Cystic Fibrosis strength in numbers

UK Cystic Fibrosis Registry
Annual Data Report 2018

Scotland

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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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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 UK. This includes people with CF, their families and clinical teams, healthcare managers, commissioners, and policy makers.

You can find a glossary of scientific and clinical terms on page **57**.

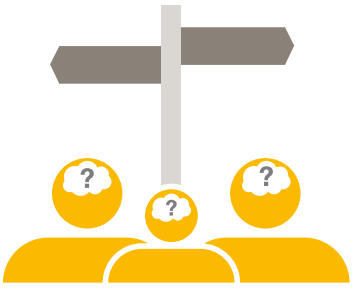
Cystic fibrosis

Cystic fibrosis is an inherited disease caused by a faulty gene known as 'CFTR'. The gene, and the protein it makes, help control the movement of salt and water in and out of cells. When the gene is faulty, it can cause thicker mucus. One of the main areas affected is the lungs; over time this thick mucus blocks and damages airways, leading to infections and making it hard to breathe. People with CF may develop other problems, such as liver disease or CF-related diabetes (CFRD). Around 85% of people with CF also have difficulty digesting food effectively.

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:

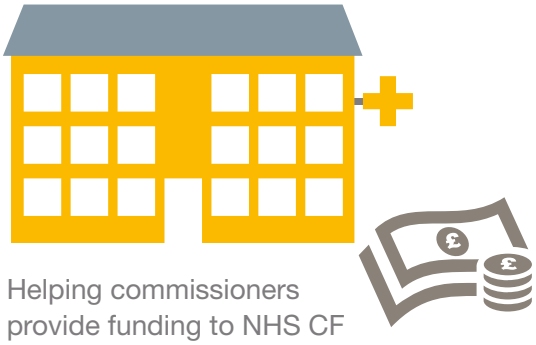


Helping people with CF and their families understand CF, and make informed decisions.

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 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 data protection legislation, and its Research Ethics Committee-approved Study Protocol. It also makes recommendations about the future development of the Registry. A sub-committee of the RSC, the Registry Research Committee, assesses applications for data and guides the Registry research strategy.

Please see Appendix 1 of the UK Cystic Fibrosis Registry 2018 Annual data Report

Data are only recorded on the UK CF Registry if explicit written consent is given by the person with CF 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 by the Registry team 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.

If requests from pharmaceutical companies are granted, for research or submissions to regulators or the NHS, the data are analysed and aggregated by Registry statisticians and only summary data are provided

Data collection

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

Where can I find more information?

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

Section 1: Scotland-wide analysis

This section provides an overview of the cystic fibrosis (CF) population, health outcomes, and care in the Scotland, with comparisons to the full CF population of United Kingdom, including CF centres in England, Northern Ireland, Scotland, and Wales.

1.1 Summary of the UK Cystic Fibrosis Registry

	2018	
	UK	**Scotland**
CF patients registered[1]	10509	927
Excluding diagnoses that year	10287	917
CF patients with an annual review; n(%)[2]	9847 (96%)	819 (89%)
Age in years; median[3]	20	22
All newly diagnosed patients (newborn screening and other)[4]	222	10
Number of patients born identified by newborn screening[4]	151	7
Age at diagnosis in months; median[3]	2	2
Adults aged 16 years and over; %[3]	60.4	62.9
Males; %[3]	53.3	53.6
Genotyped; %[3] (both mutations identified)	99.1	99.3
Total deaths reported (%)[5]	137 (1.3%)	22 (2.4%)
Age at death in years; median (95% CI)[5]	32 (29, 35)	26 (23, 34)

Notes:

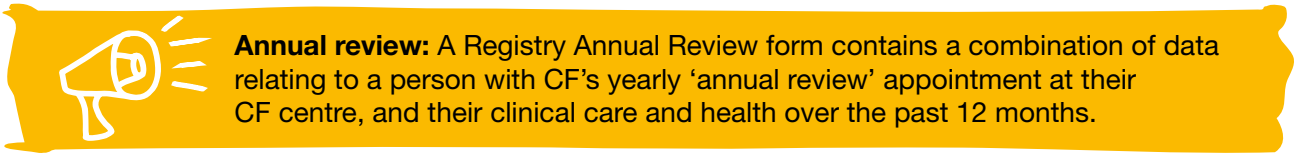
[1] Number of patients diagnosed with CF, seen in the past two years, and alive at 1 January in the given year.

[2] As patients newly diagnosed in a given year may not have their first annual review in the same year, the proportion with an annual review is calculated from the total registered excluding those diagnosed in the given year.

[3] Calculated from patients with an annual review in the given year (see footnote 2 above).

[4] Calculated from all patients registered on the database.

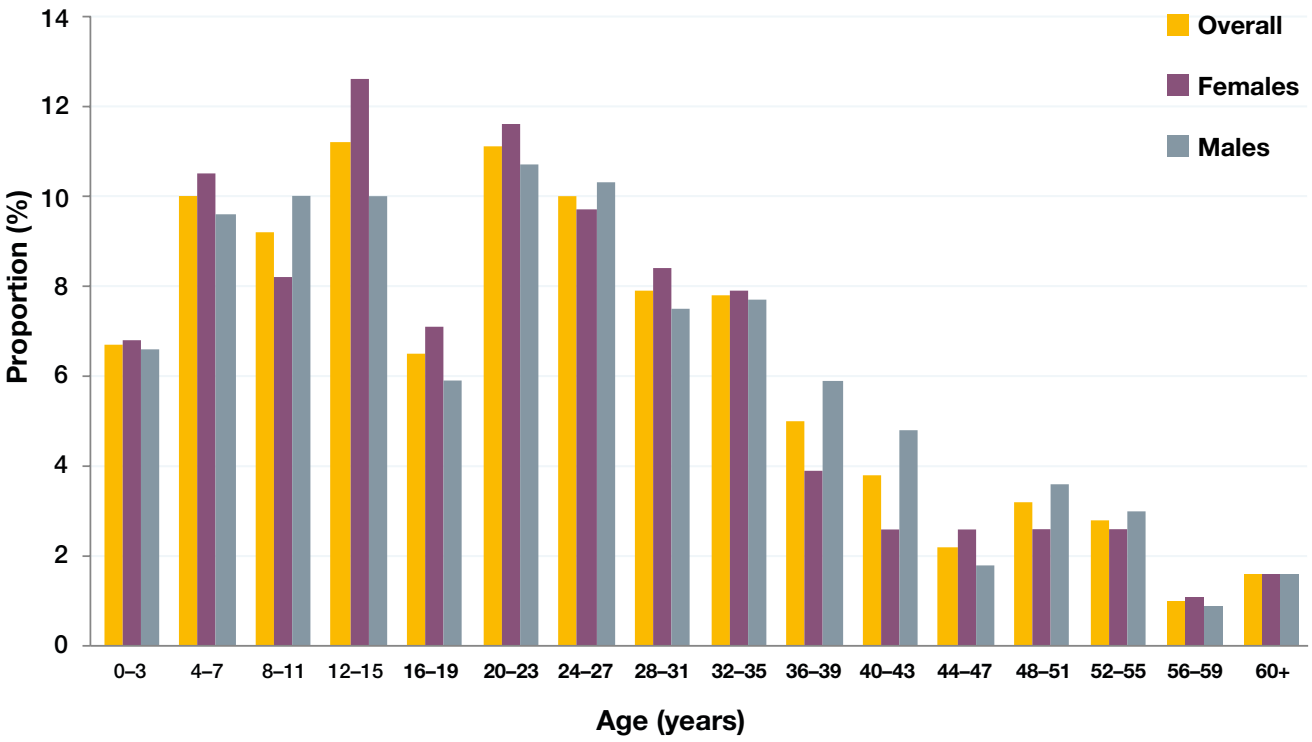
[5] Calculated from all registered patients who died in the given year.



Annual review: A Registry Annual Review form contains a combination of data relating to a person with CF's yearly 'annual review' appointment at their CF centre, and their clinical care and health over the past 12 months.

1.2 Age distribution by sex

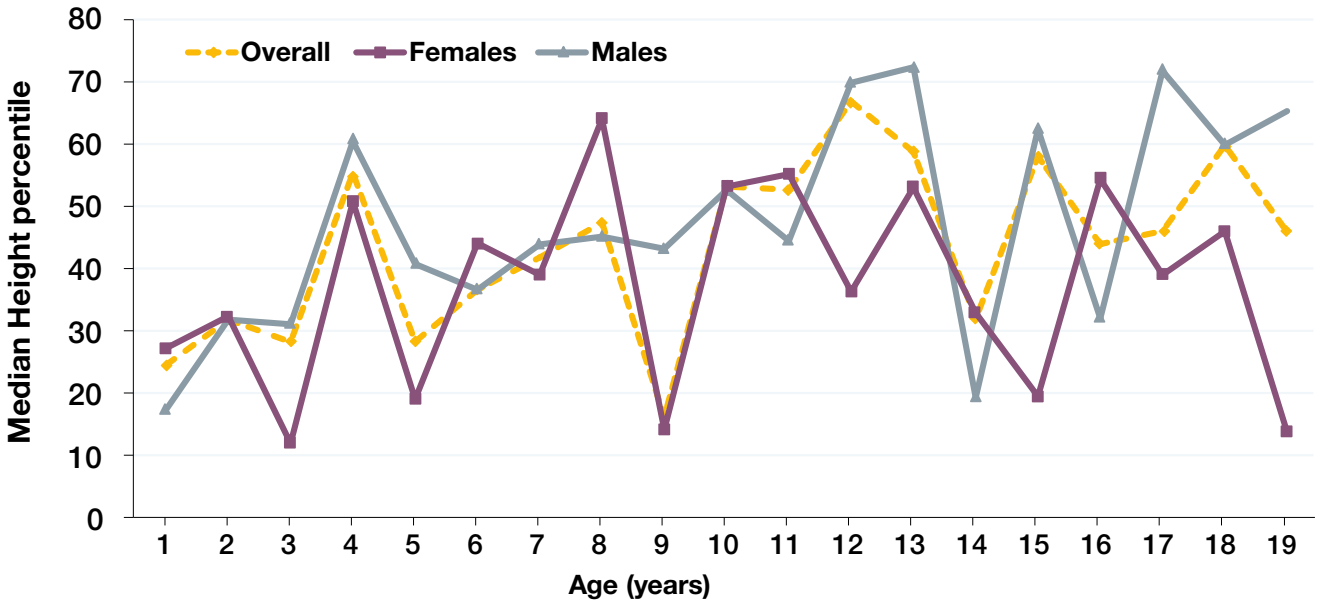
The following chart shows the mix of ages and genders in the CF population in Scotland.



Age	All; n (%)	Females; n (%)	Males; n (%)
0-3	55 (6.7)	26 (6.8)	29 (6.6)
4-7	82 (10.0)	40 (10.5)	42 (9.6)
8-11	75 (9.2)	31 (8.2)	44 (10.0)
12-15	92 (11.2)	48 (12.6)	44 (10.0)
16-19	53 (6.5)	27 (7.1)	26 (5.9)
20-23	91 (11.1)	44 (11.6)	47 (10.7)
24-27	82 (10.0)	37 (9.7)	45 (10.3)
28-31	65 (7.9)	32 (8.4)	33 (7.5)
32-35	64 (7.8)	30 (7.9)	34 (7.7)
36-39	41 (5.0)	15 (3.9)	26 (5.9)
40-43	31 (3.8)	10 (2.6)	21 (4.8)
44-47	18 (2.2)	10 (2.6)	8 (1.8)
48-51	26 (3.2)	10 (2.6)	16 (3.6)
52-55	23 (2.8)	10 (2.6)	13 (3.0)
56-59	8 (1.0)	<5 (-)	<5 (-)
60+	13 (1.6)	6 (1.6)	7 (1.6)
<16	304 (37.1)	145 (38.2)	159 (36.2)
≥16	515 (62.9)	235 (61.8)	280 (63.8)
<18	325 (39.7)	159 (41.8)	166 (37.8)
≥18	494 (60.3)	221 (58.2)	273 (62.2)
Overall	**819**	**380**	**439**

1.3 Height percentiles of children and young people (<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 UK growth data for the general population. If a person with CF is on the 40th percentile, only 40% of people the same age are their height or shorter; 60% are taller.



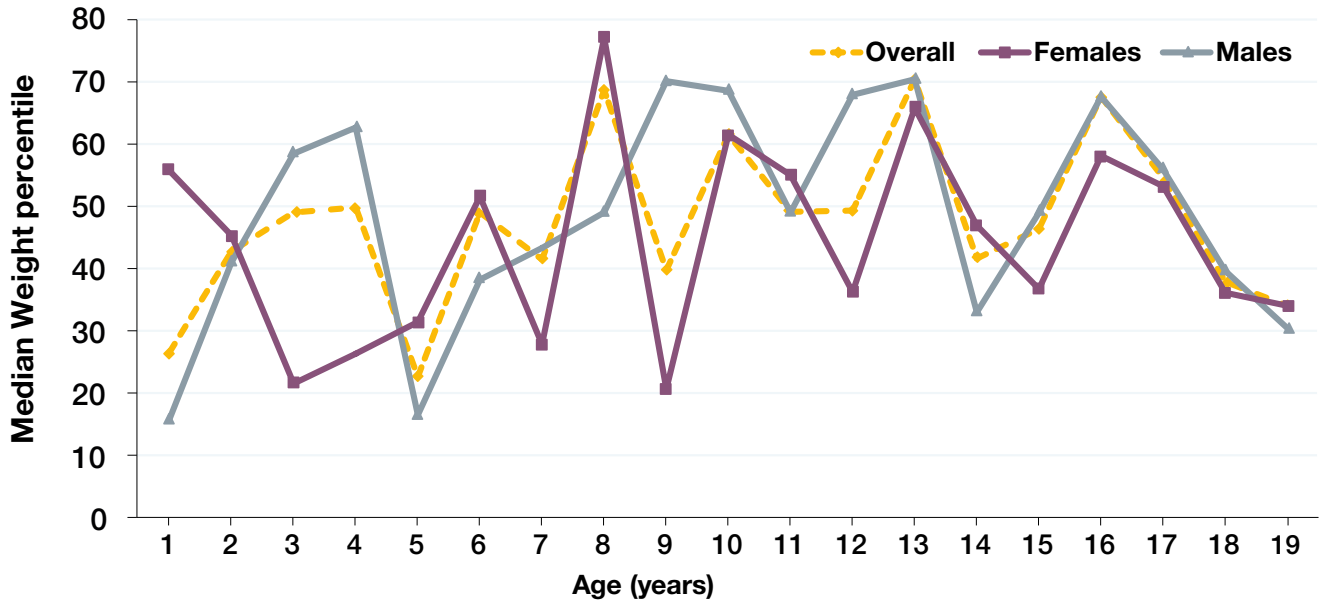
	Overall			Female			Male		
Age	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**
1	14	24.4	12.9-59.4	7	27.1	12.9-72.9	7	17.2	12.2-52.8
2	21	31.7	21.1-42.2	10	32.3	21.1-39.3	11	31.7	17.3-86.7
3	15	28.1	11.8-55.4	6	11.8	9.2-28.1	9	30.9	18.7-78.6
4	18	55.3	36.0-62.1	6	50.7	0.5-52.8	12	60.4	45.7-75.3
5	16	28.2	10.5-45.0	11	18.9	10.5-30.7	5	40.7	21.9-59.5
6	19	36.5	12.1-67.1	8	44.0	29.5-65.0	11	36.5	11.1-71.5
7	29	41.7	18.6-55.4	15	38.9	13.1-46.4	14	43.8	32.6-60.4
8	23	47.3	22.4-67.3	6	64.0	45.3-67.4	17	45.0	14.3-60.8
9	20	16.0	7.2-63.9	10	14.0	7.2-32.0	10	43.1	7.3-63.9
10	12	53.1	40.5-90.2	8	53.1	44.0-91.9	<5	52.5	25.4-77.4
11	20	52.6	19.5-66.9	7	55.1	21.0-63.3	13	44.3	14.4-68.2
12	31	66.8	25.3-77.9	18	36.2	21.8-73.6	13	69.8	45.0-85.1
13	22	58.7	52.3-84.2	9	53.0	29.1-70.6	13	72.3	53.2-84.7
14	22	31.8	14.9-63.4	13	32.8	23.3-63.4	9	19.1	9.8-56.8
15	17	57.9	19.4-71.8	8	19.4	10.6-54.7	9	62.3	57.9-72.6
16	10	43.8	17.0-60.3	5	54.3	29.0-76.6	5	31.9	14.9-49.9
17	11	46.0	16.0-71.7	-	38.9	16.0-61.0	<5	71.7	-
18	15	59.8	27.7-73.1	8	45.9	22.2-73.1	7	59.8	54.1-76.7
19	17	45.8	5.5-80.4	5	13.6	5.5-45.8	12	65.3	14.8-83.2
Overall	**352***	**44.6**	**16.5-68.5**	**-**	**37.4**	**13.6-63.4**	**-**	**51.8**	**19.9-73.1**

*number with non-missing data

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

1.4 Weight percentiles of children and young people (<20 years)[6] N=357

The following chart and table show the weight 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 people the same age are their weight or lower; 60% weigh more.



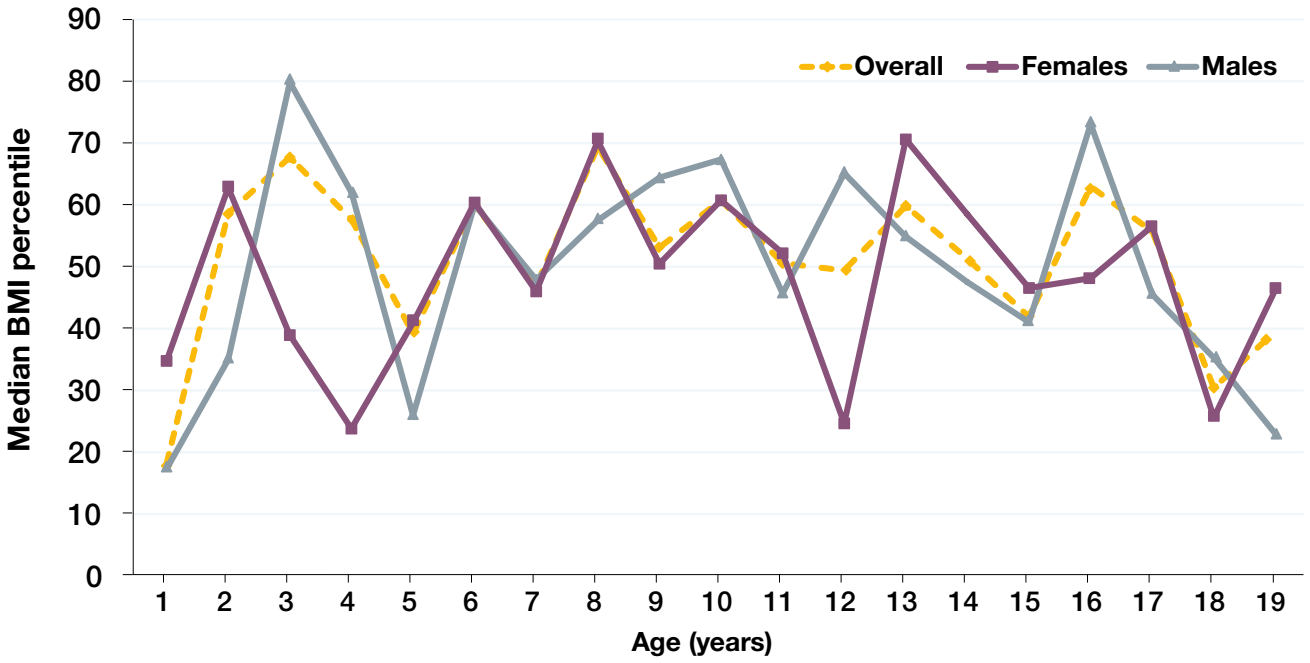
	Overall			Female			Male		
Age	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**
1	15	26.5	7.2-63.2	8	55.7	18.2-73.7	7	15.8	1.5-25.6
2	22	42.8	14.0-51.6	11	45.3	12.5-63.2	11	41.0	14.0-51.6
3	15	49.0	17.0-61.3	6	21.5	10.6-44.3	9	58.4	33.0-89.4
4	18	49.8	26.3-67.4	6	26.3	5.9-35.7	12	62.7	46.5-80.4
5	16	22.6	12.7-35.6	11	31.4	10.1-35.6	5	16.5	14.0-39.5
6	19	49.0	20.1-72.8	8	51.5	31.9-74.0	11	38.2	6.0-72.8
7	29	41.4	21.2-65.7	15	27.8	11.6-75.5	14	43.3	31.4-56.6
8	23	68.6	25.7-82.2	6	77.1	71.1-82.2	17	49.0	20.3-82.2
9	20	39.7	17.7-77.2	10	20.6	16.7-71.6	10	70.1	23.8-77.2
10	12	61.5	25.1-86.7	8	61.5	25.1-84.6	<5	68.6	28.9-89.8
11	20	49.1	24.0-66.8	7	55.0	35.3-91.7	13	49.0	23.0-55.0
12	31	49.3	30.0-67.9	18	36.2	16.6-51.2	13	67.9	49.6-85.5
13	22	70.4	37.6-80.2	9	65.8	41.1-81.1	13	70.4	27.1-80.2
14	22	41.6	12.5-61.8	13	46.9	12.5-67.7	9	33.0	13.9-56.5
15	17	46.3	26.2-57.1	8	36.7	21.2-62.3	9	49.0	35.3-57.1
16	10	67.6	22.8-87.7	5	58.1	18.4-87.7	5	67.6	35.9-86.0
17	11	54.6	37.5-73.2	-	53.2	37.5-73.2	<5	55.9	-
18	15	37.8	9.8-61.1	8	36.1	11.7-56.3	7	39.6	6.1-84.1
19	17	34.0	4.7-83.4	5	34.0	15.8-41.4	12	30.4	4.7-97.8
Overall	**354***	**46.9**	**20.1-71.8**	**-**	**44.6**	**17.4-70.6**	**-**	**50.1**	**21.3-72.8**

*number with non-missing data

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

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

The following chart and table show the BMI 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, it means that only 40% of the population at the same age are their BMI or lower; so 60% have a higher BMI.



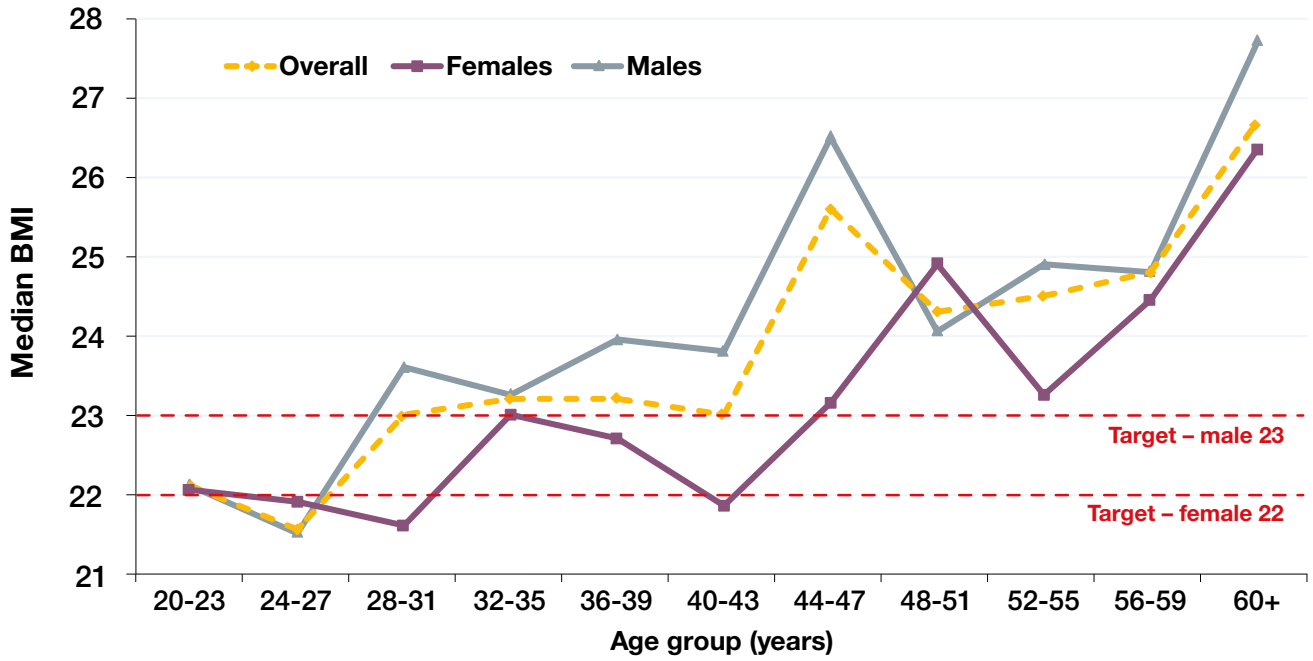
	Overall			Female			Male		
Age	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**
1	14	18.0	16.1-77.6	7	34.7	16.1-81.7	7	17.2	8.7-49.2
2	21	58.7	29.0-64.2	10	62.8	59.1-76.0	11	34.9	16.3-63.5
3	15	67.8	30.9-88.8	6	38.9	28.6-44.5	9	80.0	49.4-90.6
4	18	57.7	34.4-66.5	6	23.7	18.0-60.9	12	62.1	41.9-79.1
5	16	39.2	23.5-44.9	11	40.9	23.5-44.9	5	25.8	8.3-44.2
6	19	60.1	22.7-80.5	8	60.5	41.0-72.8	11	60.1	13.2-86.7
7	29	47.0	35.5-62.6	15	46.0	33.0-63.1	14	47.9	36.5-54.9
8	23	69.5	36.7-84.0	6	70.4	64.9-82.9	17	57.5	25.8-85.9
9	20	53.2	29.5-80.2	10	50.5	29.5-78.8	10	64.5	49.4-80.2
10	12	60.8	42.3-76.8	8	60.8	42.3-73.3	<5	67.4	37.1-86.6
11	20	50.6	32.5-65.7	7	52.2	30.2-89.2	13	45.7	34.0-59.8
12	31	49.4	21.3-66.5	18	24.5	3.6-63.8	13	65.2	49.4-76.2
13	22	59.9	39.5-78.2	9	70.7	51.8-82.8	13	54.9	26.9-78.2
14	22	51.3	17.0-66.1	13	58.5	14.0-85.3	9	47.5	29.5-59.8
15	17	41.8	25.8-57.4	8	46.6	28.5-86.8	9	41.0	22.6-44.2
16	10	63.1	31.5-90.3	5	48.2	17.9-80.1	5	73.2	46.5-91.7
17	11	55.8	45.8-71.7	-	56.6	47.8-71.7	<5	-	-
18	15	30.4	9.1-71.5	8	25.5	9.1-71.5	7	35.2	4.4-86.3
19	17	39.5	14.0-77.4	5	46.3	43.5-60.1	12	22.9	14.0-94.4
Overall	**352***	**50.6**	**26.4-74.6**	**-**	**52.2**	**25.5-74.6**	**-**	**49.3**	**26.5-76.2**

*number with non-missing data

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

1.6 Body Mass Index (BMI) in adults (20 years and over) N=462

The following chart and table show the BMI of people with CF aged 20 and over in relation to the target BMI for adults; 22 for women and 23 for men[7].



	Overall			Female			Male		
Age	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**
20-23	91	22.1	19.9-23.8	44	22.1	19.4-24.5	47	22.1	20.4-23.6
24-27	82	21.6	19.6-23.8	37	21.9	19.6-23.9	45	21.5	20.2-23.5
28-31	65	23.0	20.2-24.6	32	21.6	19.9-23.9	33	23.6	20.9-25.3
32-35	64	23.2	21.1-25.4	30	23.0	21.0-25.4	34	23.3	21.1-24.9
36-39	41	23.2	21.3-25.2	15	22.7	21.3-24.6	26	24.0	21.2-25.9
40-43	31	23.0	21.6-26.0	10	21.9	20.6-23.6	21	23.8	22.1-26.4
44-47	18	25.6	22.3-26.9	10	23.1	20.4-26.0	8	26.5	24.5-30.0
48-51	26	24.3	22.3-26.5	10	24.9	22.5-28.5	16	24.1	22.3-26.3
52-55	23	24.5	21.0-27.2	10	23.3	19.3-25.6	13	24.9	21.0-27.2
56-59	-	24.8	23.6-25.9	<5	26.4	23.5-27.1	<5	27.7	25.8-30.0
60+	13	26.7	25.8-29.5	6	26.4	23.5-27.1	7	27.7	25.8-30.0
Overall	**462***	**22.9**	**20.6-25.5**	**208**	**22.6**	**20.1-25.3**	**254**	**23.1**	**21.1-25.9**

*number with non-missing data

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

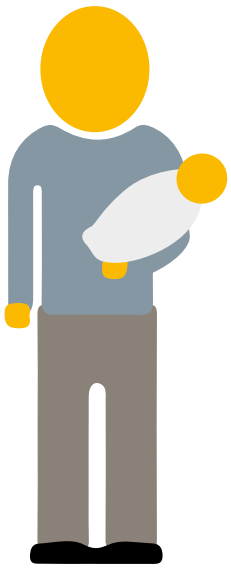
1.7 Education and employment in adults (16 years and over) N=515

The following table shows how people with CF reported their education and employment status in 2018. Please note that the groups are not mutually exclusive; for example, someone may be a student as well as working part-time.

	Overall number of patients n (%)	Male n (%)	Female n (%)
Number who completed questionnaire; n (%)	515 (100.0)	235 (100.0)	280 (100.0)
Full-time employment; n (%)	186 (36.1)	59 (25.1)	127 (45.4)
Part-time employment; n (%)	108 (21.0)	75 (31.9)	33 (11.8)
Student; n (%)	65 (12.6)	28 (11.9)	37 (13.2)
Homemaker; n (%)	15 (2.9)	15 (6.4)	0 (0.0)
Unemployed; n (%)	92 (17.9)	40 (17.0)	52 (18.6)
Disabled; n (%)	22 (4.3)	7 (3.0)	15 (5.4)
Retired; n (%)	17 (3.3)	9 (3.8)	8 (2.9)
Unknown entered; n (%)	10 (1.9)	<5 (-)	8 (2.9)
No. in work or study; n (%)	**359 (69.7)**	**162 (68.9)**	**197 (70.4)**

1.8 Pregnancy

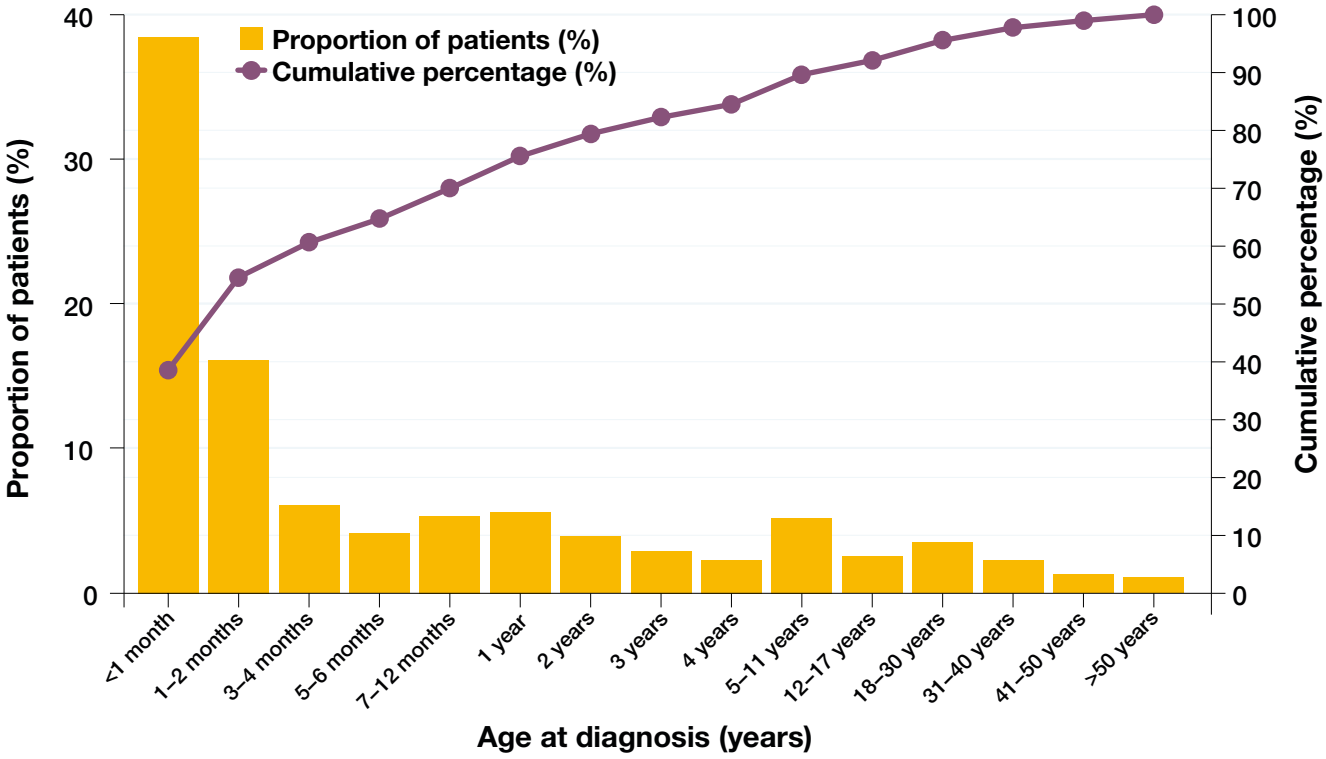
12 women with cystic fibrosis had babies in Scotland during 2018



Fewer than five men with cystic fibrosis became fathers in Scotland during 2018

Diagnosis of cystic fibrosis

1.9 Age at diagnosis and screening in children under 16 in 2018
N=304



The median (range) age at diagnosis for patients aged under 16 in 2018 is 20.5 days.

Newborn screening for CF has been done routinely in the whole of the UK since mid-2007. It 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. This means that more babies born after 2007 receive an early diagnosis than those born before.

A total of 7 patients born in 2018 were identified by newborn screening (including those without complete data).

84 (10.1%) of Scottish CF patients were diagnosed at age 16 or over. No new CF diagnoses were recorded in Scotland for people aged 16 or over during 2018.

1.10 Mode of presentation

The following table shows the number of patients diagnosed through each mode of presentation. Patients may present with multiple symptoms. The Venn diagram below shows the three most common modes of presentation excluding newborn screening (NBS), and the combinations of them.

	All patients	Age <16 at diagnosis*	Age ≥16 at diagnosis*
Total patients	819	731	84
Number diagnosed by NBS	243	243	0
Total non-NBS	**576**	**488**	**84**

Mode of presentation (excluding newborn screening)	All patients (n=819)		Under 16 (n=731)		Over 16 (n=84)	
Persistant or acute respiratory infection	209	36.3%	163	33.4%	46	54.8%
Failure to thrive/malnutrition	155	26.9%	155	31.8%	0	0.0%
Abnormal stools/fatty stool(steatorrhea)/ malabsorption	122	21.2%	117	24.0%	5	6.0%
Meconium ileus	91	15.8%	91	18.6%	0	0.0%
Family history	81	14.1%	70	14.3%	11	13.1%
Unknown	41	7.1%	33	6.8%	8	9.5%
Genotype	26	4.5%	19	3.9%	7	8.3%
Electrolyte imbalance	23	4.0%	20	4.1%	<5	3.6%
Rectal prolapse	16	2.8%	16	3.3%	0	0.0%
Bronchiectasis	-	-	<5	-	8	9.5%
Nasal polyps	5	0.9%	<5	0.6%	<5	2.4%
Prenatal	<5	-	<5	-	0	0.0%
Fertility	<5	-	0	0.0%	<5	-
Liver disease	<5	-	<5	-	0	0.0%
Pancreatitis	0	0.0%	0	0.0%	0	0.0%
Oedema	0	0.0%	0	0.0%	0	0.0%

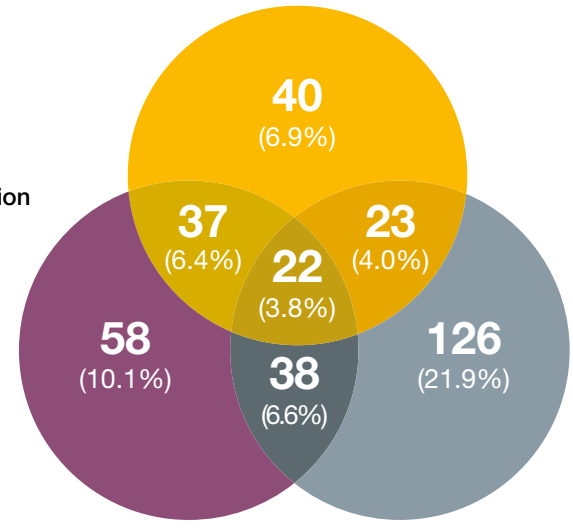
Top three non-NBS presentation routes

Abnormal stools/fatty stool (steatorrhea)/malabsorption

Failure to thrive/malnutrition

Persistent or acute respiratory infection

Other: 232 (40.3%)



*Age stratified figures are presented only for those with non-missing diagnosis date. This means that the number of people in <16 and ≥16 age groups will not necessarily add up to the 'All patients' number, which is shown for all patients, even if the diagnosis date is missing.

Lung health

For people with CF, mucus in the lungs is linked to repeat or chronic infections. This can cause permanent damage, making it harder to breathe.

In CF, the condition of the lungs is often measured using FEV_1; the Forced Expiratory Volume of air in the first second of a forced exhaled breath. In this report, an FEV_1% predicted is based on the FEV_1 we would expect for a person without CF 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. Each individual with CF will have their own FEV_1 target, based on their own lung function results and trends.

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, dietitians, and psychologists.

The FEV_1% predicted values shown in this report are calculated using an equation called Global Lungs Initiative, or 'GLI'[8]

[8] Quanjer PH et al. Eur respir J. 2012 40(6):1324-1343

1.11 FEV_1 % predicted (GLI equations) in patients aged 6 years and older who have not had a lung transplant
N=698

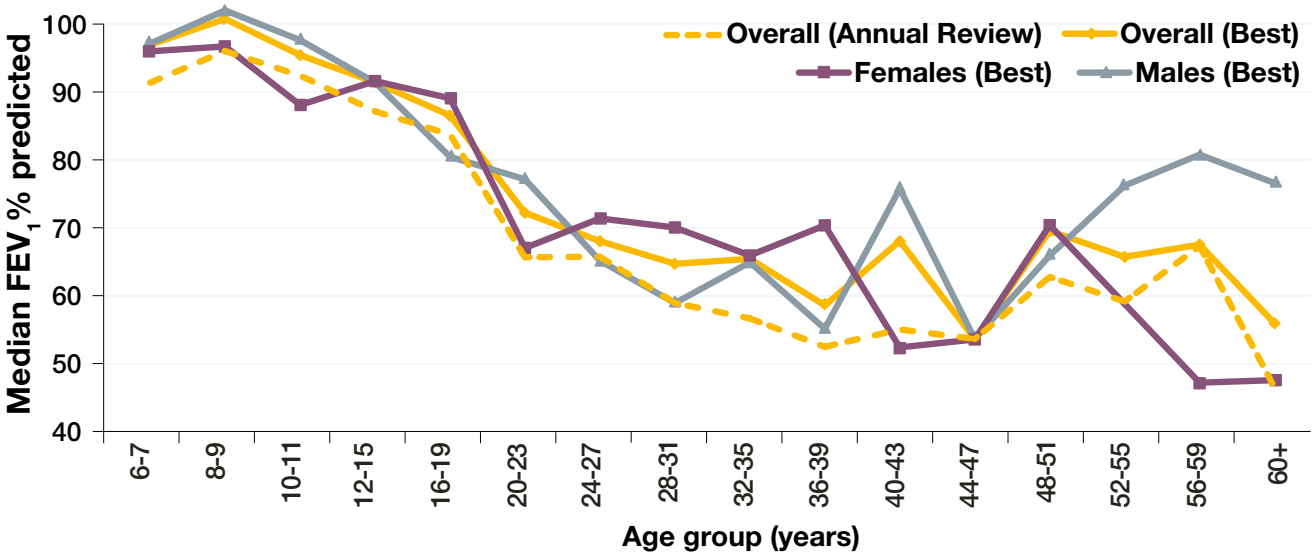
People with CF who have had lung transplants are excluded, as their new 'non-CF' lungs may have lung health similar to a person without cystic fibrosis.

	Overall			Female			Male		
Age (yrs)	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**
6-7	45	91.1	82.8-99.8	21	89.7	83.6-105.5	24	92.3	79.0-98.2
8-9	39	95.7	82.2-107.2	14	94.1	79.4-100.9	25	95.8	88.7-109.0
10-11	31	92.1	76.7-99.5	15	78.8	69.8-97.6	16	94.7	88.5-100.4
12-15	90	86.9	77.7-95.8	47	86.7	77.1-98.5	43	87.1	79.0-94.4
16-19	51	83.5	58.5-93.2	27	84.7	63.2-96.4	24	79.0	50.9-92.8
20-23	90	65.5	42.9-86.2	43	61.5	36.6-84.2	47	68.0	48.0-87.0
24-27	77	65.6	43.4-77.5	35	66.2	45.9-80.2	42	64.2	36.7-76.8
28-31	62	58.8	45.8-78.2	30	64.5	53.5-86.4	32	54.8	36.8-71.9
32-35	58	56.6	45.2-73.6	25	51.2	49.1-65.8	33	59.5	44.3-77.4
36-39	34	52.4	38.3-82.9	13	61.1	47.4-88.5	21	51.2	35.4-73.2
40-43	27	55.0	43.1-85.9	10	45.7	36.6-55.0	17	74.8	51.2-91.3
44-47	14	53.6	27.9-68.0	8	52.3	28.1-67.1	6	53.6	27.7-83.0
48-51	25	62.7	52.5-75.9	10	57.5	53.3-75.9	15	63.0	38.3-81.3
52-55	20	59.1	45.5-75.6	10	54.8	41.1-66.6	10	68.8	52.4-80.4
56-59	7	67.2	42.8-81.1	<5	46.8	42.8-67.2	<5	76.2	55.3-88.0
60+	12	46.5	35.2-74.3	5	41.0	22.1-47.5	7	73.9	44.3-76.8
<16	205	90.9	79.4-99.1	97	87.1	78.0-99.1	108	91.3	82.0-99.2
≥16	477	62.7	45.0-82.6	219	60.4	46.2-82.6	258	64.9	43.4-82.7
<18	225	89.9	78.8-98.9	111	87.1	77.1-99.1	114	91.2	80.7-98.9
≥18	457	61.7	44.3-81.1	205	58.9	45.9-79.7	252	64.2	43.0-81.3
Overall	**682***	**74.4**	**50.9-91.4**	**316**	**72.8**	**51.1-90.3**	**366**	**75.3**	**50.4-91.7**

*number with non-missing data

1.12 Best FEV_1% predicted (GLI equations) in patients aged 6 years and older who have not had a lung transplant
N=698

For the best FEV_1 calculation, where best FEV_1% was missing or less than the FEV_1% at annual review,the annual review FEV_1% was used.

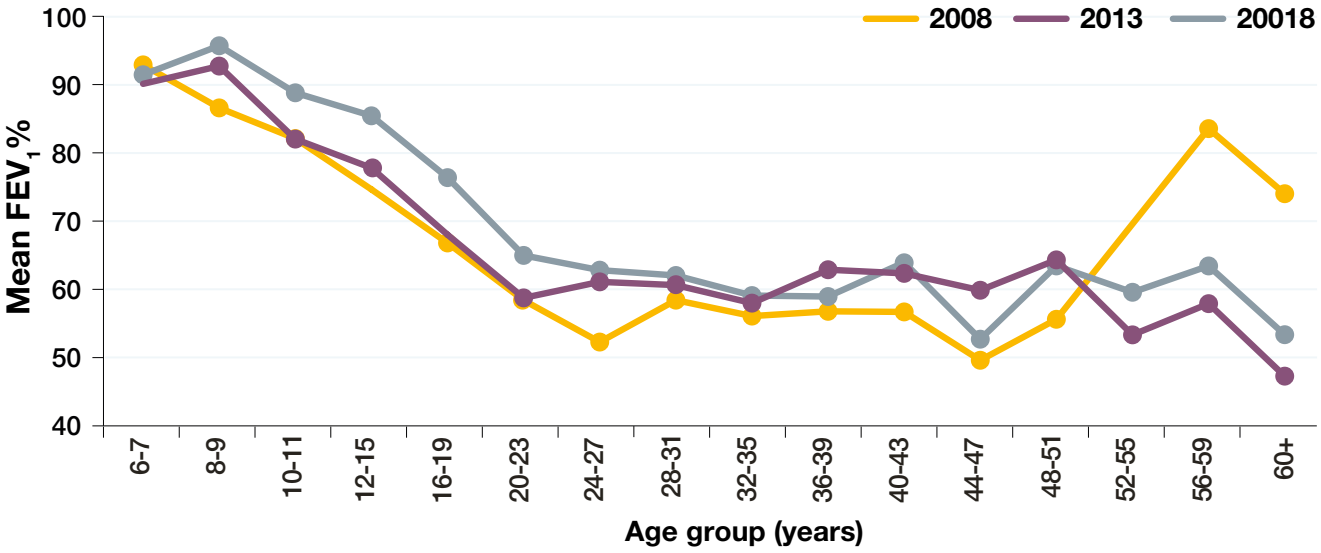


	Overall			Female			Male		
Age (yrs)	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**
6-7	47	96.7	88.5-104.6	23	95.7	88.8-105.5	24	96.8	83.6-103.9
8-9	39	100.4	86.5-109.0	14	96.4	84.2-104.1	25	101.7	91.8-109.0
10-11	31	95.1	83.3-101.2	15	87.8	80.6-99.9	16	97.3	93.1-101.4
12-15	92	91.1	84.3-98.2	48	91.3	83.4-99.7	44	91.1	84.6-95.8
16-19	53	86.3	69.8-93.9	27	88.8	73.9-96.4	26	80.2	69.5-93.0
20-23	90	72.0	50.2-91.1	43	66.9	40.1-91.1	47	76.9	54.6-93.9
24-27	78	67.9	48.6-84.4	35	71.2	56.7-87.4	43	65.0	42.7-84.4
28-31	62	64.6	50.3-79.5	30	69.9	57.4-88.3	32	58.8	43.9-75.4
32-35	58	65.3	49.0-80.1	25	65.8	52.5-77.4	33	64.8	46.5-81.2
36-39	35	58.5	42.4-88.9	13	70.2	48.0-88.9	22	55.1	40.4-73.7
40-43	28	67.9	49.0-86.2	10	52.3	38.5-58.7	18	75.6	55.5-94.1
44-47	14	53.6	32.5-70.8	8	53.5	31.2-69.8	6	53.6	39.2-95.7
48-51	25	69.4	53.3-77.3	10	70.2	53.3-75.9	15	65.8	44.1-82.3
52-55	20	65.6	49.3-78.7	10	58.7	41.1-70.1	10	76.0	54.8-93.0
56-59	7	67.4	47.2-81.7	<5	47.2	42.8-67.4	<5	80.6	64.4-88.3
60+	12	55.9	41.9-76.6	5	47.5	29.2-49.7	7	76.4	53.1-93.5
<16	209	93.5	85.1-101.6	100	92.5	84.2-100.8	109	94.7	85.5-102.0
≥16	482	68.9	49.7-87.1	219	68.1	51.2-87.1	263	69.5	48.6-87.2
<18	230	93.2	84.5-101.4	114	91.9	83.5-100.7	116	94.2	85.1-101.8
≥18	461	67.8	49.3-86.4	205	67.4	51.1-84.1	256	68.9	48.5-87.1
Overall	**691***	**80.1**	**56.0-94.5**	**319**	**79.7**	**57.0-93.7**	**372**	**80.4**	**55.7-94.6**

* number with non-missing data

1.13 FEV_1% predicted (GLI equations) over time in patients 6 years and older who have not had a lung transplant
N=698 in 2018, N=661 in 2013, N=292 in 2008*

As we learn more about CF and how to treat it, we hope to improve the outcomes of people with the condition. The chart below shows how FEV_1% in 2018 compares to Registry data from 2008 and 2013. 2008 is shown as a comparator year as this is the earliest year that we can be confident that the coverage of the Registry gives an accurate reflection of the CF population.

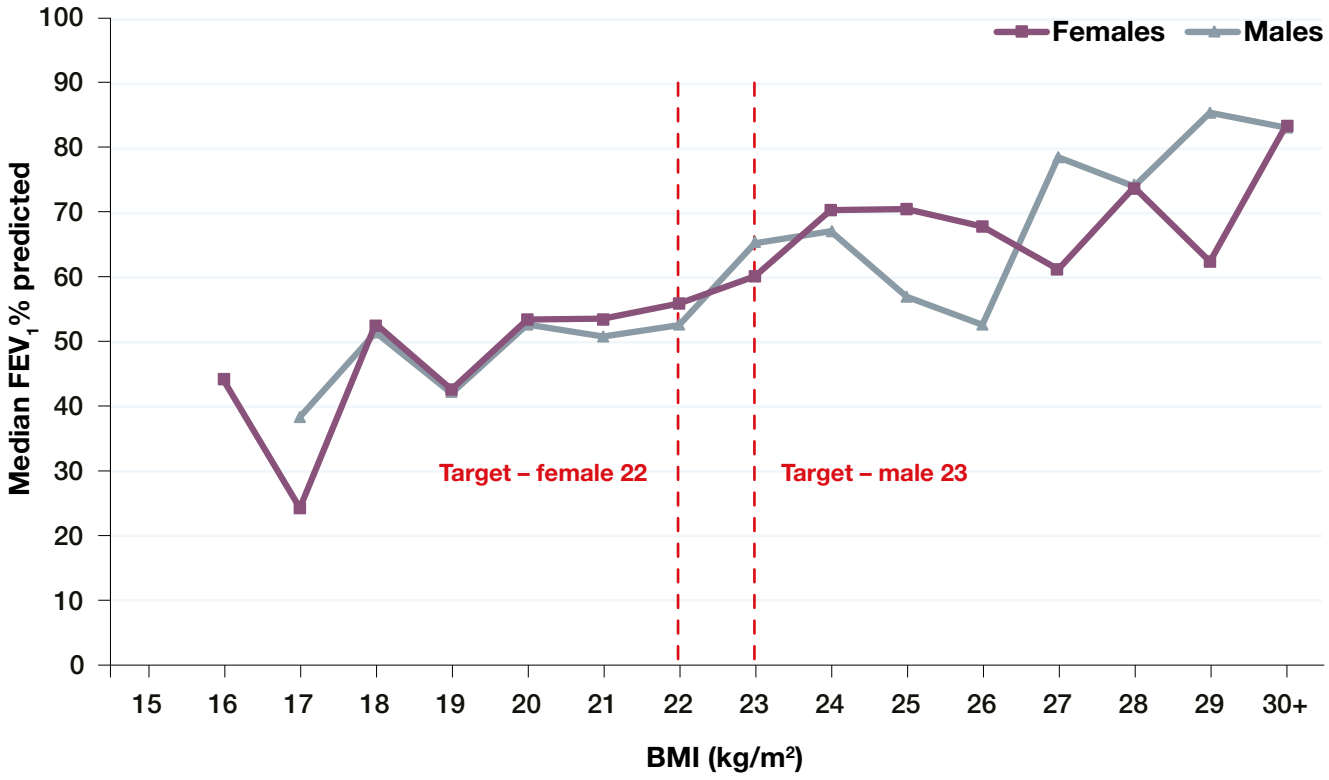


Age (years)	2008 mean	2013 mean	2018 mean	p-values (t-test) **
6-7	92.9	90.1	91.4	0.630
8-9	86.6	92.7	95.7	0.302
10-11	82.0	82.0	88.8	0.073
12-15	74.6	77.7	85.4	0.014
16-19	66.7	67.9	76.3	0.034
20-23	58.3	58.6	64.9	0.096
24-27	52.2	61.0	62.7	0.668
28-31	58.3	60.5	61.9	0.741
32-35	56.0	57.9	59.0	0.808
36-39	56.7	62.8	58.8	0.548
40-43	56.6	62.2	63.9	0.852
44-47	49.5	59.8	52.6	0.392
48-51	55.5	64.2	63.4	0.899
52-55	-	53.2	59.5	0.509
56-59	83.5	57.8	63.3	0.685
60+	73.9	47.2	53.2	-
<16	81.2	85.4	89.2	-
≥16	59.7	61.6	63.2	-
<18	80.0	82.4	88.3	-
≥18	55.6	60.5	62.5	-

* Due to missing data, means are calculated from a population of 682 in 2018, 623 in 2013 and 246 in 2008.
** t-test comparing 2018 with 2013 If the p-value is less than 0.05 then the difference in the mean is statistically significant. p-values excluded in groups where patients appear in the same group in both 2013 and 2013.

1.14 FEV_1% predicted (GLI equations) and BMI in people aged 20 years and over who have not had a transplant
N=430*

The goal BMI for adults is 22 for women, and 23 for men. The chart below shows the relationship between BMI and FEV_1% predicted. A healthy BMI can help to protect people with CF against lung infection and help to preserve lung health. This chart excludes people who have had a lung transplant.

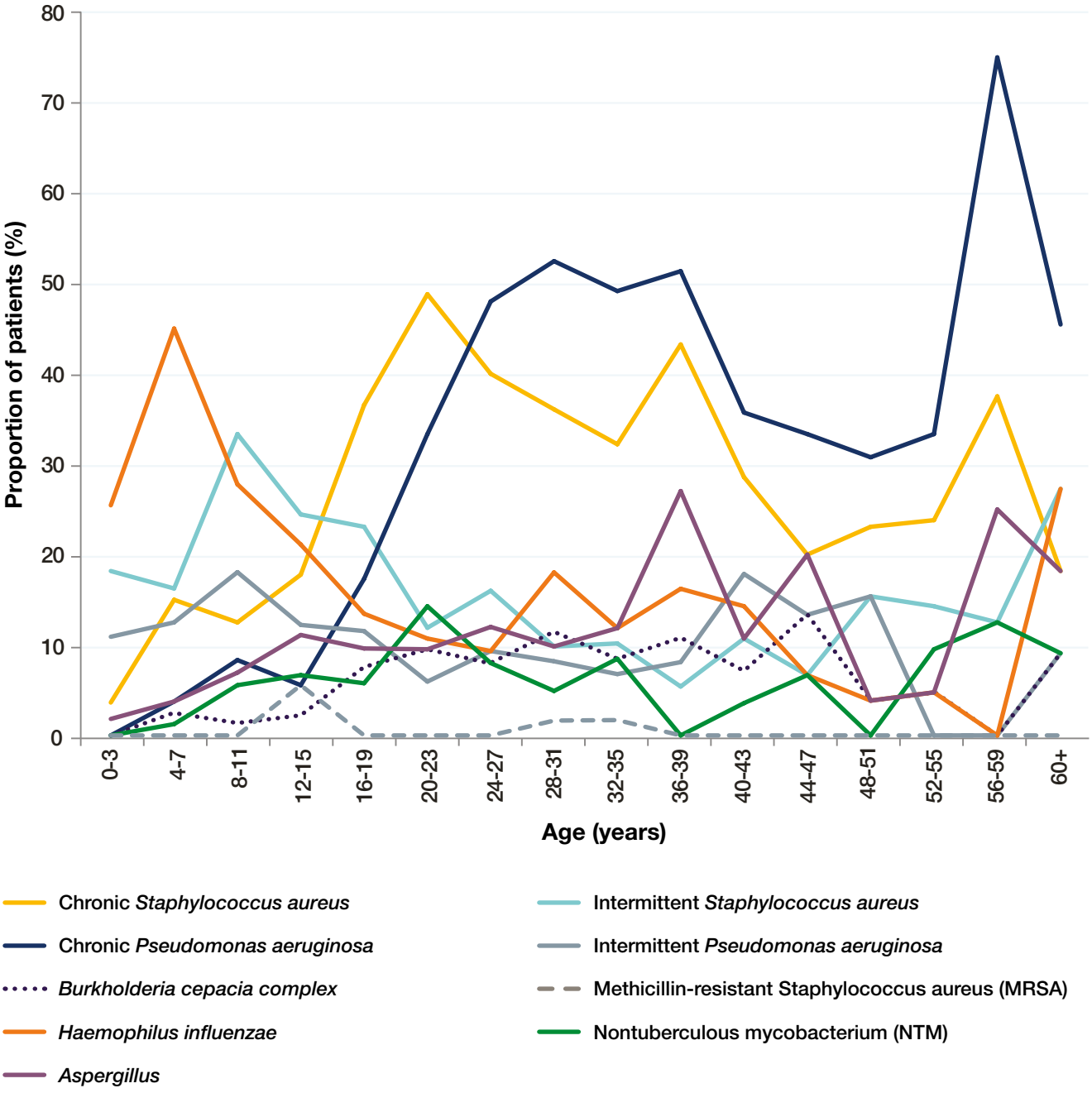


* Due to missing data, medians are calculated from a population of 426.
Each point represents the median FEV1 % predicted of patients for each given BMI value. Due to the wide range of BMIs in this population with a value of 30 of more, these are grouped into one.

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. All other infections are reported if they have occurred at least once as a positive growth in the 12 months prior to the patient’s annual review data set.

1.15 Lung infections in 2018
N=819*



*Proportions are calculated from 774 (94.5%) patients who had a culture taken in 2018.

1.16 Lung infections in 2018
<16 years N=304; ≥16 years N=515

Infections in this table reflect bugs grown in the 12 months prior to the 2018 annual review. The UK CF Registry definition of 'chronic' is three or more isolates in the last 12 months.

	Paediatric age range (Years)				Overall
	0-3	4-7	8-11	12-15	Paediatric (<16 years)
Number in age range; n	55	82	75	92	304
Number who had culture taken; n*	55	80	72	90	297
Chronic S. aureus; n (%)	<5	12 (15.0)	9 (12.5)	16 (17.8)	-
Intermittent S. aureus; n (%)	10 (18.2)	13 (16.3)	24 (33.3)	22 (24.4)	69 (23.2)
Chronic P. aeruginosa; n (%)	<5	<5	6 (8.3)	5 (5.6)	14 (4.7)
Intermittent P. aeruginosa; n (%)	6 (10.9)	10 (12.5)	13 (18.1)	11 (12.2)	40 (13.5)
B. cepacia complex; n (%)	0 (0.0)	<5	<5	<5	5 (1.7)
B. cenocepacia; n (%)	0 (0.0)	<5	<5	<5	<5
B. multivorans; n (%)	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)
B. cepacia (other); n (%)	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)
MRSA; n (%)	0 (0.0)	0 (0.0)	0 (0.0)	5 (5.6)	5 (1.7)
H. influenza; n (%)	14 (25.5)	36 (45.0)	20 (27.8)	19 (21.1)	89 (30.0)
NTM; n (%)	0 (0.0)	<5	<5	6 (6.7)	11 (3.7)
Aspergillus; n (%)	<5	<5	5 (6.9)	10 (11.1)	19 (6.4)

*Proportions are calculated from the number of people with CF who were recorded as having had a culture taken.

	Adult age range (Years)						Overall
	16-19	20-23	24-27	28-31	32-35	36-39	Adults (≥16 years)
Number in age range; n	53	91	82	65	64	41	515
Number who had culture taken; n*	52	84	75	61	59	37	477
Chronic S. aureus; n (%)	19 (36.5)	41 (48.8)	30 (40.0)	22 (36.1)	19 (32.2)	16 (43.2)	174 (36.5)
Intermittent S. aureus; n (%)	12 (23.1)	10 (11.9)	12 (16.0)	6 (9.8)	6 (10.2)	<5	63 (13.2)
Chronic P. aeruginosa; n (%)	9 (17.3)	28 (33.3)	36 (48.0)	32 (52.5)	29 (49.2)	19 (51.4)	194 (40.7)
Intermittent P. aeruginosa; n (%)	6 (11.5)	5 (6.0)	7 (9.3)	5 (8.2)	<5	<5	42 (8.8)
B. cepacia complex; n (%)	<5	8 (9.5)	6 (8.0)	7 (11.5)	5 (8.5)	<5	41 (8.6)
B. cenocepacia; n (%)	0 (0.0)	<5	<5	<5	<5	<5	14 (2.9)
B. multivorans; n (%)	<5	<5	<5	5 (8.2)	<5	<5	22 (4.6)
B. cepacia (other); n (%)	<5	<5	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)	<5
MRSA; n (%)	0 (0.0)	0 (0.0)	0 (0.0)	<5	<5	0 (0.0)	<5
H. influenza; n (%)	7 (13.5)	9 (10.7)	7 (9.3)	11 (18.0)	7 (11.9)	6 (16.2)	57 (11.9)
NTM; n (%)	<5	12 (14.3)	6 (8.0)	<5	5 (8.5)	0 (0.0)	35 (7.3)
Aspergillus; n (%)	5 (9.6)	8 (9.5)	9 (12.0)	6 (9.8)	7 (11.9)	10 (27.0)	57 (11.9)

	Adult age range (Years)						Overall
	40-43	44-47	48-51	52-55	56-59	60+	Adults (≥16 years)
Number in age range; n	31	18	26	23	8	13	515
Number who had culture taken; n*	28	15	26	21	8	11	477
Chronic S. aureus; n (%)	8 (28.6)	<5	6 (23.1)	5 (23.8)	<5	<5	174 (36.5)
Intermittent S. aureus; n (%)	<5	<5	<5	<5	<5	<5	63 (13.2)
Chronic P. aeruginosa; n (%)	10 (35.7)	5 (33.3)	8 (30.8)	7 (33.3)	6 (75.0)	5 (45.5)	194 (40.7)
Intermittent P. aeruginosa n (%)	5 (17.9)	<5	<5	0 (0.0)	0 (0.0)	<5	42 (8.8)
B. cepacia complex; n (%)	<5	<5	<5	<5	0 (0.0)	<5	41 (8.6)
B. cenocepacia; n (%)	<5	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)	14 (2.9)
B. multivorans; n (%)	<5	<5	<5	<5	0 (0.0)	0 (0.0)	22 (4.6)
B. cepacia (other); n (%)	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)	<5	<5
MRSA; n (%)	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)
H. influenza; n (%)	<5	<5	<5	<5	0 (0.0)	<5	57 (11.9)
NTM; n (%)	<5	<5	0 (0.0)	<5	<5	<5	35 (7.3)
Aspergillus; n (%)	<5	<5	<5	<5	<5	<5	57 (11.9)

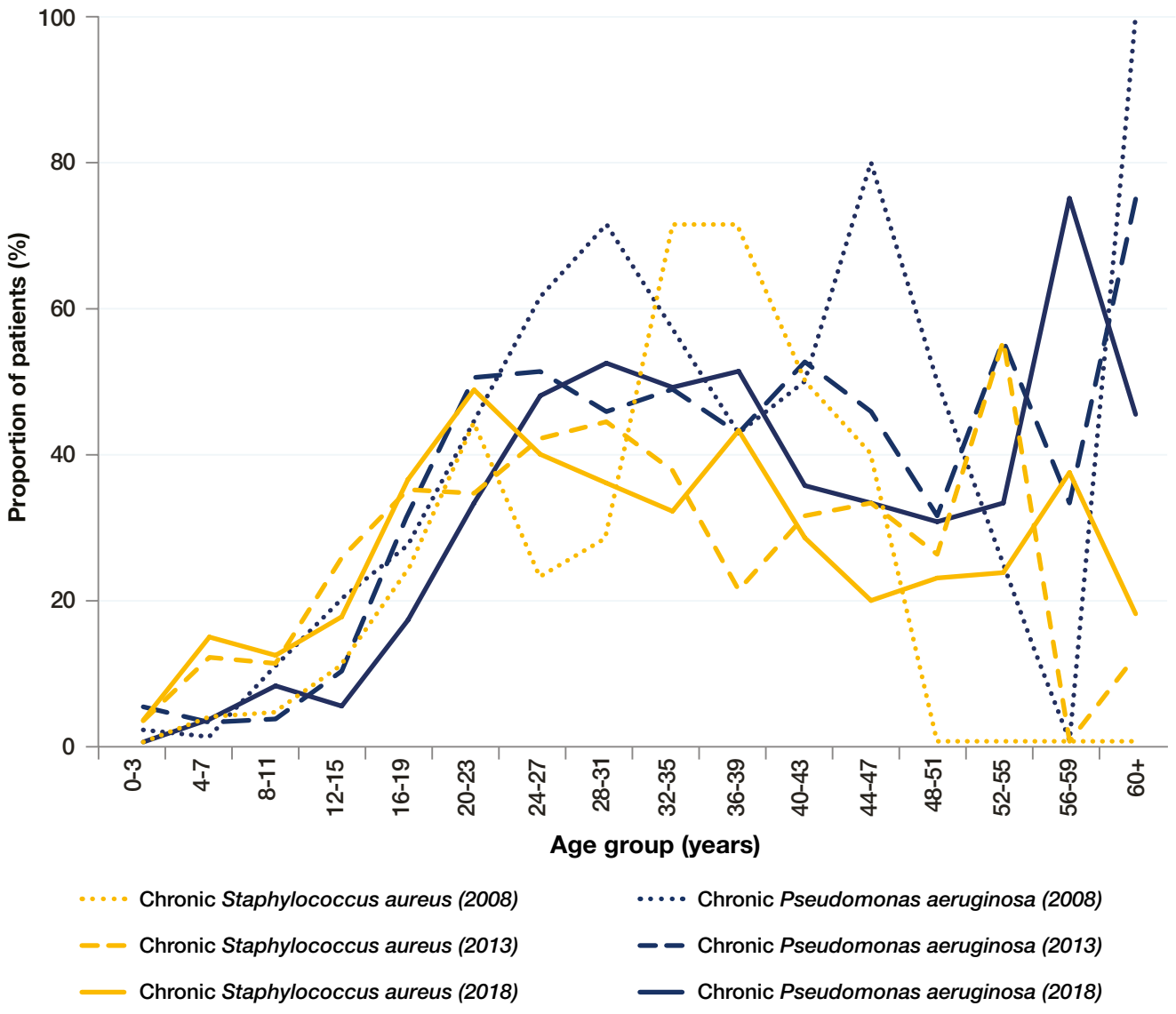
*Proportions are calculated from the number of people with CF who were recorded as having had a culture taken.

1.17 Nontuberculous mycobacteria (NTM) or atypical mycobacteria

NTM is slow to grow and takes time to treat. It may be present for several years before eradication, or may never be cleared. In the table below, 'prevalence' represents all people reported in that year as having a positive culture. 'Incidence' represents all positive cultures in individuals that have not reported having any in the previous two years of data.

	2016 (n=829)	2017 (n=858)	2018 (n=819)
NTM prevalence; n (%)	44 (5.3%)	38 (4.4%)	46 (5.6%)
On NTM treatment in the given year; n (% of NTM prevalence in given year)	16 (36%)	11 (29%)	16 (2.0%)
NTM incidence; n	20	12	19
M. abscessus prevalence; n	25	26	28
M. abscessus incidence; n	7	10	9

1.18 Lung infections over time
N=429 in 2008, N=809 in 2013, N=858 in 2018



The median age of people with chronic Pseudomonas aeruginosa infection increased from 17 years in 2008 to 29 years in 2018.

Chronic Staphylococcus aureus				
Age (years)	**2008 (%)**	**2013 (%)**	**2018 (%)**	**p-value***
0-3	1.3	1.7	3.6	0.890
4-7	5.6	5.7	15.0	0.603
8-11	7.5	9.1	12.5	0.719
12-15	13.0	13.3	17.8	0.278
16-19	19.5	19.8	36.5	0.861
20-23	21.2	28.3	48.8	0.141
24-27	20.0	27.3	40.0	0.478
28-31	21.1	25.3	36.1	0.230
32-35	23.5	20.5	32.2	0.423
36-39	22.8	18.7	43.2	0.087
40-43	14.6	19.0	28.6	0.743
44-47	20.5	22.5	20.0	0.224
48-51	26.4	21.5	23.1	0.803
52-55	10.7	26.4	23.8	0.064
56-59	6.3	19.6	37.5	- **
60+	15.0	19.2	18.2	-
<16 years	7.3	7.6	13.1	-
≥16 years	20.4	23.6	36.5	-
<18 years	8.6	8.9	14.2	-
≥18 years	20.7	24.1	36.8	-

Chronic Pseudomonas aeruginosa				
Age (years)	**2008 (%)**	**2013 (%)**	**2018 (%)**	**p-value***
0-3	2.4	2.3	0.0	0.101
4-7	6.7	3.7	3.8	0.886
8-11	14.2	7.2	8.3	0.201
12-15	24.6	17.9	5.6	0.302
16-19	43.9	33.6	17.3	0.060
20-23	61.2	48.9	33.3	0.006
24-27	64.0	57.3	48.0	0.351
28-31	68.2	60.7	52.5	0.636
32-35	64.5	61.3	49.2	0.794
36-39	60.2	58.1	51.4	0.595
40-43	62.8	53.5	35.7	0.205
44-47	62.5	52.0	33.3	0.233
48-51	43.4	50.6	30.8	0.954
52-55	57.1	51.7	33.3	0.187
56-59	31.3	39.1	75.0	- **
60+	55.0	38.5	45.5	-
<16 years	12.8	7.9	4.7	-
≥16 years	58.4	51.0	40.7	-
<18 years	15.2	10.6	5.3	-
≥18 years	61.3	53.4	41.9	-

* The proportion of people with each infection within each age group was compared between 2013 and 2018. If the p-value is less than 0.05 then the difference in the proportions is statistically significant.
**Sample size too low (<30 total) for hypothesis test

Complications

1.19 Complications in 2018

The number shown is for a complication that has been present in the preceding 12 months.

	2017			2018		
	Overall (n=858)	**<16 years (n=320)**	**≥16 years (N=538)**	**Overall (n=819)**	**<16 years (n=304)**	**≥16 years (N=515)**
Respiratory related						
Nasal polyps requiring surgery	-	<5	16 (3.0)	-	<5	11 (2.1)
Sinus disease	-	<5	122 (22.7)	-	<5	124 (24.1)
Asthma	63 (7.3)	9 (2.8)	54 (10.0)	-	<5	40 (7.8)
Allergic bronchopulmonary aspergillosis (ABPA)	37 (4.3)	7 (2.2)	30 (5.6)	29 (3.5)	5 (1.6)	24 (4.7)
Any haemoptysis	-	<5	25 (4.6)	-	<5	12 (2.3)
Massive haemoptysis	<5	0 (0.0)	<5	<5	0 (0.0)	<5
Pneumothorax requiring chest tube	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)
Pancreas & hepatobiliary disease						
Raised liver enzymes	41 (4.8)	14 (4.4)	27 (5.0)	44 (5.4)	19 (6.3)	25 (4.9)
Liver disease	68 (7.9)	18 (5.6)	50 (9.3)	121 (14.8)	32 (10.5)	89 (17.3)
Cirrhosis with no portal hypertension	-	<5	13 (2.4)	-	<5	10 (1.9)
Cirrhosis with portal hypertension	-	<5	13 (2.4)	14 (1.7)	5 (1.6)	9 (1.7)
Gall bladder disease requiring surgery	-	<5	<5	-	6 (2.0)	<5
Pancreatitis	-	<5	10 (1.9)	-	<5	<5
Upper gastrointestinal (GI)						
Gastro-oesophageal reflux disease (GERD)	-	<5	113 (21.0)	-	<5	136 (26.4)
Peptic ulcer	0 (0.0)	0	0	0 (0.0)	0	0
GI bleed (varices as source)	0 (0.0)	0	0	<5	<5	0
GI bleed (non varices as source)	<5	<5	<5	0 (0.0)	0	0
Lower gastrointestinal						
Intestinal obstruction	<5	<5	0	<5	<5	<5
Distal intestinal obstruction syndrome (DIOS)	98 (11.4)	<5	94 (17.5)	81 (9.9)	5 (1.6)	76 (14.8)
Fibrosing colonopathy/colonic stricture	0 (0.0)	0	0	<5	0	<5
Rectal prolapse	<5	<5	<5	0 (0.0)	0	0
Renal						
Kidney stones	<5	0	<5	<5	0	<5
Renal failure	13 (1.5)	0	13 (2.4)	-	<5	13 (2.5)
Musculoskeletal						
Arthritis	5 (0.6)	0	5 (0.9)	-	<5	5 (1.0)
Arthropathy	32 (3.7)	0	32 (5.9)	-	<5	32 (6.2)
Bone fracture	<5	0	<5	<5	0	<5
Osteopenia	107 (12.5)	0	107 (19.9)	86 (10.5)	0	86 (16.7)
Osteoporosis	40 (4.7)	0	40 (7.4)	49 (6.0)	0	49 (9.5)
Other						
Cancer confirmed by histology	<5	<5	<5	0 (0.0)	0	0
Port inserted or replaced	18 (2.1)	6 (1.9)	12 (2.2)	17 (2.1)	6 (2.0)	11 (2.1)
Depression	19 (2.2)	0	19 (3.5)	13 (1.6)	0	13 (2.5)
Hearing loss	-	<5	7 (1.3)	-	<5	9 (1.7)
Hypertension	17 (2.0)	0	17 (3.2)	19 (2.3)	0	19 (3.7)

1.20 Incidence of complications

The table below describes new cases of a complication that have not been reported for an individual in at least the previous two years.

	2017			2018		
	Overall (n=858)	**<16 years (n=309)**	**≥16 years (n=520)**	**Overall (n=819)**	**<16 years (n=304)**	**≥16 years (n=515)**
Allergic bronchopulmonary aspergillosis; n (%)	-	<5	8 (1.5)	6 (0.7)	<5	<5
Cirrhosis - no portal hypertension; n (%)	-	<5	11 (2.1)	<5	0	<5
Cirrhosis - with portal hypertension; n (%)	-	<5	5 (0.9)	<5	<5	<5
Cancer confirmed by histology; n (%)	<5	<5	<5	0	0	0

1.21 Cystic fibrosis-related diabetes (CFRD) N=639

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, or it may not work properly, causing CFRD. CFRD is different from type 1 and type 2 diabetes, but has features of both.

	All ≥10 years (n=639)	**10-15 years (n=124)**	**≥16 years (n=515)**
On CFRD treatment; n (%)	149 (23.3)	6 (4.8)	143 (27.8)
CFRD screening; n (%)			
Yes	339 (53.1)	100 (80.6)	239 (46.4)
Known CFRD	152 (23.8)	7 (5.6)	145 (28.2)
No	139 (21.8)	17 (13.7)	122 (23.7)
Unknown	9 (1.4)	0 (0.0)	9 (1.7)

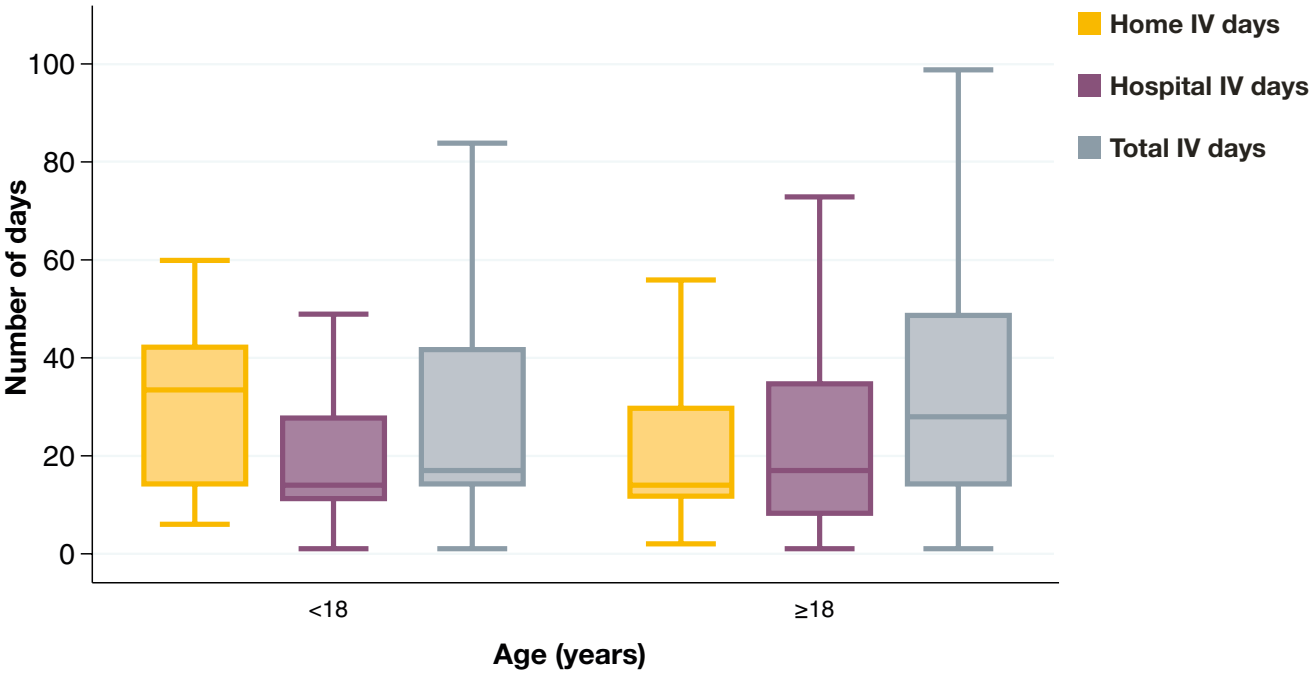
Antibiotics

1.22 Intravenous (IV) antibiotics
N=819

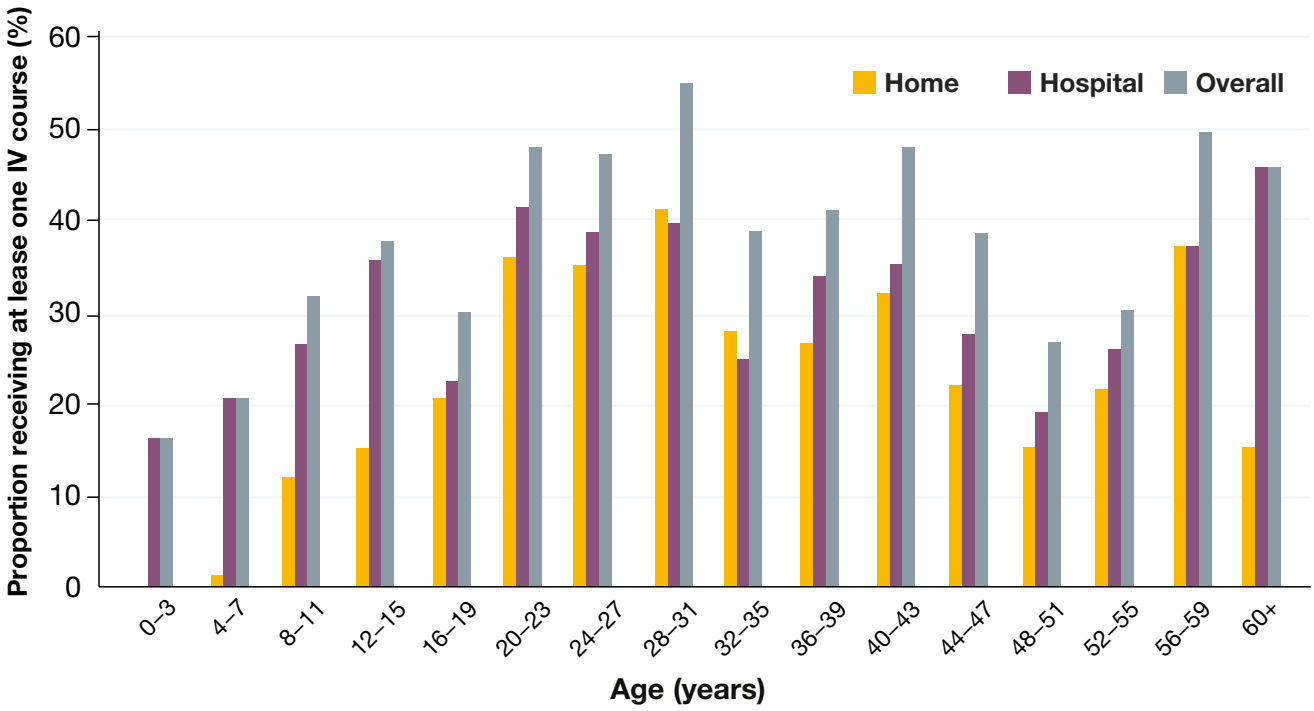
When someone with CF becomes unwell with an infection, they might be prescribed IV antibiotics. IV antibiotics are given to the patient through their veins. This treatment can take a number of days and might take place as a hospital inpatient, or at home.

		Home		Hospital		Total	
Age	**N**	**Patients n (%)**	**Median days (IQR)**	**Patients n (%)**	**Median days (IQR)**	**Patients n (%)**	**Median days (IQR)**
0-3	55	0 (0.0)	-	9 (16.4)	14 (14-14)	9 (16.4)	14 (14-14)
4-7	82	1 (1.2)	47 (47-47)	17 (20.7)	15 (13-24)	17 (20.7)	16 (13-28)
8-11	75	9 (12.0)	33 (16-42)	20 (26.7)	20 (14-41)	24 (32.0)	27 (14-49)
12-15	92	14 (15.2)	38 (14-46)	33 (35.9)	14 (6-28)	35 (38.0)	27 (14-56)
16-19	53	11 (20.8)	19 (14-50)	12 (22.6)	21 (8-36)	16 (30.2)	28 (14-42)
20-23	91	33 (36.3)	15 (12-30)	38 (41.8)	24 (12-38)	44 (48.4)	38 (26-58)
24-27	82	29 (35.4)	14 (9-28)	32 (39.0)	16 (9-41)	39 (47.6)	28 (14-58)
28-31	65	26 (40.0)	14 (11-21)	27 (41.5)	17 (6-28)	36 (55.4)	27 (14-41)
32-35	64	18 (28.1)	28 (14-48)	16 (25.0)	16 (11-23)	25 (39.1)	28 (16-42)
36-39	41	11 (26.8)	14 (11-76)	14 (34.1)	41 (14-56)	17 (41.5)	42 (23-82)
40-43	31	10 (32.3)	14 (12-49)	11 (35.5)	14 (4-14)	15 (48.4)	14 (14-49)
44-47	18	<5	29 (17-43)	5 (27.8)	14 (12-24)	-	30 (14-56)
48-51	26	<5	23 (8-41)	5 (19.2)	9 (7-14)	-	14 (7-42)
52-55	23	5 (21.7)	14 (7-23)	6 (26.1)	17 (14-35)	7 (30.4)	42 (14-46)
56-59	8	<5	12 (7-28)	<5	7 (2-14)	<5	14 (14-21)
60+	13	<5	15 (13-16)	6 (46.2)	21 (10-38)	6 (46.2)	27 (14-38)
<16	304	24 (7.9)	38 (15-45)	79 (26.0)	14 (13-28)	85 (28.0)	16 (14-43)
≥16	515	156 (30.3)	14 (12-30)	175 (34.0)	17 (8-33)	223 (43.3)	28 (14-49)
<18	325	28 (8.6)	34 (14-43)	83 (25.5)	14 (11-28)	90 (27.7)	17 (14-42)
≥18	494	152 (30.8)	14 (12-30)	171 (34.6)	17 (8-35)	218 (44.1)	28 (14-49)
Overall	**819**	**180 (22.0)**	**17 (12-38)**	**254 (31.0)**	**16 (9-32)**	**308 (37.6)**	**28 (14-47)**

This box plot graph illustrates the spread of the number of days on IV antibiotics in the Scottish CF population, stratified by age. A guide on how to correctly interpret this box plot graph can be found on page 41.



The bar graph below summarises the proportion of people receiving at least one course of IV antibiotics across different age groups within the Scottish CF population. Overall, the proportion of patients receiving at least one IV course at home was 22.0% and in hospital was 31.0%. The proportion receiving any IVs was 37.6%.



1.23 Inhaled antibiotic use among people with chronic *Pseudomonas aeruginosa*

	2008			2013			2018		
	Overall	**<16 years**	**≥16 years**	**Overall**	**<16 years**	**≥16 years**	**Overall**	**<16 years**	**≥16 years**
Patients with chronic *P. aeruginosa*; n	67	28	39	241	17	224	207	14	193
Tobramycin solution; n (%)	6 (9.0)	<5	<5	-	<5	44 (19.6)	-	<5	29 (15.0)
Other aminoglycoside; n (%)	0	0	0	0	0	0	<5	0	<5
Colistin; n (%)	21 (31.3)	15 (53.6)	6 (15.4)	92 (38.2)	13 (76.5)	79 (35.3)	34 (16.4)	6 (42.9)	28 (14.5)
Promixin; n (%)	<5	0	<5	-	<5	40 (17.9)	-	<5	26 (13.5)
Aztreonam; n (%)	-	-	-	<5	0	<5	20 (9.7)	0	20 (10.4)
Colistimethate dry powder inhaler; n (%)	-	-	-	-	-	-	-	<5	45 (23.3)
Tobramycin inhalation powder; n (%)	-	-	-	-	-	-	52 (25.1)	0	52 (26.9)
At least one of the above; n (%)	28 (41.8)	17 (60.7)	11 (28.2)	151 (62.7)	15 (88.2)	136 (60.7)	161 (77.8)	12 (85.7)	149 (77.2)

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

1.24 Long-term azithromycin use

Azithromycin is an antibiotic with some anti-inflammatory properties. It is recommended for long-term use as a prophylactic antibiotic in people with chronic *Pseudomonas aeruginosa* infection.

		Patients with chronic *P. aeruginosa*; n (%)	**Patients without chronic *P. aeruginosa*; n (%)**
2008	**Overall (n=429)**	**29 (46.0)**	**-**
	0-3 years (n=91)	0 (0)	<5
	4-15 years (n=236)	12 (35.3)	22 (64.7)
	≥16 years n=(102)	17 (60.7)	11 (39.3)
2013	**Overall (n=809)**	**-**	**-**
	0-3 years (n=70)	<5	<5
	4-15 years (n=244)	8 (17.0)	39 (83.0)
	≥16 years (n=495)	176 (54.7)	146 (45.3)
2018	**Overall (n=819)**	**171 (41.2)**	**-**
	0-3 years (n=55)	0 (0)	<5
	4-15 years (n=249)	10 (14.5)	59 (85.5)
	≥16 years (n=515)	161 (46.9)	182 (53.1)

1.25 Flucloxacillin

Flucloxacillin is an antibiotic that is used prophylactically to prevent infection with bacteria.

Age	Total patients	Patients on Flucloxacilin; n (%)
0-3	55	35 (63.6)
4-7	82	27 (32.9)
8-11	75	21 (28.0)
12-15	92	31 (33.7)
16-19	53	11 (20.8)
20-23	91	22 (24.2)
24-27	82	16 (19.5)
28-31	65	8 (12.3)
32-35	64	5 (7.8)
36-39	41	<5
40-43	31	0
44-47	18	0
48-51	26	<5
52-55	23	<5
56-59	8	0
60+	13	0
<16 years	304	114 (37.5)
≥16 years	515	69 (13.4)
<18 years	325	116 (35.7)
≥18 years	494	67 (13.6)
Overall	**819**	**183 (22.3)**

Muco-active therapies

1.26 Mannitol

Age	Total patients	Patients on Mannitol; n (%)
0-3	55	0
4-7	82	0
8-11	75	0
12-15	92	0
16-19	53	0
20-23	91	0
24-27	82	0
28-31	65	<5
32-35	64	<5
36-39	41	<5
40-43	31	<5
44-47	18	0
48-51	26	<5
52-55	23	0
56-59	8	0
60+	13	0
<16 years	304	0
≥16 years	515	9 (1.7)
<18 years	325	0
≥18 years	494	9 (1.8)
Overall	**819**	**9 (1.1)**

1.27 DNase

	2008		2013		2018	
Age	Total patients	Patients on DNase; n (%)	Total patients	Patients on DNase; n (%)	Total patients	Patients on DNase; n (%)
0-3	91	<5	70	<5	55	<5
4-7	78	9 (11.5)	101	11 (10.9)	82	22 (26.8)
8-11	66	15 (22.7)	82	22 (26.8)	75	32 (42.7)
12-15	92	33 (35.9)	61	27 (44.3)	92	51 (55.4)
16-19	32	12 (37.5)	99	41 (41.4)	53	24 (45.3)
20-23	14	<5	97	42 (43.3)	91	57 (62.6)
24-27	14	<5	75	29 (38.7)	82	39 (47.6)
28-31	11	<5	67	25 (37.3)	65	42 (64.6)
32-35	8	<5	49	12 (24.5)	64	26 (40.6)
36-39	9	<5	26	7 (26.9)	41	13 (31.7)
40-43	5	<5	19	<5	31	14 (45.2)
44-47	5	<5	22	7 (31.8)	18	7 (38.9)
48-51	<5	0	20	6 (30.0)	26	12 (46.2)
52-55	<5	0	7	<5	23	8 (34.8)
56-59	<5	0	7	0	8	<5
60+	<5	0	7	<5	13	5 (38.5)
<16 years	327	60 (18.3)	314	63 (20.1)	304	106 (34.9)
≥16 years	102	23 (22.5)	495	179 (36.2)	515	249 (48.3)
<18 years	350	71 (20.3)	363	86 (23.7)	325	115 (35.4)
≥18 years	79	12 (15.2)	446	156 (35.0)	494	240 (48.6)
Overall	**429**	**83 (19.3)**	**809**	**242 (29.9)**	**819**	**355 (43.3)**

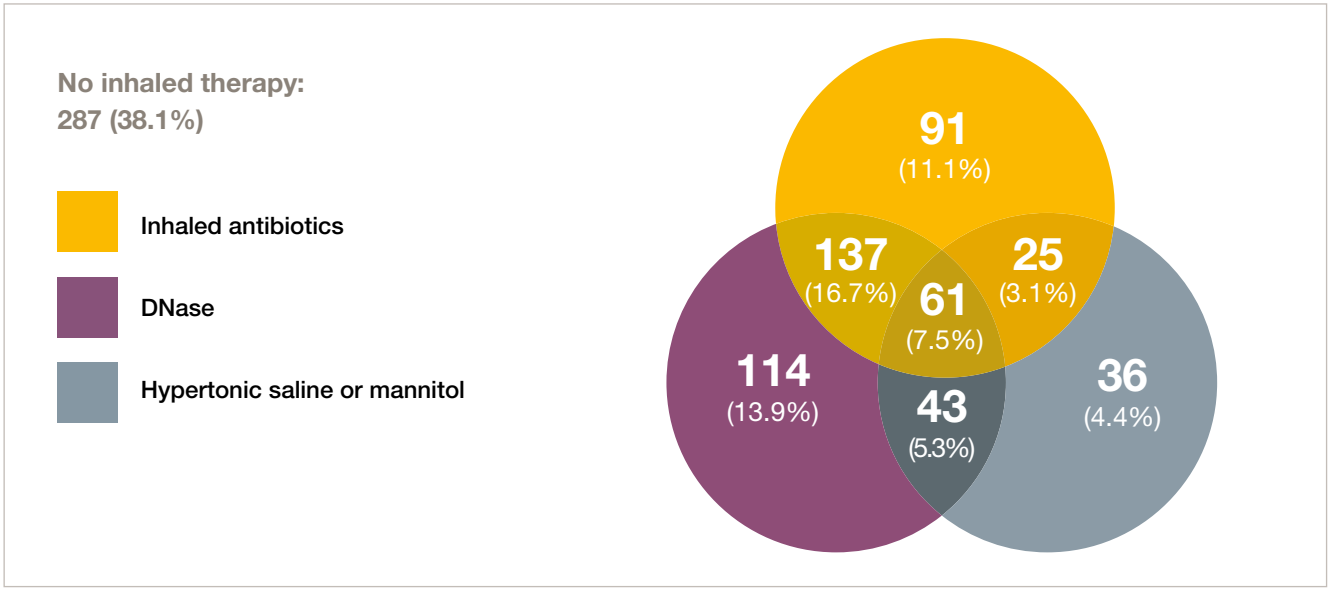
1.28 Hypertonic saline

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

	2008		2013		2018	
Age	**Total patients**	**Patients on hypertonic saline; n (%)**	**Total patients**	**Patients on hypertonic saline; n (%)**	**Total patients**	**Patients on hypertonic saline; n (%)**
0-3	91	0	70	<5	55	7 (12.7)
4-7	78	0	101	6 (5.9)	82	8 (9.8)
8-11	66	<5	82	10 (12.2)	75	17 (22.7)
12-15	92	<5	61	12 (19.7)	92	32 (34.8)
16-19	32	0	99	26 (26.3)	53	16 (30.2)
20-23	14	0	97	25 (25.8)	91	26 (28.6)
24-27	14	0	75	9 (12.0)	82	16 (19.5)
28-31	11	<5	67	7 (10.4)	65	10 (15.4)
32-35	8	0	49	5 (10.2)	64	13 (20.3)
36-39	9	0	26	0	41	5 (12.2)
40-43	5	0	19	<5	31	<5
44-47	5	0	22	<5	18	<5
48-51	<5	0	20	<5	26	<5
52-55	<5	<5	7	<5	23	<5
56-59	<5	0	7	0	8	<5
60+	<5	0	7	<5	13	0
<16 years	327	<5	314	29 (9.2)	304	64 (21.1)
≥16 years	102	<5	495	81 (16.4)	515	96 (18.6)
<18 years	350	<5	363	46 (12.7)	325	72 (22.2)
≥18 years	79	<5	446	64 (14.3)	494	88 (17.8)
Overall	**429**	**<5**	**809**	**110 (13.6)**	**819**	**160 (19.5)**

1.29 Burden of treatment

The Venn diagram shows how many people with CF are on one or more inhaled therapies and the combinations they take. A total of 287 (38.1%) people in Scotland are on no inhaled therapies.



Other Therapies

1.30 CFTR modifiers

Ivacaftor

Ivacaftor was first approved for use on the NHS in England in January 2013. Soon after, it was made available in Wales, Scotland and Northern Ireland. Since this time, ivacaftor's license has expanded across age ranges and mutation types. At the time of writing, ivacaftor is approved for use on the NHS across the UK for people aged two and older with a least one copy of nine specific CFTR mutations, known as 'gating' mutations. Ivacaftor is additionally approved for use on the NHS in Wales for people aged 18 and over with the R117H mutation.

	Age group (age at annual review)	N
Patients on Ivacaftor in Scotland	Overall	74
	<6 years	<5
	≥6 years	71
Patients stopped Ivacaftor ever	Overall	6
	<6 years	0
	≥6 years	6

People with CF tend to 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.

Tests	Age group (age at start date)	Median (IQR)	Number with complete data; n (%)
Sweat chloride before Ivacaftor	Overall	100 (95-106)	30 (40.5)
	<6 years	100 (97-102)	5 (71.4)
	≥6 years	99 (95-106)	25 (37.3)
Sweat chloride 6-8 weeks after Ivacaftor	Overall	52 (37-65)	20 (27.0)
	<6 years	54 (29-71)	<5
	≥6 years	52 (39-61)	16 (23.9)
FEV_1% before Ivacaftor	Overall	63.5 (42.1-79.8)	55 (74.30)
	<6 years	82.5 (81.6-83.5)	<5
	≥6 years	62.5 (42.1-77.2)	53 (79.1)
FEV_1% 6-8 weeks after Ivacaftor	Overall	68.8 (51.7-81.6)	51 (68.9)
	<6 years	88.1 (81.4-94.8)	<5
	≥6 years	63.9 (51.7-81.4)	49 (73.1)

Lumacaftor/ivacaftor

Lumacaftor/ivacaftor is licensed for use in patients aged 12 and over with two copies of the F508del mutation. In Autumn 2019 the Scottish Government and NHS England announced access for everyone with CF in Scotland and England who stands to benefit. Prior to this, lumacaftor/ivacaftor was only available rarely on compassionate grounds and through limited clinical trial access. In Scotland, 18 people received this drug during 2018.

	Age group (age at annual review)	N
Patients on Lumacaftor/Ivacaftor in Scotland	Overall	18
	<6 years	0
	≥6 years	18
Patients stopped Lumacaftor/Ivacaftor ever	Overall	<5
	<6 years	0
	≥6 years	<5

Tezacaftor/ivacaftor

Tezacaftor/ivacaftor was licensed in 2018 for patients aged 12 and over who have two copies of the F508del mutation, or a single copy of F508del and one of 14 specified 'residual function' mutations. In Autumn 2019 the Scottish Government and NHS England announced access for everyone with CF in Scotland and England who stands to benefit. Prior to this, tezacaftor/ivacaftor was only available rarely on compassionate grounds and through limited clinical trial access.

Fewer than five people with cystic fibrosis in Scotland are recorded as being prescribed tezacaftor/ivacaftor in 2018.

1.31 Oxygen and non-invasive ventilation

	Overall (n=819)	<16 years (n=304)	≥16 years (n=515)	<18 years (n=325)	≥18 years (n=494)
Non-invasive ventillation (NIV); n (%)	14 (1.7)	<5	11 (2.1)	<5	11 (2.2)
Long-term oxygen; n (%)	49 (6.0)	10 (3.3)	39 (7.6)	10 (3.1)	39 (7.9)
Among those who have long-term oxygen:					
Continuously	15 (30.6)	<5	13 (33.3)	<5	13 (33.3)
Nocturnal or with exertion	9 (18.4)	0 (0.0)	9 (23.1)	0 (0.0)	9 (23.1)
As required (PRN)	6 (12.2)	0 (0.0)	6 (15.4)	0 (0.0)	6 (15.4)
With exacerbation	19 (38.8)	8 (80.0)	11 (28.2)	8 (80.0)	11 (28.2)

1.32 Physiotherapy

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

	Overall (n=819)	<16 years (n=304)	≥16 years (n=515)	<18 years (n=325)	≥18 years (n=494)
Active cycle of breathing techniques; n (%)	95 (11.6)	10 (3.3)	85 (16.5)	13 (4.0)	82 (16.6)
Autogenic drainage (including assisted autogenic drainage); n (%)	362 (44.2)	70 (23.0)	292 (56.7)	75 (23.1)	287 (58.1)
Postural drainage; n (%)	<5	0 (0.0)	<5	0 (0.0)	<5
Any form of PEP; n (%)	434 (53.0)	273 (89.8)	161 (31.3)	288 (88.6)	146 (29.6)
VEST; n (%)	<5	0 (0.0)	<5	<5	<5
Exercise; n (%)	466 (56.9)	174 (57.2)	292 (56.7)	187 (57.5)	279 (56.5)
Other; n (%)	132 (16.1)	91 (29.9)	41 (8.0)	93 (28.6)	39 (7.9)

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

1.33 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.

	Overall (n=819)	<16 years (n=304)	≥16 years (n=515)	<18 years (n=494)	≥18 years (n=325)
Any supplemental feeding; n(%)	191 (23.3)	62 (20.4)	129 (25.0)	65 (20.0)	126 (25.5)
Nasogastric tube; n(%)	-	<5	8	<5	8
Gastrostomy tube/button; n(%)	33 (4.0)	15 (4.9)	18 (3.5)	15 (4.6)	18 (3.6)
Jejunal; n(%)	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)
Total parenteral nutrition (TPN); n(%)	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)

1.34 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 'bilateral sequential lung transplant'. The following table gives information about transplant activity over time.

	2013	2014	2015	2016	2017	2018
Number evaluated	21	17	19	18	22	19
Number accepted	11	12	16	8	17	7
Number receiving transplants	<5	6	<5	<5	<5	<5
Bilateral lung	<5	6	<5	<5	<5	<5
Liver	0	0	0	0	0	<5
Other	0	0	0	<5	<5	<5

Genotypes

Genotypes are part of the genetic makeup of an individual that usually control a particular characteristic, known as a phenotype. For people with CF, their genotype reveals which mutations of the CF gene causes 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.

Data completeness	n(%)
Patients genotyped with at least one variant recorded	813 (99.3)
Patients genotyped with both variants recorded	805 (98.3)
F508del mutations	
Homozygous f508del	347 (42.4)
Heterozygous f508del	391 (47.7)

1.35 Mutation combinations in Scotland

This tabulation shows the proportion (%) of patients with the most common mutation combinations. For example, 8.3% of the Scottish population have one copy of F508del and one copy of G551D in their genotype.

	Mutation 1							
Mutation 2	**F508del**	**R117H**	**G551D**	**G542X**	**621+1G->T**	**Other**	**Unknown**	**Total**
	(%)							
F508del	42.4							**89.1**
R117H	6.7	0.1						**0.7**
G551D	8.3	0.2	0.2					**2.3**
G542X	5.3	0.2	0.0	0.1				**0.9**
621+1G->T	1.0	0.0	0.1	0.0	0.0			**0.2**
Other	25.5	0.7	1.7	1.0	0.4	4.3		**6.0**
Unknown	1.0	0.0	0.0	0.0	0.0	0.0	0.7	**0.7**
Total	**90.1**	**1.3**	**2.1**	**1.1**	**0.4**	**4.3**	**0.7**	**100.0**

1.36 Mutations in the Scottish population

The table below shows the number of people with CF who carry at least one of each mutation. The groups are not mutually exclusive, as people with heterozygous mutations appear twice in the table.

These are the 20 most common mutations in the Scottish population. The full list of recorded mutations can be found in Appendix 3.

Nucleotide	Protein	Legacy name	N	%
c.1521_1523delCTT	p.Phe508del	F508del	738	90.1
c.1652G->A	p.Gly551Asp	G551D	87	10.6
c.350G->A	p.Arg117His	R117H	66	8.1
c.1624G->T	p.Gly542X	G542X	54	6.6
c.200C->T	p.Pro67Leu	P67L	39	4.8
c.1679G->C	p.Arg560Thr	R560T	16	2.0
c.1477C->T	p.Gln493X	Q493X	15	1.8
c.1585-1G->A	–	1717-1G->A	13	1.6
c.3909C->G	p.Asn1303Lys	N1303K	12	1.5
c.3454G->C	p.Asp1152His	D1152H	12	1.5
c.489+1G->T	–	621+1G->T	12	1.5
c.2657+5G->A	–	2789+5G->A	9	1.1
c.3717+12191C->T	–	3849+10kbC->T	8	1.0
c.3528delC	p.Lys1177SerfsX15	3659delC	8	1.0
c.178G->T	p.Glu60X	E60X	7	0.9
c.1558G->T	p.Val520Phe	V520F	7	0.9
c.948delT	p.Phe316LeufsX12	1078delT	6	0.7
c.1766+1G->A	–	1898+1G->A	5	0.6
c.1364C->A	p.Ala455Glu	A455E	5	0.6
c.1210-12[5](AJ574948.1:g.152T[5])	–	5T	5	0.6

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
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 review a centre's use of some medications and outcome data alongside national averages. This transparency is intended to help improve standards of care overall.

Lots of different factors can affect the outcomes of people with CF in centres, not all of which are within a 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 and 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 the data are shown in appendix 2 on page 50.

Key

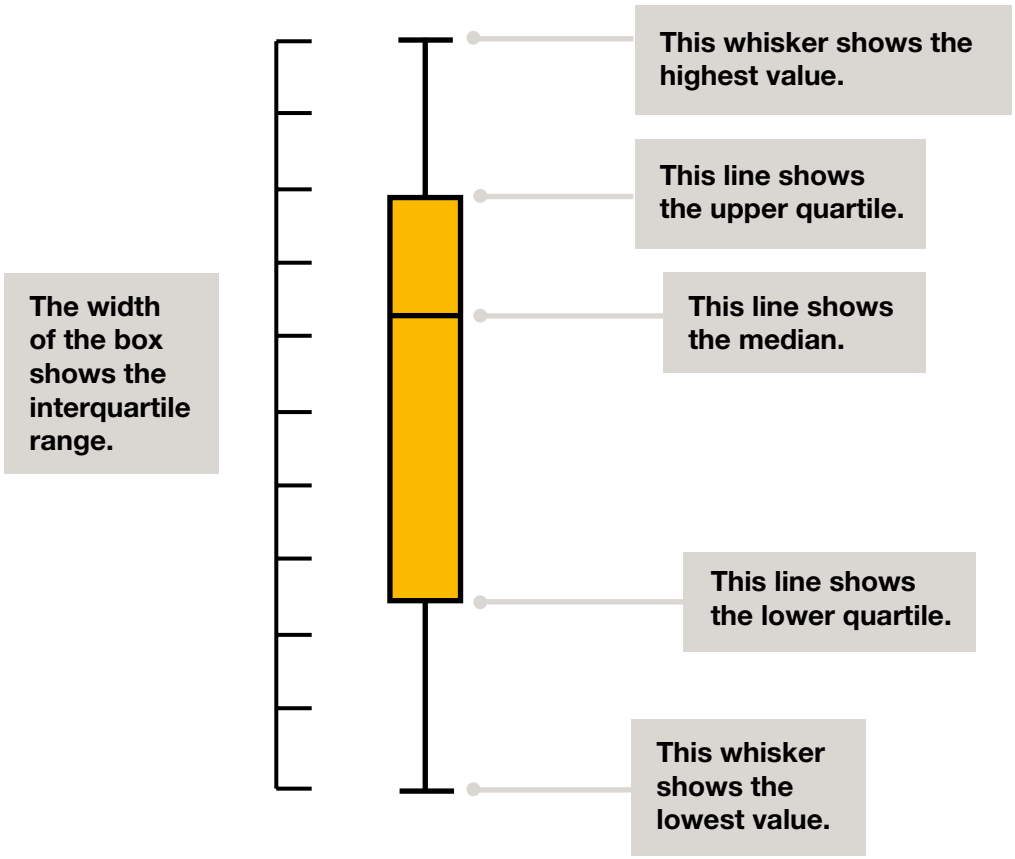
Paediatric centre

Adult centre

A guide to the charts

Some of the data in this section are shown as ‘box plots’.

Box plots

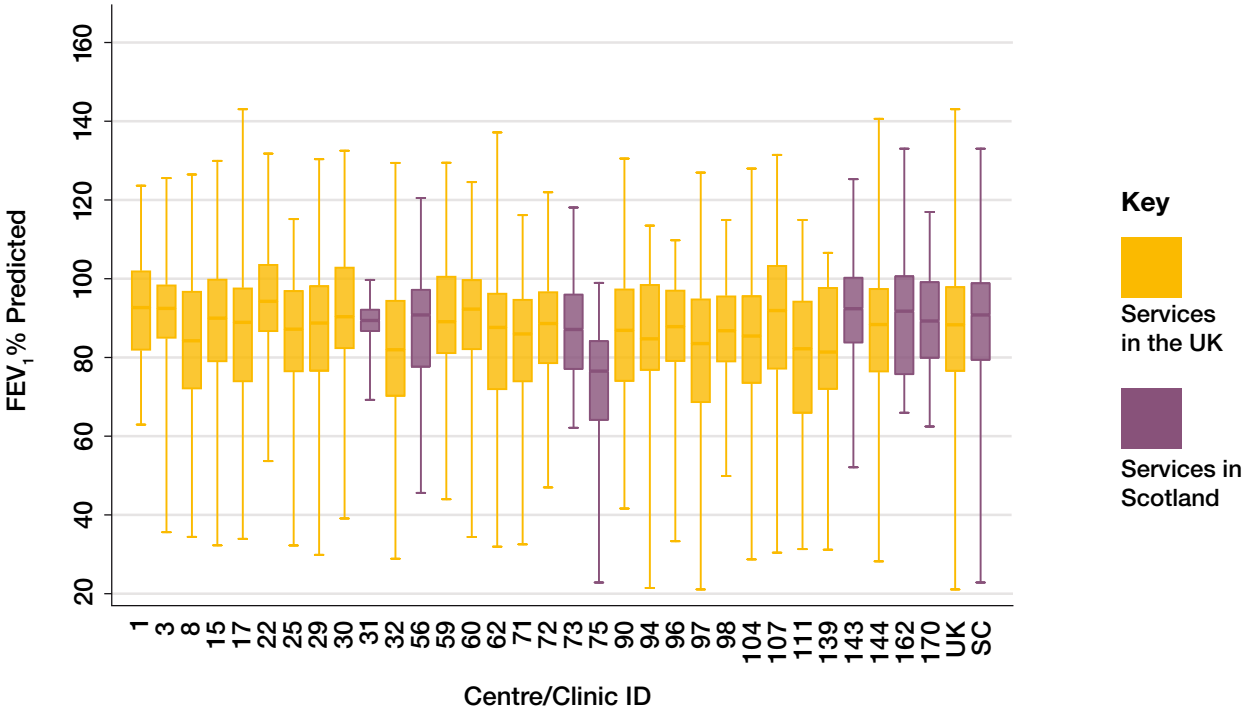


- The ‘box’ shows the middle half of the data for that centre, going from the first quartile to the third quartile. The longer the box, the more varied the data for that centre.
- The horizontal line within the box shows the median result for that centre.
- The ‘whiskers’ above and below the box show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

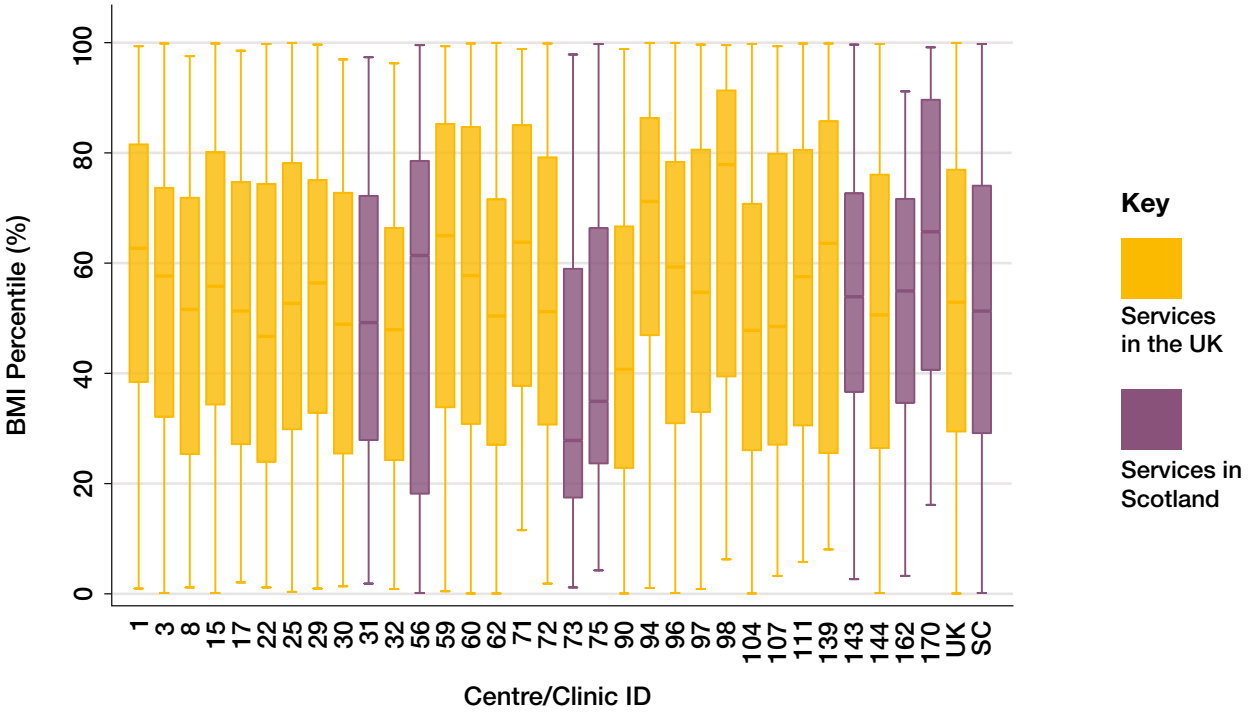
This section shows results for the five paediatric centres with their network clinics, and two stand-alone clinics.

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



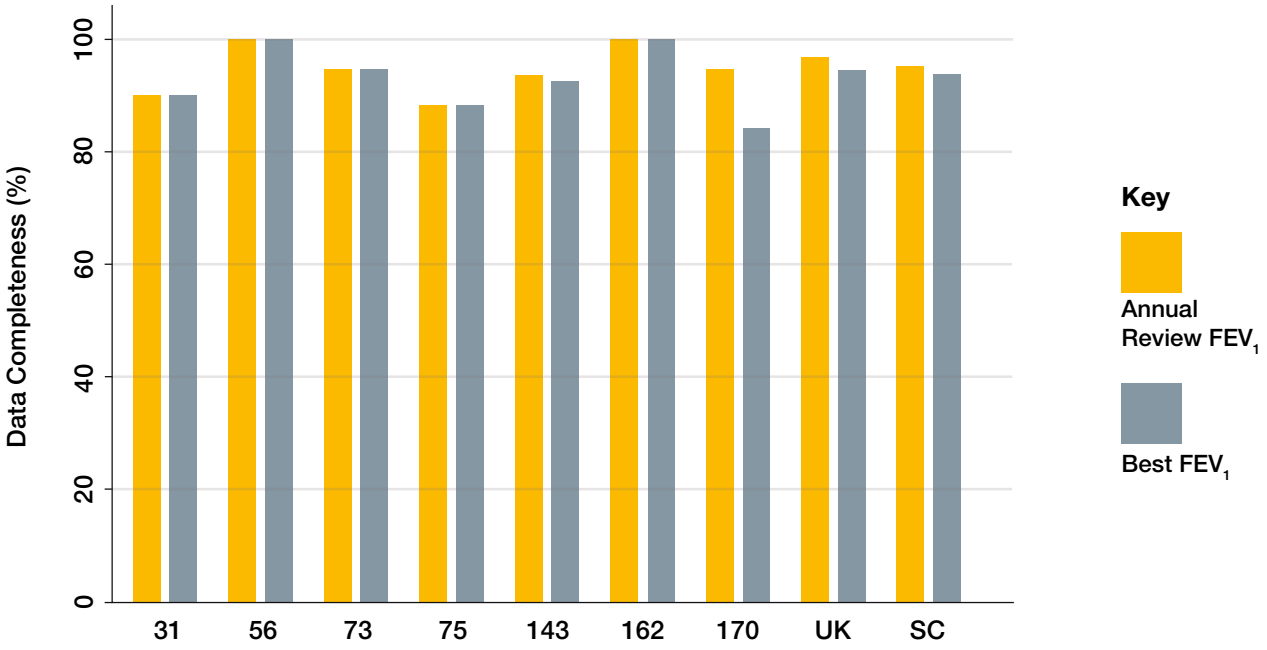
The median FEV_1% predicted of patients attending paediatric centres/clinics in Scotland is 90.9% predicted (IQR: 79.4 – 99.1).

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 51.3 (IQR: 29.0 74.2).

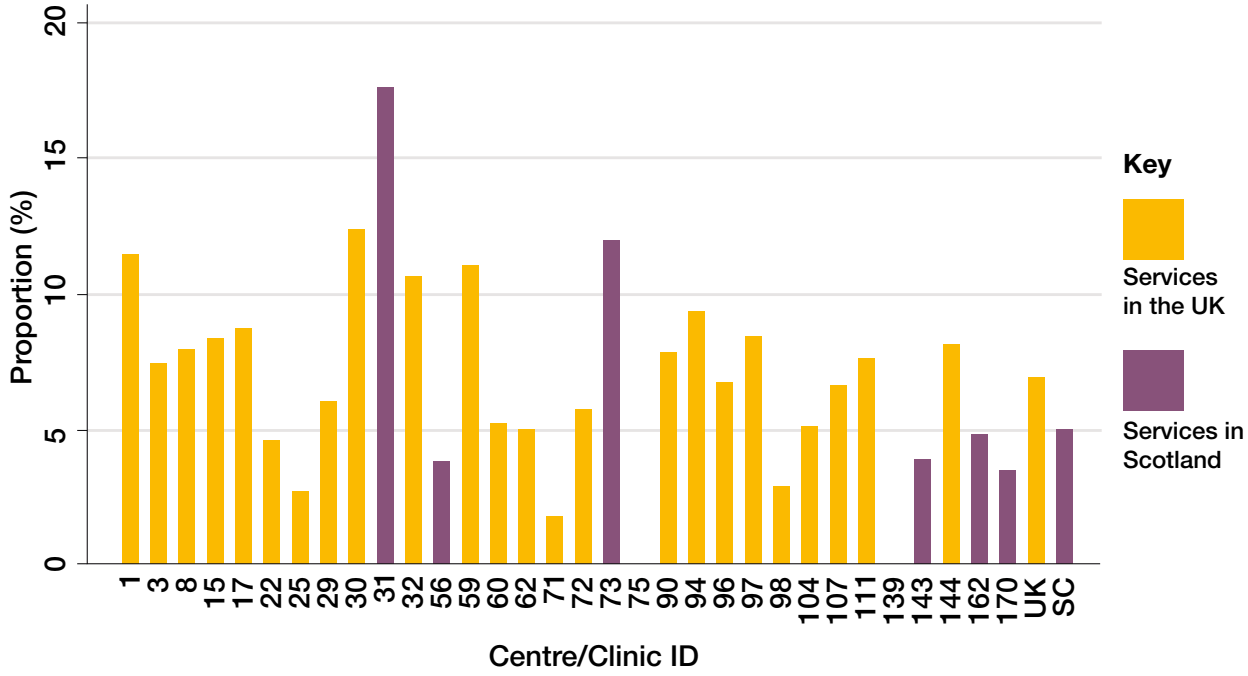
2.3 Data completeness by paediatric centre/clinic



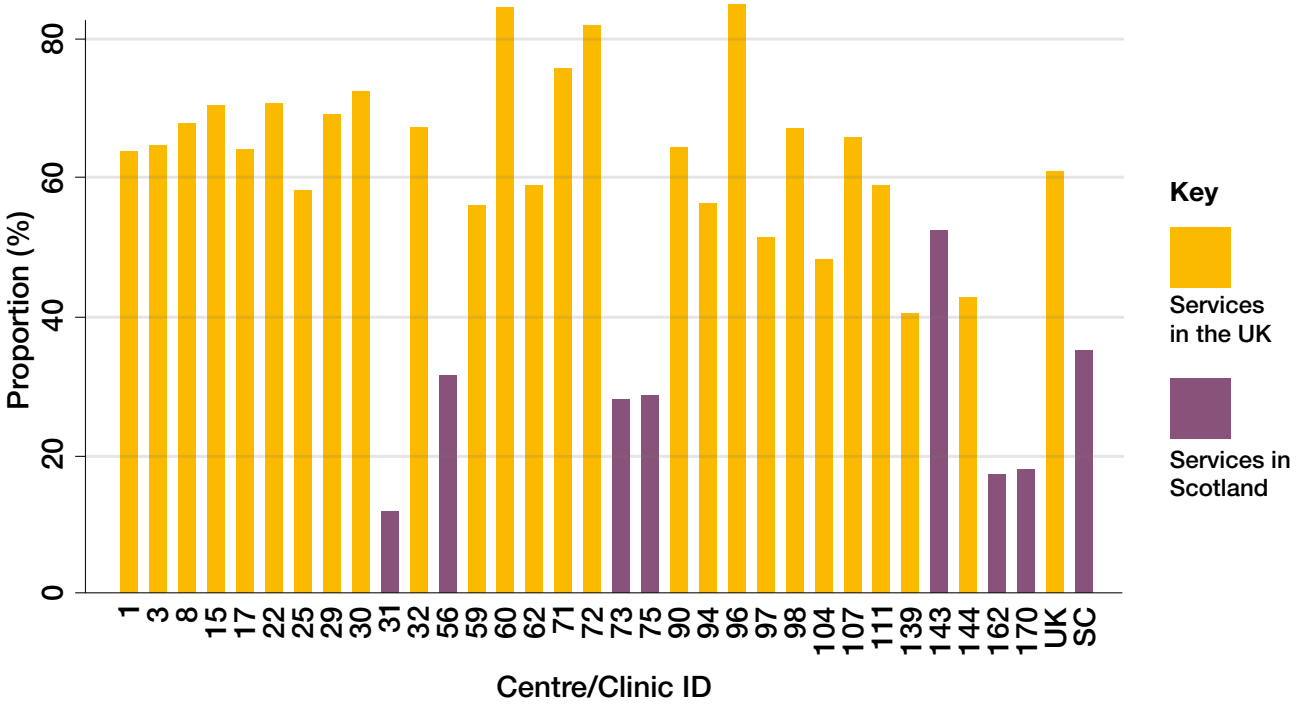
The mean data completeness for Annual Review FEV_1% predicted across Scottish paediatric centres is 95%. The mean data completeness for Best FEV_1% predicted across Scottish paediatric centres is 93.8%.

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

5.0% of paediatric patients attending clinics in Scotland during 2018 were infected with chronic *P. aeruginosa.*

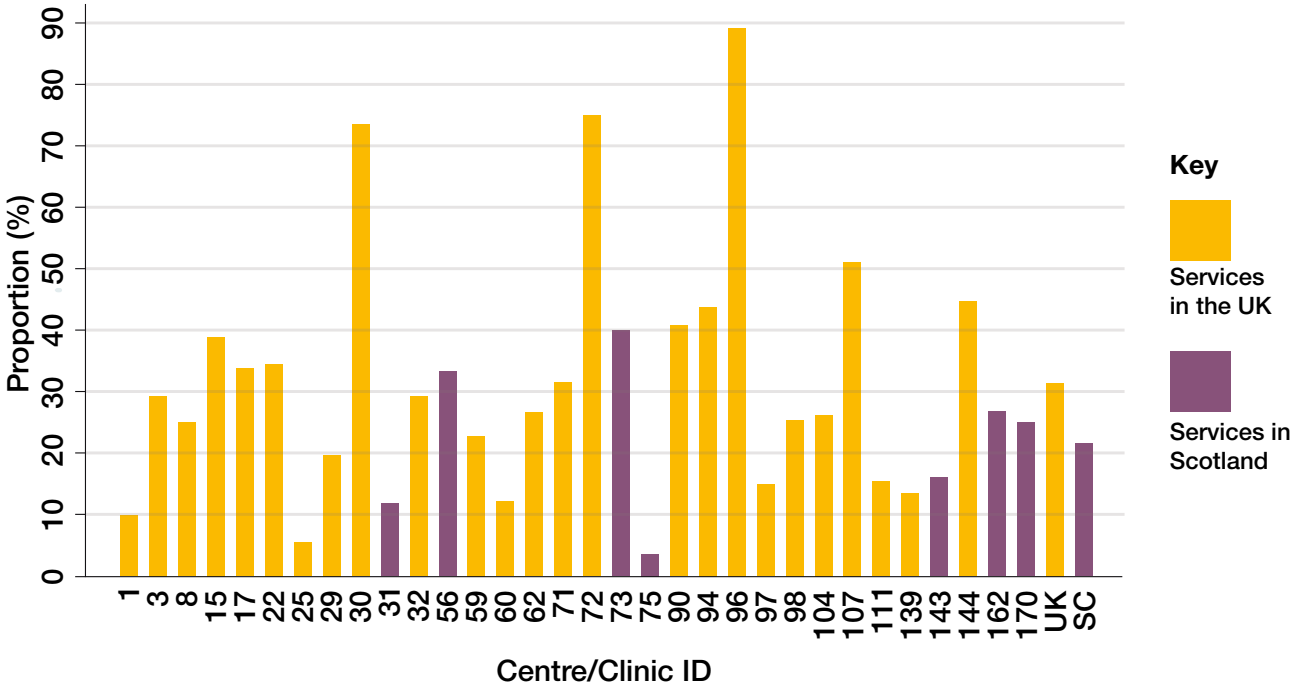


2.5 Proportion of patients receiving DNase treatment by paediatric centre/clinic



The proportion of patients attending paediatric centres/clinics in Scotland receiving DNase treatment is 35.0%.

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

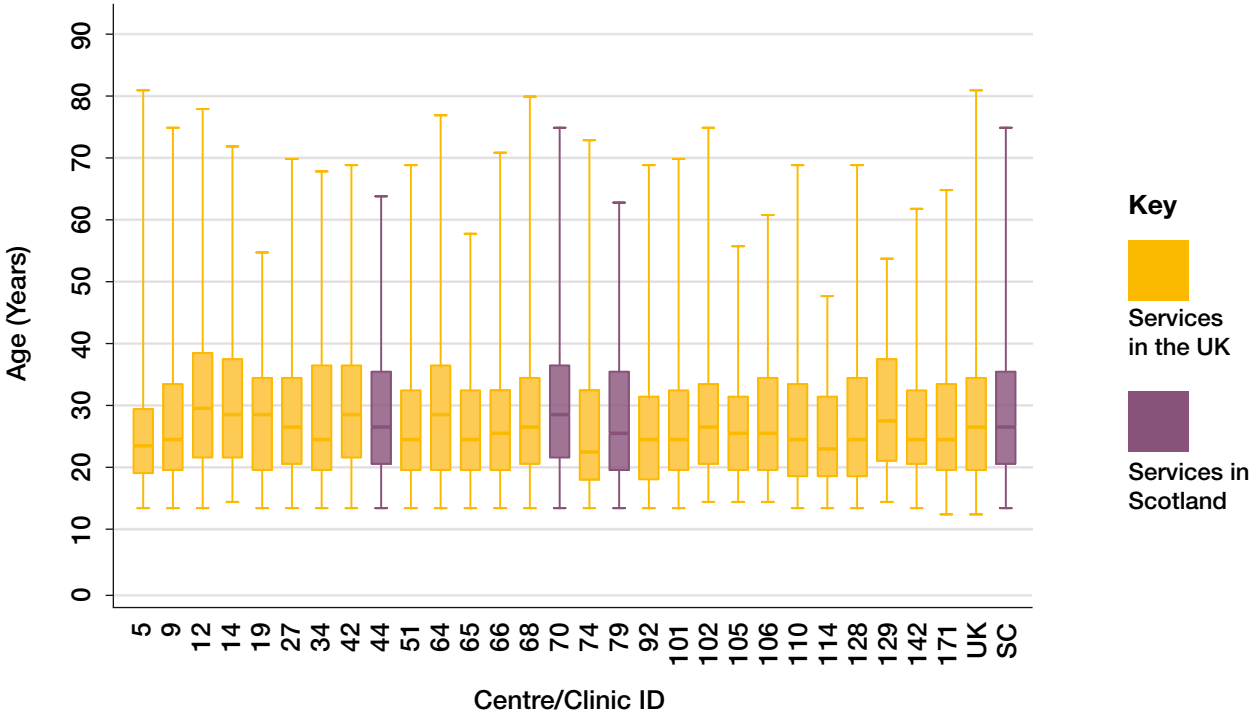


The proportion of patients attending paediatric centres/clinics in Scotland receiving hypertonic saline treatment is 21.4%.

Section 2b: Adult centre analysis

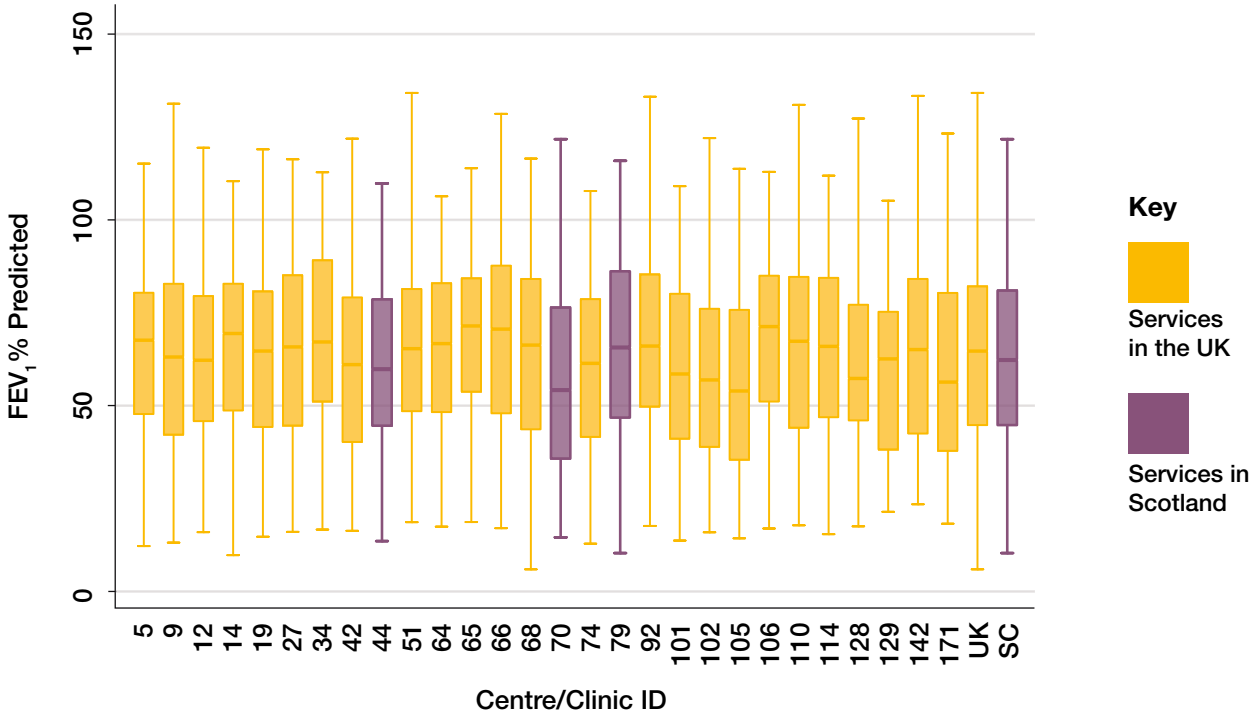
This section shows results for the three adult centres with their network clinics.

2.7 Median age (years) by adult service



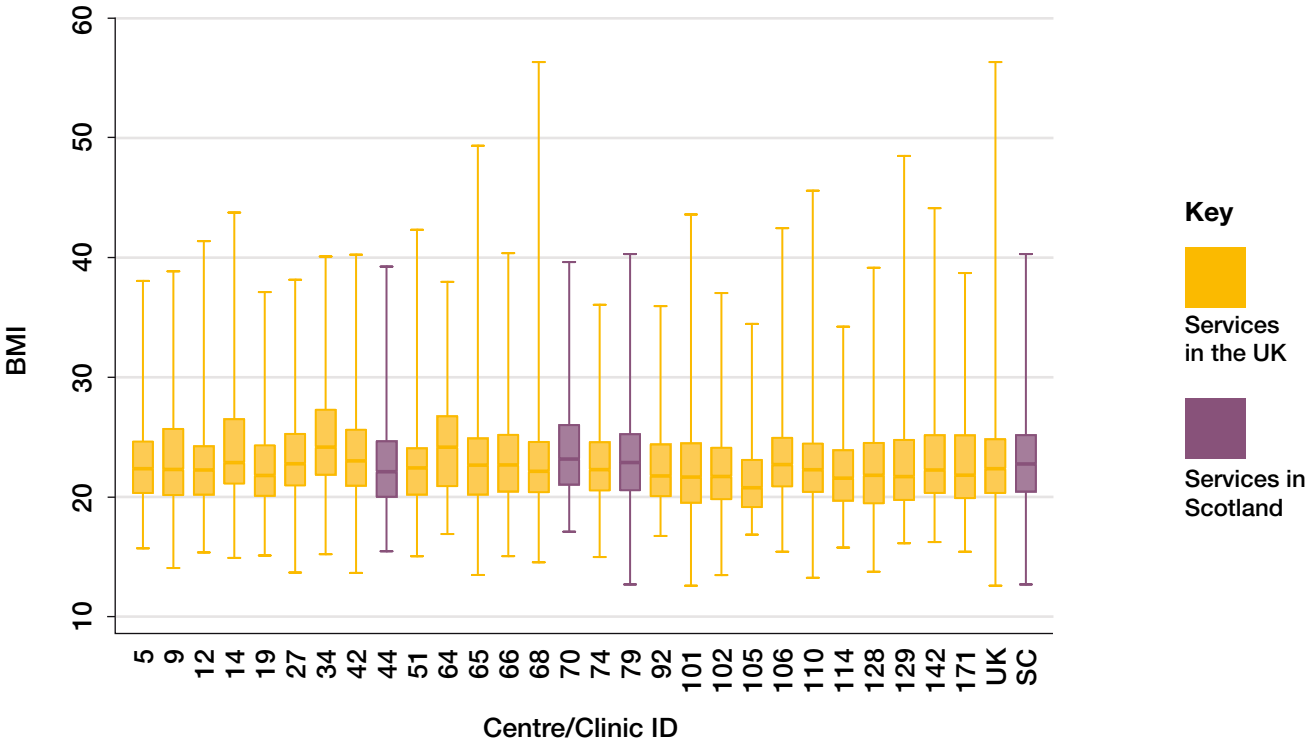
The median age of patients attending adult services in Scotland is 28 years (IQR: 22-37).

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



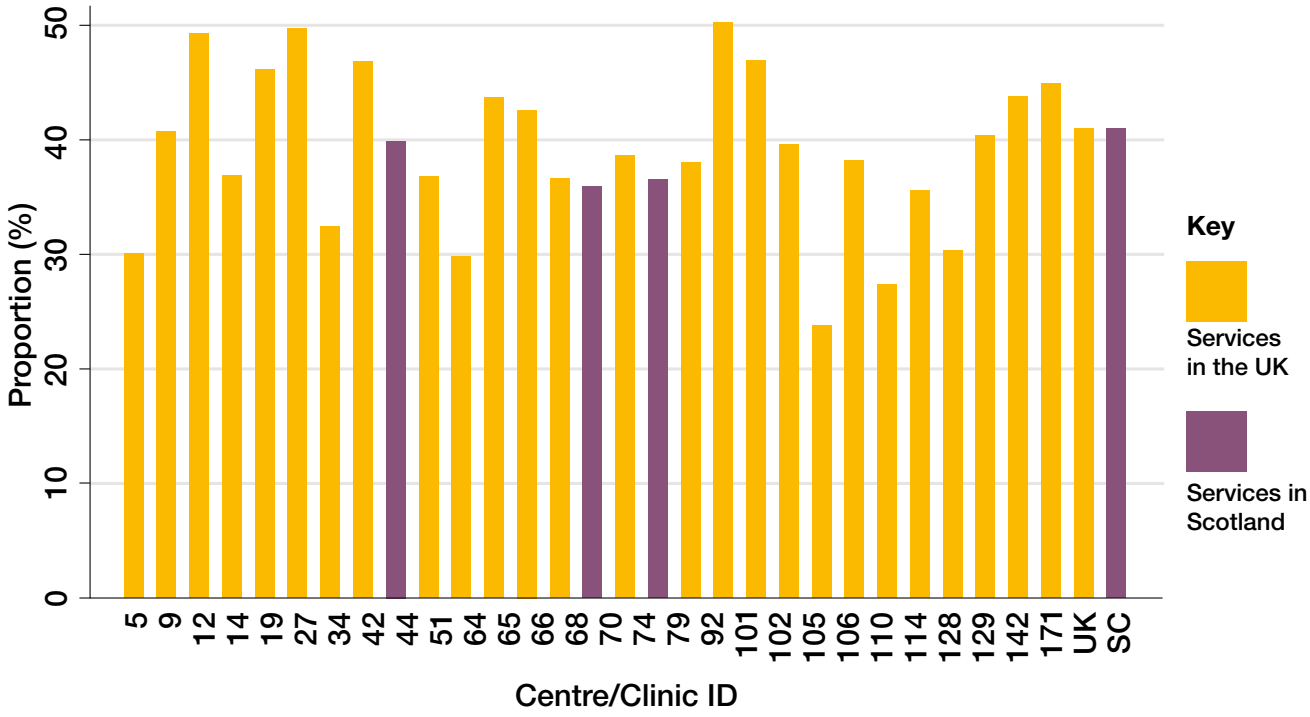
The median FEV1 % predicted of patients attending adult services in Scotland is 62.7% (IQR 45.0-82.6).

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



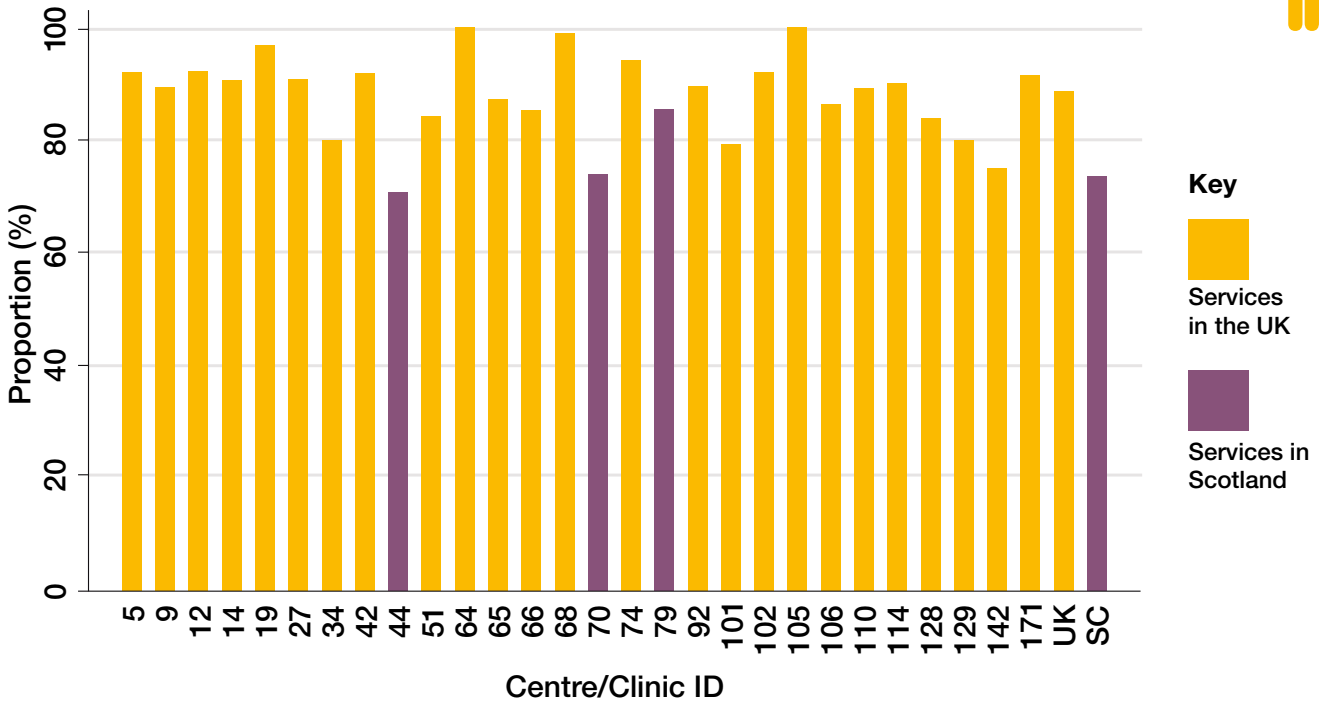
The median BMI of patients attending adult services in Scotland is 22.9 (IQR: 20.6 – 25.5).

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



The proportion of patients attending adult services in Scotland during 2018 with chronic *P. aeruginosa* is 41.0%.

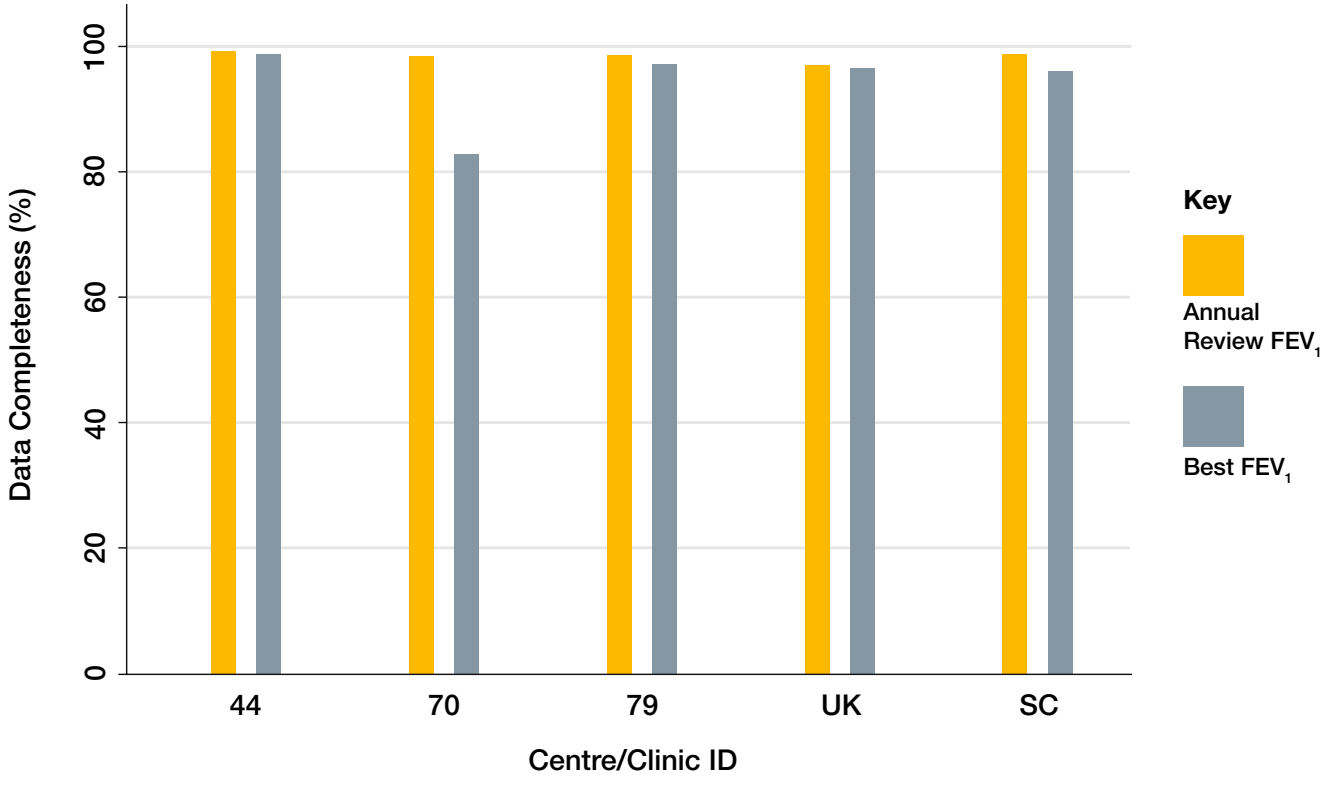
2.11 Inhaled antibiotic use for patients with chronic *Pseudomonas* by adult service



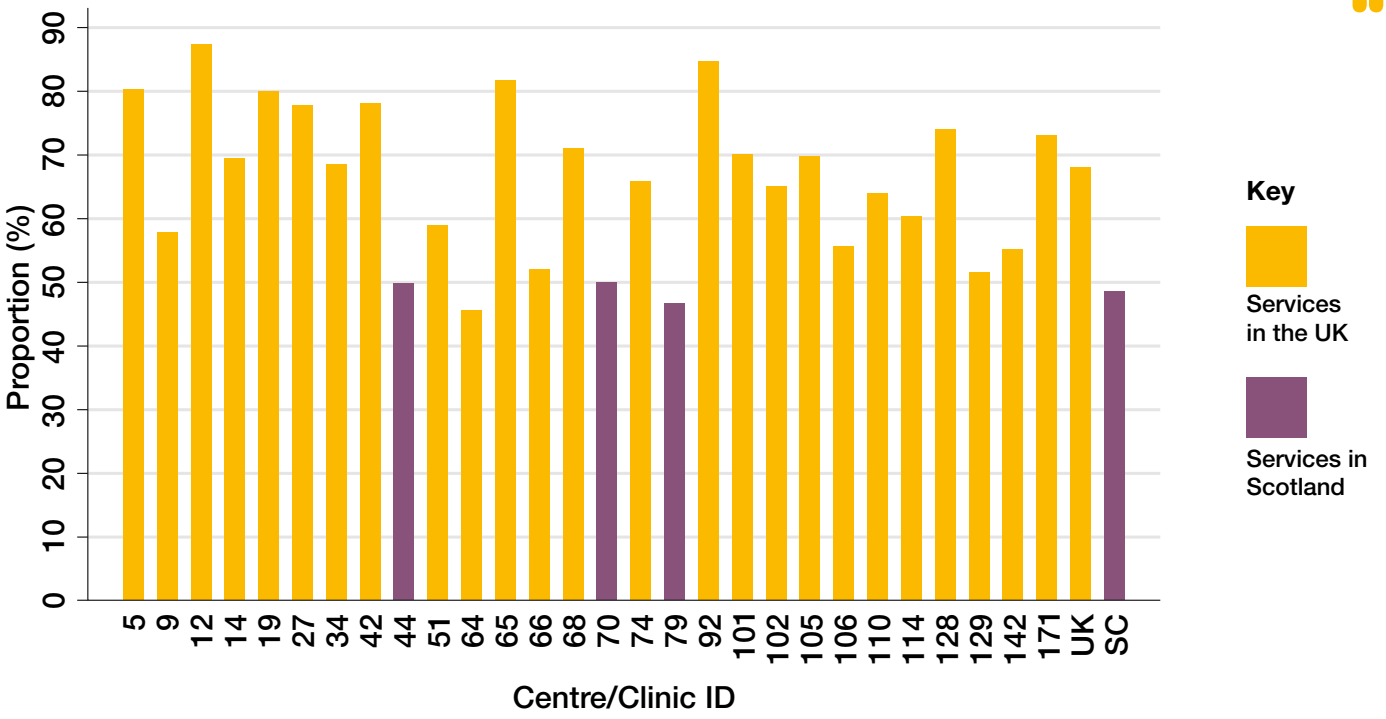
The proportion of chronic P. aeruginosa patients with receipt of inhaled antibiotics in Scotland during 2018 is 77.2%.

2.12 Data completeness by adult service

The mean data completeness for Annual Review FEV_1% predicted across Scottish adult centres in 2018 is 98.8%. The mean data completeness for Best FEV_1% predicted across adult Scottish centres is 96.0%.

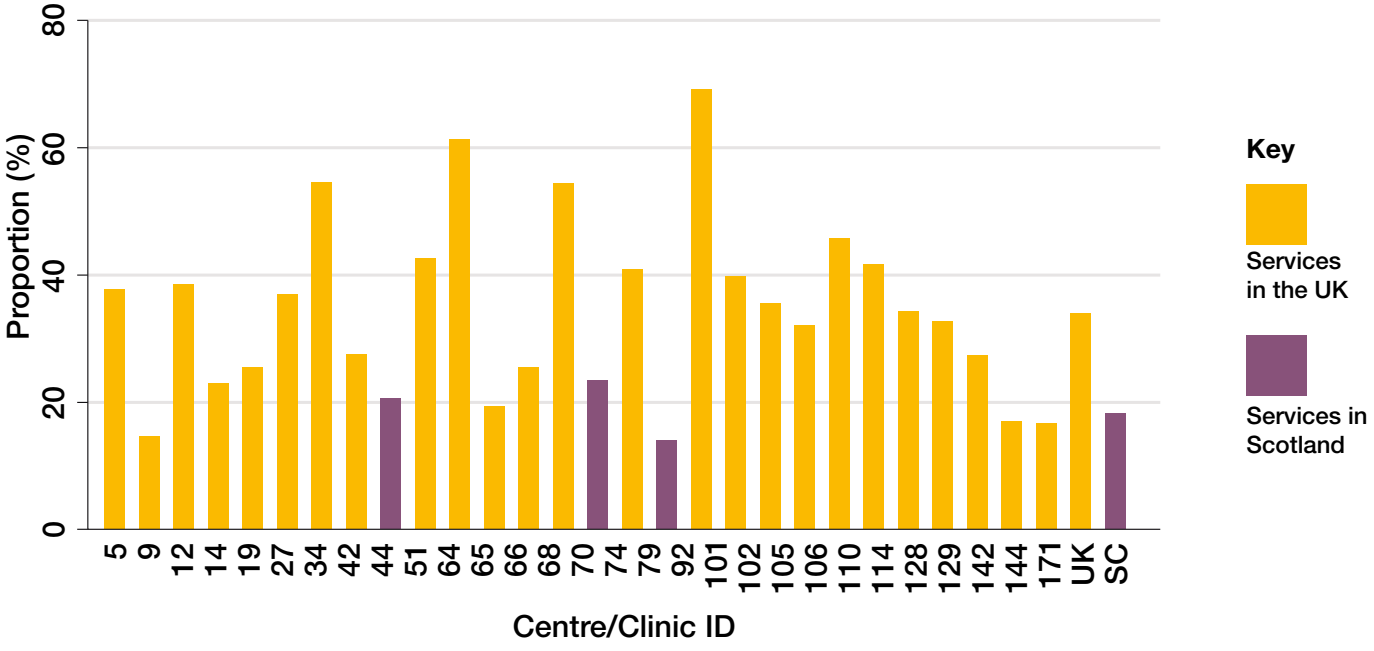


2.13 Proportion of patients receiving DNase treatment by adult service



The proportion of patients attending adult services in Scotland during 2018 receiving DNase treatment is 48.5%.

2.14 Proportion of patients receiving hypertonic saline treatment by adult service



The proportion of patients attending adult services in Scotland during 2018 receiving hypertonic saline treatment is 18.2%.

Appendices

Appendix 1: Centre-level data tables

Paediatric centres/clinics providing data in 2018 – ordered alphabetically

					Age		FEV_1% predicted at annual review	
Location	**Name**	**Clinic ID**	**Total Active**	**Number with annual review**	**Mean**	**Median**	**Number**	**Mean - unadjusted**
Scotland								
Aberdeen	Royal Aberdeen Children's Hospital	75	34	28	8.3	8.3	15	71.4
Ayr	University Hospital Crosshouse	170	28	28	8.6	7.5	18	90.3
Dundee	Ninewells Hospital	73	26	25	8.6	8.3	18	86.7
Edinburgh	Royal Hospital for Sick Children	143	136	124	9.3	9.8	88	92.2
Glasgow	Royal Hospital for Sick Children	56	93	51	8.8	8.7	36	88.7
Inverness	Raigmore Hospital	31	18	17	8.1	8.9	9	87.9
Lanarkshire	Wishaw General Hospital	162	43	41	8.9	8.3	30	90.8

Adult centres/clinics providing data in 2018 – ordered alphabetically

					Age		FEV_1% predicted at annual review	
Location	**Name**	**Clinic ID**	**Total Active**	**Number with annual review**	**Mean**	**Median**	**Number**	**Mean - unadjusted**
Scotland								
Aberdeen	Aberdeen Royal Infirmary	70	68	64	33.1	32.0	55	57.7
Edinburgh	Western General Hospital	44	249	233	32.7	30.0	213	61.8
Glasgow	Gartnavel General Hospital	79	232	208	32.9	29.0	200	65.0

*Where 'best' values were missing, or lower than FEV1% predicted taken at annual review, the annual review value was used.
**For data completeness, 'best' values were taken to be valid if they were not missing and the percent predicted was not more than 0.5% lower than FEV1% predicted taken at annual review.
† Adjusted for age - this means that the data have been fine-tuned to take account of the different spread of ages across centres and clinics. The adjusted values are intended to show what the average lung function or BMI percentile would be for that centre/clinic if the age spread is the same as the spread of age in the whole population".

FEV_1% predicted at annual review		Best FEV_1% predicted				Data completeness for FEV_1			
Mean – adjusted †	Median	Number	Mean - unadjusted	Mean - adjusted †	Median	Number with valid best FEV_1**	Percentage with valid best FEV_1	Number with FEV_1 at annual review	Percentage with FEV_1 at annual review
72.0	76.7	16	79.3	79.7	86.2	16	94.1	15	88.2
91.3	89.4	18	94.3	95.3	94.1	16	84.2	18	94.7
86.1	87.3	18	91.5	90.8	90.0	18	94.7	18	94.7
92.3	92.6	92	94.9	95.1	95.4	91	96.8	88	93.6
88.9	91.0	36	94.5	94.7	92.6	36	100.0	36	100.0
88.6	89.6	9	93.0	93.7	92.5	9	90.0	9	90.0
90.8	91.9	30	96.1	96.0	96.7	30	100.0	30	100.0

FEV_1% predicted at annual review		Best FEV_1% predicted				Data completeness for FEV_1			
Mean – adjusted †	Median	Number	Mean - unadjusted	Mean - adjusted †	Median	Number with valid best FEV_1**	Percentage with valid best FEV_1	Number with FEV_1 at annual review	Percentage with FEV_1 at annual review
57.9	54.2	56	63.4	63.5	60.5	54	84.4	63	98.4
61.8	59.5	213	66.8	66.8	68.4	230	98.7	231	99.1
65.1	65.7	203	69.8	69.9	72.3	205	98.6	205	98.6

*Where 'best' values were missing, or lower than FEV1% predicted taken at annual review, the annual review value was used.
**For data completeness, 'best' values were taken to be valid if they were not missing and the percent predicted was not more than 0.5% lower than FEV1% predicted taken at annual review.
† Adjusted for age - this means that the data have been fine-tuned to take account of the different spread of ages across centres and clinics. The adjusted values are intended to show what the average lung function or BMI percentile would be for that centre/clinic if the age spread is the same as the spread of age in the whole population".

Appendix 1: Centre-level data tables

Paediatric centres/clinics providing data in 2018 – ordered alphabetically

			BMI percentile			
Location	**Name**	**Clinic ID**	**Number**	**Mean unadjusted**	**Mean adjusted †**	**Median**
Scotland						
Aberdeen	Royal Aberdeen Children's Hospital	75	21	44.4	44.0	34.9
Ayr	University Hospital Crosshouse	170	23	62.5	62.5	63.9
Dundee	Ninewells Hospital	73	23	39.8	39.7	27.8
Edinburgh	Royal Hospital for Sick Children	143	101	54.2	54.5	54.4
Glasgow	Royal Hospital for Sick Children	56	41	51.4	51.7	61.4
Inverness	Raigmore Hospital	31	13	48.9	49.0	47.1
Lanarkshire	Wishaw General Hospital	162	37	51.9	51.8	51.4

Adult centres/clinics providing data in 2018 – ordered alphabetically

			BMI			
Location	**Name**	**Clinic ID**	**Number**	**Mean unadjusted**	**Mean adjusted †**	**Median**
Scotland						
Aberdeen	Aberdeen Royal Infirmary	70	64	23.6	23.5	23.1
Edinburgh	Western General Hospital	44	233	23.0	22.9	22.2
Glasgow	Gartnavel General Hospital	79	208	23.6	23.5	23.0

† Adjusted for age - this means that the data have been fine-tuned to take account of the different spread of ages across centres and clinics. The adjusted values are intended to show what the average lung function or BMI percentile would be for that centre/clinic if the age spread is the same as the spread of age in the whole population".

Chronic Pseudomonas		Having at least 1 IV day		Receiving DNase treatment		Receiving hypertonic saline treatment		Inhaled antibiotic use among patients with chronic Pseudomonas	
Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)
0	0	6	21.4	8	28.6	<5	-	0	0.0
<5	-	13	46.4	5	17.9	7	25.0	<5	-
<5	-	5	20.0	7	28.0	10	40.0	<5	-
5	4.0	29	23.4	65	52.4	20	16.1	5	100.0
<5	-	22	43.1	16	31.4	17	33.3	<5	-
<5	-	<5	-	<5	-	<5	-	<5	-
<5	-	9	22.0	7	17.1	11	26.8	<5	-

Chronic Pseudomonas		Having at least 1 IV day		Receiving DNase treatment		Receiving hypertonic saline treatment		Inhaled antibiotic use among patients with chronic Pseudomonas	
Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)
23	35.9	29	45.3	32	50.0	15	23.4	17	73.9
93	39.9	102	43.8	116	49.8	48	20.6	63	67.7
76	36.5	89	42.8	97	46.6	29	13.9	68	89.5

† Adjusted for age - this means that the data have been fine-tuned to take account of the different spread of ages across centres and clinics. The adjusted values are intended to show what the average lung function or BMI percentile would be for that centre/ clinic if the age spread is the same as the spread of age in the whole population".

Appendix 2: UK CF Registry Steering Committee structure

UK CF Registry Steering Committee

Role	Forename	Surname	Organisation
Commissioner, England	Kathy	Blacker	NHS England
CF Physician – paediatrics*	Siobhán	Carr	Royal Brompton Hospital
Senior Statistician †	Susan	Charman	Cystic Fibrosis Trust
Director of Data & Quality Improvement	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
Registry Clinical Data Manager †	Elaine	Gunn	Cystic Fibrosis Trust
Person with CF	Flora	Kennedy McConnel	N/A
Allied Health Professional	Alan	Peres	Royal Brompton Hospital
CF Physician - Adults	Simon	Range	Glenfield Hospital
Commissioner, Scotland	David	Steele	NHS Scotland
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	Diana	Bilton	Royal Brompton Hospital
CF physician – Adults (retired)			
Registry Consultant	Noreen	Caine	Cystic Fibrosis Trust
Pharmacovigilance PI	Siobhán	Carr	Royal Brompton Hospital
CF physician - Paediatrics			
Senior Statistician †	Susan	Charman	Cystic Fibrosis Trust
Director of Data & Quality Improvement	Rebecca	Cosgriff	Cystic Fibrosis Trust
Pharmacovigilance PI	Steve	Cunningham	Royal Hospital for Sick Children
CF Physician - Paediatrics			
Parent Representative	Marian	Dmochowska	N/A
Registry Clinical Data Manager †	Elaine	Gunn	Cystic Fibrosis Trust
Person with CF	James	Thomson	N/A
Pharmacovigilance PI	Nicholas	Simmonds	Royal Brompton Hospital
CF physician - Adults			
CF physician - Adults*#	Martin	Wildman	Northern General Hospital
Registry Development Manager †	Mary	Yip	Cystic Fibrosis Trust

*Chair † Non-voting member # Caldicott guardian

Appendix 3: Full list of mutations in the Scottish Population

The table below shows the number of people with CF who carry at least one of each mutation.
The groups are not mutually exclusive, as people with heterozygous mutations appear twice in the table.

Nucleotide	Protein	Legacy name	N	%
c.1521_1523delCTT	p.Phe508del	F508del	738	90.1
c.1652G->A	p.Gly551Asp	G551D	87	10.6
c.350G->A	p.Arg117His	R117H	66	8.1
c.1624G->T	p.Gly542X	G542X	54	6.6
c.200C->T	p.Pro67Leu	P67L	39	4.8
c.1679G->C	p.Arg560Thr	R560T	16	2.0
c.1477C->T	p.Gln493X	Q493X	15	1.8
c.1585-1G->A		1717-1G->A	13	1.6
c.3909C->G	p.Asn1303Lys	N1303K	12	1.5
c.489+1G->T		621+1G->T	12	1.5
c.3454G->C	p.Asp1152His	D1152H	12	1.5
c.2657+5G->A		2789+5G->A	9	1.1
c.3717+12191C->T		3849+10kbC->T	8	1.0
c.3528delC	p.Lys1177SerfsX15	3659delC	8	1.0
c.1558G->T	p.Val520Phe	V520F	7	0.9
c.178G->T	p.Glu60X	E60X	7	0.9
c.948delT	p.Phe316LeufsX12	1078delT	6	0.7
c.1210-12[5](AJ574948.1:g.152T[5])		5T	5	0.6
c.1766+1G->A		1898+1G->A	5	0.6
c.1364C->A	p.Ala455Glu	A455E	5	0.6
c.2657+2_2657+3insA		2789+2insA	<5	-
c.1721C->A	p.Pro574His	P574H	<5	-
c.3140-26A->G		3272-26A->G	<5	-
c.1519_1521delATC	p.Ile507del	I507del	<5	-
c.3196C->T	p.Arg1066Cys	R1066C	<5	-
c.1705T->G	p.Tyr569Asp	Y569D	<5	-
c.579+3A->G		711+3A->G	<5	-
c.509G->A	p.Arg170His	R170H	<5	-
c.2052delA	p.Lys684AsnfsX38	2184delA	<5	-
c.223C->T	p.Arg75X	R75X	<5	-
c.3846G->A	p.Trp1282X	W1282X	<5	-
c.2988G->A		3120G->A	<5	-
c.1367T->C	p.Val456Ala	V456A	<5	-
c.254G->A	p.Gly85Glu	G85E	<5	-
c.1209+1G->A		1341+1G->A	<5	-
c.1753G->T	p.Glu585X	E585X	<5	-
c.3484C->T	p.Arg1162X	R1162X	<5	-

Nucleotide	Protein	Legacy name	N	%
c.3705T->G	p.Ser1235Arg	S1235R	<5	-
c.2051_2052delAAinsG	p.Lys684SerfsX38	2183AA->G or 2183delAA->G	<5	-
c.3276C->A	p.Tyr1092X	Y1092X(C->A)	<5	-
c.2988+1G->A		3120+1G->A	<5	-
c.2158C->T	p.Gln720X	Q720X	<5	-
c.273+1G->A		405+1G->A	<5	-
c.164+2T>C		296+2T->C	<5	-
c.3158C->T	p.Thr1053Ile	T1053I	<5	-
c.2012delT	p.Leu671X	2143delT	<5	-
c.3884_3885insT	p.Ser1297PhefsX5	4016insT	<5	-
c.349C->G	p.Arg117Gly	R117G	<5	-
c.3476C->T	p.Ser1159Phe	S1159F	<5	-
c.3468G->A		3600G->A	<5	-
c.2583delT	p.Phe861LeufsX3	2711delT	<5	-
c.1055G->A	p.Arg352Gln	R352Q	<5	-
c.1327G->T	p.Asp443Tyr	D443Y	<5	-
c.2490+1G->A		2622+1G->A	<5	-
c.1647T->G	p.Ser549Arg	S549R(T->G)	<5	-
c.4147_4148insA	p.Ile1383AsnfsX3	4279insA	<5	-
c.2859_2890delACATTCTGTTCTTC AAGCACCTATGTCAACCC	p.Leu953PhefsX11	2991del32	<5	-
c.3266G->A	p.Trp1089X	W1089X	<5	-
c.1000C->T	p.Arg334Trp	R334W	<5	-
c.1006_1007insG	p.Ile336SerfsX28	1138insG	<5	-
c.443T->C	p.Ile148Thr	I148T	<5	-
c.1466C->A	p.Ser489X	S489X	<5	-
c.1657C->T	p.Arg553X	R553X	<5	-
Other selected			**67**	**8.2**

Glossary

Word/Phrase	Meaning
2018	1 January 2018 – 31 December 2018
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.
BMI (Body Mass Index)	A measure designed to show whether a person is a healthy weight for their height.
Burkholderia cepacia complex	*B. cepacia* complex are a group of bacteria, some of which threaten the health of people with cystic fibrosis.
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.
GI (Gastrointestinal)	The GI tract is the 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 gastrointestinal tract.
GLI equations	Global Lung Initiative, the equation used for calculating FEV_1% predicted from absolute FEV_1 that takes into account age, gender, height and ethnicity.
Haemophilus 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 cystic fibrosis 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.

Word/Phrase	Meaning
Homozygous	Everyone living with cystic fibrosis 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 2018 the median age of the 132 people who died was 26.
Median predicted survival	A mathematical formula predicts how long we expect half of people with CF born today will 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)	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.
NTM (nontuberculous mycobacteria)	A mycobacterium that does not cause tuberculosis, but which can cause respiratory infection. There are several known types.
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	An open sore that develops in the lining of the stomach, also known as a stomach ulcer.
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.
Prenatal	Before birth, while 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.
Staphylococcus 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.

Cystic Fibrosis Trust

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