Cystic Fibrosis Trust

# UK Cystic Fibrosis Registry
## 2021 Annual Data Report

September 2022

Uniting for a life unlimited

# UK Cystic Fibrosis Registry
# 2021 Annual Data Report

**An at-a-glance version of this report can be found at cysticfibrosis.org.uk/registry**

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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 Cystic Fibrosis Trust. We would also like to express our gratitude to the UK cystic fibrosis centres and clinics, for their continued dedication to obtaining consent and submitting data to the Registry.

## Contact information

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

Cystic Fibrosis Trust

# Contents

# Foreword

This report covers a year of continued challenge and change, with the ongoing impact of COVID-19, but also real progress for people with cystic fibrosis, as access to Kaftrio increases across the community.

As this new data reflects, during the second year of the pandemic, the way in which people with CF used health services continued to show differences from previous years. The proportion of people having annual reviews remains stable when compared to 2020 at 93%, but remains below the figures shown pre-2019. It is also interesting to note that 30% of annual reviews were recorded as taking place virtually, which indicates the persistence of some of the new ways of delivering care adopted during the first wave of COVID-19.

The scale and scope of the data collected by the UK CF Registry continued to grow, as more frequent data collection to support the reporting of the effectiveness of new medicines continued. CF teams entered over 32,000 encounters onto the system, including over 10,000 annual review datasets. I would like to thank everyone involved in making this possible. It is an amazing achievement – and we couldn't do it without the dedication of CF teams across the country.

As the CF landscape continues to change, the fact that 99% of people with CF and their families consent to their data being captured on the UK CF Registry means we can understand the increasing diversity of our community and use this to ensure that research is targeted so that nobody in the community is left behind.

It also means that throughout the year, the UK CF Registry was able to support domestic and international efforts to monitor the impact of COVID-19, receiving reports of infections and recording vaccinations. During 2021 we recorded 814 cases of infection, with a much lower rate of hospitalisation of 7.6%, compared to the 2020 figure of 24%.

The team continues to focus on ways to use the Registry to make a difference for people with cystic fibrosis. Over the last 12 months we have continued the technical development of the Registry platform to support CF STORM, a Registry-based clinical trial. The trial is focused on the safety and impact of stopping some medicines that people may not need when they are taking elexacaftor/tezacaftor/ivacaftor. We are continuing to support the forthcoming NICE (National Institute of Health and Care Excellence) appraisal of CFTR modulator therapies, through additional data collection and analysis.

In a time of profound change, this report brings so many useful insights that really can shape our work moving forwards. There are positives we can take, with an increase in the number of people with CF having families, and a much-improved picture for overall health. The predicted median survival age of people born today has increased to 53 years. But this is still far too young – and shows there is still much more work we need to do. We won't stop until everyone with CF can truly live a life unlimited: mentally well, physically well, and personally fulfilled.

I hope you enjoy reading the report and would love to hear your feedback. Please contact us on social media or by emailing **registry@cysticfibrosis.org.uk** to let us know your comments and questions.

Finally, and most importantly, I want to extend my thanks to people with cystic fibrosis for continuing to support the UK CF Registry, as well as their families and their clinical teams for coming together to make this report possible.

**David Ramsden**
Chief Executive of Cystic Fibrosis Trust

# Executive summary

The 2021 Registry data continues to be a rich resource to help CF teams, researchers and people with cystic fibrosis understand the current health of people with CF in the UK. The report will still be impacted by the effects of the COVID-19 pandemic, with some CF teams again re-deployed to other areas of the hospital and most hospital clinics still running some or all of their outpatient appointments remotely. This again is reflected in the slightly lower percentage of people having annual reviews (93%) compared to pre-pandemic.

Nevertheless, it is still a rich resource and I will try to bring to your attention some of the highlights of this year's report.

- 10,175 people had an annual review, and they form the basis of this report
- 61.9% of the population are over 16 years of age (57.9% are ≥ 18 years)
- 5.8% of the UK CF population report being non-white or of mixed ethnicity
- The slight downward trend in numbers of people newly diagnosed with CF continues, with the amended number for 2020 being 239. We have made a further update to the 2019 figure, which is now 276
- Nutritional status is improving, and a new graph (section 1.9) shows BMI trends over the last four years, with a smaller proportion now being underweight
- The median best $FEV_1$ continues to rise and is now 86.9% (section 1.15). The comparison since 2011 is shown in the next graph, with a jump apparent in the ≥ 12-year mark
- Depression in those ≥16 years old remains constant at 8.1% (section 1.22)
- 103 women had babies in 2021, nearly double the number of the previous year
- The figures for *Pseudomonas aeruginosa* infections continue to change, the intermittent figure for adults increasing (23.4%), with the chronic again decreasing (20.2%)
- Growths of NTM (6.2%) and Aspergillus (10.3%) (section 1.17, p29) have both dropped significantly, which may be related to improved health or the CFTR modulator effect of less sputum production
- The proportion of people receiving at least one course of IV antibiotics has dropped again, with only 24.3% reported compared to 39.2% in 2020. This represents 1,418 less people needing IV's in 2021 (section 1.25)
- The proportion of adults requiring oxygen has dropped in a year from 8.3% to 6.1% (section 1.35)
- The overall numbers reported as having any supplemental feeds have dropped as well, from 43.2% to 34.6% in 2021 (section 1.37)
- 7,384 people were reported as being on a CFTR modulator by December 2021 (section 1.34)
- There are quite different genotype distributions across the devolved nations, shown in section 1.45 (p49).

Sections 2 and 3 are the centre level reports which continue to show interesting differences between centres in the use of home compared to hospital IV antibiotics. They also highlight the differences in types of mucolytics used. Tables of outcome data for centres must be interpreted with caution, a lot of centres are not large enough to allow meaningful comparisons.

The trends of not just the COVID-19 pandemic but the more widespread introduction of CFTR modulators appear to be showing through in the data. This is important information that we hope you find useful. Again, I would like to thank the people with CF for consenting to have their anonymised clinical data recorded and the clinical teams for entering it into the Registry.

**Dr Siobhán B Carr**
Chair of the UK CF Registry Steering Committee

# 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 64.

An at-a-glance version of this report can be found at **cysticfibrosis.org.uk/registry**.

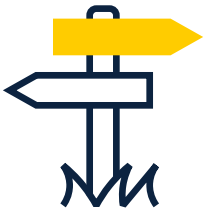
## Cystic fibrosis

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

## UK Cystic Fibrosis Registry

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

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



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

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

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

Providing data for research to find out the best ways to treat cystic fibrosis.

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

## 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 consent is given by the person with CF, or, if they're 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?

You can find out more about CF, and the UK CF Registry, at **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

| | 2016 | 2017 | 2018 | 2019 | 2020 | 2021 |
|---|---|---|---|---|---|---|
| CF patients Registered[1] | 10461 | 10469 | 10509 | 10655 | 10837 | 10908 |
| Excluding diagnoses that year | 10214 | 10255 | 10287 | 10462 | 10632 | 10720 |
| CF patients with an annual review; n(%)[2] | 9695 (95) | 9887 (96) | 9847 (96) | 10070 (96) | 9922 (93*) | 10175 (93) |
| Age in years; median[3] | 20 | 20 | 20 | 21 | 21 | 21 |
| All newly diagnosed patients (NBS and other)[4] | 247 | 214 | 222 | 193 | 205 | 188 |
| All newly diagnosed patients (amended)[5] | (322) | (304) | (301) | (276) | (239) | (TBD) |
| Number of patients born identified by NBS[4] | 216 | 192 | 167 | 150 | 152 | 134 |
| Age at diagnosis in months; median[3] | 2 | 2 | 2 | 2 | 2 | 2 |
| Adults aged 16 years and over; %[3] | 60.4 | 60.6 | 60.4 | 60.6 | 60.6 | 61.9 |
| Males; %[3] | 53.2 | 53.3 | 53 | 53.2 | 53.1 | 53.2 |
| Genotyped; %[3] | 98.4 | 99.3** | 99.1 | 99.2 | 99.2 | 99.1 |
| Total deaths reported during annual review year (%)[6] | 148 (1.5%) | 132 (1.3%) | 137 (1.3%) | 114 (1.1%) | 97 (0.9%) | 66 (0.6%) |
| Total deaths reported amended (%)[5] | 159 (1.5%) | 143 (1.4%) | 143 (1.4%) | 118 (1.1%) | 101 (1.0%) | (TBD) |
| Age at death in years; median (95% CI)[6] | 31 (29, 33) | 31(29, 35) | 32 (29, 35) | 31 (29, 34) | 36 (32, 38) | 38 (36, 42) |

**Annual review:** A Registry annual review form records 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.

Notes:

* Corrected from 2020 report.

** Corrected from 2017 report.

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

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

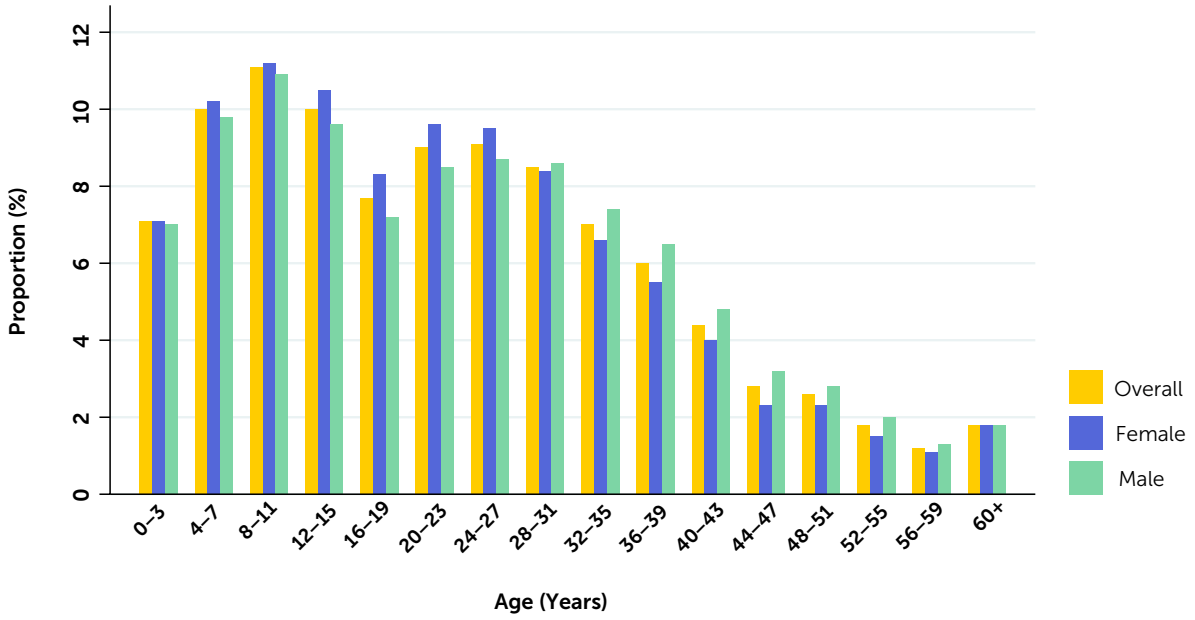
3 Calculated from patients with an annual review in the given year (see footnote 5 below).

4 Calculated from all patients registered on the database. Some diagnosis data are added after the data entry closure each year, so figures are updated the following year (see below) .

5 Amended values refer to new diagnoses or deaths that occurred within the given year but were not recorded on the Registry until after data collection closure. We first presented the amended figures in the 2019 data report. In this report we have completed an additional data cleaning exercise and so some earlier figures have also been updated.

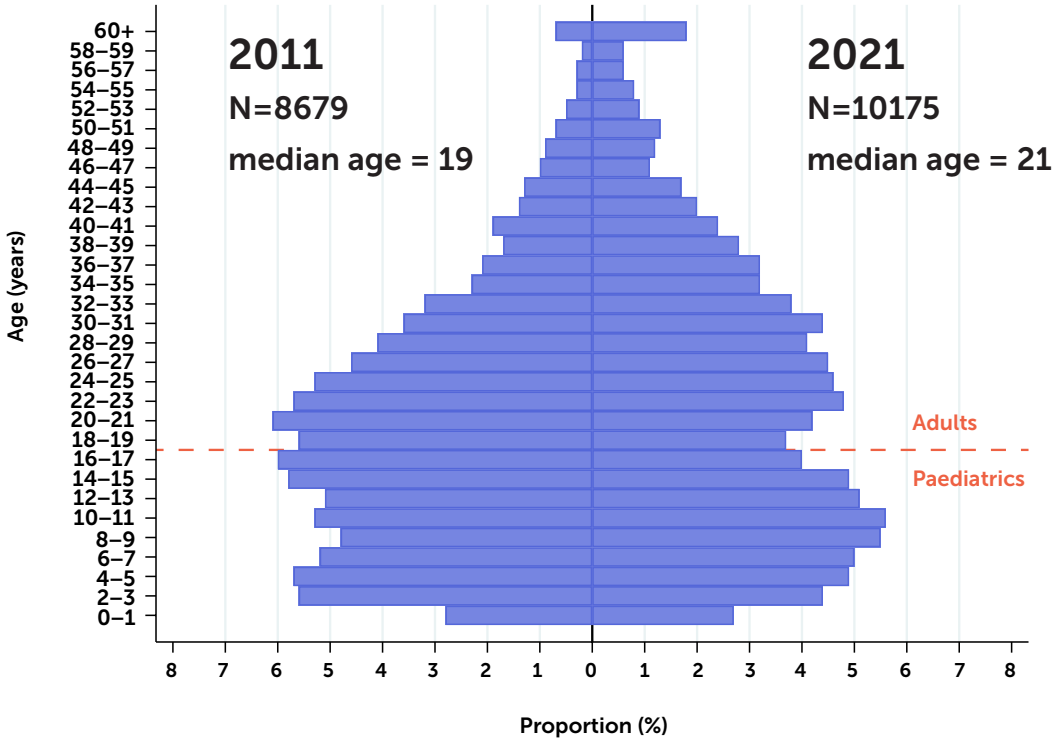
6 Calculated from all registered patients who died in the given year.

## 1.2 Age distribution by sex
## N=10175




| Age | All; n (%) | Females; n (%) | Males; n (%) |
|---|---|---|---|
| 0-3 | 718 (7.1) | 340 (7.1) | 378 (7.0) |
| 4-7 | 1013 (10.0) | 485 (10.2) | 528 (9.8) |
| 8-11 | 1127 (11.1) | 535 (11.2) | 592 (10.9) |
| 12-15 | 1020 (10.0) | 498 (10.5) | 522 (9.6) |
| 16-19 | 785 (7.7) | 397 (8.3) | 388 (7.2) |
| 20-23 | 919 (9.0) | 458 (9.6) | 461 (8.5) |
| 24-27 | 926 (9.1) | 454 (9.5) | 472 (8.7) |
| 28-31 | 865 (8.5) | 399 (8.4) | 466 (8.6) |
| 32-35 | 713 (7.0) | 315 (6.6) | 398 (7.4) |
| 36-39 | 614 (6.0) | 262 (5.5) | 352 (6.5) |
| 40-43 | 451 (4.4) | 190 (4.0) | 261 (4.8) |
| 44-47 | 280 (2.8) | 108 (2.3) | 172 (3.2) |
| 48-51 | 261 (2.6) | 111 (2.3) | 150 (2.8) |
| 52-55 | 179 (1.8) | 72 (1.5) | 107 (2.0) |
| 56-59 | 123 (1.2) | 52 (1.1) | 71 (1.3) |
| 60+ | 181 (1.8) | 85 (1.8) | 96 (1.8) |
| <16 | 3878 (38.1) | 1858 (39.0) | 2020 (37.3) |
| ≥16 | 6297 (61.9) | 2903 (61.0) | 3394 (62.7) |
| <18 | 4283 (42.1) | 2064 (43.4) | 2219 (41.0) |
| ≥18 | 5892 (57.9) | 2697 (56.6) | 3195 (59.0) |
| Overall | 10175 | 4761 | 5414 |

## 1.3 Age distribution of the UK CF population in 2011 vs 2021




## 1.4 Ethnicity

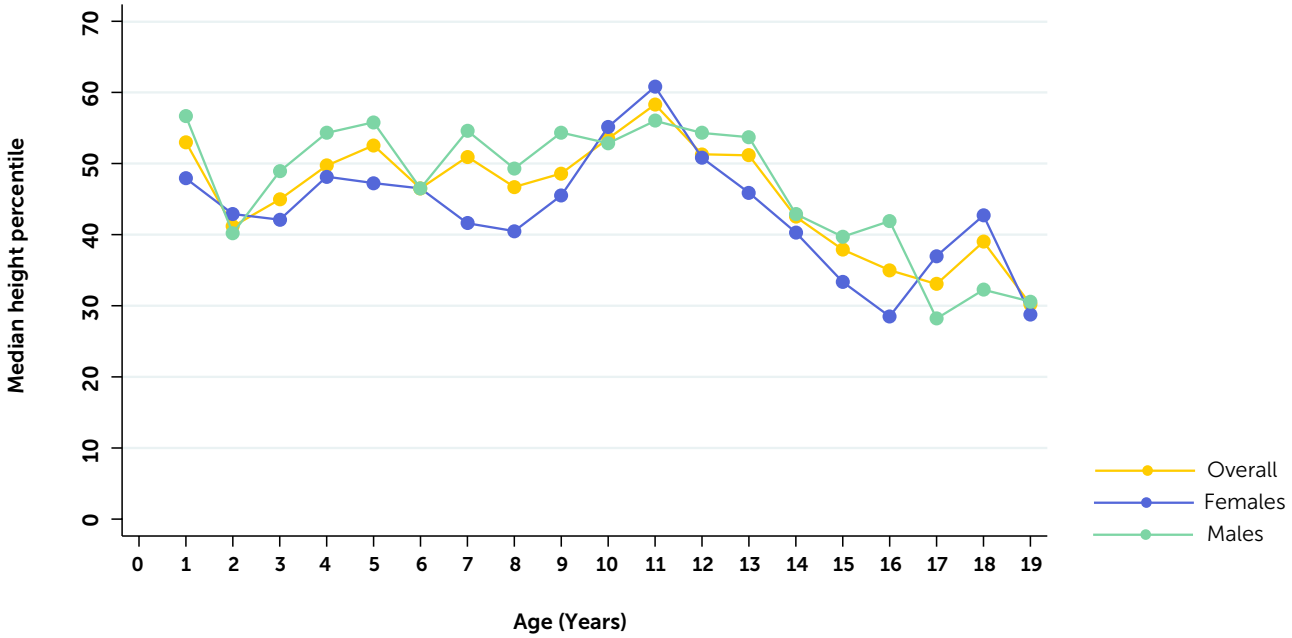
| | 2011 | 2016 | 2021 |
|---|---|---|---|
| Total | 8679 | 9695 | 10175 |
| **White** | | | |
| White | 8251 (95.1) | 9186 (94.7) | 9375 (92.1) |
| **Asian** | | | |
| Bangladeshi | 32 (0.4) | 33 (0.3) | 46 (0.5) |
| Indian | 25 (0.3) | 36 (0.4) | 48 (0.5) |
| Pakistani | 139 (1.6) | 155 (1.6) | 183 (1.8) |
| Other (Asian) | 22 (0.3) | 24 (0.2) | 32 (0.3) |
| **Black** | | | |
| Black African | -* | 12 (0.1) | 13 (0.1) |
| Black Caribbean | 14 (0.2) | 16 (0.2) | 10 (0.1) |
| Other (Black) | <5 | <5 | 5 (0.0) |
| **Mixed**** | | | |
| Mixed | 51 (0.6) | 24 (0.2) | 71 (0.7) |
| Mixed (white-Asian) | - | 6 (0.1) | 17 (0.2) |
| Mixed (white-Black African) | - | <5 | 12 (0.1) |
| Mixed (white-Black Caribbean) | - | 9 (0.1) | 24 (0.2) |
| Other (mixed) | - | 6 (0.1) | 18 (0.2) |
| **Other/Unknown** | | | |
| Other | 81 (0.9) | 92 (0.9) | 105 (1.0) |
| Unknown | 49 (0.6) | 113 (1.2) | 287 (2.8) |

* redacted to adhere to statistical disclosure guidelines.

** Further detail on mixed ethnicity categories were collected from 2016 onwards.

## 1.5 Height percentiles of children and young people (<20 years)[1] N=4663

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.



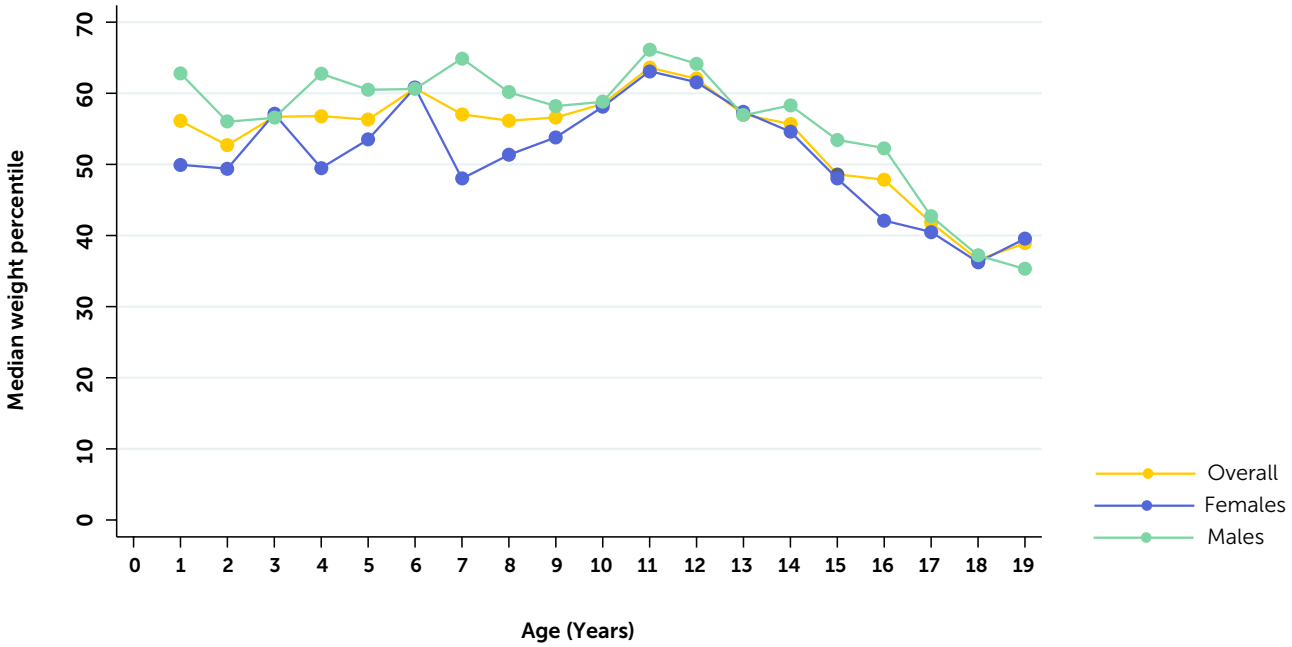

| | Overall | | | Female | | | Male | | |
|---|---|---|---|---|---|---|---|---|---|
| Age | n | Median | IQR | n | Median | IQR | n | Median | IQR |
| 1 | 165 | 53.0 | 26.6-83.7 | 83 | 47.9 | 21.6-84.0 | 82 | 56.7 | 28.5-80.9 |
| 2 | 196 | 41.2 | 19.5-73.4 | 88 | 42.9 | 17.6-72.4 | 108 | 40.2 | 19.8-73.4 |
| 3 | 190 | 44.9 | 24.1-75.2 | 79 | 42.1 | 22.9-75.2 | 111 | 48.9 | 25.4-75.3 |
| 4 | 225 | 49.7 | 21.1-81.0 | 112 | 48.2 | 17.0-75.9 | 113 | 54.3 | 25.0-84.0 |
| 5 | 230 | 52.5 | 24.9-75.1 | 116 | 47.3 | 23.8-73.2 | 114 | 55.8 | 27.2-77.0 |
| 6 | 235 | 46.5 | 22.1-76.2 | 110 | 46.5 | 22.1-75.6 | 125 | 46.5 | 24.3-76.2 |
| 7 | 244 | 50.9 | 23.8-72.4 | 117 | 41.6 | 16.4-69.1 | 127 | 54.6 | 28.7-80.8 |
| 8 | 239 | 46.7 | 22.1-77.2 | 107 | 40.5 | 20.2-78.7 | 132 | 49.3 | 24.5-76.3 |
| 9 | 281 | 48.6 | 24.5-75.2 | 141 | 45.5 | 24.1-76.3 | 140 | 54.3 | 25.8-74.8 |
| 10 | 266 | 53.5 | 27.6-78.2 | 136 | 55.2 | 23.3-78.2 | 130 | 52.8 | 30.2-77.6 |
| 11 | 259 | 58.3 | 32.8-80.0 | 120 | 60.8 | 36.6-80.4 | 139 | 56.0 | 27.5-77.1 |
| 12 | 245 | 51.3 | 25.9-76.7 | 123 | 50.8 | 25.2-75.0 | 122 | 54.3 | 26.4-78.0 |
| 13 | 248 | 51.2 | 25.6-76.6 | 117 | 45.9 | 21.7-69.4 | 131 | 53.7 | 28.6-81.1 |
| 14 | 255 | 42.5 | 19.7-71.4 | 118 | 40.3 | 19.7-70.8 | 137 | 42.9 | 21.4-71.4 |
| 15 | 223 | 37.9 | 16.7-62.0 | 114 | 33.3 | 12.3-63.1 | 109 | 39.7 | 21.1-61.5 |
| 16 | 214 | 35.0 | 16.3-60.6 | 102 | 28.5 | 11.1-45.9 | 112 | 41.9 | 20.0-66.9 |
| 17 | 172 | 33.1 | 10.9-59.7 | 93 | 37.0 | 13.9-59.7 | 79 | 28.2 | 8.9-55.2 |
| 18 | 178 | 39.0 | 10.4-65.3 | 88 | 42.7 | 15.7-62.3 | 90 | 32.3 | 7.2-65.6 |
| 19 | 193 | 30.2 | 11.9-59.0 | 98 | 28.8 | 13.6-65.3 | 95 | 30.6 | 9.0-53.9 |
| **Overall** | **4258*** | **45.9** | **21.4-73.7** | **2062** | **43.8** | **19.9-73.4** | **2196** | **48.0** | **22.7-74.4** |

* number with non-missing data.

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

## 1.6 Weight percentiles of children and young people (<20 years)[1] N=4663

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.



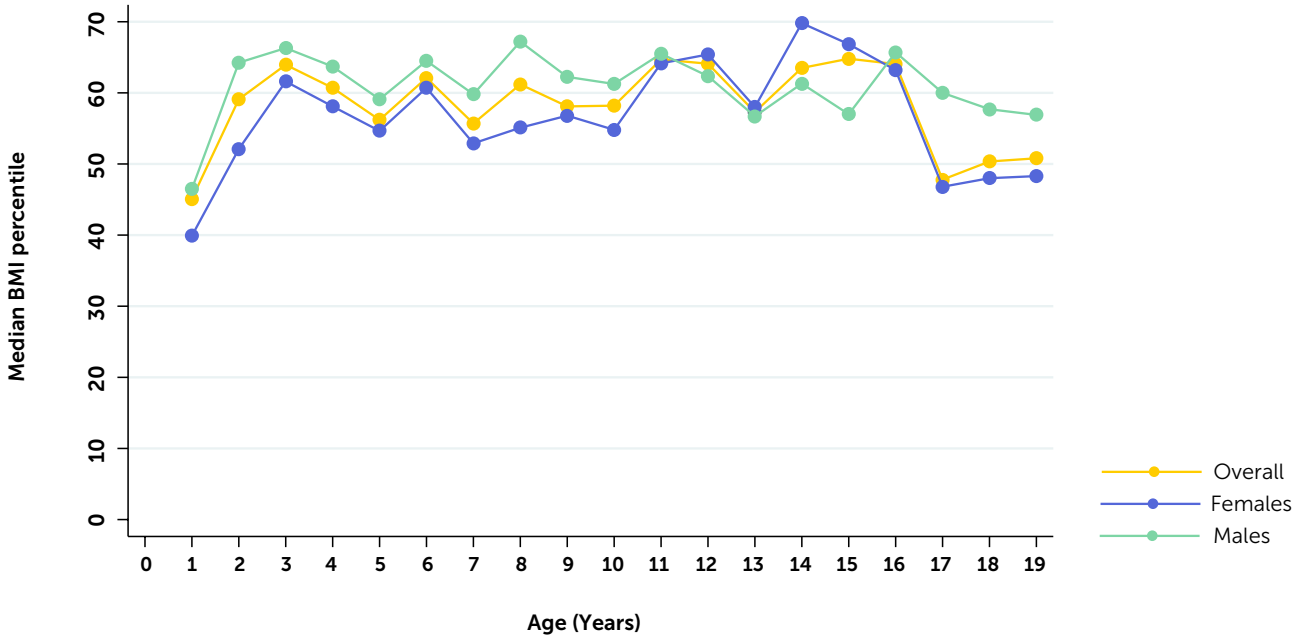

| | Overall | | | Female | | | Male | | |
|---|---|---|---|---|---|---|---|---|---|
| Age | n | Median | IQR | n | Median | IQR | n | Median | IQR |
| 1 | 183 | 56.1 | 24.2-81.9 | 92 | 50.0 | 19.5-80.8 | 91 | 62.8 | 26.1-83.8 |
| 2 | 217 | 52.7 | 28.4-77.6 | 99 | 49.4 | 21.6-73.9 | 118 | 56.0 | 32.6-79.7 |
| 3 | 203 | 56.7 | 31.5-80.1 | 83 | 57.1 | 29.1-84.1 | 120 | 56.6 | 33.3-78.8 |
| 4 | 236 | 56.8 | 30.8-81.9 | 114 | 49.5 | 28.6-75.9 | 122 | 62.8 | 37.3-85.8 |
| 5 | 236 | 56.3 | 31.8-78.9 | 119 | 53.5 | 30.3-78.3 | 117 | 60.5 | 35.0-80.4 |
| 6 | 242 | 60.7 | 32.1-80.6 | 111 | 60.8 | 32.8-79.2 | 131 | 60.6 | 29.7-83.4 |
| 7 | 248 | 57.1 | 29.0-82.7 | 118 | 48.0 | 25.5-73.7 | 130 | 64.9 | 34.8-85.4 |
| 8 | 246 | 56.2 | 31.1-82.3 | 112 | 51.3 | 22.5-82.3 | 134 | 60.2 | 38.3-82.7 |
| 9 | 285 | 56.6 | 31.3-80.8 | 142 | 53.8 | 28.5-80.1 | 143 | 58.2 | 35.4-82.3 |
| 10 | 276 | 58.5 | 29.0-83.7 | 140 | 58.1 | 22.8-83.9 | 136 | 58.8 | 32.8-83.1 |
| 11 | 265 | 63.6 | 39.2-84.6 | 121 | 63.1 | 40.8-83.1 | 144 | 66.2 | 36.0-85.1 |
| 12 | 248 | 62.1 | 35.7-83.5 | 124 | 61.6 | 30.3-81.9 | 124 | 64.2 | 39.5-85.3 |
| 13 | 254 | 57.0 | 30.2-83.6 | 121 | 57.4 | 29.3-78.1 | 133 | 56.9 | 30.8-85.4 |
| 14 | 260 | 55.7 | 25.8-85.4 | 121 | 54.6 | 27.2-85.6 | 139 | 58.3 | 25.7-84.7 |
| 15 | 225 | 48.6 | 28.1-76.1 | 115 | 48.0 | 27.1-77.0 | 110 | 53.4 | 28.1-74.0 |
| 16 | 214 | 47.8 | 26.6-74.5 | 103 | 42.1 | 26.1-69.7 | 111 | 52.3 | 27.2-77.6 |
| 17 | 173 | 41.8 | 16.8-69.1 | 94 | 40.5 | 22.6-71.9 | 79 | 42.7 | 11.6-66.0 |
| 18 | 169 | 36.6 | 12.5-68.2 | 82 | 36.3 | 12.6-65.3 | 87 | 37.2 | 11.5-72.6 |
| 19 | 175 | 38.9 | 14.5-71.0 | 90 | 39.6 | 21.0-63.6 | 85 | 35.3 | 8.2-75.3 |
| **Overall** | **4355*** | **54.9** | **28.3-80.1** | **2101** | **51.9** | **26.9-78.3** | **2254** | **57.3** | **29.6-81.6** |

* number with non-missing data.

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

## 1.7 Body Mass Index (BMI) percentiles in children and young people (<20 years)[1]
## N=4663

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 have the same BMI or lower; 60% have a higher BMI.



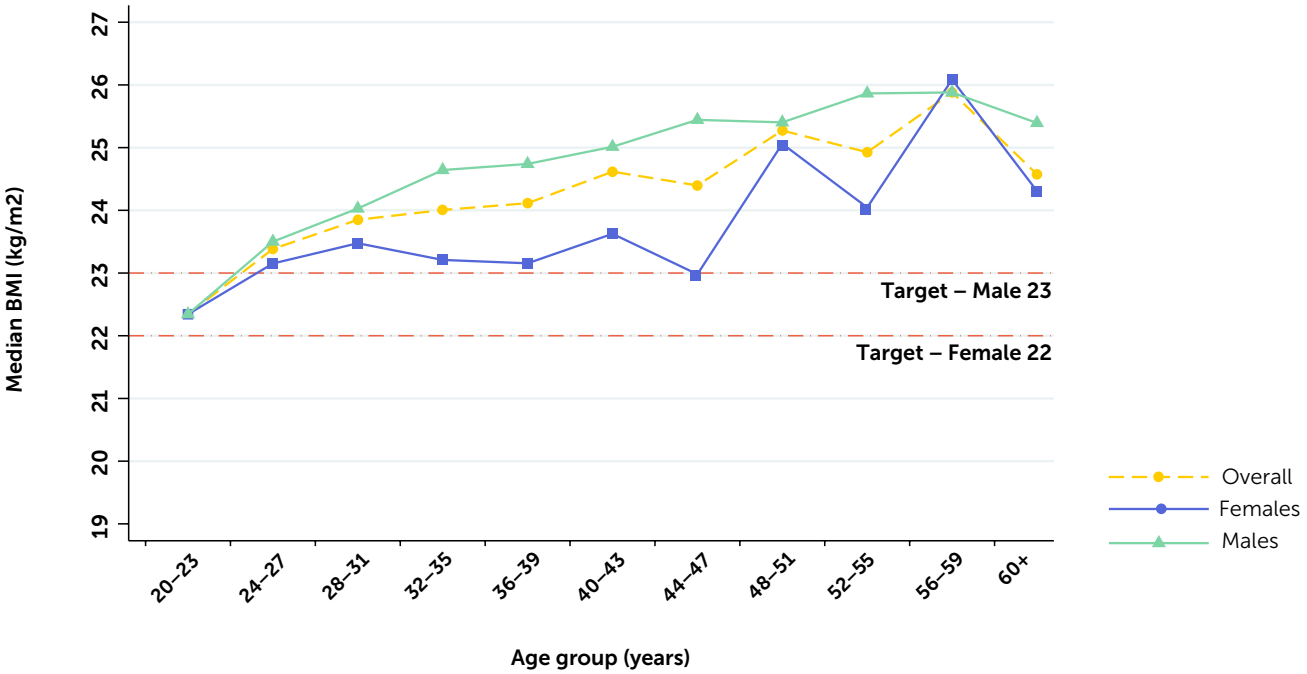

| | Overall | | | Female | | | Male | | |
|---|---|---|---|---|---|---|---|---|---|
| Age | n | Median | IQR | n | Median | IQR | n | Median | IQR |
| 1 | 165 | 45.1 | 17.5-77.5 | 83 | 39.9 | 14.5-72.1 | 82 | 46.5 | 20.5-81.0 |
| 2 | 195 | 59.1 | 33.7-81.0 | 87 | 52.1 | 28.1-76.0 | 108 | 64.3 | 39.1-83.1 |
| 3 | 190 | 63.9 | 38.4-84.2 | 79 | 61.6 | 33.8-84.2 | 111 | 66.3 | 39.3-84.2 |
| 4 | 225 | 60.7 | 33.0-81.2 | 112 | 58.1 | 31.0-77.6 | 113 | 63.7 | 33.9-86.0 |
| 5 | 229 | 56.2 | 35.2-80.7 | 116 | 54.7 | 38.4-80.4 | 113 | 59.1 | 33.8-80.7 |
| 6 | 235 | 62.1 | 39.0-84.0 | 110 | 60.7 | 39.4-79.6 | 125 | 64.5 | 37.5-86.2 |
| 7 | 244 | 55.7 | 33.8-83.2 | 117 | 52.9 | 31.0-74.0 | 127 | 59.8 | 38.5-86.3 |
| 8 | 239 | 61.2 | 38.1-84.9 | 107 | 55.1 | 36.9-83.0 | 132 | 67.2 | 41.8-85.1 |
| 9 | 281 | 58.1 | 32.0-81.4 | 141 | 56.8 | 29.3-79.4 | 140 | 62.3 | 33.9-82.6 |
| 10 | 266 | 58.2 | 30.9-84.0 | 136 | 54.8 | 29.7-80.6 | 130 | 61.3 | 31.4-86.2 |
| 11 | 259 | 64.7 | 36.7-86.7 | 120 | 64.1 | 34.8-82.9 | 139 | 65.5 | 36.7-89.4 |
| 12 | 245 | 64.1 | 34.4-85.5 | 123 | 65.4 | 34.4-80.2 | 122 | 62.3 | 33.3-87.7 |
| 13 | 248 | 57.3 | 30.8-83.4 | 117 | 58.0 | 30.1-82.6 | 131 | 56.7 | 31.5-85.0 |
| 14 | 255 | 63.5 | 36.1-85.1 | 118 | 69.8 | 37.7-85.6 | 137 | 61.3 | 33.5-84.9 |
| 15 | 223 | 64.8 | 35.1-85.6 | 114 | 66.8 | 39.5-87.0 | 109 | 57.0 | 32.5-84.4 |
| 16 | 210 | 64.1 | 38.3-83.0 | 101 | 63.2 | 41.0-83.0 | 109 | 65.7 | 35.2-84.0 |
| 17 | 171 | 47.8 | 29.3-78.6 | 93 | 46.8 | 29.5-79.9 | 78 | 60.0 | 29.3-76.9 |
| 18 | 168 | 50.3 | 26.0-74.8 | 81 | 48.0 | 18.5-70.4 | 87 | 57.7 | 30.3-77.5 |
| 19 | 174 | 50.8 | 25.2-78.7 | 89 | 48.3 | 26.2-71.6 | 85 | 56.9 | 25.2-85.4 |
| **Overall** | **4222*** | **59.7** | **32.7-82.9** | **2044** | **57.2** | **31.3-80.3** | **2178** | **61.2** | **33.8-84.7** |

* number with non-missing data.

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

## 1.8 Body Mass Index (BMI) in adults (20 years and over) N=5512

The following chart and table show the BMI of people with CF aged 20 and over in relation to the target BMI for adults; 22 for women and 23 for men[1].



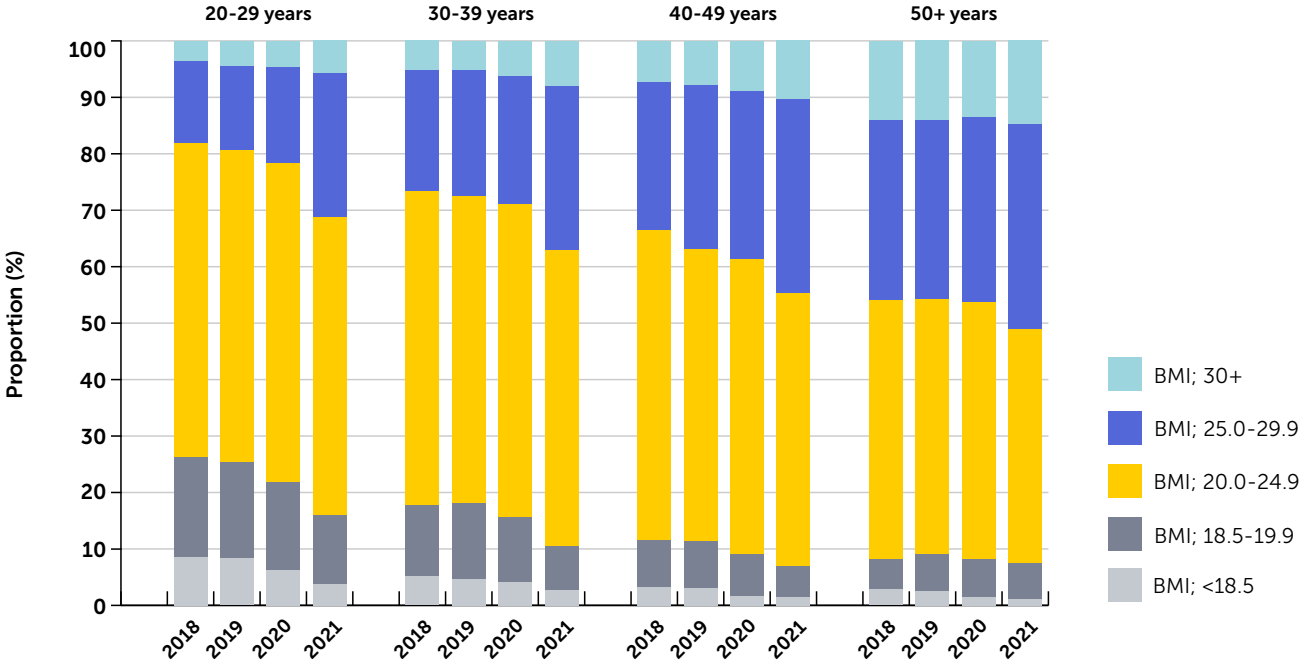

| | Overall | | | Female | | | Male | | |
|---|---|---|---|---|---|---|---|---|---|
| Age | n | Median | IQR | n | Median | IQR | n | Median | IQR |
| 20-23 | 802 | 22.3 | 20.3-25.1 | 403 | 22.3 | 20.3-24.9 | 399 | 22.3 | 20.3-25.1 |
| 24-27 | 799 | 23.4 | 21.1-26.1 | 399 | 23.2 | 20.9-25.8 | 400 | 23.5 | 21.3-26.2 |
| 28-31 | 750 | 23.8 | 21.9-26.2 | 348 | 23.5 | 21.5-26.0 | 402 | 24.0 | 22.1-26.4 |
| 32-35 | 621 | 24.0 | 21.9-26.6 | 280 | 23.2 | 21.5-26.1 | 341 | 24.6 | 22.6-27.0 |
| 36-39 | 525 | 24.1 | 22.0-26.4 | 223 | 23.2 | 21.4-25.7 | 302 | 24.7 | 22.5-26.8 |
| 40-43 | 390 | 24.6 | 22.5-27.2 | 166 | 23.6 | 21.6-26.6 | 224 | 25.0 | 23.1-27.5 |
| 44-47 | 247 | 24.4 | 22.2-26.9 | 96 | 23.0 | 21.5-25.0 | 151 | 25.4 | 23.2-27.8 |
| 48-51 | 217 | 25.3 | 23.3-28.1 | 94 | 25.1 | 22.6-28.3 | 123 | 25.4 | 23.8-27.9 |
| 52-55 | 160 | 24.9 | 22.7-28.1 | 66 | 24.0 | 21.3-26.8 | 94 | 25.9 | 23.1-28.7 |
| 56-59 | 111 | 25.9 | 23.7-29.0 | 46 | 26.1 | 22.6-30.8 | 65 | 25.9 | 24.1-28.4 |
| 60+ | 168 | 24.6 | 22.3-28.1 | 79 | 24.3 | 21.2-28.0 | 89 | 25.4 | 22.8-28.2 |
| **Overall** | **4790*** | **23.9** | **21.6-26.5** | **2200** | **23.3** | **21.2-26.1** | **2590** | **24.3** | **22.1-26.9** |

* number with non-missing data.

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

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

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




| | | Proportion (%) of age group in BMI category, by year | | | | |
|---|---|---|---|---|---|---|
| Age group | | <18.5 | 18.5-19.9 | 20.0-24.9 | 25.0-29.9 | 30+ |
| 20-29 years | 2018 | 8.6 | 17.7 | 55.6 | 14.5 | 3.6 |
| | 2019 | 8.3 | 17.1 | 55.1 | 15.0 | 4.4 |
| | 2020 | 6.3 | 15.5 | 56.6 | 16.9 | 4.7 |
| | 2021 | 3.8 | 12.2 | 52.8 | 25.5 | 5.8 |
| 30-39 years | | | | | | |
| | 2018 | 5.1 | 12.7 | 55.6 | 21.3 | 5.4 |
| | 2019 | 4.7 | 13.3 | 54.5 | 22.2 | 5.3 |
| | 2020 | 4.1 | 11.5 | 55.4 | 22.7 | 6.3 |
| | 2021 | 2.7 | 7.7 | 52.6 | 28.9 | 8.1 |
| 40-49 years | | | | | | |
| | 2018 | 3.3 | 8.2 | 55.0 | 26.1 | 7.3 |
| | 2019 | 3.1 | 8.3 | 51.6 | 29.1 | 7.9 |
| | 2020 | 1.7 | 7.4 | 52.1 | 29.8 | 9.0 |
| | 2021 | 1.5 | 5.4 | 48.5 | 34.3 | 10.4 |
| 50+ years | | | | | | |
| | 2018 | 2.8 | 5.4 | 45.9 | 31.8 | 14.1 |
| | 2019 | 2.6 | 6.5 | 45.1 | 31.8 | 14.0 |
| | 2020 | 1.5 | 6.7 | 45.4 | 32.8 | 13.6 |
| | 2021 | 1.1 | 6.4 | 41.3 | 36.4 | 14.9 |

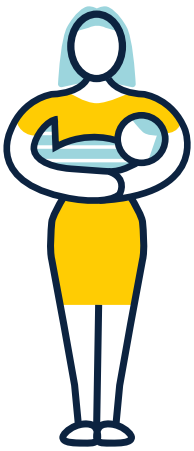
## 1.10 Education and employment in adults (16 years and over) N=6297

The following table shows how people with CF reported their education and employment status in 2021.

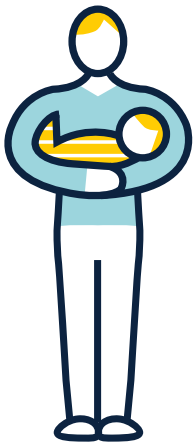
| | 2018 | 2019 | 2020 | 2021 | | |
|---|---|---|---|---|---|---|
| | Overall | Overall | Overall | Overall | Male | Female |
| Number of patients | 5952 | 6104 | 6012 | 6297 | 3394 | 2903 |
| Number who completed questionnaire; n (%) | 5950 (100.0) | 6103 (100.0) | 5968 (99.3) | 6296 (100.0) | 3394 (100.0) | 2902 (100.0) |
| Full-time employment; n (%) | 1956 (32.9) | 2048 (33.6) | 1975 (32.9) | 2097 (33.3) | 1388 (40.9) | 709 (24.4) |
| Part-time employment; n (%) | 926 (15.6) | 958 (15.7) | 894 (14.9) | 915 (14.5) | 354 (10.4) | 561 (19.3) |
| Student; n (%) | 937 (15.7) | 969 (15.9) | 1015 (16.9) | 1061 (16.8) | 515 (15.2) | 546 (18.8) |
| Homemaker; n (%) | 237 (4.0) | 231 (3.8) | 200 (3.3) | 251 (4.0) | 38 (1.1) | 213 (7.3) |
| Unemployed; n (%) | 814 (13.7) | 825 (13.5) | 847 (14.1) | 791 (12.6) | 458 (13.5) | 333 (11.5) |
| Disabled; n (%) | 359 (6.0) | 327 (5.4) | 274 (4.6) | 255 (4.0) | 144 (4.2) | 111 (3.8) |
| Retired; n (%) | 133 (2.2) | 145 (2.4) | 139 (2.3) | 162 (2.6) | 88 (2.6) | 74 (2.5) |
| Volunteer; n (%) | -* | 8 (0.1) | 11 (0.2) | 12 (0.2) | 5 (0.1) | 7 (0.2) |
| Unknown entered; n (%) | 588 (9.9) | 592 (9.7) | 613 (10.2) | 752 (11.9) | 404 (11.9) | 348 (12.0) |
| No. in work or study; n (%) | 3819 (64.2) | 3975 (65.1) | 3884 (65.1) | 4073 (64.7) | 2257 (66.5) | 1816 (62.6) |

## 1.11 Pregnancy

| | 2018 | 2019 | 2020 | 2021 |
|---|---|---|---|---|
| Women with CF who had babies; n | 65 | 58 | 56 | 103 |
| Men with CF who became fathers; n | 45 | 45 | 44 | 30 |



**103 women** with CF had babies in 2021



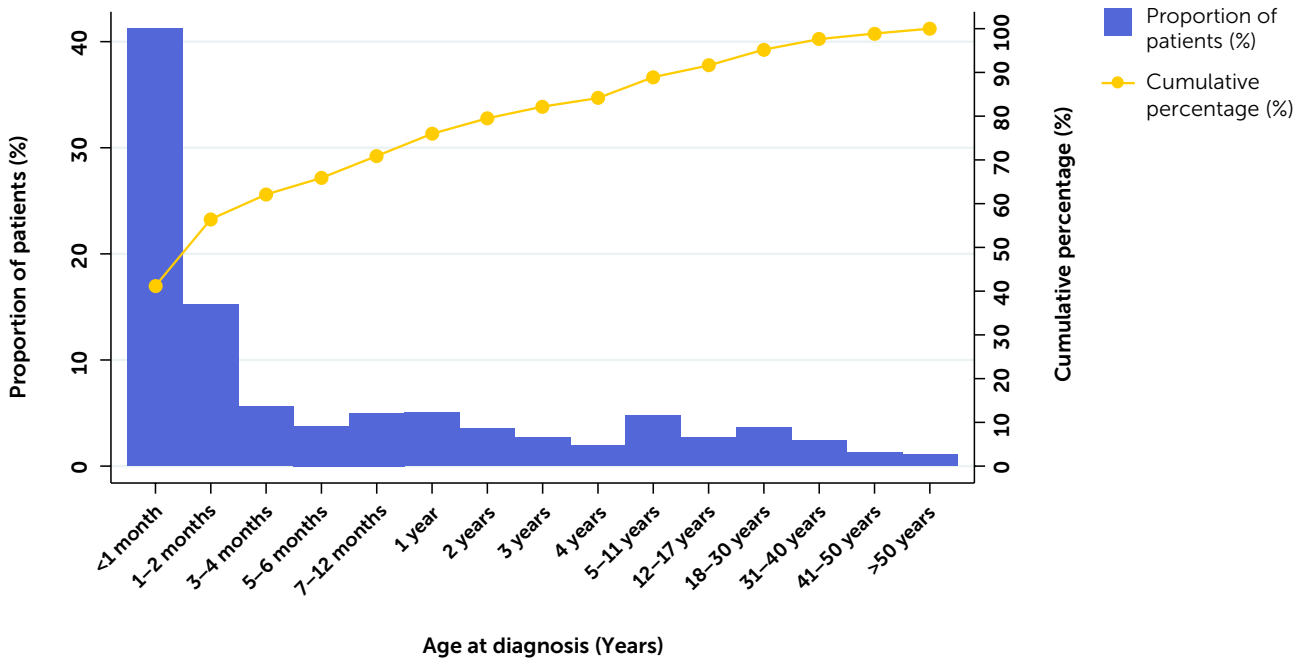
**30 men** with CF became fathers in 2021

*Newly added in 2019.

** Redacted to adhere to statistical disclosure guidelines.

# Diagnosis of cystic fibrosis

## 1.12 Age at diagnosis
## N=10174




The median age at diagnosis for patients aged under 16 in 2021 is **22 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 **134 (71%)** out of 188 patients born in 2021 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. The number of patients identified in 2020 is slightly lower (148) in this report than was recorded in the previous annual report.

**926 (14.7%)** of adults with CF in the Registry in 2021 were diagnosed at age 16 or over.

In 2021, **20** people aged 16 or over were newly diagnosed with cystic fibrosis.

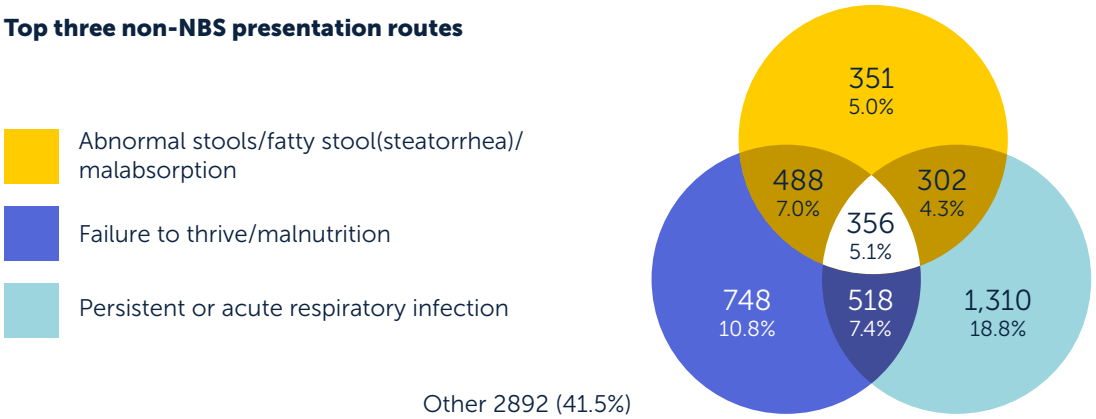
## 1.13 Mode of presentation

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 | 10175 | 9248 | 926 |
| Number diagnosed by newborn screening | 3209 | 3209 | 0 |
| **Total non-NBS** | **6966** | **6039** | **926** |

| Presentation type | All patients | Age <16 at diagnosis* | Age ≥16 at diagnosis* |
|---|---|---|---|
| Total | 6965 (100.0) | 6039 (100.0) | 926 (100.0) |
| Persistant or acute respiratory infection | 2486 (35.7) | 1987 (32.9) | 499 (53.9) |
| Failure to thrive/malnutrition | 2110 (30.3) | 2083 (34.5) | 27 (2.9) |
| Abnormal stools/fatty stool(steatorrhea)/malabsorption | 1497 (21.5) | 1445 (23.9) | 52 (5.6) |
| Meconium ileus | 1296 (18.6) | 1290 (21.4) | 6 (0.6) |
| Family history | 913 (13.1) | 783 (13.0) | 130 (14.0) |
| Genotype | 699 (10.0) | 479 (7.9) | 220 (23.8) |
| Unknown | 328 (4.7) | 271 (4.5) | 57 (6.2) |
| Rectal prolapse | -** | 236 (3.9) | <5 |
| Nasal polyps | 144 (2.1) | 78 (1.3) | 66 (7.1) |
| Bronchiectasis | 103 (1.5) | 10 (0.2) | 93 (10.0) |
| Prenatal | -** | 95 (1.6) | <5 |
| Electrolyte imbalance | 60 (0.9) | 55 (0.9) | 5 (0.5) |
| Fertility | -** | <5 | 47 (5.1) |
| Liver disease | -** | 43 (0.7) | <5 |
| Pancreatitis | 21 (0.3) | 6 (0.1) | 15 (1.6) |
| Oedema | 9 (0.1) | 9 (0.1) | 0 (0.0) |

**Top three non-NBS presentation routes**




Other 2892 (41.5%)

*Age-stratified figures are presented only for those with non-missing diagnosis date.
** redacted to adhere to statistical disclosure guidelines.

# 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, sex, 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 CF who has an $FEV_1$% predicted of 50% breathes out half the volume of air as a comparable person without cystic fibrosis.

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

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

The $FEV_1$% predicted values shown in this report are calculated using an equation called Global Lungs Initiative, or 'GLI'.[1]

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

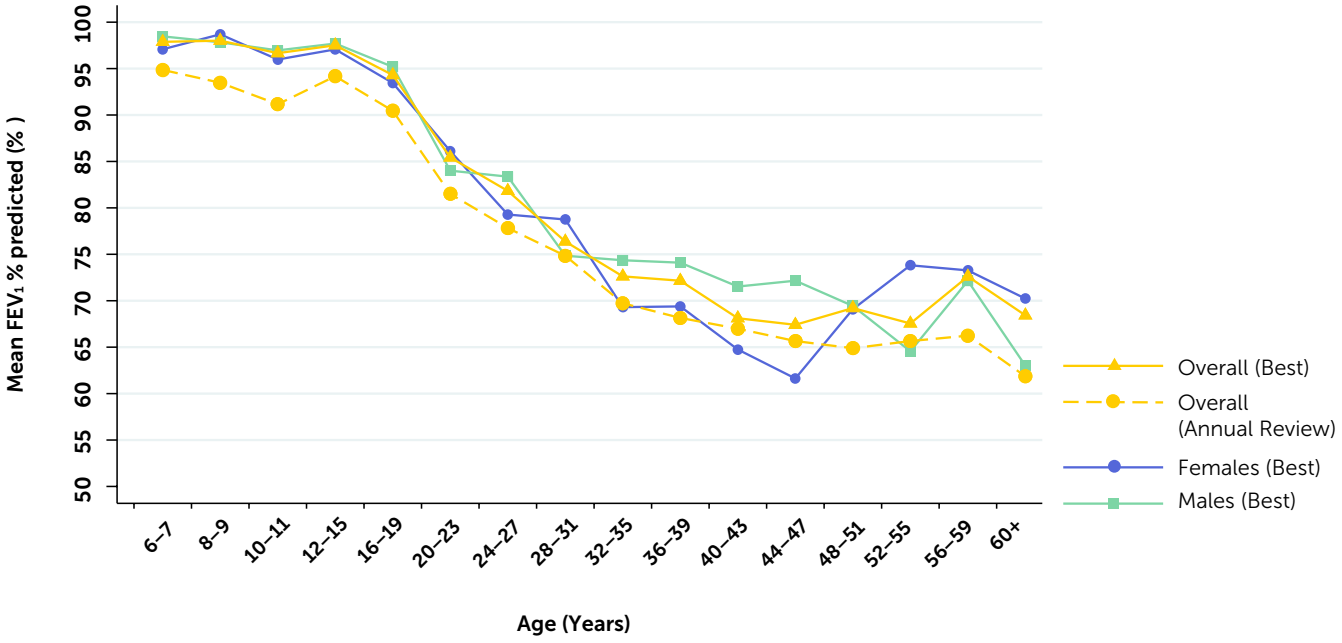
## 1.14 Annual review $FEV_1$% predicted (GLI equations) in patients age six years and older who have not had a lung transplant
## N= 8659

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

| | Overall | | | Female | | | Male | | |
|---|---|---|---|---|---|---|---|---|---|
| Age (yrs) | N | Median | IQR | N | Median | IQR | N | Median | IQR |
| 6-7 | 388 | 94.9 | 84.7-103.3 | 188 | 95.4 | 87.7-103.1 | 200 | 94.0 | 82.9-104.0 |
| 8-9 | 478 | 93.5 | 84.6-101.9 | 229 | 94.0 | 85.0-101.7 | 249 | 93.3 | 84.5-102.0 |
| 10-11 | 490 | 91.2 | 83.0-100.0 | 234 | 91.0 | 82.8-98.8 | 256 | 91.3 | 83.0-100.6 |
| 12-15 | 902 | 94.2 | 83.1-102.5 | 438 | 94.1 | 81.8-102.1 | 464 | 94.2 | 84.1-102.6 |
| 16-19 | 676 | 90.5 | 78.7-100.6 | 342 | 90.0 | 77.3-99.9 | 334 | 90.7 | 80.8-102.2 |
| 20-23 | 759 | 81.5 | 65.6-95.0 | 386 | 81.8 | 66.2-97.7 | 373 | 81.3 | 65.4-93.5 |
| 24-27 | 741 | 77.8 | 62.0-93.4 | 369 | 75.1 | 57.6-92.5 | 372 | 81.6 | 64.2-93.9 |
| 28-31 | 695 | 74.8 | 55.0-89.2 | 321 | 77.0 | 57.9-88.6 | 374 | 72.1 | 51.0-89.6 |
| 32-35 | 553 | 69.7 | 52.1-86.6 | 253 | 66.5 | 52.0-83.3 | 300 | 72.5 | 52.6-88.7 |
| 36-39 | 453 | 68.2 | 47.8-86.2 | 201 | 67.0 | 44.7-82.0 | 252 | 69.3 | 50.9-88.5 |
| 40-43 | 358 | 67.0 | 49.0-85.1 | 149 | 64.2 | 50.1-85.1 | 209 | 69.0 | 48.6-84.7 |
| 44-47 | 222 | 65.7 | 48.2-83.0 | 87 | 59.2 | 47.5-81.5 | 134 | 68.9 | 49.6-83.1 |
| 48-51 | 188 | 64.9 | 48.0-81.0 | 78 | 65.8 | 51.2-77.2 | 110 | 64.3 | 48.0-84.8 |
| 52-55 | 141 | 65.7 | 45.7-84.3 | 56 | 66.7 | 52.0-87.4 | 85 | 61.8 | 42.4-80.3 |
| 56-59 | 100 | 66.2 | 47.8-80.9 | 43 | 65.5 | 52.2-76.9 | 57 | 66.9 | 44.8-81.6 |
| 60+ | 155 | 61.9 | 44.4-79.1 | 68 | 62.7 | 47.9-76.1 | 87 | 59.2 | 40.2-79.3 |
| <16 | 2258 | 93.4 | 83.5-102.0 | 1089 | 93.6 | 83.4-101.5 | 1169 | 93.3 | 83.5-102.5 |
| ≥16 | 5040 | 76.4 | 56.2-91.3 | 2353 | 75.7 | 56.2-91.1 | 2687 | 76.8 | 56.1-91.3 |
| <18 | 2609 | 93.4 | 83.3-102.0 | 1268 | 93.6 | 83.0-101.6 | 1341 | 93.3 | 83.4-102.4 |
| ≥18 | 4689 | 74.7 | 54.6-89.9 | 2174 | 73.6 | 55.0-89.5 | 2515 | 75.4 | 54.3-90.3 |
| **Overall** | **7298*** | **83.4** | **64.2-96.4** | **3442** | **83.3** | **63.7-96.5** | **3856** | **83.5** | **64.6-96.2** |

*number with non-missing data.

## 1.15 Best* $FEV_1$% predicted (GLI equations) in patients aged six years and older who have not had a lung transplant
## N= 8659

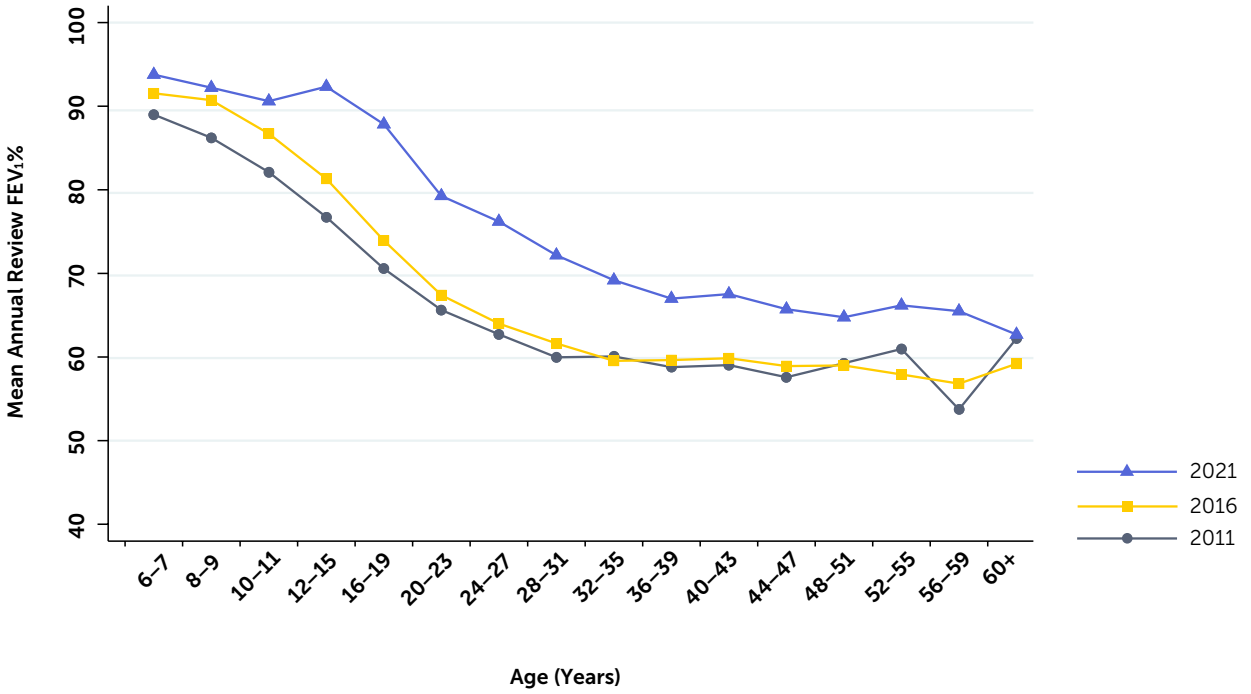



| | Overall | | | Female | | | Male | | |
|---|---|---|---|---|---|---|---|---|---|
| Age (yrs) | N | Median | IQR | N | Median | IQR | N | Median | IQR |
| 6-7 | 439 | 97.9 | 89.8-107.3 | 209 | 97.1 | 90.9-107.7 | 230 | 98.5 | 88.1-107.1 |
| 8-9 | 524 | 98.0 | 90.3-106.8 | 251 | 98.7 | 89.7-107.4 | 273 | 97.8 | 90.4-106.5 |
| 10-11 | 538 | 96.7 | 87.8-104.9 | 256 | 96.0 | 87.6-104.5 | 282 | 97.0 | 88.1-105.4 |
| 12-15 | 984 | 97.5 | 87.6-106.2 | 480 | 97.1 | 87.3-106.3 | 504 | 97.7 | 88.2-106.2 |
| 16-19 | 743 | 94.3 | 83.8-104.4 | 377 | 93.4 | 83.4-103.8 | 366 | 95.2 | 84.1-105.6 |
| 20-23 | 862 | 85.4 | 69.5-97.7 | 429 | 86.1 | 70.3-99.8 | 433 | 84.0 | 68.8-95.7 |
| 24-27 | 856 | 81.8 | 64.1-96.5 | 420 | 79.3 | 61.1-95.9 | 436 | 83.4 | 66.6-96.8 |
| 28-31 | 809 | 76.4 | 57.8-92.1 | 377 | 78.8 | 61.4-92.5 | 432 | 74.8 | 53.3-91.9 |
| 32-35 | 642 | 72.6 | 56.5-89.0 | 283 | 69.3 | 55.5-86.6 | 359 | 74.4 | 56.6-90.4 |
| 36-39 | 518 | 72.2 | 51.9-88.4 | 227 | 69.4 | 49.5-86.2 | 291 | 74.1 | 52.6-89.6 |
| 40-43 | 408 | 68.1 | 52.1-87.1 | 167 | 64.8 | 52.1-87.4 | 241 | 71.5 | 52.1-87.0 |
| 44-47 | 245 | 67.4 | 49.6-85.4 | 95 | 61.6 | 47.5-83.5 | 150 | 72.2 | 51.2-86.0 |
| 48-51 | 218 | 69.2 | 53.2-85.3 | 89 | 69.1 | 53.4-84.5 | 129 | 69.4 | 52.7-86.7 |
| 52-55 | 159 | 67.6 | 47.8-86.4 | 65 | 73.8 | 51.3-91.9 | 94 | 64.6 | 44.2-81.9 |
| 56-59 | 111 | 72.6 | 51.2-87.6 | 48 | 73.3 | 57.7-85.4 | 63 | 72.1 | 48.3-88.7 |
| 60+ | 164 | 68.4 | 47.5-84.6 | 75 | 70.2 | 51.2-84.8 | 89 | 63.0 | 43.2-84.3 |
| <16 | 2485 | 97.4 | 88.5-106.3 | 1196 | 97.1 | 88.2-106.4 | 1289 | 97.7 | 88.8-106.2 |
| ≥16 | 5735 | 79.5 | 59.2-94.3 | 2652 | 79.6 | 59.4-94.6 | 3083 | 79.3 | 58.9-94.1 |
| <18 | 2866 | 97.4 | 88.3-106.3 | 1390 | 97.1 | 88.0-106.4 | 1476 | 97.7 | 88.7-106.3 |
| ≥18 | 5354 | 77.7 | 57.9-92.8 | 2458 | 77.8 | 58.4-92.8 | 2896 | 77.6 | 57.6-92.8 |
| **Overall** | **8220**** | **86.9** | **67.7-99.7** | **3848** | **87.3** | **68.0-99.9** | **4372** | **86.7** | **67.5-99.6** |

* Where Best $FEV_1$% was missing or less than the FEV1% at annual review, annual review FEV1% was used instead.
** number with non-missing data.

## 1.16 Annual review $FEV_1$% predicted (GLI equations) over time in patients aged six years and older who have not had a lung transplant N= 8659 in 2021, N= 7977 in 2016, N= 6986 in 2011

As we learn more about CF and how to treat it, we hope to improve the outcomes of people with the condition. The chart below shows how $FEV_1$ in 2021 compares to Registry data from 2011 and 2016.



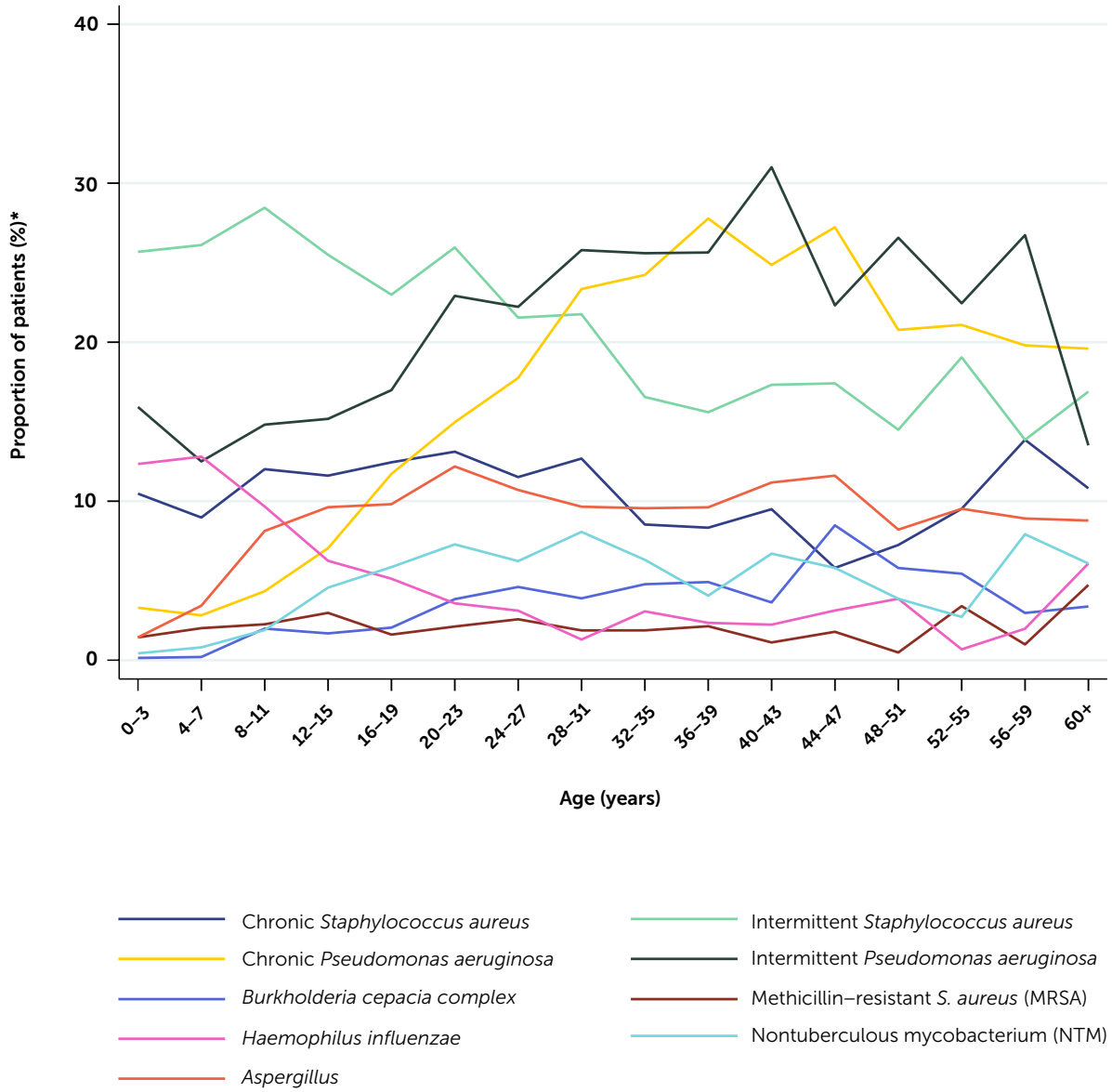

| | 2011 | | 2016 | | 2021 | | |
|---|---|---|---|---|---|---|---|
| Age (years) | n | $FEV_1$% : Mean (SD) | n | $FEV_1$% : Mean (SD) | n | $FEV_1$% : Mean (SD) | p-values (t-test) |
| 6-7 | 389 | 89.0 (15.2) | 500 | 91.5 (15.9) | 388 | 93.8 (15.3) | 0.035 |
| 8-9 | 372 | 86.2 (16.1) | 488 | 90.7 (15.7) | 478 | 92.2 (14.8) | 0.131 |
| 10-11 | 384 | 82.1 (16.1) | 440 | 86.7 (15.9) | 490 | 90.6 (15.0) | <0.001 |
| 12-15 | 920 | 76.7 (18.7) | 872 | 81.3 (17.2) | 902 | 92.3 (16.0) | <0.001 |
| 16-19 | 949 | 70.6 (21.8) | 918 | 73.9 (21.3) | 676 | 87.8 (18.9) | <0.001 |
| 20-23 | 948 | 65.6 (23.8) | 933 | 67.4 (23.0) | 759 | 79.3 (21.7) | <0.001 |
| 24-27 | 734 | 62.7 (23.6) | 881 | 64.0 (23.2) | 741 | 76.2 (22.4) | <0.001 |
| 28-31 | 589 | 60.0 (22.6) | 713 | 61.6 (23.8) | 695 | 72.2 (23.5) | <0.001 |
| 32-35 | 388 | 60.1 (22.7) | 564 | 59.5 (23.3) | 553 | 69.2 (23.3) | <0.001 |
| 36-39 | 249 | 58.8 (22.6) | 392 | 59.6 (23.4) | 453 | 67.0 (23.6) | <0.001 |
| 40-43 | 230 | 59.0 (22.8) | 256 | 59.9 (22.9) | 358 | 67.5 (23.7) | <0.001 |
| 44-47 | 165 | 57.6 (24.9) | 225 | 58.9 (22.5) | 222 | 65.7 (24.2) | 0.002 |
| 48-51 | 106 | 59.3 (22.9) | 155 | 59.0 (25.9) | 188 | 64.8 (22.7) | 0.029 |
| 52-55 | 53 | 61.0 (27.3) | 108 | 57.9 (24.2) | 141 | 66.2 (24.6) | 0.009 |
| 56-59 | 33 | 53.7 (21.8) | 59 | 56.8 (26.5) | 100 | 65.5 (22.1) | 0.028 |
| 60+ | 48 | 62.2 (24.4) | 100 | 59.2 (23.7) | 155 | 62.7 (23.3) | 0.245 |
| <16 | 2065 | 81.8 (17.9) | 2300 | 86.6 (16.9) | 2258 | 92.2 (15.4) | - |
| ≥16 | 4492 | 63.6 (23.4) | 5304 | 64.3 (23.7) | 5041 | 73.7 (23.6) | - |
| <18 | 2533 | 80.1 (18.7) | 2739 | 84.8 (17.9) | 2609 | 92.0 (15.7) | - |
| ≥18 | 4024 | 62.6 (23.5) | 4865 | 63.3 (23.7) | 4690 | 72.4 (23.6) | - |

** t-test comparing 2021 with 2016.

# 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.

## 1.17 Lung infections in 2021
## N=8913




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

## 1.17 Lung infections in 2021 (contd.)
## <16 years N=3878, ≥16 years N=6297

| | Paediatric Age Range (Years) | | | | Overall |
|---|---|---|---|---|---|
| | 0-3 | 4-7 | 8-11 | 12-15 | Paediatric (<16 years) |
| Number in age range | 718 | 1013 | 1127 | 1020 | 3878 |
| Number who had culture taken* | 697 | 992 | 1107 | 1008 | 3804 |
| Chronic *S. aureus* n (%) | 73 (10.5) | 89 (9.0) | 133 (12.0) | 117 (11.6) | 412 (10.8) |
| Intermittent *S. aureus* n (%) | 179 (25.7) | 259 (26.1) | 315 (28.5) | 257 (25.5) | 1010 (26.6) |
| Chronic *P. aeruginosa* n (%) | 23 (3.3) | 28 (2.8) | 48 (4.3) | 71 (7.0) | 170 (4.5) |
| Intermittent *P. aeruginosa* n (%) | 111 (15.9) | 124 (12.5) | 164 (14.8) | 153 (15.2) | 552 (14.5) |
| *B. cepacia* complex n (%) | <5 | <5 | 22 (2.0) | 17 (1.7) | 42 (1.1) |
| *B. cenocepacia* n (%) | <5 | <5 | 9 (0.8) | <5 | 13 (0.3) |
| *B. multivorans* n (%) | <5 | <5 | 7 (0.6) | 6 (0.6) | 14 (0.4) |
| *B. other cepacia* n (%) | <5 | <5 | <5 | <5 | 8 (0.2) |
| MRSA n (%) | 10 (1.4) | 20 (2.0) | 25 (2.3) | 30 (3.0) | 85 (2.2) |
| *H. influenza* n (%) | 86 (12.3) | 127 (12.8) | 107 (9.7) | 63 (6.3) | 383 (10.1) |
| NTM n (%) | <5 | -** | 21 (1.9) | 46 (4.6) | 78 (2.1) |
| *Aspergillus fumigatus* n (%) | 10 (1.4) | 34 (3.4) | 90 (8.1) | 97 (9.6) | 231 (6.1) |

Infections in this table reflect those grown in the 12 months prior to the 2021 annual review.
The UK CF Registry definition of 'chronic' is three or more isolates in the last 12 months.

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

** Redacted to adhere to statistical disclosure guidelines.

# Lung infections in 2021 (contd.)
## <16 years N=3878, ≥16 years N=6297

| | Adult Age Range (Years) | | | | | | Overall |
|---|---|---|---|---|---|---|---|
| | 16-19 | 20-23 | 24-27 | 28-31 | 32-35 | 36-39 | Adults (≥16 years) |
| Number in age range | 785 | 919 | 926 | 865 | 713 | 614 | 6297 |
| Number who had culture taken* | 683 | 755 | 738 | 694 | 586 | 468 | 5109 |
| Chronic *S. aureus* n (%) | 85 (12.4) | 99 (13.1) | 85 (11.5) | 88 (12.7) | 50 (8.5) | 39 (8.3) | 552 (10.8) |
| Intermittent *S. aureus* n (%) | 157 (23.0) | 196 (26.0) | 159 (21.5) | 151 (21.8) | 97 (16.6) | 73 (15.6) | 1031 (20.2) |
| Chronic *P. aeruginosa* n (%) | 80 (11.7) | 113 (15.0) | 131 (17.8) | 162 (23.3) | 142 (24.2) | 130 (27.8) | 1031 (20.2) |
| Intermittent *P. aeruginosa* n (%) | 116 (17.0) | 173 (22.9) | 164 (22.2) | 179 (25.8) | 150 (25.6) | 120 (25.6) | 1198 (23.4) |
| *B. cepacia* complex n (%) | 14 (2.0) | 29 (3.8) | 34 (4.6) | 27 (3.9) | 28 (4.8) | 23 (4.9) | 215 (4.2) |
| *B. cenocepacia* n (%) | <5 | -** | 9 (1.2) | 7 (1.0) | 9 (1.5) | 5 (1.1) | 58 (1.1) |
| *B. multivorans* n (%) | 6 (0.9) | 9 (1.2) | 15 (2.0) | 17 (2.4) | 17 (2.9) | 13 (2.8) | 105 (2.1) |
| *B. other cepacia* n (%) | <5 | 7 (0.9) | 8 (1.1) | <5 | <5 | <5 | 30 (0.6) |
| MRSA n (%) | 11 (1.6) | 16 (2.1) | 19 (2.6) | 13 (1.9) | 11 (1.9) | 10 (2.1) | 102 (2.0) |
| *H. influenza* n (%) | 35 (5.1) | 27 (3.6) | 23 (3.1) | 9 (1.3) | 18 (3.1) | 11 (2.4) | 158 (3.1) |
| NTM n (%) | 40 (5.9) | 55 (7.3) | 46 (6.2) | 56 (8.1) | 37 (6.3) | 19 (4.1) | 319 (6.2) |
| *Aspergillus fumigatus* n (%) | 67 (9.8) | 92 (12.2) | 79 (10.7) | 67 (9.7) | 56 (9.6) | 45 (9.6) | 525 (10.3) |

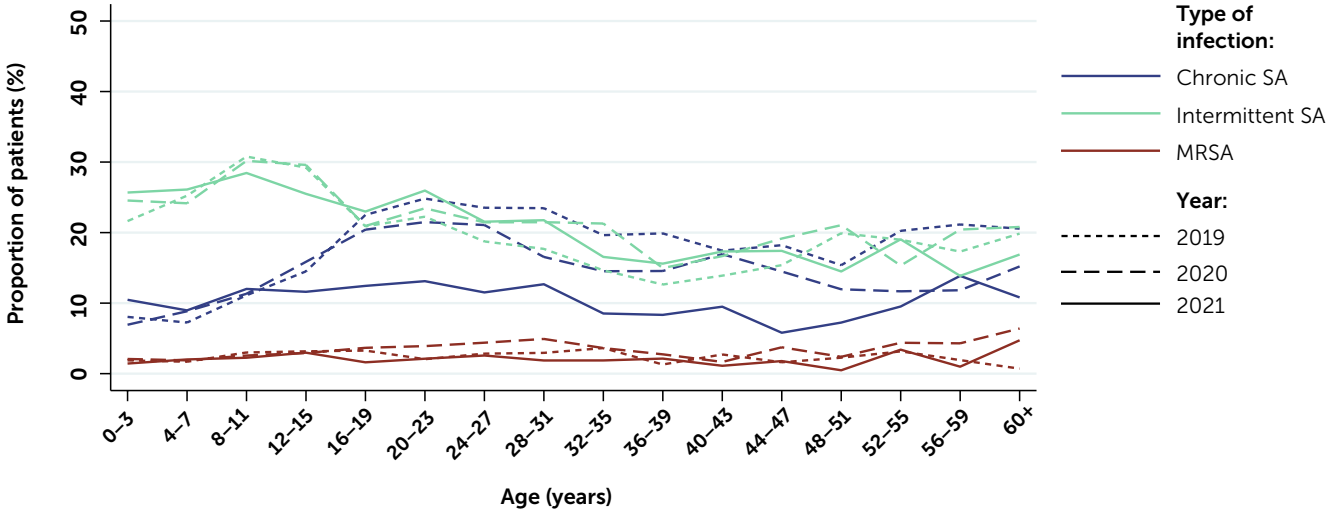
| | Adult Age Range (Years) | | | | | | Overall |
|---|---|---|---|---|---|---|---|
| | 40-43 | 44-47 | 48-51 | 52-55 | 56-59 | 60+ | Adults (≥16 years) |
| Number in age range | 451 | 280 | 261 | 179 | 123 | 181 | 6297 |
| Number who had culture taken* | 358 | 224 | 207 | 147 | 101 | 148 | 5109 |
| Chronic *S. aureus* n (%) | 34 (9.5) | 13 (5.8) | 15 (7.2) | 14 (9.5) | 14 (13.9) | 16 (10.8) | 552 (10.8) |
| Intermittent *S. aureus* n (%) | 62 (17.3) | 39 (17.4) | 30 (14.5) | 28 (19.0) | 14 (13.9) | 25 (16.9) | 1031 (20.2) |
| Chronic *P. aeruginosa* n (%) | 89 (24.9) | 61 (27.2) | 43 (20.8) | 31 (21.1) | 20 (19.8) | 29 (19.6) | 1031 (20.2) |
| Intermittent *P. aeruginosa* n (%) | 111 (31.0) | 50 (22.3) | 55 (26.6) | 33 (22.4) | 27 (26.7) | 20 (13.5) | 1198 (23.4) |
| *B. cepacia* complex n (%) | 13 (3.6) | 19 (8.5) | 12 (5.8) | 8 (5.4) | <5 | 5 (3.4) | 215 (4.2) |
| *B. cenocepacia* n (%) | 6 (1.7) | <5 | <5 | <5 | <5 | <5 | 58 (1.1) |
| *B. multivorans* n (%) | 7 (2.0) | 9 (4.0) | 7 (3.4) | <5 | <5 | <5 | 105 (2.1) |
| *B. other cepacia* n (%) | <5 | <5 | <5 | <5 | <5 | <5 | 30 (0.6) |
| MRSA n (%) | <5 | <5 | <5 | 5 (3.4) | <5 | 7 (4.7) | 102 (2.0) |
| *H. influenza* n (%) | 8 (2.2) | 7 (3.1) | 8 (3.9) | <5 | <5 | 9 (6.1) | 158 (3.1) |
| NTM n (%) | 24 (6.7) | 13 (5.8) | 8 (3.9) | <5 | -** | 9 (6.1) | 319 (6.2) |
| *Aspergillus fumigatus* n (%) | 40 (11.2) | 26 (11.6) | 17 (8.2) | 14 (9.5) | 9 (8.9) | 13 (8.8) | 525 (10.3) |

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

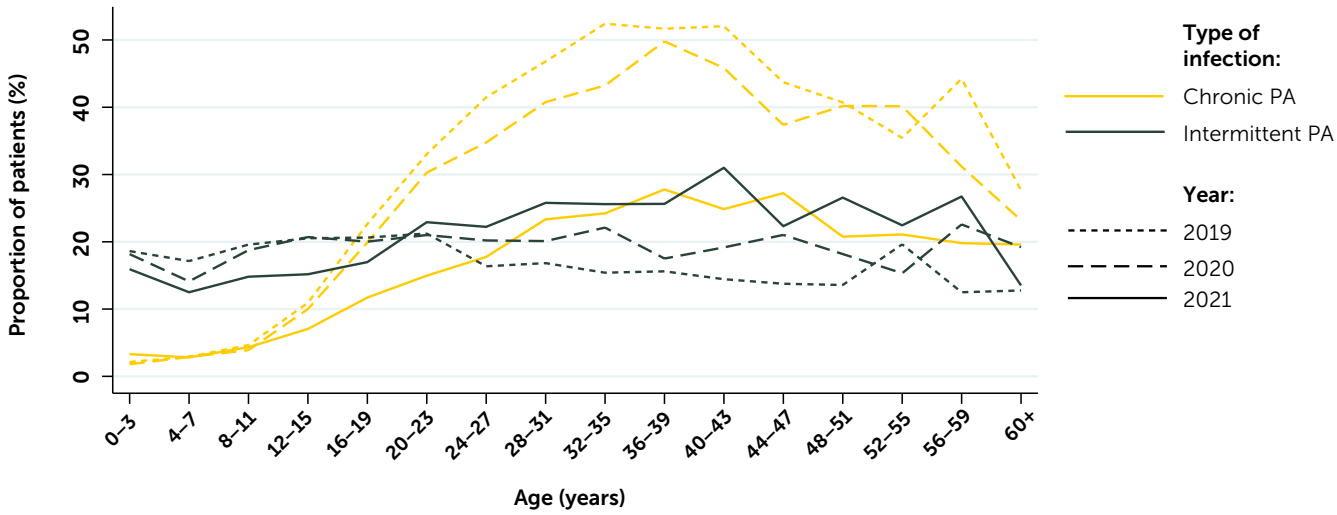
** Redacted to adhere to statistical disclosure guidelines

# 1.18 Lung infections 2019-2021

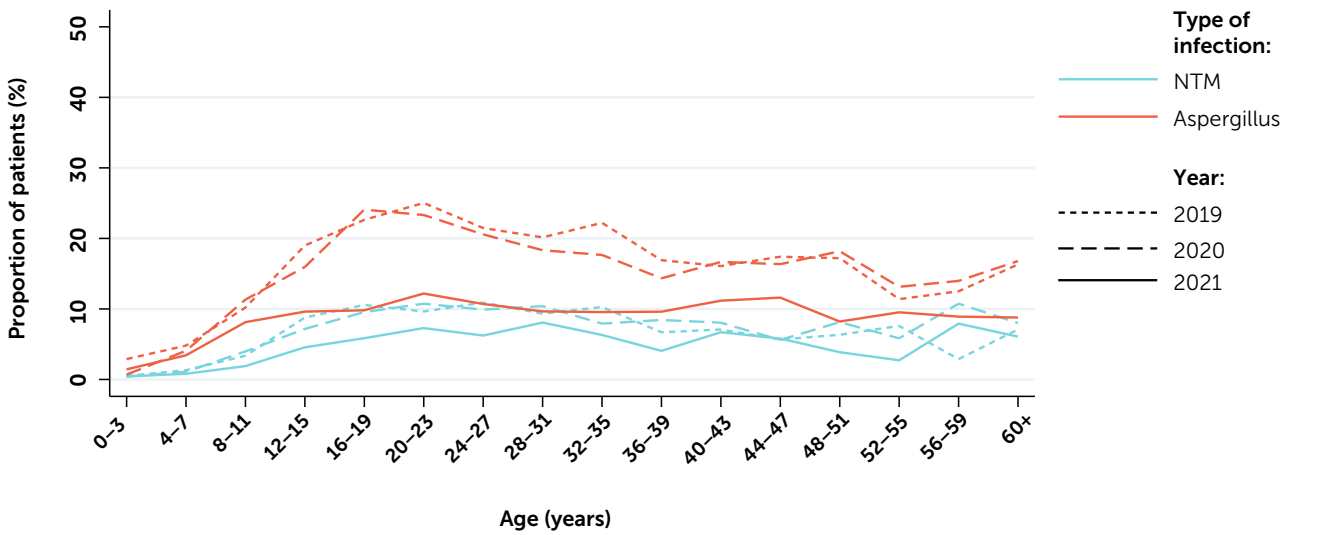
*Staphylococcus aureus* infections




*Pseudomonas aeruginosa* infections




Other infections



## 1.19 Respiratory culture sample type

| Overall | 2019 | 2020 | 2021 |
|---|---|---|---|
| Number of people with an annual review (N) | 10070 | 9922 | 10175 |
| **Number of people with at least 1 sample of any type taken N(%)*** | 9847 (97.8) | 9368 (94.4) | 8921 (87.7) |
| **Sample type**** | | | |
| Sputum; N(%) | 6865 (69.7) | 6250 (66.7) | 5196 (58.2) |
| Cough; N(%) | 6198 (62.9) | 6021 (64.3) | 6048 (67.8) |
| Bronchoalveolar lavage; N(%) | 485 (4.9) | 528 (5.6) | 224 (2.5) |
| | | | |
| **Age <16 years** | **2019** | **2020** | **2021** |
| Number of people with an annual review (N) | 3966 | 3910 | 3878 |
| **Number of people with at least 1 sample of any type taken N(%)*** | 3937 (99.3) | 3851 (98.5) | 3808 (98.1) |
| **Sample type**** | | | |
| Sputum; N(%) | 1668 (42.4) | 1437 (37.3) | 1125 (29.5) |
| Cough; N(%) | 3767 (95.7) | 3696 (96.0) | 3704 (97.3) |
| Bronchoalveolar lavage; N(%) | 343 (8.7) | 254 (6.6) | 172 (4.5) |
| | | | |
| **Age>=16 years** | **2019** | **2020** | **2021** |
| Number of people with an annual review (N) | 6104 | 6012 | 6297 |
| **Number of people with at least 1 sample of any type taken N(%)*** | 5910 (96.8) | 5517 (98.1) | 5113 (81.2) |
| **Sample type**** | | | |
| Sputum; N(%) | 5197 (87.9) | 4813 (87.2) | 4071 (79.6) |
| Cough; N(%) | 2431 (41.1) | 2325 (42.1) | 2344 (45.8) |
| Bronchoalveolar lavage; N(%) | 142 (2.4) | 274 (5.0) | 52 (1.0) |

* % is of those people with an Annual Review.

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

## 1.20 Non-tuberculous 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.

| | 2019 | 2020 | 2021 |
|---|---|---|---|
| **Number with annual review** | **(n=10070)** | **(n=9922)** | **(n=10175)** |
| NTM Prevalence; n (%) | 674 (6.7) | 620 (6.2*) | 397 (3.9) |
| On NTM treatment in the given year; n (% of NTM prevalence in given year) | 362 (53.7) | 326 (52.6) | 231 (58.1) |
| NTM Incidence[1] | 279 (3.0) | 226 (2.5) | 154 (1.7) |
| *M. abscessus* prevalence | 382 (3.8) | 361 (3.6*) | 216 (2.1) |
| *M. abscessus* incidence[2] | 126 (1.3) | 103 (1.1) | 58 (0.6) |

* correction for 2020 data

[1] Proportion based on the number of patients with non-positive NTM tests in the previous two data years

[2] Proportion based on the number of patients with non-positive *M.abscessus* tests in the previous two data years

## 1.21 COVID-19* infection in 2021

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

| | | COVID-19 Management | | | Outcomes |
|---|---|---|---|---|---|
| | Total | Symptomatic | IV antibiotics | Oral antibiotics | Hospitalised |
| **Overall; n(%)** | | | | | |
| All cases | 814 | 570 (70.0) | 41 (5.0) | 249 (30.6) | 62 (7.6) |
| **Sex; n(%)** | | | | | |
| Female | 422 (51.8) | 305 (72.3) | 26 (6.2) | 134 (31.8) | 35 (8.3) |
| Male | 392 (48.2) | 265 (67.6) | 15 (3.8) | 115 (29.3) | 27 (6.9) |
| **Ethnicity; n(%)** | | | | | |
| White | 765 (94.0) | 534 (69.8) | 36 (4.7) | 232 (30.3) | 51 (6.7) |
| Non-White | 29 (3.6) | 20 (69.0) | <5 | 9 (31.0) | 9 (31.0) |
| Unknown | 20 (2.5) | 16 (80.0) | <5 | 8 (40.0) | <5 |
| **Age; n(%)** | | | | | |
| Under 16 | 255 (31.3) | 161 (63.1) | <5 | 56 (22.0) | 10 (3.9) |
| >= 16 | 559 (68.7) | 409 (73.2) | 37 (6.6) | 193 (34.5) | 52 (9.3) |
| **Transplants; n(%)** | | | | | |
| No | 774 (95.1) | 540 (69.8) | 25 (3.2) | 239 (30.9) | 42 (5.4) |
| Yes | 40 (4.9) | 30 (75.0) | 16 (40.0) | 10 (25.0) | 20 (50.0) |
| ****BestFEV; n(%)** | | | | | |
| <40 | 38 (4.7) | 29 (76.3) | 8 (21.1) | 18 (47.4) | 10 (26.3) |
| 40-70 | 180 (22.1) | 123 (68.3) | 18 (10.0) | 53 (29.4) | 23 (12.8) |
| >70 | 596 (73.2) | 418 (70.1) | 15 (2.5) | 178 (29.9) | 29 (4.9) |

Of the 62 patients hospitalised after a positive test for COVID-19, 17 of them were also given oxygen.

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

**Patients who had a lung transplant were excluded

# Complications

## 1.22 Complications in 2021

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

| | Overall (N=10175) n (%) | <16 years (N=3878) n(%) | ≥16 years (N=6297) n(%) |
|---|---|---|---|
| **Respiratory related** | | | |
| Nasal polyps requiring surgery | 445 (4.4) | 129 (3.3) | 316 (5.0) |
| Sinus disease | 714 (7.0) | 53 (1.4) | 661 (10.5) |
| Asthma | 738 (7.3) | 183 (4.7) | 555 (8.8) |
| ABPA | 605 (5.9) | 122 (3.1) | 483 (7.7) |
| Any haemoptysis | -* | <5 | 188 (3.0) |
| Massive haemoptysis | 12 (0.1) | 0 | 12 (0.2) |
| Pneumothorax requiring chest tube | 12 (0.1) | 0 | 12 (0.2) |
| **Pancreas and hepatobiliary disease** | | | |
| Raised liver enzymes | 1243 (12.2) | 373 (9.6) | 870 (13.8) |
| Liver disease | 1553 (15.3) | 361 (9.3) | 1192 (18.9) |
| Cirrhosis with no portal hypertension | 96 (0.9) | 19 (0.5) | 77 (1.2) |
| Cirrhosis with portal hypertension | 139 (1.4) | 29 (0.7) | 110 (1.7) |
| Gall bladder disease requiring surgery | 147 (1.4) | 29 (0.7) | 118 (1.9) |
| Pancreatitis | 73 (0.7) | 9 (0.2) | 64 (1.0) |
| **Upper gastrointestinal (GI)** | | | |
| Gastro-oesphageal reflux disease (GORD) | 1764 (17.3) | 252 (6.5) | 1512 (24.0) |
| Peptic ulcer | 0 (0.0) | 0 | 0 |
| GI bleed (varices as source) | 16 (0.2) | 7 (0.2) | 9 (0.1) |
| GI bleed (non varices as source) | 17 (0.2) | 8 (0.2) | 9 (0.1) |
| **Lower gastrointestinal** | | | |
| Intestinal obstruction | 34 (0.3) | 17 (0.4) | 17 (0.3) |
| DIOS | 493 (4.8) | 102 (2.6) | 391 (6.2) |
| Fibrosing colonopathy / colonic stricture | -* | 0 | <5 |
| Rectal prolapse | 12 (0.1) | 8 (0.2) | <5 |
| **Renal** | | | |
| Kidney stones | 144 (1.4) | 10 (0.3) | 134 (2.1) |
| Renal failure | -* | <5 | 81 (1.3) |
| **Musculoskeletal** | | | |
| Arthritis | -* | <5 | 111 (1.8) |
| Arthropathy | 230 (2.3) | 7 (0.2) | 223 (3.5) |
| Bone fracture | 37 (0.4) | 16 (0.4) | 21 (0.3) |
| Osteopenia | 925 (9.1) | 7 (0.2) | 918 (14.6) |
| Osteoporosis | 408 (4.0) | 7 (0.2) | 401 (6.4) |
| **Other** | | | |
| Cancer confirmed by histology | -* | <5 | 28 (0.4) |
| Port inserted or replaced | 210 (2.1) | 64 (1.7) | 146 (2.3) |
| Depression | 522 (5.1) | 15 (0.4) | 507 (8.1) |
| Hearing loss | 358 (3.5) | 34 (0.9) | 324 (5.1) |
| Hypertension | -* | <5 | 197 (3.1) |

* Redacted to adhere to statistical disclosure guidelines.

## 1.23 Incidence of complications

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

| | 2020 | | | 2021 | | |
|---|---|---|---|---|---|---|
| | Overall (n=9922) | <16 years (n=3910) | ≥16 years (n=6012) | Overall (n=10175) | <16 years (n=3878) | ≥16 years (n=6297) |
| ABPA | 216 (2.2) | 87 (2.2) | 129 (2.2) | 153 (1.5) | 71 (1.8) | 82 (1.3) |
| Cirrhosis - no portal hypertension | 40 (0.4) | 11 (0.3) | 29 (0.5) | 57 (0.6) | 19 (0.5) | 38 (0.6) |
| Cirrhosis - with portal hypertension | 46 (0.5) | 11 (0.3) | 35 (0.6) | 39 (0.4) | 11 (0.3) | 28 (0.5) |
| Cancer confirmed by histology | * | <5 | 17 (0.3) | * | <5 | 15 (0.2) |

## 1.24 CF-related diabetes
## N= 7887

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=7887) | 10-15 years (N=1590) | ≥16 years (N=6297) |
|---|---|---|---|
| **On CFRD treatment; n (%)** | 2349 (29.8) | 132 (8.3) | 2217 (35.2) |
| Of those on treatment | | | |
| Insulin[1]; n (%) | 1999 (85.1) | 126 (95.5) | 1873 (84.5) |
| **CFRD Screening ; n(%)** | | | |
| Yes | 2993 (37.9) | 1041 (65.5) | 1952 (31.0) |
| **Screening Type** | | | |
| Continous glucose monitoring[2]; n (%) | 1069 (35.7) | 265 (25.5) | 804 (41.2) |
| Oral glucose tolerance test[2]; n (%) | 1048 (35.0) | 465 (44.7) | 583 (29.9) |
| Not screened (other) | 2258 (28.6) | 122 (7.7) | 2136 (33.9) |
| Not screened (known CFRD) | 2470 (31.3) | 349 (21.9) | 2121 (33.7) |
| Unknown | 162 (2.1) | 77 (4.8) | 85 (1.3) |

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

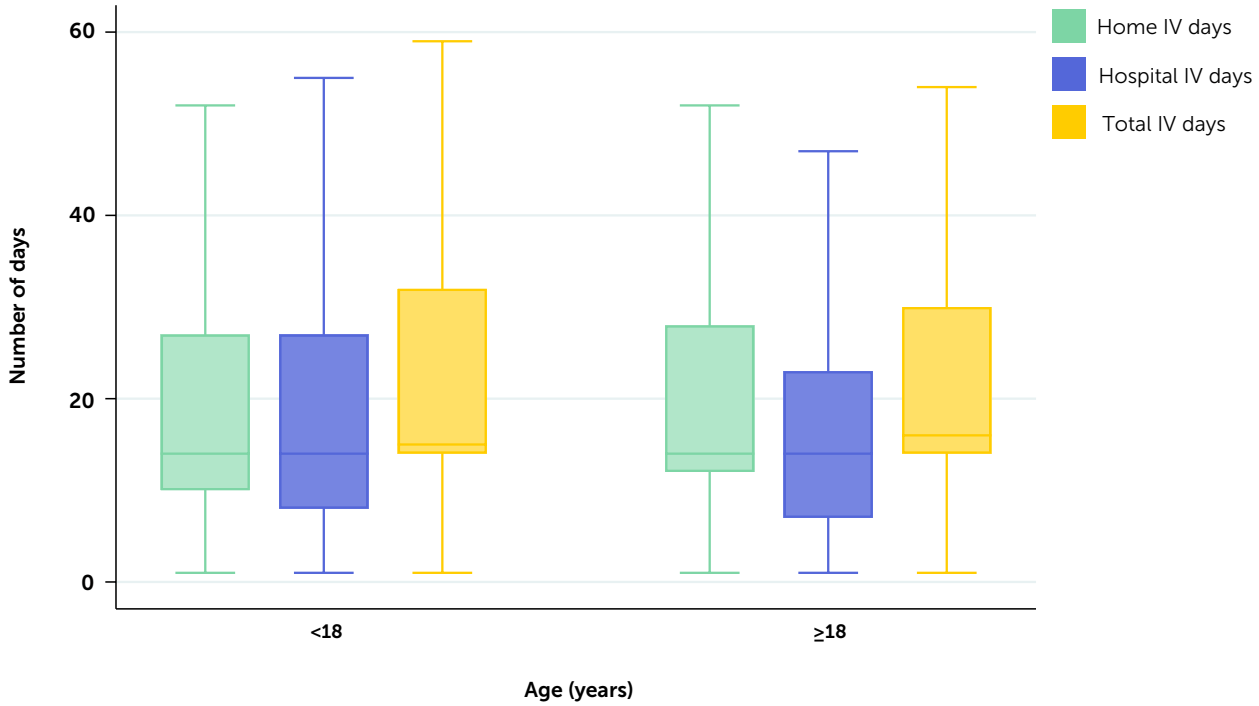
# Antibiotics

## 1.25 Intravenous (IV) antibiotics
## N=10175

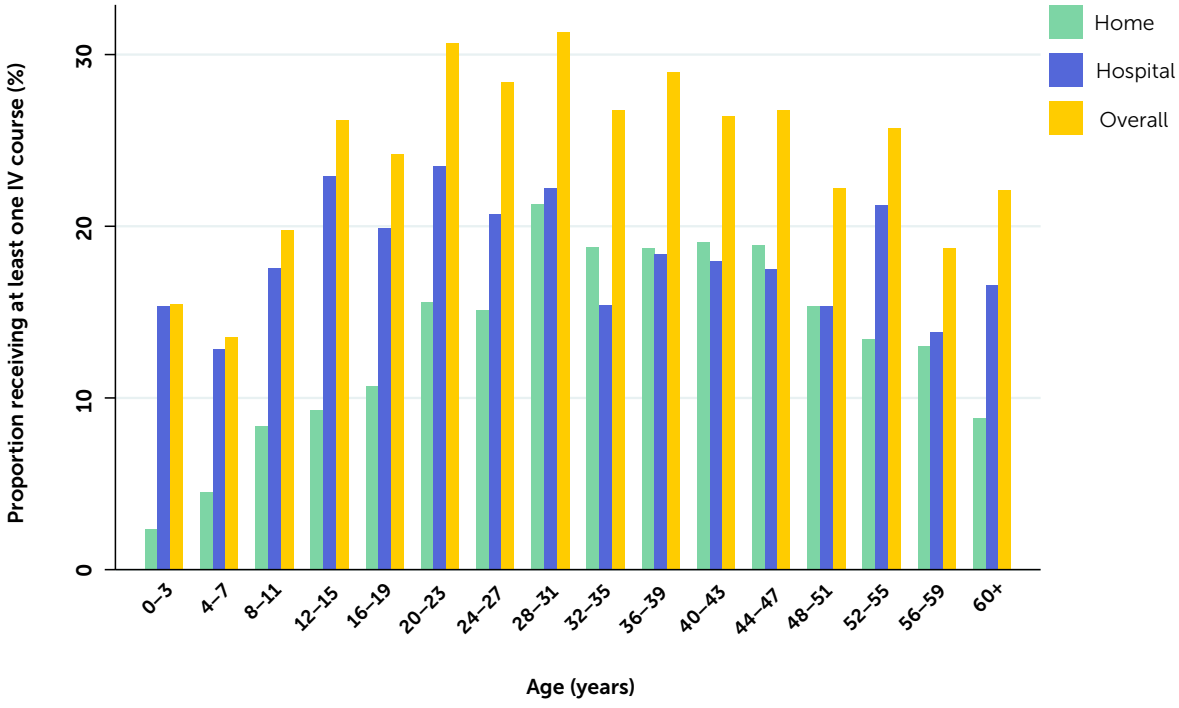
When someone with CF becomes unwell with an infection, they might be prescribed intravenous (IV) antibiotics. IV antibiotics are given to the patient through their veins. This treatment can take a number of days and may take place as a hospital inpatient, or at home.

| | | Home | | Hospital | | Total | |
|---|---|---|---|---|---|---|---|
| Age | N | Patients n (%) | Median days (IQR) | Patients n (%) | Median days (IQR) | Patients n (%) | Median days (IQR) |
| 0-3 | 718 | 17 (2.4) | 7 (5-9) | 110 (15.3) | 14 (7-19) | 111 (15.5) | 14 (8-22) |
| 4-7 | 1013 | 46 (4.5) | 13 (9-18) | 130 (12.8) | 14 (7-27) | 137 (13.5) | 14 (13-30) |
| 8-11 | 1127 | 94 (8.3) | 14 (10-27) | 198 (17.6) | 14 (8-28) | 223 (19.8) | 19 (14-38) |
| 12-15 | 1020 | 95 (9.3) | 14 (12-29) | 234 (22.9) | 14 (9-28) | 267 (26.2) | 21 (14-40) |
| 16-19 | 785 | 84 (10.7) | 14 (11-26) | 156 (19.9) | 13 (8-23) | 190 (24.2) | 14 (12-28) |
| 20-23 | 919 | 143 (15.6) | 14 (10-28) | 216 (23.5) | 14 (7-25) | 282 (30.7) | 15 (13-30) |
| 24-27 | 926 | 140 (15.1) | 14 (11-26) | 192 (20.7) | 14 (7-22) | 263 (28.4) | 16 (14-28) |
| 28-31 | 865 | 184 (21.3) | 14 (12-25) | 192 (22.2) | 13 (7-23) | 271 (31.3) | 17 (14-35) |
| 32-35 | 713 | 134 (18.8) | 14 (13-28) | 110 (15.4) | 13 (7-23) | 191 (26.8) | 16 (14-32) |
| 36-39 | 614 | 115 (18.7) | 15 (14-28) | 113 (18.4) | 14 (8-25) | 178 (29.0) | 20 (14-32) |
| 40-43 | 451 | 86 (19.1) | 14 (12-26) | 81 (18.0) | 12 (5-19) | 119 (26.4) | 17 (14-31) |
| 44-47 | 280 | 53 (18.9) | 14 (14-28) | 49 (17.5) | 14 (6-24) | 75 (26.8) | 16 (14-39) |
| 48-51 | 261 | 40 (15.3) | 16 (11-28) | 40 (15.3) | 10 (5-19) | 58 (22.2) | 18 (14-28) |
| 52-55 | 179 | 24 (13.4) | 14 (13-25) | 38 (21.2) | 14 (7-23) | 46 (25.7) | 21 (14-30) |
| 56-59 | 123 | 16 (13.0) | 14 (12-15) | 17 (13.8) | 10 (8-20) | 23 (18.7) | 14 (14-28) |
| 60+ | 181 | 16 (8.8) | 14 (10-14) | 30 (16.6) | 14 (8-20) | 40 (22.1) | 14 (11-22) |
| <16 | 3878 | 252 (6.5) | 14 (10-27) | 672 (17.3) | 14 (8-28) | 738 (19.0) | 15 (14-32) |
| ≥16 | 6297 | 1035 (16.4) | 14 (12-28) | 1234 (19.6) | 14 (7-23) | 1736 (27.6) | 16 (14-30) |
| <18 | 4283 | 298 (7.0) | 14 (10-27) | 754 (17.6) | 14 (8-27) | 841 (19.6) | 15 (14-32) |
| ≥18 | 5892 | 989 (16.8) | 14 (12-28) | 1152 (19.6) | 14 (7-23) | 1633 (27.7) | 16 (14-30) |
| **Overall** | **10175** | **1287 (12.6)** | **14 (11-28)** | **1906 (18.7)** | **14 (7-24)** | **2474 (24.3)** | **15 (14-31)** |

This box plot graph illustrates the spread of the number of days on IV antibiotics in the UK CF population, stratified by age. A guide on how to correctly interpret this box plot graph can be found on page 51.




The bar graph below summarises the proportion of people receiving at least one course of IV antibiotics across different age groups within the UK CF population. Overall, the proportion of patients receiving at least one IV course at home was 12.6% and in hospital was 18.7%. The proportion receiving any IVs was 24.3%.



## 1.26 Inhaled antibiotic use among people with chronic *Pseudomonas aeruginosa*

| | 2011 | | | 2016 | | | 2021 | | |
|---|---|---|---|---|---|---|---|---|---|
| | Overall | <16 years | ≥16 years | Overall | <16 years | ≥16 years | Overall | <16 years | ≥16 years |
| Patients with chronic *P. aeruginosa* | 3138 | 407 | 2731 | 2831 | 246 | 2585 | 1201 | 170 | 1031 |
| Tobramycin solution; n (%) | 922 (29.4) | 103 (25.3) | 819 (30.0) | 655 (23.1) | 79 (32.1) | 576 (22.3) | 360 (30.0) | 73 (42.9) | 287 (27.8) |
| Other aminoglycoside; n (%) | 63 (2.0) | 14 (3.4) | 49 (1.8) | 74 (2.6) | 12 (4.9) | 62 (2.4) | 39 (3.2) | <5 | -* |
| Colistin; n (%) | 1378 (43.9) | 231 (56.8) | 1147 (42.0) | 791 (27.9) | 117 (47.6) | 674 (26.1) | 406 (33.8) | 74 (43.5) | 332 (32.2) |
| Promixin; n (%) | 858 (27.3) | 112 (27.5) | 746 (27.3) | 888 (31.4) | 104 (42.3) | 784 (30.3) | 329 (27.4) | 70 (41.2) | 259 (25.1) |
| Aztreonam; n (%) | - | - | - | 551 (19.5) | 13 (5.3) | 538 (20.8) | 282 (23.5) | 7 (4.1) | 275 (26.7) |
| Colistimethate (DPI); n (%) | - | - | - | 513 (18.1) | 29 (11.8) | 484 (18.7) | 216 (18.0) | 14 (8.2) | 202 (19.6) |
| Tobramycin Inhalation Powder; n (%) | - | - | - | 865 (30.6) | 34 (13.8) | 831 (32.1) | 217 (18.1) | 8 (4.7) | 209 (20.3) |
| Levofloxacin ;n(%) | - | - | - | 0 | 0 | 0 | 27 (2.2) | 0 | 27 (2.6) |
| At least one of the above; n (%) | 2589 (82.5) | 366 (89.9) | 2223 (81.4) | 2519 (89.0) | 230 (93.5) | 2289 (88.5) | 1044 (86.9) | 152 (89.4) | 892 (86.5) |

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.

## 1.27 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*.

| | | Number of patients on azithromycin; n | Patients with chronic *P. aeruginosa*; n (%) | Patients without chronic *P. aeruginosa*; n (%) |
|---|---|---|---|---|
| **2011** | **Overall** | 3646 | 2109 (57.8) | 1537 (42.2) |
| | **0-3 years** | 38 | 5 (13.2) | 33(86.8) |
| | **4-15 years** | 717 | 190 (26.5) | 527 (73.5) |
| | **≥ 16 years** | 2891 | 1914 (66.2) | 977 (33.8) |
| **2016** | **Overall** | 3833 | 1859 (48.5) | 1974(51.5) |
| | **0-3 years** | 32 | 5 (15.6) | 27 (84.4) |
| | **4-15 years** | 642 | 974(14.6) | 548(85.4) |
| | **≥ 16 years** | 3159 | 1760(55.7) | 1399 (44.3) |
| **2021** | **Overall** | 4160 | 759 (18.2) | 3401 (81.8) |
| | **0-3 years** | 39 | <5 | -* |
| | **4-15 years** | 599 | 63 (10.5) | 536 (89.5) |
| | **≥ 16 years** | 3522 | 692 (19.6) | 2830 (80.4) |

* Redacted to adhere to statistical disclosure guidelines.

## 1.28 Prophylactic flucloxacillin use

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

| Age | Total patients | Patients on prophylactic flucloxacillin; n (%) |
|---|---|---|
| 0-3 | 718 | 360 (50.1) |
| 4-7 | 1013 | 346 (34.2) |
| 8-11 | 1127 | 291 (25.8) |
| 12-15 | 1020 | 242 (23.7) |
| 16-19 | 785 | 178 (22.7) |
| 20-23 | 919 | 238 (25.9) |
| 24-27 | 926 | 94 (10.2) |
| 28-31 | 865 | 65 (7.5) |
| 32-35 | 713 | 37 (5.2) |
| 36-39 | 614 | 32 (5.2) |
| 40-43 | 451 | 26 (5.8) |
| 44-47 | 280 | 14 (5.0) |
| 48-51 | 261 | 14 (5.4) |
| 52-55 | 179 | 10 (5.6) |
| 56-59 | 123 | <5 |
| 60+ | 181 | -* |
| <16 years | 3878 | 1239 (31.9) |
| ≥16 years | 6297 | 720 (11.4) |
| <18 years | 4283 | 1313 (30.7) |
| ≥18 years | 5892 | 646 (11.0) |
| **Overall** | **10175** | **1959 (19.3)** |

* Redacted to adhere to statistical disclosure guidelines.

# Bronchodilators & Corticosteroids

## 1.29 Inhaled bronchodilators & corticosteroids

| Age | Total patients | Patients on inhaled bronchodilators; n(%) | Patients on inhaled corticosteroids; n(%) | Patients on inhaled combination corticosteroids/ bronchodilators; n(%) |
|---|---|---|---|---|
| 0-3 | 718 | 120 (16.7) | 43 (6.0) | <5 |
| 4-7 | 1013 | 341 (33.7) | 151 (14.9) | -* |
| 8-11 | 1127 | 563 (50.0) | 222 (19.7) | 155 (13.8) |
| 12-15 | 1020 | 596 (58.4) | 183 (17.9) | 246 (24.1) |
| 16-19 | 785 | 533 (67.9) | 150 (19.1) | 271 (34.5) |
| 20-23 | 919 | 690 (75.1) | 205 (22.3) | 333 (36.2) |
| 24-27 | 926 | 647 (69.9) | 188 (20.3) | 310 (33.5) |
| 28-31 | 865 | 631 (72.9) | 173 (20.0) | 379 (43.8) |
| 32-35 | 713 | 529 (74.2) | 148 (20.8) | 315 (44.2) |
| 36-39 | 614 | 443 (72.1) | 123 (20.0) | 275 (44.8) |
| 40-43 | 451 | 322 (71.4) | 98 (21.7) | 204 (45.2) |
| 44-47 | 280 | 197 (70.4) | 53 (18.9) | 133 (47.5) |
| 48-51 | 261 | 177 (67.8) | 57 (21.8) | 115 (44.1) |
| 52-55 | 179 | 121 (67.6) | 40 (22.3) | 77 (43.0) |
| 56-59 | 123 | 87 (70.7) | 22 (17.9) | 47 (38.2) |
| 60+ | 181 | 130 (71.8) | 41 (22.7) | 76 (42.0) |
| <16 years | 3878 | 1620 (41.8) | 599 (15.4) | 428 (11.0) |
| ≥16 years | 6297 | 4507 (71.6) | 1298 (20.6) | 2535 (40.3) |
| <18 years | 4283 | 1890 (44.1) | 657 (15.3) | 555 (13.0) |
| ≥18 years | 5892 | 4237 (71.9) | 1240 (21.0) | 2408 (40.9) |
| **Overall** | **10175** | **6127 (60.2)** | **1897 (18.6)** | **2963 (29.1)** |

* Redacted to adhere to statistical disclosure guidelines.

# Muco-active therapies

## 1.30 Mannitol

| | 2016 | | 2021 | |
|---|---|---|---|---|
| Age | Total patients | Patients on Mannitol; n (%) | Total patients | Patients on Mannitol; n (%) |
| 0-3 | 844 | 0 | 718 | 0 |
| 4-7 | 1119 | 0 | 1013 | 0 |
| 8-11 | 975 | 0 | 1127 | 0 |
| 12-15 | 906 | <5 | 1020 | <5 |
| 16-19 | 943 | 23 (2.4) | 785 | 9 (1.1) |
| 20-23 | 998 | 59 (5.9) | 919 | 33 (3.6) |
| 24-27 | 965 | 70 (7.3) | 926 | 44 (4.8) |
| 28-31 | 802 | 57 (7.1) | 865 | 54 (6.2) |
| 32-35 | 648 | 42 (6.5) | 713 | 55 (7.7) |
| 36-39 | 443 | 28 (6.3) | 614 | 32 (5.2) |
| 40-43 | 303 | 17 (5.6) | 451 | 27 (6.0) |
| 44-47 | 267 | 16 (6.0) | 280 | 23 (8.2) |
| 48-51 | 181 | <5 | 261 | 10 (3.8) |
| 52-55 | 125 | 5 (4.0) | 179 | 5 (2.8) |
| 56-59 | 67 | <5 | 123 | <5 |
| 60+ | 109 | <5 | 181 | <5 |
| <16 years | 3844 | <5 | 3878 | <5 |
| ≥16 years | 5851 | 326 (5.6) | 6297 | 298 (4.7) |
| <18 years | 4292 | 7 (0.2) | 4283 | 5 (0.1) |
| ≥18 years | 5403 | 322 (6.0) | 5892 | 296 (5.0) |
| **Overall** | **9695** | **329 (3.4)** | **10175** | **301 (3.0)** |

## 1.31 DNase

| | 2011 | | 2016 | | 2021 | |
|---|---|---|---|---|---|---|
| Age | Total patients | Patients on DNase; n (%) | Total patients | Patients on DNase; n (%) | Total patients | Patients on DNase; n (%) |
| 0-3 | 964 | 97 (10.1) | 844 | 126 (14.9) | 718 | 124 (17.3) |
| 4-7 | 932 | 280 (30.0) | 1119 | 539 (48.2) | 1013 | 538 (53.1) |
| 8-11 | 835 | 395 (47.3) | 975 | 717 (73.5) | 1127 | 897 (79.6) |
| 12-15 | 1015 | 587 (57.8) | 906 | 719 (79.4) | 1020 | 863 (84.6) |
| 16-19 | 1016 | 575 (56.6) | 943 | 728 (77.2) | 785 | 683 (87.0) |
| 20-23 | 1003 | 576 (57.4) | 998 | 696 (69.7) | 919 | 823 (89.6) |
| 24-27 | 803 | 437 (54.4) | 965 | 623 (64.6) | 926 | 745 (80.5) |
| 28-31 | 642 | 358 (55.8) | 802 | 508 (63.3) | 865 | 620 (71.7) |
| 32-35 | 433 | 211 (48.7) | 648 | 402 (62.0) | 713 | 492 (69.0) |
| 36-39 | 293 | 120 (41.0) | 443 | 261 (58.9) | 614 | 395 (64.3) |
| 40-43 | 278 | 114 (41.0) | 303 | 162 (53.5) | 451 | 274 (60.8) |
| 44-47 | 192 | 86 (44.8) | 267 | 144 (53.9) | 280 | 175 (62.5) |
| 48-51 | 122 | 53 (43.4) | 181 | 94 (51.9) | 261 | 139 (53.3) |
| 52-55 | 61 | 24 (39.3) | 125 | 73 (58.4) | 179 | 101 (56.4) |
| 56-59 | 38 | 11 (28.9) | 67 | 34 (50.7) | 123 | 70 (56.9) |
| 60+ | 52 | 19 (36.5) | 109 | 47 (43.1) | 181 | 102 (56.4) |
| <16 years | 3746 | 1359 (36.3) | 3844 | 2101 (54.7) | 3878 | 2422 (62.5) |
| ≥16 years | 4933 | 2584 (52.4) | 5851 | 3772 (64.5) | 6297 | 4619 (73.4) |
| <18 years | 4254 | 1658 (39.0) | 4292 | 2452 (57.1) | 4283 | 2770 (64.7) |
| ≥18 years | 4425 | 2285 (51.6) | 5403 | 3421 (63.3) | 5892 | 4271 (72.5) |
| **Overall** | **8679** | **3943 (45.4)** | **9695** | **5873 (60.6)** | **10175** | **7041 (69.2)** |

## 1.32 Hypertonic saline

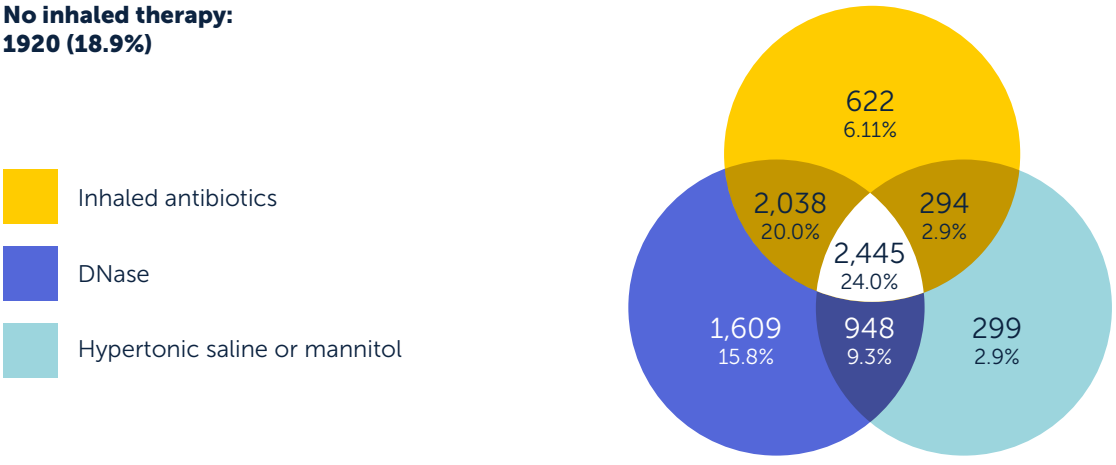
This treatment helps to thin mucus so that it is easier to cough out of the body.

| | 2011 | | 2016 | | 2021 | |
|---|---|---|---|---|---|---|
| Age | Total patients | Patients on hypertonic saline; n (%) | Total patients | Patients on hypertonic saline; n (%) | Total patients | Patients on hypertonic saline; n (%) |
| 0-3 | 964 | 32 (3.3) | 844 | 71 (8.4) | 718 | 111 (15.5) |
| 4-7 | 932 | 77 (8.3) | 1119 | 245 (21.9) | 1013 | 300 (29.6) |
| 8-11 | 835 | 163 (19.5) | 975 | 320 (32.8) | 1127 | 464 (41.2) |
| 12-15 | 1015 | 196 (19.3) | 906 | 425 (46.9) | 1020 | 494 (48.4) |
| 16-19 | 1016 | 184 (18.1) | 943 | 356 (37.8) | 785 | 404 (51.5) |
| 20-23 | 1003 | 170 (16.9) | 998 | 306 (30.7) | 919 | 524 (57.0) |
| 24-27 | 803 | 162 (20.2) | 965 | 267 (27.7) | 926 | 390 (42.1) |
| 28-31 | 642 | 138 (21.5) | 802 | 264 (32.9) | 865 | 259 (29.9) |
| 32-35 | 433 | 88 (20.3) | 648 | 221 (34.1) | 713 | 211 (29.6) |
| 36-39 | 293 | 49 (16.7) | 443 | 144 (32.5) | 614 | 207 (33.7) |
| 40-43 | 278 | 52 (18.7) | 303 | 87 (28.7) | 451 | 139 (30.8) |
| 44-47 | 192 | 36 (18.8) | 267 | 69 (25.8) | 280 | 87 (31.1) |
| 48-51 | 122 | 21 (17.2) | 181 | 49 (27.1) | 261 | 75 (28.7) |
| 52-55 | 61 | 9 (14.8) | 125 | 41 (32.8) | 179 | 45 (25.1) |
| 56-59 | 38 | 5 (13.2) | 67 | 18 (26.9) | 123 | 29 (23.6) |
| 60+ | 52 | 9 (17.3) | 109 | 27 (24.8) | 181 | 58 (32.0) |
| <16 years | 3746 | 468 (12.5) | 3844 | 1061 (27.6) | 3878 | 1369 (35.3) |
| ≥16 years | 4933 | 923 (18.7) | 5851 | 1849 (31.6) | 6297 | 2428 (38.6) |
| <18 years | 4254 | 565 (13.3) | 4292 | 1260 (29.4) | 4283 | 1562 (36.5) |
| ≥18 years | 4425 | 826 (18.7) | 5403 | 1650 (30.5) | 5892 | 2235 (37.9) |
| **Overall** | **8679** | **1391 (16.0)** | **9695** | **2910 (30.0)** | **10175** | **3797 (37.3)** |

## 1.33 Burden of treatment

The Venn diagram shows how many people with CF are on one or more inhaled therapy and the combinations they take. 1920 (18.9%) people are on no inhaled therapies.

**No inhaled therapy: 1920 (18.9%)**



# Other therapies

## 1.34 CFTR modulators

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

### Ivacaftor

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

### Lumacaftor/ivacaftor

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

### Tezacaftor/ivacaftor

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

### Elexacaftor/tezacaftor/ivacaftor

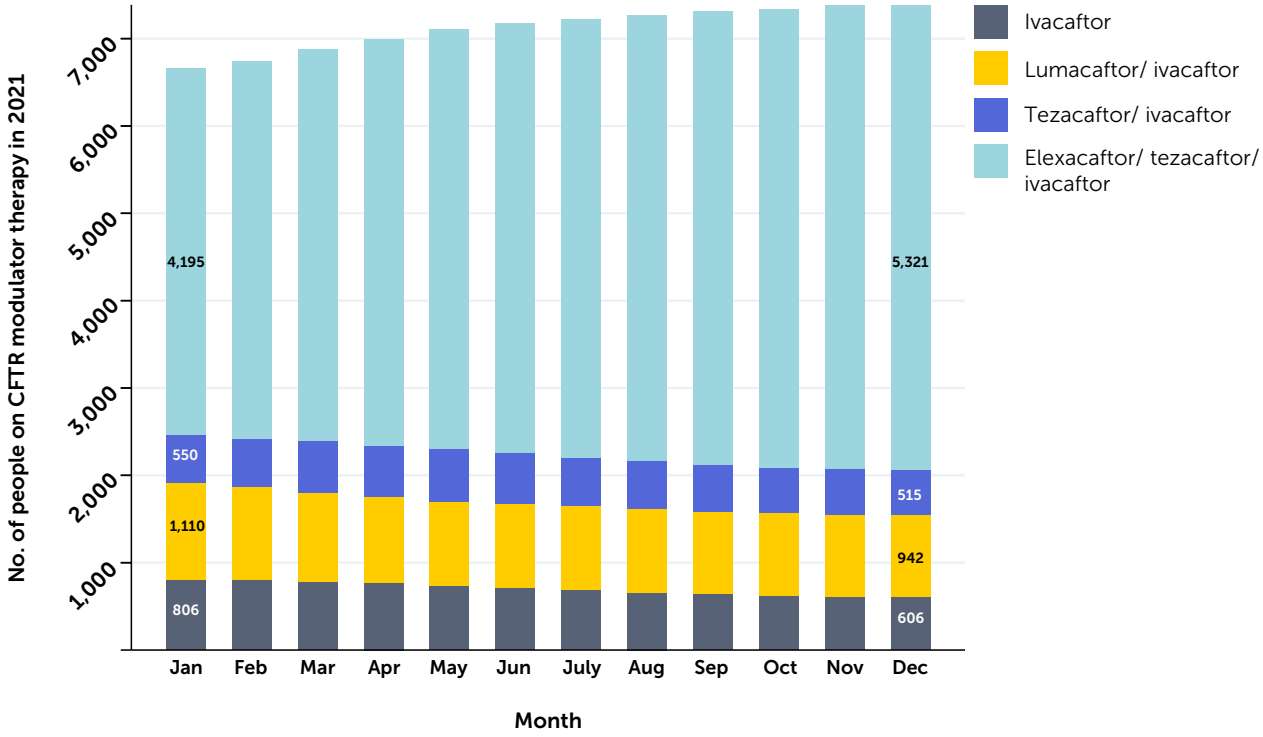
During 2021, Elexacaftor/tezacaftor/ ivacaftor was available in the UK for patients with cystic fibrosis aged 12 and over who have two copies of the F508del mutation, or a single copy of F508del and one minimal function mutation.

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

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

### CFTR modulator use in 2021

The graph below shows the number of people taking each drug by month. Where people switched modulators, the most recent prescription is counted. Only patients who had an annual review are counted. By December, 7384 people were taking a CFTR modulator.



## 1.35 Oxygen and non-invasive ventilation

| | Overall (n=10175) | <16 years (n=3878) | ≥16 years (n=6297) | <18 years (n=4283) | ≥18 years (n=5892) |
|---|---|---|---|---|---|
| Non Invasive Ventillation (NIV); n (%) | 146 (1.4) | 17 (0.4) | 129 (2.0) | 20 (0.5) | 126 (2.1) |
| Any oxygen use; n (%) | 415 (4.1) | 48 (1.2) | 367 (5.8) | 53 (1.2) | 362 (6.1) |
| Among those who had oxygen use: | | | | | |
| Continuously | -* | <5 | 57 (15.5) | <5 | 57 (15.7) |
| Nocturnal or with exertion | 200 (48.2) | 10 (20.8) | 190 (51.8) | 12 (22.6) | 188 (51.9) |
| As required (PRN) | 50 (12.0) | 5 (10.4) | 45 (12.3) | 5 (9.4) | 45 (12.4) |
| With exacerbation | 106 (25.5) | 31 (64.6) | 75 (20.4) | 34 (64.2) | 72 (19.9) |

## 1.36 Physiotherapy

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

| | Overall (n=10175) | <16 years (n=3878) | ≥16 years (n=6297) | <18 years (n=4283) | ≥18 years (n=5892) |
|---|---|---|---|---|---|
| Active cycle of breathing techniques; n (%) | 1273 (12.5) | 320 (8.3) | 953 (15.1) | 367 (8.6) | 906 (15.4) |
| Autogenic drainage (including assisted autogenic drainage); n (%) | 1810 (17.8) | 157 (4.0) | 1653 (26.3) | 205 (4.8) | 1605 (27.2) |
| Postural drainage; n (%) | 633 (6.2) | 475 (12.2) | 158 (2.5) | 505 (11.8) | 128 (2.2) |
| Any form of PEP; n (%) | 6074 (59.7) | 2930 (75.6) | 3144 (49.9) | 3241 (75.7) | 2833 (48.1) |
| VEST; n (%) | 160 (1.6) | 81 (2.1) | 79 (1.3) | 93 (2.2) | 67 (1.1) |
| Exercise; n (%) | 6095 (59.9) | 2523 (65.1) | 3572 (56.7) | 2797 (65.3) | 3298 (56.0) |
| Other; n (%) | 1794 (17.6) | 1022 (26.4) | 772 (12.3) | 1083 (25.3) | 711 (12.1) |

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

## 1.37 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=10175) | <16 years (n=3878) | ≥16 years (n=6297) | <18 years (n=4283) | ≥18 years (n=5892) |
|---|---|---|---|---|---|
| Any supplemental feeding; n(%) | 3523 (34.6) | 1016 (26.2) | 2507 (39.8) | 1149 (26.8) | 2374 (40.3) |
| Nasogastric tube; n(%) | 55 (0.5) | 14 (0.4) | 41 (0.7) | 15 (0.4) | 40 (0.7) |
| Gastrostomy tube/button; n(%) | 457 (4.5) | 177 (4.6) | 280 (4.4) | 205 (4.8) | 252 (4.3) |
| Jejunal; n(%) | 7 (0.1) | 0 (0.0) | 7 (0.1) | 0 (0.0) | 7 (0.1) |
| Total Parenteral Nutrition (TPN); n(%) | <5 | <5 | <5 | <5 | <5 |

* Redacted to adhere to statistical disclosure guidelines.

## 1.38 Transplants

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

| | 2016 | 2017 | 2018 | 2019 | 2020 | 2021 |
|---|---|---|---|---|---|---|
| Number evaluation | 221 | 235 | 247 | 241 | 175 | 78 |
| Number accepted | 96 | 121 | 104 | 96 | 66 | 23 |
| **Number receiving aged <16 years** | <5 | 5 | <5 | <5 | 0 | 0 |
| Bilateral lung | <5 | <5 | 0 | <5 | 0 | 0 |
| Liver | 0 | 0 | <5 | <5 | 0 | 0 |
| Other | 0 | <5 | 0 | 0 | 0 | 0 |
| **Number receiving aged 16+ years** | 51 | 53 | 63 | 54 | 15 | 5 |
| Bilateral lung | 46 | 51 | 58 | 49 | 12 | <5 |
| Liver | <5 | 0 | <5 | <5 | <5 | 0 |
| Other | <5 | <5 | <5 | <5 | <5 | <5 |

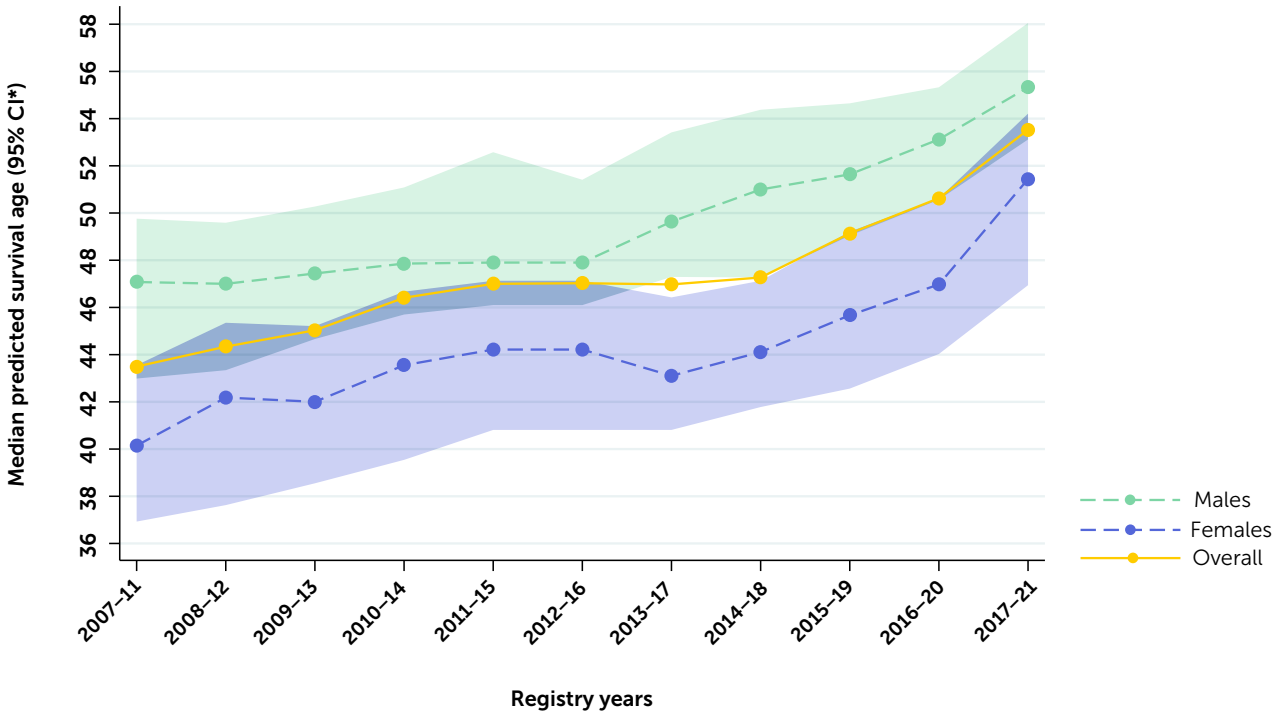
* Redacted to adhere to statistical disclosure guidelines.

# Survival

## 1.39 Median predicted survival age

The calculation of median predicted survival age is based on people with CF who are recorded in the Registry as alive in the given year. A mathematical formula[1] 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 **53.3** years. Half are therefore predicted to die before they reach that age.

Grouping together several years of data gives a better estimate of predicted survival. One-year data can show big variations in median predicted survival age from year to year, which may be due to chance alone and does not necessarily reflect a change in real-world outcomes. A rolling five-year predicted survival is therefore shown to try to smooth out these fluctuations.

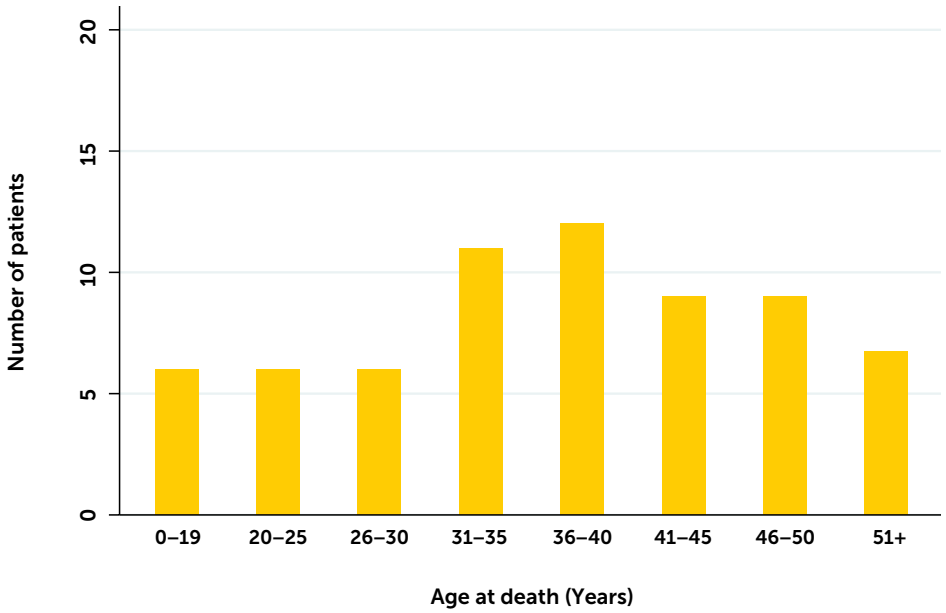



| Mean predicted survival age; years (95% CI*) | | | | |
|---|---|---|---|---|
| Years | Overall | Female | Male | p-value (males vs females) |
| 2007-11 | 43.5(41.9-45.9) | 40.1(36.9-43.6) | 47.1(43.0-49.8) | <0.001 |
| 2008-12 | 44.3(42.4-46.5) | 42.2(37.6-45.3) | 47.0(43.3-49.6) | <0.001 |
| 2009-13 | 45.0(42.8-47.0) | 42.0(38.5-45.2) | 47.4(44.7-50.3) | <0.001 |
| 2010-14 | 46.4(43.7-47.9) | 43.6(39.5-46.7) | 47.9(45.7-51.1) | <0.001 |
| 2011-15 | 47.0(44.3-48.2) | 44.2(40.8-47.1) | 47.9(46.1-52.6) | 0.004 |
| 2012-16 | 47.0(44.7-48.2) | 44.2(40.8-47.1) | 47.9(46.1-51.4) | 0.003 |
| 2013-17 | 47.0(44.8-48.2) | 43.1(40.8-46.4) | 49.6(47.3-53.4) | <0.001 |
| 2014-18 | 47.3(45.7-49.6) | 44.1(41.8-47.1) | 51.0(47.3-54.4) | <0.001 |
| 2015-19 | 49.1(47.0-51.4) | 45.7(42.6-49.2) | 51.6(49.0-54.6) | <0.001 |
| 2016-20 | 50.6(48.2-53.1) | 47.0(44.0-50.6) | 53.1(50.6-55.3) | 0.004 |
| 2017-21 | 53.5(51.5-55.2) | 51.4(46.9-54.2) | 55.3(53.1-58.1) | 0.002 |

[1] Sykes, Jenna et al. J Clin Epidemiol. 2016;70:206-213.
* Confidence interval

## 1.40 Age distribution of deaths in 2021

The table below shows the ages of the 66 people with CF who died in 2021. In 2021 the median age of the 66 people who died was 38.




| Age at death | Number of patients |
|---|---|
| 0-19 | 6 |
| 20-25 | 6 |
| 26-30 | 6 |
| 31-35 | 11 |
| 36-40 | 12 |
| 41-45 | 9 |
| 46-50 | 9 |
| 51+ | 7 |
| **Total** | **66** |

## 1.41 Causes of death

This table shows all the recorded causes of death between 2019 – 2021.

| Cause of death | Number (%) |
|---|---|
| Respiratory/cardiorespiratory | 171 (61.7) |
| Transplant-related | 36 (13.0) |
| Not known | 20 (7.2) |
| Cancer | 16 (5.8) |
| Other | 15 (5.4) |
| Liver disease/liver failure | 9 (3.2) |
| COVID-19 | -* |
| Trauma or Suicide | <5 |
| **Total** | 277 |

* redacted to adhere to statistical disclosure guidelines

# 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 cause 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 | 10801 (99.0) |
| Patients genotyped with both mutations recorded | 10503 (96.3) |
| **F508del mutations** | |
| Homozygous F508del | 5206 (47.7) |
| Heterozygous F508del | 4508 (41.3) |

## 1.42 Mutation combinations in the UK population

This tabulation shows the proportion (%) of patients with the most common mutation combinations in their genotype. For example, 4.2% of the UK population have one copy of F508del and one copy of G551D.

| Mutation | F508del | R117H | G551D | G542X | 621+1G->T | Other | Unknown | Total |
|---|---|---|---|---|---|---|---|---|
| F508del | 47.7 | | | | | | | 47.7 |
| R117H | 5.0 | 0.1 | | | | | | 5.1 |
| G551D | 4.1 | 0.2 | 0.2 | | | | | 4.5 |
| G542X | 2.5 | 0.1 | 0.1 | 0.1 | | | | 2.8 |
| 621+1G->T | 1.7 | 0.1 | 0.1 | 0.1 | 0.1 | | | 2.0 |
| Other | 26.1 | 0.6 | 0.9 | 0.7 | 0.5 | 5.3 | | 34.1 |
| Unknown | 1.9 | 0.1 | 0.1 | 0.1 | 0.0 | 0.5 | 1.0 | 3.7 |
| **Total** | **89.1** | **1.2** | **1.4** | **1.0** | **0.6** | **5.8** | **1.0** | **100.0** |

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

## 1.43 Mutations 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 because people with heterozygous mutations appear twice in the table.

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

| Nucleotide | Protein | Legacy name | N | % |
|---|---|---|---|---|
| c.1521_1523delCTT | p.Phe508del | F508del | 9714 | 89.1 |
| c.350G->A | p.Arg117His | R117H | 680 | 6.2 |
| c.1652G->A | p.Gly551Asp | G551D | 625 | 5.7 |
| c.1624G->T | p.Gly542X | G542X | 397 | 3.6 |
| c.489+1G->T | | 621+1G->T | 280 | 2.6 |
| c.3909C->G | p.Asn1303Lys | N1303K | 172 | 1.6 |
| c.1585-1G->A | | 1717-1G->A | 167 | 1.5 |
| c.1766+1G->A | | 1898+1G->A | 150 | 1.4 |
| c.3454G->C | p.Asp1152His | D1152H | 144 | 1.3 |
| c.200C->T | p.Pro67Leu | P67L | 144 | 1.3 |
| c.3140-26A->G | | 3272-26A->G | 118 | 1.1 |
| c.3528delC | p.Lys1177SerfsX15 | 3659delC | 115 | 1.1 |
| c.1679G->C | p.Arg560Thr | R560T | 102 | 0.9 |
| c.1519_1521delATC | p.Ile507del | I507del | 93 | 0.9 |
| c.1477C->T | p.Gln493X | Q493X | 92 | 0.8 |
| c.3717+12191C->T | | 3849+10kbC->T | 85 | 0.8 |
| c.1657C->T | p.Arg553X | R553X | 84 | 0.8 |
| c.254G->A | p.Gly85Glu | G85E | 82 | 0.8 |
| c.178G->T | p.Glu60X | E60X | 77 | 0.7 |
| c.2657+5G->A | | 2789+5G->A | 74 | 0.7 |

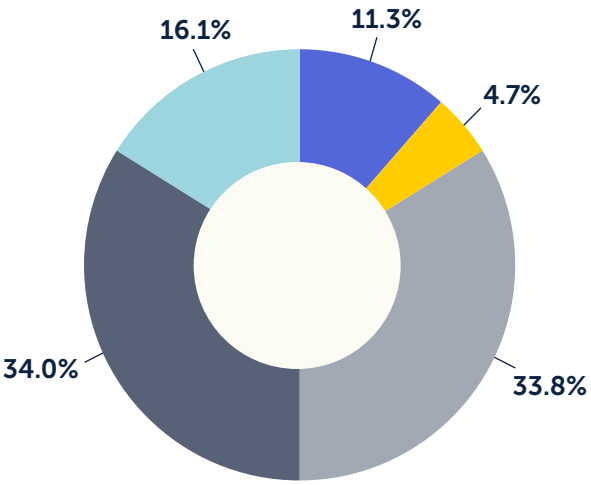
## 1.44 Mutation prevalence by devolved nation

This table shows the distribution of individual mutations across the devolved nations. The number of patients for each devolved nation is based on the location of the CF centre at which the patient receives care and does not account for patients who travel between devolved nations for care. The groups are not mutually exclusive because people with heterozygous mutations appear twice in the table.

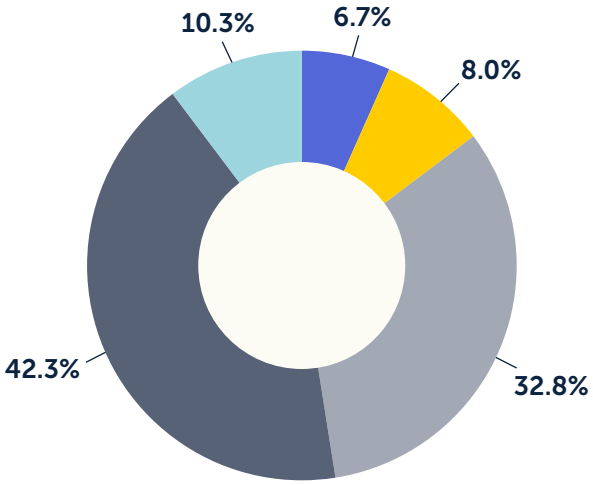
| Legacy name | England N=9044 | | Scotland N=925 | | Wales N=454 | | Northern Ireland N=485 | |
|---|---|---|---|---|---|---|---|---|
| | n | % | n | % | n | % | n | % |
| F508del | 8072 | 89.3% | 830 | 89.7% | 405 | 89.2% | 407 | 83.9% |
| R117H | 510 | 5.6% | 78 | 8.4% | 17 | 3.7% | 75 | 15.5% |
| G551D | 471 | 5.2% | 92 | 9.9% | 15 | 3.3% | 47 | 9.7% |
| G542X | 284 | 3.1% | 63 | 6.8% | 22 | 4.8% | 28 | 5.8% |
| 621+1G->T | 204 | 2.3% | 10 | 1.1% | 49 | 10.8% | 17 | 3.5% |
| N1303K | 144 | 1.6% | 12 | 1.3% | 7 | 1.5% | 9 | 1.9% |
| 1717-1G->A | 149 | 1.6% | 15 | 1.6% | <5 | - | <5 | - |
| 1898+1G->A | 119 | 1.3% | <5 | - | 27 | 5.9% | 0 | 0.0% |
| P67L | 75 | 0.8% | 49 | 5.3% | <5 | - | 18 | 3.7% |
| D1152H | 112 | 1.2% | 18 | 1.9% | <5 | - | 11 | 2.3% |

## 1.45 Genotype prevalence by devolved nation

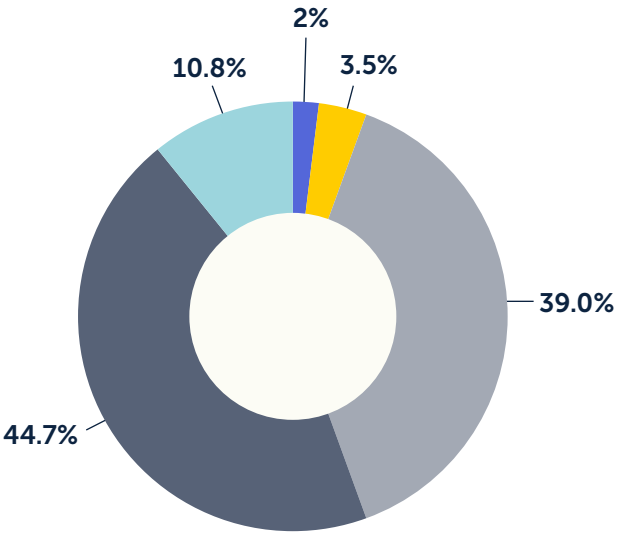
These charts show the distribution of mutation combinations across the devolved nations. The number of patients for each devolved nation is based on the location of the CF centre at which the patient receives care and does not account for patients who travel between devolved nations for care.



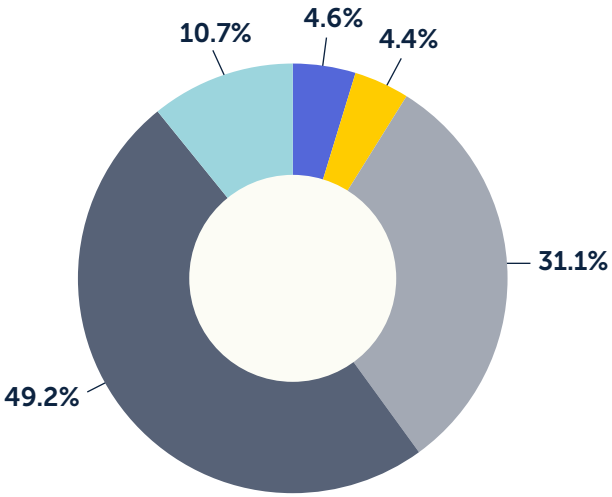

Northern Ireland
N=485




Scotland
N=925




Wales
N=454




England
N=9044

Homozygous F508del

F508del + other

F508del + gating (incl. G551D)

F508del + R117H

Non-F508del combinations

# Section 2 and 3: Centre-level analysis

Cystic fibrosis care in the UK is led by 56 regional centres, 4 stand-alone clinics and 76 networked clinics. The breakdown between centres and clinics delivering paediatric and adult care is shown below:

| | Paediatric | Adult | Total |
|---|---|---|---|
| Centres | 30 | 26 | 56 |
| Stand-alone clinics | 2 | 2 | 4 |
| Networked clinics | 69 | 7 | 76 |

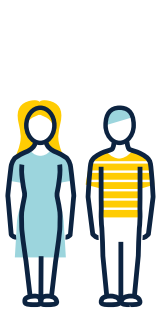
Section 2 shows analysis of data for individual CF centres. This allows people with CF, their families, and healthcare providers, to review a centre's use of some medications and outcome data alongside national averages. This transparency is intended to help improve standards of care overall.

**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, certain patient characteristics such as their gender, as well as facilities, care pathways, and the medical team providing care.**

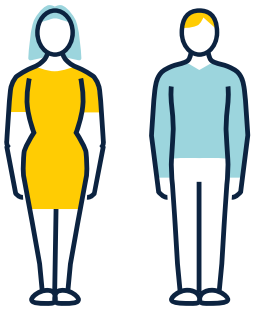
If a person with CF or a member of their family has questions about the results for their CF centre or clinic, they should discuss this with their CF team.

Full tables of the data are shown in appendix 2 on page 68.

**Key**



Paediatric centre

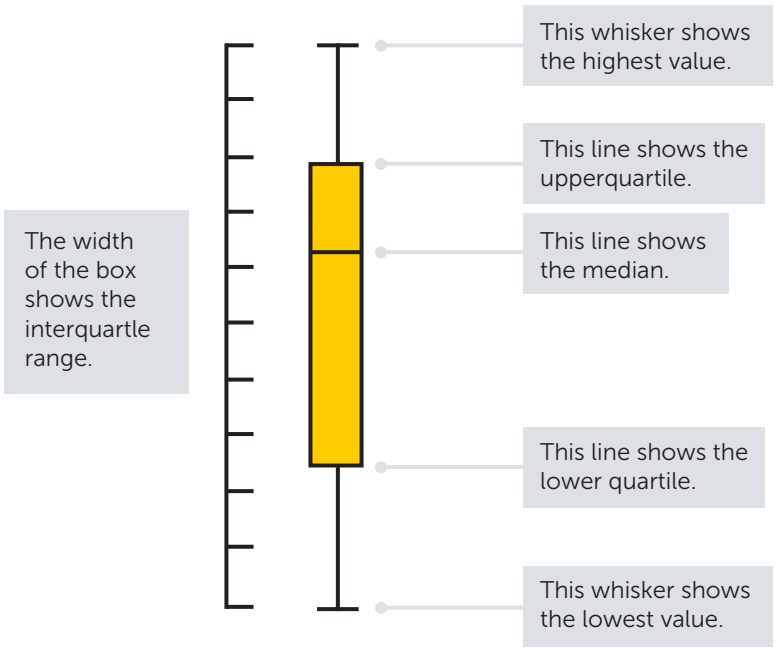


Adult centre

## 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.

## Funnel plots

The more people with CF at a care site, the closer to the national average you would expect the results to be. This is because high numbers in one centre affect the overall average across the country, 'pulling' the average towards them. When a small number people with CF are treated at a site, even a single outcome that is unusual affects the overall result for that site much more.

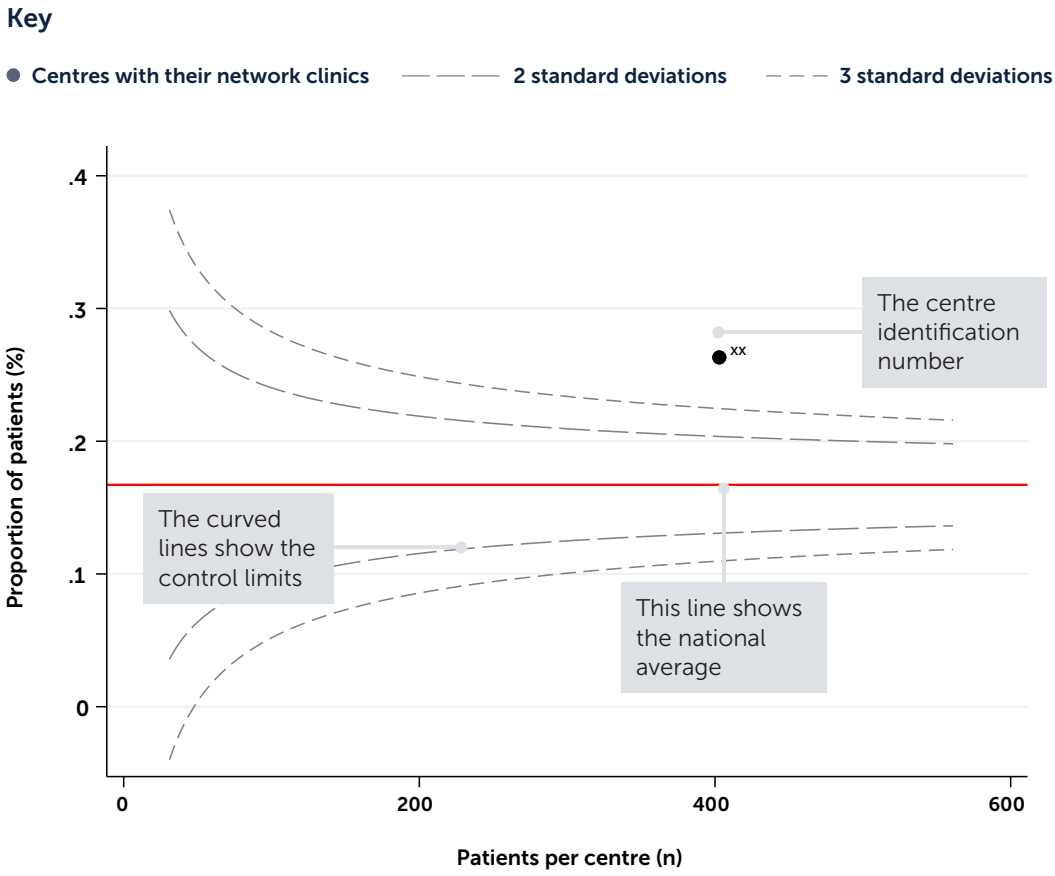
There will always be some natural variation between centres because of differences between the populations receiving care. Using only the national average as a standard can make it difficult to tell whether a survival rate that sits above the national average is higher than we would expect it to be, or not.

For this reason, the funnel plots also show 'control limits'; the curved lines on the charts that give them the 'funnel' shape. The horizontal line in the middle of the funnel shows the national average. Control limits show the rate we would expect, based on the number of people with CF at that site.

If the result for a CF centre is between the two 'control limits', it is 'as expected' and any variation above or below the national average may be due to chance alone. If a result is below the bottom control it is lower than expected, if it is above the upper control, it is higher than expected. Being outside the control limits can be a good thing, for example if a site's lung function results are exceptionally high.

A centre's data can sit outside of the control limits for a number of reasons, including patient characteristics (for example, an adult centre with younger patients might have a higher average lung function than one with older patients), problems with data submitted to the Registry, specialist practice, chance, or the care being delivered.

Where charts have been 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. Because it is difficult for adjustment to fully account for all factors that might affect clinical outcomes, we should be very careful about drawing conclusions based on adjusted outcomes alone.

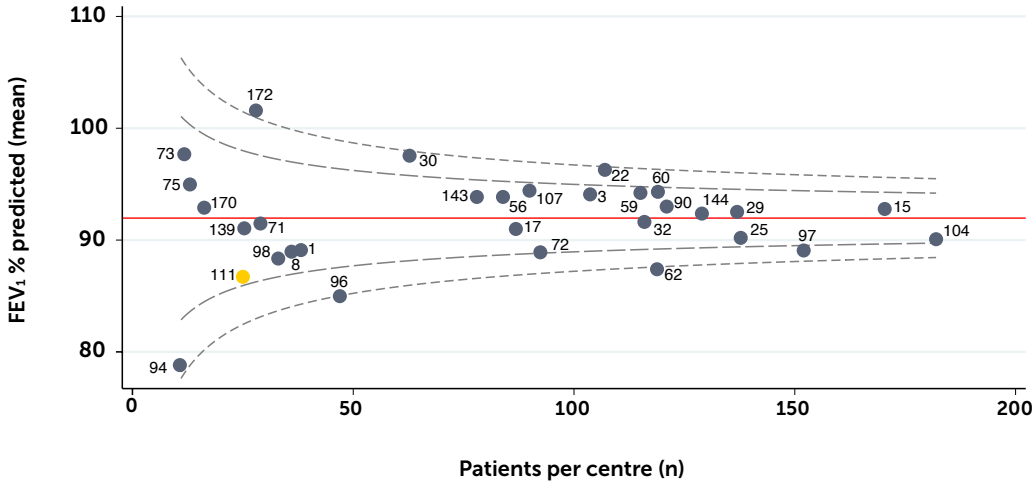


# Section 2 Paediatric centre analysis

## N=4,187

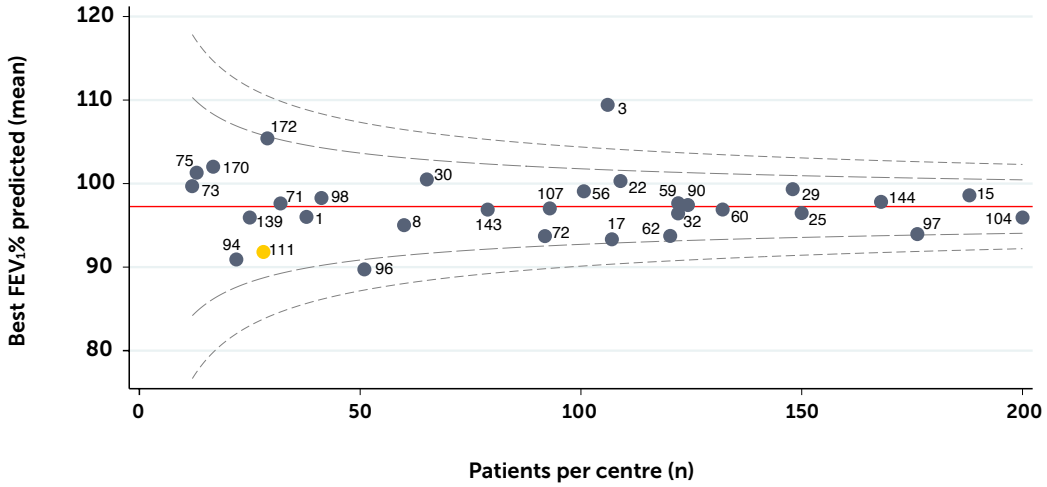
In the UK, paediatric CF care is led by 30 specialist CF centres and two stand-alone clinics (●). Some paediatric centres oversee care delivered by 69 smaller, networked clinics. Data from smaller networked clinics is included in the paediatric centre's data.

## 2.1 Age-adjusted $FEV_1$ % predicted at annual review, in patients aged six and over without a history of lung transplant, by paediatric centre/clinic



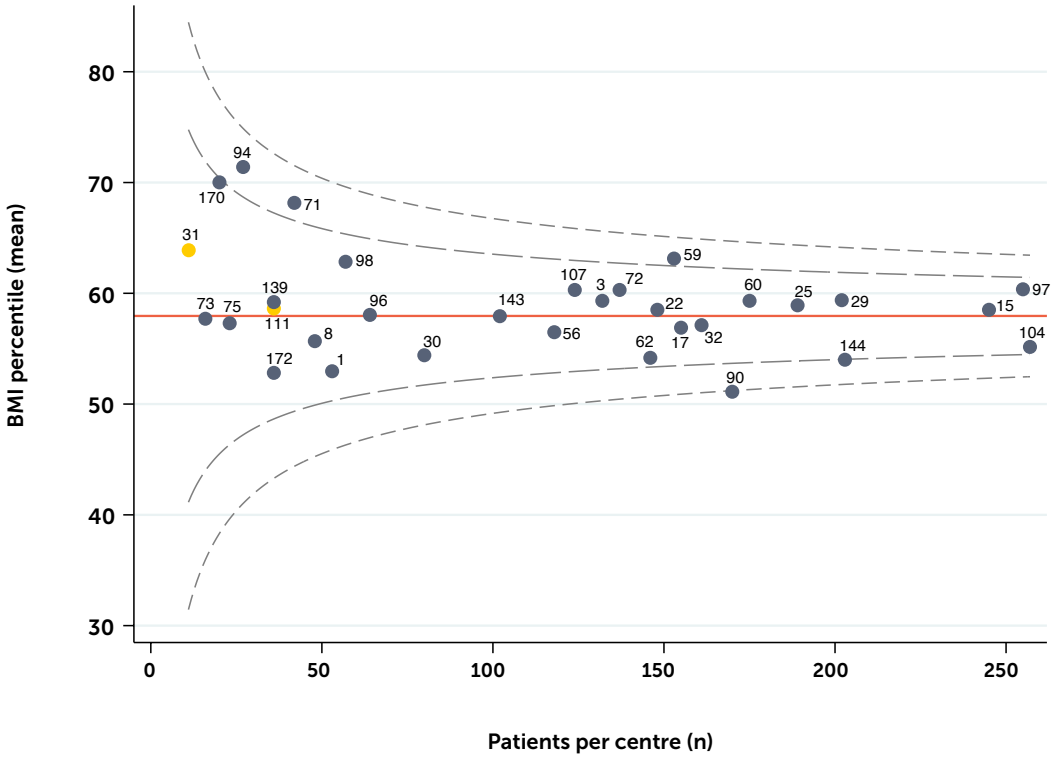

The mean $FEV_1$% predicted for patients attending paediatric centres/clinics is 92.0% predicted.

## 2.2 Age-adjusted Best $FEV_1$ % predicted at annual review, in patients aged six and over without a history of lung transplant, by paediatric centre/clinic



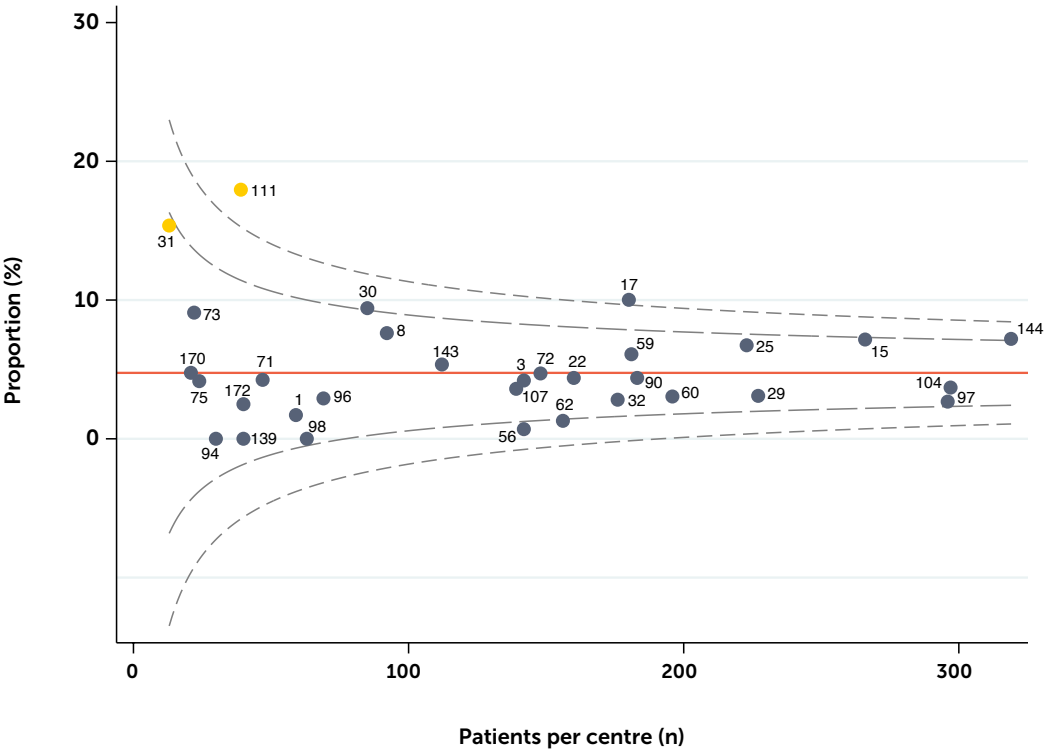

The mean Best $FEV_1$% predicted for patients attending paediatric centres/clinics is 97.3% predicted. Where Best $FEV_1$% predicted was missing, the $FEV_1$% predicted at annual review was used.

## 2.3 Age-adjusted Body Mass Index (BMI) percentile in patients aged 1-15 years by paediatric centre/clinic



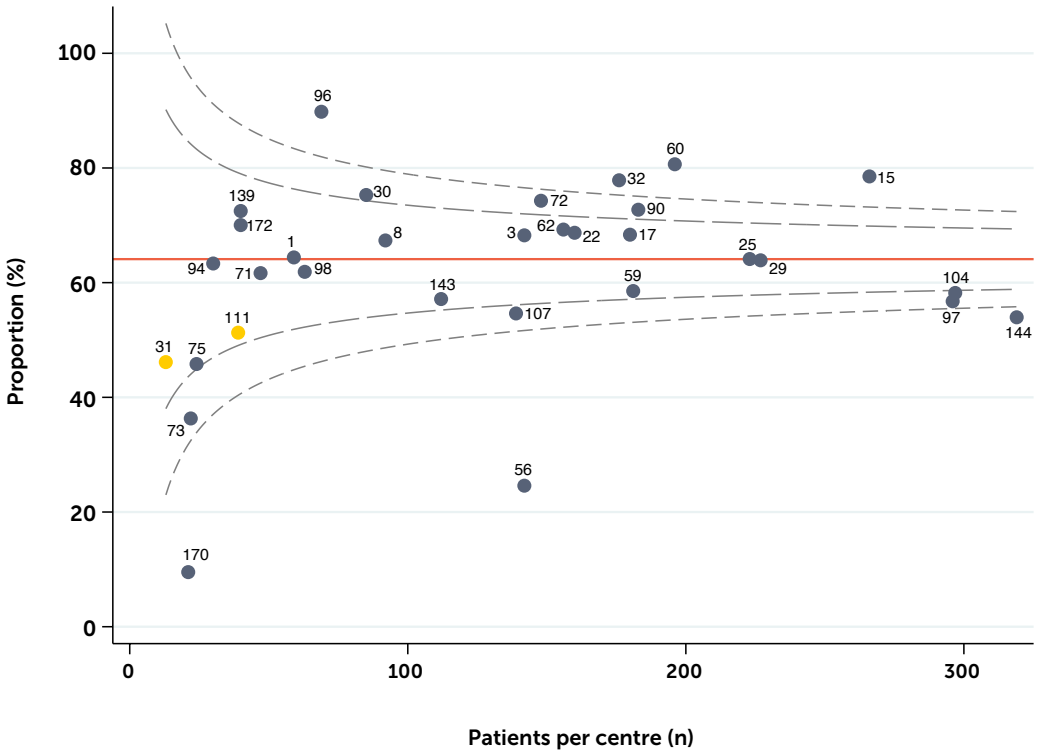

The mean BMI percentile for patients attending paediatric centres/clinics is 58.0%.

## 2.4 Proportion of patients with chronic *Pseudomonas aeruginosa* by paediatric centre/clinic



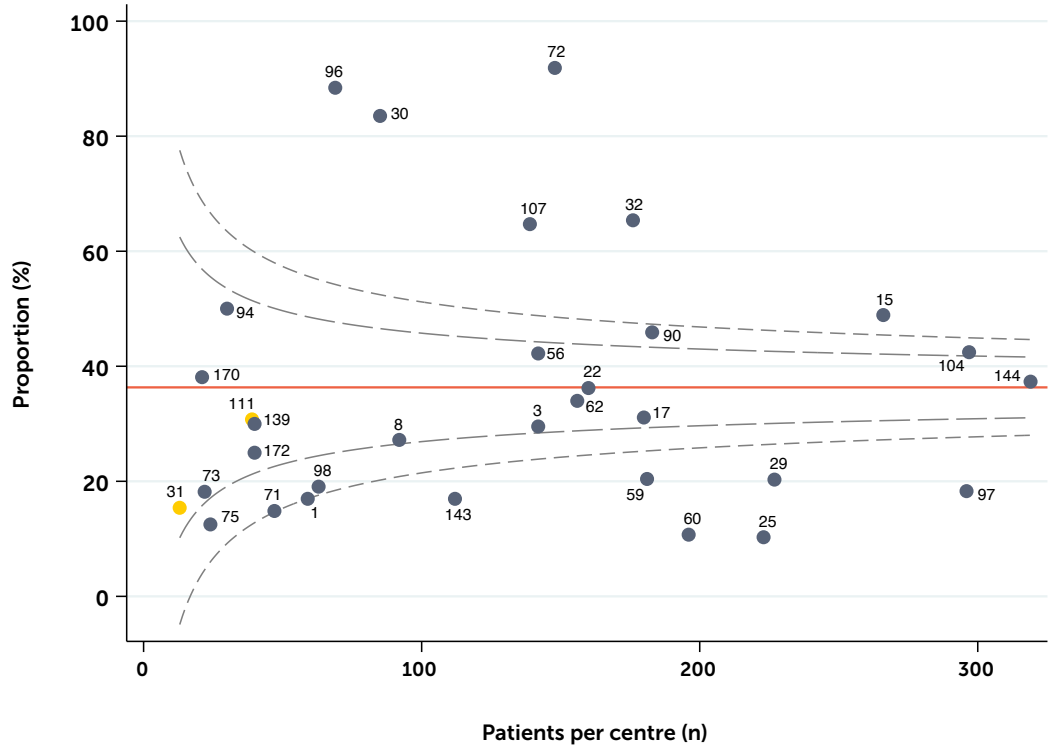

The proportion of patients with chronic *Pseudomonas aeruginosa* in paediatric centres/clinics is 4.8%.

## 2.5 Proportion of patients receiving DNAse treatment by paediatric centre/clinic



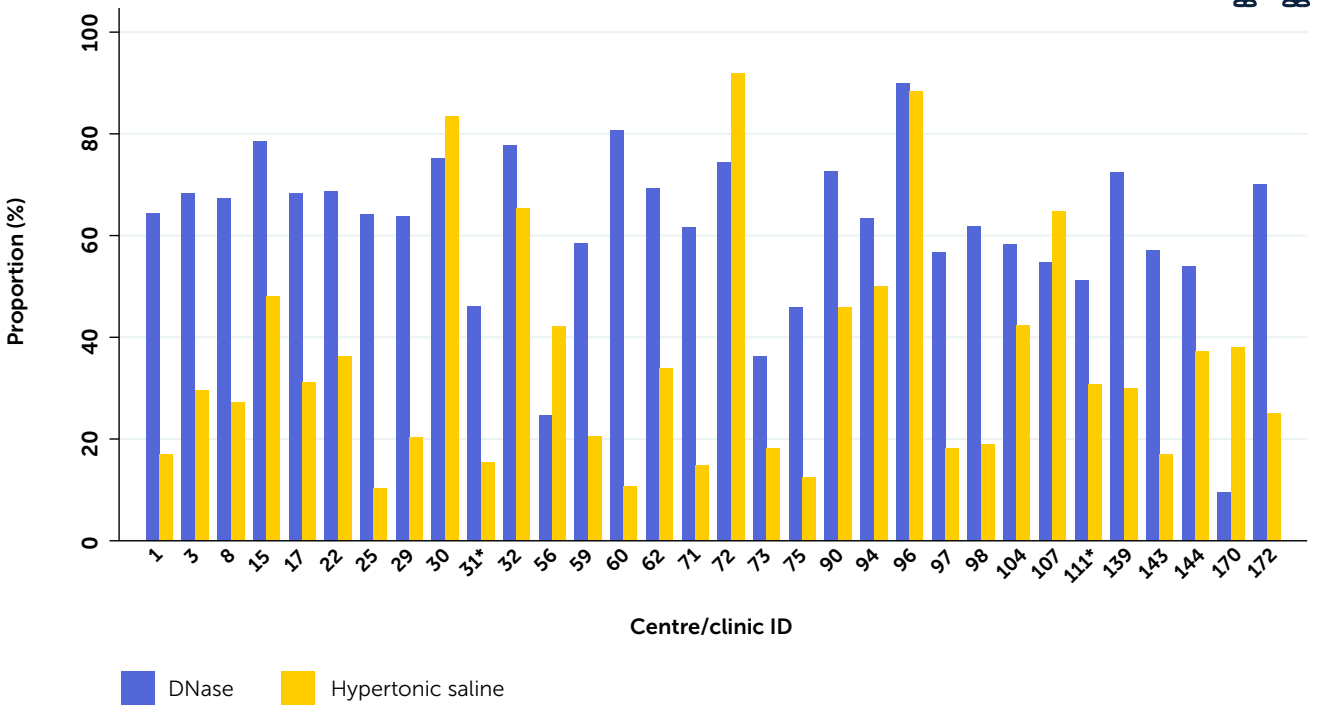

The proportion of patients receiving DNase treatment in paediatric centres/clinics is 64.1%.

## 2.6 Proportion of patients on hypertonic saline or mannitol treatment by paediatric centre/clinic




The proportion of patients receiving hypertonic saline or mannitol treatment in paediatric centres/clinics is 36.3%.

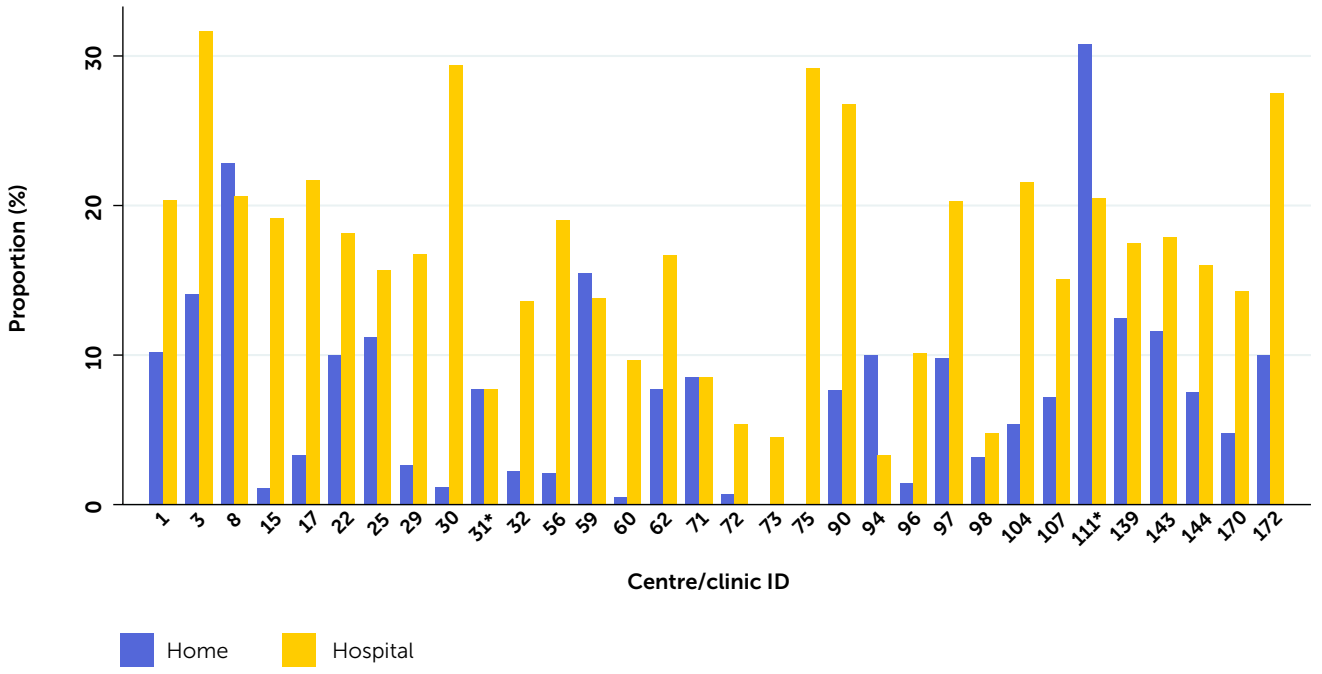
## 2.7 Proportion of patients receiving DNase/hypertonic saline/ mannitol treatment by paediatric centre/clinic




Due to the small number of paediatric patients that received mannitol (<5 across all clinics/centres), receipt of mannitol is omitted from the above graph.

## 2.8 IV use by paediatric centre/clinic

The chart below shows the proportion of patients with at least one IV day at home and/or in hospital. Patients may have a combination of home and hospital IV days.

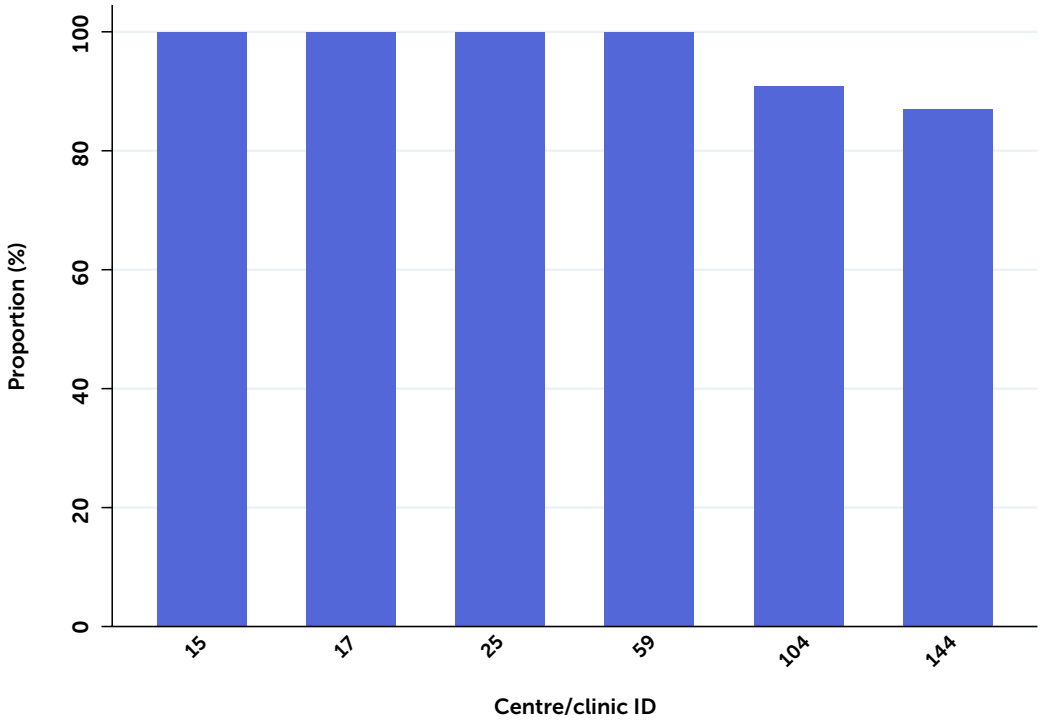



The proportion of patients receiving IVs at home was 7.0% and in hospital was 17.7%. The proportion receiving any IVs was 19.7%.

*Stand-alone clinics

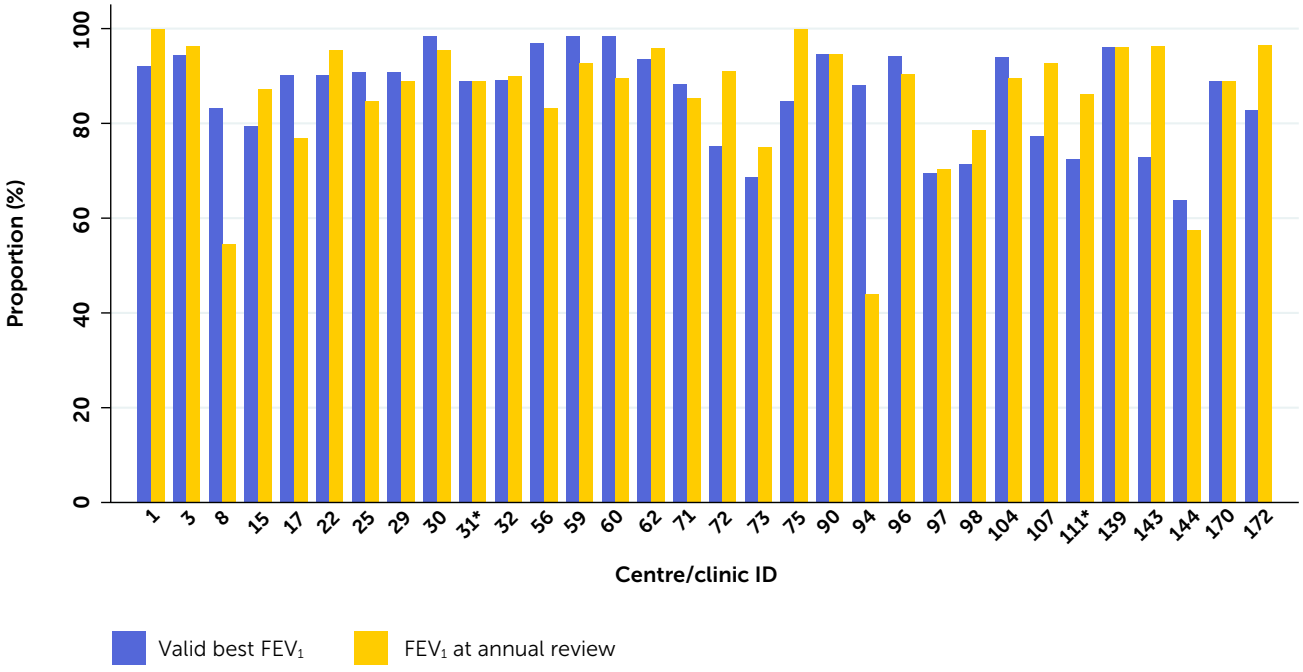
## 2.9 Inhaled antibiotic use for patients with chronic *Pseudomonas aeruginosa*, by paediatric centre/clinic

This chart excludes centres where fewer than 10 patients had chronic *P. aeruginosa*.




91.5% of patients with chronic *P. aeruginosa* received inhaled antibiotics.

## 2.10 Data completeness by paediatric centre/clinic**




*Stand-alone clinics

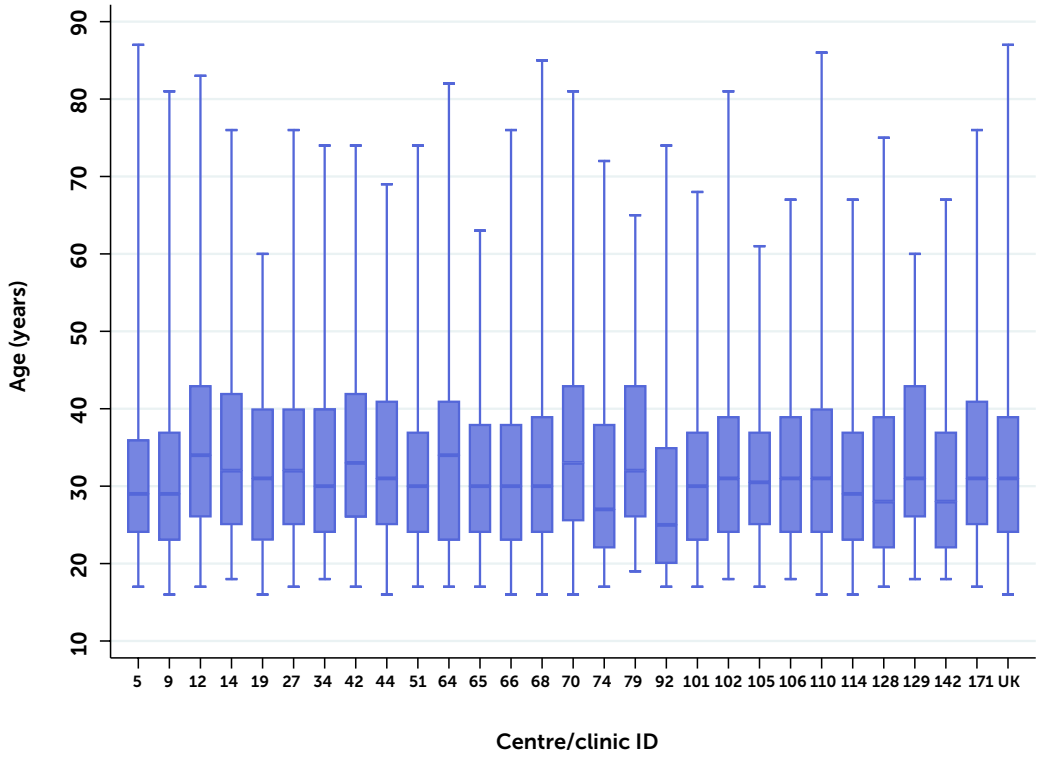
**The chart above shows the proportion of patients who had a valid best $FEV_1$% and an $FEV_1$% at annual review, excluding patients under six years of age. Best $FEV_1$% was considered valid if it was not missing, and the per cent predicted was not more than 0.5% lower than the annual review value.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

# Section 3: Adult centre analysis

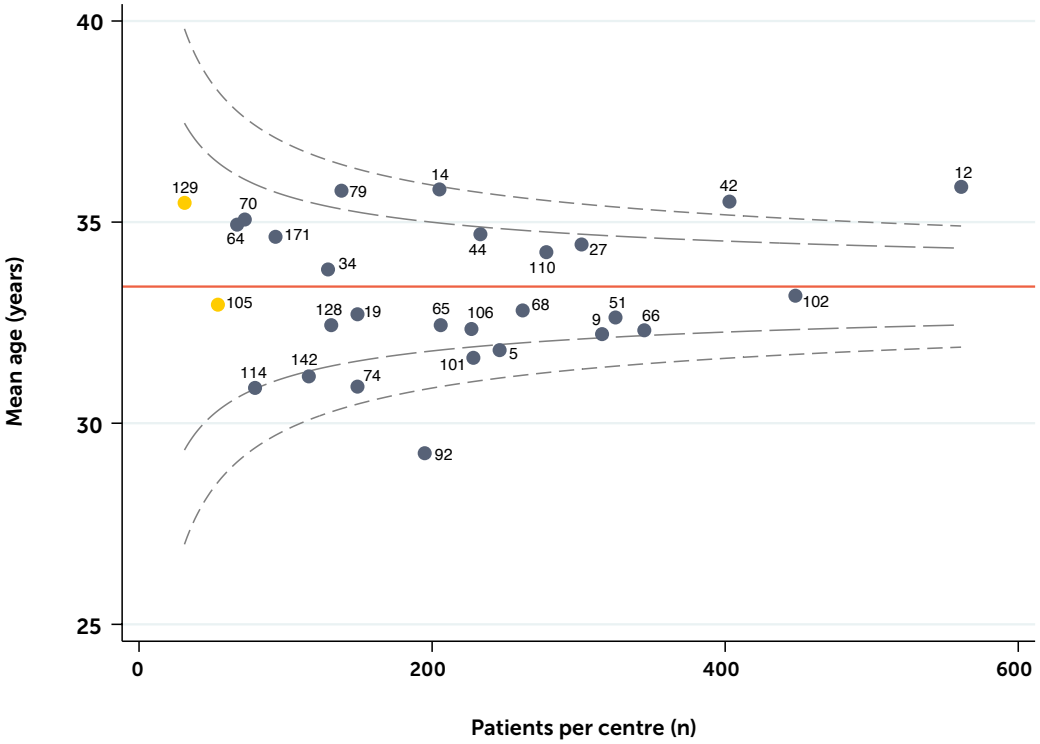
## N=5,988

In the UK, CF care is led by 26 adult specialist CF centres and two stand alone clinics (🟡). People with CF transfer to adult care centres between the ages of 16 and 18 years.

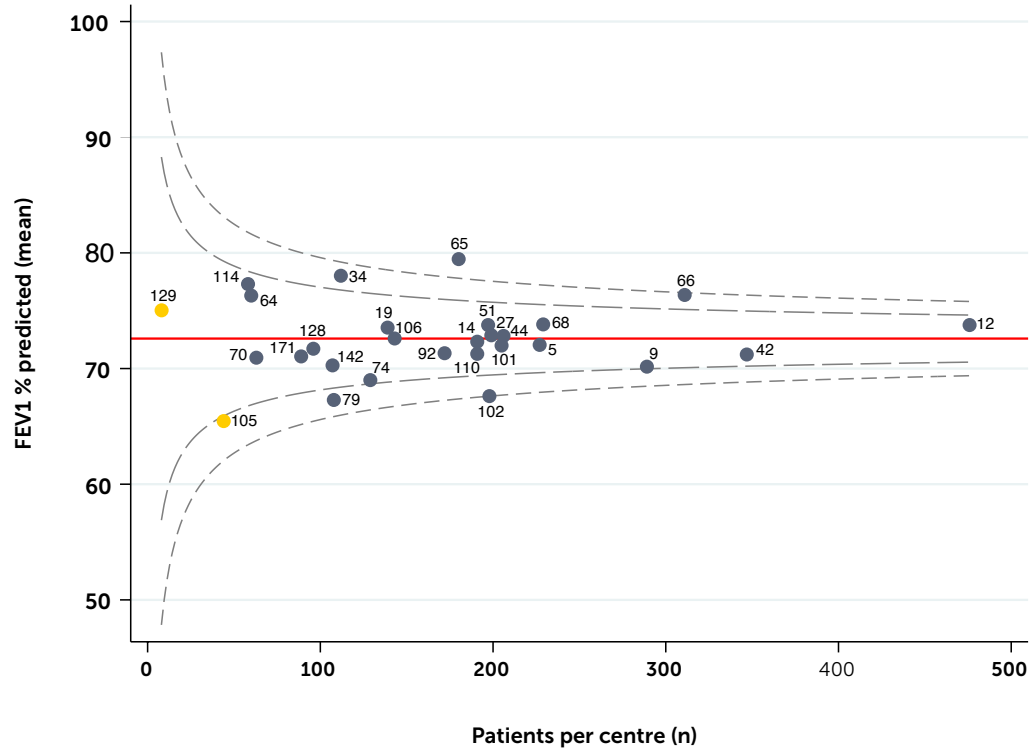
## 3.1 Age distribution by adults centre / clinic




The funnel plot below shows how the mean age in adult centres compares to the national mean. In 2021 the national mean age of patients at CF centres was 32.7 years.

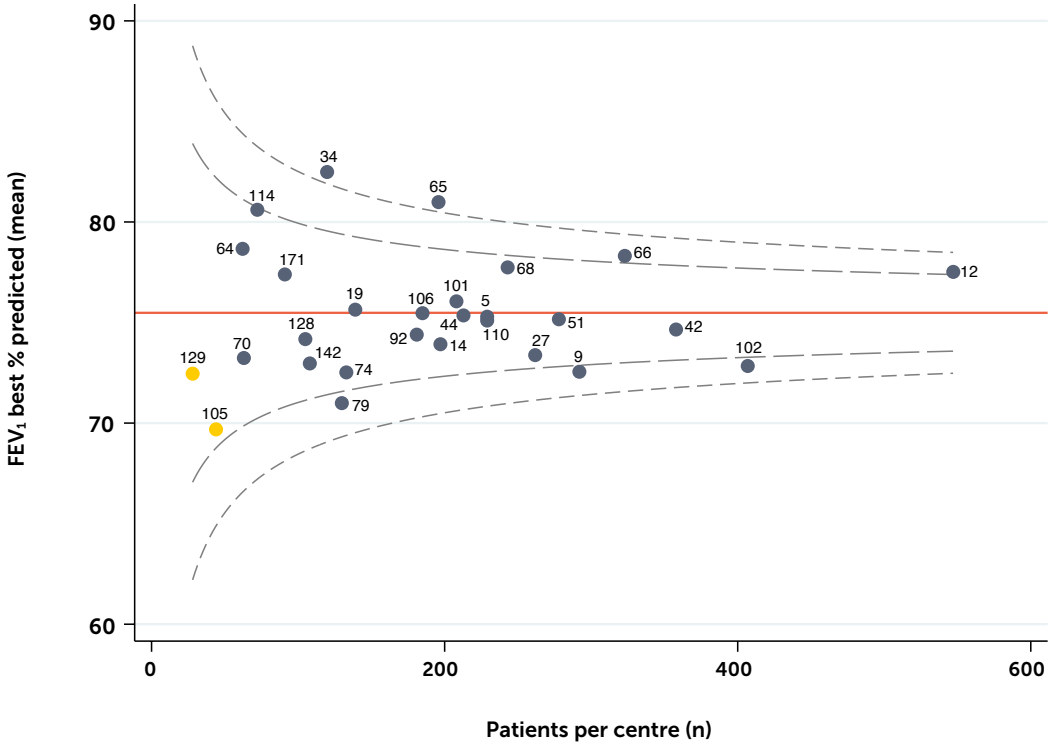


## 3.2 Age adjusted $FEV_1$ % predicted at annual review in patients without a history of lung transplant, by adult centre / clinic



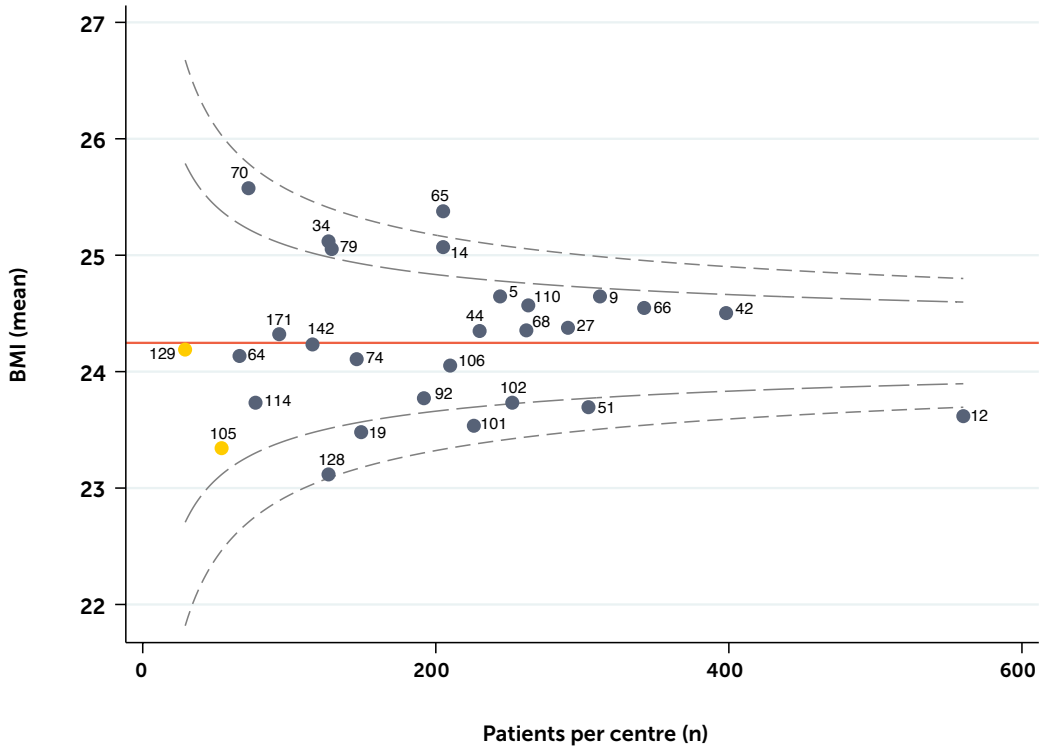

The mean $FEV_1$% predicted in adult centres/clinics is 72.6%.

## 3.3 Age adjusted Best $FEV_1$ % predicted at annual review in patients without a history of lung transplant, by adult centre / clinic



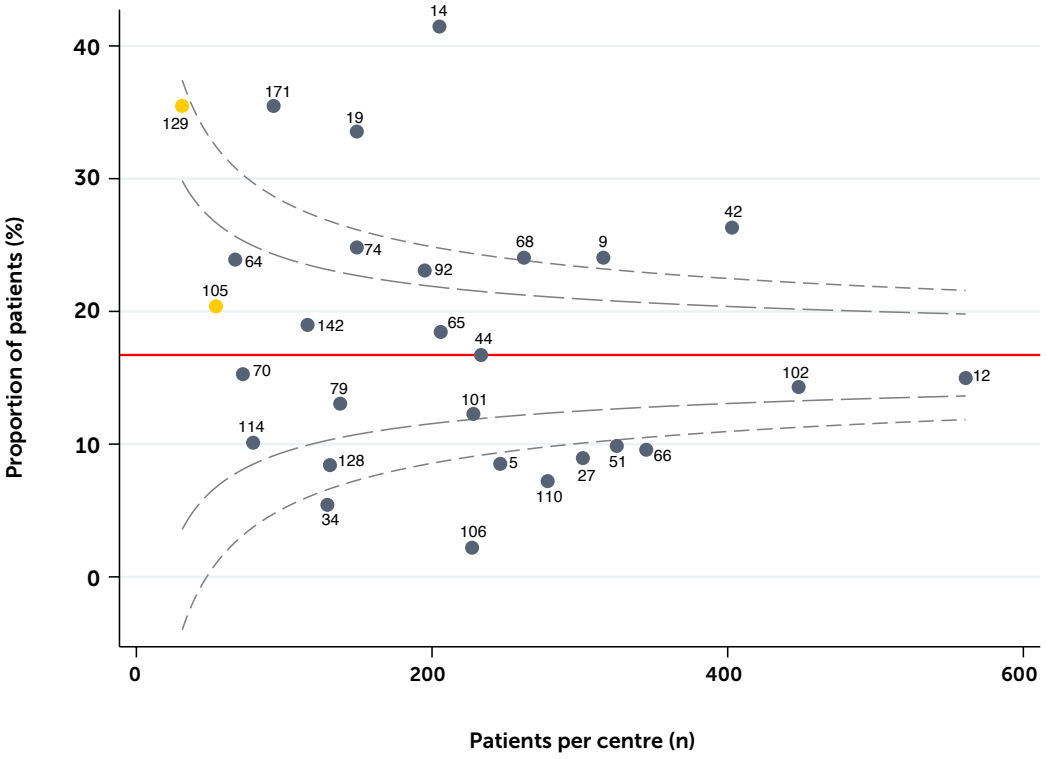

In 2021 the national mean was 75.5%. Where Best $FEV_1$% predicted was missing, or lower than the $FEV_1$ at annual review, the $FEV_1$% value at annual review was used.

## 3.4 Age-adjusted Body Mass Index (BMI) among patients aged 16 years and older by adult centre / clinic



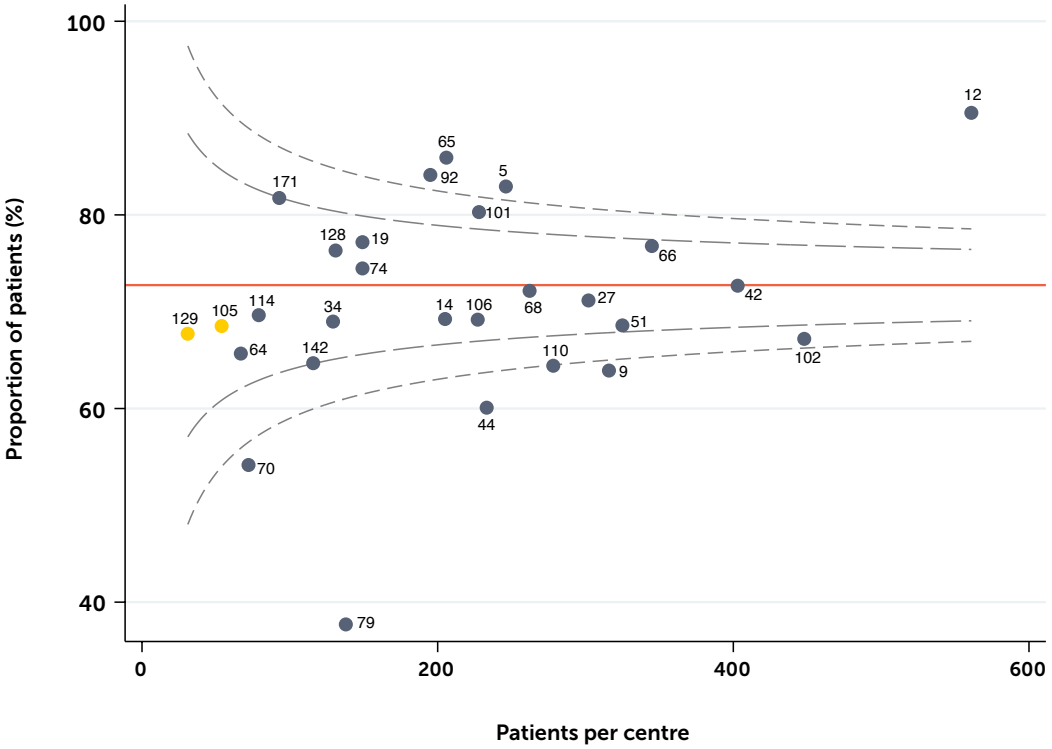

The mean BMI in adult centres/clinics is 24.2.

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



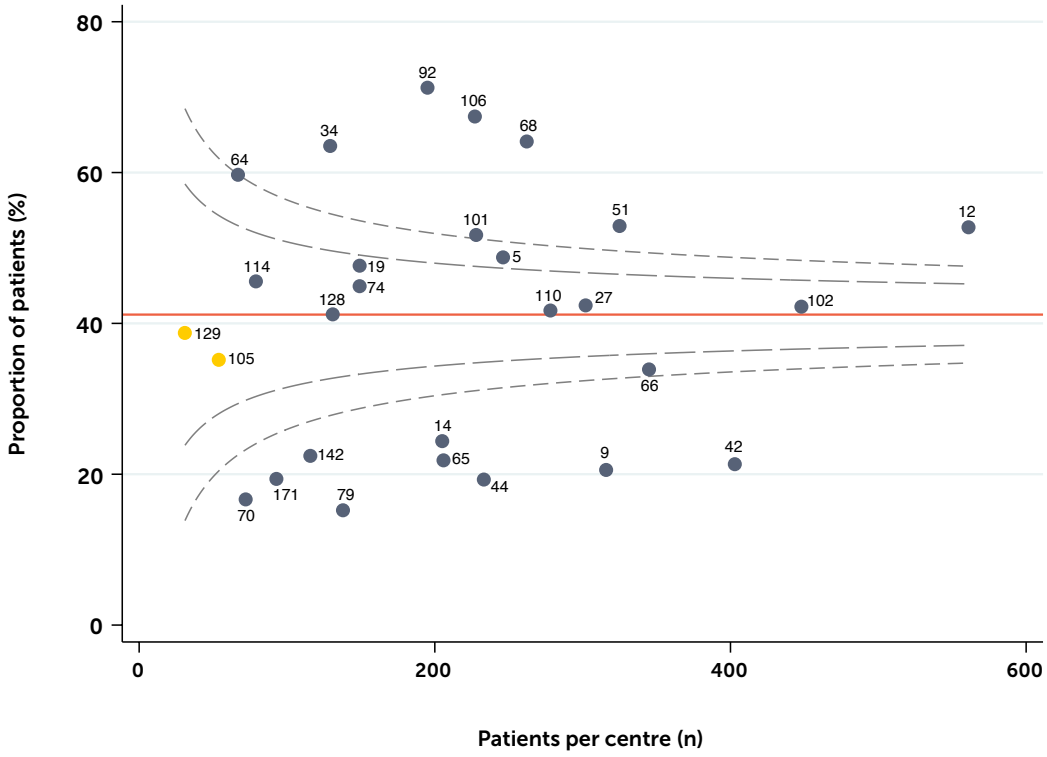

The proportion of patients with chronic *P. aeruginosa* in adult centres/clinics is 32.6%.

## 3.6 Proportion of patients receiving DNase treatment by adult centre / clinic



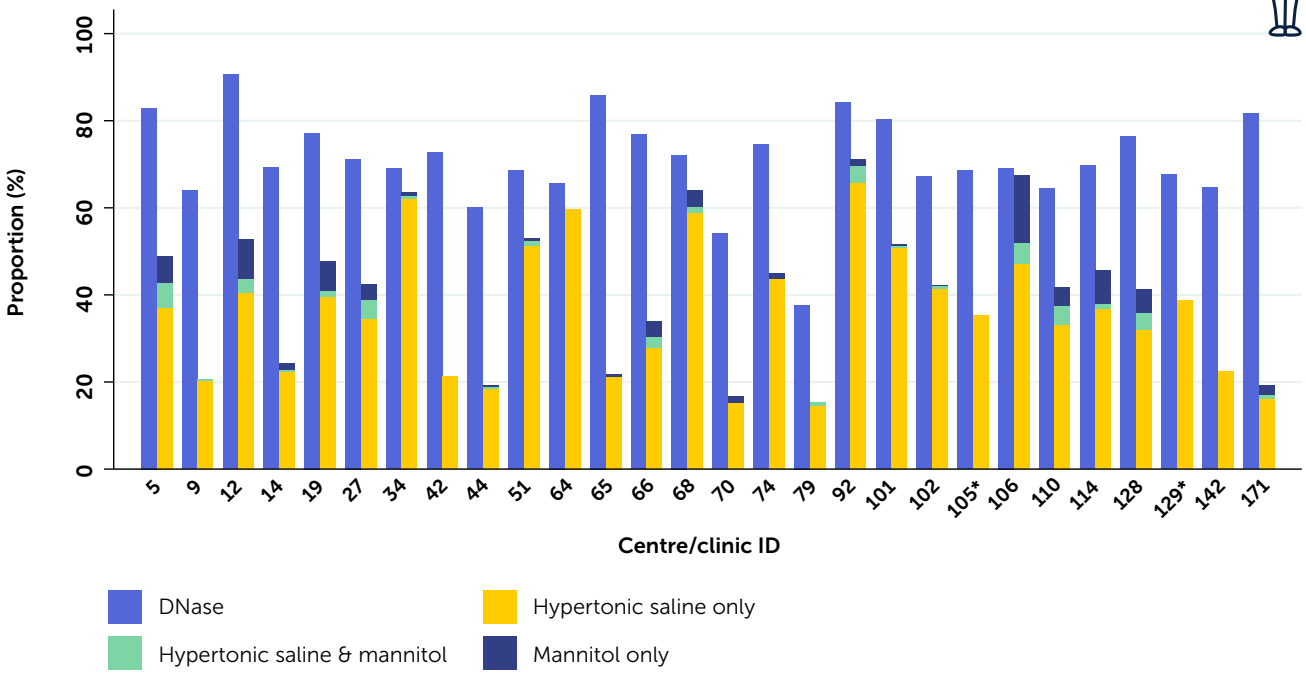

The proportion of patients receiving DNase treatment in adult centres/clinics is 72.7%.

## 3.7 Proportion of patients receiving hypertonic saline or mannitol by adult centre / clinic



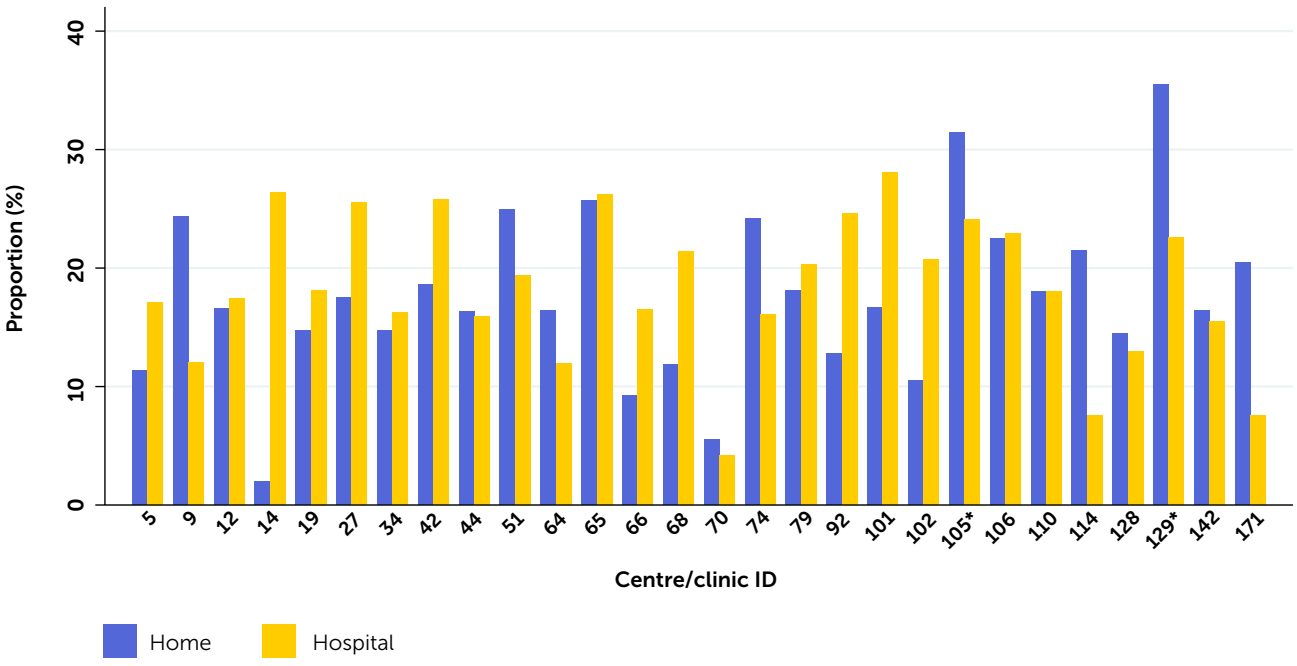

The proportion of patients receiving hypertonic saline or mannitol treatment in adult centres/ clinics is 41.2%.

## 3.8 Proportion of patients receiving DNase/hypertonic saline/ mannitol treatment by adult centre / clinic




## 3.9 Intravenous (IV) antibiotic use by adult centre / clinic

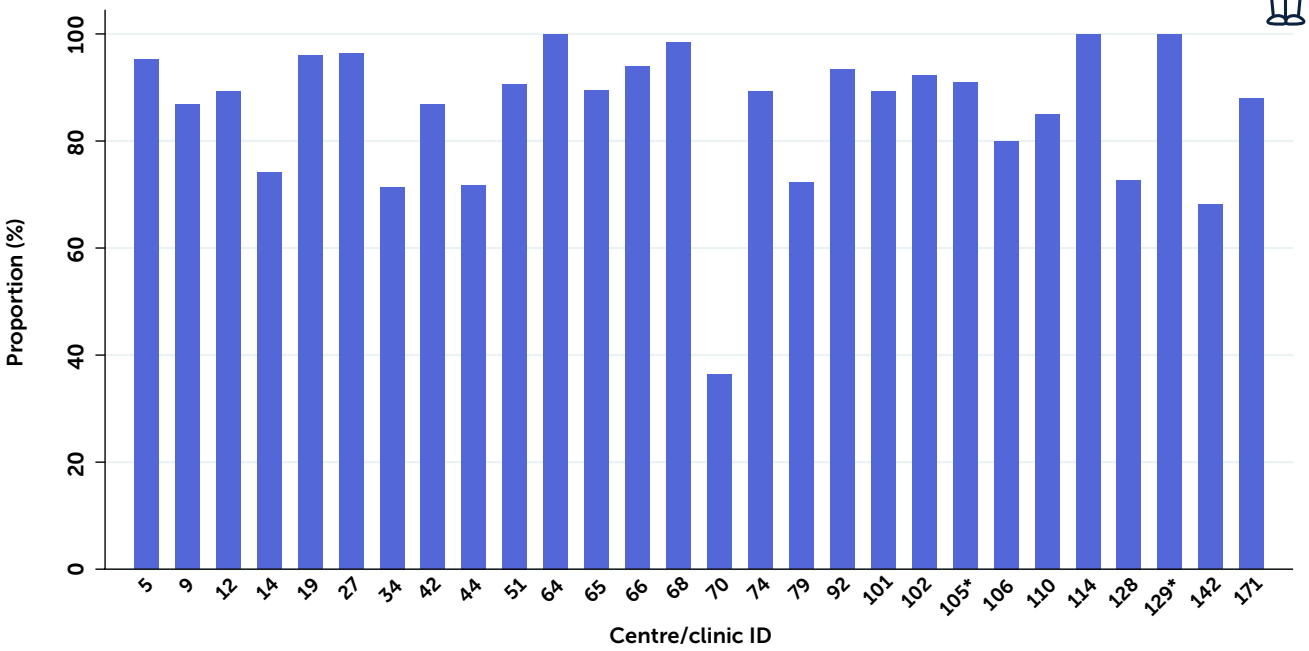
The chart below shows the proportion of patients with at least one IV day at home and/or in hospital. Patients may have a combination of home and hospital IV days.




The proportion of patients in adult centres receiving IV antibiotics at home was 17.6% and in hospital was 18.4%. The proportion receiving any IVs was 27.5%.

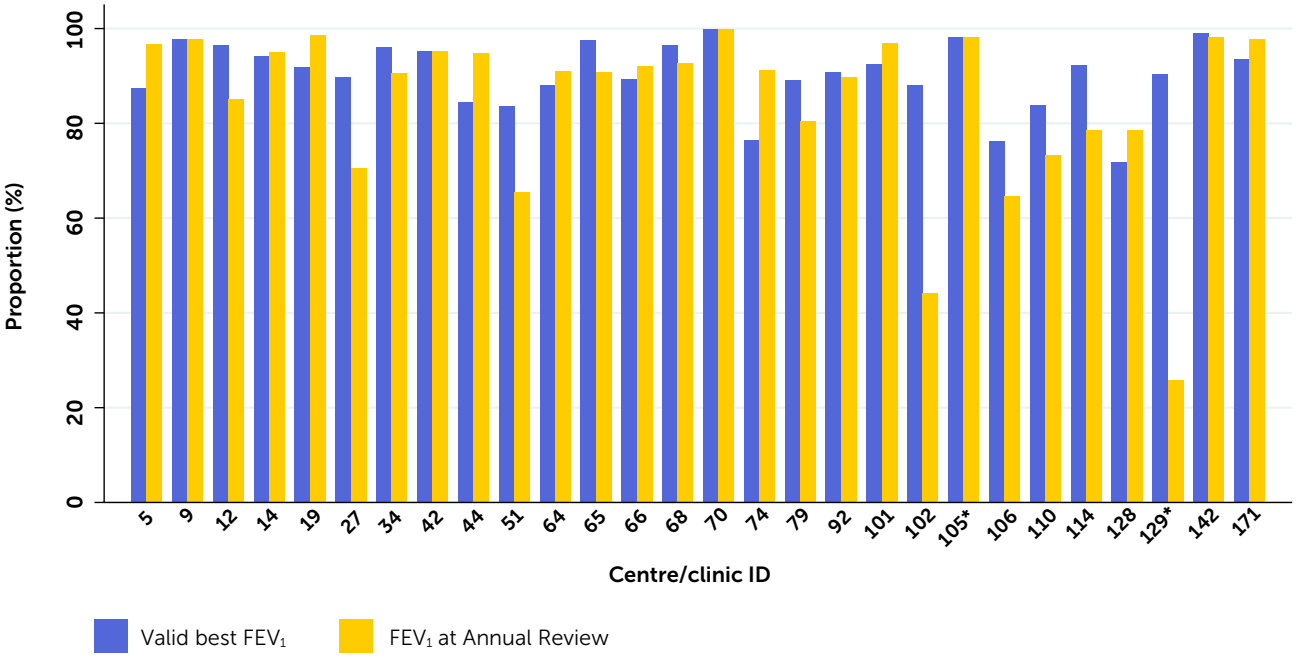
*Stand-alone clinics

## 3.10 Inhaled antibiotic use for patients with chronic *Pseudomonas aeruginosa* by adult centre / clinic




85.6% of patients in adult centres with chronic *P. aeruginosa* received inhaled antibiotics. Centres with fewer than 10 people with chronic *P. aeruginosa* were excluded.

## 3.11 Data completeness by adult centre / clinic*




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

# Glossary

| Word/Phrase | Meaning |
|---|---|
| **2021** | 1 January 2021 – 31 December 2021. |
| **ABPA (allergic bronchopulmonary aspergillosis)** | When a person develops a respiratory allergic reaction to *Aspergillus fumigatus*. |
| **Arthritis** | A condition causing pain and inflammation in the joints. |
| **Arthropathy** | A condition causing pain in the joints. |
| **Asthma** | A respiratory condition causing reversible episodes of difficulty breathing, often associated with wheezing. |
| ***B. cepacia* complex** | The *Burkholderia cepacia* complex is a group of bacteria, some of which threaten the health of people with cystic fibrosis. |
| **BMI (Body Mass Index)** | A measure designed to show whether a person is a healthy weight for their height. |
| **CF** | Cystic fibrosis. |
| **CFTR (cystic fibrosis transmembrane conductance regulator)** | A protein at the cell surface that controls the salt and water balance across a cell. The gene that causes cystic fibrosis is the blueprint for the CFTR protein. Everyone has two copies of the gene for CFTR. To be born with cystic fibrosis, both CFTR genes must be affected by a CF-causing mutation. |
| **Chronic** | Persistent, or long-lasting. |
| **Cirrhosis** | A chronic liver disease. |
| **CI (confidence interval)** | A way of expressing how certain we are about our statistical estimates of a clinical measure (eg BMI). It gives a range of results that is likely to include the 'true' value for the population. A narrow confidence interval indicates a more precise estimate. A wide confidence interval indicates more uncertainty about the true value of the clinical measure, often because a small group of patients has been studied. The confidence interval is usually stated as '95% CI', which means that the range of values has a 95 in 100 chance of including the 'true' value. |
| **Enzymes** | Biological molecules that help complex reactions, such as the digestion of food, occur in the body. |
| **$FEV_1$ (forced expiratory volume in one second)** | This is the amount of air that a person can blow out of the lungs in the first second of a forced exhaled breath. People with healthy lungs can blow out most of the air held in this time. |
| **$FEV_1$% predicted** | The $FEV_1$ can be converted from absolute litres of air blown out into a predicted percentage (%). A healthy range for % predicted is calculated from a very large population sample, and is normally considered to be between 80-120% predicted. |
| **Fibrosing colonopathy** | A condition causing narrowing of part of the colon. |
| **Gall bladder** | The small sac-shaped organ under the liver that stores bile after it is secreted by the liver, before it is released into the intestine. |
| **Gastrointestinal (GI) tract** | The GI tract is an organ system responsible for digesting food, absorbing nutrients and expelling waste. |
| **Genotype** | Part of the genetic makeup of a cell, organism or individual that usually controls a particular characteristic (known as a phenotype). |
| **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 $FEV_1$, which 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. |
| **Median predicted survival age** | A prediction of how long we expect half of the people with CF born today live for. |
| **MRSA** | Methicillin-resistant *Staphylococcus aureus* is a type of bacteria that is resistant to a number of widely used antibiotics. |
| **Mutation** | A mutation is a change in a gene. When both of a child's parents are carriers of a CF-causing mutation there is a 25% chance that the child will have cystic fibrosis. There are over 1,400 different mutations of the CFTR gene that can cause cystic fibrosis. |
| **Nasal polyps** | Small, sac-like growths of inflamed mucus membrane caused by chronic inflammation of the nasal lining. |
| **NBS (newborn screening)** | Newborn screening is part of the heel prick blood spot testing carried out on all babies at 5-7 days of age. The blood sample is tested for a number of conditions, including cystic fibrosis. |
| **NTM (non-tuberculous mycobacteria)** | A mycobacterium that does not cause tuberculosis, but which can cause respiratory infection. There are several 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; 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 on 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, whilst 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 type of bacteria 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 there is statistical evidence that the results we observe (such as a difference in median predicted survival age between males and females) are unlikely to have occurred due to chance. |

# Appendix 1: UK CF Registry Committee structure

## UK CF Registry Steering Committee

| Role | Forename | Surname | Organisation |
|---|---|---|---|
| Commissioner, England | Kathy | Blacker | NHS England |
| CF physician - Paediatrics | Malcolm | Brodlie | Newcastle Paediatric CF Centre |
| CF physician – Paediatrics* | Siobhán | Carr | Royal Brompton Hospital |
| Analytical team rep † | Susan | Charman | Cystic Fibrosis Trust |
| Head of Healthcare Data and Pharmacovigilance # | Sarah | Clarke | Cystic Fibrosis Trust |
| Director of Data & Quality Improvement | Rebecca | Cosgriff | Cystic Fibrosis Trust |
| Chair of the Research Committee | Steve | Cunningham | Royal Hospital for Sick Children, Edinburgh |
| CF Physician - Paediatrics | Gwyneth | Davies | UCL Great Ormond Street Institute of Child Health |
| CF Centre Data Manager | Lance | Dennard | Lewisham Hospital, London |
| CF physician – Adults | Jamie | Duckers | All Wales Adult CF Centre, Cardiff |
| Registry data manager † | Elaine | Gunn | Cystic Fibrosis Trust |
| Allied Health Professional | Rebecca | Heise | Kings College Adult CF Centre |
| CF Centre Data Manager | Erin | Hodgetts | North West Midlands Adults & Paediatric CF Centres |
| Person with CF | Flora | Kennedy-McConnell | N/A |
| CF physician - Adults | Simon | Range | Leicester Adult CF Centre |
| Commissioner, Wales | Andrea | Richards | Welsh Commissioning Board |
| Commissioner, Scotland | David | Steele | NHS Scotland |
| Parent representative | Vacant | | |
| Registry development manager † | Mary | Yip | Cystic Fibrosis Trust |

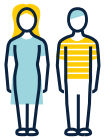
*Chair † Non-voting member # Caldicott guardian

## UK CF Registry Research Committee

| Role | Forename | Surname | Organisation |
|---|---|---|---|
| Pharmacovigilance PI | Diana | Bilton | Royal Brompton Hospital, London |
| Pharmacovigilance PI, CF physician – paediatrics | Siobhán | Carr | Royal Brompton Hospital, London |
| Analytical team rep † | Susan | Charman | Cystic Fibrosis Trust |
| Head of Healthcare Data & Pharmacovigilance # | Sarah | Clarke | Cystic Fibrosis Trust |
| Director of Data & QI | Rebecca | Cosgriff | Cystic Fibrosis Trust |
| Pharmacovigilance PI, CF physician – paediatrics * | Steve | Cunningham | Royal Hospital for Sick Children, Edinburgh |
| Parent Representative | Marian | Dmochowska | N/A |
| Registry data manager† | Elaine | Gunn | Cystic Fibrosis Trust |
| Pharmacovigilance PI, CF physician - Adults | Dilip | Nazareth | Liverpool Heart and Chest Hospital, Liverpool |
| Pharmacovigilance PI , CF physician - Adults | Nick | Simmonds | Royal Brompton Hospital, London |
| Person with CF | James | Thompson | N/A |
| Registry development manager † | Mary | Yip | Cystic Fibrosis Trust |

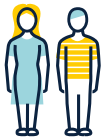
*Chair † Non-voting member # Caldicott guardian

# Appendix 2: Centre-level data tables



## Paediatric centres/clinics providing data in 2021 – ordered alphabetically by country/city

| Location | Name | Clinic ID | Total Active | Number with annual review |
|---|---|---|---|---|
| **England** | | | | |
| Birmingham | Birmingham Children's Hospital | 104 | 314 | 297 |
| Brighton | Royal Alexandra Children's Hospital | 172 | 55 | 40 |
| Bristol | Bristol Royal Hospital for Children | 32 | 187 | 176 |
| Cambridge | Addenbrooke's Hospital | 107 | 147 | 139 |
| Cornwall | Royal Cornwall Hospital | 94 | 37 | 30 |
| Exeter | Royal Devon & Exeter Hospital | 96 | 74 | 69 |
| Hull | Hull University Teaching Hospitals | 111 | 43 | 39 |
| Leeds | St James's University Hospital | 25 | 230 | 223 |
| Leicester | Leicester Royal Infirmary | 1 | 64 | 59 |
| Liverpool | Alder Hey Children's Hospital | 97 | 308 | 296 |
| London - Central | Great Ormond Street Hospital for Children | 90 | 197 | 183 |
| London - East | Royal London Hospital | 30 | 92 | 85 |
| London - South East | King's College Hospital | 17 | 194 | 180 |
| London - South West | Royal Brompton Hospital | 15 | 293 | 266 |
| Manchester | Royal Manchester Children's Hospital | 144 | 339 | 319 |
| Newcastle | Great North Children's Hospital | 59 | 194 | 181 |
| North West Midlands | University Hospital of North Midlands | 8 | 95 | 92 |
| Norwich | Norfolk & Norwich University Hospital | 98 | 66 | 63 |
| Nottingham | Nottingham University Hospitals | 62 | 165 | 156 |
| Oxford | John Radcliffe Hospital | 22 | 165 | 160 |
| Plymouth | Derriford Hospital | 139 | 41 | 40 |
| Sheffield | Sheffield Children's Hospital | 3 | 147 | 142 |
| Southampton | Southampton General Hospital | 29 | 235 | 227 |
| Teeside | James Cook University Hospital | 71 | 52 | 47 |
| **Northern Ireland** | | | | |
| Belfast | Royal Belfast Hospital for Sick Children | 60 | 212 | 196 |
| **Scotland** | | | | |
| Aberdeen | Royal Aberdeen Children's Hospital | 75 | 26 | 24 |
| Ayr | University Hospital Crosshouse | 170 | 23 | 21 |
| Dundee | Ninewells Hospital | 73 | 23 | 22 |
| Edinburgh | Royal Hospital for Sick Children | 143 | 137 | 112 |
| Glasgow | Royal Hospital for Sick Children | 56 | 164 | 142 |
| Inverness | Raigmore Hospital | 31 | 16 | 13 |
| **Wales** | | | | |
| Cardiff | Children's Hospital for Wales | 72 | 162 | 148 |



| | Age | | FEV$_1$% predicted at annual review | | | | Best FEV$_1$% predicted | | | |
|---|---|---|---|---|---|---|---|---|---|---|
| Clinic ID | Mean | Median | Number | Mean - unadjusted | Mean - adjusted | Median | Number* | Mean - unadjusted | Mean - adjusted | Median |
| | | | | | | | | | | |
| 104 | 9.2 | 8.9 | 182 | 90.1 | 90.1 | 92.6 | 200 | 95.9 | 95.9 | 97.3 |
| 172 | 8.6 | 8.5 | 28 | 101.9 | 101.6 | 103.2 | 29 | 105.6 | 105.3 | 105.5 |
| 32 | 9.2 | 9.1 | 116 | 91.7 | 91.6 | 92.9 | 122 | 96.7 | 96.6 | 96.3 |
| 107 | 8.6 | 9.0 | 90 | 94.6 | 94.4 | 95.8 | 93 | 97.3 | 97.0 | 97.2 |
| 94 | 10.3 | 9.5 | 11 | 78.5 | 78.6 | 78.6 | 22 | 91.0 | 91.0 | 90.9 |
| 96 | 9.5 | 9.0 | 47 | 85.2 | 85.0 | 88.1 | 51 | 89.8 | 89.6 | 93.6 |
| 111 | 9.1 | 9.4 | 25 | 86.9 | 86.7 | 88.1 | 28 | 92.0 | 91.9 | 93.6 |
| 25 | 9.1 | 9.3 | 138 | 90.4 | 90.3 | 93.5 | 150 | 96.7 | 96.5 | 96.5 |
| 1 | 8.0 | 8.3 | 38 | 89.4 | 89.1 | 90.5 | 38 | 96.3 | 96.0 | 96.8 |
| 97 | 9.4 | 9.3 | 152 | 89.2 | 89.2 | 90.8 | 176 | 94.0 | 94.0 | 93.8 |
| 90 | 8.8 | 9.1 | 121 | 93.1 | 93.0 | 93.5 | 124 | 97.7 | 97.5 | 97.6 |
| 30 | 9.9 | 10.7 | 63 | 97.6 | 97.6 | 98.1 | 65 | 100.7 | 100.6 | 101.3 |
| 17 | 8.3 | 7.9 | 87 | 91.1 | 91.0 | 93.4 | 107 | 93.5 | 93.3 | 95.8 |
| 15 | 9.1 | 9.1 | 170 | 92.9 | 92.8 | 94.5 | 188 | 98.9 | 98.7 | 99.2 |
| 144 | 9.1 | 9.3 | 129 | 92.4 | 92.4 | 94.7 | 168 | 97.8 | 97.8 | 98.0 |
| 59 | 8.5 | 8.8 | 115 | 94.4 | 94.3 | 97.4 | 122 | 97.7 | 97.5 | 97.7 |
| 8 | 9.8 | 10.7 | 36 | 88.9 | 89.0 | 88.8 | 60 | 95.0 | 95.0 | 96.5 |
| 98 | 8.9 | 9.0 | 33 | 88.4 | 88.4 | 85.0 | 41 | 98.3 | 98.3 | 95.6 |
| 62 | 10.1 | 10.0 | 119 | 87.4 | 87.4 | 87.7 | 120 | 93.8 | 93.8 | 94.7 |
| 22 | 9.1 | 9.5 | 107 | 96.3 | 96.3 | 95.5 | 109 | 100.4 | 100.3 | 100.2 |
| 139 | 8.2 | 8.5 | 25 | 91.4 | 91.2 | 95.9 | 25 | 96.1 | 95.9 | 99.4 |
| 3 | 9.3 | 9.3 | 104 | 94.2 | 94.1 | 95.3 | 106 | 109.5 | 109.4 | 101.0 |
| 29 | 9.1 | 9.1 | 137 | 92.6 | 92.6 | 94.9 | 148 | 99.2 | 99.2 | 100.0 |
| 71 | 9.7 | 10.8 | 29 | 91.5 | 91.5 | 91.4 | 32 | 97.5 | 97.6 | 95.6 |
| | | | | | | | | | | |
| 60 | 9.1 | 9.3 | 119 | 94.3 | 94.3 | 94.8 | 132 | 96.9 | 96.9 | 96.7 |
| | | | | | | | | | | |
| 75 | 7.8 | 7.9 | 13 | 95.0 | 95.0 | 97.6 | 13 | 101.5 | 101.3 | 100.3 |
| 170 | 10.6 | 10.1 | 16 | 92.9 | 92.9 | 98.7 | 17 | 102.1 | 102.0 | 103.3 |
| 73 | 8.7 | 9.7 | 12 | 97.8 | 97.6 | 95.4 | 12 | 99.9 | 99.7 | 99.7 |
| 143 | 9.5 | 10.0 | 78 | 93.9 | 93.9 | 92.5 | 79 | 96.7 | 96.7 | 96.9 |
| 56 | 9.1 | 9.8 | 84 | 93.9 | 93.8 | 92.7 | 101 | 99.2 | 99.1 | 98.4 |
| 31 | 8.7 | 7.9 | 8 | 95.7 | 95.7 | 92.7 | 8 | 98.7 | 98.6 | 97.1 |
| | | | | | | | | | | |
| 72 | 9.1 | 10.1 | 92 | 89.0 | 89.0 | 89.2 | 92 | 93.7 | 93.7 | 93.9 |

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

| | | | BMI | | | |
|---|---|---|---|---|---|---|
| Location | Name | Clinic ID | Number | Mean - unadjusted | Mean - adjusted | Median |
| **England** | | | | | | |
| Birmingham | Birmingham Children's Hospital | 104 | 257 | 55.2 | 55.2 | 56.0 |
| Brighton | Royal Alexandra Children's Hospital | 172 | 36 | 52.8 | 52.8 | 52.3 |
| Bristol | Bristol Royal Hospital for Children | 32 | 161 | 57.1 | 57.1 | 61.4 |
| Cambridge | Addenbrooke's Hospital | 107 | 124 | 60.3 | 60.3 | 60.2 |
| Cornwall | Royal Cornwall Hospital | 94 | 27 | 71.4 | 71.4 | 75.8 |
| Exeter | Royal Devon & Exeter Hospital | 96 | 64 | 58.0 | 58.0 | 63.7 |
| Hull | Hull University Teaching Hospitals | 111 | 36 | 58.6 | 58.6 | 57.8 |
| Leeds | St James's University Hospital | 25 | 189 | 58.9 | 58.9 | 63.8 |
| Leicester | Leicester Royal Infirmary | 1 | 53 | 53.0 | 53.0 | 56.3 |
| Liverpool | Alder Hey Children's Hospital | 97 | 255 | 60.4 | 60.3 | 65.1 |
| London - Central | Great Ormond Street Hospital for Children | 90 | 170 | 51.1 | 51.1 | 50.7 |
| London - East | Royal London Hospital | 30 | 80 | 54.4 | 54.4 | 53.1 |
| London - South East | King's College Hospital | 17 | 155 | 56.9 | 56.9 | 57.2 |
| London - South West | Royal Brompton Hospital | 15 | 245 | 58.5 | 58.5 | 58.3 |
| Manchester | Royal Manchester Children's Hospital | 144 | 203 | 54.0 | 54.0 | 53.9 |
| Newcastle | Great North Children's Hospital | 59 | 153 | 63.2 | 63.2 | 70.4 |
| North West Midlands | University Hospital of North Midlands | 8 | 48 | 55.7 | 55.7 | 59.2 |
| Norwich | Norfolk & Norwich University Hospital | 98 | 57 | 62.8 | 62.8 | 66.2 |
| Nottingham | Nottingham University Hospitals | 62 | 146 | 54.2 | 54.2 | 56.1 |
| Oxford | John Radcliffe Hospital | 22 | 148 | 58.5 | 58.5 | 57.9 |
| Plymouth | Derriford Hospital | 139 | 36 | 59.2 | 59.2 | 64.9 |
| Sheffield | Sheffield Children's Hospital | 3 | 132 | 59.3 | 59.3 | 61.0 |
| Southampton | Southampton General Hospital | 29 | 202 | 59.4 | 59.4 | 64.3 |
| Teeside | James Cook University Hospital | 71 | 42 | 68.2 | 68.2 | 76.7 |
| **Northern Ireland** | | | | | | |
| Belfast | Royal Belfast Hospital for Sick Children | 60 | 175 | 59.3 | 59.3 | 65.4 |
| **Scotland** | | | | | | |
| Aberdeen | Royal Aberdeen Children's Hospital | 75 | 23 | 57.2 | 57.3 | 61.4 |
| Ayr | University Hospital Crosshouse | 170 | 20 | 70.0 | 70.0 | 74.5 |
| Dundee | Ninewells Hospital | 73 | 16 | 57.7 | 57.7 | 62.2 |
| Edinburgh | Royal Hospital for Sick Children | 143 | 102 | 57.9 | 57.9 | 60.3 |
| Glasgow | Royal Hospital for Sick Children | 56 | 118 | 56.5 | 56.5 | 63.1 |
| Inverness | Raigmore Hospital | 31 | 11 | 63.9 | 63.9 | 65.0 |
| **Wales** | | | | | | |
| Cardiff | Children's Hospital for Wales | 72 | 137 | 60.3 | 60.3 | 65.8 |

| | Chronic *pseudomonas* | | Having at least 1 IV days | | Receiving DNase treatment | | Receiving hypertonic saline or mannitol treatment | | Inhaled antibiotic use among patients with chronic *pseudomonas* | |
|---|---|---|---|---|---|---|---|---|---|---|
| Clinic ID | Number | Proportion (%) | Number | Proportion (%) | Number | Proportion (%) | Number | Proportion (%) | Number | Proportion (%) |
| | | | | | | | | | | |
| 104 | 11 | 3.7 | 64 | 21.5 | 173 | 58.2 | 126 | 42.4 | 10 | 90.9 |
| 172 | <5 | -* | 11 | 27.5 | 28 | 70.0 | 10 | 25.0 | <5 | 100.0 |
| 32 | 5 | 2.8 | 25 | 14.2 | 137 | 77.8 | 115 | 65.3 | 5 | 100.0 |
| 107 | 5 | 3.6 | 23 | 16.5 | 76 | 54.7 | 90 | 64.7 | <5 | 80.0 |
| 94 | 0 | 0.0 | <5 | -* | 19 | 63.3 | 15 | 50.0 | 0 | 0.0 |
| 96 | <5 | -* | 7 | 10.1 | 62 | 89.9 | 61 | 88.4 | <5 | 100.0 |
| 111 | 7 | 17.9 | 13 | 33.3 | 20 | 51.3 | 12 | 30.8 | <5 | 42.9 |
| 25 | 15 | 6.7 | 45 | 20.2 | 143 | 64.1 | 23 | 10.3 | 15 | 100.0 |
| 1 | <5 | -* | 12 | 20.3 | 38 | 64.4 | 10 | 16.9 | <5 | 100.0 |
| 97 | 8 | 2.7 | 67 | 22.6 | 168 | 56.8 | 54 | 18.2 | 8 | 100.0 |
| 90 | 8 | 4.4 | 49 | 26.8 | 133 | 72.7 | 84 | 45.9 | 8 | 100.0 |
| 30 | 8 | 9.4 | 25 | 29.4 | 64 | 75.3 | 71 | 83.5 | 8 | 100.0 |
| 17 | 18 | 10.0 | 39 | 21.7 | 123 | 68.3 | 56 | 31.1 | 18 | 100.0 |
| 15 | 19 | 7.1 | 54 | 20.3 | 209 | 78.6 | 130 | 48.9 | 19 | 100.0 |
| 144 | 23 | 7.2 | 56 | 17.6 | 172 | 53.9 | 119 | 37.3 | 20 | 87.0 |
| 59 | 11 | 6.1 | 41 | 22.7 | 106 | 58.6 | 37 | 20.4 | 11 | 100.0 |
| 8 | 7 | 7.6 | 28 | 30.4 | 62 | 67.4 | 25 | 27.2 | 6 | 85.7 |
| 98 | 0 | 0.0 | <5 | -* | 39 | 61.9 | 12 | 19.0 | 0 | 0.0 |
| 62 | <5 | -* | 30 | 19.2 | 108 | 69.2 | 53 | 34.0 | <5 | 100.0 |
| 22 | 7 | 4.4 | 35 | 21.9 | 110 | 68.8 | 58 | 36.3 | 5 | 71.4 |
| 139 | 0 | 0.0 | 8 | 20.0 | 29 | 72.5 | 12 | 30.0 | 0 | 0.0 |
| 3 | 6 | 4.2 | 47 | 33.1 | 97 | 68.3 | 42 | 29.6 | 6 | 100.0 |
| 29 | 7 | 3.1 | 38 | 16.7 | 145 | 63.9 | 46 | 20.3 | 5 | 71.4 |
| 71 | <5 | -* | 5 | 10.6 | 29 | 61.7 | 7 | 14.9 | <5 | 100.0 |
| | | | | | | | | | | |
| 60 | 6 | 3.1 | 20 | 10.2 | 158 | 80.6 | 21 | 10.7 | 6 | 100.0 |
| | | | | | | | | | | |
| 75 | <5 | -* | 7 | 29.2 | 11 | 45.8 | <5 | -* | <5 | 100.0 |
| 170 | <5 | -* | <5 | -* | <5 | -* | 8 | 38.1 | <5 | 100.0 |
| 73 | <5 | -* | <5 | -* | 8 | 36.4 | <5 | -* | <5 | 50.0 |
| 143 | 6 | 5.4 | 26 | 23.2 | 64 | 57.1 | 19 | 17.0 | 6 | 100.0 |
| 56 | <5 | -* | 27 | 19.0 | 35 | 24.6 | 60 | 42.3 | 0 | 0.0 |
| 31 | <5 | -* | <5 | 7.7 | 6 | 46.2 | <5 | -* | <5 | 100.0 |
| | | | | | | | | | | |
| 72 | 7 | 4.7 | 9 | 6.1 | 110 | 74.3 | 136 | 91.9 | 7 | 100.0 |

* Redacted to adhere to statistical disclosure guidelines.

# Appendix 2: Centre-level data tables

## Adult centres/clinics providing data in 2021 – ordered alphabetically by country/city

| Location | Name | Clinic ID | Total active | Number with annual review |
|---|---|---|---|---|
| **England** | | | | |
| Birmingham | Birmingham Heartlands Hospital | 27 | 323 | 302 |
| Bristol | Bristol Royal Infirmary | 106 | 239 | 227 |
| Cambridge | The Royal Papworth Hospital | 51 | 350 | 325 |
| Cornwall | Royal Cornwall Hospital | 129 | 38 | 31 |
| Exeter | Royal Devon & Exeter Hospital | 34 | 133 | 129 |
| Frimley | Frimley Park Hospital | 19 | 151 | 149 |
| Leeds | St James's University Hospital | 42 | 410 | 403 |
| Leicester | Glenfield Hospital | 142 | 117 | 116 |
| Liverpool | Liverpool Heart and Chest Hospital | 66 | 361 | 345 |
| London - East | St. Bartholomew's Hospital | 92 | 215 | 195 |
| London - South East | University Hospital Lewisham | 105 | 55 | 54 |
| London - South East | King's College Hospital | 5 | 258 | 246 |
| London - South West | Royal Brompton Hospital | 12 | 578 | 561 |
| Manchester | Wythenshawe Hospital | 102 | 472 | 448 |
| Newcastle | Royal Victoria Infirmary | 9 | 326 | 316 |
| North West Midlands | University Hospital of North Midlands | 74 | 150 | 149 |
| Norwich | Norfolk & Norwich University Hospital | 114 | 82 | 79 |
| Nottingham | Nottingham University Hospitals | 101 | 239 | 228 |
| Oxford | Oxford University Hospitals | 128 | 149 | 131 |
| Plymouth | Derriford Hospital | 64 | 67 | 67 |
| Sheffield | Northern General Hospital | 65 | 214 | 206 |
| Southampton | Southampton General Hospital | 110 | 291 | 278 |
| York | York Hospital | 171 | 93 | 93 |
| **Northern Ireland** | | | | |
| Belfast | Belfast City Hospital | 14 | 273 | 205 |
| **Scotland** | | | | |
| Aberdeen | Aberdeen Royal Infirmary | 70 | 74 | 72 |
| Edinburgh | Western General Hospital | 44 | 243 | 233 |
| Glasgow | Queen Elizabeth University Hospital | 79 | 219 | 138 |
| **Wales** | | | | |
| Llandough | Llandough Hospital | 68 | 292 | 262 |

| | Age | | FEV$_1$% predicted at annual review | | | | Best FEV$_1$% predicted | | | |
|---|---|---|---|---|---|---|---|---|---|---|
| Clinic ID | Mean | Median | Number | Mean - unadjusted | Mean - adjusted | Median | Number* | Mean - unadjusted | Mean - adjusted | Median |
| | | | | | | | | | | |
| 27 | 34.4 | 32.0 | 199 | 72.1 | 72.9 | 74.5 | 262 | 72.9 | 73.4 | 74.5 |
| 106 | 32.3 | 31.0 | 143 | 72.9 | 72.6 | 76.6 | 185 | 75.8 | 75.5 | 78.6 |
| 51 | 32.6 | 30.4 | 197 | 74.4 | 73.8 | 75.1 | 278 | 75.7 | 75.2 | 74.7 |
| 129 | 35.5 | 31.8 | 8 | 71.4 | 75.0 | 70.1 | 28 | 72.0 | 72.5 | 71.4 |
| 34 | 33.8 | 30.9 | 112 | 77.4 | 78.0 | 79.6 | 120 | 82.2 | 82.5 | 87.6 |
| 19 | 32.7 | 31.9 | 139 | 73.9 | 73.5 | 76.8 | 139 | 76.0 | 75.6 | 77.9 |
| 42 | 35.5 | 33.6 | 347 | 70.3 | 71.2 | 73.8 | 358 | 73.7 | 74.7 | 77.9 |
| 142 | 31.2 | 28.8 | 107 | 72.1 | 70.3 | 78.4 | 108 | 74.9 | 73.0 | 79.6 |
| 66 | 32.3 | 30.8 | 311 | 76.8 | 76.4 | 79.1 | 323 | 78.7 | 78.3 | 81.1 |
| 92 | 29.2 | 25.9 | 172 | 74.0 | 71.3 | 76.2 | 181 | 77.3 | 74.4 | 80.0 |
| 105 | 32.9 | 31.0 | 44 | 65.7 | 65.5 | 64.9 | 44 | 69.9 | 69.7 | 71.2 |
| 5 | 31.8 | 29.8 | 227 | 72.8 | 72.1 | 76.0 | 229 | 76.1 | 75.3 | 82.6 |
| 12 | 35.9 | 34.2 | 476 | 72.2 | 73.8 | 72.2 | 547 | 76.2 | 77.5 | 76.2 |
| 102 | 33.2 | 31.0 | 198 | 67.5 | 67.6 | 67.2 | 407 | 72.5 | 72.9 | 72.9 |
| 9 | 32.2 | 29.9 | 289 | 71.1 | 70.1 | 73.6 | 292 | 73.6 | 72.6 | 77.3 |
| 74 | 30.9 | 27.3 | 129 | 71.4 | 69.0 | 77.3 | 133 | 74.7 | 72.5 | 81.1 |
| 114 | 30.9 | 29.1 | 58 | 78.0 | 77.3 | 82.7 | 72 | 81.9 | 80.6 | 85.4 |
| 101 | 31.6 | 30.4 | 205 | 73.0 | 72.0 | 75.3 | 208 | 77.1 | 76.1 | 79.4 |
| 128 | 32.4 | 28.3 | 96 | 73.5 | 71.7 | 73.4 | 105 | 76.0 | 74.2 | 76.4 |
| 64 | 34.9 | 34.1 | 60 | 75.9 | 76.3 | 78.4 | 62 | 78.7 | 78.7 | 81.0 |
| 65 | 32.4 | 30.7 | 180 | 79.7 | 79.5 | 83.3 | 196 | 81.4 | 81.0 | 85.9 |
| 110 | 34.3 | 31.3 | 191 | 71.3 | 71.3 | 72.4 | 229 | 75.3 | 75.1 | 76.5 |
| 171 | 34.6 | 31.3 | 89 | 70.6 | 71.1 | 70.7 | 91 | 77.1 | 77.4 | 79.4 |
| | | | | | | | | | | |
| 14 | 35.8 | 32.3 | 191 | 71.5 | 72.3 | 76.0 | 197 | 73.1 | 73.9 | 77.1 |
| | | | | | | | | | | |
| 70 | 35.1 | 33.3 | 63 | 70.7 | 71.0 | 68.7 | 63 | 73.0 | 73.2 | 73.0 |
| 44 | 34.7 | 31.9 | 206 | 72.7 | 72.8 | 73.6 | 213 | 75.1 | 75.3 | 78.0 |
| 79 | 35.8 | 32.5 | 108 | 65.4 | 67.3 | 66.7 | 130 | 69.3 | 71.0 | 71.0 |
| | | | | | | | | | | |
| 68 | 32.8 | 30.4 | 229 | 74.4 | 73.8 | 77.0 | 243 | 78.4 | 77.7 | 82.4 |

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

| | | | BMI | | | |
|---|---|---|---|---|---|---|
| Location | Name | Clinic ID | Number | Mean - unadjusted | Mean - adjusted | Median |
| **England** | | | | | | |
| Birmingham | Birmingham Heartlands Hospital | 27 | 290 | 24.5 | 24.4 | 24.2 |
| Bristol | Bristol Royal Infirmary | 106 | 210 | 24.0 | 24.1 | 23.3 |
| Cambridge | The Royal Papworth Hospital | 51 | 304 | 23.7 | 23.7 | 23.2 |
| Cornwall | Royal Cornwall Hospital | 129 | 29 | 24.4 | 24.2 | 21.9 |
| Exeter | Royal Devon & Exeter Hospital | 34 | 127 | 25.2 | 25.1 | 24.4 |
| Frimley | Frimley Park Hospital | 19 | 149 | 23.5 | 23.5 | 23.4 |
| Leeds | St James's University Hospital | 42 | 398 | 24.7 | 24.5 | 24.3 |
| Leicester | Glenfield Hospital | 142 | 116 | 24.0 | 24.2 | 23.8 |
| Liverpool | Liverpool Heart and Chest Hospital | 66 | 342 | 24.5 | 24.5 | 23.9 |
| London - East | St. Bartholomew's Hospital | 92 | 192 | 23.4 | 23.8 | 22.6 |
| London - South East | University Hospital Lewisham | 105 | 54 | 23.3 | 23.3 | 22.5 |
| London - South East | King's College Hospital | 5 | 244 | 24.5 | 24.6 | 24.0 |
| London - South West | Royal Brompton Hospital | 12 | 560 | 23.8 | 23.6 | 23.5 |
| Manchester | Wythenshawe Hospital | 102 | 252 | 23.6 | 23.7 | 23.2 |
| Newcastle | Royal Victoria Infirmary | 9 | 312 | 24.5 | 24.6 | 23.6 |
| North West Midlands | University Hospital of North Midlands | 74 | 146 | 23.9 | 24.1 | 23.6 |
| Norwich | Norfolk & Norwich University Hospital | 114 | 77 | 23.6 | 23.7 | 23.1 |
| Nottingham | Nottingham University Hospitals | 101 | 226 | 23.4 | 23.5 | 23.0 |
| Oxford | Oxford University Hospitals | 128 | 127 | 23.0 | 23.1 | 22.7 |
| Plymouth | Derriford Hospital | 64 | 66 | 24.2 | 24.1 | 24.3 |
| Sheffield | Northern General Hospital | 65 | 205 | 25.3 | 25.4 | 24.2 |
| Southampton | Southampton General Hospital | 110 | 263 | 24.6 | 24.6 | 24.0 |
| York | York Hospital | 171 | 93 | 24.4 | 24.3 | 23.7 |
| **Northern Ireland** | | | | | | |
| Belfast | Belfast City Hospital | 14 | 205 | 25.2 | 25.1 | 24.1 |
| **Scotland** | | | | | | |
| Aberdeen | Aberdeen Royal Infirmary | 70 | 72 | 25.7 | 25.6 | 24.6 |
| Edinburgh | Western General Hospital | 44 | 230 | 24.4 | 24.4 | 23.9 |
| Glasgow | Queen Elizabeth University Hospital | 79 | 129 | 25.2 | 25.1 | 24.9 |
| **Wales** | | | | | | |
| Llandough | Llandough Hospital | 68 | 262 | 24.3 | 24.4 | 23.7 |

| | Chronic *pseudomonas* | | Having at least 1 IV days | | Receiving DNase treatment | | Receiving hypertonic saline or mannitol treatment | | Inhaled antibiotic use among patients with chronic *pseudomonas* | |
|---|---|---|---|---|---|---|---|---|---|---|
| Clinic ID | Number | Proportion (%) | Number | Proportion (%) | Number | Proportion (%) | Number | Proportion (%) | Number | Proportion (%) |
| | | | | | | | | | | |
| 27 | 27 | 8.9 | 96 | 31.8 | 215 | 71.2 | 128 | 42.4 | 26 | 96.3 |
| 106 | 5 | 2.2 | 85 | 37.4 | 157 | 69.2 | 153 | 67.4 | <5 | -* |
| 51 | 32 | 9.8 | 108 | 33.2 | 223 | 68.6 | 172 | 52.9 | 29 | 90.6 |
| 129 | 11 | 35.5 | 15 | 48.4 | 21 | 67.7 | 12 | 38.7 | 11 | 100.0 |
| 34 | 7 | 5.4 | 24 | 18.6 | 89 | 69.0 | 82 | 63.6 | 5 | 71.4 |
| 19 | 50 | 33.6 | 39 | 26.2 | 115 | 77.2 | 71 | 47.7 | 48 | 96.0 |
| 42 | 106 | 26.3 | 134 | 33.3 | 293 | 72.7 | 86 | 21.3 | 92 | 86.8 |
| 142 | 22 | 19.0 | 33 | 28.4 | 75 | 64.7 | 26 | 22.4 | 15 | 68.2 |
| 66 | 33 | 9.6 | 76 | 22.0 | 265 | 76.8 | 117 | 33.9 | 31 | 93.9 |
| 92 | 45 | 23.1 | 55 | 28.2 | 164 | 84.1 | 139 | 71.3 | 42 | 93.3 |
| 105 | 11 | 20.4 | 19 | 35.2 | 37 | 68.5 | 19 | 35.2 | 10 | 90.9 |
| 5 | 21 | 8.5 | 59 | 24.0 | 204 | 82.9 | 120 | 48.8 | 20 | 95.2 |
| 12 | 84 | 15.0 | 142 | 25.3 | 508 | 90.6 | 296 | 52.8 | 75 | 89.3 |
| 102 | 64 | 14.3 | 110 | 24.6 | 301 | 67.2 | 189 | 42.2 | 59 | 92.2 |
| 9 | 76 | 24.1 | 88 | 27.8 | 202 | 63.9 | 65 | 20.6 | 66 | 86.8 |
| 74 | 37 | 24.8 | 48 | 32.2 | 111 | 74.5 | 67 | 45.0 | 33 | 89.2 |
| 114 | 8 | 10.1 | 19 | 24.1 | 55 | 69.6 | 36 | 45.6 | 8 | 100.0 |
| 101 | 28 | 12.3 | 73 | 32.0 | 183 | 80.3 | 118 | 51.8 | 25 | 89.3 |
| 128 | 11 | 8.4 | 27 | 20.6 | 100 | 76.3 | 54 | 41.2 | 8 | 72.7 |
| 64 | 16 | 23.9 | 15 | 22.4 | 44 | 65.7 | 40 | 59.7 | 16 | 100.0 |
| 65 | 38 | 18.4 | 85 | 41.3 | 177 | 85.9 | 45 | 21.8 | 34 | 89.5 |
| 110 | 20 | 7.2 | 72 | 25.9 | 179 | 64.4 | 116 | 41.7 | 17 | 85.0 |
| 171 | 33 | 35.5 | 20 | 21.5 | 76 | 81.7 | 18 | 19.4 | 29 | 87.9 |
| | | | | | | | | | | |
| 14 | 85 | 41.5 | 55 | 26.8 | 142 | 69.3 | 50 | 24.4 | 63 | 74.1 |
| | | | | | | | | | | |
| 70 | 11 | 15.3 | 5 | 6.9 | 39 | 54.2 | 12 | 16.7 | <5 | -* |
| 44 | 39 | 16.7 | 49 | 21.0 | 140 | 60.1 | 45 | 19.3 | 28 | 71.8 |
| 79 | 18 | 13.0 | 35 | 25.4 | 52 | 37.7 | 21 | 15.2 | 13 | 72.2 |
| | | | | | | | | | | |
| 68 | 63 | 24.0 | 64 | 24.4 | 189 | 72.1 | 168 | 64.1 | 62 | 98.4 |

* Redacted to adhere to statistical disclosure guidelines.

# Appendix 3: Full list of mutations in the UK CF 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 | 9645 | 89.0 |
| c.350G->A | p.Arg117His | R117H | 670 | 6.2 |
| c.1652G->A | p.Gly551Asp | G551D | 629 | 5.8 |
| c.1624G->T | p.Gly542X | G542X | 398 | 3.7 |
| c.489+1G->T | | 621+1G->T | 270 | 2.5 |
| c.1585-1G->A | | 1717-1G->A | 169 | 1.6 |
| c.3909C->G | p.Asn1303Lys | N1303K | 169 | 1.6 |
| c.1766+1G->A | | 1898+1G->A | 153 | 1.4 |
| c.200C->T | p.Pro67Leu | P67L | 141 | 1.3 |
| c.3454G->C | p.Asp1152His | D1152H | 141 | 1.3 |
| c.3528delC | p.Lys1177SerfsX15 | 3659delC | 115 | 1.1 |
| c.3140-26A->G | | 3272-26A->G | 109 | 1.0 |
| c.1679G->C | p.Arg560Thr | R560T | 102 | 0.9 |
| c.1519_1521delATC | p.Ile507del | I507del | 91 | 0.8 |
| c.1477C->T | p.Gln493X | Q493X | 89 | 0.8 |
| c.1657C->T | p.Arg553X | R553X | 88 | 0.8 |
| c.3717+12191C->T | | 3849+10kbC->T | 87 | 0.8 |
| c.254G->A | p.Gly85Glu | G85E | 80 | 0.7 |
| c.178G->T | p.Glu60X | E60X | 74 | 0.7 |
| c.1022_1023insTC | p.Phe342HisfsX28 | 1154insTC | 73 | 0.7 |
| c.2657+5G->A | | 2789+5G->A | 72 | 0.7 |
| c.3846G->A | p.Trp1282X | W1282X | 63 | 0.6 |
| c.1646G->A | p.Ser549Asn | S549N | 56 | 0.5 |
| c.948delT | p.Phe316LeufsX12 | 1078delT | 55 | 0.5 |
| c.2052delA | p.Lys684AsnfsX38 | 2184delA | 51 | 0.5 |
| c.1364C->A | p.Ala455Glu | A455E | 49 | 0.5 |
| c.617T->G | p.Leu206Trp | L206W | 46 | 0.4 |
| c.1040G->C | p.Arg347Pro | R347P | 39 | 0.4 |
| c.2657+2_2657+3insA | | 2789+2insA | 38 | 0.4 |
| c.1558G->T | p.Val520Phe | V520F | 34 | 0.3 |
| c.579+3A->G | | 711+3A->G | 33 | 0.3 |
| c.3484C->T | p.Arg1162X | R1162X | 31 | 0.3 |
| c.1000C->T | p.Arg334Trp | R334W | 29 | 0.3 |
| c.1040G->A | p.Arg347His | R347H | 29 | 0.3 |
| c.1753G->T | p.Glu585X | E585X | 27 | 0.2 |
| c.2988+1G->A | | 3120+1G->A | 26 | 0.2 |
| c.1055G->A | p.Arg352Gln | R352Q | 23 | 0.2 |
| c.1210-12[5] (AJ574948.1:g.152T[5]) | | 5T | 23 | 0.2 |

| Nucleotide | Protein | Legacy name | N | % |
|---|---|---|---|---|
| c.3718-2477C->T | | 3849+10kbC->T | 22 | 0.2 |
| c.2583delT | p.Phe861LeufsX3 | 2711delT | 22 | 0.2 |
| c.3472C->T | p.Arg1158X | R1158X | 21 | 0.2 |
| c.1006_1007insG | p.Ile336SerfsX28 | 1138insG | 21 | 0.2 |
| c.2490+1G->A | | 2622+1G->A | 20 | 0.2 |
| c.1705T->G | p.Tyr569Asp | Y569D | 20 | 0.2 |
| c.1367T->C | p.Val456Ala | V456A | 20 | 0.2 |
| c.2125C->T | p.Arg709X | R709X | 19 | 0.2 |
| c.532G->A | p.Gly178Arg | G178R | 19 | 0.2 |
| c.2834C->T | p.Ser945Leu | S945L | 19 | 0.2 |
| c.1393-1G->A | | 1525-1G->A | 19 | 0.2 |
| c.3197G->A | p.Arg1066His | R1066H | 18 | 0.2 |
| c.1523T->G | p.Phe508Cys | F508C | 18 | 0.2 |
| c.3806T->A | p.Ile1269Asn | I1269N | 17 | 0.2 |
| c.2052_2053insA | p.Gln685ThrfsX4 | 2184insA | 16 | 0.1 |
| c.658C->T | p.Gln220X | Q220X | 14 | 0.1 |
| c.2537G->A | p.Trp846X | W846X | 13 | 0.1 |
| c.292C->T | p.Gln98X | Q98X | 13 | 0.1 |
| c.3737C->T | p.Thr1246Ile | T1246I | 12 | 0.1 |
| c.1029delC | p.Cys343X | 1161delC | 12 | 0.1 |
| c.579+1G->T | | 711+1G->T | 12 | 0.1 |
| c.2988G->A | | 3120G->A | 11 | 0.1 |
| c.2875delG | p.Ala959HisfsX9 | 3007delG | 11 | 0.1 |
| c.3705T->G | p.Ser1235Arg | S1235R | 10 | 0.1 |
| c.349C->T | p.Arg117Cys | R117C | 10 | 0.1 |
| c.3208C->T | p.Arg1070Trp | R1070W | 10 | 0.1 |
| c.1466C->A | p.Ser489X | S489X | 10 | 0.1 |
| c.224G->A | p.Arg75Gln | R75Q | 9 | 0.1 |
| c.3196C->T | p.Arg1066Cys | R1066C | 9 | 0.1 |
| c.1675G->A | p.Ala559Thr | A559T | 8 | 0.1 |
| c.3468G->A | | 3600G->A | 8 | 0.1 |
| c.494T->C | p.Leu165Ser | L165S | 8 | 0.1 |
| c.1679+1G->C | | 1811+1G->C | 8 | 0.1 |
| c.695T->A | p.Val232Asp | V232D | 8 | 0.1 |
| c.2012delT | p.Leu671X | 2143delT | 7 | 0.1 |
| c.2051_2052delAAinsG | p.Lys684SerfsX38 | 2183AA->G or 2183de- lAA->G | 7 | 0.1 |
| c.1986_1989delAACT | p.Thr663ArgfsX8 | 2118del4 | 6 | 0.1 |

| Nucleotide | Protein | Legacy name | N | % |
|---|---|---|---|---|
| c.1329_1330insAGAT | p.Ile444ArgfsX3 | 1461ins4 | 6 | 0.1 |
| c.3884_3885insT | p.Ser1297PhefsX5 | 4016insT | 6 | 0.1 |
| c.2128A->T | p.Lys710X | K710X | 6 | 0.1 |
| c.1766+1G->T | | 1898+1G->T | 6 | 0.1 |
| c.1116+1G->A | | 1248+1G->A | 6 | 0.1 |
| c.3761T->G | p.Leu1254X | L1254X | 6 | 0.1 |
| c.4196_4197delTC | p.Cys1400X | 4326delTC | 6 | 0.1 |
| c.2353C->T | p.Arg785X | R785X | 6 | 0.1 |
| c.1721C->A | p.Pro574His | P574H | 6 | 0.1 |
| c.2900T->C | p.Leu967Ser | L967S | 5 | 0.0 |
| c.2551C->T | p.Arg851X | R851X | 5 | 0.0 |
| c.[1210–12[5];1210-34TG[13]] | | 5T;TG13 | 5 | 0.0 |
| c.2290C->T | p.Arg764X | R764X | 5 | 0.0 |
| c.1687T->A | p.Tyr563Asn | Y563N | 5 | 0.0 |
| c.223C->T | p.Arg75X | R75X | 5 | 0.0 |
| c.3848G->T | p.Arg1283Met | R1283M | 5 | 0.0 |
| c.[1210–12[5];1210-34TG[12]] | | 5T;TG12 | 5 | 0.0 |
| c.349C->G | p.Arg117Gly | R117G | 5 | 0.0 |
| c.3718-1G->A | | 3850-1G->A | 5 | 0.0 |
| c.2215delG | p.Val739TyrfsX16 | 2347delG | <5 | - |
| c.3353C->T | p.Ser1118Phe | S1118F | <5 | - |
| c.2249C->T | p.Pro750Leu | P750L | <5 | - |
| c.1393-2A->G | | 1525-2A->G | <5 | - |
| c.2464G->T | p.Glu822X | E822X | <5 | - |
| c.1679G->A | p.Arg560Lys | R560K | <5 | - |
| c.1680A->C | p.Arg560Ser | R560S | <5 | - |
| c.(743+1_744-1)_(1584+1_1585-1)dup | | CFTRdup6b-10 | <5 | - |
| c.3095A->G | p.Tyr1032Cys | Y1032C | <5 | - |
| c.165-3C>T | | 297-3C->T | <5 | - |
| c.595C->T | p.His199Tyr | H199Y | <5 | - |
| c.3292T->C | p.Trp1098Arg | W1098R | <5 | - |
| c.443T->C | p.Ile148Thr | I148T | <5 | - |
| c.1538A->G | p.Asp513Gly | D513G | <5 | - |
| c.850dupA | p.Met284AsnfsX3 | 977insA | <5 | - |
| c.2909G->A | p.Gly970Asp | G970D | <5 | - |
| c.262_263delTT | p.Leu88IlefsX22 | 394delTT | <5 | - |
| c.3988C->T | p.Gln1330X | Q1330X | <5 | - |
| c.1585-8G->A | | 1717-8G->A | <5 | - |
| c.2600_2601insA | p.Val868SerfsX28 | 2732insA | <5 | - |
| c.3080T->C | p.Ile1027Thr | I1027T | <5 | - |
| c.1766+5G->T | | 1898+5G->T | <5 | - |
| c.1340delA | p.Lys447ArgfsX2 | 1471delA | <5 | - |
| c.509G->A | p.Arg170His | R170H | <5 | - |

| Nucleotide | Protein | Legacy name | N | % |
|---|---|---|---|---|
| c.274G->A | p.Glu92Lys | E92K | <5 | - |
| c.1724T->A | p.Phe575Tyr | F575Y | <5 | - |
| c.1736A->G | p.Asp579Gly | D579G | <5 | - |
| c.2260G->A | p.Val754Met | V754M | <5 | - |
| c.1505T->C | p.Ile502Thr | I502T | <5 | - |
| c.2491G->T | p.Glu831X | E831X | <5 | - |
| c.1572C->A | p.Cys524X | C524X | <5 | - |
| c.2896delA | p.Thr966ArgfsX2 | 3028delA | <5 | - |
| c.91C->T | p.Arg31Cys | R31C | <5 | - |
| c.328G->C | p.Asp110His | D110H | <5 | - |
| c.2991G->C | p.Leu997Phe | L997F | <5 | - |
| c.3659delC | p.Thr1220LysfsX8 | 3791delC | <5 | - |
| c.4147_4148insA | p.Ile1383AsnfsX3 | 4279insA | <5 | - |
| c.[1521_1523delCTT;3080T->C] | p.[Phe508del;Ile- 1027Thr] | F508del;I1027T | <5 | - |
| c.350G->T | p.Arg117Leu | R117L | <5 | - |
| c.3700A->G | p.Ile1234Val | I1234V | <5 | - |
| c.577G->T | p.Glu193X | E193X | <5 | - |
| c.4046G->A | p.Gly1349Asp | G1349D | <5 | - |
| c.4004T->C | p.Leu1335Pro | L1335P | <5 | - |
| c.4111G->T | p.Glu1371X | E1371X | <5 | - |
| c.3908delA | p.Asn1303ThrfsX25 | 4040delA | <5 | - |
| c.1001G>A | p.Arg334Gln | R334Q | <5 | - |
| c.3475T->C | p.Ser1159Pro | S1159P | <5 | - |
| c.442delA | p.Ile148LeufsX5 | 574delA | <5 | - |
| c.1766+1G->C | | 1898+1G->C | <5 | - |
| c.3017C->A | p.Ala1006Glu | A1006E | <5 | - |
| c.296C->T | p.Pro99Leu | P99L | <5 | - |
| c.220C->T | p.Arg74Trp | R74W | <5 | - |
| c.[1210–12[5];1210-34TG[11]] | | 5T;TG11 | <5 | - |
| c.1651G->A | p.Gly551Ser | G551S | <5 | - |
| c.1477_1478delCA | p.Gln493ValfsX10 | 1609delCA | <5 | - |
| c.2374C->T | p.Arg792X | R792X | <5 | - |
| c.3872A->G | p.Gln1291Arg | Q1291R | <5 | - |
| c.2195T->G | p.Leu732X | L732X | <5 | - |
| c.1679+1.6kbA->G | | 1811+1.6kbA->G | <5 | - |
| c.164+2T>C | | 296+2T->C | <5 | - |
| c.3266G->A | p.Trp1089X | W1089X | <5 | - |
| c.79G->T | p.Gly27X | G27X | <5 | - |
| c.3752G->A | p.Ser1251Asn | S1251N | <5 | - |
| c.1007T->A | p.Ile336Lys | I336K | <5 | - |
| c.1727G->C | p.Gly576Ala | G576A | <5 | - |
| c.3763T->C | p.Ser1255Pro | S1255P | <5 | - |

| Nucleotide | Protein | Legacy name | N | % |
|---|---|---|---|---|
| c.2780T->C | p.Leu927Pro | L927P | <5 | - |
| c.3882_3885delTATT | p.Ile1295PhefsX32 | 4010del4 | <5 | - |
| c.2668C->T | p.Gln890X | Q890X | <5 | - |
| c.3310G->T | p.Glu1104X | E1104X | <5 | - |
| c.3205G->A | p.Gly1069Arg | G1069R | <5 | - |
| c.1046C->T | p.Ala349Val | A349V | <5 | - |
| c.1327G->T | p.Asp443Tyr | D443Y | <5 | - |
| c.4077_4080delTGTTinsAA | p.Val1360delfsX? | 4209TGTT->AA | <5 | - |
| c.3458T->A | p.Val1153Glu | V1153E | <5 | - |
| c.4231C->T | p.Gln1411X | Q1411X | <5 | - |
| c.2930C->T | p.Ser977Phe | S977F | <5 | - |
| c.3194T->C | p.Leu1065Pro | L1065P | <5 | - |
| c.2989-1G->A | | 3121-1G->A | <5 | - |
| c.1408A->G | p.Met470Val | M470V | <5 | - |
| c.3158C->T | p.Thr1053Ile | T1053I | <5 | - |
| c.613C->T | p.Pro205Ser | P205S | <5 | - |
| c.1573C->T | p.Gln525X | Q525X | <5 | - |
| c.1037T->C | p.Leu346Pro | L346P | <5 | - |
| c.1837G->A | p.Ala613Thr | A613T | <5 | - |
| c.3297C->A | p.Phe1099Leu | F1099L | <5 | - |
| c.3302T->G | p.Met1101Arg | M1101R | <5 | - |
| c.2421A->G | p.Ile807Met | I807M | <5 | - |
| c.3717G->A | | 3849G->A | <5 | - |
| c.53+1G->T | | 185+1G->T | <5 | - |
| c.1A->G | p.Met1Val | M1V | <5 | - |
| c.2645G->A | p.Trp882X | W882X | <5 | - |
| c.233dupT | p.Trp79LeufsX32 | 365-366insT | <5 | - |
| c.1021T->C | p.Ser341Pro | S341P | <5 | - |
| c.601G->A | p.Val201Met | V201M | <5 | - |
| c.3476C->T | p.Ser1159Phe | S1159F | <5 | - |
| c.1209+1G->A | | 1341+1G->A | <5 | - |
| c.3773_3774insT | p.Leu1258PhefsX7 | 3905insT | <5 | - |
| c.1418delG | p.Gly473GlufsX54 | 1548delG | <5 | - |
| c.164+1G>A | | 296+1G->A | <5 | - |
| c.263T>A or c.263T>G | p.Leu88X | L88X | <5 | - |
| c.717delG | p.Leu240X | 849delG | <5 | - |
| c.1703delT | p.Leu568CysfsX4 | 1833delT | <5 | - |
| c.3745G->A | p.Gly1249Arg | G1249R | <5 | - |
| c.413_415dupTAC | p.Leu138dup | L138ins | <5 | - |
| c.274-2A->G | | 406-2A->G | <5 | - |
| c.470_483del14 | p.Phe157X | 602del14 | <5 | - |
| c.1682C->A | p.Ala561Glu | A561E | <5 | - |

| Nucleotide | Protein | Legacy name | N | % |
|---|---|---|---|---|
| c.137C->A | p.Ala46Asp | A46D | <5 | - |
| c.2735C->A | p.Ser912X | S912X | <5 | - |
| c.859_863delAACTT | p.Asn287LysfsX19 | 991del5 | <5 | - |
| c.2620-26A->G | | 2752-26A->G | <5 | - |
| c.1654C->T | p.Gln552X | Q552X | <5 | - |
| c.(53+1_54-1)_(164+1_165-1)del | | CFTRdele2 | <5 | - |
| c.11C>A | p.Ser4X | S4X | <5 | - |
| c.273+1G->A | | 405+1G->A | <5 | - |
| c.1545_1546delTA | p.Tyr515X | 1677delTA | <5 | - |
| c.3718-3T->G | | 3850-3T->G | <5 | - |
| c.50delT | p.Phe17SerfsX8 | 182delT | <5 | - |
| c.987delA | p.Gly330GlufsX39 | 1119delA | <5 | - |
| c.1301_1307delCACTTCT | p.Ser434LeufsX6 | 1429del7 | <5 | - |
| c.1081delT | p.Trp361GlyfsX8 | 1213delT | <5 | - |
| c.3209G->A | p.Arg1070Gln | R1070Q | <5 | - |
| c.1240C->T | p.Gln414X | Q414X | <5 | - |
| c.2739T->A | p.Tyr913X | Y913X | <5 | - |
| c.2002C->T | p.Arg668Cys | R668C | <5 | - |
| c.1117-1G>A | | 1249-1G->A | <5 | - |
| c.3181G->C | p.Gly1061Arg | G1061R | <5 | - |
| c.3717+5G->A | | 3849+5G->A | <5 | - |
| c.3011_3019delCTATAGCAG or c.3009_3017delAGCTATAGC | p.Ala1004_Ala1006del | 3143del9 | <5 | - |
| c.(53+1_54-1)_(489+1_490-1)del | | CFTRdele2-4 | <5 | - |
| c.3485G->T | p.Arg1162Leu | R1162L | <5 | - |
| c.3873+2T->C | | 4005+2T->C | <5 | - |
| c.1687T->G | p.Tyr563Asp | Y563D | <5 | - |
| c.3230T->C | p.Leu1077Pro | L1077P | <5 | - |
| c.2859_2890delACATTCT-GTTCTTCAAGCACCTATGT-CAACCC | p.Leu953PhefsX11 | 2991del32 | <5 | - |
| c.4144C->T | p.Gln1382X | Q1382X | <5 | - |
| c.933_935delCTT | p.Phe312del | F311del | <5 | - |
| `Other' selected | | | 781 | 7.2 |

Cystic Fibrosis Trust

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

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

**cysticfibrosis.org.uk**



**Uniting for a life unlimited**