Cystic Fibrosis strength in numbers

UK Cystic Fibrosis Registry
2015 Annual Data Report

Published August 2016

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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

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

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Foreword

I am very pleased to introduce to you the UK Cystic Fibrosis Registry 2015 Annual Data Report.

The CF Registry plays an integral part in our wider fight for a life unlimited by cystic fibrosis (CF), by boosting world-class research, driving quality improvement in clinical care and directly empowering those living with the condition.

Yet it has the potential to provide even greater impact and we are investing significant resources to enhance its capability. The first phase of this was to move to a new software platform, completed in February this year. This will enable the Registry to collect clinical trial data from CF teams and quality of life data direct from those with cystic fibrosis who choose to provide it.

We are also developing a portal that will, in time, allow those with cystic fibrosis to access their own clinical data online. Keep an eye on our website, social media, and CF Life magazine for updates on all these developments.

It is heartening to see from this year's report the impact of the Trust's 'Genotype Matters' campaign, launched in 2015 .Today, more than 98% of people with cystic fibrosis on the UK CF Registry have a genotype recorded for both cystic fibrosis mutations. The completeness of these data are vital, to ensure people can gain access to the best medicines for them, and to further important research into new therapies. Our work continues to bring the completeness of these data as close to 100% as possible. Find out more at genotypematters.org.

As the lives and life choices of people with cystic fibrosis change, so the CF Registry needs to reflect it. For the first time this year, for example, the report includes information about pregnancies, 90 women with cystic fibrosis reported as having a live birth or still pregnant in 2015. For those wanting more information about fertility and family planning in cystic fibrosis, please download our new fertility booklet from cysticfibrosis.org.uk/fertility or contact our helpline by calling 020 3795 2184 or emailing helpline@cysticfibrosis.org.uk.

This report demonstrates the positive impact of national newborn screening for cystic fibrosis with more than 9 out of 10 five-year-olds with cystic fibrosis in 2015 having being diagnosed at birth. It also shows that 71% of people with cystic fibrosis over 16 are in work or study, with almost a third in full-time employment.

The Registry is an extraordinary asset to the UK CF community, and I would like to thank everyone who helps make it possible. From the multidisciplinary teams in CF centres throughout the UK to the thousands of generous supporters who raise money or donate and enable us to maintain the Registry – we couldn't do it without you. Above all, thank you to everyone living with cystic fibrosis and their friends and family who consent to their data being collected for this world-class resource; we will keep working to develop the potential of the Registry so that we can deliver maximum impact for everyone affected by cystic fibrosis.

Ed Owen
Chief Executive

Executive summary

The UK Registry annual report is designed to show a snap-shot of the health of people with cystic fibrosis (CF) in 2015, using data recorded at a person's annual review. This year we continue to show comparisons against previous years, demonstrating increasing numbers of people with CF, improvements in health outcomes and increased use of new therapies.

This year also sees the first publication of centre-level 'funnel plots', as well as the 'box and whisker' plots included in previous reports. These plots are designed to make it easier to understand the data shown for individual centres, and how they compare to the national medians. A guide and example plot is provided on page 39 of this report to explain how they should be read and interpreted.

2015 sees another rise in the CF population, with 10,810 now registered, allowing analysis of data from annual reviews of 9,587 people. Looking back to 2011, there has been an increase of over 1,000 people with CF in the UK, with nearly 60% of the UK CF population now aged 16 and over. With the growing importance of personalised medicines aimed at different genetic mutations, we are pleased to see such high levels of people with genotypes recorded (98%), and for the first time the top six genotypes represented across all four devolved nations are reported.

The completeness and quality of the data enables us to confidently evaluate changes in CF over time. The reduction in the rate of chronic Pseudomonas aeruginosa continues and is particularly noteworthy in the 16-19 age group, where the prevalence of this infection has reduced by 15.2% to 29.9% since 2008. Nontuberculous mycobacterium (NTM) figures for the last two years appear stable with a prevalence of 5.6% in 2015.

Comparisons with data from 2008 also show great strides in maintaining lung health, measured as FEV_1% predicted, shown in figure 1.11 as having a statistically significant improvement between these two years. There is an increase in overall treatment for CF-related diabetes (28%) since the 2011 data report (18.3%), which appears to reflect a rise in adolescents receiving treatment, up to 10% of the 10–16 year old population.

This report follows through on our previous commitment to move to new, more accurate median predicted survival figures using groups of three years' worth of data (section 1.32). The 2013–2015 group has a 47-year median predicted survival. A preliminary breakdown suggests there may also be a gender difference in survival, and an overall improvement over time. The summary table continues to show the single year calculation, as previously used in the reports.

Transplant activity has fallen, with 42 lung transplants being carried out in 2015 compared to 59 in 2014. This is in the context of a European Cystic Fibrosis Society Patient Registry Report using 2013 data that showed 3% of people in the UK living with a transplant, compared to 5.3% across Europe.

There are many other interesting examples of change in this report. We hope that you find the contents interesting and useful. We always look forward to hearing your views and welcome feedback.

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**.

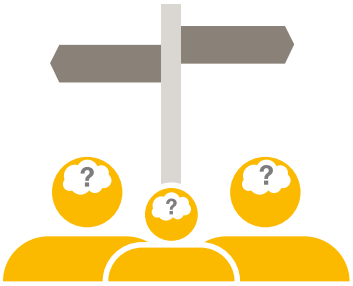
Cystic fibrosis

Cystic fibrosis is an inherited disease caused by a faulty gene known as 'CFTR'. The gene and the protein it makes control the movement of salt and water in and out of cells. When the gene is faulty, it can cause thicker mucus that blocks airways. This affects the lungs, which over time makes it hard to breathe. Around 85% people with cystic fibrosis 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 cystic fibrosis, 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 sub-committee of the RSC, known as the Registry Research Committee, assesses applications for data and guides the Registry research strategy.

Please see appendix 3 for members of each committee.

Data are only recorded on the UK CF Registry if explicit written consent is given by the person with cystic fibrosis 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 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.

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 cystic fibrosis, and the UK CF Registry, at **www.cysticfibrosis.org.uk/registry**.

A glossary of terms highlighted in teal in this report can be found on page 62.

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

	2011	2012	2013	2014	2015
CF patients registered[1] Excluding diagnoses that year	9749	10078 9804	10338 10076	10583 10356	10810 10586
CF patients with "complete" data; n(%)[2]	8679 (89%)	8794 (87%)	9052 (88%)	9432 (89%)	9587 (89%)
Age in years; median[3]	18	18	18	19	19
All newly diagnosed patients[4] **(newborn screening and other)**	261	285	301	291	224
Number of patients born identified by newborn screening[4]	203	213	177	164	112
Age at diagnosis in months; median[3]	3	3	3	2	2
Adults aged 16 yrs and over; %[3]	56.8	57.6	57.6	59.3	59.9
Males; %[3]	53.2	52.9	52.9	53.0	53.0
Genotyped; %[3]	95.6	96.2	97.2	97.7	98.1
Median predicted survival in years (95% Confidence interval)[5]	41.5 (35.7, 46.0)	43.5 (37.8, 49.9)	36.6 (34.4, 41.6)	40.1 (34.6, 46.7)	45.1 (39.9, 49.1)
Total deaths reported (%)[5]	118 (1.2%)	106 (1.1%)	146 (1.4%)	132 (1.2%)	125 (1.2%)
Age at death in years; median (95% CI)[3]	26	28 (25, 29)	29 (27, 31)	28 (25.5, 32)	28 (27, 33)

Notes:

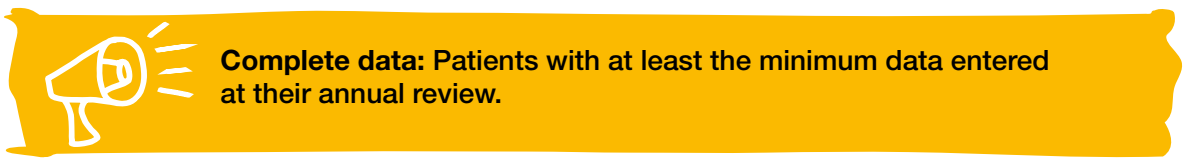
[1] The number of patients who were diagnosed with CF and had not died before 1 January in the given year.

[2] A patient has 'complete data' if their team has filled in an annual review for them for that year. Patients newly diagnosed in 2015 may not have their first annual review in the same year. If newly diagnosed patients are excluded, 91% of records are complete.

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

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

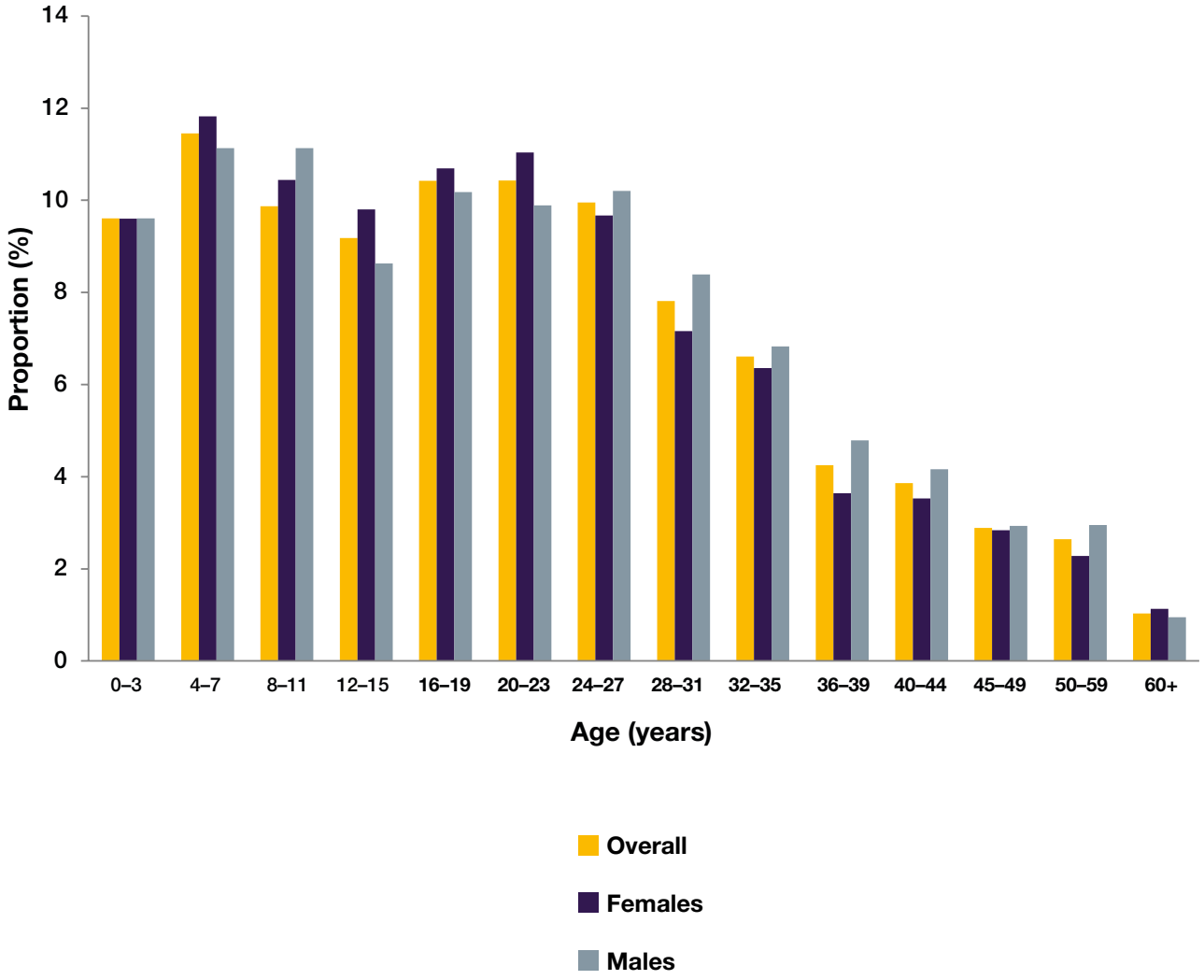
[5] Calculated from all patients registered on the database. This is the last year that survival will be shown as an annual figure. Please see section 1.31.



1.2 Age distribution by gender

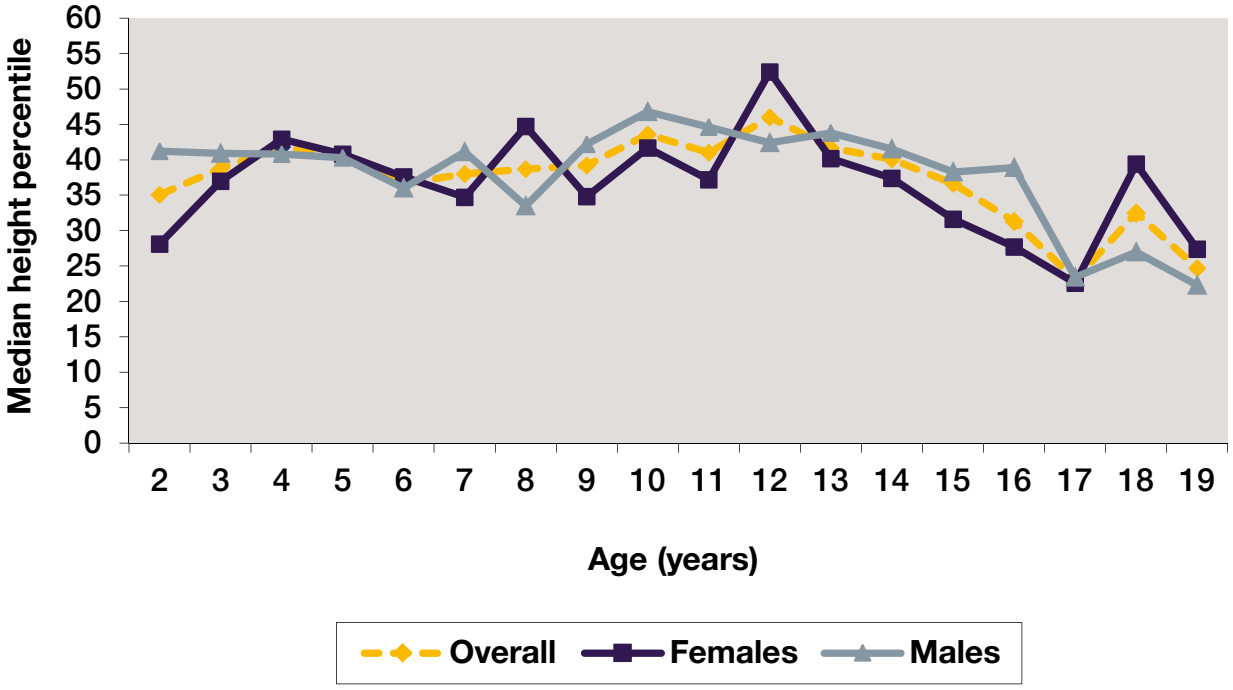
n=9587

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



1.3 Median height percentiles of children and young people (<20 years) n=4483

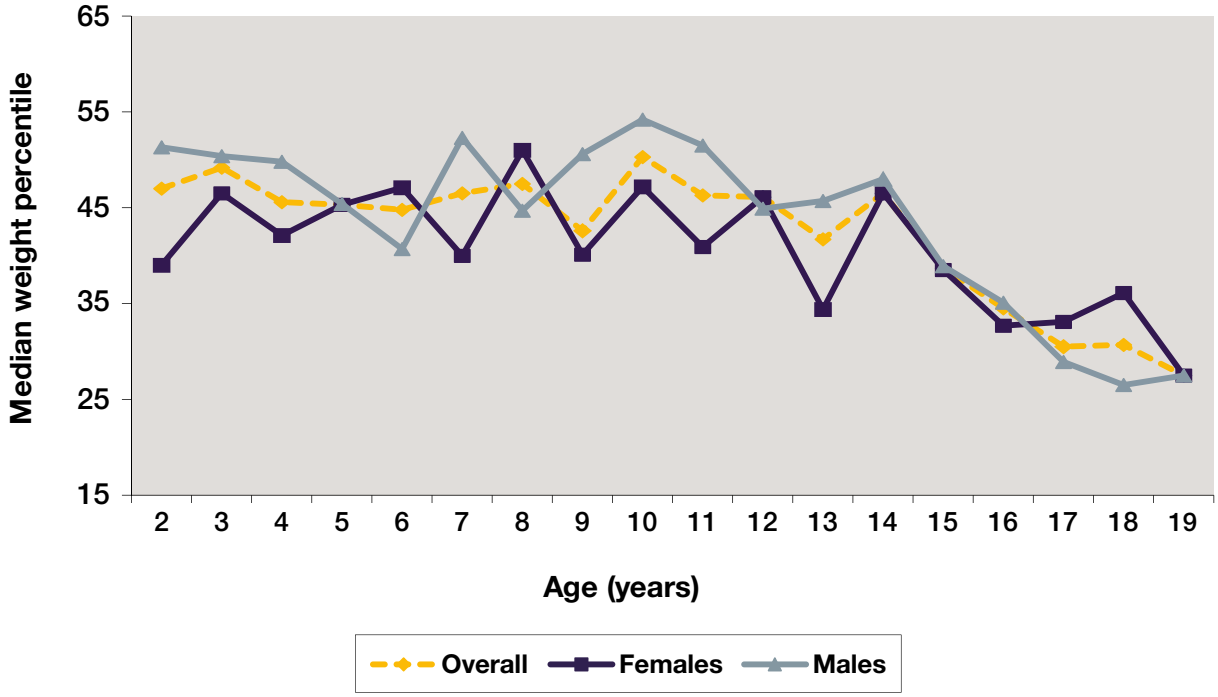
The following chart and table show the height percentiles of people with CF, aged 19 and under, in relation to the UK growth data for the general population. If a person with CF is on the 40th percentile, only 40% of people the same age are their height or shorter; 60% are taller.



	Overall			Female			Male		
Age	N	Median	IQR	N	Median	IQR	N	Median	IQR
2	271	35.1	12.6-67.0	136	28.1	10.4-65.8	135	41.2	18.1-69.0
3	290	38.8	14.8-59.9	136	37.0	16.4-58.1	154	40.9	14.1-61.2
4	289	41.8	18.8-68	139	42.9	18.0-69.8	150	40.8	20.7-66.0
5	279	40.6	18.7-63.8	130	40.8	15.6-64.3	149	40.3	19.2-63.6
6	263	36.6	16.7-62.8	141	37.6	16.3-63.0	122	36.0	16.8-62.7
7	267	38.0	17.6-61.0	123	34.7	14.5-58.6	144	41.1	19.8-62.0
8	254	38.7	15.7-65.7	125	44.7	17.0-67.8	129	33.5	13.1-62.4
9	229	39.2	15.5-63.4	114	34.8	14.1-58.7	115	42.2	16.2-64.7
10	228	43.6	23.3-65.5	104	41.7	27.1-58.0	124	46.8	22.6-73.5
11	235	41.0	17.5-70.6	128	37.2	16.7-69.8	107	44.6	17.8-73.5
12	222	46.0	14.5-71.5	111	52.4	17.7-75.0	111	42.4	13.5-69.3
13	214	41.7	14.7-67.9	112	40.2	14.5-65.0	102	43.8	15.7-71.5
14	229	40.1	14.7-65.5	111	37.4	14.8-64.7	118	41.5	14.5-68.0
15	215	36.6	11.9-67.3	108	31.6	8.1-60.9	107	38.3	17.6-70.6
16	255	31.3	11.4-60.6	126	27.7	9.7-56.3	129	38.9	13.5-67.8
17	236	23.1	7.8-49.2	106	22.6	7.2-65.9	130	23.4	8.7-41.9
18	252	32.5	11.6-59.2	115	39.4	13.7-65.3	137	27.0	9.7-54.2
19	255	24.7	9.1-57.7	134	27.4	7.7-59.0	121	22.8	9.1-53.8
Overall	**4483**	**37.1**	**14.4-63.9**	**2199**	**36.2**	**14.0-63.6**	**2284**	**37.6**	**15.0-64.2**

1.4 Median weight percentiles of children and young people (<20 years) n=4483

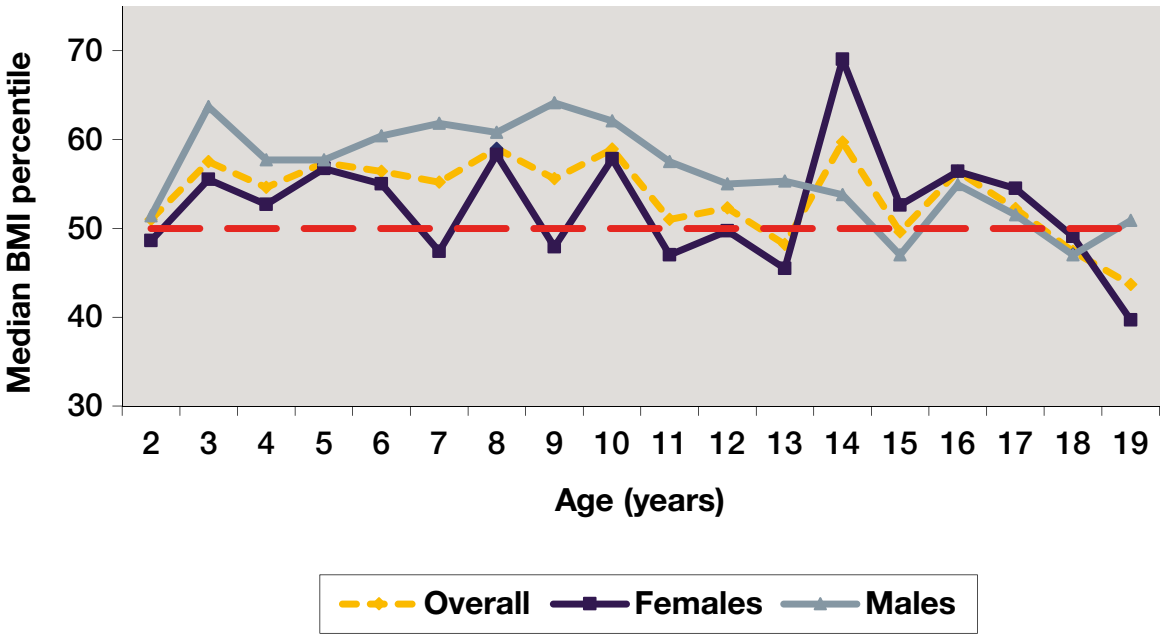
The following chart and table show the weight of people with CF, aged 19 and under, in relation to the UK growth data for the general population. If a person with CF is on the 40th percentile, only 40% of people the same age are their weight or lower; 60% weigh more.



	Overall			Female			Male		
Age	**N**	**Median**	**IQR**	**N**	**Median**	**IQR**	**N**	**Median**	**IQR**
2	271	47.0	18.9-75.6	136	39.0	13.4-73.9	135	51.3	22.3-75.8
3	290	49.2	26.6-72.4	136	46.5	26.0-71.8	154	50.4	28.7-74.1
4	289	45.6	22.6-74.1	139	42.1	21.6-76.8	150	49.8	24.4-73.2
5	279	45.3	25.2-72.3	130	45.3	24.6-70.8	149	45.4	25.3-73.8
6	263	44.8	20.1-71.7	141	47.1	22.4-70.1	122	40.7	17.5-75.8
7	267	46.5	22.3-68.0	123	40.0	20.7-64.5	144	52.3	24.2-69.9
8	254	47.5	21.3-71.4	125	51.0	21.2-74.6	129	44.7	21.0-70.3
9	229	42.6	23.6-68.4	114	40.1	16.0-64.8	115	50.6	29.0-75.9
10	228	50.3	28.7-76.9	104	47.2	26.1-72.7	124	54.2	28.9-78.9
11	235	46.3	19.4-71.9	128	40.9	16.9-70.2	107	51.5	26.2-75.0
12	222	46.1	23.8-68.7	111	46.1	19.8-66.7	111	44.9	26.7-70.2
13	214	41.7	15.9-70.8	112	34.4	14.3-67.0	102	45.7	18.6-72.4
14	229	46.5	19.6-77.7	111	46.5	22.6-78.8	118	48.0	18.4-75.2
15	215	38.7	15.2-70.0	108	38.5	14.8-72.5	107	38.9	16.7-67.3
16	255	34.5	12.7-67.2	126	32.7	14.5-61.0	129	35.1	10.1-71.6
17	236	30.5	7.8-57.9	106	33.1	9.6-67.1	130	28.9	5.4-56.7
18	252	30.7	9.6-61.8	115	36.1	11.6-62.7	137	26.5	7.3-58.3
19	255	27.5	7.7-62.3	134	27.5	7.8-54.8	121	27.5	4.9-63.8
Overall	**4483**	**43.0**	**18.7-70.5**	**2199**	**41.2**	**18.2-69.4**	**2284**	**44.9**	**19.0-71.5**

1.5 Median Body Mass Index (BMI) percentiles in children and young people (<20 years)
n=4483

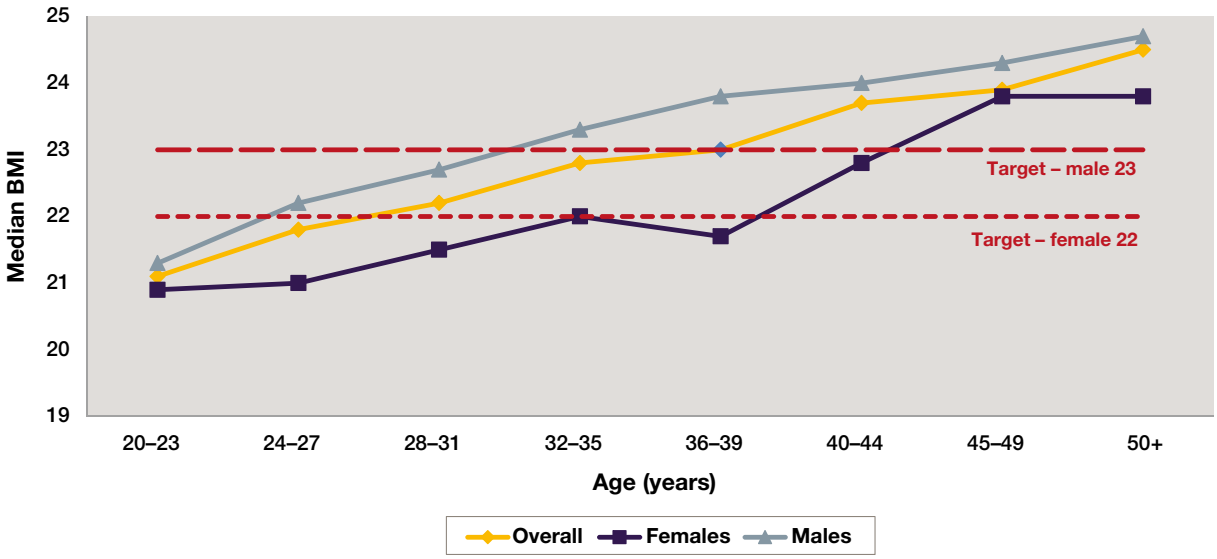
The following chart and table show the BMI percentiles of people with CF, aged 19 and under, in relation to the target BMI percentile for a healthy person of the same age (the 50th percentile, or the BMI percentile that half of the UK population people of that age has achieved). If a person with CF is 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	271	51.0	26.5-70.6	136	48.6	23.8-70.3	135	51.4	30.7-71.2
3	290	57.5	34.8-77.3	136	55.5	33.1-74.9	154	63.7	35.1-77.9
4	289	54.6	31.7-75.8	139	52.7	31.8-73.5	150	57.7	31.7-79.5
5	279	57.4	33.3-76.3	130	56.7	31.9-74.8	149	57.7	34.1-78.0
6	263	56.4	29.2-75.4	141	55.0	27.1-74.7	122	60.4	33.3-77.4
7	267	55.2	32.9-81.2	123	47.4	30.7-79.0	144	61.8	34.1-83.6
8	254	59.0	33.7-78.9	125	58.3	38.4-78.7	129	60.8	28.9-79.2
9	229	55.6	31.3-76.6	114	47.9	23.8-68.8	115	64.1	39.8-80.0
10	228	58.9	37.7-84.2	104	57.8	31.7-83.4	124	62.1	41.9-85.9
11	235	51.0	25.1-77.1	128	47.0	19.5-76.1	107	57.5	34.7-79.2
12	222	52.3	29.9-75.1	111	49.7	27.9-71.0	111	55.0	33.6-79.1
13	214	48.2	27.9-78.0	112	45.5	24.5-74.9	102	55.3	30.5-79.1
14	229	59.7	30.0-83.5	111	69.0	35.5-84.6	118	53.8	26.0-80.7
15	215	49.6	26.6-80.5	108	52.6	29.5-81.8	107	47.0	21.5-79.3
16	255	56.4	31.0-80.7	126	56.4	33.5-80.5	129	54.9	24.1-81.0
17	236	52.2	24.7-76.6	106	54.5	27.6-72.3	130	51.5	18.6-80.5
18	252	47.4	20.7-71.6	115	49.1	19.3-69.7	137	47.0	21.5-74.2
19	255	43.7	13.9-72.0	134	39.7	13.7-71.0	121	50.9	16.4-74.8
Overall	**4483**	**54.3**	**29.1-77.5**	**2199**	**52.4**	**27.9-75.2**	**2284**	**56.7**	**30.7-79.1**

1.6 Median Body Mass Index (BMI) values among adults (20 years and over)
n=4743

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[6].



	Overall			Female			Male		
Age	**N**	**Median**	**IQR**	**N**	**Median**	**IQR**	**N**	**Median**	**IQR**
20-23	1000	21.4	19.7-23.7	498	21.2	19.7-23.3	502	21.6	19.8-23.9
24-27	954	21.7	19.7-24.1	436	21.4	19.3-23.5	518	22.1	20.1-24.6
28-31	749	22.3	20.3-24.6	323	21.6	19.6-23.8	426	22.7	20.7-24.9
32-35	634	22.8	20.9-25.3	287	22.2	20.4-24.5	347	23.2	21.2-25.6
36-39	407	23.1	20.8-25.6	164	21.6	19.9-24.1	243	24.1	22.0-26.0
40-44	370	23.8	21.7-26.0	159	22.7	20.7-25.9	211	24.3	22.2-26.2
45-49	277	23.9	21.6-26.4	128	23.7	21.2-25.8	149	24.1	22.0-26.6
50+	352	24.6	22.1-27.4	154	24.2	21.2-27.4	198	24.8	22.6-27.4
Overall	**4743**	**22.4**	**20.4-24.9**	**2149**	**21.8**	**19.9-24.3**	**2594**	**22.9**	**20.9-25.3**

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

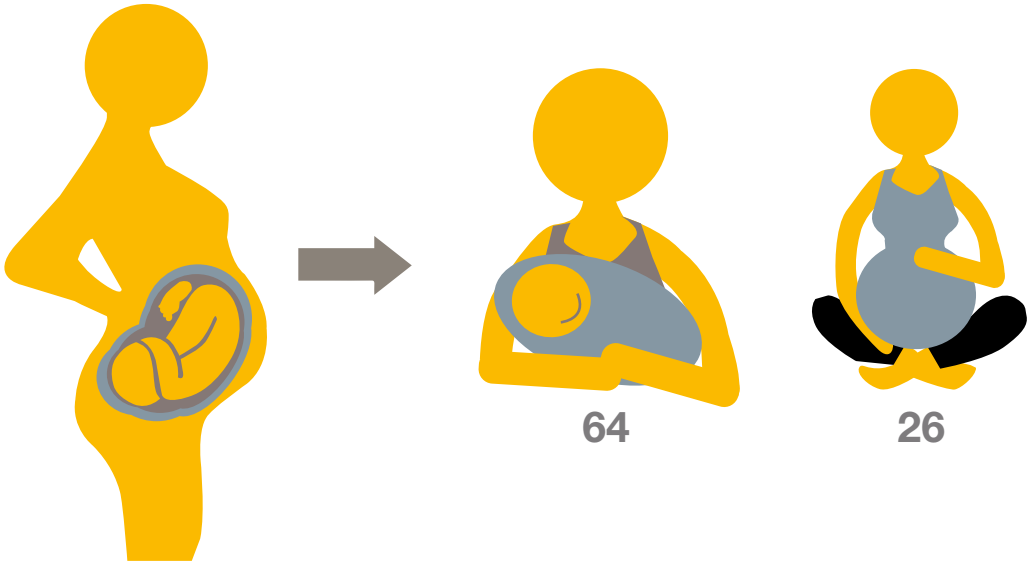
1.7 Education and employment in adults (16 years and over) n=4930

The following table shows how people with cystic fibrosis reported their education and employment status in 2015. Please note that the groups are not mutually exclusive; someone may be a student as well as working part-time, for example.

	2011	2012	2013	2014	2015
	Number of patients n (%) (n=4933)	Number of patients n (%) (n=5062)	Number of patients n (%) (n=5213)	Number of patients n (%) (n=5592)	Number of patients n (%) (n=5742)
Number of people who completed questionnaire	4406	4254	4346	4623	4930
Full-time employment	1436 (29.1)	1425 (28.2)	1502 (28.8)	1634 (29.2)	1811 (31.5)
Part-time employment	706 (14.3)	653 (12.9)	664 (12.7)	703 (12.6)	768 (13.4)
Student	933 (18.9)	917 (18.1)	922 (17.7)	976 (17.5)	927 (16.1)
Homemaker	216 (4.4)	231 (4.6)	232 (4.5)	258 (4.6)	264 (4.6)
Unemployed	793 (16.1)	684 (13.5)	685 (13.1)	821 (14.7)	761 (13.3)
Disabled	255 (5.2)	273 (5.4)	298 (5.7)	272 (4.9)	365 (6.4)
Retired	78 (1.6)	75 (1.5)	78 (1.5)	85 (1.5)	108 (1.9)
'Unknown' entered	548 (11.1)	862 (17.0)	914 (17.5)	930 (16.6)	850 (14.8)
No data recorded	76 (1.5)	38 (0.8)	21 (0.4)	39 (0.7)	27 (0.5)
Number of people in work or study	**3111 (70.6)**	**3020 (71.0)**	**3098 (71.3)**	**3243 (70.1)**	**3489 (70.8)**

1.8 Pregnancy

At the time of their 2015 annual review, 64 women with cystic fibrosis had a live birth. 26 women were still pregnant.



Diagnosis of cystic fibrosis

1.9 Age at diagnosis and screening in children under 16 in 2015 n=3780 with a diagnosis date

Newborn screening for CF has been done routinely in the whole of the UK since mid 2007. It is part of the heel prick blood spot testing done at 5-7 days of age. The blood sample is tested for a number of conditions, including cystic fibrosis. This means that more babies born after 2007 receive an early diagnosis than those born before.

Age at diagnosis	All patients <16; n (%)	Patients aged 10 years in 2015; n (%)	Patients aged 5 years in 2015; n (%)
Pre-natal	<5	0 (0)	0 (0)
Birth-3 months	2921 (77.3)	144 (63.4)	255 (92.7)
4-6 months	174 (4.6)	16 (7.0)	<5
7-12 months	130 (3.4)	14 (6.2)	<5
1 yr	187 (4.9)	15 (6.6)	5 (1.8)
2 yrs	127 (3.4)	14 (6.2)	<5
3 yrs	70 (1.9)	6 (2.6)	<5
4 yrs	48 (1.3)	5 (2.2)	<5
5 yrs	28 (0.7)	<5	<5
6 yrs	24 (0.6)	<5	-
7 yrs	13 (0.3)	<5	-
8 yrs	19 (0.5)	0 (0)	-
9 yrs	14 (0.4)	<5	-
10 yrs	9 (0.2)	<5	-
11 yrs	<5	-	-
12 yrs	<5	-	-
13 yrs	<5	-	-
14 yrs	<5	-	-
15 yrs	-	-	-
Overall	**3780**	**227**	**275**

The median (range) age at diagnosis for patients aged under 16 in 2015 is 26 days (15-92 days).

Diagnosis in the first three months of life is more common in children aged 5 years in 2015 (born after the UK-wide newborn screening programme was implemented) than in children aged 10 years in 2015 (born before the UK-wide newborn screening programme was put in place nationally).

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 2014 is higher in this report than was recorded in the report published in 2015. It is likely that the 2015 figure will be updated in the next annual report.

1.10 Age at diagnosis and screening in adults aged 16 and over in 2015 n=5684 with a diagnosis date

The table below shows the age that people aged 16 and over in 2015 were when they were diagnosed. People aged 16 or over in 2015 were born before newborn screening was done routinely in the UK.

Age at diagnosis	n (%)
Pre-natal	0 (0)
Birth-3 months	2297 (40.4)
4-6 months	525 (9.2)
7-12 months	349 (6.1)
1 yr	482 (8.5)
2 yrs	293 (5.2)
3 yrs	222 (3.9)
4 yrs	176 (3.1)
5 yrs	86 (1.5)
6 yrs	83 (1.5)
7 yrs	58 (1.0)
8 yrs	67 (1.2)
9 yrs	48 (0.8)
10 yrs	38 (0.7)
11 yrs	40 (0.7)
12 yrs	41 (0.7)
13 yrs	47 (0.8)
14 yrs	33 (0.6)
15 yrs	44 (0.8)
16-20 yrs	160 (2.8)
21-25 yrs	109 (1.9)
26-30 yrs	105 (1.8)
31-35 yrs	115 (2.0)
36-40 yrs	83(1.5)
41-45 yrs	62 (1.1)
46-50 yrs	32 (0.6)
51-60 yrs	89 (1.6)
61 yrs+	0 (0)
Overall	**5684**

In 2015 29 people aged 16 or over were newly diagnosed with CF. 755 (13.3%) of adults with CF were diagnosed after the age of 16.

Lung health

For people with cystic fibrosis mucus in the lungs is linked to repeat or chronic infections, which 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 an exhaled breath. In this report, an FEV_1% predicted is based on the FEV_1 we would expect for a person without cystic fibrosis 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, maintaining an FEV_1% predicted of 85% or higher is the target, as this indicates normal or near-normal lung health.

Most people can continue to lead a relatively normal life, including going to school or work, with 50% of their predicted FEV_1. Once FEV_1 is lower than 50% of the predicted value, it becomes difficult to lead a normal life. If FEV_1 declines to 30% or less, a patient may be considered for lung transplant.

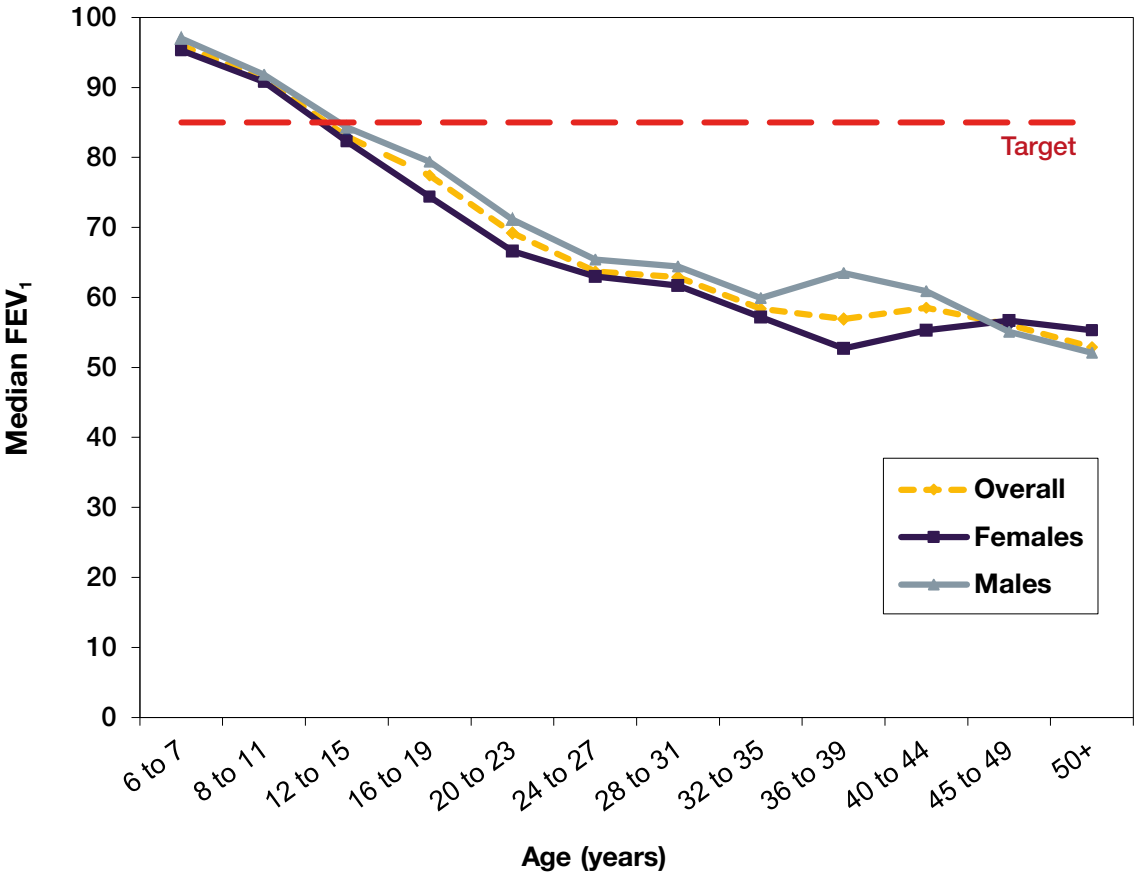
An aim of CF care is to prevent FEV_1% predicted from falling as much as possible, for as long as possible. This is a team effort between people with CF, their family, and their medical team, which can include doctors, nurses, physiotherapists, dietitians, and psychologists.

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

1.11 Median FEV_1% predicted (GLI equations) among people aged 6 years and over

n=7625

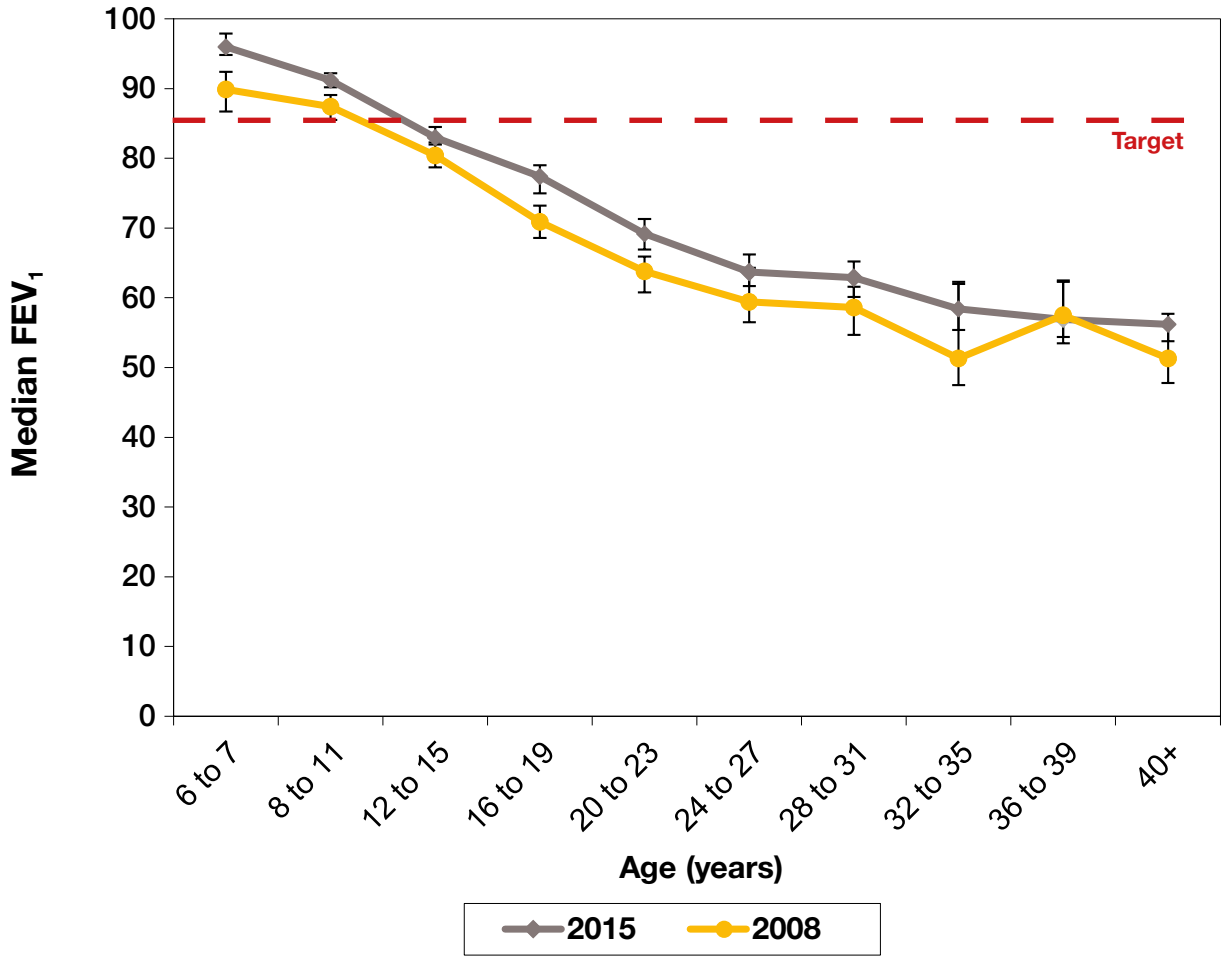
The chart and table in this section show information about those patients whose FEV_1 data were complete. People with CF who have had lung transplants are excluded, as their new 'non-CF' lungs would have lung health similar to a person without cystic fibrosis.



	Overall			Female			Male		
Age (yrs)	**N**	**Median**	**IQR**	**N**	**Median**	**IQR**	**N**	**Median**	**IQR**
6-7	518	96.0	84.3-107.3	259	95.3	83.2-107.3	259	97.0	86.1-106.4
8-11	924	91.2	80.9-100.0	461	90.8	79.6-99.3	463	91.8	82.5-100.4
12-15	866	83.0	71.2-94.9	435	82.3	70.9-93.5	431	84.3	71.7-96.1
16-19	967	77.4	58.9-90.9	463	74.4	53.9-88.9	504	79.4	61.5-92.6
20-23	955	69.2	50.7-86.1	473	66.6	49.5-83.2	482	71.1	52.3-87.8
24-27	903	63.7	45.5-83.1	413	63.0	46.4-82.8	490	65.4	45.0-83.5
28-31	690	62.9	41.7-80.2	291	61.7	41.7-83.0	399	64.4	41.6-78.4
32-35	574	58.4	40.2-79.9	261	57.2	42.2-76.8	313	59.9	38.0-80.8
36-39	370	56.9	41.4-79.1	147	52.7	39.4-74.2	223	63.5	42.3-81.4
40-44	315	58.5	41.9-77.6	132	55.3	42.0-75.3	183	60.9	41.6-79.7
45-49	232	56.1	40.5-74.4	108	56.7	43.0-73.7	124	55.1	38.5-75.3
50+	311	52.9	38.8-78.8	136	55.3	41.4-77.2	175	52.1	36.3-80.7
Overall	**7625**	**78.4**	**55.4-95.7**	**3579**	**77.4**	**54.9-94.6**	**4046**	**79.2**	**56.1-96.2**

1.12 Median FEV_1% predicted over time (GLI equations) among people aged 6 and over (excluding patients post lung transplant) n=7625 in 2015, n=4388 in 2008

As we learn more about cystic fibrosis and how to treat it, we hope to improve the outcomes of people with the condition. The chart below shows how FEV_1 in 2015 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.

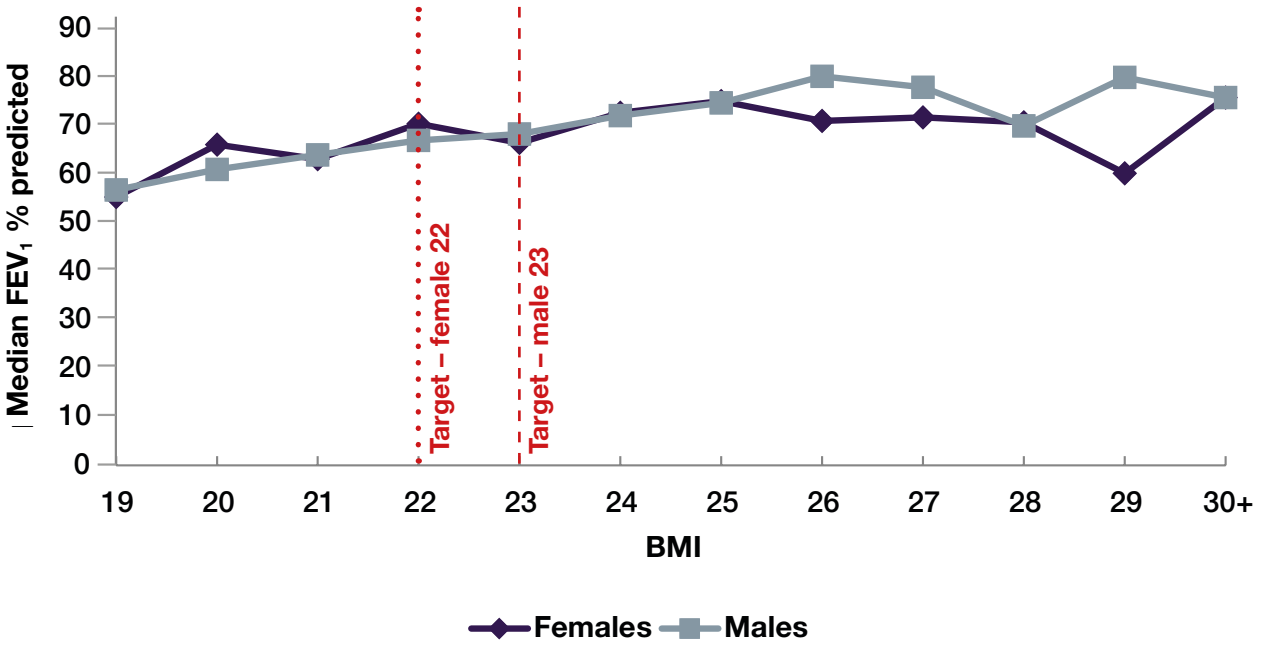


An analysis was conducted in order to determine whether there were statistically significant differences in FEV_1 (% predicted) in 2015 compared to 2008 by age category. The results show that there was a small, but statistically significant difference in the age bands where the p value is less than 0.05.

	Age (years)									
	6-7	8-11	12-15	16-19	20-23	24-27	28-31	32-35	36-39	40+
p-value	0.011	0.016	0.205	0.008	0.000	0.085	0.109	0.008	0.819	0.142

1.13 Median FEV_1% predicted and BMI in people aged 16 and over (excluding patients post-lung transplant) n=7625

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 appears to 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 all BMI≥30 are grouped into one.

1.14 Median best FEV_1% predicted in people aged 6 and over n=5970

The following table shows the highest FEV_1% predicted value recorded during the year for that patient. Other lung health calculations use the value that was taken during the person's annual review, which may not represent their best value during the year. People who are recorded as having had a lung transplant are excluded.

	Overall			Female			Male		
	N	Median	IQR	N	Median	IQR	N	Median	IQR
Overall	5970	79.0	58.7-93.4	2808	78.7	58.0-93.2	3162	79.3	59.2-93.7

Lung infections

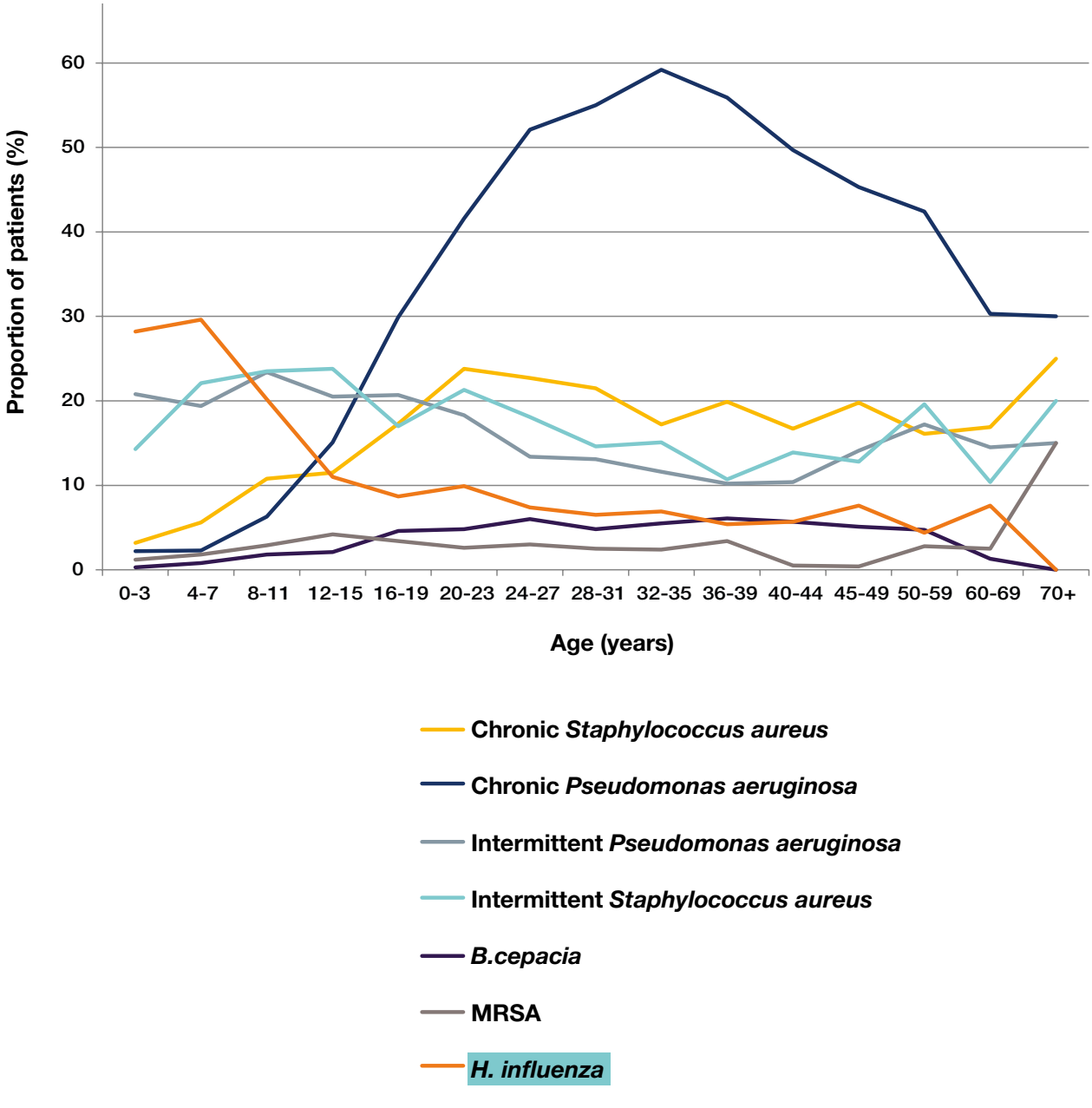
Lung infections can permanently reduce lung function in people with cystic fibrosis. Some lung infections can become 'chronic', meaning that they can't ever be removed completely using medicines.

1.15 Lung infections in 2015
n=9587

Chronic infection with *Pseudomonas aeruginosa* or *Staphylococcus aureus* is defined as three or more positive samples in the last year.

In 2015 only 6.1% of children (aged under 16) had chronic *Pseudomonas*. 46% of adults aged 16 and over were recorded as having chronic *Pseudomonas*.

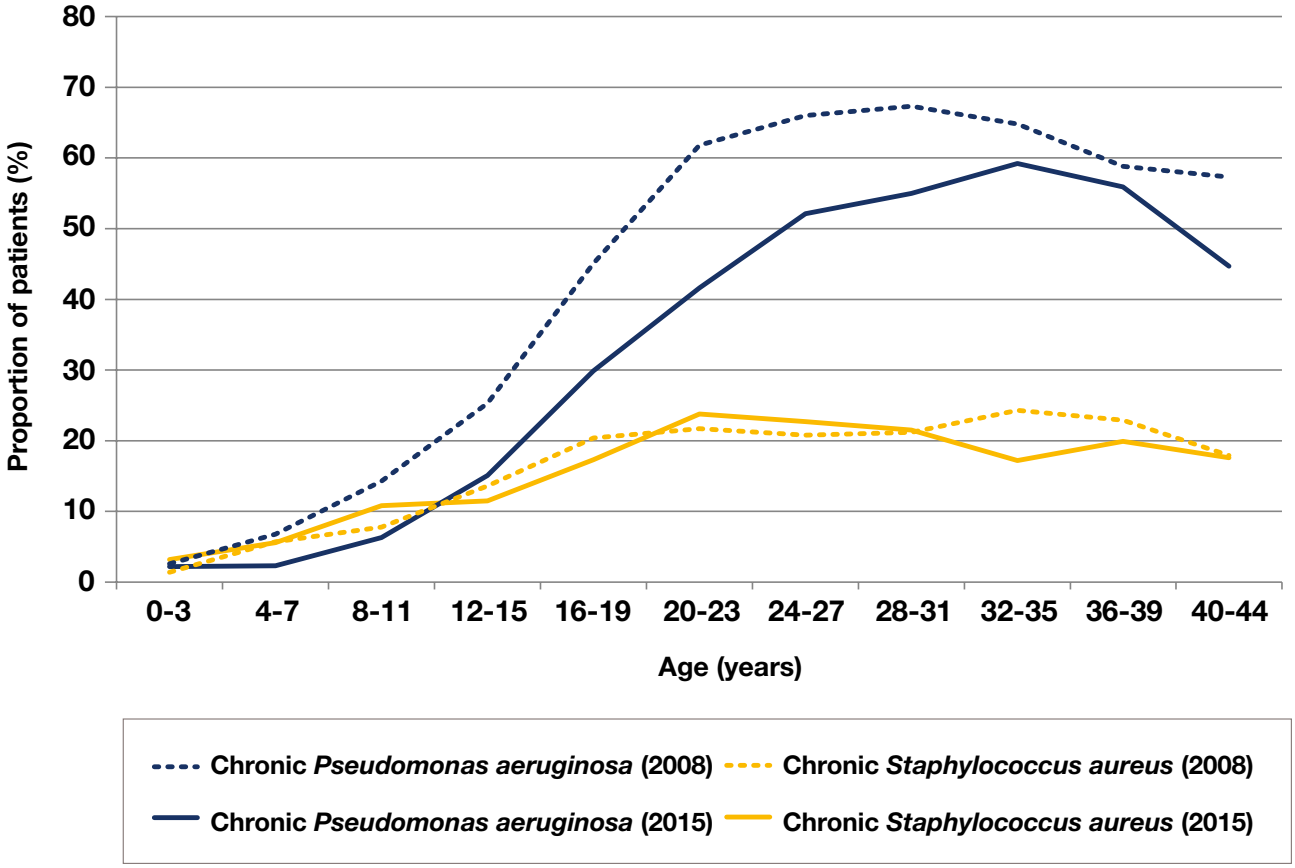
There is a steady increase with age in those with chronic *Pseudomonas*, peaking at 59% in those aged 32-35 years.



	Age (years)													Overall	
	0-3	4-7	8-11	12-15	16-19	20-23	24-27	28-31	32-35	36-39	40-44	45-49	50+	Children (<16 years)	Adults (≥16 years)
N patients in age bands	921	1098	946	880	999	1000	954	749	634	407	370	277	352	3845	5742
Chronic *S. aureus*; n (%)	29 (3.2)	60 (5.6)	100 (10.8)	100 (11.5)	170 (17.3)	234 (23.8)	212 (22.7)	158 (21.5)	106 (17.2)	80 (19.9)	61 (16.7)	54 (19.8)	58 (16.8)	289 (7.7)	1133 (20.1)
Chronic *P. aeruginosa*; n (%)	20 (2.2)	25 (2.3)	58 (6.3)	131 (15.1)	295 (29.9)	409 (41.6)	486 (52.1)	403 (55.0)	367 (59.2)	224 (55.9)	181 (49.7)	125 (45.3)	135 (39.0)	234 (6.2)	2625 (46.5)
Interrmittent *P. aeruginosa*; n (%)	189 (20.8)	208 (19.4)	216 (23.4)	178 (20.5)	204 (20.7)	180 (18.3)	125 (13.4)	96 (13.1)	72 (11.6)	41 (10.2)	38 (10.4)	39 (14.1)	57 (16.5)	791 (21.0)	852 (15.1)
Intermittent *S.aureus*; n(%)	129 (14.3)	238 (22.1)	217 (23.5)	206 (23.8)	166 (17.0)	209 (21.3)	169 (18.1)	107 (14.6)	93 (15.1)	43 (10.7)	51 (13.9)	35 (12.8)	60 (17.4)	790 (20.9)	933 (16.6)
***B. cepacia*; n (%)**	3 (0.3)	9 (0.8)	17 (1.8)	18 (2.1)	46 (4.6)	48 (4.8)	57 (6.0)	36 (4.8)	35 (5.5)	25 (6.1)	21 (5.7)	14 (5.1)	13 (3.7)	47 (1.2)	295 (5.1)
MRSA; n (%)	11 (1.2)	20 (1.82)	27 (2.9)	37 (4.2)	34 (3.4)	26 (2.6)	29 (3.0)	19 (2.5)	15 (2.4)	14 (3.4)	2 (0.5)	1 (0.4)	12 (3.4)	95 (2.5)	152 (2.7)
***H. influenza*; n (%)**	260 (28.2)	325 (29.6)	191 (20.2)	97 (11.0)	87 (8.7)	89 (9.9)	71 (7.4)	49 (6.5)	44 (6.9)	22 (5.4)	21 (5.7)	21 (7.7)	17 (4.8)	873 (22.7)	421 (7.3)

1.16 Lung infections over time

2008 n=6082 and 2015 n=9587



	Age (years)										
%	0-3	4-7	8-11	12-15	16-19	20-23	24-27	28-31	32-35	36-39	40-44
Chronic *S. aureus*; 2015	3.2	5.6	10.8	11.5	17.3	23.8	22.7	21.5	17.2	19.9	17.6
Chronic *S. aureus*; 2008	1.4	5.7	7.8	13.6	20.4	21.7	20.8	21.2	24.3	22.9	17.9
Chronic *P. aeruginosa*; 2015	2.2	2.3	6.3	15.1	29.9	41.6	52.1	55	59.2	55.9	44.7
Chronic *P. aeruginosa*; 2008	2.6	6.8	14.3	25.3	45.1	61.8	66	67.3	64.8	58.8	57.3

An analysis was conducted in order to determine whether there was a statistical difference between the proportion of people with chronic *pseudomonas aeruginosa* in 2015 compared to 2008. The results show that there is a statistically significant difference in the age bands where the p value is less than 0.05.

	Age (years)									
	0-3	4-7	8-11	12-15	16-19	20-23	24-27	28-31	32-35	36-39	40+
Chronic *P. aeruginosa*; p-value	0.772	0.000	0.000	0.000	0.000	0.000	0.000	0.000	0.156	0.546	0.000
Chronic *S. aureus*; p-value	0.063	1.000	0.068	0.251	0.132	0.342	0.426	0.956	0.026	0.446	0.999

1.17 Prevalence of complications

	Overall (n=9587)	<16 years (n=3845)	≥16 years (n=5742)
	N (%)	N (%)	N (%)
Respiratory Related			
Nasal polyps requiring surgery; n (%)	221 (2.3)	44 (1.1)	177 (3.1)
Sinus disease; n (%)	939 (9.8)	53 (1.4)	886 (15.4)
Asthma; n (%)	1382 (14.4)	497 (12.9)	885 (15.4)
ABPA; n (%)	1043 (10.9)	243 (6.3)	800 (13.9)
Haemoptysis; n (%)	762 (7.9)	38 (1.0)	724 (12.6)
Pneumothorax requiring chest tube; n (%)	59 (0.6)	<5	58 (1.0)
Nontuberculous mycobacteria or atypical mycobacteria; n (%)	536 (5.6)	113 (2.9)	423 (7.4)
Pancreas & Hepatobiliary Disease			
Raised Liver enzymes; n (%)	1116 (11.6)	264 (6.9)	852 (14.8)
Liver disease; n (%)	1371 (14.3)	340 (8.8)	1031 (18.0)
Cirrhosis with no portal hypertension; n (%)	116 (1.2)	26 (0.7)	90 (1.6)
Cirrhosis with portal hypertension; n (%)	164 (1.7)	26 (0.7)	138 (2.4)
Gall bladder disease requiring surgery; n (%)	40 (0.4)	<5	39 (0.7)
Pancreatitis; n (%)	70 (0.7)	<5	66 (1.1)
GI bleed req hosp variceal; n (%)	5 (0.1)	0 (0.0)	5 (0.1)
Upper Gastrointestinal			
GERD; n (%)	1583 (16.5)	341 (8.9)	1242 (21.6)
Peptic ulcer; n (%)	5 (0.1)	0 (0.0)	5 (0.1)
GI bleed req hosp non variceal; n (%)	11 (0.1)	<5	9 (0.2)
Lower Gastrointestinal			
Intestinal obstruction; n (%)	539 (5.6)	116 (3.0)	423 (7.4)
Fibrosing colonopathy / colonic structure; n (%)	<5	0 (0.0)	0 (0.0)
Rectal prolapse; n (%)	31 (0.3)	23 (0.6)	8 (0.1)
Renal			
Kidney stones; n (%)	96 (1.0)	12 (0.3)	84 (1.5)
Renal failure; n (%)	57 (0.6)	<5	55 (1.0)
Muscolo-Skeletal			
Arthritis; n (%)	158 (1.6)	7 (0.2)	151 (2.6)
Arthropathy; n (%)	517 (5.4)	18 (0.5)	499 (8.7)
Bone fracture; n (%)	46 (0.5)	14 (0.4)	32 (0.6)
Osteopenia; n (%)	1297 (13.5)	36 (0.9)	1261 (22.0)

Osteoporosis; n (%)	511 (5.3)	<5	507 (8.8)
Other			
Cancer confirmed by histology; n (%)	34 (0.4)	<5	32 (0.6)
Port inserted or replaced; n (%)	559 (5.8)	180 (4.7)	379 (6.6)
Depression; n (%)	452 (4.7)	<5	448 (7.8)
Hearing loss; n (%)	244 (2.5)	28 (0.7)	216 (3.8)
Hypertension; n (%)	260 (2.7)	<5	259 (4.5)
Meconium ileus; n(%)	1458 (15.2)	643 (16.7)	815 (14.2)

1.18 Incidence of complications

	2014			2015		
	Overall (n=9432)	**<16 years (n=3840)**	**≥16 (n=5592)**	**Overall (n=9587)**	**<16 years (n=3845)**	**≥16 (n=5742)**
ABPA; n (%)	143 (1.5)	67 (1.7)	76 (1.4)	99 (1.0)	38 (0.4)	61 (0.6)
Cirrhosis - no portal hypertension; n (%)	37 (0.4)	6 (0.2)	31 (0.6)	33 (0.3)	14 (0.1)	19 (0.2)
Cirrhosis - with portal hypertension; n (%)	22 (0.2)	6 (0.2)	16 (0.3)	20 (0.2)	8 (0.1)	12 (0.1)
Cancer confirmed by histology; n (%)	12 (0.1)	0 (0)	12 (0.2)	9 (0.1)	0 (0)	9 (0.1)

1.19 Nontuberculous mycobacteria (NTM) or atypical mycobacteria

	2014 n=9432	**2015 n=9587**
NTM Prevalence (%)	583 (6.2%)	536 (5.6%)
On NTM treatment in the given year (%) of total NTM prevelance	294 (50.4%)	300 (56.0%)
NTM Incidence (%)	82 (0.9%)	63 (0.7%)
***M.abscessus* complex incidence (%)**	62 (0.7%)	47 (0.5%)

1.20 CF-related diabetes

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=6970)	**10-16 years (n=1624)**	**≥16 years (n=5346)**
Treatment for CF-related diabetes; n (%)	1982 (28.0)	134 (10.0)	1848 (32.2)
Screening for CF-related diabetes			
Yes	3759 (53.1)	982 (73.1)	2777 (48.4)
No	1192 (16.8)	205 (15.3)	987 (17.2)
Known CF-related diabetes	1987 (28.0)	109 (8.1)	1878 (32.7)
Unknown	99 (1.4)	30 (2.2)	69 (1.2)

Therapies

1.21 Transplants

Lung transplantation has been available to people with cystic fibrosis for almost 30 years. Today the most common operation carried out is a double lung transplant, called a Bilateral Sequential Lung Transplant. The following table gives information about transplant activity over time.

	2011	2012	2013	2014	2015
Number of patients that year with annual review data evaluated for transplants	204	225	220	247	229
Number accepted on the transplant list	121	120	136	146	125
Number receiving transplants (<16)	<5	<5	<5	5	<5
Types of transplants received:					
Bilateral lung	<5	<5	<5	<5	<5
Heart and lung	0	0	0	0	0
Liver	0	<5	<5	<5	<5
Other	0	0	0	0	<5
Number receiving transplants (≥16)	48*	52**	54*	67**	46
Types of transplants received:					
Bilateral lung	40	43	48	59	42
Heart and lung	<5	<5	0	0	0
Liver	<5	6	<5	5	<5
Other	<5	<5	<5	5	<5

* One patient received two transplants
** Two patients had two transplants

1.22 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. In July 2015 NHS England commissioned ivacaftor for the treatment of CF in patients aged 6 years and over with at least one copy of gating mutation G187R, S549N, S549R, G551S, G1244E, S1251N, S1255P, or G139D. The table shows information about ivacaftor use and outcomes from June 2012 – December 2015.

Number of patients on ivacaftor in the UK	**439**
	Median (IQR)
Sweat chloride before ivacaftor	105 (95.6-113)
Sweat chloride 6-8 weeks after ivacaftor	49 (37-64)
FEV_1% before ivacaftor	55.4 (40.6-69.8)
FEV_1% 6-8 weeks after ivacaftor	64.1 (48.5-76.0)
Number of patients stopped ivacaftor	7

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.

1.23 Intravenous (IV) antibiotic use and outcomes
n=9587

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	921	47 (5.1)	8 (6-13)	257 (27.9)	14 (8-20)	258 (28.0)	14 (10-21)
4-7	1098	114 (10.4)	12 (7-21)	295 (26.9)	14 (9-19)	306 (27.9)	14 (14-28)
8-11	946	177 (18.7)	14 (10-28)	335 (35.4)	14 (8-28)	363 (38.4)	22 (14-43)
12-15	880	203 (23.1)	21 (10-24)	374 (42.5)	18 (11-35)	416 (47.3)	28 (14-49)
16-19	999	279 (27.9)	19 (12-35)	437 (43.7)	15 (8-40)	513 (51.4)	28 (14-56)
20-23	1000	332 (33.2)	21 (13.3-35)	448 (44.8)	16 (10-38)	547 (54.7)	28 (14-56)
24-27	954	364 (38.2)	21 (14-39)	444 (46.5)	17 (10-40)	561 (58.8)	28 (14-58)
28-31	749	284 (37.9)	22 (14-39)	313 (41.8)	18 (10-42)	430 (57.4)	28 (14-56)
32-35	634	229 (36.1)	26 (14-47)	245 (38.6)	17 (9-37)	330 (52.1)	31 (15-56)
36-39	407	147 (36.1)	24 (14-40)	147 (36.1)	16 (10-31)	208 (51.1)	28 (14-51)
40-44	370	105 (28.4)	26 (14-42)	119 (32.2)	15 (8-36)	163 (44.1)	28 (14-55)
45-49	277	74 (26.7)	23 (14-37)	87 (31.4)	18 (11-41)	111 (40.1)	35 (14-60)
50+	352	74 (21.0)	15 (11-42)	113 (32.1)	18 (10-34)	138 (39.2)	28 (14-47)
Overall	**9587**	**2429 (25.3)**	**19 (12-37)**	**3614 (37.7)**	**15 (9-33)**	**4344 (45.3)**	**28 (14-49)**

Nebulised drug treatments

Nebulised drugs are medications that are breathed in as a mist. They are changed into a mist by a pot holding liquid medication, called a nebuliser.

Nebulised medications are used because:

- The medications go straight to where they need to work (in the lung) without having to go round the body. This can reduce side-effects.
- Some medication is only available as a nebulised medication, for example DNase.
- Large doses of medication can be given compared with some types of inhaler.

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

	2008			2015		
	Overall	<16 years	≥16 years	Overall	<16 years	≥16 years
Patients with chronic *P. aeruginosa*	2098	299	1799	2859	234	2625
Tobramycin solution; n (%)	412 (19.6)	48 (16.1)	364 (20.2)	653 (22.8)	86 (36.8)	567 (21.6)
Other aminoglycoside; n (%)	43 (2.0)	5 (0.2)	38 (2.1)	95 (3.3)	15 (6.4)	80 (3.1)
Colistin; n (%)	914 (43.6)	174 (58.2)	740 (41.1)	840 (29.4)	108 (46.2)	732 (28.0)
Promixin; n (%)	490 (23.4)	73 (24.4)	417 (23.2	896 (31.3)	111 (47.4)	785 (29.9)
Aztreonam; n (%)	-	-	-	485 (17.0)	8 (3.4)	477 (18.2)
Colistimethate (DPI); n (%)	-	-	-	537 (18.8)	22 (9.4)	515 (19.6)
Tobramycin Inhalation Powder; n (%)	-	-	-	888 (31.1)	25 (10.7)	863 (32.9)
At least one of the above*; n (%)	1597 (76.1)	257 (86.0)	1340 (74.5)	2547 (89.1)	226 (96.6)	2321 (88.4)

*In 2015, 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.

Muco-active therapies

1.25 Mannitol

	2015	
Age	**Total patients**	**Patients on Mannitol**
0-3	921	<5
4-7	1098	<5
8-11	946	<5
12-15	880	<5
16-19	999	24 (2.4)
20-23	1000	57 (5.7)
24-27	954	82 (8.6)
28-31	749	49 (6.5)
32-35	634	42 (6.6)
36-39	407	25 (6.1)
40+	999	46 (4.6)
Overall	**9587**	**330 (3.4)**

1.26 DNase

	DNase; n (%)			
	2008		2015	
Age	Total patients	Patients on DNase	Total patients	Patients on DNase
0-3	605	46 (7.6)	921	121 (13.1)
4-7	621	125 (20.1)	1098	478 (43.5)
8-11	663	227 (34.2)	946	665 (70.3)
12-15	773	359 (46.4)	880	654 (74.3)
16-19	762	377 (49.5)	999	733 (73.4)
20-23	725	319 (44.0)	1000	654 (65.4)
24-27	605	288 (47.6)	954	609 (63.8)
28-31	419	182 (43.4)	749	470 (62.8)
32-35	260	108 (41.5)	634	376 (59.3)
36-39	237	83 (35.0)	407	225 (55.3)
40+	412	147 (35.7)	999	182 (51.7)
Overall	**6082**	**2261 (37.2)**	**9587**	**5495 (57.3)**

1.27 Hypertonic saline

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

	Hypertonic saline; n (%)			
	2008		2014	
Age	Number of patients	Patients on hypertonic saline	Number of patients	Patients on hypertonic saline
0-3	605	3 (0.5)	921	55 (6.0)
4-7	621	15 (2.4)	1098	198 (18.0)
8-11	663	23 (3.5)	946	282 (29.8)
12-15	773	32 (4.1)	880	359 (40.8)
16-19	762	33 (4.3)	999	354 (35.4)
20-23	725	50 (6.9)	1000	290 (29.0)
24-27	605	60 (9.9)	954	277 (29.0)
28-31	419	37 (8.8)	749	246 (32.8)
32-35	260	29 (11.2)	634	197 (31.1)
36-39	237	16 (6.8)	407	122 (30.0)
40+	204	33 (8.0)	999	248 (24.8)
Overall	**6082**	**331 (5.4)**	**9587**	**2628 (27.4)**

1.28 Long-term azithromycin use in patients with and without chronic *Pseudomonas aeruginosa*

Azithromycin is an antiobiotic with anti-inflammatory properties used to treat certain infections, including *Pseudomonas aeruginosa.*

	2008				2015			
	Overall (n=1958)	**0-3 years (n=15)**	**4-15 years (n=363)**	**≥16 years (n=1580)**	**Overall (n=3719)**	**0-3 years (n=26)**	**4-15 years (n=590)**	**≥16 years (n=3103)**
Patients with chronic *P. aeruginosa*	1246 (63.6)	<5	105 (28.9)	1139 (72.1)	1874 (50.4)	<5	94 (15.9)	1776 (57.2)
Patients without chronic *P. aeruginosa*	712 (36.4)	13 (86.7)	258 (71.1)	441 (27.9)	1845 (49.6)	22 (84.6)	496 (84.1)	1327 (42.8)

1.29 Physiotherapy

Physiotherapy helps people with cystic fibrosis clear sticky mucus from their lungs.

	Overall (n=9587)	**<16 years (n=3845)**	**≥16 years (n=5742)**
Active cycle of breathing techniques; n (%)	2708 (28.7)	1534 (40.3)	1174 (20.8)
Autogenic drainage (including assited autogenic drainage); n (%)	1431 (15.1)	196 (5.2)	1235 (21.9)
Any form of PEP; n (%)	5282 (55.9)	2706 (71.1)	2576 (45.6)
VEST; n (%)	174 (1.8)	92 (2.4)	82 (1.5)
Exercise; n (%)	3552 (37.1)	1541 (40.1)	2011 (35.0)

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

1.30 Other therapy

	Overall (n=9587)	<16 years (n=3845)	≥16 years (n=5742)
Non Invasive Ventillation (NIV); n (%)	303 (3.2)	45 (1.2)	258 (4.5)
Long-term oxygen; n (%) Among those who have long-term oxygen:	604 (6.3)	93 (2.4)	511 (8.9)
Continuously	162 (1.7)	5 (0.1)	157 (2.7)
Nocturnal or with exertion	163 (1.7)	24 (0.6)	139 (2.4)
As required (PRN)	51 (0.5)	<5	47 (0.8)
With exacerbation	228 (2.4)	60 (1.6)	168 (2.9)

1.31 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.

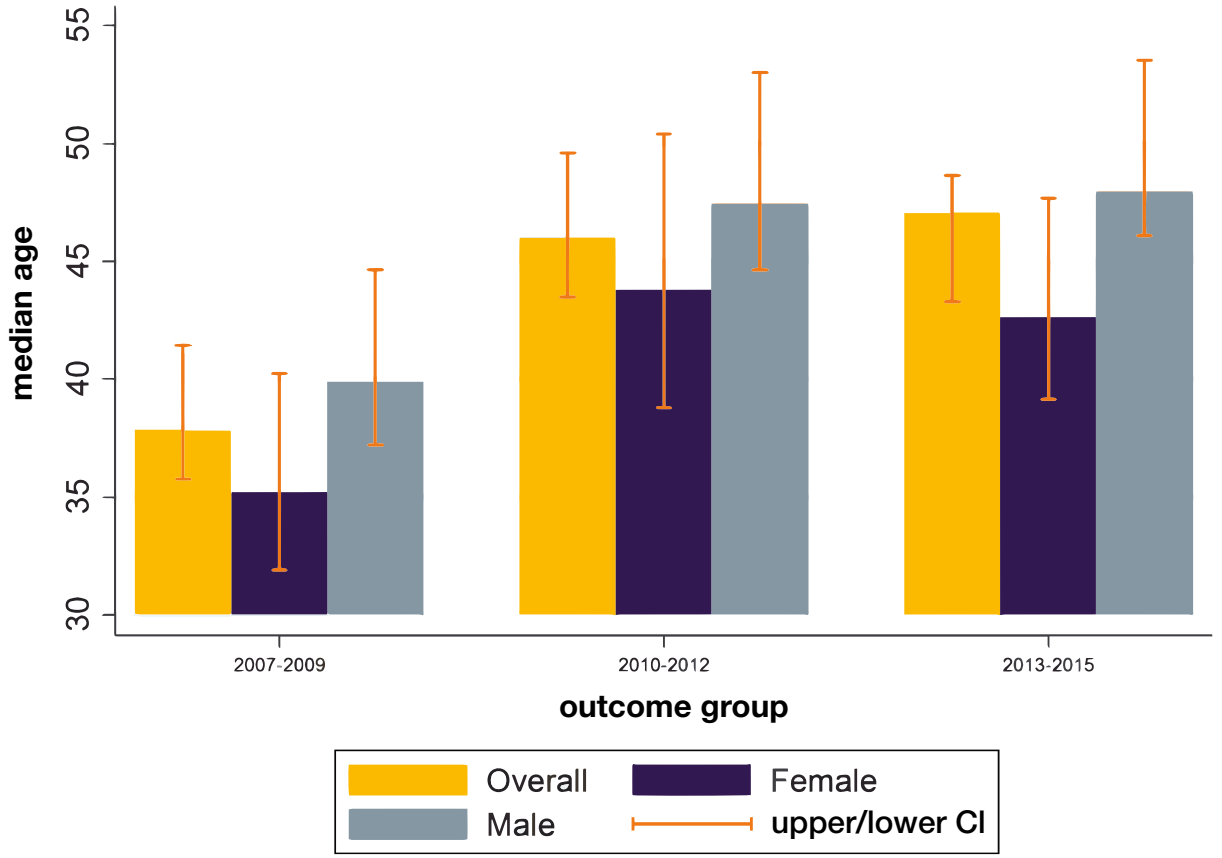
	Overall (n=9587)	<16 years (n=3845)	≥16 years (5742)
Any supplmental feeding; n (%)	3126 (32.6)	1120 (29.1)	2006 (34.9)
Nasogastric tube	109 (1.1)	15 (0.4)	94 (1.6)
Gastrostomy tube/Button	557 (5.8)	220 (5.7)	337 (5.9)
Jejunal	7 (0.1)	<5	<5
Total Parenteral Nutrition (TPN)	<5	<5	<5

1.32 Survival

Median predicted survival is a calculation based on people with CF recorded in the Registry as alive in the given year. A mathematical formula[7], which takes into account the age of those people in 2015, predicts how long we expect half of them to live. For 2015, this means that half of people registered as alive on the database are predicted to live to at least 45.1 years of age. Half of people alive today are currently predicted to die before they reach that age.

Using one year of data can show big variations in median predicted survival age each year, which can be due to chance alone and does not necessarily reflect a decline or improvement in real-world outcomes. Grouping several years together gives a better estimate of survival.

An analysis of median predicted survival grouped together in three year windows is shown below.



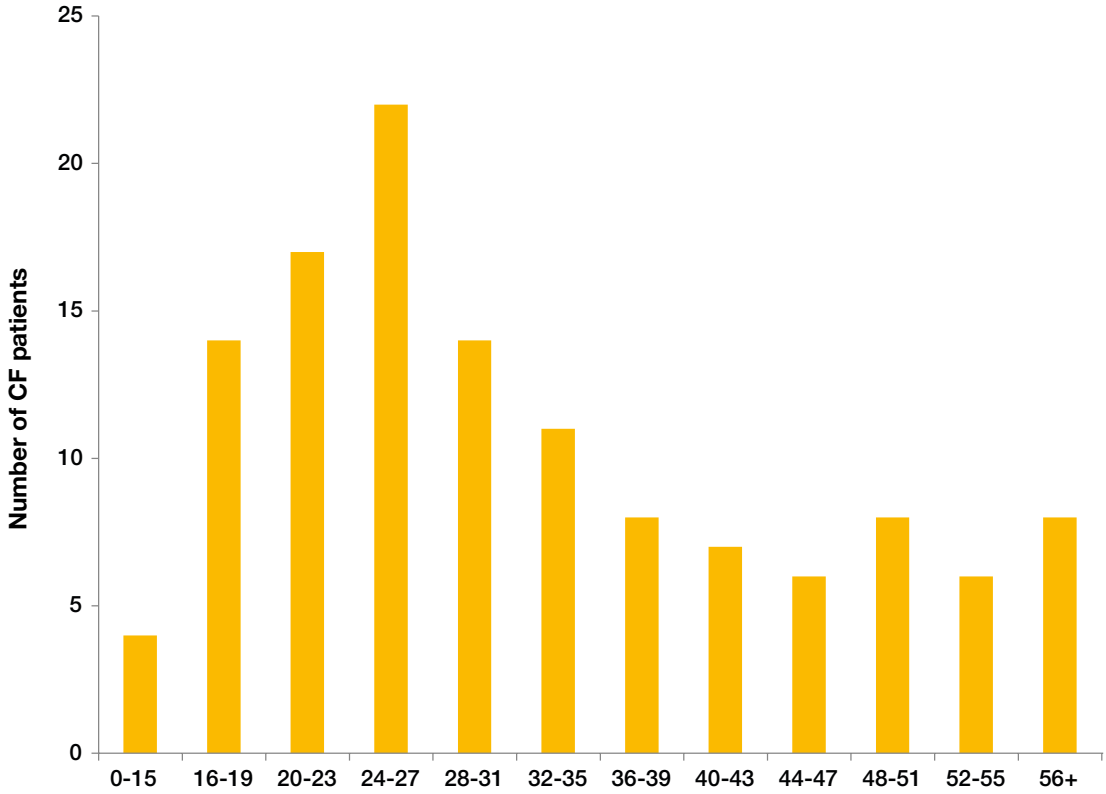
	Overall			Males			Females			Male and Female
Year	**Median age**	**Lower 95% CI**	**Upper 95% CI**	**Median age**	**Lower 95% CI**	**Upper 95% CI**	**Median age**	**Lower 95% CI**	**Upper 95% CI**	**p-values comparing survival**
2007-2009	37.87	35.76	41.43	39.88	37.22	44.65	35.18	31.92	40.24	0.0003
2010-2012	46.01	43.5	49.59	47.44	44.66	53.01	43.78	38.79	50.41	0.0143
2013-2015	47.06	43.29	48.64	47.96	46.1	53.52	42.62	39.16	47.68	0.0264

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

1.33 Age distribution of deaths in 2015

The table below shows the ages of the 125 people with cystic fibrosis who died in 2015. To protect the identities of individuals, where a number of deaths in an age group is less than five '<5' is shown, rather than the actual number.

Age at death	Number of cystic fibrosis patients
0-3	<5
4-7	<5
8-11	<5
12-15	<5
16-19	14
20-23	17
24-27	22
28-31	14
32-35	11
36-39	8
40-43	7
44-47	6
48-51	8
52-55	6
56+	8
Total	**125**



Median age of death is based on the people with CF who died in any given year. So in 2015 the median age of the 125 people who died was 28.

Genotypes

Genotypes are part of the genetic makeup of a cell, organism or individual that usually control a particular characteristic (known as a phenotype). For people with CF, their genotype reveals which mutations of the CF gene cause their CF. 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.

9401 (98.1%) patients have been genotyped with a recorded value.

DF508 Mutations; n (%)

Homozygous DF508 4728 (50.29%)

Heterozygous DF508 3779 (40.20%)

1.34 Genotypes

Mutations				
Nucleotide	**Protein**	**Legacy name**	**N**	**(%)**
c.1521_1523delCTT	p.Phe508del	^F508	8,507	90.49
Other	Other	Other	1648	17.53
c.1652G>A	p.Gly551Asp	G551D	533	5.67
Not identified	Not identified	Not identified	500	5.32
c.350G>A	p.Arg117His	R117H	433	4.61
c.1624G>T	p.Gly542X	G542X	324	3.45
c.489+1G>T	No protein name	621+1G->T	208	2.21
c.3909C>G	p.Asn1303Lys	N1303K	133	1.41
c.1585-1G>A	No protein name	1717-1G->A	117	1.24
c.1766+1G>A	No protein name	1898+1G->A	110	1.17
c.1519_1521delATC	p.Ile507del	^I507	98	1.04
c.3528delC	p.Arg560Thr	R560T	88	0.94
c.1679G>C	p.Lys1177SerfsX15	3659delC	85	0.90
c.1657C>T	p.Arg553X	R553X	77	0.82
c.3717+12191C>T	No protein name	3849+10kbC->T	76	0.81
c.254G>A	p.Asp1152His	D1152H	75	0.80
c.1477C>T	p.Gly85Glu	G85E	71	0.76
c.3454G>C	p.Gln493X	Q493X	69	0.73
c.178G>T	p.Glu60X	E60X	56	0.60
c.3846G>A	p.Trp1282X	W1282X	49	0.52
c.2052delA	p.Lys684AsnfsX38	2184delA	35	0.37
c.2657+5G>A	p.Arg347Pro	R347P	34	0.36
c.1040G>C	No protein name	2789+5G->A	32	0.34
c.1646G>A	p.Ser549Asn	S549N	29	0.31
c.1558G>T	p.Val520Phe	V520F	28	0.30
c.3484C>T	p.Arg1162X	R1162X	27	0.29
c.1364C>A	p.Ala455Glu	A455E	26	0.28
c.1000C>T	p.Arg334Trp	R334W	16	0.17

Mutations				
Nucleotide	**Protein**	**Legacy name**	**N**	**(%)**
c.1040G>A	p.Arg347His	R347H	16	0.17
c.1055G>A	p.Arg352Gln	R352Q	14	0.15
c.3472C>T	p.Arg1158X	R1158X	12	0.13
c.2988+1G>A	No protein name	3120+1G->A	11	0.12
c.532G>A	p.Gly178Arg	G178R	9	0.10
c.579+1G>T	No protein name	711+1G->T	8	0.09
-	-	R1283M	7	0.07
c.1645A>C or c.1647T>G	p.Ser549Arg	S549R	7	0.07
c.2051_2052delAAinsG	p.Lys684SerfsX38	2183delAA->G	6	0.06
c.443T>C	p.Ile148Thr	I148T	5	0.05
c.2012delT	p.Leu671X	2143delT	5	0.05
c.1675G>A	p.Ala559Thr	A559T	5	0.05
-	-	C524X	<5	-
c.2128A>T	p.Lys710X	K710X	<5	-
c.1654C>T	p.Gln552X	Q552X	<5	-

1.34a Most common genotypes by devolved nation*

	England		Scotland		Wales		Northern Ireland	
Legacy name	**N=7845**	**(%)**	**N=784**	**(%)**	**N=372**	**(%)**	**N=400**	**(%)**
^F508	7,119	91%	714	91%	334	90%	340	85%
G542X	225	3%	53	7%	24	6%	26	7%
G551D	407	5%	81	10%	13	3%	32	8%
R117H	310	4%	56	7%	13	3%	54	14%
1898+1G->A	82	1%	<5	-	26	7%	0	0%
621+1G->T	153	2%	10	1%	37	10%	8	2%

*Please note that this table was updated in August 2017.

Section 2 and 3: Centre-level analysis

Cystic fibrosis care in the UK is led by 52 regional centres, eight stand-alone clinics and 77 networked clinics. The breakdown between centres and clinics delivering paediatric and adult care is shown below:

	Paediatric	Adult	Total
Regional centres	28	24	52
Stand-alone clinics	4	4	8
Networked clinics	70	7	77

Section 2 and 3 show analysis of data for individual CF centres. This allows people with CF, their families and healthcare providers, to compare centres against one another, and the national average. This level of transparency helps to improve standards of care by giving people with CF and healthcare providers alike the chance to make informed choices about what questions to ask of their team, and which types of treatment may be best for each individual.

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.

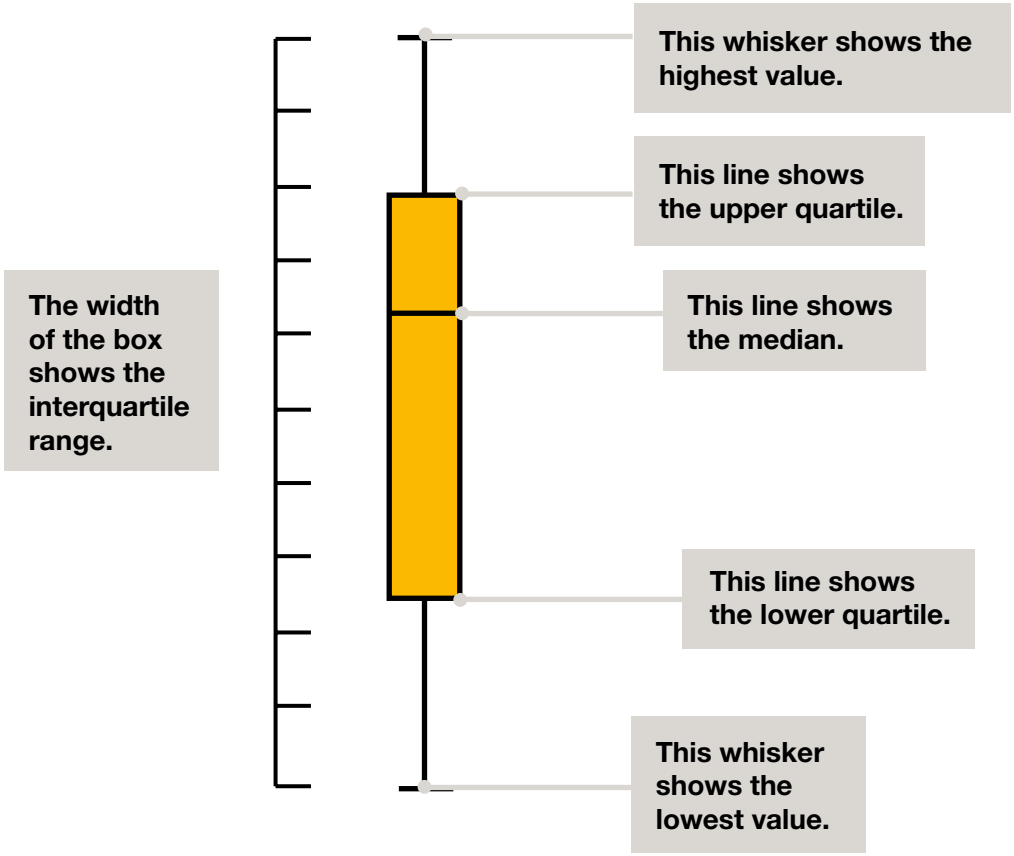
If a person with CF or a member of their family has questions about the results for their CF centre or clinic, they should discuss this with their CF team.

Full tables of the data are shown in appendix 1 on page 54.

A guide to the charts

Some of the data in this section are shown as ‘box plots’. This year, for the first time, we are also showing the data in ‘funnel plots’. We are showing the same data in two different formats so that people can view the information in a way that is most meaningful to them.

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

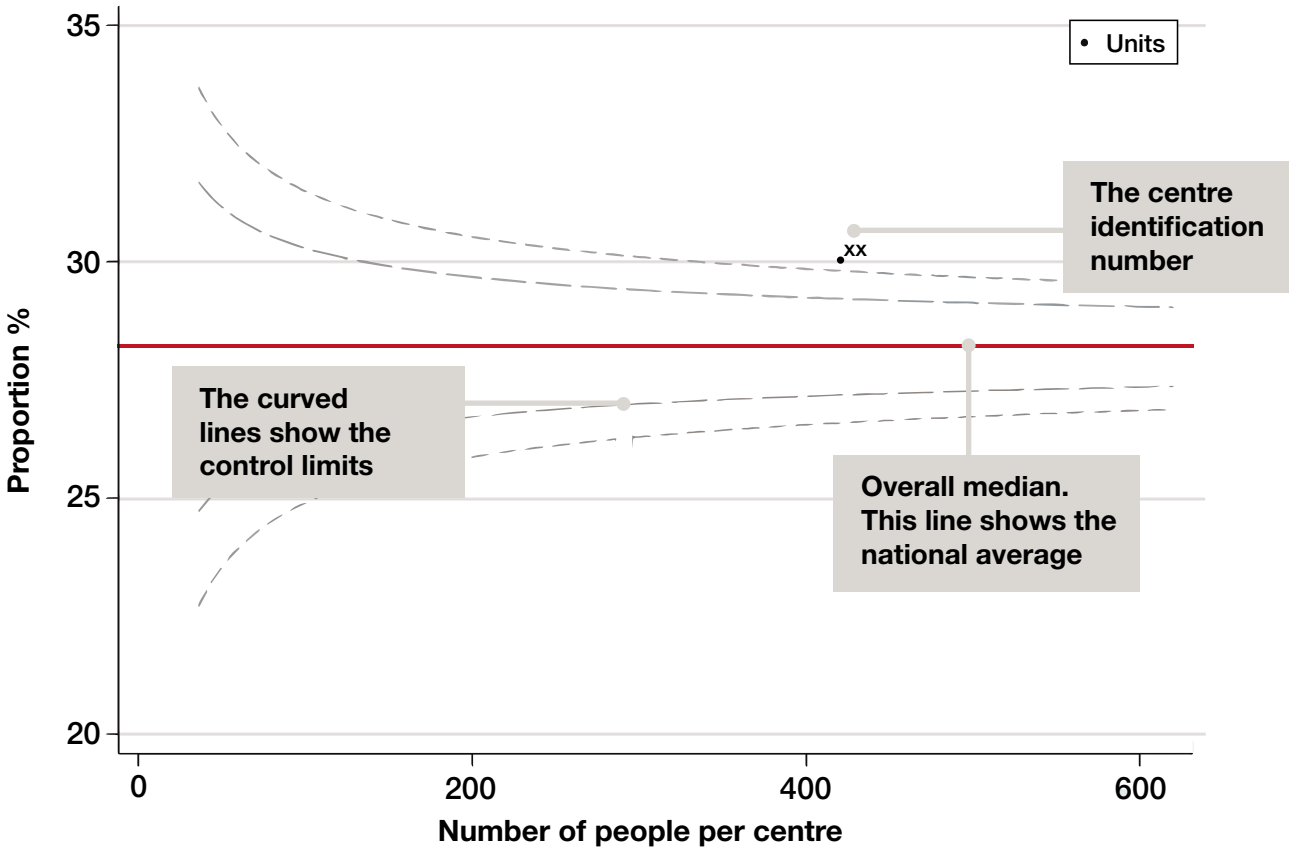
The more people with CF at a care site, the closer to the national average you would expect the results to be. This is because high numbers in one centre affect the overall average across the country, 'pulling' the average towards them. When a small number of people with CF are treated at a site, even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

If the result for a CF centre is between the '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 limit 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.

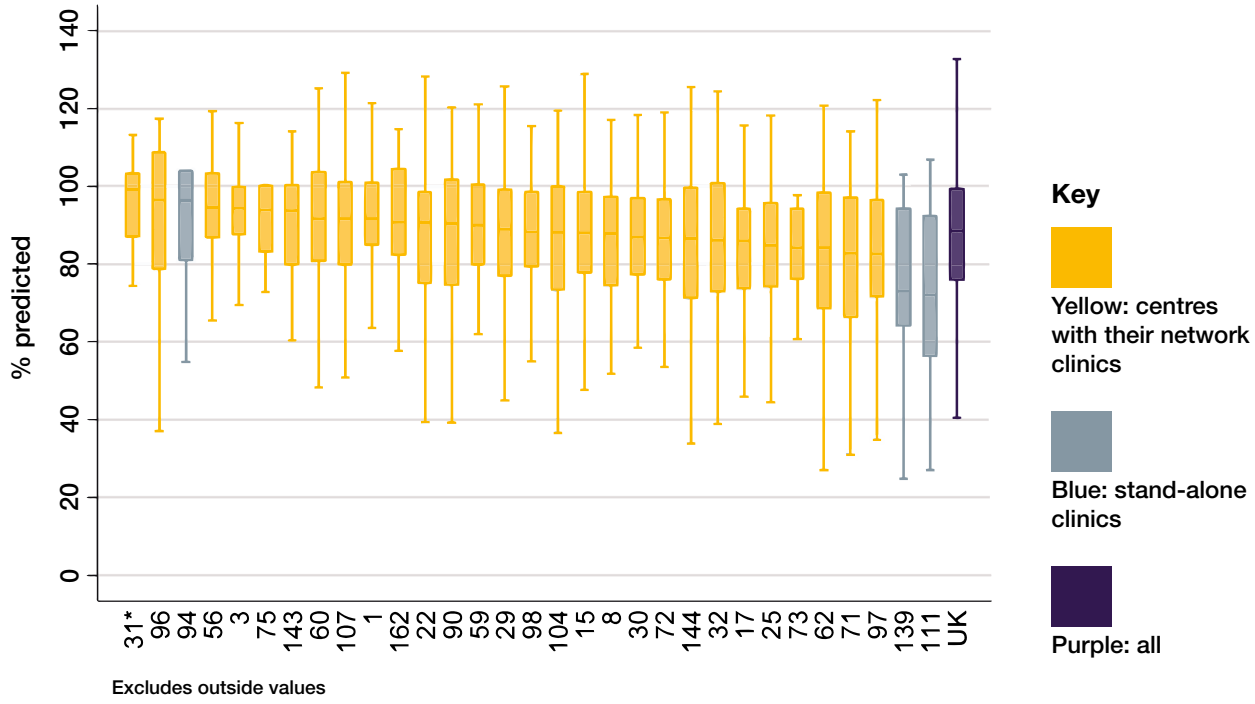


Section 2 Paediatric centre analysis

n=4188

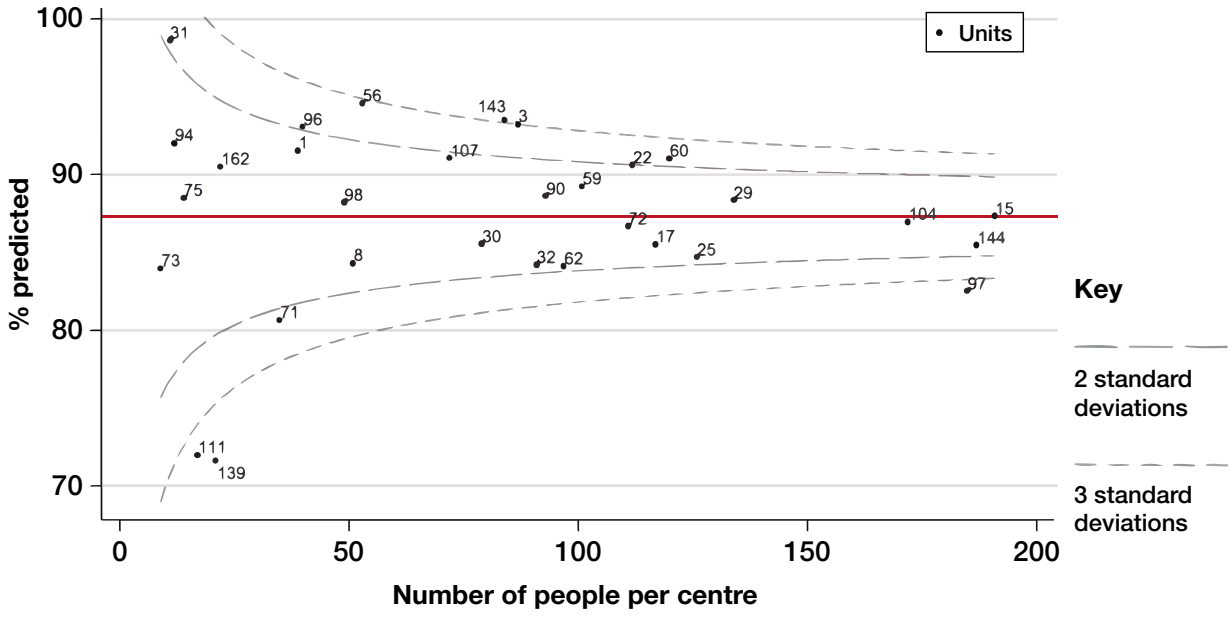
In the UK, paediatric CF care is led by 28 specialist CF centres and four stand-alone clinics. Some paediatric centres oversee care delivered by 70 smaller, networked clinics.

2.1 Median FEV_1 % predicted among patients aged 6 and over by paediatric centre/clinic (without a history of lung transplant) (GLI equations)

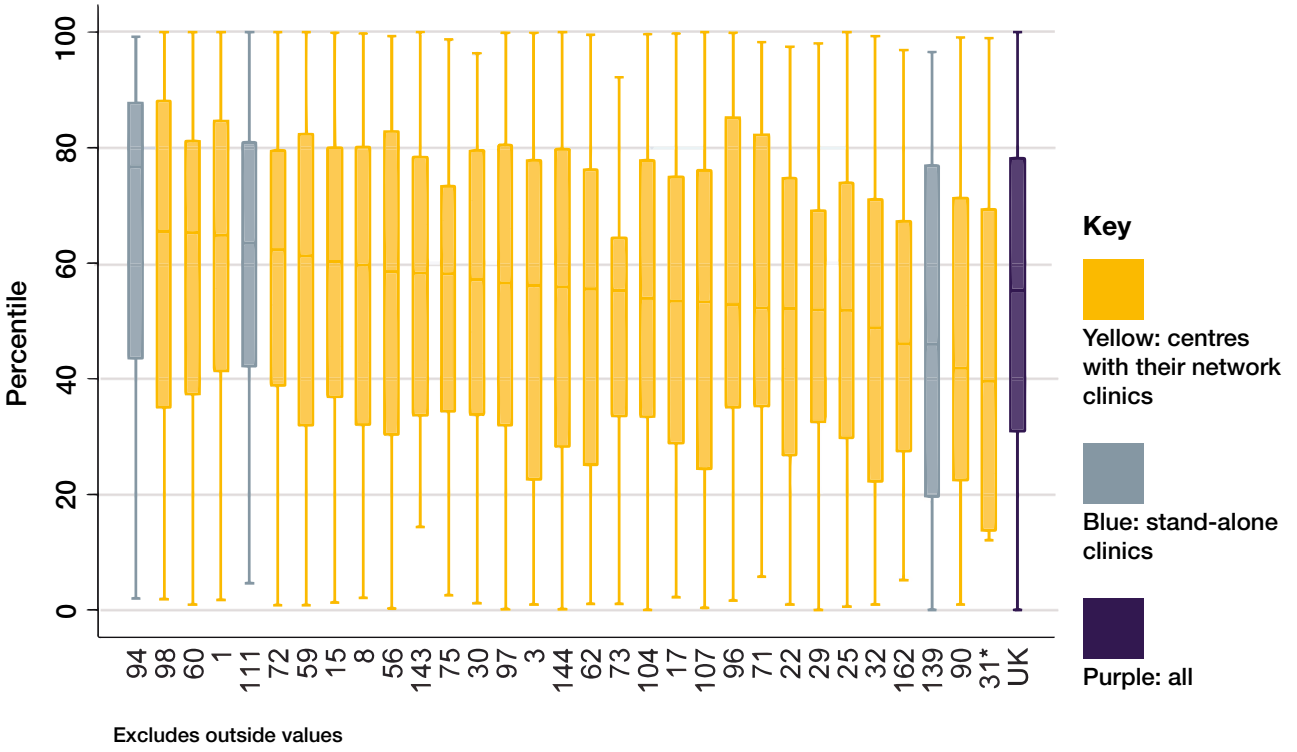


The median FEV_1 % predicted for patients attending paediatric centres/clinics is 86% predicted (IQR: 76–99).

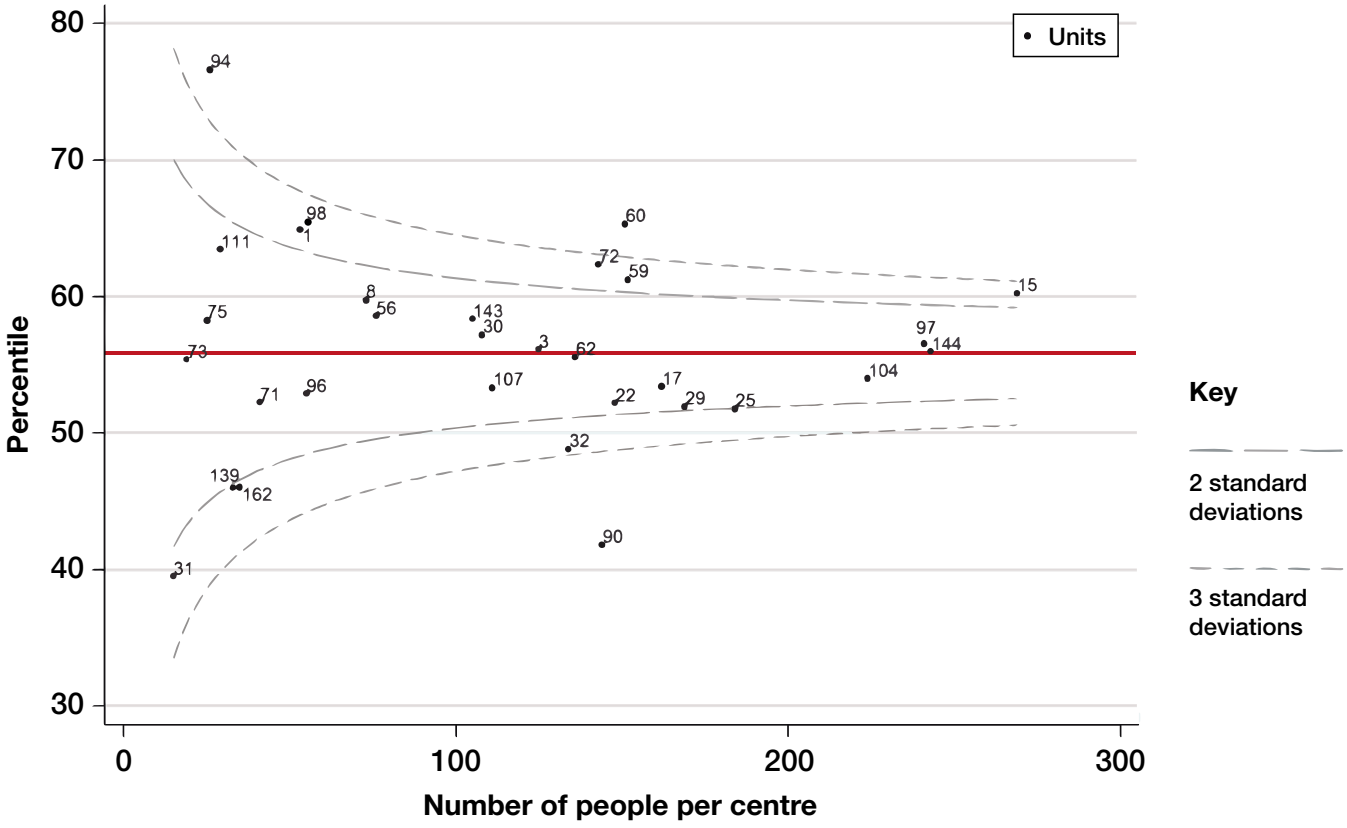
*Centre/clinic with a data set submission of fewer than 20 patients.



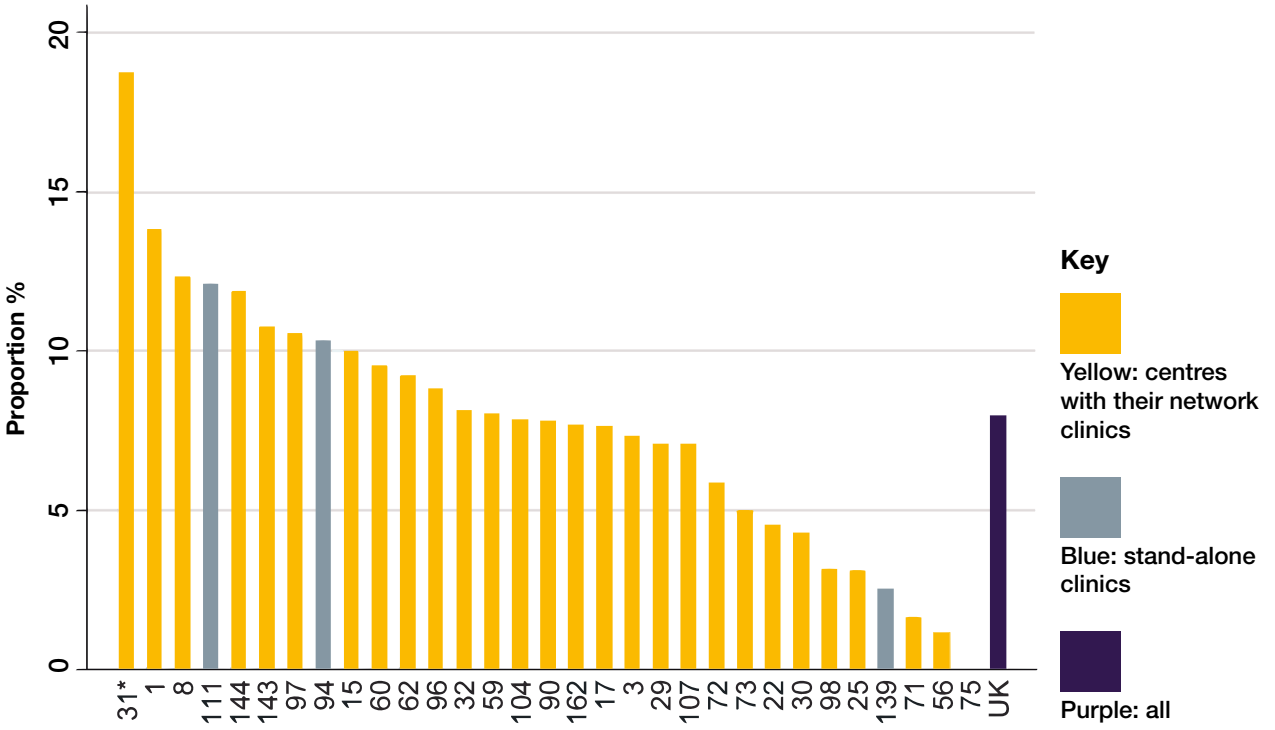
2.2 Median BMI percentile among patients aged 2 to 15 years by paediatric centre/clinic



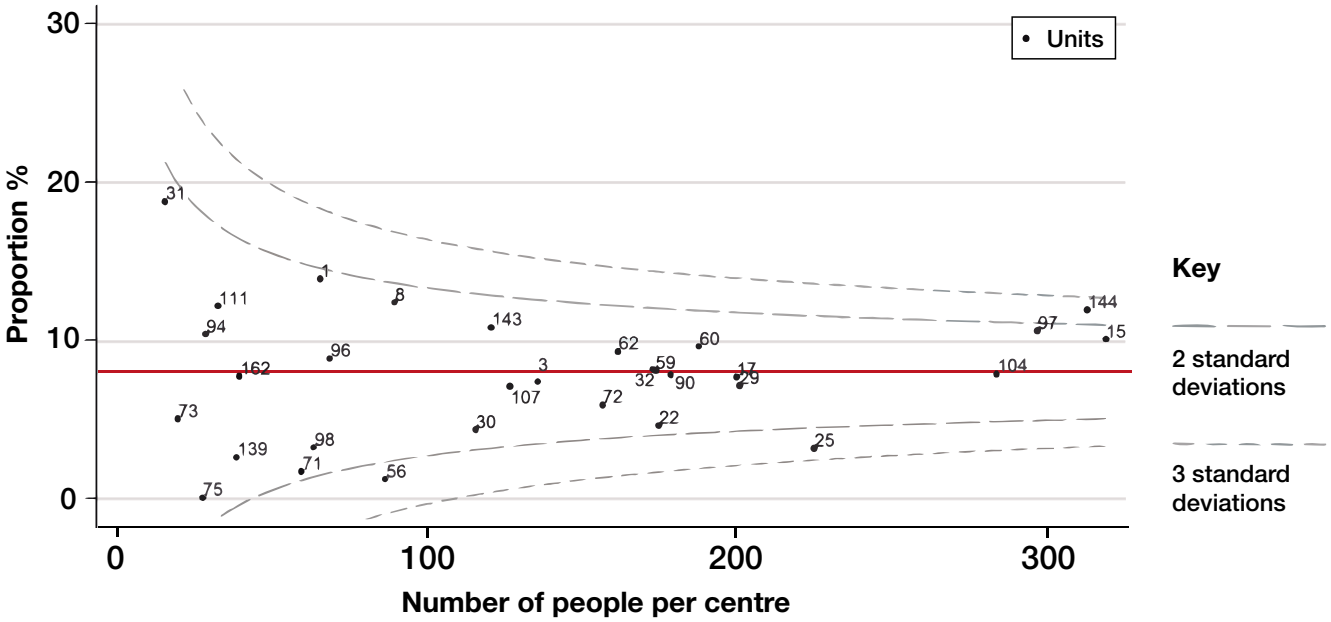
The median BMI percentile for patients attending paediatric centres/clinics is 55 (IQR: 31–78).
* Centre/clinic with a data set submission of fewer than 20 patients.



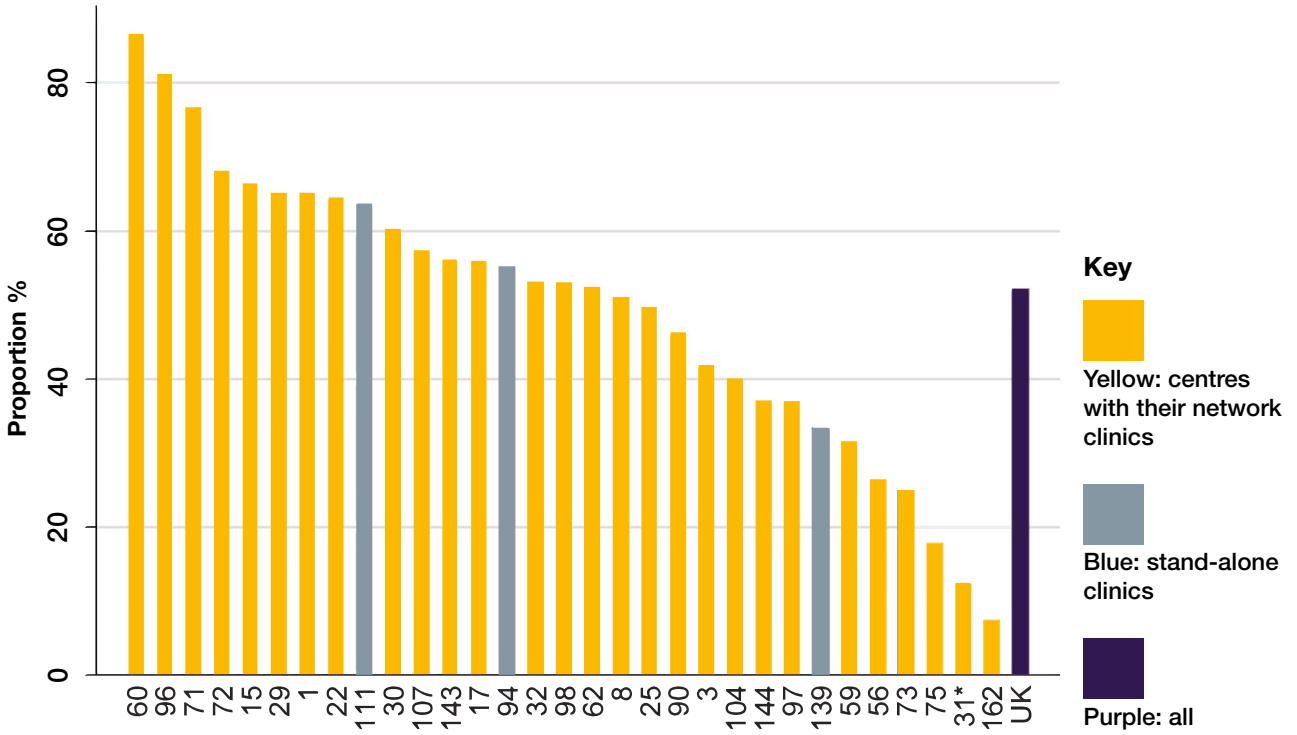
2.3 Proportion of patients with chronic *Pseudomonas aeruginosa* by paediatric centre/clinic



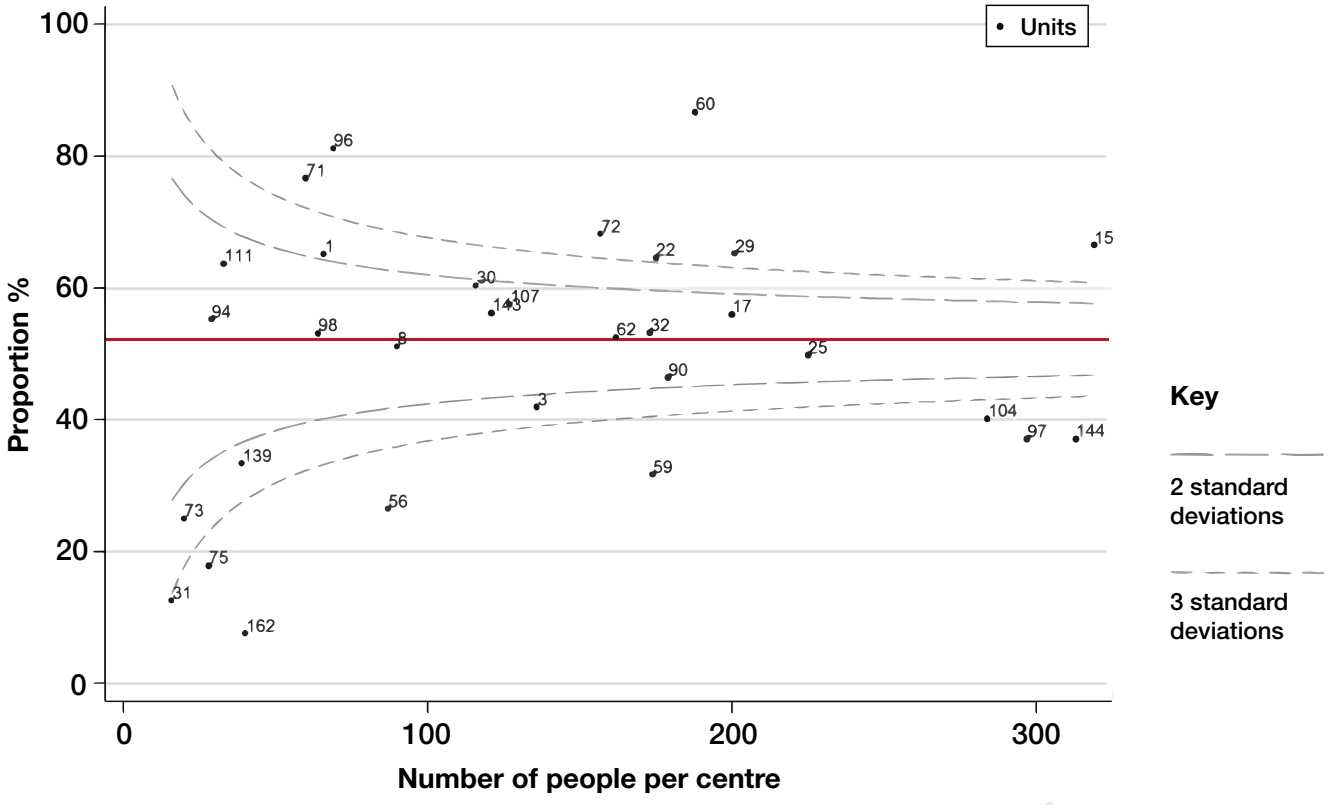
The proportion of patients with chronic *Pseudomonas aeruginosa* in paediatric centres/clinics is 8%.
* Centre/clinic with a data set submission of fewer than 20 patients.



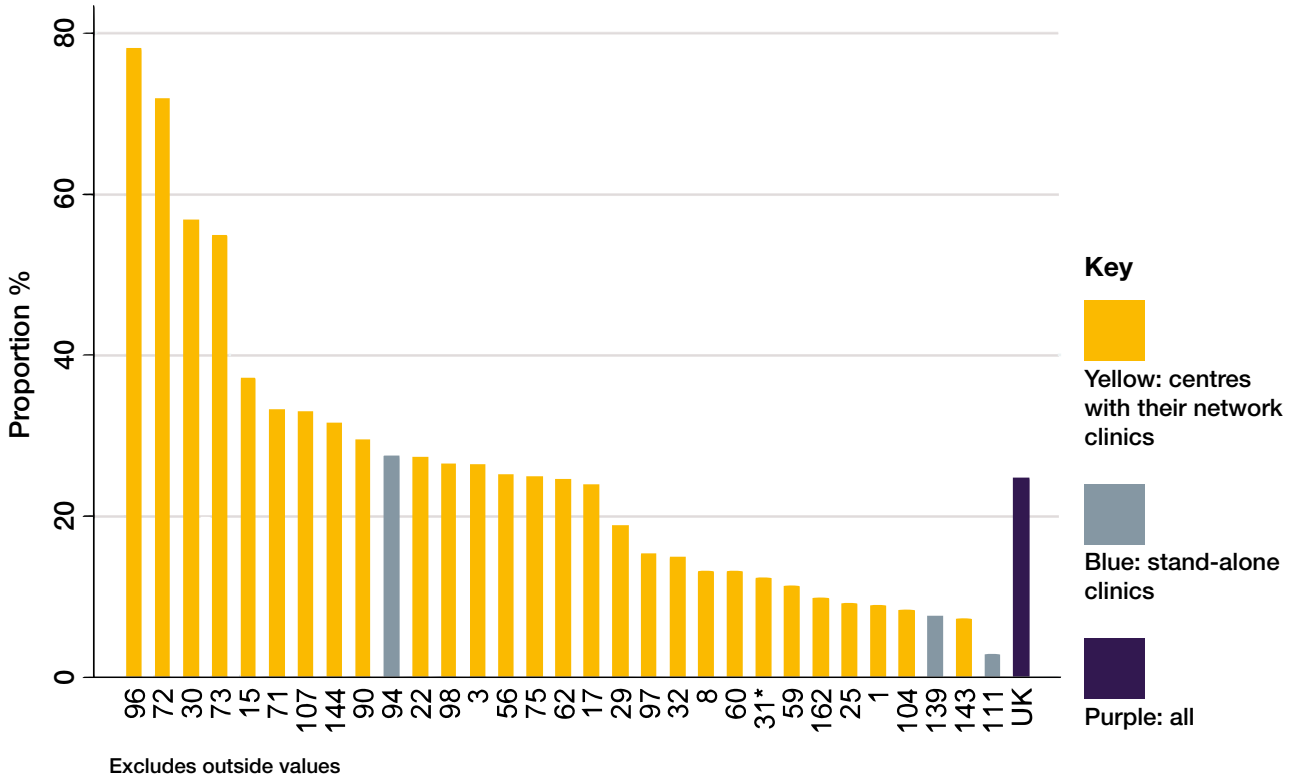
2.4 Proportion of patients receiving DNase treatment by paediatric centre/clinic



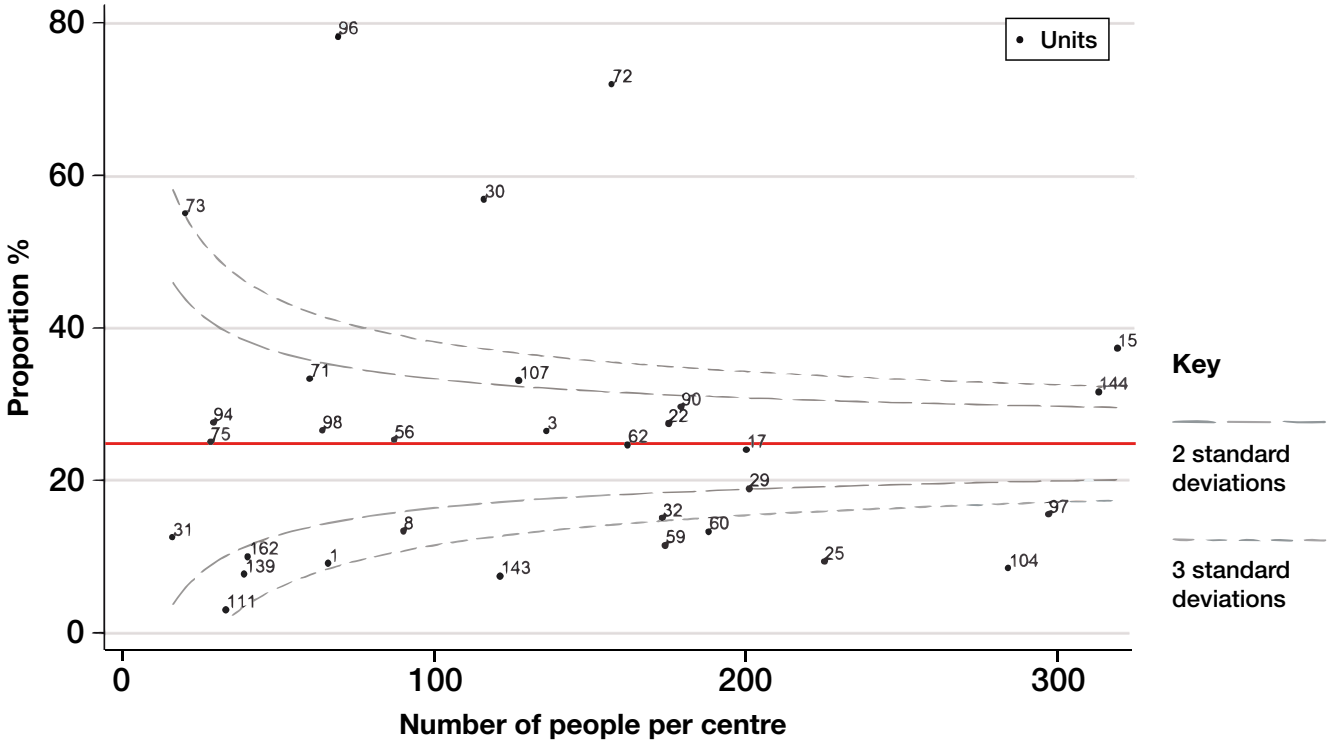
The proportion of patients receiving DNase treatment in paediatric centres/clinics is 52%.
* Centre/clinic with a data set submission of fewer than 20 patients.



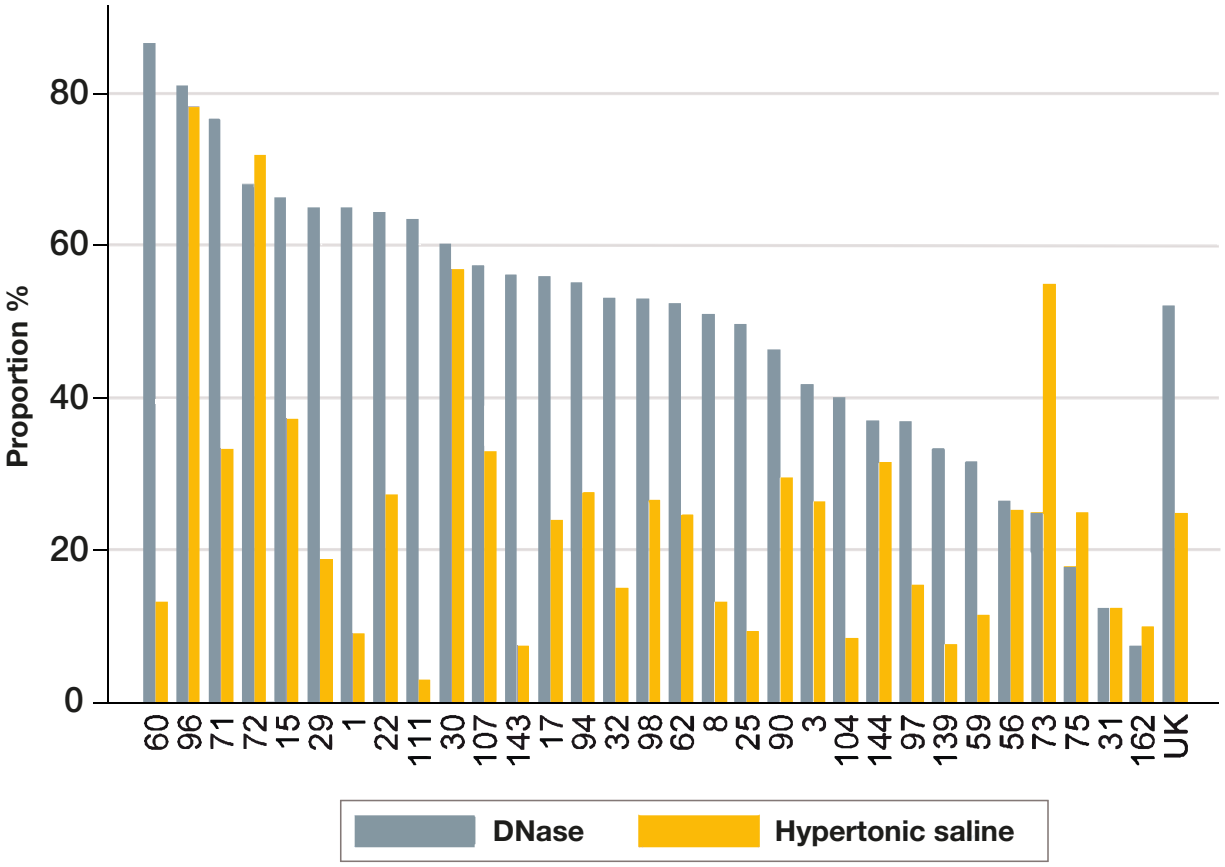
2.5 Proportion of patients receiving hypertonic saline treatment by paediatric centre/clinic



The proportion of patients receiving hypertonic saline treatment in paediatric centres/clinics is 25%.
* Centre/clinic with a data set submission of fewer than 20 patients.



2.6 DNase and hypertonic saline use by paediatric centre/clinic

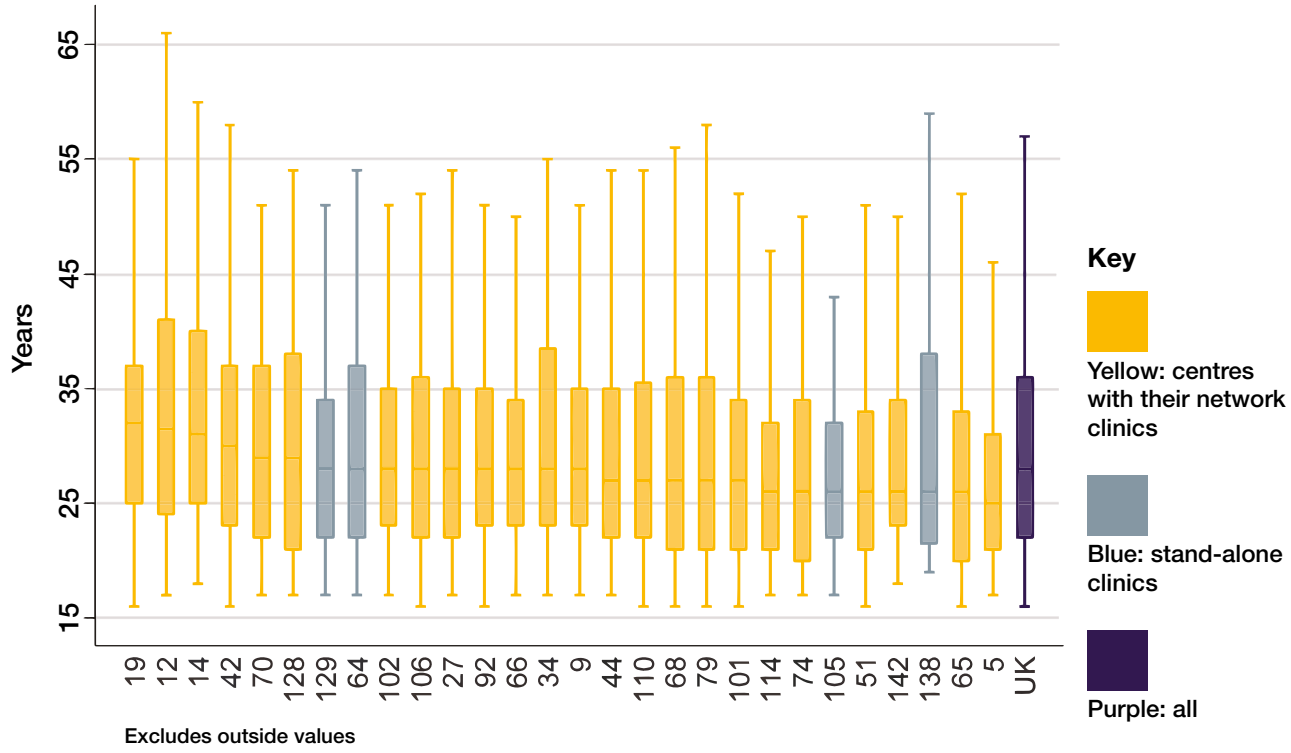


Section 3: Adult centre analyses

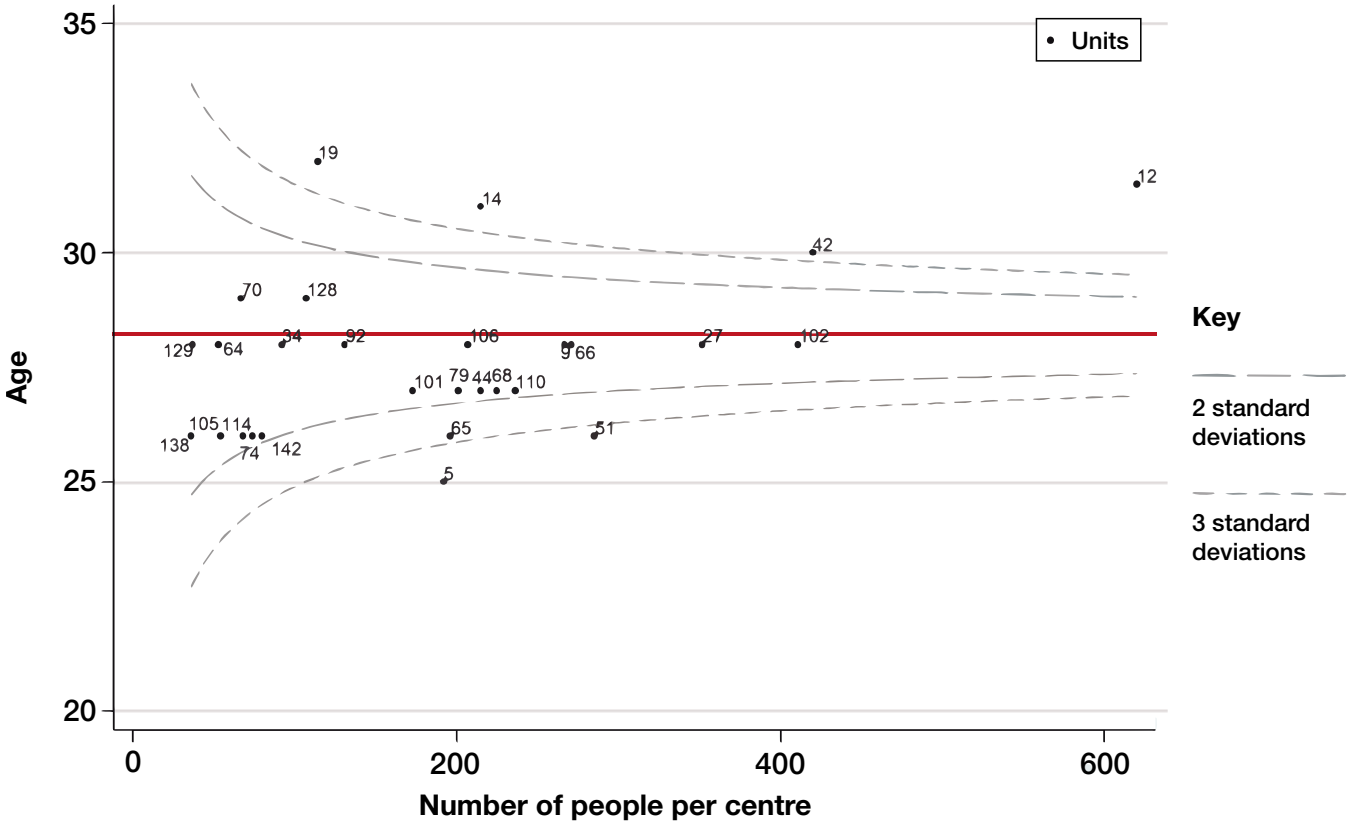
n=5399

In the UK cystic fibrosis care is led by 24 adult specialist CF centres and four stand-alone clinics. People with CF transfer to adult care centres between the ages of 16 and 18 years.

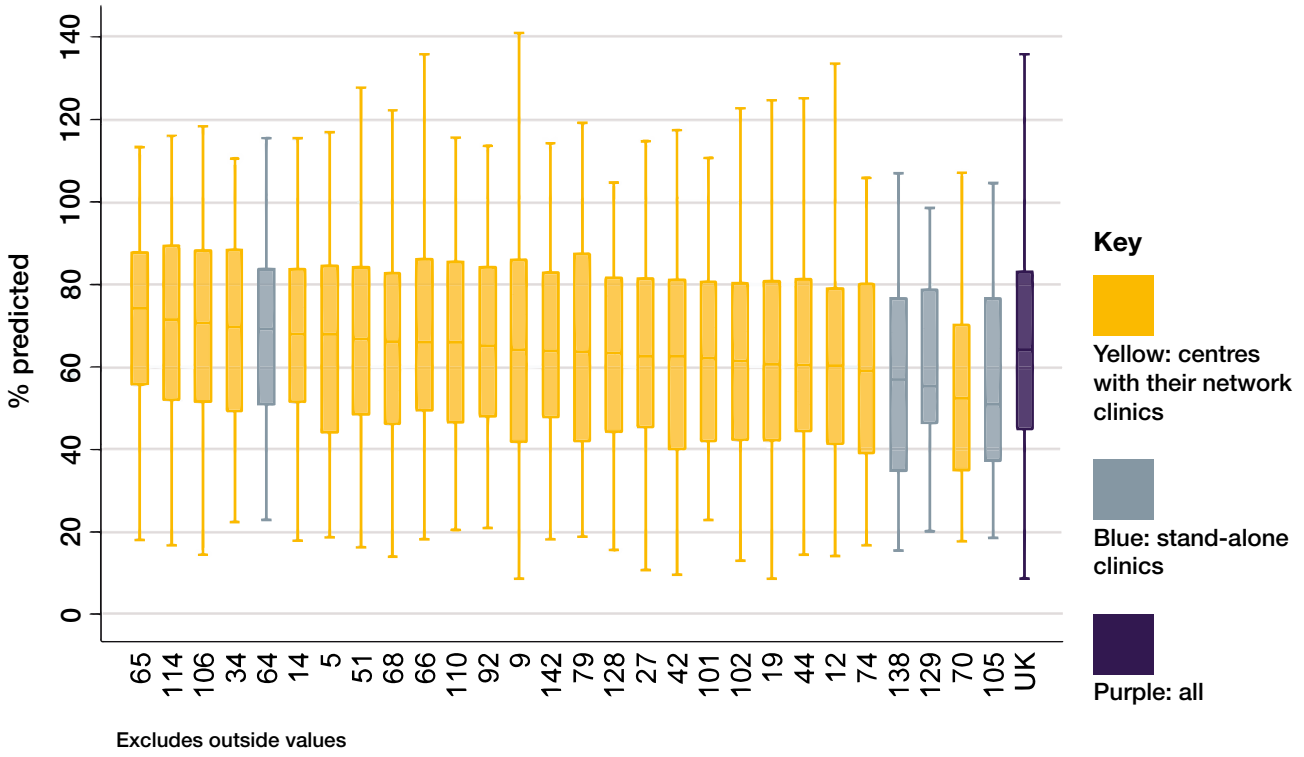
3.1 Median age (years) by adult service



The median age in adult services is 28 years (IQR: 22–36).

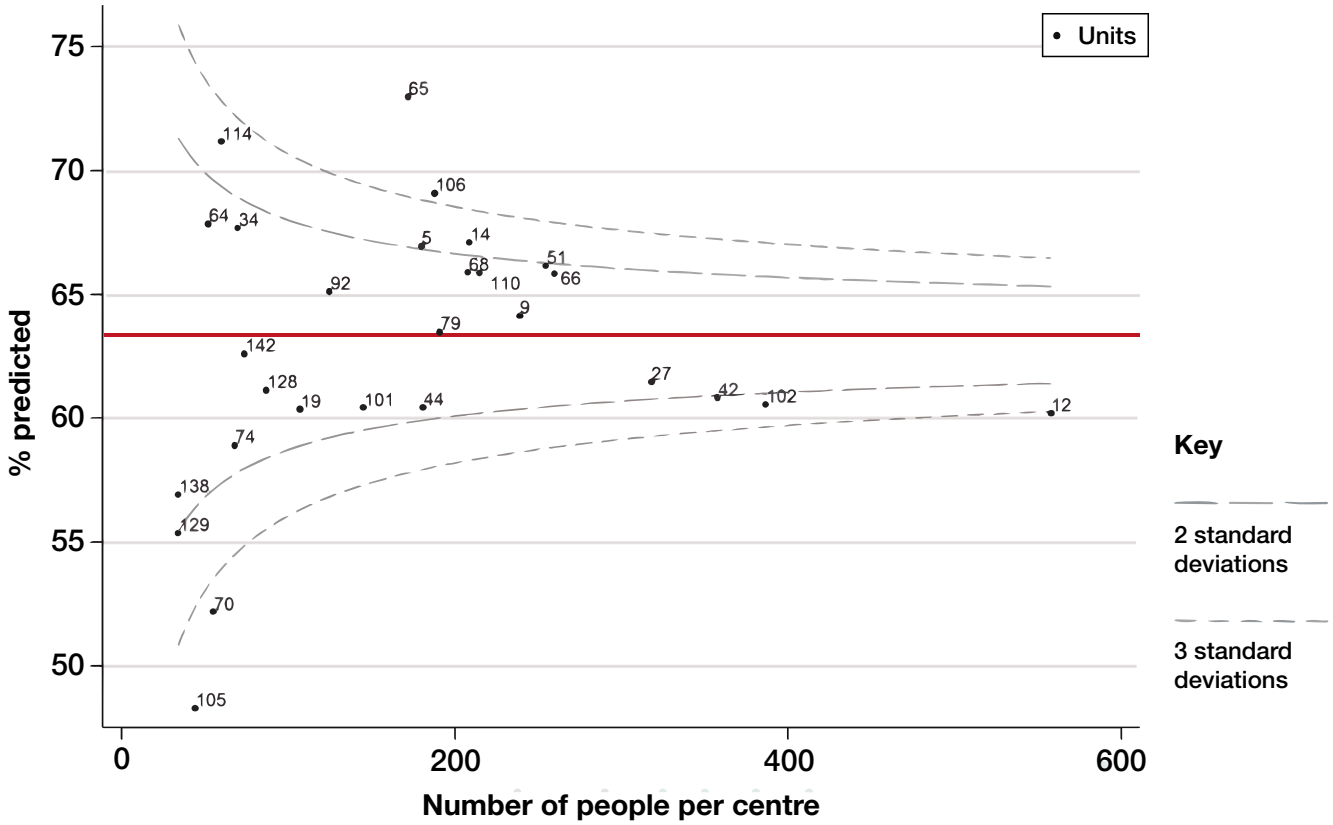


3.2 Median FEV_1 % predicted by adult service (without a history of lung transplant) (GLI equations)

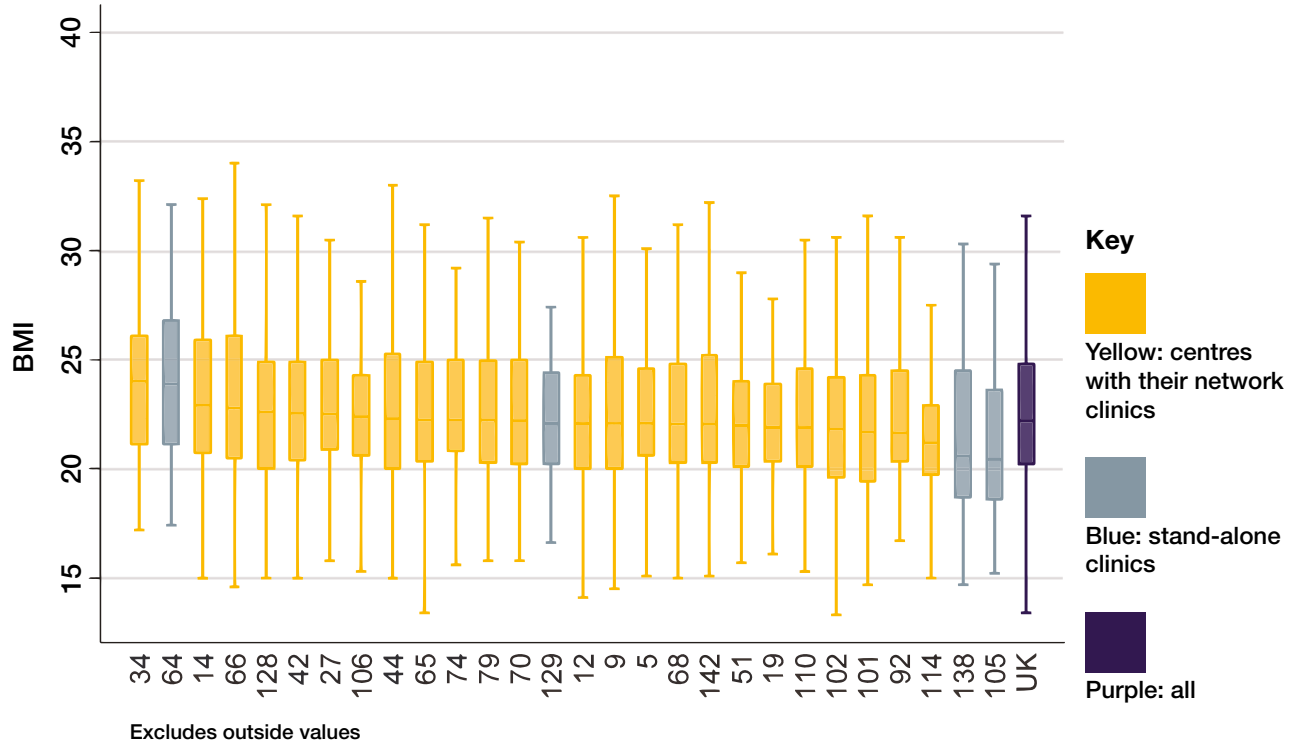


Excludes outside values

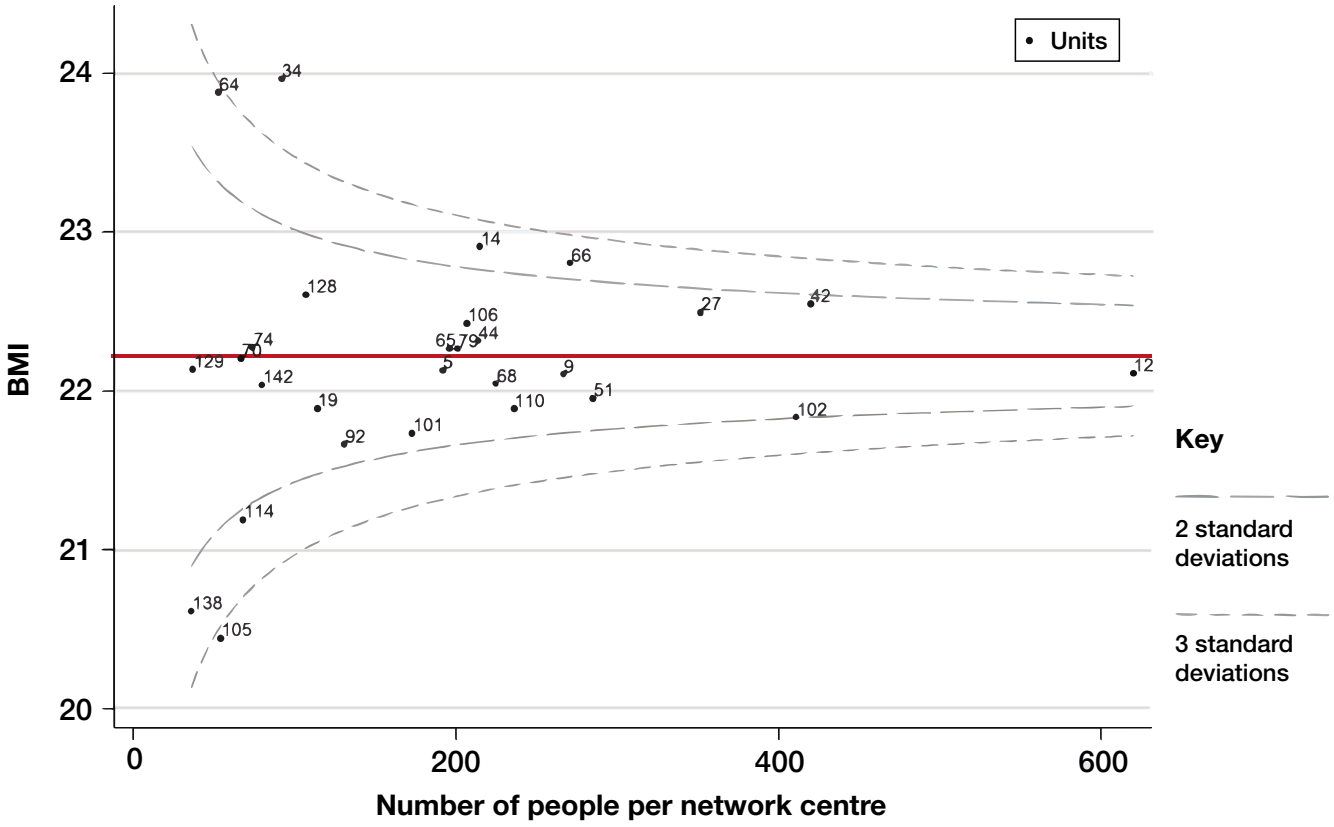
The median FEV_1 (% predicted) in adult services is 64% (IQR: 45–83).



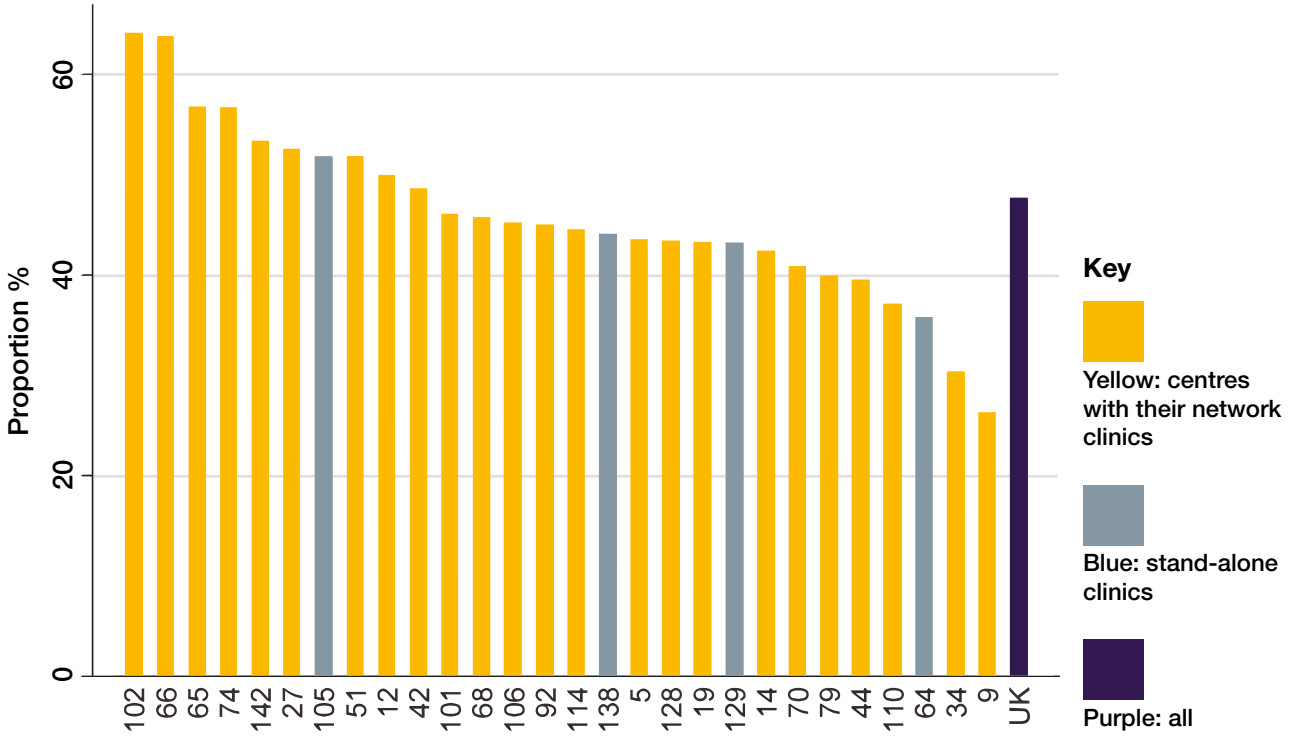
3.3 Median BMI among patients aged 16 years and older by adult service



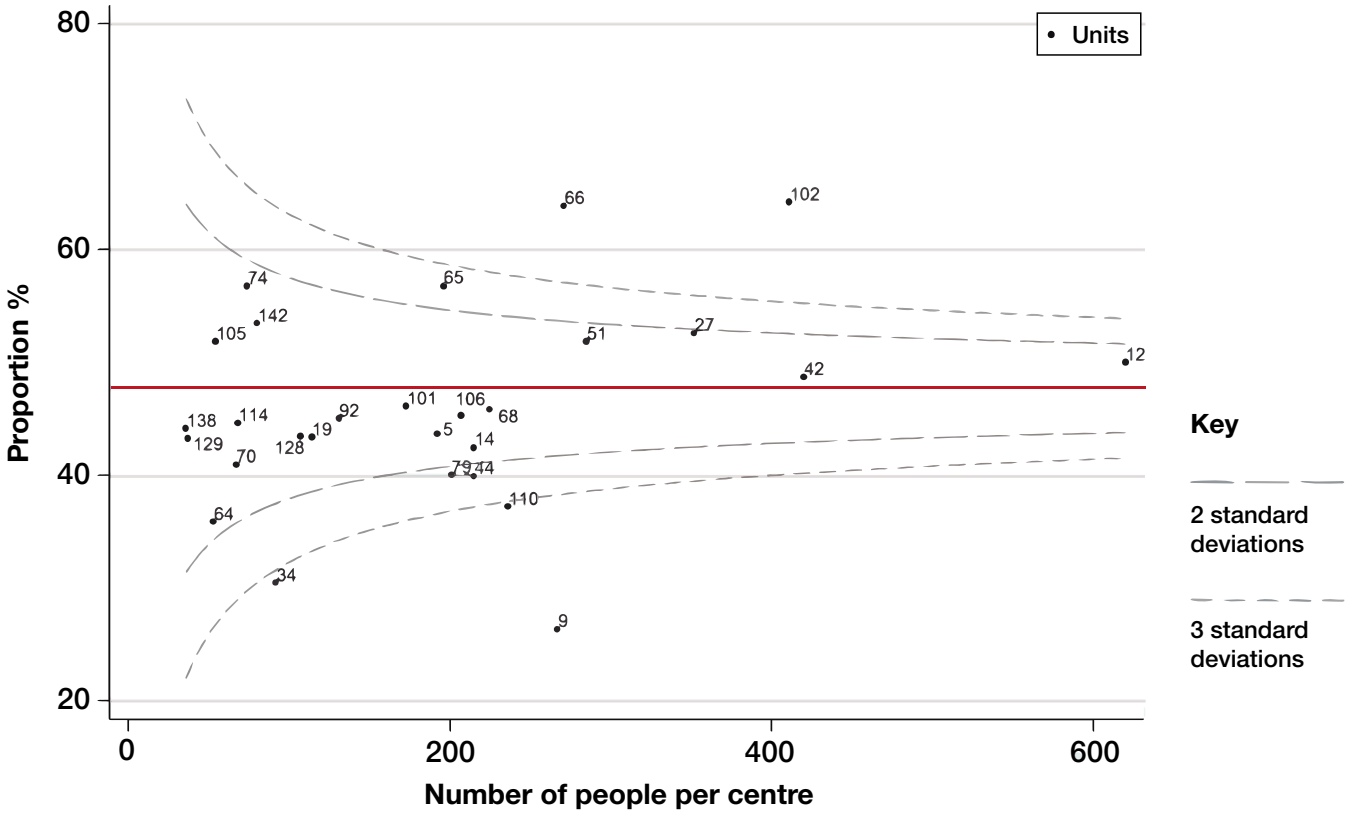
The median BMI in adult services is 22 (IQR: 20–25).



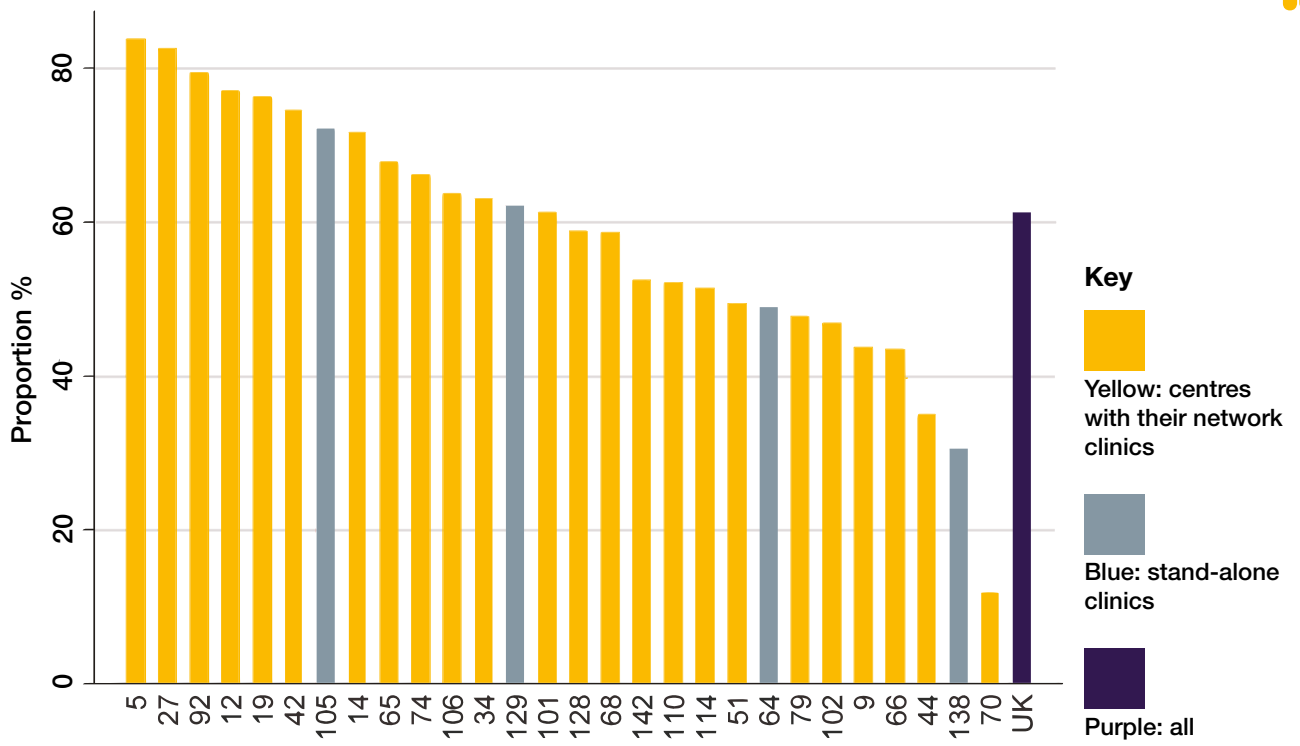
3.4 Proportion of patients with chronic *Pseudomona aeruginosa* by adult service



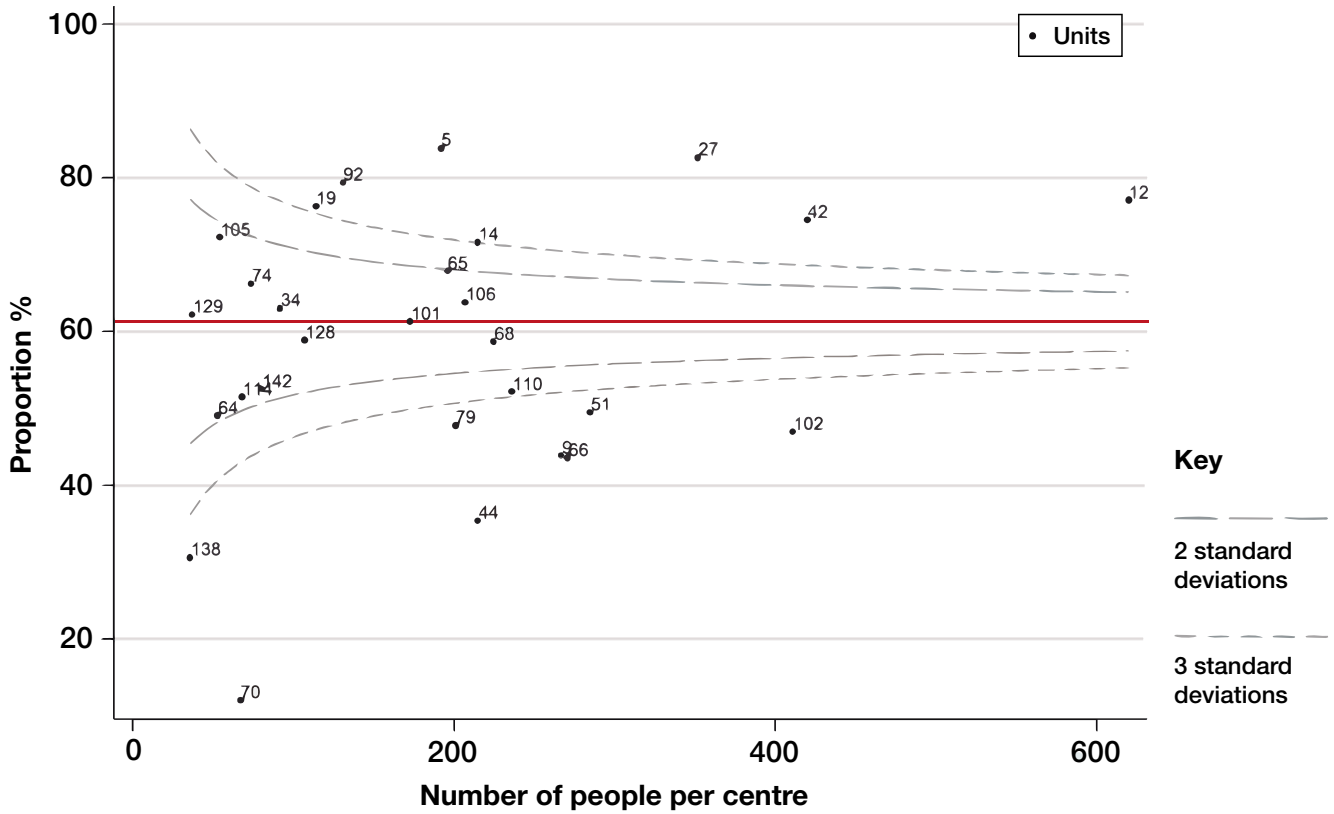
The proportion of patients with chronic *P. aeruginosa* in adult centres/clinics is 48%.



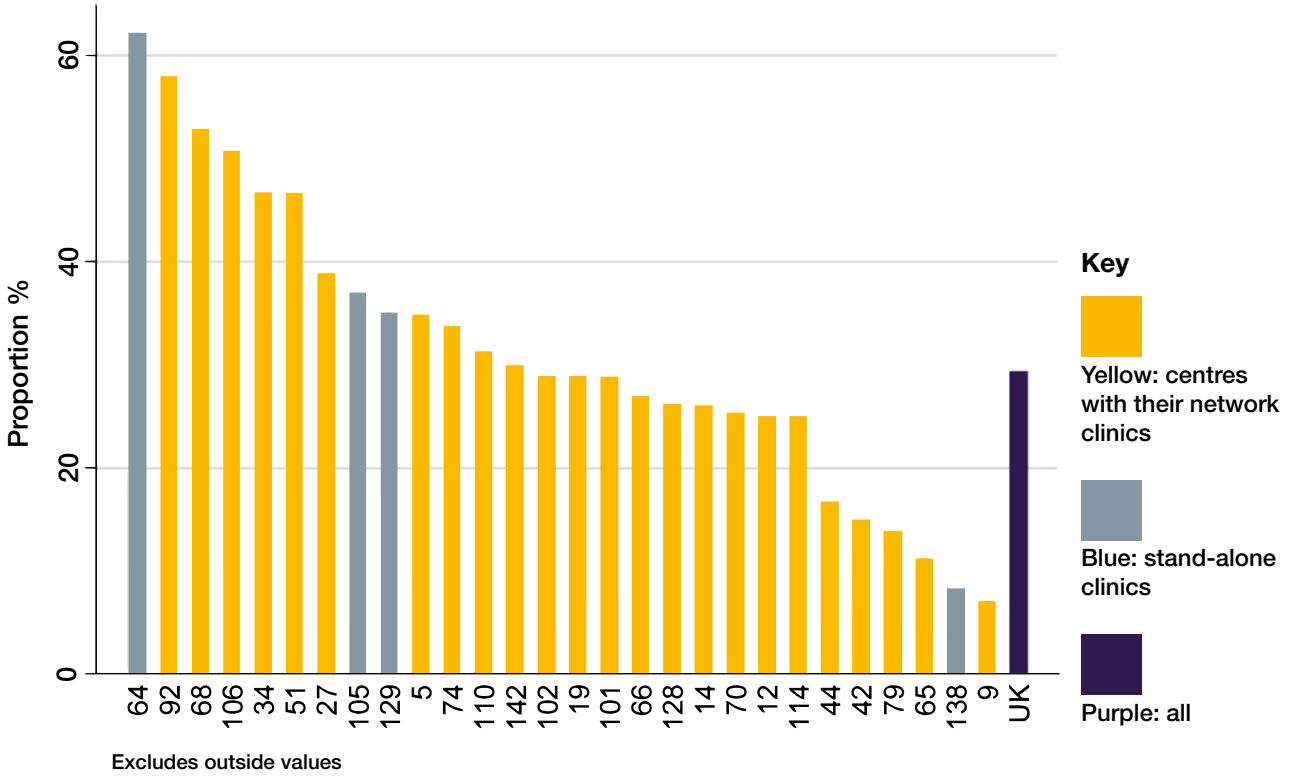
3.5 Proportion of patients receiving DNase treatment by adult service



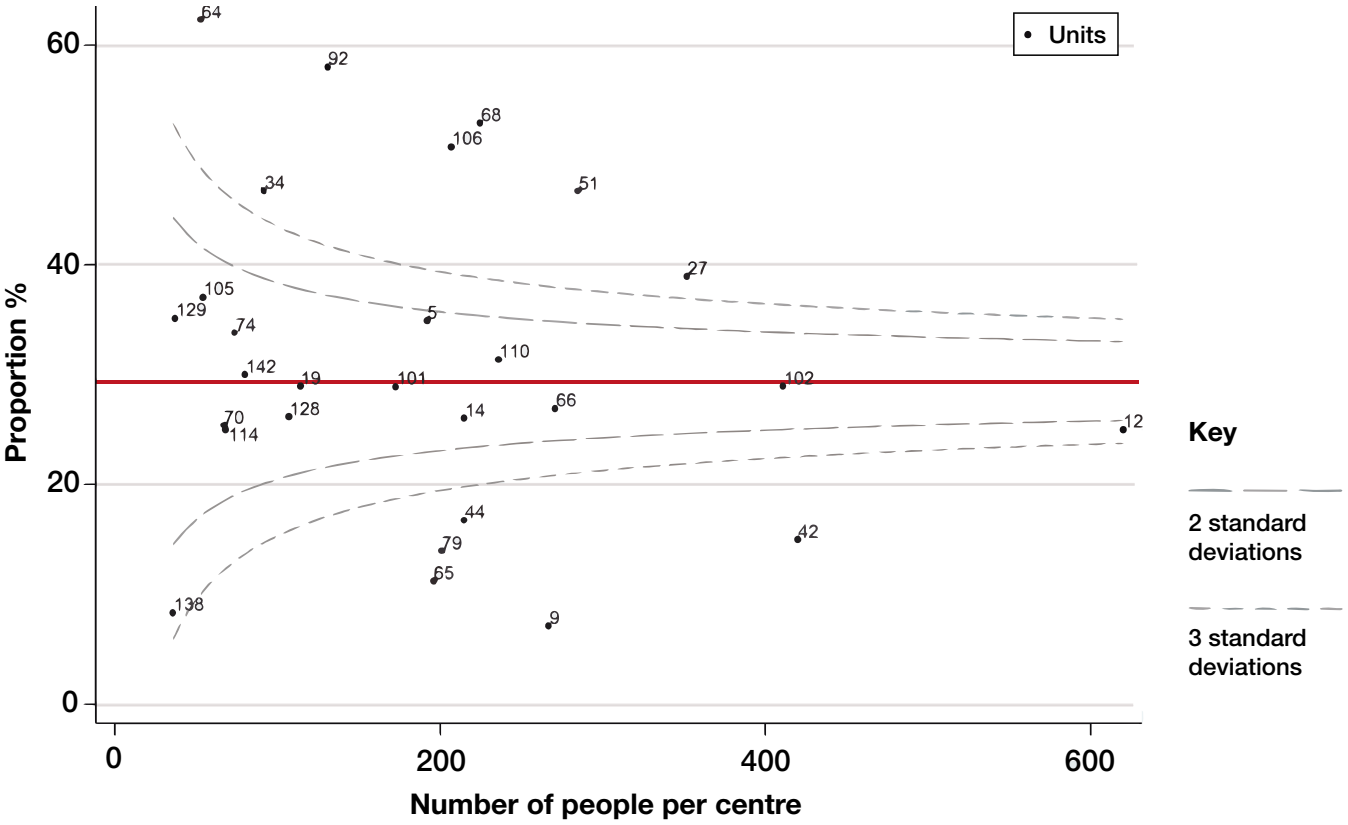
The proportion of patients receiving DNase treatment in adult centres/clinics is 61%.



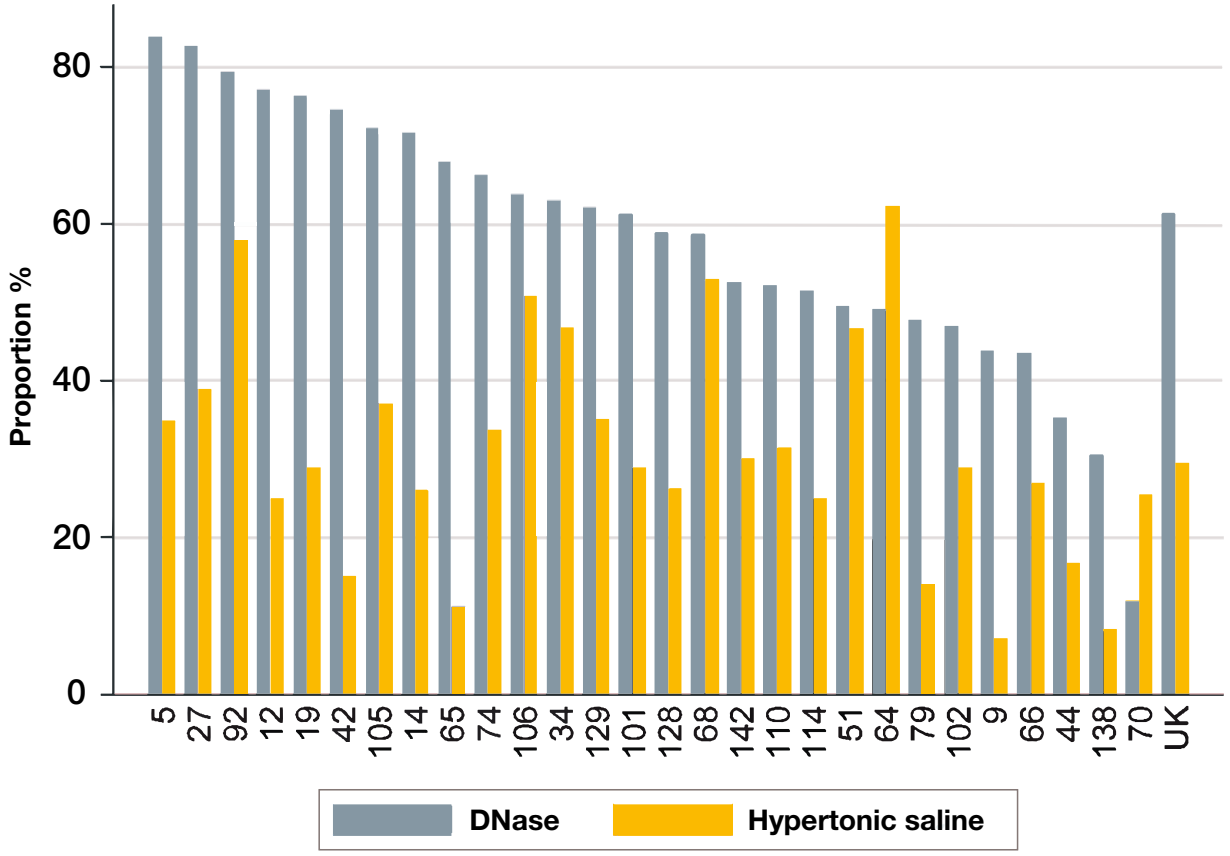
3.6 Proportion of patients receiving hypertonic saline treatment by adult service



The proportion of patients receiving hypertonic saline treatment in adult centres/clinics is 29%.



3.7 DNase and hypertonic saline use by adult service



Appendix 1: Centre level data tables

Paediatric centres/clinics providing data in 2015 – ordered by clinic ID

Country	Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2015	Median FEV_1% predicted (≥6 years)	Median BMI percentile (2-15 years)
England	Leicester	Leicester Royal Infirmary	1	67	66	91.6	64.9
England	Sheffield	Sheffield Children's Hospital	3	140	136	94.3	56.1
England	North West Staffs	North West Staffs, Stoke on Trent	8	93	90	87.8	59.7
England	London-South West	Royal Brompton Hospital	15	335	319	88.0	60.2
England	London - South East	King's College Hospital	17	205	200	86.0	53.4
England	Oxford	John Radcliff Hospital	22	181	175	90.6	52.2
England	Leeds	St James University Hospital	25	228	225	84.9	51.8
England	Southampton	Southampton General Hospital	29	208	201	88.9	51.9
England	London - East	Royal London Hospital	30	119	116	86.9	57.2
Scotland	Inverness	Raigmore Hospital	31	16	16	99.2	39.2
England	Bristol	Bristol Royal Hospital for Children	32	178	173	86.1	48.8
Scotland	Glasgow	Royal Hospital for Sick Children	56	118	87	94.6	58.6
England	Newcastle	Royal Victoria Infirmary	59	174	174	89.9	61.2
Northern Ireland	Belfast	Royal Belfast Hospital for Sick Children	60	197	188	91.7	65.3
England	Nottingham	Nottingham University Hospitals	62	168	162	84.2	55.6
England	Teeside	James Cook University Hospital	71	60	60	82.8	52.2
Wales	Cardiff	Children's Hospital for Wales	72	161	157	86.7	62.4

Country	Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2015	Median FEV_1% predicted (≥6 years)	Median BMI percentile (2-15 years)
Scotland	Dundee	Ninewells Hospital	73	22	20	84.3	55.4
Scotland	Aberdeen	Royal Aberdeen Children's Hospital	75	29	28	93.9	58.3
England	London-Central	Great Ormond Street Hospital for Children	90	185	179	90.4	41.8
England	Cornwall	Royal Cornwall Hospital	94	30	29	96.3	76.6
England	Exeter	Royal Devon & Exeter Hospital	96	71	69	96.5	52.9
England	Liverpool	Alder Hey Children's Hospital	97	301	297	82.6	56.6
England	Norwich	Norfolk & Norwich University Hospital	98	64	64	88.2	65.5
England	Birmingham	Birmingham Children's Hospital	104	297	284	88.2	54.0
England	Cambridge	Addenbrookes Hospital	107	133	127	91.6	53.3
England	Hull	Hull Royal Infirmary	111	33	33	72.0	63.4
England	Plymouth	Derriford Hospital	139	39	39	72.8	46.0
Scotland	Edinburgh	Royal Hospital for Sick Children	143	123	121	93.7	58.3
England	Manchester	Royal Manchester Children's Hospital	144	323	313	86.6	56.0
Scotland	Lanarkshire	Wishaw General Hospital	162	47	40	90.7	46.0

Adult centres/clinics providing data in 2015 – ordered by clinic ID

Country	Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2015	Median FEV_1% predicted (≥16 years)	Median BMI percentile (≥16 years)
England	London - South East	King's College Hospital	5	211	192	67.8	22.1
England	Newcastle	Royal Victoria Infirmary	9	274	267	64.1	22.1
England	London - South West	Royal Brompton Hospital	12	671	620	60.3	22.1
Northern Ireland	Belfast	Belfast City Hospital	14	256	215	67.8	22.9
England	Frimley	Frimley Park Hospital	19	119	114	60.7	21.9
England	Birmingham	Birmingham Heartlands Hospital	27	365	352	62.6	22.5
England	Exeter	Royal Devon & Exeter Hospital	34	94	92	69.7	24
England	Leeds	St James's University Hospital	42	437	420	62.5	22.6
Scotland	Edinburgh	Western General Hospital	44	228	215	61.3	22.3
England	Cambridge	Papworth Hospital	51	308	285	66.8	22.0
England	Plymouth	Derriford Hospital	64	53	53	69.1	23.9
England	Sheffield	Northern General Hospital	65	199	196	74.1	22.3
England	Liverpool	Liverpool Heart and Chest Hospital	66	286	271	66.0	22.8
Wales	Llandough	Llandough Hospital	68	225	225	66.1	22.1
Scotland	Aberdeen	Aberdeen Royal Infirmary	70	69	67	52.4	22.2
England	North West Staff	North West Staffs, Stoke on Trent	74	77	74	59.1	22.3
Scotland	Glasgow	Gartnavel General Hospital	79	217	201	63.7	22.3
England	London - East	St. Bartholomew's Hospital	92	156	131	65.1	21.7

Country	Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2015	Median FEV_1% predicted (≥16 years)	Median BMI percentile (≥16 years)
England	Nottingham	Nottingham University Hospitals	101	173	173	62.1	21.7
England	Manchester	Wythenshawe Hospital	102	426	411	61.4	21.9
England	London - South East	University Hospital Lewisham	105	54	54	50.8	20.5
England	Bristol	Bristol Royal Infirmary	106	210	207	70.6	22.4
England	Southampton	Southmapton General Hospital	110	251	236	65.9	21.9
England	Norwich	Norfolk & Norwich University Hospital	114	68	68	71.5	21.2
England	Oxford	Churchill Hospital	128	111	107	63.3	22.6
England	Cornwall	Royal Cornwall Hospital	129	38	37	55.3	22.1
England	Hull	Castle Hill Hospital	138	40	36	56.9	20.6
England	Leicester	Glenfield Hospital	142	81	80	63.8	22.1

Paediatric centres/clinics providing data in 2015 – ordered alphabetically by city

Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2015	Median FEV_1% predicted (≥6 years)	Median BMI percentile (2-15 years)
England						
Birmingham	Birmingham Children's Hospital	104	297	284	88.2	54.0
Bristol	Bristol Royal Hospital for Children	32	178	173	86.1	48.8
Cambridge	Addenbrookes Hospital	107	133	127	91.6	53.3
Cornwall	Royal Cornwall Hospital	94	30	29	96.3	76.6
Exeter	Royal Devon & Exeter Hospital	96	71	69	96.5	52.9
Hull	Hull Royal Infirmary	111	33	33	72.0	63.4
Leeds	St James University Hospital	25	228	225	84.9	51.8
Leicester	Leicester Royal Infirmary	1	67	66	91.6	64.9
Liverpool	Alder Hey Children's Hospital	97	301	297	82.6	56.6
London - East	Royal London Hospital	30	119	116	86.9	57.2
London - South East	King's College Hospital	17	205	200	86.0	53.4
London - South West	Royal Brompton Hospital	15	335	319	88.0	60.2
London - Central	Great Ormond Street Hospital for Children	90	185	179	90.4	41.8
Manchester	Royal Manchester Children's Hospital	144	323	313	86.6	56.0
Newcastle	Royal Victoria Infirmary	59	174	174	89.9	61.2
North West Staffs	North West Staffs, Stoke on Trent	8	93	90	87.8	59.7

Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2015	Median FEV_1% predicted (≥6 years)	Median BMI percentile (2-15 years)
Norwich	Norfolk & Norwich University Hospital	98	64	64	88.2	65.5
Nottingham	Nottingham University Hospitals	62	168	162	84.2	55.6
Oxford	John Radcliff Hospital	22	181	175	90.6	52.2
Plymouth	Derriford Hospital	139	39	39	72.8	46.0
Sheffield	Sheffield Children's Hospital	3	140	136	94.3	56.1
Southampton	Southampton General Hospital	29	208	201	88.9	51.9
Teeside	James Cook University Hospital	71	60	60	82.8	52.2
Northern Ireland						
Belfast	Royal Belfast Hospital for Sick Children	60	197	188	91.7	65.3
Scotland						
Aberdeen	Royal Aberdeen Children's Hospital	75	29	28	93.9	58.3
Dundee	Ninewells Hospital	73	22	20	84.3	55.4
Edinburgh	Royal Hospital for Sick Children	143	123	121	93.7	58.3
Glasgow	Royal Hospital for Sick Children	56	118	87	94.6	58.6
Inverness	Raigmore Hospital	31	16	16	99.2	39.2
Lanarkshire	Wishaw General Hospital	162	47	40	90.7	46.0
Wales						
Cardiff	Children's Hospital for Wales	72	161	157	86.7	62.4

Adult centres/clinics providing data in 2015 – ordered alphabetically by city

Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2015	Median FEV_1% predicted (≥16 years)	Median BMI percentile (≥16 years)
England						
Birmingham	Birmingham Heartlands Hospital	27	365	352	62.6	22.5
Bristol	Bristol Royal Infirmary	106	210	207	70.6	22.4
Cambridge	Papworth Hospital	51	308	285	66.8	22.0
Cornwall	Royal Cornwall Hospital	129	38	37	55.3	22.1
Exeter	Royal Devon & Exeter Hospital	34	94	92	69.7	24
Frimley	Frimley Park Hospital	19	119	114	60.7	21.9
Hull	Castle Hill Hospital	138	40	36	56.9	20.6
Leeds	St James's University Hospital	42	437	420	62.5	22.6
Leicester	Glenfield Hospital	142	81	80	63.8	22.1
Liverpool	Liverpool Heart and Chest Hospital	66	286	271	66.0	22.8
London-South East	University Hospital Lewisham	105	54	54	50.8	20.5
London-East	St. Bartholomew's Hospital	92	156	131	65.1	21.7
London-South East	King's College Hospital	5	211	192	67.8	22.1
London-South West	Royal Brompton Hospital	12	671	620	60.3	22.1
Manchester	Wythenshawe Hospital	102	426	411	61.4	21.9
Newcastle	Royal Victoria Infirmary	9	274	267	64.1	22.1
North West Staff	North West Staffs, Stoke on Trent	74	77	74	59.1	22.3

Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2015	Median FEV_1% predicted (≥16 years)	Median BMI percentile (≥16 years)
Norwich	Norfolk & Norwich University Hospital	114	68	68	71.5	21.2
Nottingham	Nottingham University Hospitals	101	173	173	62.1	21.7
Oxford	Churchill Hospital	128	111	107	63.3	22.6
Plymouth	Derriford Hospital	64	53	53	69.1	23.9
Sheffield	Northern General Hospital	65	199	196	74.1	22.3
Southampton	Southampton General Hospital	110	251	236	65.9	21.9
Northern Ireland						
Belfast	Belfast City Hospital	14	256	215	67.8	22.9
Scotland						
Aberdeen	Aberdeen Royal Infirmary	70	69	67	52.4	22.2
Edinburgh	Western General Hospital	44	228	215	61.3	22.3
Glasgow	Gartnavel General Hospital	79	217	201	63.7	22.3
Wales						
Llandough	Llandough Hospital	68	225	225	66.1	22.1

Appendix 2: Glossary

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

Word/Phrase	Meaning
2015	1 January 2015 – 31 December 2015.
ABPA (Allergic Bronchopulmonary Aspergillosis)	When a person develops a respiratory allergic reaction to the 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 wheeze.
B. cepacia	*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)	This is 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.
Confidence interval	Confidence intervals are calculated to show the range of results we would expect, based on the overall average. If a result is between the upper and lower limits of the confidence interval, it is 'as expected'.
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.

Word/Phrase	Meaning
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 (Global Lung Function Initiative) equations	An equation for calculating FEV_1% predicted that takes into account age, gender, height and ethnicity.
Haemophilus influenza	*Haemophilus influenza (H. influenzae)* is a bacterium that can cause respiratory infection.
Haemoptysis	The coughing up of blood.
Hepatobiliary disease	A liver or biliary disorder.
Heterozygous	Everyone living with cystic fibrosis has two CF-causing mutations of the gene for CFTR, one inherited from their mother and one from their father. Someone who has two different mutations is heterozygous.
Homozygous	Everyone living with cystic fibrosis has two CF-causing 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.
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 2015 the median age of the 125 people who died was 28.
Median predicted survival	Median predicted survival is a calculation based on people with CF recorded in the Registry as alive in the given year. A mathematical formula, which takes into account the age of those people in 2015, predicts how long we expect half of them to live for. For 2015, this means that half of people registered as alive on the database are predicted to live to at least 45.1. Half of people alive today are 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.

Word/Phrase	Meaning
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 have been reported.
Nasal Polyps	Small, sac-like growths of inflamed mucus caused by chronic inflammation of the nasal lining.
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 above 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 diagnosed with a condition at any time.
Pseudomonas aeruginosa	A tough bacterial strain, rarely affecting healthy people, that can cause respiratory infection.
Rectal prolapse	When the rectal wall slides through the anus.
Renal	Relating to the kidneys.
Staphylococcus aureus	*Staphylococcus aureus (S. aureus)* is a bacteria that can cause respiratory infection.
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 3: UK CF Registry Steering Committee structure

UK CF Registry Steering Committee

Role	Forename	Surname	Organisation
Allied health professional	Alan	Peres	Royal Brompton Hospital
Analytical team rep †	Stephen and/or Vian	Nyangoma Rajabzadeh-Heshejin	Imperial College London
Commissioner, England	Sue or Carrie	Sawyer Gardner	NHS England
Commissioner, Scotland	David	Steele	NHS Scotland
Commissioner, Wales †	Claire	Nelson	NHS Wales
Cystic fibrosis centre data manager	Lance	Dennard	Lewisham Hospital
CF physician - Adults	Caroline	Elston	King's College Hospital
CF physician - Adults	Simon	Range	Glenfield Hospital
CF physician - Paediatrics	Iolo	Doull	Children's Hospital for Wales
CF physician – Paediatrics*	Siobhán	Carr	Royal Brompton Hospital
Chair of the Research Committee	Martin	Wildman	Northern General Hospital
Director of Impact	Keith	Brownlee	Cystic Fibrosis Trust
Parent representative	Grant	Valentine	N/A
Person with CF	James	Thomson	N/A
Registry Data Manager †	Elaine	Gunn	Cystic Fibrosis Trust
Registry Coordinator †	Annie	Jeffery	Cystic Fibrosis Trust
Registry Lead	Rebecca	Cosgriff	Cystic Fibrosis Trust

* Chair
† Non-voting member
# Caldicott guardian

UK CF Registry Research Committee

Role	Forename	Surname	Organisation
Analytical team rep †	Stephen and/or Vian	Nyangoma Rajabzadeh-Heshejin	Imperial College London
Pharmacovigilance PI	Diana	Bilton	Royal Brompton Hospital
CF physician – adults (retired)			
Pharmacovigilance PI	Siobhán	Carr	Royal Brompton Hospital
CF physician - paediatrics			
Pharmacovigilance PI	Steve	Cunningham	Royal Hospital for Sick Children
CF physician - paediatrics			
Registry Lead	Rebecca	Cosgriff	Cystic Fibrosis Trust
Parent representative	Marian	Dmochowska	N/A
Person with CF	Dominic	Kavanagh	Cystic Fibrosis Trust
CF physician - adults*#	Martin	Wildman	Northern General Hospital
Registry consultant	Noreen	Caine	Cystic Fibrosis Trust
Registry Data Manager †	Elaine	Gunn	Cystic Fibrosis Trust
Registry Coordinator †	Annie	Jeffery	Cystic Fibrosis Trust

* Chair
† Non-voting member
# Caldicott guardian

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