Cystic Fibrosis strength in numbers

UK Cystic Fibrosis Registry
2014 Annual Data Report

Published August 2015

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An at-a-glance version of this report can be found at 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 centres and clinics, for their continued dedication to obtaining consent and submitting data to the Registry.

The Cystic Fibrosis Trust and the Registry Steering Committee would like to extend a special thank you to Professor Diana Bilton, who has been instrumental in setting up and developing the UK CF Registry as Chair of the Registry Steering Committee from 2007–2014.

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Foreword

I am delighted to introduce the UK Cystic Fibrosis Registry 2014 Annual Data Report.

We are living through a time of extraordinary hope for people with cystic fibrosis (CF) and 2014 is the first year that the UK CF Registry has been able to publish information about the effect of ivacaftor, a transformational new treatment licensed for those with the G551D mutation. This therapy is the first of a potential pipeline of personalised treatment that tackles the root cause of CF, rather than its symptoms. As more treatments become available to an increasing number of people with CF, so data from the UK CF Registry will be instrumental in informing decisions by the NHS on whether and when to fund these expensive medications.

This year, the Registry includes the data from 10,583 people, representing over 99% of people with CF in the UK. This shows an increase in the UK CF population of over 1,000 people since 2010; with an increase in the proportion of adults with the condition from 55.5% in 2010 to 59.3% in 2014.

It is tremendous news that more people with CF are living to adulthood. Yet this brings with it new challenges. Providing care and support for people who are living longer, and experiencing additional complications of CF such as CF-related diabetes (shown to be present in almost a third of adults with CF in 2014), is a problem that will take the greatest minds, together with abundant and reliable healthcare data, to solve.

The report also shows continued successes across the UK in reducing the rate of pseudomonas infections, although no real change in the incidence of nontuberculosis mycobacterium (NTM) (1.2% this year down from 1.3% in 2013).

As the population of people with CF evolves, so too does its Registry. Early in 2015 the UK CF Registry surveyed the community's views on data sharing, protection, and reporting. We were told loud and clear that people with CF and their families want all of the information we can possibly give them, in as much detail as possible. I hope we have taken the first, modest step in meeting this request with this report. Previously we have published a summary report for people with cystic fibrosis, and a detailed report for the scientific community. This year we publish a detailed report, and an at-a-glance version, both intended to be accessible to all audiences, giving people with cystic fibrosis access to as much information as they desire.

Developing the way that we report results from the UK CF Registry back to the wider CF community is a journey that we need everyone's help to complete. Any feedback on this report, positive or not, is very welcome and can be sent to us using the information in the 'contact us' section of this report (page 5).

The UK CF Registry survey report is available in detailed and at-a-glance form at **www.cysticfibrosis.org.uk/registry.**

Another important development on the horizon for the UK CF Registry is an update to the software that supports it. Due to launch in early 2016, the new Registry system will provide healthcare teams with more advanced tools to help them enter data, as well as keep track of what it shows them, to constantly work towards improving care for people with cystic fibrosis. The new platform will, in due course, also enable people with cystic fibrosis to enter and access their own data, something else people with CF have told us they want.

Finally, I want to take this opportunity to thank all those who help to make the Registry such an extraordinary asset to the CF community – from the multidisciplinary clinical teams in CF centres across the UK to the thousands of supporters whose fundraising efforts make it all possible. Most of all, I want to thank all those with cystic fibrosis and their guardians who give their consent for their data to be collected and used in this way. The Registry is a tremendous example of how our community works together for the common good, and we are committed to protecting and developing it to deliver maximum impact on the lives of people with cystic fibrosis.

Ed Owen
Chief Executive

Executive summary

The UK CF Registry holds more vital information now than ever before; not only do we have greater numbers of people with cystic fibrosis, 10,583 in 2014, but the quality of data is still improving. In 2014, 89% of people registered onto the database have complete data. To date 97.7% of people have a genotype value recorded, this is at a time when it is increasingly important for people with cystic fibrosis and their care teams to be aware of their genotypes, because of the new personalised therapies.

With these large numbers comes a greater confidence in the results we are able to communicate to the cystic fibrosis community. These include the fact that people with cystic fibrosis are now diagnosed earlier – something that is key to early intervention and better health. In 2014, after several years of plateau, the median age of diagnosis across the whole population has dropped to two months, which means we are finally seeing the effects of the newborn screening 'heel prick' programme. 91.6% of the under-five age group are now diagnosed by newborn screening. As expected with any screening programme, it will not pick up 100% of babies born with cystic fibrosis, and both adult and paediatric clinicians still need to be alert for people presenting with cystic fibrosis on clinical grounds.

We report the median predicted survival as 40.1 years in 2014. You will note this figure has fluctuated over the last few years. This variation in predicted survival is because there are small numbers of deaths relative to the whole cystic fibrosis population. The proportion of adults (>16 years) with cystic fibrosis is continuing to increase at 59.3%, with 70.1% of these adults reporting being either in work or education. This report suggests lung function (as shown by FEV_1), appears to be better across most age ranges than our comparison base year of 2008. The decrease in the prevalence of chronic *Pseudomonas aeruginosa*, also shown last year, has been maintained, with a peak of 57.7% in adults aged 28–31. However, the prevalence of *Staphylococcus aureus* remains about the same at around the 20% mark once adulthood is reached.

The prevalence of other complications such as ABPA (10.8%) remains fairly static.

CF-related diabetes continues to be well screened for and there is a slight suggestion that treatment for this complication has been increasing over the last few years, with 32.3% of adults currently recorded as on treatment.

Data on transplantation is shown over the last five years: showing a steady rise in the number evaluated for transplant (247), the number accepted on to transplant lists (146) and the number receiving transplants, with 59 people recorded as having bilateral lung transplants in 2014.

There have been impressive increases in the use of preventative inhaled medication such as DNase (54.5%) and hypertonic saline (26.1%), with peak use for DNase being in the 12–19 year age group. The consensus goal of 90% of people with chronic Pseudomonas being on a suitable suppressive inhaled antibiotic is nearly achieved this year, with 88.6% recorded on one of these therapies compared to 76.1% in 2008. The swap from nebulised antibiotics to dry powdered ones is also apparent and will be interesting to watch over time. The effect of these increases on requirement for rescue medication such as IV antibiotics is currently being explored by research groups.

Finally, centre level data is presented in the second half of the report. As ever, care needs to be taken when interpreting these charts; there is no adjustment for factors that can influence an individual centre's results, such as age of the overall population (in an adult clinic), or genotype distribution.

We hope that you find the contents of this report interesting and useful. We look forward to hearing your views about the report.

Siobhán B Carr
Acting 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

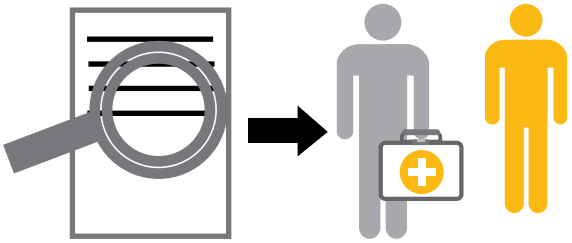
Cystic fibrosis is an inherited disease caused by a faulty gene. The gene and the protein it makes, known as 'CFTR', controls the movement of salt and water in and out of cells. When the gene is faulty, the lungs can become clogged with mucus over time. This damages the lungs. Around 85% of people with 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.

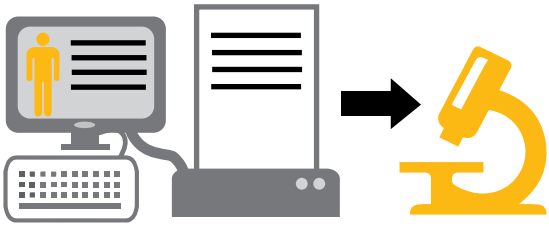
The purpose of the UK CF Registry is to improve the health of people with cystic fibrosis. This is done in a number of ways:

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

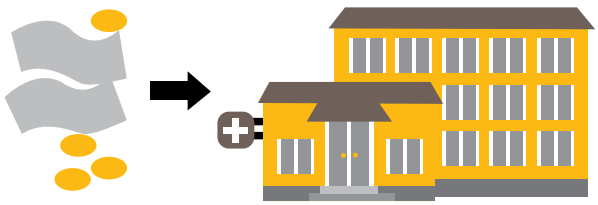


Giving clinical teams the evidence they need to improve the quality of care.

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.

Governance

The Registry Steering Committee (RSC) (Appendix 3) is responsible for making sure that the UK CF Registry is compliant with legislation like the Data Protection Act 1998, and its Research Ethics Study Protocol. The RSC also assesses applications for data, and makes recommendations about the future development of the Registry.

Data are only recorded on the Registry if explicit written consent is given by the patient or, for a child, the patient's carer.

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.

Anonymisation and pseudonymisation of the data 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 cystic fibrosis 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.**

Section 1: UK-wide analysis

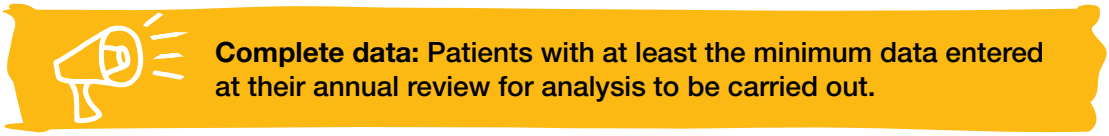
Words in this report that appear in the glossary are highlighted the first time they appear and explained in appendix 2 on page 56.

This section provides an overview of the cystic fibrosis population, health and care in the United Kingdom, including cystic fibrosis centres in England, Northern Ireland, Scotland, and Wales.

1.1 Summary of the UK Cystic Fibrosis Registry

	2010	2011	2012	2013	2014
CF patients registered[1] Excluding diagnoses that year	9385	9749	10078 9804	10338 10076	10583 10356
CF patients with "complete" data[2]**; n(%)**	7937 (85%)	8679 (89%)	8794 (87%)	9052 (88%)	9432 (89%)
Age in years; median[3]	17	18	18	18	19
All newly diagnosed patients[4] **(newborn screening and other)**	301	261	285	301	227
Number of patients born each year identified by newborn screening[4]	241	203	213	177	130
Age at diagnosis in months; median[3]	3	3	3	3	2
Adults aged 16 yrs and over; %[3]	55.5	56.8	57.6	57.6	59.3
Males; %[3]	53.1	53.2	52.9	52.9	53
Genotyped; %[3]	95.2	95.6	96.2	97.2	97.7
Median predicted survival in years (95% Confidence interval)[5]	41.4 (36.8, 46.7)	41.5 (35.7, 46.0)	43.5 (37.8, 49.9)	36.6 (34.4, 41.6)	40.1 (34.6, 46.7)
Total deaths reported (%)[5]	103 (1.1%)	118 (1.2%)	106 (1.1%)	146 (1.4%)	137 (1.5%)
Age at death in years; median (95% CI)[3]	29	26	28 (25, 29)	29 (27, 31)	28 (25.5, 32)

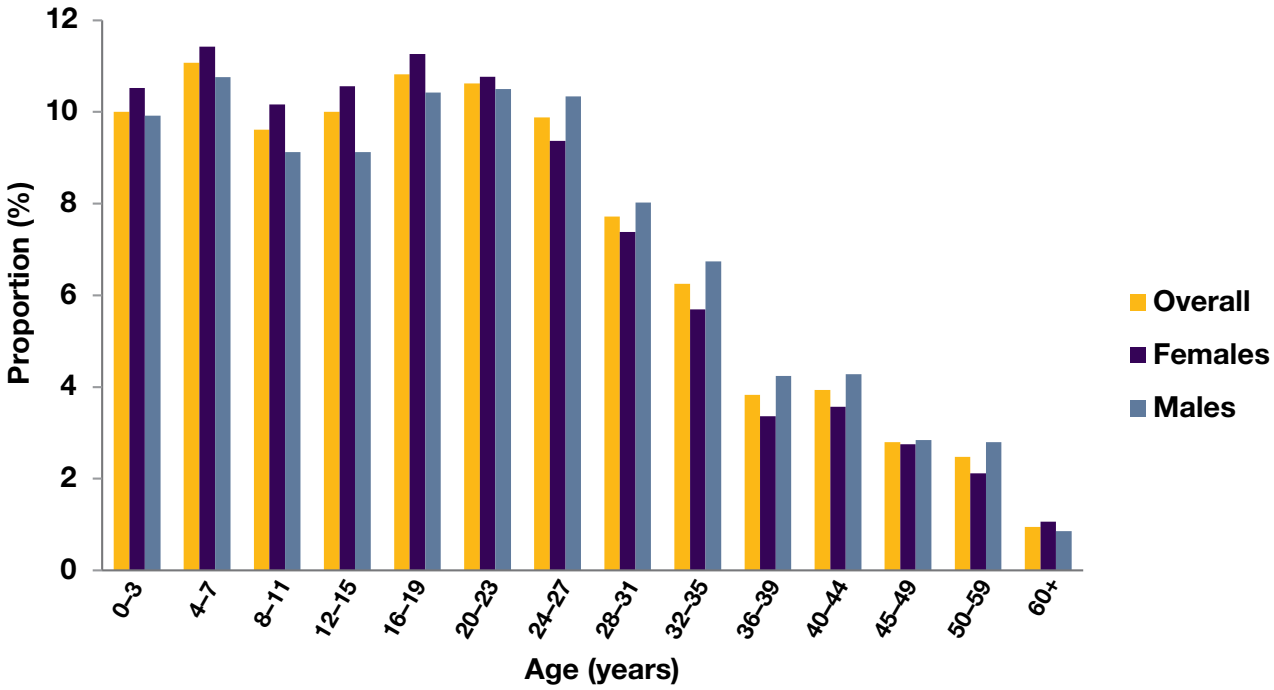
Notes:
[1] The number of patients who were diagnosed with CF and had not died before 1 January in the given year.
[2] A patient has 'complete data' if their team has filled in an annual review for them for that year. Patients newly diagnosed in 2014 may not have their first annual review in the same year. If newly diagnosed patients are excluded, 91% of records are complete.
[3] Calculated from patients with 'complete data' 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.



1.2 Age distribution by gender

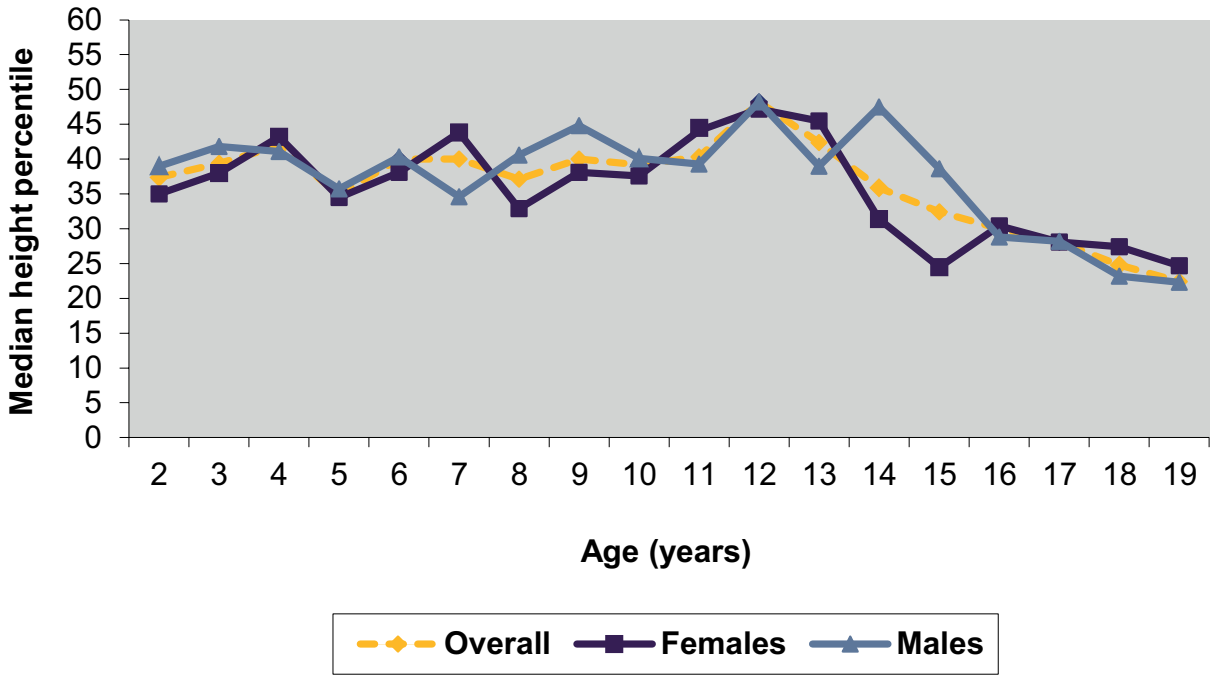
n=9432

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=4461

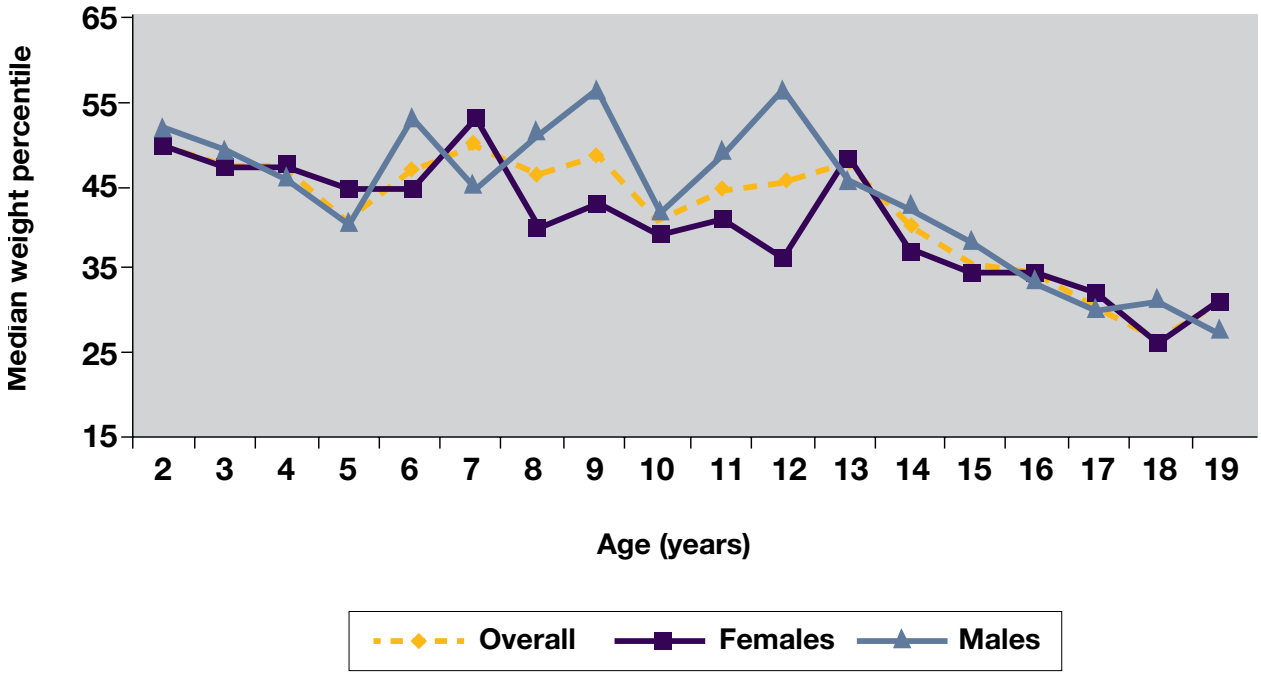
The following chart and table show the height percentiles of people with CF, aged 19 and under, in relation to the UK growth data for the general population. If a person with CF is on the 40th percentile, only 40% of the population at the same age are their height or shorter; 60% are taller.



	Overall			Female			Male		
Age	**N**	**Median**	**IQR**	**N**	**Median**	**IQR**	**N**	**Median**	**IQR**
2	288	37.4	15.6-64.8	133	35	15.5-65.0	155	39	16.3-65.2
3	276	39.4	16.2-68.3	141	38	15.3-70.5	135	41.8	18.5-65.3
4	279	41.9	21.5-63.7	128	43.2	19.5-63.3	151	41.1	22.8-64.7
5	239	34.9	15.4-64.1	121	34.5	13.1-64.6	118	35.7	18.4-63.7
6	278	39	15.4-64.6	134	38.1	15.3-64.4	144	40.3	16.0-65.6
7	248	40	15.7-68.2	123	43.8	15.2-71.0	125	34.6	16.9-67.3
8	217	37.1	16.2-62	104	32.9	15.5-59.5	113	40.6	16.9-63.8
9	238	40	19.2-60.2	112	38.1	17.9-54.9	126	44.8	20.0-74.4
10	225	39.2	16.5-63.4	124	37.6	16.5-63.7	101	40.1	16.5-63.4
11	226	41.3	12.4-65.5	110	44.1	11.0-65.4	116	39.3	13.6-66.7
12	201	48.2	15.6-73.8	102	47.2	13.7-74.2	99	48.2	18.9-73.5
13	237	42.4	17.3-69.1	121	45.5	24.1-69.6	116	39	14.5-67.5
14	231	35.9	11.1-68.5	117	31.4	8.6-59.3	114	47.5	16.1-79.5
15	258	32.4	11.1-62.4	128	24.5	9.5-55.9	130	38.6	15.0-66.3
16	246	29.6	11.1-53.3	110	30.4	8.1-70.0	136	28.8	12.2-49.1
17	242	28.1	10.0-58.3	108	28.1	6.9-55.2	134	28.2	11.0-58.9
18	267	24.8	7.9-57.3	135	27.4	7.70-59.0	132	23.2	9.7-54.7
19	265	22.4	10.3-52.4	146	24.7	10.3-52.4	119	22.3	9.1-54.8
Overall	**4461**	**36.5**	**36.5-34.9**	**2197**	**35.7**	**33.7-37.9**	**2264**	**37**	**34.9-38.9**

1.4 Median weight percentiles among children and young people (<20 years)
n=4461

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

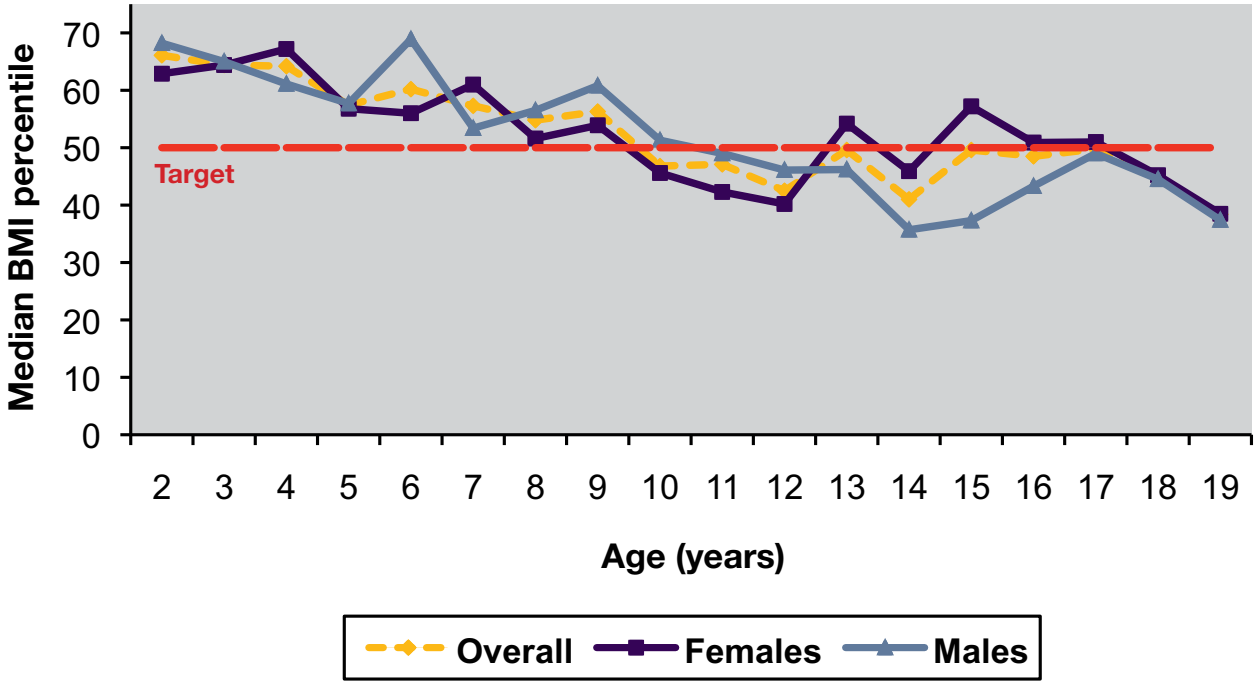


	Overall			Female			Male		
Age	**N**	**Median**	**IQR**	**N**	**Median**	**IQR**	**N**	**Median**	**IQR**
2	288	50	28.2-74.4	133	49.6	17.4-69	155	51.2	28.2-74.5
3	276	48	23.5-76.3	141	47	27.8-74.3	135	49.5	25.1-76.1
4	279	46.2	23.4-69.6	128	47.4	22.1-78.5	151	46	23.8-72.9
5	239	41	18.3-69.3	121	44.6	20.4-67.2	118	40.3	18.5-68.5
6	278	46.9	21.6-73	134	44.1	18.1-70.9	144	53.4	26.1-71.9
7	248	50.3	23.2-74	123	53	18.2-67.9	125	45.2	21.1-71.9
8	217	46.3	22-72.6	104	39.8	25.3-77	113	51.6	23.9-74.7
9	238	48.8	25.6-73.2	112	42.8	19.9-67.9	126	56.9	32.7-79.9
10	225	41	20.2-69.4	124	39.1	19-69.4	101	42.1	24-69.8
11	226	44.8	21.7-70	110	41	18.3-69.7	116	48.8	24.9-73
12	201	45.6	19.1-74.1	102	36.3	16.1-68.1	99	56.4	22.3-79.1
13	237	47.8	22.8-74.9	121	48.4	28.9-74.6	116	45.6	17.9-75.4
14	231	40.2	14.6-70	117	37.2	9-69.7	114	42.7	17.2-70.2
15	258	35.6	15.5-64.4	128	34.5	15.1-59.6	130	38.4	16.9-67.7
16	246	33.7	11.6-63.4	110	34.5	15.7-65.1	136	33.7	8.8-61.9
17	242	30.1	7.5-58.5	108	32.2	8.4-67.7	134	30.1	11.2-60.1
18	267	27	23.9-32.9	135	26.1	7.9-47.5	132	31.6	6.2-62.7
19	265	30.6	8.4-64.1	146	31.3	10-67.1	119	28.1	6.2-62
Overall	**4461**	**42.2**	**18-70.5**	**2197**	**40.8**	**17.4-69.0**	**2264**	**43.4**	**18.6-72.1**

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

n=4461

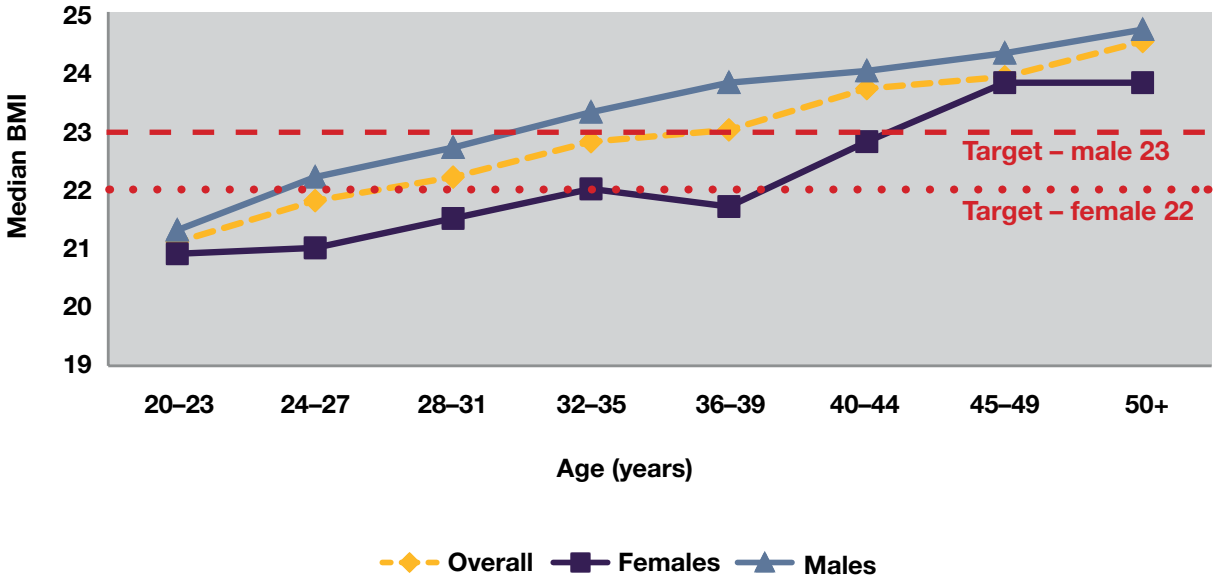
The following chart and table show the BMI percentiles of people with CF, aged 19 and under, in relation to the target BMI percentile for a person of the same age without CF (the 50th percentile, or the BMI percentile that half of the UK population of that age has achieved). If a person with CF is at the 40th percentile, it means that only 40% of people of the same age have the same BMI or lower; 60% have a higher BMI.



	Overall			Female			Male		
Age	**N**	**Median**	**IQR**	**N**	**Median**	**IQR**	**N**	**Median**	**IQR**
2	288	66.1	40.3-83.6	133	62.9	47.9-81	155	68.2	34.8-84.9
3	276	64.4	39.1-85.7	141	64.4	39-82.4	135	65	38.8-87.7
4	279	64.2	35.7-84.3	128	67.2	41.1-81.1	151	61.1	33.3-85.9
5	239	57.4	27.7-77.0	121	56.8	28.8-77.7	118	57.7	27.0-75.6
6	278	60.2	32.6-77.8	134	56	31.7-73.4	144	68.9	33.2-84.0
7	248	57.3	32.2-77.7	123	61	37.8-78.8	125	53.4	25.4-75.1
8	217	54.8	31.8-72.8	104	51.6	29.5-71.7	113	56.5	34.5-72.8
9	238	56.3	36.2-76.7	112	53.9	32.4-73	126	60.8	39.2-77.9
10	225	46.8	25.0-71.4	124	45.6	24.6-72.4	101	51.3	25.5-70.4
11	226	47.1	22.6-69.9	110	42.3	20.9-67.2	116	49	25.-72.5
12	201	42.5	23.0-70.7	102	40.2	22.7-63.6	99	46.1	23.2-79.0
13	237	49.6	23.0-72.4	121	54.2	26.7-73.9	116	46.2	18.5-72
14	231	41	21.3-70.5	117	45.9	23.9-72.9	114	35.7	16.8-61.7
15	258	49.6	21.0-72.8	128	57.2	34.7-73.4	130	37.3	17.5-72.8
16	246	48.5	22.5-76.7	110	50.9	27.1-76.9	136	43.3	15.1-75.3
17	242	49.8	21.9-75.2	108	51	25.4-77.6	134	49	20.3-74.6
18	267	44.9	16.7-73.3	135	45.2	16.3-70.7	132	44.5	17.8-77.9
19	265	38.4	15.0-71.4	146	38.5	15.7-73.8	119	37.7	14.6-68.9
Overall	**4461**	**49.4**	**23.2-74.7**	**2197**	**50.7**	**26.1-73.8**	**2264**	**54**	**26.7-79.1**

1.6 Median Body Mass Index (BMI) values among adults (20 years and over)
n=4472

The following chart and table show the BMI of people with CF aged 20 and over in relation to the target BMI for a healthy adult; 22 for women and 23 for men[1].



Age	N	Overall Median (IQR)	N	Female Median (IQR)	N	Male Median (IQR)
20-23	1002	21.1 (20.9-21.3)	477	20.9 (20.7-21.2)	525	21.3 (21.0-21.8)
24-27	932	21.8 (21.5-22.0)	415	21.0 (20.7-21.4)	517	22.2 (21.9-22.5)
28-31	728	22.2 (22.0-22.4)	327	21.5 (21.2-22.0)	401	22.7 (22.4-23.1)
32-35	589	22.8 (22.3-23.2)	252	22.0 (21.6-22.4)	337	23.3 (22.8-23.6)
36-39	361	23.0 (22.5-23.4)	149	21.7 (21.2-22.1)	212	23.8 (23.2-24.4)
40-44	372	23.7 (23.4-24.0)	158	22.8 (22.2-23.6)	214	24.0 (23.7-24.5)
45-49	264	23.9 (23.7-24.6)	122	23.8 (22.6-24.5)	142	24.3 (23.8-24.9)
50+	324	24.5 (23.8-25.1)	141	23.8 (22.8-25.2)	183	24.7 (24.2-25.2)
Overall	**4572**		**2041**		**2531**	

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

1.7 Education and employment in adults 16 years and over n=5592

The following table shows how people with cystic fibrosis reported their education and employment status in 2014. Please note that the groups are not mutually exclusive; someone may be a student as well as part-time working, for example.

Patient reported status	Number of patients (%) (n=5592)
Full-time employment	1634 (29.2)
Part-time employment	703 (12.6)
Student	976 (17.5)
Homemaker	258 (4.6)
Disabled	272 (4.9)
Unemployed	821 (14.7)
Retired	85 (1.5)
'Unknown' entered	930 (16.6)
No data recorded	39 (0.7)

Of the 4623 adults aged 16 years and older for whom an employment status questionnaire was completed, 3243 (70.1%) reported being in work or study.

Diagnosis of cystic fibrosis

1.8 Age at diagnosis and screening in children under 16
n=4105

Newborn screening for CF has been done routinely in the whole of the UK since July 2007. It is part of the heel prick blood spot testing done at five to seven 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; n (%)	Patients aged 10 years in 2014; n (%)	Patients aged 5 years in 2014; n (%)
Pre-natal	<5	0 (0)	0 (0)
Birth-3months	2822 (73.5)	135 (60.3)	219 (91.6)
4-6 months	205 (5.4)	17 (7.6)	5 (2.1)
7-12 months	139 (3.7)	11 (4.9)	<5
1 yr	203 (5.4)	22 (9.8)	<5
2 yrs	139 (3.7)	11 (4.9)	<5
3 yrs	76 (2.0)	10 (4.5)	<5
4 yrs	58 (1.5)	8 (3.6)	<5
5 yrs	29 (0.8)	<5	-
6 yrs	26 (0.7)	<5	-
7 yrs	20 (0.5)	<5	-
8 yrs	21 (0.6)	<5	-
9 yrs	11 (0.3)	-	-
10 yrs	7 (0.2)	-	-
11 yrs	<5	-	-
12 yrs	<5	-	-
13 yrs	<5	-	-
14 yrs	<5	-	-
15 yrs	<5	-	-
Overall	**3771**	**224**	**239**

The median (range) age at diagnosis for patients aged under 16 in 2014 is 27 days (0-3 months).

Diagnosis in the first three months of life is more common in children aged five years in 2014 (born after the UK-wide newborn screening programme was in place) than in children aged 10 years in 2014 (born before the UK-wide newborn screening programme was in place).

For the 50 children with complete data born in 2014, 41 (82%) were identified by newborn screening. A total of 130 patients born in 2014 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 results are updated retrospectively each year to take updated data into account. Therefore the number of patients identified in 2013 is higher in this report (177) than was recorded in the 2013 report (127). It is likely that the 2014 figure of 130 will be updated in the next annual report in 2015.

1.9 Age at diagnosis and screening in adults aged 16 and over in 2014

n=5592, 5255 with a diagnosis date

The table below shows the age of people aged 16 and over in 2014 when they were diagnosed. These people were born before newborn screening was done routinely across the UK. There were some regions with newborn screening prior to 2007.

Age at diagnosis	n (%)
Pre-natal	1 (0)
Birth-3 months	2225 (40.2)
4-6 months	508 (9.2)
7-12 months	354 (6.4)
1 yr	460 (8.3)
2 yrs	279 (5.0)
3 yrs	219 (4.0)
4 yrs	173 (3.1)
5 yrs	84 (1.5)
6 yrs	78 (1.4)
7 yrs	53 (1.0)
8 yrs	61 (1.1)
9 yrs	47 (0.9)
10 yrs	41 (0.7)
11 yrs	41 (0.7)
12 yrs	40 (0.7)
13 yrs	45 (0.8)
14 yrs	36 (0.7)
15 yrs	45 (0.8)
16-20 yrs	161 (2.9)
21-25 yrs	108 (2.0)
26-30 yrs	103 (1.9)
31-35 yrs	111 (2.0)
36-40 yrs	87 (1.6)
41-45 yrs	61 (1.1)
46-50 yrs	34 (0.6)
51-60 yrs	43 (0.8)
61 yrs+	33 (0.6)

Lung health

For people with cystic fibrosis mucus in the lungs leads to repeat or chronic infections and inflammation, which can cause permanent damage.

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, 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, 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 more difficult to lead a normal life. If FEV_1 declines to 30% or less, a patient may be considered for lung transplant.

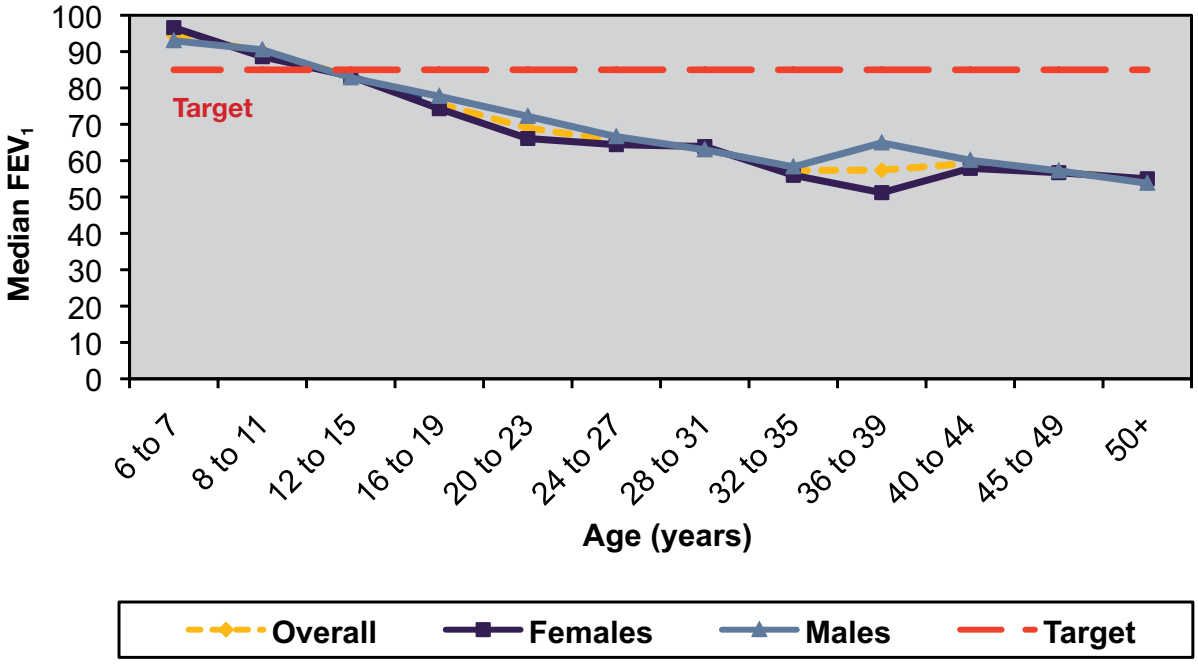
An aim of CF care is to prevent FEV_1 % predicted from falling as much as possible, for as long as possible. This is often a team effort between people with CF, their family, and their medical team, which can include doctors, nurses, physiotherapists, dieticians, and psychologists.

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

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

n=7356

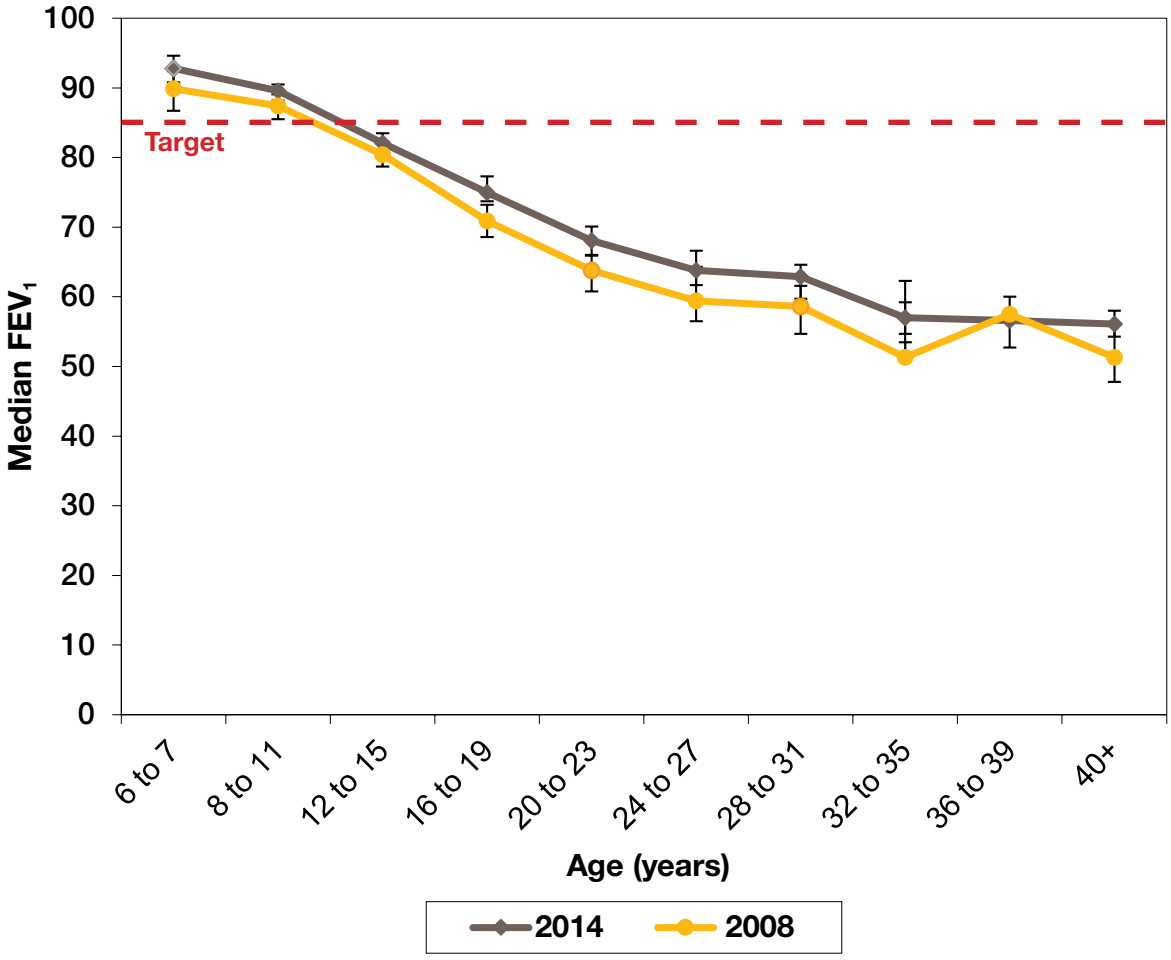
The chart and table in this section show information about those patients whose FEV_1 data were complete. People with CF who have had lung transplants are excluded, as their new 'non-CF' lungs would have lung health similar to a person without CF.



	Overall			Female			Male		
Age (yrs)	N	Median	IQR	N	Median	IQR	N	Median	IQR
6-7	248	94.4	83.3-103.7	123	96.6	82.7-107.5	125	93	84.3-102.6
8-11	904	90.1	78.9-99.9	449	88.6	77.9-98.8	455	90.5	80.3-100.7
12-15	926	83.1	67.8-93.9	468	83.2	67.6-93.7	458	82.8	67.9-94.5
16-19	1006	75.6	59.3-89.8	487	74.3	56.6-88.1	519	77.7	61.4-92.2
20-23	974	69	49.7-86.6	460	66.1	57.9-84.4	514	72.2	50.7-88.3
24-27	898	65.5	45.8-84.2	399	64.4	42.7-83.8	499	66.6	46.4-84.3
28-31	682	63.4	42.9-80.8	301	63.9	44.5-83.3	381	63	40.4-79.2
32-35	549	57.3	40.9-77.1	230	56	40.4-76.6	319	58.3	41.0-77.4
36-39	328	57.4	41.6-79.3	135	51.2	38.5-72.9	193	64.9	45.9-85.0
40-44	322	59.3	40.2-76.7	133	57.9	39.5-74.6	189	60.1	40.5-82-2
45-49	230	57	39.1-79.2	108	56.7	39.7-78.0	122	57.2	38.0-81.4
50+	289	54.8	38.9-79.3	126	55	40.9-75.3	163	53.8	37.4-82
Overall	**7356**	**73.5**	**52.6-90.2**	**3419**	**72.5**	**52.1-89.5**	**3937**	**74.1**	**73.2-75.1**

1.11 Median FEV_1% predicted over time (GLI equations) among patients aged 6 years and older (excluding patients post lung transplant) n=7356 in 2014, n=6082 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 % predicted in 2014 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. 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.



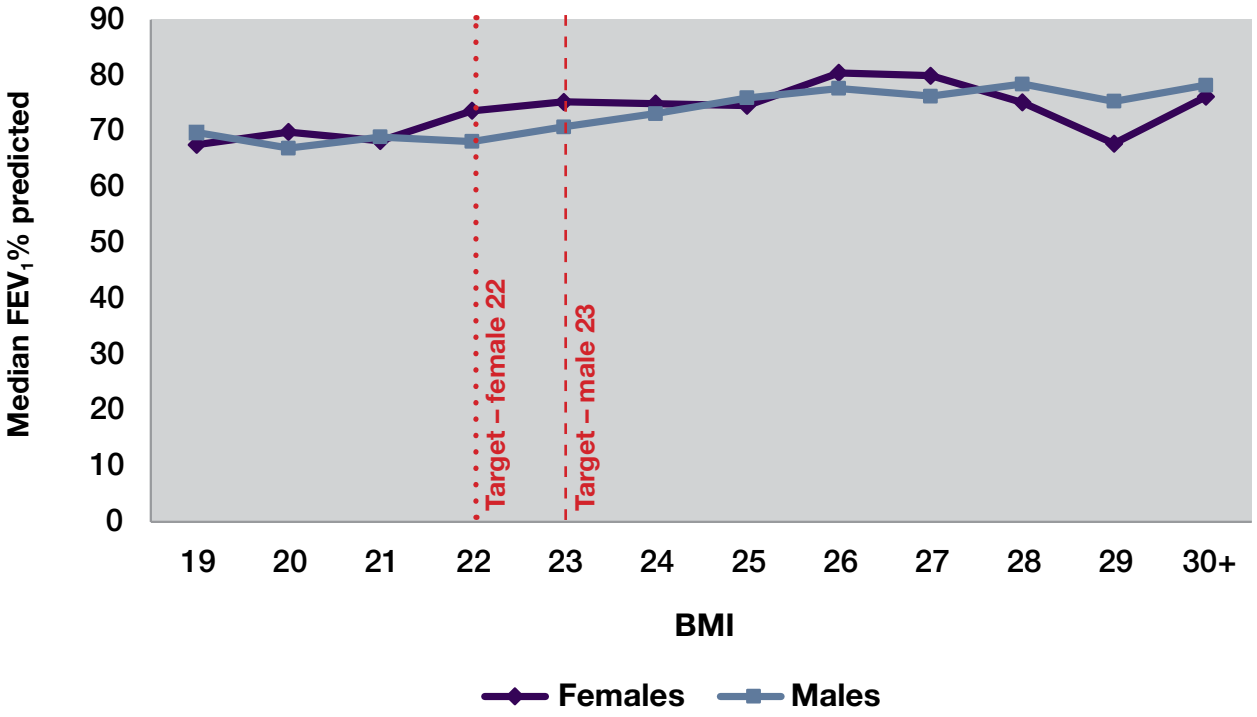
An analysis was conducted in order to determine whether there were statistically significant differences in FEV_1 (% predicted) in 2014 compared to 2008 by age category. The results show that there was a small, but statistically significant difference in the age bands where the p value is less than 0.05.

	Age (years)									
	6-7	8-11	12-15	16-19	20-23	24-27	28-31	32-35	36-39	40+
p-value	0.038	0.016	0.161	0.002	0.002	0.018	0.023	0.015	0.257	0.014

1.12 Median FEV_1% predicted and BMI in patients 16 and older (excluding patients post-lung transplant)

n=7356

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. 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.



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.

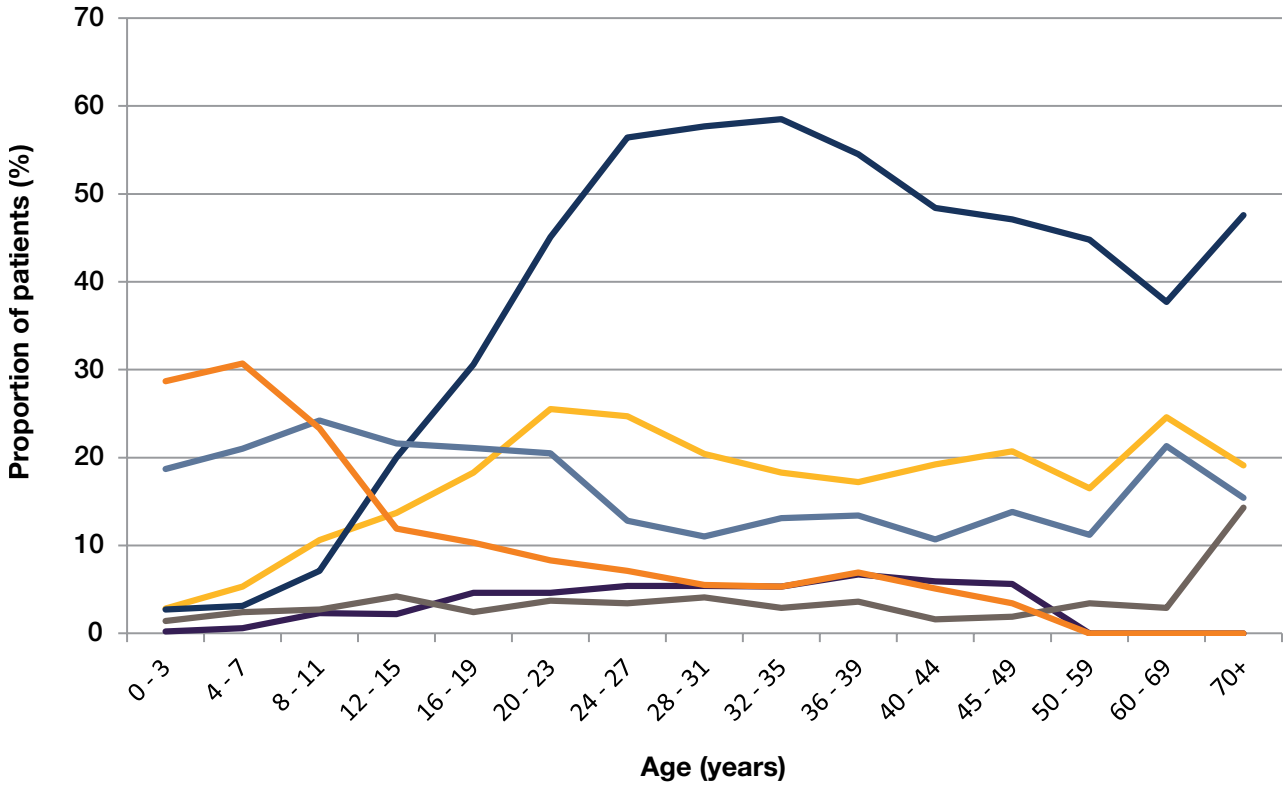
Lung infections

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.

The definition for chronic on the Registry is three or more growths in a year, and is only reported for *Pseudomonas aeruginosa* and *Staphylococcus aureus*. Other bacteria are reported if they grow at all in the year.

1.13 Lung infections in 2014

n=9432



Chronic *Staphylococcus aureus*; n=1460 (15.5%)

Chronic *Pseudomonas aeruginosa*; n=2963 (31.4%)

Intermittent *Pseudomonas aeruginosa*; n=1659 (17.6%)

***Burkholderia cepacia* complex; n=329 (3.5%)**

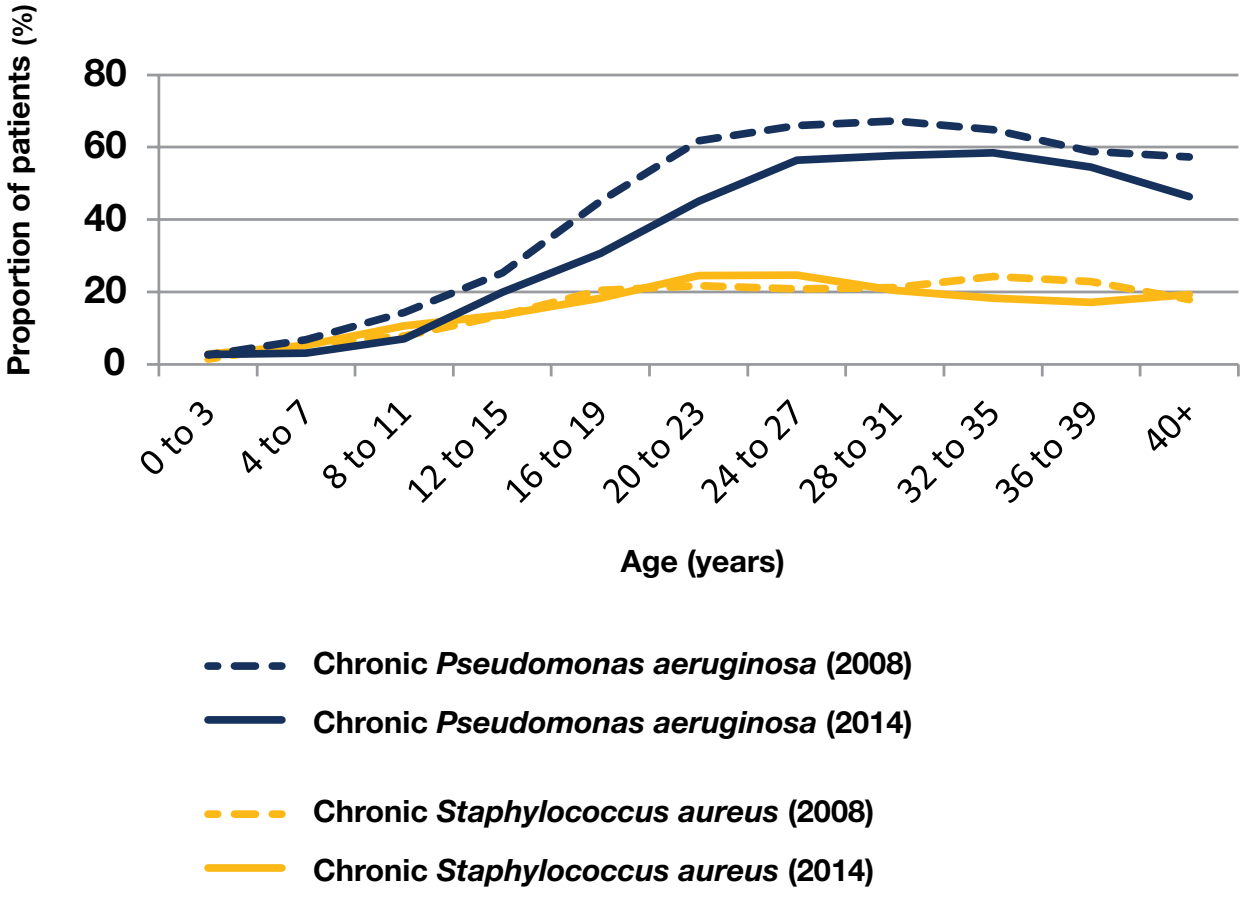
MRSA; n=278 (2.9%)

***Haemophilus Influenza*; n=1310 (13.9%)**

	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	963	1044	906	927	1020	1002	932	728	589	361	372	264	324	3840	5592
Chronic *S. aureus*; n (%)	27 (2.8)	55 (5.3)	94 (10.6)	125 (13.7)	183 (18.3)	252 (25.5)	227 (24.7)	147 (20.4)	106 (18.3)	61 (17.2)	70 (19.2)	54 (20.7)	59 (18.4)	301 (7.9)	1159 (21.0)
Chronic *P. aeruginosa*; n (%)	26 (2.7)	32 (3.1)	64 (7.1)	183 (20.0)	307 (30.6)	448 (45.1)	515 (56.4)	414 (57.7)	339 (58.5)	195 (54.5)	177 (48.4)	123 (47.1)	140 (43.5)	305 (8.0)	2658 (48.2)
Interrmittent *P. aeruginosa*; n (%)	179 (18.7)	218 (21.0)	217 (24.2)	198 (21.6)	212 (21.1)	204 (20.5)	117 (12.8)	79 (11.0)	76 (13.1)	48 (13.4)	39 (10.7)	36 (13.8)	36 (11.2)	812 (21.3)	847 (15.4)
Intermittent *S.aureus*; n(%)	152 (15.9)	184 (17.7)	196 (22.0)	193 (21.1)	191 (19.0)	180 (18.2)	161 (17.5)	107 (14.8)	83 (14.4)	57 (16.1)	43 (11.8)	30 (11.5)	48 (15.0)	725 (19.1)	900 (16.3)
***B. cepacia*; n (%)**	2 (0.2)	6 (0.6)	21 (2.3)	20 (2.2)	47 (4.6)	46 (4.6)	50 (5.4)	39 (5.4)	31 (5.3)	24 (6.7)	22 (5.9)	12 (5.6)	9 (2.8)	49 (1.3)	280 (5.0)
MRSA; n (%)	13 (1.4)	25 (2.4)	24 (2.7)	39 (4.2)	24 (2.4)	37 (3.7)	32 (3.4)	30 (4.1)	17 (2.9)	13 (3.6)	6 (1.6)	5 (1.9)	13 (4.0)	101 (2.6)	177 (3.2)
***H. influenza*; n (%)**	276 (28.7)	320 (30.7)	211 (23.3)	110 (11.9)	105 (10.3)	83 (8.3)	66 (7.1)	40 (5.5)	31 (5.3)	25 (6.9)	19 (5.1)	9 (3.4)	15 (4.6)	917 (23.9)	393 (7.0)

1.14 Lung infections over time

2008 n=6082 and 2014 n=9432



	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	0.638	0.389	0.057	0.000	0.000	0.000	0.000	0.000	0.000	0.000	0.000

An analysis was conducted in order to determine whether there was a statistical difference between the proportion of people with chronic *pseudomonas aeruginosa* in 2014 compared to 2008. The results show that there is a statistically significant difference, after the age of 12.

Complications

1.15 Prevalence of complications

	Overall (n=9432)	<16 years (n=3840)	≥16 years (n=5592)
	N (%)	N (%)	N (%)
Respiratory Related			
Nasal polyps requiring surgery; n (%)	214 (2.3)	41 (1.1)	173 (3.1)
Sinus disease; n (%)	828 (8.8)	36 (0.9)	792 (14.2)
Asthma; n (%)	1368 (14.5)	516 (13.4)	852 (15.2)
ABPA; n (%)	1017(10.8)	298 (7.8)	719 (12.9)
Haemoptysis; n (%)	775 (8.2)	41 (1.1)	734 (13.1)
Pneumothorax requiring chest tube; n (%)	67 (0.7)	3 (0.1)	64 (1.1)
Nontuberculous mycobacteria or atypical mycobacteria; n (%)	433 (4.6)	208 (5.4)	225 (4.0)
Pancreas & Hepatobiliary Disease			
Liver enzymes; n (%)	1071 (11.4)	251 (6.5)	820 (14.7)
Liver disease; n (%)	1322 (14.0)	342 (8.9)	980 (17.5)
Cirrhosis with no portal hypertension; n (%)	126 (1.43	27 (0.7)	99 (1.8)
Cirrhosis with portal hypertension; n (%)	163 (1.7)	20 (0.5)	143 (2.6)
Gall bladder disease requiring surgery; n (%)	36 (0.4)	0 (0.0)	36 (0.6)
Pancreatitis; n (%)	68 (0.7)	4 (0.1)	64 (1.1)
GI bleed req hosp variceal; n (%)	9 (0.1)	2 (0.1)	7 (0.1)
Upper Gastrointestinal			
GORD; n (%)	1494 (15.8)	348 (9.1)	1146 (20.5)
Peptic ulcer; n (%)	6 (0.1)	0 (0.0)	6 (0.1)
GI bleed req hosp non variceal; n (%)	9 (0.1)	2 (0.1)	7 (0.1)
Lower Gastrointestinal			
Intestinal obstruction; n (%)	556 (5.9)	123 (3.2)	433 (7.7)
Fibrosing colonopathy/ colonic structure; n (%)	3 (0.0)	0 (0.0)	0 (0.0)
Rectal prolapse; n (%)	31 (0.3)	24 (0.6)	7 (0.1)
Renal			
Kidney stones; n (%)	85 (0.9)	5 (0.1)	80 (1.4)
Renal failure; n (%)	50 (0.5)	3 (0.1)	47 (0.8)
Muscolo-Skeletal			
Arthritis; n (%)	163 (1.7)	11 (0.3)	152 (2.7)
Arthropathy; n (%)	532 (5.6)	19 (0.5)	513 (9.2)
Bone fracture; n (%)	46 (0.5)	12 (0.3)	34 (0.6)
Osteopenia; n (%)	1225 (13.0)	30 (0.8)	1195 (21.4)

Osteoporosis; n (%)	519 (5.5)	2 (0.5)	517 (9.2)
Other			
Cancer confirmed by histology; n (%)	33 (0.3)	2 (0.1)	31 (0.6)
Port inserted or replaced; n (%)	552 (6.0)	202 (5.3)	350 (6.3)
Absence of vas deferens (males only); n (%)	666 (7.1)	1 (0.3)	665 (12.0)
Depression; n (%)	415 (4.4)	3 (0.1)	412 (7.4)
Hearing loss; n (%)	213 (2.3)	28 (0.7)	185 (3.3)
Hypertension; n (%)	236 (2.5)	2 (0.1)	234 (4.2)

1.16 Incidence of complications

	2013			2014		
	Overall (n=9052)	**<16 years (n=3839)**	**≥16 (n=5213)**	**Overall (n=9432)**	**<16 years (n=3840)**	**≥16 (n=5592)**
Non-tuberculosis mycobacteria or atypical mycobacteria; n (%)	124 (1.3)	71 (1.8)	53 (1.0)	113 (1.2)	52 (1.4)	61 (1.1)
ABPA; n (%)	157 (1.7)	58 (1.5)	99 (1.9)	143 (1.5)	67 (1.7)	76 (1.4)
Cirrhosis - no portal hypertension; n (%)	30 (0.3)	12 (0.3)	18 (0.3)	37 (0.39)	6 (0.16)	31 (0.55)
Cirrhosis - with portal hypertension; n (%)	26 (0.3)	6 (0.2)	20 (0.4)	22 (0.23)	6 (0.16)	16 (0.29)
Cancer confirmed by histology; n (%)	13 (0.1)	1 (0.03)	12 (0.2)	12 (0.13)	0 (0)	12 (0.21)

1.17 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(%)	1924 (27.6)	195 (12.0)	1729 (32.3)
Screening for CF-related diabetes			
Yes	3939 (56.5)	1236 (76.1)	2703 (50.6)
No	1051 (15.1)	189 (11.6)	862 (16.1)
Known CF-related diabetes	1851 (26.6)	156 (9.6)	1695 (31. 7)
Unknown	128 (1.8)	43 (2.7)	85 (1.6)

1.18 Transplants

Lung transplantation has been available to people with cystic fibrosis for almost 30 years. Today the most common operation carried out is called a double lung transplant, or a Bilateral Sequential Lung transplant. Survival is constantly improving, with approximately 85% of patients surviving for at least one year following the operation, and many returning to full time work or education.

The following table shows transplant activity over time.

	2010	2011	2012	2013	2014
Number of patients that year with annual review data evaluated for transplants	169	204	225	220	247
Number accepted on the transplant list	82	121	120	136	146
Number receiving transplants (<16)	3	3	3	3	5
Types of transplants received:					
Bilateral lung	2	3	2	2	2
Heart and lung	0	0	0	0	0
Liver	1	0	1	1	3
Other	0	0	0	0	0
Number receiving transplants (≥16)	26	48*	52**	54*	67**
Types of transplants received:					
Bilateral lung	24	40	43	48	59
Heart and lung	1	4	1	0	0
Liver	0	2	6	3	5
Other	1	3	4	4	5

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

1.19 Ivacaftor

Ivacaftor is a drug that began being prescribed as a treatment for cystic fibrosis in patients aged 6 years and over with at least one copy of the genotype G551D, in June 2012. The table shows ivacaftor use and outcomes from June 2012 – December 2014.

Number of patients on ivacaftor in the UK	**402**
	Median (IQR)
Sweat chloride before ivacaftor	104 (95-114)
Sweat chloride 6-8 weeks after ivacaftor	50 (36-64)
FEV_1% before ivacaftor	61.1 (38.5-69.8)
FEV_1% 6-8 weeks after ivacaftor	71.6 (56.6-82.6)
Number of patients stopped ivacaftor	10

People with CF 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.

1.20 Intravenous (IV) antibiotic use and outcomes

n=(9432)

When someone with cystic fibrosis becomes unwell with an infection, they might be prescribed intravenous (IV) antibiotics, which 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	963	48 (0.5)	10 (8-14)	231 (24.0)	14 (8-19)	233 (24.2)	14 (11-23)
4-7	1044	113 (1.1)	12 (9-25)	302 (29.0)	14 (8-21)	313 (30.0)	14 (14-29)
8-11	906	182 (2.0)	20 (11-32)	308 (34.0)	14 (7-28)	348 (38.4)	27 (14-42)
12-15	927	239 (2.6)	21 (12-32)	446 (48.1)	15 (10-38)	497 (53.6)	28 (14-53)
16-19	1020	296 (3.1)	21 (13-32)	460 (45.1)	17 (10-40)	543 (53.2)	28 (14-56)
20-23	1002	375 (3.7)	22 (13-42)	469 (46.8)	19 (10-41)	596 (59.5)	30 (14-56)
24-27	932	355 (3.8)	24 (14-43)	454 (48.7)	19 (10-41)	567 (60.8)	30 (14-63)
28-31	728	285 (3.9)	21(14-42)	292 (40.1)	17 (8-40)	407 (55.9)	30 (14-56)
32-35	589	233 (4.0)	26 (14-43)	234 (39.7)	15 (9-34)	328 (55.7)	28 (14-56)
36-39	361	136 (3.8)	27 (14-42)	130 (36.0)	16 (9-31)	183 (50.7)	28 (15-55)
40-44	372	116 (3.1)	25 (14-38)	106 (28.5)	14 (8-30)	165 (44.4)	28 (14-50)
45-49	264	76 (2.9)	17 (12-42)	87 (33.0)	19 (9-36)	112 (42.4)	28 (15-48)
50+	324	74 (22.8)	21 (14-36)	108 (33.3)	14 (9-32)	138 (42.6)	24 (14-49)
Overall	**9432**	**2528 (26.8)**	**21 (13-39)**	**3627 (38.5)**	**15 (9-35)**	**4430 (47.0)**	**28 (14-50)**

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.
- It can be difficult to use some inhalers correctly. Using a nebuliser can mean that more of the medication gets into the lung.

1.21 Inhaled antibiotic use among patients with chronic *Pseudomonas aeruginosa*

	2008			2014		
	Overall	<16 years	≥16 years	Overall	<16 years	≥16 years
Patients with chronic *P. aeruginosa*	2098	299	1799	2963	305	2658
Tobramycin solution; n (%)	412 (19.6)	48 (16.1)	364 (20.2)	841 (28.4)	96 (31.5)	745 (28.0)
Other aminoglycoside; n (%)	43 (2.0)	5 (0.2)	38 (2.1)	139 (4.7)	22 (7.2)	117 (4.4)
Colistin; n (%)	914 (43.6)	174 (58.2)	740 (41.1)	1101 (37.2)	156 (51.1)	945 (35.6)
Promixin; n (%)	490 (23.4)	73 (24.4)	417 (23.2	919 (31.0)	134 (43.9)	785 (18.2)
Aztreonam; n (%)				395 (13.3)	10 (3.3)	385 (14.5)
Colistimethate; n (%)				433 (14.6)	21 (6.9)	412 (15.5)
Tobramycin Inhalation Powder; n (%)				802 (27.1)	24 (7.9)	778 (29.3)
At least one of the above*; n (%)	1597 (76.1)	257 (86.0)	1340 (74.5)	2625 (88.6)	288 (94.4)	2337 (87.9)

* In 2014, this includes Aztreonam.

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.

1.22 DNase

	DNase; n (%)				
	2008		2014		
Age	Total patients	Patients on Dnase	Total patients	Patients on Dnase	p-value (2008 vs 2014)
0-3	605	46 (7.6)	963	114 (11.8)	0.007
4-7	621	125 (20.1)	1044	415 (39.8)	<0.001
8-11	663	227 (34.2)	906	558 (61.6)	<0.001
12-15	773	359 (46.4)	927	648 (69.9)	<0.001
16-19	762	377 (49.5)	1020	701 (68.7)	<0.001
20-23	725	319 (44.0)	1002	632 (63.1)	<0.001
24-27	605	288 (47.6)	932	609 (65.3)	<0.001
28-31	419	182 (43.4)	728	447 (61.4)	<0.001
32-35	260	108 (41.5)	589	355 (60.3)	<0.001
36-39	237	83 (35.0)	361	186 (51.5)	0.001
40+	412	147 (35.7)	960	478 (49.8)	<0.001
Overall	**6082**	**2261 (37.2)**	**9432**	**5143 (54.5)**	

1.23 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**	**p-value (2008 vs 2014)**
0-3	605	3 (0.5)	963	64 (6.6)	<0.001
4-7	621	15 (2.4)	1044	173 (16.6)	<0.001
8-11	663	23 (3.5)	906	234 (25.8)	<0.001
12-15	773	32 (4.1)	927	340 (36.7)	<0.001
16-19	762	33 (4.3)	1020	328 (32.2)	<0.001
20-23	725	50 (6.9)	1002	285 (28.4)	<0.001
24-27	605	60 (9.9)	932	269 (28.9)	<0.001
28-31	419	37 (8.8)	728	251 (34.5)	<0.001
32-35	260	29 (11.2)	589	186 (74.1)	<0.001
36-39	237	16 (6.8)	361	96 (26.6)	<0.001
40+	412	33 (8.0)	960	240 (25.0)	<0.001
Overall	**6082**	**331 (5.4)**	**9432**	**2466 (26.1)**	

1.24 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 *P. aeruginosa*.

	2008				2014			
	Overall (n=1958)	**0-3 years (n=15)**	**4-15 years (n=363)**	**≥16 years (n=1580)**	**Overall (n=3705)**	**0-3 years (n=39)**	**4-15 years (n=594)**	**≥16 years (n=3072)**
Patients with chronic *P. aeruginosa*	1246 (63.6)	2 (13.3)	105 (28.9)	1139 (72.1)	1945 (52.5)	5 (12.8)	101 (17.0)	1839 (59.9)
Patients without chronic *P. aeruginosa*	712 (36.4)	13 (86.7)	258 (71.1)	441 (27.9)	1760 (47.5)	34 (87.2)	493 (83.0)	1233 (40.1)

1.25 Physiotherapy

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

	Overall (n=9432)	<16 years (n=3840)	≥16 years (n=5592)
Active cycle of breathing techniques; n (%)	3033 (32.5)	1720 (45.4)	1313 (23.7)
Autogenic drainage (including assited autogenic drainage); n (%)	1437 (15.4)	248 (6.54)	1189 (21.5)
Any form of PEP; n (%)	5135 (55.1)	2568 (67.8)	2567 (46.4)
VEST; n (%)	174 (1.9)	88 (2.3)	86 (1.6)
Exercise; n (%)	3229 (34.6)	1479 (39.0)	1750 (31.6)

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

1.26 Other therapy

	Overall (n=9432)	<16 years (n=3840)	≥16 years (n=5592)
NIV; n (%)	283 (3.0)	39 (1.0)	244 (4.36)
Long-term oxygen; n (%) Among those who have long-term oxygen:	653 (6.9)	94 (2.4)	559 (10.0)
Continuously	157 (1.7)	9 (0.2)	148 (2.7)
Nocturnal or with exertion	152 (1.6)	18 (0.5)	134 (2.4)
Pro re nata (PRN)	79 (0.8)	13 (0.3)	66 (1.2)
With exacerbation	265 (2.8)	54 (1.4)	211 (3.8)

1.27 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=9432)	<16 years (n=3840)	≥16 years (5592)
Any supplmental feeding; n (%)	3214 (34.1)	857 (22.3)	2136 (38.2)
Nasogastric tube	114 (1.2)	12 (0.3)	102 (1.3)
Gastrostomy tube/Button	572 (6.1)	221 (5.8)	351 (6.3)
Jejunal	6 (0.1)	0	6
TPN	2	1	1

1.28 Age distribution of deaths in 2014

The table below shows the ages of the 137 people with cystic fibrosis who died in 2014.

Age at death	Number of cystic fibrosis patients
0-3	2
4-7	1
8-11	3
12-15	0
16-19	11
20-23	25
24-27	22
28-31	13
32-35	22
36-39	9
40-43	4
44-47	4
48-51	8
52-55	5
56+	8
Total	**137**

1.29 Genotypes

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

9213 (97.7%) patients have been genotyped with a recorded value on at least one allele.

DF508 Mutations; n (%)

Homozygous DF508 4663 (50.6%)

Heterozygous DF508 3664 (39.8%)

No DF508 or both unidentified 886 (9.6%)

Mutations				
Nucleotide	**Protein**	**Legacy name**	**N**	**(%)**
c.1521_1523delCTT	p.Phe508del	^F508	8,327	90.38
other	other	Other	1528	16.59
not identified	not identified	Not Identified	555	6.02
c.1652G>A	p.Gly551Asp	G551D	525	5.70
c.350G>A	p.Arg117His	R117H	411	4.46
c.1624G>T	p.Gly542X	G542X	325	3.55
c.489+1G>T	no protein name	621+1G->T	207	2.25
c.3909C>G	p.Asn1303Lys	N1303K	125	1.36
c.1585-1G>A	no protein name	1717-1G->A	118	1.28
c.1766+1G>A	no protein name	1898+1G->A	110	1.19
c.1519_1521delATC	p.Ile507del	^I507	91	0.93
c.3528delC	p.Lys1177SerfsX15	3659delC	87	0.94
c.1679G>C	p.Arg560Thr	R560T	83	0.90
c.1657C>T	p.Arg553X	R553X	74	0.80
c.3717+12191C>T	no protein name	3849+10kbC->T	74	0.80
c.254G>A	p.Gly85Glu	G85E	70	0.76
c.1477C>T	p.Gln493X	Q493X	70	0.76
c.3454G>C	p.Asp1152His	D1152H	67	0.73
c.178G>T	p.Glu60X	E60X	61	0.66
c.3846G>A	p.Trp1282X	W1282X	50	0.54
c.2052delA	p.Lys684AsnfsX38	2184delA	34	0.37
c.2657+5G>A	no protein name	2789+5G->A	33	0.36
c.1040G>C	p.Arg347Pro	R347P	30	0.33
c.1646G>A	p.Ser549Asn	S549N	29	0.31
c.1558G>T	p.Val520Phe	V520F	28	0.30
c.3484C>T	p.Arg1162X	R1162X	26	0.28
c.1364C>A	p.Ala455Glu	A455E	25	0.27
c.1000C>T	p.Arg334Trp	R334W	16	0.17
c.1040G>A	p.Arg347His	R347H	16	0.17

Mutations				
Nucleotide	**Protein**	**Legacy name**	**N**	**(%)**
c.3472C>T	p.Arg1158X	R1158X	14	0.15
c.1055G>A	p.Arg352Gln	R352Q	14	0.15
c.2988+1G>A	no protein name	3120+1G->A	12	0.11
c.532G>A	p.Gly178Arg	G178R	9	0.09
c.579+1G>T	no protein name	711+1G->T	9	0.10
c.2051_2052delAAinsG	p.Lys684SerfsX38	2183delAA->G	7	0.08
		R1283M	7	0.08
c.1645A>C or c.1647T>G	p.Ser549Arg	S549R	6	0.07
c.1675G>A	p.Ala559Thr	A559T	5	0.05
c.443T>C	p.Ile148Thr	I148T	5	0.05
c.2012delT	p.Leu671X	2143delT	<5	-
		C524X	<5	-
c.2128A>T	p.Lys710X	K710X	<5	-
c.1654C>T	p.Gln552X	Q552X	<5	-

Section 2: 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 shows analysis of data for individual cystic fibrosis 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.

It is important to remember that lots of different factors can affect the outcomes of people with CF in centres, not all of which are within the 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 48.

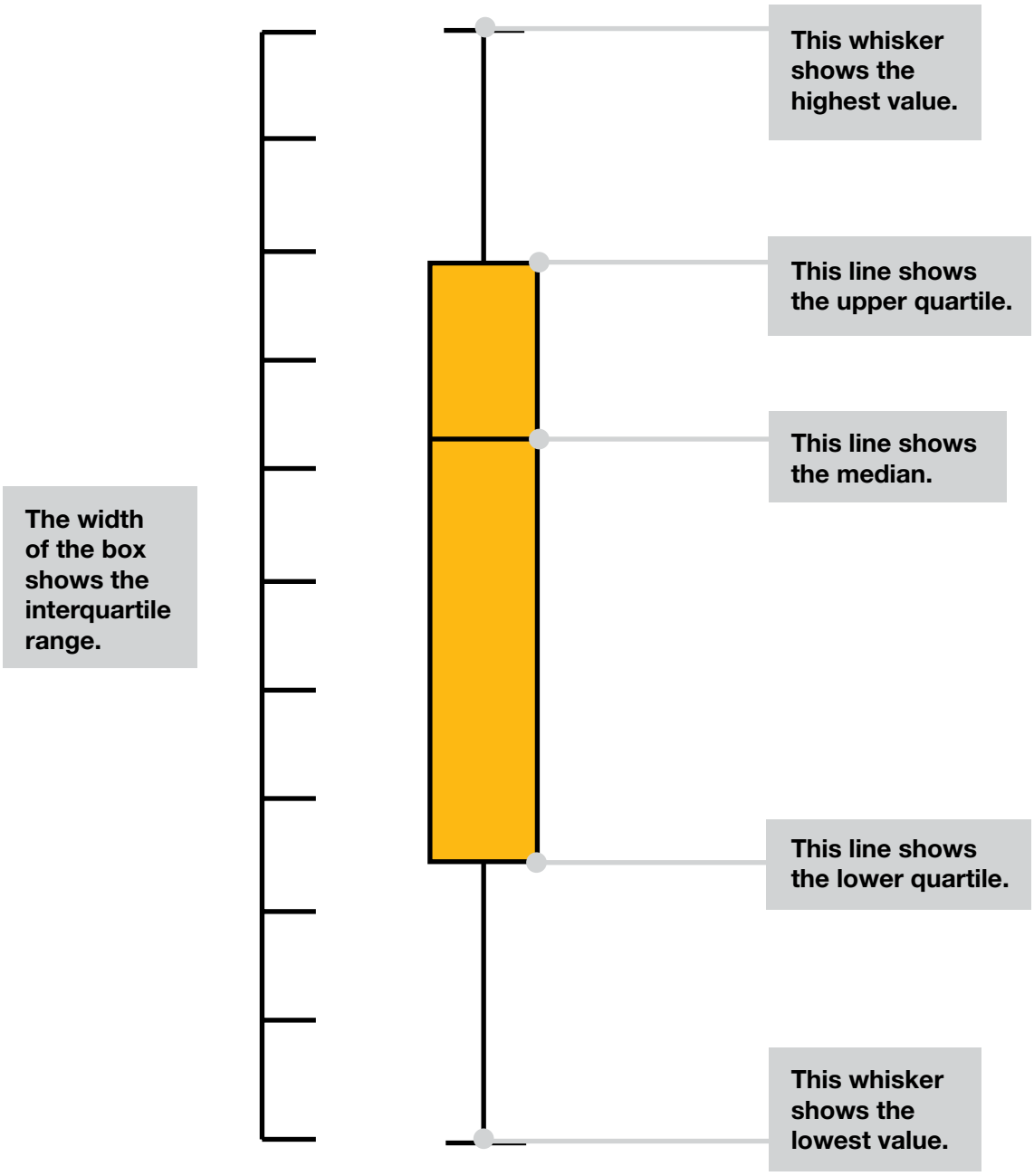
Key

 Paediatric centre

 Adult centre

A guide to the charts

Some of the charts in this section are shown as ‘box plots’. Box plots are made up of a box with a median within it, ‘whiskers’ either side with an upper adjacent.

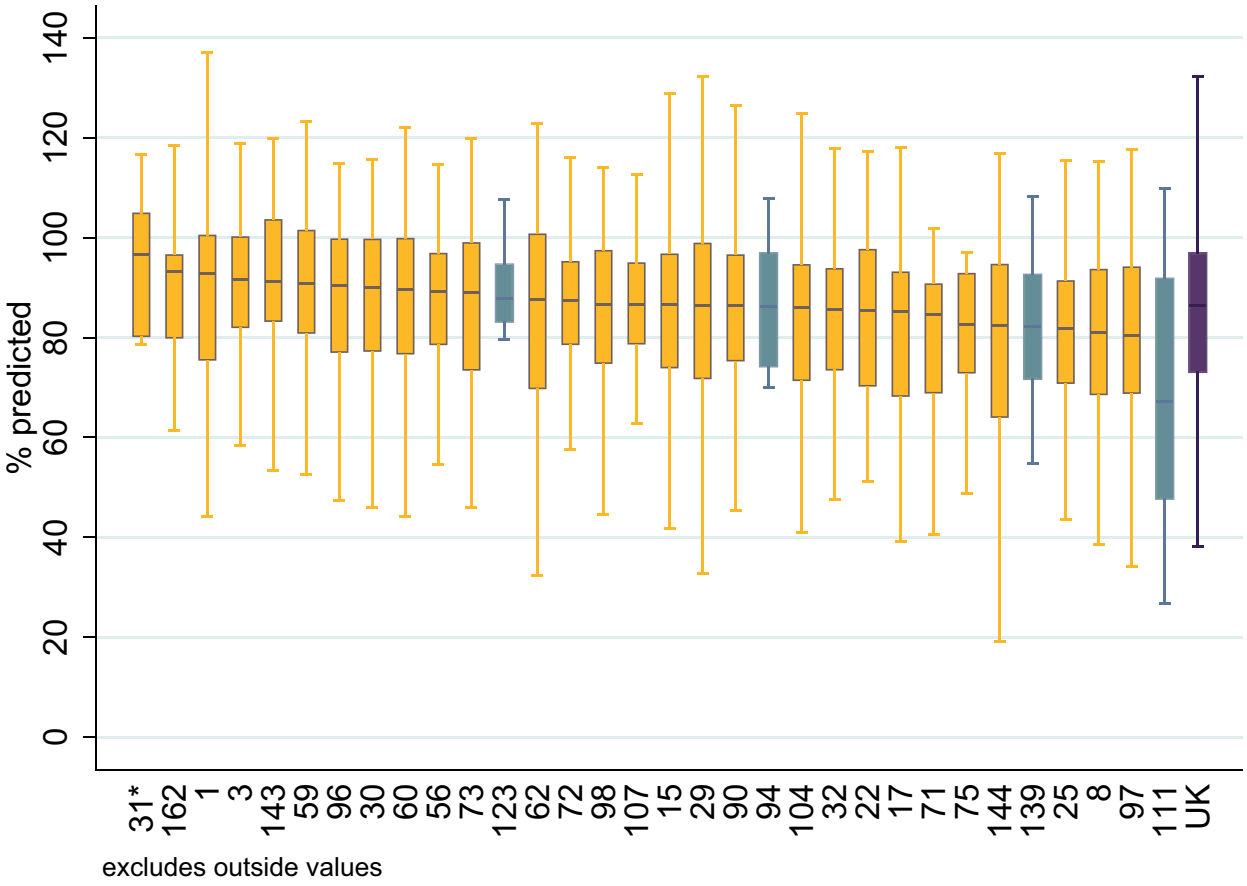


- The **‘box’** shows the middle half of the data for that centre, going from the first quartile to the third quartile. The longer the box, the more varied the data for that centre.
- The horizontal line within the box shows the **median** result for that centre.
- The **‘whiskers’** above and below 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.

Section 2a Paediatric centre analysis n=4177

This section shows results for the 28 paediatric centres with their network clinics, and four stand-alone clinics.

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



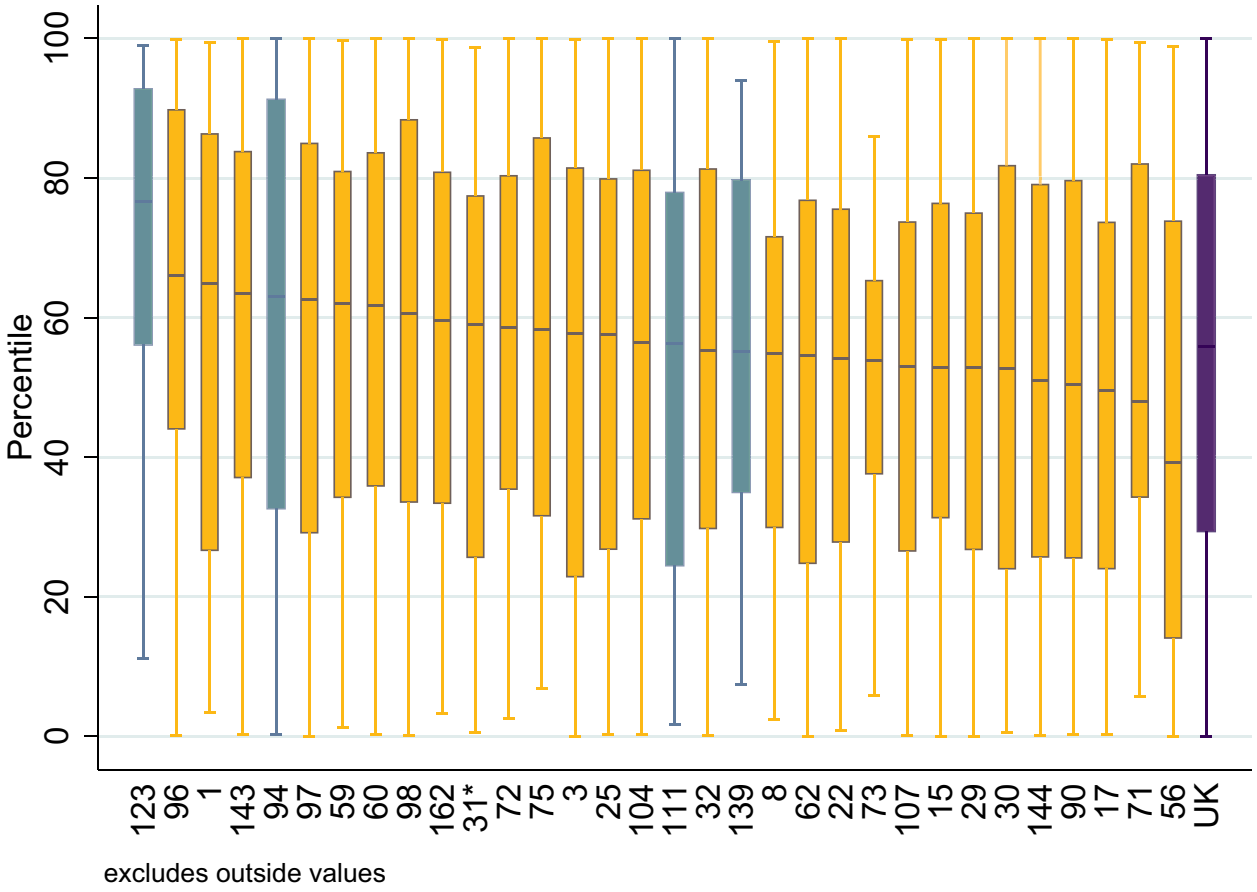
The median FEV_1 % predicted for patients attending paediatric centres/clinics is 86% predicted (IQR: 73–97).
* Centre/clinic with a data set submission of fewer than 20 patients.

Yellow: centres with their network clinics.

Blue: stand-alone clinics.

Purple: all.

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



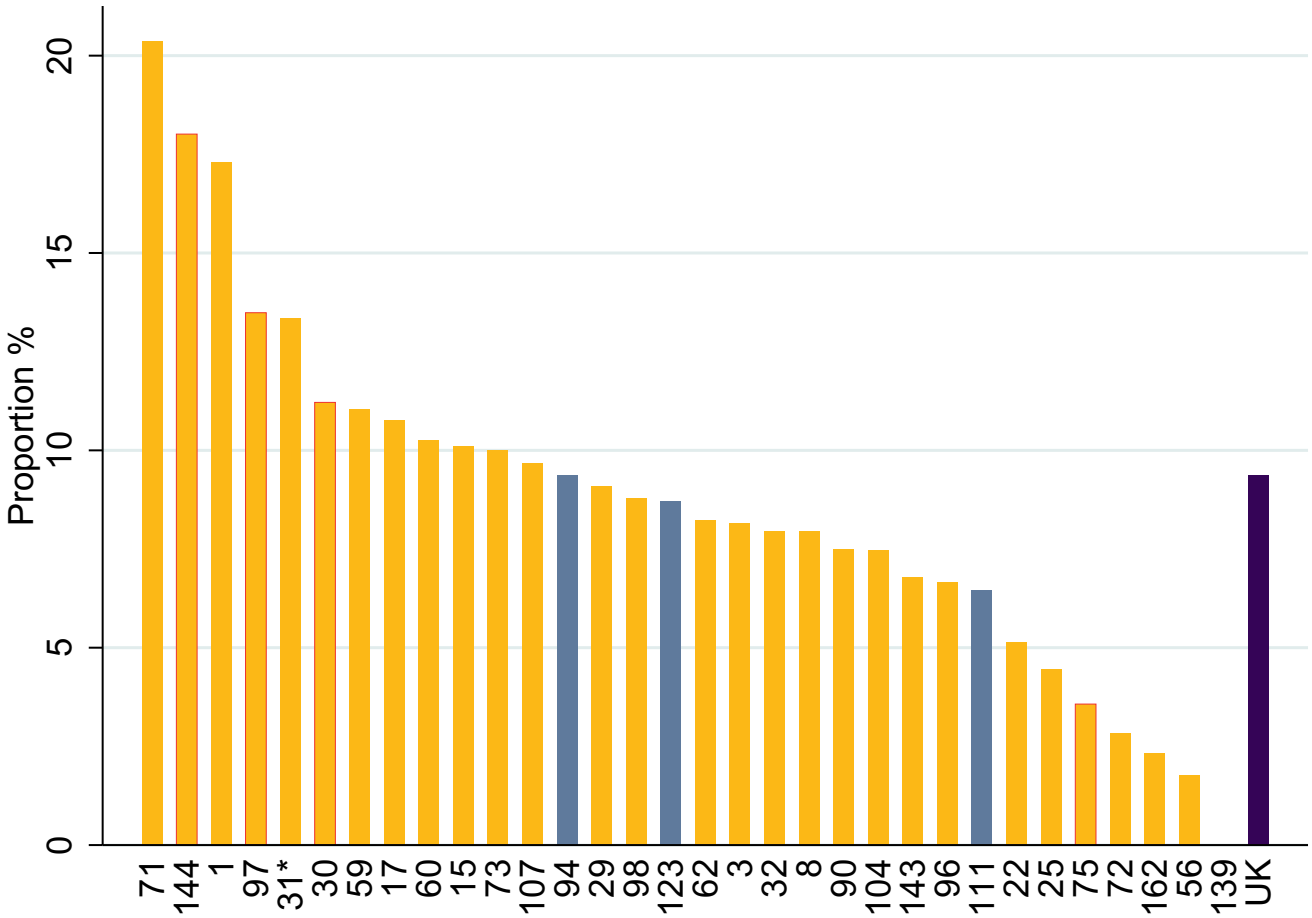
The median BMI percentile for patients attending paediatric centres/clinics is 55 (IQR: 29–77).
*Centre/clinic with a data set submission of fewer than 20 patients.

Yellow: centres with their network clinics.

Blue: stand-alone clinics.

Purple: all.

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



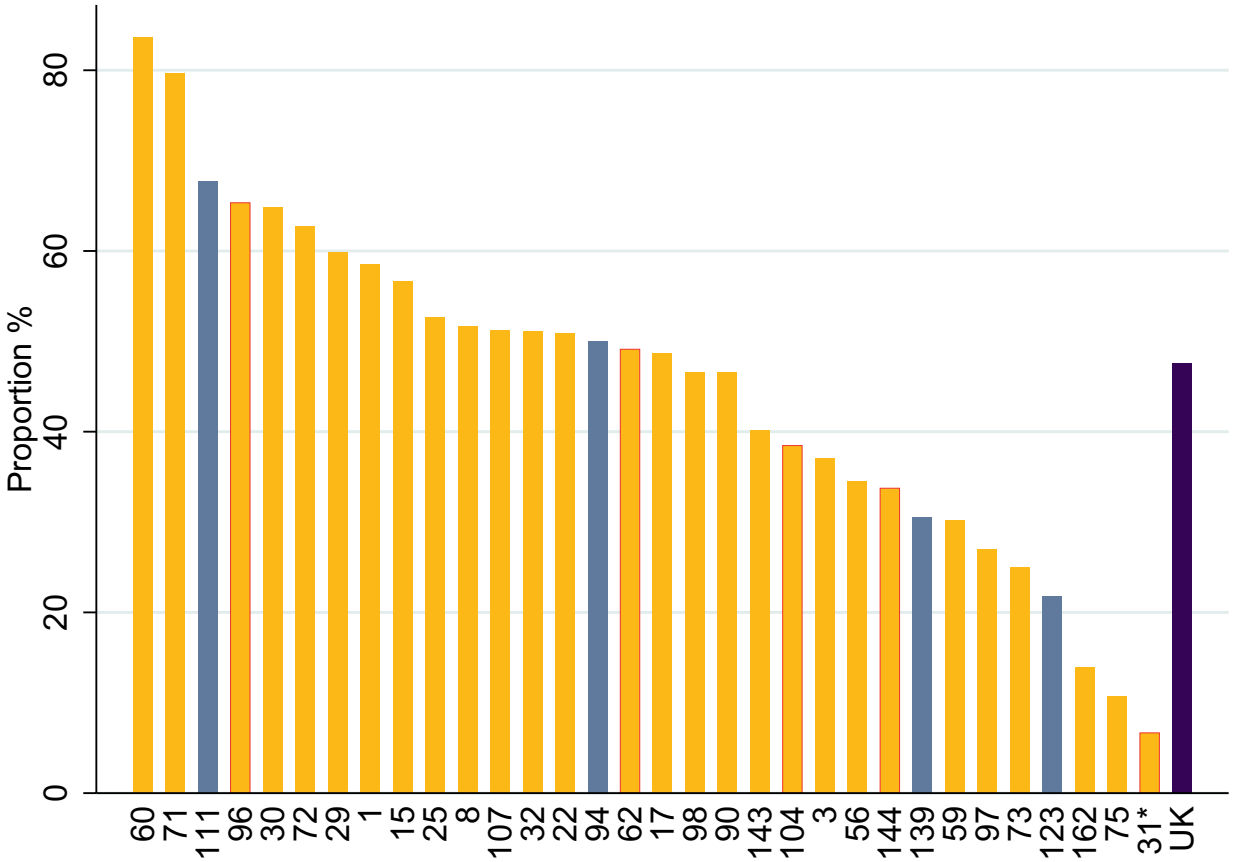
The proportion of patients with chronic *P. aeruginosa* in paediatric centres/clinics is 9%.
* Centre/clinic with a data set submission of fewer than 20 patients.

Yellow: centres with their network clinics.

Blue: stand-alone clinics.

Purple: all.

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



The proportion of patients receiving DNase treatment in paediatric centres/clinics is 48%.
* Centre/clinic with a data set submission of fewer than 20 patients.

Yellow: centres with their network clinics.

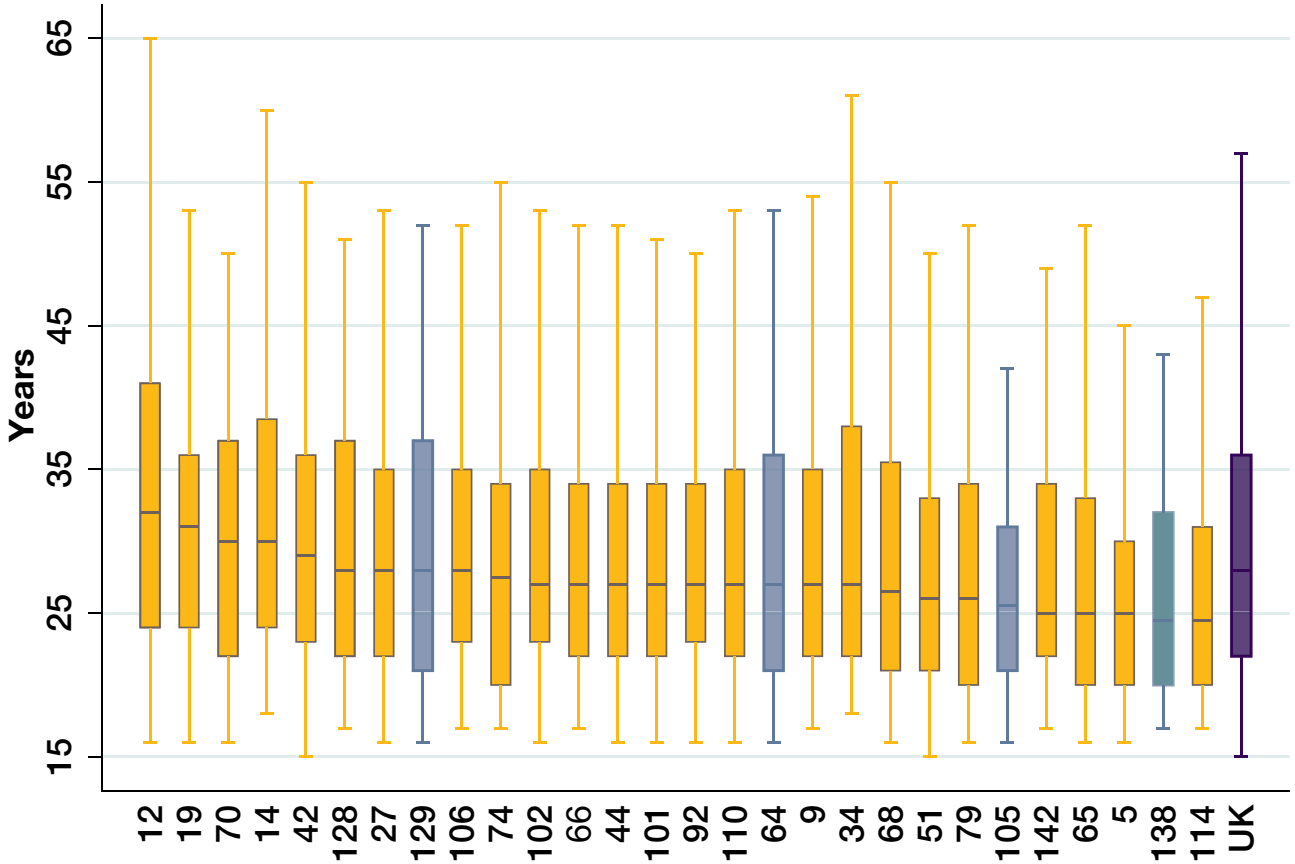
Blue: stand-alone clinics.

Purple: all.

Section 2b: Adult centre analyses n=5255

This section shows results for the 24 adult specialist cystic fibrosis centres. People with CF transfer to adult care centres between the ages of 16 and 18 years.

2.5 Median age (years) by adult service



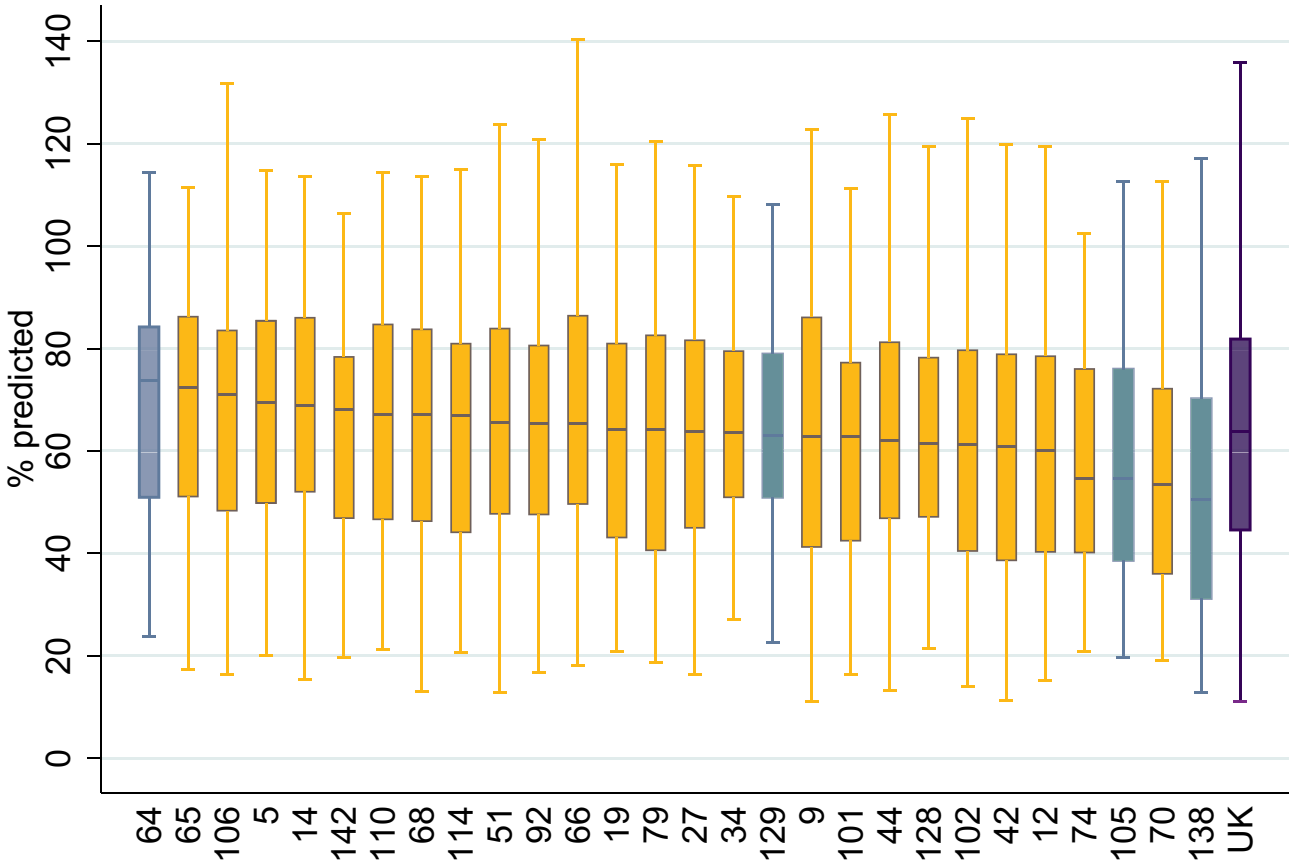
The median age in adult services is 28 years (IQR: 25–36).

Yellow: centres with their network clinics.

Blue: stand-alone clinics.

Purple: all.

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



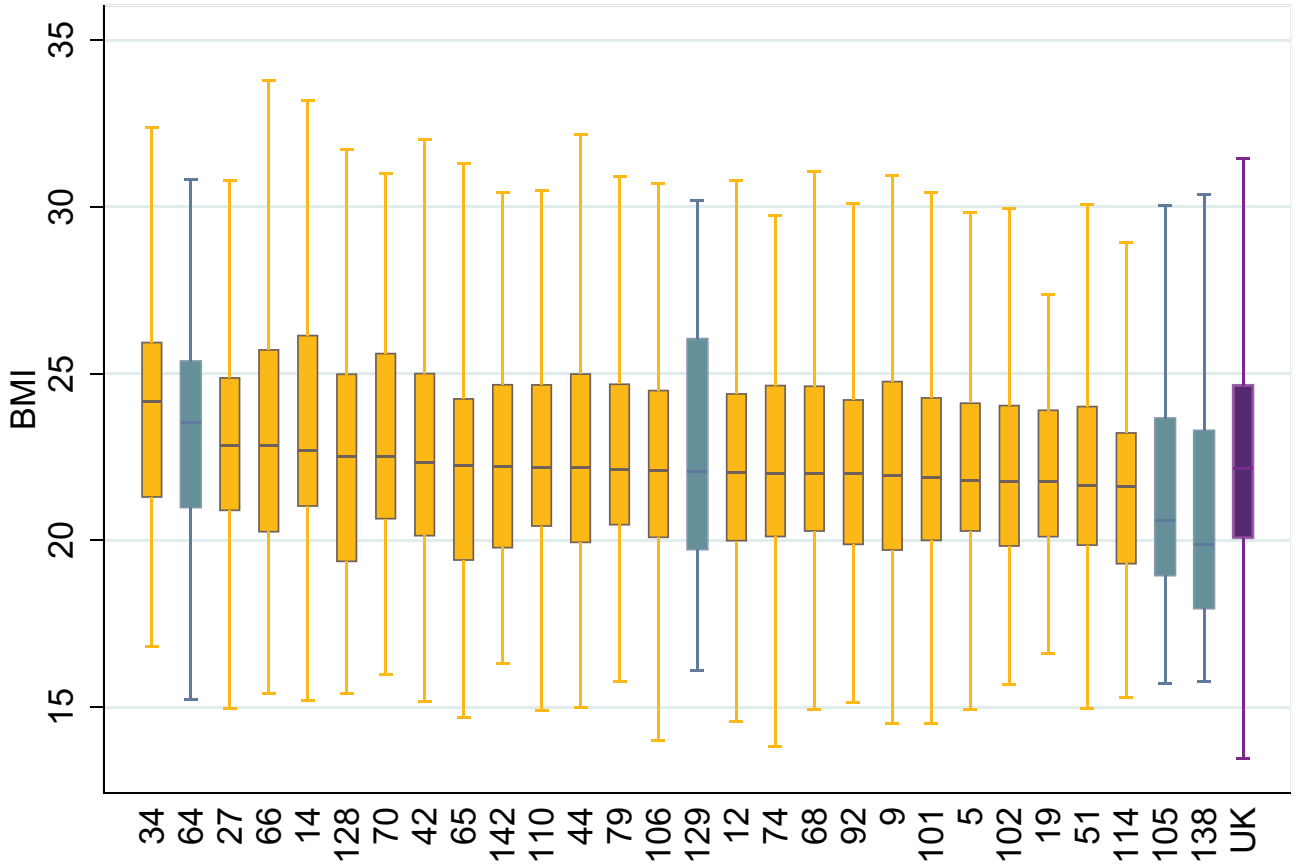
The median FEV_1 (% predicted) in adult services is 64% (IQR: 45–82).

Yellow: centres with their network clinics.

Blue: stand-alone clinics.

Purple: all.

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



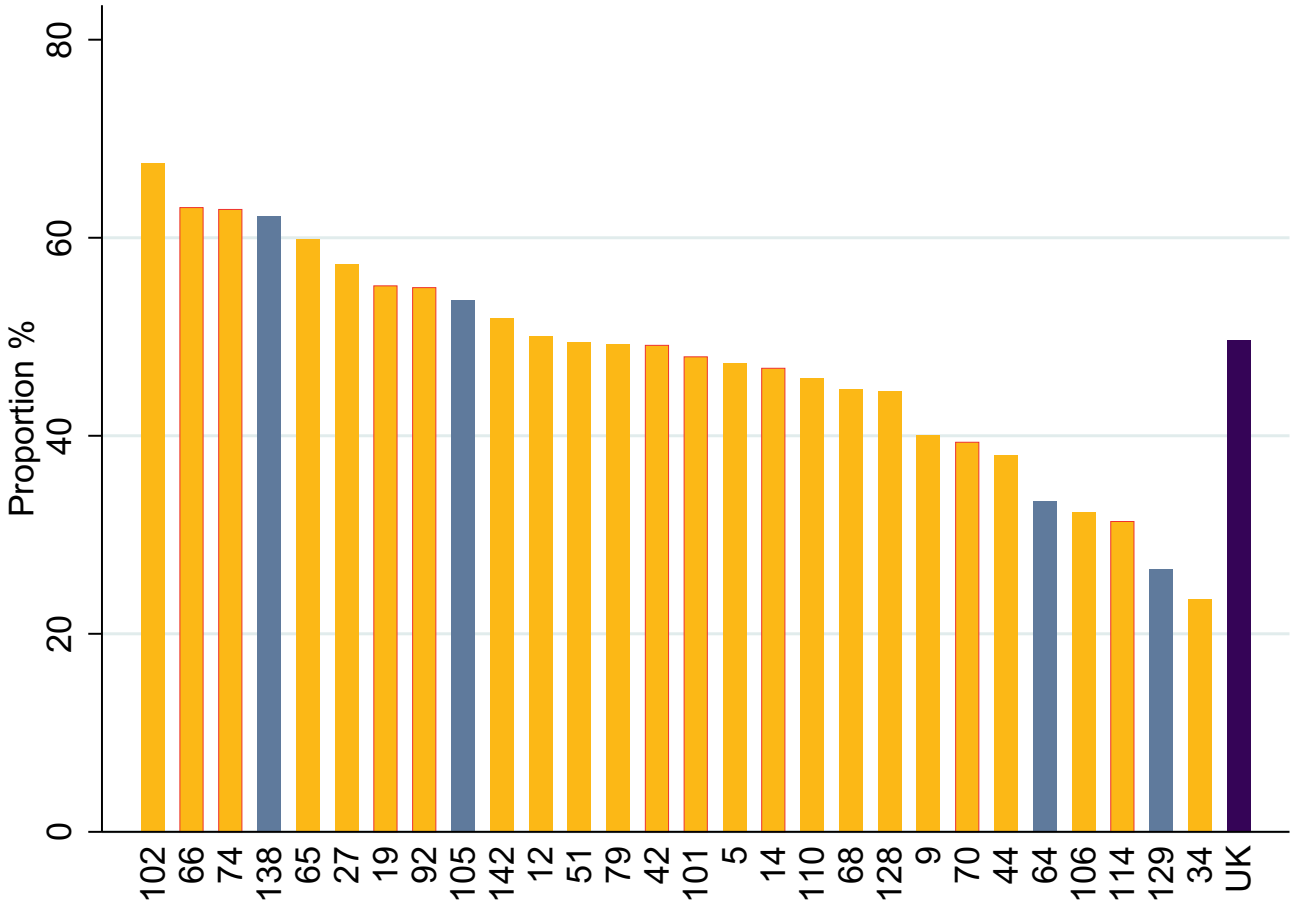
The median BMI in adult services is 22 (IQR: 20–25).

Yellow: centres with their network clinics.

Blue: stand-alone clinics.

Purple: all.

2.8 Proportion of patients with chronic *P. aeruginosa* by adult service



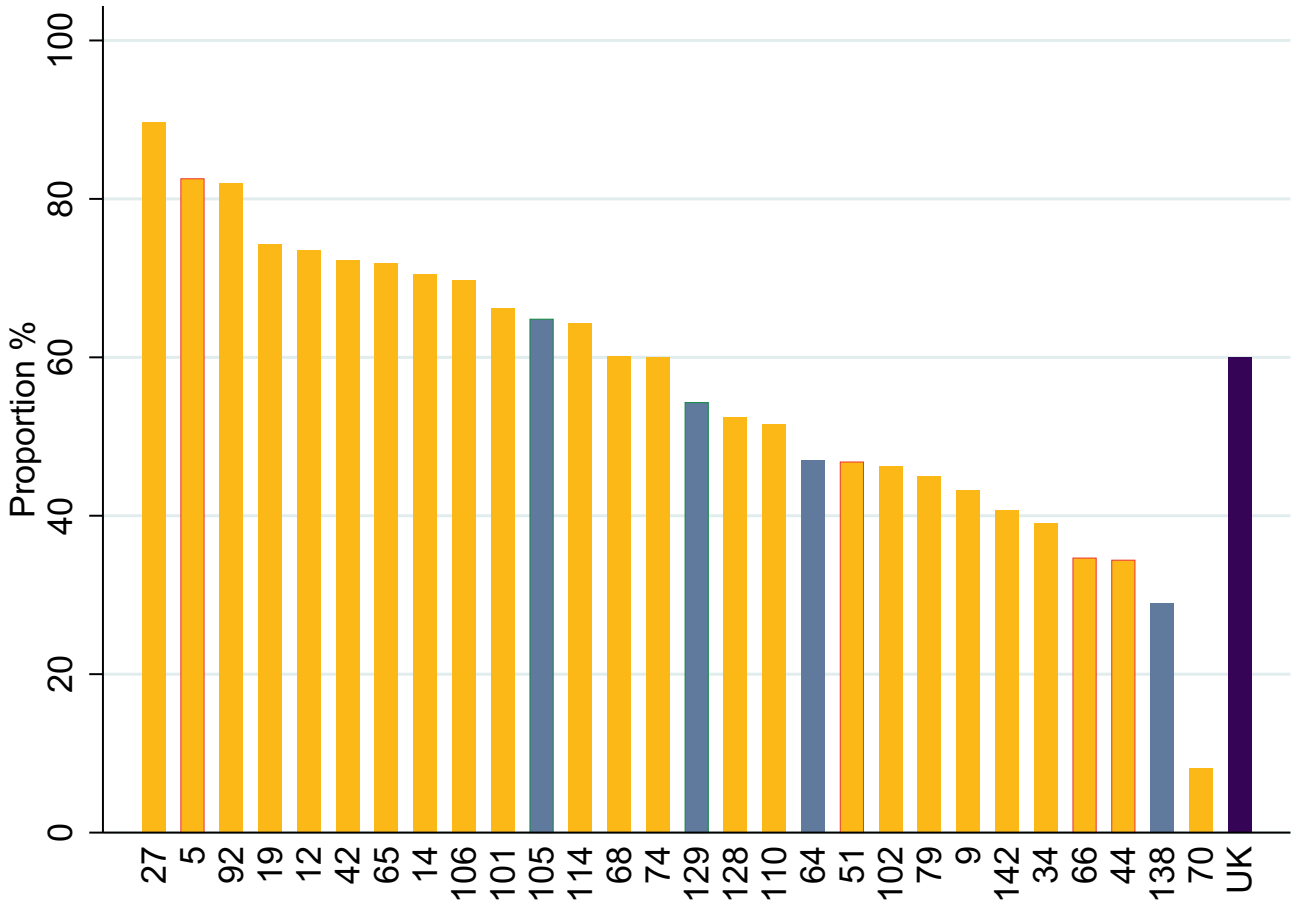
The proportion of patients with chronic *P. aeruginosa* in adult centres/clinics is 50%.

Yellow: centres with their network clinics.

Blue: stand-alone clinics.

Purple: all.

2.9 Proportion of patients receiving DNase treatment by adult service



The proportion of patients receiving DNase treatment in adult centres/clinics is 60%.

Yellow: centres with their network clinics.

Blue: stand-alone clinics.

Purple: all.

Appendix 1: Centre level data tables

Paediatric centres/clinics providing data in 2014 – ordered by clinic ID

Country	Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2014	Median FEV_1% predicted (≥6 years)	Median BMI percentile (2-15 years)
England	Leicester	Leicester Royal Infirmary	1	59	53	94.7	62.3
England	Sheffield	Sheffield Children's Hospital	3	144	135	92.8	50.6
England	North West Staffs	North West Staffs, Stoke on Trent	8	95	89	81.5	54.1
England	London - South West	Royal Brompton Hospital	15	322	318	88.2	51.6
England	London - South East	King's College Hospital	17	193	189	87.5	50.7
England	Oxford	John Radcliff Hospital	22	178	175	86.3	56.1
England	Leeds	St James's University Hospital	25	234	226	83.7	57.6
England	Southampton	Southampton General Hospital	29	208	202	88.9	53
England	London - East	Royal London Hospital	30	112	108	87.2	50.1
Scotland	Iverness	Raigmore Hospital	31	17	15	101.6	55
England	Bristol	Bristol Royal Hospital for Children	32	178	176	86.8	50.2
Scotland	Glasgow	Royal Hospital for Sick Children	56	86	58	89.4	30.5
England	Newcastle	Royal Victoria Infirmary	59	174	172	93.3	62.6
Northern Ireland	Belfast	Royal Belfast Hospital for Sick Children	60	196	195	93.2	60.6
England	Nottingham	Nottingham University Hospitals	62	175	171	88	55.2
England	Teeside	James Cook University Hospital	71	57	54	86.2	40.7
Wales	Cardiff	Children's Hospital for Wales	72	158	145	90.1	57.3

Country	Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2014	Median FEV1% predicted (≥6 years)	Median BMI percentile (2-15 years)
Scotland	Dundee	Ninewells Hospital	73	22	20	90.26	55.8
Scotland	Aberdeen	Royal Aberdeen Children's Hospital	75	29	28	80.4	50.4
England	London-Central	Great Ormond Street Hospital for Children	90	187	187	86	47.9
England	Cornwall	Royal Cornwall Hospital	94	32	32	91.6	73.2
England	Exeter	Royal Devon & Exeter Hospital	96	75	75	91	62.7
England	Liverpool	Alder Hey Children's Hospital	97	307	304	82.4	59.4
England	Norwich	Norfolk & Norwich University Hospital	98	58	58	89.4	69.9
England	Birmingham	Birmingham Children's Hospital	104	297	286	85.6	55.3
England	Cambridge	Addenbrookes Hospital	107	127	125	86.7	52.2
England	Hull	Hull Royal Infirmary	111	32	31	66.9	63.2
Scotland	Ayr/ Kilmarnock	Crosshouse Hospital	123	24	23	92.5	72.5
England	Plymouth	Derriford Hospital	139	37	36	80.7	52
Scotland	Edinburgh	Royal Hospital for Sick Children	143	124	122	92.2	63.9
England	Manchester	Royal Manchester Children's Hospital	144	340	326	84.3	53.6
Scotland	Lanarkshire	Wishaw General Hospital	162	45	43	90.8	50.6

Adult centres/clinics providing data in 2014 – ordered by clinic ID

Country	Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2014	Median FEV_1% predicted (≥16 years)	Median BMI percentile (≥16 years)
England	London-South East	King's College Hospital	5	197	189	71.4	21.8
England	Newcastle	Royal Victoria Infirmary	9	263	259	65	22
England	London-South West	Royal Brompton Hospital	12	662	643	63.7	22
Northern Ireland	Belfast	Belfast City Hospital	14	233	220	71.5	22.7
England	Frimley	Frimley Park Hospital	19	112	109	70	21.8
England	Birmingham	Birmingham Heartlands Hospital	27	352	341	65.6	22.9
England	Exeter	Royal Devon & Exeter	34	89	87	66.2	24.2
England	Leeds	St James's University Hospital	42	411	404	64.1	22.3
Scotland	Edinburgh	Western General Hospital	44	226	221	66.6	22.2
England	Cambridge	Papworth Hospital	51	290	280	68.6	21.6
England	Plymouth	Derriford Hospital	64	52	51	76.7	23.5
England	Sheffield	Northern General Hospital	65	175	174	73.7	22.3
England	Liverpool	Liverpool Heart and Chest Hospital	66	284	277	67.9	22.8
Wales	Llandough	Llandough Hospital	68	216	216	70.4	22
Scotland	Aberdeen	Aberdeen Royal Infirmary	70	66	61	57.4	22.5
England	North West Staff	North West Staffs, Stoke on Trent	74	74	70	57.9	22
Scotland	Glasgow	Gartnavel General Hospital	79	213	191	67.2	22.1
England	London-East	St. Bartholomew's Hospital	92	138	133	67.8	22

Country	Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2014	Median FEV_1% predicted (≥16 years)	Median BMI percentile (≥16 years)
England	Nottingham	Nottingham University Hospitals	101	154	151	66.3	21.9
England	Manchester	Wythenshawe Hospital	102	394	389	63.5	21.8
England	London-South East	University Hospital Lewisham	105	54	54	60.6	20.6
England	Bristol	Bristol Royal Infirmary	106	195	195	72.7	22.1
England	Southampton	Southampton General Hospital	110	227	215	69.2	22.2
England	Norwich	Norfolk & Norwich University Hospital	114	73	70	69.8	21.6
England	Oxford	Churchill Hospital	128	103	101	65.5	22.5
England	Cornwall	Royal Cornwall Hospital	129	35	35	67.1	22
England	Hull	Castle Hill Hospital	138	40	38	54	19.9
England	Leicester	Glenfield Hospital	142	81	81	70.3	22.2

Paediatric centres/clinics providing data in 2014 – ordered alphabetically by city

Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2014	Median FEV_1% predicted (≥6 years)	Median BMI percentile (2-15 years)
England						
Birmingham	Birmingham Children's Hospital	104	297	286	85.6	55.3
Bristol	Bristol Royal Hospital for Children	32	178	176	86.8	50.2
Cambridge	Addenbrookes Hospital	107	127	125	86.7	52.2
Cornwall	Royal Cornwall Hospital	94	32	32	91.6	73.2
Exeter	Royal Devon & Exeter Hospital	96	75	75	91	62.7
Hull	Hull Royal Infirmary	111	32	31	66.9	63.2
Leeds	St James's University Hospital	25	234	226	83.7	57.6
Leicester	Leicester Royal Infirmary	1	59	53	94.7	62.3
Liverpool	Alder Hey Children's Hospital	97	307	304	82.4	59.4
London - East	Royal London Hospital	30	112	108	87.2	50.1
London - South East	King's College Hospital	17	193	189	87.5	50.7
London - South West	Royal Brompton Hospital	15	322	318	88.2	51.6
London-Central	Great Ormond Street Hospital for Children	90	187	187	86	47.9
Manchester	Royal Manchester Children's Hospital	144	340	326	84.3	53.6
Newcastle	Royal Victoria Infirmary	59	174	172	93.3	62.6
North West Staffs	North West Staffs, Stoke on Trent	8	95	89	81.5	54.1
Norwich	Norfolk & Norwich University Hospital	98	58	58	89.4	69.9

Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2014	Median FEV_1% predicted (≥6 years)	Median BMI percentile (2-15 years)
Nottingham	Nottingham University Hospitals	62	175	171	88	55.2
Oxford	John Radcliff Hospital	22	178	175	86.3	56.1
Plymouth	Derriford Hospital	139	37	36	80.7	52
Sheffield	Sheffield Children's Hospital	3	144	135	92.8	50.6
Southampton	Southampton General Hospital	29	208	202	88.9	53
Teeside	James Cook University Hospital	71	57	54	86.2	40.7
Northern Ireland						
Belfast	Royal Belfast Hospital for Sick Children	60	196	195	93.2	60.6
Scotland						
Aberdeen	Royal Aberdeen Children's Hospital	75	29	28	80.4	50.4
Ayr/ Kilmarnock	Crosshouse Hospital	123	24	23	92.5	72.5
Dundee	Ninewells Hospital	73	22	20	90.26	55.8
Edinburgh	Royal Hospital for Sick Children	143	124	122	92.2	63.9
Glasgow	Royal Hospital for Sick Children	56	86	58	89.4	30.5
Iverness	Raigmore Hospital	31	17	15	101.6	55
Lanarkshire	Wishaw General Hospital	162	45	43	90.8	50.6
Wales						
Cardiff	Children's Hospital for Wales	72	158	145	90.1	57.3

Adult centres/clinics providing data in 2014 – ordered alphabetically by city

Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2014	Median FEV_1% predicted (≥16 years)	Median BMI percentile (≥16 years)
England						
Birmingham	Birmingham Heartlands Hospital	27	352	341	65.6	22.9
Bristol	Bristol Royal Infirmary	106	195	195	72.7	22.1
Cambridge	Papworth Hospital	51	290	280	68.6	21.6
Cornwall	Royal Cornwall Hospital	129	35	35	67.1	22
Exeter	Royal Devon & Exeter	34	89	87	66.2	24.2
Frimley	Frimley Park Hospital	19	112	109	70	21.8
Hull	Castle Hill Hospital	138	40	38	54	19.9
Leeds	St James's University Hospital	42	411	404	64.1	22.3
Leicester	Glenfield Hospital	142	81	81	70.3	22.2
Liverpool	Liverpool Heart and Chest Hospital	66	284	277	67.9	22.8
London-East	St. Bartholomew's Hospital	92	138	133	67.8	22
London-South East	King's College Hospital	5	197	189	71.4	21.8
London-South East	University Hospital Lewisham	105	54	54	60.6	20.6
London-South West	Royal Brompton Hospital	12	662	643	63.7	22
Manchester	Wythenshawe Hospital	102	394	389	63.5	21.8
Newcastle	Royal Victoria Infirmary	9	263	259	65	22
North West Staff	North West Staffs, Stoke on Trent	74	74	70	57.9	22
Norwich	Norfolk & Norwich University Hospital	114	73	70	69.8	21.6

Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2014	Median FEV_1% predicted (≥16 years)	Median BMI percentile (≥16 years)
Nottingham	Nottingham University Hospitals	101	154	151	66.3	21.9
Oxford	Churchill hospital	128	103	101	65.5	22.5
Plymouth	Derriford Hospital	64	52	51	76.7	23.5
Sheffield	Northern General Hospital	65	175	174	73.7	22.3
Southampton	Southampton General Hospital	110	227	215	69.2	22.2
Northern Ireland						
Belfast	Belfast City Hospital	14	233	220	71.5	22.7
Scotland						
Aberdeen	Aberdeen Royal Infirmary	70	66	61	57.4	22.5
Edinburgh	Western General Hospital	44	226	221	66.6	22.2
Glasgow	Gartnavel General Hospital	79	213	191	67.2	22.1
Wales						
Llandough	Llandough Hospital	68	216	216	70.4	22

Appendix 2: Glossary

Word/Phrase	Meaning
2014	1 January 2014 – 31 December 2014.
ABPA (Allergic Bronchopulmonary Aspergillosis)	When a person develops a respiratory allergic reaction to the Aspergillus fungus.
Absence of vas deferens	The vas deferens connect the testicles to the penis. Where a male is missing both vas deferens sperm cannot be transported.
Arthritis	A condition causing pain and inflammation in the joints.
Arthropathy	A condition causing pain in the joints.
Asthma	A respiratory condition causing episodes of difficulty breathing during attacks of spasm in the lung.
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).
GORD (Gastrooesophageal 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 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 passed away in any given year. So in 2014 the median age of the 137 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 2014, predicts how long we expect half of them to live for. For 2014, this means that half of people registered as alive on the database are predicted to live to at least 40.1. 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 can cause cystic fibrosis.
Nasal Polyps	Small, sac-like growths of inflamed mucus caused by chronic inflammation of the nasal lining.
Non-tuberculosis 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

Composition of UK CF Registry Steering Committee

Dr Janet Allen	Director of Strategic Innovation, Cystic Fibrosis Trust
Professor Diana Bilton	Adult CF Centre Director, Royal Brompton
Ms Noreen Caine	Contract Consultant, Cystic Fibrosis Trust
Dr Siobhán Carr	Paediatrician, Royal Brompton Hospital, London (Chair)
Ms Katherine Collins	Caldecott Guardian, Director NSD, Scotland
Ms Rebecca Cosgriff	Registry Lead, Cystic Fibrosis Trust
Dr Kim Cox	Lead Specialist CF Commissioner, London
Dr Steve Cunningham	Paediatrician, Royal Hospital for Sick Children, Scotland
Mrs Marian Dmochowska	Parent Representative
Dr Iolo Doull	Paediatric CF Centre Director, Cardiff Hospital, Wales
Dr Caroline Elston	Adult CF Centre Director, King's College Hospital, London
Ms Carrie Gardner	Specialised Commissioner, NHS England
Ms Elaine Gunn	Registry Manager, Cystic Fibrosis Trust
Mr Dominic Kavanagh	Patient Representative
Dr Claire Nelson	Specialised Commissioner, Wales
Dr Stephen Nyangoma	Biostatistician, Imperial College, London
Mr Ed Owen	Chief Executive, Cystic Fibrosis Trust
Ms Vian Rajabzadeh-Heshejin	Data Analyst, Imperial College, London
Dr Martin Wildman	Adult CF Centre Director, Northern General Hospital, Sheffield

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