Cystic Fibrosis strength in numbers

UK Cystic Fibrosis Registry
Annual Data Report 2019

Published August 2020

Cystic Fibrosis strength in numbers

UK Cystic Fibrosis Registry 2019 Annual Data Report

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

Report prepared by

Susan Charman	Senior Statistician	Cystic Fibrosis Trust
Andrew Lee	Medical Statistician	Cystic Fibrosis Trust
Rebecca Cosgriff	Director of Data & Quality Improvement	Cystic Fibrosis Trust
Elliot McClenaghan	Medical Statistician	Cystic Fibrosis Trust
Siobhán Carr	Consultant Respiratory Paediatrician	Royal Brompton Hospital

With assistance from

Annabel Dakin	Lead Graphic Designer	Cystic Fibrosis Trust
Elaine Gunn	Registry Data Manager	Cystic Fibrosis Trust
Kieran Earlam	Registry Coordinator	Cystic Fibrosis Trust

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.

Contact information

For more information about this report, or the UK Cystic Fibrosis Registry, please contact us:

registry@cysticfibrosis.org.uk

@CFTrust

Cystic Fibrosis strength in numbers

Contents

Foreword

The preparation of this year's UK Cystic Fibrosis (CF) Registry Annual Data Report has taken place during a time of intense challenge and hope for people with cystic fibrosis.

On the one hand, we remain in the midst of a global pandemic that has seen our community advised they are clinically extremely vulnerable and should shield for many months. The health, social and economic impact of this is huge and it has been deeply moving to see people with CF, their families, and their clinical teams come together to make sure we get through this difficult time together.

The Registry has played a key role in supporting these efforts – from reporting weekly COVID-19 statistics on the **Cystic Fibrosis Trust website** to keep the community up to date and provide advice and reassurance where we can. The UK CF Registry has also been pivotal in a global initiative to monitor COVID-19 in the CF population, in order to get targeted and tailored advice out in the public domain as quickly as possible to support evidence-based decision making. The worldwide CF community has once again demonstrated its dedication to a life unlimited, with representatives form 20 countries coming together to provide timely information at a time of real adversity.

Alongside this, we have welcomed the fantastic news that the Vertex Triple Therapy, Kaftrio, will be made available to people across the UK as soon as European Medicines Agency approval is granted. News that thousands of people will access a disease-modifying treatment for the first time highlights the importance of the Registry, which has enabled policy makers to understand the economic impact of paying for new therapies. It also allows for managed access of these treatments to be carefully monitored and independently reported, and for us to gather long-term data to help us understand how the needs of people with cystic fibrosis may change over the coming years. The UK CF Registry and the data it contains put the Cystic Fibrosis Trust and healthcare teams in the best possible position to provide optimal care for everyone.

It may feel like a lot has changed since 2019; the year that this report covers, however, the insights included in this report are an important view into the evolving world of CF at what we may come to see as a landmark year for many in the community.

Our community is larger than ever with - for the first time - more than 10,000 people with cystic fibrosis receiving an annual review in one year, amongst an overall population of 10,655. As well as growing in size, our community is, little by little, getting older. For the first time the median age of people with cystic fibrosis has reached 21 years. This is still far too young – the average age of the general population in the UK is approximately 40 years – and shows that we still have much work to do.

We are delighted to hear what you think of our reports, so please get in touch with us on social media or by emailing registry@cysticfibrosis.org.uk to let us know your feedback and questions.

Thank you again for your support of the Registry – we will work hard over the next year to gather the data needed to monitor the impact of CF therapies, and understand the needs of people not currently eligible for a CFTR modulator – whether because of their age, transplant status, genotype or drug intolerance. We will continue to monitor the short and long-term effects of the current pandemic and work with colleagues nationally and internationally to provide information, advice and support to the community.

David Ramsden
Chief Executive

Executive summary

We are pleased to share with you the annual report for UK Cystic Fibrosis Registry data entered for the calendar year 2019. The data used for the report are taken from the 10,070 Annual Reviews entered by the clinical teams across the UK. This high number of annual reviews reflects the increasing population of people with cystic fibrosis (CF) in the UK; 56.7% of people with CF are now adults and this change in age distribution is shown nicely in 1.3 (p12).

A few other things to highlight in this year's report:

Reporting the best FEV_1 (median 81.2%) in the year prior to annual review (p23) puts our reporting in line with our European Registry[1] colleagues and continues to be higher than that done at Annual Review. 1.15 (p24) shows improvements in the FEV_1 at Annual Review since 2009.

Teenage years continue to be a concern, with the weight centile for adolescents appearing to drop off rapidly (p15). This seems to be happening at the same ages as lung function steps down more rapidly, too (p23). Remembering that these figures represent age groups, not an individual's long-term data, it will be interesting to see if this weight drop-off is restricted to adolescents born before universal newborn screening was in place across the UK.

A total of 887 people who attended annual reviews in 2019 were diagnosed with CF after the age of 16 (p19-20).

New cases of Mycobacterium abscesses complex appear to remain stable over the past three years, with 126 new cases reported this year (p29). Caution must always be taken with small numbers and trends will take some time to emerge in this area.

Similarly, the rates for most complications, such as ABPA, haemoptysis and cancer, remain stable as shown in section 1.31, although a slight increase in those reporting depression at 4.4% should be noted (p31).

Assessment for transplant and numbers receiving transplants also remains static (p42). The trend for improving predicted survival continues (p44).

The classic lung infection figure in section 1.17 (p26) has changed slightly this year. We are showing the proportion with infections using the whole population as the denominator rather than only those with a sample taken. This means that the peak for chronic Pseudomonas may look a little lower at 50.4%, but the overall trend downwards over the years remains (p29).

For the first time, this year we have reported the proportion of people treated with insulin for their CF-related diabetes (p32); in the adult population this is nearly 90% and for children it is 98.5%.

We have introduced a new table (1.28) to show the proportions of people with CF on bronchodilators and inhaled corticosteroids (p37), with 50% having one form of inhaled corticosteroid.

The figures in section 1.44 (p49) nicely show the different distribution of genotypes across the devolved nations, with 35.6% being homozygous for F508del in Northern Ireland compared with 50.1% in England.

We have introduced a new Ethnicity section 1.4 (p13), showing the slight increase in those of ethnic minority backgrounds in the CF population since 2009.

The main aim of the Registry is to improve the health of people with cystic fibrosis. Our hope is that this report serves to stimulate continued improvements. Putting together the report is a team effort and has been greatly enhanced by the input of members of the Registry Steering Committee over the past few years.

In these strange times of the COVID-19 pandemic and the exciting introduction of CFTR modulators for the majority of people with CF, it will be interesting to see to how different the report may look in the coming years. Our continued gratitude goes to all people with CF that engage so enthusiastically with the Registry, thus allowing us to present this information.

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

[1] https://www.ecfs.eu/ecfspr

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 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 the 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 - and to beat - cystic fibrosis.

Helping commissioners provide funding to NHS CF centres that is proportionate to their patients' disease severity.

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, for 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

	2014	2015	2016	2017	2018	2019
CF patients registered[1]	10583	10810	10461	10469	10509	10655
Excluding diagnoses that year	10356	10586	10214	10255	10287	10462
CF patients with an annual review; n (%) [2]	9432 (91%)	9587 (91%)	9695 (95%)	9887 (96%)	9847 (96%)	10070 (96%)
Age in years; median[3]	19	19	20	20	20	21
All newly diagnosed patients (newborn screening [NBS] and other)[4]	291	224	247	214	222	193
All newly diagnosed patients (amended)[5]	(291)	(300)	(303)	(270)	(278)	(TBD)
Number of patients born identified by NBS[4]	164	168	216	192	167	137
Age at diagnosis in months; median[3]	2	2	2	2	2	2
Adults aged 16 years and over; %[3]	59.3	59.9	60.4	60.6	60.4	60.6
Males; %[3]	53.0	53.0	53.2	53.3	53.0	53.2
Genotyped; %[3]	97.7	98.1	98.4	99.3*	99.1	99.2
Total deaths reported during annual review year; n (%)[6]	132 (1.2%)	125 (1.2%)	148 (1.5%)	132 (1.3%)	137 (1.3%)	114 (1.1%)
Total deaths reported amended; n (%)[5]	139 (1.3%)	135 (1.2%)	159 (1.5%)	143 (1.4%)	143 (1.4%)	(TBD)
Age at death in years; median (95% CI)[6]	28 (25.5, 32)	28 (27, 33)	31 (29, 33)	31(29, 35)	32 (29, 35)	31 (29, 34)

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).

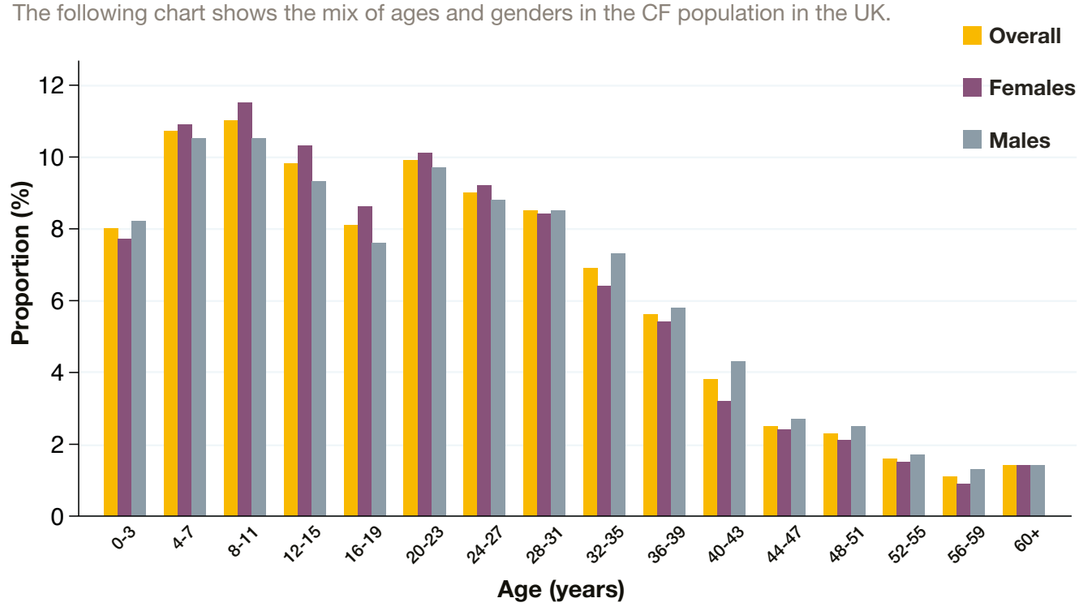
[4] Calculated from all patients registered on the database. Some diagnosis data are added after the data entry closure each year, so figures from previous years have been updated for this report.

[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.

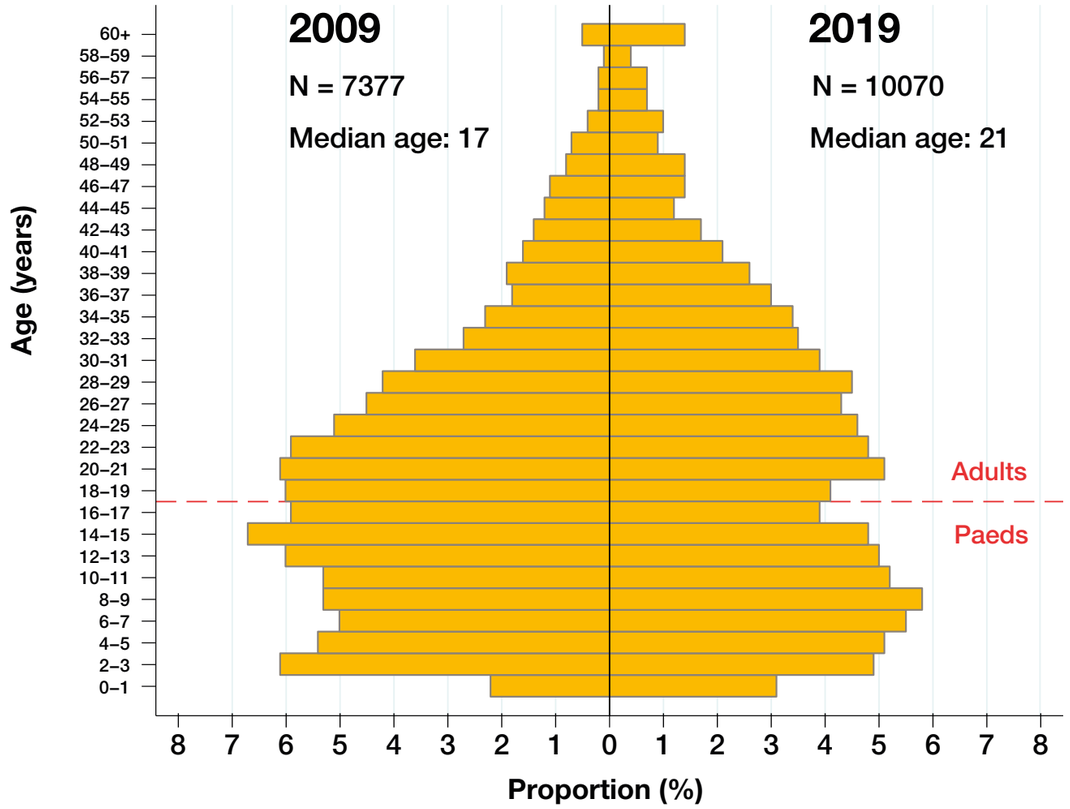
1.2 Age distribution by sex
N=10070

The following chart shows the mix of ages and genders in the CF population in the UK.



Age	All; N (%)	Females; n (%)	Males; n (%)
0-3	803 (8.0)	364 (7.7)	439 (8.2)
4-7	1075 (10.7)	514 (10.9)	561 (10.5)
8-11	1105 (11.0)	540 (11.5)	565 (10.5)
12-15	983 (9.8)	484 (10.3)	499 (9.3)
16-19	812 (8.1)	407 (8.6)	405 (7.6)
20-23	996 (9.9)	477 (10.1)	519 (9.7)
24-27	902 (9.0)	433 (9.2)	469 (8.8)
28-31	852 (8.5)	394 (8.4)	458 (8.5)
32-35	695 (6.9)	303 (6.4)	392 (7.3)
36-39	564 (5.6)	253 (5.4)	311 (5.8)
40-43	379 (3.8)	151 (3.2)	228 (4.3)
44-47	256 (2.5)	111 (2.4)	145 (2.7)
48-51	229 (2.3)	97 (2.1)	132 (2.5)
52-55	164 (1.6)	73 (1.5)	91 (1.7)
56-59	110 (1.1)	43 (0.9)	67 (1.3)
60+	145 (1.4)	68 (1.4)	77 (1.4)
<16	3966 (39.4)	1902 (40.4)	2064 (38.5)
≥16	6104 (60.6)	2810 (59.6)	3294 (61.5)
<18	4362 (43.3)	2092 (44.4)	2270 (42.4)
≥18	5708 (56.7)	2620 (55.6)	3088 (57.6)
Overall	**10070**	**4712**	**5358**

1.3 Age distribution of the UK CF population in 2009 vs 2019



(Note the different demographic distribution across the years, namely higher proportion in older age groups in 2019.)

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

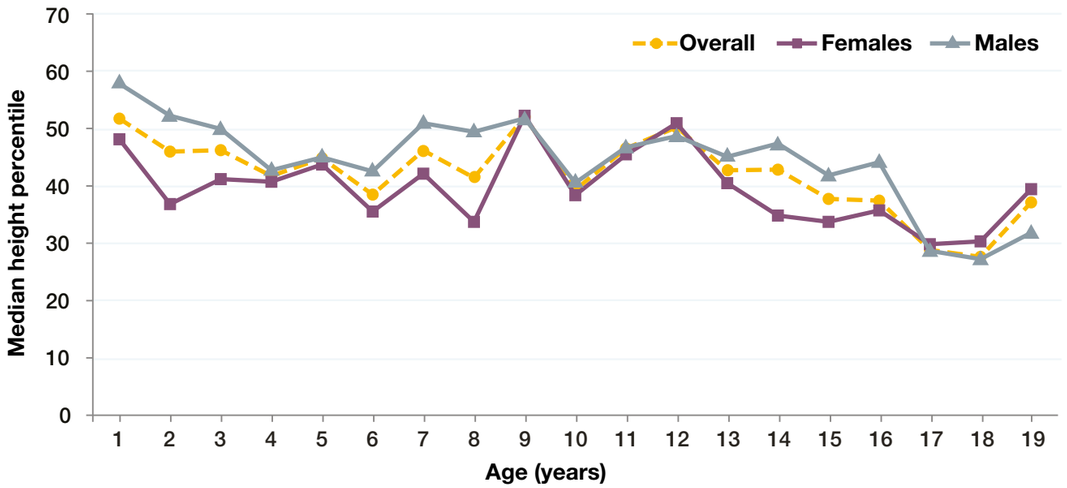
1.4 Ethnicity

	2009	2014	2019
Total	7377	9432	10070
White			
White (all); n (%)	7059 (95.7)	8939 (94.8)	9396 (93.3)
Asian			
Bangladeshi; n (%)	23 (0.3)	32 (0.3)	38 (0.4)
Indian; n (%)	24 (0.3)	33 (0.3)	45 (0.4)
Pakistani; n (%)	107 (1.5)	150 (1.6)	173 (1.7)
Other (Asian); n (%)	15 (0.2)	25 (0.3)	30 (0.3)
Black			
Black African; n (%)	10 (0.1)	12 (0.1)	12 (0.1)
Black Caribbean; n (%)	14 (0.2)	14 (0.1)	12 (0.1)
Other (Black); n (%)	<5	<5	<5
Mixed*			
Mixed (all); n (%)	31 (0.4)	74 (0.8)	58 (0.6)
Mixed (White-Asian); n (%)	-	-	15 (0.1)
Mixed (White-Black African); n (%)	-	-	9 (0.1)
Mixed (White-Black Caribbean); n (%)	-	-	19 (0.2)
Other (mixed); n (%)	-	-	15 (0.1)
Other/Unknown			
Other; n (%)	57 (0.8)	103 (1.1)	94 (0.9)
Unknown; n (%)	33 (0.4)	48 (0.5)	208 (2.1)

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

1.5 Height percentiles of children and young people (<20 years)[1] N=4778

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.

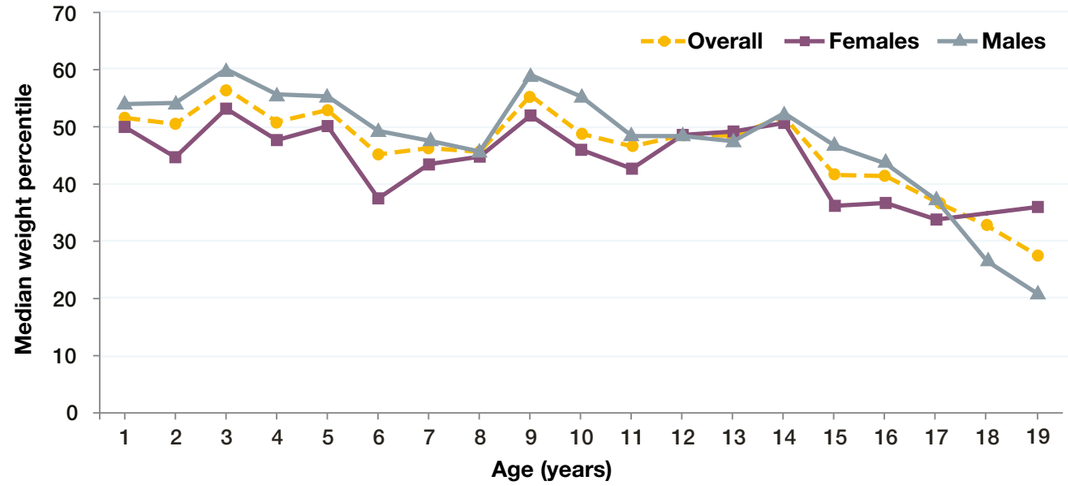


	Overall			Female			Male		
Age	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**
1	220	51.7	25.8-75.9	94	48.1	24.4-75.1	126	57.8	31.4-78.3
2	224	46.0	14.6-69.8	112	36.8	11.6-64.4	112	52.2	22.5-73.9
3	263	46.2	20.8-71.3	120	41.2	19.0-70.8	143	49.9	21.6-74.0
4	259	41.7	19.8-68.5	129	40.7	18.7-68.5	130	42.7	20.7-68.0
5	256	44.8	20.8-69.1	114	43.7	16.2-68.4	142	44.9	24.4-69.5
6	262	38.5	15.0-67.2	129	35.5	12.0-67.2	133	42.5	22.2-65.2
7	296	46.2	22.3-69.3	142	42.1	23.8-69.1	154	50.9	19.9-71.8
8	301	41.4	19.3-68.7	145	33.7	15.2-62.4	156	49.4	23.3-73.1
9	277	52.0	25.7-74.5	131	52.2	25.0-76.3	146	51.8	28.3-71.4
10	271	39.3	21.5-68.7	134	38.3	17.9-71.3	137	40.5	22.0-67.2
11	249	46.5	21.2-69.9	129	45.4	19.8-70.4	120	46.7	21.9-69.1
12	271	50.2	21.4-78.0	127	50.9	28.0-77.0	144	48.7	17.1-81.4
13	230	42.7	21.6-69.5	118	40.4	21.9-63.1	112	45.0	21.0-73.3
14	236	42.8	21.6-66.8	102	34.8	20.6-58.7	134	47.3	23.3-71.2
15	245	37.7	15.8-63.0	136	33.7	14.4-60.8	109	41.8	17.0-65.7
16	178	37.4	16.4-64.0	89	35.7	14.2-59.5	89	44.1	19.3-68.9
17	215	28.8	10.5-64.0	101	29.8	12.1-65.9	114	28.6	9.9-56.0
18	217	27.6	13.7-59.3	117	30.3	13.8-59.3	100	27.2	8.8-57.3
19	199	37.1	11.7-65.1	100	39.4	13.6-65.3	99	31.8	11.7-65.1
Overall	**4669***	**42.5**	**18.3-68.8**	**2269**	**40.0**	**17.1-67.2**	**2400**	**44.8**	**20.0-70.0**

* 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=4778

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.

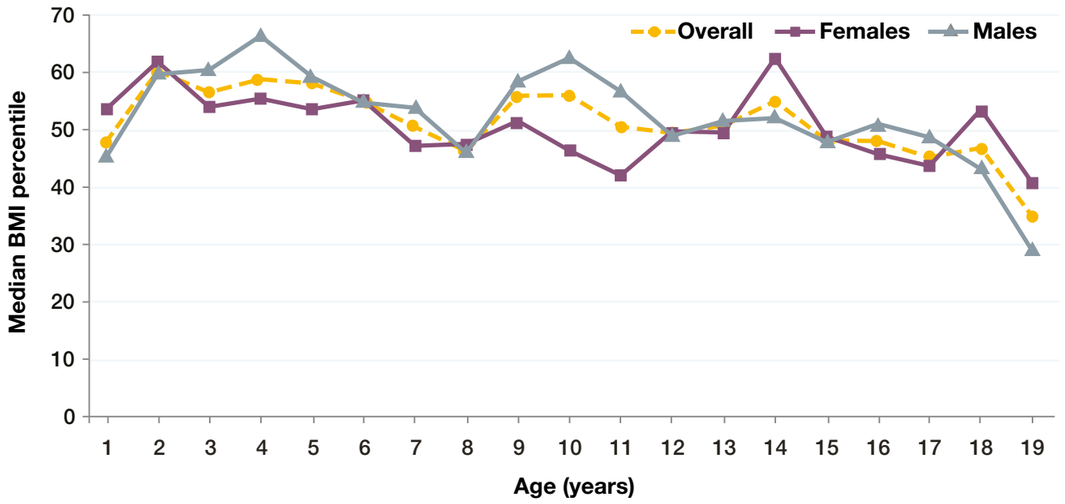


	Overall			Female			Male		
Age	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**
1	229	51.6	22.8-78.2	99	50.0	20.0-76.3	130	54.0	23.0-80.0
2	226	50.5	26.3-79.2	113	44.7	27.1-73.4	113	54.2	26.3-79.8
3	265	56.7	29.8-78.3	121	53.2	24.5-74.1	144	60.1	31.2-79.9
4	260	50.9	27.9-75.8	129	47.7	26.4-72.0	131	55.7	29.5-76.6
5	256	53.0	30.3-75.4	114	50.2	28.7-74.6	142	55.3	31.3-76.7
6	262	45.2	21.4-72.2	129	37.5	21.0-74.5	133	49.3	23.1-69.6
7	296	46.3	25.1-73.6	142	43.5	24.8-72.1	154	47.6	25.8-76.3
8	302	45.7	23.1-68.3	145	44.8	18.9-66.2	157	45.7	25.2-69.6
9	278	55.5	30.5-78.1	131	52.0	30.5-75.2	147	59.1	27.9-80.0
10	272	48.8	23.2-76.6	134	46.0	24.6-71.6	138	55.3	22.9-78.0
11	250	46.6	24.2-70.0	129	42.7	21.6-68.4	121	48.4	29.0-71.0
12	272	48.6	23.7-81.9	128	48.6	24.1-80.2	144	48.4	23.7-82.5
13	230	48.5	21.5-72.9	118	49.2	21.5-69.4	112	47.5	21.7-73.9
14	236	51.8	26.6-73.0	102	50.7	27.5-68.1	134	52.4	26.4-74.1
15	245	41.6	17.2-66.4	136	36.2	14.7-64.8	109	46.7	18.3-66.4
16	179	41.4	18.1-62.1	89	36.7	20.6-55.2	90	43.6	17.1-72.4
17	215	36.9	8.3-61.3	100	33.8	11.8-57.1	115	37.2	7.5-67.4
18	217	32.7	8.6-69.6	117	34.9	14.0-68.2	100	26.5	4.9-70.7
19	199	27.5	6.6-65.5	100	36.0	10.8-71.8	99	20.8	5.1-57.0
Overall	**4689**	**46.8**	**21.8-73.6**	**2276**	**45.1**	**21.6-70.4**	**2413**	**48.7**	**22.1-75.7**

* 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=4778

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.

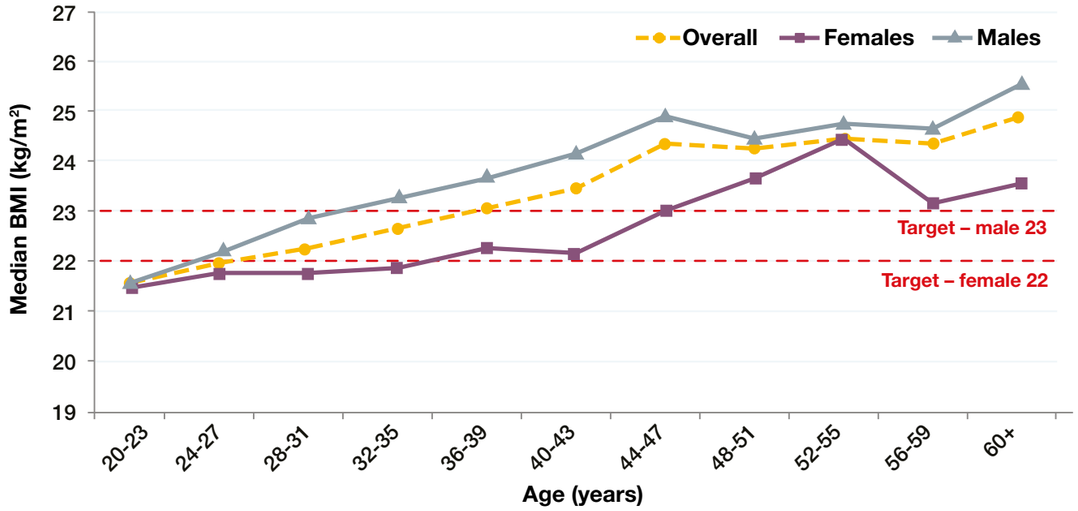


	Overall			Female			Male		
Age	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**
1	220	47.8	23.4-75.0	94	53.5	19.8-76.6	126	45.2	27.6-73.3
2	224	60.2	32.8-79.1	112	61.8	37.2-78.3	112	59.7	32.0-81.6
3	263	56.5	38.0-78.4	120	54.0	38.2-73.4	143	60.4	37.4-82.3
4	259	58.8	37.2-80.9	129	55.4	31.3-78.0	130	66.4	38.3-82.3
5	256	58.0	34.4-79.3	114	53.6	34.5-76.3	142	59.2	34.3-82.3
6	262	54.9	31.5-76.3	129	55.1	32.2-76.3	133	54.7	31.4-75.2
7	296	50.5	27.3-75.4	142	47.2	26.2-71.8	154	53.8	30.5-77.5
8	301	46.2	27.5-72.6	145	47.5	30.1-74.5	156	45.9	25.7-72.3
9	277	55.9	32.8-81.0	131	51.5	32.0-76.4	146	58.4	34.3-83.1
10	271	56.0	28.0-78.1	134	46.3	24.0-71.7	137	62.5	30.3-81.6
11	249	50.5	22.0-70.8	129	42.0	19.7-63.6	120	56.7	30.1-76.4
12	271	49.4	25.0-77.7	127	49.7	20.6-75.7	144	48.9	26.9-81.2
13	230	50.7	25.8-73.1	118	49.5	29.5-73.1	112	51.5	21.1-72.3
14	236	54.9	30.1-77.2	102	62.5	37.5-83.1	134	52.0	25.2-72.7
15	245	48.1	25.7-73.2	136	48.8	21.9-75.7	109	47.8	32.4-70.8
16	178	48.0	29.4-75.4	89	45.7	31.5-67.5	89	51.0	27.7-80.4
17	214	45.2	17.0-71.5	100	43.7	15.3-65.3	114	48.7	17.9-77.5
18	217	46.8	15.1-79.1	117	53.5	21.5-81.3	100	43.1	10.1-78.1
19	199	34.8	13.6-75.0	100	40.7	18.1-80.2	99	28.8	8.5-71.8
Overall	**4668***	**52.1**	**27.3-76.4**	**2268**	**51.3**	**27.0-74.9**	**2400**	**53.2**	**27.5-78.2**

*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=5292

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	993	21.6	19.8-23.9	476	21.5	19.7-23.8	517	21.6	20.0-23.9
24-27	899	22.0	20.0-24.4	431	21.8	20.1-24.2	468	22.2	20.0-24.7
28-31	849	22.3	20.4-24.8	392	21.8	20.0-24.3	457	22.9	20.9-25.1
32-35	695	22.7	20.8-25.4	303	21.9	19.9-24.5	392	23.3	21.3-26.0
36-39	559	23.1	21.0-25.4	249	22.3	20.4-24.8	310	23.7	21.6-25.7
40-43	378	23.5	21.3-25.9	151	22.2	20.4-24.9	227	24.2	22.0-26.3
44-47	254	24.4	22.0-27.1	110	23.0	21.0-26.3	144	25.0	22.8-27.3
48-51	229	24.3	22.1-27.0	97	23.7	21.2-26.9	132	24.5	22.4-27.0
52-55	163	24.5	21.9-27.5	72	24.5	21.6-27.7	91	24.8	22.1-27.2
56-59	109	24.4	22.4-27.3	43	23.2	21.1-26.8	66	24.7	23.0-27.5
60+	144	25.0	21.8-28.4	67	23.6	20.0-28.3	77	25.6	23.2-28.5
Overall	**5272**	**22.7**	**20.5-25.3**	**2391**	**22.1**	**20.1-24.7**	**2881**	**23.1**	**21.0-25.7**

*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=6104

The following table shows how people with CF reported their education and employment status in 2019.

	2016	2017	2018	2019		
				Overall	Male	Female
Number of patients; N	5851	5989	5952	6104	3294	2810
Number who completed questionnaire; n (%)	5791 (99.0)	5937 (99.1)	5950 (100.0)	6103 (100.0)	3294 (100.0)	2809 (100.0)
Full-time employment; n (%)	1887 (32.2)	1949 (32.5)	1956 (32.9)	2048 (33.6)	1380 (41.9)	668 (23.8)
Part-time employment; n (%)	827 (14.1)	887 (14.8)	926 (15.6)	958 (15.7)	360 (10.9)	598 (21.3)
Student; n (%)	946 (16.3)	973 (16.2)	937 (15.7)	969 (15.9)	483 (14.7)	486 (17.3)
Homemaker; n (%)	242 (4.1)	246 (4.1)	237 (4.0)	231 (3.8)	28 (0.9)	203 (7.2)
Unemployed; n (%)	784 (13.4)	837 (14.0)	814 (13.7)	825 (13.5)	480 (14.6)	345 (12.3)
Disabled; n (%)	359 (6.1)	352 (5.9)	359 (6.0)	327 (5.4)	170 (5.2)	157 (5.6)
Retired; n (%)	116 (2.0)	120 (2.0)	133 (2.2)	145 (2.4)	82 (2.5)	63 (2.2)
Volunteer; n (%)	-*	-*	-*	-**	<5	5 (0.2)
Unknown entered; n (%)	630 (10.8)	573 (9.6)	588 (9.9)	592 (9.7)	308 (9.4)	284 (10.1)
No. in work or study; n (%)	**3902 (67.5)**	**3809 (64.2)**	**3819 (64.2)**	**3975 (65.1)**	**2223 (67.5)**	**1752 (62.4)**

1.10 Pregnancy

	2016	2017	2018	2019
Women with CF that had babies; n	71	58	65	58
Men with CF who became fathers; n	48	44	45	45

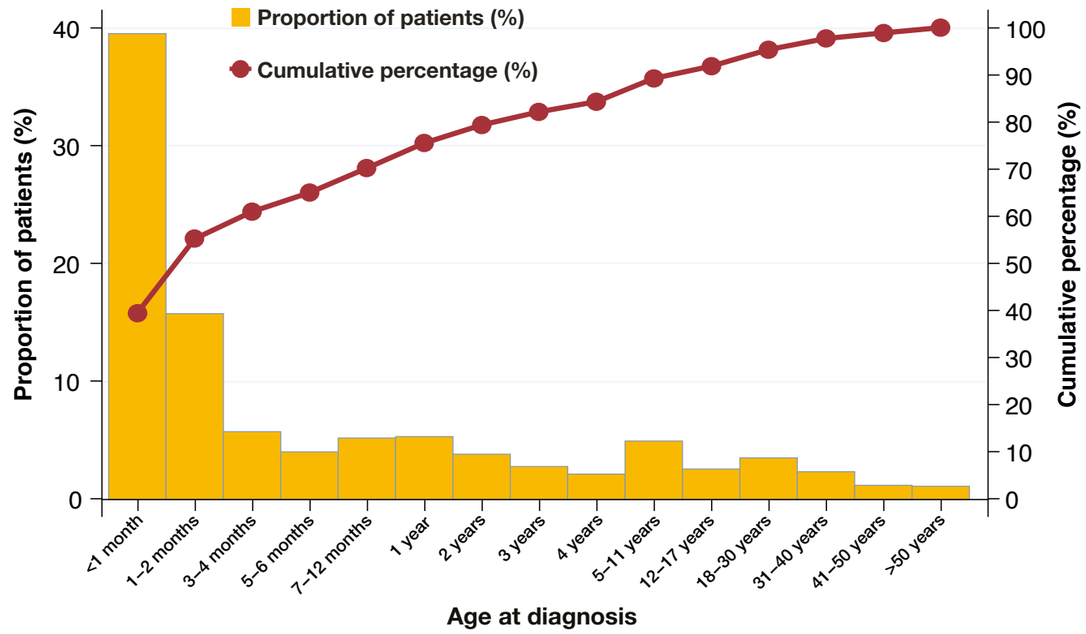
58 women with CF had babies in 2019

45 men with CF became fathers in 2019

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

Diagnosis of cystic fibrosis

1.11 Age at diagnosis in 2019
N=10070



The median age at diagnosis for patients aged under 16 in 2019 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 **137 (71.0%)** out of 193 patients born in 2019 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 2018 is higher (167) in this report than was recorded in the previous.

887 (14.9%) of adults with CF in the Registry in 2019 were diagnosed at age 16 or over.

In 2019, **28** people aged 16 or over were newly diagnosed with cystic fibrosis.

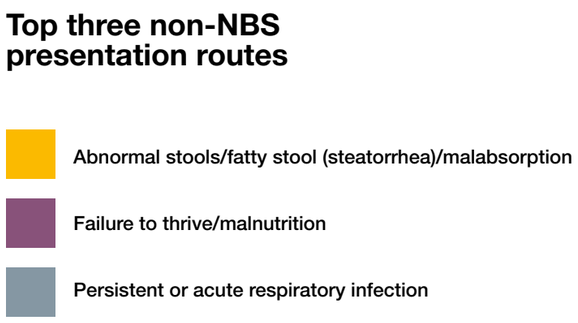
1.12 Mode of presentation

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.

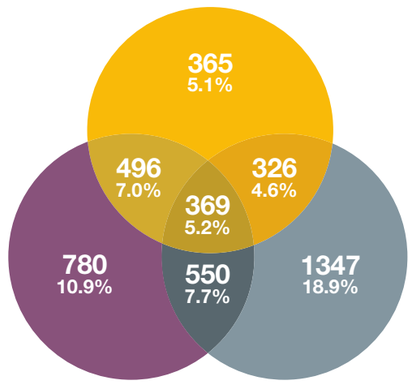
	All patients	Age <16 at diagnosis*	Age ≥16 at diagnosis*
Total patients	10070	9169	887
Number diagnosed by newborn screening (NBS)	2952	2952	0
Total non-NBS	**7118**	**6217**	**887**

Mode of presentation (excluding NBS)	All patients (N=7118)		Age <16 at diagnosis* (n=6217)		Age ≥16 at diagnosis* (n=887)	
	n	(%)	n	(%)	n	(%)
Persistent or acute respiratory infection	2592	36.3%	2101	33.7%	491	56.2%
Failure to thrive/malnutrition	2195	30.8%	2169	34.8%	26	3.0%
Abnormal stools/fatty stool (steatorrhea)/malabsorption	1556	21.8%	1503	24.1%	53	6.1%
Meconium ileus	1306	18.3%	1301	20.9%	5	0.6%
Family history	914	12.8%	789	12.7%	125	14.3%
Genotype	681	9.5%	482	7.7%	199	22.8%
Unknown	331	4.6%	278	4.5%	53	6.1%
Rectal prolapse	242	3.4%	241	3.9%	<5	0.1%
Nasal polyps	149	2.1%	80	1.3%	69	7.9%
Bronchiectasis	75	1.1%	5	0.1%	70	8.0%
Prenatal	70	1.0%	69	1.1%	0	0
Electrolyte imbalance	69	1.0%	63	1.0%	6	0.7%
Liver disease	43	0.6%	41	0.7%	<5	0.2%
Fertility	32	0.4%	<5	0.0%	31	3.5%
Pancreatitis	16	0.2%	<5	0.0%	13	1.5%
Oedema	9	0.1%	9	0.1%	0	0.0%

Top three non-NBS presentation routes



Other: 2829 (39.9%)



*Age-stratified figures are presented only for those with non-missing diagnosis date. This means that the number of people in <16 and ≥16 age groups will not necessarily add up to the 'All patients' number, which is shown for all patients, even if the diagnosis date is missing.

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=8419

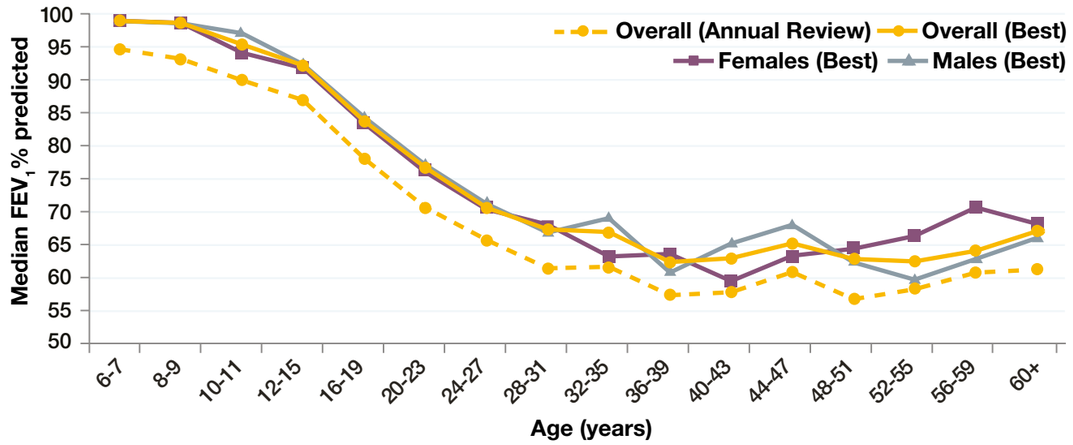
People with CF who have had lung transplants are excluded, as their new 'non-CF' lungs may have lung health similar to a person without cystic fibrosis.

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	529	94.7	82.3-103.2	258	95.3	81.5-104.0	271	94.3	83.3-102.9
8-9	555	93.2	82.0-100.7	264	92.9	82.2-100.3	291	93.3	81.8-100.9
10-11	509	89.9	80.3-98.6	260	89.3	78.5-98.2	249	90.6	82.1-99.1
12-15	962	86.9	74.0-96.7	473	86.9	72.2-96.8	489	86.8	75.4-96.6
16-19	791	78.0	60.9-89.6	400	77.9	60.4-88.5	391	78.1	61.2-90.2
20-23	954	70.6	50.3-86.3	459	69.9	48.4-86.1	495	71.7	52.0-86.8
24-27	846	65.6	47.1-83.1	409	64.0	47.1-83.2	437	68.3	47.2-82.6
28-31	776	61.3	43.4-78.9	355	62.7	45.2-80.3	421	60.3	42.0-77.1
32-35	620	61.5	42.2-78.3	266	57.1	40.5-76.6	354	64.0	43.6-79.8
36-39	487	57.2	37.9-78.8	212	57.6	41.2-79.5	275	56.6	36.7-77.0
40-43	339	57.6	40.5-76.9	128	54.6	39.4-73.6	211	58.8	41.0-79.2
44-47	220	60.7	45.0-80.2	97	56.4	42.3-74.4	123	65.3	49.2-83.4
48-51	200	56.6	40.9-75.5	82	59.1	44.5-72.0	118	56.0	38.5-76.6
52-55	146	58.1	39.3-76.3	69	59.7	41.9-75.5	77	56.3	38.5-79.2
56-59	97	60.6	44.0-78.9	38	58.2	46.3-80.3	59	62.6	35.3-77.9
60+	133	61.1	42.3-80.4	61	57.8	46.3-77.4	72	61.6	37.9-81.1
<16	2555	90.5	79.1-99.7	1255	90.5	78.1-99.5	1300	90.6	80.3-100.0
≥16	5609	65.8	45.7-83.1	2576	65.4	45.7-82.8	3033	66.3	45.8-83.2
<18	2939	89.5	77.1-99.1	1440	89.1	75.6-98.9	1499	89.8	78.3-99.3
≥18	5225	64.1	44.5-82.1	2391	63.9	44.8-82.1	2834	64.2	44.2-82.1
Overall	**8164***	**75.2**	**53.9-91.3**	**3831**	**75.1**	**53.7-91.6**	**4333**	**75.3**	**54.0-91.0**

*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=8419



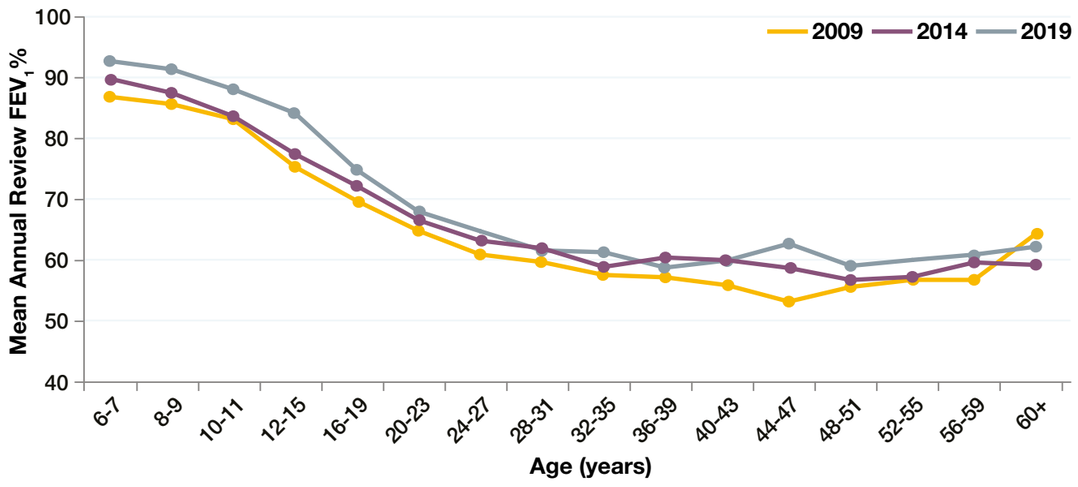
Where Best FEV_1% was missing or less than the FEV_1% at annual review, annual review FEV_1% was used instead.

	Overall			Female			Male		
Age (yrs)	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**
6-7	538	98.9	90.2-107.4	263	99.0	89.6-108.8	275	98.9	90.7-106.4
8-9	568	98.7	89.3-106.0	272	98.7	89.4-106.1	296	98.6	89.1-105.7
10-11	512	95.4	85.4-103.5	261	94.1	83.9-103.6	251	97.1	87.3-103.3
12-15	973	92.2	82.3-100.4	479	91.8	80.0-100.3	494	92.3	83.0-100.4
16-19	798	83.7	69.3-94.6	402	83.4	68.3-93.5	396	84.3	70.7-95.4
20-23	969	76.6	58.2-91.4	464	75.9	56.6-90.9	505	77.0	59.1-92.0
24-27	862	70.5	53.2-87.6	416	70.3	53.5-88.1	446	71.1	53.2-86.9
28-31	799	67.2	47.3-83.7	367	67.9	49.4-84.5	432	66.7	46.3-82.9
32-35	634	66.8	47.1-83.2	271	63.1	44.9-81.5	363	68.9	48.4-84.5
36-39	499	62.2	42.7-82.1	220	63.5	47.0-81.9	279	60.6	40.3-82.9
40-43	346	62.8	44.8-80.6	130	59.3	45.3-77.0	216	65.1	44.6-84.2
44-47	227	65.1	48.8-83.0	98	63.2	46.6-78.6	129	67.9	51.9-85.2
48-51	203	62.7	45.6-80.3	83	64.3	47.3-82.8	120	62.2	43.5-79.4
52-55	148	62.3	44.7-84.1	70	66.2	46.5-84.4	78	59.6	42.4-82.7
56-59	100	63.9	46.9-83.3	39	70.6	51.2-84.8	61	62.7	38.5-79.5
60+	137	66.9	46.3-84.9	63	68.1	51.1-81.2	74	65.9	40.2-86.3
<16	2591	95.7	85.6-103.6	1275	95.4	84.8-103.7	1316	95.9	86.6-103.6
≥16	5722	71.1	51.3-87.5	2623	70.7	51.8-87.4	3099	71.4	50.6-87.6
<18	2977	94.6	84.1-103.1	1461	94.1	83.3-103.0	1516	95.3	85.2-103.1
≥18	5336	69.8	50.1-86.6	2437	69.6	50.9-86.7	2899	70.0	49.0-86.5
Overall	**8313***	**81.2**	**59.6-95.7**	**3898**	**81.1**	**60.0-95.9**	**4415**	**81.2**	**59.4-95.6**

*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=8419 in 2019, N=7642 in 2014, N=5999 in 2009*

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 2019 compares to Registry data from 2009 and 2014. 2009 is shown as a comparator year as this is the earliest year that we can be confident that the coverage of the Registry gives an accurate reflection of the CF population.

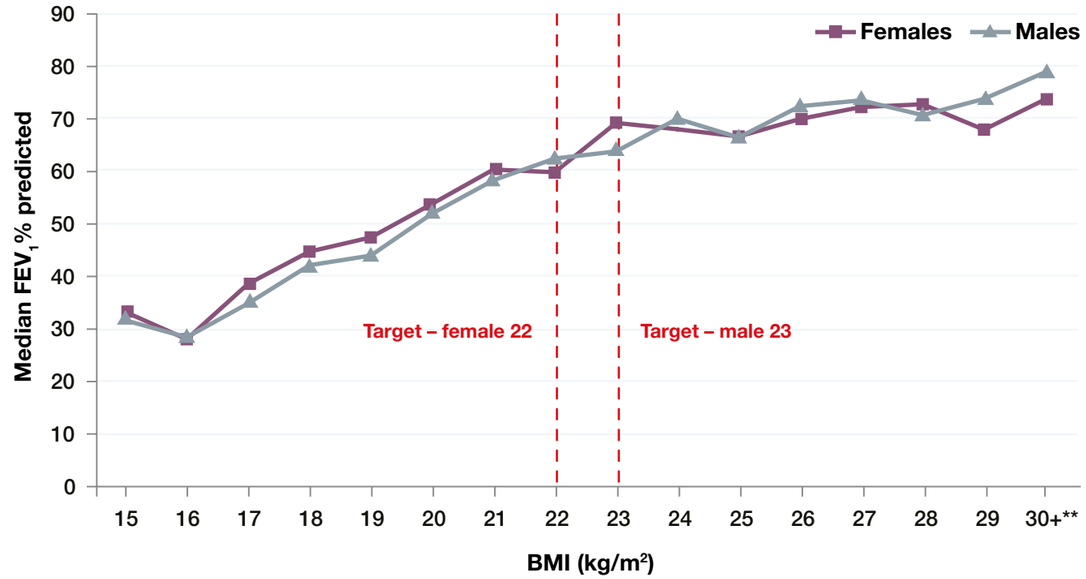


Age (years)	2009 mean FEV_1%	2014 mean FEV_1%	2019 mean FEV_1%	p-values (t-test)**
6-7	87.0	89.9	92.8	0.005
8-9	85.8	87.6	91.5	<0.001
10-11	83.3	83.8	88.2	<0.001
12-15	75.4	77.5	84.3	<0.001
16-19	69.8	72.3	74.9	0.009
20-23	64.9	66.6	68.1	0.184
24-27	61.0	63.3	64.8	0.177
28-31	59.8	62.0	61.7	0.762
32-35	57.6	59.0	61.4	0.083
36-39	57.2	60.5	58.8	0.343
40-43	55.9	60.1	60.0	0.988
44-47	53.2	58.8	62.8	0.090
48-51	55.7	56.8	59.1	0.353
52-55	56.8	57.3	60.1	0.419
56-59	56.8	59.7	61.0	0.764
60+	64.4	59.3	62.3	0.404
<16	80.8	83.3	88.4	-
16+	62.6	64.1	64.6	-
<18	79.3	81.7	87.0	-
18+	61.4	63.1	63.7	-

*Due to missing data, means are calculated from a population of 8164 in 2019, 7180 in 2014 and 5388 in 2009.
** t-test comparing 2019 with 2014.

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.

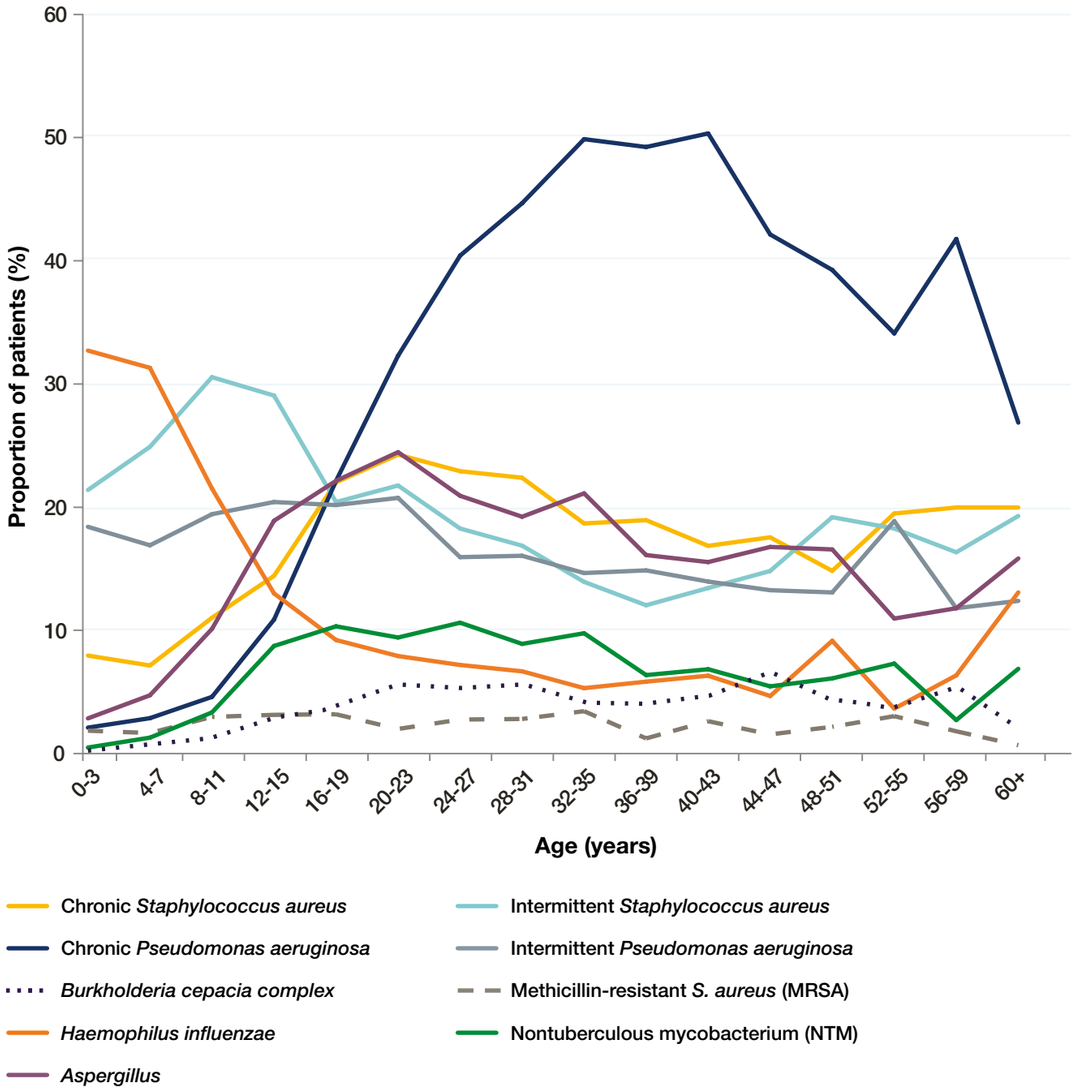


* Due to missing data, medians are calculated from a population of 4808.
** Each point represents the median FEV_1% predicted of patients for each given BMI value. Due to the wide range of BMIs in this population with a value of 30 of more, these are grouped into one.

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 2019
N=10070*



*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 2019
<16 years N=3966, ≥16 years N=6104

	Paediatric age range (years)				Overall
	0-3	4-7	8-11	12-15	<16 years
Number in age range	803	1075	1105	983	3966
Number who had culture taken*	795	1062	1098	979	3934
Chronic Staphylococcus aureus; n (%)	64 (8.0)	77 (7.2)	122 (11.0)	142 (14.4)	405 (10.2)
Intermittent Staphylococcus aureus; n (%)	172 (21.4)	268 (24.9)	338 (30.6)	286 (29.1)	1064 (26.8)
Chronic Pseudomonas aeruginosa; n (%)	17 (2.1)	31 (2.9)	51 (4.6)	107 (10.9)	206 (5.2)
Intermittent Pseudomonas aeruginosa; n (%)	148 (18.4)	182 (16.9)	215 (19.5)	201 (20.4)	746 (18.8)
Burkholderia cepacia complex; n (%)	<5	>5**	14 (1.3)	28 (2.8)	52 (1.3)
B. cenocepacia; n (%)	<5	<5	<5	6 (0.6)	12 (0.3)
B. multivorans; n (%)	<5	<5	8 (0.7)	9 (0.9)	20 (0.5)
B. other cepacia; n (%)	<5	<5	<5	8 (0.8)	14 (0.4)
MRSA; n (%)	15 (1.9)	18 (1.7)	33 (3.0)	31 (3.2)	97 (2.4)
Haemophilus influenzae; n (%)	263 (32.8)	337 (31.3)	238 (21.5)	128 (13.0)	966 (24.4)
Nontuberculous mycobacterium; n (%)	<5	14 (1.3)	37 (3.3)	86 (8.7)	141 (3.6)
Aspergillus; n (%)	23 (2.9)	51 (4.7)	112 (10.1)	186 (18.9)	372 (9.4)

Infections in this table reflect bugs grown in the 12 months prior to the 2019 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 previous years 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	812	996	902	852	695	564	6104
Number who had culture taken*	795	975	880	814	662	538	5902
Chronic S. aureus; n (%)	179 (22.0)	242 (24.3)	207 (22.9)	191 (22.4)	130 (18.7)	107 (19.0)	1282 (21.0)
Intermittent S. aureus; n (%)	166 (20.4)	217 (21.8)	165 (18.3)	144 (16.9)	97 (14.0)	68 (12.1)	1066 (17.5)
Chronic P. aeruginosa; n (%)	180 (22.2)	322 (32.3)	365 (40.5)	381 (44.7)	347 (49.9)	278 (49.3)	2403 (39.4)
Intermittent P. aeruginosa; n (%)	164 (20.2)	207 (20.8)	144 (16.0)	137 (16.1)	102 (14.7)	84 (14.9)	1017 (16.7)
B. cepacia complex; n (%)	31 (3.8)	56 (5.6)	48 (5.3)	48 (5.6)	29 (4.2)	23 (4.1)	295 (4.8)
B. cenocepacia; n (%)	7 (0.9)	10 (1.0)	18 (2.0)	9 (1.1)	7 (1.0)	6 (1.1)	77 (1.3)
B. multivorans; n (%)	15 (1.8)	31 (3.1)	22 (2.4)	27 (3.2)	17 (2.4)	14 (2.5)	153 (2.5)
B. other cepacia; n (%)	<5	9 (0.9)	<5	6 (0.7)	<5	<5	34 (0.6)
Methicillin-resistant S. aureus; n (%)	26 (3.2)	20 (2.0)	25 (2.8)	24 (2.8)	24 (3.5)	7 (1.2)	153 (2.5)
H. influenzae; n (%)	75 (9.2)	79 (7.9)	65 (7.2)	57 (6.7)	37 (5.3)	33 (5.9)	435 (7.1)
NTM; n (%)	84 (10.3)	94 (9.4)	96 (10.6)	76 (8.9)	68 (9.8)	36 (6.4)	533 (8.7)
Aspergillus; n (%)	180 (22.2)	244 (24.5)	189 (21.0)	164 (19.2)	147 (21.2)	91 (16.1)	1209 (19.8)

	Adult age range (years)						Overall
	40-43	44-47	48-51	52-55	56-59	60+	≥16 years
Number in age range	379	256	229	164	110	145	6104
Number who had culture taken*	367	247	221	158	104	141	5902
Chronic S. aureus; n (%)	64 (16.9)	45 (17.6)	34 (14.8)	32 (19.5)	22 (20.0)	29 (20.0)	1282 (21.0)
Intermittent S. aureus; n (%)	51 (13.5)	38 (14.8)	44 (19.2)	30 (18.3)	18 (16.4)	28 (19.3)	1066 (17.5)
Chronic P. aeruginosa; n (%)	191 (50.4)	108 (42.2)	90 (39.3)	56 (34.1)	46 (41.8)	39 (26.9)	2403 (39.4)
Intermittent P. aeruginosa; n (%)	53 (14.0)	34 (13.3)	30 (13.1)	31 (18.9)	13 (11.8)	18 (12.4)	1017 (16.7)
B. cepacia complex; n (%)	18 (4.7)	17 (6.6)	10 (4.4)	6 (3.7)	6 (5.5)	<5	295 (4.8)
B. cenocepacia; n (%)	6 (1.6)	<5	<5	5 (3.0)	<5	<5	77 (1.3)
B. multivorans; n (%)	9 (2.4)	7 (2.7)	6 (2.6)	<5	<5	<5	153 (2.5)
B. other cepacia; n (%)	<5	<5	<5	<5	<5	<5	34 (0.6)
MRSA; n (%)	10 (2.6)	<5	5 (2.2)	5 (3.0)	<5	<5	153 (2.5)
H. influenzae; n (%)	24 (6.3)	12 (4.7)	21 (9.2)	6 (3.7)	7 (6.4)	19 (13.1)	435 (7.1)
NTM; n (%)	26 (6.9)	14 (5.5)	14 (6.1)	12 (7.3)	<5	>5	533 (8.7)
Aspergillus; n (%)	59 (15.6)	43 (16.8)	38 (16.6)	18 (11.0)	13 (11.8)	23 (15.9)	1209 (19.8)

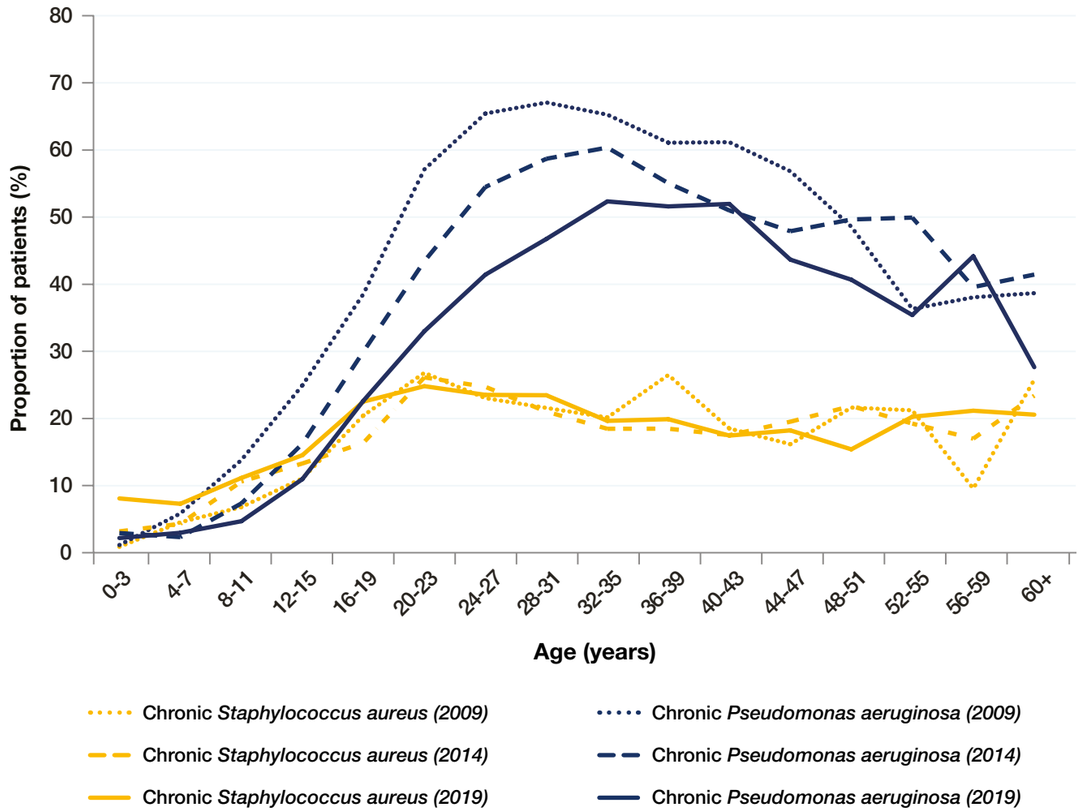
* Proportions are calculated from number in age range, whereas previous years were calculated from the number of people with CF who were recorded as having had a culture taken.

1.19 Nontuberculous 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.

	2017 (n=9887)	2018 (n=9847)	2019 (n=10070)
NTM prevalence; n (%)	592 (6.0)	697 (7.1)	674 (6.7)
On NTM treatment in the given year; n (% of NTM prevalence in given year)	352 (59.5)	343 (49.2)	362 (53.7)
NTM incidence [1]; n (%)	246 (2.7)	293 (3.2)	279 (3.0)
M. abscessus prevalence	376 (3.8)	419 (4.3)	382 (3.8)
M. abscessus incidence [2]; n (%)	136 (1.4)	157 (1.7)	126 (1.3)

1.20 Lung infections over time
N=7377 in 2009, N=9432 in 2014, N=10070 in 2019



The median age of people with chronic *Pseudomonas aeruginosa* infection increased from 24 years in 2009 to 30 years in 2019.

[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)	**2009 (%)**	**2014 (%)**	**2019 (%)**	**p-value***
0-3	0.8	3.1	8.1	<0.001
4-7	4.5	4.3	7.3	0.004
8-11	6.8	10.6	11.1	0.701
12-15	11.0	13.3	14.5	0.433
16-19	20.4	16.4	22.5	0.001
20-23	26.8	26.1	24.8	0.462
24-27	23.0	24.7	23.5	0.519
28-31	21.6	21.0	23.5	0.326
32-35	20.1	18.5	19.6	0.733
36-39	26.5	18.5	19.9	0.649
40-43	18.5	17.5	17.4	0.971
44-47	16.2	19.5	18.2	0.794
48-51	21.6	21.9	15.4	0.108
52-55	21.2	19.2	20.3	0.811
56-59	9.5	17.0	21.2	0.608
60+	25.8	23.4	20.6	0.616
<16 years	6.0	7.8	10.3	-
≥16 years	22.5	21.0	21.7	-
<18 years	7.5	8.4	11.4	-
≥18 years	23.0	21.8	21.7	-

Chronic *Pseudomonas aeruginosa*				
Age (years)	**2009 (%)**	**2014 (%)**	**2019 (%)**	**p-value***
0-3	1.1	2.9	2.1	0.349
4-7	5.8	2.3	2.9	0.363
8-11	13.8	7.3	4.6	0.011
12-15	24.9	16.2	10.9	<0.001
16-19	38.6	30.0	22.6	<0.001
20-23	57.2	43.5	33.0	<0.001
24-27	65.5	54.6	41.5	<0.001
28-31	67.2	58.8	46.8	<0.001
32-35	65.4	60.5	52.4	0.001
36-39	61.2	55.1	51.7	0.236
40-43	61.3	51.1	52.0	0.715
44-47	56.9	48.0	43.7	0.464
48-51	48.6	49.7	40.7	0.095
52-55	36.4	50.0	35.4	0.025
56-59	38.1	39.6	44.2	0.720
60+	38.7	41.5	27.7	0.030
<16 years	12.1	7.0	5.2	-
≥16 years	56.0	48.3	40.7	-
<18 years	14.1	9.3	6.3	-
≥18 years	59.6	50.5	42.4	-

*The proportion of people with each infection within each age group was compared between 2014 and 2019. If the p-value is less than 0.05 then the difference in the proportions is statistically significant.

Complications

1.21 Complications in 2019

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

	Overall (N=10070)	**<16 years (n=3966)**	**≥16 years (n=6104)**
	n (%)		
Respiratory related			
Nasal polyps requiring surgery	426 (4.2)	120 (3.0)	306 (5.0)
Sinus disease	797 (7.9)	52 (1.3)	745 (12.2)
Asthma	925 (9.2)	272 (6.9)	653 (10.7)
ABPA	754 (7.5)	138 (3.5)	616 (10.1)
Any haemoptysis	420 (4.2)	16 (0.4)	404 (6.6)
Massive haemoptysis	-*	<5	36 (0.6)
Pneumothorax requiring chest tube	27 (0.3)	0	27 (0.4)
Pancreas and hepatobiliary disease			
Raised liver enzymes	1010 (10.0)	302 (7.6)	708 (11.6)
Liver disease	1467 (14.6)	357 (9.0)	1110 (18.2)
Cirrhosis with no portal hypertension	63 (0.6)	15 (0.4)	48 (0.8)
Cirrhosis with portal hypertension	135 (1.3)	28 (0.7)	107 (1.8)
Gall bladder disease requiring surgery	143 (1.4)	24 (0.6)	119 (1.9)
Pancreatitis	65 (0.6)	9 (0.2)	56 (0.9)
Upper gastrointestinal (GI)			
GERD	1696 (16.8)	252 (6.4)	1444 (23.7)
Peptic ulcer	<5	0	<5
GI bleed (varices as source)	-*	<5	10 (0.2)
GI bleed (non varices as source)	-*	<5	11 (0.2)
Lower gastrointestinal			
Intestinal obstruction	34 (0.3)	12 (0.3)	22 (0.4)
DIOS	570 (5.7)	100 (2.5)	470 (7.7)
Fibrosing colonopathy/colonic stricture	<5	0	<5
Rectal prolapse	12 (0.1)	7 (0.2)	5 (0.1)
Renal			
Kidney stones	111 (1.1)	11 (0.3)	100 (1.6)
Renal failure	97 (1.0)	5 (0.1)	92 (1.5)
Musculoskeletal			
Arthritis	100 (1.0)	9 (0.2)	91 (1.5)
Arthropathy	250 (2.5)	14 (0.4)	236 (3.9)
Bone fracture	47 (0.5)	13 (0.3)	34 (0.6)
Osteopenia	975 (9.7)	25 (0.6)	950 (15.6)
Osteoporosis	414 (4.1)	5 (0.1)	409 (6.7)
Other			
Cancer confirmed by histology	28 (0.3)	0	28 (0.5)
Port inserted or replaced	273 (2.7)	85 (2.1)	188 (3.1)
Depression	446 (4.4)	14 (0.4)	432 (7.1)
Hearing loss	339 (3.4)	39 (1.0)	300 (4.9)
Hypertension	126 (1.3)	5 (0.1)	121 (2.0)

* Redacted to adhere to statistical disclosure guidelines.

1.22 Incidence of complications

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

	2018			2019		
	Overall (N=9847	<16 years (n=3895)	≥16 years (n=5952)	Overall (N=10070)	<16 years (n=3966)	≥16 years (n=6104)
ABPA; n (%)	219 (2.2)	70 (1.8)	149 (2.5)	225 (2.2)	62 (1.6)	163 (2.7)
Cirrhosis with no portal hypertension; n (%)	38 (0.4)	11 (0.3)	27 (0.5)	36 (0.4)	9 (0.2)	27 (0.4)
Cirrhosis with portal hypertension; n (%)	38 (0.4)	10 (0.3)	28 (0.5)	54 (0.5)	12 (0.3)	42 (0.7)
Cancer confirmed by histology; n (%)	17 (0.2)	0	17 (0.3)	20 (0.2)	0	20 (0.3)

1.23 CF-related diabetes
N=7611

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=7611)	10-15 years (n=1507)	≥16 years (n=6104)
On CFRD treatment; n (%)	2267 (29.8)	197 (13.1)	2070 (33.9)
Of those on treatment			
Insulin[1]; n (%)	2038 (89.9)	194 (98.5)	1844 (89.1)
CFRD Screening; n (%)			
Yes	4015 (52.8)	1113 (73.9)	2902 (47.5)
Screening type			
Continuous glucose monitoring[2]; n (%)	1301 (32.4)	335 (30.1)	966 (33.3)
Oral glucose tolerance test[2]; n (%)	2121 (52.8)	601 (54.0)	1520 (52.4)
Not screened (known CFRD)	2003 (26.3)	132 (8.8)	1871 (30.7)
Not screened (other)	1480 (19.4)	245 (16.3)	1235 (20.2)
Unknown	109 (1.4)	16 (1.1)	93 (1.5)

[1] Proportion of patients on treatment
[2] Proportion of patients screened

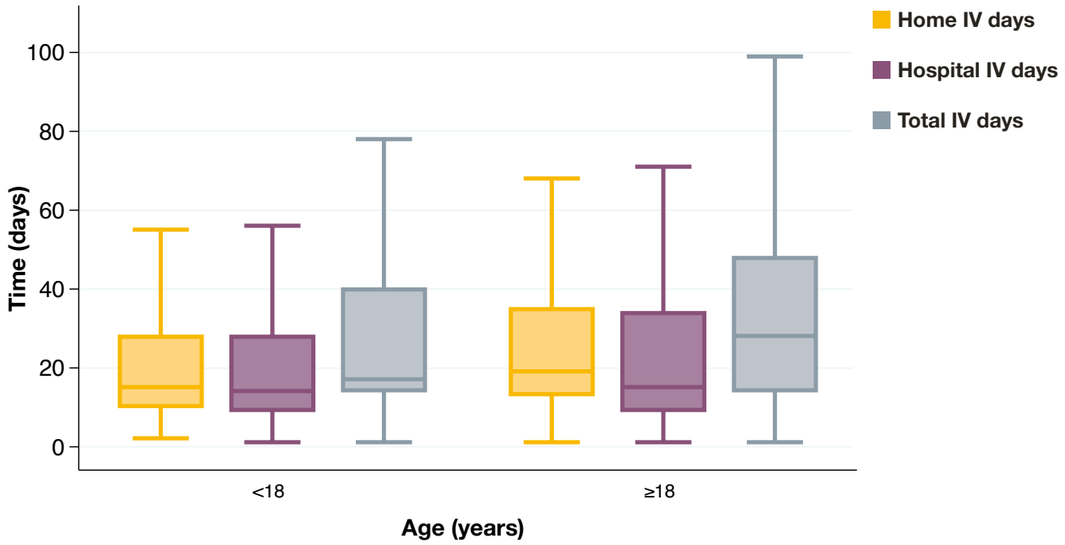
Antibiotics

1.24 Intravenous (IV) antibiotics
N=10070

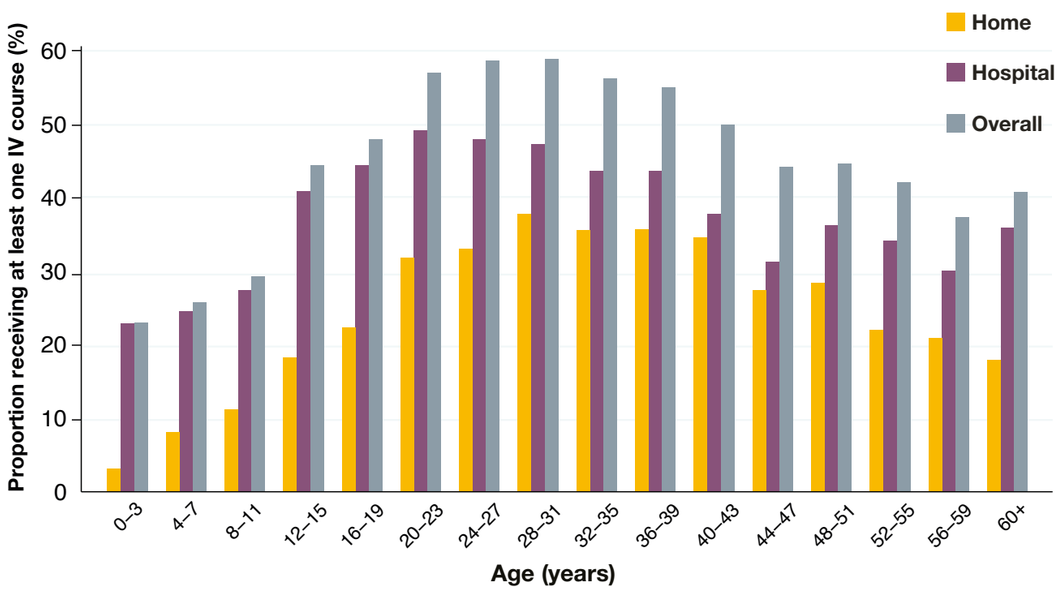
When someone with CF becomes unwell with an infection, they might be prescribed intravenous (IV) antibiotics. IV antibiotics are given to the patient through their veins. This treatment can take a number of days and might take place as a hospital inpatient, or at home.

		Home		Hospital		Total	
Age	N	Patients n (%)	Median days (IQR)	Patients n (%)	Median days (IQR)	Patients n (%)	Median days (IQR)
0-3	803	25 (3.1)	7 (5-12)	183 (22.8)	14 (5-12)	184 (22.9)	14 (9-17)
4-7	1075	86 (8.0)	14 (8-24)	263 (24.5)	14 (8-24)	276 (25.7)	14 (13-28)
8-11	1105	123 (11.1)	18 (11-28)	302 (27.3)	14 (11-28)	323 (29.2)	25 (14-41)
12-15	983	179 (18.2)	16 (11-33)	401 (40.8)	15 (11-33)	436 (44.4)	26 (14-43)
16-19	812	181 (22.3)	14 (10-28)	360 (44.3)	16 (10-28)	389 (47.9)	26 (14-42)
20-23	996	317 (31.8)	15 (11-31)	489 (49.1)	16 (11-31)	567 (56.9)	28 (14-49)
24-27	902	298 (33.0)	21 (13-39)	432 (47.9)	16 (13-39)	528 (58.5)	28 (14-49)
28-31	852	322 (37.8)	21 (13-35)	402 (47.2)	17 (13-35)	501 (58.8)	28 (14-50)
32-35	695	247 (35.5)	20 (13-35)	303 (43.6)	14 (13-35)	390 (56.1)	28 (14-49)
36-39	564	201 (35.6)	23 (14-42)	246 (43.6)	14 (14-42)	310 (55.0)	29 (14-52)
40-43	379	131 (34.6)	22 (13-42)	143 (37.7)	14 (13-42)	189 (49.9)	28 (14-47)
44-47	256	70 (27.3)	14 (9-39)	80 (31.3)	15 (9-39)	113 (44.1)	22 (14-43)
48-51	229	65 (28.4)	20 (13-36)	83 (36.2)	15 (13-36)	102 (44.5)	29 (14-47)
52-55	164	36 (22.0)	21 (14-32)	56 (34.1)	21 (14-32)	69 (42.1)	28 (14-42)
56-59	110	23 (20.9)	14 (13-21)	33 (30.0)	12 (13-21)	41 (37.3)	18 (14-40)
60+	145	26 (17.9)	14 (10-20)	52 (35.9)	13 (10-20)	59 (40.7)	17 (13-28)
<16	3966	413 (10.4)	14 (9-28)	1149 (29.0)	14 (9-28)	1219 (30.7)	16 (14-38)
≥16	6104	1917 (31.4)	19 (12-35)	2679 (43.9)	15 (12-35)	3258 (53.4)	28 (14-48)
<18	4362	493 (11.3)	15 (10-28)	1315 (30.1)	14 (10-28)	1401 (32.1)	17 (14-40)
≥18	5708	1837 (32.2)	19 (13-35)	2513 (44.0)	15 (13-35)	3076 (53.9)	28 (14-48)
Overall	**10070**	**2330 (23.1)**	**18 (12-34)**	**3828 (38.0)**	**14 (12-34)**	**4477 (44.5)**	**26 (14-43)**

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 23.1% and in hospital was 38.0%. The proportion receiving any IVs was 44.5%.



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

	2009			2014			2019		
	Overall	**<16 years**	**≥16 years**	**Overall**	**<16 years**	**≥16 years**	**Overall**	**<16 years**	**≥16 years**
Patients with chronic *P. aeruginosa*; n	2620	390	2230	2963	305	2658	2609	206	2403
Tobramycin solution; n (%)	531 (20.3)	59 (15.1)	472 (21.2)	841 (28.4)	96 (31.5)	745 (28.0)	623 (23.9)	87 (42.2)	536 (22.3)
Other aminoglycoside; n (%)	58 (2.2)	10 (2.6)	48 (2.2)	139 (4.7)	22 (7.2)	117 (4.4)	29 (1.1)	0	29 (1.2)
Colistin; n (%)	1185 (45.2)	209 (53.6)	976 (43.8)	1101 (37.2)	156 (51.1)	945 (35.6)	615 (23.6)	81 (39.3)	534 (22.2)
Promixin; n (%)	543 (20.7)	95 (24.4)	448 (20.1)	919 (31.0)	134 (43.9)	785 (18.2)	771 (29.6)	97 (47.1)	674 (28.0)
Aztreonam; n (%)	-	-	-	395 (13.3)	10 (3.3)	385 (14.5)	690 (26.4)	15 (7.3)	675 (28.1)
Colistimethate (inh) inhalation powder; n (%)	-	-	-	433 (14.6)	21 (6.9)	412 (15.5)	457 (17.5)	11 (5.3)	446 (18.6)
Tobramycin inhalation powder; n (%)	-	-	-	802 (27.1)	24 (7.9)	778 (29.3)	573 (22.0)	9 (4.4)	564 (23.5)
At least one of the above; n (%)	1946 (74.3)	325 (83.3)	1621 (72.7)	2625 (88.6)	288 (94.4)	2337 (87.9)	2315 (88.7)	189 (91.7)	2126 (88.5)

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.

1.26 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 (%)
2009	**Overall**	2574	1491 (57.9)	1083 (42.1)
	0-3 years	17	0	17 (100%)
	4-15 years	489	121 (24.7)	368 (75.3)
	≥16 years	2068	1370 (66.2)	698 (33.8)
2014	**Overall**	3705	1945 (52.5)	1760 (47.5)
	0-3 years	39	5 (12.8)	34 (87.2)
	4-15 years	594	101 (17.0)	493 (83.0)
	≥16 years	3072	1839 (59.9)	1233 (40.1)
2019	**Overall**	4130	1772 (42.9)	2358 (57.1)
	0-3 years	-*	<5	44 (95.7)
	4-15 years	672	99 (14.7)	573 (85.3)
	≥16 years	3412	1671 (49.0)	1741 (51.0)

* Redacted to adhere to statistical disclosure guidelines.

1.27 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	803	454 (56.5)
4-7	1075	297 (27.6)
8-11	1105	294 (26.6)
12-15	983	229 (23.3)
16-19	810	172 (21.2)
20-23	996	113 (11.3)
24-27	902	69 (7.6)
28-31	852	66 (7.7)
32-35	696	35 (5.0)
36-39	564	37 (6.6)
40-43	379	32 (8.4)
44-47	257	16 (6.3)
48-51	229	14 (6.1)
52-55	164	9 (5.5)
56-59	110	5 (4.5)
60+	145	8 (5.5)
<16 years	3966	1274 (32.1)
≥16 years	6104	576 (9.4)
<18 years	4360	1374 (31.5)
≥18 years	5710	476 (8.3)
Overall	**10070**	**1850 (18.4)**

Bronchodilators & Corticosteroids

1.28 Inhaled bronchodilators & corticosteroids

Age; years	Total patients	Patients on inhaled bronchodilators; n (%)	Patients on inhaled corticosteroids; n (%)	Patients on inhaled combination corticosteroids/bronchodilators; n (%)
0-3	803	128 (15.9)	52 (6.5)	0 (0.0)
4-7	1075	426 (39.6)	173 (16.1)	52 (4.8)
8-11	1105	545 (49.3)	205 (18.6)	186 (16.8)
12-15	983	583 (59.3)	165 (16.8)	302 (30.7)
16-19	810	523 (64.6)	147 (18.1)	260 (32.1)
20-23	996	683 (68.6)	197 (19.8)	348 (34.9)
24-27	902	611 (67.7)	183 (20.3)	378 (41.9)
28-31	852	632 (74.2)	167 (19.6)	421 (49.4)
32-35	696	505 (72.8)	140 (20.2)	343 (49.4)
36-39	564	407 (72.2)	125 (22.2)	279 (49.5)
40-43	379	276 (72.8)	71 (18.7)	203 (53.6)
44-47	257	170 (66.4)	47 (18.4)	134 (52.3)
48-51	229	152 (66.4)	58 (25.3)	110 (48.0)
52-55	164	113 (68.9)	39 (23.8)	76 (46.3)
56-59	110	73 (66.4)	24 (21.8)	55 (50.0)
60+	145	105 (72.4)	34 (23.4)	68 (46.9)
<16 years	3966	1682 (42.4)	595 (15.0)	540 (13.6)
≥16 years	6104	4250 (69.7)	1232 (20.2)	2675 (43.8)
<18 years	4360	1921 (44.1)	669 (15.3)	667 (15.3)
≥18 years	5710	4011 (70.3)	1158 (20.3)	2548 (44.6)
Overall	**10070**	**5932 (58.9)**	**1827 (18.1)**	**3215 (31.9)**

Muco-active therapies

1.29 Mannitol

Age; years	Total patients	Patients on Mannitol; n (%)
0-3	803	0
4-7	1075	0
8-11	1105	<5
12-15	983	<5
16-19	810	20 (2.5)
20-23	996	48 (4.8)
24-27	902	55 (6.1)
28-31	852	67 (7.9)
32-35	696	46 (6.6)
36-39	564	35 (6.2)
40-43	379	35 (9.2)
44-47	257	16 (6.3)
48-51	229	10 (4.4)
52-55	164	6 (3.7)
56-59	110	<5
60+	145	<5
<16 years	3966	<5
≥16 years	6104	342 (5.6)
<18 years	4360	9 (0.2)
≥18 years	5710	337 (5.9)
Overall	**10070**	**346 (3.4)**

1.30 DNase

	2009		2014		2019	
Age; years	Total patients	Patients on DNase; n (%)	Total patients	Patients on DNase; n (%)	Total patients	Patients on DNase; n (%)
0-3	801	55 (6.9)	963	114 (11.8)	803	155 (19.3)
4-7	742	152 (20.5)	1044	415 (39.8)	1075	584 (54.3)
8-11	838	332 (39.6)	906	558 (61.6)	1105	858 (77.6)
12-15	931	449 (48.2)	927	648 (69.9)	983	841 (85.6)
16-19	877	441 (50.3)	1020	701 (68.7)	810	682 (84.2)
20-23	861	410 (47.6)	1002	632 (63.1)	996	795 (79.8)
24-27	690	345 (50.0)	932	609 (65.3)	902	684 (75.8)
28-31	502	226 (45.0)	728	447 (61.4)	852	615 (72.2)
32-35	325	127 (39.1)	589	355 (60.3)	696	477 (68.7)
36-39	276	102 (37.0)	361	186 (51.5)	564	376 (66.7)
40-43	209	75 (35.9)	309	150 (48.5)	379	235 (62.0)
44-47	145	61 (42.1)	231	114 (49.4)	257	145 (56.6)
48-51	88	34 (38.6)	179	94 (52.5)	229	133 (58.1)
52-55	36	12 (33.3)	96	47 (49.0)	164	84 (51.2)
56-59	23	6 (26.1)	55	27 (49.1)	110	62 (56.4)
60+	33	6 (18.2)	90	46 (51.1)	145	75 (51.7)
<16 years	3312	988 (29.8)	3840	1735 (45.2)	3966	2438 (61.5)
≥16 years	4065	1845 (45.4)	5592	3408 (60.9)	6104	4363 (71.5)
<18 years	3752	1198 (31.9)	4328	2078 (48.0)	4360	2770 (63.5)
≥18 years	3625	1635 (45.1)	5104	3065 (60.1)	5710	4031 (70.6)
Overall	**7377**	**2833 (38.4)**	**9432**	**5143 (54.5)**	**10070**	**6801 (67.6)**

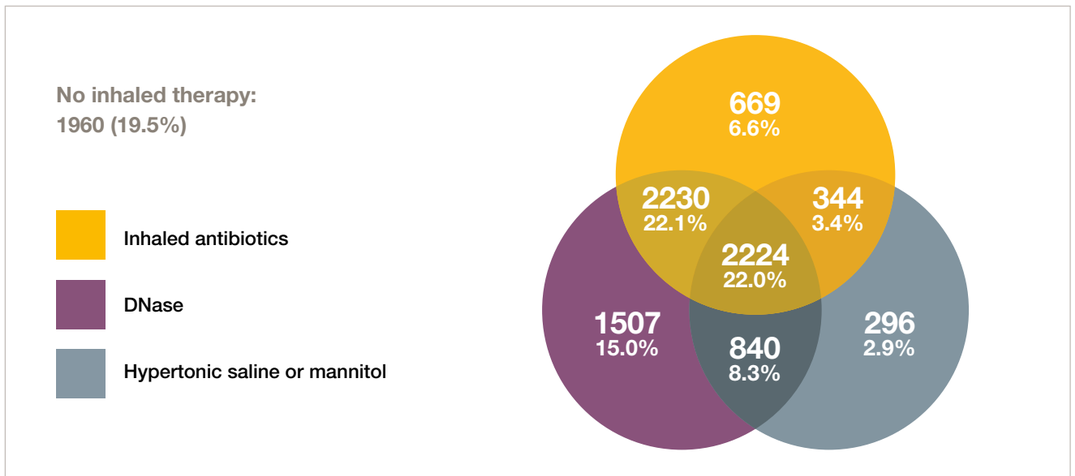
1.31 Hypertonic saline

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

	2009		2014		2019	
Age; years	Total patients; N	Patients on hypertonic saline; n (%)	Total patients; N	Patients on hypertonic saline; n (%)	Total patients; N	Patients on hypertonic saline; n (%)
0-3	801	8 (1.0)	963	64 (6.6)	803	91 (11.3)
4-7	742	19 (2.6)	1044	173 (16.6)	1075	304 (28.3)
8-11	838	44 (5.3)	906	234 (25.8)	1105	416 (37.6)
12-15	931	74 (7.9)	927	340 (36.7)	983	458 (46.6)
16-19	877	69 (7.9)	1020	328 (32.2)	810	409 (50.5)
20-23	861	63 (7.3)	1002	285 (28.4)	996	389 (39.1)
24-27	690	74 (10.7)	932	269 (28.9)	902	307 (34.0)
28-31	502	59 (11.8)	728	251 (34.5)	852	269 (31.6)
32-35	325	40 (12.3)	589	186 (31.6)	696	233 (33.6)
36-39	276	23 (8.3)	361	96 (26.6)	564	202 (35.8)
40-43	209	14 (6.7)	309	65 (21.0)	379	123 (32.5)
44-47	145	12 (8.3)	231	63 (27.3)	257	80 (31.3)
48-51	88	6 (6.8)	179	45 (25.1)	229	65 (28.4)
52-55	36	<5	96	27 (28.1)	164	46 (28.0)
56-59	23	<5	55	14 (25.5)	110	28 (25.5)
60+	33	<5	90	26 (28.9)	145	40 (27.6)
<16 years	3312	145 (4.4)	3840	811 (21.1)	3966	1269 (32.0)
≥16 years	4065	366 (9.0)	5592	1655 (29.6)	6104	2191 (35.9)
<18 years	3752	181 (4.8)	4328	970 (22.4)	4360	1473 (33.8)
≥18 years	3625	330 (9.1)	5104	1496 (29.3)	5710	1987 (34.8)
Overall	**7377**	**511 (6.9)**	**9432**	**2466 (26.1)**	**10070**	**3460 (34.4)**

1.32 Burden of treatment

The Venn diagram shows how many people with CF are on one or more inhaled therapy and the combinations they take. 1960 (19.5%) people are on no inhaled therapies.



Other therapies

1.33 CFTR modifiers

Ivacaftor

Ivacaftor was first approved for use on the NHS in England in January 2013. Soon after, it was made available in Wales, Scotland and Northern Ireland. Since this time, ivacaftor's license has expanded across age ranges and mutation types. As of 2019, ivacaftor was approved for use on the NHS across the UK for people aged one and older with at least one copy of 9 specified CFTR 'gating' mutations, and for people aged 18 and over with the R117H mutation. In June 2020, the marketing authorisation was extended to cover anyone aged 6 months and over with the R117H mutation.

	Age (at annual review)	N
Patients on ivacaftor in the UK	Overall	646
	<6 years	59
	≥6 years	587
Patients stopped ivacaftor ever	Overall	39
	<6 years	0
	≥6 years	39

Tests	Age (at start date)	Median (IQR)	Number with complete data; n(%)
Sweat chloride before ivacaftor	Overall	103 (93-113)	478 (74.0)
	<6 years	105 (97-114)	54 (80.6)
	≥6 years	103 (93-113)	424 (73.2)
Sweat chloride 6-8 weeks after ivacaftor	Overall	47 (32-60)	433 (67.0)
	<6 years	43 (29-56)	45 (67.2)
	≥6 years	48 (33-61)	388 (67.0)
FEV_1% before ivacaftor	Overall	61.6 (42.1-77.4)	458 (70.9)
	<6 years	82.6 (61.9-95.0)	19 (28.4)
	≥6 years	61.1 (41.0-77.0)	439 (75.8)
FEV_1% 6-8 weeks after ivacaftor	Overall	69.5 (49.3-86.1)	444 (68.7)
	<6 years	82.9 (70.7-95.7)	20 (29.9)
	≥6 years	68.9 (48.9-84.4)	424 (73.2)

People with CF tend to have a higher amount of chloride in their sweat than a person without cystic fibrosis. This measurement is called 'sweat chloride' and is measured in mmol/litre.

Lumacaftor/ivacaftor

Lumacaftor/ivacaftor is licensed for use in patients aged 2 and over with two copies of the F508del mutation. Managed access agreements were agreed to make lumacaftor/ivacaftor available on the NHS in Scotland from September 2019, followed by England, Wales and Northern Ireland from November 2019.

	Age (at annual review)	N
Patients on lumacaftor/ivacaftor in the UK	Overall	404
	<6 years	<5
	≥6 years	401
Patients stopped lumacaftor/ivacaftor ever	Overall	63
	<6 years	0
	≥6 years	63

Tezacaftor/ivacaftor

Tezacaftor/ivacaftor is licenced for use in patients aged 12 and over who have two copies of the F508del mutation, or a single copy of F508del and one of 14 specified 'residual function' mutations. Managed access agreements were agreed to make tezacaftor/ivacaftor available on the NHS in Scotland from September 2019, followed by England, Wales and Northern Ireland from November 2019.

	Age (at annual review)	N
Patients on tezacaftor/ivacaftor in the UK	Overall	197
	<6 years	0
	≥6 years	197
Patients stopped tezacaftor/ivacaftor	Overall	<5
	<6 years	0
	≥6 years	<5

Elexacaftor/tezacaftor/ivacaftor

At the time of writing, elexacaftor/tezacaftor/ivacaftor does not have a marketing authorisation for use in the UK and was not routinely commissioned by the NHS at any time during 2019. Elexacaftor/tezacaftor/ivacaftor is accessible through a named patient access scheme to eligible individuals who are critically ill. **13** people with cystic fibrosis in the UK are recorded as having received elexacaftor/tezacaftor/ivacaftor in 2019.

1.34 Oxygen and non-invasive ventilation

	Overall (N=10070)	<16 years (n=3966)	≥16 years (n=6104)	<18 years (n=4362)	≥18 years (n=5708)
Non-invasive ventilation (NIV); n (%)	190 (1.9)	16 (0.4)	174 (2.9)	18 (0.4)	172 (3.0)
Long-term oxygen; n (%)	615 (6.1)	72 (1.8)	543 (8.9)	88 (2.0)	527 (9.2)
Among those who have long-term oxygen:					
Continuously	131 (21.3)	<5 (2.8)	129 (23.8)	<5 (3.4)	128 (24.3)
Nocturnal or with exertion	191 (31.1)	16 (22.2)	175 (32.2)	24 (27.3)	167 (31.7)
As required (PRN)	70 (11.4)	8 (11.1)	62 (11.4)	9 (10.2)	61 (11.6)
With exacerbation	223 (36.3)	46 (63.9)	177 (32.6)	52 (59.1)	171 (32.4)

1.35 Physiotherapy

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

	Overall (N=10070)	<16 years (n=3966)	≥16 years (n=6104)	<18 years (n=4362)	≥18 years (n=5708)
Active cycle of breathing techniques; n (%)	1395 (13.9)	387 (9.8)	1008 (16.5)	431 (9.9)	964 (16.9)
Autogenic drainage (including assisted autogenic drainage); n (%)	1954 (19.4)	184 (4.6)	1770 (29.0)	247 (5.7)	1707 (29.9)
Postural drainage; n (%)	713 (7.1)	556 (14.0)	157 (2.6)	577 (13.2)	136 (2.4)
Any form of positive expiratory pressure (PEP); n (%)	6222 (61.8)	3075 (77.5)	3147 (51.6)	3397 (77.9)	2825 (49.5)
VEST; n (%)	192 (1.9)	98 (2.5)	94 (1.5)	108 (2.5)	84 (1.5)
Exercise; n (%)	6010 (59.7)	2596 (65.5)	3414 (55.9)	2852 (65.4)	3158 (55.3)
Other; n (%)	1848 (18.4)	955 (24.1)	893 (14.6)	998 (22.9)	850 (14.9)

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

1.36 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=10070)	<16 years (n=3966)	≥16 years (n=6104)	<18 years (n=4362)	≥18 years (n=5708)
Any supplemental feeding; n(%)	3683 (36.6)	1243 (31.3)	2440 (40.0)	1403 (32.2)	2280 (39.9)
Nasogastric tube; n(%)	98 (1.0)	16 (0.4)	82 (1.3)	19 (0.4)	79 (1.4)
Gastrostomy tube/button; n(%)	532 (5.3)	212 (5.3)	320 (5.2)	240 (5.5)	292 (5.1)
Jejunal; n(%)	-*	<5	10 (0.2)	<5	10 (0.2)
Total parenteral nutrition (TPN); n(%)	-*	<5	9 (0.1)	<5	9 (0.2)

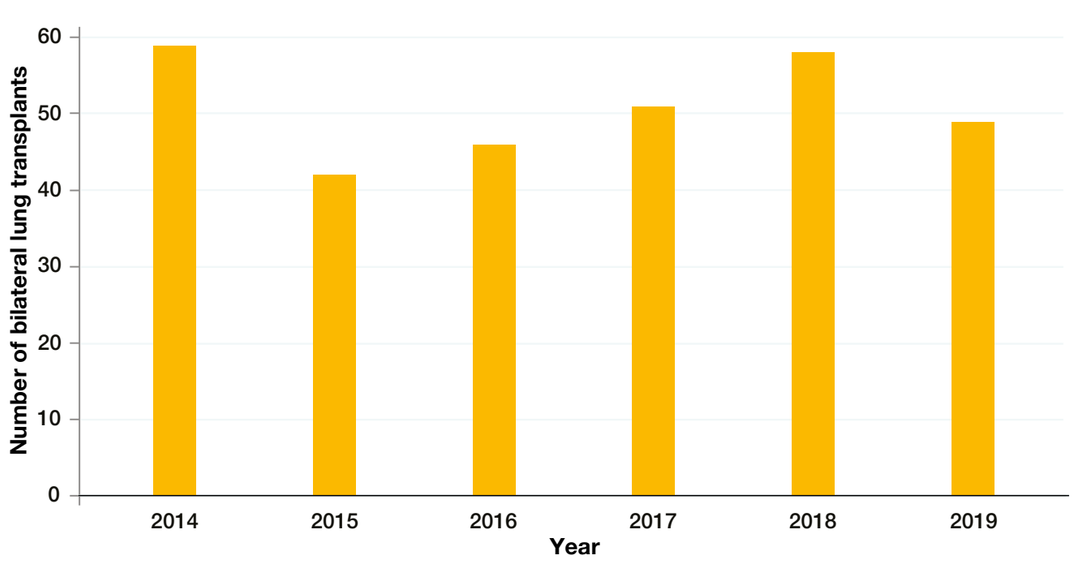
* Redacted to adhere to statistical disclosure guidelines.

1.37 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.

	2014	2015	2016	2017	2018	2019
Number evaluated	247	229	221	235	247	241
Number accepted	146	125	96	121	104	96
Number receiving aged <16 years	5	<5	<5	5	<5	<5
Bilateral lung	<5	<5	<5	<5	0	<5
Liver	<5	<5	0	0	<5	<5
Other	0	<5	0	<5	0	0
Number receiving aged ≥16 years	67	46	51	-*	63	54
Bilateral lung	59	42	46	51	58	49
Liver	5	<5	<5	0	<5	<5
Other	5	<5	<5	<5	<5	<5

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



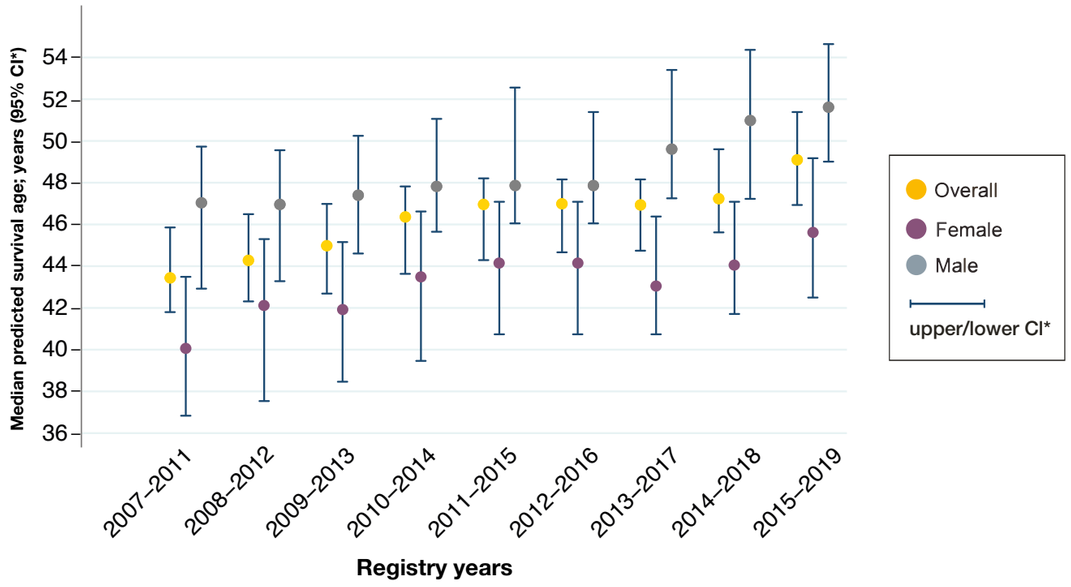
* Redacted to adhere to statistical disclosure guidelines.

Survival

1.38 Median predicted survival age

The calculation of median predicted survival 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 **49.1** years. Half are therefore predicted to die before they reach that age.

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

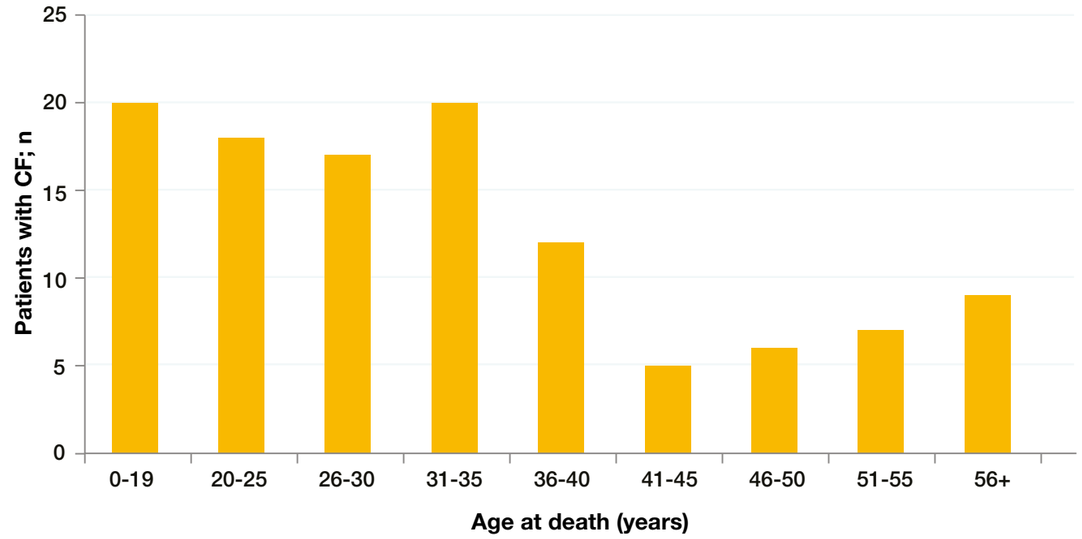


Median predicted survival age; years (95% CI*)				
Years	**Overall**	**Female**	**Male**	**p-value (males vs females)**
2007-2011	43.5 (41.9-45.9)	40.1 (36.9-43.6)	47.1 (43-49.8)	<0.001
2008-2012	44.3 (42.4-46.5)	42.2 (37.6-45.3)	47.0 (43.3-49.6)	<0.001
2009-2013	45.0 (42.8-47.0)	42.0 (38.5-45.2)	47.4 (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 (49-54.6)	<0.001

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

1.39 Age distribution of deaths in 2019

The table below shows the ages of the 137 people with CF who died in 2019. In 2019 the median age of the 114 people who died was 31. 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	20
20-25	18
26-30	17
31-35	20
36-40	12
41-45	5
46-50	6
51-55	7
56+	9
Total	**114**

1.40 Causes of death

This table shows all the recorded causes of death between 2017-2019.

Cause of death	Patients with CF; n(%)
Respiratory/cardiorespiratory	269 (70.2)
Transplant-related	36 (9.4)
Other	25 (6.5)
Cancer	19 (5.0)
Not known	17 (4.4)
Liver disease/liver failure	12 (3.1)
Trauma or suicide	5 (1.3)
Total	**383**

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	9986 (99.2)
Patients genotyped with both mutations recorded	9720 (96.5)
F508del mutations	
Homozygous F508del	4894 (48.6)
Heterozygous F508del	4141 (41.1)

1.41 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	48.6							**48.6**
R117H	4.7	0.1						**4.8**
G551D	4.2	0.2	0.2					**4.6**
G542X	2.5	0.1	0.1	0.1				**2.7**
621+1G->T	1.7	0.1	0.1	0.0	0.1			**2.1**
Other	26.1	0.6	0.9	0.7	0.5	5.0		**33.8**
Unknown	1.9	0.1	0.1	0.1	0.0	0.5	0.8	**3.5**
Total	**89.7**	**1.1**	**1.4**	**0.9**	**0.6**	**5.5**	**0.8**	**100**

1.42 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	9035	89.7
c.350G->A	p.Arg117His	R117H	587	5.8
c.1652G->A	p.Gly551Asp	G551D	581	5.8
c.1624G->T	p.Gly542X	G542X	356	3.5
c.489+1G->T		621+1G->T	258	2.6
c.3909C->G	p.Asn1303Lys	N1303K	159	1.6
c.1585-1G->A		1717-1G->A	157	1.6
c.1766+1G->A		1898+1G->A	140	1.4
c.200C->T	p.Pro67Leu	P67L	132	1.3
c.3454G->C	p.Asp1152His	D1152H	131	1.3
c.3528delC	p.Lys1177SerfsX15	3659delC	101	1.0
c.3140-26A->G		3272-26A->G	99	1.0
c.1679G->C	p.Arg560Thr	R560T	93	0.9
c.1477C->T	p.Gln493X	Q493X	87	0.9
c.1657C->T	p.Arg553X	R553X	86	0.9
c.1519_1521delATC	p.Ile507del	I507del	79	0.8
c.254G->A	p.Gly85Glu	G85E	78	0.8
c.3717+12191C->T		3849+10kbC->T	77	0.8
c.2657+5G->A		2789+5G->A	68	0.7
c.178G->T	p.Glu60X	E60X	68	0.7

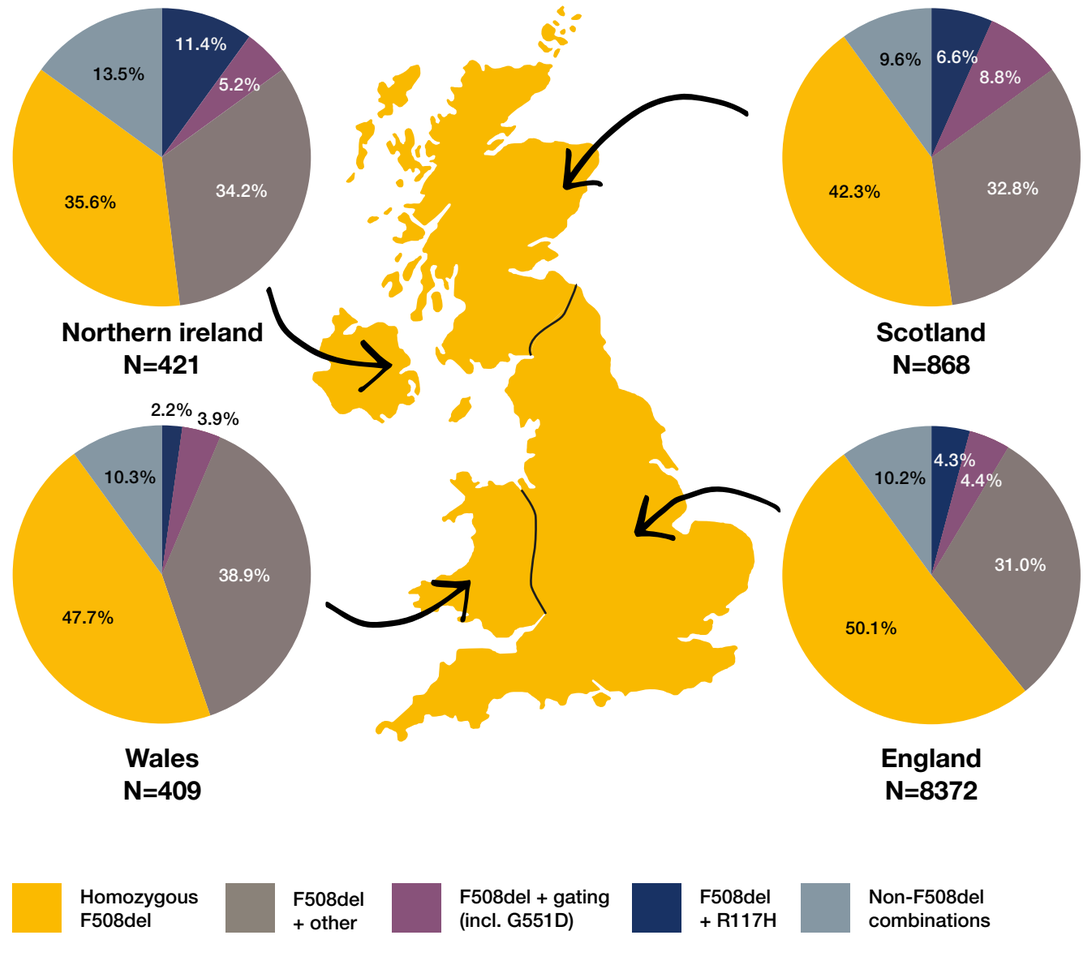
1.43 Mutation prevalence by devolved nation

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.

	England N=8372		Scotland N=868		Wales N=409		Northern Ireland N=421	
	n	%	n	%	n	%	n	%
F508del	7519	89.8	785	90.4	367	89.7	364	86.5
G551D	441	5.3	69	7.9	15	3.7	62	14.7
R117H	433	5.2	94	10.8	16	3.9	38	9
G542X	256	3.1	53	6.1	23	5.6	24	5.7
621+1G->T	191	2.3	10	1.2	45	11	12	2.9
N1303K	133	1.6	13	1.5	6	1.5	7	1.7
1717-1G->A	140	1.7	16	1.8	<5	-	0	0
1898+1G->A	110	1.3	5	0.6	25	6.1	0	0
P67L	67	0.8	46	5.3	<5	-	18	4.3
D1152H	104	1.2	15	1.7	<5	0.7	9	2.1

1.44 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 and certain patient characteristics such as their gender, as well as facilities, care pathways, and the medical team providing care.

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

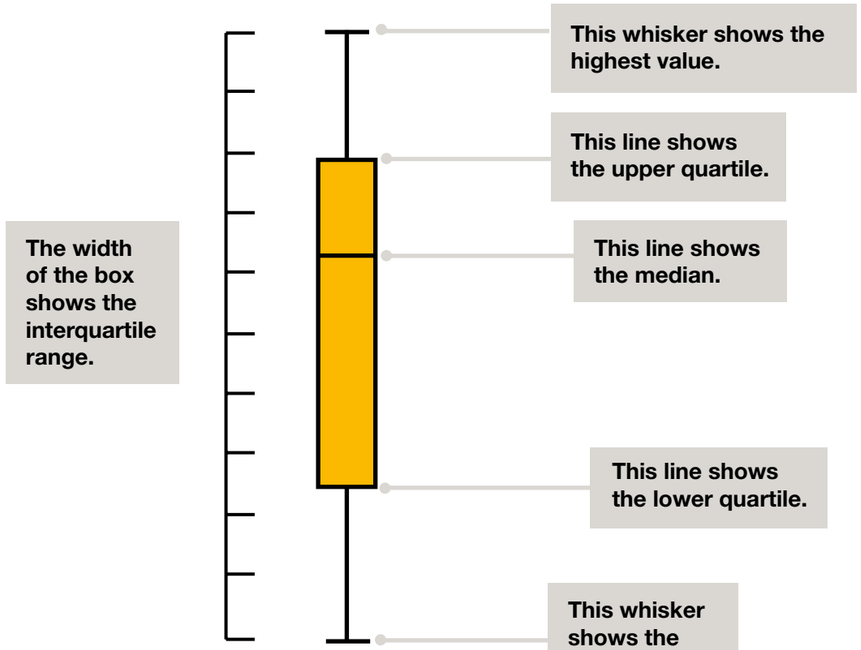
Full tables of the data are shown in appendix 2 on page 68.

Key

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.

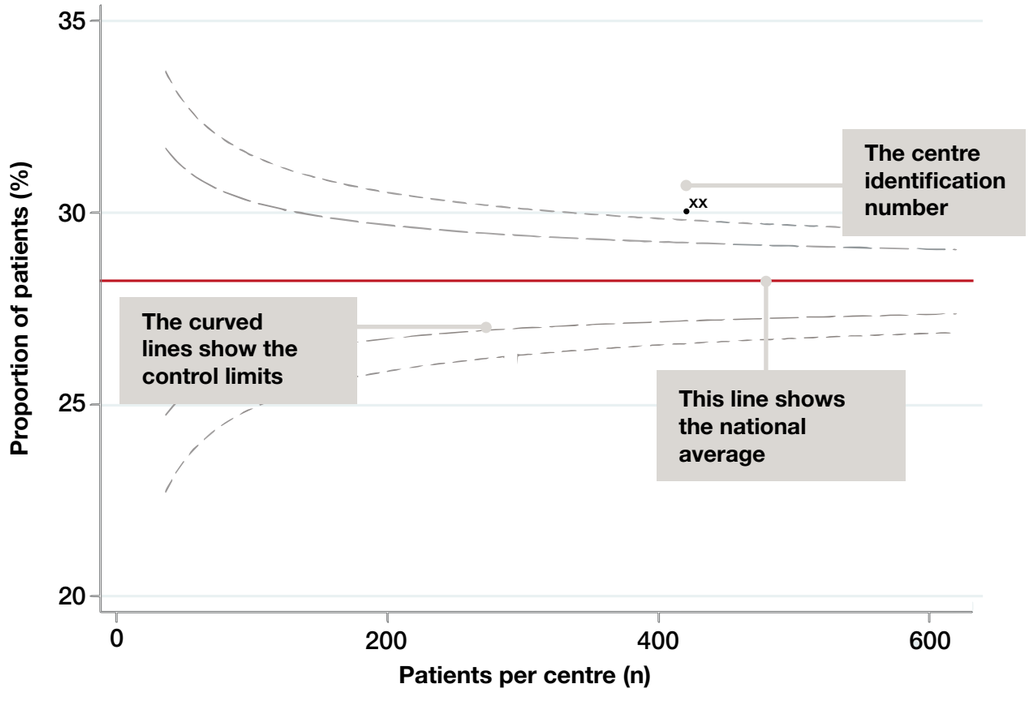
There will always be some natural variation between centres because of differences between the populations receiving care. Using only the national average as a standard can make it difficult to tell whether a survival rate that sits above the national average is higher than we would expect it to be,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



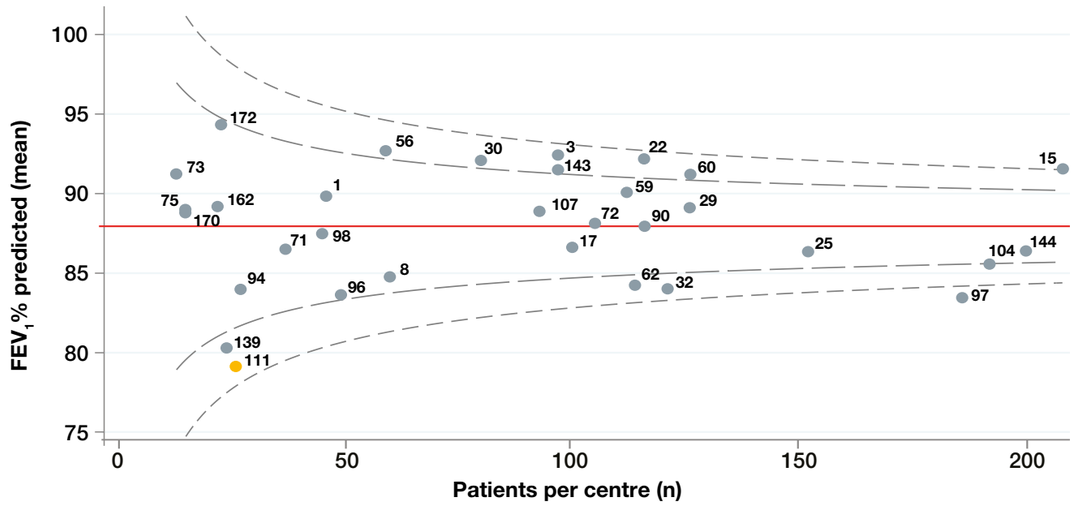
Section 2 Paediatric centre analysis

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.

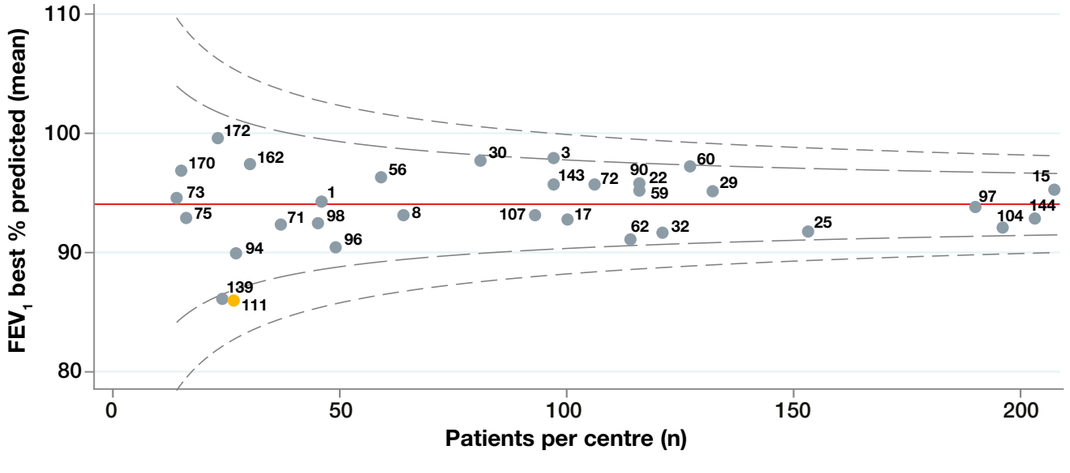
Key
Centres with their network clinics · Stand-alone clinics · — — — 2 standard deviations · - - - 3 standard deviations

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



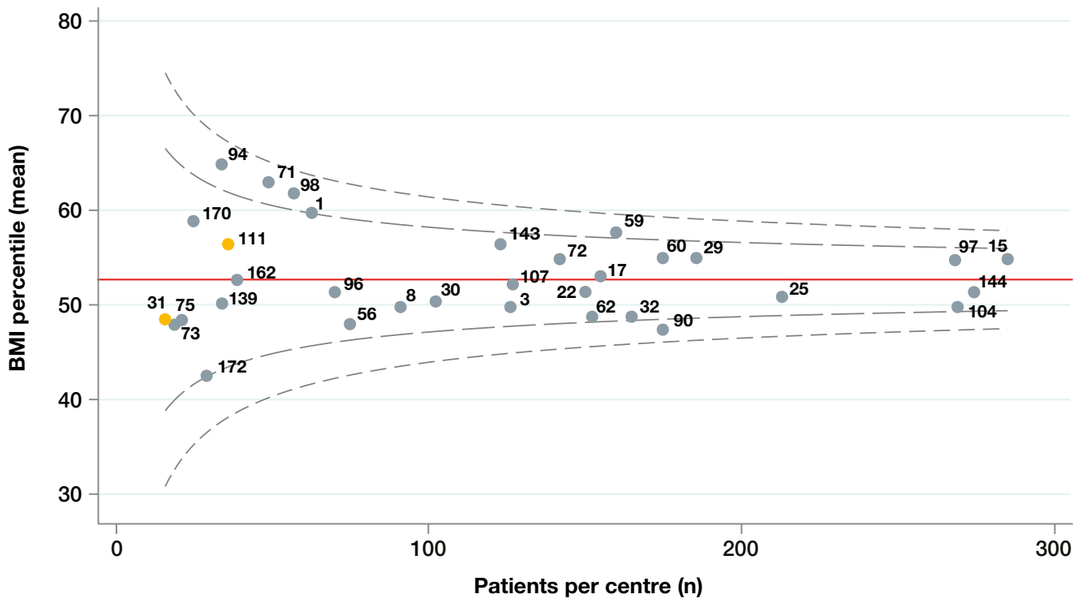
The mean FEV_1% predicted for patients attending paediatric centres/clinics is 87.9% predicted.

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



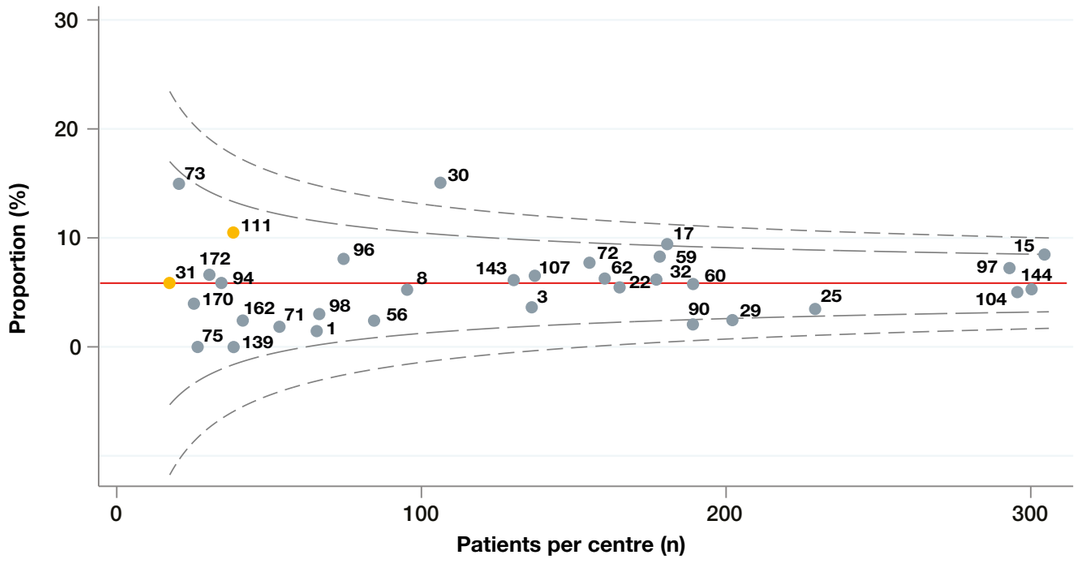
The mean Best FEV_1% predicted for patients attending paediatric centres/clinics is 94.1% predicted. Where Best FEV_1% predicted was missing, the FEV_1% predicted at annual review was used.

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



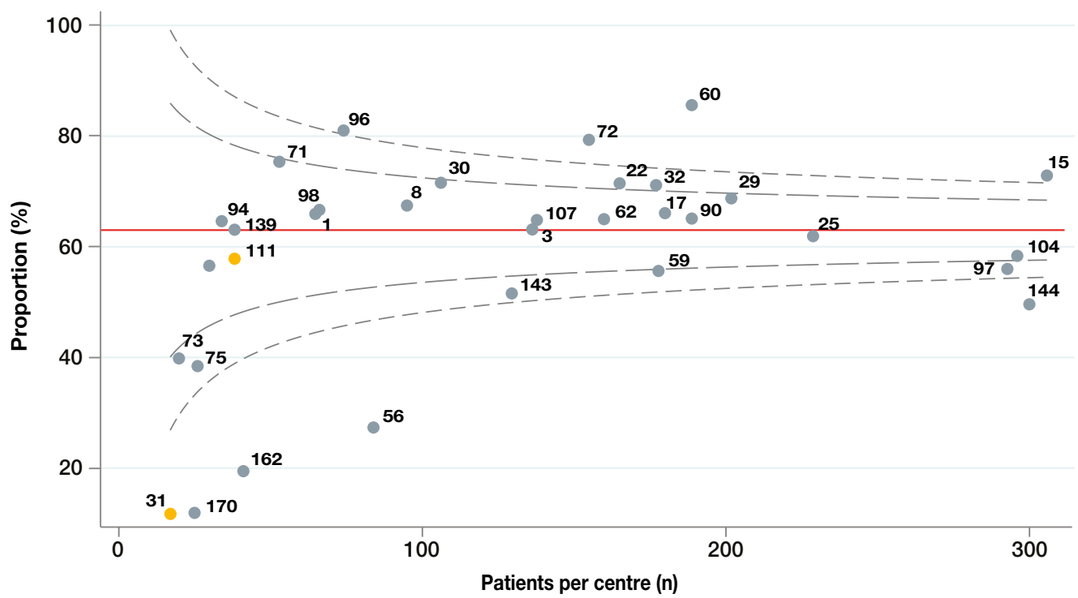
The mean BMI percentile for patients attending paediatric centres/clinics is 52.7.

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



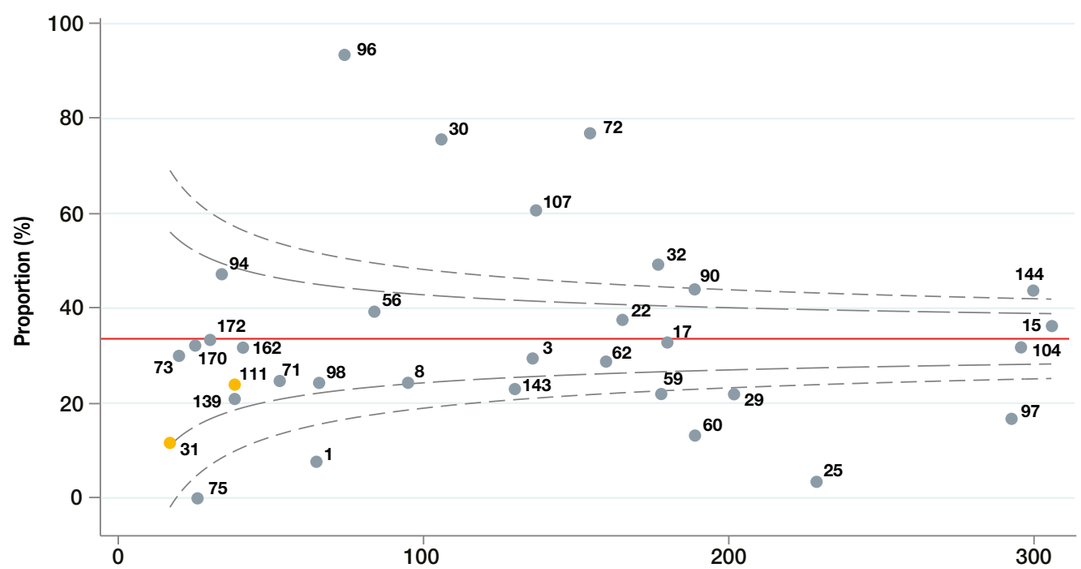
The proportion of patients with chronic *Pseudomonas aeruginosa* in paediatric centres/clinics is 5.9%.

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



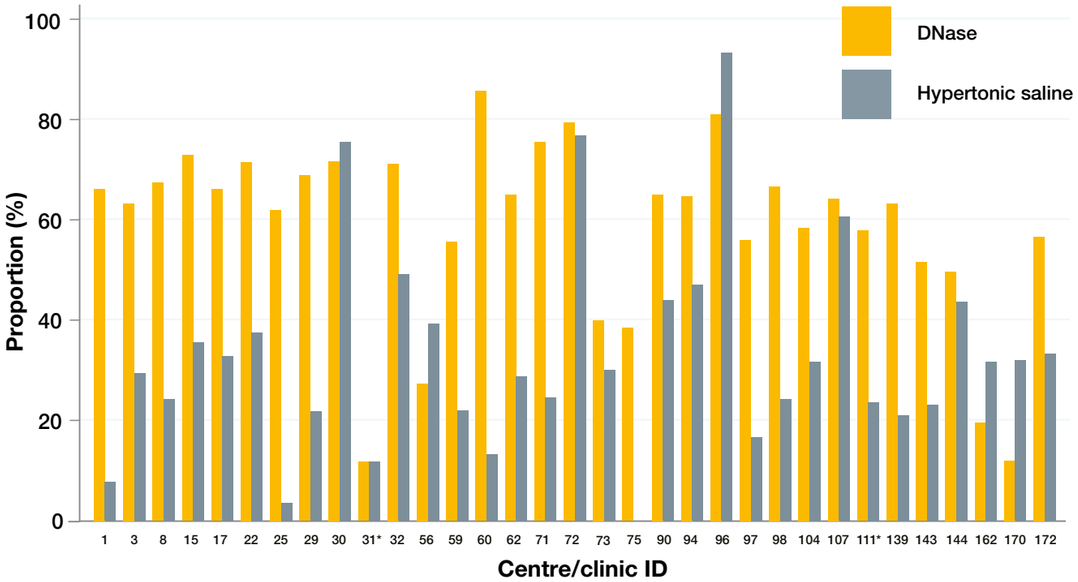
The proportion of patients receiving DNase treatment in paediatric centres/clinics is 63.1%.

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



The proportion of patients receiving hypertonic saline or mannitol treatment in paediatric centres/clinics is 33.6%.

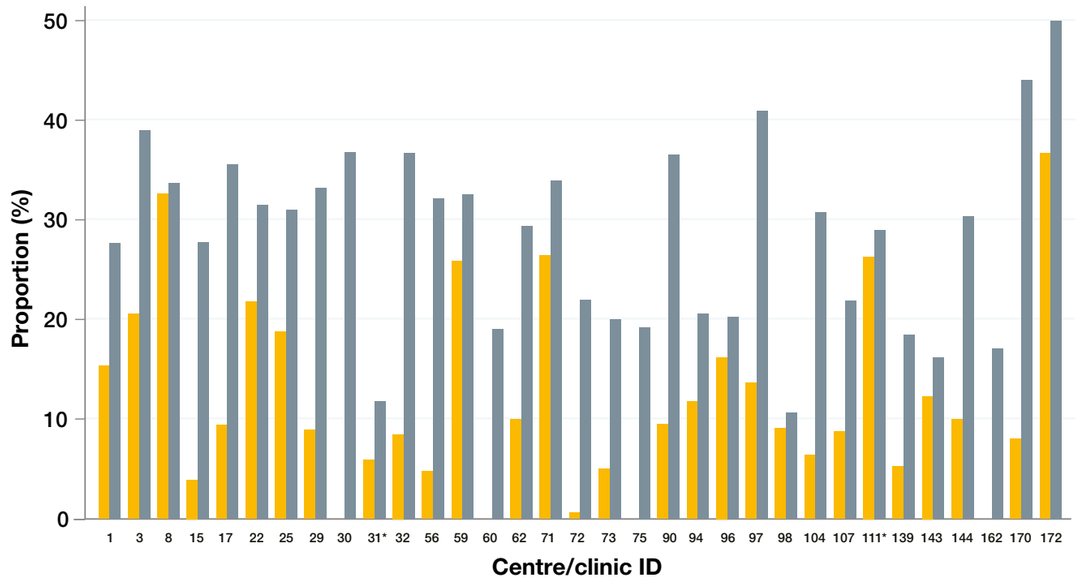
2.7 Proportion of patients receiving DNase/hypertonic saline/ mannitol treatment by paediatric centre/clinic



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 in hospital. Patients may have a combination of home and hospital IV days.

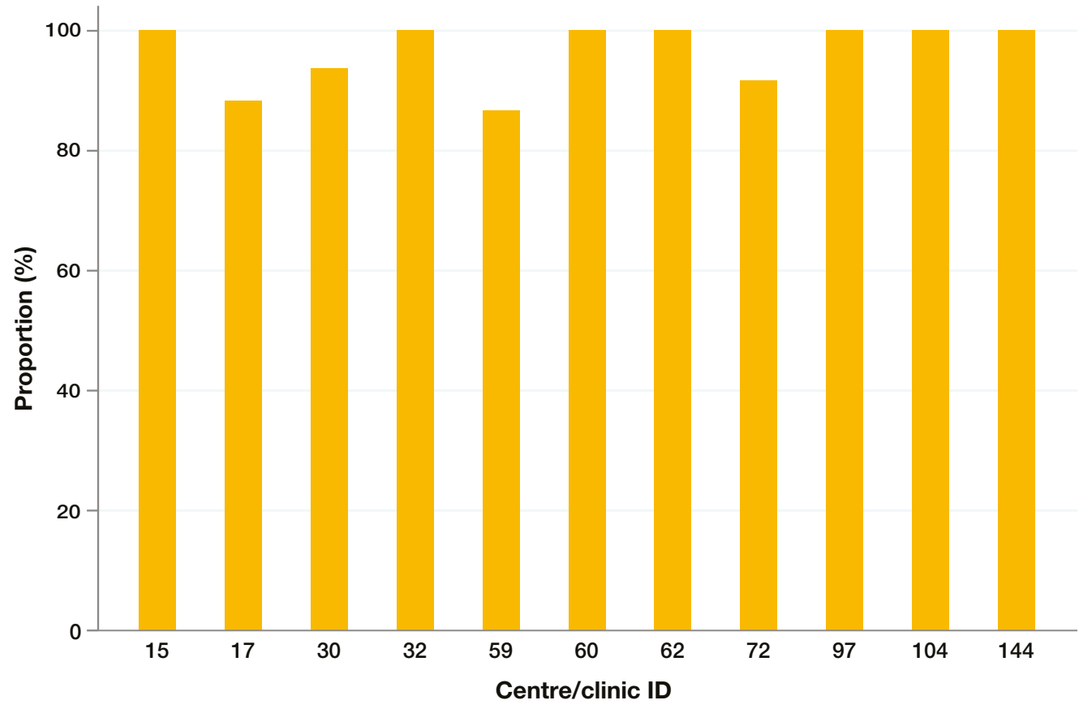


The proportion of patients receiving IVs at home was 11.2% and in hospital was 30.2%. The proportion receiving any IVs was 32.1%.

*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 front-line 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

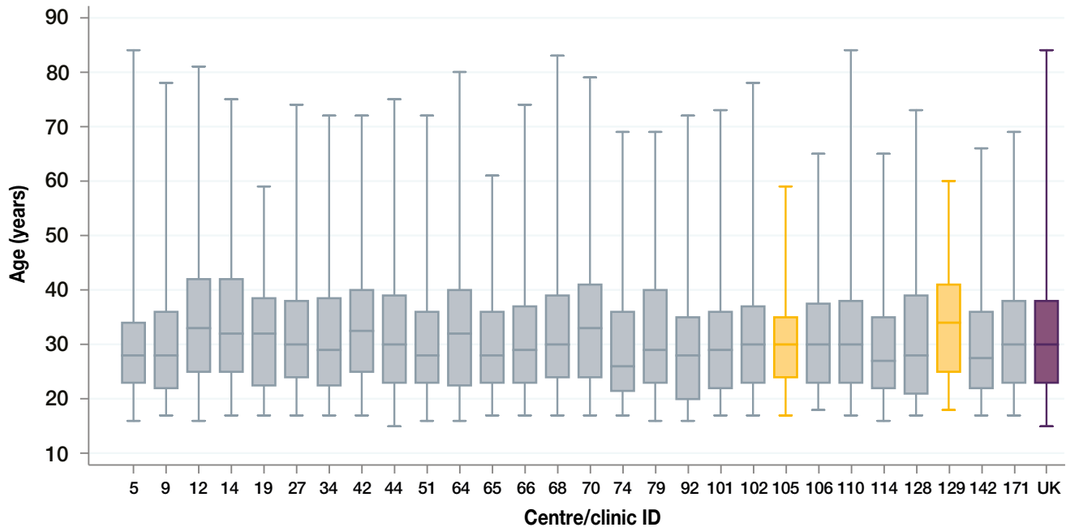
In the UK, CF care is led by 26 adult specialist CF centres. People with CF transfer to adult care centres between the ages of 16 and 18 years.

Key

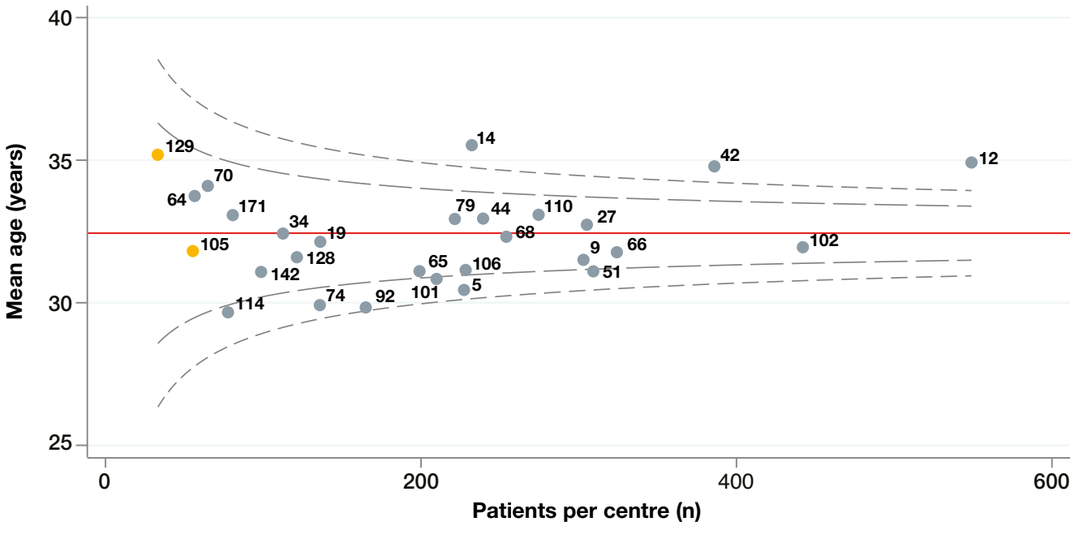
Centres with their network clinics · Stand-alone clinics · — — — 2 standard deviations · - - - 3 standard deviations

3.1 Age distribution by adults service

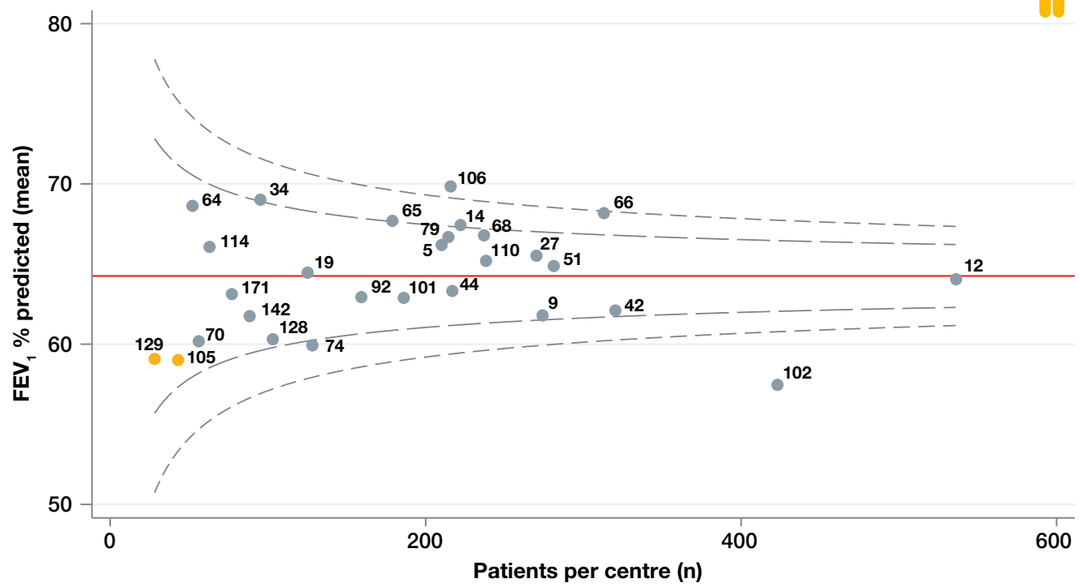
The box plot shows the age distribution of patients within each centre/clinic. In 2019 the median age in adults services was 30 years (IQR:23-38)



The funnel plot below shows how the mean age in adult centres compares to the national mean. In 2019 the national mean age of patients at CF centres was 32.5 years.

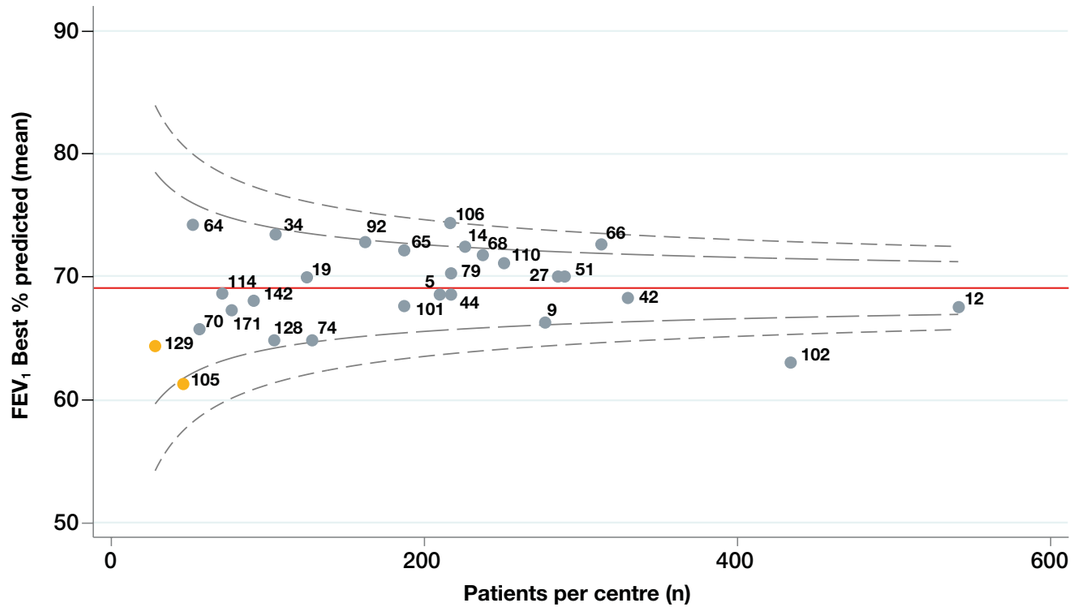


3.2 Age-adjusted FEV_1% predicted at annual review in patients without a history of lung transplant, by adult service



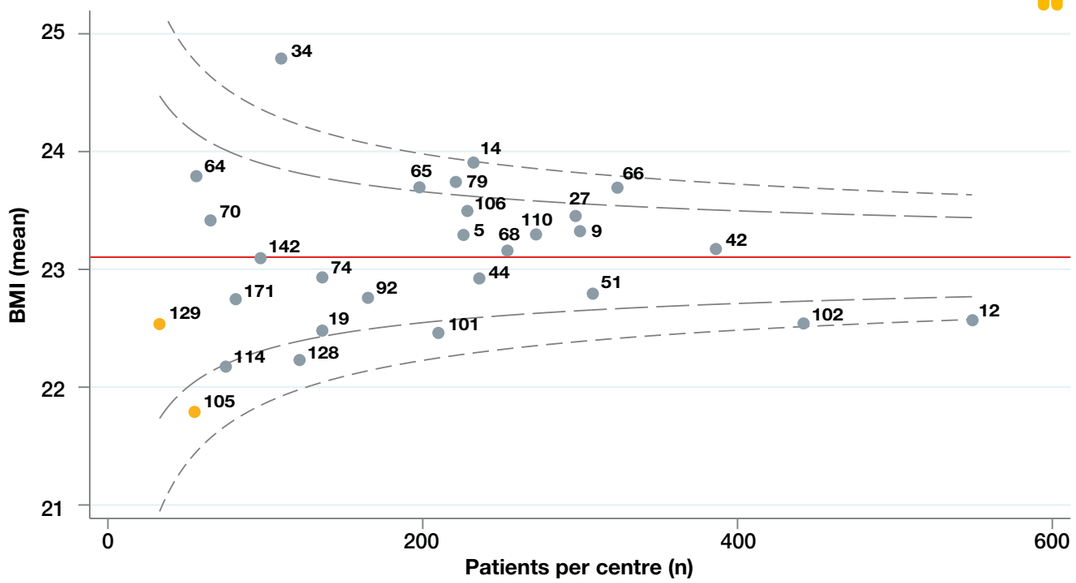
The mean FEV_1% predicted in adult services is 64.3%.

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



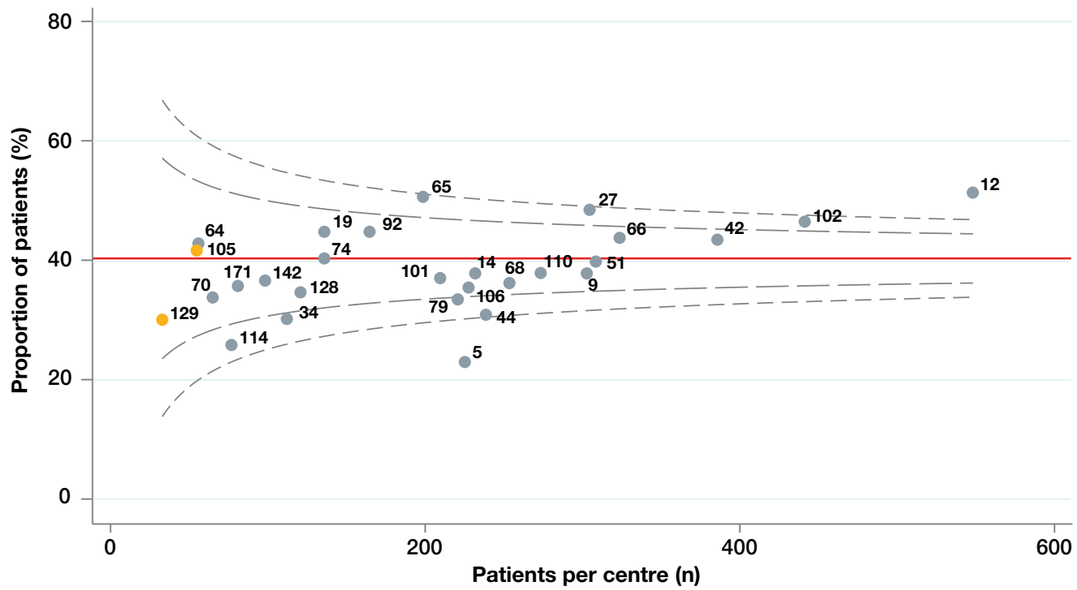
In 2019 the national mean was 69.1%. Where Best FEV_1% predicted was missing, or lower than the FEV_1 at annual review, the FEV_1% value at annual review was used.

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



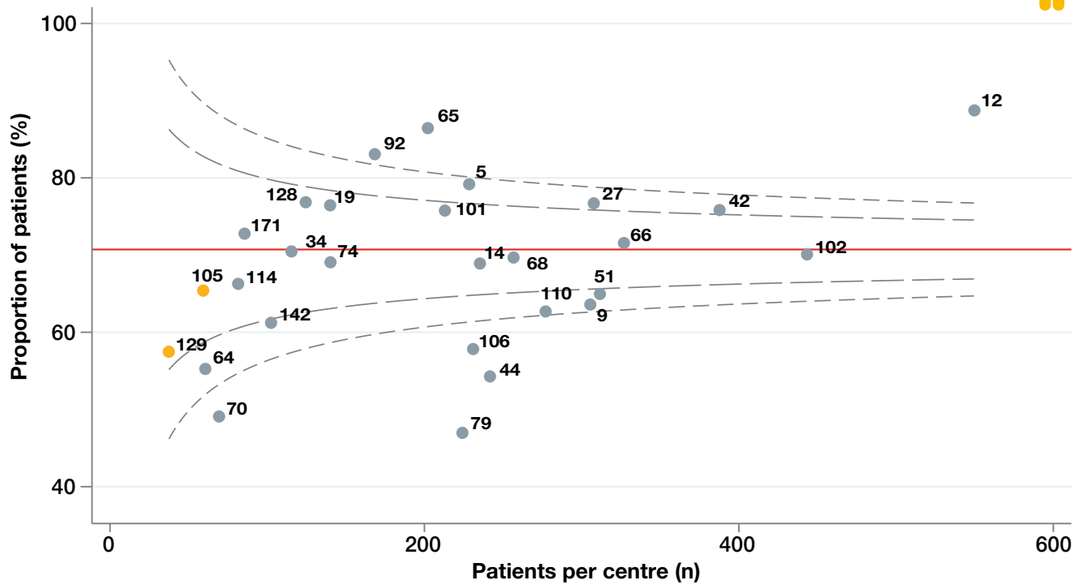
The mean BMI in adult services is 23.1.

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



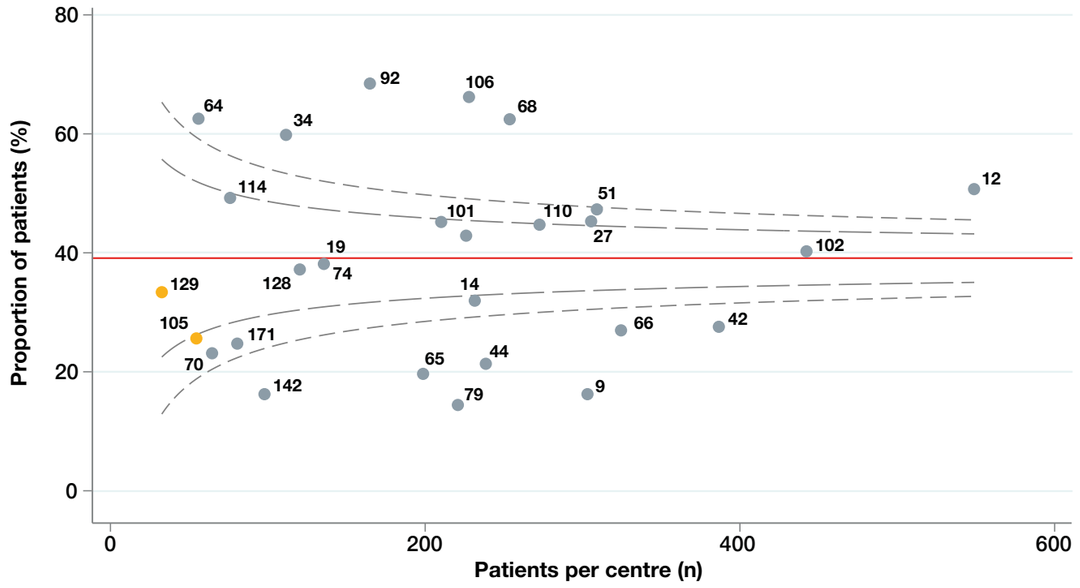
The proportion of patients with chronic *P. aeruginosa* in adult centres/clinics is 40.4%.

3.6 Proportion of patients receiving DNase treatment by adult service



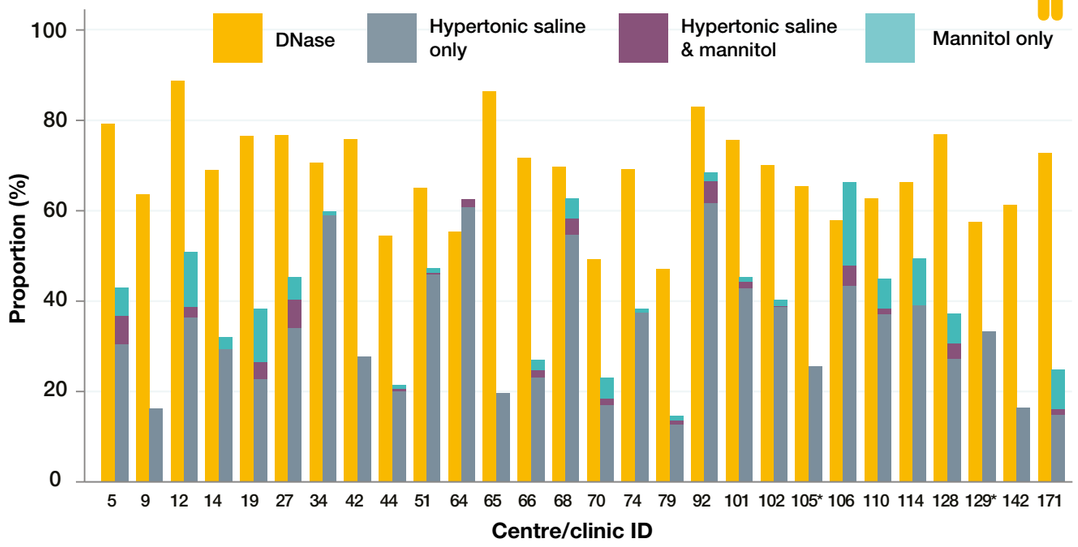
The proportion of patients receiving DNase treatment in adult centres/clinics is 70.7%.

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



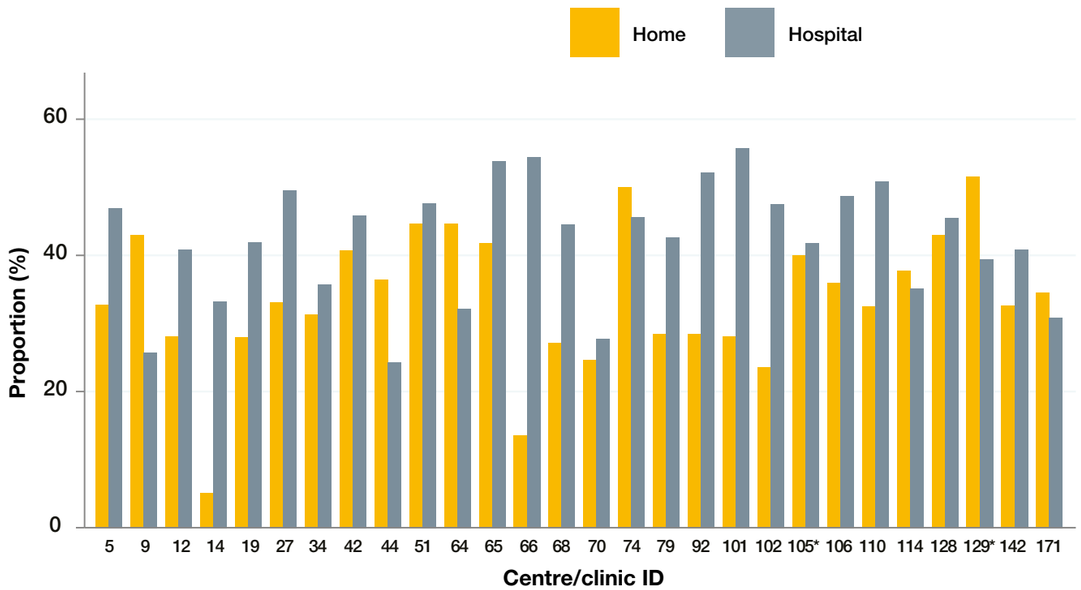
The proportion of patients receiving hypertonic saline or mannitol treatment in adult centres/clinics is 39.1%.

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



3.9 Intravenous (IV) antibiotic use by adult service

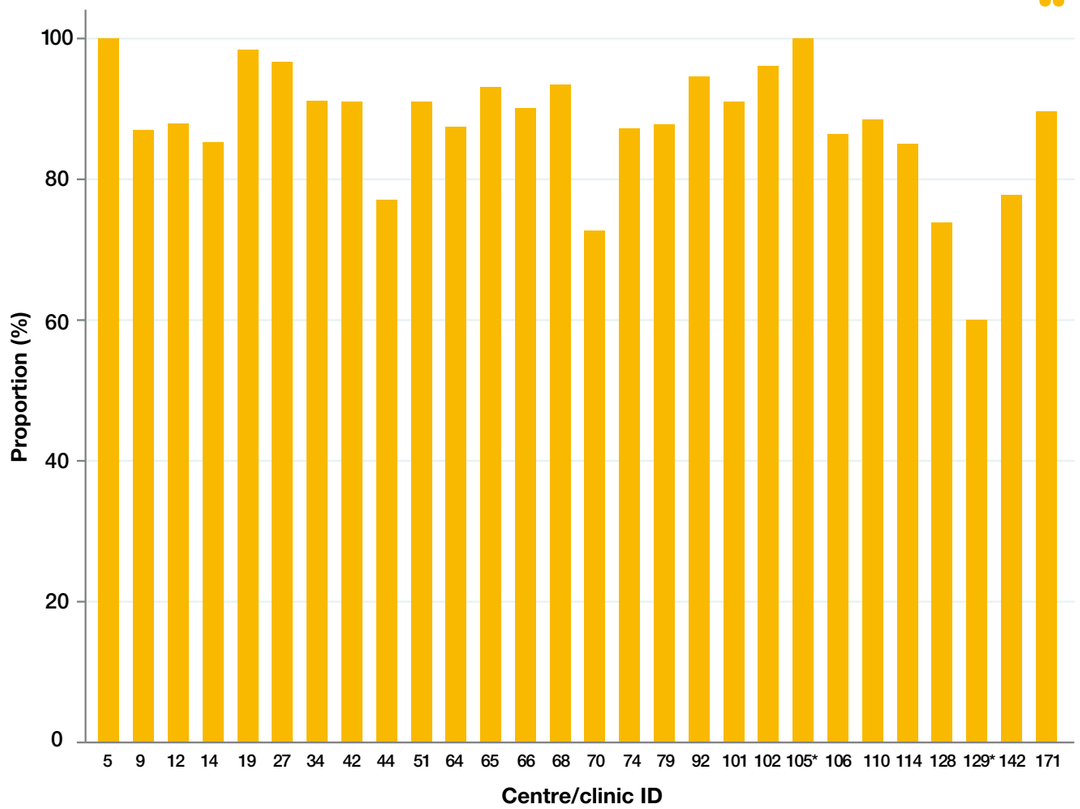
The chart below shows the proportion of patients with at least one IV day at home and in hospital. Patients may have a combination of home and hospital IV days.



The proportion of patients in adult centres receiving IV antibiotics at home was 33.6% and in hospital was 42.1%. The proportion receiving any IVs was 53.2%.

*Stand-alone clinics

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



90.0% of patients in adult centres with chronic *P. aeruginosa* received inhaled antibiotics. Centres with fewer than 10 people with chronic *Pseudomonas* were excluded.

3.11 Data completeness

Due to the COVID-19 pandemic and prioritisation of front-line 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
2019	1 January 2019 – 31 December 2019.
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)	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 2019 the median age of the 114 people who died was 31.
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 (nontuberculous 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 Data Manager †	Elaine	Gunn	Cystic Fibrosis Trust
Cystic Fibrosis Centre Data Manager	Rebecca	Heise	King's College Hospital, London
Cystic Fibrosis Cente Adminsitrator	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
Chair of the Research Committee	Martin	Wildman	Northern General Hospital, Sheffield
Registry System Development Manager †	Mary	Yip	Cystic Fibrosis Trust

*Chair † Non-voting member # Caldicott guardian

UK CF Registry Research Committee

Role	Forename	Surname	Organisation
Registry Consultant †	Noreen	Caine	Cystic Fibrosis Trust
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
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
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 2019 – ordered by clinic ID

Location	Name	Clinic ID	Total Active	Number with annual review
Leicester	Leicester Royal Infirmary	1	71	65
Sheffield	Sheffield Children's Hospital	3	141	136
North West Midlands	University Hospital of North Midlands	8	96	95
London - South West	Royal Brompton Hospital	15	321	306
London - South East	King's College Hospital	17	190	180
Oxford	John Radcliffe Hospital	22	169	165
Leeds	St James's University Hospital	25	235	229
Southampton	Southampton General Hospital	29	213	202
London - East	Royal London Hospital	30	114	106
Inverness	Raigmore Hospital	31	17	17
Bristol	Bristol Royal Hospital for Children	32	185	177
Glasgow	Royal Hospital for Children	56	102	84
Newcastle	Great North Children's Hospital	59	189	178
Belfast	Royal Belfast Hospital for Sick Children	60	196	189
Nottingham	Nottingham University Hospitals	62	161	160
Teeside	James Cook University Hospital	71	56	53
Cardiff	Children's Hospital for Wales	72	175	155
Dundee	Ninewells Hospital	73	22	20
Aberdeen	Royal Aberdeen Children's Hospital	75	33	26
London - Central	Great Ormond Street Hospital for Children	90	199	189
Cornwall	Royal Cornwall Hospital	94	36	34
Exeter	Royal Devon & Exeter Hospital	96	77	74
Liverpool	Alder Hey Children's Hospital	97	310	293
Norwich	Norfolk & Norwich University Hospital	98	72	66
Birmingham	Birmingham Children's Hospital	104	310	296
Cambridge	Addenbrookes Hospital	107	144	137
Hull	Hull University Teaching Hospitals	111	41	38
Plymouth	Derriford Hospital	139	40	38
Edinburgh	Royal Hospital for Sick Children	143	141	130
Manchester	Royal Manchester Children's Hospital	144	325	300
Lanarkshire	Wishaw General Hospital	162	43	41
Ayr	University Hospital Crosshouse	170	26	25
Brighton	Royal Alexandra Children's Hospital	172	35	30

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	9.1	46	88.7	89.8	87.4	46	93.4	94.3	93.4
8.7	8.5	97	92.4	92.4	93.7	97	98.1	98.0	98.4
8.9	9.3	60	83.9	84.7	84.7	64	92.4	93.1	95.5
8.6	8.1	208	91.3	91.6	96.3	208	95.2	95.4	99.8
7.7	7.8	100	86.1	86.6	91.0	100	92.3	92.7	94.7
8.9	9.0	116	91.8	92.2	96.0	116	95.3	95.7	96.4
8.6	8.4	152	86.2	86.3	89.7	153	91.6	91.8	95.4
8.8	8.6	126	88.2	89.1	89.9	132	94.2	95.1	94.0
10.0	10.1	80	90.5	92.1	91.8	81	96.2	97.7	98.0
8.7	10.0	9	90.7	91.7	93.6	9	94.5	95.8	93.6
9.0	8.9	121	83.0	84.0	84.3	121	90.7	91.5	92.7
8.7	9.5	59	92.6	92.7	93.3	59	96.2	96.3	97.3
8.2	7.9	112	90.1	90.1	91.3	116	95.6	95.3	94.2
8.8	9.0	126	90.4	91.1	91.6	127	96.5	97.3	96.6
9.5	9.9	114	83.3	84.2	85.3	114	90.1	91.1	91.1
10.0	9.9	37	84.0	86.5	88.0	37	89.9	92.3	91.7
9.4	9.8	105	86.6	88.1	89.7	106	94.1	95.7	94.5
8.1	8.5	13	92.6	91.2	92.9	14	96.1	94.6	99.7
7.6	7.5	15	89.4	89.0	92.0	16	93.0	92.9	92.8
8.3	7.9	116	88.1	88.0	90.6	116	96.0	95.8	96.5
9.7	8.7	27	83.7	84.0	85.8	27	89.8	89.9	94.3
8.9	8.8	49	82.5	83.6	85.1	49	89.5	90.4	89.4
8.7	8.3	186	82.9	83.5	85.4	190	93.4	93.8	92.9
9.1	10.5	45	86.0	87.5	87.0	45	90.9	92.5	90.4
8.8	8.6	192	84.5	85.5	87.9	196	91.1	92.1	92.5
8.5	8.9	93	89.2	88.9	91.0	93	93.5	93.1	95.4
8.3	7.6	26	80.2	79.2	79.0	26	87.1	86.1	88.3
8.1	8.0	24	80.5	80.3	88.6	24	86.4	86.1	91.9
9.4	9.2	97	91.1	91.5	91.5	97	95.3	95.7	95.0
9.1	8.8	200	85.5	86.4	87.1	203	92.0	92.8	94.2
9.0	9.2	22	88.4	89.2	91.5	30	97.4	97.4	98.1
8.7	8.2	15	87.9	88.8	87.2	15	95.8	96.9	93.5
9.4	8.8	23	93.6	94.3	96.2	23	99.3	99.6	100.0

* 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	63	59.8	59.8	64.2
Sheffield	Sheffield Children's Hospital	3	126	50.0	49.9	53.1
North West Midlands	University Hospital of North Midlands	8	91	50.0	49.9	51.3
London - South West	Royal Brompton Hospital	15	285	55.1	54.9	55.1
London - South East	King's College Hospital	17	155	53.4	53.1	54.9
Oxford	John Radcliffe Hospital	22	150	51.3	51.4	49.5
Leeds	St James's University Hospital	25	213	51.0	50.9	49.8
Southampton	Southampton General Hospital	29	186	55.1	55.1	56.1
London - East	Royal London Hospital	30	102	50.0	50.5	52.7
Inverness	Raigmore Hospital	31	16	48.8	48.6	43.6
Bristol	Bristol Royal Hospital for Children	32	165	48.8	48.8	47.4
Glasgow	Royal Hospital for Children	56	75	48.0	48.1	44.1
Newcastle	Great North Children's Hospital	59	160	58.0	57.7	62.1
Belfast	Royal Belfast Hospital for Sick Children	60	175	55.2	55.2	54.9
Nottingham	Nottingham University Hospitals	62	152	48.7	48.9	49.2
Teeside	James Cook University Hospital	71	49	62.3	63.0	71.3
Cardiff	Children's Hospital for Wales	72	142	54.5	54.9	57.3
Dundee	Ninewells Hospital	73	19	48.6	48.1	43.8
Aberdeen	Royal Aberdeen Children's Hospital	75	21	48.7	48.5	45.7
London - Central	Great Ormond Street Hospital for Children	90	175	47.8	47.5	44.6
Cornwall	Royal Cornwall Hospital	94	34	64.7	64.9	59.5
Exeter	Royal Devon & Exeter Hospital	96	70	51.5	51.5	55.3
Liverpool	Alder Hey Children's Hospital	97	268	54.9	54.8	53.7
Norwich	Norfolk & Norwich University Hospital	98	57	61.3	61.8	69.3
Birmingham	Birmingham Children's Hospital	104	269	49.9	49.9	50.8
Cambridge	Addenbrookes Hospital	107	127	52.4	52.2	54.7
Hull	Hull University Teaching Hospitals	111	36	56.9	56.5	54.9
Plymouth	Derriford Hospital	139	34	50.6	50.2	51.6
Edinburgh	Royal Hospital for Sick Children	143	123	56.2	56.5	56.0
Manchester	Royal Manchester Children's Hospital	144	274	51.3	51.5	51.6
Lanarkshire	Wishaw General Hospital	162	39	52.7	52.7	53.2
Ayr	University Hospital Crosshouse	170	25	59.2	58.9	59.4
Brighton	Royal Alexandra Children's Hospital	172	29	42.4	42.6	37.5

Chronic Pseudomonas		Having at least 1 IV day		Receiving DNase treatment		Receiving hypertonic saline/mannitol treatment		Inhaled antibiotic use among patients with chronic Pseudomonas	
Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)
<5	1.5	20	30.8	43	66.2	5	7.7	<5	100.0
5	3.7	53	39.0	86	63.2	40	29.4	5	100.0
5	5.3	46	48.4	64	67.4	23	24.2	<5	80.0
26	8.5	85	27.8	223	72.9	111	36.3	26	100
17	9.4	64	35.6	119	66.1	59	32.8	15	88.2
9	5.5	61	37.0	118	71.5	62	37.6	9	100.0
8	3.5	85	37.1	142	62.0	8	3.5	7	87.5
5	2.5	68	33.7	139	68.8	44	21.8	5	100.0
16	15.1	39	36.8	76	71.7	80	75.5	15	93.8
<5	5.9	<5	11.8	<5	11.8	<5	11.8	0	0.0
11	6.2	65	36.7	126	71.2	87	49.2	11	100.0
<5	2.4	27	32.1	23	27.4	33	39.3	<5	100.0
15	8.4	71	39.9	99	55.6	39	21.9	13	86.7
11	5.8	36	19.0	162	85.7	25	13.2	11	100.0
10	6.3	48	30.0	104	65.0	46	28.8	10	100.0
<5	1.9	20	37.7	40	75.5	13	24.5	<5	100.0
12	7.7	34	21.9	123	79.4	119	76.8	11	91.7
<5	15.0	<5	20.0	8	40.0	6	30.0	<5	66.7
0	0.0	5	19.2	10	38.5	0	0.0	0	0.0
<5	2.1	69	36.5	123	65.1	83	43.9	<5	100.0
<5	5.9	7	20.6	22	64.7	16	47.1	<5	100.0
6	8.1	17	23.0	60	81.1	69	93.2	6	100.0
21	7.2	125	42.7	164	56.0	49	16.7	21	100.0
<5	3.0	11	16.7	44	66.7	16	24.2	<5	100.0
15	5.1	92	31.1	173	58.4	94	31.8	15	100.0
9	6.6	32	23.4	88	64.2	83	60.6	9	100.0
<5	10.5	13	34.2	22	57.9	9	23.7	<5	100.0
0	0.0	7	18.4	24	63.2	8	21.1	0	0.0
8	6.2	25	19.2	67	51.5	30	23.1	7	87.5
16	5.3	93	31.0	149	49.7	131	43.7	16	100.0
<5	2.4	7	17.1	8	19.5	13	31.7	<5	100.0
<5	4.0	12	48.0	<5	12.0	8	32.0	<5	100.0
<5	6.7	15	50.0	17	56.7	10	33.3	<5	100.0

Appendix 2: Centre-level data tables

Adult centres/clinics providing data in 2019 – ordered by clinic ID

				Age (years)		FEV$_1$% predicted at annual review				Best FEV$_1$% predicted
Location	Name	Clinic ID	Total Active	Number with annual review	Mean	Median	Number	Mean - unadjusted	Mean - adjusted	Median
London - South East	King's College Hospital	5	240	226	30.5	28.6	210	66.5	66.2	69.0
Newcastle	Royal Victoria Infirmary	9	308	303	31.5	28.9	274	62.2	61.9	61.6
London - South West	Royal Brompton Hospital	12	559	549	34.9	33.0	536	62.8	64.0	62.7
Belfast	Belfast City Hospital	14	291	232	35.5	32.1	222	66.7	67.4	69.5
Frimley	Frimley Park Hospital	19	142	136	32.2	32.0	125	64.0	64.5	67.1
Birmingham	Birmingham Heartlands Hospital	27	318	305	32.8	30.1	270	65.2	65.5	65.0
Exeter	Royal Devon & Exeter Hospital	34	118	112	32.4	29.1	95	68.8	69.0	69.3
Leeds	St James's University Hospital	42	398	386	34.8	33.0	320	61.0	62.1	60.9
Edinburgh	Western General Hospital	44	248	239	33.0	30.8	217	63.1	63.3	63.6
Cambridge	Royal Papworth Hospital	51	330	309	31.2	28.7	281	65.1	64.9	65.8
Plymouth	Derriford Hospital	64	59	56	33.8	32.4	52	68.1	68.6	72.9
Sheffield	Northern General Hospital	65	208	199	31.1	28.8	179	67.9	67.7	72.3
Liverpool	Liverpool Heart and Chest Hospital	66	349	324	31.8	29.9	313	68.0	68.2	70.0
Llandough	Llandough Hospital	68	272	254	32.3	30.9	237	66.5	66.8	67.7
Aberdeen	Aberdeen Royal Infirmary	70	65	65	34.1	33.0	56	59.6	60.2	52.2
North West Midlands	University Hospital of North Midlands	74	140	136	30.0	26.0	128	60.8	60.0	63.2
Glasgow	Queen Elizabeth University Hospital	79	234	221	33.0	29.3	214	66.5	66.7	66.7
London - East	St Bartholomew's Hospital	92	185	165	29.9	28.1	159	63.9	63.0	62.9
Nottingham	Nottingham University Hospitals	101	213	210	30.9	29.1	186	63.4	62.9	64.1
Manchester	Wythenshawe Hospital	102	464	442	32.0	30.0	423	57.2	57.5	55.4
London - South East	University Hospital Lewisham	105	59	55	31.8	30.1	43	58.7	59.0	57.2
Bristol	Bristol Royal Infirmary	106	234	228	31.2	30.0	216	69.7	69.8	69.3
Southampton	Southampton General Hospital	110	297	274	33.1	30.1	238	65.3	65.2	65.1
Norwich	Norfolk & Norwich University Hospital	114	77	77	29.7	27.2	63	66.7	66.1	66.8
Oxford	John Radcliffe Hospital	128	137	121	31.6	28.3	103	61.1	60.3	57.6
Cornwall	Royal Cornwall Hospital	129	36	33	35.2	34.9	28	57.5	59.1	55.4
Leicester	Glenfield Hospital	142	103	98	31.1	28.0	88	62.3	61.8	64.9
York & Hull	York Hospital	171	82	81	33.1	30.0	77	62.8	63.1	61.5

Best FEV$_1$% predicted			
Number*	Mean - unadjusted	Mean - adjusted	Median
210	69.2	68.6	70.5
277	66.9	66.2	69.6
541	66.3	67.5	66.6
226	71.8	72.4	74.2
125	69.6	69.9	71.5
286	69.7	70.0	70.7
105	73.4	73.5	73.8
330	67.2	68.3	67.1
217	68.6	68.6	69.8
289	70.4	70.0	72.4
52	73.7	74.2	76.2
187	72.5	72.2	76.6
313	72.7	72.7	74.5
237	71.7	71.8	72.8
56	65.2	65.8	58.1
128	66.1	64.9	68.9
217	70.1	70.2	72.4
162	74.1	72.8	68.4
187	68.4	67.7	69.9
434	62.8	63.0	62.9
46	61.0	61.3	60.1
216	74.4	74.4	75.6
251	71.4	71.1	74.6
71	69.5	68.7	70.6
104	65.9	64.8	65.4
28	62.6	64.3	60.3
91	68.9	68.1	71.8
77	67.1	67.3	68.9

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

			BMI			
Location	Name	Clinic ID	Number	Mean - unadjusted	Mean - adjusted	Median
London - South East	King's College Hospital	5	226	23.1	23.3	22.7
Newcastle	Royal Victoria Infirmary	9	300	23.2	23.3	22.5
London - South West	Royal Brompton Hospital	12	549	22.8	22.6	22.4
Belfast	Belfast City Hospital	14	232	24.2	23.9	23.5
Frimley	Frimley Park Hospital	19	136	22.5	22.5	22.1
Birmingham	Birmingham Heartlands Hospital	27	297	23.5	23.5	23.0
Exeter	Royal Devon & Exeter Hospital	34	110	24.8	24.8	24.8
Leeds	St James's University Hospital	42	386	23.4	23.2	22.8
Edinburgh	Western General Hospital	44	236	23.0	22.9	22.5
Cambridge	Royal Papworth Hospital	51	308	22.7	22.8	22.3
Plymouth	Derriford Hospital	64	56	23.9	23.8	23.5
Sheffield	Northern General Hospital	65	198	23.6	23.7	22.9
Liverpool	Liverpool Heart and Chest Hospital	66	324	23.6	23.7	22.8
Llandough	Llandough Hospital	68	254	23.2	23.2	22.4
Aberdeen	Aberdeen Royal Infirmary	70	65	23.6	23.4	22.6
North West Midlands	University Hospital of North Midlands	74	136	22.7	22.9	22.2
Glasgow	Queen Elizabeth University Hospital	79	221	23.8	23.8	23.2
London - East	St Bartholomew's Hospital	92	165	22.5	22.8	21.8
Nottingham	Nottingham University Hospitals	101	210	22.3	22.5	21.8
Manchester	Wythenshawe Hospital	102	442	22.5	22.5	22.1
London - South East	University Hospital Lewisham	105	55	21.8	21.8	21.1
Bristol	Bristol Royal Infirmary	106	228	23.4	23.5	22.7
Southampton	Southampton General Hospital	110	272	23.3	23.3	22.5
Norwich	Norfolk & Norwich University Hospital	114	75	21.9	22.2	21.1
Oxford	John Radcliffe Hospital	128	121	22.1	22.2	21.9
Cornwall	Royal Cornwall Hospital	129	33	22.8	22.5	21.3
Leicester	Glenfield Hospital	142	97	22.9	23.1	21.9
York & Hull	York Hospital	171	81	22.8	22.8	22.0

Chronic Pseudomonas		Having at least 1 IV day		Receiving DNase treatment		Receiving hypertonic saline/mannitol treatment		Inhaled antibiotic use among patients with chronic Pseudomonas	
Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)
52	23.0	116	51.3	179	79.2	97	42.9	52	100.0
115	38.0	146	48.2	193	63.7	49	16.2	100	87.0
282	51.4	276	50.3	487	88.7	279	50.8	248	87.9
88	37.9	77	33.2	160	69.0	74	31.9	75	85.2
61	44.9	69	50.7	104	76.5	52	38.2	60	98.4
148	48.5	179	58.7	234	76.7	138	45.2	143	96.6
34	30.4	50	44.6	79	70.5	67	59.8	31	91.2
168	43.5	222	57.5	293	75.9	107	27.7	153	91.1
74	31.0	99	41.4	130	54.4	51	21.3	57	77.0
123	39.8	186	60.2	201	65.0	146	47.2	112	91.1
24	42.9	29	51.8	31	55.4	35	62.5	21	87.5
101	50.8	131	65.8	172	86.4	39	19.6	94	93.1
142	43.8	187	57.7	232	71.6	87	26.9	128	90.1
92	36.2	130	51.2	177	69.7	159	62.6	86	93.5
22	33.8	28	43.1	32	49.2	15	23.1	16	72.7
55	40.4	84	61.8	94	69.1	52	38.2	48	87.3
74	33.5	96	43.4	104	47.1	32	14.5	65	87.8
74	44.8	94	57.0	137	83.0	113	68.5	70	94.6
78	37.1	133	63.3	159	75.7	95	45.2	71	91.0
206	46.6	242	54.8	310	70.1	178	40.3	198	96.1
23	41.8	32	58.2	36	65.5	14	25.5	23	100.0
81	35.5	140	61.4	132	57.9	151	66.2	70	86.4
104	38.0	154	56.2	172	62.8	123	44.9	92	88.5
20	26.0	39	50.6	51	66.2	38	49.4	17	85.0
42	34.7	69	57.0	93	76.9	45	37.2	31	73.8
10	30.3	19	57.6	19	57.6	11	33.3	6	60.0
36	36.7	51	52.0	60	61.2	16	16.3	28	77.8
29	35.8	41	50.6	59	72.8	20	24.7	26	89.7

Appendix 2: Centre-level data tables

Paediatric centres/clinics providing data in 2019 – ordered alphabetically by country/city

Location	Name	Clinic ID	Total Active	Number with annual review	Age (years) Mean	Age (years) Median	FEV_1% predicted at annual review Number	FEV_1% predicted at annual review Mean - unadjusted	FEV_1% predicted at annual review Mean - adjusted	FEV_1% predicted at annual review Median	Best FEV_1% predicted Number*	Best FEV_1% predicted Mean - unadjusted	Best FEV_1% predicted Mean - adjusted	Best FEV_1% predicted Median
England														
Birmingham	Birmingham Children's Hospital	104	310	296	8.8	8.6	192	84.5	85.5	87.9	196	91.1	92.1	92.5
Brighton	Royal Alexandra Children's Hospital	172	35	30	9.4	8.8	23	93.6	94.3	96.2	23	99.3	99.6	100.0
Bristol	Bristol Royal Hospital for Children	32	185	177	9.0	8.9	121	83.0	84.0	84.3	121	90.7	91.5	92.7
Cambridge	Addenbrookes Hospital	107	144	137	8.5	8.9	93	89.2	88.9	91.0	93	93.5	93.1	95.4
Cornwall	Royal Cornwall Hospital	94	36	34	9.7	8.7	27	83.7	84.0	85.8	27	89.8	89.9	94.3
Exeter	Royal Devon & Exeter Hospital	96	77	74	8.9	8.8	49	82.5	83.6	85.1	49	89.5	90.4	89.4
Hull	Hull University Teaching Hospitals	111	41	38	8.3	7.6	26	80.2	79.2	79.0	26	87.1	86.1	88.3
Leeds	St James's University Hospital	25	235	229	8.6	8.4	152	86.2	86.3	89.7	153	91.6	91.8	95.4
Leicester	Leicester Royal Infirmary	1	71	65	9.2	9.1	46	88.7	89.8	87.4	46	93.4	94.3	93.4
Liverpool	Alder Hey Children's Hospital	97	310	293	8.7	8.3	186	82.9	83.5	85.4	190	93.4	93.8	92.9
London - Central	Great Ormond Street Hospital for Children	90	199	189	8.3	7.9	116	88.1	88.0	90.6	116	96.0	95.8	96.5
London - East	Royal London Hospital	30	114	106	10.0	10.1	80	90.5	92.1	91.8	81	96.2	97.7	98.0
London - South East	King's College Hospital	17	190	180	7.7	7.8	100	86.1	86.6	91.0	100	92.3	92.7	94.7
London - South West	Royal Brompton Hospital	15	321	306	8.6	8.1	208	91.3	91.6	96.3	208	95.2	95.4	99.8
Manchester	Royal Manchester Children's Hospital	144	325	300	9.1	8.8	200	85.5	86.4	87.1	203	92.0	92.8	94.2
Newcastle	Great North Children's Hospital	59	189	178	8.2	7.9	112	90.1	90.1	91.3	116	95.6	95.3	94.2
North West Midlands	University Hospital of North Midlands	8	96	95	8.9	9.3	60	83.9	84.7	84.7	64	92.4	93.1	95.5
Norwich	Norfolk & Norwich University Hospital	98	72	66	9.1	10.5	45	86.0	87.5	87.0	45	90.9	92.5	90.4
Nottingham	Nottingham University Hospitals	62	161	160	9.5	9.9	114	83.3	84.2	85.3	114	90.1	91.1	91.1
Oxford	John Radcliffe Hospital	22	169	165	8.9	9.0	116	91.8	92.2	96.0	116	95.3	95.7	96.4
Plymouth	Derriford Hospital	139	40	38	8.1	8.0	24	80.5	80.3	88.6	24	86.4	86.1	91.9
Sheffield	Sheffield Children's Hospital	3	141	136	8.7	8.5	97	92.4	92.4	93.7	97	98.1	98.0	98.4
Southampton	Southampton General Hospital	29	213	202	8.8	8.6	126	88.2	89.1	89.9	132	94.2	95.1	94.0
Teeside	James Cook University Hospital	71	56	53	10.0	9.9	37	84.0	86.5	88.0	37	89.9	92.3	91.7
Northern Ireland														
Belfast	Royal Belfast Hospital for Sick Children	60	196	189	8.8	9.0	126	90.4	91.1	91.6	127	96.5	97.3	96.6
Scotland														
Aberdeen	Royal Aberdeen Children's Hospital	75	33	26	7.6	7.5	15	89.4	89.0	92.0	16	93.0	92.9	92.8
Ayr	University Hospital Crosshouse	170	26	25	8.7	8.2	15	87.9	88.8	87.2	15	95.8	96.9	93.5
Dundee	Ninewells Hospital	73	22	20	8.1	8.5	13	92.6	91.2	92.9	14	96.1	94.6	99.7
Edinburgh	Royal Hospital for Sick Children	143	141	130	9.4	9.2	97	91.1	91.5	91.5	97	95.3	95.7	95.0
Glasgow	Royal Hospital for Children	56	102	84	8.7	9.5	59	92.6	92.7	93.3	59	96.2	96.3	97.3
Inverness	Raigmore Hospital	31	17	17	8.7	10.0	9	90.7	91.7	93.6	9	94.5	95.8	93.6
Lanarkshire	Wishaw General Hospital	162	43	41	9.0	9.2	22	88.4	89.2	91.5	30	97.4	97.4	98.1
Wales														
Cardiff	Children's Hospital for Wales	72	175	155	9.4	9.8	105	86.6	88.1	89.7	106	94.1	95.7	94.5

* 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**
England						
Birmingham	Birmingham Children's Hospital	104	269	49.9	49.9	50.8
Brighton	Royal Alexandra Children's Hospital	172	29	42.4	42.6	37.5
Bristol	Bristol Royal Hospital for Children	32	165	48.8	48.8	47.4
Cambridge	Addenbrookes Hospital	107	127	52.4	52.2	54.7
Cornwall	Royal Cornwall Hospital	94	34	64.7	64.9	59.5
Exeter	Royal Devon & Exeter Hospital	96	70	51.5	51.5	55.3
Hull	Hull University Teaching Hospitals	111	36	56.9	56.5	54.9
Leeds	St James's University Hospital	25	213	51.0	50.9	49.8
Leicester	Leicester Royal Infirmary	1	63	59.8	59.8	64.2
Liverpool	Alder Hey Children's Hospital	97	268	54.9	54.8	53.7
London - Central	Great Ormond Street Hospital for Children	90	175	47.8	47.5	44.6
London - East	Royal London Hospital	30	102	50.0	50.5	52.7
London - South East	King's College Hospital	17	155	53.4	53.1	54.9
London - South West	Royal Brompton Hospital	15	285	55.1	54.9	55.1
Manchester	Royal Manchester Children's Hospital	144	274	51.3	51.5	51.6
Newcastle	Great North Children's Hospital	59	160	58.0	57.7	62.1
North West Midlands	University Hospital of North Midlands	8	91	50.0	49.9	51.3
Norwich	Norfolk & Norwich University Hospital	98	57	61.3	61.8	69.3
Nottingham	Nottingham University Hospitals	62	152	48.7	48.9	49.2
Oxford	John Radcliffe Hospital	22	150	51.3	51.4	49.5
Plymouth	Derriford Hospital	139	34	50.6	50.2	51.6
Sheffield	Sheffield Children's Hospital	3	126	50.0	49.9	53.1
Southampton	Southampton General Hospital	29	186	55.1	55.1	56.1
Teeside	James Cook University Hospital	71	49	62.3	63.0	71.3
Northern Ireland						
Belfast	Royal Belfast Hospital for Sick Children	60	175	55.2	55.2	54.9
Scotland						
Aberdeen	Royal Aberdeen Children's Hospital	75	21	48.7	48.5	45.7
Ayr	University Hospital Crosshouse	170	25	59.2	58.9	59.4
Dundee	Ninewells Hospital	73	19	48.6	48.1	43.8
Edinburgh	Royal Hospital for Sick Children	143	123	56.2	56.5	56.0
Glasgow	Royal Hospital for Children	56	75	48.0	48.1	44.1
Inverness	Raigmore Hospital	31	16	48.8	48.6	43.6
Lanarkshire	Wishaw General Hospital	162	39	52.7	52.7	53.2
Wales						
Cardiff	Children's Hospital for Wales	72	142	54.5	54.9	57.3

Chronic Pseudomonas		Having at least 1 IV day		Receiving DNase treatment		Receiving hypertonic saline/mannitol treatment		Inhaled antibiotic use among patients with chronic Pseudomonas	
Number	**Proportion (%)**	**Number**	**Proportion (%)**	**Number**	**Proportion (%)**	**Number**	**Proportion (%)**	**Number**	**Proportion (%)**
15	5.1	92	31.1	173	58.4	94	31.8	15	100.0
<5	6.7	15	50.0	17	56.7	10	33.3	<5	100.0
11	6.2	65	36.7	126	71.2	87	49.2	11	100.0
9	6.6	32	23.4	88	64.2	83	60.6	9	100.0
<5	5.9	7	20.6	22	64.7	16	47.1	<5	100.0
6	8.1	17	23.0	60	81.1	69	93.2	6	100.0
<5	10.5	13	34.2	22	57.9	9	23.7	<5	100.0
8	3.5	85	37.1	142	62.0	8	3.5	7	87.5
<5	1.5	20	30.8	43	66.2	5	7.7	<5	100.0
21	7.2	125	42.7	164	56.0	49	16.7	21	100.0
<5	2.1	69	36.5	123	65.1	83	43.9	<5	100.0
16	15.1	39	36.8	76	71.7	80	75.5	15	93.8
17	9.4	64	35.6	119	66.1	59	32.8	15	88.2
26	8.5	85	27.8	223	72.9	111	36.3	26	100
16	5.3	93	31.0	149	49.7	131	43.7	16	100.0
15	8.4	71	39.9	99	55.6	39	21.9	13	86.7
5	5.3	46	48.4	64	67.4	23	24.2	<5	80.0
<5	3.0	11	16.7	44	66.7	16	24.2	<5	100.0
10	6.3	48	30.0	104	65.0	46	28.8	10	100.0
9	5.5	61	37.0	118	71.5	62	37.6	9	100.0
0	0.0	7	18.4	24	63.2	8	21.1	0	0.0
5	3.7	53	39.0	86	63.2	40	29.4	5	100.0
5	2.5	68	33.7	139	68.8	44	21.8	5	100.0
<5	1.9	20	37.7	40	75.5	13	24.5	<5	100.0
11	5.8	36	19.0	162	85.7	25	13.2	11	100.0
0	0.0	5	19.2	10	38.5	0	0.0	0	0.0
<5	4.0	12	48.0	<5	12.0	8	32.0	<5	100.0
<5	15.0	<5	20.0	8	40.0	6	30.0	<5	66.7
8	6.2	25	19.2	67	51.5	30	23.1	7	87.5
<5	2.4	27	32.1	23	27.4	33	39.3	<5	100.0
<5	5.9	<5	11.8	<5	11.8	<5	11.8	0	0.0
<5	2.4	7	17.1	8	19.5	13	31.7	<5	100.0
12	7.7	34	21.9	123	79.4	119	76.8	11	91.7

Appendix 2: Centre-level data tables

Adult centres/clinics providing data in 2019 – ordered alphabetically by country/city

Location	Name	Clinic ID	Total active	Number with annual review	Age (years) Mean	Age (years) Median	FEV_1% predicted at annual review Number	FEV_1% predicted at annual review Mean - unadjusted	FEV_1% predicted at annual review Mean - adjusted	FEV_1% predicted at annual review Median	Best FEV_1% predicted Number*	Best FEV_1% predicted Mean - unadjusted	Best FEV_1% predicted Mean - adjusted	Best FEV_1% predicted Median
England														
Birmingham	Birmingham Heartlands Hospital	27	318	305	32.8	30.1	270	65.2	65.5	65.0	286	69.7	70.0	70.7
Bristol	Bristol Royal Infirmary	106	234	228	31.2	30.0	216	69.7	69.8	69.3	216	74.4	74.4	75.6
Cambridge	Royal Papworth Hospital	51	330	309	31.2	28.7	281	65.1	64.9	65.8	289	70.4	70.0	72.4
Cornwall	Royal Cornwall Hospital	129	36	33	35.2	34.9	28	57.5	59.1	55.4	28	62.6	64.3	60.3
Exeter	Royal Devon & Exeter Hospital	34	118	112	32.4	29.1	95	68.8	69.0	69.3	105	73.4	73.5	73.8
Frimley	Frimley Park Hospital	19	142	136	32.2	32.0	125	64.0	64.5	67.1	125	69.6	69.9	71.5
Leeds	St James's University Hospital	42	398	386	34.8	33.0	320	61.0	62.1	60.9	330	67.2	68.3	67.1
Leicester	Glenfield Hospital	142	103	98	31.1	28.0	88	62.3	61.8	64.9	91	68.9	68.1	71.8
Liverpool	Liverpool Heart and Chest Hospital	66	349	324	31.8	29.9	313	68.0	68.2	70.0	313	72.7	72.7	74.5
London - East	St Bartholomew's Hospital	92	185	165	29.9	28.1	159	63.9	63.0	62.9	162	74.1	72.8	68.4
London - South East	King's College Hospital	5	240	226	30.5	28.6	210	66.5	66.2	69.0	210	69.2	68.6	70.5
London - South East	University Hospital Lewisham	105	59	55	31.8	30.1	43	58.7	59.0	57.2	46	61.0	61.3	60.1
London - South West	Royal Brompton Hospital	12	559	549	34.9	33.0	536	62.8	64.0	62.7	541	66.3	67.5	66.6
Manchester	Wythenshawe Hospital	102	464	442	32.0	30.0	423	57.2	57.5	55.4	434	62.8	63.0	62.9
Newcastle	Royal Victoria Infirmary	9	308	303	31.5	28.9	274	62.2	61.9	61.6	277	66.9	66.2	69.6
North West Midlands	University Hospital of North Midlands	74	140	136	30.0	26.0	128	60.8	60.0	63.2	128	66.1	64.9	68.9
Norwich	Norfolk & Norwich University Hospital	114	77	77	29.7	27.2	63	66.7	66.1	66.8	71	69.5	68.7	70.6
Nottingham	Nottingham University Hospitals	101	213	210	30.9	29.1	186	63.4	62.9	64.1	187	68.4	67.7	69.9
Oxford	John Radcliffe Hospital	128	137	121	31.6	28.3	103	61.1	60.3	57.6	104	65.9	64.8	65.4
Plymouth	Derriford Hospital	64	59	56	33.8	32.4	52	68.1	68.6	72.9	52	73.7	74.2	76.2
Sheffield	Northern General Hospital	65	208	199	31.1	28.8	179	67.9	67.7	72.3	187	72.5	72.2	76.6
Southampton	Southampton General Hospital	110	297	274	33.1	30.1	238	65.3	65.2	65.1	251	71.4	71.1	74.6
York & Hull	York Hospital	171	82	81	33.1	30.0	77	62.8	63.1	61.5	77	67.1	67.3	68.9
Northern Ireland														
Belfast	Belfast City Hospital	14	291	232	35.5	32.1	222	66.7	67.4	69.5	226	71.8	72.4	74.2
Scotland														
Aberdeen	Aberdeen Royal Infirmary	70	65	65	34.1	33.0	56	59.6	60.2	52.2	56	65.2	65.8	58.1
Edinburgh	Western General Hospital	44	248	239	33.0	30.8	217	63.1	63.3	63.6	217	68.6	68.6	69.8
Glasgow	Queen Elizabeth University Hospital	79	234	221	33.0	29.3	214	66.5	66.7	66.7	217	70.1	70.2	72.4
Wales														
Llandough	Llandough Hospital	68	272	254	32.3	30.9	237	66.5	66.8	67.7	237	71.7	71.8	72.8

* 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	297	23.5	23.5	23.0
Bristol	Bristol Royal Infirmary	106	228	23.4	23.5	22.7
Cambridge	Royal Papworth Hospital	51	308	22.7	22.8	22.3
Cornwall	Royal Cornwall Hospital	129	33	22.8	22.5	21.3
Exeter	Royal Devon & Exeter Hospital	34	110	24.8	24.8	24.8
Frimley	Frimley Park Hospital	19	136	22.5	22.5	22.1
Leeds	St James's University Hospital	42	386	23.4	23.2	22.8
Leicester	Glenfield Hospital	142	97	22.9	23.1	21.9
Liverpool	Liverpool Heart and Chest Hospital	66	324	23.6	23.7	22.8
London - East	St Bartholomew's Hospital	92	165	22.5	22.8	21.8
London - South East	King's College Hospital	5	226	23.1	23.3	22.7
London - South East	University Hospital Lewisham	105	55	21.8	21.8	21.1
London - South West	Royal Brompton Hospital	12	549	22.8	22.6	22.4
Manchester	Wythenshawe Hospital	102	442	22.5	22.5	22.1
Newcastle	Royal Victoria Infirmary	9	300	23.2	23.3	22.5
North West Midlands	University Hospital of North Midlands	74	136	22.7	22.9	22.2
Norwich	Norfolk & Norwich University Hospital	114	75	21.9	22.2	21.1
Nottingham	Nottingham University Hospitals	101	210	22.3	22.5	21.8
Oxford	John Radcliffe Hospital	128	121	22.1	22.2	21.9
Plymouth	Derriford Hospital	64	56	23.9	23.8	23.5
Sheffield	Northern General Hospital	65	198	23.6	23.7	22.9
Southampton	Southampton General Hospital	110	272	23.3	23.3	22.5
York & Hull	York Hospital	171	81	22.8	22.8	22.0
Northern Ireland						
Belfast	Belfast City Hospital	14	232	24.2	23.9	23.5
Scotland						
Aberdeen	Aberdeen Royal Infirmary	70	65	23.6	23.4	22.6
Edinburgh	Western General Hospital	44	236	23.0	22.9	22.5
Glasgow	Queen Elizabeth University Hospital	79	221	23.8	23.8	23.2
Wales						
Llandough	Llandough Hospital	68	254	23.2	23.2	22.4

Chronic *Pseudomonas*		Having at least 1 IV day		Receiving DNase treatment		Receiving hypertonic saline/mannitol treatment		Inhaled antibiotic use among patients with chronic *Pseudomonas*	
Number	**Proportion (%)**	**Number**	**Proportion (%)**	**Number**	**Proportion (%)**	**Number**	**Proportion (%)**	**Number**	**Proportion (%)**
148	48.5	179	58.7	234	76.7	138	45.2	143	96.6
81	35.5	140	61.4	132	57.9	151	66.2	70	86.4
123	39.8	186	60.2	201	65.0	146	47.2	112	91.1
10	30.3	19	57.6	19	57.6	11	33.3	6	60.0
34	30.4	50	44.6	79	70.5	67	59.8	31	91.2
61	44.9	69	50.7	104	76.5	52	38.2	60	98.4
168	43.5	222	57.5	293	75.9	107	27.7	153	91.1
36	36.7	51	52.0	60	61.2	16	16.3	28	77.8
142	43.8	187	57.7	232	71.6	87	26.9	128	90.1
74	44.8	94	57.0	137	83.0	113	68.5	70	94.6
52	23.0	116	51.3	179	79.2	97	42.9	52	100.0
23	41.8	32	58.2	36	65.5	14	25.5	23	100.0
282	51.4	276	50.3	487	88.7	279	50.8	248	87.9
206	46.6	242	54.8	310	70.1	178	40.3	198	96.1
115	38.0	146	48.2	193	63.7	49	16.2	100	87.0
55	40.4	84	61.8	94	69.1	52	38.2	48	87.3
20	26.0	39	50.6	51	66.2	38	49.4	17	85.0
78	37.1	133	63.3	159	75.7	95	45.2	71	91.0
42	34.7	69	57.0	93	76.9	45	37.2	31	73.8
24	42.9	29	51.8	31	55.4	35	62.5	21	87.5
101	50.8	131	65.8	172	86.4	39	19.6	94	93.1
104	38.0	154	56.2	172	62.8	123	44.9	92	88.5
29	35.8	41	50.6	59	72.8	20	24.7	26	89.7
88	37.9	77	33.2	160	69.0	74	31.9	75	85.2
22	33.8	28	43.1	32	49.2	15	23.1	16	72.7
74	31.0	99	41.4	130	54.4	51	21.3	57	77.0
74	33.5	96	43.4	104	47.1	32	14.5	65	87.8
92	36.2	130	51.2	177	69.7	159	62.6	86	93.5

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	9035	89.7
c.350G->A	p.Arg117His	R117H	587	5.8
c.1652G->A	p.Gly551Asp	G551D	581	5.8
c.1624G->T	p.Gly542X	G542X	356	3.5
c.489+1G->T		621+1G->T	258	2.6
c.3909C->G	p.Asn1303Lys	N1303K	159	1.6
c.1585-1G->A		1717-1G->A	157	1.6
c.1766+1G->A		1898+1G->A	140	1.4
c.200C->T	p.Pro67Leu	P67L	132	1.3
c.3454G->C	p.Asp1152His	D1152H	131	1.3
c.3528delC	p.Lys1177SerfsX15	3659delC	101	1.0
c.3140-26A->G		3272-26A->G	99	1.0
c.1679G->C	p.Arg560Thr	R560T	93	0.9
c.1477C->T	p.Gln493X	Q493X	87	0.9
c.1657C->T	p.Arg553X	R553X	86	0.9
c.1519_1521delATC	p.Ile507del	I507del	79	0.8
c.254G->A	p.Gly85Glu	G85E	78	0.8
c.3717+12191C->T		3849+10kbC->T	77	0.8
c.2657+5G->A		2789+5G->A	68	0.7
c.178G->T	p.Glu60X	E60X	68	0.7
c.1022_1023insTC	p.Phe342HisfsX28	1154insTC	67	0.7
c.3846G->A	p.Trp1282X	W1282X	56	0.6
c.948delT	p.Phe316LeufsX12	1078delT	54	0.5
c.1646G->A	p.Ser549Asn	S549N	53	0.5
c.2052delA	p.Lys684AsnfsX38	2184delA	48	0.5
c.1364C->A	p.Ala455Glu	A455E	47	0.5
c.617T->G	p.Leu206Trp	L206W	44	0.4
c.1040G->C	p.Arg347Pro	R347P	37	0.4
c.579+3A->G		711+3A->G	34	0.3
c.1558G->T	p.Val520Phe	V520F	31	0.3
c.3484C->T	p.Arg1162X	R1162X	31	0.3
c.2657+2_2657+3insA		2789+2insA	31	0.3
c.1000C->T	p.Arg334Trp	R334W	25	0.2
c.1040G->A	p.Arg347His	R347H	25	0.2
c.2988+1G->A		3120+1G->A	24	0.2
c.1055G->A	p.Arg352Gln	R352Q	22	0.2
c.1753G->T	p.Glu585X	E585X	22	0.2
c.1006_1007insG	p.Ile336SerfsX28	1138insG	21	0.2
c.2834C->T	p.Ser945Leu	S945L	21	0.2
c.2583delT	p.Phe861LeufsX3	2711delT	20	0.2
c.3472C->T	p.Arg1158X	R1158X	19	0.2
c.2490+1G->A		2622+1G->A	19	0.2
c.2125C->T	p.Arg709X	R709X	18	0.2
c.532G->A	p.Gly178Arg	G178R	18	0.2
c.1367T->C	p.Val456Ala	V456A	17	0.2
c.1210-12[5] (AJ574948.1:g.152T[5])		5T	17	0.2
c.1705T->G	p.Tyr569Asp	Y569D	16	0.2
c.1393-1G->A		1525-1G->A	16	0.2
c.3718-2477C->T		3849+10kbC->T	16	0.2
c.3806T->A	p.Ile1269Asn	I1269N	15	0.1
c.3197G->A	p.Arg1066His	R1066H	15	0.1
c.658C->T	p.Gln220X	Q220X	14	0.1
c.292C->T	p.Gln98X	Q98X	12	0.1
c.2052_2053insA	p.Gln685ThrfsX4	2184insA	12	0.1
c.349C->T	p.Arg117Cys	R117C	12	0.1
c.2537G->A	p.Trp846X	W846X	11	0.1
c.3737C->T	p.Thr1246Ile	T1246I	11	0.1
c.2875delG	p.Ala959HisfsX9	3007delG	11	0.1
c.579+1G->T		711+1G->T	11	0.1
c.2988G->A		3120G->A	10	0.1
c.1029delC	p.Cys343X	1161delC	9	0.1
c.3705T->G	p.Ser1235Arg	S1235R	9	0.1
c.1466C->A	p.Ser489X	S489X	9	0.1
c.4196_4197delTC	p.Cys1400X	4326delTC	8	0.1
c.224G->A	p.Arg75Gln	R75Q	8	0.1
c.695T->A	p.Val232Asp	V232D	8	0.1
c.3196C->T	p.Arg1066Cys	R1066C	8	0.1
c.3276C->A	p.Tyr1092X	Y1092X(C->A)	8	0.1
c.1679+1G->C		1811+1G->C	8	0.1
c.1675G->A	p.Ala559Thr	A559T	7	0.1
c.2051_2052delAAinsG	p.Lys684SerfsX38	2183AA->G or 2183de-lAA->G	7	0.1
c.494T->C	p.Leu165Ser	L165S	7	0.1
c.2353C->T	p.Arg785X	R785X	6	0.1
c.1986_1989delAACT	p.Thr663ArgfsX8	2118del4	6	0.1
c.3208C->T	p.Arg1070Trp	R1070W	6	0.1

Nucleotide	Protein	Legacy name	N	%
c.1721C->A	p.Pro574His	P574H	6	0.1
c.3468G->A		3600G->A	6	0.1
c.2128A->T	p.Lys710X	K710X	6	0.1
c.1329_1330insAGAT	p.Ile444ArgfsX3	1461ins4	6	0.1
c.1766+1G->T		1898+1G->T	6	0.1
c.3761T->G	p.Leu1254X	L1254X	5	0.0
c.2290C->T	p.Arg764X	R764X	5	0.0
c.1523T->G	p.Phe508Cys	F508C	5	0.0
c.223C->T	p.Arg75X	R75X	5	0.0
c.3848G->T	p.Arg1283Met	R1283M	5	0.0
c.2551C->T	p.Arg851X	R851X	5	0.0
c.1687T->A	p.Tyr563Asn	Y563N	<5	-
c.1538A->G	p.Asp513Gly	D513G	<5	-
c.1572C->A	p.Cys524X	C524X	<5	-
c.2249C->T	p.Pro750Leu	P750L	<5	-
c.2012delT	p.Leu671X	2143delT	<5	-
c.1116+1G->A		1248+1G->A	<5	-
c.2909G->A	p.Gly970Asp	G970D	<5	-
c.3718-1G->A		3850-1G->A	<5	-
c.443T->C	p.Ile148Thr	I148T	<5	-
c.3884_3885insT	p.Ser1297PhefsX5	4016insT	<5	-
c.1680A->C	p.Arg560Ser	R560S	<5	-
c.1393-2A->G		1525-2A->G	<5	-
c.262_263delTT	p.Leu88IlefsX22	394delTT	<5	-
c.2215delG	p.Val739TyrfsX16	2347delG	<5	-
c.2464G->T	p.Glu822X	E822X	<5	-
c.595C->T	p.His199Tyr	H199Y	<5	-
c.1645A->C	p.Ser549Arg	S549R(A->C)	<5	-
c.1679G->A	p.Arg560Lys	R560K	<5	-
c.850dupA	p.Met284AsnfsX3	977insA	<5	-
c.349C->G	p.Arg117Gly	R117G	<5	-
c.3095A->G	p.Tyr1032Cys	Y1032C	<5	-
c.165-3C>T		297-3C->T	<5	-
c.2600_2601insA	p.Val868SerfsX28	2732insA	<5	-
c.1736A->G	p.Asp579Gly	D579G	<5	-
c.2900T->C	p.Leu967Ser	L967S	<5	-
c.3659delC	p.Thr1220LysfsX8	3791delC	<5	-
c.1505T->C	p.Ile502Thr	I502T	<5	-
c.2491G->T	p.Glu831X	E831X	<5	-
c.1585-8G->A		1717-8G->A	<5	-
c.3292T->C	p.Trp1098Arg	W1098R	<5	-
c.577G->T	p.Glu193X	E193X	<5	-
c.2260G->A	p.Val754Met	V754M	<5	-

Nucleotide	Protein	Legacy name	N	%
c.4147_4148insA	p.Ile1383AsnfsX3	4279insA	<5	-
c.2896delA	p.Thr966ArgfsX2	3028delA	<5	-
c.1647T->G	p.Ser549Arg	S549R(T->G)	<5	-
c.4004T->C	p.Leu1335Pro	L1335P	<5	-
c.3908delA	p.Asn1303ThrfsX25	4040delA	<5	-
c.54-5940_273+10250del21kb	p.Ser18ArgfsX16	CFTRdele2,3	<5	-
c.3353C->T	p.Ser1118Phe	S1118F	<5	-
c.2991G->C	p.Leu997Phe	L997F	<5	-
c.328G->C	p.Asp110His	D110H	<5	-
c.350G->T	p.Arg117Leu	R117L	<5	-
c.4046G->A	p.Gly1349Asp	G1349D	<5	-
c.274G->A	p.Glu92Lys	E92K	<5	-
c.1766+5G->T		1898+5G->T	<5	-
c.[1210–12[5];1210-34TG[12]]		5T;TG12	<5	-
c.[1210–12[5];1210-34TG[13]]		5T;TG13	<5	-
c.3475T->C	p.Ser1159Pro	S1159P	<5	-
c.1679+1.6kbA->G		1811+1.6kbA->G	<5	-
c.3266G->A	p.Trp1089X	W1089X	<5	-
c.442delA	p.Ile148LeufsX5	574delA	<5	-
c.1007T->A	p.Ile336Lys	I336K	<5	-
c.1651G->A	p.Gly551Ser	G551S	<5	-
c.3181G->C	p.Gly1061Arg	G1061R	<5	-
c.79G->T	p.Gly27X	G27X	<5	-
c.1046C->T	p.Ala349Val	A349V	<5	-
c.2780T->C	p.Leu927Pro	L927P	<5	-
c.3882_3885delTATT	p.Ile1295PhefsX32	4010del4	<5	-
c.91C->T	p.Arg31Cys	R31C	<5	-
c.1001G>A	p.Arg334Gln	R334Q	<5	-
c.3080T->C	p.Ile1027Thr	I1027T	<5	-
c.164+2T>C		296+2T->C	<5	-
c.1327G->T	p.Asp443Tyr	D443Y	<5	-
c.2374C->T	p.Arg792X	R792X	<5	-
c.1766+1G->C		1898+1G->C	<5	-
c.3700A->G	p.Ile1234Val	I1234V	<5	-
c.2195T->G	p.Leu732X	L732X	<5	-
c.3310G->T	p.Glu1104X	E1104X	<5	-
c.3752G->A	p.Ser1251Asn	S1251N	<5	-
c.1340delA	p.Lys447ArgfsX2	1471delA	<5	-
c.4077_4080delTGTTinsAA	p.Val1360delfsX?	4209TGTT->AA	<5	-
c.2668C->T	p.Gln890X	Q890X	<5	-
c.3988C->T	p.Gln1330X	Q1330X	<5	-
c.4147_4148insA	p.Ile1383AsnfsX3	4279insA	<5	-

Nucleotide	Protein	Legacy name	N	%
c.2620-26A->G		2752-26A->G	<5	-
c.1724T->A	p.Phe575Tyr	F575Y	<5	-
c.1418delG	p.Gly473GlufsX54	1548delG	<5	-
c.1682C->A	p.Ala561Glu	A561E	<5	-
c.92G>T	p.Arg31Leu	R31L	<5	-
c.3011_3019delCTATAGCAG or c.3009_3017delAGCTATAGC	p.Ala1004_Ala1006del	3143del9	<5	-
c.53+1G->T		185+1G->T	<5	-
c.1647T->A	p.Ser549Arg	S549R	<5	-
c.1687T->G	p.Tyr563Asp	Y563D	<5	-
c.859_863delAACTT	p.Asn287LysfsX19	991del5	<5	-
c.1240C->T	p.Gln414X	Q414X	<5	-
c.470_483del14	p.Phe157X	602del14	<5	-
c.3476C->T	p.Ser1159Phe	S1159F	<5	-
c.2645G->A	p.Trp882X	W882X	<5	-
c.3209G->A	p.Arg1070Gln	R1070Q	<5	-
	p.Ser466X	S466X	<5	-
c.1727G->C	p.Gly576Ala	G576A	<5	-
c.(53+1_54-1)_(489+1_490-1)del		CFTRdele2-4	<5	-
c.220C->T	p.Arg74Trp	R74W	<5	-
c.137C->A	p.Ala46Asp	A46D	<5	-
c.1703delT	p.Leu568CysfsX4	1833delT	<5	-
c.1654C->T	p.Gln552X	Q552X	<5	-
c.263T>A or c.263T>G	p.Leu88X	L88X	<5	-
c.2739T->A	p.Tyr913X	Y913X	<5	-
c.1545_1546delTA	p.Tyr515X	1677delTA	<5	-
c.233dupT	p.Trp79LeufsX32	365-366insT	<5	-
c.717delG	p.Leu240X	849delG	<5	-
c.1477_1478delCA	p.Gln493ValfsX10	1609delCA	<5	-
c.2421A->G	p.Ile807Met	I807M	<5	-
c.274-2A->G		406-2A->G	<5	-
c.3205G->A	p.Gly1069Arg	G1069R	<5	-
c.164+1G>A		296+1G->A	<5	-
c.296C->T	p.Pro99Leu	P99L	<5	-
c.1573C->T	p.Gln525X	Q525X	<5	-
c.3873+2T->C		4005+2T->C	<5	-
c.601G->A	p.Val201Met	V201M	<5	-
c.1202G->A	p.Trp401X	W401X(TAG)	<5	-
c.2859_2890delACATTCT-GTTCTTCAAGCACCTATGT-CAACCC	p.Leu953PhefsX11	2991del32	<5	-
c.1203G->A	p.Trp401X	W401X(TGA)	<5	-
c.3302T->G	p.Met1101Arg	M1101R	<5	-

Nucleotide	Protein	Legacy name	N	%
c.4231C->T	p.Gln1411X	Q1411X	<5	-
c.3194T->C	p.Leu1065Pro	L1065P	<5	-
c.1037T->C	p.Leu346Pro	L346P	<5	-
c.613C->T	p.Pro205Ser	P205S	<5	-
c.1081delT	p.Trp361GlyfsX8	1213delT	<5	-
c.1021T->C	p.Ser341Pro	S341P	<5	-
c.3611G->A	p.Trp1204X	W1204X(3743G->A)	<5	-
c.2002C->T	p.Arg668Cys	R668C	<5	-
c.(743+1_744-1)_(1584+1_1585-1)dup		CFTRdup6b-10	<5	-
c.(53+1_54-1)_(164+1_165-1)del		CFTRdele2	<5	-
c.3158C->T	p.Thr1053Ile	T1053I	<5	-
c.1837G->A	p.Ala613Thr	A613T	<5	-
c.3773_3774insT	p.Leu1258PhefsX7	3905insT	<5	-
c.3717+5G->A		3849+5G->A	<5	-
c.2989-1G->A		3121-1G->A	<5	-
c.3017C->A	p.Ala1006Glu	A1006E	<5	-
c.2735C->A	p.Ser912X	S912X	<5	-
c.11C>A	p.Ser4X	S4X	<5	-
c.3230T->C	p.Leu1077Pro	L1077P	<5	-
c.3872A->G	p.Gln1291Arg	Q1291R	<5	-
c.4111G->T	p.Glu1371X	E1371X	<5	-
c.3745G->A	p.Gly1249Arg	G1249R	<5	-
c.50delT	p.Phe17SerfsX8	182delT	<5	-
c.3718-3T->G		3850-3T->G	<5	-
c.3297C->A	p.Phe1099Leu	F1099L	<5	-
c.1209+1G->A		1341+1G->A	<5	-
'Other' selected			**749**	**7.4**

Cystic Fibrosis Trust

cysticfibrosis.org.uk

Fighting for a Life Unlimited