Cystic Fibrosis our focus

UK Cystic Fibrosis Registry
Annual data report 2013

July 2014

Executive Summary

It is a pleasure to introduce the 2013 UK Cystic Fibrosis (CF) Registry data report, which provides information on more than 10000 people receiving care in CF centres across the UK. Collection of this data relies on the consent of parents of children with CF and adults with CF themselves, as well as the hard work of CF teams in collecting and recording the data as part of the care they provide. The Registry is managed and underwritten by the Cystic Fibrosis Trust as part of the strategic aims of beating the disease.

This report is published during the Trust's 50th year and is a timely opportunity to consider the initiatives that have had major influences on CF care to inspire us for future steps forward. The initial decision by the Trust to provide a research grant to Dr Anil Mehta, a visionary academic in Dundee, to set up a registry of people with CF has proved to be an important milestone in CF care in the UK. In 2006, the Trust took over the management of the Registry to guarantee high-quality data collection and analysis to underpin research and development in cystic fibrosis. A web-based system was adopted that was developed from the platform successfully used by the Cystic Fibrosis Foundation in the US, who generously gifted the licence for use to the Trust.

The Registry will continue to grow and adapt too. An external review conducted by Prof Kathy Rowan made a number of recommendations to build on its success to date, and the Trust will be taking these forward in the coming year.

Since the early days of the Registry, patient numbers have expanded, and now include more than 99% of all the people with CF in the UK. The Registry work has grown not only in numbers of patients but also in terms of the contracts and active projects supported, all in pursuit of improved care and outcomes for people with cystic fibrosis.

The Registry has become the key source of data for those commissioning CF services and, in the last year, has been recognised by the Federal Drug Administration in the US and the European Medicines Agency (the bodies responsible for licencing and regulating medicines) as one of the best registries for facilitating the safe and effective monitoring of new CF therapies. In doing so the Registry has provided an unrivalled way for the Trust to provide a service to people with CF, by ensuring new therapies can be made available in the UK while guaranteeing the careful monitoring of everyone receiving them.

The annual report is a central pillar in the Registry work and now represents a fulfilment of the contracts held between NHS commissioners and the Cystic Fibrosis Trust to provide detailed data on the health outcomes of people with cystic fibrosis and the provision of care.

Our ability as a community to provide accurate data has facilitated the funding decisions for introduction of new CF therapies. For example, we have been able to report to commissioners on the uptake of ivacaftor and will be able to report on the improvements in health seen as a result of this new therapy. Furthermore, the provision of centre-specific data facilitates the review of services and links to commissioner reviews of local service provisions.

We all want to see people with CF living longer and achieving their life ambitions without the need for burdensome treatments. In common with earlier publications, this report includes a calculation of the median predicted survival of the current population in the UK. This has increased in each of the last four years, to 43.5 years in 2012. In 2013 however, the figure dropped to 36.6 years.

This survival measure is related to variations in the number of deaths occurring in any one year, and experts analysing the data report that the trend over time is upwards, and that this year's figure is part of the usual variation.

Future reports will report this measure by showing five-year trends, in line with US Registry reports. We are also working on new models to be able to more accurately estimate survival for people born in any one year with CF, based on the steady improvements we are seeing.

It is particularly important to highlight the progress achieved in outcomes for people with cystic fibrosis. We know that chronic infection with *Pseudomonas aeruginosa* leads to faster decline in lung function and a higher requirement for treatment in people with cystic fibrosis. We also know that chronic infection can be prevented by strict adherence to cross-infection guidelines and aggressive application of recommended approaches to treat early infection.

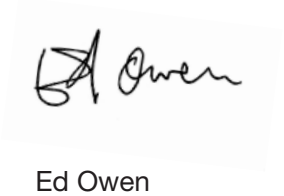
We are delighted that a trend identified last year has been confirmed and that, when comparing 2008 data with 2013, the proportion of patients with chronic *P. aeruginosa* infection is lower across all age groups from four to 31. The differences are statistically significant, and the largest difference is in adults. We believe this relates to the application of guidelines regarding infection control and eradication protocols for new
P. aeruginosa infection and should, over time, lead to an overall healthier CF population requiring less hospital treatment.

The 2013 report also shows that there has been an increase in the use of DNase (Pulmozyme®) over the last five years. This mucolytic agent is one of the therapies designed to make it easier to clear infected mucus and improve lung function, reducing the need for intravenous antibiotics. This and other therapies are being monitored to ensure equity of access to therapies and appropriate adherence to guidelines. Future reports are to demonstrate a broader range of therapies.

The Registry is therefore demonstrating that guidelines, information and investment are enabling more people with cystic fibrosis to receive preventative treatments rather than relying on rescue therapy in the form of intravenous antibiotics.

We hope you will take time to read the report carefully and join in the discussions that the data generate. These are exciting times in cystic fibrosis as new treatments are being developed and we consider how care is delivered to the increasing number of people with the condition. This increase equates to the challenge of a virtual new CF centre each year, and we will be engaging in conversations about sustainable models of care for the future based on evidence provided by the Registry.

Professor Diana Bilton
Chair, Registry Steering Committee



Ed Owen
Chief Executive, Cystic Fibrosis Trust

The Annual Data Report 2013: Summary can be downloaded from the Cystic Fibrosis Trust's website at: cysticfibrosis.org.uk/Registry

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Section 1: All UK patients

1.1 Summary of the UK Cystic Fibrosis Registry

	2009	2010	2011	2012	2013
CF patients registered Excluding diagnoses that year	9029[1]	9385[1]	9749[1]	10078[1] 9804	10338[1] 10076
CF patients with "complete" data; n(%) Rate of completeness excluding diagnoses that year	7377[2] (82%)	7937[2] (85%)	8679[2] (89%)	8794[2] (87%) 90%	9052[2] (88%) 90%
Age in years; median	17[3]	17[3]	18[3]	18[3]	18[3]
All newly diagnosed patients (newborn screening and other)	261[4]	301[4]	261[4]	274[4]	261[4]
Number of patients born each year identified by newborn screening Earlier data are updated as diagnoses data are updated (see full analysis in Section 1.3)		241	203	202	127
Age at diagnosis in months; median	3[3]	3[3]	3[3]	3[3]	3[3]
Adults aged 16 yrs and older; %	55.1[3]	55.5[3]	56.8[3]	57.6[3]	57.6[3]
Males; %	53.1[3]	53.1[3]	53.2[3]	52.9[3]	52.9[3]
Genotyped; %	94.3[3]	95.2[3]	95.6[3]	96.2[3]	97.2[3]
Median predicted survival in years (95% confidence interval)	34.4[5] (30.7, 37.0)	41.4[5] (36.8, 46.7)	41.5[5] (35.7, 46.0)	43.5[5] (37.8, 49.9)	36.6[5] (34.4, 41.6)
Total deaths reported	141 (1.6%)	103 (1.1%)	118 (1.2%)	106 (1.1%)	146 (1.4%)
Age at death in years; median (95% CI)[6]	27	29	26	28 (25, 29)	29 (27, 31)

Notes:

1 This is calculated as the number of patients on the database who satisfied the following criteria:
- were born and diagnosed with CF prior to 1 January 2010/2011/2012/2013/2014; and
- had no recorded date of death before 1 January 2009/2010/2011/2012/2013.

2 "Complete" data is defined as having a clinical encounter when "well".

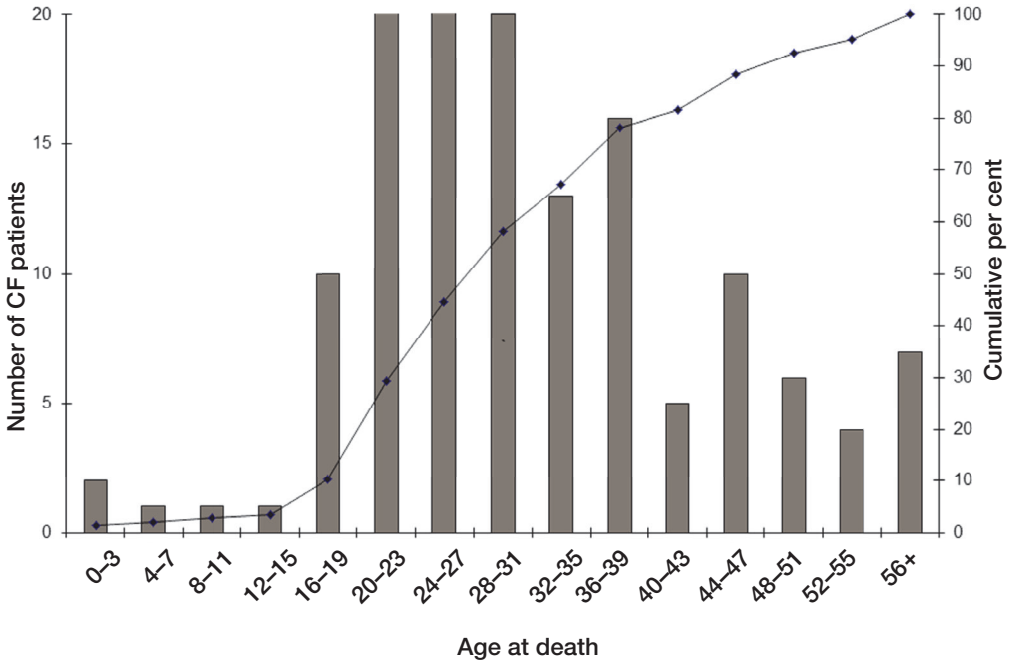
3 Calculated for patients with "complete" data in that given year.

4 Calculated for all patients registered.

5 This represents the age beyond which half of the current UK CF Registry patients would be expected to live, given the ages of CF patients in the Registry and the mortality distribution of deaths in the same year.

6 Confidence interval estimated using the bias-corrected and accelerated (BCa) bootstrap method.

1.2 Age distribution of deaths in 2013



There were 146 recorded deaths in 2013. The median age at death was 29 years (min = 0 yrs; max = 74 years; 95% confidence interval: 27–31 years).

Analyses based on 9052 patients with complete* data at 2013 annual review

1.3 Age at diagnosis and screening statistics among children

Age at diagnosis	All patients; n (%)	Patients aged 10 years in 2013; n (%)	Patients aged 5 years in 2013; n (%)
Pre-natal	3 (0.1)	0 (0.0)	0 (0.0)
Birth–3 months	2746 (72.6)	125 (59.8)	251(90.6)
4–6 months	228 (6.0)	13 (6.2)	5 (1.8)
7–12 months	155 (4.1)	11 (5.3)	6 (2.2)
1 yr	221 (5.8)	21 (10.1)	5 (1.8)
2 yrs	150 (4.0)	9 (4.3)	6 (2.2)
3 yrs	87 (2.3)	8 (3.8)	2 (0.7)
4 yrs	62 (1.6)	6 (2.9)	2 (0.7)
5 yrs	32 (0.8)	6 (2.9)	-
6 yrs	24 (0.6)	2 (1.0)	-
7 yrs	21 (0.6)	1 (0.5)	-
8 yrs	23 (0.6)	5 (2.4)	-
9 yrs	11 (0.3)	2 (1.0)	-
10 yrs	6 (0.2)	-	-
11 yrs	2 (0.1)	-	-
12 yrs	2 (0.1)	-	-
13 yrs	6 (0.2)	-	-
14 yrs	1 (0.0)	-	-
15 yrs	2 (0.1)	-	-
Overall	3782	209	277

The median (range) age at diagnosis is 30 days (0–185 months).

Diagnosis in the first three months of life was more common in children aged 5 years in 2013 (born in 2008) than in children aged 10 years in 2013 (born in 2003).

Of the 54 children with complete data born in 2013, 39 (72%) were identified by newborn screening.

A total of 127 patients born in 2013 were identified by newborn screening (including patients with and without complete data). In 2012 this figure was 202 and in 2011 it was 203. As there is a delay between when newborn screening tests are performed and the results inputted on to the Registry, these statistics are continuously updated as the Registry is updated. It is therefore anticipated that the number of patients born in 2013 and identified by newborn screening in 2013 will increase when new data become available on the Registry in 2014.

* “Complete” data refers to the minimum data required to produce the range of clinical outcomes presented in this report.

1.4 Age at diagnosis and screening statistics among current adults

Age at diagnosis	n (%)
Pre-natal	1 (0.02)
Birth–3 months	2047 (39.7)
4–6 months	479 (9.3)
7–12 months	333 (6.5)
1 yr	445 (8.6)
2 yrs	270 (5.2)
3 yrs	196 (3.8)
4 yrs	165 (3.2)
5 yrs	88 (1.7)
6 yrs	68 (1.3)
7 yrs	51 (1.0)
8 yrs	59 (1.1)
9 yrs	49 (1.0)
10 yrs	42 (0.8)
11 yrs	38 (0.7)
12 yrs	36 (0.7)
13 yrs	39 (0.8)
14 yrs	35 (0.7)
15 yrs	43 (0.8)
16–20 yrs	145 (2.8)
21–25 yrs	105 (2.0)
26–30 yrs	87 (1.7)
31–35 yrs	107 (2.1)
36–40 yrs	76 (1.5)
41–45 yrs	58 (1.1)
46–50 yrs	34 (0.7)
51–60 yrs	31 (0.6)
61 yrs+	32 (0.6)

The median (range) age at diagnosis is 7 months (0–79 years).

Of the 5213 adults with complete data in 2013, 413 were diagnosed by neonatal screening and 32 adults were diagnosed in 2013.

1.5 Genotyping

8799 (97.2%) patients have been genotyped with a recorded value.

DF508 Mutations; n (%)	
Homozygous DF508	4511 (51.3%)
Heterozygous DF508	3479 (39.5%)
No DF508 or both unidentified	809 (9.2%)

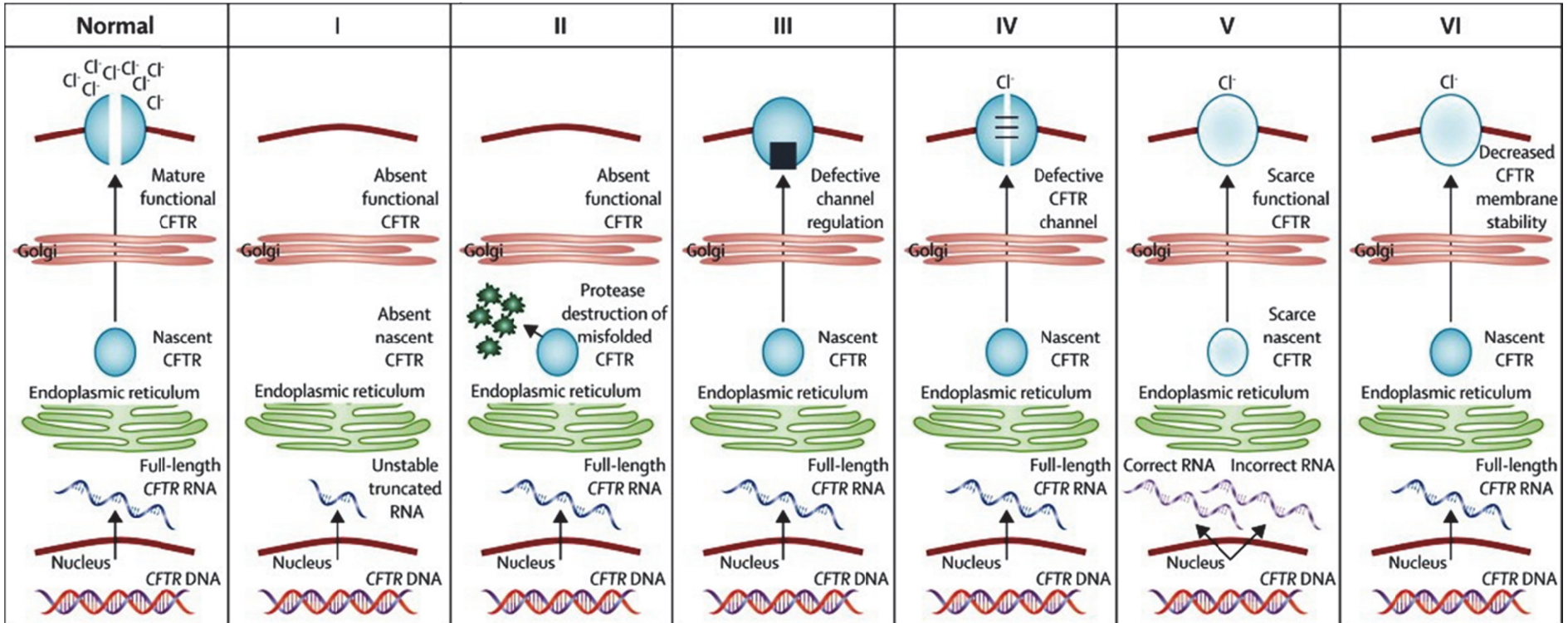
783 (8.9%) patients have at least one unknown genotype.

Mutations[1]

All mutations Current name	New name	N	(%)
DF508	p.Phe508del	7990	90.81
G551D	p.Gly551Asp	514	5.84
R117H	p.Arg117His	398	4.52
G542X	p.Gly542X	318	3.61
621+1G->T	c.489+1G>T	186	2.11
1717-1G->A	c.1585-1G>A	120	1.36
N1303K	p.Asn1303Lys	115	1.31
2789+5G->A	c.2657+5G>A	104	1.18
1898+1G->A	c.1766+1G>A	97	1.10
I507	p.Ile507del	91	1.03
3659delC	c.3528delC	89	1.01

[1] Only mutations that were observed in more than 1% of patients with complete data in 2013 were reported. Further information on CF mutations can be found at: http://www.cftr2.org/. For further information on the UK CF population, please contact the Trust – see cysticfibrosis.org.uk

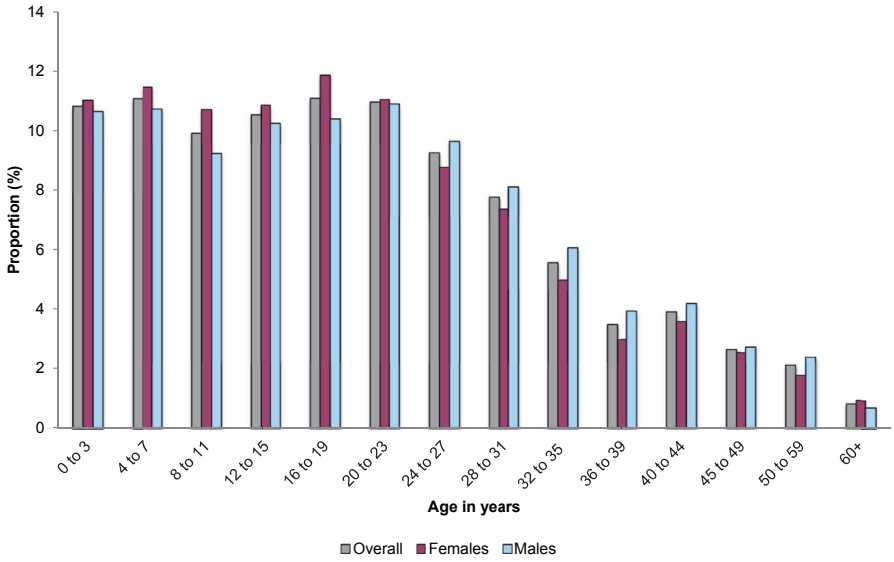
Cystic fibrosis mutations and their functional effects



	Normal	I	II	III	IV	V	VI
CFTR defect		No functional CFTR protein	CFTR trafficking defect	Defective channel regulation	Decreased channel conductance	Reduced synthesis of CFTR	Decreased CFTR stability
Type of mutations		Nonsense; frameshift; canonical splice	Missense; aminoacid deletion	Missense; aminoacid change	Missense; aminoacid change	Splicing defect; missense	Missense; aminoacid change
Specific mutation examples[11]		Gly542X Trp1282X Arg553X 621+1G→T	Phe508del Asn1303Lys Ile507del Arg560Thr	Gly551Asp Gly178Arg Gly551Ser Ser549Asn	Arg117His Arg347Pro Arg117Cys Arg334Trp	3849+10kbC→T 2789+5G→A 3120+1G→A 5T	4326delTC Gln1412X 4279insA

Courtesy of Boyle, DeBoeck, Lancet Respiratory Medicine 2013, 1: 158–63

1.6 Age distribution by gender

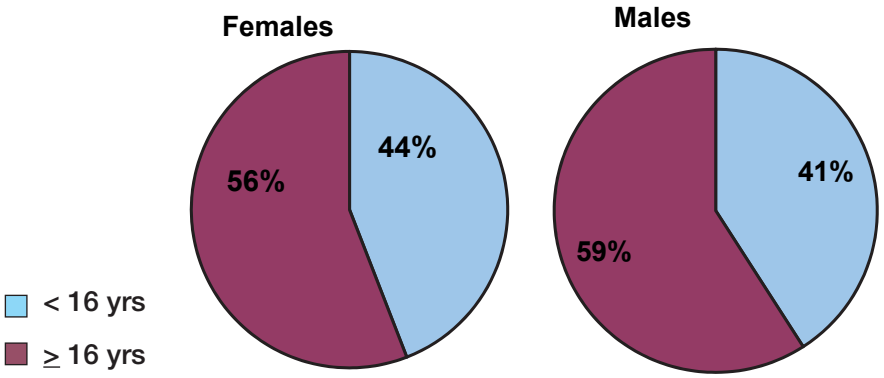


Age is calculated as the age at annual review encounter.

1.7 Age and sex distribution

Age	Overall N=9052	Female N=4268	Male N=4784
0–3 yrs	981 (10.8)	471 (11.0)	510 (10.7)
4–7	1004 (11.1)	490 (11.5)	514 (10.7)
8–11	899 (9.9)	458 (10.7)	441 (9.2)
12–15	955 (10.6)	464 (10.9)	491 (10.3)
16–19	1005 (11.1)	507 (11.9)	498 (10.4)
20–23	994 (11.0)	472 (11.1)	522 (10.9)
24–27	836 (9.2)	374 (8.8)	462 (9.7)
28–31	703 (7.8)	315 (7.4)	388 (8.1)
32–35	503 (5.6)	213 (5.0)	290 (6.1)
36–39	315 (3.5)	127 (3.0)	188 (3.9)
40–44	353 (3.9)	153 (3.6)	200 (4.2)
45–49	240 (2.7)	109 (2.6)	131 (2.7)
50–59	190 (2.1)	75 (1.8)	115 (2.4)
60+	74 (0.8)	40 (0.9)	34 (0.7)
Median (IQR)	18 (9–28)	18 (8–27)	19 (9–29)

1.8 Age distribution by sex



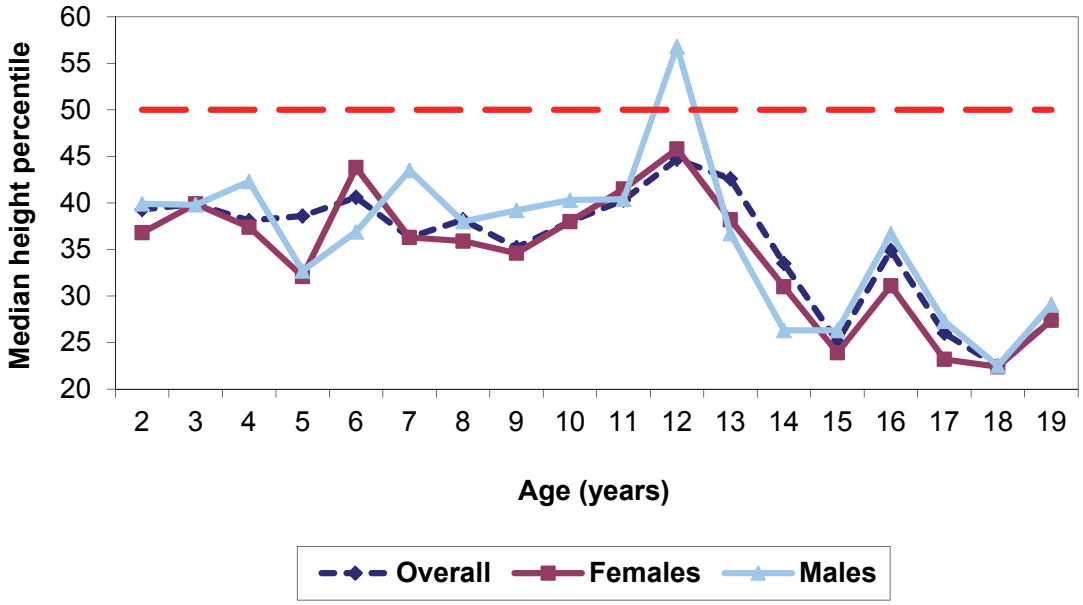
1.9 Employment and education status among adults aged 16 years and older

	Number of patients
Full-time working	1502
Part-time working	664
Student	922
Homemaker	232
Unemployed	685
“Disabled”	298
Retired	78
Unknown	914
No data	7

Note that these groups are not mutually exclusive.

Of the 4278 adults aged 16 years and older for whom an employment status questionnaire was completed (excluding “unknown”), **3031 (70.9%) reported being in work or study**. In 2009, this figure was 68.8%.

1.10 Median height percentiles among children and young people (<20 years) (n=4346)



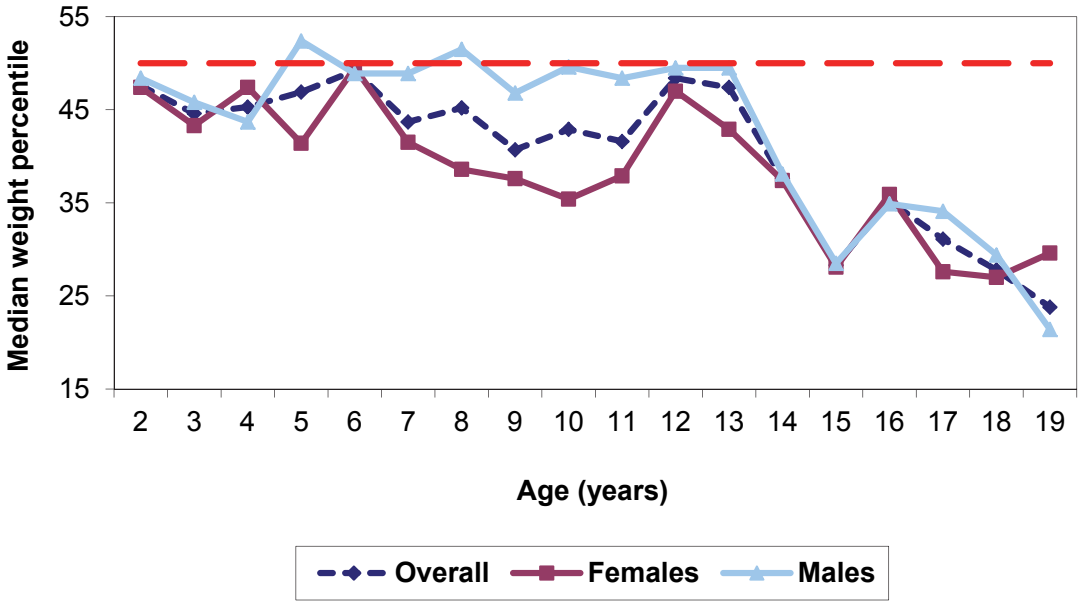
N refers to the number of patients in each age/sex category who had non-missing height data.

The red dotted line indicates the 50th percentile, which is a marker used to target growth in children. The aim is to monitor and maintain growth as close to the 50th percentile as possible.

Age	N	Overall Median (IQR)	N	Female Median (IQR)	N	Male Median (IQR)
2	265	39.3 (15.1,60.1)	132	36.8 (14.1,59.9)	133	39.9 (19.2,60.5)
3	268	39.9 (18.7, 62.9)	127	39.9 (14.9, 60.6)	141	39.8 (19.9, 65.0)
4	247	38.1 (17.6, 63.6)	124	37.4 (16.5, 65.5)	123	38.6 (17.9, 63.0)
5	273	38.6 (14.7, 60.3)	132	32.1 (11.5, 60.1)	141	42.3 (17.5, 61.5)
6	254	40.6 (18.2, 64.9)	126	43.8 (17.8, 64.9)	128	32.7 (18.9. 65.6)
7	217	36.3 (15.2, 62.7)	101	36.3 (15.0, 60.9)	116	36.9 (15.3, 63.5)
8	236	38.2 (17.3, 59.8)	110	35.9 (15.4, 53.8)	126	43.5 (18.4, 70.1)
9	235	35.2 (13.7, 65.3)	132	34.6 (13.9, 62.6)	103	38.0 (13.6, 67.9)
10	208	38.0 (12.0, 65.2)	103	38.0 (14.1, 64.7)	105	39.2 (9.9, 67.7)
11	212	40.3 (17.0, 71.6)	109	41.5 (15.6, 71.0)	103	40.3 (19.7, 71.7)
12	224	44.7 (19.8, 75.0)	111	45.8 (26.0, 76.0)	113	40.4 (15.8, 74.8)
13	223	42.6 (17.4, 74.0)	117	38.2 (14.4, 66.1)	106	56.8 (20.9, 76.8)
14	274	33.5 (11.7, 64.1)	128	31.0 (9.7, 61.8)	146	36.7 (12.9, 68.2)
15	228	25.1 (9.4, 53.4)	106	23.9 (7.5, 54.8)	122	26.3 (13.1, 51.8)
16	263	34.9 (10.2,59.1)	118	31.1 (7.8, 59.6)	145	36.7 (14.0, 58.7)
17	237	26.0 (8.1, 50.0)	121	23.2 (6.4, 50.7)	116	27.3 (12.6, 50.0)
18	254	22.5 (9.1, 54.1)	142	22.4 (9.9, 52.7)	112	22.5 (8.4, 56.2)
19	228	27.4 (10.3,53.9)	115	27.4 (9.7, 52.4)	113	29.1 (11.6, 54.0)

Age	N	Overall Median (IQR)	N	Female Median (IQR)	N	Male Median
Overall	4346	35.4 (13.8, 62.7)	2154	34.5 (12.5, 60.9)	2192	36.7 (14.9, 63.6)
2–4 yrs	780	39.1 (17.1, 61.4)	383	39.1 (15.1, 60.8)	397	39.1 (19.1, 61.9)
5–7 yrs	744	37.7 (15.5, 63.2)	359	37.3 (15.2, 63.0)	385	38.5 (15.9, 63.6)
8–10 yrs	679	37.6 (14.7, 63.0)	345	35.2 (14.8, 60.4)	334	41.3 (14.4, 68.0)
11–13 yrs	659	42.5 (18.0, 72.9)	337	42.2 (16.9, 71.7)	322	43.8 (18.5, 74.6)
14–15 yrs	502	29.2 (11.0, 59.6)	234	28.1 (9.4, 59.6)	268	29.8 (13.0, 59.6)
16–19 yrs	982	27.4 (9.8, 54.1)	496	27.3 (7.8, 53.6)	486	28.3 (11.7, 54.6)

1.11 Median weight percentiles among children and young people (<20 years) (n=4365)

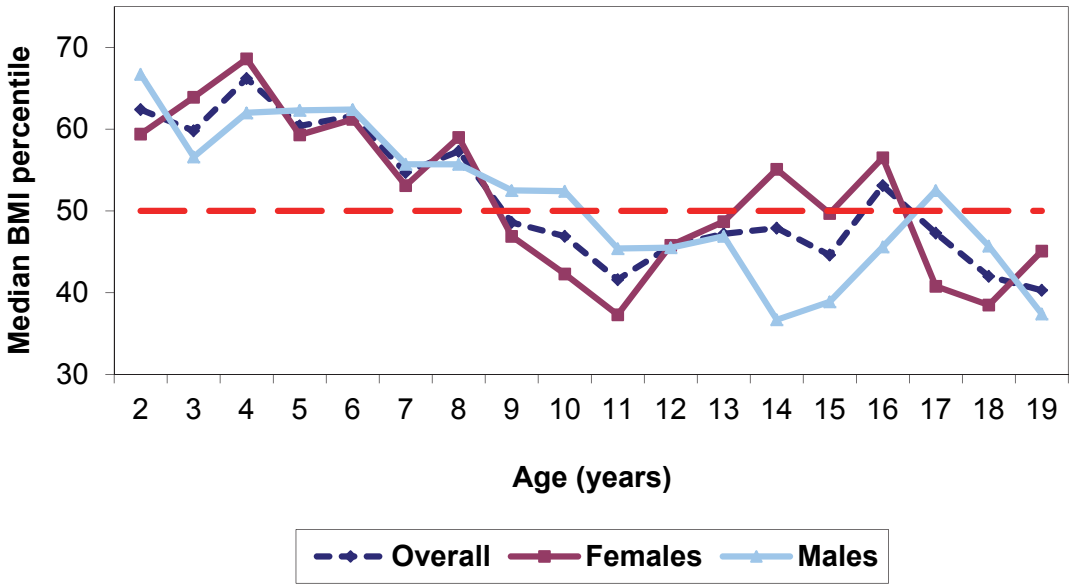


N refers to the number of patients in each age/sex category who had non-missing weight data.

The red dotted line indicates the 50th percentile, which is a marker used to target weight in children. The aim is to monitor and maintain weight as close to the 50th percentile as possible.

Age	N	Overall Median (IQR)	N	Female Median (IQR)	N	Male Median (IQR)
2	269	47.6 (23.8, 72.1)	134	47.4 (19.0, 72.5)	135	48.4 (24.7, 71.8)
3	271	44.6 (22.3, 71.8)	128	43.3 (18.7, 72.1)	143	45.8 (22.4, 71.8)
4	247	45.3 (20.4, 71.6)	124	47.4 (23.2, 71.0)	123	43.7 (18.9, 71.8)
5	276	46.9 (24.8, 71.0)	133	41.4 (18.8, 63.6)	143	52.4 (28.2, 75.5)
6	254	49.2 (23.8, 73.9)	126	49.5 (26.2, 74.4)	128	48.9 (23.1, 72.1)
7	217	43.7 (18.4, 69.0)	101	41.5 (19.0, 64.5)	116	48.9 (17.9, 70.8)
8	235	45.2 (23.9, 70.6)	110	38.6 (18.9, 66.5)	125	51.5 (26.7, 80.2)
9	234	40.7 (19.6, 70.7)	132	37.6 (17.0, 69.7)	102	46.8 (27.2, 70.7)
10	209	42.9 (19.4, 64.8)	103	35.4 (16.9, 59.5)	106	49.6 (24.2, 71.2)
11	213	41.6 (20.7, 70.4)	110	37.9 (15.1, 66.4)	103	48.4 (29.1, 75.2)
12	224	48.4 (23.4, 76.5)	111	47.0 (23.1, 70.4)	113	49.5 (23.7, 81.7)
13	224	47.4 (18.0, 76.7)	118	42.9 (14.3, 71.5)	106	49.5 (26.7, 79.9)
14	275	37.6 (15.7, 66.6)	129	37.4 (15.6, 66.7)	146	38.1 (15.7, 66.2)
15	229	28.3 (10.6, 58.0)	106	28.1 (11.5, 58.2)	123	28.5 (10.3, 57.0)
16	266	35.2 (14.3, 65.5)	121	35.9 (13.5, 70.3)	145	34.9 (14.2, 62.9)
17	240	31.1 (10.3, 60.4)	122	27.6 (7.2, 49.7)	118	34.1 (11.3, 65.9)
18	254	27.8 (8.7, 63.5)	142	27.0 (8.7, 60.7)	112	29.4 (8.3, 65.2)
19	228	23.8 (8.0, 59.2)	115	29.6 (13.5, 59.5)	113	21.4 (5.7, 55.7)
Overall	4365	41.0 (16.9, 68.4)	2165	38.8 (16.2, 66.6)	2200	43.3 (18.0, 70.3)
2–4 yrs	787	45.9 (22.3, 71.8)	386	45.8 (19.8, 72.2)	401	45.9 (22.4, 71.8)
5–7 yrs	747	46.8 (22.4, 70.9)	360	43.9 (21.2, 68.8)	387	50 (23.5, 72.3)
8–10 yrs	678	43.0 (21.2, 69.8)	345	37.0 (18.0, 65.5)	333	48.8 (26.4, 72.2)
11–13 yrs	661	45.6 (20.7, 73.3)	339	42.5 (16.3, 69.2)	322	49.4 (26.3, 79.0)
14–15 yrs	504	33.8 (12.9, 63.9)	235	33.0 (13.2, 65.0)	269	33.9 (12.0, 59.5)
16–19 yrs	988	30.0 (9.9, 62.6)	500	29.6 (10.3, 61.1)	488	30.5 (9.0, 63.4)

1.12 Median BMI percentiles among children and young people (<20 years) (n=4237)



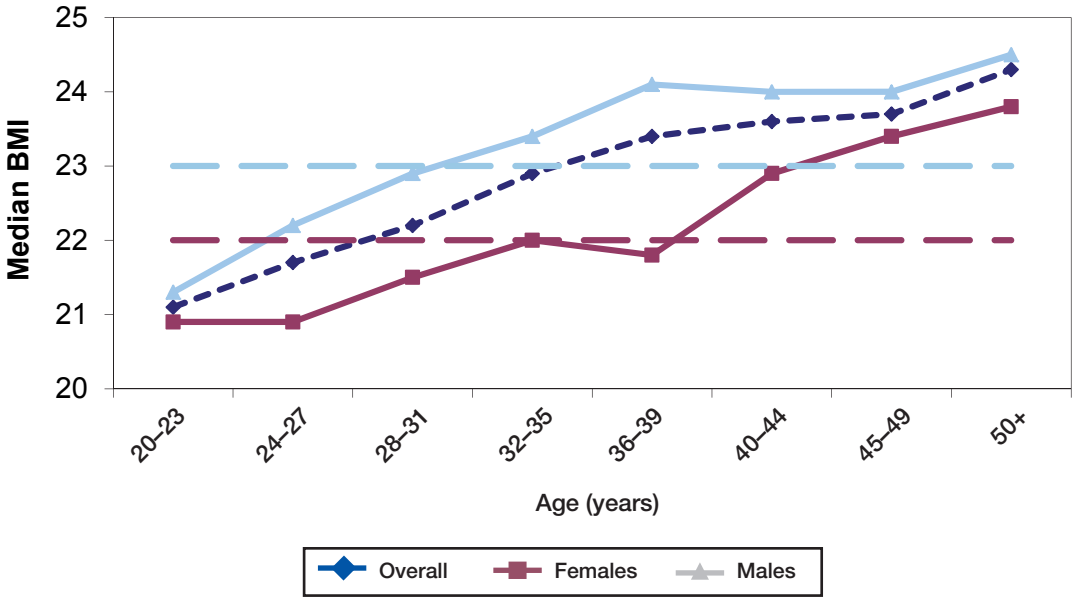
N refers to the number of patients in each age/sex category who had non-missing BMI data.

The red dotted line indicates the 50th percentile, which is a marker used to target weight for height in children. The aim is to monitor and maintain weight for height as close to the 50th percentile as possible. BMI percentiles for young people aged 16 to 19 years were calculated separately using LMS Growth software with British 1990 reference values.

Age	N	Overall Median (IQR)	N	Female Median (IQR)	N	Male Median (IQR)
2	265	62.4 (31.9, 82.4)	132	59.4 (26.0, 79.7)	133	66.7 (41.0, 85.2)
3	268	59.8 (34.7, 81.0)	127	63.9 (39.0, 77.5)	141	56.6 (31.8, 82.8)
4	247	66.2 (36.3, 82.8)	124	68.6 (40.9, 82.3)	123	62.0 (33.1, 83.9)
5	273	60.4 (38.1, 82.1)	132	59.3 (37.4, 77.6)	141	62.3 (38.8, 86.1)
6	254	61.6 (35.0, 80.0)	126	61.2 (41.3, 80.5)	128	62.4 (30.0, 78.7)
7	217	54.7 (29.6, 74.6)	101	53.1 (28.9, 75.1)	116	55.7 (30.0, 74.2)
8	235	57.3 (34.2, 77.2)	110	59.0 (32.8, 76.7)	125	55.7 (36.2, 77.4)
9	234	48.6 (28.9, 71.3)	132	46.9 (25.5, 72.6)	102	52.5 (30.7, 69.4)
10	208	46.9 (25.1, 66.9)	103	42.3 (22.1, 62.3)	105	52.4 (31.4, 71.1)
11	212	41.6 (23.3, 68.2)	109	37.3 (21.2, 61.8)	103	45.4 (26.0, 74.6)
12	224	45.7 (22.6, 71.4)	111	45.8 (20.1, 70.8)	113	45.5 (23.7, 73.2)
13	223	47.2 (21.5, 72.5)	117	48.7 (25.7, 73.7)	106	46.9 (20.6, 70.9)
14	274	47.9 (19.0, 68.6)	128	55.1 (36.2, 70.9)	146	36.7 (11.2, 63.7)
15	123	44.6 (16.3, 69.3)	62	49.7 (28.9, 73.6)	61	38.9 (8.7, 58.8)
16	263	53.1 (23.4, 75.6)	118	56.5 (29.6, 79.4)	145	45.6 (19.4, 72.7)
17	236	47.3 (20.6,76.0)	120	40.8 (20.3, 72.4)	116	52.5 (21.7, 80.8)
18	253	42.0 (16.9, 72.1)	142	38.5 (18.1, 72.1)	111	45.7 (15.1, 72.1)
19	228	40.3 (11.8, 67.3)	115	45.1 (17.0, 67.4)	113	37.4 (9.0, 68.3)
Overall	4237	51.8 (26.5, 75.6)	2109	52.0 (27.1, 75.0)	2128	51.4 (25.7, 76.4)

Age	N	Overall Median (IQR)	N	Female Median (IQR)	N	Male Median (IQR)
2–4 yrs	780	62.6 (34.4, 82.3)	383	63.0 (33.8, 80.5)	397	61.7 (34.6, 84.0)
5–7 yrs	744	59.4 (35.3, 79.0)	359	58.3 (36.2, 77.8)	385	60.1 (33.0, 80.3)
8–10 yrs	677	50.6 (29.2, 73.2)	345	47.4 (26.2, 71.7)	332	54.4 (33.7, 75.0)
11–13 yrs	659	44.4 (22.5, 70.0)	337	43.7 (22.4, 69.0)	322	45.6 (22.5, 73.3)
14–15 yrs	397	46.9 (18.3, 68.8)	190	52.7 (33.7, 71.4)	207	38.5 (9.9, 62.2)
16–19 yrs	980	45.4 (19.2, 73.7)	495	45.2 (20.5, 73.4)	485	45.6 (17.0, 74.0)

1.13 Median BMI values among adults aged 20 years and older (n=4086)

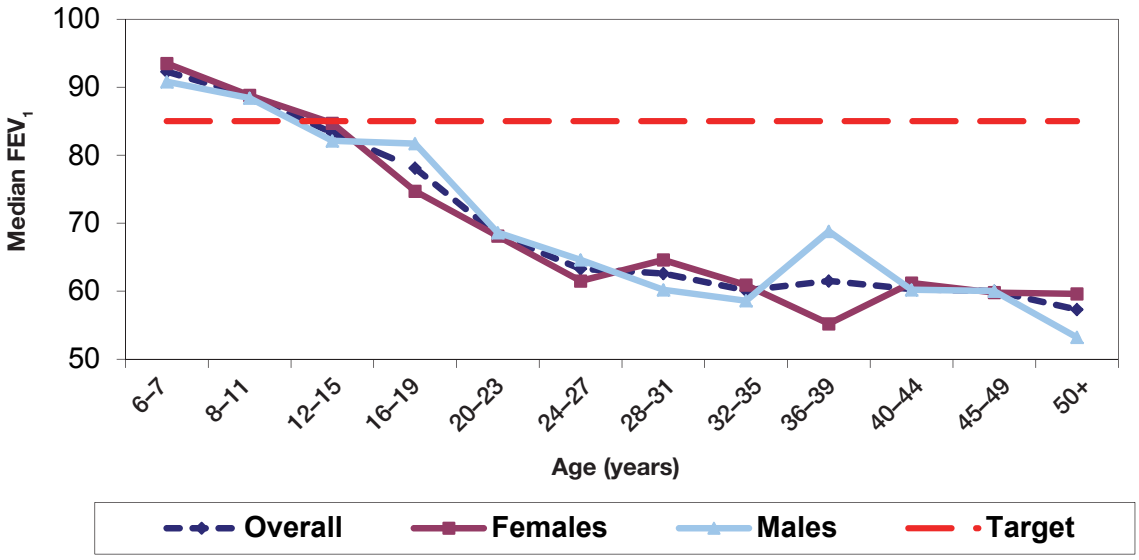


N refers to the number of patients in each age/sex category with non-missing BMI data.

The purple dotted line indicates a BMI of 22, which is a marker used to target BMI in adult women; the blue dotted line indicates a BMI of 23, which is a marker used for adult men. Individuals aged between 16 and 19 are not included in this graph because the absolute BMI value can be misleading for this age group.

Age	N	Overall Median (IQR)	N	Female Median (IQR)	N	Male Median (IQR)
20–23	966	21.1 (19.1, 23.3)	460	20.9 (18.9, 23.0)	506	21.3 (19.3, 23.6)
24–27	812	21.7 (19.7, 23.8)	363	20.9 (19.1, 22.9)	449	22.2 (20.3, 24.6)
28–31	685	22.2 (20.2, 24.4)	309	21.5 (19.9, 23.6)	376	22.9 (20.7, 25.1)
32–35	487	22.9 (20.8, 24.9)	205	22.0 (20.1, 24.5)	282	23.4 (21.3, 25.1)
36–39	306	23.4 (21.2, 25.6)	122	21.8 (20.4, 24.3)	184	24.1 (22.3, 26.0)
40–44	339	23.6 (21.3, 25.7)	144	22.9 (20.3, 25.3)	195	24.0 (22.2, 25.8)
45–49	230	23.7 (21.7, 26.7)	102	23.4 (20.9, 25.7)	128	24.0 (22.3, 27.2)
50+	261	24.3 (21.8, 27.1)	114	23.8 (21.0, 27.5)	147	24.5 (22.7, 26.8)
Overall	4086	22.3 (20.2, 24.7)	1819	21.6 (19.7, 24.0)	2267	22.9 (20.8, 25.2)

1.14a Median FEV_1 (% predicted) among patients aged 6 years and older, excluding patients post lung transplant (n=6923)



N refers to the number of patients in each age/sex category among those with non-missing FEV_1 % predicted data.

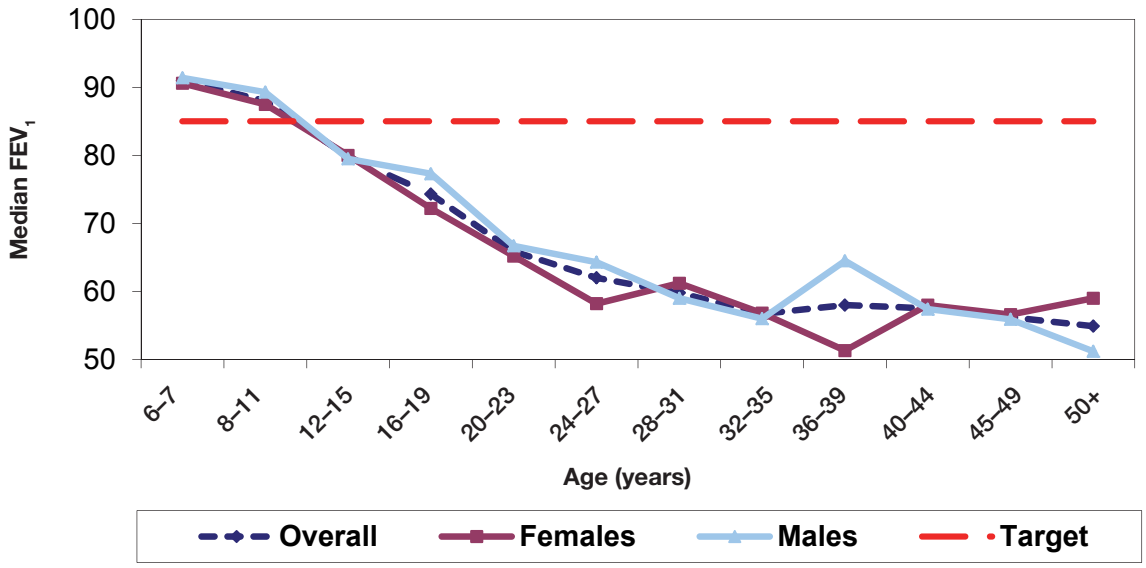
The dotted line in this figure illustrates a target FEV_1 % predicted of 85%. Anything above this indicates normal or near-normal lung function values.

Age	N	Overall Median (IQR)	N	Female Median (IQR)	N	Male Median (IQR)
6–7	421	92.3 (80.1,101.6)	201	93.5 (82.2, 102.5)	220	90.8 (78.5, 100.2)
8–11	858	88.7 (77.0, 97.9)	441	88.8 (76.9, 97.8)	417	88.4 (77.2, 97.9)
12–15	919	83.3 (69.1, 94.8)	447	84.7 (70.1, 96.5)	472	82.1 (68.6, 93.7)
16–19	952	78.1 (58.9, 92.6)	489	74.7 (54.5, 89.1)	473	81.7 (64.6, 96.4)
20–23	933	68.3 (48.8, 85.5)	443	68.1 (47.1, 84.5)	490	68.6 (49.7, 86.1)
24–27	771	63.3 (43.8, 82.5)	346	61.5 (43.8, 85.2)	425	64.6 (44.0, 81.3)
28–31	636	62.6 (42.6, 82.3)	281	64.6 (45.4, 86.0)	355	60.2 (40.6, 81.2)
32–35	446	60.1 (42.8, 77.8)	186	60.9 (45.0, 81.3)	260	58.6 (41.6, 77.5)
36–39	267	61.5 (44.0, 78.5)	108	55.2 (41.3, 70.9)	159	68.8 (45.4, 80.7)
40–44	294	60.3 (41.8, 79.8)	127	61.2 (42.6, 80.2)	167	60.2 (39.6, 79.1)
45–49	203	60.0 (41.2, 82.0)	94	59.8 (44.9, 74.4)	109	60.0 (37.8, 89.2)
50+	223	57.3 (41.4, 80.7)	95	59.6 (45.6, 80.5)	128	53.2 (38.8, 80.8)
Overall	6923	75.0 (53.9, 91.0)	3249	75.4 (53.8, 91.4)	3674	74.8 (53.9, 90.5)

The aim of good CF care is to preserve normal lung function for as long as possible among the paediatric population and to maintain stable lung function in adulthood. This is important for the latter as lung function at 50% and above will facilitate all of the normal activities of daily living, including attendance at work and college.

The proportion of patients aged 6 and older with a value of FEV_1 less than 85% predicted was 65%.

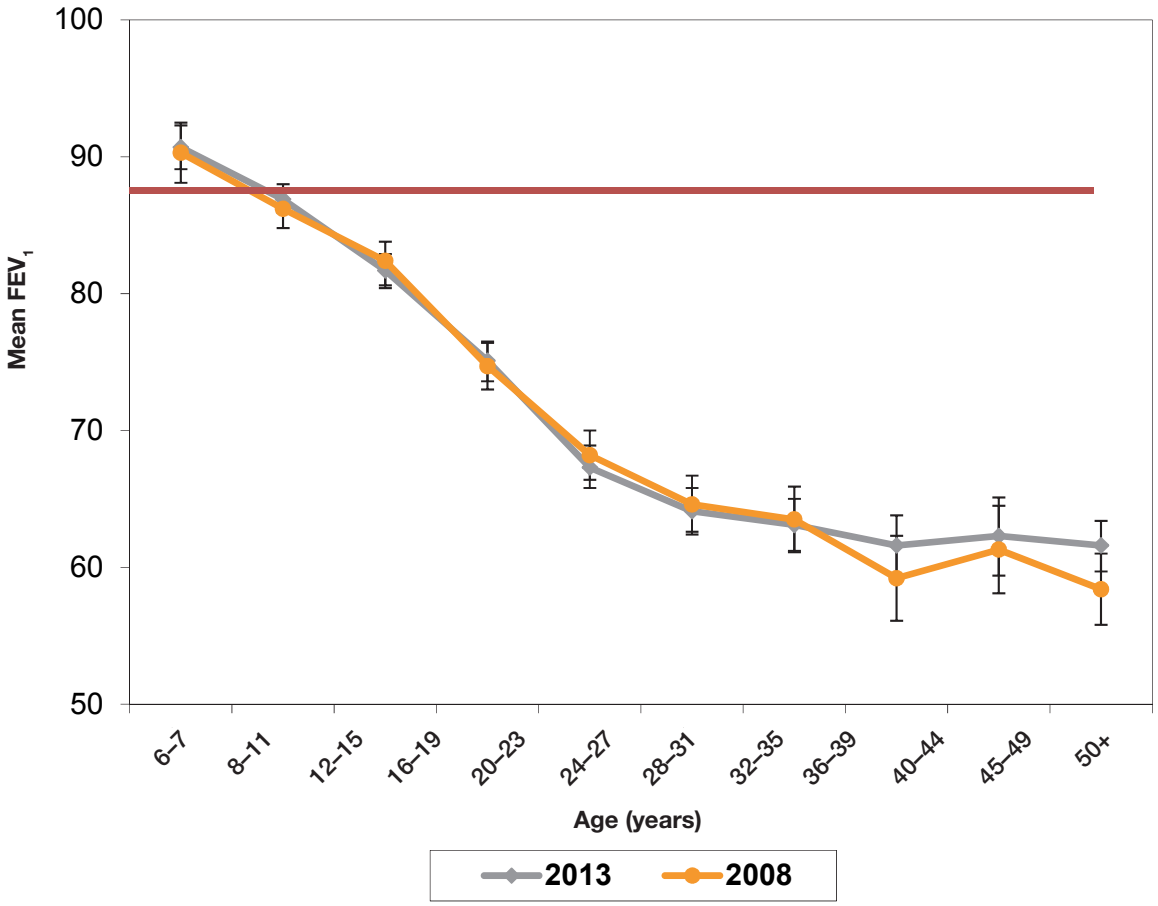
1.14b Median FEV_1 (% predicted, GLI equations) among patients aged 6 years and older, excluding patients post lung transplant (n=6923)



Age	N	Overall Median (IQR)	N	Female Median (IQR)	N	Male Median (IQR)
6–7	421	91.0 (78.3, 99.9)	201	90.6 (78.5,100.1)	220	91.4 (77.7,99.6)
8–11	858	88.0 (77.1,98.0)	441	87.5 (75.6,97.0)	417	89.3 (78.1,99.7)
12–15	919	79.8 (67.1, 91.3)	447	80.0 (66.5, 91.0)	472	79.5 (67.3, 91.4)
16–19	952	74.3 (56.4, 88.4)	480	72.2 (53.0, 86.3)	472	77.3 (60.6, 90.5)
20–23	933	65.9 (47.4, 82.5)	443	65.2 (45.3, 80.8)	490	66.7 (48.8, 84.0)
24–27	771	62.0 (42.7, 79.9)	346	58.2 (42.0, 80.5)	425	64.3 (43.7, 79.8)
28–31	636	59.8 (40.3, 79.0)	281	61.2 (43.0, 81.2)	355	59.0 (39.0, 78.3)
32–35	446	56.7 (41.2, 75.3)	186	56.8 (41.6, 75.3)	260	56.0 (40.9, 75.5)
36–39	267	58.0 (41.4, 76.2)	108	51.3 (38.5, 66.9)	159	64.5 (44.7, 78.0)
40–44	294	57.5 (39.7, 75.6)	127	58.0 (40.0, 75.4)	167	57.4 (38.5, 76.3)
45–49	203	56.2 (39.7, 80.5)	94	56.6 (42.5, 70.5)	109	55.9 (36.5, 83.4)
50+	223	54.9 (39.7, 79.0)	95	59.0 (44.5, 79.1)	128	51.2 (36.9, 78.8)
Overall	6923	72.2 (51.8, 88.3)	3249	71.8 (51.3, 87.8)	3674	72.5 (52.4, 89.1)

N refers to the number of patients in each age/sex category among those with non-missing FEV_1 % predicted data.

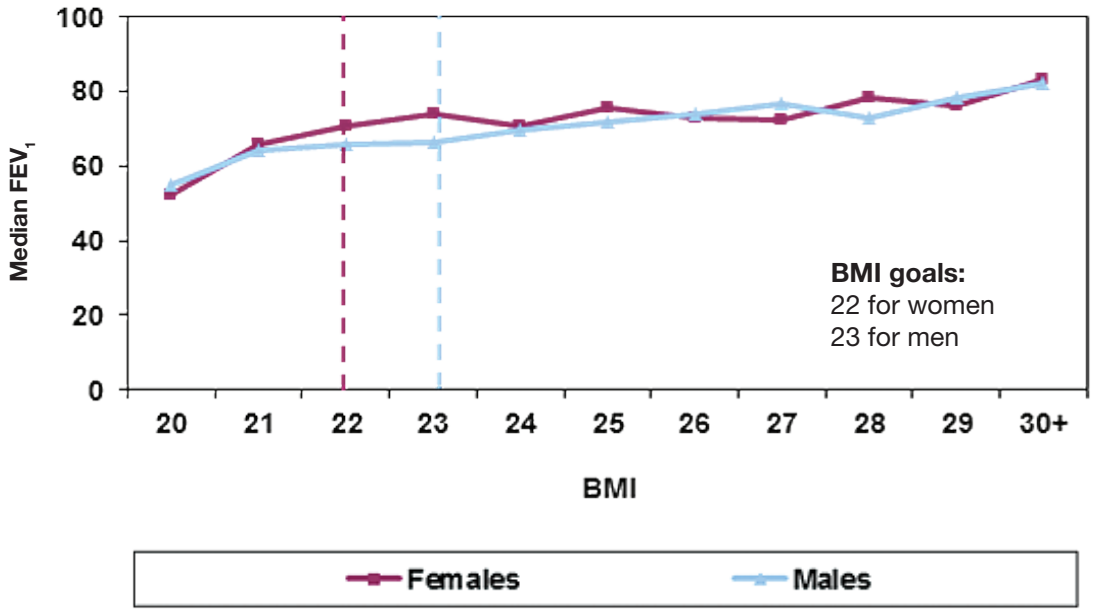
1.15 Mean FEV_1 (% predicted) among patients aged 6 years and older by year in 2008 and 2013 (excluding patients post lung transplant)



An analysis was conducted in order to determine whether there were statistically significant differences in FEV_1 (% predicted) in 2013 compared to 2008 by age category. The results show that there was a small but statistically significant difference among patients aged 40 years and older, with FEV_1 levels being higher in 2013.

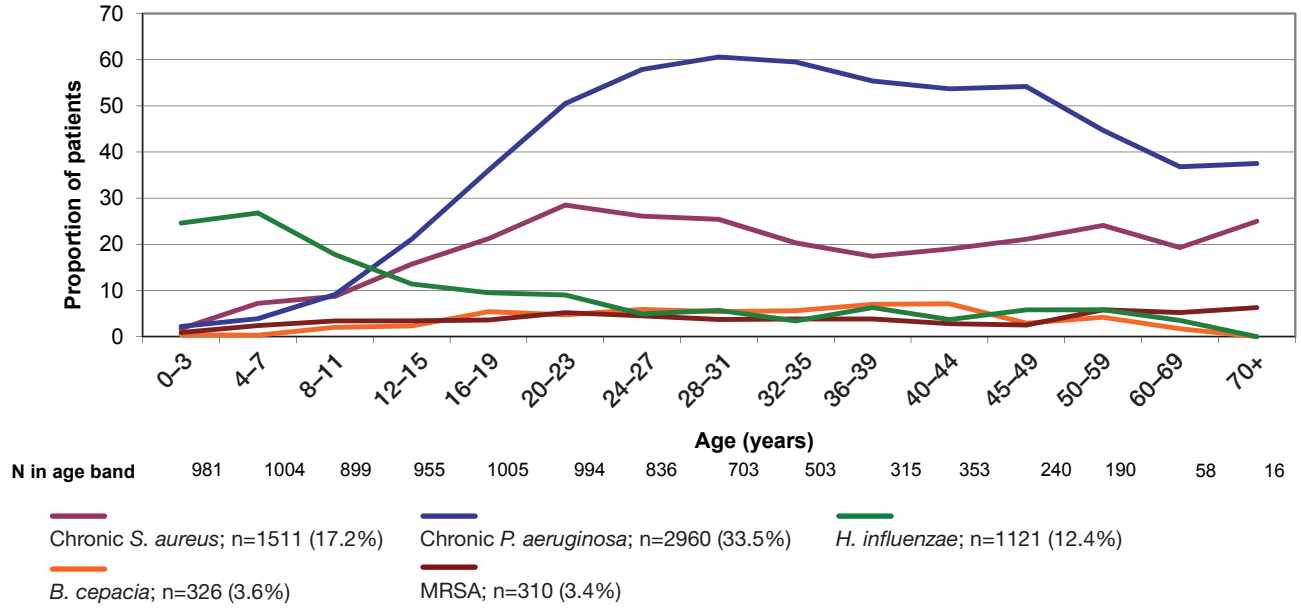
	Age (years)									
	6–7	8–11	12–15	16–19	20–23	24–27	28–31	32–35	36–39	40+
p-value	0.783	0.307	0.758	0.429	0.764	0.741	0.401	0.096	0.357	0.003

1.16 Median FEV_1 (% predicted) vs BMI among patients aged 16 years and older (excluding patients post 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 we grouped all BMI≥30 into one group.

1.17 Lung infections in 2013



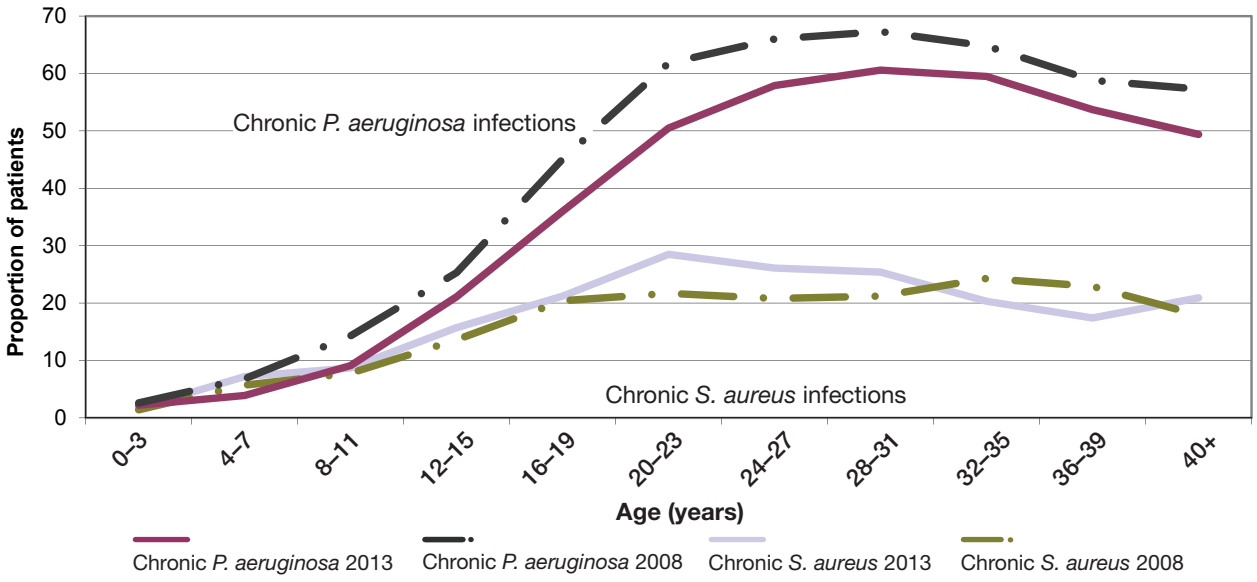
Chronic infection with *S. aureus* or *P. aeruginosa* were identified from annual review. Data on *B. cepacia, MRSA* and *H. influenzae* were collected from culture results at annual review.

Current treatments and good cross-infection measures mean that we can aim to reduce the number of people with CF transferring from paediatric to adult care with chronic *P. aeruginosa* infection, and currently the aim is for less than 30% of paediatric patients to be chronically infected at the time of transfer. A future aim is to see this reduce to less than 20%.

	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+	All	Children (<16 years)	Adults (≥16 years)
N patients in age band	981	1004	899	955	1005	994	836	703	503	315	353	240	264	9052	3839 (42.4)	5213 (57.6)
Chronic *S. aureus*; n(%)	16 (1.7)	70 (7.2)	75 (8.7)	142 (15.7)	207 (21.2)	280 (28.5)	217 (26.1)	175 (25.4)	100 (20.3)	54 (17.4)	66 (19.0)	49 (21.1)	60 (23.1)	1511 (17.2)	303 (8.2)	1208 (23.6)
Chronic *P. aeruginosa*; n(%)	21 (2.2)	38 (3.9)	78 (9.1)	192 (21.1)	356 (36.0)	496 (50.5)	479 (57.9)	421 (60.6)	294 (59.5)	169 (53.7)	188 (54.2)	117 (50.0)	111 (42.5)	2960 (33.5)	329 (8.9)	2631 (51.1)
Intermittent *P. aeruginosa*; n(%)	183 (19.2)	215 (22.2)	173 (20.1)	219 (24.1)	187 (18.9)	167 (17.0)	106 (12.8)	77 (11.1)	66 (13.4)	43 (13.7)	34 (9.8)	29 (12.4)	32 (12.3)	1531 (17.3)	790 (21.4)	741 (14.4)
Intermittent *S. aureus*; n(%)	158 (16.6)	193 (19.9)	216 (25.1)	163 (18.0)	181 (18.5)	169 (17.2)	128 (15.4)	102 (14.8)	55 (11.2)	50 (16.1)	41 (11.8)	27 (11.6)	34 (13.1)	1517 (17.2)	730 (19.8)	787 (15.4)
B. cepacia; n(%)	4 (0.4)	3.0 (0.3)	18 (2.0)	22.0 (2.3)	54 (5.4)	47 (4.7)	49 (5.9)	38 (5.4)	28 (5.6)	22 (7.0)	25 (7.1)	7 (2.9)	9 (3.4)	326 (3.6)	47 (1.2)	279 (5.4)
MRSA; n(%)	9 (0.9)	24 (2.4)	31 (3.4)	32.0 (3.4)	36 (3.6)	52 (5.2)	38 (4.5)	26 (3.7)	19 (3.8)	12 (3.8)	10 (2.8)	6 (2.5)	15 (5.7)	310 (3.4)	96 (2.5)	214 (4.1)
H. influenza; n(%)	241 (24.6)	269 (26.8)	160 (17.8)	109 (11.4)	95 (9.5)	89 (9.0)	41 (4.9)	40 (5.7)	17 (3.4)	20 (6.3)	13 (3.7)	14 (5.8)	13 (4.9)	1121 (12.4)	779 (20.3)	342 (6.6)

Age is calculated as age at annual review

1.18 Lung infections in 2008 and 2013



		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	0.633	0.014	0.002	0.051	0.000	0.000	0.003	0.030	0.172	0.247	0.061
Chronic *S. aureus;* p-value	0.690	0.252	0.527	0.262	0.694	0.002	0.024	0.127	0.225	0.123	0.160

1.19a Prevalence of complications

	Overall (n=9052)	<16 years (n=3839)	≥16 years (n=5213)
Respiratory related			
Nasal polyps requiring surgery; n(%)	168 (1.9)	36 (0.9)	132 (2.5)
Sinus disease; n(%)	746 (8.2)	48 (1.3)	698 (13.4)
Asthma; n(%)	1391 (15.4)	545 (14.2)	846 (16.2)
ABPA; n(%)	948 (10.5)	277 (7.2)	671 (12.9)
Haemoptysis; n(%)	76 (0.8)	0 (0.0)	76 (1.5)
Pneumothorax requiring chest tube; n(%)	53 (0.6)	2 (0.1)	51 (1.0)
Non-tuberculous mycobacteria or atypical mycobacteria; n(%)	512 (5.7)	107 (2.8)	405 (7.8)
Pancreas and hepatobiliary disease			
Liver enzymes; n(%)	1061 (11.7)	259 (6.7)	802 (15.4)
Liver disease; n(%)	1204 (13.3)	347 (9.0)	857 (16.4)
Cirrhosis with no portal hypertension; n(%)	129 (1.4)	31 (0.8)	98 (1.9)
Cirrhosis with portal hypertension; n(%)	160 (1.8)	20 (0.5)	140 (2.7)

	Overall (n=9052)	<16 years (n=3839)	≥16 years (n=5213)
Gallbladder disease requiring surgery; n(%)	33 (0.4)	0 (0.0)	33 (0.6)
Pancreatitis; n(%)	76 (0.8)	4 (0.1)	72 (1.4)
GI bleed req. hosp variceal; n(%)	9 (0.1)	2 (0.1)	7 (0.1)
Upper gastrointestinal			
GERD; n(%)	1482 (16.4)	336 (8.8)	1146 (22.0)
Peptic ulcer; n(%)	7 (0.1)	1 (0.0)	6 (0.1)
GI bleed req. hosp non variceal n(%)	5 (0.1)	2 (0.1)	3 (0.1)
Lower gastrointestinal			
Intestinal obstruction; n(%)	551 (6.1)	128 (3.3)	423 (8.1)
Fibrosing colonopathy/colonic structure; n(%)	1 (0.0)	1 (0.0)	0 (0.0)
Rectal prolapse; n(%)	28 (0.3)	25 (0.7)	3 (0.1)
Renal			
Kidney stones; n(%)	85 (0.9)	7 (0.2)	78 (1.5)
Renal failure; n(%)	18 (0.2)	2 (0.1)	16 (0.3)
Musculoskeletal			
Arthritis; n(%)	144 (1.6)	9 (0.2)	135 (2.6)
Arthropathy; n(%)	506 (5.6)	18 (0.5)	488 (9.4)
Bone fracture; n(%)	55 (0.6)	12 (0.3)	43 (0.8)
Osteopenia; n(%)	1085 (12.0)	22 (0.6)	1063 (20.4)
Osteoporosis; n(%)	469 (5.2)	7 (0.2)	462 (8.9)
Other			
Cancer confirmed by histology; n(%)	27 (0.3)	3 (0.1)	24 (0.5)
Port inserted or replaced; n(%)	548 (6.1)	220 (5.7)	328 (6.3)
Absence of vas deferens*; n(%)	670 (14.0)	3 (0.2)	667 (23.6)
Depression; n(%)	410 (4.5)	9 (0.2)	401 (7.7)
Hearing loss; n(%)	186 (2.1)	26 (0.7)	160 (3.1)
Hypertension; n(%)	200 (2.2)	4 (0.1)	196 (3.8)

* The denominator is restricted to male patients

For patients who are reported to have had non-tuberculous mycobacteria/atypical mycobacteria, cirrhosis (with/without portal hypertension), cancer or ABPA in 2013, we explored their clinical history to determine if this was the first year in which such a complication was reported. This historical search was not limited to annual review encounters and where no clinical history was available it is assumed that 2013 was the year the complication first developed.

1.19b Incidence of key complications

	Newly identified in 2012			Newly identified in 2013		
	Overall (n=8794)	<16 years (n=3732)	≥16 years (n=5062)	Overall (n=9052)	<16 years (n=3839)	≥16 years (n=5213)
Non-tuberculous mycobacteria or atypical mycobacteria; n(%)	159 (1.8)	30 (0.8)	129 (2.5)	134 (1.5)	33 (0.9)	101 (1.9)
ABPA; n(%)	169 (1.9)	64 (1.7)	105 (2.1)	157 (1.7)	58 (1.5)	99 (1.9)
Cirrhosis with no portal hypertension; n(%)	33 (0.4)	9 (0.2)	24 (0.5)	30 (0.3)	12 (0.3)	18 (0.3)
Cirrhosis with portal hypertension; n(%)	18 (0.2)	6 (0.2)	12 (0.2)	26 (0.3)	6 (0.2)	20 (0.4)
Cancer confirmed by histology; n(%)	10 (0.1)	1 (0.02)	9 (0.2)	13 (0.1)	1 (0.03)	12 (0.2)

1.20 CF-related diabetes

	All ≥10 years (n=6594)	10–16 years (n=1381)	≥16 years (n=5213)
Treatment for CF-related diabetes*; n(%)	1711 (26.0)	127 (9.2)	1584 (30.4)
Screening for CF-related diabetes Yes No Known CF-related diabetes Unknown	 3733 (56.6) 1017 (15.4) 1622 (24.6) 222 (3.4)	 1049 (76.0) 167 (12.1) 80 (5.8) 85 (6.2)	 2684 (51.5) 850 (16.4) 1542 (29.6) 137 (2.6)

*Treatment for CF-related diabetes was enquired about in an annual review questionnaire which was completed by 6585 of the 6594 patients aged 10 years and older with "complete" annual review encounter data. Among patients aged 10–16 years this represents 1379 patients and in patients 16 years and older 5206.

1.21 Transplants

	2009	2010	2011	2012	2013
Number of patients that year with annual review data evaluated for transplants	143	169	204	225	220
Number accepted on the transplant list	79	82	121	120	136
Number receiving transplants (<16) Types of transplants received: Bilateral lung Heart and lung Liver Other	3 3 0 0 0	3 2 0 1 0	3 3 0 0 0	3 2 0 1 0	3 2 0 1 0
Number receiving transplants (≥16) Types of transplants received: Bilateral lung Heart and lung Liver Other	22* 16 0 5 2	26 24 1 0 1	48* 40 4 2 3	52** 43 1 6 4	54* 48 0 3 4

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

1.22 Other therapy

	Overall (n=9052)	<16 years (n=3839)	≥16 years (n=5213)
NIV; n(%)	221(2.5)	24 (0.6)	197 (3.8)
Long-term oxygen; n(%) Among those who had long-term oxygen therapy: Continuously Nocturnal+exertion PRN With exacerbation	610 (6.8) 152 (24.9) 142 (23.3) 83 (13.6) 233 (38.2)	93 (2.4) 8 (8.6) 16 (17.2) 9 (9.7) 60 (64.5)	517 (10.0) 144 (27.9) 126 (24.4) 74 (14.3) 173 (33.5)

1.23 Feeding

	Overall (n=9052)	<16 years (n=3839)	≥16 years (n=5213)
Any supplemental feeding; n(%) Nasogastric tube Gastrostomy tube/Button Jejunal TPN	2826 (31.9) 110 548 9 9	1020 (27.5) 13 204 2 4	1806 (35.2) 97 344 7 5

1.24 Days on IV antibiotics in the last 12 months

	Home		Hospital		Total	
Age	N (%)	Median (IQR)	N (%)	Median (IQR)	N (%)	Median (IQR)
0–3	57 (5.8)	10 (7–13)	271 (27.7)	14 (8–21)	274 (28.0)	14 (12–26)
4–7	136 (13.5)	14 (10–26)	309 (30.8)	14 (7–21)	330 (32.9)	14 (14–29)
8–11	184 (20.5)	18 (12–38)	321 (35.8)	14 (9–28)	372 (41.4)	27 (14–44)
12–15	287 (30.1)	21 (12–35)	466 (48.9)	15 (9–42)	527 (55.3)	31 (14–56)
16–19	303 (30.2)	20 (12–38)	418 (41.7)	16 (9–39)	511 (51.0)	28 (14–54)
20–23	402 (40.4)	26 (14–42)	486 (48.9)	16 (10–40)	617 (62.1)	30 (14–56)
24–27	329 (39.4)	26 (14–42)	407 (48.7)	18 (11–39)	513 (61.4)	34 (14–59)
28–31	297 (42.2)	23 (14–43)	306 (43.5)	16 (9–35)	421 (59.9)	29 (14–56)
32–35	213 (42.5)	28 (14–43)	178 (35.5)	16 (11–35)	284 (56.7)	28 (14–56)
36–39	128 (40.6)	21 (13–42)	120 (38.1)	16 (9–30)	165 (52.4)	28 (14–52)
40–44	110 (31.2)	22 (14–49)	112 (31.7)	14 (9–26)	160 (45.3)	28 (14–49)
45–49	68 (28.6)	15 (12–43)	70 (29.4)	16 (11–44)	98 (41.2)	28 (14–59)
50+	56 (21.2)	19 (12–34)	84 (31.8)	15 (10–29)	103 (39.0)	22 (14–51)
Overall	2570 (28.4)	21 (13–41)	3548 (39.3)	15 (9–33)	4375 (48.3)	28 (14–52)

1.25a Nebulised drug treatment: DNase

Age	DNase treatment; n(%)		
	2008	2013	p-value (2008 vs 2013)
0–3	46 (7.6)	100 (10.2)	<0.001
4–7	125 (20.1)	332 (33.1)	<0.001
8–11	227 (34.2)	496 (55.2)	<0.001
12–15	359 (46.4)	627 (65.7)	<0.001
16–19	377 (49.5)	635 (63.2)	<0.001
20–23	319 (44.0)	625 (62.9)	<0.001
24–27	288 (47.6)	537 (64.2)	<0.001
28–31	182 (43.4)	413 (58.7)	<0.001
32–35	108 (41.5)	283 (56.3)	<0.001
36–39	83 (35.0)	157 (49.8)	0.001
40–44	147 (35.7)*	168 (47.6)	<0.001**
45–49		113 (47.1)	
50+		129 (48.9)	
Overall	2261 (37.2)	4615(51.0)	

* In 2008 all patients aged 40 years and older were grouped together.
** All patients aged 40 years and older were grouped together for this comparison.

1.25b Nebulised drug treatment: Hypertonic saline

Age	Hypertonic saline; n(%)		
	2008	2013	p-value (2008 vs 2013)
0–3	3 (0.5)	49 (5.0)	<0.001
4–7	15 (2.4)	157 (15.6)	<0.001
8–11	23 (3.5)	225 (25.0)	<0.001
12–15	32 (4.1)	303 (31.7)	<0.001
16–19	33 (4.3)	287 (28.6)	<0.001
20–23	50 (6.9)	263 (26.5)	<0.001
24–27	60 (9.9)	220 (26.3)	<0.001
28–31	37 (8.8)	206 (29.3)	<0.001
32–35	29 (11.2)	131 (26.0)	<0.001
36–39	16 (6.8)	76 (24.1)	<0.001
40–44	33 (8.0)*	75 (21.3)	<0.001**
45–49		56 (23.3)	
50+		69 (26.1)	
Overall	331 (5.4)	2117 (23.4)	

* In 2008 all patients aged 40 years and older were grouped together.
** All patients aged 40 years and older were grouped together for this comparison.

1.25c Inhaled antibiotic use among patients with chronic *Pseudomonas aeruginosa*

	2008			2013		
	Overall	<16 years	≥16 years	Overall	<16 years	≥16 years
Patients with chronic *P. aeruginosa*	2098	299	1799	2960	329	2631
Tobramycin solution; n(%)	412 (19.6)	48 (16.1)	364 (20.2)	929 (31.4)	103 (31.3)	826 (31.4)
Other aminoglycoside; n(%)	43 (2.0)	5 (0.2)	38 (2.1)	108 (3.6)	13 (4.0)	95 (3.6)
Colistin; n(%)	914 (43.6)	174 (58.2)	740 (41.1)	1173 (39.6)	176 (53.5)	997 (37.9)
Promixin; n(%)	490 (23.4)	73 (24.4)	417 (23.2)	881 (29.8)	140 (42.6)	741 (28.2)
Aztreonam; n(%)				201 (6.8)	2 (0.6)	199 (7.6)
At least one of the above*; n(%)	1597 (76.1)	257 (86.0)	1340 (74.5)	2368 (80.0)	302 (91.8)	2066 (78.5)

* In 2013, this includes Aztreonam.

The consensus view in the UK is that 90% of patients chronically infected with *P. aeruginosa* should be prescribed at least one of the above nebulised antibiotics.

1.25d Long-term use of azithromycin among patients with and without chronic *Pseudomonas aeruginosa*

	2008				2013			
	Overall (n=6082)	0–3 years (n=605)	4–15 years (n=2057)	≥16 years (n=3420)	Overall (n=9052)	0–3 years (n=981)	4–15 years (n=2858)	≥16 years (n=5213)
Patients with chronic *P. aeruginosa*	1246 (59.4)	2 (15.4)	105 (36.7)	1139 (63.3)	2022 (68.3)	2 (9.5)	141 (45.8)	1879 (71.4)
Patients without chronic *P. aeruginosa*	712 (22.1)	13 (2.7)	258 (17.3)	441 (35.3)	1597 (27.2)	25 (2.7)	479 (19.7)	1093 (43.5)

1.26 Physiotherapy techniques

	Overall (n=9052)	<16 years (n=3839)	≥16 years (n=5213)
Active cycle of breathing techniques; n(%)	3244 (36.1)	1820 (47.9)	1424 (27.5)
Autogenic drainage (including assisted autogenic drainage); n(%)	1400 (15.6)	265 (7.0)	1135 (21.9)
Any form of PEP; n(%)	4685 (52.2)	2388 (62.8)	2297 (44.4)
VEST; n(%)	171 (1.9)	90 (2.4)	81 (1.6)

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

Section 2: Analyses by paediatric care centre/clinic

(based on 4206 patients from paediatric care centres with complete* data at 2013 annual review)

* “Complete” data refers to the minimum data required to produce the range of clinical outcomes presented in this report.

How to interpret the graphs presented in Sections 2 and 3

Continuous outcomes such as age, BMI and FEV_1 in each centre are presented in the form of box plots. These graphs are commonly used to illustrate the spread of continuous measures in different groups.

Box plots in general are composed of a box, two whiskers, two adjacent values and some marker symbols for outside values. The lower border of the box denotes the first quartile, Q_1 (or 25th percentile); the upper border denotes the third quartile, Q_3 (or 75th percentile). The line in the middle of the box is the median (the 50th percentile). An upper whisker extends from the third quartile to the value that corresponds to the third quartile plus 1.5 times the inter-quartile range (Q_3+1.5xIQR). Likewise, a lower whisker extends from the first quartile to the value corresponding to the first quartile minus 1.5 times the inter-quartile range (Q_1-1.5xIQR). "Outside values" (or outliers) refer to values that are unusually distant from the rest of the data. For the report, we did not include the outside values as this would have created a great deal of spread.

When the data are normally distributed, the median lies in the middle of the box and the plot looks symmetrical. If the distribution is skewed then the median shifts towards the top or bottom of the box. In the picture below, the median is closer to Q_1, implying that the distribution is skewed to the right.

Reference: Kohler, U., Kreuter, F. (2012) Data Analysis Using Stata, STATA Press, Texas

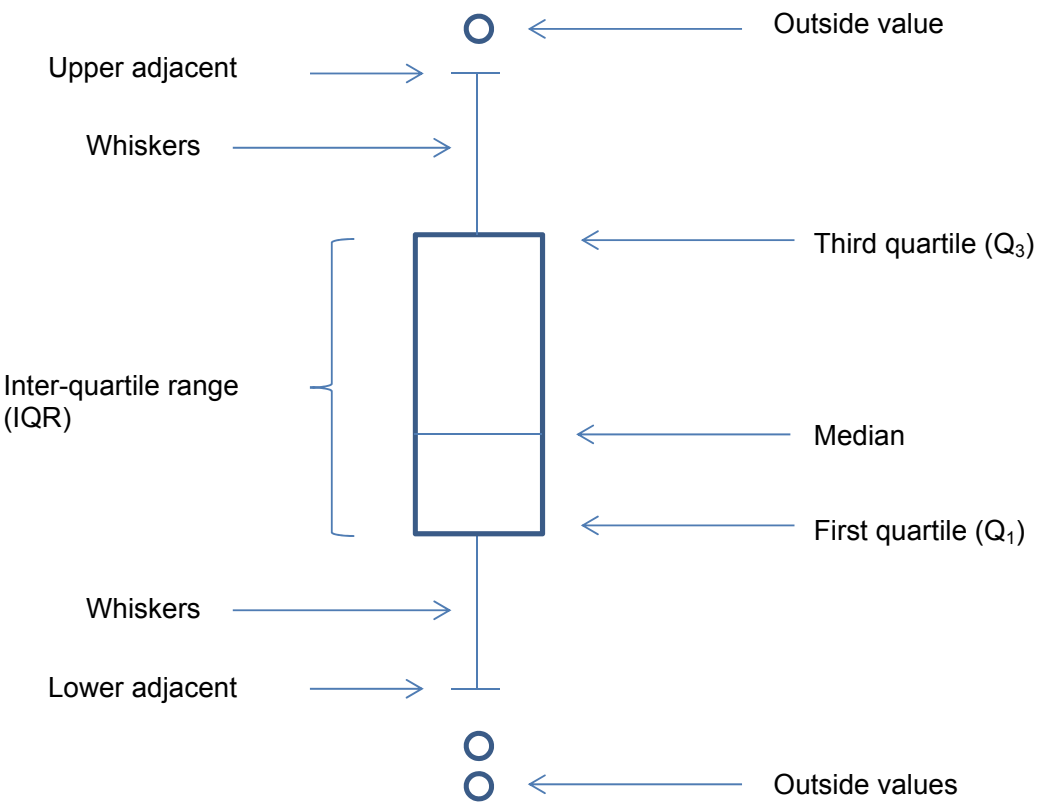
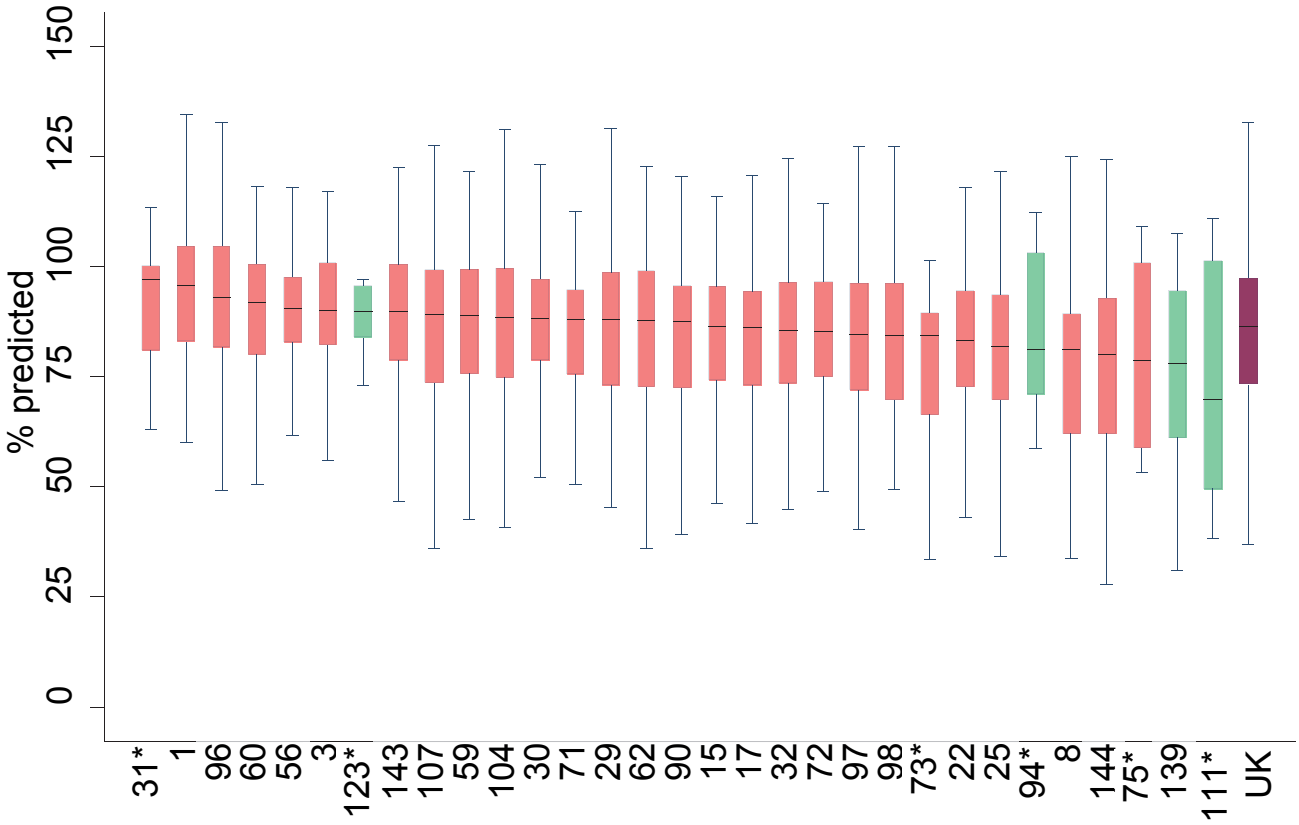


Figure 2.1 Median FEV_1 % predicted among patients aged 6 years and older by paediatric centre/clinic (without a history of lung transplant)



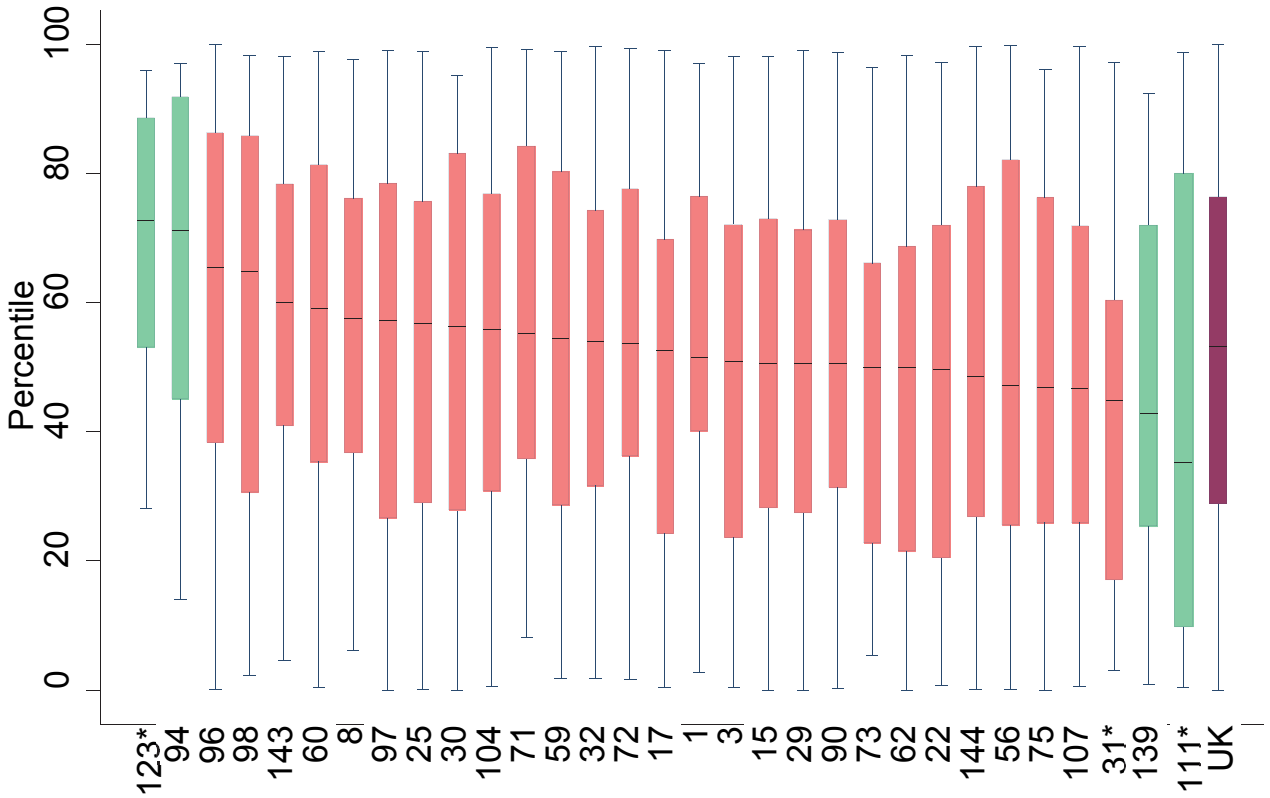
Excludes outside values

The median FEV_1 % predicted for patients attending paediatric centres/clinics is 87% predicted (IQR: 73–97).

Red: centres with their network clinics. Green: stand-alone clinics. Purple: all.

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

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



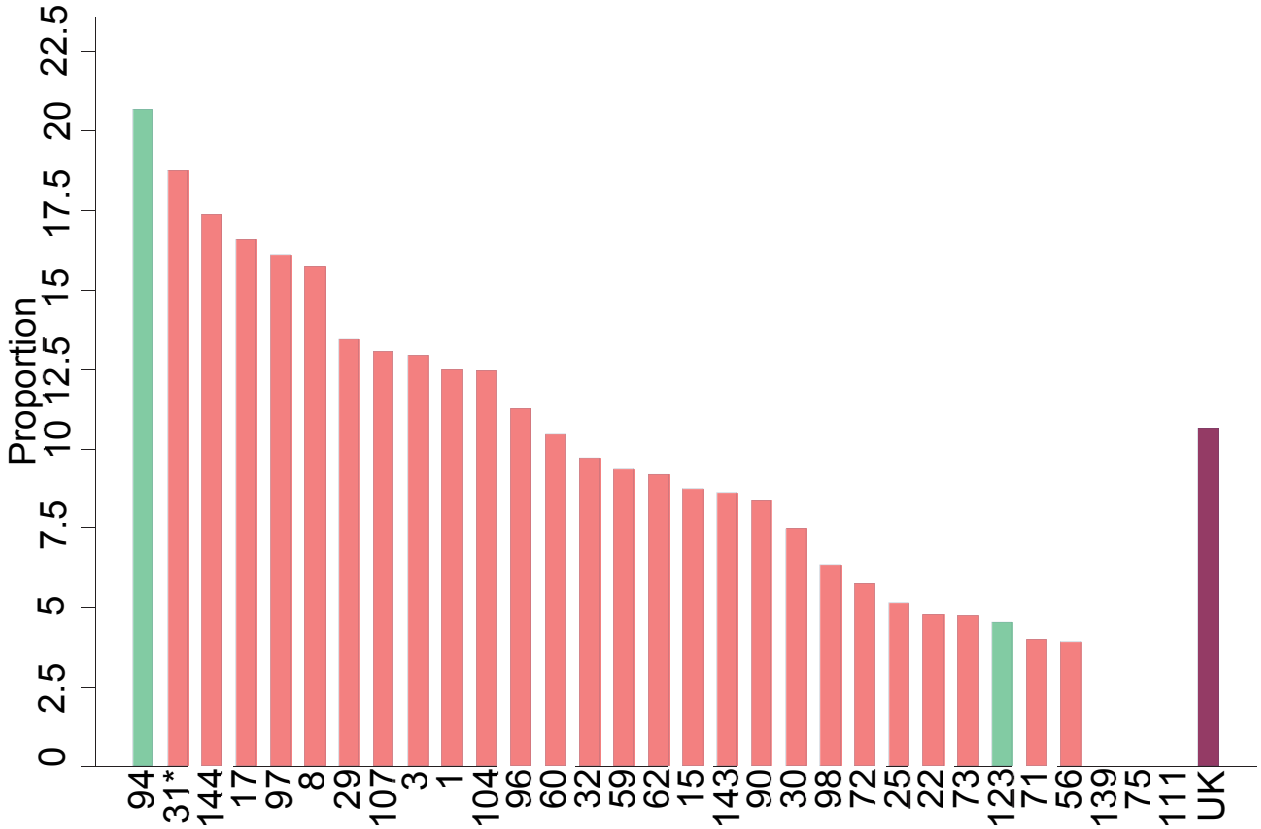
Excludes outside values

The median BMI percentile in paediatric centres/clinics is 53 (IQR: 29–76).

Red: centres with their network clinics. Green: stand-alone clinics. Purple: all.

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

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

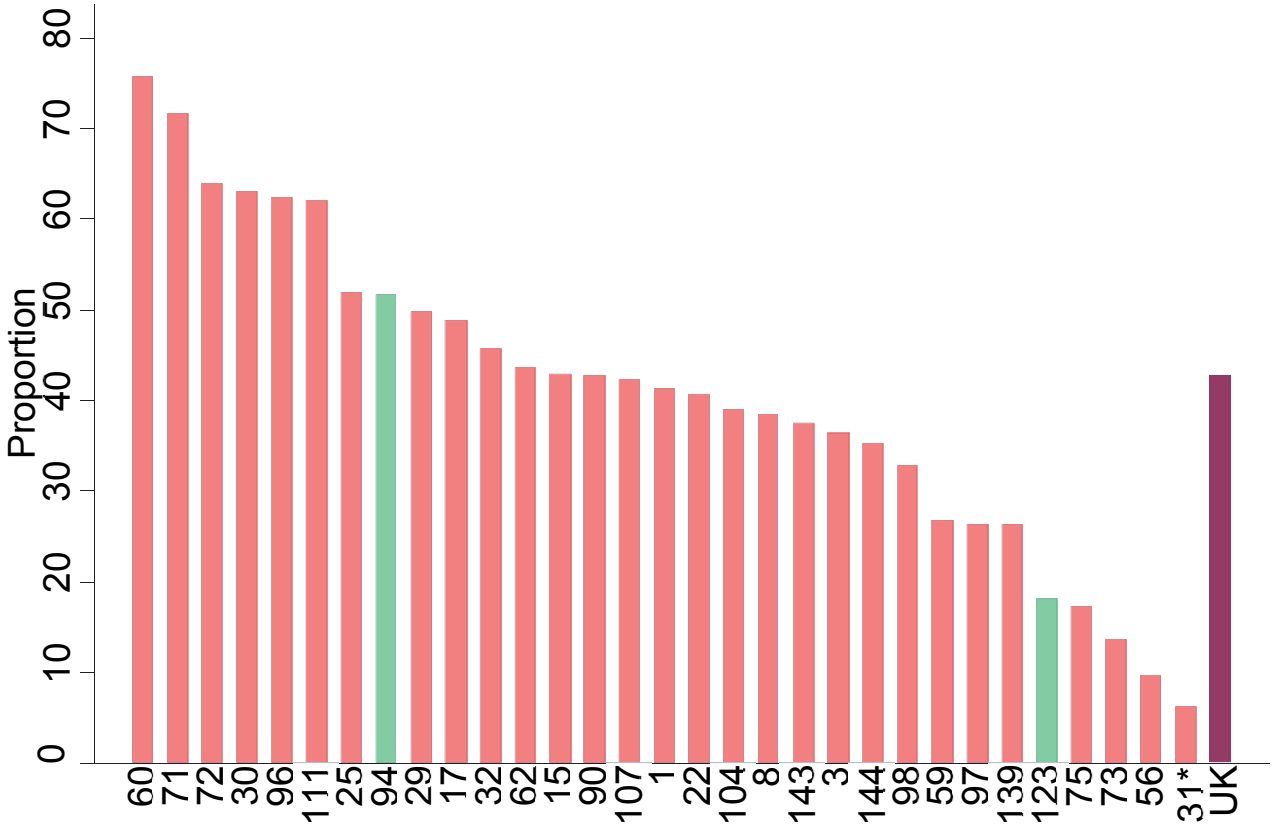


The proportion of patients with chronic *P. aeruginosa* in paediatric centres/clinics is 11%.

Red: centres with their network clinics. Green: stand-alone clinics. Purple: all.

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

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



The proportion of patients receiving DNase treatment in paediatric centres/clinics is 43%.

Red: centres with their network clinics. Green: stand-alone clinics. Purple: all.

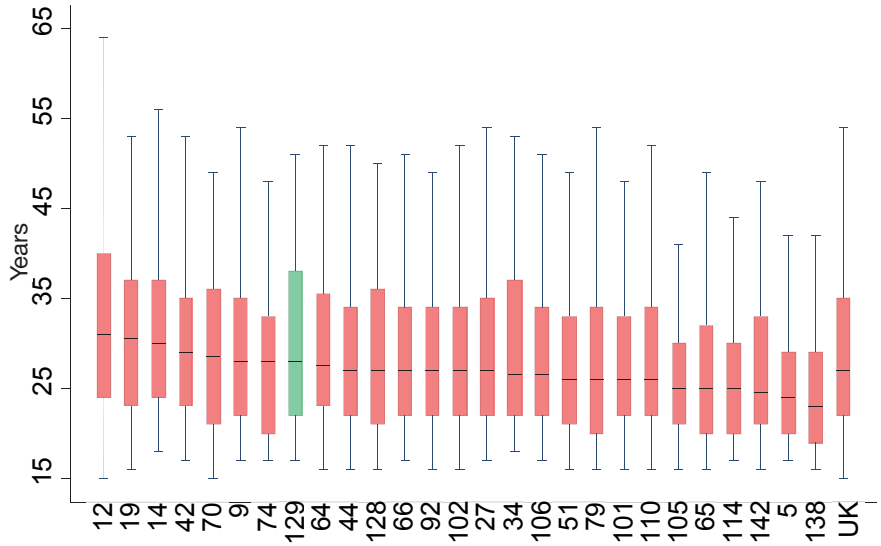
* Centre/clinic with a data set submission of fewer than 20 patients.

Section 3: Analyses by adult service

(based on 4846 patients from adult services with complete* data at 2013 annual review)

* “Complete” data refers to the minimum data required to produce the range of clinical outcomes presented in this report.

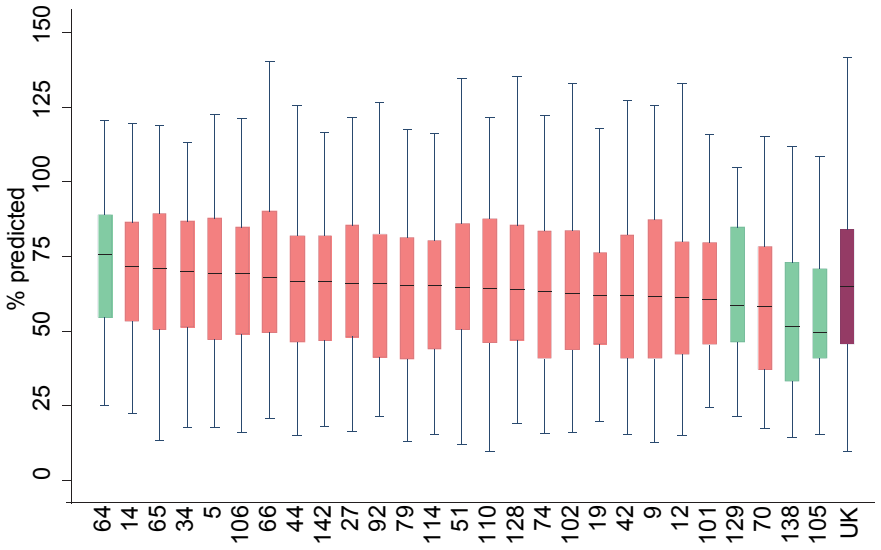
Figure 3.1 Median age (years) by adult service



Excludes outside values

The median age in adult services is 27 years (IQR: 22–34).
Red: centres. Green: other clinics. Purple: all.

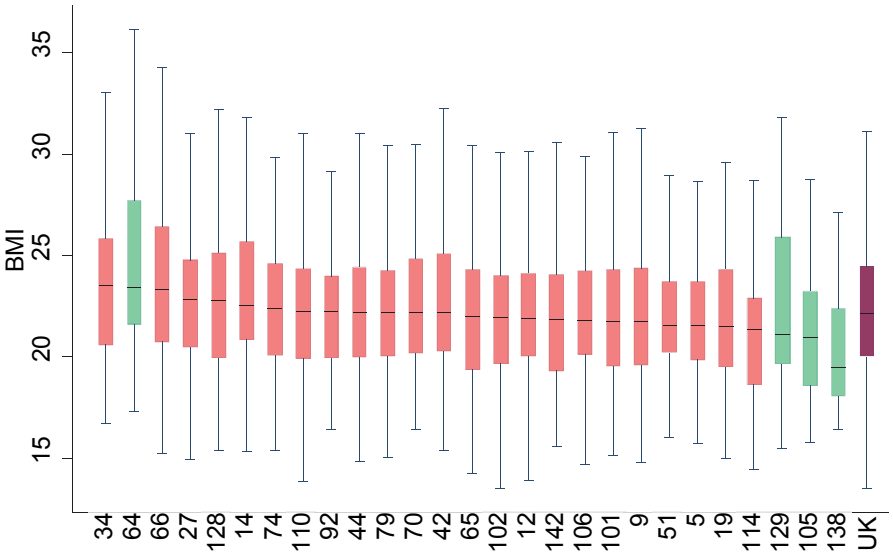
Figure 3.2 Median FEV_1 (% predicted) by adult service (without a history of lung transplant)



Excludes outside values

The median FEV_1 (% predicted) in adult services is 65% (IQR: 45–84).
Red: centres. Green: other clinics. Purple: all.

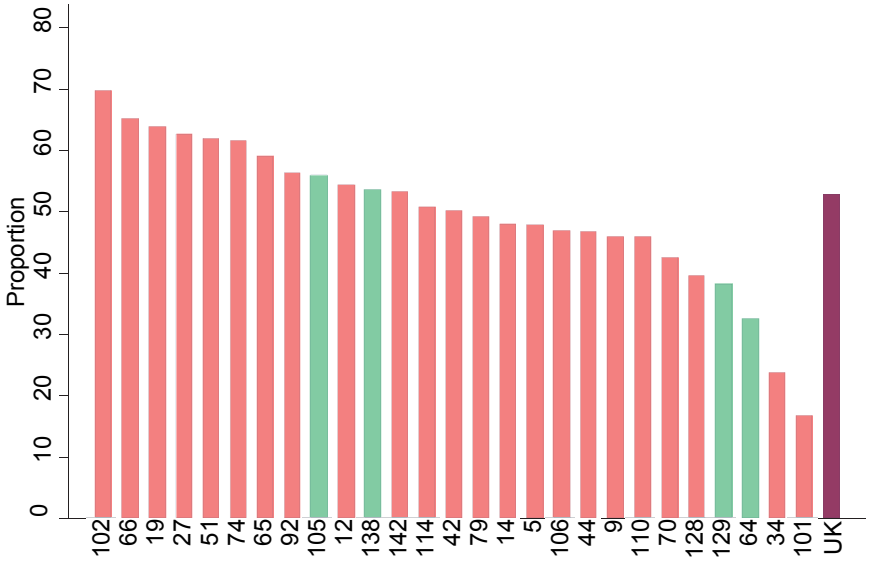
Figure 3.3 Median BMI among patients aged 16 years and older by adult service



Excludes outside values

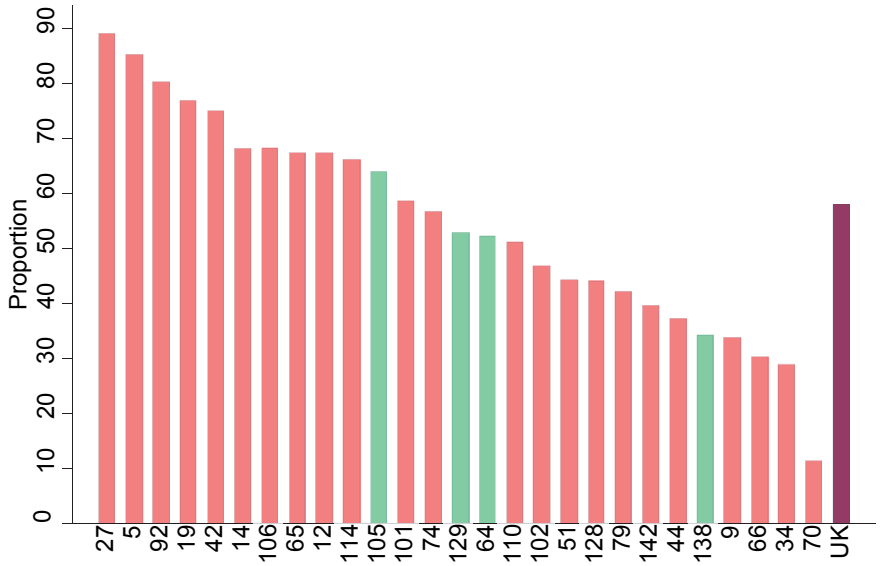
The median BMI in adult services is 22 (IQR: 20–24).
Red: centres. Green: other clinics. Purple: all.

Figure 3.4 Proportion of patients with chronic *P. aeruginosa* by adult service



The proportion of patients with chronic *P. aeruginosa* in adult services is 53%.
Red: centres. Green: other clinics. Purple: all.

Figure 3.5 Proportion of patients receiving DNase treatment by adult service



The proportion of patients receiving DNase treatment in adult services is 58%.
Red: centres. Green: other clinics. Purple: all.

Section 4: Care centres/clinics providing data in 2013

4.1 Paediatric centres/clinics providing data in 2013 – ordered by clinic ID

Country	Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2013	Median FEV_1 % predicted (≥6 years)	Median BMI percentile (2–15 years)
England	Leicester	Leicester Royal Infirmary	1	58	58	95.7	51.6
England	Sheffield	Sheffield Children's Hospital	3	141	140	90.0	50.9
England	Stoke	University Hospital of North Staffordshire	8	95	91	81.1	57.5
England	London – South West	Royal Brompton Hospital	15	319	312	86.5	50.7
England	London	King's College Hospital	17	190	188	86.0	52.5
England	Oxford	John Radcliffe Hospital	22	171	167	83.4	49.7
England	Leeds	St James's University Hospital	25	236	233	81.9	56.8
England	Southampton	Southampton General Hospital	29	216	211	87.9	50.7
England	London – East	Royal London Hospital	30	116	111	88.1	56.3
Scotland	Inverness	Raigmore Hospital	31	16	16	97.2	44.7
England	Bristol	Bristol Royal Hospital for Children	32	175	175	85.5	54.0
Scotland	Glasgow	Royal Hospital for Sick Children	56	133	123	90.7	47.1
England	Newcastle	Royal Victoria Infirmary	59	188	168	89.0	54.4
Northern Ireland	Belfast	Royal Belfast Hospital for Sick Children	60	202	194	91.9	59.0
England	Nottingham	Nottingham Children's Hospital	62	175	174	87.7	49.9

Country	Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2013	Median FEV_1 % predicted (≥6 years)	Median BMI percentile (2–15 years)
England	Teesside	James Cook University Hospital	71	55	53	87.9	55.3
Wales	Cardiff	Children's Hospital for Wales	72	180	164	85.1	53.7
Scotland	Dundee	Ninewells Hospital	73	23	22	84.3	50.0
Scotland	Aberdeen	Royal Aberdeen Children's Hospital	75	29	29	78.7	47.0
England	London – Central	Great Ormond Street Hospital for Children	90	181	180	87.5	50.5
England	Truro	Royal Cornwall Hospital	94	30	29	81.2	71.3
England	Exeter	Royal Devon & Exeter Hospital	96	74	72	93.1	65.5
England	Liverpool	Alder Hey Children's Hospital	97	305	300	84.5	57.3
England	Norwich	Norfolk & Norwich University Hospital	98	64	64	84.4	64.9
England	Birmingham	Birmingham Children's Hospital	104	287	282	88.5	55.8
England	Cambridge	Addenbrookes Hospital	107	132	130	89.1	46.6
England	Hull	Hull Royal Infirmary	111	29	29	69.8	35.4
Scotland	Ayr/ Kilmarnock	Crosshouse Hospital	123	22	22	89.9	72.8
England	Plymouth	Derriford Hospital	139	38	38	78.0	42.8
Scotland	Edinburgh	Royal Hospital for Sick Children	143	118	117	89.8	60.1
England	Manchester	Royal Manchester Children's Hospital	144	326	314	80.0	48.6

4.2 Adult centres/clinics providing data in 2013 – ordered by clinic ID

Country	Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2013	Median FEV_1 % predicted (≥16 years)	Median BMI (≥16 years)
England	London – South East	King's College Hospital	5	182	169	69.4	21.5
England	Newcastle	Royal Victoria Infirmary	9	255	248	61.8	21.7
England	London – South West	Royal Brompton Hospital	12	656	643	61.1	21.9
Northern Ireland	Belfast	Belfast City Hospital	14	231	205	71.5	22.5
England	Frimley	Frimley Park Hospital	19	117	108	62.0	21.5
England	Birmingham	Birmingham Heartlands Hospital	27	350	339	66.0	22.8
England	Exeter	Royal Devon & Exeter Hospital	34	89	86	70.0	23.5
England	Leeds	St James's University Hospital	42	416	409	62.0	22.2
Scotland	Edinburgh	Western General Hospital	44	219	212	66.7	22.2
England	Cambridge	Papworth Hospital	51	273	235	64.7	21.5
England	Plymouth	Derriford Hospital	64	48	44	75.6	23.4
England	Sheffield	Northern General Hospital	65	169	166	71.2	22.0
England	Liverpool	Liverpool Heart and Chest Hospital	66	272	257	68.1	23.3
Scotland	Aberdeen	Aberdeen Royal Infirmary	70	64	62	57.0	22.2
England	Stoke-on-Trent	University Hospital of North Staffordshire	74	69	67	63.5	22.4
Scotland	Glasgow	Gartnavel General Hospital	79	221	206	65.3	22.2

Country	Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2013	Median FEV_1 % predicted (≥16 years)	Median BMI (≥16 years)
England	London – East	London Chest Hospital	92	151	127	65.9	22.2
England	Nottingham	Nottingham City Hospital	101	143	133	60.7	21.8
England	Manchester	Wythenshawe Hospital	102	387	376	62.7	21.9
England	London – South East	University Hospital Lewisham	105	52	50	49.4	20.9
England	Bristol	Bristol Royal Infirmary	106	187	182	69.3	21.8
England	Southampton	Southampton General Hospital	110	230	211	64.2	22.3
England	Norwich	Norfolk & Norwich University Hospital	114	65	65	65.0	21.4
England	Oxford	Churchill Hospital	128	98	93	64.0	22.8
England	Truro	Royal Cornwall Hospital	129	35	34	58.5	21.1
England	Hull	Castle Hill Hospital	138	42	41	51.4	19.5
England	Leicester	Glenfield Hospital	142	78	78	66.6	21.9

4.3 Paediatric centres/clinics providing data in 2013 – ordered alphabetically

Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2013	Median FEV_1 % predicted (≥6 years)	Median BMI percentile (2–15 years)
England						
Birmingham	Birmingham Children's Hospital	104	287	282	88.5	55.8
Bristol	Bristol Royal Hospital for Children	32	175	175	85.5	54.0
Cambridge	Addenbrookes Hospital	107	132	130	89.1	46.6
Exeter	Royal Devon & Exeter Hospital	96	74	72	93.1	65.5
Hull	Hull Royal Infirmary	111	29	29	69.8	35.4
Leeds	St James's University Hospital	25	236	233	81.9	56.8
Leicester	Leicester Royal Infirmary	1	58	58	95.7	51.6
Liverpool	Alder Hey Children's Hospital	97	305	300	84.5	57.3
London – Central	Great Ormond Street Hospital for Children	90	181	180	87.5	50.5
London – East	Royal London Hospital	30	116	111	88.1	56.3
London – South East	King's College Hospital	17	190	188	86.0	52.5
London – South West	Royal Brompton Hospital	15	319	312	86.5	50.7
Manchester	Royal Manchester Children's Hospital	144	326	314	80.0	48.6
Newcastle	Royal Victoria Infirmary	59	188	168	89.0	54.4
Norwich	Norfolk & Norwich University Hospital	98	64	64	84.4	64.9
Nottingham	Nottingham Children's Hospital	62	175	174	87.7	49.9
Oxford	John Radcliffe Hospital	22	171	167	83.4	49.7
Plymouth	Derriford Hospital	139	38	38	78.0	42.8
Sheffield	Sheffield Children's Hospital	3	141	140	90.0	50.9
Southampton	Southampton General Hospital	29	216	211	87.9	50.7
Stoke	University Hospital of North Staffordshire	8	95	91	81.1	57.5

Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2013	Median FEV_1 % predicted (≥6 years)	Median BMI percentile (2–15 years)
Teesside	James Cook University Hospital	71	55	53	87.9	55.3
Truro	Royal Cornwall Hospital	94	30	29	81.2	71.3
Northern Ireland						
Belfast	Royal Belfast Hospital for Sick Children	60	202	194	91.9	59.0
Scotland						
Aberdeen	Royal Aberdeen Children's Hospital	75	29	29	78.7	47.0
Ayr/ Kilmarnock	Crosshouse Hospital	123	22	22	89.9	72.8
Dundee	Ninewells Hospital	73	23	22	84.3	50.0
Edinburgh	Royal Hospital for Sick Children	143	118	117	89.8	60.1
Glasgow	Royal Hospital for Sick Children	56	133	123	90.7	47.1
Inverness	Raigmore Hospital	31	16	16	97.2	44.7
Wales						
Cardiff	Children's Hospital for Wales	72	180	164	85.1	53.7

4.4 Adult centres/clinics providing data in 2013 – ordered alphabetically

Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2013	Median FEV_1 % predicted (≥16 years)	Median BMI (≥16 years)
England						
Birmingham	Birmingham Heartlands Hospital	27	350	339	66.0	22.8
Bristol	Bristol Royal Infirmary	106	187	182	69.3	21.8
Cambridge	Papworth Hospital	51	273	235	64.7	21.5
Exeter	Royal Devon & Exeter Hospital	34	89	86	70.0	23.5
Frimley	Frimley Park Hospital	19	117	108	62.0	21.5
Hull	Castle Hill Hospital	138	42	41	51.4	19.5
Leeds	St James's University Hospital	42	416	409	62.0	22.2
Leicester	Glenfield Hospital	142	78	78	66.6	21.9
Liverpool	Liverpool Heart and Chest Hospital	66	272	257	68.1	23.3
London – East	London Chest Hospital	92	151	127	65.9	22.2
London – South East	King's College Hospital	5	182	169	69.4	21.5
London – South East	University Hospital Lewisham	105	52	50	49.4	20.9
London – South West	Royal Brompton Hospital	12	656	643	61.1	21.9
Manchester	Wythenshawe Hospital	102	387	376	62.7	21.9
Newcastle	Royal Victoria Infirmary	9	255	248	61.8	21.7
Norwich	Norfolk & Norwich University Hospital	114	65	65	65.0	21.4
Nottingham	Nottingham City Hospital	101	143	133	60.7	21.8
Oxford	Churchill Hospital	128	98	93	64.0	22.8
Plymouth	Derriford Hospital	64	48	44	75.6	23.4
Sheffield	Northern General Hospital	65	169	166	71.2	22.0
Southampton	Southampton General Hospital	110	230	211	64.2	22.3
Stoke-on-Trent	University Hospital of North Staffordshire	74	69	67	63.5	22.4
Truro	Royal Cornwall Hospital	129	35	34	58.5	21.1

Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2013	Median FEV_1 % predicted (≥16 years)	Median BMI (≥16 years)
Northern Ireland						
Belfast	Belfast City Hospital	14	231	205	71.5	22.5
Scotland						
Aberdeen	Aberdeen Royal Infirmary	70	64	62	57.0	22.2
Edinburgh	Western General Hospital	44	219	212	66.7	22.2
Glasgow	Gartnavel General Hospital	79	221	206	65.3	22.2

Section 5: UK CF Registry Steering Committee

5.1 Composition of UK CF Registry Steering Committee

Professor Diana Bilton (Chair)	Adult CF Centre Director, Royal Brompton Hospital, London
Dr Caroline Elston	Adult CF Centre Director, King's College Hospital, London
Dr Iolo Doull	Paediatric CF Centre Director, Cardiff Hospital, Wales
Dr Siobhan Carr	Paediatrician, Royal Brompton Hospital, London
Dr Steve Cunningham	Paediatrician, Edinburgh Royal Infirmary, Scotland
Dr Martin Wildman	Adult CF Centre Director, Northern General Hospital, Sheffield
Professor Stuart Elborn	Adult CF Centre Director, Belfast, NI and Trustee of the Trust
Dr Stephanie MacNeill	Biostatistician, Imperial College, London
Mr George Vamvakas	Biostatistician, Imperial College, London
Mrs Marian Dmochowska	Parent Representative
Mr Dominic Kavanagh	Patient Representative
Ms Katherine Collins	Director NSD, Scotland
Ms Carrie Gardner	Specialist Commissioner, NHS England
Dr Kim Cox	Lead Specialist CF Commissioner, London
Dr Lisa Davies	Specialist Commissioner, Wales
Mr Ed Owen	Chief Executive, Cystic Fibrosis Trust
Dr Janet Allen	Director of Research, Cystic Fibrosis Trust
Ms Elaine Gunn	Registry Manager, Cystic Fibrosis Trust

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