Cystic Fibrosis Trust
Annual data report 2011

January 2013

Executive Summary

The 2011 annual data report for the UK CF Registry represents the culmination of dedication and hard work by CF specialist teams in collecting and submitting data from the people they care for with CF. This effort is a strong representation of the commitment to seeking the best outcomes possible for the CF community. The work would not be possible without the consent of patients and parents who also wish to see progressive improvement in outcomes.

The CF Trust is to be applauded for their vision and foresight in funding and facilitating the UK CF Registry at a time when others questioned the benefit of the investment. Seven years on the registry is a model for other specialist conditions and the power of having complete data on the population requiring care has been demonstrated. Furthermore the registry is now supported financially by the NHS in England as well as Scotland.

The registry data is used in a number of ways to help drive improvements in CF care. In the last year UK CF Registry data has helped the NHS to identify with some detail the requirements for services for people with CF in paediatric and adult care. In addition the registry has been a key resource for the teams involved in health technology appraisals of all the new drugs licenced and now available for CF. The comprehensive data set which illustrates lung function decline over time, specific to genotype, facilitated evidence based discussions on cost effectiveness which assisted in the approval for NHS funding.

The registry data supports critical analysis of CF specialist service outcomes at several levels. Each specialist team can analyse their own patient data at any time. At peer review the registry data plays a key role in assessing provision and standards of care, and in the annual report we chart centre outcomes.

It is clear to us that outcomes in terms of lung function can be affected by many factors including those related to care (such as use of CF therapies) and those related to the person with CF (such as genotype, age and socioeconomic status). We would like to be able to provide better comparisons of centres so that we can learn from each other and ensure optimum care for the community. As a step forward in this strategy we are delighted that a research team led by Dr Stephanie MacNeill, an Imperial College Statistician who has worked on the annual report for the last few years has been successful in obtaining a grant from the NIHR (National Institute for Health research) to develop better ways of comparing centres from registry data. We will expect to see changes in the annual report over the next three years as a result of that project.

For this year it is impressive to see that we have 9749 patients registered and that 89% of these have what we define as a complete dataset compared to 71% in 2008. This represents a year on year improvement in data quality and we continue to aim for 100%.

The registry works closely with the CF UK New-born Screening Programme Centre and we can see that 155 patients were added to the registry in 2011 as a result of a positive diagnosis from new-born screening. The registry provides data to the screening programme centre to ensure the best audit of outcomes of screening for CF is performed.

Improved survival of people with CF is a key goal for all of us and we know that this is best judged over long periods to really be sure of the significance of changes. The introduction of new CF correcting therapies like Kalydeco, which are known to improve lung function significantly are likely to have significant impact on survival but this may take many years to show in registry data. The median predicted survival for 2011 is 41.5 years which is in keeping with the 2010 data.

We have shown data for important outcomes such as FEV1, BMI and infections for the national data set over several years. We hope to see improvements in lung function over time as our goal is to ensure children graduating to adult services have lung function in the normal range and then maintain that level as they grow older.

Finally we note the increase in the number of lung transplants in 2011 in CF patients.
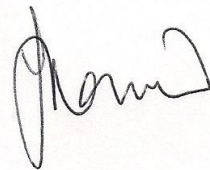
This report seeks to summarise important headline data in the UK population. There is a wealth of data behind this report which can be mined in order to gain insights into all aspects of CF therapies, complications and therapies. Data application forms can be found on the CF Trust web site or by contacting Elaine Gunn the registry manager at egunn@cftrust.org.

We want to thank all who continue to be involved in the UK CF Registry. The patients and families who consent to data collection, the clinical teams who continue to collect and submit such robust and complete data, as well as the registry team at the CF trust supported by the registry steering committee.

The registry has become a recognised tool in NHS care for people with CF and we look forward to continued success in using the registry to improve standards and outcomes in UK CF care.

Diana Bilton   Joanne Osmond
Chair UK CF Registry Steering Committee   Director of Clinical Care

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Section 1: All UK Patients

1.1 Summary of the UK Cystic Fibrosis Registry

	2007	2008	2009	2010	2011
CF patients registered	8080[1]	8513[1]	9029[1]	9385[1]	9749[1]
CF patients with "complete" data; n(%)	4408[2] (55%)	6082[2] (71%)	7377[2] (82%)	7937[2] (85%)	8679[2] (89%)
Age in years; median	18[3]	18[3]	17[3]	17[3]	18[3]
All newly diagnosed patients (newborn screening and other)	239[4]	235[4]	261[4]	301[4]	261[4]
Newly diagnosed patients identified through newborn screening				189	155
Age at diagnosis in months; median	5[3]	4[3]	3[3]	3[3]	3[3]
Adults aged 16 yrs and over; %	56.7[3]	56.2[3]	55.1[3]	55.5[3]	56.8[3]
Males; %	53.9[3]	53.3[3]	53.1[3]	53.1[3]	53.2[3]
Genotyped; %	92.6[3]	93.7[3]	94.3[3]	95.2[3]	95.6[3]
Median predicted survival in years (95% Confidence interval)	35.2[5] (31.0, 42.6)	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)
Total deaths reported	106	100	141	103	118
Age at death in years; median	24	27	27	29	26

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 2008/2009/2010/2011/2012; and
- had no recorded date of death before 1 January 2007/2008/2009/2010/2011

[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.

1.2 Age distribution of deaths in 2011
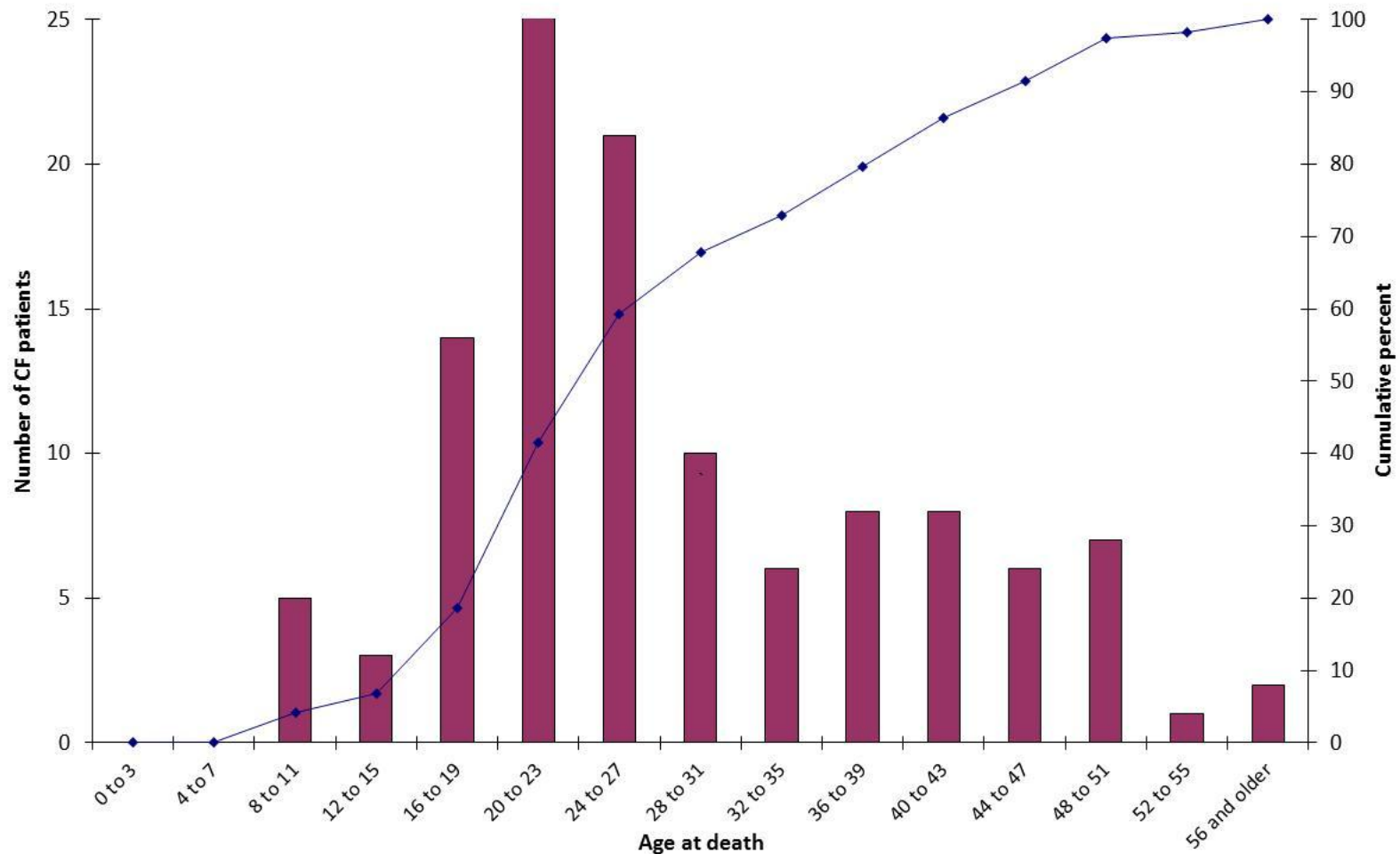
There were 118 recorded deaths in 2011. The median age at death was 26 years (min = 9 yrs; max = 73 years).

Analyses based on 8679 patients with complete* data at 2011 annual review

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

1.3 Age at diagnosis and screening statistics among children

Age at diagnosis	All patients; n (%)	Patients aged 10 years in 2011; n(%)	Patients aged 5 years in 2011; n(%)
Pre-natal	0	0	0
Birth – 3 months	2497 (67.7)	103 (50.2)	148 (65.8)
4-6 months	251 (6.8)	24 (11.7)	17 (7.6)
7-12 months	184 (5.0)	16 (7.8)	17 (7.6)
1 yr	257 (7.0)	22 (10.7)	18 (8.0)
2 yrs	174 (4.7)	15 (7.3)	12 (5.3)
3 yrs	108 (2.9)	5 (2.4)	3 (1.3)
4 yrs	68 (1.8)	7 (3.4)	6 (2.7)
5 yrs	41 (1.1)	5 (2.4)	4 (1.8)
6 yrs	22 (0.6)	2 (1.0)	-
7 yrs	18 (0.5)	0	-
8 yrs	27 (0.7)	1 (0.5)	-
9 yrs	16 (0.4)	3 (1.5)	-
10 yrs	8 (0.2)	2 (1.0)	-
11 yrs	9 (0.2)	-	-
12 yrs	3 (0.1)	-	-
13 yrs	1 (0.03)	-	-
14 yrs	3 (0.1)	-	-
15 yrs	0	-	-

The median (range) age at diagnosis is 1 month (0-14 years).

Diagnosis in the first three months of life was more common in children aged 5 years in 2011 (born in 2006) than in children aged 10 years in 2011 (born in 2001).

Of the 106 children with complete data born in 2011, 78 (74%) were identified by newborn screening.

In 2011, a total of 155 patients were identified by newborn screening (including patients with and without complete data). In 2010 this figure was 189 and in 2009 it was 151.

1.4 Age at diagnosis and screening statistics among adults

Age at diagnosis	n(%)
Pre-natal	0
Birth-3 months	1941 (40.0)
4-6 months	469 (9.7)
7-12 months	308 (6.4)
1 yr	411 (8.5)
2 yrs	253 (5.2)
3 yrs	191 (3.9)
4 yrs	166 (3.4)
5 yrs	80 (1.7)
6 yrs	70 (1.4)
7 yrs	51 (1.1)
8 yrs	56 (1.2)
9 yrs	44 (0.9)
10 yrs	38 (0.8)
11 yrs	34 (0.7)
12 yrs	36 (0.7)
13 yrs	39 (0.8)
14 yrs	33 (0.7)
15 yrs	35 (0.7)
16 – 20 yrs	147 (3.0)
21 – 25 yrs	97 (2.0)
26 – 30 yrs	74 (1.5)
31 – 35 yrs	83 ((1.7)
36 – 40 yrs	63 (1.3)
41 - 45 yrs	48 (1.0)
46-50 yrs	27 (0.6)
51 yrs +	60 (1.2)

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

Of the 4933 adults with complete data in 2011, 376 were diagnosed by neonatal screening and 26 adults were diagnosed in 2011.

1.5 Genotyping

8294 (95.6%) patients have been genotyped with a recorded value

DF508 Mutations; n (%)

Homozygous DF508	4311 (52.0%)
Heterozygous DF508	3205 (38.6%)
No DF508 or both unidentified	778 (9.4%)

All mutations and their classes

All mutations

Current name	New name	Class	N	(%)
DF508	p.Phe508del	II	7516	(90.6)
G551D	p.Gly551Asp	III	466	(5.6)
R117H	p.Arg117His	IV	336	(4.1)
G542X	p.Gly542X	I	295	(3.6)
621+1G->T	c.489+1G>T	I	193	(2.3)
N1303K	p.Asn1303Lys	II	113	(1.4)
1717-1G->A	c.1585-1G>A	I	108	(1.3)
1898+1G->A	c.1766+1G>A	I	104	(1.3)
R560T	p.Arg560Thr	III	82	(1.0)
DI507	p.Ile507del	II	82	(1.0)
3659delC	c.3528delC	II	76	(0.9)
R553X	p.Arg553X	I	73	(0.9)
3849+10kbC->T	c.3717+10kbC>T	V	62	(0.8)
G85E	p.Gly85Glu	IV	57	(0.7)
E60X	p.Glu60X	I	53	(0.6)
D1152H	p.Asp1152His	IV	51	(0.6)
Q493X	p.Gln493X	I	50	(0.6)
W1282X	p.Trp1282X	I	42	(0.5)
1078delT	c.948delT	II	35	(0.4)
2184delA	c.2052delA	II	32	(0.4)
2789+5G->A	c.2657+5G>A	V	28	(0.3)
V520F	p.Val520Phe	III	27	(0.3)
R347P	p.Arg347Pro	IV	22	(0.3)
R1162X	p.Arg1162X	I	21	(0.3)
A455E	p.Ala455Glu	V	21	(0.3)
S549N	p.Ser549Asn	II	19	(0.2)
R347H	p.Arg347His	IV	14	(0.2)
R1158X	p.Arg1158X	I	12	(0.1)
711+1G->T	c.579+1G>T	I	11	(0.1)
3120+1G->A	c.2988+1G>A	I	11	(0.1)
R334W	p.Arg334Trp	IV	9	(0.1)
1161delC	c.1029delC	I	7	(0.1)
I148T	p.Ile148Thr	V	7	(0.1)

L206W	p.Leu206Trp	II	6	*(0.1)*
R352Q	p.Arg352Gln	unknown	6	*(0.1)*
2183delAA->G	c.2051_2052delAAinsG	I	5	*(0.1)*
R1066C	p.Arg1066Cys	unknown	5	*(0.1)*
S549R	p.Ser549Arg	II/III	5	*(0.1)*
A559T	p.Ala559Thr	unknown	4	*(0.1)*
R117C	p.Arg117Cys	IV/V	4	*(0.1)*
R1283M	p.Arg1283Met	unknown	4	*(0.1)*
G178R	p.Gly178Arg	unknown	4	*(0.1)*
Y1092X	p.Tyr1092X	I	4	*(0.1)*
P574H	p.Pro574His	IV	3	*(0.04)*
Y563D	p.Tyr563Asp	unknown	3	*(0.04)*
2143delT	c.490-1G>A	II	3	*(0.04)*
3120G->A	c.2988G>A	V	2	*(0.02)*
2043delG	c.1911delG	unknown	2	*(0.02)*
K710X	p.Lys710X	I	1	*(0.01)*
S549I			1	*(0.01)*
1677delTA	p.Tyr515x	II	1	*(0.01)*
1609delCA	c.1477_1478delCA	unknown	1	*(0.01)*
1898+5G->T	c.1766+5G>T	V	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)*
574delA	c.442delA	unknown	1	*(0.01)*
C524X	p.Cys524X	I	1	*(0.01)*
G330X		I	1	*(0.01)*
Q552X	p.Gln552X	I	1	*(0.01)*
S1251N	p.Ser1251Asn	unknown	1	*(0.01)*
G480C	p.Gly480Cys	unknown	1	*(0.01)*
S364P	p.Ser364Pro	unknown	1	*(0.01)*
W1089X	p.Trp1089X	I	1	*(0.01)*

Other   855 *(10.3)*
Not identified   1025 *(12.4)*

Cystic Fibrosis mutations and their functional effects
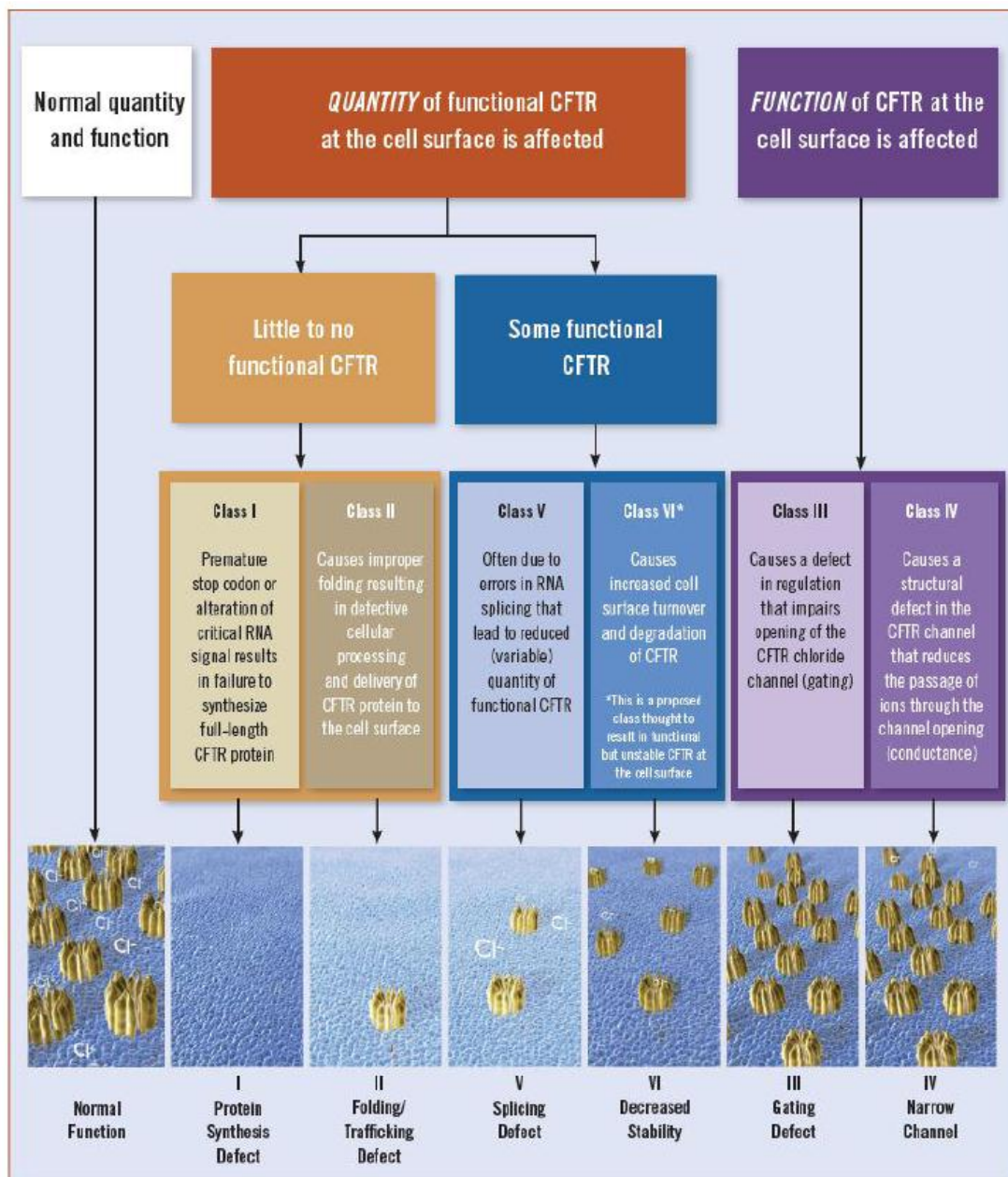
Courtesy of Vertex Pharmaceuticals Incorporated

1.6 Age distribution
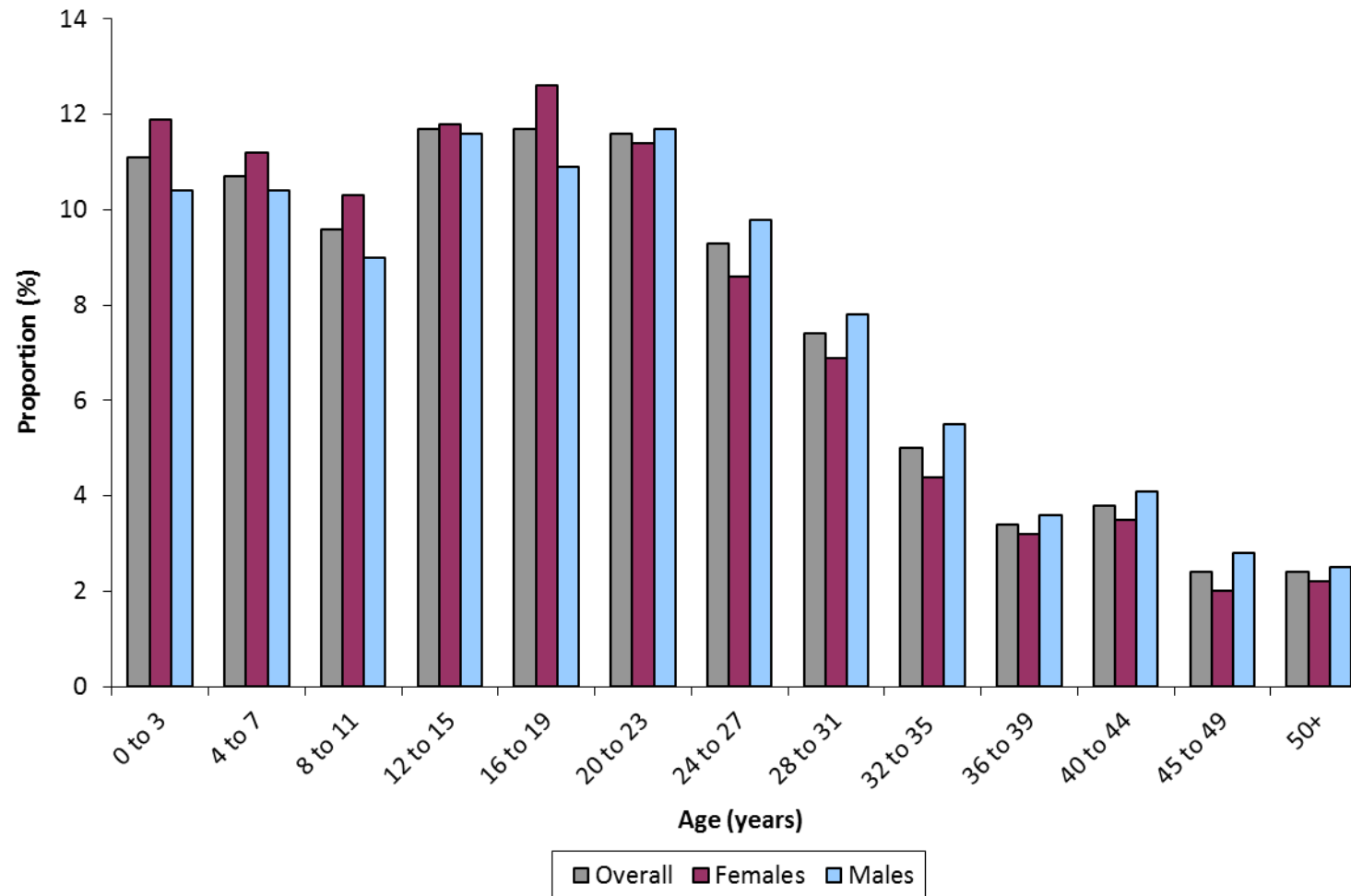
Age is calculated as the age at annual review encounter.

1.7 Age and sex distribution

Age	Overall N=8679	Female N=4058	Male N=4621
0-3 yrs	964 (11.1)	482 (11.9)	482 (10.4)
4-7	932 (10.7)	453 (11.2)	479 (10.4)
8-11	835 (9.6)	419 (10.3)	416 (9.0)
12-15	1015 (11.7)	480 (11.8)	535 (11.6)
16-19	1016 (11.7)	512 (12.6)	504 (10.9)
20-23	1003 (11.6)	461 (11.4)	542 (11.7)
24-27	803 (9.3)	349 (8.6)	454 (9.8)
28-31	642 (7.4)	281 (6.9)	361 (7.8)
32-35	433 (5.0)	180 (4.4)	253 (5.5)
36-39	293 (3.4)	128 (3.2)	165 (3.6)
40-44	328 (3.8)	140 (3.5)	188 (4.1)
45-49	209 (2.4)	82 (2.0)	127 (2.8)
50+	206 (2.4)	91 (2.2)	115 (2.5)
Median (range)	18 (0-81)	17 (0-80)	19 (0-81)

1.8 Age distribution by sex

Females

45% < 16 yrs
55% 16 yrs +

Males

41% < 16 yrs
59% 16 yrs +
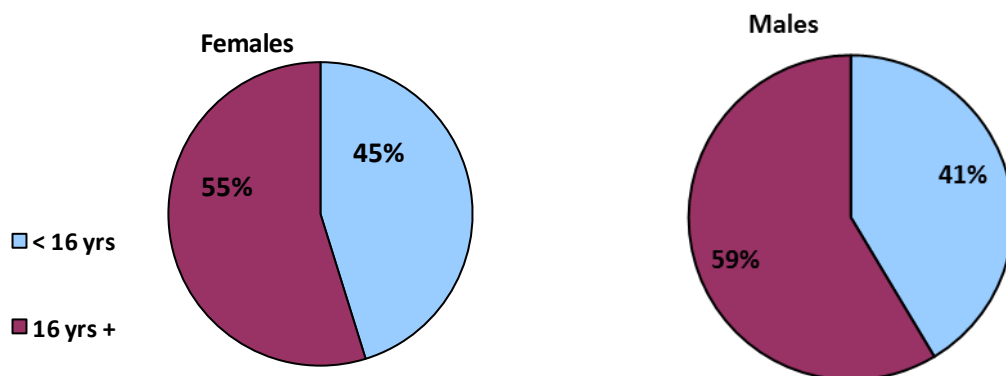
1.9 Employment and education status among adults aged 16 years and older

	Number of patients
Full-time working	1436
Part-time working	706
Student	933
Homemaker	216
Unemployed	793
"Disabled"	255
Retired	78
Unknown	548
No data	*624*

Note that these groups are not mutually exclusive.

Of the 4309 adults aged 16 years and older for whom an employment status questionnaire was completed (excluding "unknown"), <u>3014 (70.0%) reported being in work or study</u>.

1.10 Median height percentiles among children (n=3221)
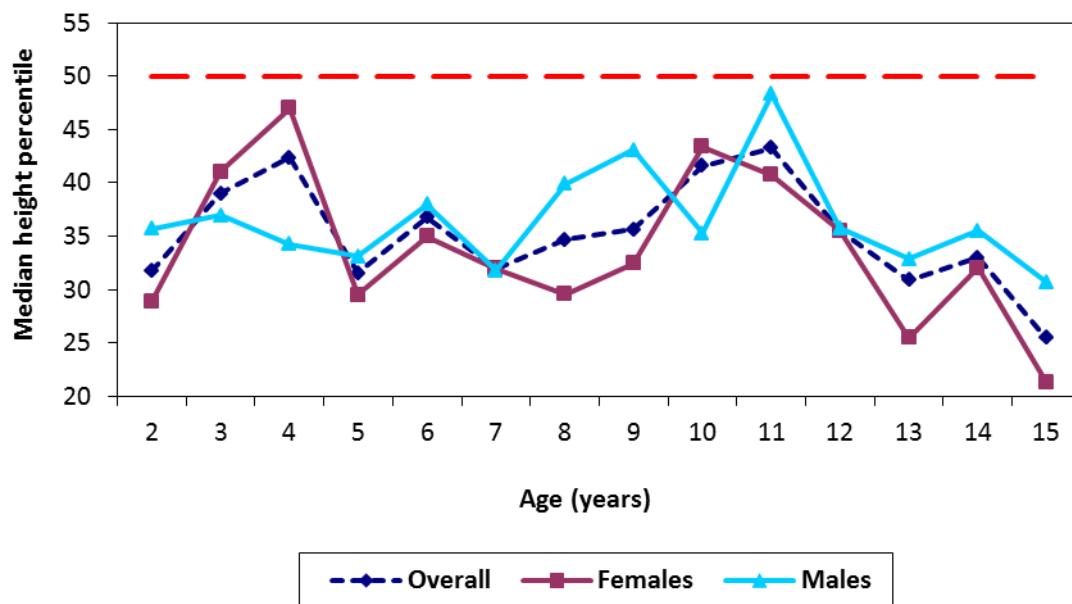
Age	Overall		Female		Male	
	N	**Median (IQR)**	**N**	**Median (IQR)**	**N**	**Median (IQR)**
2	224	31.7 (14.2-57.5)	106	28.8 (12.5-54.9)	118	35.7 (18.7-59.4)
3	260	39.0 (16.1-65.2)	137	41.0 (17.2-65.8)	123	36.9 (15.4-65.2)
4	246	42.4 (17.2-65.0)	113	47.0 (18.7-70.6)	133	34.2 (15.2-63.0)
5	227	31.5 (13.6-57.1)	107	29.5 (13.6-52.5)	120	33.1 (13.5-62.8)
6	213	36.8 (16.8-66.1)	98	35.0 (17.0-61.3)	115	38.0 (16.1-68.8)
7	234	32.0 (14.4-58.1)	128	32.0 (15.3-58.9)	106	31.8 (13.4-57.2)
8	198	34.7 (14.1-62.3)	105	29.6 (14.3-59.2)	93	39.9 (13.4-69.2)
9	208	35.7 (15.0-64.9)	107	32.4 (10.7-64.5)	101	43.1 (18.1-65.3)
10	206	41.6 (15.7-68.5)	98	43.4 (19.1-69.6)	108	35.2 (11.9-66.9)
11	210	43.3 (15.4-73.6)	105	40.7 (12.9-65.2)	105	48.3 (19.3-79.8)
12	258	35.6 (11.6-65.5)	126	35.5 (11.5-66.1)	132	35.7 (11.5-64.7)
13	221	30.9 (11.5-60.3)	91	25.4 (8.7-64.2)	130	32.9 (13.5-58.0)
14	271	33.0 (12.0-60.1)	126	32.0 (8.6-53.9)	145	35.5 (14.5-66.9)
15	245	25.5 (9.9-55.9)	127	21.3 (6.5-53.0)	118	30.7 (13.2-62.2)
Overall	3221	34.8 (14.1-63.3)	1574	33.6 (13.3-61.1)	1647	35.7 (14.8-64.5)
2-4 yrs	730	36.8 (16.0-63.3)	356	38.0 (16.4-64.0)	374	36.6 (15.9-63.1)
5-7 yrs	674	33.0 (14.9-59.3)	333	31.9 (15.2-57.7)	341	34.3 (14.5-63.8)
8-10 yrs	612	36.7 (15.2-65.1)	310	35.1 (14.9-64.3)	302	38.9 (15.3-66.9)
11-13 yrs	689	36.0 (12.9-65.4)	322	35.5 (11.4-65.1)	367	37.5 (14.7-66.5)
14-15 yrs	516	30.7 (10.8-59.1)	253	28.2 (6.9-53.4)	263	33.2 (13.7-64.6)

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=3236)
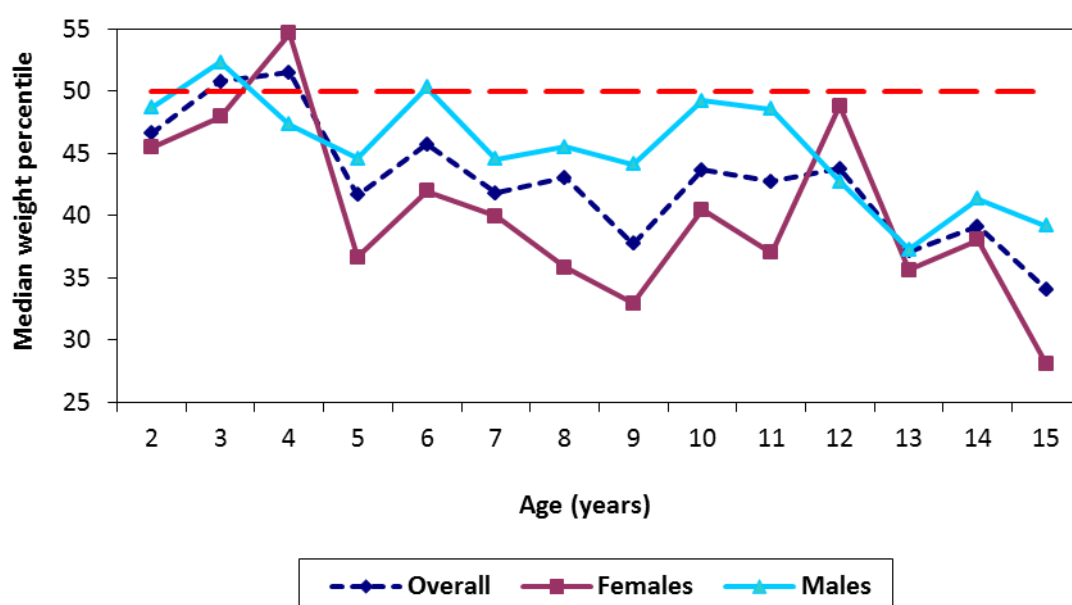
Age	Overall		Female		Male	
	N	Median (IQR)	N	Median (IQR)	N	Median (IQR)
2	227	46.6 (19.2-70.2)	106	45.5 (17.4-66.2)	121	48.7 (22.0-73.8)
3	264	50.8 (22.7-75.2)	139	47.9 (22.7-71.6)	125	52.3 (22.6-79.2)
4	247	51.5 (24.2-75.7)	114	54.7 (23.8-76.8)	133	47.3 (24.3-74.9)
5	228	41.6 (18.7-64.9)	108	36.6 (17.4-60.7)	120	44.6 (21.6-73.5)
6	214	45.7 (21.3-71.5)	99	42.0 (18.9-68.0)	115	50.3 (25.4-72.1)
7	234	41.8 (19.1-69.1)	128	39.9 (19.0-71.0)	106	44.5 (19.6-67.4)
8	199	43.1 (20.2-60.3)	105	35.8 (15.9-57.2)	94	45.5 (28.5-70.0)
9	208	37.7 (20.0-69.1)	107	32.9 (16.2-64.3)	101	44.1 (23.3-74.7)
10	206	43.7 (21.8-73.3)	98	40.5 (21.0-67.7)	108	49.2 (23.8-81.3)
11	210	42.7 (15.8-77.0)	105	37.0 (13.4-70.2)	105	48.6 (25.1-80.6)
12	259	43.8 (16.4-69.7)	126	48.8 (14.3-75.3)	133	42.7 (17.8-66.0)
13	222	37.1 (15.4-61.9)	92	35.6 (14.2-66.7)	130	37.3 (15.7-61.9)
14	273	39.1 (17.2-68.8)	128	38.0 (16.4-68.5)	145	41.4 (17.8-68.9)
15	245	34.1 (13.7-68.7)	127	28.1 (10.1-66.8)	118	39.2 (17.6-71.7)
Overall	3236	42.9 (18.9-70.5)	1582	40.7 (17.1-68.7)	1654	44.8 (21.3-72.6)
2-4 yrs	738	49.2 (22.3-73.9)	359	49.5 (21.3-73.5)	379	48.8 (22.6-75.1)
5-7 yrs	676	43.4 (19.3-69.8)	335	39.8 (18.7-66.6)	341	46.0 (20.7-71.2)
8-10 yrs	613	42.1 (21.0-68.6)	310	36.8 (17.8-62.7)	303	47.0 (25.0-74.6)
11-13 yrs	691	41.5 (15.7-70.3)	323	38.2 (13.7-71.8)	368	42.3 (18.1-69.0)
14-15 yrs	518	37.2 (15.9-68.4)	255	35.6 (13.5-66.8)	263	41.1 (17.9-70.1)

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.
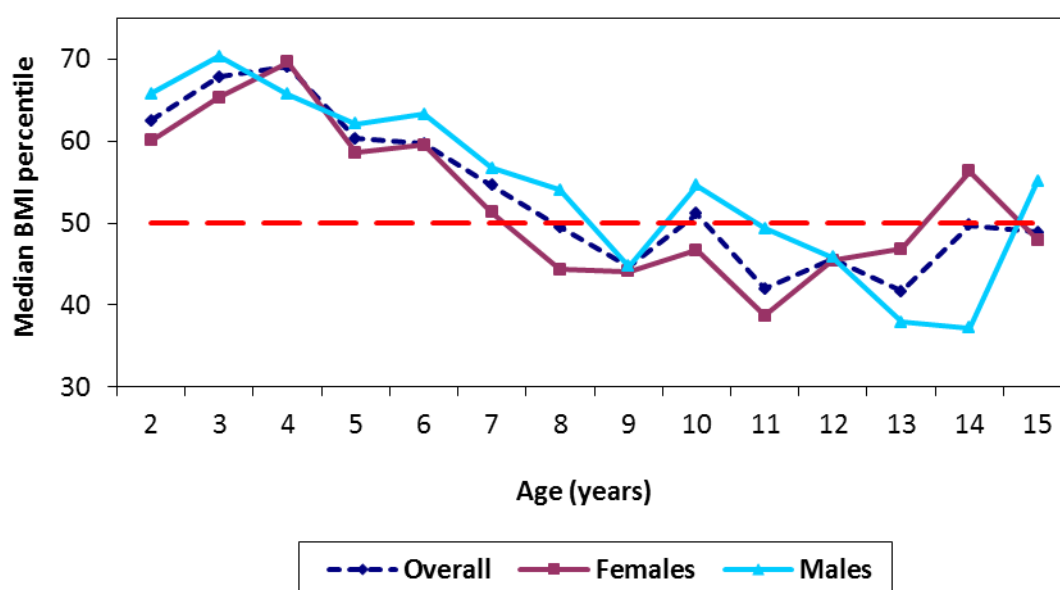
1.12 Median BMI percentiles among children (n=3104)

Age	Overall		Female		Male	
	N	Median (IQR)	N	Median (IQR)	N	Median (IQR)s
2	224	62.5 (31.3-80.6)	106	60.1 (30.1-78.6)	118	65.8 (34.4-81.1)
3	259	67.8 (34.0-86.0)	136	65.3 (34.9-84.0)	123	70.3 (31.7-88.5)
4	246	69.1 (43.6-85.9)	113	69.6 (46.1-87.5)	133	65.7 (38.9-83.2)
5	227	60.3 (32.4-78.2)	107	58.6 (32.1-76.0)	120	62.1 (34.4-81.2)
6	213	59.7 (38.8-77.2)	98	59.5 (38.8-74.8)	115	63.3 (38.5-79.5)
7	234	54.6 (30.8-75.3)	128	51.3 (25.7-77.1)	106	56.7 (34.5-73.3)
8	198	49.5 (29.3-70.4)	105	44.3 (27.1-66.2)	93	54.0 (32.6-73.6)
9	208	44.6 (23.4-71.0)	107	44.1 (26.1-70.6)	101	44.7 (20.8-77.2)
10	206	51.1 (24.8-72.4)	98	46.7 (19.8-65.1)	108	54.6 (27.2-77.6)
11	210	42.0 (19.1-69.5)	105	38.7 (19.1-70.8)	105	49.3 (19.5-69.5)
12	258	45.8 (22.4-68.0)	126	45.4 (24.0-70.6)	132	45.8 (21.2-64.7)
13	221	41.7 (22.4-66.9)	91	46.8 (27.4-68.9)	130	37.9 (18.8-65.6)
14	271	49.7 (24.0-73.0)	126	56.3 (32.8-74.7)	145	37.2 (18.8-71.4)
15	129	48.9 (22.6-75.8)	66	47.9 (24.1-74.9)	63	55.1 (20.0-76.9)
Overall	3104	53.6 (27.8-76.7)	1512	53.7 (28.4-75.9)	1592	53.5 (27.1-77.5)
2-4 yrs	729	66.7 (37.4-83.5)	355	66.2 (38.3-82.8)	374	67.7 (35.0-84.8)
5-7 yrs	674	58.6 (33.9-77.1)	333	56.6 (31.7-75.9)	341	60.6 (34.9-78.1)
8-10 yrs	612	47.5 (26.2-71.3)	310	44.5 (24.8-66.5)	302	52.0 (27.5-76.2)
11-13 yrs	689	42.9 (21.0-68.1)	322	43.2 (21.8-69.0)	367	42.8 (20.3-66.5)
14-15 yrs	400	49.3 (23.4-73.5)	192	52.6 (31.1-74.5)	208	44.2 (19.5-72.4)

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.

1.13 Median BMI values among adults (n=4849)
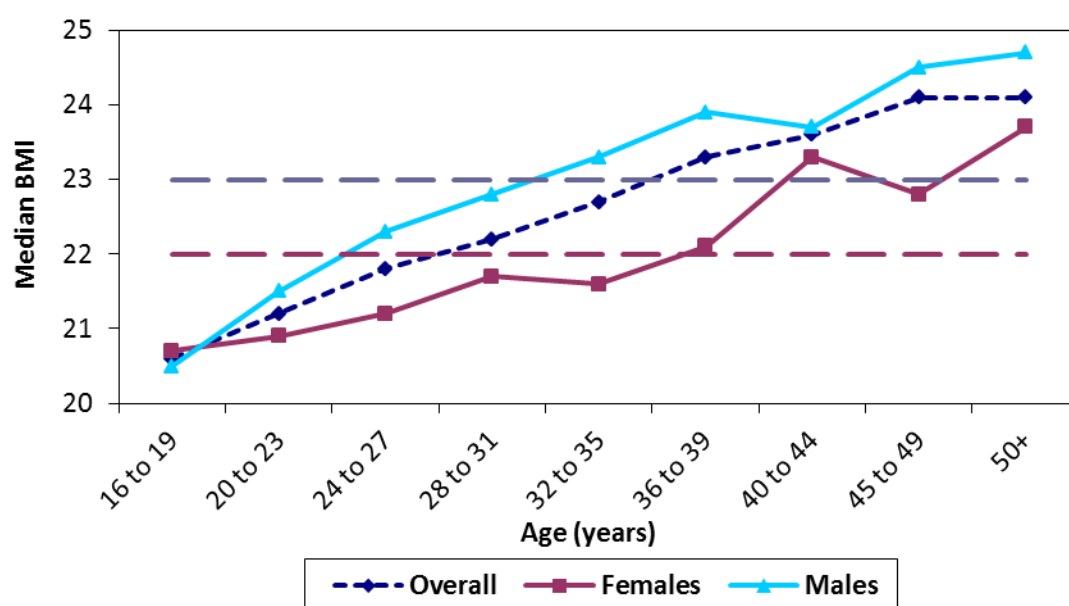
Age	Overall		Female		Male	
	N	Median (IQR)	N	Median (IQR)	N	Median (IQR)
16-19	1000	20.6 (18.8-22.6)	503	20.7 (18.9-22.5)	497	20.5 (18.6-22.6)
20-23	986	21.2 (19.4-23.2)	451	20.9 (19.3-22.7)	535	21.5 (19.5-23.6)
24-27	791	21.8 (19.9-23.9)	346	21.2 (19.4-23.2)	445	22.3 (20.2-24.2)
28-31	630	22.2 (20.3-24.7)	280	21.7 (19.8-23.9)	350	22.8 (20.7-25.2)
32-35	426	22.7 (20.6-24.8)	176	21.6 (20.1-24.2)	250	23.3 (21.4-25.3)
36-39	287	23.3 (21.1-25.9)	126	22.1 (20.6-25.4)	161	23.9 (22.0-26.0)
40-44	322	23.6 (21.7-26.0)	139	23.3 (21.0-25.8)	183	23.7 (22.0-26.0)
45-49	204	24.1 (21.2-26.4)	80	22.8 (20.4-25.7)	124	24.5 (22.1-26.7)
50+	203	24.1 (21.8-27.2)	89	23.7 (21.2-27.4)	114	24.7 (22.4-27.1)
Overall	4849	21.9 (19.9-24.3)	2190	21.4 (19.6-23.6)	2659	22.4 (20.2-24.8)

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=6902)
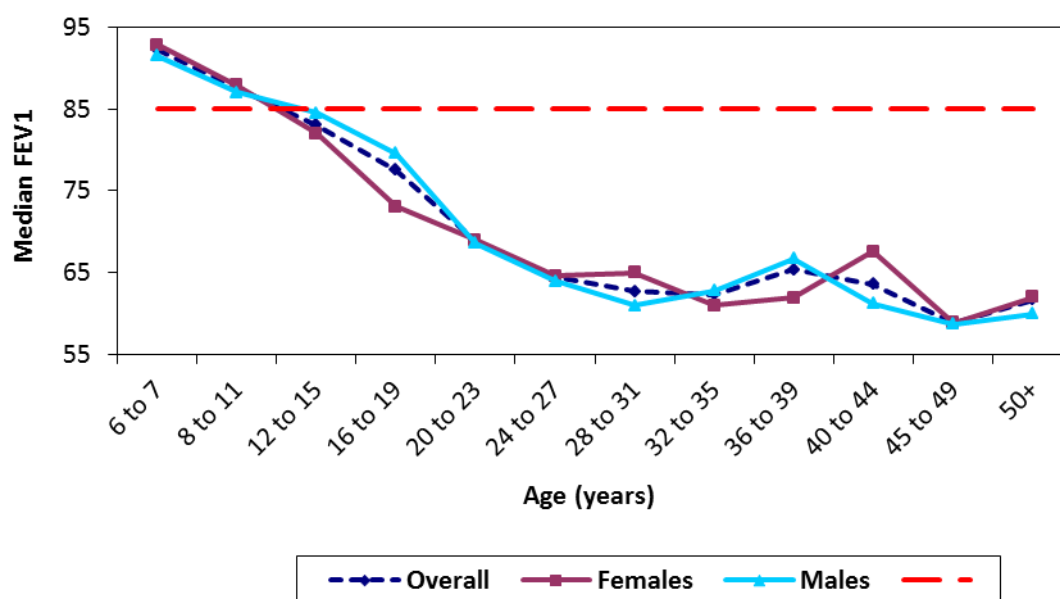
| Age	Overall		Female		Male	
	N	Median (IQR)	N	Median (IQR)	N	Median (IQR)
6-7	403	92.3 (83.1-102.7)	203	92.8 (83.7-103.1)	200	91.5 (83.0-102.4)
8-11	770	87.5 (76.3-96.4)	389	87.9 (76.6-96.7)	381	87.0 (75.7-96.1)
12-15	941	83.0 (69.0-95.6)	444	82.0 (66.5-95.2)	497	84.5 (71.1-96.2)
16-19	981	77.5 (58.4-91.6)	495	73.1 (55.5-88.3)	486	79.6 (61.4-96.0)
20-23	975	68.8 (49.1-87.1)	445	69.0 (48.8-88.0)	530	68.6 (49.9-86.5)
24-27	779	64.5 (44.5-83.1)	340	64.6 (44.9-86.9)	439	64.0 (44.0-81.6)
28-31	627	62.7 (44.9-80.9)	277	65.0 (47.7-81.0)	350	61.0 (40.9-80.5)
32-35	421	62.3 (46.0-81.6)	172	61.0 (46.2-78.5)	249	62.8 (45.9-84.6)
36-39	285	65.4 (47.0-82.9)	126	61.9 (47.0-83.9)	159	66.7 (47.0-82.6)
40-44	315	63.6 (43.8-83.0)	135	67.6 (44.3-85.7)	180	61.2 (43.3-81.3)
45-49	202	58.8 (40.5-76.1)	79	58.9 (39.8-75.9)	123	58.7 (40.6-77.2)
50+	203	61.7 (40.6-83.6)	89	62.0 (39.9-78.1)	114	60.0 (41.3-87.2)
Overall	6902	74.7 (54.3-91.1)	3194	74.7 (54.7-91.5)	3708	74.7 (53.8-90.8)

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.

1.15 Median FEV$_1$ (% predicted) among patients aged 6 years and older by year since 2008
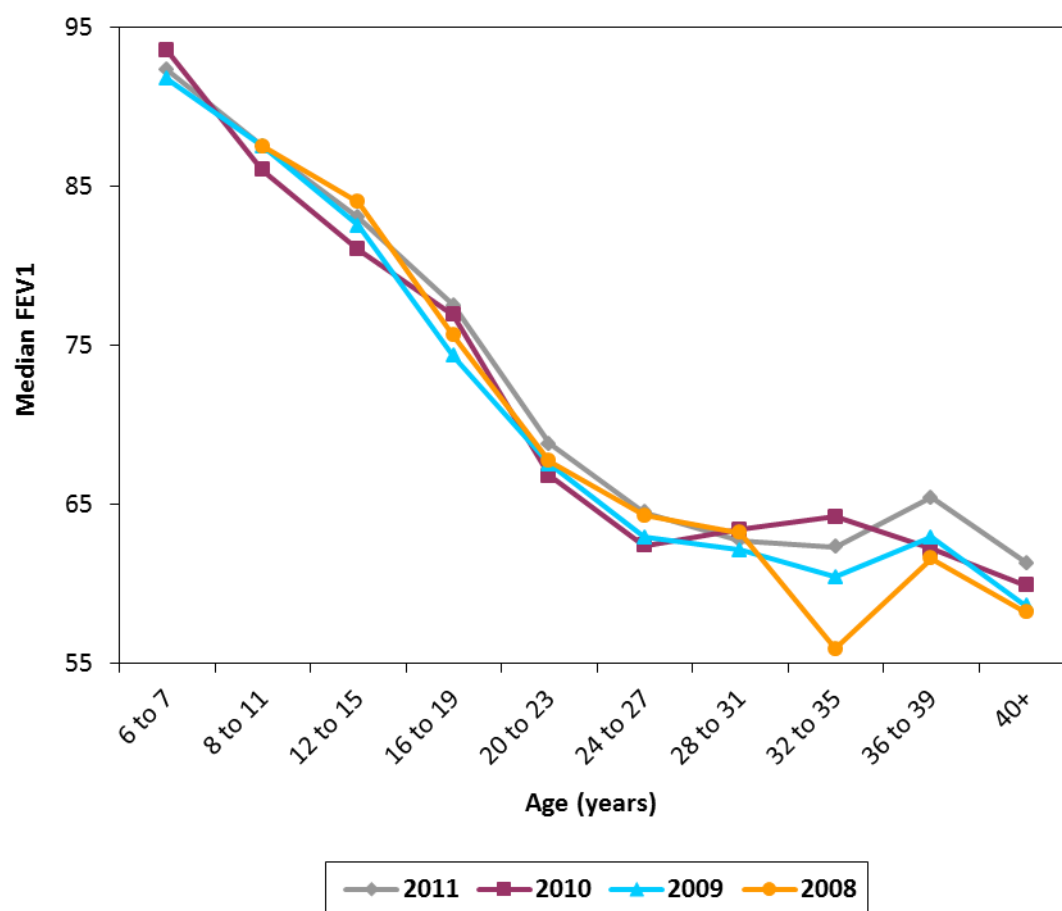
1.16 Median FEV$_1$ (% predicted) vs BMI among patients aged 16 years and older
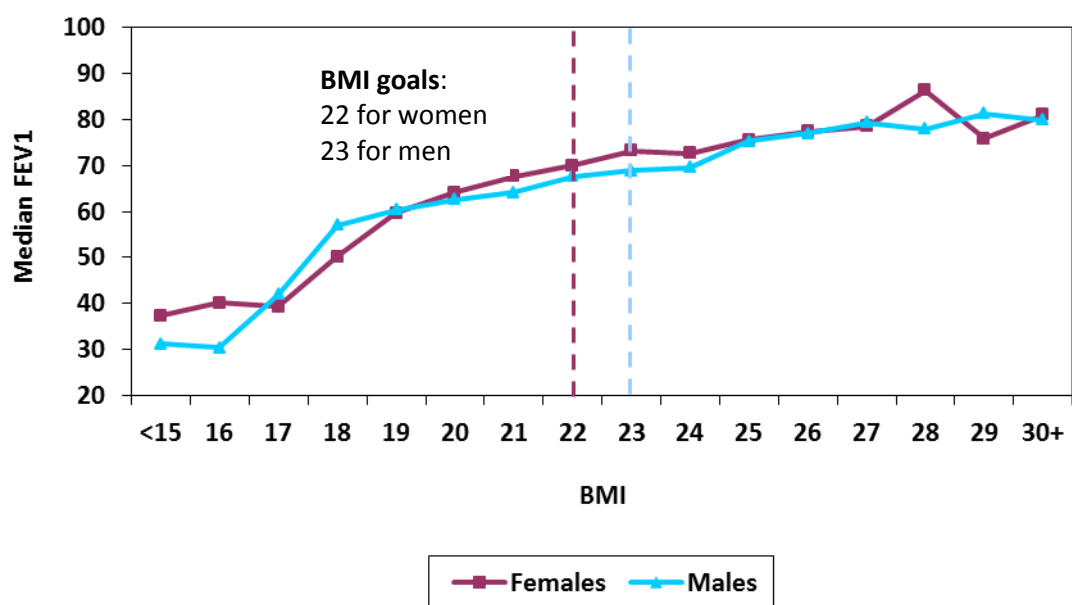
Each point represents the median FEV$_1$ % predicted of patients for each given BMI value. Due to the wide range of BMIs in this population we grouped all BMI≤15 into one group and BMI≥30 into another.

1.17 Lung infections in 2011

	0 to 3	4 to 7	8 to 11	12 to 15	16 to 19	20 to 23	24 to 27	28 to 31	32 to 35	36 to 39	40 to 44	45 to 49	50+
N in age band	964	932	835	1015	1016	1003	803	642	433	293	328	209	206
N cultures taken	939	919	823	997	987	969	768	614	411	270	297	191	193
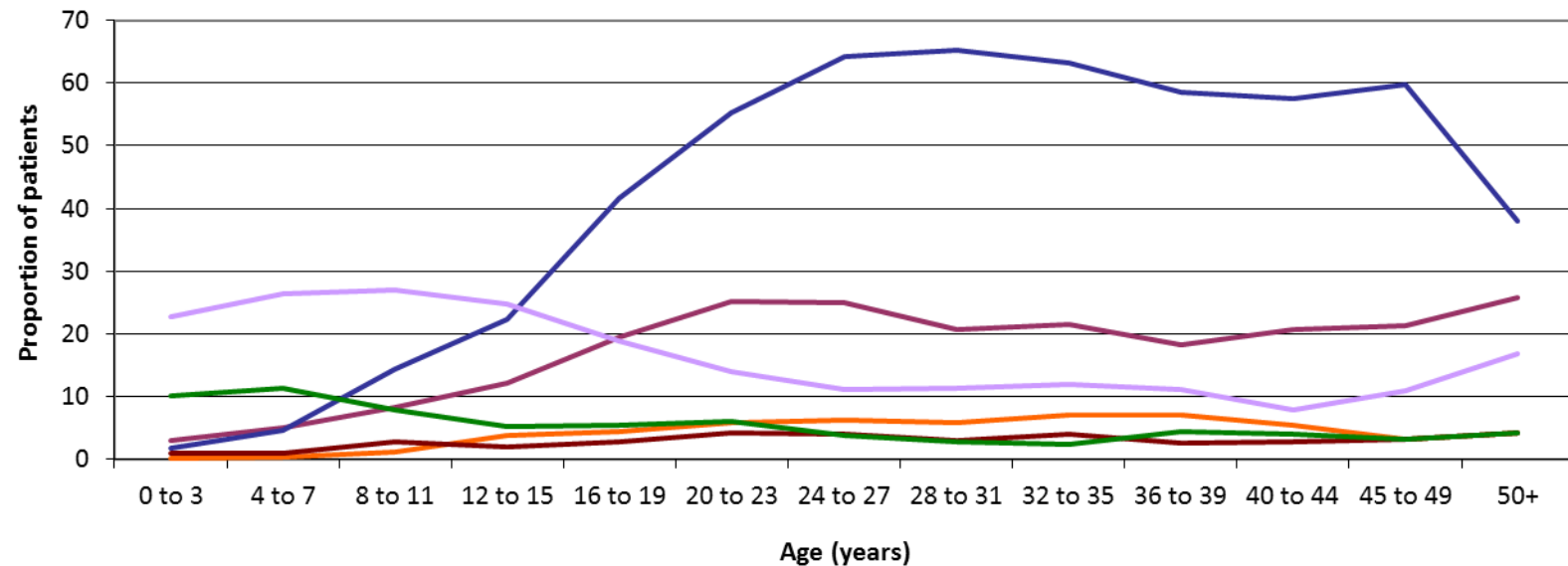
— Chronic *S.aureus*; n=1312 (15.7%)   — Chronic *P.aeruginosa*; n=3058 (36.5%)   — Intermittent *P.aeruginosa*; n=1547 (18.4%)

— *B.cepacia*; n=314 (3.8%)   — MRSA; n=221 (2.6%)   — *H.influenzae*; n=523 (6.2%)

Chronic and intermittent 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%.

Lung infections in 2011

|	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	964	932	835	1015	1016	1003	803	642	433	293	328	209	206	8679	3746	4933
N cultures taken at clinic	939	919	823	997	987	969	768	614	411	270	297	191	193	8378	3678	4700
Chronic *S.aureus*; n(%)	28 (3.0)	46 (5.1)	66 (8.2)	119 (12.2)	187 (19.4)	243 (25.1)	193 (24.9)	129 (20.6)	89 (21.4)	52 (18.3)	65 (20.7)	43 (21.3)	52 (25.7)	1312 (15.7)	259 (7.2)	1053 (22.2)
Chronic *P.aeruginosa*; n(%)	16 (1.7)	43 (4.7)	116 (14.3)	217 (22.3)	402 (41.7)	540 (55.3)	502 (64.2)	411 (65.3)	265 (63.3)	167 (58.6)	182 (57.6)	120 (59.7)	77 (37.9)	3058 (36.5)	392 (10.9)	2666 (55.8)
Intermittent *P.aeruginosa*; n(%)	209 (22.7)	239 (26.4)	219 (27.0)	241 (24.7)	181 (18.8)	137 (14.0)	87 (11.1)	71 (11.3)	50 (11.9)	32 (11.2)	25 (7.9)	22 (11.0)	34 (16.8)	1547 (18.4)	908 (25.1)	639 (13.4)
B.cepacia; n(%)	2 (0.2)	4 (0.4)	9 (1.1)	37 (3.7)	44 (4.5)	56 (5.8)	48 (6.3)	36 (5.9)	29 (7.1)	19 (7.0)	16 (5.4)	6 (3.1)	8 (4.2)	314 (3.8)	52 (1.4)	262 (5.6)
MRSA; n(%)	9 (1.0)	9 (1.0)	22 (2.7)	20 (2.0)	27 (2.7)	41 (4.2)	30 (3.9)	18 (2.9)	16 (3.9)	7 (2.6)	8 (2.7)	6 (3.1)	8 (4.2)	221 (2.6)	60 (1.6)	161 (3.4)
H.influenza; n(%)	95 (10.1)	105 (11.4)	65 (7.9)	53 (5.3)	53 (5.4)	58 (6.0)	29 (3.8)	17 (2.8)	10 (2.4)	12 (4.4)	12 (4.0)	6 (3.1)	8 (4.2)	523 (6.2)	318 (8.7)	205 (4.4)

Age is calculated as age at annual review

1.18 Trends in lung infections since 2008
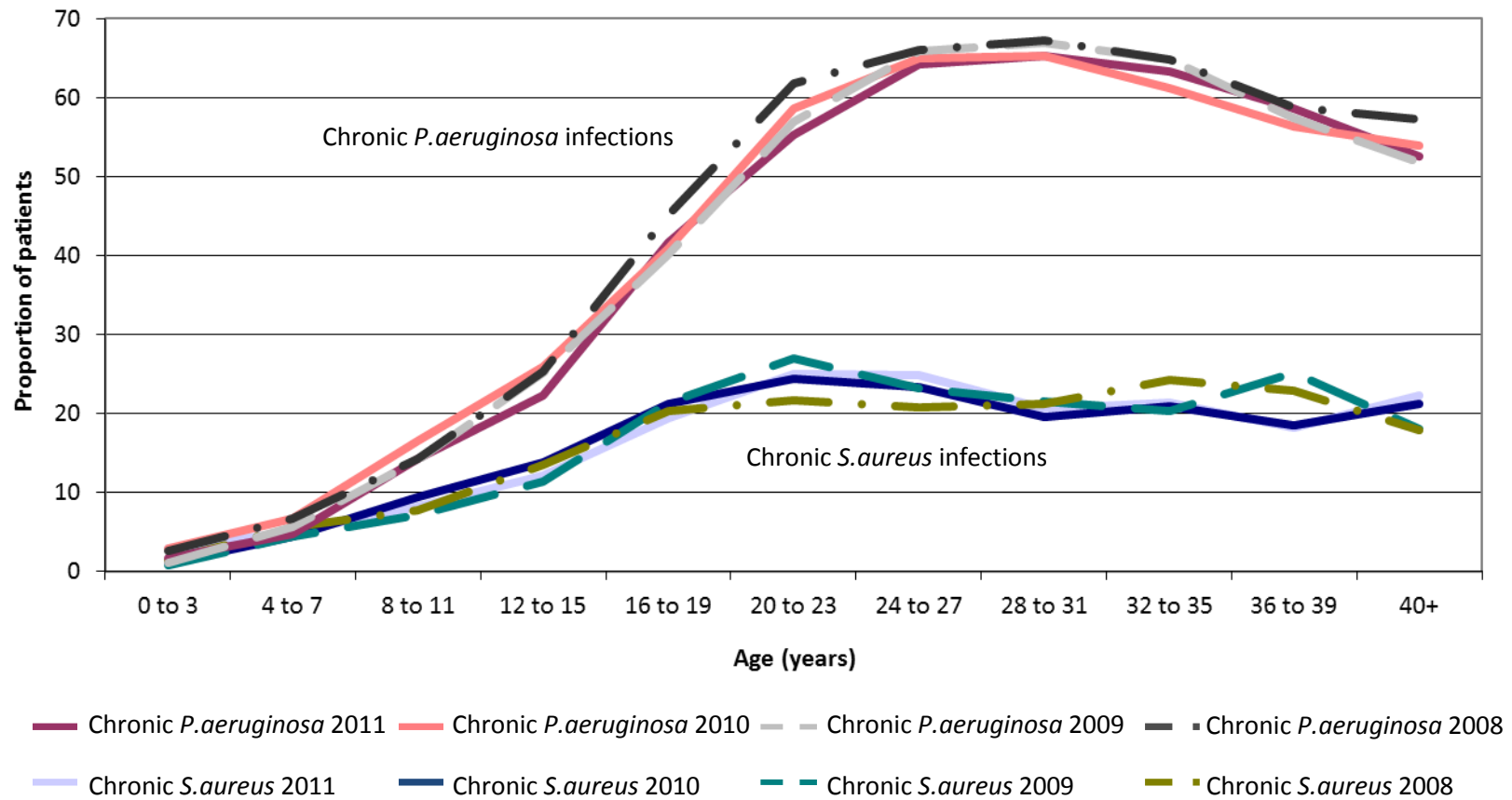
1.19 Prevalence of complications

	Overall (n=8679)	<16 years (n=3746)	≥16 years (n=4933)
Nontuberculous mycobacteria or atypical mycobacteria; *n(%)*	338 (3.9)	87 (2.3)	251 (5.1)
*Newly identified in 2011**	*131 (1.5)*	*33 (0.9)*	*98 (2.0)*
Cirrhosis with no portal hypertension; *n(%)*	113 (1.3)	22 (0.6)	91 (1.8)
*Newly identified in 2011**	*44 (0.5)*	*10 (0.3)*	*34 (0.7)*
Cirrhosis with portal hypertension; *n(%)*	171 (2.0)	22 (0.6)	149 (3.0)
*Newly identified in 2011**	*34 (0.4)*	*8 (0.2)*	*26 (0.5)*
Gallbladder disease requiring surgery; *n(%)*	42 (0.5)	2 (0.1)	40 (0.8)
Nasal polyps requiring surgery; *n(%)*	252 (2.9)	54 (1.4)	198 (4.0)
Pneumothorax requiring chest tube; *n(%)*	43 (0.5)	2 (0.1)	41 (0.8)
Cancer confirmed by histology; *n(%)*	14 (0.2)	3 (0.1)	11 (0.2)
Fibrosing colonopathy/ colonic stricture; *n(%)*	4 (0.05)	2 (0.1)	2 (0.04)
ABPA; *n(%)*	847 (9.8)	293 (7.8)	554 (11.2)
*Newly identified in 2011**	*190 (2.2)*	*78 (2.1)*	*112 (2.3)*
Port inserted or replaced; *n(%)*	583 (6.7)	211 (5.6)	372 (7.5)

* For patients who are reported to have had non-tuberculous mycobacteria/atypical mycobacteria, cirrhosis (with/without portal hypertension) or ABPA in 2011 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 2011 was the year the complication first developed.

1.20 CF-related diabetes

	Overall* (n=8561)	<16 years * (n=3669)	≥16 years * (n=4892)
Treatment for CF-related diabetes; n(%)	1569 (18.3)	131 (3.6)	1438 (29.4)

* *Treatment for CF-related diabetes was enquired about in an annual review questionnaire which was completed by 8561 of the 8679 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

	2011	2010	2009
Number of patients that year with annual review data evaluated for transplants	204	169	143
Number accepted on the transplant list	121	82	79
Number receiving transplants **Types of transplants received:**	51*	29	25*
Bilateral lung	43	26	19
Heart and lung	4	1	0
Liver	2	1	5
Kidney	2	1	2
Pancreas	1	-	-

One patient received two transplants

1.22 Other therapy

	Overall (n=8679)	<16 years (n=3746)	≥16 years (n=4933)
NIV; n(%)	197 (2.4)	39 (1.1)	158 (3.3)
Long-term oxygen; n(%)	568 (6.8)	106 (2.9)	462 (9.6)
Among those who had long-term oxygen therapy:			
Continuously	111 (19.5)	7 (6.6)	104 (22.5)
Nocturnal+exertion	156 (27.5)	27 (25.5)	129 (27.9)
PRN	85 (15.0)	10 (9.4)	75 (16.2)
With exacerbation	216 (38.0)	62 (58.5)	154 (33.3)

1.23 Feeding

	All patients (n=8679)	<16 years (n=3746)	≥16 years (n=4933)
Any supplemental feeding; n(%)	2684 (30.9)	967 (25.8)	1717 (34.8)
Nasogastric Tube	112 (4.2)	20 (2.1)	92 (5.4)
Gastrostomy Tube / Button	540 (20.1)	209 (21.6)	331 (19.3)
Jejunal	6 (0.2)	1 (0.1)	5 (0.3)
TPN	3 (0.1)	1 (0.1)	2 (0.1)

1.24 Days on IV antibiotics

Age	Home		Hospital		Total	
	N (%)	Median (IQR)	N (%)	Median (IQR)	N (%)	Median (IQR)
0-3	54 (5.9)	13 (7-25)	277 (30.0)	14 (10-20)	289 (31.3)	14 (11-25)
4-7	138 (15.1)	14 (11-36)	289 (31.7)	14 (8-25)	327 (35.9)	15 (14-42)
8-11	188 (23.1)	21 (12-40)	328 (40.3)	14 (9-28)	374 (46.0)	28 (14-45)
12-15	276 (27.8)	28 (14-45)	445 (44.9)	16 (10-42)	518 (52.2)	40 (14-56)
16-19	346 (35.1)	16 (11-39)	474 (48.0)	16 (8-41)	571 (57.9)	28 (14-56)
20-23	400 (40.6)	25 (14-42)	512 (51.9)	18 (10-40)	615 (62.4)	34 (14-61)
24-27	330 (41.4)	24 (14-42)	395 (49.5)	18 (10-36)	512 (64.2)	28 (14-56)
28-31	285 (44.7)	28 (14-49)	295 (46.3)	15 (7-34)	397 (62.3)	35 (15-58)
32-35	197 (46.2)	27 (14-45)	187 (43.9)	14 (7-28)	260 (61.0)	28 (14-55)
36-39	93 (32.2)	28 (14-52)	104 (36.0)	14 (7-31)	139 (48.1)	28 (14-59)
40-44	112 (34.6)	21 (14-35)	110 (34.0)	16 (10-28)	163 (50.3)	27 (14-42)
45-49	64 (31.2)	18 (12-42)	82 (40.0)	14 (9-28)	101 (49.3)	30 (14-56)
50+	50 (24.4)	20 (12-42)	64 (31.2)	14 (8-28)	79 (38.5)	28 (14-46)
Overall	2533 (29.8)	23 (14-42)	3562 (41.9)	14 (9-31)	4345 (51.1)	28 (14-54)

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**
0-3 yrs	46 (7.6)	55 (6.9)	87 (9.5)	97 (10.1)
4-7	125 (20.1)	152 (20.5)	172 (21.9)	280 (30.0)
8-11	227 (34.2)	332 (39.6)	367 (42.8)	395 (47.3)
12-15	359 (46.4)	449 (48.2)	528 (54.5)	587 (57.8)
16-19	377 (49.5)	441 (50.3)	509 (54.0)	575 (56.6)
20-23	319 (44.0)	410 (47.6)	491 (53.5)	576 (57.4)
24-27	288 (47.6)	345 (50.0)	389 (52.9)	437 (54.4)
28-31	182 (43.4)	226 (45.0)	304 (52.7)	358 (55.8)
32-35	108 (41.5)	127 (39.1)	155 (44.8)	211 (48.7)
36-39	83 (35.0)	102 (37.0)	127 (44.1)	120 (41.0)
40-44	147 (35.7)*	194 (36.3) *	123 (43.5)	132 (40.2)
45-49			73 (45.3)	100 (47.9)
50+			66 (42.0)	75 (36.4)
Overall	2261 (37.2)	2833 (38.4)	3391 (42.7)	3943 (45.4)

Antibiotic use among patients with chronic *Pseudomonas aeruginosa*

	Overall	<16 years	≥16 years
Patients with chronic pseudomonas	3058	392	2666
Tobramycin solution; n(%)	910 (29.8)	109 (27.8)	801 (30.0)
Other aminoglycoside; n(%)	64 (2.1)	13 (3.3)	51 (1.9)
Colistin; n(%)	1302 (42.6)	211 (53.8)	1091 (40.9)
Promixin; n(%)	857 (28.0)	114 (29.1)	743 (27.9)
At least one of the above; n(%)	2515 (82.2)	354 (90.3)	2161 (81.1)

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.

Section 2: Analyses by paediatric care centre/clinic

(based on 4081 patients from paediatric care centres with complete* data at 2011 annual review)

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

Figure 2.1 Median FEV$_1$ % predicted by paediatric centre/clinic
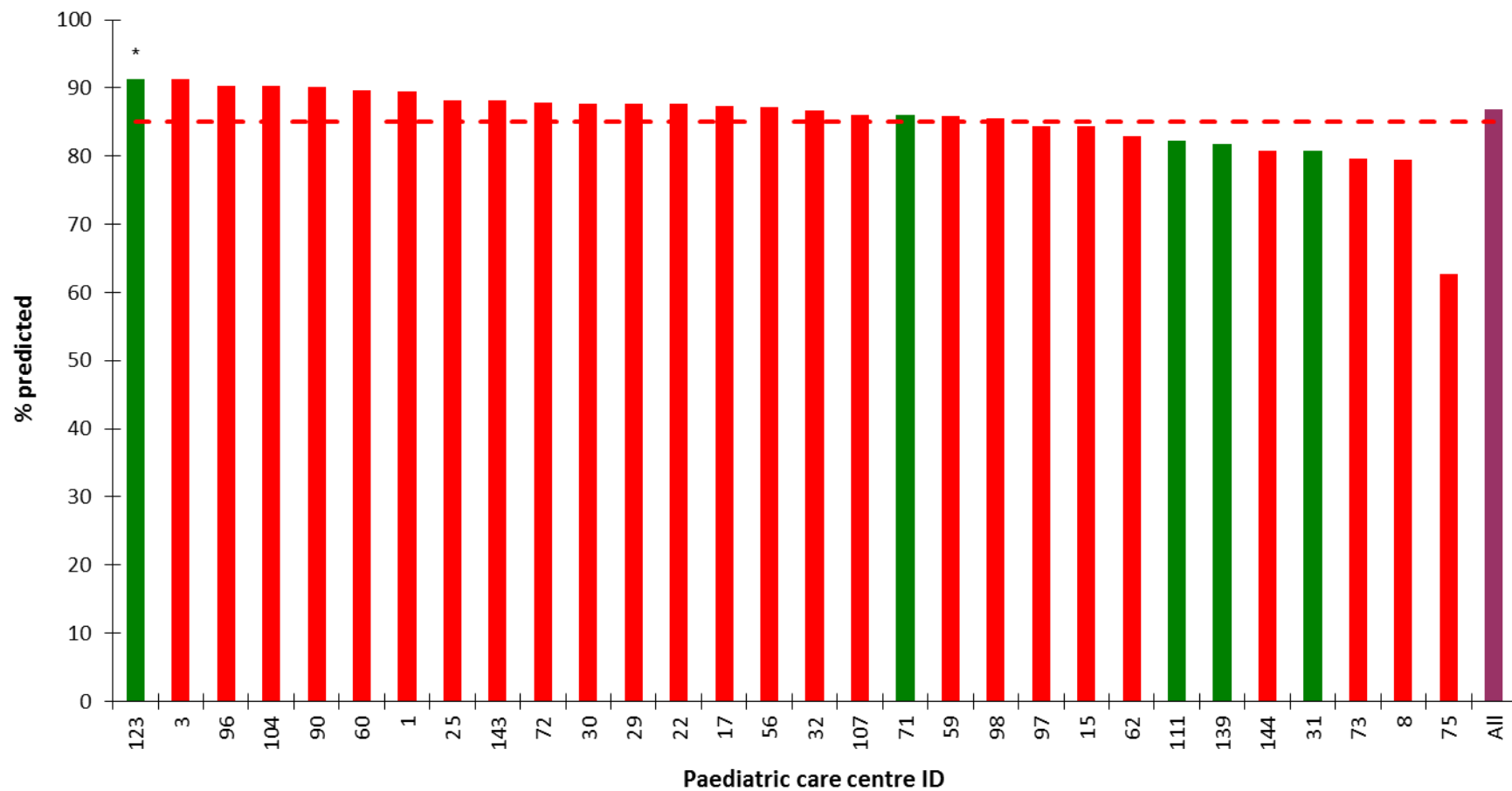
The median FEV1 % predicted for paediatric centres/clinics is 86.5% predicted (IQR: 73.3-97.2).
*Red: centres with their network clinics. Green: stand-alone clinics . Plum: all. * Centre/clinic with a dataset submission of less than 20 patients.*

Figure 2.2 Median BMI percentile by paediatric centre/clinic
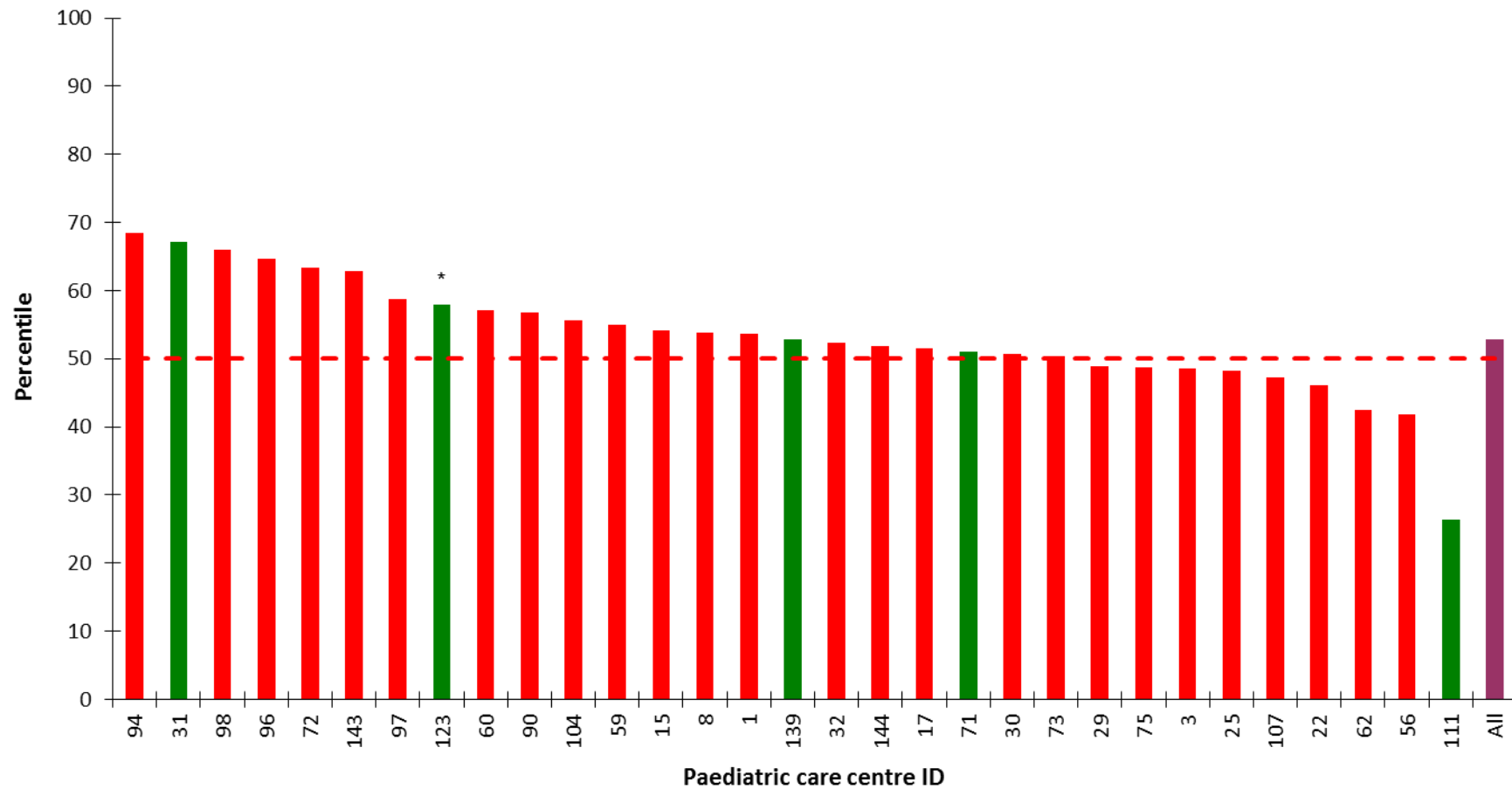
The median BMI percentile for paediatric centres/clinics is 53.5 (IQR: 27.8 – 76.7).
*Red: centres with their network clinics. Green: stand-alone clinics . Plum: all. * Centre/clinic with a dataset submission of less than 20 patients.*

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

The proportion of patients with chronic *P.aeruginosa* for paediatric centres/clinics is 12.6%.

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

Section 3: Analyses by Adult Service

(based on 4598 patients from adult services with complete* data at 2011 annual review)

* "Complete" data refers to the minimum data required to produce the range of clinical outcomes presented in this report.
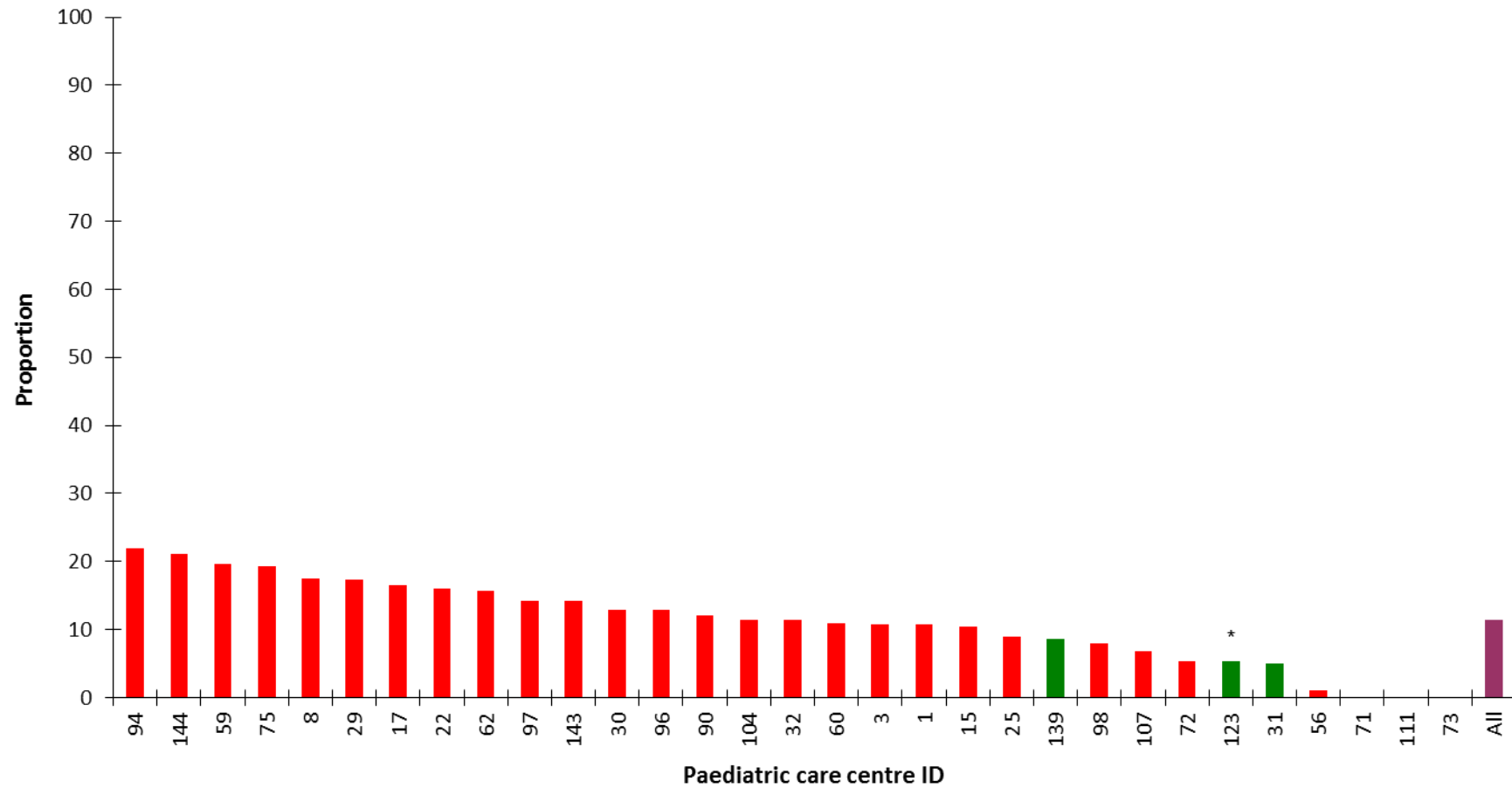
Figure 3.1 Median age (years) by adult services
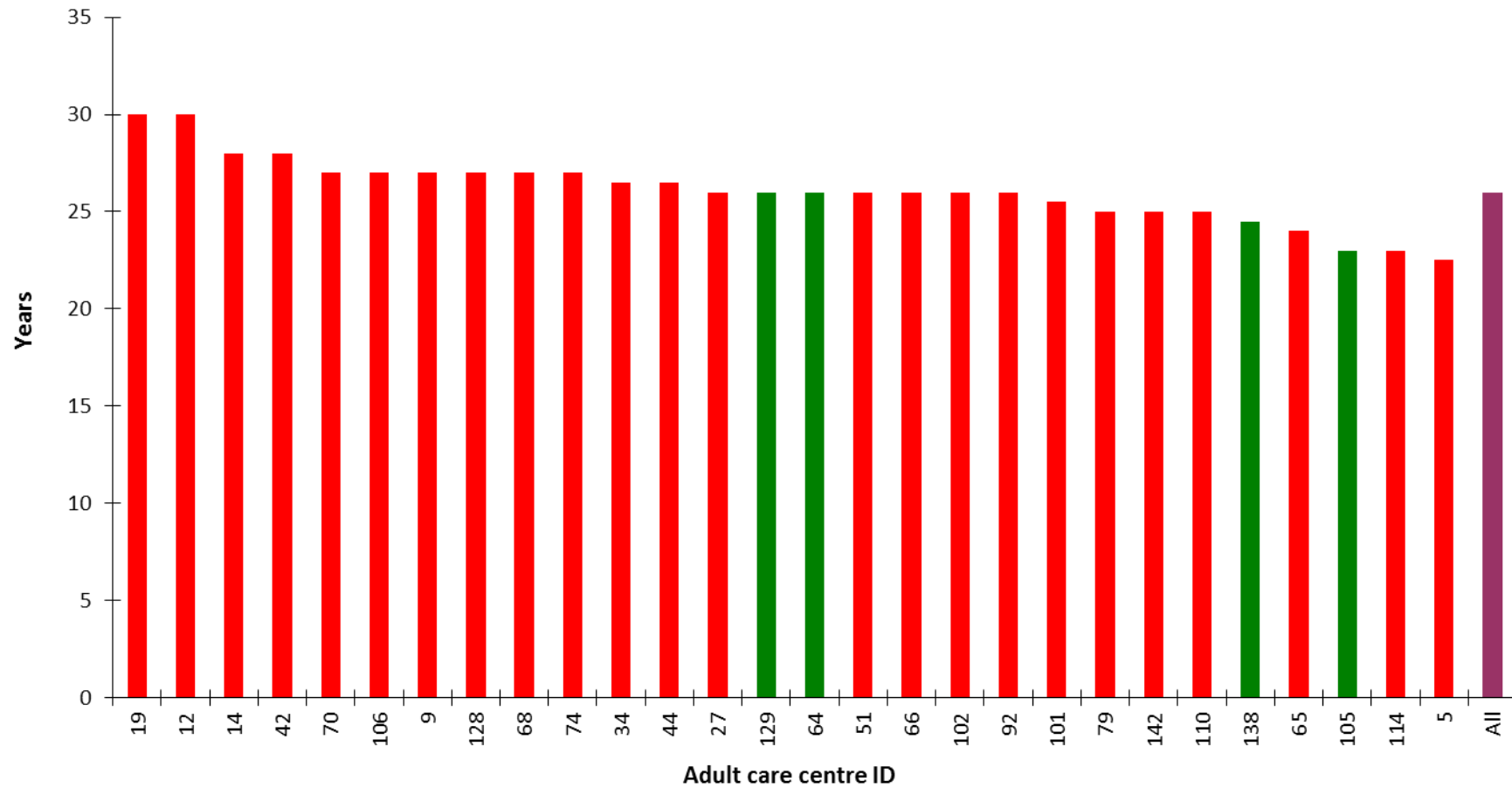
The median age in adult services is 26 years (IQR: 21-34).
Red: centres. Green: other clinics. Plum: all.

Figure 3.2 Median FEV$_1$ (% predicted) by adult services
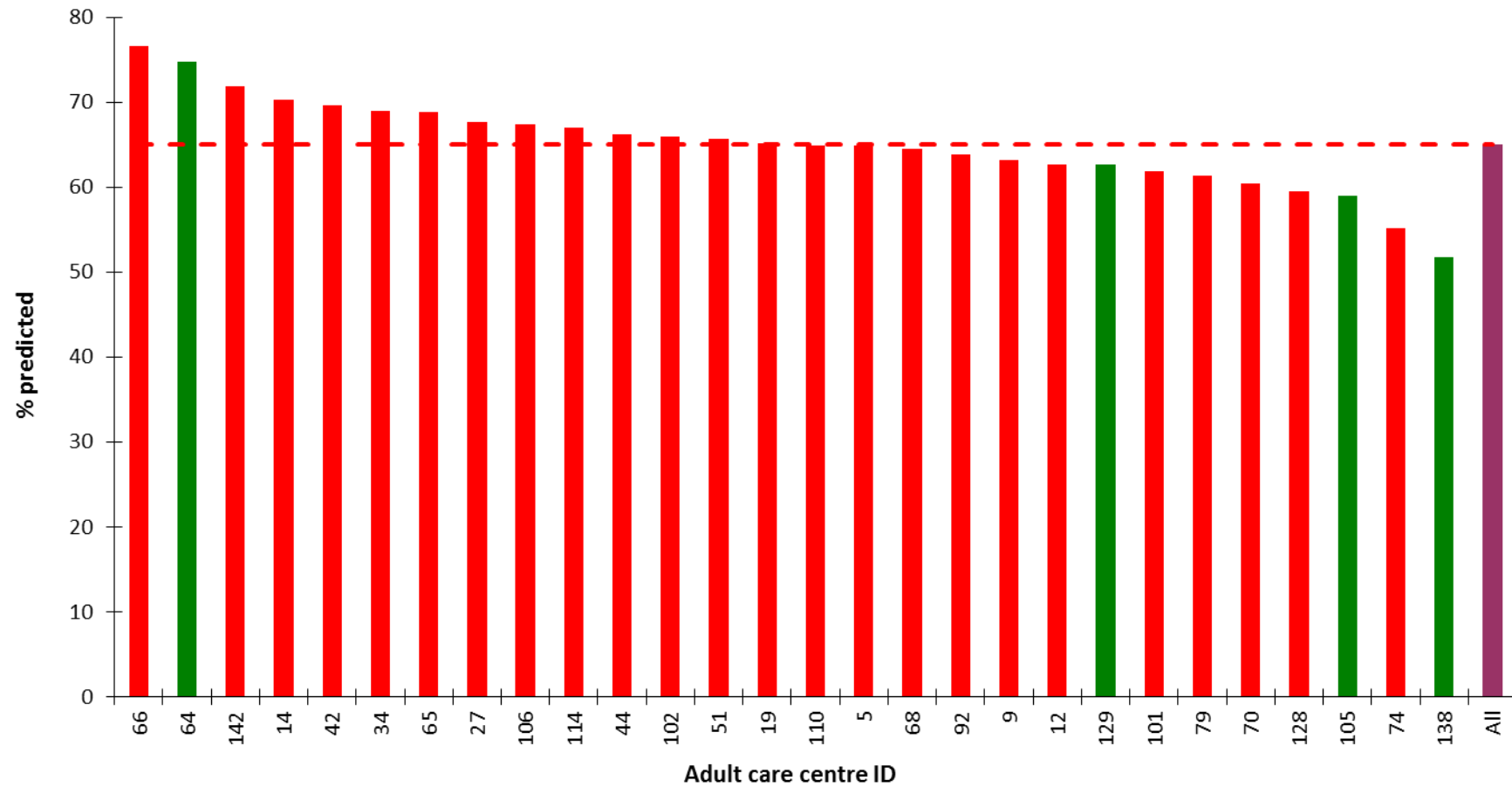
The median FEV$_1$ (% predicted) in adult services is 65.9% (IQR: 46.4 – 84.6).
Red: centres. Green: other clinics. Plum: all. Red line: median across all adult services.

Figure 3.3 Median BMI by adult services
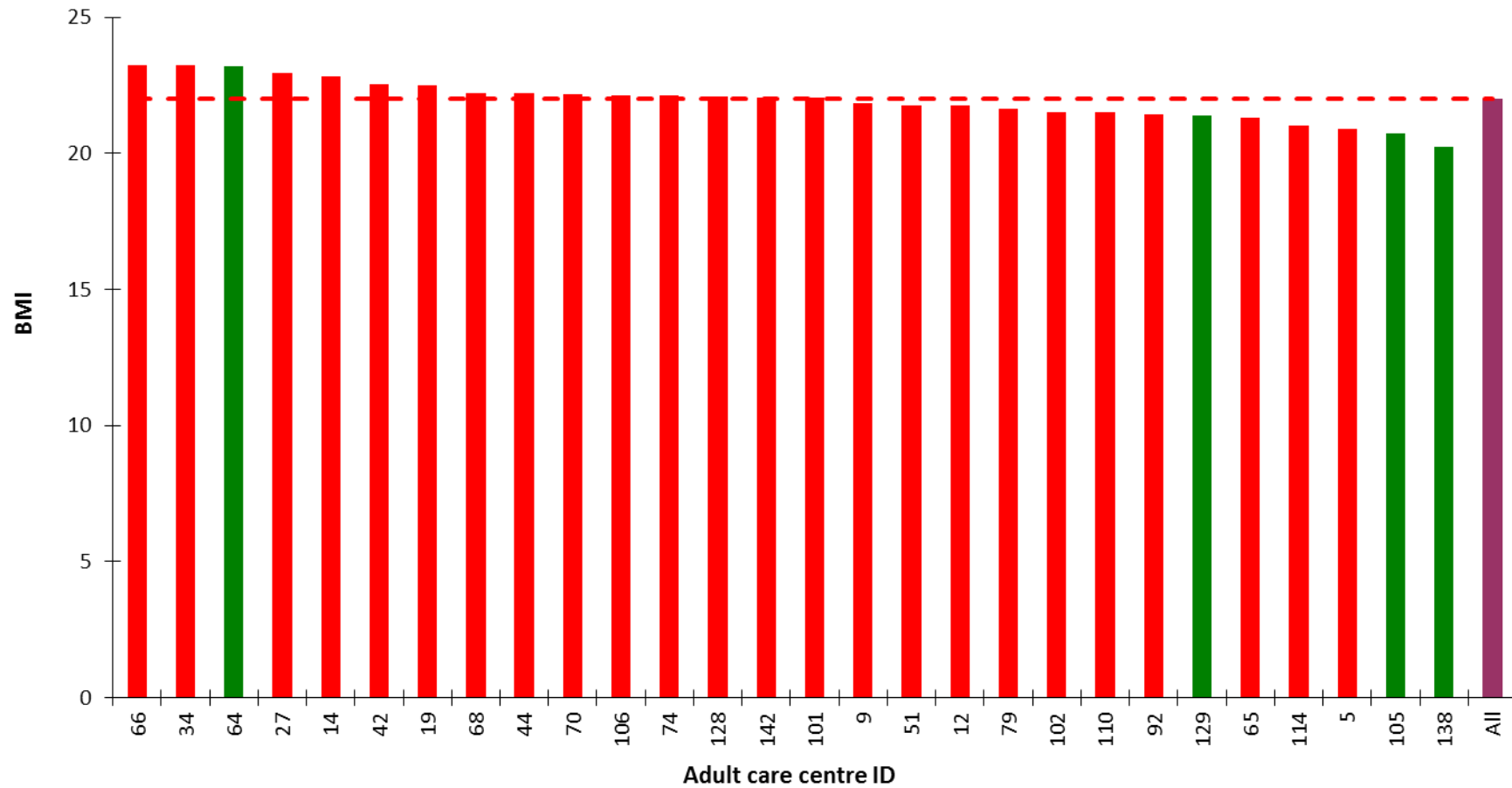
The median BMI in adult services is 22.0 (IQR: 20.0 – 24.4).

Red: centres. Green: other clinics. Plum: all. Red line: median across all adult services.

Figure 3.4 Proportion of patients with chronic *P.aeruginosa* by adult services
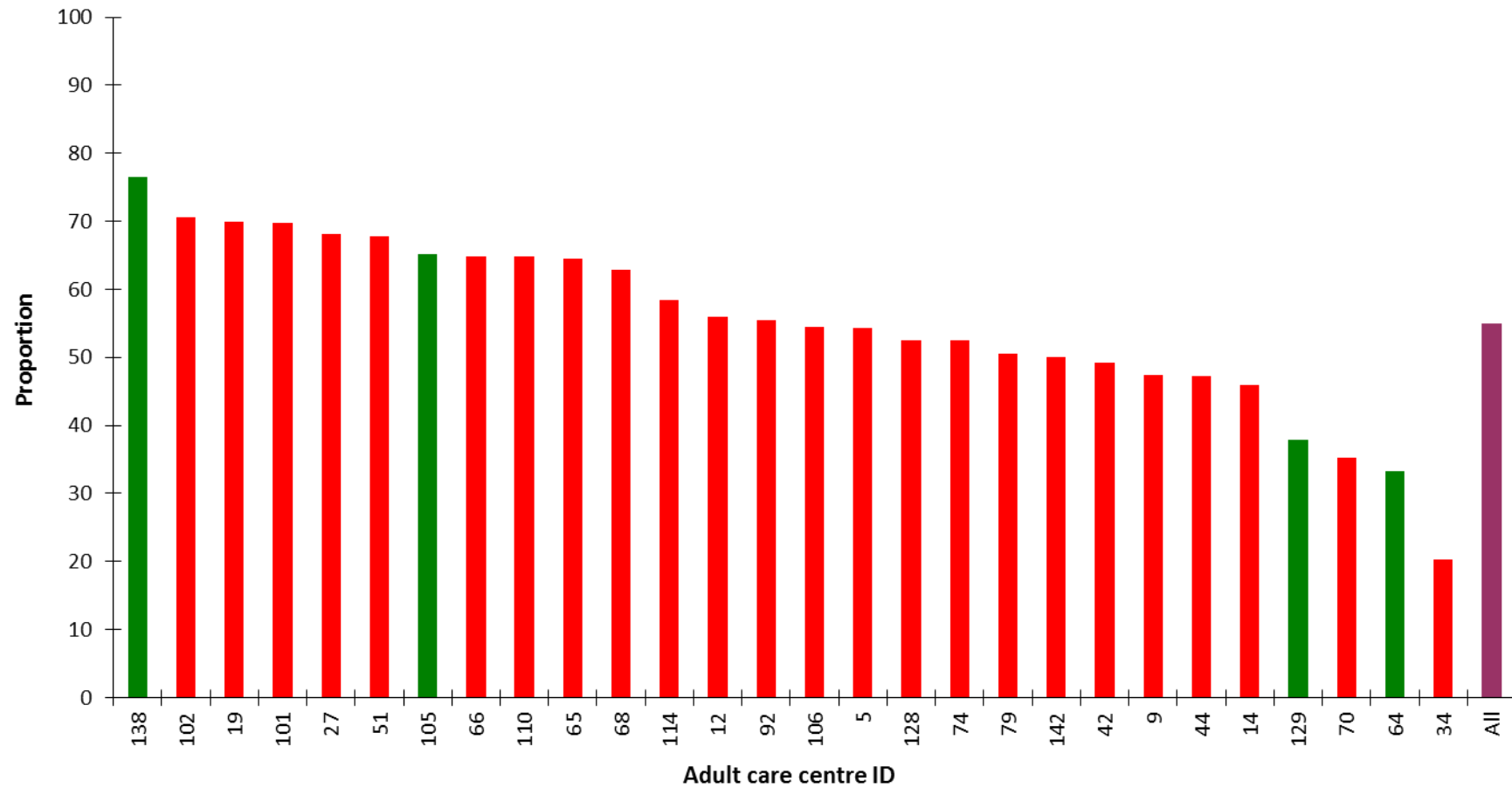
The proportion of patients with chronic *P.aeruginosa* for adult services is 57.5%.
Red: centres. Green: other clinics. Plum: all.

Section 4: Care centres/clinics providing data in 2011

4.1 Paediatric centres/clinics providing data in 2011 – 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 2011 and, for those patients alive in 2011 who did not have an annual review encounter, where they had their last encounter in between 2010-2011. 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 2011
England	Leicester	Leicester Royal Infirmary	1	58	57
England	Sheffield	Sheffield Children's Hospital	3	135	131
England	Stoke	University Hospital of North Staffordshire	8	94	94
England	London - South West	Royal Brompton Hospital	15	309	309
England	London	King's College Hospital	17	198	198
England	Oxford	John Radcliffe Hospital	22	169	167
England	Leeds	St James's University Hospital	25	236	230
England	Southampton	Southampton General hospital	29	181	179
England	London - East	Royal London Hospital	30	121	121
Scotland	Inverness	Raigmore Hospital	31	20	20

England	Bristol	Bristol Royal Hospital for Children	32	152	150
Scotland	Glasgow	Royal Hospital for Sick Children	56	136	106
England	Newcastle	Royal Victoria Infirmary	59	166	164
Northern Ireland	Belfast	Royal Belfast Hospital for Sick Children	60	188	181
England	Nottingham	Nottingham City Hospital	62	173	168
England	Middlesborough	James Cook University Hospital	71	55	54
Wales	Cardiff	Children's Hospital for Wales	72	180	172
Scotland	Dundee	Ninewells Hospital	73	24	21
Scotland	Aberdeen	Royal Aberdeen Children's Hospital	75	33	31
England	London – Central	Great Ormond Street Hospital for Sick Children	90	166	161
England	Portsmouth	Queen Alexandra Hospital	91	23	22
England	Truro	Royal Cornwall Hospital	94	42	41
England	Exeter	Royal Devon & Exeter Hospital	96	64	64
England	Liverpool	Alder Hey Children's Hospital	97	291	291
England	Norwich	Norfolk & Norwich University Hospital	98	64	63

England	Birmingham	Birmingham Children's Hospital	104	293	291
England	Cambridge	Addenbrookes Hospital	107	120	120
England	Hull	Hull Royal Infirmary	111	35	35
Scotland	Ayr/Kilmarnock	Crosshouse Hospital	123	20	19
England	Plymouth	Derriford Hospital	139	35	35
Scotland	Edinburgh	Royal Hospital for Sick Children	143	106	96
England	Manchester	Royal Manchester Children's Hospital (previously: University of Central Manchester Hospital)	144	320	312

4.2 Adult centres/clinics providing data in 2011 – ordered by clinic ID

Country	Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2011
England	London - South East	King's College Hospital	5	148	142
England	Newcastle	Royal Victoria Infirmary	9	217	217
England	London - South West	Royal Brompton Hospital	12	611	599
Northern Ireland	Belfast	Belfast City Hospital	14	210	197
England	Frimley	Frimley Park Hospital	19	107	106
England	Birmingham	Birmingham Heartlands Hospital	27	311	304
England	Exeter	Royal Devon & Exeter Hospital	34	81	78
England	Leeds	St James's University Hospital	42	395	387
Scotland	Edinburgh	Western General Hospital	44	196	166
England	Cambridge	Papworth Hospital	51	250	245
England	Plymouth	Derriford Hospital	64	40	39
England	Sheffield	Northern General Hospital	65	143	139
England	Liverpool	Liverpool Heart and Chest Hospital	66	252	245
Wales	Llandough	Llandough Hospital	68	172	166

Scotland	Aberdeen	Aberdeen Royal Infirmary	70	53	53
England	Stoke-on-Trent	University Hospital of North Staffordshire	74	61	61
Scotland	Glasgow	Gartnavel General Hospital	79	217	203
England	London - East	London Chest Hospital	92	132	124
England	Nottingham	Nottingham City hospital	101	126	122
England	Manchester	Wythenshawe Hospital	102	364	344
England	London - South East	University Hospital Lewisham	105	44	44
England	Bristol	Bristol Royal Infirmary	106	152	141
England	Southampton	Southampton General hospital	110	215	208
England	Norwich	Norfolk & Norwich University Hospital	114	59	56
England	Oxford	Churchill Hospital	128	82	79
England	Truro	Royal Cornwall Hospital	129	31	31
England	Hull	Castle Hill Hospital	138	35	34
England	Leicester	Glenfield Hospital	142	72	68

4.3 Paediatric centres/clinics providing data in 2011 – alphabetical order

England

Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2011
Birmingham	Birmingham Children's Hospital	104	293	291
Bristol	Bristol Royal Hospital for Children	32	152	150
Cambridge	Addenbrookes Hospital	107	120	120
Exeter	Royal Devon & Exeter Hospital	96	64	64
Hull	Hull Royal Infirmary	111	35	35
Leeds	St James's University Hospital	25	236	230
Leicester	Leicester Royal Infirmary	1	58	57
Liverpool	Alder Hey Children's Hospital	97	291	291
London - Central	Great Ormond Street Hospital for Sick Children	90	166	161
London - East	Royal London Hospital	30	121	121
London - South East	King's College Hospital	17	198	198
London - South West	Royal Brompton Hospital	15	309	309
Manchester	Royal Manchester Children's Hospital (previously: University of Central Manchester Hospital)	144	320	312

Newcastle	Royal Victoria Infirmary	59	166	164
Newcastle	James Cook University Hospital	71	55	54
Norwich	Norfolk & Norwich University Hospital	98	64	63
Nottingham	Nottingham City Hospital	62	173	168
Oxford	John Radcliffe Hospital	22	169	167
Plymouth	Derriford Hospital	139	35	35
Sheffield	Sheffield Children's Hospital	3	135	131
Southampton	Southampton General hospital	29	181	179
Stoke	University Hospital of North Staffordshire	8	94	94
Truro	Royal Cornwall Hospital	94	42	41

Northern Ireland

Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2011
Belfast	Royal Belfast Hospital for Sick Children	60	188	181

Scotland

Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2011
Aberdeen	Royal Aberdeen Children's Hospital	75	33	31

Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2011
Ayr/Kilmarnock	Crosshouse Hospital	123	20	19
Dundee	Ninewells Hospital	73	24	21
Edinburgh	Royal Hospital for Sick Children	143	106	96
Glasgow	Royal Hospital for Sick Children	56	136	106
Inverness	Raigmore Hospital	31	20	20

Wales

Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2011
Cardiff	Children's Hospital for Wales	72	180	172

4.4 Adult centres/clinics providing data in 2011 – alphabetical order

England

Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2011
Birmingham	Birmingham Heartlands Hospital	27	311	304
Bristol	Bristol Royal Infirmary	106	152	141
Cambridge	Papworth Hospital	51	250	245
Exeter	Royal Devon & Exeter Hospital	34	81	78
Frimley	Frimley Park Hospital	19	107	106
Hull	Castle Hill Hospital	138	35	34
Leeds	St James's University Hospital	42	395	387
Leicester	Glenfield Hospital	142	72	68
Liverpool	Liverpool Heart and Chest Hospital	66	252	245
London - East	London Chest Hospital	92	132	124
London - South East	King's College Hospital	5	148	142
London - South East	University Hospital Lewisham	105	44	44
London - South West	Royal Brompton Hospital	12	611	599

Manchester	Wythenshawe Hospital	102	364	344
Newcastle	Royal Victoria Infirmary	9	217	217
Norwich	Norfolk & Norwich University Hospital	114	59	56
Nottingham	Nottingham City Hospital	101	126	122
Oxford	Churchill Hospital	128	82	79
Plymouth	Derriford Hospital	64	40	39
Sheffield	Northern General Hospital	65	143	139
Southampton	Southampton General hospital	110	215	208
Stoke-on-Trent	University Hospital of North Staffordshire	74	61	61
Truro	Royal Cornwall Hospital	129	31	31

Northern Ireland

Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2011
Belfast	Belfast City Hospital	14	210	197

Scotland

Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2011
Aberdeen	Aberdeen Royal Infirmary	70	53	53
Edinburgh	Western General Hospital, Edinburgh	44	196	166
Glasgow	Gartnavel General Hospital, Glasgow	79	217	203

Wales

Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2011
Llandough	Llandough Hospital	68	172	166

Section 5: UK CF Registry Steering Committee

UK CF Registry Steering Committee

Dr Diana Bilton (Chair) - Director Adult CF Centre and Honorary Senior Lecturer, Royal Brompton Hospital NHS Trust, London

Dr Iolo Doull – Director Wales Paediatric CF Centre, Cardiff

Dr Keith Brownlee – Director Paediatric CF Centre, Leeds

Dr Siobhan Carr - Director Paediatric CF Centre, Royal London Hospital

Dr Steve Cunningham - CF Paediatrician, Edinburgh

Dr Caroline Elston – Director Adult CF Centre, Kings College Hospital

Dr Martin Wildman – Director Adult CF Centre, Northern General Hospital, Sheffield

Prof Stuart Elborn – Director Adult CF Centre, Belfast and Chair of RAC

Dr Stephanie MacNeill – Medical Biostatistician, Imperial College, London

Mrs Marian Dmochowska - Parent Representative

Mr Dominic Kavanagh - Patient Representative

Dr Sarah Walters - Patient Representative

Dr Kim Cox - Lead Specialist CF Commissioner, Yorks and Humber

Ms Katherine Collins - Director NSD, Scotland

Ms Sandra Tribe - Specialist CF Commissioner, London

Dr Kevin Smith - Public Health Doctor, Specialist Commissioning

Dr Geoffrey Carroll - Specialist CF Commissioner, Wales

Ms Joanne Osmond - Director Clinical Care, Cystic Fibrosis Trust

Ms Elaine Gunn - Registry Manager, Cystic Fibrosis Trust

Mr Ed Owen - CEO, Cystic Fibrosis Trust

Dr Janet Allen – Director of Research, Cystic Fibrosis Trust