Cystic Fibrosis strength in numbers

UK Cystic Fibrosis Registry
Annual Data Report 2017

Scotland

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UK Cystic Fibrosis Registry 2017 Annual Data Report - Scotland

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Acknowledgements

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

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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 **53**.

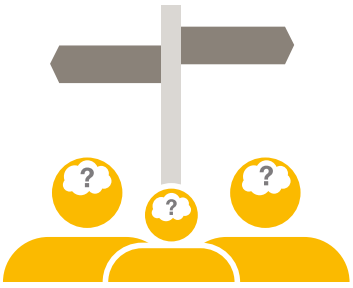
Cystic fibrosis

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

UK Cystic Fibrosis Registry

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

The purpose of the UK CF Registry is to improve the health of people with cystic fibrosis. This is done in a number of ways:

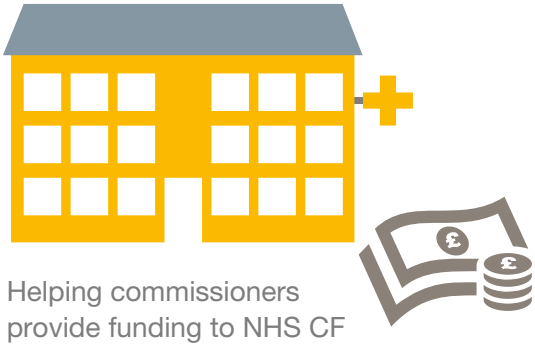


Helping people with CF and their families understand CF, and make informed decisions.

Giving clinical teams the evidence they need to improve the quality of care.

Monitoring the safety and effectiveness of new treatments for cystic fibrosis.

Providing data for research to find out the best ways of treating and beating cystic fibrosis.



Helping commissioners provide funding to NHS CF centres that is proportionate to their patients' disease severity.

Governance

The Registry Steering Committee (RSC) is responsible for making sure that the UK CF Registry is compliant with data protection legislation, and its Research Ethics Committee-approved Study Protocol. It also makes recommendations about the future development of the Registry. A sub-committee of the RSC, the Registry Research Committee, assesses applications for data and guides the Registry research strategy.

Please see Appendix 1: UK CF Registry Committee Structure.

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

When data are provided to third parties such as the NHS or university researchers, they are either anonymised (all identifiable data removed completely) or pseudonymised (all identifiable data replaced with a unique identification number). Pseudonymisation is used so that data can be traced back to what is in the 'live' database by the Registry team for the purposes of updating the data or answering queries. This means that the Registry data used for research, and the results that come from it, cannot identify the people whose data are stored on the UK CF Registry.

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

Data collection

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

Where can I find more information?

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

Section 1: UK-wide analysis

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

1.1 Summary of the UK Cystic Fibrosis Registry

	2017	
	UK	**Scotland**
CF patients registered[1]	10,469	923
Excluding diagnoses that year	10,255	915
CF patients with an annual review; n(%)[2]	9,887 (96%)	858 (93%)
Age in years; median[3]	20	20
All newly diagnosed patients (newborn screening and other)[4]	214	8
Number of patients born identified by newborn screening[4]	172	7
Age at diagnosis in months; median[3]	2	3
Adults aged 16 years and over; %[3]	60.6	61.4
Males; %[3]	53.3	54.2
Genotyped; %[3] **(both mutations identified)**	98.4	99.4
Total deaths reported (%)[5]	132 (1.3%)	19 (2.1%)
Age at death in years; median (95% CI)[5]	31 (29, 35)	32 (27, 35)

Notes:

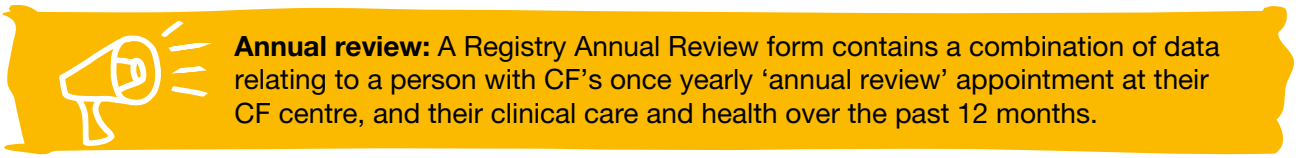
[1] Number of patients diagnosed with CF, seen in the past two years, and alive at 1 January in the given year. This number reduced in 2016 as a result of a data cleaning exercise. We followed up on patients who were registered but did not have data submitted in 2016. If they were no longer being cared for within the NHS (eg they had moved abroad), they were marked as 'inactive' and excluded from this number.

[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. Some diagnosis data are added after the data entry closure each year, so figures from previous years have been updated for this report.

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

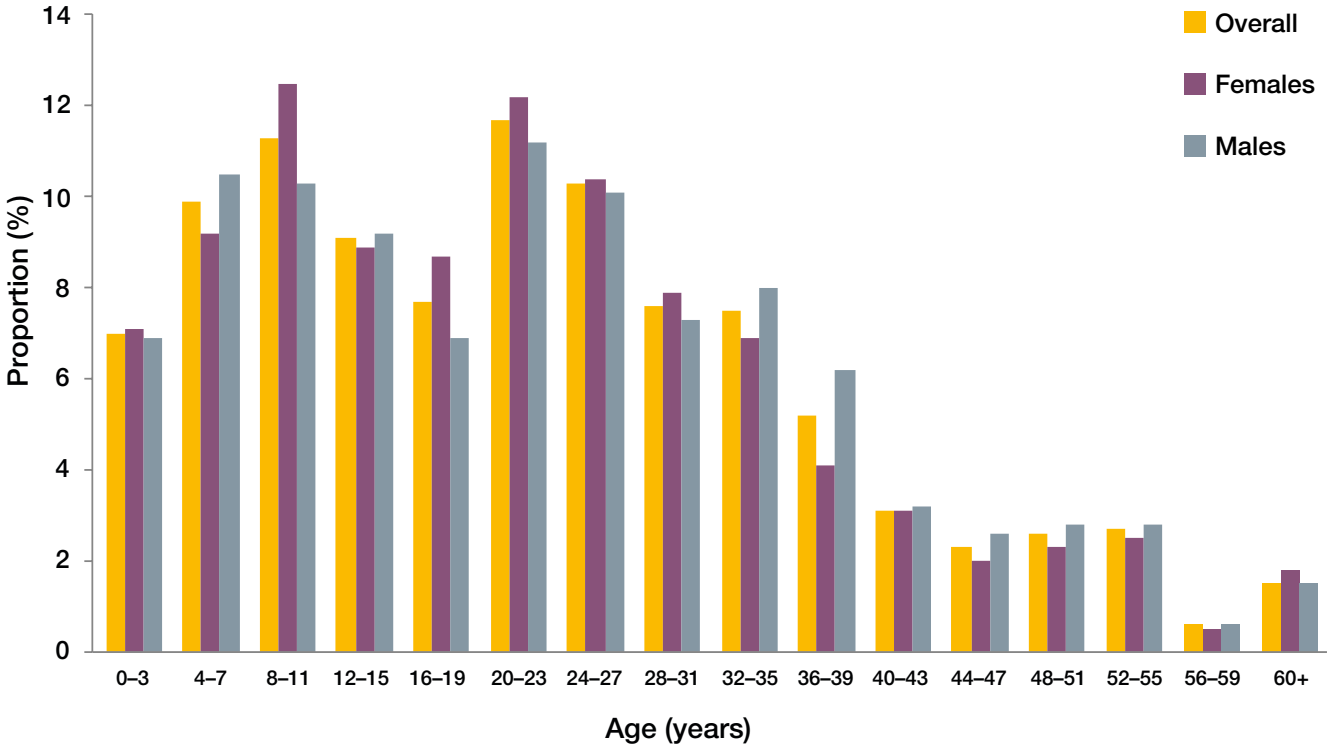


Annual review: A Registry Annual Review form contains a combination of data relating to a person with CF's once yearly 'annual review' appointment at their CF centre, and their clinical care and health over the past 12 months.

1.2 Age distribution by gender

N=858

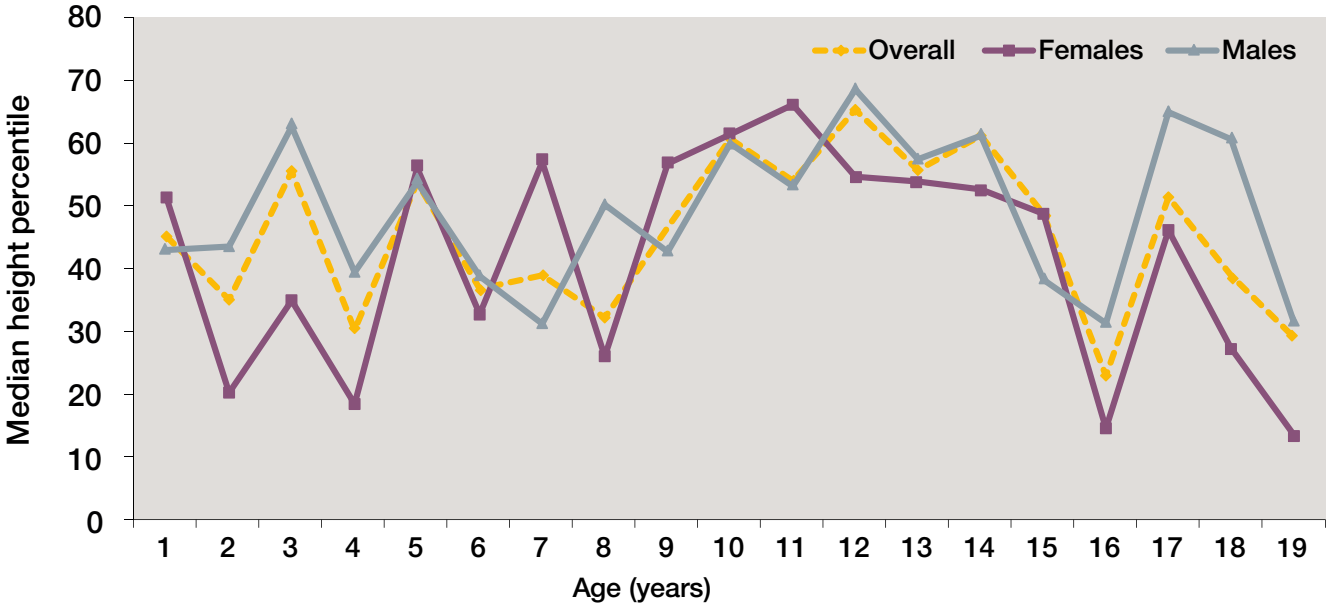
The following chart shows the mix of ages and genders in the CF population in Scotland.



Age	All; n (%)	Females; n (%)	Males; n (%)
0-3	60 (7.0)	28 (7.1)	32 (6.9)
4-7	85 (9.9)	36 (9.2)	49 (10.5)
8-11	97 (11.3)	49 (12.5)	48 (10.3)
12-15	78 (9.1)	35 (8.9)	43 (9.2)
16-19	66 (7.7)	34 (8.7)	32 (6.9)
20-23	100 (11.7)	48 (12.2)	52 (11.2)
24-27	88 (10.3)	41 (10.4)	47 (10.1)
28-31	65 (7.6)	31 (7.9)	34 (7.3)
32-35	64 (7.5)	27 (6.9)	37 (8.0)
36-39	45 (5.2)	16 (4.1)	29 (6.2)
40-43	27 (3.1)	12 (3.1)	15 (3.2)
44-47	20 (2.3)	8 (2.0)	12 (2.6)
48-51	22 (2.6)	9 (2.3)	13 (2.8)
52-55	23 (2.7)	10 (2.5)	13 (2.8)
56-59	5 (0.6)	<5	<5
60+	13 (1.5)	7 (1.8)	6 (1.3)
<16	320 (37.3)	148 (37.7)	172 (37.0)
≥16	538 (62.7)	245 (62.3)	293 (63.0)
<18	348 (40.6)	166 (42.2)	182 (39.1)
≥18	510 (59.4)	227 (57.8)	283 (60.9)
Overall	858	-	-

1.3 Height percentiles of children and young people (<20 years)[6] N=386

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.



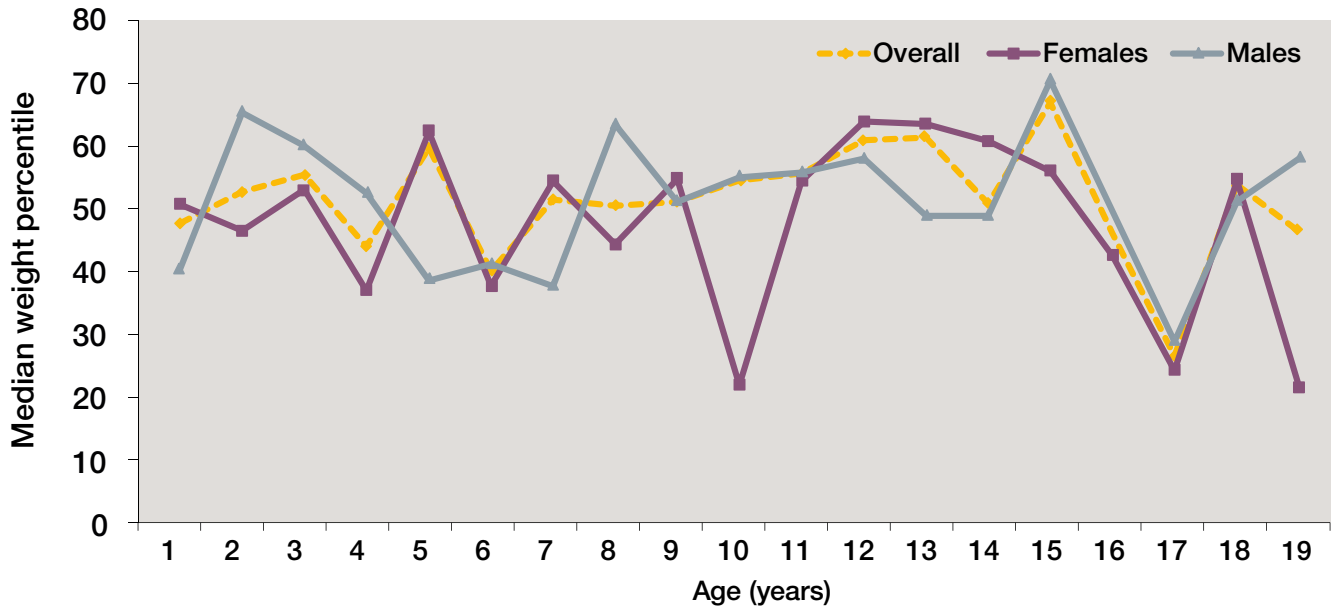
	Overall			Female			Male		
Age	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**
1	17	45.2	29.0-60.1	7	51.3	25.7-62.8	10	43.1	29.0-60.1
2	18	35.2	20.3-64.5	9	20.3	10.9-51.5	9	43.6	31.7-82.6
3	17	55.6	31.8-72.6	7	35.0	22.2-56.0	10	62.7	38.1-76.1
4	14	30.3	15.2-62.5	7	18.5	15.2-69.1	7	39.5	6.8-62.5
5	23	53.8	17.7-70.7	12	56.1	30.2-70.9	11	53.8	10.7-70.6
6	30	36.8	19.3-53.1	12	32.8	18.8-43.4	18	38.8	19.3-61.0
7	18	39.0	16.3-57.6	5	57.1	38.8-74.4	13	31.3	6.8-53.9
8	31	32.3	11.4-58.5	19	26.1	12.1-50.2	12	50.3	8.4-77.1
9	15	46.7	21.4-68.6	6	56.8	22.1-68.6	9	42.8	18.7-63.0
10	17	60.8	9.3-73.6	7	61.5	5.9-83.0	10	60.0	31.1-73.6
11	34	54.1	20.0-79.3	17	66.2	19.1-79.3	17	53.2	35.5-79.2
12	21	65.4	36.5-78.3	9	54.7	21.2-80.7	12	68.7	45.8-77.9
13	26	55.8	17.5-75.7	13	54.0	13.4-75.7	13	57.5	25.5-70.5
14	18	61.3	39.9-81.3	6	52.7	8.8-72.7	12	61.3	42.0-81.9
15	13	48.8	14.9-62.1	7	48.8	14.9-91.1	6	38.3	13.4-62.1
16	14	23.0	11.2-60.9	≥5	14.6	11.2-60.9	<5	-	-
17	14	51.5	28.1-69.8	7	46.2	27.7-69.8	7	65.0	49.8-76.7
18	20	39.0	16.7-82.1	9	27.4	17.8-59.0	11	60.7	15.6-83.6
19	18	29.2	13.6-48.3	7	13.6	13.6-71.1	11	31.6	18.2-48.3
Overall	**378***	**45.7**	**17.8-70.6**	**-**	**38.8**	**14.9-68.6**	**-**	**49.8**	**25.6-73.6**

*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=386

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.



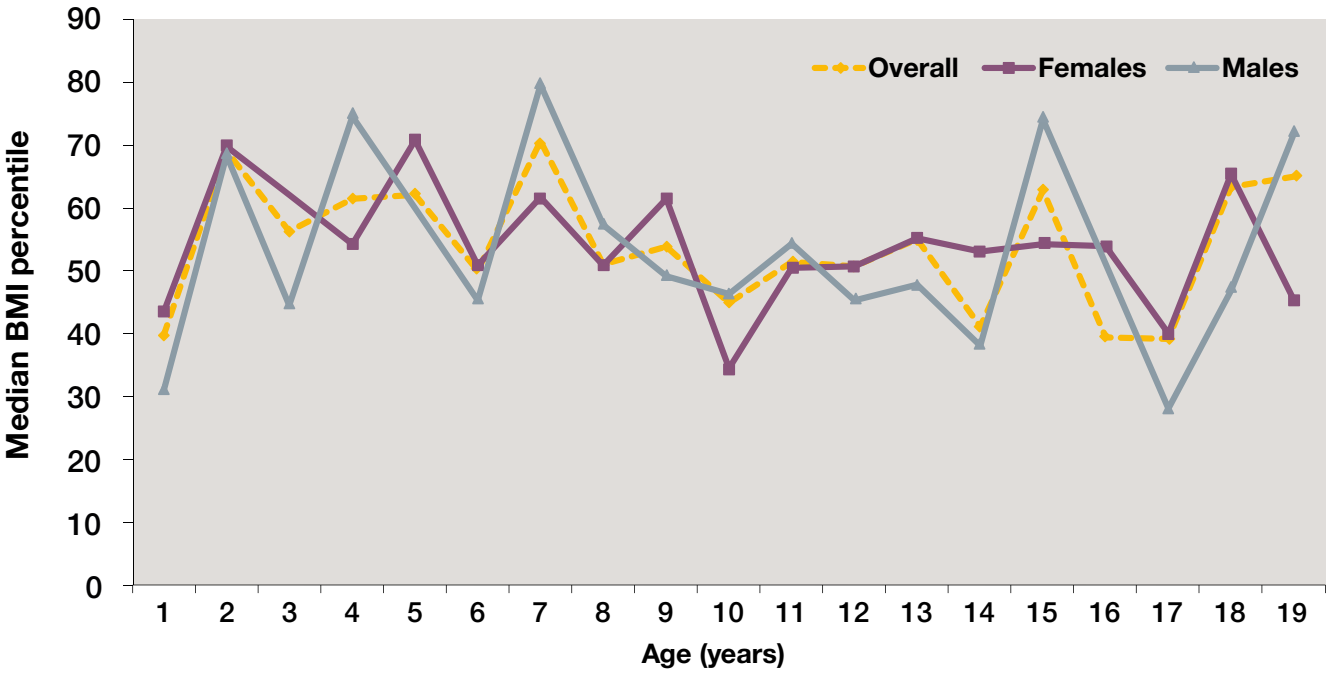
	Overall			Female			Male		
Age	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**
1	17	47.6	21.9-61.6	7	50.6	14.3-56.5	10	40.3	21.9-65.5
2	19	52.6	24.0-84.9	10	46.5	17.0-52.6	9	65.2	60.2-84.9
3	17	55.3	39.5-67.6	7	53.0	30.0-59.2	10	59.9	39.5-68.7
4	14	44.0	20.6-63.8	7	36.9	6.2-63.8	7	52.4	20.6-74.6
5	23	59.6	18.9-77.5	12	62.1	34.3-76.7	11	38.5	13.7-80.8
6	30	40.1	23.0-61.7	12	37.7	14.8-58.0	18	41.2	27.5-61.7
7	18	51.3	22.7-90.0	5	54.3	53.6-92.2	13	37.6	22.7-77.5
8	31	50.4	20.6-66.7	19	44.3	17.3-61.3	12	63.3	31.3-69.8
9	15	51.0	25.6-85.0	6	54.7	16.0-85.0	9	51.0	27.2-74.5
10	17	54.4	19.4-65.4	7	21.9	7.5-89.9	10	55.0	35.9-65.4
11	34	55.6	33.5-70.4	17	54.3	33.5-65.4	17	55.7	38.9-70.6
12	21	60.8	38.6-68.1	9	63.8	38.6-67.5	12	57.9	38.4-71.1
13	26	61.3	34.9-79.4	13	63.4	40.9-79.4	13	48.8	29.7-72.2
14	18	50.8	23.6-68.7	6	60.7	20.5-64.2	12	48.8	23.7-71.8
15	13	66.8	29.8-78.6	7	55.9	16.4-79.5	6	70.3	29.8-78.6
16	14	30.0	5.8-53.7	≥5	42.5	12.7-69.8	<5	-	-
17	14	26.5	18.4-73.0	7	24.2	19.0-76.2	7	28.8	16.2-73.0
18	20	53.7	28.8-87.8	9	54.4	44.6-62.5	11	51.1	19.9-99.5
19	18	46.3	14.6-70.9	7	21.7	10.5-46.0	11	57.9	39.0-70.9
Overall	**379***	**51.6**	**24.0-70.4**	**-**	**51.1**	**21.6-66.7**	**-**	**52.4**	**27.2-72.1**

*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=386

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.



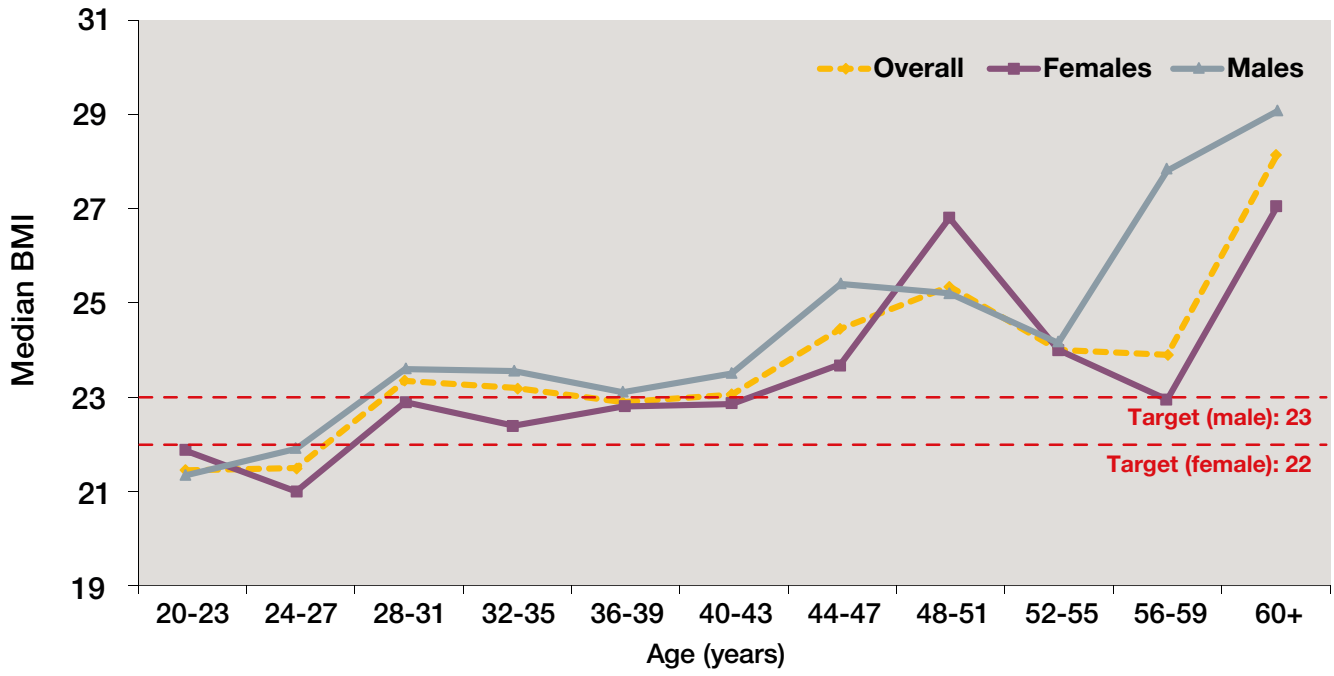
	Overall			Female			Male		
Age	n	Median	IQR	n	Median	IQR	n	Median	IQR
1	17	39.8	20.7-55.2	7	43.5	12.9-55.2	10	31.0	20.7-64.1
2	18	69.0	45.9-80.6	9	69.7	49.8-80.4	9	68.3	45.3-80.6
3	17	56.2	33.0-68.8	7	62.0	17.2-74.5	10	44.4	33.0-68.8
4	14	61.4	19.7-78.0	7	54.2	19.7-72.1	7	74.5	9.1-88.8
5	23	62.0	32.3-85.0	12	70.7	38.7-77.6	11	59.9	10.8-89.0
6	30	50.0	34.4-68.9	12	50.8	32.1-64.6	18	45.3	34.4-73.4
7	18	70.6	41.2-87.3	5	61.6	49.4-87.3	13	79.5	41.2-86.6
8	31	51.0	35.7-65.3	19	50.8	32.2-63.5	12	57.3	44.4-67.1
9	15	53.8	35.5-84.8	6	61.4	21.1-88.3	9	49.1	43.4-80.1
10	17	44.9	23.8-66.8	7	34.5	19.0-96.4	10	46.3	23.8-66.8
11	34	51.3	32.1-74.0	17	50.4	32.1-65.1	17	54.3	33.5-74.0
12	21	50.7	32.8-74.2	9	50.7	41.0-72.7	12	45.3	21.0-83.7
13	26	55.0	34.5-84.5	13	55.1	25.3-86.9	13	47.8	42.3-71.4
14	18	41.2	27.9-59.7	6	53.0	33.9-64.3	12	38.1	19.0-55.7
15	13	62.7	36.9-76.0	7	54.2	31.8-88.0	6	74.0	56.9-76.0
16	14	39.4	5.7-64.5	≥5	53.9	32.0-74.9	<5	-	-
17	14	39.2	18.0-64.0	7	40.0	34.9-73.5	7	28.1	7.0-64.0
18	20	63.3	39.7-87.3	9	65.3	61.7-73.9	11	46.8	29.4-98.3
19	18	64.9	16.0-86.8	7	45.3	6.5-71.7	11	71.8	18.2-91.8
Overall	**378***	**51.7**	**32.2-74.3**	**-**	**52.9**	**33.6-72.4**	**-**	**51.4**	**30.6-74.8**

*Number with non-missing data

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

1.6 Body Mass Index (BMI) in adults (20 years and over) N=472

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



	Overall			Female			Male		
Age	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**
20-23	100	21.4	19.1-23.8	48	21.9	18.8-24.4	52	21.4	19.3-23.0
24-27	83	21.5	19.8-23.6	40	21.0	18.9-23.1	43	21.9	20.3-24.0
28-31	64	23.4	21.1-25.2	31	22.9	21.0-25.2	33	23.6	21.8-25.2
32-35	60	23.2	20.7-25.3	24	22.4	21.1-25.5	36	23.6	20.5-25.1
36-39	45	22.9	21.4-25.0	16	22.8	21.1-24.7	29	23.1	21.5-26.4
40-43	26	23.0	22.2-25.7	12	22.9	21.9-25.7	14	23.5	22.5-25.7
44-47	18	24.5	22.3-26.3	7	23.7	20.3-25.9	11	25.4	23.7-30.0
48-51	20	25.4	22.5-26.9	9	26.8	20.7-27.8	11	25.2	23.3-26.0
52-55	22	24.0	21.1-28.0	10	24.0	19.8-30.0	12	24.1	21.3-27.5
56-59	5	23.9	23.8-27.8	<5	-	-	<5	-	-
60+	13	28.1	26.6-29.1	7	27.0	24.9-28.7	6	29.1	26.8-30.0
Overall	**456**	**22.9**	**20.5-25.3**	**-**	**22.6**	**19.8-24.8**	**-**	**23.1**	**21.0-25.7**

*Number with non-missing data

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

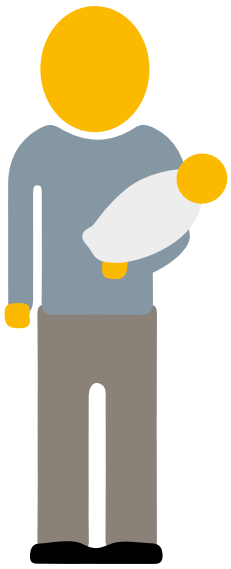
1.7 Education and employment in adults (16 years and over) N=538

The following table shows how people with CF reported their education and employment status in 2017. Please note that the groups are not mutually exclusive; someone may be a student as well as working part-time, for example.

	Number of patients n (%)
Number who completed questionnaire; n (%)	525 (97.6)
Full-time employment; n (%)	189 (35.1)
Part-time employment; n (%)	95 (17.7)
Student; n (%)	78 (14.5)
Homemaker; n (%)	16 (3.0)
Unemployed; n (%)	101 (18.8)
Disabled; n (%)	20 (3.7)
Retired; n (%)	15 (2.8)
Unknown entered; n (%)	11 (2.0)
No data recorded; n (%)	-*
No. in work or study; n (%)	**362 (69.0)**

1.8 Pregnancy

8 women with cystic fibrosis had babies in 2017



7 men with cystic fibrosis became fathers in 2017

*'No data recorded' is no longer available to select.

Diagnosis of cystic fibrosis

1.9 Age at diagnosis and screening in children under 16 in 2017 N=320

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.

Age at diagnosis	All patients <16; n (%)	Patients aged 10 years; n (%)	Patients aged 5 years; n (%)
Prenatal	0 (0)	0 (0)	0 (0)
Birth-3 months	281 (88.9)	17 (100.0)	21 (91.3)
4-6 months	9 (2.8)	-	<5
7-12 months	8 (2.5)	-	-
1 yr	<5	-	-
2 yrs	8 (2.5)	-	<5
3 yrs	<5	-	-
4 yrs	<5	-	-
5 yrs	-	-	-
6 yrs	-	-	-
7 yrs	-	-	-
8 yrs	-	-	-
9 yrs	-	-	-
10 yrs	<5	-	-
11 yrs	<5	-	-
12 yrs	-	-	-
13 yrs	-	-	-
14 yrs	-	-	-
15 yrs	-	-	-
Overall	**316***	**17**	**23**

The median (range) age at diagnosis for patients aged under 16 in 2017 is 46 days (0-192 months).

Diagnosis in the first three months of life is more common in children aged five years in 2017 (born after the UK-wide newborn screening programme was in place) than in children aged 10 years in 2017 (born during the final year of the introduction of universal newborn screening in the UK).

A total of 7 patients born in 2017 were identified by newborn screening (including those without complete data). As there is a delay between newborn screening tests being performed and the results entering the Registry, these statistics are updated retrospectively each year to take updated data into account. Therefore the number of patients identified in 2017 is higher (6) in this report than was recorded in the previous. It is likely that the 2017 figure will be updated in the next annual report in 2019.

*Number with non-missing data

1.10 Age at diagnosis and screening in adults aged 16 and over in 2017 N=538

The table below shows the age at diagnosis for people aged 16 and over in 2017. People aged 16 or over in 2017 were born when newborn screening was carried out done in a few areas of the UK, before it became universal in mid-2007.

Age at diagnosis	n (%)
Birth-3 months	186 (34.7)
4-6 months	50 (9.3)
7-12 months	33 (6.2)
1 year	40 (7.5)
2 years	35 (6.5)
3 years	25 (4.7)
4 years	17 (3.2)
5 years	10 (1.9)
6 years	7 (1.3)
7 years	7 (1.3)
8 years	8 (1.5)
9 years	6 (1.1)
10 years	<5
11 years	6 (1.1)
12 years	6 (1.1)
13 years	4 (0.7)
14 years	<5
15 years	7 (1.3)
16-19 years	14 (2.6)
20-23 years	9 (1.7)
24-27 years	10 (1.9)
28-31 years	16 (3.0)
32-35 years	11 (2.1)
36-39 years	8 (1.5)
40-43 years	<5
44-47 years	<5
48-51 years	<5
52-55 years	<5
56-59 years	<5
60+ years	<5
Overall	**536***

Overall, **85** (14.5%) adults with CF in the Registry in 2017 were diagnosed at age 16 or over.

In 2017, **5** people aged 16 or over were newly diagnosed with cystic fibrosis

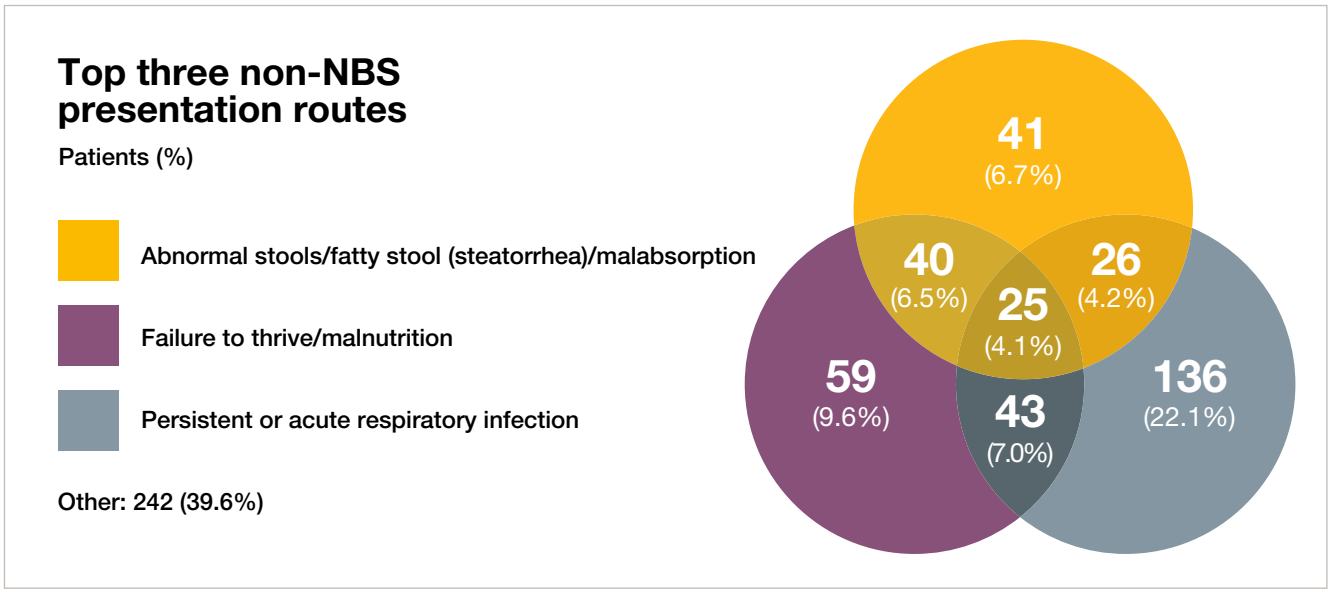
*Number with non-missing data

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	858	767	91
Number diagnosed by NBS	242	242	0
Total non-NBS	**614**	**526**	**90**

Mode of presentation (excluding NBS)	All patients (n=614)	Age <16 at diagnosis* (n=526)	Age ≥16 at diagnosis* (n=86)
Persistent or acute respiratory infection	230 (37.6)	182 (34.6)	48 (55.8)
Failure to thrive/malnutrition	167 (27.3)	167 (31.7)	0 (0)
Abnormal stools/fatty stool (steatorrhea)/ malabsorption	132 (21.6)	127 (24.1)	5 (5.8)
Meconium ileus	102 (16.7)	102 (19.4)	0 (0)
Family history	86 (14.1)	74 (14.1)	12 (14.0)
Genotype	29 (4.7)	21 (4.0)	8 (9.3)
Unknown	45 (7.4)	33 (6.3)	10 (11.6)
Rectal prolapse	18 (2.9)	18 (3.4)	0 (0)
Nasal polyps	5 (0.8)	<5	<5
Electrolyte imbalance	24 (3.9)	≥5	<5
Prenatal	<5	<5	<5
Bronchiectasis	7 (1.1)	<5	≥5
Liver disease	<5	<5	<5
Fertility	<5	<5	<5
Pancreatitis	<5	<5	<5
Oedema	<5	<5	<5



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

Lung health

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

In CF the condition of the lungs is often measured using FEV_1; the Forced Expiratory Volume of air in the first second of a forced exhaled breath. In this report, an FEV_1% predicted is based on the FEV_1 we would expect for a person without CF of the same age, gender, height, and ethnicity.

A person with CF who has FEV_1% predicted of 100% can breathe out the same amount of air in the first second of an exhaled breath as we would expect from a comparable person without cystic fibrosis. A person with FEV_1% predicted of 50% breathes out half the volume of air as a comparable person without cystic fibrosis.

For people with CF, an FEV_1% predicted of 85% or higher is the target, as this indicates normal or near-normal lung health. Each individual with CF will have their own FEV_1 target, based on their own lung function results and trends.

An aim of CF care is to prevent FEV_1% predicted from falling as much as possible, for as long as possible. This is often a team effort between people with CF, their family, and their medical team, which can include doctors, nurses, physiotherapists, dietitians, and psychologists.

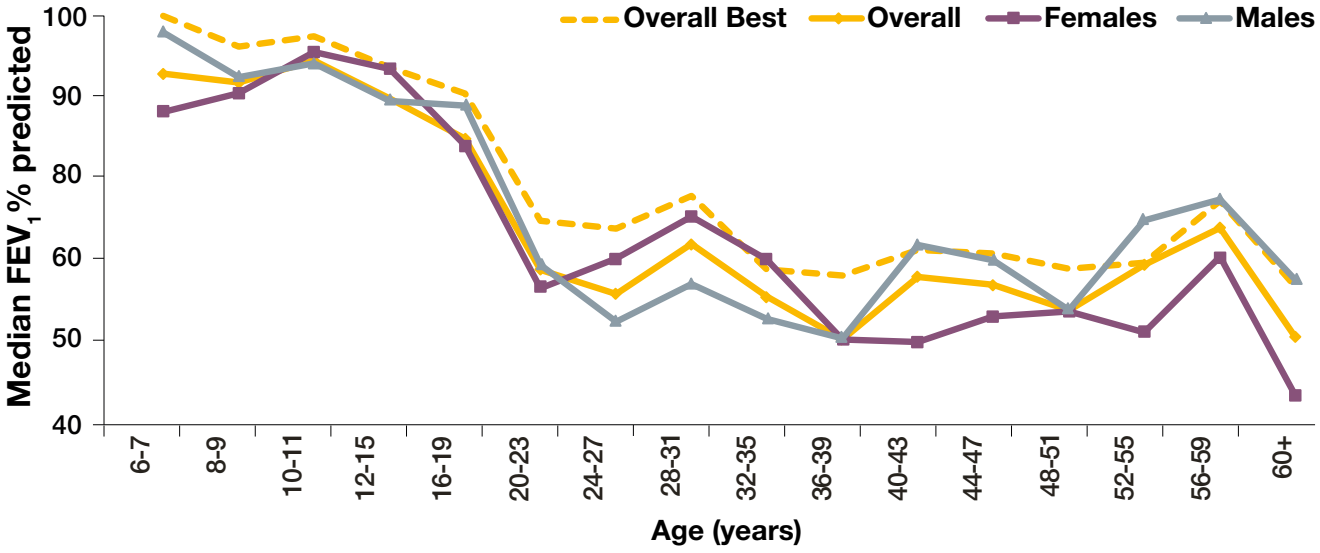
The FEV_1% predicted values shown in this report are calculated using an equation called Global Lungs Initiative, or 'GLI'[8]

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

1.12 FEV_1% predicted (GLI equations) in patients aged six years and older who have not had a lung transplant
N=761

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.

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	42	91.4	83.3-100.3	14	85.9	80.1-90.7	28	97.5	86.6-104.3
8-9	44	90.2	81.6-96.8	24	88.6	81.7-100.7	20	90.9	81.6-95.1
10-11	50	93.4	80.2-98.8	23	94.6	82.1-100.4	27	93.0	78.9-98.0
12-15	76	87.8	77.6-97.5	35	92.2	79.7-98.1	41	87.4	75.4-93.4
16-19	64	81.9	60.2-93.5	34	81.0	63.6-89.4	30	86.8	51.1-94.8
20-23	94	62.7	42.2-83.9	44	60.0	33.5-82.5	50	63.4	46.5-85.3
24-27	78	59.2	42.7-77.8	37	64.4	50.7-77.8	41	55.1	40.2-77.5
28-31	57	66.5	44.2-79.6	25	70.5	54.0-80.8	32	60.6	40.8-77.1
32-35	56	58.8	41.1-74.8	22	64.2	49.6-75.0	34	55.6	35.9-74.6
36-39	39	52.5	39.5-87.4	15	52.5	35.4-88.9	24	52.7	39.7-80.1
40-43	24	61.7	43.9-78.9	11	52.2	43.3-72.3	13	66.4	56.3-84.7
44-47	15	60.6	34.2-76.9	6	55.9	30.7-66.9	9	64.1	51.6-82.6
48-51	19	56.8	42.8-76.7	9	56.7	53.7-71.9	10	56.9	40.1-80.3
52-55	19	63.5	48.3-78.4	10	53.6	48.3-73.6	9	70.0	48.6-84.5
56-59	<5	-	-	<5	-	-	<5	-	-
60+	12	52.9	37.9-72.3	6	44.3	25.3-55.6	6	61.3	49.4-81.2
<16	212	89.8	80.1-98.3	96	88.6	81.2-98.6	116	90.0	79.5-98.2
≥16	481	63.5	43.6-83.2	220	64.8	47.6-81.0	261	61.5	41.1-84.5
<18	239	88.5	79.4-97.9	114	87.8	79.6-97.8	125	89.9	79.1-97.9
≥18	454	62.6	42.7-81.7	202	63.8	46.9-79.8	252	60.9	40.9-83.8
Overall	**693***	**75.3**	**52.2-91.1**	**316**	**75.6**	**53.0-89.5**	**377**	**74.3**	**51.5-92.1**

*Number with non-missing data

1.13 Best FEV_1% predicted (GLI equations) in patients aged six years and older who have not had a lung transplant
N=761

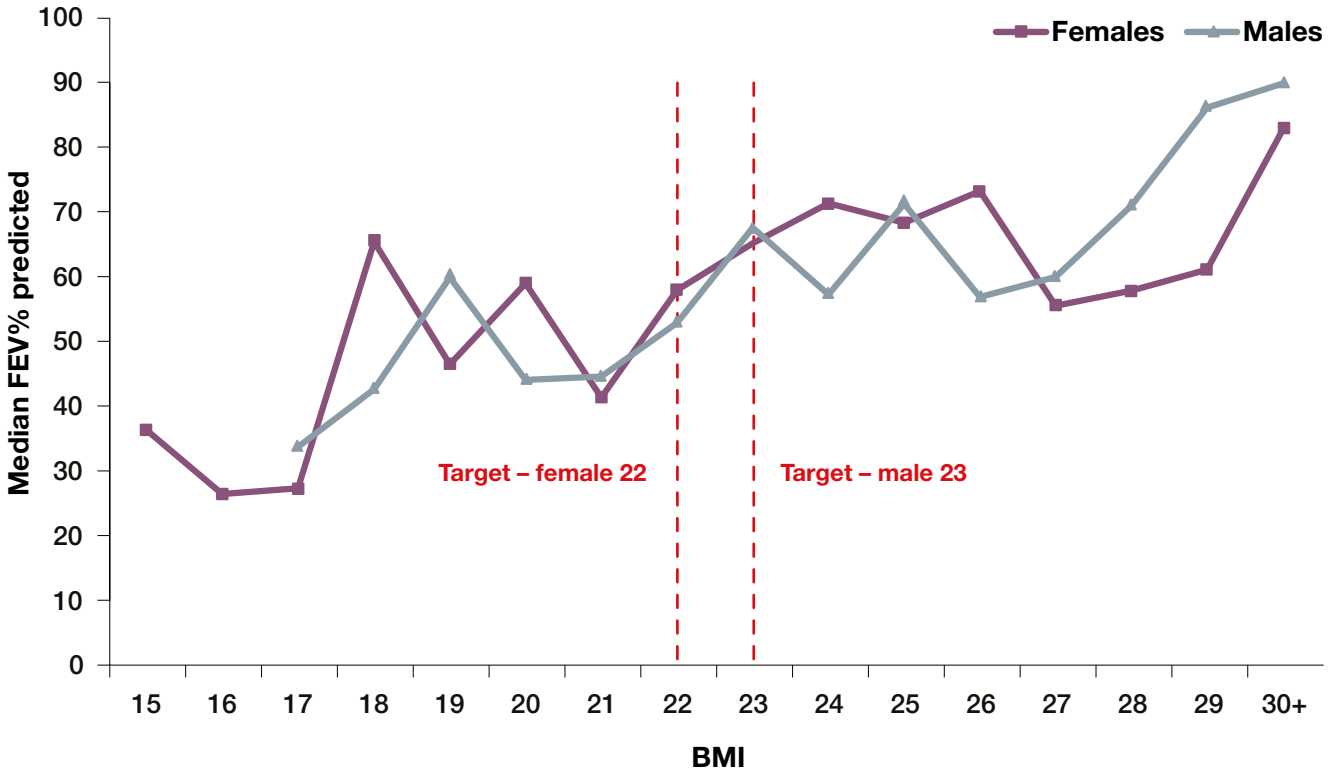
	Overall			Female			Male		
Age (yrs)	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**	**n**	**Median**	**IQR**
6-7	44	99.9	88.0-103.9	15	90.7	80.1-99.5	29	100.8	96.3-107.4
8-9	44	95.4	85.5-104.6	24	95.1	86.4-106.2	20	95.6	84.9-102.7
10-11	51	96.9	85.4-103.8	24	97.3	84.4-104.6	27	96.9	85.5-102.3
12-15	78	92.4	85.0-102.2	35	93.9	85.0-103.1	43	89.9	83.5-97.5
16-19	65	88.6	72.2-95.7	34	86.8	76.8-94.3	31	90.4	60.0-102.6
20-23	97	69.9	47.3-87.3	46	68.2	45.8-86.5	51	70.5	52.3-91.1
24-27	78	68.7	51.3-86.3	37	72.3	55.6-80.7	41	62.8	45.1-86.3
28-31	59	73.5	52.2-83.2	27	75.3	62.4-86.0	32	68.0	43.6-80.0
32-35	56	62.8	45.2-81.0	22	66.8	57.6-76.5	34	56.9	42.5-83.3
36-39	39	61.9	49.0-87.4	15	63.7	47.2-88.9	24	60.4	49.6-82.3
40-43	24	65.7	48.0-89.0	11	53.4	44.5-89.7	13	72.8	62.2-88.4
44-47	15	65.1	51.6-76.9	6	57.9	36.2-66.9	9	66.1	54.5-85.4
48-51	19	62.9	47.6-80.3	9	64.3	56.7-78.1	10	58.5	44.3-80.3
52-55	19	63.8	49.9-83.2	10	62.7	49.0-75.4	9	79.7	50.9-87.2
56-59	<5	-	-	<5	-	-	<5	-	-
60+	12	60.3	42.2-76.6	6	52.4	29.7-62.9	6	70.7	56.6-81.2
<16	217	94.9	85.4-103.4	98	94.1	85.0-103.2	119	95.8	85.9-103.6
≥16	487	70.3	50.8-87.5	224	70.9	53.5-87.1	263	69.3	47.5-88.4
<18	244	93.9	84.6-102.9	116	92.8	83.3-102.5	128	95.3	85.5-103.3
≥18	460	68.4	50.1-87.1	206	68.2	52.9-86.0	254	68.7	47.2-87.4
Overall	**704***	**80.2**	**59.0-95.8**	**322**	**80.1**	**61.6-94.3**	**382**	**80.3**	**57.5-96.6**

*Where Best FEV_1% was missing or less than the FEV_1% at annual review, annual review FEV_1% was used instead.

*Number with non-missing data

1.14 FEV_1% predicted (GLI equations) and BMI in people aged 20 years and over who have not had a transplant N=417*

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

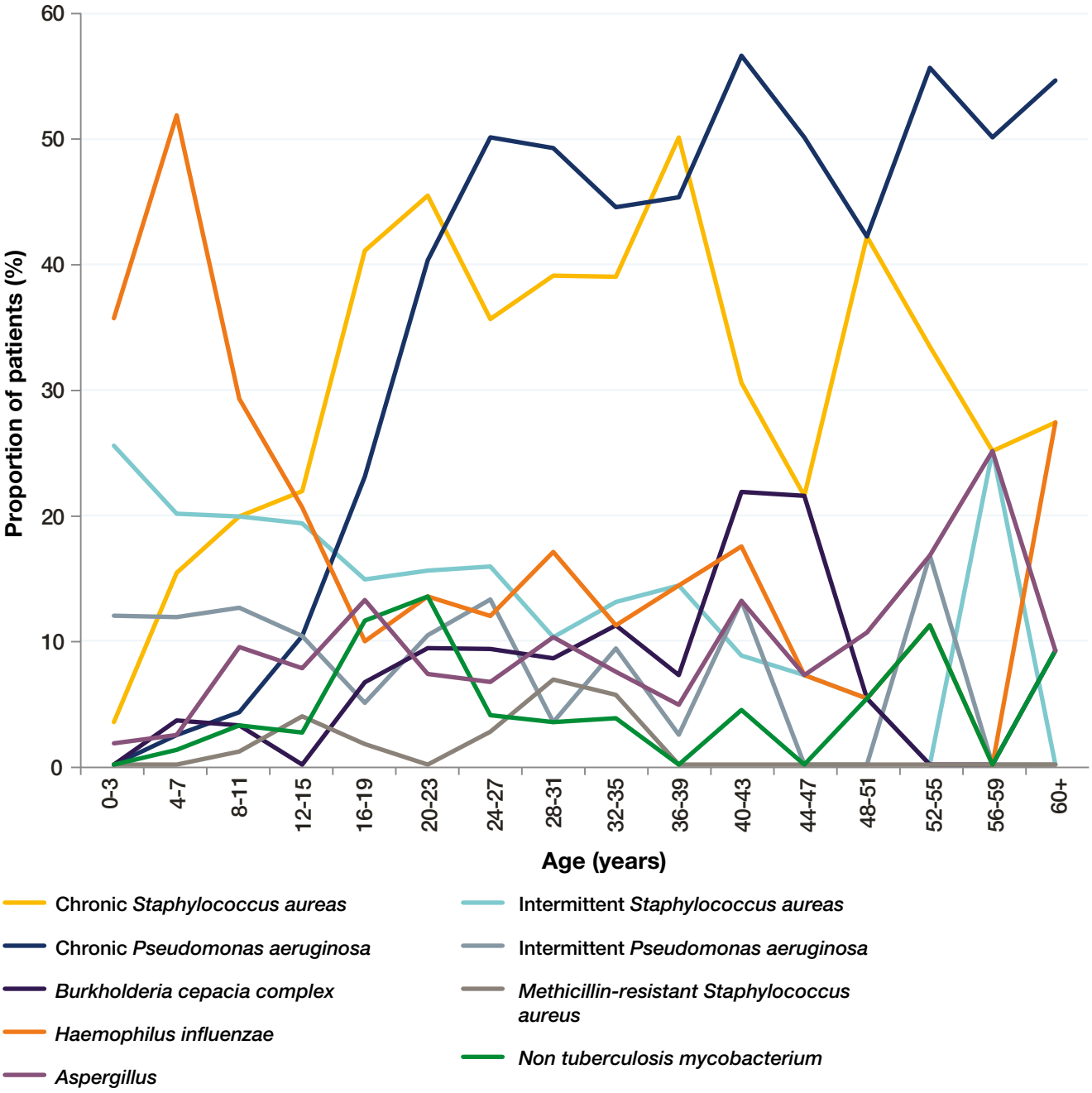


*Due to missing data, medians are calculated from a population of 417.
Each point represents the median FEV_1 % predicted of patients for each given BMI value. Due to the wide range of BMIs in this population with a value of 30 of more, these are grouped into one.

Lung infections

Lung infections can permanently reduce lung function in people with cystic fibrosis. Some lung infections can become ‘chronic’, meaning that they can’t ever be removed completely using medicines. All other infections are reported if they have occurred at least once as a positive growth in the 12 months prior to the patient’s annual review data set.

1.15 Lung infections in 2017
N=858



1.16 Lung infections in 2017
<16 years N=320; ≥16 years N=538

Infections in this table reflect bugs grown in the 12 months prior to the 2017 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	60	85	97	78	320
Number who had culture taken	59	85	96	78	318
Chronic *S. aureus* n (%)	<5	13 (15.3)	19 (19.8)	17 (21.8)	51 (16.0)
Intermittent *S. aureus* n (%)	15 (25.4)	17 (20.0)	19 (19.8)	15 (19.2)	66 (20.8)
Chronic *P. aeruginosa* n (%)	<5	<5	<5	8 (10.3)	14 (4.4)
Intermittent *P. aeruginosa* n (%)	7 (11.9)	10 (11.8)	12 (12.5)	8 (10.3)	37 (11.6)
B. cepacia complex n (%)	<5	<5	<5	<5	6 (1.9)
B. cenocepacia n (%)	<5	<5	<5	<5	<5
B. multivorans n (%)	<5	<5	<5	<5	<5
B. cepacia (other) n (%)	<5	<5	<5	<5	<5
MRSA n (%)	<5	<5	<5	<5	<5
H. influenza n (%)	21 (35.6)	44 (51.8)	28 (29.2)	16 (20.5)	109 (34.3)
NTM n (%)	<5	<5	<5	<5	6 (1.9)
Aspergillus n (%)	<5	<5	9 (9.4)	6 (7.7)	18 (5.7)

	Adult Age Range (Years)						Overall
	16-19	20-23	24-27	28-31	32-35	36-39	Adults (≥16 years)
Number in age range	66	100	88	65	64	45	538
Number who had culture taken	61	97	76	59	54	42	478
Chronic *S. aureus* n (%)	25 (41.0)	44 (45.4)	27 (35.5)	23 (39.0)	21 (38.9)	21 (50.0)	189 (39.5)
Intermittent *S. aureus* n (%)	9 (14.8)	15 (15.5)	12 (15.8)	6 (10.2)	7 (13.0)	6 (14.3)	60 (12.6)
Chronic *P. aeruginosa* n (%)	14 (23.0)	39 (40.2)	38 (50.0)	29 (49.2)	24 (44.4)	19 (45.2)	209 (43.7)
Intermittent *P. aeruginosa* n (%)	<5	10 (10.3)	10 (13.2)	<5	5 (9.3)	<5	37 (7.7)
B. cepacia complex n (%)	<5	9 (9.3)	7 (9.2)	5 (8.5)	6 (11.1)	<5	44 (9.2)
B. cenocepacia n (%)	<5	<5	<5	<5	<5	<5	11 (2.3)
B. multivorans n (%)	<5	<5	<5	<5	<5	<5	25 (5.2)
B. cepacia (other) n (%)	<5	<5	<5	<5	<5	<5	5 (1.0)
MRSA n (%)	<5	<5	<5	<5	<5	<5	10 (2.1)
H. influenza n (%)	6 (9.8)	13 (13.4)	9 (11.8)	10 (16.9)	6 (11.1)	6 (14.3)	61 (12.8)
NTM n (%)	7 (11.5)	13 (13.4)	<5	<5	<5	<5	32 (6.7)
Aspergillus n (%)	8 (13.1)	7 (7.2)	5 (6.6)	6 (10.2)	<5	<5	43 (9.0)
	Adult Age Range (Years)						Overall
	40-43	44-47	48-51	52-55	56-59	60+	Adults (≥16 years)
Number in age range	27	20	22	23	5	13	538
Number who had culture taken	23	14	19	18	<5	11	478
Chronic *S. aureus* n (%)	7 (30.4)	<5	8 (42.1)	6 (33.3)	<5	<5	189 (39.5)
Intermittent *S. aureus* n (%)	<5	<5	<5	<5	<5	<5	60 (12.6)
Chronic *P. aeruginosa* n (%)	13 (56.5)	7 (50.0)	8 (42.1)	10 (55.6)	<5	6 (54.5)	209 (43.7)
Intermittent *P. aeruginosa* n (%)	<5	<5	<5	<5	<5	<5	37 (7.7)
B. cepacia complex n (%)	5 (21.7)	<5	<5	<5	<5	<5	44 (9.2)
B. cenocepacia n (%)	<5	<5	<5	<5	<5	<5	11 (2.3)
B. multivorans n (%)	<5	<5	<5	<5	<5	<5	25 (5.2)
B. cepacia (other) n (%)	<5	<5	<5	<5	<5	<5	5 (1.0)
MRSA n (%)	<5	<5	<5	<5	<5	<5	10 (2.1)
H. influenza n (%)	<5	<5	<5	<5	<5	<5	61 (12.8)
NTM n (%)	<5	<5	<5	<5	<5	<5	32 (6.7)
Aspergillus n (%)	<5	<5	<5	<5	<5	<5	43 (9.0)

1.17 Nontuberculous mycobacteria (NTM) or atypical mycobacteria

Non-tuberculous mycobacterium 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.

	2015 (n=795)	2016 (n=829)	2017 (n=858)
NTM Prevalence (%)	37 (4.7%)	44 (5.3%)	38 (4.4%)
On NTM treatment in the given year (% of NTM prevalence in given year)	12 (32%)	16 (36%)	11 (29%)
NTM Incidence	20	20	12
M. abscessus prevalence	23	25	26
M. abscessus incidence*	-	7	10

1.18 Lung infections over time
N=429 in 2008, N=809 in 2013, N=858 in 2017

*M. abscessus incidence cannot be evaluated prior to 2016 as enhanced NTM reporting was not available before 2014.

Chronic *Staphylococcus aureus*				
	2008 (%)	2013 (%)	2017 (%)	p-value
0-3	0.0	3.6	3.4	0.960
4-7	4.1	12.2	15.3	0.517
8-11	4.8	11.4	19.8	0.076
12-15	11.2	25.9	21.8	0.698
16-19	24.1	35.2	41.0	0.653
20-23	44.4	34.7	45.4	0.175
24-27	23.1	42.1	35.5	0.129
28-31	28.6	44.4	39.0	0.311
32-35	71.4	37.8	38.9	0.652
36-39	71.4	21.4	50.0	0.018
40-43	50.0	31.6	30.4	0.758
44-47	40.0	33.3	21.4	- *
48-51	0.0	26.3	42.1	0.491
52-55	0.0	55.6	33.3	0.115
56-59	0.0	0.0	25.0	- *
60+	0.0	12.5	27.3	- *
<16 years	5.1	13.1	16.0	N/A
≥16 years	34.9	35.9	39.5	N/A
<18 years	6.0	15.0	17.1	N/A
≥18 years	41.0	36.9	40.1	N/A

Chronic *Pseudomonas aeruginosa*				
	2008 (%)	2013 (%)	2017 (%)	p-value
0-3	2.3	5.5	0.0	0.087
4-7	1.4	3.3	2.4	0.716
8-11	11.1	3.8	4.2	0.799
12-15	20.2	10.3	10.3	0.935
16-19	27.6	31.9	23.0	0.163
20-23	44.4	50.5	40.2	0.101
24-27	61.5	51.3	50.0	0.298
28-31	71.4	45.8	49.2	0.945
32-35	57.1	48.9	44.4	0.279
36-39	42.9	42.9	45.2	0.848
40-43	50.0	52.6	56.5	0.900
44-47	80.0	45.8	50.0	0.467
48-51	50.0	31.6	42.1	0.747
52-55	0.0	55.6	55.6	0.538
56-59	0.0	33.3	50.0	- *
60+	100.0	75.0	54.5	0.195
<16 years	9.0	5.3	4.4	N/A
≥16 years	47.0	45.4	43.7	N/A
<18 years	10.2	8.0	5.8	N/A
≥18 years	54.1	47.5	45.0	N/A

*Sample size too low for hypothesis test

Complications

1.19 Prevalence of complications

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

	2016			2017*		
	Overall (n=795)	**<16 years (n=298)**	**≥16 years (n=497**	**Overall (n=858)**	**<16 years (n=320)**	**≥16 years (N=538)**
Respiratory related						
Nasal polyps requiring surgery	-	<5	9 (1.7)	-	<5	16 (3.0)
Sinus disease	-	<5	164 (31.5)	-	<5	122 (22.7)
Asthma	79 (9.5)	14 (4.5)	64 (12.5)	63 (7.3)	9 (2.8)	54 (10.0)
Allergic bronchopulmonary aspergillosis (ABPA)	-	<5	33 (6.3)	37 (4.3)	7 (2.2)	30 (5.6)
Any haemoptysis	23 (2.8)	0	23 (4.4)	-	<5	25 (4.6)
Massive haemoptysis	-			-	-	-
Pneumothorax requiring chest tube	0	0	0	0	0	0
Pancreas & hepatobiliary disease						
Raised liver enzymes	-	<5	25 (4.8)	41 (4.8)	14 (4.4)	27 (5.0)
Liver disease	104 (12.5)	23 (7.4	81 (15.6)	68 (7.9)	18 (5.6)	50 (9.3)
Cirrhosis with no portal hypertension	-	<5	12 (2.3)	-	<5	13 (2.4)
Cirrhosis with portal hypertension	19 (2.3)	6 (1.9)	13 (2.5)	-	<5	13 (2.4)
Gall bladder disease requiring surgery	<5	<5	<5	-	<5	<5
Pancreatitis	-	<5	7 (1.3)	-	<5	10 (1.9)
Upper gastrointestinal						
Gastroeosophageal reflux disease (GERD)	-	<5	99 (19.0)	-	<5	113 (21.0)
Peptic ulcer	0	0	0	0	0	0
GI bleed (varices as source)	0	0	0	0	0	0
GI bleed (non varices as source)	-	0	<5	-	<5	<5
Lower gastrointestinal						
Intestinal obstruction	0	0	0	-	<5	0
Distal intestinal obstruction syndrome	-	-	-	-	<5	94 (17.5)
Fibrosing colonopathy/colonic stricture	-	<5	88 (16.9)	0	0	0
Rectal prolapse	0	0	0	-	<5	<5
Renal						
Kidney stones	-	<5	<5	-	0	<5
Renal failure	13 (1.6)	0	13 (2.5)	13 (1.5)	0	13 (2.4)
Musculo-skeletal						
Arthritis	-	<5	<5	5 (0.6)	0	5 (0.9)
Arthropathy	33 (4.0)	0	44 (6.3)	32 (3.7)	0	32 (5.9)
Bone fracture	0	0	0	-	0	<5
Osteopenia	107 (12.9)	0	107 (20.6)	107 (12.5)	0	107 (19.9)
Osteoporosis	42 (5.1)	0	42 (8.1)	40 (4.7)	0	40 (7.4)
Other						
Cancer confirmed by histology	0	0	0	-	<5	<5
Port inserted or replaced	-	<5	<5	18 (2.1)	6 (1.9)	12 (2.2)
Depression	21 (2.5)	0	21 (4.0)	19 (2.2)	0	19 (3.5)
Hearing loss	-	<5	9 (1.7)	-	<5	7 (1.3)
Hypertension	17 (2.1)	0	17 (3.3)	17 (2.0)	0	17 (3.2)

Please note that in 2017 the data entry pathway for complications changed, which has resulted in a drop in prevalence for several complications. Results in 2017 should be interpreted with caution until 2018, when we will confirm whether this is a true decrease.

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.

	Newly identified in 2016			Newly identified in 2017		
	Overall (n=829)	<16 years (n=309)	≥16 years (n=520)	Overall (n=858)	<16 years (n=320)	≥16 years (n=538)
Allergic bronchopulmonary aspergillosis (ABPA); n (%)	**11 (1.3)**	**<5**	**≥5**	**10 (1.2)**	**<5**	**≥5**
Cirrhosis - no portal hypertension; n (%)	13 (1.6)	<5	≥5	9 (1.0)	<5	≥5
Cirrhosis - with portal hypertension; n (%)	7 (0.8)	<5	<5	6 (0.7)	<5	≥5
Cancer confirmed by histology; n (%)	<5	<5	<5	<5	<5	5

1.21 CF-related diabetes
N=667

Cystic fibrosis-related diabetes (CFRD) is common in adults and adolescents with cystic fibrosis. This is because, for many people with CF, the pancreas does not work properly. This can mean that not enough insulin is produced, 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=667)	10-15 years (n=129)	≥16 years (n=538)
On CFRD treatment; n (%)	150 (22.5)	7 (5.4)	143 (26.6)
CFRD screening; n (%)			
Yes	354 (53.1)	115 (89.1)	239 (44.4)
No	133 (19.9)	6 (4.7)	127 (23.6)
Existing CFRD diagnosis	152 (22.8)	7 (5.4)	145 (27.0)
Unknown	9 (1.3)	<5	≥5

Antibiotics

1.22 Intravenous (IV) antibiotics
N=858

When someone with CF becomes unwell with an infection, they might be prescribed intravenous (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	60	<5	-	11 (18.3)	13 (13-13)	11 (18.3)	14 (7-18)
4-7	85	5 (5.9)	25 (12-34)	18 (21.2)	14 (12-34)	20 (23.5)	15 (14-40)
8-11	97	9 (9.3)	40 (28-47)	28 (28.9)	15 (28-47)	30 (30.9)	35 (14-68)
12-15	78	8 (10.3)	26 (23-45)	23 (29.5)	17 (23-45)	27 (34.6)	26 (14-42)
16-19	66	13 (19.7)	18 (14-37)	19 (28.8)	16 (14-37)	24 (36.4)	26 (14-49)
20-23	100	33 (33.0)	14 (10-28)	42 (42.0)	14 (10-28)	52 (52.0)	25 (14-44)
24-27	88	26 (29.5)	14 (14-35)	29 (33.0)	14 (14-35)	38 (43.2)	26 (14-42)
28-31	65	18 (27.7)	27 (10-42)	18 (27.7)	15 (10-42)	24 (36.9)	33 (25-51)
32-35	64	12 (18.8)	25 (14-39)	16 (25.0)	19 (14-39)	22 (34.4)	29 (14-42)
36-39	45	15 (33.3)	16 (13-49)	7 (15.6)	25 (13-49)	16 (35.6)	37 (14-52)
40-43	27	9 (33.3)	17 (14-28)	8 (29.6)	13 (14-28)	14 (51.9)	14 (13-36)
44-47	20	<5	-	<5	16 (10-31)	5 (25.0)	14 (13-44)
48-51	22	<5	-	<5	41 (49-68)	5 (22.7)	61 (55-63)
52-55	23	5 (21.7)	14 (12-14)	9 (39.1)	14 (12-14)	11 (47.8)	14 (14-14)
56-59	5	0 (0.0)	-	<5	-	<5	-
60+	13	<5	9 (4-32)	6 (46.2)	10 (4-32)	6 (46.2)	14 (14-39)
<16	320	23 (7.2)	28 (22-41)	80 (25.0)	14 (22-41)	88 (27.5)	23 (14-43)
≥16	538	142 (26.4)	18 (13-39)	161 (29.9)	14 (13-39)	218 (40.5)	27 (14-44)
<18	348	29 (8.3)	27 (18-40)	89 (25.6)	14 (18-40)	100 (28.7)	23 (14-42)
≥18	510	136 (26.7)	17 (13-40)	152 (29.8)	14 (13-40)	206 (40.4)	27 (14-45)
Overall	**858**	**165 (19.2)**	**21 (13-40)**	**241 (28.1)**	**14 (13-40)**	**306 (35.7)**	**26 (14-44)**

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

	2008			2013			2017		
	Overall	<16 years	≥16 years	Overall	<16 years	≥16 years	Overall	<16 years	≥16 years
Patients with chronic *P. aeruginosa*	67	28	39	241	17	224	223	14	209
Tobramycin solution; n (%)	6 (9.0)	<5	<5	48 (19.9)	<5	≥5	34 (15.2)	<5	≥5
Other aminoglycoside; n (%)	0	0	0	0	0	0	<5	0	<5
Colistin; n (%)	21 (31.3)	15 (53.6)	6 (15.4)	92 (38.2)	13 (76.5)	79 (35.3)	55 (24.7)	6 (42.9)	49 (23.4)
Promixin; n (%)	<5	0	<5	41 (17.0)	<5	≥5	35 (15.7)	5 (35.7)	30 (14.4)
Aztreonam; n (%)	-	-	-	<5	0	<5	17 (7.6)	0	17 (8.1)
Colistimethate (DPI); n (%)	-	-	-	-	-	-	54 (24.2)	<5	≥5
Tobramycin Inhalation Powder; n (%)	-	-	-	-	-	-	64 (28.7)	0	64 (30.6)
At least one of the above; n (%)	28 (41.8)	17 (60.7)	11 (28.2)	151 (62.7)	15 (88.2)	136 (60.7)	183 (82.1)	12 (85.7)	171 (81.8)

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

1.24 Long-term azithromycin use

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

		Patients with chronic *P. aeruginosa*; n (%)	Patients without chronic *P. aeruginosa*; n (%)
2008	**Overall (n=429)**	**29 (46.0)**	**34 (54.0)**
	0-3 years (n=91)	<5	<5
	4-15 years (n=236)	12 (35.3)	22 (64.7)
	≥16 years n=(102)	17 (60.7)	11 (39.3)
2013	**Overall (n=809)**	**185 (49.9)**	**186 (50.1)**
	0-3 years (n=70)	<5	<5
	4-15 years (n=244)	8 (17.0)	39 (83.0)
	≥16 years (n=495)	176 (54.7)	146 (45.3)
2017	**Overall (n=858)**	**190 (43.0)**	**252 (57.0)**
	0-3 years (n=60)	0 (0)	<5
	4-15 years (n=260)	11 (13.3)	72 (86.7)
	≥16 years (n=538)	179 (50.1)	178 (49.9)

*In 2013, this includes Aztreonam. In 2017 it includes Aztreonam, Colistimethate and Tobramycin Inhalation Powder.

1.25 Flucloxacillin

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

Age	Total patients	Patients on Flucloxacilin; n (%)
0-3	58	32 (55.2)
4-7	82	28 (34.1)
8-11	92	31 (33.7)
12-15	77	26 (33.8)
16-19	64	20 (31.3)
20-23	100	29 (29.0)
24-27	82	17 (20.7)
28-31	64	8 (12.5)
32-35	59	7 (11.9)
36-39	42	<5
40-43	25	<5
44-47	17	<5
48-51	19	<5
52-55	22	<5
56-59	5	0
60+	13	0
<16 years	309	117 (37.9)
≥16 years	512	89 (17.4)
<18 years	337	121 (35.9)
≥18 years	484	85 (17.6)
Overall	**821**	**206 (25.1)**

Muco-active therapies

1.26 Mannitol

Age	Total patients	Patients on Mannitol; n (%)
0-3	58	0
4-7	82	0
8-11	92	0
12-15	77	0
16-19	64	0
20-23	100	<5
24-27	82	<5
28-31	64	<5
32-35	59	<5
36-39	42	<5
40-43	25	0
44-47	17	<5
48-51	19	<5
52-55	22	0
56-59	5	0
60+	13	0
<16 years	309	0
≥16 years	512	10 (2.0)
<18 years	337	0
≥18 years	484	10 (2.1)
Overall	**821**	**10 (1.2)**

1.27 DNase

	2008		2013		2017	
Age	Total patients	Patients on DNase; n (%)	Total patients	Patients on DNase; n (%)	Total patients	Patients on DNase; n (%)
0-3	91	<5	70	<5	58	<5
4-7	78	9 (11.5)	101	11 (10.9)	82	18 (22.0)
8-11	66	15 (22.7)	82	22 (26.8)	92	42 (45.7)
12-15	92	33 (35.9)	61	27 (44.3)	77	38 (49.4)
16-19	32	12 (37.5)	99	41 (41.4)	64	33 (51.6)
20-23	14	<5	97	42 (43.3)	100	61 (61.0)
24-27	14	<5	75	29 (38.7)	82	41 (50.0)
28-31	11	<5	67	25 (37.3)	64	30 (46.9)
32-35	8	<5	49	12 (24.5)	59	25 (42.4)
36-39	9	<5	26	7 (26.9)	42	14 (33.3)
40-43	5	<5	19	<5	25	12 (48.0)
44-47	5	<5	22	7 (31.8)	17	7 (41.2)
48-51	<5	0	20	6 (30.0)	19	8 (42.1)
52-55	<5	0	7	<5	22	7 (31.8)
56-59	<5	0	7	0	5	<5
60+	<5	0	7	<5	13	<5
<16 years	327	60 (18.3)	314	63 (20.1)	309	99 (32.0)
≥16 years	102	23 (22.5)	495	179 (36.2)	512	242 (47.3)
<18 years	350	71 (20.3)	363	86 (23.7)	337	112 (33.2)
≥18 years	79	12 (15.2)	446	156 (35.0)	484	229 (47.3)
Overall	**429**	**83 (19.3)**	**809**	**242 (29.9)**	**821**	**341 (41.5)**

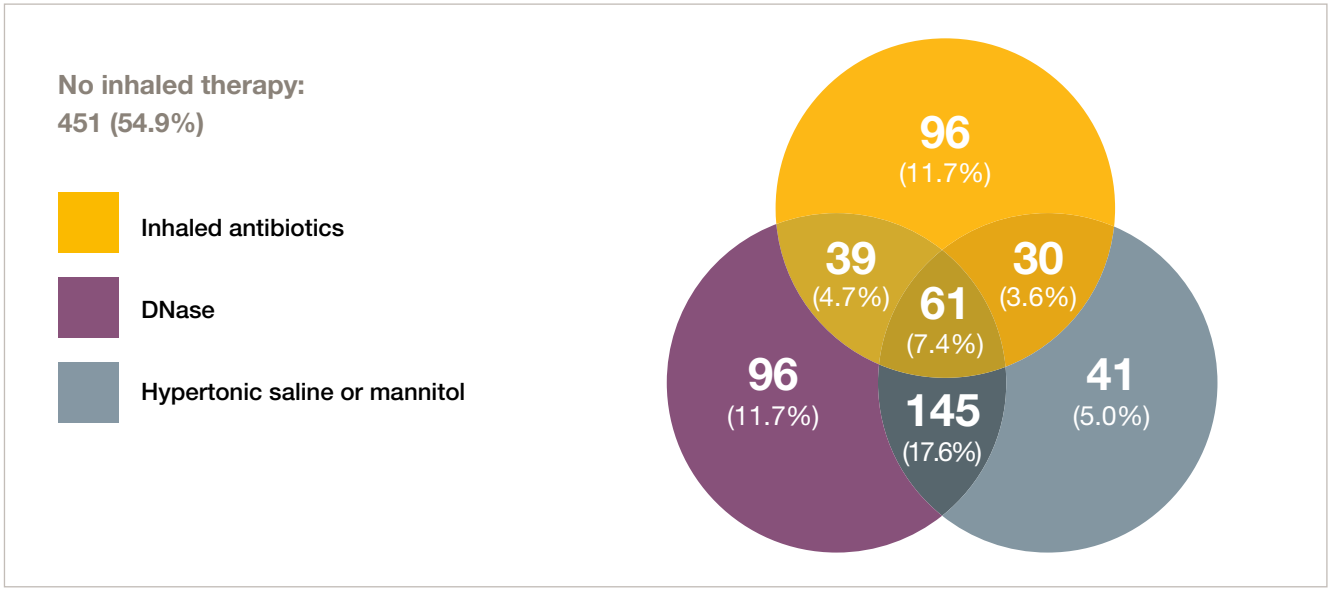
1.28 Hypertonic saline

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

	2008		2013		2017	
Age	**Total patients**	**Patients on hypertonic saline; n (%)**	**Total patients**	**Patients on hypertonic saline; n (%)**	**Total patients**	**Patients on hypertonic saline; n (%)**
0-3	91	0	70	<5	58	6 (10.3)
4-7	78	0	101	6 (5.9)	82	12 (14.6)
8-11	66	<5	82	10 (12.2)	92	22 (23.9)
12-15	92	<5	61	12 (19.7)	77	25 (32.5)
16-19	32	0	99	26 (26.3)	64	19 (29.7)
20-23	14	0	97	25 (25.8)	100	24 (24.0)
24-27	14	0	75	9 (12.0)	82	14 (17.1)
28-31	11	<5	67	7 (10.4)	64	16 (25.0)
32-35	8	0	49	5 (10.2)	59	11 (18.6)
36-39	9	0	26	0	42	5 (11.9)
40-43	5	0	19	<5	25	<5
44-47	5	0	22	<5	17	<5
48-51	<5	0	20	<5	19	<5
52-55	<5	<5	7	<5	22	<5
56-59	<5	0	7	0	5	0
60+	<5	0	7	<5	13	<5
<16 years	327	<5	314	29 (9.2)	309	65 (21.0)
≥16 years	102	<5	495	81 (16.4)	512	100 (19.5)
<18 years	350	<5	363	46 (12.7)	337	74 (22.0)
≥18 years	79	<5	446	64 (14.3)	484	91 (18.8)
Overall	**429**	**<5**	**809**	**110 (13.6)**	**821**	**165 (20.1)**

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 451 (54.9%) 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.

Number of patients on ivacaftor in Scotland	70
Sweat chloride before ivacaftor	99 (95-104)
Sweat chloride 6-8 weeks after ivacaftor	49 (40-67)
FEV_1% before ivacaftor	62.5 (47.5-79.8)
FEV_1% 6-8 weeks after ivacaftor	72.5 (53.2-81.6)
Number of patients stopped ivacaftor ever	7

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.

Ivacaftor/Lumacaftor

Ivacaftor/Lumacaftor is licensed for use in patients aged 12 and over with two copies of the F508del mutation. In 2017 it was available to specific people with CF in the UK through a named patient access scheme. In Scotland, seven people received this drug in 2017.

1.31 Oxygen and non-invasive ventilation

	Overall (n=858)	<16 years (n=320)	≥16 years (n=538)	<18 years (n=348)	≥18 years (n=510)
Non-invasive ventilation (NIV); n (%)	9 (1.0)	<5	≥5	<5	≥5
Long-term oxygen; n (%)	44 (5.1)	5 (1.6)	39 (7.2)	6 (1.7)	38 (7.5)
Among those who have long-term oxygen:					
Continuously	10 (22.7)	<5	≥5	< 5	≥5
Nocturnal or with exertion	12 (27.3)	0 (0.0)	12 (30.8)	0 (0.0)	12 (31.6)
As required (PRN)	7 (15.9)	0 (0.0)	7 (17.9)	<5	≥5
With exacerbation	15 (34.1)	<5	≥5	<5	≥5

1.32 Physiotherapy

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

	Overall (n=858)	<16 years (n=320)	≥16 years (n=538)	<18 years (n=348)	≥18 years (n=510)
Active cycle of breathing techniques; n (%)	137 (16.0)	23 (7.2)	114 (21.2)	29 (8.3)	108 (21.2)
Autogenic drainage (including assisted autogenic drainage); n (%)	380 (44.3)	73 (22.8)	307 (57.1)	86 (24.7)	294 (57.6)
Postural drainage; n (%)	7 (0.8)	≥5	<5	≥5	<5
Any form of positive expiratory pressure (PEP); n (%)	421 (49.1)	268 (83.8)	153 (28.4)	284 (81.6)	137 (26.9)
VEST; n (%)	<5	<5	<5	<5	<5
Exercise; n (%)	457 (53.3)	141 (44.1)	316 (58.7)	155 (44.5)	302 (59.2)
Other; n (%)	188 (21.9)	138 (43.1)	50 (9.3)	143 (41.1)	45 (8.8)

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=858)	<16 years (n=320)	≥16 years (n=538)	<18 years (n=348)	≥18 years (n=510)
Any supplemental feeding; n (%)	191 (22.3)	67 (20.9)	124 (23.0)	70 (20.1)	121 (23.7)
Nasogastric tube; n (%)	15 (1.7)	5 (1.6)	10 (1.9)	5 (1.4)	10 (2.0)
Gastrostomy tube/button; n (%)	26 (3.0)	13 (4.1)	13 (2.4)	13 (3.7)	13 (2.5)
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.

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

Genotypes

Genotypes are part of the genetic makeup of an individual that usually control a particular characteristic, known as a phenotype. For people with CF, their genotype reveals which mutations of the CF gene causes their cystic fibrosis. Everyone living with CF has two mutations of the gene for CFTR; one on each allele. One is inherited from their mother, and one from their father. If both mutations (or genotypes) are the same, the person is said to be homozygous. Someone who has two different variants is heterozygous.

852 (99.3%) patients have been genotyped with at least one recorded value.

F508del Mutations: n (%)

Homozygous F508del: 365 (42.8%)

Heterozygous F508del: 399 (46.8%)

1.35 Genotypes in the UK 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	765	89.2
c.1652G->A	p.Gly551Asp	G551D	91	10.6
c.350G->A	p.Arg117His	R117H	71	8.3
c.1624G->T	p.Gly542X	G542X	62	7.2
c.200C->T	p.Pro67Leu	P67L	35	4.1
c.1679G->C	p.Arg560Thr	R560T	17	2.0
c.1585-1G->A		1717-1G->A	14	1.6
c.1477C->T	p.Gln493X	Q493X	14	1.6
c.3454G->C	p.Asp1152His	D1152H	12	1.4
c.489+1G->T		621+1G->T	12	1.4
c.3909C->G	p.Asn1303Lys	N1303K	12	1.4
c.2657+5G->A		2789+5G->A	10	1.2
c.3528delC	p.Lys1177SerfsX15	3659delC	10	1.2
c.178G->T	p.Glu60X	E60X	8	0.9
c.1558G->T	p.Val520Phe	V520F	7	0.8
c.1210-12[5](AJ574948.1:g.152T[5])		5T	6	0.7
c.948delT	p.Phe316LeufsX12	1078delT	6	0.7
c.3717+12191C->T		3849+10kbC->T	5	0.6
c.1705T->G	p.Tyr569Asp	Y569D	5	0.6
c.1364C->A	p.Ala455Glu	A455E	5	0.6
c.1519_1521delATC	p.Ile507del	I507del	<5	-
c.1766+1G->A		1898+1G->A	<5	-
c.3140-26A->G		3272-26A->G	<5	-
c.2657+2_2657+3insA		2789+2insA	<5	-
c.254G->A	p.Gly85Glu	G85E	<5	-

Nucleotide	Protein	Legacy name	N	%
c.3846G->A	p.Trp1282X	W1282X	<5	-
c.223C->T	p.Arg75X	R75X	<5	-
c.2988G->A		3120G->A	<5	-
c.2988+1G->A		3120+1G->A	<5	-
c.2012delT	p.Leu671X	2143delT	<5	-
c.1657C->T	p.Arg553X	R553X	<5	-
c.3196C->T	p.Arg1066Cys	R1066C	<5	-
c.273+1G->A		405+1G->A	<5	-
c.274G->A	p.Glu92Lys	E92K	<5	-
c.579+3A->G		711+3A->G	<5	-
c.2583delT	p.Phe861LeufsX3	2711delT	<5	-
c.3266G->A	p.Trp1089X	W1089X	<5	-
c.3884_3885insT	p.Ser1297PhefsX5	4016insT	<5	-
c.443T->C	p.Ile148Thr	I148T	<5	-
c.3484C->T	p.Arg1162X	R1162X	<5	-
c.1466C->A	p.Ser489X	S489X	<5	-
c.1000C->T	p.Arg334Trp	R334W	<5	-
c.2490+1G->A		2622+1G->A	<5	-
c.579+1G->T		711+1G->T	<5	-
c.1753G->T	p.Glu585X	E585X	<5	-
c.1209+1G->A		1341+1G->A	<5	-
c.3705T->G	p.Ser1235Arg	S1235R	<5	-
c.3276C->A	p.Tyr1092X	Y1092X(C->A)	<5	-
c.2052delA	p.Lys684AsnfsX38	2184delA	<5	-
Other selected			107	12.5

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 2 on page 62.

Key

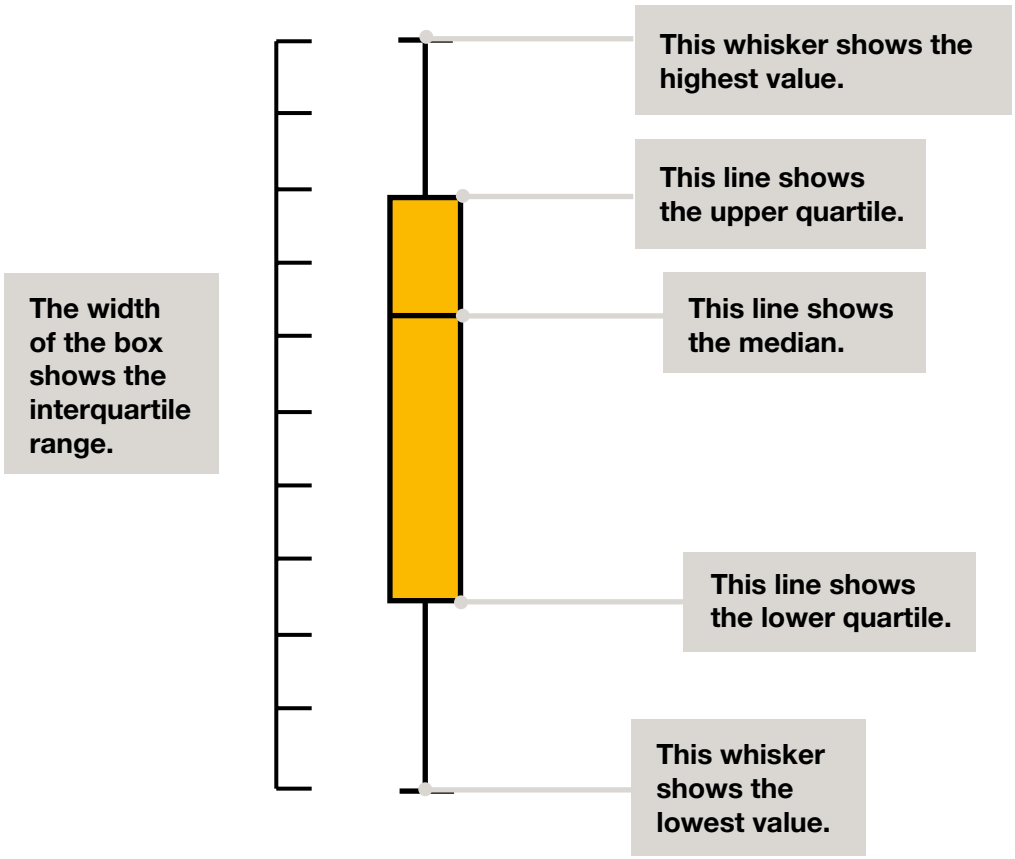

Paediatric centre


Adult centre

A guide to the charts

Some of the data in this section are shown as ‘box plots’. We also show the data in ‘funnel plots’.

Box plots

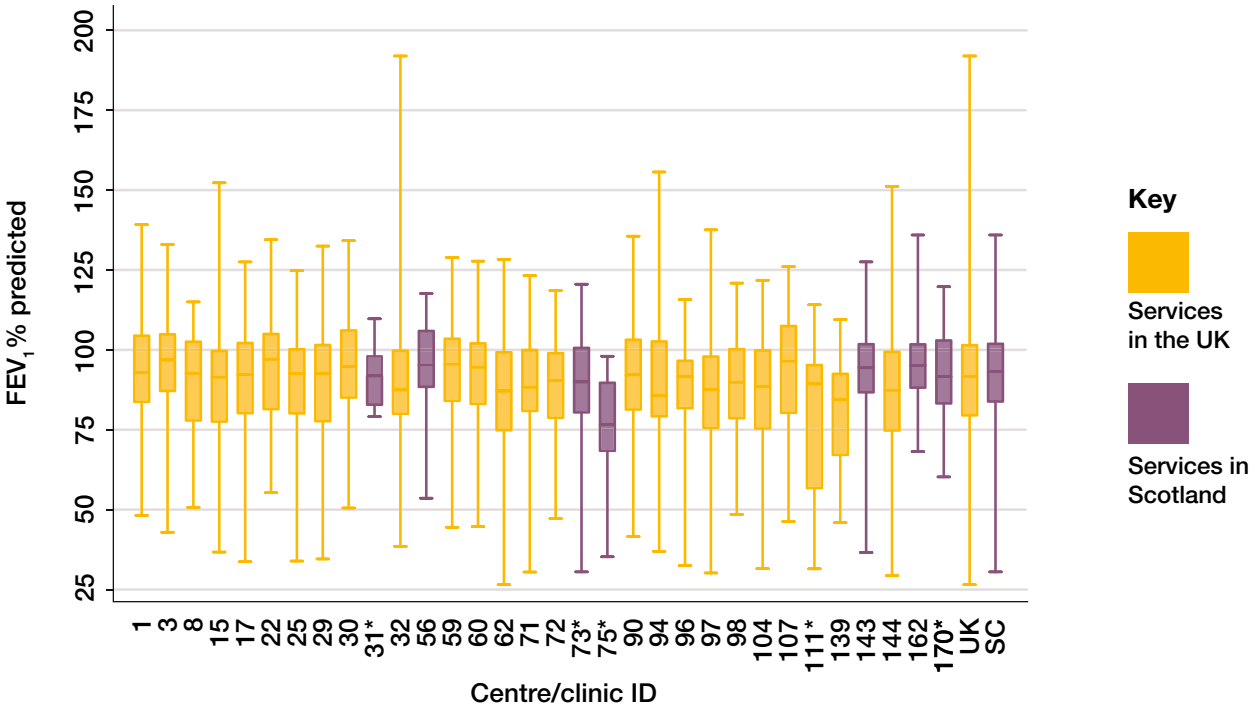


- The ‘box’ shows the middle half of the data for that centre, going from the first quartile to the third quartile. The longer the box, the more varied the data for that centre.
- The horizontal line within the box shows the median result for that centre.
- The ‘whiskers’ above and below the box show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

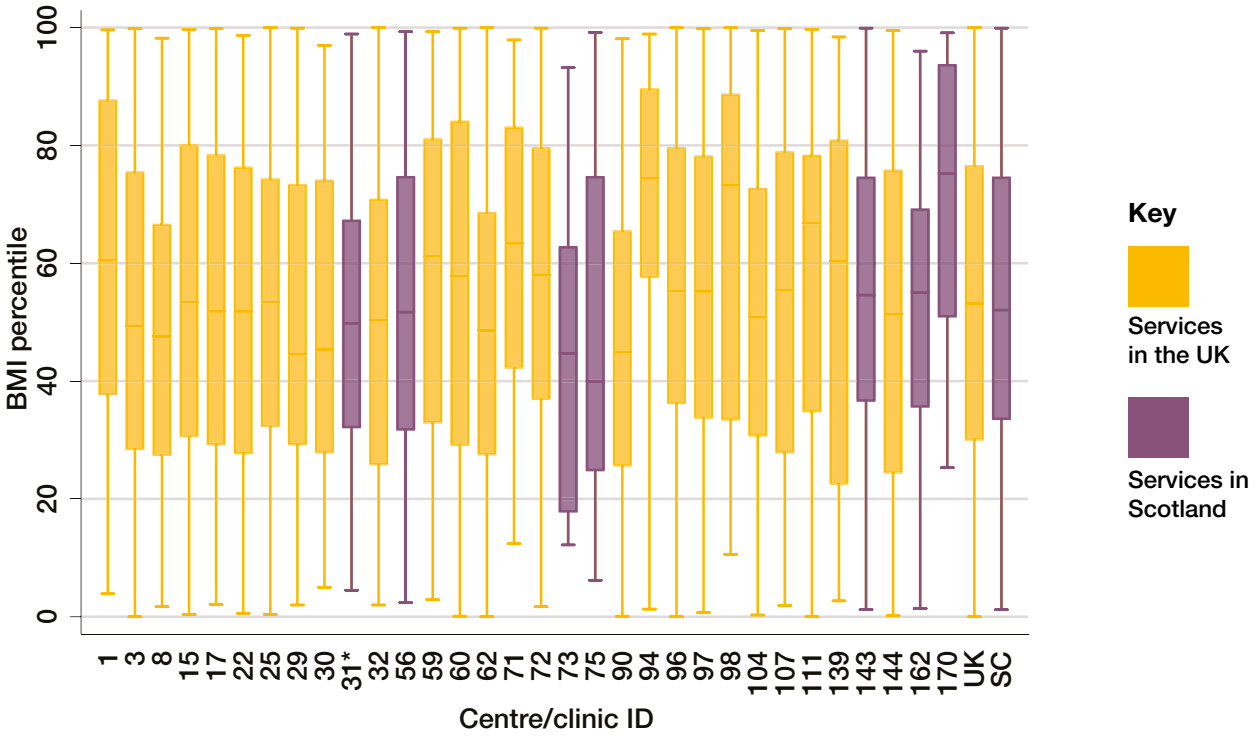
This section shows results for the six paediatric centres with their network clinics, and one stand-alone clinic.

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



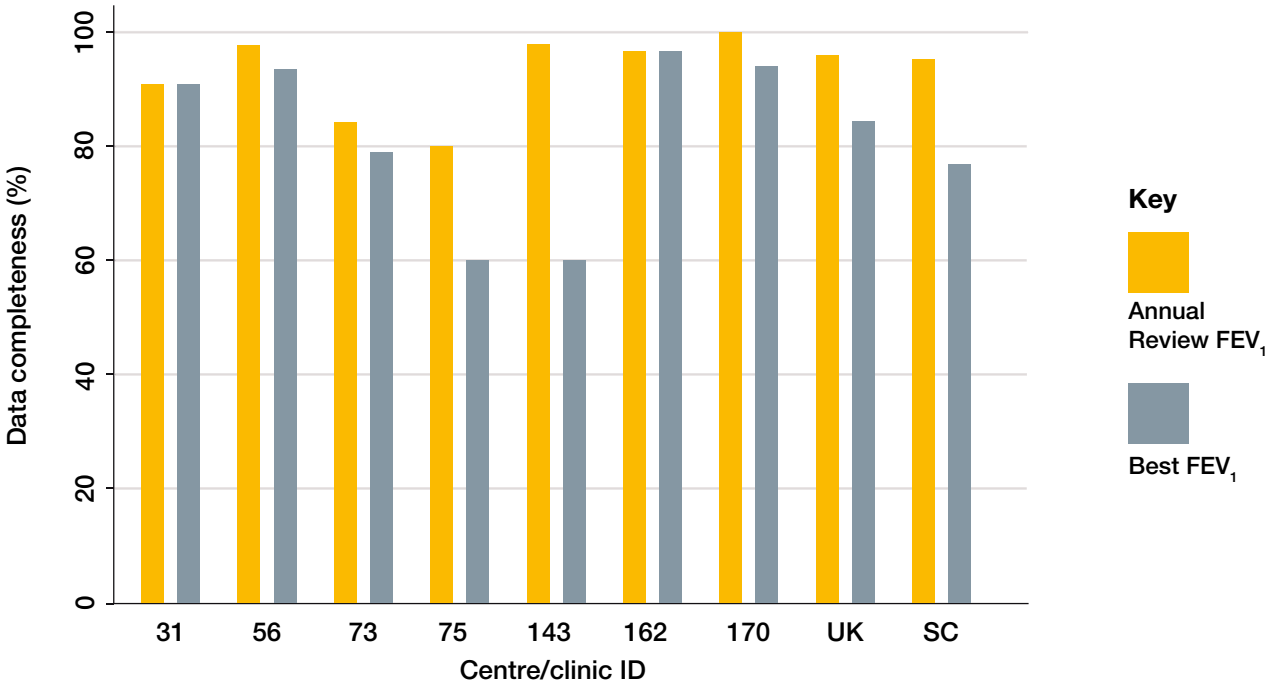
The median FEV_1 % predicted of patients attending paediatric centres/clinics in Scotland is 89.5% predicted (IQR: 80.0-98.2).

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



The median BMI percentile of patients attending paediatric centres/clinics in Scotland is 52 (IQR: 34-75).

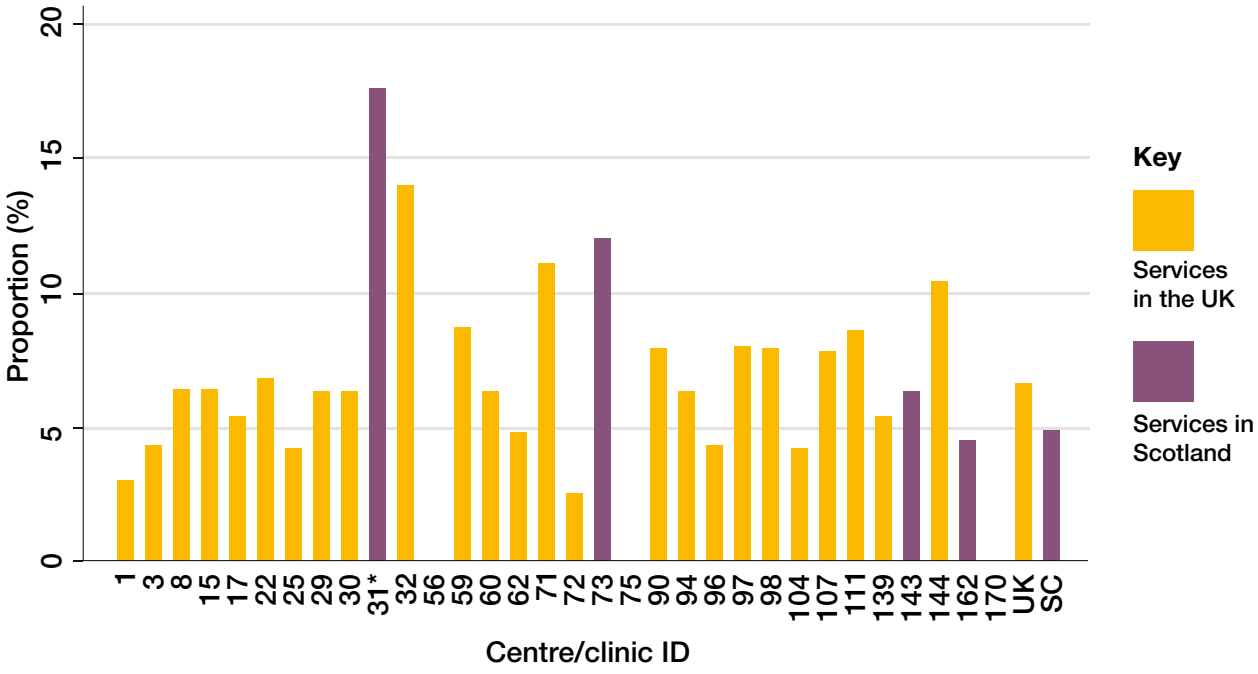
2.3 Data completeness by paediatric centre/clinic



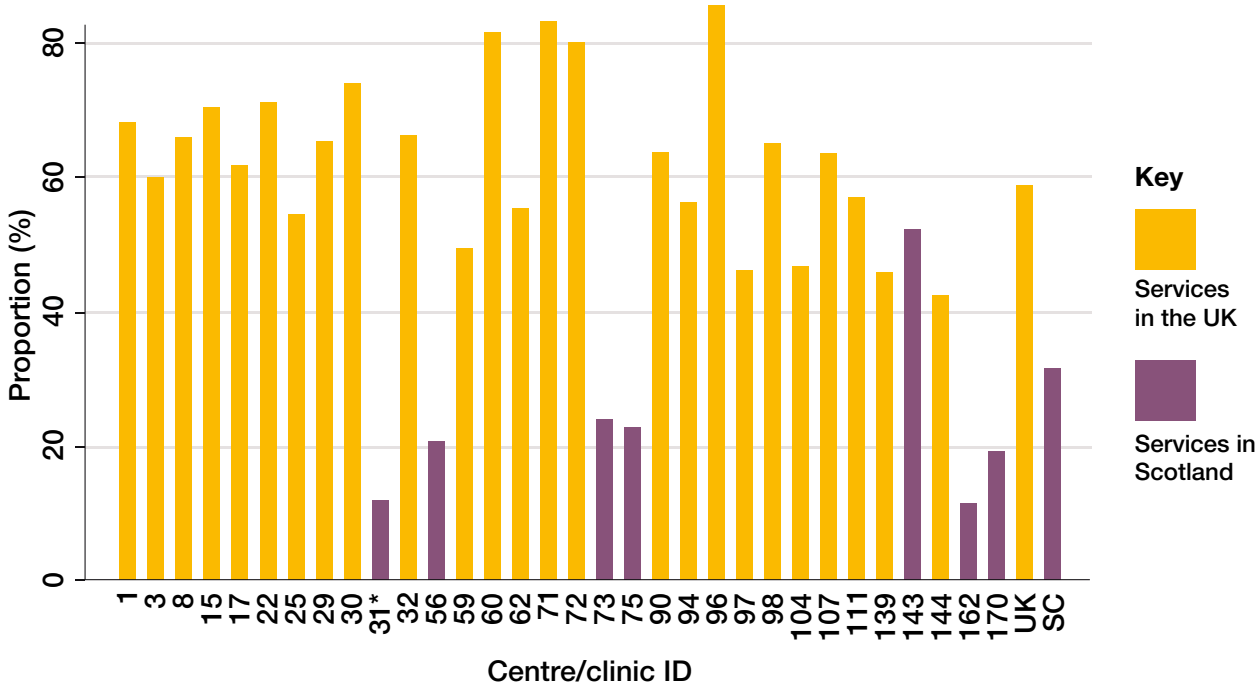
The mean data completeness for Annual Review FEV_1% predicted across Scottish paediatric centres is 95%. The mean data completeness for Best FEV_1 % predicted across Scottish paediatric centres is 77%.

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

4.8% of paediatric patients attending clinics in Scotland during 2017 were infected with chronic P. aeruginosa.

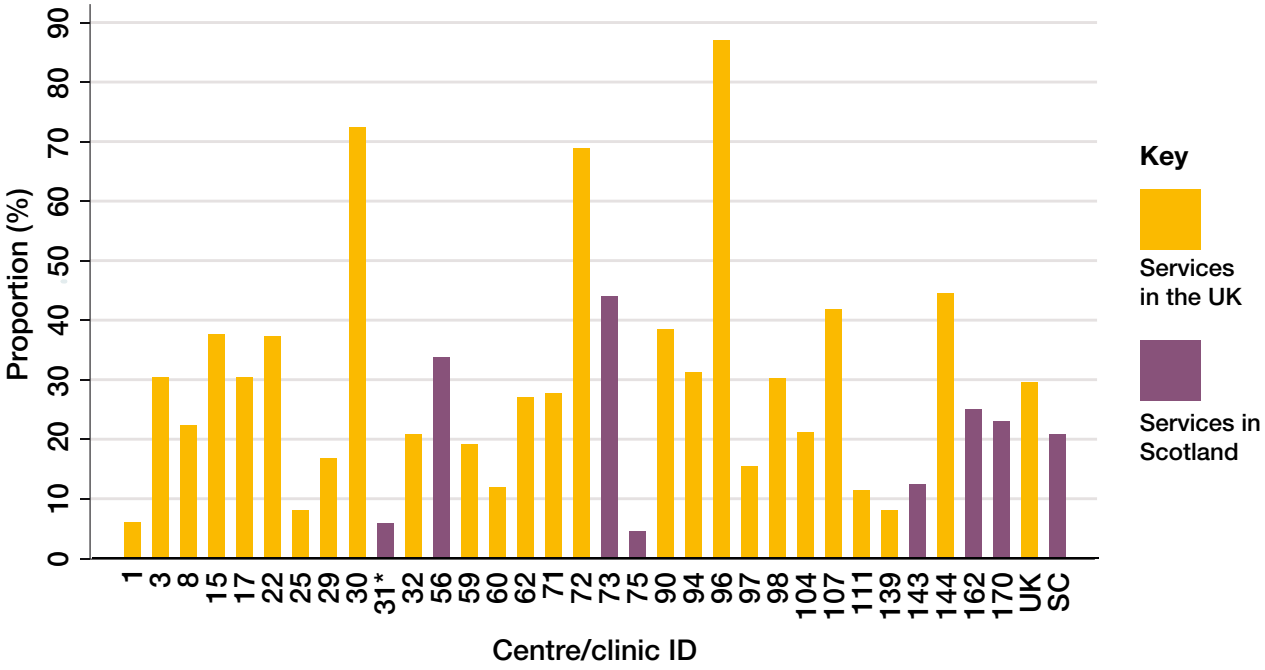


2.5 Proportion of patients receiving DNase treatment by paediatric centre/clinic



The proportion of patients attending paediatric centres/clinics in Scotland receiving DNase treatment is 31.5%.

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

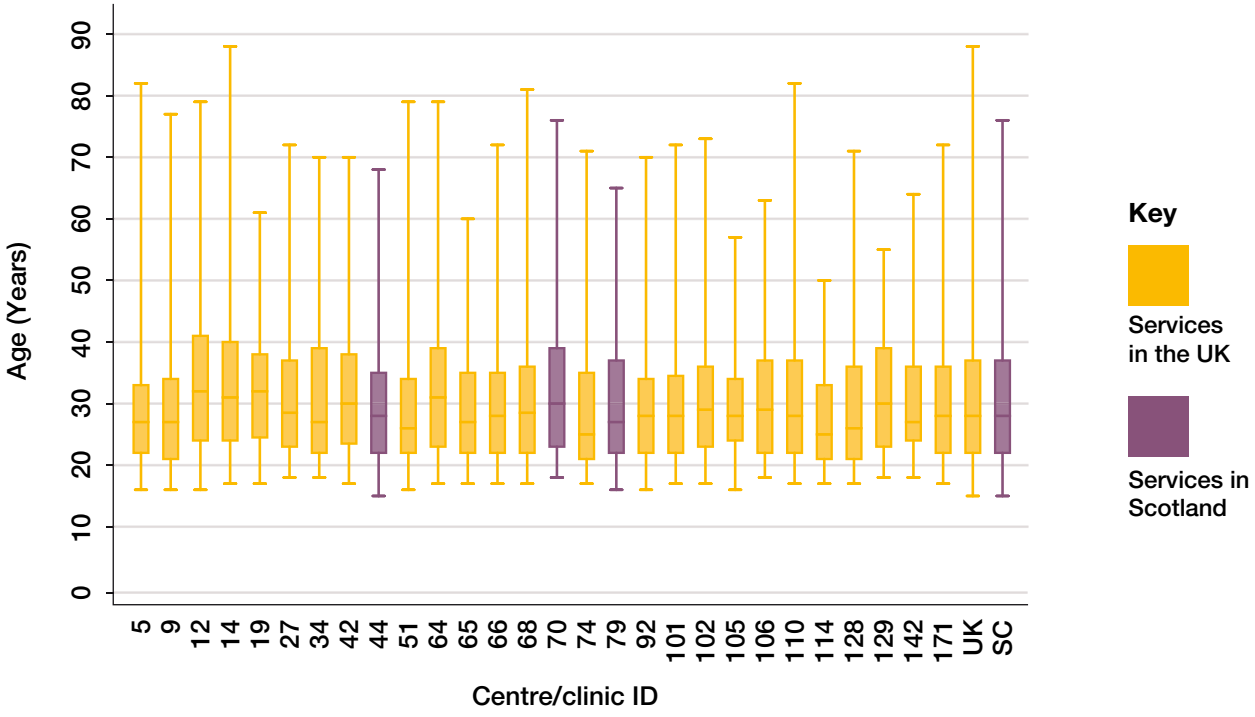


The proportion of patients attending paediatric centres/clinics in Scotland receiving DNase treatment is 20.9%.

Section 2b Adult centre analysis

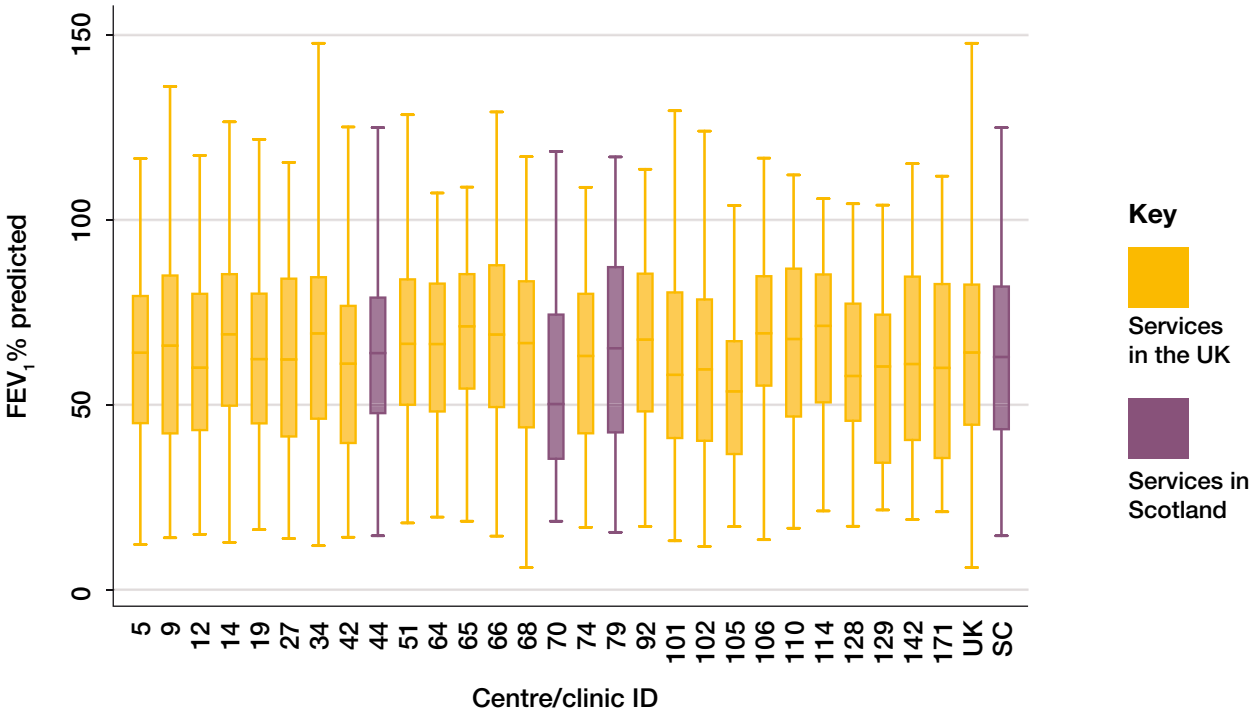
This section shows results for the three adult centres with their network clinics.

2.7 Median age (years) by adult service



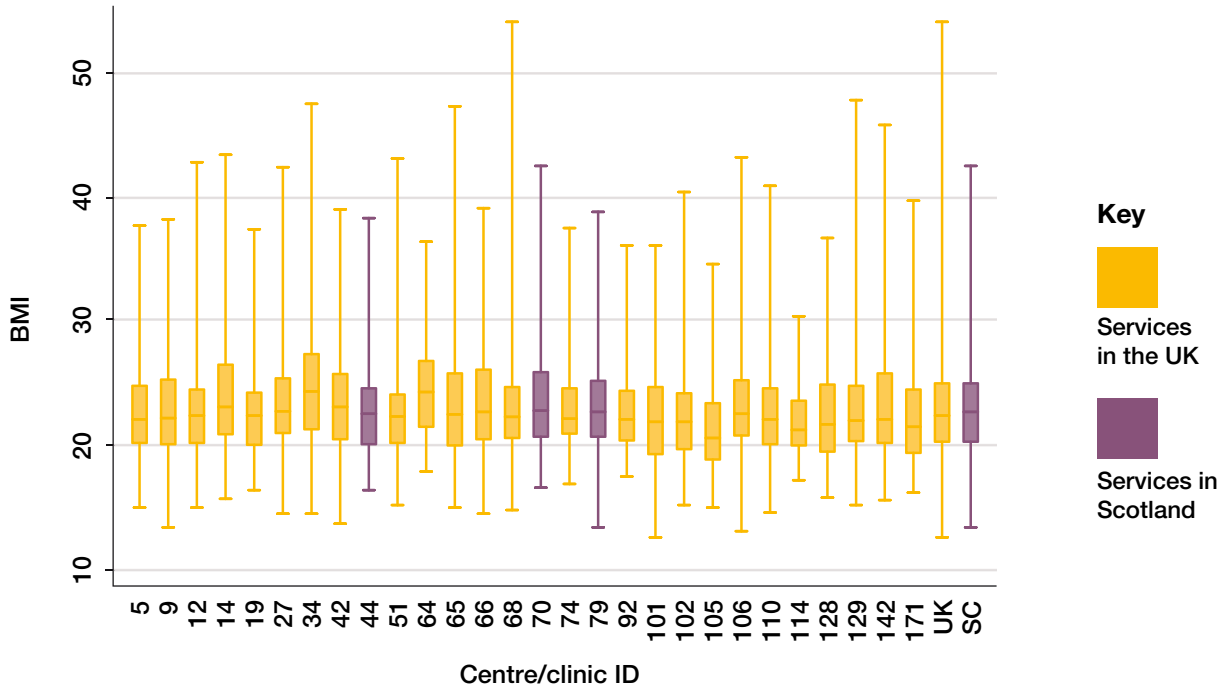
The median age of patients attending adult services in Scotland is 28 years (IQR: 22-37).

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



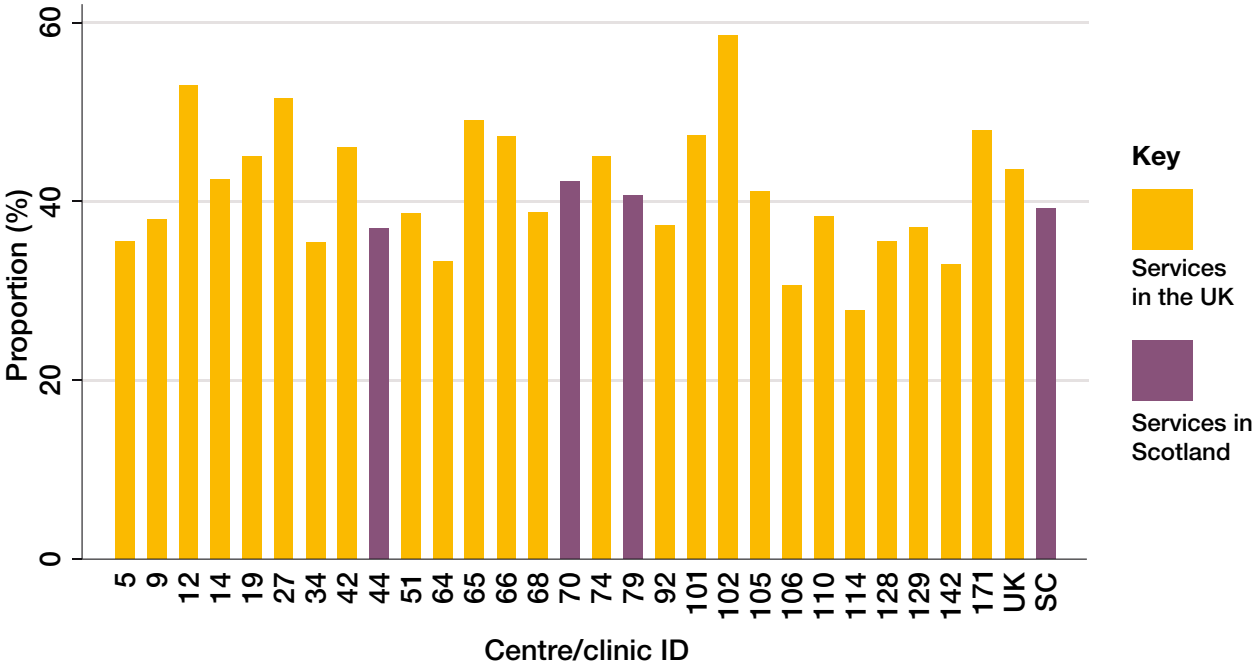
The median FEV_1 % predicted of patients attending adult services in Scotland is 63% (IQR: 43 – 82).

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



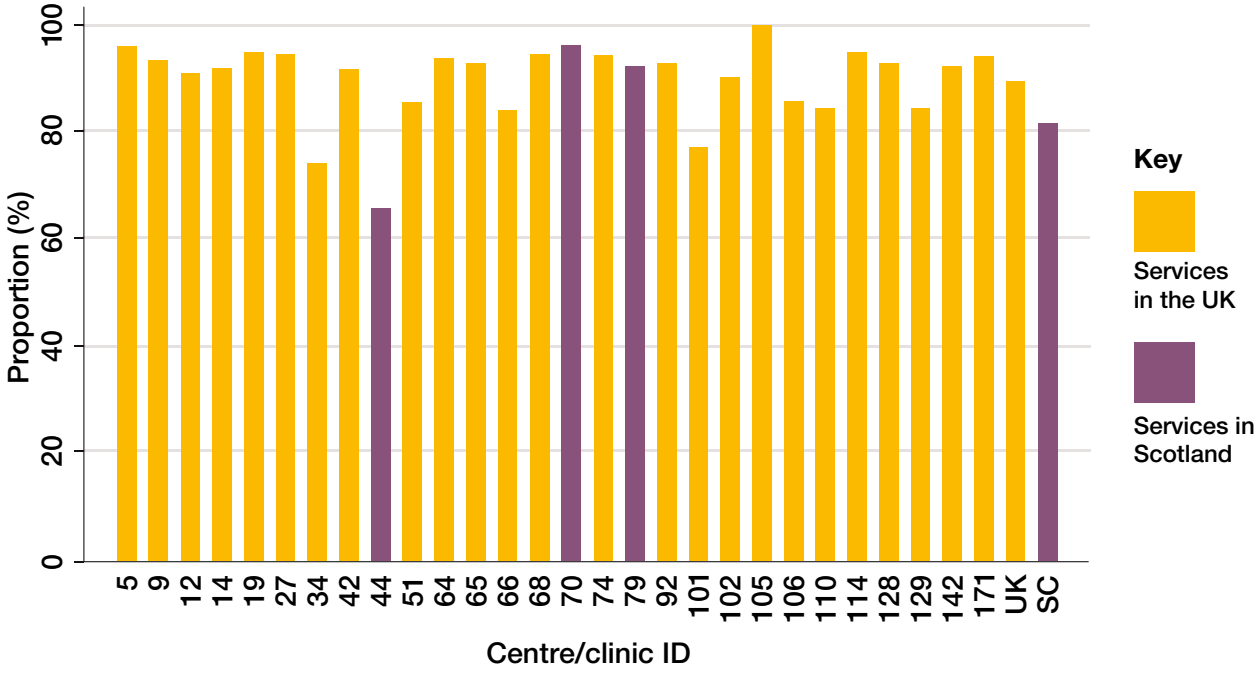
The median BMI of patients attending adult services in Scotland is 23 (IQR: 20 – 25).

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



The proportion of patients attending adult services in Scotland during 2017 with chronic *P. aeruginosa* is 39%.

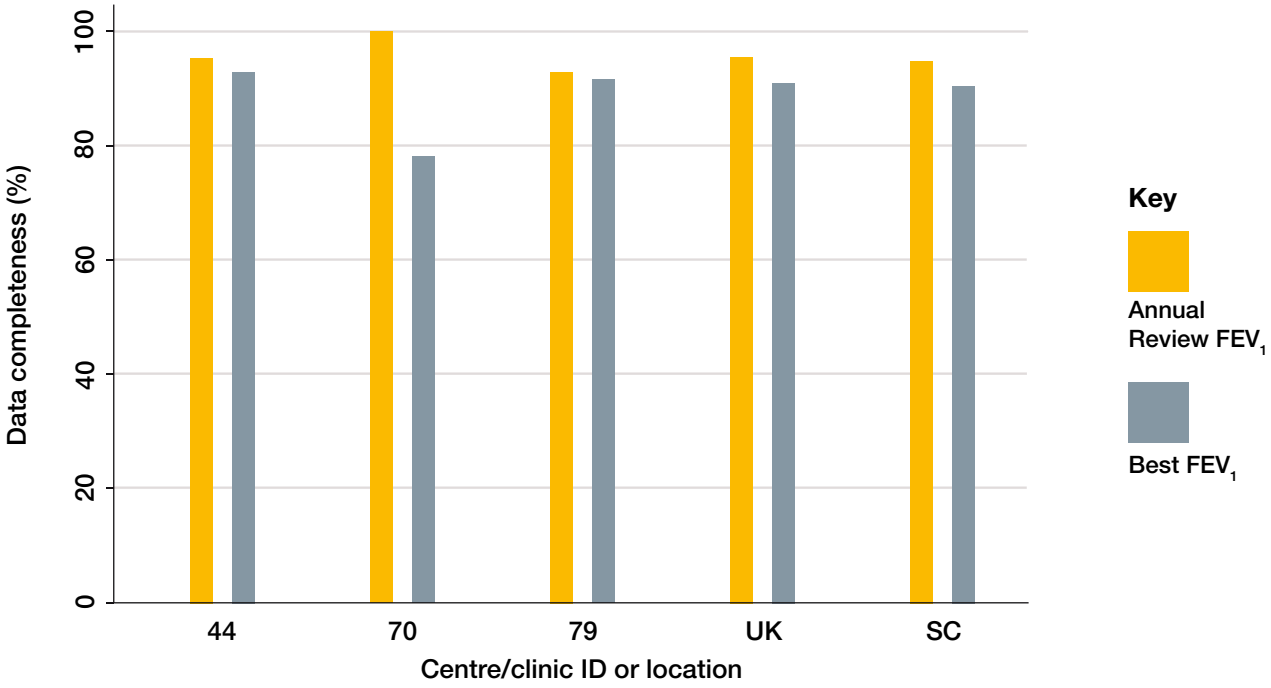
2.11 Inhaled antibiotic use for patients with chronic *P. aeruginosa* by adult service



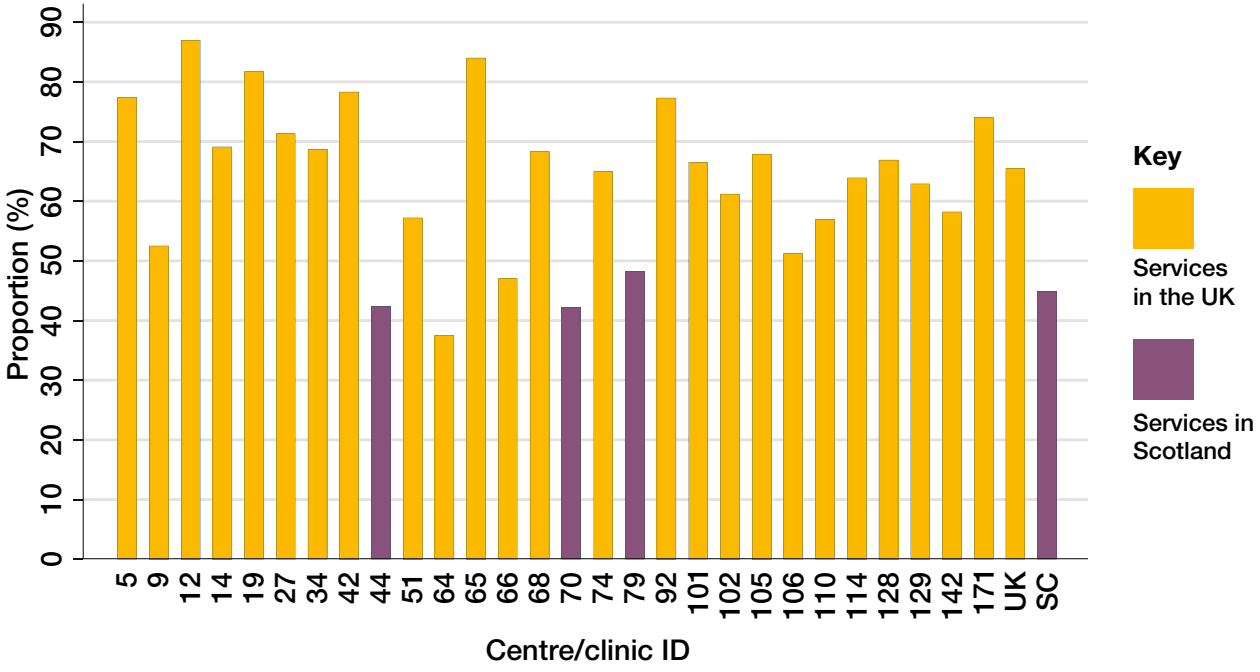
The proportion of chronic *P. aeruginosa* patients attending clinics in Scotland during 2017 is 81.6%.

2.12 Data completeness by adult service

The mean data completeness for Annual Review FEV_1% predicted across Scottish adult centres in 2017 is 95%. The mean data completeness for Best FEV_1 % predicted across adult Scottish centres is 91%.

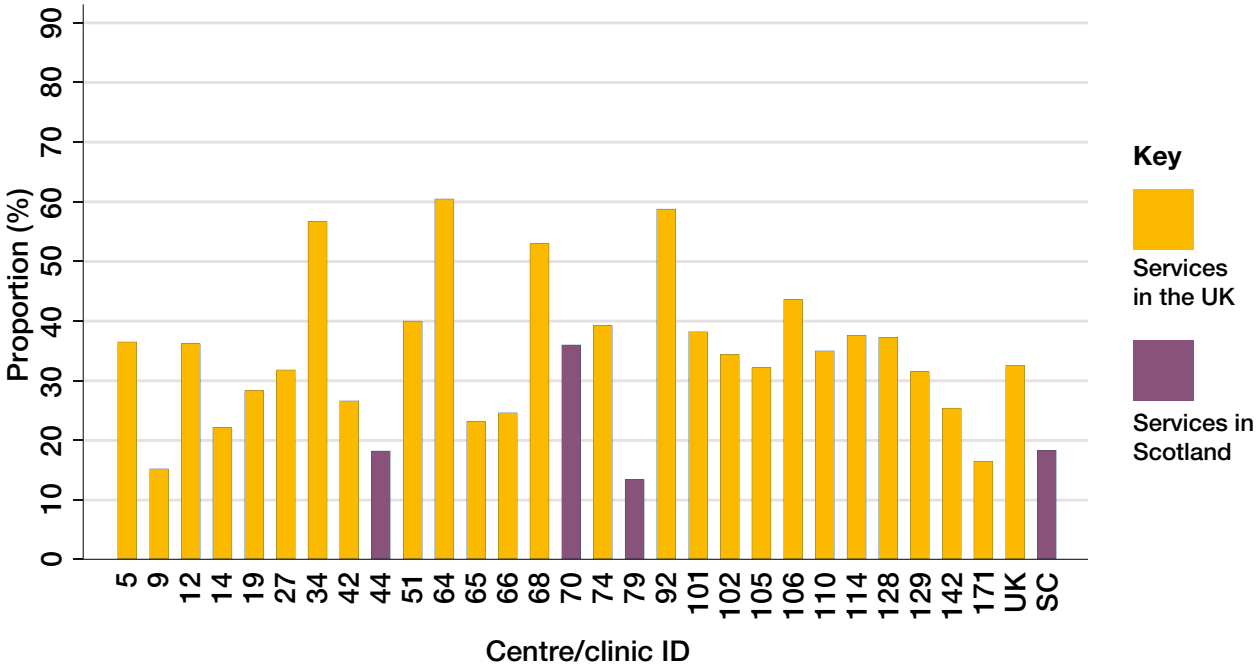


2.13 Proportion of patients receiving DNase treatment by adult service



The proportion of patients attending adult services in Scotland during 2017 receiving DNase treatment is 44.8%.

2.14 Proportion of patients receiving hypertonic saline treatment by adult service



The proportion of patients attending adult services in Scotland during 2017 receiving hypertonic saline treatment is 18%.

Appendices

Appendix 1: Centre-level data tables

Paediatric centres/clinics providing data in 2017 – ordered alphabetically

Location	Name	ID	Total active	Number with annual review	Age		FEV$_1$% predicted at annual review			
					Mean	Median	Number	Mean unadjusted	Mean adjusted †	Median
Scotland										
Aberdeen	Royal Aberdeen Children's Hospital	75	31	22	8.7	9.1	12	72.2	73.1	72.8
Ayr	University Hospital Crosshouse	170	26	26	8.7	8.6	17	87.7	88.4	87.9
Dundee	Ninewells Hospital	73	25	25	8.1	7.2	16	84.7	83.9	86.3
Edinburgh	Royal Hospital for Sick Children	143	131	128	8.9	9.0	93	90.6	90.0	90.7
Glasgow	Royal Hospital for Sick Children	56	95	68	8.5	8.8	45	89.8	89.7	91.5
Inverness	Raigmore Hospital	31	18	17	8.0	8.9	10	88.4	88.5	88.1
Lanarkshire	Wishaw General Hospital	162	48	44	8.3	7.7	29	91.2	90.4	91.4

Adult centres/clinics providing data in 2017 – ordered alphabetically

Location	Name	ID	Total active	Number with annual review	Age		FEV$_1$% predicted at annual review			
					Mean	Median	Number	Mean unadjusted	Mean adjusted †	Median
Scotland										
Aberdeen	Aberdeen Royal Infirmary	70	67	64	32.6	31.5	57	56.2	56.8	50.2
Edinburgh	Western General Hospital	44	245	238	32.1	29.9	209	63.6	63.4	64.0
Glasgow	Gartnavel General Hospital	79	237	226	31.6	27.7	205	64.9	64.6	65.3

*Where 'best' values were missing, or lower than FEV$_1$% predicted taken at annual review, the annual review value was used.
**For data completeness, 'best' values were taken to be valid if they were not missing and the percent predicted was not more than 0.5% lower than FEV$_1$% predicted taken at annual review.
† Adjusted for age - please see full UK CF Annual Report 2017.

Best FEV_1 % predicted				Data completeness for FEV_1			
Number*	Mean unadjusted	Mean adjusted †	Median	Number with valid best FEV_1**	Percentage with valid best FEV_1	Number with FEV_1 at annual review	Percentage with FEV_1 at annual review
15	84.8	85.2	89.3	9	60.0	12	80.0
17	97.6	98.2	101.8	16	94.1	17	100.0
16	90.6	89.7	95.8	15	78.9	16	84.2
94	103.6	103.0	93.0	57	60.0	93	97.9
45	96.0	95.7	96.8	43	93.5	45	97.8
10	94.3	94.4	94.0	10	90.9	10	90.9
30	95.8	94.8	96.4	29	96.7	29	96.7

Best FEV_1 % predicted				Data completeness for FEV_1			
Number*	Mean unadjusted	Mean adjusted †	Median	Number with valid best FEV_1**	Percentage with valid best FEV_1	Number with FEV_1 at annual review	Percentage with FEV_1 at annual review
57	60.8	61.5	57.8	50	78.1	64	100.0
209	70.8	70.6	70.3	221	92.9	227	95.4
211	70.9	70.5	72.4	207	91.6	210	92.9

Appendix 1: Centre-level data tables

Paediatric centres/clinics providing data in 2017 – ordered alphabetically

Location	Name	ID	BMI percentile				Chronic *pseudomonas*	
			Number	Mean unadjusted	Mean adjusted †	Median	Number	Proportion (%)
Scotland								
Aberdeen	Royal Aberdeen Children's Hospital	75	19	47.9	48.2	39.9	0	0
Ayr	University Hospital Crosshouse	170	17	67.4	67.7	70.5	0	0
Dundee	Ninewells Hospital	73	19	43.7	43.3	44.7	<5	12
Edinburgh	Royal Hospital for Sick Children	143	111	56.3	56.4	54.3	8	6.3
Glasgow	Royal Hospital for Sick Children	56	62	52.9	52.9	51.8	0	0
Inverness	Raigmore Hospital	31	10	58.0	57.9	61.5	<5	17.6
Lanarkshire	Wishaw General Hospital	162	32	52.5	52.3	55.0	<5	4.5

Adult centres/clinics providing data in 2017 – ordered alphabetically

Location	Name	ID	BMI				Chronic *pseudomonas*	
			Number	Mean unadjusted	Mean adjusted †	Median	Number	Proportion (%)
Scotland								
Aberdeen	Aberdeen Royal Infirmary	70	64	23.7	23.5	22.6	27	42.2
Edinburgh	Western General Hospital	44	229	22.7	22.6	22.5	88	37.0
Glasgow	Gartnavel General Hospital	79	218	22.7	22.8	22.7	92	40.7

† Adjusted for age - please see full UK CF Annual Report 2017.

Having at least 1 IV day		Receiving DNase treatment		Receiving hypertonic saline treatment		Inhaled antibiotic use among patients with chronic *pseudomonas*	
Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)
6	27.3	5	22.7	<5	4.5	0	0
6	23.1	5	19.2	6	23.1	0	0
6	24.0	6	24.0	11	44.0	<5	100
30	23.4	67	52.3	16	12.5	7	87.5
28	41.2	14	20.6	23	33.8	0	0
<5	23.5	<5	11.8	<5	5.9	<5	66.7
11	25.0	5	11.4	11	25.0	<5	100

Having at least 1 IV day		Receiving DNase treatment		Receiving hypertonic saline treatment		Inhaled antibiotic use among patients with chronic *pseudomonas*	
Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)	Number	Proportion (%)
26	40.6	27	42.2	23	35.9	26	96.3
101	42.4	101	42.4	43	18.1	58	65.9
88	38.9	109	48.2	30	13.3	85	92.4

Glossary

Word/Phrase	Meaning
2017	1 January 2017 – 31 December 2017
ABPA (allergic bronchopulmonary aspergillosis)	When a person develops a respiratory allergic reaction to Aspergillus fumigatus.
Arthritis	A condition causing pain and inflammation in the joints.
Arthropathy	A condition causing pain in the joints.
Asthma	A respiratory condition causing reversible episodes of difficulty breathing, often associated with wheezing.
B. cepacia complex	Burkholderia cepacia complex are a group of bacteria, some of which threaten the health of people with cystic fibrosis.
BMI (Body Mass Index)	A measure designed to show whether a person is a healthy weight for their height.
CF	Cystic fibrosis
CFTR (cystic fibrosis transmembrane conductance regulator)	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.
H. influenza	Haemophilus influenza is a bacterium that can cause serious illness.
Haemoptysis	The coughing up of blood.
Hepatobiliary disease	A liver or biliary disorder.
Heterozygous	Everyone living with cystic fibrosis has two mutations of the gene for CFTR, one inherited from their mother and one from their father. Someone who has two different mutations is heterozygous.

Word/Phrase	Meaning
Homozygous	Everyone living with cystic fibrosis has two mutations of the gene for CFTR, one inherited from their mother and one from their father. If both mutations (or genotypes) are the same, the person is said to be homozygous.
Hypertension	High blood pressure.
Incidence	The number of people newly diagnosed with a condition in the given year.
IQR (Interquartile range)	Also called the mid-spread, or middle fifty, IQR is a measure of the spread of data. It shows the difference between the upper and lower quartiles. IQR = Q3 – Q1.
Mean	A type of average, calculated by adding up all the values and dividing by the number of values.
Median	The middle number, when all numbers are arranged from smallest to largest.
Median age of death	Median age of death is based on the people with CF who died in any given year. So in 2017 the median age of the 132 people who died was 31.
Median predicted survival	A mathematical formula predicts how long we expect half of people with CF born today will live. Half of people born today are predicted to live to at least 47 years. Half of people are therefore predicted to die before they reach that age.
MRSA	Methicillin-resistant Staphylococcus aureus is a type of bacteria that is resistant to a number of widely used antibiotics.
Mutation	A mutation is a change in a gene. When both of a child's parents are carriers of a CF-causing mutation there is a 25% chance that the child will have cystic fibrosis. There are over 1,400 different mutations of the CFTR gene that can cause cystic fibrosis.
Nasal polyps	Small, sac-like growths of inflamed mucus caused by chronic inflammation of the nasal lining.
NBS (newborn screening)	Newborn screening is part of the heel prick blood spot testing done at 5-7 days of age. The blood sample is tested for a number of conditions, including cystic fibrosis.
NTM (Nontuberculous Mycobacteria)	A mycobacterium that does not cause tuberculosis, but which can cause respiratory infection. There are several types known.
Osteopenia	A medical condition less severe than osteoporosis, where the mineral content of bone is reduced.
Osteoporosis	A condition where the bones become brittle from loss of tissue.
Pancreas	An organ in the digestive system that produces insulin and digestive enzymes.
Pancreatitis	Inflammation of the pancreas.
Peptic ulcer	Or, stomach ulcer, is an open sore that develops in the lining of the stomach.
Percentile	A percentile shows where a value stands, relative to the rest of the data. If a value is higher than 90% of the rest of the data, it is at the 90th percentile.
Pneumothorax	A collection of air in the cavity between the lungs and the chest wall causing collapse of the lung on the affected side.
Portal hypertension	High blood pressure in the portal vein system, which is the blood system of the liver.
Prenatal	Before birth, while the baby is still in the womb.
Prevalence	The overall number of people with the condition in the last 12 months.
Pseudomonas aeruginosa	A tough bacterial strain. Rarely affecting healthy people, it can cause a wide range of infections, particularly in those with a weakened immune system.
Rectal prolapse	When the rectal wall slides through the anus.
Renal	Relating to the kidneys.
Staphylococcus aureus	Staphylococcus aureus is a bacterium that can cause disease if it enters the body.
Sinus disease	When the sinuses, which are usually filled with air, are typically full of thick sticky mucus.
Statistically significant	This phrase means that after careful calculations there is a definite difference between two groups, which is not simply a result of chance.

Appendix 2: UK CF Registry Steering Committee structure

UK CF Registry Steering Committee

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

UK CF Registry Research Committee

Role	Forename	Surname	Organisation
Pharmacovigilance PI CF physician – adults (retired)	Diana	Bilton	Royal Brompton Hospital
Registry consultant	Noreen	Caine	Cystic Fibrosis Trust
Pharmacovigilance PI CF physician - paediatrics	Siobhán	Carr	Royal Brompton Hospital
Senior Statistician †	Susan	Charman	Cystic Fibrosis Trust
Director of Data & Quality Improvement	Rebecca	Cosgriff	Cystic Fibrosis Trust
Pharmacovigilance PI CF physician - paediatrics	Steve	Cunningham	Royal Hospital for Sick Children
Parent representative	Marian	Dmochowska	N/A
Registry Clinical Data Manager †	Elaine	Gunn	Cystic Fibrosis Trust
Person with CF	Dominic	Kavanagh	Cystic Fibrosis Trust
Pharmacovigilance PI CF physician - adults	Nicholas	Simmonds	Royal Brompton Hospital
CF physician - adults*#	Martin	Wildman	Northern General Hospital
Registry Development Manager †	Mary	Yip	Cystic Fibrosis Trust

*Chair † Non-voting member # Caldicott guardian

Cystic Fibrosis Trust

cysticfibrosis.org.uk

Fighting for a Life Unlimited