Cystic Fibrosis strength in numbers

UK Cystic Fibrosis Registry
2015 Annual Data Report

Scotland

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An at-a-glance version of this report can be found at **www.cysticfibrosis.org.uk/registryreports**

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Acknowledgements

First and foremost, the UK Cystic Fibrosis Registry team would like to thank people with cystic fibrosis and their families for their support, as well as anyone who has generously donated to the Cystic Fibrosis Trust. We would also like to express our gratitude to the UK cystic fibrosis centres and clinics, for their continued dedication to obtaining consent and submitting data to the Registry.

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Cystic Fibrosis strength in numbers

Contents

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.

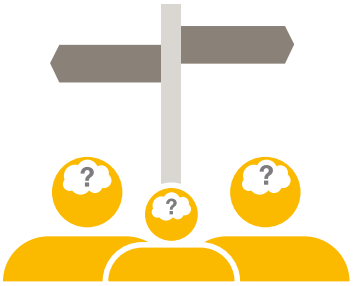
Cystic fibrosis

Cystic fibrosis is an inherited disease caused by a faulty gene known as 'CFTR'. The gene and the protein it makes control the movement of salt and water in and out of cells. When the gene is faulty, it can cause thicker mucus that blocks airways. This affects the lungs, which over time make it hard to breathe. 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 in England provide funding to NHS CF centres that is proportionate to their patients' disease severity.

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 makes recommendations about the future development of the Registry. A sub-committee of the RSC, known as the Registry Research Committee, assesses applications for data and guides the Registry research strategy.

Please see appendix 3 for members of each committee.

Data are only recorded on the UK CF Registry if explicit written consent is given by the person with cystic fibrosis or, for a child, their parent or guardian.

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. This 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**.

i **Words in this report that appear in the glossary are highlighted the first time they appear and explained in appendix 2 on page 46.**

Section 1: Scotland-wide analysis

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

1.1 Summary of the UK Cystic Fibrosis Registry

	2015	
	UK	**Scotland**
CF patients registered[1] Excluding 2014 diagnoses	10810[1] 10586	966[1] 959
CF patients with "complete" data [2]**; n(%)** Rate of completeness excluding 2014 diagnoses	9587 (91%)	795 (82%)
Age in years; median[3]	19	20
All newly diagnosed patients (newborn screening and other)[4]	224	7
Newly diagnosed patients identified through newborn screening[4]	112	3
Age at diagnosis in months; median[3]	2	2
Adults aged 16 yrs and over; %[3]	59.9	62.5
Males; %	53	52
Genotyped; %[3]	98.1	98.6
Total deaths reported[4]	125 (1.2%)	11 (1.1%)
Age at death in years; median (95%CI)	28 (25.5,32.0)	27 (18.7-55.9)

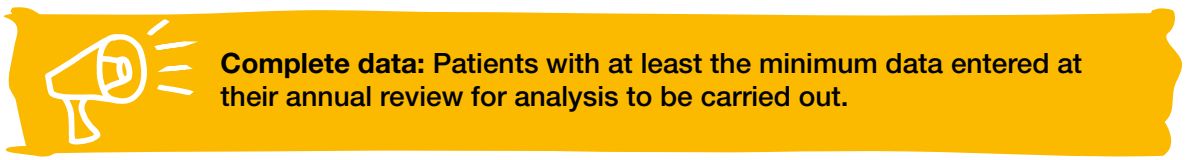
Notes:

[1] This is calculated as the number of patients on the database who were diagnosed with CF and had not died before 1 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 2015 may not have their first annual review in the same year. If newly diagnosed patients are excluded, 82% of records are complete.

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

[4] Calculated from 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.

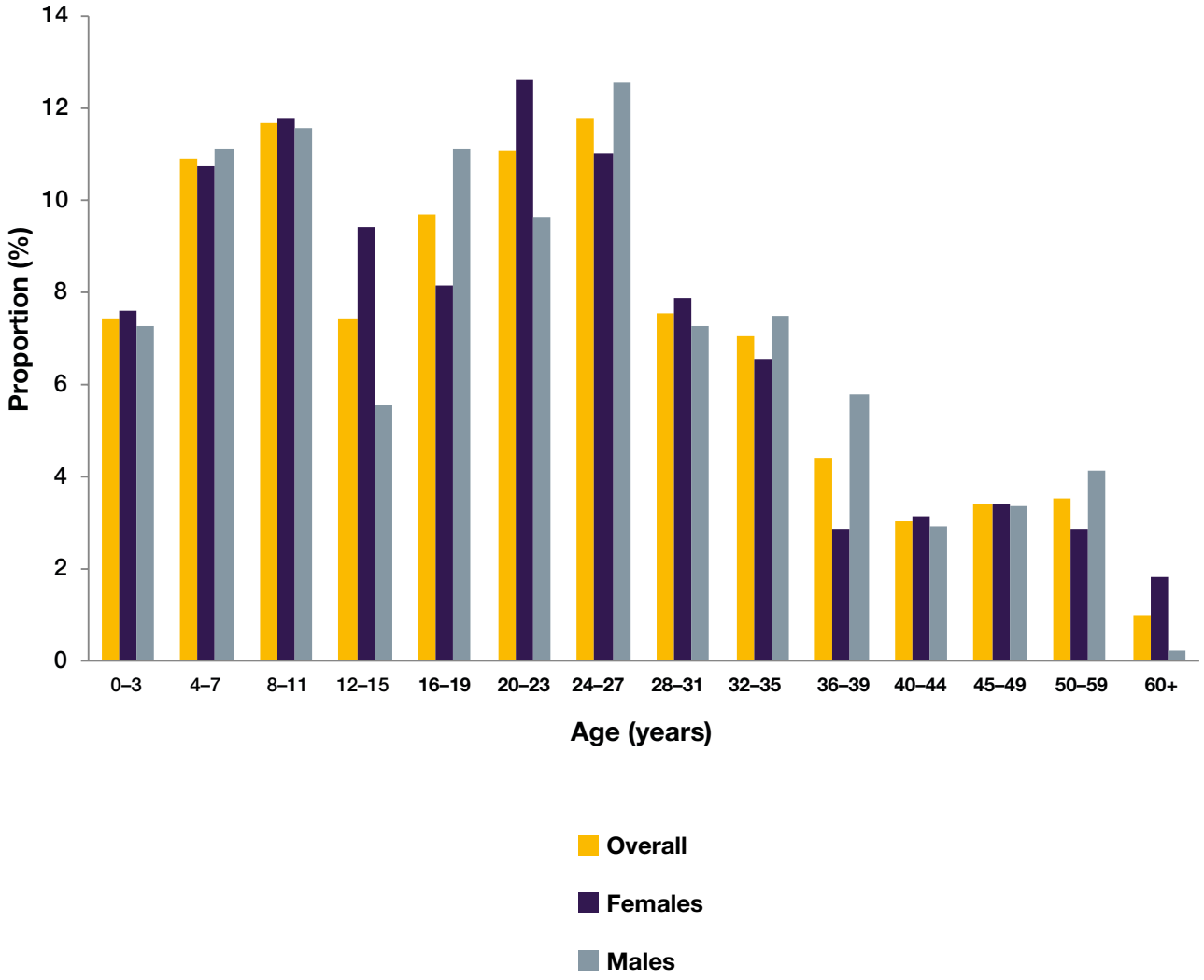


Complete data: Patients with at least the minimum data entered at their annual review for analysis to be carried out.

1.2 Age distribution by gender

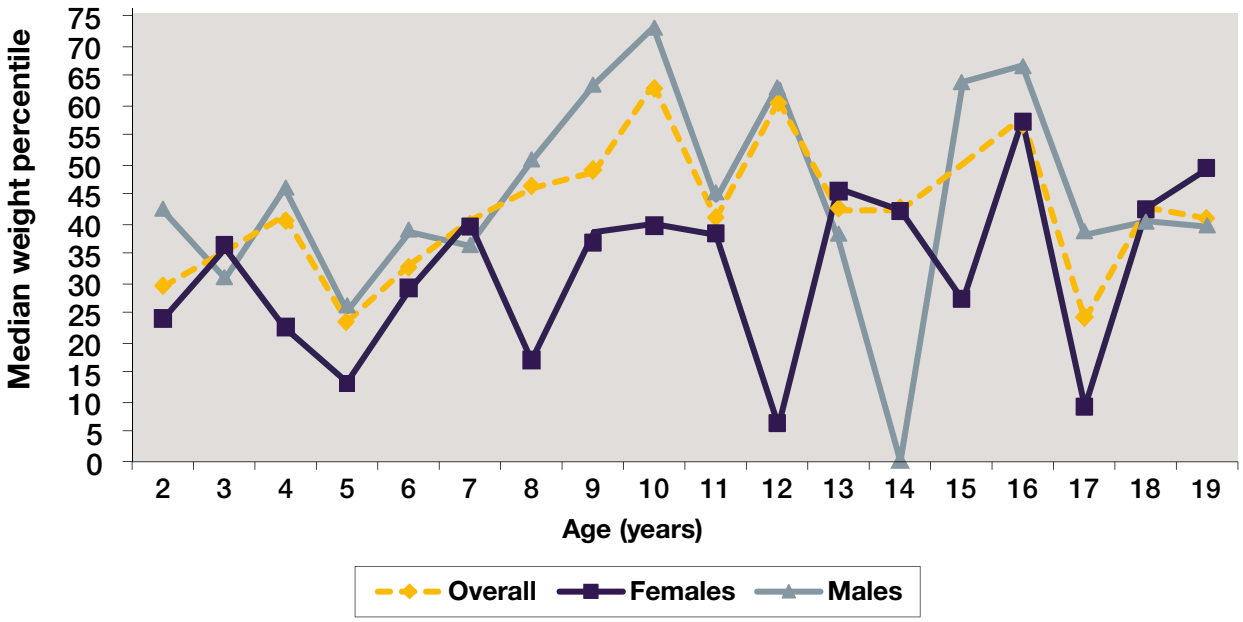
n=795

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=353

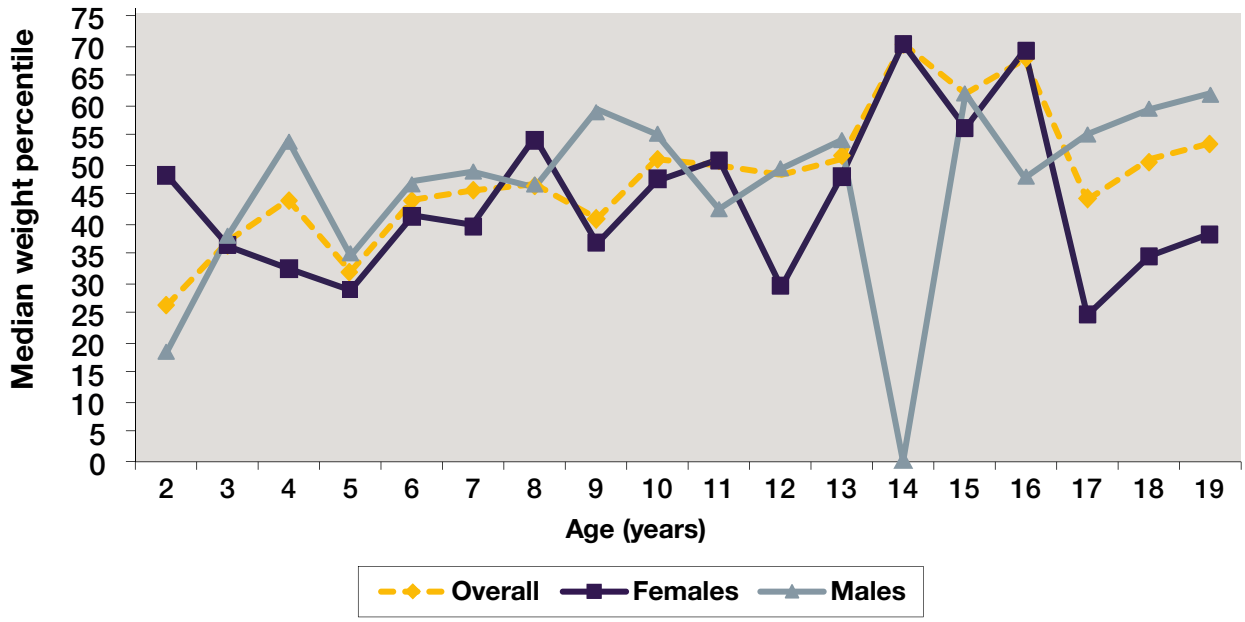
The following chart and table show the height percentiles of people with CF, aged 19 and under, in relation to the UK growth data for the general population. If a person with CF is on the 40th percentile, only 40% of the population at the same age are their height or shorter, 60% are taller



	Overall			Female			Male		
Age	**N**	**Median**	**IQR**	**N**	**Median**	**IQR**	**N**	**Median**	**IQR**
2	16	29.6	15.1-58.0	9	24.0	12.6-69.7	7	42.5	22.6-64.3
3	21	35.4	8.2-49.2	11	36.0	8.7-53.0	10	31.0	2.4-41.3
4	22	41.7	7.4-66.8	8	22.8	5.5-70.9	14	46.2	7.4-67.3
5	26	23.7	5.2-57.9	13	13.6	4.4-52.3	13	25.6	6.5-67.5
6	24	33.3	14.2-74.4	11	28.9	13.8-77.7	13	38.6	10.1-71.5
7	15	40.8	27.0-47.1	9	40.8	28.6-46.7	6	36.3	17.8-60.7
8	24	46.1	12.0-66.8	9	17.2	11.6-60.3	15	50.8	25.5-72.6
9	28	49.0	20.6-73.9	18	39.0	20.8-63.6	10	63.6	12.4-83.3
10	16	63.7	40.4-84.0	5	40.4	1.9-52.6	11	73.5	59.3-88.6
11	25	41.0	27.2-74.6	13	38.2	27.2-71.8	12	44.4	23.7-80.4
12	16	60.9	19.3-74.6	5	6.1	5.3-95.3	11	63.7	43.1-74.4
13	20	42.6	28.3-68.1	13	46.1	33.1-77.7	7	38.0	14.7-46.2
14	7	42.5	20.5-75.7	7	42.5	20.5-75.7	0	0	0
15	16	50.1	10.3-71.2	11	27.4	6.8-66.8	5	64.1	37.6-73.6
16	17	58.1	20.6-79.7	6	57.5	18.7-64.8	11	67.0	21.3-86.9
17	13	24.0	9.3-44.9	<5	9.3	4.4-32.8	9	38.4	23.5-52.7
18	27	42.9	18.4-59.7	11	43.0	7.7-52.4	16	40.5	19.8-67.7
19	20	41.0	18.3-71.1	10	49.2	28.8-76.0	10	39.8	14.5-66.7
Overall	**353**	**37.1**	**14.4-63.9**	**173**	**35.7**	**13.2-62.4**	**180**	**45.4**	**21.9-70.8**

1.4 Median weight percentiles of children and young persons (<20 years) n=353

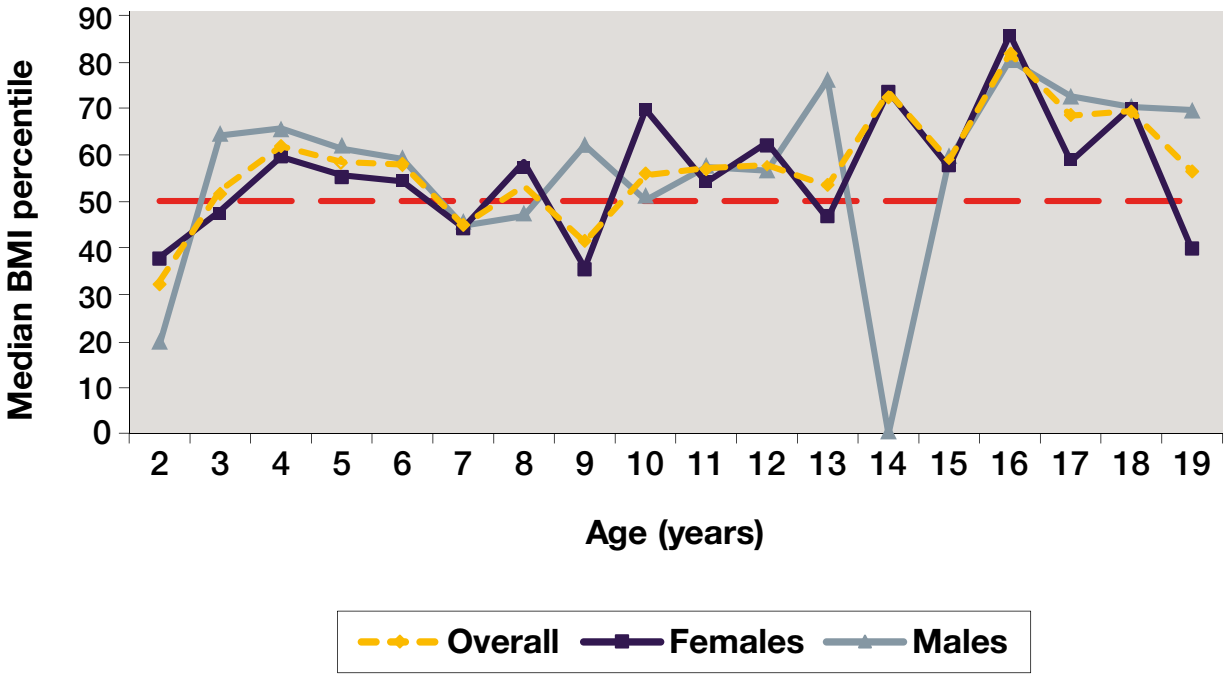
The following chart and table show the weight percentiles of people with CF, aged 19 and under, in relation to the UK growth data for the general population. If a person with CF is at the 40th percentile, only 40% of the population at the same age are their weight or lower, 60% weigh more.



	Overall			Female			Male		
Age	**N**	**Median**	**IQR**	**N**	**Median**	**IQR**	**N**	**Median**	**IQR**
2	16	26.2	12.0-59.4	9	48.1	17.8-65.7	7	18.1	9.3-32.3
3	21	37.2	8.8-56.5	11	36.2	3.6-50.8	10	38.2	17.2-59.6
4	22	43.8	9.5-84.7	8	32.6	13.7-80.0	14	54.1	8.4-85.2
5	26	31.8	9.8-86.5	13	28.8	5.6-76.9	13	34.7	11.8-87.9
6	24	44.1	27.0-73.9	11	41.3	28.9-76.6	13	47.0	21.9-71.7
7	15	45.7	20.9-55.2	9	40.0	24.3-62.3	6	49.0	6.6-59.1
8	24	46.9	31.0-73.9	9	54.7	25.3-85.6	15	46.3	30.3-69.9
9	28	41.1	18.7-68.7	18	36.8	15.3-53.7	10	59.5	19.5-77.5
10	16	50.9	33.2-83.5	5	47.3	20.7-68.9	11	55.1	35.6-84.3
11	25	49.6	28.9-76.6	13	51.0	28.1-71.2	12	42.7	28.9-89.9
12	16	48.2	30.5-73.3	5	29.5	15.0-73.8	11	49.5	42.7-74.1
13	20	50.9	25.5-80.5	13	47.8	28.8-86.1	7	54.0	17.4-70.8
14	7	70.5	45.7-79.2	7	70.5	45.7-79.2	0	0.0	0
15	16	62.1	24.9-80.4	11	56.1	17.1-80.8	5	62.1	40.1-82.1
16	17	67.9	27.7-91.9	6	69.4	56.9-90.6	11	47.7	18.4-98.1
17	13	44.3	17.0-63.0	<5	24.6	14.5-41.3	9	55.3	31.1-69.2
18	27	50.9	17.7-80.7	11	34.6	1.7-80.7	16	59.5	26.4-84.4
19	20	53.7	18.4-79.7	10	38.4	15.2-85.6	10	61.7	40.8-76.4
Overall	**353**	**47.2**	**24.4-73.9**	**173**	**44.0**	**21.2-70.8**	**180**	**48.3**	**26.9-75.8**

1.5 Median Body Mass Index (BMI) percentiles among children and young people (<20 years)
n=353

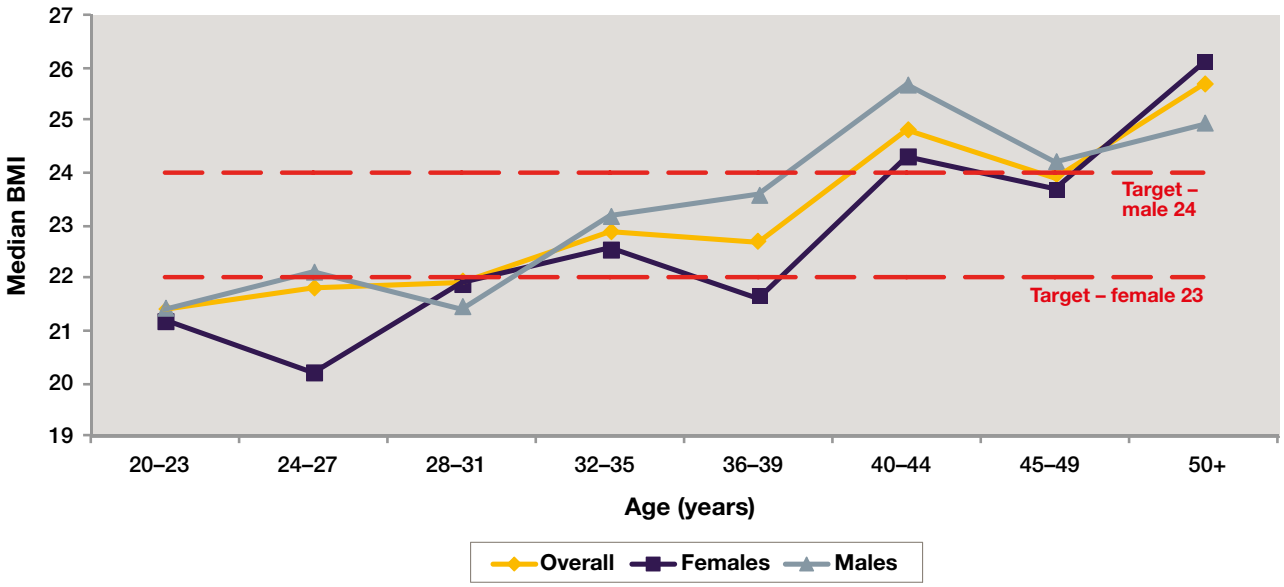
The following chart and table show the body mass index (BMI) percentiles of people with CF, aged 19 years and younger, in relation to the target BMI percentile for a person of the same age without CF (the 50th percentile, or the BMI percentile that half of the UK population of that age achieved). If 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	16	32.5	14.4-59.0	9	38.2	25.5-68.2	7	19.6	9.7-38.3
3	21	52.3	20.0-72.5	11	47.9	19.7-61.0	10	64.1	20.8-89.9
4	22	62.0	34.8-83.4	8	59.8	41.5-77.6	14	65.8	29.5-87.8
5	26	58.2	35.4-89.8	13	55.4	32.5-89.7	13	61.7	37.8-90.4
6	24	58.1	34.1-76.6	11	54.4	39.5-81.3	13	59.3	21.7-74.0
7	15	44.4	31.6-77.2	9	44.4	30.2-77.9	6	45.0	23.8-73.1
8	24	53.4	35.9-81.4	9	58.8	44.3-86.4	15	47.2	28.8-75.0
9	28	41.2	23.3-64.0	18	35.0	19.1-59.7	10	62.4	24.5-77.6
10	16	55.8	35.3-89.6	5	70.0	43.7-90.7	11	50.4	33.8-87.0
11	25	57.1	33.6-92.7	13	54.1	24.6-75.4	12	57.6	47.7-95.4
12	16	57.5	29.6-65.7	5	62.5	17.9-66.8	11	56.7	33.6-66.2
13	20	53.3	29.9-84.2	13	46.3	32.4-81.7	7	76.0	28.2-85.4
14	7	73.8	66.8-91.4	7	73.8	66.8-91.4	0	0.0	0
15	16	58.6	36.9-84.9	11	57.9	33.4-76.9	5	59.3	40.0-92.7
16	17	81.6	46.6-93.4	6	86.0	62.6-92.3	11	80.7	46.2-96.6
17	13	68.2	38.8-86.5	<5	58.8	25.6-80.6	9	72.7	38.8-88.9
18	27	69.7	23.4-92.4	11	69.7	2.9-92.4	16	70.4	44.4-91.9
19	20	56.5	12.3-90.0	10	40.3	8.2-80.1	10	69.8	41.6-90.7
Overall	**353**	**57.9**	**32.9-90.7**	**173**	**55.4**	**33.9-73.5**	**180**	**59.0**	**32.2-84.1**

1.6 Median Body Mass Index (BMI) values among adults (20 years and over)
n=420

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	88	21.4	18.8-23.4	48	21.2	18.4-23.4	40	21.4	19.0-23.5
24-27	94	21.8	19.9-24.1	42	20.2	18.9-24.0	52	22.1	21.0-25.3
28-31	60	21.9	20.0-24.2	30	21.9	20.2-25.0	30	21.4	19.8-24.2
32-35	56	22.9	20.5-24.4	25	22.6	19.2-25.0	31	23.2	21.1-24.2
36-39	35	22.7	21.3-26.6	11	21.6	20.6-22.7	24	23.6	21.8-27.7
40-44	24	24.8	22.1-27.9	12	24.3	22.0-27.3	12	25.7	22.2-28.7
45-49	27	23.9	21.7-26.1	13	23.7	21.4-27.2	14	24.2	22.0-25.6
50+	36	25.7	22.3-29.0	18	26.1	23.1-29.8	18	24.9	21.1-28.3
Overall	**420**	**22.3**	**20.2-25.2**	**199**	**22.1**	**20.0-25.0**	**221**	**22.6**	**20.8-25.4**

[1] Stallings et al, J Am Diet Assoc. 2008;108:832-839

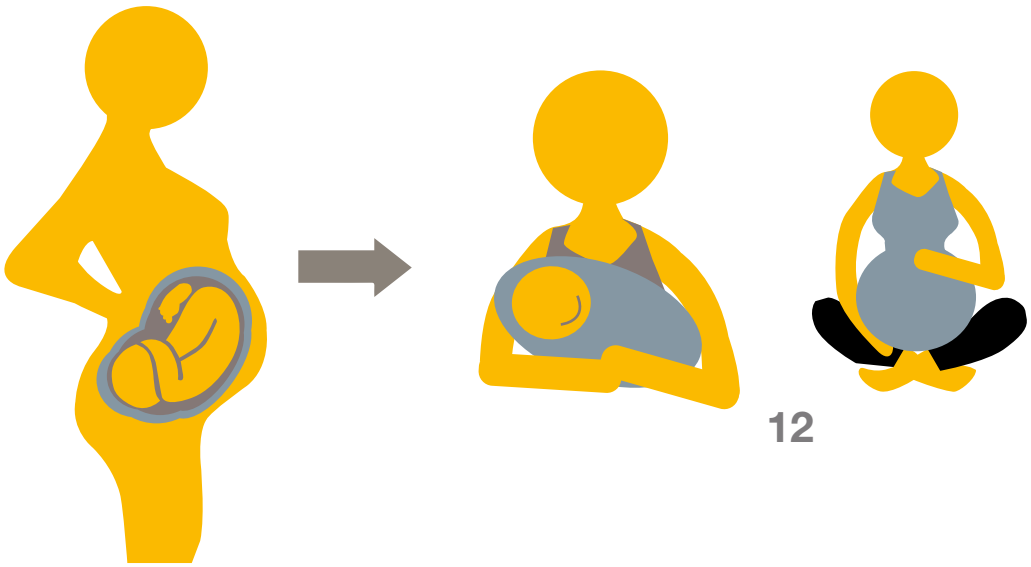
1.7 Education and employment in adults aged 16 years and over n=497

The following table shows how people with cystic fibrosis reported their education and employment status in 2015. Please note that the groups are not mutually exclusive; someone may be a student as well as part-time working, for example.

	Number of patients n (%)
Full-time working	172 (34.6)
Part-time working	76 (15.3)
Student	90 (18.1)
Homemaker	29 (5.8)
Unemployed	91 (18.3)
"Disabled"	15 (3.0)
Retired	11 (2.2)
Unknown	22 (4.4)
No data	9 (1.8)
Number of people in work or study	**326 (70%)**
Number of people who completed questionnaire	**466 (93%)**

Pregnancy

In 2015, 12 women with cystic fibrosis in Scotland had either had a live birth, or were still pregnant at the time of reporting.



1.8 Age at Diagnosis in children under 16
n= 298*

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 2015; n (%)	Patients aged 5 years in 2015; n (%)
Pre-natal	<5	<5	<5
Birth-3months	253 (85.2)	13 (81.3)	24 (96.0)
4-6 months	8 (2.7)	<5	<5
7-12 months	7 (2.4)	<5	-
1 yr	9 (3.0)	<5	-
2 yrs	6 (2.0)	-	-
3 yrs	<5	-	-
4 yrs	<5	-	-
5 yrs	<5	-	-
6 yrs	<5	-	-
7 yrs	<5	-	-
8 yrs	<5	-	-
9 yrs	<5	-	-
10 yrs	-	-	-
11 yrs	-	-	-
12 yrs	-	-	-
13 yrs	-	-	-
14 yrs	-	-	-
15 yrs	-	-	-
Overall	**297**	**16**	**25**

*n=297 – had exact diagnosis dates

Diagnosis in the first three months of life is more common in children aged 5 years in 2015 (born after the UK-wide newborn screening programme was implemented) than in children aged 10 years in 2015 (born before the UK-wide newborn screening programme was put in place nationally).

There is a delay between newborn screening tests being performed and the results entering the Registry, these statistics are updated retrospectively each year to take updated data into account. Therefore the number of patients identified in 2014 is higher in this report than was recorded in the report published in 2015. It is likely that the 2015 figure will be updated in the next annual report.

1.9 Age at Diagnosis in adults aged 16 years and over in 2015 n=497

The table below shows the age of people aged 16 and over in 2014 when they were diagnosed. These people were born before newborn screening was done routinely across the UK. There were some regions with newborn screening prior to 2007

Age at diagnosis	n (%)
Pre-natal	0 (0)
Birth-3months	181 (36.4)
4-6 months	49 (9.9)
7-12 months	30 (6.0)
1 yr	37 (7.4)
2 yrs	33 (6.6)
3 yrs	24 (4.8)
4 yrs	13 (2.6)
5 yrs	10 (2.0)
6 yrs	8 (1.6)
7 yrs	7 (1.4)
8 yrs	6 (1.2)
9 yrs	<5
10 yrs	<5
11 yrs	5 (1.0)
12 yrs	6 (1.2)
13 yrs	<5
14 yrs	5 (1.0)
15 yrs	5 (1.0)
16-20 yrs	15 (3.0)
21-25 yrs	6 (1.2)
26-30 yrs	17 (3.4)
31-35 yrs	9 (1.8)
36-40 yrs	6 (1.2)
41-45 yrs	<5
46-50 yrs	<5
51-60 yrs	10 (2.0)
61 yrs+	-
Overall	**497**

Lung health

For people with cystic fibrosis mucus in the lungs is linked to repeat or chronic, which can cause permanent damage, making it harder to breathe.

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.

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.

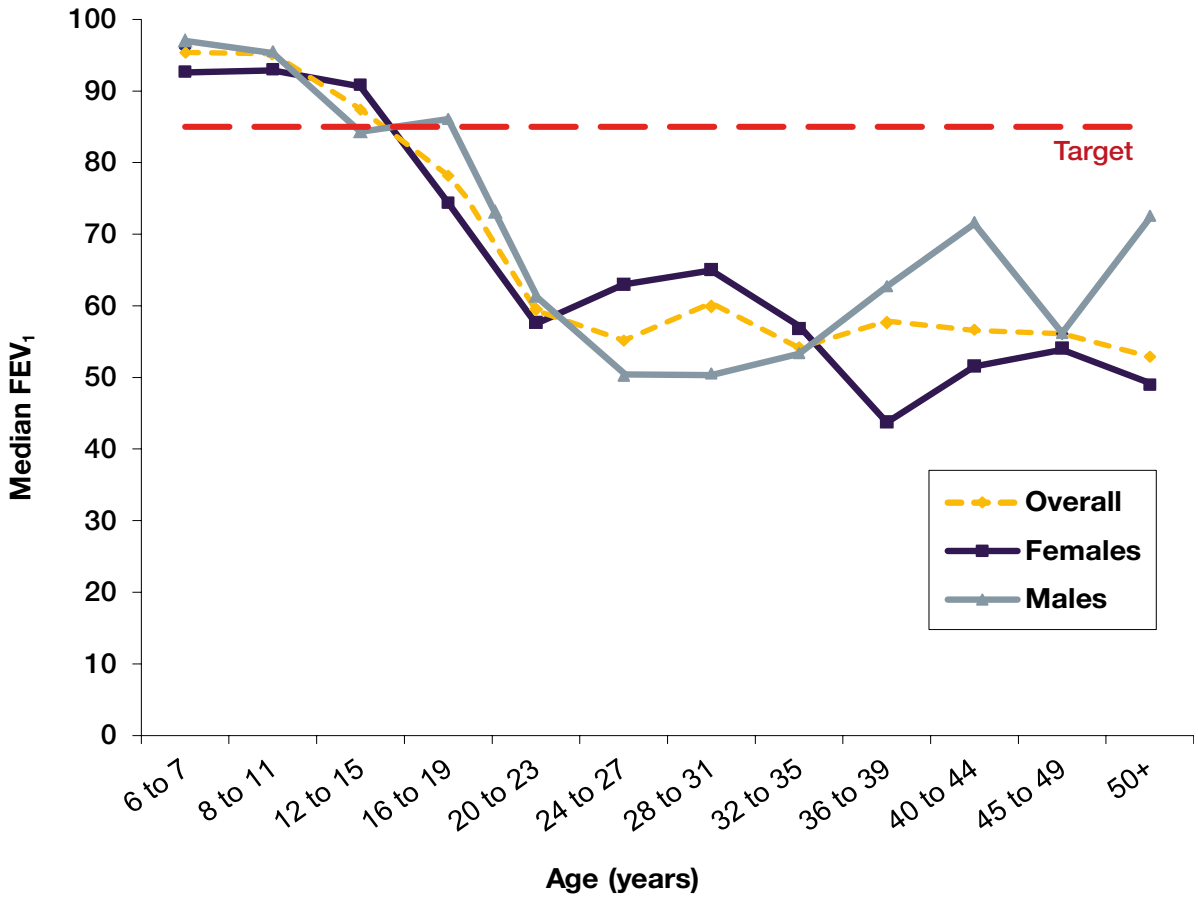
This is often a team effort between people with CF, their family, and their medical team, which can include doctors, nurses, physiotherapists, dieticians, and psychologists.

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, GLI equations) among patients aged 6 years and over, excluding patients post lung transplant

n=620

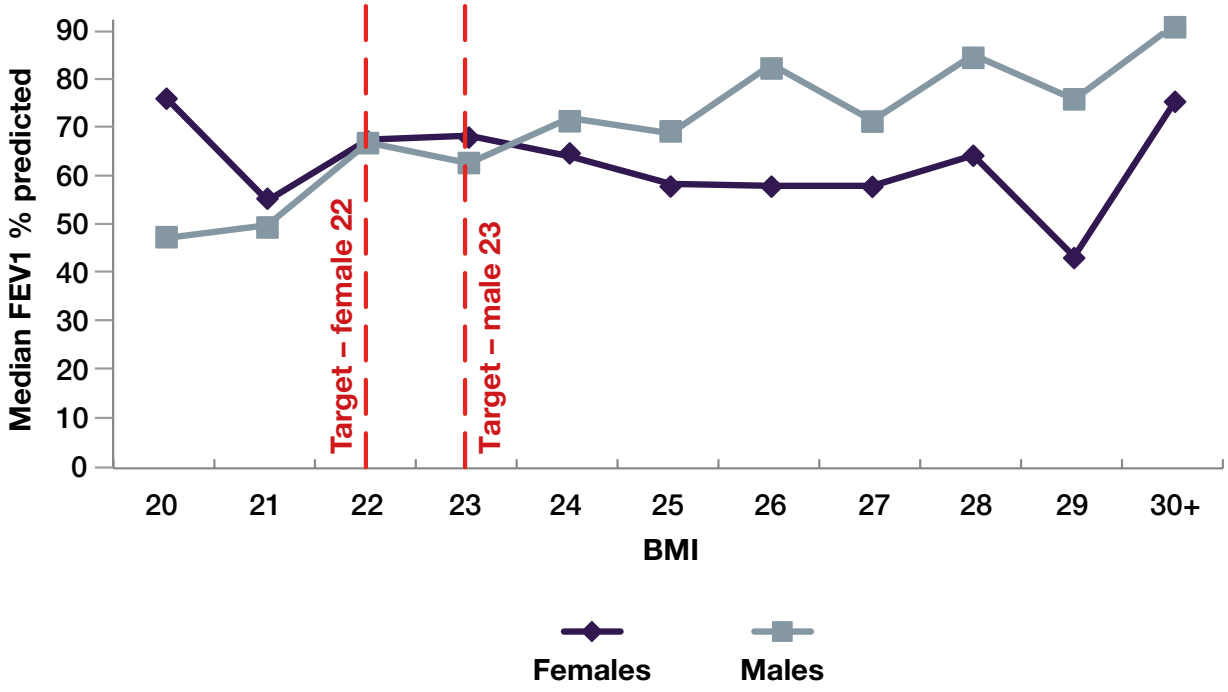
The chart and table in this section show the information about those patients whose FEV_1 data were complete. People with CF who have had lung transplants are excluded, as their new 'non-CF' lungs would have lung health similar to a person without CF.



	Overall			Female			Male		
Age (yrs)	N	Median	IQR	N	Median	IQR	N	Median	IQR
6-7	35	95.6	83.8-103.6	20	92.7	83.5-108.4	15	96.6	86.8-102
8-11	88	95.1	88.7-100.5	40	93.2	88.4-100.4	48	95.5	89.3-101.0
12-15	57	87.6	73.1-100.3	34	90.9	75.0-102.6	23	84.5	71.1-100.2
16-19	76	78.3	60.2-94.5	31	74.5	35.5-88.8	45	86.0	66.5-97.1
20-23	83	59.8	40.3-76.8	47	58.2	43.5-75.6	36	61.4	35.7-81.1
24-27	83	55.1	41.9-76.5	37	63.7	49.2-77.6	46	50.6	40.6-71.6
28-31	55	60.3	34.4-74.9	25	65.1	44.1-78.3	30	50.6	31.7-74.7
32-35	42	54.0	39.2-72.9	19	57.1	43.2-82.6	23	53.5	34.4-71.7
36-39	28	57.7	36.4-84.5	10	44.1	30.9-77.2	18	62.5	41.2-86.7
40-44	19	56.5	45.1-90.2	10	51.7	40.4-58.9	9	71.3	56.4-96.6
45-49	24	55.9	43.4-72.0	12	54.3	44.2-71.6	12	56.1	41.1-72.0
50+	30	52.8	42.2-81.4	15	49.4	42.3-69.7	15	72.5	42.1-85.6
Overall	**620**	**52.8**	**42.2-81.4**	**300**	**72.8**	**51.4-92.1**	**320**	**72.7**	**48.0-92.5**

1.11 Median FEV_1 % predicted and BMI among patients aged 16 years and older (excluding patients post-lung transplant) n=448

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.

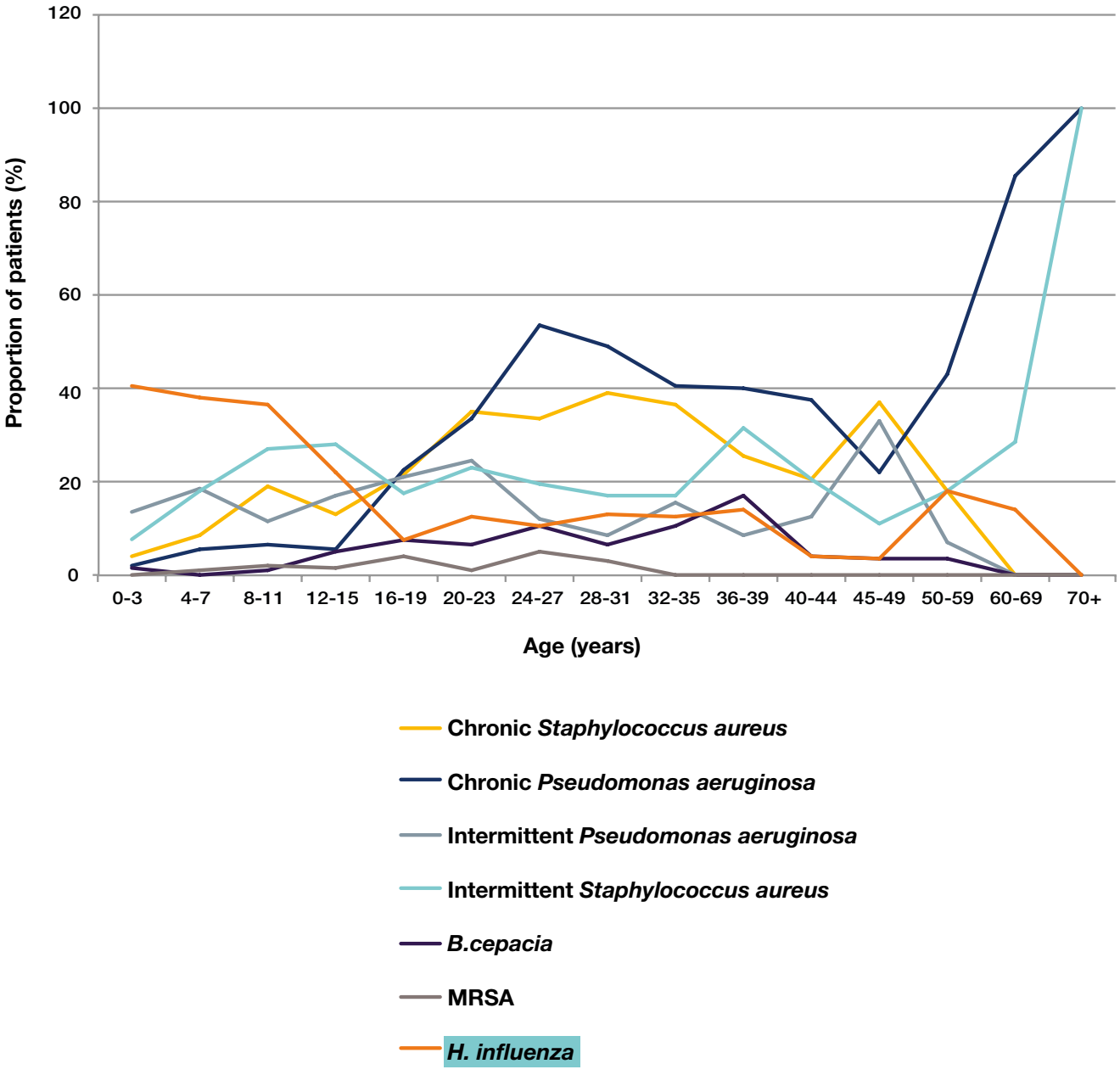


Each point represents the median FEV_1% predicted of patients for each given BMI value. Due to the wide range of BMIs in this population we grouped all BMI≤16 into one group and BMI≥26 into another.

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.

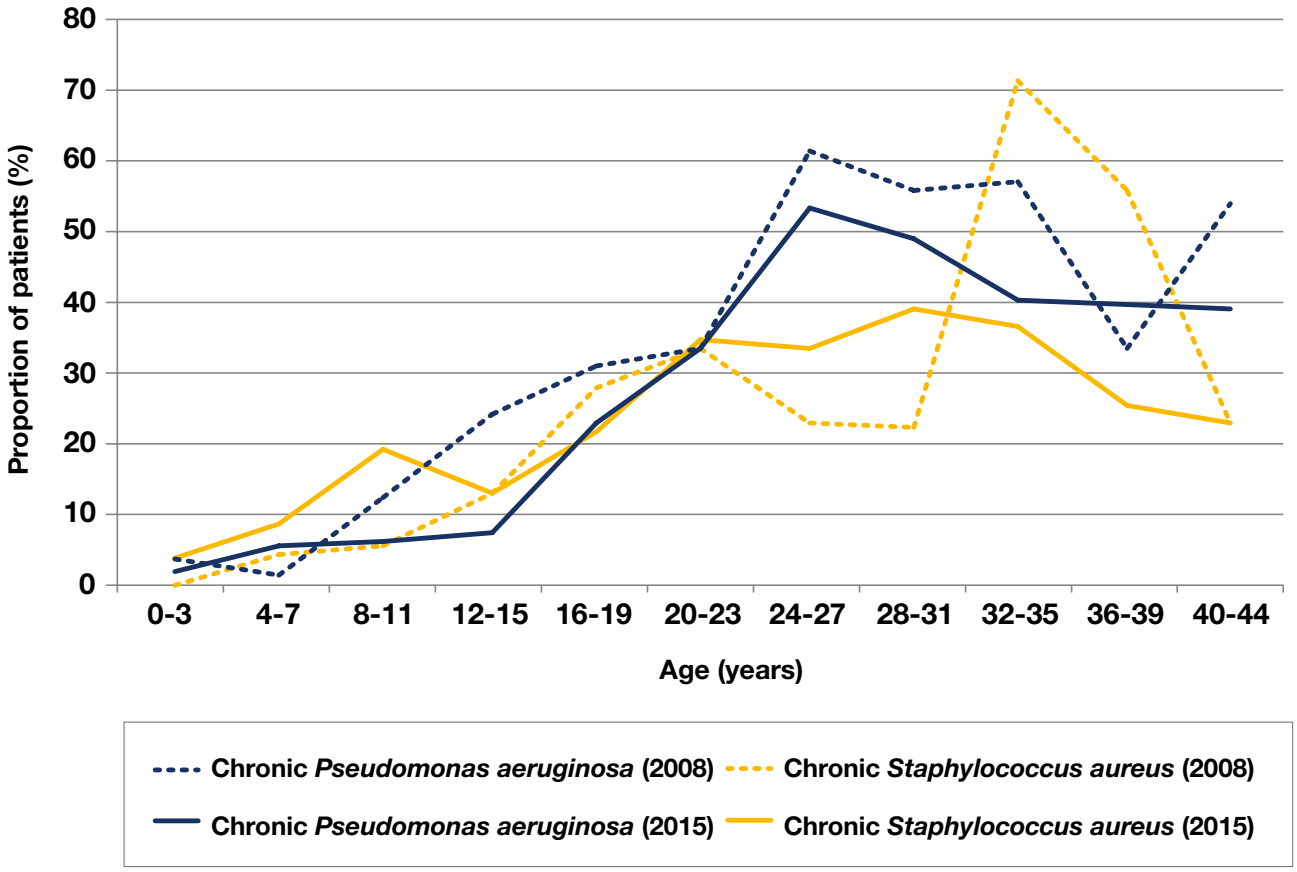
1.12 Lung infections in 2015



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+	Children (<16 years)	Adults (≥16 years)
N patients in age bands	59	87	93	59	77	88	94	60	56	35	24	27	36	298	497
Chronic S.aureus; n(%)	2 (3.9)	6 (8.5)	15 (19.2)	7 (13.2)	16 (21.6)	30 (34.9)	31 (33.7)	23 (39)	19 (36.5)	9 (25.7)	5 (20.8)	10 (37)	5 (13.9)	30 (11.8)	148 (30.5)
Chronic P.aeruginosa; n(%)	1 (2.0)	4 (5.7)	5 (6.4)	4 (7.4)	17 (22.7)	29 (33.7)	49 (53.3)	29 (49.2)	21 (40.4)	14 (40)	9 (37.5)	6 (22.2)	19 (52.8)	14 (5.6)	193 (39.7)
Intermittent P.aeruginosa; n(%)	7 (13.7)	13 (18.6)	9 (11.5)	9 (17.0)	16 (21.3)	21 (24.4)	11 (12.0)	5 (8.5)	8 (15.4)	3 (8.6)	3 (12.5)	9 (33.3)	2 (5.6)	38 (15.1)	78 (16.1)
Intermittent S.aureus; n(%)	4 (7.7)	13 (18.3)	21 (26.9)	15 (28.3)	13 (17.6)	20 (23.3)	18 (19.6)	10 (17.0)	9 (17.3)	11 (31.4)	5 (20.8)	3 (11.1)	8 (22.2)	53 (20.9)	97 (20.0)
B.cepacia; n(%)	1 (1.69)	0 (0)	1 (1.1)	3 (5.1)	6 (7.8)	6 (6.8)	10 (10.6)	4 (6.7)	6 (10.7)	6 (17.1)	1 (4.2)	1 (3.7)	1 (2.8)	5 (1.7)	41 (8.3)
MRSA; n(%)	0 (0)	1 (1.2)	2 (2.2)	1 (1.7)	3 (3.9)	1 (1.1)	5 (5.3)	2 (3.3)	0 (0)	0 (0)	0 (0)	0 (0)	0 (0)	4 (1.3)	11 (2.2)
H. influenza; n(%)	24 (40.7)	33 (37.9)	34 (36.6)	13 (22.0)	6 (7.8)	11 (12.5)	10 (10.6)	8 (13.3)	7 (12.5)	5 (14.3)	1 (4.2)	1 (3.7)	6 (16.7)	104 (34.9)	55 (11.1)

1.13 Lung infections over time



	Age (years)										
%	**0-3**	**4-7**	**8-11**	**12-15**	**16-19**	**20-23**	**24-27**	**28-31**	**32-35**	**36-39**	**40-44**
Chronic S.aureus; 2015	3.9	8.5	19.2	13.2	21.6	34.9	33.7	39	36.5	25.7	23
Chronic P.aeruginosa; 2015	2	5.7	6.4	7.6	22.7	33.7	53.3	49.2	40.4	40	39.1
Chronic S.aureus; 2008	0	4.5	5.5	13.3	28	33.3	23.1	22.2	71.4	55.6	23.1
Chronic P.aeruginosa; 2008	3.6	1.5	12.7	24	30.8	33.3	61.5	55.6	57.1	33.3	53.9

1.14 Prevalence of complications

	Overall (n=795)	<16 years (n=298)	≥16 years (n=497)
	N (%)	N (%)	N (%)
Respiratory Related			
Nasal polyps requiring surgery	10 (1.3)	<5	7 (1.4)
Sinus disease	200 (25.2)	<5	197 (39.6)
Asthma	99 (12.5)	19 (6.4)	80 (16.1)
ABPA	55 (6.9)	12 (4.0)	43 (8.7)
Haemoptysis	15 (1.9)	<5	14 (2.8)
Pneumothorax requiring chest tube	<5	<5	<5
Nontuberculous mycobacteria or atypical mycobacteria	37 (4.7)	11 (3.7)	26 (5.2)
Pancreas & Hepatobiliary Disease			
Liver enzymes	19 (2.4)	8 (2.7)	11 (2.2)
Liver disease	127 (16.0)	36 (12.1)	91 (18.3)
Cirrhosis with no portal hypertension	<5	<5	<5
Cirrhosis with portal hypertension	15 (1.9)	<5	11 (2.2)
Gallbladder disease requiring surgery	6 (0.8)	0 (0.0)	6 (1.2)
Pancreatitis	7 (0.9)	<5	6 (1.2)
GI bleed req hosp variceal	0 (0.0)	<5	0 (0.0)
Upper Gastrointestinal			
GERD	137 (17.2)	<5	135 (27.2)
Peptic ulcer	0 (0.0)	0 (0.0)	0 (0.0)
GI bleed req hosp non variceal	0 (0.0)	0 (0.0)	0 (0.0)
Lower Gastrointestinal			
Intestinal obstruction	105 (13.2)	11 (3.7)	94 (18.9)
Fibrosing colonopathy/ colonic structure	0 (0.0)	0 (0.0)	0 (0.0)
Rectal prolapse	<5	<5	<5
Meconium ileus	118 (14.8)	47 (15.8)	71 (14.3)
Renal			
Kidney stones	<5	<5	<5
Renal failure	9 (1.1)	0 (0.0)	9 (1.8)
Muscolo-Skeletal			
Arthritis	5 (0.6)	<5	<5
Arthropathy	38 (4.8)	<5	36 (7.2)
Bone fracture	<5	<5	0 (0.0)
Osteopenia	95 (11.9)	0 (0.0)	95 (19.1)

Osteoporosis	42 (5.3)	0 (0.0)	42 (8.5)
Other			
Cancer confirmed by histology	<5	<5	<5
Port inserted or replaced	17 (2.1)	9 (3.0)	8 (1.6)
Depression	28 (3.5)	0 (0.0)	28 (5.6)
Hearing loss	12 (1.5)	<5	9 (1.8)
Hypertension	26 (3.3)	0 (0.0)	26 (5.2)

1.15 Incidence of complications

	Newly identified in 2014			Newly identified in 2015		
	Overall (n=782)	**<16 years (n=297)**	**≥16 years (n=485)**	**Overall (n=795)**	**<16 years (n=298)**	**≥16 years (n=497)**
ABPA; n(%)	11 (1.4)	<5	10 (2.1)	7 (0.9)	<5	<5
Cirrhosis with no portal hypertension; n(%)	<5	<5	<5	<5	0	<5
Cirrhosis with portal hypertension; n(%)	6 (0.8)	<5	5 (1.0)	14 (1.8)	<5	10 (2.0)
Cancer confirmed by histology *; n(%)	<5	0	<5	<5	0	<5

1.15a Incidence of NTM

	2014 n=782	2015 n=795
NTM Prevalence (%)	31 (4.0)	37 (4.7)
On NTM treatment in the given year (%) of total NTM prevalence	8 (25.8)	12 (32.4)
NTM Incidence (%)	<5	5 (3.5)
M.abscessus complex incidence (%)	<5	<5

1.16 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.

	All ≥ 10 years (n=597)	10-16 years (n=100)	≥16 years (n=497)
Treatment for CF-related diabetes; n(%)	142 (23.8)	<5	138 (2.4)
Screening for CF-related diabetes			
Yes	335 (56.1)	77 (77.0)	258 (4.5)
No	96 (16.1)	16 (16.0)	80 (1.4)
Known CF-related diabetes	141 (23.6)	<5	137 (2.4)
Unknown	15 (2.5)	<5	13 (0.2)

1.17 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.

	2011	2012	2013	2014	2015
Number of patients that year with annual review data evaluated for transplants	12	13	21	17	19
Number accepted on the transplant list	5	7	11	12	6
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

1.18 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 2015.

Number of patients on ivacaftor in the UK	**58**
	Median (IQR)
Sweat chloride pre	107 (92-117)
Sweat chloride post	60 (33.3-70.8)
Fev_1% pre	52.6 (46.7-60.2)
Fev_1% post	63.8 (51.9-72.3)
Number of patients stopped Ivacaftor	<5

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.

1.19 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.

		Home		Hospital		Total	
Age	N	N (%)	Median (IQR)	N (%)	Median (IQR)	N (%)	Median (IQR)
0-3	59	2 (0.3)	12 (6-17)	16 (2.0)	15 (8-31)	16 (2.0)	17 (11-31)
4-7	87	7 (0.9)	33 (9-46)	23 (2.9)	14 (9-36)	24 (3.0)	28 (14-52)
8-11	93	11 (1.4)	17 (10-46)	30 (3.8)	14 (7-36)	30 (3.8)	28 (14-53)
12-15	59	11 (1.4)	13 (10-51)	25 (3.1)	19 (7-62)	25 (3.1)	56 (15-75)
16-19	77	15 (1.9)	35 (14-49)	16 (2.0)	14 (7-21)	23 (2.9)	20 (14-56)
20-23	88	30 (3.8)	14 (12-30)	29 (3.6)	14 (7-30)	38 (4.8)	28 (14-52)
24-27	94	32 (4.0)	14 (13-28)	26 (3.3)	13 (8-42)	39 (4.9)	28 (14-50)
28-31	60	24 (3.0)	45 (13-57)	21 (2.6)	9 (5-35)	30 (3.8)	46 (14-66)
32-35	56	14 (1.8)	14 (13-27)	11 (1.4)	14 (4-28)	18 (2.3)	19 (14-34)
36-39	35	12 (1.5)	28 (14-42)	8 (1.0)	12 (6-21)	13 (1.6)	28 (14-65)
40-44	24	7 (0.9)	28 (14-42)	3 (0.4)	5 (2-7)	8 (1.0)	21 (14-42)
45-49	27	5 (0.6)	21 (14-40)	3 (0.4)	14 (14-38)	6 (0.8)	25 (14-57)
50+	36	7 (0.9)	14 (12-20)	8 (1.0)	19 (10-27)	11 (1.4)	14 (14-40)
Overall	**795**	**177 (22.3)**	**14 (13-42)**	**219 (27.5)**	**14 (7-33)**	**281 (35.3)**	**28 (14-54)**

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.

1.20 Inhaled Antibiotic use among patients with chronic Pseudomonas aeruginosa

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.

	2008			2015		
	Overall	<16 years	≥16 years	Overall	<16 years	≥16 years
Patients with chronic pseudomonas	67	28	39	207	14	193
Tobramycin solution; n(%)	6 (9.0)	<5	<5	33 (15.9)	5 (35.7)	28 (14.5)
Other aminoglycoside; n(%)	0	0	0	<5	0	<5
Colistin; n(%)	21 (31.3)	15 (53.6)	6 (15.4)	64 (30.9)	8 (57.1)	56 (29.0)
Promixin; n(%)	<5	0	<5	37 (17.9)	<5	34 (17.6)
Aztreonam; n(%)*	-	-	-	<5	0	<5
Colistimethate(DPI); n(%)*	-	-	-	34 (16.4)	<5	33 (17.1)
Tobramycin Inhalation Powder; n(%)*	-	-	-	59 (28.5)	<5	58 (30.1)
At least one of the above; n(%)*	-	-	-	163 (78.7)	12 (85.7)	151 (78.2)

*Not reported for 2008

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.

1.21 DNase

	2015		2015		
Age	**Total patients**	**Patients on Dnase**	**Total patients**	**Patients on Dnase**	**p-value (2008 vs 2015)**
0-3	91	<5	59	5 (8.5)	0.314
4-7	78	9 (11.5)	87	18 (20.7)	0.169
8-11	66	15 (22.7)	93	43 (46.2)	0.004
12-15	92	33 (35.9)	59	50 (50.9)	<0.001
16-19	32	12 (37.5)	77	36 (40.9)	0.500
20-23	14	<5	88	45 (47.9)	0.005
24-27	14	<5	94	19 (31.7)	0.420
28-31	11	<5	60	14 (25.0)	1.000
32-35	8	<5	56	10 (28.6)	1.000
36-39	9	<5	35	<5	1.000
40+	14	5 (35.7)	87	12 (44.4)	0.795
Overall	**429**	**83 (19.4)**	**795**	**286 (36.0)**	

1.22 Hypertonic saline

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

Age	Hypertonic saline; n (%)				p-value (2008 vs 2015)
	2008		2015		
	Number of patients	Patients on hypertonic saline	Number of patients	Patients on hypertonic saline	
0-3	91	0 (0)	59	<5	n/a
4-7	78	0 (0)	87	11 (12.6)	n/a
8-11	66	<5	93	18 (19.4)	<0.001
12-15	92	<5	59	19 (32.2)	<0.001
16-19	32	0 (0)	77	19 (24.7)	0.002
20-23	14	0 (0)	88	20 (22.7)	n/a
24-27	14	0 (0)	94	15 (16.0)	n/a
28-31	11	<5	60	12 (20.0)	0.400
32-35	8	0 (0)	56	6 (10.7)	n/a
36-39	9	0 (0)	35	<5	n/a
40+	14	0 (0)	87	9 (10.34)	n/a
Overall	429	<5	795	286 (36.0)	<0.001

1.23 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				2015			
	Overall (n=429)	0-3 years (n=91)	4-15 years (n=236)	≥16 years (n=102)	Overall (n=795)	0-3 years (n=59)	4-15 years (n=239)	≥16 years (n=497)
Patients with chronic P.aeruginosa	29 (43.3)	0 (0)	12 (46.2)	17 (43.6)	176 (44.6)	0 (0)	10 (18.5)	166 (48.8)
Patients without chronic P.aeruginosa	34 (12.4)	<5	22 (12.9)	11 (22.0)	219 (55.4)	<5	44 (81.5)	174 (51.2)

1.24 Physiotherapy

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

	Overall (n=795)	<16 years (n=298)	≥16 years (n=497)
Active cycle of breathing techniques; n(%)	218 (27.8)	70 (23.6)	148 (30.4)
Autogenic drainage (including assisted autogenic drainage); n(%)	289 (36.9)	77 (25.9)	212 (43.5)
Any form of PEP; n(%)	414 (52.8)	237 (79.8)	177 (36.3)
VEST; n(%)	<5	<5	<5
Exercise; n(%)	262 (33.0)	61 (20.5)	201 (40.4)

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

1.25 Other therapy

	Overall (n=795)	<16 years (n=298)	≥16 years (n=497)
Non Invasive Ventilation (NIV); n (%)	9 (1.1)	<5	5 (1.0)
Long-term oxygen; n (%) Among those who have long-term oxygen:	38 (4.8)	8 (2.7)	30 (6.0)
Continuously	7 (0.9)	<5	6 (1.2)
Nocturnal or with exertion	11 (1.4)	<5	10 (2.0)
As required (PRN)	<5	0 (0)	<5
With exacerbation	19 (2.4)	6 (2.0)	13 (2.6)

1.26 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=795)	<16 years (n=298)	≥16 years (n=497)
Any supplemental feeding; n(%)	201 (25.3)	61 (20.5)	140 (28.2)
Nasogastric Tube	13 (1.6)	<5	10 (2.0)
Gastrostomy Tube / Button	42 (5.3)	15 (5.0)	27 (5.4)
Jejunal	0 (0)	0 (0)	0 (0)
TPN	0 (0)	0 (0)	0 (0)

1.27 Age distribution of deaths in 2015

There were 11 recorded deaths in 2015.
The median age at death was Median 28 years range (22.5-42.0)

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.

784 (98.62%) patients have been genotyped with a recorded value.

DF508 Mutations; n (%)	
Homozygous DF508	**349 (44.52)**
Heterozygous DF508	**365 (46.56)**
No DF508 or both unidentified	**70 (8.93)**

1.28 Genotypes

Mutations		
All mutations Current Name	**N**	**(%)**
^F508	714	91.07
Other	149	19.01
G551D	81	10.33
R117H	56	7.14
G542X	53	6.76
R560T	17	2.17
1717-1G->A	14	1.79
Q493X	13	1.66
Not Identified	10	1.28
621+1G->T	10	1.28
N1303K	8	1.02
3659delC	8	1.02
3849+10kbC->T	7	0.89
V520F	7	0.89
D1152H	6	0.77
2789+5G->A	6	0.77
E60X	5	0.64
^I507	<5	-
A455E	<5	-
2143delT	<5	-
1898+1G->A	<5	-
G85E	<5	-
W1282X	<5	-
2184delA	<5	-
R1162X	<5	-
R553X	<5	-

Section 2: Centre Level Analysis

Cystic fibrosis care in 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	2	0	2

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 44.

Key

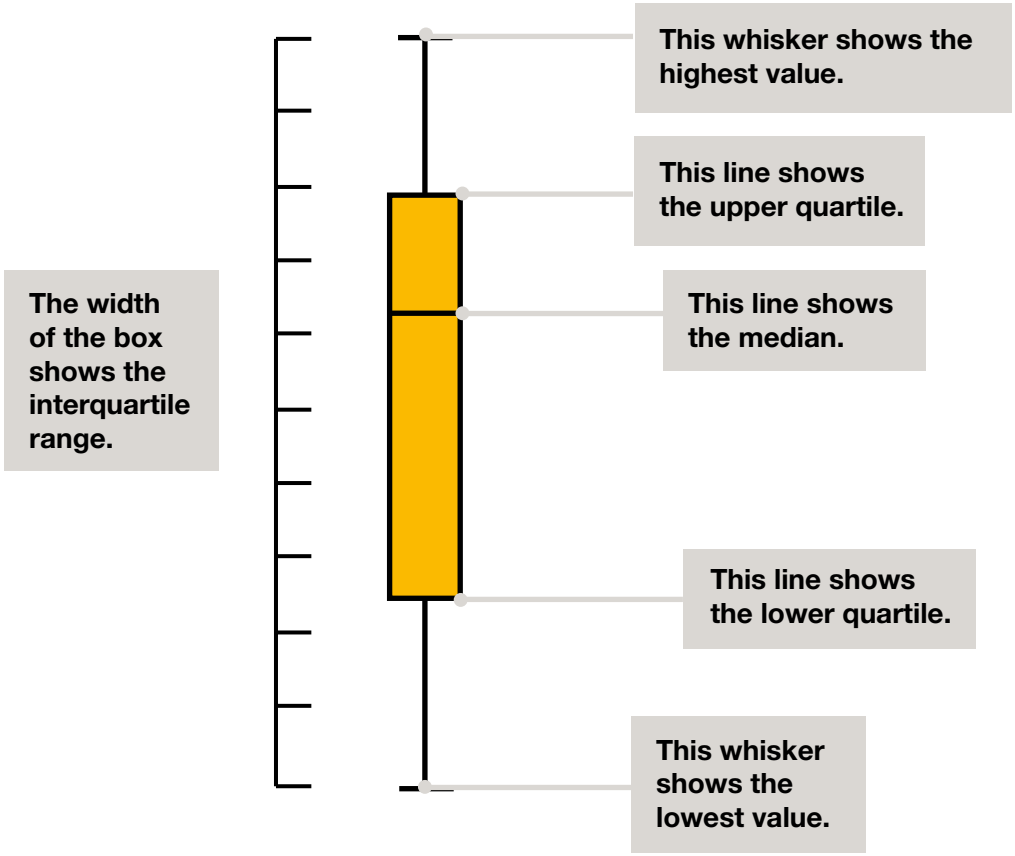
 Paediatric centre

 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.

Box plots



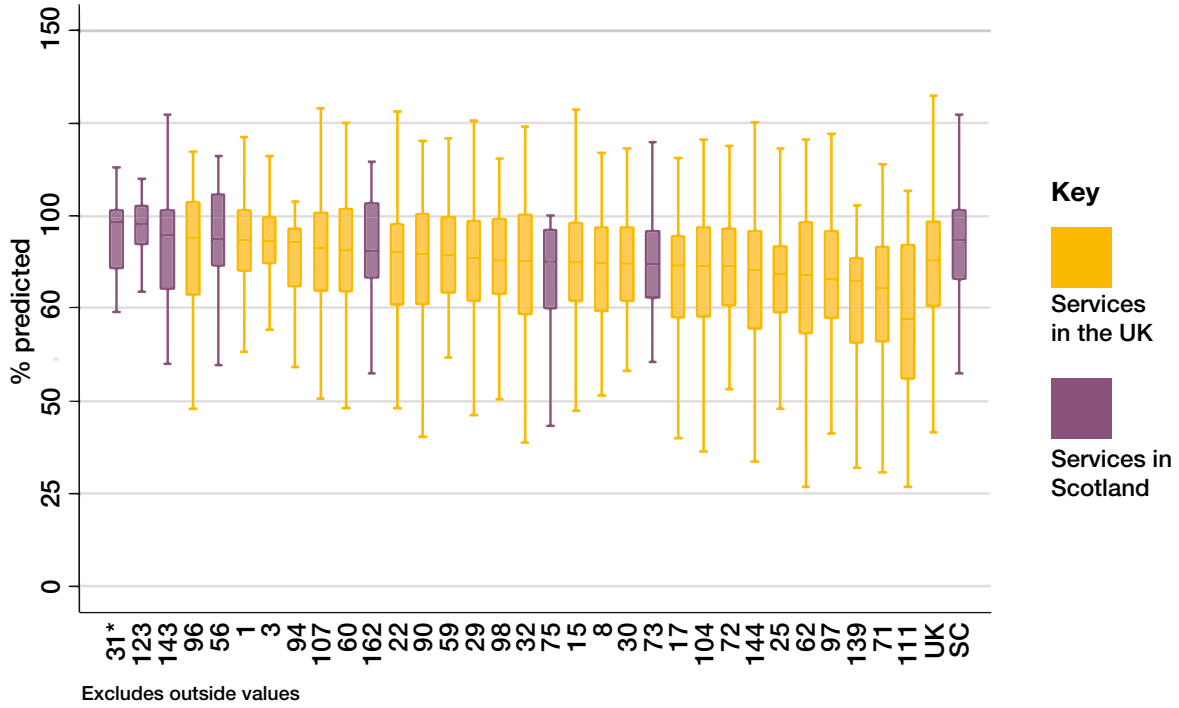
- The ‘box’ shows the middle half of the data for that centre, going from the first quartile to the third quartile. The longer the box, the more varied the data for that centre.
- The horizontal line within the box shows the median result for that centre.
- The ‘whiskers’ above and below the box shows 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=312

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

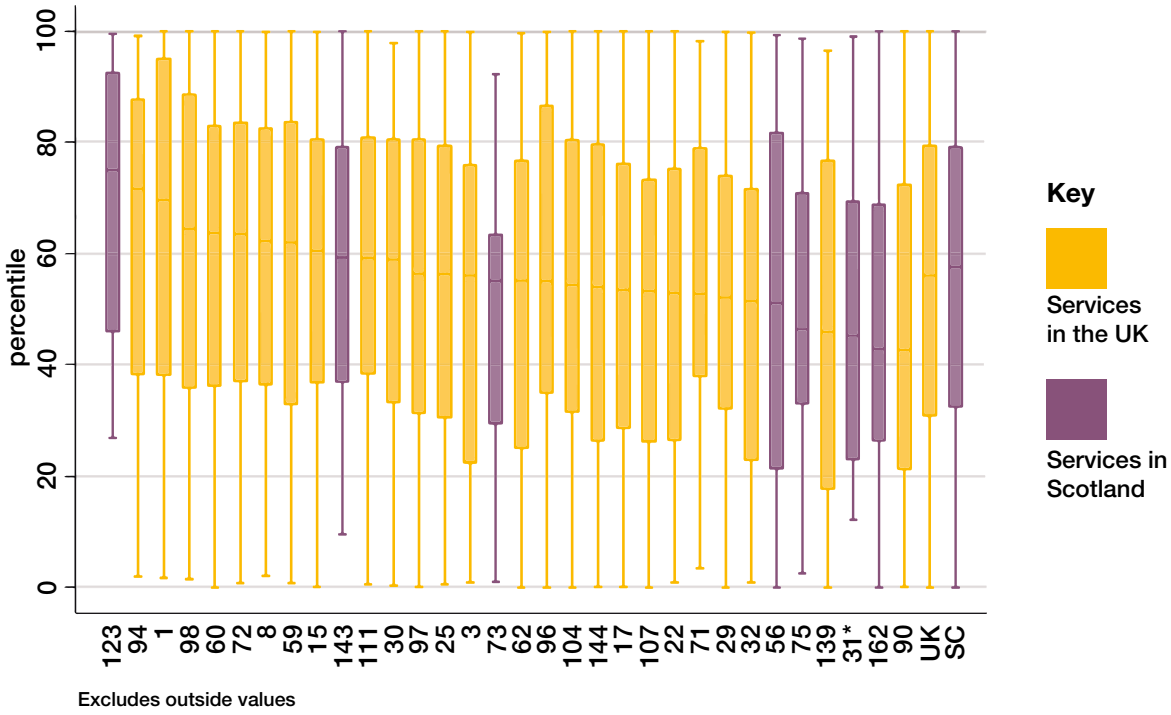
2.1 Median FEV_1 % predicted among patients aged 6 and older by paediatric centre/clinic (without a history of lung transplant) (GLI equations)



The median FEV_1 % predicted of patients attending paediatric centres/clinics in Scotland is 94% predicted (IQR: 82-102).

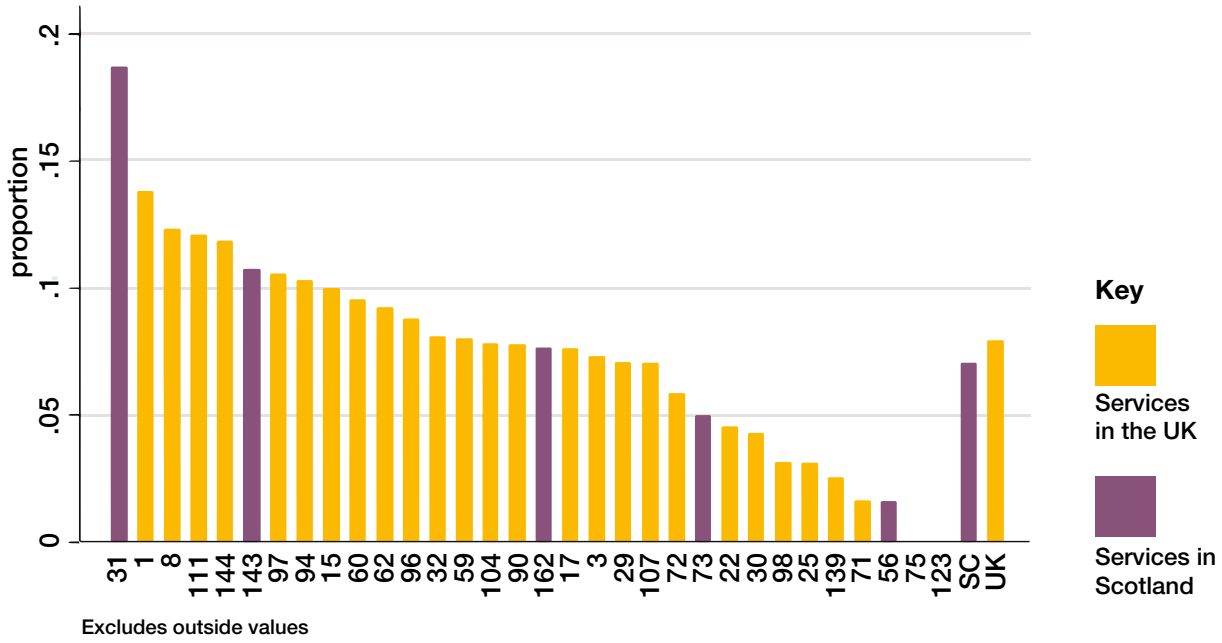
** Centre/clinic with a dataset submission of less than 20 patients.*

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



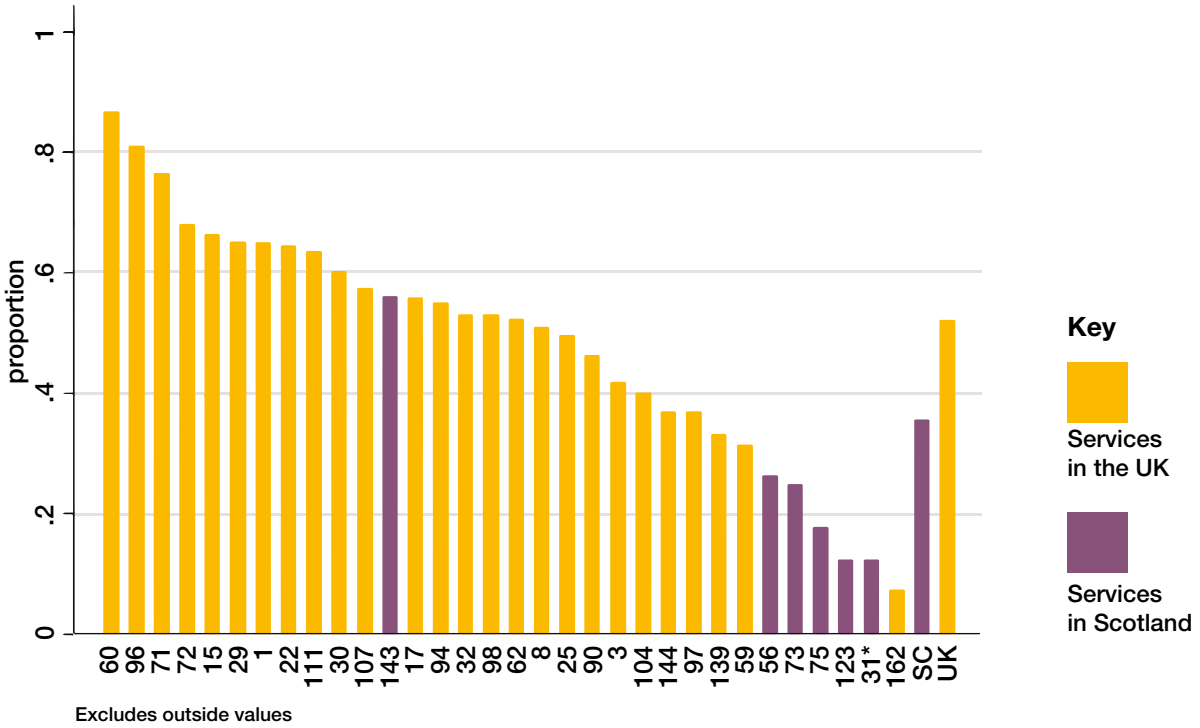
The median BMI percentile of patients attending paediatric centres/clinics in Scotland is 56 (IQR: 33 – 75).
** Centre/clinic with a dataset submission of less than 20 patients*

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.

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



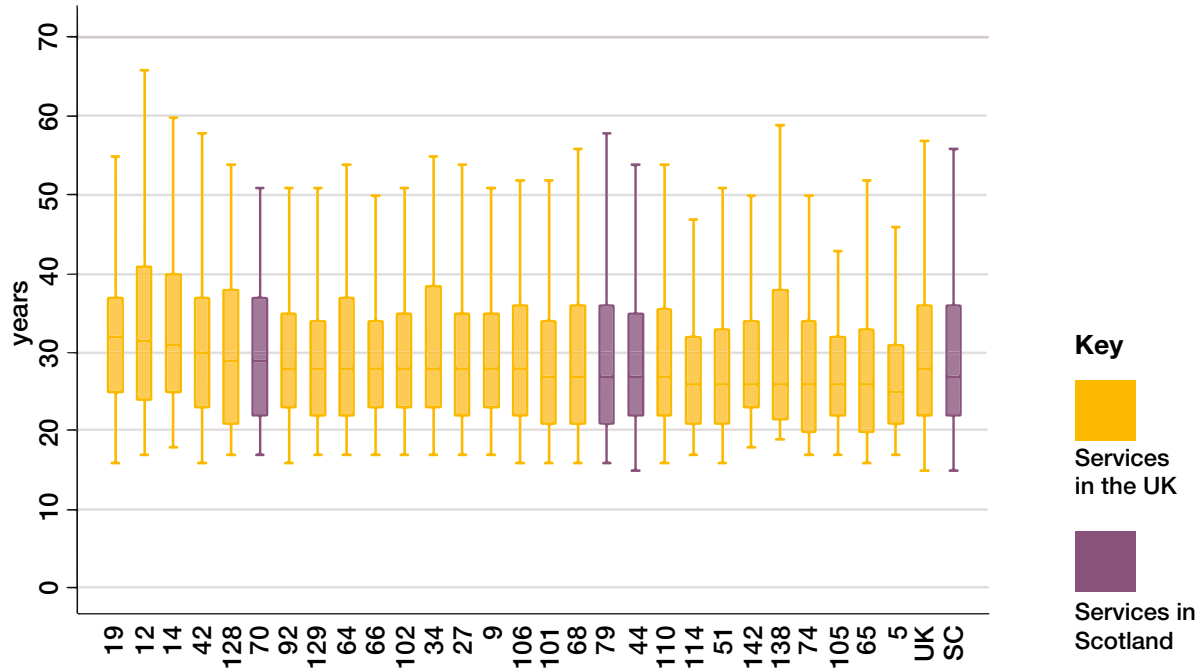
The proportion of patients attending paediatric centres/clinics in Scotland receiving DNase treatment is 34%.
* Centre/clinic with a dataset submission of less than 20 patients.

Section 2b: Adult Centres Analysis

n=483

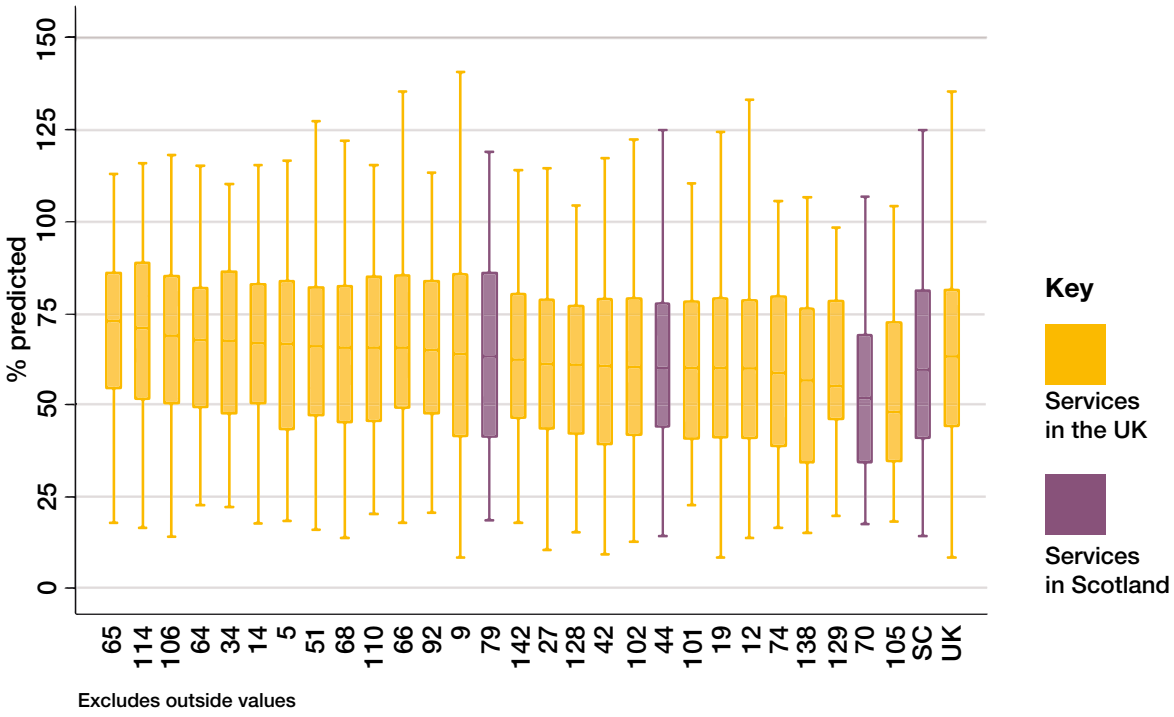
This section shows results for the three adult centres with any network clinics.

2.5 Median age (years) by adult service



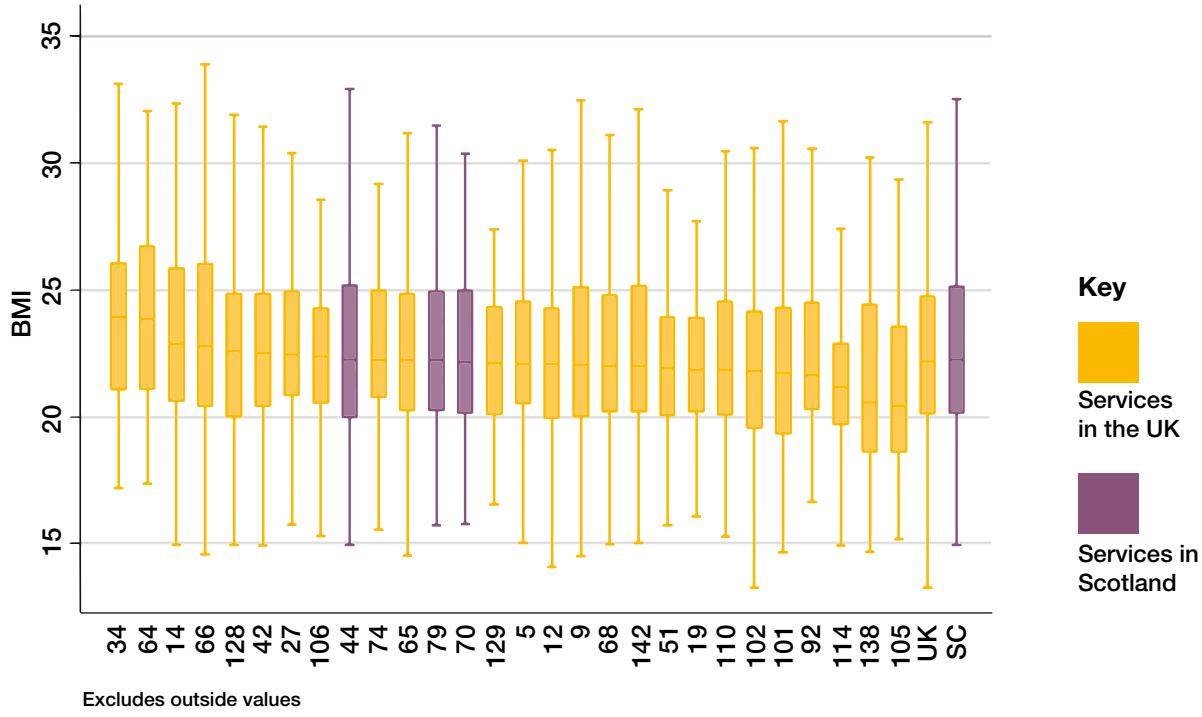
The median age of patients attending adult services in Scotland is 26 years (IQR: 21-35).

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



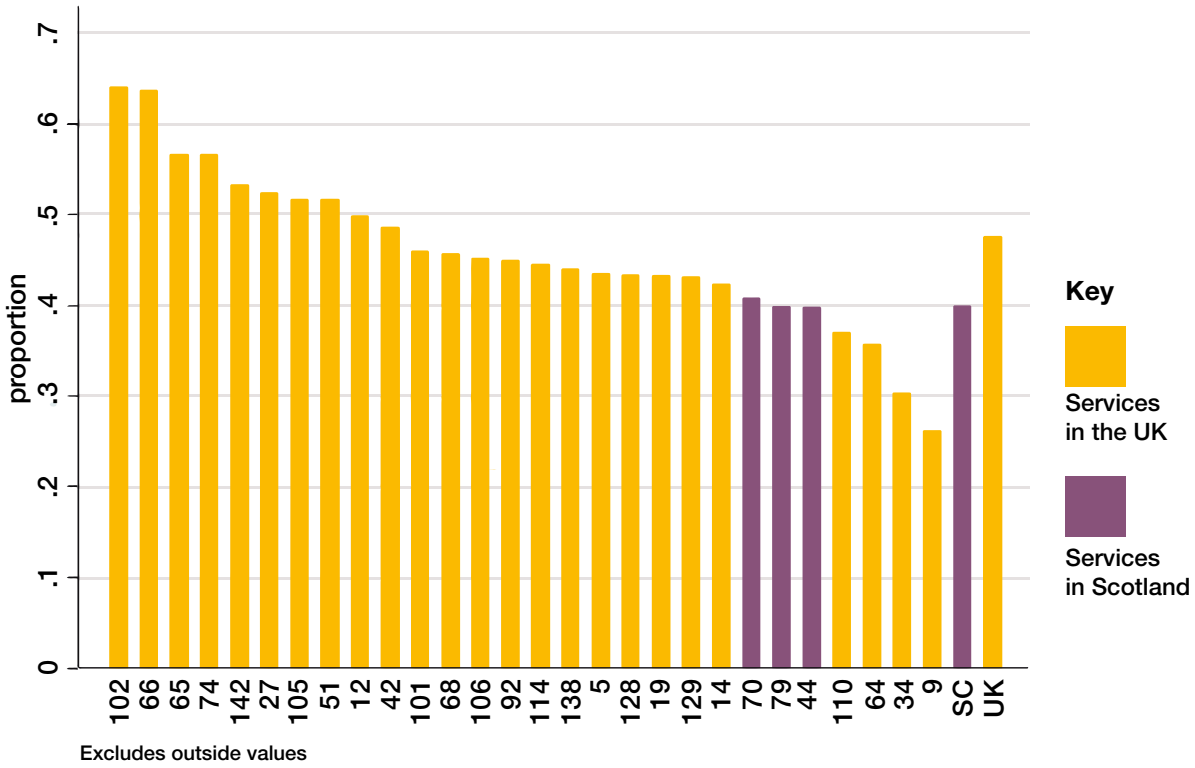
The median FEV_1 (% predicted) of patients attending adult services in Scotland is 60% (IQR: 42 – 83).

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



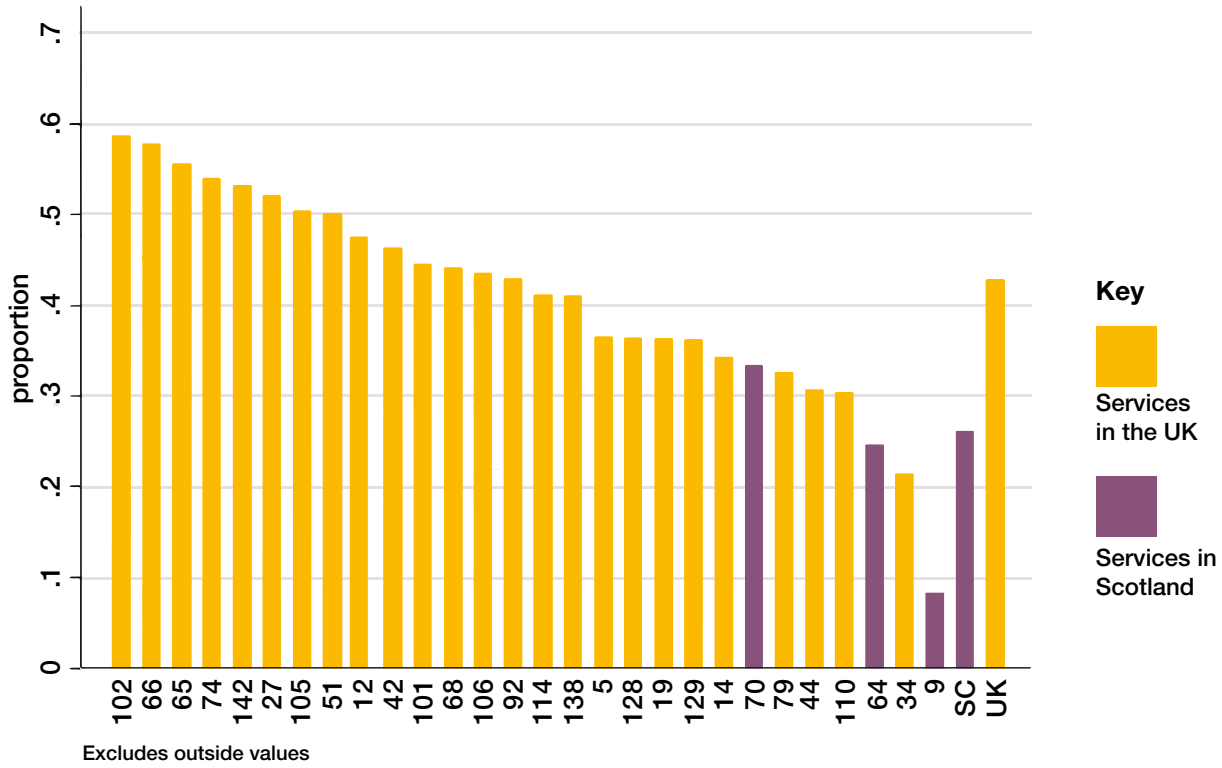
The median BMI of patients attending adult services in Scotland is 22 (IQR: 20 – 25).

2.8 Proportion of patients with chronic P.aeruginosa by adult service



The proportion of patients attending adult services in Scotland with chronic P.aeruginosais 42%.

2.9 Proportion of patients receiving DNase treatment by adult service



The proportion of patients attending adult services in Scotland receiving DNase treatment is 36%.

Appendix 1: Centre level data tables

Paediatric centres/clinics providing data in 2015 – ordered by clinic ID

Country	Location	Centre/ Clinic	Clinic ID	Number of active patients	Number of patients providing data in 2015	Median FEV_1% predicted (≥6 years)	Median BMI percentile (2-15 years)
Scotland	Inverness	Raigmore Hospital	31	17	13	99.2	44.7
Scotland	Glasgow	Royal Hospital for Sick Children	56	89	64	92.8	47.6
Scotland	Ayr	Crosshouse Hospital, Ayr	123	24	23	98.7	72.5
Scotland	Dundee	Ninewells Hospital	73	22	19	84.3	61.1
Scotland	Aberdeen	Aberdeen Royal Infirmary	75	29	23	93.9	72.4
Scotland	Edinburgh	Royal Hospital for Sick Children	143	125	108	93.7	64.8
Scotland	Lanarkshire	Wishaw General Hospital	162	44	32	90.7	48.6

Adult centres/clinics providing data in 2015 – ordered by clinic ID

Country	Location	Centre /Clinic	Clinic ID	Number of active patients	Number of patients providing data in 2015	Median FEV_1% predicted (≥16 years)	Median BMI (≥16 years)
Scotland	Edinburgh	Western General Hospital	44	226	215	61.3	22.2
Scotland	Aberdeen	Aberdeen Royal Infirmary	70	67	67	52.4	22.2
Scotland	Glasgow	Gartnavel General Hospital	79	219	201	63.7	22.2

Appendix 1: Centre level data tables

Paediatric centres/clinics providing data in 2015 – ordered alphabetically

Country	Location	Centre/ Clinic	Clinic ID	Number of active patients	Number of patients providing data in 2015	Median FEV_1% predicted (≥6 years)	Median BMI percentile (2-15 years)
Scotland	Aberdeen	Aberdeen Royal Infirmary	75	29	23	93.9	72.4
Scotland	Ayr	Crosshouse Hospital, Ayr	123	24	23	98.7	72.5
Scotland	Dundee	Ninewells Hospital	73	22	19	84.3	61.1
Scotland	Edinburgh	Royal Hospital for Sick Children	143	125	108	93.7	64.8
Scotland	Glasgow	Royal Hospital for Sick Children	56	89	64	92.8	47.6
Scotland	Inverness	Raigmore Hospital	31	17	13	99.2	44.7
Scotland	Lanarkshire	Wishaw General Hospital	162	44	32	90.7	48.6

Adult centres/clinics providing data in 2015 – ordered alphabetically

Country	Location	Centre /Clinic	Clinic ID	Number of active patients	Number of patients providing data in 2015	Median FEV_1% predicted (≥16 years)	Median BMI (≥16 years)
Scotland	Aberdeen	Aberdeen Royal Infirmary	70	67	67	52.4	22.2
Scotland	Edinburgh	Western General Hospital	44	226	215	61.3	22.2
Scotland	Glasgow	Gartnavel General Hospital	79	219	201	63.7	22.2

Appendix 2: Glossary

Words in this report that appear in this glossary are highlighted the first time they appear.

Word/Phrase	Meaning
2015	1 January 2015 – 31 December 2015.
ABPA (Allergic Bronchopulmonary Aspergillosis)	When a person develops a respiratory allergic reaction to the Aspergillus fumigatus.
Arthritis	A condition causing pain and inflammation in the joints.
Arthropathy	A condition causing pain in the joints.
Asthma	A respiratory condition causing reversible episodes of difficulty breathing often associated with wheeze.
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.
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.

Word/Phrase	Meaning
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).
GERD (Gastroesophageal 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 Function 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.
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 died in any given year. So in 2015 the median age of the 125 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 2015, predicts how long we expect half of them to live for. For 2015, this means that half of people registered as alive on the database are predicted to live to at least 45.1. 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.

Word/Phrase	Meaning
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 have been reported.
Nasal Polyps	Small, sac-like growths of inflamed mucus caused by chronic inflammation of the nasal lining.
Nontuberculous 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.
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 structure

UK CF Registry Steering Committee

Role	Forename	Surname	Organisation
Allied health professional	Alan	Peres	Royal Brompton Hospital
Analytical team rep †	Stephen and/or Vian	Nyangoma Rajabzadeh-Heshejin	Imperial College London
Commissioner, England	Sue or Carrie	Sawyer Gardner	NHS England
Commissioner, Scotland	David	Steele	NHS Scotland
Commissioner, Wales †	Claire	Nelson	NHS Wales
Cystic fibrosis centre data manager	Lance	Dennard	Lewisham Hospital
CF physician - Adults	Caroline	Elston	King's College Hospital
CF physician - Adults	Simon	Range	Glenfield Hospital
CF physician - Paediatrics	Iolo	Doull	Children's Hospital for Wales
CF physician – Paediatrics*	Siobhán	Carr	Royal Brompton Hospital
Chair of the Research Committee	Martin	Wildman	Northern General Hospital
Director of Impact	Keith	Brownlee	Cystic Fibrosis Trust
Parent representative	Grant	Valentine	N/A
Person with CF	James	Thomson	N/A
Registry Data Manager †	Elaine	Gunn	Cystic Fibrosis Trust
Registry Coordinator †	Annie	Jeffery	Cystic Fibrosis Trust
Registry Lead	Rebecca	Cosgriff	Cystic Fibrosis Trust

* Chair
† Non-voting member
# Caldicott guardian

UK CF Registry Research Committee

Role	Forename	Surname	Organisation
Analytical team rep †	Stephen and/or Vian	Nyangoma Rajabzadeh-Heshejin	Imperial College London
Pharmacovigilance PI	Diana	Bilton	Royal Brompton Hospital
CF physician – adults (retired)			
Pharmacovigilance PI	Siobhán	Carr	Royal Brompton Hospital
CF physician - paediatrics			
Pharmacovigilance PI	Steve	Cunningham	Royal Hospital for Sick Children
CF physician - paediatrics			
Registry Lead	Rebecca	Cosgriff	Cystic Fibrosis Trust
Parent representative	Marian	Dmochowska	N/A
Person with CF	Dominic	Kavanagh	Cystic Fibrosis Trust
CF physician - adults*#	Martin	Wildman	Northern General Hospital
Registry consultant	Noreen	Caine	Cystic Fibrosis Trust
Registry Data Manager †	Elaine	Gunn	Cystic Fibrosis Trust
Registry Coordinator †	Annie	Jeffery	Cystic Fibrosis Trust

* Chair
† Non-voting member
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Cystic Fibrosis Trust

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