Cystic Fibrosis strength in numbers

UK Cystic Fibrosis Registry

2014 Annual Data Report – Scotland

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Contents

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UK Cystic Fibrosis Registry 2014 Annual Data Report - Scotland

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

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Introduction

This report is aimed at anyone who is interested in the health, care, and outcomes of people with cystic fibrosis (CF) in the Scotland. This includes people with CF, their families and clinical teams, healthcare managers, commissioners, and policymakers.

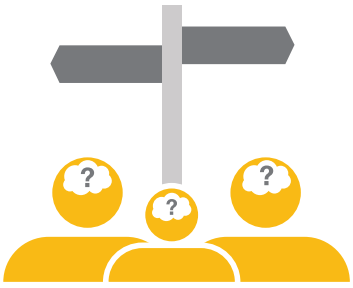
Cystic Fibrosis

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

UK Cystic Fibrosis Registry

The UK CF Registry has been sponsored and hosted by the Cystic Fibrosis Trust since 2007. It is a database of consenting people with CF in the UK. The Registry collects demographic, treatment, and health outcomes data. You can find a full list of the data items we collect at www.cysticfibrosis.org.uk/registry.

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

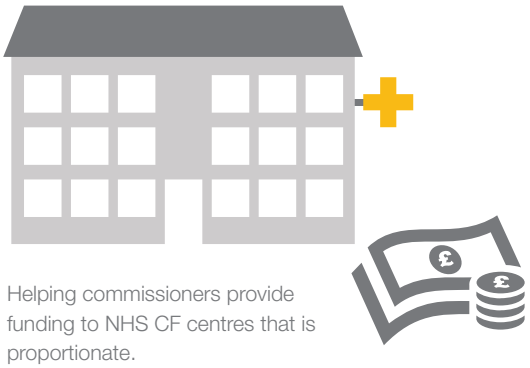


Helping people with CF and their families understand cystic fibrosis, and make informed decisions.

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

Monitoring the safety and effectiveness of new treatments for cystic fibrosis.

Providing data for research to find out the best ways of treating, and beating cystic fibrosis.



Helping commissioners provide funding to NHS CF centres that is proportionate.

Governance

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

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

When data are provided to third parties, such as the NHS or university researchers, they are either anonymised (all identifiable data removed completely) or pseudonymised (all identifiable data replaced with a unique identification number). Pseudonymisation is used so that data can be traced back to what is in the 'live' database for the purposes of updating the data or answering queries.

Anonymisation and pseudonymisation of the data means that the Registry data used for research, and the results that come from it, cannot identify the people whose data are stored on the UK CF Registry.

Data collection

Data are entered onto the UK CF Registry by NHS employees at cystic fibrosis centres in the UK, using a secure web portal.

Where can I find more information?

You can find out more about cystic fibrosis, and the UK CF Registry, at www.cysticfibrosis.org.uk/registry.

Words in this report that appear in the glossary are highlighted the first time they appear and explained in appendix 2 on page **47**.

Section 1: All Patients in Scotland

This section provides an overview of the cystic fibrosis population, health and care in Scotland.

1.1 Summary of the UK Cystic Fibrosis Registry 2014

	UK	Scotland
CF patients registered[1] Excluding 2014 diagnoses	10583 10356	970 962
CF patients with "complete" data[2]**; n(%)** Rate of completeness excluding 2014 diagnoses	9432 (89%) 91%	782(81%) 81%
Age in years; median[3]	19	20
All newly diagnosed patients (newborn screening and other)[4]	227	8
Newly diagnosed patients identified through newborn screening[4]	130	5
Age at diagnosis in months; median[3]	2	2
Adults aged 16 yrs and over; %	59.3	62
Males; %	53	53.3
Genotyped; %[3]	97.7	98.3
Total deaths reported[5]	137 (1.3%)	15 (1.5%)
Age at death in years; median (95%CI) [3]	28 (25.5,32.0)	26 (20.4,33.5)

Notes:

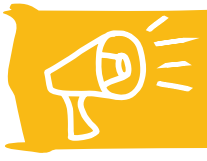
[1] This is calculated as the number of patients on the database who were diagnosed with CF and had not died before 1st January in the given year.

[2] A patient has "Complete data" if their team has filled in an annual review for that year. Patients newly diagnosed in 2014 may not have their first annual review in the same year. If newly diagnosed patients are excluded, 81% of records are complete.

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

[4] Calculated for all patients registered. Some diagnosis data are added after the data entry closure each year, so the figures from previous years have been updated for this report.

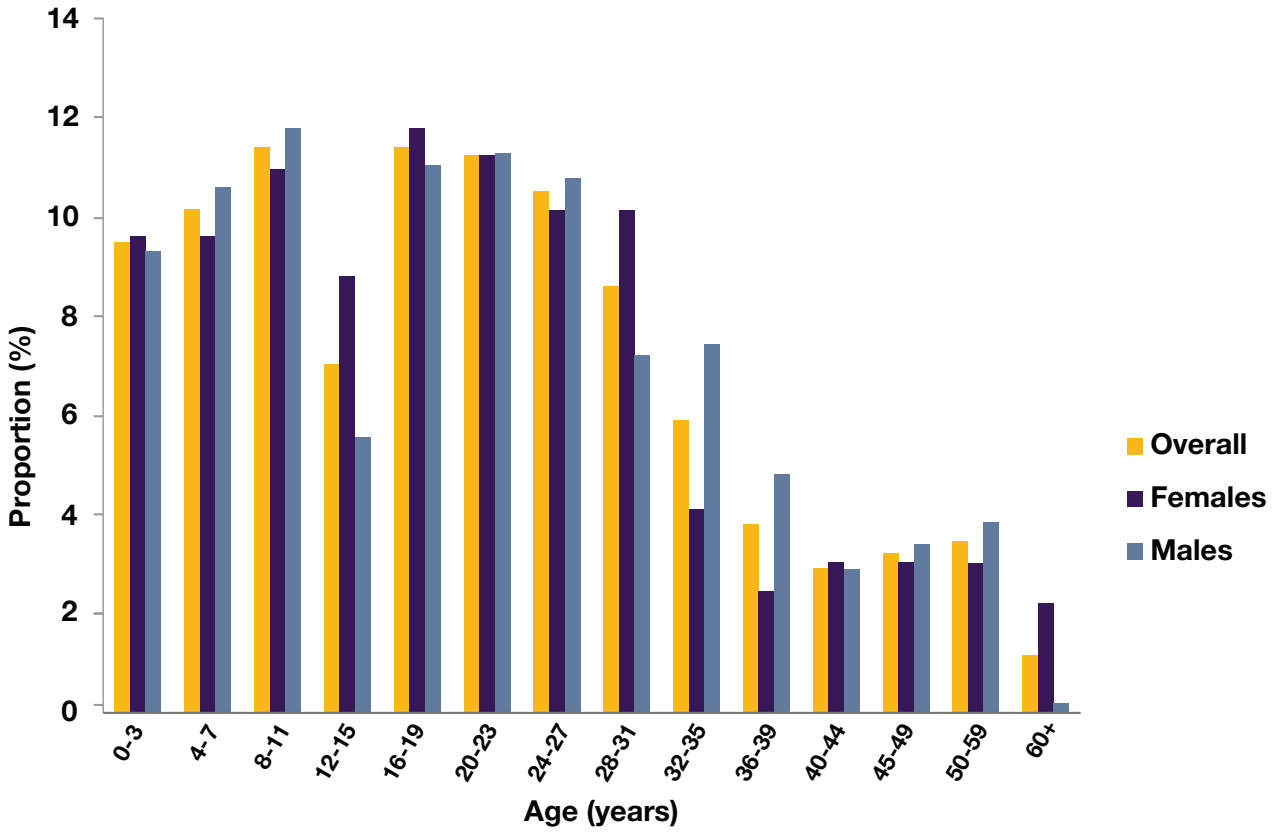
[5] Calculated for all patients registered.



Complete data: Patients with at least the minimum data entered at their annual review for analysis to be carried out.

1.2 Age distribution by gender n=782

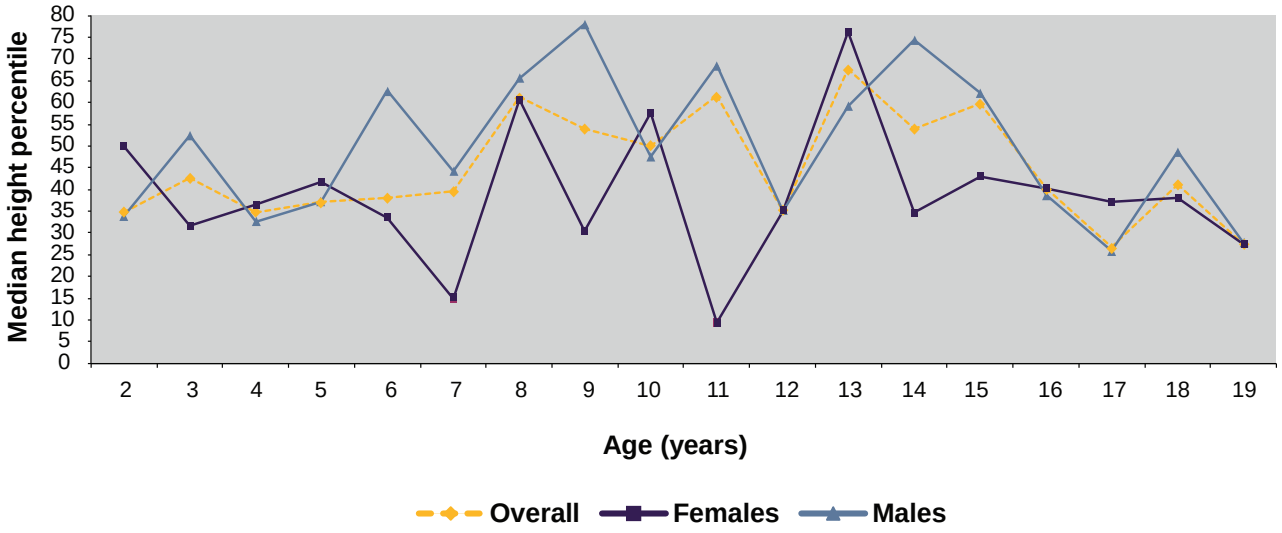
The following chart shows the mix of ages and genders in the cystic fibrosis population in Scotland.



1.3 Median height percentiles among children and young persons (<20 years)

n=349

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

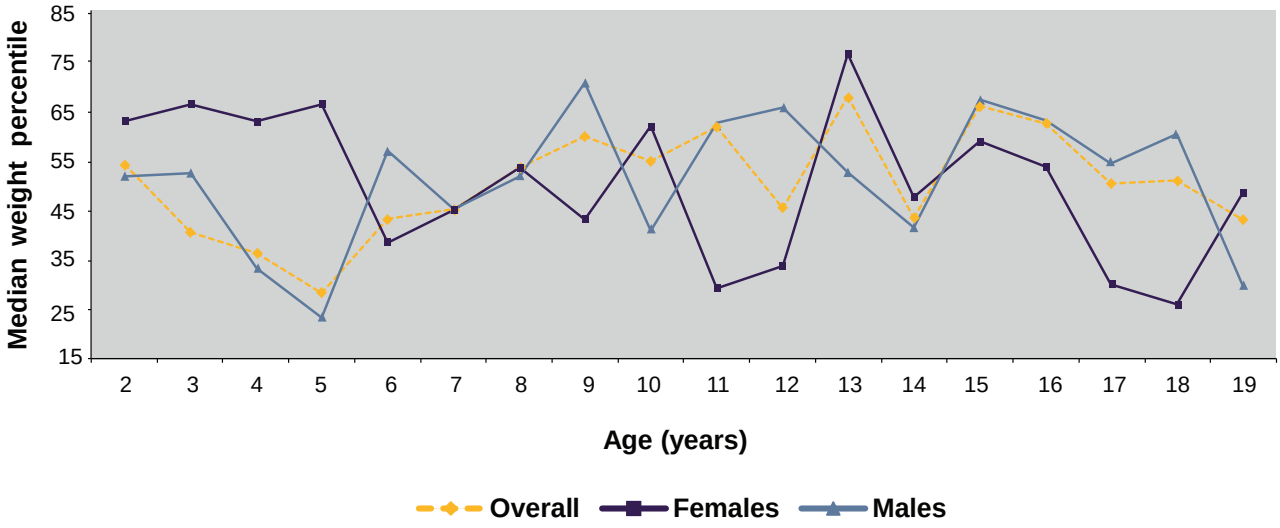


	Overall			Female			Male		
Age	N	Median	IQR	N	Median	IQR	N	Median	IQR
2	18	34.7	14.1-64.4	7	49.9	14.7-70.5	11	33.7	12.3-55.6
3	24	42.7	11.8-67.1	11	31.8	12.7-57.5	13	52.5	8.3-74.4
4	21	34.2	26.5-48.8	7	36.4	8.6-52.6	14	32.6	27.8-43.7
5	18	37.1	6.2-74.1	8	41.7	10.8-77.2	10	37.1	3.4-61.9
6	19	38.1	27.2-72.6	10	33.5	22.2-55.5	9	62.8	32.7-80.3
7	20	39.7	13.7-65.1	9	14.8	13.3-61.7	11	44.3	23.5-66.2
8	25	61.4	17.6-77.6	11	60.6	17.4-78.8	14	65.4	17.2-77.1
9	21	53.8	30.5-79.6	10	30.5	2.3-54.0	11	78	38.1-86.4
10	25	49.9	19.1-72.3	14	57.6	16.5-74.2	11	47.6	21.5-62.4
11	17	61.7	32.3-79.8	5	9.3	2.9-66.5	12	68.5	52.3-81.1
12	16	35.1	26.3-70.5	6	35.1	30.3-88.9	10	35.1	22.6-69.8
13	12	67.7	37.9-84.5	11	76.3	37.4-85.6	<5	59.1	59.1-59.1
14	17	54	30.1-82.1	9	34.7	12.9-71.5	8	74.5	32.5-82.9
15	10	59.6	15.2-76.8	6	42.9	15.2-78.0	<5	62.1	19.9-81.6
16	19	40.3	27.0-77.1	7	40.3	11.1-77.1	12	38.8	30.2-83.7
17	19	26.7	15.6-59.3	6	37.1	8.2-52.3	13	25.9	15.7-64.1
18	24	41.2	11.5-67.7	10	38	9.7-71.0	14	48.7	13.0-66.5
19	24	27.4	10.3-80.1	18	27.4	9.6-81.8	6	27.8	12.8-65.0
Overall	349	40.3	17.9-71.8	165	36.2	14.8-71.8	184	47.5	24.1-71.8

1.4 Median weight percentiles among children and young persons (<20 years)

n=350

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

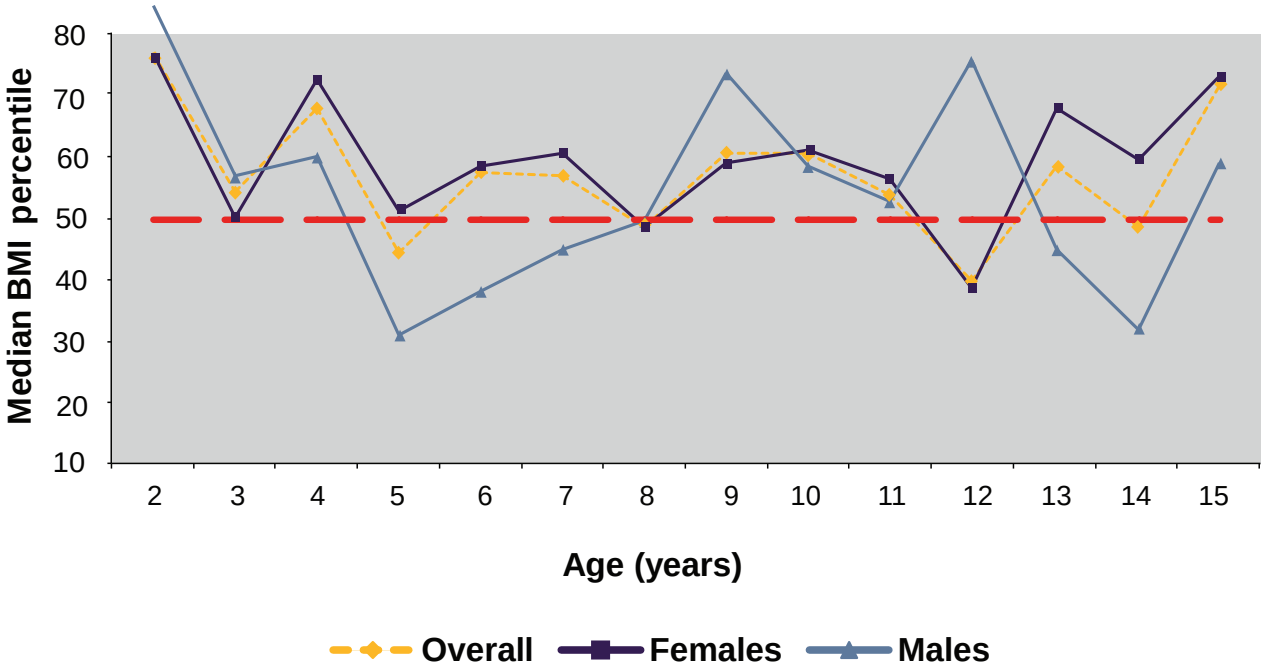


	Overall			Female			Male		
Age	N	Median	IQR	N	Median	IQR	N	Median	IQR
2	19	54.4	40.4-76.3	7	63.2	11.6-87.8	11	51.9	22.4-76.3
3	24	40.7	8.3-83.9	11	66.5	12.6-81.3	13	52.6	8.6-89.1
4	21	36.4	19.0-75.4	7	63.2	11.6-87.8	14	33.3	22.0-56.2
5	18	28.3	9.6-73.2	8	66.5	12.6-81.3	10	23.4	3.4-63.6
6	19	43.4	30.5-73.1	10	38.5	28.5-76.8	9	57.2	25.1-73.8
7	20	45.3	17.4-67.8	9	45.2	24.2-73.8	11	45.4	16.4-68.9
8	25	53.7	25.7-74.1	11	53.7	22.0-73.7	14	52	25.6-76.3
9	21	60.1	38.2-77.3	10	43.3	15.8-62.7	11	70.9	45.5-86.0
10	24	55	28.3-80.6	14	62.2	27.2-82.8	10	40.9	26.1-79.4
11	18	62	27.9-71.0	5	29.3	12.4-68.1	13	62.8	47.0-72.4
12	15	45.5	27.5-93.4	6	33.9	29.2-94.0	9	65.9	22.6-86.0
13	12	68.2	38.4-79.7	11	76.7	33.7-79.8	<5	52.7	52.7-52.7
14	17	43.5	27.1-75.2	9	47.8	23.3-80.9	8	41.6	29.7-59.4
15	10	66.2	32.7-77.0	6	59.1	30.9-77.0	<5	67.5	23.2-84.5
16	19	62.6	20.1-83.4	7	53.9	19.5-96.6	12	63.3	25.3-81.5
17	19	50.5	9.7-72.4	6	30.1	8.2-57.4	13	54.5	12.4-73.2
18	25	51.1	22.6-72.8	10	26.1	16.6-87.2	14	60.5	27.1-72.4
19	24	43.3	11.3-77.9	18	48.5	13.8-79.9	6	30	1.6-53.1
Overall	350	52	23.2-74.2	165	47.8	21.6-74.4	183	48.9	22.4-73.9

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

n=345

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

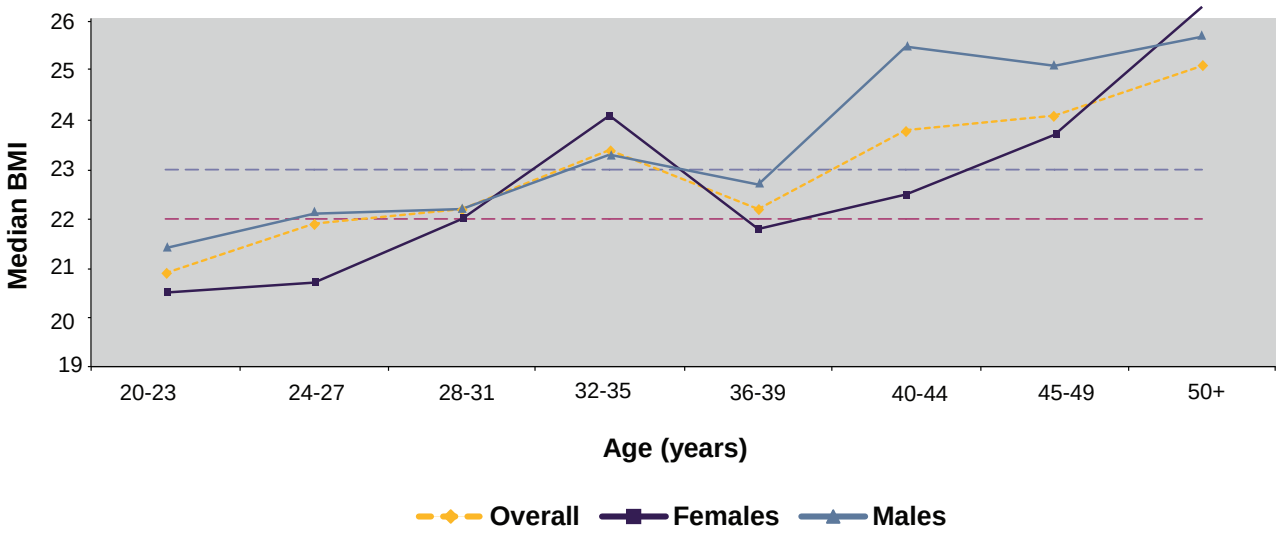


	Overall			Female			Male		
Age	N	Median	IQR	N	Median	IQR	N	Median	IQR
2	18	77.3	46.9-87.9	7	77.1	56.8-87.4	11	84.9	29.5-93.0
3	24	54.8	21.6-83.3	11	50.4	16.2-81.0	13	57.1	28.6-85.6
4	21	68.1	35.8-84.6	7	73.2	42.1-91.7	14	60.3	26.7-82.7
5	18	44.7	12.2-73.2	8	51.6	21.4-73.8	10	31.1	0.7-72.4
6	19	57.9	31.2-73.9	10	58.8	42.4-75.4	9	38.1	23.3-74.1
7	19	57.1	27.5-71.0	9	61.1	36.6-83.9	10	44.9	22.5-70.4
8	25	48.7	29.2-73.7	11	48.7	29.5-72.4	14	49.8	27.9-76.5
9	21	60.8	40.3-79.1	10	59.5	39.2-81.7	11	73.9	38.8-79.2
10	24	60.8	18.7-81.0	14	61.5	23.2-82.9	10	58.7	11.9-82.3
11	17	53.8	43.0-62.4	5	56.5	32.2-63.7	12	53.2	40.4-63.1
12	15	40	27.8-91.9	6	38.8	30.4-84.1	9	76	25.3-95.1
13	12	58.9	39.0-84.6	11	68.2	37.2-89.2	<5	44.9	44.9-44.9
14	16	48.8	31.4-74.6	9	59.9	43.1-79.4	7	32.2	26.5-52.1
15	10	72	27.0-80.6	6	73.5	22.9-80.6	<5	59.4	26.7-86.4
16	19	72.6	28.9-90.7	7	76.9	28.9-98.0	12	71	34.9-90.2
17	19	51.5	27.9-75.5	6	54	24.2-64.2	13	51.5	23.3-81.5
18	24	64	47.8-82.7	10	60.4	24.9-78.6	14	65.9	53.1-84.5
19	24	54.1	8.8-83.1	18	69.5	11.8-84.9	6	24.6	5.5-56.3
Overall	345	57.6	29.7-79.2	165	59.7	33.7-78.8	180	55.3	27.9-79.8

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

n=383

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



	Overall			Female			Male		
Age	N	Median	IQR	N	Median	IQR	N	Median	IQR
20-23	86	20.9	18.8-23.3	40	20.5	18.6-22.3	46	21.4	18.8-24.0
24-27	78	21.9	19.9-23.9	35	20.7	19.6-24.6	43	22.1	20.5-23.7
28-31	65	22.2	20.0-24.7	35	22	20.0-25.2	30	22.2	20.0-24.2
32-35	42	23.4	21.5-25.4	14	24.1	21.7-25.8	28	23.3	21.1-25.4
36-39	29	22.2	20.6-25.7	9	21.8	20.1-24.6	20	22.7	21.6-26.4
40-44	23	23.8	22.1-26.1	11	22.5	21.6-25.0	12	25.5	22.3-29.3
45-49	25	24.1	21.6-27.9	11	23.7	21.4-27.1	14	25.1	21.6-30.4
50+	35	25.1	22.2-27.5	18	26.3	22.1-28.4	17	24.3	21.8-27.1
Overall	**383**	**22.3**	**20.3-25.1**	**173**	**22.1**	**20.0-24.9**	**210**	**22.5**	**20.6-25.4**

1.7 Education and employment in adults aged 16 years and over

n = 506

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

Patient reported status	Number of patients
Full-time working	164 (32.4%)
Part-time working	73 (14.4%)
Student	95 (18.8%)
Homemaker	28 (5.5%)
Unemployed	93 (18.4%)
“Disabled”	12 (2.4%)
Retired	10 (2%)
Unknown	20 (4%)
No data	<5

Of the 485 adults aged 16 years and older for whom an employment status questionnaire was completed (excluding “unknown”), 321 (69.2%) reported being in work or study.

Diagnosis of cystic fibrosis

1.8 Age at Diagnosis in children under 16

n= 289*

Newborn screening for CF has been done routinely in the whole of the UK since July 2007. It is part of the heel prick blood spot testing done at five to seven days of age. The blood sample is tested for a number of conditions, including cystic fibrosis. This means that more babies born after 2007 receive an early diagnosis than those born before.

Age at diagnosis	All patients; n (%)	Patients aged 10 years in 2014; n (%)	Patients aged 5 years in 2014; n (%)
Pre-natal	<5	-	-
Birth-3months	237 (72.0)	22 (88.0)	9 (100)
4-6 months	9 (3.1)	<5	-
7-12 months	5 (1.7)	<5	-
1 yr	8 (2.8)	-	-
2	8 (2.8)	-	-
3	6 (2.1)	-	-
4	7 (2.4)	-	-
5	<5	-	-
6	<5	-	-
7	<5	-	-
8	<5	-	-
9	<5	-	-
10	-	-	-
11	-	-	-
12	-	-	-
13	-	-	-
14	-	-	-
15	-	-	-
Overall	**289* (37.4)**	**25**	**19**

*Note: 8 children with missing data

The median (range) age at diagnosis is 89 days (61 -120 days).

The median (range) age at diagnosis for patients aged under 16 in 2014 is 22 days (9-56)

Diagnosis in the first three months of life was more common in children aged 5 years in 2014 (born after the UK-wide newborn screening programme was in place) than in children aged 10 years in 2014 (born before the UK-wide newborn screening programme was in place).

For the 2 children with complete data born in 2014, none were identified by newborn screening. In 2014, a total of 4 patients were identified by newborn screening (including patients with and without complete data). As there is a delay between newborn screening tests being performed and the results entering the Registry, these results are updated retrospectively each year to take updated data into account. In 2013 this figure was 8 and in 2012 it was 12.

1.9 Age at Diagnosis in adults aged 16 years and over in 2014

n=485

Age at diagnosis	n (%)
Pre-natal	0
Birth-3months	185 (36.2)
4-6 months	48 (9.9)
7-12 months	27 (5.6)
1 yr	42 (8.7)
2 yrs	30 (6.2)
3 yrs	21 (4.35)
4 yrs	13 (2.7)
5 yrs	9 (1.9)
6 yrs	8 (1.7)
7 yrs	6 (1.2)
8 yrs	<5
9 yrs	5 (1.0)
10 yrs	<5
11 yrs	6 (1.2)
12 yrs	5 (1.0)
13 yrs	<5)
14 yrs	5 (1.0)
15 yrs	5 (1.0)
16-20 yrs	16 (3.3)
21-25 yrs	6 (1.2)
26-30 yrs	17 (3.5)
31-35 yrs	10 (2.1)
36-40 yrs	7 (1.5)
41-45 yrs	<5
46-50 yrs	<5
51 yrs	9 (1.9)
Overall	**485 (62.0)**

Lung Health

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

In CF the condition of the lungs is often measured using FEV_1; the forced expiratory volume of air in the first second of an exhaled breath. In this report, FEV_1% predicted is based on the FEV_1 we would expect for a person without cystic fibrosis of the same age, gender, height, and ethnicity.

A person with CF who has FEV_1% predicted of 100 can breathe out the same amount of air in the first second of an exhaled breath as we would expect from a comparable person without cystic fibrosis.

A person with FEV_1% predicted of 50 breathes out half the volume of air as a comparable person without cystic fibrosis.

For people with CF, an FEV_1% predicted of 85 or higher is the target, as this indicates normal or near-normal lung health.

Most people can continue to lead a relatively normal life, including going to school or work, with 50% of their predicted FEV_1. Once FEV_1 is lower than 50% of the predicted value, it becomes more difficult to lead a normal life. If FEV_1 declines to 30% or less, a patient may be considered for lung transplant.

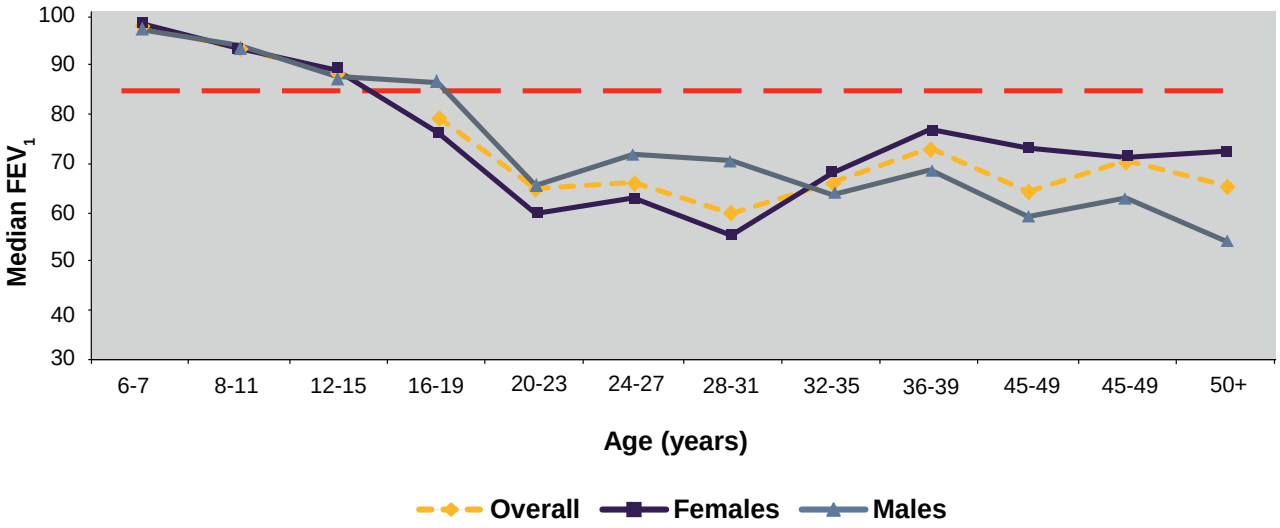
An aim of CF care is to prevent FEV_1% predicted from falling as much as possible, for as long as possible. This is often a team effort between people with CF, their family, and their medical team, which can include doctors, nurses, physiotherapists, dieticians, and psychologists.

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

1.10 Median FEV_1 (% predicted) among patients aged 6 years and over, excluding patients post lung transplant

n=636

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

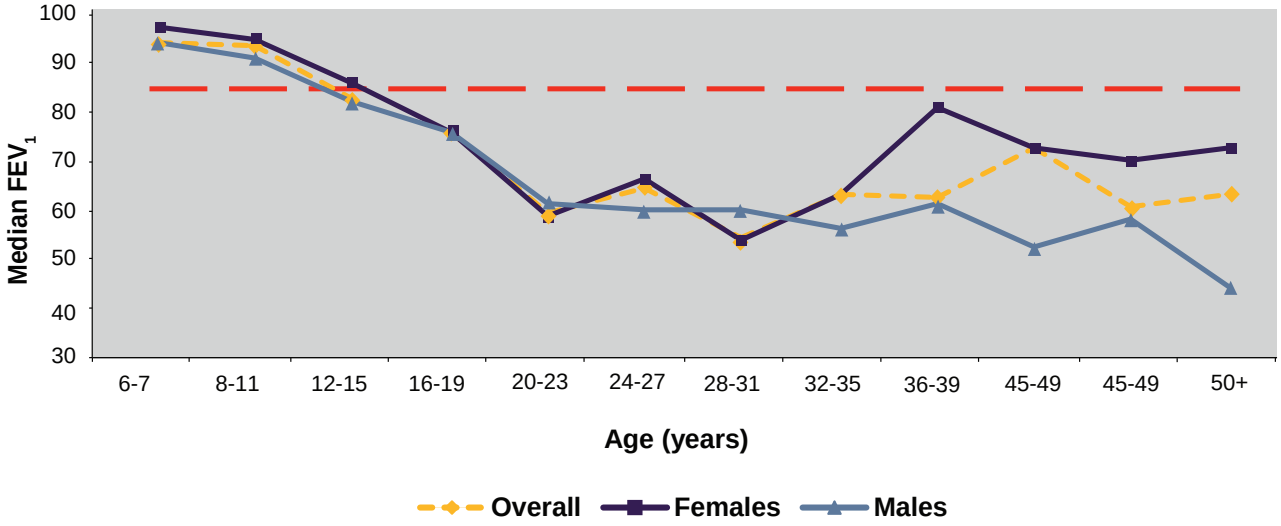


	Overall			Female			Male		
Age	N	Median	IQR	N	Median	IQR	N	Median	IQR
6 to 7	38	97.7	83.4-108.1	17	98.5	86.0-110.7	21	97	78.0-105.0
8 to 11	84	93.2	85.9-102.7	38	93.2	86.3-98.9	46	93.8	85.2-105.8
12 to 15	54	88	76.1-99.9	27	89.1	72.3-102.0	27	87.5	79.3-99.0
16 to 19	89	79.7	56.0-98.2	45	76.3	51.8-97.7	44	86.2	60.5-100.0
20 to 23	86	64.7	47.9-83.1	37	59.7	47.1-78.2	49	65.7	48.3-88.0
24 to 27	75	65.8	44.3-87.6	40	62.8	42.5-88.8	35	72.1	44.3-85.0
28 to 31	58	59.5	41.5-83.2	28	55	36.8-74.5	30	70.2	46.2-91.9
32 to 35	45	65.8	39.4-93.8	21	68.2	48.7-88.6	24	63.3	38.4-98.1
36 to 39	26	72.9	52.3-91.0	16	76.7	56.7-98.7	10	68.4	42.2-79.4
40 to 44	23	64.3	47.9-78.8	14	73.3	50.8-79.8	9	58.9	43.9-79.6
45 to 49	25	70.5	52.2-78.2	12	71	56.4-78.5	13	63.1	40.6-85.2
50+	33	65.7	44.4-88.4	17	72.2	53.4-94.1	16	54.1	31.8-77.6
Overall	**636**	**78.2**	**54.0-94.6**	**312**	**76.1**	**53.2-93.4**	**324**	**80.3**	**54.9-96.4**

1.11 Median FEV_1 (% predicted, GLI equations) among patients aged 6 years and older, excluding patients post lung transplant

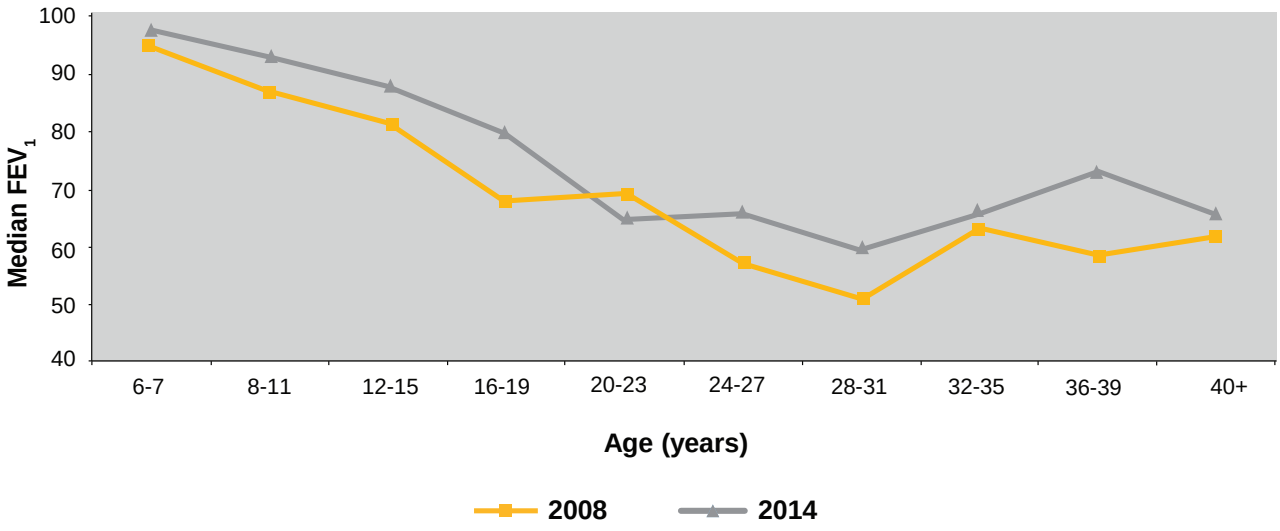
n=602

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



	Overall			Female			Male		
Age	N	Median	IQR	N	Median	IQR	N	Median	IQR
6 to 7	36	94.1	83.3-104.8	15	97.5	84.1-108.2	21	93.9	77.9-104.4
8 to 11	76	93.1	86.6-101.5	35	94.8	90.8-101.8	41	90.8	85.3-100.9
12 to 15	53	82.5	73.0-95.9	26	86	68.8-96.8	27	82.1	75.2-93.6
16 to 19	85	75.6	52.3-92.4	43	75.6	51.9-99.0	42	75.7	52.2-92.0
20 to 23	84	59.4	46.1-78.5	37	58.6	50.2-81.4	47	60.8	41.2-75.1
24 to 27	71	64.3	47.0-77.8	39	66	48.7-85.1	32	59.7	37.3-72.3
28 to 31	53	53.8	39.4-73.6	26	53.5	37.1-72.3	27	59.6	39.9-74.6
32 to 35	41	62.6	40.2-85.5	19	63	51.8-85.3	22	55.6	34.0-85.8
36 to 39	24	72.3	43.4-88.7	15	80.9	52.0-107.5	9	61.2	39.9-72.3
40 to 44	23	62.3	43.9-87.2	14	72.3	47.5-89.1	9	51.9	38.0-73.4
45 to 49	25	60.2	49.3-78.7	12	70.1	53.0-83.9	13	57.9	35.3-74.6
50+	31	63.2	34.2-83.4	15	72.2	51.5-88.4	16	43.9	29.6-72.5
Overall	**602**	**74**	**51.7-90.8**	**296**	**90.4**	**73.9-103.4**	**306**	**85.2**	**67.2-97.5**

1.11a Median FEV_1 among patients aged 6 years and older by year in 2008 (n=420) and 2014 (n=636) (excluding patients post lung transplant)

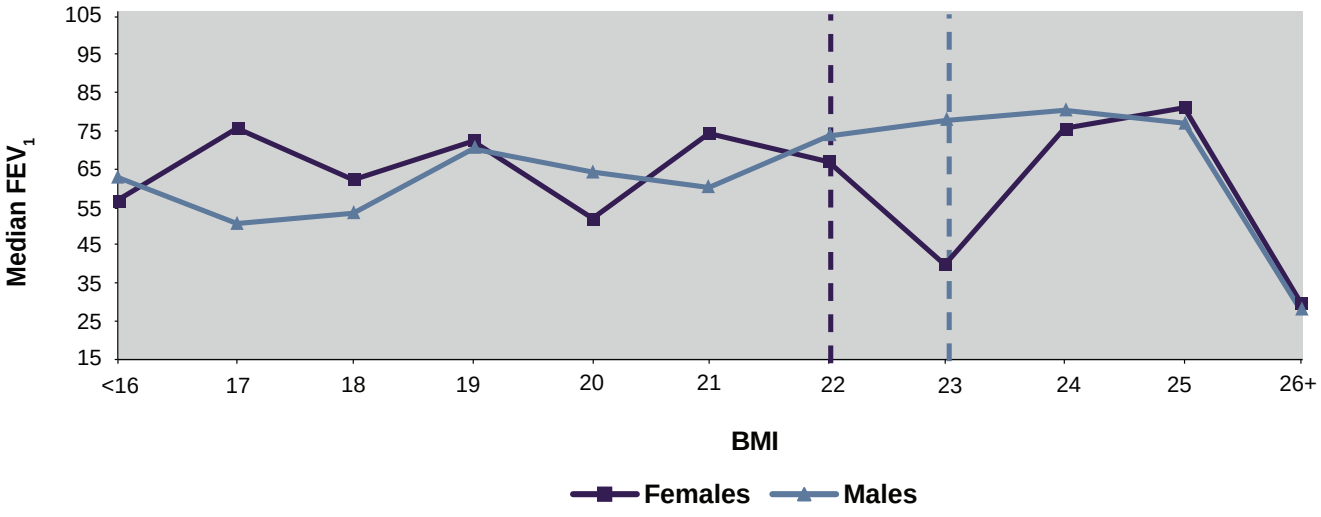


An analysis was conducted in order to determine whether there were statistically significant differences in FEV_1 (% predicted) in 2014 compared to 2008 by age category.

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

n=461

The goal BMI for adults is 22 for women and 23 for men. The chart shows the relationship between BMI and FEV_1% predicted. A healthy BMI can protect people with CF against lung infection, and help to preserve lung health. People with CF who have had lung transplants are excluded, as their new 'non-CF' lungs would have lung health similar to a person without cystic fibrosis.



Each point represents the median FEV_1 % predicted of patients for each given BMI value. Due to the wide range of BMIs in this population we grouped all BMI≤16 into one group and BMI≥26 into another.

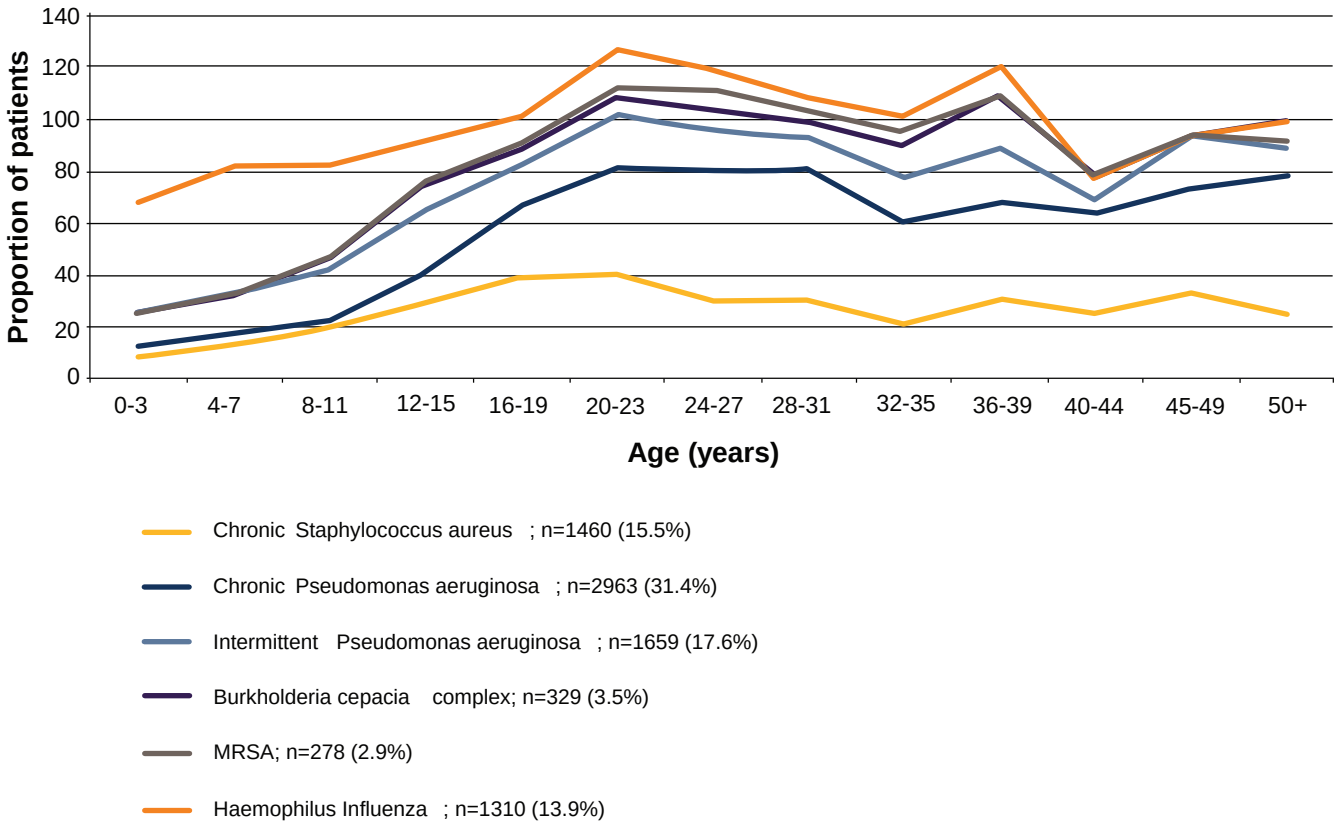
Lung infections

Lung infections can permanently reduce lung function in people with cystic fibrosis. Some lung infections can become 'chronic', meaning that they can't ever be removed completely using medicines.

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

1.13 Lung infections in 2014

n=782



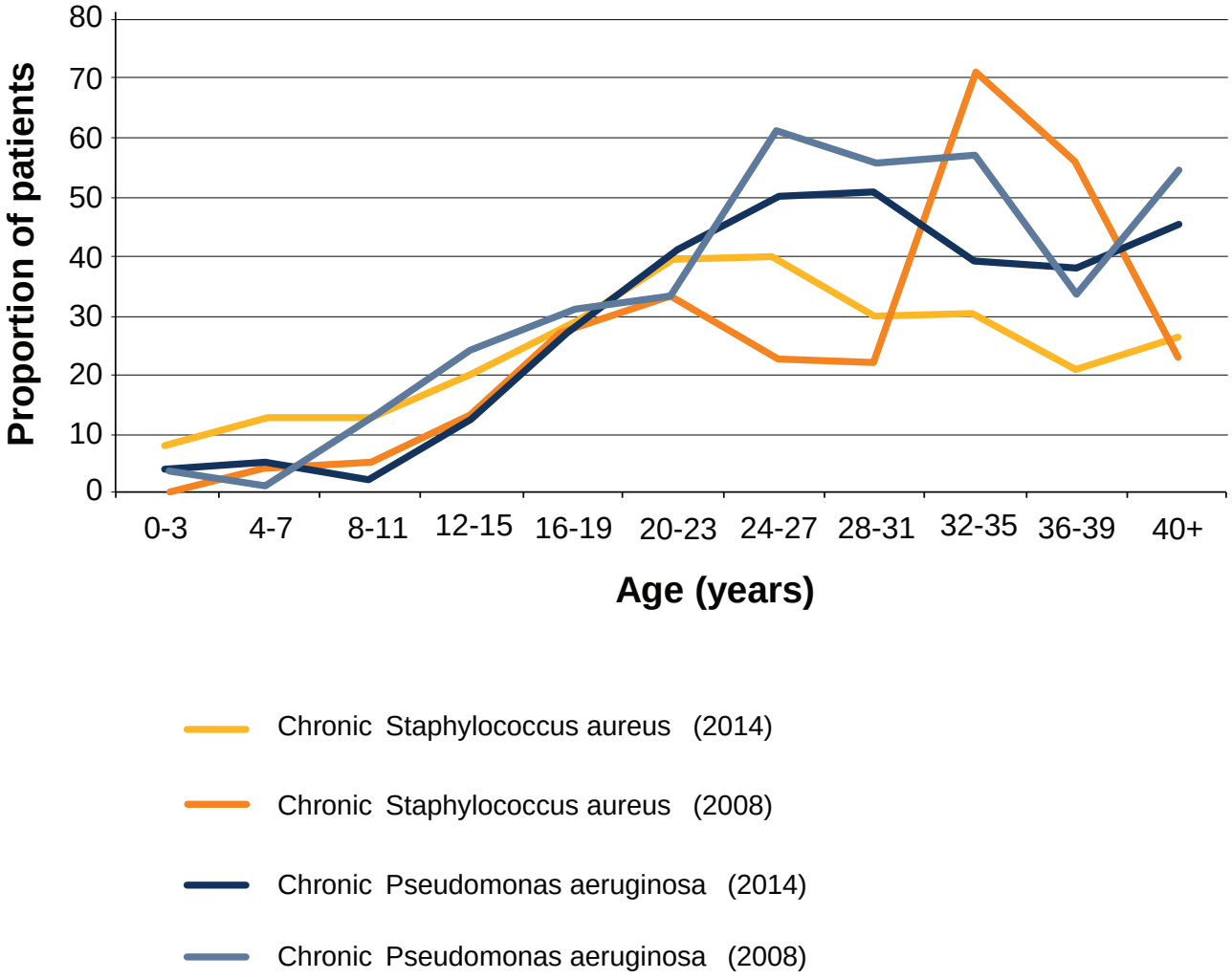
Chronic infection with S. aureus or P. aeruginosa were identified from annual review. Data on B.cepacia, MRSA and H.influenzae were collected from culture results at annual review.

	Age (years)													Overall		
	0-3	4-7	8-11	12-15	16-19	20-23	24-27	28-31	32-35	36-39	40-44	45-49	50+	All	Children (<16 years)	Adults (≥16 years)
N patients in age band	74	79	89	55	89	88	82	67	46	29	23	25	36	782	297	485
Chronic S.aureus; n(%)	6 -8.1	10 -12.8	11 -12.8	11 -20	25 -28.7	33 -39.3	33 -40.2	20 -29.9	14 -30.4	6 -20.7	7 -30.4	6 -25	9 -25	191 -24.8	38 -13	153 -32
Chronic P.aeruginosa; n(%)	<5	<5	<5	7 -12.7	24 -27.6	36 -40.9	41 -50	34 -50.8	18 -39.1	11 -37.9	9 -39.1	10 -40	19 -52.8	218 -28.1	16 -5.5	202 -41.8
Intermittent S.auerus; n(%)	16 -21.6	15 -19.2	20 -23.3	17 -30.9	16 -18.4	14 -16.7	15 -18.3	11 -16.4	6 -13	8 -27.6	<5	<5	<5	149 -19.3	68 -23.2	81 -17
Intermittent P.aeruginosa; n(%)	9 -12.3	11 -14.1	17 -19.8	13 -23.6	13 -14.9	18 -20.5	13 -15.9	8 -11.9	8 -17.4	6 -20.7	<5	5 -20	<5	126 -16.3	50 -17.1	76 -15.7
B.cepacia; n(%)	0	0	<5 -4.5	5 -9.1	6 -6.7	6 -6.8	7 -8.5	<5 -6	6 -13	6 -20.7	<5	0	<5	47 -6	9 -3	38 -7.8
MRSA; n(%)	0	<5	<5	<5	<5	<5	5 -6.2	<5	<5	0	0	0	0	18 -2.3	<5	15 -3.1
H.influenza; n(%)	32 -43.2	38 -48.1	31 (34.8)	9 -16.4	9 -10.1	13 -14.8	7 -8.5	<5	<5	<5	0	0	<5	151 -19.3	110 -37	41 -8.5

1.14 Lung infection over time

2008 n=429 and 2014

n=782



Complications

1.15 Prevalence of key complications

	Overall (n=782)	<16 years (n=297)	≥16 years (n=485)
	N(%)	N(%)	N(%)
Respiratory Related			
Asthma; n(%)	107 (13.7)	22 (7.4)	85 (17.5)
Sinus disease; n(%)	180 (23.0)	<5	177 (36.5)
Nasal polyps requiring surgery; n(%)	13 (1.8)	<5	11 (2.3)
Nontuberculous mycobacteria or atypical mycobacteria; n(%)	39 (5.0)	15 (5.1)	24 (5.0)
Pneumothorax requiring chest tube; n(%)	<5	0	<5
ABPA; n(%)	57 (7.3)	13 (4.4)	44 (9.1)
Hemoptysis; n(%)	13 (1.7)	2 (0.7)	11 (2.3)
Pancreas and Hepatobiliary Disease			
Cirrhosis with no portal hypertension; n(%)	6 (0.77)	<5	5 (1.0)
Cirrhosis with portal hypertension; n(%)	17 (2.2)	5 (1.7)	12 (2.5)
Liver disease; n(%)	131 (16.8)	42 (14.1)	89 (18.4)
Liver enzymes; n(%)	19 (2.4)	11 (3.7)	8 (1.6)
Gall bladder disease requiring surgery; n(%)	<5	0	<5
Pancreatitis; n(%)	8 (10.2)	<5	7 (1.4)
Gastrointestinal			
Intestinal obstruction; n(%)	91 (11.6)	7 (2.4)	84 (17.3)
GERD; n(%)	117 (15.0)	<5	113 (23.3)
Fibrosing colonopathy/ colonic stricture; n(%)	0	0	0
Rectal prolapse; n(%)	0	0	0
Renal			
Kidney stones; n(%)	<5	<5	<5
Renal failure; n(%)	<5	0	<5
Musculo-Skeletal			
Arthritis; n(%)	<5	0	<5
Arthropathy; n(%)	34 (4.3)	<5	33 (6.8)
Bone fracture; n(%)	<5	0	<5
Osteopenia; n(%)	82 (10.5)	0	82 (16.9)
Osteoporosis; n(%)	34 (4.3)	0	34 (7.0)
Other			
Cancer confirmed by histology; n(%)	<5	0	<5
Port inserted or replaced; n(%)	16 (2.0)	9 (3.0)	7 (1.4)
Absence of vas deferens; n(%)	10 (1.3)	0	10 (2.1)
Depression; n(%)	25 (3.2)	0	25 (5.6)
Hearing loss; n(%)	10 (1.3)	<5	8 (1.6)
Hypertension; n(%)	24 (3.1)	0	24 (4.9)
CFRD; n(%)	**132 (16.9)**	**6 (2.0)**	**126 (26.0)**

1.16 Incidence of key complications

	Newly identified in 2013			Newly identified in 2014		
	Overall (n=809)	<16 years (n=314)	≥16 years (n=495)	Overall (n=782)	<16 years (n=297)	≥16 years (n=485)
Nontuberculous mycobacteria or atypical mycobacteria; n(%)	<5	0	<5	<5	<5	<5
ABPA; n(%)	54 (6.7)	10 (3.2)	44 (0.9)	11 (1.4)	1 (0.3)	10 (2.1)
Cirrhosis with no portal hypertension; n(%)	<5	<5	<5	<5	0	<5
Cirrhosis with portal hypertension; n(%)	<5	0	<5	6 (0.8)	<5	5 (1.0)
Cancer confirmed by histology *; n(%)	0	0	0	<5	0	<5

1.17 CF-related diabetes

Cystic fibrosis-related diabetes (CFRD) is common in adults and adolescents with CF. This is because, for many people with CF, the pancreas does not work properly. This can mean that not enough insulin is produced, causing CFRD. CFRD is different from type 1 and type 2 diabetes, but has features of both.

	Overall* (n=782)	<16 years * (n=297)	≥16 years * (n=485)
Treatment for CF-related diabetes; n(%)	127 (16.2)	9 (3.0)	118 (24.3)
Screening for CF-related diabetes			
Yes	427 (55.1)	171 (56.4)	256 (52.9)
No	191 (24.7)	109 (37.5)	82 (17.0)
Known CF-related diabetes	145 (18.7)	6 (2.1)	139 (28.7)
Unknown	11 (1.4)	5 (1.7)	6 (1.2)

1.18 Transplants

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

The following table shows transplant activity over time.

	2010	2011	2012	2013	2014
Number of patients that year with annual review data evaluated for transplants	16	12	13	21	17
Number accepted on the transplant list	9	5	7	11	12
Number receiving transplants	<5	<5	5	<5	5
Types of transplants received:					
Bilateral lung	<5	<5	5	<5	5
Heart and lung	0	0	0	0	0
Liver	0	0	0	0	0
Other	0	0	0	0	0

1.19 Ivacaftor

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

Number of patients on Ivacaftor in Scotland	60
	Median (IQR)
Sweat chloride pre	106 (99-117)
Sweat chloride post	53 (38-68)
FEV_1% pre	59.1 (55.2-67.7)
FEV_1% post	73.8 (66.1-78.3)
Also number of patients stopped Ivacaftor	4

People with CF have a higher amount of chloride in their sweat than a person without cystic fibrosis. This measurement is called 'sweat chloride' and is measured in mmol/litre.

1.20 Intravenous (IV) antibiotic use

When someone with cystic fibrosis becomes unwell with an infection, they might be prescribed intravenous (IV) antibiotics which are given to the patient through their veins. This treatment can take a number of days and might take place in hospital or at home.

Age	Home				Hospital				Total			
	N	%	Median days	IQR	N	%	Median days	IQR	N	%	Median days	IQR
0-3	<5	1.4	49	49-49	16	21.6	14	12.3-23.5	16	21.6	15	13-28.5
4-7	6	7.6	39.5	17.5-59	26	32.9	14	10.8-57.3	26	32.9	39	14-61
8-11	11	12.4	39	10-53	27	30.3	15	12-45	28	31.5	45.5	14-59
12-15	8	14.6	39.5	23.5-51.8	28	50.9	26.5	14-42	28	50.9	42	18.3-64.5
16-19	22	24.7	20	13.5-56.8	20	22.5	14.5	9-33	28	31.5	28	14-65.3
20-23	29	33	22	14-42	27	30.7	14	7-42	40	45.5	28	14-55.3
24-27	31	37.8	28	14-65	24	29.3	16.5	9.3-27.8	39	47.6	35	14-77
28-31	22	32.8	37	14-66.5	17	25.4	16	6.5-27	27	40.3	42	17-70
32-35	9	19.6	14	14-42.5	<5	8.7	13.5	7-21.5	11	23.9	14	14-49
36-39	11	37.9	42	14-82	6	20.7	10	5.8-16.8	12	41.4	45.5	14-80.5
40-44	6	26.1	14	14-31.5	<5	13	14	8-28	8	34.8	14	14-38.5
45-49	9	36	14	9.5-28	<5	16	29.5	9-77.8	9	36	28	14-52
50+	9	25	15	13.5-33.5	9	25	10	3.5-13	12	33.3	17.5	14-38.5
Overall	**174**	**22.3**	**28**	**14-49.3**	**211**	**27**	**14**	**9-41**	**284**	**36.3**	**29**	**14-59**

Nebulised drug treatments

Nebulised drugs are medications that are breathed in as a mist. They are changed into a mist by a pot holding liquid medication, called a nebuliser.

Nebulised medications are used because:

- The medications go straight to where they need to work (in the lung) without having to go around the body. This can reduce side effects.
- Some medication is only available as a nebulised medication, for example, DNase.
- Large doses of medication can be given compared with some types of inhaler.
- It can be difficult to use some inhalers correctly. Using a nebuliser can mean that more of the medication gets into the lung.

1.21 Inhaled Antibiotic use among patients with chronic Pseudomonas aeruginosa

	2008			2014		
	Overall	<16 years	≥16 years	Overall	<16 years	≥16 years
Patients with chronic pseudomonas	67	28	39	218	16	202
Tobramycin solution; n(%)	6 (9.0)	<5	<5	43 (19.7)	<5	40 (19.8)
Other aminoglycoside; n(%)	0	0	0	7 (3.2)	0	7 (3.5)
Colistin; n(%)	21 (31.3)	15 (53.6)	6 (15.4)	74 (33.9)	11 (68.8)	63 (31.2)
Promixin; n(%)	<5	0	<5	29 (13.3)	<5	28 (13.9)
Aztreonam; n(%)*				18 (85.8)	10 (62.5)	177 (87.6)
Tobramycin Dry Powder; n(%)*				50 (22.9)	0	50 (24.8)
Colistimethate; n(%)*				22 (10.1)	<5	21 (10.4)
At least one of the above; n(%)**	-	-	-	**164 (75.2)**	**13 (81.3)**	**151 (74.8)**

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

1.22 DNase

	DNase; n(%)				
Age	2008		2014		
	Total patients	Patients on Dnase	Total patients	Patients on Dnase	p-value (2008 vs 2014)
0-3	91	3 (3.3)	74	5 (6.8)	0.652
4-7	78	9 (11.5)	79	18 (22.8)	0.378
8-11	66	15 (22.7)	89	31 (34.8)	0.232
12-15	92	33 (35.9)	55	28 (50.9)	<0.001
16-19	32	12 (37.5)	89	38 (42.7)	1
20-23	14	1 (7.1)	88	35 (39.8)	0.003
24-27	14	1 (7.1)	82	41 (50.0)	<0.001
28-31	11	2 (18.2)	67	21 (31.3)	0.116
32-35	8	1 (12.5)	46	8 (17.4)	0.58
36-39	9	1 (1.1)	29	9 (31.0)	0.479
40+	14	5 (35.7)	84	22 (26.2)	0.604
Overall	**429**	**83 (19.4)**	**782**	**256 (32.7)**	

1.23 Hypertonic saline

This treatment helps to thin mucus so that it is easier to cough out of the body

Age	Hypertonic saline; n(%)				
	2010	2011	2012	2013	2014
0-3	**0**	**0**	**<5**	**<5**	**<5**
4-7	0	<5	<5	6 (5.9)	9 (11.4)
8-11	<5	7 (12.1)	8 (13.6)	10 (12.2)	14 (15.7)
12-15	9 (10.7)	10 (15.2)	10 (15.2)	12 (19.7)	15 (27.3)
16-19	8 (9.0)	12 (12.2)	19 (19.8)	26 (26.3)	28 (31.5)
20-23	5 (7.7)	7 (8.3)	16 (15.8)	25 (25.8)	35 (39.8)
24-27	8 (13.3)	9 (13.0)	14 (19.4)	9 (12.0)	16 (18.2)
28-31	<5)	<5	6 (9.2)	7 (10.5)	15 (18.3)
32-35	<5 [1]	<5	6 (13.3)	5 (10.2)	14 (2.9)
36-39		<5	<5	0	<5
40-44		6 (8.1)	<5	<5	0
45-49			<5	<5	6 (24.0)
50+			<5)	<5	8 (22.2)
Overall	**40 (6.1)**	**59 (8.3)**	**95 (13.0)**	**110 (13.6)**	**134 (17.1)**

[1] In 2010 all patients aged 32 years and older were grouped together.

1.24 Long-term use of azithromycin among patients with and without chronic Pseudomonas aeruginosa

Azithromycin is an antibiotic with anti-inflammatory properties used to treat certain infections, including P.aeruginosa.

	2008				2014			
	Overall (n=429)	0-3 years (n=91)	4-15 years (n=236)	≥16 years (n=102)	Overall (n=782)	0-3 years (n=74)	4-15 years (n=223)	≥16 years (n=485)
Among patients with chronic P.aeruginosa	29 (43.3)	0	12 (46.2)	17 (43.6)	187 (85.8)	<5	9 (69.2)	177 (87.6)
Among patients without chronic P.aeruginosa	34 (12.4)	<5	22 (12.9)	11 (22.0)	216 (38.8)	<5	55 (26.7)	159 (56.6)

1.25 Physiotherapy

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

	Overall (n=782)	<16 years (n=297)	≥16 years (n=485)
Active cycle of breathing techniques; n(%)	199 (25.7)	79 (27.2)	120 (24.8)
Autogenic drainage (including assisted autogenic drainage); n(%)	290 (37.5)	87 (29.9)	203 (42.0)
Any form of PEP; n(%)	395 (51.0)	223 (76.6)	172 (35.6)
VEST; n(%)	5 (0.7)	<5	<5

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

1.26 Other therapy

	Overall (n=782)	<16 years (n=297)	≥16 years (n=485)
NIV; n(%)	9 (1.2)	<5	5 (1.0)
Long-term oxygen; n(%)	39 (5.0)	10 (3.4)	29 (6.0)
Among those who had long-term oxygen therapy:			
Continuously	9 (23.1)	2 (20.0)	7 (24.1)
Nocturnal+exertion	11 (28.2)	1 (10.0)	10 (34.5)
PRN	<5	0	<5
With exacerbation	17 (43.6)	7 (70.0)	10 (34.5)

1.27 Feeding

Supplementary feeding, often using a nasogastric (via the nose) or gastrostomy (via the abdomen) tube directly to the stomach, is considered when a person with CF has poor weight gain, or progressive weight loss, despite efforts to increase oral intake.

	All patients (n=782)	<16 years (n=297)	≥16 years (n=485)
Any supplemental feeding; n(%)	175 (22.4)	52 (17.5)	123 (25.4)
Nasogastric Tube			
Gastrostomy Tube / Button	10 (5.7)	0	10 (8.1)
Jejunal	40 (22.9)	14 (26.9)	26 (21.1)
TPN	0	0	0
	0	0	0

1.28 Age distribution of deaths in 2014

There were 15 recorded deaths in 2014. The median age at death was 26 years (range: 16 – 55 years; 95% confidence interval: 20.3-33.5)

Age at death	0 - 3	4 - 7	8 - 11	12 - 15	16 - 19	20 - 23	24 - 27	28 - 31	32 - 35	36 - 39	40 - 43	44 - 47	48 - 51	52 - 55	56+	Total
Number of CF patients	0	0	0	0	<5	<5	<5	<5	<5	0	0	0	0	<5		15

1.5 Genotypes

Genotypes are part of the genetic makeup of a cell, organism, or individual that usually controls a particular characteristic (known as phenotype). For people with cystic fibrosis, their genotype reveals which mutations of the CF gene cause their cystic fibrosis. Everyone living with cystic fibrosis has two mutations of the gene for CFTR; one on each allele. One is inherited from their mother, and one from their father. If both mutations,(or genotypes) are the same, the person is said to be homozygous. Someone who has two different variants is heterozygous.

766 (98.0%) patients have been genotyped with a recorded value

DF508 Mutations; n (%)

Homozygous DF508	336 (43.9%)
Heterozygous DF508	356 (46.5%)
No DF508 or both unidentified 886 (9.6%)	74 (9.7%)

Mutations[1]			
All mutations Current Name	**New Name**	**N**	**(%)**
DF508	p.Phe508del	692	90.3
G551D	p.Gly551Asp	82	10.7
G542X	p.Gly542X	53	6.9
R117H	p.Arg117His	51	6.7
1717-1G->A	c.1585-1G>A	15	2
3659delC	c.3528delC	9	1.2
N1303K	p.Asn1303Lys	8	1
2789+5G->A	c.2657+5G>A	6	0.8
1898+1G->A	c.1766+1G>A	<5	

Section 2: Centre Level Analysis

Cystic fibrosis care in the Scotland is led by eight regional centres, two stand-alone clinics and three networked clinics. The breakdown between centres and clinics delivering paediatric and adult care is shown below:

	Paediatric	Adult	Total
Regional centres	5	3	8
Stand-alone clinics	1	0	1
Networked clinics	2	1	3

Section 2 shows analysis of data for individual cystic fibrosis centres. This allows people with CF, their families, and healthcare providers to compare centres against one another, and the national average. This level of transparency helps to improve standards of care by giving people with CF and healthcare providers alike the chance to make informed choices about what questions to ask of their team, and which types of treatment may be best for each individual.

It is important to remember that lots of different factors can affect the outcomes of people with CF in centres, not all of which are within the centre's control. This might include the economic profile of the area, the age at which the person with CF was diagnosed and referred to the centre, certain patient characteristics such as their gender, as well as facilities, care pathways, and the medical team providing care.

If a person with CF or a member of their family has questions about the results for their CF centre or clinic, they should discuss this with their CF team.

Full tables of data are shown in appendix 1 on page XX.

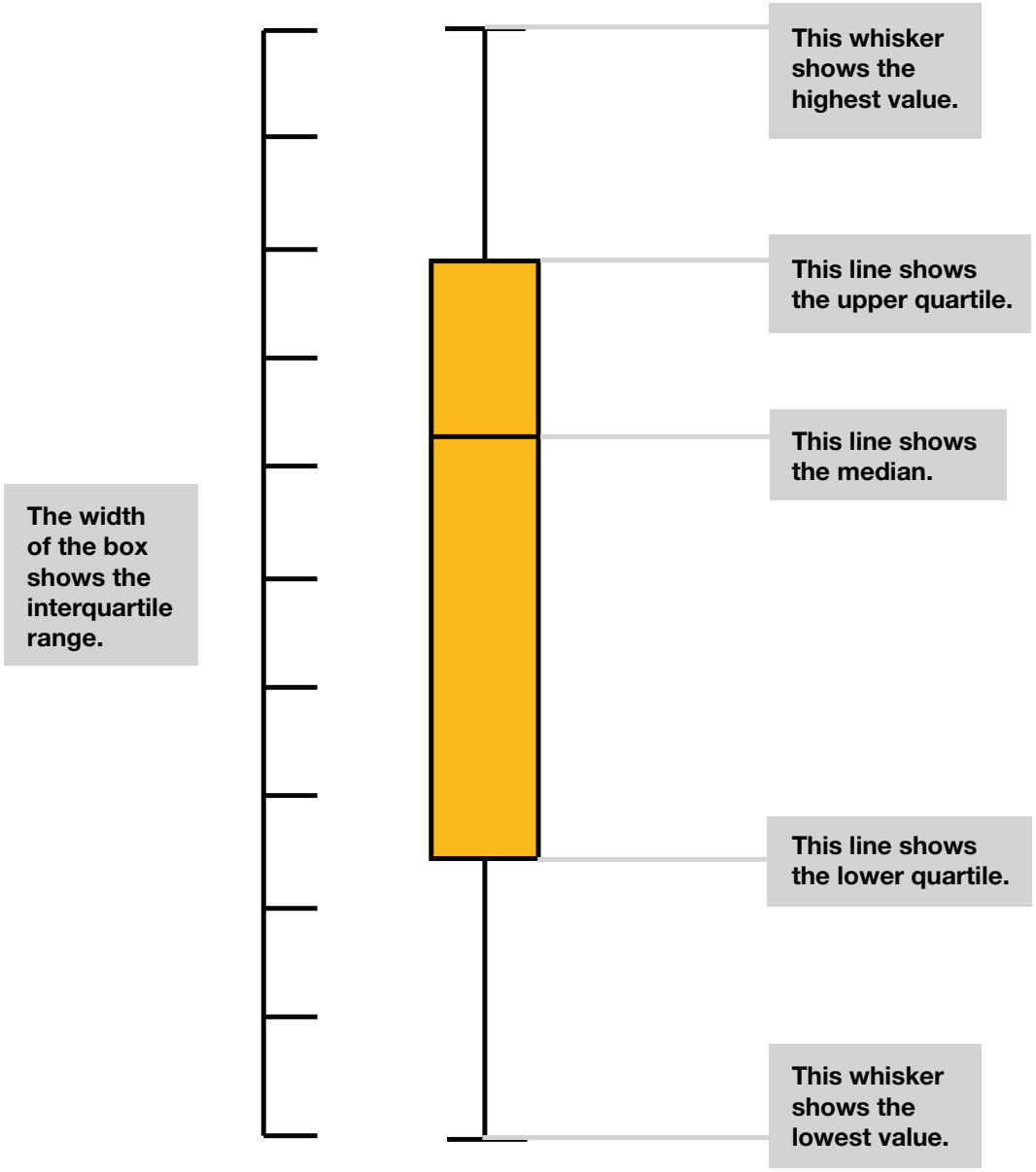
Key

Paediatric centre

Adult centre

A guide to the charts

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



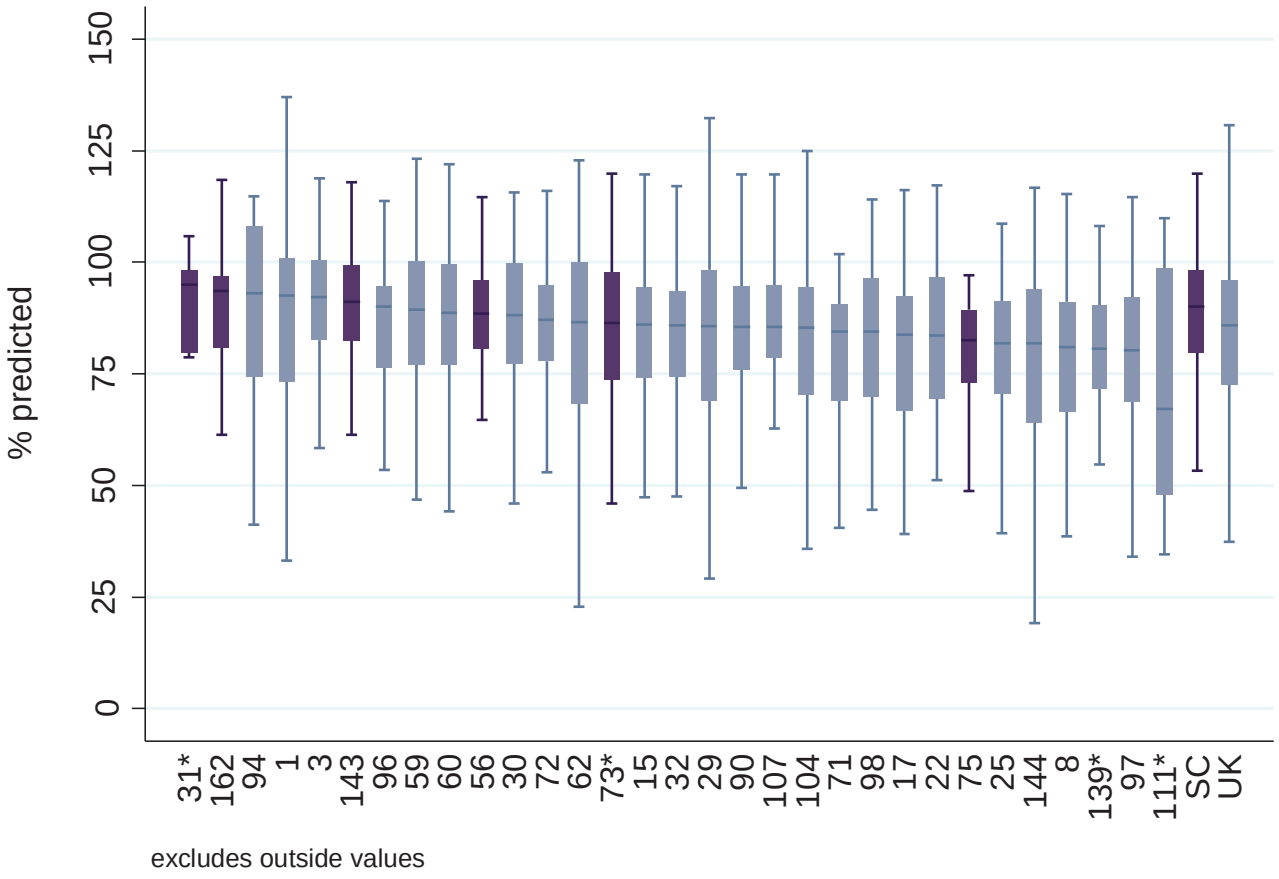
- The ‘box’ shows the middle half of the data for that centre, going from the first quartile to the third quartile. The longer the box, the more varied the data for that centre
- The horizontal line within the box shows the median result for that centre
- The ‘whiskers’ above and below the box shows the highest and lowest values for that centre, excluding any outliers
- The position of the box between the whiskers shows any skew in the data. If a box is towards the top of the whisker, more of the people for this centre were recorded at the high end of the scale

Section 2a Paediatric centre analysis

n = 309

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

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



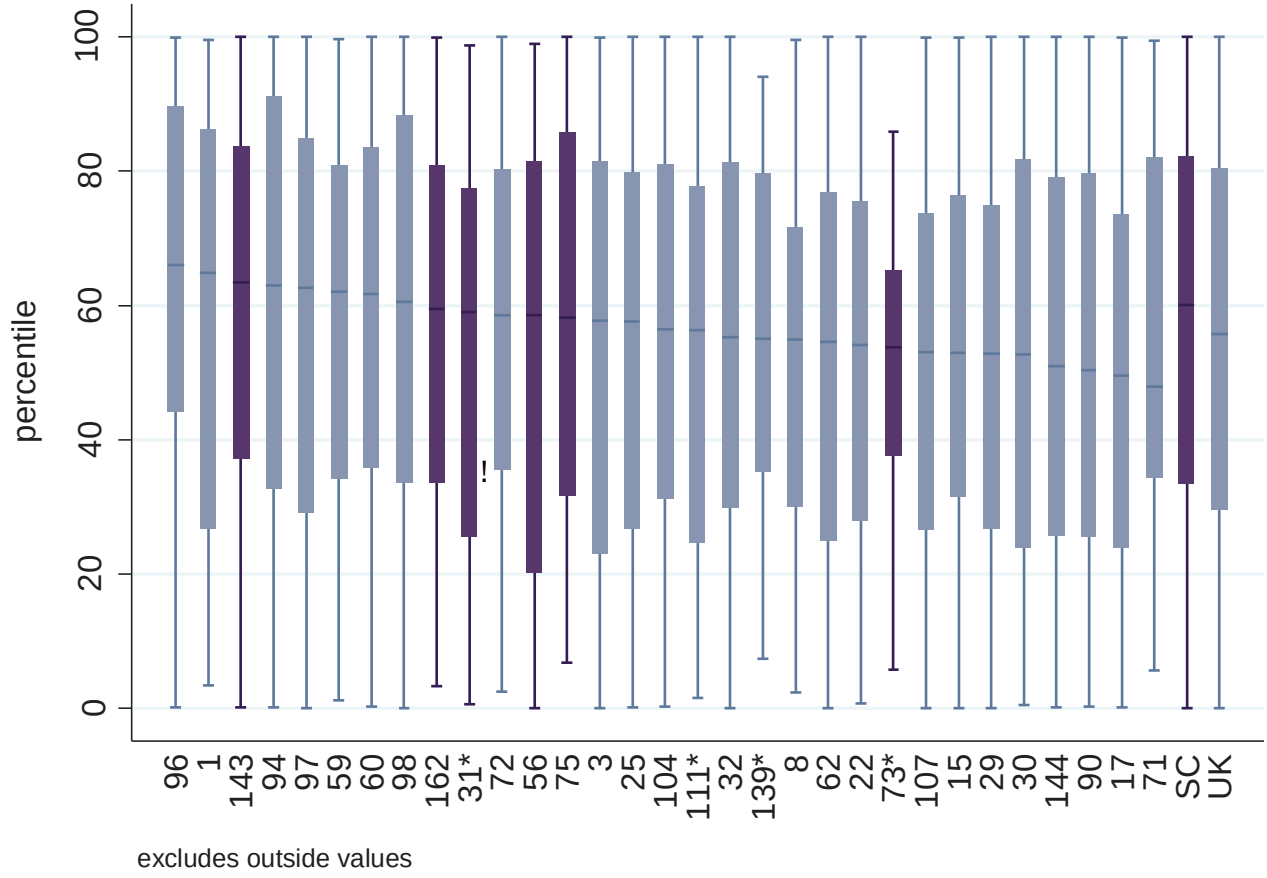
The median FEV_1 % predicted of patients attending paediatric centres/clinics in Scotland is 86% predicted (IQR: 73-96).

* Centre/clinic with a dataset submission of less than 20 patients.

Blue: Services in the UK

Purple: Services in Scotland

Figure 2.2 Median Body Mass Index (BMI) percentile among patients aged 2 to 15 years by paediatric centre/clinic



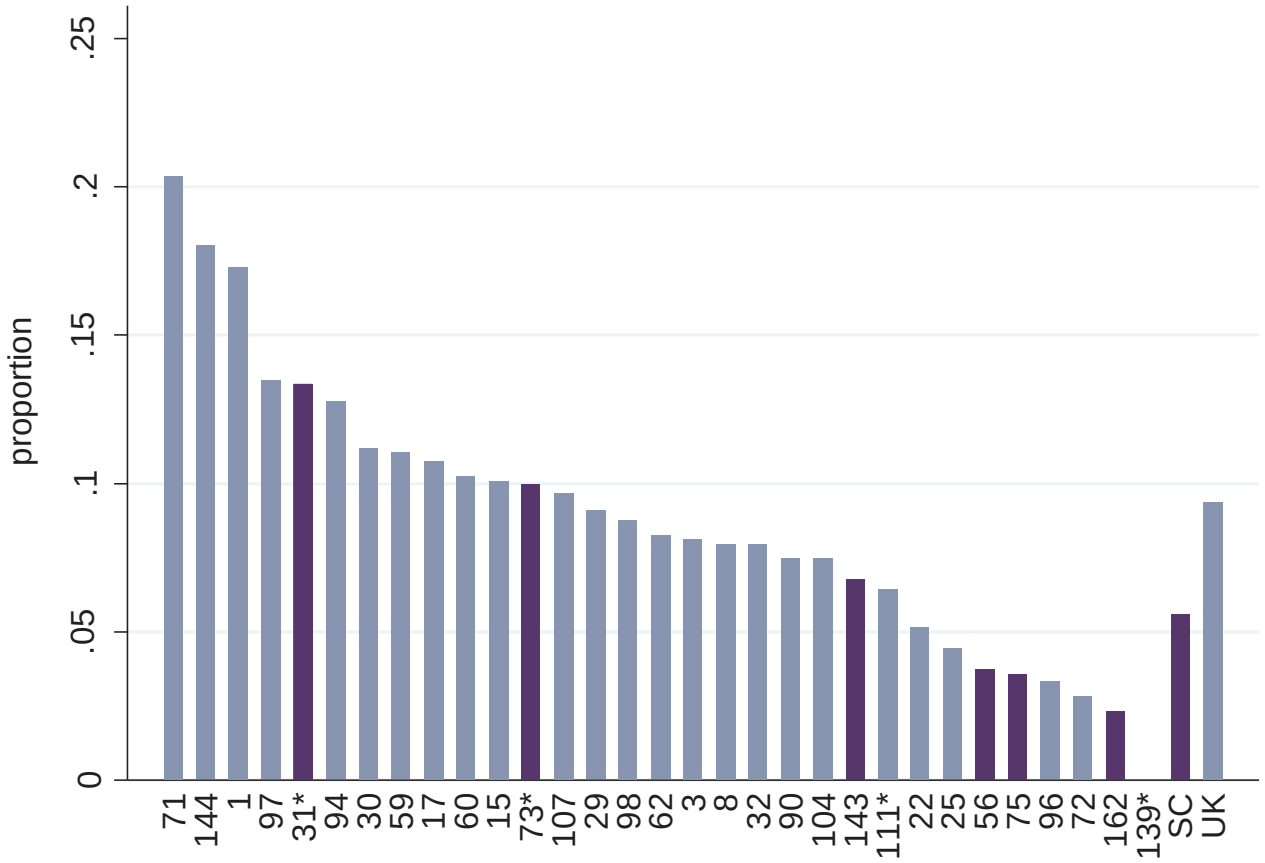
The median BMI percentile of patients attending paediatric centres/clinics in Scotland is 56 (IQR: 30 – 80).

* Centre/clinic with a dataset submission of less than 20 patients.

Blue: Services in the UK

Purple: Services in Scotland

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



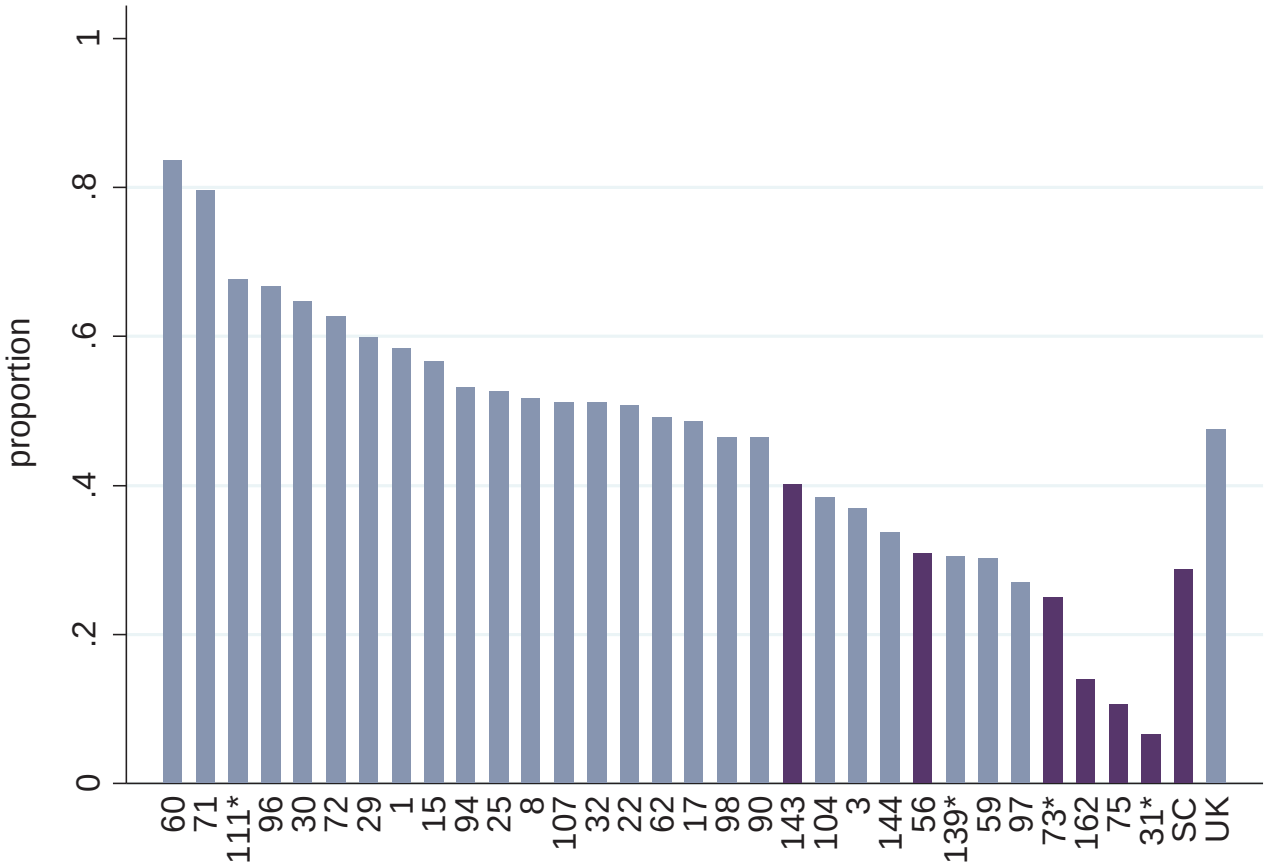
The proportion of patients attending paediatric centres/clinics in Scotland with chronic P.aeruginosais 6%.

* Centre/clinic with a dataset submission of less than 20 patients.

Blue: Services in the UK

Purple: Services in Scotland

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



The proportion of patients attending paediatric centres/clinics in Scotland receiving DNase treatment is 29%.
* Centre/clinic with a dataset submission of less than 20 patients.

Blue: Services in the UK

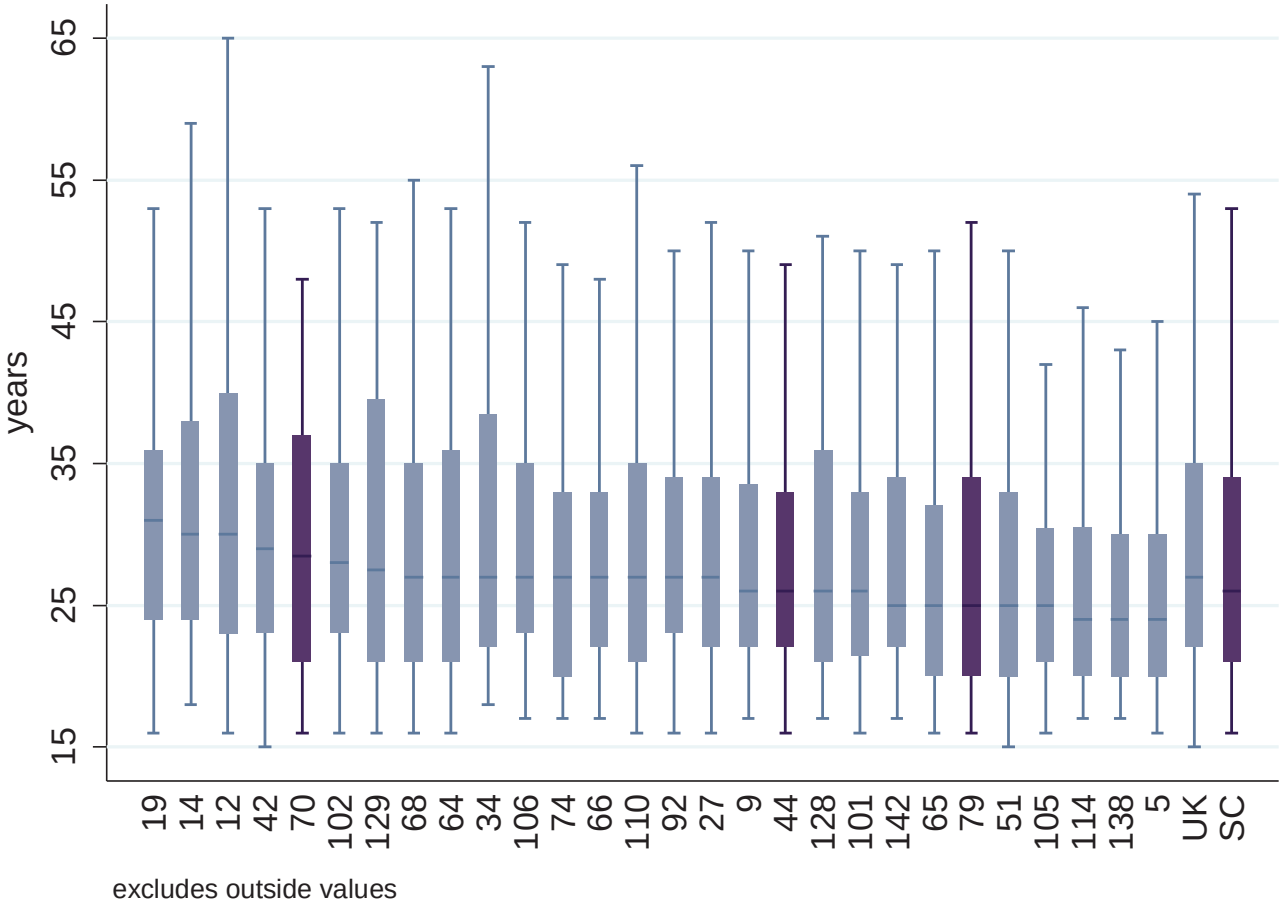
Purple: Services in Scotland

Section 2b Adult Centres Analysis

n = 473

This section shows results for the three adult centres with any network clinics.

Figure 2.5 Median age (years) by adult service

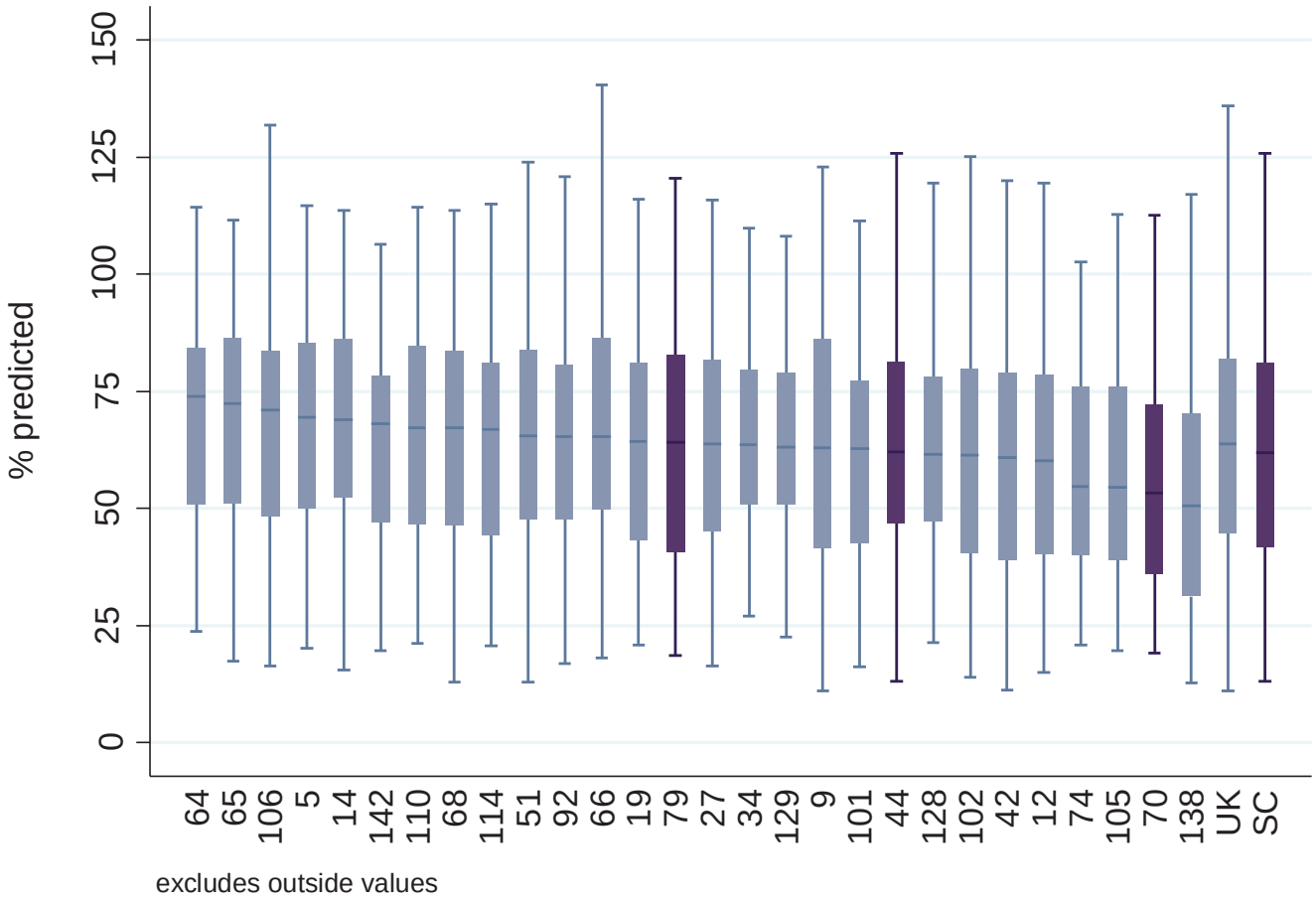


The median age of patients attending adult services in Scotland is 27 years (IQR: 21-35).

Blue: Services in the UK

Purple: Services in Scotland

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

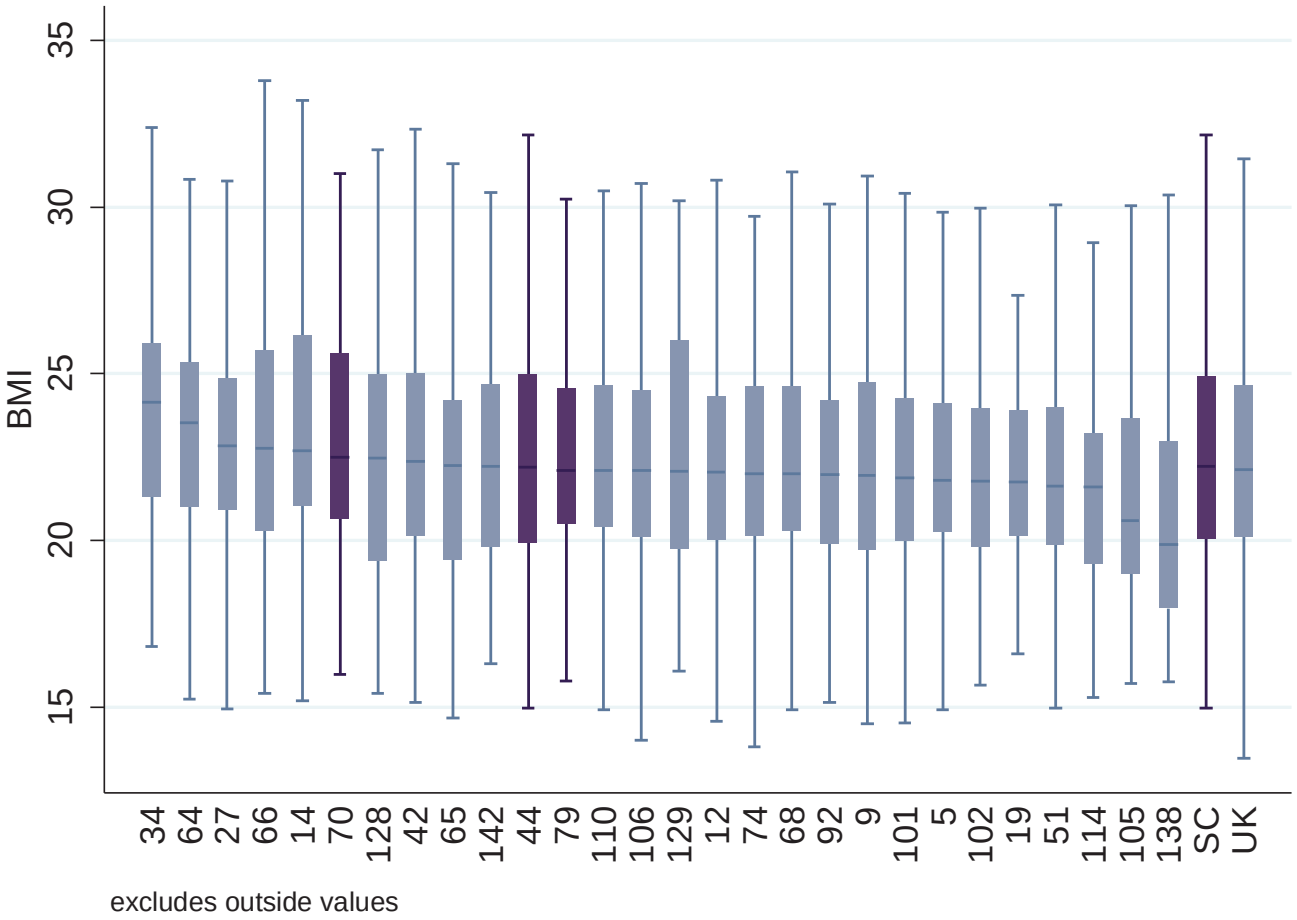


The median FEV_1 (% predicted) of patients attending adult services in Scotland is 62% (IQR: 42 – 81).

Blue: Services in the UK

Purple: Services in Scotland

Figure 2.7 Median Body Mass Index (BMI) among patients aged 16 years and older by adult service

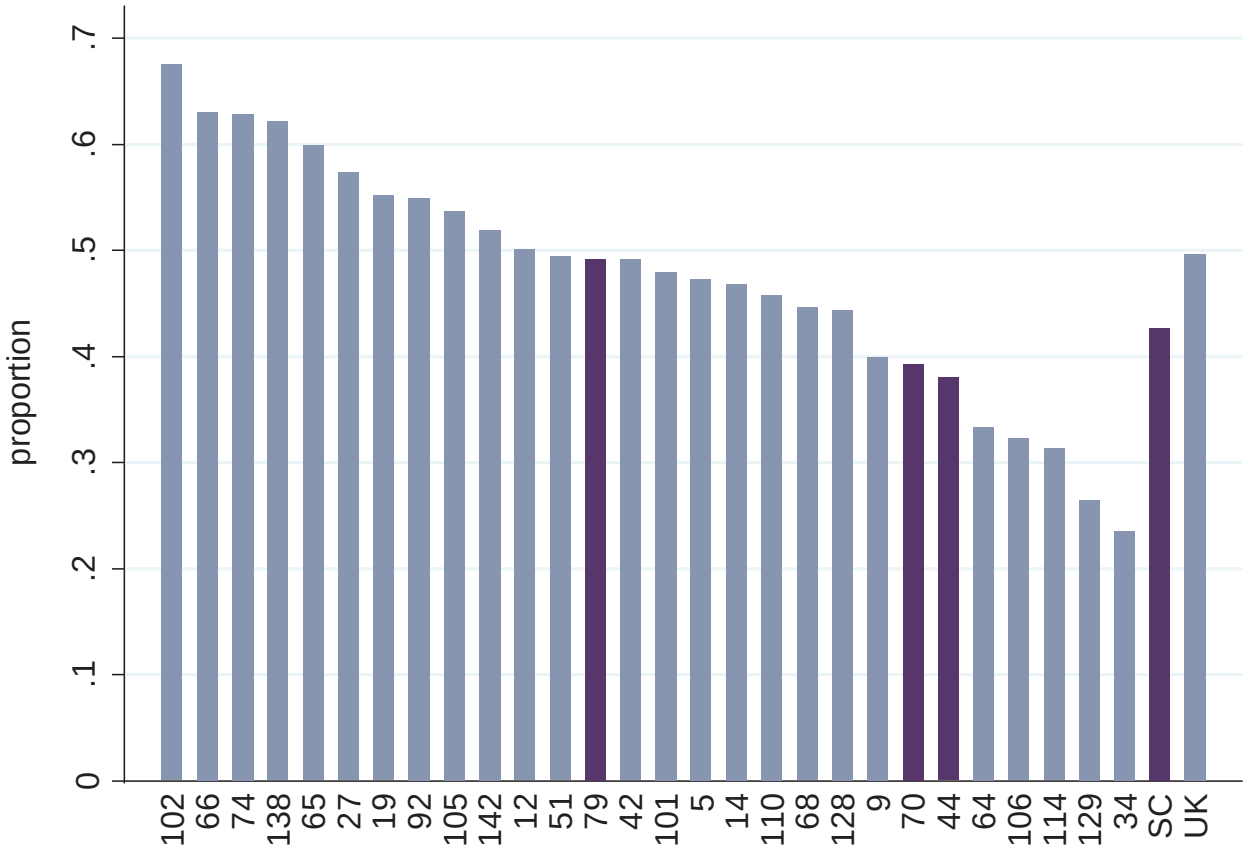


The median BMI of patients attending adult services in Scotland is 22 (IQR: 20 – 25).

Blue: Services in the UK

Purple: Services in Scotland

Figure 2.8 Proportion of patients with chronic P.aeruginosaby adult service

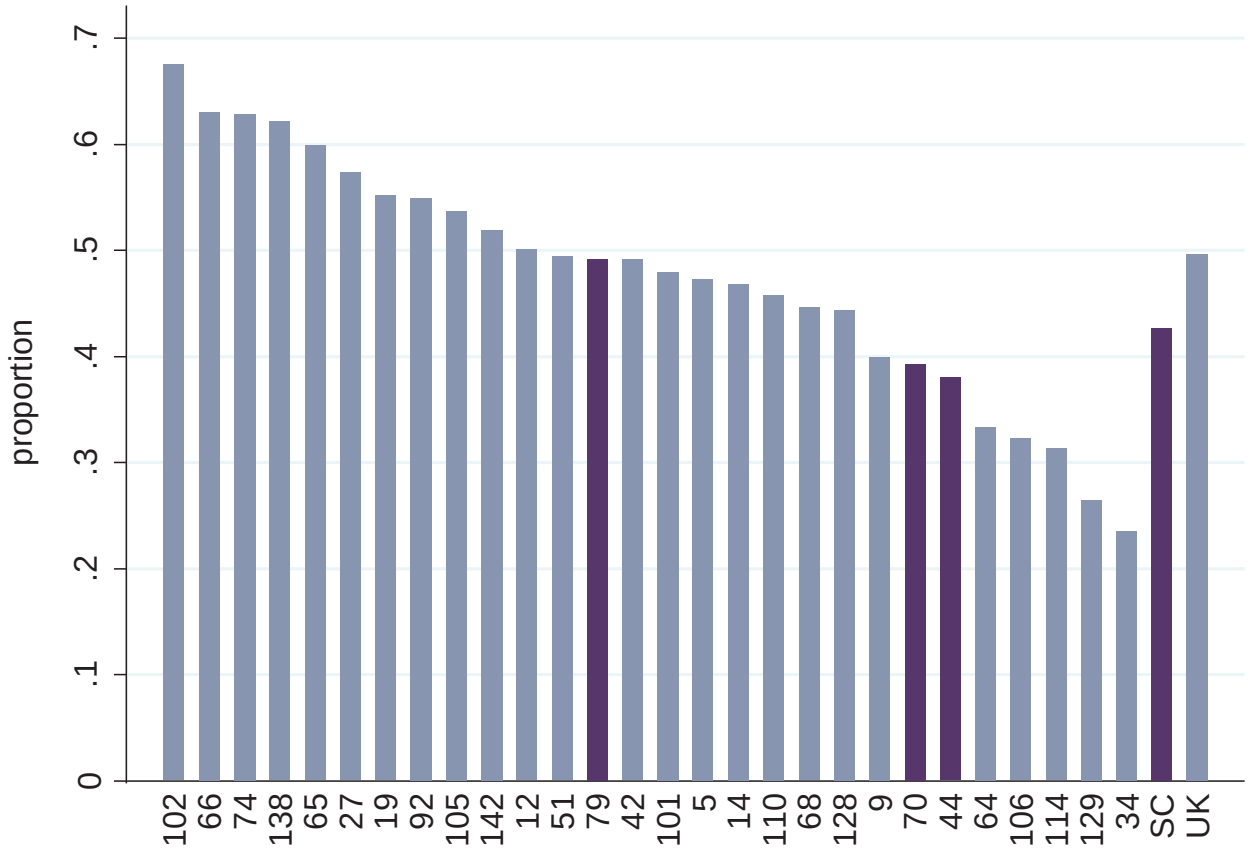


The proportion of patients attending adult services in Scotland with chronic P.aeruginosais 43%.

Blue: Services in the UK

Purple: Services in Scotland

Figure 2.9 Proportion of patients receiving DNase treatment by adult service



The proportion of patients attending adult services in Scotland receiving DNase treatment is 35%.

Blue: Services in the UK

Purple: Services in Scotland

Appendix 1: Centre level data tables

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

Scotland Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2014	Median FEV_1% predicted (≥6 years)	Median BMI percentile (2-15 years)
Inverness	Raigmore Hospital	31	17	15	101.6	55
Glasgow	Royal Hospital for Sick Children	56	110	81	88.5	74.9
Dundee	Ninewells Hospital	73	22	20	90.26	55.8
Aberdeen	Royal Aberdeen Children's Hospital	75	29	28	80.4	50.4
Edinburgh	Royal Hospital for Sick Children	143	124	122	92.2	63.9
Lanarkshire	Wishaw General Hospital	162	45	43	90.8	50.6

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

Scotland Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2014	Median FEV_1% predicted (≥16 years)	Median BMI percentile (≥16 years)
Edinburgh	Western General Hospital	44	226	221	66.6	22.2
Aberdeen	Aberdeen Royal Infirmary	70	66	61	57.4	22.5
Glasgow	Gartnavel General Hospital	79	213	191	67.2	22.1

Paediatric centres/clinics providing data in 2014 – ordered alphabetically

Scotland Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2014	Median FEV_1% predicted (≥6 years)	Median BMI percentile (2-15 years)
Aberdeen	Royal Aberdeen Children's Hospital	75	29	28	80.4	50.4
Dundee	Ninewells Hospital	73	22	20	90.26	55.8
Edinburgh	Royal Hospital for Sick Children	143	124	122	92.2	63.9
Glasgow	Royal Hospital for Sick Children	56	110	81	88.5	74.9
Inverness	Raigmore Hospital	31	17	15	101.6	55
Lanarkshire	Wishaw General Hospital	162	45	43	90.8	50.6

Adult centres/clinics providing data in 2014 – ordered alphabetically

Scotland Location	Centre/clinic	Clinic ID	Number of active patients	Number of patients providing data in 2014	Median FEV_1% predicted (≥16 years)	Median BMI percentile (≥16 years)
Aberdeen	Aberdeen Royal Infirmary	70	66	61	57.4	22.5
Edinburgh	Western General Hospital	44	226	221	66.6	22.2
Glasgow	Gartnavel General Hospital	79	213	191	67.2	22.1

Appendix 2: Glossary

Word/Phrase	Meaning
2014	1 January 2014 – 31 December 2014.
ABPA (Allergic Bronchopulmonary Aspergillosis)	When a person develops a respiratory allergic reaction to the Aspergillus fungus.
Absence of vas deferens	The vas deferens connect the testicles to the penis. Where a male is missing both vas deferens sperm cannot be transported.
Arthritis	A condition causing pain and inflammation in the joints.
Arthropathy	A condition causing pain in the joints.
Asthma	A respiratory condition causing episodes of difficulty breathing during attacks of spasm in the lung.
B. cepacia	Burkholderia cepacia complex are a group of bacteria, some of which threaten the health of people with cystic fibrosis.
BMI (Body Mass Index)	A measure designed to show whether a person is a healthy weight for their height.
CF	Cystic fibrosis.
CFTR (Cystic Fibrosis Transmembrane conductance Regulator)	This is a protein at the cell surface that controls the salt and water balance across a cell. The gene that causes cystic fibrosis is the blueprint for the CFTR protein. Everyone has two copies of the gene for CFTR. To be born with cystic fibrosis, both CFTR genes must be affected by a CF-causing mutation.
Chronic	Persistent, or long-lasting.
Cirrhosis	A chronic liver disease.
Confidence interval	Confidence intervals are calculated to show the range of results we would expect, based on the overall average. If a result is between the upper and lower limits of the confidence interval, it is 'as expected'.
Enzymes	Biological molecules that help complex reactions, such as digestion of food, occur in the body.
FEV_1 (Forced Expiratory Volume in one second)	This is the amount of air that a person can blow out of the lungs in the first second of a forced exhaled breath. People with healthy lungs can blow out most of the air held in this time.
FEV_1 % predicted	The FEV_1 can be converted from absolute litres of air blown out into a predicted percentage (%). A healthy range for % predicted is calculated from a very large population sample, and is normally considered to be between 80-120% predicted.

Word/Phrase	Meaning
Fibrosing colonopathy	A condition causing narrowing of part of the colon.
Gall bladder	The small sac-shaped organ under the liver that stores bile after it is secreted by the liver, before it is released into the intestine.
Gastrointestinal (GI)	The GI tract is an organ system responsible for digesting food, absorbing nutrients and expelling waste.
Genotype	Part of the genetic makeup of a cell, organism, or individual, that usually controls a particular characteristic (known as a phenotype).
GORD (Gastrooesophageal Reflux Disease)	A chronic symptom of damage caused by stomach acid coming up from the stomach into the oesophagus.
GI bleed	Bleeding in the gastro-intestinal tract.
GLI (Global Lung Initiative) equations	An equation for calculating FEV_1% predicted that takes into account age, gender, height and ethnicity.
Haemophilus influenza	Haemophilus influenza (H. influenzae) is a bacterium that can cause respiratory infection.
Haemoptysis	The coughing up of blood.
Hepatobiliary disease	A liver or biliary disorder.
Heterozygous	Everyone living with cystic fibrosis has two CF-causing mutations of the gene for CFTR, one inherited from their mother and one from their father. Someone who has two different mutations is heterozygous.
Homozygous	Everyone living with cystic fibrosis has two CF-causing mutations of the gene for CFTR, one inherited from their mother and one from their father. If both mutations (or genotypes) are the same, the person is said to be homozygous.
Hypertension	High blood pressure.
Incidence	The number of people newly diagnosed with a condition in the given year.

Word/Phrase	Meaning
IQR (InterQuartile Range)	Also called the mid-spread, or middle fifty, IQR is a measure of the spread of data. It shows the difference between the upper and lower quartiles. IQR = Q3 – Q1.
Median	The middle number, when all numbers are arranged from smallest to largest.
Median age of death	Median age of death is based on the people with CF who passed away in any given year. So in 2014 the median age of the 137 people who died was 28.
Median predicted survival	Median predicted survival is a calculation based on people with CF recorded in the Registry as alive in the given year. A mathematical formula, which takes into account the age of those people in 2014, predicts how long we expect half of them to live for. For 2014, this means that half of people registered as alive on the database are predicted to live to at least 40.1. Half of people alive today are predicted to die before they reach that age.
MRSA	Methicillin-resistant staphylococcus aureus is a type of bacteria that is resistant to a number of widely used antibiotics.
Mutation	A mutation is a change in a gene. When both of a child's parents are carriers of a CF-causing mutation there is a 25% chance that the child will have cystic fibrosis. There are over 1,400 different mutations of the CFTR gene that can cause cystic fibrosis.
Nasal Polyps	Small, sac-like growths of inflamed mucus caused by chronic inflammation of the nasal lining.
Non-tuberculosis Mycobacteria (NTM)	A mycobacterium that does not cause tuberculosis, but which can cause respiratory infection. There are several types known.
Osteopenia	A medical condition less severe than osteoporosis, where the mineral content of bone is reduced.
Osteoporosis	A condition where the bones become brittle from loss of tissue.
Pancreas	An organ in the digestive system that produces insulin and digestive enzymes.
Pancreatitis	Inflammation of the pancreas.

Word/Phrase	Meaning
Peptic ulcer	Or, stomach ulcer, is an open sore that develops in the lining of the stomach.
Percentile	A percentile shows where a value stands, relative to the rest of the data. If a value is higher than 90% of the rest of the data, it is above the 90th percentile.
Pneumothorax	A collection of air in the cavity between the lungs and the chest wall causing collapse of the lung on the affected side.
Portal hypertension	High blood pressure in the portal vein system, which is the blood system of the liver.
Pre-natal	Before birth, whilst the baby is still in the womb.
Prevalence	The overall number of people diagnosed with a condition at any time.
Pseudomonas aeruginosa	A tough bacterial strain, rarely affecting healthy people, that can cause respiratory infection.
Rectal prolapse	When the rectal wall slides through the anus.
Renal	Relating to the kidneys.
Staphylococcus aureus	Staphylococcus aureus (S. aureus) is a bacteria that can cause respiratory infection.
Sinus disease	When the sinuses, which are usually filled with air, are typically full of thick sticky mucus.
Statistically significant	This phrase means that after careful calculations there is a definite difference between two groups, which is not simply a result of chance.

Appendix 3: UK CF Registry Steering Committee

Composition of UK CF Registry Steering Committee

Dr Janet Allen Director of Strategic Innovation, Cystic Fibrosis Trust

Professor Diana Bilton Adult CF Centre Director, Royal Brompton

Ms Noreen Caine Contract Consultant, Cystic Fibrosis Trust

Dr Siobhán Carr Paediatrician, Royal Brompton Hospital, London (Chair)

Ms Katherine Collins Caldecott Guardian, Director NSD, Scotland

Ms Rebecca Cosgriff Registry Lead, Cystic Fibrosis Trust

Dr Kim Cox Lead Specialist CF Commissioner, London

Dr Steve Cunningham Paediatrician, Royal Hospital for Sick Children, Scotland

Mrs Marian Dmochowska Parent Representative

Dr Iolo Doull Paediatric CF Centre Director, Cardiff Hospital, Wales

Dr Caroline Elston Adult CF Centre Director, King's College Hospital, London

Ms Carrie Gardner Specialised Commissioner, NHS England

Ms Elaine Gunn Registry Manager, Cystic Fibrosis Trust

Mr Dominic Kavanagh Patient Representative

Dr Claire Nelson Specialised Commissioner, Wales

Dr Stephen Nyangoma Biostatistician, Imperial College, London

Mr Ed Owen Chief Executive, Cystic Fibrosis Trust

Ms Vian Rajabzadeh-Heshejin Data Analyst, Imperial College, London

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

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