Cystic Fibrosis strength in numbers

UK Cystic Fibrosis Registry
Annual Data Report 2016

Published August 2017

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

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Acknowledgements

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Foreword

I am delighted to write my first UK CF Registry Annual Report Foreword as Chief Executive of the Cystic Fibrosis Trust, on this, the 10th anniversary of the UK CF Registry being hosted here. It is fantastic to see the improvements in the health and outcomes of people with cystic fibrosis (CF) compared to the 2015 report.

The UK CF Registry is a vital tool for the CF community in working toward a life unlimited for people with cystic fibrosis. It stimulates ground-breaking research, supports clinical teams to improve care, monitors the safety of new medicines, and, through this report, gives people with CF and their families up-to-date information about the condition.

The UK CF Registry is also playing a vital role in advancing clinical trials, and answering questions about CF care that may otherwise go unanswered. Traditional clinical trials are expensive to run and slow to set up. Running clinical studies within the UK CF Registry, in specially designed modules, is faster and more cost effective. In 2016 the UK CF Registry launched three CF Registry-based studies, involving people with CF who consent to take part. The studies are looking at antibiotic prescribing in young children, monitoring people with two copies of the DF508 mutation, and collecting quality of life information direct from adults with cystic fibrosis.

In 2016 the UK CF Registry moved to a new software platform, and we are already seeing the benefits of this cutting-edge technology. Overall data completeness is now 95%, covering 9,695 people with cystic fibrosis. 119 people with CF are recorded as having a child in 2016. More information about fertility and family planning in CF is available from cysticfibrosis.org.uk/fertility or through our helpline by calling 020 3795 2184 or emailing helpline@cysticfibrosis.org.uk.

I would like to give my thanks to people with CF and their families, whose consent to participate in the CF Registry makes all of this invaluable work possible. I would also like to express my gratitude to CF teams across the UK who work extraordinarily hard to ensure that accurate and complete data are provided to the UK CF Registry every year.

David Ramsden
Chief Executive

Executive summary

This 2016 UK CF Registry report is the first to be generated since the move to our new software platform at the beginning of 2016. I must thank all of the CF centre staff and central CF Registry team for making this transition so smooth and successful.

Keen eyed people will notice a drop in overall numbers registered to 10,461. This does not reflect a drop in new cases of people with CF, but is a result of quality improvement work where we have clarified the current status of those registered but not having an annual review recorded in 2015 and 2016. In fact, there continues to be an increase in the total number of people in the UK living with CF, with more people receiving a new diagnosis of CF each year than dying of the disease. This is reflected in the increase in those with a 'complete' data set entered for 2016, to 9,695. It is from this 'complete' data set that the results for this report are generated. A reminder that the newly diagnosed numbers from the previous year's report are updated in the summary table, showing that there were 300 newly diagnosed people in 2015 This appears fairly stable over the last few years.

The proportion of those over 16 years of age continues to increase, breaking through the 60% barrier in this report. This does not reflect the distribution between adult and paediatric CF clinics but is the cut-off value assigned in the UK Registry report for an 'adult'. The median age of death has also increased this year to 31 years, although this figure may fluctuate when only a yearly snapshot is used. The survival calculation used in fig 1.32 now comes into line with other international CF Registries and gives 47 years as the median predicted survival of a child born today with CF, with males looking as if they do better than females. This calculation does not account for the new generation of CFTR modifiers that are coming through and are likely to have a significant effect on this over the coming years.

The median age of diagnosis is two months, reflecting the impact of Newborn Screening (NBS) and the success of picking up cases early. It should be noted that the NBS programme does not pick up all cases; of the 295 children who were five years old (diagnosed in the era of NBS), 7.4% were diagnosed following a clinical presentation (table 1.9). There were also 16 adults newly diagnosed in 2016.

The proportion of those with chronic *Pseudomonas aeruginosa* (defined on the registry as three or more growths in the preceding 12 months) continues to decrease a little, with the over 16s now standing at 44.2%, down from 48.2% in 2014. The under 16 age group remains static at 6.4%. The intermittent rate also appears to be dropping with 12.6% in this age range having a growth of *Pseudomonas aeruginosa* reported. There appears to be a continued improvement in the proportion (87.6%) receiving an inhaled antibiotic for chronic infection. Prevalence of non-tuberculous mycobacterium (NTM) recorded in the previous year appears to remain stable at 5.8% with a slight trend towards a higher proportion starting treatment for this infection; 58.7% are reported as currently on treatment.

Transplant numbers have been static over the last few years with only 46 adults receiving a lung transplant in 2016. Referrals for transplant remain similar to the last few years at 221 despite an increasing population. There also appears to be a drop in numbers being accepted onto transplant lists at; 96 in 2016.

Trends in types of physiotherapy performed continue to change with the more traditional active cycle of breathing techniques dropping from 29% in 2015 to 14% this year. 57% of people included exercise as one of their physiotherapy options, up from 37%.

For the first time we are presenting centre-level data for best FEV_1 in the age adjusted funnel plots. This data field is now 92% complete and brings our reporting into line with the European CF Registry. With a median FEV_1 of 78.8%, it is 4% higher than that recorded at annual review.

These are a few highlights of this year's report, we hope you find it a useful resource and we will endeavour to enhance the depth of reporting into the future.

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.

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

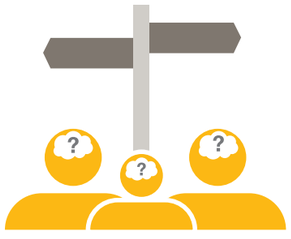
Cystic fibrosis

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. 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:

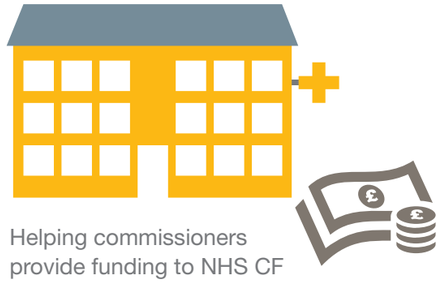


Helping people with CF and their families understand CF, and make informed decisions.

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 legislation like the Data Protection Act 1998, and its Research Ethics Committee-approved Study Protocol. It also makes recommendations about the future development of the Registry. A separate sub-committee of the RSC, the 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**.

i A glossary of terms highlighted in teal in this report can be found on page 49.

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

	2012	2013	2014	2015	2016
CF patients registered[1]	10078	10338	10583	10810	10461
Excluding diagnoses that year	9804	10076	10356	10586	10214
CF patients with 'complete' data; n (%)[2]	8794 (90%)	9052 (90%)	9432 (91%)	9587 (91%)	9695 (95%)
Age in years; median[3]	18	18	19	19	20
All newly diagnosed patients (NBS and other)	285	301	291	300	247[4]
Number diagnosed by NBS	213	177	164	168	180[4]
Age at diagnosis in months; median[3]	3	3	2	2	2
Adults aged 16 years and over; %[3]	57.6	57.6	59.3	59.9	60.4
Males; %[3]	52.9	52.9	53.0	53.0	53.2
Genotyped; %[3]	96.2	97.2	97.7	98.1	98.4
Total deaths reported (%)[5]	106 (1.1%)	146 (1.4%)	132 (1.2%)	125 (1.2%)	148 (1.5%)
Age at death in years; median (95% CI)[5]	28 (25, 29)	29 (27, 31)	28 (25.5, 32)	28 (27, 33)	31 (29, 33)

Complete data: Patients with at least the minimum data entered at their annual review.

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 has 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] A patient has 'complete data' if their team has filled in an annual review for them for that year. As patients newly diagnosed in a given year may not have their first annual review in the same year, the proportion with complete data is calclulated from the total registered excluding those diagnosed in the given year.

[3] Calculated from patients with complete data in the given year (see footnote 2 above).

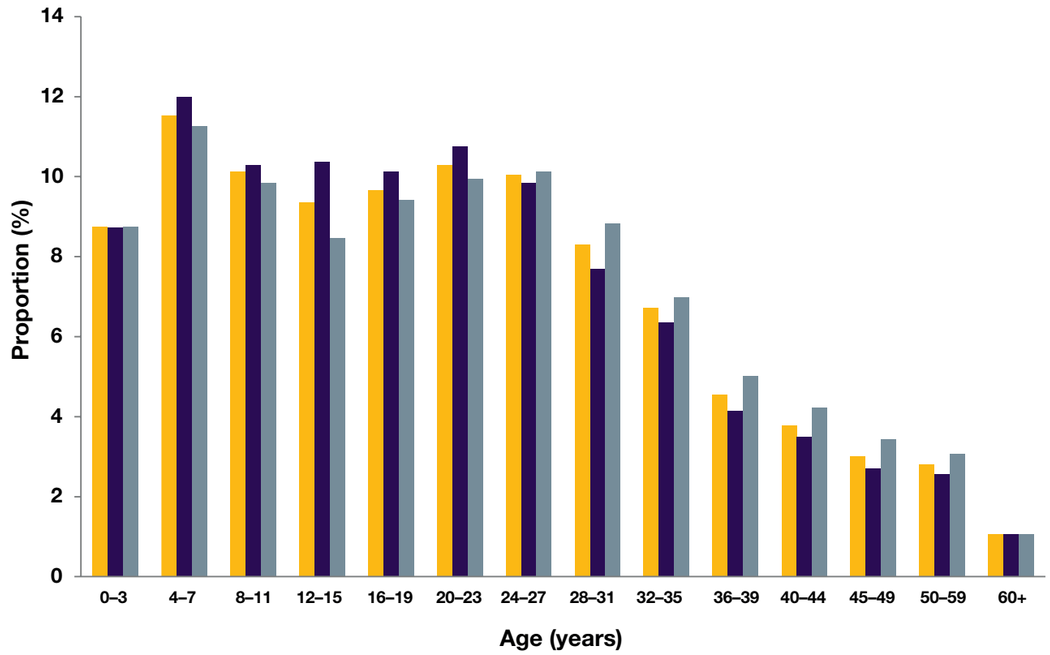
[4] Calculated from all patients registered on the database. Some diagnosis data are added after the data entry closure each year, so figures from previous years have been updated for this report.

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

1.2 Age distribution by gender

n=9695

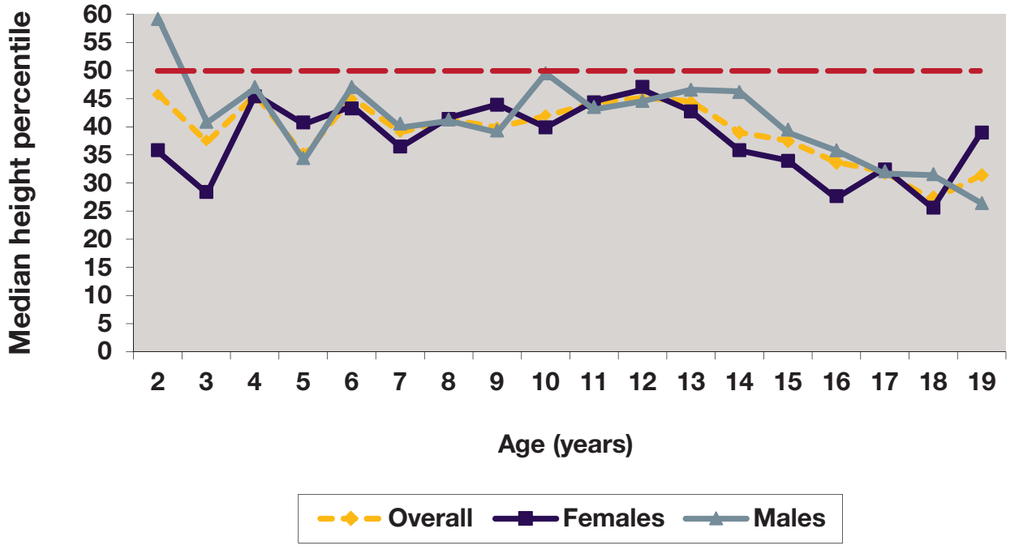
The following chart shows the mix of ages and genders in the CF population in the UK.



Age	All; n (%)	Females; n (%)	Males; n (%)
0-3	844 (8.7)	394 (8.7)	450 (8.7)
4-7	1119 (11.5)	542 (11.9)	577 (11.2)
8-11	975 (10.1)	467 (10.3)	508 (9.8)
12-15	906 (9.3)	470 (10.4)	436 (8.5)
16-19	943 (9.7)	459 (10.1)	484 (9.4)
20-23	998 (10.3)	486 (10.7)	512 (9.9)
24-27	965 (10.0)	446 (9.8)	519 (10.1)
28-31	802 (8.3)	350 (7.7)	452 (8.8)
32-35	648 (6.7)	289 (6.4)	359 (7.0)
36-39	443 (4.6)	187 (4.1)	256 (5.0)
40-44	373 (3.8)	157 (3.5)	216 (4.2)
45-49	295 (3.0)	121 (2.7)	174 (3.4)
50-59	275 (2.8)	116 (2.6)	159 (3.1)
60+	109 (1.1)	52 (1.1)	57 (1.1)
Overall	**9695**	**4536**	**5159**

1.3 Median height percentiles of children and young people (<20 years)[6] n=4457

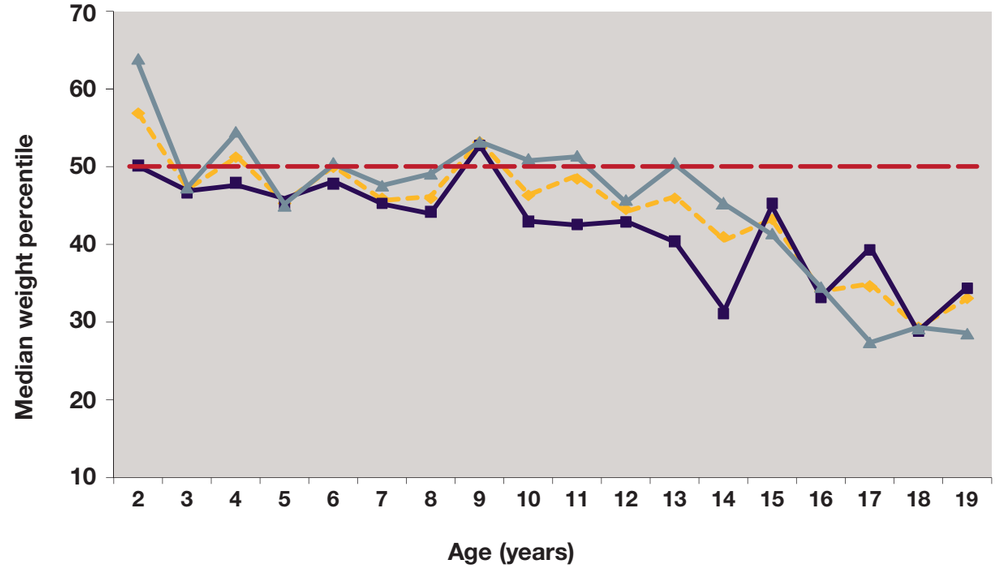
The following chart and table show the height percentiles of people with CF, aged 19 and under, in relation to UK growth data for the general population. If a person with CF is on the 40th percentile, only 40% of people the same age are their height or shorter; 60% are taller.



	Overall			Female			Male		
Age	n	Median	IQR	n	Median	IQR	n	Median	IQR
2	250	46.0	20.5-75.0	118	35.9	15.2-64.8	132	59.3	30.9-77.2
3	264	37.6	16.7-67.7	125	28.6	13.9-64.9	139	41.0	20.4-69.1
4	283	45.9	20.0-69.2	139	45.9	21.1-72.4	144	47.2	15.2-68.0
5	292	35.4	18.2-61.3	140	40.8	16.5-62.3	152	34.5	20.1-60.8
6	281	45.6	22.8-68.9	128	44.0	21.0-69.9	153	47.4	26.7-67.2
7	263	39.5	16.2-63.1	135	36.7	13.6-64.9	128	40.3	19.6-62.6
8	261	41.8	18.3-64.8	124	41.9	19.6-66.1	137	41.4	17.3-63.6
9	259	40.2	17.0-70.3	130	44.3	16.2-74.0	129	39.4	17.8-68.0
10	224	42.2	18.6-67.8	111	40.2	18.9-66.3	113	49.9	18.3-69.2
11	231	44.5	24.0-67.8	102	44.9	21.6-61.7	129	43.4	25.5-73.1
12	245	45.4	19.8-70.0	139	46.8	21.1-69.6	106	44.9	18.0-70.3
13	211	44.9	14.6-72.5	103	43.1	17.5-72.0	108	47.0	12.4-73.7
14	207	39.3	15.4-66.0	103	36.3	11.3-62.4	104	46.7	19.0-70.3
15	243	37.9	12.5-65.2	125	34.3	12.6-65.2	118	39.3	11.6-66.8
16	216	34.2	14.6-66.1	102	27.7	9.8-60.4	114	36.2	18.6-69.5
17	232	32.5	8.0-59.6	114	33.1	7.9-59.6	118	32.0	10.5-60.7
18	240	27.6	9.1-56.1	120	25.9	9.1-59.3	120	31.9	9.1-51.3
19	255	31.8	9.1-59.0	123	39.3	7.7-67.0	132	26.9	9.2-53.9
Overall	**4457**	**39.8**	**16.2-67.0**	**2181**	**39.1**	**15.4-66.1**	**2276**	**41.0**	**17.3-67.6**

1.4 Median weight percentiles of children and young people (<20 years)[6] n=4457

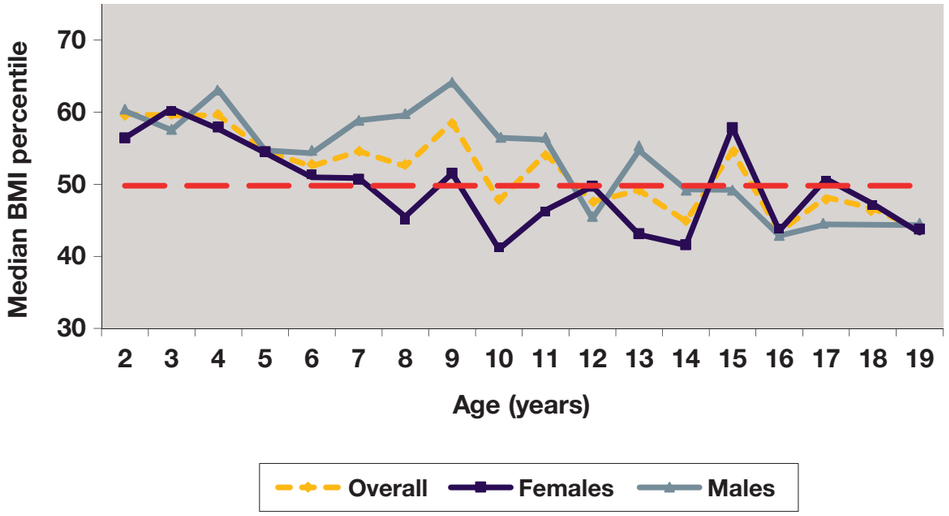
The following chart and table show the weight of people with CF, aged 19 and under, in relation to the UK growth data for the general population. If a person with CF is on the 40th percentile, only 40% of people the same age are their weight or lower; 60% weigh more.



	Overall			Female			Male		
Age	n	Median	IQR	n	Median	IQR	n	Median	IQR
2	250	57.4	31.6-80.7	118	50.5	25.7-76.9	132	63.9	40.2-81.3
3	264	47.7	22.4-74.7	125	47.4	17.7-74.3	139	47.9	25.4-74.7
4	283	51.9	28.0-74.3	139	48.1	29.1-74.3	144	55.1	25.6-74.6
5	292	45.8	21.8-69.4	140	46.4	21.8-71.8	152	45.7	21.8-69.2
6	281	50.5	24.2-76.2	128	48.7	23.0-76.8	153	50.7	25.3-76.2
7	263	46.2	21.6-72.5	135	45.5	18.9-69.6	128	48.0	25.3-77.5
8	261	46.5	21.7-73.9	124	44.3	20.0-66.8	137	49.5	24.9-76.1
9	259	53.8	23.8-78.1	130	53.3	20.8-80.0	129	53.8	24.8-76.5
10	224	46.9	23.3-70.8	111	43.5	20.8-65.5	113	51.3	26.9-74.7
11	231	49.4	26.7-76.0	102	43.0	24.5-73.3	129	51.9	36.0-79.1
12	245	44.6	23.4-72.9	139	43.5	20.7-75.6	106	45.9	25.6-70.9
13	211	46.5	20.4-73.7	103	40.7	17.3-72.1	108	50.8	22.5-76.4
14	207	40.9	15.4-68.6	103	31.7	10.5-63.9	104	45.5	23.1-72.7
15	243	43.6	18.1-75.7	125	45.4	21.3-76.8	118	41.6	16.8-75.1
16	216	34.3	11.2-64.6	102	33.5	7.8-63.7	114	34.8	15.0-68.4
17	232	35.3	8.0-63.1	114	39.8	15.0-59.3	118	27.5	4.5-68.8
18	240	29.4	6.9-62.4	120	28.9	6.8-64.4	120	29.4	7.4-61.5
19	255	33.3	6.8-62.3	123	34.8	9.9-63.3	132	28.8	5.8-60.8
Overall	**4457**	**45.2**	**19.3-72.5**	**2181**	**43.1**	**18.1-71.1**	**2276**	**46.8**	**20.1-73.9**

1.5 Median Body Mass Index (BMI) percentiles in children and young people (<20 years)[6]
n=4457

The following chart and table show the BMI percentiles of people with CF, aged 19 and under, in relation to the target BMI percentile for a healthy person of the same age; the 50th percentile; or the BMI percentile that half of the UK population people of that age has achieved. 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.

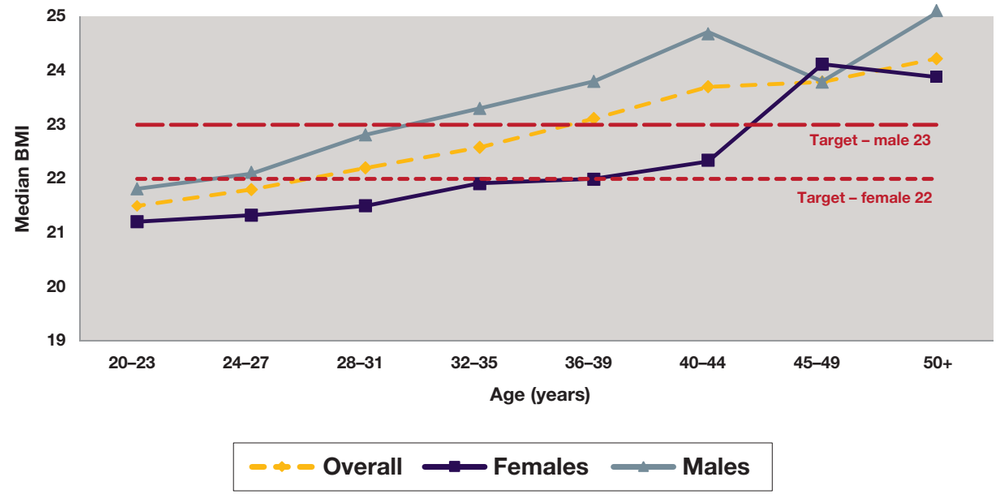


	Overall			Female			Male		
Age	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**
2	250	59.6	34.6-79.0	118	56.5	33.2-78.8	132	60.4	35.6-81.0
3	264	59.0	35.6-77.6	125	60.1	35.8-79.1	139	57.5	35.5-76.8
4	283	59.5	37.2-80.2	139	57.7	35.6-76.9	144	63.0	37.3-81.8
5	292	54.8	34.6-72.3	140	54.7	36.1-70.0	152	55.3	32.5-75.6
6	281	52.5	31.7-77.8	128	50.8	33.8-77.9	153	54.3	24.8-77.6
7	263	54.5	30.3-76.6	135	50.7	29.7-68.4	128	58.5	31.3-80.7
8	261	52.4	29.8-76.8	124	44.7	25.4-70.8	137	59.6	32.3-79.8
9	259	58.4	30.3-81.1	130	52.2	26.3-80.2	129	63.9	35.4-81.6
10	224	47.6	24.9-73.0	111	41.3	22.1-72.3	113	56.2	28.6-73.9
11	231	53.9	33.1-81.5	102	45.8	26.4-79.9	129	56.5	36.0-81.9
12	245	47.6	22.8-72.7	139	50.1	21.7-75.2	106	45.1	26.2-68.8
13	211	49.2	25.9-76.6	103	43.2	21.0-68.5	108	54.8	33.2-77.1
14	207	44.9	24.4-71.5	103	41.7	24.4-67.6	104	49.3	25.6-72.6
15	243	54.8	25.9-83.8	125	58.4	34.5-85.9	118	49.4	21.5-80.2
16	216	43.0	18.8-72.1	102	43.0	19.5-71.3	114	42.7	18.7-72.9
17	232	48.0	17.3-76.6	114	50.8	19.9-78.8	118	44.5	8.9-73.6
18	240	46.4	18.5-71.4	120	46.8	20.4-69.8	120	44.5	17.0-76.6
19	255	43.3	14.9-71.2	123	43.3	12.3-72.2	132	44.1	16.6-69.3
Overall	**4457**	**52.2**	**27.5-76.9**	**2181**	**49.9**	**26.8-75.9**	**2276**	**54.2**	**27.9-77.4**

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

1.6 Median Body Mass Index (BMI) in adults (20 years and over)
n=4908

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	998	21.5	19.5-23.6	486	21.2	19.3-23.2	512	21.8	19.7-23.9
24-27	965	21.8	19.8-24.1	446	21.4	19.5-23.6	519	22.1	20.1-24.5
28-31	802	22.2	20.1-24.7	350	21.5	19.7-24.0	452	22.8	20.7-25.1
32-35	648	22.6	20.7-25.1	289	21.9	20.1-24.4	359	23.3	21.4-25.7
36-39	443	23.1	21.0-25.2	187	22.0	20.0-24.2	256	23.8	21.7-25.6
40-44	373	23.7	21.3-26.5	157	22.3	20.3-25.8	216	24.7	22.4-26.9
45-49	295	23.8	21.8-26.1	121	24.2	21.2-26.9	174	23.8	22.1-25.7
50+	384	24.2	22.3-27.2	168	23.9	21.0-26.7	216	25.0	22.7-27.7
Overall	**4908**	**22.4**	**20.3-25.0**	**2204**	**21.8**	**19.8-24.3**	**2704**	**23.0**	**20.9-25.4**

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

1.7 Education and employment in adults (16 years and over) n=5851

The following table shows how people with CF reported their education and employment status in 2016. Please note that the groups are not mutually exclusive; someone may be a student as well as working part-time, for example.

	2012	2013	2014	2015	2016
Number of Patients n (%)	**5062**	**5213**	**5592**	**5742**	**5851**
Number who completed questionnaire	4254 (84.0)	4346 (83.4)	4623 (82.7)	4930 (85.9)	5791 (99.0)
Full-time employment	1425 (28.2)	1502 (28.8)	1634 (29.2)	1811 (31.5)	1887 (32.2)
Part-time employment	653 (12.9)	664 (12.7)	703 (12.6)	768 (13.4)	827 (14.1)
Student	917 (18.1)	922 (17.7)	976 (17.5)	927 (16.1)	946 (16.3)
Homemaker	231 (4.6)	232 (4.5)	258 (4.6)	264 (4.6)	242 (4.1)
Unemployed	684 (13.5)	685 (13.1)	821 (14.7)	761 (13.3)	784 (13.4)
Disabled	273 (5.4)	5.7 (298)	272 (4.9)	365 (6.4)	359 (6.1)
Retired	75 (1.5)	78 (1.5)	85 (1.5)	108 (1.9)	116 (2.0)
Unknown' entered	862 (17.0)	914 (17.5)	930 (16.6)	850 (14.8)	630 (10.8)
No data recorded	38 (0.8)	21 (0.4)	39 (0.7)	27 (0.5)	-*
No. in work or study (% calculated from number who completed questionnaire)	**3020 (71.0)**	**3098 (71.3)**	**3242 (70.1)**	**3489 (70.8)**	**3902 (67.5)****

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

1.8 Pregnancy

71 women with cystic fibrosis had babies in 2016

48 men with cystic fibrosis became fathers in 2016

Diagnosis of cystic fibrosis

1.9 Age at diagnosis and screening in children under 16 in 2016 n=3762

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 in 2016; n (%)	Patients aged 10 years in 2016; n (%)	Patients aged 5 years in 2016; n (%)
Prenatal	<5	0 (0.0)	0 (0.0)
Birth-3 months	2988 (79.4)	145 (65.6)	264 (92.6)
4-6 months	163 (4.3)	16 (7.2)	7 (2.5)
7-12 months	135 (3.6)	15 (6.8)	6 (2.1)
1 yr	141 (3.7)	17 (7.7)	<5
2 yrs	112 (3.0)	11 (5.0)	<5
3 yrs	62 (1.6)	5 (2.3)	<5
4 yrs	44 (1.2)	<5	<5
5 yrs	27 (0.7)	<5	<5
6 yrs	21 (0.6)	<5	-
7 yrs	13 (0.3)	0 (0.0)	-
8 yrs	17 (0.5)	<5	-
9 yrs	12 (0.3)	<5	-
10 yrs	9 (0.2)	-	-
11 yrs	5 (0.1)	-	-
12 yrs	<5	-	-
13 yrs	<5	-	-
14 yrs	<5	-	-
15 yrs	0 (0.0)	-	-
	3762	**221**	**285**

The median (range) age at diagnosis for patients aged under 16 in 2016 is 25 days (0-180 months).

Diagnosis in the first three months of life is more common in children aged 5 years in 2016 (born after the UK-wide newborn screening programme was in place) than in children aged 10 years in 2016 (born before the UK-wide newborn screening programme was in place).

A total of **180** patients born in 2016 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 2015 is higher in this report than was recorded in the previous. It is likely that the 2016 figure will be updated in the next annual report in 2018.

Of the 247 newly diagnosed patients in 2016, **28 (11%)** presented with Meconium Ileus. Of everyone with complete data in 2016, **1,540 (16%)** presented with Meconium ileus.

1.10 Age at diagnosis and screening in adults aged 16 and over in 2016 n=5802 with a diagnosis date

The table below shows the age at diagnosis for people aged 16 and over in 2016. People aged 16 or over in 2016 were born when newborn screening was only done in a few areas of the UK, before it becoming universal in mid-2007.

Age at diagnosis	n (%)
Birth-3 months	2331 (40.2)
4-6 months	535 (9.2)
7-12 months	418 (7.2)
1 yr	425 (7.3)
2 yrs	292 (5.0)
3 yrs	223 (3.8)
4 yrs	171 (3.0)
5 yrs	88 (1.5)
6 yrs	81 (1.4)
7 yrs	56 (1.0)
8 yrs	68 (1.2)
9 yrs	47 (0.8)
10 yrs	42 (0.7)
11 yrs	42 (0.7)
12 yrs	44 (0.8)
13 yrs	48 (0.8)
14 yrs	36 (0.6)
15 yrs	51 (0.9)
16-20 yrs	176 (3.0)
21-25 yrs	115 (2.0)
26-30 yrs	112 (2.0)
31-35 yrs	120 (2.1)
36-40 yrs	95 (1.6)
41-45 yrs	68 (1.2)
46-50 yrs	36 (0.6)
51-60 yrs	49 (0.8)
61 yrs+	33 (0.6)
Overall	**5802**

Overall, 935 (16.1%) adults with CF in the Registry in 2016 were diagnosed at age 16 or over.

In 2016, 26 people aged 16 or over were newly diagnosed with cystic fibrosis.

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, dieticians, and psychologists.

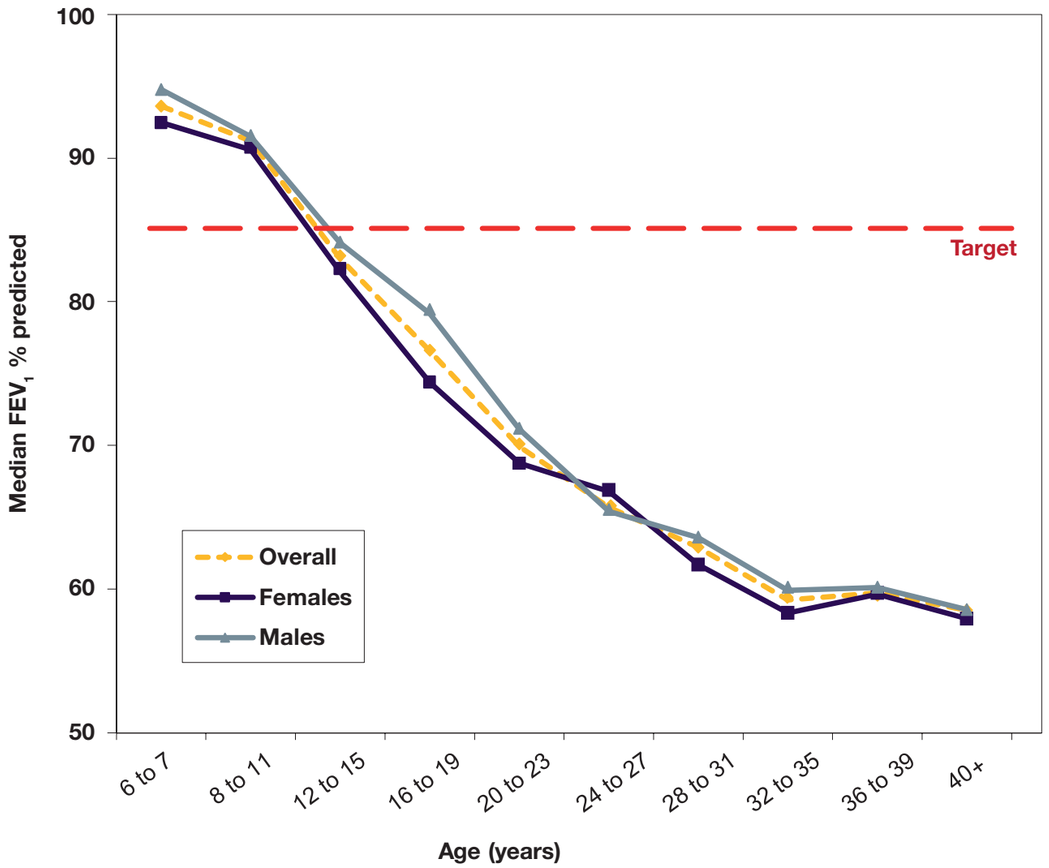
The FEV_1% predicted values shown in this report are calculated using an equation called Global Lungs Initiative, or 'GLI'*.

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

1.11 Median FEV_1% predicted (GLI equations) among patients aged 6 years and older

n=7830

The chart and table in this section show information about those patients whose FEV_1 data were complete. 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 CF.

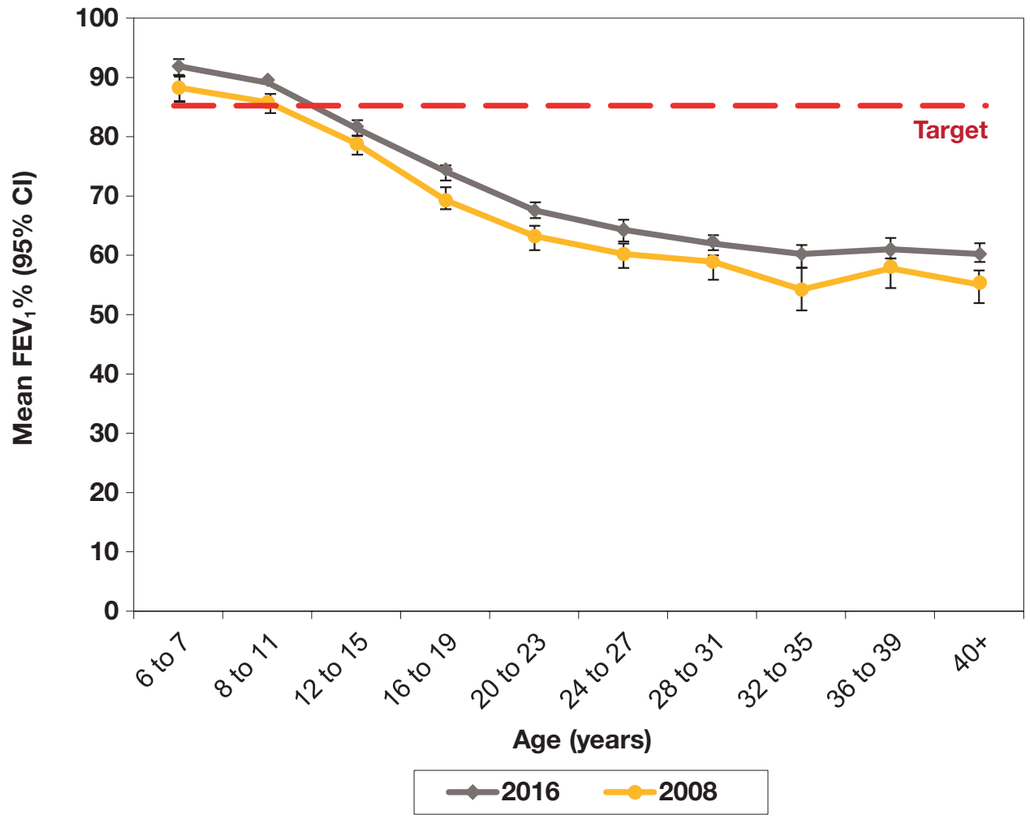


	Overall			Female			Male		
Age (yrs)	**N**	**Median**	**IQR**	**N**	**Median**	**IQR**	**N**	**Median**	**IQR**
6-7	500	93.2	82.7-101.4	263	91.8	82.8-100.2	237	94.4	82.6-103.3
8-11	929	90.2	79.5-99.5	446	90.0	79.5-99.6	483	90.7	79.6-99.3
12-15	872	82.5	71.2-93.3	454	81.3	69.7-93.1	418	83.5	73.7-93.5
16-19	923	76.8	60.2-90.2	451	74.6	57.9-89.2	472	78.6	62.7-91.0
20-23	949	69.9	50.6-84.3	459	68.6	49.8-84	490	71.3	52.1-85.2
24-27	903	65.8	47.2-81.5	416	66.5	48-80.3	487	64.7	45.5-82.2
28-31	747	62.6	42.1-79.3	325	61.4	42.4-80.5	422	63.6	41.4-77.9
32-35	599	58.8	39.6-77.8	271	58.0	39.3-74	328	59.8	39.9-79.6
36-39	419	59.2	42.3-76.9	174	58.9	41.4-73.7	245	59.7	42.9-77.7
40+	988	58.3	40.7-78.3	423	58.0	42.8-77.1	565	58.4	39.5-78.9
Overall	**7830**	**74.0**	**53.5-89.7**	**3663**	**73.4**	**53.3-89.5**	**4167**	**74.4**	**53.6-89.8**

1.12 Mean FEV_1% predicted over time (using GLI equations) in patients 6 years and older

n=7830 in 2016, n=4388 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 2016 compares to Registry data from 2008. 2008 is shown as the 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.

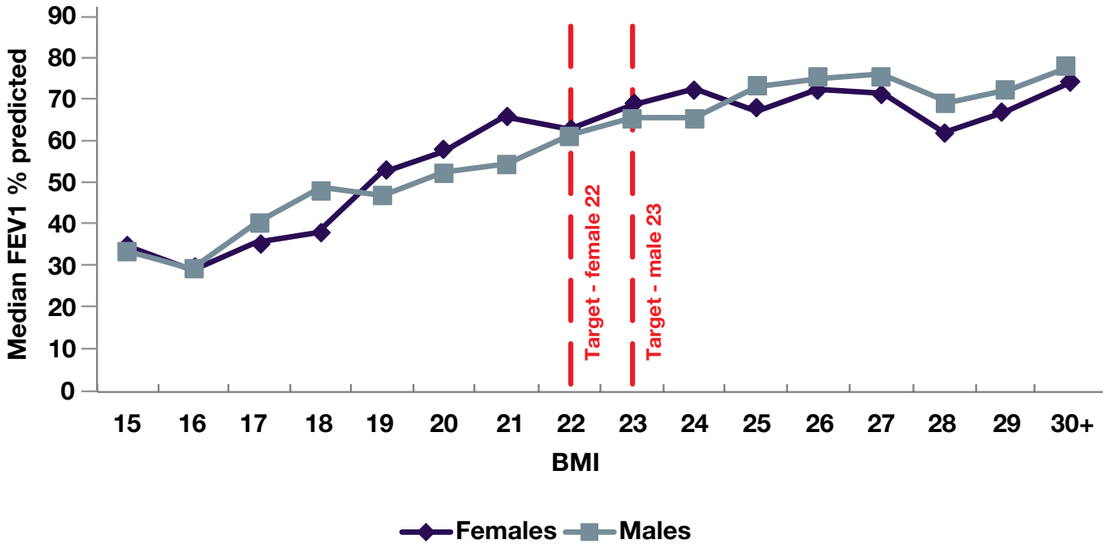


	Age (years)									
	6-7	**8-11**	**12-15**	**16-19**	**20-23**	**24-27**	**28-31**	**32-35**	**36-39**	**40+**
2008 mean FEV_1%	88.2	85.5	78.3	69.7	63	60.1	58.5	54.1	57.8	54.8
2016 mean FEV_1%	91.5	88.8	81.3	73.9	67.5	64.1	61.8	60	60.4	60.2
p-values (t-test)*	0.008	<0.001	<0.001	<0.001	<0.001	0.003	0.04	0.003	0.217	<0.001

* We have presented means with 95% confidence intervals in this section as these are more appropriate alongside the t-test (statistical hypothesis test) to compare FEV_1% between 2008 and 2016. If the p-value is less than 0.05 then the difference in FEV_1% is statistically significant.

1.13 Median FEV_1% predicted and BMI in people aged 20 years and over
n=4863

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.



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.

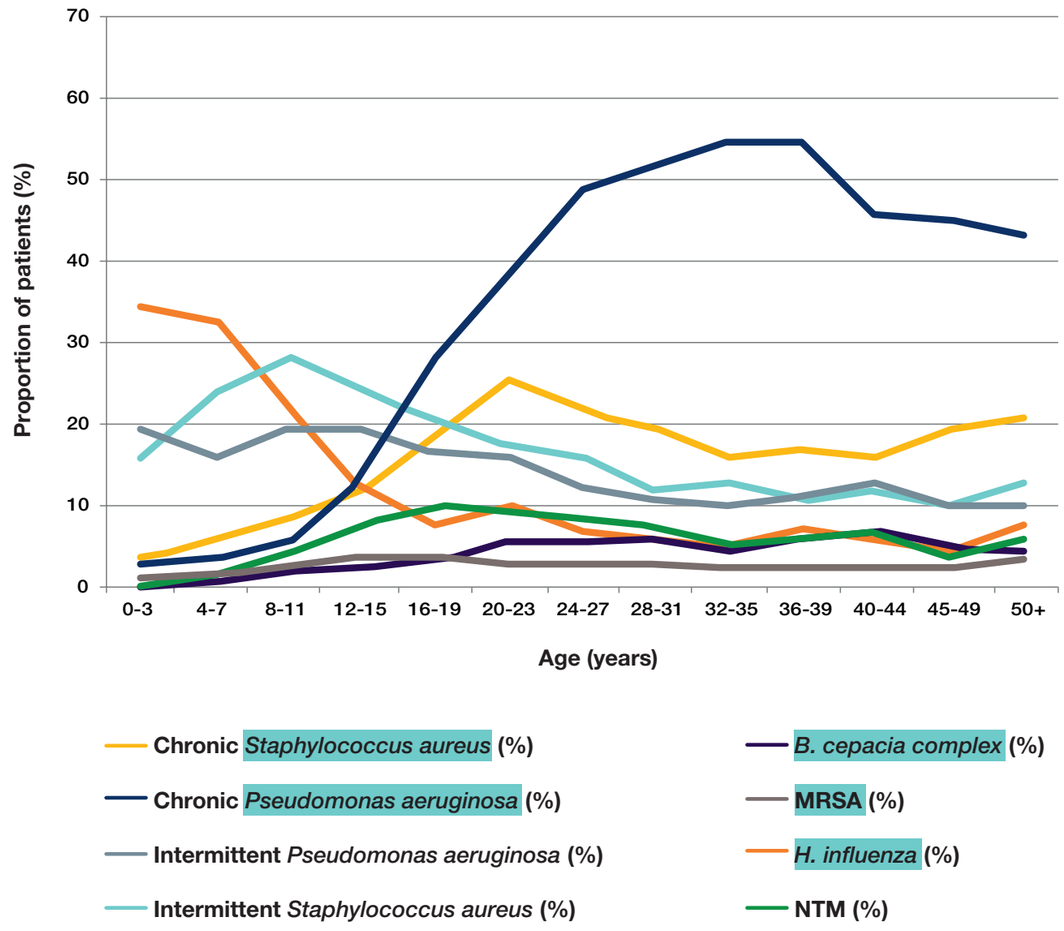
1.14 Median best FEV_1% predicted (GLI equations) among patients aged 6 years and older.

	Overall			Female			Male		
Age (yrs)	N	Median	IQR	N	Median	IQR	N	Median	IQR
6-7	460	99.0	89.3 -106.8	228	97.8	89.6 - 106.3	232	99.2	89.3 - 107.1
8-11	857	95.4	86.2 -103.6	415	95.1	84.8 - 103.2	442	96.0	87.2 – 104.0
12-15	821	87.2	76.9 - 97.4	430	87.1	75.7 - 96.7	391	88.3	78.0 - 97.9
16-19	810	83.1	66.5 - 94.4	402	81.0	63.9 - 92.8	408	83.8	69.3 - 95.5
20-23	882	74.4	54.8 - 89.1	436	71.9	50.4 - 86.4	446	76.8	58.8 – 91.0
24-27	837	69.5	51.0 - 86.8	385	69.8	51.5 - 86.1	452	69.3	50.9 - 87.1
28-31	686	67.5	47.5 - 84.2	300	65.8	47.0 - 84.8	386	68.2	47.9 - 83.3
32-35	547	60.9	43.5 - 80.4	253	60.8	42.7 - 76.9	294	61.6	44.2 - 82.5
36-39	401	63.4	47.8 - 81.3	171	62.0	46.8 - 80.3	230	64.9	48.4 - 83.4
40+	917	61.2	44.3 - 80.7	398	61.0	46.1 - 79.7	519	61.3	41.7 - 81.6
Overall	**7218**	**78.8**	**58 - 94.3**	**3418**	**78.2**	**57.1 - 93.9**	**3800**	**79.6**	**58.4 - 94.6**

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.

1.15 Lung infections in 2016
n=9695



1.15 Lung infections in 2016
<16 years n=3844, ≥16 years n=5851

	Paediatric Age Range (Years)				Overall
	0-3	4-7	8-11	12-15	Paediatric (<16 years)
Number in age range	844	1119	975	906	3844
Chronic *S.aureus* n (%)	27 (3.2%)	67 (6%)	84 (8.6%)	112 (12.4%)	290 (7.5%)
Intermittent *S.aureus* n (%)	135 (16%)	265 (23.7%)	274 (28.1%)	216 (23.8%)	890 (23.2%)
Chronic *P. aeruginosa* n (%)	26 (3.1%)	39 (3.5%)	57 (5.8%)	124 (13.7%)	246 (6.4%)
Intermittent *P. aeruginosa* n (%)	161 (19.1%)	180 (16.1%)	190 (19.5%)	176 (19.4%)	707 (18.4%)
B. cepacia complex n (%)	<5	7 (0.6%)	18 (1.8%)	21 (2.3%)	- (1.2%)
MRSA n (%)	9 (1.1%)	21 (1.9%)	27 (2.8%)	34 (3.8%)	91 (2.4%)
H. influenza n (%)	290 (34.4%)	363 (32.4%)	209 (21.4%)	108 (11.9%)	970 (25.2%)
NTM n (%)	<5	14 (1.3%)	40 (4.1%)	65 (7.2%)	- (3.1%)

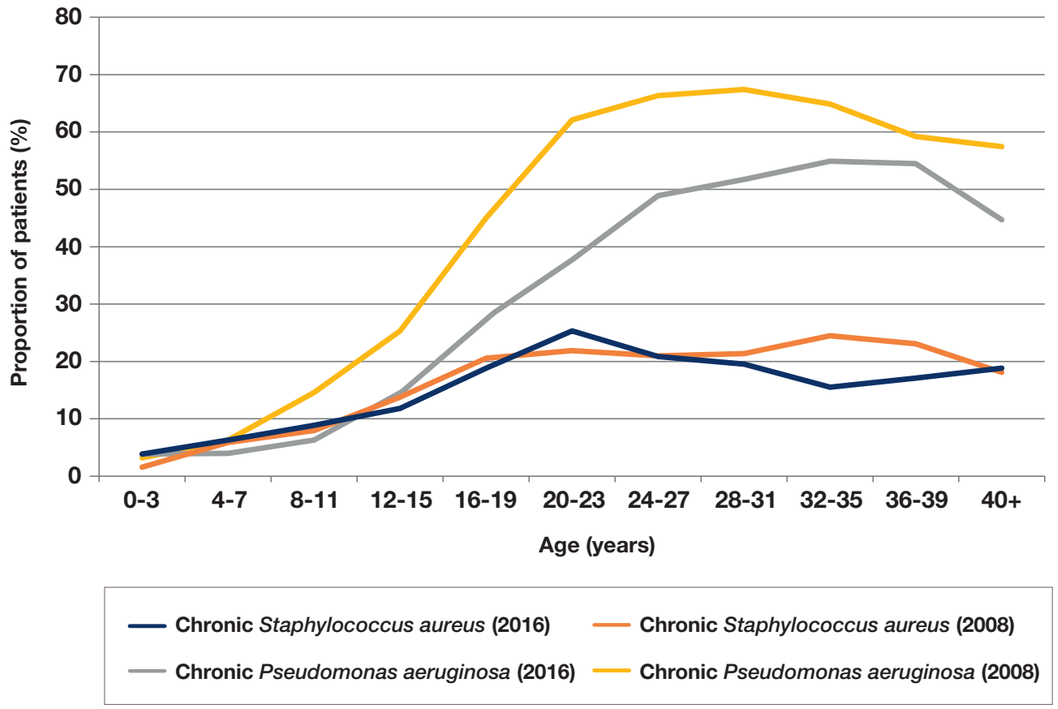
	Adult Age Range (Years)									Overall
	16-19	20-23	24-27	28-31	32-35	36-39	40-44	45-49	50+	Adults (≥16 years)
Number in age range	944	998	965	802	648	443	373	295	384	5851
Chronic *S.aureus* n (%)	179 (19%)	255 (25.6%)	206 (21.3%)	154 (19.2%)	104 (16%)	74 (16.7%)	59 (15.8%)	57 (19.3%)	79 (20.6%)	1167 (19.9%)
Intermittent *S.aureus* n (%)	191 (20.2%)	172 (17.2%)	154 (16%)	94 (11.7%)	83 (12.8%)	48 (10.8%)	44 (11.8%)	30 (10.2%)	50 (13%)	866 (14.8%)
Chronic *P. aeruginosa* n (%)	260 (27.5%)	379 (38%)	468 (48.5%)	414 (51.6%)	354 (54.6%)	240 (54.2%)	170 (45.6%)	133 (45.1%)	167 (43.5%)	2585 (44.2%)
Intermittent *P. aeruginosa* n (%)	156 (16.5%)	153 (15.3%)	117 (12.1%)	84 (10.5%)	63 (9.7%)	49 (11.1%)	48 (12.9%)	30 (10.2%)	39 (10.2%)	739 (12.6%)
B. cepacia complex n (%)	33 (3.5%)	57 (5.7%)	51 (5.3%)	48 (6%)	31 (4.8%)	27 (6.1%)	26 (7%)	15 (5.1%)	16 (4.2%)	304 (5.2%)
MRSA n (%)	36 (3.8%)	30 (3%)	33 (3.4%)	26 (3.2%)	14 (2.2%)	9 (2%)	8 (2.1%)	6 (2%)	13 (3.4%)	175 (3%)
H. influenza n (%)	77 (8.2%)	95 (9.5%)	67 (6.9%)	44 (5.5%)	34 (5.2%)	32 (7.2%)	22 (5.9%)	13 (4.4%)	27 (7%)	411 (7%)
NTM n (%)	96 (10.2%)	92 (9.2%)	80 (8.3%)	58 (7.2%)	32 (4.9%)	29 (6.5%)	24 (6.4%)	12 (4.1%)	23 (6%)	446 (7.6%)

1.16 Nontuberculous mycobacteria (NTM) or atypical mycobacteria**

	2014 (n=9532)	2015 (n=9587)	2016 (n=9695)
NTM Prevalence (%)	583 (6.1%)	536 (5.6%)	567 (5.8%)
On NTM treatment in the given year (% of NTM prevalence in given year)	294 (50.4%)	300 (56.0%)	333 (58.7%)
NTM Incidence	336	222	228
M.abscessus prevalence*	300	321	337

* M.abscessus incidence in 2016 was 110. Prior years cannot be evaluated as enhanced NTM reporting was not available prior to 2014.

1.17 Lung infections over time
n=6082 in 2008, n=9695 in 2016



Chronic *Staphylococcus aureus*	Age (years)										
	0-3	4-7	8-11	12-15	16-19	20-23	24-27	28-31	32-35	36-39	40-44
2016 (%)	3.2	6	8.6	12.4	19	25.6	21.3	19.2	16	16.7	18.5
2008 (%)	1.4	5.7	7.8	13.6	20.4	21.7	20.8	21.2	24.3	22.9	17.9
p-value*	0.031	0.805	0.581	0.496	0.516	0.103	0.834	0.457	0.008	0.076	0.809

Chronic *Pseudomonas aeruginosa*	Age (years)										
	0-3	4-7	8-11	12-15	16-19	20-23	24-27	28-31	32-35	36-39	40+
2016 (%)	3.1	3.5	5.8	13.7	27.5	38	48.5	51.6	54.6	54.2	44.7
2008 (%)	2.6	6.8	14.3	25.3	45.1	61.8	66	67.3	64.8	58.8	57.3
p-value*	0.581	0.002	<0.001	<0.001	<0.001	<0.001	<0.001	<0.001	0.067	0.444	0.002

* The proportion of people with each infection within each age group was compared between 2008 and 2016. If the p-value is less than 0.05 then the difference in the proportions is statistically significant.

****Reference to figure 1.16 (page 24):** Non-tuberculous mycobacterium is slow to grow and slow to treat. It may be present for several years before eradication, or may never be cleared. In the table on page 24, '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 2 years of data.

1.18 Prevalence of complications

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

	Overall (n=9587)	<16 years (n=3845)	≥16 years (n=5742)
	N (%)	N (%)	N (%)
Respiratory Related			
Nasal polyps requiring surgery; n (%)	379 (3.9)	96 (2.5)	283 (4.8)
Sinus disease; n (%)	949 (9.8)	62 (1.6)	887 (15.2)
Asthma; n (%)	1306 (13.5)	437 (11.4)	869 (14.9)
ABPA; n (%)	646 (6.7)	145 (3.8)	501 (8.6)
Haemoptysis; n (%)	697 (7.2)	35 (0.9)	662 (11.3)
Pneumothorax requiring chest tube; n (%)	33 (0.3)	5 (0.1)	28 (0.5)
Pancreas & Hepatobiliary Disease			
Raised liver enzymes; n (%)	962 (9.9)	234 (6.1)	728 (12.4)
Liver disease; n (%)	1332 (13.7)	289 (7.5)	1043 (17.8)
Cirrhosis with no portal hypertension; n (%)	119 (1.2)	27 (0.7)	92 (1.6)
Cirrhosis with portal hypertension; n (%)	146 (1.5)	24 (0.6)	122 (2.1)
Gall bladder disease requiring surgery; n (%)	99 (1.0)	9 (0.2)	90 (1.5)
Pancreatitis; n (%)	43 (0.4)	5 (0.1)	38 (0.6)
Upper Gastrointestinal			
GERD; n (%)	1118 (11.5)	178 (4.6)	940 (16.1)
Peptic ulcer; n (%)	<5	<5	<5
GI bleed (varices as source); n (%)	-	<5	5 (0.1)
GI bleed (non varices as source); n (%)	-	<5	9 (0.2)
Lower Gastrointestinal			
Intestinal obstruction; n (%)	31 (0.3)	16 (0.4)	15 (.3)
DIOS; n(%)	444 (4.6)	76 (2)	368 (6.3)
Fibrosing colonopathy / colonic stricture; n (%)	<5	<5	<5
Rectal prolapse; n (%)	12 (0.1)	7 (0.2)	5 (0.1)
Renal			
Kidney stones; n(%)	77 (0.8)	7 (0.2)	70 (1.2)
Renal failure; n(%)	-	<5	75 (1.3)
Muscolo-Skeletal			
Arthritis; n (%)	179 (1.8)	5 (0.1)	174 (3.0)
Arthropathy; n (%)	526 (5.4)	21 (0.5)	505 (8.6)
Bone fracture; n (%)	26 (0.3)	8 (0.2)	18 (0.3)
Osteopenia; n (%)	1339 (13.8)	41 (1.1)	1298 (22.2)
Osteoporosis; n (%)	481 (5.0)	7 (0.2)	474 (8.1)
Other			
Cancer confirmed by histology; n (%)	24 (0.2)	0 (0)	24 (0.4)
Port inserted or replaced; n (%)	293 (3.0)	98 (2.5)	195 (3.3)
Depression; n (%)	500 (5.2)	13 (0.3)	487 (8.3)
Hearing loss; n (%)	275 (2.8)	33 (0.9)	242 (4.1)
Hypertension; n (%)	100 (1.0)	0 (0)	100 (1.7)

1.19 Incidence of complications
n=9695

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

	2015			2016		
	Overall (n=9587)	<16years (n=3845)	>=16 years (n=5742)	Overall (n=9695)	<16years (n=3844)	>=16 years (n=5851)
ABPA: n (%)	99 (1.0)	38 (0.4)	61 (0.6)	188 (1.9)	68 (1.8)	120 (2.1)
Cirrhosis - no portal hypertension; n (%)	33 (0.3)	14 (0.1)	19 (0.2)	49 (0.5)	13 (0.1)	36 (0.6)
Cirrhosis - with hypertension; n (%)	20 (0.2)	8 (0.1)	12 (0.1)	38 (0.4)	7 (0.2)	31 (0.5)
Cancer confirmed by histology : n (%)	9 (0.1)	0 (0)	9 (0.1)	16 (0.2)	0 (0)	16 (0.3)

1.20 CF-related diabetes
n=7212

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, causing CFRD. CFRD is different from type 1 and type 2 diabetes, but has features of both.

	All ≥ 10 years (n=7212)		10-15 years (n=1361)		≥16 years (n=5851)	
On CFRD treatment; n (%)	2124	29.5%	157	11.5%	1967	33.6%
Screening:						
Yes	3783	52.5%	952	70.0%	2831	48.4%
No	1363	18.9%	274	20.1%	1089	18.6%
Known CFRD	1832	25.4%	49	3.6%	1783	30.5%
Unknown	118	1.6%	39	2.9%	79	1.4%

Antibiotics

1.21 Intravenous (IV) antibiotics
n=9695

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	844	37 (4.4)	7 (6-10)	228 (27.0)	14 (8-19)	232 (27.5)	14 (11-20)
4-7	1119	107 (9.6)	14 (11-24)	306 (27.3)	14 (8-20)	330 (29.5)	14 (13-28)
8-11	975	150 (15.4)	14 (10-30)	337 (34.6)	14 (9-28)	368 (37.7)	12 (14-41)
12-15	906	192 (21.2)	21 (12-40)	382 (42.2)	15 (10-35)	431 (47.6)	28 (14-43)
16-19	943	232 (24.6)	14 (10-35)	418 (44.3)	15 (9-36)	477 (50.5)	28 (14-45)
20-23	998	325 (32.6)	19 (13-33)	458 (45.9)	17 (9-40)	559 (56.0)	28 (14-55)
24-27	965	361 (37.4)	18 (13-36)	456 (47.3)	19 (10-37)	565 (58.5)	29 (14-56)
28-31	802	316 (39.4)	21 (14-33)	340 (42.4)	18 (9-36)	465 (58.0)	28 (14-50)
32-35	648	220 (34.0)	24 (14-41)	257 (39.7)	21 (10-45)	353 (54.5)	31 (14-57)
36-39	443	156 (35.2)	25 (13-42)	143 (32.3)	14 (8-28)	218 (49.2)	28 (14-46)
40-44	373	98 (26.3)	20 (12-32)	118 (31.6)	15 (8-34)	158 (42.4)	27 (14-46)
45-49	295	89 (30.2)	26 (14-40)	82 (27.8)	15 (9-29)	121 (41.0)	29 (14-44)
50+	384	85 (22.1)	21 (13-36)	131 (34.1)	14 (9-32)	167 (43.5)	21 (13-50)
Overall	**9695**	**2368 (24.4)**	**19 (13-35)**	**3656 (37.7)**	**15 (9-33)**	**4444 (45.8)**	**27 (14-45)**

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

	2008			2016		
	Overall	< 16 years	≥16 years	Overall	< 16 years	≥16 years
Patients with chronic *P. aeruginosa*	2098	299	1799	2831	246	2585
Tobramycin solution; n (%)	412 (19.6)	48 (16.1)	364 (20.2)	591 (20.9)	75 (26.9)	516 (20.0)
Other aminoglycoside; n (%)	43 (2.0)	5 (0.2)	38 (2.1)	73 (2.6)	11 (3.9)	62 (2.4)
Colistin; n (%)	914 (43.6)	174 (58.2)	740 (41.1)	716 (25.3)	109 (39.1)	607 (23.5)
Promixin; n (%)	490 (23.4)	73 (24.4)	417 (23.2)	844 (29.8)	98 (35.1)	746 (28.9)
Aztreonam; n (%)	-	-	-	531 (18.8)	12 (4.3)	519 (20.1)
Colistimethate (DPI); n (%)	-	-	-	472 (16.7)	29 (10.4)	443 (17.1)
Tobramycin Inhalation Powder; n (%)	-	-	-	787 (27.8)	34 (12.2)	753 (29.1)
At least one of the above*; n (%)	1597 (76.1)	257 (86.0)	1340 (74.5)	2480 (87.6)	230 (93.5)	2251 (87.1)

*In 2016, this includes Aztreonam, Colistimethate and Tobramycin Inhalation Powder.

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.

1.23 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*.

	2008				2016			
	Overall (n=1958)	0-3 years (n=15)	4-15 years (n=363)	≥16 years (n=1580)	Overall (n=3833)	0-3 years (n=32)	4-15 years (n=642)	≥16 years (n=3159)
Patients with chronic *P. aeruginosa*; n (%)	1246 (63.6)	<5	105 (28.9)	1139 (72.1)	1859 (48.5)	5 (15.6)	94 (14.6)	1760 (55.7)
Patients without chronic *P. aeruginosa;* n (%)	712 (36.4)	13 (86.7)	258 (71.1)	441 (27.9)	1974 (51.5)	27 (84.4)	548 (85.4)	1399 (44.3)

Muco-active therapies

1.24 Mannitol

	2016	
Age	**Total Patients**	**Patients on Mannitol**
0-3	844	0
4-7	1119	0
8-11	975	0
12-15	906	<5
16-19	943	23 (2.4)
20-23	998	49 (4.9)
24-27	965	68 (7.0)
28-31	802	49 (4.9)
32-35	648	38 (5.9)
36-39	443	25 (5.6)
40+	1052	43 (4.1)
Overall	**9695**	**298 (3.1)**

1.25 DNase

	DNase; n (%)			
	2008		2016	
Age	**Total patients**	**Patients on DNase**	**Total patients**	**Patients on DNase**
0-3	605	46 (7.6)	844	126 (14.9)
4-7	621	125 (20.1)	1119	539 (48.2)
8-11	663	227 (34.2)	975	714 (73.2)
12-15	773	359 (46.4)	906	719 (79.4)
16-19	762	377 (49.5)	943	720 (76.4)
20-23	725	319 (44.0)	998	681 (68.2)
24-27	605	288 (47.6)	965	617 (63.9)
28-31	419	182 (43.4)	802	498 (62.1)
32-35	260	108 (41.5)	648	391 (60.3)
36-39	237	83 (35.0)	443	255 (57.6)
40+	412	147 (35.7)	1052	531 (50.5)
Overall	**6082**	**2261(37.2)**	**9695**	**5791 (59.7)**

1.26 Hypertonic saline

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

	Hypertonic saline; n (%)			
	2008		2016	
Age	**Total patients**	**Patients on hypertonic saline**	**Total patients**	**Patients on hypertonic saline**
0-3	605	<5	844	69 (8.2)
4-7	621	15 (2.4)	1119	241 (21.5)
8-11	663	23 (3.5)	975	311 (31.9)
12-15	773	32 (4.1)	906	425 (46.9)
16-19	762	33 (4.3)	943	350 (37.1)
20-23	725	50 (6.9)	998	294 (29.5)
24-27	605	60 (9.9)	965	260 (26.9)
28-31	419	37 (8.8)	802	256 (31.9)
32-35	260	29 (11.2)	648	219 (33.8)
36-39	237	16 (6.8)	443	140 (31.6)
40+	412	33 (8.0)	1052	285 (27.1)
Overall	**6082**	**331 (5.4)**	**9695**	**2850 (29.4)**

Other Therapies

1.27 CFTR modifiers

1.27a Ivacaftor

Ivacaftor began being prescribed as a treatment for CF in patients aged 6 years and over with at least one copy of the genotype G551D in June 2012. The table shows information about ivacaftor use and outcomes from June 2012 – December 2016.

Number of patients on ivacaftor in the UK	**473**
Sweat chloride before Ivacaftor	104 (93, 113)
Sweat chloride 6-8 weeks after Ivacaftor	46 (31, 60)
FEV_1% before Ivacaftor	61.9 (47.3, 74.1)
FEV_1% 6-8 weeks after Ivacaftor	69.8 (55.2, 84.1)
Number of patients stopped Ivacaftor ever	27

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.

1.27b Ivacaftor/Lumacaftor

Ivacaftor/Lumacaftor is licensed for use in patients aged 12 and over with two copies of the F508del mutation. In 2016 it was available to specific people with CF in the UK through a named patient access scheme. 78 people received this drug in 2016.

1.28 Oxygen and non-invasive ventilation

	Overall (n=9695)	**<16 years (n=3844)**	**≥16 years (n=5851)**
Non Invasive Ventillation (NIV); n (%)	236 (2.4)	26 (0.7)	210 (3.6)
Long-term oxygen; n (%) **Among those who have long-term oxygen:**	640 (6.6)	95 (2.5)	545 (9.3)
Continuously	146 (1.5)	10 (0.3)	136 (2.3)
Nocturnal or with exertion	196 (2.0)	25 (0.7)	171 (2.9)
As required (PRN)	77 (0.8)	8 (0.2)	69 (1.2)
With exacerbation	221 (2.3)	52 (1.4)	169 (2.9)

1.29 Physiotherapy

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

	Overall (n=9695)	<16 years (n=3844)	≥16 years (n=5851)
Active cycle of breathing techniques; n (%)	1388 (14.3)	442 (11.5)	946 (16.2)
Autogenic drainage (including assisted autogenic drainage); n (%)	1695 (14.5)	235 (6.1)	1460 (25.0)
Any form of PEP; n (%)	5577 (57.5)	2838 (73.8)	2739 (46.8)
VEST; n (%)	187 (1.9)	100 (2.6)	87 (1.5)
Exercise; n (%)	5544 (57.2)	2323 (60.4)	3221 (55.1)

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

1.30 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=9695)	<16 years (n=3844)	≥16 years (n=5851)
Any supplemental feeding; n (%)	3091 (31.9)	1114 (29.0)	1977 (33.8)
Nasogastric tube	94 (1.0)	10 (0.3)	84 (1.4)
Gastrostomy tube/Button	543 (5.6)	223 (5.8)	320 (5.5)
Jejunal	14 (0.1)	<5	12 (0.2)
Total Parenteral Nutrition (TPN)	<5	<5	<5

1.31 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.

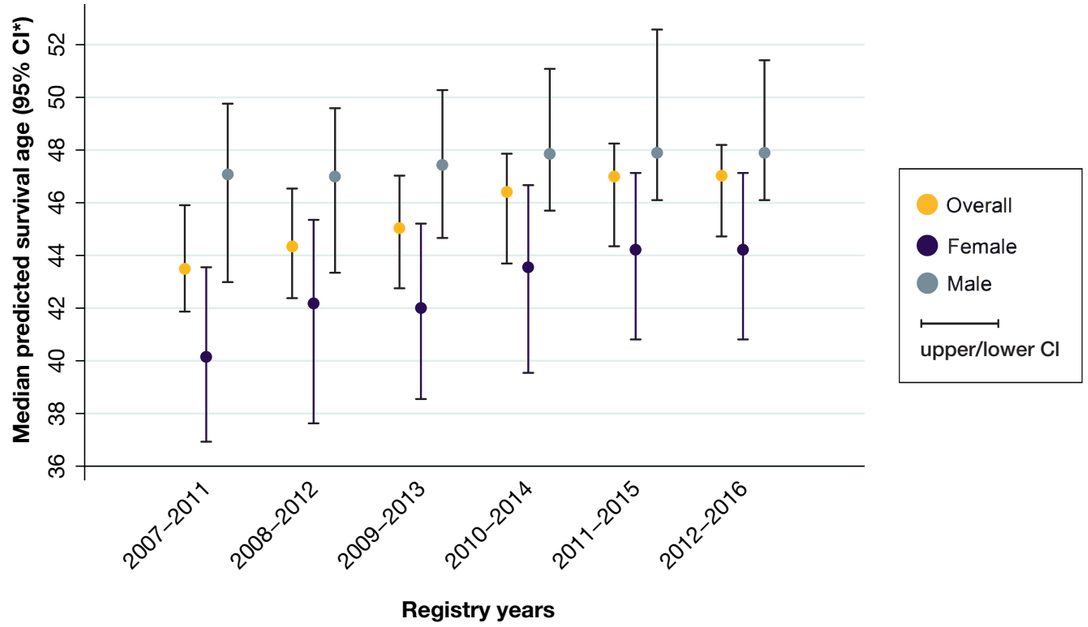
	2012	2013	2014	2015	2016
Number evaluation	225	220	247	229	221
Number accepted	120	136	146	125	96
Number receiving aged <16 yrs	<5	<5	5	<5	<5
Bilateral lung	<5	<5	<5	<5	<5
Heart and lung	0	0	0	0	0
Liver	<5	<5	<5	<5	0
Other	0	0	0	<5	0
Number receiving aged 16+ yrs	52	54	67	46	51
Bilateral lung	43	48	59	42	46
Heart and lung	<5	0	0	0	0
Liver	6	<5	5	<5	<5
Other	<5	<5	5	<5	<5

Survival

1.32 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 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.



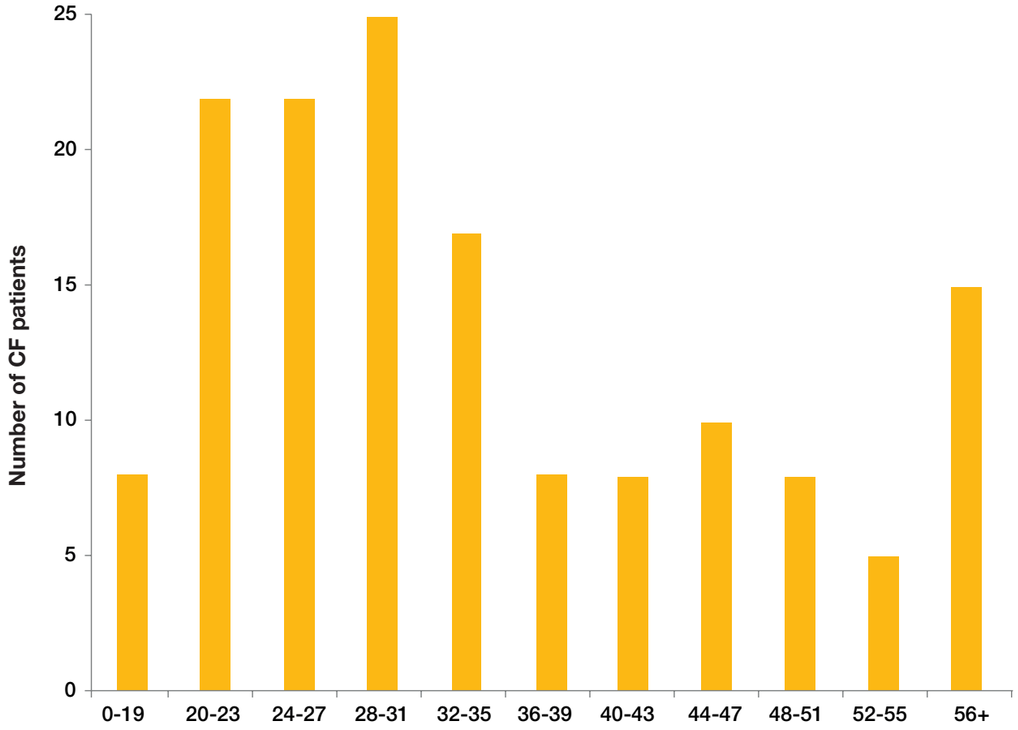
Median predicted survival age (95% CI*)				
Registry years	**Overall**	**Female**	**Male**	**p-values comparing survival**
2007-2011	43.5 (41.9-45.9)	40.1 (36.9-43.6)	47.1 (43.0-49.8)	<0.001
2008-2012	44.3 (42.4-46.5)	42.2 (37.6-45.3)	47.0 (43.3-49.6)	<0.001
2009-2013	45.0 (42.8-47.0)	42.0 (38.5-45.2)	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

8 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

1.33 Age distribution of deaths in 2016

The table below shows the ages of the 148 people with CF who died in 2016. 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 cystic fibrosis patients
0-19	8
20-23	22
24-27	22
28-31	25
32-35	17
36-39	8
40-43	8
44-47	10
48-51	8
52-55	5
56+	15
Total	**148**

Median age of death is based on the people with CF who died in any given year. In 2016 the median age of the 148 people who died was 31.

Genotypes

Genotypes are part of the genetic makeup of an individual that usually controls 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.

9544 (98.4%) patients have been genotyped with a recorded value.

DF508 Mutations: n (%)

Homozygous DF508: 4789 (50.2%)

Heterozygous DF508: 3882 (40.7%)

1.34 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.

Mutations				
Nucleotide	**Protein**	**Legacy name**	**N**	**%**
c.1521_1523delCTT	p.Phe508del	F508del	8671	90.9
c.1652G->A	p.Gly551Asp	G551D	561	5.9
c.350G->A	p.Arg117His	R117H	489	5.1
c.1624G->T	p.Gly542X	G542X	341	3.6
c.489+1G->T	-	621+1G->T	244	2.6
c.3909C->G	p.Asn1303Lys	N1303K	158	1.7
c.1585-1G->A	-	1717-1G->A	135	1.4
c.1766+1G->A	-	1898+1G->A	123	1.3
c.3528delC	p.Lys1177SerfsX15	3659delC	104	1.1
c.200C->T	p.Pro67Leu	P67L	101	1.1
c.3454G->C	p.Asp1152His	D1152H	100	1.0
c.1679G->C	p.Arg560Thr	R560T	96	1.0
c.1519_1521delATC	p.Ile507del	I507del	86	0.9
c.1477C->T	p.Gln493X	Q493X	85	0.9
c.3140-26A->G	-	3272-26A->G	79	0.8
c.1657C->T	p.Arg553X	R553X	78	0.8
c.254G->A	p.Gly85Glu	G85E	78	0.8
c.3717+12191C->T	-	3849+10kbC->T	74	0.8
c.178G->T	p.Glu60X	E60X	67	0.7
c.1022_1023insTC	p.Phe342HisfsX28	1154insTC	56	0.6
c.3846G->A	p.Trp1282X	W1282X	54	0.6
c.2657+5G->A	-	2789+5G->A	52	0.5
c.948delT	p.Phe316LeufsX12	1078delT	52	0.5
c.1646G->A	p.Ser549Asn	S549N	41	0.4
c.2052delA	p.Lys684AsnfsX38	2184delA	38	0.4
c.1040G->C	p.Arg347Pro	R347P	37	0.4
c.1364C->A	p.Ala455Glu	A455E	37	0.4
c.617T->G	p.Leu206Trp	L206W	33	0.3
c.3484C->T	p.Arg1162X	R1162X	32	0.3
c.1558G->T	p.Val520Phe	V520F	28	0.3
c.579+3A->G	-	711+3A->G	27	0.3
c.1210-12[5](AJ574948.1:g.152T[5])	-	5T	24	0.3
c.2657+2_2657+3insA	-	2789+2insA	23	0.2
c.2988+1G->A	-	3120+1G->A	22	0.2
c.1055G->A	p.Arg352Gln	R352Q	21	0.2

Nucleotide	Protein	Legacy name	N	%
c.1040G->A	p.Arg347His	R347H	18	0.2
c.1753G->T	p.Glu585X	E585X	18	0.2
c.2583delT	p.Phe861LeufsX3	2711delT	18	0.2
c.1000C->T	p.Arg334Trp	R334W	16	0.2
c.1393-1G->A	-	1525-1G->A	15	0.2
c.3472C->T	p.Arg1158X	R1158X	14	0.1
c.2834C->T	p.Ser945Leu	S945L	14	0.1
c.532G->A	p.Gly178Arg	G178R	14	0.1
c.1705T->G	p.Tyr569Asp	Y569D	14	0.1
c.2125C->T	p.Arg709X	R709X	13	0.1
c.2052_2053insA	p.Gln685ThrfsX4	2184insA	11	0.1
c.3196C->T	p.Arg1066Cys	R1066C	11	0.1
c.579+1G->T	-	711+1G->T	11	0.1
c.3197G->A	p.Arg1066His	R1066H	11	0.1
c.1466C->A	p.Ser489X	S489X	11	0.1
c.3705T->G	p.Ser1235Arg	S1235R	10	0.1
c.1679+1G->C	-	1811+1G->C	9	0.1
c.349C->T	p.Arg117Cys	R117C	7	0.1
c.292C->T	p.Gln98X	Q98X	7	0.1
c.1675G->A	p.Ala559Thr	A559T	6	0.1
c.223C->T	p.Arg75X	R75X	6	0.1
c.224G->A	p.Arg75Gln	R75Q	6	0.1
c.1645A->C	p.Ser549Arg	S549R	5	0.1
c.2128A->T	p.Lys710X	K710X	5	0.1
c.2012delT	p.Leu671X	2143delT	<5	-
c.443T->C	p.Ile148Thr	I148T	<5	-
c.274G->A	p.Glu92Lys	E92K	<5	-
c.2290C->T	p.Arg764X	R764X	<5	-
c.2260G->A	p.Val754Met	V754M	<5	-
c.2051_2052delAAinsG	p.Lys684SerfsX38	2183AA->G or 2183delAA->G	<5	-
c.2195T->G	p.Leu732X	L732X	<5	-
c.1329_1330insAGAT	p.Ile444ArgfsX3	1461ins4	<5	-
c.3181G->C	p.Gly1061Arg	G1061R	<5	-
c.3773_3774insT	p.Leu1258PhefsX7	3905insT	<5	-
c.1545_1546delTA	p.Tyr515X	1677delTA	<5	-
c.1727G->C	p.Gly576Ala	G576A	<5	-
c.1654C->T	p.Gln552X	Q552X	<5	-
c.3209G->A	p.Arg1070Gln	R1070Q	<5	-
c.2780T->C	p.Leu927Pro	L927P	<5	-
c.3310G->T	p.Glu1104X	E1104X	<5	-
c.3718-1G->A	-	3850-1G->A	<5	-
c.166G->A	p.Glu56Lys	E56K	<5	-
c.2739T->A	p.Tyr913X	Y913X	<5	-
'Other' selected			1345	14.1

1.35 Genotypes by devolved nation

Country	England		Scotland		Wales		N. Ireland	
	n=7890	(%)	n=823	(%)	n=433	(%)	N=398	(%)
^F508	7149	90.6	745	90.5	392	90.5	335	84.2
G542X	233	3.0	59	7.2	24	5.5	24	6.0
G551D	422	5.3	87	10.6	21	4.8	31	7.8
R117H	350	4.4	61	7.4	14	3.2	58	14.6
1898+1G->A	89	1.1	<5	-	29	6.7	<5	-
621`+1G->T	177	2.2	12	1.5	42	9.7	13	3.3

Section 2 and 3: Centre-level analysis

Cystic fibrosis care in the UK is led by 54 regional centres, five 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	24	54
Stand-alone clinics	2	3	5
Networked clinics	68	7	75

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

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

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

Full tables of the data are shown in appendix 2 on page 52.

Key

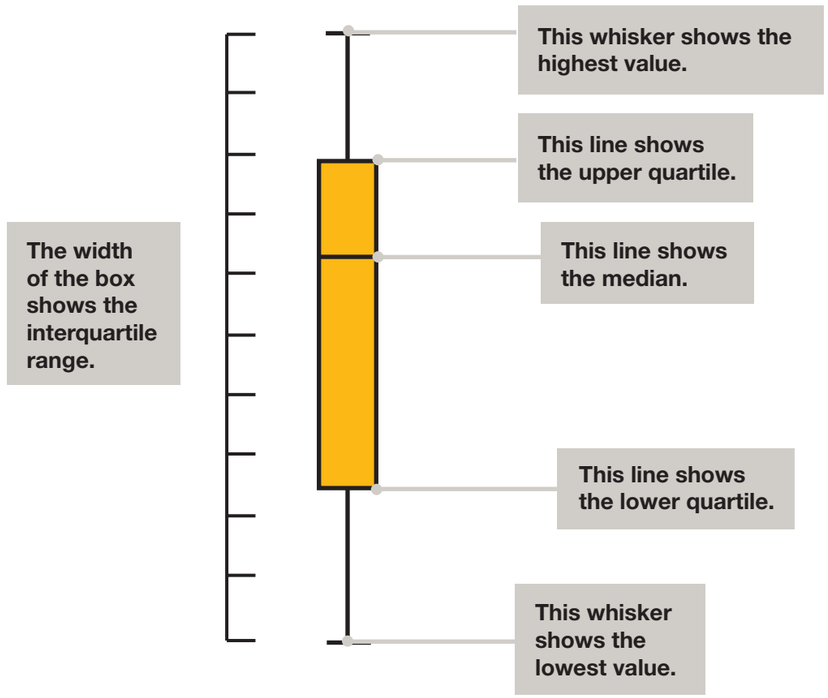
Paediatric centre

Adult centre

A guide to the charts

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

Box plots



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

Funnel plots

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

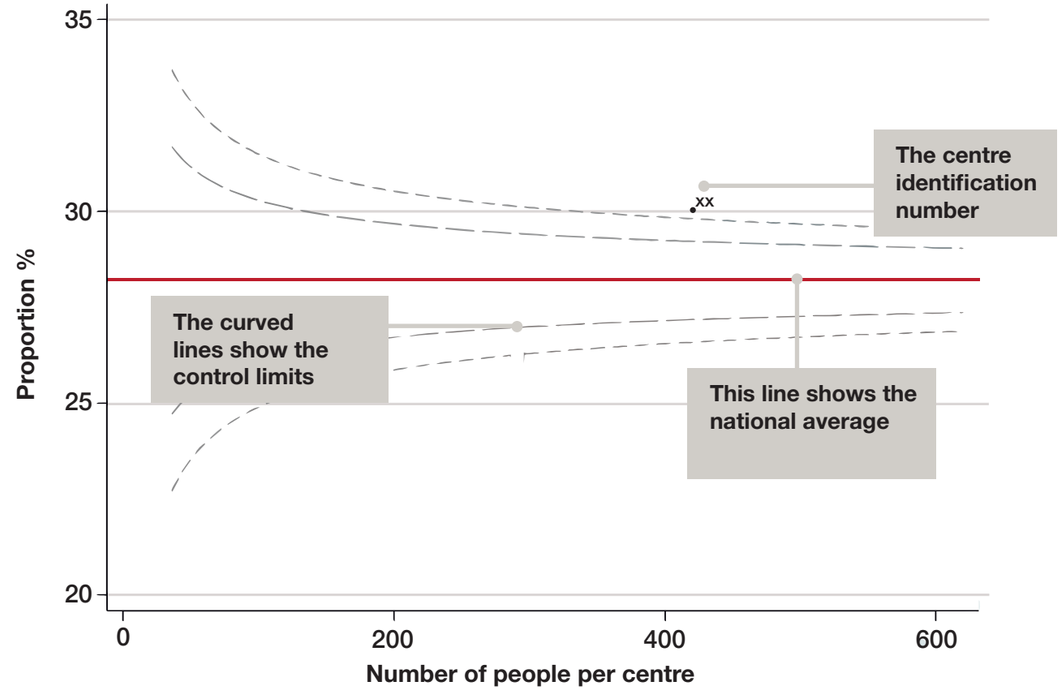
There will always be some natural variation between centres, because of differences between the population receiving care. Using only the national average as a standard can make it difficult to tell whether a survival rate that sits above the national average is higher than we would expect it to be,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

We have followed the standard approach for funnel plots, using means as the summary measure and standard deviations to calculate the control limits. This is because, for the CF-related outcomes we report, the data are appropriately distributed.



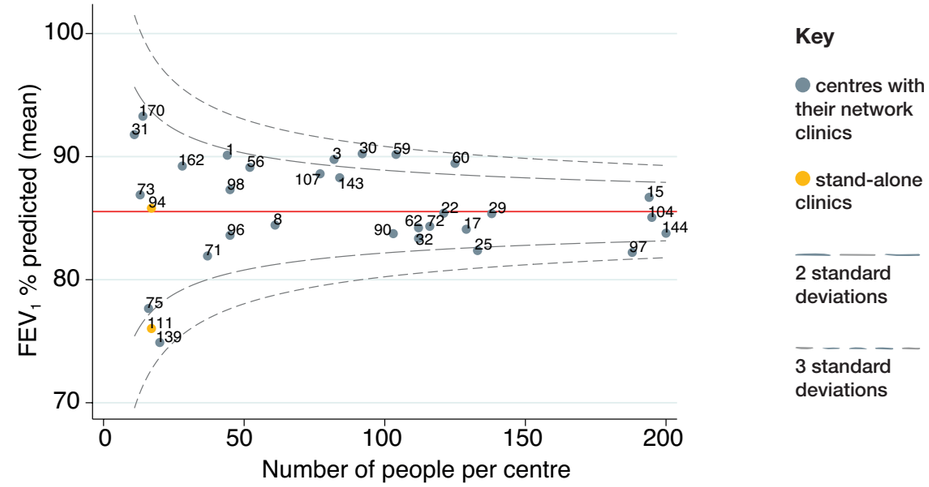
Section 2 Paediatric centre analysis

n=4149

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.

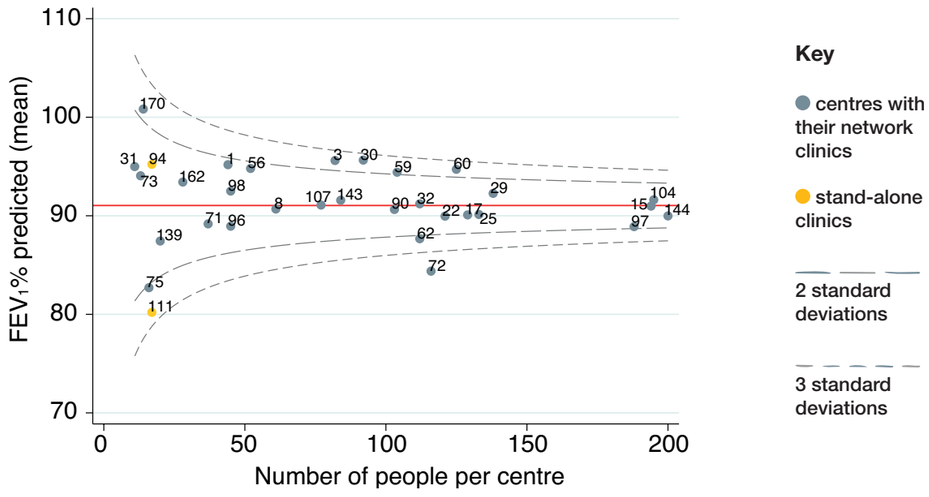
2.1a, 2.1b and 2.2 have been adjusted for age (using a linear term for age). This means that the data have been fine-tuned to take account of the different spread of ages across centres and clinics. The adjusted values are intended to show what the average lung function or BMI percentile would be for 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.

2.1a Age adjusted FEV_1% predicted at annual review, among patients aged 6 and over, by paediatric centre/clinic (without a history of lung transplant)



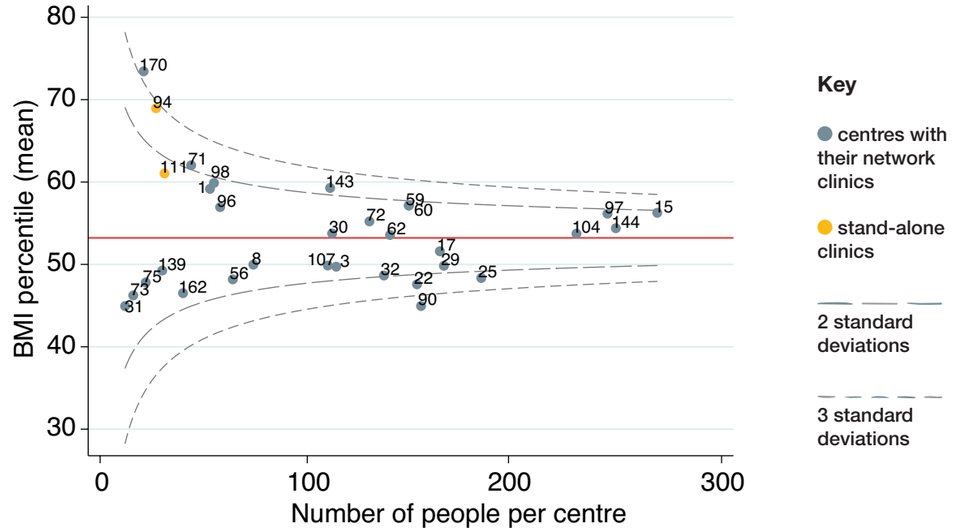
The mean FEV_1 % predicted for patients attending paediatric centres/clinics is 85% predicted.

2.1b Age adjusted best FEV_1% predicted (GLI) among patients aged 6 and over, by paediatric centre /clinic (without a history of lung transplant)



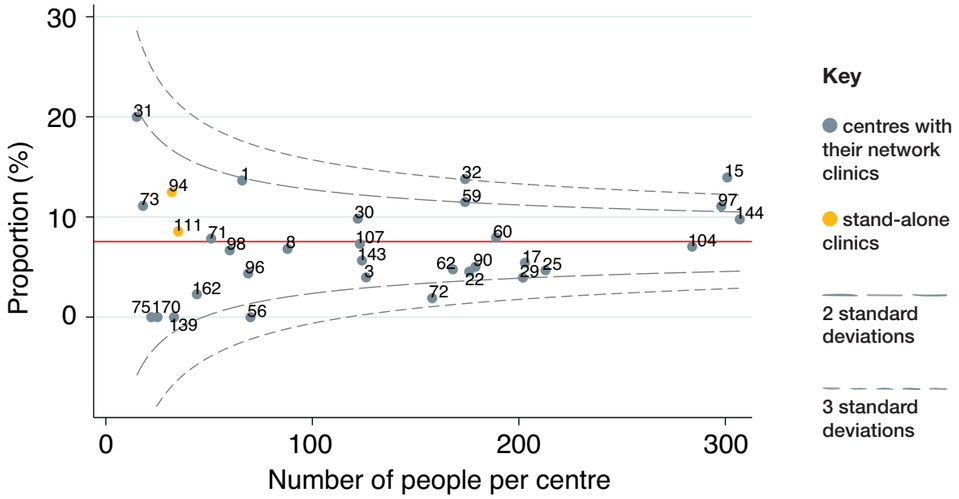
The mean best FEV_1 % predicted for patients attending paediatric centres/clinics is 91% predicted. Where best FEV_1 % predicted was missing, the FEV_1% predicted at annual review was used.

2.2 Age adjusted BMI percentile among patients aged 2-15 years by paediatric centre/clinic



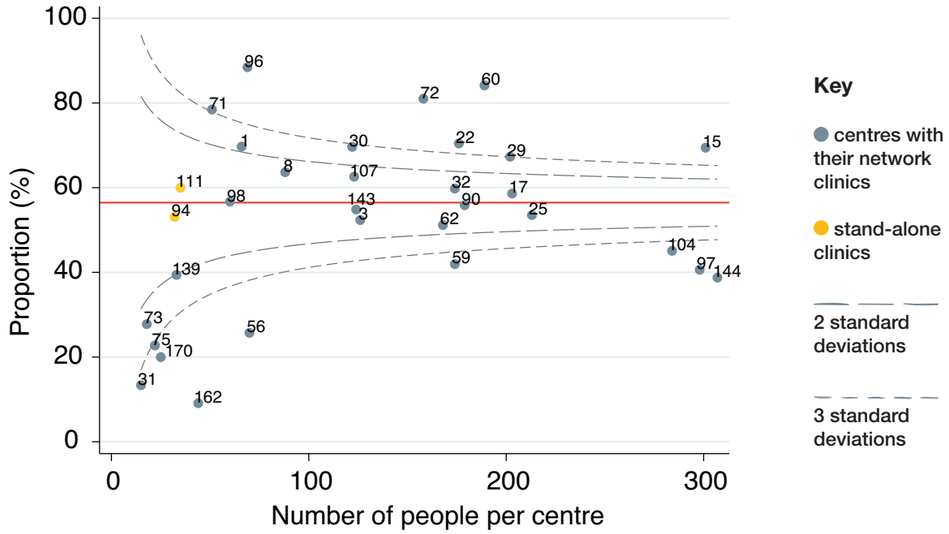
The mean BMI percentile for patients attending paediatric centres/clinics is 53.2%.

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



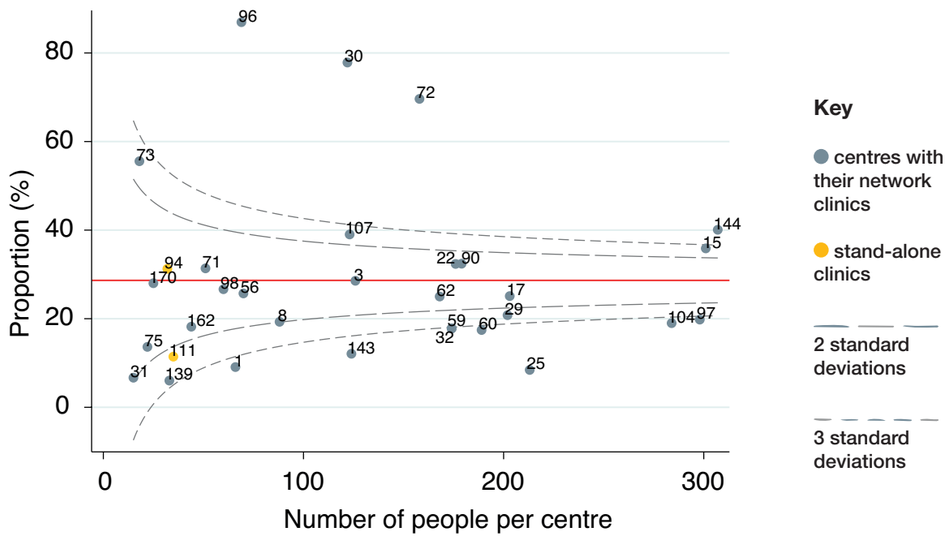
The proportion of patients with chronic *Pseudomonas aeruginosa* in paediatric centres/clinics is 7.5%.

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



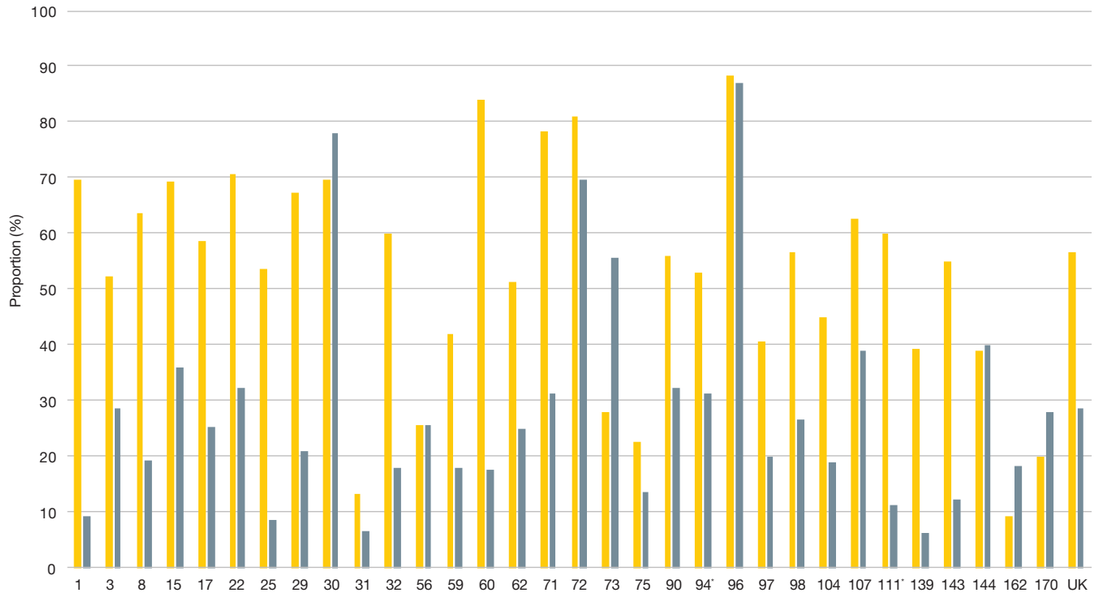
The proportion of patients receiving DNase treatment in paediatric centres/clinics is 56.5%.

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



The proportion of patients receiving DNase treatment in paediatric centres/clinics is 28.7%.

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



* Stand-alone clinics

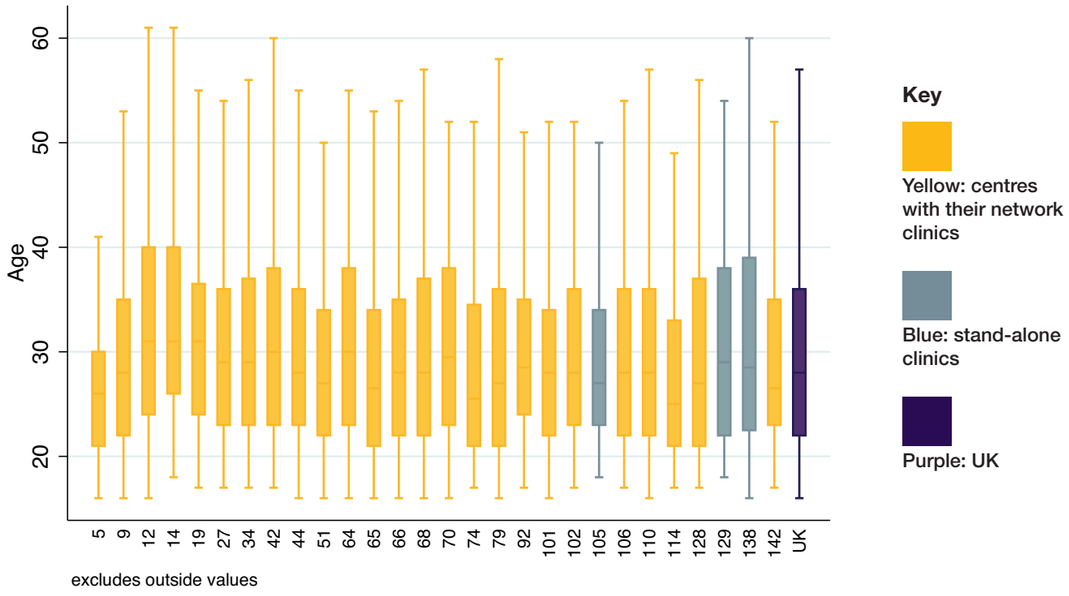
Section 3: Adult centre analyses

n=5546

In the UK CF care is led by 24 adult specialist CF centres. People with CF transfer to adult care centres between the ages of 16 and 18 years.

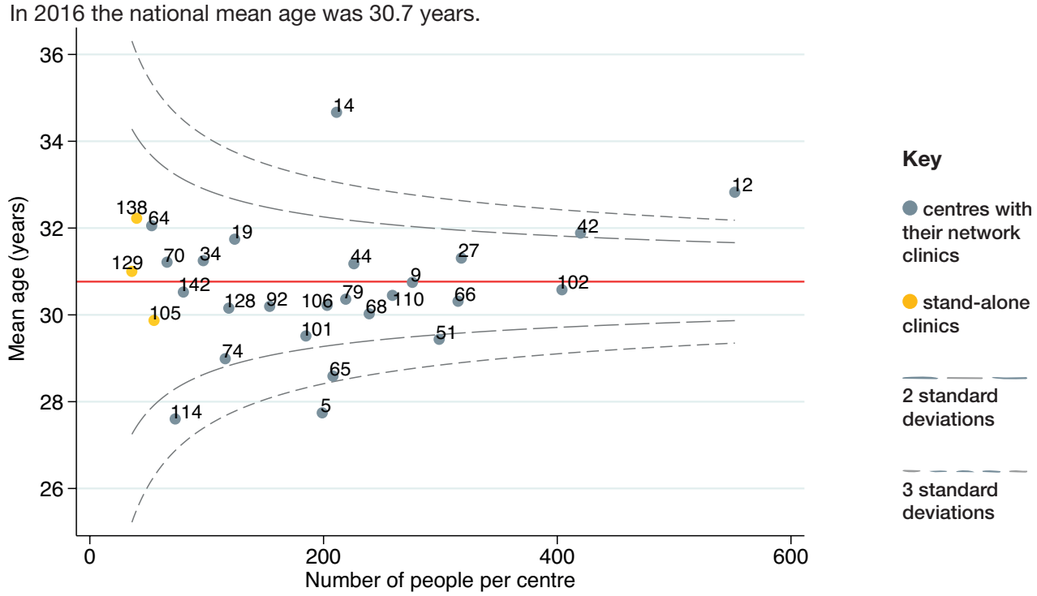
3.1 Mean age by adult services

The box-whisker plot shows the age distribution of patients within each centre/clinic.
In 2016 the median age in adults services was 29 years (IQR:22-36)



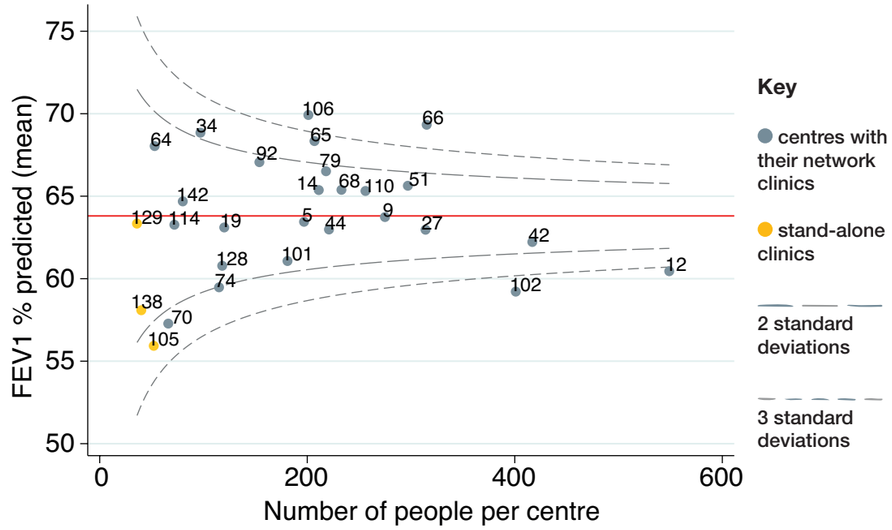
excludes outside values

The funnel plot below shows how the mean age in adult centres compares to the national mean.
In 2016 the national mean age was 30.7 years.



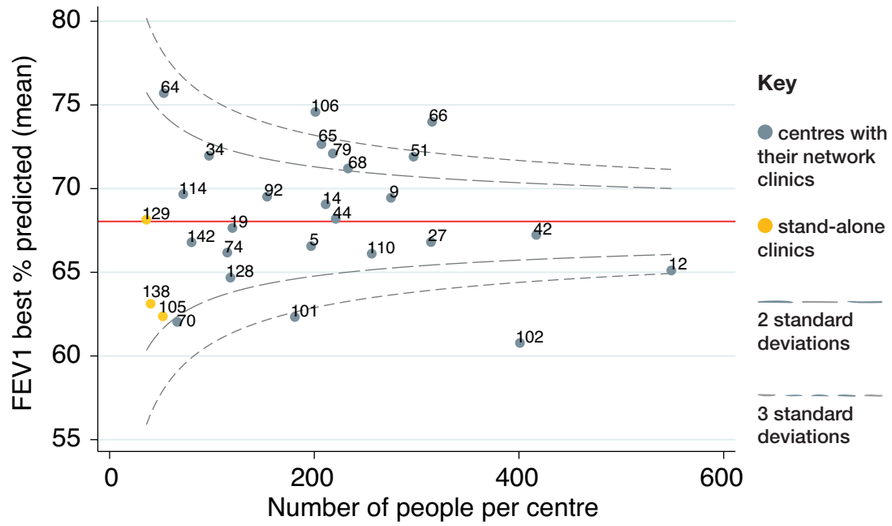
3.2a, 3.2b and 3.3 have been adjusted for age (using a linear term for age). This means that the data have been fine-tuned to take account of the different spread of ages across centres and clinics. The adjusted values are intended to show what the average lung function or BMI would be for that centre/clinic 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.

3.2a Age adjusted FEV_1% predicted at annual review by adults service (without a history of lung transplant)



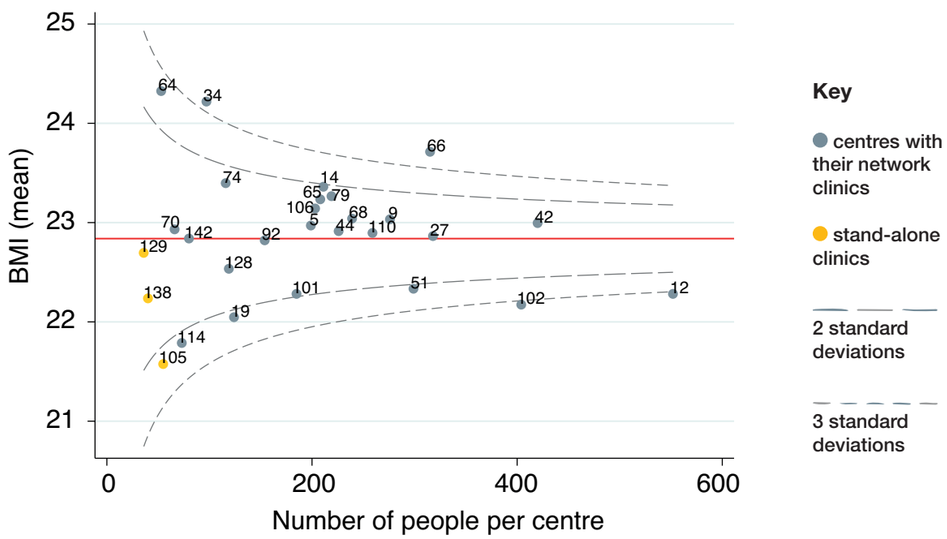
The mean FEV_1 (% predicted) in adult services is 63.8%.

3.2b Age adjusted best FEV_1% predicted by adults services (without a history of lung transplant).



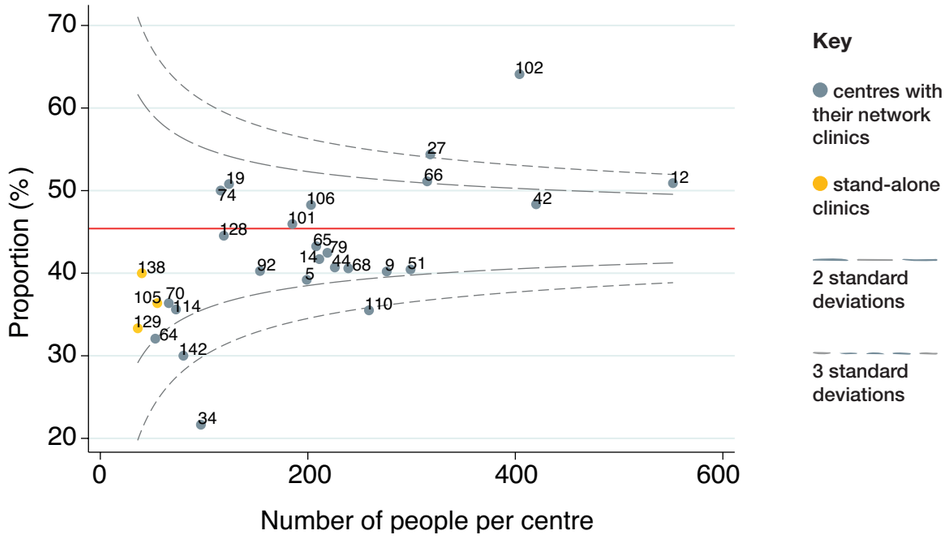
In 2016 the national mean was 68%. Where best FEV_1% predicted was missing the FEV_1% value at annual review was used.

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



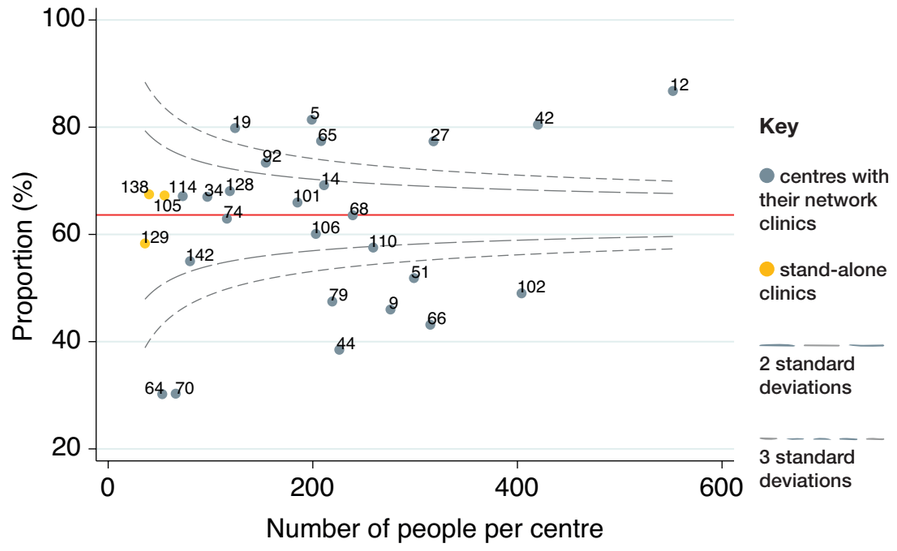
The mean BMI in adult services is 22.8.

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



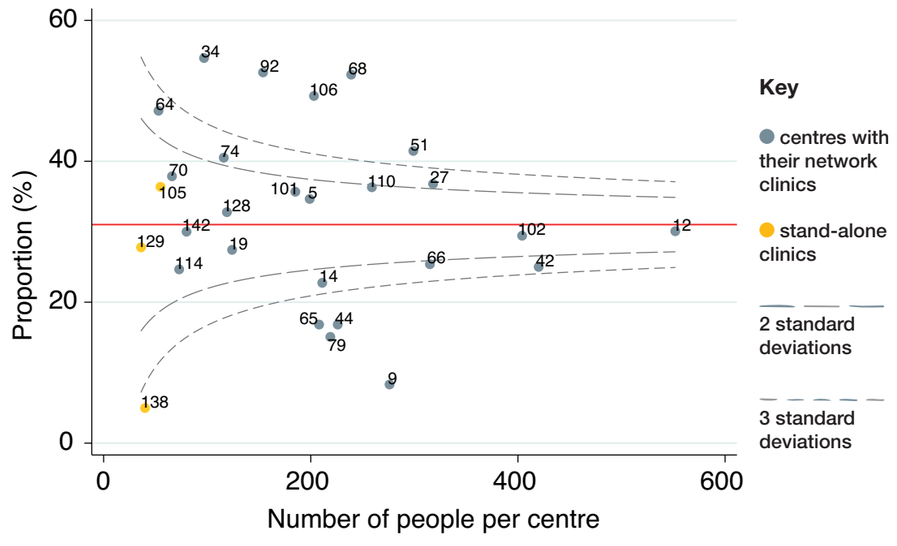
The proportion of patients with chronic *P. aeruginosa* in adult centres/clinics is 45.4%.

3.5 Proportion of patients receiving DNase treatment by adult service



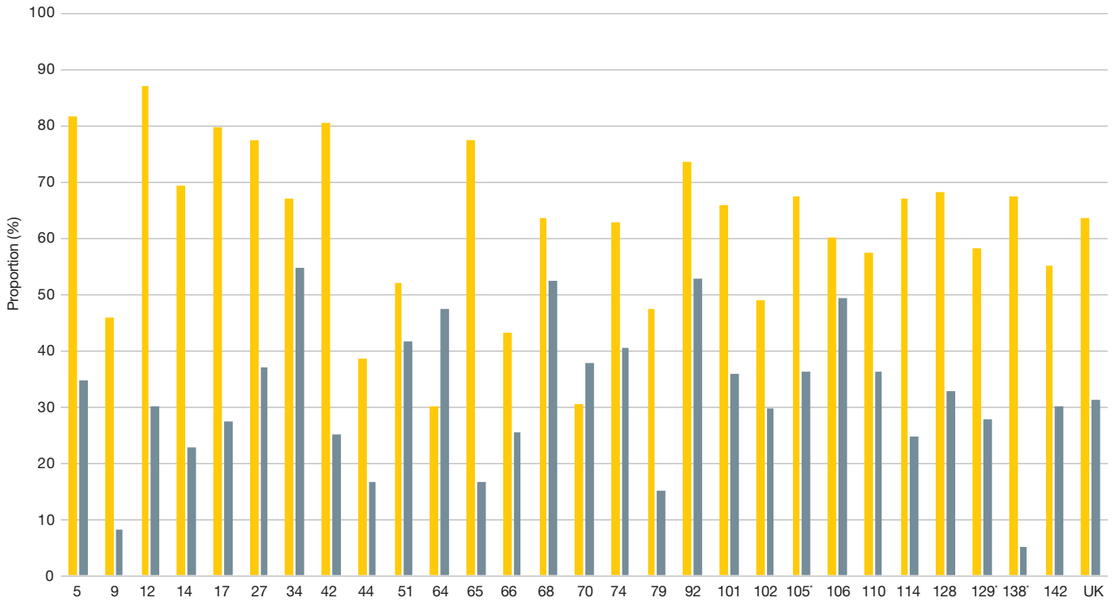
The proportion of patients receiving DNase treatment in adult centres/clinics is 63.6%.

3.6 Proportion of patients receiving hypertonic saline by adults service



The proportion of patients receiving hypertonic saline treatment in adult centres/clinics is 31%.

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



* Stand-alone clinics

Glossary

Words in this report that appear in this glossary are highlighted the first time they appear.

Word/Phrase	Meaning
2016	1 January 2016 – 31 December 2016
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. influenza	Haemophilus influenza is a bacterium that can cause serious illness.
Haemoptysis	The coughing up of blood.
Hepatobiliary disease	A liver or biliary disorder.

Heterozygous	Everyone living with cystic fibrosis has two mutations of the gene for CFTR, one inherited from their mother and one from their father. Someone who has two different mutations is heterozygous.
Word/Phrase	**Meaning**
Homozygous	Everyone living with cystic fibrosis has two mutations of the gene for CFTR, one inherited from their mother and one from their father. If both mutations (or genotypes) are the same, the person is said to be homozygous.
Hypertension	High blood pressure.
Incidence	The number of people newly diagnosed with a condition in the given year.
IQR (InterQuartile Range)	Also called the mid-spread, or middle fifty, IQR is a measure of the spread of data. It shows the difference between the upper and lower quartiles. IQR = Q3 – Q1.
Mean	A type of average, calculated by adding up all the values and dividing by the number of values.
Median	The middle number, when all numbers are arranged from smallest to largest.
Median age of death	Median age of death is based on the people with CF who died in any given year. So in 2016 the median age of the 148 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	Carrie or Sue	Gardner Sawyer	NHS England
Registry Data Manager †	Elaine	Gunn	Cystic Fibrosis Trust
Registry Development Manager †	Annie	Jeffery	Cystic Fibrosis Trust
Commissioner, Wales †	Claire	Nelson	NHS Wales
Analytical team representative †	Stephen and/or Margaret	Nyangoma Shi	Imperial College London
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 Data Manager †	Elaine	Gunn	Cystic Fibrosis Trust
Registry Development Manager †	Annie	Jeffery	Cystic Fibrosis Trust
Person with CF	Dominic	Kavanagh	Cystic Fibrosis Trust
Analytical team rep †	Stephen and/or Margaret	Nyangoma Shi	Imperial College London
Pharmacovigilance PI	Nicholas	Simmonds	Royal Brompton Hospital
CF physician - Adults			
CF physician - Adults*#	Martin	Wildman	Northern General Hospital

* Chair † Non-voting member # Caldicott guardian

Appendix 2: Data tables

Paediatric centres/clinics providing data in 2016 – ordered by clinic ID

Location	Centre/Clinic	ID	Active patients	Patients with 2016 data	FEV_1% predicted (≥6 years) (at annual review)		Best FEV_1% predicted (≥6 years)		BMI percentile (2-15 years)		Chronic Pseudomonas		Receiving DNase treatment		Receiving Hypertonic Saline treatment	
					Mean	Median	Mean	Median	Mean	Median	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)
Leicester	Leicester Royal Infirmary	1	71	66	89.9	92.6	95.0	96.4	59.0	64.4	9	13.6	46	69.7	6	9.1
Sheffield	Sheffield Children's Hospital	3	134	126	90.5	91.4	96.4	98.0	49.8	53.2	5	4.0	66	52.4	36	28.6
Stoke-on-Trent	Royal Stoke University Hospital	8	96	88	85.0	84.4	91.3	93.3	50.0	52.0	6	6.8	56	63.6	17	19.3
London: South West	Royal Brompton Hospital	15	337	301	86.7	89.7	91.0	91.7	56.4	59.1	42	14.0	209	69.4	108	35.9
London: South East	King's College Hospital	17	222	203	84.1	85.7	90.0	91.2	51.5	50.8	11	5.4	119	58.6	51	25.1
Oxford	John Radcliff Hospital	22	185	176	85.7	86.4	90.3*	91.5	47.6	48.4	8	4.5	124	70.5	57	32.4
Leeds	St James's University Hospital	25	224	213	83.1	87.0	90.9	93.6	48.6	50.3	10	4.7	114	53.5	18	8.5
Southampton	Southampton General Hospital	29	223	202	85.1	88.1	92.0	93.1	49.7	48.7	8	4.0	136	67.3	42	20.8
London: East	Royal London Hospital	30	139	122	90.4	88.2	95.7	95.2	53.3	55.8	12	9.8	85	69.7	95	77.9
Inverness	Raigmore Hospital	31	17	15	92.0	94.7	95.2	96.5	44.8	43.6	<5	20.0	<5	13.3	<5	6.7
Bristol	Bristol Royal Hospital for Children	32	194	174	83.0	82.6	90.8	91.3	48.7	46.1	24	13.8	104	59.8	31	17.8
Glasgow	Royal Hospital for Sick Children	56	93	70	90.7	90.9	96.4	94.3	47.9	43.3	0	0.0	18	25.7	18	25.7
Newcastle	Royal Victoria Infirmary	59	184	174	89.7	91.4	94.4	95.7	57.3	56.7	20	11.5	73	42.0	31	17.8
Belfast	Royal Belfast Hospital for Sick Children	60	205	189	88.9	90.3	94.1	95.0	57.2	57.4	15	7.9	159	84.1	33	17.5
Nottingham	Nottingham University Hospitals	62	173	168	83.7	86.4	87.1	88.3	53.5	56.4	8	4.8	86	51.2	42	25.0
Teeside	James Cook University Hospital	71	53	51	81.0	82.5	88.1	87.9	61.9	60.7	<5	7.8	40	78.4	16	31.4
Cardiff	Children's Hospital for Wales	72	172	158	84.3	84.5	84.3*	84.5	55.0	56.3	<5	1.9	128	81.0	110	69.6
Dundee	Ninewells Hospital	73	23	18	87.9	88.8	95.1	97.4	46.0	46.8	<5	11.1	5	27.8	10	55.6
Aberdeen	Royal Aberdeen Children's Hospital	75	30	22	78.3	78.8	83.4	85.5	47.6	42.3	0	0.0	5	22.7	<5	13.6
London: Central	Great Ormond Street Hospital for Children	90	188	179	84.3	84.3	91.3	93.3	45.1	40.4	9	5.0	100	55.9	58	32.4
Cornwall	Royal Cornwall Hospital	94	32	32	86.0	87.8	96.1	101.0	69.6	80.6	<5	12.5	17	53.1	10	31.3
Exeter	Royal Devon & Exeter Hospital	96	74	69	82.7	84.6	88.1	89.9	56.9	58.0	<5	4.3	61	88.4	60	87.0
Liverpool	Alder Hey Children's Hospital	97	316	298	82.4	85.6	89.2	92.0	56.2	55.7	33	11.1	121	40.6	59	19.8
Norwich	Norfolk & Norwich University Hospital	98	64	60	88.2	89.6	93.4	93.3	59.5	71.1	<5	6.7	34	56.7	16	26.7
Birmingham	Birmingham Children's Hospital	104	308	284	84.2	85.4	90.7	90.3	53.6	52.5	20	7.0	128	45.1	54	19.0
Cambridge	Addenbrookes Hospital	107	130	123	89.9	90.2	92.4	94.5	50.2	52.1	9	7.3	77	62.6	48	39.0
Hull	Hull Royal Infirmary	111	35	35	78.5	76.5	82.8	85.5	61.7	64.3	<5	8.6	21	60.0	<5	11.4
Plymouth	Derriford Hospital	139	39	33	75.3	71.8	88.0	93.3	49.1	40.2	0	0.0	13	39.4	<5	6.1
Edinburgh	Royal Hospital for Sick Children	143	132	124	89.2	89.6	92.5*	94.2	59.3	58.5	7	5.6	68	54.8	15	12.1
Manchester	Royal Manchester Children's Hospital	144	345	307	82.5	83.9	88.7	93.1	54.5	54.8	30	9.8	119	38.8	123	40.1
Lanarkshire	Wishaw General Hospital	162	45	44	90.8	91.5	95.0	96.9	46.5	42.3	<5	2.3	<5	9.1	8	18.2
Ayr	University Hospital Crosshouse	170	27	25	92.5	97.6	99.9	98.7	73.4	77.7	0	0.0	5	20.0	7	28.0

* 'Best' FEV_1% predicted data for these sites was less than 80% complete. Where 'Best' values were missing, FEV_1% predicted taken at annual review was used.

Appendix 2: Data tables

Adult centres/clinics providing data in 2016 – ordered by clinic ID

Location	Centre/Clinic	ID	Active patients	Patients with 2016 data	FEV_1% predicted (≥16 years) (at annual review)		Best FEV_1% predicted (≥16 years)		BMI percentile (≥16 years)		Chronic Pseudomonas		Receiving DNase treatment		Receiving Hypertonic Saline treatment	
					Mean	Median	Mean	Median	Mean	Median	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)
London: South East	King's College Hospital	5	222	199	64.3	67.1	67.6	71.8	22.7	22.1	78	39.2	162	81.4	69	34.7
Newcastle	Royal Victoria Infirmary	9	293	276	63.7	63.8	69.4	70.0	23.0	22.3	111	40.2	127	46.0	23	8.3
London South West	Royal Brompton Hospital	12	650	552	59.8	59.9	64.4	64.1	22.5	22.2	281	50.9	479	86.8	166	30.1
Belfast	Belfast City Hospital	14	271	211	64.1	64.7	67.7*	68.8	23.8	22.5	88	41.7	146	69.2	48	22.7
Frimley	Frimley Park Hospital	19	133	124	62.7	62.8	67.2	68.0	22.1	22.0	63	50.8	99	79.8	34	27.4
Birmingham	Birmingham Heartlands Hospital	27	338	318	62.8	63.5	66.6	67.0	22.9	22.3	173	54.4	246	77.4	117	36.8
Exeter	Royal Devon & Exeter Hospital	34	103	97	68.6	69.6	71.8	72.3	24.3	23.9	21	21.6	65	67.0	53	54.6
Leeds	St James's University Hospital	42	429	420	61.9	62.7	66.9	68.3	23.1	22.6	203	48.3	338	80.5	105	25.0
Edinburgh	Western General Hospital	44	235	226	62.9	63.5	68.1	68.1	22.9	22.4	92	40.7	87	38.5	38	16.8
Cambridge	Papworth Hospital	51	322	299	66.0	66.5	72.4	73.1	22.2	22.1	121	40.5	155	51.8	124	41.5
Plymouth	Derriford Hospital	64	55	53	67.6	70.2	75.2	78.5	24.5	23.9	17	32.1	16	30.2	25	47.2
Sheffield	Northern General Hospital	65	211	208	69.0	71.6	73.4*	77.2	23.0	22.1	90	43.3	161	77.4	35	16.8
Liverpool	Liverpool Heart and Chest Hospital	66	318	315	69.5	69.2	74.2	74.1	23.7	22.9	161	51.1	136	43.2	80	25.4
Llandough	Llandough Hospital	68	246	239	65.6	68.6	71.4	74.0	23.0	22.1	97	40.6	152	63.6	125	52.3
Aberdeen	Aberdeen Royal Infirmary	70	71	66	57.1	56.0	61.9	58.6	23.0	22.2	24	36.4	20	30.3	25	37.9
Stoke-on-Trent	Royal Stoke University Hospital	74	120	116	60.0	63.3	66.8	69.1	23.2	22.7	58	50.0	73	62.9	47	40.5
Glasgow	Gartnavel General Hospital	79	235	219	66.7	68.7	72.3	72.6	23.2	22.5	93	42.5	104	47.5	33	15.1
London: East	St. Bartholomew's Hospital	92	159	154	67.2	67.5	69.7	69.9	22.8	21.9	62	40.3	113	73.4	81	52.6
Nottingham	Nottingham University Hospitals	101	189	185	61.5	61.4	62.8*	64.3	22.2	22.0	85	45.9	122	65.9	66	35.7
Manchester	Wythenshawe Hospital	102	458	404	59.3	57.6	60.8	58.7	22.2	21.8	259	64.1	198	49.0	119	29.5
London: South East	University Hospital Lewisham	105	57	55	56.1	55.7	62.5	66.9	21.5	20.5	20	36.4	37	67.3	20	36.4
Bristol	Bristol Royal Infirmary	106	213	203	70.1	72.3	74.8	77.6	23.1	22.8	98	48.3	122	60.1	100	49.3
Southampton	Southampton General Hospital	110	266	259	65.4	68.6	66.3*	69.8	22.9	22.1	92	35.5	149	57.5	94	36.3
Norwich	Norfolk & Norwich University Hospital	114	74	73	64.3	64.4	70.7	72.8	21.5	21.0	26	35.6	49	67.1	18	24.7
Oxford	Churchill Hospital	128	124	119	61.0	59.0	65.0	64.8	22.5	22.0	53	44.5	81	68.1	39	32.8
Cornwall	Royal Cornwall Hospital	129	37	36	63.1	61.5	68.0	65.4	22.8	21.4	12	33.3	21	58.3	10	27.8
Hull	Castle Hill Hospital	138	42	40	57.7	53.5	62.7	58.0	22.4	21.6	16	40.0	27	67.5	2	5.0
Leicester	Glenfield Hospital	142	80	80	64.7	66.4	66.8	67.4	22.8	22.1	24	30.0	44	55.0	24	30.0

* 'Best' FEV_1% predicted data for these sites was less than 80% complete. Where 'Best' values were missing, FEV_1% predicted taken at annual review was used.

Appendix 2: Data tables

Paediatric centres/clinics providing data in 2016 – ordered alphabetically by country/city

Location	Centre/Clinic	ID	Active patients	Patients with 2016 data	FEV_1% predicted (≥6 years) (at annual review)		Best FEV_1% predicted (≥6 years)		BMI percentile (2-15 years)		Chronic Pseudomonas		Receiving DNase treatment		Receiving Hypertonic Saline treatment	
					Mean	Median	Mean	Median	Mean	Median	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)
England																
Birmingham	Birmingham Children's Hospital	104	308	284	84.2	85.4	90.7	90.3	53.6	52.5	20	7.0	128	45.1	54	19.0
Bristol	Bristol Royal Hospital for Children	32	194	174	83.0	82.6	90.8	91.3	48.7	46.1	24	13.8	104	59.8	31	17.8
Cambridge	Addenbrookes Hospital	107	130	123	89.9	90.2	92.4	94.5	50.2	52.1	9	7.3	77	62.6	48	39.0
Cornwall	Royal Cornwall Hospital	94	32	32	86.0	87.8	96.1	101.0	69.6	80.6	<5	12.5	17	53.1	10	31.3
Exeter	Royal Devon & Exeter Hospital	96	74	69	82.7	84.6	88.1	89.9	56.9	58.0	<5	4.3	61	88.4	60	87.0
Hull	Hull Royal Infirmary	111	35	35	78.5	76.5	82.8	85.5	61.7	64.3	<5	8.6	21	60.0	<5	11.4
Leeds	St James's University Hospital	25	224	213	83.1	87.0	90.9	93.6	48.6	50.3	10	4.7	114	53.5	18	8.5
Leicester	Leicester Royal Infirmary	1	71	66	89.9	92.6	95.0	96.4	59.0	64.4	9	13.6	46	69.7	6	9.1
Liverpool	Alder Hey Children's Hospital	97	316	298	82.4	85.6	89.2	92.0	56.2	55.7	33	11.1	121	40.6	59	19.8
London: South East	King's College Hospital	17	222	203	84.1	85.7	90.0	91.2	51.5	50.8	11	5.4	119	58.6	51	25.1
London: South West	Royal Brompton Hospital	15	337	301	86.7	89.7	91.0	91.7	56.4	59.1	42	14.0	209	69.4	108	35.9
London: Central	Great Ormond Street Hospital for Children	90	188	179	84.3	84.3	91.3	93.3	45.1	40.4	9	5.0	100	55.9	58	32.4
London: East	Royal London Hospital	30	139	122	90.4	88.2	95.7	95.2	53.3	55.8	12	9.8	85	69.7	95	77.9
Manchester	Royal Manchester Children's Hospital	144	345	307	82.5	83.9	88.7	93.1	54.5	54.8	30	9.8	119	38.8	123	40.1
Newcastle	Royal Victoria Infirmary	59	184	174	89.7	91.4	94.4	95.7	57.3	56.7	20	11.5	73	42.0	31	17.8
Norwich	Norfolk & Norwich University Hospital	98	64	60	88.2	89.6	93.4	93.3	59.5	71.1	<5	6.7	34	56.7	16	26.7
Nottingham	Nottingham University Hospitals	62	173	168	83.7	86.4	87.1	88.3	53.5	56.4	8	4.8	86	51.2	42	25.0
Oxford	John Radcliff Hospital	22	185	176	85.7	86.4	90.3*	91.5	47.6	48.4	8	4.5	124	70.5	57	32.4
Plymouth	Derriford Hospital	139	39	33	75.3	71.8	88.0	93.3	49.1	40.2	0	0.0	13	39.4	<5	6.1
Sheffield	Sheffield Children's Hospital	3	134	126	90.5	91.4	96.4	98.0	49.8	53.2	5	4.0	66	52.4	36	28.6
Stoke-on-Trent	Royal Stoke University Hospital	8	96	88	85.0	84.4	91.3	93.3	50.0	52.0	6	6.8	56	63.6	17	19.3
Southampton	Southampton General Hospital	29	223	202	85.1	88.1	92.0	93.1	49.7	48.7	8	4.0	136	67.3	42	20.8
Teeside	James Cook University Hospital	71	53	51	81.0	82.5	88.1	87.9	61.9	60.7	<5	7.8	40	78.4	16	31.4
Northern Ireland																
Belfast	Royal Belfast Hospital for Sick Children	60	205	189	88.9	90.3	94.1	95.0	57.2	57.4	15	7.9	159	84.1	33	17.5
Scotland																
Aberdeen	Royal Aberdeen Children's Hospital	75	30	22	78.3	78.8	83.4	85.5	47.6	42.3	0	0.0	5	22.7	<5	13.6
Ayr	University Hospital Crosshouse	170	27	25	92.5	97.6	99.9	98.7	73.4	77.7	0	0.0	5	20.0	7	28.0
Dundee	Ninewells Hospital	73	23	18	87.9	88.8	95.1	97.4	46.0	46.8	<5	11.1	5	27.8	10	55.6
Edinburgh	Royal Hospital for Sick Children	143	132	124	89.2	89.6	92.5*	94.2	59.3	58.5	7	5.6	68	54.8	15	12.1
Glasgow	Royal Hospital for Sick Children	56	93	70	90.7	90.9	96.4	94.3	47.9	43.3	0	0.0	18	25.7	18	25.7
Inverness	Raigmore Hospital	31	17	15	92.0	94.7	95.2	96.5	44.8	43.6	<5	20.0	<5	13.3	<5	6.7
Lanarkshire	Wishaw General Hospital	162	45	44	90.8	91.5	95.0	96.9	46.5	42.3	<5	2.3	<5	9.1	8	18.2
Wales																
Cardiff	Children's Hospital for Wales	72	172	158	84.3	84.5	84.3*	84.5	55.0	56.3	<5	1.9	128	81.0	110	69.6

* 'Best' FEV_1% predicted data for these sites was less than 80% complete. Where 'Best' values were missing, FEV_1% predicted taken at annual review was used.

Appendix 2: Data tables

Adult centres/clinics providing data in 2016 – ordered alphabetically by country/city

Location	Centre/Clinic	ID	Active patients	Patients with 2016 data	FEV_1% predicted (≥16 years) (at annual review)		Best FEV_1% predicted (≥16 years)		BMI percentile (≥16 years)		Chronic Pseudomonas		Receiving DNase treatment		Receiving Hypertonic Saline treatment	
					Mean	Median	Mean	Median	Mean	Median	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)
England																
Birmingham	Birmingham Heartlands Hospital	27	338	318	62.8	63.5	66.6	67.0	22.9	22.3	173	54.4	246	77.4	117	36.8
Bristol	Bristol Royal Infirmary	106	213	203	70.1	72.3	74.8	77.6	23.1	22.8	98	48.3	122	60.1	100	49.3
Cambridge	Papworth Hospital	51	322	299	66.0	66.5	72.4	73.1	22.2	22.1	121	40.5	155	51.8	124	41.5
Cornwall	Royal Cornwall Hospital	129	37	36	63.1	61.5	68.0	65.4	22.8	21.4	12	33.3	21	58.3	10	27.8
Exeter	Royal Devon & Exeter Hospital	34	103	97	68.6	69.6	71.8	72.3	24.3	23.9	21	21.6	65	67.0	53	54.6
Frimley	Frimley Park Hospital	19	133	124	62.7	62.8	67.2	68.0	22.1	22.0	63	50.8	99	79.8	34	27.4
Hull	Castle Hill Hospital	138	42	40	57.7	53.5	62.7	58.0	22.4	21.6	16	40.0	27	67.5	2	5.0
Leeds	St James's University Hospital	42	429	420	61.9	62.7	66.9	68.3	23.1	22.6	203	48.3	338	80.5	105	25.0
Leicester	Glenfield Hospital	142	80	80	64.7	66.4	66.8	67.4	22.8	22.1	24	30.0	44	55.0	24	30.0
Liverpool	Liverpool Heart and Chest Hospital	66	318	315	69.5	69.2	74.2	74.1	23.7	22.9	161	51.1	136	43.2	80	25.4
London: South East	University Hospital Lewisham	105	57	55	56.1	55.7	62.5	66.9	21.5	20.5	20	36.4	37	67.3	20	36.4
London: South East	King's College Hospital	5	222	199	64.3	67.1	67.6	71.8	22.7	22.1	78	39.2	162	81.4	69	34.7
London South West	Royal Brompton Hospital	12	650	552	59.8	59.9	64.4	64.1	22.5	22.2	281	50.9	479	86.8	166	30.1
London: East	St Bartholomew's Hospital	92	159	154	67.2	67.5	69.7	69.9	22.8	21.9	62	40.3	113	73.4	81	52.6
Manchester	Wythenshawe Hospital	102	458	404	59.3	57.6	60.8	58.7	22.2	21.8	259	64.1	198	49.0	119	29.5
Newcastle	Royal Victoria Infirmary	9	293	276	63.7	63.8	69.4	70.0	23.0	22.3	111	40.2	127	46.0	23	8.3
Norwich	Norfolk & Norwich University Hospital	114	74	73	64.3	64.4	70.7	72.8	21.5	21.0	26	35.6	49	67.1	18	24.7
Nottingham	Nottingham University Hospitals	101	189	185	61.5	61.4	62.8*	64.3	22.2	22.0	85	45.9	122	65.9	66	35.7
Oxford	Churchill Hospital	128	124	119	61.0	59.0	65.0	64.8	22.5	22.0	53	44.5	81	68.1	39	32.8
Plymouth	Derriford Hospital	64	55	53	67.6	70.2	75.2	78.5	24.5	23.9	17	32.1	16	30.2	25	47.2
Sheffield	Northern General Hospital	65	211	208	69.0	71.6	73.4*	77.2	23.0	22.1	90	43.3	161	77.4	35	16.8
Southampton	Southampton General Hospital	110	266	259	65.4	68.6	66.3*	69.8	22.9	22.1	92	35.5	149	57.5	94	36.3
Stoke-on-Trent	Royal Stoke University Hospital	74	120	116	60.0	63.3	66.8	69.1	23.2	22.7	58	50.0	73	62.9	47	40.5
Northern Ireland																
Belfast	Belfast City Hospital	14	271	211	64.1	64.7	67.7*	68.8	23.8	22.5	88	41.7	146	69.2	48	22.7
Scotland																
Aberdeen	Aberdeen Royal Infirmary	70	71	66	57.1	56.0	61.9	58.6	23.0	22.2	24	36.4	20	30.3	25	37.9
Edinburgh	Western General Hospital	44	235	226	62.9	63.5	68.1	68.1	22.9	22.4	92	40.7	87	38.5	38	16.8
Glasgow	Gartnavel General Hospital	79	235	219	66.7	68.7	72.3	72.6	23.2	22.5	93	42.5	104	47.5	33	15.1
Wales																
Llandough	Llandough Hospital	68	246	239	65.6	68.6	71.4	74.0	23.0	22.1	97	40.6	152	63.6	125	52.3

* 'Best' FEV_1% predicted data for these sites was less than 80% complete. Where 'Best' values were missing, FEV_1% predicted taken at annual review was used.

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