Cystic Fibrosis our focus

UK Cystic Fibrosis Registry
Annual data report 2012

September 2013

Executive Summary

We are delighted to present the annual report of the UK Cystic Fibrosis (CF) Registry from data entered in 2012.

The Registry is a vital tool for improving care for people with CF in the UK made possible by the important collaboration between the CF Trust, NHS, clinicians, allied health professionals, and people with cystic fibrosis and their families.

This latest report demonstrates there are now 10,078 people with CF registered; 274 of these were newly registered in 2012, of which 157 were diagnosed on newborn screening. Over 57% of the patients registered are over 16 and there are now over 800 people with CF on the Registry aged over 40.

Over the last few years we have worked hard with the CF community to ensure all patients with CF have their complete data, including genotype, lung function, height and weight, captured on the Registry to ensure robust data on which to base planning and assessment of CF care, epidemiological studies and key trials of new therapies.

Indeed, for the first year, we have published a summary version of the data report, to present the findings in a more user-friendly form to ensure people with cystic fibrosis and those who care for them, as well as others, also have full access to this information.

The Registry data was particularly useful in the last year as a tool to aid the introduction of ivacaftor (Kalydeco), the first mutation specific therapy for CF. The Registry gave commissioners accurate figures on numbers of patients with the G551D mutation and provide data to those responsible for constructing health economic models in order to calculate the impact of the introduction of the new therapy.

The Registry is also utilised in peer review visits designed to ensure that care in CF centres is delivered in line with designated standards. We are pleased to see data on chronic *Pseudomonas aeruginosa* (PA) infection (Fig 1.18) showing that significantly less people with CF between age 16 and 23 have chronic PA infection in 2012 compared to 2008. This suggests that implementation of strategies of aggressive early treatment of *Pseudomonas* infection coupled with adherence to segregation guidelines have been successful in reducing chronic PA infection. In the future we hope this will lead to better outcomes and lower requirements for intravenous antibiotic therapy.

The 2012 data also demonstrates a rise in rates of therapies designed to maintain lung function and reduce exacerbations. Use of DNase has increased from 2008 to 2012 (Fig 1.25). Furthermore, centres are clearly adhering to guidelines recommending treatment of chronic PA infection with inhaled antibiotic therapy. In 2012, 90.2% of children with chronic PA infection were receiving antibiotic therapy via the inhaled route. The figure in adults is lower at 79% perhaps reflecting choice of patients

in avoiding the burden of nebulised therapy. It will be helpful to monitor these figures as options of antibiotics in dry powder inhaled form as an alternative to nebulised therapy have been made available and approved by NICE.

This large dataset also allows us to calculate median survival of the registered patient cohort and this year we see the median survival reaching 43.5 years, a steadily improving picture in keeping with registry data from around the world.

However, there are still a significant number of people with CF who die in their third decade, and there is still much to do in identifying and treating the factors responsible for decline in health in young adults. We expect researchers to utilise the Registry data in this endeavour and we hope to see further shifts in lung function and median age of death over the next few years.

The Annual Data Report 2012: Summary can be downloaded from the Cystic Fibrosis Trust's website at: cysticfibrosis.org.uk/registry

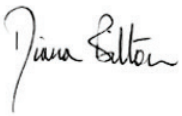
The number of patients receiving bilateral lung transplants is similar to 2011 at 45 (a definite improvement on 29 in 2012) but still less than needed as patients continue to die waiting for donor organs. It has been an important goal of the Registry to help identify the best outcomes for patients with CF and to ensure that all patients have access to the best care possible.

We are delighted that the National Institute for Health Research (NIHR) has awarded a grant to a multidisciplinary team led by Dr Stephanie MacNeill and Professor Paul Cullinan from Imperial College London (who provide the statistical and epidemiological input for this annual report) to utilise a new way of comparing Registry data in centre outcomes in order to truly identify best care. We look forward to that research changing our presentation of centre comparisons in future reports.

The Registry now provides a powerful tool for tracking the effects of new CF therapies. We have begun producing pharmacovigilence data for the European Medicines Agency in relation to new therapies and can also provide feedback to health commissioners on the effects of outcomes within the population treated.

The success of the Registry relies on the robust data submitted by the teams in centres caring for people with CF, the consent of all adults with CF and parents of children with CF, and the generous financial support of the wider cystic fibrosis community.

It is a tribute to all involved that the Registry has become an integral part of CF care in the UK, and we look forward to further development of the Registry to help deliver further improvements to the lives of people with CF



Dr Diana Bilton
Chair, Registry Steering Committee

Ed Owen
Chief Executive,
Cystic Fibrosis Trust

Contents

Section 1: All UK patients

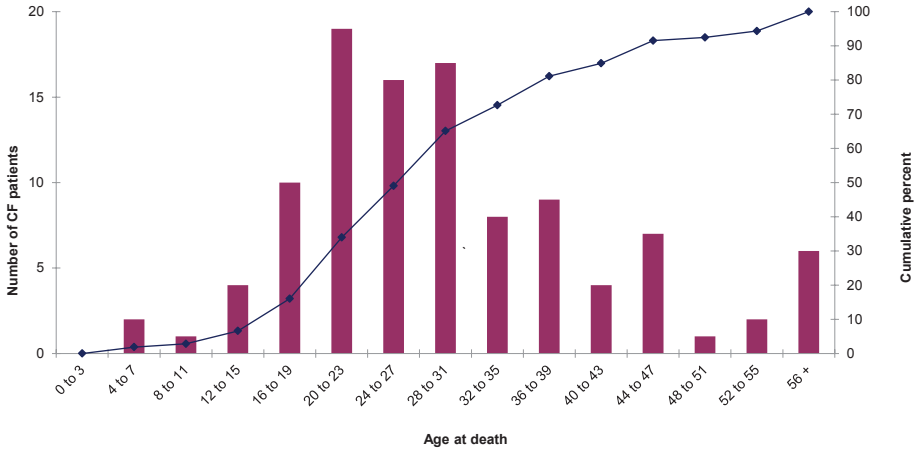
1.1 Summary of the UK Cystic Fibrosis Registry

	2008	2009	2010	2011	2012
CF patients registered Excluding 2012 diagnoses	8513[1]	9029[1]	9385[1]	9749[1]	10078[1] 9804
CF patients with "complete" data; n(%) Rate of completeness excluding 2012 diagnoses	6082[2] (71%)	7377[2] (82%)	7937[2] (85%)	8679[2] (89%)	8794[2] (87%) 90%
Age in years; median	18[3]	17[3]	17[3]	18[3]	18[3]
All newly diagnosed patients (newborn screening and other)	235[4]	261[4]	301[4]	261[4]	274[4]
Newly diagnosed patients identified through newborn screening			189	155	157
Age at diagnosis in months; median	4[3]	3[3]	3[3]	3[3]	3[3]
Adults aged 16 yrs and over; %	56.2[3]	55.1[3]	55.5[3]	56.8[3]	57.6[3]
Males; %	53.3[3]	53.1[3]	53.1[3]	53.2[3]	52.9[3]
Genotyped; %	93.7[3]	94.3[3]	95.2[3]	95.6[3]	96.2[3]
Median predicted survival in years (95% confidence interval)	38.8[5] (34.2, 47.3)	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)
Total deaths reported	100	141	103	118	106
Age at death in years; median (95% CI)[6]	27	27	29	26	28 (25, 29)

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 2009/2010/2011/2012/2013; and
   - had no recorded date of death before 1 January 2008/2009/2010/2011/2012
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 2012



There were 106 recorded deaths in 2012. The median age at death was 28 years (min = 5 yrs; max = 72 years; 95% confidence interv There were 106 recorded deaths in 2012. The median age at death was 28 years (min = 5 yrs; max = 72 years; 95% confidence interval: 25–29 years).
al: 25–29 years).

Analyses based on 8794 patients with complete* data at 2012 annual review

1.3 Age at diagnosis and screening statistics among children

Age at diagnosis	All patients; n (%)	Patients aged 10 years in 2012; n(%)	Patients aged 5 years in 2012; n(%)
Pre-natal	0	0	0
Birth to 3 months	2567 (70.0)	107 (51.9)	181 (71.8)
4–6 months	230 (6.3)	14 (6.8)	13 (5.2)
7–12 months	177 (4.8)	18 (8.7)	12 (4.8)
1 yr	243 (6.6)	22 (10.7)	20 (7.9)
2 yrs	159 (4.3)	18 (8.7)	13 (5.2)
3 yrs	91 (2.5)	9 (4.4)	6 (2.4)
4 yrs	64 (1.7)	5 (2.4)	6 (2.4)
5 yrs	37 (1.0)	2 (1.0)	1 (0.4)
6 yrs	24 (0.7)	1 (0.5)	-
7 yrs	18 (0.5)	3 (1.5)	-
8 yrs	25 (0.7)	6 (2.9)	-
9 yrs	12 (0.3)	0	-
10 yrs	8 (0.2)	1 (0.5)	-
11 yrs	5 (0.1)	-	-
12 yrs	2 (0.1)	-	-
13 yrs	6 (0.2)	-	-
14 yrs	1 (0.03)	-	-
15 yrs	1 (0.03)	-	-

The median (range) age at diagnosis is 1 month (0–15 years).

Diagnosis in the first three months of life was more common in children aged 5 years in 2012 (born in 2007) than in children aged 10 years in 2012 (born in 2002).

Of the 63 children with complete data born in 2012, 50 (79%) were identified by newborn screening.

In 2012, a total of 157 patients were identified by newborn screening (including patients with and without complete data). In 2011 this figure was 155 and in 2010 it was 189.

* “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	0
Birth to 3 months	1967 (39.5)
4–6 months	494 (9.9)
7–12 months	318 (6.4)
1 yr	427 (8.6)
2 yrs	269 (5.4)
3 yrs	191 (3.8)
4 yrs	162 (3.3)
5 yrs	83 (1.7)
6 yrs	72 (1.4)
7 yrs	48 (1.0)
8 yrs	58 (1.2)
9 yrs	48 (1.0)
10 yrs	37 (0.7)
11 yrs	36 (0.7)
12 yrs	38 (0.8)
13 yrs	41 (0.8)
14 yrs	33 (0.7)
15 yrs	39 (0.8)
16–20 yrs	146 (2.9)
21–25 yrs	95 (1.9)
26–30 yrs	86 (1.7)
31–35 yrs	89 (1.8)
36–40 yrs	66 (1.3)
41–45 yrs	50 (1.0)
46–50 yrs	28 (0.6)
51–60 yrs	38 (0.8)
61 yrs+	25 (0.5)

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

Of the 5062 adults with complete data in 2012, 378 were diagnosed by neonatal screening and 24 adults were diagnosed in 2012.

1.5 Genotyping

8462 (96.2%) patients have been genotyped with a recorded value.

DF508 Mutations; n (%)	
Homozygous DF508	4371 (51.7%)
Heterozygous DF508	3300 (39.0%)
No DF508 or both unidentified	791 (9.4%)

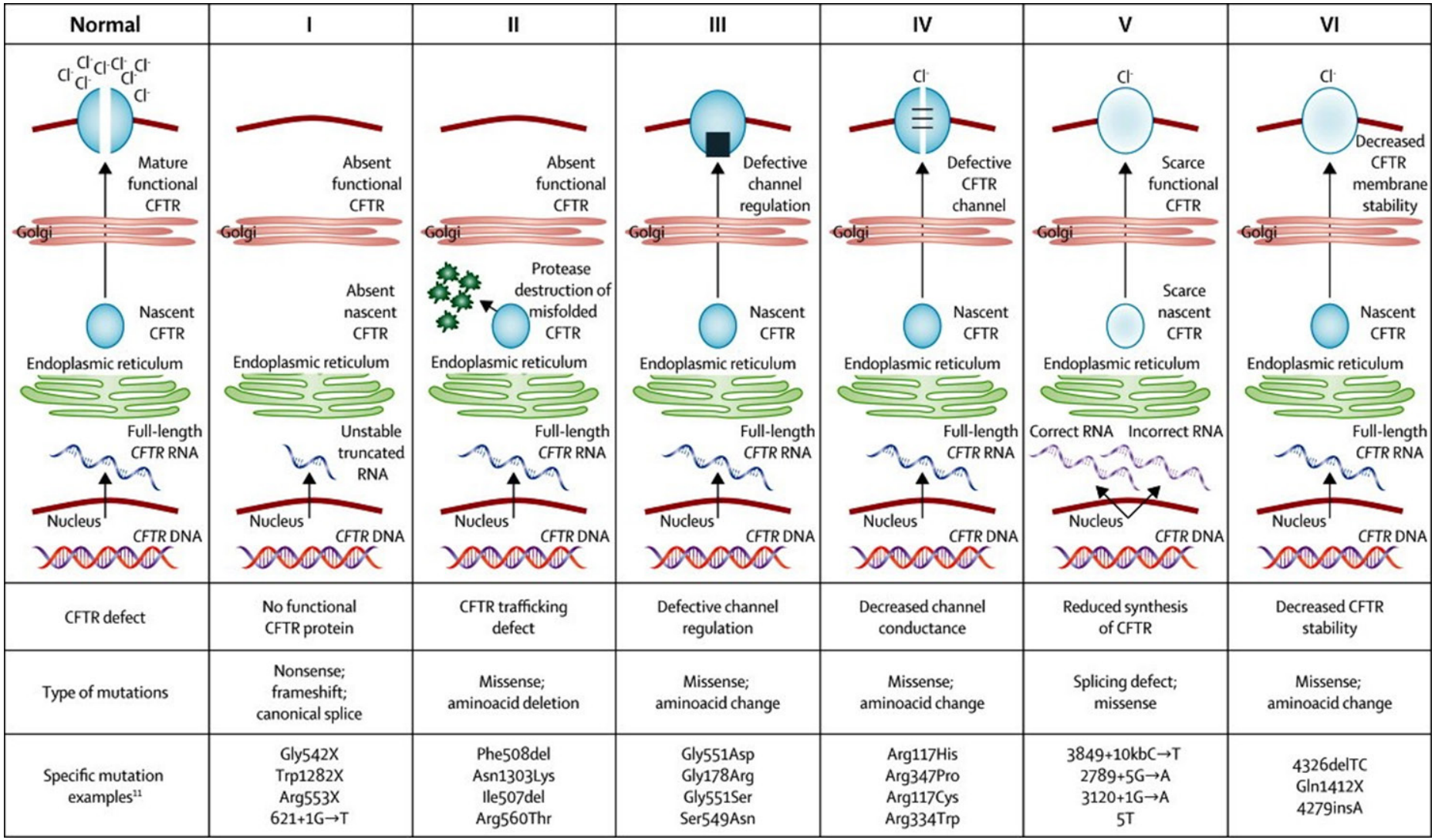
908 (10.7%) patients have at least one unknown genotype.

All mutations and their classes

All mutations Current name	New name	Class	N (%)
DF508	p.Phe508del	II	7671 (90.7)
G551D	p.Gly551Asp	III	471 (5.6)
R117H	p.Arg117His	IV	361 (4.3)
G542X	p.Gly542X	I	307 (3.6)
621+1G->T	c.489+1G>T	I	180 (2.1)
N1303K	p.Asn1303Lys	II	111 (1.3)
1717-1G->A	c.1585-1G>A	I	106 (1.3)
1898+1G->A	c.1766+1G>A	I	95 (1.1)
DI507	p.Ile507del	II	85 (1.0)
R560T	p.Arg560Thr	III	83 (1.0)
3659delC	c.3528delC	II	82 (1.0)
R553X	p.Arg553X	I	71 (0.8)
3849+10kbC->T	c.3717+10kbC>T	V	64 (0.8)
Q493X	p.Gln493X	I	58 (0.7)
G85E	p.Gly85Glu	IV	56 (0.7)
E60X	p.Glu60X	I	54 (0.6)
D1152H	p.Asp1152His	IV	54 (0.6)
W1282X	p.Trp1282X	I	45 (0.5)
1078delT	c.948delT	II	37 (0.4)
2789+5G->A	c.2657+5G>A	V	30 (0.4)
2184delA	c.2052delA	II	29 (0.3)
V520F	p.Val520Phe	III	29 (0.3)
R347P	p.Arg347Pro	IV	26 (0.3)
R1162X	p.Arg1162X	I	23 (0.3)
A455E	p.Ala455Glu	V	21 (0.3)

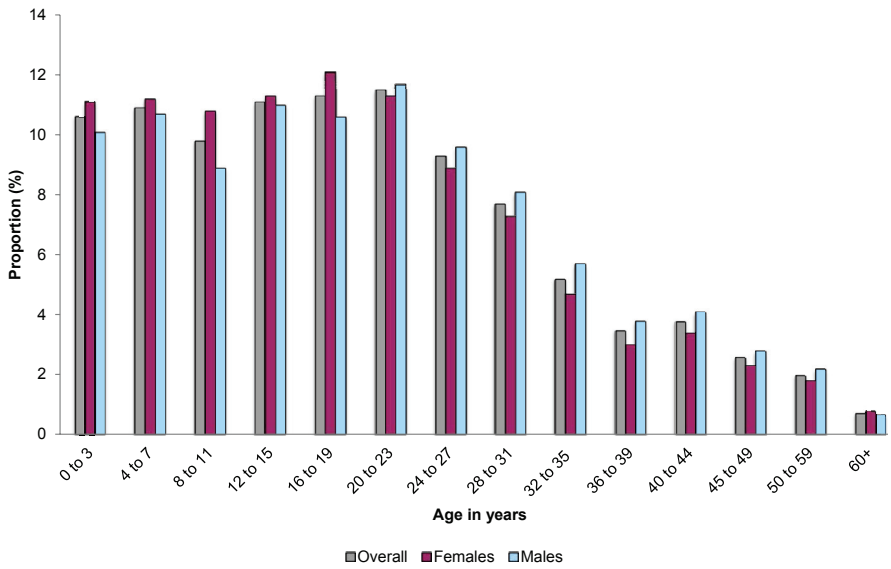
All mutations Current name	New name	Class	N (%)
S549N	p.Ser549Asn	II	21 (0.3)
R347H	p.Arg347His	IV	14 (0.2)
R1158X	p.Arg1158X	I	14 (0.2)
711+1G->T	c.579+1G>T	I	11 (0.1)
3120+1G->A	c.2988+1G>A	I	10 (0.1)
R334W	p.Arg334Trp	IV	10 (0.1)
L206W	p.Leu206Trp	unknown	10 (0.1)
R352Q	p.Arg352Gln	unknown	9 (0.1)
1161delC	c.1029delC	I	7 (0.1)
I148T	p.Ile148Thr	V	7 (0.1)
R1066C	p.Arg1066Cys	unknown	7 (0.1)
Y1092X	p.Tyr1092X	I	6 (0.1)
2183delAA->G	c.2051_2052delAAinsG	I	5 (0.1)
S549R	p.Ser549Arg	II/III	4 (0.1)
A559T	p.Ala559Thr	unknown	4 (0.1)
R117C	p.Arg117Cys	IV/V	4 (0.1)
G178R	p.Gly178Arg	unknown	4 (0.1)
P574H	p.Pro574His	IV	4 (0.1)
R1283M	p.Arg1283Met	unknown	3 (0.04)
Y563D	p.Tyr563Asp	unknown	3 (0.04)
2143delT	c.490-1G>A		3 (0.04)
3120G->A	c.2988G>A	V	2 (0.02)
2043delG	c.1911delG		2 (0.02)
1677delTA	p.Tyr515x		2 (0.02)
574delA	c.442delA	unknown	2 (0.02)
C524X	p.Cys524X	I	2 (0.02)
S1251N	p.Ser1251Asn	unknown	2 (0.02)
K710X	p.Lys710X	I	1 (0.01)
S549I			1 (0.01)
1609delCA	c.1477_1478delCA	unknown	1 (0.01)
2869insG	c.2737_2738insG	I	1 (0.01)
3662delA	c.3530delA	unknown	1 (0.01)
3849+4A->G	c.3717+4A>G	V	1 (0.01)
G330X			1 (0.01)
Q552X	p.Gln552X	I	1 (0.01)
2711delT			1 (0.01)
S364P	p.Ser364Pro		1 (0.01)
W1089X	p.Trp1089X	I	1 (0.01)
Other			1013 (12.0)
Not identified			908 (10.7)

Cystic Fibrosis mutations and their functional effects



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

1.6 Age distribution by gender

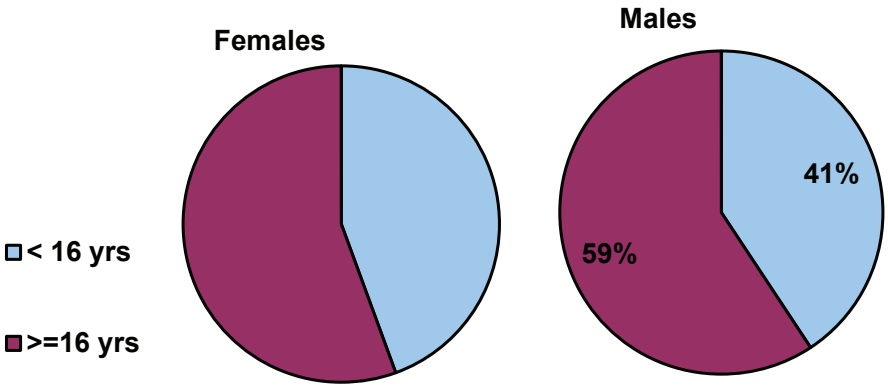


Age is calculated as the age at annual review encounter.

1.7 Age and sex distribution

Age	Overall N=8794	Female N=4143	Male N=4651
0–3 yrs	929 (10.6%)	458 (11.1%)	471 (10.1%)
4–7	962 (10.9%)	465 (11.2%)	497 (10.7%)
8–11	862 (9.8%)	448 (10.8%)	414 (8.9%)
12–15	979 (11.1%)	467 (11.3%)	512 (11.0%)
16–19	993 (11.3%)	502 (12.1%)	491 (10.6%)
20–23	1011 (11.5%)	466 (11.3%)	545 (11.7%)
24–27	816 (9.3%)	369 (8.9%)	447 (9.6%)
28–31	679 (7.7%)	301 (7.3%)	378 (8.1%)
32–35	459 (5.2%)	196 (4.7%)	263 (5.7%)
36–39	303 (3.5%)	125 (3.0%)	178 (3.8%)
40–44	332 (3.8%)	142 (3.4%)	190 (4.1%)
45–49	227 (2.6%)	97 (2.3%)	130 (2.8%)
50–59	178 (2.0%)	74 (1.8%)	104 (2.2%)
60+	64 (0.7%)	33 (0.8%)	31 (0.7%)
Median (min-max)	18 (0–83)	17 (0–81)	19 (0–83)

1.8 Age distribution by sex



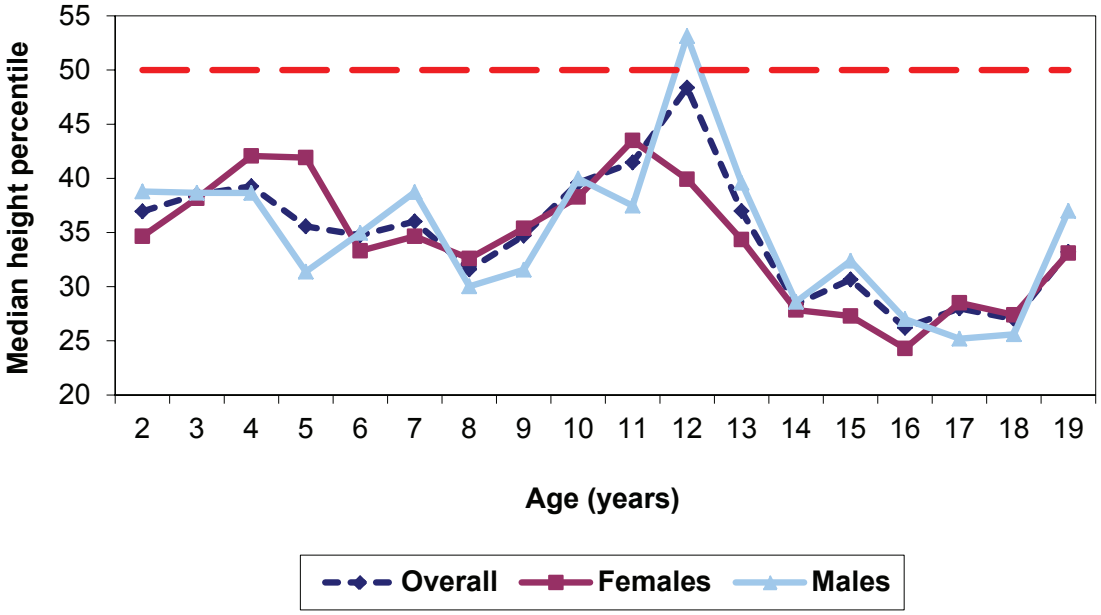
1.9 Employment and education status among adults aged 16 years and older

	Number of patients
Full-time working	1425
Part-time working	653
Student	917
Homemaker	231
Unemployed	684
“Disabled”	273
Retired	75
Unknown	862
No data	38

Note that these groups are not mutually exclusive.

Of the 4162 adults aged 16 years and older for whom an employment status questionnaire was completed (excluding “unknown”), 2928 (70.4%) reported being in work or study. In 2008, this figure was 69.9%.

1.10 Median height percentiles among children (n=4256)



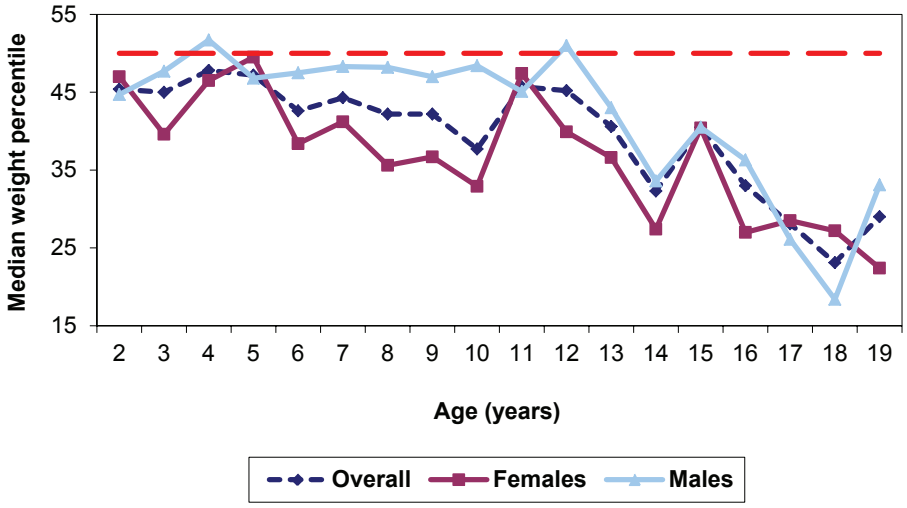
Age	N	Overall Median (IQR)	N	Female Median (IQR)	N	Male Median (IQR)
2	267	37.0 (17.5, 59.7)	122	34.6 (15.2, 58.7)	145	38.8 (21.4, 62.5)
3	241	38.5 (17.0, 61.7)	121	38.2 (14.4, 62.5)	120	38.7 (19.2, 61.0)
4	265	39.3 (15.8, 64.5)	136	42.1 (17.4, 64.2)	129	38.7 (13.1, 66.9)
5	254	35.6 (15.4, 64.0)	125	41.9 (16.5, 68.5)	129	31.4 (14.9, 61.4)
6	217	34.7 (14.1, 58.0)	99	33.3 (13.5, 55.4)	118	35.0 (14.7, 62.0)
7	216	36.0 (17.5, 63.2)	100	34.6 (17.4, 59.7)	116	38.7 (17.8, 66.0)
8	219	31.6 (12.9, 61.1)	124	32.6 (13.0, 61.1)	95	30.0 (12.5, 62.8)
9	212	34.7 (12.2, 63.3)	103	35.4 (11.9, 60.4)	109	31.6 (12.3, 65.1)
10	211	39.6 (16.1, 68.0)	117	38.3 (15.3, 68.3)	94	40.0 (19.5, 68.1)
11	210	41.5 (16.4, 69.5)	99	43.5 (16.6, 69.5)	111	37.5 (15.7, 69.5)
12	222	48.4 (16.7, 71.9)	113	39.9 (13.0, 68.0)	109	53.1 (19.7, 79.1)
13	254	37.0 (12.9, 66.3)	124	34.3 (12.0, 66.2)	130	39.6 (13.5, 67.6)
14	236	28.4 (8.9, 51.8)	104	27.8 (7.8, 57.4)	132	28.6 (14.0, 49.9)
15	257	30.7 (9.9, 60.2)	122	27.3 (6.9, 56.4)	135	32.4 (14.2, 63.3)
16	257	26.2 (8.5, 53.4)	130	24.3 (5.8, 52.3)	127	27.0 (11.9, 56.3)
17	255	28.0 (9.4, 53.2)	138	28.5 (10.7, 53.2)	117	25.2 (8.2, 54.8)
18	238	27.0 (9.3, 53.0)	116	27.4 (10.3, 58.7)	122	25.6 (7.2, 48.7)
19	225	33.2 (13.6, 61.0)	111	33.1 (13.3, 59.0)	114	37.0 (14.8, 64.9)

Age	N	Overall Median (IQR)	N	Female Median (IQR)	N	Male Median
Overall	4256	33.8 (13.2, 61.8)	2104	33.2 (12.3, 60.9)	2152	34.8 (14.4, 62.9)
2–4 yrs	773	38.4 (17.0, 62.1)	379	38.2 (16.0, 62.3)	394	38.7 (18.3, 61.5)
5–7 yrs	687	35.1 (15.5, 61.3)	324	35.7 (15.4, 60.4)	363	34.9 (15.8, 63.6)
8–10 yrs	642	35.7 (13.7, 64.5)	344	35.8 (13.2, 62.4)	298	35.5 (14.2, 66.0)
11–13 yrs	686	40.2 (15.5, 69.5)	336	39.1 (14.4, 67.5)	350	41.9 (16.2, 71.3)
14–15 yrs	493	29.6 (9.6, 57.0)	226	27.3 (7.6, 56.6)	267	31.0 (14.2, 57.1)
16–19 yrs	475	27.5 (9.8, 55.3)	495	27.4 (9.6, 54.2)	480	28.0 (10.6, 55.4)

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.

1.11 Median weight percentiles among children (n=4270)



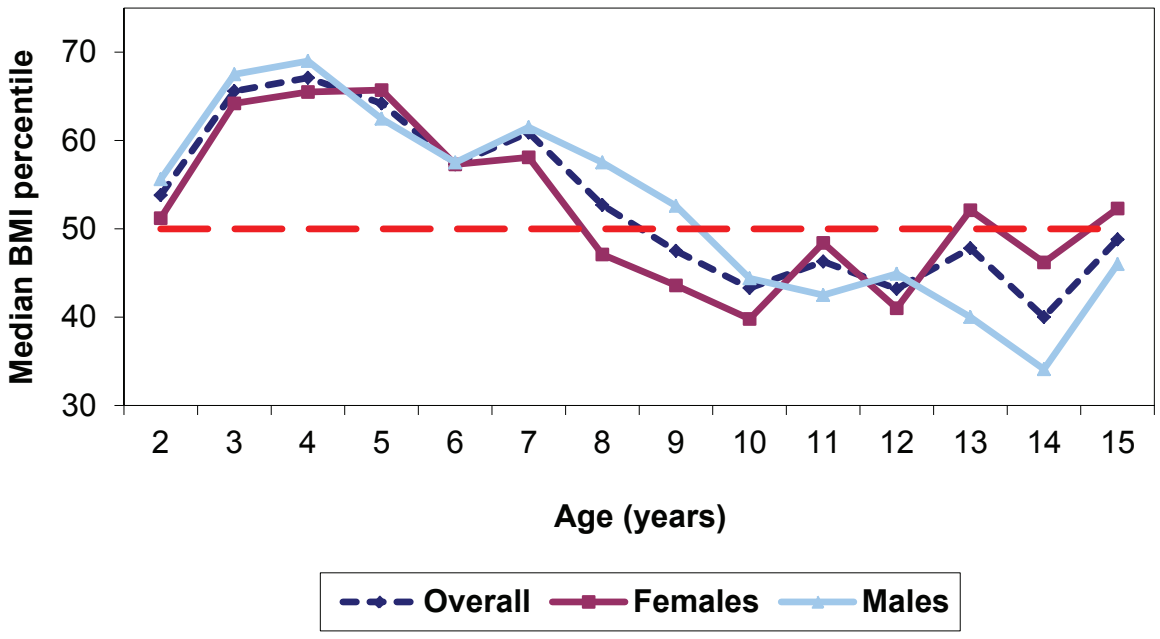
Age	N	Overall Median (IQR)	N	Female Median (IQR)	N	Male Median (IQR)
2	272	45.4 (21.4, 72.2)	124	47.0 (18.0, 70.1)	148	44.7 (24.0, 75.4)
3	243	45.0 (19.9, 72.7)	122	39.6 (18.9, 69.8)	121	47.7 (22.4, 75.6)
4	267	47.8 (21.2, 70.8)	137	46.5 (25.5, 67.6)	130	51.7 (19.6, 74.9)
5	254	47.2 (23.7, 73.6)	125	49.5 (27.3, 72.5)	129	46.8 (20.3, 74.7)
6	216	42.6 (18.2, 67.4)	99	38.4 (20.1, 59.1)	117	47.5 (18.1, 71.3)
7	217	44.3 (20.6, 68.9)	100	41.2 (19.3, 69.6)	117	48.3 (27.9, 68.6)
8	220	42.2 (20.4, 71.1)	124	35.6 (18.2, 71.1)	96	48.2 (21.2, 71.1)
9	213	42.2 (19.7, 65.2)	104	36.7 (17.9, 60.6)	109	47.0 (24.0, 67.4)
10	210	37.7 (20.9, 70.0)	116	32.9 (18.9, 64.3)	94	48.4 (26.5, 75.0)

Age	N	Overall Median (IQR)	N	Female Median (IQR)	N	Male Median (IQR)
11	211	45.7 (21.9, 74.1)	99	47.4 (19.0, 67.6)	112	45.1 (23.3, 76.9)
12	222	45.2 (20.5, 76.9)	113	39.9 (12.1, 69.4)	109	51.0 (25.1, 81.0)
13	254	40.6 (20.5, 69.5)	124	36.6 (17.5, 71.5)	130	43.0 (21.7, 68.5)
14	237	32.3 (14.1, 55.7)	105	27.4 (11.9, 56.9)	132	33.6 (14.4, 54.2)
15	257	40.5 (14.0, 69.0)	122	40.3 (11.0, 69.8)	135	40.5 (15.1, 68.6)
16	259	33.0 (12.0, 63.0)	131	27.0 (10.2, 58.1)	128	36.3 (16.2, 68.7)
17	255	28.1 (9.2, 58.8)	137	28.5 (8.8, 57.4)	118	26.1 (9.8, 59.8)
18	242	23.1 (4.8, 59.4)	117	27.2 (5.8, 61.2)	125	18.4 (3.8, 58.7)
19	221	29.0 (6.9, 60.1)	109	22.4 (5.4, 48.1)	112	33.1 (8.0, 67.4)
Overall	4270	39.9 (16.5, 68.6)	2108	36.5 (15.4,66.9)	2162	42.0 (17.7, 69.9)
2–4 yrs	782	46.6 (21.1, 72.1)	383	45.3 (19.6,69.5)	399	47.4 (22.2, 75.4)
5–7 yrs	687	45.7 (20.9, 69.6)	324	44.0 (21.0,69.2)	363	47.0 (20.8, 70.7)
8–10 yrs	643	41.3 (20.6, 69.2)	344	34.8 (18.3,65.2)	299	47.6 (23.6, 70.6)
11–13 yrs	687	43.4 (21.1, 72.7)	336	39.6 (17.3,69.3)	351	45.1 (22.8, 76.7)
14–15 yrs	494	35.2 (14.2, 63.7)	227	34.0 (11.5,65.9)	267	35.5 (14.9, 58.7)
16–19 yrs	977	28.3 (8.3, 60.9)	494	27.1 (8.0, 56.7)	483	30.6 (9.0, 63.0)

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.

1.12 Median BMI percentiles among children (n=3164)

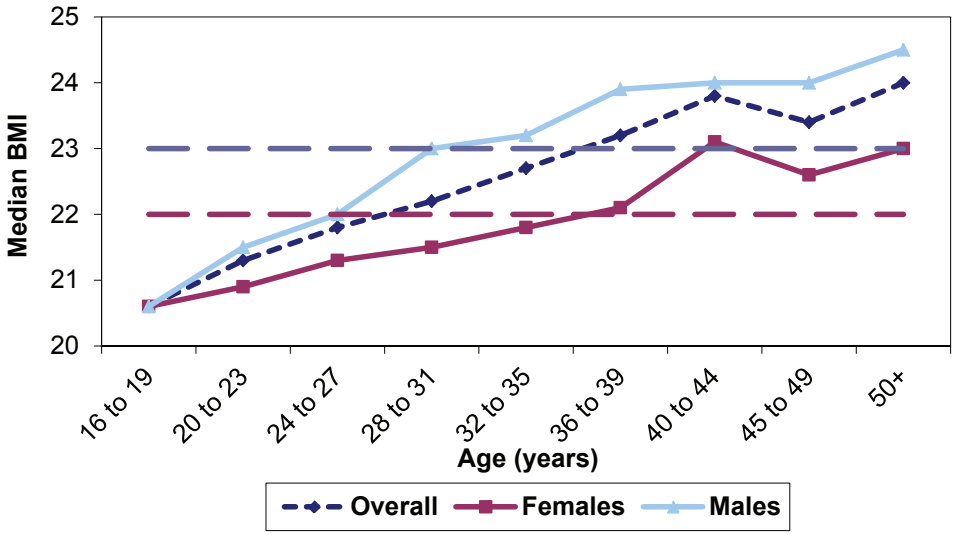


Age	N	Overall Median (IQR)	N	Female Median (IQR)	N	Male Median (IQR)
2	265	53.8 (32.3, 78.0)	122	51.2 (31.8,75.6)	143	55.6 (32.5, 80.3)
3	241	65.6 (38.9, 81.5)	121	64.2 (37.8,79.7)	120	67.5 (39.7, 83.5)
4	264	67.1 (36.1, 84.6)	135	65.5 (33.3,82.2)	129	69.0 (38.3, 87.1)
5	254	64.2 (40.2, 81.5)	125	65.7 (45.8,80.0)	129	62.5 (35.6, 83.4)
6	216	57.3 (31.0, 76.2)	99	57.3 (30.9,75.3)	117	57.5 (30.0, 77.0)
7	216	60.9 (34.8, 74.3)	100	58.1 (31.9,73.7)	116	61.5 (38.6, 75.2)
8	219	52.7 (32.2, 74.5)	124	47.1 (28.2,74.4)	95	57.5 (33.9, 74.7)
9	212	47.5 (27.4, 70.5)	103	43.6 (25.5,68.5)	109	52.6 (30.9, 74.6)
10	210	43.3 (26.0, 67.3)	116	39.8 (23.6,63.5)	94	44.4 (31.3, 69.7)
11	210	46.3 (23.1, 72.9)	99	48.4 (21.1,69.9)	111	42.5 (25.0, 74.3)
12	222	43.2 (17.6, 70.6)	113	41.0 (19.4,69.1)	109	44.9 (16.2, 73.8)
13	254	47.8 (23.1, 70.4)	124	52.1 (29.5,73.2)	130	40.0 (18.6, 67.6)
14	236	40.0 (18.5, 65.5)	104	46.2 (25.9,68.0)	132	34.1 (12.5, 60.8)
15	145	48.8 (22.8, 70.3)	75	52.3 (23.2,75.3)	70	46.0 (21.6, 66.9)
Overall	3164	52.7 (28.4, 75.5)	1560	52.9 (28.3,74.7)	1604	52.6 (28.4, 76.2)
2–4 yrs	770	62.7 (35.6, 81.9)	378	59.7 (33.3,80.0)	392	65.2 (37.3, 84.4)
5–7 yrs	686	61.3 (35.9, 77.6)	324	61.6 (36.9,77.1)	362	61.1 (34.2, 77.9)
8–10 yrs	641	46.5 (27.7, 70.6)	343	43.9 (25.9,69.5)	298	51.6 (32.4, 72.4)
11–13 yrs	686	45.9 (22.0, 71.1)	336	48.6 (23.6,70.9)	350	42.2 (20.6, 71.2)
14–15 yrs	381	41.5 (21.0, 67.5)	179	49.8 (25.8,69.0)	202	37.2 (14.3, 61.6)

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 as close to the 50th percentile as possible.

1.13 Median BMI values among adults (n=4907)

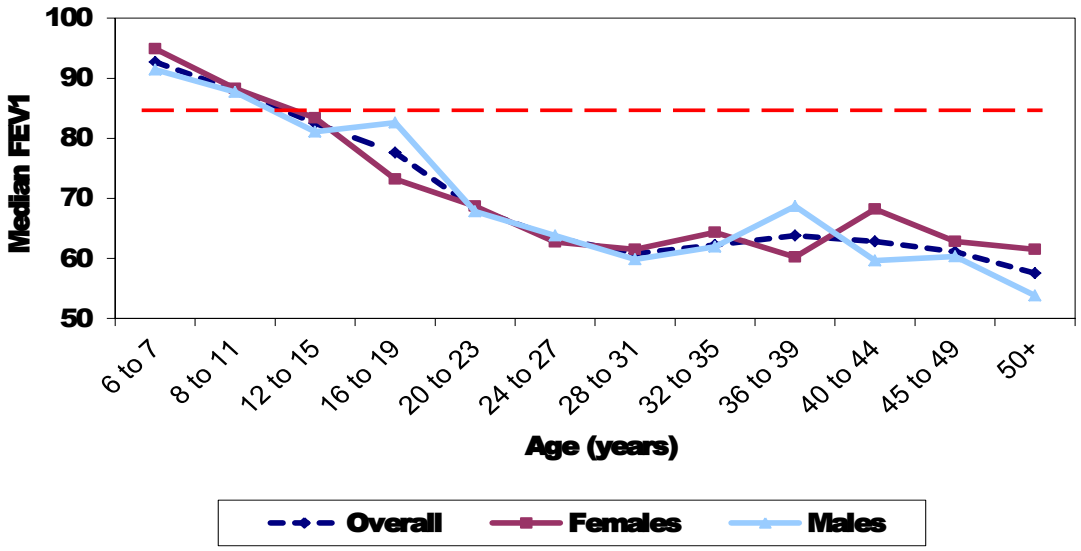


Age	N	Overall Median (IQR)	N	Female Median (IQR)	N	Male Median (IQR)
16–19	967	20.6 (18.8, 22.7)	490	20.6 (18.7,22.7)	477	20.6 (18.8, 22.8)
20–23	973	21.3 (19.3, 23.1)	452	20.9 (19.2,22.8)	521	21.5 (19.3, 23.5)
24–27	797	21.8 (19.9, 23.6)	362	21.3 (19.3,23.3)	435	22.0 (20.3, 24.0)
28–31	655	22.2 (20.4, 24.7)	289	21.5 (19.8,23.7)	366	23.0 (21.0, 25.1)
32–35	438	22.7 (20.6, 25.0)	184	21.8 (20.2,24.5)	254	23.2 (21.3, 25.1)
36–39	296	23.2 (21.2, 25.7)	123	22.1 (20.6,24.5)	173	23.9 (21.6, 26.3)
40–44	321	23.8 (21.5, 25.9)	139	23.1 (20.5,25.6)	182	24.0 (22.1, 25.9)
45–49	224	23.4 (21.2, 26.5)	97	22.6 (20.5,25.7)	127	24.0 (22.1, 26.8)
50+	236	24.0 (21.6, 27.1)	103	23.0 (20.8,26.7)	133	24.5 (22.5, 27.2)
Overall	4907	21.9 (19.9, 24.3)	2239	21.4 (19.5,23.5)	2668	22.4 (20.3, 24.7)

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.

1.14 Median FEV_1 (% predicted) among patients aged 6 years and older (n=6935)



Age	N	Overall Median (IQR)	N	Female Median (IQR)	N	Male Median (IQR)
6–7	389	92.7 (82.3, 102.0)	180	94.9 (84.7, 102.1)	209	91.4 (80.0, 101.7)
8–11	811	88.0 (76.9, 97.2)	424	88.3 (76.7, 97.2)	387	87.7 (77.6, 97.2)
12–15	931	82.5 (68.2, 94.5)	447	83.4 (66.1, 96.3)	484	81.1 (69.6, 92.2)
16–19	947	77.6 (58.6, 92.5)	478	73.2 (54.9, 88.5)	469	82.6 (64.2, 95.9)
20–23	944	68.3 (47.9, 85.7)	437	68.7 (49.0, 85.6)	507	67.8 (47.4, 85.9)
24–27	781	63.4 (44.2, 81.7)	358	62.7 (45.7, 84.2)	423	63.8 (43.6, 80.4)
28–31	640	60.8 (42.2, 79.5)	284	61.5 (45.9, 80.8)	356	59.8 (39.6, 78.1)
32–35	434	62.2 (44.3, 81.1)	187	64.3 (44.7, 80.7)	247	61.9 (44.1, 81.6)
36–39	292	63.8 (43.9, 80.8)	123	60.2 (43.7, 76.6)	169	68.7 (44.0, 81.9)
40–44	318	62.8 (41.8, 80.8)	137	68.2 (42.8, 83.0)	181	59.6 (40.4, 76.7)
45–49	218	61.1 (40.5, 80.9)	94	62.8 (41.5, 74.2)	124	60.3 (38.9, 82.9)
50+	230	57.5 (39.3, 77.9)	101	61.5 (39.1, 75.1)	129	53.8 (39.3, 79.6)
Overall	6935	74.2 (53.5, 90.6)	3250	74.3 (54.1, 91.3)	3685	74.1 (53.1, 90.1)

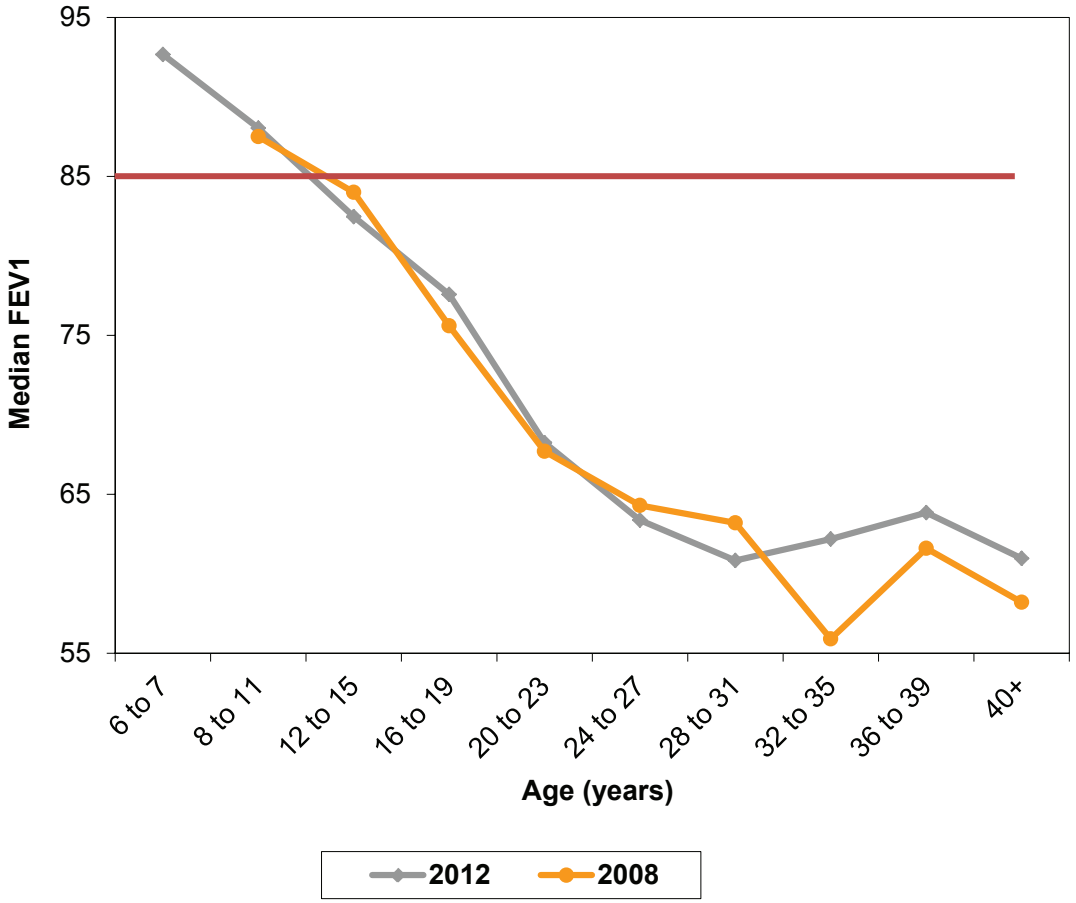
N refers to the number of patients in each age/sex category among those with non-missing FEV_1 % predicted data

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.

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 was 66%.

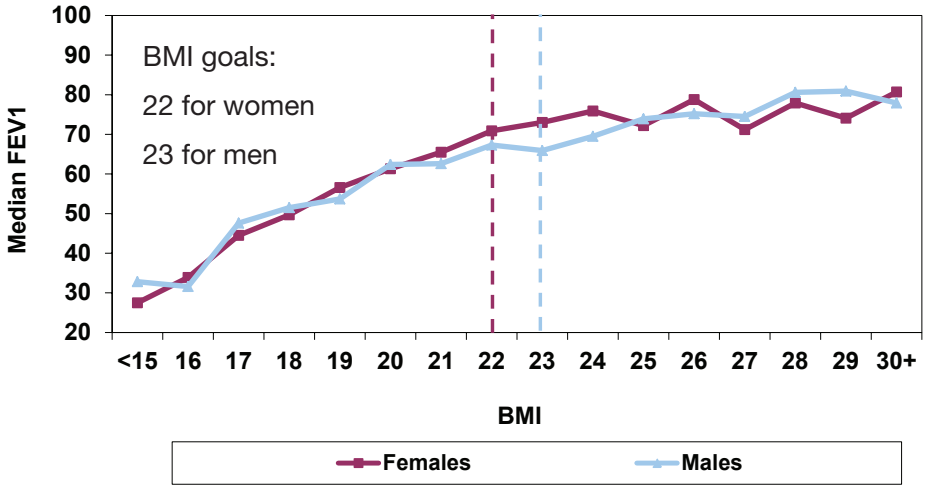
1.15 Median FEV_1 (% predicted) among patients aged 6 years and older by year in 2008 and 2012



An analysis was conducted in order to determine whether there were statistically significant differences in FEV_1 (% predicted) in 2012 compared to 2008 by age category. The results showed that none of the differences reached statistical significance as the p-values below show:

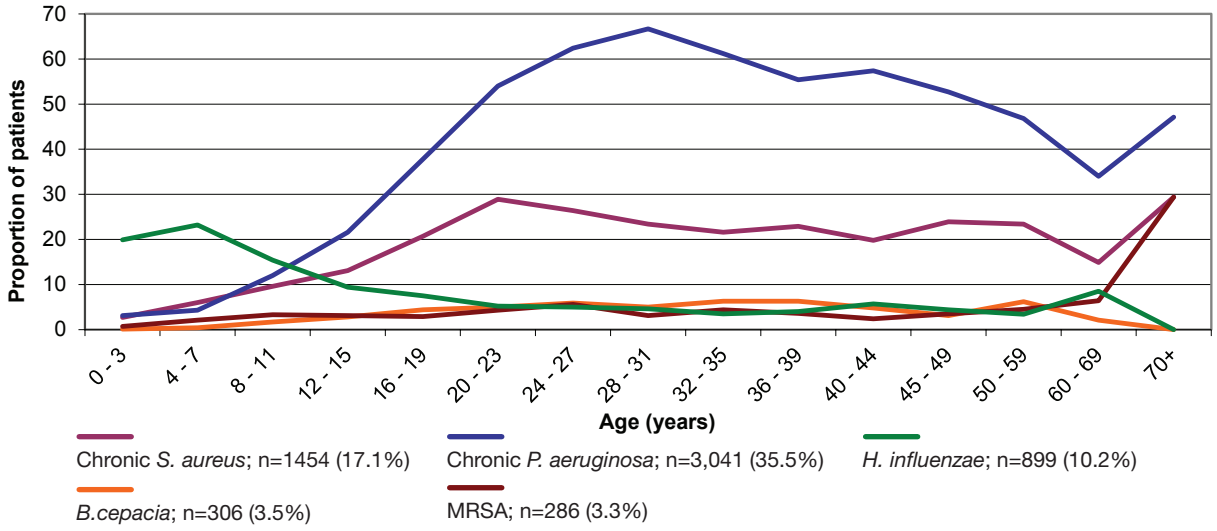
	6–7	8–11	12–15	16–19	20–23	24– 27	28– 31	32–35	36–39	40+
p-value	0.208	0.874	0.131	0.468	0.935	0.939	0.605	0.073	0.606	0.144

1.16 Median FEV_1 (% predicted) vs BMI among patients aged 16 years and older



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≤15 into one group and BMI≥30 into another.

1.17 Lung infections in 2012



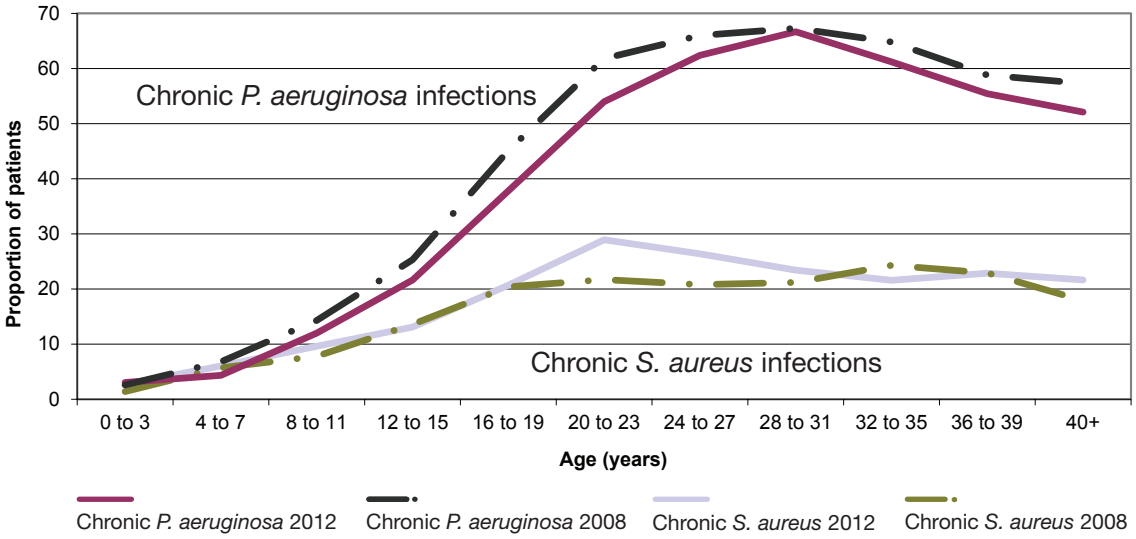
Chronic infection with *S. aureus* or *P. aeruginosa* were identified from annual review. Data on *B. cepacia*, MRSA and *H. influenzae* were collected from culture results at annual review.

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 *Pseudomonas 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 (yrs) Age (yrs)											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 yrs)	Adults (≥16 yrs)
N patients in age band	929	962	862	979	993	1011	816	679	459	303	332	227	242	8794	3732 (42.4)	5062 (57.6)
Chronic *S. aureus*; n(%)	25 (2.7)	57 (6.0)	80 (9.6)	125 (13.1)	197 (20.7)	283 (28.9)	206 (26.4)	153 (23.4)	95 (21.6)	67 (22.9)	62 (19.8)	52 (23.9)	52 (22.1)	1454 (17.1)	287 (7.9)	1167 (24.0)
Chronic *P. aeruginosa*; n(%)	28 (3.1)	41 (4.3)	101 (12.0)	207 (21.6)	361 (37.8)	528 (54.0)	491 (62.4)	440 (66.7)	276 (61.2)	163 (55.4)	183 (57.4)	118 (52.7)	104 (44.3)	3041 (35.5)	377 (10.3)	2664 (54.3)
Intermittent *P. aeruginosa*; n(%)	222 (24.1)	235 (24.9)	221 (26.3)	256 (26.8)	201 (21.1)	149 (15.2)	98 (12.5)	70 (10.6)	48 (10.6)	34 (11.6)	24 (7.5)	19 (8.5)	35 (14.9)	1612 (18.8)	934 (25.5)	678 (13.8)
Intermittent *S.Aureus*;n(%)	137 (14.9)	201 (21.3)	202 (24.3)	214 (22.4)	184 (19.4)	162 (16.6)	127 (16.3)	105 (16.1)	47 (10.7)	36 (12.3)	34 (10.8)	23 (10.6)	22 (9.4)	1494 (17.6)	754 (20.7)	740 (15.2)
B. cepacia; n(%)	1 (0.1)	4 (0.4)	15 (1.7)	27 (2.8)	44 (4.4)	50 (5.0)	48 (5.9)	34 (5.0)	29 (6.3)	19 (6.3)	16 (4.8)	7 (3.1)	12 (5.0)	306 (3.5)	47 (1.3)	259 (5.1)
MRSA; n(%)	6 (0.7)	20 (2.1)	28 (3.3)	30 (3.1)	29 (2.9)	43 (4.3)	46 (5.6)	21 (3.1)	20 (4.4)	11 (3.6)	8 (2.4)	8 (3.5)	16 (6.6)	286 (3.3)	84 (2.3)	202 (4.0)
H.influenza; n(%)	185 (19.9)	223 (23.2)	133 (15.4)	92 (9.4)	74 (7.5)	53 (5.2)	41 (5.0)	31 (4.6)	16 (3.5)	12 (4.0)	19 (5.7)	10 (4.4)	10 (4.1)	899 (10.2)	633 (17.0)	266 (5.3)

Age is calculated as age at annual review

1.18 Lung infections in 2008 and 2012



When comparing pseudomonas infections between 2008 with 2012 among young adults – 16 to 19 years and 20 to 23 years – we observed a statistically significant reduction in the proportion of patients chronically colonised (p-value= 0.004 and 0.014, respectively).

1.19 Prevalence of complications

	Overall (n=8794)	<16 years (n=3732)	≥16 years (n=5062)
Asthma; n(%)	1280 (14.6)	488 (13.1)	792 (15.6)
Sinus disease; n(%)	688 (7.8)	44 (1.2)	644 (12.7)
Nasal polyps requiring surgery; n(%)	205 (2.3)	47 (1.3)	158 (3.1)
Non-tuberculous mycobacteria or atypical mycobacteria; n(%)	465 (5.3)	94 (2.5)	371 (7.3)
Pneumothorax requiring chest tube; n(%)	39 (0.4)	1 (0.03)	38 (0.8)
ABPA; n(%)	909 (10.3)	278 (7.4)	631 (12.5)
Hemoptysis; n(%)	76 (0.9)	1 (0.03)	75 (1.5)
Cirrhosis with no portal hypertension; n(%)	124 (1.4)	25 (0.7)	99 (2.0)
Cirrhosis with portal hypertension; n(%)	164 (1.9)	24 (0.6)	140 (2.8)
Liver disease; n(%)	1181 (13.4)	366 (9.8)	815 (18.1)
Liver enzymes; n(%)	976 (11.1)	242 (6.5)	734 (14.5)

	Overall (n=8794)	<16 years (n=3732)	≥16 years (n=5062)
Gallbladder disease requiring surgery; n(%)	32 (0.4)	2 (0.1)	30 (0.6)
Intestinal obstruction; n(%)	569 (6.5)	144 (3.9)	425 (8.4)
Peptic ulcer; n(%)	6 (0.1)	1 (0.03)	5 (0.1)
GERD; n(%)	1345 (15.3)	318 (8.5)	1027 (20.3)
GI bleed req hosp non variceal n(%)	9 (0.1)	3 (0.1)	6 (0.1)
GI bleed req hosp variceal; n(%)	8 (0.1)	2 (0.1)	6 (0.1)
Kidney stones; n(%)	79 (0.9)	8 (0.2)	71 (1.4)
Renal failure; n(%)	18 (0.2)	1 (0.0.3)	17 (0.3)
Pancreatitis; n(%)	73 (0.8)	2 (0.1)	71 (1.4)
Fibrosing colonopathy/ colonic stricture; n(%)	5 (0.1)	2 (0.1)	3 (0.1)
Rectal prolapse; n(%)	27 (0.3)	24 (0.6)	3 (0.1)
Arthritis ; n(%)	147 (1.7)	13 (0.3)	134 (2.6)
Arthropathy; n(%)	463 (5.3)	18 (0.5)	445 (8.8)
Bone fracture; n(%)	66 (0.8)	11 (0.3)	55 (1.1)
Osteopenia; n(%)	1011 (11.5)	26 (0.7)	985 (19.5)
Osteoporosis; n(%)	493 (5.6)	7 (0.2)	486 (9.6)
Cancer confirmed by histology; n(%)	18 (0.2)	3 (0.1)	15 (0.3)
Port inserted or replaced; n(%)	560 (6.4)	225 (6.0)	335 (6.6)
Absence of vas deferens; n(%)	593 (6.7)	2 (0.1)	591 (11.7)
Depression; n(%)	380 (4.3)	8 (0.2)	372 (7.3)
Hearing loss; n(%)	169 (1.8)	33 (0.9)	136 (2.7)
Hypertension; n(%)	181 (2.1)	0	181 (3.6)

For patients who are reported to have had non-tuberculous mycobacteria/atypical mycobacteria, cirrhosis (with/without portal hypertension), cancer or ABPA in 2012 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 2012 was the year the complication first developed.

	Newly identified in 2011			Newly identified in 2012		
	Overall (n=8679)	<16 years (n=3746)	≥16 years (n=4933)	Overall (n=8794)	<16 years (n=3732)	≥16 years (n=5062)
Nontuberculous mycobacteria or atypical mycobacteria; n(%)	131 (1.5)	33 (0.9)	98 (2.0)	159 (1.8)	30 (0.8)	129 (2.5)
ABPA; n(%)	190 (2.2)	78 (2.1)	112 (2.3)	169 (1.9)	64 (1.7)	105 (2.1)
Cirrhosis with no portal hypertension; n(%)	44 (0.5)	10 (0.3)	34 (0.7)	33 (0.4)	9 (0.2)	24 (0.5)
Cirrhosis with portal hypertension; n(%)	34 (0.4)	8 (0.2)	26 (0.5)	18 (0.2)	6 (0.2)	12 (0.2)
Cancer confirmed by histology *; n(%)				10 (0.1)	1 (0.02)	9 (0.2)

*New cases of cancer confirmed by histology were not included in the 2011 annual report

1.20 CF-related diabetes

	Overall* (n=8772)	<16 years * (n=3715)	≥16 years * (n=5057)
Treatment for CF-related diabetes; n(%)	1641 (18.7)	153 (4.1)	1488 (29.4)
Screening for CF-related diabetes	190 (2.2)	78 (2.1)	112 (2.3)
Yes	5068 (58.3)	2500 (67.8)	2568 (51.4)
No	1825 (21.0)	989 (26.8)	836 (16.7)
Known CF-related diabetes	1605 (18.5)	106 (2.9)	1499 (30.0)
Unknown	189 (2.2)	92 (2.5)	97 (1.9)

*Treatment for CF-related diabetes was enquired about in an annual review questionnaire which was completed by 8772 of the 8794 patients with “complete” annual review encounter data. For this reason the number of patients in each age group differs to section 1.19.

1.21 Transplants

	2008	2009	2010	2011	2012
Number of patients that year with annual review data evaluated for transplants	126	143	169	204	225
Number accepted on the transplant list	55	79	82	121	120
Number receiving transplants	24	25*	29	51*	55**
Types of transplants received: Bilateral lung Heart and lung Liver Other	 16 1 6 1	 19 0 5 2	 26 1 1 1	 43 4 2 3	 45 1 7 4

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

1.22 Other therapy

	Overall (n=8794)	<16 years (n=3732)	≥16 years (n=5062)
NIV; n(%)	234 (2.7)	43 (1.2)	191 (3.9)
Long-term oxygen; n(%)	625 (7.3)	106 (2.9)	519 (10.5)
Among those who had long-term oxygen therapy:			
Continuously	149 (23.8)	14 (13.2)	135 (26.0)
Nocturnal+exertion	139 (22.2)	19 (17.9)	120 (23.1)
PRN	87 (13.9)	7 (6.6)	80 (15.4)
With exacerbation	250 (40.0)	66 (62.3)	184 (35.5)

1.23 Feeding

	All patients (n=8794)	<16 years (n=3732)	≥16 years (n=5062)
Any supplemental feeding; n(%)	2763 (31.4)	1000 (26.8)	1763 (34.8)
Nasogastric tube	115 (4.2)	10 (1.0)	105 (6.0)
Gastrostomy tube / Button	551 (19.9)	212 (21.2)	339 (19.2)
Jejunal	4 (0.1)	1 (0.1)	3 (0.2)
TPN	6 (0.2)	4 (0.4)	2 (0.1)

1.24 Days on IV antibiotics

Age	N (%)	Home Median (IQR)	N (%)	Hospital Median (IQR)	N (%)	Total Median (IQR)
0–3 yrs	54 (5.8)	12 (7–20)	265 (28.7)	14 (9–21)	271 (29.3)	14 (12–24)
4–7	147 (15.4)	14 (10–28)	306 (32.0)	14 (8–24)	337 (35.2)	16 (14–35)
8–11	195 (22.7)	24 (14–36)	316 (36.8)	14 (8–28)	369 (43.0)	28 (14–42)
12–15	295 (30.3)	24 (13–42)	457 (46.9)	16 (10–38)	524 (53.8)	35 (14–56)
16–19	312 (31.5)	21 (13–39)	464 (46.8)	15 (9–36)	539 (54.4)	28 (14–56)
20–23	413 (40.9)	22 (14–42)	505 (50.0)	20 (11–44)	626 (61.9)	31 (15–59)
24–27	344 (42.3)	25 (13–47)	395 (48.5)	17 (10–36)	501 (61.6)	30 (14–57)
28–31	282 (41.5)	26 (14–45)	297 (43.7)	20 (10–42)	406 (59.8)	33 (14–63)
32–35	182 (39.7)	28 (14–46)	169 (36.9)	15 (8–29)	242 (52.8)	29 (14–56)
36–39	112 (37.0)	24 (13–40)	122 (40.3)	14 (7–26)	156 (51.5)	28 (14–45)
40–44	102 (30.7)	26 (14–44)	110 (33.1)	14 (9–27)	150 (45.2)	28 (14–50)
45–49	70 (30.8)	25 (14–51)	81 (35.7)	18 (10–37)	103 (45.4)	35 (14–64)
50+	61 (25.2)	20 (11–35)	83 (34.3)	14 (9–31)	105 (43.4)	27 (14–43)
Overall	2569 (29.3)	23 (13–42)	3570 (40.7)	15 (9–33)	4329 (49.4)	28 (14–55)

N refers to the number of patients in each age category who had IV antibiotics

1.25 Nebulised drug treatment

Age	DNase treatment; n(%)					
	2008	2009	2010	2011	2012	p-value (2008 vs 2012)
0–3 yrs	46 (7.6)	55 (6.9)	87 (9.5)	97 (10.1)	97 (10.4)	0.067
4–7	125 (20.1)	152 (20.5)	172 (21.9)	280 (30.0)	306 (31.8)	<0.001
8–11	227 (34.2)	332 (39.6)	367 (42.8)	395 (47.3)	445 (51.6)	<0.001
12–15	359 (46.4)	449 (48.2)	528 (54.5)	587 (57.8)	612 (62.5)	<0.001
16–19	377 (49.5)	441 (50.3)	509 (54.0)	575 (56.6)	587 (59.1)	<0.001
20–23	319 (44.0)	410 (47.6)	491 (53.5)	576 (57.4)	626 (61.9)	<0.001
24–27	288 (47.6)	345 (50.0)	389 (52.9)	437 (54.4)	475 (58.2)	<0.001
28–31	182 (43.4)	226 (45.0)	304 (52.7)	358 (55.8)	395 (58.2)	<0.001
32–35	108 (41.5)	127 (39.1)	155 (44.8)	211 (48.7)	237 (51.6)	0.009
36–39	83 (35.0)	102 (37.0)	127 (44.1)	120 (41.0)	141 (46.5)	0.007
40–44	147	194 (36.3)	123 (43.5)	132 (40.2)	156 (47.0)	<0.001**
45–49	(35.7)*	*	73 (45.3)	100 (47.9)	105 (46.3)	28 (14–55)
50+			66 (42.0)	75 (36.4)	104 (43.0)	
Overall	2261 (37.2)	2833 (38.4)	3391 (42.7)	3943 (45.4)	4286 (48.7)	

* In 2008 and 2009 all patients aged 40 years and older were grouped together
** All patients aged 40 years and older were grouped together for this comparison

Antibiotic use among patients with chronic *Pseudomonas aeruginosa*

	2008			2012		
	Overall	<16 years	≥16 years	Overall	<16 years	≥16 years
Patients with chronic pseudomonas	2098	299	1799	3041	377	2664
Tobramycin solution; n(%)	412 (19.6)	48 (16.1)	364 (20.2)	1018 (33.5)	120 (31.8)	898 (33.7)
Other aminoglycoside; n(%)	43 (2.0)	5 (0.2)	38 (2.1)	104 (3.4)	11 (2.9)	93 (3.5)
Colistin; n(%)	914 (43.6)	174 (58.2)	740 (41.1)	1326 (43.6)	214 (56.8)	1112 (41.7)
Promixin; n(%)	490 (23.4)	73 (24.4)	417 (23.2)	810 (26.6)	133 (35.3)	677 (25.4)
At least one of the above; n(%)	1597 (76.1)	257 (86.0)	1340 (74.5)	2444 (80.4)	340 (90.2)	2104 (79.0)

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

1.26 Physiotherapy techniques

	All patients (n=8794)	<16 years (n=3732)	≥16 years (n=5062)
Active cycle of breathing techniques; n(%)	3525 (40.8)	1861 (51.6)	1664 (33.1)
Autogenic drainage (including assisted autogenic drainage); n(%)	971 (11.2)	137 (3.8)	834 (16.6)
Any form of PEP; n(%)	4330 (50.1)	2207 (61.2)	2123 (42.2)
VEST; n(%)	157 (1.8)	94 (2.6)	63 (1.3)

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 4084 patients from paediatric care centres with complete* data at 2012 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 (or 25th percentile); the upper border denotes the third quartile (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 (Q3+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 (Q1-1.5xIQR). "Outside values" (or outliers) refer to values which 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.

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

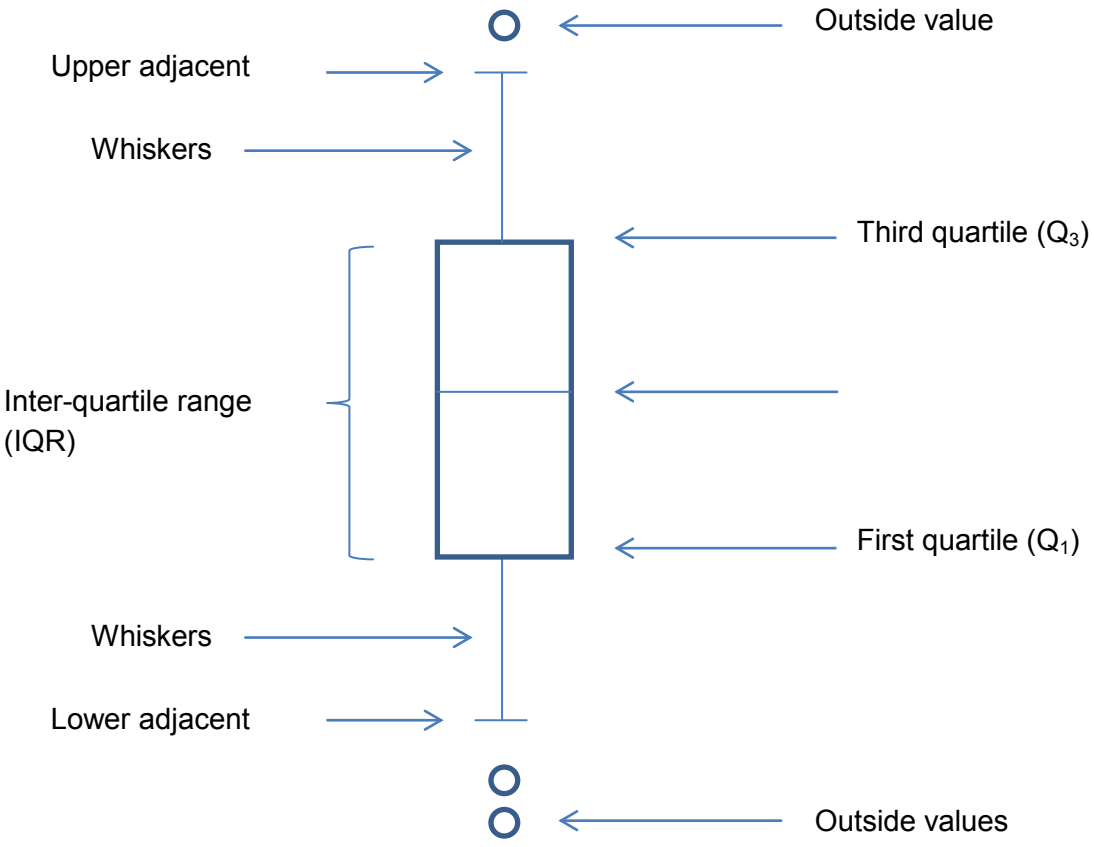
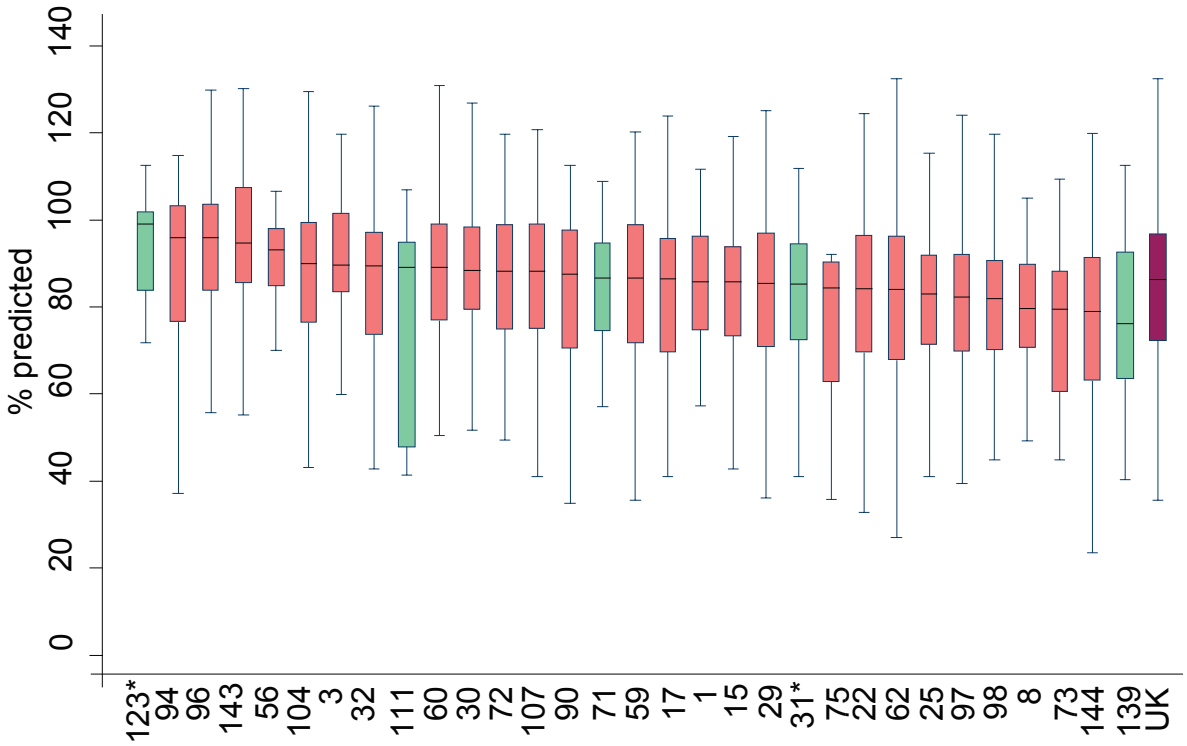


Figure 2.1 Median FEV_1 % predicted by paediatric centre/clinic

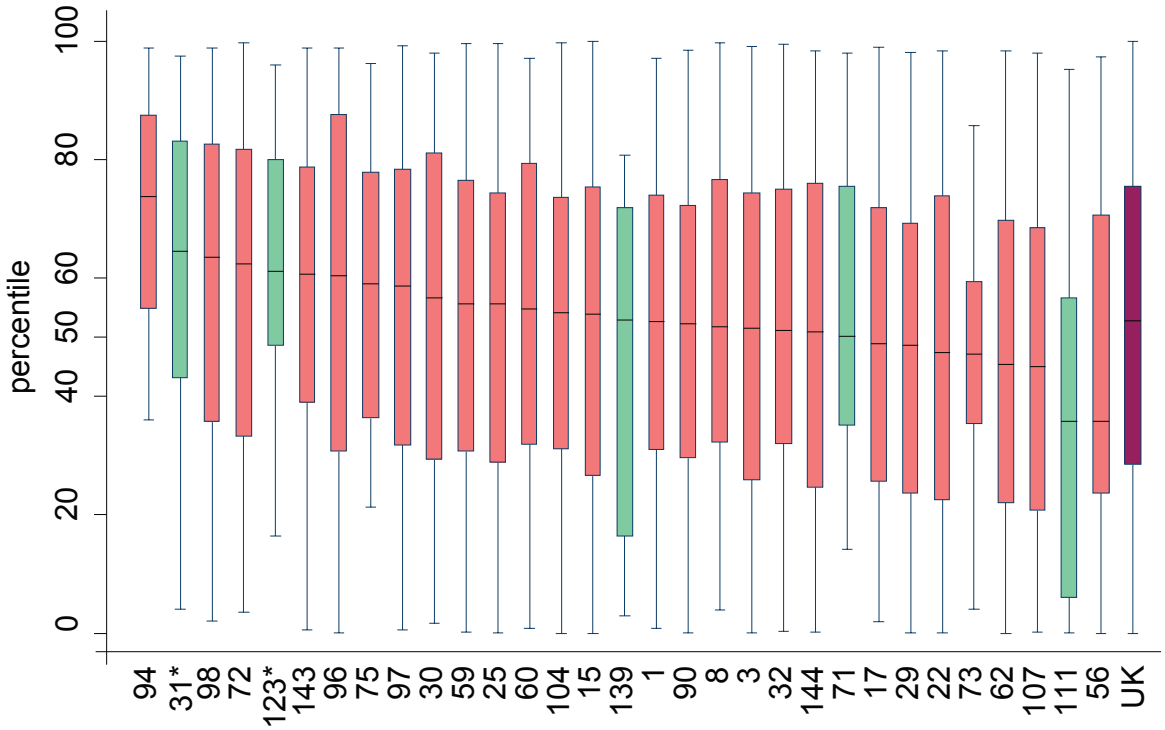


Excludes oustide values

The median FEV_1 % predicted for patients attending paediatric centres/clinics is 86% predicted (IQR: 72–97).

Red: centres with their network clinics. Green: stand-alone clinics. Purple: all. *Centre/clinic with a dataset submission of fewer than 20 patients.

Figure 2.2 Median BMI percentile by paediatric centre/clinic

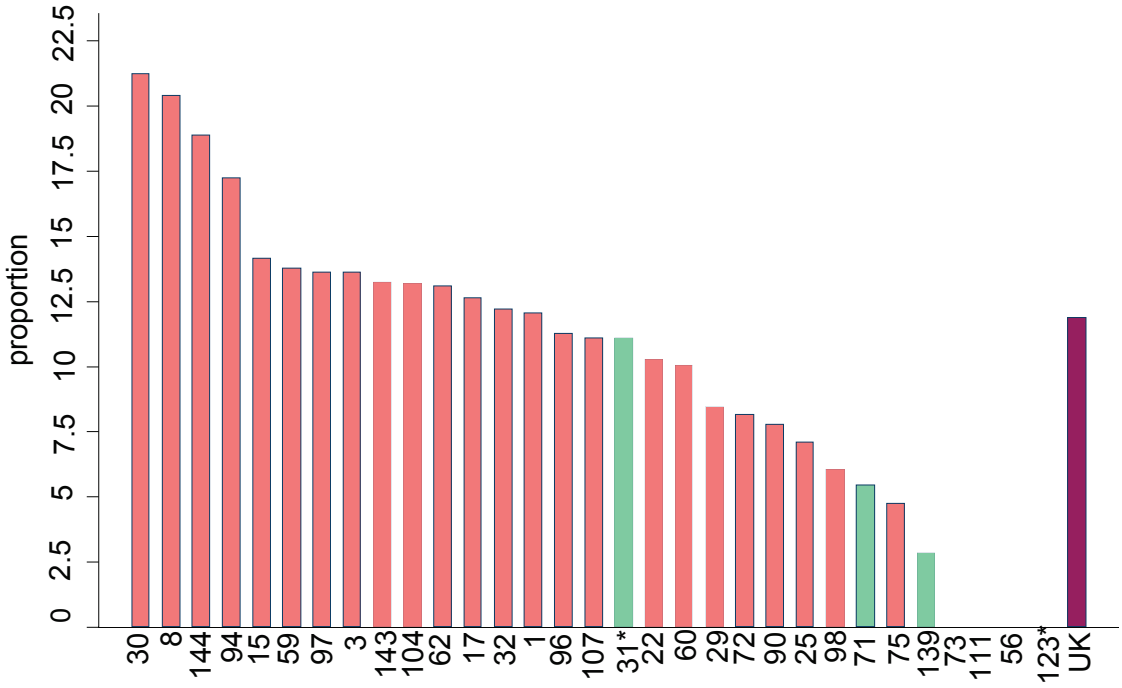


Excludes oustide values

The median BMI percentile for paediatric centres/clinics is 53 (IQR: 28–75).

Red: centres with their network clinics. Green: stand-alone clinics. Purple: all. *Centre/clinic with a dataset submission of less 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* for paediatric entres/clinics is 12%.

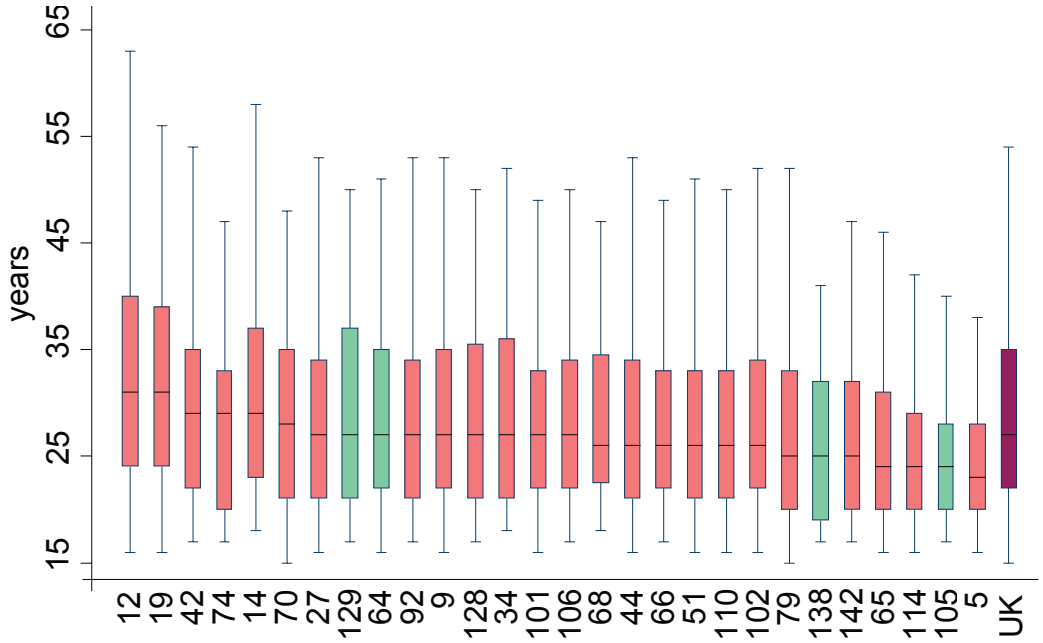
Red: centres with their network clinics. Green: stand-alone clinics. Purple: all. *Centre/clinic with a dataset submission of fewer than 20 patients.

Section 3: Analyses by adult

(based on 4710 patients from adult services with complete* data at 2012 annual review)

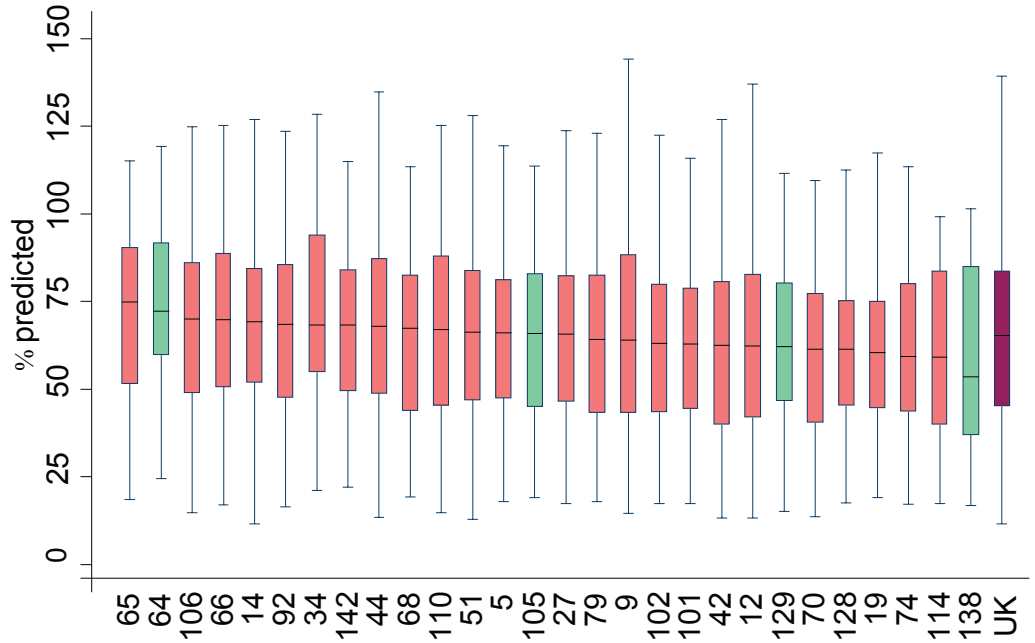
* “Complete” data refers to the minimum data required to produce the range of clinical outcomes presented in this report.

Figure 3.1 Median age (years) by adult services



The median age in adult services is 27 years (IQR: 22–35).
Red: centres. Green: other clinics. Purple: all.

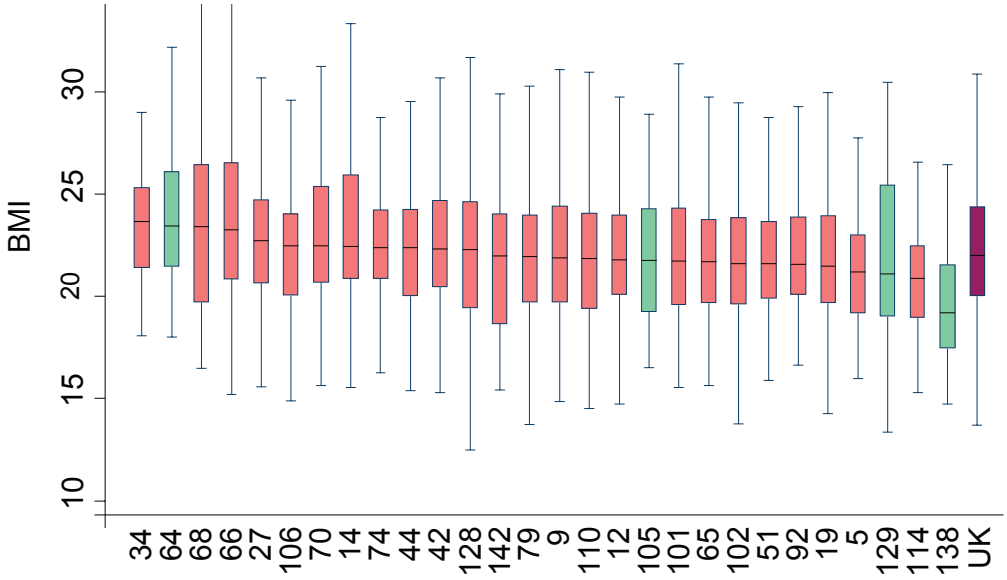
Figure 3.2 Median FEV_1 (% predicted) by adult services



Excludes oustide values

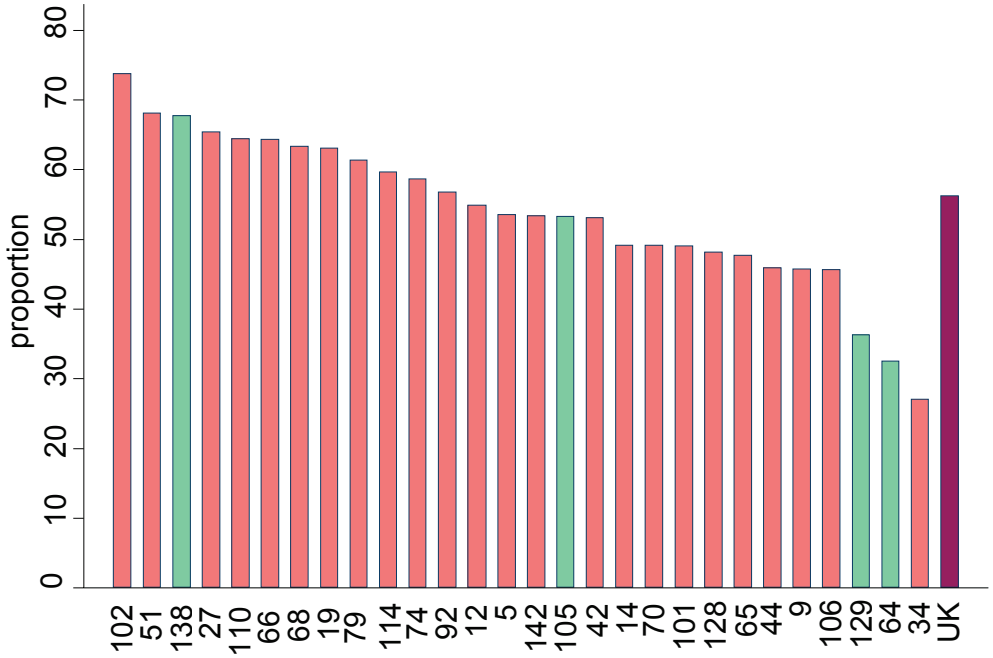
The median FEV_1 (% predicted) in adult services is 65% (IQR: 45–84).
Red: centres. Green: other clinics. Purple: all.

Figure 3.3 Median BMI by adult services



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 services



Excludes oustide values

The proportion of patients with chronic *P. aeruginosa* for adult services is 56%. Red: centres. Green: other clinics. Purple: all.

Section 4: Care centres/clinics providing data in 2012

4.1 Paediatric centres/clinics providing data in 2012 – ordered by clinic ID

The number of active patients at any given centre will fluctuate throughout the year based on new diagnoses, deaths, and transfers. The figures quoted for each centre (in this and all subsequent sections) are therefore estimates based on where patients had their annual review encounter in 2012 and, for those patients alive in 2012 who did not have an annual review encounter, where they had their last encounter in 2011–2012. Patients in the latter group who have transferred from where they had their last clinical encounter are not included.

Country	Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2012
England	Leicester	Leicester Royal Infirmary	1	58	58
England	Sheffield	Sheffield Children's Hospital	3	138	136
England	Stoke	University Hospital of North Staffordshire	8	99	98
England	London – South West	Royal Brompton Hospital	15	313	312
England	London	King's College Hospital	17	191	191
England	Oxford	John Radcliffe Hospital	22	171	165
England	Leeds	St James's University Hospital	25	245	239
England	Southampton	Southampton General Hospital	29	200	194
England	London – East	Royal London Hospital	30	119	114
Scotland	Inverness	Raigmore Hospital	31	18	18
England	Bristol	Bristol Royal Hospital for Children	32	177	174
Scotland	Glasgow	Royal Hospital for Sick Children	56	115	83

Country	Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2012
England	Newcastle	Royal Victoria Infirmary	59	170	157
Northern Ireland	Belfast	Royal Belfast Hospital for Sick Children	60	196	183
England	Nottingham	Nottingham City Hospital	62	174	170
England	Teesside	James Cook University Hospital	71	57	55
Wales	Cardiff	Children's Hospital for Wales	72	183	165
Scotland	Dundee	Ninewells Hospital	73	22	22
Scotland	Aberdeen	Royal Aberdeen Children's Hospital	75	25	21
England	London – Central	Great Ormond Street Hospital for Children	90	176	175
England	Truro	Royal Cornwall Hospital	94	31	29
England	Exeter	Royal Devon & Exeter Hospital	96	76	74
England	Liverpool	Alder Hey Children's Hospital	97	293	287
England	Norwich	Norfolk & Norwich University Hospital	98	68	67
England	Birmingham	Birmingham Children's Hospital	104	292	291
England	Cambridge	Addenbrookes Hospital	107	127	127
England	Hull	Hull Royal Infirmary	111	35	35
Scotland	Ayr/Kilmarnock	Crosshouse Hospital	123	20	18
England	Plymouth	Derriford Hospital	139	38	35
Scotland	Edinburgh	Royal Hospital for Sick Children	143	106	83
England	Manchester	Royal Manchester Children's Hospital	144	313	308

4.2 Adult centres/clinics providing data in 2012 – ordered by clinic ID

Country	Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2012
England	London – South East	King's College Hospital	5	164	156
England	Newcastle	Royal Victoria Infirmary	9	240	238
England	London – South West	Royal Brompton Hospital	12	638	622
Northern Ireland	Belfast	Belfast City Hospital	14	215	187
England	Frimley	Frimley Park Hospital	19	113	111
England	Birmingham	Birmingham Heartlands Hospital	27	329	322
England	Exeter	Royal Devon & Exeter Hospital	34	79	76
England	Leeds	St James's University Hospital	42	404	395
Scotland	Edinburgh	Western General Hospital	44	217	210
England	Cambridge	Papworth Hospital	51	264	255
England	Plymouth	Derriford Hospital	64	43	43
England	Sheffield	Northern General Hospital	65	158	155
England	Liverpool	Liverpool Heart and Chest Hospital	66	261	247
Wales	Llandough	Llandough Hospital	68	169	32
Scotland	Aberdeen	Aberdeen Royal Infirmary	70	62	59

Country	Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2012
England	Stoke-on-Trent	University Hospital of North Staffordshire	74	63	63
Scotland	Glasgow	Gartnavel General Hospital	79	220	215
England	London – East	London Chest Hospital	92	144	129
England	Nottingham	Nottingham City Hospital	101	129	122
England	Manchester	Wythenshawe Hospital	102	379	369
England	London – South East	University Hospital Lewisham	105	47	45
England	Bristol	Bristol Royal Infirmary	106	166	163
England	Southampton	Southampton General Hospital	110	226	215
England	Norwich	Norfolk & Norwich University Hospital	114	61	60
England	Oxford	Churchill Hospital	128	85	84
England	Truro	Royal Cornwall Hospital	129	33	33
England	Hull	Castle Hill Hospital	138	36	31
England	Leicester	Glenfield Hospital	142	74	73

4.3 Paediatric centres/clinics providing data in 2012 – alphabetical order

England

Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2012
Birmingham	Birmingham Children's Hospital	104	292	291
Bristol	Bristol Royal Hospital for Children	32	177	174
Cambridge	Addenbrookes Hospital	107	127	127
Exeter	Royal Devon & Exeter Hospital	96	76	74
Hull	Hull Royal Infirmary	111	35	35
Leeds	St James's University Hospital	25	245	239
Leicester	Leicester Royal Infirmary	1	58	58
Liverpool	Alder Hey Children's Hospital	97	293	287
London – Central	Great Ormond Street Hospital for Children	90	176	175
London – East	Royal London Hospital	30	119	114
London – South East	King's College Hospital	17	191	191
London – South West	Royal Brompton Hospital	15	313	312
Manchester	Royal Manchester Children's Hospital	144	313	308
Newcastle	Royal Victoria Infirmary	59	170	157
Norwich	Norfolk & Norwich University Hospital	98	68	67
Nottingham	Nottingham City Hospital	62	174	170
Oxford	John Radcliffe Hospital	22	171	165
Plymouth	Derriford Hospital	139	38	35
Sheffield	Sheffield Children's Hospital	3	138	136
Southampton	Southampton General Hospital	29	200	194
Stoke	University Hospital of North Staffordshire	8	99	98
Teesside	James Cook University Hospital	71	57	55
Truro	Royal Cornwall Hospital	94	31	29

Northern Ireland

Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2012
Belfast	Royal Belfast Hospital for Sick Children	60	196	183

Scotland

Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2012
Aberdeen	Royal Aberdeen Children's Hospital	75	25	21
Ayr/Kilmarnock	Crosshouse Hospital	123	20	18
Dundee	Ninewells Hospital	73	22	22
Edinburgh	Royal Hospital for Sick Children	143	106	83
Glasgow	Royal Hospital for Sick Children	56	115	83
Inverness	Raigmore Hospital	31	18	18

Wales

Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2012
Cardiff	Children's Hospital for Wales	72	183	165

4.4 Adult centres/clinics providing data in 2012 – alphabetical order

England

Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2012
Birmingham	Birmingham Heartlands Hospital	27	329	322
Bristol	Bristol Royal Infirmary	106	166	163
Cambridge	Papworth Hospital	51	264	255
Exeter	Royal Devon & Exeter Hospital	34	79	76
Frimley	Frimley Park Hospital	19	113	111
Hull	Castle Hill Hospital	138	36	31
Leeds	St James's University Hospital	42	404	395
Leicester	Glenfield Hospital	142	74	73
Liverpool	Liverpool Heart and Chest Hospital	66	261	247
London – East	London Chest Hospital	92	144	129
London – South East	King’s College Hospital	5	164	156
London – South East	University Hospital Lewisham	105	47	45
London – South West	Royal Brompton Hospital	12	638	622
Manchester	Wythenshawe Hospital	102	379	369
Newcastle	Royal Victoria Infirmary	9	240	238
Norwich	Norfolk & Norwich University Hospital	114	61	60
Nottingham	Nottingham City Hospital	101	129	122
Oxford	Churchill Hospital	128	85	84
Plymouth	Derriford Hospital	64	43	43
Sheffield	Northern General Hospital	65	158	155
Southampton	Southampton General Hospital	110	226	215
Stoke-on-Trent	University Hospital of North Staffordshire	74	63	63
Truro	Royal Cornwall Hospital	129	33	33

Northern Ireland

Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2012
Belfast	Belfast City Hospital	14	215	187

Scotland

Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2012
Aberdeen	Aberdeen Royal Infirmary	70	62	59
Edinburgh	Western General Hospital, Edinburgh	44	217	210
Glasgow	Gartnavel General Hospital, Glasgow	79	220	215

Wales

Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2012
Llandough	Llandough Hospital	68	169	32

Section 5: Registry Steering Committee membership

Registry Steering Committee membership:

Dr Diana Bilton (Chair)	Adult CF Centre Director, Royal Brompton Hospital, London
Dr Caroline Elston	Adult CF Centre Director, Kings Hospital, London
Dr Iolo Doull	Paediatric CF Centre Director, Cardiff Hospital, Wales
Dr Keith Brownlee	Paediatric CF Centre Director, St James Hospital, Leeds
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 & Trustee of CF Trust
Dr Stephanie MacNeill	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, London
Dr Kim Cox	Lead Specialist CF Commissioner, London
Dr Lisa Davies	Specialist Commissioner, Wales
Mr Ed Owen	Chief Executive, Cystic Fibrosis Trust
Ms Joanne Osmond	Director of Clinical Care, Cystic Fibrosis Trust
Mr Stephen Williams	Director of Finance, Cystic Fibrosis Trust
Dr Janet Allen	Director of Research, Cystic Fibrosis Trust
Ms Elaine Gunn	Registry Manager, Cystic Fibrosis Trust

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