51.0	70	68.6	21	20.6	24	75.0
34	39.1	44	50.6	68	78.2	18	20.7	29	85.3

Appendix 2: Centre-level data tables

Paediatric centres/clinics providing data in 2020 – ordered alphabetically by country/city

Location	Name	Clinic ID	Total Active	Number with annual review*
England				
Birmingham	Birmingham Children's Hospital	104	311	298
Brighton	Royal Alexandra Children's Hospital	172	47	38
Bristol	Bristol Royal Hospital for Children	32	179	171
Cambridge	Addenbrookes Hospital	107	153	141
Cornwall	Royal Cornwall Hospital	94	37	36
Exeter	Royal Devon & Exeter Hospital	96	75	74
Hull	Hull University Teaching Hospitals	111	43	42
Leeds	St James's University Hospital	25	234	225
Leicester	Leicester Royal Infirmary	1	75	65
Liverpool	Alder Hey Children's Hospital	97	320	303
London - Central	Great Ormond Street Hospital for Children	90	196	188
London - East	Royal London Hospital	30	108	101
London - South East	King's College Hospital	17	189	181
London - South West	Royal Brompton Hospital	15	311	294
Manchester	Royal Manchester Children's Hospital	144	342	287
Newcastle	Great North Children's Hospital	59	190	173
North West Midlands	University Hospital of North Midlands	8	97	91
Norwich	Norfolk & Norwich University Hospital	98	70	68
Nottingham	Nottingham University Hospitals	62	173	161
Oxford	John Radcliffe Hospital	22	180	171
Plymouth	Derriford Hospital	139	41	39
Sheffield	Sheffield Children's Hospital	3	148	140
Southampton	Southampton General Hospital	29	228	211
Teeside	James Cook University Hospital	71	52	49
Northern Ireland				
Belfast	Royal Belfast Hospital for Sick Children	60	205	188
Scotland				
Aberdeen	Royal Aberdeen Children's Hospital	75	29	24
Ayr	University Hospital Crosshouse	170	25	23
Dundee	Ninewells Hospital	73	20	8
Edinburgh	Royal Hospital for Sick Children	143	142	117
Glasgow	Royal Hospital for Children	56	118	104
Inverness	Raigmore Hospital	31	16	13
Lanarkshire	Wishaw General Hospital	162	46	40
Wales				
Cardiff	Children's Hospital for Wales	72	176	157

* Annual review data may represent a construct derived from a recorded clinic encounter
** Where 'Best' values were missing, or lower than FEV_1% predicted taken at annual review, the annual review value was used.

Age (years)		FEV_1% predicted at annual review				Best FEV_1% predicted			
Mean	Median	Number	Mean - unadjusted	Mean - adjusted	Median	Number**	Mean - unadjusted	Mean - adjusted	Median
9.2	8.8	129	83.5	84.6	85.6	202	91.3	92.0	94.0
9.2	9.4	20	97.5	98.0	97.2	26	97.9	98.7	98.4
8.7	8.8	79	88.4	88.5	90.6	108	92.0	92.0	94.1
8.7	9.0	74	91.8	91.2	95.4	95	95.3	94.9	95.7
9.7	9.3	13	80.9	83.0	82.3	26	85.7	86.0	89.5
9.5	9.0	30	90.9	91.3	89.1	52	91.4	91.9	90.5
8.6	8.6	28	82.3	81.7	90.3	28	86.3	85.9	91.3
9.1	9.2	93	87.1	86.7	91.6	159	90.8	90.6	93.5
8.5	8.5	30	88.4	89.1	93.2	40	94.2	94.6	94.7
9.0	8.7	108	85.2	85.9	86.6	165	89.7	90.1	89.3
8.4	8.3	73	86.4	86.7	88.4	120	95.1	94.6	96.1
10.1	10.7	56	88.3	89.2	89.2	73	96.1	97.1	97.8
8.1	7.1	53	85.9	85.6	89.4	106	92.9	92.9	94.8
9.1	9.0	149	92.8	92.8	93.1	207	95.6	95.6	97.6
9.3	9.3	76	84.4	85.1	86.8	172	92.1	92.7	95.9
8.3	8.1	96	92.9	92.1	93.7	112	97.1	96.6	97.6
9.2	9.9	29	86.9	86.4	86.8	62	91.9	92.3	94.4
9.1	10.1	28	85.4	86.8	87.4	43	92.5	94.0	89.1
10.0	10.0	87	82.3	83.1	84.4	115	88.7	89.4	89.4
9.5	9.7	117	91.5	92.1	94.8	120	93.6	94.4	96.6
7.5	8.0	21	89.7	88.7	88.0	22	95.3	94.4	97.6
9.1	9.0	65	92.8	92.9	93.7	101	98.8	98.8	99.3
9.0	9.0	98	86.9	87.3	89.5	125	93.6	94.5	95.1
10.3	10.2	23	84.8	86.8	82.1	36	89.0	90.4	87.5
9.3	9.4	104	88.8	89.3	90.0	126	94.3	94.8	94.5
7.1	7.0	13	94.2	92.9	90.0	14	102.2	100.8	101.2
9.2	8.8	9	89.0	88.1	86.9	11	99.7	98.5	95.7
9.2	10.0	<5	79.2	81.4	79.2	<5	88.0	88.3	88.2
9.8	9.9	58	91.8	92.1	93.4	79	92.8	93.2	94.2
9.2	9.8	61	92.1	92.5	94.8	68	94.6	95.1	95.8
10.0	11.0	8	85.3	84.8	85.3	9	87.2	87.0	86.6
9.2	9.1	11	89.4	87.3	91.1	30	95.0	94.7	95.4
9.2	9.8	71	85.8	86.5	83.8	88	91.3	92.2	92.2

			BMI percentile			
Location	**Name**	**Clinic ID**	**Number**	**Mean - unadjusted**	**Mean - adjusted**	**Median**
England						
Birmingham	Birmingham Children's Hospital	104	208	53.6	53.8	51.1
Brighton	Royal Alexandra Children's Hospital	172	30	48.3	48.3	43.8
Bristol	Bristol Royal Hospital for Children	32	147	55.5	55.2	56.9
Cambridge	Addenbrookes Hospital	107	108	56.1	55.9	57.1
Cornwall	Royal Cornwall Hospital	94	32	70.2	70.4	75.1
Exeter	Royal Devon & Exeter Hospital	96	48	57.0	56.9	56.2
Hull	Hull University Teaching Hospitals	111	38	61.1	60.9	62.6
Leeds	St James's University Hospital	25	173	57.3	57.0	55.5
Leicester	Leicester Royal Infirmary	1	39	60.7	61.5	64.0
Liverpool	Alder Hey Children's Hospital	97	200	57.7	57.9	58.5
London - Central	Great Ormond Street Hospital for Children	90	167	51.7	51.4	52.9
London - East	Royal London Hospital	30	71	53.9	54.4	59.6
London - South East	King's College Hospital	17	127	52.9	52.5	50.4
London - South West	Royal Brompton Hospital	15	272	58.9	58.7	60.0
Manchester	Royal Manchester Children's Hospital	144	165	52.8	53.2	55.4
Newcastle	Great North Children's Hospital	59	144	62.9	62.5	70.5
North West Midlands	University Hospital of North Midlands	8	50	50.5	50.6	47.8
Norwich	Norfolk & Norwich University Hospital	98	43	62.3	62.6	65.9
Nottingham	Nottingham University Hospitals	62	127	50.3	50.7	52.4
Oxford	John Radcliffe Hospital	22	158	55.6	55.7	56.8
Plymouth	Derriford Hospital	139	34	60.9	60.1	63.5
Sheffield	Sheffield Children's Hospital	3	124	57.1	57.0	57.6
Southampton	Southampton General Hospital	29	177	59.3	59.2	62.2
Teeside	James Cook University Hospital	71	36	69.5	70.8	71.9
Northern Ireland						
Belfast	Royal Belfast Hospital for Sick Children	60	164	59.0	59.0	62.8
Scotland						
Aberdeen	Royal Aberdeen Children's Hospital	75	22	55.6	54.2	60.2
Ayr	University Hospital Crosshouse	170	15	59.0	58.4	70.3
Dundee	Ninewells Hospital	73	6	51.8	51.6	59.0
Edinburgh	Royal Hospital for Sick Children	143	100	55.8	56.0	57.3
Glasgow	Royal Hospital for Children	56	75	57.7	58.1	56.8
Inverness	Raigmore Hospital	31	12	46.1	46.1	39.8
Lanarkshire	Wishaw General Hospital	162	24	46.3	45.7	42.3
Wales						
Cardiff	Children's Hospital for Wales	72	89	56.9	57.1	56.5

Chronic *P. aeruginosa*		Having at least 1 IV day		Receiving DNase treatment		Receiving hypertonic saline/mannitol treatment		Inhaled antibiotic use among patients with chronic *P. aeruginosa*	
Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)
11	3.7	86	28.9	178	59.7	115	38.6	10	90.9
<5	5.3	16	42.1	23	60.5	8	21.1	<5	50.0
9	5.3	49	28.7	126	73.7	105	61.4	8	88.9
8	5.7	35	24.8	82	58.2	92	65.2	8	100.0
<5	8.3	5	13.9	24	66.7	19	52.8	<5	100.0
<5	4.1	14	18.9	64	86.5	69	93.2	<5	100.0
5	11.9	14	33.3	19	45.2	12	28.6	<5	80.0
16	7.1	58	25.8	148	65.8	23	10.2	16	100.0
<5	3.1	20	30.8	42	64.6	8	12.3	<5	100.0
20	6.6	102	33.7	166	54.8	52	17.2	20	100.0
10	5.3	61	32.4	133	70.7	80	42.6	9	90.0
14	13.9	41	40.6	74	73.3	75	74.3	14	100.0
10	5.5	53	29.3	122	67.4	63	34.8	10	100.0
19	6.5	63	21.4	228	77.6	133	45.2	20	105.3
24	8.4	73	25.4	146	50.9	112	39.0	21	87.5
14	8.1	53	30.6	103	59.5	38	22.0	13	92.9
5	5.5	41	45.1	57	62.6	26	28.6	<5	80.0
0	0.0	10	14.7	42	61.8	17	25.0	0	0.0
9	5.6	47	29.2	112	69.6	59	36.6	6	66.7
11	6.4	55	32.2	122	71.3	65	38.0	9	81.8
0	0.0	8	20.5	27	69.2	11	28.2	0	0.0
6	4.3	54	38.6	89	63.6	46	32.9	6	100.0
9	4.3	50	23.7	141	66.8	50	23.7	6	66.7
<5	4.1	14	28.6	34	69.4	9	18.4	<5	100.0
9	4.8	29	15.4	154	81.9	25	13.3	9	100.0
0	0.0	5	20.8	11	45.8	0	0.0	0	0.0
<5	4.3	5	21.7	<5	8.7	9	39.1	<5	100.0
<5	25.0	<5	12.5	<5	50.0	<5	12.5	<5	50.0
10	8.5	36	30.8	73	62.4	31	26.5	8	80.0
<5	2.9	38	36.5	30	28.8	43	41.3	<5	100.0
<5	7.7	<5	7.7	<5	30.8	<5	15.4	<5	100.0
0	0.0	5	12.5	9	22.5	15	37.5	0	0.0
<5	2.5	28	17.8	119	75.8	138	87.9	<5	100.0

Appendix 2: Centre-level data tables

Adult centres/clinics providing data in 2020 – ordered alphabetically by country/city

Location	Name	Clinic ID	Total active	Number with annual review*
England				
Birmingham	Birmingham Heartlands Hospital	27	322	308
Bristol	Bristol Royal Infirmary	106	242	230
Cambridge	Royal Papworth Hospital	51	335	309
Cornwall	Royal Cornwall Hospital	129	36	35
Exeter	Royal Devon & Exeter Hospital	34	123	122
Frimley	Frimley Park Hospital	19	143	140
Leeds	St James's University Hospital	42	401	390
Leicester	Glenfield Hospital	142	108	102
Liverpool	Liverpool Heart and Chest Hospital	66	355	335
London - East	St Bartholomew's Hospital	92	196	185
London - South East	University Hospital Lewisham	105	56	53
London - South East	King's College Hospital	5	246	235
London - South West	Royal Brompton Hospital	12	567	550
Manchester	Wythenshawe Hospital	102	466	441
Newcastle	Royal Victoria Infirmary	9	322	308
North West Midlands	University Hospital of North Midlands	74	143	141
Norwich	Norfolk & Norwich University Hospital	114	79	73
Nottingham	Nottingham University Hospital	101	222	216
Oxford	John Radcliffe Hospital	128	135	101
Plymouth	Derriford Hospital	64	66	64
Sheffield	Northern General Hospital	65	212	202
Southampton	Southampton General Hospital	110	292	281
York & Hull	York Hospital	171	89	87
Northern Ireland				
Belfast	Belfast City Hospital	14	291	232
Scotland				
Aberdeen	Aberdeen Royal Infirmary	70	74	72
Edinburgh	Western General Hospital	44	239	215
Glasgow	Queen Elizabeth University Hospital	79	226	218
Wales				
Llandough	Llandough Hospital	68	276	240

Age (years)		FEV$_1$% predicted at annual review				Best FEV$_1$% predicted			
Mean	Median	Number	Mean - unadjusted	Mean - adjusted	Median	Number**	Mean - unadjusted	Mean - adjusted	Median
34.0	31.5	158	65.7	66.5	66.0	270	69.0	69.6	68.9
31.5	30.0	212	69.3	69.2	71.1	212	72.9	72.7	74.9
31.6	29.3	215	67.0	67.0	66.8	287	70.4	70.2	70.3
35.7	33.2	24	66.5	67.8	68.6	27	70.4	71.4	70.1
33.1	30.1	62	65.3	66.2	64.4	109	73.4	73.7	74.0
32.6	32.0	122	68.9	69.4	69.8	129	71.8	72.2	71.7
35.3	33.6	183	65.0	65.9	67.2	330	65.3	66.6	66.9
31.5	28.8	55	60.8	59.8	66.9	87	63.5	62.8	67.5
32.2	30.0	287	69.1	69.3	69.5	321	73.2	73.2	74.2
29.3	27.1	147	68.5	66.8	69.1	171	70.4	68.6	72.5
32.7	31.3	39	61.6	61.8	57.8	44	65.6	66.1	63.6
31.4	29.3	204	69.5	69.3	70.3	208	71.4	70.9	71.9
35.4	33.6	472	67.0	68.4	68.0	536	68.9	70.2	70.6
32.5	30.9	95	63.0	62.6	62.8	419	64.4	64.6	62.6
31.7	29.0	272	63.7	63.2	65.4	282	67.5	66.9	70.1
30.4	26.9	112	63.3	62.1	64.2	124	67.0	65.7	69.1
30.8	28.3	27	67.7	68.1	69.1	66	72.6	72.3	72.8
31.2	29.7	135	64.0	63.5	65.1	187	68.8	68.0	69.3
31.9	27.5	66	63.7	62.5	61.7	90	67.3	66.1	66.4
34.7	33.5	50	71.1	71.7	70.9	58	78.1	78.3	79.5
31.5	29.8	77	65.8	65.5	66.9	191	73.0	72.8	77.9
34.0	30.6	99	67.9	69.0	68.5	240	71.0	71.0	71.1
33.9	30.8	81	66.0	66.7	64.5	83	70.6	71.0	71.7
33.6	30.7	43	57.5	58.1	56.8	48	60.6	61.0	56.4
34.7	33.6	65	65.4	66.1	65.7	65	68.0	68.5	65.7
33.4	30.8	157	64.0	64.2	63.8	196	69.8	70.0	69.8
34.0	30.4	123	65.5	66.0	64.9	196	66.3	66.9	66.0
32.3	29.9	197	66.2	66.1	68.4	225	72.4	72.1	73.9

* Annual review data may represent a construct derived from a recorded clinic encounter
** 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	295	23.6	23.5	23.4
Bristol	Bristol Royal Infirmary	106	230	23.5	23.6	22.8
Cambridge	Royal Papworth Hospital	51	300	23.1	23.2	22.8
Cornwall	Royal Cornwall Hospital	129	35	24.1	23.8	22.5
Exeter	Royal Devon & Exeter Hospital	34	119	25.0	25.0	24.6
Frimley	Frimley Park Hospital	19	140	22.9	22.9	22.5
Leeds	St James's University Hospital	42	377	24.1	23.8	23.3
Leicester	Glenfield Hospital	142	98	23.2	23.3	22.2
Liverpool	Liverpool Heart and Chest Hospital	66	335	23.7	23.8	23.0
London - East	St Bartholomew's Hospital	92	182	22.7	23.1	22.3
London - South East	King's College Hospital	5	53	23.0	22.9	22.6
London - South East	University Hospital Lewisham	105	222	23.5	23.7	22.8
London - South West	Royal Brompton Hospital	12	550	23.0	22.8	22.5
Manchester	Wythenshawe Hospital	102	407	23.0	23.1	22.5
Newcastle	Royal Victoria Infirmary	9	299	23.5	23.7	22.8
North West Midlands	University Hospital of North Midlands	74	130	22.8	23.1	22.8
Norwich	Norfolk & Norwich University Hospital	114	72	22.2	22.4	21.8
Nottingham	Nottingham University Hospitals	101	213	22.8	22.9	22.0
Oxford	John Radcliffe Hospital	128	99	22.6	22.7	22.1
Plymouth	Derriford Hospital	64	62	24.3	24.2	24.0
Sheffield	Northern General Hospital	65	200	24.1	24.2	23.3
Southampton	Southampton General Hospital	110	277	23.8	23.8	23.1
York & Hull	York Hospital	171	86	23.7	23.6	23.3
Northern Ireland						
Belfast	Belfast City Hospital	14	48	23.8	23.8	23.0
Scotland						
Aberdeen	Aberdeen Royal Infirmary	70	72	24.8	24.6	23.3
Edinburgh	Western General Hospital	44	214	23.3	23.2	22.7
Glasgow	Queen Elizabeth University Hospital	79	189	24.3	24.2	23.7
Wales						
Llandough	Llandough Hospital	68	240	23.4	23.5	22.8

Chronic *P. aeruginosa*		Having at least 1 IV day		Receiving DNase treatment		Receiving hypertonic saline/mannitol treatment		Inhaled antibiotic use among patients with chronic *P. aeruginosa*	
Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)
118	38.3	156	50.6	224	72.7	134	43.5	115	97.5
38	16.5	128	55.7	159	69.1	158	68.7	32	84.2
84	27.2	170	55.0	222	71.8	174	56.3	78	92.9
10	28.6	14	40.0	21	60.0	12	34.3	10	100.0
38	31.1	47	38.5	86	70.5	79	64.8	34	89.5
55	39.3	66	47.1	113	80.7	71	50.7	55	100.0
155	39.7	215	55.1	289	74.1	89	22.8	137	88.4
32	31.4	52	51.0	70	68.6	21	20.6	24	75.0
122	36.4	171	51.0	254	75.8	118	35.2	111	91.0
54	29.2	81	43.8	157	84.9	125	67.6	53	98.1
19	35.8	26	49.1	36	67.9	17	32.1	18	94.7
52	22.1	102	43.4	195	83.0	116	49.4	52	100.0
244	44.4	252	45.8	500	90.9	308	56.0	229	93.9
157	35.6	203	46.0	309	70.1	190	43.1	148	94.3
117	38.0	130	42.2	194	63.0	64	20.8	103	88.0
43	30.5	80	56.7	102	72.3	66	46.8	38	88.4
20	27.4	36	49.3	53	72.6	35	47.9	18	90.0
73	33.8	111	51.4	171	79.2	112	51.9	67	91.8
44	43.6	60	59.4	84	83.2	47	46.5	36	81.8
14	21.9	25	39.1	42	65.6	42	65.6	14	100.0
93	46.0	113	55.9	173	85.6	48	23.8	84	90.3
67	23.8	134	47.7	182	64.8	127	45.2	59	88.1
34	39.1	44	50.6	68	78.2	18	20.7	29	85.3
21	43.8	22	45.8	41	85.4	24	50.0	19	90.5
15	20.8	18	25.0	42	58.3	22	30.6	13	86.7
52	24.2	77	35.8	136	63.3	42	19.5	43	82.7
9	4.1	81	37.2	97	44.5	31	14.2	9	100.0
78	32.5	111	46.3	174	72.5	159	66.3	76	97.4

Appendix 3: Full list of 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, as people with heterozygous mutations appear twice in the table.

Nucleotide	Protein	Legacy name	N	%
c.1521_1523delCTT	p.Phe508del	F508del	9645	89.0
c.350G->A	p.Arg117His	R117H	670	6.2
c.1652G->A	p.Gly551Asp	G551D	629	5.8
c.1624G->T	p.Gly542X	G542X	398	3.7
c.489+1G->T		621+1G->T	270	2.5
c.1585-1G->A		1717-1G->A	169	1.6
c.3909C->G	p.Asn1303Lys	N1303K	169	1.6
c.1766+1G->A		1898+1G->A	153	1.4
c.200C->T	p.Pro67Leu	P67L	141	1.3
c.3454G->C	p.Asp1152His	D1152H	141	1.3
c.3528delC	p.Lys1177SerfsX15	3659delC	115	1.1
c.3140-26A->G		3272-26A->G	109	1.0
c.1679G->C	p.Arg560Thr	R560T	102	0.9
c.1519_1521delATC	p.Ile507del	I507del	91	0.8
c.1477C->T	p.Gln493X	Q493X	89	0.8
c.1657C->T	p.Arg553X	R553X	88	0.8
c.3717+12191C->T		3849+10kbC->T	87	0.8
c.254G->A	p.Gly85Glu	G85E	80	0.7
c.178G->T	p.Glu60X	E60X	74	0.7
c.1022_1023insTC	p.Phe342HisfsX28	1154insTC	73	0.7
c.2657+5G->A		2789+5G->A	72	0.7
c.3846G->A	p.Trp1282X	W1282X	63	0.6
c.1646G->A	p.Ser549Asn	S549N	56	0.5
c.948delT	p.Phe316LeufsX12	1078delT	55	0.5
c.2052delA	p.Lys684AsnfsX38	2184delA	51	0.5
c.1364C->A	p.Ala455Glu	A455E	49	0.5
c.617T->G	p.Leu206Trp	L206W	46	0.4
c.1040G->C	p.Arg347Pro	R347P	39	0.4
c.2657+2_2657+3insA		2789+2insA	38	0.4
c.1558G->T	p.Val520Phe	V520F	34	0.3
c.579+3A->G		711+3A->G	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	27	0.2
c.2988+1G->A		3120+1G->A	26	0.2
c.1055G->A	p.Arg352Gln	R352Q	23	0.2
c.1210-12[5] (AJ574948.1:g.152T[5])		5T	23	0.2

Nucleotide	Protein	Legacy name	N	%
c.3718-2477C->T		3849+10kbC->T	22	0.2
c.2583delT	p.Phe861LeufsX3	2711delT	22	0.2
c.3472C->T	p.Arg1158X	R1158X	21	0.2
c.1006_1007insG	p.Ile336SerfsX28	1138insG	21	0.2
c.2490+1G->A		2622+1G->A	20	0.2
c.1705T->G	p.Tyr569Asp	Y569D	20	0.2
c.1367T->C	p.Val456Ala	V456A	20	0.2
c.2125C->T	p.Arg709X	R709X	19	0.2
c.532G->A	p.Gly178Arg	G178R	19	0.2
c.2834C->T	p.Ser945Leu	S945L	19	0.2
c.1393-1G->A		1525-1G->A	19	0.2
c.3197G->A	p.Arg1066His	R1066H	18	0.2
c.1523T->G	p.Phe508Cys	F508C	18	0.2
c.3806T->A	p.Ile1269Asn	I1269N	17	0.2
c.2052_2053insA	p.Gln685ThrfsX4	2184insA	16	0.1
c.658C->T	p.Gln220X	Q220X	14	0.1
c.2537G->A	p.Trp846X	W846X	13	0.1
c.292C->T	p.Gln98X	Q98X	13	0.1
c.3737C->T	p.Thr1246Ile	T1246I	12	0.1
c.1029delC	p.Cys343X	1161delC	12	0.1
c.579+1G->T		711+1G->T	12	0.1
c.2988G->A		3120G->A	11	0.1
c.2875delG	p.Ala959HisfsX9	3007delG	11	0.1
c.3705T->G	p.Ser1235Arg	S1235R	10	0.1
c.349C->T	p.Arg117Cys	R117C	10	0.1
c.3208C->T	p.Arg1070Trp	R1070W	10	0.1
c.1466C->A	p.Ser489X	S489X	10	0.1
c.224G->A	p.Arg75Gln	R75Q	9	0.1
c.3196C->T	p.Arg1066Cys	R1066C	9	0.1
c.1675G->A	p.Ala559Thr	A559T	8	0.1
c.3468G->A		3600G->A	8	0.1
c.494T->C	p.Leu165Ser	L165S	8	0.1
c.1679+1G->C		1811+1G->C	8	0.1
c.695T->A	p.Val232Asp	V232D	8	0.1
c.2012delT	p.Leu671X	2143delT	7	0.1
c.2051_2052delAAinsG	p.Lys684SerfsX38	2183AA->G or 2183de-lAA->G	7	0.1
c.1986_1989delAACT	p.Thr663ArgfsX8	2118del4	6	0.1

c.1329_1330insAGAT	p.Ile444ArgfsX3	1461ins4	6	0.1
c.3884_3885insT	p.Ser1297PhefsX5	4016insT	6	0.1
c.2128A->T	p.Lys710X	K710X	6	0.1
c.1766+1G->T		1898+1G->T	6	0.1
c.1116+1G->A		1248+1G->A	6	0.1
c.3761T->G	p.Leu1254X	L1254X	6	0.1
c.4196_4197delTC	p.Cys1400X	4326delTC	6	0.1
c.2353C->T	p.Arg785X	R785X	6	0.1
c.1721C->A	p.Pro574His	P574H	6	0.1
c.2900T->C	p.Leu967Ser	L967S	5	0.0
c.2551C->T	p.Arg851X	R851X	5	0.0
c.[1210–12[5];1210-34TG[13]]		5T;TG13	5	0.0
c.2290C->T	p.Arg764X	R764X	5	0.0
c.1687T->A	p.Tyr563Asn	Y563N	5	0.0
c.223C->T	p.Arg75X	R75X	5	0.0
c.3848G->T	p.Arg1283Met	R1283M	5	0.0
c.[1210–12[5];1210-34TG[12]]		5T;TG12	5	0.0
c.349C->G	p.Arg117Gly	R117G	5	0.0
c.3718-1G->A		3850-1G->A	5	0.0
c.2215delG	p.Val739TyrfsX16	2347delG	<5	-
c.3353C->T	p.Ser1118Phe	S1118F	<5	-
c.2249C->T	p.Pro750Leu	P750L	<5	-
c.1393-2A->G		1525-2A->G	<5	-
c.2464G->T	p.Glu822X	E822X	<5	-
c.1679G->A	p.Arg560Lys	R560K	<5	-
c.1680A->C	p.Arg560Ser	R560S	<5	-
c.(743+1_744-1)_(1584+1_1585-1)dup		CFTRdup6b-10	<5	-
c.3095A->G	p.Tyr1032Cys	Y1032C	<5	-
c.165-3C>T		297-3C->T	<5	-
c.595C->T	p.His199Tyr	H199Y	<5	-
c.3292T->C	p.Trp1098Arg	W1098R	<5	-
c.443T->C	p.Ile148Thr	I148T	<5	-
c.1538A->G	p.Asp513Gly	D513G	<5	-
c.850dupA	p.Met284AsnfsX3	977insA	<5	-
c.2909G->A	p.Gly970Asp	G970D	<5	-
c.262_263delTT	p.Leu88IlefsX22	394delTT	<5	-
c.3988C->T	p.Gln1330X	Q1330X	<5	-
c.1585-8G->A		1717-8G->A	<5	-
c.2600_2601insA	p.Val868SerfsX28	2732insA	<5	-
c.3080T->C	p.Ile1027Thr	I1027T	<5	-
c.1766+5G->T		1898+5G->T	<5	-
c.1340delA	p.Lys447ArgfsX2	1471delA	<5	-
c.509G->A	p.Arg170His	R170H	<5	-

Nucleotide	Protein	Legacy name	N	%
c.274G->A	p.Glu92Lys	E92K	<5	-
c.1724T->A	p.Phe575Tyr	F575Y	<5	-
c.1736A->G	p.Asp579Gly	D579G	<5	-
c.2260G->A	p.Val754Met	V754M	<5	-
c.1505T->C	p.Ile502Thr	I502T	<5	-
c.2491G->T	p.Glu831X	E831X	<5	-
c.1572C->A	p.Cys524X	C524X	<5	-
c.2896delA	p.Thr966ArgfsX2	3028delA	<5	-
c.91C->T	p.Arg31Cys	R31C	<5	-
c.328G->C	p.Asp110His	D110H	<5	-
c.2991G->C	p.Leu997Phe	L997F	<5	-
c.3659delC	p.Thr1220LysfsX8	3791delC	<5	-
c.4147_4148insA	p.Ile1383AsnfsX3	4279insA	<5	-
c.[1521_1523delCTT;3080T->C]	p.[Phe508del;Ile-1027Thr]	F508del;I1027T	<5	-
c.350G->T	p.Arg117Leu	R117L	<5	-
c.3700A->G	p.Ile1234Val	I1234V	<5	-
c.577G->T	p.Glu193X	E193X	<5	-
c.4046G->A	p.Gly1349Asp	G1349D	<5	-
c.4004T->C	p.Leu1335Pro	L1335P	<5	-
c.4111G->T	p.Glu1371X	E1371X	<5	-
c.3908delA	p.Asn1303ThrfsX25	4040delA	<5	-
c.1001G>A	p.Arg334Gln	R334Q	<5	-
c.3475T->C	p.Ser1159Pro	S1159P	<5	-
c.442delA	p.Ile148LeufsX5	574delA	<5	-
c.1766+1G->C		1898+1G->C	<5	-
c.3017C->A	p.Ala1006Glu	A1006E	<5	-
c.296C->T	p.Pro99Leu	P99L	<5	-
c.220C->T	p.Arg74Trp	R74W	<5	-
c.[1210–12[5];1210-34TG[11]]		5T;TG11	<5	-
c.1651G->A	p.Gly551Ser	G551S	<5	-
c.1477_1478delCA	p.Gln493ValfsX10	1609delCA	<5	-
c.2374C->T	p.Arg792X	R792X	<5	-
c.3872A->G	p.Gln1291Arg	Q1291R	<5	-
c.2195T->G	p.Leu732X	L732X	<5	-
c.1679+1.6kbA->G		1811+1.6kbA->G	<5	-
c.164+2T>C		296+2T->C	<5	-
c.3266G->A	p.Trp1089X	W1089X	<5	-
c.79G->T	p.Gly27X	G27X	<5	-
c.3752G->A	p.Ser1251Asn	S1251N	<5	-
c.1007T->A	p.Ile336Lys	I336K	<5	-
c.1727G->C	p.Gly576Ala	G576A	<5	-
c.3763T->C	p.Ser1255Pro	S1255P	<5	-

Nucleotide	Protein	Legacy name	N	%
c.2780T->C	p.Leu927Pro	L927P	<5	-
c.3882_3885delTATT	p.Ile1295PhefsX32	4010del4	<5	-
c.2668C->T	p.Gln890X	Q890X	<5	-
c.3310G->T	p.Glu1104X	E1104X	<5	-
c.3205G->A	p.Gly1069Arg	G1069R	<5	-
c.1046C->T	p.Ala349Val	A349V	<5	-
c.1327G->T	p.Asp443Tyr	D443Y	<5	-
c.4077_4080delTGTTinsAA	p.Val1360delfsX?	4209TGTT->AA	<5	-
c.3458T->A	p.Val1153Glu	V1153E	<5	-
c.4231C->T	p.Gln1411X	Q1411X	<5	-
c.2930C->T	p.Ser977Phe	S977F	<5	-
c.3194T->C	p.Leu1065Pro	L1065P	<5	-
c.2989-1G->A		3121-1G->A	<5	-
c.1408A->G	p.Met470Val	M470V	<5	-
c.3158C->T	p.Thr1053Ile	T1053I	<5	-
c.613C->T	p.Pro205Ser	P205S	<5	-
c.1573C->T	p.Gln525X	Q525X	<5	-
c.1037T->C	p.Leu346Pro	L346P	<5	-
c.1837G->A	p.Ala613Thr	A613T	<5	-
c.3297C->A	p.Phe1099Leu	F1099L	<5	-
c.3302T->G	p.Met1101Arg	M1101R	<5	-
c.2421A->G	p.Ile807Met	I807M	<5	-
c.3717G->A		3849G->A	<5	-
c.53+1G->T		185+1G->T	<5	-
c.1A->G	p.Met1Val	M1V	<5	-
c.2645G->A	p.Trp882X	W882X	<5	-
c.233dupT	p.Trp79LeufsX32	365-366insT	<5	-
c.1021T->C	p.Ser341Pro	S341P	<5	-
c.601G->A	p.Val201Met	V201M	<5	-
c.3476C->T	p.Ser1159Phe	S1159F	<5	-
c.1209+1G->A		1341+1G->A	<5	-
c.3773_3774insT	p.Leu1258PhefsX7	3905insT	<5	-
c.1418delG	p.Gly473GlufsX54	1548delG	<5	-
c.164+1G>A		296+1G->A	<5	-
c.263T>A or c.263T>G	p.Leu88X	L88X	<5	-
c.717delG	p.Leu240X	849delG	<5	-
c.1703delT	p.Leu568CysfsX4	1833delT	<5	-
c.3745G->A	p.Gly1249Arg	G1249R	<5	-
c.413_415dupTAC	p.Leu138dup	L138ins	<5	-
c.274-2A->G		406-2A->G	<5	-
c.470_483del14	p.Phe157X	602del14	<5	-
c.1682C->A	p.Ala561Glu	A561E	<5	-

Nucleotide	Protein	Legacy name	N	%
c.137C->A	p.Ala46Asp	A46D	<5	-
c.2735C->A	p.Ser912X	S912X	<5	-
c.859_863delAACTT	p.Asn287LysfsX19	991del5	<5	-
c.2620-26A->G		2752-26A->G	<5	-
c.1654C->T	p.Gln552X	Q552X	<5	-
c.(53+1_54-1)_(164+1_165-1)del		CFTRdele2	<5	-
c.11C>A	p.Ser4X	S4X	<5	-
c.273+1G->A		405+1G->A	<5	-
c.1545_1546delTA	p.Tyr515X	1677delTA	<5	-
c.3718-3T->G		3850-3T->G	<5	-
c.50delT	p.Phe17SerfsX8	182delT	<5	-
c.987delA	p.Gly330GlufsX39	1119delA	<5	-
c.1301_1307delCACTTCT	p.Ser434LeufsX6	1429del7	<5	-
c.1081delT	p.Trp361GlyfsX8	1213delT	<5	-
c.3209G->A	p.Arg1070Gln	R1070Q	<5	-
c.1240C->T	p.Gln414X	Q414X	<5	-
c.2739T->A	p.Tyr913X	Y913X	<5	-
c.2002C->T	p.Arg668Cys	R668C	<5	-
c.1117-1G>A		1249-1G->A	<5	-
c.3181G->C	p.Gly1061Arg	G1061R	<5	-
c.3717+5G->A		3849+5G->A	<5	-
c.3011_3019delCTATAGCAG or c.3009_3017delAGCTATAGC	p.Ala1004_Ala1006del	3143del9	<5	-
c.(53+1_54-1)_(489+1_490-1)del		CFTRdele2-4	<5	-
c.3485G->T	p.Arg1162Leu	R1162L	<5	-
c.3873+2T->C		4005+2T->C	<5	-
c.1687T->G	p.Tyr563Asp	Y563D	<5	-
c.3230T->C	p.Leu1077Pro	L1077P	<5	-
c.2859_2890delACATTCT-GTTCTTCAAGCACCTATGT-CAACCC	p.Leu953PhefsX11	2991del32	<5	-
c.4144C->T	p.Gln1382X	Q1382X	<5	-
c.933_935delCTT	p.Phe312del	F311del	<5	-
`Other' selected			**781**	**7.2**

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