Cystic Fibrosis strength in numbers

UK Cystic Fibrosis Registry
Annual Data Report 2017

Published August 2018

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

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The UK CF Registry Steering Committee

Acknowledgements

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

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@CFTrust

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Foreword

I am very pleased to present to you the 2017 data report of the UK Cystic Fibrosis Registry. The Registry is critical to improving the health of people with cystic fibrosis (CF), while at the same time it is the CF community who make the Registry the world-class resource it is, by sharing their data. The Registry supports funding of CF services that reflects the complexity of the needs of individual people with the condition, and the excellent quality data it holds enables innovatively designed clinical trials and ground-breaking research. Through this report, the Registry and the data shared by people with CF enable us to communicate important trends in CF in the UK.

It has been a busy year for research using Registry data. This includes scientific publications using Registry data to evaluate the effect of CF treatments[1], and to produce conditional estimates of survival for people who have reached certain age-milestones[2].

Another paper this year suggests an association between higher IV antibiotic use in a CF centre and higher lung function[3]. You'll see in Sections 2 and 3 of this report we have, for the first time, published IV use by individual CF centres. A publication describing Anti-Staphylococcal Antibiotic Prophylaxis in young children with CF[4] highlights the importance of the Registry-based clinical trial CF START in finding definitive answers to questions highlighted by observational research. You can read more about this clinical trial on the Cystic Fibrosis Trust website.

Through the Registry, the CF community is an exemplar of the power of patient data. With our work on new drug safety monitoring, it has been possible to study the real-world effects of precision medicines, leading to a publication supporting disease modification with ivacaftor treatment[5]. We hope that this capability will encourage decision makers to put their trust in the robust data of the Registry when making future decisions about making CF therapies available in the UK.

Reading through the report this year, I am really encouraged to see improvements in lung function, incidence of chronic *Pseudomonas*, and prescribing of recommended medicines like DNase. However, despite these positive changes, we have a lot of work still to do. Registry data shows us that the burden of treatment for people with CF is high, and with breakthrough medicines still not reimbursed by the NHS, too few are able to benefit from disease modifying treatments.

As CF evolves over the coming years, and new treatments continue to become available, we will need high quality Registry data more than ever, to monitor the present, predict the future, and make the right choices. I want to take this opportunity to express my personal gratitude, and that of the entire CF community, to those people who consent to participate in the UK CF Registry, and the clinical teams who work tirelessly to support the Registry as well as the people they care for.

David Ramsden
Chief Executive

[1] Newsome SJ et al. Estimating long-term treatment effects in observational data: A comparison of the performance of different methods under real-world uncertainty, Stat Med. 2018 37(15):2367-2390

[2] Keogh RH et al. Up-to-date and projected estimates of survival for people with cystic fibrosis using baseline characteristics: A longitudinal study using UK patient registry data, J Cyst Fibros. 2018 17(2):218-227

[3] 'Hoo ZH et al. Do cystic fibrosis centres with the lowest FEV_1 still use the least amount of intravenous antibiotics? A registry-based comparison of intravenous antibiotic use among adult CF centres in the UK, J Cyst Fibros. 2017 17(3):360-367

[4] Hurley MN et al. Early Respiratory Bacterial Detection and Antistaphylococcal Antibiotic Prophylaxis in Young Children with Cystic Fibrosis, Ann Am Thorac Soc. 2018 15(1):42-48

[5] Bessonova L et al. Data from the US and UK cystic fibrosis registries support disease modification by CFTR modulation with ivacaftor, Thorax. 2018 73(8):731-740

Executive summary

Ten and a half thousand people with cystic fibrosis (or their parents) have consented to having their anonymised data captured by the UK CF Registry, and this report contains summary information on 9887 (96%) annual review visits in the calendar year 2017. The report uses amalgamated data from all the adult and paediatric centres in the UK, as well as summary data at an individual centre level.

Some of the important highlights of this report include:

- 53.3% of the total population are male, although in the childhood years there are more females (section 1.2)
- Over 98% of the population have two genotype values recorded (section 1.38)
  - 49.1% are homozygous for delta F508
  - 40.4% are heterozygous for delta F508
- 2640 (27%) were diagnosed through newborn screening (section 1.11). For children age five who were born after the introduction of universal CF newborn screening in the UK, this rises to nearly 93%.
- Respiratory infections were the commonest other presentation (37%) in those not identified through new born screening (section 1.11)
- 78% of people with cystic fibrosis are taking at least one inhaled treatment, with 20% on three different inhaled therapies (section 1.30)
- The median FEV_1% predicted for children under 16 at annual review is 89%, for those 16 and over it is 65% (section 1.12)
- Chronic *Pseudomonas aeruginosa* infection increases with age, with 28% of the total population classified as this. When we look specifically at the adult population, this increases to 44.5% (section 1.17)
- The incidence of new NTM infection is similar to the last two years at 2.5%, with Mycobacterium abscessus complex accounting for 55% of these (section 1.18)
- We have for the first time published IV use and data completeness by individual CF centre. Overall completeness for key outcome FEV_1 is 95.6%. You can find these results in Section 2 and 3 of the report

Eleven years of good quality long-term data has allowed us to add 2013 as a benchmark year into many of our charts and tables alongside 2008, which we have used previously. This allows some easy comparisons of changes in outcome measures and treatments over time.

In response to requests from the CF community, we have introduced two age cut offs for 'adult' and 'paediatric' data, with breaks at both 16 and 18 years presented. In the centre level reports there is no age cut off, all individuals cared for by a centre are included. We hope this will make the report more useful for people with CF, students, clinical teams, researchers and policy makers.

It is important that we continue to receive feedback on these reports so that we can improve them each year. Please let us know your thoughts by emailing **registry@cysticfibrosis.org.uk** or tweeting **@CFTrust.**

Thank you to everyone who consents to participate in the UK CF Registry, making this report possible, and to the clinical teams who have supported the Cystic Fibrosis Registry over the past 11 years.

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

Introduction

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

You can find a Glossary of scientific and clinical terms on page 59.

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

Cystic fibrosis

Cystic fibrosis is an inherited disease caused by a faulty 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. They may 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**.

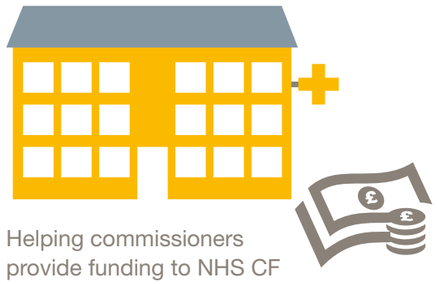
The purpose of the UK CF Registry is to improve the health of people with cystic fibrosis. This is done in a number of ways:

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 of treating, and beating 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 written 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 is 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

	2013	2014	2015	2016	2017
CF patients registered[1]	10338	10583	10810	10461	10469
Excluding diagnoses that year	10076	10356	10586	10214	10255
CF patients with an annual review; n(%)[2]	9052 (90%)	9432 (91%)	9587 (91%)	9695 (95%)	9887 (96%)
Age in years; median[3]	18	19	19	20	20
All newly diagnosed patients (NBS and other)[4]	301	291	300	303	214
Number of patients born identified by NBS[4]	177	164	168	216	172
Age at diagnosis in months; median[3]	3	2	2	2	2
Adults aged 16 years and over; %[3]	57.6	59.3	59.9	60.4	60.6
Males; %[3]	52.9	53	53	53.2	53.3
Genotyped; %[3] (both mutations identified)	97.2	97.7	98.1	98.4	98.4
Total deaths reported (%)[5]	146 (1.4%)	132 (1.2%)	125 (1.2%)	148 (1.5%)	132 (1.3%)
Age at death in years; median (95% CI)[5]	29 (27, 31)	28 (25.5, 32)	28 (27, 33)	31 (29, 33)	31 (29, 35)

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:

[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.

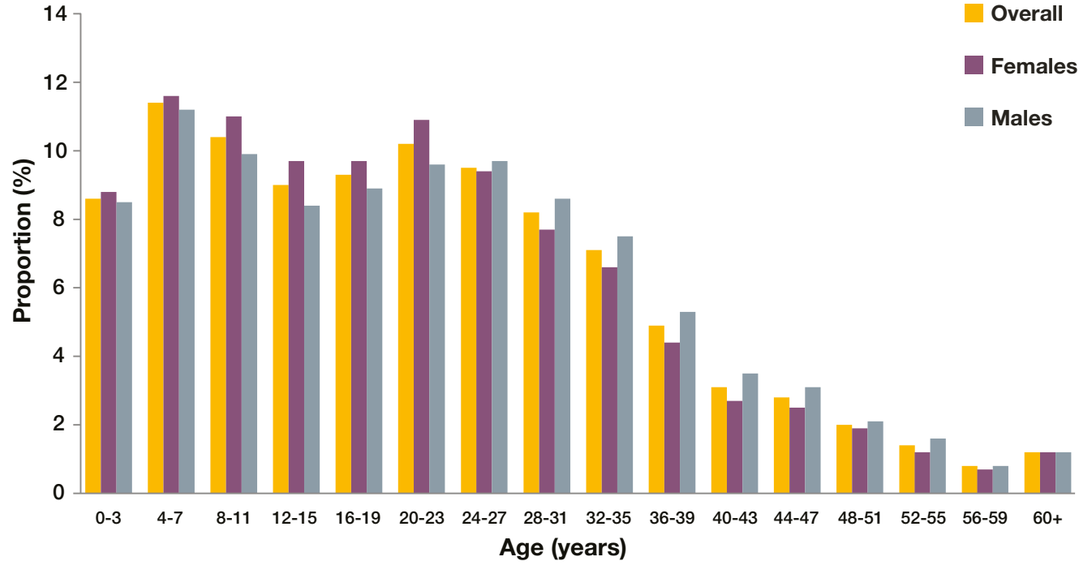
[3] Calculated from patients with an annual review in the given year (see footnote 2 above).

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

1.2 Age distribution by gender
N=9887

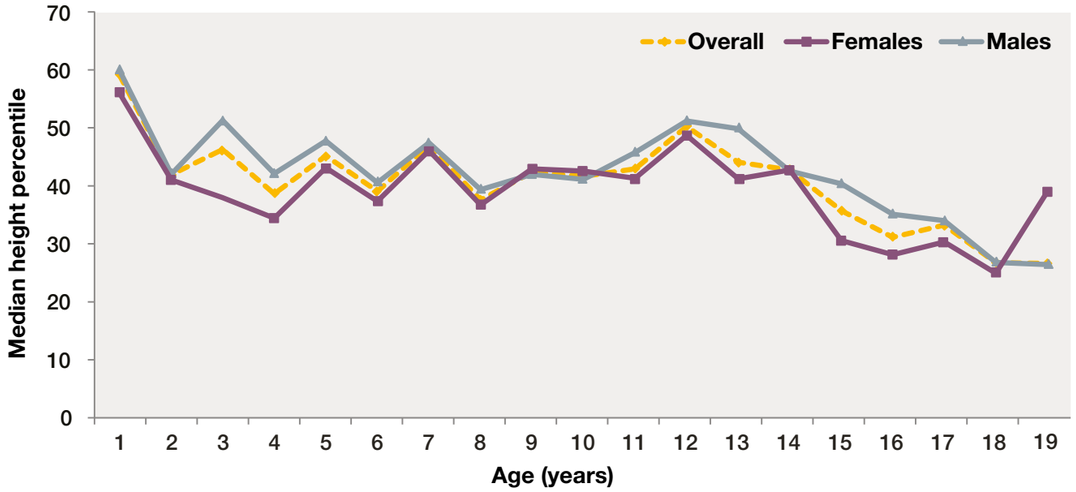
The following chart shows the mix of ages and genders in the CF population in the UK.



Age	All; n (%)	Females; n (%)	Males; n (%)
0-3	854 (8.6)	405 (8.8)	449 (8.5)
4-7	1124 (11.4)	534 (11.6)	590 (11.2)
8-11	1031 (10.4)	508 (11.0)	523 (9.9)
12-15	889 (9.0)	447 (9.7)	442 (8.4)
16-19	918 (9.3)	450 (9.7)	468 (8.9)
20-23	1010 (10.2)	502 (10.9)	508 (9.6)
24-27	942 (9.5)	433 (9.4)	509 (9.7)
28-31	810 (8.2)	356 (7.7)	454 (8.6)
32-35	698 (7.1)	304 (6.6)	394 (7.5)
36-39	483 (4.9)	203 (4.4)	280 (5.3)
40-43	311 (3.1)	127 (2.7)	184 (3.5)
44-47	280 (2.8)	115 (2.5)	165 (3.1)
48-51	201 (2.0)	90 (1.9)	111 (2.1)
52-55	138 (1.4)	56 (1.2)	82 (1.6)
56-59	76 (0.8)	34 (0.7)	42 (0.8)
60+	122 (1.2)	57 (1.2)	65 (1.2)
<16	3898 (39.4)	1894 (41.0)	2004 (38.1)
≥16	5989 (60.6)	2727 (59.0)	3262 (61.9)
<18	4329 (43.8)	2108 (45.6)	2221 (42.2)
≥18	5558 (56.2)	2513 (54.4)	3045 (57.8)
Overall	9887	4621	5266

1.3 Height percentiles of children and young people (<20 years)[6] N=4730

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.

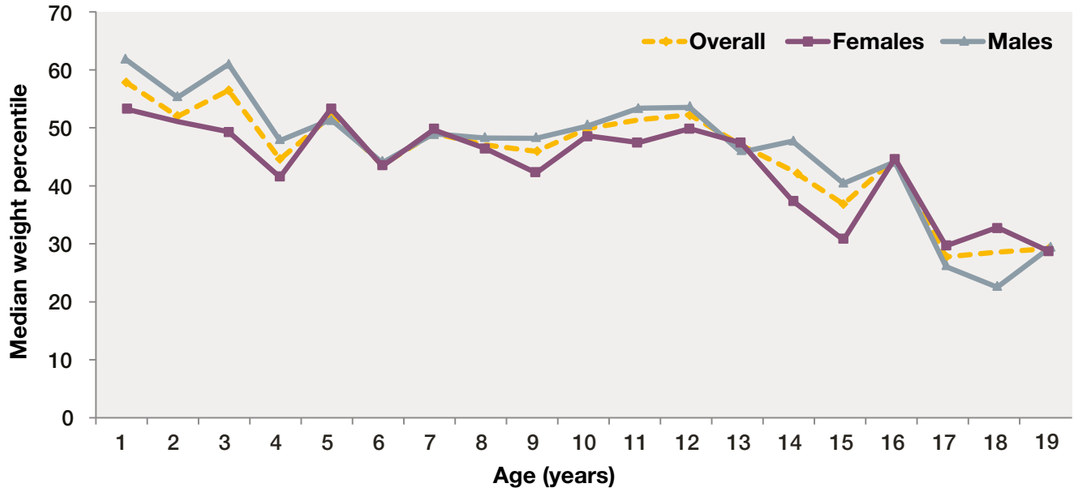


	Overall			Female			Male		
Age	n	Median	IQR	n	Median	IQR	n	Median	IQR
1	239	59.3	28.3-83.0	111	56.2	23.4-80.4	128	60.1	28.9-84.1
2	257	42.2	20.7-65.1	122	41.3	20.3-66.9	135	42.3	20.9-64.7
3	252	46.5	22.6-75.8	118	38.3	17.9-69.9	134	51.5	26.7-76.6
4	265	38.9	16.4-65.1	125	34.7	13.3-65.1	140	42.4	21.3-66.2
5	280	45.5	23.2-69.3	132	43.4	24.3-68.8	148	48.0	20.0-69.3
6	318	39.3	19.3-64.2	155	37.6	17.0-63.3	163	40.8	20.9-66.9
7	256	47.3	24.3-70.7	120	46.3	21.9-71.3	136	47.7	28.3-70.2
8	282	37.8	15.1-65.7	145	37.0	15.1-64.0	137	39.6	18.3-66.5
9	247	42.6	17.7-67.3	121	43.2	18.5-67.3	126	42.3	17.7-65.3
10	251	41.9	17.9-73.3	121	42.8	19.7-76.1	130	41.4	16.6-69.9
11	245	43.2	25.6-72.3	118	41.6	26.0-69.1	127	46.0	25.5-74.4
12	234	50.5	26.7-71.8	110	49.0	27.0-69.0	124	51.4	26.3-73.3
13	232	44.3	15.9-70.2	125	41.5	15.2-68.6	107	50.1	16.3-75.6
14	207	43.0	12.6-67.3	103	43.0	11.6-66.2	104	42.8	13.5-69.6
15	211	36.0	13.8-61.5	108	30.9	11.4-61.6	103	40.6	14.2-60.2
16	233	31.4	11.5-63.2	121	28.4	11.2-61.8	112	35.3	11.6-64.3
17	191	33.5	10.5-65.8	89	30.6	10.7-65.6	102	34.3	9.6-66.6
18	235	27.0	7.7-59.0	117	25.2	5.6-56.5	118	27.0	9.2-59.2
19	249	26.9	10.3-53.8	118	39.3	10.3-65.2	131	26.7	10.3-48.1
Overall	**4684***	**41.0**	**17.1-67.7**	**2279**	**40.3**	**16.0-67.0**	**2405**	**42.4**	**18.5-68.3**

*number with non-missing data

1.4 Weight percentiles of children and young people (<20 years)[6] N=4730

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.

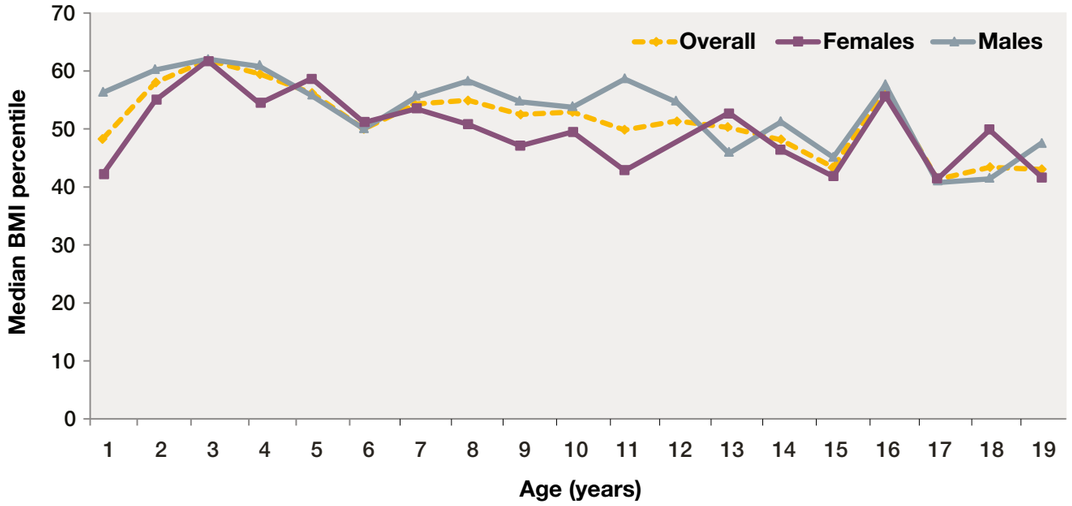


	Overall			Female			Male		
Age	n	Median	IQR	n	Median	IQR	n	Median	IQR
1	244	58.0	29.0-79.1	114	53.3	25.2-72.9	130	61.9	35.4-84.9
2	260	52.1	25.3-77.8	123	51.2	24.2-78.5	137	55.4	26.3-77.6
3	253	56.6	30.1-77.7	118	49.5	22.5-77.6	135	61.1	39.8-79.6
4	265	44.7	22.9-71.9	125	41.6	18.6-69.9	140	47.9	26.8-74.3
5	280	52.2	25.6-74.1	132	53.5	29.7-74.7	148	51.6	23.9-73.0
6	318	43.7	22.2-70.1	155	43.5	21.7-70.9	163	44.1	22.2-69.6
7	257	49.2	27.2-77.5	120	49.8	28.1-79.2	137	49.1	27.2-77.1
8	283	47.2	24.2-73.5	145	46.6	24.0-65.8	138	48.3	26.8-75.9
9	248	46.1	19.2-73.8	121	42.3	17.2-70.9	127	48.3	25.6-75.4
10	253	50.0	21.4-77.8	122	48.8	20.3-79.5	131	50.4	22.3-72.7
11	245	51.5	30.0-75.4	118	47.5	27.7-68.5	127	53.5	30.5-78.2
12	235	52.4	29.3-75.3	111	50.0	27.8-74.1	124	53.7	34.5-80.7
13	232	47.2	21.0-71.8	125	47.5	25.2-71.5	107	45.9	20.9-72.0
14	207	42.7	19.3-66.4	103	37.5	18.0-64.2	104	47.8	22.7-74.3
15	211	36.9	13.8-63.0	108	30.8	11.6-55.3	103	40.5	18.8-65.7
16	233	44.8	13.3-75.6	121	44.8	15.2-69.8	112	44.3	11.9-76.1
17	192	27.8	9.4-67.8	89	29.7	7.2-75.8	103	26.1	9.9-61.2
18	235	28.6	5.9-61.8	117	32.9	9.9-61.6	118	22.5	3.8-62.5
19	249	29.2	7.1-56.8	118	28.8	7.9-56.8	131	29.2	6.5-57.3
Overall	**4700***	**46.1**	**20.6-73.0**	**2285**	**44.5**	**20.2-71.0**	**2415**	**48.0**	**21.6-74.2**

*number with non-missing data
[6] Based on UK-WHO growth charts, 1990 (updated 1996)

1.5 Body Mass Index (BMI) percentiles in children and young people (<20 years)[6]
N=4730

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 are their BMI or lower; so 60% have a higher BMI.

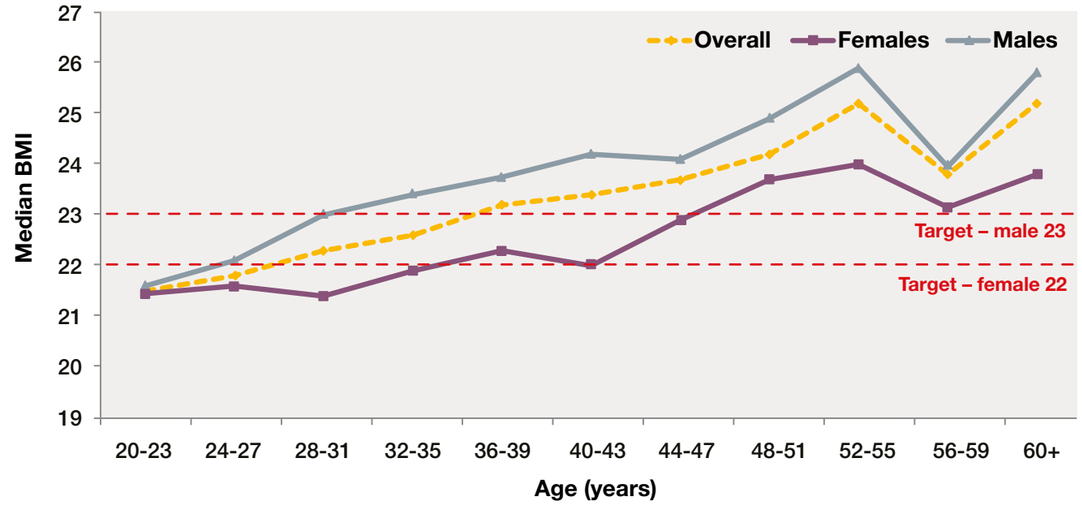


	Overall			Female			Male		
Age	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**
1	239	48.4	28.4-74.9	111	42.3	21.5-69.2	128	56.3	30.4-81.3
2	257	58.0	31.9-80.3	122	55.0	34.6-79.5	135	60.2	29.0-80.6
3	252	61.9	36.1-78.9	118	61.8	36.1-78.5	134	62.0	36.1-79.7
4	265	59.5	35.8-78.3	125	54.4	34.4-74.4	140	60.8	36.8-80.8
5	280	56.2	34.2-78.3	132	58.7	35.7-76.6	148	55.8	33.1-78.8
6	318	50.0	29.9-70.6	155	51.1	32.2-68.9	163	50.0	28.3-72.6
7	256	54.3	31.0-80.3	120	53.5	33.9-78.2	136	55.5	27.8-82.3
8	282	55.0	33.5-75.8	145	50.8	31.9-72.3	137	58.3	37.4-79.8
9	247	52.5	31.5-75.2	121	47.1	25.5-66.5	126	54.7	38.1-78.9
10	251	52.9	24.4-80.1	121	49.5	24.4-79.6	130	53.8	24.5-80.1
11	245	49.8	29.3-77.5	118	42.8	26.7-66.8	127	58.6	33.5-80.6
12	234	51.4	28.5-78.0	110	47.8	23.6-76.8	124	54.6	30.9-82.4
13	232	50.3	25.5-76.4	125	52.8	25.2-82.2	107	45.8	26.8-71.9
14	207	48.1	26.9-70.2	103	46.4	26.6-67.3	104	51.2	27.5-76.8
15	211	43.4	23.0-67.8	108	41.8	26.2-66.6	103	45.1	19.8-69.9
16	233	56.8	24.0-84.0	121	55.7	29.5-83.5	112	57.6	16.8-85.4
17	192	41.3	15.8-73.2	90	41.3	16.3-74.2	102	40.8	15.2-70.7
18	235	43.4	15.8-72.5	117	49.8	21.6-73.9	118	41.4	12.7-68.9
19	249	43.0	16.7-67.9	118	41.7	17.5-65.6	131	47.6	13.6-75.2
Overall	**4685***	**51.7**	**27.9-76.2**	**2280**	**50.2**	**27.3-74.3**	**2405**	**53.6**	**28.1-78.1**

*number with non-missing data
[6] Based on UK-WHO growth charts, 1990 (updated 1996)

1.6 Body Mass Index (BMI) in adults (20 years and over)
N=5071

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[7].



	Overall			Female			Male		
Age	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**
20-23	996	21.5	19.6-23.7	494	21.4	19.4-23.6	502	21.6	19.7-23.8
24-27	914	21.8	19.9-24.3	426	21.6	19.7-23.9	488	22.1	20.1-24.5
28-31	789	22.3	20.2-24.7	350	21.4	19.7-23.7	439	23.0	21.1-25.4
32-35	684	22.6	20.6-25.3	299	21.9	20.2-24.3	385	23.4	21.1-25.9
36-39	473	23.2	21.1-25.4	199	22.3	20.2-24.8	274	23.8	21.8-25.6
40-43	304	23.4	21.3-25.9	123	22.0	20.2-24.5	181	24.2	22.4-26.4
44-47	271	23.7	21.6-26.3	111	22.9	20.5-26.2	160	24.1	22.6-26.3
48-51	194	24.2	21.9-26.7	87	23.7	21.1-26.8	107	24.9	22.4-26.7
52-55	135	25.2	22.2-27.7	55	24.0	21.9-26.5	80	25.9	22.9-27.9
56-59	76	23.8	22.3-25.8	34	23.1	21.2-25.3	42	23.9	22.7-26.4
60+	121	25.2	22.5-28.1	57	23.8	21.6-28.1	64	25.8	23.3-27.9
Overall	**4957***	**22.6**	**20.4-25.1**	**2235**	**21.9**	**19.9-24.3**	**2722**	**23.1**	**20.9-25.6**

*number with non-missing data
[7] Stallings et al, J Am Diet Assoc. 2008;108:832-839

1.7 Education and employment in adults (16 years and over)
N=5989

The following table shows how people with CF reported their education and employment status in 2017. Please note that the groups are not mutually exclusive; someone may be a student as well as working part-time, for example.

	2013	2014	2015	2016	2017
Number of patients	5213	5592	5742	5851	5989
Number who completed questionnaire; n (%)	4346 (83.4)	4623 (82.7)	4930 (85.9)	5791 (99.0)	5937 (99.1)
Full-time employment; n (%)	1502 (28.8)	1634 (29.2)	1811 (31.5)	1887 (32.2)	1949 (32.5)
Part-time employment; n (%)	664 (12.7)	703 (12.6)	768 (13.4)	827 (14.1)	887 (14.8)
Student; n (%)	922 (17.7)	976 (17.5)	927 (16.1)	946 (16.3)	973 (16.2)
Homemaker; n (%)	232 (4.5)	258 (4.6)	264 (4.6)	242 (4.1)	246 (4.1)
Unemployed; n (%)	685 (13.1)	821 (14.7)	761 (13.3)	784 (13.4)	837 (14.0)
Disabled; n (%)	298 (5.7)	272 (4.9)	365 (6.4)	359 (6.1)	352 (5.9)
Retired; n (%)	78 (1.5)	85 (1.5)	108 (1.9)	116 (2.0)	120 (2.0)
Unknown entered; n (%)	914 (17.5)	930 (16.6)	850 (14.8)	630 (10.8)	573 (9.6)
No data recorded; n (%)	**21 (0.4)**	**39 (0.7)**	**27 (0.5)**	**-***	**-***
No. in work or study; n (%)	**3098 (71.3)**	**3242 (70.1)**	**3489 (70.8)**	**3902 (67.5)****	**4055 (68.3)**

1.8 Pregnancy

58 women with cystic fibrosis had babies in 2017

44 men with cystic fibrosis became fathers in 2017

*'No data recorded' is no longer available to select.
**The total number of people with CF in work or study increased in 2016. However, because more patients have employment data recorded overall, the percentage is lower.

Diagnosis of cystic fibrosis

1.9 Age at diagnosis and screening in children under 16 in 2017
N=3898

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.

Age at diagnosis	All patients <16; n (%)	Patients aged 10 years; n (%)	Patients aged 5 years; n (%)
Prenatal	9 (0.2)	-	-
Birth-3 months	3153 (82.2)	182 (72.5)	261 (92.9)
4-6 months	134 (3.5)	13 (5.2)	<5
7-12 months	119 (3.1)	14 (5.6)	<5
1 yr	124 (3.2)	12 (4.8)	<5
2 yrs	99 (2.6)	13 (5.2)	7 (2.5)
3 yrs	55 (1.4)	<5	<5
4 yrs	42 (1.1)	5 (2.0)	<5
5 yrs	23 (0.6)	<5	-
6 yrs	21 (0.6)	<5	-
7 yrs	10 (0.3)	0	-
8 yrs	15 (0.4)	<5	-
9 yrs	15 (0.4)	<5	-
10 yrs	6 (0.2)	<5	-
11 yrs	5 (0.1)	-	-
12 yrs	5 (0.1)	-	-
13 yrs	<5	-	-
14 yrs	-	-	-
15 yrs	-	-	-
Overall	**3837***	**251**	**281**

The median (range) age at diagnosis for patients aged under 16 in 2017 is **24** days (0-162 months).

Diagnosis in the first three months of life is more common in children aged five years in 2017 (born after the UK-wide newborn screening programme was in place) than in children aged 10 years in 2017 (born during the final year of the introduction of universal newborn screening in the UK).

A total of **172** patients born in 2017 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 2016 is higher (303) in this report than was recorded in the previous. It is likely that the 2017 figure will be updated in the next annual report in 2019.

*number with non-missing data

1.10 Age at diagnosis and screening in adults aged 16 and over in 2017 N=5989

The table below shows the age at diagnosis for people aged 16 and over in 2017. People aged 16 or over in 2017 were born when newborn screening was only done in a few areas of the UK, before it became universal in mid-2007.

Age at diagnosis	n (%)
Birth-3 months	2394 (40.3)
4-6 months	550 (9.3)
7-12 months	421 (7.1)
1 yr	429 (7.2)
2 yrs	294 (4.9)
3 yrs	228 (3.8)
4 yrs	179 (3.0)
5 yrs	91 (1.5)
6 yrs	74 (1.2)
7 yrs	58 (1.0)
8 yrs	66 (1.1)
9 yrs	52 (0.9)
10 yrs	43 (0.7)
11 yrs	43 (0.7)
12 yrs	45 (0.8)
13 yrs	52 (0.9)
14 yrs	37 (0.6)
15 yrs	46 (0.8)
16-19 yrs	145 (2.4)
20-23 yrs	110 (1.8)
24-27 yrs	100 (1.7)
28-31 yrs	94 (1.6)
32-35 yrs	101 (1.7)
36-39 yrs	76 (1.3)
40-43 yrs	66 (1.1)
44-47 yrs	39 (0.7)
48-51 yrs	28 (0.5)
52-55 yrs	27 (0.5)
56-59 yrs	23 (0.4)
60+ yrs	39 (0.6)
Overall	**5953***

Overall, **848** (14.2%) adults with CF in the Registry in 2017 were diagnosed at age 16 or over.

In 2017, **16** people aged 16 or over were newly diagnosed with cystic fibrosis

*number with non-missing data

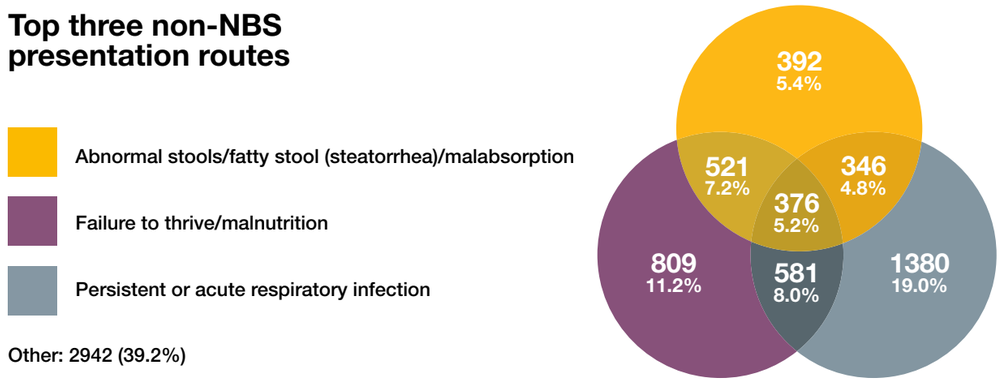
1.11 Mode of presentation

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	9887	8942	848
Number diagnosed by newborn screening	2640	2634	0
Total non-NBS	**7247**	**6308**	**848**

Mode of presentation (excluding newborn screening)	All patients (n=7247)		Age <16 at diagnosis* (n=6308)		Age ≥16 at diagnosis* (n=848)	
Persistant or acute respiratory infection	2683	37.0%	2188	34.7%	493	58.1%
Failure to thrive/malnutrition	2287	31.6%	2258	35.8%	27	3.2%
Abnormal stools/fatty stool (steatorrhea)/malabsorption	1635	22.6%	1579	25.0%	55	6.5%
Meconium Ileus	1346	18.6%	1334	21.1%	5	0.6%
Family history	913	12.6%	777	12.3%	135	15.9%
Genotype	602	8.3%	432	6.8%	165	19.5%
Unknown	299	4.1%	238	3.8%	53	6.3%
Rectal prolapse	-	-	245	3.9%	<5	-
Nasal polyps	155	2.1%	83	1.3%	72	8.5%
Electrolyte imbalance	74	1.0%	68	1.1%	6	0.7%
Prenatal	-	-	53	0.8%	<5	-
Bronchiectasis	-	-	<5	-	39	4.6%
Liver disease	-	-	38	0.6%	<5	-
Fertility	-	-	<5	-	21	2.5%
Pancreatitis	-	-	<5	-	8	0.9%
Oedema	8	0.1%	8	0.1%	0	0.0%

Top three non-NBS presentation routes



Other: 2942 (39.2%)

*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.

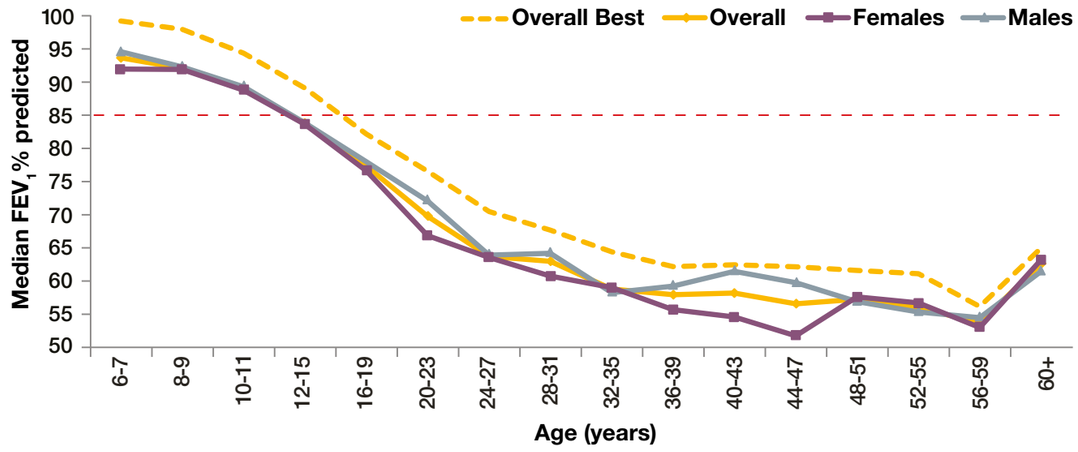
The FEV_1% predicted values shown in this report are calculated using an equation called Global Lungs Initiative, or 'GLI'[8]

[8] Quanjer PH et al. Eur respir J. 2012 Dec; 40(6):1324-1343

1.12 FEV_1% predicted (GLI equations) in patients aged six years and older who have not had a lung transplant N=8168

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 the 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	526	93.6	81.4-101.9	249	91.9	80.3-100.8	277	94.5	82.7-103.0
8-9	511	92.0	82.4-101.1	256	91.9	81.8-100.8	255	92.2	83.1-101.9
10-11	488	89.0	77.3-98.2	235	88.7	75.7-97.8	253	89.2	79.8-98.3
12-15	862	83.7	71.6-93.7	438	83.6	70.7-94.2	424	83.8	72.0-93.4
16-19	887	77.3	60.3-89.4	440	76.6	58.5-88.3	447	78.0	63.1-90.5
20-23	945	69.8	50.6-86.6	466	66.9	49.4-85.3	479	72.1	53.1-87.6
24-27	860	63.7	46.2-82.0	402	63.6	48.0-80.6	458	63.9	45.5-83.1
28-31	725	63.0	42.7-80.3	315	60.8	42.4-79.0	410	64.2	44.2-81.4
32-35	609	58.8	38.6-77.4	268	59.1	39.0-76.8	341	58.3	38.2-78.3
36-39	436	58.0	41.0-76.8	179	55.7	41.7-76.4	257	59.3	40.5-77.1
40-43	276	58.2	41.9-78.3	109	54.6	41.0-74.6	167	61.5	43.3-81.2
44-47	237	56.6	39.1-77.0	97	51.8	40.4-71.6	140	59.8	37.8-79.7
48-51	172	57.3	38.9-79.9	79	57.6	40.2-76.8	93	56.9	37.1-82.9
52-55	119	56.0	42.1-77.1	51	56.7	39.5-74.3	68	55.4	42.2-78.2
56-59	68	53.8	38.4-72.3	30	53.0	46.1-69.0	38	54.5	33.2-77.7
60+	114	62.6	42.0-79.1	54	63.2	45.9-78.1	60	61.5	39.6-79.5
<16	2387	89.0	76.9-98.4	1178	88.1	76.2-98.0	1209	89.7	77.7-98.9
≥16	5448	65.2	45.5-83.1	2490	63.7	45.4-81.5	2958	66.5	45.6-84.3
<18	2802	87.6	75.4-97.6	1386	86.6	74.5-97.2	1416	88.2	75.8-98.2
≥18	5033	63.7	44.2-82.1	2282	62.4	44.2-80.5	2751	65.1	44.2-83.3
Overall	**7835***	**74.6**	**53.1-90.4**	**3668**	**74.2**	**52.5-89.8**	**4167**	**74.9**	**53.6-91.0**

*number with non-missing data

1.13 Best FEV_1% predicted (GLI equations) in patients aged six years and older who have not had a lung transplant
N=8168

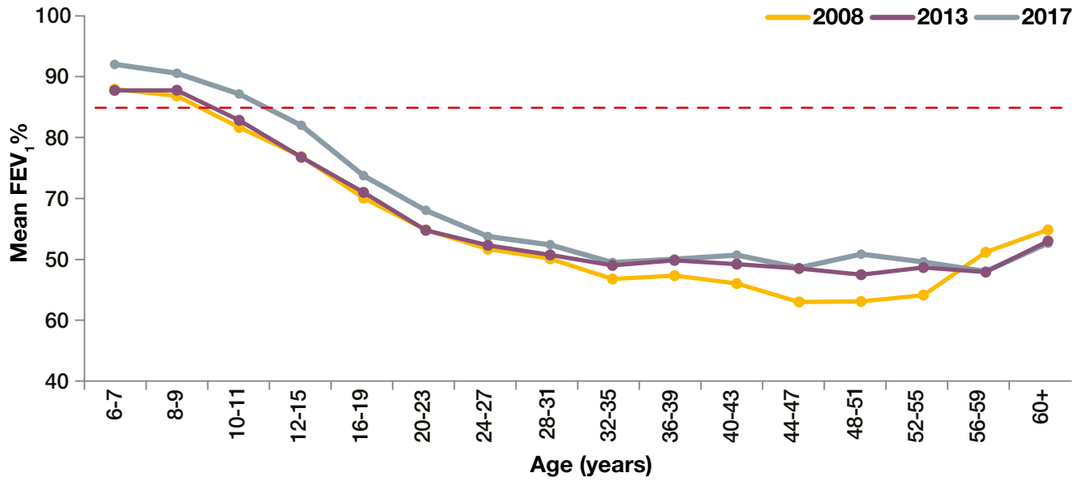
	Overall			Female			Male		
Age (yrs)	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**
6-7	546	99.1	89.8-106.5	259	98.1	89.9-105.6	287	100.4	89.6-107.3
8-9	517	97.9	88.7-106.2	259	96.6	88.5-106.2	258	98.9	89.1-105.9
10-11	492	94.3	85.4-102.0	238	93.4	82.1-101.1	254	95.4	87.2-102.3
12-15	875	89.0	78.0-98.3	443	89.4	76.1-98.1	432	88.8	79.0-98.3
16-19	902	82.1	67.2-93.6	446	81.5	65.2-92.4	456	83.3	69.7-94.4
20-23	963	76.6	57.8-91.3	474	74.2	55.8-89.9	489	79.0	59.4-92.5
24-27	873	70.5	51.5-86.5	405	70.5	53.5-86.3	468	70.4	49.9-86.7
28-31	735	67.7	47.4-84.9	320	65.5	45.8-84.5	415	70.0	49.5-85.0
32-35	622	64.4	44.6-82.2	272	64.4	45.5-80.0	350	64.4	43.7-83.7
36-39	439	62.2	45.7-81.5	181	62.7	45.7-83.0	258	61.3	45.7-81.2
40-43	279	62.5	45.8-81.6	111	58.3	44.5-81.1	168	64.9	47.8-83.0
44-47	239	62.2	44.6-79.2	98	61.4	44.8-74.0	141	63.1	43.5-82.0
48-51	174	61.6	42.4-83.3	81	61.9	45.9-81.8	93	60.5	42.0-87.1
52-55	119	61.1	44.8-81.6	51	61.1	48.3-81.3	68	60.3	43.5-85.4
56-59	68	56.2	44.0-76.9	30	56.9	47.7-75.6	38	55.0	34.5-80.5
60+	115	65.0	45.1-81.4	54	64.4	49.6-81.8	61	65.0	40.7-80.2
<16	2430	94.4	84.0-103.3	1199	94.1	83.2-102.6	1231	94.9	85.0-103.9
≥16	5528	71.0	50.5-87.7	2523	69.9	50.7-86.9	3005	71.5	50.3-88.5
<18	2852	93.2	82.0-102.4	1410	92.8	81.3-101.7	1442	93.5	82.8-103.1
≥18	5106	69.4	49.3-86.8	2312	68.1	49.5-86.0	2794	70.4	49.2-87.6
Overall	**7958***	**80.3**	**59.2-95.0**	**3722**	**80.0**	**59.3-94.6**	**4236**	**80.6**	**59.1-95.2**

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

*number with non-missing data

1.14 FEV_1% predicted (GLI equations) over time in patients six years and older who have not had a lung transplant
N=8168 in 2017, N=7268 in 2013, N=5049 in 2008*

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 2017 compares to Registry data from 2008 and 2013. 2008 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.

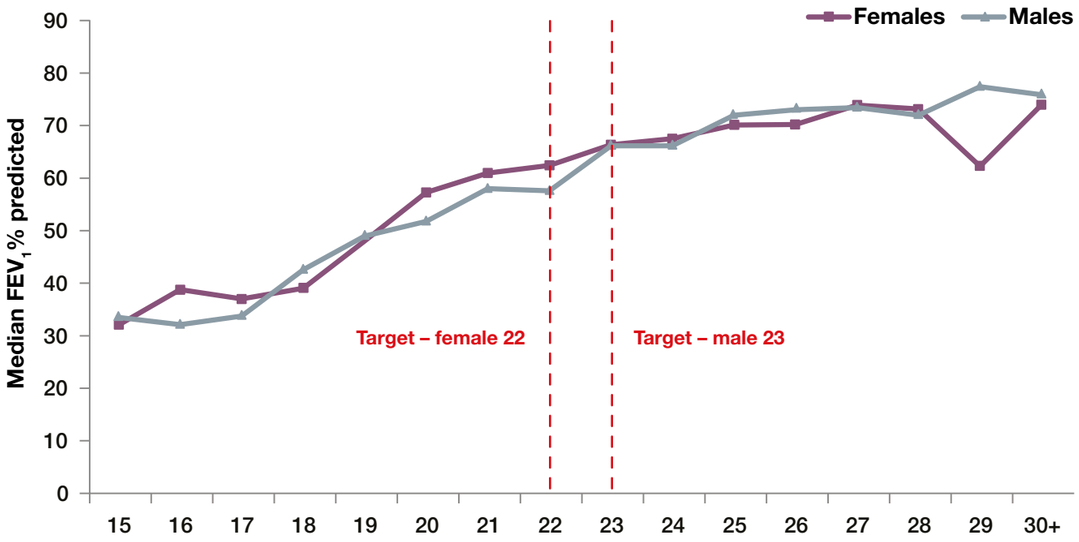


Age (years)	2008 mean FEV_1%	2013 mean FEV_1%	2017 mean FEV_1%	p-values (t-test)**
6-7	87.9	87.7	92.0	<0.001
8-9	86.8	87.8	90.6	0.006
10-11	81.7	82.8	87.2	<0.001
12-15	76.9	76.7	82.0	<0.001
16-19	70.0	71.0	73.7	0.008
20-23	64.9	64.8	68.0	0.003
24-27	61.7	62.3	63.7	0.214
28-31	60.1	60.7	62.4	0.194
32-35	56.8	59.0	59.5	0.752
36-39	57.3	59.8	60.0	0.900
40-43	56.0	59.2	60.7	0.471
44-47	53.0	58.5	58.6	0.964
48-51	53.1	57.5	60.9	0.252
52-55	54.1	58.6	59.6	0.812
56-59	61.1	57.9	58.0	0.981
60+	64.8	63.0	62.6	0.931
<16	81.5	82.2	87.1	N/A
≥16	62.7	63.3	64.5	N/A
<18	79.8	80.4	85.5	N/A
≥18	61.6	62.3	63.5	N/A

*Due to missing data, means are calculated from a population of 7835 in 2017, 6825 in 2013 and 4613 in 2008.
** t-test comparing 2017 with 2013

1.15 FEV_1% predicted (GLI equations) and BMI in people aged 20 years and over who have not had a transplant
N=4764*

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.

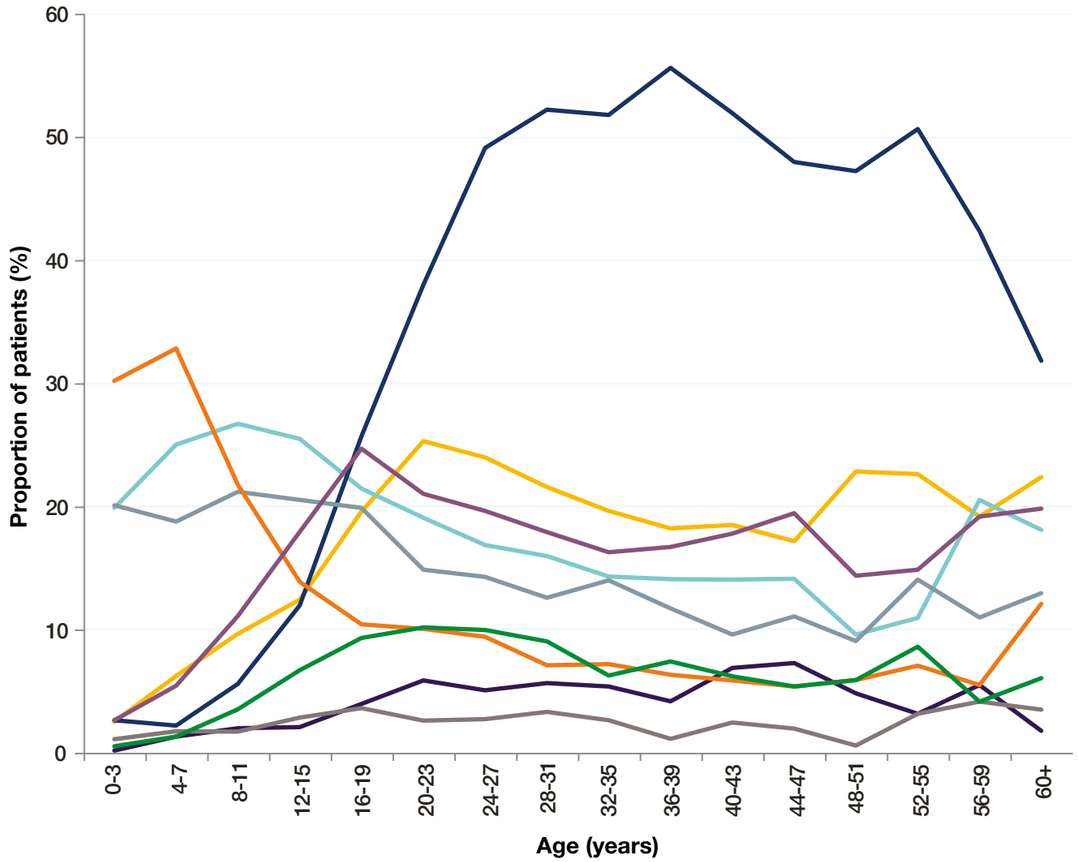


*Due to missing data, medians are calculated from a population of 4553.
Each point represents the median FEV_1 % predicted of patients for each given BMI value. Due to the wide range of BMIs in this population with a value of 30 of more, these are grouped into one.

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.

1.16 Lung infections in 2017
N=9887



1.17 Lung infections in 2017
<16 years N=3898, ≥16 years N=5989

	Paediatric Age Range (Years)				Overall
	0-3	4-7	8-11	12-15	Paediatric (<16 years)
Number in age range	854	1124	1031	889	3898
Number who had culture taken	850	1113	1028	886	3877
Chronic S. *aureus* n (%)	21 (2.5)	69 (6.2)	99 (9.6)	110 (12.4)	299 (7.7)
Intermittent S. *aureus* n (%)	169 (19.9)	279 (25.1)	275 (26.8)	226 (25.5)	949 (24.5)
Chronic P. *aeruginosa* n (%)	22 (2.6)	24 (2.2)	57 (5.5)	106 (12.0)	209 (5.4)
Intermittent P. *aeruginosa* n (%)	171 (20.1)	209 (18.8)	218 (21.2)	182 (20.5)	780 (20.1)
B. *cepacia complex* n (%)	<5	14 (1.3)	20 (1.9)	18 (2.0)	53 (1.4)
B. *cenocepacia* n (%)	<5	<5	5 (0.5)	<5	12 (0.3)
B. *multivorans* n (%)	<5	5 (0.4)	6 (0.6)	6 (0.7)	17 (0.4)
B. *cepacia* (other) n (%)	<5	<5	6 (0.6)	5 (0.6)	13 (0.3)
MRSA n (%)	9 (1.1)	19 (1.7)	17 (1.7)	25 (2.8)	70 (1.8)
H. *influenzae* n (%)	257 (30.2)	366 (32.9)	224 (21.8)	123 (13.9)	970 (25.0)
NTM n (%)	<5	14 (1.3)	36 (3.5)	59 (6.7)	113 (2.9)
Aspergillus n (%)	22 (2.6)	60 (5.4)	114 (11.1)	159 (17.9)	355 (9.2)

Infections in this table reflect bugs grown in the 12 months prior to the 2017 annual review.

The UK CF Registry definition of 'chronic' is three or more isolates in the last 12 months.

	Adult Age Range (Years)						Overall
	16-19	20-23	24-27	28-31	32-35	36-39	Adults (≥16 years)
Number in age range	918	1010	942	810	698	483	5989
Number who had culture taken	894	975	896	764	657	461	5706
Chronic S. *aureus* n (%)	175 (19.6)	247 (25.3)	215 (24.0)	165 (21.6)	129 (19.6)	84 (18.2)	1226 (21.5)
Intermittent S. *aureus* n (%)	192 (21.5)	186 (19.1)	151 (16.9)	122 (16.0)	94 (14.3)	65 (14.1)	956 (16.8)
Chronic P. *aeruginosa* n (%)	230 (25.7)	371 (38.1)	441 (49.2)	400 (52.4)	341 (51.9)	257 (55.7)	2540 (44.5)
Intermittent P. *aeruginosa* n (%)	178 (19.9)	145 (14.9)	128 (14.3)	96 (12.6)	92 (14.0)	54 (11.7)	808 (14.2)
B. *cepacia complex* n (%)	35 (3.9)	57 (5.8)	45 (5.0)	43 (5.6)	35 (5.3)	19 (4.1)	292 (5.1)
B. *cenocepacia* n (%)	7 (0.8)	11 (1.1)	14 (1.6)	11 (1.4)	8 (1.2)	6 (1.3)	78 (1.4)
B. *multivorans* n (%)	17 (1.9)	34 (3.5)	23 (2.6)	25 (3.3)	19 (2.9)	11 (2.4)	153 (2.7)
B. *cepacia* (other) n (%)	7 (0.8)	9 (0.9)	7 (0.8)	5 (0.7)	7 (1.1)	<5	45 (0.8)
MRSA n (%)	32 (3.6)	25 (2.6)	24 (2.7)	25 (3.3)	17 (2.6)	5 (1.1)	152 (2.7)
H. *influenzae* n (%)	93 (10.4)	98 (10.1)	84 (9.4)	54 (7.1)	47 (7.2)	29 (6.3)	474 (8.3)
NTM n (%)	83 (9.3)	99 (10.2)	89 (9.9)	69 (9.0)	41 (6.2)	34 (7.4)	479 (8.4)
Aspergillus n (%)	221 (24.7)	205 (21.0)	176 (19.6)	137 (17.9)	107 (16.3)	77 (16.7)	1109 (19.4)

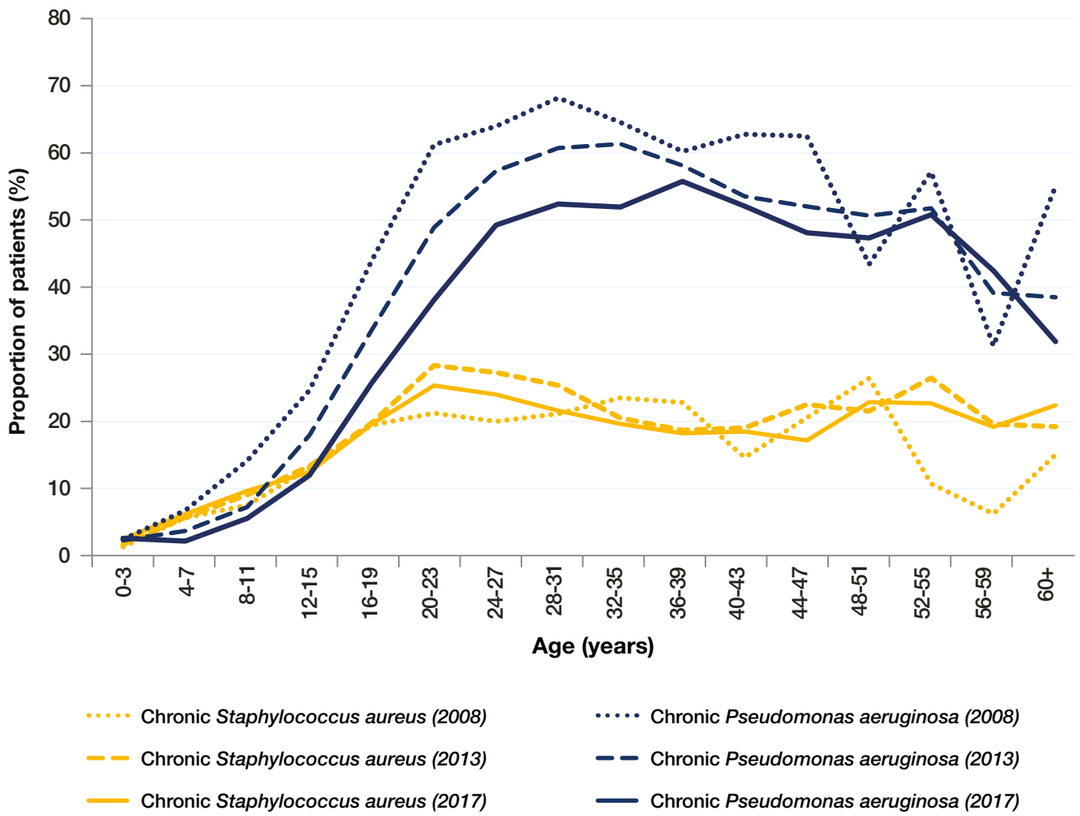
	Adult Age Range (Years)						Overall
	40-43	44-47	48-51	52-55	56-59	60+	Adults (≥16 years)
Number in age range	311	280	201	138	76	122	5989
Number who had culture taken	292	262	188	128	73	116	5706
Chronic S. *aureus* n (%)	54 (18.5)	45 (17.2)	43 (22.9)	29 (22.7)	14 (19.2)	26 (22.4)	1226 (21.5)
Intermittent S. *aureus* n (%)	41 (14.0)	37 (14.1)	18 (9.6)	14 (10.9)	15 (20.5)	21 (18.1)	956 (16.8)
Chronic P. *aeruginosa* n (%)	152 (52.1)	126 (48.1)	89 (47.3)	65 (50.8)	31 (42.5)	37 (31.9)	2540 (44.5)
Intermittent P. *aeruginosa* n (%)	28 (9.6)	29 (11.1)	17 (9.0)	18 (14.1)	8 (11.0)	15 (12.9)	808 (14.2)
B. *cepacia complex* n (%)	20 (6.8)	19 (7.3)	9 (4.8)	<5	<5	<5	292 (5.1)
B. *cenocepacia* n (%)	8 (2.7)	6 (2.3)	5 (2.7)	<5	<5	<5	78 (1.4)
B. *multivorans* n (%)	9 (3.1)	7 (2.7)	<5	<5	<5	<5	153 (2.7)
B. *cepacia* (other) n (%)	<5	<5	<5	<5	<5	<5	45 (0.8)
MRSA n (%)	7 (2.4)	5 (1.9)	<5	<5	<5	<5	152 (2.7)
H. *influenzae* n (%)	17 (5.8)	14 (5.3)	11 (5.9)	9 (7.0)	<5	14 (12.1)	474 (8.3)
NTM n (%)	18 (6.2)	14 (5.3)	11 (5.9)	11 (8.6)	<5	7 (6.0)	479 (8.4)
Aspergillus n (%)	52 (17.8)	51 (19.5)	27 (14.4)	19 (14.8)	14 (19.2)	23 (19.8)	1109 (19.4)

1.18 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.

	2015 (n=9587)	2016 (n=9695)	2017 (n=9887)
NTM Prevalence (%)	536 (5.6%)	567 (5.8%)	592 (6.0%)
On NTM treatment in the given year (% of NTM prevalence in given year)	300 (56.0%)	333 (58.7%)	352 (59.5%)
NTM Incidence	222	228	246
M. *abscessus* prevalence	321	337	376
M. *abscessus* incidence	-*	110	136

1.19 Lung infections over time
N=6082 in 2008, N=9052 in 2013, N=9887 in 2017



M. abscessus incidence cannot be evaluated prior to 2016 as enhanced NTM reporting was not available before 2014.

Chronic *Staphylococcus aureus*				
Age (years)	**2008 (%)**	**2013 (%)**	**2017 (%)**	**p-value***
0-3	1.3	1.7	2.5	0.235
4-7	5.6	5.7	6.2	0.640
8-11	7.5	9.1	9.6	0.628
12-15	13.0	13.3	12.4	0.590
16-19	19.5	19.8	19.6	0.791
20-23	21.2	28.3	25.3	0.067
24-27	20.0	27.3	24.0	0.044
28-31	21.1	25.3	21.6	0.039
32-35	23.5	20.5	19.6	0.534
36-39	22.8	18.7	18.2	0.844
40-43	14.6	19.0	18.5	0.762
44-47	20.5	22.5	17.2	0.123
48-51	26.4	21.5	22.9	0.901
52-55	10.7	26.4	22.7	0.398
56-59	6.3	19.6	19.2	0.994
60+	15.0	19.2	22.4	0.722
<16 years	7.3	7.6	7.7	n/a
≥16 years	20.4	23.6	21.5	n/a
<18 years	8.6	8.9	8.6	n/a
≥18 years	20.7	24.1	21.9	n/a

Chronic *Pseudomonas aeruginosa*				
Age (years)	**2008 (%)**	**2013 (%)**	**2017 (%)**	**p-value***
0-3	2.4	2.3	2.6	0.641
4-7	6.7	3.7	2.2	0.033
8-11	14.2	7.2	5.5	0.147
12-15	24.6	17.9	12.0	<0.001
16-19	43.9	33.6	25.7	<0.001
20-23	61.2	48.9	38.1	<0.001
24-27	64.0	57.3	49.2	<0.001
28-31	68.2	60.7	52.4	<0.001
32-35	64.5	61.3	51.9	<0.001
36-39	60.2	58.1	55.7	0.464
40-43	62.8	53.5	52.1	0.514
44-47	62.5	52.0	48.1	0.297
48-51	43.4	50.6	47.3	0.361
52-55	57.1	51.7	50.8	0.611
56-59	31.3	39.1	42.5	0.650
60+	55.0	38.5	31.9	0.235
<16 years	12.8	7.9	5.4	n/a
≥16 years	58.4	51.0	44.5	n/a
<18 years	15.2	10.6	7.1	n/a
≥18 years	61.3	53.4	46.3	n/a

*The proportion of people with each infection within each age group was compared between 2013 and 2017. If the p-value is less than 0.05 then the difference in the proportions is statistically significant.

Complications

1.20 Prevalence of complications

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

	2016			2017*		
	Overall (n=9695)	<16 years (n=3844)	≥16 years (n=5851)	Overall (n=9887)	<16 years (n=3898)	≥16 years (N=5989)
	N (%)					
Respiratory related						
Nasal polyps requiring surgery	379 (3.9)	96 (2.5)	283 (4.8)	333 (3.4)	90 (2.3)	243 (4.1)
Sinus disease	949 (9.8)	62 (1.6)	887 (15.2)	694 (7.0)	42 (1.1)	652 (10.9)
Asthma	1306 (13.5)	437 (11.4)	869 (14.9)	777 (7.9)	228 (5.8)	549 (9.2)
ABPA	646 (6.7)	145 (3.8)	501 (8.6)	664 (6.7)	129 (3.3)	535 (8.9)
Any haemoptysis	697 (7.2)	35 (0.9)	662 (11.3)	890 (9.0)	40 (1.0)	850 (14.2)
Massive haemoptysis	Not reported in 2016			31 (0.3)	0	31 (0.5)
Pneumothorax requiring chest tube	33 (0.3)	5 (0.1)	28 (0.5)	-	<5	31 (0.5)
Pancreas & Hepatobiliary disease						
Raised liver enzymes	962 (9.9)	234 (6.1)	728 (12.4)	835 (8.4)	213 (5.5)	622 (10.4)
Liver disease	1332 (13.7)	289 (7.5)	1043 (17.8)	1278 (12.9)	307 (7.9)	971 (16.2)
Cirrhosis with no portal hypertension	119 (1.2)	27 (0.7)	92 (1.6)	124 (1.3)	17 (0.4)	107 (1.8)
Cirrhosis with portal hypertension	146 (1.5)	24 (0.6)	122 (2.1)	162 (1.6)	27 (0.7)	135 (2.3)
Gall bladder disease requiring surgery	99 (1.0)	9 (0.2)	90 (1.5)	140 (1.4)	26 (0.7)	114 (1.9)
Pancreatitis	43 (0.4)	5 (0.1)	38 (0.6)	42 (0.4)	6 (0.2)	36 (0.6)
Upper Gastrointestinal						
GERD	1118 (11.5)	178 (4.6)	940 (16.1)	1414 (14.3)	236 (6.1)	1178 (19.7)
Peptic ulcer	<5	<5	<5	7 (0.1)	0	7 (0.1)
GI bleed (varices as source)	-	<5	5 (0.1)	-	<5	8 (0.1)
GI bleed (non varices as source)	-	<5	9 (0.2)	15 (0.2)	5 (0.1)	10 (0.2)
Lower Gastrointestinal						
Intestinal obstruction	31 (0.3)	16 (0.4)	15 (.3)	48 (0.5)	21 (0.5)	27 (0.5)
DIOS	444 (4.6)	76 (2)	368 (6.3)	528 (5.3)	92 (2.4)	436 (7.3)
Fibrosing colonopathy/colonic stricture	<5	<5	<5	5 (0.1)	<5	<5
Rectal prolapse	12 (0.1)	7 (0.2)	5 (0.1)	15 (0.2)	8 (0.2)	7 (0.1)
Renal						
Kidney stones	77 (0.8)	7 (0.2)	70 (1.2)	97 (1.0)	6 (0.2)	91 (1.5)
Renal failure	-	<5	75 (1.3)	-	<5	85 (1.4)
Musculo-Skeletal						
Arthritis	179 (1.8)	5 (0.1)	174 (3.0)	118 (1.2)	9 (0.2)	109 (1.8)
Arthropathy	526 (5.4)	21 (0.5)	505 (8.6)	328 (3.3)	14 (0.4)	314 (5.2)
Bone fracture	26 (0.3)	8 (0.2)	18 (0.3)	29 (0.3)	5 (0.1)	24 (0.4)
Osteopenia	1339 (13.8)	41 (1.1)	1298 (22.2)	1131 (11.4)	26 (0.7)	1105 (18.5)
Osteoporosis	481 (5.0)	7 (0.2)	474 (8.1)	448 (4.5)	10 (0.3)	438 (7.3)
Other						
Cancer confirmed by histology	24 (0.2)	0 (0)	24 (0.4)	34 (0.3)	<5	33 (0.6)
Port inserted or replaced	293 (3.0)	98 (2.5)	195 (3.3)	236 (2.4)	107 (2.7)	129 (2.2)
Depression	500 (5.2)	13 (0.3)	487 (8.3)	428 (4.3)	12 (0.3)	416 (6.9)
Hearing loss	275 (2.8)	33 (0.9)	242 (4.1)	220 (2.2)	19 (0.5)	201 (3.4)
Hypertension	100 (1.0)	0 (0)	100 (1.7)	106 (1.1)	<5	104 (1.7)

*Please note that in 2017 the data entry pathway for complications changed, which has resulted in a drop in prevelance for several complications. Results in 2017 should be interpreted with caution until 2018, when we will confirm whether this is a true decrease.

1.21 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.

	2016			2017		
	Overall (n=9695)	<16 years (n=3844)	≥16 years (n=5851)	Overall (n=9887)	<16 years (n=3989)	≥16 years (n=5989)
ABPA; n (%)	188 (1.9)	68 (1.8)	120 (2.1)	165 (1.7)	64 (1.6)	101 (1.7)
Cirrhosis - no portal hypertension; n (%)	49 (0.5)	13 (0.1)	36 (0.6)	58 (0.6)	7 (0.2)	51 (0.9)
Cirrhosis - with portal hypertension; n (%)	38 (0.4)	7 (0.2)	31 (0.5)	43 (0.4)	14 (0.4)	29 (0.5)
Cancer confirmed by histology; n (%)	16 (0.2)	0 (0)	16 (0.3)	18 (0.2)	<5	17 (0.3)

1.22 CF-related diabetes
N=7378

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=7378)	10-15 years (n=1389)	≥16 years (n=5989)
On CFRD treatment; n (%)	2195 (29.8)	169 (12.2)	2026 (33.8)
CFRD screening; n (%)			
Yes	3906 (52.9)	983 (70.8)	2923 (48.8)
No	1421 (19.3)	272 (19.6)	1149 (19.2)
Existing CFRD diagnosis	1823 (24.7)	94 (6.8)	1729 (28.9)
Unknown	106 (1.4)	38 (2.7)	68 (1.1)

Antibiotics

1.23 Intravenous (IV) antibiotics

N=9887

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	854	43 (5.0)	12 (7-13)	229 (26.8)	13 (7-13)	233 (27.3)	14 (8-22)
4-7	1124	102 (9.1)	12 (8-23)	301 (26.8)	14 (8-23)	317 (28.2)	14 (13-28)
8-11	1031	137 (13.3)	15 (10-37)	324 (31.4)	14 (10-37)	348 (33.8)	22 (14-43)
12-15	889	183 (20.6)	19 (11-31)	373 (42.0)	14 (11-31)	413 (46.5)	28 (14-42)
16-19	918	253 (27.6)	20 (12-34)	407 (44.3)	19 (12-34)	474 (51.6)	28 (14-48)
20-23	1010	312 (30.9)	20 (14-34)	468 (46.3)	17 (14-34)	566 (56.0)	28 (14-50)
24-27	942	333 (35.4)	22 (14-39)	425 (45.1)	17 (14-39)	526 (55.8)	28 (14-54)
28-31	810	292 (36.0)	23 (13-39)	357 (44.1)	16 (13-39)	458 (56.5)	29 (14-53)
32-35	698	232 (33.2)	21 (14-42)	283 (40.5)	15 (14-42)	369 (52.9)	28 (14-52)
36-39	483	170 (35.2)	20 (14-35)	186 (38.5)	14 (14-35)	247 (51.1)	28 (14-49)
40-43	311	99 (31.8)	21 (12-36)	120 (38.6)	15 (12-36)	155 (49.8)	28 (14-45)
44-47	280	82 (29.3)	20 (11-35)	99 (35.4)	14 (11-35)	127 (45.4)	27 (14-48)
48-51	201	42 (20.9)	23 (14-38)	62 (30.8)	21 (14-38)	80 (39.8)	32 (16-50)
52-55	138	34 (24.6)	16 (12-51)	51 (37.0)	14 (12-51)	62 (44.9)	18 (14-41)
56-59	76	13 (17.1)	14 (10-22)	25 (32.9)	18 (10-22)	30 (39.5)	24 (14-39)
60+	122	26 (21.3)	14 (10-28)	38 (31.1)	11 (10-28)	45 (36.9)	24 (14-39)
<16	3898	465 (11.9)	14 (10-28)	1227 (31.5)	14 (10-28)	1311 (33.6)	16 (14-39)
≥16	5989	1888 (31.5)	21 (13-37)	2521 (42.1)	16 (13-37)	3139 (52.4)	28 (14-51)
<18	4329	559 (12.9)	15 (10-30)	1417 (32.7)	14 (10-30)	1526 (35.3)	19 (14-41)
≥18	5558	1794 (32.3)	21 (13-37)	2331 (41.9)	16 (13-37)	2924 (52.6)	28 (14-51)
Overall	**9887**	**2353 (23.8)**	**20 (13-35)**	**3748 (37.9)**	**14 (13-35)**	**4450 (45.0)**	**27 (14-46)**

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

	2008			2013			2017		
	Overall	**<16 years**	**≥16 years**	**Overall**	**<16 years**	**≥16 years**	**Overall**	**<16 years**	**≥16 years**
Patients with chronic *P. aeruginosa*	2098	299	1799	2960	329	2631	2749	209	2540
Tobramycin solution; n (%)	412 (19.6)	48 (16.1)	364 (20.2)	929 (31.4)	103 (31.3)	826 (31.4)	626 (22.8)	72 (34.4)	554 (21.8)
Other aminoglycoside; n (%)	43 (2.0)	5 (1.7)	38 (2.1)	108 (3.6)	13 (4.0)	95 (3.6)	52 (1.9)	<5	51 (2.0)
Colistin; n (%)	914 (43.6)	174 (58.2)	740 (41.1)	1173 (39.6)	176 (53.5)	997 (37.9)	680 (24.7)	79 (37.8)	601 (23.7)
Promixin; n (%)	490 (23.4)	73 (24.4)	417 (23.2)	881 (29.8)	140 (42.6)	741 (28.2)	859 (31.2)	98 (46.9)	761 (30.0)
Aztreonam; n (%)	-	-	-	201 (6.8)	<5	199 (7.6)	628 (22.8)	10 (4.8)	618 (24.3)
Colistimethate (DPI); n (%)	-	-	-	-	-	-	531 (19.3)	15 (7.2)	516 (20.3)
Tobramycin Inhalation Powder; n (%)	-	-	-	-	-	-	782 (28.4)	26 (12.4)	756 (29.8)
At least one of the above; n (%)	1597 (76.1)	257 (86.0)	1340 (74.5)	2368 (80.0)	302 (91.8)	2066 (78.5)	2469 (89.8)	191 (91.4)	2278 (89.7)

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.

1.25 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*.

		Patients with chronic *P. aeruginosa*; n (%)	Patients without chronic *P. aeruginosa*; n (%)
2008	**Overall (n=1958)**	1246 (63.6)	712 (36.4)
	0-3 years (n=15)	<5	13 (86.7)
	4-15 years (n=363)	105 (28.9)	258 (71.1)
	≥ 16 years (n=1580)	1139 (72.1)	441 (27.9)
2013	**Overall (n=3619)**	2022 (55.9)	1597 (44.1)
	0-3 years (n=27)	<5	25 (92.6)
	4-15 years (n=620)	141 (22.7)	479 (77.3)
	≥ 16 years (n=2972)	1879 (63.2)	1093 (36.8)
2017	**Overall (n=4103)**	1922 (46.8)	2181 (53.2)
	0-3 years (n=37)	<5	34 (91.9)
	4-15 years (n=676)	89 (13.2)	587 (86.8)
	≥16 years (n=3390)	1830 (54.0)	1560 (46.0)

*In 2013, this includes Aztreonam. In 2017 it includes Aztreonam, Colistimethate and Tobramycin Inhalation Powder.

1.26 Prophylactic flucloxacillin use

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

Age	Total patients	Patients on prophylactic flucloxacillin; n (%)
0-3	854	531 (62.2)
4-7	1124	310 (27.6)
8-11	1031	283 (27.4)
12-15	889	225 (25.3)
16-19	918	166 (18.1)
20-23	1010	97 (9.6)
24-27	942	81 (8.6)
28-31	810	53 (6.5)
32-35	698	50 (7.2)
36-39	483	40 (8.3)
40-43	311	23 (7.4)
44-47	280	17 (6.1)
48-51	201	15 (7.5)
52-55	138	5 (3.6)
56-59	76	7 (9.2)
60+	122	6 (4.9)
<16 years	3898	1349 (34.6)
≥16 years	5989	560 (9.4)
<18 years	4329	1439 (33.2)
≥18 years	5558	470 (8.5)
Overall	**9887**	**1909 (19.3)**

Muco-active therapies

1.27 Mannitol

Age	Total patients	Patients on Mannitol; n (%)
0-3	854	0
4-7	1124	0
8-11	1031	0
12-15	889	<5
16-19	918	21 (2.3)
20-23	1010	60 (5.9)
24-27	942	63 (6.7)
28-31	810	58 (7.2)
32-35	698	47 (6.7)
36-39	483	33 (6.8)
40-43	311	22 (7.1)
44-47	280	14 (5.0)
48-51	201	5 (2.5)
52-55	138	<5
56-59	76	<5
60+	122	<5
<16 years	3898	<5
≥16 years	5989	330 (5.5)
<18 years	4329	6 (0.1)
≥18 years	5558	325 (5.8)
Overall	**9887**	**331 (3.3)**

1.28 DNase

	2008		2013		2017	
Age	Total patients	Patients on DNase; n (%)	Total patients	Patients on DNase; n (%)	Total patients	Patients on DNase; n (%)
0-3	605	46 (7.6)	981	100 (10.2)	854	128 (15.0)
4-7	621	125 (20.1)	1004	332 (33.1)	1124	579 (51.5)
8-11	663	227 (34.2)	899	496 (55.2)	1031	790 (76.6)
12-15	773	359 (46.4)	955	627 (65.7)	889	725 (81.6)
16-19	762	377 (49.5)	1005	635 (63.2)	918	742 (80.8)
20-23	725	319 (44.0)	994	625 (62.9)	1010	728 (72.1)
24-27	605	288 (47.6)	836	537 (64.2)	942	641 (68.0)
28-31	419	182 (43.4)	703	413 (58.7)	810	536 (66.2)
32-35	260	108 (41.5)	503	283 (56.3)	698	431 (61.7)
36-39	237	83 (35.0)	315	157 (49.8)	483	292 (60.5)
40-43	165	58 (35.2)	294	141 (48.0)	311	179 (57.6)
44-47	120	56 (46.7)	213	102 (47.9)	280	153 (54.6)
48-51	59	18 (30.5)	152	79 (52.0)	201	105 (52.2)
52-55	29	9 (31.0)	76	32 (42.1)	138	68 (49.3)
56-59	17	<5	48	24 (50.0)	76	41 (53.9)
60+	22	<5	74	32 (43.2)	122	55 (45.1)
<16 years	2662	757 (28.4)	3839	1555 (40.5)	3898	2222 (57.0)
≥16 years	3420	1504 (44.0)	5213	3060 (58.7)	5989	3971 (66.3)
<18 years	3014	932 (30.9)	4354	1891 (43.4)	4329	2564 (59.2)
≥18 years	3068	1329 (43.3)	4698	2724 (58.0)	5558	3629 (65.3)
Overall	**6082**	**2261 (37.2)**	**9052**	**4615 (51.0)**	**9887**	**6193 (62.6)**

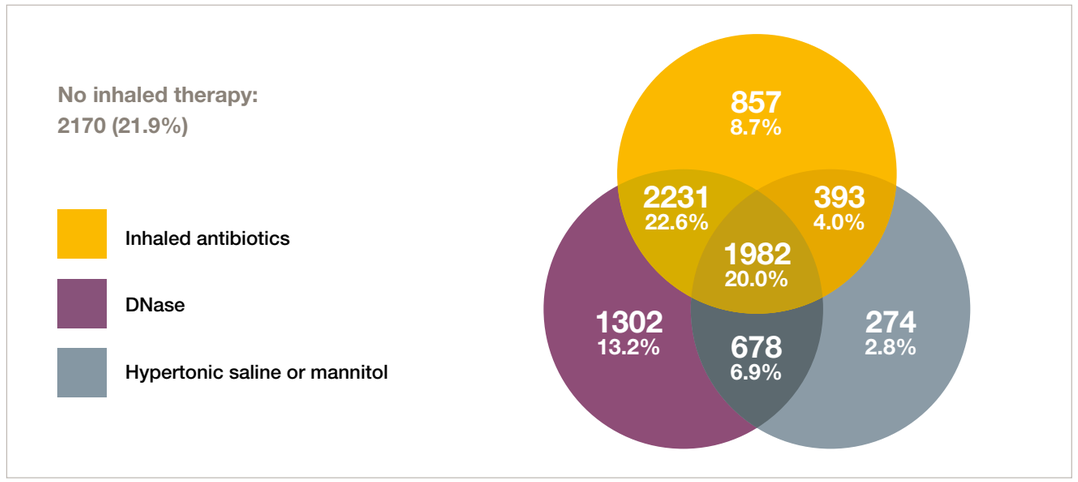
1.29 Hypertonic saline

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

	2008		2013		2017	
Age	**Total patients**	**Patients on hypertonic saline; n (%)**	**Total patients**	**Patients on hypertonic saline; n (%)**	**Total patients**	**Patients on hypertonic saline; n (%)**
0-3	605	<5	981	49 (5.0)	854	70 (8.2)
4-7	621	15 (2.4)	1004	157 (15.6)	1124	271 (24.1)
8-11	663	23 (3.5)	899	225 (25.0)	1031	352 (34.1)
12-15	773	32 (4.1)	955	303 (31.7)	889	397 (44.7)
16-19	762	33 (4.3)	1005	287 (28.6)	918	415 (45.2)
20-23	725	50 (6.9)	994	263 (26.5)	1010	331 (32.8)
24-27	605	60 (9.9)	836	220 (26.3)	942	265 (28.1)
28-31	419	37 (8.8)	703	206 (29.3)	810	256 (31.6)
32-35	260	29 (11.2)	503	131 (26.0)	698	251 (36.0)
36-39	237	16 (6.8)	315	76 (24.1)	483	160 (33.1)
40-43	165	13 (7.9)	294	64 (21.8)	311	103 (33.1)
44-47	120	13 (10.8)	213	50 (23.5)	280	78 (27.9)
48-51	59	<5	152	35 (23.0)	201	50 (24.9)
52-55	29	<5	76	23 (30.3)	138	38 (27.5)
56-59	17	0	48	9 (18.8)	76	23 (30.3)
60+	22	0	74	19 (25.7)	122	29 (23.8)
<16 years	2662	73 (2.7)	3839	734 (19.1)	3898	1090 (28.0)
≥16 years	3420	258 (7.5)	5213	1383 (26.5)	5989	1999 (33.4)
<18 years	3014	86 (2.9)	4354	879 (20.2)	4329	1304 (30.1)
≥18 years	3068	245 (8.0)	4698	1238 (26.4)	5558	1785 (32.1)
Overall	**6082**	**331 (5.4)**	**9052**	**2117 (23.4)**	**9887**	**3089 (31.2)**

1.30 Burden of treatment

The Venn diagram shows how many people with CF are on one or more inhaled therapy and the combinations they take. 2170 (21.9%) people are on no inhaled therapies.



Other Therapies

1.31 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. At the time of writing, ivacaftor is approved for use on the NHS across the UK for people aged two and older with a least one copy of 9 specific CFTR mutations, known as 'gating' mutations. Ivacaftor is additionally approved for use on the NHS in Wales for people aged 18 and over with the R117H mutation.

Number of patients on ivacaftor in the UK	**557**
Sweat chloride before ivacaftor	104 (94-113)
Sweat chloride 6-8 weeks after ivacaftor	47 (33-60)
FEV_1% before ivacaftor	62.4 (45.2-77.2)
FEV_1% 6-8 weeks after ivacaftor	71.6 (53.8-88.1)
Number of patients stopped ivacaftor ever	30

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.

Ivacaftor/Lumacaftor

Ivacaftor/Lumacaftor is licensed for use in patients aged 12 and over with two copies of the F508del mutation. In 2017 it was available to specific people with CF in the UK through a named patient access scheme. **243** people received this drug in 2017.

1.32 Oxygen and non-invasive ventilation

	Overall (n=9887)	**<16 years (n=3898)**	**≥16 years (n=5989)**	**<18 years (n=4329)**	**≥18 years (n=5558)**
Non Invasive Ventilation (NIV); n (%)	212 (2.1)	27 (0.7)	185 (3.1)	34 (0.8)	178 (3.2)
Long term oxygen; n (%)	621 (6.3)	81 (2.1)	540 (9.0)	103 (2.4)	518 (9.3)
Among those who have long-term oxygen:					
Continuously	137 (22.1)	8 (9.9)	129 (23.9)	9 (8.7)	128 (24.7)
Nocturnal or with exertion	176 (28.3)	14 (17.3)	162 (30.0)	24 (23.3)	152 (29.3)
As required (PRN)	82 (13.2)	7 (8.6)	75 (13.9)	9 (8.7)	73 (14.1)
With exacerbation	226 (36.4)	52 (64.2)	174 (32.2)	61 (59.2)	165 (31.9)

1.33 Physiotherapy

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

	Overall (n=9887)	<16 years (n=3898)	≥16 years (n=5989)	<18 years (n=4329)	≥18 years (n=5558)
Active cycle of breathing techniques; n (%)	1493 (15.1)	442 (11.3)	1051 (17.5)	496 (11.5)	997 (17.9)
Autogenic drainage (including assisted autogenic drainage); n (%)	1829 (18.5)	198 (5.1)	1631 (27.2)	272 (6.3)	1557 (28.0)
Postural drainage; n (%)	851 (8.6)	651 (16.7)	200 (3.3)	666 (15.4)	185 (3.3)
Any form of PEP; n (%)	5842 (59.1)	2934 (75.3)	2908 (48.6)	3262 (75.4)	2580 (46.4)
VEST; n (%)	178 (1.8)	91 (2.3)	87 (1.5)	104 (2.4)	74 (1.3)
Exercise; n (%)	5957 (60.3)	2549 (65.4)	3408 (56.9)	2802 (64.7)	3155 (56.8)
Other; n (%)	1650 (16.7)	876 (22.5)	774 (12.9)	923 (21.3)	727 (13.1)

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

1.34 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=9887)	<16 years (n=3898)	≥16 years (n=5989)	<18 years (n=4329)	≥18 years (n=5558)
Any supplemental feeding; n (%)	3112 (31.5)	1202 (30.8)	1910 (31.9)	1351 (31.2)	1761 (31.7)
Nasogastric tube; n (%)	105 (1.1)	16 (0.4)	89 (1.5)	19 (0.4)	86 (1.5)
Gastrostomy tube/Button; n (%)	537 (5.4)	202 (5.2)	335 (5.6)	239 (5.5)	298 (5.4)
Jejunal; n (%)	-	<5	13 (0.2)	6 (0.1)	11 (0.2)
Total Parenteral Nutrition (TPN); n (%)	7 (0.1)	<5	5 (0.1)	<5	5 (0.1)

1.35 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.

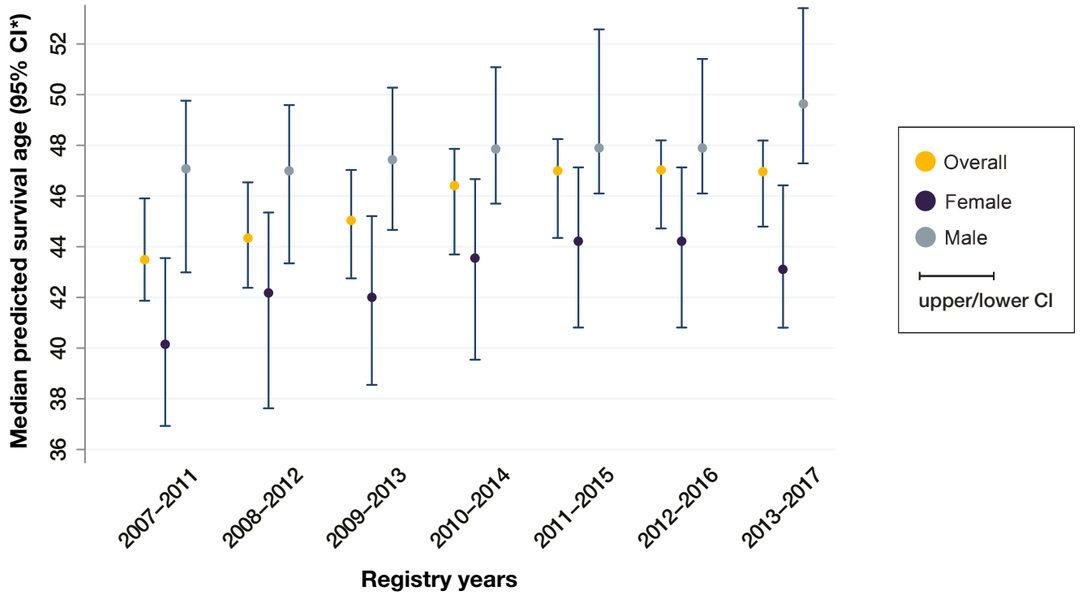
	2013	2014	2015	2016	2017
Number evaluated	220	247	229	221	235
Number accepted	136	146	125	96	121
Number receiving aged <16 years	<5	5	<5	<5	5
Bilateral lung	<5	<5	<5	<5	<5
Liver	<5	<5	<5	0	0
Other	0	0	<5	0	<5
Number receiving aged 16+ years	54	67	46	51	53
Bilateral lung	48	59	42	46	51
Liver	<5	5	<5	<5	0
Other	<5	5	<5	<5	<5

Survival

1.36 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[9] 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 **47** years. Half of people 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.



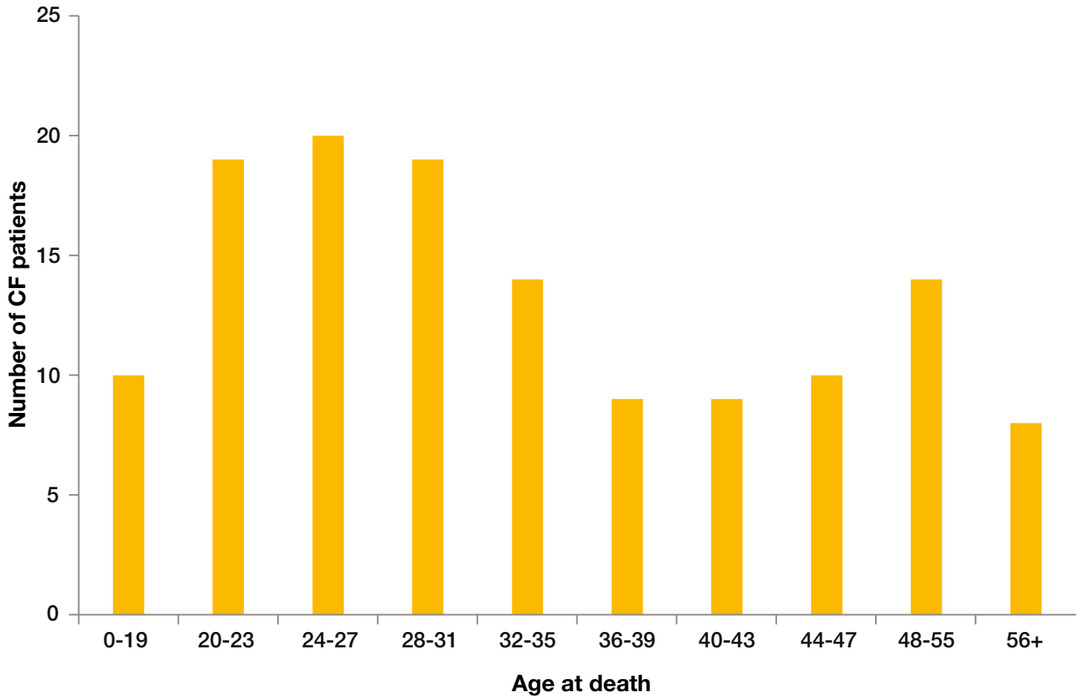
Median predicted survival age (95% CI*)				
Years	Overall	Female	Male	p-values (males vs females)
2007-2011	43.5 (41.9-45.9)	40.1 (36.9-43.6)	47.1 (43.0-49.8)	<0.001
2008-2012	44.3 (42.4-46.5)	42.2 (37.6-45.3)	47.0 (43.3-49.6)	<0.001
2009-2013	45.0 (42.8-47.0)	42.0 (38.5-45.2)	47.4 (44.7-50.3)	<0.001
2010-2014	46.4 (43.7-47.9)	43.6 (39.5-46.7)	47.9 (45.7-51.1)	<0.001
2011-2015	47.0 (44.3-48.2)	44.2 (40.8-47.1)	47.9 (46.1-52.6)	0.004
2012-2016	47.0 (44.7-48.2)	44.2 (40.8-47.1)	47.9 (46.1-51.4)	0.003
2013-2017	47.0 (44.8-48.2)	43.1 (40.8-46.4)	49.6 (47.3-53.4)	<0.001

[9] Sykes, Jenna et al. A standardized approach to estimating survival statistics for population-based cystic fibrosis cohorts, Journal of Clinical Epidemiology. 2016, Volume 70, 206-213

*Confidence Interval

1.37 Age distribution of deaths in 2017

The table below shows the ages of the 132 people with CF who died in 2017. To protect the identities of individuals, where a number of deaths in an age group is less than five, '<5' is shown in the table, and age groups are grouped together in the chart.



Age at death	Number of CF patients
0-19	10
20-23	19
24-27	20
28-31	19
32-25	14
36-39	9
40-43	9
44-47	10
48-55	14
56+	8
Total	**132**

Median age of death is based on the people with CF who died in any given year. In 2017 the median age of the 132 people who died was 31.

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 causes 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.

9818 (99.3%) patients have been genotyped with at least one recorded value.

F508del Mutations: n (%)

Homozygous F508del: 4856 (49.1%)

Heterozygous F508del: 3990 (40.4%)

1.38 Genotypes 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	8846	89.5
c.1652G->A	p.Gly551Asp	G551D	579	5.9
c.350G->A	p.Arg117His	R117H	544	5.5
c.1624G->T	p.Gly542X	G542X	362	3.7
c.489+1G->T	-	621+1G->T	252	2.5
c.3909C->G	p.Asn1303Lys	N1303K	158	1.6
c.1585-1G->A	-	1717-1G->A	138	1.4
c.1766+1G->A	-	1898+1G->A	128	1.3
c.200C->T	p.Pro67Leu	P67L	117	1.2
c.3454G->C	p.Asp1152His	D1152H	112	1.1
c.3528delC	p.Lys1177SerfsX15	3659delC	107	1.1
c.1679G->C	p.Arg560Thr	R560T	94	1.0
c.3140-26A->G	-	3272-26A->G	91	0.9
c.1519_1521delATC	p.Ile507del	I507del	86	0.9
c.1657C->T	p.Arg553X	R553X	84	0.8
c.1477C->T	p.Gln493X	Q493X	81	0.8
c.254G->A	p.Gly85Glu	G85E	81	0.8
c.3717+12191C->T	-	3849+10kbC->T	78	0.8
c.178G->T	p.Glu60X	E60X	68	0.7
c.1022_1023insTC	p.Phe342HisfsX28	1154insTC	60	0.6
c.2657+5G->A	-	2789+5G->A	56	0.6
c.3846G->A	p.Trp1282X	W1282X	54	0.5
c.948delT	p.Phe316LeufsX12	1078delT	52	0.5
c.1646G->A	p.Ser549Asn	S549N	48	0.5
c.1364C->A	p.Ala455Glu	A455E	40	0.4

Nucleotide	Protein	Legacy name	N	%
c.2052delA	p.Lys684AsnfsX38	2184delA	37	0.4
c.617T->G	p.Leu206Trp	L206W	36	0.4
c.1040G->C	p.Arg347Pro	R347P	35	0.4
c.3484C->T	p.Arg1162X	R1162X	31	0.3
c.1558G->T	p.Val520Phe	V520F	30	0.3
c.579+3A->G		711+3A->G	30	0.3
c.2657+2_2657+3insA		2789+2insA	26	0.3
c.1210-12[5](AJ574948.1:g.152T[5])		5T	25	0.3
c.2988+1G->A		3120+1G->A	23	0.2
c.1040G->A	p.Arg347His	R347H	23	0.2
c.1055G->A	p.Arg352Gln	R352Q	22	0.2
c.1753G->T	p.Glu585X	E585X	22	0.2
c.2583delT	p.Phe861LeufsX3	2711delT	19	0.2
c.3472C->T	p.Arg1158X	R1158X	19	0.2
c.1000C->T	p.Arg334Trp	R334W	19	0.2
c.1705T->G	p.Tyr569Asp	Y569D	16	0.2
c.2834C->T	p.Ser945Leu	S945L	16	0.2
c.2490+1G->A		2622+1G->A	15	0.2
c.1393-1G->A		1525-1G->A	15	0.2
c.532G->A	p.Gly178Arg	G178R	15	0.2
c.2125C->T	p.Arg709X	R709X	14	0.1
c.3197G->A	p.Arg1066His	R1066H	13	0.1
c.2537G->A	p.Trp846X	W846X	13	0.1
c.1466C->A	p.Ser489X	S489X	12	0.1
c.579+1G->T		711+1G->T	12	0.1
c.2052_2053insA	p.Gln685ThrfsX4	2184insA	12	0.1
c.658C->T	p.Gln220X	Q220X	12	0.1
c.2875delG	p.Ala959HisfsX9	3007delG	11	0.1
c.3196C->T	p.Arg1066Cys	R1066C	9	0.1
c.292C->T	p.Gln98X	Q98X	9	0.1
c.3705T->G	p.Ser1235Arg	S1235R	9	0.1
c.4196_4197delTC	p.Cys1400X	4326delTC	8	0.1
c.1675G->A	p.Ala559Thr	A559T	8	0.1
c.3276C->A	p.Tyr1092X	Y1092X(C->A)	8	0.1
c.2988G->A		3120G->A	8	0.1
c.1679+1G->C		1811+1G->C	7	0.1
c.224G->A	p.Arg75Gln	R75Q	7	0.1
c.349C->T	p.Arg117Cys	R117C	7	0.1
c.1645A->C	p.Ser549Arg	S549R(A->C)	6	0.1
c.223C->T	p.Arg75X	R75X	6	0.1
c.2128A->T	p.Lys710X	K710X	6	0.1
c.2290C->T	p.Arg764X	R764X	5	0.1
c.1986_1989delAACT	p.Thr663ArgfsX8	2118del4	5	0.1
c.443T->C	p.Ile148Thr	I148T	5	0.1
c.274G->A	p.Glu92Lys	E92K	<5	-
c.3208C->T	p.Arg1070Trp	R1070W	<5	-

Nucleotide	Protein	Legacy name	N	%
c.2353C->T	p.Arg785X	R785X	<5	-
c.2051_2052delAAinsG	p.Lys684SerfsX38	2183AA->G or 2183delAA->G	<5	-
c.1679G->A	p.Arg560Lys	R560K	<5	-
c.3718-1G->A		3850-1G->A	<5	-
c.2012delT	p.Leu671X	2143delT	<5	-
c.328G->C	p.Asp110His	D110H	<5	-
c.3884_3885insT	p.Ser1297PhefsX5	4016insT	<5	-
c.1329_1330insAGAT	p.Ile444ArgfsX3	1461ins4	<5	-
c.1736A->G	p.Asp579Gly	D579G	<5	-
c.2551C->T	p.Arg851X	R851X	<5	-
c.262_263delTT	p.Leu88IlefsX22	394delTT	<5	-
c.2464G->T	p.Glu822X	E822X	<5	-
c.2260G->A	p.Val754Met	V754M	<5	-
c.3659delC	p.Thr1220LysfsX8	3791delC	<5	-
c.3080T->C	p.Ile1027Thr	I1027T	<5	-
c.1007T->A	p.Ile336Lys	I336K	<5	-
c.2215delG	p.Val739TyrfsX16	2347delG	<5	-
c.595C->T	p.His199Tyr	H199Y	<5	-
c.2668C->T	p.Gln890X	Q890X	<5	-
c.2780T->C	p.Leu927Pro	L927P	<5	-
c.3310G->T	p.Glu1104X	E1104X	<5	-
c.2491G->T	p.Glu831X	E831X	<5	-
c.220C->T	p.Arg74Trp	R74W	<5	-
c.54-5940_273+10250del21kb	p.Ser18ArgfsX16	CFTRdele2,3	<5	-
c.2374C->T	p.Arg792X	R792X	<5	-
c.442delA	p.Ile148LeufsX5	574delA	<5	-
c.3752G->A	p.Ser1251Asn	S1251N	<5	-
c.1679+1.6kbA->G		1811+1.6kbA->G	<5	-
c.3700A->G	p.Ile1234Val	I1234V	<5	-
c.1585-8G->A		1717-8G->A	<5	-
c.3209G->A	p.Arg1070Gln	R1070Q	<5	-
c.1116+1G->A		1248+1G->A	<5	-
c.2195T->G	p.Leu732X	L732X	<5	-
c.1766+1G->C		1898+1G->C	<5	-
c.1340delA	p.Lys447ArgfsX2	1471delA	<5	-
c.3266G->A	p.Trp1089X	W1089X	<5	-
c.91C->T	p.Arg31Cys	R31C	<5	-
c.2991G->C	p.Leu997Phe	L997F	<5	-
c.3181G->C	p.Gly1061Arg	G1061R	<5	-
c.1727G->C	p.Gly576Ala	G576A	<5	-
c.1203G->A	p.Trp401X	W401X(TGA)	<5	-
c.273+1G->A		405+1G->A	<5	-
c.3194T->C	p.Leu1065Pro	L1065P	<5	-
c.1647T->G	p.Ser549Arg	S549R(T->G)	<5	-
c.1418delG	p.Gly473GlufsX54	1548delG	<5	-

Nucleotide	Protein	Legacy name	N	%
c.4111G->T	p.Glu1371X	E1371X	<5	-
	p.Ser549Arg	S549R	<5	-
c.1573C->T	p.Gln525X	Q525X	<5	-
c.1651G->A	p.Gly551Ser	G551S	<5	-
c.580-1G->T		712-1G->T	<5	-
c.137C->A	p.Ala46Asp	A46D	<5	-
c.1654C->T	p.Gln552X	Q552X	<5	-
c.1682C->A	p.Ala561Glu	A561E	<5	-
c.4077_4080delTGTTinsAA	p.Val1360delfsX?	4209TGTT->AA	<5	-
c.613C->T	p.Pro205Ser	P205S	<5	-
c.3485G->T	p.Arg1162Leu	R1162L	<5	-
c.1240C->T	p.Gln414X	Q414X	<5	-
c.933_935delCTT	p.Phe312del	F311del	<5	-
c.3205G->A	p.Gly1069Arg	G1069R	<5	-
c.1210-12[5](AJ574948.1:g.152T[7])		7T	<5	-
c.2989-1G->A		3121-1G->A	<5	-
c.1081delT	p.Trp361GlyfsX8	1213delT	<5	-
c.1A->G	p.Met1Val	M1V	<5	-
c.4046G->A	p.Gly1349Asp	G1349D	<5	-
c.2735C->A	p.Ser912X	S912X	<5	-
c.1209+1G->A		1341+1G->A	<5	-
c.1545_1546delTA	p.Tyr515X	1677delTA	<5	-
c.3773_3774insT	p.Leu1258PhefsX7	3905insT	<5	-
c.166G->A	p.Glu56Lys	E56K	<5	-
c.2739T->A	p.Tyr913X	Y913X	<5	-
c.2002C->T	p.Arg668Cys	R668C	<5	-
c.3611G->A	p.Trp1204X	W1204X(3743G->A)	<5	-
c.1202G->A	p.Trp401X	W401X(TAG)	<5	-
c.3230T->C	p.Leu1077Pro	L1077P	<5	-
c.1021T->C	p.Ser341Pro	S341P	<5	-
'Other' selected		-	1249	12.6

1.39 Genotypes by devolved nation

	England		Scotland		Wales		Northern Ireland	
	n = 8201	%	n = 858	%	n = 401	%	n = 427	%
F508del	7361	89.8%	765	89.2%	359	89.5%	361	84.5%
G542X	251	3.1%	62	7.2%	22	5.5%	27	6.3%
G551D	429	5.2%	91	10.6%	19	4.7%	40	9.4%
R117H	402	4.9%	71	8.3%	12	3.0%	59	13.8%
621+1G->T	182	2.2%	12	1.4%	45	11.2%	13	3.0%
1898+1G->A	97	1.2%	<5	-	26	6.5%	<5	-

Section 2 and 3: Centre-level analysis

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

	Paediatric	Adult	Total
Centres	30	26	56
Stand-alone clinics	2	2	4
Networked clinics	68	7	75

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

Lots of different factors can affect the outcomes of people with CF in centres, not all of which are within a centre's control. This might include the economic profile of the area, the age at which the person with CF was diagnosed and referred to the centre 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 62.

Key

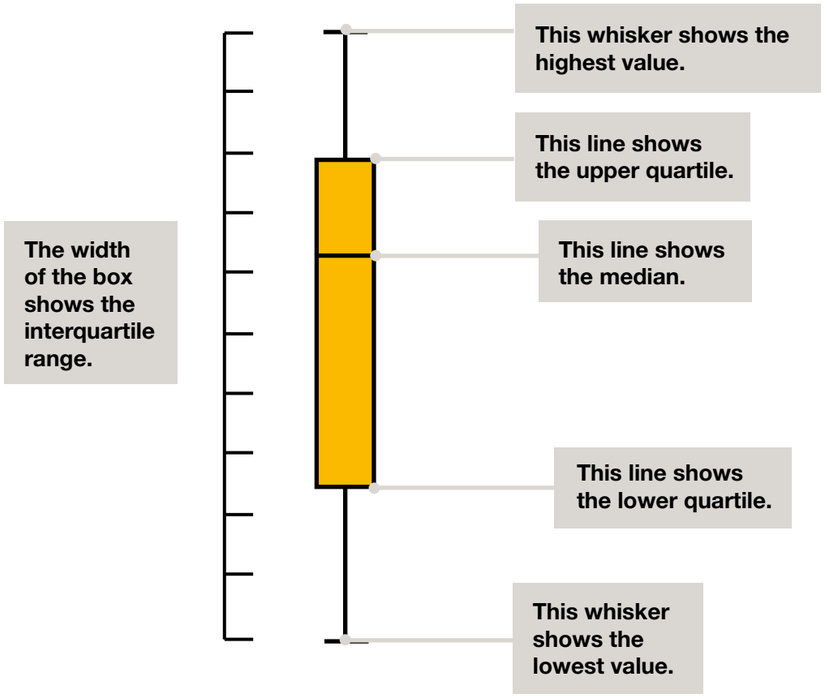
Paediatric centre

Adult centre

A guide to the charts

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

Box plots



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

Funnel plots

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

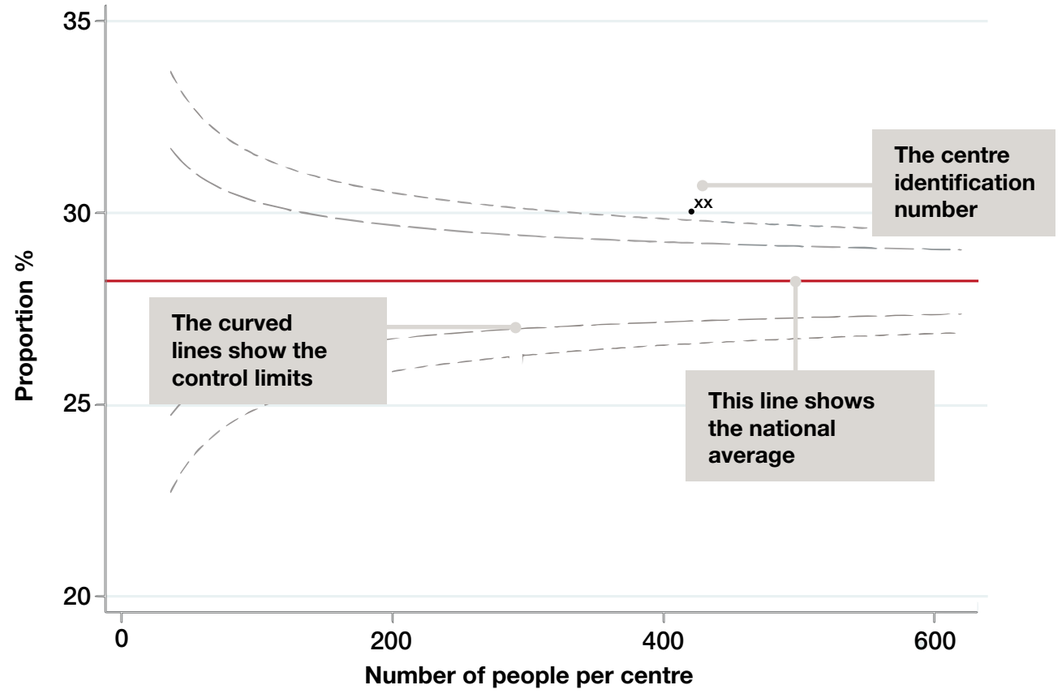
There will always be some natural variation between centres, because of differences between the population receiving care. Using only the national average as a standard can make it difficult to tell whether a survival rate that sits above the national average is higher than we would expect it to be,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

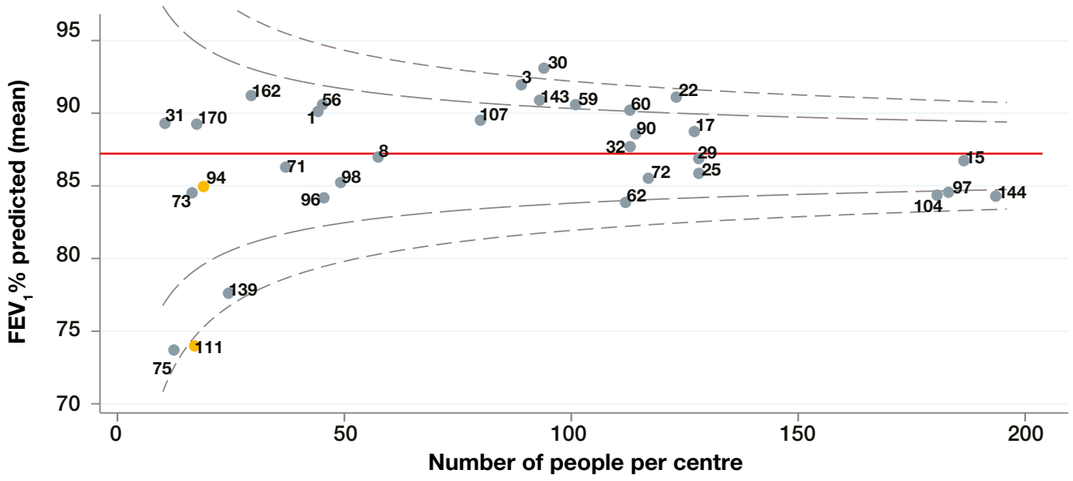
N=4224

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

Key

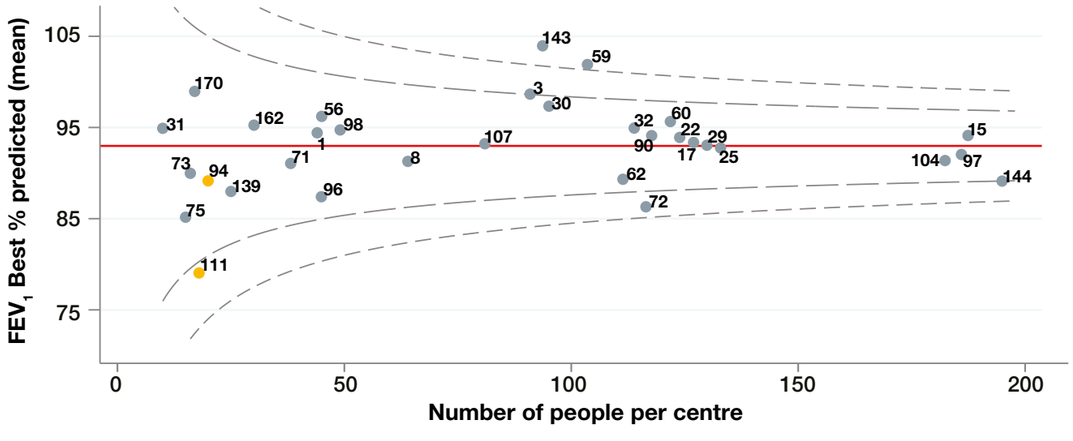
Centres with their network clinics · Stand-alone clinics · 2 standard deviations · 3 standard deviations

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



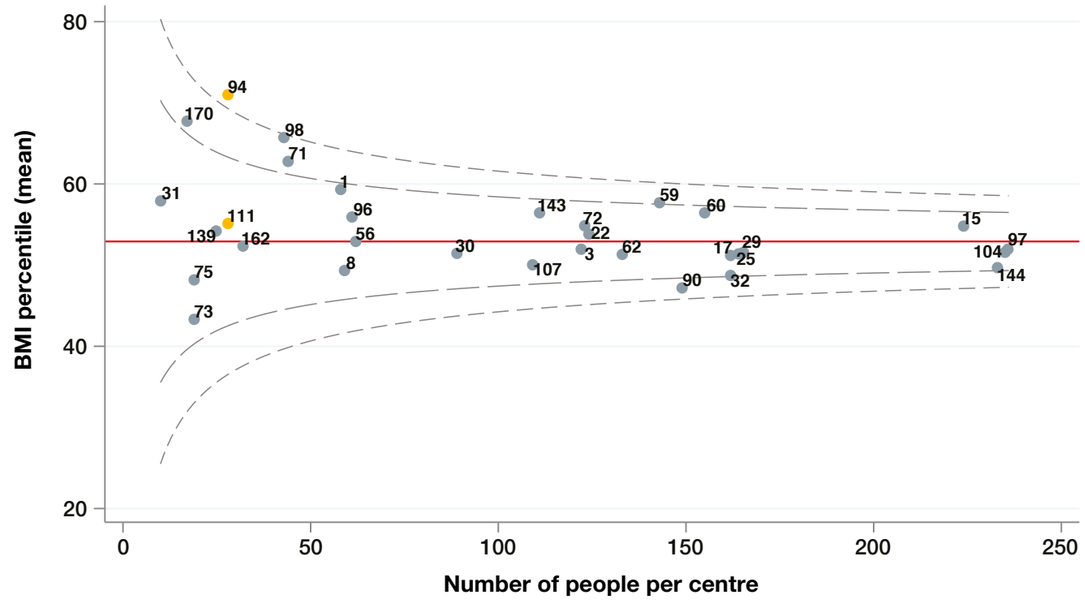
The mean FEV_1% predicted for patients attending paediatric centres/clinics is 86.5% predicted.

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



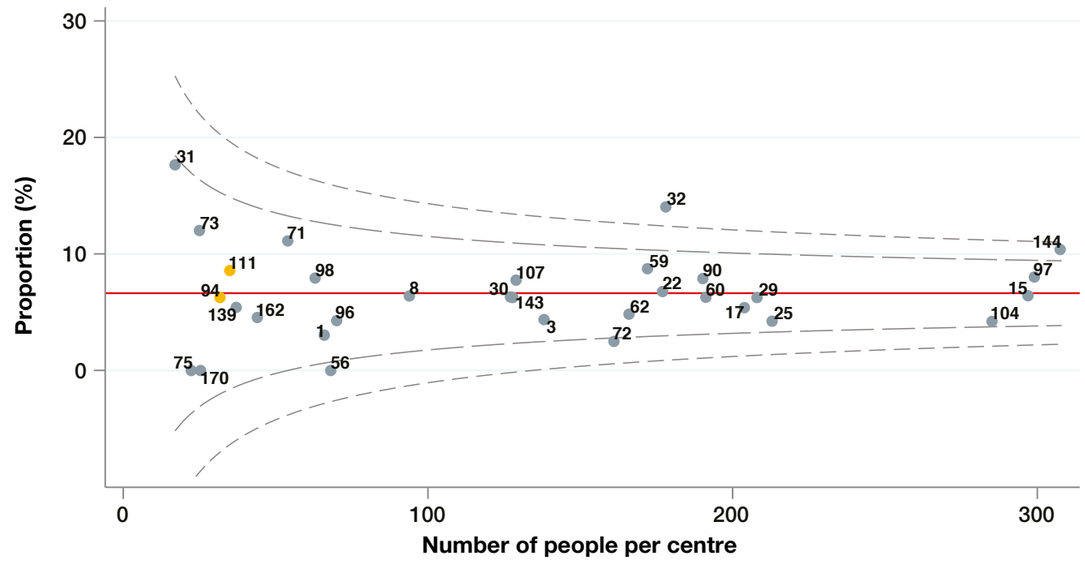
The mean Best FEV_1% predicted for patients attending paediatric centres/clinics is 93.0% predicted. Where Best FEV_1% predicted was missing, the FEV_1% predicted at annual review was used.

2.3 Age adjusted BMI percentile in patients aged 1-15 years by paediatric centre/clinic



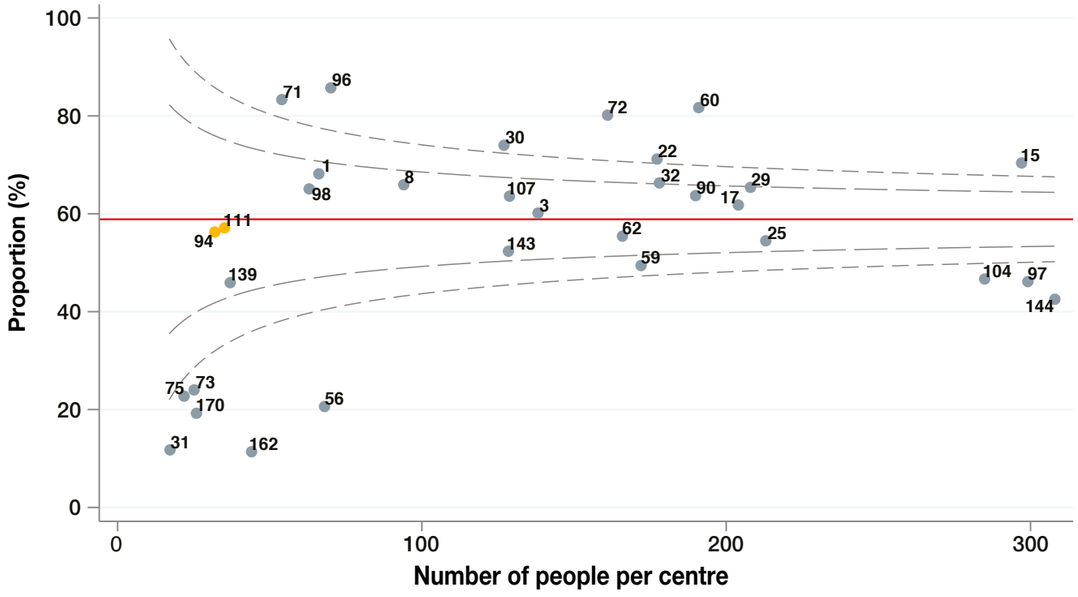
The mean BMI percentile for patients attending paediatric centres/clinics is 52.9.

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



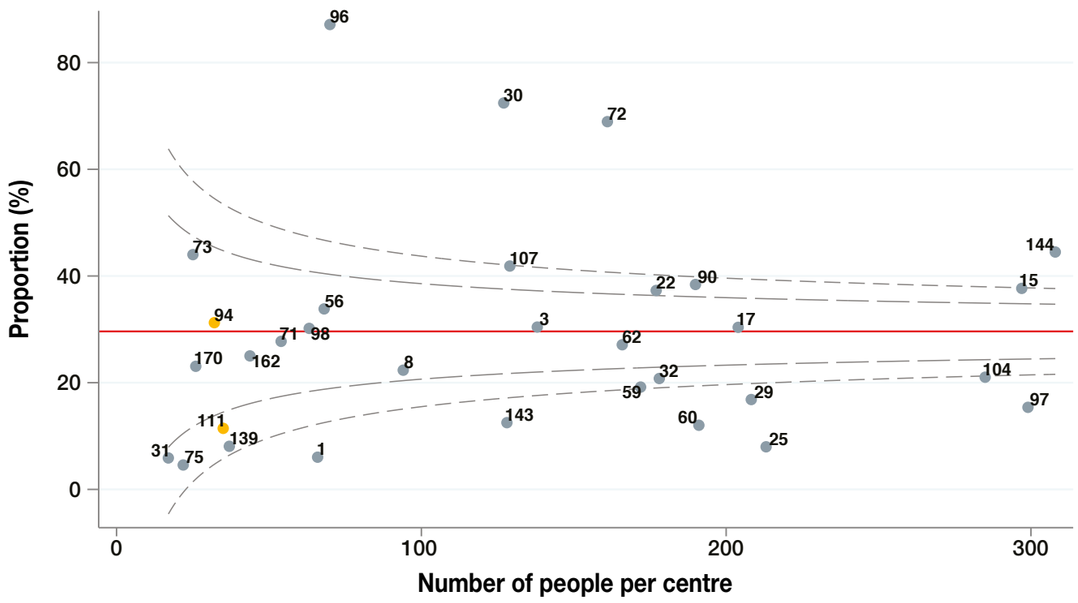
The proportion of patients with chronic *Pseudomonas aeruginosa* in paediatric centres/clinics is 6.6%.

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



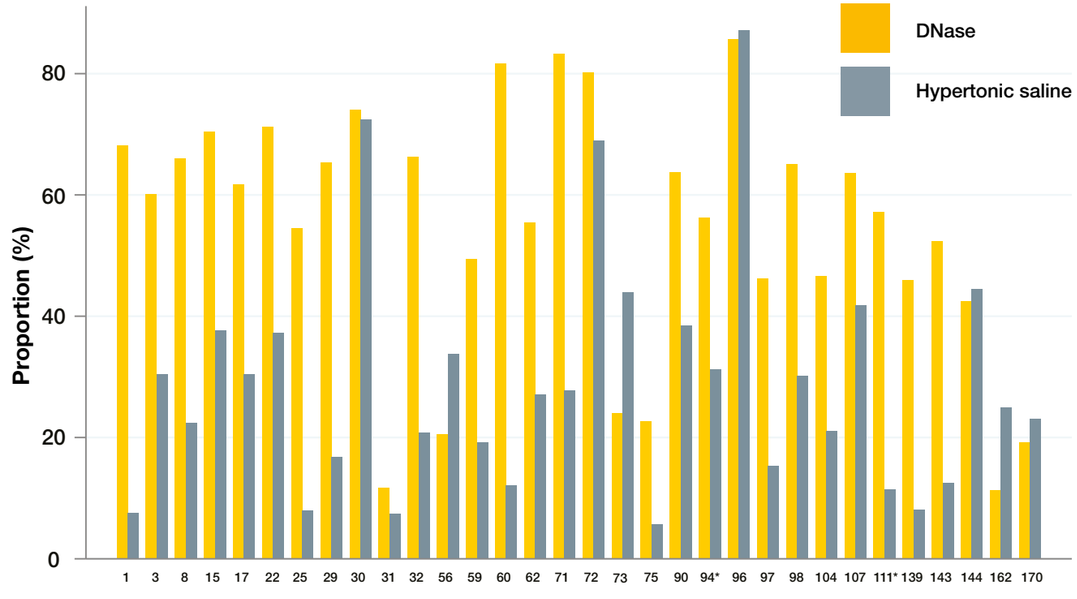
The proportion of patients receiving DNase treatment in paediatric centres/clinics is 58.9%.

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



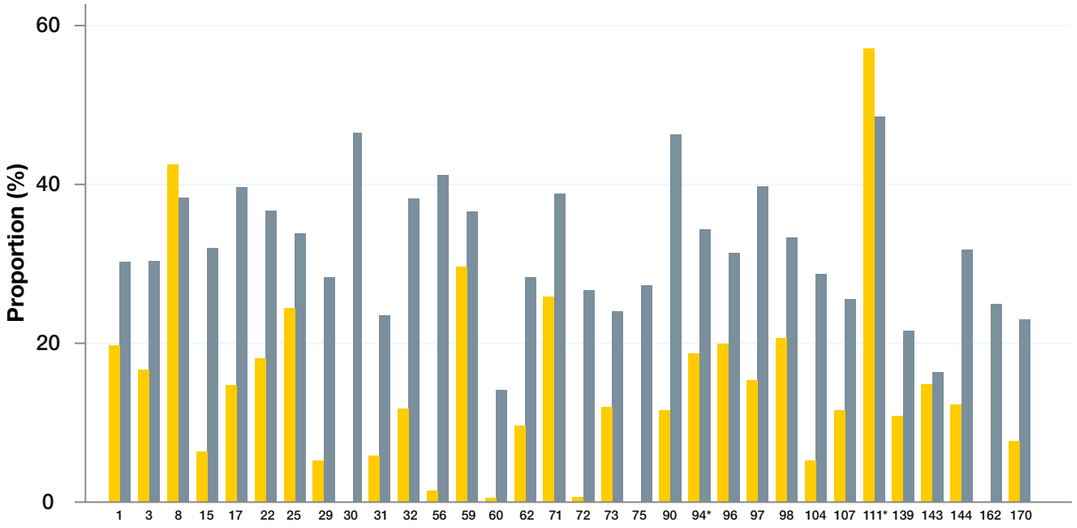
The proportion of patients receiving hypertonic saline treatment in paediatric centres/clinics is 29.6%.

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



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.



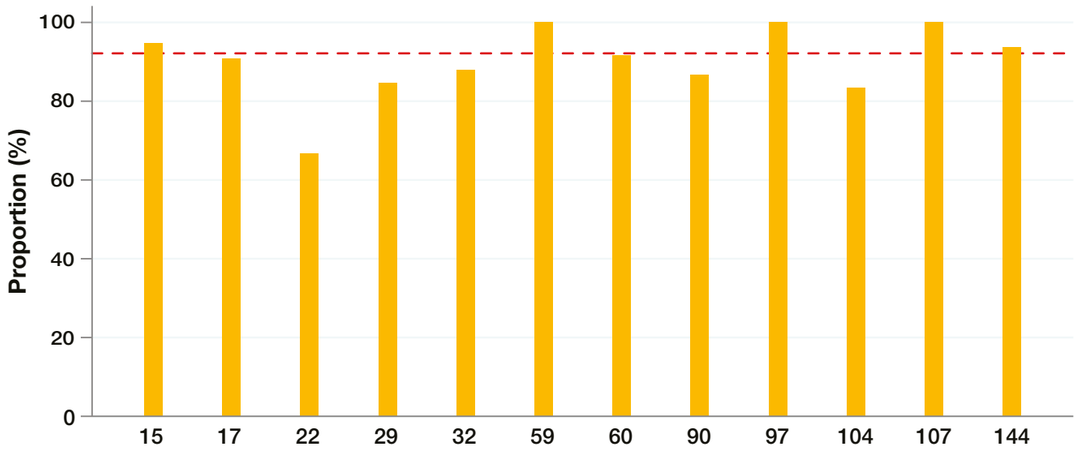
The proportion of patients receiving IVs at home was 12.9% and in hospital was 32.6%. The proportion receiving any IVs was 35.2%.

Home
Hospital

* Stand-alone clinics

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

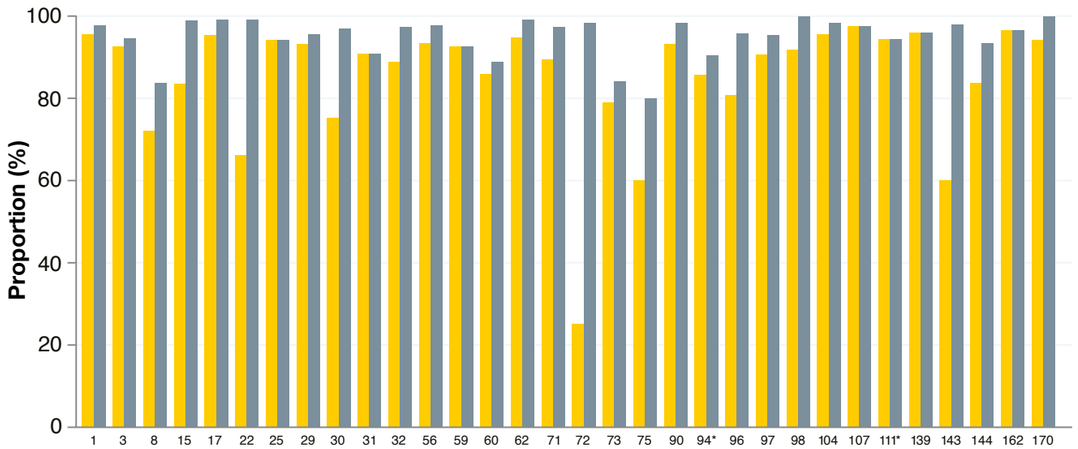
This chart excludes centres where less than 10 patients had chronic *P. aeruginosa*.



92.1% of patients with chronic *P. aeruginosa* received inhaled antibiotics.

2.10 Data completeness by paediatric centre/clinic

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



The proportion of patients with a valid Best FEV_1% was 84.8%.
The proportion with an FEV_1% at annual review was 96.0%.

* Stand-alone clinics

Section 3: Adult centre analysis

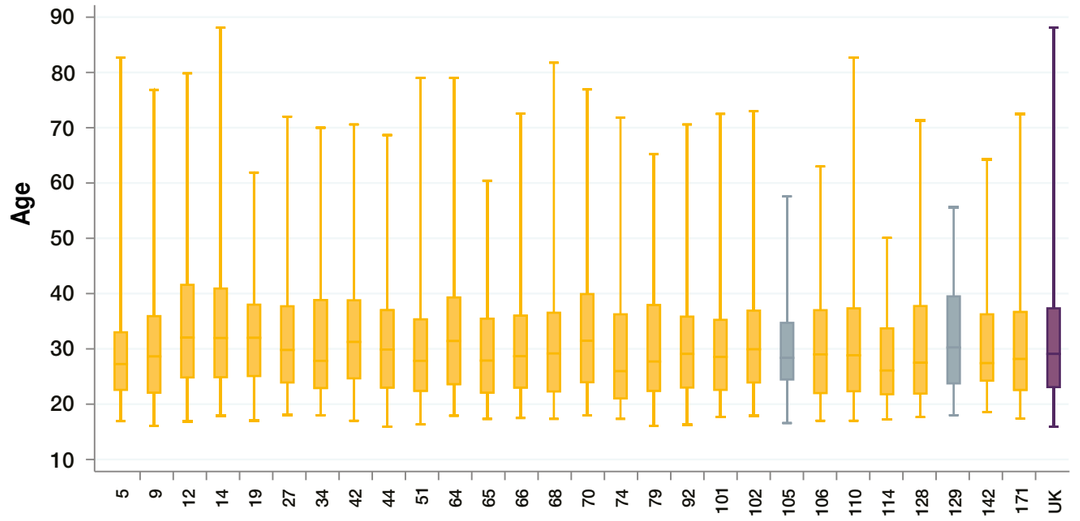
N=5663

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.

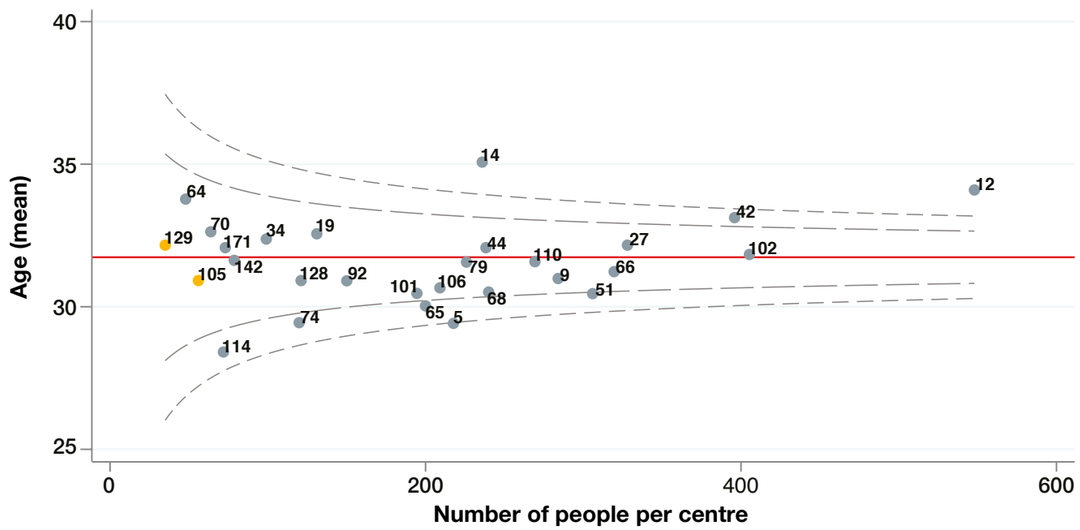
Key
Centres with their network clinics · Stand-alone clinics · — — — 2 standard deviations · — — — 3 standard deviations

3.1 Age distribution by adults service

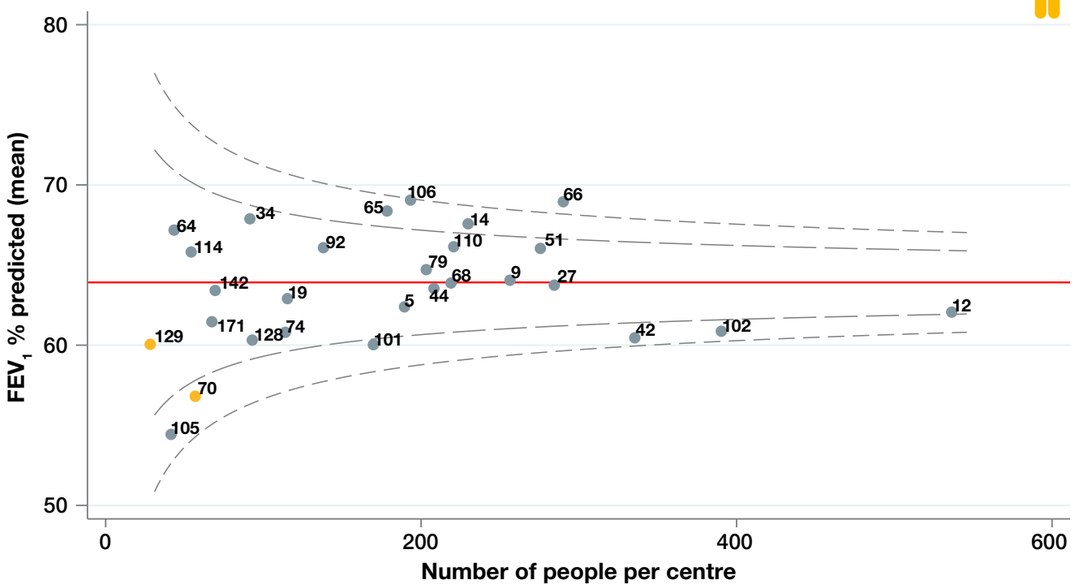
The box-whisker plot shows the age distribution of patients within each centre/clinic.
In 2017 the median age in adults services was 29 years (IQR:23-37)



The funnel plot below shows how the mean age in adult centres compares to the national mean. In 2017 the national mean age was 31.7 years.

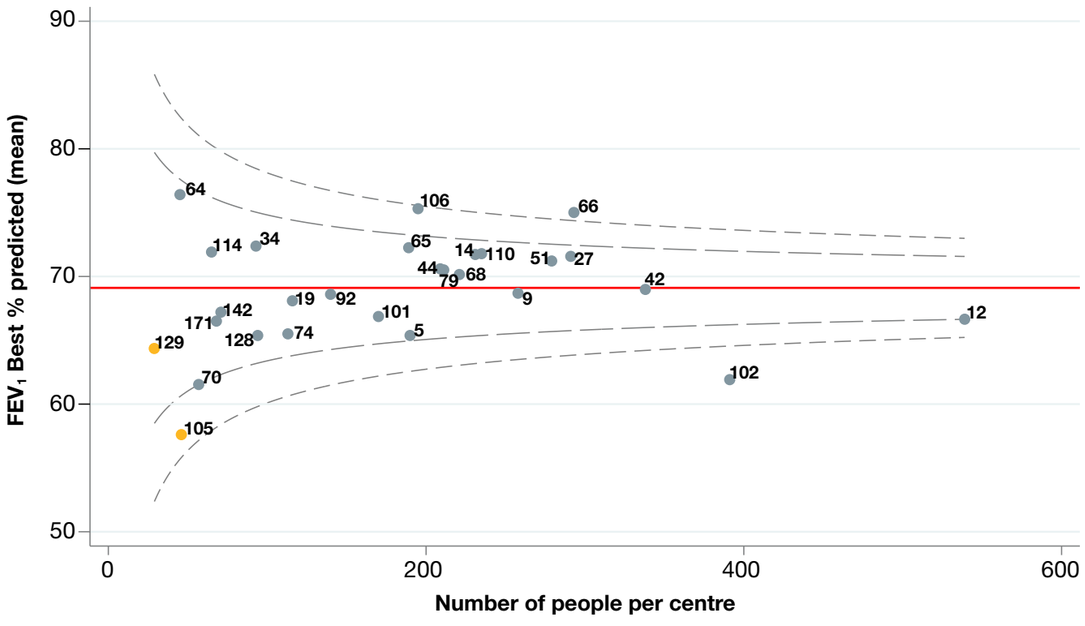


3.2 Age adjusted FEV_1% predicted at annual review in patients without a history of lung transplant, by adult service



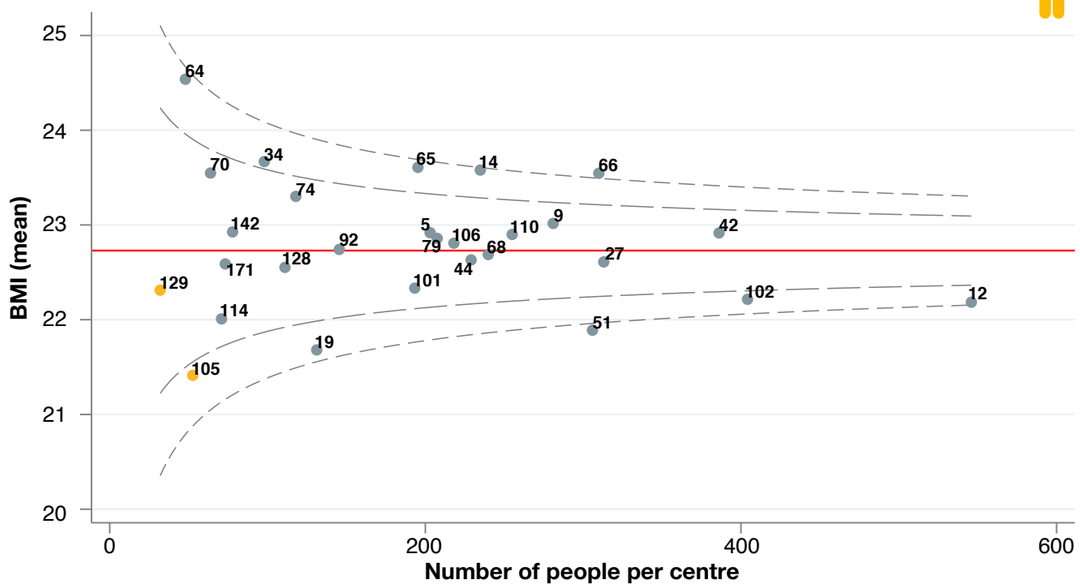
The mean FEV_1% predicted in adult services is 63.8%.

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



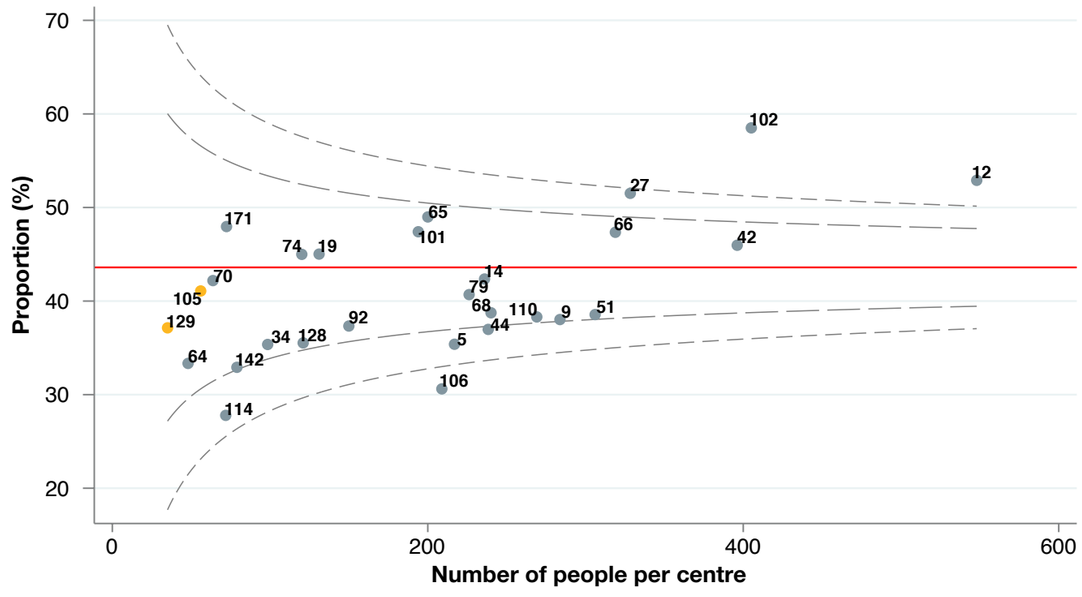
In 2017 the national mean was 69.1%. Where Best FEV_1% predicted was missing, or lower than the FEV_1 at annual review the FEV_1% value at annual review was used.

3.4 Age adjusted BMI among patients aged 16 years and older by adult service



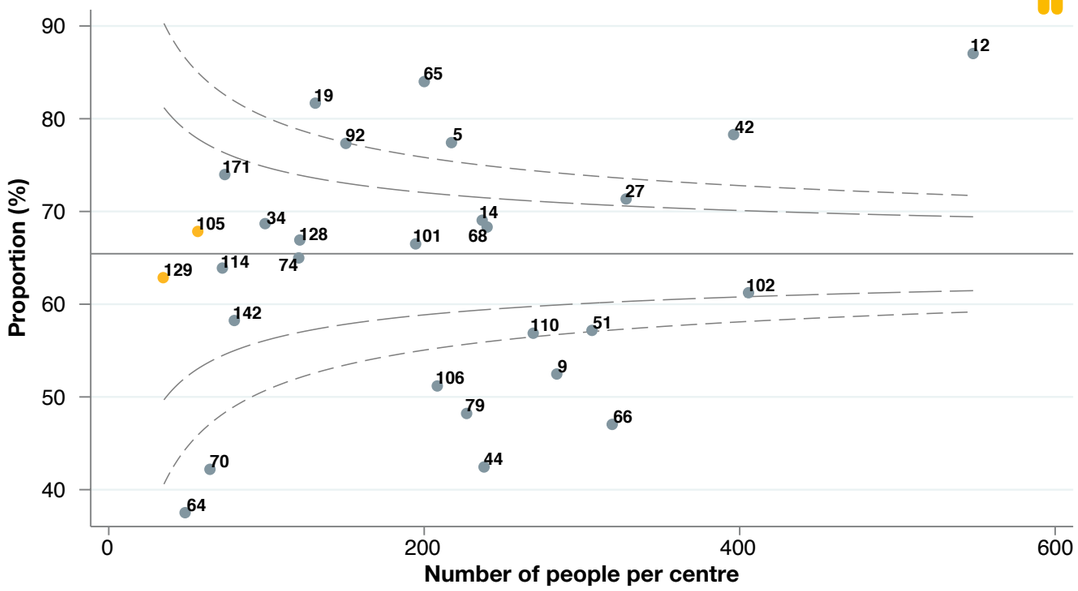
The mean BMI in adult services is 22.7.

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



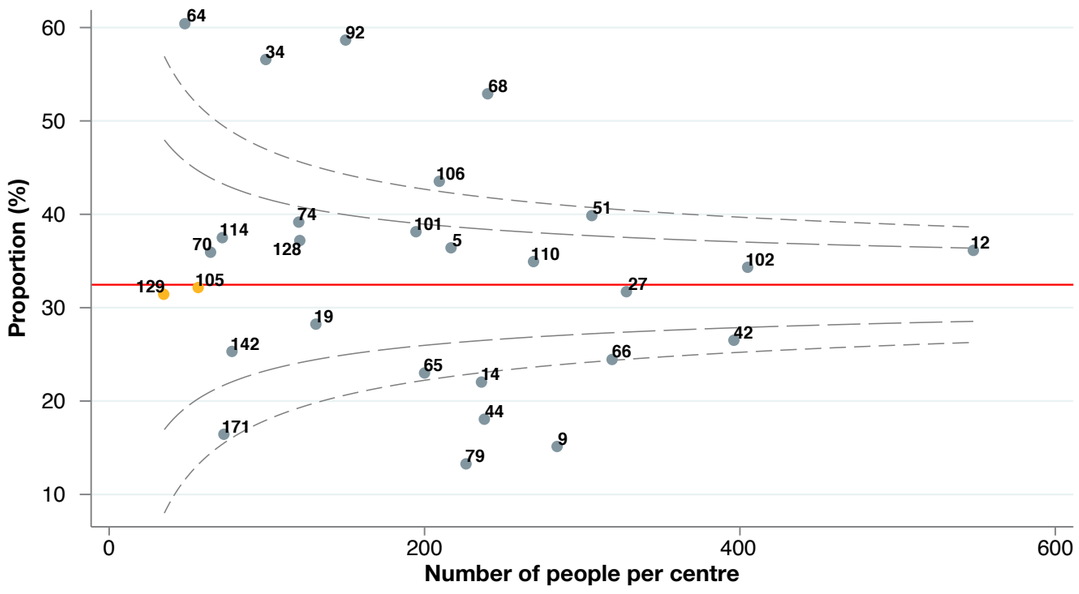
The proportion of patients with chronic *P. aeruginosa* in adult centres/clinics is 43.6%.

3.6 Proportion of patients receiving DNase treatment by adult service



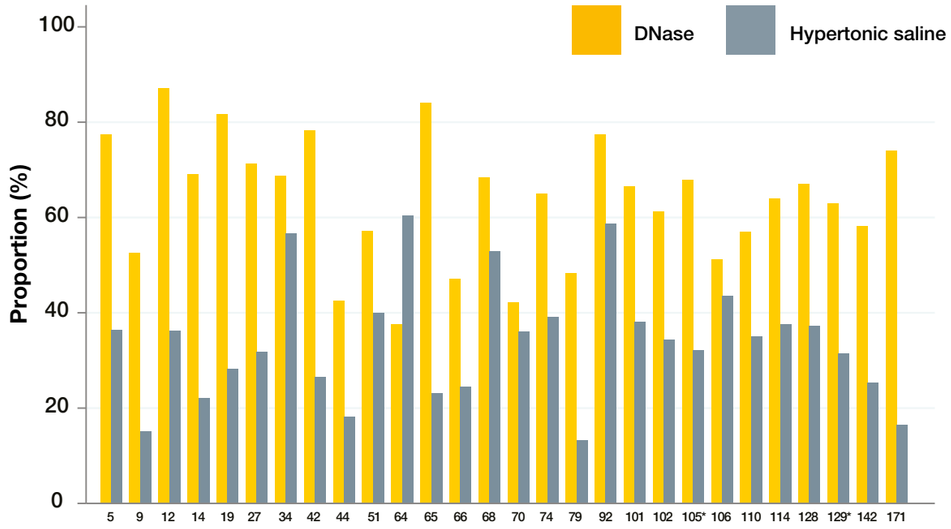
The proportion of patients receiving DNase treatment in adult centres/clinics is 65.4%.

3.7 Proportion of patients receiving hypertonic saline by adult service



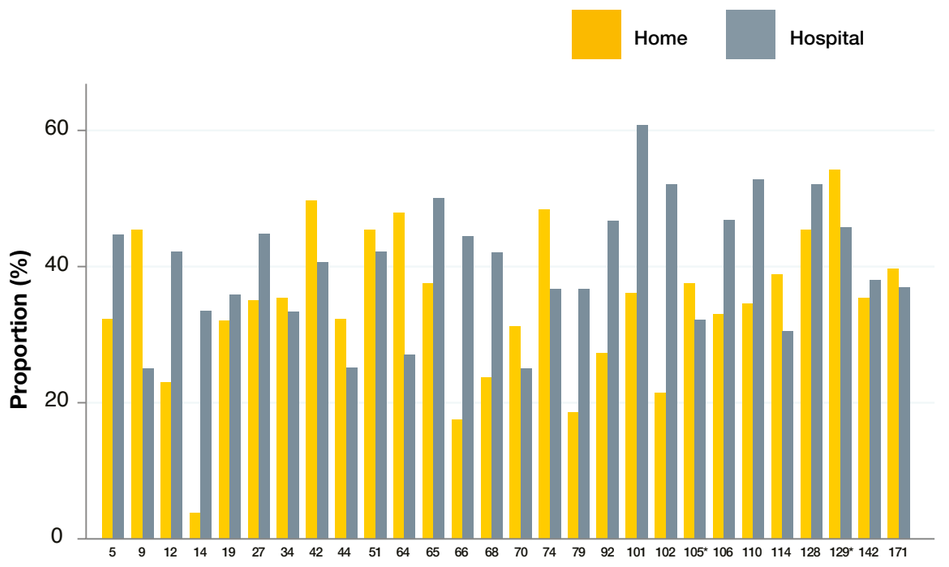
The proportion of patients receiving hypertonic saline treatment in adult centres/clinics is 32.5%.

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



3.9 IV use by adult service

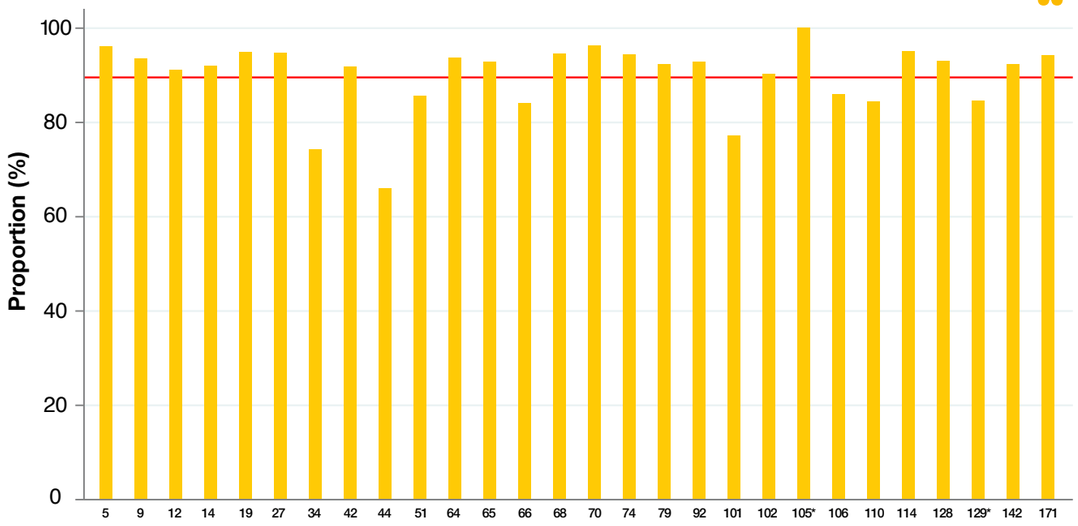
The chart below shows the proportion of patients with at least 1 IV day at home and in hospital. Patients may have a combination of home and hospital IV days.



The proportion of patients in adult centres receiving IV antibiotics at home was 32.0% and in hospital was 41.8%. The proportion receiving any IVs was 52.4%.

* Stand-alone clinics

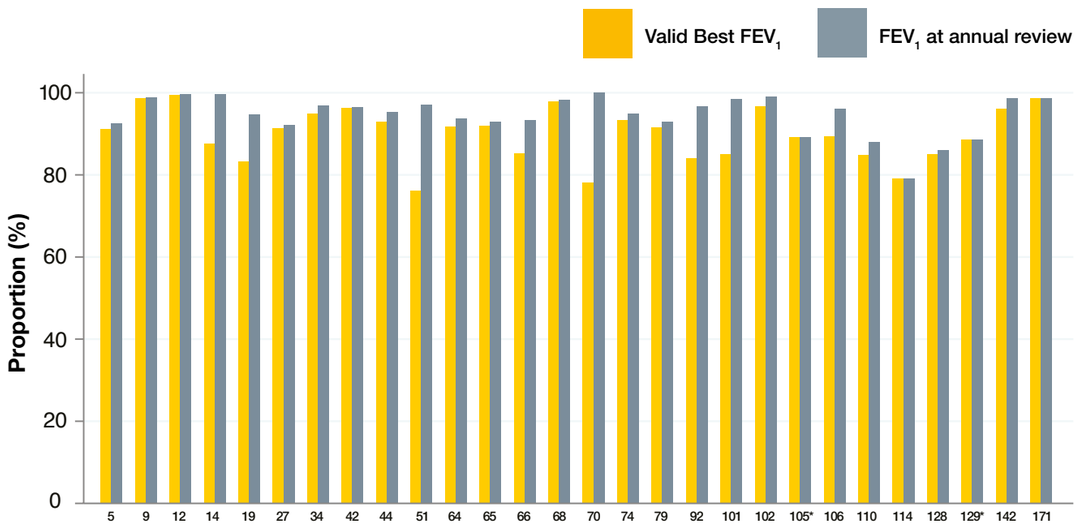
3.10 Inhaled antibiotic use for patients with chronic Pseudomonas by adult service



89.6% 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 by adult service

FEV_1 was considered valid if it was not missing, and the percent predicted was not more than 0.5% lower than the annual review value. For some patients there may be medical reasons why FEV_1 could not be taken, so centres may not be able to get 100% completeness.



The proportion of patients with a valid Best FEV_1 was 91.1%. The proportion with an FEV_1 at annual review was 95.6%.

* Stand-alone clinics

Glossary

Word/Phrase	Meaning
2017	1 January 2017 – 31 December 2017
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	Burkholderia cepacia complex are 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 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 gastro-intestinal tract.
GLI equations	Global Lung Initiative, the equation used for calculating FEV_1% predicted from absolute FEV_1 that takes into account age, gender, height and ethnicity.
H. influenzae	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 2017 the median age of the 132 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 47 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 caused by chronic inflammation of the nasal lining.
NBS	Newborn screening 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.
Nontuberculous Mycobacteria (NTM)	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, is 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 at 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.
Pre-natal	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.
S. aureus	*Staphylococcus aureus* is a bacterium 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 that after careful calculations there is a definite difference between two groups, which is not simply a result of chance.

Appendix 1: UK CF Registry Steering Committee structure

UK CF Registry Steering Committee

Role	Forename	Surname	Organisation
Director of Impact †	Keith	Brownlee	Cystic Fibrosis Trust
CF physician – Paediatrics*	Siobhán	Carr	Royal Brompton Hospital
Senior Statistician †	Susan	Charman	Cystic Fibrosis Trust
Registry Lead	Rebecca	Cosgriff	Cystic Fibrosis Trust
Cystic fibrosis centre data manager	Lance	Dennard	Lewisham Hospital
CF physician - Paediatrics	Iolo	Doull	Children's Hospital for Wales
CF physician - Adults	Caroline	Elston	King's College Hospital
Commissioner, England	Kathy	Blacker	NHS England
Registry Clinical Data Manager †	Elaine	Gunn	Cystic Fibrosis Trust
Registry Development Manager †	Mary	Yip	Cystic Fibrosis Trust
Commissioner, Wales †	Claire	Nelson	NHS Wales
Allied health professional	Alan	Peres	Royal Brompton Hospital
CF physician - Adults	Simon	Range	Glenfield Hospital
Commissioner, Scotland	David	Steele	NHS Scotland
Person with CF	James	Thomson	N/A
Parent representative	Grant	Valentine	N/A
Chair of the Research Committee #	Martin	Wildman	Northern General Hospital

UK CF Registry Research Committee

Role	Forename	Surname	Organisation
Pharmacovigilance PI	Diana	Bilton	Royal Brompton Hospital
CF physician – Adults (retired)			
Registry consultant	Noreen	Caine	Cystic Fibrosis Trust
Pharmacovigilance PI	Siobhán	Carr	Royal Brompton Hospital
CF physician - Paediatrics			
Senior Statistician †	Susan	Charman	Cystic Fibrosis Trust
Registry Lead	Rebecca	Cosgriff	Cystic Fibrosis Trust
Pharmacovigilance PI	Steve	Cunningham	Royal Hospital for Sick Children
CF physician - Paediatrics			
Parent representative	Marian	Dmochowska	N/A
Registry Clinical Data Manager †	Elaine	Gunn	Cystic Fibrosis Trust
Person with CF	Dominic	Kavanagh	Cystic Fibrosis Trust
Pharmacovigilance PI	Nicholas	Simmonds	Royal Brompton Hospital
CF physician - Adults			
CF physician - Adults*#	Martin	Wildman	Northern General Hospital
Registry Development Manager †	Mary	Yip	Cystic Fibrosis Trust

*Chair † Non-voting member # Caldicott guardian

Appendix 2: Data tables

Paediatric centres/clinics providing data in 2017 – ordered by clinic ID

* Where 'Best' values were missing, or lower than FEV_1% predicted taken at annual review, the annual review value was used.
** For data completeness, 'Best' values were taken to be valid if they were not missing and the percent predicted was not more than 0.5% lower than FEV_1% predicted taken at annual reviews.

					Age		FEV_1% predicted at annual review		FEV_1% predicted at annual review		Best FEV_1% predicted				Data completeness for FEV_1			
Location	Name	Clinic ID	Total Active	Number with annual review	Mean	Median	Number	Mean - unadjusted	Mean - adjusted	Median	Number*	Mean - unadjusted	Mean - adjusted	Median	Number with valid Best FEV_1**	Percentage with valid Best FEV_1	Number with FEV_1 at annual review	Percentage with FEV_1 at annual review
Leicester	Leicester Royal Infirmary	1	69	66	8.8	8.2	44	88.5	89.3	89.1	44	93.5	94.2	93.3	43	95.6	44	97.8
Sheffield	Sheffield Children's Hospital	3	142	138	8.5	8.3	89	91.5	91.2	93.2	91	98.3	98.0	98.7	87	92.6	89	94.7
Stoke-on-Trent	University Hospitals of North Midlands	8	100	94	8.6	8.0	57	86.4	86.2	89.0	64	91.0	91.0	93.5	49	72.1	57	83.8
London - South West	Royal Brompton Hospital	15	315	297	8.6	8.1	187	85.3	85.9	87.7	188	93.1	93.7	93.4	158	83.6	187	98.9
London - South East	King's College Hospital	17	218	204	8.4	7.9	127	86.9	88.0	88.6	127	91.9	92.9	94.3	122	95.3	127	99.2
Oxford	John Radcliffe Hospital	22	184	177	9.0	9.0	123	89.7	90.3	93.3	124	93.0	93.4	95.6	82	66.1	123	99.2
Leeds	St James's University Hospital	25	229	213	8.0	7.3	128	85.7	85.2	88.8	130	93.3	92.7	95.6	130	94.2	130	94.2
Southampton	Southampton General Hospital	29	218	208	8.6	8.8	128	85.1	86.1	88.8	131	91.8	92.8	95.1	125	93.3	128	95.5
London - East	Royal London Hospital	30	130	127	9.3	9.1	94	91.4	92.2	91.0	95	96.2	96.8	95.2	73	75.3	94	96.9
Inverness	Raigmore Hospital	31	18	17	8.0	8.9	10	88.4	88.6	88.1	10	94.3	94.4	94.0	10	90.9	10	90.9
Bristol	Bristol Royal Hospital for Children	32	185	178	8.7	8.0	113	85.5	86.9	83.7	114	93.7	94.9	92.6	103	88.8	113	97.4
Glasgow	Royal Hospital for Sick Children	56	95	68	8.5	8.8	45	89.8	89.7	91.5	45	96.0	95.7	96.8	43	93.5	45	97.8
Newcastle	Royal Victoria Infirmary	59	185	172	8.3	7.9	101	89.3	89.7	91.8	104	100.8	101.1	96.5	102	92.7	102	92.7
Belfast	Royal Belfast Hospital for Sick Children	60	205	191	8.5	8.1	113	88.3	89.4	90.8	122	94.7	95.2	95.3	109	85.8	113	89.0
Nottingham	Nottingham University Hospitals	62	172	166	9.2	9.4	112	82.0	83.3	83.4	112	87.9	89.1	89.8	108	94.7	113	99.1
Teeside	James Cook University Hospital	71	56	54	9.5	9.5	37	83.3	85.5	84.6	38	89.0	90.9	89.6	34	89.5	37	97.4
Cardiff	Children's Hospital for Wales	72	168	161	8.8	9.0	117	84.9	84.8	86.6	117	86.4	86.2	89.0	30	25.2	117	98.3
Dundee	Ninewells Hospital	73	25	25	8.1	7.2	16	84.7	83.9	86.3	16	90.6	89.7	95.8	15	78.9	16	84.2
Aberdeen	Royal Aberdeen Children's Hospital	75	31	22	8.7	9.1	12	72.2	73.1	72.8	15	84.8	85.2	89.3	9	60.0	12	80.0
London - Central	Great Ormond Street Hospital for Children	90	202	190	8.1	7.3	114	87.8	87.8	88.4	115	94.5	94.4	94.2	111	93.3	117	98.3
Cornwall	Royal Cornwall Hospital	94	32	32	7.7	6.8	19	85.2	84.3	81.9	20	90.2	88.9	96.2	18	85.7	19	90.5
Exeter	Royal Devon & Exeter Hospital	96	72	70	9.2	9.2	45	81.7	83.4	87.9	45	85.9	87.4	89.1	38	80.9	45	95.7
Liverpool	Alder Hey Children's Hospital	97	318	299	8.6	8.1	183	82.8	83.9	83.8	187	90.6	91.6	91.7	174	90.6	183	95.3
Norwich	Norfolk & Norwich University Hospital	98	69	63	9.1	9.8	49	84.6	84.5	86.1	49	94.5	94.2	98.2	45	91.8	49	100.0
Birmingham	Birmingham Children's Hospital	104	305	285	8.8	8.8	181	82.5	83.6	84.7	183	90.1	91.1	91.3	177	95.7	182	98.4
Cambridge	Addenbrookes Hospital	107	141	129	7.8	7.2	80	89.5	88.6	92.8	81	94.0	92.9	95.7	80	97.6	80	97.6
Hull	Hull Royal Infirmary	111	36	35	7.5	6.4	17	74.2	73.6	85.6	18	80.0	79.3	86.3	17	94.4	17	94.4
Plymouth	Derriford Hospital	139	41	37	8.4	8.0	24	76.9	77.0	80.7	25	88.0	87.8	89.0	24	96.0	24	96.0
Edinburgh	Royal Hospital for Sick Children	143	131	128	8.9	9.0	93	90.6	90.0	90.7	94	103.6	103.1	93.0	57	60.0	93	97.9
Manchester	Royal Manchester Children's Hospital	144	335	308	9.1	8.7	194	82.1	83.6	83.6	196	87.5	88.8	88.6	176	83.8	196	93.3
Lanarkshire	Wishaw General Hospital	162	48	44	8.3	7.7	29	91.2	90.4	91.4	30	95.8	94.8	96.4	29	96.7	29	96.7
Ayr	University Hospital Crosshouse	170	26	26	8.7	8.6	17	87.7	88.5	87.9	17	97.6	98.2	101.8	16	94.1	17	100.0

			BMI percentile			
Location	**Name**	**Clinic ID**	**Number**	**Mean - unadjusted**	**Mean - adjusted**	**Median**
Leicester	Leicester Royal Infirmary	1	58	59.3	59.3	61.0
Sheffield	Sheffield Children's Hospital	3	122	52.3	52.0	53.5
Stoke-on-Trent	University Hospitals of North Midlands	8	59	49.3	49.3	47.5
London - South West	Royal Brompton Hospital	15	224	54.9	54.8	56.3
London - South East	King's College Hospital	17	162	51.3	51.2	50.5
Oxford	John Radcliffe Hospital	22	124	53.6	53.8	54.4
Leeds	St James's University Hospital	25	164	51.9	51.4	53.4
Southampton	Southampton General Hospital	29	165	51.5	51.6	47.4
London - East	Royal London Hospital	30	89	51.1	51.4	49.6
Inverness	Raigmore Hospital	31	10	58.0	57.9	61.5
Bristol	Bristol Royal Hospital for Children	32	162	48.7	48.7	49.5
Glasgow	Royal Hospital for Sick Children	56	62	52.9	52.9	51.8
Newcastle	Royal Victoria Infirmary	59	143	57.7	57.7	61.0
Belfast	Royal Belfast Hospital for Sick Children	60	155	56.4	56.4	57.8
Nottingham	Nottingham University Hospitals	62	133	51.2	51.3	56.5
Teeside	James Cook University Hospital	71	45	62.3	62.8	63.0
Cardiff	Children's Hospital for Wales	72	123	54.6	54.8	57.0
Dundee	Ninewells Hospital	73	19	43.7	43.3	44.7
Aberdeen	Royal Aberdeen Children's Hospital	75	19	47.9	48.2	39.9
London - Central	Great Ormond Street Hospital for Children	90	149	47.4	47.2	45.3
Cornwall	Royal Cornwall Hospital	94	28	71.4	71.0	77.9
Exeter	Royal Devon & Exeter Hospital	96	61	55.8	55.9	56.5
Liverpool	Alder Hey Children's Hospital	97	236	51.9	52.0	54.1
Norwich	Norfolk & Norwich University Hospital	98	43	65.2	65.7	77.4
Birmingham	Birmingham Children's Hospital	104	235	51.5	51.6	50.6
Cambridge	Addenbrookes Hospital	107	109	50.5	50.1	50.6
Hull	Hull Royal Infirmary	111	28	55.6	55.1	66.1
Plymouth	Derriford Hospital	139	25	54.5	54.2	60.7
Edinburgh	Royal Hospital for Sick Children	143	111	56.3	56.4	54.3
Manchester	Royal Manchester Children's Hospital	144	233	49.6	49.7	49.4
Lanarkshire	Wishaw General Hospital	162	32	52.5	52.3	55.0
Ayr	University Hospital Crosshouse	170	17	67.4	67.7	70.5

Chronic *pseudomonas*		Having at least 1 IV days		Receiving Dnase treatment		Receiving hypertonic saline treatment		Inhaled antibiotic use among patients with chronic *pseudomonas*	
Number	**Proportion (%)**	**Number**	**Proportion (%)**	**Number**	**Proportion (%)**	**Number**	**Proportion (%)**	**Number**	**Proportion (%)**
<5	3.0	23	34.8	45	68.2	<5	6.1	<5	100.0
6	4.3	48	34.8	83	60.1	42	30.4	5	83.3
6	6.4	52	55.3	62	66.0	21	22.3	6	100.0
19	6.4	95	32.0	209	70.4	112	37.7	18	94.7
11	5.4	81	39.7	126	61.8	62	30.4	10	90.9
12	6.8	74	41.8	126	71.2	66	37.3	8	66.7
9	4.2	88	41.3	116	54.5	17	8.0	9	100.0
13	6.3	59	28.4	136	65.4	35	16.8	11	84.6
8	6.3	59	46.5	94	74.0	92	72.4	8	100.0
<5	17.6	<5	23.5	<5	11.8	<5	5.9	<5	66.7
25	14.0	68	38.2	118	66.3	37	20.8	22	88.0
0	0.0	28	41.2	14	20.6	23	33.8	0	0.0
15	8.7	71	41.3	85	49.4	33	19.2	15	100.0
12	6.3	28	14.7	156	81.7	23	12.0	11	91.7
8	4.8	50	30.1	92	55.4	45	27.1	7	87.5
6	11.1	23	42.6	45	83.3	15	27.8	6	100.0
<5	2.5	43	26.7	129	80.1	111	68.9	<5	100.0
<5	12.0	6	24.0	6	24.0	11	44.0	<5	100.0
0	0.0	6	27.3	5	22.7	<5	4.5	0	0.0
15	7.9	88	46.3	121	63.7	73	38.4	13	86.7
<5	6.3	11	34.4	18	56.3	10	31.3	<5	100.0
<5	4.3	24	34.3	60	85.7	61	87.1	<5	100.0
24	8.0	129	43.1	138	46.2	46	15.4	24	100.0
5	7.9	24	38.1	41	65.1	19	30.2	5	100.0
12	4.2	85	29.8	133	46.7	60	21.1	10	83.3
10	7.8	34	26.4	82	63.6	54	41.9	10	100.0
<5	8.6	24	68.6	20	57.1	<5	11.4	<5	100.0
<5	5.4	10	27.0	17	45.9	<5	8.1	<5	100.0
8	6.3	30	23.4	67	52.3	16	12.5	7	87.5
32	10.4	103	33.4	131	42.5	137	44.5	30	93.8
<5	4.5	11	25.0	5	11.4	11	25.0	<5	100.0
0	0.0	6	23.1	5	19.2	6	23.1	0	0.0

Appendix 2: Data tables

Adult centres/clinics providing data in 2017 – ordered by clinic ID

					Age		FEV_1% predicted at annual review	
Location	Name	Clinic ID	Total Active	Number with annual review	Mean	Median	Number	Mean - unadjusted
London - South East	King's College Hospital	5	224	217	29.4	27.3	190	63.1
Newcastle	Royal Victoria Infirmary	9	287	284	31.0	28.6	258	64.5
London - South West	Royal Brompton Hospital	12	637	548	34.1	32.0	539	61.1
Belfast	Belfast City Hospital	14	281	236	35.1	32.0	231	66.7
Frimley	Frimley Park Hospital	19	133	131	32.6	32.0	115	62.1
Birmingham	Birmingham Heartlands Hospital	27	332	328	32.2	29.8	285	63.5
Exeter	Royal Devon & Exeter Hospital	34	104	99	32.4	27.8	92	68.0
Leeds	St James's University Hospital	42	399	396	33.1	31.3	337	59.7
Edinburgh	Western General Hospital	44	245	238	32.1	29.9	209	63.6
Cambridge	Papworth Hospital	51	320	306	30.4	27.8	276	66.8
Plymouth	Derriford Hospital	64	54	48	33.8	31.4	44	66.4
Sheffield	Northern General Hospital	65	208	200	30.0	27.9	179	68.9
Liverpool	Liverpool Heart and Chest Hospital	66	324	319	31.2	28.7	291	68.9
Llandough	Llandough Hospital	68	260	240	30.5	29.2	219	64.1
Aberdeen	Aberdeen Royal Infirmary	70	67	64	32.6	31.5	57	56.2
Stoke-on-Trent	University Hospitals of North Midlands	74	120	120	29.4	26.0	109	61.7
Glasgow	Gartnavel General Hospital	79	237	226	31.6	27.7	205	64.9
London - East	St. Bartholomew's Hospital	92	158	150	30.9	29.1	139	66.2
Nottingham	Nottingham University Hospitals	101	197	194	30.5	28.5	170	60.3
Manchester	Wythenshawe Hospital	102	431	405	31.8	29.9	391	60.2
London - South East	University Hospital Lewisham	105	58	56	30.9	28.4	41	54.2
Bristol	Bristol Royal Infirmary	106	223	209	30.7	29.0	193	69.0
Southampton	Southampton General Hospital	110	275	269	31.6	28.8	222	66.6
Norwich	Norfolk & Norwich University Hospital	114	75	72	28.4	26.1	54	66.7
Oxford	Churchill Hospital	128	126	121	30.9	27.5	93	60.9
Cornwall	Royal Cornwall Hospital	129	36	35	32.2	30.3	28	59.6
Leicester	Glenfield Hospital	142	83	79	31.6	27.4	71	63.0
York	York & Hull Adult CF Centre	171	74	73	32.1	28.2	68	61.7

FEV_1% predicted at annual review		Best FEV_1% predicted				Data completeness for FEV_1			
Mean - adjusted	Median	Number*	Mean - unadjusted	Mean - adjusted	Median	Number with valid Best FEV_1**	Percentage with valid Best FEV_1	Number with FEV_1 at annual review	Percentage with FEV_1 at annual review
62.3	64.1	190	66.4	65.4	69.8	198	91.2	201	92.6
63.9	66.0	258	69.5	68.7	72.0	280	98.6	281	98.9
62.0	60.0	539	65.4	66.6	64.6	545	99.5	546	99.6
67.5	69.1	231	70.7	71.7	73.6	207	87.7	235	99.6
62.8	62.4	116	67.3	68.1	70.4	109	83.2	124	94.7
63.7	62.3	291	71.3	71.6	68.1	300	91.5	302	92.1
67.9	69.3	93	72.5	72.4	76.1	94	94.9	96	97.0
60.3	61.1	338	68.2	69.0	66.8	381	96.2	382	96.5
63.4	64.0	209	70.8	70.6	70.3	221	92.9	227	95.4
66.0	66.5	279	72.2	71.2	73.3	233	76.1	297	97.1
67.0	66.4	45	75.7	76.4	76.8	44	91.7	45	93.8
68.3	71.2	189	73.0	72.3	77.5	184	92.0	186	93.0
68.9	69.0	293	75.0	75.0	77.2	272	85.3	298	93.4
63.8	66.7	221	70.7	70.2	73.7	235	97.9	236	98.3
56.8	50.2	57	60.8	61.5	57.8	50	78.1	64	100.0
60.5	63.2	113	66.9	65.5	69.8	112	93.3	114	95.0
64.6	65.3	211	70.9	70.5	72.4	207	91.6	210	92.9
66.0	67.6	140	68.8	68.6	71.2	126	84.0	145	96.7
59.9	58.1	170	67.4	66.9	66.0	165	85.1	191	98.5
60.7	59.6	391	61.3	61.9	60.7	392	96.8	401	99.0
54.3	53.6	46	57.5	57.6	57.4	50	89.3	50	89.3
69.0	69.4	195	75.3	75.3	76.4	187	89.5	201	96.2
66.0	67.8	235	72.5	71.8	76.1	228	84.8	237	88.1
65.8	71.3	65	73.1	71.9	71.9	57	79.2	57	79.2
60.2	57.8	94	66.2	65.4	64.5	103	85.1	104	86.0
60.0	60.4	29	64.1	64.4	63.0	31	88.6	31	88.6
63.3	61.0	71	66.7	67.2	66.6	76	96.2	78	98.7
61.4	60.0	68	66.8	66.5	66.7	72	98.6	72	98.6

* Where 'Best' values were missing, or lower than FEV_1% predicted taken at annual review, the annual review value was used.
** For data completeness, 'Best' values were taken to be valid if they were not missing and the percent predicted was not more than 0.5% lower than FEV_1% predicted taken at annual review

			BMI			
Location	Name	Clinic ID	Number	Mean - unadjusted	Mean - adjusted	Median
London - South East	King's College Hospital	5	203	22.6	22.9	22.1
Newcastle	Royal Victoria Infirmary	9	281	22.9	23.0	22.2
London - South West	Royal Brompton Hospital	12	546	22.5	22.2	22.4
Belfast	Belfast City Hospital	14	235	23.9	23.6	23.2
Frimley	Frimley Park Hospital	19	131	21.9	21.7	22.0
Birmingham	Birmingham Heartlands Hospital	27	313	22.7	22.6	22.7
Exeter	Royal Devon & Exeter Hospital	34	98	23.7	23.7	24.3
Leeds	St James's University Hospital	42	386	23.1	22.9	22.9
Edinburgh	Western General Hospital	44	229	22.7	22.6	22.5
Cambridge	Papworth Hospital	51	306	21.7	21.9	22.2
Plymouth	Derriford Hospital	64	48	24.7	24.5	24.3
Sheffield	Northern General Hospital	65	195	23.4	23.6	22.5
Liverpool	Liverpool Heart and Chest Hospital	66	310	23.5	23.5	22.7
Llandough	Llandough Hospital	68	240	22.5	22.7	22.3
Aberdeen	Aberdeen Royal Infirmary	70	64	23.7	23.5	22.6
Stoke-on-Trent	University Hospitals of North Midlands	74	118	23.0	23.3	22.3
Glasgow	Gartnavel General Hospital	79	218	22.7	22.8	22.7
London - East	St. Bartholomew's Hospital	92	145	22.7	22.7	22.1
Nottingham	Nottingham University Hospitals	101	193	22.2	22.3	21.6
Manchester	Wythenshawe Hospital	102	404	22.3	22.2	21.9
London - South East	University Hospital Lewisham	105	52	21.4	21.4	20.4
Bristol	Bristol Royal Infirmary	106	208	22.8	22.8	22.4
Southampton	Southampton General Hospital	110	255	22.8	22.9	22.0
Norwich	Norfolk & Norwich University Hospital	114	71	21.6	22.0	21.2
Oxford	Churchill Hospital	128	111	22.5	22.5	22.1
Cornwall	Royal Cornwall Hospital	129	32	22.4	22.3	21.8
Leicester	Glenfield Hospital	142	78	22.9	22.9	21.7
York	York & Hull Adult CF Centre	171	73	22.6	22.6	21.5

Chronic *pseudomonas*		Having at least 1 IV days		Receiving Dnase treatment		Receiving hypertonic saline treatment		Inhaled antibiotic use among patients with chronic *pseudomonas*	
Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)
77	35.5	111	51.2	168	77.4	79	36.4	74	96.1
108	38.0	134	47.2	149	52.5	43	15.1	101	93.5
290	52.9	264	48.2	477	87.0	198	36.1	264	91.0
100	42.4	80	33.9	163	69.1	52	22.0	92	92.0
59	45.0	61	46.6	107	81.7	37	28.2	56	94.9
169	51.5	184	56.1	234	71.3	104	31.7	160	94.7
35	35.4	46	46.5	68	68.7	56	56.6	26	74.3
182	46.0	243	61.4	310	78.3	105	26.5	167	91.8
88	37.0	101	42.4	101	42.4	43	18.1	58	65.9
118	38.6	183	59.8	175	57.2	122	39.9	101	85.6
16	33.3	25	52.1	18	37.5	29	60.4	15	93.8
98	49.0	120	60.0	168	84.0	46	23.0	91	92.9
151	47.3	154	48.3	150	47.0	78	24.5	127	84.1
93	38.8	118	49.2	164	68.3	127	52.9	88	94.6
27	42.2	26	40.6	27	42.2	23	35.9	26	96.3
54	45.0	69	57.5	78	65.0	47	39.2	51	94.4
92	40.7	88	38.9	109	48.2	30	13.3	85	92.4
56	37.3	74	49.3	116	77.3	88	58.7	52	92.9
92	47.4	133	68.6	129	66.5	74	38.1	71	77.2
237	58.5	239	59.0	248	61.2	139	34.3	214	90.3
23	41.1	29	51.8	38	67.9	18	32.1	23	100.0
64	30.6	121	57.9	107	51.2	91	43.5	55	85.9
103	38.3	151	56.1	153	56.9	94	34.9	87	84.5
20	27.8	35	48.6	46	63.9	27	37.5	19	95.0
43	35.5	71	58.7	81	66.9	45	37.2	40	93.0
13	37.1	21	60.0	22	62.9	11	31.4	11	84.6
26	32.9	44	55.7	46	58.2	20	25.3	24	92.3
35	47.9	40	54.8	54	74.0	12	16.4	33	94.3

Appendix 2: Data tables

Paediatric centres/clinics providing data in 2017 – ordered alphabetically by country/city

					Age		FEV_1% predicted at annual review	
Location	**Name**	**Clinic ID**	**Total Active**	**Number with annual review**	**Mean**	**Median**	**Number**	**Mean - unadjusted**
England								
Birmingham	Birmingham Children's Hospital	104	305	285	8.8	8.8	181	82.5
Bristol	Bristol Royal Hospital for Children	32	185	178	8.7	8.0	113	85.5
Cambridge	Addenbrookes Hospital	107	141	129	7.8	7.2	80	89.5
Cornwall	Royal Cornwall Hospital	94	32	32	7.7	6.8	19	85.2
Exeter	Royal Devon & Exeter Hospital	96	72	70	9.2	9.2	45	81.7
Hull	Hull Royal Infirmary	111	36	35	7.5	6.4	17	74.2
Leeds	St James's University Hospital	25	229	213	8.0	7.3	128	85.7
Leicester	Leicester Royal Infirmary	1	69	66	8.8	8.2	44	88.5
Liverpool	Alder Hey Children's Hospital	97	318	299	8.6	8.1	183	82.8
London - East	Royal London Hospital	30	130	127	9.3	9.1	94	91.4
London - South East	King's College Hospital	17	218	204	8.4	7.9	127	86.9
London - South West	Royal Brompton Hospital	15	315	297	8.6	8.1	187	85.3
London - Central	Great Ormond Street Hospital for Children	90	202	190	8.1	7.3	114	87.8
Manchester	Royal Manchester Children's Hospital	144	335	308	9.1	8.7	194	82.1
Newcastle	Royal Victoria Infirmary	59	185	172	8.3	7.9	101	89.3
Norwich	Norfolk & Norwich University Hospital	98	69	63	9.1	9.8	49	84.6
Nottingham	Nottingham University Hospitals	62	172	166	9.2	9.4	112	82.0
Oxford	John Radcliffe Hospital	22	184	177	9.0	9.0	123	89.7
Plymouth	Derriford Hospital	139	41	37	8.4	8.0	24	76.9
Sheffield	Sheffield Children's Hospital	3	142	138	8.5	8.3	89	91.5
Southampton	Southampton General Hospital	29	218	208	8.6	8.8	128	85.1
Stoke-on-Trent	University Hospitals of North Midlands	8	100	94	8.6	8.0	57	86.4
Teeside	James Cook University Hospital	71	56	54	9.5	9.5	37	83.3
Northern Ireland								
Belfast	Royal Belfast Hospital for Sick Children	60	205	191	8.5	8.1	113	88.3
Scotland								
Aberdeen	Royal Aberdeen Children's Hospital	75	31	22	8.7	9.1	12	72.2
Ayr	University Hospital Crosshouse	170	26	26	8.7	8.6	17	87.7
Dundee	Ninewells Hospital	73	25	25	8.1	7.2	16	84.7
Edinburgh	Royal Hospital for Sick Children	143	131	128	8.9	9.0	93	90.6
Glasgow	Royal Hospital for Sick Children	56	95	68	8.5	8.8	45	89.8
Inverness	Raigmore Hospital	31	18	17	8.0	8.9	10	88.4
Lanarkshire	Wishaw General Hospital	162	48	44	8.3	7.7	29	91.2
Wales								
Cardiff	Children's Hospital for Wales	72	168	161	8.8	9.0	117	84.9

FEV_1% predicted at annual review		Best FEV_1% predicted				Data completeness for FEV_1			
Mean - adjusted	**Median**	**Number***	**Mean - unadjusted**	**Mean - adjusted**	**Median**	**Number with valid Best FEV_1****	**Percentage with valid Best FEV_1**	**Number with FEV_1 at annual review**	**Percentage with FEV_1 at annual review**
83.6	84.7	183	90.1	91.1	91.3	177	95.7	182	98.4
86.9	83.7	114	93.7	94.9	92.6	103	88.8	113	97.4
88.6	92.8	81	94.0	92.9	95.7	80	97.6	80	97.6
84.3	81.9	20	90.2	88.9	96.2	18	85.7	19	90.5
83.4	87.9	45	85.9	87.4	89.1	38	80.9	45	95.7
73.6	85.6	18	80.0	79.3	86.3	17	94.4	17	94.4
85.2	88.8	130	93.3	92.7	95.6	130	94.2	130	94.2
89.3	89.1	44	93.5	94.2	93.3	43	95.6	44	97.8
83.8	83.8	187	90.6	91.6	91.7	174	90.6	183	95.3
92.2	91.0	95	96.2	96.8	95.2	73	75.3	94	96.9
87.9	88.6	127	91.9	92.8	94.3	122	95.3	127	99.2
85.9	87.7	188	93.1	93.7	93.4	158	83.6	187	98.9
87.8	88.4	115	94.5	94.4	94.2	111	93.3	117	98.3
83.6	83.6	196	87.5	88.8	88.6	176	83.8	196	93.3
89.7	91.8	104	100.8	101.1	96.5	102	92.7	102	92.7
84.5	86.1	49	94.5	94.2	98.2	45	91.8	49	100.0
83.3	83.4	112	87.9	89.1	89.8	108	94.7	113	99.1
90.3	93.3	124	93.0	93.4	95.6	82	66.1	123	99.2
77.0	80.7	25	88.0	87.7	89.0	24	96.0	24	96.0
91.2	93.2	91	98.3	98.0	98.7	87	92.6	89	94.7
86.1	88.8	131	91.8	92.8	95.1	125	93.3	128	95.5
86.2	89.0	64	91.0	91.0	93.5	49	72.1	57	83.8
85.5	84.6	38	89.0	90.8	89.6	34	89.5	37	97.4
89.4	90.8	122	94.7	95.2	95.3	109	85.8	113	89.0
73.1	72.8	15	84.8	85.2	89.3	9	60.0	12	80.0
88.4	87.9	17	97.6	98.2	101.8	16	94.1	17	100.0
83.9	86.3	16	90.6	89.7	95.8	15	78.9	16	84.2
90.0	90.7	94	103.6	103.0	93.0	57	60.0	93	97.9
89.7	91.5	45	96.0	95.7	96.8	43	93.5	45	97.8
88.5	88.1	10	94.3	94.4	94.0	10	90.9	10	90.9
90.4	91.4	30	95.8	94.8	96.4	29	96.7	29	96.7
84.8	86.6	117	86.4	86.2	89.0	30	25.2	117	98.3

* Where 'Best' values were missing, or lower than FEV_1% predicted taken at annual review, the annual review value was used.
** For data completeness, 'Best' values were taken to be valid if they were not missing and the percent predicted was not more than 0.5% lower than FEV_1% predicted taken at annual review

			BMI percentile			
Location	**Name**	**Clinic ID**	**Number**	**Mean - unadjusted**	**Mean - adjusted**	**Median**
England						
Birmingham	Birmingham Children's Hospital	104	235	51.5	51.6	50.6
Bristol	Bristol Royal Hospital for Children	32	162	48.7	48.7	49.5
Cambridge	Addenbrookes Hospital	107	109	50.5	50.1	50.6
Cornwall	Royal Cornwall Hospital	94	28	71.4	71.0	77.9
Exeter	Royal Devon & Exeter Hospital	96	61	55.8	55.9	56.5
Hull	Hull Royal Infirmary	111	28	55.6	55.1	66.1
Leeds	St James's University Hospital	25	164	51.9	51.4	53.4
Leicester	Leicester Royal Infirmary	1	58	59.3	59.3	61.0
Liverpool	Alder Hey Children's Hospital	97	236	51.9	52.0	54.1
London - East	Royal London Hospital	30	89	51.1	51.4	49.6
London - South East	King's College Hospital	17	162	51.3	51.2	50.5
London - South West	Royal Brompton Hospital	15	224	54.9	54.8	56.3
London - Central	Great Ormond Street Hospital for Children	90	149	47.4	47.2	45.3
Manchester	Royal Manchester Children's Hospital	144	233	49.6	49.7	49.4
Newcastle	Royal Victoria Infirmary	59	143	57.7	57.7	61.0
Norwich	Norfolk & Norwich University Hospital	98	43	65.2	65.7	77.4
Nottingham	Nottingham University Hospitals	62	133	51.2	51.3	56.5
Oxford	John Radcliffe Hospital	22	124	53.6	53.8	54.4
Plymouth	Derriford Hospital	139	25	54.5	54.2	60.7
Sheffield	Sheffield Children's Hospital	3	122	52.3	52.0	53.5
Southampton	Southampton General Hospital	29	165	51.5	51.6	47.4
Stoke-on-Trent	University Hospitals of North Midlands	8	59	49.3	49.3	47.5
Teeside	James Cook University Hospital	71	45	62.3	62.8	63.0
Northern Ireland						
Belfast	Royal Belfast Hospital for Sick Children	60	155	56.4	56.4	57.8
Scotland						
Aberdeen	Royal Aberdeen Children's Hospital	75	19	47.9	48.2	39.9
Ayr	University Hospital Crosshouse	170	17	67.4	67.7	70.5
Dundee	Ninewells Hospital	73	19	43.7	43.3	44.7
Edinburgh	Royal Hospital for Sick Children	143	111	56.3	56.4	54.3
Glasgow	Royal Hospital for Sick Children	56	62	52.9	52.9	51.8
Inverness	Raigmore Hospital	31	10	58.0	57.9	61.5
Lanarkshire	Wishaw General Hospital	162	32	52.5	52.3	55.0
Wales						
Cardiff	Children's Hospital for Wales	72	123	54.6	54.8	57.0

Chronic *pseudomonas*		Having at least 1 IV days		Receiving Dnase treatment		Receiving hypertonic saline treatment		Inhaled antibiotic use among patients with chronic *pseudomonas*	
Number	**Proportion (%)**	**Number**	**Proportion (%)**	**Number**	**Proportion (%)**	**Number**	**Proportion (%)**	**Number**	**Proportion (%)**
12	4.2	85	29.8	133	46.7	60	21.1	10	83.3
25	14.0	68	38.2	118	66.3	37	20.8	22	88.0
10	7.8	34	26.4	82	63.6	54	41.9	10	100.0
<5	6.3	11	34.4	18	56.3	10	31.3	<5	100.0
<5	4.3	24	34.3	60	85.7	61	87.1	<5	100.0
<5	8.6	24	68.6	20	57.1	<5	11.4	<5	100.0
9	4.2	88	41.3	116	54.5	17	8.0	9	100.0
<5	3.0	23	34.8	45	68.2	<5	6.1	<5	100.0
24	8.0	129	43.1	138	46.2	46	15.4	24	100.0
8	6.3	59	46.5	94	74.0	92	72.4	8	100.0
11	5.4	81	39.7	126	61.8	62	30.4	10	90.9
19	6.4	95	32.0	209	70.4	112	37.7	18	94.7
15	7.9	88	46.3	121	63.7	73	38.4	13	86.7
32	10.4	103	33.4	131	42.5	137	44.5	30	93.8
15	8.7	71	41.3	85	49.4	33	19.2	15	100.0
5	7.9	24	38.1	41	65.1	19	30.2	5	100.0
8	4.8	50	30.1	92	55.4	45	27.1	7	87.5
12	6.8	74	41.8	126	71.2	66	37.3	8	66.7
<5	5.4	10	27.0	17	45.9	<5	8.1	<5	100.0
6	4.3	48	34.8	83	60.1	42	30.4	5	83.3
13	6.3	59	28.4	136	65.4	35	16.8	11	84.6
6	6.4	52	55.3	62	66.0	21	22.3	6	100.0
6	11.1	23	42.6	45	83.3	15	27.8	6	100.0
12	6.3	28	14.7	156	81.7	23	12.0	11	91.7
0	0.0	6	27.3	5	22.7	<5	4.5	0	0.0
0	0.0	6	23.1	5	19.2	6	23.1	0	0.0
<5	12.0	6	24.0	6	24.0	11	44.0	<5	100.0
8	6.3	30	23.4	67	52.3	16	12.5	7	87.5
0	0.0	28	41.2	14	20.6	23	33.8	0	0.0
<5	17.6	<5	23.5	<5	11.8	<5	5.9	<5	66.7
<5	4.5	11	25.0	5	11.4	11	25.0	<5	100.0
<5	2.5	43	26.7	129	80.1	111	68.9	<5	100.0

Appendix 2: Data tables

Adult centres/clinics providing data in 2017 – ordered alphabetically by country/city

					Age		FEV_1% predicted at annual review	
Location	**Name**	**Clinic ID**	**Total Active**	**Number with annual review**	**Mean**	**Median**	**Number**	**Mean - unadjusted**
England								
Birmingham	Birmingham Heartlands Hospital	27	332	328	32.2	29.8	285	63.5
Bristol	Bristol Royal Infirmary	106	223	209	30.7	29.0	193	69.0
Cambridge	Papworth Hospital	51	320	306	30.4	27.8	276	66.8
Cornwall	Royal Cornwall Hospital	129	36	35	32.2	30.3	28	59.6
Exeter	Royal Devon & Exeter Hospital	34	104	99	32.4	27.8	92	68.0
Frimley	Frimley Park Hospital	19	133	131	32.6	32.0	115	62.1
Leeds	St James's University Hospital	42	399	396	33.1	31.3	337	59.7
Leicester	Glenfield Hospital	142	83	79	31.6	27.4	71	63.0
Liverpool	Liverpool Heart and Chest Hospital	66	324	319	31.2	28.7	291	68.9
London - East	St. Bartholomew's Hospital	92	158	150	30.9	29.1	139	66.2
London - South East	King's College Hospital	5	224	217	29.4	27.3	190	63.1
London - South East	University Hospital Lewisham	105	58	56	30.9	28.4	41	54.2
London - South West	Royal Brompton Hospital	12	637	548	34.1	32.0	539	61.1
Manchester	Wythenshawe Hospital	102	431	405	31.8	29.9	391	60.2
Newcastle	Royal Victoria Infirmary	9	287	284	31.0	28.6	258	64.5
Norwich	Norfolk & Norwich University Hospital	114	75	72	28.4	26.1	54	66.7
Nottingham	Nottingham University Hospitals	101	197	194	30.5	28.5	170	60.3
Oxford	Churchill Hospital	128	126	121	30.9	27.5	93	60.9
Plymouth	Derriford Hospital	64	54	48	33.8	31.4	44	66.4
Sheffield	Northern General Hospital	65	208	200	30.0	27.9	179	68.9
Southampton	Southampton General Hospital	110	275	269	31.6	28.8	222	66.6
Stoke-on-Trent	University Hospitals of North Midlands	74	120	120	29.4	26.0	109	61.7
York	York & Hull Adult CF Centre	171	74	73	32.1	28.2	68	61.7
Northern Ireland								
Belfast	Belfast City Hospital	14	281	236	35.1	32.0	231	66.7
Scotland								
Aberdeen	Aberdeen Royal Infirmary	70	67	64	32.6	31.5	57	56.2
Edinburgh	Western General Hospital	44	245	238	32.1	29.9	209	63.6
Glasgow	Gartnavel General Hospital	79	237	226	31.6	27.7	205	64.9
Wales								
Llandough	Llandough Hospital	68	260	240	30.5	29.2	219	64.1

FEV_1% predicted at annual review		Best FEV_1% predicted				Data completeness for FEV_1			
Mean - adjusted	**Median**	**Number***	**Mean - unadjusted**	**Mean - adjusted**	**Median**	**Number with valid Best FEV_1****	**Percentage with valid Best FEV_1**	**Number with FEV_1 at annual review**	**Percentage with FEV_1 at annual review**
63.7	62.3	291	71.3	71.6	68.1	300	91.5	302	92.1
69.0	69.4	195	75.3	75.3	76.4	187	89.5	201	96.2
66.0	66.5	279	72.2	71.2	73.3	233	76.1	297	97.1
60.0	60.4	29	64.1	64.4	63.0	31	88.6	31	88.6
67.9	69.3	93	72.5	72.4	76.1	94	94.9	96	97.0
62.8	62.4	116	67.3	68.1	70.4	109	83.2	124	94.7
60.3	61.1	338	68.2	69.0	66.8	381	96.2	382	96.5
63.3	61.0	71	66.7	67.2	66.6	76	96.2	78	98.7
68.9	69.0	293	75.0	75.0	77.2	272	85.3	298	93.4
66.0	67.6	140	68.8	68.6	71.2	126	84.0	145	96.7
62.3	64.1	190	66.4	65.4	69.8	198	91.2	201	92.6
54.3	53.6	46	57.5	57.6	57.4	50	89.3	50	89.3
62.0	60.0	539	65.4	66.6	64.6	545	99.5	546	99.6
60.7	59.6	391	61.3	61.9	60.7	392	96.8	401	99.0
63.9	66.0	258	69.5	68.7	72.0	280	98.6	281	98.9
65.8	71.3	65	73.1	71.9	71.9	57	79.2	57	79.2
59.9	58.1	170	67.4	66.9	66.0	165	85.1	191	98.5
60.2	57.8	94	66.2	65.4	64.5	103	85.1	104	86.0
67.0	66.4	45	75.7	76.4	76.8	44	91.7	45	93.8
68.3	71.2	189	73.0	72.3	77.5	184	92.0	186	93.0
66.0	67.8	235	72.5	71.8	76.1	228	84.8	237	88.1
60.5	63.2	113	66.9	65.5	69.8	112	93.3	114	95.0
61.4	60.0	68	66.8	66.5	66.7	72	98.6	72	98.6
67.5	69.1	231	70.7	71.7	73.6	207	87.7	235	99.6
56.8	50.2	57	60.8	61.5	57.8	50	78.1	64	100.0
63.4	64.0	209	70.8	70.6	70.3	221	92.9	227	95.4
64.6	65.3	211	70.9	70.5	72.4	207	91.6	210	92.9
63.8	66.7	221	70.7	70.2	73.7	235	97.9	236	98.3

* Where 'Best' values were missing, or lower than FEV_1% predicted taken at annual review, the annual review value was used.
** For data completeness, 'Best' values were taken to be valid if they were not missing and the percent predicted was not more than 0.5% lower than FEV_1% predicted taken at annual review

			BMI			
Location	**Name**	**Clinic ID**	**Number**	**Mean - unadjusted**	**Mean - adjusted**	**Median**
England						
Birmingham	Birmingham Heartlands Hospital	27	313	22.7	22.6	22.7
Bristol	Bristol Royal Infirmary	106	208	22.8	22.8	22.4
Cambridge	Papworth Hospital	51	306	21.7	21.9	22.2
Cornwall	Royal Cornwall Hospital	129	32	22.4	22.3	21.8
Exeter	Royal Devon & Exeter Hospital	34	98	23.7	23.7	24.3
Frimley	Frimley Park Hospital	19	131	21.9	21.7	22.0
Leeds	St James's University Hospital	42	386	23.1	22.9	22.9
Leicester	Glenfield Hospital	142	78	22.9	22.9	21.7
Liverpool	Liverpool Heart and Chest Hospital	66	310	23.5	23.5	22.7
London - East	St. Bartholomew's Hospital	92	145	22.7	22.7	22.1
London - South East	King's College Hospital	5	203	22.6	22.9	22.1
London - South East	University Hospital Lewisham	105	52	21.4	21.4	20.4
London - South West	Royal Brompton Hospital	12	546	22.5	22.2	22.4
Manchester	Wythenshawe Hospital	102	404	22.3	22.2	21.9
Newcastle	Royal Victoria Infirmary	9	281	22.9	23.0	22.2
Norwich	Norfolk & Norwich University Hospital	114	71	21.6	22.0	21.2
Nottingham	Nottingham University Hospitals	101	193	22.2	22.3	21.6
Oxford	Churchill Hospital	128	111	22.5	22.5	22.1
Plymouth	Derriford Hospital	64	48	24.7	24.5	24.3
Sheffield	Northern General Hospital	65	195	23.4	23.6	22.5
Southampton	Southampton General Hospital	110	255	22.8	22.9	22.0
Stoke-on-Trent	University Hospitals of North Midlands	74	118	23.0	23.3	22.3
York	York & Hull Adult CF Centre	171	73	22.6	22.6	21.5
Northern Ireland						
Belfast	Belfast City Hospital	14	235	23.9	23.6	23.2
Scotland						
Aberdeen	Aberdeen Royal Infirmary	70	64	23.7	23.5	22.6
Edinburgh	Western General Hospital	44	229	22.7	22.6	22.5
Glasgow	Gartnavel General Hospital	79	218	22.7	22.8	22.7
Wales						
Llandough	Llandough Hospital	68	240	22.5	22.7	22.3

Chronic *pseudomonas*		Having at least 1 IV days		Receiving Dnase treatment		Receiving hypertonic saline treatment		Inhaled antibiotic use among patients with chronic *pseudomonas*	
Number	**Proportion (%)**	**Number**	**Proportion (%)**	**Number**	**Proportion (%)**	**Number**	**Proportion (%)**	**Number**	**Proportion (%)**
169	51.5	184	56.1	234	71.3	104	31.7	160	94.7
64	30.6	121	57.9	107	51.2	91	43.5	55	85.9
118	38.6	183	59.8	175	57.2	122	39.9	101	85.6
13	37.1	21	60.0	22	62.9	11	31.4	11	84.6
35	35.4	46	46.5	68	68.7	56	56.6	26	74.3
59	45.0	61	46.6	107	81.7	37	28.2	56	94.9
182	46.0	243	61.4	310	78.3	105	26.5	167	91.8
26	32.9	44	55.7	46	58.2	20	25.3	24	92.3
151	47.3	154	48.3	150	47.0	78	24.5	127	84.1
56	37.3	74	49.3	116	77.3	88	58.7	52	92.9
77	35.5	111	51.2	168	77.4	79	36.4	74	96.1
23	41.1	29	51.8	38	67.9	18	32.1	23	100.0
290	52.9	264	48.2	477	87.0	198	36.1	264	91.0
237	58.5	239	59.0	248	61.2	139	34.3	214	90.3
108	38.0	134	47.2	149	52.5	43	15.1	101	93.5
20	27.8	35	48.6	46	63.9	27	37.5	19	95.0
92	47.4	133	68.6	129	66.5	74	38.1	71	77.2
43	35.5	71	58.7	81	66.9	45	37.2	40	93.0
16	33.3	25	52.1	18	37.5	29	60.4	15	93.8
98	49.0	120	60.0	168	84.0	46	23.0	91	92.9
103	38.3	151	56.1	153	56.9	94	34.9	87	84.5
54	45.0	69	57.5	78	65.0	47	39.2	51	94.4
35	47.9	40	54.8	54	74.0	12	16.4	33	94.3
100	42.4	80	33.9	163	69.1	52	22.0	92	92.0
27	42.2	26	40.6	27	42.2	23	35.9	26	96.3
88	37.0	101	42.4	101	42.4	43	18.1	58	65.9
92	40.7	88	38.9	109	48.2	30	13.3	85	92.4
93	38.8	118	49.2	164	68.3	127	52.9	88	94.6

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