Cystic Fibrosis strength in numbers

UK Cystic Fibrosis Registry
Annual Data Report 2019

Scotland

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Annual Data Report 2019 - Scotland

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Acknowledgements

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Introduction

This report is aimed at anyone who is interested in the health, care, and outcomes of people with cystic fibrosis (CF) in the 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 50.

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 the body to produce 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. 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.
The Registry Research Committee, which is a subcommittee of the RSC, assesses applications for data and guides the Registry research strategy.

Please see Appendix 1 of the UK Cystic Fibrosis Registry 2019 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 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?

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 Scotland, with comparisons to the full CF population of the United Kingdom, including CF centres in England, Northern Ireland, Scotland, and Wales.

1.1 Summary of the UK Cystic Fibrosis Registry

	2019	
	UK	Scotland
CF patients registered[1]	10655	931
Excluding diagnoses that year	10462	924
CF patients with an annual review; n(%)[2]	10070 (96%)	868 (94%)
Age in years; median[3]	21	22
All newly diagnosed patients (newborn screening and other)[4]	193	7
Number of patients born identified by newborn screening[4]	137	6
Age at diagnosis in months; median[3]	2	2
Adults aged 16 years and over; %[3]	60.6	61.3
Males; %[3]	53.2	54.0
Genotyped; %[3] (both mutations identified)	99.2	99.2
Total deaths reported (%)[5]	114 (1.1%)	12 (1.3%)
Age at death in years; median (95% CI)[5]	31 (29, 34)	26 (11, 42)

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.

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.

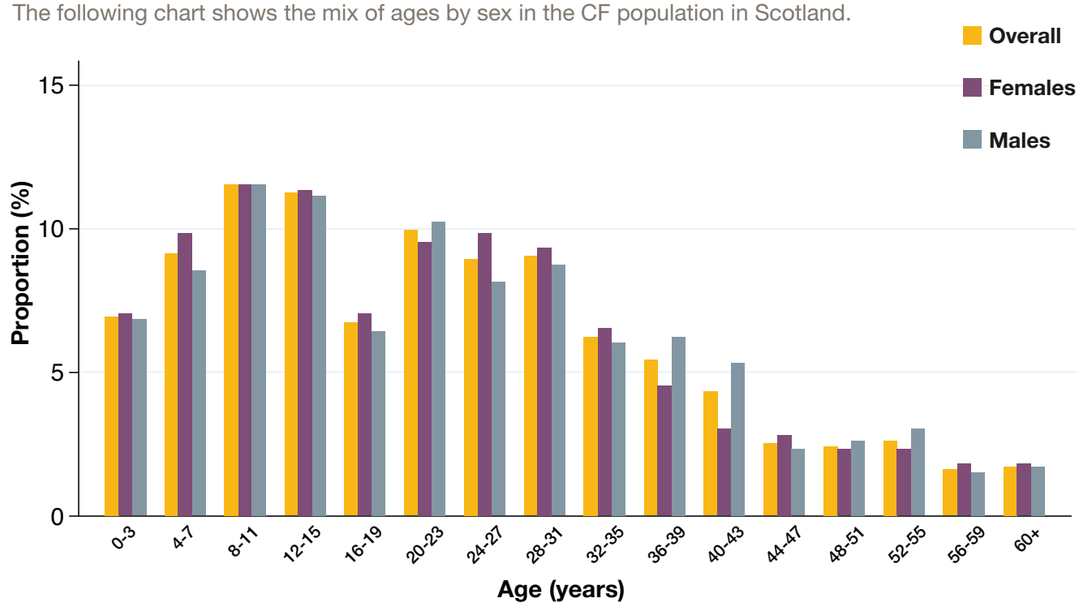
[3] Calculated from patients with an annual review in the given year (see footnote 2 above).

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

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

1.2 Age distribution by sex

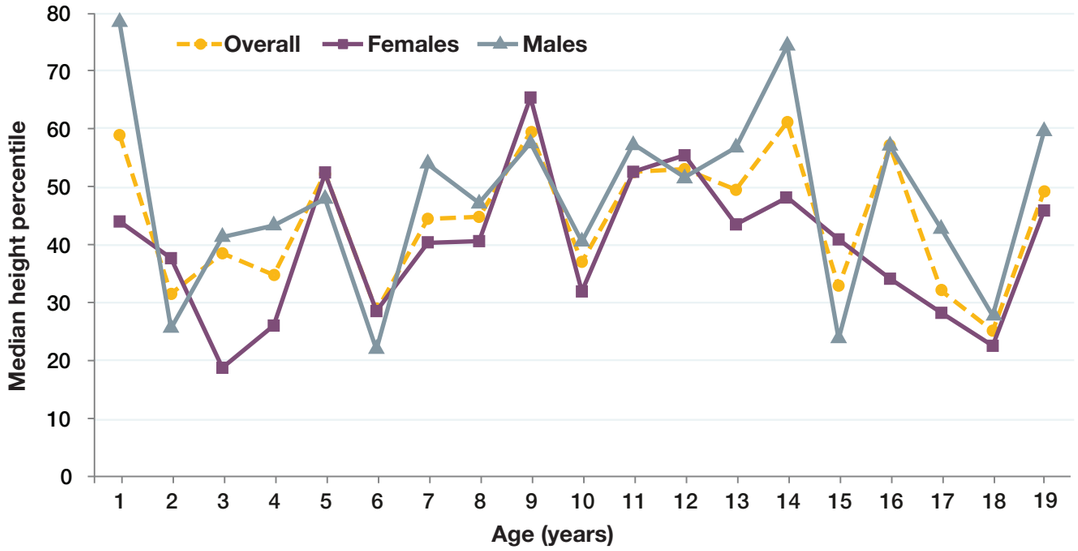
The following chart shows the mix of ages by sex in the CF population in Scotland.



Age	All; n (%)	Females; n (%)	Males; n (%)
0-3	60 (6.9)	28 (7.0)	32 (6.8)
4-7	79 (9.1)	39 (9.8)	40 (8.5)
8-11	100 (11.5)	46 (11.5)	54 (11.5)
12-15	97 (11.2)	45 (11.3)	52 (11.1)
16-19	58 (6.7)	28 (7.0)	30 (6.4)
20-23	86 (9.9)	38 (9.5)	48 (10.2)
24-27	77 (8.9)	39 (9.8)	38 (8.1)
28-31	78 (9.0)	37 (9.3)	41 (8.7)
32-35	54 (6.2)	26 (6.5)	28 (6.0)
36-39	47 (5.4)	18 (4.5)	29 (6.2)
40-43	37 (4.3)	12 (3.0)	25 (5.3)
44-47	22 (2.5)	11 (2.8)	11 (2.3)
48-51	21 (2.4)	9 (2.3)	12 (2.6)
52-55	23 (2.6)	9 (2.3)	14 (3.0)
56-59	14 (1.6)	7 (1.8)	7 (1.5)
60+	15 (1.7)	7 (1.8)	8 (1.7)
<16	336 (38.7)	158 (39.6)	178 (38.0)
≥16	532 (61.3)	241 (60.4)	291 (62.0)
<18	366 (42.2)	170 (42.6)	196 (41.8)
≥18	502 (57.8)	229 (57.4)	273 (58.2)
Overall	**868**	**399**	**469**

1.3 Height percentiles of children and young people (<20 years)[6] N=394

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.

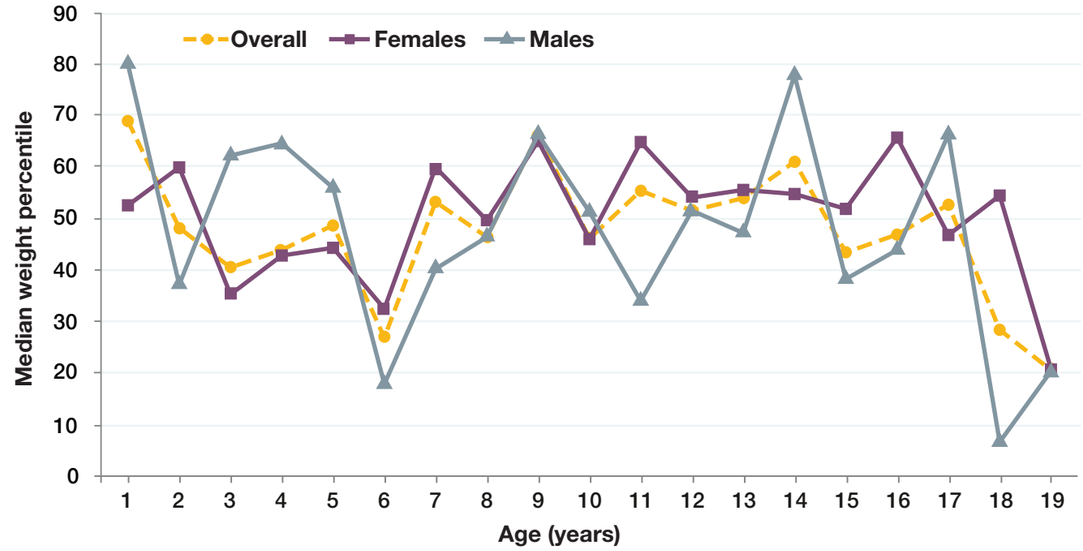


Age (years)	Overall			Female			Male		
	n	Median	IQR	n	Median	IQR	n	Median	IQR
1	22	58.8	22.6-90.7	9	43.9	22.6-74.2	13	78.3	29.8-94.5
2	17	31.4	11.6-58.6	10	37.5	7.3-59.8	7	25.4	11.6-58.6
3	18	38.4	15.8-60.1	7	18.6	2.9-52.0	11	41.2	37.2-88.6
4	22	34.7	23.6-68.8	13	25.9	22.5-57.7	9	43.2	29.7-88.3
5	15	52.2	30.8-65.2	5	52.2	28.2-65.9	10	47.8	31.4-60.7
6	16	28.4	8.1-42.0	8	28.4	10.8-40.5	8	21.8	7.9-43.6
7	26	44.3	18.2-62.4	13	40.2	20.4-60.0	13	53.8	16.1-64.3
8	33	44.7	19.0-58.4	16	40.5	8.9-58.1	17	47.1	21.9-59.8
9	24	59.3	39.5-76.3	6	65.3	59.9-79.9	18	57.5	33.7-73.5
10	25	36.9	13.8-52.0	14	31.8	14.3-68.7	11	40.4	8.2-52.0
11	18	52.5	26.5-82.3	10	52.5	26.5-88.2	8	57.3	20.5-76.4
12	22	53.0	12.3-65.1	10	55.3	5.0-65.1	12	51.5	14.5-72.8
13	29	49.4	32.6-81.5	12	43.4	29.0-80.5	17	56.7	32.6-94.4
14	19	61.1	30.1-87.4	8	48.1	9.6-82.6	11	74.3	56.5-90.4
15	27	32.7	13.7-61.7	15	40.8	13.7-69.0	12	23.6	13.8-47.4
16	10	56.9	51.9-68.9	<5	-	-	-	57.0	53.0-68.9
17	20	31.9	15.2-61.1	11	28.1	13.9-59.7	9	42.6	22.6-62.4
18	14	25.0	7.8-52.8	9	22.4	10.4-52.6	5	27.6	7.1-57.8
19	14	49.1	14.6-65.1	7	45.8	13.6-65.3	7	59.5	14.6-65.1
Overall	**391***	**43.5**	**17.7-67.7**	**184**	**40.1**	**15.1-62.1**	**207**	**50.2**	**20.1-71.2**

*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=394

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.

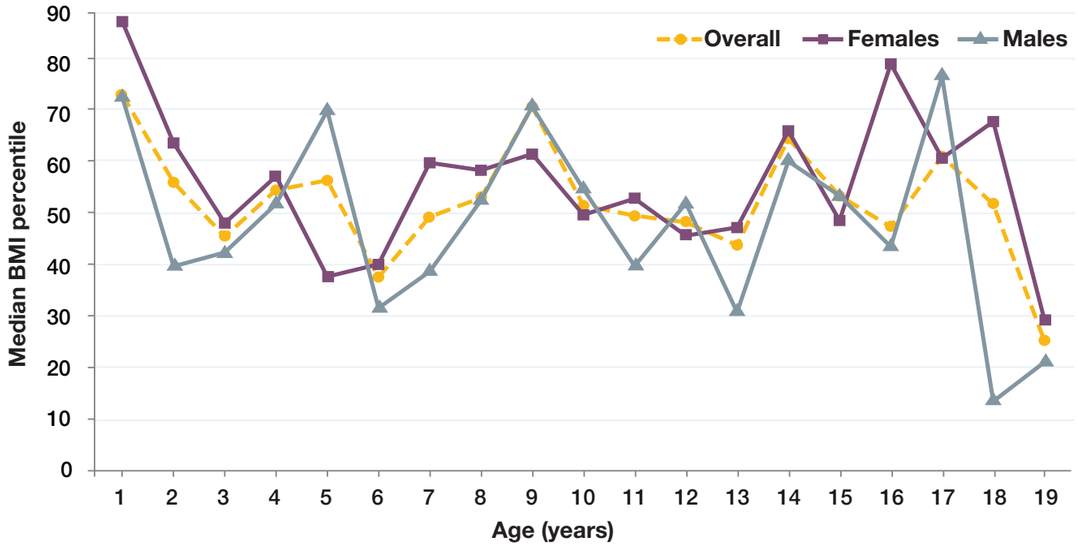


Age (years)	Overall			Female			Male		
	n	Median	IQR	n	Median	IQR	n	Median	IQR
1	22	69.1	24.7-92.2	9	52.6	46.9-72.9	13	80.1	22.8-93.2
2	17	48.2	24.0-68.1	10	60.0	39.0-72.7	7	37.1	13.8-48.2
3	18	40.5	14.8-69.3	7	35.2	8.3-92.0	11	62.3	22.6-69.3
4	22	43.9	23.8-75.7	13	42.8	23.8-66.6	9	64.7	43.3-76.6
5	15	48.7	32.6-64.5	5	44.3	43.5-48.7	10	56.0	32.6-69.6
6	16	26.8	12.8-36.5	8	32.4	19.0-41.5	8	17.7	8.1-32.5
7	26	53.3	18.3-73.2	13	60.0	22.1-72.1	13	40.2	18.3-81.1
8	33	46.4	15.8-66.3	16	49.7	12.3-71.2	17	46.4	20.6-61.1
9	24	66.4	38.8-87.7	6	65.1	41.5-81.0	18	66.4	36.2-88.3
10	25	46.1	19.7-71.9	14	46.0	19.7-58.1	11	51.3	16.0-75.4
11	18	55.5	27.0-80.4	10	64.9	28.2-80.4	8	34.0	23.4-75.2
12	22	51.5	19.4-75.7	10	54.1	13.6-90.1	12	51.5	30.8-72.9
13	29	54.0	29.1-67.3	12	55.6	36.0-64.3	17	47.2	15.3-76.7
14	19	61.1	47.4-82.2	8	54.8	43.6-72.6	11	78.0	54.2-84.6
15	27	43.4	24.5-58.7	15	51.9	24.1-76.9	12	38.2	26.2-52.8
16	10	46.9	32.4-72.4	<5	-	-	-	43.9	32.4-72.4
17	20	52.8	18.5-81.8	11	46.8	8.2-84.1	9	66.4	20.9-73.6
18	14	28.3	8.3-69.6	9	54.5	17.0-71.5	5	6.4	4.1-8.3
19	14	20.5	6.4-59.0	7	20.7	6.4-84.1	7	20.2	5.0-44.0
Overall	**391***	**48.0**	**20.7-73.2**	**184**	**49.8**	**21.0-72.4**	**207**	**47.2**	**20.6-75.7**

*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=394

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.

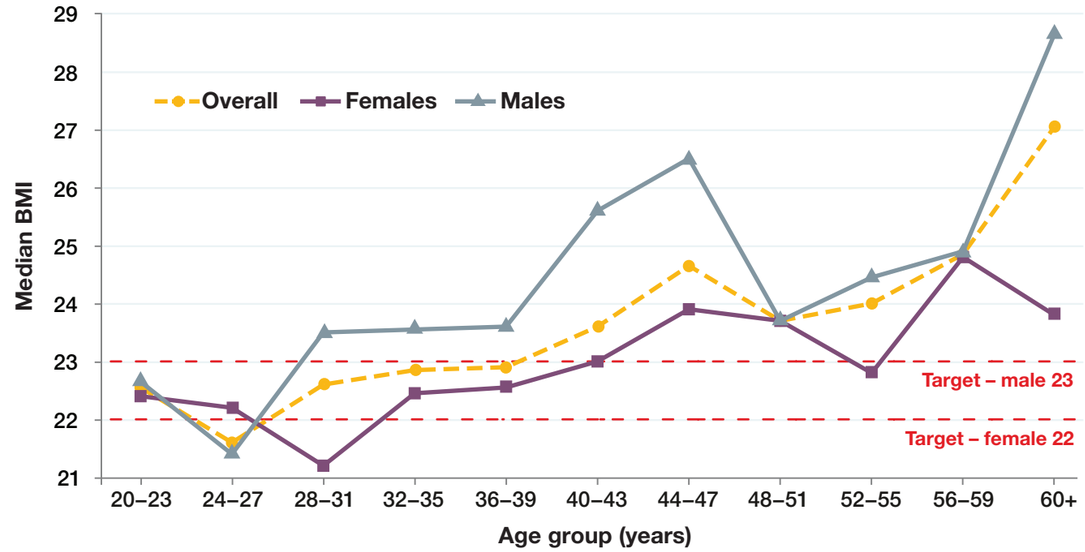


Age (years)	Overall			Female			Male		
	n	Median	IQR	n	Median	IQR	n	Median	IQR
1	22	72.8	22.8-91.5	9	87.0	17.9-92.3	13	72.3	31.8-87.3
2	17	55.8	37.6-71.1	10	63.4	42.9-95.4	7	39.5	32.0-69.8
3	18	45.3	27.0-72.2	7	47.9	44.9-95.9	11	42.2	20.4-66.9
4	22	54.3	26.2-70.1	13	57.0	26.2-68.1	9	51.6	43.2-91.1
5	15	56.2	25.3-70.0	5	37.5	29.7-45.1	10	69.7	25.3-72.0
6	16	37.4	23.8-52.0	8	39.9	31.1-60.4	8	31.4	17.9-42.2
7	26	49.1	22.6-82.7	13	59.6	18.7-75.5	13	38.5	25.0-82.9
8	33	52.8	27.5-71.7	16	58.2	24.5-74.2	17	52.3	32.4-61.8
9	24	70.5	44.8-83.8	6	61.3	43.9-75.9	18	70.5	45.7-91.3
10	25	51.3	41.9-75.2	14	49.5	36.5-63.2	11	54.5	42.8-78.8
11	18	49.3	24.3-77.2	10	52.7	40.2-80.1	8	39.5	21.3-64.8
12	22	48.1	27.8-81.7	10	45.5	31.2-91.8	12	51.8	26.4-80.0
13	29	43.6	12.0-65.0	12	47.0	11.6-66.9	17	30.6	12.0-65.0
14	19	64.3	51.7-84.8	8	65.8	55.6-72.7	11	59.9	33.3-88.4
15	27	53.2	29.5-71.7	15	48.4	20.0-86.1	12	53.2	44.3-65.2
16	10	47.2	35.2-78.8	<5	-	-	-	43.3	35.2-56.0
17	20	60.8	17.1-80.9	11	60.5	12.4-77.1	9	76.4	21.9-82.8
18	14	51.5	22.7-81.5	9	67.6	29.5-92.5	5	13.3	9.3-47.5
19	14	25.0	4.7-80.0	7	29.1	4.7-81.9	7	20.9	3.0-41.1
Overall	**391***	**51.7**	**26.5-76.8**	**184**	**53.2**	**29.3-77.4**	**207**	**48.9**	**25.0-76.1**

*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) N=474

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].



Age (years)	Overall			Female			Male		
	n	Median	IQR	n	Median	IQR	n	Median	IQR
20-23	86	22.5	20.6-24.9	38	22.4	20.3-25.5	48	22.7	20.9-24.4
24-27	77	21.6	19.1-24.4	39	22.2	19.0-24.9	38	21.4	19.1-23.8
28-31	78	22.6	20.3-25.6	37	21.2	20.2-24.3	41	23.5	21.2-25.7
32-35	54	22.9	21.4-25.1	26	22.5	20.8-24.5	28	23.5	22.0-26.9
36-39	47	22.9	20.6-25.2	18	22.5	20.0-25.2	29	23.6	21.4-25.1
40-43	37	23.6	21.7-26.8	12	23.0	21.8-23.9	25	25.6	21.5-27.4
44-47	22	24.7	22.5-27.2	11	23.9	20.3-25.7	11	26.5	23.3-30.0
48-51	21	23.7	22.4-26.8	9	23.7	22.0-25.1	12	23.7	22.5-27.1
52-55	23	24.0	22.3-27.0	9	22.8	21.9-24.5	14	24.5	22.8-27.0
56-59	14	24.8	22.6-26.6	7	24.8	22.6-30.0	7	24.9	21.1-26.6
60+	14	27.0	22.9-30.0	6	23.8	17.0-28.2	8	28.6	25.8-30.0
Overall	**473***	**22.9**	**20.7-25.7**	**212**	**22.6**	**20.2-24.9**	**261**	**23.4**	**21.2-26.3**

*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)
N=532

The following table shows how people with CF reported their education and employment status in 2019. Please note that the groups are not mutually exclusive; for example, someone may be a student as well as working part-time.

	All patients; n (%)	Male; n (%)	Female; n (%)
Number of patients	532	291	241
Number who completed questionnaire; n (%)	532 (100.0)	291 (100.0)	241 (100.0)
Full-time employment; n (%)	199 (37.4)	135 (46.4)	64 (26.6)
Part-time employment; n (%)	101 (19.0)	35 (12.0)	66 (27.4)
Student; n (%)	69 (13.0)	37 (12.7)	32 (13.3)
Homemaker; n (%)	18 (3.4)	<5	-
Unemployed; n (%)	92 (17.3)	57 (19.6)	35 (14.5)
Disabled; n (%)	22 (4.1)	8 (2.7)	14 (5.8)
Retired; n (%)	19 (3.6)	10 (3.4)	9 (3.7)
Unknown entered; n (%)	10 (1.9)	-	<5
Total patients in work or study; n (%)	369 (69.4)	207 (71.1)	162 (67.2)

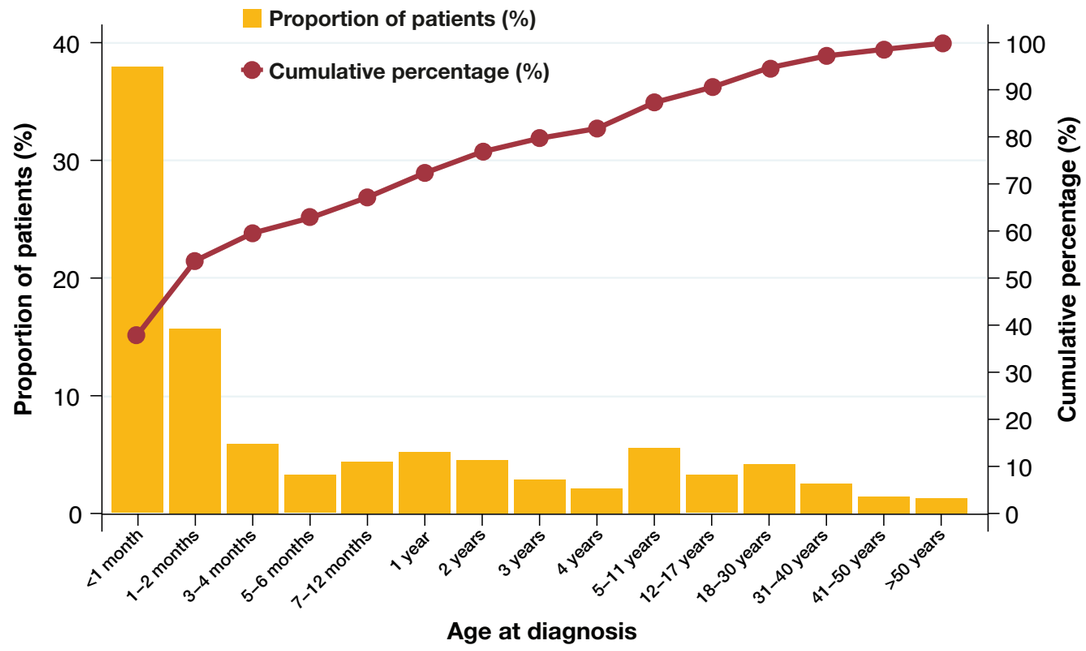
1.8 Pregnancy

seven women with cystic fibrosis had babies in Scotland during 2019

Fewer than five men with cystic fibrosis became fathers in Scotland during 2019

Diagnosis of cystic fibrosis

1.9 Age at diagnosis and screening in children under 16 in 2019
N=346



The median (range) age at diagnosis for patients aged under 16 in 2019 is **20 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 six patients born in 2019 were identified by newborn screening (including those without complete data).

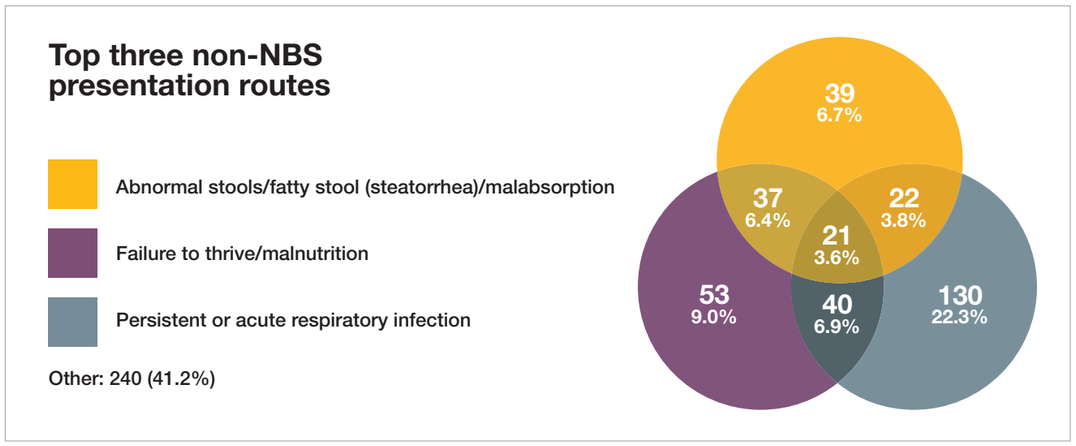
87 (10.0%) of Scottish CF patients were diagnosed at age 16 or over. No new CF diagnoses were recorded in Scotland for people aged 16 or over during 2019.

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	868	779	87
Number diagnosed by NBS	284	284	0
Total non-NBS	**584=**	**495**	**87**

Mode of presentation (excluding NBS)	All patients (N=868)		Age <16 at diagnosis* (n=779)		Age ≥16 at diagnosis* (n=87)	
	n	(%)	n	(%)	n	(%)
Persistent or acute respiratory infection	213	36.3	167	33.7	46	52.9
Failure to thrive/malnutrition	151	25.8	151	30.5	0	0.0
Abnormal stools/fatty stool (steatorrhea)/malabsorption	119	20.3	114	23.0	5	5.7
Meconium ileus	94	16.0	94	19.0	0	0.0
Family history	83	14.2	72	14.5	11	12.6
Genotype	43	7.3	34	6.9	9	10.3
Unknown	27	4.6	20	4.0	7	8.0
Rectal prolapse	22	3.8	-	-	<5	-
Nasal polyps	16	2.7	16	3.2	0	0.0
Bronchiectasis	11	1.9	<5	-	-	-
Prenatal	5	0.9	<5	-	<5	-
Electrolyte imbalance	<5	-	0	0.0	<5	-
Liver disease	<5	-	<5	-	0	0.0
Fertility	<5	-	<5	-	0	0.0
Pancreatitis	<5	-	<5	-	0	0.0
Oedema	0	0.0	0	0.0	0	0.0



*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, thickened 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) at Annual Review in patients aged 6 years and older who have not had a lung transplant
N=737

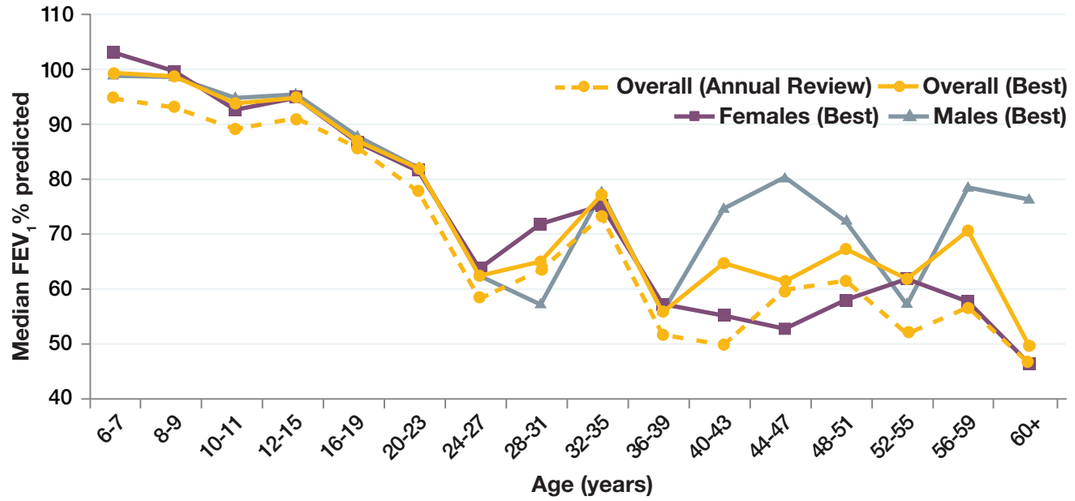
People with CF who have had lung transplants are excluded, as their new 'non-CF' lungs may have lung health similar to a person without cystic fibrosis.

	Overall			Female			Male		
Age (yrs)	n	Median	IQR	n	Median	IQR	n	Median	IQR
6-7	37	94.6	87.1-103.2	19	97.8	90.0-109.8	18	93.8	73.2-97.7
8-9	51	93.0	86.4-99.3	19	93.6	89.0-102.9	32	92.7	82.1-98.8
10-11	42	89.1	81.0-97.9	24	85.6	78.4-104.6	18	90.2	82.6-95.5
12-15	94	91.0	82.8-98.8	43	88.8	83.1-96.3	51	92.8	82.7-100.1
16-19	56	85.7	69.2-93.2	28	84.6	69.2-93.1	28	86.1	66.3-94.1
20-23	84	77.7	51.3-92.2	37	76.6	49.4-94.0	47	79.4	55.0-88.6
24-27	75	58.2	36.1-80.2	38	58.3	38.2-76.6	37	54.8	34.6-82.3
28-31	73	63.3	44.1-76.8	35	66.0	53.0-79.5	38	54.2	37.2-74.2
32-35	48	73.2	51.6-82.4	21	66.2	51.5-81.5	27	75.0	51.7-85.3
36-39	40	51.6	38.1-67.1	16	52.7	40.2-67.1	24	50.6	37.1-67.1
40-43	32	49.8	36.7-89.1	11	43.7	34.4-85.2	21	70.1	39.7-89.6
44-47	17	59.8	48.5-80.7	9	48.5	42.3-59.8	8	82.7	57.6-87.5
48-51	21	61.5	45.5-70.2	9	53.3	45.5-64.1	12	67.8	48.7-81.3
52-55	21	51.8	44.9-75.7	9	55.7	44.9-74.2	12	48.4	39.2-89.2
56-59	13	56.6	44.2-76.0	6	53.9	46.3-58.4	7	76.0	35.3-84.7
60+	13	46.4	40.7-75.7	5	40.7	24.8-46.3	8	67.5	45.5-82.2
<16	224	92.1	82.5-99.3	105	91.6	83.1-102.9	119	92.5	82.1-98.5
≥16	493	65.5	43.7-84.9	224	63.8	44.8-82.9	269	66.0	42.5-85.8
<18	253	91.6	82.0-98.9	117	91.6	82.0-102.4	136	91.9	81.9-98.3
≥18	464	62.5	42.5-83.2	212	60.1	44.2-81.1	252	63.1	42.2-84.8
Overall	**717***	**77.7**	**51.7-92.5**	**329**	**76.6**	**51.4-92.9**	**388**	**78.9**	**52.0-92.1**

*number with non-missing data

1.12 Best FEV1% predicted (GLI equations) in patients aged 6 years and older who have not had a lung transplant
N=737

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.

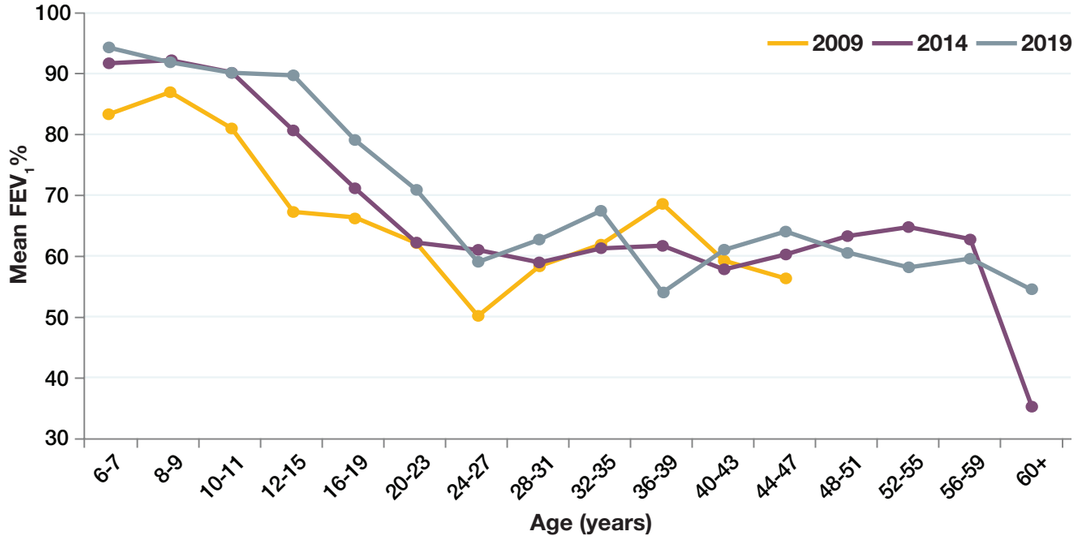


	Overall			Female			Male		
Age (yrs)	n	Median	IQR	n	Median	IQR	n	Median	IQR
6-7	38	99.3	90.0-110.3	19	103.0	91.3-113.7	19	98.7	82.0-104.3
8-9	57	98.7	89.8-104.8	22	99.6	89.8-105.6	35	98.5	89.2-104.4
10-11	42	93.7	84.8-104.7	24	92.5	85.0-108.8	18	94.7	83.6-97.9
12-15	97	94.7	85.6-102.2	45	94.7	85.6-99.8	52	95.3	85.3-102.7
16-19	56	86.9	73.2-98.5	28	86.6	75.9-96.4	28	87.7	73.2-101.6
20-23	85	81.9	55.6-93.0	37	81.4	51.3-94.0	48	82.0	61.0-92.0
24-27	75	62.3	44.6-85.5	38	63.6	49.9-83.3	37	62.3	42.9-85.5
28-31	73	64.9	49.5-80.4	35	71.8	59.1-86.7	38	57.1	40.6-78.4
32-35	49	77.1	55.6-83.6	21	75.1	59.9-82.2	28	77.4	52.6-86.4
36-39	40	55.7	44.3-72.0	16	57.2	49.1-80.3	24	55.7	42.0-68.9
40-43	32	64.7	44.0-90.5	11	55.1	35.5-91.5	21	74.6	44.8-89.6
44-47	18	61.3	51.5-80.7	9	52.7	47.5-63.7	9	80.3	60.8-86.8
48-51	21	67.2	53.2-77.7	9	58.0	47.8-67.2	12	72.2	59.3-84.8
52-55	21	61.8	45.1-81.8	9	61.8	45.1-80.8	12	57.0	39.5-89.2
56-59	13	70.6	47.1-78.4	6	57.6	47.1-70.6	7	78.4	45.2-85.8
60+	13	49.6	46.3-76.8	5	46.3	27.6-49.6	8	76.2	48.2-85.8
<16	234	96.1	87.8-103.5	110	95.9	88.4-104.4	124	96.7	86.0-102.7
≥16	496	70.9	49.6-87.6	224	70.6	51.6-87.1	272	71.8	47.1-88.3
<18	263	95.8	86.4-103.5	122	95.1	88.3-103.9	141	96.0	85.8-102.7
≥18	467	68.8	49.0-86.7	212	67.5	51.2-86.0	255	69.2	46.4-87.1
Overall	**730***	**82.5**	**58.0-96.3**	**334**	**82.4**	**59.1-96.2**	**396**	**82.7**	**56.3-96.5**

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=737 in 2019, N=640 in 2014, N=375 in 2009*

The chart below shows how FEV_1% in 2019 compares to Registry data from 2009 and 2014. 2009 is shown as a comparator year.

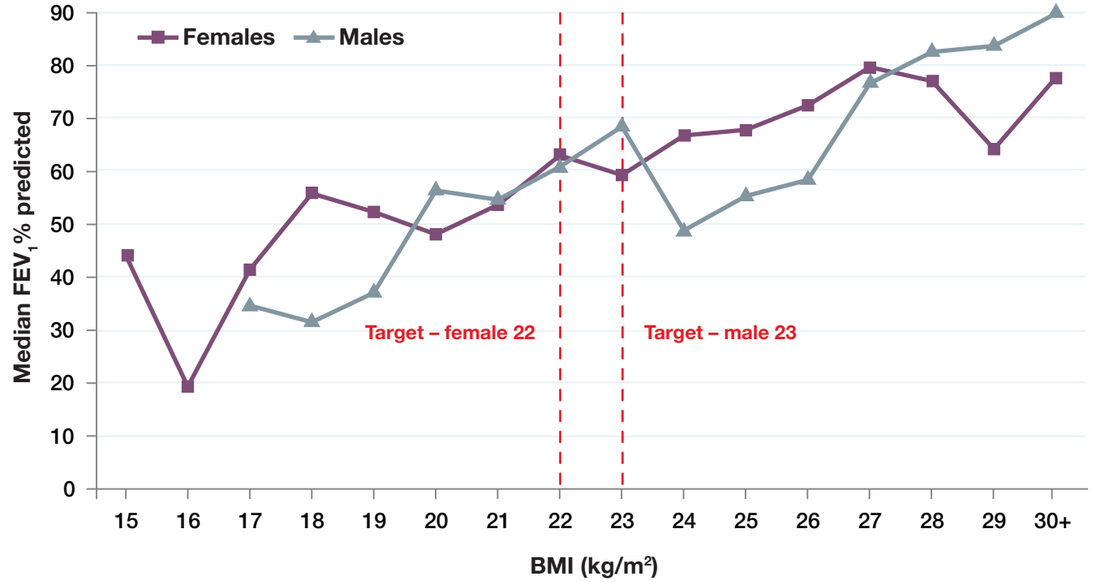


Age (years)	2009 mean FEV_1%	2014 mean FEV_1%	2019 mean FEV_1%	p-values (t-test)**
6-7	83.3	91.6	94.2	0.495
8-9	86.9	92.2	91.8	0.901
10-11	81.0	90.2	90.1	0.986
12-15	67.3	80.6	89.7	<0.001
16-19	66.3	71.1	79.1	0.052
20-23	62.1	62.2	70.9	0.018
24-27	50.2	61.0	59.0	0.637
28-31	58.3	59.0	62.7	0.365
32-35	61.9	61.3	67.4	0.249
36-39	68.5	61.7	54.0	0.200
40-43	59.2	57.8	61.0	0.722
44-47	56.3	60.3	64.0	0.601
48-51	***	63.3	60.6	0.702
52-55	***	64.8	58.1	**
56-59	***	62.8	59.6	**
60+	***	35.3	54.5	**
<16	74.8	87.9	91.0	N/A
16+	60.7	62.7	64.5	N/A
<18	73.9	86.4	90.0	N/A
18+	59.9	61.1	63.4	N/A

* due to missing data, means are calculated from a population of 717 in 2019, 604 in 2014 and 273 in 2009.
** t-test comparing 2019 with 2014. If the p-value is less than 0.05 then the difference in the mean is statistically significant.
***due to small numbers in these age groups, the mean for the age group marked 44-47 is for all people aged 44 and older.

1.14 FEV_1% predicted (GLI equations) and BMI in people aged 20 years and over who have not had a transplant
N=441

The goal BMI for adults is 22 for women, and 23 for men[1]. 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.

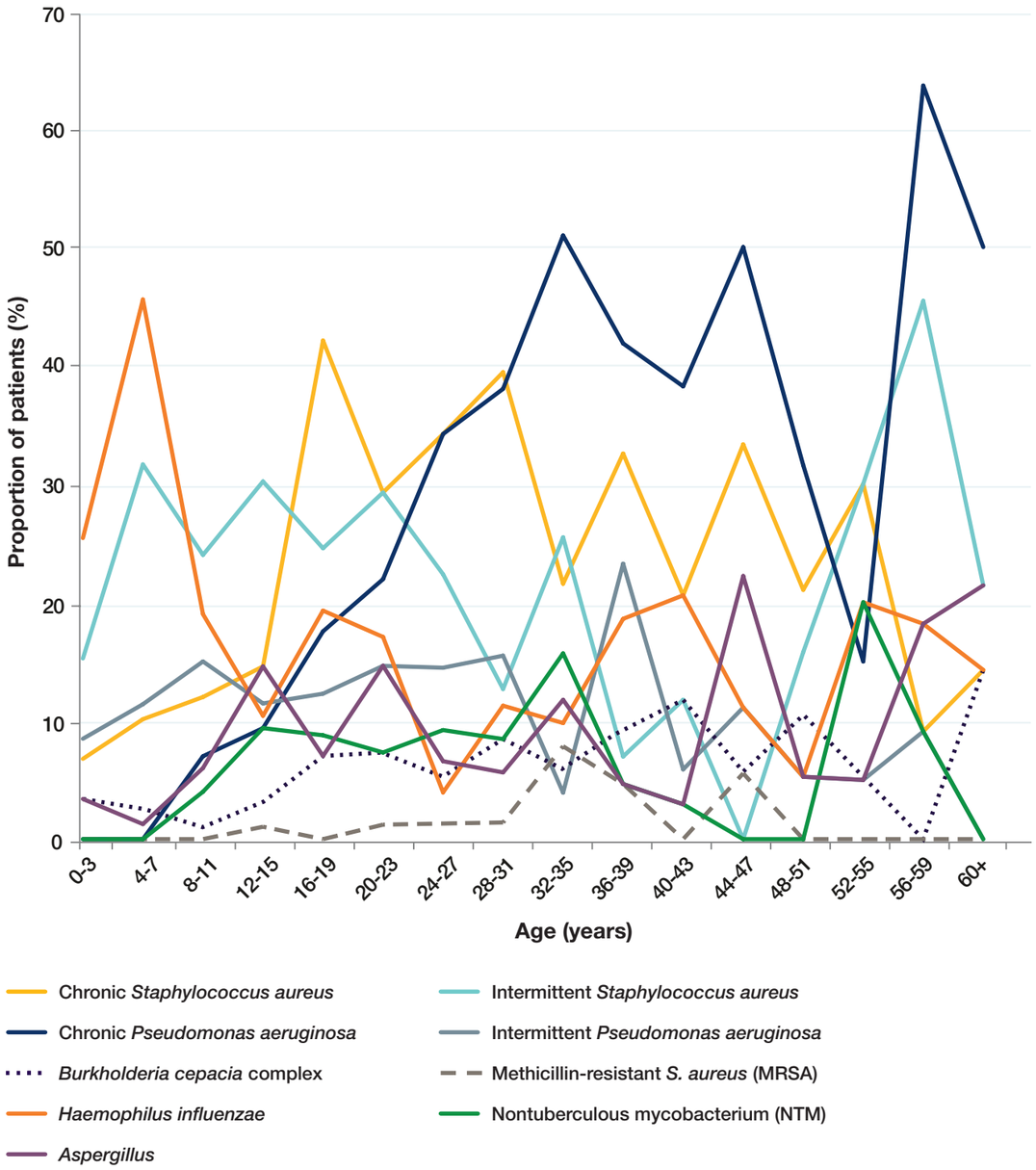


[1] Stallings et al. J Am Diet Assoc. 2008 08:832-839

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 2019
N=868



1.16 Lung infections in 2019
<16 years N=336; ≥16 years N=532

Infections in this table reflect bugs grown in the 12 months prior to the 2019 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	<16 years
Patients in age range; n	60	79	100	97	336
Patients who had culture taken; n*	59	79	100	96	334
Chronic *Staphylococcus aureus*; n (%)	<5	-	12 (12.0)	14 (14.6)	38 (11.4)
Intermittent *Staphylococcus aureus*; n (%)	9 (15.3)	25 (31.6)	24 (24.0)	29 (30.2)	87 (26.0)
Chronic *Pseudomonas aeruginosa*; n (%)	0 (0.0)	0 (0.0)	7 (7.0)	9 (9.4)	16 (4.8)
Intermittent *Pseudomonas aeruginosa*; n (%)	5 (8.5)	9 (11.4)	15 (15.0)	11 (11.5)	40 (12.0)
Burkholderia cepacia complex; n (%)	<5	<5	<5	<5	8 (2.4)
B. cenocepacia; n (%)	0 (0.0)	<5	0 (0.0)	<5	3 (0.9)
B. multivorans; n (%)	0 (0.0)	0 (0.0)	<5	<5	<5
Other Burkolderia; n (%)	<5	<5	0 (0.0)	0 (0.0)	<5
MRSA; n (%)	0 (0.0)	0 (0.0)	0 (0.0)	<5	<5
Haemophilus influenzae; n (%)	15 (25.4)	36 (45.6)	19 (19.0)	10 (10.4)	80 (24.0)
Nontuberculous mycobacterium; n (%)	0 (0.0)	0 (0.0)	<5	-	13 (3.9)
Aspergillus; n (%)	<5	<5	6 (6.0)	14 (14.6)	23 (6.9)

	Adult age range (years)						Overall
	16-19	20-23	24-27	28-31	32-35	36-39	≥16 years
Patients in age range; n	58	86	77	78	54	47	532
Patients who had culture taken; n	57	82	76	71	51	43	496
Chronic *S. aureus*; n (%)	24 (42.1)	24 (29.3)	26 (34.2)	28 (39.4)	11 (21.6)	14 (32.6)	153 (30.8)
Intermittent *S. aureus*; n (%)	14 (24.6)	24 (29.3)	17 (22.4)	9 (12.7)	13 (25.5)	3 (7.0)	101 (20.4)
Chronic *P. aeruginosa*; n (%)	10 (17.5)	18 (22.0)	26 (34.2)	27 (38.0)	26 (51.0)	18 (41.9)	170 (34.3)
Intermittent *P. aeruginosa*; n (%)	7 (12.3)	12 (14.6)	11 (14.5)	11 (15.5)	<5	10 (23.3)	60 (12.1)
B. cepacia complex; n (%)	<5	6 (7.3)	<5	6 (8.5)	<5	<5	37 (7.5)
B. cenocepacia; n (%)	<5	<5	<5	<5	0 (0.0)	<5	7 (1.4)
B. multivorans; n (%)	<5	<5	<5	<5	0 (0.0)	<5	18 (3.6)
Other Burkholderia; n (%)	0 (0.0)	<5	0 (0.0)	<5	0 (0.0)	0 (0.0)	<5
MRSA; n (%)	0 (0.0)	<5	<5	<5	<5	<5	10 (2.0)
H. influenzae; n (%)	11 (19.3)	14 (17.1)	<5	8 (11.3)	<5	8 (18.6)	67 (13.5)
Nontuberculous mycobacterium; n (%)	5 (8.8)	6 (7.3)	7 (9.2)	6 (8.5)	8 (15.7)	<5	40 (8.1)
Aspergillus; n (%)	<5	12 (14.6)	5 (6.6)	<5	6 (11.8)	<5	45 (9.1)

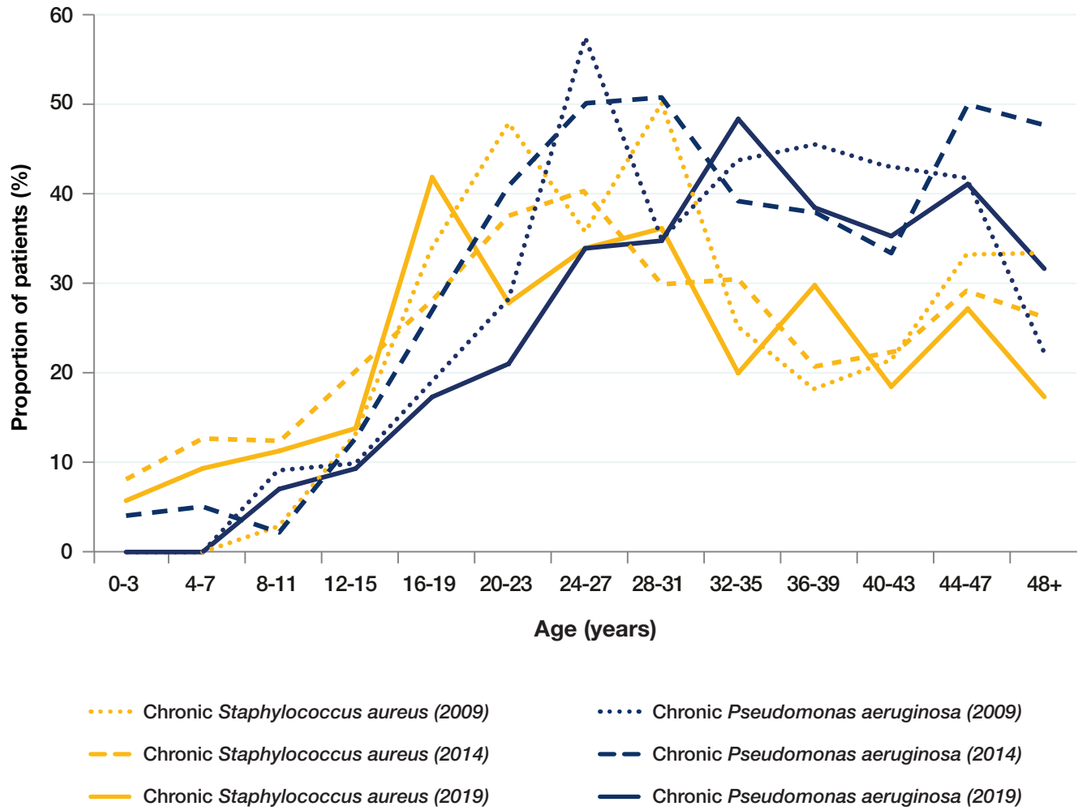
	Adult age range (years)						Overall
	40-43	44-47	48-51	52-55	56-59	60+	≥16 years
Patients in age range; n	37	22	21	23	14	15	532
Patients who had culture taken; n	34	18	19	20	11	14	496
Chronic *S. aureus*; n (%)	7 (20.6)	6 (33.3)	<5	6 (30.0)	<5	<5	153 (30.8)
Intermittent *S. aureus*; n (%)	<5	0 (0.0)	<5	6 (30.0)	5 (45.5)	<5	101 (20.4)
Chronic *P. aeruginosa*; n (%)	13 (38.2)	9 (50.0)	6 (31.6)	<5	7 (63.6)	7 (50.0)	170 (34.3)
Intermittent *P. aeruginosa*; n (%)	<5	<5	<5	<5	<5	0 (0.0)	60 (12.1)
B. cepacia complex; n (%)	<5	<5	<5	<5	0 (0.0)	<5	37 (7.5)
B. cenocepacia; n (%)	<5	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)	7 (1.4)
B. multivorans; n (%)	<5	<5	<5	<5	0 (0.0)	<5	18 (3.6)
Other Burkholderia; n (%)	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)	<5	<5
MRSA; n (%)	0 (0.0)	1 (5.6)	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)	10 (2.0)
H. influenzae; n (%)	7 (20.6)	<5	<5	<5	<5	<5	67 (13.5)
Nontuberculous mycobacterium; n (%)	<5	0 (0.0)	0 (0.0)	<5	<5	0 (0.0)	40 (8.1)
Aspergillus; n (%)	<5	<5	<5	<5	<5	<5	45 (9.1)

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.

	2017 (n=858)	2018 (n=819)	2019 (n=868)
NTM prevalence; n (%)	38 (4.4)	46 (5.6)	53 (6.1)
On NTM treatment in the given year; n (% of NTM prevalence in given year)	11 (29.0)	16 (34.9)	29 (3.3)
NTM incidence1	12 (1.4)	19 (2.5)	23 (2.8)
M. ycobacterium abscessus prevalence (%)	26 (3.0)	28 (3.4)	32 (3.7)

1.18 Lung infections over time
N=524 in 2009, N=782 in 2014, N=868 in 2019



Chronic *Staphylococcus aureus*				
Age (years)	**2009 (%)**	**2014 (%)**	**2019 (%)**	**p-value***
0-3	0.0	8.1	6.7	0.752
4-7	0.0	12.7	10.1	0.617
8-11	3.0	12.4	12.0	0.940
12-15	13.2	20.0	14.4	0.374
16-19	34.0	28.1	41.4	0.095
20-23	47.8	37.5	27.9	0.178
24-27	35.7	40.2	33.8	0.398
28-31	50.0	29.9	35.9	0.440
32-35	25.0	30.4	20.4	0.247
36-39	18.2	20.7	29.8	0.382
40-43	21.4	22.2	18.9	0.774
44-47	33.3	29.2	27.3	0.887
48+	33.3	26.2	17.8	0.287
<16	4.4	12.8	11.3	N/A
16+	36.5	31.5	28.8	N/A
<18	5.8	14.6	13.1	N/A
18+	38.0	31.8	28.5	N/A

Chronic *Pseudomonas aeruginosa*				
Age (years)	**2009 (%)**	**2014 (%)**	**2019 (%)**	**p-value***
0-3	0.0	4.1	0.0	0.115
4-7	0.0	5.1	0.0	0.043
8-11	9.1	2.2	7.0	0.126
12-15	9.9	12.7	9.3	0.506
16-19	19.1	27.0	17.2	0.172
20-23	28.3	40.9	20.9	0.004
24-27	57.1	50.0	33.8	0.038
28-31	35.0	50.7	34.6	0.050
32-35	43.8	39.1	48.1	0.365
36-39	45.5	37.9	38.3	0.974
40-43	42.9	33.3	35.1	0.895
44-47	41.7	50.0	40.9	0.536
48+	22.2	47.6	31.5	0.086
<16	4.7	5.4	4.8	N/A
16+	34.5	41.6	32.0	N/A
<18	5.2	6.9	5.5	N/A
18+	37.4	43.6	33.1	N/A

* The proportion of people with each infection within each age group was compared between 2014 and 2019. If the p-value is less than 0.05 then the difference in the proportions is statistically significant.

Complications

1.19 Complications in 2019

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

	Overall n (%)	<16 years n (%)	≥16 years n (%)
Respiratory related			
Nasal polyps requiring surgery	26 (3.0)	5 (1.5)	21 (3.9)
Sinus disease	-	<5	103 (19.4)
Asthma	-	<5	46 (8.6)
Allergic bronchopulmonary aspergillosis (ABPA)	39 (4.5)	8 (2.4)	31 (5.8)
Any haemoptysis	15 (1.7)	0	15 (2.8)
Massive haemoptysis	<5	0	<5
Pneumothorax requiring chest tube	<5	0	<5
Pancreas and hepatobiliary related			
Raised liver enzymes	61 (7.0)	22 (6.5)	39 (7.3)
Liver disease	133 (15.3)	33 (9.8)	100 (18.8)
Cirrhosis with no portal hypertension	-	<5	7 (1.3)
Cirrhosis with portal hypertension	15 (1.7)	5 (1.5)	10 (1.9)
Gall bladder disease requiring surgery	7 (0.8)	<5	<5
Pancreatitis	-	<5	6 (1.1)
Upper gastrointestinal (GI)			
Gastroesophageal reflux disease (GERD)	-	<5	147 (27.6)
Peptic ulcer	0 (0.0)	0	0
GI bleed (varices as source)	<5	<5	0
GI bleed (non varices as source)	<5	0	<5
Lower GI			
Intestinal obstruction	<5	<5	<5
Distal intestinal obstruction syndrome (DIOS)	87 (10.0)	7 (2.1)	80 (15.0)
Fibrosing colonopathy/colonic stricture	0 (0.0)	0	0
Rectal prolapse	0 (0.0)	0	0
Renal			
Kidney stones	<5	<5	<5
Renal failure	8 (0.9)	0	8 (1.5)
Musculoskeletal			
Arthritis	-	<5	6 (1.1)
Arthropathy	-	<5	38 (7.1)
Bone fracture	<5	0	<5
Osteopenia	89 (10.3)	0	89 (16.7)
Osteoporosis	33 (3.8)	0	33 (6.2)
Other			
Cancer confirmed by histology	0 (0.0)	0	0
Port inserted or replaced	12 (1.4)	6 (1.8)	6 (1.1)
Depression	23 (2.6)	0	23 (4.3)
Hearing loss	-	<5	13 (2.4)
Hypertension	10 (1.2)	0	10 (1.9)

-Redacted to adhere to statistical disclosure guidelines.

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.

	2018			2019		
	Overall (n=858)	<16 years (n=309)	≥16 years (n=520)	Overall (n=868)	<16 years (n=336)	≥16 years (n=532)
Allergic bronchopulmonary aspergillosis; n (%)	10 (1.2)	<5	≥5	17 (2.0)	11 (2.1)	6 (1.8)
Cirrhosis - no portal hypertension; n (%)	9 (1.0)	<5	≥5	2 (0.2)	1 (0.2)	1 (0.3)
Cirrhosis - with portal hypertension; n (%)	6 (0.7)	<5	≥5	6 (0.7)	3 (0.6)	3 (0.9)
Cancer confirmed by histology; n (%)	<5	<5	<5	0	0	0

1.21 Cystic fibrosis-related diabetes (CFRD)

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=672)	10-15 years (n=140)	≥16 years (n=532)
On CFRD treatment; n (%)	135 (20.1)	6 (4.3)	129 (24.2)
Of those on treatment			
Insulin; n (%)	131 (97.0)	6 (100.0)	125 (96.9)
CFRD Screening; n(%)			
Yes	348 (51.8)	118 (84.3)	230 (43.2)
Screening type; n(%)			
Continous glucose monitoring; n (%)	79 (22.7)	17 (14.4)	62 (27.0)
Oral glucose tolerance test; n (%)	261 (75.0)	93 (78.8)	168 (73.0)
Not screened (known CFRD)	144 (21.4)	4 (2.9)	140 (26.3)
Not screened (other)	163 (24.3)	17 (12.1)	146 (27.4)
Unknown	17 (2.5)	1 (0.7)	16 (3.0)

[1] Proportion of patients on treatment
[2] Proportion of patients screened

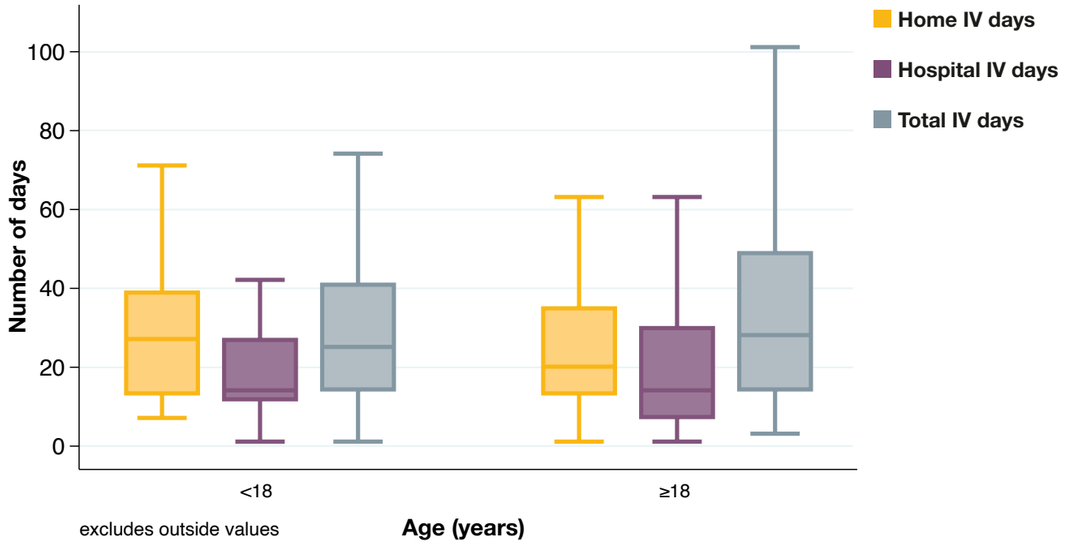
Antibiotics

1.22 Intravenous (IV) antibiotics
N=868

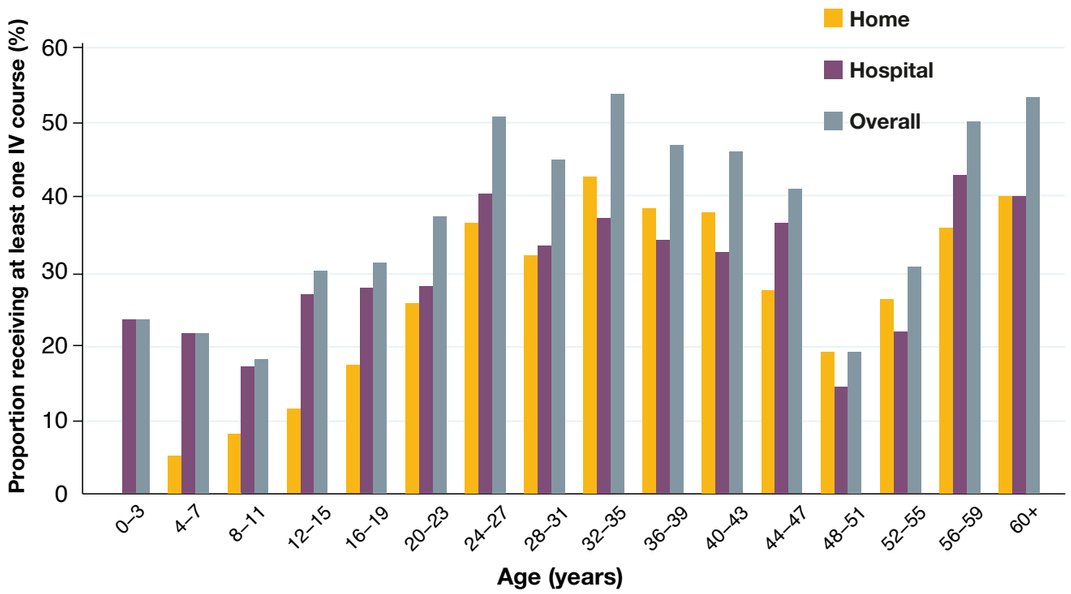
When someone with CF becomes unwell with an infection, they might be prescribed IV antibiotics. IV antibiotics are given to the patient through their veins. This treatment can take a number of days and might take place as a hospital inpatient, or at home.

		Home		Hospital		Total	
Age (years)	N	Patients n (%)	Median days (IQR)	Patients n (%)	Median days (IQR)	Patients n (%)	Median days (IQR)
0-3	60	0 (0.0)	N/A	14 (23.3)	16 (14-29)	14 (23.3)	16 (14-29)
4-7	79	4 (5.1)	37 (21-40)	17 (21.5)	13 (21-40)	17 (21.5)	14 (13-32)
8-11	100	8 (8.0)	27 (15-42)	17 (17.0)	16 (15-42)	18 (18.0)	34 (14-56)
12-15	97	11 (11.3)	28 (12-39)	26 (26.8)	14 (12-39)	29 (29.9)	28 (14-41)
16-19	58	10 (17.2)	24 (22-30)	16 (27.6)	16 (22-30)	18 (31.0)	38 (14-76)
20-23	86	22 (25.6)	29 (14-39)	24 (27.9)	14 (14-39)	32 (37.2)	32 (14-50)
24-27	77	28 (36.4)	19 (14-37)	31 (40.3)	24 (14-37)	39 (50.6)	40 (14-56)
28-31	78	25 (32.1)	18 (13-28)	26 (33.3)	14 (13-28)	35 (44.9)	28 (14-42)
32-35	54	23 (42.6)	17 (11-33)	20 (37.0)	9 (11-33)	29 (53.7)	28 (14-52)
36-39	47	18 (38.3)	18 (12-26)	16 (34.0)	13 (12-26)	22 (46.8)	25 (14-42)
40-43	37	14 (37.8)	37 (14-56)	12 (32.4)	11 (14-56)	17 (45.9)	39 (14-56)
44-47	22	6 (27.3)	15 (13-29)	8 (36.4)	16 (13-29)	9 (40.9)	22 (18-43)
48-51	21	4 (19.0)	49 (28-57)	3 (14.3)	19 (28-57)	4 (19.0)	60 (28-81)
52-55	23	6 (26.1)	6 (6-24)	5 (21.7)	10 (6-24)	7 (30.4)	21 (14-42)
56-59	14	5 (35.7)	14 (11-17)	6 (42.9)	8 (11-17)	7 (50.0)	14 (13-22)
60+	15	6 (40.0)	13 (10-14)	6 (40.0)	23 (10-14)	8 (53.3)	19 (14-43)
<16	336	23 (6.8)	28 (12-39)	74 (22.0)	14 (12-39)	78 (23.2)	24 (14-41)
≥16	532	167 (31.4)	21 (13-35)	173 (32.5)	14 (13-35)	227 (42.7)	28 (14-49)
<18	366	25 (6.8)	27 (13-39)	80 (21.9)	14 (13-39)	85 (23.2)	25 (14-41)
≥18	502	165 (32.9)	20 (13-35)	167 (33.3)	14 (13-35)	220 (43.8)	28 (14-49)
Overall	**868**	**190 (21.9)**	**21 (13-36)**	**247 (28.5)**	**14 (13-36)**	**305 (35.1)**	**28 (14-43)**

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 21.9% and in hospital was 28.5%. The proportion receiving any IVs was 35.1%.



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

	2009			2014			2019		
	Overall	<16 years	≥16 years	Overall	<16 years	≥16 years	Overall	<16 years	≥16 years
Patients with chronic *P. aeruginosa*; n	84	17	67	228	16	212	186	16	170
Tobramycin solution; n (%)	15 (17.9)	1 (5.9)	14 (20.9)	45 (19.7)	3 (18.8)	42 (19.8)	29 (15.6)	4 (25.0)	25 (14.7)
Other aminoglycoside; n (%)	<5	0	<5	7 (3.1)	0	7 (3.3)	<5	0	<5
Colistin; n (%)	37 (44.0)	10 (58.8)	27 (40.3)	80 (35.1)	10 (62.5)	70 (33.0)	32 (17.2)	7 (43.8)	25 (14.7)
Promixin; n (%)	<5	0	<5	30 (13.2)	<5	-	25 (13.4)	<5	-
Aztreonam; n (%)	-	-	-	2 (0.9)	0	2 (0.9)	22 (11.8)	0	22
Colistimethate (inh) inhalation powder; n (%)	-	-	-	20 (8.8)	<5	-	46 (24.7)	<5	-
Tobramycin inhalation powder; n (%)	-	-	-	49 (21.5)	0	49 (23.1)	40 (21.5)	0	40 (23.5)
At least one of the above; n (%)	52 (61.9)	11 (64.7)	41 (61.2)	168 (73.7)	12 (75.0)	156 (73.6)	150 (80.6)	14 (87.5)	136 (80.0)

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.

	Age	Number of patients on azithromycin; n	Patients with chronic *P. aeruginosa*; n (%)	Patients without chronic *P. aeruginosa*; n (%)
2009	Overall	139	51 (36.7)	88 (63.3)
	0-3 years	0	0	0
	4-15 years	-	<5	31 (88.6)
	≥16 years	104	47 (45.2)	57 (54.8)
2014	Overall	404	187 (46.3)	217 (53.7)
	0-3 years	<5	0	<5
	4-15 years	64	9 (14.1)	55 (85.9)
	≥16 years	338	178 (52.7)	160 (47.3)
2019	Overall	454	163 (35.9)	291 (64.1)
	0-3 years	7	0	7 (100.0)
	4-15 years	84	11 (13.1)	73 (86.9)
	≥16 years	363	152 (41.9)	211 (58.1)

1.25 Flucloxacillin

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

Age; years	Patients; n	Patients on prophylactic flucloxacillin; n (%)
0-3	60	39 (65.0)
4-7	79	31 (39.2)
8-11	100	37 (37.0)
12-15	97	32 (33.0)
16-19	58	12 (20.7)
20-23	86	28 (32.6)
24-27	77	13 (16.9)
28-31	78	10 (12.8)
32-35	54	<5
36-39	47	<5
40-43	37	<5
44-47	22	0
48-51	21	<5
52-55	23	<5
56-59	14	<5
60+	15	0
<16	336	139 (41.4)
≥16	532	76 (14.3)
<18	366	148 (40.4)
≥18	502	67 (13.3)
Overall	**868**	**215 (24.8)**

Muco-active therapies

1.26 Mannitol

Age; years	Patients; n	Patients on Mannitol; n (%)
0-3	60	0
4-7	79	0
8-11	100	0
12-15	97	0
16-19	58	<5
20-23	86	0
24-27	77	<5
28-31	78	<5
32-35	54	<5
36-39	47	<5
40-43	37	<5
44-47	22	0
48-51	21	<5
52-55	23	0
56-59	14	0
60+	15	0
<16	336	0
≥16	532	11 (2.1)
<18	366	0
≥18	502	11 (2.2)
Overall	**868**	**11 (1.3)**

1.27 DNase

	2009		2014		2019	
Age; years	Patients; n	Patients on DNase; n (%)	Patients; n	Patients on DNase; n (%)	Patients; n	Patients on DNase; n (%)
0-3	88	3 (3.4)	74	5 (6.8)	60	6 (10.0)
4-7	76	6 (7.9)	79	18 (22.8)	79	17 (21.5)
8-11	66	15 (22.7)	89	31 (34.8)	100	42 (42.0)
12-15	91	36 (39.6)	55	28 (50.9)	97	52 (53.6)
16-19	47	22 (46.8)	89	38 (42.7)	58	31 (53.4)
20-23	46	10 (21.7)	88	35 (39.8)	86	51 (59.3)
24-27	28	8 (28.6)	82	41 (50.0)	77	42 (54.5)
28-31	20	<5	67	21 (31.3)	78	49 (62.8)
32-35	16	<5	46	8 (17.4)	54	24 (44.4)
36-39	11	<5	29	9 (31.0)	47	22 (46.8)
40-43	14	<5	18	<5	37	14 (37.8)
44-47	12	<5	24	9 (37.5)	22	9 (40.9)
48-51	3	0	19	6 (31.6)	21	9 (42.9)
52-55	2	0	7	<5	23	9 (39.1)
56-59	1	<5	7	0	14	6 (42.9)
60+	3	0	9	<5	15	<5
<16	321	60 (18.7)	297	82 (27.6)	336	117 (34.8)
≥16	203	54 (26.6)	485	174 (35.9)	532	270 (50.8)
<18	345	70 (20.3)	335	98 (29.3)	366	134 (36.6)
≥18	179	44 (24.6)	447	158 (35.3)	502	253 (50.4)
Overall	**524**	**114 (21.8)**	**782**	**256 (32.7)**	**868**	**387 (44.6)**

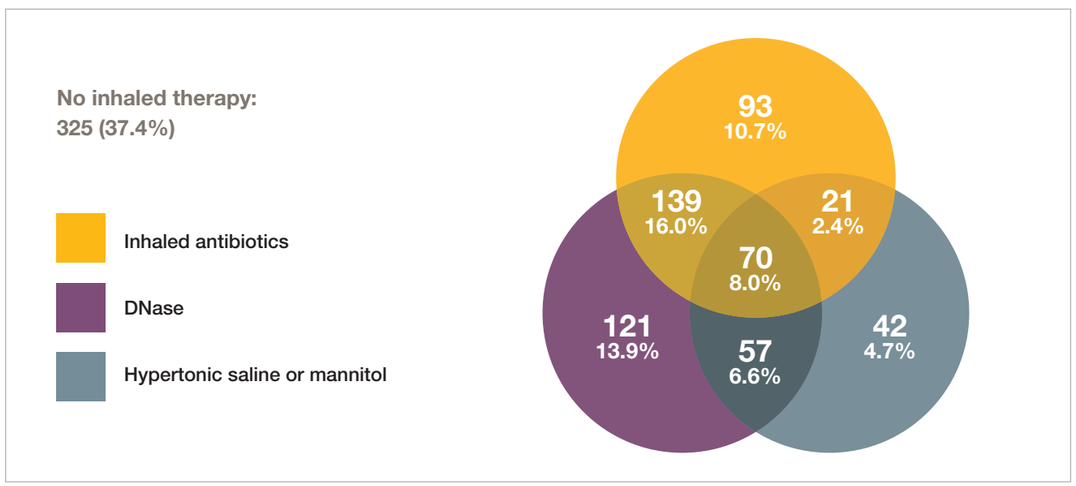
1.28 Hypertonic saline

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

	2009		2014		2019	
Age; years	**Patients; n**	**Patients on hypertonic saline; n (%)**	**Patients; n**	**Patients on hypertonic saline; n (%)**	**Patients; n**	**Patients on hypertonic saline; n (%)**
0-3	88	0	74	<5	60	10 (16.7)
4-7	76	0	79	9 (11.4)	79	14 (17.7)
8-11	66	<5	89	14 (15.7)	100	27 (27.0)
12-15	91	7 (7.7)	55	15 (27.3)	97	38 (39.2)
16-19	47	<5	89	28 (31.5)	58	17 (29.3)
20-23	46	0	88	16 (18.2)	86	18 (20.9)
24-27	28	<5	82	15 (18.3)	77	19 (24.7)
28-31	20	<5	67	14 (20.9)	78	11 (14.1)
32-35	16	0	46	<5	54	10 (18.5)
36-39	11	0	29	<5	47	8 (17.0)
40-43	14	0	18	0	37	<5
44-47	12	0	24	5 (20.8)	22	<5
48-51	<5	0	19	<5	21	<5
52-55	<5	0	7	0	23	<5
56-59	<5	0	7	<5	14	<5
60+	<5	0	9	<5	15	<5
<16	321	8 (2.5)	297	41 (13.8)	336	89 (26.5)
≥16	203	3 (1.5)	485	93 (19.2)	532	94 (17.7)
<18	345	9 (2.6)	335	50 (14.9)	366	98 (26.8)
≥18	179	2 (1.1)	447	84 (18.8)	502	85 (16.9)
Overall	**524**	**11 (2.1)**	**782**	**134 (17.1)**	**868**	**183 (21.1)**

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 325 (37.4%) 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 (at annual review)	Patients; n
Patients on ivacaftor in Scotland	Overall	81
	<6 years	4
	≥6 years	77
Patients stopped ivacaftor ever	Overall	7
	<6 years	0
	≥6 years	7

Tests	Age (at start date)	Median (IQR)	Number with complete data; n (%)
Sweat chloride before ivacaftor	Overall	99mmol/l (94-106)	34 (42.0)
	<6 years	100mmol/l (97-106)	9 (75.0)
	≥6 years	99mmol/l (94-106)	25 (36.2)
Sweat chloride 6-8 weeks after ivacaftor	Overall	52mmol/l (40-67)	22 (27.2)
	<6 years	47mmol/l (40-68)	5 (41.7)
	≥6 years	52mmol/l (44-62)	17 (24.6)
FEV_1% before ivacaftor	Overall	63.5 (47.7-79.9)	57 (70.4)
	<6 years	85.5 (82.5-92.0)	4 (33.3)
	≥6 years	61.4 (47.5-77.2)	53 (76.8)
FEV_1% 6-8 weeks after ivacaftor	Overall	70.3 (51.7-88.4)	53 (65.4)
	<6 years	94.8 (81.4-99.3)	3 (25.0)
	≥6 years	66.4 (50.3-81.6)	50 (72.5)

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.

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, 26 people received this drug during 2019.

	Age (at annual review)	Patients; n
Patients on lumacaftor/ivacaftor in Scotland	Overall	-
	<6 years	<5
	≥6 years	25
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.

A total of 45 people with cystic fibrosis in Scotland are recorded as being prescribed tezacaftor/ivacaftor in 2019 and no one has stopped taking it.

1.31 Oxygen and non-invasive ventilation

	Overall (N=868)	<16 years (n=336)	≥16 years (n=532)	<18 years (n=366)	≥18 years (n=502)
Non-invasive ventilation (NIV); n (%)	9 (1.0)	0 (0.0)	9 (1.7)	0 (0.0)	9 (1.8)
Long-term oxygen; n (%)	45 (5.2)	<5	-	<5	-
Among those who have long-term oxygen:					
Continuously	21 (46.7)	0 (0.0)	21 (47.7)	0 (0.0)	21 (47.7)
Nocturnal or with exertion	9 (20.0)	0 (0.0)	9 (20.5)	0 (0.0)	9 (20.5)
As required (PRN)	6 (13.3)	0 (0.0)	6 (13.6)	0 (0.0)	6 (13.6)
With exacerbation	9 (20.0)	<5	8 (18.2)	<5	-

1.32 Physiotherapy

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

	Overall (N=868)	<16 years (n=336)	≥16 years (n=532)	<18 years (n=366)	≥18 years (n=502)
Active cycle of breathing techniques; n (%)	91 (10.5)	8 (2.4)	83 (15.6)	11 (3.0)	80 (15.9)
Autogenic drainage (including assisted autogenic drainage); n (%)	390 (44.9)	70 (20.8)	320 (60.2)	81 (22.1)	309 (61.6)
Postural drainage; n (%)	<5	0 (0.0)	<5	0 (0.0)	<5
Any form of positive expiratory pressure (PEP); n (%)	491 (56.6)	309 (92.0)	182 (34.2)	331 (90.4)	160 (31.9)
VEST; n (%)	<5	<5	<5	<5	<5
Exercise; n (%)	453 (52.2)	167 (49.7)	286 (53.8)	188 (51.4)	265 (52.8)
Other; n (%)	176 (20.3)	120 (35.7)	56 (10.5)	123 (33.6)	53 (10.6)

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=868)	<16 years (n=336)	≥16 years (n=532)	<18 years (n=366)	≥18 years (n=502)
Any supplemental feeding; n (%)	219 (25.2)	87 (25.9)	132 (24.8)	92 (25.1)	127 (25.3)
Nasogastric tube; n (%)	10 (1.2)	<5	-	<5	-
Gastrostomy tube/button; n (%)	32 (3.7)	14 (4.2)	18 (3.4)	14 (3.8)	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 (%)	<5	0 (0.0)	<5	0 (0.0)	<5

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.

	2014	2015	2016	2017	2018	2019
Patients evaluated; n	17	19	18	22	19	21
Patients accepted; n	12	16	8	17	7	10
Patients receiving transplants; n	6	<5	<5	<5	<5	5
Bilateral lung	6	<5	<5	<5	<5	<5
Liver	0	0	0	0	<5	<5
Other	0	0	<5	<5	<5	0

The graph below shows the total number of bilateral lung transplants over time in patients aged 16 and over.

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 mutation recorded	861 (99.2)
Patients genotyped with both mutations recorded	853 (98.3)
F508del mutations	
Homozygous F508del	367 (42.3)
Heterozygous F508del	418 (48.2)

1.35 Mutation combinations in Scotland

This tabulation shows the proportion (%) of patients with the most common mutation combinations. For example, 8.6% 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	42.3							**42.3**
R117H	6.6	0.1						**6.7**
G551D	8.6	0.1	0.2					**9.0**
G542X	5.0	0.2	0.1	0.1				**5.4**
621+1G->T	0.7	0.0	0.1	0.0	0.0			**0.8**
Other	26.4	0.9	1.6	0.7	0.3	4.1		**34.1**
Unknown	0.9	0.0	0.0	0.0	0.0	0.0	0.8	**1.7**
Total	**90.4**	**1.4**	**2.1**	**0.8**	**0.3**	**4.1**	**0.8**	**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 2.

Nucleotide	Protein	Legacy name	N	%
c.1521_1523delCTT	p.Phe508del	F508del	785	90.4
c.1652G->A	p.Gly551Asp	G551D	94	10.8
c.350G->A	p.Arg117His	R117H	69	7.9
c.1624G->T	p.Gly542X	G542X	53	6.1
c.200C->T	p.Pro67Leu	P67L	46	5.3
c.1679G->C	p.Arg560Thr	R560T	16	1.8
c.1585-1G->A		1717-1G->A	16	1.8
c.1477C->T	p.Gln493X	Q493X	15	1.7
c.3454G->C	p.Asp1152His	D1152H	15	1.7
c.3909C->G	p.Asn1303Lys	N1303K	13	1.5
c.2657+5G->A		2789+5G->A	13	1.5
c.489+1G->T		621+1G->T	10	1.2
c.1558G->T	p.Val520Phe	V520F	8	0.9
c.3528delC	p.Lys1177SerfsX15	3659delC	8	0.9
c.1364C->A	p.Ala455Glu	A455E	6	0.7
c.948delT	p.Phe316LeufsX12	1078delT	6	0.7
c.1766+1G->A		1898+1G->A	5	0.6
c.178G->T	p.Glu60X	E60X	5	0.6
c.2657+2_2657+3insA		2789+2insA	<5	-
c.3846G->A	p.Trp1282X	W1282X	<5	-

Section 2 Centre-level analysis

Cystic fibrosis care in Scotland is led by eight regional centres, two stand-alone clinics and three networked clinics. The breakdown 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 1.

Key

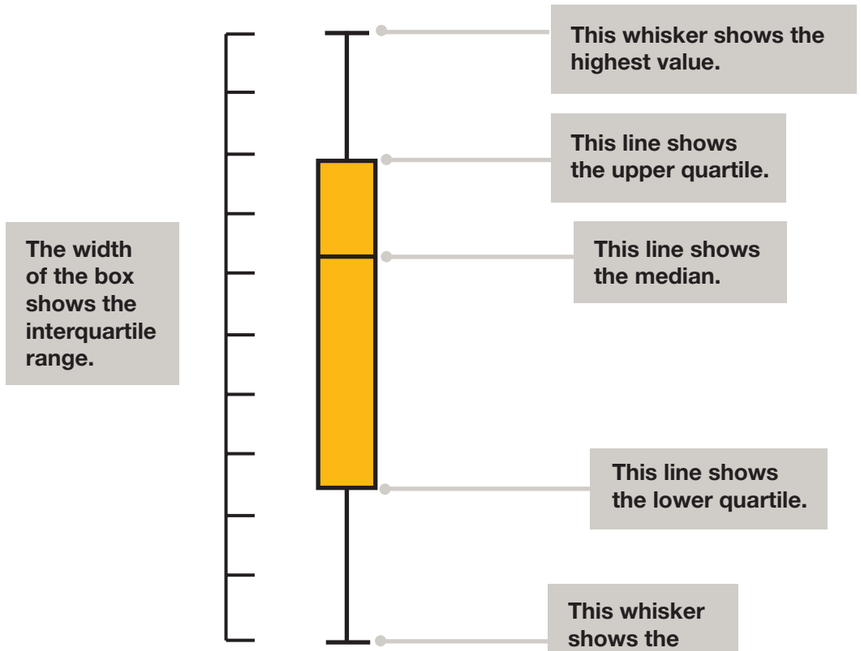
Paediatric centre

Adult centre

A guide to the charts

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

Box plots



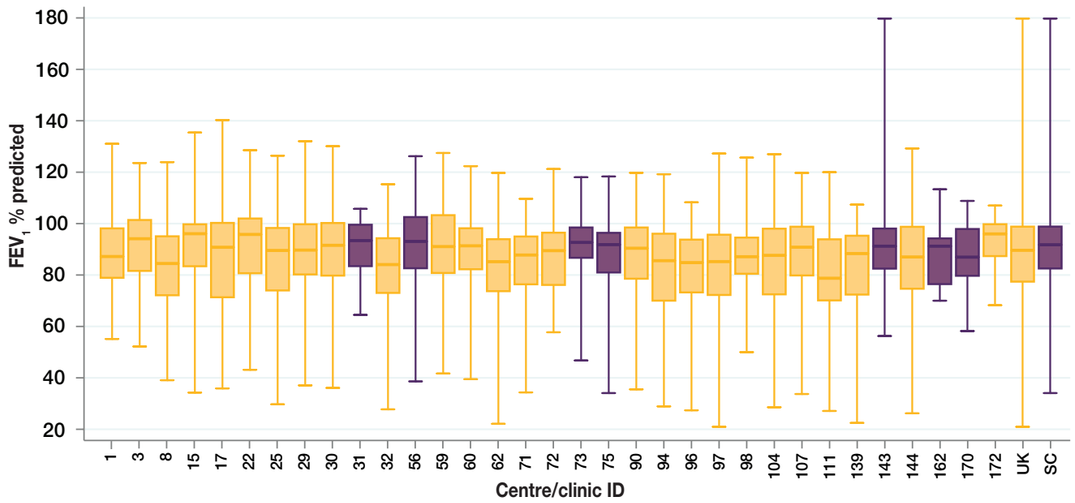
- The 'box' shows the middle half of the data for that centre, going from the first quartile to the third quartile. The longer the box, the more varied the data for that centre.
- The horizontal line within the box shows the median result for that centre.
- The 'whiskers' above and below the box show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

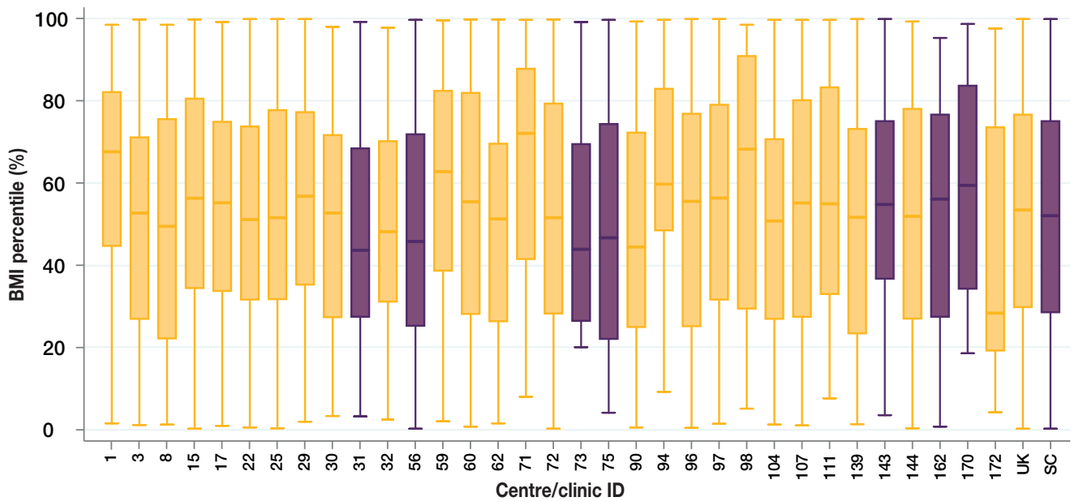
Key ● Services in the UK ● Services in Scotland

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



The median FEV_1% predicted of patients attending paediatric centres/clinics in Scotland is 92.0% predicted (IQR: 85.0 – 99.1).

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

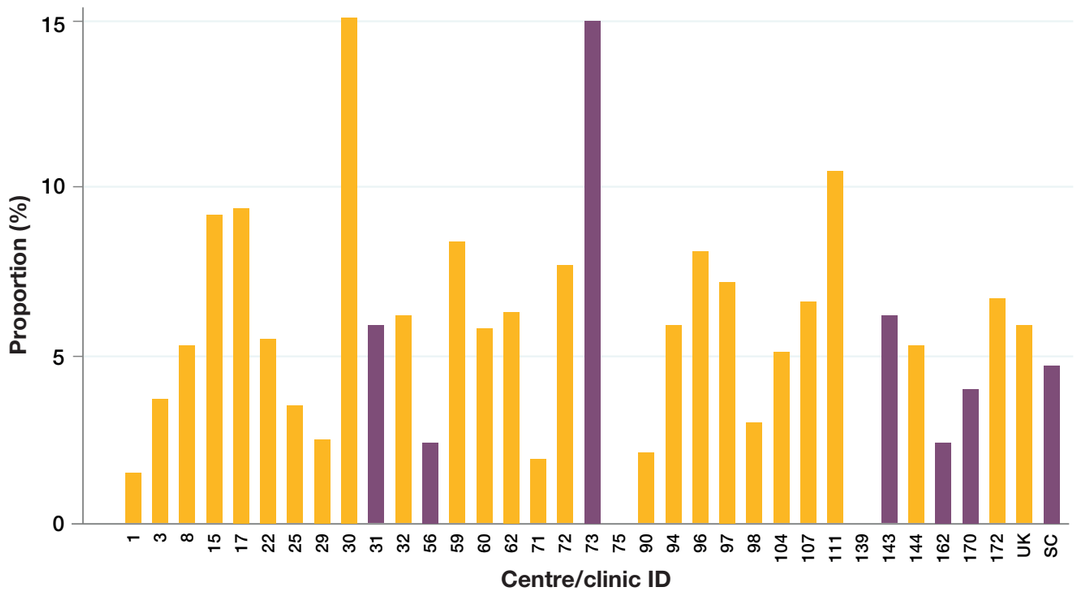


The median BMI percentile of patients attending paediatric centres/clinics in Scotland is 51.3 (IQR: 29.0-74.2).

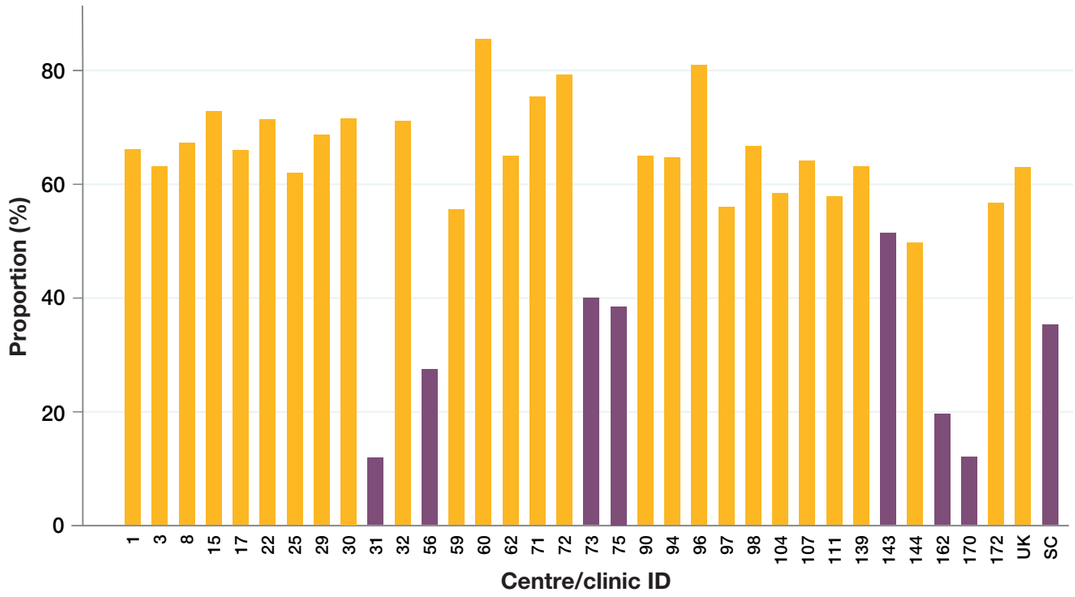
2.3 Data completeness by paediatric centre/clinic

Due to the COVID-19 pandemic and prioritisation of front-line services, some sites were unable to complete data cleaning. As a result, the data completeness section has been omitted this year.

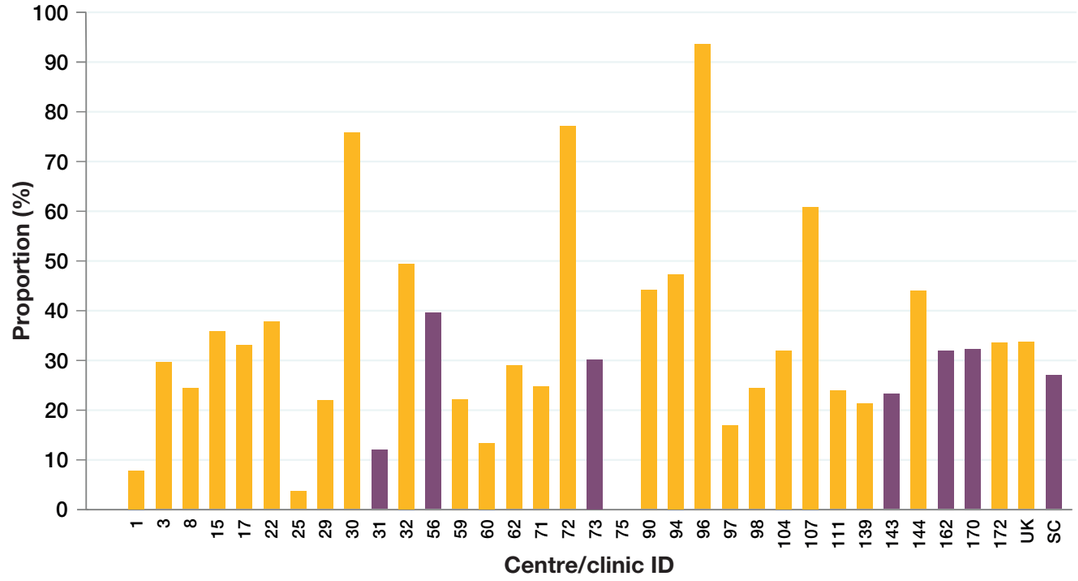
2.4 Proportion of patients with chronic *P. aeruginosa* by paediatric centre/clinic



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



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

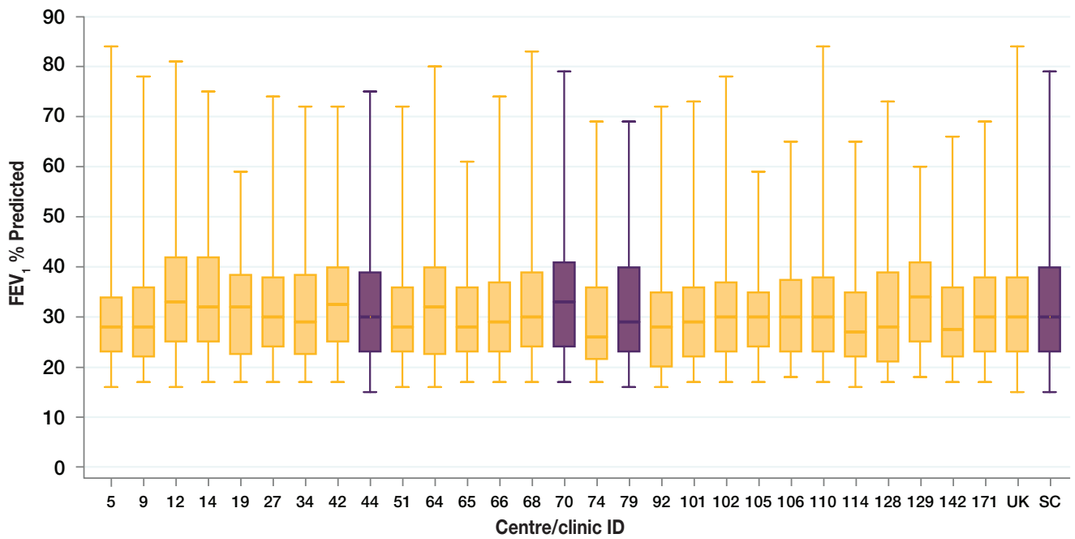


Section 2b: Adult centre analysis

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

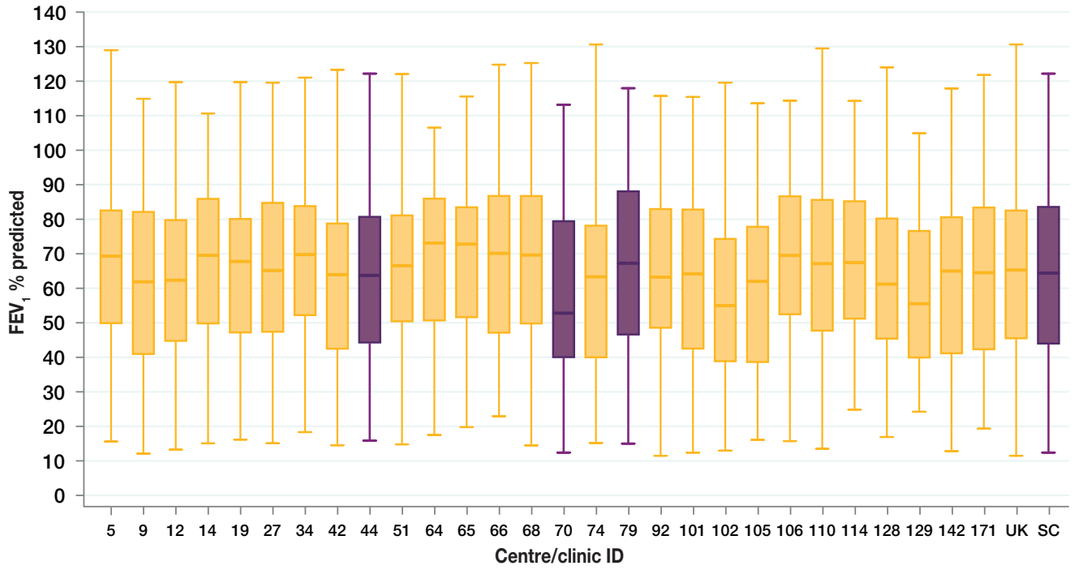
Key ● Services in the UK ● Services in Scotland

2.7 Age distribution by adult centre/clinic



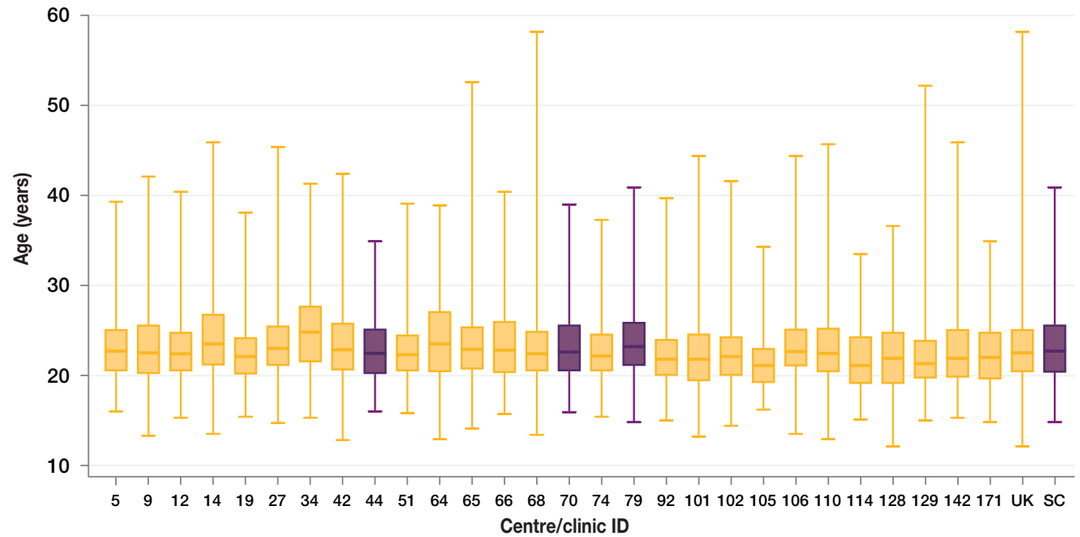
The median age of patients attending adult services in Scotland is 28 years (IQR: 22-37).

2.8 FEV_1 % predicted (GLI equations) by adult centre/clinic (without a history of lung transplant)



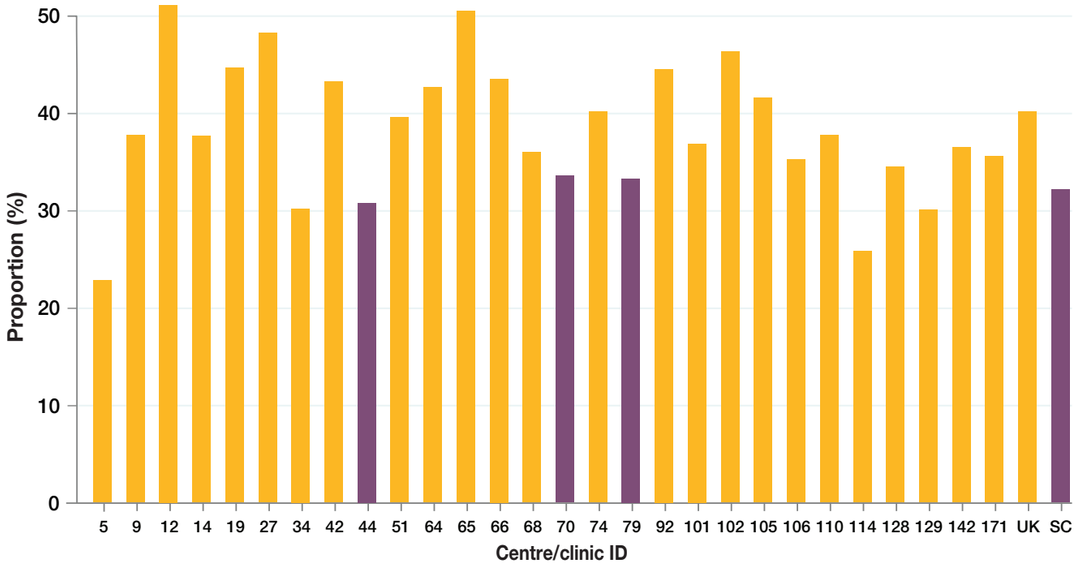
The median FEV_1 % predicted of patients attending adult services in Scotland is 62.7% (IQR 45.0-82.6).

2.9 Body Mass Index (BMI) distribution among patients aged 16 years and older



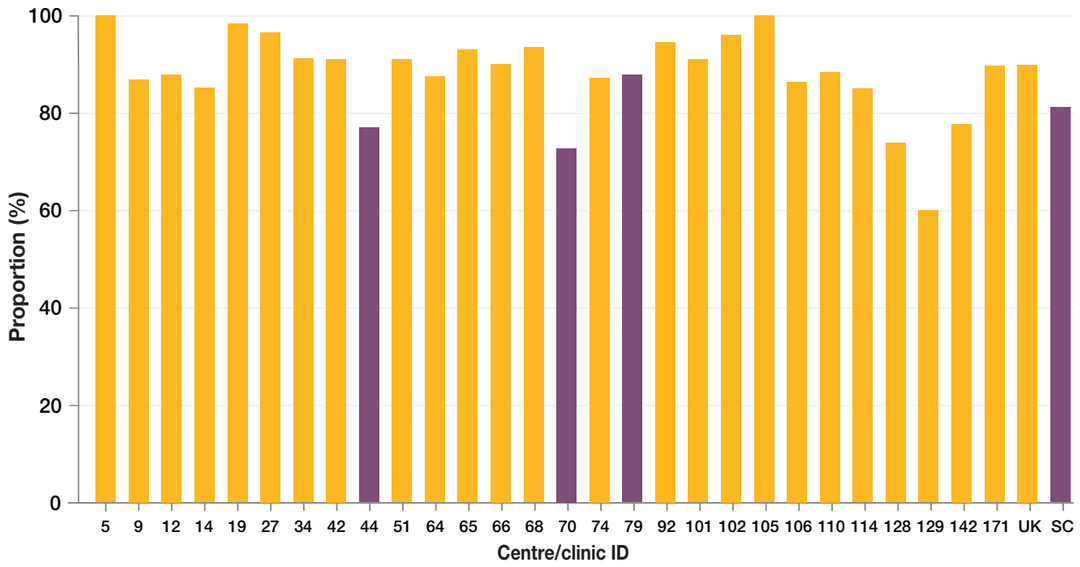
The median BMI of patients attending adult services in Scotland is 22.9 (IQR: 20.6-25.5).

2.10 Proportion of patients with chronic *Pseudomonas aeruginosa* by adult



The proportion of patients with chronic *P.aeruginosa* attending adult services in Scotland is 32%.

2.11 Inhaled antibiotic use for patients with chronic *Pseudomonas aeruginosa* by centre/clinic

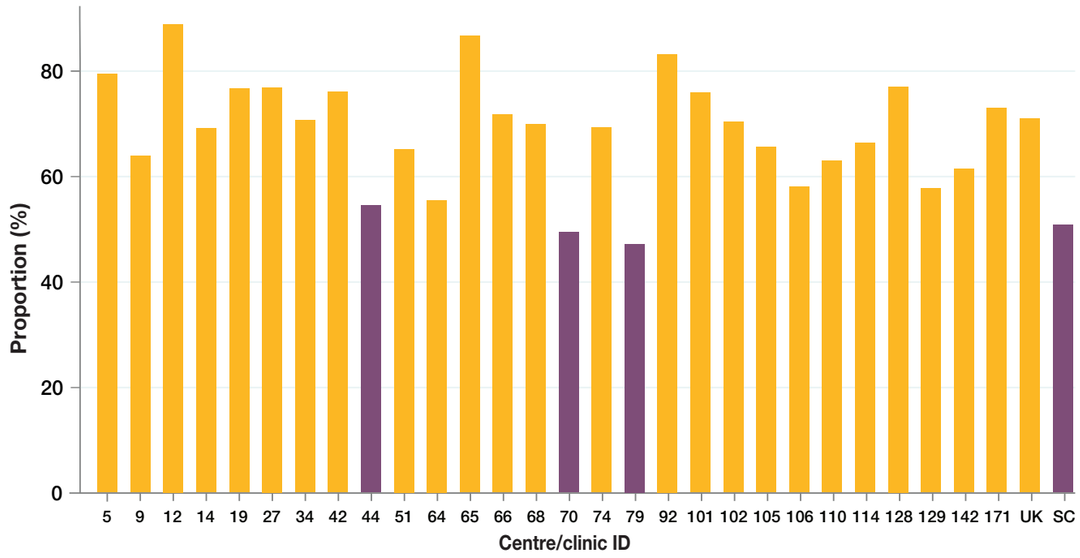


The proportion of chronic P. aeruginosa patients on inhaled antibiotics in Scotland is 81%.

2.12 Data completeness by adult centre/clinic

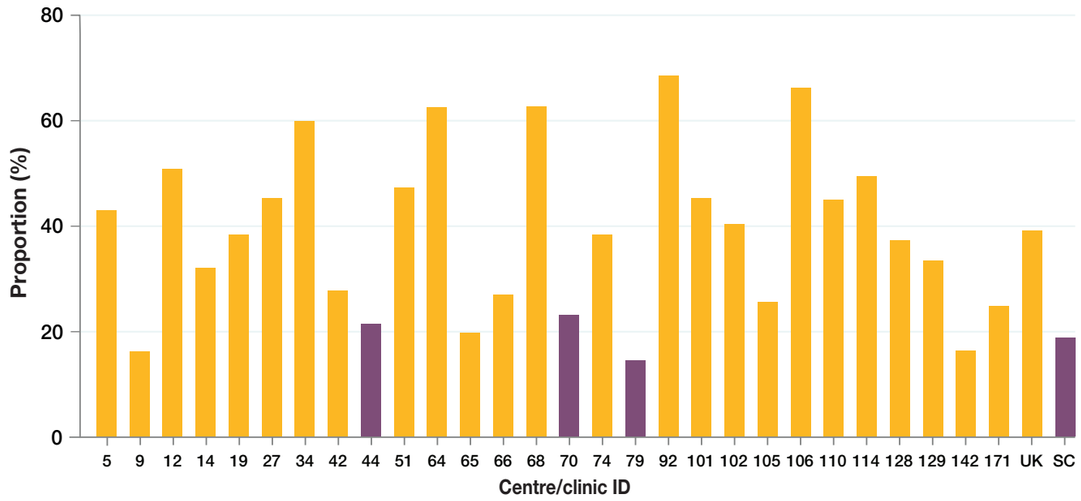
Due to the COVID-19 pandemic and prioritisation of front-line services, some sites were unable to complete data cleaning. As a result, the data completeness section has been omitted this year.

2.13 Proportion of patients receiving DNase treatment by adult centre/clinic



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

2.14 Proportion of patients receiving hypertonic saline or mannitol treatment by adult centre/clinic



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

Glossary

Word/Phrase	Meaning
2019	1 January 2019 – 31 December 2019
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	*H. 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.
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 2019 the median age of the 114 people who died (in UK) was 31.
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 1400 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	*S. 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 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 2019 – ordered alphabetically by location

					Age (years)	
Location	**Name**	**Clinic ID**	**Total Active**	**Number with annual review**	**Mean**	**Median**
Scotland						
Aberdeen	Royal Aberdeen Children's Hospital	75	33	26	7.6	7.5
Ayr	University Hospital Crosshouse	170	26	25	8.7	8.2
Dundee	Ninewells Hospital	73	22	20	8.1	8.5
Edinburgh	Royal Hospital for Sick Children	143	141	130	9.4	9.2
Glasgow	Royal Hospital for Sick Children	56	102	84	8.7	9.5
Inverness	Raigmore Hospital	31	17	17	8.7	10.0
Lanarkshire	Wishaw General Hospital	162	43	41	9.0	9.2

FEV_1% predicted at annual review				Best FEV_1% predicted			
Number	**Mean - unadjusted**	**Mean – adjusted †**	**Median**	**Number**	**Mean - unadjusted**	**Mean - adjusted †**	**Median**
15	89.4	89.0	92.0	16	93.0	92.9	92.8
15	87.9	88.8	87.2	15	95.8	96.9	93.5
13	92.6	91.2	92.9	14	96.1	94.6	99.7
97	91.1	91.5	91.5	97	95.3	95.7	95.0
59	92.6	92.7	93.3	59	96.2	96.3	97.3
9	90.7	91.7	93.6	9	94.5	95.8	93.6
22	88.4	89.2	91.5	30	97.4	97.4	98.1

Adult centres/clinics providing data in 2019 – ordered alphabetically by location

					Age (years)	
Location	**Name**	**Clinic ID**	**Total Active**	**Number with annual review**	**Mean**	**Median**
Scotland						
Aberdeen	Royal Aberdeen Children's Hospital	70	65	65	34.1	33.0
Edinburgh	Royal Hospital for Sick Children	44	248	239	33.0	30.8
Glasgow	Royal Hospital for Sick Children	79	234	221	33.0	29.3

FEV_1% predicted at annual review				Best FEV_1% predicted			
Number	**Mean - unadjusted**	**Mean – adjusted †**	**Median**	**Number**	**Mean - unadjusted**	**Mean - adjusted †**	**Median**
56	59.6	60.2	52.2	56	65.2	65.8	58.1
217	63.1	63.3	63.6	217	68.6	68.6	69.8
214	66.5	66.7	66.7	217	70.1	70.2	72.4

* where 'best' values were missing, or lower than FEV_1% predicted taken at annual review, the annual review value was used.
† 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 2019 – ordered alphabetically by location

			BMI percentile			
Location	**Name**	**Clinic ID**	**Number**	**Mean - unadjusted**	**Mean - adjusted †**	**Median**
Scotland						
Aberdeen	Royal Aberdeen Children's Hospital	75	21	48.7	48.5	45.7
Ayr	University Hospital Crosshouse	170	25	59.2	58.9	59.4
Dundee	Ninewells Hospital	73	19	48.6	48.1	43.8
Edinburgh	Royal Hospital for Sick Children	143	123	56.2	56.5	56.0
Glasgow	Royal Hospital for Sick Children	56	75	48.0	48.1	44.1
Inverness	Raigmore Hospital	31	16	48.8	48.6	43.6
Lanarkshire	Wishaw General Hospital	162	39	52.7	52.7	53.2

Chronic *Pseudomonas*		Having at least 1 IV day		Receiving DNase treatment		Receiving hypertonic saline/mannitol treatment		Inhaled antibiotic use among patients with chronic *Pseudomonas*	
Number	**Proportion (%)**	**Number**	**Proportion (%)**	**Number**	**Proportion (%)**	**Number**	**Proportion (%)**	**Number**	**Proportion (%)**
0	0.0	5	19.2	10	38.5	0	0.0	0	0.0
<5	4.0	12	48.0	<5	12.0	8	32.0	<5	100.0
<5	15.0	<5	20.0	8	40.0	6	30.0	<5	66.7
8	6.2	25	19.2	67	51.5	30	23.1	7	87.5
<5	2.4	27	32.1	23	27.4	33	39.3	<5	100.0
<5	5.9	<5	11.8	<5	11.8	<5	11.8	0	0.0
<5	2.4	7	17.1	8	19.5	13	31.7	<5	100.0

Adult centres/clinics providing data in 2019 – ordered alphabetically by location

			BMI; kg/m^2			
Location	**Name**	**Clinic ID**	**Number**	**Mean - unadjusted**	**Mean - adjusted †**	**Median**
Scotland						
Aberdeen	Aberdeen Royal Infirmary	70	65	23.6	23.4	22.6
Edinburgh	Western General Hospital	44	236	23.0	22.9	22.5
Glasgow	Gartnavel General Hospital	79	221	23.8	23.8	23.2

Chronic *Pseudomonas*		Having at least 1 IV day		Receiving DNase treatment		Receiving hypertonic saline/mannitol treatment		Inhaled antibiotic use among patients with chronic *Pseudomonas*	
Number	**Proportion (%)**	**Number**	**Proportion (%)**	**Number**	**Proportion (%)**	**Number**	**Proportion (%)**	**Number**	**Proportion (%)**
22	33.8	28	43.1	32	49.2	15	23.1	16	72.7
74	31.0	99	41.4	130	54.4	51	21.3	57	77.0
74	33.5	96	43.4	104	47.1	32	14.5	65	87.8

† 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: 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	-
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_2890delACATTCT-GTTCTTC AAGCACCTATGT-CAACCC	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			**67**	**8.2**

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