Cystic Fibrosis strength in numbers

UK Cystic Fibrosis Registry
2016 Annual Data Report

Scotland

Cystic Fibrosis strength in numbers

UK Cystic Fibrosis Registry 2016 Annual Data Report

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

Report prepared by

Siobhán Carr	Consultant Respiratory Paediatrician	Royal Brompton Hospital
Susan Charman	Registry Senior Statistician	Cystic Fibrosis Trust
Roisin Connon	Registry Data Analyst	Cystic Fibrosis Trust
Rebecca Cosgriff	UK Cystic Fibrosis Registry Lead	Cystic Fibrosis Trust
Andrew Lee	Registry Statistician	Cystic Fibrosis Trust

With assistance from

The UK CF Registry Steering Committee

Chloe Ainsley	Senior Graphic Designer	Cystic Fibrosis Trust
Elaine Gunn	UK Cystic Fibrosis Registry Data Manager	Cystic Fibrosis Trust

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.

Contact information

For more information about this report, or the UK Cystic Fibrosis Registry, please contact us:

registry@cysticfibrosis.org.uk

@CFTrust

Cystic Fibrosis strength in numbers

Contents

Acknowledgements 3
Contact information 3

Introduction 6
- Cystic fibrosis 6
- UK Cystic Fibrosis Registry 7
- Governance 7
- Data collection 7

Section 1: Scotland-wide analysis 8
- 1.1 Summary of the UK Cystic Fibrosis Registry 8
- 1.2 Age distribution by gender 9
- 1.3 Median height percentiles among children and young persons (<20 years) 10
- 1.4 Median weight percentiles among children and young persons (<20 years) 11
- 1.5 Median BMI percentiles among children and young people (<20 years) 12
- 1.6 Median BMI values among adults (>20 years) 13
- 1.7 Education and employment in adults (>16 years) 14
- 1.8 Pregnancy 14

Diagnosis of cystic fibrosis 15
- 1.9 Age at diagnosis and screening statistics among children (<16 years) 15
- 1.10 Age at diagnosis and screening statistics among adults (≥16 in 2016) 16

Lung health 17
- 1.11 Median FEV_1 % predicted, GLI equations among patients aged 6 years and over 18
- 1.12 Median FEV_1 % predicted vs BMI among patients aged 20 years and older 19
- 1.13 Median best FEV_1 % predicted (GLI equations) among patients aged 6 years and older 19

Lung infections 20
- 1.14 Lung infections in 2016 20
- 1.15 Non-tuberculous mycobacteria (NTM) or atypical mycobacteria 21
- 1.16 Lung infections over time 22

Complications 23
- 1.17 Prevalence of complications 23
- 1.18 Incidence of complications 24
- 1.19 Cystic fibrosis-related diabetes 24

Antibiotics 25
1.20 Intravenous (IV) antibiotiocs 25
1.21 Inhaled antibiotic use among patients with chronic Pseudomonas aeruginosa 25
1.22 Long-term use of azithromycin among patients with and without chronic Pseudomonas aeruginosa 26
Muco-active therapies 27
1.23 Mannitol 27
1.24 DNase 27
1.25 Hypertonic saline 27
Other therapies 28
1.26 CFTR modifiers 28
1.27 Oxygen and non-invasive ventilation 28
1.28 Physiotherapy 29
1.29 Feeding 29
1.30 Transplants 29
Genotypes 30
1.31 Genotypes 30

Section 2: Centre-level analysis 31
A guide to charts 32
Paediatric centre analysis 33
2.1 Median FEV_1 % predicted among patients aged 6 years and older by paediatric centre/clinic 33
2.2 Median BMI percentile among patients aged 2 to 15 years by paediatric centre/clinic 33
2.3 Proportion of patients with chronic *P. aeruginosa* by paediatric centre/clinic 34
2.4 Proportion of patients receiving DNase treatment by paediatric centre/clinic 35
2.5 Proportion of patients receiving hypertonic saline by paediatric centre/clinic 35
Adult centre analysis 36
2.6 Median age (years) by adult service 36
2.7 Median FEV_1 % predicted by adult service 36
2.8 Median BMI among patients aged 16 years and older by adult service 37
2.9 Proportion of patients with chronic *P. aeruginosa* by adult service 37
2.10 Proportion of patients receiving DNase treatment by adult service 38
2.11 Proportion of patients receiving hypertonic saline treatment by adult service 38

Appendices 39
Glossary 39
Appendix 1: Centre-level data tables 42
Paediatric centres/clinics providing data in 2016 – ordered by clinic ID 42
Adult centres/clinics providing data in 2016 – ordered by clinic ID 42
Paediatric centres/clinics providing data in 2016 – ordered alphabetically 44
Adult centres/clinics providing data in 2016 – ordered alphabetically 44
Appendix 2: Composition of UKCystic Fibrosis 46
Registry Steering Committee 46

Introduction

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

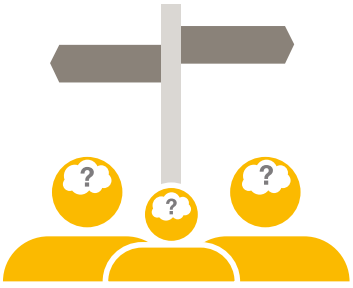
Cystic fibrosis

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 a build-up of thick mucus that blocks airways. Over time, this makes it harder to breathe and increases the risk of lung infections. Around 85% of people with CF also have difficulty digesting food due to the build-up of mucus in the pancreas and digestive system.

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 via 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 CF, 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 2 on page 46 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 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 on page 39.**

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

	2016	
	UK	**Scotland**
CF patients registered[1] Excluding 2016 diagnoses	10461 10214	908 901
CF patients with "complete" data[2]**, n (%)**	9695 (95%)	829 (92%)
Age in years, median[2]	20	21
All newly diagnosed patients (newborn screening and other)[3]	247	7
Newly diagnosed patients identified through newborn screening[4]	180	6
Age at diagnosis in months; median[3]	2	2
Adults aged 16 years and over, %[3]	60.4	62.7
Males, %	53.2	53
Genotyped, %[3]	98.4	98.6
Total deaths reported[4]	148 (1.5%)	17 (1.9%)
Age at death in years, median[5] **(95%CI)**	31 (29, 33)	31 (28, 39)

Notes:

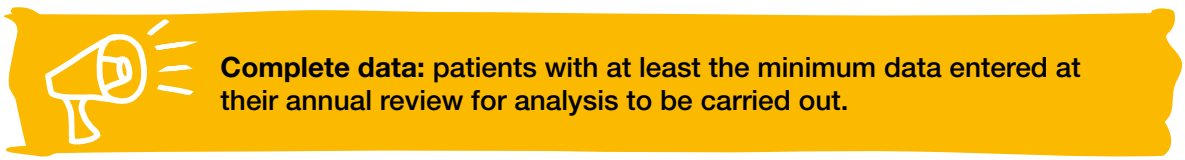
[1] This is calculated as the number of patients on the database who were diagnosed with CF and had not died before 1 January in the given year.

[2] A patient has "Complete" data if their team has filled in an annual review for that year. Patients newly diagnosed in 2016 may not have their first annual review in the same year. If newly diagnosed patients are excluded, 92% 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.

[5] Calculated from patients who died in the given year.

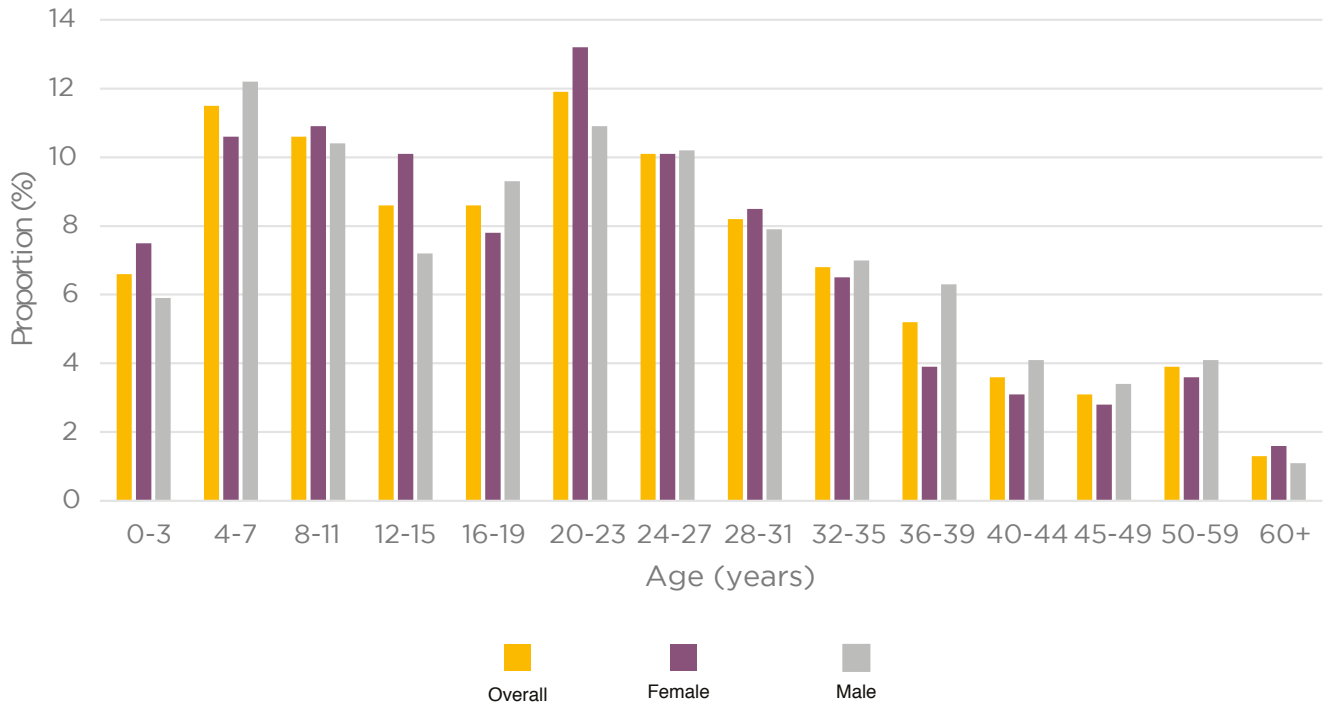


Complete data: patients with at least the minimum data entered at their annual review for analysis to be carried out.

1.2 Age distribution by gender

n=829

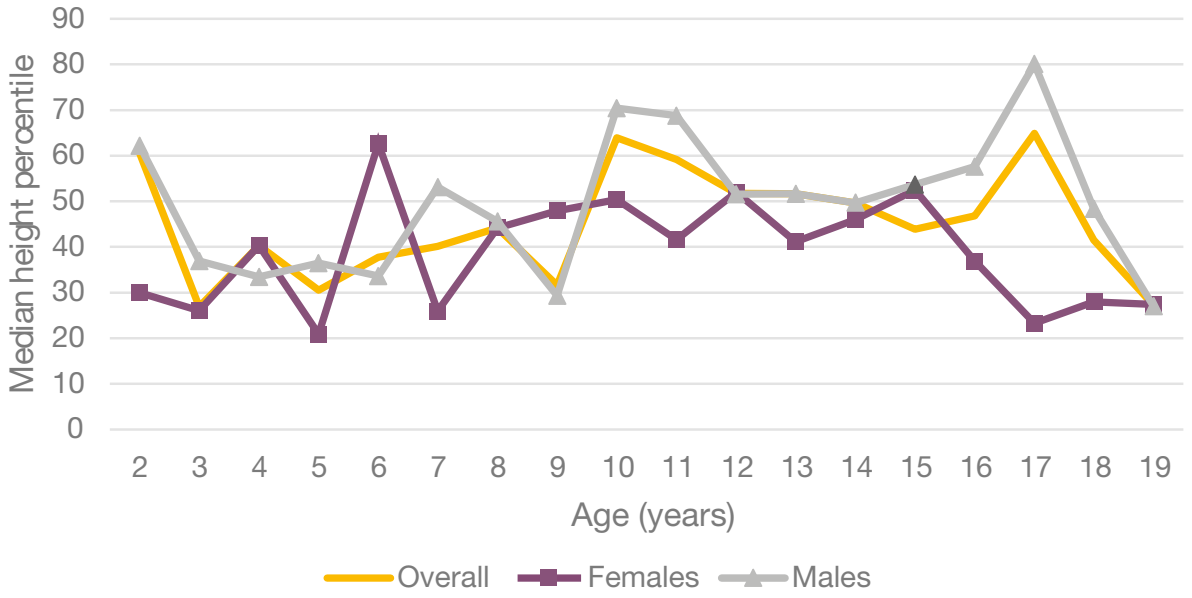
This chart shows the mix of ages and genders in the cystic fibrosis population in Scotland.



Age	All; n (%)	Female; n (%)	Male; n (%)
0-3	55 (6.6)	29 (7.5)	26 (5.9)
4-7	95 (11.5)	41 (10.6)	54 (12.2)
8-11	88 (10.6)	42 (10.9)	46 (10.4)
12-15	71 (8.6)	39 (10.1)	32 (7.2)
16-19	71 (8.6)	30 (7.8)	41 (9.3)
20-23	99 (11.9)	51 (13.2)	48 (10.9)
24-27	84 (10.1)	39 (10.1)	45 (10.2)
28-31	68 (8.2)	33 (8.5)	35 (7.9)
32-35	56 (6.8)	25 (6.5)	31 (7.0)
36-39	43 (5.2)	15 (3.9)	28 (6.3)
40-44	30 (3.6)	12 (3.1)	18 (4.1)
45-49	26 (3.1)	11 (2.8)	15 (3.4)
50-59	32 (3.9)	14 (3.6)	18 (4.1)
60+	11 (1.3)	6 (1.6)	5 (1.1)
Overall Total	**829**	**387**	**442**

1.3 Median height percentiles among children and young persons (<20 years) [6] n=357

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.



▲ Where there are fewer than 5 people in an age group the plotted value on the graph is the midpoint of the two values at either side.

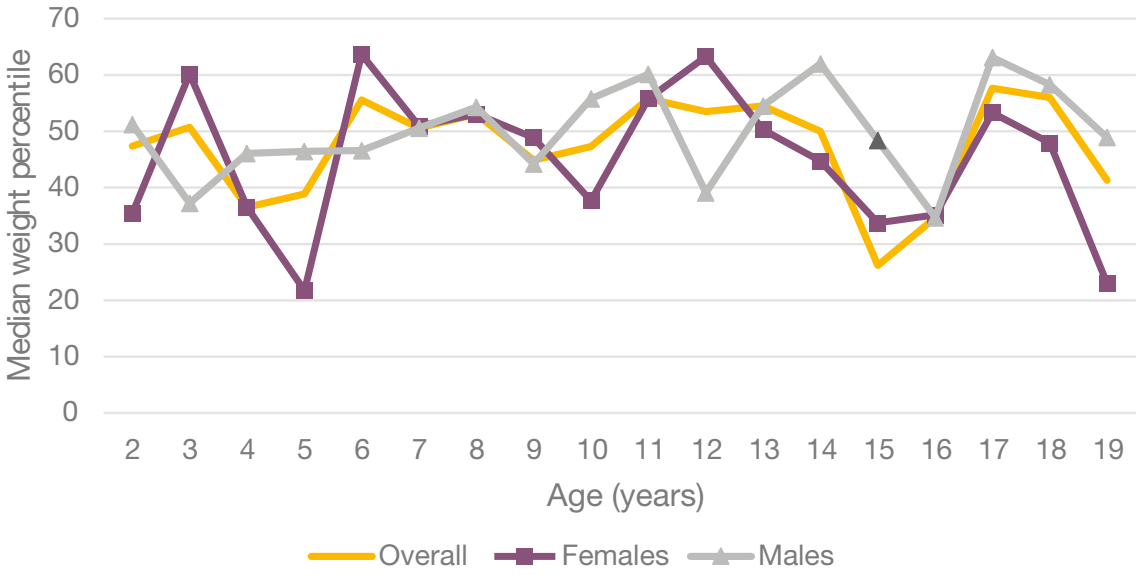
	Overall			Female			Male		
Age (years)	N	Median	IQR*	N	Median	IQR*	N	Median	IQR*
2	15	60.5	30.0-63.7	5	30.0	16.3-40.4	10	62.2	47.0-68.6
3	17	26.9	18.6-44.4	11	26.0	18.6-69.8	6	37.0	16.7-44.4
4	17	40.3	15.2-65.2	9	40.3	29.0-68.7	8	33.5	10.4-64.2
5	28	30.5	14.6-54.2	12	20.9	10.9-49.9	16	36.4	21.4-55.7
6	26	37.8	16.5-73.0	8	62.6	20.0-73.7	18	33.7	16.5-53.0
7	24	40.1	9.3-68.0	12	26.0	9.5-61.0	12	53.1	5.7-72.6
8	18	44.2	30.9-71.4	8	44.2	34.0-72.8	10	45.7	30.9-71.4
9	21	31.6	11.1-64.7	10	47.9	12.1-68.5	11	29.3	8.9-62.4
10	29	63.9	29.2-74.4	14	50.3	22.5-69.1	15	70.4	29.8-78.4
11	20	59.2	23.7-78.6	10	41.6	14.4-66.4	10	68.8	39.0-89.2
12	26	51.8	28.8-70.3	13	52.0	33.7-69.3	13	51.6	28.8-70.3
13	17	51.7	25.1-82.3	8	41.2	18.0-84.6	9	51.7	47.5-82.3
14	15	49.7	21.1-60.6	6	45.9	29.8-52.4	9	49.7	11.6-60.6
15	13	43.9	16.9-72.5	12	52.5	15.5-74.2	<5	N/A	N/A
16	15	46.8	21.6-68.7	6	37.0	13.9-70.6	9	57.6	21.9-67.7
17	14	64.9	18.1-82.1	7	23.3	5.8-70.1	7	80.1	33.5-84.3
18	18	41.6	27.0-49.5	8	28.0	15.7-47.5	10	48.4	37.2-75.5
19	24	27.2	11.0-56.0	9	27.4	8.9-52.4	15	27.0	11.6-59.5
Overall	**357**	**43.1**	**18.6-69.1**	**168**	**38.0**	**17.1-68.3**	**189**	**49.5**	**24.7-70.4**

[6] Based on UK-WHO growth charts 1990 (updated 1996)

*Interquartile range

1.4 Median weight percentiles of children and young persons (<20 years)[6] n=357

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.



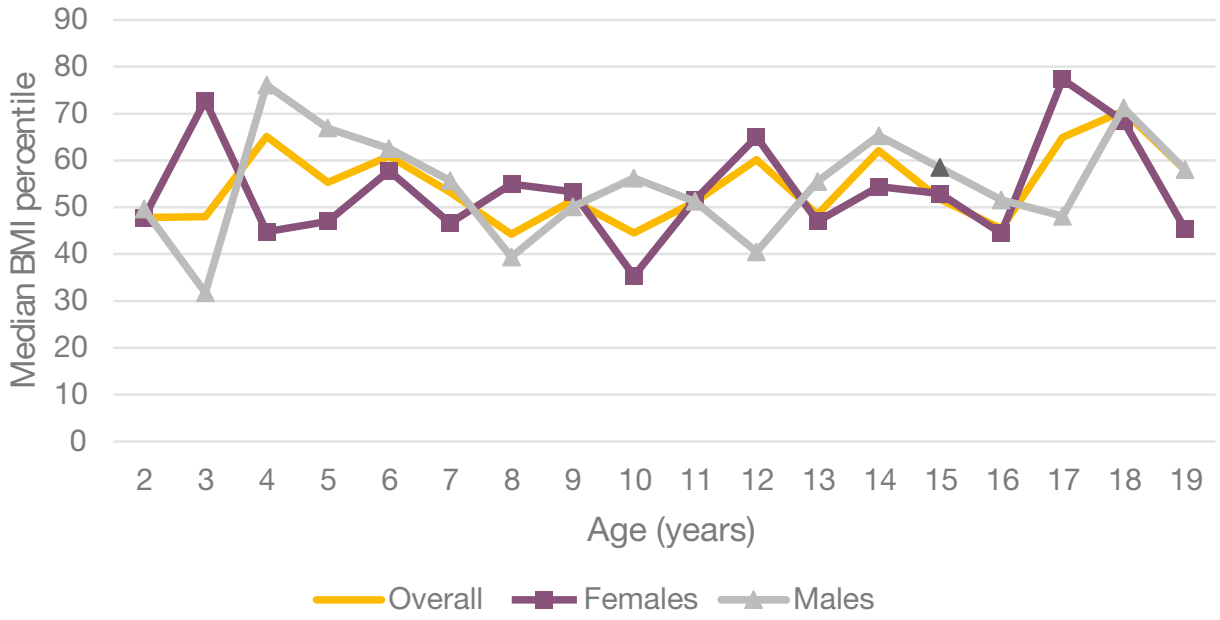
▲ Where there are fewer than 5 people in an age group the plotted value on the graph is the midpoint of the two values at either side

	Overall			Female			Male		
Age (years)	**N**	**Median**	**IQR***	**N**	**Median**	**IQR***	**N**	**Median**	**IQR***
2	15	47.4	27.0-64.3	5	35.4	32.6-49.3	10	51.2	27.0-70.4
3	17	50.7	14.3-66.3	11	60.2	14.3-80.6	6	37.3	10.5-50.7
4	17	36.5	28.9-77.0	9	36.5	28.9-44.4	8	46.1	21.7-89.6
5	28	38.9	18.5-73.8	12	21.8	10.4-60.3	16	46.4	25.2-77.7
6	26	55.5	20.1-82.4	8	63.6	22.9-82.9	18	46.6	17.7-82.4
7	24	50.6	25.8-77.5	12	50.9	9.0-76.4	12	50.6	37.8-77.5
8	18	53.1	33.0-74.1	8	53.1	39.0-73.3	10	54.3	16.7-74.1
9	21	44.8	21.0-67.8	10	48.9	13.7-91.6	11	44.3	22.3-60.7
10	29	47.3	29.9-64.1	14	37.7	21.2-59.7	15	55.8	31.8-67.1
11	20	55.8	37.3-72.2	10	55.8	30.0-63.5	10	60.1	40.9-93.3
12	26	53.5	31.3-77.6	13	63.3	36.4-77.6	13	39.1	29.0-72.9
13	17	54.5	31.7-76.8	8	50.3	22.9-74.9	9	54.5	51.2-87.6
14	15	50.0	27.6-85.1	6	44.8	27.6-68.2	9	62.0	30.2-85.1
15	13	26.2	18.1-67.0	12	33.8	18.0-76.6	<5	N/A	N/A
16	15	34.7	12.3-59.2	6	35.2	10.3-59.2	9	34.7	20.1-59.1
17	14	57.7	35.9-80.7	7	53.3	49.8-80.7	7	63.1	25.5-98.9
18	18	56.0	33.9-67.1	8	47.9	19.7-64.4	10	58.3	36.5-75.7
19	24	41.3	11.2-65.1	9	23.0	13.6-48.6	15	48.9	4.2-69.6
Overall	**357**	**49.3**	**23.0-73.1**	**168**	**47.1**	**19.8-69.8**	**189**	**50.6**	**27.0-75.7**

[6] Based on UK-WHO growth charts 1990 (updated 1996)
*Interquartile range

1.5 Median Body Mass Index (BMI) percentiles among children and young people (<20 years)[6]
n=357

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.



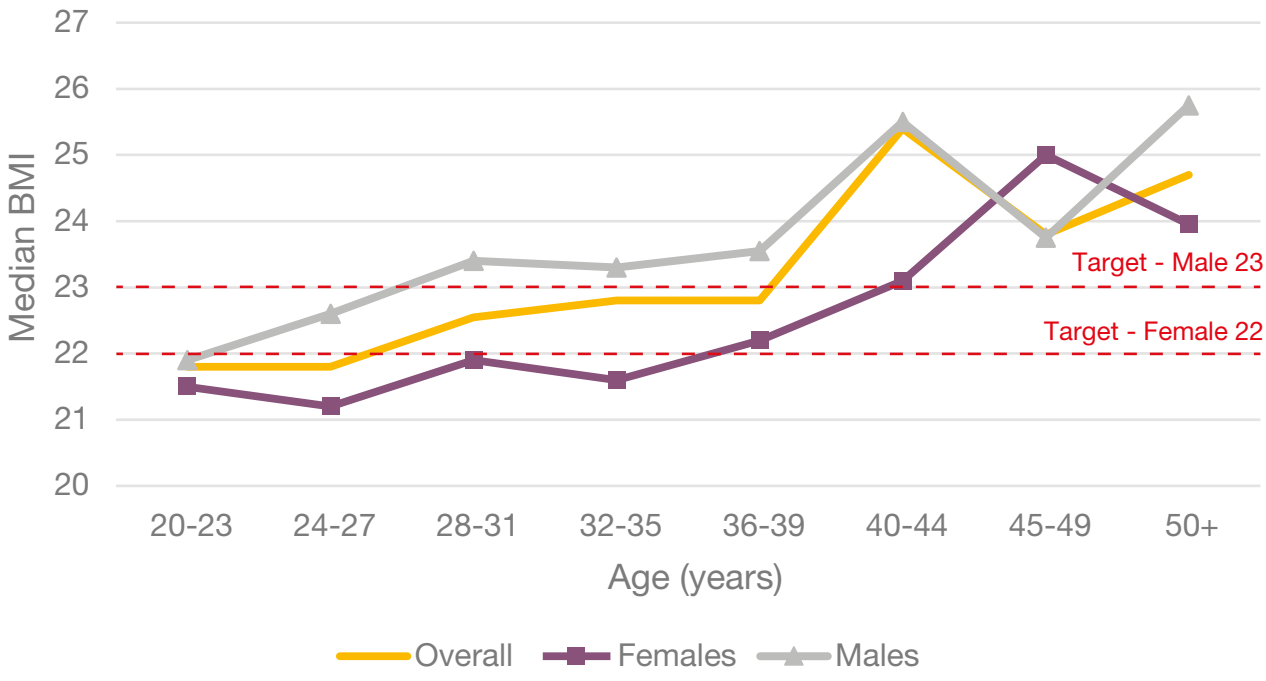
▲ Where there are less than 5 people in an age group the plotted value on the graph is the midpoint of the two values at either side.

	Overall			Female			Male		
Age	**N**	**Median**	**IQR**	**N**	**Median**	**IQR**	**N**	**Median**	**IQR**
2	15	47.8	23.6-68.9	5	47.8	11.8-75.6	10	49.6	29.6-66.9
3	17	48.0	25.6-79.7	11	72.8	26.8-82.6	6	31.8	17.9-67.4
4	17	65.1	35.6-89.8	9	44.8	29.7-74.5	8	76.1	47.2-94.3
5	28	55.3	39.3-74.5	12	47.0	36.8-63.0	16	66.9	40.5-81.8
6	26	60.9	39.1-89.5	8	57.8	40.6-78.7	18	62.6	33.2-90.6
7	24	53.2	31.3-84.9	12	46.6	28.8-79.3	12	55.7	34.1-84.9
8	18	44.3	34.1-74.7	8	55.0	35.0-79.7	10	39.4	28.3-65.6
9	21	51.5	34.4-74.1	10	53.3	26.3-97.6	11	50.1	39.9-70.5
10	29	44.5	16.4-71.5	14	35.3	9.4-71.5	15	56.2	16.4-71.8
11	20	51.3	34.3-84.1	10	51.7	35.3-77.7	10	51.3	33.2-92.2
12	26	60.2	27.5-69.3	13	65.0	27.5-69.3	13	40.5	31.7-66.8
13	17	48.5	42.8-64.5	8	47.0	37.5-57.2	9	55.5	42.8-65.9
14	15	62.1	30.3-82.3	6	54.4	39.0-78.6	9	65.3	30.3-82.3
15	13	51.8	19.2-67.6	12	52.9	23.4-78.5	<5	N/A	N/A
16	15	45.4	21.6-66.4	6	44.5	17.9-66.4	9	51.6	26.0-63.7
17	14	64.9	35.1-80.7	7	77.3	50.8-80.7	7	48.1	32.1-97.1
18	18	70.6	19.1-76.6	8	68.4	22.0-73.3	10	71.3	19.1-87.1
19	24	57.9	9.5-82.2	9	45.4	6.9-78.0	15	58.1	9.6-86.3
Overall	**357**	**53.2**	**30.3-76.7**	**168**	**51.6**	**28.0-76.1**	**189**	**54.5**	**31.7-77.6**

[6] Based on UK-WHO growth charts 1990 (updated 1996)

1.6 Median Body Mass Index (BMI) values among adults (20 years and over)
n=449

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	99	21.8	19.7-23.4	51	21.5	18.4-23.7	48	21.9	19.7-23.4
24-27	84	21.8	20.1-24.0	39	21.2	19.4-23.4	45	22.6	20.9-24.6
28-31	68	22.5	20.5-25.3	33	21.9	20.7-25.2	35	23.4	20.3-26.1
32-35	56	22.8	20.4-24.8	25	21.6	20.1-24.6	31	23.3	21.3-25.3
36-39	43	22.8	21.3-26.6	15	22.2	20.1-25.1	28	23.5	21.7-27.0
40-44	30	25.4	21.6-27.3	12	23.1	21.2-27.4	18	25.5	22.6-27.1
45-49	26	23.8	22.1-25.8	11	25.0	20.9-27.8	15	23.8	22.1-25.4
50+	43	24.7	22.0-28.1	20	24.0	22.0-29.0	23	25.8	22.1-28.0
Overall	**449**	**22.6**	**20.4-25.3**	**206**	**22.0**	**20.1-24.8**	**243**	**23.0**	**21.1-25.8**

[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=520

The following table shows how people with CF reported their education and employment status in 2016. Please note that the groups are not mutually exclusive; someone may be a student as well as working part-time, for example.

	Number of people n (%)
Number of people who completed questionnaire	506
Full-time employment	177 (35.0)
Part-time employment	92 (18.2)
Student	72 (14.2)
Homemaker	19 (3.8)
Unemployed	100 (19.8)
Disabled	18 (3.6)
Retired	13 (2.6)
Unknown entered	15 (3.0)
Number of people in work or study	**341 (67.4)**

1.8 Pregnancy

7 women with cystic fibrosis had babies in 2016

10 men with cystic fibrosis became fathers in 2016

Diagnosis of cystic fibrosis

1.9 Age at diagnosis in children under 16
n= 309*

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 between five and 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 2016; n (%)	Patients aged 5 years in 2016; n (%)
Pre-natal	0 (0.0)	0 (0.0)	0 (0.0)
Birth-3months	270 (88.2)	26 (89.7)	25 (89.3)
4-6 months	9 (2.9)	<5	<5
7-12 months	6 (2.0)	<5	<5
1 years	<5	<5	<5
2 years	9 (2.9)	<5	<5
3 years	<5	<5	<5
4 years	<5	<5	<5
5 years	<5	<5	<5
6 years	<5	<5	-
7 years	<5	<5	-
8 years	<5	<5	-
9 years	<5	<5	-
10 years	<5	<5	-
11 years	<5	-	-
12 years	<5	-	-
13 years	<5	-	-
14 years	<5	-	-
15 years	<5	-	-
Overall	**306**	**29**	**28**

*n=306 – had exact diagnosis dates

The median (range) age at diagnosis for patients under 16 in 2016 is 20 days (0-140 months)

1.10 Age at diagnosis in adults aged 16 years and over in 2016 n=520

The table below shows the age at diagnosis of people aged 16 and over in 2016. 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 (0.0)
Birth-3 months	186 (35.8)
4-6 months	51 (9.8)
7-12 months	33 (6.3)
1 years	39 (7.5)
2 years	34 (6.5)
3 years	24 (4.6)
4 years	17 (3.3)
5 years	9 (1.7)
6 years	8 (1.5)
7 years	6 (1.2)
8 years	8 (1.5)
9 years	5 (1.0)
10 years	<5
11 years	6 (1.2)
12 years	6 (1.2)
13 years	<5
14 years	<5
15 years	7 (1.3)
16-20 years	16 (3.1)
21-25 years	7 (1.3)
26-30 years	18 (3.5)
31-35 years	11 (2.1)
36-40 years	6 (1.2)
41-45 years	5 (1.0)
46-50 years	<5
51-60 years	7 (1.3)
61 years+	<5
Overall	**520**

Lung health

For people with CF, mucus in the lungs is linked to repeated or chronic infections, 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 people with 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.

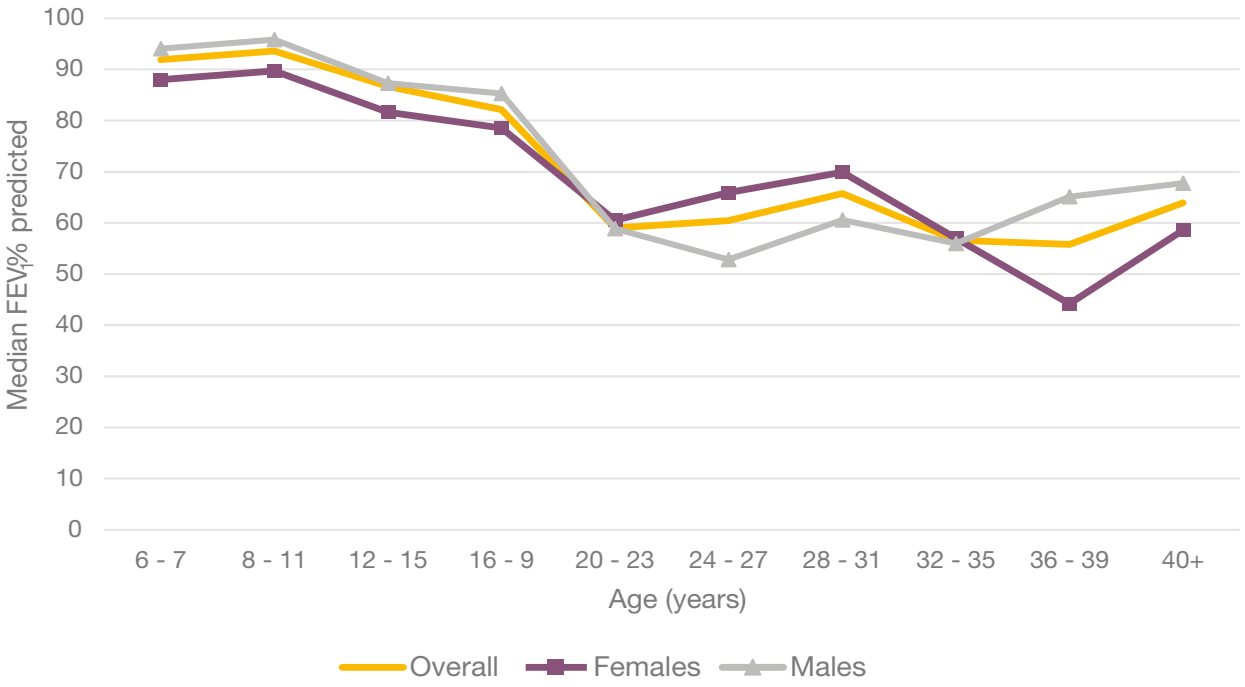
This is often a team effort between people with CF, their family, and their medical team, which can include doctors, nurses, physiotherapists, dieticians, and psychologists.

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

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

n=692

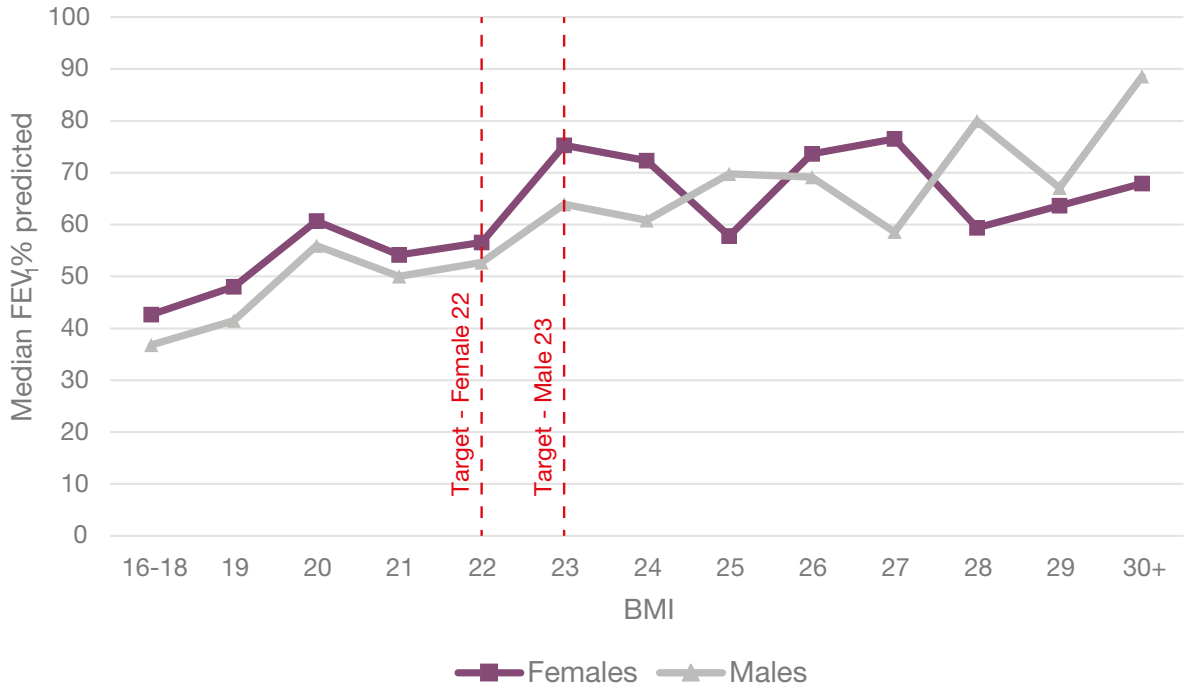
The chart and table in this section show the information about those patients whose FEV_1 data were complete. People with CF who have had lung transplants are excluded, as their new 'non-CF' lungs would have lung health similar to a person without Cystic Fibrosis.



	Overall			Female			Male		
Age (years)	**N**	**Median**	**IQR**	**N**	**Median**	**IQR**	**N**	**Median**	**IQR**
6-7	46	91.9	83.7-104.3	18	88.0	78.2-107.3	28	94.0	87.1-102.9
8-11	84	93.6	86.4-101.8	39	89.7	86.0-101.6	45	95.8	88.1-103.1
12-15	71	86.7	75.7-92.0	39	81.7	71.2-93.6	32	87.3	79.5-91.7
16-19	71	82.1	63.5-93.1	30	78.6	55.6-89.6	41	85.3	69.3-93.4
20-23	94	59.0	37.6-76.4	49	60.5	44.0-79.1	45	58.9	36.7-74.0
24-27	79	60.4	42.2-80.8	36	65.9	47.7-80.0	43	52.8	39.7-80.9
28-31	62	65.8	42.9-79.3	32	69.9	60.8-77.8	30	60.6	34.5-79.9
32-35	51	56.6	44.1-78.4	23	57.0	50.2-73.5	28	56.0	39.1-81.3
36-39	40	55.8	40.6-78.1	14	44.2	34.8-60.7	26	65.1	46.7-80.1
40+	94	63.9	45.0-78.1	41	58.6	44.3-73.8	53	67.7	48.9-79.4
Overall	**692**	**75.0**	**53.0-90.5**	**321**	**73.7**	**54.1-89.6**	**371**	**76.4**	**52.8-91.2**

1.12 Median FEV_1 % predicted and BMI among patients aged 20 years and older
n=445

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 patients with BMI ≥30 together.

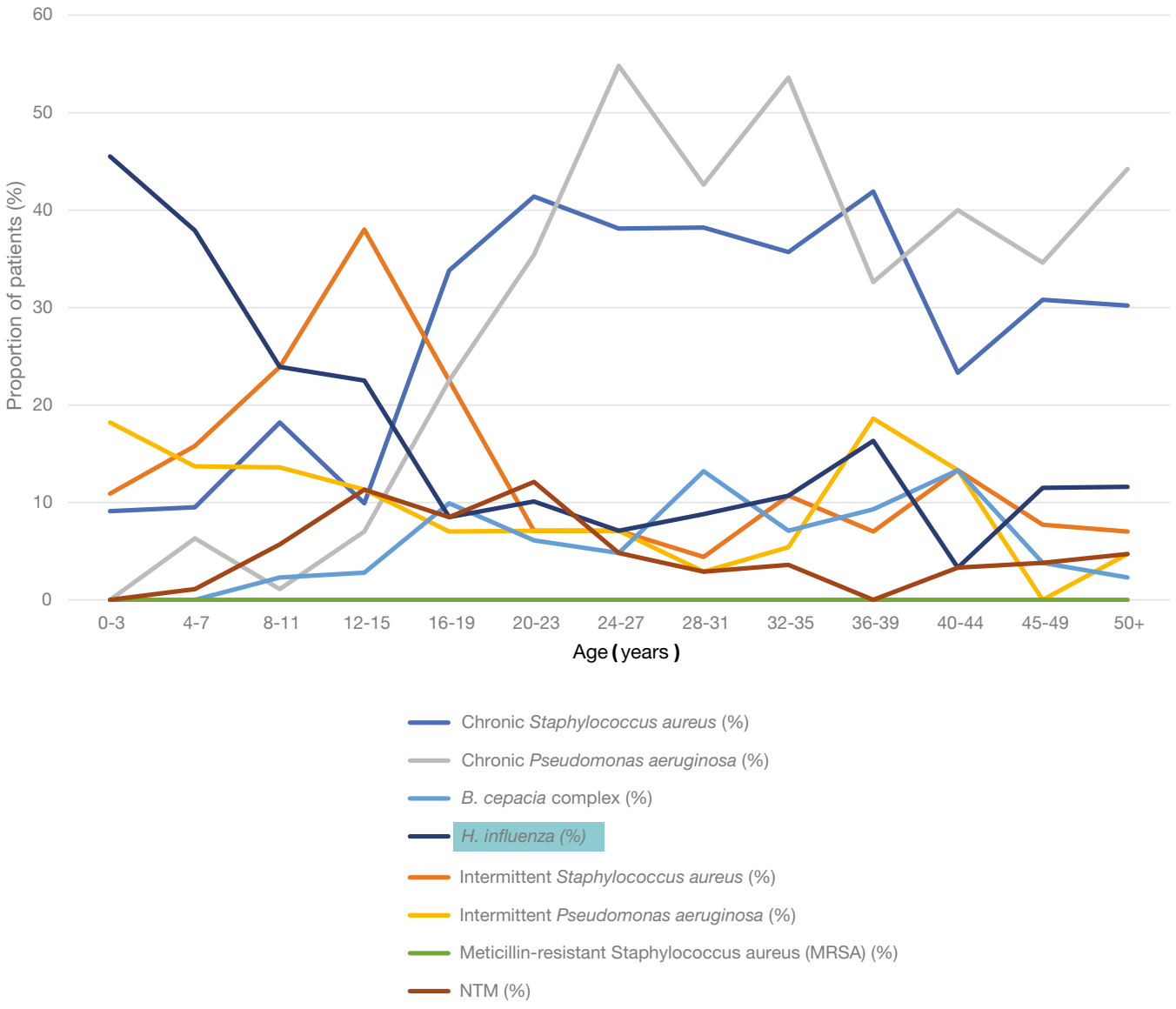
1.13 Median best FEV_1 % predicted (GLI equations) among patients aged 6 years and older
n=657

	Overall			Female			Male		
Age (years)	**N**	**Median**	**IQR**	**N**	**Median**	**IQR**	**N**	**Median**	**IQR**
6-7	38	96.7	89.3 - 105.6	17	96.8	86 - 111.4	21	96.6	91.6 - 105
8-11	74	97.6	89.1 - 106.4	36	97.2	84.6 - 104.6	38	98.8	91.8 - 106.6
12-15	63	88.5	77.1 - 97.4	33	87.7	74.5 - 93.2	30	89.8	82.4 - 97.4
16-19	68	89.5	69.9 - 99.3	28	86.4	58.2 - 95.9	40	90.1	76.8 - 103.1
20-23	94	65.3	45.8 - 83.3	49	67.9	45.8 - 83.3	45	65.3	46.5 - 80.9
24-27	80	64.9	48.5 - 83.5	37	72.3	55.7 - 82.7	43	60.0	43.5 - 83.8
28-31	60	72.2	50.9 - 83.3	31	75.2	62.3 - 84.4	29	62.3	40.4 - 82.1
32-35	50	60.6	47.1 - 78.6	22	61.9	53.6 - 78.3	28	57.4	39.7 - 81.8
36-39	39	62.2	50.5 - 83.7	13	58.0	41.6 - 62.2	26	68.8	51.2 - 84.7
40+	91	65.2	52.8 - 79.6	41	63.6	49.8 - 75.4	50	69.9	54.8 - 79.9
Overall	**657**	**77.1**	**57.6 - 94.4**	**307**	**76.0**	**58.7 - 94.2**	**350**	**78.4**	**56.6 - 94.7**

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.

1.14 Lung infections in 2016
<16 years n=309, ≥16 years n=520



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.

	Paediatric Age Range (Years)				Overall
	0-3	4-7	8-11	12-15	Paediatric (<16 years)
Number in age range	55	95	88	71	309
Chronic *S.aureus*, n (%)	5 (9.1%)	9 (9.5%)	16 (18.2%)	7 (9.9%)	37 (12%)
Intermittent *S.aureus*, n (%)	6 (10.9%)	15 (15.8%)	21 (23.9%)	27 (38%)	69 (22.3%)
Chronic *P. aeruginosa*, n (%)	0 (0%)	6 (6.3%)	<5	5 (7%)	- (3.9%)
Intermittent *P. aeruginosa*, n (%)	10 (18.2%)	13 (13.7%)	12 (13.6%)	8 (11.3%)	43 (13.9%)
B. cepacia complex, n (%)	0 (0%)	0 (0%)	<5	<5	- (1.3%)
MRSA, n (%)	0 (0%)	0 (0%)	0 (0%)	0 (0%)	0 (0%)
H. influenza, n (%)	25 (45.5%)	36 (37.9%)	21 (23.9%)	16 (22.5%)	98 (31.7%)
NTM, n (%)	0 (0%)	<5	5 (5.7%)	8 (11.3%)	- (4.5%)

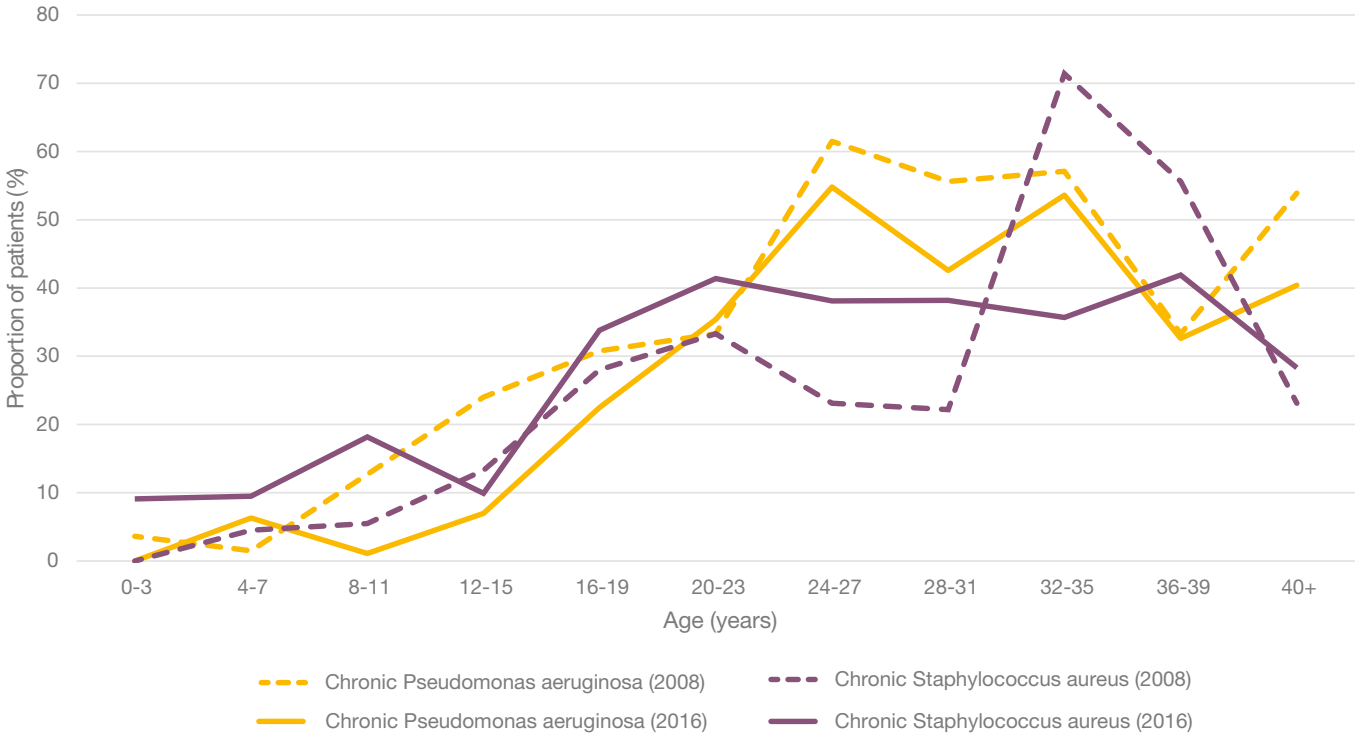
	Adult Age Range (Years)									Overall
	16-19	20-23	24-27	28-31	32-35	36-39	40-44	45-49	50+	Adults (≥16 years)
Number in age range	71	99	84	68	56	43	30	26	43	520
Chronic *S.aureus*, n (%)	24 (33.8%)	41 (41.4%)	32 (38.1%)	26 (38.2%)	20 (35.7%)	18 (41.9%)	7 (23.3%)	8 (30.8%)	13 (30.2%)	189 (36.3%)
Intermittent *S.aureus*, n (%)	16 (22.5%)	7 (7.1%)	6 (7.1%)	<5	6 (10.7%)	<5	<5	<5	<5	50 (9.6%)
Chronic *P. aeruginosa*, n (%)	16 (22.5%)	35 (35.4%)	46 (54.8%)	29 (42.6%)	30 (53.6%)	14 (32.6%)	12 (40%)	9 (34.6%)	19 (44.2%)	210 (40.4%)
Intermittent *P. aeruginosa*, n (%)	5 (7%)	7 (7.1%)	6 (7.1%)	<5	<5	8 (18.6%)	<5	0 (0%)	<5	37 (7.1%)
B. cepacia complex, n (%)	7 (9.9%)	6 (6.1%)	<5	9 (13.2%)	<5	<5	<5	<5	<5	40 (7.7%)
MRSA, n (%)	0 (0%)	0 (0%)	0 (0%)	0 (0%)	0 (0%)	0 (0%)	0 (0%)	0 (0%)	0 (0%)	0 (0%)
H. influenza, n (%)	6 (8.5%)	10 (10.1%)	6 (7.1%)	6 (8.8%)	6 (10.7%)	7 (16.3%)	<5	<5	5 (11.6%)	50 (9.6%)
NTM, n (%)	6 (8.5%)	12 (12.1%)	<5	<5	<5	0 (0%)	<5	<5	<5	30 (5.8%)

1.15 Nontuberculous mycobacteria (NTM) or atypical mycobacteria*

	2014 (n=782)	2015 (n=795)	2016 (n=829)
NTM prevalence (%)	34 (4.3%)	37(4.7%)	44 (5.3%)
On NTM treatment in the given year (% of NTM prevalence in given year)	8 (24)	12(32)	16(36)
NTM incidence	29	20	20
M.abscessus prevalence**	26	23	25

* Non tuberculous mycobacterium is slow to grow and slow to treat and may be present for several years before eradication, or may never be cleared.In the table 1.15, 'prevalence' represents all people reported in that years as having a positive culture. 'Incidence' represents all positive cultures in individual who have not reported any in the previous 2 years.
** M.abscessus incidence in 2016 was 11. Prior years cannot be evaluated as enhanced NTM reporting was not available prior to 2014.

1.16 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+**
Chronic *S. aureus*; 2016	9.1	9.5	18.2	9.9	33.8	41.4	38.1	38.2	35.7	41.9	28.3
Chronic *P. aeruginosa*; 2016	0.0	6.3	1.1	7.0	22.5	35.4	54.8	42.6	53.6	32.6	40.4
Chronic *S. aureus*; 2008	0.0	4.5	5.5	13.3	28.0	33.3	23.1	22.2	71.4	55.6	23.1
Chronic *P. aeruginosa*; 2008	3.6	1.5	12.7	24.0	30.8	33.3	61.5	55.6	57.1	33.3	53.9

1.17 Prevalence of complications

	Overall (n=795)	<16 years (n=298)	≥16 years (n=497)
	N (%)	N (%)	N (%)
Respiratory Related			
Nasal polyps requiring surgery	-	<5	9 (1.7)
Sinus disease	-	<5	164 (31.5)
Asthma	79 (9.5)	14 (4.5)	65 (12.5)
Allergic bronchopulmonary aspergillosis (ABPA)	-	<5	33 (6.3)
Haemoptysis	23 (2.8)	0 (0.0)	23 (4.4)
Pneumothorax requiring chest tube	0 (0.0)	0 (0.0)	0 (0.0)
Pancreas & Hepatobiliary Disease			
Elevated Liver enzymes	-	<5	25 (4.8)
Liver disease	104 (12.5)	23 (7.4)	81 (15.6)
Cirrhosis with no portal hypertension	-	<5	12 (2.3)
Cirrhosis with portal hypertension	19 (2.3)	6 (1.9)	13 (2.5)
Gallbladder disease requiring surgery	<5	<5	<5
Pancreatitis	-	<5	7 (1.3)
Upper Gastrointestinal (GI)			
Gastroesophageal reflux disease (GERD)	100 (12.1)	<5	99 (19.0)
Peptic ulcer	0 (0.0)	0 (0.0)	0 (0.0)
GI bleed req hospital variceal	0 (0.0)	0 (0.0)	0 (0.0)
GI bleed req hospital non variceal	<5	0 (0.0)	<5
Lower GI			
Intestinal obstruction	0 (0.0)	0 (0.0)	0 (0.0)
Fibrosing colonopathy/colonic structure	-	<5	88 (16.9)
Rectal prolapse	0 (0.0)	0 (0.0)	0 (0.0)
Meconium ileus	<5	<5	0 (0.0)
Renal			
Kidney stones	<5	<5	<5
Renal failure	13 (1.6)	0 (0.0)	13 (2.5)
Musculoskeletal			
Arthritis	<5	<5	<5
Arthropathy	33 (4.0)	0 (0.0)	33 (6.3)
Bone fracture	0 (0.0)	0 (0.0)	0 (0.0)
Osteopenia	107 (12.9)	0 (0.0)	107 (20.6)
Osteoporosis	42 (5.1)	0 (0.0)	42 (8.1)
Other			
Cancer confirmed by histology	0 (0.0)	0 (0.0)	0 (0.0)
Port inserted or replaced	7 (0.8)	<5	<5
Depression	21 (2.5)	0 (0.0)	21 (4.0)
Hearing loss	-	<5	9 (1.7)
Hypertension	17 (2.1)	0 (0.0)	17 (3.3)

1.18 Incidence of complications

	Newly identified in 2015			Newly identified in 2016		
	Overall (n=795)	<16 years (n=298)	≥16 years (n=497)	Overall (n=829)	<16 years (n=309)	≥16 years (n520)
ABPA; n (%)	7 (0.9)	<5	<5	11 (1.3)	<5	8 (1.5)
Cirrhosis with no portal hypertension; n (%)	<5	0	<5	13 (1.6)	<5	11 (2.1)
Cirrhosis with portal hypertension; n (%)	14 (1.8)	<5	10 (2.0)	7 (0.8)	<5	<5
Cancer confirmed by histology; n (%)	<5	0	<5	<5	<5	<5

1.19 CF-related diabetes

Cystic fibrosis-related diabetes (CFRD) is common in adults and adolescents with cystic fibrosis. 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=640)	10-15 years (n=120)	≥ 16 years (n=520)
Treatment for CF-related diabetes; n(%)	155 (24.2)	11 (9.2)	144 (27.7)
Screening for CF-related diabetes			
Yes	357 (55.8)	101 (84.2)	256 (49.2)
No	117 (18.3)	13 (10.8)	104 (20.0)
Known CF-related diabetes	131 (20.5)	5 (4.2)	126 (24.2)
Unknown	13 (2.0)	<5	12 (2.3)

Antibiotics

1.20 Intravenous (IV) antibiotic use

When someone with CF 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	Number in age band	N (%)	Median days (IQR)	N (%)	Median days (IQR)	N (%)	Median days (IQR)
0-3	55	<5		10 (18.2)	13 (7-20)	10 (18.2)	14 (12-20)
4-7	95	8 (8.4)	25 (13-48)	20 (21.1)	21 (14-33)	23 (24.2)	27 (14-52)
8-11	88	11 (12.5)	14 (9-48)	30 (34.1)	19 (14-43)	32 (36.4)	37 (14-54)
12-15	71	12 (16.9)	33 (16-42)	21 (29.6)	39 (14-70)	25 (35.2)	43 (26-70)
16-19	71	14 (19.7)	14 (12-49)	22 (31.0)	14 (6-28)	27 (38.0)	27 (14-50)
20-23	99	35 (35.4)	28 (14-39)	37 (37.4)	14 (7-28)	50 (50.5)	26 (14-42)
24-27	84	26 (31.0)	14 (13-36)	30 (35.7)	14 (7-23)	41 (48.8)	14 (14-53)
28-31	68	24 (35.3)	24 (14-53)	22 (32.4)	15 (13-29)	35 (51.5)	28 (14-51)
32-35	56	12 (21.4)	17 (14-35)	15 (26.8)	19 (11-35)	21 (37.5)	26 (15-42)
36-39	43	10 (23.3)	35 (29-56)	10 (23.3)	13 (8-14)	13 (30.2)	42 (29-55)
40-44	30	10 (33.3)	13 (8-20)	6 (20.0)	13 (6-14)	12 (40.0)	14 (12-28)
45-49	26	6 (23.1)	38 (14-41)	<5		8 (30.8)	38 (14-43)
50+	43	8 (18.6)	16 (9-29)	13 (30.2)	13 (3-14)	15 (34.9)	14 (12-28)
Overall	**829**	**177 (21.4)**	**20 (13-41)**	**239 (28.8)**	**14 (10-32)**	**312 (37.6)**	**27 (14-50)**

1.21 Inhaled antibiotic use among patients with chronic *Pseudomonas aeruginosa*

	2008			2016		
	Overall	<16 years	≥16 years	Overall	<16 years	≥16 years
Patients with chronic pseudomonas	207	14	193	222	12	210
Tobramycin solution, n(%)	33 (15.9)	5 (35.7)	28 (14.5)		<5	35 (16.7)
Other aminoglycoside, n(%)	<5	0	<5	<5	0	<5
Colistin, n(%)	64 (30.9)	8 (57.1)	56 (29.0)	76 (34.2)	9 (75.0)	67 (31.9)
Promixin, n(%)	37 (17.9)	<5	34 (17.6)		<5	31 (14.8)
Aztreonam, n(%)*	<5	0	<5	10 (4.5)	0	10 (4.8)
Colistimethate(DPI), n(%)*		<5	33 (17.1)		<5	44 (21.0)
Tobramycin Inhalation Powder, n(%)*		<5	58 (30.1)	68 (30.6)	0	68 (32.4)
At least one of the above, n(%)*	163 (78.7)	12 (85.7)	151 (78.2)	173 (77.9)	11 (91.7)	162 (77.1)

*Not reported for 2008

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

1.22 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				2016			
	Overall (n=63*)	**0-3 years (n<5)**	**4-15 years (n=34)**	**≥16 years (n=28)**	**Overall**	**0-3 years (n=59)**	**4-15 years (n=239)**	**≥16 years (n=497)**
Patients with chronic P. aeruginosa; n (%)	29 (46.0)	0 (0)	12 (35.3)	17 (60.7)	187 (43.4)	0 (0)	10 (11.2)	177 (51.8)
Patients without chronic P. aeruginosa; n (%)	34 (54.0)	<5	22 (64.7)	11 (39.3)	244 (56.6)	0 (0)	79 (88.8)	165 (48.2)

*number on azithromycin in the given year

Muco-active therapies

1.23 Mannitol

	2016	
Age	Total Patients	Patients on Mannitol
0-3	55	0
4-7	95	0
8-11	88	0
12-15	71	0
16-19	71	0
20-23	99	<5
24-27	84	<5
28-31	68	<5
32-35	56	<5
36-39	43	0
40+	99	<5
Overall	**829**	**11 (1.3)**

1.24 DNase

	DNase; n (%)			
	2008		2016	
Age	Total patients	Patients on DNase; n (%)	Total patients	Patients on DNase; n (%)
0-3	91	<5	55	6 (10.9)
4-7	78	9 (11.5)	95	19 (20.0)
8-11	66	15 (22.7)	88	40 (45.5)
12-15	92	33 (35.9)	71	36 (50.7)
16-19	32	12 (37.5)	71	40 (56.3)
20-23	14	<5	99	53 (53.5)
24-27	14	<5	84	39 (46.4)
28-31	11	<5	68	28 (41.2)
32-35	8	<5	56	17 (30.4)
36-39	9	<5	43	14 (32.6)
40+	14	5 (35.7)	99	26 (26.3)
Overall	**429**	**83 (19.4)**	**829**	**318 (38.4)**

1.25 Hypertonic saline

This treatment helps to thin mucus so that it is easier to cough up and expel from the body.

	Hypertonic saline; n (%)			
	2008		2016	
Age	Number of patients	Patients on hypertonic saline; n (%)	Number of patients	Patients on hypertonic saline; n (%)
0-3	91	0	55	<5
4-7	78	0	95	8 (8.4)
8-11	66	<5	88	20 (22.7)
12-15	92	<5	71	27 (38.0)
16-19	32	0	71	17 (23.9)
20-23	14	0	99	27 (27.3)
24-27	14	0	84	18 (21.4)
28-31	11	<5	68	17 (25.0)
32-35	8	0	56	5 (8.9)
36-39	9	0	43	5 (11.6)
40+	14	0	99	10 (10.1)
Overall	**429**	**<5**	**829**	**158 (19.1)**

Other therapies

1.26 CFTR modifiers

1.26a Ivacaftor

Ivacaftor is a drug that has been prescribed as a treatment for cystic fibrosis in patients aged six years and over with at least one copy of the genotype G551D since June 2012.

Number of patients on Ivacaftor in Scotland	66
Sweat chloride before Ivacaftor	106 (96, 106)
Sweat chloride 6-8 weeks after Ivacaftor	53 (44, 67)
FEV_1 % before Ivacaftor	61.4 (49.9, 72.8)
FEV_1 % 6-8 weeks after Ivacaftor	68.9 (53.7, 81.4)
Number of patients stopped Ivacaftor ever	<5

People with CF tend to have more chloride in their sweat than people without cystic fibrosis. This measurement is called 'sweat chloride' and is measured in mmol/litre.

1.26b Ivacaftor/Lumacaftor

Ivacaftor/Lumacaftor is licensed for use in patients aged 12 and over with two copies of the F508del mutation. In 2016 it was available to specific people with CF in the UK through a named patient access scheme. In Scotland one person received this drug in 2016.

1.27 Oxygen and non-invasive ventilation

	Overall (n=829)	<16 years (n=309)	≥16 years (n=520)
Non-invasive ventilation (NIV), n (%)		<5	5 (1.0)
Long-term oxygen, n (%)	39 (4.7)	7 (2.3)	32 (6.2)
Among those who have long-term oxygen:			
Continuous		<5	5 (1.0)
Nocturnal or with exertion		<5	11 (2.1)
As required (PRN)	<5	0	<5
With exacerbation	17 (2.1)	5 (1.6)	12 (2.3)

1.28 Physiotherapy

Physiotherapy helps people with CF clear sticky mucus from their lungs.

	Overall (n=829)	<16 years (n=309)	≥16 years (n=520)
Active cycle of breathing techniques, n (%)	160 (19.3)	36 (11.7)	124 (23.8)
Autogenic drainage (including assisted autogenic drainage), n (%)	340 (41.0)	93 (30.1)	247 (47.5)
Any form of PEP, n (%)	400 (48.3)	251 (81.2)	149 (28.7)
VEST, n (%)	<5	<5	<5
Exercise, n (%)	362 (43.7)	84 (27.2)	278 (53.5)

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

1.29 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 food intake.

	Overall (n=829)	<16 years (n=309)	≥16 years (n=520)
Any supplemental feeding, n (%)	176 (21.2)	52 (16.8)	124 (23.8)
Nasogastric tube, n (%)		<5	9 (1.7)
Gastronomy tube/button, n (%)	33 (4.0)	14 (4.5)	19 (3.7)
Jejunal	0	0	0
Total parenteral nutrition (TPN)	0	0	0

1.30 Transplants

Lung transplantation has been available to people with CF for almost 30 years. Today, the most common operation carried out is a double lung transplant, or a Bilateral Sequential Lung transplant.
Post-transplant survival rates are 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	2016
Number evaluated	12	13	21	17	18	18
Number accepted	5	7	11	12	6	8
Number receiving transplants	<5	5	<5	5	<5	6
Type of transplant received:						
Bilateral lung	<5	5	<5	5	<5	<5
Heart and lung	0	0	0	0	0	0
Liver	0	0	0	0	0	0
Other	0	0	0	0	0	<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 CF, their genotype reveals which mutations of the CF gene cause their cystic fibrosis. Everyone living with CF 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.

824 (99.4%) patients have been genotyped with a recorded value.

DF508 Mutations; n (%)	
Homozygous DF508	359 (43.6)
Heterozygous DF508	387 (47.0)
No DF508 or both unidentified	78 (9.5)

1.31 Genotypes

Mutations				
Nucleotide	**Protein**	**Legacy name**	**N**	**%**
c.1521_1523delCTT	p. Phe508del	F508del	746	90.0
c.1652G->A	p. Gly551Asp	G551D	87	10.5
c.350G->A	p. Arg117His	R117H	61	7.4
c.1624G->T	p. Gly542X	G542X	59	7.1
c.200C->T	p. Pro67Leu	P67L	33	4.0
c.1679G->C	p. Arg560Thr	R560T	19	2.3
c.1585-1G->A		1717-1G->A	15	1.8
c.1477C->T	p. Gln493X	Q493X	14	1.7
c.489+1G->T		621+1G->T	12	1.4
c.3909C->G	p. Asn1303Lys	N1303K	12	1.4
c.3454G->C	p. Asp1152His	D1152H	11	1.3
c.2657+5G->A		2789+5G->A	9	1.1
c.3528delC	p. Lys1177SerfsX15	3659delC	9	1.1
c.178G->T	p. Glu60X	E60X	8	1.0
c.1558G->T	p. Val520Phe	V520F	6	0.7
c.948delT	p. Phe316LeufsX12	1078delT	6	0.7
c.3717+12191C->T		3849+10kbC->T	5	0.6
c.1210-12[5 (AJ574948.1:g.152T[5])		5T	5	0.6
c.1705T->G	p. Tyr569Asp	Y569D	5	0.6
c.1364C->A	p. Ala455Glu	A455E	<5	0.5
c.1766+1G->A		1898+1G->A	<5	0.5
c.1519_1521delATC	p. Ile507del	I507del	<5	0.5
c.3196C->T	p. Arg1066Cys	R1066C	<5	0.4
c.3140-26A->G		3272-26A->G	<5	0.2
c.3484C->T	p. Arg1162X	R1162X	<5	0.2
c.2988G->A		3120G->A	<5	0.2
c.254G->A	p. Gly85Glu	G85E	<5	0.2
c.2012delT	p. Leu671X	2143delT	<5	0.2
c.223C->T	p. Arg75X	R75X	<5	0.2
c.2657+2_2657+3insA		2789+2insA	<5	0.2
c.2988+1G->A		3120+1G->A	<5	0.2
c.2052delA	p. Lys684AsnfsX38	2184delA	<5	0.2
c.3846G->A	p. Trp1282X	W1282X	<5	0.2
c.1657C->T	p. Arg553X	R553X	<5	0.1
c.1466C->A	p. Ser489X	S489X	<5	0.1
c.2583delT	p. Phe861LeufsX3	2711delT	<5	0.1
c.579+3A->G		711+3A->G	<5	0.1
c.1209+1G->A		1341+1G->A	<5	0.1
c.3705T->G	p. Ser1235Arg	S1235R	<5	0.1
c.274G->A	p. Glu92Lys	E92K	<5	0.1
c.3884_3885insT	p. Ser1297PhefsX5	4016insT	<5	0.1
c.579+1G->T		711+1G->T	<5	0.1
c.3276C->A	p. Tyr1092X	Y1092X(C->A)	<5	0.1
c.1329_1330insAGAT	p. Ile444ArgfsX3	1461ins4	<5	0.1
c.273+1G->A		405+1G->A	<5	0.1
c.2490+1G->A		2622+1G->A	<5	0.1
Other selected			101	12.2

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 of 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 CF centres. This allows people with CF, their families, and healthcare providers to compare centres with 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 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 42.

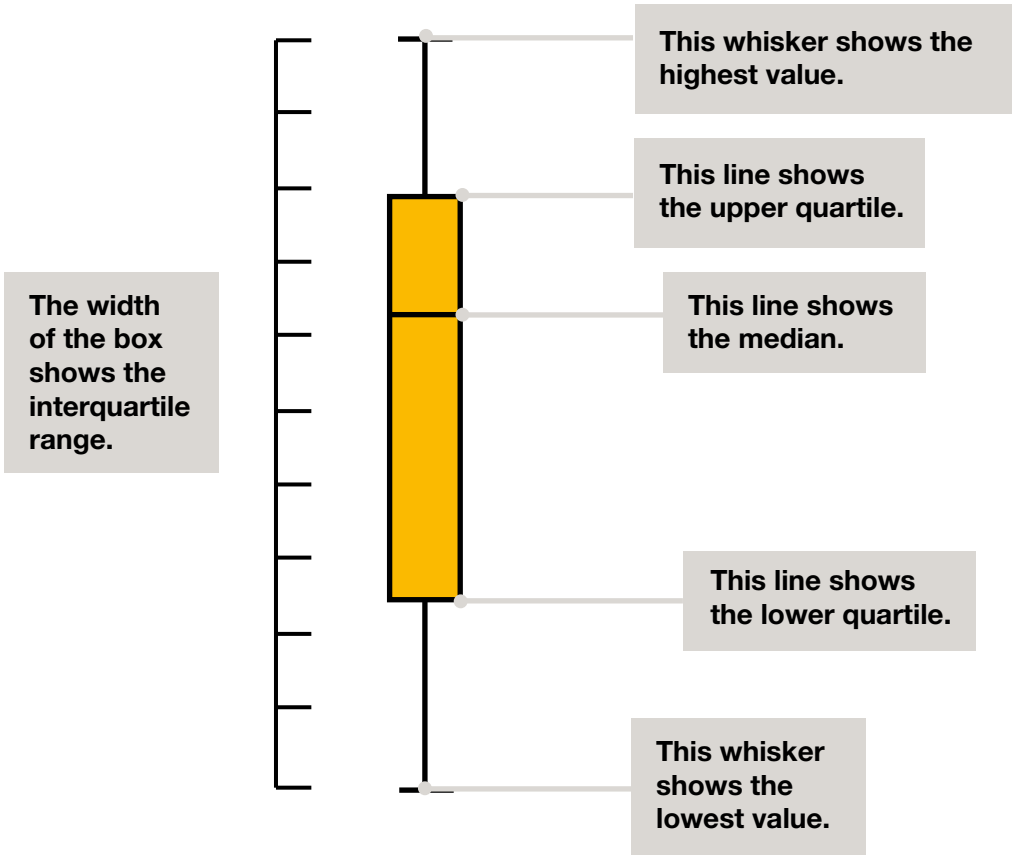
Key


Paediatric centre


Adult centre

A guide to the charts

Box plots

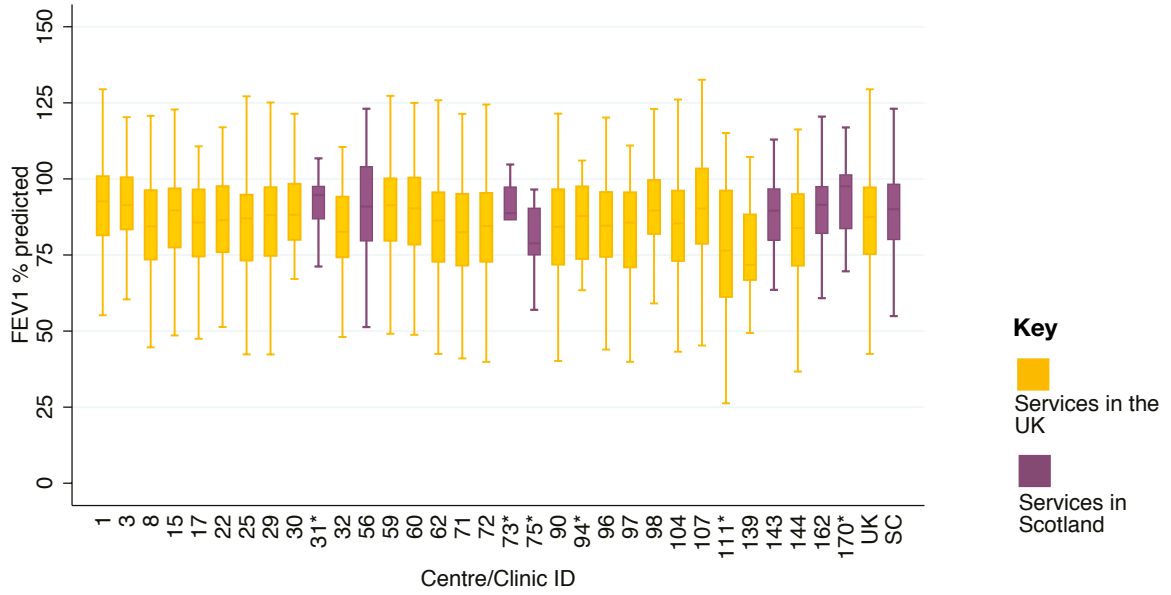


- The ‘box’ shows the middle half of the data for that centre, going from the first quartile to the third quartile. The longer the box, the more varied the data for that centre.
- The horizontal line within the box shows the median result for that centre.
- The ‘whiskers’ above and below the box shows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

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

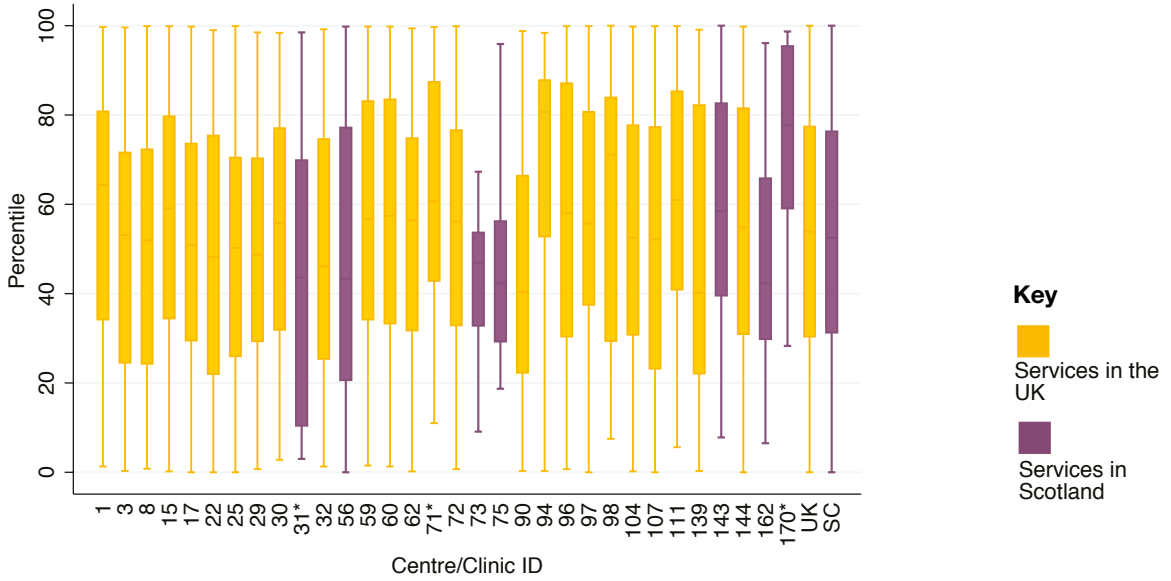
2.1 Median FEV_1 % predicted among patients aged 6 and older by paediatric centre/clinic (without a history of lung transplant) (GLI equations)



The median FEV_1 % predicted of patients attending paediatric centres/clinics in Scotland is 90% predicted (IQR: 80-98).

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

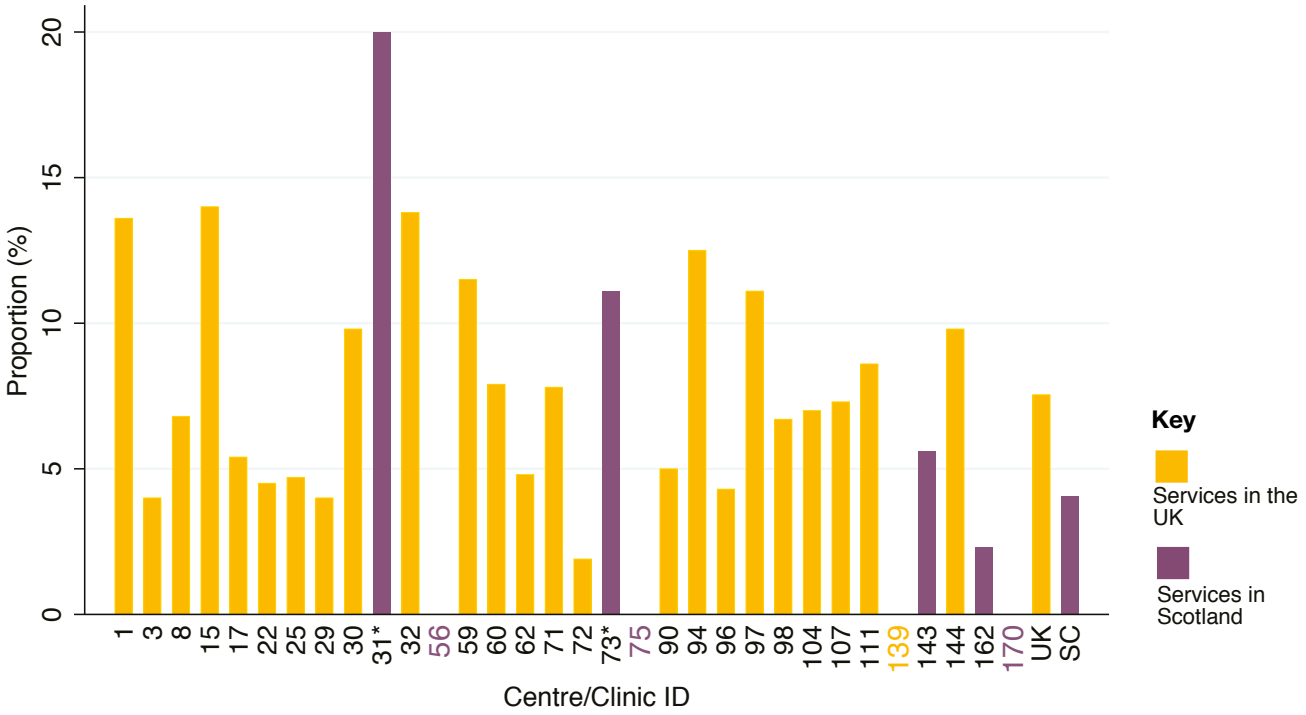
2.2 Median Body Mass Index (BMI) percentile among patients aged 2-15 years by paediatric centre/clinic



The median BMI percentile of patients attending paediatric centres/clinics in Scotland is 53 (IQR: 31-76).

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

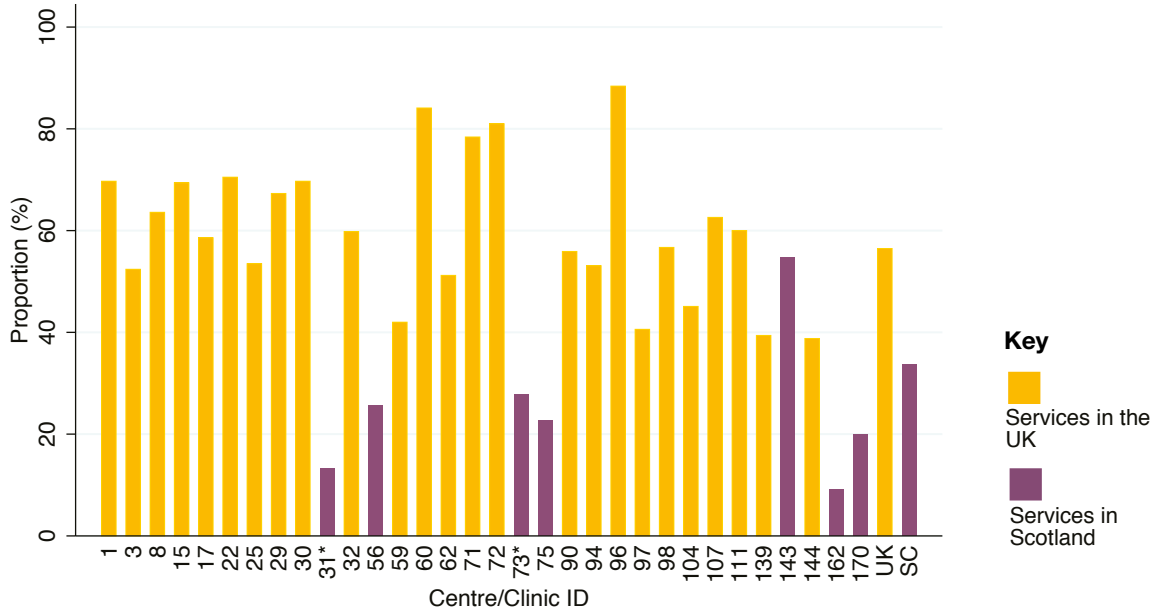
2.3 Proportion of patients with chronic *P. aeruginosa* by paediatric centre/clinic



The proportion of patients attending paediatric centres/clinics in Scotland with chronic *P. aeruginosa* is 4%.

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

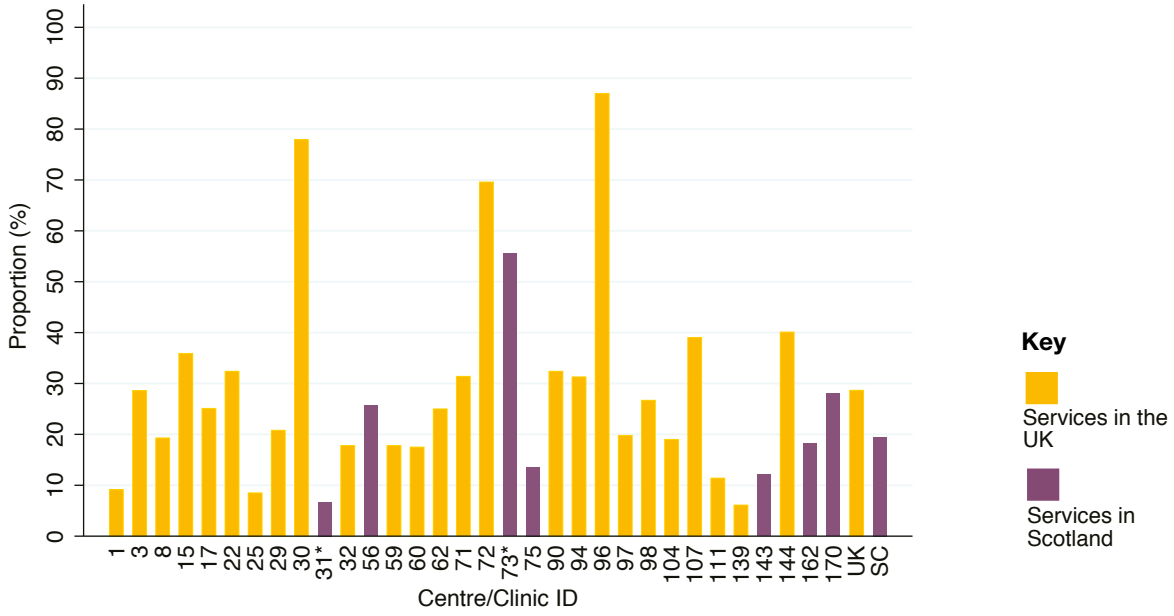
2.4 Proportion of patients receiving DNase treatment by paediatric centre/clinic



The proportion of patients attending paediatric centres/clinics in Scotland receiving DNase treatment is 33%.

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

2.5 Proportion of patients receiving hypertonic saline treatment by paediatric centre/clinic

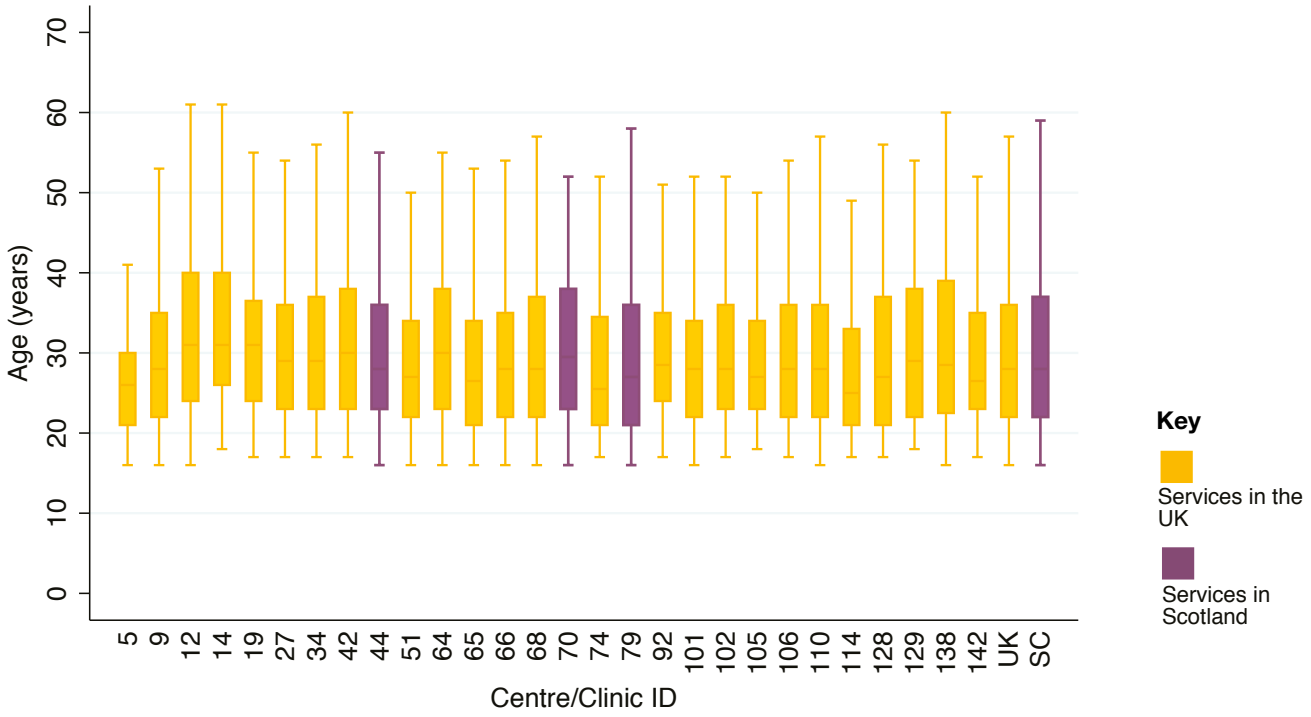


The proportion of patients attending paediatric centres/clinics in Scotland receiving hypertonic saline treatment is 19.5%.

Section 2b: Adult Centres Analysis

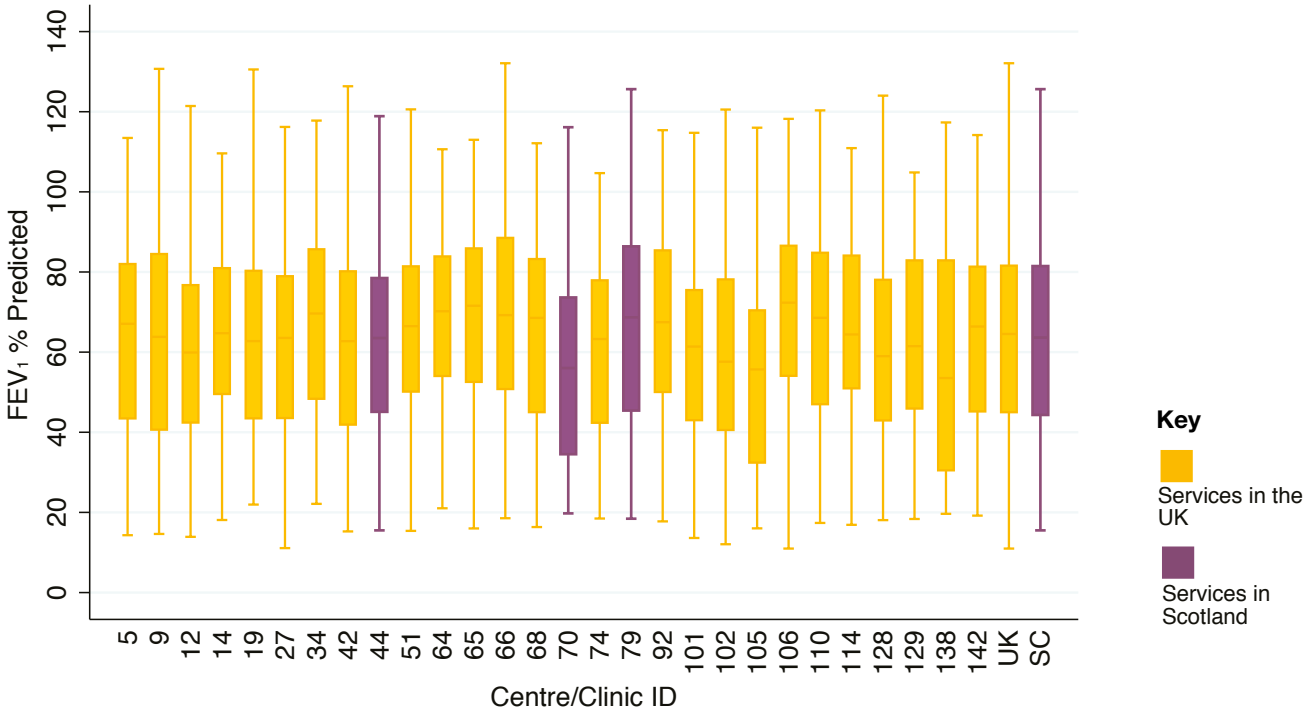
This section shows results for the three adult centres with any network clinics.

2.6 Median age (years) by adult service



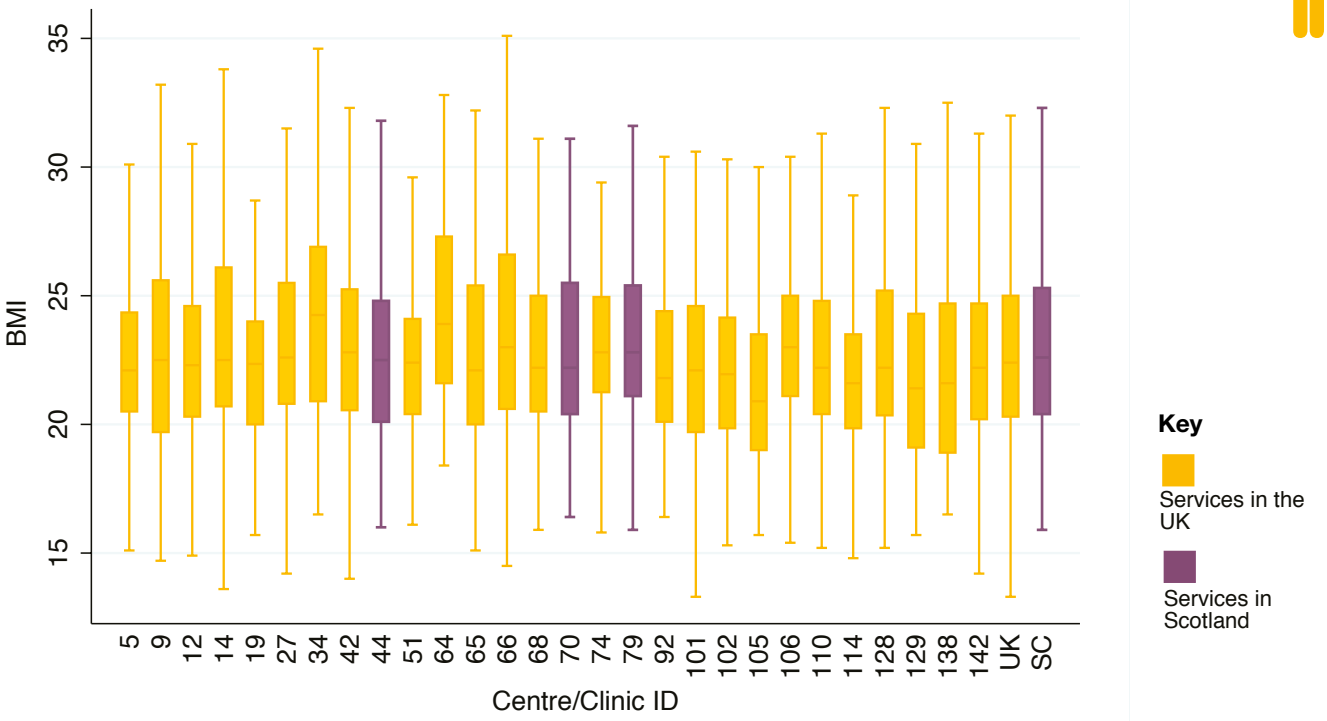
The median age of patients attending adult services in Scotland is 28 years (IQR: 22-37).

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



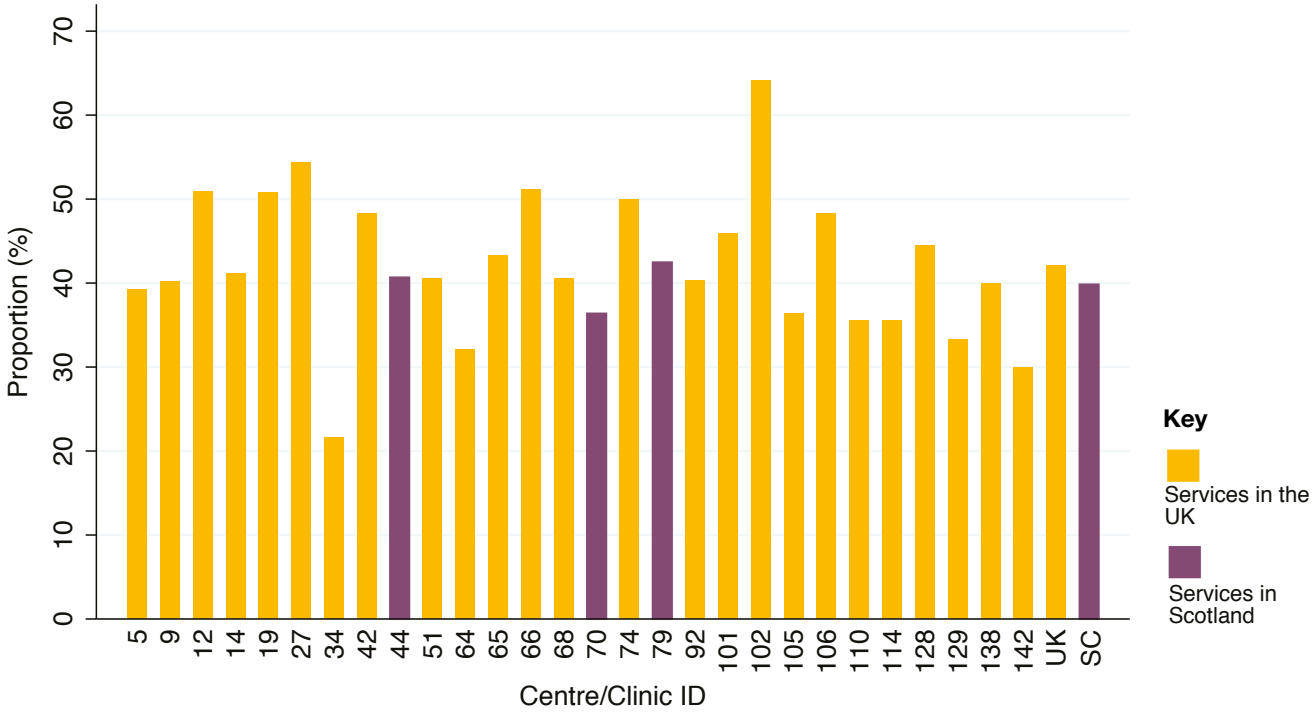
The median FEV_1 % predicted of patients attending adult services in Scotland is 65% (IQR: 45 – 82).

2.8 Median BMI among patients aged 16 years and older by adult service



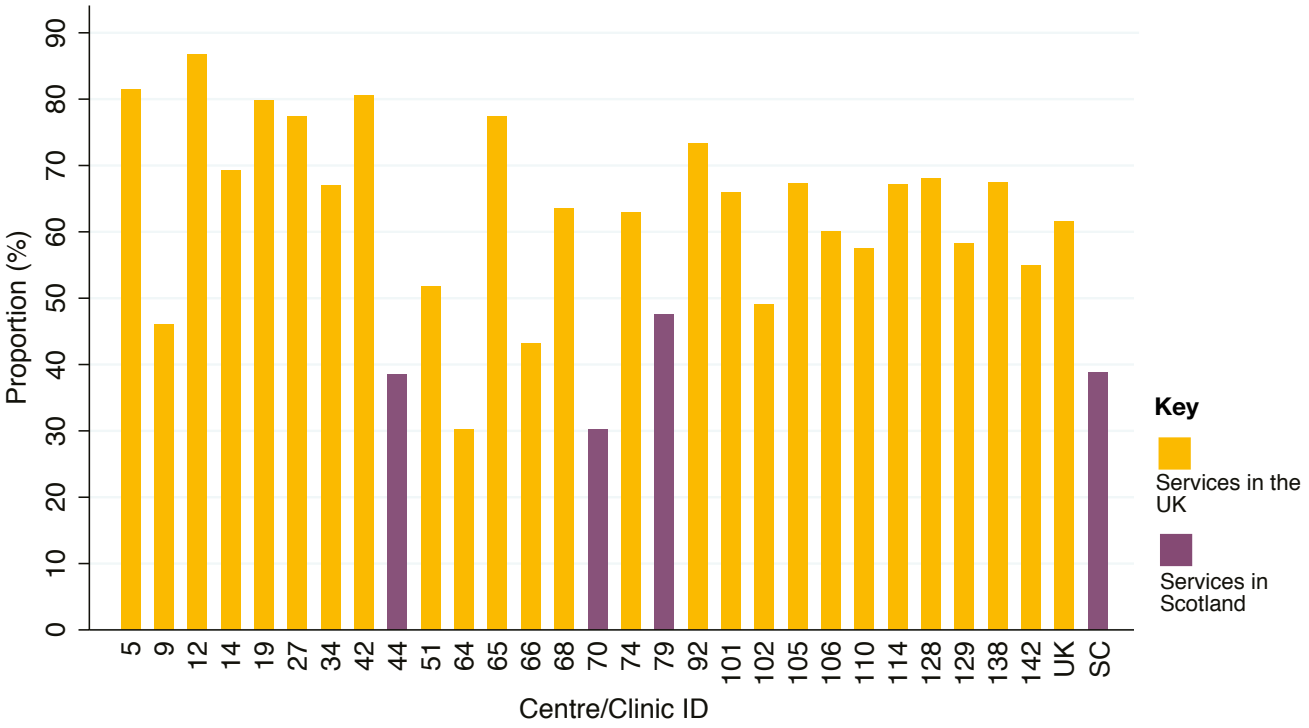
The median BMI of patients attending adult services in Scotland is 23 (IQR: 20 – 25).

2.9 Proportion of patients with chronic *P. aeruginosa* by adult service



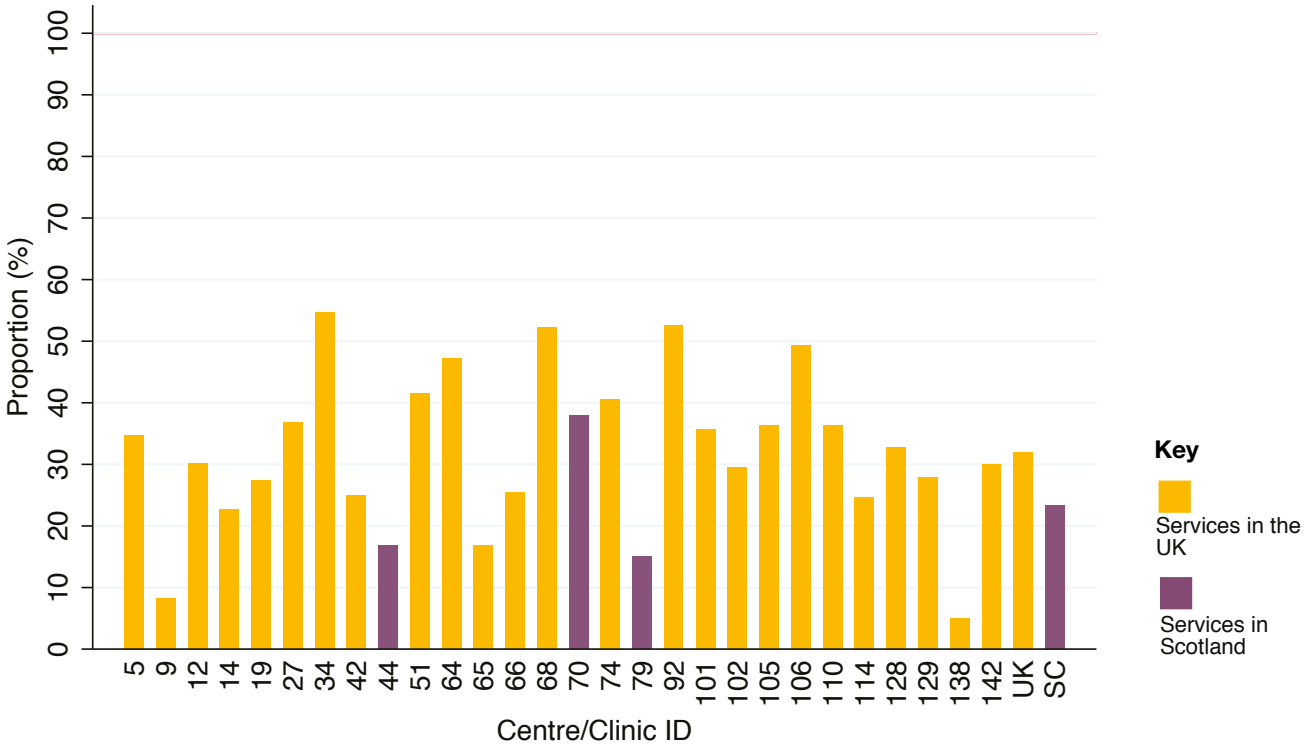
The proportion of patients attending adult services in Scotland with chronic *P. aeruginosa* is 40%.

2.10 Proportion of patients receiving DNase treatment by adult service



The proportion of patients attending adult services in Scotland receiving DNase treatment is 39%.

2.11 Proportion of patients receiving hypertonic saline treatment by adult service



The proportion of patients attending adult services in Scotland receiving hypertonic saline treatment is 23%.

Glossary

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

Word/Phrase	Meaning
2016	1 January 2016 – 31 December 2016.
ABPA (Allergic Bronchopulmonary Aspergillosis)	When a person develops a respiratory allergic reaction to 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 wheezing.
***B. cepacia* complex**	*Burkholderia cepacia* complex is 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)	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.
CI (Confidence Interval)	A way of expressing how certain we are about our statistical estimates of a clinical measure (eg BMI). It gives a range of results that is likely to include the 'true' value for the population. A narrow confidence interval indicates a more precise estimate. A wide confidence interval indicates more uncertainty about the true value of the clinical measure - often because a small group of patients has been studied. The confidence interval is usually stated as '95% CI', which means that the range of values has a 95 in 100 chance of including the 'true' value.
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 equations	Global Lung Function Initiative (GLI) is the equation used for calculating FEV_1% predicted from absolute FEV_1 that takes into account age, gender, height and ethnicity.
H. influenza	*Haemophilus influenza* is a bacterium that can cause serious illness.
Haemoptysis	The coughing up of blood.
Hepatobiliary disease	A liver or biliary disorder.
Heterozygous	Everyone living with CF has two 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 CF has two 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.
Mean	A type of average, calculated by adding up all the values and dividing by the number of values.
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 2016 the median age of the 148 people who died was 31.
Median predicted survival	A mathematical formula that predicts how long we expect half of people with CF born today to live. Half of people born today are predicted to live to at least 47 years. Half of people are therefore 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.
NBS	Newborn screening is part of the heel prick blood spot testing done at 5-7 days of age. The blood sample is tested for a number of conditions, including cystic fibrosis.
Nontuberculous Mycobacteria (NTM)	A mycobacterium that does not cause tuberculosis, but which can cause respiratory infection. There are several types known.

Word/Phrase	Meaning
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	Also '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 at 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 with the condition in the last 12 months.
Pseudomonas aeruginosa	A tough bacterial strain. Rarely affecting healthy people, it can cause a wide range of infections, particularly in those with a weakened immune system.
Rectal prolapse	When the rectal wall slides through the anus.
Renal	Relating to the kidneys.
S. aureus	*Staphylococcus aureus* is a bacterium that can cause disease if it enters the body.
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 1: Centre-level data tables

Paediatric centres/clinics providing data in 2016 – ordered by clinic ID

Location	Centre/clinic	ID	Active patients	Patients with 2016 data	FEV_1% predicted (≥6 years) (at annual review)		Best FEV_1% predicted (≥6 years)	
					Mean	Median	Mean	Median
Inverness	Raigmore Hospital	31	17	15	92.0	94.7	95.2	96.5
Glasgow	Royal Hospital for Sick Children	56	93	70	90.7	90.9	96.4	94.3
Dundee	Ninewells Hospital	73	23	18	87.9	88.8	95.1	97.4
Aberdeen	Royal Aberdeen Children's Hospital	75	30	22	78.3	78.8	83.4	85.5
Edinburgh	Royal Hospital for Sick Children	143	132	124	89.2	89.6	92.5*	94.2
Lanarkshire	Wishaw General Hospital	162	45	44	90.8	91.5	95.0	96.9
Ayr	University Hospital Crosshouse	170	27	25	92.5	97.6	99.9	98.7

* 'Best' FEV_1% predicted data for these sites was less than 80% complete. Where 'Best' values were missing, FEV_1% predicted taken at annual review was used.

Adult centres/clinics providing data in 2016 – ordered by clinic ID

Location	Centre/clinic	ID	Active patients	Patients with 2016 data	FEV_1% predicted (≥16 years) (at annual review)		Best FEV_1% predicted (≥16 years)	
					Mean	Median	Mean	Median
Edinburgh	Western General Hospital	44	235	226	62.9	63.5	68.1	68.1
Aberdeen	Aberdeen Royal Infirmary	70	71	66	57.1	56.0	61.9	58.6
Glasgow	Gartnavel General Hospital	79	235	219	66.7	68.7	72.3	72.6

* 'Best' FEV_1% predicted data for these sites was less than 80% complete. Where 'Best' values were missing, FEV_1% predicted taken at annual review was used.

	BMI percentile (2-15 years)		Chronic Pseudomonas		Receiving DNase treatment		Receiving hypertonic saline treatment	
	Mean	Median	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)
	44.8	43.6	<5	20.0	<5	13.3	<5	6.7
	47.9	43.3	0	0.0	18	25.7	18	25.7
	46.0	46.8	<5	11.1	5	27.8	10	55.6
	47.6	42.3	0	0.0	5	22.7	<5	13.6
	59.3	58.5	7	5.6	68	54.8	15	12.1
	46.5	42.3	<5	2.3	<5	9.1	8	18.2
	73.4	77.7	0	0.0	5	20.0	7	28.0

	BMI percentile (≥16 years)		Chronic Pseudomonas		Receiving DNase treatment		Receiving hypertonic saline treatment	
	Mean	Median	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)
	22.9	22.4	92	40.7	87	38.5	38	16.8
	23.0	22.2	24	36.4	20	30.3	25	37.9
	23.2	22.5	93	42.5	104	47.5	33	15.1

Appendix 1: Centre-level data tables

Paediatric centres/clinics providing data in 2016 – ordered alphabetically

Location	Centre/clinic	ID	Active patients	Patients with 2016 data	FEV_1% predicted (≥6 years) (at annual review)		Best FEV_1% predicted (≥6 years)	
					Mean	Median	Mean	Median
Aberdeen	Royal Aberdeen Children's Hospital	75	30	22	78.3	78.8	83.4	85.5
Ayr	University Hospital Crosshouse	170	27	25	92.5	97.6	99.9	98.7
Dundee	Ninewells Hospital	73	23	18	87.9	88.8	95.1	97.4
Edinburgh	Royal Hospital for Sick Children	143	132	124	89.2	89.6	92.5*	94.2
Glasgow	Royal Hospital for Sick Children	56	93	70	90.7	90.9	96.4	94.3
Inverness	Raigmore Hospital	31	17	15	92.0	94.7	95.2	96.5
Lanarkshire	Wishaw General Hospital	162	45	44	90.8	91.5	95.0	96.9

* 'Best' FEV_1% predicted data for these sites was less than 80% complete. Where 'Best' values were missing, FEV_1% predicted taken at annual review was used.

Adult centres/clinics providing data in 2016 – ordered alphabetically

Location	Centre/clinic	ID	Active patients	Patients with 2016 data	FEV_1% predicted (≥16 years) (at annual review)		Best FEV_1% predicted (≥16 years)	
					Mean	Median	Mean	Median
Aberdeen	Aberdeen Royal Infirmary	70	71	66	57.1	56.0	61.9	58.6
Edinburgh	Western General Hospital	44	235	226	62.9	63.5	68.1	68.1
Glasgow	Gartnavel General Hospital	79	235	219	66.7	68.7	72.3	72.6

* 'Best' FEV_1% predicted data for these sites was less than 80% complete. Where 'Best' values were missing, FEV_1% predicted taken at annual review was used.

	BMI percentile (2-15 years)		Chronic Pseudomonas		Receiving DNase treatment		Receiving hypertonic saline treatment	
	Mean	Median	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)
	47.6	42.3	0	0.0	5	22.7	<5	13.6
	73.4	77.7	0	0.0	5	20.0	7	28.0
	46.0	46.8	<5	11.1	5	27.8	10	55.6
	59.3	58.5	7	5.6	68	54.8	15	12.1
	47.9	43.3	0	0.0	18	25.7	18	25.7
	44.8	43.6	<5	20.0	<5	13.3	<5	6.7
	46.5	42.3	<5	2.3	<5	9.1	8	18.2

	BMI percentile (≥16 years)		Chronic Pseudomonas		Receiving DNase treatment		Receiving hypertonic saline treatment	
	Mean	Median	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)
	23.0	22.2	24	36.4	20	30.3	25	37.9
	22.9	22.4	92	40.7	87	38.5	38	16.8
	23.2	22.5	93	42.5	104	47.5	33	15.1

Appendix 2: UK CF Registry Steering Committee structure

UK CF Registry Steering Committee

Role	Forename	Surname	Organisation
Director of Impact	Keith	Brownlee	Cystic Fibrosis Trust
CF physician – paediatrics*	Siobhán	Carr	Royal Brompton Hospital
Senior Statistician †	Susan	Charman	Cystic Fibrosis Trust
Registry lead	Rebecca	Cosgriff	Cystic Fibrosis Trust
Cystic fibrosis centre data manager	Lance	Dennard	Lewisham Hospital
CF physician - paediatrics	Iolo	Doull	Children's Hospital for Wales
CF physician - adults	Caroline	Elston	King's College Hospital
Commissioner, England	Carrie or Sue	Gardner Sawyer	NHS England
CF physician - adults	Caroline	Elston	King's College Hospital
Registry data manager †	Elaine	Gunn	Cystic Fibrosis Trust
Commissioner, Wales †	Claire	Nelson	NHS Wales
Allied health professional	Alan	Peres	Royal Brompton Hospital
CF physician - adults	Simon	Range	Glenfield Hospital
Commissioner, Scotland	David	Steele	NHS Scotland
Person with CF	James	Thomson	N/A
Parent representative	Grant	Valentine	N/A
Chair of the Research Committee #	Martin	Wildman	Northern General Hospital
Registry Development Manager †	Mary	Yip	Cystic Fibrosis Trust

UK CF Registry Research Committee

Role	Forename	Surname	Organisation
Pharmacovigilance PI CF physician – adults (retired)	Diana	Bilton	Royal Brompton Hospital
Registry consultant	Noreen	Caine	Cystic Fibrosis Trust
Pharmacovigilance PI CF physician - paediatrics	Siobhán	Carr	Royal Brompton Hospital
Senior statistician †	Susan	Charman	Cystic Fibrosis Trust
Registry lead	Rebecca	Cosgriff	Cystic Fibrosis Trust
Pharmacovigilance PI CF physician - paediatrics	Steve	Cunningham	Royal Hospital for Sick Children
Parent representative	Marian	Dmochowska	N/A
Registry data manager †	Elaine	Gunn	Cystic Fibrosis Trust
Person with CF	Dominic	Kavanagh	Cystic Fibrosis Trust
CF physician - adults*#	Martin	Wildman	Northern General Hospital
Registry coordinator †	Mary	Yip	Cystic Fibrosis Trust

* Chair † Non-voting member # Caldicott guardian

Cystic Fibrosis Trust

cysticfibrosis.org.uk

Fighting for a Life Unlimited