



UK CF Registry Annual Data Report 2008

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UK CF Registry
Annual Data Report 2008**

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PREFACE



The *Annual Data Report 2008* marks another significant step forward for the UK CF Registry.

The number of patients registered has increased to 8513; and with more clinics starting to use Port CF in 2009, we anticipate that this number will rise over the next couple of years.

The number of patients with 'complete data' has risen to 6082 in 2008 – over 70% of patients registered. Our thanks are due to the care teams at CF Centres and clinics for their continuing hard work collecting and entering the data. The challenge for 2009 and the future is to increase the number and proportion of patients with 'complete data'.

With more patient data from more CF centres and clinics, we have been able to expand the range of analyses provided in the report: section 1 contains more detail about complications and therapies for all patients in the UK; sections 2 and 3 contain more comparisons of outcomes between centres and clinics. For paediatric care, comparisons are provided both by care networks and by individual centre/clinic. Thus every CF service can now begin to use the *Annual Data Report* to benchmark and improve patient care.

The highlight from the data is that median predicted survival for 2008 is 38.8 years (compared to 35.2 years in 2007). The year on year increase is likely to have arisen from more complete data being available rather than from a change in practice; it is however encouraging that this figure remains comparable to prior year and to other CF registries.

Care must also be taken when interpreting the charts in sections 2 and 3 comparing outcomes between centres and clinics, particularly when comparing a clinic with fewer than 20 patients to a large CF Centre. However, if used carefully now – and with more sophisticated analysis of lung function and other trends over time – these charts will be invaluable in improving care, both at individual CF centres/clinics and throughout the UK.

As ever, we would like to acknowledge our gratitude to all who have contributed to the production of this report: the patients and families who agree to contribute their data; the care teams at CF centres and clinics for collecting and entering the data; the biostatisticians at the National Heart and Lung Institute, Imperial College for providing the analyses and charts; and the Registry team at the Cystic Fibrosis Trust.

We hope that you find the *Annual Data Report 2008* interesting and helpful. Please share it widely among colleagues – and anyone who can use it to help improve care for people with Cystic Fibrosis throughout the UK.

A handwritten signature in black ink, appearing to read 'Diana Bilton'.

Dr Diana Bilton
Chair
CF Registry Steering Committee

A handwritten signature in black ink, appearing to read 'Alan Larsen'.

Alan Larsen
Director of Research and Finance
Cystic Fibrosis Trust

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Summary of the UK Cystic Fibrosis Registry

	<u>2002</u>	<u>2003</u>	<u>2004</u>	<u>2007</u>	<u>2008</u>
CF patients registered	6932	6861	7046	8080 ¹	8513 ¹
CF patients with “complete” data	5301	4875	5561	4408 ²	6082 ²
Age in years; median	17.4	16.1	16.4	18 ³	18 ³
Newly diagnosed patients	159	142	164	239 ⁴	235 ⁴
Age at diagnosis in months; median	5	5	5	5 ^{3,5}	4 ^{3,5}
Adults aged 16 yrs and over; %	50.1	50.8	51.4	56.7 ³	56.2 ³
Males; %	53.9	53.8	53.4	53.9 ³	53.3 ³
Genotyped; %	95	95	95	92.6 ³	93.7 ³
Median predicted survival in years (95% Confidence interval)				35.2 ⁶ (31.0, 42.6)	38.8 ⁶ (34.2, 47.3)
Total deaths reported	94	103	123	106	100
Age at death in years; median	23	24.2	25.6	24	27

Starting in 2007, data were entered on the newly established Port CF system. Definitions may not be consistent with previous years.

Notes:

¹ From 2007, this is calculated as the number of patients on the database who satisfied the following criteria:

- were born and diagnosed with CF on or before 31 December; and
- had no recorded date of death before 1 January of the reporting year

² In 2007, “complete data” is defined as having a clinical encounter when “well” that year. In 2008 this question was changed in Port CF from “well, unwell, unknown” to “stable or unstable”. “Complete data” in 2008 is defined as having a clinical encounter when “stable” that year.

³ Calculated for patients with “complete” data.

⁴ Calculated for all patients registered. Among the 6082 patients with complete data in 2008, 137 were diagnosed in 2008.

⁵ Three patients diagnosed prenatally had age at diagnosis set to 0 months.

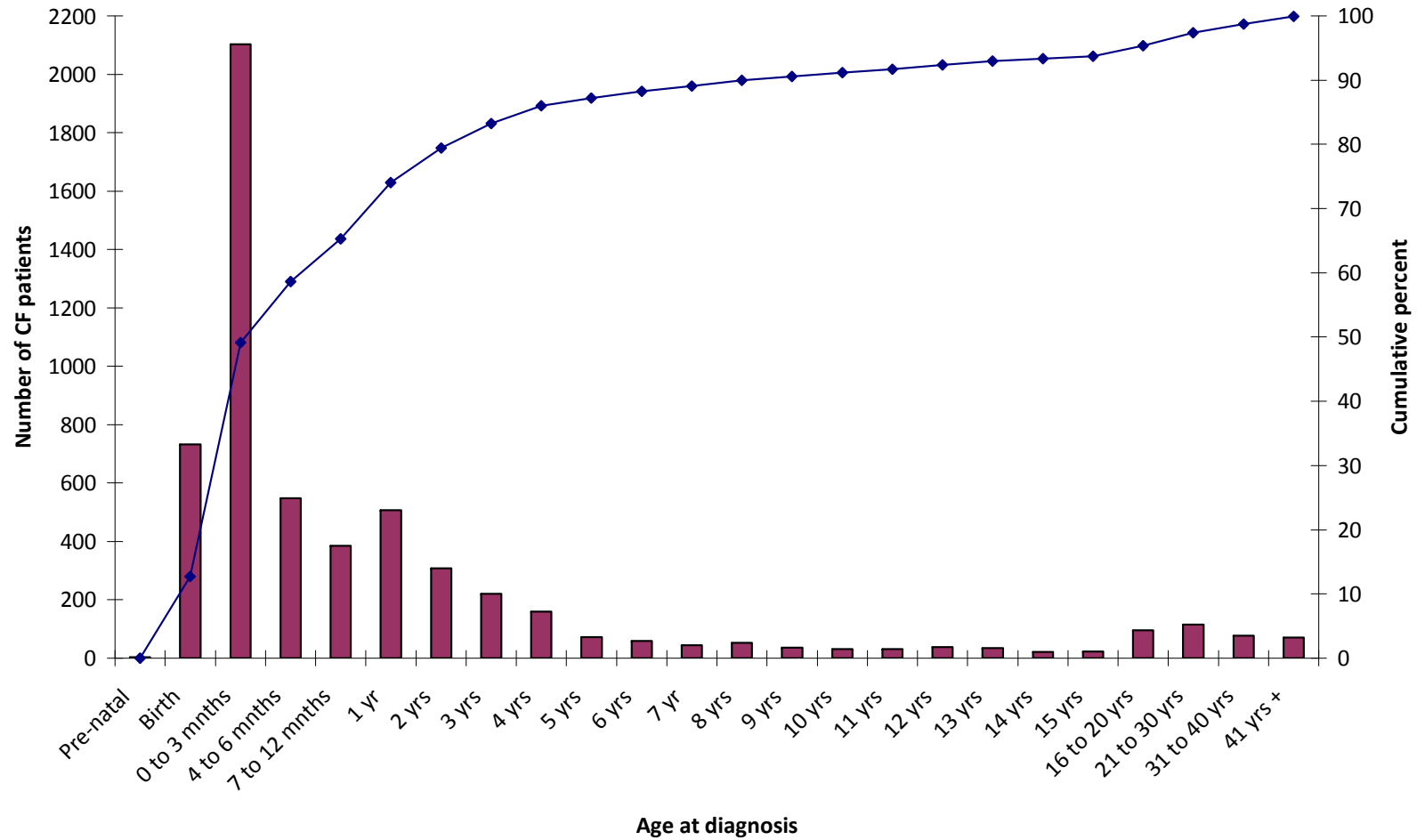
⁶ This represents the age beyond which half of the current UK CF Registry patients would be expected to live, given the ages of CF patients in the Registry and the mortality distribution of deaths in the same year.

Section 1:

All UK Patients

**(based on 6082 patients with complete
annual review data in 2008)**

1.1 Age at diagnosis



Date of diagnosis is available for 5771 of the 6082 patients with annual review data. Three patients were diagnosed using prenatal screening.

1.2 Diagnosis and screening statistics

Age at diagnosis; n (%)

<i>Pre-natal</i>	3 (0.1)
<i>Birth</i>	733 (12.7)
<i>0-3 months</i>	2103 (36.4)
<i>4-6 months</i>	548 (9.5)
<i>7-12 months</i>	385 (6.7)
<i>1 yr</i>	506 (8.8)
<i>2 yrs</i>	308 (5.3)
<i>3 yrs</i>	221 (3.8)
<i>4 yrs</i>	159 (2.8)
<i>5 yrs</i>	72 (1.3)
<i>6 yrs</i>	60 (1.0)
<i>7 yrs</i>	45 (0.8)
<i>8 yrs</i>	53 (0.9)
<i>9 yrs</i>	36 (0.6)
<i>10 yrs</i>	32 (0.6)
<i>11 yrs</i>	31 (0.5)
<i>12 yrs</i>	38 (0.7)
<i>13 yrs</i>	35 (0.6)
<i>14 yrs</i>	22 (0.4)
<i>15 yrs</i>	23 (0.4)
<i>16 – 20 yrs</i>	95 (1.7)
<i>21 – 30 yrs</i>	115 (2.0)
<i>31 – 40 yrs</i>	77 (1.3)
<i>41 yrs +</i>	71 (1.2)

The median (range) age at diagnosis is 4 months (0-79 years).

880 patients were diagnosed by neonatal screening.

Of the 79 patients born in 2008, 56 were identified by neo-natal screening.

1.3 Genotyping

5701 patients have been genotyped with a recorded value

DF508 Mutations; n (%)

Homozygous DF508	3095 (54.3%)
Heterozygous DF508	2149 (37.7%)
No DF508 or both unidentified	457 (8.0%)

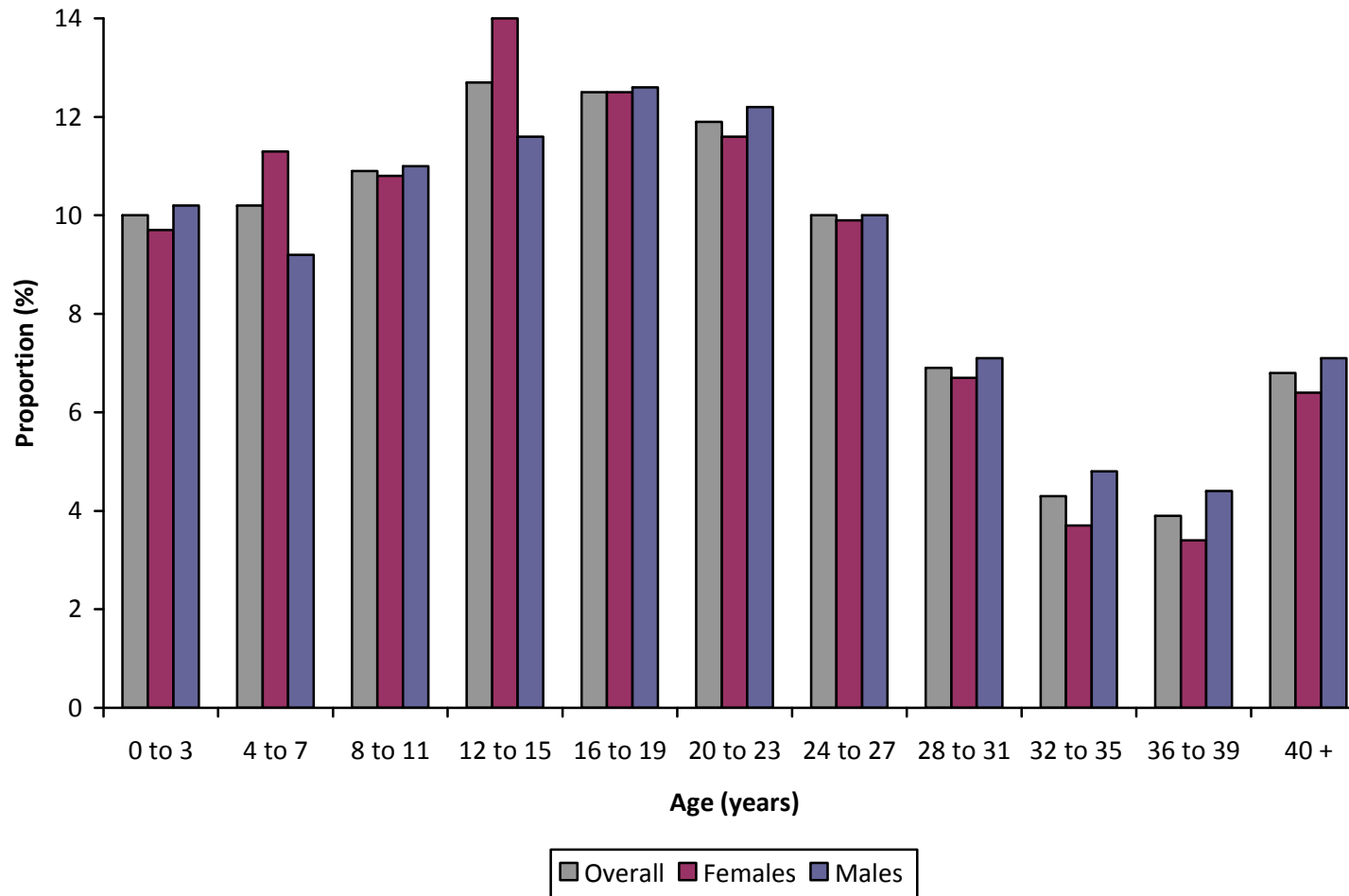
All mutations; n(%)

DF508	5244	(92.0%)
G551D	318	(5.6%)
G542X	190	(3.3%)
R117H	184	(3.2%)
621+1G->T	156	(2.7%)
1898+1G->A	77	(1.4%)
1717-1G->A	67	(1.2%)
N1303K	69	(1.2%)
DI507	62	(1.1%)
R560T	49	(0.9%)
3659delC	52	(0.9%)
R553X	45	(0.8%)
G85E	37	(0.7%)
Q493X	37	(0.7%)
E60X	39	(0.7%)
3849+10kbC->T	33	(0.6%)
W1282X	27	(0.5%)
2184delA	21	(0.4%)
1078delT	22	(0.4%)
V520F	16	(0.3%)
2789+5G->A	18	(0.3%)
D1152H	18	(0.3%)
R347P	18	(0.3%)
3120+1G->A	10	(0.2%)
A455E	10	(0.2%)
711+1G->T	11	(0.2%)
S549N	12	(0.2%)
R1162X	13	(0.2%)
1161delC	3	(0.1%)
2183delAA->G	3	(0.1%)
I148T	3	(0.1%)
K710X	3	(0.1%)
L206W	3	(0.1%)
S549R	3	(0.1%)
A559T	4	(0.1%)

R352Q	4	(0.1%)
R334W	5	(0.1%)
R1158X	8	(0.1%)
R347H	8	(0.1%)
2143delT	2	(0.04%)
3662delA	2	(0.04%)
G178R	2	(0.04%)
P574H	2	(0.04%)
R117C	2	(0.04%)
R1283M	2	(0.04%)
1609delCA	1	(0.02%)
1677delTA	1	(0.02%)
1898+5G->T	1	(0.02%)
2869insG	1	(0.02%)
3120G->A	1	(0.02%)
3550-T->G	1	(0.02%)
3849+4A->G	1	(0.02%)
574delA	1	(0.02%)
C524X	1	(0.02%)
D579Y	1	(0.02%)
E585X	1	(0.02%)
Q552X	1	(0.02%)
R1066C	1	(0.02%)
S1251N	1	(0.02%)
V201M	1	(0.02%)
W1089X	1	(0.02%)
Y1092X	1	(0.02%)
Y563D	1	(0.02%)
<i>Other</i>	<i>389</i>	<i>(6.8%)</i>
<i>Not identified</i>	<i>825</i>	<i>(14.5%)</i>

Table shows the number of patients (%) with at least one copy of the mutation.

1.4 Age distribution

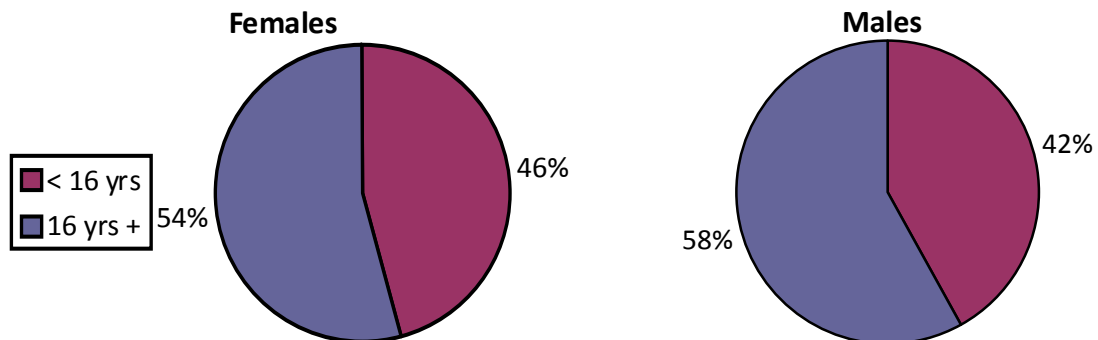


Age is calculated as the age at annual review.

1.5 Age and sex distribution

Age	Overall N=6082	Female N=2844	Male N=3239
0-3 yrs	605 (10.0)	276 (9.7)	329 (10.2)
4-7	621 (10.2)	322 (11.3)	299 (9.2)
8-11	663 (10.9)	308 (10.8)	355 (11.0)
12-15	773 (12.7)	397 (14.0)	376 (11.6)
16-19	762 (12.5)	355 (12.5)	407 (12.6)
20-23	725 (11.9)	331 (11.6)	394 (12.2)
24-27	605 (10.0)	281 (9.9)	324 (10.0)
28-31	419 (6.9)	190 (6.7)	229 (7.1)
32-35	260 (4.3)	105 (3.7)	155 (4.8)
36-39	237 (3.9)	96 (3.4)	141 (4.4)
40+	412 (6.8)	182 (6.4)	230 (7.1)
Median (range)	18 yrs (0 - 80 yrs)	17 yrs (0 – 80 yrs)	18 yrs (0 – 77yrs)

1.6 Age distribution by sex



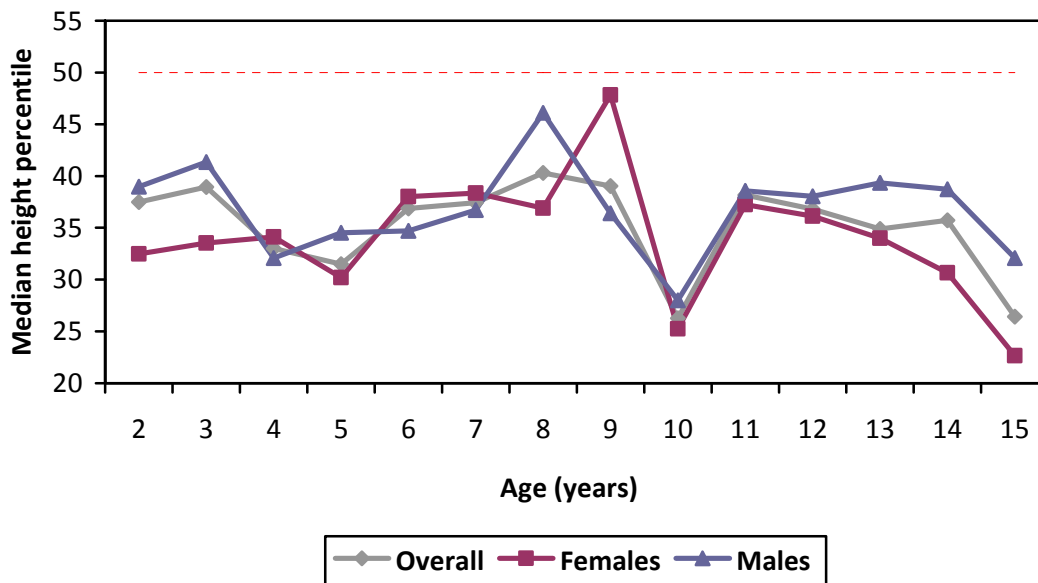
1.7 Employment status among adults aged 16yrs+

	<u>Number of patients</u>
Full-time working	1053
Part-time working	410
Student	618
Homemaker	156
Unemployed	568
“Disabled”	142
Retired	40
Unknown	169
<i>No data</i>	494

Note that these groups are not mutually exclusive.

Of the 2926 adults aged 16yrs and older for whom an employment status questionnaire was completed (excluding “unknown”), 2046 (69.9%) reported being in work or study.

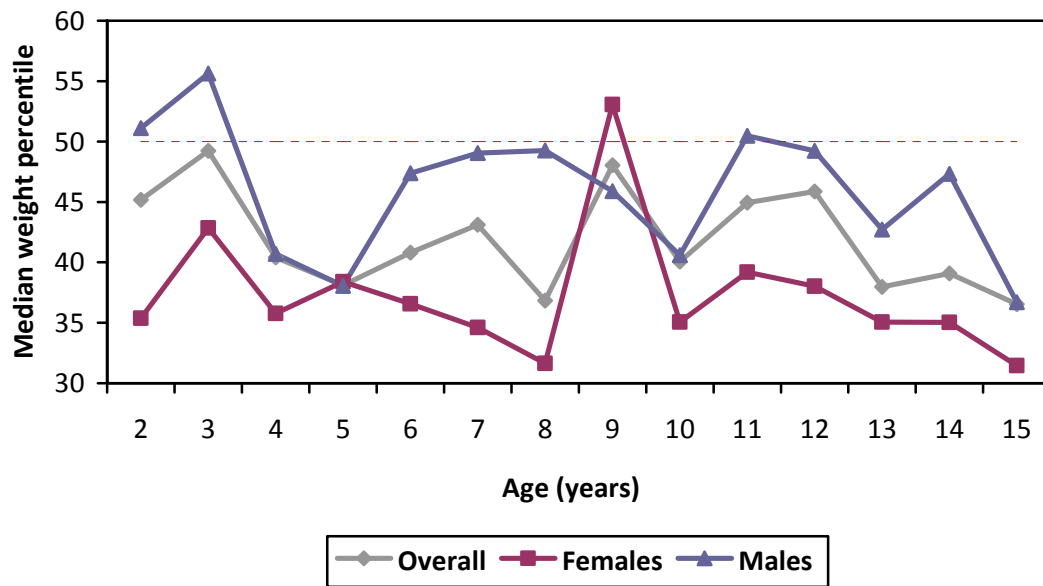
1.8 Median height percentiles among children (n=2256)



Age	Overall		Female		Male	
	N	Median (range)	N	Median (range)	N	Median (range)
2	149	37.5 (0.6, 99.6)	73	32.5 (0.8, 99.6)	76	39.0 (0.6, 99.2)
3	128	38.9 (0.03, 99.5)	57	33.5 (0.03, 97.9)	71	41.4 (1.1, 99.5)
4	167	33.0 (0.3, 99.1)	86	34.1 (0.3, 98.2)	81	32.1 (0.4, 99.1)
5	135	31.5 (0.04, 97.6)	71	30.2 (0.04, 86.8)	64	34.5 (0.9, 97.6)
6	146	36.9 (0.1, 99.7)	80	38.0 (0.1, 98.4)	66	34.7 (0.06, 99.7)
7	156	37.4 (0.01, 95.6)	74	38.3 (0.01, 95.6)	82	36.7 (0.1, 95.3)
8	142	40.3 (0.2, 98.7)	74	36.9 (0.2, 98.7)	68	46.1 (0.5, 98.7)
9	166	39.0 (0.3, 100.0)	76	47.8 (0.4, 99.2)	90	36.4 (0.3, 100.0)
10	160	26.3 (0.1, 92.9)	66	25.3 (0.2, 92.9)	94	28.0 (0.1, 92.5)
11	171	38.2 (0.3, 98.6)	79	37.3 (0.3, 98.6)	92	38.6 (1.0, 98.1)
12	202	36.8 (0.1, 98.9)	107	36.1 (0.1, 98.9)	95	38.1 (0.7, 97.9)
13	184	34.9 (0.3, 97.3)	104	34.0 (0.3, 97.3)	80	39.3 (0.4, 91.1)
14	173	35.7 (0, 99.4)	79	30.7 (0.3, 97.7)	94	38.7 (0, 99.4)
15	177	26.4 (0, 99.5)	84	22.7 (0, 99.5)	93	32.1 (0, 98.0)
Overall	2256	34.8 (0, 100.0)	1110	32.7 (0, 99.6)	1146	36.8 (0, 100.0)

N refers to the number of patients in each age/sex category

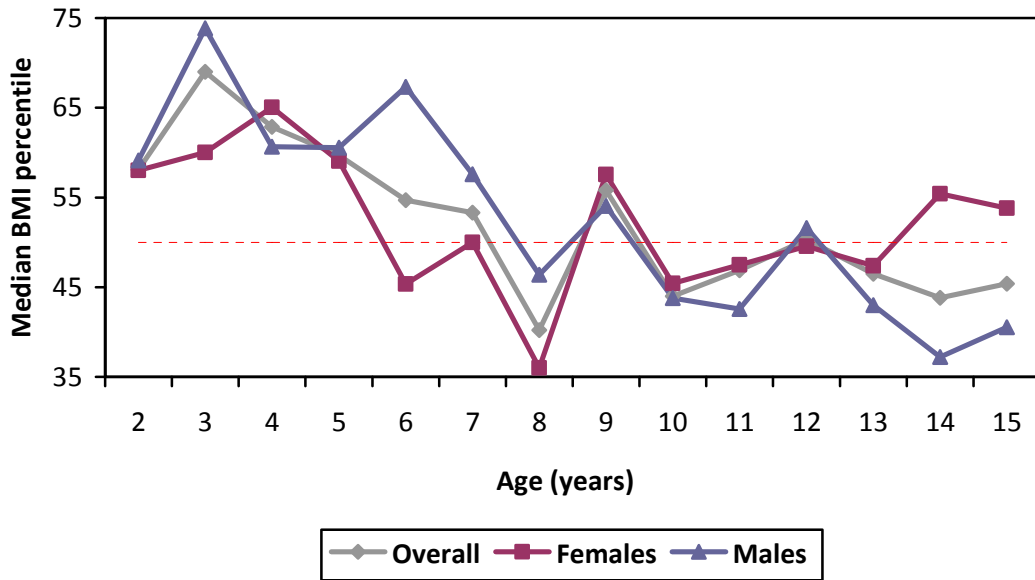
1.9 Median weight percentiles among children (n=2317)



Age	Overall		Female		Male	
	N	Median (range)	N	Median (range)	N	Median (range)
2	157	45.2 (0.2, 100.0)	76	35.4 (0.2, 99.2)	81	51.1 (0.6, 100.0)
3	132	49.2 (0.1, 99.7)	58	42.8 (0.1, 96.3)	74	55.6 (0.6, 99.7)
4	169	40.4 (0.9, 99.6)	88	35.8 (1.3, 98.2)	81	40.7 (0.9, 99.6)
5	138	38.1 (0.6, 97.2)	73	38.4 (1.0, 89.5)	65	38.1 (0.6, 97.2)
6	150	40.8 (0.02, 99.6)	82	36.6 (0.4, 98.7)	68	47.4 (0.02, 99.6)
7	158	43.1 (0, 99.8)	75	34.6 (0, 97.9)	83	49.0 (0, 99.8)
8	148	36.8 (0.1, 100.0)	76	31.6 (0.1, 100.0)	72	49.3 (0.6, 95.3)
9	169	48.0 (1.2, 99.9)	80	53.1 (1.2, 99.2)	89	45.9 (1.5, 99.9)
10	160	40.1 (0.1, 97.6)	66	35.1 (0.1, 94.6)	94	40.6 (0.9, 97.6)
11	174	44.9 (1.3, 99.6)	81	39.2 (1.3, 99.6)	93	50.5 (1.6, 99.6)
12	205	45.9 (0.2, 99.5)	109	38.0 (0.3, 98.5)	96	49.2 (0.2, 99.5)
13	188	38.0 (0.2, 99.3)	106	35.1 (0.2, 99.3)	82	42.7 (1.4, 94.5)
14	181	39.1 (0, 99.9)	84	35.0 (0, 99.9)	97	47.3 (0, 98.2)
15	188	36.5 (0, 99.4)	92	31.5 (0, 99.0)	96	36.7 (0, 99.4)
Overall	2317	41.6 (0, 100.0)	1146	37.1 (0, 100.0)	1171	45.7 (0, 100.0)

N refers to the number of patients in each age/sex category

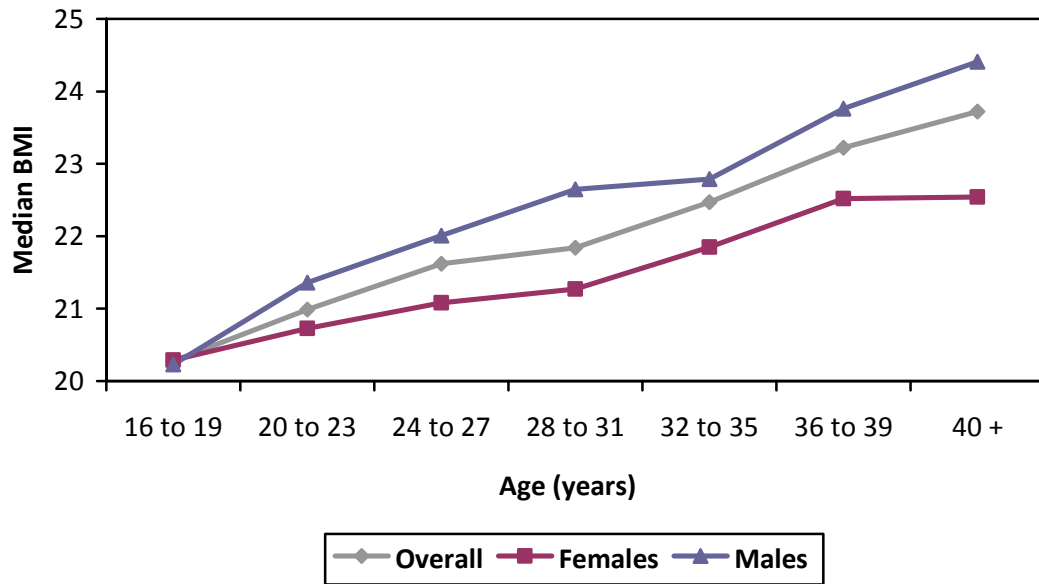
1.10 Median BMI percentiles among children (n=2301)



Age	Overall		Female		Male	
	N	Median (range)	N	Median (range)	N	Median (range)
2	150	58.1 (0.01, 99.1)	73	58.0 (0.01, 99.1)	77	59.1 (0.7, 98.8)
3	130	69.0 (0.1, 99.8)	58	60.0 (1.5, 99.8)	72	73.8 (0.1, 99.7)
4	169	62.9 (0.8, 99.5)	88	65.0 (0.8, 96.5)	81	60.6 (1.1, 99.5)
5	137	59.7 (0.4, 96.2)	72	59.0 (0.5, 96.2)	65	60.6 (0.4, 94.4)
6	148	54.7 (0.04, 98.4)	82	45.3 (0.04, 98.0)	66	67.3 (0.1, 98.4)
7	158	53.3 (0, 98.8)	75	50.0 (0, 96.5)	83	57.6 (1.5, 98.8)
8	148	40.2 (0.3, 99.1)	76	36.0 (0.3, 99.1)	72	46.4 (0.9, 93.8)
9	168	55.8 (0.6, 98.5)	79	57.6 (1.4, 96.9)	89	54.1 (0.6, 98.5)
10	160	44.0 (0.2, 95.5)	66	45.4 (0.2, 95.5)	94	43.8 (0.2, 95.3)
11	173	46.9 (1.8, 98.8)	80	47.5 (1.8, 98.8)	93	42.6 (2.6, 98.1)
12	204	50.3 (0.8, 97.1)	108	49.5 (0.8, 97.1)	96	51.6 (0.9, 97.1)
13	188	46.5 (0.4, 97.5)	106	47.4 (0.4, 97.5)	82	43.0 (0.8, 91.5)
14	180	43.8 (0, 98.0)	84	55.4 (0, 98.0)	96	37.2 (0.1, 95.5)
15	188	45.4 (0, 97.6)	92	53.8 (0, 95.3)	96	40.5 (0.2, 97.9)
Overall	2301	51.7 (0, 99.8)	1139	51.6 (0, 99.8)	1162	51.7 (0.1, 99.7)

N refers to the number of patients in each age/sex category

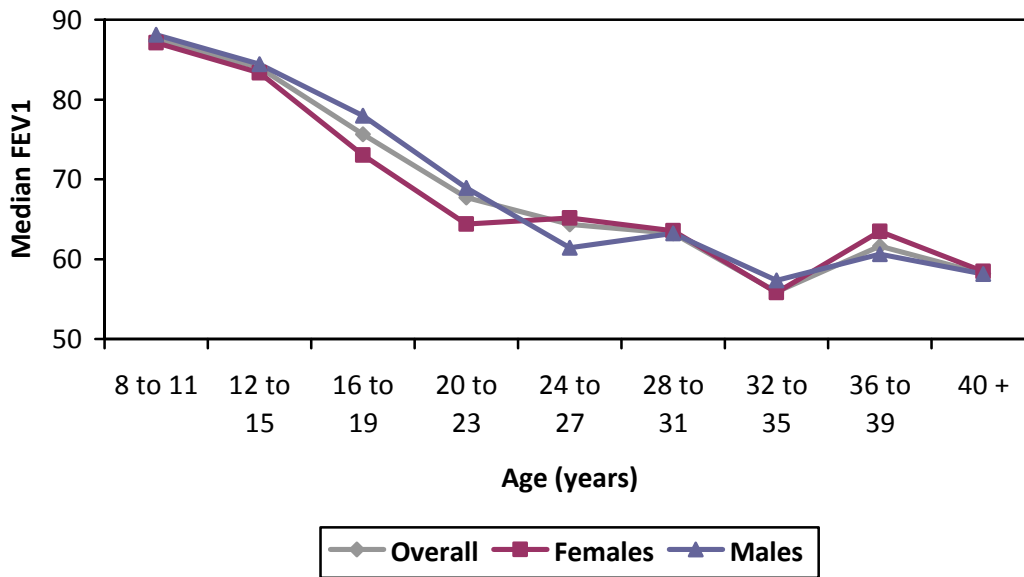
1.11 Median BMI values among adults (n=3284)



Age	Overall		Female		Male	
	N	Median (range)	N	Median (range)	N	Median (range)
16-19	739	20.3 (13.9, 41.5)	346	20.3 (13.8, 36.4)	393	20.2 (14.7, 41.5)
20-23	695	21.0 (14.7, 37.1)	317	20.7 (14.7, 36.0)	378	21.4 (15.0, 37.1)
24-27	581	21.6 (13.5, 40.0)	268	21.1 (15.3, 40.0)	313	22.0 (13.5, 36.4)
28-31	400	21.8 (15.2, 34.7)	179	21.3 (15.2, 34.7)	221	22.6 (16.1, 31.5)
32-35	248	22.5 (16.1, 48.5)	101	21.9 (17.1, 39.9)	147	22.8 (16.1, 48.5)
36-39	229	23.2 (15.6, 42.6)	92	22.5 (15.6, 39.6)	137	23.8 (16.0, 42.6)
40+	392	23.7 (15.5, 41.5)	171	22.5 (15.5, 41.5)	221	24.4 (17.0, 38.0)
Overall	3284	21.6 (13.5, 48.5)	1474	21.1 (13.8, 41.5)	1810	22.0 (13.5, 48.5)

N refers to the number of patients in each age/sex category

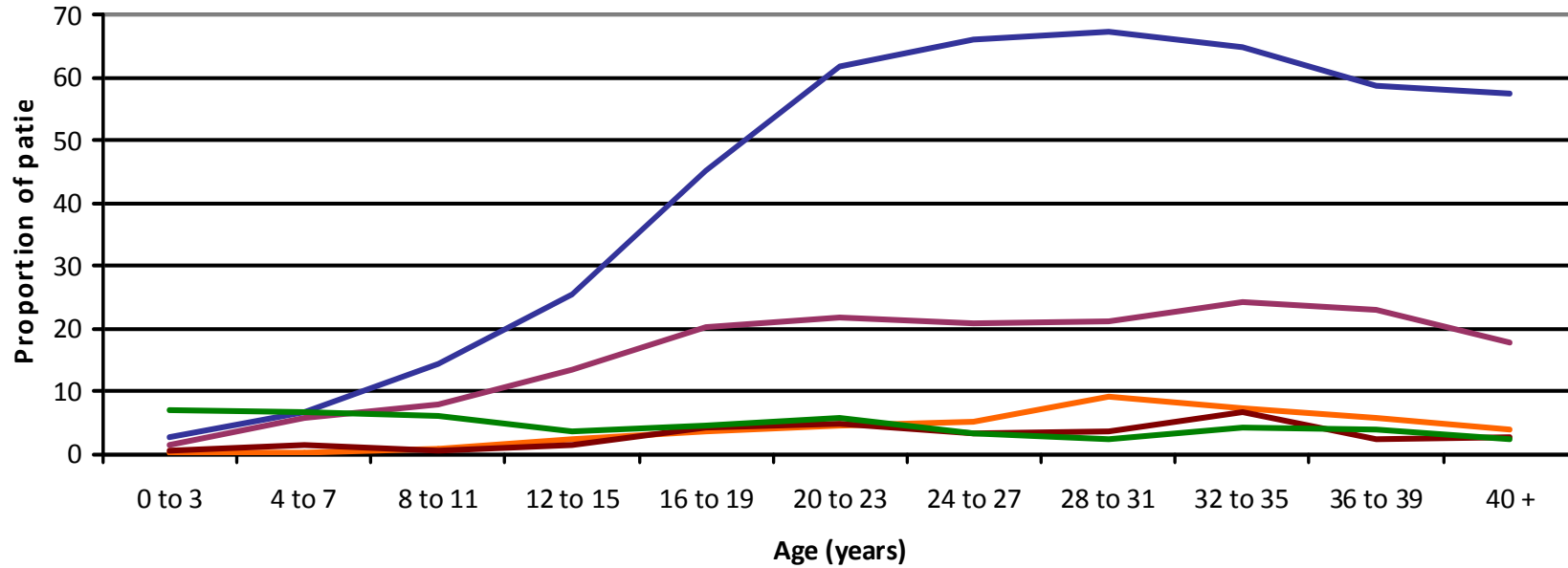
1.12 Median FEV₁ (% predicted) among patients aged 8 years and older (n=4554)



Age	Overall		Female		Male	
	N	Median (range)	N	Median (range)	N	Median (range)
8-11	605	87.5 (33.9, 131.7)	288	87.1 (33.9, 131.7)	317	88.1 (35.2, 128.7)
12-15	719	84.0 (17.9, 127.0)	368	83.4 (17.9, 125.3)	351	84.4 (28.2, 127.0)
16-19	721	75.6 (14.7, 129.4)	337	73.1 (15.5, 126.3)	384	78.0 (14.7, 129.4)
20-23	682	67.7 (13.9, 131.1)	310	64.4 (18.9, 131.1)	372	68.9 (13.9, 130.8)
24-27	570	64.3 (14.8, 133.2)	265	65.2 (15.4, 133.2)	305	61.4 (14.8, 125.3)
28-31	394	63.2 (12.8, 142.8)	180	63.5 (15.1, 142.8)	214	63.2 (12.8, 127.6)
32-35	245	55.9 (15.3, 124.7)	101	55.8 (22.6, 120.7)	144	57.3 (15.3, 124.7)
36-39	223	61.6 (14.8, 140.7)	91	63.5 (14.8, 140.7)	132	60.6 (18.6, 116.7)
40+	395	58.2 (13.9, 147.3)	174	58.5 (17.5, 143.7)	221	58.1 (13.9, 147.3)
Overall	4554	73.2 (12.8, 147.3)	2114	73.0 (14.8, 143.7)	2440	73.2 (12.8, 147.3)

N refers to the number of patients in each age/sex category

1.13 Infections



N in age band	605	621	663	773	762	725	605	419	260	237	412
N cultures taken	539	549	571	681	694	650	555	374	234	206	368

— Chronic *S. aureus*; n=799 (15.2%)
 — Chronic *P. aeruginosa*; n=2098 (39.4%)
 — *B. cepacia*; n=184 (3.4%)
— MRSA; n=147 (2.7%)
 — *H. influenzae*; n=256 (4.7%)

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

	Age (yrs)											Overall
	0-3	4-7	8-11	12-15	16-19	20-23	24-27	28-31	32-35	36-39	40+	
<i>N</i> patients in age band	605	621	663	773	762	725	605	419	260	237	412	6082
<i>N</i> cultures taken at clinic	539	549	571	681	694	650	555	374	234	206	368	5421
Chronic <i>S.aureus</i> at annual review; n(%)	7 (1.4)	31 (5.7)	43 (7.8)	89 (13.6)	135 (20.4)	138 (21.7)	111 (20.8)	79 (21.2)	55 (24.3)	47 (22.9)	64 (17.9)	799 (15.2)
Chronic <i>P.aeruginosa</i> at annual review; n(%)	13 (2.6)	37 (6.8)	81 (14.3)	168 (25.3)	305 (45.1)	398 (61.8)	355 (66.0)	255 (67.3)	151 (64.8)	124 (58.8)	211 (57.3)	2098 (39.4)
<i>B.cepacia</i> at clinic; n(%)	1 (0.2)	2 (0.4)	5 (0.9)	16 (2.4)	25 (3.6)	29 (4.5)	29 (5.2)	34 (9.1)	17 (7.3)	12 (5.8)	14 (3.8)	184 (3.4)
MRSA at clinic; n(%)	3 (0.6)	8 (1.5)	3 (0.5)	10 (1.5)	30 (4.3)	31 (4.8)	18 (3.2)	13 (3.5)	16 (6.8)	5 (2.4)	10 (2.7)	147 (2.7)
<i>H.influenzae</i> at clinic; n(%)	38 (7.1)	37 (6.7)	34 (6.0)	24 (3.5)	32 (4.6)	37 (5.7)	18 (3.2)	9 (2.4)	10 (4.3)	8 (3.9)	9 (2.5)	256 (4.7)

Age is calculated as age at annual review.

Chronic *P. aeruginosa* is defined as where there have been 3 or more isolates in the twelve months.

1.14 Complications identified in 2008

	Overall (n=6082)	<16 years (n=2662)	≥16 years (n=3420)
Nontuberculous mycobacteria or atypical mycobacteria; <i>n</i> (%)	140 (2.3)	26 (1.0)	114 (3.3)
Cirrhosis with no portal hypertension; <i>n</i> (%)	76 (1.2)	21 (0.8)	55 (1.6)
Cirrhosis with portal hypertension; <i>n</i> (%)	123 (2.0)	20 (0.8)	103 (3.0)
Gallbladder disease requiring surgery; <i>n</i> (%)	25 (0.4)	2 (0.1)	23 (0.7)
Nasal polyps requiring surgery; <i>n</i> (%)	247 (4.1)	44 (1.7)	203 (5.9)
Pneumothorax requiring chest tube; <i>n</i> (%)	40 (0.7)	0	40 (1.2)
Cancer confirmed by histology; <i>n</i> (%)	10 (0.2)	1 (0.04)	9 (0.3)
Fibrosing colonopathy/colonic stricture; <i>n</i> (%)	2 (0.03)	1 (0.04)	1 (0.03)
Port inserted or replaced; <i>n</i> (%)	337 (5.5)	97 (3.6)	240 (7.0)

1.15 Transplants

Of those with complete data in 2008, 126 patients had been evaluated and 55 accepted onto the transplant list.

24 received transplants: 16 bilateral lung
 1 heart and lung
 6 liver
 1 renal

1.16 Other therapy

	All (n=6082)	<16 years (n=2662)	≥16 years (n=3420)
NIV; n(%)	85 (1.6)	12 (0.6)	73 (2.4)
Long-term oxygen; n(%)	334 (6.3)	53 (2.4)	281 (9.2)
<i>Among those who had long-term oxygen therapy:</i>			
<i>Continuously</i>	59 (17.7)	2 (3.8)	57 (20.3)
<i>Nocturnal+exertion</i>	87 (26.1)	12 (22.6)	75 (26.7)
<i>When required</i>	43 (12.9)	2 (3.8)	41 (14.6)
<i>With exacerbation</i>	145 (43.4)	37 (69.8)	108 (38.4)

1.17 Feeding

	Overall (n=6082)	<16 years (n=2662)	≥16 years (n=3420)
Any supplemental feeding; n(%)	1769 (32.4)	618 (26.5)	1151 (40.0)
<i>Nasogastric Tube</i>	97 (1.8)	13 (0.6)	84 (2.7)
<i>Gastrostomy Tube / Button</i>	354 (6.5)	160 (6.8)	194 (6.2)
<i>Jejunal</i>	2 (0.04)	1 (0.04)	1 (0.03)
<i>Total parental nutrition</i>	2 (0.04)	1 (0.04)	1 (0.03)

1.18 Days on IV antibiotics

Age	Home		Hospital		Total	
	N (%)	Median (range)	N (%)	Median (range)	N (%)	Median (range)
0-3	24 (4.5)	10.5 (1, 70)	160 (30.3)	14 (1, 135)	165 (31.2)	14 (1, 140)
4-7	72 (12.2)	15 (1, 112)	201 (34.0)	14 (1, 112)	219 (37.1)	14 (2, 119)
8-11	137 (22.1)	28 (1, 156)	197 (31.8)	14 (1, 270)	239 (38.6)	28 (1, 270)
12-15	205 (28.5)	28 (1, 360)	309 (42.9)	14 (1, 184)	387 (53.8)	28 (1, 365)
16-19	258 (37.4)	22 (1, 280)	342 (49.6)	14 (1, 208)	427 (61.9)	28 (1, 287)
20-23	248 (38.0)	28 (3, 351)	303 (46.4)	14 (1, 229)	391 (59.9)	28 (2, 365)
24-27	245 (45.3)	25 (3-112)	259 (47.9)	20 (1, 177)	354 (65.4)	31 (6, 177)
28-31	170 (44.0)	28 (4, 230)	146 (37.8)	14 (1, 361)	228 (59.1)	28 (4, 361)
32-35	103 (42.6)	24 (1, 98)	89 (36.8)	16 (1, 134)	138 (57.0)	28 (4, 169)
36-39	84 (38.4)	28 (4, 168)	69 (31.5)	14 (3, 233)	119 (54.3)	28 (4, 233)
40+	123 (32.5)	27 (3, 350)	119 (31.5)	15 (1, 225)	170 (45.0)	28 (7, 350)
Overall	1669 (30.0)	25 (3, 360)	2194 (39.4)	14 (1, 361)	2837 (50.9)	28 (1, 365)

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

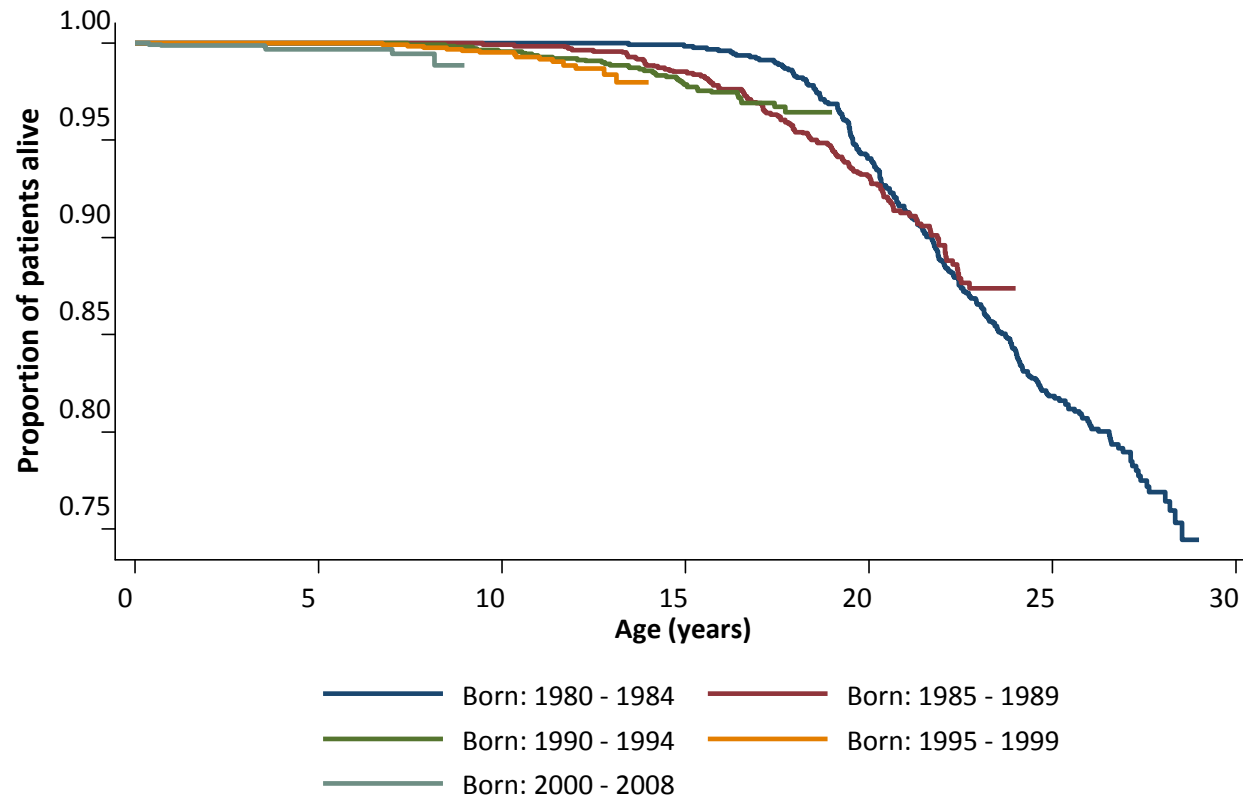
1.19 Drug treatment

Age	DNase treatment; n(%)
0-3 yrs	46 (7.6)
4-7	125 (20.1)
8-11	227 (34.2)
12-15	359 (46.4)
16-19	377 (49.5)
20-23	319 (44.0)
24-27	288 (47.6)
28-31	182 (43.4)
32-35	108 (41.5)
36-39	83 (35.0)
40+	147 (35.7)
Overall	2261 (37.2)

Antibiotic use among patients with chronic *P. aeruginosa*

	Overall	<16 years	≥16 years
Patients with chronic <i>P. aeruginosa</i>	2098	299	1799
<i>Nebulised tobramycin solution (TOBI/Bramitob)</i>	412 (19.6)	48 (16.1)	364 (20.2)
<i>Other aminoglycoside</i>	43 (2.0)	5 (0.2)	38 (2.1)
<i>Nebulised colistin (Colomycin)</i>	914 (43.6)	174 (58.2)	740 (41.1)
<i>Nebulised colistin (Promixin)</i>	490 (23.4)	73 (24.4)	417 (23.2)
<i>Chronic macrolide</i>	1267 (60.4)	110 (36.8)	1157 (64.3)

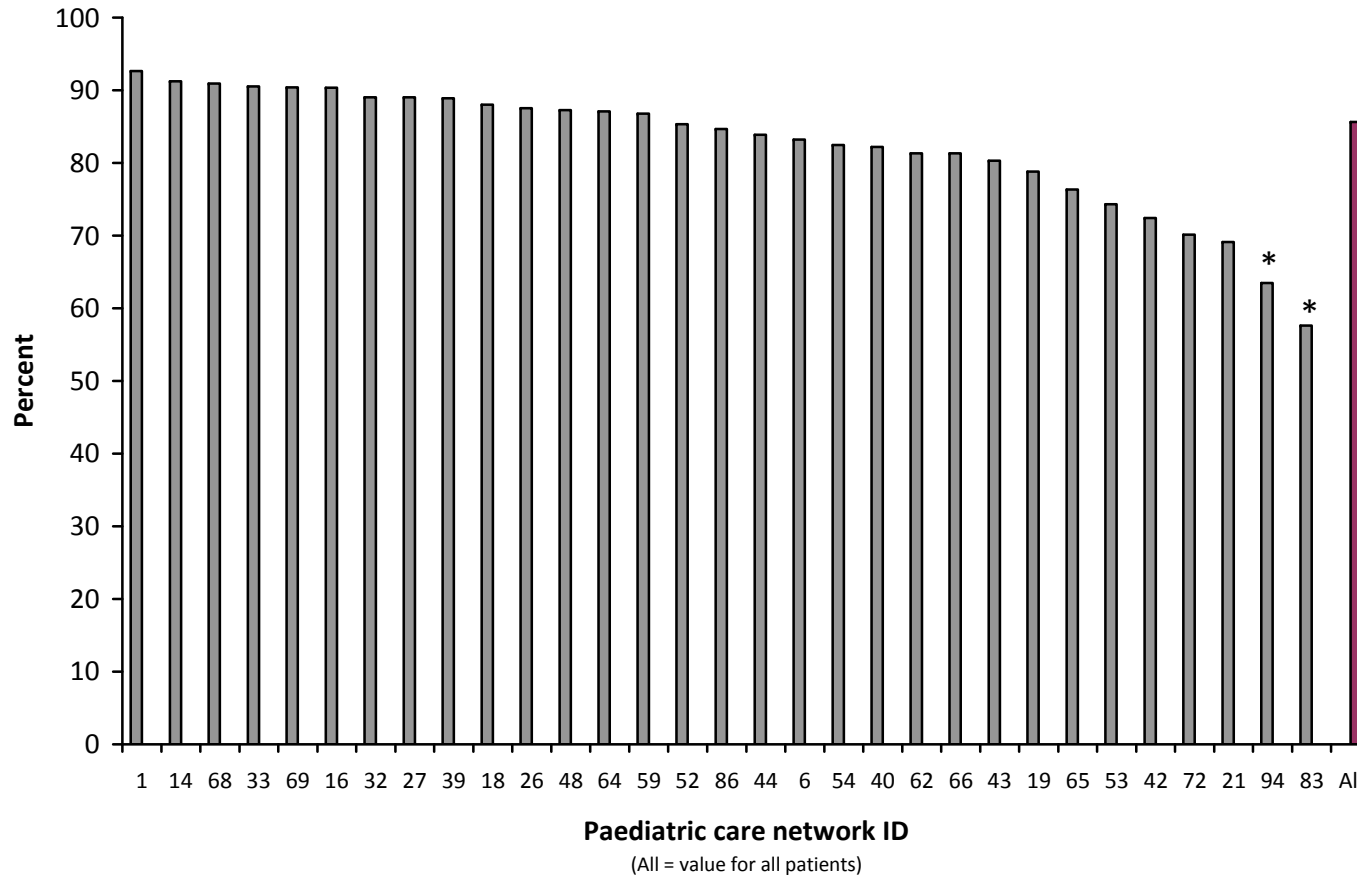
1.20 Actuarial survival by birth cohort



Section 2:

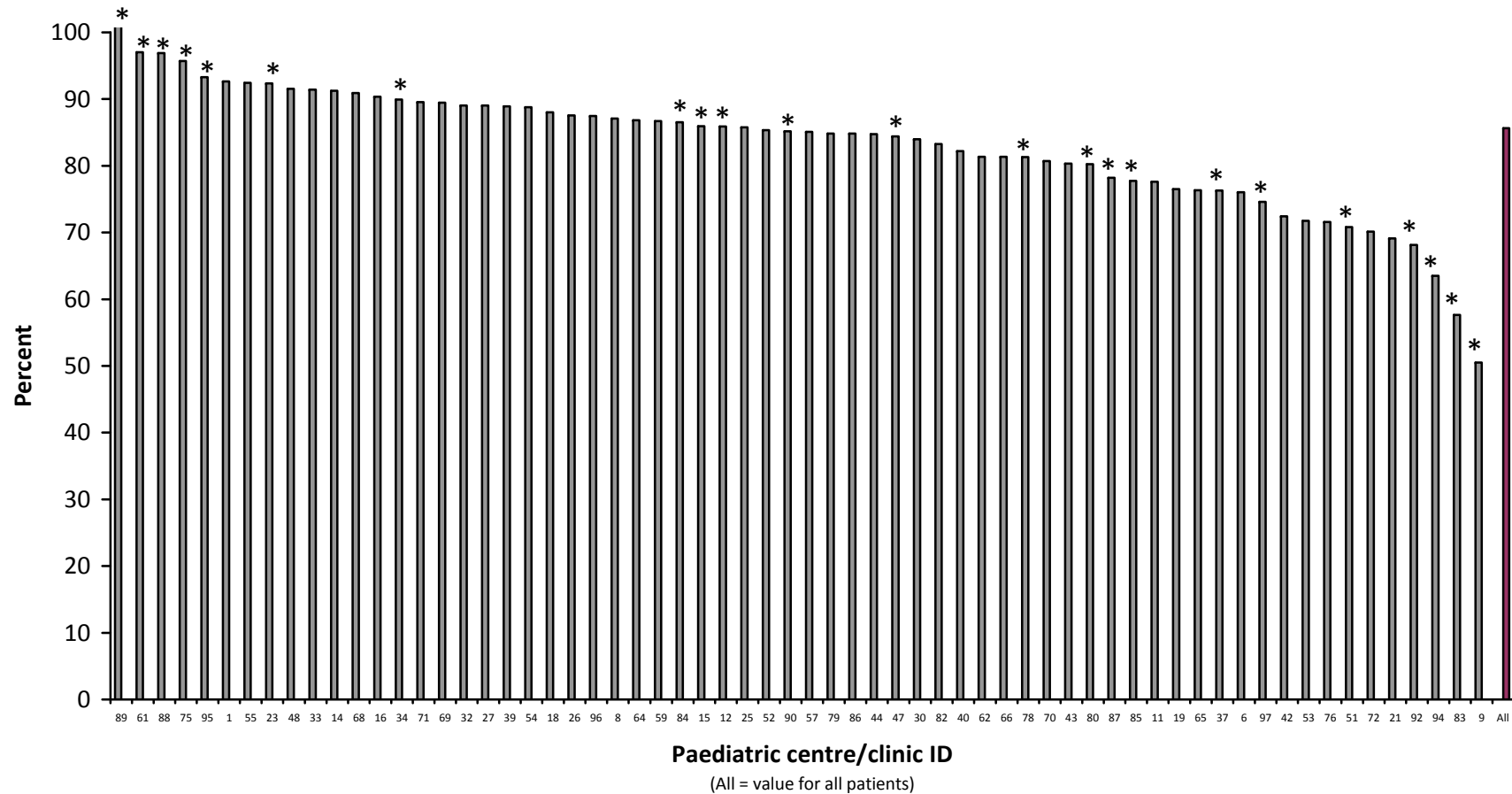
Comparison of outcomes for paediatric care networks, centres and clinics (based on 2900 patients with complete annual review data in 2008)

2.1 Median FEV₁ (% predicted) by paediatric care network and lone centre/clinic



Network clinics are reported under the ID of the lead specialist centre for the network.
 The median FEV₁ (% predicted) for paediatric care networks and lone centres/clinics is 84.7% (min=57.6%, max=92.7%).
 * Centre/clinic with fewer than 20 patients at annual review

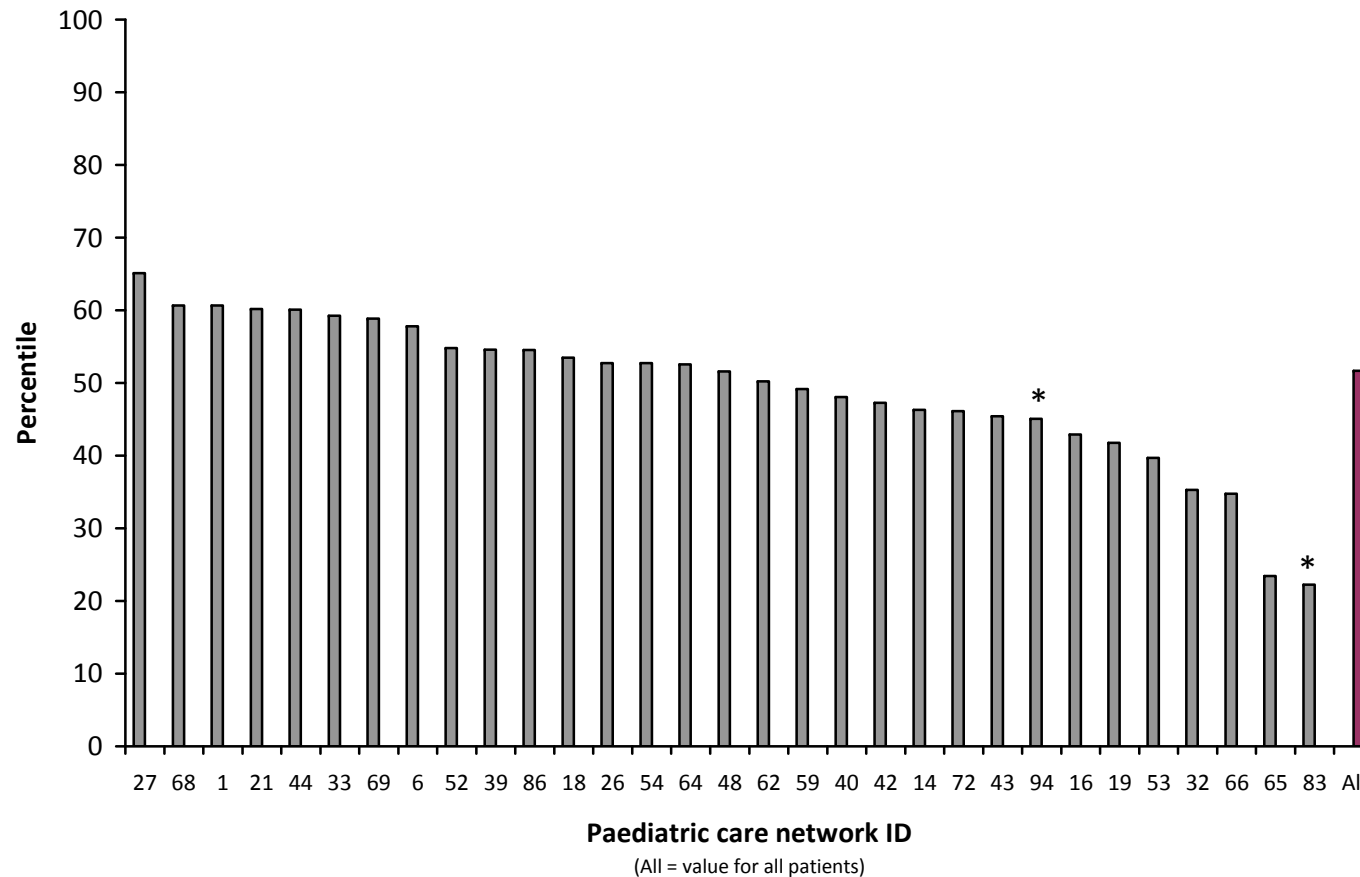
2.2 Median FEV₁ (% predicted) by paediatric centre/clinic



The median FEV₁ (% predicted) for paediatric care centres/clinics is 85.1% (min=50.5%, max=101.8%). Data were missing for centre 98*.

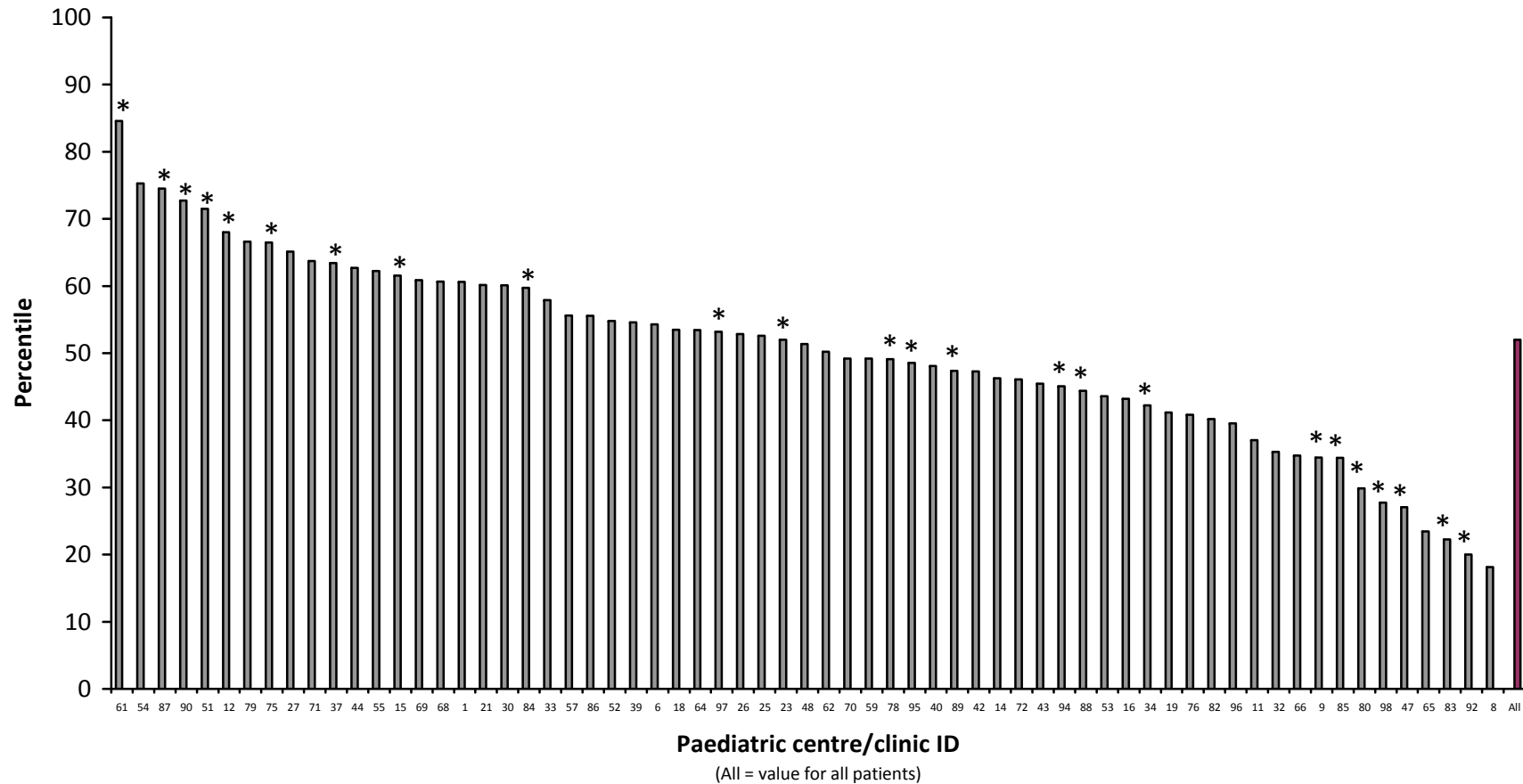
* Centre/clinic with fewer than 20 patients at annual review

2.3 Median BMI (percentile) by paediatric care network and lone centre/clinic



Network clinics are reported under the ID of the lead specialist centre for the network.
 The median BMI (percentile) for paediatric care networks and lone centres/clinics is 51.6 (min=22.3, max=65.1).
 * Centre/clinic with fewer than 20 patients at annual review

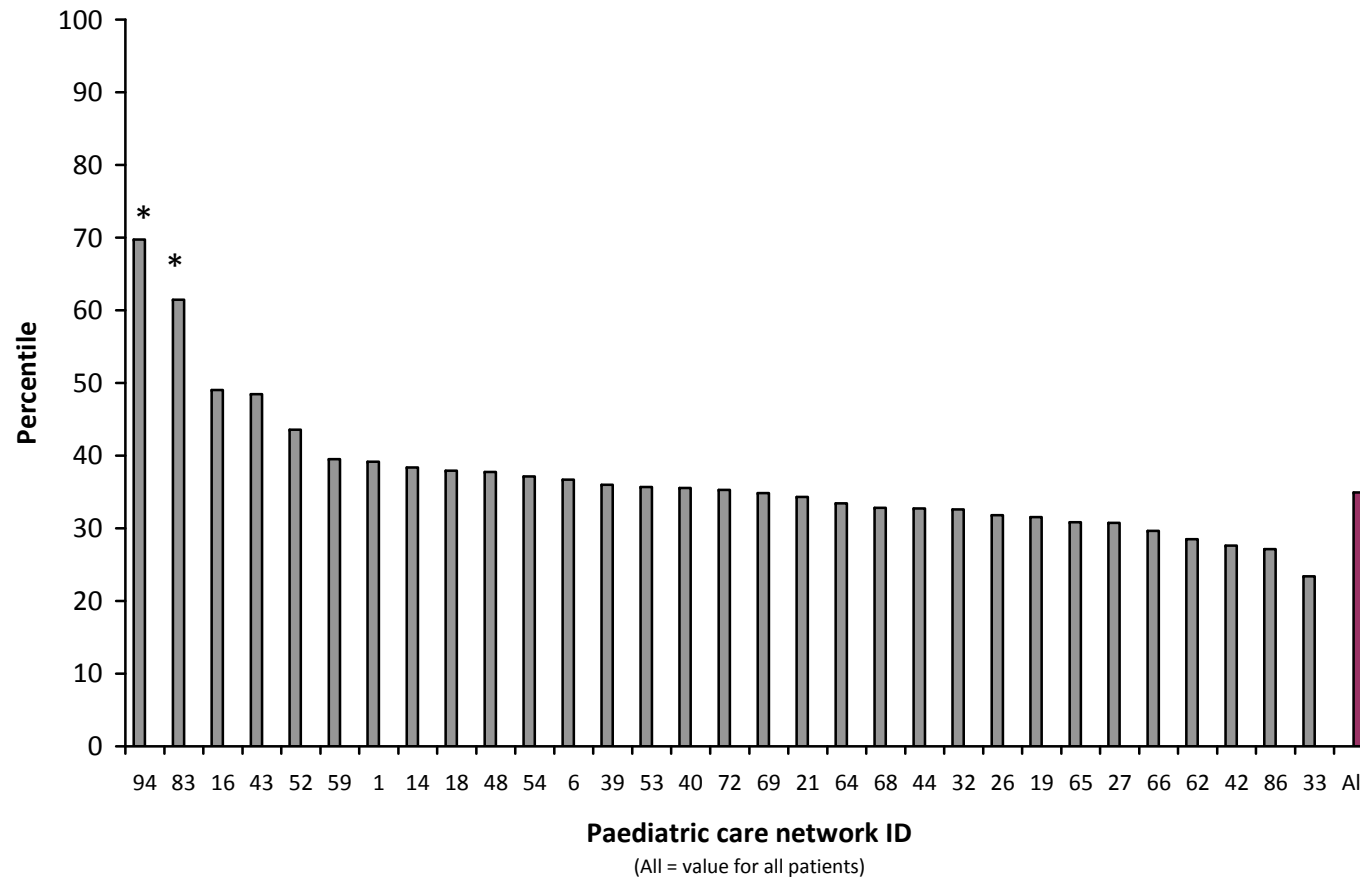
2.4 Median BMI (percentile) by paediatric centre/clinic



The median BMI (percentile) for paediatric care centres/clinics is 51.4 (min=18.1, max=84.6).

* Centre/clinic with fewer than 20 patients at annual review

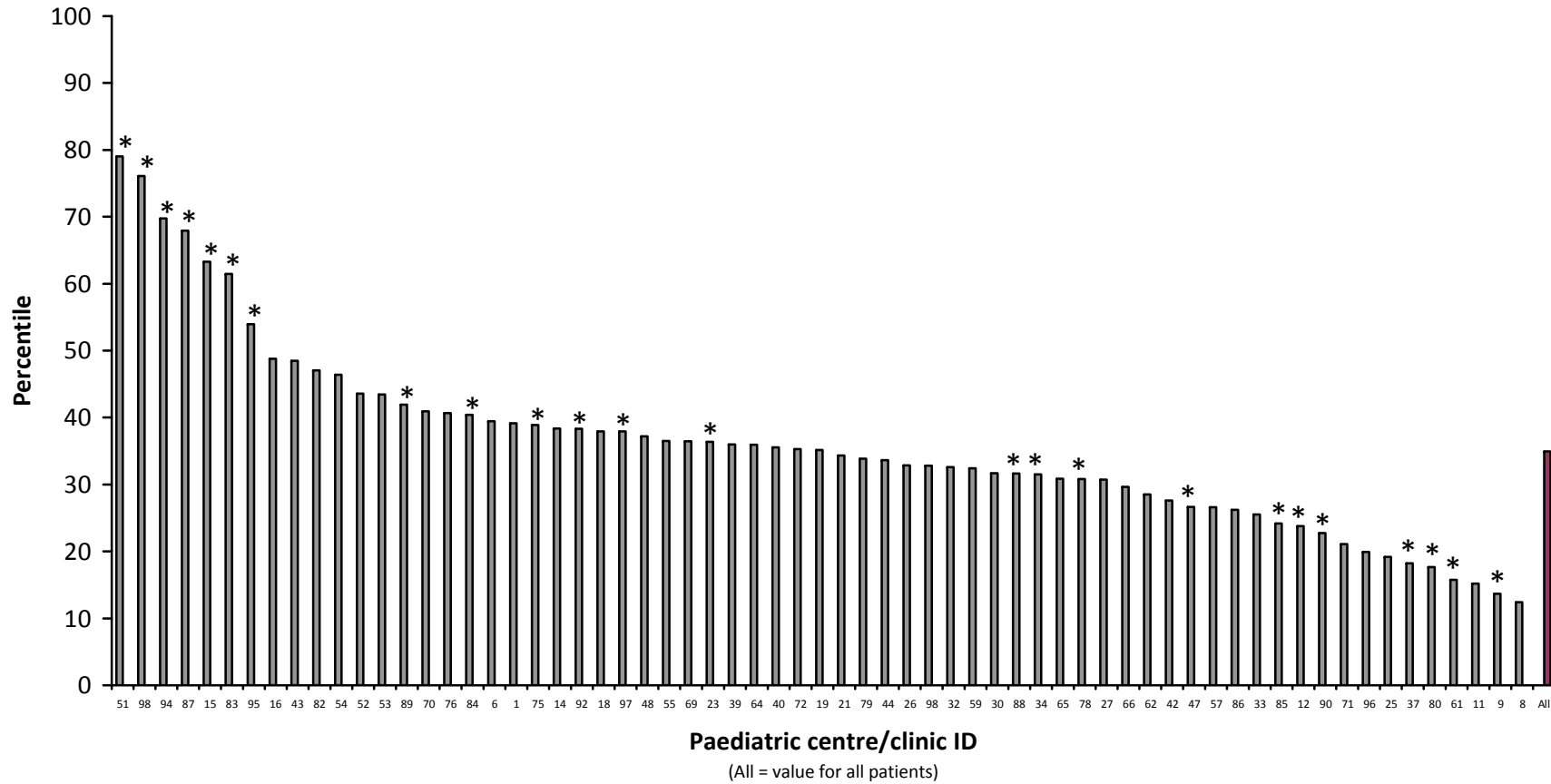
2.5 Median height (percentile) by paediatric care network and lone centre/clinic



Network clinics are reported under the ID of the lead specialist centre for the network.
The median height (percentile) for paediatric care networks and lone centres/clinics is 35.3 (min=23.4, max=69.7).

** Centre/clinic with fewer than 20 patients at annual review*

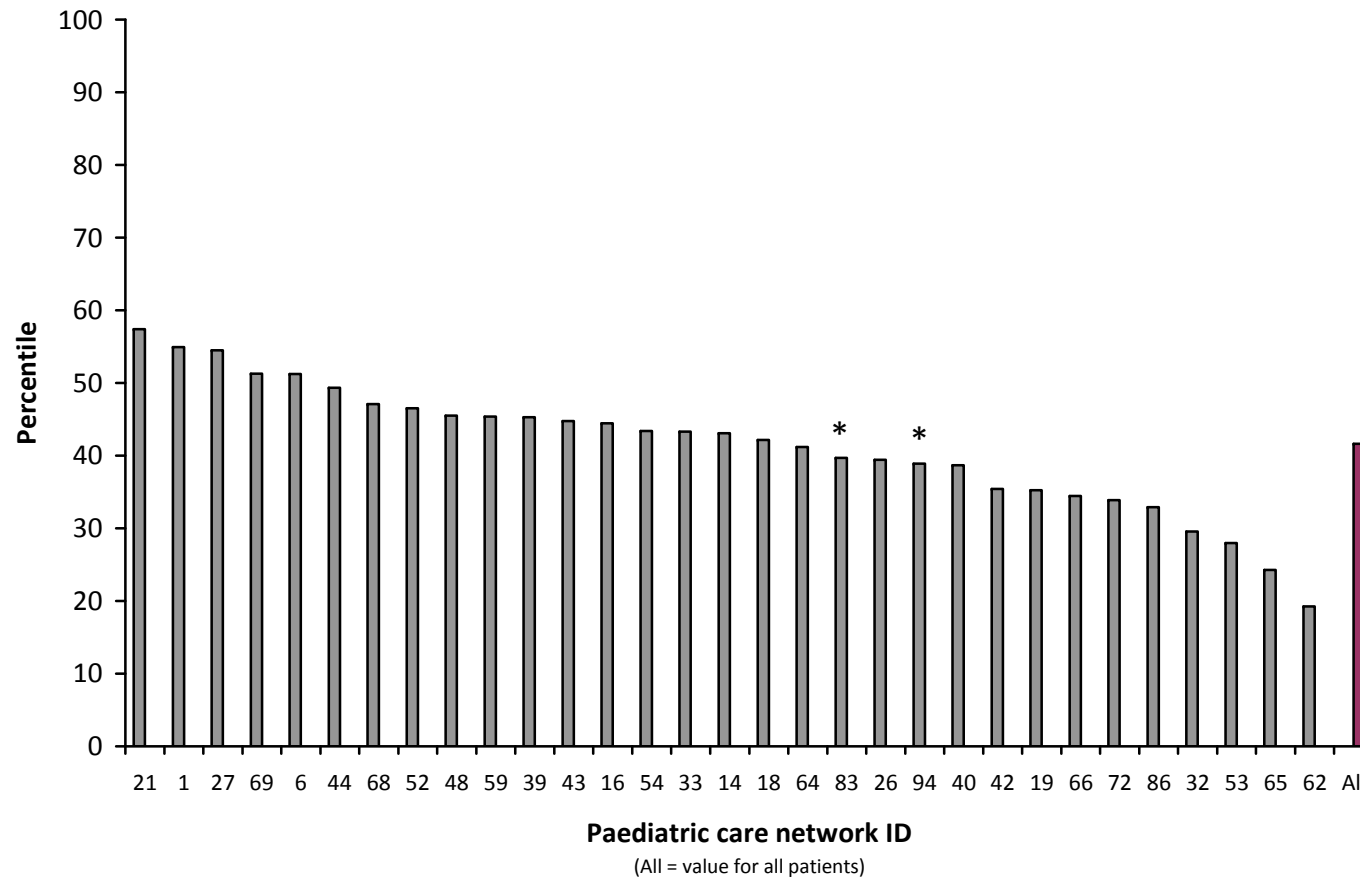
2.6 Median height (percentile) by paediatric centre/clinic



The median height (percentile) for paediatric care centres/clinics is 35.2 (min=12.4, max=79.0).

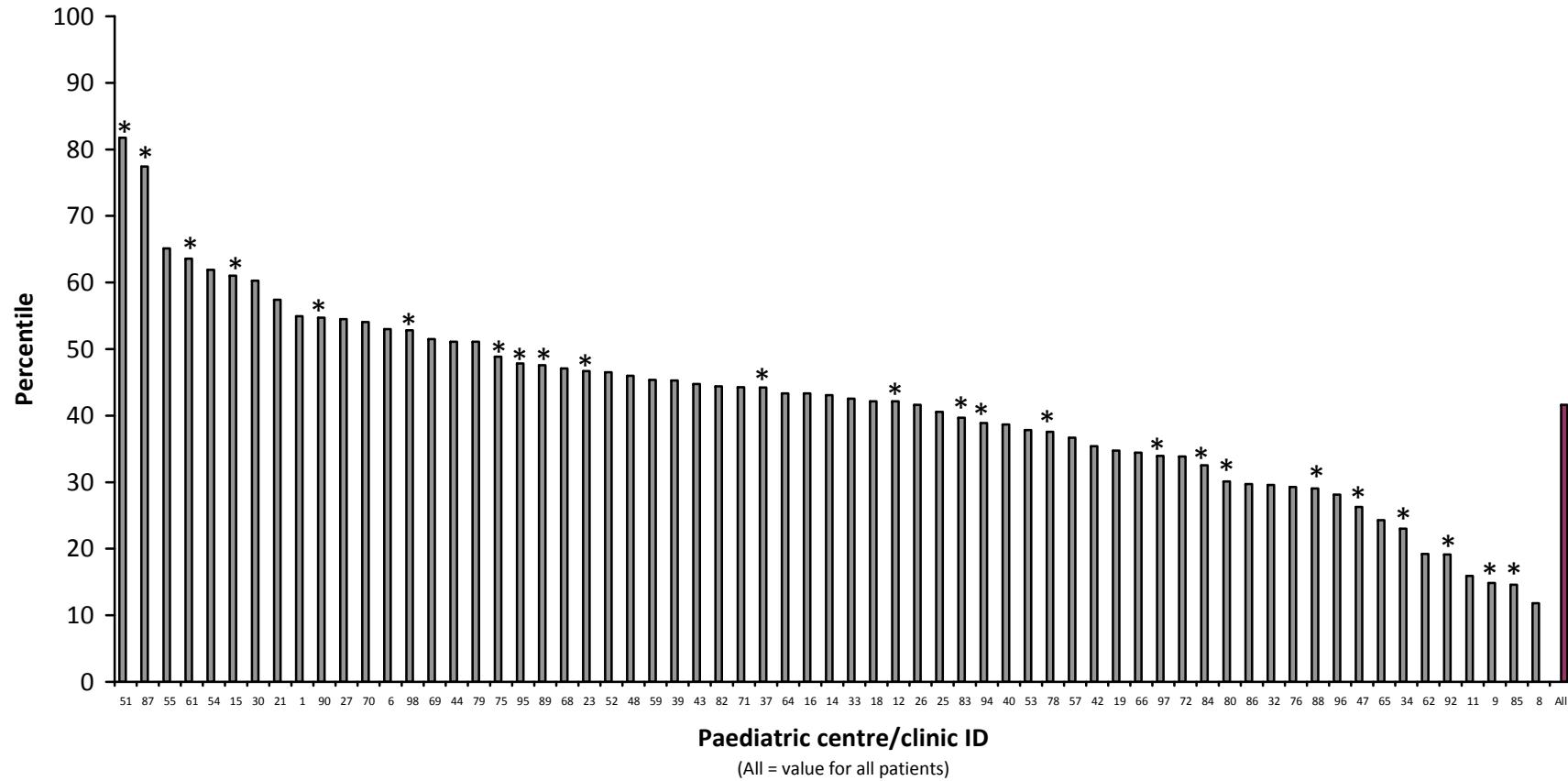
* Centre/clinic with fewer than 20 patients at annual review

2.7 Median weight (percentile) by paediatric care network and lone centre/clinic



Network clinics are reported under the ID of the lead specialist centre for the network.
 The median weight (percentile) for paediatric care networks and lone centres/clinics is 43.1 (min=19.2, max=57.4).
 * Centre/clinic with fewer than 20 patients at annual review

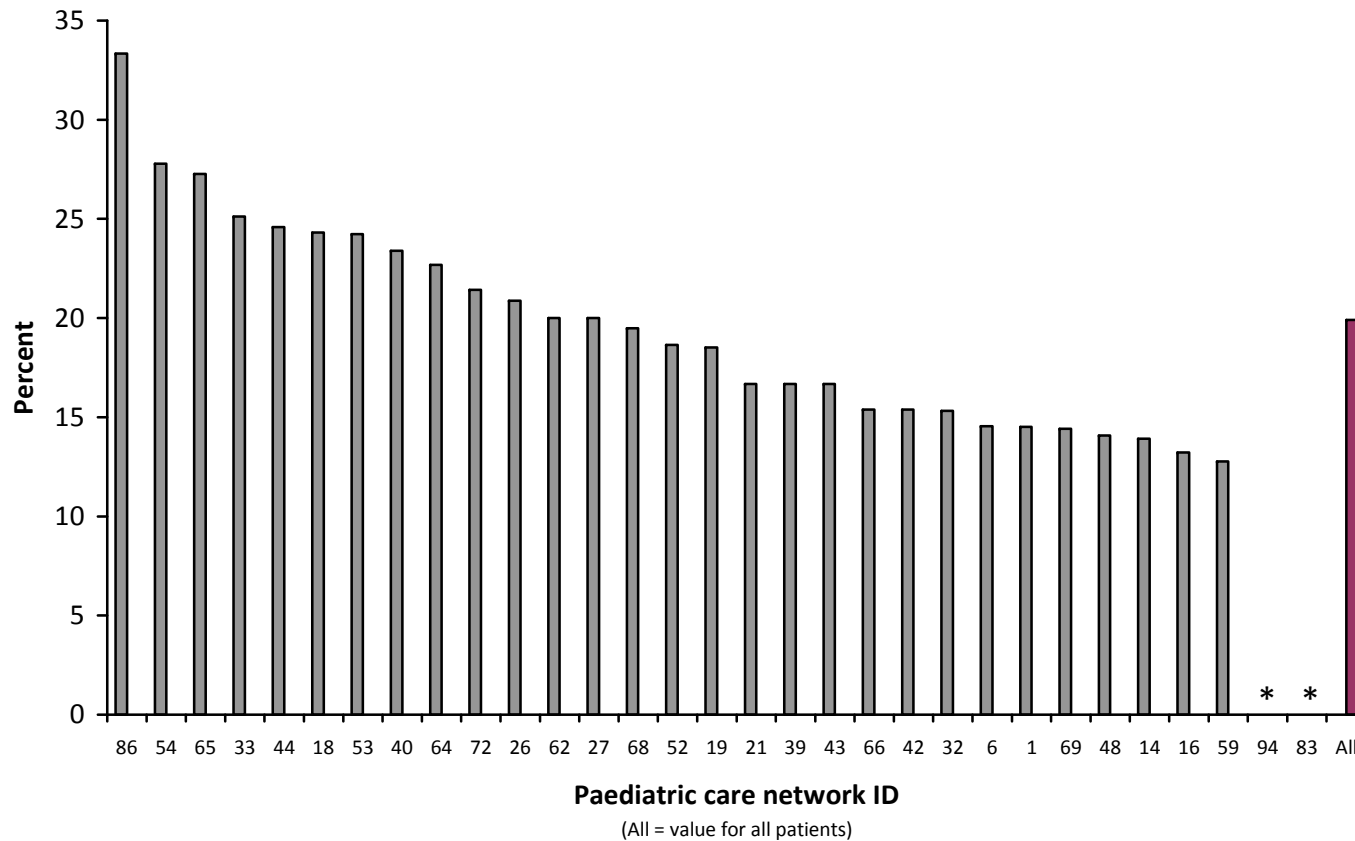
2.8 Median weight (percentile) by paediatric centre/clinic



The median weight (percentile) for paediatric care centres/clinics is 43.1 (min=11.8, max=81.8).

* Centre/clinic with fewer than 20 patients at annual review

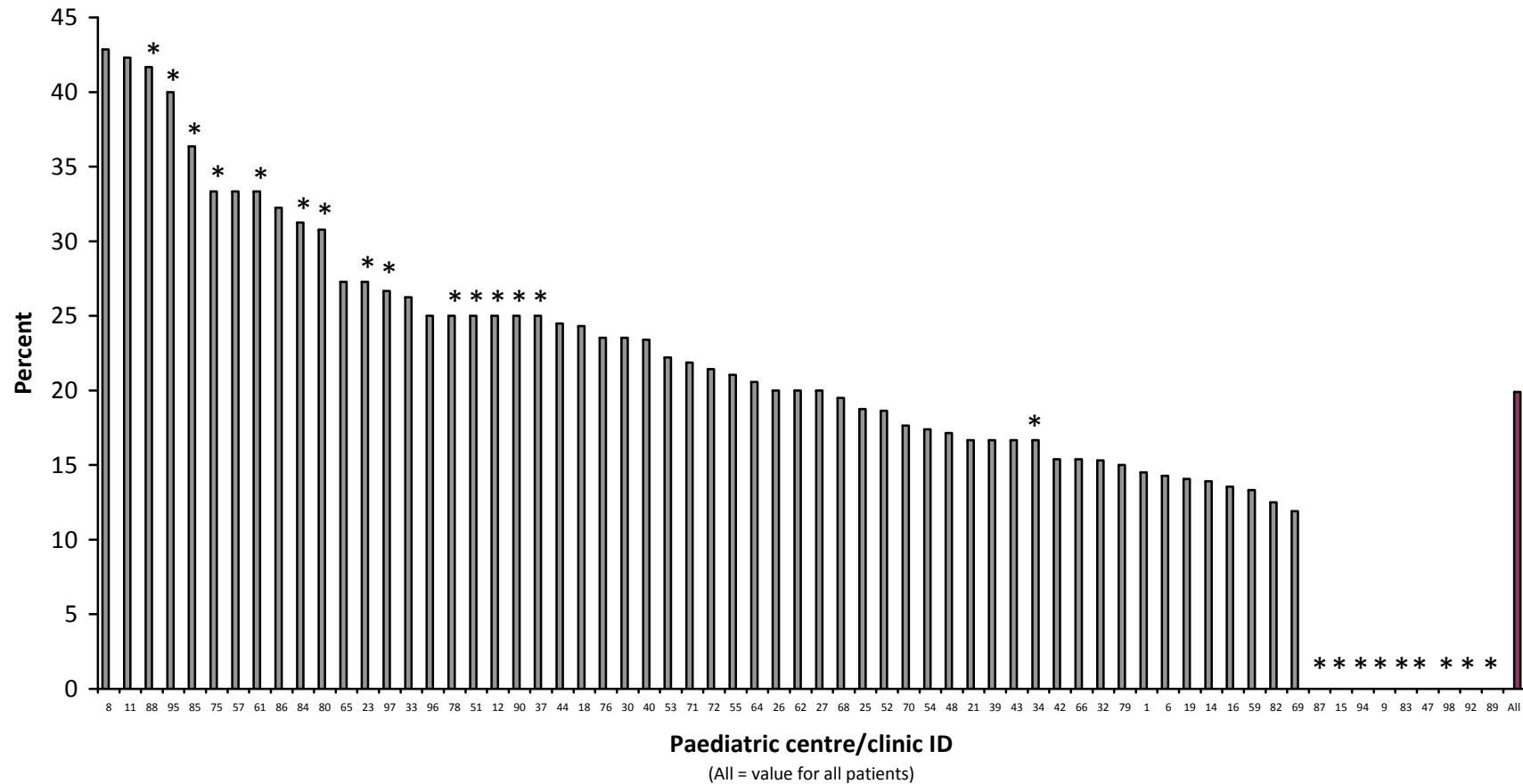
2.9 Proportion of patients below 10th percentile for height by paediatric care network and lone centre/clinic



Network clinics are reported under the ID of the lead specialist centre for the network.
The median proportion of patients below the 10th percentile for height for paediatric care networks and lone centres/clinics is 18.5% (min=0%, max=33.3%).

** Centre/clinic with fewer than 20 patients at annual review*

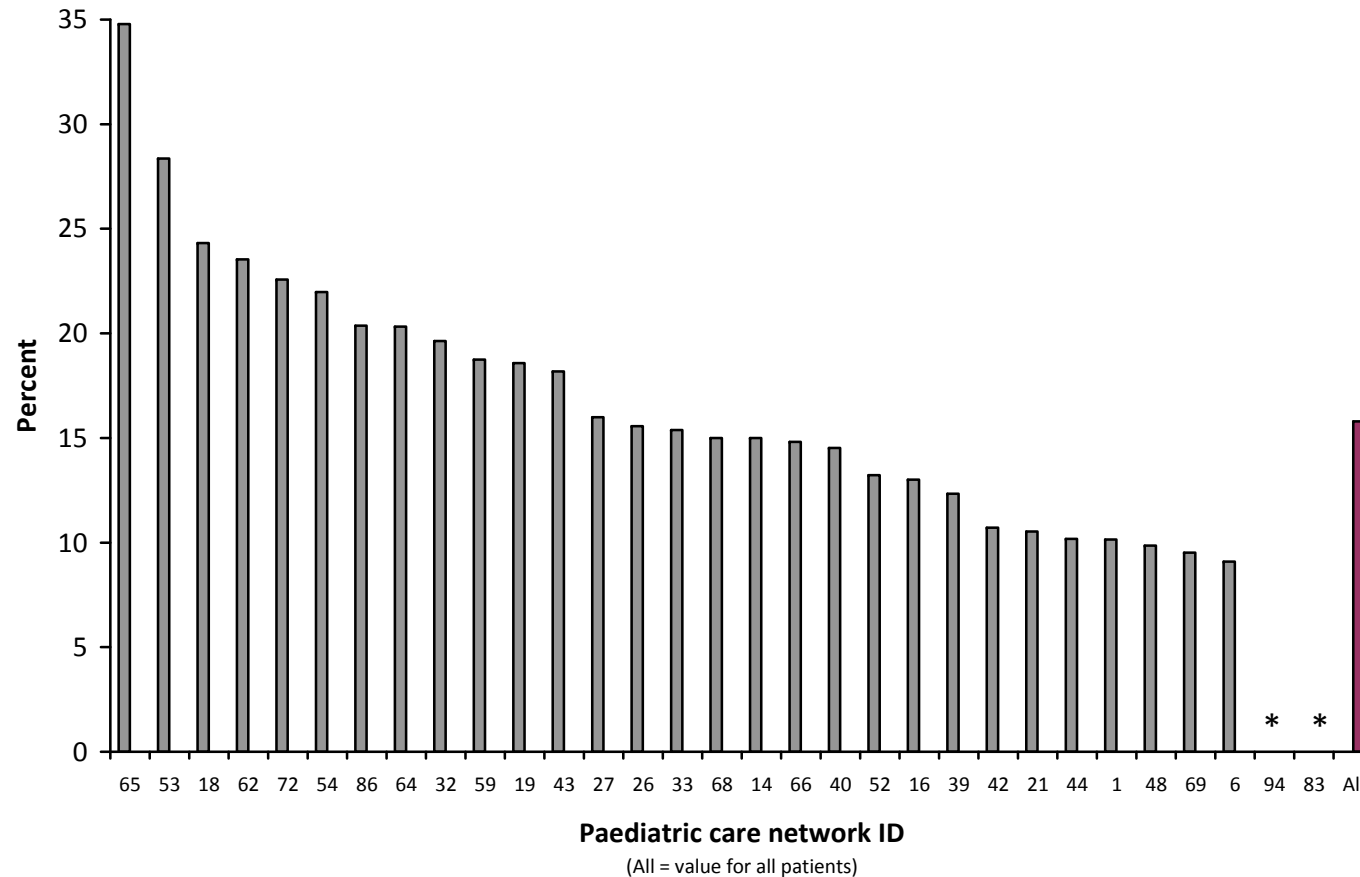
2.10 Proportion of patients below 10th percentile for height by paediatric centre/clinic



The median proportion of patients below the 10th percentile for height for paediatric care centres/clinics is 20.0% (min=0%, max=42.9%).

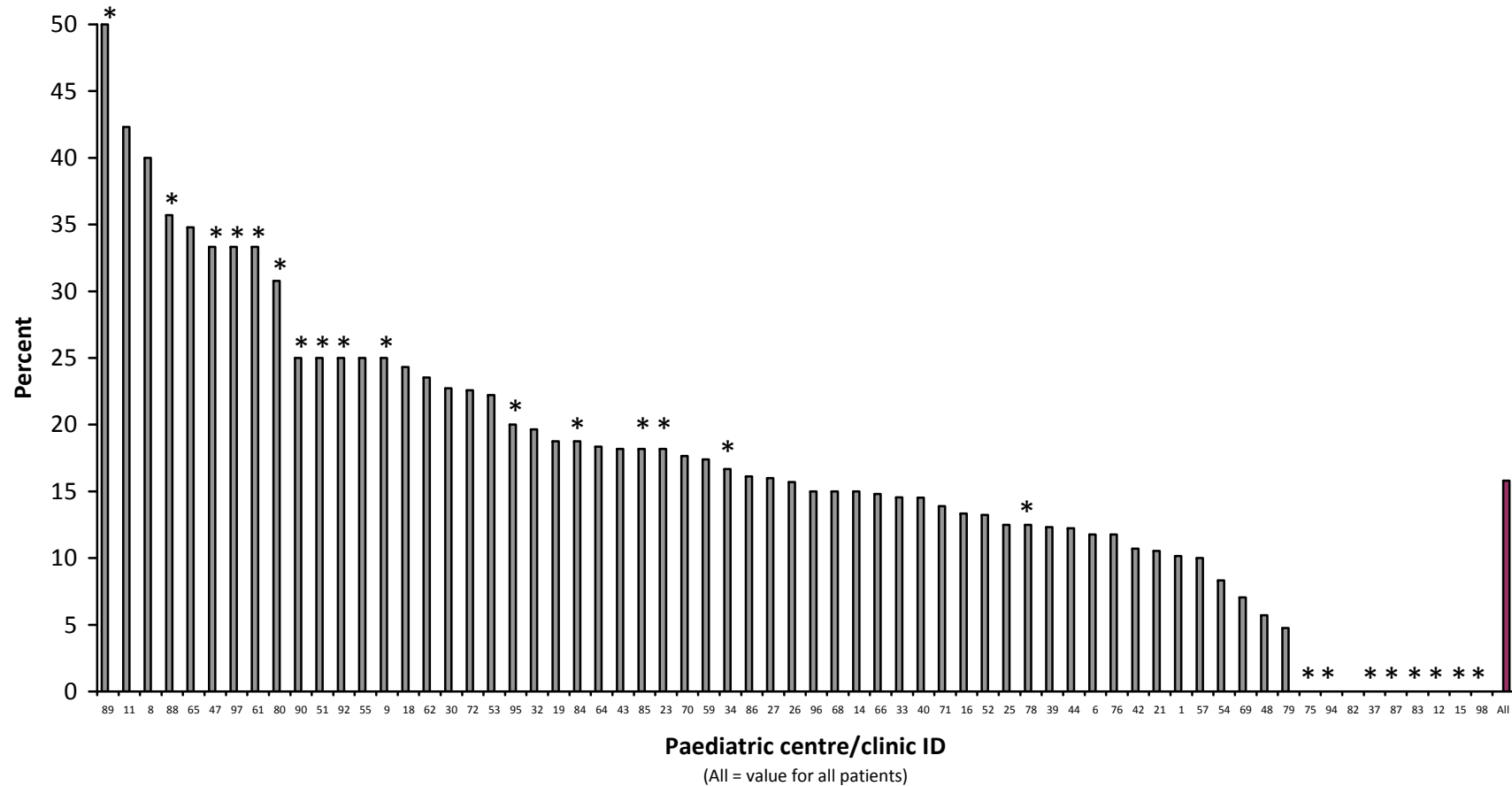
* Centre/clinic with fewer than 20 patients at annual review

2.11 Proportion of patients below 10th percentile for weight by paediatric care network and lone centre/clinic



Network clinics are reported under the ID of the lead specialist centre for the network.
 The median proportion of patients below the 10th percentile for weight for paediatric care networks and lone centres/clinics is 15.0% (min=0%, max=34.8%). * Centre/clinic with fewer than 20 patients at annual review

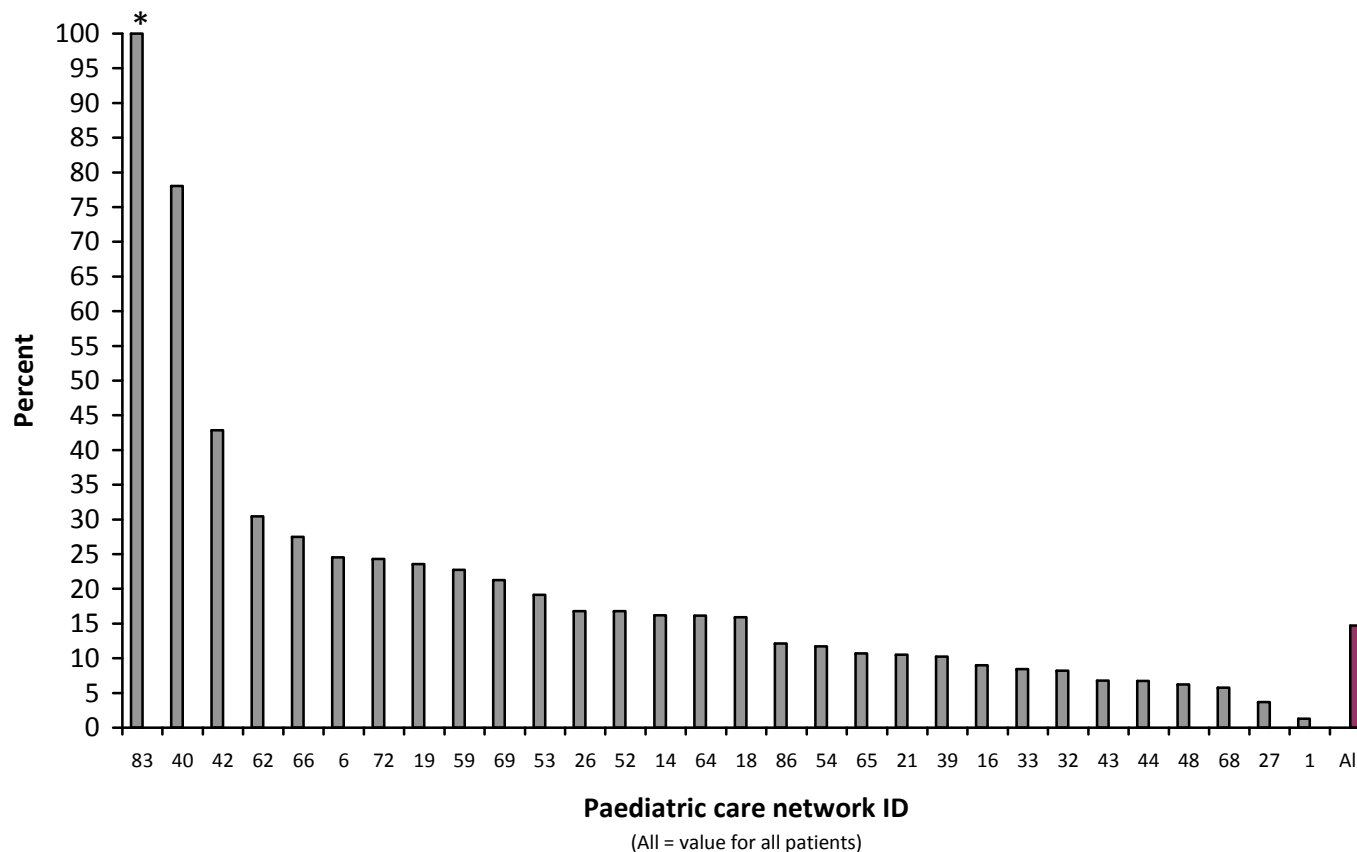
2.12 Proportion of patients below 10th percentile for weight by paediatric centre/clinic



The median proportion of patients below the 10th percentile for weight for paediatric care centres/clinics is 15.7% (min=0%, max=50%).

* Centre/clinic with fewer than 20 patients at annual review

2.13 Proportion of patients with chronic *P. aeruginosa* by paediatric care network and lone centre/clinic



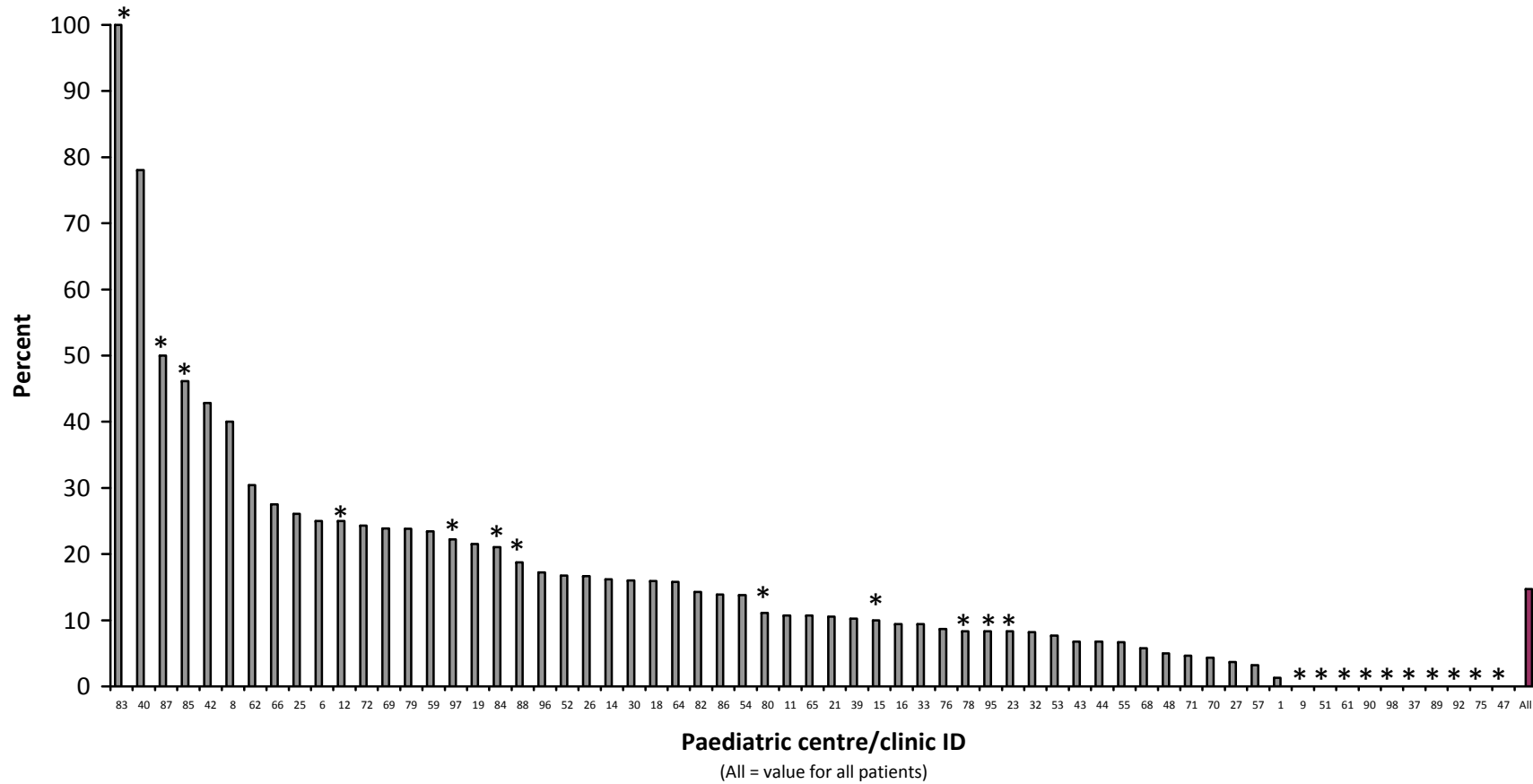
Network clinics are reported under the ID of the lead specialist centre for the network.

The median proportion of patients with chronic *P. aeruginosa* for paediatric care networks and lone centres/clinics is 16.0% (min=1.3%, max=100%).

Data were missing for centre 94*.

* Centre/clinic with fewer than 20 patients at annual review

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



The median proportion of patients with chronic *P. aeruginosa* for paediatric care centres/clinics is 10.7% (min=0%, max=100%).

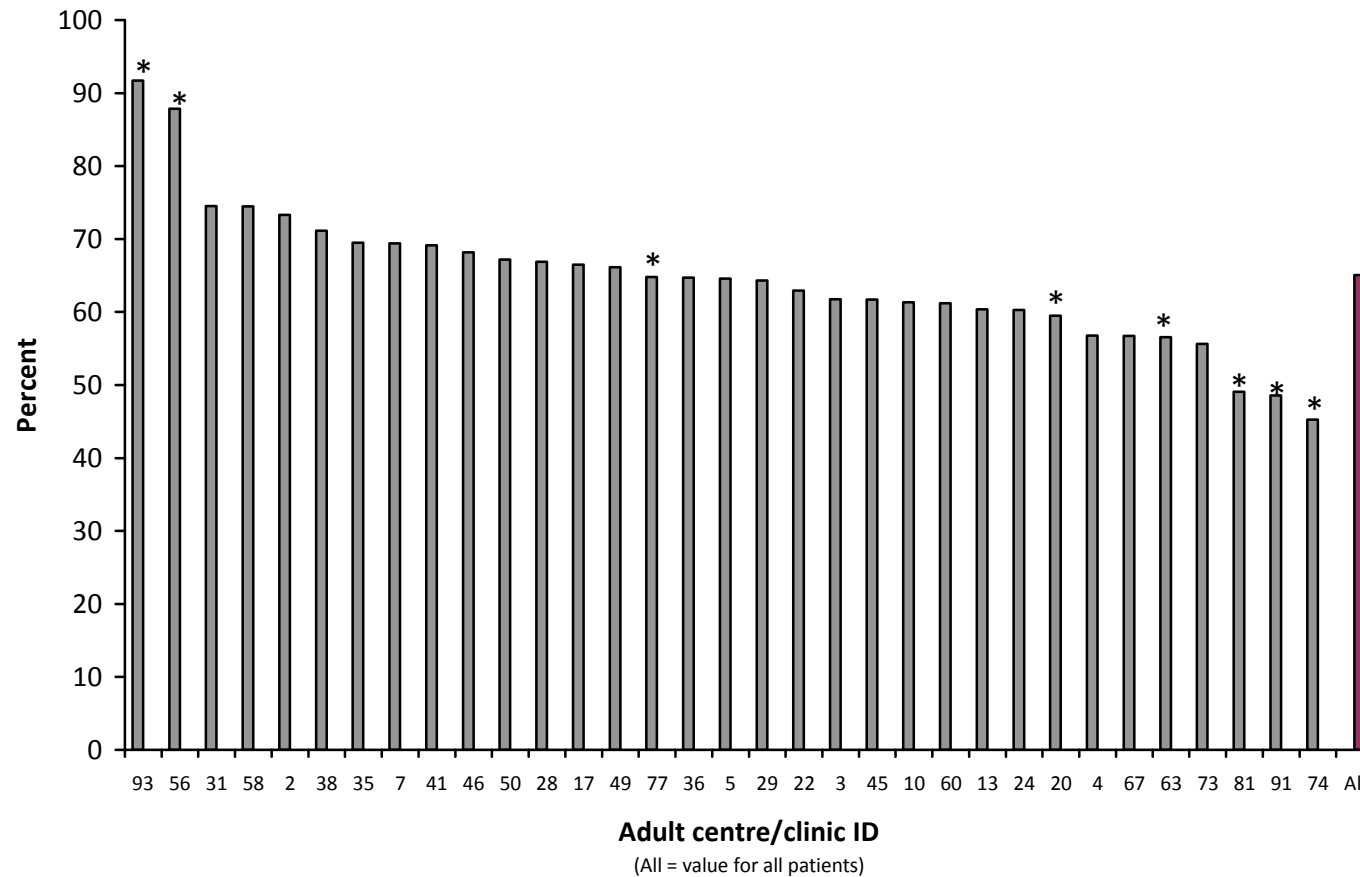
Data were missing for centres 34* and 94*.

* Centre/clinic with fewer than 20 patients at annual review

Section 3:

Comparison of outcomes for adult care centres and clinics (based on 3182 patients with complete annual review data in 2008)

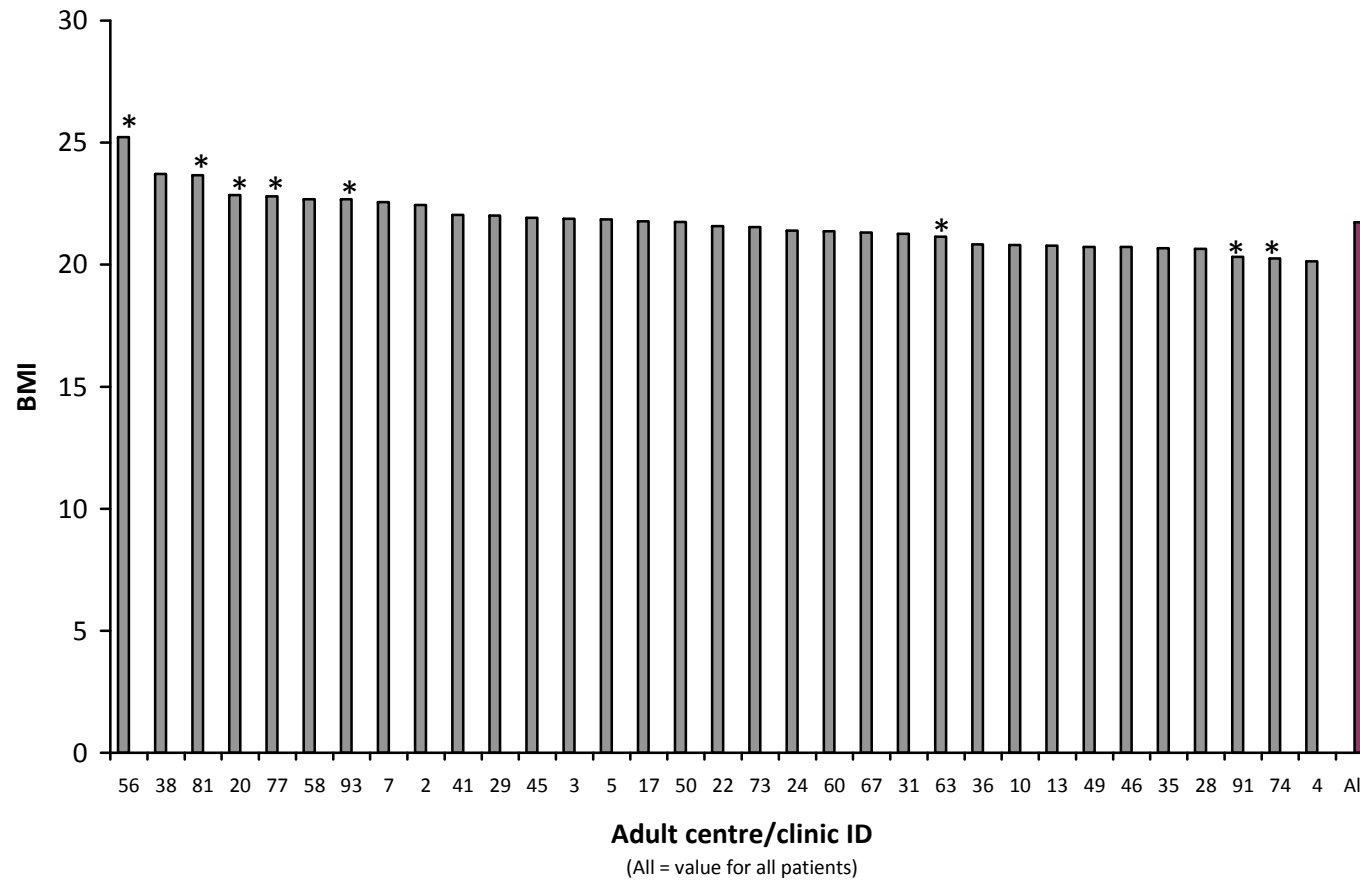
3.1 Median FEV₁ (% predicted) by adult centre/clinic



The median FEV₁ (% predicted) for adult care centres is 64.6% (min=45.3%, max=91.7%).

* Centre/clinic with fewer than 20 patients at annual review

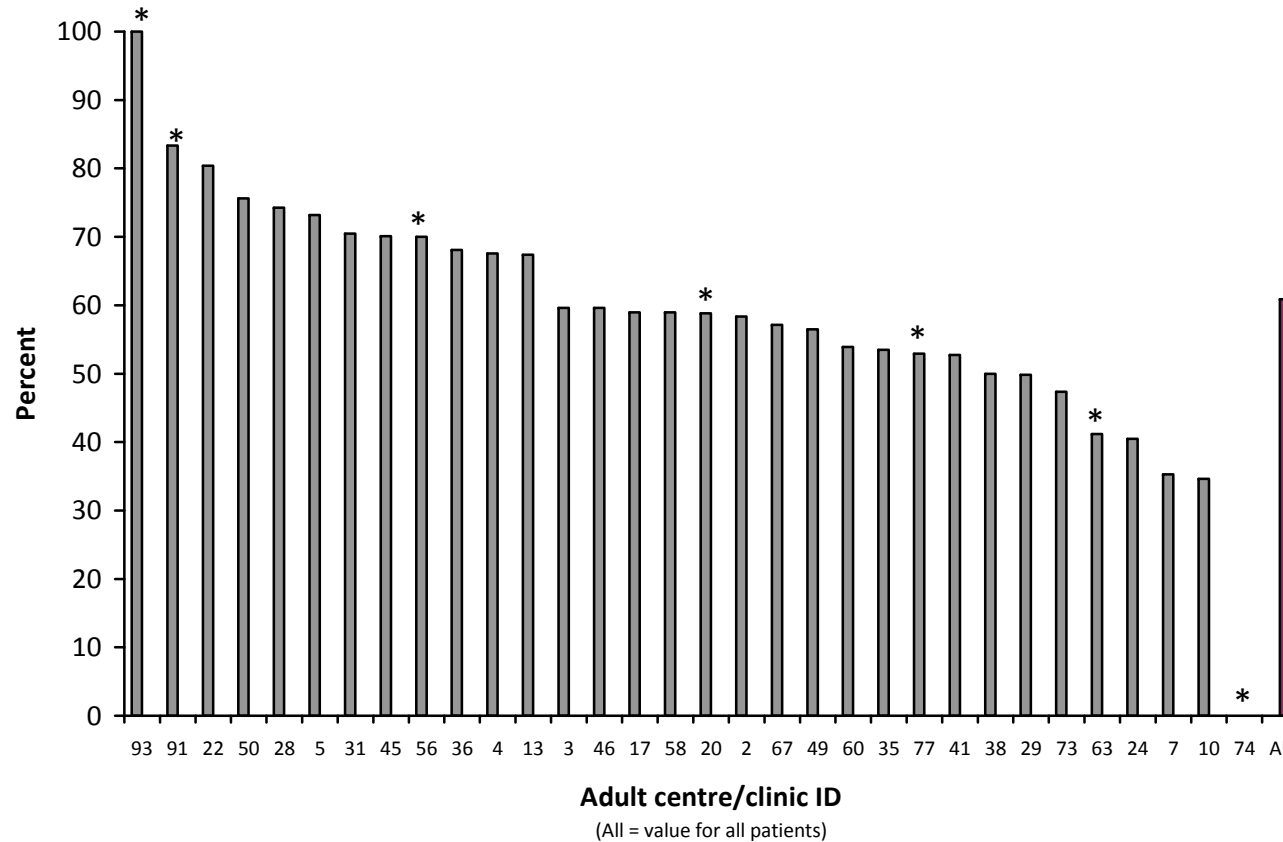
3.2 Median BMI by adult centre/clinic



The median BMI value for adult care centres/clinics is 21.6 (min=20.1, max=25.2).

* Centre/clinic with fewer than 20 patients at annual review

3.3 Proportion of patients with chronic *P. aeruginosa* by adult centre/clinic



The median proportion of patients with chronic *P. aeruginosa* for adult care centres/clinics with is 58.9% (min=0%, max=100%).

Data were missing for centre 81*.

* Centre/clinic with fewer than 20 patients at annual review

Section 4:

Care centres/clinics providing data in 2008

4.1 Paediatric care centres/clinics providing data in 2008

	Patients registered	Patients with "complete data"
England		
Addenbrookes Hospital, Cambridge	47	41
Hinchingsbrooke Hospital, Huntingdon	11	11
Ipswich General Hospital	9	0
Peterborough District General Hospital	25	23
West Suffolk General Hospital, Bury St Edmunds	9	9
Alder Hey Children's Hospital, Liverpool	93	79
Arrowe Park Hospital, Wirral	21	0
Leighton Hospital, Crewe	14	13
Glan Clwyd Hospital, Rhyl	10	0
Royal Albert Edward Infirmary, Wigan	30	28
Warrington District General Hospital	26	23
Birmingham Children's Hospital	256	209
New Cross Hospital, Wolverhampton	47	45
Walsgrave Hospital, Coventry	34	23
Booth Hall Children's Hospital, Manchester	306	1
Bristol Royal Hospital for Children	85	65
Cheltenham General and Gloucestershire Royal Hospital	6	2
Great Ormond Street Hospital for Sick Children, London	165	120
Luton and Dunstable Hospital	27	18
Hull Royal Infirmary	39	31
James Cook University Hospital, Middlesbrough	26	0
John Radcliffe Hospital, Oxford	166	141
King's College Hospital, London	141	123
Royal Alexandra Children's Hospital, Brighton	31	30
Leicester Royal Infirmary	65	61
Norfolk and Norwich University Hospital	36	36
James Paget Hospital, Great Yarmouth	16	12
Queen Elizabeth Hospital, Kings Lynn	18	18
Nottingham City Hospital	82	28
Chesterfield Hospital	10	8
Derby Hospital	32	0
Kings Mill Hospital	15	2
Lincoln Hospital	25	19
Pilgrim Hospital, Boston	23	22
Queen Mary's Hospital for Children, Carshalton	47	42
Royal Brompton Hospital, London	336	205
Royal Devon and Exeter Hospital, Exeter	39	34
Derriford Hospital, Plymouth	33	30
North Devon District Hospital, Barnstaple	13	13
Royal Cornwall Hospital, Truro	24	23
Torbay Hospital	20	19
Royal London Hospital	104	92
Royal Victoria Infirmary, Newcastle	123	107
Bishop Auckland General Hospital	14	13

James Cook University Hospital, Middlesbrough	3	0
Sunderland Royal Hospital	18	5
West Cumberland Hospital, Whitehaven	11	7
Sheffield Children's Hospital	157	109
St James's University Hospital, Leeds	206	152
St. Luke's Hospital, Bradford	17	6
Southampton General Hospital	161	48
University Hospital Lewisham, London	41	38
University Hospital of North Staffordshire, Stoke-on-Trent	66	44
Princess Royal Hospital, Telford / Royal	25	23
Shrewsbury Hospital		
Northern Ireland		
Royal Belfast Hospital for Sick Children	230	162
Scotland		
Crosshouse Hospital, Ayr	39	23
Ninewells Hospital, Dundee	29	27
Raigmore Hospital, Inverness	25	23
Royal Aberdeen Children's Hospital	42	36
Royal Hospital for Sick Children, Edinburgh	129	93
Royal Hospital for Sick Children, Glasgow	201	136
Dumfries and Galloway Royal Infirmary	4	4
Wales		
Children's Hospital for Wales, Cardiff	86	61
Bronglais Hospital, Aberystwyth	5	3
Hereford County Hospital	8	5
Nevill Hall Hospital, Abergavenny	1	0
Princess of Wales Hospital, Bridgend	7	5
Royal Glamorgan Hospital, Llantrisant	7	5
Royal Gwent Hospital, Newport	36	21
Singleton Hospital, Swansea	37	35
West Wales General Hospital, Carmarthen	11	4
Withybush General Hospital, Haverfordwest	7	6

4.2 Adult care centres/clinics providing data in 2008

	Patients registered	Patients with "complete data"
England		
Bath Royal United Hospital	4	1
Birmingham Heartlands Hospital	280	242
New Cross Hospital, Wolverhampton	15	12
Bristol Royal Infirmary	103	83
Castle Hill Hospital, Hull	26	0
Churchill Hospital, Oxford	66	61
Derriford Hospital, Plymouth	36	36
Frimley Park Hospital, Camberley	109	82
Glenfield Hospital, Leicester	48	47
Kings College Hospital, London	102	81
Liverpool Heart and Chest Hospital	220	209
London Chest Hospital	136	116
Norfolk & Norwich University Hospital, Norwich	51	46
Northern General Hospital, Sheffield	111	103
Nottingham City Hospital	118	103
Papworth Hospital	222	136
Royal Brompton Hospital, London	658	435
Royal Cornwall Hospital, Truro	30	26
Royal Devon and Exeter Hospital, Exeter	45	28
North Devon District Hospital, Barnstaple	4	0
Royal Victoria Infirmary, Newcastle	206	186
Southampton General Hospital	124	110
Poole Hospital	17	17
Salisbury Hospital	3	0
St James's University Hospital, Leeds	359	338
Torbay Hospital	6	6
University Hospital Lewisham, London	40	38
University Hospital of North Staffordshire, Stoke-on-Trent	47	25
Wythenshawe Hospital, Manchester	313	239
Northern Ireland		
Belfast City Hospital	192	149
Scotland		
Aberdeen Royal Infirmary	53	47
Gartnavel General Hospital, Glasgow	130	17
Western General Hospital, Edinburgh	107	1
Dumfries and Galloway Royal Infirmary, Dumfries	4	0
Ninewells Hospital, Dundee	48	3
Raigmore Hospital, Inverness	20	19
Wales		
Llandough Hospital	152	140

Section 5:

Acknowledgements

6.1 UK CF Registry Steering Committee

Dr Diana Bilton (<i>Chair</i>)	Consultant Physician, Adult CF Unit Royal Brompton Hospital, London
Mrs Rosie Barnes	Chief Executive, Cystic Fibrosis Trust
Dr Keith Brownlee	Consultant Paediatrician, Regional CF Unit St James's University Hospital, Leeds
Ms Shona Charlton	Senior Commissioner South West Specialist Commissioning Group
Dr Geoffrey Carroll	Medical Director, Health Commission Wales
Ms Kathy Collins (<i>Caldicott Guardian</i>)	Nursing and Quality Adviser National Services Division, Scotland
Mrs Marian Dmochowska	Parent Representative
Dr Iolo Doull	Consultant Paediatrician, Paediatric CF Unit Children's Hospital for Wales, Cardiff
Professor Stuart Elborn	Consultant Physician, Adult CF Unit Belfast City Hospital
Dr Caroline Elston	Consultant Physician, Adult CF Unit King's College Hospital, London
Mr Alan Larsen	Director of Research and Finance, Cystic Fibrosis Trust
Dr Jim Littlewood	Chairman, Cystic Fibrosis Trust Retired Consultant Paediatrician
Ms Sue McLellen	Head of Specialised Commissioning, London Specialised Commissioning Group
Ms Helen Tilley	Lead Commissioner London Specialised Commissioning Group
Dr Sarah Walters	Patient Representative Epidemiologist, Birmingham University
Dr Martin Wildman	Consultant Physician, Adult CF Centre Northern General Hospital, Sheffield

6.2 Data analysis

Data analysis was performed by:

Paul Cullinan MD, FRCP – Professor in Occupational and Environmental Respiratory Disease

Stephanie MacNeil PhD – Medical Statistician

Department of Occupational and Environmental Medicine

National Heart and Lung Institute

Imperial College

London

Section 6:

Patient data questionnaire 2008

6.1 Demographic Data

Last Name		Last Name at birth (if diff)	
First Name			
County of Birth		Date of Birth	
Gender	<input type="checkbox"/> male	<input type="checkbox"/> female	
Race	<input type="checkbox"/> Caucasian	<input type="checkbox"/> Black African	<input type="checkbox"/> Black Caribbean
	<input type="checkbox"/> Black Other	<input type="checkbox"/> Indian	<input type="checkbox"/> Pakistani
	<input type="checkbox"/> Bangladeshi	<input type="checkbox"/> Chinese	<input type="checkbox"/> Asian Other
	<input type="checkbox"/> Mixed Race	<input type="checkbox"/> Individual preferred not to Answer	<input type="checkbox"/> Clinician preferred not to ask question
	<input type="checkbox"/> Other (please specify) _____		
NHS number	_____	CHI number	_____
Full Postcode	_____	GP Postcode	_____
Biological Mother's Ht		Biological Father's Ht.	
Full Postcode		_____	
Genotyped?	<input type="checkbox"/> Yes <input type="checkbox"/> No	Date	unknown
Mutation 1		Mutation 2	
Complications @ birth	<input type="checkbox"/> Meconium ileus/intestinal obstruction managed medically		
	<input type="checkbox"/> Meconium ileus/intestinal obstruction managed surgically		
	<input type="checkbox"/> None <input type="checkbox"/> Unknown		
Date of Death	_____	Date is an estimate	<input type="checkbox"/> Yes
Cause of Death	<input type="checkbox"/> Resp./cardioresp.	<input type="checkbox"/> Liver Disease/Failure	<input type="checkbox"/> Trauma
	<input type="checkbox"/> Suicide	<input type="checkbox"/> Transplant	<input type="checkbox"/> Other(spec)
Date of Diagnosis _____			
Diagnosis Suggested By	<input type="checkbox"/> Acute or persistent respiratory symptoms		<input type="checkbox"/> Oedema
	<input type="checkbox"/> Electrolyte imbalance		<input type="checkbox"/> Family history
	<input type="checkbox"/> Failure to thrive/malnutrition		<input type="checkbox"/> Genotype
	<input type="checkbox"/> Meconium ileus/other intestinal obstruction		<input type="checkbox"/> Liver problems
	<input type="checkbox"/> Nasal polyps/sinus disease		<input type="checkbox"/> Prenatal screening
	<input type="checkbox"/> Steatorrhea/abnormal stools/malabsorption		<input type="checkbox"/> Neonatal screening
	<input type="checkbox"/> Other (specify)		<input type="checkbox"/> Rectal prolapse
			<input type="checkbox"/> Unknown
Chloride Sweat Test Value	_____	<input type="checkbox"/> Unknown	
CF Diagnosis Reversed	<input type="checkbox"/>		
if yes, select reasons	<input type="checkbox"/> Transepithelial Potential Differences		<input type="checkbox"/> DNA Analysis
	<input type="checkbox"/> Repeat Normal Sweat Testing		<input type="checkbox"/> Other

6.2 Annual Survey

Information of Patient :

Patient Name _____

Birth Date _____

Patient ID _____

Annual Survey Year _____

PATIENT STATUS :

Since last annual survey, this patient was Seen at this Centre/Clinic

Transferred to another Centre/Clinic (seen or not seen)

Patient's date of death was _____

Full Postcode _____ foreign unknown Confirm full postcode

PULMONARY :

Oxygen therapy

Yes, Continuously
 Yes, Nocturnal and/or with exertion
 Yes, During exacerbation
 Yes, prn
 No
 Unknown

Non-invasive ventilation Yes No Unknown

Chest XRay Yes No Unknown

Dexa Scan Normal Abnormal Not Done

Liver USS Normal Abnormal Not Done

Pseudomonas Yes No Unknown

Intermittent Chronic

Staph.aureus Yes No Unknown

Intermittent Chronic

Flu Vaccine Yes No Unknown

Pneumovax Yes No Unknown

No
 Yes, Occasionally
 Yes, Regularly, less than 1 ppd
 Yes, Regularly, 1 ppd or more
 Decline to answer
 Not Known
 Not Applicable

Smoke Cigarettes

GROWTH AND NUTRITION :

pancreatic Enzyme Supplements Yes No Unknown

Fat sol. vitamin levels meas. Yes No Unknown

Testosterone (male only) Yes No Unknown

Oestrogen (female only) Yes No Unknown

daily Ursodeoxycholic acid Yes No Unknown

UPDATE ON CFRD STATUS :

Not done
 Normal Glucose Metabolism (includes normal, random, fasting, or OGTT)
 Impaired Glucose Tolerance (FBG 6.1-6.9, 2-h PG 7.8-11.0 mmol/L)
 CF-related diabetes
 CF-related diabetes w/out fasting hyperglycemia (FBG < 6.0, 2-h PG >= 11.1mmol/L)
 CF-related diabetes with fasting hyperglycemia (FBG >=6.0 mmol/L)

Hgb A1C Val _____ Not Done Unknown

Retinopathy Yes No Unknown

Microalbuminuria Yes No Unknown

Receive treatment for CFRD Yes No

Dietary change Oral hypoglyc. agnts
 Intermittent Insulin Chronic req. Insulin

TRANSPLANTATION :

evaluated at transplant centre Yes No Unknown

If yes, were they accepted declined deferred

receive a transplant Yes No Unknown

number of transplants

type of transplants

Lung: Bilateral Lung: Heart/Lung Lung: Lobar/cadaveric

Lung: Lobar/living donor Liver

Other (spec) _____

month and day of first lung transplant _____

post transplant complications? Yes No

If Yes, types of complications

Bronchiolitis obliterans syndrome

Lympho-proliferative disorder

Atypical infection

Renal failure

Other (spec) _____

CLINICAL TRIALS :

participated in any ethically approved protocols? Yes No Unknown

if yes, which? CF Trust Funded Studies In house studies

Commercial Studies Other (please specify) _____

IV DAYS :

Hospital IV Days _____

Home IV Days _____

SOCIO-ECONOMIC STATUS :

Patient Education Level	<input type="checkbox"/> Less than GCSE	Mother's Education Level	<input type="checkbox"/> Less than GCSE
	<input type="checkbox"/> GCSE or equivalent		<input type="checkbox"/> GCSE or equivalent
	<input type="checkbox"/> A Level or equivalent		<input type="checkbox"/> A Level or equivalent
	<input type="checkbox"/> College		<input type="checkbox"/> College
	<input type="checkbox"/> University		<input type="checkbox"/> University
	<input type="checkbox"/> Unknown		<input type="checkbox"/> Unknown
	<input type="checkbox"/> Not Applicable		<input type="checkbox"/> Not Applicable
Father's Education Level	<input type="checkbox"/> Less than GCSE	Spouse/Partner's Education Level (Age 16 and older only)	<input type="checkbox"/> Less than GCSE
	<input type="checkbox"/> GCSE or equivalent		<input type="checkbox"/> GCSE or equivalent
	<input type="checkbox"/> A Level or equivalent		<input type="checkbox"/> A Level or equivalent
	<input type="checkbox"/> College		<input type="checkbox"/> College
	<input type="checkbox"/> University		<input type="checkbox"/> University
	<input type="checkbox"/> Unknown		<input type="checkbox"/> Unknown
	<input type="checkbox"/> Not Applicable		<input type="checkbox"/> Not Applicable

AGE 16 AND OLDER :

Marital Status Single (never married) Living Together Married

Separated Divorced Widowed

Unknown

Employment Full time Part time Full time homemaker

Student Unemployed Disabled

Retired Unknown

PREGNANCY :

Since last annual review, was patient pregnant? Yes No Unknown

If yes, Outcome Live Birth Still Birth Spontaneous Abortion Therapeutic Abortion

Undelivered Unknown

B.CEPACIA : If B.Cepacia complex was isolate, was the identification validated at a B.cepacia reference lab? Yes No Unknown

6.3 Encounter: Nutrition

Information of Patient :

Patient Name _____
 Birth Date _____
 Patient ID _____

Encounter Date _____
 Encounter Type Annual Review encounter Other encounter
 This encounter is part of A Clinical Visit
 A Hospitalisation Care Episode
 A Home IV Care Episode

PATIENT STATUS:

At the time of clinical visit, the patient was stable not stable
 HEIGHT: _____ cm. in. Not Measured
 WEIGHT: _____ kg. lb. Not Measured

NUTRITIONAL ASSESSMENT:

Patient was seen by a Dietitian Yes No
 Assessment of oral intake Done Not Done
 Is patient currently receiving supplemental feeding? Yes No Unknown
 If Yes, indicate Feeding Route:
 oral supplementation
 nasogastric tube
 gastrostomy tube/button
 jejunal tube
 total parenteral nutrition
 Is patient currently receiving pancreatic enzyme supplements? Yes No Unknown
 Creon 5000 scoop
 Creon 8000
 Creon 10000
 Creon 25000
 Creon 40000
 Pancrease 5000
 Nutrizyme
 Other, Please specify _____
 Number per day _____
 Acid Blocker (Daily use. Check all that apply since last visit)
 Antacids
 H2 Blocker
 Proton Pump Inhibitor
 None
 Unknown

6.4 Encounter: Pulmonary Assessment

Information of Patient :

Patient Name _____

Birth Date _____

Patient ID _____

Encounter Date _____

Encounter Type Annual Review encounter Other encounter

This encounter is part of A Clinical Visit

A Hospitalisation Care Episode

A Home IV Care Episode

PATIENT STATUS:

At the time of clinical visit, the patient was stable not stable

HEIGHT: _____ cm. in. Not Measured

WEIGHT: _____ kg. lb. Not Measured

PULMONARY FUNCTION TESTS (PFTs):

If no values given, select reason Not Done Unable to perform reliable test

FVC measure _____ (L)

FEV1 measure _____ (L)

FEF25-75 Measure _____ (L)

PULMONARY ASSESSMENT

Was the patient seen by a doctor at this visit? Yes No Unknown

Based on the assessment of the care team at this visit, was the patient experiencing an increase in respiratory symptoms or a pulmonary exacerbation? Yes No Unknown

If the patient was experiencing an increase in respiratory symptoms or a pulmonary exacerbation at this visit (i.e., the question above was answered yes), then select the treatment option below that best matches the prescribed treatment plan.

- Incr. airway clearance exercise and/or bronchodilators
- Oral non-quinolone antibiotic
- Oral macrolide antibiotic
- Oral quinolone antibiotic
- Inh. Antibiotic
- Inh. antibi. & oral non-quinolone antibi.
- Inh. antibi. & oral macrolide antibiotic
- Inh. antibi. & oral quinolone antibiotic
- hosp/home administ. IV antibi. (w/in 2 weeks of visit)
- None of the above
- Other (specify) _____

6.5 Encounter: Pulmonary Therapies

Information of Patient :

Patient Name _____

Birth Date _____

Patient ID _____

Encounter Date _____

Encounter Type Annual Review encounter Other encounter

This encounter is part of A Clinical Visit

A Hospitalisation Care Episode

A Home IV Care Episode

PATIENT STATUS:

At the time of clinical visit, the patient was stable not stable

HEIGHT: _____ cm. in. Not Measured

WEIGHT: _____ kg. lb. Not Measured

	Technique	Primary (select one)	Secondary (select all that apply)
Airway Clearance Techniques (check only one primary means of airway clearance and all secondary forms of airway clearance that apply)	Positive Expiratory Pressure (PEP)	<input type="checkbox"/>	<input type="checkbox"/>
	Postural drainage with clapping	<input type="checkbox"/>	<input type="checkbox"/>
	Forced expiration techniques (eg, autogenic drainage, huff cough, active cycle breathing)	<input type="checkbox"/>	<input type="checkbox"/>
	Oscillating PEP (eg, Flutter, acapella, IPV)	<input type="checkbox"/>	<input type="checkbox"/>
	High frequency chest wall compression (eg, Vest)	<input type="checkbox"/>	<input type="checkbox"/>
	Exercise	<input type="checkbox"/>	<input type="checkbox"/>
	Other (specify) _____	<input type="checkbox"/>	<input type="checkbox"/>
	None of the above	<input type="checkbox"/>	<input type="checkbox"/>
Pulmonary Medications used since last encounter (check all that applied as of last clinical encounter)	Mucolytics:	Frequency:	
	<input type="checkbox"/> DNase	<input type="checkbox"/> 2.5 mg BID	<input type="checkbox"/> 2.5 mg od
	<input type="checkbox"/> Acetylcysteine	<input type="checkbox"/> other regimen (different dose or freq)	
	Inhaled antibiotics:		
	<input type="checkbox"/> Tobramycin solution for inhalation	<input type="checkbox"/> 300 mg BID alternate month schedule	<input type="checkbox"/> 300 mg BID continuous
	<input type="checkbox"/> Other aminoglycoside	<input type="checkbox"/> Other regimen (different dose or freq)	
	<input type="checkbox"/> Colistin	<input type="checkbox"/> Alternate Month	<input type="checkbox"/> Continuous
	<input type="checkbox"/> Promixin	<input type="checkbox"/> Other regimen (different dose or freq)	
	<input type="checkbox"/> Chronic macrolide antibiotic	<input type="checkbox"/> Alternate Month	<input type="checkbox"/> Continuous
	<input type="checkbox"/> Chronic oral antibiotic (i.e. not prescribed to treat an exacerbation)	<input type="checkbox"/> Other regimen (different dose or freq)	
	Check all that apply:		
	<input type="checkbox"/> Quinolone		
	<input type="checkbox"/> Cephalosporin		
	<input type="checkbox"/> Amoxicillin		
	<input type="checkbox"/> Tetracycline		
	<input type="checkbox"/> Flucloxacillin		
	<input type="checkbox"/> Cotrimoxazole		
	<input type="checkbox"/> Other		
	<input type="checkbox"/> High-dose ibuprofen (e.g. (Total mg/dose) _____mg 25-30 mg/kg)		
	<input type="checkbox"/> Hypertonic saline Concentration % _____ (3 to 10)		

	Frequency
	<input type="checkbox"/> QD <input type="checkbox"/> BID <input type="checkbox"/> Other
<i>Bronchodilators (oral):</i>	
<input type="checkbox"/>	Beta agonist
<input type="checkbox"/>	Theophylline product
<i>Bronchodilators (inhaled)</i>	
Check all that apply:	
<input type="checkbox"/>	Short acting beta agonist
<input type="checkbox"/>	Long acting beta agonist
<input type="checkbox"/>	Short acting anticholinergic
<input type="checkbox"/>	Long acting anticholinergic
<input type="checkbox"/>	Combination beta agonist and anticholinergic
<i>Corticosteroids</i>	
<input type="checkbox"/>	Oral
<input type="checkbox"/>	Inhaled
<input type="checkbox"/>	Inhaled in combination with a bronchodilator
<input type="checkbox"/>	Leukotriene modifiers
<input type="checkbox"/>	Mast cell stabilizers
<input type="checkbox"/>	Antifungals
<i>Drug Intolerance</i>	
<input type="checkbox"/>	DNase
<input type="checkbox"/>	Tobramycin solution for inhalation
<input type="checkbox"/>	Colistin
<input type="checkbox"/>	Macrolide antibiotics
<input type="checkbox"/>	High-dose ibuprofen
<input type="checkbox"/>	Hypertonic saline
<input type="checkbox"/>	IV antibiotics, Please Specify : _____
<input type="checkbox"/>	This patient is not on any of the above pulmonary medications

6.6 Encounter: Respiratory Microbiology

Information of Patient :

Patient Name _____

Birth Date _____

Patient ID _____

Encounter Date _____

Encounter Type Annual Review encounter Other encounter

This encounter is part of A Clinical Visit
 A Hospitalisation Care Episode
 A Home IV Care Episode

PATIENT STATUS:

At the time of clinical visit, the patient was stable not stable

HEIGHT: _____ cm. in. Not Measured

WEIGHT: _____ kg. lb. Not Measured

RESPIRATORY MICROBIOLOGY:

If culture was performed, where was it performed? At CF Centre/clinic Elsewhere

Date of culture _____

Type of Culture sputum
 throat/nasal
 bronchoscopy
 no growth/sterile culture
 normal flora
 Pseudomonas aeruginosa
 mucoid
 non-mucoid
 unknown
 Burkholderia cepacia complex
 B. Cenocepacia
 B. Multivorans
 Other Burkholderia Cepacia
 Stenotrophomonas (Xanthomonas) maltophilia
 Other pseudomonas species
 Staphylococcus aureus
 MRSA (methicillin resistant Staph aureus)
 Haemophilus influenzae (any species)
 Aspergillus (any species)
 non-tuberculous mycobacterium
 Escherichia coli (E coli)
 Klebsiella (any species)
 Other gram negative (e.g. Burkholderia gladioli is NOT included in B. cepacia complex)
 Alcaligenes (Achromobacter) xylosoxidans
 Pandoria
 Other, Specify _____

6.7 Encounter: Laboratory

Information of Patient :

Patient Name _____

Birth Date _____

Patient ID _____

Encounter Date _____

Encounter Type Annual Review encounter Other encounter

This encounter is part of A Clinical Visit

A Hospitalisation Care Episode

A Home IV Care Episode

PATIENT STATUS:

At the time of clinical visit, the patient was stable not stable

HEIGHT: _____ cm. in. Not Measured

WEIGHT: _____ kg. lb. Not Measured

SERUM CREATININE Level _____ mmol/dl Not Measured

LIVER ENZYMES DRAWN Yes No Unknown

CFRD SCREENING

Was this patient screened for CFRD? Yes No No-patient has known CFRD Unknown

random blood glucoseX _____ mmol/litre

fasting blood glucose _____ mmol/litre

If OGTT performed

fasting blood glucose _____ mmol/litre

2 hour _____ mmol/litre

6.8 Encounter: Complications

Information of Patient :

Patient Name _____

Birth Date _____

Patient ID _____

Encounter Date _____

Encounter Type Annual Review encounter Other encounter

This encounter is part of A Clinical Visit

A Hospitalisation Care Episode

A Home IV Care Episode

PATIENT STATUS:

At the time of clinical visit, the patient was stable not stable

HEIGHT: _____ cm. in. Not Measured

WEIGHT: _____ kg. lb. Not Measured

- | | |
|--|---|
| <input type="checkbox"/> Allergic Bronchial Pulmonary Aspergillosis (ABPA) | <input type="checkbox"/> Absence of Vas Deferens |
| <input type="checkbox"/> Arthritis | <input type="checkbox"/> Arthropathy |
| <input type="checkbox"/> Asthma | <input type="checkbox"/> Atyp. Mycobact. Inf. (req. Rx) |
| <input type="checkbox"/> Bone fracture | <input type="checkbox"/> Cancer confirmed by histology |
| <input type="checkbox"/> CFRD | <input type="checkbox"/> Chronic Pseudomonas aeruginosa |
| <input type="checkbox"/> Chronic Staph Aureus | <input type="checkbox"/> Cirrhosis with portal hypertension |
| <input type="checkbox"/> Cirrhosis with no portal hypertension | <input type="checkbox"/> Depression |
| <input type="checkbox"/> Dist Int Obst Synd (DIOS) | <input type="checkbox"/> Fib colonopathy/colonic strict.(report incidence only) |
| <input type="checkbox"/> Gallbladder Dis req surgery | <input type="checkbox"/> GERD (Gastro-Esoph. Ref. Dis.) |
| <input type="checkbox"/> GI Bleed req hosp non variceal | <input type="checkbox"/> GI Bleed req hosp variceal |
| <input type="checkbox"/> Hearing loss | <input type="checkbox"/> Hemoptysis, massive |
| <input type="checkbox"/> Hypertension | <input type="checkbox"/> Kidney Stones |
| <input type="checkbox"/> Liver disease, non-cirrhosis | <input type="checkbox"/> Liver enzymes elevated |
| <input type="checkbox"/> Nasal polyps req surgery | <input type="checkbox"/> NTM |
| <input type="checkbox"/> Osteopenia | <input type="checkbox"/> Osteoporosis |
| <input type="checkbox"/> Pancreatitis | <input type="checkbox"/> Peptic ulcer disease |
| <input type="checkbox"/> Pneumothorax req chest tube | <input type="checkbox"/> Port inserted or replaced |
| <input type="checkbox"/> Rectal prolapse | <input type="checkbox"/> Renal failure req dialysis |
| <input type="checkbox"/> Sinus Disease (symptomatic) | <input type="checkbox"/> Other (specify) _____ |