Cystic Fibrosis Trust

UK Cystic Fibrosis Registry

Annual Data Report 2021 — Scotland

December 2022

Uniting for a life unlimited

UK Cystic Fibrosis Registry
Annual Data Report 2021 — Scotland

Report prepared by

Ciarán Haugh, Medical Statistician, Cystic Fibrosis Trust
Susan Charman, Senior Statistician, Cystic Fibrosis Trust

With assistance from

Siâron Hughes, Freelance Graphic Designer, su-ma.com
Elaine Gunn, Registry Data Manager, Cystic Fibrosis Trust
Sarah Clarke, Associate Director of Data and Quality Improvement, Cystic Fibrosis Trust
The UK CF Registry Steering Committee

Acknowledgements

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

Contact information

For more information about this report, or the UK Cystic Fibrosis Registry, please contact us: **registry@cysticfibrosis.org.uk @CFTrust**

Cystic Fibrosis Trust

Contents

Executive summary

The 2021 UK CF Scottish Registry report once again demonstrates the incredible commitment of CF teams in Scotland to deliver this rich dataset to help support the care of people with CF. The most obvious challenge to the data in 2021 was the continuing COVID-19 pandemic. Although 2021 saw a move back towards pre-pandemic care, many CF teams in Scotland continued to deliver appointments remotely.

Despite these circumstances, the CF Registry data has held up remarkably well and I hope you find the information in this report both interesting and stimulating. Here are some key highlights:

- 777 people had an annual review (the basis of this report). This represents 84% of 915 people eligible for annual review. As this is a reduction on the 89% achieved in 2020, a point of focus for clinicals teams should be ensuring an annual review for all people with CF as the impact of the pandemic wanes.
- Most people with CF in Scotland are over 16 years of age (58.2%). We anticipate this number will continue to increase with improvements in care and the impact of CFTR modulator therapies across all age groups.
- There were 10 new diagnoses of CF in 2021, with nine identified by new-born screening, supporting the case for new-born screening to be more widely available to enable earlier diagnosis.
- In this age of genetic-based therapies, it is promising that 99.5% of people have both CF mutations identified in the Registry. Those currently without access to suitable CFTR modulator therapies, need to be quickly and accurately identifiable so that potential novel therapies can be offered to them without delay.
- There were fewer respiratory cultures made in 2021 compared with previous years. This reduction is mostly in those over 16 years of age, where there was a reduction in both sputum, cough and bronchoalveolar lavage sampling in 2021 compared with previous years. Whether this was a reduction in sample requests due to remote clinic visits, or a reduction in sample acquisition due to the impact of CFTR modulators on sputum production is not yet clear.
- There was a reduction in how frequently key respiratory pathogens were identified in 2021 compared with previous years, including non-tuberculous mycobacteria and chronic *Pseudomonas aeruginosa* infection. The pandemic or the decline in sampling frequency (as noted above) could be factors.
- As expected, triple modulator therapies continued to be rolled out rapidly in 2021, which is thanks to the hard work and dedication of CF clinical teams.

Despite many challenges in 2021, both for clinical teams and people with CF, the 2021 UK CF Scottish Registry report provides a wealth of stimulating data. If you are a person with CF or a family member, thank you so much for consenting that your data can deliver this detailed report. If you are a member of a clinical or data team, thank you for continuing to input data through truly extenuating circumstances – your support is truly valued. I hope that everyone reading this report finds something of interest and can identify different things to take forward and help improve the lives and care of people with CF.

With very best wishes,

Prof. Steve Cunningham
Professor of Paediatric Respiratory Medicine
Chair of the UK CF Registry Research Committee
University of Edinburgh

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 52.

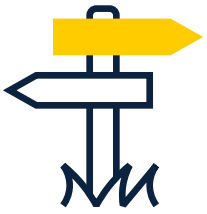
Cystic fibrosis

Cystic fibrosis is an inherited disease caused by a faulty version of a 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, and the protein it makes, 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 also develop other problems, such as liver disease or CF-related diabetes (CFRD). Around 85% of people with CF also have difficulty digesting food.

UK Cystic Fibrosis Registry

The UK CF Registry has been sponsored and hosted by the Cystic Fibrosis Trust since 2007. It is a database of consenting people with CF in the UK. The Registry collects demographic, treatment and health outcomes data. You can find a full list of the data items we collect at **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 to treat 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 2021 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 **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

	2021	
	UK	Scotland
CF patients registered[1]	10908	925
Excluding diagnoses that year	10720	915
CF patients with an annual review; n(%)[2]	10175 (93.3)	777 (84%)
Age in years; median[3]	21	21
All newly diagnosed patients (newborn screening and other)[4]	188	10
Number of patients born identified by newborn screening[4]	134	9
Age at diagnosis in months; median[3]	2	2
Adults aged 16 years and over; %[3]	61.9	58.2
Males; %[3]	53.2	52.4
Genotyped; %[3] (both mutations identified)	99.1	99.5
Total deaths reported (%)[5]	66 (0.6%)	9 (1%)
Age at death in years; median (95% CI)[5]	38 (36, 42)	35 (25, 43)

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.

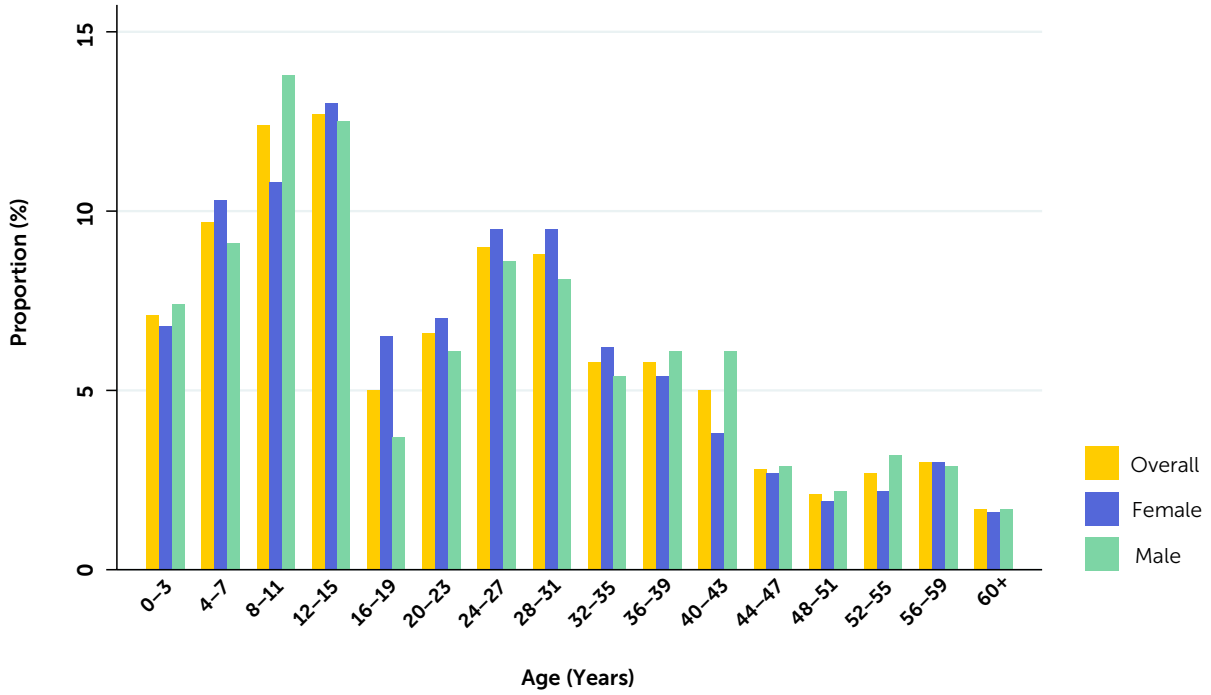
3 Calculated from patients with an annual review in the given year (see footnote 2 above).

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

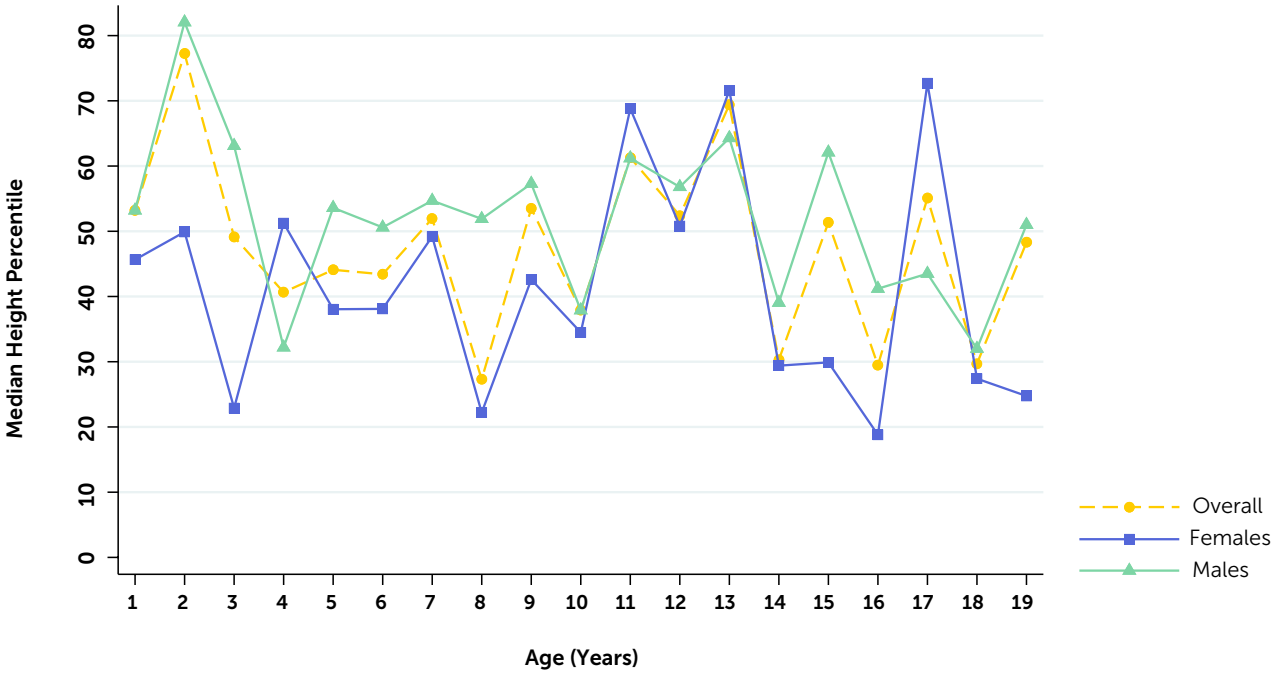
The following chart shows the mix of ages by sex in the CF population in Scotland.



Age	All; n (%)	Females; n (%)	Males; n (%)
0-3	55 (7.1)	25 (6.8)	30 (7.4)
4-7	75 (9.7)	38 (10.3)	37 (9.1)
8-11	96 (12.4)	40 (10.8)	56 (13.8)
12-15	99 (12.7)	48 (13.0)	51 (12.5)
16-19	39 (5.0)	24 (6.5)	15 (3.7)
20-23	51 (6.6)	26 (7.0)	25 (6.1)
24-27	70 (9.0)	35 (9.5)	35 (8.6)
28-31	68 (8.8)	35 (9.5)	33 (8.1)
32-35	45 (5.8)	23 (6.2)	22 (5.4)
36-39	45 (5.8)	20 (5.4)	25 (6.1)
40-43	39 (5.0)	14 (3.8)	25 (6.1)
44-47	22 (2.8)	10 (2.7)	12 (2.9)
48-51	16 (2.1)	7 (1.9)	9 (2.2)
52-55	21 (2.7)	8 (2.2)	13 (3.2)
56-59	23 (3.0)	11 (3.0)	12 (2.9)
60+	13 (1.7)	6 (1.6)	7 (1.7)
<16	325 (41.8)	151 (40.8)	174 (42.8)
≥16	452 (58.2)	219 (59.2)	233 (57.2)
<18	345 (44.4)	164 (44.3)	181 (44.5)
≥18	432 (55.6)	206 (55.7)	226 (55.5)
Overall	777	370	407

1.3 Height percentiles of children and young people (<20 years)[1] N=364

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	8	53.2	33.1-62.2	<5	-*	-*	<5	-*	-*
2	14	77.3	19.6-92.2	<5	-*	-*	10	82.1	19.6-93.0
3	14	49.2	12.3-86.1	5	22.9	6.3-59.2	9	63.1	33.6-88.9
4	20	40.7	10.4-75.9	8	51.3	24.7-69.8	12	32.2	8.0-81.4
5	19	44.1	21.4-70.3	10	38	18.3-54.8	9	53.6	23.3-82.4
6	15	43.4	20.7-75.6	10	38.1	20.7-44.9	5	50.6	26.4-79.1
7	14	52	35.1-73.0	7	49.2	35.9-73.0	7	54.7	16.7-75.6
8	15	27.3	19.3-53.2	6	22.2	13.1-27.3	9	51.9	22.4-55.2
9	24	53.5	25.3-73.4	9	42.6	30.7-67.7	15	57.3	18.1-78.4
10	29	37.9	21.3-56.0	16	34.5	19.8-74.6	13	37.9	24.3-53.1
11	23	61.3	53.3-74.6	8	68.8	33.8-77.3	15	61.2	53.3-72.1
12	21	52.4	31.3-74.7	13	50.8	31.3-69.5	8	56.8	38.3-77.1
13	19	69.4	33.4-80.8	9	71.6	42.7-79.0	10	64.3	33.4-82.0
14	22	30.3	16.2-70.8	10	29.4	10.2-70.8	12	39	23.9-70.7
15	34	51.3	25.4-74.7	16	29.9	22.4-54.8	18	62.1	33.4-79.3
16	10	29.4	5.8-75.3	7	18.8	4.0-75.3	<5	-*	-*
17	8	55.1	20.6-88.1	<5	-*	-*	<5	-*	-*
18	8	29.7	18.2-58.4	5	27.4	13.8-62.5	<5	-*	-*
19	11	48.3	22.1-59.0	6	24.8	14.3-52.4	5	51	48.3-61.4
Overall	**328****	**49.8**	**22.3-73.9**	**157**	**42.4**	**20.5-70.8**	**171**	**53.3**	**26.4-76.3**

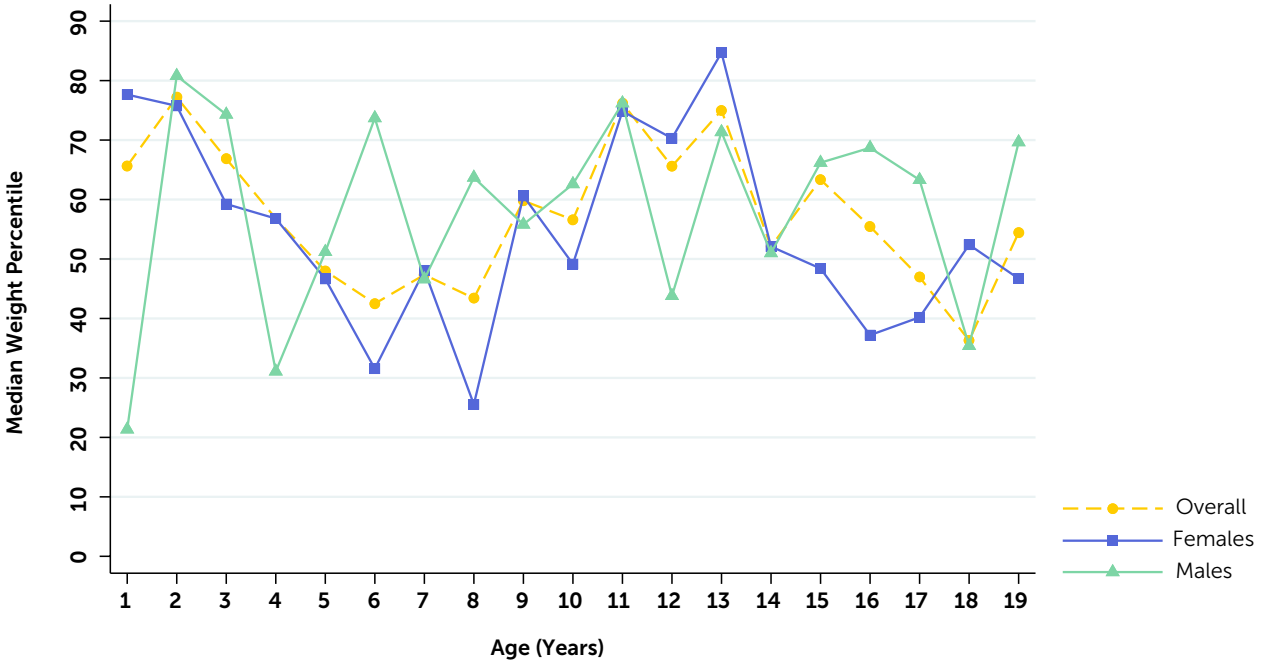
1 Based on UK-WHO growth charts, 1990 (updated 1996)

* Redacted to adhere to statistical disclosure guidelines

** number with non-missing data

1.4 Weight percentiles of children and young people (<20 years)[1] N=364

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.

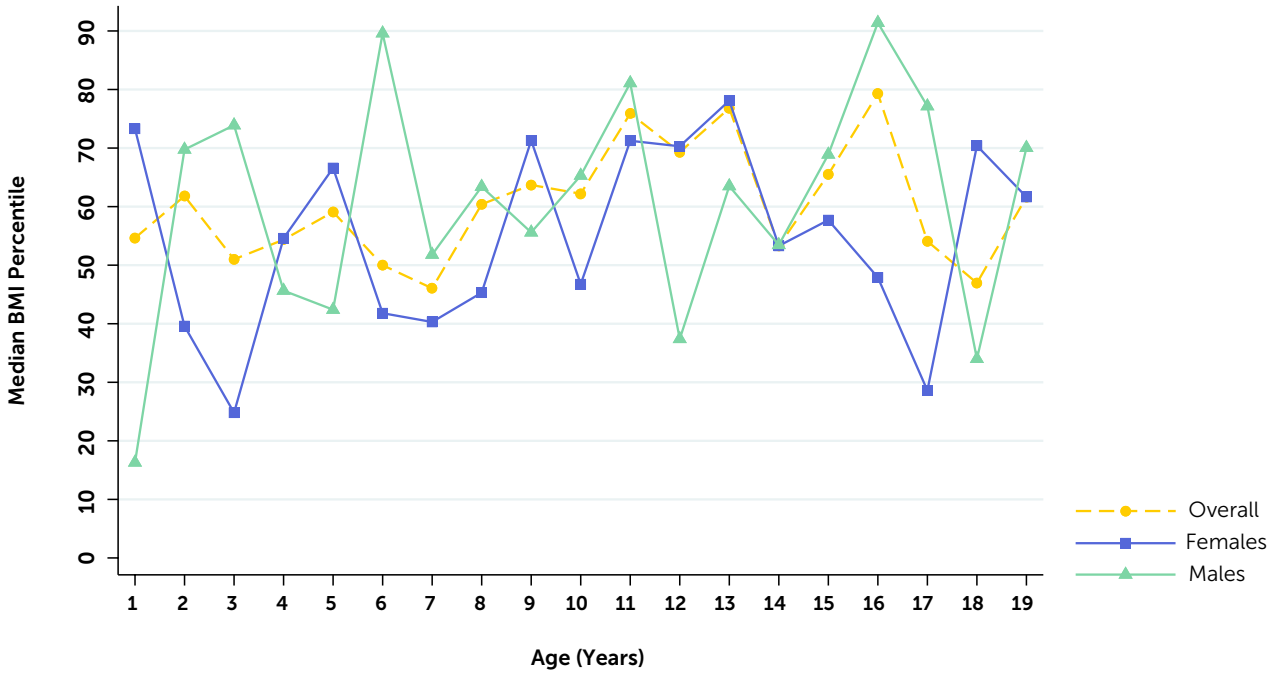


	Overall			Female			Male		
Age	n	Median	IQR	n	Median	IQR	n	Median	IQR
1	10	65.6	27.5-80.1	6	77.6	59.6-81.4	<5	-*	-*
2	17	77.2	40.7-89.6	6	75.8	28.1-89.6	11	80.8	40.7-91.3
3	16	66.9	11.3-82.6	6	59.3	15.6-68.9	10	74.3	7.0-89.0
4	21	56.8	24.8-71.2	8	56.8	39.5-67.8	13	31.1	23.8-71.2
5	19	48.0	28.0-85.6	10	46.7	34.1-75.0	9	51.2	28.0-85.6
6	15	42.5	28.2-73.7	10	31.5	22.4-65.0	5	73.7	69.6-95.2
7	14	47.3	34.8-65.1	7	48.1	40.9-65.1	7	46.6	32.6-92.4
8	15	43.4	15.8-63.7	6	25.5	15.8-43.4	9	63.7	19.5-68.9
9	25	59.8	35.4-89.7	9	60.6	48.0-80.2	16	55.8	32.3-90.4
10	29	56.6	23.4-72.1	16	49.2	19.5-73.6	13	62.6	51.8-71.6
11	23	76.2	35.9-91.4	8	74.8	20.1-88.4	15	76.2	65.3-91.5
12	21	65.6	41.8-91.6	13	70.3	62.9-91.6	8	43.8	36.5-74.9
13	19	75.0	34.1-91.2	9	84.7	55.1-91.2	10	71.3	33.3-91.0
14	22	52.1	32.3-66.3	10	52.1	42.7-66.3	12	51.0	27.8-69.6
15	34	63.3	45.5-75.0	16	48.4	20.9-73.9	18	66.2	54.7-76.4
16	10	55.4	37.2-68.7	7	37.2	23.3-66.9	<5	-*	-*
17	8	46.9	38.0-89.7	<5	-*	-*	<5	-*	-*
18	8	36.3	19.1-68.5	5	52.4	18.9-84.6	<5	-*	-*
19	10	54.4	39.7-80.2	6	46.7	39.7-60.6	<5	-*	-*
Overall	**336****	**59.6**	**30.7-80.1**	**162**	**54.0**	**28.1-75.9**	**174**	**62.4**	**32.9-83.3**

1 Based on UK-WHO growth charts, 1990 (updated 1996)
* Redacted to adhere to statistical disclosure guidelines
**number with non-missing data

1.5 Body Mass Index (BMI) percentiles in children and young people (<20 years)[1]
N=364

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.

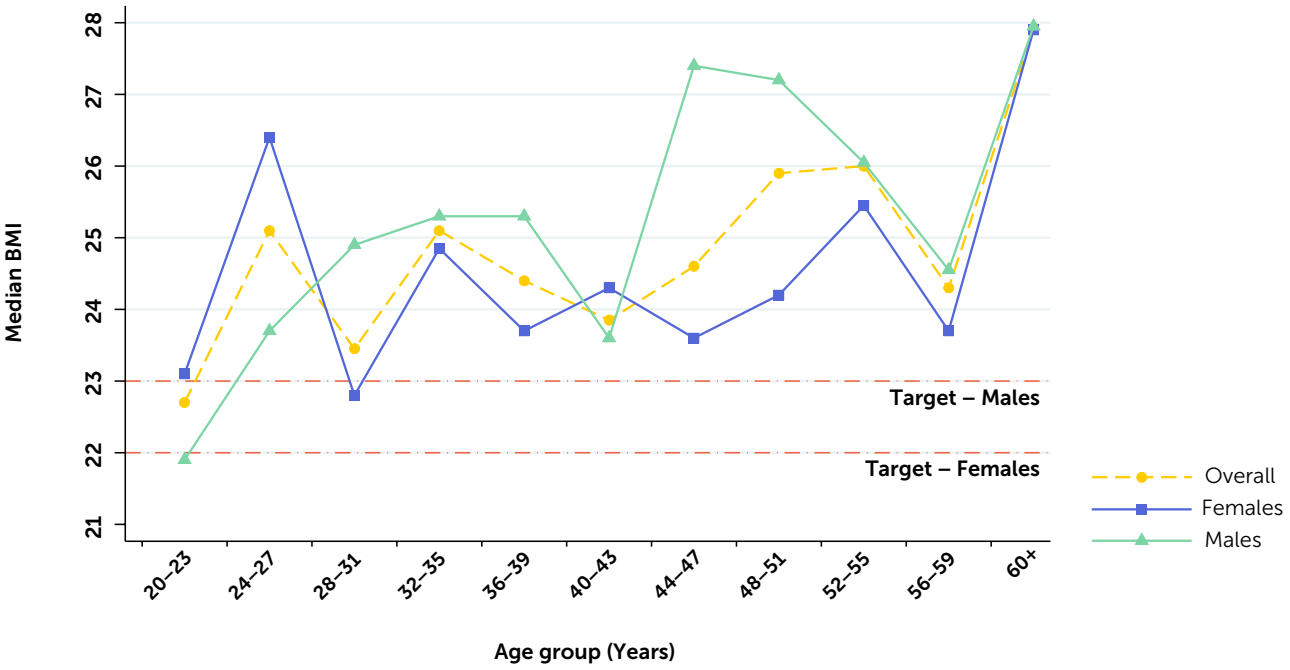


	Overall			Female			Male		
Age	n	Median	IQR	n	Median	IQR	n	Median	IQR
1	8	54.7	16.3-78.2	<5	-*	-*	<5	-*	-*
2	14	61.8	38.1-81.3	<5	-*	-*	10	69.8	59.1-85.2
3	14	51.0	19.5-87.2	5	24.8	19.5-62.7	9	73.9	27.4-87.2
4	20	54.3	25.2-80.2	8	54.6	23.8-77.6	12	45.7	26.6-81.6
5	19	59.1	40.4-74.9	10	66.6	48.4-74.9	9	42.4	31.1-74.3
6	15	50.0	34.6-89.6	10	41.8	26.7-56.9	5	89.6	59.0-91.5
7	14	46.0	36.7-71.6	7	40.3	12.0-71.6	7	51.8	38.5-91.7
8	15	60.4	23.6-67.7	6	45.3	23.6-60.4	9	63.4	28.0-85.1
9	24	63.7	41.5-89.2	9	71.3	52.0-88.3	15	55.6	37.3-90.1
10	29	62.2	42.3-79.5	16	46.8	28.5-69.2	13	65.3	55.2-85.1
11	23	75.9	28.6-92.0	8	71.3	16.5-86.8	15	81.1	63.2-96.1
12	21	69.3	41.5-94.4	13	70.3	67.6-94.4	8	37.4	18.9-86.4
13	19	76.8	36.3-91.3	9	78.1	62.7-91.3	10	63.5	36.3-90.8
14	22	53.3	34.5-74.3	10	53.3	34.5-82.5	12	53.5	32.9-67.2
15	34	65.5	29.0-83.4	16	57.7	24.9-77.9	18	68.9	29.0-84.4
16	10	79.3	34.2-91.4	7	47.9	27.5-86.1	<5	-*	-*
17	8	54.1	27.8-96.9	<5	-*	-*	<5	-*	-*
18	8	47.0	20.9-74.7	5	70.4	13.2-78.9	<5	-*	-*
19	10	61.7	39.6-83.0	6	61.7	39.6-67.1	<5	-*	-*
Overall	**327****	**62.7**	**31.9-85.1**	**157**	**60.9**	**29.1-79.4**	**170**	**63.5**	**33.9-86.8**

1 Based on UK-WHO growth charts, 1990 (updated 1996)
* Redacted to adhere to statistical disclosure guidelines
**number with non-missing data

1.6 Body Mass Index BMI in adults (≥ 20 years) N=413

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[1].



	Overall			Female			Male		
Age	n	Median	IQR	n	Median	IQR	n	Median	IQR
20-23	46	22.7	20.2-25.5	23	23.1	21.1-25.5	23	21.9	20.0-26.1
24-27	60	25.1	21.9-27.3	29	26.4	22.3-28.2	31	23.7	21.2-26.1
28-31	64	23.5	22.0-26.2	34	22.8	21.3-24.7	30	24.9	23.2-27.9
32-35	40	25.1	22.0-27.8	20	24.8	22.1-27.8	20	25.3	22.0-27.8
36-39	43	24.4	22.4-27.9	20	23.7	22.3-27.6	23	25.3	22.4-27.9
40-43	34	23.9	22.2-26.9	13	24.3	22.2-26.9	21	23.6	22.2-26.5
44-47	19	24.6	21.7-27.8	9	23.6	22.9-26.6	10	27.4	21.7-28.4
48-51	12	25.9	23.5-28.3	<5	-*	-*	8	27.2	24.7-29.4
52-55	16	26	24.2-29.0	6	25.5	24.1-30.0	10	26	24.3-28.9
56-59	20	24.3	22.7-27.2	10	23.7	22.6-26.7	10	24.5	23.4-27.7
60+	11	27.9	26.5-30.0	5	27.9	24.7-28.5	6	28	26.6-30.0
Overall	**365****	**24.4**	**22.1-27.6**	**173**	**24.3**	**22.0-27.0**	**192**	**24.5**	**22.1-27.8**

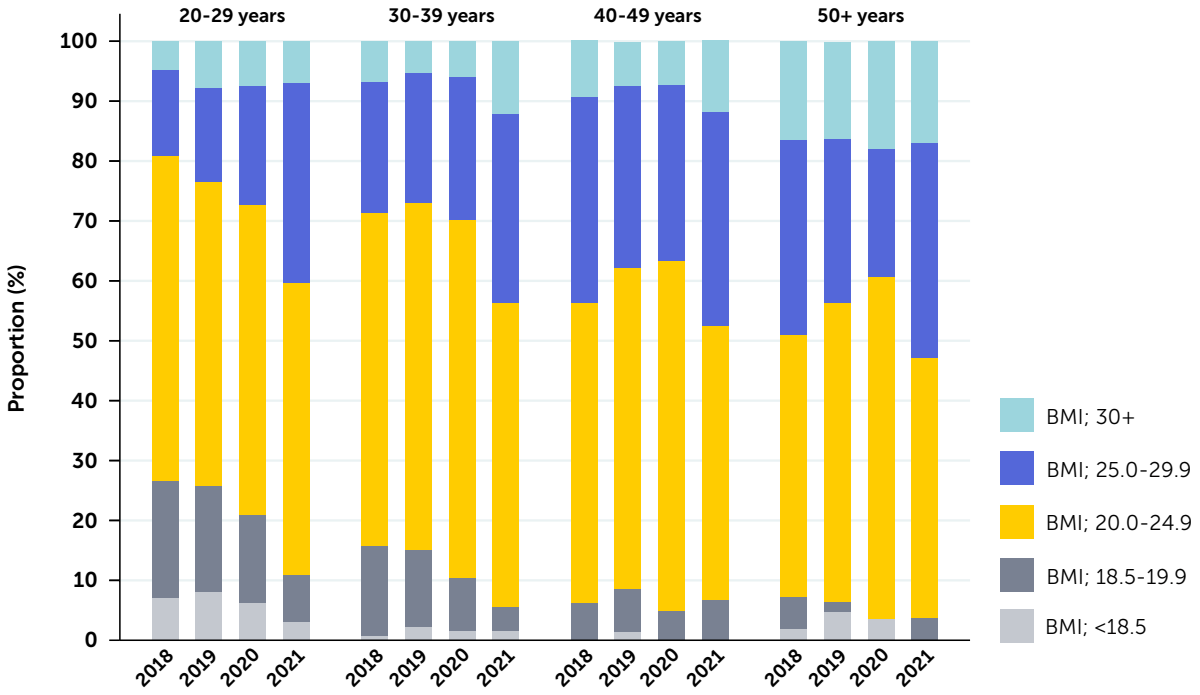
1 Stallings et al. J Am Diet Assoc. 2008 108:832-839

* Redacted to adhere to statistical disclosure guidelines

**number with non-missing data

1.7 Body Mass Index (BMI) in adults for 2018 – 2021

The following graph shows the change in the proportion of people in each BMI group from 2018 to 2021.



		Proportion (%) of age group in BMI category, by year				
Age group		**<18.5**	**18.5-19.9**	**20.0-24.9**	**25.0-29.9**	**30+**
20-29 years	**2018**	7.1	19.5	54.3	14.3	<5
	2019	8.1	17.7	50.7	15.8	7.7
	2020	6.3	14.7	51.6	20	7.4
	2021	<5	7.8	48.8	33.3	7
30-39 years						
	2018	<5	15	55.6	21.8	6.8
	2019	<5	12.8	57.9	21.8	5.3
	2020	<5	9	59.7	23.9	6
	2021	<5	<5	50.8	31.5	12.1
40-49 years						
	2018	0	6.3	50	34.4	9.4
	2019	<5	7.2	53.6	30.4	7.2
	2020	0	<5	58.5	29.3	7.3
	2021	0	6.8	45.8	35.6	11.9
50+ years						
	2018	<5	5.5	43.6	32.7	16.4
	2019	<5	<5	50	27.4	16.1
	2020	<5	0	57.1	21.4	17.9
	2021	0	<5	43.4	35.8	17

1.8 Education and employment in adults (≥16 years) N=452

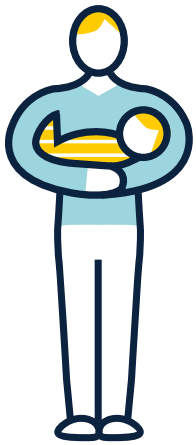
The following table shows how people with CF reported their education and employment status in 2021. Please note that the groups are not mutually exclusive; for example, someone may be a student as well as working part-time.

	Overall numbers of patients; n (%)	Male; n (%)	Female; n (%)
Number of patients	452	233	219
Number who completed questionnaire; n (%)	451 (99.8)	233 (100.0)	218 (99.5)
Full-time employment; n (%)	170 (37.7)	107 (45.9)	63 (28.9)
Part-time employment; n (%)	76 (16.9)	27 (11.6)	49 (22.5)
Student; n (%)	62 (13.7)	27 (11.6)	35 (16.1)
Homemaker; n (%)	11 (2.4)	<5	-*
Unemployed; n (%)	64 (14.2)	36 (15.5)	28 (12.8)
Disabled; n (%)	10 (2.2)	-*	<5
Retired; n (%)	14 (3.1)	8 (3.4)	6 (2.8)
Unknown entered; n (%)	43 (9.5)	20 (8.6)	23 (10.6)
No. in work or study; n (%)	308 (68.3)	161 (69.1)	147 (67.4)

* Redacted to adhere to statistical disclosure guidelines

1.9 Pregnancy

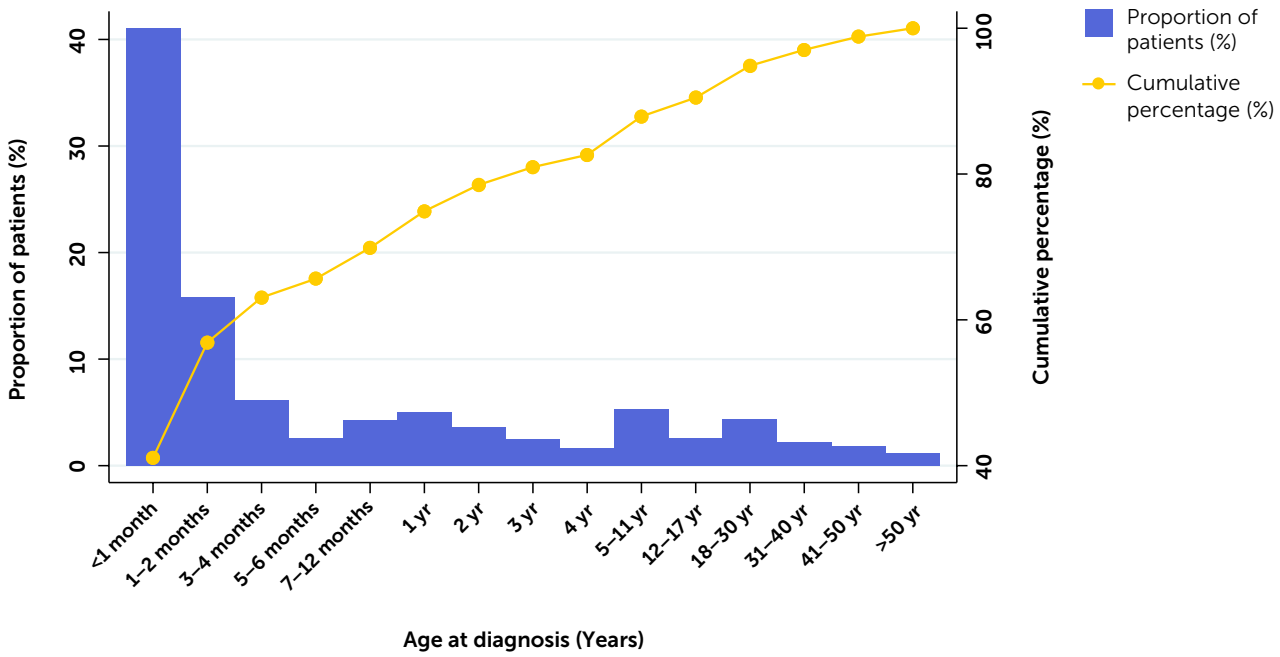
Seven women with cystic fibrosis had babies in Scotland during 2021



Fewer than five men with cystic fibrosis became fathers in Scotland during 2021

Diagnosis of cystic fibrosis

1.10 Age at diagnosis
N=777



The median age at diagnosis for patients aged under 16 in 2021 is **21 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 **nine patients** born in 2021 were identified by newborn screening (including those without complete data).

79 (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 2021.

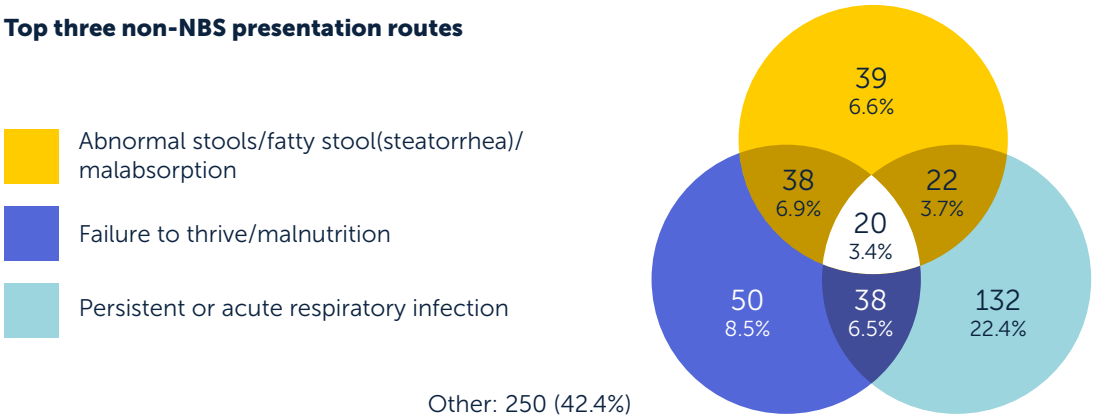
1.11 Mode of presentation

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	925	835	90
Number diagnosed by NBS	336	336	0
Total non-NBS	**589**	**499**	**90**

Mode of presentation (excluding NBS)	All patients (n=589)		Age <16 at diagnosis* (n=499)		Age ≥16 at diagnosis* (n=90)	
	n	(%)	n	(%)	n	(%)
Persistant or acute respiratory infection	212	36.0%	165	33.1%	47	52.2%
Failure to thrive/malnutrition	146	24.8%	146	29.3%	0	0.0%
Abnormal stools/fatty stool(steator- rhea)/malabsorption	119	20.2%	114	22.8%	5	5.6%
Meconium ileus	101	17.1%	101	20.2%	0	0.0%
Family history	85	14.4%	72	14.4%	13	14.4%
Unknown	45	7.6%	34	6.8%	11	12.2%
Genotype	30	5.1%	22	4.4%	8	8.9%
Electrolyte imbalance	22	3.7%	19	3.8%	<5	3.3%
Rectal prolapse	15	2.5%	15	3.0%	0	0.0%
Bronchiectasis	12	2.0%	<5	0.4%	10	11.1%
Fertility	9	1.5%	0	0.0%	9	10.0%
Nasal polyps	5	0.8%	5	1.0%	0	0.0%
Liver disease	<5	0.7%	<5	0.4%	<5	2.2%
Prenatal	<5	0.5%	<5	0.6%	0	0.0%
Oedema	<5	0.2%	<5	0.2%	0	0.0%
Pancreatitis	0	0.0%	0	0.0%	0	0.0%

Top three non-NBS presentation routes



Other: 250 (42.4%)

*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'[1].

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

1.12 FEV_1% predicted (GLI equations) at annual review in patients aged 6 years and older who have not had a lung transplant N= 652

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.

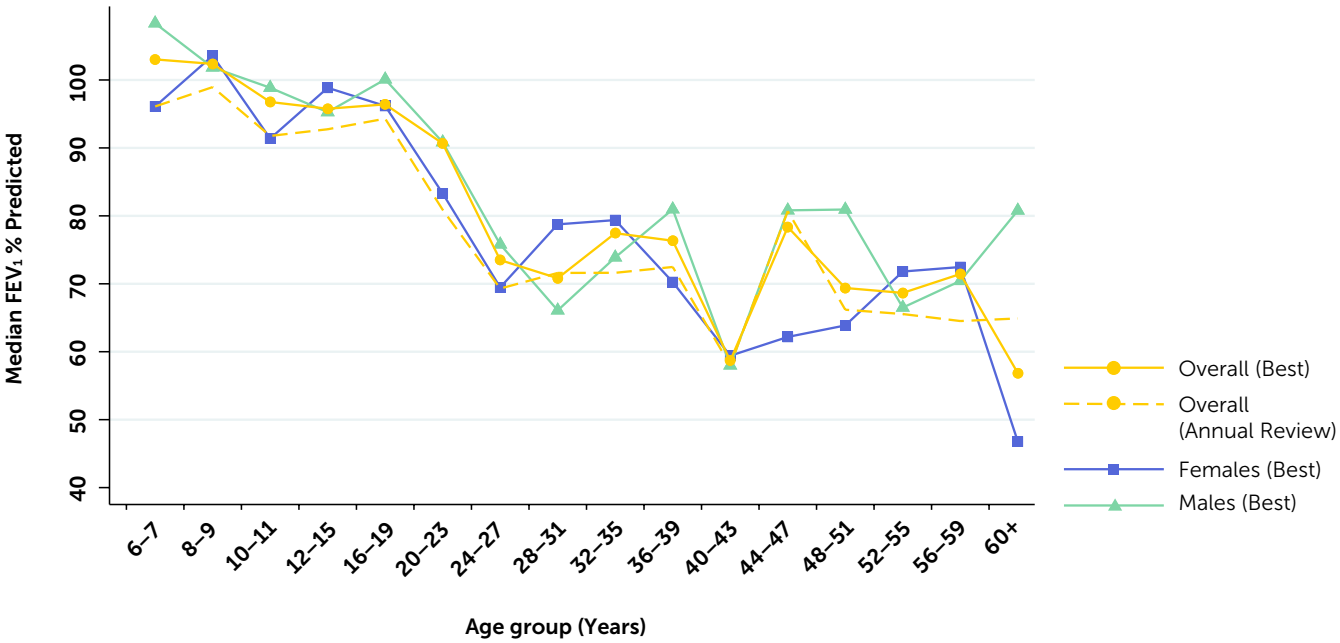
	Overall			Female			Male		
Age (yrs)	N	Median	IQR	N	Median	IQR	N	Median	IQR
6-7	24	96.1	90.7-107.4	13	95.2	92.1-99.9	11	98.4	90.2-109.6
8-9	38	98.9	88.1-102.3	15	98.2	87.5-105.8	23	99.3	88.4-102.3
10-11	50	91.8	85.9-100.0	23	88.6	83.9-96.8	27	94.6	89.2-106.7
12-15	90	92.7	85.9-102.6	45	94.5	86.0-103.2	45	91.7	85.7-100.1
16-19	37	94.3	85.4-102.8	23	92.3	83.7-101.5	14	99.6	89.3-106.4
20-23	43	80.9	58.7-97.8	22	76.6	60.5-100.3	21	90.8	44.5-97.8
24-27	63	69.3	40.6-83.3	32	68.2	41.9-88.7	31	70.5	40.6-77.8
28-31	60	71.6	48.3-89.3	31	78.8	59.9-91.1	29	64.7	46.4-80.2
32-35	39	71.6	65.5-84.8	19	74.9	69.3-84.8	20	71.0	46.4-84.0
36-39	41	72.5	53.3-85.7	19	64.6	48.8-80.7	22	73.6	53.6-89.7
40-43	25	58.0	45.3-72.0	9	63.3	52.6-85.8	16	53.8	41.9-71.2
44-47	16	80.7	50.9-95.5	7	52.1	48.0-95.6	9	83.0	78.3-95.3
48-51	14	66.2	43.9-77.9	6	61.5	43.9-66.3	8	72.0	40.5-86.6
52-55	20	65.6	48.2-69.1	7	65.7	53.6-86.4	13	65.4	44.0-68.7
56-59	20	64.5	48.7-77.6	11	63.5	52.2-72.6	9	70.4	40.6-81.6
60+	8	64.9	45.7-83.0	<5	-*	-*	6	79.9	50.7-85.4
<16	202	94.3	87.2-102.6	96	93.6	85.8-102.6	106	94.7	88.4-104.1
≥16	386	71.6	50.6-88.4	188	71.8	54.7-89.0	198	71.4	46.0-88.2
<18	222	94.3	87.2-102.6	109	93.4	85.8-102.6	113	94.8	88.5-104.1
≥18	366	70.5	49.2-86.7	175	70.3	53.3-87.5	191	70.8	46.0-86.6
Overall	**588****	**83.7**	**63.3-96.9**	**284**	**83.6**	**64.5-96.9**	**304**	**83.8**	**60.6-96.8**

* Redacted to adhere to statistical disclosure guidelines

**number with non-missing data

1.13 Best FEV_1% predicted (GLI equations) in patients aged 6 years and older who have not had a lung transplant
N=652

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.

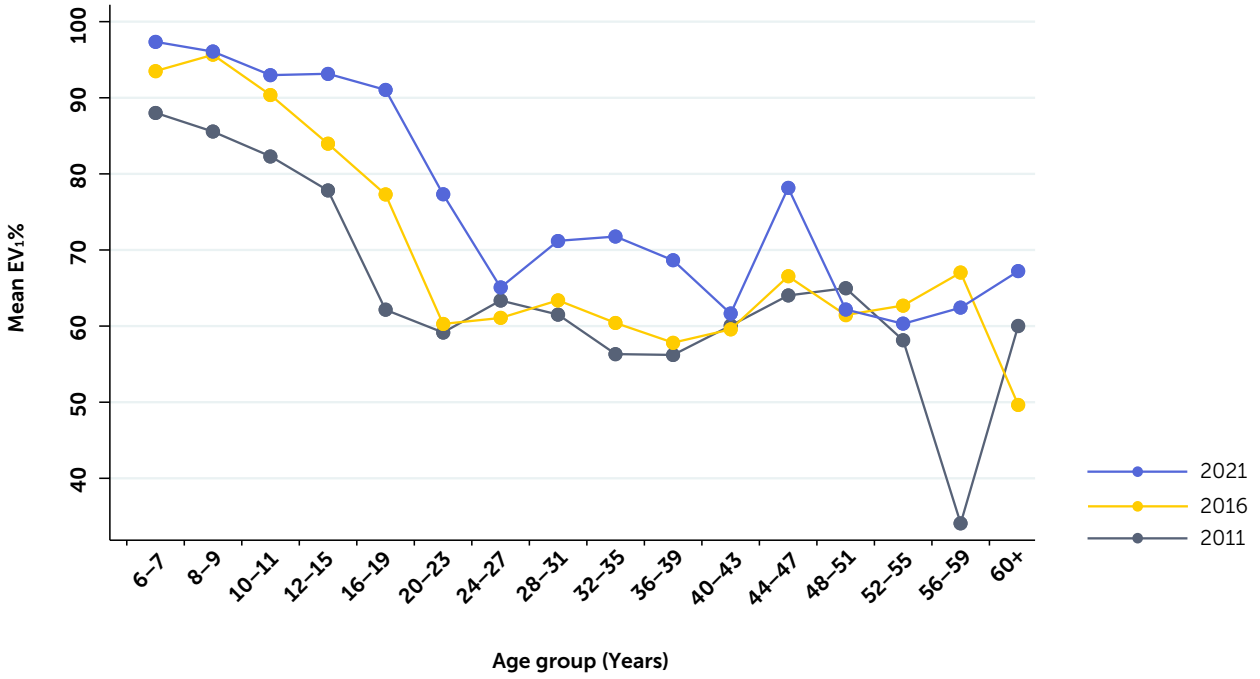


	Overall			Female			Male		
Age (yrs)	N	Median	IQR	N	Median	IQR	N	Median	IQR
6-7	31	103.0	90.2-109.9	16	96.1	87.1-109.0	15	108.3	91.1-115.3
8-9	41	102.4	94.1-107.0	15	103.6	88.1-113.3	26	101.8	94.1-106.8
10-11	52	96.8	88.9-107.8	23	91.4	85.0-105.8	29	98.9	92.5-108.2
12-15	97	95.8	89.5-107.5	47	98.8	88.9-109.3	50	95.3	90.1-105.8
16-19	39	96.4	87.3-106.4	24	96.2	86.8-102.4	15	100.1	87.3-107.4
20-23	47	90.7	59.9-101.2	24	83.3	63.5-103.2	23	90.8	51.9-101.2
24-27	67	73.5	45.2-88.8	33	69.5	44.0-96.9	34	75.8	47.2-84.8
28-31	64	70.8	53.0-90.4	33	78.8	59.9-90.5	31	66.1	48.1-83.6
32-35	41	77.4	68.6-83.2	21	79.4	69.3-83.2	20	73.9	46.4-84.5
36-39	41	76.3	53.3-87.6	19	70.3	48.8-83.8	22	81.0	57.6-91.7
40-43	32	58.7	46.6-75.8	11	59.4	47.3-89.1	21	58.0	46.0-71.8
44-47	19	78.4	52.1-95.3	8	62.2	50.9-92.3	11	80.8	64.1-95.3
48-51	14	69.4	43.9-81.7	6	63.9	43.9-69.3	8	80.9	51.2-91.1
52-55	21	68.7	49.5-74.0	8	71.8	52.1-84.3	13	66.5	47.8-72.2
56-59	20	71.4	51.7-83.6	11	72.5	52.2-77.6	9	70.4	48.3-86.0
60+	10	56.9	46.8-82.2	<5	-*	-*	7	80.8	50.7-85.4
<16	221	98.2	89.9-108.2	101	98.2	87.4-109.3	120	98.7	91.4-107.9
≥16	415	76.2	53.3-91.1	201	77.4	56.0-92.0	214	75.2	50.7-90.3
<18	241	98.4	89.9-107.6	114	98.2	88.1-108.0	127	98.9	90.8-107.6
≥18	395	73.8	52.4-90.2	188	74.1	55.0-90.4	207	73.5	50.1-89.3
Overall	**636****	**86.5**	**66.7-100.0**	**302**	**85.9**	**67.4-99.7**	**334**	**88.1**	**66.1-100.1**

* Redacted to adhere to statistical disclosure guidelines
**number with non-missing data

1.14 FEV_1% predicted (GLI equations) over time in patients aged 6 years and older who have not had a lung transplant N=652 in 2021, N=695 in 2016, N=575 in 2011*

The chart below shows how FEV_1% in 2021 compares to Registry data from 2011 and 2016. 2011 is shown as a comparator year.



Age (yrs)	n	2011 mean FEV_1%: Mean (SD)	n	2016 mean FEV_1%: Mean (SD)	n	2021 mean FEV_1%: Mean (SD)	p-values (t-test)**
6-7	39	88.0 (9.8)	46	93.5 (14.6)	24	97.4 (13.2)	0.283
8-9	25	85.6 (15.7)	37	95.6 (12.6)	38	96.1 (12.7)	0.888
10-11	27	82.3 (17.3)	47	90.4 (13.1)	50	93.0 (13.4)	0.335
12-15	60	77.8 (18.7)	71	84.0 (14.0)	90	93.2 (14.1)	<0.001
16-19	93	62.1 (21.3)	71	77.3 (24.0)	37	91.0 (20.2)	0.004
20-23	79	59.2 (23.9)	94	60.3 (23.7)	43	77.3 (24.3)	<0.001
24-27	61	63.4 (23.9)	77	61.1 (25.0)	63	65.1 (24.1)	0.340
28-31	44	60.7 (20.8)	57	63.4 (22.9)	60	71.2 (24.8)	0.080
32-35	27	56.3 (24.6)	48	60.4 (21.8)	39	71.8 (19.3)	0.013
36-39	17	56.2 (23.2)	32	57.8 (25.4)	41	68.6 (20.9)	0.050
40-43	19	60.1 (24.6)	19	59.6 (25.9)	25	61.6 (24.3)	0.788
44-47	15	64.0 (25.5)	18	66.5 (22.9)	16	78.2 (29.7)	0.207
48-51	15	65.0 (17.9)	18	61.5 (21.7)	14	62.2 (22.8)	0.927
52-55	8	58.1 (28.9)	14	62.7 (16.6)	20	60.3 (17.7)	0.693
56-59	6	34.1 (14.0)	5	67.0 (22.5)	20	62.4 (18.5)	***
60+	<5	60.0 (27.2)	9	49.7 (20.2)	8	67.2 (25.3)	***
<16	151	82.5 (16.5)	201	89.8 (14.4)	202	94.2 (13.6)	N/A
≥16	387	60.4 (22.9)	462	63.5 (24.1)	386	70.8 (24.0)	N/A
<18	200	78.1 (19.3)	230	88.7 (15.6)	222	94.0 (14.0)	N/A
≥18	338	59.9 (23.2)	433	62.3 (23.9)	366	69.6 (23.7)	N/A

* due to missing data, means are calculated from a population of 588 in 2021, 663 in 2016 and 538 in 2011.

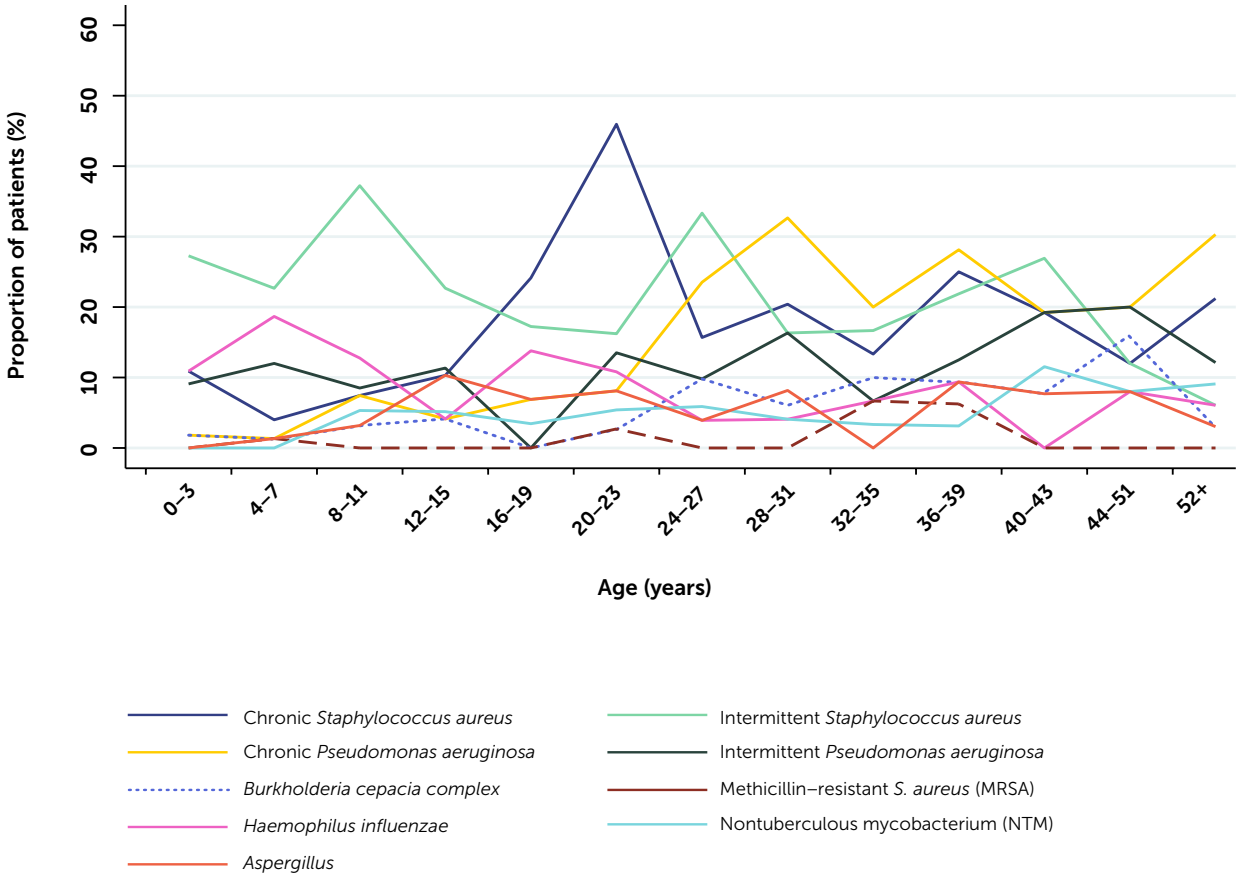
** t-test comparing 2021 with 2016. If the p-value is less than 0.05 then the difference in the mean is statistically significant.

***t-test not performed due to small numbers in these age groups.

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 2021
N=633*



* Proportions are calculated from the number of patients with at least one sample taken in the relevant age group. This is a change from the 2020 data report where they were calculated from the number of people with Annual reviews in the age group.

1.16 Lung infections in 2021
<16 years N=321; ≥16 years N=312

Infections in this table reflect bugs grown in the 12 months prior to the 2020 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	55	75	96	99	325
Number who had culture taken*	55	75	94	97	321
Chronic *S. aureus* n (%)	6 (10.9)	<5	7 (7.4)	10 (10.3)	26 (8.1)
Intermittent *S. aureus* n (%)	15 (27.3)	17 (22.7)	35 (37.2)	22 (22.7)	89 (27.7)
Chronic *P. aeruginosa* n (%)	<5	<5	7 (7.4)	<5	13 (4.0)
Intermittent *P. aeruginosa* n (%)	5 (9.1)	9 (12.0)	8 (8.5)	11 (11.3)	33 (10.3)
B. cepacia complex n (%)	<5	<5	<5	<5	9 (2.8)
B. cenocepacia n (%)	0	0	<5	<5	<5
B. multivorans n (%)	0	0	0	<5	<5
B. other cepacia n (%)	0	<5	<5	0	<5
MRSA n (%)	0	<5	0	0	<5
H. influenza n (%)	6 (10.9)	14 (18.7)	12 (12.8)	<5	36 (11.2)
NTM n (%)	0	0	5 (5.3)	5 (5.2)	10 (3.1)
Aspergillus fumigatus n (%)	0	<5	<5	10 (10.3)	14 (4.4)

* Proportions are calculated from the number of people who were recorded as having at least one respiratory culture sample taken.

** Redacted to adhere to statistical disclosure guidelines.

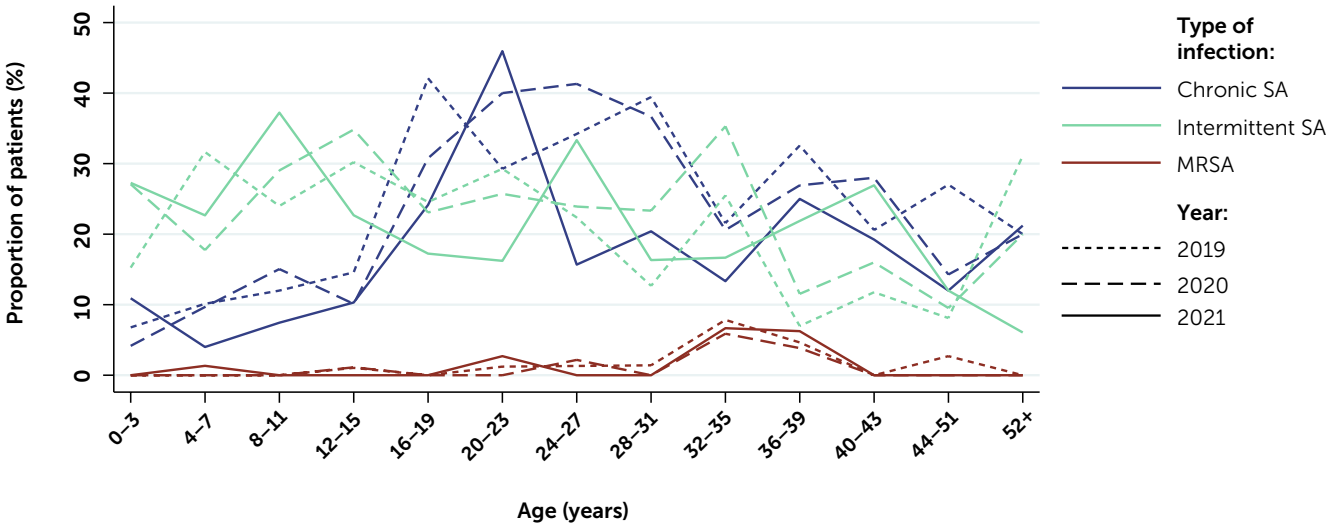
Lung infections in 2021 (contd.)
<16 years N=321; ≥16 years N=312

	Adult Age Range (Years)					Overall
	16-19	20-23	24-27	28-31	32-35	Adults (≥16 years)
Number in age range	39	51	70	68	45	452
Number who had culture taken*	29	37	51	49	30	312
Chronic *S. aureus* n (%)	7 (24.1)	17 (45.9)	8 (15.7)	10 (20.4)	<5	69 (22.1)
Intermittent *S. aureus* n (%)	5 (17.2)	6 (16.2)	17 (33.3)	8 (16.3)	5 (16.7)	60 (19.2)
Chronic *P. aeruginosa* n (%)	<5	<5	12 (23.5)	16 (32.7)	6 (20.0)	68 (21.8)
Intermittent *P. aeruginosa* n (%)	0	5 (13.5)	5 (9.8)	8 (16.3)	<5	38 (12.2)
B. cepacia complex n (%)	0	<5	5 (9.8)	<5	<5	22 (7.1)
B. cenocepacia n (%)	0	0	<5	<5	<5	8 (2.6)
B. multivorans n (%)	0	<5	<5	<5	<5	10 (3.2)
B. other cepacia n (%)	0	0	<5	0	0	<5
MRSA n (%)	0	<5	0	0	<5	5 (1.6)
H. influenza n (%)	<5	<5	<5	<5	<5	21 (6.7)
NTM n (%)	<5	<5	<5	<5	<5	18 (5.8)
Aspergillus n (%)	<5	<5	<5	<5	0	19 (6.1)

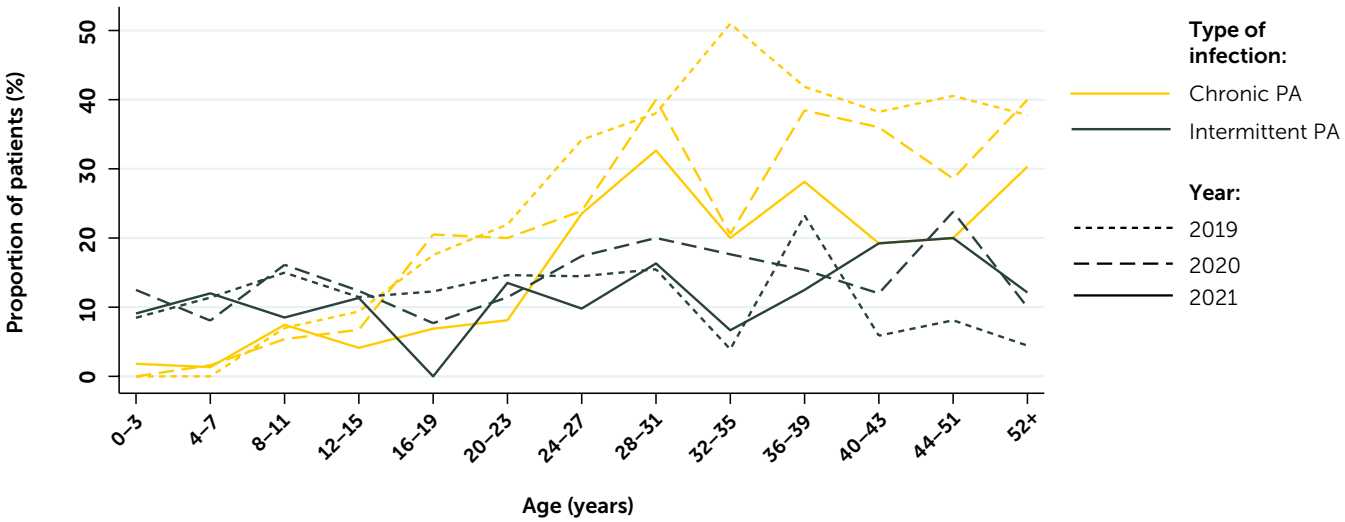
		Adult Age Range (Years)			Overall
	36-39	40-43	44-51	52+	Adults (≥16 years)
Number in age range	45	39	38	57	452
Number who had culture taken*	32	26	25	33	312
Chronic *S. aureus* n (%)	8 (25.0)	5 (19.2)	<5	7 (21.2)	69 (22.1)
Intermittent *S. aureus* n (%)	7 (21.9)	7 (26.9)	<5	<5	60 (19.2)
Chronic *P. aeruginosa* n (%)	9 (28.1)	5 (19.2)	5 (20.0)	10 (30.3)	68 (21.8)
Intermittent *P. aeruginosa* n (%)	<5	5 (19.2)	5 (20.0)	<5	38 (12.2)
B. cepacia complex n (%)	<5	<5	<5	<5	22 (7.1)
B. cenocepacia n (%)	<5	0	<5	0	8 (2.6)
B. multivorans n (%)	0	<5	<5	0	10 (3.2)
B. other cepacia n (%)	<5	0	0	<5	<5
MRSA n (%)	<5	0	0	0	5 (1.6)
H. influenza n (%)	<5	0	<5	<5	21 (6.7)
NTM n (%)	<5	<5	<5	<5	18 (5.8)
Aspergillus n (%)	<5	<5	<5	<5	19 (6.1)

1.17 Lung infections 2019-2021

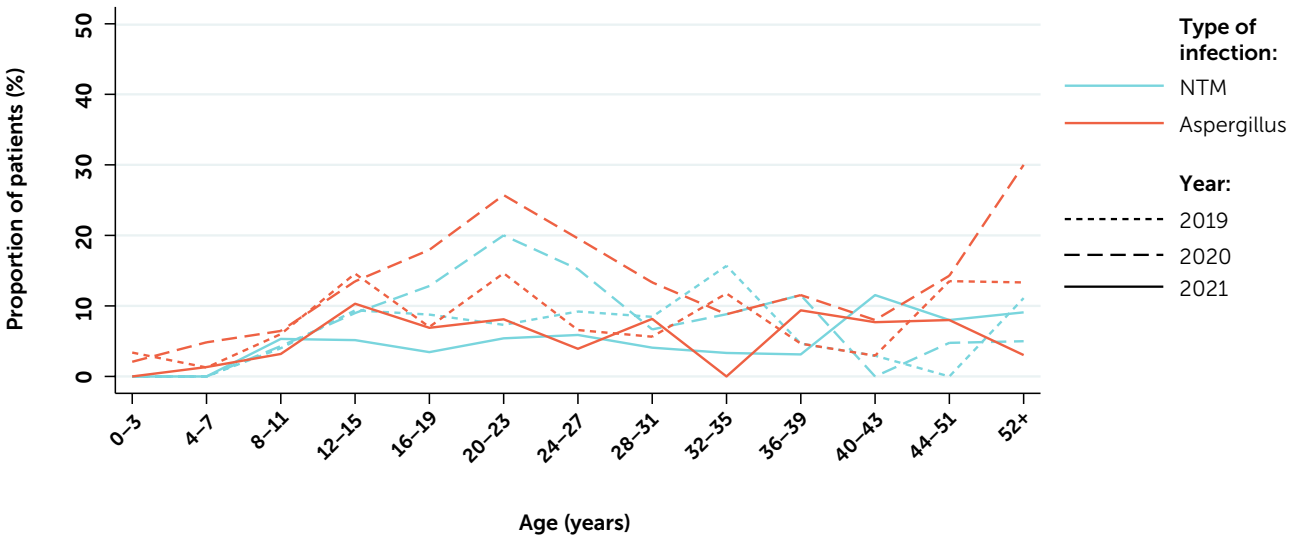
Staphylococcus aureus infections



Pseudomonas aeruginosa infections



Other infections



1.18 Respiratory culture sample type

Overall	2019	2020	2021
Number of people with an annual review (N)	868	814	777
Number people with any sample taken N(%)*	831 (95.7)	753 (92.5)	635 (81.7)
Sample type**			
Sputum; N(%)	609 (73.3)	543 (72.1)	329 (51.8)
Cough; N(%)	421 (50.7)	536 (71.2)	439 (69.1)
Bronchoalveolar lavage; N(%)	25 (3.0)	202 (26.8)	18 (2.8)
Age<16 years	**2019**	**2020**	**2021**
Number of people <16 with an annual review (N)	336	298	325
Number people <16 with any sample taken N(%)*	3937 (99.7)	294 (98.7)	323 (99.4)
Sample type**			
Sputum; N(%)	129 (38.5)	95 (32.3)	69 (21.4)
Cough; N(%)	326 (97.3)	285 (96.9)	323 (99.4)
Bronchoalveolar lavage; N(%)	19 (5.7)	19 (6.5)	14 (4.3)
Age>=16 years	**2019**	**2020**	**2021**
Number of people >=16 with an annual review (N)	532	516	452
Number people >=16 with any sample taken N(%)*	496 (93.2)	459 (90.0)	312 (69.0)
Sample type**			
Sputum; N(%)	480 (96.8)	448 (97.6)	260 (83.3)
Cough; N(%)	95 (19.2)	251 (54.7)	116 (37.2)
Bronchoalveolar lavage; N(%)	6 (1.2)	183 (39.9)	<5

* % is of those people with an annual review.

** Patients can have more than one sample taken so the % total may not add up to 100%.

1.19 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.

	2019 (n=868)	2020 (n=814)	2021 (n=777)
NTM Prevalence; n (%)	53 (6.1)	41 (5.0)	28 (3.6)
On NTM treatment in the given year; n (% of NTM prevalence in given year)	29 (54.7)	19 (46.3)	11 (39.3)
NTM Incidence	23 (2.8)	12 (1.6)	13 (1.8)
Mycobacterium abscessus prevalence	32 (3.7)	27 (3.3)	14 (1.8)
Mycobacterium abscessus incidence	13 (1.6)	6 (0.8)	<5

1.20 COVID-19* infection in 2021

COVID-19 management and outcomes for people with CF infected with COVID-19 during the calendar year of 2021 are described below. Information is stratified by sex, ethnicity, age, organ transplant status and Best FEV_1% prior to catching COVID-19.

			COVID-19 Management	Outcomes
Categorical	Total	Symptomatic	Oral antibiotics	Hospitalised
Overall; n(%)				
All cases	56 (100.0)	41 (73.2)	21 (37.5)	10 (17.9)
Sex; n(%)				
Female	26 (46.4)	21 (80.8)	8 (30.8)	<5
Male	30 (53.6)	20 (66.7)	13 (43.3)	6 (20.0)
Ethnicity; n(%)				
White	53 (94.6)	38 (71.7)	20 (37.7)	10 (18.9)
**Any other	<5	<5	<5	0 (0.0)
Age; n(%)				
Under 16	23 (41.1)	14 (60.9)	11 (47.8)	<5
>= 16	33 (58.9)	27 (81.8)	10 (30.3)	9 (27.3)
Transplants; n(%)				
No	50 (89.3)	35 (70.0)	20 (40.0)	5 (10.0)
Yes	6 (10.7)	6 (100.0)	<5	5 (83.3)
***BestFEV; n(%)**				
<40	5 (10.6)	<5	<5	<5
40-70	14 (29.8)	10 (71.4)	<5	<5
>70	28 (59.6)	21 (75.0)	11 (39.3)	<5

Fewer than five people had IV antibiotics. Fewer than five people had oxygen. Fewer than five people were hospitalised and had oxygen.

* COVID-19 cases confirmed with positive PCR or lateral flow tests

** Including unknown ethnicity

*** Patients who had a lung transplant were excluded

Complications

1.21 Complications in 2021

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

Complications	Overall	<16 years	≥16 years
Respiratory related			
Nasal polyps requiring surgery	15 (1.9)	7 (2.2)	8 (1.8)
Sinus disease	92 (11.8)	0	92 (20.4)
Asthma	50 (6.4)	0	50 (11.1)
ABPA	37 (4.8)	6 (1.8)	31 (6.9)
Any haemoptysis	8 (1.0)	<5	7 (1.5)
Massive haemoptysis	<5	0	<5
Pneumothorax requiring chest tube	0 (0.0)	0	0
Pancreas and hepatobiliary disease			
Raised liver enzymes	70 (9.0)	11 (3.4)	59 (13.1)
Liver disease	118 (15.2)	37 (11.4)	81 (17.9)
Cirrhosis with no portal hypertension	19 (2.4)	7 (2.2)	12 (2.7)
Cirrhosis with portal hypertension	15 (1.9)	<5	13 (2.9)
Gall bladder disease requiring surgery	16 (2.1)	<5	13 (2.9)
Pancreatitis	11 (1.4)	0	11 (2.4)
Upper gastrointestinal (GI)			
GORD	152 (19.6)	11 (3.4)	141 (31.2)
Peptic ulcer	0 (0.0)	0	0
GI bleed (varices as source)	5 (0.6)	<5	<5
GI bleed (non varices as source)	<5	0	<5
Lower gastrointestinal			
Intestinal obstruction	0 (0.0)	0	0
DIOS	89 (11.5)	5 (1.5)	84 (18.6)
Fibrosing colonopathy / colonic stricture	0 (0.0)	0	0
Rectal prolapse	<5	<5	0
Renal			
Kidney stones	6 (0.8)	<5	5 (1.1)
Renal failure	7 (0.9)	0	7 (1.5)
Musculoskeletal			
Arthritis	8 (1.0)	0	8 (1.8)
Arthropathy	28 (3.6)	<5	27 (6.0)
Bone fracture	0 (0.0)	0	0
Osteopenia	78 (10.0)	0	78 (17.3)
Osteoporosis	40 (5.1)	0	40 (8.8)
Other			
Cancer confirmed by histology	<5	0	<5
Port inserted or replaced	9 (1.2)	5 (1.5)	<5
Depression	22 (2.8)	<5	21 (4.6)
Hearing loss	12 (1.5)	<5	11 (2.4)
Hypertension	25 (3.2)	0	25 (5.5)

* Redacted to adhere to statistical disclosure guidelines.

1.22 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.

	2020			2021		
	Overall (n=632)	<16 years (n=299)	≥16 years (n=333)	Overall (n=777)	<16 years (n=325)	≥16 years (n=452)
ABPA; n (%)	7 (2.1)	<5	10 (1.6)	21 (2.7)	16 (3.5)	5 (1.5)
Cirrhosis - no portal hypertension; n (%)	0 (0)	0 (0)	0 (0)	15 (1.9)	8 (1.8)	7 (2.2)
Cirrhosis - with portal hypertension; n (%)	<5	<5	6 (.9)	<5	<5	0
Cancer confirmed by histology; n (%)	<5	<5	<5	<5	<5	0

1.23 CF-related diabetes
N= 7887

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=605)	10-15 years (N=153)	≥16 years (N=452)
On CFRD treatment; n (%)	129 (21.3)	12 (7.8)	117 (25.9)
Of those on treatment			
Insulin[1]; n (%)	128 (99.2)	11 (91.7)	117 (100.0)
CFRD Screening ; n(%)			
Yes	251 (41.5)	94 (61.4)	157 (34.7)
Screening type ;n(%)			
Continous glucose monitoring[2]; n (%)	60 (23.9)	30 (31.9)	30 (19.1)
Oral glucose tolerance test[2]; n (%)	150 (59.8)	70 (74.5)	80 (51.0)
Not screened (known CFRD)	145 (24.0)	10 (6.5)	135 (29.9)
Not screened (other)	192 (31.7)	48 (31.4)	144 (31.9)
Unknown	16 (2.6)	<5	16 (3.5)

1 Proportion of patients on treatment
2 Proportion of patients screened
* redacted to adhere to statistical disclosure guidelines

Antibiotics

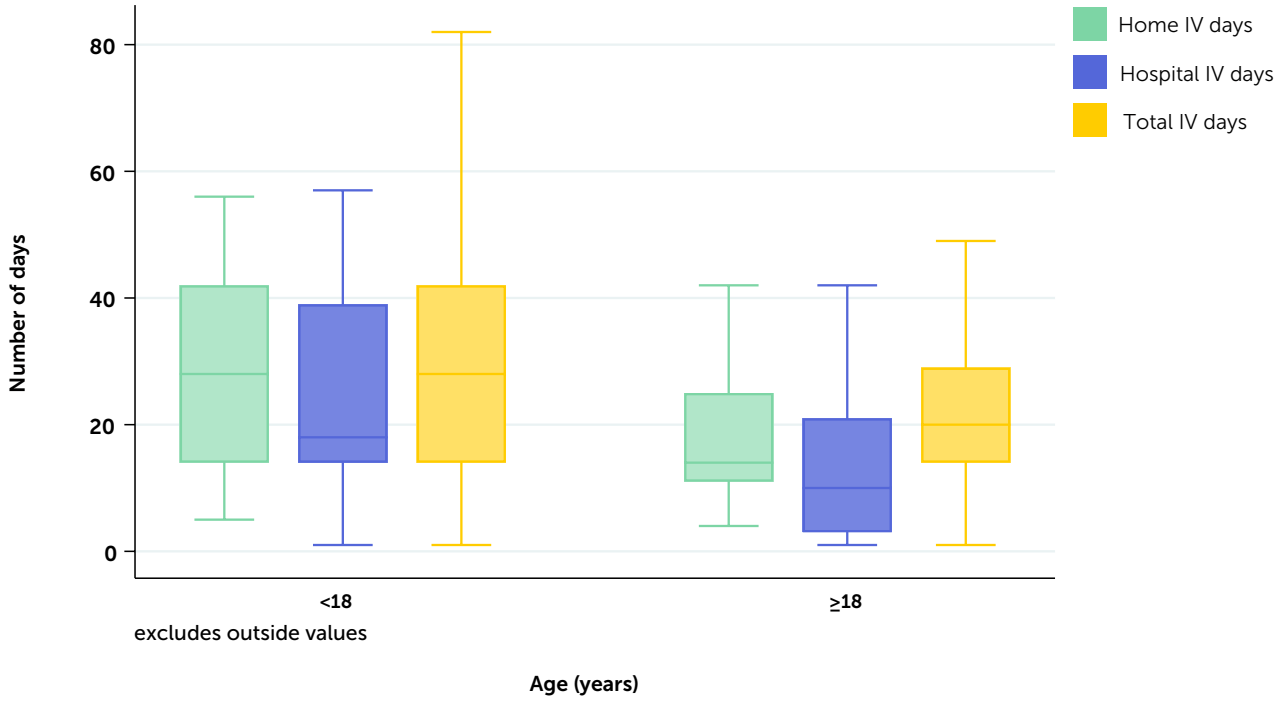
1.24 Intravenous (IV) antibiotics
N=777

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.

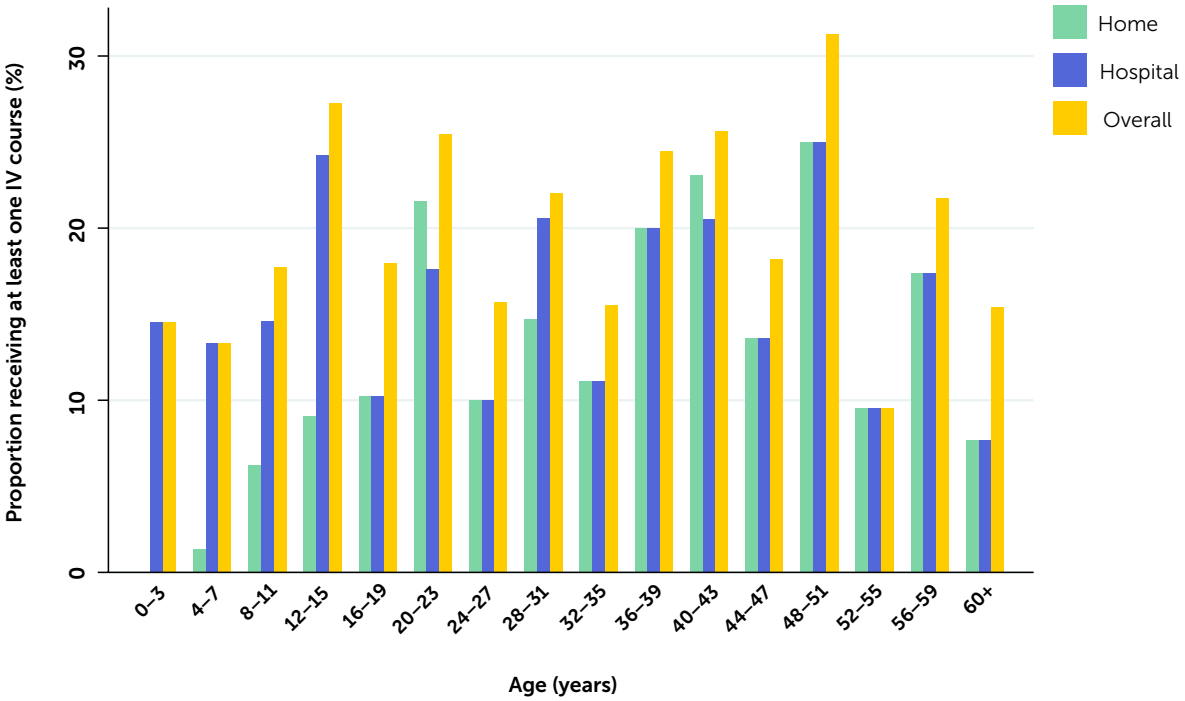
		Home		Hospital		Total	
Age	N	Patients n (%)	Median days (IQR)	Patients n (%)	Median days (IQR)	Patients n (%)	Median days (IQR)
0-3	55	0	-*	8 (14.5)	14 (6-15)	8 (14.5)	14 (6-15)
4-7	75	<5	-*	10 (13.3)	19 (14-42)	10 (13.3)	29 (14-42)
8-11	96	6 (6.3)	42 (28-50)	14 (14.6)	30 (14-39)	17 (17.7)	38 (22-56)
12-15	99	9 (9.1)	28 (14-40)	24 (24.2)	21 (14-42)	27 (27.3)	31 (15-44)
16-19	39	<5	-*	<5	-*	7 (17.9)	14 (9-14)
20-23	51	11 (21.6)	14 (10-28)	9 (17.6)	10 (4-14)	13 (25.5)	14 (14-28)
24-27	70	7 (10.0)	15 (11-28)	7 (10.0)	13 (3-14)	11 (15.7)	14 (14-28)
28-31	68	10 (14.7)	16 (10-24)	14 (20.6)	7 (4-27)	15 (22.1)	28 (7-42)
32-35	45	5 (11.1)	14 (14-25)	5 (11.1)	14 (3-17)	7 (15.6)	17 (14-28)
36-39	45	9 (20.0)	19 (13-28)	9 (20.0)	6 (3-26)	11 (24.4)	28 (14-41)
40-43	39	9 (23.1)	20 (13-28)	8 (20.5)	22 (9-33)	10 (25.6)	35 (20-42)
44-47	22	<5	-*	<5	-*	<5	-*
48-51	16	<5	-*	<5	-*	5 (31.3)	15 (14-22)
52-55	21	<5	-*	<5	-*	<5	-*
56-59	23	<5	-*	<5	-*	5 (21.7)	25 (14-28)
60+	13	<5	-*	<5	-*	<5	-*
<16	325	16 (4.9)	33 (15-42)	56 (17.2)	19 (14-40)	62 (19.1)	30 (14-42)
≥16	452	69 (15.3)	14 (11-25)	70 (15.5)	10 (3-21)	92 (20.4)	18 (14-28)
<18	345	19 (5.5)	28 (14-42)	59 (17.1)	18 (14-39)	67 (19.4)	28 (14-42)
≥18	432	66 (15.3)	14 (11-25)	67 (15.5)	10 (3-21)	87 (20.1)	20 (14-29)
Overall	**777**	**85 (10.9)**	**14 (12-28)**	**126 (16.2)**	**14 (5-28)**	**154 (19.8)**	**23 (14-42)**

* Redacted to adhere to statistical disclosure guidelines.

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 44.



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 10.9% and in hospital was 16.2%. The proportion receiving any IVs was 19.8%.



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

	2011			2016			2021		
	Overall	<16 years	≥16 years	Overall	<16 years	≥16 years	Overall	<16 years	≥16 years
Patients with chronic *P. aeruginosa*	197	16	181	220	12	208	81	13	68
Tobramycin solution; n (%)	32 (16.2)	0	32 (17.7)	38 (17.3)	<5	35 (16.8)	15 (18.5)	<5	13 (19.1)
Other aminoglycoside; n (%)	<5	0	<5	<5	0	<5	<5	0	<5
Colistin; n (%)	64 (32.5)	11 (68.8)	53 (29.3)	76 (34.5)	9 (75.0)	67 (32.2)	26 (32.1)	9 (69.2)	17 (25.0)
Promixin; n (%)	62 (31.5)	<5	61 (33.7)	32 (14.5)	<5	31 (14.9)	8 (9.9)	<5	6 (8.8)
Aztreonam; n (%)	-	-	-	10 (4.5)	0	10 (4.8)	5 (6.2)	0	5 (7.4)
Colistimethate (DPI); n (%)	-	-	-	45 (20.5)	<5	44 (21.2)	9 (11.1)	<5	7 (10.3)
Tobramycin Inhalation Powder; n (%)	-	-	-	68 (30.9)	0	68 (32.7)	13 (16.0)	0	13 (19.1)
Levofloxacin ;n(%)	-	-	-	-	-	-	<5	0	<5
At least one of the above; n (%)	152 (77.2)	12 (75.0)	140 (77.3)	173 (78.6)	11 (91.7)	162 (77.9)	55 (67.9)	12 (92.3)	43 (63.2)

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.

1.26 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.

		Number of patients on azithromycin; n	Patients with chronic *P. aeruginosa*; n (%)	Patients without chronic *P. aeruginosa*; n (%)
2011	**Overall**	329	170 (51.7)	159 (48.3)
	0-3 years	<5	<5	<5
	4-15 years	43	6 (14.0)	37 (86.0)
	≥ 16 years	284	163 (57.4)	121 (42.6)
2016	**Overall**	431	187 (43.4)	244 (56.6)
	0-3 years	0	0 (0)	0 (0)
	4-15 years	89	10 (11.2)	79 (88.8)
	≥ 16 years	342	177 (51.8)	165 (48.2)
2021	**Overall**	384	64 (16.7)	320 (83.3)
	0-3 years	6	0 (0.0)	6 (100.0)
	4-15 years	92	6 (6.5)	86 (93.5)
	≥ 16 years	286	58 (20.3)	228 (79.7)

1.27 Flucloxacillin use

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

Age	Total patients	Patients on prophylactic flucloxacillin; n (%)
0-3	55	35 (63.6)
4-7	74	35 (47.3)
8-11	96	30 (31.3)
12-15	97	25 (25.8)
16-19	37	7 (18.9)
20-23	51	15 (29.4)
24-27	67	11 (16.4)
28-31	66	8 (12.1)
32-35	45	<5
36-39	44	<5
40-43	39	0
44-47	22	0
48-51	15	<5
52-55	20	<5
56-59	22	<5
60+	13	0
<16 years	322	125 (38.8)
≥16 years	441	48 (10.9)
<18 years	341	128 (37.5)
≥18 years	422	45 (10.7)
Overall	**763**	**173 (22.7)**

* Redacted to adhere to statistical disclosure guidelines.

Muco-active therapies

1.28 Mannitol

Age	Total patients	Patients on Mannitol; n (%)
<16 years	322	0
≥16 years	441	<5
<18 years	341	0
≥18 years	422	<5
Overall	**763**	**<5**

1.29 DNase

	2011		2016		2021	
Age	Total patients	Patients on DNase; n (%)	Total patients	Patients on DNase; n (%)	Total patients	Patients on DNase; n (%)
0-3	69	<5	54	6 (11.1)	55	<5
4-7	90	10 (11.1)	91	19 (20.9)	74	18 (24.3)
8-11	58	20 (34.5)	86	40 (46.5)	96	42 (43.8)
12-15	66	23 (34.8)	69	36 (52.2)	97	57 (58.8)
16-19	98	41 (41.8)	66	40 (60.6)	37	26 (70.3)
20-23	84	37 (44.0)	97	53 (54.6)	51	31 (60.8)
24-27	69	19 (27.5)	79	39 (49.4)	67	47 (70.1)
28-31	52	18 (34.6)	65	28 (43.1)	66	44 (66.7)
32-35	34	8 (23.5)	49	17 (34.7)	45	20 (44.4)
36-39	21	0	39	14 (35.9)	44	19 (43.2)
40-43	21	6 (28.6)	21	6 (28.6)	39	14 (35.9)
44-47	19	5 (26.3)	19	6 (31.6)	22	9 (40.9)
48-51	17	5 (29.4)	16	8 (50.0)	15	5 (33.3)
52-55	8	<5	16	5 (31.3)	20	9 (45.0)
56-59	6	<5	5	0	22	10 (45.5)
60+	<5	0	10	<5	13	<5
<16 years	283	56 (19.8)	300	101 (33.7)	322	121 (37.6)
≥16 years	432	143 (33.1)	482	217 (45.0)	441	236 (53.5)
<18 years	335	81 (24.2)	326	117 (35.9)	341	134 (39.3)
≥18 years	380	118 (31.1)	456	201 (44.1)	422	223 (52.8)
Overall	**715**	**199 (27.8)**	**782**	**318 (40.7)**	**763**	**357 (46.8)**

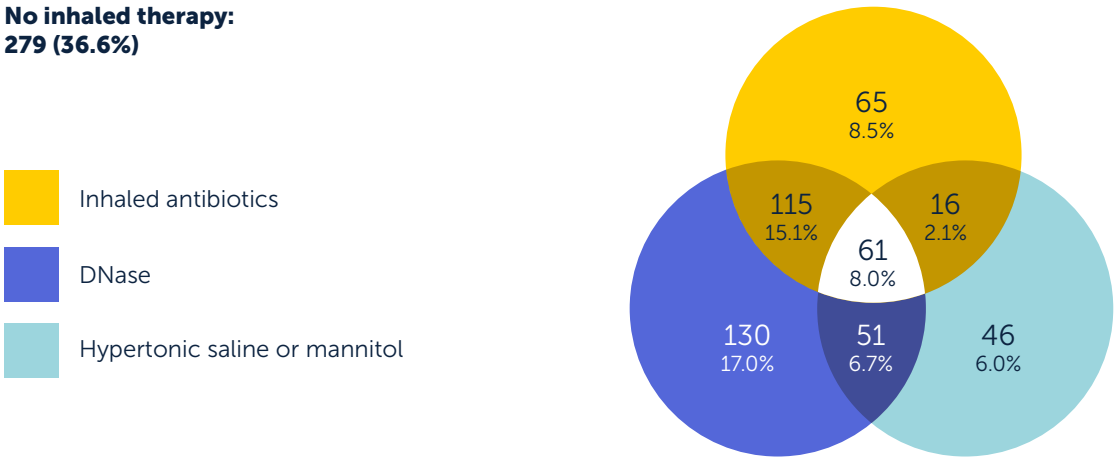
1.30 Hypertonic saline

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

	2011		2016		2021	
Age	Total patients	Patients on hypertonic saline; n (%)	Total patients	Patients on hypertonic saline; n (%)	Total patients	Patients on hypertonic saline; n (%)
0-3	69	0	54	<5	55	9 (16.4)
4-7	90	<5	91	8 (8.8)	74	15 (20.3)
8-11	58	7 (12.1)	86	20 (23.3)	96	31 (32.3)
12-15	66	10 (15.2)	69	27 (39.1)	97	38 (39.2)
16-19	98	12 (12.2)	66	17 (25.8)	37	12 (32.4)
20-23	84	7 (8.3)	97	27 (27.8)	51	12 (23.5)
24-27	69	9 (13.0)	79	18 (22.8)	67	25 (37.3)
28-31	52	<5	65	17 (26.2)	66	10 (15.2)
32-35	34	<5	49	5 (10.2)	45	6 (13.3)
36-39	21	<5	39	5 (12.8)	44	6 (13.6)
40-43	21	<5	21	<5	39	5 (12.8)
44-47	19	<5	19	<5	22	0
48-51	17	0	16	<5	15	<5
52-55	8	<5	16	<5	20	<5
56-59	6	0	5	0	22	<5
60+	<5	0	10	<5	13	0
<16 years	283	18 (6.4)	300	59 (19.7)	322	93 (28.9)
≥16 years	432	41 (9.5)	482	99 (20.5)	441	79 (17.9)
<18 years	335	25 (7.5)	326	67 (20.6)	341	99 (29.0)
≥18 years	380	34 (8.9)	456	91 (20.0)	422	73 (17.3)
Overall	**715**	**59 (8.3)**	**782**	**158 (20.2)**	**763**	**172 (22.5)**

1.31 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 279 (36.6%) people in Scotland are on no inhaled therapies.



Other therapies

1.32 CFTR modulators

In 2021, the CFTR modulators were available to the following people across the UK with cystic fibrosis under a managed access agreement:

Ivacaftor

In 2021, ivacaftor has approval for use for people aged four months and older with at least one copy of a CFTR 'gating' mutation, and for people aged six months and over with the R117H mutation.

Lumacaftor/ivacaftor

Lumacaftor/ivacaftor is licensed for use in the UK for patients aged two and over with two copies of the F508del mutation.

Tezacaftor/ivacaftor

Tezacaftor/ivacaftor is licenced for use in patients aged six and over who have two copies of the F508del mutation, or a single copy of F508del and one of 14 residual function mutations.

Elexacaftor/tezacaftor/ivacaftor

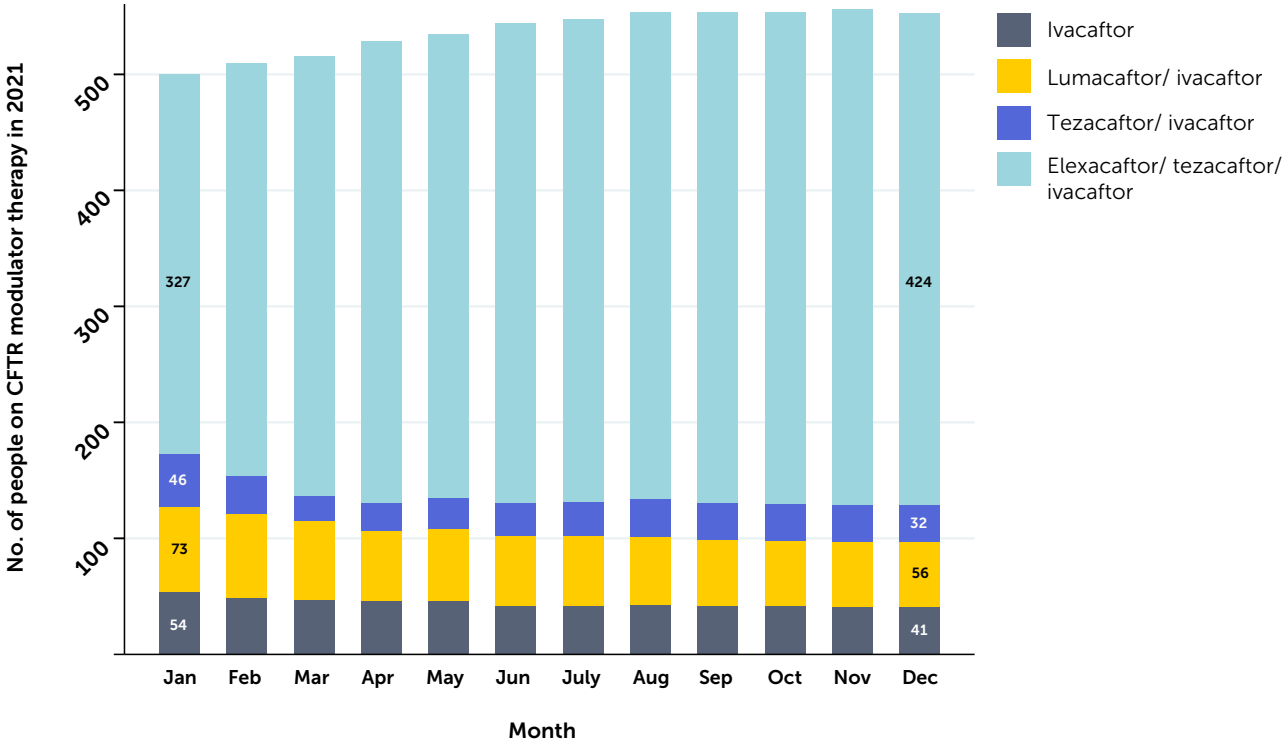
During 2021, Elexacaftor/tezacaftor/ ivacaftor was available in the UK for patients with cystic fibrosis aged 12 and over who have two copies of the F508del mutation, or a single copy of F508del and one minimal function mutation.

In January 2021 guidance was issued to clinicians supporting prescribing of a CFTR modulator to people with one copy of a F508del mutation but for whom the drug was not currently licensed.

The access arrangements prior to 2021 are described in previous annual reports.

CFTR modulator use in 2021

The graph below shows the number of people taking each drug by month. Where people switched modulators, the most recent prescription is counted. Only patients who had an annual review are counted. By December, 553 people were taking a CFTR modulator in Scotland.



1.33 Oxygen and non-invasive ventilation

	Overall (n=814)	<16 years (n=298)	≥16 years (n=516)	<18 years (n=321)	≥18 years (n=493)
Non-Invasive Ventilation (NIV); n (%)	<5	0 (0.0)	<5	0 (0.0)	<5
Any oxygen use; n (%)	23 (3.0)	<5	21 (4.6)	<5	21 (4.9)
Among those who had oxygen use:					
Continuously	<5	0 (0.0)	<5	0 (0.0)	<5
Nocturnal or with exertion	8 (34.8)	0 (0.0)	8 (38.1)	0 (0.0)	8 (38.1)
As required (PRN)	<5	0 (0.0)	<5	0 (0.0)	<5
With exacerbation	8 (34.8)	<5	6 (28.6)	<5	6 (28.6)

1.34 Physiotherapy

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

	Overall (n=814)	<16 years (n=298)	≥16 years (n=516)	<18 years (n=321)	≥18 years (n=493)
Active cycle of breathing techniques; n (%)	88 (11.3)	8 (2.5)	80 (17.7)	8 (2.3)	80 (18.5)
Autogenic drainage (including assisted autogenic drainage); n (%)	307 (39.5)	59 (18.2)	248 (54.9)	68 (19.7)	239 (55.3)
Postural drainage; n (%)	<5	0 (0.0)	<5	0 (0.0)	<5
Any form of PEP; n (%)	462 (59.5)	311 (95.7)	151 (33.4)	329 (95.4)	133 (30.8)
VEST; n (%)	<5	0 (0.0)	<5	0 (0.0)	<5
Exercise; n (%)	439 (56.5)	183 (56.3)	256 (56.6)	197 (57.1)	242 (56.0)
Other; n (%)	153 (19.7)	120 (36.9)	33 (7.3)	123 (35.7)	30 (6.9)

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

1.35 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=777)	<16 years (n=325)	≥16 years (n=452)	<18 years (n=345)	≥18 years (n=432)
Any supplemental feeding; n(%)	257 (33.1)	48 (14.8)	209 (46.2)	53 (15.4)	204 (47.2)
Nasogastric tube; n(%)	5 (0.6)	<5	<5	<5	<5
Gastrostomy tube/button; n(%)	20 (2.6)	11 (3.4)	9 (2.0)	12 (3.5)	8 (1.9)
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

1.36 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.

	2016	2017	2018	2019	2020	2021
Patients evaluated; n	18	22	19	21	11	5
Patients accepted; n	8	17	7	10	<5	<5
Patients receiving transplants; n	<5	<5	<5	5	0	<5
Bilateral lung	<5	<5	<5	<5	0	<5
Liver	0	0	<5	<5	0	0
Other	<5	<5	<5	0	0	0

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	920 (99.5)
Patients genotyped with both mutations recorded	911 (98.5)
F508del mutations	
Homozygous F508del	391 (42.3)
Heterozygous F508del	439 (47.5)

1.37 Mutation combinations in Scotland

This table shows the proportion (%) of patients with the most common mutation combinations. For example, 7.9% 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	6.7	0.1						6.8
G551D	7.9	0.1	0.2					8.2
G542X	5.4	0.2	0.1	0.1				5.8
621+1G->T	0.6	0.0	0.1	0.0	0.0			0.8
Other	26.1	1.2	1.5	1.0	0.3	4.5		34.6
Unknown	0.8	0.1	0.0	0.0	0.0	0.1	0.5	1.5
Total	**89.7**	**1.7**	**1.9**	**1.1**	**0.3**	**4.6**	**0.5**	**100.0**

* In this section, we include everyone who is registered (see table 1.1) and where mutations are available.

1.38 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	830	89.7
c.1652G->A	p.Gly551Asp	G551D	92	9.9
c.350G->A	p.Arg117His	R117H	78	8.4
c.1624G->T	p.Gly542X	G542X	63	6.8
c.200C->T	p.Pro67Leu	P67L	49	5.3
c.1679G->C	p.Arg560Thr	R560T	18	1.9
c.3454G->C	p.Asp1152His	D1152H	18	1.9
c.1585-1G->A		1717-1G->A	15	1.6
c.1477C->T	p.Gln493X	Q493X	13	1.4
c.2657+5G->A		2789+5G->A	13	1.4
c.3909C->G	p.Asn1303Lys	N1303K	12	1.3
c.489+1G->T		621+1G->T	10	1.1
c.3717+12191C->T		3849+10kbC->T	8	0.9
c.3528delC	p.Lys1177SerfsX15	3659delC	8	0.9
c.1364C->A	p.Ala455Glu	A455E	8	0.9
c.1558G->T	p.Val520Phe	V520F	8	0.9
c.2657+2_2657+3insA		2789+2insA	7	0.8
c.3140-26A->G		3272-26A->G	6	0.6
c.178G->T	p.Glu60X	E60X	6	0.6
c.948delT	p.Phe316LeufsX12	1078delT	6	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	1	0	1

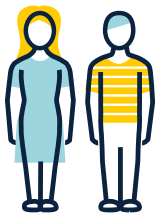
Section 2 shows analysis of data for individual CF centres. This allows people with CF, their families, and healthcare providers, to review a centre's use of some medications and outcome data alongside national averages. This transparency is intended to help improve standards of care overall.

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.

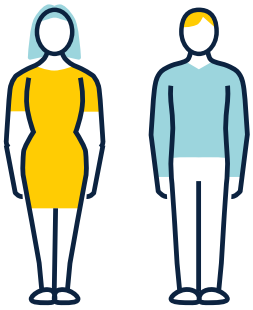
If a person with CF or a member of their family has questions about the results for their CF centre or clinic, they should discuss this with their CF team.

Full tables of the data are shown in appendix 1.

Key



Paediatric centre

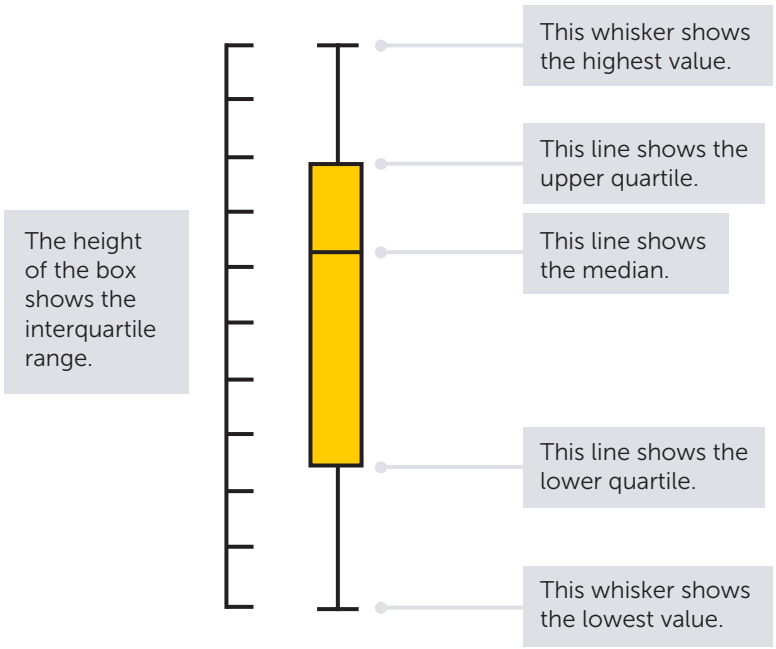


Adult centre

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.

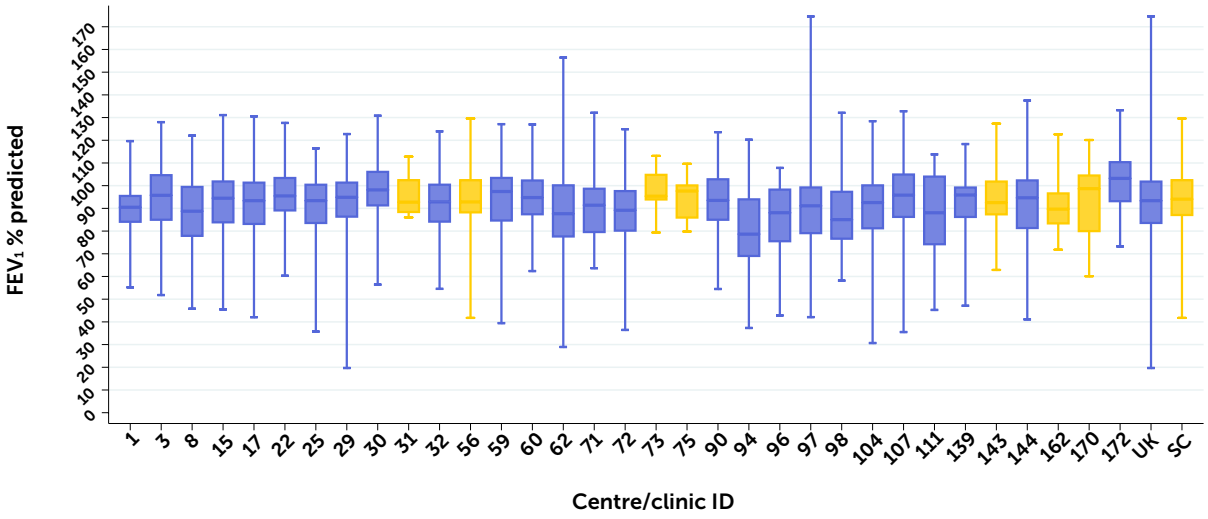
Section 2a: Paediatric centre analysis

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

Key

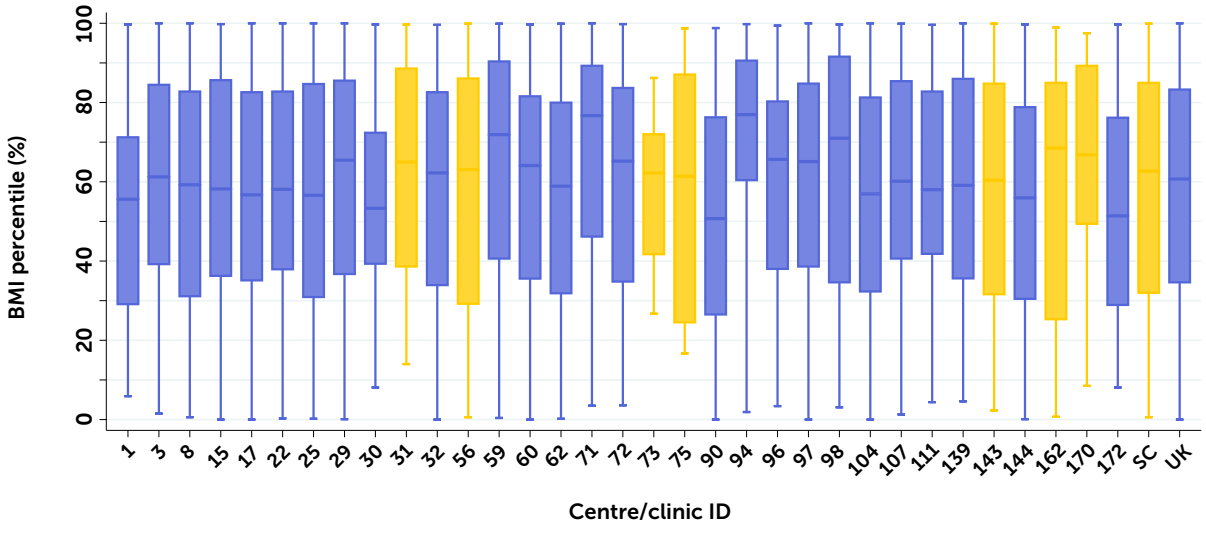
● Services in the UK ● Services in Scotland

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



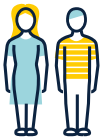
The mean FEV_1% predicted of patients attending paediatric centres/clinics in Scotland is 94.1% predicted (IQR: 86.9 - 102.6).

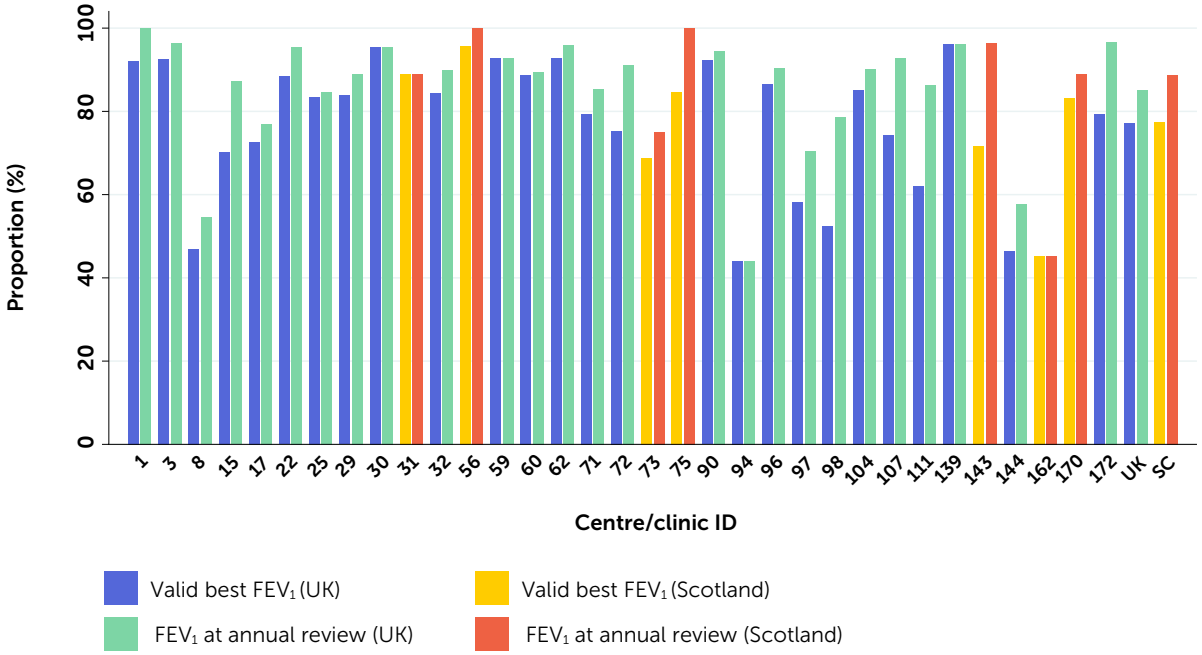
2.2 Body Mass Index (BMI) percentile among patients aged 2-15 years by paediatric centre/clinic



The median BMI percentile of patients attending paediatric centres/clinics in Scotland is 56.4 (IQR: 31.0-79.8).

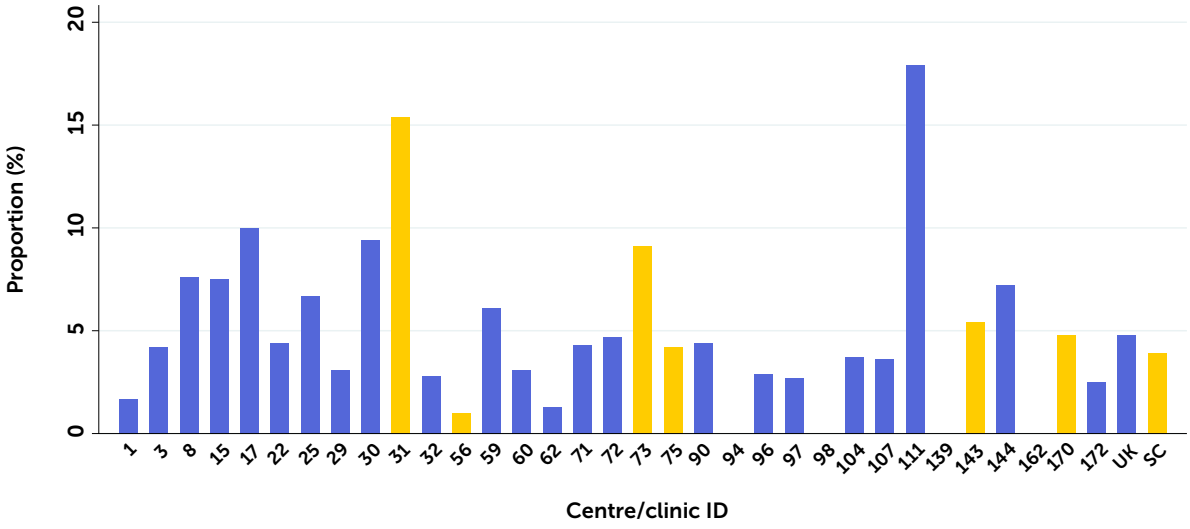
2.3 Data completeness by paediatric centre/clinic**



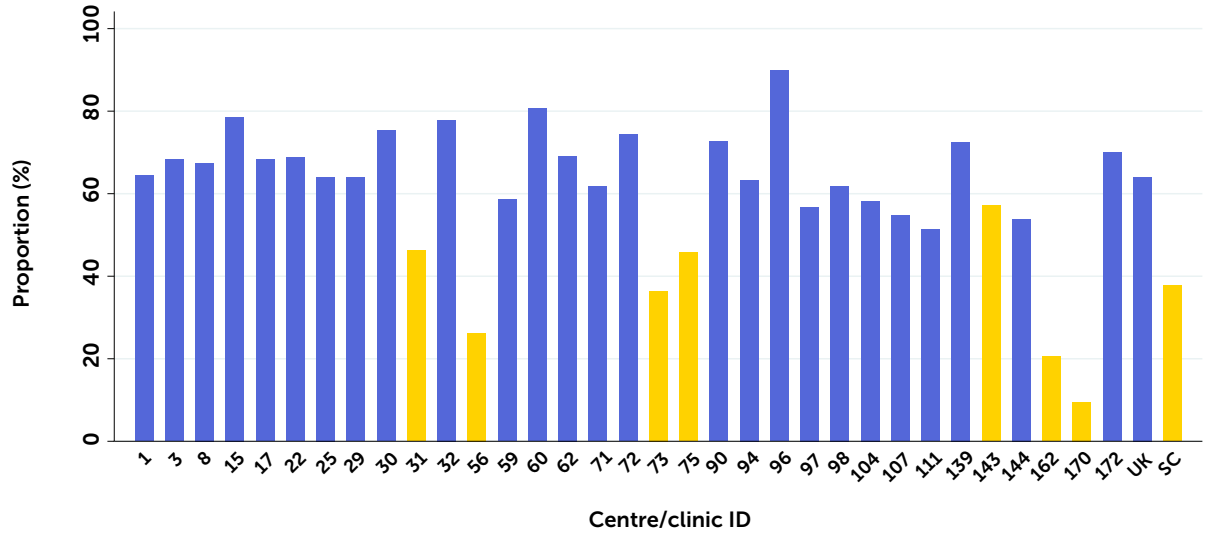


**The chart above shows the proportion of patients who had a valid best FEV_1% and an FEV_1% at annual review, excluding patients under six years of age. Best FEV_1% was considered valid if it was not missing, and the per cent predicted was not more than 0.5% lower than the annual review value. For some patients there may be medical reasons why FEV_1 could not be taken, so centres may not be able to get 100% completeness.

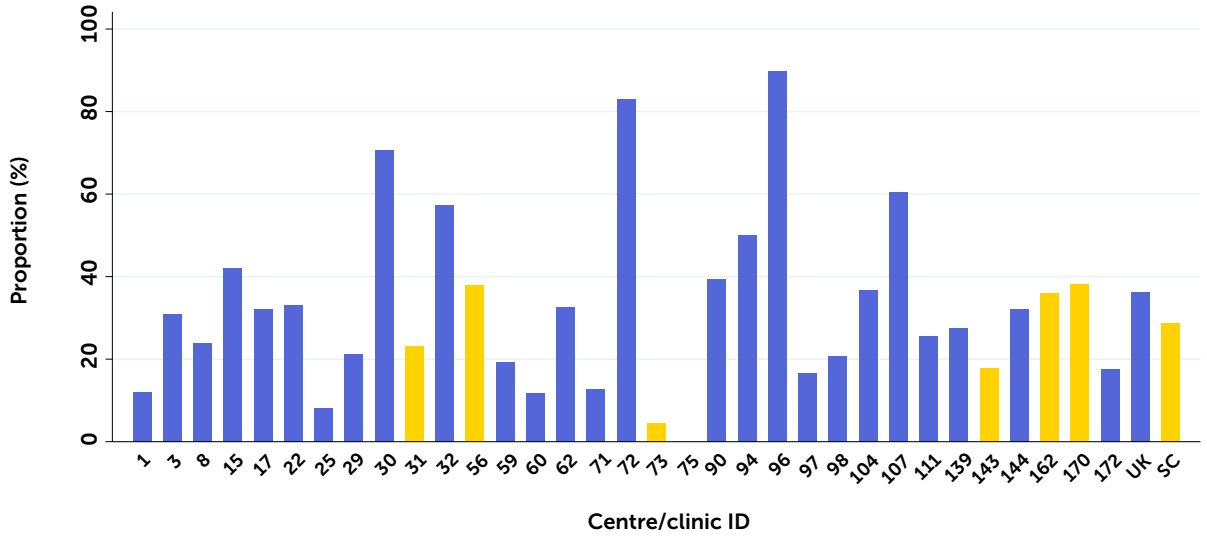
2.4 Proportion of patients with chronic *P. aeruginosa* by paediatric centre/clinic



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



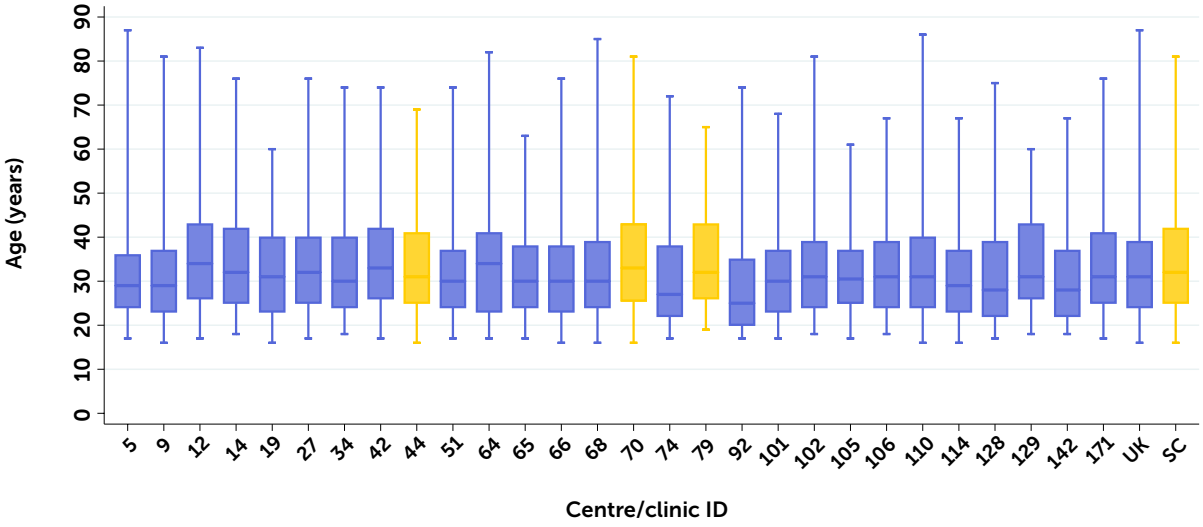
Section 2b: Adult centre analysis

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

Key

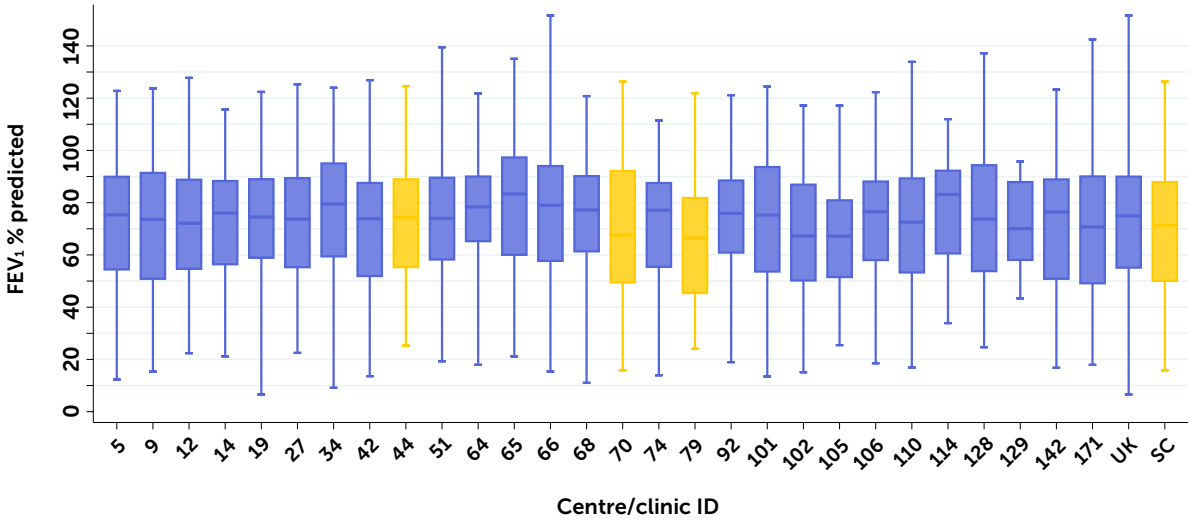
● Services in the UK ● Services in Scotland

2.7 Age distribution by adult centre/clinic



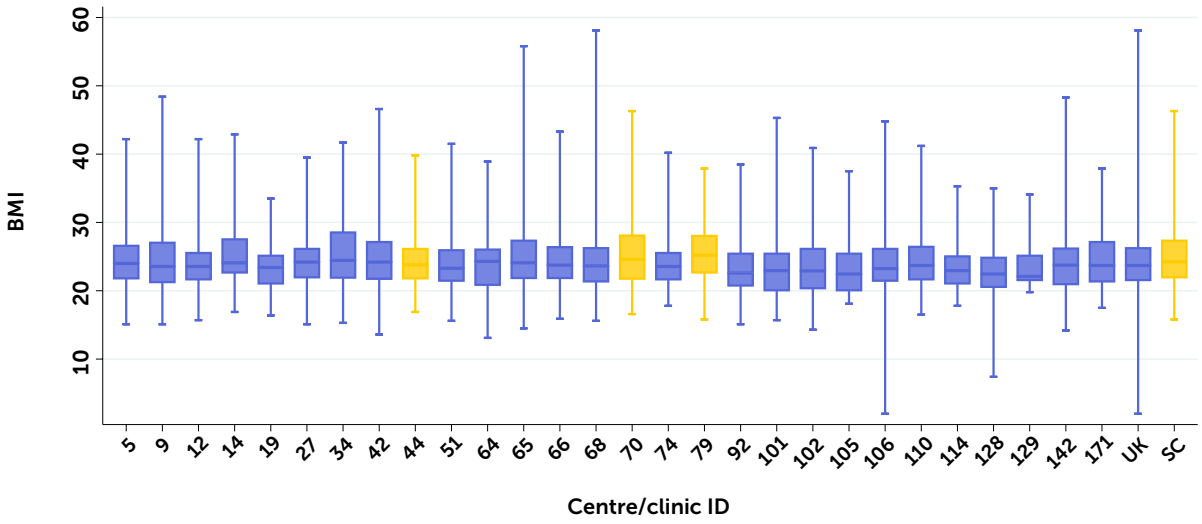
The median age of patients attending adult services in Scotland is 32 years (IQR: 25-42).

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



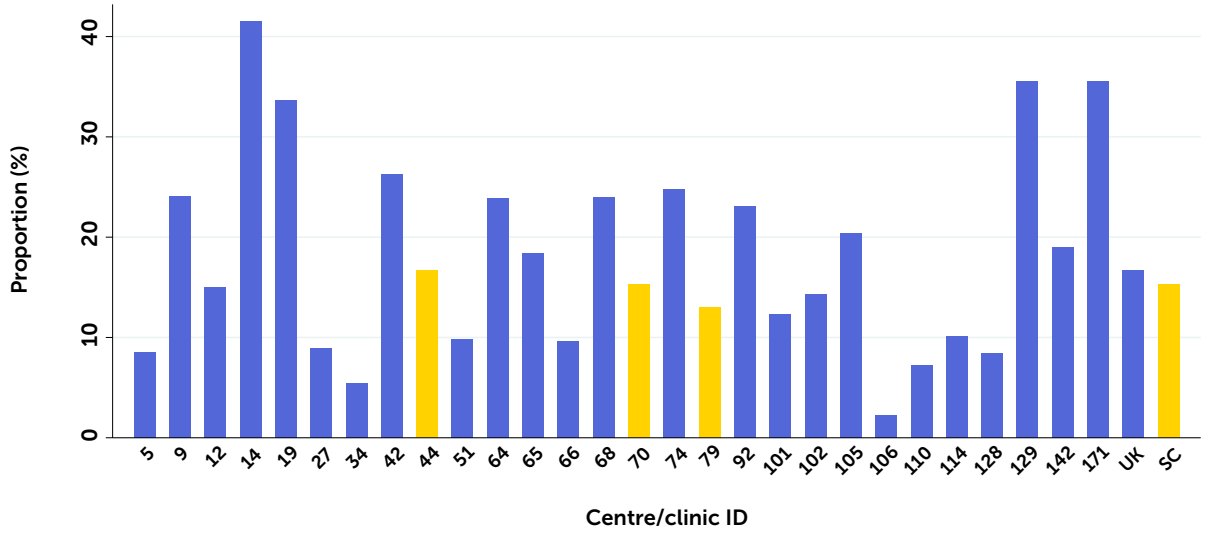
The median FEV_1 % predicted of patients attending adult services in Scotland is 71.3% (IQR 49.9-88.1).

2.9 Body Mass Index (BMI) distribution among patients aged 16 years and older by adult centre/clinic



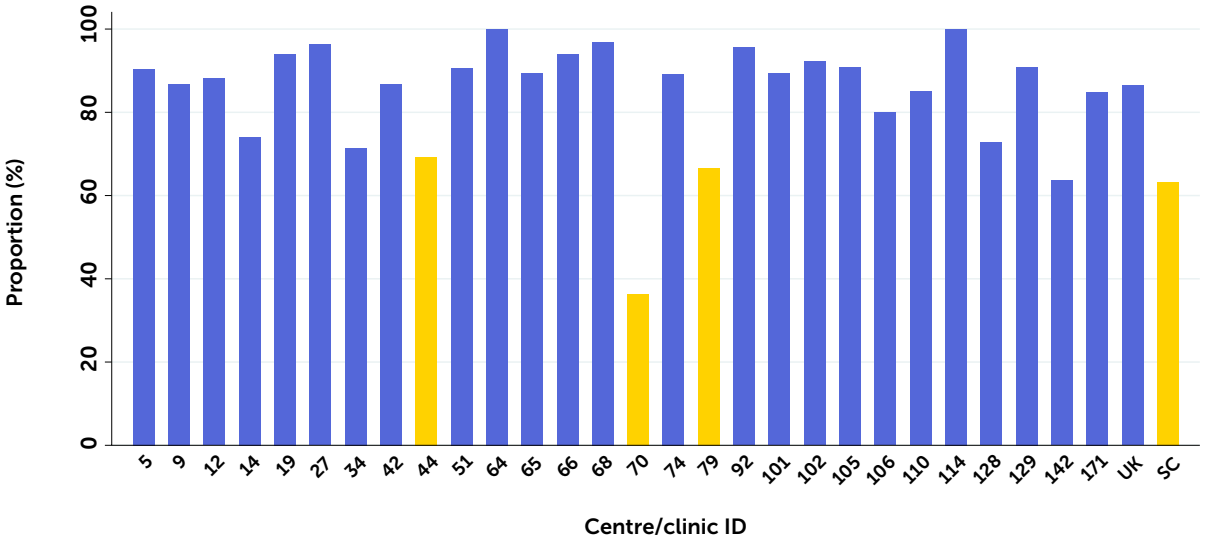
The median BMI of patients attending adult services in Scotland is 24.3 (IQR: 21.9-27.4).

2.10 Proportion of patients with chronic *Pseudomonas aeruginosa* by adult centre/clinic



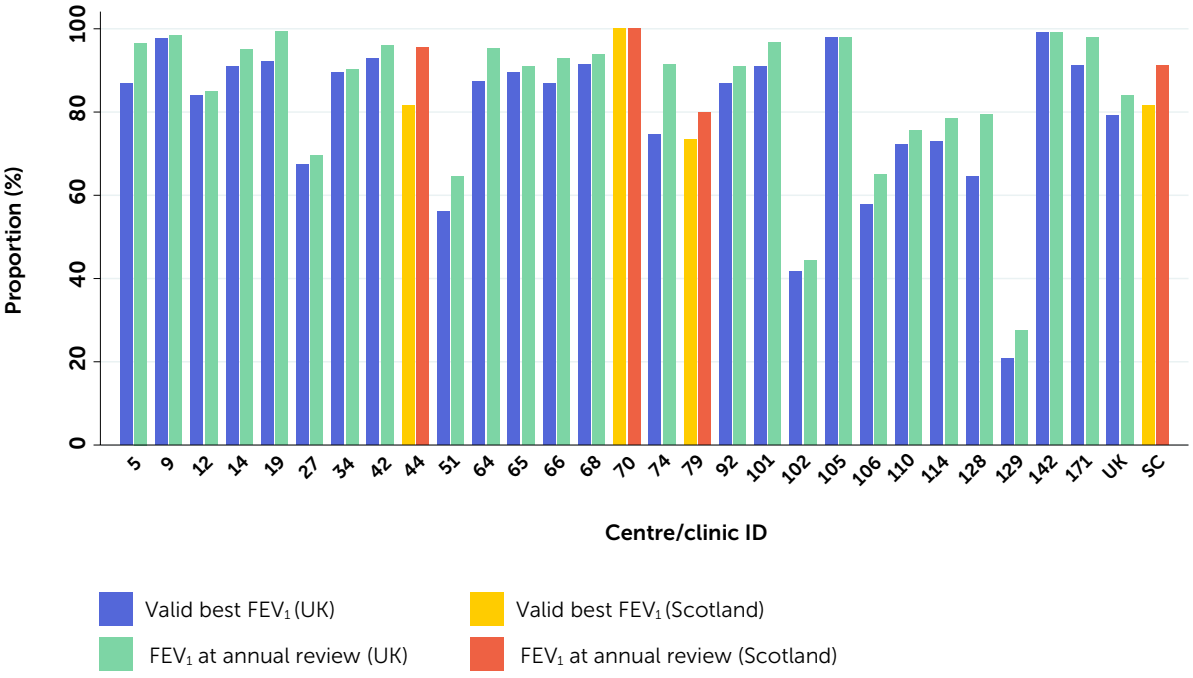
The proportion of patients with chronic *P.aeruginosa* attending adult services in Scotland is 15.1%.

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



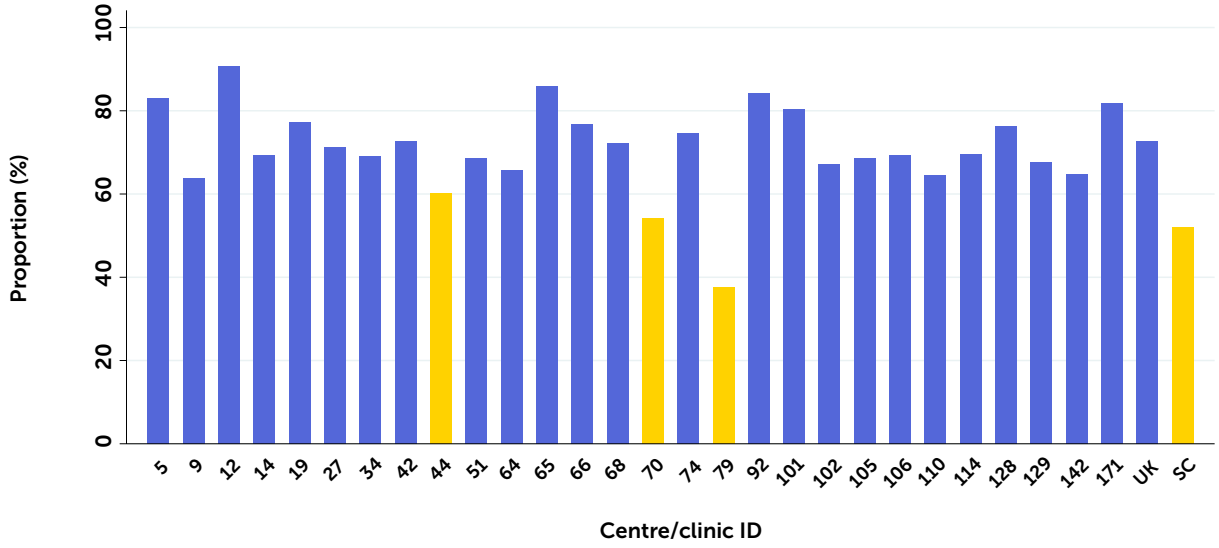
The proportion of chronic *P. aeruginosa* patients on inhaled antibiotics in Scotland is 63.2%.

2.12 Data completeness by adult centre/clinic*



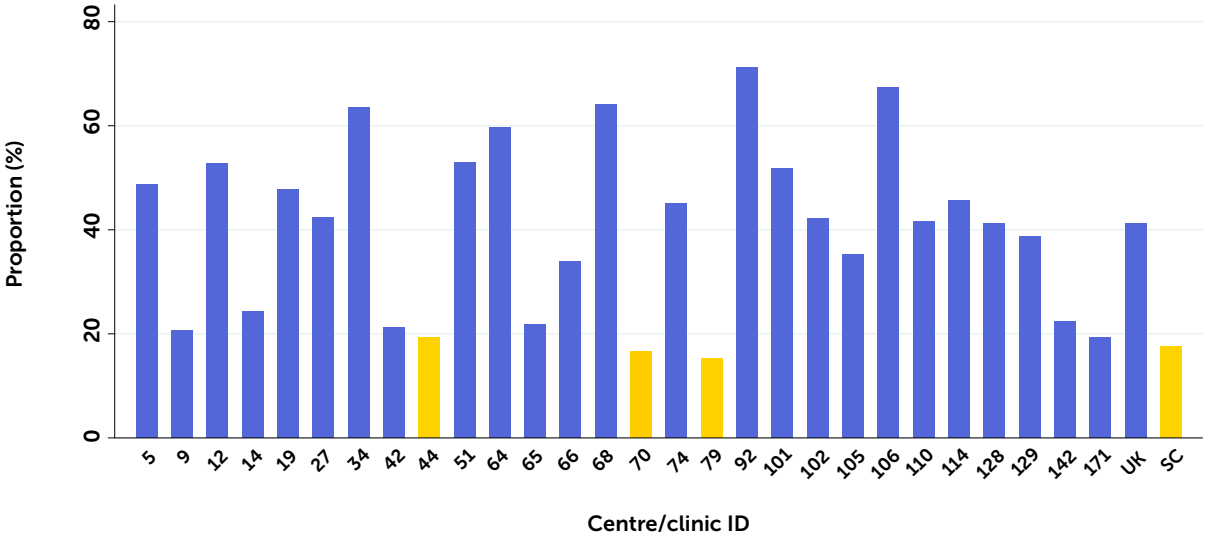
*FEV_1 was considered valid if it was not missing, and the percent predicted was not more than 0.5% lower than the annual review value. For some patients there may be medical reasons why FEV_1 could not be taken, so centres may not be able to get 100% completeness.

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 52.1%.

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



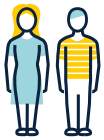
The proportion of patients attending adult services in Scotland receiving hypertonic saline or mannitol treatment is 17.6%.

Glossary

Word/Phrase	Meaning
2021	1 January 2021 – 31 December 2021.
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.
***Burkholderia cepacia* complex**	*B. cepacia* complex is a group of bacteria, some of which threaten the health of people with cystic fibrosis.
BMI (Body Mass Index)	A measure designed to show whether a person is a healthy weight for their height.
CF	Cystic fibrosis.
CFTR (cystic fibrosis transmembrane conductance regulator)	A protein at the cell surface that controls the salt and water balance across a cell. The gene that causes cystic fibrosis is the blueprint for the CFTR protein. Everyone has two copies of the gene for CFTR. To be born with cystic fibrosis, both CFTR genes must be affected by a CF-causing mutation.
Chronic	Persistent, or long-lasting.
Cirrhosis	A chronic liver disease.
CI (confidence interval)	A way of expressing how certain we are about our statistical estimates of a clinical measure (eg BMI). It gives a range of results that is likely to include the 'true' value for the population. A narrow confidence interval indicates a more precise estimate. A wide confidence interval indicates more uncertainty about the true value of the clinical measure - often because a small group of patients has been studied. The confidence interval is usually stated as '95% CI', which means that the range of values has a 95 in 100 chance of including the 'true' value.
Enzymes	Biological molecules that help complex reactions, such as digestion of food, occur in the body.
FEV_1 (forced expiratory volume in one second)	This is the amount of air that a person can blow out of the lungs in the first second of a forced exhaled breath. People with healthy lungs can blow out most of the air held in this time.
FEV_1% predicted	The FEV_1 can be converted from absolute litres of air blown out into a predicted percentage (%). A healthy range for % predicted is calculated from a very large population sample, and is normally considered to be between 80-120% predicted.
Fibrosing colonopathy	A condition causing narrowing of part of the colon.
Gall bladder	The small sac-shaped organ under the liver that stores bile after it is secreted by the liver, before it is released into the intestine.
Gastrointestinal (GI) tract	The GI tract is an organ system responsible for digesting food, absorbing nutrients and expelling waste.
Genotype	Part of the genetic makeup of a cell, organism or individual that usually controls a particular characteristic (known as a phenotype).
GORD (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, which 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.

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.
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 membrane caused by chronic inflammation of the nasal lining.
NBS (newborn screening)	Newborn screening is part of the heel prick blood spot testing carried out on all babies at 5-7 days of age. The blood sample is tested for a number of conditions, including cystic fibrosis.
NTM (non tuberculous 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

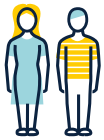


Paediatric centres/clinics providing data in 2021 – ordered alphabetically by location

Location	Name	Clinic ID	Total Active	Number with annual review
Scotland				
Aberdeen	Royal Aberdeen Children's Hospital	75	26	24
Ayr	University Hospital Crosshouse	170	23	21
Dundee	Ninewells Hospital	73	23	22
Edinburgh	Royal Hospital for Sick Children	143	137	112
Glasgow	Royal Hospital for Sick Children	56	164	142
Inverness	Raigmore Hospital	31	16	13

Paediatric centres/clinics providing data in 2021 – ordered alphabetically by location

			BMI			
Location	Name	Clinic ID	Number	Mean - unadjusted	Mean - adjusted	Median
Scotland						
Aberdeen	Royal Aberdeen Children's Hospital	75	23	57.3	57.3	61.4
Ayr	University Hospital Crosshouse	170	20	70.0	70.0	74.5
Dundee	Ninewells Hospital	73	16	57.7	57.7	62.2
Edinburgh	Royal Hospital for Sick Children	143	102	57.9	57.9	60.3
Glasgow	Royal Hospital for Sick Children	56	118	56.5	56.5	63.1
Inverness	Raigmore Hospital	31	11	63.9	63.9	65.0



	Age		FEV$_1$% predicted at annual review				Best FEV$_1$% predicted			
Clinic ID	Mean	Median	Number	Mean - unadjusted	Mean - adjusted	Median	Number**	Mean - unadjusted	Mean - adjusted	Median
75	7.8	7.9	13	95.0	95.0	97.6	13	101.5	101.3	100.3
170	10.6	10.1	16	92.9	92.9	98.7	17	102.1	102.0	103.3
73	8.7	9.7	12	97.8	97.6	95.4	12	99.9	99.7	99.7
143	9.5	10.0	78	93.9	93.9	92.5	79	96.7	96.7	96.9
56	9.1	9.8	84	93.9	93.8	92.7	101	99.2	99.1	98.4
31	8.7	7.9	8	95.7	95.7	92.7	8	98.7	98.6	97.1

	Chronic *pseudomonas*		Having at least 1 IV days		Receiving DNase treatment		Receiving hypertonic saline or mannitol treatment		Inhaled antibiotic use among patients with chronic *Pseudomonas*	
Clinic ID	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)
75	<5	-*	7	29.2	11	45.8	<5	-*	<5	100.0
170	<5	-*	<5	-*	<5	-*	8	38.1	<5	100.0
73	<5	-*	<5	-*	8	36.4	<5	-*	<5	50.0
143	6	5.4	26	23.2	64	57.1	19	17.0	6	100.0
56	<5	-*	27	19.0	35	24.6	60	42.3	0	0.0
31	<5	-*	<5	7.7	6	46.2	<5	-*	<5	100.0

* Redacted to adhere to statistical disclosure guidelines.

** Where 'Best' values were missing, or lower than FEV$_1$% predicted taken at annual review, the annual review value was used.

Appendix 1: Centre-level data tables

Adult centres/clinics providing data in 2021 – ordered alphabetically by location

Location	Name	Clinic ID	Total active	Number with annual review
Scotland				
Aberdeen	Aberdeen Royal Infirmary	70	74	72
Edinburgh	Western General Hospital	44	243	233
Glasgow	Queen Elizabeth University Hospital	79	219	138

Adult centres/clinics providing data in 2021 – ordered alphabetically by location

			BMI			
Location	Name	Clinic ID	Number	Mean - unadjusted	Mean - adjusted	Median
Scotland						
Aberdeen	Aberdeen Royal Infirmary	70	72	25.7	25.6	24.6
Edinburgh	Western General Hospital	44	230	24.4	24.4	23.9
Glasgow	Queen Elizabeth University Hospital	79	129	25.2	25.1	24.9

	Age		FEV$_1$% predicted at annual review				Best FEV$_1$% predicted			
Clinic ID	Mean	Median	Number	Mean - unadjusted	Mean - adjusted	Median	Number**	Mean - unadjusted	Mean - adjusted	Median
70	35.1	33.3	63	70.7	71.0	68.7	63	73.0	73.2	73.0
44	34.7	31.9	206	72.7	72.8	73.6	213	75.1	75.3	78.0
79	35.8	32.5	108	65.4	67.3	66.7	130	69.3	71.0	71.0

	Chronic *pseudomonas*		Having at least 1 IV days		Receiving DNase treatment		Receiving hypertonic saline or mannitol treatment		Inhaled antibiotic use among patients with chronic *pseudomonas*	
Clinic ID	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)
70	11	15.3	5	6.9	39	54.2	12	16.7	<5	-*
44	39	16.7	49	21.0	140	60.1	45	19.3	28	71.8
79	18	13.0	35	25.4	52	37.7	21	15.2	13	72.2

* Redacted to adhere to statistical disclosure guidelines.

** Where 'Best' values were missing, or lower than FEV$_1$% predicted taken at annual review, the annual review value was used.

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	830	89.7
c.1652G->A	p.Gly551Asp	G551D	92	9.9
c.350G->A	p.Arg117His	R117H	78	8.4
c.1624G->T	p.Gly542X	G542X	63	6.8
c.200C->T	p.Pro67Leu	P67L	49	5.3
c.1679G->C	p.Arg560Thr	R560T	18	1.9
c.3454G->C	p.Asp1152His	D1152H	18	1.9
c.1585-1G->A		1717-1G->A	15	1.6
c.1477C->T	p.Gln493X	Q493X	13	1.4
c.2657+5G->A		2789+5G->A	13	1.4
c.3909C->G	p.Asn1303Lys	N1303K	12	1.3
c.489+1G->T		621+1G->T	10	1.1
c.3717+12191C->T		3849+10kbC->T	8	0.9
c.3528delC	p.Lys1177SerfsX15	3659delC	8	0.9
c.1364C->A	p.Ala455Glu	A455E	8	0.9
c.1558G->T	p.Val520Phe	V520F	8	0.9
c.2657+2_2657+3insA		2789+2insA	7	0.8
c.3140-26A->G		3272-26A->G	6	0.6
c.178G->T	p.Glu60X	E60X	6	0.6
c.948delT	p.Phe316LeufsX12	1078delT	6	0.6
c.1210-12[5] (AJ574948.1:g.152T[5])		5T	5	0.5
c.3846G->A	p.Trp1282X	W1282X	<5	-
c.1721C->A	p.Pro574His	P574H	<5	-
c.1766+1G->A		1898+1G->A	<5	-
c.1523T->G	p.Phe508Cys	F508C	<5	-
c.3196C->T	p.Arg1066Cys	R1066C	<5	-
c.1705T->G	p.Tyr569Asp	Y569D	<5	-
c.2012delT	p.Leu671X	2143delT	<5	-
c.579+3A->G		711+3A->G	<5	-
c.3468G->A		3600G->A	<5	-
c.1519_1521delATC	p.Ile507del	I507del	<5	-
c.2988G->A		3120G->A	<5	-
c.2052delA	p.Lys684AsnfsX38	2184delA	<5	-
c.2988+1G->A		3120+1G->A	<5	-
c.223C->T	p.Arg75X	R75X	<5	-
c.2490+1G->A		2622+1G->A	<5	-
c.164+2T>C		296+2T->C	<5	-
c.254G->A	p.Gly85Glu	G85E	<5	-
c.1680A->C	p.Arg560Ser	R560S	<5	-

Nucleotide	Protein	Legacy name	N	%
c.3884_3885insT	p.Ser1297PhefsX5	4016insT	<5	-
c.1367T->C	p.Val456Ala	V456A	<5	-
c.274G->A	p.Glu92Lys	E92K	<5	-
c.3484C->T	p.Arg1162X	R1162X	<5	-
c.3737C->T	p.Thr1246Ile	T1246I	<5	-
c.3476C->T	p.Ser1159Phe	S1159F	<5	-
c.2859_2890delACATTCTGTT CTTCAAGCACCTATGTCAACCC	p.Leu953PhefsX11	2991del32	<5	-
c.3266G->A	p.Trp1089X	W1089X	<5	-
c.1466C->A	p.Ser489X	S489X	<5	-
c.1040G->C	p.Arg347Pro	R347P	<5	-
c.54-5940_273+10250del21kb	p.Ser18ArgfsX16	CFTRdele2,3	<5	-
c.1006_1007insG	p.Ile336SerfsX28	1138insG	<5	-
c.3276C->A	p.Tyr1092X	Y1092X(C->A)	<5	-
c.3705T->G	p.Ser1235Arg	S1235R	<5	-
c.1055G->A	p.Arg352Gln	R352Q	<5	-
c.273+1G->A		405+1G->A	<5	-
c.509G->A	p.Arg170His	R170H	<5	-
c.1657C->T	p.Arg553X	R553X	<5	-
c.2583delT	p.Phe861LeufsX3	2711delT	<5	-
c.1753G->T	p.Glu585X	E585X	<5	-
c.4004T->C	p.Leu1335Pro	L1335P	<5	-
c.1327G->T	p.Asp443Tyr	D443Y	<5	-
c.2051_2052delAAinsG	p.Lys684SerfsX38	2183AA->G or 2183delAA->G	<5	-
c.1585-8G->A		1717-8G->A	<5	-
c.443T->C	p.Ile148Thr	I148T	<5	-
c.349C->G	p.Arg117Gly	R117G	<5	-
c.1000C->T	p.Arg334Trp	R334W	<5	-
c.349C->T	p.Arg117Cys	R117C	<5	-
c.[1210–12[5];1210-34TG[13]]		5T;TG13	<5	-
c.3158C->T	p.Thr1053Ile	T1053I	<5	-
c.292C->T	p.Gln98X	Q98X	<5	-
c.4147_4148insA	p.Ile1383AsnfsX3	4279insA	<5	-
c.1022_1023insTC	p.Phe342HisfsX28	1154insTC	<5	-
c.3208C->T	p.Arg1070Trp	R1070W	<5	-
c.1209+1G->A		1341+1G->A	<5	-
`Other' selected			14	1.5

Cystic Fibrosis Trust

Cystic Fibrosis Trust is the charity uniting people to stop cystic fibrosis. Our community will improve care, speak out, support each other and fund vital research as we race towards effective treatments for all.

We won't stop until everyone can live without the limits of cystic fibrosis.

cysticfibrosis.org.uk

Uniting for a life unlimited