MONASH University

# ANNUAL REPORT 2025

## AUSTRALIAN CYSTIC FIBROSIS DATA REGISTRY

**Data Extract Period**

The data contained in this report was extracted from the ACFDR on May 4th 2026, and pertains to data related to patient events from January 1st to December 31st 2025. As the registry does not capture data in real time, there can be a lag between the occurrence of an event and its capture in the ACFDR.

**Abbreviations**

| | |
|---|---|
| ABS | Australian Bureau of Statistics |
| ACFDR | Australian Cystic Fibrosis Data Registry |
| AWESCORE | The Alfred Wellness Score |
| BAL | Broncho Alveolar Lavage |
| BMI | Body Mass Index |
| CDC | Centers for Disease Control and Prevention |
| CF | Cystic Fibrosis |
| CFQ-R | Cystic Fibrosis Questionnaire Revised |
| CFTR | Cystic Fibrosis Transmembrane Conductance Regulator |
| CI | Confidence Interval |
| ETI | Elexacaftor/tezacaftor/ivacaftor |
| FEV | Forced Expiratory Volume |
| FEV1 pp | Forced Expiratory Volume (litres) in 1 second percent predicted |
| GOR | Gastroesophageal Reflux |
| GLI | Global Lung Initiative |
| IV | Intravenous |
| MRSA | Methicillin-resistant Staphylococcus aureus |
| NTM | Nontuberculous Mycobacteria |
| PBS | Pharmaceutical Benefits Scheme |
| PROM | Patient-Reported Outcome Measure |
| pwCF | People with Cystic Fibrosis |
| WHO | World Health Organization |

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CYSTIC FIBROSIS Australia

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# FOREWORDS

## FROM THE CYSTIC FIBROSIS AUSTRALIA CEO

It is my pleasure to introduce the 2025 Australian Cystic Fibrosis Data Registry (ACFDR) Annual Report.

Each year, the ACFDR provides invaluable insights into the health and wellbeing of people living with cystic fibrosis across Australia. More than a collection of statistics, the registry tells the story of a community, a healthcare system, and a research effort working together to achieve better outcomes for people with cystic fibrosis.

This year's report reflects a remarkable period in the history of cystic fibrosis care. For the first time, there are now more than 3,900 people living with cystic fibrosis in Australia. This milestone is a testament to decades of scientific discovery, clinical excellence, advocacy, and the determination of people with cystic fibrosis and their families. It also reflects a reality that would have been difficult to imagine only a generation ago, now that more people with cystic fibrosis are living longer, healthier and fuller lives than ever before.

The data presented in this report continues to demonstrate overwhelmingly positive trends across many key health indicators. Improvements in lung function, nutritional outcomes, survival, and quality of life are increasingly evident, particularly following the introduction of highly effective CFTR modulator therapies. These innovative treatments have changed the trajectory of cystic fibrosis for many Australians, offering new hope and new possibilities for the future.

Importantly, the ACFDR allows us to measure this progress. It enables clinicians, researchers, policymakers and advocates to understand what is working, identify emerging challenges and continue striving for better outcomes. The strength of the registry lies not only in the quality of the data it captures, but also in the commitment of the people and healthcare teams who contribute to it each year.

While there is much to celebrate, this report also reminds us that our work is far from complete. Not everyone with cystic fibrosis benefits from currently available modulator therapies. For some individuals, effective treatment options remain limited. Others continue to face significant health challenges and treatment burdens. The data reinforces the importance of continuing to invest in research, innovation, clinical care and equitable access to treatment so that every person with cystic fibrosis can share in the progress being made.

At Cystic Fibrosis Australia, we remain committed to advocating for all people living with cystic fibrosis. We will continue to work alongside researchers, clinicians, governments, industry partners and the broader cystic fibrosis community to drive improvements in care, and support the development of new therapies.

The future of cystic fibrosis is brighter than it has ever been, but our ambition remains unchanged. We want every person with cystic fibrosis to have the opportunity to live a longer, healthier and more fulfilling life.

I would like to thank the many individuals and organisations whose contributions make the ACFDR possible, including people with cystic fibrosis and their families, clinicians, data managers, researchers, healthcare teams and our funding partners. Your commitment continues to advance knowledge, improve care and shape the future of cystic fibrosis in Australia.

Together, we are witnessing extraordinary progress, and we will continue the work that lies ahead.

**Dr Jo Armstrong**
Chief Executive Officer,
Cystic Fibrosis Australia

CYSTIC FIBROSIS Australia

# FROM THE MONASH UNIVERSITY ACFDR LEAD

I am delighted to present the Australian Cystic Fibrosis Data Registry's (ACFDR) 2025 Annual Report, a comprehensive analysis of data relating to the activities and outcomes of people with CF who participate in the ACFDR.

The ACFDR is approaching 4,000 people with CF, mainly due to people with CF living longer than ever before. The registry reported 65 new diagnoses in 2025, and in line with recent years, an increasing proportion of these are over 18 years of age (21.5%). More people with CF live full social lives, with 50% completing a tertiary education and 75% reporting being employed. A further 81 pregnancies were reported for women with CF in 2025.

People with CF continue to break new records, with median FEV1 in adults for the first time being greater than 80pp. Overall infection rates continue to decline, as well as many complications associated with CF. People with CF are spending less time in hospital. Paediatric hospitalisations have reduced from 0.7 per person in 2020 to 0.4 per person in 2025, and 75% of adults reported not having any hospitalisations in 2025. The data is also showing shifts in resourcing requirements – although the number of clinic visits per adult is declining, the total number of adult clinic visits increased in 2025 due to the greater overall adult numbers.

Outcomes for people with CF continue to be reflected in reduced numbers of deaths, and higher predicted survival. However, statistics do not describe the experience of living with CF. Five to six percent of adults with CF are not eligible for a CFTR modulator, and a further 8-9% were eligible but not prescribed a modulator at time of reporting due to switching between modulators or treatment side effects. This means that people with CF require personalised care depending on their age and individual circumstances.

As well as informing clinical care, the ACFDR is a data asset that supports a significant amount of research and quality improvement activity, with four publications, two conference presentations and fifteen data requests in 2025 alone.

I would like to extend my sincere thanks to the ACFDR team, Steering Committee members, participating clinicians and CF centre staff, people with CF, CFA and the broader CF community for your commitment and ongoing support of the Australian Cystic Fibrosis Data Registry.

**Professor Susannah Ahern**
Head, Clinical Outcomes data Reporting and Research,
School of Public Health and Preventive Medicine,
Monash University

## FROM THE REGISTRY CLINICAL LEAD

As Chair of the ACFDR, I always receive the draft annual report with a sense of anticipation. It is a privilege to be involved in this work and to see outcomes for people in Australia living with CF continue to improve. The 2025 report is both encouraging and promising.

The CF community in Australia continues to grow. This report presents data on 3,916 Australians living with CF, with the proportion aged 18 years and older now reaching 62%. We can also report that 150 people identify as Aboriginal and/or Torres Strait Islander. While the overall number of people receiving a new diagnosis of CF remains relatively stable, an increasing proportion are being diagnosed later in life. In 2020, 10.8% of people newly diagnosed with CF were aged 18 years or older; this has now increased to 21.5%. This likely reflects greater awareness of CF in the community and increased access to genetic testing, leading to diagnosis in more people with conditions such as bronchiectasis, chronic sinusitis or fertility concerns. These individuals now have an explanation for their symptoms and may also be eligible for treatment with CFTR modulators.

The steady improvement in lung function also continues. Among children aged 6–17 years, median lung function has now nearly reached the healthy population mean at 98%. For people aged 18 years and older, it has exceeded 80% for the first time. Infection with serious pathogens such as Pseudomonas has also declined substantially, falling from 34.1% in 2020 to 15.3%.

It is not only lung disease that is improving. For the first time, we may be seeing a fall in the prevalence of diabetes, a difficult and significant additional burden for people with CF. Among children aged 12–17 years, the prevalence of diabetes was 21.8% in 2020 and has fallen to 10.7% in 2025. In adults aged 18–29 years, prevalence appears to have plateaued: it was 24.9% in 2020, appeared to peak at 30.2% in 2022, and is now 21.8%. Similar trends have been seen with impaired glucose tolerance, the precursor to diabetes. This may indicate that early use of CFTR modulators in children is helping to protect the pancreas from further damage; if so, we should see this trend continue.

Once again, I hope you find the report as valuable as I have. I extend my sincere thanks to the clinical teams and to people with CF for taking the time to enter their data and make this report possible.

**Professor Peter Wark**
Director of Cystic Fibrosis Service, Respiratory and Sleep Physician, Alfred Health, Victoria,
Conjoint Professor of Medicine, Monash University, School of Translational Medicine,
Adjunct Professor, University of Newcastle,
Honorary Senior Staff Specialist, Respiratory and Sleep Medicine John Hunter Hospital, New South Wales

## FROM THE REGISTRY DEPUTY CLINICAL LEAD

It is an honour to be included as Deputy Clinical Lead of the Australian Cystic Fibrosis Data Registry. As a paediatric respiratory physician, the transformation in cystic fibrosis over the course of my career has been tremendous, and progress is further described in the 2025 Annual Report.

Our national registry, and the data within it, represents an incredible opportunity to develop future-focused CF care. In this 2025 Annual Report, the role of the registry in promoting quality care nationally through benchmarking and registry-led initiatives is evident in the description of clinical variation amongst CF centres (section 4) and the collection of patient-reported outcome measures (section 5), and I am looking forward to future developments in this space.

The registry data has, and will continue to, enable the targeting of CF treatment and interventions to maximise clinical outcomes while minimising burden of care where possible, and it is exciting to consider the research occurring in this space.

It is a privilege to be a part of this team, and I am grateful to all those who contribute their data, and their time and effort to maintain and develop the registry.

**Dr Katherine Frayman**
Respiratory Paediatrician, Department of Respiratory and Sleep Medicine, Royal Children's Hospital
Honorary Senior Fellow, Department of Paediatrics, University of Melbourne
Honorary Fellow Manager, Respiratory Diseases Group, Murdoch Children's Research Institute

# SUMMARY OF REGISTRY DATA

| | 2020 | 2021 | 2022 | 2023 | 2024 | 2025 |
|---|---|---|---|---|---|---|
| **PEOPLE WITH CYSTIC FIBROSIS (PWCF)** | | | | | | |
| Active population in the ACFDR* | 3,538 | 3,616 | 3,738 | 3,798 | 3,788 | 3,916 |
| Age (median) | 20.2 years | 20.6 years | 21.1 years | 21.7 years | 22.4 years | 23.0 years |
| Age (mean) | 22.6 years | 23.0 years | 23.4 years | 24.0 years | 24.6 years | 25.3 years |
| Adults (≥18 years) number (%); | 1,965 (55.5%) | 2,019 (55.8%) | 2,124 (56.8%) | 2,212 (58.2%) | 2,274 (60.0%) | 2,437 (62.2%) |
| Adults: Males % | 52.8% | 52.8% | 53.5% | 52.5% | 52.3% | 52.8% |
| Aboriginal and/or Torres Strait Islander (n) | 135 | 141 | 150 | 151 | 148 | 150 |
| **CF DIAGNOSIS & GENOTYPING** | | | | | | |
| Newly diagnosed pwCF | 74 | 92 | 90 | 68 | 48 | 65 |
| % New diagnoses <1 year | 82.4% | 82.6% | 78.9% | 77.9% | 83.3% | 76.9% |
| % New diagnoses ≥18 years | 10.8% | 12.0% | 11.1% | 13.2% | 16.7% | 21.5% |
| Genotyped: One or more known allele | 98.4% | 98.4% | 99.1% | 99.2% | 99.8% | 99.5% |
| Genotyped: Two known alleles | 92.2% | 94.8% | 95.9% | 96.3% | 99.7% | 99.4% |
| % F508del Homozygous | 47% | 47% | 46% | 46% | 46% | 46% |
| % F508del Heterozygous | 43% | 43% | 44% | 44% | 44% | 44% |
| **CLINICAL MEASURES (LUNG FUNCTION & NUTRITION)** | | | | | | |
| Median FEV1 pp 6-17 years | 91.0 | 93.0 | 93.2 | 96.6 | 97.7 | 98.0 |
| Median FEV1 pp 18 years and older | 70.0 | 73.0 | 75.6 | 78.6 | 79.0 | 80.6 |
| Median weight for length percentile <2 years | 51st | 51st | 50th | 54th | 51st | 48th |
| Median BMI percentile 2-17 years | 57th | 58th | 57th | 60th | 61st | 61st |
| Median BMI 18 years and older | 22.9 | 23.0 | 23.4 | 23.9 | 23.8 | 23.9 |
| Number of lung function tests (mean) | | | | | | |
| Paediatric | 3.4 | 3.8 | 3.1 | 2.9 | 2.7 | 2.7 |
| Adult | 3.6 | 3.6 | 2.9 | 2.8 | 2.7 | 2.4 |
| **RESPIRATORY MICROBIOLOGY** | | | | | | |
| *P. aeruginosa* (%) | 34.1% | 32.8% | 28.4% | 19.1% | 17.9% | 15.3% |
| *S. aureus* (%) | 39.4% | 40.7% | 38.4% | 32.5% | 32.5% | 29.7% |
| *Aspergillus* spp (%) | 15.9% | 15.1% | 11.6% | 7.1% | 6.6% | 5.9% |
| Non-tuberculous mycobacterium (%) | 5.2% | 6.9% | 3.8% | 2.6% | 2.6% | 3.1% |
| Number of Microbiology samples (mean) | | | | | | |
| Paediatric | 3.5 | 3.6 | 3.6 | 3.5 | 3.5 | 3.4 |
| Adult | 2.2 | 2.5 | 2.3 | 1.5 | 1.6 | 1.4 |
| **COMPLICATIONS** | | | | | | |
| % with CF related Diabetes <12 years | 3.2% | 3.0% | 3.8% | 2.2% | 2.0% | 1.0% |
| % with CF related Diabetes 12-17 years | 21.8% | 16.1% | 17.9% | 12.8% | 11.7% | 10.7% |
| % with CF related Diabetes 18-29 years | 24.9% | 26.0% | 30.2% | 26.6% | 24.7% | 21.8% |
| % with CF related Diabetes 30+ years | 28.6% | 27.9% | 30.8% | 28.9% | 30.0% | 29.5% |

| | 2020 | 2021 | 2022 | 2023 | 2024 | 2025 |
|---|---|---|---|---|---|---|
| **MULTIDISCIPLINARY CARE** | | | | | | |
| % with Physiotherapy annual review | N/A | 85.7% | 83.9% | 85.6% | 88.0% | 89.3% |
| % with Dietitian annual review | N/A | 74.7% | 76.6% | 77.5% | 76.8% | 78.7% |
| % with Mental Health annual review ≥12 years | N/A | 25.3% | 26.8% | 23.6% | 24.9% | 27.7% |
| % with Social work review | N/A | N/A | 43.2% | 46.2% | 48.6% | 46.1% |
| % with Gastroenterologist annual review | N/A | N/A | 24.0% | 23.8% | 21.4% | 21.2% |
| % with Endocrinologist annual review | N/A | N/A | 19.5% | 19.6% | 22.0% | 20.7% |
| **CFTR MODULATORS** | | | | | | |
| % Eligible and prescribed a CFTR modulator | | | | | | |
| Total cohort | N/A | N/A | 68.6% | 77.8% | 80.6% | 81.5% |
| Paediatric | N/A | N/A | 51.9% | 65.6% | 74.0% | 75.0% |
| Adult | N/A | N/A | 81.8% | 87.3% | 85.2% | 85.6% |
| **CF MANAGEMENT AND HEALTHCARE UTILISATION** | | | | | | |
| Clinical visits per person (mean) | | | | | | |
| Paediatric | 4.7 | 4.9 | 4.4 | 4.3 | 3.9 | 3.9 |
| Adult | 5.1 | 5.0 | 4.9 | 4.3 | 4.0 | 3.8 |
| Hospitalisations (mean) | | | | | | |
| Paediatric | 0.7 | 0.7 | 0.7 | 0.6 | 0.5 | 0.4 |
| Adult | 0.7 | 0.7 | 0.6 | 0.4 | 0.5 | 0.5 |
| % with zero hospitalisations | 62.5% | 61.2% | 64.2% | 70.4% | 71.9% | 74.6% |
| **PREGNANCY** | | | | | | |
| Total pregnancies | 42 | 48 | 69 | 85 | 86 | 81 |
| **LUNG TRANSPLANTS AND SURVIVAL** | | | | | | |
| Total pwCF living with a lung transplant | 129 | 123 | 117 | 117 | 107 | 98 |
| Bilateral lung transplants performed | 15 | 9 | 6 | 9 | 3 | 4 |
| Deaths (total CF deaths) | 18 | 19 | 10 | 18 | 11 | 10 |
| Deaths (lung transplant recipients only) | 6 | 11 | 5 | 8 | 8 | 4 |
| Median age of death (lung transplant recipients included) | 30.7 years | 36.8 years | 44.2 years | 40.6 years | 36.6 years | 40.1 years |
| Median age of death (lung transplant recipients excluded) | 21.1 years | 29.9 years | 34.1 years | 48.3 years | 25.5 years | 55.3 years |
| Survival median (cohort, birth years) | 51.2 years (2015-2019) | 54.7 years (2016-2020) | 56.9 years (2017-2021) | 60.6 years (2018-2022) | 64.0 years (2019-2023) | 70.2 years (2020-2024) |

* Defined as number of pwCF who met the diagnostic inclusion criteria and had acceptable data quality
**Note:** pwCF with lung transplants may not be captured in the ACFDR post-transplant.

# 1.

# AUSTRALIANS WITH CYSTIC FIBROSIS

# 1. AUSTRALIANS WITH CYSTIC FIBROSIS

## 1.1 OVERVIEW

Cystic fibrosis (CF) is a recessively inherited genetic condition, a multisystem disorder associated with reduced life expectancy, mostly due to respiratory failure. This report highlights the epidemiological and clinical characteristics of children and adults with CF that are captured in the Australian Cystic Fibrosis Data Registry (ACFDR), as of December 31st 2025.

The ACFDR collects data from people with CF (pwCF) from the time of their diagnosis and throughout their life, or until they have undergone a lung transplant, when many pwCF are discharged from CF-centre care. Most pwCF are cared for by specialist clinicians and teams in public hospital CF Centres. These centres may be associated with paediatric health services, adult health services or both. The ACFDR 2025 Report is structured in four main sections in recognition that the experience of living with CF changes over time as do the treatments and outcomes. The sections relate to:

1. High level overview of aggregate data for pwCF, including survival outcomes
2. Diagnosis and management of children and adolescents with CF
3. Diagnosis and management of adults with CF
4. Benchmarking of site-level data

For the first time this report includes early Patient Reported Outcomes data for pwCF, available in chapter 5.

This 2025 report includes data from 3,916 pwCF across Australia. The 2025 cohort, also described as the 'active population' encompasses pwCF who accessed specialist CF care in Australia during 2025. This cohort comprises returning patients continuing their CF care, newly diagnosed pwCF, and those diagnosed in previous years who were new to the ACFDR during the reporting period. Individuals with CF may be considered 'inactive' if no specialist CF care record was submitted to the ACFDR during the reporting period. This does not necessarily indicate that no care was received. Furthermore, individuals may return to the active population in future reporting years. As such, minor fluctuations in the total number of pwCF can be expected year-to-year and are reflected in the sample sizes and analyses presented within this report.

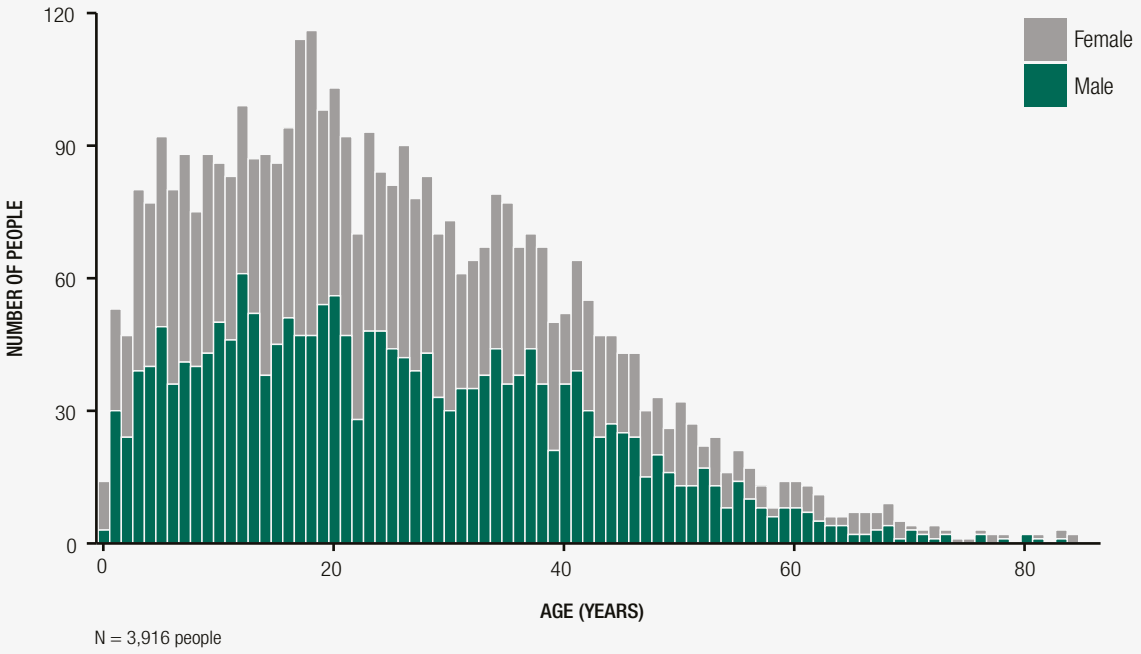
As per the 2023 and 2024 reports, this report follows international registry reporting practice to **only include data from lung transplant recipients in the following sections - Demographics, Diagnostic and Genotype Information**. This is so that information regarding clinical care and outcomes reflects the majority of pwCF who have not had a lung transplant. **Survival analyses will be presented in relation to pwCF with and without transplants separately**. As of December 31st 2025, the ACFDR contains data from **98 pwCF** who have undergone a lung transplant.

## 1.2 DEMOGRAPHICS

### Age and Sex of People with CF in the Registry

As of December 31st 2025, the ACFDR held records of **3,916 active pwCF**, collected from 23 CF centres in Australia. Of the 3,916 pwCF, **2,437 were adults** (18+ years) and **1,479 were children and adolescents** (0-17 years) (Figure 1.1). Throughout this report, age is calculated based on age at December 31st of the reporting year.

**FIGURE 1.1:** ACFDR 2025: PEOPLE WITH CF IN AUSTRALIA BY AGE AND SEX




As of December 31st 2025, the proportion of males in the ACFDR was 52.1% and females were 47.9%. The largest age grouping for pwCF was those aged 18-29 years which comprises 26.7% of pwCF, followed by pwCF aged ≥40 years (18.6%) and pwCF aged 30-39 (16.9%) (Figure 1.1 and Table 1.1).

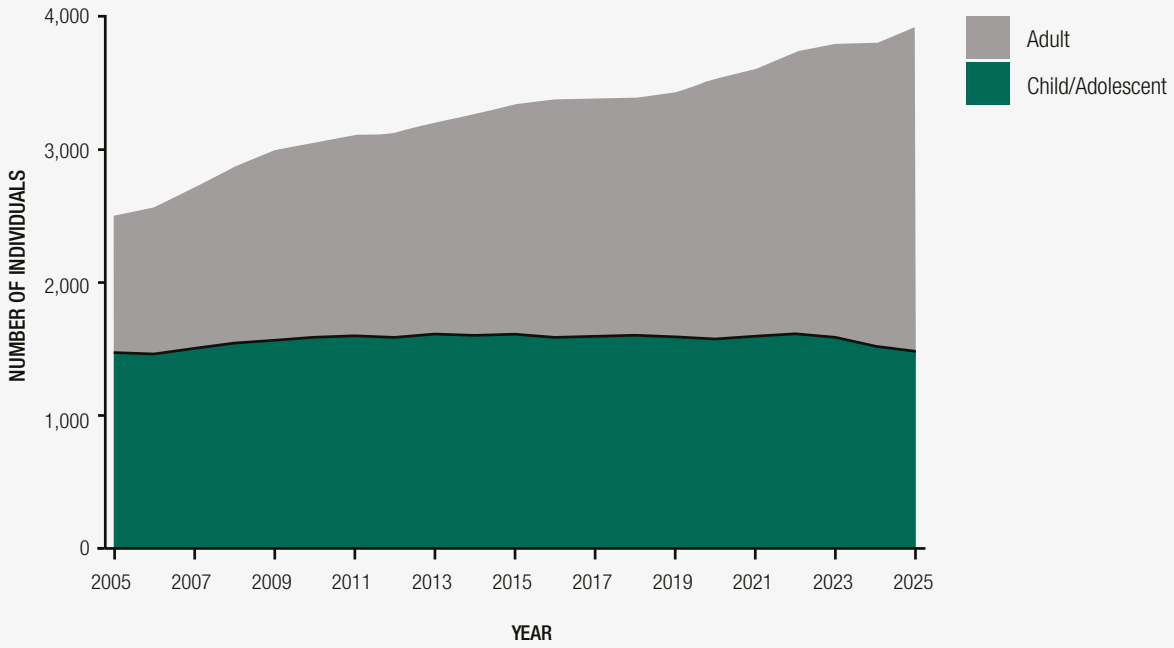
**TABLE 1.1:** ACFDR 2025: PEOPLE WITH CF BY AGE AND SEX

| Age | Female | Male | Total |
|---|---|---|---|
| <2 | 48.9% (45) | 51.1% (47) | 92 |
| 2-5 | 49.0% (154) | 51.0% (160) | 314 |
| 6-11 | 48.4% (242) | 51.6% (258) | 500 |
| 12-17 | 49.2% (282) | 50.8% (291) | 573 |
| 18-29 | 49.8% (521) | 50.2% (525) | 1,046 |
| 30-39 | 46.0% (305) | 54.0% (358) | 663 |
| ≥40 | 44.6% (325) | 55.4% (403) | 728 |
| **Total** | 47.9% (1,874) | 52.1% (2,042) | 3,916 |

The proportion of the registry population who were adults in 2025 was 62.2%, compared with 60.0% in 2024 (Figure 1.2). In 2025, the median age of the CF population was 23.0 years, with a mean age of 25.3 years. The median age for males at 23.6 years (22.9 years in 2024) remained higher than that for females at 22.3 years in 2025 (21.8 years in 2024).

In 2025, the number of Indigenous Australians with CF was 150.

**FIGURE 1.2:** ACFDR 2005-2025: PAEDIATRIC VS ADULTS PROPORTION

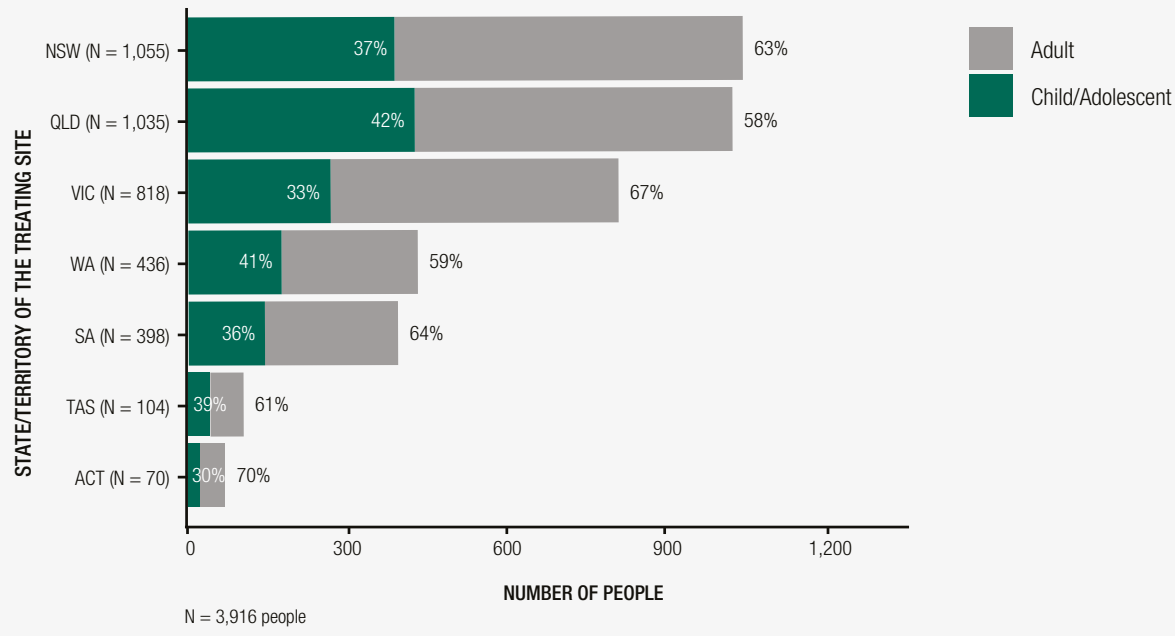



**Note:** Population size in 2017 was estimated based on the populations in years 2016 and 2018

## Geographical Distribution of People with CF in Australia

Figure 1.3 shows the number of pwCF and the jurisdiction in which they received their care in 2025, as well as the proportion of these who were children/adolescents and adults. CF centres in New South Wales provided care to the most pwCF, followed by those in Queensland, Victoria, Western Australia, South Australia, Tasmania and the Australian Capital Territory.

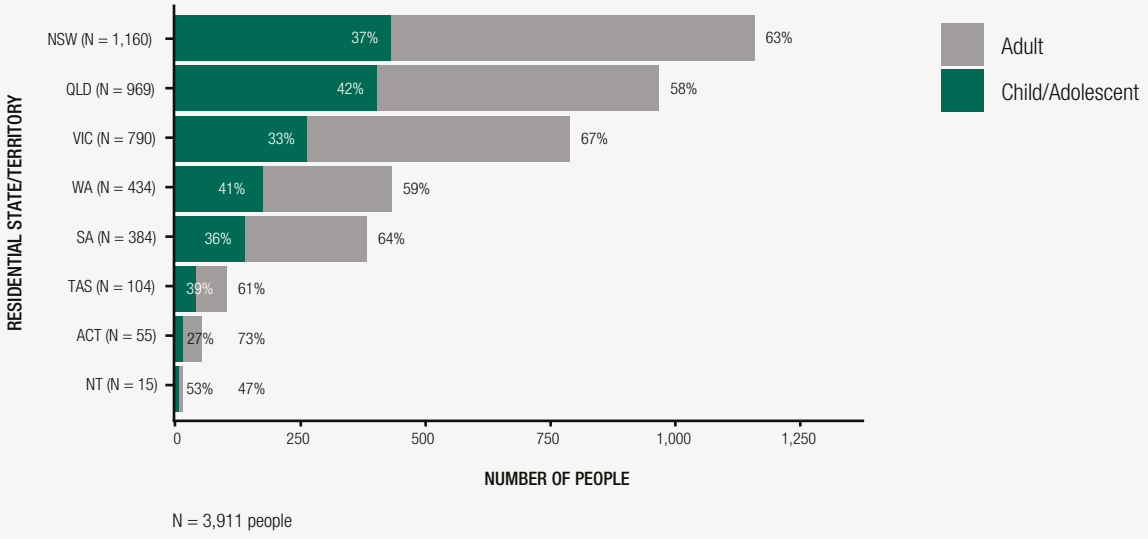
**FIGURE 1.3:** ACFDR 2025: DISTRIBUTION BY STATE AND TERRITORY WHERE CARE WAS PROVIDED




**Note:** PwCF who were shared or transferred between CF Centres in 2025 may be represented more than once

Figure 1.4 shows the jurisdictions where pwCF lived in 2025 (or the most recent prior year), derived from postcode information collected by the registry. New South Wales had the highest number and proportion (1,160/30% of residing pwCF), followed by Queensland (969/25%), Victoria (790/20%), Western Australia (434/11%), South Australia (384/10%), Tasmania (104/3%), the Australian Capital Territory (55/1%) and the Northern Territory (15/<1%). Five pwCF did not have their postcode recorded. In 2025, there were minimal differences between jurisdiction of residence and jurisdiction of care with the exception of the Northern Territory and the Australian Capital Territory. This suggests the vast majority of pwCF are accessing care within their state or territory of residence.

**FIGURE 1.4:** ACFDR 2025: DISTRIBUTION BY PATIENT RESIDENCE




N = 3,911 people

## 1.3 DIAGNOSTIC AND GENOTYPE INFORMATION

### Diagnostic Information

#### New Diagnoses

The number of new CF diagnoses notified to the registry in 2025 was 65, (compared to 48 in 2024) including 50 people (compared to 40 in 2024) diagnosed at less than one year of age and 14 people diagnosed over 18 years. New diagnoses refer to individuals diagnosed with CF between July 1st 2024 and December 31st 2025 who were not represented in previous ACFDR Annual Reports. In addition to those newly diagnosed, this report includes pwCF diagnosed prior to July 2024 who contributed data to the ACFDR for the first time in 2025. This brings the total 2025 cohort to 3,916.

Over half of all new diagnoses, and approximately three-quarters of diagnoses within 8 weeks, were made by newborn screening (Table 1.2). Compared to those in 2024, a lower proportion of 2025 new diagnoses occurring within 8 weeks were made by newborn screening (89.2% in 2024 vs 72.7% in 2025). This suggests that some paediatric diagnoses are taking longer to confirm. Compared to the eight adults diagnosed with CF in 2024, fourteen adults were diagnosed in 2025 (21.5% of new diagnoses). This is consistent with an increasing trend in the proportion of adult CF diagnoses (Table 1.3).

**TABLE 1.2:** ACFDR 2025: AGE OF NEW DIAGNOSES

| Age | Total (%) | Diagnosis via newborn screening test | Diagnosis by non-newborn screening test indication |
|---|---|---|---|
| Birth-8 weeks | 44 (67.7%) | 32 (72.7%) | 12 (27.3%) |
| 8 weeks-12 months | 6 (9.2%) | 5 (83.3%) | <5 |
| 1-17 years | <5 | 0 (0.0%) | <5 |
| 18+ years | 14 (21.5%) | 0 (0.0%) | 14 (100.0%) |
| **Total** | 65 (100.0%) | 37 (56.9%) | 28 (43.1%) |

**TABLE 1.3:** ACFDR 2020-2025: NEW DIAGNOSES BY AGE

| | 2020 | 2021 | 2022 | 2023 | 2024 | 2025 |
|---|---|---|---|---|---|---|
| Total (N) | 74 | 92 | 90 | 68 | 48 | 65 |
| <1 year (%) | 82.4% | 82.6% | 78.9% | 77.9% | 83.3% | 76.9% |
| 1-17 years (%) | 6.8% | 5.4% | 10.0% | 8.8% | 0.0% | 1.6% |
| 18+ years (%) | 10.8% | 12.0% | 11.1% | 13.2% | 16.7% | 21.5% |

## Factors Associated with the Diagnosis of CF

For the total ACFDR cohort (3916 pwCF), a diagnosis of CF was confirmed or suggested by newborn screening (58.5%), clinical signs/symptoms (35.3%), family history (8.8%), and prenatal screening (2.9%) (Figure 1.5 and Table 1.4). Of those with clinical symptoms, the most common presentations were meconium ileus/intestinal obstruction (33.1%), respiratory signs and symptoms (27.0%), and gastrointestinal symptoms (14.3%). With multiple responses available, often there is an overlap in clinical symptoms and newborn screening or family history in the registry.

**FIGURE 1.5:** ACFDR 2025: FACTORS ASSOCIATED WITH DIAGNOSIS OF CF (WHOLE COHORT)

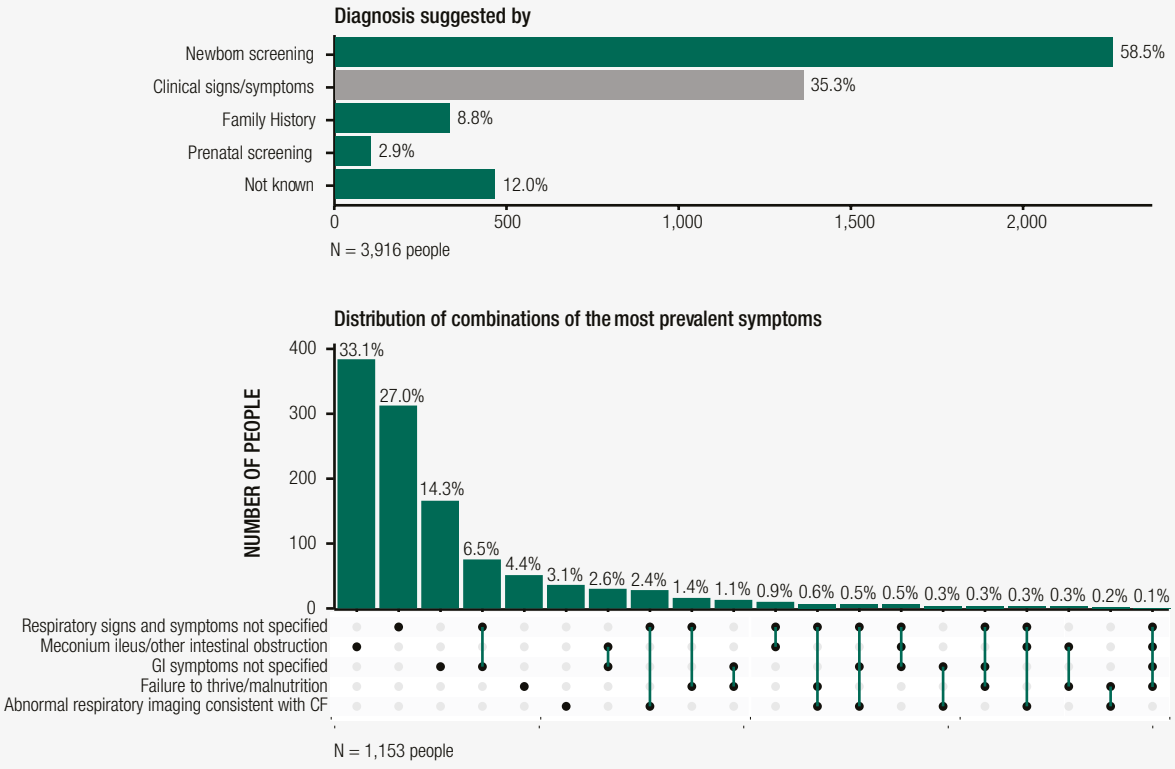


Table 1.4 shows the factors associated with CF diagnoses for the total CF cohort for 2025 compared with the new diagnoses for 2025. The proportion of diagnoses indicated by newborn screening or clinical signs/symptoms were similar for both newly diagnosed individuals and the whole cohort. New diagnoses were more likely to have a sweat chloride value recorded than the broader CF population (84.6% vs 63.9%). Compared to the total cohort, new diagnoses in 2025 were less likely to arise from clinical symptoms or signs (35.3% vs 32.3%) and more likely to arise from family history (8.8% vs 13.8%). Notably, the proportion of diagnoses associated with prenatal screening has increased from 2.9% (cohort average) to 10.8% in 2025. This may reflect changes to subsidisation of genetic testing in November 2023. Additionally, an increase in genetic testing may also lead to a reduction in continuing pregnancies that test positive for CF and a reduction in diagnoses attributed to NBS.

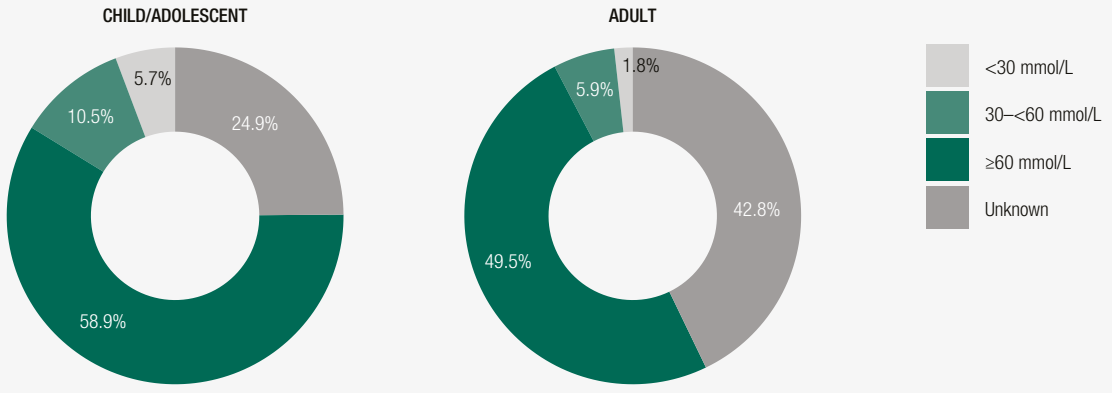
**TABLE 1.4:** ACFDR 1998-2025: FACTORS ASSOCIATED WITH DIAGNOSIS OF CF (2025 VS WHOLE COHORT)

| Diagnosis by | Total cohort N = 3,916 (%) | 2025 New diagnosis N = 65 (%) |
|---|---|---|
| Newborn screening | 2,291 (58.5%) | 37 (56.9%) |
| Clinical signs/symptoms | 1,382 (35.3%) | 21 (32.3%) |
| Family history | 345 (8.8%) | 9 (13.8%) |
| Prenatal screening | 114 (2.9%) | 7 (10.8%) |
| Not known | 468 (12.0%) | 0 (0.0%) |
| Sweat chloride value available | 2,504 (63.9%) | 55 (84.6%) |

Alongside indications for diagnosis, Table 1.4 shows the proportion of pwCF with a sweat chloride value at diagnosis. For pwCF diagnosed in 2025, the documentation of sweat chloride in the registry was 84.6% compared to 63.9% for all pwCF. Sweat chloride testing is important in the diagnosis of CF. In addition to assisting the diagnosis of CF, repeat sweat chloride measures can indicate disease progression/adherence to treatments.

Figure 1.6 shows the proportion of child/adolescent and adult pwCF with sweat chloride values (pre-CFTR modulator use) of <30 mmol/L, 30-<60 mmol/L, ≥60 mmol/L, and unknown. Fewer children and adolescents had unknown sweat chloride than adults (24.9% compared to 42.8%).

**FIGURE 1.6:** ACFDR 2025: SWEAT CHLORIDE VALUES AT DIAGNOSIS BY AGE

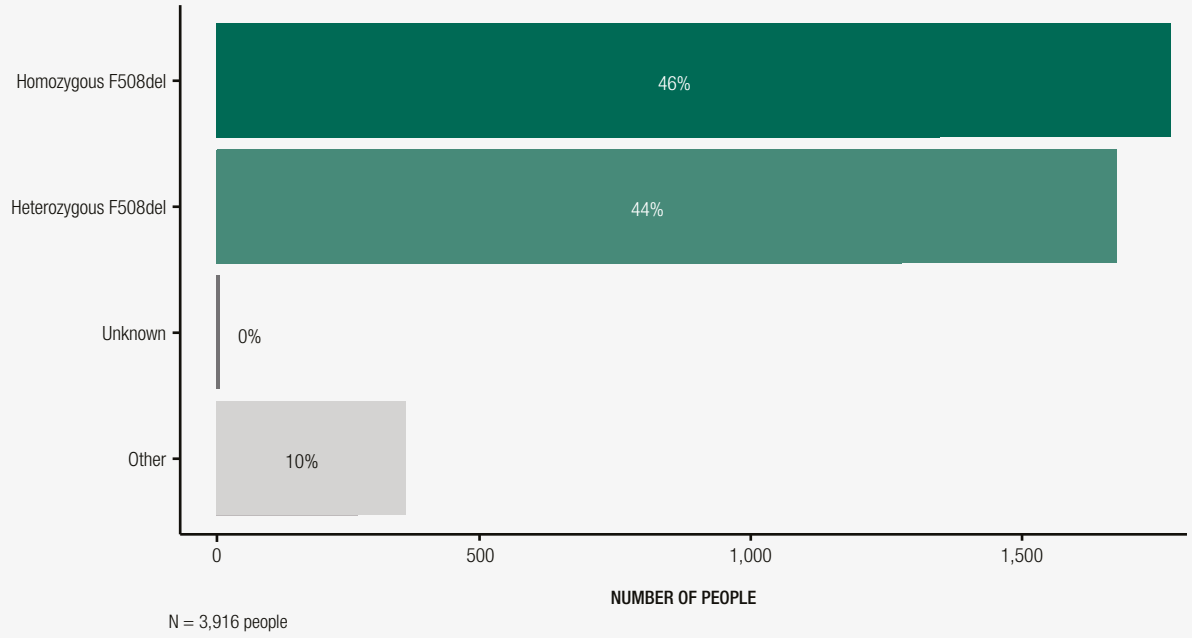


## Genotype Information

Information regarding the specific genotype of individuals diagnosed with CF is critical, due to current treatments that are available for specific alleles or allele combinations. The CFTR gene has two alleles, and variations may exist in both alleles that are related to the development of CF symptoms (phenotype). Of the 3,916 people in the registry in 2025, 99.5% had at least one allele known and 99.4% had both alleles known. Only 18 (0.5%) pwCF had two unknown alleles, with 13 of these being adults.

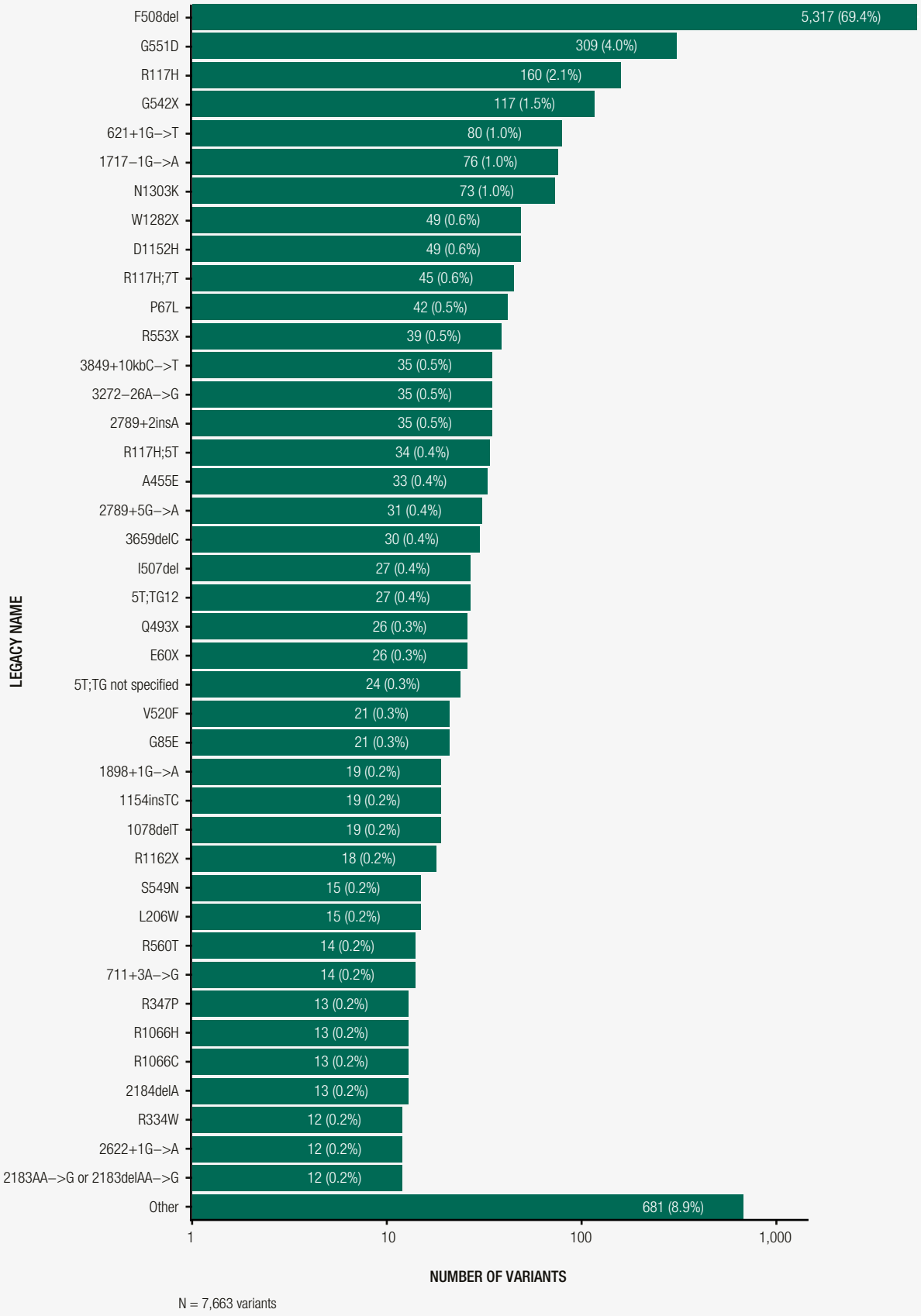
The most common CF-causing variant is F508del. PwCF can either be homozygous to F508del (have two F508del alleles) or heterozygous (have one F508del allele). In 2025, the proportion of Australian pwCF who were homozygous for the F508del variant was 46%, and the proportion who are heterozygous was 44%, thus 90% of pwCF in Australia have F508del variants. In 2025, one in ten pwCF do not have a F508del allele (Figure 1.7).

**FIGURE 1.7:** ACFDR 2025: MOST COMMON CFTR VARIANT COMBINATIONS



The most common variant alleles other than F508del in 2025 were G551D (4.0%), R117H (2.1%), and G542X (1.5%) (Figure 1.8).

**FIGURE 1.8:** ACFDR 2025: MOST COMMON INDIVIDUAL ALLELE CFTR VARIANTS IN THE ACFDR




N = 7,663 variants

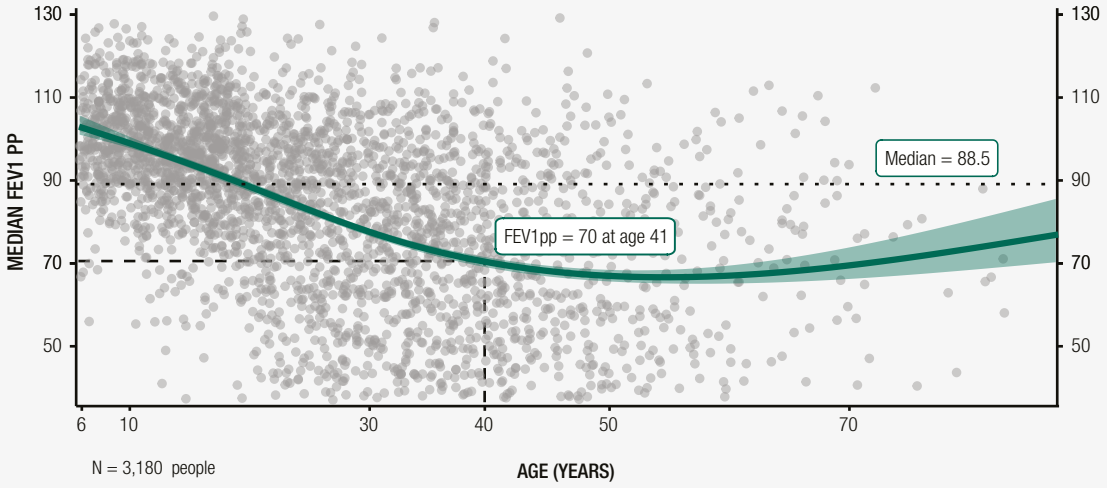
## 1.4 LUNG FUNCTION

For monitoring lung function in pwCF, the average of the highest Forced Expiratory Volume (litres) in 1 second percent predicted (FEV1 pp) is calculated in each quarter of the year. Predicted values are based on the Global Lung Initiative (GLI) formulae. Lung function measures are aligned with methods used in the United States Cystic Fibrosis Foundation Patient Registry.

Over 80% of active pwCF in the ACFDR have lung function information for 2025 (3,180 people). Four hundred and six (10.6%) pwCF in the registry are children younger than 6 years of age who do not routinely have lung function information recorded, and a further 7.5% of registry participants did not have lung function information recorded in 2025.

For 2025, the median lung function for pwCF, measured as FEV1 pp, is within the normal range for young children and adolescents (0-17) (Figure 1.9). At 41 years of age, the median FEV1 pp is 70.0.

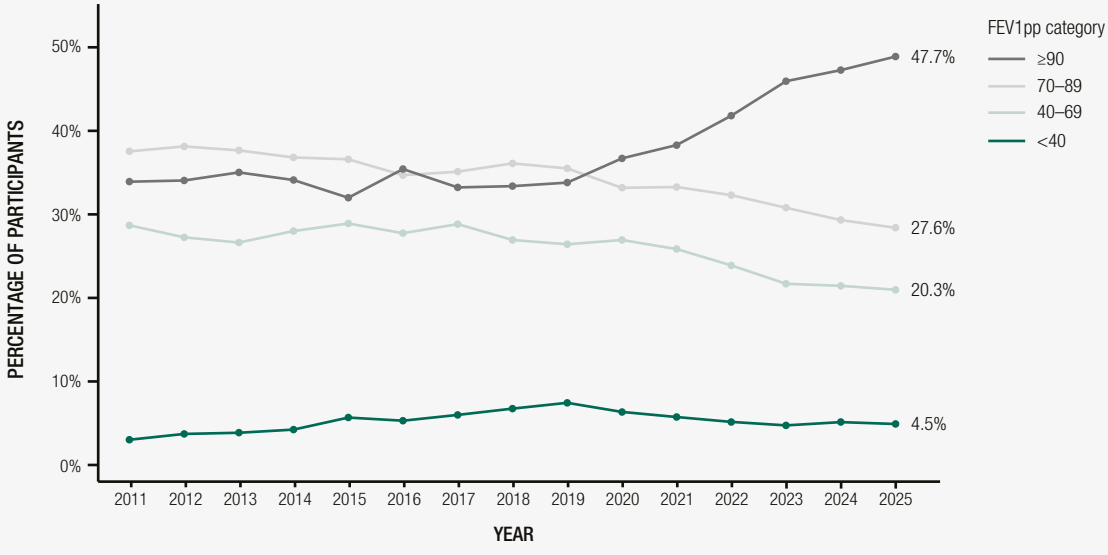
**FIGURE 1.9:** ACFDR 2025: MEDIAN FEV1 PP BY AGE




The solid trend line was estimated using a natural cubic spline with 3 degrees of freedom.
The dotted horizontal line shows the overall median FEV1pp.
Shaded area represents the 95% confidence interval.

Figure 1.10 depicts the trends in annual FEV1 pp for pwCF in four categories (FEV1 pp of ≥90; 70-89; 40-69; <40). The figure shows a significant increase since 2019 (from 32.8 to 47.7) in the proportion of pwCF who have an FEV1 pp of ≥90; and a concomitant decrease in the proportion of pwCF with FEV1 pp in the range of 40-90. The proportion of pwCF who have median FEV1 pp of <40 remains relatively stable, affecting 4.5% of pwCF in 2025.

**FIGURE 1.10:** ACFDR 2011-2025: FEV1 PP DISTRIBUTION OVER TIME

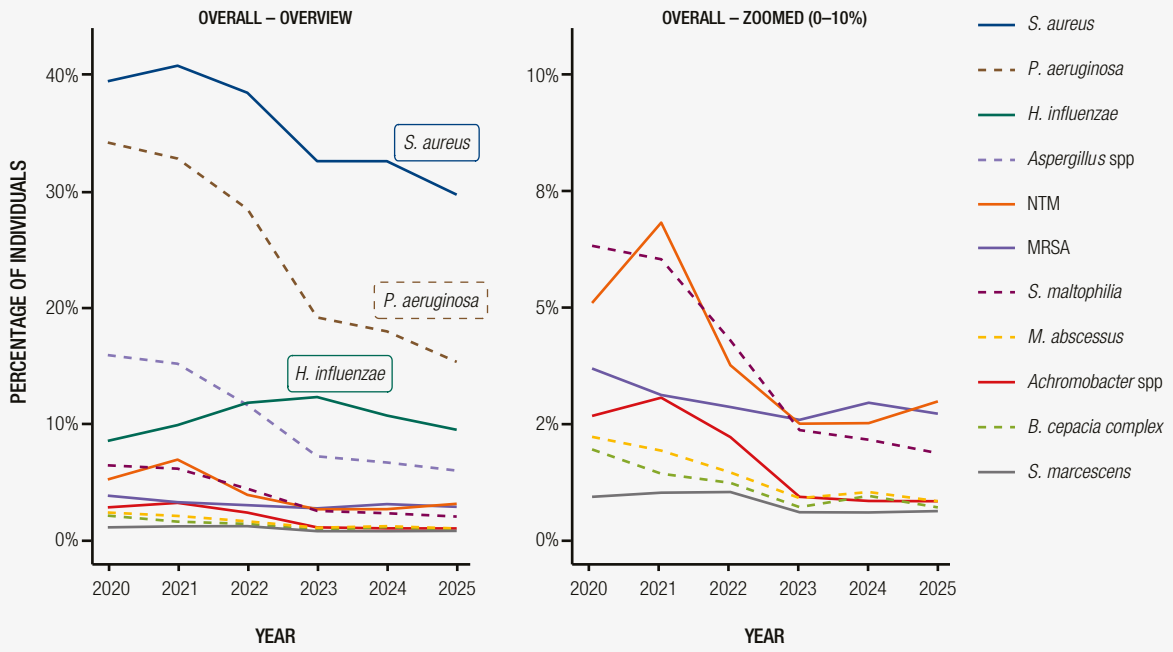


## 1.5 MICROBIOLOGY

Figure 1.11 shows the proportion of pwCF who tested positive for respiratory organisms commonly pathogenic to pwCF on at least one occasion. There has been a notable reduction over a number of years in many high prevalence organisms, including *S. aureus*, *P. aeruginosa* and *Aspergillus* spp.

As of December 2025, the overall prevalence recorded in the registry of *S. aureus* was 29.7%; *P. aeruginosa* was 15.3%; *H. influenzae* was 9.4%; *Aspergillus* spp was 5.9%; Non-tuberculous Mycobacteria (NTM) was 3.1%; *Methicillin-resistant Staphylococcus aureus* (MRSA) was 2.8%; *S. maltophilia* was 2.0%; *Achromobacter* spp was 0.9%; *M. abscessus* was 0.9%; *B. cepacia complex* was 0.8%; and *S. marcescens* had a prevalence of 0.7%.

**FIGURE 1.11:** ACFDR 2020-2025: PREVALENCE OF RESPIRATORY MICROORGANISMS IN SPUTUM, BAL BRONCHOSCOPY AND UPPER AIRWAY SAMPLES




## 1.6 CFTR MODULATORS

Disease-modifying therapies have the potential to dramatically reduce symptoms and increase survival for an increasing number of pwCF. Different therapies target different genetic variants, and not all pwCF may be eligible to receive Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) modulators. Additionally, CFTR modulators are high-cost medicines and are generally available initially in Australia via special access schemes before being approved for listing on the Pharmaceutical Benefits Scheme (PBS). In 2025, the following CFTR modulators were available to pwCF meeting the defined eligibility criteria. Further information on modulator eligibility can be found via the relevant Therapeutic Goods Administration Product Information sheets.

### Ivacaftor (KALYDECO®)

Ivacaftor is available on the PBS for pwCF aged 1 month and older (extended in September 2025 from the previous eligibility of 4 months and older) who have at least one mutation in the CFTR gene that is responsive to Ivacaftor potentiation based on clinical and/or in vitro data.

### Lumacaftor/Ivacaftor (ORKAMBI®)

Lumacaftor/ivacaftor is a combination therapy available on the PBS for pwCF aged one year and older who have two copies of the F508del gene change in the CFTR gene.

### Tezacaftor/Ivacaftor And Ivacaftor (SYMDEKO®)

Tezacaftor/ivacaftor is also a combination therapy available on the PBS for pwCF aged 12 years and older with either two copies of the F508del mutation or at least one copy of a responsive mutation in the CFTR gene based on in-vitro data.

## Elexacaftor/Tezacaftor/Ivacaftor (TRIKAFTA®)

Elexacaftor/tezacaftor/ivacaftor (ETI) is a triple combination therapy available on the PBS for pwCF aged 2 years and older, who have at least one mutation in the CFTR gene that is responsive based on clinical and/or in vitro data.

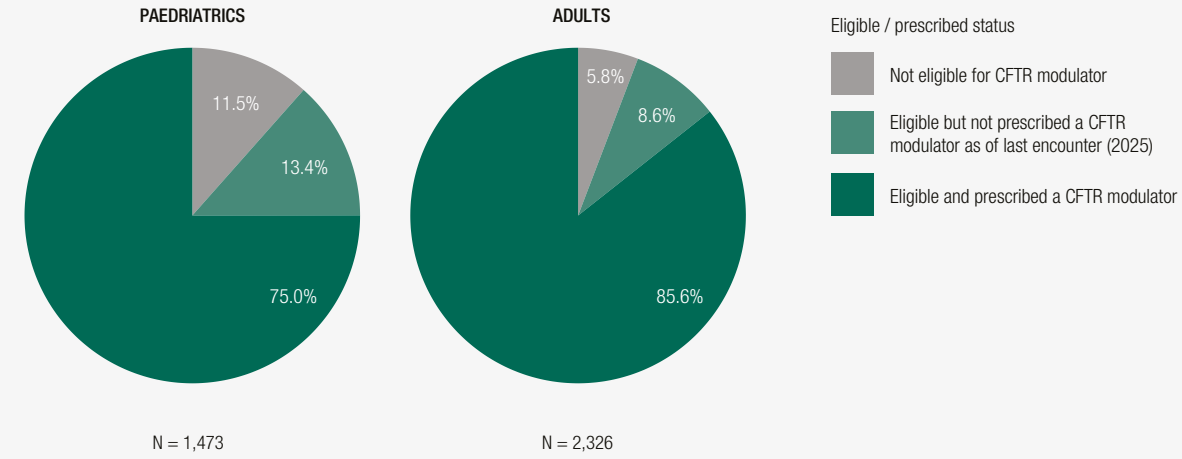
Trikafta was initially available on the PBS in April 2022 for pwCF aged 12 years and older with at least one copy of the F508del gene change in the CFTR gene. In May 2023 pwCF aged 6-11 years were eligible to receive Trikafta, in August 2024 pwCF aged 2-5 years were able to access the treatment, and in July 2025 the eligible CFTR gene mutations were expanded to include pwCF with at least one responsive mutation in the CFTR gene.

To account for delays between CFTR modulator eligibility criteria updates and medication reviews for pwCF, the CFTR modulator information presented within this report considers eligibility based on the criteria as of June 30th 2025.

A total of 3,799 pwCF (99.5%) in the registry had known eligibility status for CFTR modulators based on their genotype (Figure 1.12). Of these, 3,097 (81.5%) pwCF were prescribed at least one modulator during 2025.

In 2025, the number of pwCF receiving a modulator prescription increased by 70; up from 3,027 in 2024.

**FIGURE 1.12:** ACFDR 2025: ELIGIBILITY AND PRESCRIBING OF CFTR MODULATORS BY AGE




As of December 31st 2025

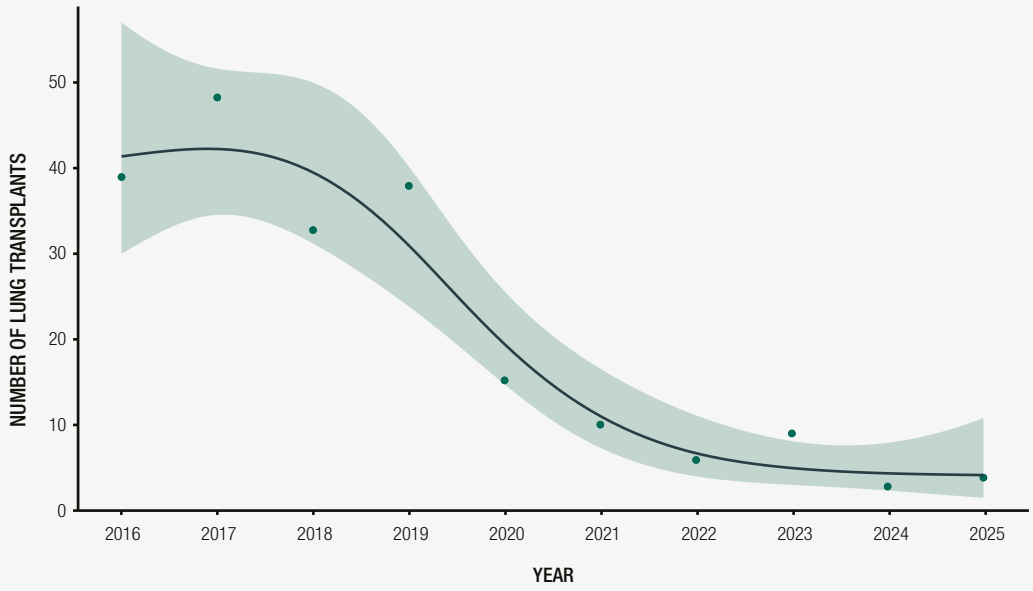
## 1.7 TRANSPLANTATION AND SURVIVAL

### Transplantation

The most common transplantation procedure for pwCF is a bilateral (double) lung transplant. As CF is a systemic disease, other organs may also be severely affected by either the underlying disease or its related complications and require transplantation, including the kidney, liver or pancreas. Occasionally multi-organ transplants are required.

In 2025, there were a total of 5 transplants for pwCF; 4 were bilateral lung transplants of which all were performed in adults. Thirty-seven people were evaluated for a transplant in 2025; 3 (8.1%) were waitlisted, and 4 (10.8%) were deferred from the waiting list. The number of annual bilateral lung transplants undertaken over the last decade is shown in Figure 1.13. There has been a substantial decline in bilateral lung transplants over the last few years among pwCF in Australia, consistent with international trends.

**FIGURE 1.13:** ACFDR 2016-2025: BILATERAL LUNG TRANSPLANTS




Solid trend line: Quasi-Poisson GLM with natural cubic spline (df = 3).
Shaded area: 95% CI on the mean count (log link, back-transformed).

### Status of People with CF in the ACFDR

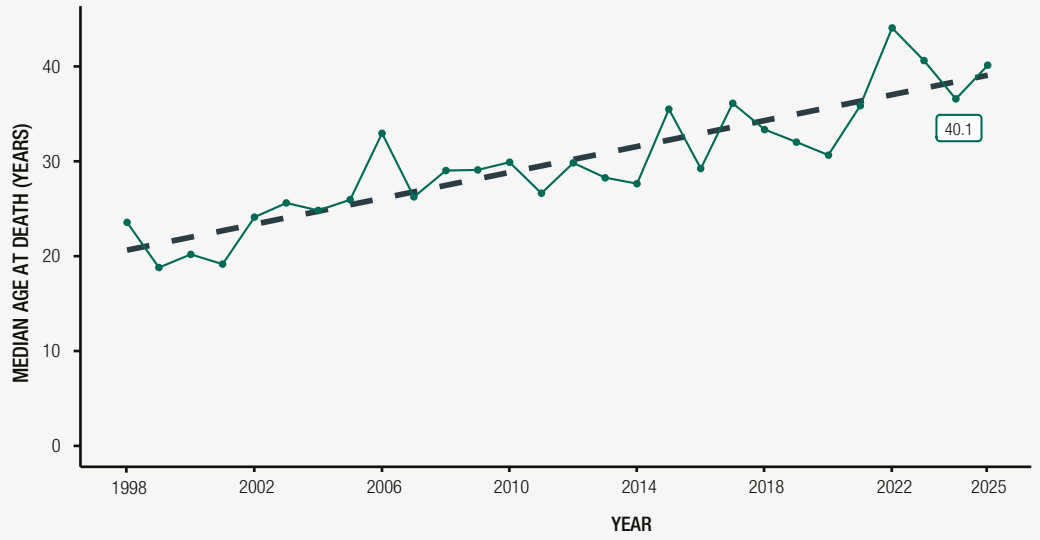
The (follow-up) status of people in the ACFDR is updated annually by CF centres. Many pwCF who have undergone organ transplantation may not have been followed up by the ACFDR, and their deaths may not be captured in the registry.

In 2025, the ACFDR recorded the deaths of 10 pwCF, of which four (40.0%) were pwCF who had received a lung transplant. Seventy percent of deaths occurred in pwCF aged 30 years or older. In 2025, the deaths among individuals with CF were caused by pulmonary manifestations (3), causes unrelated to CF (3), post-transplant complications (2), unspecified CF-related causes (1), and unknown cause of death (1).

#### Median Age of Death

The median age of death in 2025 was 40.1 years of age for pwCF (Figure 1.14). Median age may vary from year to year given the relatively small number of deaths per annum. The median age of death differs from estimated survival, which projects the lifespan of a person with CF born in a specific year.

**FIGURE 1.14:** ACFDR 1998-2025: MEDIAN AGE OF DEATH




Straight dashed line represents the overall trend estimated by a linear regression model

## Survival

The median estimated survival for pwCF is determined on the basis of the individuals who are alive in the ACFDR in a given year. Internationally, CF registries have documented steady increases in median survival over recent years, attributed to advancements in treatments. This positive trend is expected to persist, with further improvements anticipated as more individuals with cystic fibrosis are managed with CFTR modulators.

**Table 1.5 is inclusive of pwCF who have undergone a lung transplant, while Table 1.6 excludes pwCF who have undergone a lung transplant**. Table 1.5 shows that the estimated survival has increased from 47.4 years for pwCF born in 2011-15, to 70.2 years for pwCF born in 2020-24. For pwCF, excluding those with lung transplants, survival is even higher, with a median survival of 82.1 years for pwCF born in 2020-24. These estimates are based on currently observed mortality patterns, and as such, should be interpreted cautiously. The model does not fully account for future age-related comorbidities that many pwCF have not yet reached in large numbers. Future survival modelling may need to incorporate ageing-related disease burden as the CF population continues to age.

The ACFDR is reporting survival data one year in arrears to allow for late notification of recent deaths to be captured by the registry. The N/A value for the upper 95% confidence limit indicates that the upper bound of the confidence interval for the median survival time could not be estimated. This is likely due to a combination of a high number of censored observations (pwCF who were still alive at the end of the study period) and a limited number of events (deaths). Continued follow-up will help improve the accuracy of our estimates.

**TABLE 1.5:** ACFDR 2011-2024: MEDIAN SURVIVAL OF PEOPLE WITH CF IN AUSTRALIA (LUNG TRANSPLANT RECIPIENTS INCLUDED)

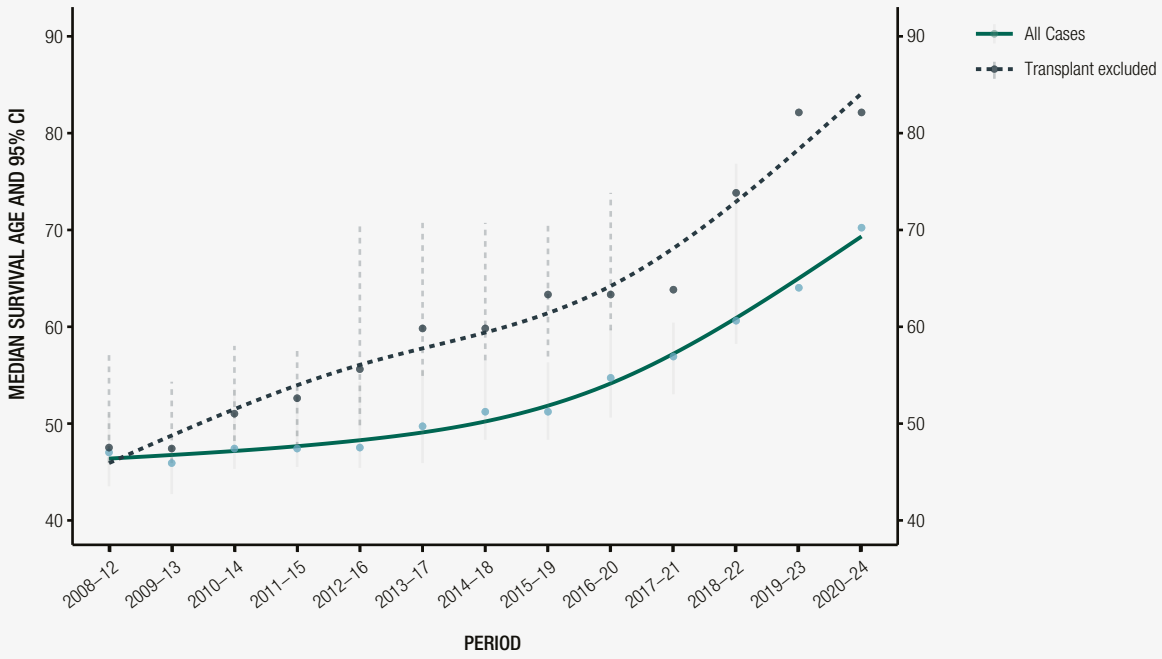
| Period | Median age and 95% CI (years) | Number of deaths/Number at risk |
|---|---|---|
| 2011-15 | 47.4 (45.5 - 53.6) | 174 / 3,495 |
| 2012-16 | 47.5 (45.4 - 54.3) | 181 / 3,556 |
| 2013-17 | 49.7 (45.9 - 55.6) | 179 / 3,587 |
| 2014-18 | 51.2 (48.3 - 56.9) | 175 / 3,720 |
| 2015-19 | 51.2 (48.0 - 56.3) | 180 / 3,789 |
| 2016-20 | 54.7 (50.6 - 59.8) | 171 / 3,821 |
| 2017-21 | 56.9 (53.0 - 60.4) | 154 / 3,861 |
| 2018-22 | 60.6 (58.2 - 76.8) | 120 / 3,898 |
| 2019-23 | 64.0 (60.4 - NA) | 97 / 3,938 |
| 2020-24 | 70.2 (61.1 - NA) | 80 / 3,972 |

**TABLE 1.6:** ACFDR 2011-2024: MEDIAN SURVIVAL OF PEOPLE WITH CF IN AUSTRALIA (LUNG TRANSPLANT RECIPIENTS EXCLUDED)

| Period | Median age and 95% CI (years) | Number of deaths/Number at risk |
|---|---|---|
| 2011-15 | 52.6 (47.5 - 58.0) | 99 / 3,071 |
| 2012-16 | 54.3 (48 - 70.7) | 106 / 3,166 |
| 2013-17 | 56.9 (54 - 70.7) | 104 / 3,216 |
| 2014-18 | 59.8 (56.3 - 70.7) | 99 / 3,359 |
| 2015-19 | 63.3 (56.9 - 70.7) | 97 / 3,447 |
| 2016-20 | 63.3 (59.2 - 73.8) | 95 / 3,529 |
| 2017-21 | 63.8 (59.8 - NA) | 84 / 3,601 |
| 2018-22 | 73.8 (63.8 - NA) | 62 / 3,672 |
| 2019-23 | 82.1 (70.2 - NA) | 51 / 3,747 |
| 2020-24 | 82.1 (70.2 - NA) | 41 / 3,809 |

Survival is also shown graphically below for all pwCF in the registry (Figure 1.15).

**FIGURE 1.15:** ACFDR 2008-2024: MEDIAN SURVIVAL OF PEOPLE WITH CF IN AUSTRALIA




Each point shows the estimated median survival age.
Vertical bars show the two-sided 95% CI where available.
Smoothed lines are natural cubic splines (df = 3).

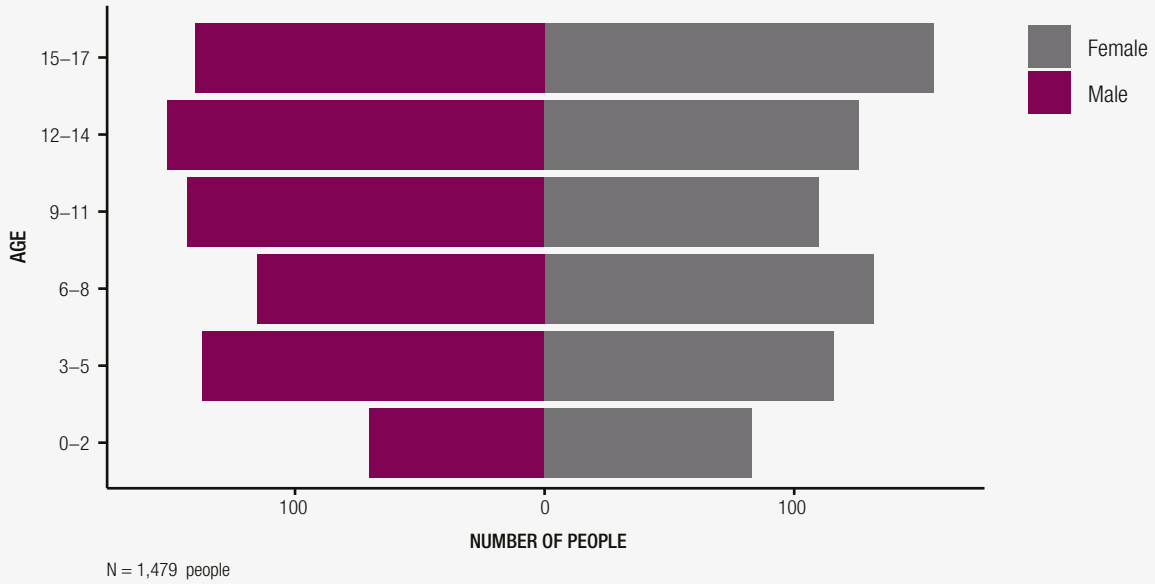
# 2. PAEDIATRIC DATA

# 2. PAEDIATRIC DATA

## 2.1 DEMOGRAPHIC INFORMATION

As of December 31st 2025, the ACFDR held data regarding 1,479 children and adolescents (0-17 years old) with CF, comprising 723 females and 756 males (Figure 2.1).

**FIGURE 2.1:** ACFDR PAEDIATRICS 2025: BY AGE AND SEX



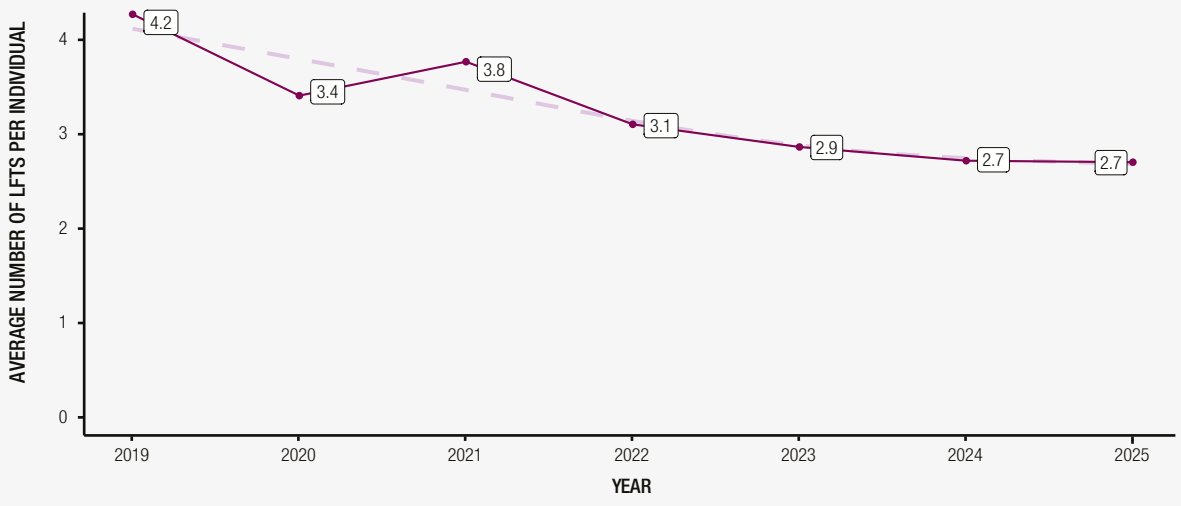

N = 1,479 people

## 2.2 CLINICAL MEASURES

### Lung Function

Figure 2.2 illustrates the average number of spirometry tests per child recorded annually in the registry from 2019 to 2025. In 2019, children and adolescents averaged 4.2 tests each, however this declined to 2.7 tests per child in 2024 and has since remained stable.

**FIGURE 2.2:** ACFDR PAEDIATRICS 2019-2025: AVERAGE NUMBER OF LUNG FUNCTION TESTS ENTERED PER INDIVIDUAL

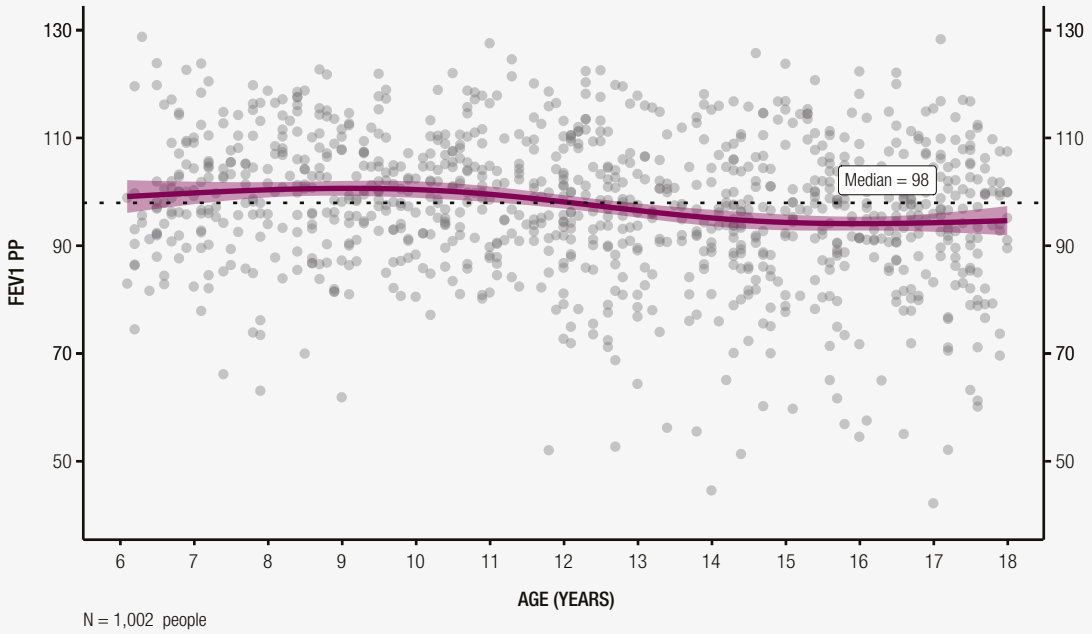



Dashed curve represents the smoothed trend (natural cubic spline, df = 3)

For the monitoring of lung function in pwCF, the average of the highest FEV1 pp is recorded in each quarter of the year. Predicted values are based on the GLI formulae. Lung function measures are aligned with methods used in the United States Cystic Fibrosis Foundation's Patient Registry, whereby annual measures of lung function, weight, and height are reported as an average of the maximum value from each quarter where measurements have been recorded.

The 2025 median lung function is shown in the scatterplot below for 1,002 children and adolescents. 10.6% of participants in the registry are children younger than 6 years of age who do not routinely have lung function information recorded, and a further 7.5% of paediatric registry participants did not have lung function information recorded in 2025 (Figure 2.3).

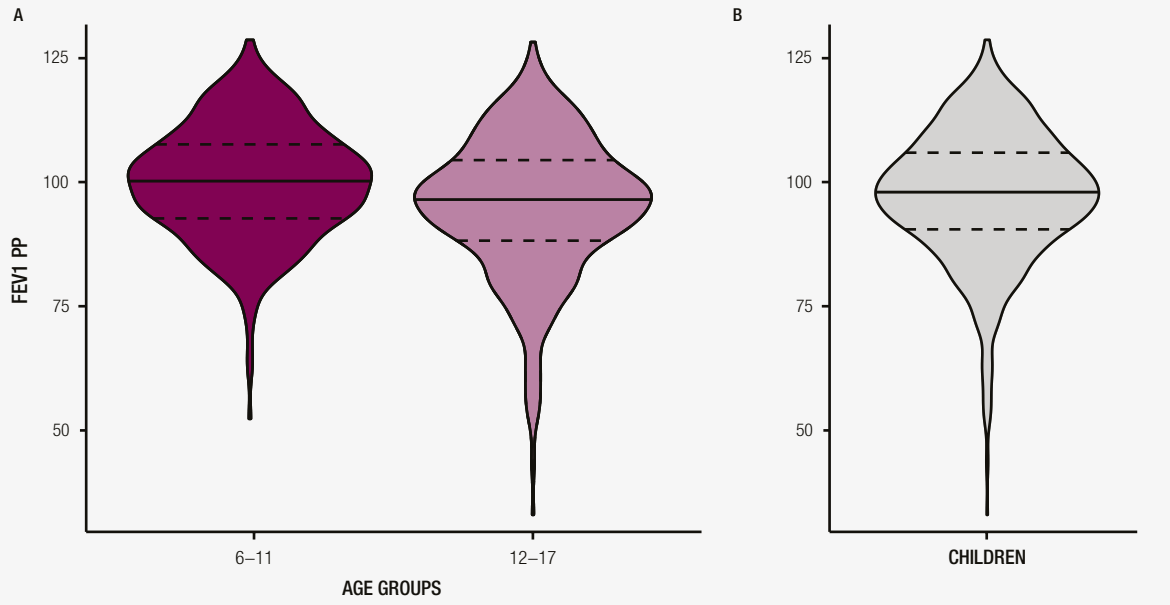
**FIGURE 2.3:** ACFDR PAEDIATRICS 2025: FEV1 PP BY AGE




The solid trend line was estimated using a natural cubic spline with 3 degrees of freedom.
The dotted horizontal line shows the median FEV1pp.
Shaded area represent the 95% confidence intervals.

In 2025, the median FEV1 pp for 6-11 year-olds was 100.2 and for 12-17 year-olds was 96.5. The median FEV1 pp for children and adolescents aged 6-17 years in 2025 was 98.0 (Figure 2.4).

**FIGURE 2.4:** ACFDR PAEDIATRICS 2025: MEDIAN FEV1 PP BY AGE GROUP




The median FEV1 pp for children and adolescents has increased over time. For 6-year-olds, it has remained stable from 98.8 in 2020 to 98.2 in 2025; for 12-year-olds it has increased from 93.5 to 98.5; and for 17-year-olds, it has increased from 81.5 to 96.6 (Figure 2.5).

**FIGURE 2.5:** ACFDR PAEDIATRICS 2020-2025: MEDIAN FEV1 PP OVER TIME

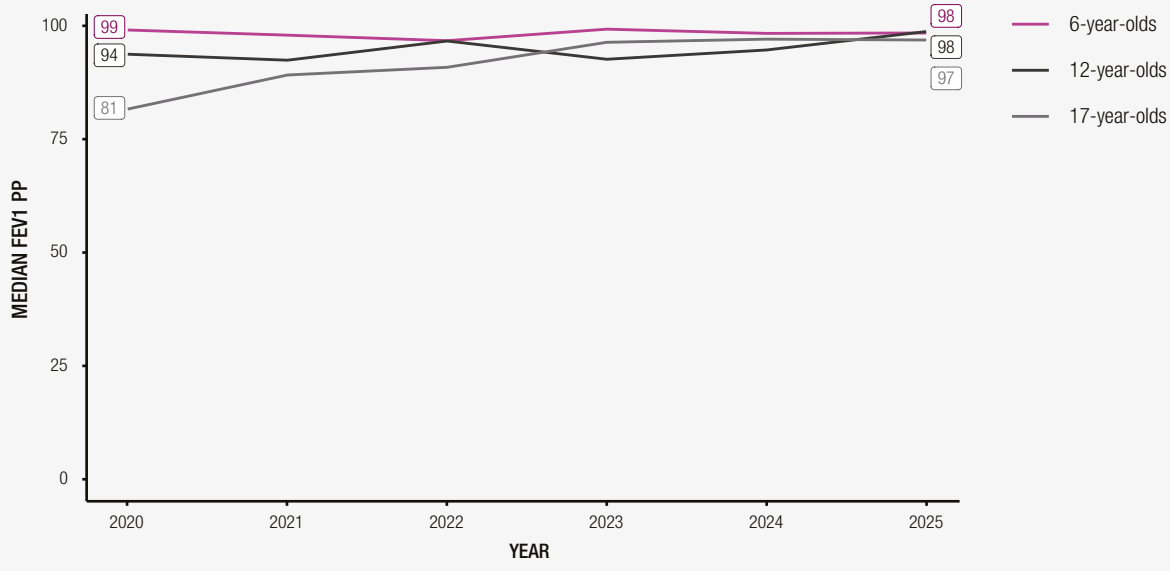
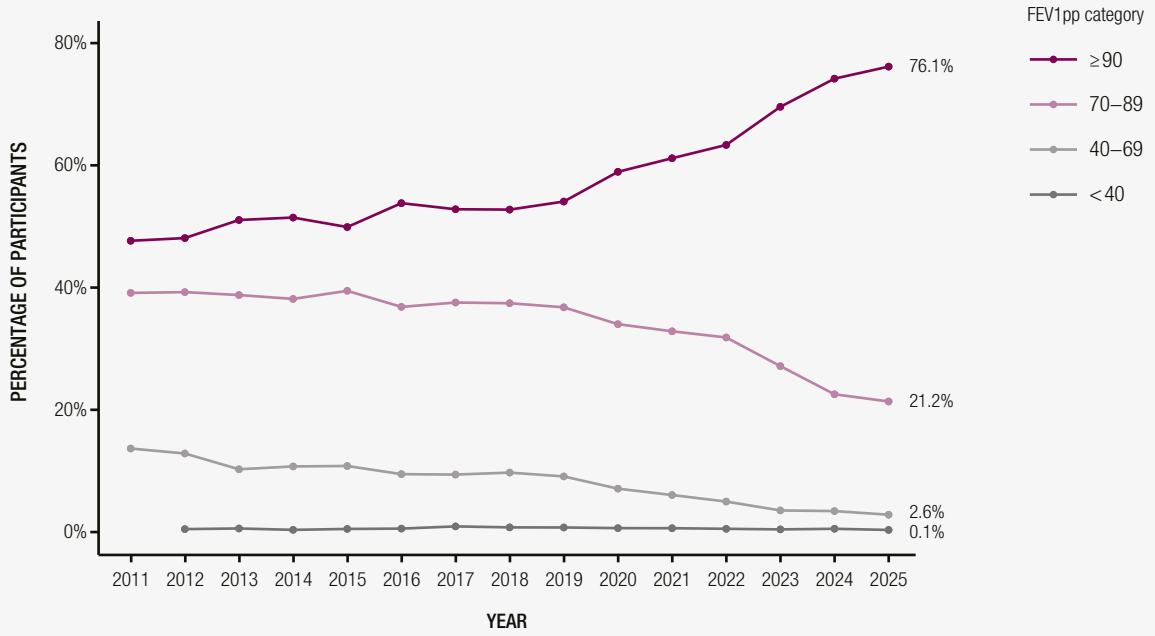


Figure 2.6 depicts the trend in FEV1 pp for children and adolescents with CF since 2011. In recent years, there has been a significant increase in the proportion of pwCF with FEV1 pp scores above 90, from 46.5% in 2011 to 76.1% in 2025.

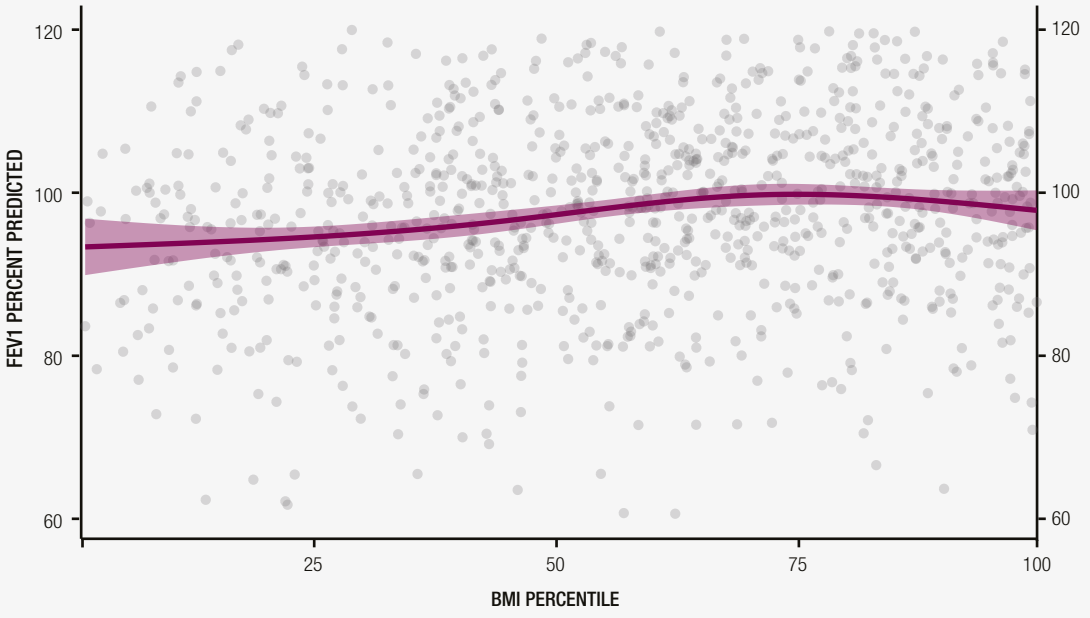
**FIGURE 2.6:** ACFDR PAEDIATRICS 2011-2025: FEV1 PP DISTRIBUTION OVER TIME




## Lung Function and Body Mass Index

The relationship between FEV1 pp and Body Mass Index (BMI) for 2025 is shown in Figure 2.7. In general, as BMI increases, so does FEV1 pp.

**FIGURE 2.7:** ACFDR PAEDIATRICS 2025: FEV1 PP VS BMI PERCENTILE

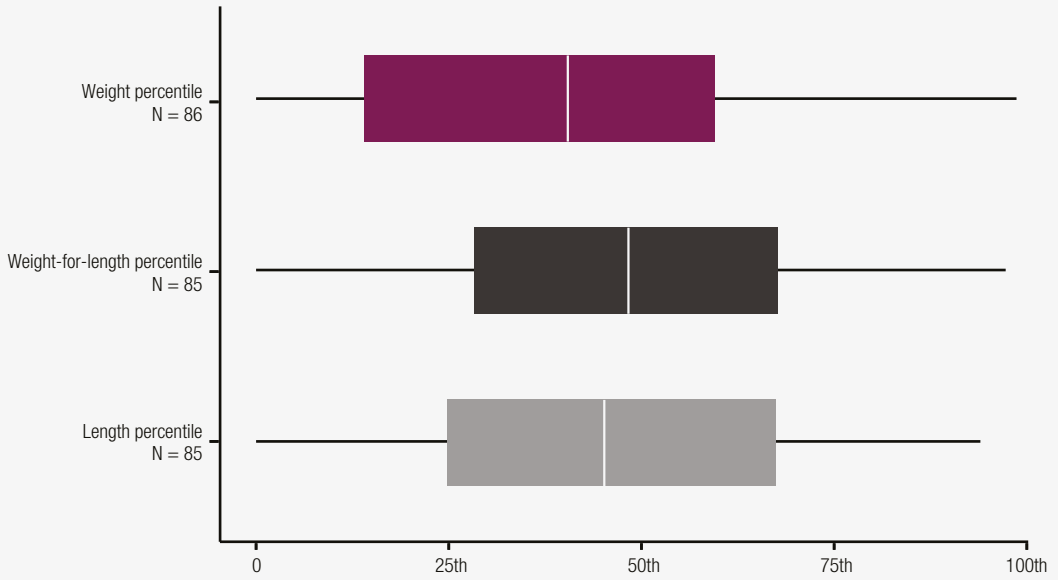



Solid line was estimated using a natural cubic spline with 3 degrees of freedom
Shaded area represents 95% confidence interval

## Nutrition

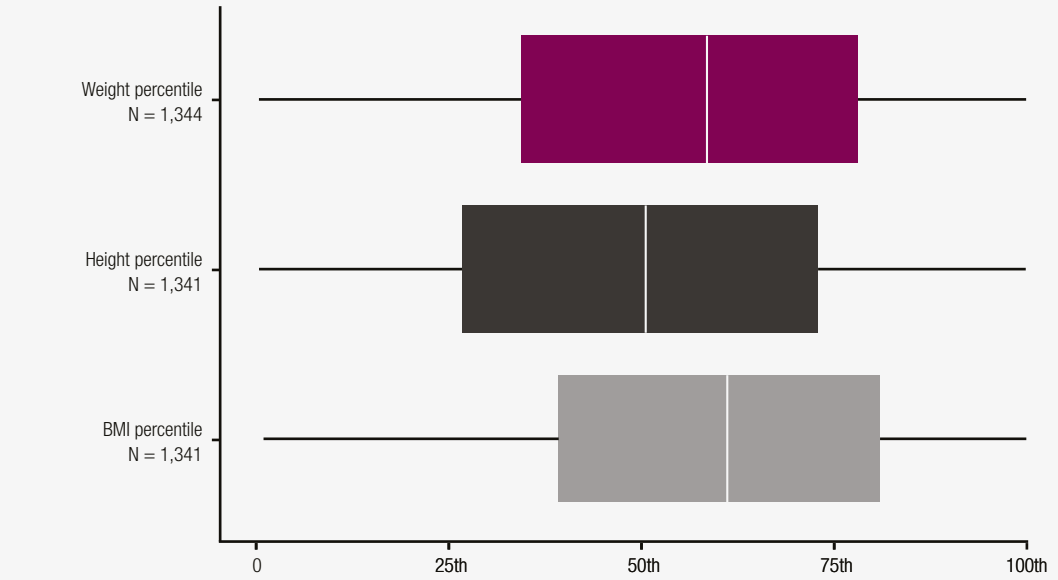
For 2025, infants (<24 months of age) had a median weight on the 40th percentile. The median length was on the 45th percentile; and the median weight-for-length was on the 48th percentile (Figure 2.8).

**FIGURE 2.8:** ACFDR PAEDIATRICS 2025: WEIGHT, LENGTH, AND WEIGHT-FOR-LENGTH PERCENTILES LESS THAN 24 MONTHS




In 2025, for children and adolescents aged 2-17 years, the median weight was on the 58th percentile, the median height was on the 50th percentile, and the median BMI was on the 61st percentile. These figures represent the best weight, height, and BMI per individual averaged over a 12-month period (Figure 2.9).

**FIGURE 2.9:** ACFDR PAEDIATRICS 2025: WEIGHT, HEIGHT, AND BMI PERCENTILES AGE 2-17 YEARS




Children 2–17
Height and BMI percentiles were calculated using WHO growth chart.
Weight percentiles were calculated using CDC growth chart

Nutritional status for the majority of children and adolescents with CF was in the optimal and acceptable BMI percentile ranges for 2025 (Table 2.1), with 17.5% being overweight/obese, and 12.7% having suboptimal nutritional status. The proportion of overweight/obese children/adolescents with CF has reduced slightly from 18.2% in 2024 to 17.5% in 2025.

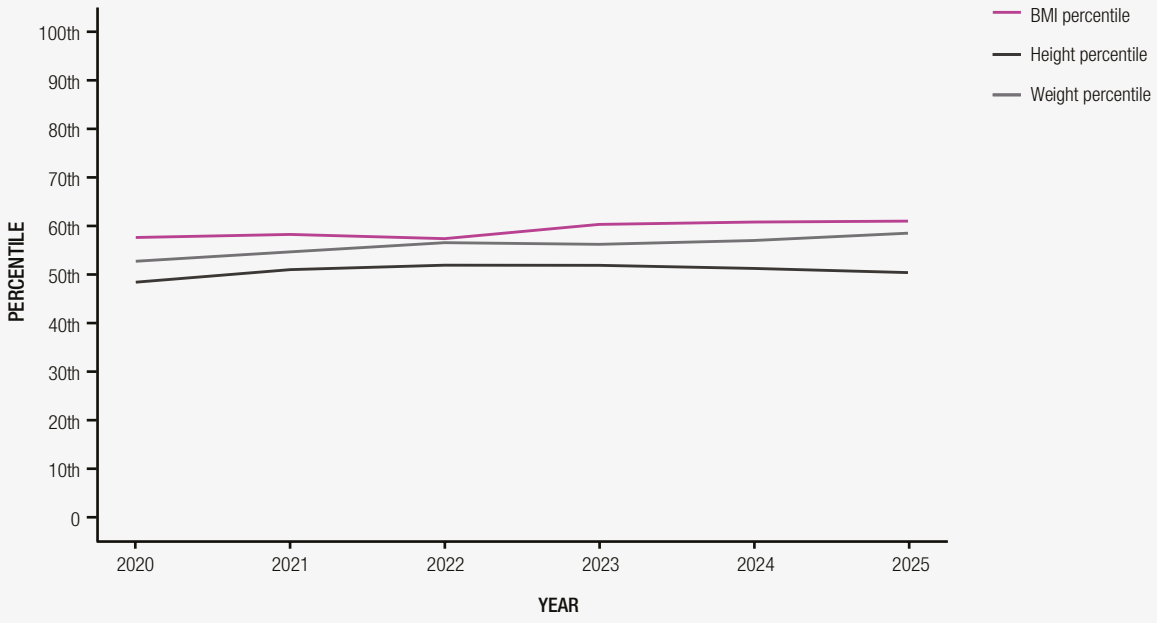
**TABLE 2.1:** ACFDR PAEDIATRICS 2025: <2-17 YEARS NUTRITIONAL STATUS

| Nutritional Status* | <2 years | 2-5 years | 6-11 years | 12-17 years | Total (n = 1,421) |
|---|---|---|---|---|---|
| Optimal/Acceptable | 66 (82.5%) | 194 (67.8%) | 337 (68.5%) | 395 (70.2%) | 992 (69.8%) |
| Overweight/Obese | 0 (0.0%) | 71 (24.8%) | 94 (19.1%) | 84 (14.9%) | 249 (17.5%) |
| Suboptimal/Undernourished | 14 (17.5%) | 21 (7.3%) | 61 (12.4%) | 84 (14.9%) | 180 (12.7%) |

* High BMI (obese range): BMI >95th percentile using CDC growth chart (children and adolescents 2-18 years).
High BMI (overweight range): BMI 85th-95th percentile using CDC growth chart (children and adolescents 2-18 years).
Optimal: weight-for-lengths >50th percentile (infants 0-1 years); BMI 50th-85th percentile using CDC growth chart (children and adolescents 2-18 years).
Acceptable: weight-for-lengths 25th-50th percentile (infants 0-1 years); BMI 25th-50th percentile (children and adolescents 2-18 years).
Suboptimal: weight-for-length 10th-25th percentile (infants 0-1 years); BMI 10th-25th percentile (children and adolescents 2-18 years).
Undernourished: persistent weight for length <10th percentile (infants 0-1 years); BMI <10th percentile (children and adolescents 2-18 years)

Over the last 6 years the median height for children and adolescents aged 2-17 has increased by 2 percentile points (from the 48th percentile to the 50th percentile), and weight has increased by 5 percentile points (from the 53rd percentile to the 58th percentile). As a result, the average BMI also increased by 3 percentile points during this time, from the 58th percentile to the 61st percentile (Figure 2.10).

**FIGURE 2.10:** ACFDR PAEDIATRICS 2020-2025: 2-17 YEARS MEDIAN NUTRITIONAL PERCENTILES

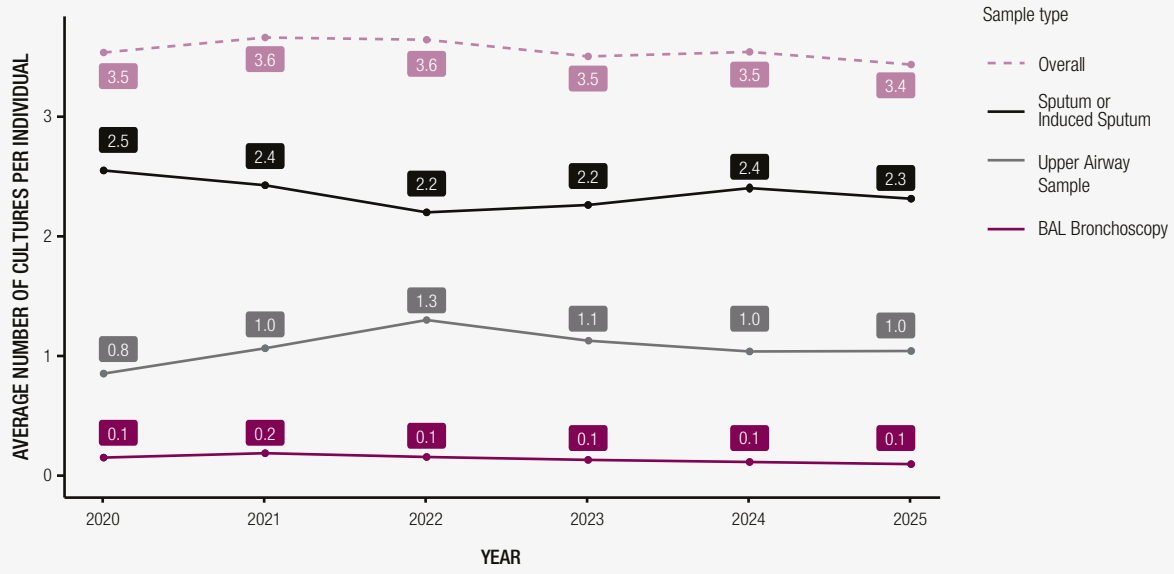


## Microbiology

The average number of respiratory samples collected per child each year over a 5-year period is shown in Figure 2.11. The overall number of respiratory samples stayed stable from 3.5 per person in 2020 to an average of 3.4 samples in 2025.

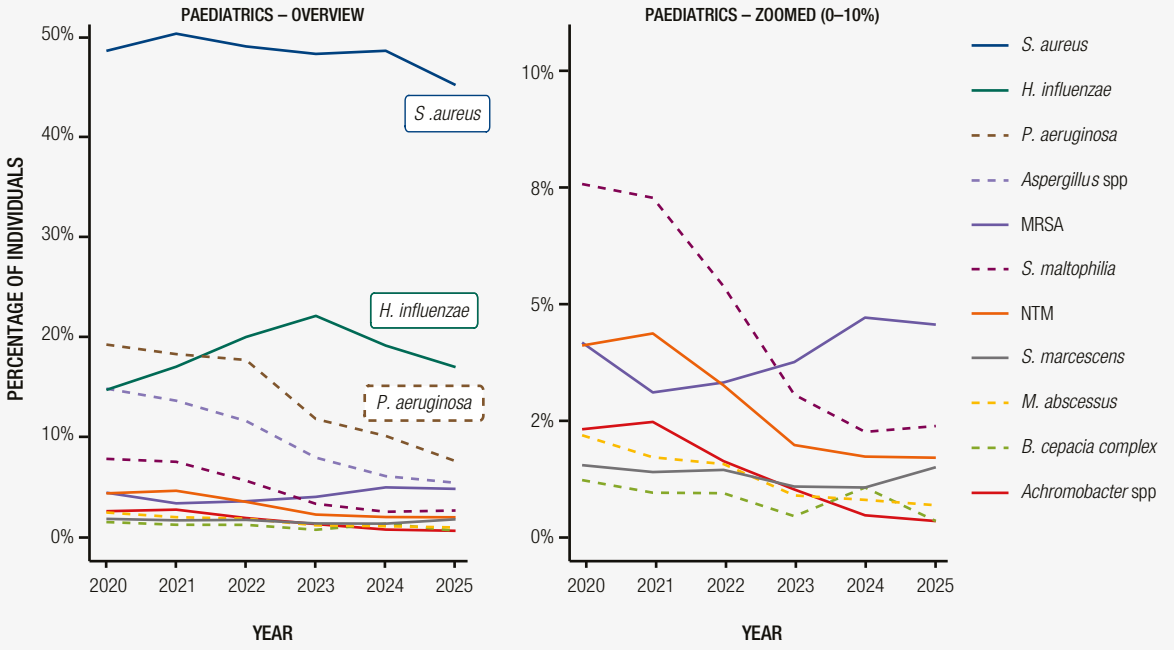
When considering the specific sample types, sputum/induced sputum samples accounted for the greatest proportion, at 2.3 samples per person in 2025. Upper airway samples averaged 1 per person (throat swabs, cough swabs, nasopharyngeal swabs and sinus wash), and bronchoscopy/BAL samples averaged 0.1 in 2025.

**FIGURE 2.11:** ACFDR PAEDIATRICS 2020-2025: MEAN NUMBER OF RESPIRATORY SAMPLES PER INDIVIDUAL



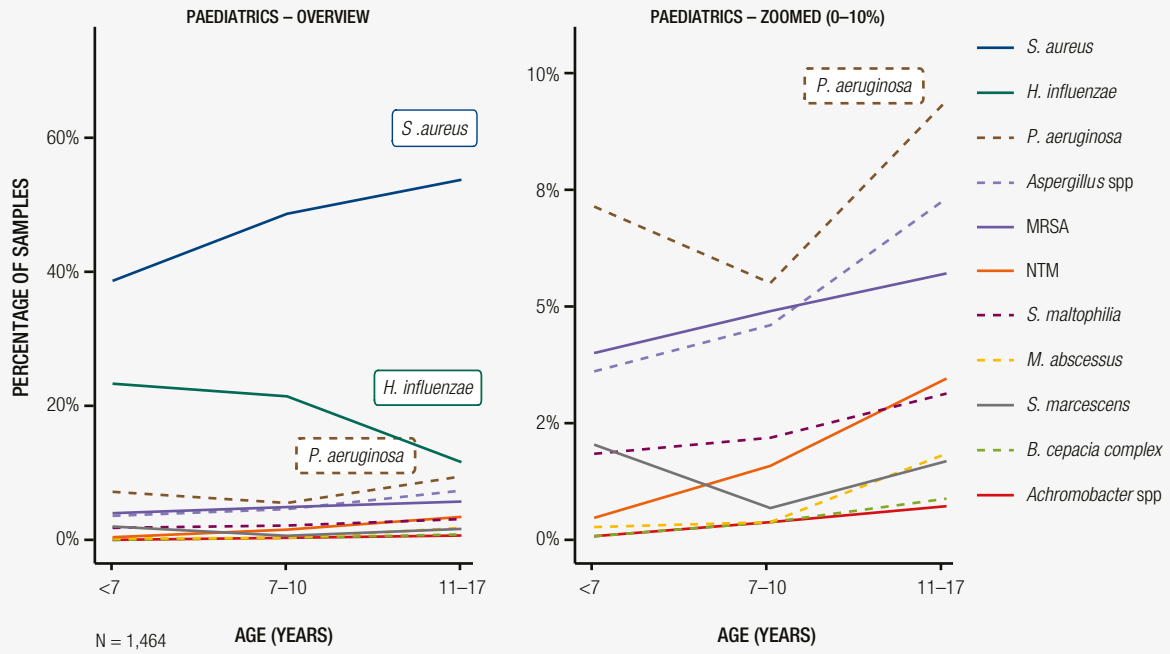

The prevalence of some of the most common paediatric microorganisms has changed over the last 6 years. The prevalence of *S. aureus* has decreased to 44.9% in 2025; the prevalence of *H. influenzae* has decreased to 16.7%; the prevalence of *P. aeruginosa* has decreased to 7.3% in 2025; and *Aspergillus* spp has decreased to 5.1% in 2025. The prevalence of less common paediatric microorganisms has mostly decreased (Figure 2.12).

**FIGURE 2.12:** ACFDR PAEDIATRICS 2020-2025: PREVALENCE OF RESPIRATORY MICROBIOLOGY



Of the 1,464 microbiology culture samples collected in 2025, the most common pathogen across all age groups was *S. aureus*, with the prevalence increasing with age (Figure 2.13, Table 2.2). The next most common pathogen was *H. influenzae*, followed by *P. aeruginosa*. The prevalence of *S. aureus* increases with increasing age, with a prevalence of 38.0% for samples from children <7 years, 47.9% in children and adolescents 7-10 years and 52.9% in adolescents 11-17 years. The prevalence of *H. influenzae* decreases with age, being 22.9% of samples from children <7 years, 21.1% for children and adolescents 7-10 years, and 11.4% for adolescents aged 11-17 years.

**FIGURE 2.13:** ACFDR PAEDIATRICS 2025: PREVALENCE OF RESPIRATORY MICROORGANISMS BY AGE



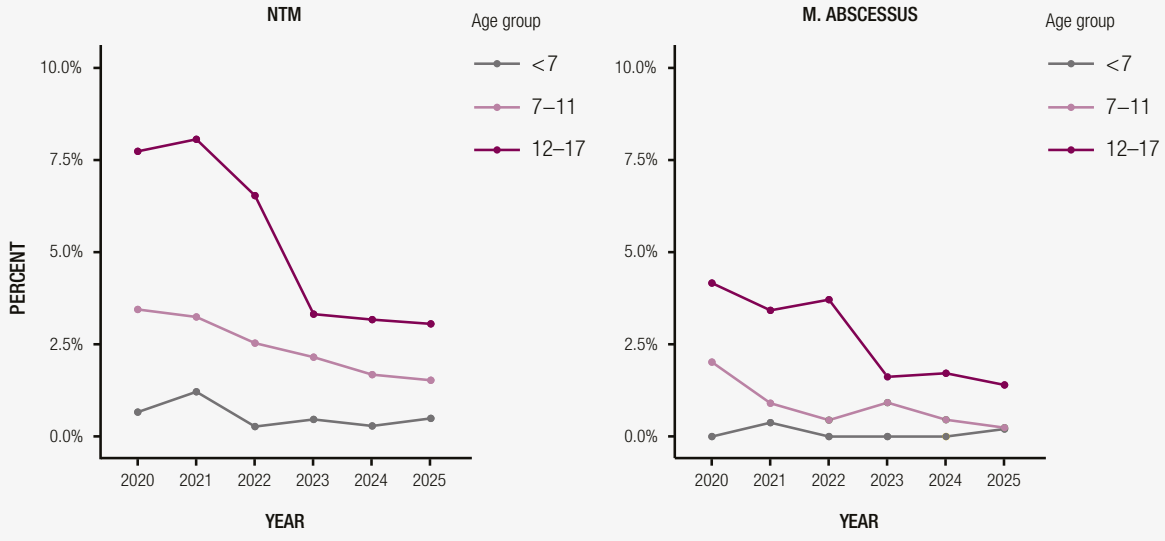
For children younger than seven years, 75 lower airway samples were collected by bronchoalveolar lavage (BAL) in 2025. The most common organisms identified in this age group in 2025 included *S. aureus* (26.7%), *H. influenzae* (24.0%), *Aspergillus* spp (16.0%), and *P. aeruginosa* (8.0%) (Table 2.2).

**TABLE 2.2:** ACFDR PAEDIATRICS 2025: PREVALENCE OF RESPIRATORY MICROORGANISMS BY AGE

| | BAL samples | All samples | | |
|---|---|---|---|---|
| | <7 years | <7 years | 7-10 years | 11-17 years |
| Total patients in the age range | 490 | 490 | 334 | 654 |
| Samples taken | 75 | 510 | 332 | 622 |
| Patients who provided a sample | 75 | 449 | 316 | 598 |
| *S. aureus* | 20 / 75 (26.7%) | 194 / 510 (38.0%) | 159 / 332 (47.9%) | 329 / 622 (52.9%) |
| *H. influenzae* | 18 / 75 (24.0%) | 117 / 510 (22.9%) | 70 / 332 (21.1%) | 71 / 622 (11.4%) |
| *P. aeruginosa* | 6 / 75 (8.0%) | 36 / 510 (7.1%) | 18 / 332 (5.4%) | 58 / 622 (9.3%) |
| *Aspergillus spp* | 12 / 75 (16.0%) | 18 / 510 (3.5%) | 15 / 332 (4.5%) | 45 / 622 (7.2%) |
| *B. cepacia complex* | 0 / 75 (0.0%) | 0 / 510 (0.0%) | <5 | 5 / 622 (0.8%) |
| MRSA | <5 | 20 / 510 (3.9%) | 16 / 332 (4.8%) | 35 / 622 (5.6%) |
| *Achromobacter spp* | 0 / 75 (0.0%) | 0 / 510 (0.0%) | <5 | <5 |
| *S. maltophilia* | <5 | 9 / 510 (1.8%) | 7 / 332 (2.1%) | 19 / 622 (3.1%) |
| *S. marcescens* | 0 / 75 (0.0%) | 10 / 510 (2.0%) | <5 | 10 / 622 (1.6%) |
| *Nontuberculous mycobacteria* | 0 / 75 (0.0%) | <5 | 5 / 332 (1.5%) | 21 / 622 (3.4%) |
| *M. abscessus* | 0 / 75 (0.0%) | <5 | <5 | 11 / 622 (1.8%) |

In 2025, the prevalence of NTM among children <7 years was 0.4%, in 7-11 year-olds was 1.4%, and in 12-17 year-olds was 3.0%. The prevalence of *M. abscessus* among children <7 years was 0.2%, among 7-11 year-olds was 0.2% and among 12-17 year-olds was 1.4%. Since 2020, there has been a decline in isolation of NTM (Figure 2.14).

**FIGURE 2.14:** ACFDR PAEDIATRICS 2019-2025: PREVALENCE OF NTM INFECTIONS BY AGE



## 2.3 CF MANAGEMENT

### Clinic Visits

Table 2.3 and Figure 2.15 show the total number of clinical visits for the paediatric population in the registry per year over the last 4 years. The total number of clinical visits was highest in 2022 (7,103 encounters), with visits declining since then to a total of 5,686 in 2025.

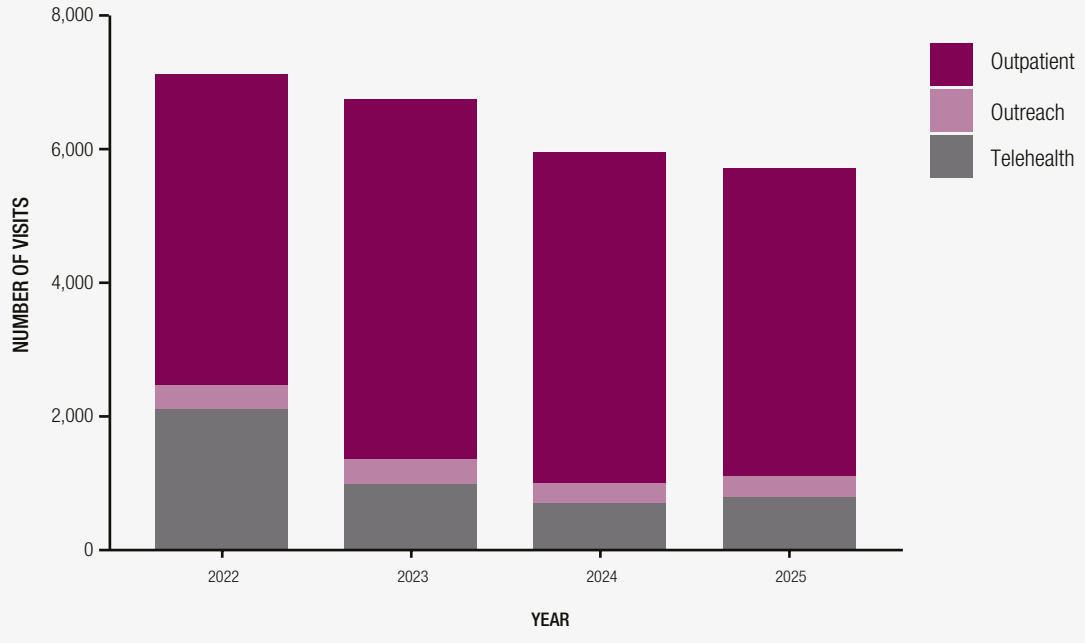
The nature of clinical visits has also changed during this time. The proportion of telehealth visits (audio and/or visual visits conducted at home or in a healthcare setting) for children and adolescents were 29.4% in 2022, decreasing to a low level of 11.4% in 2024, and then increasing slightly to 13.5% in 2025. This change reflects an increase in the proportion of outpatient visits, from 65.6% in 2022 to 81.0% in 2025 (Table 2.3 and Figure 2.15).

Historical results have been recalculated using year-specific active populations and age classifications based on age at December 31st of each reporting year, rather than age at clinic visit. This change in methodology has slightly reduced the number of pwCF classified as paediatric and therefore the number of paediatric clinical visits.

**TABLE 2.3:** ACFDR PAEDIATRICS 2022-2025: TOTAL NUMBER OF VISITS RECORDED PER YEAR

| Visit type | 2022 | 2023 | 2024 | 2025 |
|---|---|---|---|---|
| Outpatient | 4,658 (65.6%) | 5,383 (80.1%) | 4,950 (83.5%) | 4,603 (81.0%) |
| Outreach | 355 (5.0%) | 363 (5.4%) | 303 (5.1%) | 315 (5.5%) |
| Telehealth | 2,090 (29.4%) | 972 (14.5%) | 675 (11.4%) | 768 (13.5%) |
| Total | 7,103 (100.0%) | 6,718 (100.0%) | 5,928 (100.0%) | 5,686 (100.0%) |

**FIGURE 2.15:** ACFDR PAEDIATRICS 2022-2025: TOTAL NUMBER OF VISITS RECORDED BY YEAR



## CF Standards of Care

The Australian CF Standards of Care for pwCF recommend four clinic visits per year. In 2025 the number of children/adolescents with CF who had at least 4 clinic visits per year was 891 (60.3% of all children with CF) overall, compared to 1,056 (65.3%) in 2022 (Table 2.4). In 2025, the proportion of children and adolescents receiving four or more clinical encounters were highest among children <2 years old at 73.9%, followed by 64.6% for 2-6 year-olds, 61.8% for 7-11 year-olds and 54.0% for adolescents.

**TABLE 2.4:** ACFDR PAEDIATRICS 2022-2025: NUMBER AND PROPORTION WITH 4 OR MORE CLINICAL VISITS BY AGE

| | NUMBER (%) WITH 4+ CLINIC VISITS PER YEAR | | | |
|---|---|---|---|---|
| Age | 2022 | 2023 | 2024 | 2025 |
| <2 | 101 (70.0%) | 100 (79.0%) | 61 (69.0%) | 68 (73.9%) |
| 2-6 | 263 (66.0%) | 274 (68.0%) | 263 (65.0%) | 257 (64.6%) |
| 7-11 | 277 (62.0%) | 298 (68.0%) | 278 (63.0%) | 257 (61.8%) |
| 12-17 | 415 (66.0%) | 402 (65.0%) | 320 (55.0%) | 309 (54.0%) |
| **Total** | 1,056 (65.3%) | 1,074 (68.0%) | 922 (61.0%) | 891 (60.3%) |

While the majority of children/adolescents had at least 4 visits per year, 13.0% of children aged under 2 years, 16.8% of children aged 2-6 years, 20.0% of children aged 7-11 years, and 26.9% of adolescents aged 12-17 had 3 visits in 2025. Less than 10% of all children and adolescents with CF had one or no clinical visits in 2025 (Figure 2.16).

**FIGURE 2.16:** ACFDR PAEDIATRICS 2025: PROPORTION OF PARTICIPANTS HAVING 4 OR MORE CLINICAL VISITS

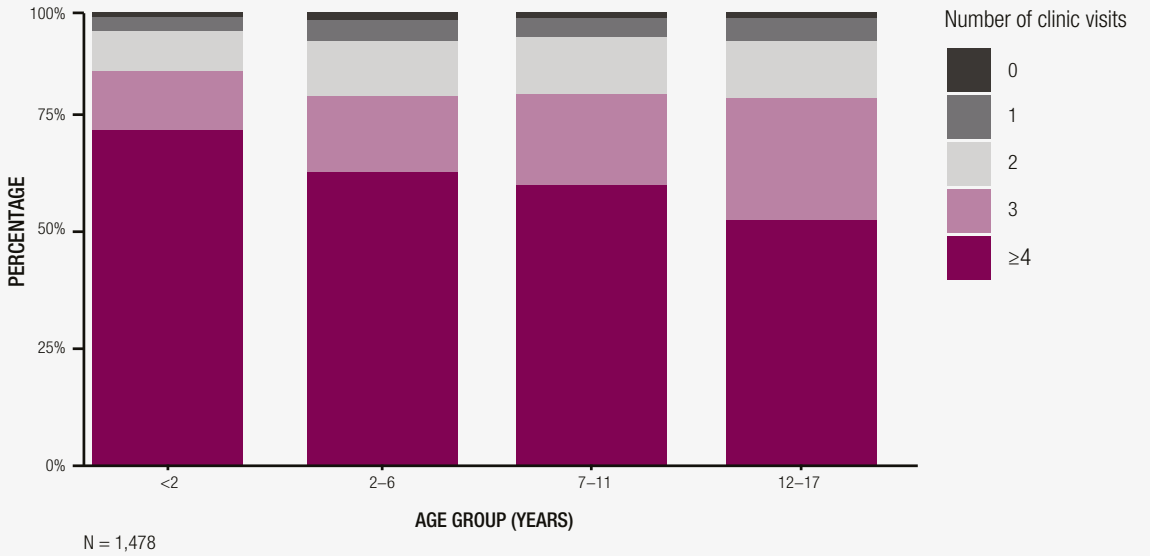


Table 2.5 shows the average number of clinical encounters per child or adolescent with CF in 2025. The average number of clinic visits decreased with age, with children aged less than two having the highest average number of clinic visits per person (5.3) and 12-17 year-olds having the fewest visits (3.7).

**TABLE 2.5:** ACFDR PAEDIATRICS 2025: AVERAGE NUMBER OF CLINICAL ENCOUNTERS PER PERSON

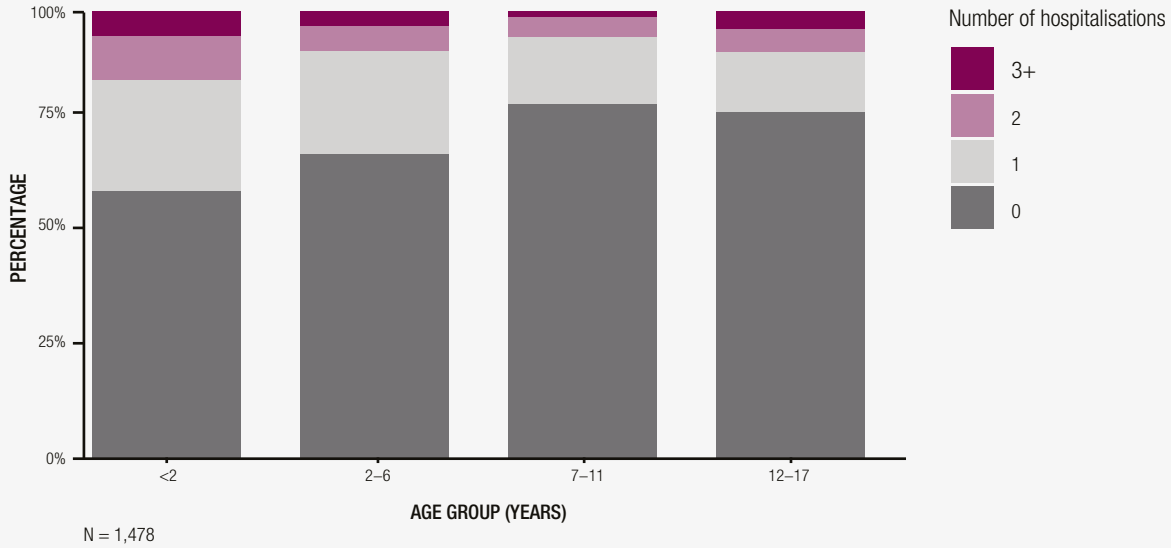
| Age | Average number of clinic visits |
|---|---|
| <2 | 5.3 |
| 2-6 | 3.8 |
| 7-11 | 3.9 |
| 12-17 | 3.7 |
| **Total** | 3.9 |

## Hospitalisations

There was a total of 665 hospitalisations for children and adolescents in 2025. A majority (59.8%) of children younger than 2 years of age did not have any hospitalisations in 2025, while 25.0% had 1 hospitalisation, 9.8% had 2 hospitalisations, and 5.4% had 3 or more hospitalisations (Figure 2.17). In the 2-6 year age range in 2025, 68.1% had no hospitalisations, with 23.1% having 1, 5.5% having 2, and 3.3% having 3 or more hospitalisations.

Lower proportions of hospitalisations were noted for older children and adolescents. For the 7-11 age group, 79.3% had no hospitalisations, while 14.9% had 1, 4.6% had 2, and 1.2% had at least 3 hospitalisations. Similarly, among adolescents aged 12-17, 77.6% had no hospitalisations, while 13.3% had 1, 5.1% had 2, and 4.0% had at least 3 hospitalisations (Figure 2.17).

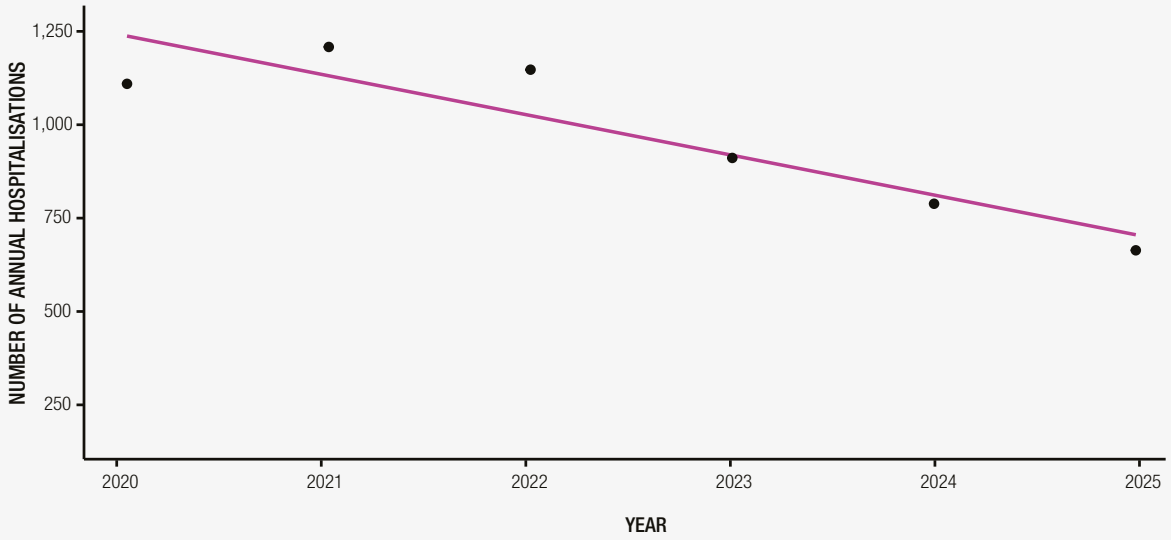
**FIGURE 2.17:** ACFDR PAEDIATRICS 2025: HOSPITALISATIONS BY AGE GROUP



## Paediatric Hospitalisations Over Time

Paediatric hospitalisations have reduced significantly since 2020, with a decrease from 1,112 admissions in 2020 to 665 admissions in 2025, a reduction of 40.2% over the last 6 years (Figure 2.18).

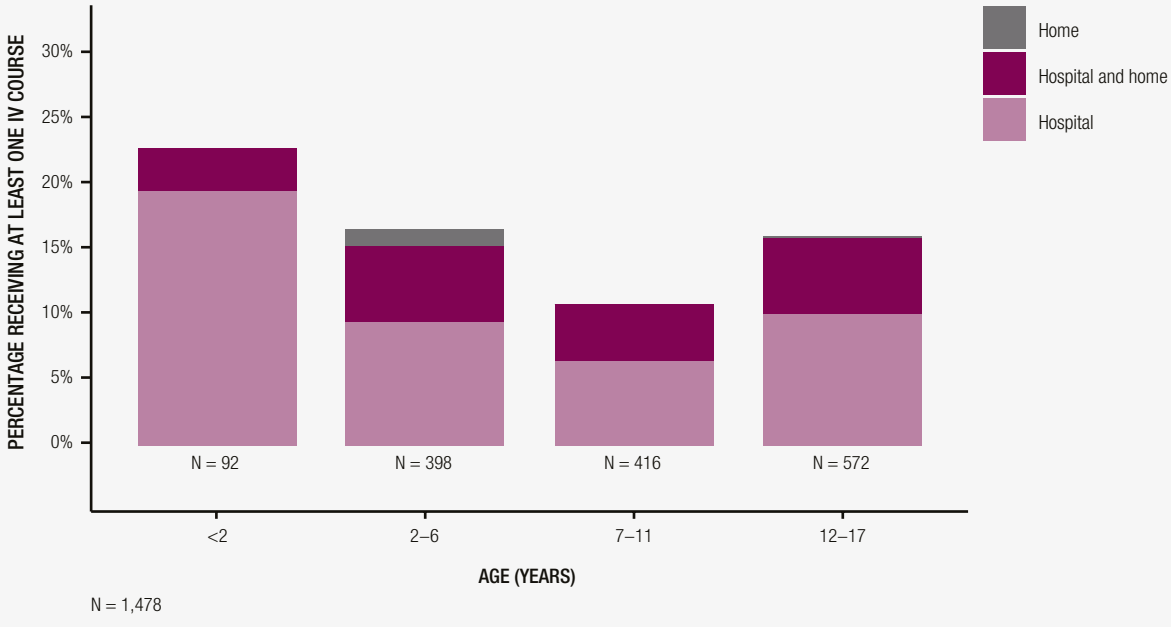
**FIGURE 2.18:** ACFDR PAEDIATRICS 2020-2025: HOSPITALISATIONS PER YEAR




## IV Antibiotic Therapy

In 2025, the vast majority of intravenous (IV) therapy treatments for children occurred in hospital, with 19.6% of children <2 years, 9.5% of children 2-6 years, 6.5% of 7-11 year-olds, and 10.1% of 12-17 year-olds reporting receiving hospital IV therapy. The use of combined hospital and home IV antibiotics was highest among both 2-6 year-olds and 12-17 year-olds at 5.8%, followed by 4.3% of 7-11 year-olds and 3.3% of children less than 2 years of age. In 2025, very few children or adolescents (0.4%) received solely home-based IV therapy (Figure 2.19).

**FIGURE 2.19:** ACFDR PAEDIATRICS 2025: PARTICIPANTS RECEIVING AT LEAST ONE COURSE OF IV ANTIBIOTIC THERAPY



The median duration of IV antibiotic therapy in hospital, at home, and for combined therapy at home and in hospital was 14 days (Table 2.6).

**TABLE 2.6:** ACFDR PAEDIATRICS 2025: MEDIAN AND MEAN DAYS IV ANTIBIOTIC THERAPY

| IV antibiotic therapy location | N | Median days receiving therapy | Mean days receiving therapy | Range (days) |
|---|---|---|---|---|
| Home | 6 | 14 | 12.8 | 7 - 21 |
| Hospital | 139 | 14 | 16.0 | 1 - 116 |
| Hospital and home | 77 | 14 | 16.7 | 4 - 38 |

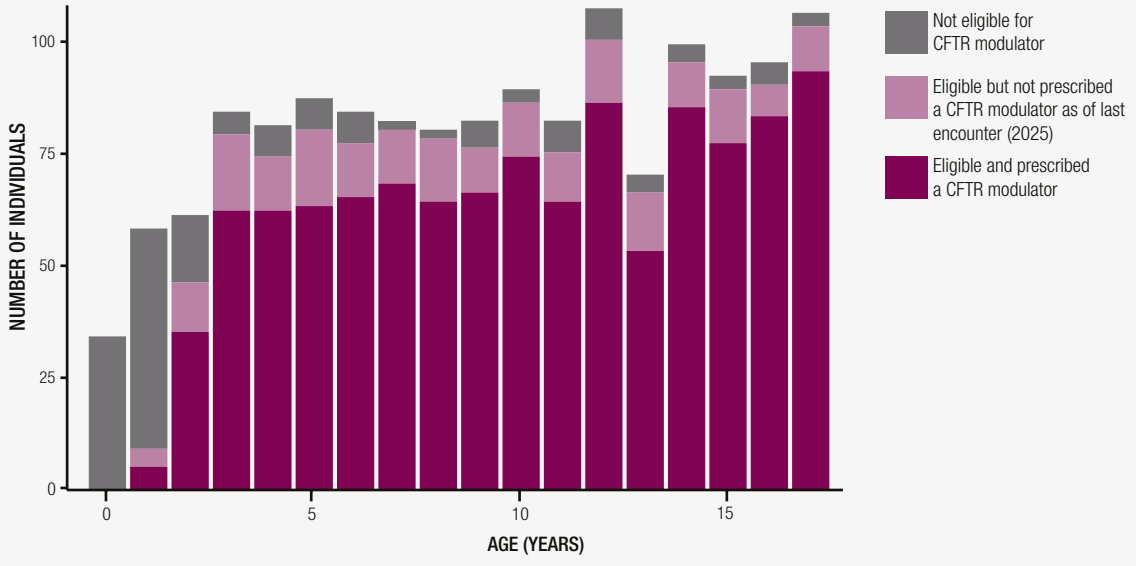
## CFTR Modulators

Data were calculated from pwCF who were on a modulator as of December 31st 2025. Data presented here reflect only those pwCF (excluding lung transplant recipients) who had CFTR modulator data entered into the registry, which generally includes those prescribed modulators available via the PBS. Please refer to section 1.6 for more information regarding CFTR modulator eligibility criteria.

Of the children and adolescents in the registry who have not had a lung transplant, eligibility status for a CFTR modulator was known/recorded for 99.7% (1,473 people) in 2025.

Of these, 170 children and adolescents (11.5%) were not eligible for a modulator; 75.0% (1,105 children and adolescents) were eligible and prescribed a modulator; and 13.4% (198 children and adolescents) were eligible but not prescribed a modulator in 2025. Eligibility was determined based on age at the start of the year to identify those eligible during that year, whereas the age groups in the data are defined by age at the end of the year (Figure 2.20).

**FIGURE 2.20:** ACFDR PAEDIATRICS 2025: CFTR ELIGIBILITY AND PRESCRIBING BY AGE

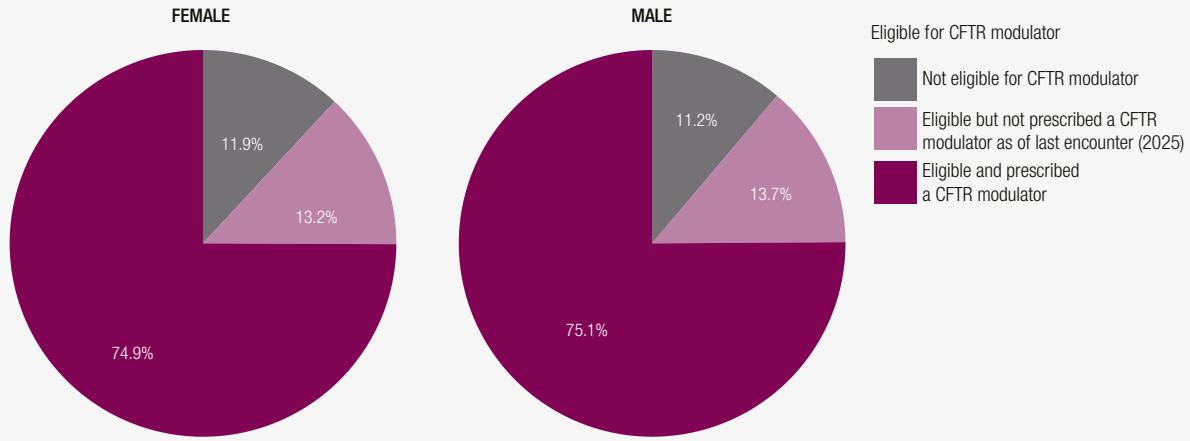



N = 1,473 individuals with known eligibility

Figure 2.21 below shows the eligibility and prescribing information for male and female children and adolescents with CF. The denominator is 1,473 children and adolescents with CF where eligibility for a modulator is recorded in the registry based on genotype. On December 31st 2025, 11.9% of females and 11.2% of males were not eligible for a CFTR modulator.

Of the paediatric population, 74.9% of females and 75.1% of males were eligible and prescribed a modulator, whereas 13.2% of females and 13.7% of males were eligible but not prescribed a modulator, for the first time showing similar prescribing rates for both males and females.

**FIGURE 2.21:** ACFDR PAEDIATRICS 2025: ELIGIBILITY AND PRESCRIBING OF CFTR MODULATORS BY GENDER




N = 1,473 individuals with known eligibility

Among children with CF, uptake of CFTR modulators – particularly Trikafta (ETI) – increases markedly with age. In children aged <6 years, over half (53.3%) were receiving Trikafta, and less than one fifth (18.0%) were not eligible for any CFTR modulator therapy, reflecting historical regulatory and reimbursement restrictions for this age range. As children enter higher age bands where eligibility criteria broaden, Trikafta use becomes dominant: 77.8% of those aged 6-11 and 81.9% of those aged 12-17 were receiving Trikafta. Across all paediatric groups, only a small proportion were receiving older-generation modulators (3.9%-13.6%), and between 11.6% and 15.1% were eligible but not receiving treatment, suggesting potential delays in access or clinical considerations despite eligibility (Table 2.7).

**Due to the majority of pwCF accessing Trikafta in 2025, the following modulator data has been categorised as 'Trikafta' or 'other CFTR modulator'.**

**TABLE 2.7:** ACFDR PAEDIATRICS 2025: MODULATOR USE BY AGE

| Age (Years) | Total N | Trikafta N (%) | Other CFTR modulator N (%) | Eligible but not receiving CFTR modulator N (%) | Not eligible for CFTR modulator N (%) |
|---|---|---|---|---|---|
| <6 | 405 | 216 (53.3%) | 55 (13.6%) | 61 (15.1%) | 73 (18.0%) |
| 6-11 | 499 | 388 (77.8%) | 24 (4.8%) | 70 (14.0%) | 17 (3.4%) |
| 12-17 | 569 | 466 (81.9%) | 22 (3.9%) | 66 (11.6%) | 15 (2.6%) |

**Note**: Table 2.7 describes CFTR modulator use as of December 31st 2025, with age categories based on age at year end. Eligibility was assessed using age at the beginning of the year; therefore, counts may differ slightly from the eligibility/prescribing summaries presented elsewhere in the report.

Table 2.8 describes the **reasons for permanent or temporary cessation and/or change to modulator prescriptions** for children and adolescents with CF throughout 2025. **Switching from another modulator to Trikafta was the most common reason for discontinuation** (51 pwCF). Eleven pwCF discontinued a modulator due to their involvement in a clinical trial. Some pwCF discontinued a modulator due to adverse effects, primarily liver impairment or intolerance (17 pwCF), mental health reasons (9 pwCF), or another intolerance or adverse event otherwise unspecified (25 pwCF). Of all pwCF who discontinued Trikafta in 2025, 34.4% discontinued the modulator due to an intolerance or adverse event otherwise unspecified.

**TABLE 2.8:** ACFDR PAEDIATRICS 2025: REASON FOR DISCONTINUATION/CHANGE OF MODULATOR

| Reasons for discontinuation/change | Total (N=126) | Trikafta (N=61) | Other CFTR modulator (N=65) |
|---|---|---|---|
| Switch to other CFTR modulator | 52 (41.3%) | <5 | 51 (78.5%) |
| Other intolerance/adverse event | 25 (19.8%) | 21 (34.4%) | <5 |
| Liver impairment/intolerance | 17 (13.5%) | 15 (24.6%) | <5 |
| Clinical trial | 11 (8.7%) | 9 (14.8%) | <5 |
| Mental health | 9 (7.1%) | 6 (9.8%) | <5 |
| Other reason (non-adverse) | 6 (4.8%) | <5 | <5 |
| Rash | <5 | <5 | 0 (0.0%) |
| Patient preference/non-adherence | <5 | <5 | 0 (0.0%) |
| Weight gain | <5 | <5 | 0 (0.0%) |

In 2025, **94.5% of children and adolescents on modulators were prescribed a standard dose**. Figure 2.22 shows the proportion of patients aged <6, 6-11, and 12-17 years prescribed standard and non-standard doses of Trikafta and other CFTR modulators. Six percent of all children and adolescents on Trikafta were prescribed non-standard doses. All children and adolescents who received a non-Trikafta modulator were prescribed standard doses.

**FIGURE 2.22:** ACFDR PAEDIATRICS 2025: STANDARD AND NON-STANDARD DOSE BY AGE AND MODULATOR


**Note**: Based on modulator status as of December 31st 2025. Percentages exclude missing dose information.

## 2.4 COMPLICATIONS AND THERAPIES

### CF Pulmonary Disease

In 2025 there were 13 paediatric hospitalisations for haemoptysis, none of which required embolisation. No children or adolescents with CF required hospitalisation for pneumothorax in 2025.

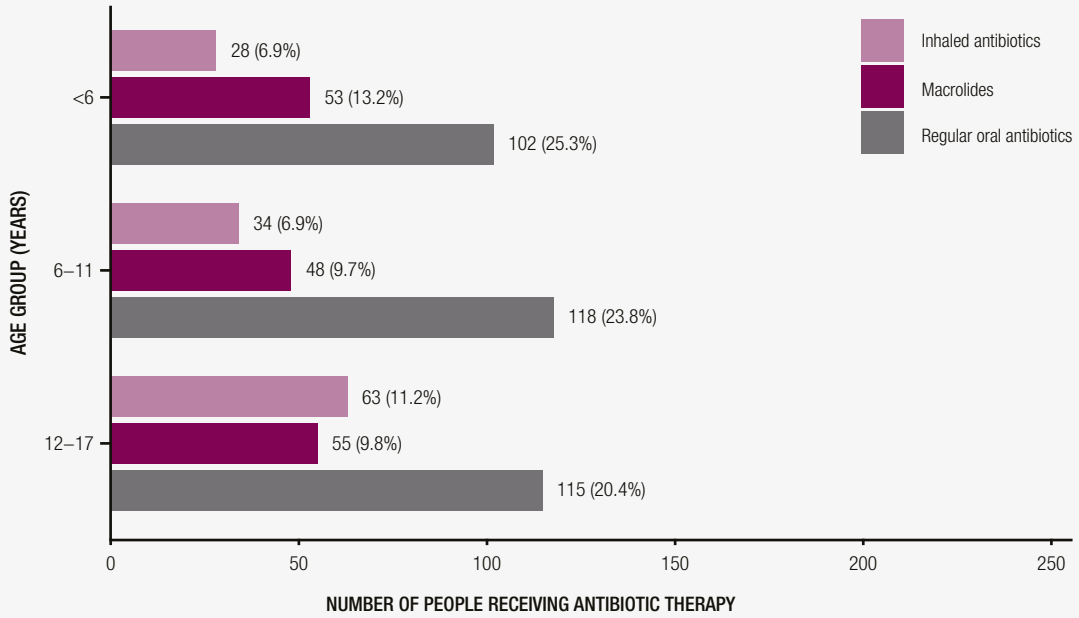
### CF Pulmonary Therapies — Maintenance Antibiotics

The use of maintenance antibiotic therapy for children and adolescents is depicted in figures 2.23 and 2.24A, B, and C. In 2025, use of inhaled antibiotics were more commonly used by older children, with 28 (6.9%) children under 6, 34 (6.9%) children aged 6-11 and 63 (11.2%) adolescents aged 12-17 being administered inhaled antibiotics.

Macrolide use was similar regardless of age with 53 (13.2%) children under 6 years, 48 (9.7%) children 6-11, and 55 (9.8%) 12-17 year-olds using macrolides in 2025.

Regular oral antibiotics use was similar across all age groups, with 102 (25.3%) children under 6,118 (23.8%) children 6-11, and 115 (20.4%) adolescents aged 12-17 being prescribed regular oral antibiotics.

**FIGURE 2.23:** ACFDR PAEDIATRICS 2025: MAINTENANCE ANTIBIOTIC THERAPY BY AGE GROUP




N = 1,462

Given that the majority of pwCF are now receiving modulator treatments, some adjuvant antibiotic treatment use has reduced over time. Figures 2.24A, B, and C show **usage of regular oral antibiotics, macrolides and inhaled antibiotics over a six-year period**.

For children less than 6 years, regular oral antibiotics use has decreased over time, while inhaled antibiotics and macrolide use has remained relatively stable (at around 10-15%). For children aged 6-11 years, there has been a continuing decrease in regular oral antibiotics and inhaled antibiotics, while macrolide use has remained stable at around 10%. For adolescents aged 12-17 years, use of regular oral antibiotics, inhaled antibiotics, and macrolides have all decreased since 2020. Inhaled antibiotic use showed the most significant reduction for this cohort, decreasing by approximately two-thirds by 2025.

**FIGURE 2.24A:** ACFDR PAEDIATRICS 2020-2025: MAINTENANCE ANTIBIOTIC THERAPY FOR CHILDREN UNDER 6 YEARS

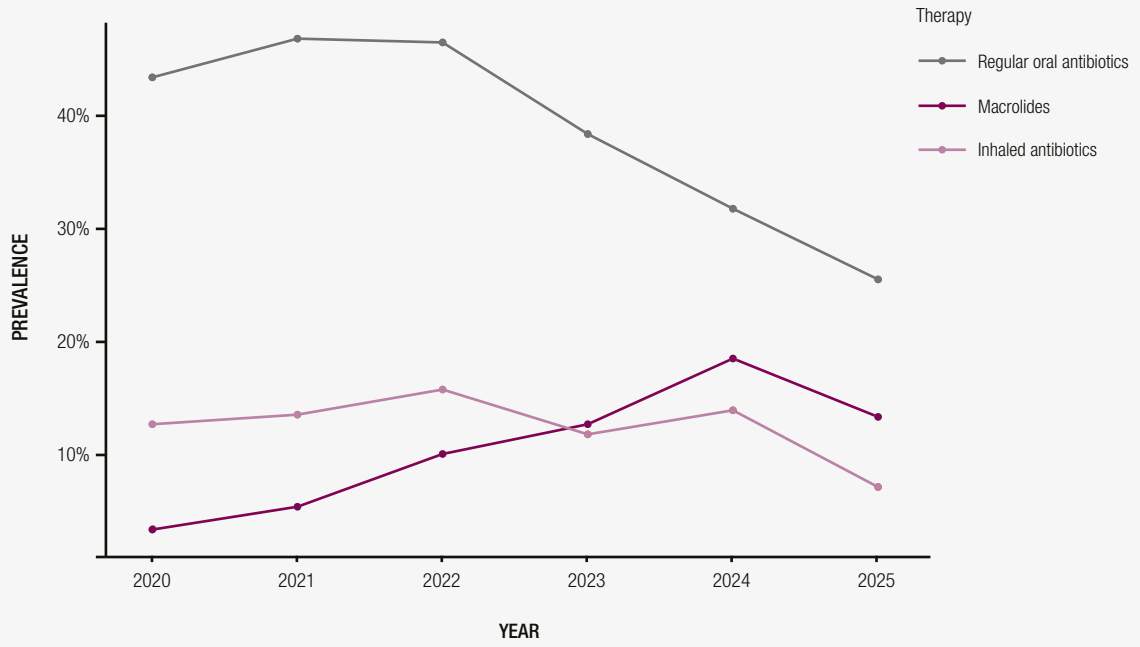



**FIGURE 2.24B:** ACFDR PAEDIATRICS 2020-2025: MAINTENANCE ANTIBIOTIC THERAPY FOR CHILDREN AGED 6-11 YEARS

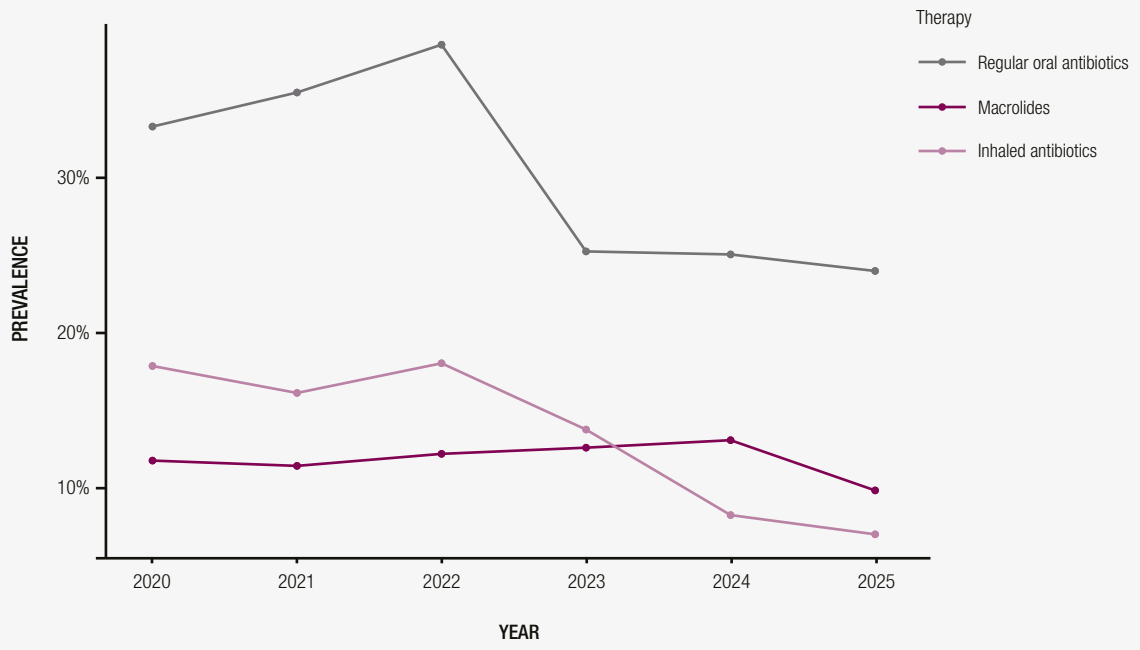
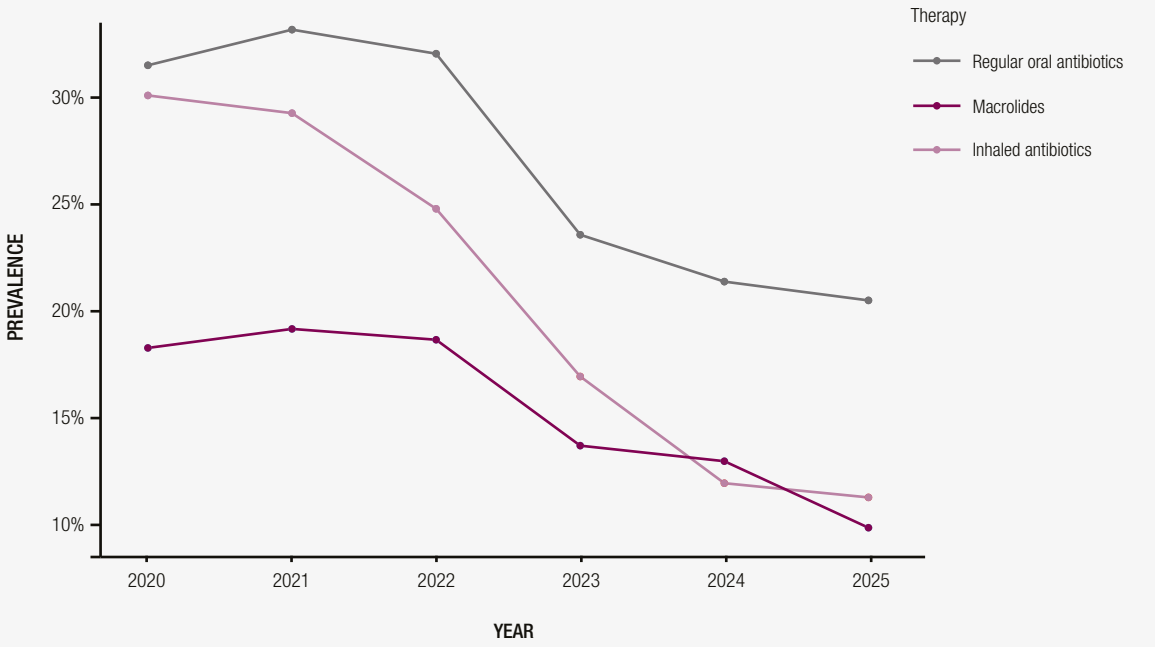


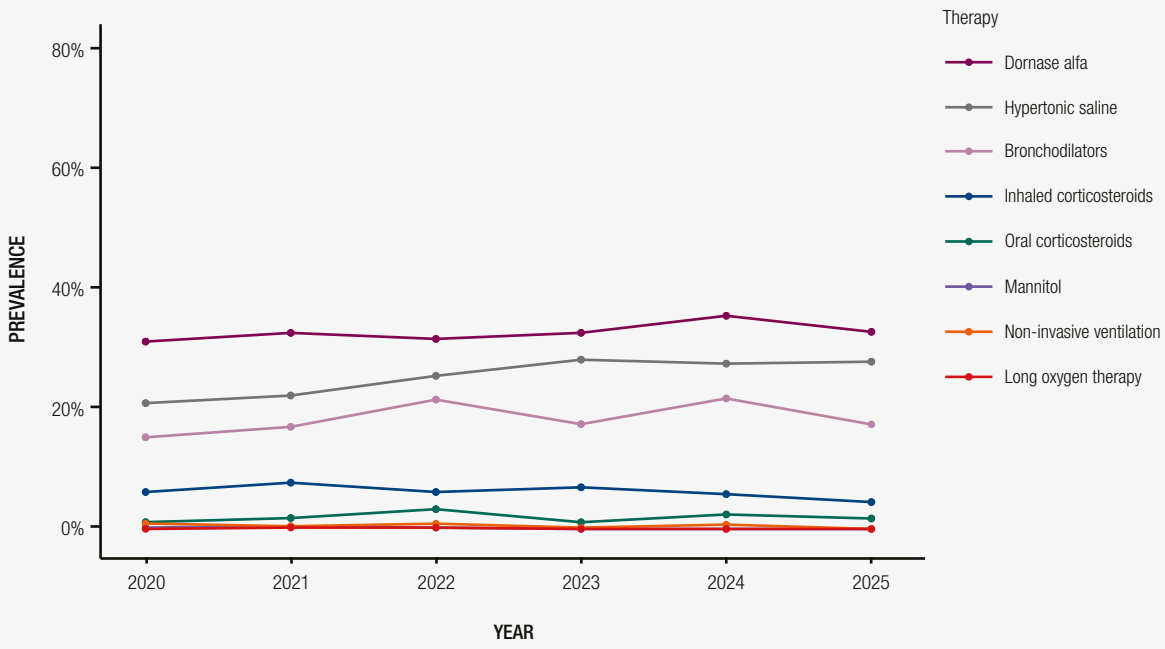
**FIGURE 2.24C:** ACFDR PAEDIATRICS 2020-2025: MAINTENANCE ANTIBIOTIC THERAPY FOR CHILDREN AGED 12-17 YEARS




## CF Lung Therapies — Non-Antibiotic Management

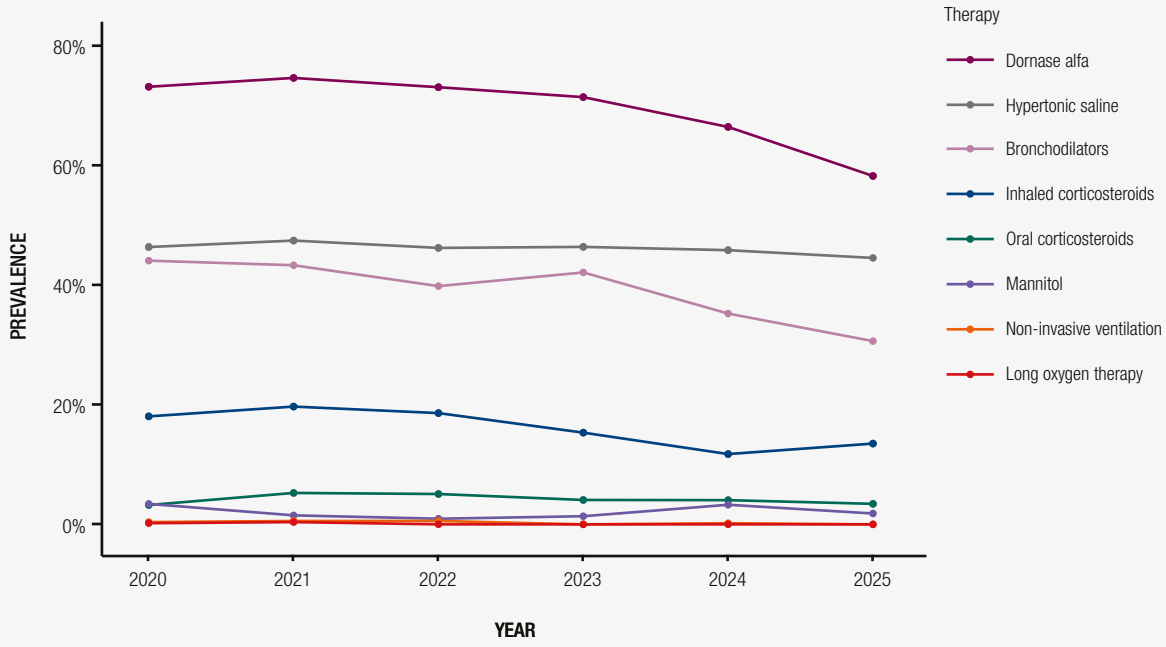
In 2025, the most used adjuvant lung therapies among children and adolescents, regardless of age, were dornase alfa, followed by hypertonic saline and bronchodilators. Use of all adjuvant pulmonary treatments were higher in older children and adolescents. **For children <6 years of age**, use of dornase alfa and hypertonic saline slightly increased over the last six years, with bronchodilator use remaining relatively stable. In 2025, the proportions of children <6 years using these were 32.8%, 27.8% and 17.4% respectively. Usage of inhaled and oral corticosteroids, non-invasive ventilation, long-term oxygen therapy and mannitol remained stable, with less than 5% of pwCF younger than 6 years using these adjuvant lung therapies in 2025 (Figure 2.25A).

**FIGURE 2.25A:** ACFDR PAEDIATRICS 2020-2025: NON-ANTIBIOTIC LUNG THERAPIES FOR CHILDREN UNDER 6 YEARS



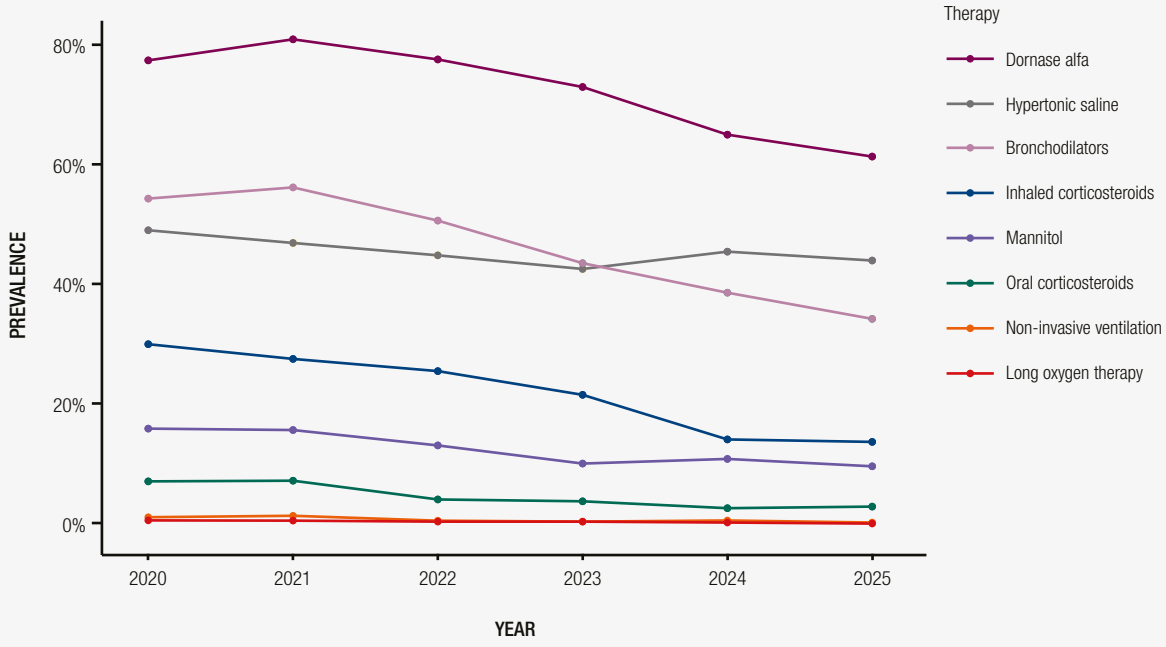
**For older children (6-11 years)**, use of dornase alfa and bronchodilators has decreased over the 6-year period, with 58.4% using dornase alfa and 30.7% using bronchodilators in 2025. Inhaled corticosteroid use has decreased until 2024, however, increased to 13.5% in 2025, while other therapies remained fairly stable during this period (Figure 2.25B).

**FIGURE 2.25B:** ACFDR PAEDIATRICS 2020-2025: NON-ANTIBIOTIC LUNG THERAPIES FOR CHILDREN AGED 6-11 YEARS




**For adolescents (12-17 years)**, use of most adjuvant treatments has declined over the last 5 years. Usage of adjuvant treatments in 2025 for this age group were 61.3% for dornase alfa, 44.0% for hypertonic saline, 34.2% for bronchodilators, and 13.7% for inhaled corticosteroids. The remainder were used by less than 10% of this age group (Figure 2.25C).

**FIGURE 2.25C:** ACFDR PAEDIATRICS 2020-2025: NON-ANTIBIOTIC LUNG THERAPIES FOR CHILDREN AGED 12-17 YEARS



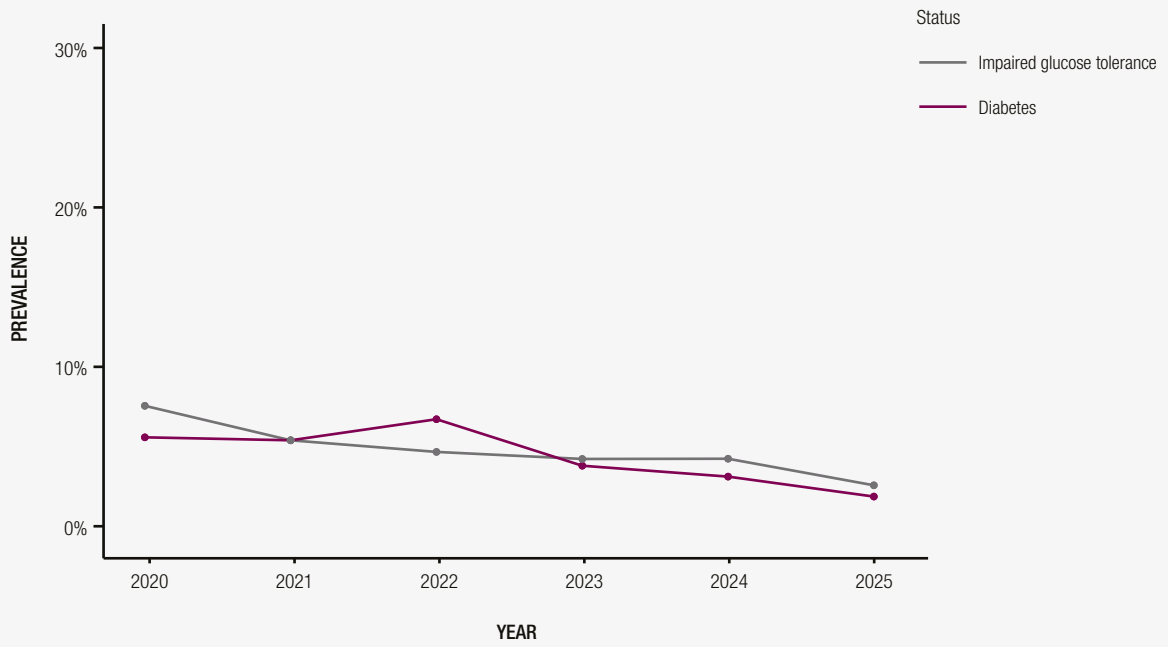
## CF-Related Diabetes

CF-related diabetes is a common complication in adults with CF, and may manifest in older children. Screening for diabetes and impaired glucose tolerance is recommended for pwCF aged 10 years and older. Figures 2.26A and B show the incidence of impaired glucose tolerance and diabetes over the last six years.

The incidence of impaired glucose tolerance reported in 2025 was very low for young children <6 years, with 0.6% reporting impaired glucose tolerance and 0.0% reporting diabetes. This has remained stable over time.

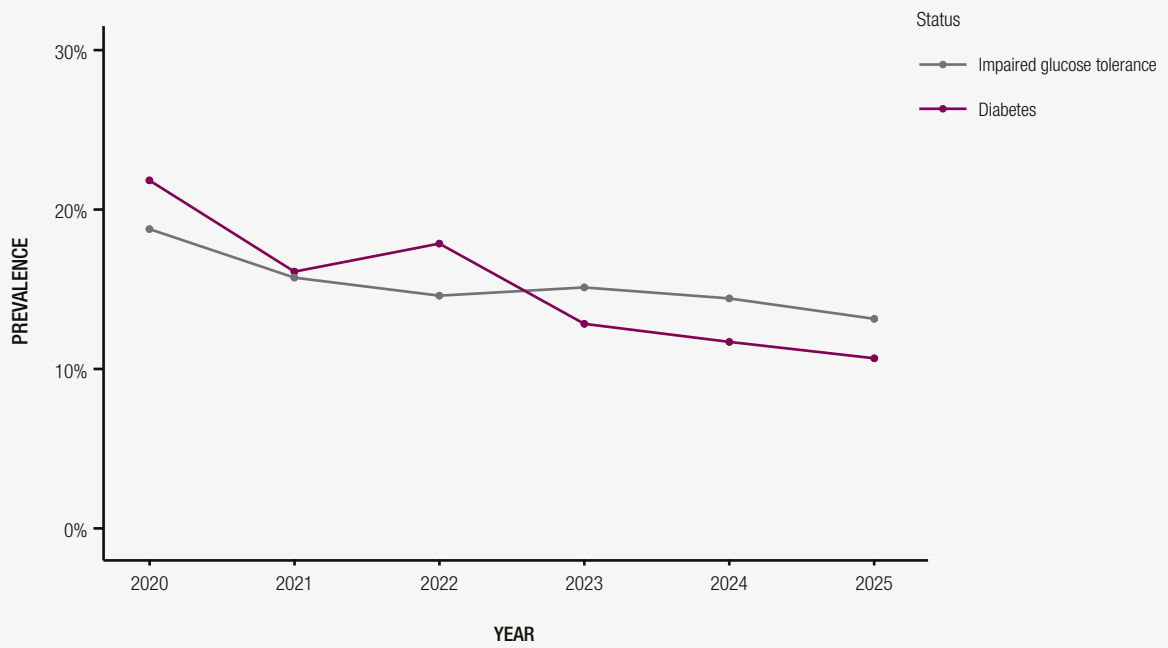
**For children aged 6-11 years**, the incidence of impaired glucose intolerance and diabetes has **reduced over time**. In 2025, the proportion affected were 2.6% and 1.9% respectively.

**FIGURE 2.26A:** ACFDR PAEDIATRICS 2020-2025: DIABETIC STATUS FOR CHILDREN AGED 6-11 YEARS




**For adolescents aged 12-17 years**, there has been a decline in the prevalence of these conditions over time. Impaired glucose tolerance has decreased from 18.8% in 2020 to 13.1% in 2025, and diabetes has reduced from 21.8% to 10.7% respectively.

**FIGURE 2.26B:** ACFDR PAEDIATRICS 2020-2025: DIABETIC STATUS FOR CHILDREN AGED 12-17 YEARS



Over 85% of children with diabetes were treated with insulin. Most insulin use by children and adolescents was long term (chronic) use (Table 2.9).

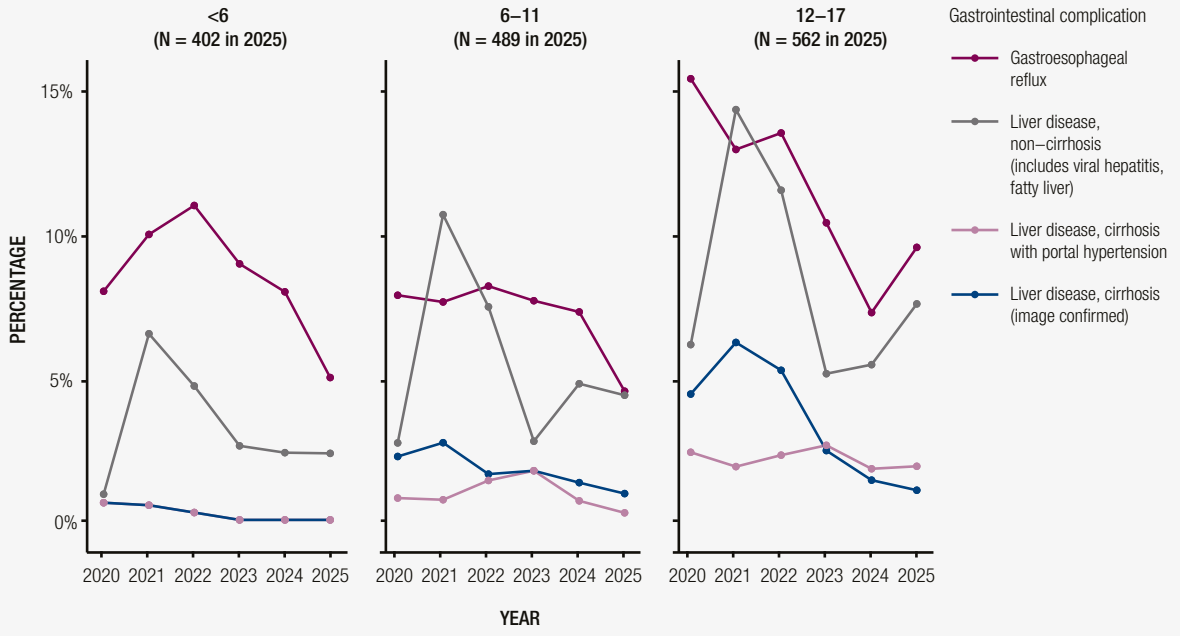
**TABLE 2.9:** ACFDR PAEDIATRICS 2025: DIABETIC TREATMENT BY AGE GROUP

| Diabetes treatment type | <12 (N = 8) | 12-17 (N = 56) |
|---|---|---|
| Insulin | 8 (100.0%) | 47 (83.9%) |
| Hypoglycaemics | 0 (0.0%) | <5 |
| Insulin and hypoglycaemics | 0 (0.0%) | <5 |
| Diet/lifestyle management only | 0 (0.0%) | 5 (8.9%) |
| No treatment for diabetes | 0 (0.0%) | <5 |
| **Insulin use** | **<12 (N = 8)** | **12-17 (N = 53)** |
| Chronic insulin use | 8 (100.0%) | 41 (77.4%) |
| Intermittent insulin use | 0 (0.0%) | <5 |
| Insulin use, duration unknown | 0 (0.0%) | 8 (15.1%) |
| No insulin use | 0 (0.0%) | 0 (0.0%) |

## CF Gastrointestinal Disease

Gastrointestinal complications of CF are not common in children and adolescents, and rates have been decreasing over the last 6 years. In 2025, the most common gastrointestinal complication among children was gastroesophageal reflux (GOR), at 5.0% of children aged 6 or younger, 4.5% of 6-11 year-olds, and 9.4% of 12-17 year-olds. The incidence of cirrhosis without portal hypertension has reduced significantly for 12-17 year-olds to 1.1% in 2025, while cirrhosis with portal hypertension was less than 2%. Non-cirrhotic liver disease affected approximately 7.5% of this age group in 2025 (Figure 2.27).

**FIGURE 2.27:** ACFDR PAEDIATRICS 2020-2025: STOMACH AND LIVER DISEASE



In 2025, pancreatitis was very uncommon among children, with <1% of children and adolescents with CF reporting pancreatitis.

In 2025, pancreatic insufficiency affected 75.7% of children <6 years, 78.9% of children 6-11 years, and 77.7% of adolescents 12-17 years (Table 2.10).

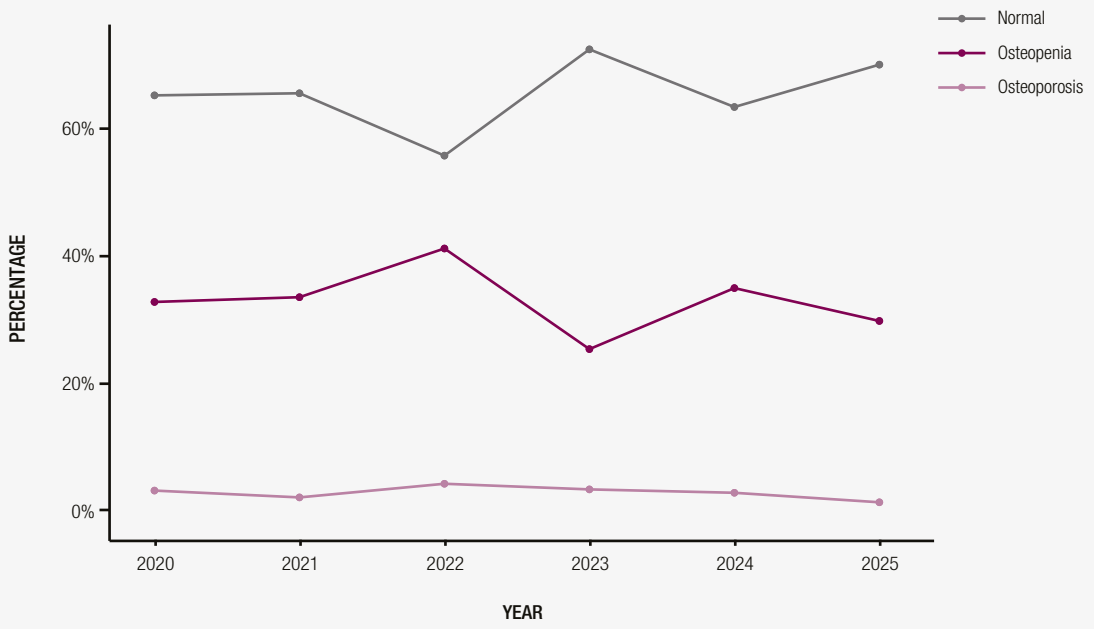
**TABLE 2.10:** ACFDR PAEDIATRICS 2025: PANCREATIC STATUS BY AGE

| Pancreatic Status | <6 (N = 395) | 6-11 (N = 483) | 12-17 (N = 557) |
|---|---|---|---|
| Insufficient | 299 (75.7%) | 381 (78.9%) | 433 (77.7%) |

## Osteopenia and Bone Density

Bone mineral density scans are not routinely performed on children younger than 10 years unless clinically indicated. In 2025, 102 adolescents older than 10 years had their bone density status reported to the ACFDR. Of these, 29.4% had osteopenia. This proportion has decreased slightly over time from 32.4% in 2020 to 29.4% in 2025. Osteoporosis was very uncommon for children with CF aged 10-17 in 2025 (Figure 2.28). Fracture status was recorded for 722 young pwCF in 2025, with fractures reported for 28 (3.9%) 10-17 year-olds.

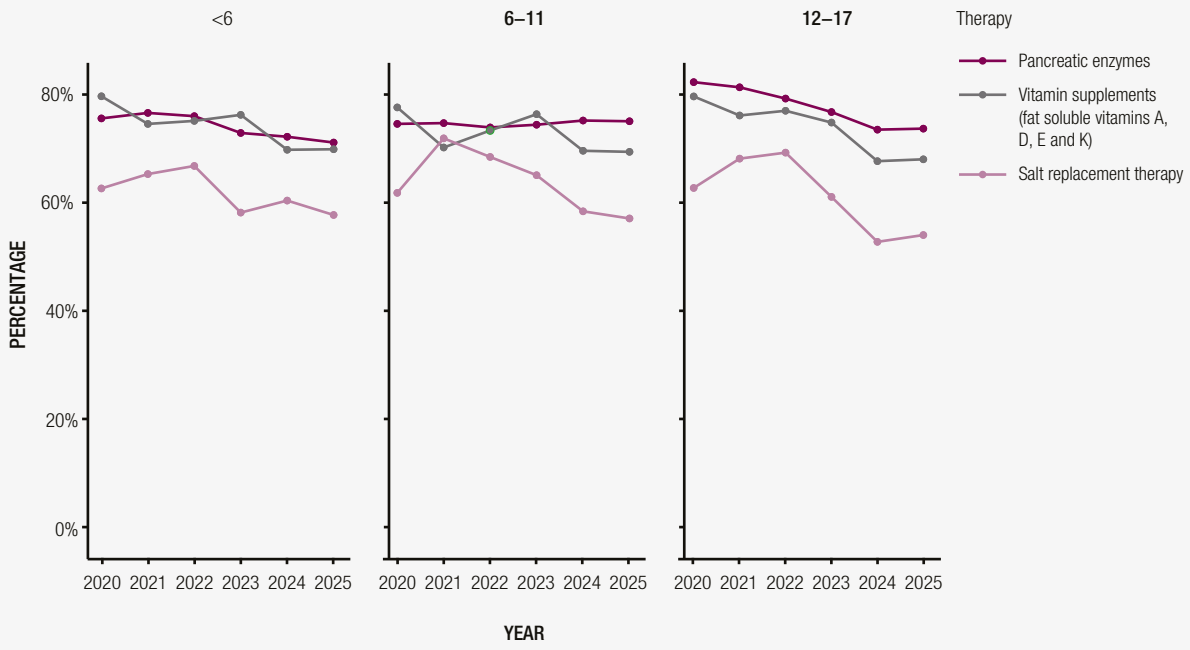
**FIGURE 2.28:** ACFDR PAEDIATRICS 2020-2025: BONE DENSITY FOR ADOLESCENTS



## Nutritional Supplementation

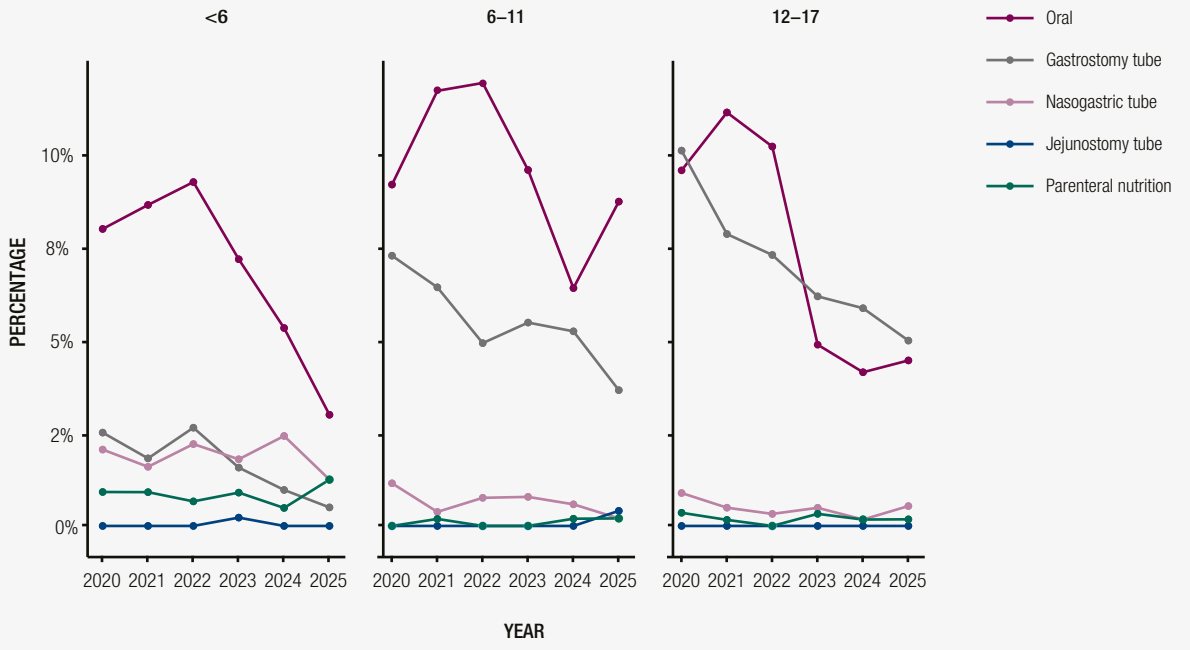
Figure 2.29 reveals the usage of pancreatic enzymes and nutritional supplements across age groups from 2020-2025. In 2025, for those under 6, 71.2% used pancreatic enzymes, 70.0% used fat-soluble vitamin supplements, and 57.8% used salt replacement therapy. Among 6-11 year-olds, 75.2% used pancreatic enzymes, 69.5% took vitamin supplements, and 57.2% used salt replacement therapy. Among 12-17 year-olds, 74.1% used pancreatic enzymes, 68.4% took vitamin supplements, and 54.4% used salt replacement therapy. Since 2020, use of nutritional supplementation, particularly salt replacement, has decreased over time, with pancreatic enzyme use being more stable other than for the 12-17 year age group.

**FIGURE 2.29:** ACFDR PAEDIATRICS 2020-2025: NUTRITIONAL SUPPLEMENTS BY AGE




Since 2020, the use of nutritional support among children and adolescents has decreased (Figure 2.30). In 2025, three percent of children aged less than 6 years, 8.7% of children aged 6-11 and 4.3% of children aged 12 or older respectively required oral supplements. In 2025, 3.6% of children aged 6-11 and 5.0% of adolescents received nutritional support via a gastrostomy tube. Other forms of nutritional support were uncommon.

**FIGURE 2.30:** ACFDR PAEDIATRICS 2020-2025: NUTRITIONAL SUPPORT BY AGE

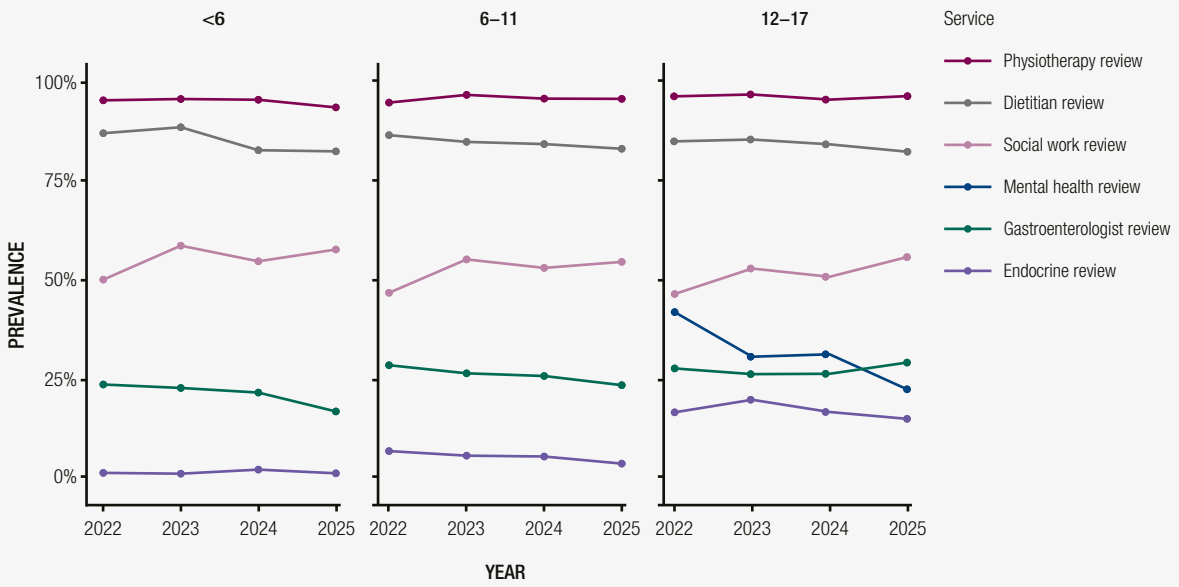


## Multidisciplinary Care

Multidisciplinary care is a cornerstone of effective, contemporary CF management. In 2025, a majority of children and adolescents participated in annual physiotherapy and dietitian reviews. Approximately half of children with CF had a social work review in 2025, approximately one quarter had a gastroenterology review or a mental health screen or review (for children 12 years or older). Proportions who had reviews are generally similar for younger children and adolescents except for endocrine reviews, which were performed for 14.6% of adolescents but only 1-3.4% of younger children (Figure 2.31).

From 2022 to 2025, there was minimal variation in the proportion of young pwCF accessing allied health therapies with the exception of mental health reviews for adolescents aged 12-17 which decreased from 41.3% in 2022 to 22.0% in 2025.

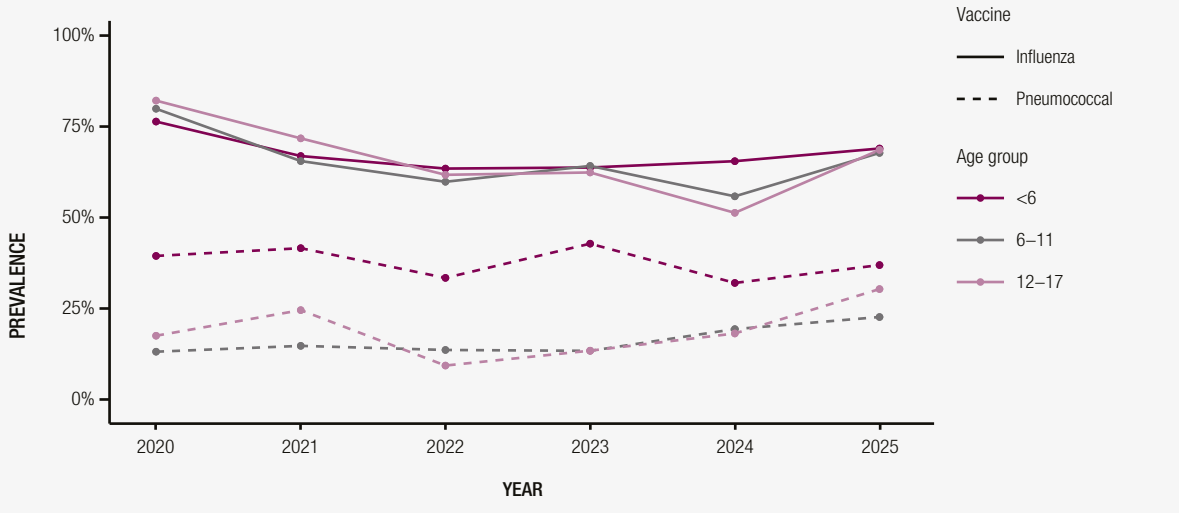
**FIGURE 2.31:** ACFDR PAEDIATRICS 2022-2025: ANNUAL MULTIDISCIPLINARY CARE APPOINTMENTS BY AGE GROUP




## Vaccinations

Influenza immunisation is recommended for individuals with CF aged six months and older on an annual basis. In 2025, 73.8% and 59.8% of children and adolescents with CF recorded vaccination status for influenza and pneumococcal respectively. Of these, 69.6% of children aged <6, 68.4% of children aged 6-11 years, and 69.2% of adolescents aged 12-17 years were recorded as being immunised against influenza, compared to 77.0% of children <6, 80.6% of children aged 6-11 years, and 82.8% of adolescents aged 12-17 in 2020 (Figure 2.32).

**FIGURE 2.32:** ACFDR PAEDIATRICS 2020-2025: VACCINATIONS BY AGE GROUP



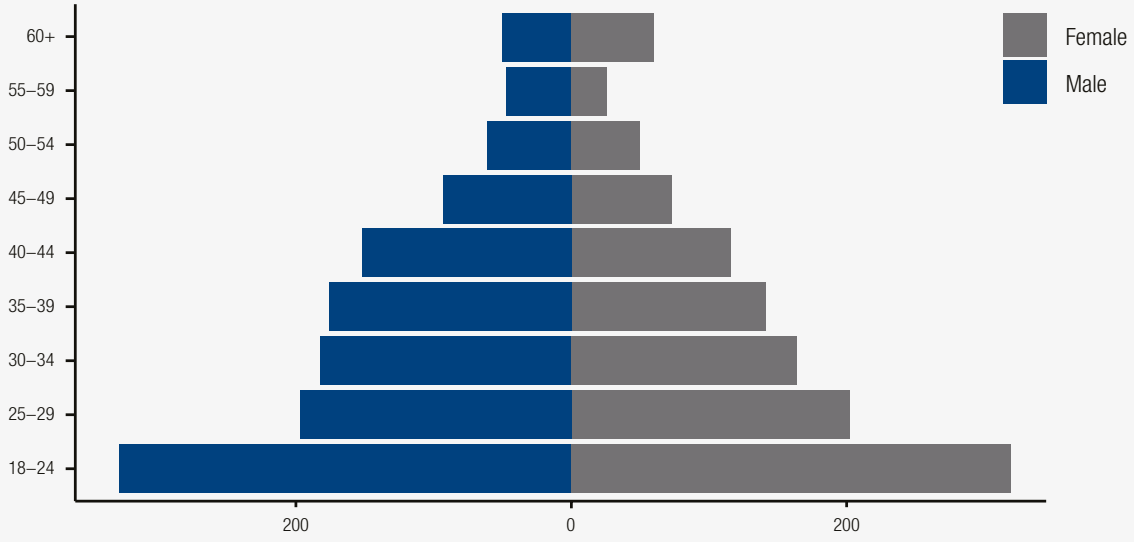
# 3.
# ADULT DATA

# 3. ADULT DATA

## 3.1 ADULTS WITH CF

As of December 31st 2025, the ACFDR held data regarding 2,437 adults with CF, of which 1,151 (47%) were female and 1,286 (53%) were male. The most common adult age group was 18-24 years for both females (17%) and males (16%), followed by 25-29 years for females (11%) and males (10%). Seventeen percent of females and 19% of males were aged 40 and older. This chapter discusses management and clinical outcomes of adult patients in the registry (Figure 3.1).

**FIGURE 3.1:** ACFDR ADULTS 2025: AGE AND SEX




N = 2,437 people

### Socioeconomic Characteristics

#### Educational Attainment

Figure 3.2 shows the highest educational attainment for adults with CF over the last six years. Of the 1,738 (71.3%) adults with CF for whom education information was available in the ACFDR in 2025, the most common educational attainment was senior secondary school (39%), followed by a University/Bachelor degree (25%), and a Tertiary certificate/diploma (22%). In 2025, 9% of adults nominated junior secondary school as their highest educational attainment, a decrease from 13% in 2020 (Figure 3.2).

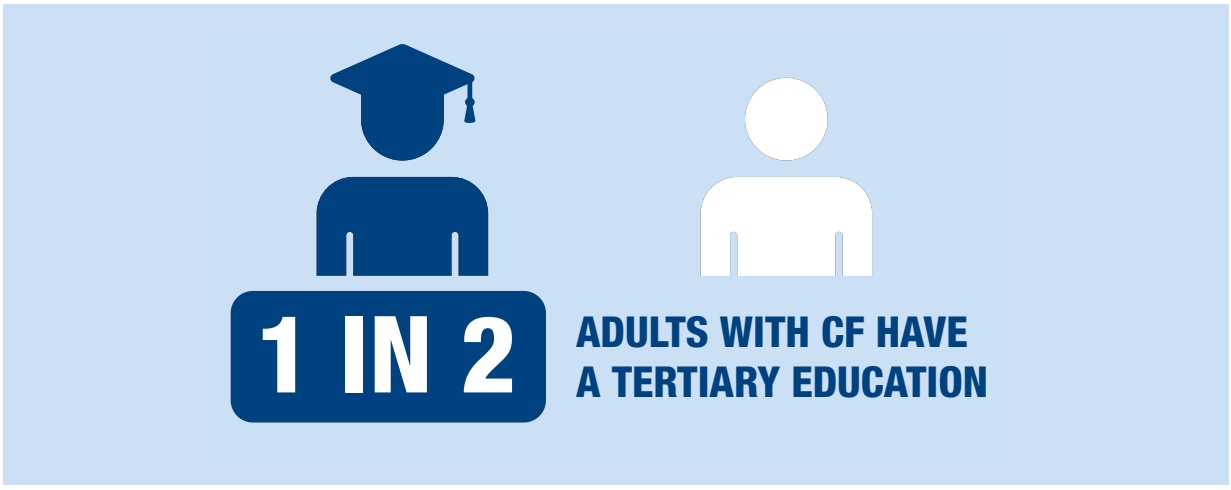


**FIGURE 3.2:** ACFDR ADULTS 2020-2025: HIGHEST EDUCATIONAL ATTAINMENT

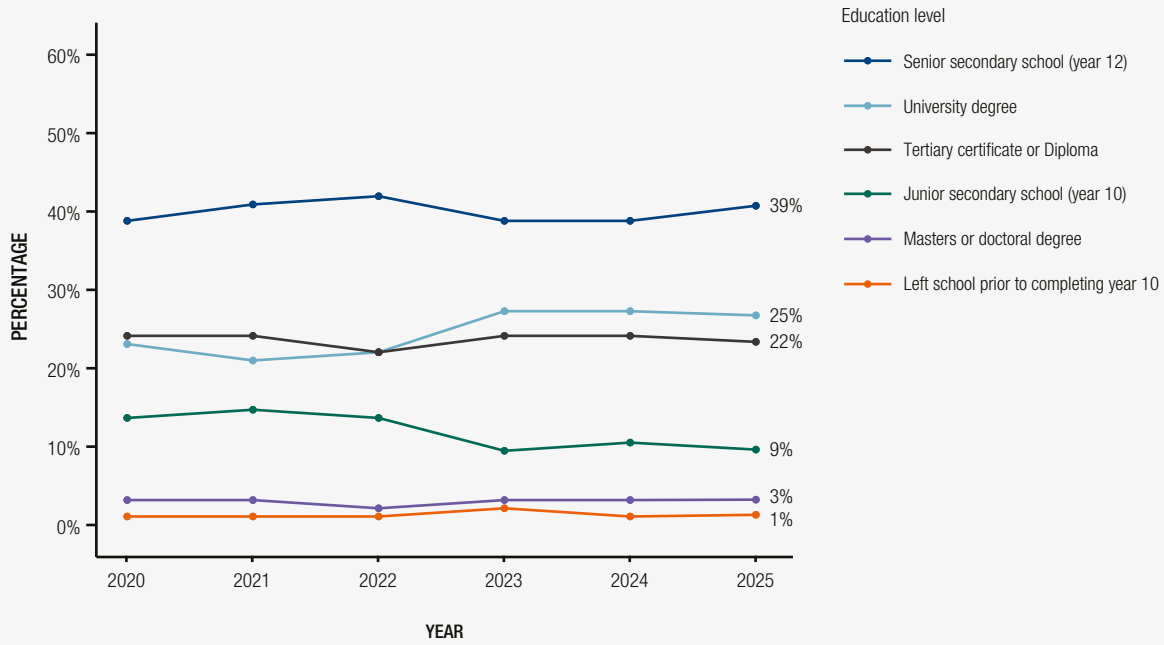



Table 3.1 compares the level of educational attainment of pwCF to the broader Australian population (Australian Bureau of Statistics (ABS) data as of May 2025). Educational attainment for pwCF has been increasing in recent years, and in 2025, **25% of pwCF had completed a university bachelor** degree, which is higher than in the **general population** (21%). Similarly, **fewer pwCF are leaving school prior to completing year 12** than in the general population (10% of pwCF compared to 19% of Australians) (Table 3.1).

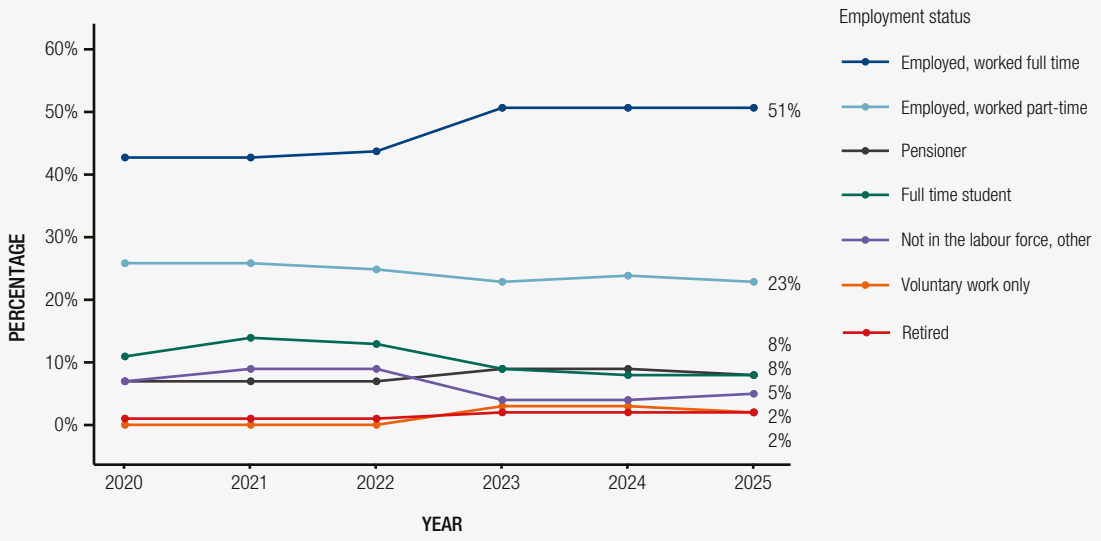
**TABLE 3.1:** ACFDR ADULTS 2025: HIGHEST EDUCATIONAL ATTAINMENT

| Highest educational attainment | ACFDR (N = 1,738) | ABS Data (%), May 2025 for 15-74 year- olds |
|---|---|---|
| Masters or doctoral degree | 53 (3%) | 8.9% |
| University/Bachelor degree | 443 (25%) | 21.3% |
| Tertiary certificate or Diploma | 387 (22%) | 28.6% |
| Senior secondary school (year 12) | 675 (39%) | 18.6% |
| Junior secondary school (year 10) | 159 (9%) | 13.8% |
| Left school prior to completing year 10 | 21 (1%) | 5.3% |


Figure 3.3 shows the employment status of adults with CF over the last six years. Within the ACFDR dataset, information on employment status was available for 2,262 adults with CF (92.8%). Among them, 51% were engaged in full-time employment in 2025, an increase from 43% in 2020.

Most other employment statuses remained stable or declined slightly over the last 6 years. In 2025, 23% were in part-time employment, 8% were pensioners, and 8% were enrolled in full-time study. The proportion of adults with CF not in the labour force has decreased from 7% in 2020 to 5% in 2025. Adults engaged in voluntary work only, or who were retired accounted for 4% of all adults with CF (Figure 3.3).

**FIGURE 3.3:** ACFDR ADULTS 2020-2025: EMPLOYMENT STATUS



## Marital Status

Information regarding marital status was available for 2,262 adults in the registry (92.8%). Of these, 52% of women and 50% of men were either married or in a de facto relationship.

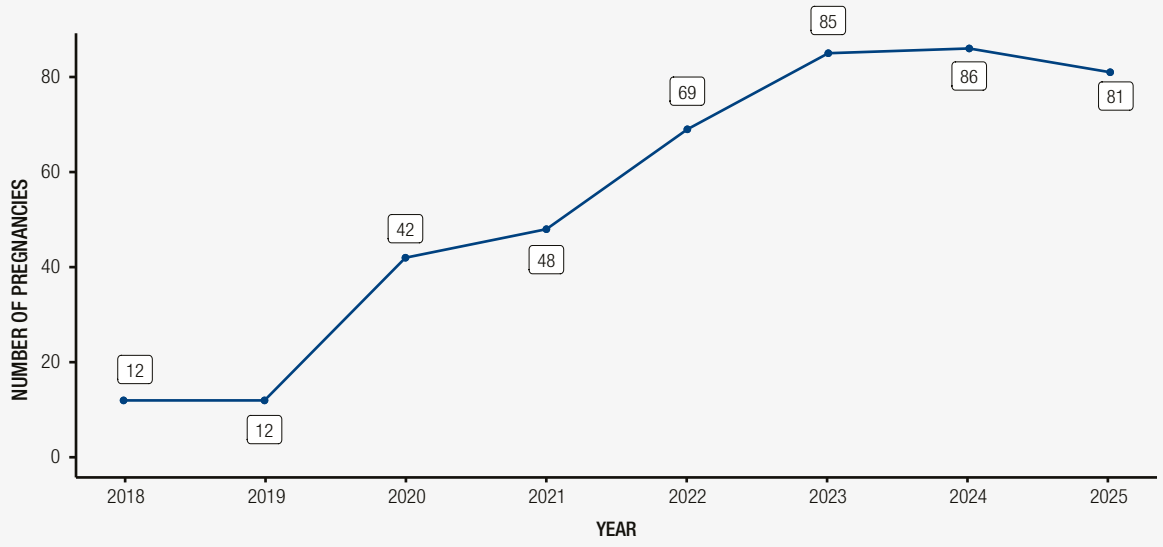

## Pregnancies for People with CF

**From this point in the clinical data, adults with lung transplants (n= 97) have been excluded from the analysis.**

In 2025, there were 81 pregnancies to women with CF, similar to the number of pregnancies recorded in 2023 and 2024 (Figure 3.4).

Historical results have been recalculated using year-specific active populations and consolidated to reflect a single pregnancy outcome per person. For pwCF recording conflicting pregnancy outcomes across multiple CF centres, event-based outcomes (e.g., live births) have been retained over an ongoing status (i.e., currently pregnant).

**FIGURE 3.4:** ACFDR ADULTS 2018-2025: PREGNANCY



Of the 81 pregnancies recorded in 2025, by December 31st 2025 there were 43 (53.1%) live births and 25 (30.9%) people still pregnant (Table 3.2).

**TABLE 3.2:** ACFDR ADULTS 2025: PREGNANCY STATUS

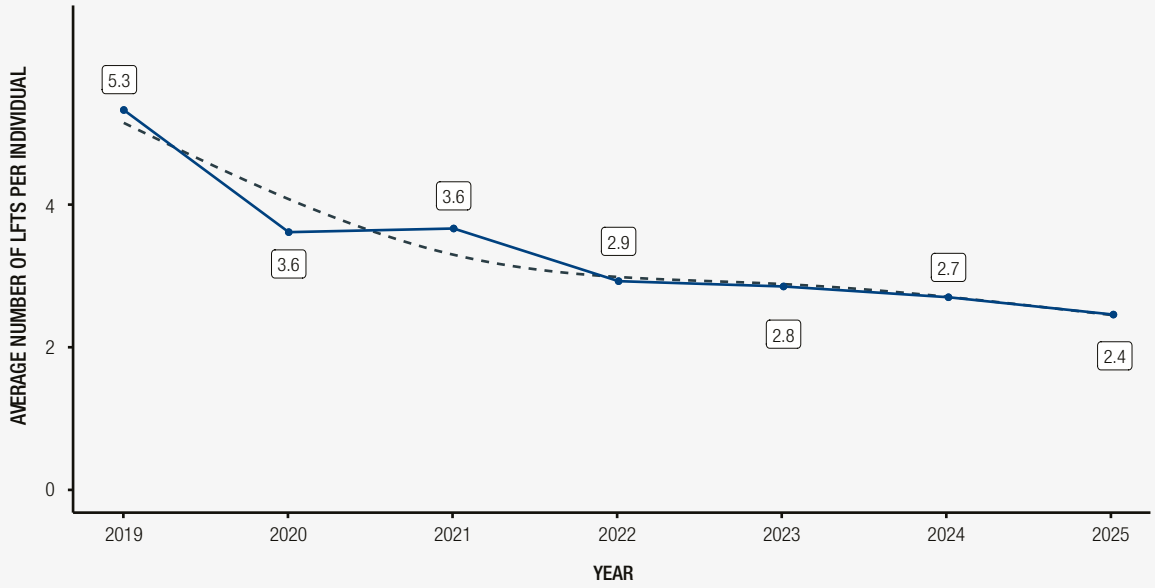
| Status | ≥18 (N = 81) |
|---|---|
| Currently pregnant | 25 (30.9%) |
| Live birth | 43 (53.1%) |
| Other (Miscarriage, stillbirth or termination) | 13 (16.0%) |

## 3.2 CLINICAL MEASURES

### Lung Function

Figure 3.5 depicts the annual average number of lung function (spirometry) tests per adult recorded in the registry from 2019 to 2025. This has halved in recent years, with adults averaging 2.4 tests in 2025 compared to 5.3 tests in 2019.

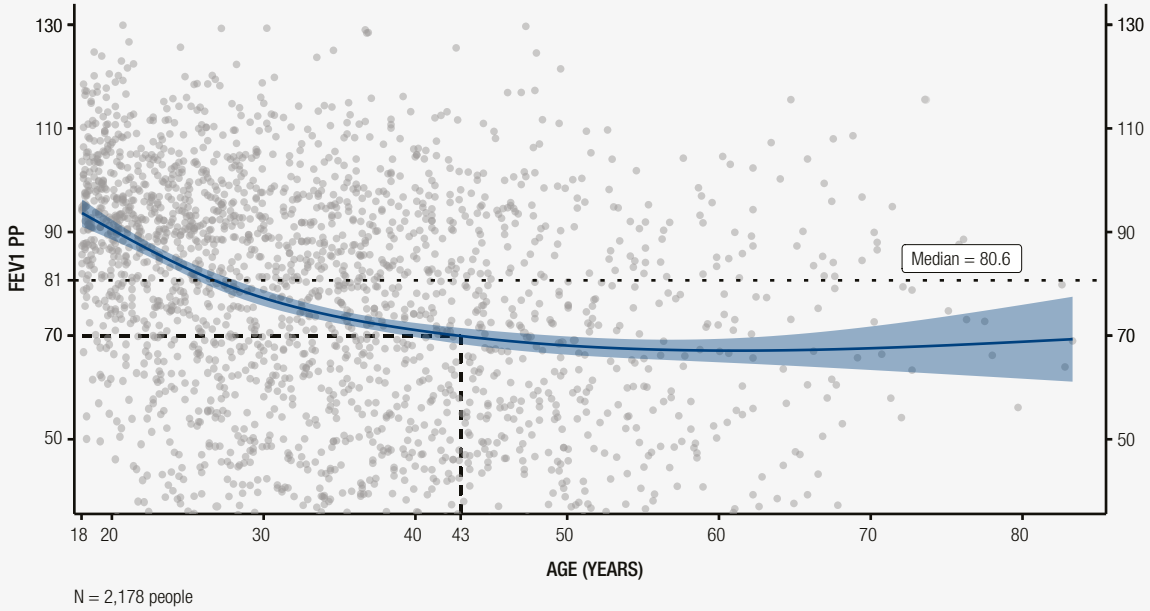
**FIGURE 3.5:** ACFDR ADULTS 2019-2025: AVERAGE NUMBER OF LUNG FUNCTION TESTS ENTERED PER INDIVIDUAL




Dashed curve represents the smoothed trend (natural cubic spline, df = 3)

There were 2,178 (89.4%) adults in 2025 in the registry who had their lung function measures recorded. The adult lung function results reveal a progressive decline in median FEV1 pp with increasing age (Figure 3.6). The median age at which an FEV1 pp of 70.0 is recorded is 43 years in 2025, an increase from 36 years in 2022.

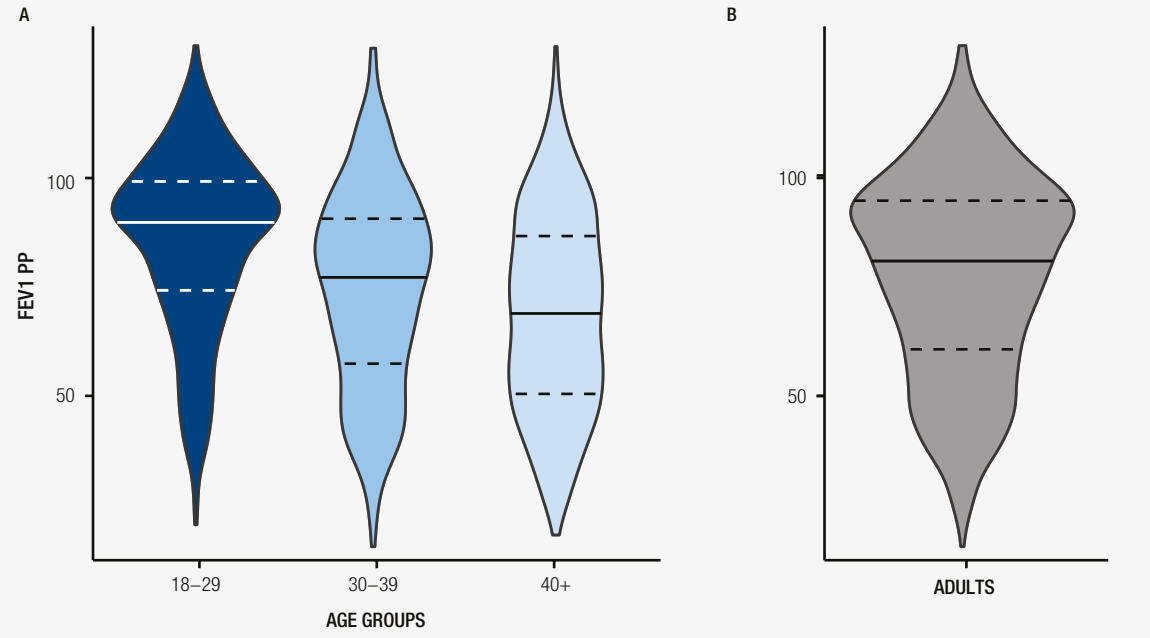
**FIGURE 3.6:** ACFDR ADULTS 2025: FEV1 PP BY AGE




The solid trend line was estimated using a natural cubic spline with 3 degrees of freedom.
The dotted horizontal line shows the median FEV1pp.
Shaded area represents the 95% confidence interval.

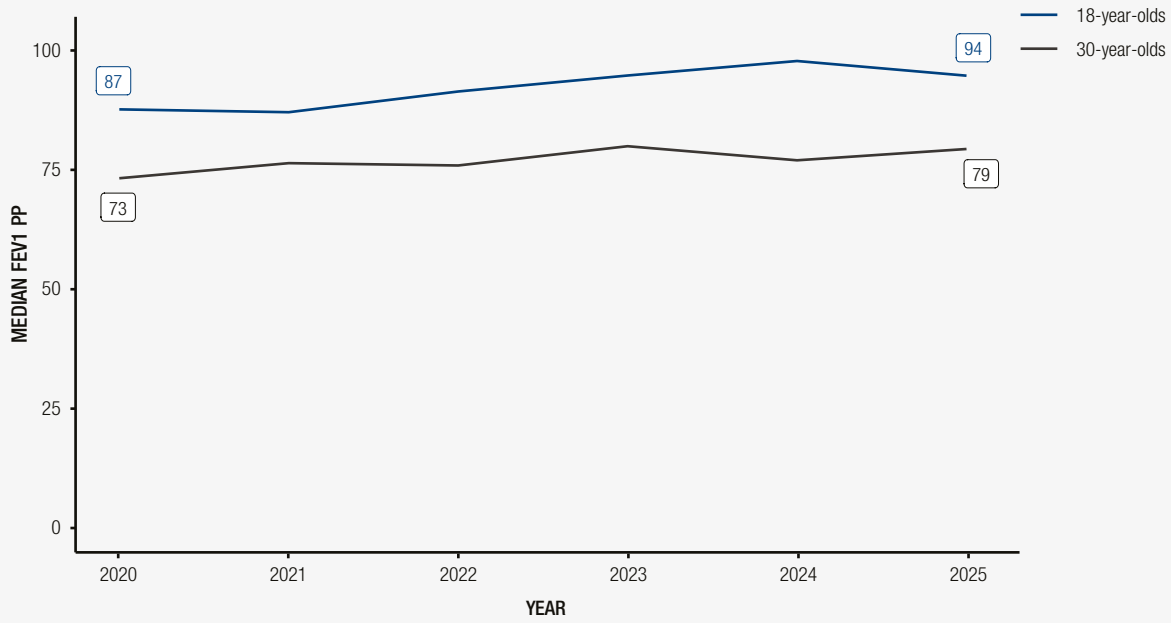
In 2025, the median FEV1 pp for adults overall was 80.6, with the median FEV1 pp for 18-29 year-olds being 89.4, for 30-39 year-olds being 76.9, and for 40+ year-olds being 68.7 (Figure 3.7).

**FIGURE 3.7:** ACFDR ADULTS 2025: MEDIAN FEV1 PP



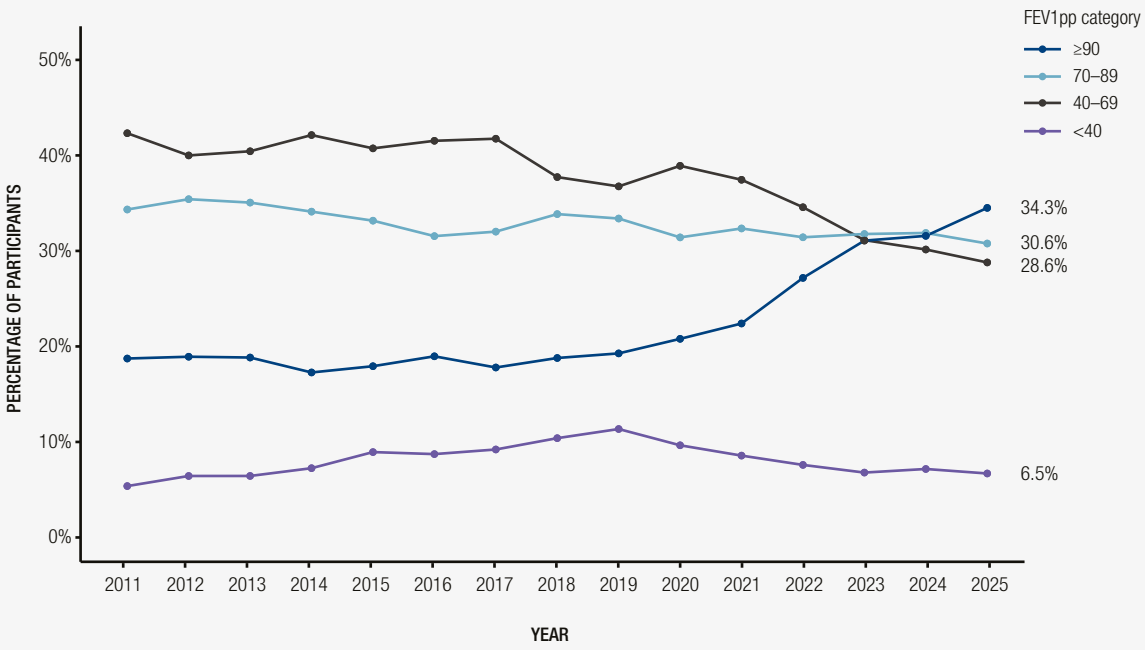
The median FEV1 pp has increased over time. For 18-year-olds, it has increased from 87.3 in 2020 to 94.4 in 2025, a 7.1 increase. For pwCF of age 30 years, the median FEV1 pp in 2025 was 79.0, an increase of 6.1 from 72.9 in 2020 (Figure 3.8).

**FIGURE 3.8:** ACFDR ADULTS 2020-2025: MEDIAN FEV1 PP OVER TIME




FEV1 pp has increased since 2011 (Figure 3.9). The proportion of adults with an FEV1 pp of ≥90 has increased from 18.7% in 2011 to 34.3% in 2025, and is now the largest group. The proportion of adults with FEV1 pp of 70-89 has decreased from 34.1% in 2011 to 30.6% in 2025, and the proportion of adults with FEV1 pp of 40-69 has decreased from 41.8% to 28.6%. The proportion of pwCF with an FEV1 pp of <40 has remained between 5-10% during the last 5 years.

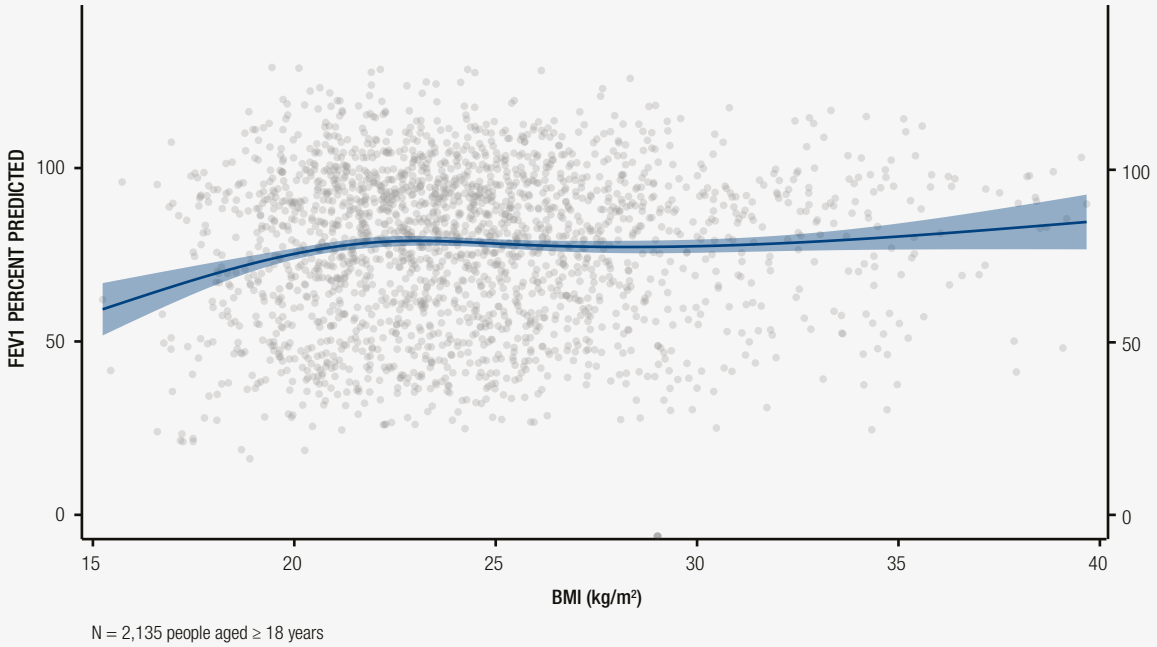
**FIGURE 3.9:** ACFDR ADULTS 2011-2025: FEV1 PP DISTRIBUTION OVER TIME



## Lung Function and Body Mass Index

For pwCF aged 18-40 years, FEV1 pp increased with increasing BMI, although, at BMIs into the high 20s, this appears to variably affect FEV1 pp. PwCF over 40 years are not included due to fewer numbers which makes the data difficult to interpret due to increased variability (Figure 3.10).

**FIGURE 3.10:** ACFDR ADULTS 2025 : FEV1 PP VS BMI

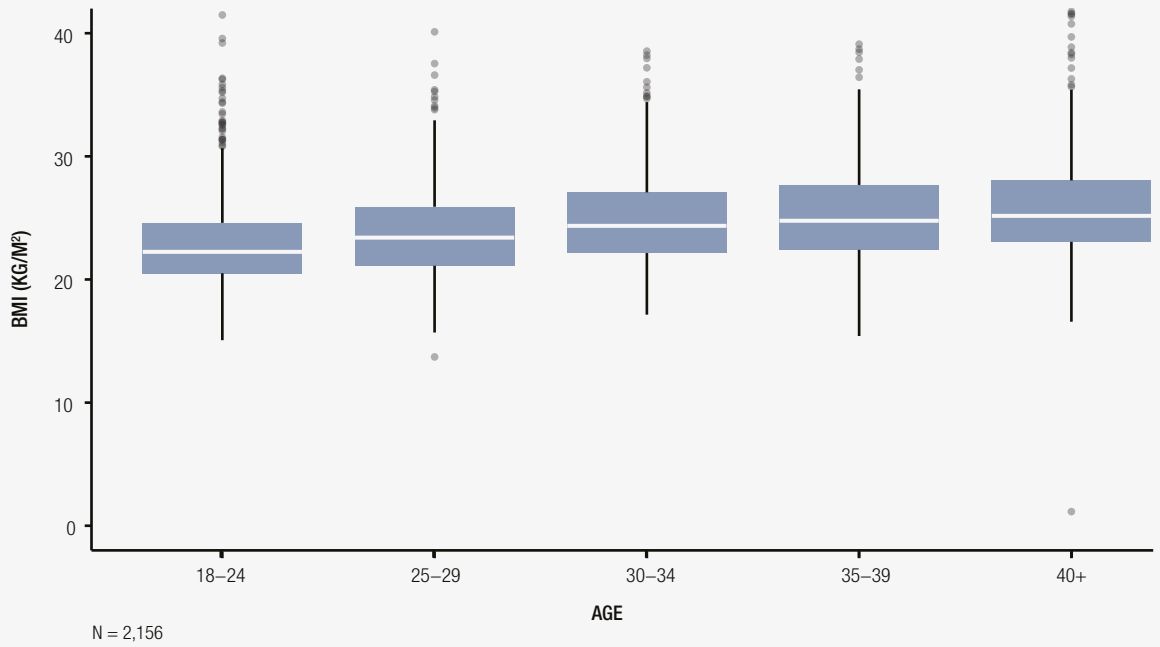



Solid line was calculated using a natural cubic spline with 3 degrees of freedom
Shaded area represents 95% confidence interval

## Nutrition

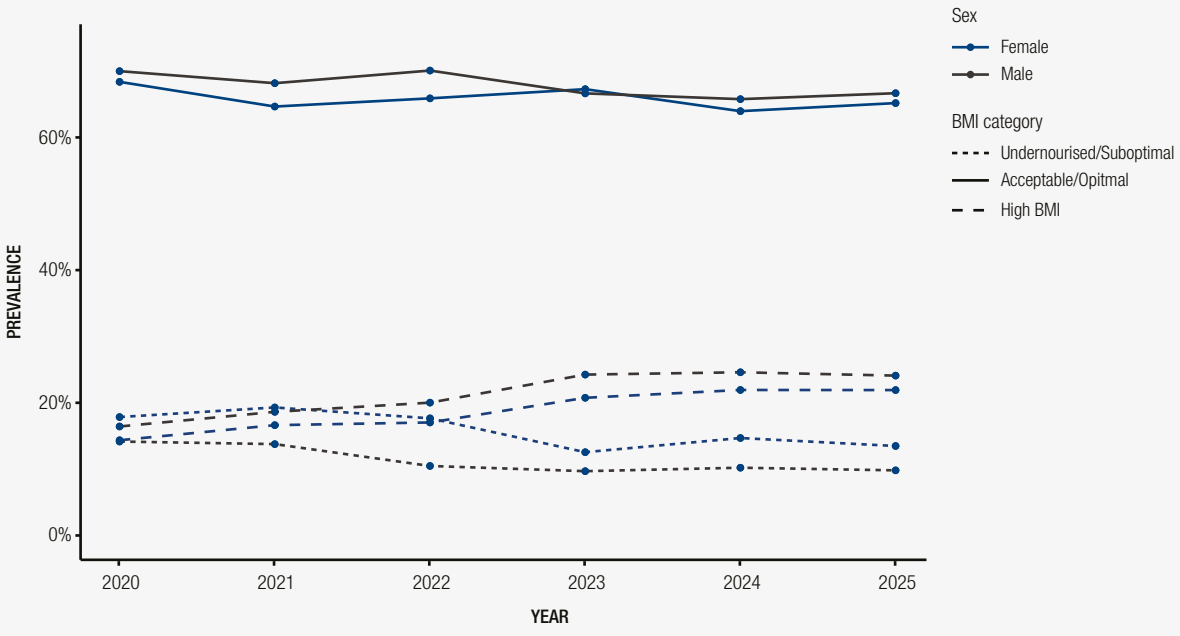
BMI for adults with CF increases with increasing age. For 2025, in the 18-24 age group, the median BMI was 22.3, for the 25-29 age group it was 23.4, for 30-34 age group it was 24.4 and for 35-39 age group it was 24.8. The highest median BMI was observed in the 40 and older age category, with a value of 25.2 (Figure 3.11).

**FIGURE 3.11:** ACFDR ADULTS 2025: BMI BY AGE



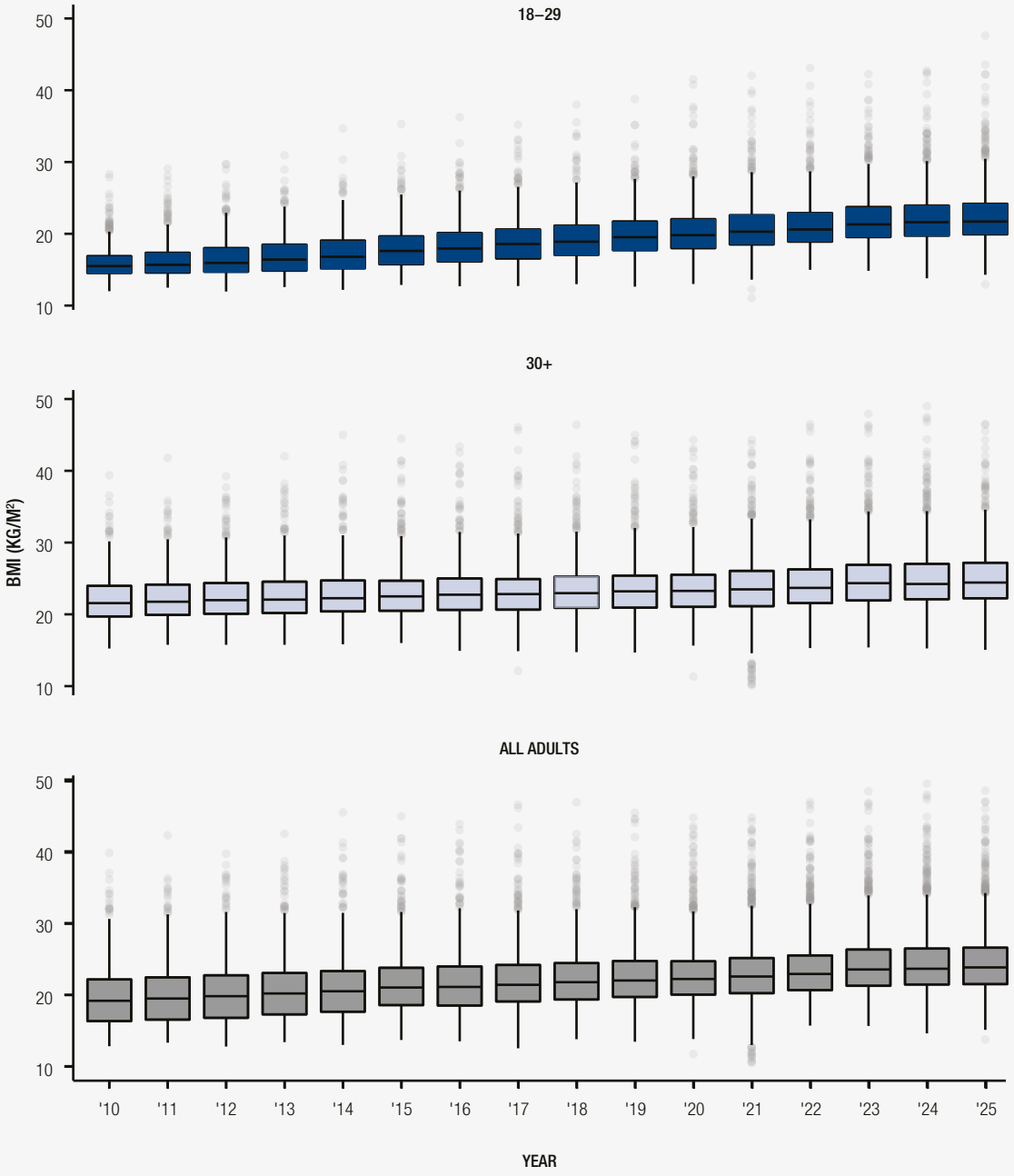

Approximately two-thirds of adults with CF had a BMI in the optimal/acceptable range recorded in 2025 (66.4% of males and 64.9% of females). 23.9% of males and 21.8% of females had a high BMI, while 13.4% of females and 9.7% of males had a suboptimal BMI (Figure 3.12).

**FIGURE 3.12:** ACFDR ADULTS 2020-2025: BMI BY SEX



The BMI data for the adult CF population revealed a consistent upward trajectory over the years. In the 18-29 age group, the median BMI has risen steadily from 16.3 in 2010 to 22.6 in 2025. A similar pattern was observed in the 30-year age group, where the median BMI increased from 22.0 in 2010 to 24.9 in 2025. When considering all adults with CF, the overall median BMI has shown a continuous increase from 19.2 in 2010 to 23.9 in 2025 (Figure 3.13).

**FIGURE 3.13:** ACFDR ADULTS 2010-2025: MEDIAN BMI

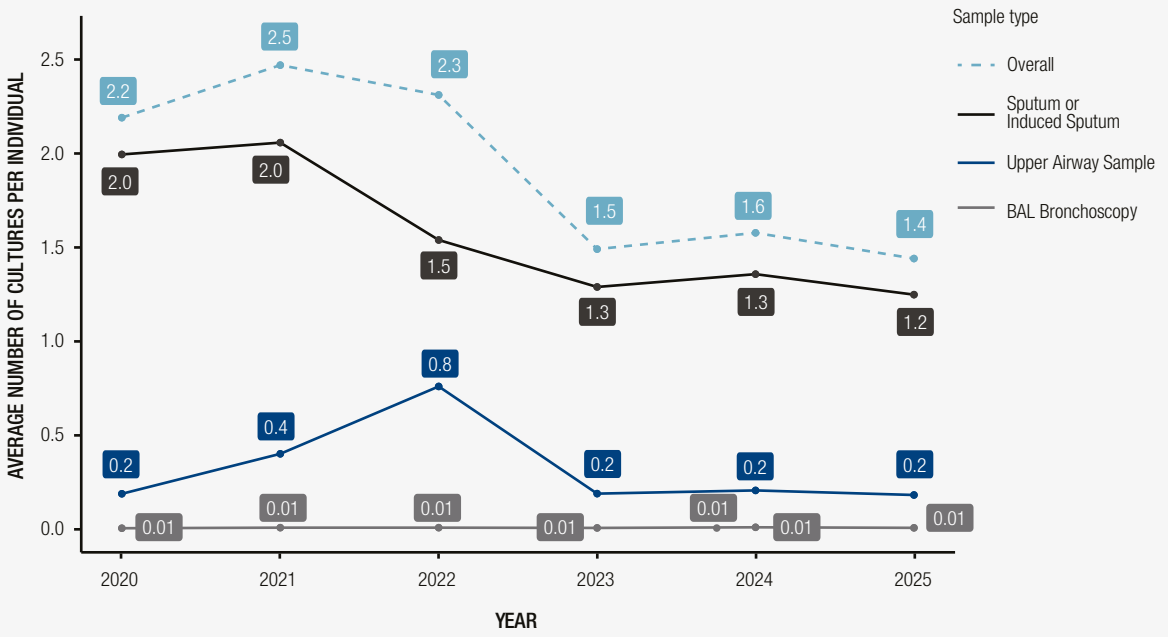


## Microbiology

### Microbiology Samples

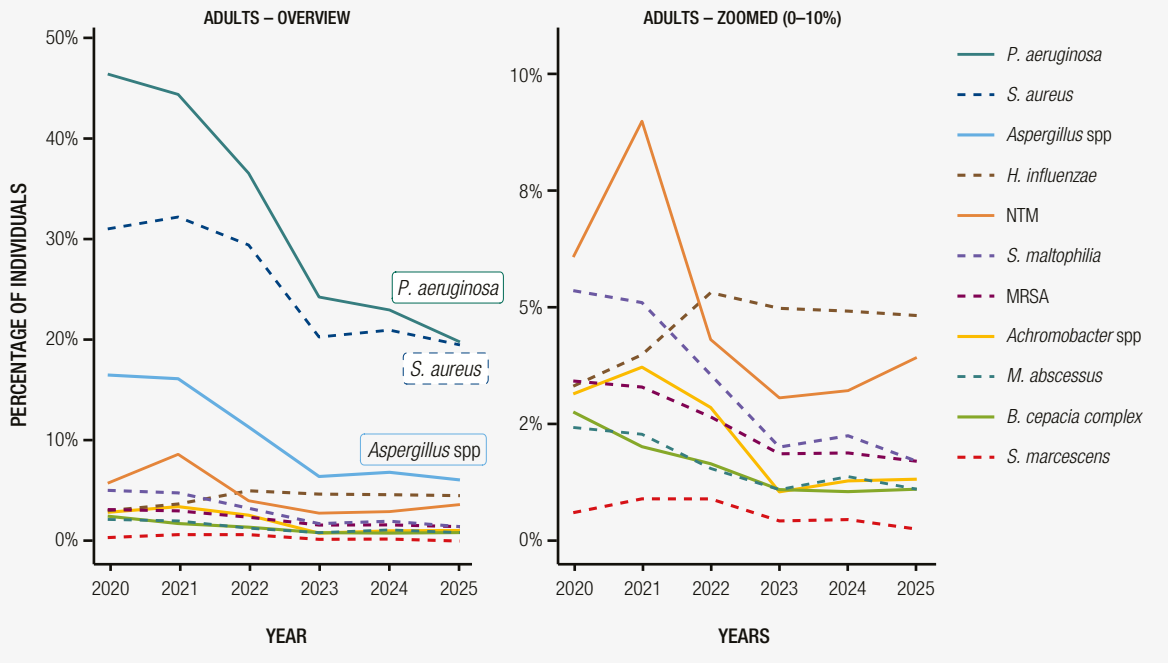
The average number of respiratory samples collected per adult from 2020 to 2025 is depicted in Figure 3.14. From 2020 to 2025 there was a reduction in the overall number of respiratory samples per adult, peaking at 2.5 in 2021 before decreasing to a low of 1.4 samples in 2025. The most common respiratory sample types collected for adults are sputum/induced sputum, which has followed this trend.

**FIGURE 3.14:** ACFDR ADULTS 2020-2025: AVERAGE NUMBER OF RESPIRATORY SAMPLES PER INDIVIDUAL




The prevalence of specific organisms commonly affecting pwCF has generally declined over the last six years (Figure 3.15). The prevalence of *P. aeruginosa* has decreased from 47.3% in 2020 to 20.3% by 2025. *S. aureus* has decreased from a peak of 32.9% in 2021 to 20.0% in 2025. The prevalence of *Aspergillus* spp has declined from 17.0% in 2020 to 6.4% in 2025. The prevalence of the less common microorganisms NTM, MRSA, *S. maltophilia*, *Achromobacter* spp, *B. cepacia complex*, *M. abscessus*, and *S. marcescens* generally decreased, all with prevalence of less than 5% in 2025.

**FIGURE 3.15:** ACFDR ADULTS 2020-2025: PREVALENCE OF RESPIRATORY MICROBIOLOGY



Microbiology results for adults from 1,246 samples collected in 2025 show the prevalence of specific microorganisms for different age groups of pwCF. The largest variations with age are for *P. aeruginosa*, which had a prevalence of 20.5% among 18-24 year-olds, increasing to a prevalence of 54.7% for 35-44 year-olds before decreasing slightly to 49.3% for pwCF aged 45 and older. Conversely, the prevalence of *S. aureus* decreased from 50.8% for 18-24 year-olds to 24.9% for pwCF at 45+ years of age (Figure 3.16 and Table 3.3). The prevalence of less common organisms is fairly consistent, at less than 10% across the age groups, with the exception of *Aspergillus* spp and *H. influenzae*.

**FIGURE 3.16:** ACFDR ADULTS 2025: PREVALENCE OF RESPIRATORY MICROORGANISMS BY AGE

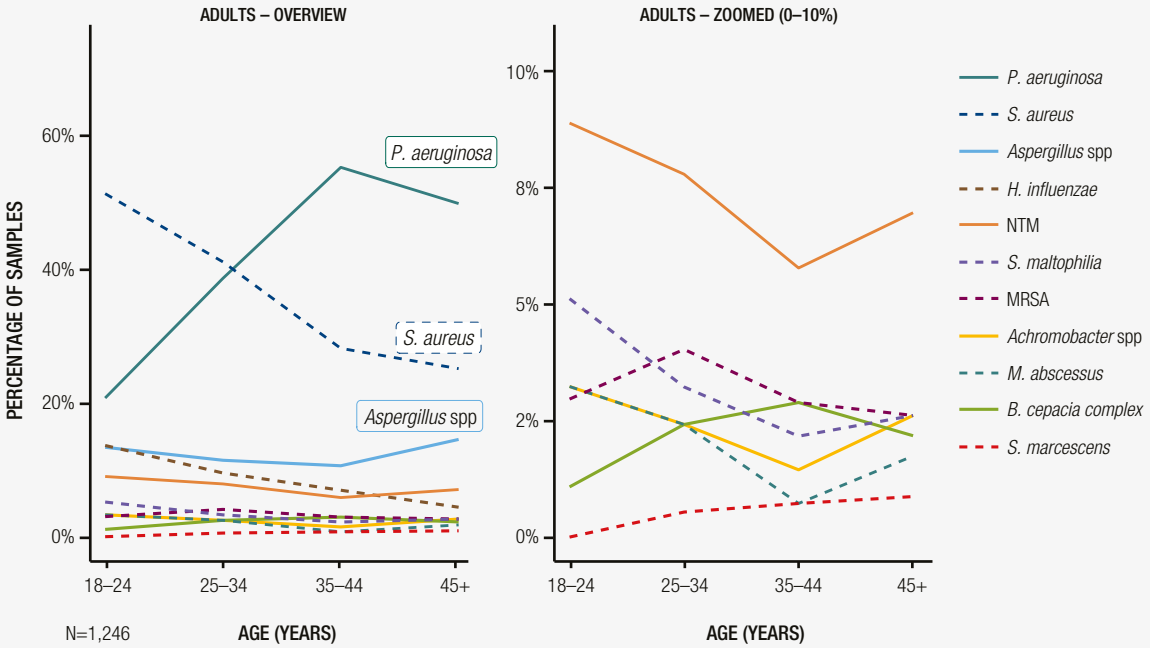


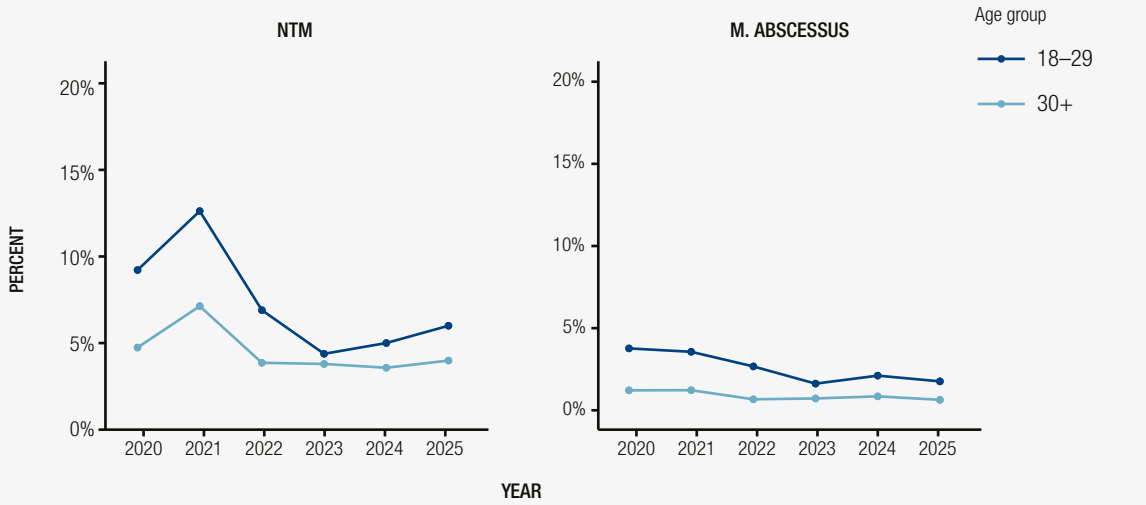

**TABLE 3.3:** ACFDR ADULTS 2025: PREVALENCE OF RESPIRATORY MICROORGANISMS BY AGE

| All samples | | | | |
|---|---|---|---|---|
| | 18-24 | 25-34 | 35-44 | 45+ |
| Total patients in the age range | 643 | 728 | 550 | 419 |
| Samples taken | 370 | 371 | 276 | 229 |
| Patients who provided a sample | 362 | 364 | 276 | 226 |
| *P. aeruginosa* | 76 / 370 (20.5%) | 142 / 371 (38.3%) | 151 / 276 (54.7%) | 113 / 229 (49.3%) |
| *H. influenzae* | 50 / 370 (13.5%) | 35 / 371 (9.4%) | 19 / 276 (6.9%) | 10 / 229 (4.4%) |
| *B. cepacia complex* | <5 | 9 / 371 (2.4%) | 8 / 276 (2.9%) | 5 / 229 (2.2%) |
| *S. aureus* | 188 / 370 (50.8%) | 151 / 371 (40.7%) | 77 / 276 (27.9%) | 57 / 229 (24.9%) |
| MRSA | 11 / 370 (3.0%) | 15 / 371 (4.0%) | 8 / 276 (2.9%) | 6 / 229 (2.6%) |
| *Achromobacter* spp | 12 / 370 (3.2%) | 9 / 371 (2.4%) | <5 | 6 / 229 (2.6%) |
| *S. maltophilia* | 19 / 370 (5.1%) | 12 / 371 (3.2%) | 6 / 276 (2.2%) | 6 / 229 (2.6%) |
| *S. marcescens* | 0 / 370 (0.0%) | <5 | <5 | <5 |
| *Aspergillus* spp | 49 / 370 (13.2%) | 42 / 371 (11.3%) | 29 / 276 (10.5%) | 33 / 229 (14.4%) |
| *M. abscessus* | 12 / 370 (3.2%) | 9 / 371 (2.4%) | <5 | <5 |
| NTM | 33 / 370 (8.9%) | 29 / 371 (7.8%) | 16 / 276 (5.8%) | 16 / 229 (7.0%) |

### Nontuberculous Mycobacterium (NTM) Prevalence

NTM, particularly *M. abscessus* infection, has been associated with poorer outcomes for pwCF. NTM infection rates, including by *M. abscessus* for pwCF peaked in 2021 and have since decreased. In 2025, NTM was most prevalent for 18-29 year-olds at 5.1%, followed by 3.0% of 30+ year-olds. *M. abscessus* infection rates during 2025 were 1.7% for 18-29 year-olds and 0.6% for 30+ year-olds (Figure 3.17).

**FIGURE 3.17:** ACFDR ADULTS 2020-2025: PREVALENCE OF NTM INFECTIONS BY AGE



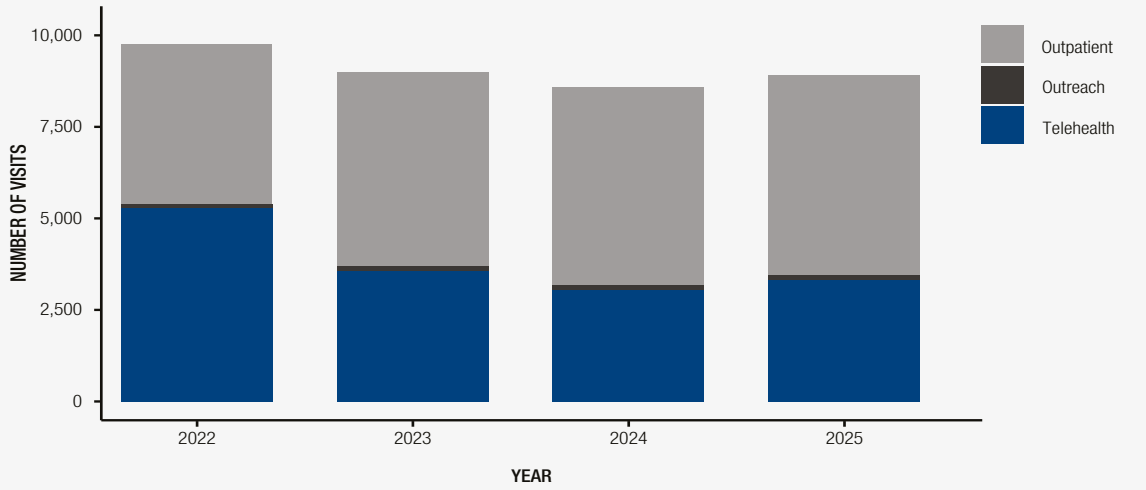

## 3.3 CF MANAGEMENT

### Clinic Visits

Traditionally, pwCF have had regular clinical visits with multidisciplinary teams. Standards of care for pwCF have recommended four clinical visits per year.

The total number of clinical visits for adults with CF has declined since 2022 (9733 visits) (Figure 3.18 and Table 3.4). For the first time since 2022, the total number of visits slightly increased in 2025. This may become a resourcing issue for adult centres over time as more pwCF reach adulthood. In 2025, there were a total of 8,915 clinical visits of which 61.2% were in a clinic, 37.3% were telehealth, and 1.5% were outreach. Overall, the proportion of visits that were telehealth have remained between 35-40% over 2023-2025.

**FIGURE 3.18:** ACFDR ADULTS 2022-2025: TOTAL NUMBER OF VISITS RECORDED PER YEAR



Historical results have been recalculated using year-specific active populations and age classifications based on age at December 31st of each reporting year, rather than age at clinic visit. Consequently, some values presented in Table 3.4 may differ slightly from those reported in previous annual reports.

**TABLE 3.4:** ACFDR ADULTS 2022-2025: TOTAL NUMBER OF CLINICAL VISITS

| Visit type | 2022 | 2023 | 2024 | 2025 |
|---|---|---|---|---|
| Outpatient | 4,338 (44.6%) | 5,292 (58.8%) | 5,410 (62.9%) | 5,454 (61.2%) |
| Outreach | 99 (1.0%) | 135 (1.5%) | 128 (1.5%) | 132 (1.5%) |
| Telehealth | 5,296 (54.4%) | 3,568 (39.7%) | 3,058 (35.6%) | 3,329 (37.3%) |
| **Total** | 9,733 (100.0%) | 8,995 (100.0%) | 8,596 (100.0%) | 8,915 (100.0%) |

## Standards of Care

The Australian CF Standards of Care for pwCF recommend four clinical visits per year. In 2025 the number of adults with CF who had at least 4 clinic visits was 1,120 (47.9%) overall (Figure 3.19). This was similar among pwCF of 18-29 years (46.1%) and those who were 30+ years old (49.2%). This proportion has declined over time for both age groups (Table 3.5).

The average number of clinical visits per adult with CF has also declined over time and is 3.8 for adults between the ages of 18-29 and 3.9 for adults 30 years or older (Table 3.6).

**FIGURE 3.19:** ACFDR ADULTS 2025: PROPORTION OF PATIENTS WITH 4 OR MORE CLINICAL VISITS

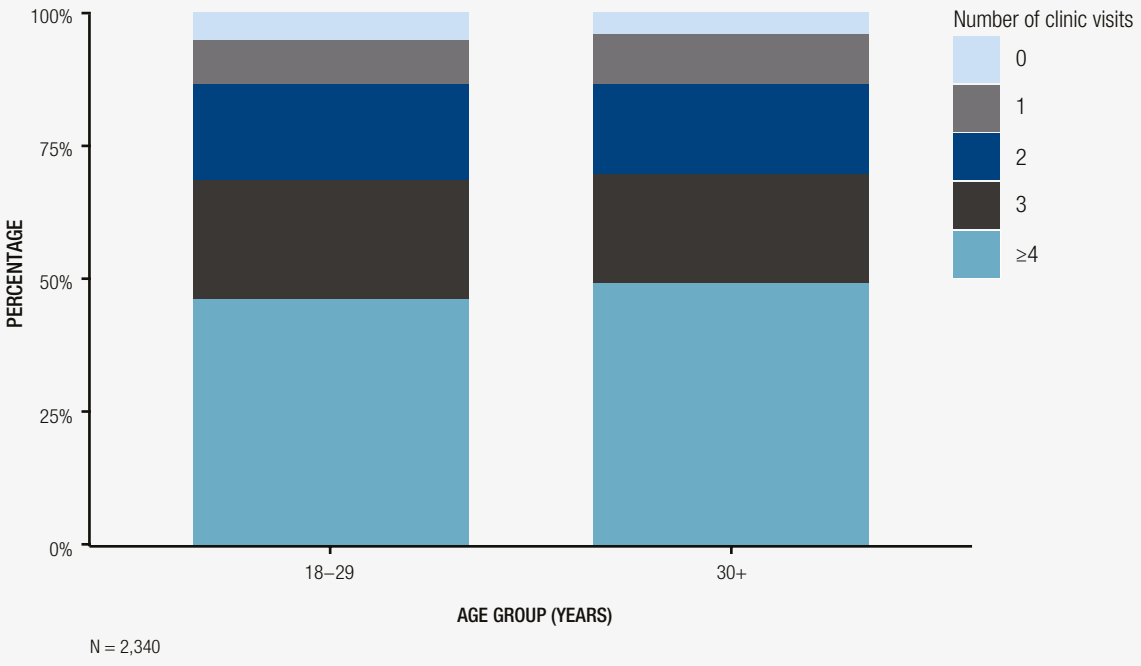



**TABLE 3.5:** ACFDR ADULTS 2022-2025: NUMBER AND PROPORTION WITH 4 OR MORE CLINICAL VISITS BY AGE

| | Number (%) with 4+ visits | | | |
|---|---|---|---|---|
| **Age** | 2022 | 2023 | 2024 | 2025 |
| 18-29 | 568 (61.0%) | 537 (57.0%) | 468 (48.6%) | 479 (46.1%) |
| 30+ | 735 (62.0%) | 646 (56.0%) | 624 (51.7%) | 641 (49.2%) |
| **Total** | 1,303 (61.5%) | 1,183 (56.0%) | 1,092 (50.4%) | 1,120 (47.9%) |

**TABLE 3.6:** ACFDR ADULTS 2025: AVERAGE NUMBER OF CLINICAL ENCOUNTERS PER PERSON

| Age | Average number of clinic visits |
|---|---|
| 18-29 | 3.8 |
| 30+ | 3.9 |
| **Total** | 3.8 |

## Hospitalisations

In 2025, data regarding adult hospitalisations was reported for all adults, excluding those with lung transplants (97), totalling 2,340 adults.

Approximately three quarters (74.8%) of adults with CF did not have any hospitalisations in 2025. Of those aged 18-29 years, 15.8% had 1 hospitalisation, 3.9% had 2 hospitalisations, and 4.7% had 3 or more hospitalisations. For adults with CF aged 30 years or more, 17.7% had 1, 5.0% had 2, and 3.3% had 3 or more hospitalisations during 2025 (Figure 3.20 and Table 3.7).

**FIGURE 3.20:** ACFDR ADULTS 2025: HOSPITALISATIONS BY AGE

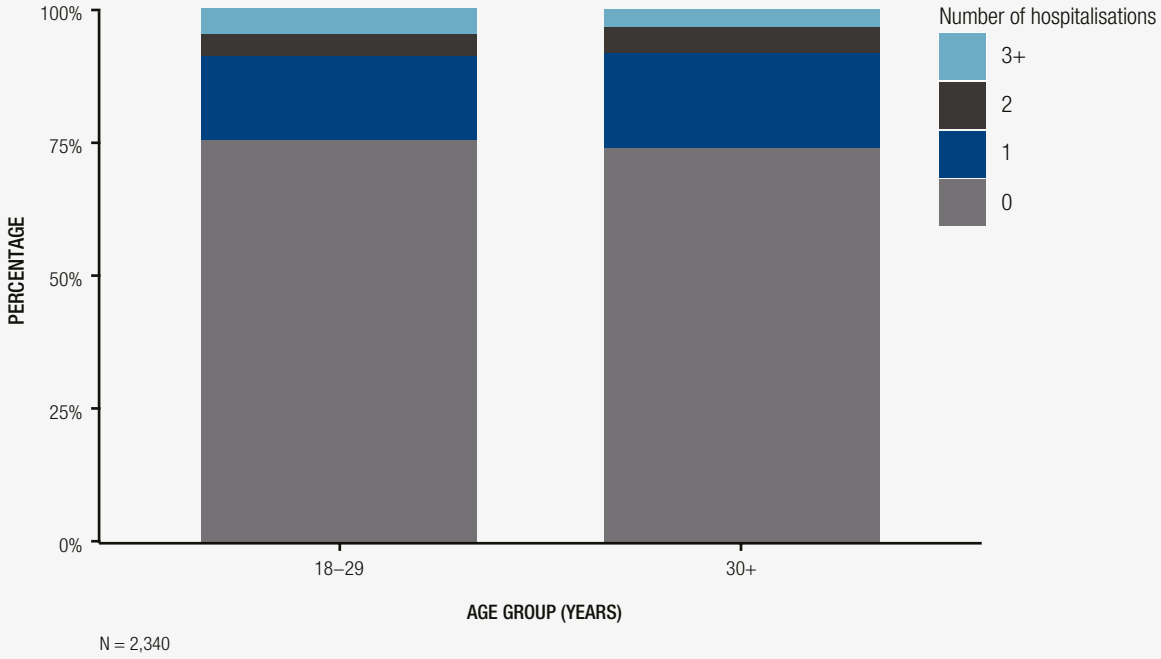



**TABLE 3.7:** ACFDR ADULTS 2025: HOSPITALISATIONS BY AGE

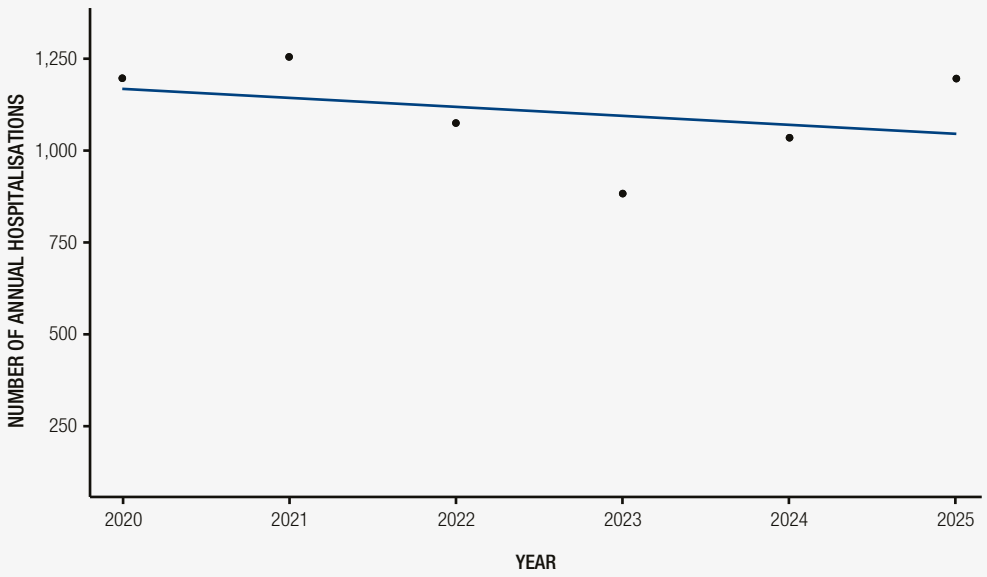
| Age | Hospitalisations | N (%) |
|---|---|---|
| 18-29 | 0 | 784 (75.5%) |
| | 1 | 164 (15.8%) |
| | 2 | 41 (3.9%) |
| | 3+ | 49 (4.7%) |

| Age | Hospitalisations | N (%) |
|---|---|---|
| 30+ | 0 | 963 (74.0%) |
| | 1 | 231 (17.7%) |
| | 2 | 65 (5.0%) |
| | 3+ | 43 (3.3%) |

## Adult Hospitalisations Over Time

Adult hospitalisations per year continued to fluctuate from 2020 to 2025, from 1,188 admissions in 2020 to a low of 874 admissions in 2023. Hospitalisations increased slightly to 1,187 in 2025, however, the overall trend has seen a reduction over the last six years (Figure 3.21).

**FIGURE 3.21:** ACFDR ADULTS 2020-2025: HOSPITALISATIONS PER YEAR

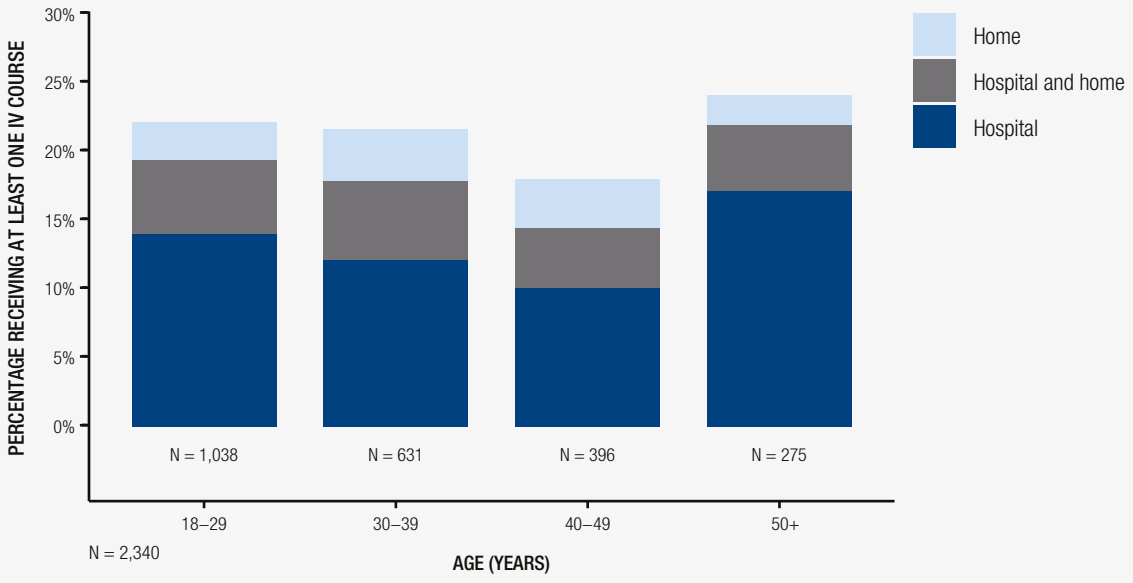



## IV Antibiotic Therapy

In 2025, there were 493 adults who received at least one course of IV antibiotic therapy; this represents 21.5% of all adults with CF (excluding lung transplant recipients). For adults who received IV antibiotic therapy in 2025, the most common setting for treatment was hospital only (61.4%), followed by combined hospital and home (24.1%), and home only (14.5%).

The age groups most likely to receive IV antibiotics were those aged 50+ years (24.0% of the cohort), followed by 18-29 years (22.1% of the cohort), 30-39 years (21.5% of the cohort), and 40-49 years (17.9% of the cohort) (Figure 3.22).

**FIGURE 3.22:** ACFDR ADULTS 2025: IV ANTIBIOTIC THERAPY BY AGE



In 2025, the median duration of IV antibiotic therapy in hospital only was 11 days, with a median of 14 days of therapy for adults receiving therapy at home or a combination of hospital and home-based therapy (Table 3.8).

**TABLE 3.8:** ACFDR ADULTS 2025: MEDIAN AND MEAN DAYS IV ANTIBIOTIC THERAPY

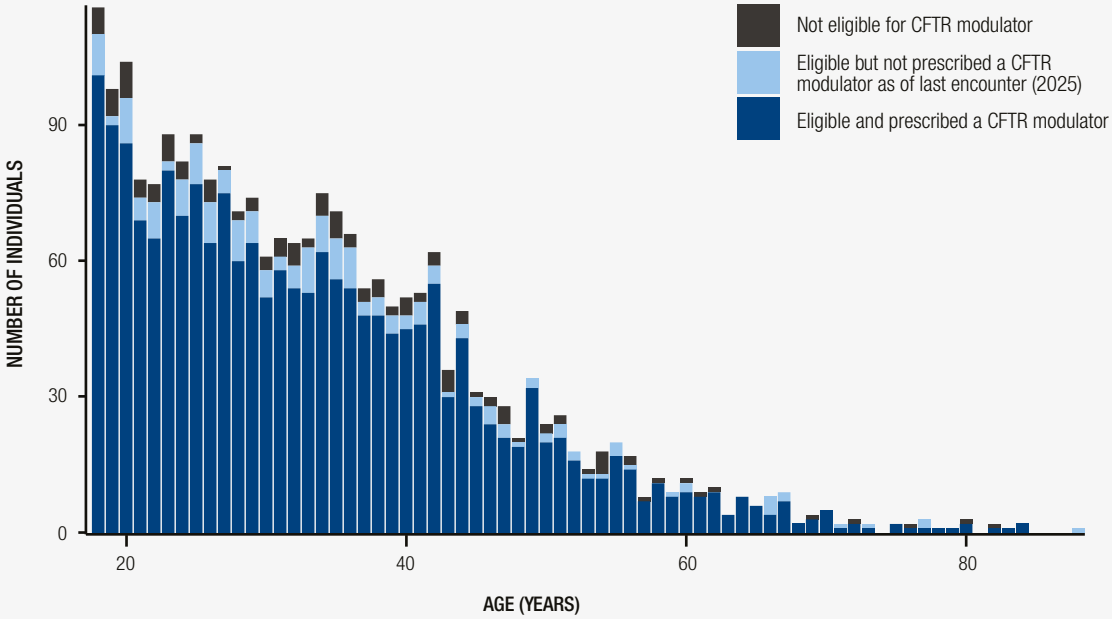
| IV antibiotic therapy location | N | Median days receiving therapy | Mean days receiving therapy | Range (days) |
|---|---|---|---|---|
| Home | 72 | 14 | 16.1 | 1 - 51 |
| Hospital | 300 | 11 | 15.6 | 1 – 105 |
| Hospital and home | 121 | 14 | 25.0 | 1 - 279 |

## CFTR Modulators

Disease-modifying therapies reduce pulmonary exacerbations, improve quality of life, and improve nutritional parameters for an increasing number of pwCF. Different therapies target different genetic variants, and not all pwCF may be eligible to receive CFTR modulators. Data were calculated from pwCF who were on a modulator as of December 31st 2025. This is generally those pwCF on modulators available via the PBS. Please refer to section 1.6 for more information regarding CFTR modulator eligibility criteria.

In 2025, 2,326 adults had their eligibility for a CFTR modulator (based on genotype) known (99.4% of adults who have not had a transplant). Figure 3.23 shows that the vast majority of these adults with known eligibility were prescribed a modulator at the end of 2025.

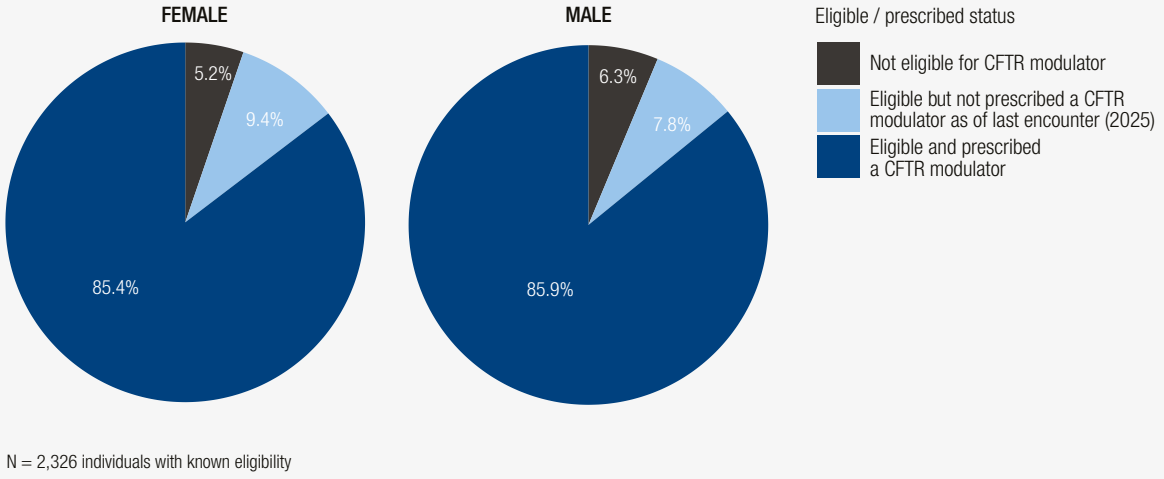
**FIGURE 3.23:** ACFDR ADULTS 2025: CFTR MODULATOR ELIGIBILITY AND PRESCRIBING BY AGE




N = 2,326 individuals with known eligibility

Of the 2,326 adults in the registry in 2025 who had known eligibility for a modulator, 5.8% were not eligible for a CFTR modulator, 85.6% were eligible and prescribed a modulator, and 8.6% were eligible but not prescribed a modulator. There were minimal differences in modulator eligibility between females and males with 5.2% of females and 6.3% of males not eligible for a CFTR modulator, 85.4% of females and 85.9% of males eligible and prescribed a modulator, and 9.4% of females and 7.8% of males eligible and not prescribed a modulator (Figure 3.24). There has been a continuing gradual increase since 2022 in the proportion of adults who are prescribed a modulator.

**FIGURE 3.24:** ACFDR ADULTS 2025: COMBINED ELIGIBILITY AND PRESCRIPTION OF CFTR MODULATORS




N = 2,326 individuals with known eligibility

The most common CFTR modulator for pwCF is Trikafta. Among those aged 18-29 years, 85.0% were receiving Trikafta, and this remained high (80.9%) in adults aged ≥30 years. Use of other CFTR modulators was low across adult groups (4.2%-6.4%), indicating a transition away from previous-generation therapies. Only a small minority were eligible but not prescribed a modulator (8.1% of 18-29 year-olds and 9.1% of adults aged 30+), and very few were classified as not eligible for modulators (2.7%-3.6%), reflecting broader eligibility criteria and near-universal access in adults. Overall, the adult data highlight widespread adoption of Trikafta, with most adults who are eligible receiving treatment, consistent with current clinical practice focused on early initiation and sustained modulator therapy (Table 3.9).

**Due to the majority of pwCF accessing Trikafta in 2025, the following modulator data has been categorised as 'Trikafta' or 'other CFTR modulator'.**

**TABLE 3.9:** ACFDR ADULTS 2025: MODULATOR USE BY AGE

| Age (Years) | Total N | Trikafta N (%) | Other CFTR modulator N (%) | Eligible but not receiving CFTR modulator N (%) | Not eligible for CFTR modulator N (%) |
|---|---|---|---|---|---|
| 18-29 | 1,035 | 880 (85.0%) | 43 (4.2%) | 84 (8.1%) | 28 (2.7%) |
| 30+ | 1,291 | 1,044 (80.9%) | 83 (6.4%) | 117 (9.1%) | 47 (3.6%) |

**Note**: Table 3.9 describes CFTR modulator use as of December 31st 2025, with age categories based on age at year end. Eligibility was assessed using age at the beginning of the year; therefore, counts may differ slightly from the eligibility/prescribing summaries presented elsewhere in the report.

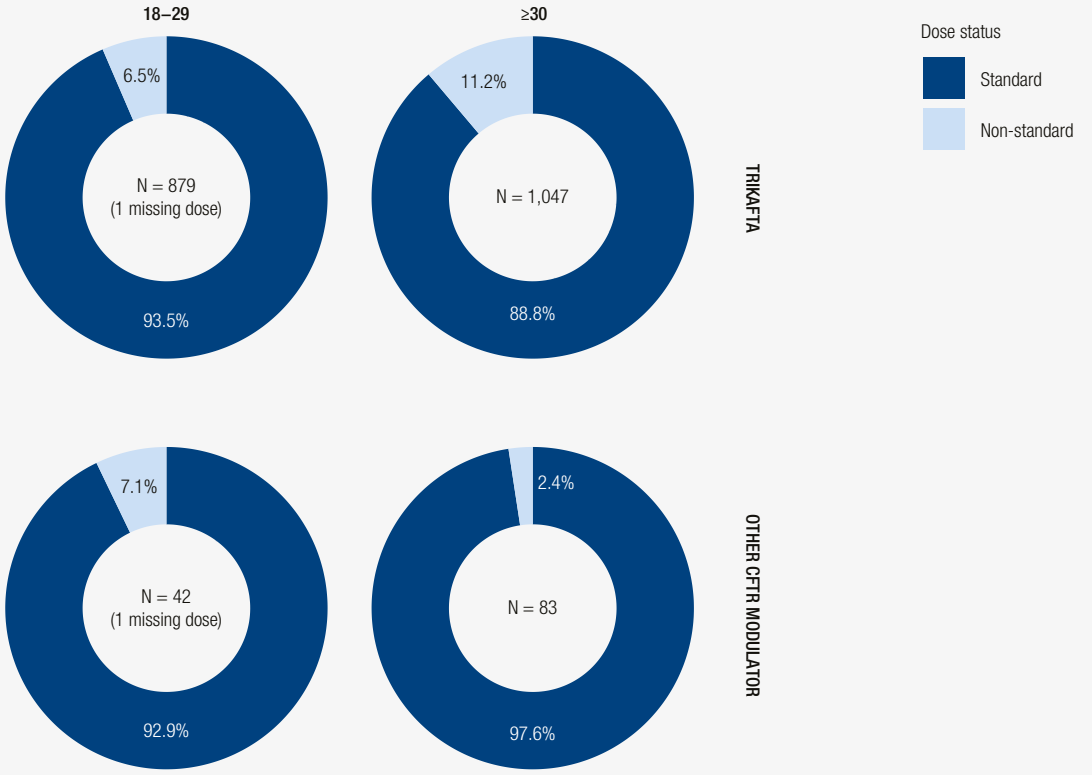
Table 3.10 shows the reasons for permanent or temporary cessation and change to modulator prescriptions. Throughout 2025, **switching from a modulator other than Trikafta to another modulator was the most common reason for discontinuation** (44 pwCF), followed by liver impairment/intolerance (14 pwCF). Unspecified intolerances or adverse events resulted in a further 10 CFTR modulator discontinuations. Discontinuations were less common for other adverse reasons such as mental health, weight gain, rash, or pulmonary side effects. A small proportion of pwCF discontinued a CFTR modulator due to other reasons including patient preference, lack of benefit, and pregnancy or pregnancy planning.

**TABLE 3.10:** ACFDR ADULTS 2025: REASONS FOR DISCONTINUATION/SWITCH FROM MODULATORS

| Reasons for discontinuation/change | Total (N = 98) | Trikafta (N = 50) | Other CFTR modulator (N = 48) |
|---|---|---|---|
| Switch to other CFTR modulator | 44 (44.9%) | 0 (0.0%) | 44 (91.7%) |
| Liver impairment/intolerance | 14 (14.3%) | 13 (26.0%) | <5 |
| Other intolerance/adverse event | 10 (10.2%) | 10 (20.0%) | 0 (0.0%) |
| Other reason (non-adverse) | <5 | <5 | <5 |
| Mental health | <5 | <5 | 0 (0.0%) |
| Weight gain | <5 | <5 | 0 (0.0%) |
| Rash | <5 | <5 | 0 (0.0%) |
| Patient preference | <5 | <5 | 0 (0.0%) |
| Pregnancy/Pregnancy planning | <5 | <5 | 0 (0.0%) |
| Lack of benefit | <5 | <5 | 0 (0.0%) |
| Pulmonary side effect/intolerance | <5 | 0 (0.0%) | <5 |

In 2025, **91% of adults on modulators were prescribed a standard dose**. Figure 3.25 shows the proportion of patients aged 18-29 and 30 years and older who were prescribed standard and non-standard doses of Trikafta and other CFTR modulators. Of those taking Trikafta, 6.5% of adults aged 18-29 and 11.2% of adults aged 30 or older were prescribed non-standard doses. Less than 5% of all adults on other CFTR modulators were prescribed non-standard doses.

**FIGURE 3.25:** ACFDR ADULTS 2025: STANDARD AND NON-STANDARD DOSE BY AGE AND MODULATOR




**Note**: Based on modulator status as of December 31st 2025. Percentages exclude missing dose information.

## 3.4 COMPLICATIONS AND THERAPIES

### CF Pulmonary Disease

In 2025, the rate of haemoptysis requiring hospital admission was 5.4% for adults aged 18-29 years including 0.9% requiring embolisation, and 6.1% for those aged 30+, including 0.7% requiring embolisation. Pneumothorax occurred in 0.3% of 18-29 year-olds and 0.1% of 30+ year-olds.

### CF Pulmonary Therapies — Maintenance Antibiotics

A mainstay of medical treatment for CF lung disease is preventive and therapeutic antibiotic therapy that may be administered orally or inhaled. Figures 3.26A and B show changes in use of adjuvant pulmonary therapies over the last six years. Overall, there has been a significant reduction in use of inhaled antibiotics and macrolides by adults with CF.

In 2025, among individuals aged 18-29, 13.9% were prescribed inhaled antibiotics (approximately one-third of the proportion recorded in 2020); 15.9% were using macrolides (approximately two-fifths of the proportion recorded in 2020); and 9.7% were on regular oral antibiotics (approximately two-fifths of the proportion recorded in 2020) (Figure 3.26A).

In the 30+ age group maintenance antibiotic use has halved since 2020. In 2025, 23.0% of the cohort were on inhaled antibiotics, 8.9% were on regular oral antibiotics, and 34.1% were using macrolides (Figure 3.26B).

**FIGURE 3.26A:** ACFDR ADULTS 2020-2025: MAINTENANCE ANTIBIOTIC THERAPY FOR ADULTS AGED 18-29 YEARS

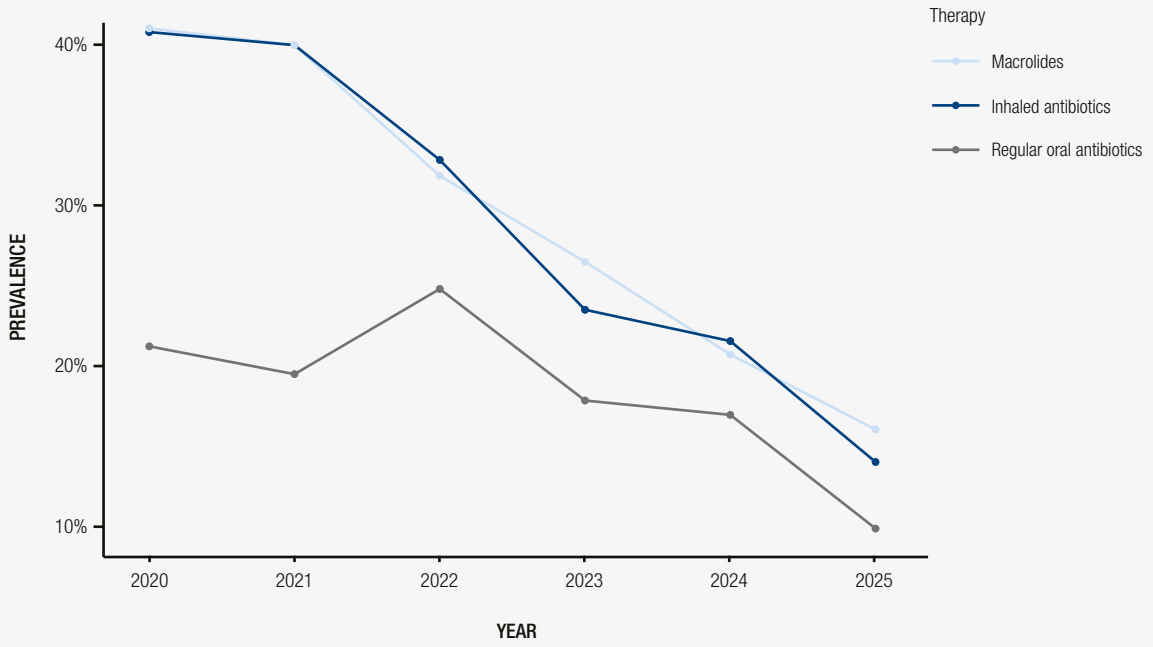
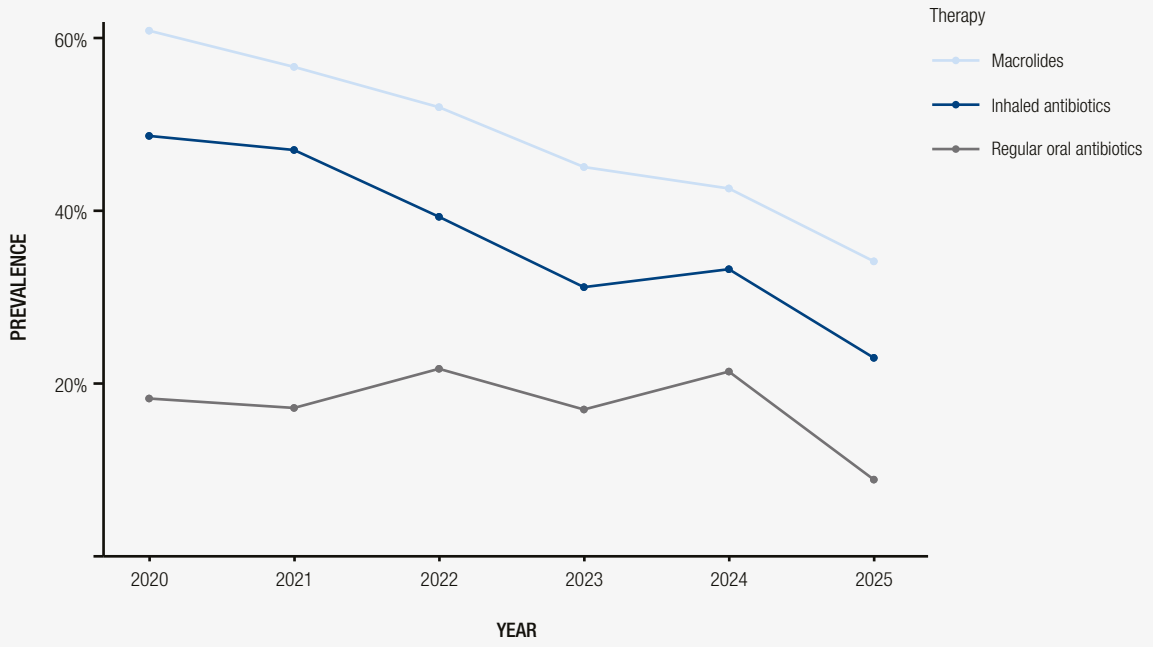



**FIGURE 3.26B:** ACFDR ADULTS 2020-2025: MAINTENANCE ANTIBIOTIC THERAPY FOR ADULTS AGED 30 YEARS AND OLDER



## CF Lung Therapies — Non-Antibiotic Management

Figures 3.27A and B show a similar reduction in many other (non-antibiotic) adjuvant pulmonary therapies from 2020 to 2025. Among both age groups there has been a reduction in use of bronchodilators, dornase alfa, hypertonic saline, and inhaled corticosteroids. There has been little change in use of the remaining therapies, which are generally required for those pwCF who have more severe disease/exacerbations.

In 2025, among adults 18-29 years, 43.8% used bronchodilators, 40.9% used dornase alfa, 35.0% used hypertonic saline and 27.6% used inhaled corticosteroids. Less commonly used drugs were inhaled mannitol (4.4%), oral corticosteroids (2.7%), non-invasive ventilation (1.9%) and long-term oxygen therapy (0.2%) (Figure 3.27A).

In 2025, among adults aged 30 years or older there was a slightly higher use of bronchodilators (50.0%), inhaled corticosteroids (39.9%), dornase alfa (34.2%) and hypertonic saline (31.5%). Uncommonly, adults required oral corticosteroids (4.4%), non-invasive ventilation (3.5%) long-term oxygen therapy (2.0%) or mannitol (1.9%) (Figure 3.27B).

**FIGURE 3.27A:** ACFDR ADULTS 2020-2025: NON-ANTIBIOTIC LUNG THERAPIES FOR ADULTS AGED 18-29 YEARS

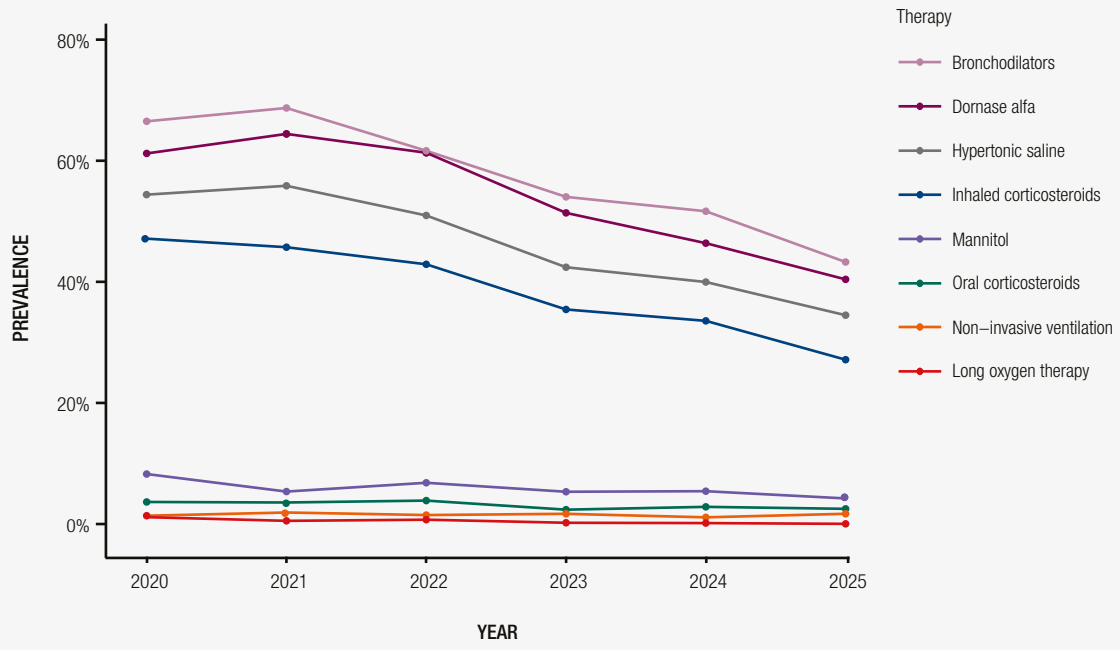
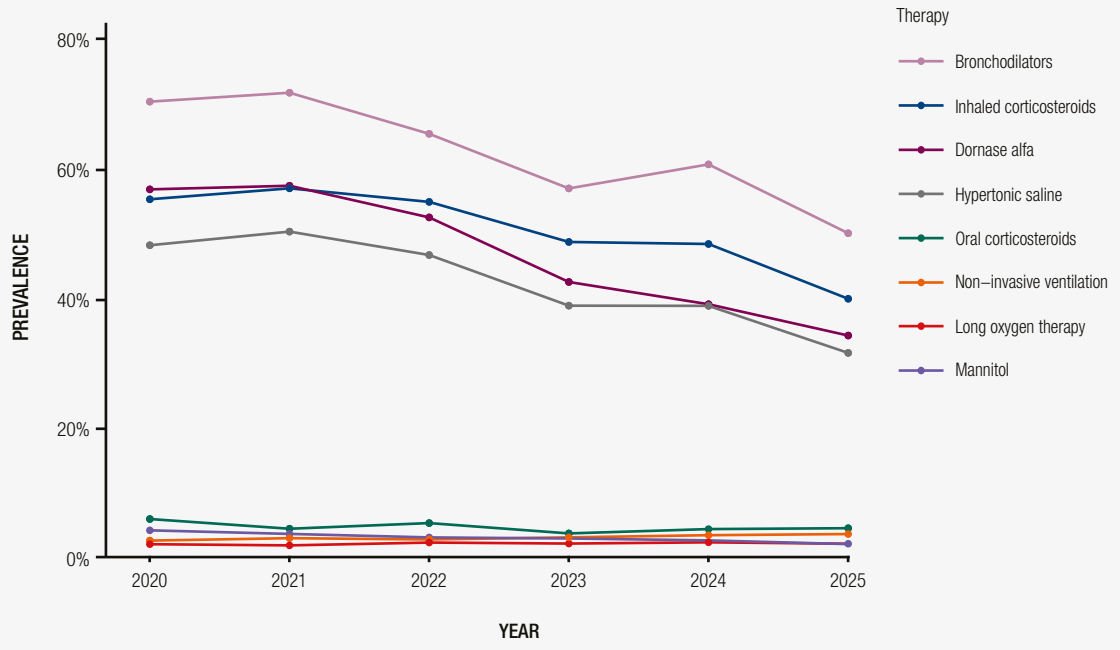



**FIGURE 3.27B:** ACFDR ADULTS 2020-2025: NON-ANTIBIOTIC LUNG THERAPIES FOR ADULTS AGED 30 YEARS AND OLDER



## CF-Related Diabetes

People with CF are at risk of impaired glucose tolerance and diabetes due to CF-related pancreatic disease. Since the introduction of CFTR modulators, there has been some reduction in the proportion of adults with impaired glucose tolerance. The proportion of pwCF aged 18-29 years with impaired glucose tolerance has decreased from 14.8% in 2020 to 11.0% in 2025. For pwCF aged 30 years and older, the proportion with impaired glucose tolerance has decreased from 15.3% in 2020 to 12.9% in 2025. In 2025, the proportion of those with diabetes was 21.8% for 18-29 year-olds and 29.5% for those aged 30 years or older. While there appears to be a downward trend for diabetes prevalence for 18-29 year-olds, the rate for those 30+ years seems more consistent (Figures 3.28A and B).

**FIGURE 3.28A:** ACFDR ADULTS 2020-2025: DIABETIC STATUS FOR ADULTS AGED 18-29 YEARS

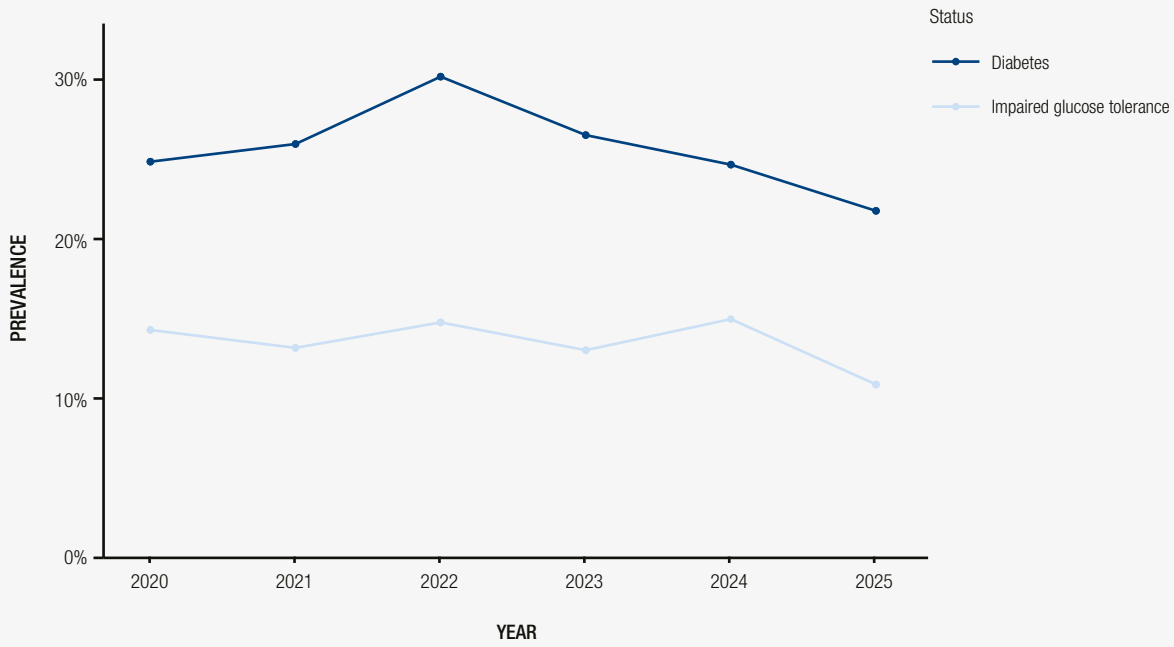
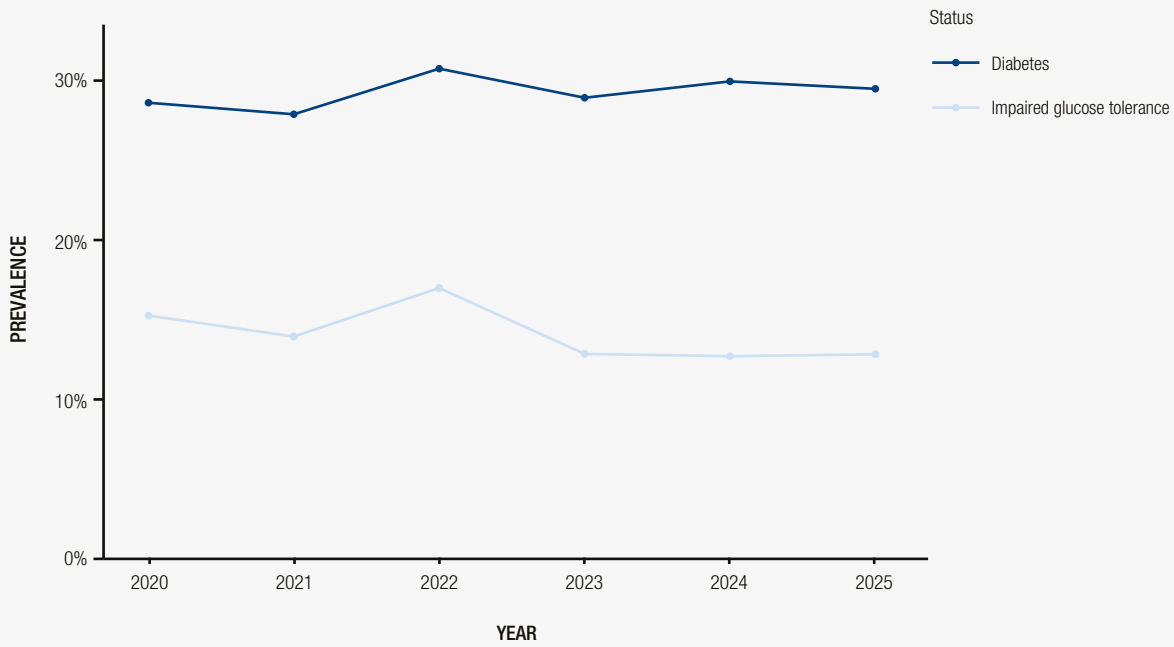



**FIGURE 3.28B:** ACFDR ADULTS 2020-2025: DIABETIC STATUS FOR ADULTS AGED 30 YEARS AND OLDER



Of those with diabetes, the vast majority are treated with insulin: 82.0% of 18-29 year-olds and 71.0% of those 30 years or older. Diet and lifestyle management only was the second most frequent treatment, with 10.0% of 18-29 year-olds and 19.5% of those 30+ years of age using this treatment. Hypoglycemics on their own and in conjunction with insulin were less common, as was using no treatment for diabetes (Table 3.11).

**TABLE 3.11:** ACFDR ADULTS 2025: DIABETIC TREATMENT BY AGE

| Diabetes treatment type | 18-29 (N = 211) | 30+ (N = 365) |
|---|---|---|
| Insulin | 173 (82.0%) | 259 (71.0%) |
| Hypoglycemics | 6 (2.8%) | 20 (5.5%) |
| Insulin and hypoglycemics | <5 | 6 (1.6%) |
| Diet/lifestyle management only | 21 (10.0%) | 71 (19.5%) |
| No treatment for diabetes | 9 (4.3%) | 9 (2.5%) |

Of those that use insulin, the vast majority (90.3% of 18-29 year-olds and 88.7% of 30+ year-olds) required chronic insulin administration. Only 3.0% of 18-29 year-olds and 3.8% of 30+ year-olds use insulin intermittently (Table 3.12).

**TABLE 3.12:** ACFDR ADULTS 2025: INSULIN USE BY AGE

| Insulin use | 18-29 (N = 165) | 30+ (N = 291) |
|---|---|---|
| Chronic insulin use | 149 (90.3%) | 258 (88.7%) |
| Intermittent insulin use | 5 (3.0%) | 11 (3.8%) |
| Insulin use, duration unknown | 11 (6.7%) | 22 (7.6%) |

## CF Gastrointestinal Disease

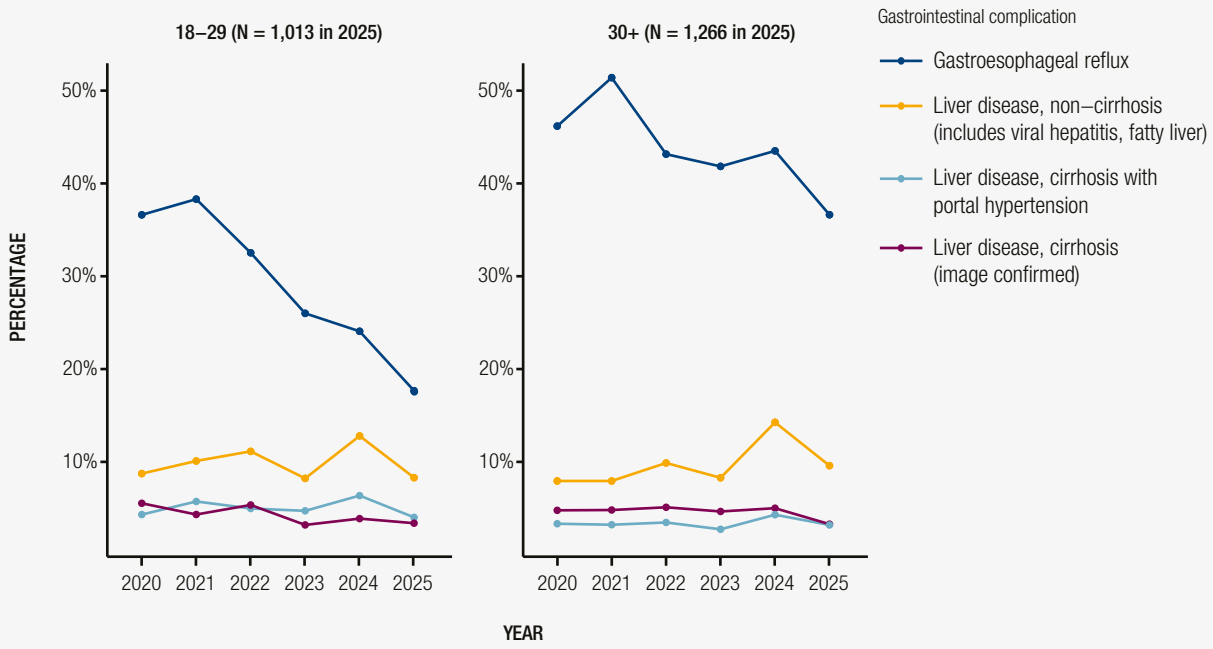
### Stomach and Liver

Gastrointestinal complications for people with CF include those related to the stomach, pancreas and liver.

Gastroesophageal reflux (GOR) has reduced significantly among adults with CF. In 2025, among adults aged 18-29 years, 17.0% experienced GOR (compared with 36.3% in 2020), and 36.3% of those aged 30 years and older experienced GOR (compared to 45.9% in 2020).

CF-associated liver disease is uncommon, and overall trends over time are less clear. The most common liver disease for pwCF is acute (non-cirrhotic) liver disease which affected 7.4% of 18-29 year-olds (similar at 7.9% in 2020) and 9.0% of 30+ year-olds in 2025 (an increase from 7.4% in 2020). Cirrhotic liver disease affected 5.6% of 18-29 year-olds (a decrease from 8.0% in 2020) and 5.3% of 30+ year-olds in 2025 (a decrease from 6.9% in 2020) (Figure 3.29).

**FIGURE 3.29:** ACFDR ADULTS 2020-2025: GASTROINTESTINAL COMPLICATIONS BY AGE




### Pancreatic Disease

The majority of adult participants in the registry were pancreatic insufficient. In 2025, 84.3% of pwCF aged 18-29, and 80.2% of those aged 30 years or older, were pancreatic insufficient. The vast majority of pwCF do not have a history of acute or chronic pancreatitis. (Table 3.13).

Compared to 2023, rates of recurrent pancreatitis have slightly reduced (from 2.2% for the 18-29 age group and 3.7% for the 30+ age group).

**TABLE 3.13:** ACFDR ADULTS 2025: PANCREATIC DISEASE BY AGE

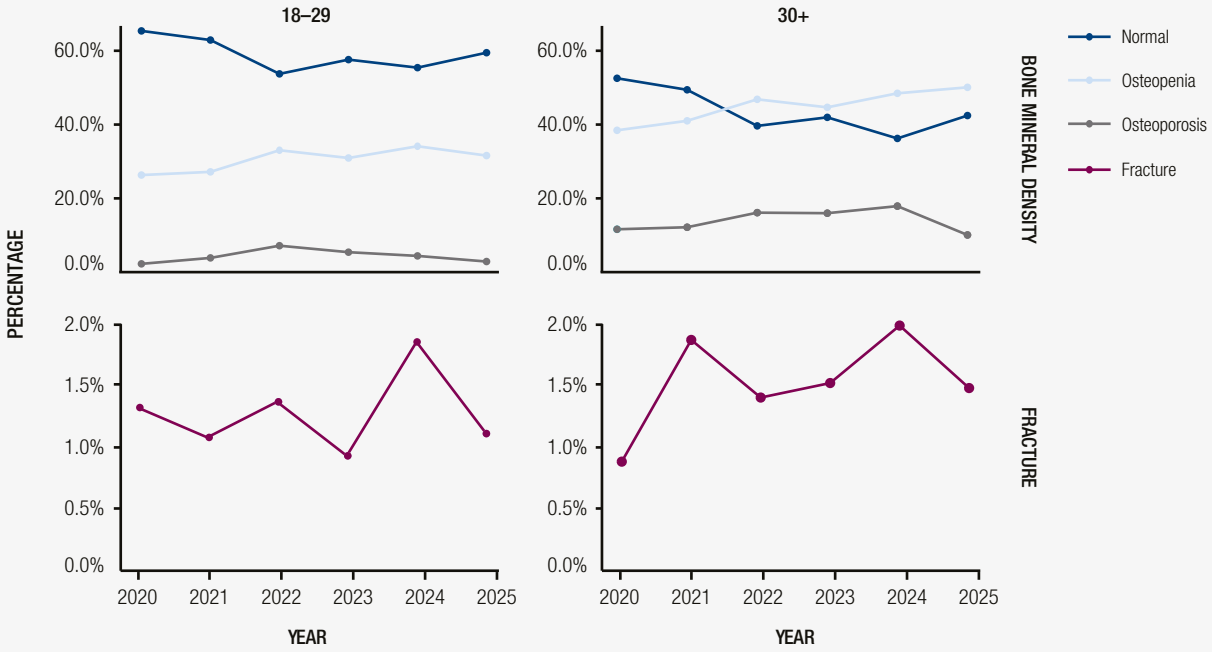
| Pancreatic status | 18-29 (N = 1,012) | 30+ (N = 1,178) |
|---|---|---|
| Insufficient | 853 (84.3%) | 945 (80.2%) |
| **Pancreatitis** | **18-29 (N = 1,009)** | **30+ (N = 1,265)** |
| No history of pancreatitis | 989 (98.0%) | 1231 (97.3%) |
| Acute (first pancreatitis event this current year) | <5 | 0 (0.0%) |
| Recurrent pancreatitis (history of more than one event of pancreatitis) | 14 (1.4%) | 31 (2.5%) |
| Pancreatitis not otherwise specified | 5 (0.5%) | <5 |

## Bone Density Status and Osteopenia

CF can cause reduced bone mineral density (osteopenia) or osteoporosis, which may increase the risk of bone fractures.

In 2025, 580 adults with CF recorded results from bone mineral density scans. Of these, 61.1% of 18-29 year-olds reported bone mineral density within the normal range, 33.6% reported osteopenia, and 5.3% reported osteoporosis. For pwCF aged 30 years or older, osteopenia was more common, reported by 49.4%, followed by 41.7% with normal range bone density and 8.9% with osteoporosis. Twenty-nine fractures were reported, with 11 (1.1%) among 18-29 year-olds and 18 (1.4%) among 30+ year-olds (Figure 3.30).

**FIGURE 3.30:** ACFDR ADULTS 2020-2025: BONE DENSITY STATUS



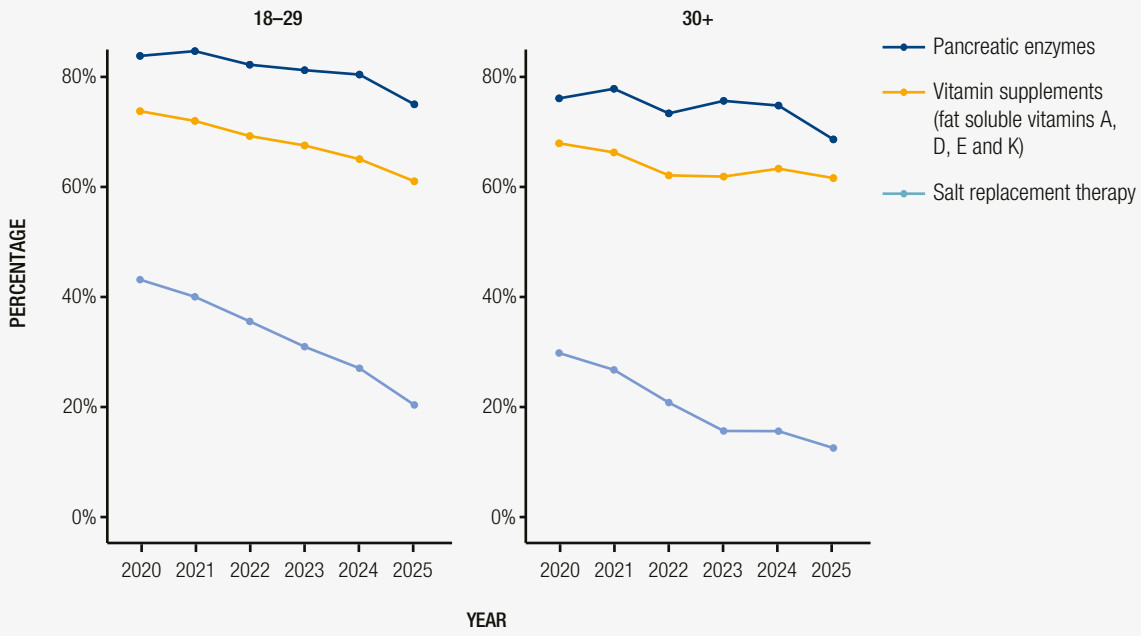

## Cancer

Of the ACFDR's adult cohort, four pwCF recorded cancer during 2025, a reduction from eleven new cancer diagnoses in 2024.

## Nutritional Supplementation

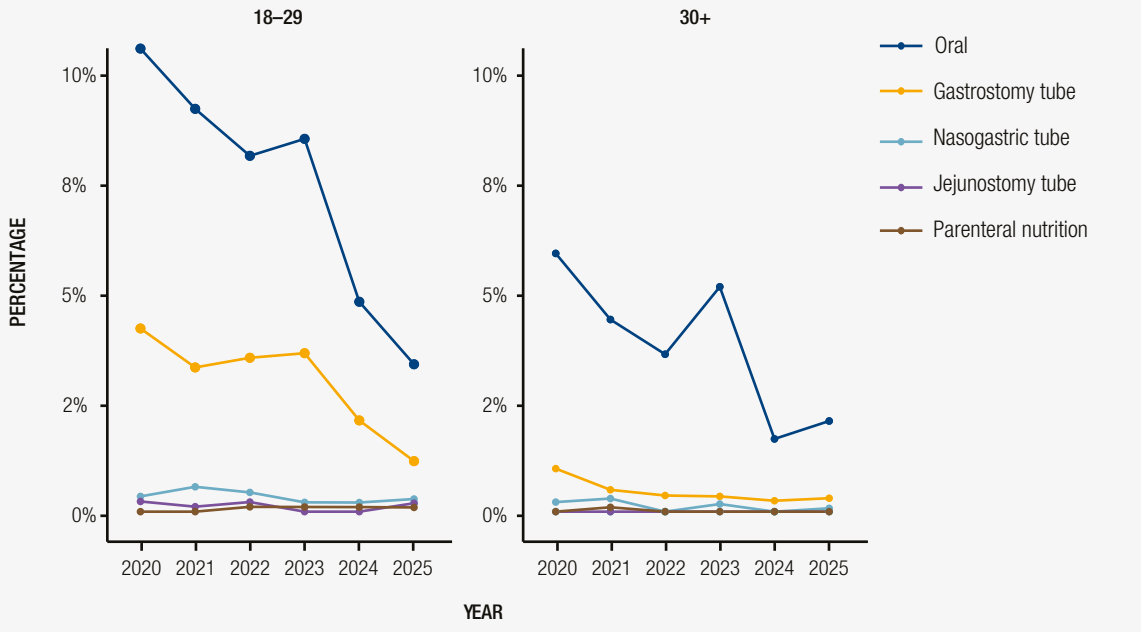
Pancreatic enzymes have been a mainstay of treatment for pancreatic insufficiency for pwCF. In 2025, 75.3% of those aged 18-29 and 69.3% of those aged 30 years or older used pancreatic enzymes, a reduction from 83.7% and 76.4% respectively in 2020. Similarly there has been a reduction in the use of fat-soluble vitamin supplements during this time from 74.2% to 62.0% for 18-29 year-olds and from 68.6% to 62.6% for those aged 30 years or older. There has been a large reduction in adults with CF using salt replacements, from 45.1% to 23.4% for 18-29 year-olds and 32.4% to 16.0% of 30+ year-olds respectively using salt replacement in 2025 (Figure 3.31).

**FIGURE 3.31:** ACFDR ADULTS 2020-2025: NUTRITIONAL SUPPLEMENTS BY AGE




Use of nutritional support has declined amongst adults with CF since 2020. Between 2020 and 2025, oral supplementation decreased from 10.6% to 3.4% of 18-29 year-olds and 5.9% to 2.1% of 30+ year-olds. Supplementation via a gastrostomy tube has decreased with both time and age, with 1.2% of 18-29 year-olds and 0.3% of 30+ year-olds accessing the treatment in 2025 compared with 4.2% and 1.0% respectively in 2020. Supplementation via a nasogastric tube, jejunostomy tube, or parenteral nutrition were accessed by less than 10 adults with CF combined in 2025 (Figure 3.32).

**FIGURE 3.32:** ACFDR ADULTS 2020-2025: NUTRITIONAL SUPPORT BY AGE



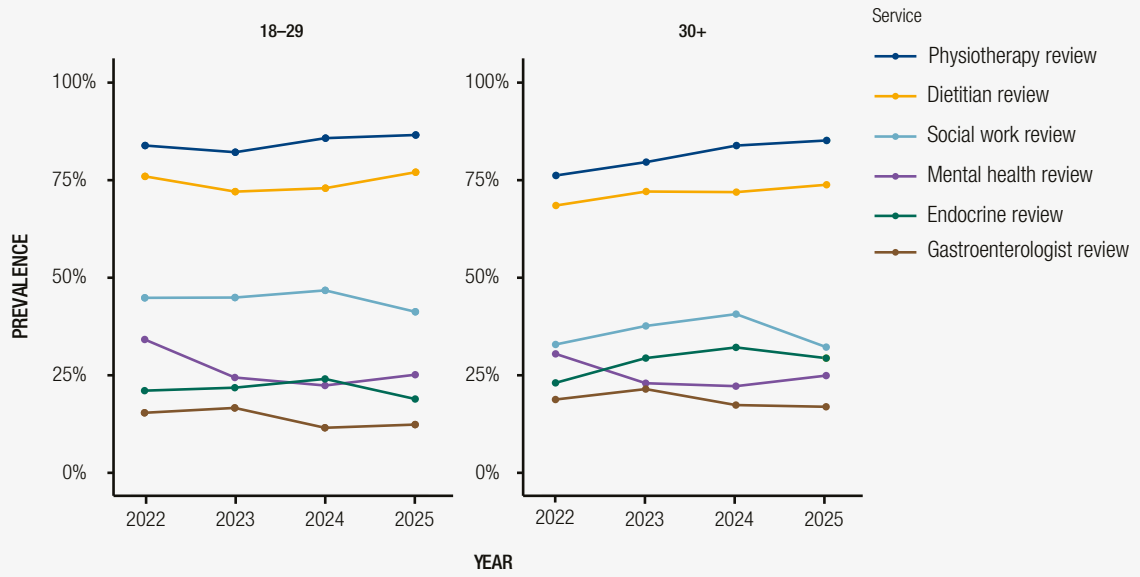
## Multidisciplinary Care

Multidisciplinary care is a mainstay of CF treatment, and for the last few years the ACFDR has been recording the proportion of pwCF who have annual reviews by allied health and medical specialists.

Figure 3.33 shows that the majority of people with CF participate in annual physiotherapy and dietitian reviews. Of pwCF aged 18-29 in 2025, over two-thirds participated in annual social work reviews; over one-quarter participated in mental health reviews, one-fifth participated in endocrine reviews and approximately one-sixth participated in gastrointestinal reviews. In general, prevalence of annual reviews has been similar for this cohort over the last few years, with a slight reduction in mental health and gastrointestinal reviews.

Of adults with CF who are 30 years or older, approximately one-third participated in social work and endocrine reviews, over one-quarter participated in mental health reviews and one-fifth participated in gastrointestinal reviews. For this age group, physiotherapy, dietitian and endocrine reviews have increased slightly over time, while mental health reviews have declined slightly.

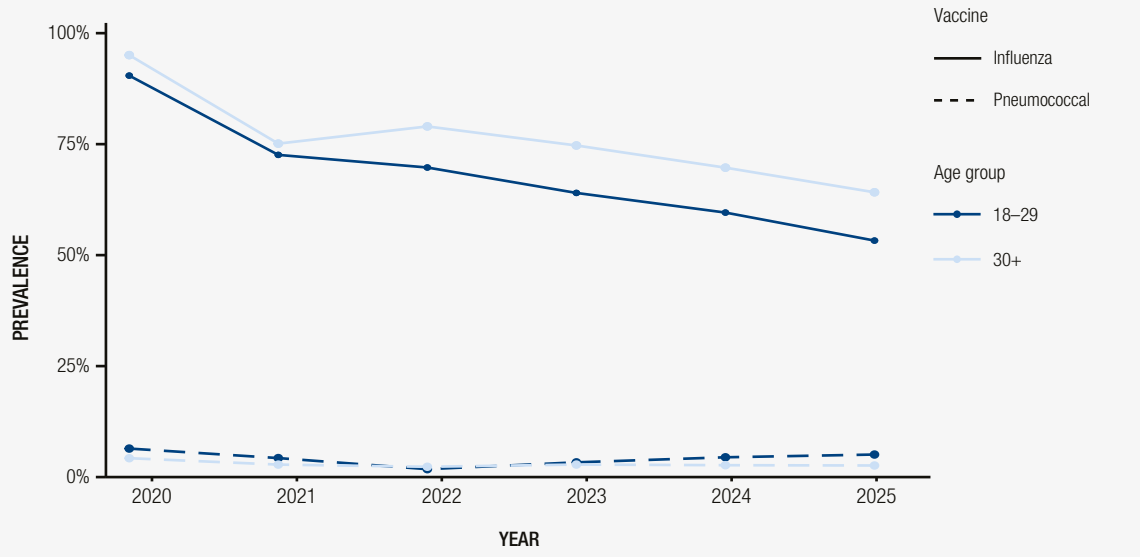
**FIGURE 3.33:** ACFDR ADULTS 2022-2025: MULTIDISCIPLINARY CARE REVIEWS BY AGE




## Vaccination

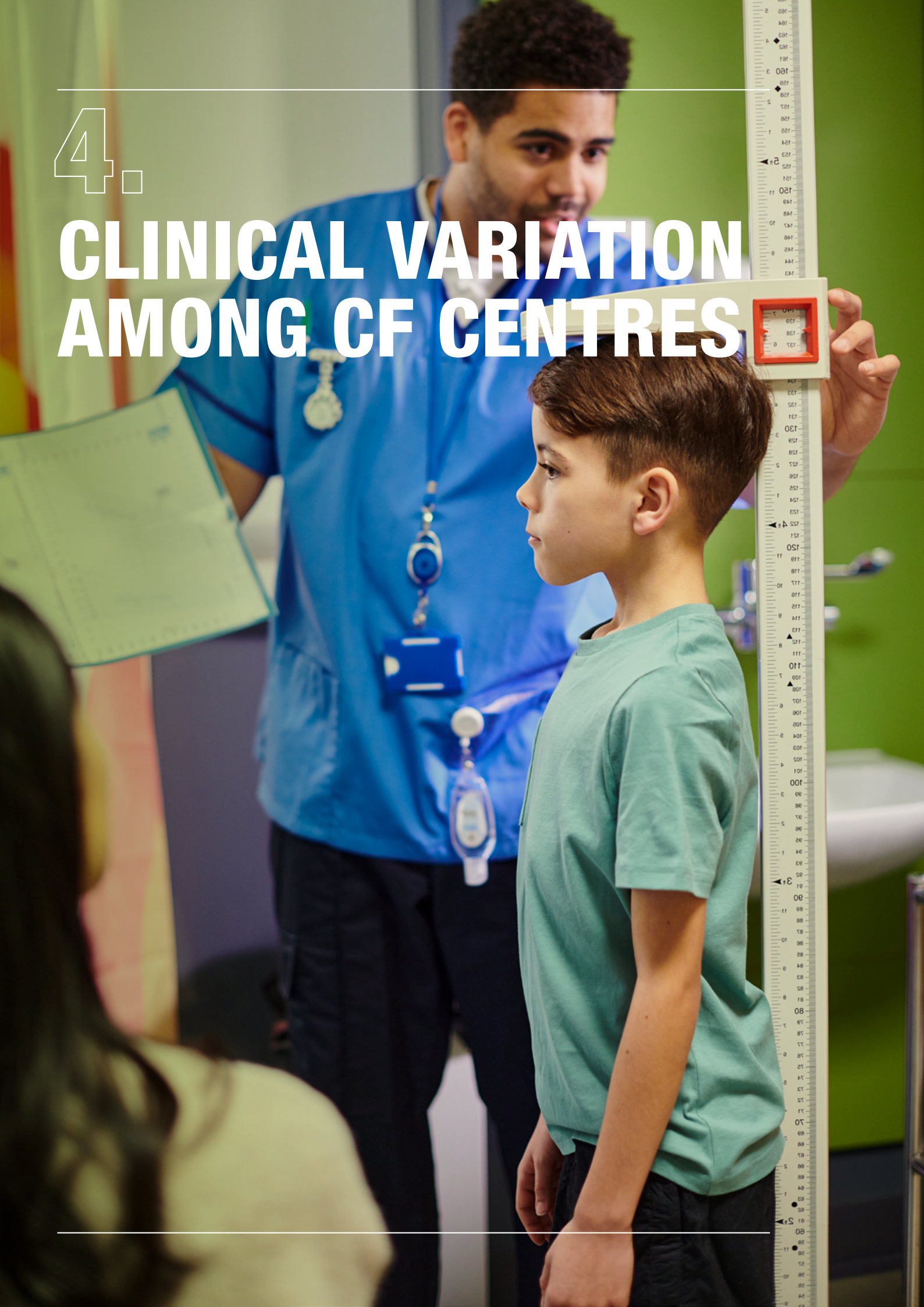
In 2025, 68.5% and 65.6% of adults with CF recorded influenza and pneumococcal vaccination status respectively. Of these, 52.9% of adults aged 18-29 years, and 63.7% of adults aged 30 years or older reported receiving an influenza vaccination. This is a decrease from 2020 when 90.0% of 30+ year-olds and 94.6% of 18-29 year-olds reported receiving an influenza vaccination (Figure 3.34)

**FIGURE 3.34:** ACFDR ADULTS 2020-2025: VACCINATIONS BY AGE



# 4.

# CLINICAL VARIATION AMONG CF CENTRES



# 4. CLINICAL VARIATION AMONG CF CENTRES

## 4.1 RISK-ADJUSTED CLINICAL INDICATORS (FUNNEL PLOTS)

This section of the report shows variation in processes and outcomes of care between CF centres, where paediatric patients are compared against other paediatric services, and adult patients are compared with those from other adult services. By reporting this variation in practice, the registry aims to improve clinical care.

Risk-adjusted funnel plots that identify clinical variation between CF centres are a new addition to the ACFDR annual report. Risk-adjusted funnel plots were generated to enable identification of variation in clinical outcomes and CFTR modulator uptake across participating sites.

When interpreting funnel plots:

— The horizontal axis (x-axis) shows the number of patients seen at each site being examined.

— The vertical axis (y-axis) shows the mean of each quality indicator by site.

— The overall mean across all sites is shown by the horizontal green line.

— The two contour lines above and below this red line represents the 95% and 99.8% control limits.

Any site crossing the 99.8% control limit may be deemed a statistical outlier and further evaluation may be necessary to identify the cause of this variation in outcome from the rest.

FEV 1pp plots are risk adjusted for age, sex and height, when calculating FEV1 pp for pwCF homozygous for F508del.

FEV1 pp is used for all CF centres; BMI for adult CF centres; and BMI percentiles for paediatric CF centres.

Age groups are based on patient age at December 31st 2025. Measures are aligned with methods used in the United States Cystic Fibrosis Foundation's Patient Registry, whereby annual measures of lung function, weight and height are reported as an average of the maximum value from each quarter.

For pwCF who had measurements taken at more than one centre, annual values contributing to the charts and tables for each of these centres were compiled from all clinical measurements reported by any centre.

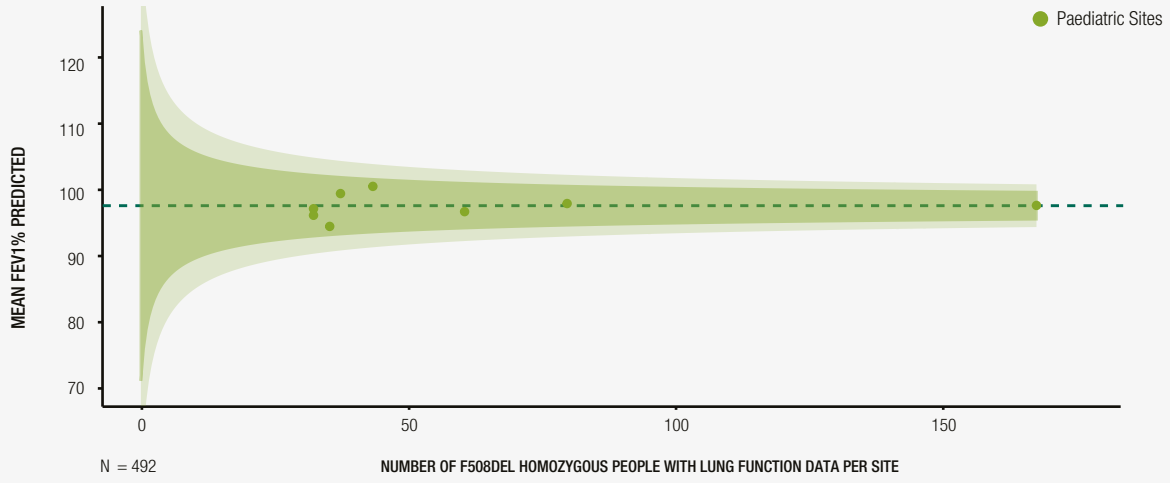
Height and BMI percentiles were calculated using the World Health Organization (WHO) growth chart, and Weight percentiles were calculated using the Centers for Disease Control and Prevention (CDC) growth chart. The included population at each centre comprises patients whose status in the registry overall is current at the end of the reference year and who had clinical measurements taken at the centre and reported to the registry during that year. Figures and tables in the report are shown for those with the data available.

## Paediatrics: Variation in Lung Function

To support accurate benchmarking, the lung function for this comparative plot includes only pwCF from paediatric CF centres (n=8) who are homozygous for F508del (Figure 4.1).

Each circle represents a site, with the x-axis showing the number of F508del homozygous individuals at that site, and the y-axis showing the mean FEV1 pp. The solid horizontal line at 97% represents the overall average FEV1 pp across all sites. The shaded areas indicate the 95% and 99.8% control limits, which reflect the expected variation due to chance. The data of two paediatric sites are not shown due to small numbers (<20). The remaining sites display a range of mean FEV1 pp values from 94.0 to 99.9, with patient numbers varying between 33 and 167 per site.

**FIGURE 4.1:** ACFDR CLINICAL VARIATION AMONG CF CENTRES 2025: MEAN FEV1 PP AT PAEDIATRIC SITES



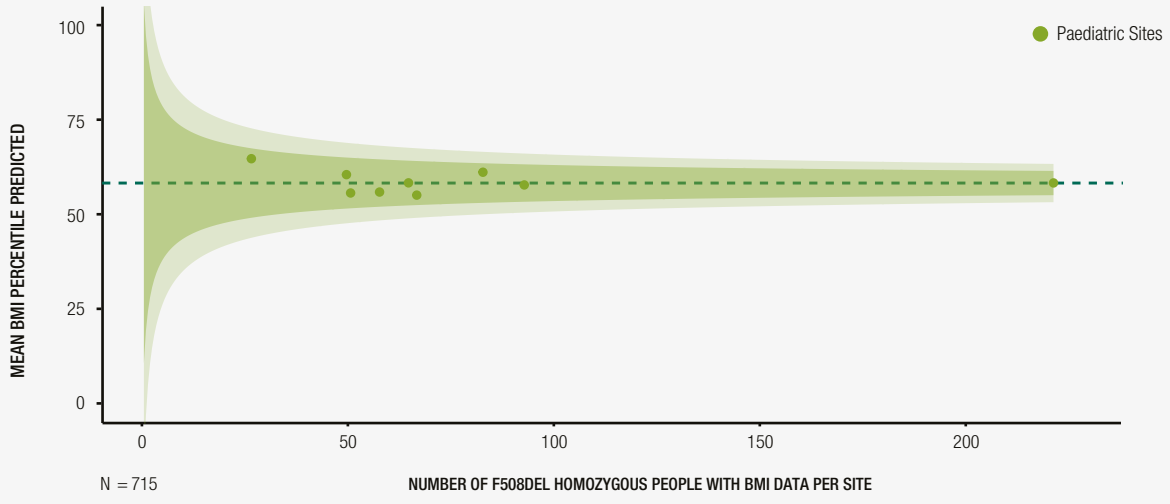

Shaded areas represent 95 and 99.8% control limits
Mean FEV1% predicted in all paediatric sites = 97%
Mean was calculated across F508del homozygous individuals with available data

* FEV1% predicted is standardised for age, sex and height.
Sites with fewer than 20 cases are not shown.

## Paediatrics: Variation in BMI

Figure 4.2 illustrates the average BMI percentiles across different paediatric sites (n=9). The horizontal line at 58% indicates the overall average BMI percentile across all paediatric sites. One site's data is not displayed due to the small number of pwCF at that site. Among the remaining sites, the mean BMI percentiles were within narrow control limits (95%), ranging from 55.0 to 64.6. The number of people for these sites varied from 27 to 221.

**FIGURE 4.2:** ACFDR CLINICAL VARIATION AMONG CF CENTRES 2025: MEAN BMI% AT PAEDIATRIC SITES



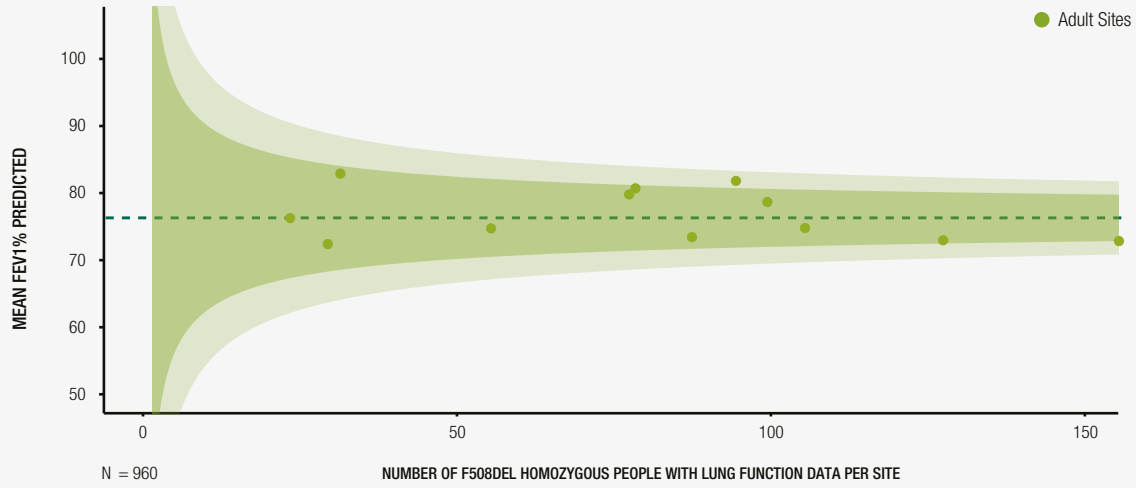

Shaded areas represent 95 and 99.8% control limits
Mean BMI percentile in all paediatric sites = 58%
Mean was calculated across F508del homozygous individuals with available data

Sites with fewer than 20 cases are not shown.

## Adults: Variation in Lung Function

Figure 4.3 illustrates ACFDR clinical variation at adult sites, focusing on mean FEV1 pp (n = 12). The plot shows the relationship between the number of F508del homozygous people with lung function data per site, with the overall mean FEV1 pp across all adult sites at 76. Most sites fall within the expected variation. One site was removed from analysis due to low numbers. The remaining sites display a range of mean FEV1 pp values from 72.4 to 82.9, with patient numbers varying between 23 and 155 per site.

**FIGURE 4.3:** ACFDR CLINICAL VARIATION AMONG CF CENTRES 2025: MEAN FEV1 PP AT ADULT SITES



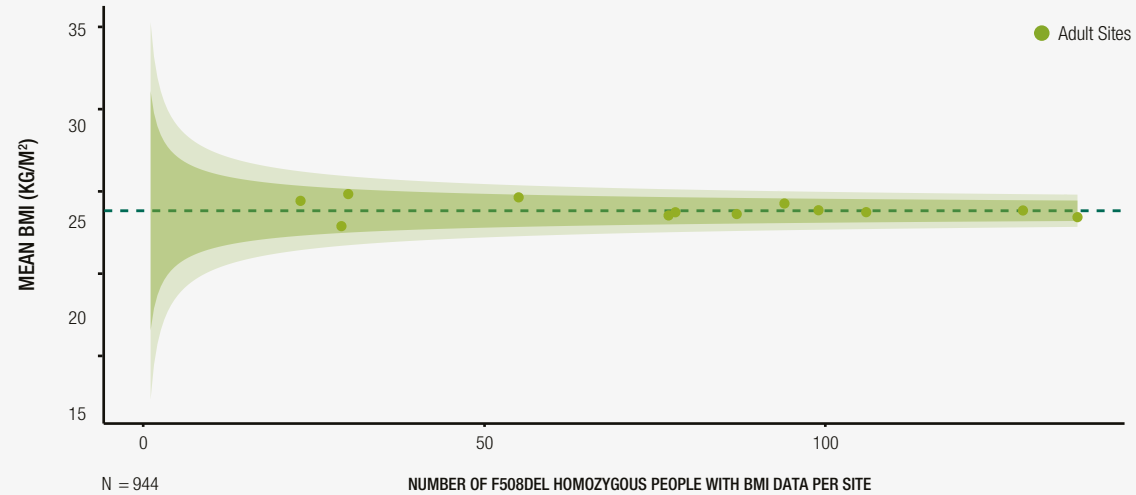

Shaded areas represent 95 and 99.8% control limits
Mean FEV1% predicted in all adult sites = 76%
Mean was calculated across F508del homozygous individuals with available data

* FEV1% predicted is standardised for age, sex and height.
Sites with fewer than 20 cases are not shown.

## Adults: Variation in BMI

Figure 4.4 presents clinical variation of mean BMI at adult sites for F508del homozygous individuals, illustrating the relationship between the number of people with BMI data per site and the mean BMI (n = 12). The overall mean BMI across all adult sites is 23.8 kg/m$^2$. Most sites fall within the expected variation limits, with one site omitted from the analysis due to low numbers. Among the remaining sites, the mean BMI were predominantly within 95% control limits, ranging from approximately 22.9 kg/m$^2$ to 24.8 kg/m$^2$. The number of people for these sites varied from 23 to 137.

**FIGURE 4.4:** ACFDR CLINICAL VARIATION AMONG CF CENTRES 2025: MEAN BMI AT ADULT SITES



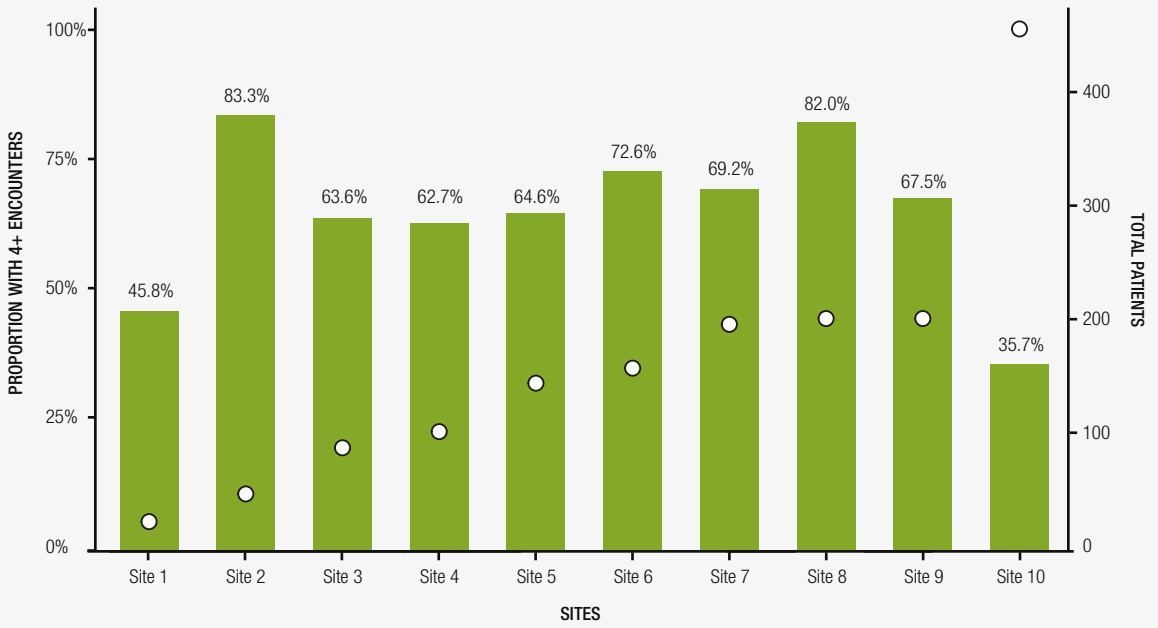

Shaded areas represent 95 and 99.8% control limits
Mean BMI in all adult sites = 23.8
Mean was calculated across F508del homozygous individuals with available data

Sites with fewer than 20 cases are not shown.

## Paediatrics: Proportion of Individuals with four or more visits per year

The proportion of pwCF with 4 or more clinical visits per year, across 10 different paediatric sites is shown in Figure 4.5. The proportions vary between sites, ranging from a low of 35.7% at Site 10 to a high of 83.3% at Site 2. Most sites have between 60-85% of pwCF receiving at least 4 annual clinical visits. The white dots indicate the total number of pwCF at each site, with the scale shown at the right.

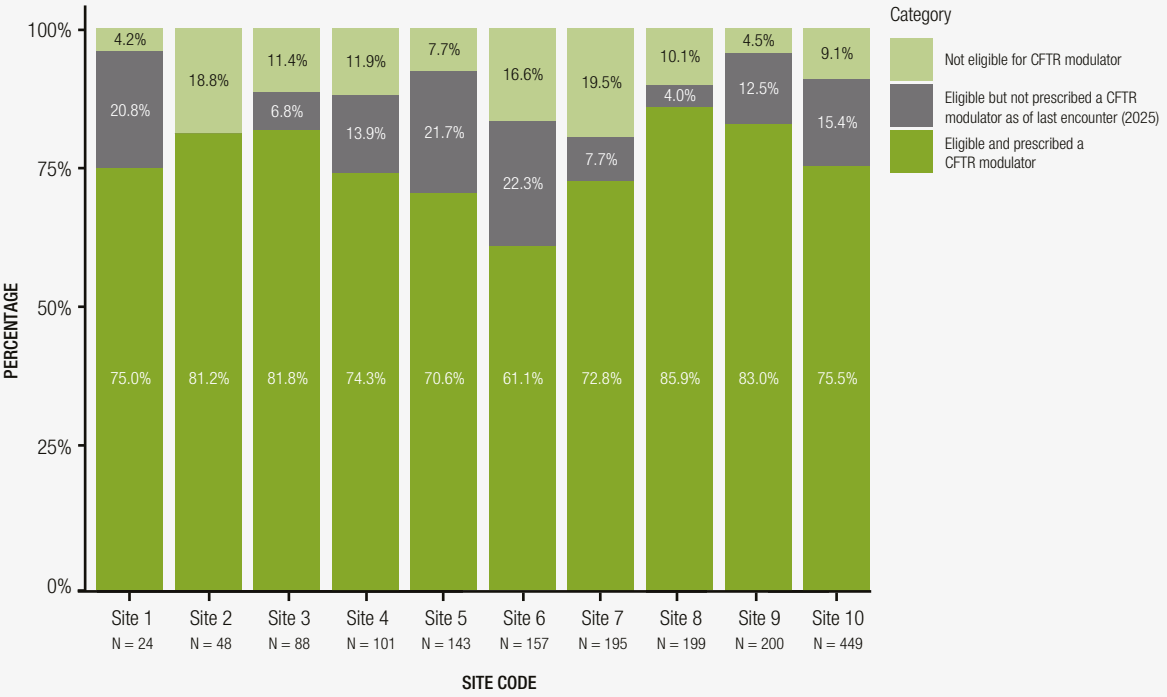
**FIGURE 4.5:** ACFDR CLINICAL VARIATION AMONG CF CENTRES 2025: PAEDIATRIC SITES WITH 4 ANNUAL CLINICAL VISITS




## Paediatrics: Proportion of eligible Individuals prescribed a modulator

The proportion of paediatric pwCF eligible for and prescribed CFTR modulators varies across 10 sites (Figure 4.6). The proportion of pwCF eligible and prescribed a CFTR modulator ranges from 61.1% (Site 6) to 85.9% (Site 8), with most sites falling between 70-85%. The proportion of pwCF who are not eligible for CFTR modulators ranges from 4.2% (Site 1) to 19.5% (Site 7). The proportion of pwCF at each site who are eligible but not prescribed a modulator vary from 0.0% (Site 2) to 22.3% (Site 6).

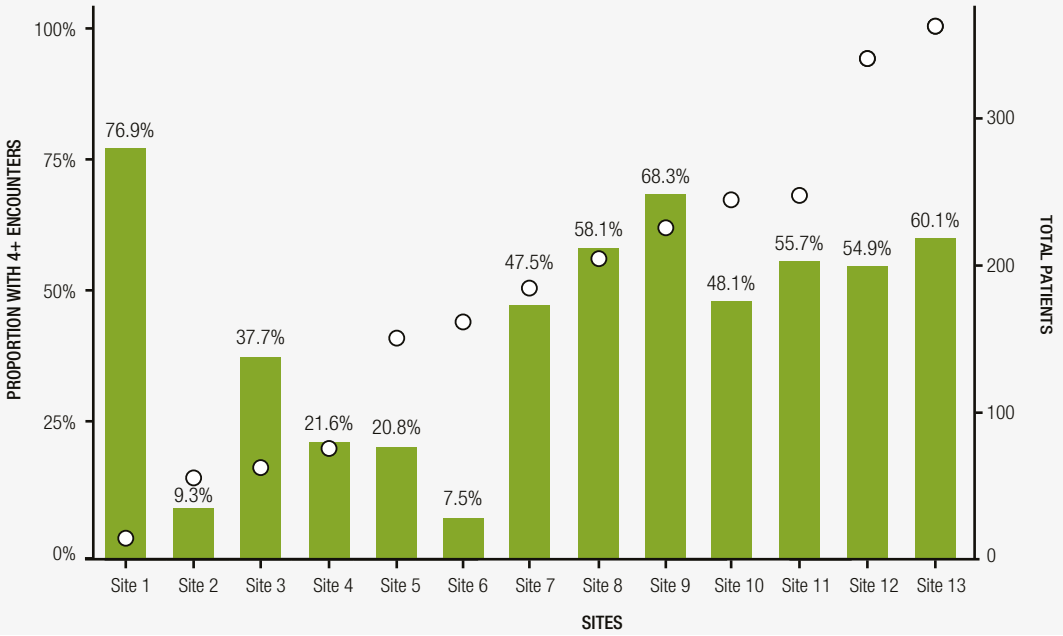
**FIGURE 4.6:** ACFDR CLINICAL VARIATION AMONG CF CENTRES 2025: PROPORTION AT PAEDIATRIC SITES THAT ARE ELIGIBLE AND PRESCRIBED A CFTR MODULATOR



## Adults: Proportion of Individuals with four visits per year

Figure 4.7 shows the proportion of pwCF with 4 annual clinical visits across 13 adult sites. The proportions range from a low of 7.5% at Site 6 to a high of 76.9% at Site 1. The majority of sites fall between 45% and 70%. Site 13 has the highest total number of pwCF, while Site 1 has the lowest number. The white dots indicate the total number of patients at each site, with the scale shown on the right.

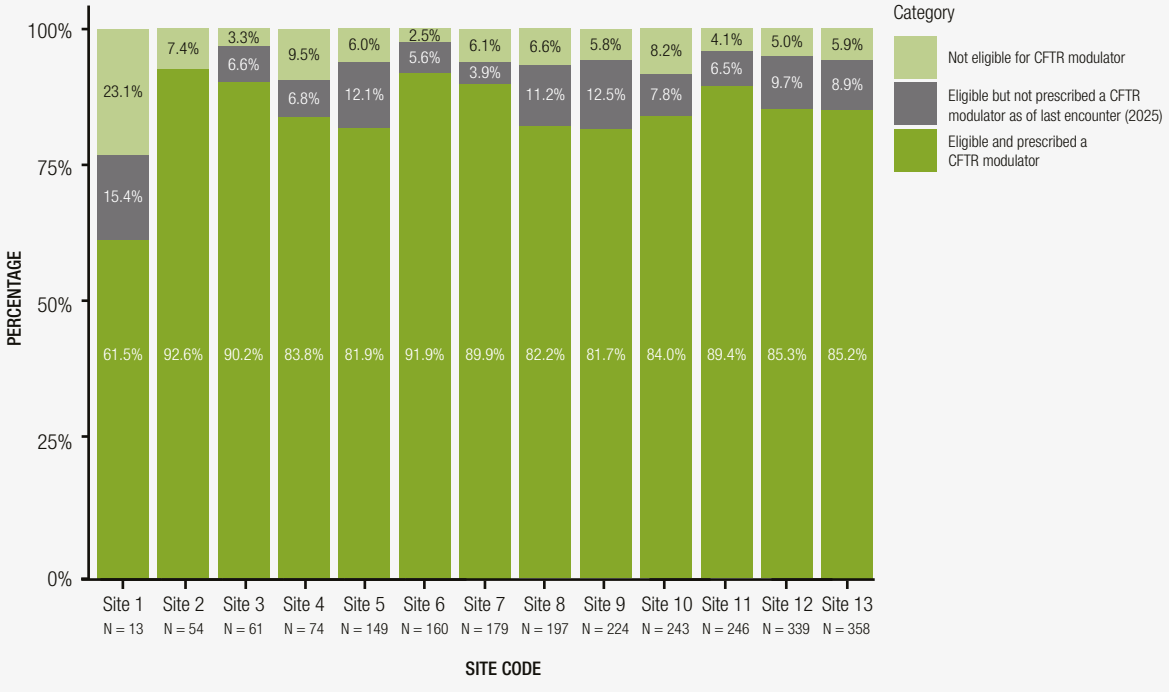
**FIGURE 4.7:** ACFDR CLINICAL VARIATION AMONG CF CENTRES 2025: ADULT SITES WITH 4 ANNUAL CLINICAL VISITS




## Adults: Proportion of eligible Individuals prescribed a modulator

The proportion of pwCF by category of eligibility and prescription for CFTR modulators across 13 adult sites is shown in Figure 4.8. The majority of pwCF at all sites are eligible and prescribed a CFTR modulator, with percentages ranging from 61.5% (Site 1) to 92.6% (Site 2). The proportion of pwCF who are not eligible to be prescribed a modulator ranges from 2.5% (Site 6) to 23.1% (Site 1). A smaller proportion of pwCF are eligible but not prescribed a CFTR modulator, varying from 0.0% (Site 2) to 15.4% (Site 1).

**FIGURE 4.8:** ACFDR CLINICAL VARIATION AMONG CF CENTRES 2025: PROPORTION AT ADULT SITES THAT ARE ELIGIBLE AND PRESCRIBED A CFTR MODULATOR



# 5. PATIENT-REPORTED OUTCOME MEASURES

Patient-reported outcome measures (PROMs) are questionnaires that capture health outcomes such as symptoms and health related quality of life directly from patients. The collection of PROMs in the ACFDR can assist with understanding patient experiences of CF and associated treatment. Furthermore, PROMs can support clinical care by promoting regular psychosocial wellbeing reviews in conjunction with the continuing health needs of pwCF.

In 2025, the ACFDR commenced a pilot of PROM surveys. This project was funded by the Department of Health, Disability and Ageing. Guided by qualitative interviews and focus groups with clinicians, pwCF and their parents/caregivers, two tools were selected for use in the ACFDR:

### Cystic Fibrosis Questionnaire Revised (CFQ-R)

The CFQ-R assesses the health-related quality of life of pwCF. Through its adult, paediatric, and parent/proxy versions, the CFQ-R is comprised of 12 subscales: physical functioning, vitality, emotion, eating, treatment burden, health perceptions, social functioning, body image, role, weight, respiratory functioning and digestion. Scores range from 0 to 100, with higher scores representing better functioning in these domains.

### Alfred Wellness Score (AWESCORE)

AWESCORE is a validated instrument for use amongst adults with CF. It consists of 10 questions under five key themes, which include, respiratory, physical, nutritional, psychological and general health. Each question in the PROM is scored using a numerical rating scale of 0 to 10. The total score ranges between 0 and 100, where a higher score demonstrates higher quality of life.

Commencing on 12th August 2025, pwCF aged 3 and older with identifiable data and/or the parents/guardians of children and adolescents with CF were invited via email to participate in PROM surveys. A maximum of three reminders were sent at fortnightly intervals to eligible participants who had not yet completed the survey.

In 2025, a total of 9 sites, including 3 paediatric sites, 5 adult sites and 1 combined paediatric/adult site participated in the collection of PROM data. Of the 1,052 people invited, 197 participants completed the survey (response rate: 18.7%). Participants included 153 adults with CF, 12 children and adolescents with CF, and 32 parents or guardians of young pwCF. While participation demonstrated feasibility of national implementation, response rates varied across sites and highlighted challenges in engagement and data completeness.

Findings from the CFQ-R indicated that adults with CF reported relatively high health-related quality of life in domains such as eating, weight, body image, and respiratory function, but lower scores in health perceptions, vitality, and treatment burden. AWESCORE results showed moderate overall wellbeing, with higher scores observed in males. For children, parent-reported outcomes were generally higher than self-reported scores, particularly for physical functioning, though differences were not statistically significant.

PROM collection will continue in 2026 for any registry participants providing a valid email address. Due to the chronic nature of CF, PROMs will be collected every 6 months to capture recent information about the wellbeing of pwCF. The PROMs program will be evaluated by the ACFDR Steering Committee in late 2026/early 2027.

# 6. ACADEMIC OUTPUTS

## Publications

Below are publications from 2025 that have used data from the Australian Cystic Fibrosis Data Registry:

Gardiner A, McGuiness O, Menadue C, Visser S, Jo H, Yozghatlian V, Aquino-Salomon T, Bortoft K, Hammond K, Yee B, Wong K, Piper A, Sivam S. Non-invasive ventilation in cystic fibrosis: the Australian experience over the past 24 years. Intern Med J. 2025 Mar;55(3):530-532. doi: 10.1111/imj.16658

Evans IES, Duplancic CM, Kidd TA, Ballard EL, Thomson RM, Wainwright CE, Bell SC, National NTM in CF study group. The changing face of cystic fibrosis research: challenges of multi-centre microbiology cohort studies. Journal of Cystic Fibrosis. 2025 Sept;24(5):903-908. doi: 10.1016/j.jcf.2025.07.002

Burgel PR, Orenti A, Cromwell E, Macek M, Gutierrez HH, Karadag B, Faro A, van Rens JG, Naehrlich L, Bakkeheim E, Carr SB, Lindblad A, Zolin A, Lammertyn E, **Ruseckaite R***, Zampoli M, Byrnes CA, da Silva-Filho L, Elbert A, Cheng SY, Stephenson AL; on the behalf of the ECFSPR scientific committee and the Global CF Registry Collaboration. Global prevalence of CFTR variants with respect to their responsiveness to elexacaftor-tezacaftor-ivacaftor. Journal of Cystic Fibrosis. 2025 Nov;24(6):1017-1026. doi: 10.1016/j.jcf.2025.10.007

**Douglas TA***, **Pourghaderi AR***, **Ahern S***, Corda J, Earnest A, **Mulrennan S***, Ranganathan S, **Ruseckaite R***, Shanthikumar S. The impact of telehealth on clinical outcomes in adults and children with cystic fibrosis in Australia. Journal of Cystic Fibrosis. Published online December 2025. doi: 10.1016/j.jcf.2025.12.017

*Current members of the ACFDR Steering Committee and/or coordinating team.

## Conference Presentations

Below are conference presentations from 2025 by members of the ACFDR coordinating team:

NZ CF Symposium 2025, Auckland, NZ, 2025. Ruseckaite R. Using CF data to improve care: How real-world registry data is shaping treatment approaches and outcomes. Invited keynote talk.

ACTA 2025 Clinical Trials and Registries Symposium, Melbourne, AUS, 2025. Hall I, Ruseckaite R, Pourghaderi AR, Honardoost MA, Liman J, Ahern S. Leveraging the Australian cystic fibrosis data registry to drive change through peer review: a pilot study. Oral presentation.

# 7. DATA ACCESS REQUESTS

**The ACFDR encourages the secondary use of its data for research and related purposes. Fifteen data access requests were received and approved for the ACFDR in 2025.**

| Date | Name | Organisation | Request type | Request |
|---|---|---|---|---|
| 7-Jan | Elena Schneider-Futschik / Rasa Ruseckaite | NSW Health Pathology / Monash University | Research | Current infection rates of NTMs for pwCF in Australia |
| 31-Jan | Claire Wainwright | Queensland Children's Hospital | Non-research | Breakdown of the number of patients and their basic FEV1 and BMI data by paediatric/adult clinics (The Prince Charles Hospital, Mater Hospital, Gold Coast University Hospital) |
| 31-Jan | Maxine Orre | Vertex Pharmaceuticals | Non-research | Update on new mutation data |
| 30-Apr | Jane Willis | Monash Health (Monash Medical Centre) | Research | A breakdown of Monash Health patients by age, diabetes status and liver disease |
| 30-Apr | Megan Window | Patient Safety and Quality, Clinical Excellence Queensland \| Queensland Health | Non-research | Request to access 2023 site reports for Queensland CF centres |
| 8-May | Maxine Orre | Vertex Pharmaceuticals | Non-research | CF patient population by age and mutation |
| 4-Jun | Lorena Rodrigues Sabino | The University of Newcastle | Research | Non F508DEL patient list |
| 11-Jul | Arul Earnest / Simran Chawla | Monash University | Research | Machine learning modelling of specific infections, poor lung function (FEV1%), and optimal BMI% among cystic fibrosis patients in Victoria and area level determinants |
| 16-Jul | Nick Mann | Australian Institute of Health and Welfare | Non-research | CF prevalence data for the Australian Burden of Disease 2026 Study |
| 29-Jul | Toni Douglas / Rasa Ruseckaite | Queensland Children's Hospital / Monash University | Research | Real world outcome in Australian children after the initiation of triple modulators |
| 29-Aug | Claire Wainwright / Kara Brady | Queensland Children's Hospital / The Prince Charles Hospital | Research | Finding the Optimal Regimen for Mycobacterium Abscessus Treatment (FORMAT) Trial |
| 22-Sep | Jo Harrison | Royal Children's Hospital | Non-research | Comparison of RCH median FEV1 for Royal Children's Hospital patients with the Australian median for years 2015 to 2023 |
| 17-Oct | Ellie Johnson / Daniel Smith | The Prince Charles Hospital | Research | Description of the health of pwCF aged 65+ with a focus on comorbidities, pharmacotherapies, and psychosocial functioning. |
| 31-Oct | Simon Visser | Royal Prince Alfred Hospital | Research | Reducing the frequency of outpatient review in people with cystic fibrosis on high-efficacy modulator therapy (REFORM) |
| 3-Nov | Elena Schneider-Futschik / Rasa Ruseckaite | The University of Melbourne / Monash University | Research | ACFDR pregnancies - Data before and after pregnancy for pwCF on ETI vs non-modulator patients |

## How can I request data from the ACFDR?

Data access requests are subject to approval by the registry's Steering Committee and relevant ethics committees, and Monash University's conditions of use. Interested researchers/individuals are advised to contact Monash University for details and to arrange consideration of their research proposal. In accordance with the ACFDR data access policy, a fee may be charged to recover the costs of data extraction and/or analysis.

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## ACFDR STEERING COMMITTEE MEMBERSHIP 2025

| Steering Committee Members | Role/Specialisation | Institution/Association |
|---|---|---|
| Professor Susannah Ahern | Coordinating Investigator/Academic Lead | Monash University, VIC |
| Professor Peter Wark | Clinical Lead ACFDR/CF Adult Physician | The Alfred Hospital, VIC |
| Professor Andre Schultz | Deputy Lead/CF Paediatric Physician | Perth Children's Hospital, WA |
| Dr Katherine Frayman | Deputy Lead/CF Paediatric Physician | Royal Children's Hospital, VIC |
| A/Professor Rasa Ruseckaite | Deputy Monash Academic Lead ACFDR | Monash University, VIC |
| Dr Jo Armstrong | CEO Cystic Fibrosis Australia | Cystic Fibrosis Australia |
| Dr Siobhain Mulrennan | CF Adult Physician | Sir Charles Gairdner Hospital, WA |
| Dr Judith Morton | CF Adult Physician | Royal Adelaide Hospital, SA |
| Dr Tonia Douglas | CF Paediatric Physician | Queensland Children's Hospital, QLD |
| Dr Bernadette Prentice | CF Paediatric Physician | Sydney Children's Hospital, NSW |
| Dr Nathan Ward | Physiotherapist | Royal Adelaide Hospital, SA |
| Sue Morey OAM | Nurse Practitioner | Sir Charles Gairdner Hospital, WA |
| Caz Boyd | Consumer Representative | WA |
| Sophie Worthington | Consumer Representative | WA |
| Penny Jones | Consumer Representative | NSW |
| Rebecca Edwards | Consumer Representative | NSW |
| Charlotte Roseby | Consumer Representative | VIC |
| Kate Ellis | Consumer Representative | VIC |

## LIST OF PARTICIPATING SITES

| Site | |
|---|---|
| Centenary Hospital for Women & Children (CHW) | Paediatric |
| Gold Coast University Hospital (GCH) | Adult |
| Gosford Hospital (GOS) | Paediatric and Adult |
| John Hunter Children's Hospital (JHC) | Paediatric |
| John Hunter Hospital (JHH) | Adult |
| Launceston General Hospital (LGH) | Paediatric |
| Mater Hospital (MAH) | Adult |
| Monash Medical Centre (MMC) | Paediatric and Adult |
| North West Regional Hospital (BUR) | Paediatric |
| Perth Children's Hospital (PCH) | Paediatric |
| Queensland Children's Hospital (QCH) | Paediatric |
| Royal Adelaide Hospital (RAH) | Adult |
| Royal Children's Hospital (RCH) | Paediatric |
| Royal Hobart Hospital (RHH) | Paediatric and Adult |
| Royal Prince Alfred Hospital (RPA) | Adult |
| Sir Charles Gairdner Hospital (SCG) | Adult |
| Sydney Children's Hospital (SCH) | Paediatric |
| The Alfred Hospital (ALF) | Adult |
| The Canberra Hospital (CHA) | Adult |
| The Children's Hospital, Westmead (CHW) | Paediatric |
| The Prince Charles Hospital (PCH) | Adult |
| Westmead Hospital (WMH) | Adult |
| Women's and Children's Hospital (WCH) | Paediatric |

## ACFDR COORDINATING CENTRE, MONASH UNIVERSITY

The ACFDR coordinating team encourages contact regarding all registry related activities and operations, including access to data through the email account below.

Email: med–acfdregistry@monash.edu
Registry Academic Lead: Professor Susannah Ahern
Deputy Monash Academic Lead: A/Professor Rasa Ruseckaite
Principal Data Science Lead: Dr Ahmad Reza Pourghaderi
Registry Coordinator: Ms Isabella Hall
Phone: +61 (0) 3 9903 1656

## ACCESS TO REGISTRY DATA

Requests for information from the ACFDR are welcome.
Application should be made to the ACFDR Coordinating Centre, Monash University.
Email: med–acfdregistry@monash.edu

## ELECTRONIC DATA CAPTURE

Study data were collected and managed using REDCap electronic data capture tool hosted and managed by Helix (Monash University).[1, 2]

REDCap (Research Electronic Data Capture) is a secure, web-based software platform designed to support data capture for research studies, providing 1) an intuitive interface for validated data capture; 2) audit trails for tracking data manipulation and export procedures; 3) automated export procedures for seamless data downloads to common statistical packages; and 4) procedures for data integration and interoperability with external sources.

1. PA Harris, R Taylor, R Thielke, J Payne, N Gonzalez, JG. Conde, Research electronic data capture (REDCap) – A metadata-driven methodology and workflow process for providing translational research informatics support, J Biomed Inform. 2009 Apr;42(2):377-81..
2. PA Harris, R Taylor, BL Minor, V Elliott, M Fernandez, L O'Neal, L McLeod, G Delacqua, F Delacqua, J Kirby, SN Duda, REDCap Consortium, The REDCap consortium: Building an international community of software partners, J Biomed Inform. 2019 May 9 [doi: 10.1016/j.jbi.2019.103208]

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MONASH University

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