Cystic Fibrosis strength in numbers

UK Cystic Fibrosis Registry Annual Data Report 2020

Scotland Version 2 – amended May 2022

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UK Cystic Fibrosis Registry Annual Data Report 2020 - Scotland

Report prepared by

Francis Adams Susan Charman Andrew Lee

With assistance from

Medical Statistician Senior Statistician Medical Statistician Cystic Fibrosis Trust Cystic Fibrosis Trust Cystic Fibrosis Trust

| Siaron Hughes | Freelance Graphic Designer | www.su-ma.com |
|---------------|-----------------------------|-----------------------|
| Elaine Gunn | Registry Data Manager | Cystic Fibrosis Trust |
| Sarah Clarke | Head of Healthcare Data and | Cystic Fibrosis Trust |
| | Pharmacovigilance | |

The UK CF Registry Steering Committee

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Contact information

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

registry@cysticfibrosis.org.uk



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3

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Contents

| Report prepared by Acknowledgements Contact information | 3 3 3 |
|--|--|
| Introduction | 6 |
| Cystic fibrosis UK Cystic Fibrosis Registry Governance Data collection Where can I find more information? How the Covid-19 Pandemic affected data entry 2020 | 6 6 7 7 7 8 |
| Section 1: Scotland-wide analysis | 10 |
| 1.1 Summary of the UK Cystic Fibrosis Registry 1.2 Age distribution by sex 1.3 Height percentiles of children and young people (<20 years) 1.4 Weight percentiles of children and young people (<20 years) 1.5 Body Mass Index BMI percentiles in children and young people (<20 years) 1.6 Body Mass Index BMI in adults (≥ 20 years) 1.7 Education and employment in adults (≥ 16 years) 1.8 Pregnancy | 10 11 12 13 14 15 16 16 |
| Diagnosis of cystic fibrosis | 17 |
| 1.9 Age at diagnosis | 17 |
| Lung health | 10 |
| 1.11 FEV % predicted (GLI equations) at Annual Review in patients aged 6 years and older | 20 |
| who have not had a lung transplant $1.12 \text{ Best FEV}_1\%$ predicted (GLI equations) in patients aged six years and older who have not had a lung transplant | 21 |
| 1.13 FEV ₁ % predicted (GLI equations) over time in patients aged six years and older who have not had a lung transplant | 22 |
| 1.14 FEV $_1$ % predicted (GLI equations) and BMI in people aged 20 years and over who have not had a transplant | 23 |
| Lung infections | 24 |
| 1.15 Lung infections in 2020 (graph) | 24 |
| 1.16 Lung infections in 2020 (table) | 25 |
| 1.17 Nontuberculous mycobacteria (NTM) or atypical mycobacteria | 27 |
| 1.18 Lung infections over time | 27 |
| Complications | 29 |
| 1.19 Complications in 2020 | 29 |
| 1.20 Incidence of complications | 30 |
| 1.21 Cystic tibrosis-related diabetes | 30 |
| 1.22 Intravenous (IV) antibiotics | 31 31 |
| 1.22 Initiavenous (IV) antibiotics | 23 |
| 1.24 Long-term azithromycin use | 33 |
| 1 25 Flucloxacillin | 34 |
| Muco-active therapies | 35 |
| 1.26 Mannitol | 35 |
| 1.27 DNase | 35 |
| 1.28 Hypertonic saline | 36 |
| 1.29 Burden of treatment | 36 |
| | |

| Other therapies 1.30 CFTR modulators 1.31 Oxygen and non-invasive ventilation 1.32 Physiotherapy 1.33 Feeding 1.34 Transplants | 37 37 38 38 39 39 |
|--|--|
| Genotypes 1.35 Mutation combinations in Scotland 1.36 Mutations in the Scottish population | 40 40 41 |
| Section 2 and 3: Centre-level analysis | 42 |
| A guide to the charts Box plots Section 2a Paediatric centre analysis 2.1 FEV, % predicted (GLI equations) among patients aged 6 and older by paediatric centre/clinic (without a history of lung transplant) 2.2 Body mass Index (BMI) percentile among patients aged 2-15 years by paediatric centre/clinic 2.3 Data completeness by paediatric centre/clinic 2.4 Proportion of patients with chronic <i>P. aeruginosa</i> 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 2.7 Age distribution by adult centre/clinic 2.8 FEV, % predicted (GLI equations) by adult centre/clinic (without a history of lung transplant) 2.9 Body Mass Index (BMI) distribution among patients aged 16 years and older by adult centre/clinic 2.10 Proportion of patients with chronic <i>Pseudomonas aeruginosa</i> by adult centre/clinic 2.11 Inhaled antibiotic use for patients with chronic <i>Pseudomonas aeruginosa</i> by centre/clinic 2.12 Data completeness by adult centre/clinic 2.13 Proportion of patients receiving DNase treatment by adult centre/clinic 2.10 Proportion of patients with chronic <i>Pseudomonas aeruginosa</i> by adult centre/clinic 2.11 Inhaled antibiotic use for patients with chronic <i>Pseudomonas aeruginosa</i> by centre/clinic 2.13 Proportion of patients receiving DNase treatment by adult centre/clinic 2.14 Proportion of patients receiving DNase treatment by adult centre/clinic 2.14 Proportion of patients receiving DNase treatment by adult centre/clinic 2.14 Proportion of patients receiving DNase treatment by adult centre/clinic | 43 43 44 44 45 45 46 47 48 48 49 50 50 51 51 |
| Glossary | 52 |
| Appendix 1: Centre-level data tables Paediatric centres/clinics providing data in 2020 – ordered alphabetically by location Adult centres/clinics providing data in 2020 – ordered alphabetically by location Paediatric centres/clinics providing data in 2020 – ordered alphabetically by location Adult centres/clinics providing data in 2020 – ordered alphabetically by location Adult centres/clinics providing data in 2020 – ordered alphabetically by location Adult centres/clinics providing data in 2020 – ordered alphabetically by location Appendix 2: Full list of mutations in the Scottish Population | 54 54 56 56 58 |

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

UK Cystic Fibrosis Registry

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

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



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



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



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



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



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 2020 Annual Data Report

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

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

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

Data collection

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

Where can I find more information?

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

How the pandemic affected data entry in 2020

We think it's important to show as much data as possible from 2020 about clinical care and health outcomes over-time, as well as a snapshot of how things were in that year. However, data entry into the Registry was affected by the COVID-19 pandemic in five key ways:

- **1.** Less face-to-face appointments
- **2.** Less testing of lung function and for lung infection
- **3.** CF team members were diverted to COVID-19 care
- **4.** More virtual appointments
- 5. More home spirometry equipment available to people with CF

This means it's hard for us to tell whether any changes we see in the 2020 data are:

- 'Real' (associated with the health and treatment of people with CF)
- Caused one or more of the five 'Pandemic factors'

...or a mixture of the two. Because of this, it is very important that results from 2020 are interpreted with caution. In the next year or two it will become clearer whether 2020 represents an unusual single year, or a new normal for people with cystic fibrosis. We have marked results that are at particular risk of being distorted because of the pandemic with one of these symbols:



Results for outcomes like lung function, complication rates, and infection prevalence may be different to normal because:

- There was less data available, and clinical teams had less time to enter it onto the system. This is a trend we have seen reflected in CF Registries around the world.
- For extended periods people with CF were advised to shield, and not leaving their homes. Clinical teams who care for people with cystic fibrosis were also diverted to front-line COVID-19 care. This meant that in-person appointments, and the clinical measurements that would happen during then, took place less often than normal.
- Not everyone with CF had access to spirometry equipment at home. Although funding was provided to support this, those that did get access to this equipment may not have done until after their 2020 Annual Review. We aren't sure if measurements taken at home are equivalent to those taken in clinic.
- Most CF centres didn't measure lung function in hospital during the pandemic because it's an Aerosol Generating Procedure and might have spread COVID-19.

We helped CF centres as much as we could to represent a true picture of the health of their patients during 2020. This included re-assigning an outpatient clinic appointment as an Annual Review, so that they could be included in this report. Where key data was not available at Annual review we used information provided at an encounter where it could sensibly be used as a substitute.

COVID-19 cases in Scotland

Please refer to the UK cystic fibrosis registry 2020 annual data report on page 32 for descriptive statistics of COVID-19 management and outcomes for people in the UK with CF infected with COVID-19 during the calendar year of 2020 (01 of January 2020–31 of December 2020). 15 of the 217 reported cases were in Scotland.

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

| | 2020 | | | | |
|---|--------------|-------------|--|--|--|
| | UK | Scotland | | | |
| CF patients registered ¹ | 10837 | 915 | | | |
| Excluding diagnoses that year | 10632 | 903 | | | |
| CF patients with an annual review; $n(\%)^2$ | 9922 (91.6%) | 814 (89%) | | | |
| Age in years; median ³ | 21 | 23 | | | |
| All newly diagnosed patients (newborn screening and other) ⁴ | 205 | 12 | | | |
| Number of patients born identified by newborn screening ⁴ | 152 | 9 | | | |
| Age at diagnosis in months; median ³ | 2 | 2 | | | |
| Adults aged 16 years and over; $\%^3$ | 60.6 | 63.4 | | | |
| Males; % ³ | 53.1 | 52.7 | | | |
| Genotyped; % ³ (both mutations identified) | 99.2 | 99.8 | | | |
| Total deaths reported (%) ⁵ | 97 (0.9%) | 18 (2%) | | | |
| Age at death in years; median (95% CI) 5 | 36 (32, 38) | 35 (27, 46) | | | |



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 (years)

| Age | All; n (%) | Females; n (%) | Males; n (%) |
|---------|------------|----------------|--------------|
| 0-3 | 48 (5.9) | 21 (5.5) | 27 (6.3) |
| 4-7 | 64 (7.9) | 33 (8.6) | 31 (7.2) |
| 8-11 | 95 (11.7) | 43 (11.2) | 52 (12.1) |
| 12-15 | 91 (11.2) | 44 (11.4) | 47 (11.0) |
| 16-19 | 52 (6.4) | 29 (7.5) | 23 (5.4) |
| 20-23 | 67 (8.2) | 30 (7.8) | 37 (8.6) |
| 24-27 | 88 (10.8) | 45 (11.7) | 43 (10.0) |
| 28-31 | 74 (9.1) | 33 (8.6) | 41 (9.6) |
| 32-35 | 53 (6.5) | 28 (7.3) | 25 (5.8) |
| 36-39 | 44 (5.4) | 19 (4.9) | 25 (5.8) |
| 40-43 | 45 (5.5) | 15 (3.9) | 30 (7.0) |
| 44-47 | 17 (2.1) | 11 (2.9) | 6 (1.4) |
| 48-51 | 26 (3.2) | 11 (2.9) | 15 (3.5) |
| 52-55 | 16 (2.0) | 7 (1.8) | 9 (2.1) |
| 56-59 | 22 (2.7) | 10 (2.6) | 12 (2.8) |
| 60+ | 12 (1.5) | 6 (1.6) | 6 (1.4) |
| <16 | 298 (36.6) | 141 (36.6) | 157 (36.6) |
| ≥16 | 516 (63.4) | 244 (63.4) | 272 (63.4) |
| <18 | 321 (39.4) | 154 (40.0) | 167 (38.9) |
| ≥18 | 493 (60.6) | 231 (60.0) | 262 (61.1) |
| Overall | 814 | 385 | 429 |

1.3 Height percentiles of children and young people (<20 years)¹ N=350

The following chart and table show the height percentiles of people with CF, aged 19 and under, in relation to UK growth data for the general population. If a person with CF is on the 40th percentile, only 40% of people the same age are their height or shorter; 60% are taller.



| Age | Overall | | Female | Female | | | Male | | |
|---------|---------|--------|-----------|--------|--------|-----------|------|--------|-----------|
| (years) | n | Median | IQR | n | Median | IQR | n | Median | IQR |
| 1 | 9 | 57.2 | 45.6-78.7 | <5 | - | - | - | 67.9 | 45.6-88.9 |
| 2 | 14 | 44.8 | 27.4-81.8 | 7 | 32.4 | 8.2-61.0 | 7 | 77.2 | 27.4-89.7 |
| 3 | 12 | 55.8 | 36.0-67.3 | 6 | 57.9 | 51.4-60.7 | 6 | 55.3 | 20.6-73.9 |
| 4 | 11 | 29.6 | 20.4-61.6 | 6 | 22.3 | 20.4-37.9 | 5 | 56.9 | 29.6-82.9 |
| 5 | 15 | 40.6 | 9.7-80.7 | 9 | 35.5 | 9.1-64.3 | 6 | 55.3 | 28.4-89.5 |
| 6 | 9 | 42.3 | 36.9-66.3 | 3 | 40.0 | 0.7-86.3 | 6 | 47.3 | 36.9-66.3 |
| 7 | 15 | 23.4 | 19.4-68.6 | 9 | 23.4 | 21.0-47.7 | 6 | 30.8 | 17.2-68.6 |
| 8 | 20 | 42.5 | 13.8-58.3 | 10 | 32.3 | 15.6-51.5 | 10 | 49.6 | 11.9-70.8 |
| 9 | 26 | 38.0 | 16.9-62.1 | 12 | 33.7 | 14.3-59.3 | 14 | 44.3 | 17.7-62.1 |
| 10 | 16 | 50.5 | 17.4-63.7 | <5 | - | - | - | 51.1 | 19.6-56.1 |
| 11 | 22 | 48.5 | 24.6-68.8 | 13 | 51.0 | 31.6-67.0 | 9 | 46.0 | 24.6-69.1 |
| 12 | 18 | 68.4 | 32.5-85.8 | 9 | 83.1 | 51.9-86.2 | 9 | 52.7 | 32.5-74.4 |
| 13 | 18 | 52.8 | 14.7-69.1 | 8 | 38.6 | 9.4-66.1 | 10 | 56.4 | 15.2-69.7 |
| 14 | 25 | 58.4 | 27.5-75.2 | 13 | 35.1 | 16.4-58.4 | 12 | 66.7 | 53.2-90.8 |
| 15 | 15 | 69.6 | 34.7-89.5 | 8 | 56.8 | 38.2-92.1 | 7 | 73.3 | 28.2-84.2 |
| 16 | 12 | 24.8 | 2.8-51.7 | 7 | 14.2 | 3.5-48.2 | 5 | 42.2 | 1.1-55.1 |
| 17 | 7 | 33.9 | 23.3-49.9 | <5 | - | - | <5 | - | - |
| 18 | 14 | 43.9 | 22.2-72.7 | 5 | 27.7 | 22.2-52.4 | 9 | 49.5 | 27.0-72.7 |
| 19 | 13 | 26.8 | 13.6-51.2 | - | 17.6 | 13.6-36.2 | <5 | - | - |
| Overall | 291 | 46.0 | 19.4-69.6 | 144 | 39.0 | 16.0-63.5 | 147 | 53.2 | 22.1-73.9 |

*number with non-missing data

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

1.4 Weight percentiles of children and young people (<20 years)¹ N=350

The following chart and table show the weight of people with CF, aged 19 and under, in relation to the UK growth data for the general population. If a person with CF is on the 40th percentile, only 40% of people the same age are their weight or lower; 60% weigh more.



| Age | Overall | | | Female | e. | | Male | | |
|---------|---------|--------|-----------|--------|--------|-----------|------|--------|-----------|
| (years) | n | Median | IQR | n | Median | IQR | n | Median | IQR |
| 1 | 11 | 67.7 | 21.3-89.3 | <5 | - | - | - | 67.7 | 21.3-94.9 |
| 2 | 17 | 67.9 | 34.0-77.1 | 8 | 48.6 | 25.5-68.0 | 9 | 76.0 | 52.4-87.2 |
| 3 | 16 | 60.8 | 12.7-81.1 | 7 | 74.4 | 52.8-85.8 | 9 | 31.4 | 11.6-70.4 |
| 4 | 13 | 59.5 | 20.5-68.2 | 6 | 28.3 | 19.1-67.8 | 7 | 59.7 | 28.4-85.2 |
| 5 | 18 | 48.1 | 21.5-76.8 | 10 | 44.2 | 14.7-66.7 | 8 | 58.5 | 37.7-95.0 |
| 6 | 10 | 37.7 | 22.7-70.9 | <5 | - | - | - | 30.0 | 22.7-72.2 |
| 7 | 15 | 40.9 | 18.3-65.1 | 9 | 38.6 | 28.1-51.5 | 6 | 44.8 | 17.3-65.1 |
| 8 | 19 | 60.8 | 15.6-87.2 | 10 | 55.6 | 17.9-64.9 | 9 | 63.2 | 15.6-92.0 |
| 9 | 27 | 49.2 | 20.6-63.3 | 12 | 45.9 | 26.1-65.6 | 15 | 49.2 | 19.6-63.3 |
| 10 | 16 | 65.3 | 35.7-84.2 | <5 | - | - | - | 68.8 | 41.5-85.9 |
| 11 | 22 | 54.1 | 33.9-81.8 | 13 | 55.9 | 25.3-87.8 | 9 | 52.2 | 41.5-78.2 |
| 12 | 19 | 56.6 | 40.4-84.2 | 10 | 69.3 | 45.1-87.8 | 9 | 45.2 | 40.4-77.3 |
| 13 | 19 | 56.4 | 18.6-65.0 | 9 | 45.0 | 8.1-62.1 | 10 | 57.2 | 22.0-65.0 |
| 14 | 26 | 59.7 | 37.2-70.4 | 13 | 51.7 | 24.2-69.3 | 13 | 67.1 | 46.4-70.4 |
| 15 | 18 | 58.8 | 42.9-82.7 | 9 | 50.2 | 42.9-68.2 | 9 | 81.2 | 56.8-85.0 |
| 16 | 12 | 44.2 | 28.1-89.5 | 7 | 40.8 | 2.1-92.0 | 5 | 47.5 | 30.9-82.4 |
| 17 | 7 | 17.9 | 9.0-63.8 | <5 | - | - | <5 | - | - |
| 18 | 12 | 49.8 | 10.0-90.4 | <5 | - | - | - | 57.0 | 17.1-92.8 |
| 19 | 13 | 28.0 | 3.1-75.2 | - | 37.5 | 6.7-75.2 | <5 | - | - |
| Overall | 310 | 52.0 | 24.9-76.9 | 150 | 48.3 | 20.5-75.2 | 160 | 56.9 | 28.1-80.6 |

*number with non-missing data

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

1.5 Body Mass Index BMI percentiles in children and young people (<20 years)¹ N=350

The following chart and table show the BMI percentiles of people with CF, aged 19 and under, in relation to the UK growth data for the general population. If a person with CF is on the 40th percentile, it means that only 40% of the population at the same age are their BMI or lower; so 60% have a higher BMI.



| Age | Overall | | Femal | Female | | | Male | | |
|---------|---------|--------|-----------|--------|--------|-----------|------|--------|-----------|
| (years) | n | Median | IQR | n | Median | IQR | n | Median | IQR |
| 1 | 9 | 50.1 | 35.0-84.8 | <5 | - | - | - | 61.8 | 32.1-86.6 |
| 2 | 14 | 66.5 | 41.4-84.1 | 7 | 42.8 | 16.1-70.0 | 7 | 77.9 | 65.7-86.9 |
| 3 | 12 | 51.1 | 27.8-92.1 | 6 | 66.1 | 47.3-89.0 | 6 | 39.6 | 19.6-95.2 |
| 4 | 11 | 66.6 | 29.6-72.1 | 6 | 38.3 | 29.6-67.0 | 5 | 69.5 | 66.6-72.1 |
| 5 | 15 | 58.5 | 42.6-78.1 | 9 | 51.1 | 42.6-62.8 | 6 | 65.3 | 45.2-85.7 |
| 6 | 9 | 39.4 | 15.6-77.2 | <5 | - | - | - | 41.8 | 15.6-77.2 |
| 7 | 15 | 57.4 | 24.2-72.4 | 9 | 61.1 | 24.2-72.4 | 6 | 55.3 | 29.7-68.1 |
| 8 | 18 | 59.6 | 25.0-84.9 | 10 | 59.6 | 24.0-75.5 | 8 | 55.7 | 33.8-90.9 |
| 9 | 26 | 57.6 | 29.3-74.6 | 12 | 64.1 | 27.8-74.3 | 14 | 50.3 | 31.2-74.6 |
| 10 | 16 | 69.2 | 33.5-91.0 | <5 | - | - | - | 74.3 | 48.1-94.6 |
| 11 | 21 | 68.7 | 43.8-84.5 | 12 | 76.8 | 27.8-92.2 | 9 | 65.7 | 50.3-80.0 |
| 12 | 18 | 54.9 | 38.5-82.1 | 9 | 54.6 | 40.0-82.1 | 9 | 55.2 | 31.0-67.3 |
| 13 | 18 | 49.8 | 25.0-66.3 | 8 | 51.4 | 13.9-75.1 | 10 | 49.8 | 27.8-66.3 |
| 14 | 24 | 50.1 | 23.9-73.1 | 13 | 50.1 | 23.5-88.0 | 11 | 48.1 | 24.3-70.2 |
| 15 | 14 | 55.6 | 39.1-71.7 | 8 | 55.6 | 44.3-65.1 | 6 | 56.1 | 23.7-81.7 |
| 16 | 11 | 79.6 | 26.2-96.2 | - | 79.6 | 22.2-96.2 | <5 | - | - |
| 17 | 7 | 20.4 | 16.9-57.7 | <5 | - | - | <5 | - | - |
| 18 | 12 | 55.2 | 14.1-93.1 | <5 | - | - | - | 54.4 | 14.1-95.3 |
| 19 | 13 | 45.3 | 18.1-75.3 | - | 61.8 | 22.3-75.3 | <5 | - | - |
| Overall | 283 | 55.9 | 29.6-80.5 | 141 | 54.9 | 26.2-79.6 | 142 | 56.5 | 31.0-81.5 |

*number with non-missing data

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

1.6 Body Mass Index BMI in adults (≥ 20 years) N=464

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 men¹.



| Age | Overall | | | Female | | | Male | | |
|---------|---------|--------|-----------|--------|--------|-----------|------|--------|-----------|
| (years) | n | Median | IQR | n | Median | IQR | n | Median | IQR |
| 20-23 | 60 | 22.2 | 20.8-25.2 | 25 | 22.8 | 20.7-25.1 | 35 | 21.9 | 20.8-25.3 |
| 24-27 | 71 | 22.6 | 20.4-25.8 | 34 | 23.1 | 20.6-26.0 | 37 | 22.5 | 20.4-25.1 |
| 28-31 | 56 | 23.2 | 21.1-26.4 | 22 | 22.1 | 20.8-24.3 | 34 | 23.8 | 21.2-27.2 |
| 32-35 | 42 | 23.9 | 20.8-25.2 | 23 | 23.5 | 20.4-25.3 | 19 | 24.1 | 21.6-24.7 |
| 36-39 | 39 | 23.2 | 20.7-26.5 | 17 | 21.6 | 20.5-25.6 | 22 | 23.9 | 21.7-26.5 |
| 40-43 | 37 | 23.8 | 21.2-26.1 | 12 | 22.2 | 21.0-25.0 | 25 | 24.3 | 21.9-26.5 |
| 44-47 | 15 | 25.3 | 21.7-28.1 | 9 | 25.3 | 24.1-26.8 | 6 | 25.2 | 21.7-28.1 |
| 48-51 | 18 | 24.5 | 22.4-28.6 | 7 | 22.4 | 21.4-23.6 | 11 | 25.4 | 24.3-30.0 |
| 52-55 | 13 | 24.1 | 23.1-27.9 | 6 | 23.6 | 23.0-25.5 | 7 | 24.2 | 23.1-28.4 |
| 56-59 | 17 | 24.9 | 23.0-29.5 | 8 | 25.8 | 21.5-30.0 | 9 | 24.9 | 23.8-28.9 |
| 60+ | 9 | 26.6 | 23.5-30.0 | - | 24.3 | 23.0-26.6 | <5 | - | - |
| Overall | 377 | 23.4 | 21.2-26.2 | 168 | 23.1 | 20.8-25.7 | 209 | 23.8 | 21.6-26.6 |

*number with non-missing data

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

1.7 Education and employment in adults (\geq **16 years)** N=516

The following table shows how people with CF reported their education and employment status in 2020. Please note that the groups are not mutually exclusive; for example, someone may be a student as well as working part-time.

| | All patients; n (%) | Male; n (%) | Female; n (%) |
|---|---------------------|-------------|---------------|
| Number of patients | 516 | 272 | 244 |
| Number who completed questionnaire; n (%) | 471 (91.3) | 245 (90.1) | 226 (92.6) |
| Full-time employment; n (%) | 182 (38.6) | 120 (49.0) | 62 (27.4) |
| Part-time employment; n (%) | 81 (17.2) | 27 (11.0) | 54 (23.9) |
| Student; n (%) | 65 (13.8) | 29 (11.8) | 36 (15.9) |
| Homemaker; n (%) | 15 (3.2) | 0 (0.0) | 15 (6.6) |
| Unemployed; n (%) | 83 (17.6) | 47 (19.2) | 36 (15.9) |
| Disabled; n (%) | 20 (4.2) | 9 (3.7) | 11 (4.9) |
| Retired; n (%) | 16 (3.4) | 8 (3.3) | 8 (3.5) |
| Unknown entered; n (%) | 7 (1.5) | 3 (1.2) | 4 (1.8) |
| Total patients in work or study; n (%) | 328 (69.6) | 176 (71.8) | 152 (67.3) |

1.8 Pregnancy



Fewer than five women with cystic fibrosis had babies in Scotland during 2020.



No men with cystic fibrosis became fathers in Scotland during 2020.

Diagnosis of cystic fibrosis

1.9 Age at diagnosis N=814



The median (range) age at diagnosis for patients aged under 16 in 2020 is 19 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 2020 were identified by newborn screening (including those without complete data).

78 (9.6%) 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 2020.

1.10 Mode of presentation

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

| | All patients | Age <16 at diagnosis* | Age ≥16 at diagnosis* |
|-------------------------|--------------|-----------------------|-----------------------|
| Total patients | 915 | 824 | 90 |
| Number diagnosed by NBS | 316 | 316 | 0 |
| Total non-NBS | 599 | 508 | 90 |

| Mode of presentation | All patients (n=551) | | Age <16 at diagnosis* (n=469) | | Age ≥16 at diagnosis* (n=82) | |
|---|-------------------------|------|----------------------------------|------|---------------------------------|------|
| (excluding NBS) | n | (%) | n | (%) | n | (%) |
| Persistant or acute respiratory infection | 201 | 36.5 | 156 | 33.3 | 45 | 54.9 |
| Failure to thrive/malnutrition | 144 | 26.1 | 144 | 30.7 | 0 | 0.0 |
| Abnormal stools/fatty stool(steator- rhea)/malabsorption | 117 | 21.2 | 112 | 23.9 | 5 | 6.1 |
| Meconium ileus | 90 | 16.3 | 90 | 19.2 | 0 | 0.0 |
| Family history | 80 | 14.5 | 68 | 14.5 | 12 | 14.6 |
| Unknown | 42 | 7.6 | 32 | 6.8 | 10 | 12.2 |
| Genotype | 24 | 4.4 | 19 | 4.1 | 5 | 6.1 |
| Electrolyte imbalance | 21 | 3.8 | - | - | <5 | - |
| Rectal prolapse | 14 | 2.5 | 14 | 3.0 | 0 | 0.0 |
| Bronchiectasis | 9 | 1.6 | <5 | - | - | - |
| Fertility | 7 | 1.3 | 0 | 0.0 | 7 | 8.5 |
| Nasal polyps | <5 | - | <5 | - | <5 | 2.4 |
| Liver disease | <5 | - | <5 | - | 0 | 0.0 |
| Prenatal | <5 | - | <5 | - | 0 | 0.0 |
| Oedema | <5 | - | <5 | - | 0 | 0.0 |
| Pancreatitis | 0 | 0.0 | 0 | 0.0 | 0 | 0.0 |



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

Lung health

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

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

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

For people with CF, an $\text{FEV}_1\%$ predicted of 85% or higher is the target, as this indicates normal or nearnormal 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 FEV₁% 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 FEV₁% predicted values shown in this report are calculated using an equation called Global Lungs Initiative, or 'GLI'¹.



1.11 FEV_1 % predicted (GLI equations) at Annual Review in patients aged 6 years and older who have not had a lung transplant N=501

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 | Female | | | Male | | |
|-----------|---------|--------|------------|--------|--------|------------|------|--------|------------|
| Age (yrs) | n | Median | IQR | n | Median | IQR | n | Median | IQR |
| 6-7 | 16 | 96.7 | 89.0-103.5 | 8 | 96.7 | 92.3-103.5 | 8 | 97.4 | 84.9-103.7 |
| 8-9 | 37 | 93.4 | 83.7-101.8 | 19 | 98.1 | 86.9-102.1 | 18 | 88.6 | 81.1-97.6 |
| 10-11 | 31 | 94.2 | 82.3-104.4 | 13 | 95.0 | 85.1-103.1 | 18 | 92.1 | 82.2-105.7 |
| 12-15 | 66 | 90.6 | 80.0-98.2 | 33 | 88.4 | 78.6-98.3 | 33 | 91.0 | 81.7-97.2 |
| 16-19 | 42 | 85.1 | 59.3-99.4 | 22 | 88.3 | 59.3-100.9 | 20 | 81.7 | 62.4-93.8 |
| 20-23 | 50 | 79.0 | 58.7-93.9 | 19 | 71.6 | 57.3-83.2 | 31 | 82.5 | 58.7-95.2 |
| 24-27 | 62 | 60.0 | 38.7-82.6 | 30 | 64.3 | 39.0-89.1 | 32 | 57.3 | 37.8-67.3 |
| 28-31 | 43 | 59.8 | 40.9-76.5 | 19 | 64.9 | 49.6-81.1 | 24 | 53.9 | 33.7-75.0 |
| 32-35 | 36 | 67.4 | 53.1-80.1 | 18 | 68.3 | 56.1-83.1 | 18 | 66.5 | 50.1-79.0 |
| 36-39 | 30 | 56.4 | 45.2-81.3 | 13 | 56.2 | 48.7-69.7 | 17 | 56.5 | 38.8-88.4 |
| 40-43 | 28 | 57.6 | 39.1-86.5 | 11 | 56.1 | 38.9-81.8 | 17 | 58.7 | 41.8-103.1 |
| 44-47 | 12 | 63.7 | 45.2-81.0 | - | 47.0 | 43.3-70.3 | <5 | - | - |
| 48-51 | 18 | 59.8 | 46.7-67.7 | 8 | 62.8 | 55.7-67.8 | 10 | 57.3 | 39.1-67.7 |
| 52-55 | 11 | 64.2 | 43.1-77.8 | 6 | 57.2 | 43.1-75.7 | 5 | 65.5 | 64.2-77.8 |
| 56-59 | 11 | 55.1 | 46.5-68.1 | - | 67.0 | 52.3-78.5 | <5 | - | - |
| 60+ | 8 | 41.4 | 31.3-47.0 | <5 | - | - | <5 | - | - |
| <16 | 150 | 92.6 | 82.2-99.8 | 73 | 92.9 | 82.9-102.0 | 77 | 91.0 | 81.7-99.3 |
| ≥16 | 351 | 65.4 | 44.4-83.9 | 165 | 67.0 | 48.6-83.1 | 186 | 65.0 | 41.5-84.8 |
| <18 | 167 | 91.8 | 81.7-99.8 | 83 | 92.4 | 82.7-102.0 | 84 | 90.6 | 81.4-99.3 |
| ≥18 | 334 | 64.7 | 43.7-82.6 | 155 | 64.9 | 48.2-81.8 | 179 | 64.4 | 41.4-83.7 |
| Overall | 501* | 77.8 | 53.5-92.3 | 238 | 78.3 | 55.3-92.8 | 263 | 76.9 | 50.1-92.0 |

*number with non-missing data

1.12 Best FEV_1 % predicted (GLI equations) in patients aged 6 years and older who have not had a lung transplant N=657

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.



Age (years)

| | Overal | I | | Female | | | Male | | |
|-----------|--------|--------|------------|--------|--------|------------|------|--------|------------|
| Age (yrs) | n | Median | IQR | n | Median | IQR | n | Median | IQR |
| 6-7 | 25 | 101.3 | 95.4-109.3 | 12 | 99.3 | 89.1-109.0 | 13 | 106.4 | 95.7-109.3 |
| 8-9 | 44 | 96.9 | 86.5-103.0 | 22 | 98.3 | 90.8-102.1 | 22 | 94.6 | 83.7-105.2 |
| 10-11 | 41 | 94.2 | 86.0-106.9 | 18 | 93.5 | 86.0-103.2 | 23 | 96.2 | 83.4-108.2 |
| 12-15 | 82 | 91.8 | 81.7-98.8 | 40 | 91.2 | 80.8-100.9 | 42 | 93.6 | 83.2-98.6 |
| 16-19 | 50 | 88.2 | 74.9-99.9 | 28 | 89.6 | 82.5-100.5 | 22 | 85.6 | 66.6-99.4 |
| 20-23 | 64 | 80.5 | 58.7-94.0 | 28 | 77.0 | 58.7-87.8 | 36 | 84.9 | 58.6-95.3 |
| 24-27 | 81 | 62.8 | 41.4-83.7 | 42 | 61.1 | 44.3-89.7 | 39 | 64.4 | 40.1-75.5 |
| 28-31 | 68 | 63.5 | 43.5-80.0 | 30 | 64.7 | 52.7-82.2 | 38 | 57.8 | 40.5-78.5 |
| 32-35 | 48 | 71.4 | 60.0-80.1 | 24 | 71.8 | 60.7-80.3 | 24 | 70.1 | 55.0-80.1 |
| 36-39 | 37 | 58.5 | 50.3-81.3 | 16 | 58.5 | 49.3-76.2 | 21 | 57.5 | 54.2-88.4 |
| 40-43 | 37 | 61.4 | 44.1-85.4 | 13 | 58.0 | 49.4-85.4 | 24 | 63.4 | 43.9-84.8 |
| 44-47 | 15 | 63.0 | 45.8-81.8 | 10 | 47.7 | 44.7-63.0 | 5 | 81.8 | 74.5-81.8 |
| 48-51 | 22 | 62.8 | 56.9-69.3 | 9 | 63.5 | 61.6-67.8 | 13 | 61.7 | 56.8-73.4 |
| 52-55 | 15 | 64.2 | 47.8-78.5 | 6 | 61.4 | 53.1-79.7 | 9 | 64.2 | 47.8-77.8 |
| 56-59 | 18 | 68.0 | 48.6-82.7 | 9 | 67.9 | 52.3-72.6 | 9 | 68.8 | 43.5-92.9 |
| 60+ | 10 | 43.2 | 33.8-52.7 | 5 | 37.2 | 26.4-46.6 | 5 | 52.7 | 39.7-80.6 |
| <16 | 192 | 95.2 | 83.7-103.2 | 92 | 93.8 | 82.8-103.1 | 100 | 95.7 | 84.3-104.8 |
| ≥16 | 465 | 67.8 | 49.6-86.4 | 220 | 67.9 | 52.4-86.4 | 245 | 67.7 | 47.0-86.4 |
| <18 | 213 | 93.9 | 83.6-103.1 | 104 | 93.7 | 82.9-102.8 | 109 | 94.2 | 83.7-103.7 |
| ≥18 | 444 | 67.3 | 48.5-85.6 | 208 | 67.4 | 52.3-83.9 | 236 | 67.3 | 44.9-86.5 |
| Overall | 657* | 79.7 | 57.2-93.9 | 312 | 79.6 | 58.5-93.9 | 345 | 80.3 | 56.8-93.9 |

*number with non-missing data

1.13 FEV₁% predicted (GLI equations) over time in patients 6 years and older who have not had a lung transplant N=701 in 2020, N=655 in 2015, N=513 in 2010*

The chart below shows how FEV_1 % in 2020 compares to Registry data from 2010 and 2015. 2010 is shown as a comparator year.



Age (years)

| Age (years) | 2010 mean FEV ₁ % | 2015 mean FEV ₁ % | 2020 mean FEV₁%* | p-values (t-test)** |
|-------------|------------------------------|------------------------------|------------------------------------|---------------------|
| 6-7 | 89.1 | 92.4 | 96.5 | 0.345 |
| 8-9 | 89.2 | 92.4 | 92.6 | 0.940 |
| 10-11 | 79.8 | 90.5 | 92.2 | 0.614 |
| 12-15 | 73.1 | 83.6 | 88.8 | 0.080 |
| 16-19 | 65.0 | 73.7 | 78.7 | 0.333 |
| 20-23 | 59.6 | 59.7 | 76.0 | <0.001 |
| 24-27 | 58.6 | 60.3 | 59.7 | 0.885 |
| 28-31 | 56.1 | 57.8 | 60.5 | 0.580 |
| 32-35 | 66.0 | 57.3 | 65.8 | 0.075 |
| 36-39 | 51.8 | 60.6 | 60.3 | 0.958 |
| 40-43 | 67.2 | 58.5 | 64.5 | 0.484 |
| 44-47 | 54.8 | 61.2 | 62.0 | 0.926 |
| 48-51 | 62.0 | 61.7 | 59.6 | 0.799 |
| 52-55 | 57.3 | 63.6 | 61.6 | *** |
| 56-59 | 46.8 | 66.2 | 57.5 | *** |
| 60+ | 64.7 | 45.6 | 42.9 | *** |
| <16 | 80.4 | 89.2 | 91.2 | N/A |
| 16+ | 60.9 | 61.9 | 65.1 | N/A |
| <18 | 77.7 | 87.8 | 90.5 | N/A |
| 18+ | 59.4 | 60.7 | 64.2 | N/A |

* due to missing data, means are calculated from a population of 501 in 2020, 617 in 2015 and 428 in 2010.

** t-test comparing 2020 with 2015. 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.

1.14 FEV₁% predicted (GLI equations) and BMI in people aged 20 years and over who have not had a transplant N=434

The goal BMI for adults is 22 for women, and 23 for men¹. The chart below shows the relationship between BMI and FEV₁% predicted. A healthy BMI can help to protect people with CF against lung infection and help to preserve lung health. This chart excludes people who have had a lung transplant.



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.



24 UK Cystic Fibrosis Registry Annual Data Report 2020



1.16 Lung infections in 2020 <16 years N=298; ≥16 years N=516

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 | 48 | 64 | 95 | 91 | 298 |
| Number who had culture taken* | 48 | 62 | 93 | 89 | 292 |
| Chronic S. aureus n (%) | <5 | - | 14 (14.7) | 9 (9.9) | 31 (10.4) |
| Intermittent S. aureus n (%) | 13 (27.1) | 11 (17.2) | 27 (28.4) | 31 (34.1) | 82 (27.5) |
| Chronic P. aeruginosa n (%) | 0 (0.0) | <5 | 5 (5.3) | <5 | 12 (4.0) |
| Intermittent P. aeruginosa n (%) | 6 (12.5) | 5 (7.8) | 15 (15.8) | 11 (12.1) | 37 (12.4) |
| B. cepacia complex n (%) | <5 | <5 | <5 | <5 | 8 (2.7) |
| <i>B. cenocepacia</i> n (%) | 0 (0.0) | 0 (0.0) | 0 (0.0) | <5 | <5 |
| <i>B. multivorans</i> n (%) | 0 (0.0) | 0 (0.0) | 0 (0.0) | <5 | <5 |
| B. other cepacia n (%) | <5 | 0 (0.0) | <5 | 0 (0.0) | <5 |
| MRSA n (%) | 0 (0.0) | 0 (0.0) | 0 (0.0) | <5 | <5 |
| H. influenza n (%) | 15 (31.3) | 20 (31.3) | 10 (10.5) | 11 (12.1) | 56 (18.8) |
| NTM n (%) | 0 (0.0) | 0 (0.0) | <5 | - | 12 (4.0) |
| Aspergillus n (%) | <5 | <5 | 6 (6.3) | 12 (13.2) | 22 (7.4) |

| | Adult Age Range (Years) | | | | | | Overall |
|--|-------------------------|-----------|-----------|-----------|-----------|-----------|-----------------------|
| | 16-19 | 20-23 | 24-27 | 28-31 | 32-35 | 36-39 | Adults (≥16 years) |
| Number in age range | 52 | 67 | 88 | 74 | 53 | 44 | 516 |
| Number who had culture taken* | 39 | 35 | 46 | 30 | 34 | 26 | 276 |
| Chronic S. aureus n (%) | 12 (23.1) | 14 (20.9) | 19 (21.6) | 11 (14.9) | 7 (13.2) | 7 (15.9) | 84 (16.3) |
| Intermittent <i>S. aureus</i> n (%) | 9 (17.3) | 9 (13.4) | 11 (12.5) | 7 (9.5) | 12 (22.6) | <5 | 61 (11.8) |
| Chronic <i>P. aeruginosa</i> n (%) | 8 (15.4) | 7 (10.4) | 11 (12.5) | 12 (16.2) | 7 (13.2) | 10 (22.7) | 78 (15.1) |
| Intermittent <i>P.</i> aeruginosa n (%) | <5 | <5 | 8 (9.1) | 6 (8.1) | 6 (11.3) | <5 | 41 (7.9) |
| <i>B. cepacia complex</i> n (%) | 0 (0.0) | <5 | <5 | <5 | 6 (11.3) | <5 | 23 (4.5) |
| <i>B. cenocepacia</i> n (%) | 0 (0.0) | <5 | 0 (0.0) | <5 | <5 | <5 | 6 (1.2) |
| B. multivorans n (%) | 0 (0.0) | <5 | <5 | 0 (0.0) | <5 | <5 | 11 (2.1) |
| B. other cepacia n (%) | 0 (0.0) | <5 | <5 | <5 | <5 | 0 (0.0) | 6 (1.2) |
| <i>M</i> RSA n (%) | 0 (0.0) | 0 (0.0) | <5 | 0 (0.0) | <5 | <5 | 4 (0.8) |
| <i>H. influenza</i> n (%) | 13 (25.0) | <5 | 7 (8.0) | 5 (6.8) | 5 (9.4) | <5 | 48 (9.3) |
| <i>NTM</i> n (%) | 5 (9.6) | 7 (10.4) | 7 (8.0) | <5 | <5 | <5 | 29 (5.6) |
| Aspergillus n (%) | 7 (13.5) | 9 (13.4) | 9 (10.2) | <5 | <5 | <5 | 46 (8.9) |

| | Adult age range (years) | | | | | | Overall |
|--|-------------------------|---------|---------|---------|---------|---------|-----------------------|
| | 40-43 | 44-47 | 48-51 | 52-55 | 56-59 | 60+ | Adults (≥16 years) |
| Number in age range | 45 | 17 | 26 | 16 | 22 | 12 | 516 |
| Number who had culture taken* | 25 | 9 | 12 | 8 | 9 | 3 | 276 |
| Chronic S. aureus n (%) | 7 (15.6) | <5 | 0 (0.0) | <5 | <5 | 0 (0.0) | 84 (16.3) |
| Intermittent <i>S. aureus</i> n (%) | <5 | 0 (0.0) | <5 | <5 | <5 | <5 | 61 (11.8) |
| Chronic <i>P. aeruginosa</i> n (%) | 9 (20.0) | <5 | <5 | <5 | <5 | <5 | 78 (15.1) |
| Intermittent <i>P.</i> aeruginosa n (%) | <5 | <5 | <5 | 0 (0.0) | <5 | 0 (0.0) | 41 (7.9) |
| <i>B. cepacia complex</i> n (%) | <5 | <5 | <5 | 0 (0.0) | 0 (0.0) | 0 (0.0) | 23 (4.5) |
| <i>B. cenocepacia</i> n (%)) | 0 (0.0) | <5 | 0 (0.0) | 0 (0.0) | 0 (0.0) | 0 (0.0) | 6 (1.2) |
| B. multivorans n (%) | <5 | <5 | <5 | 0 (0.0) | 0 (0.0) | 0 (0.0) | 11 (2.1) |
| B. other cepacia n (%)) | <5 | 0 (0.0) | 0 (0.0) | 0 (0.0) | 0 (0.0) | 0 (0.0) | 6 (1.2) |
| <i>M</i> RSA n (%) | 0 (0.0) | 0 (0.0) | 0 (0.0) | 0 (0.0) | 0 (0.0) | 0 (0.0) | 4 (0.8) |
| <i>H. influenza</i> n (%) | 5 (11.1) | <5 | 0 (0.0) | 0 (0.0) | <5 | 0 (0.0) | 48 (9.3) |
| <i>NTM</i> n (%) | 0 (0.0) | 0 (0.0) | <5 | 0 (0.0) | <5 | 0 (0.0) | 29 (5.6) |
| Aspergillus n (%) | <5 | <5 | <5 | <5 | <5 | <5 | 46 (8.9) |

1.17 Nontuberculous mycobacteria (NTM) or atypical mycobacteria

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

| | 2018 (n=819) | 2019 (n=868) | 2020 (n=814) |
|---|--------------|--------------|--------------|
| NTM prevalence; n (%) | 46 (5.6) | 53 (6.1) | 41 (5.0) |
| On NTM treatment in the given year; n (% of NTM prevalence in given year) | 16 (34.8) | 29 (54.7) | 19 (46.3) |
| NTM incidence | 19 (2.5) | 23 (2.8) | 12 (1.6) |
| Mycobacterium abscessus prevalence (%) | 28 (3.4) | 32 (3.7) | 27 (3.3) |
| <i>Mycobacterium abscessus incidence;</i> n (%) | 9 (1.1) | 13 (1.6) | 6 (0.8) |



1.18 Lung infections over time

N=661 in 2010, N=795 in 2015, N=814 in 2020



| Chronic Staphylococcus aureus | | | | | | | | |
|-------------------------------|---------|---------|---------|----------|--|--|--|--|
| Age (years) | 2010(%) | 2015(%) | 2020(%) | p-value* | | | | |
| 0-3 | 2.2 | 3.4 | 4.2 | 0.833 | | | | |
| 4-7 | 2.3 | 6.9 | 9.4 | 0.578 | | | | |
| 8-11 | 15.4 | 16.1 | 14.7 | 0.792 | | | | |
| 12-15 | 23.8 | 11.9 | 9.9 | 0.702 | | | | |
| 16-19 | 34.8 | 20.8 | 23.1 | 0.756 | | | | |
| 20-23 | 32.3 | 34.1 | 20.9 | 0.071 | | | | |
| 24-27 | 30.0 | 33.0 | 21.6 | 0.085 | | | | |
| 28-31 | 34.1 | 38.3 | 14.9 | 0.002 | | | | |
| 32-35 | 38.9 | 33.9 | 13.2 | 0.011 | | | | |
| 36-39 | 50.0 | 25.7 | 15.9 | 0.281 | | | | |
| 40-43 | 21.1 | 23.8 | 15.6 | 0.418 | | | | |
| 44-47 | 18.2 | 33.3 | 17.6 | 0.275 | | | | |
| 48+ | 58.8 | 17.8 | 5.3 | 0.026 | | | | |
| <16 | 10.4 | 10.1 | 10.4 | N/A | | | | |
| 16+ | 34.1 | 29.8 | 16.3 | N/A | | | | |
| <18 | 13.9 | 11.6 | 11.2 | N/A | | | | |
| 18+ | 33.3 | 30.0 | 16.0 | N/A | | | | |

| Chronic Pseudomonas aeruginosa | | | | | | | |
|--------------------------------|---------|---------|---------|----------|--|--|--|
| Age (years) | 2010(%) | 2015(%) | 2020(%) | p-value* | | | |
| 0-3 | 1.1 | 1.7 | 0.0 | 0.365 | | | |
| 4-7 | 2.3 | 4.6 | 1.6 | 0.303 | | | |
| 8-11 | 6.2 | 5.4 | 5.3 | 0.972 | | | |
| 12-15 | 14.3 | 6.8 | 6.6 | 0.964 | | | |
| 16-19 | 37.1 | 22.1 | 15.4 | 0.345 | | | |
| 20-23 | 55.4 | 33.0 | 10.4 | 0.001 | | | |
| 24-27 | 40.0 | 52.1 | 12.5 | <0.001 | | | |
| 28-31 | 46.3 | 48.3 | 16.2 | <0.001 | | | |
| 32-35 | 50.0 | 37.5 | 13.2 | 0.004 | | | |
| 36-39 | 35.7 | 40.0 | 22.7 | 0.097 | | | |
| 40-43 | 47.4 | 42.9 | 20.0 | 0.052 | | | |
| 44-47 | 45.5 | 28.6 | 11.8 | 0.206 | | | |
| 48+ | 47.1 | 42.2 | 15.8 | 0.001 | | | |
| <16 | 5.8 | 4.7 | 4.0 | N/A | | | |
| 16+ | 44.3 | 38.8 | 15.1 | N/A | | | |
| <18 | 8.0 | 5.8 | 5.0 | N/A | | | |
| 18+ | 47.6 | 40.3 | 15.0 | N/A | | | |

* The proportion of people with each infection within each age group was compared between 2015 and 2020. If the p-value is less than 0.05 then the difference in the proportions is statistically significant.

Complications

1.19 Complications in 2020

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

| | Overall | <16 years | ≥16 years |
|--|------------|-----------|------------|
| | n (%) | n (%) | n (%) |
| Respiratory related | | | |
| Nasal polyps requiring surgery | 21 (2.6) | 8 (2.7) | 13 (2.5) |
| Sinus disease | 109 (13.4) | 0 | 109 (21.1) |
| Asthma | 55 (6.8) | <5 | - |
| Allergic bronchopulmonary aspergillosis (ABPA) | 37 (4.5) | 7 (2.3) | 30 (5.8) |
| Any haemoptysis | 14 (1.7) | 0 | 14 (2.7) |
| Massive haemoptysis | <5 | 0 | <5 |
| Pneumothorax requiring chest tube | 0 (0.0) | 0 | 0 |
| Pancreas and hepatobiliary related | | | |
| Raised liver enzymes | 47 (5.8) | 11 (3.7) | 36 (7.0) |
| Liver disease | 150 (18.4) | 35 (11.7) | 115 (22.3) |
| Cirrhosis with no portal hypertension | 5 (0.6) | 0 | 5 (1.0) |
| Cirrhosis with portal hypertension | 19 (2.3) | 5 (1.7) | 14 (2.7) |
| Gall bladder disease requiring surgery | 7 (0.9) | <5 | - |
| Pancreatitis | 8 (1.0) | <5 | - |
| Upper gastrointestinal (GI) | | | |
| Gastroesophageal reflux disease (GERD) | 231 (28.4) | <5 | - |
| Peptic ulcer | 0 (0.0) | 0 | 0 |
| GI bleed (varices as source) | <5 | 0 | <5 |
| GI bleed (non varices as source) | 0 (0.0) | 0 | 0 |
| Lower GI | | | |
| Intestinal obstruction | <5 | <5 | 0 |
| Distal intestinal obstruction syndrome (DIOS) | 104 (12.8) | 5 (1.7) | 99 (19.2) |
| Fibrosing colonopathy / colonic stricture | 0 (0.0) | 0 | 0 |
| Rectal prolapse | <5 | <5 | 0 |
| Renal | | | |
| Kidney stones | 5 (0.6) | <5 | <5 |
| Renal failure | 11 (1.4) | 0 | 11 (2.1) |
| Musculoskeletal | | | |
| Arthritis | <5 | 0 | <5 |
| Arthropathy | 35 (4.3) | <5 | 33 (6.4) |
| Bone fracture | <5 | <5 | 0 |
| Osteopenia | 111 (13.6) | 0 | 111 (21.5) |
| Osteoporosis | 48 (5.9) | 0 | 48 (9.3) |
| Other | | | |
| Cancer confirmed by histology | <5 | <5 | <5 |
| Port inserted or replaced | 12 (1.5) | <5 | - |
| Depression | 24 (2.9) | <5 | - |
| Hearing loss | 12 (1.5) | <5 | - |
| Hypertension | 13 (1.6) | 0 | 13 (2.5) |

-Redacted to adhere to statistical disclosure guidelines.

1.20 Incidence of complications

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

| | 2019 | | | 2020 | | |
|---|--------------------|----------------------|----------------------|--------------------|----------------------|----------------------|
| | Overall (n=868) | <16 years (n=336) | ≥16 years (n=532) | Overall (n=814) | <16 years (n=298) | ≥16 years (n=516) |
| Allergic bronchopulmonary aspergillosis; n (%) | 20 (2.3) | 6 (1.8) | 14 (2.6) | 12 (1.5) | <5 | - |
| Cirrhosis - no portal hypertension; n (%) | 0 (0) | 0 (0) | 0 (0) | <5 | 0 (0) | <5 |
| Cirrhosis - with portal hypertension; n (%) | 7 (.8) | <5 | <5 | 5 (0.6) | <5 | <5 |
| Cancer confirmed by histology; n (%) | 0 (0) | 0 (0) | 0 (0) | <5 | <5 | <5 |

1.21 Cystic fibrosis-related diabetes (CFRD)

CFRD is common in adults and adolescents with cystic fibrosis. This is because, for many people with CF, the pancreas does not work properly. This can mean that not enough insulin is produced, or it may not work properly, causing CFRD. CFRD is different from type 1 and type 2 diabetes, but has features of both.

| | All ≥10 years (n=651) | 10-15 years (n=135) | ≥16 years (n=516) |
|-------------------------------------|--------------------------|------------------------|----------------------|
| On CFRD treatment; n (%) | 96 (14.7) | <5 | - |
| Of those on treatment | | | |
| Insulin; n (%) | 95 (99.0) | <5 | - |
| CFRD Screening; n(%) | | | |
| Yes | 190 (29.2) | 80 (59.3) | 110 (21.3) |
| Screening type ; n (%) | | | |
| Continous glucose monitoring; n (%) | 35 (18.4) | 14 (17.5) | 21 (19.1) |
| Oral glucose tolerance test; n (%) | 136 (71.6) | 55 (68.8) | 81 (73.6) |
| Not screened(known CFRD) | 141 (21.7) | <5 | - |
| Not screened (other) | 169 (26.0) | 49 (36.3) | 120 (23.3) |
| Unknown | 8 (1.2) | <5 | - |



Antibiotics

1.22 Intravenous (IV) antibiotics N=814

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

| | | Home | | Hospital | | Total | | |
|-------------|-----|-------------------|----------------------|-------------------|----------------------|-------------------|----------------------|--|
| Age (years) | N | Patients n (%) | Median days (IQR) | Patients n (%) | Median days (IQR) | Patients n (%) | Median days (IQR) | |
| 0-3 | 48 | 0 (0.0) | N/A | 9 (18.8) | 14 () | 9 (18.8) | 14 (14-27) | |
| 4-7 | 64 | <5 | 14 (14-14) | 15 (23.4) | 14 (14-14) | 16 (25.0) | 14 (9-35) | |
| 8-11 | 95 | 5 (5.3) | 42 (31-42) | 21 (22.1) | 14 (31-42) | 23 (24.2) | 15 (14-51) | |
| 12-15 | 91 | 10 (11.0) | 21 (11-41) | 30 (33.0) | 25 (11-41) | 33 (36.3) | 28 (18-61) | |
| 16-19 | 52 | 9 (17.3) | 14 (6-49) | 9 (17.3) | 18 (6-49) | 12 (23.1) | 15 (11-93) | |
| 20-23 | 67 | 17 (25.4) | 14 (14-27) | 16 (23.9) | 10 (14-27) | 22 (32.8) | 14 (14-40) | |
| 24-27 | 88 | 26 (29.5) | 14 (11-36) | 32 (36.4) | 11 (11-36) | 37 (42.0) | 21 (14-42) | |
| 28-31 | 74 | 21 (28.4) | 19 (12-24) | 20 (27.0) | 9 (12-24) | 26 (35.1) | 21 (14-35) | |
| 32-35 | 53 | 18 (34.0) | 14 (13-26) | 14 (26.4) | 15 (13-26) | 20 (37.7) | 26 (14-40) | |
| 36-39 | 44 | 14 (31.8) | 19 (14-33) | 11 (25.0) | 6 (14-33) | 17 (38.6) | 25 (14-42) | |
| 40-43 | 45 | 17 (37.8) | 13 (10-46) | 17 (37.8) | 7 (10-46) | 19 (42.2) | 14 (14-56) | |
| 44-47 | 17 | <5 | - | <5 | - | <5 | - | |
| 48-51 | 26 | <5 | - | <5 | - | 5 (19.2) | 23 (14-28) | |
| 52-55 | 16 | 5 (31.3) | 28 (14-28) | <5 | - | 5 (31.3) | 28 (21-28) | |
| 56-59 | 22 | 6 (27.3) | 17 (11-28) | 6 (27.3) | 8 (11-28) | 7 (31.8) | 14 (14-28) | |
| 60+ | 12 | <5 | - | 5 (41.7) | 14 (10-28) | 5 (41.7) | 14 (14-42) | |
| <16 | 298 | 17 (5.7) | 27 (14-42) | 75 (25.2) | 15 (14-42) | 81 (27.2) | 22 (14-42) | |
| ≥16 | 516 | 142 (27.5) | 15 (11-28) | 139 (26.9) | 10 (11-28) | 179 (34.7) | 21 (14-41) | |
| <18 | 321 | 20 (6.2) | 28 (13-42) | 79 (24.6) | 15 (13-42) | 86 (26.8) | 21 (14-51) | |
| ≥18 | 493 | 139 (28.2) | 14 (11-28) | 135 (27.4) | 10 (11-28) | 174 (35.3) | 21 (14-40) | |
| Overall | 814 | 159 (19.5) | 15 (11-30) | 214 (26.3) | 14 (11-30) | 260 (31.9) | 21 (14-42) | |

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



The bar graph below summarises the proportion of people receiving at least one course of IV antibiotics across different age groups within the Scottish CF population. Overall, the proportion of patients receiving at least one IV course at home was 19.5% and in hospital was 26.3%. The proportion receiving any IVs was 31.9%.



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

| | 2010 2 | | 2015 | | | 2020 | | | |
|--|--------------|--------------|--------------|----------------|---------------|----------------|--------------|--------------|--------------|
| | Overall | <16 years | ≥16 years | Overall | <16 years | ≥16 years | Overall | <16 years | ≥16 years |
| Patients with chronic <i>P. aeruginosa</i> ; n | 136 | 18 | 118 | 215 | 14 | 201 | 90 | 12 | 78 |
| Tobramycin solution; n (%) | 18 (13.2) | <5 | - | 35 (16.3) | 3 (21.4) | 32 (15.9) | 19 (21.1) | <5 | - |
| Other aminoglycoside; n (%) | <5 | 0 | <5 | <5 | 0 | <5 | <5 | 0 | <5 |
| Colistin; n (%) | 58 (42.6) | 10 (55.6) | 48 (40.7) | 68 (31.6) | 8 (57.1) | 60 (29.9) | 25 (27.8) | 5 (41.7) | 20 (25.6) |
| Promixin; n (%) | 23 (16.9) | 0 | 23 (19.5) | 39 (18.1) | <5 | - | 12 (13.3) | <5 | - |
| Aztreonam; n (%) | - | - | - | 215 (100.0) | 14 (100.0) | 201 (100.0) | 10 (11.1) | 0 | 10 (12.8) |
| Colistimethate (inh) in- halation powder; n (%) | - | - | - | 35 (16.3) | <5 | - | 20 (22.2) | <5 | - |
| Tobramycin Inhalation Powder; n (%) | - | - | - | 59 (27.4) | <5 | - | 17 (18.9) | 0 | 17 (21.8) |
| Levofloxacin; n(%) | - | - | - | - | - | - | <5 | 0 | <5 |
| At least one of the above; n (%) | 94 (69.1) | 11 (61.1) | 83 (70.3) | 167 (77.7) | 11 (78.6) | 156 (77.6) | 77 (85.6) | 10 (83.3) | 67 (85.9) |

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

1.24 Long-term azithromycin use

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

| | Age | Number of patients on azithromycin; n | Patients with chronic <i>P. aeruginosa</i> ; n (%) | Patients without chronic <i>P. aeruginosa</i> ; n (%) |
|------|------------|---------------------------------------|---|--|
| | Overall | 293 | 133 (45.4) | 160 (54.6) |
| 2010 | 0-3 years | <5 | 0 (0.0) | <5 |
| 2010 | 4-15 years | 55 | 9 (16.4) | 46 (83.6) |
| | ≥ 16 years | 237 | 124 (52.3) | 113 (47.7) |
| | Overall | 420 | 176 (41.9) | 244 (58.1) |
| 2015 | 0-3 years | <5 | 0 (0.0) | <5 |
| 2015 | 4-15 years | 66 | 9 (13.6) | 57 (86.4) |
| | ≥ 16 years | 353 | 167 (47.3) | 186 (52.7) |
| | Overall | 461 | 73 (15.8) | 388 (84.2) |
| 2020 | 0-3 years | 6 | 0 (0.0) | 6 (100.0) |
| 2020 | 4-15 years | 92 | 6 (6.5) | 86 (93.5) |
| | ≥ 16 years | 363 | 67 (18.5) | 296 (81.5) |

1.25 Flucloxacillin

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

| Age; years | Patients; n | Patients on prophylactic flucloxacillin; n (%) |
|------------|-------------|--|
| 0-3 | 48 | 34 (70.8) |
| 4-7 | 64 | 27 (42.2) |
| 8-11 | 95 | 39 (41.1) |
| 12-15 | 91 | 26 (28.6) |
| 16-19 | 52 | 14 (26.9) |
| 20-23 | 67 | 24 (35.8) |
| 24-27 | 88 | 13 (14.8) |
| 28-31 | 74 | 11 (14.9) |
| 32-35 | 53 | <5 |
| 36-39 | 44 | <5 |
| 40-43 | 45 | <5 |
| 44-47 | 17 | 0 |
| 48-51 | 26 | <5 |
| 52-55 | 16 | 0 |
| 56-59 | 22 | <5 |
| 60+ | 12 | 0 |
| <16 years | 298 | 126 (42.3) |
| ≥16 years | 516 | 72 (14.0) |
| <18 years | 321 | 130 (40.5) |
| ≥18 years | 493 | 68 (13.8) |
| Overall | 814 | 198 (24.3) |

Muco-active therapies

1.26 Mannitol

| Age; years | Patients; n | Patients on Mannitol; n (%) |
|------------|-------------|-----------------------------|
| 0-3 | 48 | 0 |
| 4-7 | 64 | 0 |
| 8-11 | 95 | 0 |
| 12-15 | 91 | 0 |
| 16-19 | 52 | 0 |
| 20-23 | 67 | 0 |
| 24-27 | 88 | <5 |
| 28-31 | 74 | <5 |
| 32-35 | 53 | <5 |
| 36-39 | 44 | 0 |
| 40-43 | 45 | <5 |
| 44-47 | 17 | 0 |
| 48-51 | 26 | <5 |
| 52-55 | 16 | 0 |
| 56-59 | 22 | 0 |
| 60+ | 12 | 0 |
| <16 years | 298 | 0 |
| ≥16 years | 516 | 8 (1.6) |
| <18 years | 321 | 0 |
| ≥18 years | 493 | 8 (1.6) |
| Overall | 814 | 8 (1.0) |

1.27 DNase

| | 2010 | | 2015 | | 2020 | | |
|------------|-------------|-----------------------------|-------------|-----------------------------|-------------|-----------------------------|--|
| Age; years | Patients; n | Patients on DNase; n (%) | Patients; n | Patients on DNase; n (%) | Patients; n | Patients on DNase; n (%) | |
| 0-3 | 90 | 4 (4.4) | 59 | 5 (8.5) | 48 | 7 (14.6) | |
| 4-7 | 88 | 12 (13.6) | 87 | 18 (20.7) | 64 | 11 (17.2) | |
| 8-11 | 65 | 26 (40.0) | 93 | 43 (46.2) | 95 | 42 (44.2) | |
| 12-15 | 84 | 37 (44.0) | 59 | 30 (50.8) | 91 | 56 (61.5) | |
| 16-19 | 89 | 36 (40.4) | 77 | 40 (51.9) | 52 | 31 (59.6) | |
| 20-23 | 65 | 26 (40.0) | 88 | 36 (40.9) | 67 | 42 (62.7) | |
| 24-27 | 60 | 16 (26.7) | 94 | 45 (47.9) | 88 | 56 (63.6) | |
| 28-31 | 41 | 14 (34.1) | 60 | 19 (31.7) | 74 | 50 (67.6) | |
| 32-35 | 18 | 3 (16.7) | 56 | 14 (25.0) | 53 | 26 (49.1) | |
| 36-39 | 14 | 1 (7.1) | 35 | 10 (28.6) | 44 | 20 (45.5) | |
| 40-43 | 19 | 8 (42.1) | 21 | <5 | 45 | 16 (35.6) | |
| 44-47 | 11 | <5 | 21 | 8 (38.1) | 17 | 10 (58.8) | |
| 48-51 | 7 | <5 | 16 | 6 (37.5) | 26 | 10 (38.5) | |
| 52-55 | 5 | 0 | 14 | 5 (35.7) | 16 | 7 (43.8) | |
| 56-59 | <5 | <5 | 7 | <5 | 22 | 9 (40.9) | |
| 60+ | <5 | 0 | 8 | <5 | 12 | 5 (41.7) | |
| <16 years | 327 | 79 (24.2) | 298 | 96 (32.2) | 298 | 116 (38.9) | |
| ≥16 years | 334 | 111 (33.2) | 497 | 190 (38.2) | 516 | 282 (54.7) | |
| <18 years | 373 | 102 (27.3) | 328 | 114 (34.8) | 321 | 130 (40.5) | |
| ≥18 years | 288 | 88 (30.6) | 467 | 172 (36.8) | 493 | 268 (54.4) | |
| Overall | 661 | 190 (28.7) | 795 | 286 (36.0) | 814 | 398 (48.9) | |

1.28 Hypertonic saline

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

| | 2010 | | 2015 | | 2020 | | |
|------------|-------------|--|-------------|--|-------------|--|--|
| Age; years | Patients; n | Patients on hypertonic saline; n (%) | Patients; n | Patients on hypertonic saline; n (%) | Patients; n | Patients on hypertonic saline; n (%) | |
| 0-3 | 90 | 0 | 59 | <5 | 48 | 8 (16.7) | |
| 4-7 | 88 | 0 | 87 | 11 (12.6) | 64 | 15 (23.4) | |
| 8-11 | 65 | <5 | 93 | 18 (19.4) | 95 | 32 (33.7) | |
| 12-15 | 84 | 9 (10.7) | 59 | 19 (32.2) | 91 | 41 (45.1) | |
| 16-19 | 89 | 8 (9.0) | 77 | 19 (24.7) | 52 | 18 (34.6) | |
| 20-23 | 65 | 5 (7.7) | 88 | 20 (22.7) | 67 | 18 (26.9) | |
| 24-27 | 60 | 8 (13.3) | 94 | 15 (16.0) | 88 | 24 (27.3) | |
| 28-31 | 41 | <5 | 60 | 12 (20.0) | 74 | 12 (16.2) | |
| 32-35 | 18 | 0 | 56 | 6 (10.7) | 53 | 8 (15.1) | |
| 36-39 | 14 | <5 | 35 | <5 | 44 | 7 (15.9) | |
| 40-43 | 19 | <5 | 21 | 0 | 45 | <5 | |
| 44-47 | 11 | 0 | 21 | <5 | 17 | 0 | |
| 48-51 | 7 | <5 | 16 | <5 | 26 | <5 | |
| 52-55 | 5 | 0 | 14 | <5 | 16 | 0 | |
| 56-59 | <5 | 0 | 7 | 0 | 22 | <5 | |
| 60+ | <5 | 0 | 8 | <5 | 12 | <5 | |
| <16 years | 327 | 13 (4.0) | 298 | 51 (17.1) | 298 | 96 (32.2) | |
| ≥16 years | 334 | 27 (8.1) | 497 | 85 (17.1) | 516 | 95 (18.4) | |
| <18 years | 373 | 17 (4.6) | 328 | 58 (17.7) | 321 | 105 (32.7) | |
| ≥18 years | 288 | 23 (8.0) | 467 | 78 (16.7) | 493 | 86 (17.4) | |
| Overall | 661 | 40 (6.1) | 795 | 136 (17.1) | 814 | 191 (23.5) | |

1.29 Burden of treatment

The Venn diagram shows how many people with CF are on one or more inhaled therapies and the combinations they take. A total of 253 (31.6%) people in Scotland are on no inhaled therapies.



Other therapies 1.30 CFTR modulators

Ivacaftor

As of November 2020, ivacaftor was approved for use on the NHS across the UK for people aged four months and older with at least one copy of a CFTR 'gating' mutation, and for people aged 6 months and over with the R117H. Additionally patients in Scotland and NI can access from age 4 months with specific rare mutations.

Lumacaftor/ivacaftor

Lumacaftor/ivacaftor is licensed for use in patients aged 2 and over with two copies of the F508del mutation. Managed access was agreed to make lumacaftor/ivacaftor available on the NHS from Autumn/Winter 2019.

Tezacaftor/ivacaftor

Tezacaftor/ivacaftor is licenced for use in patients aged 6 and over who have two copies of the F508del mutation, or a single copy of F508del and one of 14 residual function mutations. Managed access was agreed to make tezacaftor/ivacaftor available on the NHS from Autumn/Winter 2019. Patients aged 6+ with rare mutations gained access in Scotland and NI from December 2020.

Elexacaftor/tezacaftor/ivacaftor

As an extension of the managed access agreement, from August 2020 Elexacaftor/tezacaftor/ ivacaftor was made available in the UK for those ages 12 and over who have two copies of the F508del mutation, or a single copy of F508del and one minimal function mutation.

CFTR modulator use in 2020

A total of 485 people in Scotland received one CFTR modulator treatment in 2020. The graph below shows the number of people on each drug by month. Where people switched modulators, the most recent prescription is counted.

1.31 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 Ventillation (NIV); n (%) | 5 (0.6) | <5 | <5 | <5 | <5 | | | | | |
| Long term oxygen; n (%) | 23 (2.8) | <5 | - | 5 (1.6) | 18 (3.7) | | | | | |
| Anong those who have long-term oxygen: | | | | | | | | | | |
| Continuously | 5 (21.7) | 0 (0.0) | 5 (26.3) | 0 (0.0) | 5 (27.8) | | | | | |
| Nocturnal or with exertion | 10 (43.5) | 0 (0.0) | 10 (52.6) | 0 (0.0) | 10 (55.6) | | | | | |
| As required (PRN) | <5 | 0 (0.0) | <5 | <5 | <5 | | | | | |
| With exacerbation | 5 (21.7) | <5 | <5 | <5 | <5 | | | | | |

1.32 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 (%) | 93 (11.4) | 6 (2.0) | 87 (16.9) | 7 (2.2) | 86 (17.4) |
| Autogenic drainage (including assisted autogenic drainage); n (%) | 352 (43.2) | 53 (17.8) | 299 (57.9) | 60 (18.7) | 292 (59.2) |
| Postural drainage; n (%) | <5 | 0 (0.0) | <5 | 0 (0.0) | <5 |
| Any form of PEP; n (%) | 459 (56.4) | 276 (92.6) | 183 (35.5) | 295 (91.9) | 164 (33.3) |
| VEST; n (%) | <5 | 0 (0.0) | <5 | 0 (0.0) | <5 |
| Exercise; n (%) | 431 (52.9) | 161 (54.0) | 270 (52.3) | 176 (54.8) | 255 (51.7) |
| Other; n (%) | 170 (20.9) | 126 (42.3) | 44 (8.5) | 128 (39.9) | 42 (8.5) |

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=814) | <16 years (n=298) | ≥16 years (n=516) | <18 years (n=321) | ≥18 years (n=493) |
|---|--------------------|----------------------|----------------------|----------------------|----------------------|
| Any supplemental feeding; n (%) | 273 (33.5) | 49 (16.4) | 224 (43.4) | 56 (17.4) | 217 (44.0) |
| Nasogastric tube; n (%) | 6 (0.7) | <5 | <5 | <5 | <5 |
| Gastrostomy tube/Button; n (%) | 29 (3.6) | 12 (4.0) | 17 (3.3) | 13 (4.0) | 16 (3.2) |
| Jejunal; n (%) | 0 (0.0) | 0 (0.0) | 0 (0.0) | 0 (0.0) | 0 (0.0) |
| Total Parenteral Nutrition (TPN); n (%) | <5 | <5 | 0 (0.0) | <5 | 0 (0.0) |

1.34 Transplants

Lung transplantation has been available to people with CF for almost 30 years. Today, the most common operation carried out is a double-lung transplant, or 'bilateral sequential lung transplant'. The following table gives information about transplant activity over time.

| | 2015 | 2016 | 2017 | 2018 | 2019 | 2020 |
|-----------------------------------|------|------|------|------|------|------|
| Patients evaluated; n | 19 | 18 | 22 | 19 | 21 | 11 |
| Patients accepted; n | 16 | 8 | 17 | 7 | 10 | 2 |
| Patients receiving transplants; n | <5 | <5 | <5 | <5 | 5 | 0 |
| Bilateral lung | <5 | <5 | <5 | <5 | <5 | 0 |
| Liver | 0 | 0 | 0 | <5 | <5 | 0 |
| Other | 0 | <5 | <5 | <5 | 0 | 0 |

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

Genotypes

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

| Data completeness | n (%) |
|--|------------|
| Patients genotyped with at least one mutation recorded | 812 (99.8) |
| Patients genotyped with both mutations recorded | 811 (99.6) |
| F508del mutations | |
| Homozygous F508del | 357 (43.9) |
| Heterozygous F508del | 380 (46.7) |

1.35 Mutation combinations in Scotland

This tabulation shows the proportion (%) of patients with the most common mutation combinations. For example, 8.5% 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 | 43.9 | | | | | | | 43.9 | |
| R117H | 6.4 | 0.1 | | | | | | 6.5 | |
| G551D | 8.5 | 0.1 | 0.2 | | | | | 8.8 | |
| G542X | 5.5 | 0.2 | 0.1 | 0.1 | | | | 6.0 | |
| 621+1G->T | 0.5 | 0.0 | 0.1 | 0.0 | 0.0 | | | 0.6 | |
| Other | 25.7 | 1.2 | 1.2 | 0.7 | 0.2 | 4.7 | | 33.8 | |
| Unknown | 0.1 | 0.0 | 0.0 | 0.0 | 0.0 | 0.0 | 0.2 | 0.4 | |
| Total | 90.5 | 1.7 | 1.7 | 0.9 | 0.2 | 4.7 | 0.2 | 100.0 | |

1.36 Mutations in the Scottish population

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

These are the 20 most common mutations in the Scottish population. The full list of recorded mutations can be found in Appendix 2.

| Nucleotide | Protein | Legacy name | N | % |
|---------------------|-------------------|---------------|-----|------|
| c.1521_1523delCTT | p.Phe508del | F508del | 737 | 90.5 |
| c.1652G->A | p.Gly551Asp | G551D | 84 | 10.3 |
| c.350G->A | p.Arg117His | R117H | 66 | 8.1 |
| c.1624G->T | p.Gly542X | G542X | 55 | 6.8 |
| c.200C->T | p.Pro67Leu | P67L | 41 | 5.0 |
| c.1679G->C | p.Arg560Thr | R560T | 18 | 2.2 |
| c.1585-1G->A | | 1717-1G->A | 13 | 1.6 |
| c.1477C->T | p.Gln493X | Q493X | 12 | 1.5 |
| c.3909C->G | p.Asn1303Lys | N1303K | 11 | 1.4 |
| c.3454G->C | p.Asp1152His | D1152H | 10 | 1.2 |
| c.2657+5G->A | | 2789+5G->A | 10 | 1.2 |
| c.1558G->T | p.Val520Phe | V520F | 8 | 1.0 |
| c.3717+12191C->T | | 3849+10kbC->T | 7 | 0.9 |
| c.489+1G->T | | 621+1G->T | 7 | 0.9 |
| c.178G->T | p.Glu60X | E60X | 6 | 0.7 |
| c.948delT | p.Phe316LeufsX12 | 1078delT | 6 | 0.7 |
| c.2657+2_2657+3insA | | 2789+2insA | 5 | 0.6 |
| c.1364C->A | p.Ala455Glu | A455E | 5 | 0.6 |
| c.3528delC | p.Lys1177SerfsX15 | 3659delC | 5 | 0.6 |
| c.1721C->A | p.Pro574His | P574H | <5 | - |

Section 2 Centre-level analysis

Cystic fibrosis care in Scotland is led by eight regional centres, two stand-alone clinics and three networked clinics. The breakdown of centres and clinics delivering paediatric and adult care is shown below:

| | Paediatric | Adult | Total |
|---------------------|------------|-------|-------|
| Centres | 5 | 3 | 8 |
| Stand-alone clinics | 2 | 0 | 2 |

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

Lots of different factors can affect the outcomes of people with CF in centres, not all of which are within a centre's control. This might include the economic profile of the area, the age at which the person with CF was diagnosed and referred to the centre and certain patient characteristics such as their gender, as well as facilities, care pathways, and the medical team providing care.

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

Full tables of the data are shown in appendix 1.

Key

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 Oservices in the UK Oservices in Scotland

The median FEV_1 % predicted of patients attending paediatric centres/clinics in Scotland is 92.4% predicted (IQR: 82.1 – 99.7).

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

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

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

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

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

Section 2b: Adult centre analysis

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

Key Services in the UK Services in Scotland

2.7 Age distribution by adult centre/clinic

The median age of patients attending adult services in Scotland is 30 years (IQR: 24-40).

2.8 FEV₁ % predicted (GLI equations) by adult centre/clinic (without a history of lung transplant)

The median FEV₁ % predicted of patients attending adult services in Scotland is 65.3% (IQR 44.4-82.9).

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 23.2 (IQR: 21.0-26.1).

The proportion of patients with chronic Paeruginosa attending adult services in Scotland is 15%.

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 85.5%.

2.12 Data completeness by adult centre/clinic

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

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

The proportion of patients attending adult services in Scotland receiving DNase treatment is 54.5%.

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

The proportion of patients attending adult services in Scotland receiving hypertonic saline or mannitol treatment is 18.8%.

Glossary

| Word/Phrase | Meaning |
|--|--|
| 2020 | 1 January 2020 – 31 December 2020 |
| ABPA (allergic bronchopulmonary aspergillosis) | When a person develops a respiratory allergic reaction to Aspergillus fumigatus. |
| Arthritis | A condition causing pain and inflammation in the joints. |
| Arthropathy | A condition causing pain in the joints. |
| Asthma | A respiratory condition causing reversible episodes of difficulty breathing, often associated with wheezing. |
| BMI (body mass index) | A measure designed to show whether a person is a healthy weight for their height. |
| <i>Burkholderia cepacia</i> complex | <i>B. cepacia</i> complex are a group of bacteria, some of which threaten the health of people with cystic fibrosis. |
| CF | Cystic fibrosis. |
| CFTR (cystic fibrosis transmembrane conductance regulator) | A protein at the cell surface that controls the salt and water balance across a cell. The gene that causes cystic fibrosis is the blueprint for the CFTR protein. Everyone has two copies of the gene for CFTR. To be born with cystic fibrosis, both CFTR genes must be affected by a CF-causing mutation. |
| Chronic | Persistent, or long-lasting. |
| Cirrhosis | A chronic liver disease. |
| CI (confidence interval) | A way of expressing how certain we are about our statistical estimates of a clinical measure (eg BMI). It gives a range of results that is likely to include the 'true' value for the population. A narrow confidence interval indicates a more precise estimate. A wide confidence interval indicates more uncertainty about the true value of the clinical measure - often because a small group of patients has been studied. The confidence interval is usually stated as '95% Cl', which means that the range of values has a 95 in 100 chance of including the 'true' value. |
| Enzymes | Biological molecules that help complex reactions, such as digestion of food, occur in the body. |
| FEV ₁ (forced expiratory volume in one second) | This is the amount of air that a person can blow out of the lungs in the first second of a forced exhaled breath. People with healthy lungs can blow out most of the air held in this time. |
| FEV ₁ % predicted | The FEV ₁ can be converted from absolute litres of air blown out into a predicted percentage (%). A healthy range for % predicted is calculated from a very large population sample, and is normally considered to be between 80-120% predicted. |
| Fibrosing colonopathy | A condition causing narrowing of part of the colon. |
| Gall bladder | The small sac-shaped organ under the liver that stores bile after it is secreted by the liver, before it is released into the intestine. |
| GI (gastrointestinal) | The GI tract is the organ system responsible for digesting food, absorbing nutrients and expelling waste. |
| Genotype | Part of the genetic makeup of a cell, organism or individual that usually controls a particular characteristic (known as a phenotype). |
| GERD (gastroesophageal reflux disease) | A chronic symptom of damage caused by stomach acid coming up from the stomach into the oesophagus. |
| GI bleed | Bleeding in the gastrointestinal tract. |
| GLI equations | Global Lung Initiative, the equation used for calculating FEV_1 % predicted from absolute FEV_1 that takes into account age, gender, height and ethnicity. |
| Haemophilus influenza | <i>H. influenza</i> 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 2020 the median age of the 97 people who died (in UK) was 36. |
| MRSA | Methicillin-resistant <i>Staphylococcus aureus</i> is a type of bacteria that is resistant to a number of widely used antibiotics. |
| Mutation | A mutation is a change in a gene. When both of a child's parents are carriers of a CF-causing mutation there is a 25% chance that the child will have cystic fibrosis. There are over 1400 different mutations of the CFTR gene that can cause cystic fibrosis. |
| Nasal polyps | Small, sac-like growths of inflamed mucus caused by chronic inflammation of the nasal lining. |
| NBS (newborn screening) | Newborn screening is part of the heel prick blood spot testing done at 5-7 days of age. The blood sample is tested for a number of conditions, including cystic fibrosis. |
| NTM (nontuberculous mycobacteria) | A mycobacterium that does not cause tuberculosis, but which can cause respiratory infection. There are several known types. |
| Osteopenia | A medical condition less severe than osteoporosis, where the mineral content of bone is reduced. |
| Osteoporosis | A condition where the bones become brittle from loss of tissue. |
| Pancreas | An organ in the digestive system that produces insulin and digestive enzymes. |
| Pancreatitis | Inflammation of the pancreas. |
| Peptic ulcer | An open sore that develops in the lining of the stomach, also known as a stomach ulcer. |
| Percentile | A percentile shows where a value stands, relative to the rest of the data. If a value is higher than 90% of the rest of the data, it is at the 90th percentile. |
| Pneumothorax | A collection of air in the cavity between the lungs and the chest wall causing collapse of the lung on the affected side. |
| Portal hypertension | High blood pressure in the portal vein system, which is the blood system of the liver. |
| Prenatal | Before birth, while the baby is still in the womb. |
| Prevalence | The overall number of people with the condition in the last 12 months. |
| Pseudomonas aeruginosa | A tough bacterial strain. Rarely affecting healthy people, it can cause a wide range of infections, particularly in those with a weakened immune system. |
| Rectal prolapse | When the rectal wall slides through the anus. |
| Renal | Relating to the kidneys. |
| Staphylococcus aureus | S. aureus is a bacterium that can cause disease if it enters the body. |
| Sinus disease | When the sinuses, which are usually filled with air, are full of thick sticky mucus. |
| Statistically significant | This phrase means that after careful calculations there is a definite difference between two groups, which is not simply a result of chance. |

Paediatric centres/clinics providing data in 2020 – ordered alphabetically by location

| | | | | | | ears) | | | |
|-------------|------------------------------------|-----------|-----------------|---------------------------|------|--------|--|--|--|
| Location | Name | Clinic ID | Total Active | Number with annual review | Mean | Median | | | |
| Scotland | Scotland | | | | | | | | |
| Aberdeen | Royal Aberdeen Children's Hospital | 75 | 29 | 24 | 7.1 | 7.0 | | | |
| Ayr | University Hospital Crosshouse | 170 | 25 | 23 | 9.2 | 8.8 | | | |
| Dundee | Ninewells Hospital | 73 | 20 | 8 | 9.2 | 10.0 | | | |
| Edinburgh | Royal Hospital for Sick Children | 143 | 142 | 117 | 9.8 | 9.9 | | | |
| Glasgow | Royal Hospital for Sick Children | 56 | 118 | 104 | 9.2 | 9.8 | | | |
| Inverness | Raigmore Hospital | 31 | 16 | 13 | 10.0 | 11.0 | | | |
| Lanarkshire | Wishaw General Hospital | 162 | 46 | 40 | 9.2 | 9.1 | | | |

Adult centres/clinics providing data in 2020 – ordered alphabetically by location

| | Α | | | Age (ye | Age (years) | |
|-----------|----------------------------|-----------|-----------------|---------------------------|-------------|--------|
| Location | Name | Clinic ID | Total Active | Number with annual review | Mean | Median |
| Scotland | | | | | | |
| Aberdeen | Aberdeen Royal Infirmary | 70 | 74 | 72 | 34.7 | 33.6 |
| Edinburgh | Western General Hospital | 44 | 239 | 215 | 33.4 | 30.8 |
| Glasgow | Gartnavel General Hospital | 79 | 226 | 218 | 34.0 | 30.4 |

| FEV ₁ % predicted at annual review | | | Best FEV ₁ % predicted | | | | |
|---|----------------------|----------------------|-----------------------------------|--------|----------------------|----------------------|--------|
| Number | Mean - unadjusted | Mean – adjusted † | Median | Number | Mean - unadjusted | Mean - adjusted † | Median |
| | | | | | | | |
| 13 | 94.2 | 92.9 | 90.0 | 14 | 102.2 | 100.8 | 101.2 |
| 9 | 89.0 | 88.1 | 86.9 | 11 | 99.7 | 98.5 | 95.7 |
| <5 | 79.2 | 81.4 | 79.2 | <5 | 88.0 | 88.3 | 88.2 |
| 58 | 91.8 | 92.1 | 93.4 | 79 | 92.8 | 93.2 | 94.2 |
| 61 | 92.1 | 92.5 | 94.8 | 68 | 94.6 | 95.1 | 95.8 |
| 8 | 85.3 | 84.8 | 85.3 | 9 | 87.2 | 87.0 | 86.6 |
| 11 | 89.4 | 87.3 | 91.1 | 30 | 95.0 | 94.7 | 95.4 |

| FEV ₁ % predicted at annual review | | | Best FEV,% predicted | | | | |
|---|----------------------|----------------------|----------------------|--------|----------------------|---------------------------------|--------|
| Number | Mean - unadjusted | Mean – adjusted † | Median | Number | Mean - unadjusted | Mean - adjusted [†] | Median |
| | | | | | | | |
| 65 | 65.4 | 66.1 | 65.7 | 65 | 68.0 | 68.5 | 65.7 |
| 157 | 64.0 | 64.2 | 63.8 | 196 | 69.8 | 70.0 | 69.8 |
| 123 | 65.5 | 66.0 | 64.9 | 196 | 66.3 | 66.9 | 66.0 |

 ^{*} where 'best' values were missing, or lower than FEV, % predicted taken at annual review, the annual review value was used.
 † Adjusted for age - this means that the data have been fine-tuned to take account of the different spread of ages across centres and clinics. The adjusted values are intended to show what the average lung function or BMI percentile would be for that centre/ clinic if the age spread is the same as the spread of age in the whole population.

Paediatric centres/clinics providing data in 2020 – ordered alphabetically by location

| | BMI percentile | | | | | | |
|-------------|---------------------------------------|-----------|--------|----------------------|---------------------------------|--------|--|
| Location | Name | Clinic ID | Number | Mean - unadjusted | Mean - adjusted [†] | Median | |
| Scotland | | | | | | | |
| Aberdeen | Royal Aberdeen Children's Hospital | 75 | 22 | 55.6 | 54.2 | 60.2 | |
| Ayr | University Hospital Crosshouse | 170 | 15 | 59.0 | 58.4 | 70.3 | |
| Dundee | Ninewells Hospital | 73 | 6 | 51.8 | 51.6 | 59.0 | |
| Edinburgh | Royal Hospital for Sick Children | 143 | 100 | 55.8 | 56.0 | 57.3 | |
| Glasgow | Royal Hospital for Sick Children | 56 | 75 | 57.7 | 58.1 | 56.8 | |
| Inverness | Raigmore Hospital | 31 | 12 | 46.1 | 46.1 | 39.8 | |
| Lanarkshire | Wishaw General Hospital | 162 | 24 | 46.3 | 45.7 | 42.3 | |

Adult centres/clinics providing data in 2020 – ordered alphabetically by location

| | | | BMI; kg/m² | | | |
|-----------|----------------------------|-----------|------------|----------------------|---------------------------------|--------|
| Location | Name | Clinic ID | Number | Mean - unadjusted | Mean - adjusted [†] | Median |
| Scotland | | | | | | |
| Aberdeen | Aberdeen Royal Infirmary | 70 | 72 | 24.8 | 24.6 | 23.3 |
| Edinburgh | Western General Hospital | 44 | 215 | 24.0 | 23.9 | 22.7 |
| Glasgow | Gartnavel General Hospital | 79 | 189 | 24.3 | 24.2 | 23.7 |

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

| Chronic Pseudomonas | | Having at least 1 IV day | | Receiving DNase treatment | | Receiving hypertonic saline/mannitol treatment | | Inhaled antibiotic use among patients with chronic <i>Pseudomonas</i> | |
|------------------------|-------------------|-----------------------------|-------------------|------------------------------|-------------------|--|-------------------|---|-------------------|
| Number | Proportion (%) | Number | Proportion (%) | Number | Proportion (%) | Number | Proportion (%) | Number | Proportion (%) |
| | | | | | | | | | |
| 0 | 0.0 | 5 | 20.8 | 11 | 45.8 | 0 | 0.0 | 0 | 0.0 |
| <5 | 4.3 | 5 | 21.7 | <5 | 8.7 | 9 | 39.1 | <5 | 100.0 |
| <5 | 25.0 | <5 | 12.5 | <5 | 50.0 | <5 | 12.5 | <5 | 50.0 |
| 10 | 8.5 | 36 | 30.8 | 73 | 62.4 | 31 | 26.5 | 8 | 80.0 |
| <5 | 2.9 | 38 | 36.5 | 30 | 28.8 | 43 | 41.3 | <5 | 100.0 |
| <5 | 7.7 | <5 | 7.7 | <5 | 30.8 | <5 | 15.4 | <5 | 100.0 |
| 0 | 0.0 | 5 | 12.5 | 9 | 22.5 | 15 | 37.5 | 0 | 0.0 |

| Chronic Pseudomonas | | Having at least 1 IV day | | Receiving DNase treatment | | Receiving hypertonic saline/mannitol treatment | | Inhaled antibiotic use among patients with chronic <i>Pseudomonas</i> | |
|------------------------|-------------------|-----------------------------|-------------------|------------------------------|-------------------|--|-------------------|---|-------------------|
| Number | Proportion (%) | Number | Proportion (%) | Number | Proportion (%) | Number | Proportion (%) | Number | Proportion (%) |
| | | | | | | | | | |
| 15 | 20.8 | 18 | 25.0 | 42 | 58.3 | 22 | 30.6 | 13 | 86.7 |
| 52 | 24.2 | 77 | 35.8 | 136 | 63.3 | 42 | 19.5 | 43 | 82.7 |
| 9 | 4.1 | 81 | 37.2 | 97 | 44.5 | 31 | 14.2 | 9 | 100.0 |

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 | Ν | % |
|---------------------|-------------------|---------------|-----|------|
| c.1521_1523delCTT | p.Phe508del | F508del | 737 | 90.5 |
| c.1652G->A | p.Gly551Asp | G551D | 84 | 10.3 |
| c.350G->A | p.Arg117His | R117H | 66 | 8.1 |
| c.1624G->T | p.Gly542X | G542X | 55 | 6.8 |
| c.200C->T | p.Pro67Leu | P67L | 41 | 5.0 |
| c.1679G->C | p.Arg560Thr | R560T | 18 | 2.2 |
| c.1585-1G->A | | 1717-1G->A | 13 | 1.6 |
| c.1477C->T | p.Gln493X | Q493X | 12 | 1.5 |
| c.3909C->G | p.Asn1303Lys | N1303K | 11 | 1.4 |
| c.3454G->C | p.Asp1152His | D1152H | 10 | 1.2 |
| c.2657+5G->A | | 2789+5G->A | 10 | 1.2 |
| c.1558G->T | p.Val520Phe | V520F | 8 | 1.0 |
| c.3717+12191C->T | | 3849+10kbC->T | 7 | 0.9 |
| c.489+1G->T | | 621+1G->T | 7 | 0.9 |
| c.178G->T | p.Glu60X | E60X | 6 | 0.7 |
| c.948delT | p.Phe316LeufsX12 | 1078delT | 6 | 0.7 |
| c.2657+2_2657+3insA | | 2789+2insA | 5 | 0.6 |
| c.1364C->A | p.Ala455Glu | A455E | 5 | 0.6 |
| c.3528delC | p.Lys1177SerfsX15 | 3659delC | 5 | 0.6 |
| c.1721C->A | p.Pro574His | P574H | <5 | - |
| c.1766+1G->A | | 1898+1G->A | <5 | - |
| c.3196C->T | p.Arg1066Cys | R1066C | <5 | - |
| c.3846G->A | p.Trp1282X | W1282X | <5 | - |
| c.1705T->G | p.Tyr569Asp | Y569D | <5 | - |
| c.2012delT | p.Leu671X | 2143delT | <5 | - |
| c.1523T->G | p.Phe508Cys | F508C | <5 | - |
| c.579+3A->G | | 711+3A->G | <5 | - |
| c.223C->T | p.Arg75X | R75X | <5 | - |
| c.3140-26A->G | | 3272-26A->G | <5 | - |
| c.1367T->C | p.Val456Ala | V456A | <5 | - |
| c.2988+1G->A | | 3120+1G->A | <5 | - |
| c.2052delA | p.Lys684AsnfsX38 | 2184delA | <5 | - |
| c.3468G->A | | 3600G->A | <5 | - |
| c.1680A->C | p.Arg560Ser | R560S | <5 | - |
| c.254G->A | p.Gly85Glu | G85E | <5 | - |
| c.2988G->A | | 3120G->A | <5 | - |

| Nucleotide | Protein | Legacy name | Ν | % |
|--|------------------|-------------|----|---|
| c.1210-12[5] (AJ574948.1:g.152T[5]) | | 5T | <5 | - |
| c.3884_3885insT | p.Ser1297PhefsX5 | 4016insT | <5 | - |
| c.3158C->T | p.Thr1053lle | T1053I | <5 | - |
| c.3705T->G | p.Ser1235Arg | S1235R | <5 | - |
| c.1327G->T | p.Asp443Tyr | D443Y | <5 | - |
| c.274G->A | p.Glu92Lys | E92K | <5 | - |
| c.3484C->T | p.Arg1162X | R1162X | <5 | - |
| c.509G->A | p.Arg170His | R170H | <5 | - |
| c.4004T->C | p.Leu1335Pro | L1335P | <5 | - |
| c.1209+1G->A | | 1341+1G->A | <5 | - |
| c.3476C->T | p.Ser1159Phe | S1159F | <5 | - |
| c.1006_1007insG | p.lle336SerfsX28 | 1138insG | <5 | - |
| c.1657C->T | p.Arg553X | R553X | <5 | - |
| c.1055G->A | p.Arg352Gln | R352Q | <5 | - |
| c.273+1G->A | | 405+1G->A | <5 | - |
| c.2490+1G->A | | 2622+1G->A | <5 | - |
| c.2859_2890delACATTCT- GTTCTTCAAGCACCTATGT- CAACCC | p.Leu953PhefsX11 | 2991del32 | <5 | - |
| c.2583delT | p.Phe861LeufsX3 | 2711delT | <5 | - |
| c.1519_1521delATC | p.lle507del | 1507del | <5 | - |
| c.3266G->A | p.Trp1089X | W1089X | <5 | - |
| c.1466C->A | p.Ser489X | S489X | <5 | - |
| c.1585-8G->A | | 1717-8G->A | <5 | - |
| c.292C->T | p.Gln98X | Q98X | <5 | - |
| Other | | | <5 | - |

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